Adbry, Dupixent
Prior Authorization Guidelines

Affected Medication(s)

- Dupixent (dupilimab) subcutaneous solution
- Adbry (tralokinumab) subcutaneous solution

FDA Approved Indication(s)

- Dupixent:
  - Treatment of patients ages 6 months and older with moderate-to-severe atopic dermatitis whose disease is not adequately controlled with topical prescription therapies or when those therapies are not advisable
  - As an add-on maintenance treatment in patient with moderate-to-severe asthma ages 6 years and older with an eosinophilic phenotype or with oral corticosteroid dependent asthma
  - As an add-on maintenance treatment in adult patients with inadequately controlled chronic rhinosinusitis with nasal polyposis (CRSwNP)
  - Treatment of pediatric patients 12 years and older weighing at least 40 kg with eosinophilic esophagitis (EoE)
  - Treatment of adult patients with prurigo nodularis
- Adbry:
  - Treatment of moderate-to-severe atopic dermatitis in adult patients whose disease is not adequately controlled with topical prescription therapies or when those therapies are not advisable

Dosing

- Refer to respective package insert for dosing information

Initial Authorization Criteria

1. Is the request for continuation of the same therapy for the same condition?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Will the requested drug be used concurrently with other biologic therapy? (Examples: Actemra, Enbrel, Cimzia, Humira, Otezla, Cosentyx, etc.)
   a. If yes, clinical review required
   b. If no continue to #4
4. What is the diagnosis that the drug is being requested for?
   a. Atopic dermatitis, continue to corresponding criteria
   b. Moderate to severe asthma, continue to corresponding criteria
   c. Chronic rhinosinusitis, continue to corresponding criteria
   d. Eosinophilic esophagitis, continue to corresponding criteria
   e. Prurigo nodularis, continue to corresponding criteria

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f. Other indication, clinical review required

Atopic Dermatitis

1. Does the member have at least 10% body surface area involvement? (Provide documentation of body surface area affected)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the request for Dupixent (dupilimub)?
    a. If yes, continue to #4
    b. If no, continue to #3
3. Does the member have a documented trial with insufficient response, intolerance or contraindication to Dupixent (dupilimab)? (Provide documentation to support insufficient response, intolerance, and/or contraindication)
    a. If yes, continue to #4
    b. If no, continue to #5
4. Does the member have a documented trial with insufficient response, or intolerance, or contraindication to TWO of the following therapies: topical agent (high-potency steroid/calcineurin inhibitor), immunomodulatory agent (azathioprine, cyclosporine, methotrexate, mycophenolate), or phototherapy? (Provide supporting documentation of all therapies tried)
    a. If yes, continue to #8
    b. If no, clinical review required
5. Does the member have a documented trial with insufficient response, or intolerance, or contraindication to both a high-potency topical steroid (i.e. clobetasol 0.05%, fluocinonide 0.1%, halobetasol 0.05%, or betamethasone dipropionate 0.05%) and a topical calcineurin inhibitor (i.e. tacrolimus or pimecrolimus)? (Provide supporting documentation of all therapies tried)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a documented trial with insufficient response or intolerance or contraindication to at least one systemic immunomodulatory agent (i.e. azathioprine, cyclosporine, methotrexate, or mycophenolate)? (Provide supporting documentation of all therapies tried)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have a documented trial with insufficient response, intolerance, or contraindication to phototherapy? (Provide documentation to support insufficient response, intolerance, and/or contraindication)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Is the requested drug being prescribed by or in consultation with a dermatologist, allergist, or immunologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Moderate to severe asthma

1. Does the member have a baseline forced expiratory volume in 1 second (FEV1) less than 80% of predicted normal for adults or FEV1 of less than 90% in adolescents despite adherence to asthma maintenance regimen? (Provide baseline FEV1)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member experienced at least one severe exacerbation within the last 12 months that requires an urgent care visit, or hospitalization OR at least two exacerbations requiring oral corticosteroids despite adherence to asthma maintenance regimen? (Provide supporting documentation of past medical history)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a baseline eosinophil count greater than 150 cells/mcL?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member currently on a high-dose inhaled corticosteroids (ICS)?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the member currently on a long-acting beta agonist (LABA)? (Provide documentation of medication history)
    a. If yes, continue to #7
    b. If no, continue to #6
6. Does the member have a history of intolerance or contraindication to LABA and is on a leukotriene modifier (LTRA)? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the requested treatment dose appropriate?
    a. If yes, continue to #8
    b. If no, clinical review required
8. Is Dupixent (dupilumab) being prescribed by or in consultation with an allergist or pulmonologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Chronic rhinosinusitis with nasal polyposis

1. Does the member bilateral nasal polyposis and chronic symptoms of sinusitis?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member had history of sinus surgery but developed recurrent refractory disease?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have documented treatment failure with systemic corticosteroids after the sinus surgery?

a. If yes, continue to #4
b. If no, clinical review required

4. Is the requested treatment dose appropriate?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is Dupixent (dupilumab) being prescribed by or in consultation with an otolaryngologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Eosinophilic Esophagitis (EoE)

1. Is the member 12 years of age or older?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a diagnosis of eosinophilic esophagitis (confirmed by endoscopic biopsy) defined as greater than or equal to 15 intraepithelial eosinophils per high-power field (eos/hpf)? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member currently have on-going symptoms of dysphagia (pain when swallowing, drooling, sensation of food getting stuck in the throat, chest pain) despite dietary modifications? (Provide documentation of symptoms)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the patient weigh greater than or equal to 40 kg? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a documented trial with an insufficient response, intolerance or contraindication to a minimum 8-week trial of at least one proton pump inhibitor (omeprazole, lansoprazole, pantoprazole, etc.)? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a documented trial with insufficient response, intolerance or contraindication to at least one topical (budesonide, fluticasone) or oral glucocorticoid therapy? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is Dupixent (dupilumab) being prescribed or in consultation with a gastroenterologist or other appropriate specialist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Prurigo Nodularis

1. Is the member 18 years of age or older?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have at least 10% body surface area involvement? (Provide documentation of body surface area affected)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a documented trial with insufficient response, or intolerance, or contraindication to TWO of the following therapies: topical agent (high-potency steroid/calcineurin inhibitor), immunomodulatory agent (cyclosporine, methotrexate), or phototherapy? (Provide supporting documentation of all therapies tried)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the requested drug being prescribed by or in consultation with a dermatologist, allergist, or immunologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Will the requested medication be used with other biologic therapy? (Examples: Enbrel, Actemra, Cimzia, Simponi, Orencia, Taltz, Cosentyx, Otezla, etc)
    a. If yes, clinical review required
    b. If no, continue to #3
3. Were updated chart notes (dated within 1 year) provided with documentation of significant clinical response to therapy? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the requested drug being prescribed by or in consultation with a dermatologist, allergist, immunologist, or other appropriate specialist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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Effective Date: 1/1/2019, 1/1/20, 9/1/21, 3/1/22, 9/1/22, 11/20/23

References:

1. Dupixent (dupilumab) [Prescribing Information]. Tarrytown, NY: Sanofi-aventis U.S. LLC. December 2021.
2. Dupixent. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed September 14, 2018.
3. Adbry (tralokinumab-ldrm) [Prescribing Information]. Ballerup, Denmark: LEO Pharma A/S. May 2023.
4. Weston MD, Howe MD. Treatment of atopic dermatitis (eczema). Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed September 14, 2018.
5. Halilos MD, Holbrook MD. Chronic rhinosinusitis: Management. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed November 6, 2019
6. Cloutier MM, et al. 2020 Focused Updates to the Asthma Management Guidelines: A Report from the National Asthma Education and Prevention Program Coordinating Committee Expert Panel Working Group. J Allergy Clin Immunol. 2020 Dec;146(6):1217-1270. doi: 10.1016/j.jaci.2020.10.003. Erratum in: J Allergy Clin Immunol. 2021 Apr;147(4):1528-1530. PMID: 33280709; PMCID: PMC7924476.
7. Hirano, Ikuo, et al. "AGA institute and the joint task force on allergy-immunology practice parameters clinical guidelines for the management of eosinophilic esophagitis." Gastroenterology 158.6 (2020): 1776-1786.
8. Watsky, K. Prurigo Nodularis. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed August 29, 2023.

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Allergen Extracts
Prior Authorization Guidelines

Affected Medication(s)

- Grastek sublingual tablet
- Odactra sublingual tablet
- Oralair sublingual tablet
- Ragwitek sublingual tablet

FDA Approved Indication(s)

- **Grastek:** As an immunotherapy for the treatment of grass pollen-induced allergic rhinitis with or without conjunctivitis confirmed by positive skin test or *in vitro* testing for pollen-specific IgE antibodies for Timothy grass or cross-reactive grass pollens for patients between 5 and 65 years of age
- **Odactra:** As an immunotherapy for house dust mite (HDM)-induced allergic rhinitis, with or without conjunctivitis, confirmed by *in vitro* testing for IgE antibodies to *Dermatophagoides farinae* or *Dermatophagoides pteronyssinus* house dust mites, or skin testing to licensed house dust mite allergen extracts for adult patients between 18 and 65 years of age
- **Oralair:** As an immunotherapy for the treatment of grass pollen-induced allergic rhinitis with or without conjunctivitis confirmed by positive skin test or *in vitro* testing for pollen-specific IgE antibodies for any of the five grass species contained in this product for patients between 5 and 65 years of age
- **Ragwitek:** As an immunotherapy for the treatment of short ragweed pollen-induced allergic rhinitis, with or without conjunctivitis, confirmed by positive skin test or *in vitro* testing for pollen-specific IgE antibodies for short ragweed pollen for patients between 5 and 65 years of age

Dosing

- Refer to corresponding package insert for specific dosing recommendations

Authorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member remain symptomatic despite treatment with a nasal steroid AND oral antihistamine at the maximum indicated doses? (Provide supporting documentation of relevant past/current medication history and symptom history)
    a. If yes, continue to #3
    b. If no, clinical review required
3. For Grastek, Oralair, and Ragwitek: Will the treatment begin prior to the start of allergy season and continue throughout the allergy season? (12 weeks prior for Grastek or Ragwitek, 16 weeks prior for Oralair)
    a. If yes, continue to #4
    b. If no, clinical review required
    c. If not applicable, continue to #4

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4. Does the member have a positive skin test or *in vitro* testing for pollen specific IgE antibodies to the corresponding allergen listed below? (Provide relevant test results for review)

    - Grastek: Timothy grass or cross-reactive grass
    - Odactra: *Dermatophagoides farinae* or *Dermatophagoides pteronyssinus* house dust mites
    - Oralair: Sweet vernal, Orchard, perennial Rye, Timothy, or Kentucky blue grass
    - Ragwitek: Short Ragweed

    a. If yes, continue to #5
    b. If no, clinical review required

5. Does the member have any of the following contraindications to the requested treatment?

    - Severe, unstable, or uncontrolled asthma
    - History of any severe systemic allergic reaction or severe local reaction after taking any sublingual allergen immunotherapy
    - History of eosinophilic esophagitis
    - Hypersensitivity to any of the inactive ingredients

    a. If yes, clinical review required
    b. If no, continue to #6

6. Is the requested drug Grastek?

    a. If yes, go to #7
    b. If no, continue to #8

7. Has the member completed 3 consecutive years of therapy (including intervals between grass pollen seasons)?

    a. If yes, clinical review required
    b. If no, continue to #8

8. Is the treatment being prescribed by, or in consult with an allergist, immunologist, or otolaryngologist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Odactra (dermatophagoides pteronyssinus and dermatophagoides farinae) [Prescribing Information]. Horsholm, Denmark: ALK-Abello A/S. March 2020.
2. Oralair (anthoxanthum odoratum pollen, dactylis glomerata pollen, lolium perenne pollen, phleum pratense pollen, and poa pratensis) [Prescribing Information]. Antony, France: Stallergenes. January 2021.

Last Reviewed: 10/17/18, 11/20/19, 7/21/21, 9/21/22, 9/15/23
Effective Date: 1/1/19, 1/1/20, 9/1/21

3. Ragwitek (ambrosia artemisiifolia pollen) [Prescribing Information]. Horsholm, Denmark: ALK-Abello A/S. April 2021.
4. Grastek (timothy grass pollen allergen extract) [Prescribing Information]. Horsholm, Denmark: ALK-Abello A/S. August 2020.
5. Greenhawt, M, Oppenheimer J, Nelson M, et al. Sublingual immunotherapy: a focused allergen immunotherapy practice parameter update. Annals of Allergy, Asthma & Immunology 118(2017) 276-282. https://doi.org/10.1016/j.anai.2016.12.009. Accessed October 2018.

Amyotrophic Lateral Sclerosis (ALS) Agents
Prior Authorization Guidelines

Affected Medication(s)

- Radicava ORS (edavarone) oral suspension
-

FDA Approved Indication(s)

- Treatment of amyotrophic lateral sclerosis (ALS) in adults

Dosing

- Radicava: 105 mg orally once daily for 14 days, followed by 14-day drug-free period, then 105 mg once daily for 10 days within a 14-day period, followed by a 14-day drug-free period
-

Initial Authorization Criteria

1. Is the request for continuation of Radicava ORS (edavarone) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member at least 18 years of age?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment prescribed by or in consultation with a neurologist?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member been diagnosed with ALS defined by the revised El Escorial criteria, Awaji criteria, or Gold Coast criteria? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the patient currently on riluzole therapy or have a documented contraindication or intolerance to riluzole? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have a disease duration of less than or equal to 2 years? (Provide supporting documentation)

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a. If yes, continue to #8
b. If no, clinical review required

8. Does the member have a forced vital capacity (%FVC) of greater than or equal to 80% of predicted? (Provide supporting documentation)
    a. If yes, continue to #9
    b. If no, clinical review required
9. Does the member have a baseline documentation of the revised ALS Functional Rating Scale (ALSFRS-R) score with greater than 2 points in each of the 12 items? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member experienced a documented positive response to therapy? (ex. no tracheostomy or assisted ventilation required) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Miller, Robert G., et al. "Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology." Neurology 73.15 (2009): 1218-1226.
2. EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, et al. "EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)–revised report of an EFNS task force." European journal of neurology 19.3 (2012): 360-375.
3. Radicava ORS (edaravone) oral suspension [package insert]. Jersey City, NJ: Mitsubishi Tanabe Pharma Corporation; 2023

Last Reviewed: 11/16/22, 5/19/23, 5/17/24
Effective Date:1/1/23, 7/15/23, 8/1/24

4. Drugs@FDA: FDA Approved Drug Products. 2023. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 18 Apr. 2023].
5. Writing Group; Edaravone (MCI-186) ALS 19 Study Group. Safety and efficacy of edaravone in well defined patients with amyotrophic lateral sclerosis: a randomised, double-blind, placebo-controlled trial. Lancet Neurol. 2017;16(7):505-512.

Amicar® (aminocaproic acid)
Prior Authorization Guidelines

Affected Medication(s)

- Amicar oral solution
- Amicar oral tablet
- Aminocaproic acid oral solution
- Aminocaproic acid oral tablet

FDA Approved Indication(s)

- Enhancing hemostasis when fibrinolysis contributes to bleeding
- Hemorrhage secondary to various disorders including aplastic anemia, abruption placentae, hepatic cirrhosis, and neoplastic diseases

Dosing

- 4 to 5 g administered during the first hour of treatment, followed by a continuing rate of 1 to 1.25 g per hour, continued for about 8 hours or until the bleeding situation has been controlled (Maximum daily dose: 30g)

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a trial with inadequate response, intolerance, or contraindication to tranexamic acid? (Provide documentation of trial with inadequate response, intolerance, or contraindication)
    a. If yes, approve for 1 month unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Amicar. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed October 14, 2023.
2. Amicar (aminocaproic acid) [Prescribing Information]. Lake Forest, IL: Clover Pharmaceuticals, Corp. June 2022.

Last Reviewed: 11/21/18, 11/20/19, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 1/1/20, 9/1/21, 12/20/23

Anti-Asthmatic Agent Policy
Prior Authorization Guidelines

Affected Medication(s)

- Fasenra (benralizumab) subcutaneous solution
- Nucala (mepolizumab) auto-injector solution
- Nucala (mepolizumab) syringe
- Tezspire (tezepelumab) auto-injector solution
- Tezspire (tezepelumab) syringe
- Xolair (omalizumab) subcutaneous solution

FDA Approved Indication(s)

Fasenra

- As add-on maintenance treatment of patients 12 years of age and older with severe asthma, eosinophilic phenotype

Nucala

- As an add-on maintenance therapy for patients 6 years of age and older with severe asthma, eosinophilic phenotype
- For the treatment of adult patients with eosinophilic granulomatosis with polyangiitis (EGPA)
- For the treatment of adult and pediatric patients aged 12 years and older with hypereosinophilic syndrome (HES) for ≥6 months without an identifiable non-hematologic secondary cause.

Tezspire

- As add-on maintenance treatment of adult and pediatric patients aged 12 years and older with severe asthma

Xolair

- For patients 6 years of age and older with moderate to severe persistent asthma who have a positive skin test or *in vitro* reactivity to a perennial aeroallergen and whose symptoms are inadequately controlled with inhaled corticosteroids
- For the treatment of nasal polyps in adult patients 18 years of age and older with inadequate response to nasal corticosteroids
- For the treatment of adults and adolescents 12 years of age and older with chronic idiopathic urticaria who remain symptomatic despite H1 antihistamine treatment

Dosing

- Reference dosing recommendations in package insert

Initial Authorization Criteria

1. Is the request for continuation of injectable anti-asthmatic therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication and age? (Provide documentation of diagnosis)
    a. If yes, continue to #3

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b. If no, clinical review required

3. Which diagnosis the injectable anti-asthmatic being requested for?

   a. Moderate to severe persistent asthma, continue to corresponding criteria
   b. Chronic Idiopathic Urticaria (CIU), continue to corresponding criteria
   c. Eosinophilic granulomatosis with polyangiitis (EGPA), continue to corresponding criteria
   d. Hypereosinophilic syndrome (HES), continue to the corresponding criteria
   e. Nasal polyps, continue to corresponding criteria

<u>Moderate to severe persistent asthma</u>

1. Does the member have a reduced lung function at baseline defined as pre-bronchodilator FEV1 less than 80%? (Provide supporting documentation)

   a. If yes, continue to #2
   b. If no, clinical review required

2. Is the member currently on a high-dose inhaled corticosteroid (ICS)? (Provide documentation of medication history)

   c. If yes, continue to #3
   d. If no, clinical review required

3. Is the member on a long acting beta agonist (LABA)? (Provide documentation of medication history)

   a. If yes, continue to #5
   b. If no, continue to #4

4. Does the member have a history of intolerance or contraindication to LABA and is on a leukotriene modifier (LTRA)? (Provide supporting documentation)

   a. If yes, continue to #5
   b. If no, clinical review required

5. Has the member experienced 2 or more exacerbations within the last 12 months that required systemic steroid treatment, an urgent care visit, or hospitalization despite adherence to asthma maintenance therapy? (Provide documentation of exacerbation history)

   a. If yes, continue to #6
   b. If no, clinical review required

6. Does the member have eosinophilic phenotype asthma defined as baseline eosinophil count greater than equal to 150 cells/ul within 12 months? (Provide supporting documentation)

   a. If yes, continue to #11
   b. If no, continue to #7

7. Does the member have a baseline immunogloblin E (IgE) level between 30 and 700 IU/mL? (Provide baseline IgE lab results)

   a. If yes, continue to #8
   b. If no, continue to #10

8. Does the member have body weight less than 150 kg?

    a. If yes, continue to #9
    b. If no, continue to #10

9. Does the member have a positive skin test or *in vitro* reactivity to a perennial aeroallergen? (Provide allergen test results)

    a. If yes, continue to #11
    b. If no, continue to #10

10. Is the request for Tezspire?

    a. If yes, continue to #11
    b. If no, clinical review required

11. Is the requested injectable anti-asthmatic to be used in combination with current asthma treatment regimen? (Provide documentation of planned treatment regimen)

    a. If yes, continue to #12
    b. If no, clinical review required

12. Is the injectable anti-asthmatic prescribed by, or in consult with, an allergist or pulmonologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Chronic Idiopathic Urticaria (CIU)

1. Is the member 12 years of age or older?

    a. If yes, continue to #2
    b. If no, clinical review required

2. Did the member have an insufficient response to TWO high dose antihistamines with a duration of at least 2 weeks each? (Provide documentation of past medications used along with response to therapy)

    a. If yes, continue to #5
    b. If no, continue to #3

3. Does the member have a contraindication to antihistamines? (Provide supporting documentation)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Did the member have an insufficient response or contraindication to leukotriene modifiers (LTRA)? (Provide supporting documentation)

    a. If yes, continue to #6
    b. If no, clinical review required

5. Did the member have an insufficient response to combination therapy with high dose second-generation antihistamines with at least one of the following for a minimum of 4 weeks trial? (Provide documentation of past medications used along with response to therapy)

- A H2-antagonist
- A 1st generation antihistamine at bedtime
- A leukotriene modifier (LTRA)

a. If yes, continue to #6
b. If no, clinical review required

6. Does the member have an insufficient response, intolerance, or contraindication to hydroxyzine or doxepin? (Provide documentation of response to therapy)

a. If yes, continue to #7
b. If no, clinical review required

7. Is Xolair (omalizumab) prescribed by, or in consult with, an allergist or dermatologist?

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

<u>Eosinophilic granulomatosis with polyangiitis (EGPA)</u>

1. Does the member have relapsing or refractory eosinophilic granulomatosis with polyangiitis with at least one of the following characteristics?

- Asthma
- Sinusitis
- Pulmonary infiltrates
- Neuropathy
- Eosinophilic vasculitis of one or more end-organs

a. If yes, continue to #2
b. If no, clinical review required

2. Does the member have relapsing or refractory disease despite treatment with TWO separate trials of the following therapies in combination glucocorticoid: azathioprine, methotrexate, leflunomide?

a. If yes, continue to #3
b. If no, clinical review required

3. Has the member been established on stable dose of oral steroid therapy for 4 weeks or more?

a. If yes, continue to #4
b. If no, clinical review required

4. Does the member have a baseline blood eosinophil level of 10% and higher OR an absolute eosinophil count of 1000 cells/ul or higher? (Provide supporting lab for review)

a. If yes, continue to #5
b. If no, clinical review required

5. Is the requested medication being prescribed by or in consult with a specialist who is experienced in treating EGPA?

a. If yes, approve for 6 months, unless otherwise specified
b. If no, clinical review required

Hypereosinophilic Syndrome (HES)

1. Does the member have documentation of primary HES without non-hematologic secondary causes? (i.e. drug hypersensitivity, parasitic helminth infection, HIV infection, non-hematologic malignancy)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a blood eosinophil count of 1,000 cells/mcL or higher?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member trialed HES therapy consisting of chronic/episodic oral corticosteroids, immunosuppressive, or cytotoxic therapy?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has the member experienced at least 2 HES flares within the past 12 months? (defined as HES-related worsening of clinical symptoms or blood eosinophil counts requiring an escalation in therapy)
    a. If yes, continue to #3
    b. If no, clinical review required
5. Is the requested medication being prescribed by, or in consult with, a specialist who is experienced in treating HES?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Nasal Polyps

1. Does the member have documentation of bilateral nasal polyps confirmed by endoscopy with a total nasal polyp score (NPS) of 5 or greater and NPS score of 2 or greater per nostril? (NPS range 0-4 per nostril, 0-8 total)
    - 0= no polyps
    - 1=small polyps in the middle meatus not reaching below the inferior border of the middle turbinate
    - 2=polyps reaching below the lower border of the middle turbinate
    - 3=large polyps reaching the lower border of the inferior turbinate or polyps medial to the middle turbinate
    - 4=large polyps causing complete obstruction of the inferior nasal cavity)

    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have documented moderate to severe nasal congestion?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member previously trialed nasal corticosteroids at the maximum recommended dose?

a. If yes, continue to #4
b. If no, clinical review required

4. Is the requested medication being prescribed by, or in consultation with, an ENT specialist?

a. If yes, approve for 6 months
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

a. If yes, continue to #2
b. If no, clinical review required

2. Which diagnosis is the requested medicaiton being used for?

a. Moderate to severe persistent asthma, please see corresponding criteria
b. Chronic Idiopathic Urticaria (CIU), please see corresponding criteria
c. Eosinophilic granulomatosis with polyangiitis (EGPA), continue to corresponding criteria
d. Hyperesosinophilic Syndrome, continue to corresponding criteria
e. Nasal polyps, continue to corresponding criteria

<u>Moderate to severe persistent asthma</u>

1. Is the member adherent to asthma maintenance therapy defined as a high-dose ICS plus a LABA or LTRA?

a. If yes, continue to # 2
b. If no, clinical review required

2. Is the member responding positively to therapy defined as reduction in exacerbations, reductions in corticosteroid dose, or improvement in FEV1 compared to baseline? (Provide supporting documentation)

a. If yes, continue to #3
b. If no, clinical review required

3. Is the requested medication being prescribed by, or in consult with an allergist or pulmonologist?

a. If yes, approve for 12 months unless otherwise specified
b. If no, clinical review required

<u>Chronic Idiopathic Urticaria (CIU)</u>

1. Is the member responding positively to therapy defined as reduction in symptoms compared to baseline? (Provide supporting documentation)

a. If yes, continue to #2
b. If no, clinical review required

2. Is Xolair (omalizumab) prescribed by, or in consult with, an allergist or dermatologist?

a. If yes, approve for 12 months unless otherwise specified
b. If no, clinical review required

<u>Eosinophilic granulomatosis with polyangiitis (EGPA)</u>

1. Is the member responding positively to therapy defined as reduction in relapse and or ability to taper down on glucocorticoid use? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is Nucala (omalizumab) prescribed by, or in consult with, an allergist or dermatologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Hyperesosinophilic Syndrome

1. Is the member responding positively to therapy defined by an improvement in symptoms and/or a reduction in the frequency of exacerbation? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the medication being prescribed by, or in consultation with, a specialist who is experienced in treating HES?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Nasal Polyps

1. Is the member responding positively to therapy defined as an improvement in Nasal Polyps Score (NPS) and a decrease in the severity of nasal congestion?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the medication being prescribed by, or in consult with, an ENT specialist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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Last Reviewed: 11/7/18, 3/18/20, 5/19/21, 7/20/22, 5/19/23
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Apokyn®, Inbrija® (levodopa) Prior Authorization Guidelines

Affected Medication(s)

- Apokyn subcutaneous solution
- Apomorphine subcutaneous solution
- Inbrija inhalation powder

FDA Approved Indication(s)

- For the acute, intermittent treatment of hypomobility, "off" episodes ("end-of-dose wearing off" and unpredictable "on/off" episodes) in patients with advanced Parkinson's disease

Dosing

- Apokyn: Initially 0.2 ml (2 mg) titrated up to a maximum of 0.6 ml (6 mg)
- Inbrija:
  - 84mg (contents of two capsules) inhaled, as needed, for OFF symptoms up to five times a day
  - The maximum dose per OFF period is 84mg, and the maximum recommended daily dosage of Inbrija is 420mg

Initial Authorization Criteria

1. Is the request for continuation of Apokyn (apomorphine hydrochloride) or Inbrija (levodopa) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the medication being requested for an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have a diagnosis of advanced Parkinson's disease? (Provide supporting documentation)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is the member 18 years of age or older?
   a. If yes, continue to #5
   b. If no, clinical review required
5. Does the member have significant hypomobility or "off" episodes that last at least 2 hours? (Provide supporting documentation)
   a. If yes, continue to #6
   b. If no, clinical review required
6. Is the member on a maximally tolerated dose of levodopa AND one additional agent from one of the following classes? (Provide relevant medication history)
   - Dopamine agonist (Examples include: ropinirole, rotigotine)

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- Catechol-O-methyl transferase (COMT) inhibitor (Examples include: entacapone, tolcapone)
- Monoamine oxidase type B inhibitor (MAO-B) (Examples include: rasagiline, safinamide, selegiline)

a. If yes, continue to #7
b. If no, clinical review required

7. Is the request for Apokyn or apomorphine?

a. If yes, continue to #8
b. If no, continue to #9

8. Does the member have a trial with inadequate response to, intolerance, or contraindication to Inbrija? (Provide supporting documentation)

a. If yes, continue to #9
b. If no, clinical review required

9. Is the treatment being prescribed by, or in consultation with, a neurologist?

a. If yes, approve for 12 months unless otherwise specified
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

a. If yes, continue to #2
b. If no, clinical review required

2. Is the member demonstrating positive clinical response to therapy defined by a decrease in frequency of hypomobility or "off" episodes? (Provide supporting documentation)

a. If yes, continue to #3
b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with, a neurologist?

a. If yes, approve for 12 months reauthorization unless otherwise specified
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

Last Reviewed: 10/17/18, 1/21/20, 11/18/20, 11/17/21, 1/20/23, 11/17/23
Effective Date: 1/1/19, 2/15/20, 12/15/20, 1/1/22, 3/15/23, 12/20/23

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Aranesp® (darbepoetin alfa)
Prior Authorization Guidelines

Affected Medication(s)

- Aranesp subcutaneous injection solution

FDA Approved Indication(s)

- Treatment of anemia due to chronic kidney disease (CKD), including patients on dialysis and patients not on dialysis
- Treatment of anemia in patients with non-myeloid malignancies where anemia is due to the effect of concomitant myelosuppressive chemotherapy, and upon initiation, there is a minimum of two additional months of planned chemotherapy

Dosing

- Refer to package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of Aranesp (darbepoetin alfa) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the medication being requested for an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Have serum ferritin, transferrin saturation, hematocrit (Hct), and hemoglobin (Hb) lab values been completed within 30 days of planned administration? (Provide labs for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a serum ferritin ≥100 ng/mL (mcg/L) and transferrin saturation (TSAT) ≥20%? (Provide labs for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the member's hemoglobin (Hb) <10 g/dL and/or Hematocrit (Hct) <30%?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Have other causes of anemia (e.g. hemolysis, bleeding, vitamin deficiency, etc.) been ruled out?
    a. If yes, continue to #7
    b. If no, clinical review required
7. Which indication is Aranesp (darbepoetin alfa) being requested for? (Record submitted diagnosis and review all criteria based on the submitted diagnosis)
    a. Anemia secondary to myelodysplastic syndrome (MDS), continue to corresponding criteria

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b. Anemia secondary to Myeloproliferative Neoplasms (MPN) – Myelofibrosis, continue to corresponding criteria
c. Anemia secondary to chemotherapy treatment, continue to corresponding criteria
d. Anemia secondary to chronic kidney disease (non-dialysis patients), approve for 3 months unless otherwise specified
e. Other indication, continue to corresponding criteria

Anemia secondary to myelodysplastic syndrome (MDS)

1. Does the member have symptomatic anemia? (Examples include: exertional dyspnea, dyspnea at rest, fatigue, lethargy, confusion, etc.) (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is member's endogenous serum erythropoietin level ≤500 mUnits/mL? (Provide lab for review)

    a. If yes, approve for 45 days unless otherwise specified
    b. If no, clinical review required

Anemia secondary to Myeloproliferative Neoplasms (MPN) – Myelofibrosis

1. Is member's endogenous serum erythropoietin level <500 mUnits/mL? (Provide lab for review)

    a. If yes, approve for 45 days unless otherwise specified
    b. If no, clinical review required

Anemia secondary to chemotherapy treatment

1. Is the member receiving concurrent myelosuppressive chemotherapy for non-myeloid malignancies?

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the therapy intention of the chemotherapy curative?

    a. If yes, clinical review required
    b. If no, continue to #3

3. Are there two or more additional months of planned chemotherapy remaining? (Provide documentation of treatment plan)

    a. If yes, approve for 6 months or until completion of chemotherapy course, whichever is less
    b. If no, clinical review required

Other Indications

1. Is the requested use supported by major compendia? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member tried and had an inadequate response OR dose the member have a contradiction to ALL standard treatment options for the requested indication (Provide all prior treatment history, contraindication if appropriate, and treatment plan)

a. If yes, approve for 45 days unless otherwise specified
b. If no, clinical review required

Reauthorization Criteria

1. Was the last dose of Aranesp (darbepoetin alfa) less than 60 days ago? (Provide date of last dose)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within 1 year) provided with documentation of significant clinical response to therapy? (Provide updated clinical documentation for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there documentation of an absence of unacceptable toxicity from the drug? (Examples include pure red cell aplasia, severe allergic reactions (anaphylaxis, angioedema, bronchospasm, etc), severe cardiovascular events (stroke, myocardial infarction, congestive heart failure, thromboembolism, uncontrolled hypertension), seizures, increased risk of tumor progression/recurrence in members with cancer, etc)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Were lab values obtained within 30 days of the date of administration (unless otherwise indicated)? (Provide updated lab result for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have adequate iron stores as demonstrated by serum ferritin ≥100 ng/mL (mcg/L) and transferrin saturation (TSAT) ≥20% measured within the previous 3 months? (Provide lab result for review)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Have other causes of anemia (e.g. hemolysis, bleeding, vitamin deficiency, etc.) been ruled out?
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member meet the diagnosis and clinical requirements for at least one of the following below? (Provide supporting clinical documentation)
    - Anemia secondary to myelodysplastic syndrome (MDS) with hemoglobin (Hb) <12 g/dL and/or Hematocrit (Hct) <36%
    - Anemia secondary to myeloproliferative neoplasms (MF, post-PV myelofibrosis, post-ET myelofibrosis) with hemoglobin (Hb) <10 g/dL and/or hematocrit (Hct) <30%
    - Anemia secondary to palliative myelosuppressive chemotherapy for non-myeloid malignancies with hemoglobin (Hb) <10 g/dL and/or hematocrit (Hct) <30% and requesting Aranesp to be used concurrently with chemotherapy with minimum two additional months of therapy remaining
    - Anemia secondary to chronic kidney disease with hemoglobin (Hb) <12 g/dL and/or hematocrit (Hct) <36% in pediatric patients OR hemoglobin (Hb) <11 g/dL and/or hematocrit (Hct) <33% in adult patients
    - Use supported by major compendia

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a. If yes, approve for 45 days unless otherwise specified
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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Arcalyst® (rilonacept)
Prior Authorization Guidelines

Affected Medication(s)

- Arcalyst powder for reconstitution for subcutaneous solution

FDA Approved Indication(s)

- For treatment of Cryopyrin-Associated Periodic Syndromes (CAPS), including Familial Cold Auto-inflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) in adults and children 12 and older
- Maintenance of remission of deficiency of interleukin-1 receptor antagonist in adults and pediatric patients weighing ≥10 kg
- Treatment of recurrent pericarditis and reduction in risk of recurrence in adults and pediatric patients ≥12 years of age

Dosing

- Refer to corresponding package insert for information

Initial Authorization Criteria

1. Is the request for continuation of Arcalyst (rilonacept) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Will Arcalyst (rilonacept) be used with other biologic agent(s)? (Examples: Kineret, Ilaris, Actemra)
    a. If yes, clinical review required
    b. If no, continue to #4
4. What is the diagnosis that the medication is being requested for?
    a. Cryopyrin-Associated Periodic Syndromes (CAPS), continue to corresponding criteria
    b. Deficiency of Interleukin-1 Receptor Antagonist (DIRA), continue to corresponding criteria
    c. Pericarditis, continue to corresponding criteria
    d. Other indication, continue to corresponding criteria

Cryopyrin-Associated Periodic Syndromes (CAPS)

1. Is the genetic testing result confirming diagnosis of CAPS received? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 12 years of age or older?
    a. If yes, continue to #3

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b. If no, clinical review required

3. Is the treatment being prescribed by or in consultation with a rheumatologist?

   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

Deficiency of Interleukin-1 Receptor Antagonist (DIRA)

1. Does the member weigh at least 10kg?

   a. If yes, continue to #2
   b. If no, clinical review required

2. Has the diagnosis been confirmed by a mutation in the *IL1RN* gene?

   a. If yes, continue to #3
   b. If no, clinical review required

3. Has the member demonstrated prior clinical benefit with Kineret (anakinra)? (Examples of clinical benefit include: normalized acute phase reactants, resolution of fever, skin rash and bone pain or reduced dosage of corticosteroids)

   a. If yes, continue to #4
   b. If no, clinical review required

4. Is the treatment being prescribed by or in consultation with a rheumatologist, dermatologist or a physician specializing in the treatment of autoinflammatory disorders?

   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

Recurrent Pericarditis

1. Does the member have recurrent pericarditis as defined as at least three (3) episodes within the past year?

   a. If yes, continue to #2
   b. If no, clinical review required

2. Is the member receiving standard treatment for pericarditis (i.e. NSAIDs, colchicine, and/or systemic corticosteroids) and still symptomatic?

   a. If yes, continue to #4
   b. If no, continue to #3

3. Does the member have a documented inadequate response, intolerance, or contraindication to standard treatment options? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

   a. If yes, continue to #4
   b. If no, clinical review required

4. Is the treatment being prescribed by or in consultation with a rheumatologist or cardiologist?

   a. If yes, approve for 3 months unless otherwise specified
   b. If no, clinical review required

Other Indications

1. Has the member tried and had an inadequate response OR dose the member have a contradiction to ALL standard treatment options for the requested indication (Provide all prior treatment history, contraindication if appropriate, and treatment plan)?
    a. If yes, approve for 3 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member responding positively to therapy as defined by the following: (Provide supporting documentation)
    - Cryopyrin-Associated Periodic Syndromes (CAPS): decrease in symptoms from baseline and/or improvement in serum levels of inflammatory proteins (i.e. CRP)
    - Deficiency of Interleukin-1 Receptor Antagonist (DIRA): sustained clinical remission, resolution of fever, skin rash, and bone pain or normalized acute phase reactants
    - Recurrent Pericarditis: absence of chest pain with normalization of inflammatory biomarkers such as erythrocyte sedimentation rate and/or C-reactive protein, continued resolution of fever and bone pain
    - Other Indication: Clinical documentation confirming disease responsiveness to therapy provided

    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with the appropriate specialist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Arcalyst Prescribing Information. Tarrytown, NY: Regeneron Pharmaceuticals, Inc.; November 2020. Available at https://www.regeneron.com/sites/default/files/Arcalyst_FPI.pdf.
2. Hoffman HM, Throne ML, Amar NJ, et al. Efficacy and safety of rilonacept (interleukin-1 trap) in patients with cryopyrin-associated periodic syndromes. Arthritis and Rheum. 2008;58(8): 2443-2452.
3. Nigrovic, P. Cryoprin-associated periodic syndromes and related disorders. Orange S, Kaplan S, eds. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed May, 2020.

Last Reviewed: 10/17/18, 5/20/20, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 7/1/20, 9/1/21, 12/20/23

4. Garg M, de Jesus AA, Chapelle D, et al. Rilonacept maintains long-term inflammatory remission in patients with deficiency of the IL-1 receptor antagonist. JCI Insight. 2017 Aug 17;2(16):e94838. doi: 10.1172/jci.insight.94838.
5. Klein AL, Imazio M, Cremer P, et al. Phase 3 trial of interleukin-1 trap rilonacept in recurrent pericarditis. N Engl J Med. 2021;384(1):31-41.

Arikayce® (amikacin)
Prior Authorization Guidelines

Affected Medication(s)

- Arikayce oral suspension for inhalation

FDA Approved Indication(s)

- In adults who have limited or no alternative treatment options, for the treatment of *Mycobacterium avium* complex (MAC) lung disease as part of a combination antibacterial drug regimen in patients who do not achieve negative sputum cultures after a minimum of 6 consecutive months of a multidrug background regimen therapy

Dosing

- Once daily inhalation of the contents of one 590 mg/8.4 mL ARIKAYCE vial (590 mg of amikacin) using the Lamira Nebulizer System

Initial Authorization Criteria

1. Is the request for continuation of Arikayce (amikacin) therapy?

    a. If yes, continue to Reauthorization
    b. If no, continue to #2

2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Does the member have *Mycobacterium avium* complex (MAC) lung disease as confirmed by positive sputum culture? (Provide positive sputum culture for review)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is the member 18 years of age or older?

    a. If yes, continue to #5
    b. If no, clinical review required

5. Has the member trialed a minimum of 6-months of a multidrug background regimen (listed below) with failure confirmed by sputum culture? (Note: Failure defined as continued positive sputum culture) (Provide supporting documentation)

    - Clarithromycin/azithromycin + ethambutol + rifampin/rifabutin
    - Clarithromycin/azithromycin + ethambutol + rifampin/rifabutin + parenteral streptomycin/amikacin

    a. If yes, continue to #6
    b. If no, clinical review required

Last Reviewed: 9/18/19, 11/18/20, 9/15/21, 11/16/22, 11/17/23
Effective Date: 10/15/19, 12/15/20

6. Is the treatment being prescribed by or in consultation with an ID specialist or pulmonologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within the past 6 months) with documentation of negative sputum cultures received? (Note: Treatment should be continued until sputum cultures are consecutively negative for at least 12 months) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with an ID specialist or pulmonologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. ARIKAYCE (amikacin liposome inhalation suspension) for oral inhalation [package insert]. Midlothian, VA: PARI Pharma; July 2022.
2. Drugs@FDA: FDA Approved Drug Products. 2018. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed Nov 5. 2018].
3. Griffith, David E., et al. "An official ATS/IDSA statement: diagnosis, treatment, and prevention of nontuberculous mycobacterial diseases." American journal of respiratory and critical care medicine 175.4 (2007): 367-416.
4. Daley, Charles L., et al. "Treatment of nontuberculous mycobacterial pulmonary disease: an official ATS/ERS/ESCMID/IDSA clinical practice guideline: executive summary." Clinical Infectious Diseases 71.4 (2020): e1-e36.

Last Reviewed: 9/18/19, 11/18/20, 9/15/21, 11/16/22, 11/17/23
Effective Date: 10/15/19, 12/15/20

Banzel® (rufinamide)
Prior Authorization Guidelines

Affected Medication(s)

- Banzel (rufinamide) oral tablet
- Banzel (rufinamide) oral suspension
- rufinamide oral tablet
- rufinamide oral suspension

FDA Approved Indication(s)

- Adjunctive treatment of seizures associated with Lennox-Gastaut Syndrome (LGS) in adults and pediatric patients 1 year of age and older

Dosing

- Maximum dose of 45 mg/kg per day in two divided doses, not to exceed 3200 mg per day

Initial Authorization Criteria

1. Is the request for continuation of rufinamide (Banzel) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member currently taking at least one other antiepileptic drug with inadequate response? (i.e. valproic acid, lamotrigine, topiramate, felbamate, cannabidiol) (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Will the member continue therapy with at least one other antiepileptic drug in combination with rufinamide (Banzel)?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have familial short QT syndrome?
    a. If yes, clinical review required
    b. If no. continue to #6
6. Is the treatment being prescribed by or in consultation with a neurologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)

Last Reviewed: 9/19/2018, 11/20/19, 7/21/21, 11/16/22, 9/15/23
Effective Date: 1/1/2019, 9/1/21

a. If yes, continue to #2
b. If no, clinical review required

2. Were updated chart notes (within 1 year) provided with documentation of significant clinical response to therapy defined as at least 20% reduction in seizure frequency? (Provide documentation of decreased seizure frequency)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with a neurologist?
    a. If yes, approve for 12 months reauthorization unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Banzel (rufinamide) [Prescribing Information]. Woodcliff Lake, NJ: Eisai Inc. December 2021.
2. Banzel. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed June 25, 2018.
3. National Institute for Health and Care Excellence (NICE): Epilepsies: diagnosis and management. National Institute for Health and Care Excellence (NICE). London, United Kingdom. Available at: https://www.nice.org.uk/guidance/cg137/resources/epilepsies-diagnosis-and-management-35109515407813. Accessed June 27, 2018.
4. Biton V, Krauss G, Vasquez-Santana B, et al. A randomized, double-blind, placebo-controlled, parallel-group study of ralfinamide as adjunctive therapy for refractory partial-onset seizures. Epilepsia. 2011 Feb;52(2):234-42. Available at: https://www.ncbi.nlm.nih.gov/pubmed/20887365. Accessed August 6, 2018.
5. Ohtsuka Y, Yoshinaga H, Shirasaka Y, et al. Rufinamide as an adjuctive therapy for Lennox-Gastaut syndrome: a randomized double-blind placebo-controlled trial in Japan. Epilepsy Res. 2014 Nov;108(9):1627-36. Available at: https://www.ncbi.nlm.nih.gov/pubmed/25219353. Accessed August 6, 2018.
6. National Institute of Neurological Disorders and Stroke. Lennox-Gastaut Syndrome Information Page. Available at: https://www.ninds.nih.gov/Disorders/All-Disorders/Lennox-Gastaut-Syndrome-Information-Page.
7. Debopam Samanta, Management of Lennox-Gastaut syndrome beyond childhood: A comprehensive review, Epilepsy & Behavior, Volume 114, Part A, 2021,107612, ISSN 1525-5050, https://doi.org/10.1016/j.yebeh.2020.107612.

Last Reviewed: 9/19/2018, 11/20/19, 7/21/21, 11/16/22, 9/15/23
Effective Date: 1/1/2019, 9/1/21

Benlysta® (belimumab)
Prior Authorization Guidelines

Affected Medication(s)

- Benlysta subcutaneous solution

FDA Approved Indication(s)

- Treatment of adult patients with active, autoantibody-positive, systemic lupus erythematosus (SLE) who are receiving standard therapy
- Treatment of adults with active lupus nephritis who are receiving standard therapy

Dosing

- Systemic lupus erythematosus: 200 mg subcutaneously once weekly
- Lupus nephritis: 400 mg subcutaneously once weekly for 4 doses, then 200 mg once weekly thereafter

Initial Authorization Criteria

1. Is the request for continuation of Benlysta (belimumab) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years or older with a positive autoantibody test? (Antinuclear antibody (ANA) titer ≥1:80 OR Anti-dsDNA autoantibodies ≥30 IU/mL) (Provide test result for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have an active infection?
    a. If yes, clinical review required
    b. If no, continue to #5
5. Does the member have any of the following exclusion criteria?
    - Severe active central nervous system lupus
    - Use of other biologics or IV cyclophosphamide
    - Received a live vaccine within 30 days before starting or concurrently with Benlysta

    a. If yes, clinical review required
    b. If no, continue to #6
6. What is the indication that the medication is being requested for?
    a. Systemic lupus erythematosus (SLE), see corresponding criteria

Last Reviewed: 9/19/2018, 5/20/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/2019, 7/1/20, 1/1/22, 3/1/24

b. Lupus nephritis, see corresponding criteria

Systemic lupus erythematosus (SLE)

1. Has the member failed to respond adequately to at least TWO (2) standard therapies (anti-malarials, corticosteroids, non-steroidal anti-inflammatory drugs, immunosuppressives (excluding intravenous cyclophosphamide))? (Provide documentation of treatment history)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member have one of the following? (Provide assessment score for review)

    - Safety of Estrogens in Lupus Erythematosus National Assessment – Systemic Lupus Erythematosus Disease Activity Index (SELENA-SLEDAI) score of ≥6
    - Two or more British Isles Lupus Assessment Group (BILAG) B organ domain scores

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is Benlysta (belimumab) being prescribed by or in consultation with a rheumatologist or specialist experienced in treatment of SLE?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Lupus Nephritis

1. Does the member have active lupus nephritis Class III, IV, or V as confirmed by a kidney biopsy?

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member had an inadequate response, intolerance or contraindication to standard therapies including corticosteroids AND either cyclophosphamide or mycophenolate mofetil?

    a. If yes, continue to #3
    b. If no, clinical review required

3. Have baseline measurements for one or more of the following been provided: urine protein:creatinine ratio (uPCR), estimated glomerular filtration rate (eGFR), or urine protein? (Document baseline measurements)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is Benlysta (belimumab) being prescribed by or in consultation with a rheumatologist or specialist experienced in treatment of SLE?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Does member continue to meet initial authorization criteria as outlined above?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there documentation of an absence of unacceptable toxicity from the medication? (Examples of unacceptable toxicity include the following: depression, suicidal thoughts, serious infections, signs or symptoms of progressive multifocal leukoencephalopathy (PML), malignancy, severe hypersensitivity reaction, etc.)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is Benlysta (belimumab) being prescribed by or in consultation with a rheumatologist or specialist experienced in treatment of SLE?
    a. If yes, continue to #4
    b. If no, clinical review required
4. What is the indication that the medication is being requested for?
    a. Systemic lupus erythematosus (SLE), see corresponding criteria
    b. Lupus nephritis, see corresponding criteria

<u>Systemic lupus erythematosus (SLE)</u>

1. Were updated chart notes (within 1 year) provided with documentation of disease stability and/or improvement as indicated by one or more of the following when compared to pre-treatment baseline? (Provide updated assessment score for review)
    - Improvement in the SELENA-SLEDAI score of ≥4 points; OR
    - No new BILAG-A organ domain score or 2 new BILAG-B organ domain scores; OR
    - No worsening (<0.30-point increase) in Physician's Global Assessment (PGA) score; OR
    - Seroconverted (negative) or had a 20% reduction in autoantibody level;

    a. If yes, approve for 12 months reauthorization unless otherwise specified
    b. If no, clinical review required

<u>Lupus nephritis</u>

1. Has documentation of disease stability and/or improvement (from pre-treatment baseline) for one or more of the following measurements been provided: urine protein:creatinine ratio (uPCR), estimated glomerular filtration rate (eGFR), or urine protein? (Document updated measurements)

    a. If yes, approve for 12 months reauthorization unless otherwise specified
    b. If no, clinical review required

<u>Note:</u>

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as

medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Benlysta [package insert]. Rockville, MD; Human Genome Sciences/GlaxoSmithKline; January 2020. Accessed May 2020.
2. Boyce EG, Fusco BE. Belimumab: review of use in systemic lupus erythematosus. Clin Ther. 2012 May;34(5):1006-22. doi: 10.1016/j.clinthera.2012.02.028. Epub 2012 Mar 30.
3. Navarra SV, Guzmán RM, Gallacher AE, et al. Efficacy and safety of belimumab in patients with active systemic lupus erythematosus: a randomised, placebo-controlled, phase 3 trial. Lancet. 2011 Feb;377(9767):721-31. doi: 10.1016/SO140-6736(10)61354-2. Epub 2011 Feb 4.
4. Furie R, Petri M, Zamani O, et al. A phase III, randomized, placebo-controlled study of belimumab, a monoclonal antibody that inhibits B lymphocyte stimulator, in patients with systemic lupus erythematosus. Arthritis Rheum. 2011 Dec;63(12):3918-30. doi: 10.1002/art.30613.
5. Petri M, Orbai AM, Alarcón GS, et al. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. Arthritis Rheum. 2012 Aug;64(8):2677-86. doi: 10.1002/art.34473.
6. Furie R, Stohl W, Ginzler EM, et al. Biologic activity and safety of belimumab, a neutralizing anti-B-lymphocyte stimulator (BLyS) monoclonal antibody: a phase I trial in patients with systemic lupus erythematosus. Arthritis Res Ther. 2008;10(5):R109. doi: 10.1186/ar2506. Epub 2008 Sep 11.
7. Kim SS, Kirou KA, Erkan D. Belimumab in systemic lupus erythematosus: an update for clinicians. Ther Adv Chronic Dis. 2012 Jan;3(1):11-23. doi: 10.1177/2040622311424806.
8. Calvo-Alén J1, Silva-Fernández L, Úcar-Angulo E, et al. SER consensus statement on the use of biologic therapy for systemic lupus erythematosus. Reumatol Clin. 2013 Sep-Oct;9(5):281-96.
9. Gordon C, Amissah-Arthur MB, Gayed M, et al. The British Society for Rheumatology guideline for the management of systemic lupus erythematosus in adults. Rheumatol 2017 Oct 6. doi: 10.1093/rheumatology/kex286.
10. NICE. Belimumab for treating active autoantibody-positive systemic lupus erythematosus: Technology Appraisal Guidance [TAG397]. https://www.nice.org.uk/guidance/ta397/ Accessed November 2017.
11. Wisconsin Physician Service Insurance Corp. Local Coverage Determination (LCD): Drugs and Biologics (Non-chemotherapy) (L34741). Centers for Medicare & Medicare Services. Updated on 10/17/2017 with effective dates 11/1/2017. Accessed November 2017.
12. Stohl W, Schwarting A, Okada M, et al. Efficacy and Safety of Subcutaneous Belimumab in Systemic Lupus Erythematosus: A Fifty-Two-Week Randomized, Double-Blind, Placebo-Controlled Study. *Arthritis Rheumatol*. 2017;69(5):1016–1027. doi:10.1002/art.40049

Last Reviewed: 9/19/2018, 5/20/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/2019, 7/1/20, 1/1/22, 3/1/24

Besremi® (ropeginterferon alfa-2b-njft)
Prior Authorization Guidelines

Affected Medication(s)

- Besremi (ropeginterferon alfa-2b-njft) subcutaneous solution

FDA Approved Indication(s)

- Treatment of adults with polycythemia vera

Dosing

- Patients not already on hydroxyurea: Starting dose of 100 mcg subcutaneous injection every two weeks, increasing dose by 50mcg every two weeks (up to a maximum of 500 mcg), until hematological parameters are stabilized
- Patients transitioning from hydroxyurea: Starting dose of 50 mcg subcutaneous injection every two weeks in combination with hydroxyurea. Gradually taper off the hydroxyurea (see package insert for taper). Increase Besremi dose by 50 mcg every two weeks (up to a maximum of 500 mcg), until hematological parameters are stabilized
- If hematological stability achieved for at least 1 year, the dosing interval may be expanded to every 4 weeks

Initial Authorization Criteria

1. Is the request for continuation of a previously approved Besremi (ropeginterferon alfa-2b-njft) prior authorization with the same indication as the previous approval?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #4
    b. If no, continue to #3
3. Is the medication being requested for an indication supported by the National Comprehensive Cancer Network (NCCN) recommendation with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #4
    b. If not, clinical review required
4. Is the treatment being prescribed by, or in consultation with, an oncologist or hematologist?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required

Last Reviewed: 3/16/22, 3/17/23, 1/19/24
Effective Date: 4/15/22

6. Has the member previously trialed hydroxyurea with inadequate response, OR has a previous intolerance, contraindication, or medical rationale to avoid therapy with hydroxyurea been noted? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Has the member previously trialed Pegasys with inadequate response or has a documented intolerance or contraindication to treatment with Pegasys been provided? (Provide supporting documentation)
    a. If yes, approve for 4 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication or supported by NCCN recommendation with an evidence level of 2A or higher? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by, or in consultation with, an oncologist or hematologist?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there clinical documentation confirming disease responsiveness to therapy provided? (Examples include reduction in hematocrit <45% without phlebotomy, platelets ≤400 × 10^9/L and leukocytes ≤10 × 10^9/L, normal spleen size, etc.) (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. BESREMi (ropeginterferon alfa-2b-njft) injection, [package insert]. Burlington, MA: PharmaEssentia Corp; 2022.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 01 Feb. 2022].
3. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Myeloproliferative Neoplasms. Version 3.2022 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/pdf/mpn.pdf. Accessed Feb 13, 2023.
4. Gisslinger, Heinz, et al. "Ropeginterferon alfa-2b versus standard therapy for polycythaemia vera (PROUD-PV and CONTINUATION-PV): a randomised, non-inferiority, phase 3 trial and its extension study." The Lancet Haematology 7.3 (2020): e196-e208.

Last Reviewed: 3/16/22, 3/17/23, 1/19/24
Effective Date: 4/15/22

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Bylvay® (odevixibat)
Prior Authorization Guidelines

Affected Medication(s)

- Bylvay capsule/pellet

FDA Approved Indication(s)

- Treatment of pruritus in patients ≥3 months of age with progressive familial intrahepatic cholestasis (PFIC) (NOTE: May not be effective in PFIC type 2 patients with ABCB11 variants resulting in non-functional or complete absence of bile salt export pump protein (BSEP-3))
- Treatment of cholestatic pruritis in patients 12 months of age and older with Alagille Syndrome (ALGS)

Dosing

- Refer to package insert for specific dosing recommendations

Initial Authorization Criteria

1. Has the requested medication previously been approved by OHSU for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. What diagnosis is the medication being requested for?
    a. Progressive Familial intrahepatic cholestasis (PFIC), continue to corresponding criteria.
    b. Alagille syndrome, continue to corresponding criteria

Progressive Familial Intrahepatic Cholestasis

1. Does the member have a diagnosis of familial intrahepatic cholestasis (PFIC) Type I or II confirmed by presence of a mutation in the *ATP8B1 (FIC_1)* or *ABCB11* gene, liver biopsy or ultrasound or biliary lipid analysis? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 3 months of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a clinical history of cholestasis and have the other main causes of cholestasis been ruled out (e.g. biliary atresia, Alagille syndrome, alpha1antitrypsine deficiency, cystic fibrosis, sclerosing cholangitis and extrahepatic bile duct obstruction)? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of an inadequate response, intolerance, or contraindication to at least TWO of the following: ursodiol, cholestyramine, or rifampin? (Provide supporting documentation)
    a. If yes, continue to #5

Last Reviewed: 9/15/21, 3/17/23, 9/15/23
Effective Date: 11/1/21, 11/20/23

b. If no, clinical review required

5. Has the member previously undergone partial or total biliary diversion surgery that has been ineffective in relieving pruritus OR is medical rationale provided why the member cannot undergo surgery? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the medication prescribed by, or in consultation with, a hepatologist or an appropriate biliary specialist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Alagille Syndrome

1. Is the patient 12 months of age or older?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have documentation of Alagille syndrome confirmed by genetic testing or liver biopsy? (genetic testing with JAG1 or NOTCH2 mutation present) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member previously trialed a maximum tolerated dose of all of the following for at least 4 weeks with treatment failure, or is there a documented intolerance/contraindication to all of the following: rifampin, ursodiol, and cholestyramine/colesevelam? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has the member previously trialed a maximum tolerated dose of Livmarli (maralixibat) for at least 4 weeks with treatment failure, or is there a documented intolerance/contraindication to Livmarli (maraliximab)? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is there documentation member is experiencing moderate to severe pruritus despite current therapy? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is Bylvay (odevixibat) being prescribed by, or in consultation with, a gastroenterologist or specialist experienced in treating Alagille syndrome?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?

a. If yes, continue to #2
b. If no, clinical review required

2. Were updated chart notes (within 1 year) with documentation of improvement in pruritis symptoms from baseline received? (Provide supporting documentation)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with, a hepatologist or an appropriate biliary specialist?

    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. BYLVAY (odevixibat) capsule/pellets [package insert]. Boston, MA: Albireo Pharma, Inc.; 2023.
2. Roy-Chowdhury J, Roy-Chowdhury N. Inherited disorders associated with conjugated hyperbilirubinemia. In: UpToDate, K. Lindor, S. Grover (Eds), UpToDate, Waltham, MA. (Accessed on August 27, 2021.)
3. Davit-Spraul, Anne, et al. "Progressive familial intrahepatic cholestasis." Orphanet journal of rare diseases 4.1 (2009): 1-12.
4. Gunaydin, Mithat, and Asudan Tugce Bozkurter Cil. “Progressive familial intrahepatic cholestasis: diagnosis, management, and treatment.” Hepatic medicine: evidence and research vol. 10 95-104. 10 Sep. 2018, doi:10.2147/HMER.S137209

Last Reviewed: 9/15/21, 3/17/23, 9/15/23
Effective Date: 11/1/21, 11/20/23

Cablivi® (caplacizumab-yhdp)
Prior Authorization Guidelines

Affected Medication(s)

- Cablivi (caplacizumab-yhdp) subcutaneous solution

FDA Approved Indication(s)

- Treatment of adult patients with acquired thrombotic thrombocytopenic purpura (aTTP), in combination with plasma exchange and immunosuppressive therapy

Dosing

Should be administered upon initiation of plasma exchange therapy:

- First day of treatment: 11 mg bolus intravenous (IV) injection at least 15 minutes prior to plasma exchange followed by an 11 mg subcutaneous (SC) injection after completion of plasma exchange on day 1
- Subsequent days of treatment during daily plasma exchange: 11 mg SC injection once daily following plasma exchange
- Treatment after plasma exchange period: 11 mg SC injection once daily continuing for 30 days following the last daily plasma exchange. Treatment may be extended for a maximum of 28 days if patient have signs of persistent underlying disease such as suppressed ADAMTS13 activity levels remain present after initial treatment course.

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a diagnosis of acquired TTP confirmed by severe ADAMTS13 deficiency with ADAMTS13 activity levels of less than 10% and thrombocytopenia and/or microangiopathic hemolytic anemia OR a PLASMIC scored of 6-7? (Provide ADAMTS13 activity level or PLASMIC score for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Was the therapy started upon initiation of plasma exchange therapy in combination with corticosteroids?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member received, or planning to receive, the IV bolus dose?
    a. If yes, continue to #6
    b. If no, clinical review required

Last Reviewed: 5/20/20, 3/10/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 7/1/20, 5/1/21, 4/15/22, 6/15/24

6. Is this medication being prescribed by, or in consultation with, a hematologist?
    a. If yes, approve up to 30 days following the last day of plasma exchange therapy
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Caplacizumab-yhdp (Cablivi) [package insert]. Cambridge, MA: Genzyme Corporation; March 2022.
2. Cully M, Hunt BJ, Benjamin S, et al. Guidelines on the diagnosis and management of thrombotic thrombocytopenic purpura and other thrombotic microangiopathies. Br J Haematol. 2012;158(3):323-35.
3. Schwartz J, Padmanabhan A, Aqui N, et al. Guidelines on the use of therapeutic apheresis in clinical practice-evidence-based approach from the writing committee of the American society for apheresis: The Seventh special issue. J Clin Pher. 2016 Jun;31(3):149-62.
4. Bendapudi PK, Hurwitz S, Fry A, et al. Derivation and external validation of the PLASMIC score for rapid assessment of adults with thrombotic microangiopathies: a cohort study. Lancet Haematology. 2017;4(4):e157
5. Zheng ZL, Vesely SK, Cataland SR, et al. ISTH guidelines for treatment of thrombotic thrombocytopenic purpura. J Thromb Haemost. 2020;18(10):2496-2502.

Last Reviewed: 5/20/20, 3/10/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 7/1/20, 5/1/21, 4/15/22, 6/15/24

Camzyos® (mavacamten)
Prior Authorization Guidelines

Affected Medication(s)

- Camzyos (mavacamten) oral capsule

FDA Approved Indication(s)

- Treatment of adults with symptomatic New York Heart Association (NYHA) class II-III obstructive hypertrophic cardiomyopathy (HCM) to improve functional capacity and symptoms

Dosing

- Recommended starting dose: 5 mg once daily
- Allowable subsequent doses with titration are 2.5, 5, 10, or 15 mg once daily

Initial Authorization Criteria

1. Is the request for continuation of Camzyos (mavacamten) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member aged 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of NYHA class II to III obstructive hypertrophic cardiomyopathy (HCM) and is experiencing symptoms? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have documentation of a left ventricular ejection fraction (LVEF) of 55% or greater? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have documentation of a LVOT peak gradient of 50 mmHg or greater at rest or with provocation? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Has the member previous trialed all of the following classes of medications at maximally indicated doses unless intolerance or contraindication is present? (Provide supporting documentation)
    - Non-vasodilating beta-blocker (i.e. atenolol, bisoprolol, metoprolol, propranolol)
    - Non-dihydropyridine calcium channel blocker (i.e. verapamil, diltiazem)

Last Reviewed: 7/20/22, 7/21/23
Effective Date: 9/1/22

- Disopyramide

a. If yes, continue to #8
b. If no, clinical review is required

8. Is Camzyos® (mavacamten) being prescribed by, or in consult with, a cardiologist?

a. If yes, approve for 6 months
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)

a. If yes, continue to #2
b. If no, clinical review required

2. Is the treatment being prescribed by or in consultation with a cardiologist?

a. If yes, continue to #3
b. If no, clinical review required

3. Were updated chart notes (within the past year) provided with documentation of significant clinical response to prior therapy received? (i.e. improvement NYHA Class II-III symptoms) (Provide supporting documentation)

a. If yes, continue to #4
b. If no, clinical review required

4. Does the member have an updated echocardiogram (within that past year and since starting therapy) showing LVEF of at least 50%?

a. If yes, approve for 12 months
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. CAMZYOS (mavacamten) capsules, [package insert]. Brisbane, CA: MyoKardia, Inc.; 2022.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 26 May. 2022].
3. Olivotto, Iacopo, et al. "Mavacamten for treatment of symptomatic obstructive hypertrophic cardiomyopathy (EXPLORER-HCM): a randomised, double-blind, placebo-controlled, phase 3 trial." The Lancet 396.10253 (2020): 759-769.
4. Ommen, Steve R., et al. "2020 AHA/ACC guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines." Journal of the American College of Cardiology 76.25 (2020): e159-e240.

Last Reviewed: 7/20/22, 7/21/23
Effective Date: 9/1/22

Carbaglu® (carglumic acid)
Prior Authorization Guidelines

Affected Medication(s)

- Carbaglu oral tablet
- Carglumic acid oral tablet

FDA Approved Indication(s)

- Adjunctive therapy in pediatric and adult patients for treatment of acute hyperammonemia due to deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS)
- Maintenance therapy in pediatric and adult patients for treatment of chronic hyperammonemia due to deficiency of the hepatic enzyme N-acetylglutamate synthase (NAGS)
- Adjunctive treatment in adult and pediatric patients for the treatment of acute hyperammonemia due to propionic acidemia (PA) or methylmalonic acidemia (MMA)

Dosing

NAGS Deficiency:

- Acute: 100 mg/kg to 250 mg/kg divided into 2 to 4 doses (rounded to the nearest 100 mg)
- Maintenance: 10 mg/kg to 100 mg/kg divided into 2 to 4 doses (rounded to the nearest 100 mg)

Acute Hyperammonemia due to PA or MMA:

- Patients ≤15 kg: 150 mg/kg/day in two equal doses administered 12 hours apart
- Patients >15 kg: 3.3 g/m^2/day in two equal doses administered 12 hours apart

Initial Authorization Criteria

1. Is the request for continuation of Carbaglu (carglumic acid) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the diagnosis of NAGS deficiency, MMA, or PA confirmed by plasma amino acid/urine orotic acid or enzyme analysis? (Provide lab report confirming diagnosis)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is baseline plasma ammonia level provided? (Provide lab for review)
    a. If yes, continue to #5
    b. If no, clinical review required

Last Reviewed: 11/21/18, 11/18/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/19, 1/1/22, 6/1/23, 3/1/24

5. Will Carbaglu (carglumic acid) be used with other methods to lower plasma ammonia level? (i.e. hemodialysis, sodium phenylacetate and sodium benzoate)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the member adherent to a protein restrictive diet? (Provide documentation of restrictive diet)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the treatment being initiated by a provider that specializes in the treatment of inherited metabolic disorders? (Examples include a medical geneticist or an endocrinologist)
    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member show a positive clinical response to therapy as defined be a decrease in plasma ammonia levels from baseline? (Provide documentation of current and baseline plasma ammonia levels for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being initiated by a provider that specializes in the treatment of inherited metabolic disorders? (Examples include a medical geneticist or an endocrinologist)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Carbaglu [prescribing information]. Puteaux, France: Orphan Europe, SARL; August 2021.
2. Lee, B. Urea cycle disorder: clinical features and diagnosis. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed December 2022.
3. Lee, B. Urea cycle disorder: management. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed December 2022.

Last Reviewed: 11/21/18, 11/18/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/19, 1/1/22, 6/1/23, 3/1/24

Cayston® (aztreonam)
Prior Authorization Guidelines

Affected Medication(s)

- Cayston powder for inhalation solution

FDA Approved Indication(s)

- To improve respiratory symptoms in cystic fibrosis (CF) patients with *Pseudomonas aeruginosa*

Dosing

- For adults and children 7 years of age and older:
  - One vial (75mg) reconstituted with 1 mL of sterile diluent administered 3 times a day for a 28 day course

Initial Authorization Criteria

1. Is the request for continuation of Cayston (aztreonam) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have cystic fibrosis and a lung infection with a positive culture demonstrating *Pseudomonas aeruginosa* infection? (Provide supporting documentation of cystic fibrosis diagnosis and positive culture for *Pseudomonas aeruginosa*)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Does the member have baseline FEV1 greater than 25%? (Provide FEV1 for review)
   a. If yes, continue to #5
   b. If no, clinical review required
5. Does the member have a previous trial with inadequate response, contraindication, or intolerance to tobramycin inhaled solution? (Provide supporting documentation for review)
   a. If yes, approve for 1 month unless otherwise specified
   b. If no, continue to # 6
6. Does the member have a culture showing resistance to tobramycin? (Provide culture results for review)
   a. If yes, approve for 1 month unless otherwise specified
   b. If no, clinical review required

Last Reviewed: 11/21/18, 11/18/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/19, 3/1/24

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is documentation confirming improvement in respiratory symptoms provided? (Provide supporting documentation for review)
    a. If yes approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Cayston (aztreonam) [Prescribing Information]. Foster City, CA: Gilead Sciences, Inc. December 2021.
2. Oermann, CM, Retsch-Bogart, GZ, Ouittner, AL, et al. An 18-month Study of the Safety and Efficacy of Repeated Course of Inhaled Aztreonam Lysine in Cystic Fibrosis. Pediatric Pulmonology. 2010 November; 45(11): doi:10.1002/ppul.21301.
3. Wainwright, CE, Quittner AL, Geller, DE, et al. Aztreonam for inhalation (AZLI) in patients with cystic fibrosis, mild lung impairment, and *P.aeruginosa*. Journal of Cycstic Fibrosis 10(2011) 234-242.
4. Mogayzel Jr, Peter J., et al. "Cystic Fibrosis Foundation pulmonary guideline. Pharmacologic approaches to prevention and eradication of initial Pseudomonas aeruginosa infection." Annals of the American Thoracic Society 11.10 (2014): 1640-1650.

Last Reviewed: 11/21/18, 11/18/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/19, 3/1/24

Chenodal® (chenodiol)
Prior Authorization Guidelines

Affected Medication(s)

- Chenodal oral tablet

FDA Approved Indication(s)

- For patients with radiolucent stones in well-opacifying gallbladders, in whom selective surgery would be undertaken except for the presence of increased surgical risk due to systemic disease or age

Dosing

- Initially: 250mg twice daily for 2 weeks
- Then, increase by 250 mg/day each week until recommended or maximum tolerated dose is reached
- Refer to package insert for recommended dosing specifications

Initial Authorization Criteria

1. Is the request for continuation of Chenodal (chenodiol) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there documentation that the member is not a candidate for surgery? (Provide rationale stating why member is not a candidate for surgery)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of a previous inadequate response, contraindication or intolerance to ursodiol? (Provide history of ursodiol use or contraindication to therapy)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Does the member continue to meet the above criteria?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member exceeded 24 months of therapy in this treatment course?
    a. If yes, clinical review required
    b. If no, approve for up to 12 months unless otherwise specified (Not to exceed 24 months of treatment)

Last Reviewed: 10/3/18, 11/18/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/19, 3/1/24

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Chenodal tablets [prescribing information]. San Diego, CA: Retrophin; November 2021.

2. Chenodal. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed September 21, 2021.

3. Zakko MD. Nonsurgical treatment of gallstones. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed September 19, 2018.

Last Reviewed: 10/3/18, 11/18/20, 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/19, 3/1/24

Cholbam® (cholic acid)
Prior Authorization Guidelines

Affected Medication(s)

- Cholbam oral capsule

FDA Approved Indication(s)

- Treatment of bile acid synthesis disorders due to single enzyme defects (SEDs)
- Adjunctive treatment of peroxisomal disorders (PDs) including Zellweger spectrum disorders in patients who exhibit manifestations of liver disease, steatorrhea or complications from decreased fat soluble vitamin absorption

Dosing

- Refer to package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of Cholbam (cholic acid) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a diagnosis of peroxisomal disorder? (Provide documentation to support confirmation of diagnosis)
    a. If yes, continue to #4
    b. If no, continue to #7
4. Does the member have manifestations of at least one of the following? (Provide supporting documentation)
    - Liver disease (ex. jaundice or elevated liver enzymes)
    - Steatorrhea
    - Complications from decreased fat-soluble vitamin absorption

    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment intended for extrahepatic signs and/or symptoms of peroxisomal disorders? (Examples include psychomotor retardation, neurologic dysfunctions, hearing loss, visual abnormalities, and/or osteoporosis)
    a. If yes, clinical review required
    b. If no, continue to #6
6. Was a baseline liver function test and INR received? (Provide lab results)

Last Reviewed: 10/17/18, 1/20/21, 3/16/22, 3/17/23
Effective Date: 1/1/19, 6/1/23

a. If yes, continue to #7
b. If no, clinical review required

7. Is the treatment being prescribed by, or in consult with, a medical geneticist, a pediatric gastroenterologist, a hepatologist, or a specialist experienced in treating inborn errors of metabolism?

    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member demonstrated a positive clinical response to therapy defined as decreased signs and/or symptoms from baseline? (Provide supporting documentation and updated liver function tests)

    a. If yes, continue to #3
    b. If no, clinical response required

3. Is the treatment being prescribed by, or in consult with, a medical geneticist, a pediatric gastroenterologist, a hepatologist, or a specialist experienced in treating inborn errors of metabolism?

    a. If yes, approve for 1 year unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Cholbam [Prescribing Information]. Baltimore, MD: Asklepion Pharmaceuticals LLC; May 2021.
2. Wanders, R. Peroxisomal disorders. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed January 10, 2023.

Last Reviewed: 10/17/18, 1/20/21, 3/16/22, 3/17/23
Effective Date: 1/1/19, 6/1/23

Compounded Medications
Prior Authorization Guidelines

Affected Medication(s)

- All compounded medications that are not commercially available

FDA Approved Indication(s)

- Refer to indications associated with active ingredient(s)

Dosing

- Refer to dosing associated with active ingredient(s)

Initial & Reauthorization Authorization Criteria

1. Are all active ingredients in the compounded medication FDA-approved?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Are all active ingredients being used for an FDA approved or major compendia supported indication? (Provide documentation of diagnosis and treatment plan)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there an FDA-approved commercially available medication on the market for treatment of the requested condition?
    a. If yes, continue to #4
    b. If no, approve for 3 months unless otherwise specified
4. Is there documentation to support medical necessity over commercially available products? (Provide documentation supporting use over commercially available product)
    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

Last Reviewed: 9/19/2018, 11/18/20, 11/17/21, 3/17/23, 3/15/24
Effective Date: 1/1/2019

Corlanor® (ivabradine)
Prior Authorization Guidelines

Affected Medication(s)

- Corlanor oral tablet
- Corlanor oral solution

FDA Approved Indication(s)

- To reduce the risk of hospitalization for worsening heart failure in patients with stable, symptomatic chronic heart failure with left ventricular ejection fraction ≤ 35%, who are in sinus rhythm with resting heart rate ≥ 70 beats per minute and either are on maximally tolerated doses of beta-blockers or have a contraindication to beta-blocker use
- Treatment of stable symptomatic heart failure due to dilated cardiomyopathy (DCM) in pediatric patients aged 6 months and older, who are in sinus rhythm with an elevated heart rate

Dosing

- Adults:
  - Initial: 5 mg twice daily with meals
  - After 2 weeks, adjust dose to achieve a resting heart rate between 50-60 beats per minute (reference dosage adjustments in package insert)
- Pediatrics:
  - Please see package insert for specific pediatric weight-based dosing

Initial Authorization Criteria

1. Is the request for continuation of Corlanor (ivabradine) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member aged 6 months or older?
   a. If yes, continue to #4
   b. If no, clinical review required
4. What is the indication the medication is being requested for?
   a. Chronic heart failure, continue to question #5
   b. Inappropriate sinus tachycardia, continue to #8
   c. Other indication, clinical review required
5. Does the member have a left ventricular ejection fraction of 35% or less for adults or 45% or less for pediatrics? (Provide documentation for review)
   a. If yes, continue to #6
   b. If no, clinical review required

Last Reviewed: 10/3/18, 11/18/20, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 12/15/20, 7/15/22, 6/15/24

6. Is the member currently in sinus rhythm with a resting heart rate meeting one of the below? (Provide heart rate for review)
    - ≥ 70 bpm for ages 5 years and older
    - ≥75 bpm for ages 3-5 years
    - ≥95 bpm for ages 1-3 years
    - ≥105 bpm for ages 6-12 months

    a. If yes, continue to # 7
    b. If no, clinical review required
7. Is the member on a maximally tolerated dose of a beta-blocker (i.e. metoprolol succinate, carvedilol, or bisoprolol) or have contraindication to their use? (Provide history of beta-blocker use or contraindication to therapy)

    a. If yes, continue to #10
    b. If no, clinical review required
8. Does the member have confirmation of inappropriate sinus tachycardia defined as a sinus heart rate greater than 100 bpm at rest (with a mean 24-hour heart rate >90 bpm not due to primary causes) AND is associated with distressing symptoms of palpitations (i.e. presyncope, headache, dyspnea, etc.)? (Provide supporting documentation)

    a. If yes, continue to #9
    b. If no, clinical review required
9. Have all other causes of sinus tachycardia have been ruled out (examples include thyroid disease or drug induced)?

    a. If yes, continue to #10
    b. If no, clinical review required
10. Is the treatment being prescribed by or in consult with a cardiologist or other appropriate specialist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by or in consultation with a cardiologist or other appropriate specialist?

    a. If yes, continue to #3
    b. If no, clinical review required
3. Were updated chart notes provided with documentation of significant clinical response to therapy? (Provide updated clinical information for review such as heart rate stabilization, improvement in HF symptoms, etc.)

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 10/3/18, 11/18/20, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 12/15/20, 7/15/222, 6/15/24

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Corlanor. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed October 23, 2020
2. Corlanor (ivabradine) [Prescribing Information]. Thousand Oaks, CA: Amgen. April 2019.
3. Yancy, Clyde W., et al. "2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America." Journal of the American College of Cardiology 70.6 (2017): 776-803.
4. Sheldon, Robert S., et al. "2015 heart rhythm society expert consensus statement on the diagnosis and treatment of postural tachycardia syndrome, inappropriate sinus tachycardia, and vasovagal syncope." Heart rhythm 12.6 (2015): e41-e63.

Last Reviewed: 10/3/18, 11/18/20, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 12/15/20, 7/15/222, 6/15/24

Cresemba® (isavuconazonium)
Prior Authorization Guidelines

Affected Medication(s)

- Cresemba oral capsule

FDA Approved Indication(s)

- For patients 6 years of age and older who weigh 16 kilograms and greater for the treatment of invasive aspergillosis
- For patients 6 years of age and older who weigh 16 kilograms and greater for the treatment of invasive mucormycosis

Dosing

- Adults:
  - Loading dose: Two 186 mg-capsules (372 mg) orally every 8 hours for 48 hours
  - Maintenance dose: Two 186 mg-capsules (372 mg) orally once daily
- Pediatrics:
  - See package insert for specific pediatric weight based dosing

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #2
   b. If no, clinical review required
2. Is the member 6 years of age or older?
   a. If yes, continue to #3
   b. If no, clinical review required
3. What is the requested diagnosis?
   a. Invasive aspergillosis, continue to #4
   b. Invasive mucormycosis, continue to #6
4. Is documentation of rationale for avoidance or contraindication to both voriconazole and posaconazole received? (Provide supporting documentation)
   a. If yes, continue to #9
   b. If no, continue to #5
5. Did the member initiate Cresemba (isavuconazonium) therapy during the inpatient stay?
   a. If yes, continue to #9
   b. If no, clinical review required
6. Did the member have a trial with response to amphotericin B or documentation of intolerance or contraindication? (Provide supporting documentation)
   a. If yes, continue to #7
   b. If no, continue to #8

Last Reviewed: 10/17/18, 1/20/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 4/15/22, 6/15/24

7. Is documentation of rationale for avoidance or contraindication to posaconazole received? (Provide supporting documentation)
    a. If yes, continue to #9
    b. If no, continue to #8

8. Did the member initiate Cresemba (isavuconazonium) therapy during the inpatient stay?
    a. If yes, continue to #9
    b. If no, clinical review required

9. Is the treatment being initiated by an infectious disease specialist?
    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Cresemba Prescribing Information. Northbrook, IL: Astellas, Inc.; June 2015. Available at: www.cresemba.com.
2. Patterson TF, Thompson GR, Denning DW et al. Practice Guidelines for the Diagnosis and Management of Aspergillosis: 2016 Update by the Infectious Diseases Society of America. Clin Infect Dis. 2016 Aug 15;63(4):e1-e60. doi: 10.1093/cid/ciw326.
3. Tissot F, Agrawal S, Pagano L, et al. ECIL-6 guidelines for the treatment of invasive candidiasis, aspergillosis and mucormycosis in leukemia and hematopoietic stem cell transplant patients. Haematologica. Mar 2017, 102(3) 433-444.
4. Centers for Disease Control and Prevention. Fungal Diseases: Treatment of Mucormycosis. Last updated December 30, 2015. Available at: https://www.cdc.gov/fungal/diseases/mucormycosis/treatment.html.
5. Kauffman, CA. Treatment and prevention of invasive aspergillosis. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed September 2018.
6. Cox, GM. Mucormycosis (zygomycosis). Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed September 2018.

Last Reviewed: 10/17/18, 1/20/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 4/15/22, 6/15/24

Cystagon®, Procysbi® (cysteamine)
Prior Authorization Guidelines

Affected Medication(s)

- Cystagon oral capsule
- Procysbi oral capsule delayed release
- Procysbi DR granule packet

FDA Approved Indication(s)

- Cystagon: Management of nephropathic cystinosis in children and adults
- Procysbi: Treatment of nephropathic cystinosis in adults and pediatric patients 1 year of age and older

Dosing

- Cystagon:
  - Initially: Start at 1/4 to 1/6 of the maintenance dose and increase over 4-6 weeks
  - Maintenance dose: 1.30 grams/m^2/day divided into four doses/daily
  - Use chart in package insert for weight-based dosing
- Procysbi:
  - Initially: Start at 1/4 to 1/6 of the maintenance dose and increase over 4-6 weeks
  - Maintenance dose: 1.30 grams/m^2/day divided into two doses/daily
  - Use chart in package insert for weight-based dosing

Initial Authorization Criteria

1. Is the request for continuation of Cystagon (cysteamine) or Procysbi (cysteamine) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have confirmation of nephropathic cystinosis defined by elevated leukocyte cysteine levels (LCL) or presence of the CTNS gene mutation? (Provide documentation of elevated LCL or CTNS gene mutation)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is the request for Procysbi (cysteamine)?
   a. If yes, continue to #5
   b. If no, continue to #7
5. Is clinical rationale for avoiding Cystagon (cysteamine) provided? (Provide supporting documentation with inadequate response or intolerance)
   a. If yes, continue to #6
   b. If no, clinical review required

Last Reviewed: 11/21/18, 3/18/20, 7/21/21, 3/17/23
Effective Date: 1/1/19, 5/1/20

6. Is the member at least one year of age or older?
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the treatment being initiated by a specialist experienced in the management of nephropathic cystinosis? (Examples include endocrinologist, nephrologist, or urologist)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have documentation of a positive clinical response to therapy as evidenced by a reduction in WBC cysteine levels compared to pre-treatment? (Provide supporting documentation for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being initiated by a specialist experienced in the management of nephropathic cystinosis? (examples include endocrinologist, nephrologist, or urologist)
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Procysbi [package insert]. Novato, CA: Raptor Pharmaceuticals, Inc.; February 2022.
2. Cystagon [package insert]. Morgantown, WV: Mylan Pharmaceuticals, Inc.; August 2021.
3. Langman CB, Greenbaum LA, Sarwal M, et al. A randomized controlled crossover trial with delayed-release cysteamine bitartrate in nephropathic cystinosis: effectiveness on white blood cell cysteine levels and comparison of safety. Clin J Am Soc Nephrol 2012;7:1112-1120.
4. Niaudet P. Cystinosis. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed June 2021.

Last Reviewed: 11/21/18, 3/18/20, 7/21/21, 3/17/23
Effective Date: 1/1/19, 5/1/20

Cystaran®, Cystadrops® (cysteamine hydrochloride) Prior Authorization Guidelines

Affected Medication(s)

- Cystaran 0.44% ophthalmic solution
- Cystadrops 0.37% ophthalmic solution

FDA Approved Indication(s)

- Treatment of corneal cystine crystal accumulation in patients with cystinosis

Dosing

- Cystaran:
  - One drop in each eye, every waking hour
- Cystadrops
  - One drop in each eye 4 times daily during waking hours

Initial Authorization Criteria

1. Is the request for continuation of Cystaran (cysteamine hydrochloride) or Cystadrops (cysteamine) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have a presence of corneal cysteine accumulation? (Provide supporting documentation)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is the request for use of Cystadrops?
   a. If yes, continue to #5
   b. If no, continue to #6
5. Has the member had a previous trial with inadequate response or intolerance to treatment with Cystaran?
   a. If yes, continue to #6
   b. If no, clinical review required
6. Is the treatment being prescribed by, or in consultation with an ophthalmologist?
   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

Reauthorization Criteria

Last Reviewed: 10/17/18, 11/18/20, 1/20/21,3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 12/15/20, 3/1/21, 4/15/22, 6/15/24

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member currently have a presence of corneal cysteine accumulation OR did the member previously have corneal cysteine accumulation prior to the start of Cystaran or Cystadrops therapy? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member demonstrated a positive clinical response to therapy? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by, or in consultation with an ophthalmologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Cystaran Prescribing Information. Amityville, NY: Hi-Tech Pharmacal Co., Inc., October 2012. Available at http://www.cystaran.com/. Accessed October 23, 2020.
2. Cystadrops Prescribing Information. Lebanon, NJ: Recordati Rare Diseases Inc., September 2020. Accessed January 15, 2021.
3. Cystinosis. National Organization for Rare Disorders website. https://rarediseases.org/rare-diseases/cystinosis/. Published 1986. Updated 2017. Accessed September 2018.

Last Reviewed: 10/17/18, 11/18/20, 1/20/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 12/15/20, 3/1/21, 4/15/22, 6/15/24

Daliresp® (roflumilast)
Prior Authorization Guidelines

Affected Medication(s)

- Daliresp (roflumilast) oral tablet
- roflumilast oral tablet

FDA Approved Indication(s)

- Treatment to reduce the risk of COPD exacerbations in patients with severe COPD associated with chronic bronchitis and a history of exacerbations.

Dosing

- 500 mcg tablet daily
- Patients may initially start at 250 mcg daily for 4 weeks

Initial Authorization Criteria

1. Is the request for continuation of Daliresp (roflumilast) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is Daliresp (roflumilast) being requested for COPD with associated bronchitis in a member with a history of exacerbations? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a FEV1 of ≤ 50% predicted? (Provide documentation of baseline FEV1)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of minimum 3 months trial or, intolerance, or contraindication to maintenance triple therapy with a long acting beta agonist, a long acting anti-muscarinic agonist, and an inhaled corticosteroid? (Provide supporting documentation of all therapies tried)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by or in consultation with a pulmonologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is Daliresp (roflumilast) being requested for COPD with associated bronchitis in a member with a history of exacerbations? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required

Last Reviewed: 9/19/2018, 1/20/2021, 3/16/22, 11/16/22, 11/17/23
Effective Date: 1/1/2019, 1/1/23

2. Were updated chart notes (within 1 year) provided with documentation of significant clinical response to therapy defined as a decrease in COPD exacerbations from baseline? (Provide documentation of decreased COPD exacerbations)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with a pulmonologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Daliresp (roflumilast)[Prescribing Information]. Wilmington, DE: AstraZeneca Pharmaceuticals LP. March 2020.
2. Daliresp. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed March 8, 2022.
3. Ferguson MD, Make MD. Management of refractory chronic obstructive pulmonary disease. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed March 8, 2022.
4. "Global strategy for the diagnosis, management, and prevention of chronic obstructive pulmonary disease: 2022 Report." (2022).

Last Reviewed: 9/19/2018, 1/20/2021, 3/16/22, 11/16/22, 11/17/23
Effective Date: 1/1/2019, 1/1/23

Daybue® (trofinetide)
Prior Authorization Guidelines

Affected Medication(s)

- Daybue (trofinetide) oral tablet

FDA Approved Indication(s)

- Treatment of Rett syndrome in adults and pediatric patients 2 years of age and older

Dosing

- Refer to package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is Daybue® (trofinetide) being requested for an FDA approved indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 2 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of typical Rett syndrome according to the Rett Syndrome Diagnostic Criteria with a disease-causing mutation in the MECP2 gene as confirmed by genetic testing? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a current clinically significant cardiovascular, endocrine (such as hypo- or hyperthyroidism, Type 1 diabetes mellitus, or uncontrolled Type 2 diabetes mellitus), renal, hepatic, respiratory or gastrointestinal disease (such as celiac disease or inflammatory bowel disease)? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #6
6. Does the member have a history of or current cerebrovascular disease or brain trauma? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #7
7. Does the member have significant, uncorrected visual or hearing impairment? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #8

Last Reviewed: 5/19/23
Effective Date: 7/15/23

8. Is the treatment being prescribed by, or in consultation with, a clinical geneticist or neurologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within past year) provided with documentation of clinical response to prior therapy received?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a clinical geneticist or neurologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. DAYBUE (trofinetide) oral solution, [package insert]. San Diego, CA: Acadia Pharmaceuticals Inc.; 2023.
2. Drugs@FDA: FDA Approved Drug Products. 2023. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 27 Mar. 2023].
3. Neul, J. L., Percy, A. K., Benke, T. A., Berry-Kravis, E. M., Glaze, D. G., Peters, S. U., Jones, N. E., & Youakim, J. M. (2022). Design and outcome measures of LAVENDER, a phase 3 study of trofinetide for Rett syndrome. Contemporary clinical trials, 114, 106704.
4. Fu, C., Armstrong, D., Marsh, E., Lieberman, D., Motil, K., Witt, R., Standridge, S., Nues, P., Lane, J., Dinkel, T., Coenraads, M., von Hehn, J., Jones, M., Hale, K., Suter, B., Glaze, D., Neul, J., Percy, A., & Benke, T. (2020). Consensus guidelines on managing Rett syndrome across the lifespan. BMJ paediatrics open, 4(1), e000717.
5. Jeffrey L. Neul, , Kaufmann, W.E., Glaze, D.G., Christodoulou, J., Clarke, A.J., Bahi-Buisson, N., Leonard, H., Bailey, M.E.S., Schanen, N.C., Zappella, M., Renieri, A., Huppke, P., Percy, A.K. and (2010), Rett syndrome: Revised diagnostic criteria and nomenclature. Ann Neurol., 68: 944-950.

Diacomit® (stiripentol)
Prior Authorization Guidelines

Affected Medication(s)

- Diacomit oral capsule
- Diacomit oral powder for suspension

FDA Approved Indication(s)

- Treatment of seizures associated with Dravet syndrome (DS) in patients 6 months of age and older and weighing 7 kg or more taking clobazam

Note: There is no clinical data to support use of Diacomit as monotherapy in Dravet syndrome

Dosing

- 50 mg/kg/day administered in 2 or 3 divided doses

Initial Authorization Criteria

1. Is the request for continuation of Diacomit (stiripentol) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member currently have a diagnosis of Dravet syndrome?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has the patient previously trialed valproate, topiramate, and clobazam, unless intolerance or contraindication, and continued to have 4 or more generalized tonic-clonic seizures per month despite optimized therapy?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Will Diacomit (stiripentol) be used in conjunction with clobazam?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Last Reviewed: 9/18/19, 11/20/19, 7/21/21, 11/16/22, 9/15/23
Effective Date: 10/15/19, 1/1/23, 11/30/23

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within the past 6 months) with documentation of at least a 50% decrease in the frequency of generalized clonic and tonic-clonic seizures?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Will Diacomit (stiripentol) be used in combination with clobazam?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by or in consultation with a neurologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Lexicomp [internet database]. Hudson, OH: Wolters Kluwer. Updated periodically. Accessed August 13, 2019.
2. Stiripentol (Diacomit) Capsule and powder for suspension [package insert]. Redwood City, USA: Bicodex Inc; July 2022.
3. Drugs@FDA: FDA Approved Drug Products. 2019. accessdatafda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 13 Aug. 2019].
4. National Institute for Health and Care Excellence; Epilepsies: Diagnosis and Management Clinical Guidelines. *NICE Guideline*. May 2021. Available at: https://www.nice.org.uk/guidance/cg137
5. Wilmhurst JM, Gaillard WD, Vinayan KP, et al. Summary of recommendations for the management of infantile seizures: Task force report for the ILAE commission of pediatrics. Epilepsia. 2015; 56(8):1185-1197

Last Reviewed: 9/18/19, 11/20/19. 7/21/21, 11/16/22, 9/15/23
Effective Date: 10/15/19, 1/1/23, 11/20/23

Dibenzyline® (phenoxybenzamine)
Prior Authorization Guidelines

Affected Medication(s)

- Dibenzyline (phenoxybenzamine) oral capsule
- phenoxybenzamine oral capsule

FDA Approved Indication(s)

- Treatment of sweating and hypertension associated with pheochromocytoma

Dosing

- 20-40 mg orally two to three times daily

Initial Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have confirmed pheochromocytoma by imaging? (Provide supporting documentation for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is phenoxybenzamine being requested as preoperative management? (Provide treatment plan/duration and planned surgical date)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is documentation with rationale to avoid other alpha-blockers received? (i.e. prazosin, terazosin, doxazosin)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by or in consultation with an endocrinologist?
    a. If yes, approve for 1 month unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Dibenzyline [package insert]. Chestnut Ridge, NY: Par Pharma; August 2021.

Last Reviewed: 11/20/19, 5/19/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 2/1/20, 6/15/21, 6/15/24

2. Lenders JWM, Duh QY, Eisenhofer G, et al. Phenochromocytoma and paraganglioma: an endocrine society clinical practice guideline. The Journal of Clinical Endocrinology & Metabolism. 2014;6:1915-1942. Available at: https://doi.org/10.1210/jc.2014-1498.
3. Fassnacht, M., et al. "Adrenocortical carcinomas and malignant phaeochromocytomas: ESMO–EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up." Annals of Oncology 31.11 (2020): 1476-1490.

Last Reviewed: 11/20/19, 5/19/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 2/1/20, 6/15/21, 6/14/24

Doptelet® (avatrombopag maleate) & Mulpleta® (lusutrombopag) Prior Authorization Guidelines

Affected Medication(s)

- Doptelet (avatrombopag) oral tablet
- Mulpleta (lusutrombopag) oral tablet

FDA Approved Indication(s)

- Doptelet:
  - For the treatment of thrombocytopenia in adult patients with chronic liver disease who are scheduled to undergo a procedure
  - The treatment of thrombocytopenia in adult patients with chronic immune thrombocytopenia who have had an insufficient response to a previous treatment
- Mulpleta:
  - For the treatment of thrombocytopenia in adult patients with chronic liver disease who are scheduled to undergo a procedure

Dosing

- Doptelet:
  - Chronic liver disease:
    - Platelet count <40: 60 mg for 5 days
    - Platelet count 40-50: 40 mg for 5 days
  - Chronic ITP
    - Starting dose 20mg once daily, refer to package insert for dose adjustments based on platelet counts
- Mulpleta
  - 3 mg for 7 days 8 to 14 days prior to a scheduled procedure

Authorization Criteria

1. Is the request for continuation of Doptelet for the treatment of chronic immune thrombocytopenia?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member 18 years of age or older?
   a. If yes, continue to #4
   b. If no, clinical review required
4. What indication is the medication being requested for?
   a. Thrombocytopenia in adult with chronic liver disease, continue to corresponding criteria

Last Reviewed: 11/21/18, 5/19/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 6/15/21, 1/1/23

b. Thrombocytopenia in adult with chronic immune thrombocytopenia, continue to corresponding criteria (Doptelet only)

Thrombocytopenia in adult with chronic liver disease

1. Does the member have a platelet count of less than 50 x 10^9? (Provide documentation of platelet count)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a planned medical or dental procedure with intermediate-to-high bleeding risk within the next 30 days? (Provide date and type of scheduled procedure for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a hematologist, hepatologist, or gastroenterologist?
    a. If yes, continue to #4
    b. If no, clinical review required
4. What is the requested medication?
    a. Doptelet (avatrombopag maleate), continue to #5
    b. Mulpleta (lusutrombopag), continue to #6
5. Is the treatment plan to begin therapy 10-13 days prior to the scheduled procedure and undergo the procedure within 5 to 8 days after the last dose? (Provide documentation of treatment plan and date of scheduled procedure)
    a. If yes, approve for 5 days
    b. If no, clinical review required
6. Does the member have a previous trial with inadequate response, intolerance, or contraindication to Doptelet? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the treatment plan to begin therapy 8-14 days prior to the scheduled procedure and undergo the procedure 2-8 days after the last dose? (Provide documentation of treatment plan and date of scheduled procedure)
    a. If yes, approve for 7 days
    b. If no, clinical review required

Thrombocytopenia in adults with chronic immune thrombocytopenia (ITP)

1. Does the member have a platelet count less than 30 x 109/L (30,000/mm) that was taken within the last 30 days? (Provide platelet count for review)
    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member had an inadequate response, intolerance, or contraindication to glucocorticoids AND splenectomy or rituximab or immunoglobulins for ITP (Inadequate response defined as platelet count fails to each greater than or equal to 50 x 10^9/L (50,000/mm))?? (Provide supporting documentation)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the medication being prescribed by, or in consultation with, a hematologist?

    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Were updated chart notes (within previous 6 months) provided with documentation of significant clinical response to prior therapy received (i.e. platelet count greater than or equal to 50 x 10^9/L (50,000/mm))? (Provide supporting documentation)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with, a hematologist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Doptelet Prescribing Information. Durham, NC: Dova Pharmaceuticals, Inc.; May 2018. Available at: https://www.doptelet.com.
2. Mulpleta Prescribing Information. Florham Park, NJ: Shionogi Inc.; August 2018. Available at: https://www.mulpleta.com/
3. Hayashi H, Beppu T, Shirabe K, Maehara Y, and Baba H. Management of thrombocytopenia due to liver cirrhosis: a review. World J Gastroenterol. 2014; 20(10): 2595-2605.
4. George, PhD, Arnold, MD. Immune thrombocytopenia (ITP) in adults: Second-line and subsequent therapies. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed September 28, 2022.

Last Reviewed: 11/21/18, 5/19/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 6/15/21, 1/1/23

Egrifta SV® (tesamorelin)
Prior Authorization Guidelines

Affected Medication(s)

- Egrifta SV subcutaneous injection

FDA Approved Indication(s)

- Reduction of excess abdominal fat in HIV-infected adult patients with lipodystrophy

Dosing

- 1.4 mg subcutaneously once daily

Initial Authorization Criteria

1. Is the request for continuation of Egrifta SV (tesamorelin) therapy?

    a. If yes, continue to Reauthorization
    b. If no, continue to #2

2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the member currently 18 years of age or older?

    a. If yes, continue to #4
    b. If no, clinical review required

4. Does the member have a confirmed diagnosis of HIV-associated lipodystrophy?

    a. If yes, continue to #5
    b. If no, clinical review required

5. Does the member meet one of the following? (Provide supporting documentation)
    - Male: waist circumference is ≥ 95 cm (37.4 in) and waist-to-hip ratio is ≥ 0.94
    - Female: waist circumference is ≥ 94 cm (37 in) and waist-to-hip ratio is ≥ 0.88

    a. If yes, continue to #6
    b. If no, clinical review required

6. Is the member established on an antiretroviral regimen for at least 8 weeks? (Provide documentation of regimen)

    a. If yes, continue to #7
    b. If no, clinical review required

7. Is the treatment being prescribed by, or in consultation with, an endocrinologist or a physician specializing in the treatment of HIV infection?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 11/16/22, 11/17/23
Effective Date: 1/1/23

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within 1 year) provided with documentation of significant clinical response to therapy such as reduction in visceral adipose tissue measured by waist circumference or computed tomography (CT) scan? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, an endocrinologist or a physician specializing in the treatment of HIV infection?
    a. If yes, approve for 12 months reauthorization unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Egrifta SV® injection [prescribing information]. Montreal, Quebec, Canada: Thera technologies; July 2019.
2. HIV and Lipodystrophy. US Department of Health and Human Services. Available at: https://aidsinfo.nih.gov/understanding-hiv-aids/fact-sheets/22/61/hiv-and-lipodystrophy. Accessed October 18, 2022.
3. Falutz J, Potvin D, Mamputu JC, et al. Effects of tesamorelin, a growth hormone-releasing factor, in HIV infected patients with abdominal fat accumulation: a randomized placebo-controlled trial with a safety extension. J Acquir Immune Defic Syndr. 2010;53(3):311-322.
4. Stanley T, Falutz J, Marsolais C, et al. Reduction in visceral adiposity is associated with an improved metabolic profile in HIV-infected patients receiving tesamorelin.
5. Clin Infect Dis. 2012 Jun;54(11):1642-51.Makimura H, Feldpausch MN, Rope AM, et al. Metabolic effects of a growth hormone-releasing factor in obese subjects with reduced growth hormone secretion: a randomized controlled trial. J Clin Endocrinol Metab. 2012;97(12):4769-4779.

Last Reviewed: 11/16/22, 11/17/23
Effective Date: 1/1/23

Enspryng® (satralizumab-mwge)
Prior Authorization Guidelines

Affected Medication(s)

- Enspryng subcutaneous injection solution

FDA Approved Indication(s)

- Treatment of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive

Dosing

- Recommended loading dose for first three administrations is 120mg by subcutaneous injection at weeks 0, 2, and 4, followed by maintenance dosage of 120mg every four weeks

Initial Authorization Criteria

1. Is the request for continuation of a previously approved Enspryng (satralizumab-mwge) prior authorization with the same indication as the previous approval?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, continue to #4
    b. If not, clinical review required
4. Does the member currently have documented diagnosis of neuromyelitis optica spectrum disorder (NMOSD) and are they anti-aquaporin-4 (AQP4) antibody positive? (Provide supporting documentation of diagnosis and AQP4 status)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the member 18 years of age or older?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Has the member previously trialed at least TWO of the following for 12 weeks or greater with inadequate response, intolerance, or contraindication: azathioprine, methotrexate, and/or mycophenolate? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required

Last Reviewed: 1/20/21, 1/19/22, 3/16/22, 3/17/23, 1/19/24
Effective Date: 3/1/21, 4/15/22, 3/1/24

7. Has the member previously trialed rituximab with inadequate response, intolerance, or contraindication? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Were updated chart notes (within the past 6 months) provided with documentation of significant clinical response? (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. ENSPRYNG™ (satralizumab-mwge) injection,solution, [package insert]. San Francisco, CA: Genentech, Inc.; 2021.
2. Sellner, Johann, et al. "EFNS guidelines on diagnosis and management of neuromyelitis optica." European journal of neurology 17.8 (2010): 1019-1032.
3. Traboulsee, A., et al. "Efficacy and safety of satralizumab monotherapy for relapse prevention in neuromyelitis optica spectrum disorder (NMOSD): Results from SAkuraStar, a double-blind placebo-controlled phase 3 clinical study." Journal of the Neurological Sciences 405 (2019): 171.
4. Yamamura, Takashi, et al. "Efficacy of satralizumab (SA237) in subgroups of patients in SAkuraSky: a phase III double-blind, placebo-controlled, add-on study in patients with neuromyelitis optica spectrum disorder (NMOSD)(S43. 008)." (2019): S43-008.

Last Reviewed: 1/20/21, 1/19/22, 3/16/22, 3/17/23, 1/19/24
Effective Date: 3/1/21, 4/15/22, 3/1/24

Epidiolex® (cannabidiol)
Prior Authorization Guidelines

Affected Medication(s)

- Epidiolex (cannabidiol) solution

FDA Approved Indication(s)

- Treatment of seizures associated with Lennox-Gastaut syndrome (LGS), Dravet syndrome (DS), or tuberous sclerosis complex (TSC) in patients 1 year of age and older

Dosing

- Starting dose: 2.5mg/kg taken twice daily for one week
- Maintenance dose:
  - LGS or DS: 5mg/kg twice daily up to maximum dose 10mg/kg twice daily
  - TSC:12.5 mg/kg twice daily

Initial Authorization Criteria

1. Is the request for continuation of Epidiolex (cannabidiol) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member 1 year of age or older?
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is the member currently taking at least one other antiepileptic drug with inadequate response? (Provide documentation of antiepileptic therapy and seizure frequency)
   a. If yes, continue to #5
   b. If no, clinical review required
5. Is Epidiolex being requested for management of Lennox-Gastaut syndrome (LGS)?
   a. If yes, continue to #6
   b. If no, continue to #7
6. Has the member had a previous trial with inadequate response, intolerance, or contraindication to clobazam? (Provide documentation of trial with inadequate response)
   a. If yes, continue to #7
   b. If no, clinical review required
7. Will the member continue therapy with at least one other antiepileptic drug in combination with Epidiolex (cannabidiol)?

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a. If yes, continue to #8
b. If no, clinical review required

8. Is the medication prescribed by, or in consultation with, a neurologist?

    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Were updated chart notes (within 1 year) with documentation of significant clinical response to prior therapy received? (Significant clinical response is defined by a decrease in seizure frequency compared to pre- treatment baseline)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with, a neurologist?

    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. EPIDIOLEX (cannabidiol) oral solution [package insert]. Carlsbad, CA: Greenwich Biosciences, Inc.; 2018.
2. National Institute of Neurological Disorders and Stroke. Dravet Syndrome Information Page. Available at: https://www.ninds.nih.gov/Disorders/All-Disorders/Dravet-Syndrome-Information-Page.
3. National Institute for Health and Care Excellance (NICE). Epilepsies: diagnosis and management. Available at: https://www.nice.org.uk/guidance/CG137/chapter/Appendix-E-Pharmacological-treatment.
4. Ferrie CD, Patel A. Treatment of lennox-gastaut syndrome. Eur J Paediatr Neurol. 2009 Nov;13(6):493-504.
5. American Academy of Neurology and the American Epilepsy Society. Treatments for Refractory Epilepsy; Guideline Summary for Clinicians. Available at: http://tools.aan.com/professionals/practice/pdfs/clinician_ep_treatment_e.pdf .
6. National Institute of Neurological Disorders and Stroke. Lennox-Gastaut Syndrome Information Page. Available at: https://www.ninds.nih.gov/Disorders/All-Disorders/Lennox-Gastaut-Syndrome-Information-Page.

Last Reviewed: 1/16/19, 5/19/21, 11/16/22, 9/15/23
Effective Date: 2/1/19, 6/15/21, 1/1/23

7. Hancock EC, Cross JH. Treatment of Lennox-Gastaut syndrome. Cochrane Database of Systematic Reviews 2013, Issue 2. Art. No.: CD003277.
8. Wirrell EC. Treatment of Dravet Syndrome. Can J Neurol Sci. 2016 Jun;43 Suppl 3:S13-8. doi: 10.1017/cjn.2016.249. https://www.ncbi.nlm.nih.gov/pubmed/27264138
9. Kim HJ, Kim SH, MD, Kang HC, et al. Adjunctive Levetiracetam Treatment in Pediatric Lennox-Gastaut Syndrome. Pediatr Neurol. 2014 Oct;51(4):527-31. doi: 10.1016/j.pediatrneurol.2014.06.004. Epub 2014 Jun 25. https://www.ncbi.nlm.nih.gov/pubmed/25266616
10. Asadi-Pooya AA. Lennox-Gastaut syndrome: a comprehensive review. Neurol Sci. 2018 Mar;39(3):403-414. doi: 10.1007/s10072-017-3188-y. Epub 2017 Nov 9. https://www.ncbi.nlm.nih.gov/pubmed/29124439

Epogen®, Procrit®, Retacrit® (epoetin alfa)
Prior Authorization Guidelines

Affected Medication(s)

- Epogen (epoetin alfa) injection solution
- Procrit (epoetin alfa) injection solution
- Retacrit (epoetin alfa-epbx) injection solution

FDA Approved Indication(s)

- Treatment of anemia due to chronic kidney disease (CKD), including patients on dialysis and not on dialysis to decrease the need for red blood cell (RBC) transfusion
- Treatment of anemia due to zidovudine administered at ≤ 4200 mg/week in patients with HIV-infection with endogenous serum erythropoietin levels of ≤ 500 mUnits/mL
- Treatment of anemia in patients with non-myeloid malignancies where anemia is due to the effect of concomitant myelosuppressive chemotherapy, and upon initiation, there is a minimum of two additional months of planned chemotherapy
- To reduce the need for allogeneic red blood cell (RBC) transfusions among patients with perioperative hemoglobin > 10 to ≤ 13 g/dL who are at high risk for perioperative blood loss from elective, non-cardiac, nonvascular surgery. Epoetin alfa is not indicated for patients who are willing to donate autologous blood pre-operatively

Dosing

- Refer to package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of epoetin alfa therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the medication being requested for an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Have serum ferritin, transferrin saturation, hemoglobin (Hb), and hematocrit (Hct) labs been completed within 30 days of planned administration? (Provide labs for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a serum ferritin ≥ 100 ng/mL (mcg/L) and transferrin saturation (TSAT) ≥ 20%? (Provide labs for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a hemoglobin (Hb) < 10 g/dL and/or Hematocrit (Hct) < 30%?
    a. If yes, continue to #7
    b. If no, continue to #6

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Effective Date: 1/1/19, 9/1/21, 4/15/24

6. Is the medication being requested to reduce allogeneic blood transfusions in elective, non-cardiac, non-vascular surgery?
    a. If yes, continue to #7
    b. If no, clinical review required
7. Have other causes of anemia (e.g. hemolysis, bleeding, vitamin deficiency, etc.) been ruled out?
    a. If yes, continue to #8
    b. If no, clinical review required
8. Is epoetin alfa-apbx being requested?
    a. If yes, continue to #10
    b. If no, continue to #9
9. Does the member have a contraindication or history of intolerance to a trial of epoetin alfa-apbx? (Provide supporting documentation of contraindication and/or intolerance)
    a. If yes, continue to #10
    b. If no, clinical review required
10. Which indication is epoetin alfa being requested for? (Record submitted diagnosis and review all criteria based on the submitted diagnosis)
    a. Anemia secondary to myelodysplastic syndrome (MDS), continue to corresponding criteria
    b. Anemia secondary to Myeloproliferative Neoplasms (MPN) – Myelofibrosis, continue to corresponding criteria
    c. Anemia secondary to chemotherapy treatment, continue to corresponding criteria
    d. Anemia secondary to chronic kidney disease (non-dialysis patients), approve for 3 months unless otherwise specified
    e. Anemia secondary to zidovudine treated, HIV-infected patients, continue to corresponding criteria
    f. Reduction of allogeneic blood transfusions in elective, non-cardiac, non-vascular surgery, continue to corresponding criteria
    g. Other Indication, continue to corresponding criteria

<u>Anemia secondary to myelodysplastic syndrome (MDS)</u>

1. Does the member have symptomatic anemia? (Examples include: exertional dyspnea, dyspnea at rest, fatigue, lethargy, confusion, etc.) (Provide supporting documentation of symptoms)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member's endogenous serum erythropoietin level ≤ 500 mUnits/mL? (Provide serum erythropoietin level for review)
    a. If yes, approve for 45 days unless otherwise specified
    b. If no, clinical review required

<u>Anemia secondary to Myeloproliferative Neoplasms (MPN) – Myelofibrosis</u>

1. Is the members endogenous serum erythropoietin level < 500 mUnits/mL? (Provide serum erythropoietin level for review)
    a. If yes, approve for 45 days unless otherwise specified
    b. If no, clinical review required

Anemia secondary to chemotherapy treatment

1. Is the member receiving concurrent myelosuppressive chemotherapy for non-myeloid malignancies?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the therapy intention of the chemotherapy curative?
    a. If yes, clinical review required
    b. If no, continue to #3
3. Are there two or more additional months of planned chemotherapy remaining? (Provide documentation of treatment plan)
    a. If yes, approve for 6 months or until completion of chemotherapy course, whichever is less
    b. If no, clinical review required

Anemia secondary to zidovudine treated, HIV-infected patients

1. Does the member have an endogenous serum erythropoietin level ≤ 500 mUnits/mL AND is the member currently receiving zidovudine administered at ≤ 4200 mg/week? (Provide serum erythropoietin level or current zidovudine dose for review)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reduction of allogeneic blood transfusions in elective, non-cardiac, non-vascular surgery

1. Does the member have a hemoglobin (Hb) level between 10 g/dL and 13 g/dL and/or is the hematocrit (Hct) between 30% and 39%? (Provide supporting documentation with Hb and Hct lab values for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the surgery high-risk for perioperative blood loss? (i.e. expected to lose >2 units of blood)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is patient unwilling to donate autologous blood pre-operatively?
    a. If yes, approve for 45 days unless otherwise specified
    b. If no, clinical review required

Other Indications

1. Is the requested use supported by major compendia? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member tried and had an inadequate response OR dose the member have a contradiction to ALL standard treatment options for the requested indication? (Provide all prior treatment history, contraindication if appropriate, and treatment plan)
    a. If yes, approve for 45 days unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Was the last dose of epoetin alfa less than 60 days ago? (Provide date of last dose)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Were updated chart notes (within 1 year) provided with documentation of significant clinical response to therapy? (Provide documentation of clinical response)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is there documentation of an absence of unacceptable toxicity from the drug? (Examples include severe cardiovascular events (stroke, myocardial infarction, thromboembolism, uncontrolled hypertension), tumor progression or recurrence in members with cancer, seizures, pure red cell aplasia, severe cutaneous reactions (erythema multiforme, Stevens-Johnson syndrome/toxic epidermal necrolysis), “gasping syndrome” (central nervous system depression, metabolic acidosis, gasping respirations) due to benzyl alcohol preservative, etc.)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Were lab values obtained within 30 days of the date of administration (unless otherwise indicated)? (Provide updated lab result for review)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have adequate iron stores as demonstrated by serum ferritin ≥ 100 ng/mL (mcg/L) and transferrin saturation (TSAT) ≥ 20% measured within the previous 3 months? (Provide lab result for review)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Have other causes of anemia (e.g. hemolysis, bleeding, vitamin deficiency, etc.) been ruled out?
    a. If yes, continue to #8
    b. If no, clinical review required
8. Does the member meet the clinical requirements for their corresponding diagnosis as defined below? (Provide supporting documentation)
    - Anemia secondary to myelodysplastic syndrome (MDS) with Hemoglobin (Hb) < 12 g/dL and/or Hematocrit (Hct) < 36%
    - Anemia secondary to myeloproliferative neoplasms (MF, post-PV myelofibrosis, post-ET myelofibrosis) with Hemoglobin (Hb) < 10 g/dL and/or Hematocrit (Hct) < 30%
    - Reduction of allogeneic blood transfusions in elective, non-cardiac, non-vascular surgery with Hemoglobin (Hb) between 10 g/dL and 13 g/dL and/or Hematocrit (Hct) between 30% and 39%
    - Anemia secondary to palliative myelosuppressive chemotherapy for non-myeloid malignancies with Hemoglobin (Hb) <10 g/dL and/or Hematocrit (Hct) < 30% and requesting

epoetin alfa to be used concurrently with chemotherapy with minimum two additional months of therapy remaining
- Anemia secondary to zidovudine treated, HIV-infected patients with Hemoglobin (Hb) < 12 g/dL and/or Hematocrit (Hct) < 36% AND receiving zidovudine administered at ≤ 4200 mg/week
- Anemia secondary to chronic kidney disease with hemoglobin (Hb) <12 g/dL and/or hematocrit (Hct) <36% in pediatric patients OR hemoglobin (Hb) <11 g/dL and/or hematocrit (Hct) <33% in adult patients
- Hemoglobin (Hb) < 11 g/dL and/or Hematocrit (Hct) < 33% for all other indications
- Use supported my major compendia

a. If yes, approve for 1 year unless otherwise specified
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Procrit [package insert]. Horsham, PA; Janssen Products, LP; September 2017. Accessed March 2018.
2. Epogen [package insert]. Thousand Oaks, CA; Amgen, Inc; September 2017. Accessed March 2018.
3. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) epoetin alfa. National Comprehensive Cancer Network, 2018. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed March 2018.
4. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Cancer-and Chemotherapy-Induced Anemia Version 2.2018. National Comprehensive Cancer Network, 2017. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed March 2018.
5. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Myelodysplastic Syndrome Version 2.2018. National Comprehensive Cancer Network, 2017. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed March 2018.
6. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Myeloproliferative Neoplasms Version 2.2018. National Comprehensive Cancer Network, 2017. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed March 2018

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Evenity® (romosozumab-aqqg)
Prior Authorization Guidelines

Affected Medication(s)

- Evenity subcutaneous solution

FDA Approved Indication(s)

- Treatment of osteoporosis in postmenopausal women at high risk for fracture, defined as a history of osteoporotic fracture, or multiple risk factors for fracture; or patients who have failed or are intolerant to other available osteoporosis therapy.

Dosing

- 210 mg subcutaneously every month (limited to 12 months cumulative use per lifetime)

Initial Authorization Criteria

1. Is the request for continuation of Evenity therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member a postmenopausal female? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have osteoporosis defined as one of the following? (Provide supporting documentation including DXA report within the last 2 years)
    - Hip DXA (femoral neck or total hip) or lumbar spine T-score less than or equal to -2.5 and/or forearm DXA 33% (one-third) radius
    - T-score less than or equal to -1 or low bone mass AND a history of fragility fracture to the hip or spine
    - T-score between -1 and -2.5 with a FRAX 10-year probability for major fracture ≥20% or hip fracture ≥3%

    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a high risk for fracture as defined by one or more of the following? (Provide supporting documentation)

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- History of an osteoporotic fracture as an adult
- Parental history of hip fracture
- Low BMI
- Rheumatoid arthritis
- Alcohol intake of 3 or more drinks per day
- Current smoking
- History of oral glucocorticoids ≥ 5 mg/day of prednisone (or equivalent) for > 3 months in lifetime
- Early Menopause

a. If yes, continue to #7
b. If no, clinical review required

7. Does the member have five years of cumulative treatment with bisphosphonates? (Provide all prior therapy history)

    a. If yes, continue to #11
    b. If no, continue to #8

8. Does the member have a trial with insufficient response to at least 12 months of bisphosphonate therapy (oral or IV) as defined by a decrease in T-score from baseline or member had a fracture while on bisphosphonate therapy? (Provide past relevant medication list with documentation of response to therapy)

    a. If yes, continue to #11
    b. If no, continue to #9

9. Does the member have a contraindication or intolerance to oral bisphosphonates? (Provide supporting documentation)

    a. If yes, continue to #10
    b. If no, clinical review required

10. Does the member have a contraindication or intolerance to IV bisphosphonates? (Provide supporting documentation)

    a. If yes, continue to #11
    b. If no, clinical review required

11. Does the member have a previous trial with insufficient response to at least 12 months of Prolia therapy as defined by a decrease in T-score from baseline or member had a fracture while on Prolia therapy OR an intolerance or contraindication to Prolia? (Provide past relevant medication list with documentation of response to therapy)

    a. If yes, continue to #12
    b. If no, clinical review required

12. Is the member currently supplementing with at least 1,000 mg of calcium and 400 IU of vitamin D daily that will be continued throughout therapy? (Provide list of current relevant medications)

    a. If yes, continue to #13
    b. If no, clinical review required

13. Is the requested medication being prescribed by, or in consultation with, an endocrinologist, rheumatologist, or specialist experienced in treatment of osteoporosis?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member previously had a cumulative lifetime treatment duration of 12 months of Evenity?

    a. If yes, clinical review required
    b. If no, continue to #3

3. Does the member demonstrate positive clinical response to therapy as defined by absence of fractures and/or an increase in bone mineral density from pretreatment baseline? (Provide updated DXA report and other supporting documentation)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is the requested medication being prescribed by, or in consultation with, an endocrinologist, rheumatologist, or specialist experienced in treatment of osteoporosis?

    a. If yes, approve for a maximum of 6 additional months (lifetime max is 1 year of therapy)
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm
2. EVENITY (romosozumab-aqqg) Injectable solution [package insert]. Thousand Oaks, CA: Amgen Inc.; October 2021.
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Last Reviewed: 11/20/19, 5/19/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/20, 7/15/22, 6/15/24

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Evrysdi® (risdiplam)
Prior Authorization Guidelines

Affected Medication(s)

- Evrysdi (risdiplam) oral powder for solution

FDA Approved Indication(s)

- Treatment of spinal muscular atrophy (SMA) in pediatric and adult patients

Dosing

- Less than 2 months of age: 0.15 mg/kg/day
- 2 months to less than 2 years of age: 0.2 mg/kg/day
- 2 years of age and older, weighing less than 20kg: 0.25 mg/kg/day
- 2 years of age and older, weight 20kg or more: 5 mg/day

Initial Authorization Criteria

1. Is the request for renewal of a previously approved Evrysdi (risdiplam) prior authorization with the same indication?

    a. If yes, continue to Reauthorization
    b. If no, continue to #2

2. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Does the member have a confirmed diagnosis of SMA type 1, 2, or 3, with four or fewer copies of SMN2? (Provide supporting documentation)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Does the member have advanced SMA disease defined as ventilator dependence >16 hours/day or tracheostomy? (Provide supporting documentation)

    a. If yes, clinical review required
    b. If no, continue to #5

5. Was baseline motor function assessed by one of the following? (Provide supporting documentation)

    - Hammersmith Infant Neurological Examination (HINE-2)
    - Motor Function Measure 32 (MFM32)
    - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)
    - Upper Limb Module (ULM)
    - Revised Upper Limb Module (RULM)
    - Hammersmith Functional Motor Scale (HFMS)
    - Bayley Scales of Infant and Toddler Development – Third Edition (BSID-III)

    a. If yes, continue to #6
    b. If no, clinical review required

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6. Does the member have a history of prior treatment with Zolgensma or will this medication be used in combination with Spinraza?
    a. If yes, clinical review required
    b. If no, continue to #7
7. Is the requested medication being prescribed by, or in consultation with, a neurologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is Evrysdi (risdiplam) being requested for an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within 1 month of reauthorization request date) with documentation of significant clinical response to therapy defined as improvement from baseline in one of the following received? (Provide supporting documentation)
        - Hammersmith Infant Neurological Examination (HINE-2)
        - Motor Function Measure 32 (MFM32)
        - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)
        - Upper Limb Module (ULM)
        - Revised Upper Limb Module (RULM)
        - Hammersmith Functional Motor Scale (HFMS)
        - Bayley Scales of Infant and Toddler Development – Third Edition (BSID-III)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the requested medication being prescribed by, or in consultation with, a neurologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Drugs@FDA: FDA Approved Drug Products. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 22 Mar 2023].
2. EVRYSDI™ (risdiplam)oral solution [package insert]. San Francisco, CA: Genentech, Inc 2022.
3. Glascock, Jacqueline, et al. "Treatment algorithm for infants diagnosed with spinal muscular atrophy through newborn screening." Journal of neuromuscular diseases 5.2 (2018): 145-158.

Last Reviewed: 3/17/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 5/1/21, 7/15/22, 7/1/23, 6/15/24

4. Glascock, Jacqueline, et al. "Revised recommendations for the treatment of infants diagnosed with spinal muscular atrophy via newborn screening who have 4 copies of SMN2." Journal of neuromuscular diseases 7.2 (2020): 97.
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Last Reviewed: 3/17/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 5/1/21, 7/15/22, 7/1/23, 6/15/24

Exjade®, Jadenu® (deferasirox)
Prior Authorization Guidelines

Affected Medication(s)

- Exjade oral tablet for suspension
- deferasirox oral tablet for suspension
- Jadenu oral tablet/sprinkle granules
- deferasirox oral tablet/sprinkle granules

FDA Approved Indication(s)

- Treatment of chronic iron overload due to blood transfusions (transfusional hemosiderosis) in patients 2 years of age and older
- Treatment of chronic iron overload in patients 10 years of age and older with non-transfusion-dependent thalassemia (NTDT) syndromes and with a liver iron concentration (LIC) of at least 5 milligrams of iron per gram of liver dry weight (mg Fe/g dw) and a serum ferritin greater than 300 mcg/L

Dosing

- Refer to package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of Exjade or Jadenu (deferasirox) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consult with, a hematologist?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member concurrently using other iron chelators (Examples include: Ferriprox, Desferal, Depen, and Cuprimine)?
    a. If yes, clinical review required
    b. If no, continue to #5
5. What is the indication that the medication is being requested for?
    a. Chronic iron overload due to blood transfusions, see corresponding criteria
    b. Chronic iron overload due to non-transfusion-dependent thalassemia (NTDT), see corresponding criteria

Chronic iron overload due to blood transfusions

1. Is the member 2 years of age or older?
    a. If yes, continue to #2

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b. If no, clinical review required

2. Does the member have a history of transfusion with at least 100 mL/kg of packed red blood cells? (Provide supporting documentation for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is serum ferritin consistently greater than 1000 mcg/L? (Provide supporting labs for review)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Chronic iron overload due to non-transfusion-dependent thalassemia (NTDT)

1. Is the member 10 years of age or older?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a serum ferritin level of > 300 mcg/L and a liver iron concentration of ≥ 5 mg Fe/g of liver dry weight? (Provide documentation of lab result)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by, or in consult with a hematologist?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member concurrently using other iron chelators?
    a. If yes, clinical review required
    b. If no, continue to #4
4. What is the indication that the medication is being requested for?
    a. Chronic iron overload due to blood transfusions, see corresponding criteria
    b. Chronic iron overload due to non-transfusion-dependent thalassemia (NTDT), see corresponding criteria

Chronic iron overload due to blood transfusions

1. Does the member have a serum ferritin level ≥ 500 mcg/L? (Provide documentation of serum ferritin level within the past 30 days)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 10/17/18, 1/21/20, 9/15/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 4/15/24

Chronic iron overload due to non-transfusion-dependent thalassemia (NTDT)

1. Does the member have a liver iron concentration of ≥ 3 mg Fe/g of liver dry weight? (Provide documentation of lab result within the past 90 days)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Exjade Prescribing Information. East Hanover, NJ: Novartis Pharmaceuticals Corporation; July 2020. Available at http://www.us.exjade.com/.
2. Jadenu Prescribing Information. East Hanover, NJ: Novartis Pharmaceuticals Corporation; July 2020. Available at https://www.jadenu.com/.
3. Oken M, Creech R, Tormey D, et al. Toxicity and response criteria of the Eastern Cooperative Oncology Group. Am J Clin Oncol. 1982;5:649-655. http://ecog-acrin.org/resources/ecog-performance-status.
4. Musallam KM, Angastiniotis M, Eleftheriou A, Porter JB. Cross-talk between available guidelines for the management of patients with beta-thalassemia major. Acta Haematol. 2013; 130: 64-73. DOI: 10.1159/000345734.
5. Hoffbrand AV, Taher A, Cappellini MD. How I treat tranfusional iron overload. Blood. November 1, 2012; 120(18): 3657-3669.
6. Taher AT, Viprakasit V, Musallam KM, Cappellini MD. Treating iron overload in patients with non-transfusion-dependent thalassemia: Critical Reivew. Am J Hematol. 2013; 88: 409-415. DOI: 10.1002/ajh.23405.

Last Reviewed: 10/17/18, 1/21/20, 9/15/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 4/15/24

Ferriprox® (deferiprone)
Prior Authorization Guidelines

Affected Medication(s)

- Ferriprox oral solution
- Ferriprox oral tablet
- Deferiprone oral tablet
- Ferriprox Twice-A-Day tablet

FDA Approved Indication(s)

- Treatment of transfusional iron overload in adults and pediatric patients ≥8 years of age (tablets) or adults and pediatric patients ≥3 years of age (solution) with thalassemia syndromes, sickle cell disease, or other anemias

Dosing

- 25 mg/kg to 33 mg/kg orally three times daily
- Twice daily formulation: 37.5 mg/kg to 50 mg/kg orally twice daily

Initial Authorization Criteria

1. Is the request for continuation of Ferriprox (deferiprone) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have an ANC greater than 1.5 x 10^9/L and a baseline serum ferritin level greater than 1,000 mcg/L? (Provide ANC and serum ferritin level for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a trial with inadequate response, intolerance, or contraindication to an iron chelator? (Examples include Desferal, Exjade, or Jadenu) (Inadequate response defined as serum ferritin > 2,500 mcg/L) (Provide documentation of trial and response, intolerance, or contraindication)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, a hematologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

Last Reviewed: 11/21/18, 11/18/20, 3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 12/15/20, 4/15/22, 7/1/23, 6/15/24

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member had a positive clinical response to therapy as defined by a ≥20% decline in serum ferritin within one year of starting therapy and is ANC being monitored weekly while on therapy? (Provide supporting documentation for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a hematologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Ferriprox [prescribing information]. Toronto, Ontario: ApoPharma USA, Inc.; April 2021.
2. Galanello R. Deferiprone in the treatment of transfusion-dependent thalassemia: a review and perspective. Therapeutics and Clinical Risk Management 2007;3(5):795- 805
3. Porter JB, Shah FT. Iron overload in thalassemia and related conditions: therapeutic goals and assessment of response to chelation therapies. Hematol Oncol Clin N Am 2010; 24;1109-1130
4. Cappellini MD, Cohen A, Porter, J, et al. A short guide for the management of transfusion dependent thalassaemia. 3rd Edition. Available at: Short-GUIDE-low-res.pdf (thalassaemia.org.cy). Accessed on March, 2022.

Last Reviewed: 11/21/18, 11/18/20, 3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 12/15/20, 4/15/22, 7/1/23, 6/15/24

Fertility Agents
Prior Authorization Guidelines

Affected Medication(s)

- Cetrotide (cetrorelix acetate) subcutaneous powder for solution
- chorionic gonadotropin intramuscular powder for solution
- clomiphene oral tablet
- Crinone 8% (progesterone) vaginal gel
- Endometrin (progesterone) vaginal tablet
- Follistim AQ (follitropin beta) subcutaneous solution
- Fyremadel (ganirelix acetate) subcutaneous solution
- ganirelix acetate subcutaneous solution
- Gonal-F (follitropin alfa) subcutaneous powder for solution
- Menopur (menotropins) subcutaneous powder for solution
- Novarel (chorionic gonadotropin) intramuscular powder for solution
- Ovidrel (chorionic gonadotropin alfa, recombinant) subcutaneous solution
- Pregnyl (chorionic gonadotropin) intramuscular powder for solution
- Saizen (somatropin) subcutaneous powder for solution

FDA Approved Indication(s)

- Refer to package insert for specific indications for each medication

Dosing

- Refer to corresponding package insert for specific dosing recommendations

Fertility Benefit Note

- Services for removal and preservation of oocytes and sperm prior to gonadotoxic treatment are not subject to the lifetime infertility maximum. The Plan covers these services at the applicable Member Cost Share when Prior Authorization is obtained by the Plan

Authorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member meet one or more of the following conditions? (**Please note:** Per plan provisions, coverage of medications under the fertility benefit is limited to services rendered by OHSU Fertility Clinic)

    Female or assigned female at birth:
    - 35 years of age or younger with failure to conceive after regular unprotected sexual intercourse for 1 year or more
    - 35 years of age or older with failure to conceive after regular unprotected sexual intercourse for 6 months or more

Last Reviewed: 12/19/18, 4/2/19, 5/19/21, 1/1/22, 1/20/23, 1/19/24
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- Requiring medical assistance to conceive (e.g. oligoovulation due to PCOS) including same-sex couples and single persons using partner or donor gametes
- Prior diagnosis of infertility
- Recurrent pregnancy loss defined as two or more pregnancy losses (miscarriages) prior to 20 weeks gestation
- Prior cycle of in vitro fertilization or intracytoplasmic sperm injection with failure
- Premature ovarian insufficiency or decreased ovarian reserve due to gonadotoxic therapy
- History of bilateral oophorectomy
- Carrier of a genetic disease (or affected by a genetic disease) and/or has a partner who is a carrier of a genetic disease (or affected by a genetic disease) and at risk of having a child with a genetic disease

Male partner or assigned male at birth:

- Infertility due to gonadotoxic therapy (e.g., orchiectomy, chemotherapy, or lupus therapy)
- Non-obstructive azoospermia or severe oligospermia
- Paraplegia and sperm retrieval required to achieve pregnancy
- Carrier of a genetic disease (or affected by a genetic disease) and/or has a partner who is a carrier of a genetic disease (or affected by a genetic disease) and at risk of having a child with a genetic disease

a. If yes, approve for 12 months or up to duration of benefit
b. If no, continue to #3

3. Is the male partner (or assigned male at birth) HIV positive and meet BOTH the following requirements?

- Adherent with antiretroviral therapy regimen
- Washed sperm needed for insemination to prevent HIV transmission to female partner

a. If yes, approve for 12 months or up to duration of benefit
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Cetrotide (cetrorelix acetate) [Prescribing Information]. Rockland, MA: EMD Serono, Inc. September 2018.
2. Chorionic gonadotropin [Prescribing Information]. Lake Zurich, IL: Fresenius Kabi USA, LLC. July 2017.
3. Clomiphene [Prescribing Information]. Chestnut Ridge, NY: Par Pharmaceuticals, Inc. October 2017.
4. Crinone (progesterone) [Prescribing Information]. Parsippany, NJ: Actavis Pharma, Inc. November 2017 2016.
5. Endometrin (progesterone) [Prescribing Information]. Parsippany, NJ: Ferring Pharmaceuticals Inc. October 2018.

Last Reviewed: 12/19/18, 4/2/19, 5/19/21, 1/1/22, 1/20/23, 1/19/24
Effective Date: 1/1/19, 4/2/19, 7/1/21, 1/1/22, 3/15/23, 3/1/24

6. Follistim AQ (follitropin beta) [Prescribing Information]. Whitehouse Station, NJ: Organon USA Inc. December 2014.
7. Ganirelix acetate [Prescribing Information]. Whitehouse Station, NJ: Organon USA Inc. May 2018.
8. Gonal-F (follitropin alfa) [Prescribing Information]. Rockland, MA: EMD Serono, Inc. May 2018.
9. Menopur (menotropins) [Prescribing Information]. Parsippany, NJ: Ferring Pharmaceuticals Inc. May 2018.
10. Novarel (chorionic gonadotropin) [Prescribing Information]. Parsippany, NJ: Ferring Pharmaceuticals Inc. November 2018.
11. Ovidrel (chorionic gonadotropin alfa, recombinant) [Prescribing Information]. Rockland, MA: EMD Serono, Inc. June 2018.
12. Pregnyl (chorionic gonadotropin) [Prescribing Information]. Whitehouse Station, NJ: Organon USA Inc. January 2015.

Last Reviewed: 12/19/18, 4/2/19, 5/19/21, 1/1/22, 1/20/23, 1/19/24
Effective Date: 1/1/19, 4/2/19, 7/1/21, 1/1/22, 3/15/23, 3/1/24

Filspari (sparsentan)
Prior Authorization Guidelines

Affected Medication(s)

- Filspari (sparsentan) oral tablet

FDA Approved Indication(s)

- To reduce proteinuria in adults with primary immunoglobulin A nephropathy (IgAN) at risk of rapid disease progression

Dosing

- 200 mg once daily for 14 days, then increase to the recommended dose of 400 mg daily if tolerated

Initial Authorization Criteria

1. Is the request for continuation of Filspari (sparsentan) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of primary immunoglobulin A nephropathy as proven by biopsy? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have documentation of estimated glomerular filtration rate (eGFR) of 30 mL/min/1.73m^2 or greater? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have documentation of a total urine protein greater than or equal to 1 gram/day? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Did the member have an inadequate response to a 12 week trial of the following classes of medications at maximally indicated doses unless intolerance or contraindication is present to all three classes? (Provide supporting documentation)
    - Angiotensin-converting enzyme inhibitor (i.e. lisinopril, benazepril, enalapril); OR

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- Angiotensin receptor blocker (i.e. irbesartan, losartan); AND
- Sodium-glucose Cotransporter-2 inhibitor (i.e. Farxiga)

a. If yes, continue to #8
b. If no, clinical review is required

8. Is Filspari (sparsentan) being prescribed by, or in consult with, a nephrologist?

a. If yes, approve for 6 months
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)

a. If yes, continue to #2
b. If no, clinical review required

2. Were updated chart notes (within past year) provided with documentation of significant clinical response to prior therapy received? (i.e. reduction in UPCR or UACR, increased or stable eGFR)

a. If yes, continue to #3
b. If no, clinical review required

3. Is Filspari (sparsentan) being prescribed by, or in consult with, a nephrologist?

a. If yes, approve for 12 months
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Filspari (sparsentan) tablets, [package insert]. San Diego, CAC:\Users\gouldl\Desktop\Rebate Compiles\Commercial: Travere Therapeutics, Inc; 2023.

2. Drugs@FDA: FDA Approved Drug Products. 2023. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 22 Mar. 2023].

3. KDIGO Glomerular Diseases Work Group. 2021. KDIGO 2021 Clinical Practice Guideline for the Management of Glomerular Diseases. Kidney International Supplements. 2021;11(2):1-221.

4. Wheeler, D. C., Toto, R. D., Stefansson, B. V., Jongs, N., Chertow, G. M., Greene, T., & Committees, D. C. T. (2021). A pre-specified analysis of the DAPA-CKD trial demonstrates the effects of dapagliflozin on major adverse kidney events in patients with IgA nephropathy. Kidney International, 100(1), 215-224.

Last Reviewed: 5/19/23, 5/17/24
Effective Date: 7/15/23, 6/15/24

Fintepla® (fenfluramine)
Prior Authorization Guidelines

Affected Medication(s)

- Fintepla oral solution

FDA Approved Indication(s)

- Treatment of seizures associated with Dravet syndrome and Lennox-Gastaut syndrome in patients 2 years of age and older

Dosing

- Initial starting dose: 0.1 mg/kg twice daily, which can be increased weekly based on efficacy and tolerability
- Patients not on concomitant Diacomit (stiripentol): The maximum daily maintenance dosage of Fintepla is 0.35 mg/kg twice daily (maximum daily dosage of 26 mg)
- Patients taking concomitant Diacomit (stiripentol) plus clobazam: The maximum daily maintenance dosage of Fintepla for patients taking these medications is 0.2 mg/kg twice daily (maximum daily dosage of 17 mg)

Initial Authorization Criteria

1. Is the request for continuation of a previously approved Fintepla (fenfluramine) prior authorization and indication is for the same as previous approval?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member age 2 years or older?
    a. If yes, continue #4
    b. If no, clinical review required
4. Will the member continue therapy with at least one other antiepileptic drug or antiepileptic treatment (i.e. vagal nerve stimulation or ketogenic diet) in combination with Fintepla? (Provide treatment regimen)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, continue to #6
    b. If no, clinical review required
6. What is the indication Fintepla is being requested for?
    a. Dravet syndrome, continue to corresponding criteria
    b. Lennox-Gastaut syndrome, continue to corresponding criteria

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Dravet Syndrome

1. Has the member previously trialed valproate, topiramate, and clobazam (unless intolerance or contraindication provided) and continued to have 4 or more convulsive seizures per month despite optimized therapy? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member previously had an inadequate response, intolerance or contraindication to Diacomit in combination with clobazam? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Lennox-Gastaut Syndrome

1. Has the member previously had an inadequate response, intolerance or contraindication to at least two other antiseizure medications (i.e. felbamate, clobazam, lamotrigine, topiramate, valproic acid, levetiracetam, zonisamide, etc.)? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within the past 12 months) with documentation of at least a 50% decrease in the frequency of convulsive seizures compared to pre-therapy baseline? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member continued therapy with at least one other antiepileptic drug or antiepileptic treatment (i.e. vagal nerve stimulation or ketogenic diet) in combination with Fintepla? (Provide treatment regimen)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by or in consultation with a neurologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

Last Reviewed: 9/16/20, 9/15/21, 1/20/23, 1/19/24
Effective Date: 11/15/20, 11/1/21, 3/15/23, 3/1/24

References:

1. FINTEPLA® (fenfluramine) oral solution [package insert]. Emeryville, CA: Zogenix, Inc; September 2023.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 26 November. 2023].
3. Lagae, Lieven, et al. "Fenfluramine hydrochloride for the treatment of seizures in Dravet syndrome: a randomised, double-blind, placebo-controlled trial." The Lancet 394.10216 (2019): 2243-2254.
4. Nabbout, Rima, et al. "Fenfluramine for treatment-resistant seizures in patients with Dravet syndrome receiving stiripentol-inclusive regimens: a randomized clinical trial." JAMA neurology 77.3 (2020): 300-308.
5. National Institute for Health and Care Excellence; Epilepsies: Diagnosis and Management Clinical Guidelines. NICE Guideline. May 2021. Available at: https://www.nice.org.uk/guidance/cg137
6. Wilmhurst JM, Gaillard WD, Vinayan KP, et al. Summary of recommendations for the management of infantile seizures: Task force report for the ILAE commission of pediatrics. Epilepsia. 2015; 56(8):1185-1197
7. Wirrell, Elaine C., et al. "Optimizing the diagnosis and management of Dravet syndrome: recommendations from a North American consensus panel." Pediatric neurology 68 (2017): 18-34.
8. Asadi-Pooya AA. Lennox-Gastaut syndrome: a comprehensive review. Neurol Sci. 2018 Mar;39(3):403-414. doi: 10.1007/s10072-017-3188-y. Epub 2017 Nov 9. https://www.ncbi.nlm.nih.gov/pubmed/29124439
9. National Institute of Neurological Disorders and Stroke. Lennox-Gastaut Syndrome Information Page. Available at: https://www.ninds.nih.gov/Disorders/All-Disorders/Lennox-Gastaut-Syndrome-Information-Page.
10. Hancock EC, Cross JH. Treatment of Lennox-Gastaut syndrome. Cochrane Database of Systematic Reviews 2013, Issue 2. Art. No.: CD003277.

Last Reviewed: 9/16/20, 9/15/21, 1/20/23, 1/19/24
Effective Date: 11/15/20, 11/1/21, 3/15/23, 3/1/24

Fuzeon® (enfuvirtide), Rukobia® (fostemsavir tromethamine), Sunleca® (lenacapavir) Prior Authorization Guidelines

zAffected Medication(s)

- Fuzeon (enfuvirtide) subcutaneous solution
- Rukobia (fostemsavir tromethamine) oral tablet
- Sunleca (lenacapavir) oral tablet

FDA Approved Indication(s)

- **Fuzeon:** Treatment of HIV-1 infection, in combination with other antiretroviral agents, in treatment-experienced patients with evidence of HIV-1 replication despite ongoing antiretroviral therapy
- **Rukobia, Sunlenca:** Treatment of HIV-1 infection, in combination with other antiretroviral agents, in heavily treatment-experienced adults with multidrug-resistant HIV-1 infection failing their current antiretroviral regimen due to resistance, intolerance, or safety considerations

Dosing

- Refer to corresponding package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of the same antiretroviral agent for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for an FDA approved or major compendia supported indication?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member currently have documented resistance or contraindications to 3 or more different classes of antiretrovirals? (examples include: NRTIs, INSTIs, PIs, NNRTIs, CCR5 antagonist) (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Will the requested medication be taken in combination with an optimized antiviral background regimen including one or more other antiretroviral medications? (Provide treatment regimen)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, an infectious disease specialist or provider experienced in the treatment of HIV?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

Last Reviewed: 9/16/20, 9/15/21, 3/17/23, 3/15/24
Effective Date: 11/15/20, 11/1/21, 6/1/23, 4/15/24

1. Is the requested indication Food and Drug Administration (FDA) approved or supported by major compendia?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within the past 6 months) provided with documentation of decreased viral load compared to pre-therapy baseline? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Will the requested medication continue to be taken in combination with an optimized antiviral background regimen including one or more other antiretroviral medications? (Provide treatment regimen)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by or in consultation with an infectious disease specialist or provider experienced in the treatment of HIV?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. RUKOBIA (fostemsavir) oral extended-release tablet [package insert]. Triangle Park, NC: Viiv Healthcare; January 2022.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 27 July. 2020].
3. US Department of Health and Human Services. "Guidelines for the use of antiretroviral agents in adults and adolescents with HIV." (2019).
4. World Health Organization. "Guidelines for managing advanced HIV disease and rapid initiation of antiretroviral therapy, July 2017." (2017).
5. Kozal M, Aberg J, Pialoux G, et al. Fostemsavir in Adults with Multidrug-Resistant HIV-1 Infection. N Engl J Med. 2020;382(13):1232-1243. doi:10.1056/NEJMoa1902493
6. Lagishetty C, Moore K, Ackerman P, Llamoso C, Magee M. Effects of Temsavir, Active Moiety of Antiretroviral Agent Fostemsavir, on QT Interval: Results from a Phase I Study and an Exposure-Response Analysis. Clin Transl Sci. 2020;13(4):769-776. doi:10.1111/cts.12763

Last Reviewed: 9/16/20, 9/15/21, 3/17/23, 3/15/24
Effective Date: 11/15/20, 11/1/21, 6/1/23, 4/15/24

Galafold® (migalastat hydrochloride)
Prior Authorization Guidelines

Affected Medication(s)

- Galafold oral capsule

FDA Approved Indication(s)

- Treatment of adults with a confirmed diagnosis of Fabry disease and an amenable galactosidase alpha gene (GLA) variant based on in vitro assay data

Dosing

- 123 mg orally once every other day

Initial Authorization Criteria

1. Is the request for continuation of Galafold (migalastat hydrochloride) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a diagnosis of Fabry disease that is confirmed by biochemical and/or molecular genetic testing? (Provide genetic testing results for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Were baseline plasma or urinary globotriaosylceramide (Gb3/GL-3), globotriaosylsphingosine (lyso-Gb3), or kidney interstitial capillary cell globotriaosylceramide (KIC GL-3) labs provided?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a GLA variant based on in vitro assay that is considered amenable? (Provide documentation of amenable GLA variant)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the member female?
    a. If yes, continue to #8
    b. If no, continue to #9

Last Reviewed: 12/19/18, 11/18/20, 9/15/21, 11/16/22, 9/15/23, 5/17/24
Effective Date: 1/1/19, 6/15/24

8. Does the member have documented clinical manifestations of Fabry disease? (Provide documentation of disease manifestations e.g. cardiac, renal, neurologic)
    a. If yes, continue to #9
    b. If no, clinical review required
9. Will Galafold (migalastat hydrochloride) be used in combination with other enzyme replacement therapy for treatment of Fabry disease?
    a. If yes, clinical review required
    b. If no, continue to #10
10. Is the treatment being prescribed by, or in consultation with, a clinical geneticist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a positive clinical response to therapy as defined as reduction in levels of plasma or urinary globotriaosylceramide (Gb3/GL-3), globotriaosylsphingosine (lyso-Gb3), or kidney interstitial capillary cell globotriaosylceramide (KIC GL-3) and/or an improvement or stabilization of clinical signs and symptoms (e.g., dermatologic, gastrointestinal, pulmonary, vascular, renal, cardiac, neurologic manifestations)? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the patient have documentation of estimated glomerular filtration rate (eGFR) of 30 mL/min/1.73m2 or greater? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by, or in consultation with, a clinical geneticist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Galafold [Product Information], Amicus Therapeutics U.S., Inc. Cranbury, NJ. December 2021.

Last Reviewed: 12/19/18, 11/18/20, 9/15/21, 11/16/22, 9/15/23, 5/17/24
Effective Date: 1/1/19, 6/15/24

2. Germain DP, Hughes DA, Nicholls K, et al.: Treatment of Fabry's Disease with the Pharmacologic Chaperone Migalastat. NEJM 2016; 375:545-555.
3. Desnick RJ, Brady R, Barranger J, et al. Fabry disease, an Under-Recognized Multisystemic Disorder: Expert Recommendations for Diagnosis, Management, and Enzyme Replacement Therapy. Ann Intern Med. 2003; 138(4):338-46.
4. Ortiz, Alberto, et al. "Fabry disease revisited: management and treatment recommendations for adult patients." Molecular genetics and metabolism 123.4 (2018): 416-427.
5. Müntze J, Gensler D, Maniuc O, et al. Oral Chaperone Therapy Migalastat for Treating Fabry Disease: Enzymatic Response and Serum Biomarker Changes After 1 Year. Clin Pharmacol Ther. 2019;105(5):1224-1233.

Last Reviewed: 12/19/18, 11/18/20, 9/15/21, 11/16/22, 9/15/23, 5/17/24
Effective Date: 1/1/19, 6/15/24

Gattex® (teduglutide)
Prior Authorization Guidelines

Affected Medication(s)

- Gattex kit for subcutaneous administration

FDA Approved Indication(s)

- Treatment of adult and pediatric patients 1 year of age or older with Short Bowel Syndrome (SBS) who are dependent on parenteral support

Dosing

- 0.05 mg/kg body weight administered by subcutaneous injection once daily

Initial Authorization Criteria

1. Is the request for continuation of Gattex (teduglutide) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member at least 1 year of age?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has the member been dependent on parenteral nutrition support at least 3 times a week for at least 12 consecutive months prior to planned date of Gattex (teduglutide) initiation? (Provide documentation of parenteral nutritional support history and frequency)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, a gastroenterologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required

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Effective Date: 1/1/19, 12/15/20, 4/15/23

2. Has the member's requirement for parenteral nutritional support decreased at least 1 day per week from pre-treatment baseline? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a gastroenterologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Gattex [package insert]. Lexington, MA: Shire-NPS Pharmaceuticals, Inc.; November 2022.
2. Jeppesen P, Pertkiewicz M, Messing B, et al. Teduglutide reduces need for parenteral support among patients with short bowel syndrome with intestinal failure. Gastroenterology. 2012;143:1473-1481.
3. Parrish, Carol Rees, and John K. DiBaise. "Managing the adult patient with short bowel syndrome." Gastroenterology & hepatology 13.10 (2017): 600.

Last Reviewed: 11/21/18, 11/18/20, 11/17/21, 3/17/23, 3/15/23
Effective Date: 1/1/19, 12/15/20, 4/15/23

Glucosylceramide Synthase Inhibitors
Prior Authorization Guidelines

Affected Medication(s)

- Cerdelga (eliglustat) oral capsule
- miglustat oral capsule
- Opfolda (miglustat) oral capsule
- Yargesa (miglustat) oral capsule
- Zavesca (miglustat) oral capsule

FDA Approved Indication(s)

- Cerdelga: Long-term treatment of adult patients with Gaucher disease type 1 (GD1) who are CYP2D6 extensive metabolizers (EMs), intermediate metabolizers (IMs), or poor metabolizers (PMs) as detected by an FDA-cleared test
- Opfolda: Treatment, in combination with cipaglucosidase alfa, of adult patients with late-onset Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency) weighing ≥40 kg and who are not improving on their current enzyme replacement therapy (ERT)
- Yargesa, Zavesca: Monotherapy for the treatment of adult patients with mild to moderate type 1 Gaucher disease for whom enzyme replacement therapy (examples include: imiglucerase, velaglucerase alfa, or taliglucerase alpha) is not a therapeutic option (e.g. due to allergy, hypersensitivity, or poor venous access)

Dosing

- Refer to corresponding package insert for specific dosing recommendations
  -

Initial Authorization Criteria

1. Is the request for continuation of the same glucosylceramide synthase inhibitor for the same indication?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Has the member had a previous inadequate response, intolerance, or contraindication (i.e. due to allergy, hypersensitivity, or poor venous access) to enzyme replacement therapy (i.e. velaglucerase alfa, imiglucerase, or taliglucerase alpha for Gaucher disease or alglucosidase alfa for Pompe disease)? (Provide history of enzyme replacement therapy or contraindication to use)
   a. If yes, continue to #3
   b. If no, clinical review required
4. Is the treatment being initiated by a provider that specializes in the treatment of inherited metabolic disorders? (Examples include a medical geneticist or an endocrinologist)
   a. If yes, continue to #5

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Effective Date: 1/1/19, 3/1/22, 4/15/24

b. If no, clinical review required

5. What is the indication that the medication is being request for? (Provide genetic testing result for review)

    a. Type 1 Gaucher disease, continue to corresponding criteria
    b. Pompe disease (lysosomal acid alpha-glucosidase deficiency), approve for 3 months unless otherwise specified
    c. Other Indication, clinical review required

Type 1 Gaucher Disease

1. Is the request for Cerdelga (eliglustat)?

    a. If yes, continue to #2
    b. If no, approve for 3 months unless otherwise specified

2. For approval of Cerdelga (eliglustat): Is the member a CYP2D6 poor metabolizer, extensive metabolizer, or intermediate metabolizer as confirmed using an FDA-cleared test?

    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the treatment being prescribed by or in consultation with a provider who specializes in the treatment of inherited metabolic disorders? (Examples include a medical geneticist or an endocrinologist)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is there documentation that the member has a clinical response to therapy defined by an improvement in symptoms and quality of life?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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GnRH Agonists
Prior Authorization Guidelines

Affected Medication(s)

- Eligard (leuprolide acetate) subcutaneous powder for suspension
- Fensolvi (leuprolide acetate) subcutaneous powder for suspension
- Leuprolide acetate solution for subcutaneous injection
- Lupron (leuprolide acetate) subcutaneous powder for suspension
- Lupron Depot (leuprolide acetate) for intramuscular injection
- Lupron Depot- Ped (leuprolide acetate) for intramuscular injection
- Synarel (nafarelin acetate) nasal spray
- Trelstar (triptorelin pamoate) intramuscular powder for suspension
- Triptodur (triptorelin) intramuscular powder for suspension

FDA Approved Indication(s)

- **Eligard**: For the palliative treatment of advanced prostate cancer
- **Fensolvi**: For the treatment of pediatric patients 2 years of age and older with central precocious puberty (CPP)
- **Leuprolide acetate**: For the palliative treatment of advanced prostate cancer
- **Lupron**: For the palliative treatment of advanced prostatic cancer
- **Lupron Depot**: (endometriosis, duration is 6 months, preop, duration is 1 dose)
    - For management of endometriosis, including pain relief and reduction of endometriotic lesions
    - For initial management of the painful symptoms of endometriosis and for management of recurrence of symptoms when use in combination with norethindrone acetate
    - For the preoperative hematologic improvement of patients with anemia caused by uterine leiomyomata when used concomitantly with iron therapy
- **Lupron Depot-Ped**: For the treatment of children with central precocious puberty (CPP)
- **Synarel**:
    - For the treatment of central precocious puberty (gonadotropin-dependent precocious puberty) in children of both sexes
    - For management of endometriosis, including pain relief and reduction of endometriotic lesions in adult patient
- **Trelstar**: For the palliative treatment of advanced prostate cancer
- **Triptodur**: For the treatment of pediatric patients 2 years of age and older with central precocious puberty

Dosing

- Refer to corresponding package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of the same GnRH agonist therapy?

    a. If yes, continue to Reauthorization
    b. If no, continue to #2

2. Is the request for use to treat an FDA approved indication or major compendia supported indication? (Provide documentation of diagnosis)

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a. If yes, continue to #3
b. If no, clinical review required

3. What indication is the medication being requested for?

    a. Anemia associated with uterine leiomyomata (fibroids), continue to corresponding criteria
    b. Endometriosis, continue to corresponding criteria
    c. Central precocious puberty, continue to corresponding criteria
    d. Gender affirming treatment, continue to corresponding criteria
    e. Oncology indication, continue to corresponding criteria
    f. Other indication, continue to corresponding criteria

Anemia Associated with Uterine Leiomyomata (fibroids)

1. Is the member's hematocrit (Hct) ≤30% and/or hemoglobin (Hb) ≤ 10.2g/dL?

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the member currently taking adequate iron supplementation with insufficient response? (Provide documentation trial with inadequate response)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Will the member continue to take iron supplementation throughout therapy? (Provide documentation of complete treatment regimen)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is the member scheduled to have surgical removal of fibroids within the next 6 months? (Provide documentation of surgical date for review)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Is the medication being prescribed by, or in consultation with, a gynecologist?

    a. If yes, approve for up to 6 months unless otherwise specified
    b. If no, clinical review required

Endometriosis

1. Does the member have a previous trial with inadequate response, intolerance or contraindication to continuous contraception? (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the medication being prescribed by, or in consultation with, a gynecologist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Central Precocious Puberty

1. Is the member less than 8 years old if female or less than 9 years old if male?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have confirmation of diagnosis by measurement of serum luteinizing hormone (LH), follicle-stimulating hormone (FSH), and estradiol and/or testosterone? (Provide documentation of LH, FSH, and estradiol and/or testosterone levels)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the request for Triptodur, or Synarel?
    a. If yes, continue to #4
    b. If no, continue to #5
4. Does the member have a previous trial with inadequate response, intolerance, or contraindication to treatment with Lupron?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the medication prescribed by, or in consultation with, a pediatric endocrinologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

<u>Gender Affirming Treatment</u>

1. Does the member have a diagnosis of gender dysphoria by a qualified mental health professional? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

<u>Oncology Indication</u>

1. Is the medication being requested for an indication supported by the National Comprehensive Cancer Network (NCCN) with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide performance status for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Other Indications

1. Is the request supported by current medical guidelines?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member tried and had an inadequate response or have a contraindication to all preferred treatment options for the treatment of the requested condition?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with an appropriate specialist experienced in treating the requested condition?
    a. If yes, approve for standard treatment duration OR up to 12 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the request for reauthorization of Synarel?
    a. If yes, clinical review required
    b. If no, continue to #3
3. What indication is the medication being requested for?
    a. Anemia associated with uterine leiomyomata (fibroids), clinical review required
    b. Endometriosis, clinical review required
    c. Central precocious puberty, continue to corresponding criteria
    d. Gender affirming treatment, approve x 1 year
    e. Oncology indication, continue to corresponding criteria
    f. Other indication, continue to corresponding criteria

Central Precocious Puberty

1. Does the member show a positive clinical response to therapy? (Examples include: adequate hormone suppression, cessation of menses in girls, normalization and stabilization of linear growth and bone age advancement, and stabilization in the clinical signs/symptoms of puberty) (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the medication being prescribed by, or in consultation with, a pediatric endocrinologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Oncology Indication

1. Does the member show a positive clinical response to therapy defined by documentation of disease responsiveness? (Examples include reduction in PSA to normal values, serum testosterone level ≤50 ng/dL, etc.) (Provide supporting documentation of disease response)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the medication being prescribed by, or in consultation with, an oncologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Other Indications

1. Were updated chart notes (dated within 1 year) provided with documentation of significant clinical response to therapy? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the medication being prescribed by or in consultation with an appropriate specialist experienced in treating the requested condition?
    a. If yes, approve for standard treatment duration OR up to 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Eligard [Product Information], Tolmar Pharmaceuticals, Inc. Fort Collins, CO. April 2019
2. Fensolvi [Product Information], Tolmar Pharmaceuticals, Inc. Fort Collins, CO. April 2022
3. Lupron [Product Information], TAP Pharmaceutical Products Inc. Lake Forest, IL. February 2008
4. Lupron Depot [Product Information], AbbVie Inc. North Chicago, IL. February 2021
5. Lupron Depot- Ped [Product Information], AbbVie Inc. North Chicago, IL. November 2018
6. Synarel [Product Information], G.D. Searle LLC Division of Pfizer Inc. New York, NY. April 2022
7. Trelstar [Product Information], Verity Pharmaceuticals, Inc. Wayne, PA. October 2020
8. Triptodur [Product Information], Arbor Pharmaceuticals, LLC. Atlanta, GA. April 2022
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Growth Hormone
Prior Authorization Guidelines

Affected Medication(s)

- Genotropin
- Humatrope
- Norditropin
- Nutropin
- Omnitrope
- Saizen
- Serostim
- Skytrofa
- Zomacton

Indication(s)

- For the treatment of children with growth failure due to growth hormone deficiency (GHD), Prader-Willi syndrome (PWS), Small for Gestational Age (SGA), Turner syndrome (TS), and Idiopathic Short Stature (ISS)
  - Genotropin, Omnitrope
- For the treatment of children with short stature or growth failure associated with GHD, TS, ISS, short stature homeobox-containing gene (SHOX) deficiency, and failure to catch up in height after SGA
  - Humatrope
- For the treatment of pediatric members with growth failure due to inadequate secretion of endogenous growth hormone (GH), short stature associated with Noonan syndrome (NS), short stature associated with TS, SGA with no catch-up growth by age 2 to 4 years, ISS, and growth failure due to PWS
  - Norditropin
- For the treatment of children with growth failure due to GHD, ISS, TS, and chronic kidney disease (CKD) up to the time of renal transplantation
  - Nutropin AQ
- For the treatment of pediatric members with growth failure due to inadequate secretion of endogenous GH, short stature associated with TS, ISS, short stature or growth failure in SHOX deficiency, and short stature born SGA with no catch-up growth by 2 years to 4 years
  - Zomacton
- For the treatment of children with growth failure due to GHD
  - Saizen, Skytrofa
- For the treatment of adults with either adult onset or childhood onset GHD
  - Genotropin, Humatrope, Nutropin AQ, Omnitrope, Saizen
- For replacement of endogenous GH is adults with GH deficiency
  - Norditropin, Zomacton
- For the treatment of patients with HIV associated wasting or cachexia to increase lean body mass and body weight, and improve physical endurance. Concomitant antiretroviral therapy is necessary
  - Serostim

Dosing

- Refer to corresponding package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for renewal of a previously approved prior authorization for the same medication with the same indication?
   a. If yes, continue to Reauthorization

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b. If no, continue to #2

2. Is the treatment being prescribed by, or in consultation with, an appropriate specialist (e.g. endocrinologist, HIV specialist, gastroenterologist, etc.)?

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the requested medication either Nutropin or Omnitrope?

    a. If yes, continue to #5
    b. If no, continue to #4

4. Does the member have a trial with insufficient response, an intolerance, or contraindication to both Nutropin and Omnitrope OR use is inappropriate for FDA approved indication? (Provide supporting documentation)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Is the request for one of the below indications AND matches the medication's FDA approved indication for use? (Provide documentation of diagnosis, patient weight, and requested dosing/dosing frequency)

    - Growth failure due to growth hormone deficiency (GHD) in pediatrics
    - Growth failure due to Prader-Willi Syndrome (PWS) in pediatrics
    - Short stature born small for gestational age (SGA) with no catch-up growth by age 2 to 4 years in pediatrics
    - Short stature associated with Turner's Syndrome (TS) in pediatrics
    - Idiopathic short stature (ISS)
    - Short stature homeobox-containing gene (SHOX) deficiency in pediatrics
    - Short stature associated with Noonan Syndrome (NS) in pediatrics
    - Chronic renal insufficiency in pediatrics
    - Adult acquired GHD
    - GHD in adults who had childhood onset GHD
    - Acquired immunodeficiency syndrome (AIDS) wasting or cachexia
    - Short bowel syndrome

    a. If yes, continue to corresponding criteria
    b. If no, clinical review required

<u>Growth hormone deficiency (GHD) in pediatrics</u>

1. Does the member have auxologic evidence of short stature or growth failure defined by one of the following? (Provide supporting documentation)

    - "Severe" short stature (height < −2.5 SD below mean for age)
    - Height more than 2 SD below mid-parental height (average of mother's/father's heights)
    - Height < −2 SD below mean AND a 1-year height velocity < −1 SD below the mean for chronologic age or (in children 2 years of age or older) a 1-year decrease of > 0.5 SD in height

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member have a diagnosis of GHD confirmed by any of the following? (Provide supporting documentation)

- Insulin-like growth factor 1 (IGF-1) and insulin-like growth factor binding protein-3 (IGFBP-3) are <-2 SD with delayed bone age
- Positive for PROP1 or POU1F1 mutation
- When newborn, history of hypoglycemia, serum GH concentration <5 mcg/L, and deficiency ≥ 1 other pituitary hormone or classical imaging triad (ectopic posterior pituitary and pituitary hypoplasia with abnormal stalk
- Known pituitary abnormality (e.g. congenital anomaly, tumor, irradiation) and deficiency ≥ 1 other pituitary hormone

a. If yes, continue to #5
b. If no, continue to #3

3. Has the member completed GH stimulation testing? (Provide supporting documentation)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Upon provocative testing, was GH < 10 mcg/L for two different stimuli? (Provide supporting documentation)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Have other causes of short stature or growth failure been ruled out? (i.e. hypothyroidism, chronic systemic disease, and skeletal disorders)

    a. If yes, continue to #6
    b. If no, clinical review required

6. Is there documentation of open epiphyses? (Provide supporting documentation)

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Prader-Willi Syndrome (PWS) in pediatrics

1. Does the member have a diagnosis of PWS confirmed by genetic testing? (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member have uncontrolled diabetes, severe obesity, severe sleep apnea, or respiratory compromise? (Provide supporting documentation)

    a. If yes, clinical review required
    b. If no, continue to #3

3. Does the member have evidence of short stature or growth failure as defined as any of the following? (Provide supporting documentation)

    - Height < −2 SD below mid-parental height (average of mother's/father's heights)
    - Height < −2 SD below mean of same gender and chronological age
    - Height velocity < −2 SD below mean over 1 year OR < −1.5 SD below mean over 2 years

    a. If yes, clinical review required
    a. If no, continue to #4

4. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Small for gestational age (SGA) in pediatrics

1. Was the member born SGA as defined as weight and/or length <-2 SD below age mean? (Provide documentation of birth weight and length)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 2 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Did the member fail to achieve postnatal catch-up growth with height remaining <-2 SD below age mean? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Turner's Syndrome (TS) in pediatrics

1. Does the member have a diagnosis of TS confirmed by karyotype analysis? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member's height below the 5th percentile of the normal female growth curve? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Idiopathic short stature (ISS) in pediatrics

1. Does the member have ISS as defined as height $\leq$-2.25 SDs of mean for age in the absence of any endocrinal, metabolic, or other cause that explains short stature? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member's predicted adult height below the normal range (less than 63 inches for males or less than 59 inches for females)? (Provide documentation of predicted adult height)

a. If yes, continue to #3
b. If no, clinical review required

3. Has the provider thoroughly informed the member/member's family of the risks versus benefits and limitations of growth hormone therapy? (Provide supporting documentation)

a. If yes, continue to #4
b. If no, clinical review required

4. Is there documentation of open epiphyses? (Provide supporting documentation)

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

<u>SHOX deficiency in pediatrics</u>

1. Does the member have SHOX deficiency as diagnosed by DNA analysis? (Provide supporting documentation)

a. If yes, continue to #2
b. If no, clinical review required

2. Does the member have evidence of short stature or growth failure as defined by any of the following? (Provide supporting documentation)

- Height < −2 SD below mid-parental height (average of mother's/father's heights)
- Height < −2 SD below mean of same gender and chronological age
- Height velocity < −2 SD below mean over 1 year OR < −1.5 SD below mean over 2 years

a. If yes, continue to #3
b. If no, clinical review required

3. Is there documentation of open epiphyses? (Provide supporting documentation)

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

<u>Noonan Syndrome in pediatrics</u>

1. Does the member have Noonan syndrome confirmed by genetic testing? (Provide supporting documentation)

a. If yes, continue to #2
b. If no, clinical review required

2. Does the member have evidence of short stature or growth failure as defined as any of the following? (Provide supporting documentation)

- Height < −2 SD below mid-parental height (average of mother's/father's heights)
- Height < −2 SD below mean of same gender and chronological age

a. Height velocity < −2 SD below mean over 1 year OR < −1.5 SD below mean over 2 yearsIf yes, continue to #3
b. If no, clinical review required

3. Has severe hypertrophic cardiomyopathy been ruled out? (Examples of probable HCM include an abnormal electrocardiogram, an echocardiography showing left ventricle hypertrophy or systolic anterior motion of the mitral valve, or abnormal exercise testing) (Provide supporting documentation)

a. If yes, continue to #4
b. If no, clinical review required

4. Has the member been screened for thyroid abnormalities? (Examples include autoimmune thyroiditis with presence of thyroid autoantibodies) (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Chronic renal insufficiency in pediatrics

1. Does the member have chronic renal insufficiency with an estimated GFR <75 mL/min per 1.73 m^2? (Provide documentation of GFR)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there evidence of growth impairment, defined as height Z-score <-1.88 (3^{rd} percentile) or a height velocity <-2 SDs for age? *[Note: Z-score calculated using CDC height chart]* (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Have other causes of short stature been ruled out and/or corrected prior to consideration of GH therapy? (i.e. acidosis, secondary hyperparathyroidism, malnutrition, zinc deficiency)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have severe hyperparathyroidism (CKD stage 2-4: PTH >400 pg/mL or CKD stage 5: PTH >900 pg/mL)?
    a. If yes, clinical review required
    b. If no, continue to #5
5. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the member pre-kidney transplantation? (Provide supporting documentation)
    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Adult onset GHD

1. Does the member have all of the following? (Provide supporting documentation)
    - Confirmed panhypopituitarism (deficiency in ≥ 3 pituitary hormones)
    - Serum IGF-1 levels < 2.5 percentile
    - Irreversible pituitary disease or physical trauma (e.g. pituitary tumor, pituitary surgical damage, irradiation, sarcoidosis)

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, continue to #2

2. Does the member have documentation of at least TWO of the following? (Provide supporting documentation)
    - Deficient in at least ≥ 1 other pituitary hormone and IGF-1 <50th percentile
    - Insulin tolerance test (ITT) with peak growth hormone (GH) ≤ 5.0 mcg/L
    - Glucagon stimulation test with serum GH < 3.0 mcg/L (or <1 mcg/L if obese) and arginine-L-DOPA stimulation test with serum GH < 1.5 mcg/L
    - Macimorelin stimulation test with serum GH <2.8 ng/mL

    a. If yes, continue to #3
    b. If no, clinical review required
3. Was member's GHD is caused by traumatic brain injury or subarachnoid hemorrhage?
    a. If yes, continue to #4
    b. If no, approve for 6 months unless otherwise specified
4. Was GH stimulation test performed at least 12 months after the event? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Adults with childhood onset GHD

1. Was the member treated with GH replacement for conditions other than for GHD (i.e. TS, ISS, PWS, etc)?
    a. If yes, clinical review required
    b. If no, continue to #2
2. Does the member have childhood onset GHD from a known genetic mutation, embryopathic/congenital defect, or irreversible hypothalamic-pituitary structural damage AND has panhypopituitarism (≥ 3 pituitary hormone deficiencies) with IGF-1 < 2.5 percentile? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, continue to #3
3. Has the member completed retesting with GH stimulation test? *[Note: Re-testing should be completed after final height achieved and GH stopped for at least 1 month]* (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Upon re-testing, does the member have confirmation of GHD through at least TWO of the following? (Provide supporting documentation)
    - Deficient in at least one other pituitary hormone and IGF-1 <50th percentile
    - Insulin tolerance test (ITT) with peak growth hormone (GH) ≤ 5.0 mcg/L
    - Glucagon stimulation test with serum GH < 3.0 mcg/L (or <1 mcg/L if BMI is ≥ 25 kg/m^2) and arginine-L-DOPA stimulation test with serum GH < 1.5 mcg/L
    - Macimorelin stimulation test with serum GH <2.8 ng/mL

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

HIV-associated wasting or cachexia

1. Does the member have HIV and one the following? (Provide supporting documentation)
    - Unintentional weight loss of ≥10% over 12 months
    - Weight < 90% of the lower limit of ideal body weight
    - BMI <20 kg/m^2

    a. If yes, continue to #2
    b. If no, clinical review required
2. Have other possible illnesses aside from HIV/AIDS been ruled out as the cause of weight loss?

    a. If yes, continue to #3
    b. If no, clinical review required
3. Will the member receive antiretroviral therapy for HIV/AIDS concomitantly with the requested medication? (Provide supporting documentation of treatment plan)

    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a documented trial with insufficient response, an intolerance, or contraindication to at least one appetite stimulants and/or anabolic agents? (Provide supporting documentation)

    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the member less than 18 years of age?

    a. If yes, clinical review required
    b. If no, approve for 3 months unless otherwise specified

<u>Short bowel syndrome (SBS)</u>

1. Does the member have nutritional malabsorption due to loss of function or portion of the intestines? (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member currently receiving specialized nutrition support (i.e. high carbohydrate, low-fat diet, enteral feedings, parenteral nutrition)? (Provide supporting documentation of nutrition support)

    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member tried and failed traditional therapies for the management of SBS (e.g. acid suppressing agents, antidiarrheals, or octreotide)? (Provide supporting documentation)

    a. If yes, approve for 1 month unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for one of the below indications AND matches the medication's FDA-labeled indications for use? (Provide documentation of diagnosis, patient weight, and requested dosing/dosing frequency)

    a. GHD, PWS, SGA, ISS, TS, SHOX, NS in pediatrics, continue to the corresponding criteria
    b. CKD in pediatrics, continue to corresponding criteria

c. Adults with GHD, continue to corresponding criteria
d. Acquired immunodeficiency syndrome (AIDS) wasting or cachexia, continue to corresponding criteria
e. Short bowel syndrome, clinical review required

GHD, PWS, SGA, ISS, TS, SHOX, NS in pediatrics

1. Is there documentation of member responding to therapy (i.e. growth velocity ≥ 2 cm/year)? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member still have potential to grow (i.e. has not reached expected final adult height)? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member's IGF-I level maintained between 0 to +2 SD for age? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

CKD in pediatrics

1. Is the member pre-renal transplantation? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there documentation of member responding to therapy (i.e. growth velocity ≥ 2 cm/year)? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member still below target height based on midparental height or 50th percentile for age? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have severe hyperparathyroidism (CKD stage 2-4: PTH >400 pg/mL or CKD stage 5: PTH >900 pg/mL)?
    a. If yes, clinical review required

b. If no, approve for 12 months unless otherwise specified

Adults with GHD (childhood or adult onset)

1. Is the member's IGF-1 concentration within the age-specific range of normal? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there documentation the member is benefiting from GH therapy (e.g. increase quality of life, improvements in body composition, cardiovascular risk markers, etc)? (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

HIV-associated wasting or cachexia

1. Is there documentation of positive response from therapy (i.e. increase in body weight or BCM)? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member continue to have evidence of wasting? (Provide supporting documentation)
    a. If yes, approve for 12 weeks unless otherwise specified (maximum total duration 48 weeks)
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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Hepatitis C Direct Acting Antivirals
Prior Authorization Guidelines

Affected Medication(s)

- Epclusa (sofosbuvir/velpatasvir) oral tablet / pellets
- Sofosbuvir/velpatasvir oral tablet
- Mavyret (glecaprevir/pibrentasvir) oral tablet / pellets
- Vosevi (sofosbuvir/velpatasvir/voxilaprevir) oral tablet

FDA Approved Indication(s)

- **Epclusa**: Treatment of chronic hepatitis C (HCV) genotype 1, 2, 3, 4, 5, or 6 infection in adults and pediatric patients ≥3 years of age without cirrhosis or with compensated cirrhosis or in combination with ribavirin in patients with decompensated cirrhosis
- **Mavyret**: Treatment of chronic HCV genotype 1, 2, 3, 4, 5, or 6 infection in adults and pediatric patients ≥3 years of age without cirrhosis or with compensated cirrhosis (Child-Pugh A); HCV genotype 1 infection in adults and pediatric patients ≥3 years of age previously treated with a regimen containing an HCV NS5A inhibitor or an NS3/4A protease inhibitor, but not both
- **Vosevi**: Treatment of adults with chronic HCV infection without cirrhosis or with compensated cirrhosis (Child-Pugh A) who have genotype 1, 2, 3, 4, 5, or 6 infection and have previously been treated with an HCV regimen containing an NS5A inhibitor or who have genotype 1a or 3 infection and have previously been treated with an HCV regimen containing sofosbuvir without an NS5A inhibitor

Dosing

- Refer to indication specific compendia supported dosing

Initial Authorization Criteria

1. Is the request for use to treat chronic hepatitis C? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the request for continuation of a hepatitis C regimen?
    a. If yes, clinical review required
    b. If no, continue to #3

3. Have all of the following pre-treatment test results been submitted for regimen appropriateness review? (Provide documentation of all requested test results)
    - HCV genotype for members with cirrhosis
    - Fibrosis staging if appropriate
    - Quantifiable baseline HCV RNA (within the last 12 months or rationale indicating why HCV RNA labs were completed >12 months prior)
    - HBV serology
    - Treatment history and outcome

    a. If yes, continue to #4
    b. If no, clinical review required

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4. If positive HBsAg or positive anti-HBc with negative HBsAb: Is there a documented monitoring plan? (Provide documentation of monitoring plan)

    a. If yes, continue to #5
    b. If no, clinical review required
    c. N/A, continue to #5

5. Is the requested treatment being prescribed by a provider who meets one of the following criteria:

    - No specialist required: Fibrosis score F0-F2, treatment naive with no prior Hepatitis B exposure
    - ECHO participant: Fibrosis score F3-F4 or prior Hepatitis B exposure
    - Specialist (i.e. gastroenterologist, hepatologist, ID): Decompensated cirrhosis or multiple DAA-failures

    a. If yes, continue to #6
    b. If no, clinical review required

6. Is this medication appropriate considering comorbid conditions or contraindications for use (e.g. pregnancy, malignancy outside of the liver not meeting oncologic criteria for cure, etc.)? (Provide documentation of comorbid conditions)

    a. If yes, continue to #7
    b. If no, clinical review required

7. Does the requested regimen match one of the approved regimens below or is it supported by current AASLD-IDSA guidelines with an evidence rating of Class I, Level B and higher? (Provide documentation of treatment regimen)

    a. If yes, approve for appropriate duration
    b. If no, clinical review required

REGIMENS

Table 1: Recommended Treatment Regimens for Adults, and Adolescents 12 years of age and older with Hepatitis C virus

Treatment History	Cirrhosis Status	Recommended Regimen
Treatment Naïve (Genotype 1-6)		
Treatment naïve or confirmed reinfection	Non-Cirrhotic or Compensated Cirrhosis	• G/P x 8 weeks • SOF/VEL x 12 weeks (Baseline resistance testing recommended for GT3 patien with compensated cirrhosis)

	Decompensated Cirrhosis	• SOF/VEL + RBV x 12 weeks • SOF/VEL x 24 weeks if ribavirin ineligible
Treatment Experienced		
Sofosbuvir based regimen treatment failures, including: Sofosbuvir + ribavirin Ledipasvir/sofosbuvir Velpatasvir/sofosbuvir	Non-cirrhotic or compensated cirrhosis	• SOF/VEL/VOX x12 weeks • G/P x 16 weeks (except GT3)
	Decompensated cirrhosis	• SOF/VEL + RBV x 24 weeks
Elbasvir/grazoprevir treatment failures	Non-cirrhotic or compensated cirrhosis	• SOF/VEL/VOX x 12 weeks
Glecaprevir/pibrentasvir treatment failures	Non-cirrhotic or compensated cirrhosis	• G/P + SOF + RBV x 16 weeks • SOF/VEL/VOX x 12 weeks (plus RBV if compensated cirrhosis)
Multiple DAA Treatment Failures, including: sofosbuvir/velpatasvir/voxilaprevir glecaprevir/pibrentasvir + sofosbuvir	Non-cirrhotic or compensated cirrhosis	• G/P + SOF + RBV x 16-24 weeks • SOF/VEL/VOX + RBV x 24 weeks

Abbreviations: DAA = direct acting antiviral; G/P = glecaprevir and pibrentasvir; PEG= pegylated interferon; RBV = ribavirin; SOF = sofosbuvir; SOF/VEL = sofosbuvir/velpatasvir; SOF/VEL/VOX = sofosbuvir/velpatasvir/voxilaprevir

Ribavirin ineligible/intolerance may include: 1) neutrophils < 750 mm3, 2) hemoglobin < 10 g/dl, 3) platelets <50,000 cells/mm3, autoimmune hepatitis or other autoimmune condition, hypersensitivity or allergy to ribavirin

Rarely, genotyping assays may indicate the presence of a mixed infection (e.g., genotypes 1a and 2). Treatment data for mixed genotypes with direct-acting antivirals are limited. However, in these cases, a pangenotypic regimen is appropriate.

Ribavirin-containing regimens are absolutely contraindicated in pregnant women and in the male partners of women who are pregnant. Documented use of two forms of birth control in patients and sex partners for whom a ribavirin containing regimen is chosen is required.

All regimens containing a protease inhibitor (elbasvir, glecaprevir, simeprevir, paritaprevir, voxilaprevir) should not be used in patients with moderate to severe hepatic impairment (CTP B and C).

Table 2: Recommended Treatment Regimens for children ages 3 - 12 years of age with Hepatitis C virus

Treatment History	Cirrhosis Status	Recommended Regimen
Treatment Naïve Genotype 1-6		
Treatment naïve, confirmed reinfection or prior treatment with PEG/RBV	Non-cirrhotic or compensated cirrhosis	• SOF/VEL x 12 weeks • G/P x 8 weeks

	Decompensated Cirrhosis	• SOF/VEL + RBV x 12 weeks
Treatment Experienced with DAA regimen		
Note: Efficacy and safety data extremely limited in treatment experienced to other DAAs in this population. Can consider recommended treatment regimens in adults if FDA approved for pediatric use. Recommend consulting with hepatologist.		
Abbreviations: DAA = direct acting antiviral; G/P = glecaprevir and pibrentasvir; RBV = ribavirin; SOF/VEL = sofosbuvir/velpatasvir • All regimens containing a protease inhibitor (elbasvir, glecaprevir, simeprevir, paritaprevir, voxilaprevir) should not be used in patients with moderate to severe hepatic impairment (CTP B and C)		

Table 3: Recommended dosage of sofosbuvir/velpatasvir in pediatric patients 3 years of age and older:

Body Weight	Dosing of sofosbuvir/velpatasvir
Less than 17 kg	One 150 mg/37.5 mg pellet packet once daily
17 kg to less than 30 kg	One 200 mg/50 mg pellet packet OR tablet once daily
At least 30 kg	Two 200 mg/50 mg pellet packets once daily OR one 400 mg/100 mg tablet once daily

Table 4: Recommended dosage of glecaprevir/pibrentasvir in pediatric patients 3 years of age and older:

Body Weight	Dosing of sofosbuvir/velpatasvir
Less than 20 kg	Three 50mg/20 mg pellet packets once daily
20 kg to less than 30 kg	Four 50 mg/20 mg pellet packets once daily
30 kg to less than 45 kg	Five 50 mg/20 mg pellet packets once daily
45 kg and greater OR 12 years of age and older	Three 100mg/40 mg tablets once daily

Table 5: Unique Populations

Characteristics	Cirrhosis Status	Recommended Regimen
POST LIVER TRANSPLANT: GENOTYPE 1-6 in the allograft		
Treatment-naïve or -experienced	Non-cirrhotic or compensated cirrhosis	• SOF/VEL x 12 weeks • G/P x 12 weeks
	Decompensated cirrhosis	• SOF/VEL + RBV x 12 weeks (naïve) • SOF/VEL + RBV x 24 weeks (experienced)

Treatment experienced with direct acting antiviral (DAA) regimen	Non-cirrhotic or compensated cirrhosis	• SOF/VEL/VOX x 12 weeks
POST KIDNEY TRANSPLANT: GENOTYPE 1-6		
Treatment-naive or non-DAA experienced	Non-cirrhotic or compensated cirrhosis	• SOF/VEL x 12 week • G/P x 12 weeks
Treatment experienced with direct acting antiviral (DAA) regimen	Non-cirrhotic or compensated cirrhosis	• SOF/VEL/VOX +/- RBV x 12 weeks
HIV/HCV - COINFECTION		
HIV/HCV-coinfected persons should be treated and retreated the same as persons without HIV infection, after recognizing and managing interactions with antiretroviral medications		

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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Hereditary Angioedema Agents Policy
Prior Authorization Guidelines

Affected Medication(s)

- Berinert (human c1-esterase inhibitor) intravenous solution
- Cinryze (human c1-esterase inhibitor) intravenous solution
- Firazyr (icatibant acetate) subcutaneous solution
- Icatibant acetate subcutaneous solution
- Haegarda (human c1-esterase inhibitor) subcutaneous solution
- Kalbitor (ecallantide) subcutaneous solution
- Orladeyo (berotralstat) oral capsule
- Ruconest (c1-esterase inhibitor recombinant) intravenous solution
- Sajazir (icatibant acetate) subcutaneous solution
- Takhzyro (lanadelumab-flyo) subcutaneous solution

Indication(s)

- **Berinert**: Treatment of acute abdominal, facial, or laryngeal hereditary angioedema (HAE) attacks in adult and pediatric patients 6 years of age and older
- **Cinryze**: Routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years old and above) with Hereditary Angioedema (HAE)
- **Firazyr (icatibant, Sajazir)**: Treatment of acute attacks of hereditary angioedema (HAE) in adults 18 years of age and older
- **Haegarda**: Routine prophylaxis to prevent Hereditary Angioedema (HAE) attacks in adolescent and adult patients 6 years of age and older
- **Kalbitor(:** Treatment of acute attacks of hereditary angioedema (HAE) in patients 12 years and older
- **Orladeyo:** Prophylaxis to prevent attacks of hereditary angioedema (HAE) in adults and pediatric patients 12 years and older
- **Ruconest**: Treatment of acute attacks in adult and adolescent patients with hereditary angioedema (HAE) 13 years of age and older
- **Takhzyro**: Prophylaxis to prevent attacks of hereditary angioedema (HAE) in patients 2 years and older

Dosing

- **Berinert**: 20 IU/kg intravenously
- **Cinryze**:
  - Adults and adolescents 12 years and older: 1,000 U IV every 3 or 4 days
    - For inadequate response: Dose may be increased every 3 or 4 days up to 2,000 U (not to exceed 80 U/kg)
  - Pediatric patients 6 to 11 years old: 500 U IV every 3 or 4 days
    - Dose may be adjusted according to individual response up to 1,000 U every 3 or 4 days
- **Firazyr, Sajazir**: 30 mg subcutaneously for attack, up to 2 additional doses may be administered at intervals of at least 6 hours if attack persists or symptoms recur
- **Haegarda**: Self-administer 60 IU/kg body weight subcutaneously twice weekly
- **Kalbitor:** 30 mg subcutaneously for attack, may repeat an additional 30 mg within 24 hours if attack persists

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- **Orladeyo: 110-**150mg orally once daily
- **Ruconest**:
  - Less than 84 kg: 50 U/kg intravenously
  - Greater than or equal to 84 kg: 4200 U (2 vials) intravenously
  - No more than two doses should be administered within a 24 hour period
- **Takhyzyro:**
  - 300mg subcutaneously every two weeks. Consider dosing every 4 weeks when patient is attack free for greater than 6 months

Initial Authorization Criteria

1. Is the request for continuation of the therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the medication being requested for an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member avoiding possible triggers for HAE attacks? Possible triggers include:
   - Helicobacter pylori infections (confirmed by lab test)
   - Systemic estrogen products
   - Antihypertensive agents containing ACE inhibitors
   - Dipeptidyl peptidase IV (DPP-IV) inhibitors (e.g., sitagliptin)
   - Neprilysin inhibitors (e.g., sacubitril)

   a. If yes, continue to #4
   b. If no, clinical review required
4. Does the member have at least 2 HAE attacks per month at baseline? (Provide supporting documentation for review)
   a. If yes, continue to #6
   b. If no, continue to #5
5. Does the member have a history of moderate to severe cutaneous or abdominal attacks OR mild to severe airway swelling attacks of HAE? (i.e. debilitating cutaneous/gastrointestinal symptoms OR laryngeal/pharyngeal/tongue swelling) (Provide supporting documentation for review)
   a. If yes, continue to #6
   b. If no, clinical review required
6. Does the member have one of the following clinical presentations consistent with HAE subtype? (Provide supporting documentation)
   - For HAE I (C1-inhibitor deficiency):
     - Low C1 inhibitor (C1-INH) antigenic level (C1-INH antigenic level below the lower limit of normal as defined by the laboratory performing the test); AND

- Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); AND
- Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test); AND
  - Patient has a family history of HAE OR
  - Normal C1q level

- For HAE II (C1-inhibitor dysfunction):
  - Normal to elevated C1-INH antigenic level; AND
  - Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); AND
  - Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test)

a. If yes, continue to #7
b. If no, clinical review required

7. Is the request for either Takhzyro, Cinryze, or Orladeyo?

   a. If yes, continue to #8
   b. If no, continue to #9

8. Has the member had a previous trial with inadequate response, intolerance, or contraindication to therapy with Haegarda? (Provide supporting documentation) (NOTE: Takhzyro can be used if member is <6 years old)

   a. If yes, continue to #11
   b. If no, clinical review required

9. Is the request for Berinert, Sajazir, Firazyr, Kalbitor, or Ruconest?

   a. If yes, continue to #10
   b. If no, continue to #11

10. Has the member had a previous trial with inadequate response, intolerance, or contraindication to therapy with generic icatibant? (Provide supporting documentation)

    a. If yes, continue to #11
    b. If no, clinical review required

11. Is the medication being prescribed by or in consultation with a specialist in allergy, immunology, hematology, pulmonology, or medical genetics?

    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Does the member continue to meet criteria above?

   a. If yes, continue to #2
   b. If no, clinical review required

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2. Has the member demonstrated significant improvement in severity and duration of attacks that has been achieved and sustained? (Provide supporting documentation of improvement in severity or duration of attacks)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there documentation that there has been an absence of unacceptable toxicity from the drug. (Examples of unacceptable toxicity include the following: hypersensitivity reactions, serious thrombotic events, laryngeal attacks, etc.) (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the medication being prescribed by or in consultation with a specialist in allergy, immunology, hematology, pulmonology, or medical genetics?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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18. Maurer M, Mager M, Ansotegui I, et al. The international WAO/EAACI guideline for the management of hereditary angioedema-The 2017 revision and update. Allergy. 2018 Jan 10. doi: 10.1111/all.13384.

Hetlioz® (tasimelteon)
Prior Authorization Guidelines

Affected Medication(s)

- Hetlioz oral capsule
- Hetlioz LQ oral suspension
- Tasimelteon capsule

FDA Approved Indication(s)

- Hetlioz capsules:
  - For the treatment of Non-24-Hour Sleep-Wake Disorder (Non-24) in adults
  - For the treatment of nighttime sleep disturbances in SMS in patients 16 years of age and older.
- Hetlioz LQ:
  - For the treatment of nighttime sleep disturbances in SMS in pediatric patients 3 to 15 years of age

Dosing

- 20 mg by mouth taken before bedtime at the same time every night

Initial Authorization Criteria

1. Is the request for continuation of Hetlioz (tasimelteon) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have documentation of a minimum trial of 12 weeks of melatonin with insufficient response, intolerance, or contraindication to therapy? (Provide relevant current/past medication history)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is Hetlioz (tasimelteon) being requested for nighttime sleep disturbances related to Smith-Magenis syndrome (SMS)?
   a. If yes, continue to #6
   b. If no, continue to #5
5. Is the member completely blind with no light perception? (Provide supporting documentation)
   a. If yes, continue to #6
   b. If no, clinical review required
6. Is the treatment being prescribed by or in consult with a specialist specialized in sleep disorders?
   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

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Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member had a positive clinical response to therapy as defined as decreased symptoms? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consult with a specialist in sleep disorders?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Hetlioz Prescribing Information. Washington, D.C.: Vanda Pharmaceuticals Inc.; February 2021. Available at: www.hetlioz.com.
2. Auger RR, Burgess HJ, Emens JS, Deriy LV, Thomas SM, and Sharkey KM. Clinical practice guideline for the treatment of intrinsic circadian rhythm sleep-wake disorders: advanced sleep-wake phase disorder (ASWPD), delayed sleep-wake phase disorder (DSWPD), non-24-hour sleep-wake rhythm disorder (N24SWD), and irregular sleep-wake rhythm disorder (ISWRD) - an update for 2015. J Clin Sleep Med. 2015; 11(10): 1199-1236.
3. Abbott SM. Non-24-hour sleep-wake rhythm disorder. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed October 2018.

Last Reviewed: 10/17/18, 7/21/21, 7/20/22, 3/17/23, 3/15/24
Effective Date: 1/1/19, 9/1/21, 9/1/22, 6/1/23, 4/15/24

Impavido® (miltefosine)
Prior Authorization Guidelines

Affected Medication(s)

- Impavido oral capsule

FDA Approved Indication(s)

- In adults and adolescents 12 years of age or older and weighing greater or equal to 30kg for treatment of:
  - Visceral leishmaniasis caused by *Leishmania donovani*
  - Cutaneous leishmaniasis caused by *Leishmania braziliensis*, *Leishmania guyanensis*, and *Leishmania panamensis*
  - Mucosal leishmaniasis caused by *Leishmania braziliensis*

Dosing

- Patients weighing 30 to 44 kg: 50 mg twice daily with food for 28 days
- Patients weighing 45 kg or greater: 50 mg three times daily with food for 28 days

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, continue to #2
2. Does the member currently have a free-living ameba infection? (Examples include: primary amebic meningoencephalitis (PAM) due to *Naegleria fowleri*, granulomatous amebic encephalitis (GAE) due to *Balamuthia mandrillaris,* or keratitis due to *Acanthamoeba spp.*)? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member at least 12 years of age and weighs at least 30 kg? (Provide member weight for review)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is the requested dosing appropriate for the member's weight?
   a. If yes, continue to #5
   b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, an infectious disease specialist?
   a. If yes, approve for 1 month
   b. If no, clinical review required

Note:

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Effective Date: 1/1/19, 3/1/24

medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Impavido (prescribing information). Paladin Therapeutics Inc. Wilmington, DE. May 2021.
2. CDC Guidelines. Naegleria fowleri – Primary Amebic Meningoencephalitis (PAM) –Amebic Encephalitis. http://www.cdc.gov/parasites/naegleria/index.html. February 2017.
3. CDC Guidelines. Balamuthia mandrillaris - Granulomatous Amebic Encephalitis (GAE). http://www.cdc.gov/parasites/balamuthia/index.html. February 2016.
4. CDC Guidelines. Parasites – Acanthamoeba - Granulomatous Amebic Encephalitis (GAE); Keratitis. https://www.cdc.gov/parasites/acanthamoeba/index.html. Accessed December 2022.
5. Centers for Disease Control and Prevention: Leishmaniasis – Resources for Health Professionals. Available at: https://www.cdc.gov/parasites/leishmaniasis/health_professionals/index.html. Last updated: September 2021. Accessed on January 4, 2022.

Last Reviewed: 11/21/18, 1/20/21, 1/19/22, 1/20/23, 1/19/24
Effective Date: 1/1/19, 3/1/24

Increlex® (mecasermin)
Prior Authorization Guidelines

Affected Medication(s)

- Increlex subcutaneous solution

FDA Approved Indication(s)

Children ≥ 2 years old and adolescents:

- Treatment of growth failure with severe primary IGF-1 deficiency
- Treatment of growth hormone (GH) gene deletion who have developed neutralizing antibodies to GH

Dosing

- Recommended starting dose: 0.04 to 0.08 mg/kg twice daily
- Maximum dose: 0.12 mg/kg twice daily

Initial Authorization Criteria

1. Is the request for continuation of Increlex (mecasermin) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have documentation of open epiphyses demonstrated on bone radiograph? (Provide documentation of open epiphyses)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have any of the following secondary forms of IGF-1 deficiency?
    - Growth Hormone deficiency (GHD)
    - Malnutrition
    - Hypothyroidism
    - Chronic treatment with pharmacologic doses of steroidal anti-inflammatories

    a. If yes, clinical review required
    b. If no, continue to #5
5. Is the medication being prescribed by, or in consultation with, a pediatric endocrinologist?
    a. If yes, continue to #6
    b. If no, clinical review required
6. What diagnosis is Increlex being requested for?
    a. Severe IGF-1 deficiency, continue to corresponding criteria

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Effective Date: 1/1/19

b. Growth hormone (GH) gene deletion, continue to corresponding criteria

Severe IGF-1 Deficiency

1. Does the member have a height standard deviation score of less than or equal to -3.0? (Provide documentation of height standard deviation score for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a basal IGF-1 standard deviation score of less than or equal to -3.0? (Provide documentation of basal IGF-1 standard deviation score for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have normal or elevated growth hormone levels? (Provide documentation of growth hormone level for review)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Growth hormone (GH) Gene Deletion

1. Does the member have a basal IGF-1 level below normal range? (Provide documentation of basal IGF-1 level for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a presence of neutralizing antibodies to GH as confirmed by serum testing or genetic testing? (Provide supporting documentation of neutralizing antibodies)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a positive clinical response to therapy as defined by a height velocity of at least 2cm per year? (Provide documentation of height velocity for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member met their expected adult height goal? (Provide documentation of expected adult height and current height for review)
    a. If yes, clinical review required
    b. If no, approve for 12 months unless otherwise specified

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Increlex (mecasermin [rDNA origin] injection) [Product Information], Tercica, Inc. Brisbane, CA. August 2005.
2. American Association of Clinical Endocrinologists Medical Guidelines for Clinical Practice for Growth Hormone Use in Adults and Children – 2003 Update. Endocrine Practice 2003; 64-76.

Last Reviewed: 12/19/18, 7/21/21, 7/20/22, 7/21/23
Effective Date: 1/1/19

Injectable CGRP Antagonists
Prior Authorization Guidelines

Affected Medication(s)

- Aimovig (erenumab-aooe) subcutaneous solution
- Ajovy (fremanezumab-vfrm) subcutaneous solution
- Emgality (galcanezumab-gnlm) subcutaneous solution

FDA Approved Indication(s)

- Aimovig, Ajovy, Emgality: As the preventive therapy of migraine in adults
- Emgality: For treatment of episodic cluster headache in adults

Dosing

- Aimovig: 70 mg to140 mg subcutaneously once monthly
- Ajovy: 225 mg subcutaneously once monthly or 675 mg subcutaneously every 3 months
- Emgality:
  - Migraine: 240 mg subcutaneously once as loading dose, then 120 mg subcutaneously once monthly
  - Cluster headache: 300 mg at the onset of the cluster period and then monthly until the end of the cluster period

Initial Authorization Criteria

1. Has the requested medication previously been approved by OHSU?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member 18 years of age or older?
   a. If yes, continue to #4
   b. If no, clinical review required
4. What is the requested drug being used for?
   a. Migraine, continue to #5
   b. Cluster Headache, continue to #8
   c. Other indication, clinical review required
5. Has the member experienced 4 or more migraine headache days per month?
   a. If yes, continue to #6
   b. If no, clinical review required
6. Has the member had a two-month trial and failure or intolerance to (2) of the following alternative agents with differing mechanisms of action for migraine prophylaxis: topiramate, divalproex, metoprolol, propranolol, timolol, atenolol, nadolol, amitriptyline, nortriptyline, venlafaxine, duloxetine?
   a. If yes, continue to #11

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b. If no, continue to #7

7. Does the member have contraindications to all of the following alternative agents used for migraine prophylaxis: topiramate, divalproex, metoprolol, propranolol, timolol, atenolol, nadolol, amitriptyline, nortriptyline, venlafaxine, duloxetine?

    a. If yes, continue to #11
    b. If no, clinical review required

8. Does this member have at least two cluster periods lasting at least seven days to one year with at least five attacks?

    a. If yes, continue to #9
    b. If no, clinical review required

9. Are this member's cluster periods separated by at least three (3) months of pain-free remission?

    a. If yes, continue to #10
    b. If no, clinical review required

10. Has the member had a trial and failure or intolerance to a two-month trial of verapamil?

    a. If yes, continue to #11
    b. If no, clinical review required

11. Will CGRP antagonist to be used in combination with Botox?

    a. If yes, clinical review required
    b. If no, continue to #12

12. Will the injectable CGRP antagonist be used in combination with another CGRP antagonist? (including oral CGRP antagonists)

    a. If yes, clinical review required
    b. If no, approve for 6 months unless otherwise specified

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member have a positive clinical response to therapy as defined by a reduction in the frequency of migraine days per month or 50% or more reduction in weekly cluster headache attack frequency from pre-treatment baseline?

    a. If yes, continue to #3
    b. If no, clinical review required

3. Will the injectable CGRP antagonist be used in combination with another CGRP antagonist? (including oral CGRP antagonists)

    a. If yes, clinical review required
    b. If no, approve for 12 months unless otherwise specified

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Effective Date: 1/1/19, 9/1/19, 7/1/20, 11/15/20, 12/15/20, 1/1/22, 1/1/23, 12/20/23

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Aimovig [Prescribing Information]. Thousand Oaks, CA: Amgen Inc.; May 2018.
2. Ajovy [Prescribing Information]. North Wales, PA: Teva Pharmaceuticals USA, Inc.; July 2018.
3. Emgality [Prescribing Information]. Indianaplois, IN: Eli Lilly and Company: December 2019.
4. Silberstein SD, Holland S, Freitag F, et al. American Academy of Neurology: Evidence-based guideline update: Pharmacologic treatment for episodic migraine prevention in adults. Neurology 2012; 78: 1337-45.
5. Institute for clinical and economic review (ICER). Calcitonin Gene-Related Peptide (CGRP) inhibitors as preventive treatments for patients with episodic or chronic migraine: effectiveness and value. May 2018. Accessed October 11, 2018.
6. Robbins MS, Starling AJ, Pringsheim TM, et al. Treatment of Cluster Headache: The American Headache Society Evidence-Based Guidelines. Headache. 2016;56:1096-1106. Available at: https://headachejournal.onlinelibrary.wiley.com/doi/epdf/10.1111/head.12866.
7. The American Headache Society Position Statement on Integrating New Migraine Treatments Into Clinical Practice. Headache. 2019;59:1-18. Available at: https://headachejournal.onlinelibrary.wiley.com/doi/epdf/10.1111/head.13456.
8. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalagia. 2018;38(1);1-211. Available at: https://journals.sagepub.com/doi/pdf/10.1177/0333102417738202.

Last Reviewed: 12/19/18, 7/17/19, 5/20/20, 9/16/20, 11/18/20, 11/17/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 9/1/19, 7/1/20, 11/15/20, 12/15/20, 1/1/22, 1/1/23, 12/20/23

Inqovi® (decitabine and cedazuridine), Onureg® (azacitidine) Prior Authorization Guidelines

Affected Medication(s)

- Inqovi oral tablet
- Onureg oral tablet

FDA Approved Indication(s)

- Inqovi:
  - Treatment of adult patients with myelodysplastic syndromes (MDS), including previously treated and untreated, de novo and secondary MDS with the following French-American-British subtypes (refractory anemia, refractory anemia with ringed sideroblasts, refractory anemia with excess blasts, and chronic myelomonocytic leukemia [CMML]) and intermediate-1, intermediate-2, and high-risk International Prognostic Scoring System groups.
- Onureg:
  - Continued treatment of adult patients with acute myeloid leukemia (AML) who achieved first complete remission (CR) or complete remission with incomplete blood count recovery (CRi) following intensive induction chemotherapy and are not able to complete intensive curative therapy

Dosing

- Inqovi:
  - One tablet (35mg decitabine and 100mg cedazuridine) by mouth one time daily on Days 1 through 5 of each 28-day cycle
- Onureg:
  - 300mg orally once time daily on days 1 through 14 of each 28-day cycle

Initial Authorization Criteria

1. Is the request for continuation of a previously approved Inqovi or Onureg prior authorization with the same indication as the previous approval?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #4
   b. If no, continue to #3
3. Is the medication being requested for an indication supported by the National Comprehensive Cancer Network (NCCN) recommendation with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
   a. If yes, continue to #4
   b. If not, clinical review required
4. Is the treatment being prescribed by, or in consultation with, an oncologist?
   a. If yes, continue to #5
   b. If no, clinical review required

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Effective Date: 3/1/21

5. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is there medical rationale why the member cannot use generic IV formulation? (Provide supporting documentation)
    a. If yes, approve for 4 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication or supported by NCCN recommendation with an evidence level of 2A or higher? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by, or in consultation with, an oncologist?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there clinical documentation confirming disease responsiveness to therapy provided? (Example include reduction in tumor size, objective response, delay in progression, partial response, etc.) (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 05 Oct. 2020].
2. INQOVI® (decitabine and cedazuridine) tablets, [package insert]. Princeton, NJ: Taiho Oncology, Inc; 2020.
3. ONUREG (azacitidine) tablets, [package insert]. Summit, NJ: Celegene Corp.; 2020.
4. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Myelodysplastic Syndromes. Version 1.2021 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/default.aspx. Accessed December 8, 2020.
5. Garcia-Manero, Guillermo, et al. "Oral cedazuridine/decitabine: a phase 2, pharmacokinetic/pharmacodynamic, randomized, crossover study in MDS and CMML." Blood (2020).
6. Wei, Andrew H., et al. "The QUAZAR AML-001 Maintenance Trial: results of a phase III international, randomized, double-blind, placebo-controlled study of CC-486 (oral formulation of azacitidine) in patients with acute myeloid leukemia (AML) in first remission." (2019): LBA-3.

Last Reviewed: 1/20/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 3/1/21

7. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Acute Myeloid Leukemia. Version 2.2021 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/default.aspx. Accessed December 15, 2020.

Idiopathic Pulmonary Fibrosis (IPF) agents
Prior Authorization Guidelines

Affected Medication(s)

- Esbriet (pirfenidone) oral capsule/tablet
- Ofev (nintedanib) oral capsule
- pirfenidone oral capsule/tablet

FDA Approved Indication(s)

- Esbriet: For the treatment of idiopathic pulmonary fibrosis (IPF)
- Ofev:
  - For the treatment of adult with idiopathic pulmonary fibrosis (IPF)
  - For the treatment of adult with chronic fibrosing interstitial lung diseases (ILDs) with a progressive phenotype
  - To slow the rate of decline in pulmonary function in adult patients with systemic sclerosis-associated interstitial lung disease (SSc-ILD).

Dosing

- Esbriet: 801 mg three times daily (refer to package insert for titration recommendations)
- Ofev: 150 mg twice daily

Initial Authorization Criteria

1. Is the request for continuation of Esbriet (pirfenidone) or Ofev (nintedanib) therapy for the same indication?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with a pulmonologist?
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is the member 18 years of age or older?
   a. If yes, continue to #5
   b. If no, clinical review required
5. What is the requested indication?
   a. Idiopathic pulmonary fibrosis, continue to corresponding criteria
   b. Chronic fibrosing interstitial lung disease with progressive phenotype (Ofev only), continue to corresponding criteria
   c. To slow the rate of decline in pulmonary function in patients with systemic sclerosis-associated interstitial lung disease (SSC-ILD) (Ofev only), continue to corresponding criteria

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d. Other indication, clinical review required

Idiopathic pulmonary fibrosis

1. Is documentation confirming diagnosis of idiopathic pulmonary fibrosis (IPF) including presence of usual interstitial pneumonia (UIP), high resolution computed tomography (HRCT) result, or surgical lung biopsy result provided? (Provide supporting documentation for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Have known causes of interstitial lung disease been ruled out? (i.e. rheumatic disease, drug induced UIP, occupational exposures)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the requested medication Ofev (nintedanib) capsules?
    a. If yes, continue to #4
    b. If no, approve for 6 months
4. Has the member had a trial with insufficient response, intolerance, or contraindication to pirfenidone (Esbriet) tablets or capsules? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Chronic fibrosing interstitial lung disease with progressive phenotype (Ofev only)

1. Does the member have pulmonary fibrosis that is confirmed by high resolution computed tomography (HRCT)?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have any of the following clinical signs of progression?
    - FVC decline ≥10%
    - FVC decline ≥5% and <10% with worsening respiratory symptoms or imaging
    - Worsening respiratory symptoms and worsening imaging

    a. If yes, approve for 6 months
    b. If no, clinical review required

Systemic sclerosis-associated interstitial lung disease (Ofev only)

1. Does the member have pulmonary fibrosis that is confirmed by high resolution computed tomography (HRCT)?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have signs of systemic sclerosis? [Signs include: skin thickening of the fingers, fingertip lesions, telangiectasia, abnormal nailfold capillaries, pulmonary arterial hypertension, Raynaud's phenomenon, SSc-related antibodies (anticentromere, anti-topoisomerase I, anti-RNA polymerase III)]
    a. If yes, approve for 6 months

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b. If no, clinical review required

Reauthorization Criteria

1. Is the request to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member demonstrate a positive clinical response to therapy as defined by no disease progression, a slowed rate of progression, or improved respiratory symptoms? (Provide FVC test result and supportive documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with a pulmonologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Esbriet Prescribing Information. South San Francisco, CA: Genentech USA, Inc.; February 2022. Available at: https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=2e8c3537-36d7-4de5-9b5c-7a624b9a9e6e
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Last Reviewed: 10/17/18, 1/20/21, 3/16/22, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 3/1/21, 4/15/22, 7/15/22, 6/15/24

Isturisa® (osilodrostat)
Prior Authorization Guidelines

Affected Medication(s)

- Isturisa (osilodrostat) oral tablet

FDA Approved Indication(s)

- Treatment of adult patients with Cushing's Disease for whom pituitary surgery is not an option or has not been curative

Dosing

- Initiate dosage at 2 mg orally twice daily. Titrate dosage by 1 to 2 mg twice daily no more frequently than every 2 weeks based on rate of cortisol changes, individual tolerability and improvement in signs and symptoms. Maximum recommended dosage is 30 mg twice daily

Initial Authorization Criteria

1. Is the request for continuation of Isturisa (osilodrostat) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment prescribed by or in consultation with an endocrinologist?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has the member been diagnosed with endogenous Cushing's Disease and has either failed pituitary surgery or is not a candidate for surgery? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a 24-hour mean urinary free cortisol level greater than 1.5 times the upper limit of normal? (Above 67 µg/24 hours) (Provide supporting lab values)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a previous trial with inadequate response, intolerance, or contraindication to ketoconazole? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Last Reviewed: 11/18/20, 11/17/21, 3/17/23, 3/15/24
Effective Date: 12/15/20, 4/15/24

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the treatment prescribed by or in consultation with an endocrinologist?

    a. If yes, continue to #3
    b. If no, clinical review required

3. Has the member been diagnosed with endogenous Cushing's Disease and has either failed pituitary surgery or is not a candidate for surgery? (Provide supporting documentation)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Has the member experienced a documented positive response to therapy defined by a reduction in 24-hour urinary free cortisol levels to normal levels and/or improvement in signs or symptoms? (Provide supporting documentation) Note: For subsequent renewals documented maintenance of initial response is required.

    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

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3. Pivonello, Rosario, et al. "Efficacy and safety of osilodrostat in patients with Cushing's disease (LINC 3): a multicentre phase III study with a double-blind, randomised withdrawal phase." The Lancet Diabetes & Endocrinology 8.9 (2020): 748-761.
4. Lynnette K. Nieman, Beverly M. K. Biller, James W. Findling, M. Hassan Murad, John Newell-Price, Martin O. Savage, Antoine Tabarin, Treatment of Cushing's Syndrome: An Endocrine Society Clinical Practice Guideline, The Journal of Clinical Endocrinology & Metabolism, Volume 100, Issue 8, 1 August 2015, Pages 2807–2831, https://doi.org/10.1210/jc.2015-1818

Last Reviewed: 11/18/20, 11/17/21, 3/17/23, 3/15/24
Effective Date: 12/15/20, 4/15/24

Iwilfin (eflornithine)
Prior Authorization Guidelines

Affected Medication(s)

- Iwilfin (eflornithine) oral tablet

FDA Approved Indication(s)

- To reduce the risk of relapse in adult and pediatric patients with high-risk neuroblastoma (HRNB) who have demonstrated at least a partial response to prior multiagent, multimodality therapy including anti-GD2 immunotherapy

Dosing

- 0.75 to 1.5 m2: 576 mg by mouth twice daily
- Greater than 1.5 m2: 768 mg by mouth twice daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being requested for an indication supported by the National Comprehensive Cancer Network (NCCN) recommendation with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the medication being prescribed by, or in consultation with, an oncologist?
    a. If yes, approve for 4 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required

Last Reviewed: 3/15/24
Effective Date: 4/15/24

2. Is there clinical documentation confirming disease responsiveness to therapy provided? (Example include reduction in tumor size, objective response, delay in progression, partial response, etc.) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. IWILFIN™ (eflornithine) tablets [package insert]. Louisville, KY: USWM, LLC 2024.
2. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Neuroblastoma. Version 1.2024 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/pdf/neuroblastoma.pdf. [Accessed February 8, 2024.]
3. Oesterheld J, Ferguson W, Kraveka JM, et al. Eflornithine as Postimmunotherapy Maintenance in High-Risk Neuroblastoma: Externally Controlled, Propensity Score-Matched Survival Outcome Comparisons. J Clin Oncol. 2024;42(1):90-102.

Janus Kinase Inhibitor
Prior Authorization Guidelines

Affected Medication(s)

- Cibinqo™ (abrocitinib) oral tablet
- Olumiant® (baricitinib) oral tablet
- Rinvoq® (upadacitinib) oral tablet
- Xeljanz® (tofacitinib citrate) oral tablet/solution
- Xeljanz® XR oral tablet

FDA Approved Indication(s)

- Drug Compendia supported indications may be covered

Drug Name	RA	JIA	PsA	AS	UC	CD	AD	Other
Cibinqo							X	
Rinvoq	X		X	X	X	X	X	
Xeljanz	X	X	X	X	X			
Xeljanz Solution		X						
Xeljanz XR	X		X	X	X			
Olumiant	X							X

Dosing

- Refer to corresponding package insert for information

Initial Authorization Criteria

1. Is the request for continuation of therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication or a major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Will the requested medication be used concurrently with any other biologic therapy? (Examples: Enbrel, Actemra, Cimzia, Simponi, Orencia, Taltz, Cosentyx, Otezla, etc)
    a. If yes, clinical review required
    b. If no, continue to #4
4. What is the diagnosis that the medication is being requested for?
    a. Rheumatoid arthritis, continue to corresponding criteria
    b. Juvenile idiopathic arthritis, continue to corresponding criteria
    c. Psoriatic arthritis, continue to corresponding criteria
    d. Ankylosing spondylitis, continue to corresponding criteria
    e. Ulcerative colitis, continue to corresponding criteria

Last Reviewed: 11/16/22, 7/21/23, 3/15/24
Effective Date: 1/1/23, 9/15/23, 4/15/24

f. Crohn's disease, continue to corresponding criteria
g. Atopic Dermatitis, continue to corresponding criteria
h. Other indication, continue to corresponding criteria

Rheumatoid Arthritis (RA)

1. Is the diagnosis of rheumatoid arthritis (RA) confirmed by ACR/EULAR classification criteria AND has the diagnosis been documented for greater for 6 months? (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member have moderate to severe active RA confirmed by one of the tests below and despite the current RA management regimen? (Provide current RA management regimen and test result for review)

    - Routine Assessment of Patient Index Data 3 (RAPID3) of 2.0 or higher
    - Clinical Disease Activity Index (CDAI) of 10 or higher
    - Disease Activity Score (DAS) 28 erythrocyte sedimentation rate (ESR) of 3.2 or higher
    - Simplified Disease Activity Index (SDAI) of 11 or higher

    a. If yes, continue to #3
    b. If no, clinical review required.

3. Did the member have an inadequate response to a 12 week trial of methotrexate? (Provide documentation of inadequate response to methotrexate)

    a. If yes, continue to #6
    b. If no, continue to #4

4. Does the member have a contraindication or history of intolerance to methotrexate? (Provide documentation of contraindication and/or intolerance. Note: 1. Alcohol consumption is not considered a contraindication 2. Nausea to oral formulation is not considered an intolerance)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Does the member have a contraindication to all of the following OR an inadequate response to one 12 week trial with the following disease-modifying antirheumatic drugs: leflunomide, sulfasalazine, or hydroxychloroquine? (Provide documentation of contraindication or inadequate response to therapy)

    a. If yes, continue to #6
    b. If no, clinical review required

6. Is the request for Olumiant (baricitinib)?

    a. If yes, continue to #7
    b. If no, continue to #9

7. Does the member have a documented inadequate response, contraindication, or intolerance to TWO of the following agents: preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Rinvoq (upadacitinib), Simponi (golimumab), or Xeljanz/Xeljanz XR (tofacitinib citrate)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #8
    b. If no, clinical review required

8. Does the member have a documented inadequate response, contraindication, or intolerance to Kevzara (sarilumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)
    a. If yes, continue to #9
    b. If no, clinical review required
9. Is the medication being prescribed by or in consultation with a rheumatologist?
    a. If yes, approve 6 months unless otherwise specified
    b. If no, clinical review required

Juvenile Idiopathic Arthritis (JIA/PJIA)

1. Does the member have moderate to severe active polyarticular JIA defined as greater or equal to 5 swollen joints and at least 3 joints with limitation in motion? (Provide documentation of affected joints and current treatment regimen)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Did the member have an inadequate response to a 12 week trial of methotrexate? (Provide documentation of trial with inadequate response)
    a. If yes, continue to #5
    b. If no, continue to #3
3. Does the member have a contraindication or history of intolerance to methotrexate? (Provide documentation of contraindication and/or intolerance. Note: 1. Alcohol consumption is not considered a contraindication 2. Nausea to oral formulation is not considered an intolerance)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Did the member have a contraindication or history of intolerance to leflunomide? (Provide documentation of contraindication and/or intolerance)
    a. If yes, continue to #5
    b. If no, deny. Clinical criteria not met
5. Does the member have documentation of an inadequate response, intolerance, or contraindication to a preferred adalimumab biosimilar? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the request for Xeljanz oral tablet or Xeljanz oral solution? (Note: Xeljanz XR tablet is not FDA approved for treatment of PJIA)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the medication being prescribed by or in consultation with a rheumatologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Psoriatic Arthritis (PsA)

1. Does the member currently have active psoriatic arthritis defined as greater or equal to 3 swollen joints AND greater or equal to 3 tender or painful joints despite the current treatment regimen? (Provide documentation of affected joints and current treatment regimen)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member have a contraindication or an inadequate response to a 12 week trial with one of the following: methotrexate, leflunomide, cyclosporine, sulfasalazine? (Provide documentation of trial with inadequate response or contraindication)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Does the member have a documented inadequate response, contraindication, or intolerance to TWO of the following agents: preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Simponi (golimumab), Skyrizi (risankizumab) or Stelara (ustekinumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #6
    b. If no, continue to # 4

4. Is the request for Xeljanz (tofacitinib)?

    a. If yes, continue to #5
    b. If no, continue to #6

5. Does the member have documented needle phobia to the degree that the member has previously refused any injectable therapy or medical procedure? (refer to DSM-V-TR F40.2 for specific phobia diagnostic criteria)

    a. If yes, continue to #7
    b. If no, clinical review required

6. Does the member have a documented inadequate response, contraindication, or intolerance to BOTH of the following agents: Taltz (ixekizumab) and Xeljanz (tofacitinib)? (Provide documentation of inadequate responses, contraindication, and/or intolerance) [n/a if requested drug is Xeljanz]

    a. If n/a, continue to #7
    b. If yes, continue to # 7
    c. If no, clinical review required

7. Is the medication being prescribed by or in consultation with a rheumatologist or dermatologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

<u>Ankylosing Spondylitis (AS)</u>

1. Does the member currently have active AS despite a current treatment regimen as defined by the below? (Provide supporting documentation)

    - Bath ankylosing spondylitis disease activity index (BASDAI) greater or equal to 4 <u>OR</u>
    - Ankylosing Spondylitis Disease Activity Score (ASDAS) greater or equal to 2.1 <u>AND</u>
    - Elevated CRP, positive MRI, or Radiographic sacroiliitis

    a. If yes, continue to #2
    b. If no, clinical review required

2. Did the member have an inadequate response or intolerance to TWO separate 4 week trials of prescription strength oral nonsteroidal anti-inflammatory drugs (NSAIDs)? (Provide documentation of NSAIDs tried, examples: ibuprofen, naproxen, diclofenac, meloxicam, etc.)
    a. If yes, continue to #4
    b. If no, continue to #3
3. Does the member have a contraindication to oral NSAIDs? (Provide documentation of contraindication)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have isolated sacroilitis, or enthesitis disease? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, continue to #7
5. Did the member have an inadequate response to a parenteral glucocorticoid injection? (Provide documentation of trial with inadequate response)
    a. If yes, continue to #9
    b. If no, continue to #6
6. Does the member have a contraindication to a parenteral glucocorticoid injection? (Provide documentation of contraindication)
    a. If yes, continue to #9
    b. If no, clinical review required
7. Did the member have an inadequate response to a 12 week trial with sulfasalazine? (Provide documentation of trial with inadequate response)
    a. If yes, continue to #9
    b. If no, continue to #8
8. Does the member have a contraindication or history of intolerance to sulfasalazine? (Provide documentation of contraindication and/or intolerance)
    a. If yes, continue to #9
    b. If no, clinical review required
9. Does the member have a documented inadequate response, intolerance, or contraindication to TWO of the following agents: preferred adalimumab biosimilar, Cimzia (certolizumab pegol), or Simponi (golimumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)
    a. If yes, continue to #10
    b. If no, clinical review required
10. Is Rinvoq (upadacitinib) the requested medication?
    a. If yes, continue to #11
    b. If no, continue to #12
11. Does the member have a documented inadequate response, intolerance, or contraindication to Taltz (ixekizumab) or Xeljanz (tofacitinib)? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #12
    b. If no, clinical review required

12. Is the medication being prescribed by or in consultation with a rheumatologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Ulcerative Colitis (UC)

1. Does the member currently have active Ulcerative Colitis? (Provide documentation of diagnosis confirmed by endoscopy, colonoscopy, or sigmoidoscopy with Mayo score of greater than 2)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a documented inadequate response, intolerance, or contraindication to TWO of the following: preferred adalimumab biosimilar, Simponi (golimumab), or Stelara (ustekinumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)
    a. If yes, continue to #5
    b. If no, continue to #3
3. What is the requested medication?
    a. Xeljanz (tofacitinib), continue to #4
    b. Rinvoq (upadacitinib), clinical review required
4. Does the member with needle phobia to the degree that the member has previously refused any injectable therapy or medical procedure? (refer to DSM-V-TR F40.2 for specific phobia diagnostic criteria)
    a. If yes, continue to #6
    b. If no, clinical review required
5. Does the member have a documented inadequate response, intolerance, or contraindication to Xeljanz (tofacitinib)? (Provide documentation of inadequate response, contraindications, and/or intolerances)
    a. If yes, continue to #6
    b. If n/a, continue to #6
    c. If not, clinical review required
6. Is the medication being prescribed by, or in consultation with, a gastroenterologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Crohn's Disease (CD)

1. Does the member currently have active CD defined as a Crohn's Disease Activity Index (CDAI) greater than 220 despite the current treatment regimen? (Provide documentation of CDAI and current treatment regimen)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Did the member have an inadequate response to TWO of the following oral agents for a minimum trial of 12 weeks each: 6-mercaptopurine, azathioprine, corticosteroid, methotrexate, mesalamine, or sulfasalazine? (Provide documentation of 12-week trials with inadequate responses)
    a. If yes, continue to #4
    b. If no, continue to #3
3. Does the member have a contraindication or history of intolerance to at least TWO of the following oral

agents: 6-mercaptopurine, azathioprine, corticosteroids, methotrexate, mesalamine, or sulfasalazine? (Provide documentation of contraindications and/or intolerances)

a. If yes, continue to #4
b. If no, clinical review required

4. Does the member have a documented inadequate response, contraindication, or intolerance to TWO of the following agents: preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Skyrizi (risankizumab) or Stelara (ustekinumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

a. If yes, continue to #5
b. If no, clinical review required

5. Is the medication being prescribed by, or in consultation with, a gastroenterologist?

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

Atopic Dermatitis

1. Does the member have at least 10% body surface area involvement? (Provide documentation of body surface area affected)

a. If yes, continue to #2
b. If no, clinical review required

2. Is the request for Dupixent (dupilumab)?

a. If yes, continue to #4
b. If no, continue to #3

3. Does the member have a documented trial with insufficient response, intolerance or contraindication to Dupixent (dupilimab)? (Provide documentation to support insufficient response, intolerance, and/or contraindication)

a. If yes, continue to #4
b. If no, continue to #5

4. Does the member have a documented trial with insufficient response, or intolerance, or contraindication to TWO of the following therapies: topical agent (high-potency steroid/calcineurin inhibitor), immunomodulatory agent (azathioprine, cyclosporine, methotrexate, mycophenolate), or phototherapy? (Provide supporting documentation of all therapies tried)

a. If yes, continue to #8
b. If no, clinical review required

5. Does the member have a documented trial with insufficient response, or intolerance, or contraindication to both a high-potency topical steroid (i.e. clobetasol 0.05%, fluocinonide 0.1%, halobetasol 0.05%, or betamethasone dipropionate 0.05%) and a topical calcineurin inhibitor (i.e. tacrolimus or pimecrolimus)? (Provide supporting documentation of all therapies tried)

a. If yes, continue to #6
b. If no, clinical review required

6. Does the member have a documented trial with insufficient response or intolerance or contraindication to at least one systemic immunomodulatory agent (i.e. azathioprine, cyclosporine, methotrexate, or mycophenolate)? (Provide supporting documentation of all therapies tried)

a. If yes, continue to #7
b. If no, clinical review required

7. Does the member have a documented trial with insufficient response, intolerance, or contraindication to phototherapy? (Provide documentation to support insufficient response, intolerance, and/or contraindication)

    a. If yes, continue to #8
    b. If no, clinical review required

8. Is the requested drug being prescribed by or in consultation with a dermatologist, allergist, or immunologist?

    a. If yes, approve for 6 months unless otherwise
    b. If no, clinical review required

Other Indications

1. Is the request for a FDA approved indication?

    a. If yes, continue to #4
    b. If no, continue to #2

2. Is the requested use supported by major compendia not otherwise excluded by plan design?

    a. If yes, continue to #3
    b. If no, clinical review required

3. Has the member tried and had an inadequate response OR does the member have a contraindication to ALL standard treatment options for the requested indication? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is the treatment being prescribed by or in consultation with an appropriate specialist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication FDA-approved or supported by major compendia? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Were updated chart notes (dated within 1 year) provided with documentation of significant clinical response to therapy? (Provide supporting documentation)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Will the requested medication be used with other biologic therapy? (Examples: Enbrel, Actemra, Cimzia, Simponi, Orencia, Taltz, Cosentyx, Otezla, etc)

    a. If yes, clinical review required
    b. If no, continue to #4

4. Is the treatment being prescribed by or in consultation with an appropriate specialist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:
Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Cibinqo [prescribing information]. New York, NY: Pfizer Inc., February 2022.
2. Rinvoq [prescribing information]. North Chicago, IL: AbbVie Inc., May 2023.
3. Olumiant [prescribing information]. Indianapolis, IN: Eli Lilly and Company., June 2022.
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Joenja® (leniolisib)
Prior Authorization Guidelines

Affected Medication(s)

- Joenja (leniolisib) oral tablet

FDA Approved Indication(s)

- Treatment of activated phosphoinositide 3-kinase delta (PI3Kδ) syndrome in adult and pediatric patients 12 years of age and older

Dosing

- 70 mg orally twice daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is Joenja® (leniolisib) being requested for an FDA approved indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 12 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have confirmed APDS-associated PI3Kδ mutation with a documented variant in PIK3CD or PIK3R1? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have nodal and/or extranodal lymphoproliferation with measurable index lesions? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have clinical findings and manifestations compatible with APDS such as recurrent sinopulmonary infections, intermittent herpesvirus viremia, and/or organ dysfunction? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Will Joenja be used in combination with immunosuppressive medications, PI3Kδ inhibitors, or B-cell depleters such as rituximab? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #8

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8. Is the treatment being prescribed by, or in consultation with, an immunologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within past year) provided with documentation of clinical response to prior therapy received (ex. decrease in size of index lesions, decrease in infections)?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, an immunologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

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Juxtapid® (lomitapide mesylate)
Prior Authorization Guidelines

Affected Medication(s)

- Juxtapid oral capsule

FDA Approved Indication(s)

- Indicated as an adjunct to a low-fat diet and other lipid-lowering treatments, including LDL apheresis where available, to reduce low-density lipoprotein cholesterol (LDL-C), total cholesterol (TC), apolipoprotein B (apo B), and non-high-density lipoprotein cholesterol (non-HDL-C) in patients with homozygous familial hypercholesterolemia (HoFH)

Dosing

- Initially 5mg orally once daily
- Refer to package insert for dose titration chart

Initial Authorization Criteria

1. Has the requested medication previously been approved by OHSU?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the medication being prescribed by or in consultation with cardiologist, endocrinologist, or lipid specialist?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a diagnosis of Homozygous Familial Hypercholesterolemia (HoFH) as supported by one of the following? (Provide supporting documentation)
    - Genetic mutation testing (mutations in LDL receptor gene, proprotein convertase subtilisin kexin9 [PCSK9] gene, apo B gene, LDL receptor adaptor protein 1 [LDLRAP1] gene)
    - Treated LDL-C ≥ 300 mg/dL or untreated LDL-C ≥ 500 mg/dL AND one of the following (i or ii):
        i. Tendon or cutaneous xanthoma prior to age 10 years
        ii. Evidence of HeFH in both parents (e.g., documented history of elevated LDL-C ≥ 190 mg/dL prior to lipid-lowering therapy)

    a. If yes, continue to #5
    b. If no, clinical review required
5. Is all of the following documentation received? (Provide supporting documentation)
    - Complete lipid panel performed within the last 3 months

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- Baseline LDL-C
- Documentation of dietary measures being undertaken to lower cholesterol

a. If yes, continue to #6
b. If no, clinical review required

6. Has the member been on high-intensity statin therapy for the last 3 consecutive months and will continue with high-intensity statin therapy? (High-intensity statin therapy includes: atorvastatin 40-80 mg or rosuvastatin 20-40 mg) (Document current statin regimen with initiation date and verify adherence)

    a. If yes, continue to #11
    b. If no, continue to #7

7. What is the rationale provided for avoiding high-intensity statin therapy? (Provide supporting documentation for review)

    a. Statin intolerance due to myalgia or myopathy, continue to #8
    b. History of rhabdomyolysis with creatinine kinase (CK) levels greater than 10-times upper limit of normal (document date occurred) OR labeled contraindication to all statin therapy, continue to #11
    c. All other rationale, clinical review required

8. Is the member currently receiving a maximally tolerated dose of a statin AND ezetimibe and will continue statin and ezetimibe with the requested medication?

    a. If yes, continue to #11
    b. If no, continue to #9

9. Is documentation of persistent myalgia or myopathy on two separate 8-week trials with pravastatin, rosuvastatin, or fluvastatin provided?

    a. If yes, continue to #10
    b. If no, clinical review required

10. Has the member been on ezetimibe for 3 consecutive months and will continue with the requested medication?

    a. If yes, continue to #11
    b. If no, clinical review required

11. Does the member have a documented trial with insufficient response, intolerance, or contraindication to a PCSK9 inhibitor (Examples: Repatha or Praluent)? (Provide supporting documentation)

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the medication being prescribed by or in consultation with cardiologist, endocrinologist, or lipid specialist?

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)

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a. If yes, continue to #3
b. If no, clinical review required

3. Does the member have a diagnosis of Homozygous Familial Hypercholesterolemia (HoFH) as supported by one of the following? (Provide supporting documentation)

    - Genetic mutation testing (mutations in LDL receptor gene, proprotein convertase subtilisin kexin9 [PCSK9] gene, apo B gene, LDL receptor adaptor protein 1 [LDLRAP1] gene)
    - Treated LDL-C ≥ 300 mg/dL or untreated LDL-C ≥ 500 mg/dL AND one of the following (i or ii):
        i. Tendon or cutaneous xanthoma prior to age 10 years
        ii. Evidence of HeFH in both parents (e.g., documented history of elevated LDL-C ≥ 190 mg/dL prior to lipid-lowering therapy)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Has the member demonstrated a positive clinical response to therapy? (Provide documentation of an LDL-C reduction since the initiation of therapy)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Is an updated lipid panel result received? (Provide lipid panel results for review)

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Juxtapid Prescribing Information. Cambridge, MA: Aegerion Pharmaceuticals, Inc.; January 2020. Available at http://www.juxtapidpro.com/prescribing-information.
2. Stone NJ, Robinson JG, Lichtenstein AH, et al. 2013 ACC/AHA guideline on the treatment of blood cholesterol to reduce atherosclerotic cardiovascular risk in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. Circulation. 2014 June 24; 129[suppl 2]: S1-S45.
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Last Reviewed: 11/7/18, 9/16/20, 9/15/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 11/15/20, 3/15/23, 3/1/24

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Jynarque® (tolvaptan)
Prior Authorization Guidelines

Affected Medication(s)

- Jynarque oral tablet

FDA Approved Indication(s)

- To slow kidney function decline in adults at risk of rapidly progressing autosomal dominant polycystic kidney disease (ADPKD)

Dosing

- Initially: 60 mg orally per day as 45 mg taken on waking and 15 mg taken 8 hours later
- Titrate to 90 mg taken on waking and 30 mg taken 8 hours later if tolerated
- Refer to package insert for dose adjustments for patients taking moderate CYP 3A inhibitors

Initial Authorization Criteria

1. Is the request for continuation of Jynarque (tolvaptan) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a diagnosis of autosomal dominant polycystic kidney disease confirmed by ultrasonography, MRI/CT scan, or genetic testing? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the member at risk of rapidly-progressing autosomal dominant polycystic kidney disease (ADPKD) defined by any of the following? (Provide supporting documentation)
    - MAYO class 1D or 1E
    - MAYO class 1C with additional evidence of rapid disease progression
    - eGFR decline ≥ 3.0 mL/min/1.73 m^2 over a period of ≥ 4 years
    - An ultrasound determined kidney length of > 16.5 cm
    - PROPKD score >6
    - Age of ≤ 55 with CKD stage 3

    a. If yes, continue to #6
    b. If no, clinical review required

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6. Does the member have a contraindication to Jynarque (tolvaptan)? (Contraindications include: History of signs or symptoms of significant liver impairment or injury, use of Jynarque with strong CYP 3A inhibitors, uncorrected abnormal blood sodium concentrations, unable to sense or respond to thirst, hypovolemia, uncorrected urinary outflow obstruction, or anuria)
    a. If yes, clinical review required
    b. If no, continue to #7
7. Does the member have baseline liver function (ALT and AST) and bilirubin levels within normal range? (Provide documentation of AST, ALT and bilirubin levels taken within the previous 3 months for review)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Is the treatment being prescribed by or in consultation with a nephrologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member had a positive clinical response to therapy as defined by a slowing in the decline in kidney function and/or an improvement in kidney pain? (Provide supporting documentation of positive clinical response)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member experienced an increase in ALT, AST, or bilirubin to greater than 2 times the upper limit of normal? (Provide updated ALT, AST, and bilirubin levels for review)
    a. If yes, clinical review required
    b. If no, continue to #4
4. Is the treatment being prescribed by or in consultation with a nephrologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

Last Reviewed: 11/21/18, 11/18/20, 9/15/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 3/15/23, 3/1/24

References:

1. Jynarque [package insert]. Rockville, MD: Otsuka America Pharmaceutical, Inc.; November 2022.
2. Chapman AB, Devuyst O, Eckardt KU, et al. Autosomal Dominant Polycystic Kidney Disease (ADPKD): Report from a Kidney Disease: Improving Global Outcomes (KDIGO) Controversies Conference. 2/2017.
3. Srivastava A, Patel N. Autosomal dominant polycystic kidney disease. American Academy of Family Physician. 2014;90(5):303-307
4. Torres VE, Chapman AB, Devuyst O, et al. Tolvaptan in patients with autosomal dominant polycystic kidney disease. The New England Journal of Medicine. 2012;367:2407-18.
5. Müller, Roman-Ulrich, et al. "An update on the use of tolvaptan for autosomal dominant polycystic kidney disease: consensus statement on behalf of the ERA Working Group on Inherited Kidney Disorders, the European Rare Kidney Disease Reference Network and Polycystic Kidney Disease International." Nephrology Dialysis Transplantation 37.5 (2022): 825-839.

Last Reviewed: 11/21/18, 11/18/20, 9/15/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 3/15/23, 3/1/24

Kalydeco® (ivacaftor)
Prior Authorization Guidelines

Affected Medication(s)

- Kalydeco oral tablet
- Kalydeco oral granules

FDA Approved Indication(s)

- Treatment of cystic fibrosis (CF) in patients age 1 month and older who have one mutation in the CFTR gene that is responsive to ivacaftor potentiation based on clinical and/or in vitro assay data

Dosing

- For patients 6 years and older: One 150 mg tablet twice daily
- For patients 6 months to less than 6 years of age:
  - 5 kg to less than 7 kg: One 25 mg packet twice daily
  - 7 kg to less than 14 kg: One 50 mg packet twice daily
  - 14 kg or greater: One 75 mg packet twice daily
- For patients 4 months to less than 6 months:
  - 5 kg or greater: One 25 mg packet twice daily
- For patients 2 months to less than 4 months:
  - 3 kg or greater: One 13.4 mg packet twice daily
- For patients 1 month to less than 2 months:
  - 3kg or greater: One 5.8 mg packet twice daily

Initial Authorization Criteria

1. Is the request for continuation of Kalydeco (ivacaftor) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is there documentation that the member has a cystic fibrosis transmembrane conductance regulator (CFTR) gene mutation that is responsive to ivacaftor based on in vitro data and/or clinical data? (Provide documentation of specific mutation for review)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is there documentation of the member's pulmonary status (baseline FEV1) and liver function (ALT and AST) and are the liver enzymes within normal limits? (Provide documentation of pulmonary and liver tests for review)
   a. If yes, continue to #5
   b. If no, clinical review required

Last Reviewed: 10/3/18, 5/19/21, 5/18/22, 5/19/23, 5/17/24
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5. Is the member at least 1 month of age?

    a. If yes, continue to #6
    b. If no, clinical review required

6. Is Kalydeco (ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Were updated chart notes (within past year) provided with documentation of clinical response to prior therapy received? (Provide documentation of improvement of FEV1 from baseline and/or a reduction in the number of pulmonary exacerbations)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Has documentation been provided of liver function tests (ALT and AST) within the last year and are they within normal limits? (Provide ALT and AST levels for review)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is Kalydeco (ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis?

    a. If yes, approve for 12 months unless otherwise
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Kalydeco® (ivacaftor) [Prescribing Information]. Boston, MA: Vertex Pharmaceuticals Inc. August 2018.
2. Kalydeco®. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed August 31, 2018.

Last Reviewed: 10/3/18, 5/19/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 6/15/21, 7/1/23, 6/15/24

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Last Reviewed: 10/3/18, 5/19/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 6/15/21, 7/1/23, 6/15/24

Keveyis® (dichlorphenamide)
Prior Authorization Guidelines

Affected Medication(s)

- dichlorphenamide oral tablet
- Keveyis oral tablet
- Ormalvi oral tablet

FDA Approved Indication(s)

- Treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants

Dosing

- Initially: 50 mg twice daily
- Titrate as needed based on response with maximum dose of 200 mg per day

Initial Authorization Criteria

1. Is the request for continuation of dichlorphenamide therapy for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Are both baseline serum potassium and bicarbonate levels received? (Provide supporting labs for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a previous trial with inadequate response, intolerance, or contraindication to treatment with acetazolamide? (Provide documentation of trial with inadequate response, intolerance, or contraindication)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, continue to #6
    b. If no, clinical review required
6. What diagnosis is the medication being requested for?
    a. Hypokalemic periodic paralysis, continue to corresponding criteria
    b. Hyperkalemic periodic paralysis, continue to corresponding criteria

Hypokalemic periodic paralysis

Last Reviewed: 11/21/18, 3/17/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 6/15/23

1. Does the member have a history of two or more attacks of muscle weakness with documented serum potassium <3.5 mEq/L? (Provide supporting documentation of attacks with muscle weakness and serum potassium levels for review)

    a. If yes, continue to #4
    b. If no, continue to #2

2. Does the member have a history of one attack of muscle weakness and one attack of weakness in one family relative with documented serum potassium <3.5 mEq/L? (Provide supporting documentation of members' and their family relatives' attack with muscle weakness and serum potassium levels for review)

    a. If yes, continue to #4
    b. If no, continue to #3

3. Does the member have three or more of the following six clinical/laboratory features?

    - Onset in the first or second decade of life (childhood or teenage years)
    - Duration of attack (muscle weakness involving ≥1 limbs) longer than two hours
    - The presence of triggers (previous carbohydrate rich meal, symptom onset during rest after exercise, stress)
    - Improvement in symptoms with potassium intake
    - A family history of the condition or genetically confirmed skeletal calcium or sodium channel mutation
    - Positive long exercise test

    a. If yes, continue to #4
    b. If no, clinical review required

4. Have other causes of hypokalemia been ruled out? (i.e. renal, adrenal, thyroid dysfunction; renal tubular acidosis; diuretic and laxative abuse)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Have dietary interventions been trialed with inadequate response? (Provide documentation of high potassium intake and low sodium intake)

    a. If yes, continue to #6
    b. If no, clinical review required

6. Has the member been taking and will continue to take a potassium supplement throughout therapy? (Provide supporting documentation)

    a. If yes, approve for 6 months, unless otherwise specified
    b. If no, clinical review required

<u>Hyperkalemic periodic paralysis</u>

1. Has the member's diagnosis been confirmed by electromyography, genetic testing, or provocative testing? (Provide supporting documentation of testing done to confirm diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member had an ECG performed to exclude prolonged QT and/or ventricular arrhythmias? (Provide supporting documentation)

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a. If yes, continue to #3
b. If no, clinical review required

3. Have other causes of hyperkalemia been ruled out? (i.e. Andersen-Tawil syndrome, adrenal insufficiency, renal dysfunction, or drug abuse)

   a. If yes, continue to #4
   b. If no, clinical review required

4. Have dietary and exercise restrictions been trialed with inadequate response? (Provide documentation of low potassium intake and exercise restrictions)

   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)

   a. If yes, continue to #2
   b. If no, clinical review required

2. Has the member had a positive clinical response to therapy as defined by a decrease in the frequency in paralytic attacks? (Provide supporting documentation for review)

   a. If yes, continue to #3
   b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with, a neurologist?

   a. If yes, approve for 12 months unless otherwise specified
   b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Keveyis [prescribing information]. Taro Pharmaceuticals U.S.A., Inc. Hawthorne, NY. December 2021.
2. Vicart S, Sternberg D, Arzel-Hézode M, et al. Hypokalemic Periodic Paralysis. 2002 Apr 30 [Updated 2014 Jul 31]. In: Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2015. Available from: http://www.ncbi.nlm.nih.gov/books/NBK1338/?report=classic
3. Jurkat-Rott K, Lehmann-Horn F. Hyperkalemic Periodic Paralysis Type 1. 2003 Jul 18 [Updated 2011 May 31]. In: Pagon RA, Adam MP, Ardinger HH, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2015. Available from: http://www.ncbi.nlm.nih.gov/books/NBK1496/

Last Reviewed: 11/21/18, 3/17/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 6/15/24

Korlym® (mifepristone)
Prior Authorization Guidelines

Affected Medication(s)

- Korlym oral tablet
- mifepristone 300 mg oral tablet

FDA Approved Indication(s)

- To control hyperglycemia secondary to hypercortisolism in adult patients with endogenous Cushing's syndrome who have type 2 diabetes mellitus or glucose intolerance and have failed surgery or are not candidates for surgery

Dosing

- 300 mg orally once daily

Initial Authorization Criteria

1. Is the request for continuation of Korlym (mifepristone) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member a female of reproductive age?
    a. If yes, continue to #5
    b. If no, continue to #6
5. Has pregnancy been excluded by a negative pregnancy test and will the member use contraception while on therapy? (Provide documentation of negative pregnancy test and contraception use)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have endogenous hypercortisolism? (i.e. not hypercortisolism due to chronic high dose glucocorticoids) (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have type 2 diabetes or glucose intolerance secondary to hypercortisolism confirmed by HbA1c? (Provide supporting documentation including current HbA1c)

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a. If yes, continue to #8
b. If no, clinical review required

8. Has the member failed surgery or is the member not a surgical candidate? (Trans-sphenoidal surgery for pituitary dependent Cushing's or surgical removal of an adrenocortical tumor in malignant Cushing's) (Provide supporting documentation of surgery or rationale for avoiding surgery)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Has the member had a trial with inadequate response or an intolerance to a steroidogenesis inhibitor? (i.e. ketoconazole, metyrapone)

    a. If yes, continue to #10
    b. If no, clinical review required

10. Does the member have a trial with inadequate response, an intolerance, or contraindication to treatment with maximum dose of metformin? Note: If patient has GI discomfort, metformin ER (Glucophage XR) should be trialed (Provide documentation of trial, intolerance, or contraindication)

    a. If yes, continue to #11
    b. If no, clinical review required

11. Is the treatment being prescribed by, or in consultation with, an endocrinologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the member a female of reproductive age?

    a. If yes, continue to #3
    b. If no, continue to #6

3. Have more than 14 days passed between the last dose and the next scheduled dose? (Provide documentation of date of last dose and date of next scheduled dose)

    a. If yes, continue to #4
    b. If no, continue to #5

4. Has pregnancy been excluded by a negative pregnancy test? (Provide documentation of date of last dose and negative pregnancy test if indicated)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Does the member use contraception while on therapy? (Provide documentation of contraception use)

    a. If yes, continue to #6

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b. If no, clinical review required

6. Has the member had a positive clinical response to therapy as defined by improved glucose control? (Provide supporting documentation including an updated HbA1c)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the treatment being prescribed by, or in consultation with, an endocrinologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Korlym [package insert]. Corcept Therapeutics Inc., Menlo Park, CA, December 2022.
2. Johanssen S, Allolio B. Mifeprisone (RU 486) in Cushing's syndrome. European Journal of Endocrinology/European Federation of Endocrine Societies 2007; 157: 561-569.
3. Barbot M, Ceccato F, Scaroni C. Diabetes Mellitus Secondary to Cushing's Disease. Front Endocrinol (Lausanne). 2018;9:284. Published 2018 Jun 5. doi:10.3389/fendo.2018.00284
4. Nieman LK, Biller BM, Findling JW. Treatment of Cushing's Syndrome: An Endocrine Society Clinical Practice Guideline. J Clin Endocrinol Metab. 2015;100(8):2807-2831.

Last Reviewed: 11/21/18, 5/19/21, 7/20/22, 3/17/23, 3/15/24
Effective Date: 1/1/19, 4/15/24

Koselugo® (selumetinib)
Prior Authorization Guidelines

Affected Medication(s)

- Koselugo oral capsule

FDA Approved Indication(s)

- Treatment of pediatric patients 2 years of age and older with neurofibromatosis type 1 (NF1) who have symptomatic, inoperable plexiform neurofibromas (PN)

Dosing

- Refer to corresponding package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of a previously approved Koselugo (selumetinib) prior authorization and indication is for the same as previous approval?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member currently have a diagnosis of neurofibromatosis type 1 confirmed through genetic testing or 2 or more of the following diagnostic criteria? (Provide supporting documentation)
        i. 6 or more café-au-lait macules (greater than or equal to 0.5 cm in prepubertal subjects or greater than or equal to 1.5 cm in post pubertal subjects)
        ii. Two or more neurofibromas of any type or one plexiform neurofibroma
        iii. Freckling in the axillary or inguinal region
        iv. Optic glioma
        v. Two or more Lisch nodules (iris hamartomas)
        vi. A distinctive osseous lesion such as sphenoid dysplasia or thinning of long bone cortex with or without pseudarthrosis
        vii. A heterozygous pathogenic *NF1* variant with a variant allele fraction of 50% in apparently normal tissue such as white blood cells
        viii. A first-degree relative (parent, sibling, or offspring) with NF-1 by the above criteria
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have at least one measurable (at least 3cm in one dimension) plexiform neurofibroma that is inoperable? (provide supporting documentation)
    a. If yes, continue to #5

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b. If no, clinical review required

5. Is the member's plexiform neurofibroma symptomatic, disfiguring, or growing in size? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the treatment being prescribed by, or in consultation with, an oncologist or neurologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within the past 6 months) provided with documentation of a clinical positive response defined as a decrease in or maintenance of plexiform neurofibroma volume compared to pre-therapy baseline? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with an oncologist or neurologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Koselugo [selumetinib] capsules. AstraZeneca Pharmaceuticals, LP. Wilmington, DE; December 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 26 May. 2020].
3. Widemann, Brigitte C., et al. "Phase I study of the MEK1/2 inhibitor selumetinib (AZD6244) hydrogen sulfate in children and young adults with neurofibromatosis type 1 (NF1) and inoperable plexiform neurofibromas (PNs)." (2014): 10018-10018.
4. Gross AM, Wolters P, Baldwin A et al. SPRINT: Phase II study of the MEK ½ inhibitor selumetinib (AZD6244, ARRY142886) in children with neurofibromatosis type 1 (NF1) and inoperable plexiform neurofibromas (PN).

Last Reviewed: 9/16/20, 9/15/21, 11/16/22, 11/17/23
Effective Date: 11/15/20, 11/1/21, 1/1/23, 12/20/23

Journal of Clinical Oncology. 2018; 36(15): 10503. Available from: http://ascopubs.org/doi/abs/10.1200/JCO.2018.36.15_suppl.10503. Accessed August 12, 2020.
5. Miller DT, Freedenberg D, Schorry E, et al. Health Supervision for Children With Neurofibromatosis Type 1. Pediatrics. 2019;143(5):e20190660
6. Dombi E, Baldwin A, Marcus L, et al. Activity of selumetinib in neurofibromatosis type-1 related plexiform neurofibromas. N Engl J Med. 2016; 375(26): 2550-2560.
7. Ferner, Rosalie E., et al. "Guidelines for the diagnosis and management of individuals with neurofibromatosis 1." Journal of medical genetics 44.2 (2007): 81-88.
8. National Institutes of Health Consensus Development Conference Statement: neurofibromatosis. Bethesda, Md., USA, July 13-15, 1987. Neurofibromatosis 1:172-178,1988
9. Legius E, Messiaen L, Wolkenstein P, et al. Revised diagnostic criteria for neurofibromatosis type 1 and Legius syndrome: An international consensus recommendation. Genet Med 2021; 23:1506.

Kuvan® (sapropterin dihydrochloride)
Prior Authorization Guidelines

Affected Medication(s)

- Kuvan powder for oral solution
- Kuvan oral tablet
- Sapropterin powder for oral solution
- Sapropterin oral tablet
- Javygtor powder for oral solution
- Javygtor oral tablet

FDA Approved Indication(s)

- To reduce blood phenylalanine (Phe) levels in patients with hyperphenylalaninemia (HPA) due to tetrahydrobiopterin- (BH4-) responsive Phenylketonuria (PKU)

Dosing

- Patients 1 month to 6 years: Starting dose of 10mg/kg once daily then dose adjust based on response
- Patients 7 years and older: Starting dose of 10 to 20 mg/kg once daily then dose adjust based on response

Initial Authorization Criteria

1. Is the request for continuation of Kuvan (sapropterin dihydrochloride) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Will Kuvan (sapropterin dihydrochloride) be used in conjunction with a phenylalanine-restricted diet (i.e. foods with high protein such as meat, fish, eggs, and milk products should be avoided)? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the baseline phenylalanine level provided and does it exceed 360 µmol/L? (Provide baseline phenylalanine)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Will the member have a phenylalanine blood level measured after 1 week of therapy and then periodically for up to 2 months of therapy?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the treatment being prescribed by or in consultation with a specialist experienced in treatment of hyperphenylalaninemia?
    a. If yes, approve for 2 months unless otherwise specified

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b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is Kuvan (sapropterin dihydrochloride) being used in conjunction with a phenylalanine-restricted diet (i.e. foods with high protein such as meat, fish, eggs, and milk products should be avoided)? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member demonstrated a positive clinical response to therapy as defined by a decrease in average blood Phenylalanine levels by at least 30% below pretreatment baseline? (Provide pretreatment and updated phenylalanine levels)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Will the member's blood phenylalanine levels continue to be monitored throughout therapy?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by or in consultation with a specialist experienced in treatment of hyperphenylalaninemia?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Kuvan (sapropterin dihydrochloride) [Prescribing Information]. Novato, CA: BioMarin Pharmaceutical, Inc. February 2021 .
2. Vockley, Jerry, et al. "Phenylalanine hydroxylase deficiency: diagnosis and management guideline." Genetics in Medicine 16.2 (2014): 188.

Last Reviewed: 11/21/18, 11/18/20, 1/19/22, 11/16/22, 7/21/23
Effective Date: 1/1/19, 12/15/20, 1/1/23

Leukine® (sargramostim)
Prior Authorization Guidelines

Affected Medication(s)

- Leukine for subcutaneous injection

FDA Approved Indication(s)

- In adult patients with cancer undergoing autologous hematopoietic stem cell transplantation for the mobilization of hematopoietic progenitor cells into peripheral blood for collection by leukapheresis
- To increase survival in adult and pediatric patients from birth to 17 years of age acutely exposed to myelosuppressive doses of radiation (Hematopoietic Syndrome of Acute Radiation Syndrome [H-ARS])
- To shorten time to neutrophil recovery and to reduce the incidence of severe, life-threatening, or fatal infections following induction chemotherapy in adult patients 55 years and older with acute myeloid leukemia (AML)
- For the acceleration of myeloid reconstitution following autologous peripheral blood progenitor cell (PBPC) or bone marrow transplantation in adult and pediatric patients 2 years of age and older with non-Hodgkin's lymphoma (NHL), acute lymphoblastic leukemia (ALL) and Hodgkin's lymphoma (HL)
- For the acceleration of myeloid reconstitution in adult and pediatric patients 2 years of age and older undergoing allogeneic bone marrow transplantation from HLA-matched related donors
- For the treatment of adult and pediatric patients 2 years and older who have undergone allogeneic or autologous bone marrow transplantation in whom neutrophil recovery is delayed or failed

Dosing

- Refer to package insert for specific dosing recommendations

Authorization Criteria

1. Is the medication being requested for an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is Leukine (sargramostim) being requested for one of the following FDA approved indications? (For intravenous administration request, contact medical benefit administer for coverage)

    - Peripheral Blood Progenitor Cell (PBPC) mobilization, collection, or transplantation
    - Patients acutely exposed to myelosuppressive doses of radiation (Hematopoietic Subsyndrome of Acute Radiation Syndrome [H-ARS])

    a. If yes, continue to #3
    b. If no, clinical review required

3. Does the member have a documented trial with insufficient response, intolerance, or contraindication to myeloid growth factors (Examples: Zarxio, Neupogen, etc)? (Provide supporting documentation)

    a. If yes, continue to #4
    b. If no, clinical review required

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Effective Date: 1/1/19, 6/15/24

4. Is the treatment being prescribed by or in consultation with an oncologist, hematologist, or a transplant specialist?

    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Leukine [package insert]. Bridgewater, NJ; sanofi-aventis US LLC; May 2021. Accessed April 2022.
2. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) sargramostim. National Comprehensive Cancer Network, 2022. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed April 2022.
3. Wisconsin Physicians Service Insurance Corporation. Local Coverage Determination (LCD): Human Granulocyte/Macrophage Colony Stimulating Factors (L34699). Centers for Medicare & Medicaid Services, Inc. Updated on 1/23/2018 with effective date 02/1/2018. Accessed March 2018.
4. Palmetto GBA. Local Coverage Determination (LCD): White Cell Colony Stimulating Factors (L37176). Centers for Medicare & Medicaid Services, Inc. Updated on 12/7/2017 with effective date 2/26/2018. Accessed March 2018.

Last Reviewed: 11/7/18, 3/17/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 615/24

Livmarli® (maralixibat)
Prior Authorization Guidelines

Affected Medication(s)

- Livmarli (maralixibat) oral tablet

FDA Approved Indication(s)

- Treatment of cholestatic pruritus in patients with Alagille syndrome (ALGS) 3 months of age and older

Dosing

- Initiate dose at 190 mcg/kg orally once daily, then after one week, increase to 380 mcg/kg as tolerated.

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is Livmarli (maralixibat) being requested for an FDA approved indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the patient 3 months of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of Alagille syndrome confirmed by genetic testing or liver biopsy? (genetic testing with JAG1 or NOTCH2 mutation present) (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member previously trialed a maximum tolerated dose of all of the following for at least 4 weeks with treatment failure, or is there a documented intolerance/contraindication to all of the following: rifampin, ursodiol, and cholestyramine/colesevelam? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is there documentation that member is experiencing moderate to severe pruritus despite current therapy? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is Livmarli (maralixibat) being prescribed by, or in consult with, a gastroenterologist or specialist experienced in treating Alagille syndrome?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Last Reviewed: 5/18/22, 5/19/23, 11/17/23
Effective Date: 7/15/22, 7/1/23, 12/20/23

Reauthorization Criteria

1. Is the documented indication approved by the FDA? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within past year) provided with documentation of significant clinical response to prior therapy received? (i.e. decrease in severity of pruritus)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is Livmarli (maralixibat) being prescribed by, or in consult with, a gastroenterologist or specialist experienced in treating Alagille syndrome?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. LIVMARLI™ (maralixibat) oral solution, [package insert]. Foster City, CA: Mirum Pharmaceuticals, Inc; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 23 Nov. 2021].
3. Gonzales, Emmanuel, et al. "Efficacy and safety of maralixibat treatment in patients with Alagille syndrome and cholestatic pruritus (ICONIC): a randomized phase 2 study." The Lancet 398.10311 (2021): 1581-1592.
4. Poupon R, Chopra S. Pruritus associated with cholestasis. In: UpToDate, K. Lindor, S. Grover (Eds), UpToDate, Waltham, MA. (Accessed on April 14, 2022.)

Last Reviewed: 5/18/22, 5/19/23, 11/17/23
Effective Date: 7/15/22, 7/1/23, 12/20/23

Lucemyra® (lofexidine)
Prior Authorization Guidelines

Affected Medication(s)

- Lucemyra oral tablet

FDA Approved Indication(s)

- For mitigation of opioid withdrawal symptoms to facilitate abrupt opioid discontinuation in adults

Dosing

- Recommended starting dose: is 0.54 mg 4 times daily during the period of peak withdrawal symptoms
- Continue for up to 14 days
- Gradual dose reduction over 2-4 day period

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Was the therapy with Lucemyra (lofexidine) been initiated at the inpatient setting?
    a. If yes, approve for 14 days
    b. If no, continue to #4
4. Is there medical rationale why an opioid taper cannot be used? (Examples of opioid taper medications include: buprenorphine, methadone, other opioids)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member had a trial with inadequate response, intolerance, or contraindication to clonidine?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the medication being prescribed by, or in consultation with, a provider specializing in pain management, or addiction medicine?
    a. If yes, approve for 14 days
    b. If no, clinical review required

Last Reviewed: 12/19/18, 7/21/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 4/15/24

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Lucemyra (lofexidine) [Prescribing Information]. Louisville, KY: US WorldMeds, LLC. July 2018.
2. Berna C, Kulich RJ, Rathmell JP. Tapering Long-term Opioid Therapy in Chronic Noncancer Pain: Evidence and Recommendations for Everyday Practice. Mayo Clin Proc 2015; 90:828.
3. ASAM: The ASAM National Practice Guideline for the Use of Medications in the Treatment of Addiction Involving Opioid Use, American Society of Addiction Medicine. http://www.asam.org/docs/default-source/practice-support/guidelines-and-consensus-docs/national-practice-guideline.pdf (Accessed on December 11, 2018).
4. Meader, Nicholas. "A comparison of methadone, buprenorphine and alpha2 adrenergic agonists for opioid detoxification: a mixed treatment comparison meta-analysis." *Drug and alcohol dependence* 108.1-2 (2010): 110-114.
5. Cunningham, Chinazo, et al. The ASAM National Practice Guideline for the Treatment of Opioid Use Disorder: 2020 Focused Update.Journal of Addiction Medicine: March/April 2020 - Volume 14 - Issue 2S - p 1-91.
6. Perry, Christopher, et al. "The management of substance use disorders: synopsis of the 2021 US Department of Veterans Affairs and US Department of Defense clinical practice guideline." Annals of internal medicine 175.5 (2022): 720-731.

Last Reviewed: 12/19/18, 7/21/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 4/15/24

Lupkynis® (voclosporin)
Prior Authorization Guidelines

Affected Medication(s)

- Lupkynis (voclosporin) oral capsule

FDA Approved Indication(s)

- In combination with a background immunosuppressive therapy regimen for treatment of adult patients with active lupus nephritis

Dosing

- 23.7 mg twice daily

Initial Authorization Criteria

1. Is the request for renewal of a previously approved Lupkynis (voclosporin) prior authorization with the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a confirmed diagnosis of active lupus nephritis? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member 18 years of age or older?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member trialed at least 2 of the following medications with an inadequate response: cyclosporine, tacrolimus, mycophenolate mofetil, azathioprine? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, continue to #6
6. Does the member have documentation of intolerance and/or contraindication to all of the following: cyclosporine, tacrolimus, mycophenolate mofetil, azathioprine? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have a previous trial with inadequate response, intolerance, or contraindication to Benlysta (belimumab)? (Provide documentation of trial, intolerance, or contraindication)
    a. If yes, continue to #7
    b. If no, clinical review required
8. Will the member be using Lupkynis in combination with mycophenolate and corticosteroids? (Provide treatment plan)

Last Reviewed: 3/17/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 5/1/21, 7/15/22, 7/1/23, 6/15/24

a. If yes, continue to #8
b. If no, clinical review required

9. Is the requested medication being prescribed by, or in consultation with, a rheumatologist or a specialist experienced in treatment of SLE?

    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is Lupkynis (voclosporin) being requested for an FDA approved or major compendia supported indication? (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Were updated chart notes (within 1 year) with documentation of significant clinical response to therapy defined as one of the following received? (Provide supporting documentation)

    - A decrease in urine protein-to-creatinine ratio to ≤0.5 mg/mg plus a eGFR >60mL/min/1.73m^2
    - No decrease of 20% or more from baseline eGFR

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the requested medication being prescribed by, or in consultation with, a rheumatologist or specialist experienced in treatment of SLE?

    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. LUPKYNIS (voclosporin) capsules, [package insert]. Rockville, MD: Aurinia Pharmaceuticals, Inc.; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 12 Jan. 2021].
3. Rovin, Brad H., et al. "A randomized, controlled double-blind study comparing the efficacy and safety of dose-ranging voclosporin with placebo in achieving remission in patients with active lupus nephritis." Kidney international 95.1 (2019): 219-231.
4. Mok, Chi Chiu. "Calcineurin inhibitors in systemic lupus erythematosus." Best Practice & Research Clinical Rheumatology 31.3 (2017): 429-438.
5. Kidney Disease: Improving Global Outcomes (KDIGO) Lupus Nephritis Work Group. KDIGO 2024 Clinical Practice Guideline for the management of LUPUS NEPHRITIS. *Kidney Int*. 2024;105(1S):S1-S69.

Last Reviewed: 3/17/21, 5/18/22, 5/19/23, 5/17/24
Effective Date: 5/1/21, 7/15/22, 4/1/23, 6/15/24

Medical Necessity
Prior Authorization Guidelines

Affected Medication(s)

- Abilify Mycite Kit
- Absorica/ LD
- Apap-caffeine-dihydrocodeine 325
- Acthar gel
- Acticlate 150mg tablet
- Actoplus Met XR
- Adlarity patch
- Admelog
- Adzenys XR-ODT
- Aemcolo
- Afrezza
- Agamree suspension
- Airsupra
- Alkindi
- Aplenzin
- Amphetamine sulfate
- Akynzeo
- Ala-scalp lotion
- Altoprev
- Amcinonide cream and ointment
- Amzeeq
- Anzemet
- Apadaz
- Apexicon E cream
- Apidra
- Aplenzin
- Aptensio XR
- Aspruzyo sprinkle ER
- Atorvaliq
- Auvelity
- Auvi-Q
- Azstarys
- Baclofen suspension
- Baxdela
- Beconase AQ
- Belbuca Film
- Belsomra
- Benzhydroco-apap
- Bevespi Aerosphere
- Bismuth-metronidazole-tetracycline capsule
- Dyanavel XR
- Dyrenium
- Elepsia XR
- Elyxyb solution
- Emflaza
- Emsam patch
- Entadfi capsule
- Eohilia
- Equetro capsule
- Ergomar
- Ertaczo 2% cream
- Eulexin
- Evamist spray
- Evekeo, Evekeo ODT
- Exelderm 1% solution & cream
- Exservan
- Eysuvis
- Ezetimibe-atorvastatin
- Fenoglide (fenofibrate)
- Fenoprofen
- Fibricor (fenofibrate)
- Fioricet w/ Codeine
- Firdapse
- Flagyl 375mg capsule
- Flector patch
- Fleqsuvy
- FloLipid
- Fluoxetine DR 90 mg capsule
- Flurandrenolide cream
- Furoscix
- Gabapentin ER tablet
- Gimoti
- Glumetza ER
- Glycate
- Glycopyrrolate 1.5mg tablet
- Gocovri
- Gralise
- Granisetron tablet
- H.P. Acthar
- Halobetasol foam
- Lyumjev
- Lyvispah packet
- Meloxicam capsule
- Meperidine tablet/solution
- Metaxalone 400mg tab
- Metformin 625 mg tab
- Metformin ER OSM/gastr
- Methamphetamine tablet
- Methocarbamol 1000mg
- Methoxsalen 10 mg cap
- Methsuximide
- Methylphenidate ER 10mg, 15mg, 20mg, 30mg, 40mg, 50mg, 60mg capsules (Aptensio generic)
- Methylphenidate ER 45mg, 63mg, and 72 mg tablet (Relexxi generic)
- Metoclopramide ODT
- Metronidazole 375mg capsule
- Miacalcin
- Miconazole-zinc-petro 0.25-15%
- Migergot suppository
- Mondoxyne 75mg capsule
- Monodox 75mg capsule
- Motofen
- Motpoly XR capsule
- Mydayis ER
- Nalfon
- Nalocet
- Namzaric
- Naprelan
- Naproxen CR
- Naproxen-esomeprazole
- Nascobal spray
- Natesto
- Nexiclon XR
- Qudexy XR
- Quillichew ER
- Quillivant XR
- Quviviq
- Rasuvo
- Rayos DR
- Reltone capsule
- Regranex
- Relexxii ER
- Reyvow
- Ribavirin
- Ridaura
- Rocklatan
- Roxybond
- Sancuso
- Scalacort lotion
- Seglentis
- Sernivo spray
- Sertraline capsule
- Seysara
- Sivextro
- Soliqua 100-33
- Soltamox
- Sorilux 0.005% foam
- Sovaldi tablet/ pellet
- Sovuna
- Spritam
- Strensiq
- Sular ER tablet
- Sulconazole nitrate 1% soln & cream
- Sumatriptan-naproxen
- Suprax chewable tablet
- Symax duotab
- Sympazan Film
- Syprine
- Tadliq suspension
- Talicia DR
- Tasmar
- Tavaborole 5% solution
- Tarpeyo (budesonide)
- Teglutek suspension
- Terbutaline

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Medical Necessity
Prior Authorization Guidelines

- Bonjesta
- Brexafemme
- Breztri Aerosphere
- Bryhali lotion
- Bupap 50-300
- Butalbital-acetaminophen 50-300
- Butalb-apap-caffeine w/ codeine 50-300-40-30 capsule
- Butorphanol Spray
- Cabtreo gel
- Calcipotriene 0.005% foam
- Calcitonin-Salmon
- Cambia packet
- Carisoprodol-aspirin-codeine tablet
- Celontin
- Chlorzoxazone 250/375/750mg tablets
- Citalopram capsule
- Clocortilone 0.1% cream pump
- Cloderm cream pump
- Clonidine ER 0.17 mg
- Condylox 0.5% gel
- Conjupri
- Cordran cream, ointment, and tape
- Cortisone tablet
- Cortrophin Gel
- Cotempla XR-ODT
- Coxanto capsule
- Cuvrior
- Cyanocobalamin spray
- Cycloset
- Cystadane
- Daraprim
- Dartisla ODT
- Daytrana (methylphenidate)
- Deflazacort
- Desonide gel
- Desoxyn (methamphetamine)
- Halocinonide cream
- Halog cream, ointment, and solution
- Harvoni tablet/ pellet
- Hemady
- Hemlibra
- Horizant
- Humalog Mix
- Humalog U-100/200/JR
- Humalog Tempo Pen
- Humulin 70/30
- Humulin N, Humulin R
- Hydrocodone ER
- Hysingla ER
- Ibsrela
- Igalmi film
- Impoyz cream
- Inderal XL
- Indocin suspension
- Indocin suppository
- Indomethacin suppository
- Indomethacin suspension
- Intrarosa
- Innopran XL
- Insulin lispro Pen
- Insulin lispro Vial
- Isordil 40mg tablet
- Isosorbide dinitrate 40mg tablet
- Isotretinoin 25mg & 35 mg capsules (Absorica generic)
- Iyuzeh drops
- Jalyn (dutasteride-tamsulosin)
- Jornay PM
- Jublia
- Kapvay
- Kerendia
- Kerydin
- Konvomep
- Lampit
- Lanso-amox-clarithro
- Nexletol
- Nexlizet
- Niacor (niacin)
- Nisoldipine ER
- Nocdurna
- Norgesic tablet
- Norgesic Forte
- Noritate
- Noxafil 300 mg powdermix susp
- Nucynta
- Nuzyra
- Omnaris
- Omeclamox-pak combo pack
- Omeprazole-bicarb packet
- Opzelura
- Orapred ODT
- Oravig
- Orphenadrine-asa-caf
- Orphengesic forte
- Osmolex
- Osphena
- Otrexup
- Oxaprozin capsule
- Oxbryta
- Oxistat 1% cream & lotion
- Oxybutynin 2.5 mg tab
- Oxycodone-apap 2.5/300, 5/300, 7.5/300 10/300, 5/325/5ml soln
- Oxtellar XR
- Pandel cream
- Pennsaid pump
- Pegasys
- Pegintron
- Pegintron Readiclick
- Pentazocine-naloxone
- Phospholine iodide 0.125% drop
- Pioglitazone-glimepiride
- Podofilox 0.5% gel
- Pokonza packet
- Tetracycline tablet
- Texacort solution
- Thalitone 15mg tablet
- Tiagabine
- Tlando
- Tolcapone
- Tolsura
- Topiramate ER
- Tramadol ER TBMP
- Tramadol 25 mg tablet
- Tramadol solution
- Treximet tablet
- Triamterene
- Trientine hcl 500mg capsule
- Trokendi XR
- Ultravate lotion
- Ursodiol 200/400 cap
- Valtoco
- Varubi
- Venlafaxine besylate ER
- Veozah
- Verapamil PM
- Verdeso foam
- Verelan PM
- Viberzi
- Voquezna
- Vtama
- Vusion ointment
- Wynzora
- Xdemvy
- Xelstrym
- Xenleta
- Xerese cream
- Xhance spray
- Ximino
- Xphozah
- Xultophy 100-3.6
- Xuriden
- Zcort
- Zegerid packet
- Zembrace Symtouch
- Zelnorm
- Zenzedi except 5 and 10 mg

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- Dextroamphetamine oral tablet (2.5mg, 7.5mg, 15mg, 20mg, 30mg)
- Dextroamphatemine-amphetamine ER capsules (Mydayis generic)
- Diclofenac patch
- Diclofenac 25 mg, 35mg cap
- Diclofenac pot 25mg tablet
- Diclofenac powd pk
- Diclofenac 2% pump
- Diclofenac-misoprostol tablet
- Diflorasone cream and ointment
- Dihydroergotamine ampule
- Dipentum
- Doral
- Doxycycline hyclate 150mg
- Doxycycline mono 75mg and 150mg capsule
- Drizalma
- Duaklir Pressair
- Duobrii lotion
- Ledipasvir-sofosbuvir oral tablet
- Levamlodipine
- Levorphanol
- Lexette foam
- Licart 1.3% Patch
- Likmez suspension
- Lipofen (fenofibrate)
- Liqrev oral suspension
- Livtencity
- Lodoco
- Lofena
- Loreev XR
- Lorzone 375/750mg tablets
- Lybalvi
- Pramipexole ER tablet
- Prednisolone ODT
- Prednisolone tablet
- ProAir Respiclick/Digihaler
- Prolate tablet/solution
- Pylera
- Pyrimethamine
- Qbrexza
- Qdolo
- Qelbree
- Qnasl/Qnasl Children
- Quazepam
- Zepatier
- Zetonna
- Zileuton ER tablet
- Zipsor
- Zoryve cream and foam
- Ztlido
- Zyflo Filmtab

Authorization Criteria

1. Is the requested medication being used for an FDA approved or major compendia supported indication? (Verify regimen and dosing)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the request supported by current medical guidelines?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Did the member exhaust all formulary alternative products for treatment of the requested condition? (Provide supporting documentation)
    a. If yes, approve for lesser of 12 months OR standard treatment duration

Last Reviewed: 12/19/18, 5/15/19, 7/17/19, 9/18/19, 11/20/19, 3/18/20, 7/15/20, 9/16/20, 11/18/20, 1/20/21, 3/17/21, 7/21/21, 9/15/21, 11/17/21, 1/19/22, 3/16/22, 7/20/22, 9/21/22, 11/16/22, 3/17/23, 5/19/23, 7/21/23, 9/15/23, 11/19/23, 1/19/24, 3/15/24, 5/17/24

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b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

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Mesnex® (mesna)
Prior Authorization Guidelines

Affected Medication(s)

- Mesnex oral tablet

FDA Approved Indication(s)

- Prophylactic agent to reduce the incidence of ifosfamide-induced hemorrhagic cystitis

Dosing

- Two oral doses after bolus IV injection
- Oral doses of 40% of the ifosfamide dose at 2 and 6 hours after ifosfamide administration
- Repeat on each day ifosfamide is administered

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member currently receiving or planning to receive ifosfamide containing chemotherapy regimen? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member's body surface area and treatment plan provided for review of appropriate dosing? (Provide BSA and treatment plan for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by, or in consultation with, an oncologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Mesnex (mesna) [Prescribing Information]. Deerfield, IL: Baxter Healthcare Corporation.December 2021.

Last Reviewed: 11/21/18, 7/21/21, 3/17/23, 1/19/24
Effective Date: 1/1/19, 3/1/24

Methyltestosterone Products
Prior Authorization Guidelines

Affected Medication(s)

- Methyltestosterone oral capsule
- Methitest oral tablet

FDA Approved Indication(s)

- In males:
  - Primary hypogonadism (congenital or acquired)
  - Hypogonadotropic hypogonadism (congenital or acquired)
  - To stimulate puberty in carefully selected males with clearly delayed puberty
- In females:
  - May be used secondarily in women with advancing inoperable metastatic (skeletal) mammary cancer who are 1 to 5 years postmenopausal
  - Premenopausal women with breast cancer who have benefitted from oophorectomy and are considered to have a hormone-responsive tumor

Dosing

- In males: initial dosage of 10-50 mg daily
- In females: 50-200 mg daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same methyltestosterone product?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. What is the indication that the methyltestosterone product is being requested for?
   a. Male member with delayed puberty, continue to corresponding criteria
   b. Male member with hypogonadism, continue to corresponding criteria
   c. Female member with breast cancer, continue to corresponding criteria
   d. Gender affirming treatment, continue to corresponding criteria

Assigned male at birth member with delayed puberty

1. Is the member 14 years of age or older?
   a. If yes, continue to #2
   b. If no, clinical review required
2. Does the member have Tanner Staging of III or below?
   a. If yes, approve for 6 months unless otherwise specified

Last Reviewed: 12/19/18, 7/21/21, 7/20/22, 7/21/23, 11/17/23
Effective Date:1/1/19, 9/21/21, 9/1/22, 12/20/23

b. If no, clinical review required

Assigned male at birth member with hypogonadism

1. Does the member have documentation of TWO baseline total testosterone levels < 300 ng/dL that were taken in the morning on different days? (Provide documentation of total testosterone levels)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have at least TWO signs/symptoms of hypogonadism? (e.g. sleep disturbances, gynecomastia, decreased lean body mass, visceral obesity, hot flashes, changes in mood) (Provide supporting documentation of signs/symptoms)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there medical rationale why the member cannot use a generic injectable AND topical testosterone product? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Assigned female at birth member with Breast Cancer

1. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Gender Affirming Treatment

1. Does the member have a diagnosis of gender dysphoria by a qualified mental health professional? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have documentation of a positive clinical response to therapy defined by one of the following? (Provide supporting documentation)
    - For members with delayed puberty, documentation of progression into puberty AND with Tanner Staging of IV or less
    - For members with hypogonadism, documentation of normal testosterone levels
    - For members seeking gender affirming treatment, approve x12 months

    a. If yes, approve for 12 months unless otherwise specified

b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Methitest Prescribing Information. Global Pharmaceuticals, Division of Impax Laboratories Inc. May 2019.
2. Hembree WC, Cohen-Kettenis P, Delemarre-van de Waal HA, et al. Endocrine Society. Endocrine treatment of transsexual persons: an Endocrine Society clinical practice guideline. J Clin Endocrinol Metab. 2009 Sep;94(9):3132-3154.
3. Biro FM, Chan YM. Normal puberty. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com.
4. Olson-Kennedy J, Rosenthal SM, Hastings J, et al. UCSF Transgender Care & Treatment Guidelines: Health consideration for gender non-forming children and transgender adolescents. 2016 Jun. Available at: https://transcare.ucsf.edu/guidelines/youth

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Effective Date: 1/1/19, 9/1/21, 9/1/22, 12/20/23

Mircera® (methoxy polyethylene glycol-epoetin beta)
Prior Authorization Guidelines

Affected Medication(s)

- Mircera injection solution

FDA Approved Indication(s)

- Treatment of anemia associated with chronic kidney disease (CKD) in:
  - Adult patients on dialysis and patients not on dialysis
  - Pediatric patients 5 to 17 years of age on hemodialysis who are converting from another ESA after their hemoglobin level was stabilized with an ESA (IV route only, medical benefit)

Dosing

- Adults with CKD:
  - Recommended starting dose for patients who are not currently treated with an ESA: 0.6 mcg/kg administered as a single IV or SC injection once every two weeks
  - Refer to package insert for specific dose titration recommendations
  - Once hemoglobin stabilized, may be administered once monthly
- Pediatrics with CKD on hemodialysis:
  - Administer IV once every 4 weeks at a dose based on total weekly ESA dose at time of conversion (see table in package insert)

Initial Authorization Criteria

1. Is the request for continuation of Mircera (methoxy polyethylene glycol-epoetin beta) therapy in which the last dose was received less than 60 days ago? (Provide documentation of date of last administration)
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the requested medication planning to be administered through subcutaneous route? (For intravenous routes of administration request, contact medical benefit provider)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Is the member on hemodialysis? (Note: IV administration is preferred for members on hemodialysis due to lower immunogenicity)
   a. If yes, clinical review required
   b. If no, continue to #5
5. Does the member have a hemoglobin (Hb) level less than 10 g/dL and/or hematocrit (Hct) less than 30%? (Provide documentation of hemoglobin and hematocrit lab values taken within 30 days prior to planned administration)

Last Reviewed: 11/21/18, 7/21/21, 11/16/22, 11/17/23
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a. If yes, continue to #6
b. If no, clinical review required

6. Does the member have adequate iron stores as defined by serum ferritin ≥ 100 ng/mL and transferrin saturation (TSAT) ≥ 20%? (Provide documentation of serum ferritin and transferrin saturation levels taken within 30 days of planned administration)

    a. If yes, continue to #7
    b. If no, clinical review required

7. Have other causes of anemia (hemolysis, bleeding, vitamin deficiency, etc.) been ruled out? (Provide supporting documentation)

    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication) and the last dose was less than 60 days ago? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member had a positive clinical response to therapy as defined by a stabilization in hemoglobin and/or a reduction in the need for RBC transfusion? (Provide documentation of stabilization of hemoglobin or reduction in the need for RBC transfusions)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Does the member have a hemoglobin (Hb) level less than 11 g/dL and/or hematocrit (Hct) less than 33%? (Provide documentation of hemoglobin and hematocrit lab values taken within 30 days prior to planned administration)

    a. If yes, continue to #5
    b. If no, continue to #4

4. Does the member have a hemoglobin (Hb) level between 11 and 12 g/dL and will the Mircera dose be interrupted or reduced to the lowest dose sufficient to reduce the need for RBC transfusions? (Provide documentation of hemoglobin lab values taken within 30 days prior to planned administration and documentation of plan to interrupt therapy or reduce dose)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Does the member have adequate iron stores as defined by serum ferritin ≥ 100 ng/mL and transferrin saturation (TSAT) ≥ 20%? (Provide documentation of serum ferritin and transferrin saturation levels taken within 30 days of planned administration)

    a. If yes, approve for 1 year unless otherwise specified
    b. If no, clinical review required

Note:

Last Reviewed: 11/21/18, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 9/1/21

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Mircera [package insert]. San Francisco, CA; Genentech, Inc: August 2019.
2. Kidney Disease: Improving Global Outcomes (KDIGO) Anemia Work Group. KDIGO Clinical Practice Guideline for Anemia in Chronic Kidney Disease, Kidney Int, 2012; Suppl 2:279-335
3. National Kidney Foundation. K/DOQI Clinical Practice Guidelines for Anemia of Chronic Kidney Disease. http://kidneyfoundation.cachefly.net/professionals/KDOQI/guidelines_anemiaUP/guide1.htm. Accessed November 9, 2018

Last Reviewed: 11/21/18, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 9/1/21

Multiple Sclerosis Agents
Prior Authorization Guidelines

Affected Medication(s)

- Aubagio (teriflunomide)
- Avonex (interferon beta-1a)
- Bafiertam (monoethyl fumarate)
- Betaseron (interferon beta-1b)
- Copaxone (glatiramer acetate)
- Dimethyl fumarate
- Extavia (interferon beta-1b)
- Fingolimod
- Gilenya (fingolimod)
- Glatiramer, Glatopa
- Kesimpta (ofatumumab)
- Mavenclad (cladribine)
- Mayzent (siponimod)
- Plegridy (interferon beta-1a)
- Ponvory (ponesimod)
- Rebif (interferon beta-1a)
- Tascenso ODT (fingolimod)
- Tecfidera (dimethyl fumarate)
- Teriflunomide
- Vumerity (diroximel fumarate)

FDA Approved Indication(s)

- For treatment of adult patients with relapsing forms of multiple sclerosis, to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease:
  - Avonex, Aubagio, Bafiertam, Betaseron, Copaxone, Extavia, Kesimpta, Mayzent, Plegridy, Ponvory, Rebif, Tecfidera, Vumerity
- For treatment of patients 10 years of age and older with relapsing forms of multiple sclerosis, to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease:
  - Gilenya, Tascenso ODT
- For the treatment of adult patients relapsing forms of multiple sclerosis, to include relapsing-remitting disease and active secondary progressive disease
  - Mavenclad

Dosing

- **Avonex**: 30 mcg IM injection once a week
- **Aubagio**: 7 mg or 14 mg orally once daily
- **Bafiertam**: 190 mg orally twice daily
- **Betaseron**: 0.25 mg SC injection every other day
- **Extavia**: 0.25 mg SC injection every other day
- **Gilenya, Tascenso ODT**: 0.25 mg to 0.5 mg orally once daily
- **Kesimpta**: 20 mg SC injection once monthly
- **Copaxone**: 20 mg SC injection daily OR 40 mg SC injection three times weekly
- **Mavenclad**: Cumulative dosage of 3.5mg/kg orally and divided into 2 yearly treatment courses (1.75mg/kg per treatment course)

Last Reviewed: 10/3/18, 5/15/19, 11/20/19, 5/20/20, 9/16/20, 11/18/20, 1/19/22, 11/16/22, 5/19/23, 7/21/23, 5/17/24
Effective Date: 1/1/19, 7/1/19, 1/1/20, 7/1/20, 11/15/20, 12/15/20, 3/1/22, 1/1/23, 7/15/23, 9/15/23, 6/15/24

- **Mayzent**: 1mg to 2mg once daily (reference PI for specific dosing based on CYP2C9 genotype)
- **Plegridy**: 125 mcg subcutaneous injection every 14 days
- **Ponvory:** 20 mg orally once daily (refer to package insert for titration schedule)
- **Rebif**: 22 mcg or 44 mcg SC injection three times per week
- **Tecfidera**: 240 mg orally twice daily
- **Vumerity**: 462 mg orally twice daily

Initial Authorization Criteria

1. Is the request for continuation of a multiple sclerosis agent therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is an MRI result consistent with multiple sclerosis received? (Provide MRI for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member 18 years of age or older? (Note: Gilenya/Tascenso ODT are the only drugs approved for use in adolescents)
    a. If yes, continue to #5
    b. If no, continue to #7
5. Is the requested medication dimethyl fumarate, teriflunomide, fingolimod, or glatiramer acetate?
    a. If yes, continue to #7
    b. If no, continue to #6
6. Does the member have a history of inadequate response, intolerance or contraindication to at least one drug from each of the following groups? (Provide supporting documentation)
        - Glatiramer acetate
        - Dimethyl fumarate or Teriflunomide
        - Fingolimod
    a. If yes, continue to #7
    b. If no, clinical review required
7. Will the requested medication be used with other disease-modifying therapy for multiple sclerosis?
    a. If yes, clinical review required
    b. If no, continue to #8
8. Is the requested multiple sclerosis agent being prescribed by or in consultation with a neurologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 10/3/18, 5/15/19, 11/20/19, 5/20/20, 9/16/20, 11/18/20, 1/19/22, 11/16/22, 5/19/23, 7/21/23, 5/17/24
Effective Date: 1/1/19, 7/1/19, 1/1/20, 7/1/20, 11/15/20, 12/15/20, 3/1/22, 1/1/23, 7/15/23, 9/15/23, 6/15/24

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Will the requested medication be used with other disease-modifying therapy for multiple sclerosis?
    a. If yes, clinical review required
    b. If no, continue to #3
3. Is clinical documentation confirming responsiveness to therapy provided? (Provide documentation of disease stability)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the requested multiple sclerosis agent being prescribed by or in consultation with a neurologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Avonex (interferon beta-1a) [Prescribing Information]. Cambridge, MA: Biogen, Inc. July 2019.
2. Aubagio (teriflunomide) [Prescribing Information]. Cambridge, MA: Genzyme Corporation, A Sanofi Company. September 2019
3. Bafiertam (monoethyl fumarate) [Prescribing Information]. High Point NC, NJ: Banner Life Sciences LLC. April 2020.
4. Betaseron (interferon beta-1b) [Prescribing Information]. Whippany, NJ: Bayer HealthCare Pharmaceuticals Inc. August 2019.
5. Extavia (interferon beta-1b) [Prescribing Information]. East Hanover, NJ: Novartis Pharmaceuticals Corporation. October 2019.
6. Gilenya (fingolimod) [Prescribing Information]. East Hanover, NJ: Novartis Pharmaceuticals Corporation. August 2019.
7. Kesimpta (ofatumumab) [Prescribing Information]. East Hanover, NJ: Novartis Pharmaceuticals Corporation. August 2020.
8. Copaxone (glatiramer acetate) [Prescribing Information]. North Wales, PA: Teva Pharmaceuticals USA, Inc. July 2019.
9. Mavenclad (cladribine) [Prescribing Information]. Rockland, MA: EMD Serono, Inc. April 2019.
10. Mayzent (siponimod) tablets [package insert]. East Hanover, NJ: Novartis Pharmaceuticals Corp.; March 2019

Last Reviewed: 10/3/18, 5/15/19, 11/20/19, 5/20/20, 9/16/20, 11/18/20, 1/19/22, 11/16/22, 5/19/23, 7/21/23, 5/17/24
Effective Date: 1/1/19, 7/1/19, 1/1/20, 7/1/20, 11/15/20, 12/15/20, 3/1/22, 1/1/23, 7/15/23, 9/15/23, 6/15/24

11. Plegridy (peginterferon beta-1a) [Prescribing Information]. Cambridge, MA: Biogen, Inc. July 2019.
12. Ponvory (ponesimod) tablet [Prescribing Information]. Titusville, NJ: Janssen Pharmaceuticals. August 2023.
13. Rebif (interferon beta-1a) [Prescribing Information]. Rockland, MA: EMD Serono, Inc. October 2019.
14. Tascenso ODT (fingolimod) [Prescribing Information]. Cambridge, United Kingdom: Cycle Pharmaceuticals Ltd. December 2022.
15. Tecfidera (dimethyl fumarate) [Prescribing Information]. Cambridge, MA: Biogen, Inc. July 2019.
16. Vumerity (diroximel fumarate) delayed-release capsules, [package insert]. Cambridge, MA: Biogen, Inc.; 2019.
17. Rae-Grand A, Day GS, Marrie RA, et al. Practice guideline recommendations summary: Disease-modifying therapies for adults with multiple sclerosis. Neurology. 2018;90(17):777-788. Available at: http://n.neurology.org/content/neurology/90/17/777.full.pdf.

Myalept® (metreleptin)
Prior Authorization Guidelines

Affected Medication(s)

- Myalept powder for subcutaneous injection

FDA Approved Indication(s)

- As an adjunct to diet as replacement therapy to treat the complications of leptin deficiency in patients with congenital or acquired generalized lipodystrophy

Dosing

- Refer to package insert for recommended doses based on gender and weight

Initial Authorization Criteria

1. Is the request for continuation of Myalept (metreleptin) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have at least one of the following complications associated with leptin deficiency in patients with congenital or acquired generalized lipodystrophy? (Provide supporting documentation)
        - Diabetes mellitus
        - Hypertriglyceridemia
        - Increased fasting insulin levels
    a. If yes, continue to #4
    b. If no, clinical review required
4. Have baseline HbA1c, fasting glucose, and fasting triglycerides levels been received? (Provide labs for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have documentation of a trial with insufficient response to the following conventional therapies to treat each metabolic complication present?
        - Diabetes mellitus or insulin resistance
            - Diet modification
            - Optimized insulin therapy at maximally tolerated doses
        - Hypertriglyceridemia
            - Diet modification
            - Optimized therapy with at least two triglyceride lowering agents from different classes (i.e. fibrates, statins) at maximally tolerated doses

Last Reviewed: 11/7/18, 1/20/21, 7/20/22, 7/21/23
Effective Date: 1/1/19, 9/1/22

a. If yes, continue to #6
b. If no, clinical review required

6. Is the requested medication being used for any of the following conditions? (Provide clinical documentation for review)

    - Partial lipodystrophy
    - Nonalcoholic steatohepatitis (NASH)
    - HIV-related lipodystrophy
    - Metabolic disease without concurrent evidence of congenital or acquired generalized lipodystrophy

    a. If yes, clinical review required
    b. If no, continue to #7

7. Is the treatment being prescribed by, or in consultation with an endocrinologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member demonstrated a positive clinical response to therapy as defined by improvement in at least one metabolic parameter? (Provide documentation and updated HbA1c, fasting glucose, or fasting triglycerides level for review)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with an endocrinologist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Myalept. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed September 27, 2018.
2. Myalept (metreleptin) [Prescribing Information]. Cambridge, MA: Aegerion Pharmaceuticals, Inc. September 2021.

Last Reviewed: 11/7/18, 1/20/21, 7/20/22, 7/21/23
Effective Date: 1/1/19, 9/1/22

3. Mantzoros, C. Lipodystrophic sundromes. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed October 2018.

Last Reviewed: 11/7/18, 1/20/21, 7/20/22, 7/21/23
Effective Date: 1/1/19, 9/1/22

Myeloid Growth Factors
Prior Authorization Guidelines

Affected Medication(s)

- Fulphila (pegfilgrastim-jmdb) subcutaneous solution
- Fylnetra (pegfilgrastim-pbbk) subcutaneous solution
- Granix (tbo-filgrastim) injection solution
- Neulasta (pegfilgrastim) subcutaneous solution
- Neupogen (filgrastim) injection solution
- Nivestym (filgrastim-aafi) injection solution
- Nyvepria (pegfilgrastim-apgf) subcutaneous solution
- Releuko (filgrastim-ayow) injection solution
- Stimufend (pegfilgrastim-fbgk) subcutaneous solution
- Udenyca (pegfilgrastim-cbqv) subcutaneous solution
- Zarxio (filgrastim-sndz) injection solution
- Ziextenzo (pegfilgrastim-bmez) subcutaneous solution

FDA Approved Indication(s)

- To reduce the duration of severe neutropenia in adult and pediatric patients 1 month and older with non-myeloid malignancies receiving myelosuppressive anticancer drugs associated with a clinically significant incidence of febrile neutropenia
  - Granix
- To decrease the incidence of infection, as manifested by febrile neutropenia, in patients with non-myeloid malignancies receiving myelosuppressive anti-cancer drugs associated with a clinically significant incidence of febrile neutropenia
  - Fulphila, Fylnetra, Neulasta, Neupogen, Nivestym, Releuko, Stimufend, Udenyca, Ziextenzo, Zarxio
- For reducing the time to neutrophil recovery and the duration of fever, following induction or consolidation chemotherapy treatment of patients with acute myeloid leukemia (AML)
  - Neupogen, Nivestym, Releuko, Zarxio
- To reduce the duration of neutropenia and neutropenia-related clinical sequelae, e.g., febrile neutropenia, in patients with non-myeloid malignancies undergoing myeloablative chemotherapy followed by bone marrow transplantation
  - Neupogen, Nivestym, Releuko, Zarxio
- For the mobilization of autologous hematopoietic progenitor cells into the peripheral blood for collection by leukapheresis
  - Neupogen, Nivestym, Zarxio
- For chronic administration to reduce the incidence and duration of sequelae of neutropenia (e.g., fever, infections, oropharyngeal ulcers) in symptomatic patients with congenital neutropenia, cyclic neutropenia, or idiopathic neutropenia
  - Neupogen, Nivestym, Releuko, Zarxio
- To increase survival in patients acutely exposed to myelosuppressive doses of radiation
  - Neulasta, Neupogen, Stimufend, Udenyca

Dosing

- Refer to package insert for recommended dosing for corresponding diagnosis

Authorization Criteria

Last Reviewed: 11/7/18, 7/17/19, 3/17/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 9/1/19, 5/1/21, 3/1/24

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the medication being prescribed by or in consultation with an oncologist/hematologist or an appropriate specialist?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the submitted diagnosis for hematopoietic radiation injury syndrome?
    a. If yes, continue to #4
    b. If no, continue to #6
4. Is Neupogen, Stimufend, or Udenyca being requested?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, continue to #5
5. Does the member have a documented clinical rationale for avoiding Neupogen (filgrastim), Stimufend (pegfilgrastim-fbgk), AND Udenyca (pegfilgrastim-cbqv)? (Provide supporting documentation)
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required
6. Is the request for one of the following preferred agents: Nivestym (filgrastim-aafi), Releuko (filgrastim-ayow), Fulphila (pegfilgrastim-jmdb), or Fylnetra (pegfilgrastim-pbbk)?
    a. If yes, continue to #11
    b. If no, continue to #7
7. Is the requested drug one of the following: Granix (tbo-filgrastim), Neupogen (filgrastim), or Zarxio (filgrastim-sndz)?
    a. If yes, continue to #8
    b. If no, continue to #9
8. Does the member have a documented clinical rationale for avoiding Releuko (filgrastim-ayow) AND Nivestym (filgrastim-aafi)? (Provide supporting documentation)
    a. If yes, continue to #10
    b. If no, clinical review required
9. Is the requested drug one of the following: Neulasta (pegfilgrastim), Nyvepria (pegfilgrastim-apgf), Stimufend (pegfilgrastim-fbgk), Udenyca (pegfilgrastim-cbqv), or Ziextenzo (pegfilgrastim-bmez)?
    a. If yes, continue to #10

b. If no, clinical review required

10. Does the member have a documented clinical rationale for avoiding Fulphila (pegfilgrastim-jmdb) AND Fylnetra (pegfilgrastim-pbbk)? (Provide supporting documentation)

    a. If yes, continue to #11
    b. If no, clinical review required

11. What is the medication being requested for? (Provide clinical documentation to support diagnosis)

    a. Bone Marrow Transplantation, approve for 4 months unless otherwise specified
    b. Peripheral Blood Progenitor cell (PBPC) mobilization, approve for 4 months unless otherwise specified
    c. Acute myeloid leukemia (AML) patient undergoing induction or consolidation chemotherapy, approve for 4 months unless otherwise specified
    d. Prophylaxis of febrile neutropenia in patients with non-myeloid malignancy, continue to corresponding criteria
    e. Treatment of chemotherapy-induced febrile neutropenia, continue to corresponding criteria
    f. Severe Chronic Neutropenia, continue to corresponding criteria
    g. Other indication, continue to corresponding criteria

<u>Prophylaxis of febrile neutropenia in patients with non-myeloid malignancy</u>

1. Does the planned chemotherapy regimen have a high risk (greater than 20% risk) of febrile neutropenia?

    a. If yes, continue to #2
    b. If no, continue to #4

2. Is the planned chemotherapy regimen for curative treatment intent?

    a. If, yes approve for 4 months unless otherwise specified
    b. If no, continue to #3

3. Is clinical rationale provided to support the use of a high-risk regimen in the palliative setting? (Provide supporting documentation)

    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

4. Does the member have at least one of the following risk factors for febrile neutropenia? (Provide supporting documentation)

    - 65 years or older and receiving full chemotherapy dose intensity
    - Prior chemotherapy or radiotherapy
    - Persistent neutropenia
    - Tumor involvement in the bone marrow
    - Recent surgery and/or open wounds
    - Renal dysfunction (creatinine clearance <50)
    - Liver dysfunction (bilirubin >2.0)

    a. If yes, continue to #5
    b. If no, continue to #6

5. Does the planned chemotherapy regimen have an intermediate risk (10 to 20% risk) of febrile neutropenia?
    a. If yes, continue to #7
    b. If no, clinical review required
6. Is the member continuing the same chemotherapy regimen which induced a dose-limiting neutropenic event on a previous cycle? (Provide supporting clinical documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the planned chemotherapy regimen for curative treatment intent?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Treatment of chemotherapy-induced febrile neutropenia

1. Has the member received a prophylaxis regimen for febrile neutropenia with a granulocyte colony stimulating factor on the current chemotherapy cycle?
    a. If yes, clinical review required
    b. If no, continue to #2
2. Does the member have an absolute neutrophil count (ANC) <500/mm^3? (Provide documentation of ANC lab value)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have one or more of the following risk factors for developing infection-related complications? (Provide supporting documentation)
    - Sepsis Syndrome
    - Age >65
    - Absolute neutrophil count [ANC] <100/mcL
    - Duration of neutropenia expected to be greater than 10 days
    - Pneumonia or other clinically documented infections
    - Invasive fungal infection
    - Hospitalization at the time of fever
    - Prior episode of febrile neutropenia

    a. If yes, approve for 1 month unless otherwise specified
    b. If no, clinical review required

Severe chronic neutropenia

1. Does the member have an absolute neutrophil count (ANC) <500/mm^3? (Provide CBC with differential for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a diagnosis of one of the following? (Provide documentation of diagnosis)
    - Congenital neutropenia

- Cyclic neutropenia
- Idiopathic neutropenia

a. If yes, continue to #3
b. If no, clinical review required

3. Does the member have neutropenia symptoms (i.e. fever, infections, etc.)? (Provide documentation of symptoms)

a. If yes, approve for 4 months unless otherwise specified
b. If no, clinical review required

Other Indication

1. Is the requested use supported by major compendia? (Examples: Micromedex, Clinical Pharmacology, NCCN, etc.) (Provide supporting documentation confirming diagnosis)

a. If yes, continue to #2
b. If no, clinical review required

2. Has the member tried and had an inadequate response OR does the member have a contraindication to ALL standard treatment options for the requested indication (Provide all prior treatment history, contraindication if appropriate, and treatment plan)

a. If yes, approve for 4 months unless otherwise specified
b. If no, clinical review required

Note:

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References:

1. Neupogen [package insert]. Thousand Oaks, CA; Amgen Inc; June 2016. Accessed July 2019.
2. Zarxio [package insert]. Princeton, NJ; Sandoz Inc; April 2018. Accessed July 2019.
3. Granix [package insert]. North Wales, PA; Teva Pharmaceuticals USA, Inc.; April 2019. Accessed July 2019.
4. Neulasta [package insert]. Thousand Oaks, CA; Amgen Inc; April 2019. Accessed July 2019.
5. Fulphila [package insert]. Rockford, IL; Mylan Institutional LLC; May 2019. Accessed July 2019.
6. Nivestym [package insert]. Lake Forest, IL; Hospira, Inc., a Pfizer Company; March 2019. Accessed July 2019.
7. Udenyca [package insert]. Redwood City, CA; Coherus BioSciences, Inc; February 2019. Accessed July 2019.
8. Fylnetra [package insert]. Piscataway, NJ; Kashiv BioSciences, Inc; May 2022. Accessed December 2023.
9. Stimufend [package insert]. Lake Geneva, IL; Fresenius Kabi; October 2023. Accessed December 2023.
10. Releuko [package insert]. Piscataway, NJ; Kashiv BioSciences; August 2023. Accessed December 2023.

Last Reviewed: 11/7/18, 7/17/19, 3/17/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 9/1/19, 5/1/21, 3/1/24

11. Neupogen. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed September 2018.
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13. pegfilgrastim. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed July 2019.
14. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) filgrastim. National Comprehensive Cancer Network, 2018. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed March 2018.
15. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) Hematopoietic Growth Factors. Version 2.2019. National Comprehensive Cancer Network, 2019. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc." To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed July 2019.
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17. Wisconsin Physicians Service Insurance Corporation. Local Coverage Determination (LCD): Human Granulocyte/Macrophage Colony Stimulating Factors (L34699). Centers for Medicare & Medicaid Services, Inc. Updated on 1/23/2018 with effective date 02/1/2018. Accessed March 2018.
18. First Coast Service Options, Inc. Local Coverage Determination (LCD): G-CSF (Neupogen®, Granix™, Zarxio™) (L34002). Centers for Medicare & Medicaid Services, Inc. Updated on 6/10/2016 with effective date 7/5/2016. Accessed March 2018.
19. National Government Services, Inc. Local Coverage Article: Filgrastim, Pegfilgrastim, Tbofilgrastim, Filgrastim-sndz (e.g., Neupogen®, Neulasta™, Granix™, Zarxio™) - Related to LCD L33394 (A52408). Centers for Medicare & Medicaid Services, Inc. Updated on 9/23/2016 with effective date 10/1/2016. Accessed March 2018.
20. Palmetto GBA. Local Coverage Determination: White Cell Colony Stimulating Factors (L37176).Centers for Medicare & Medicaid Services, Inc. Updated on 12/7/2017 with effective date 2/26/2018. Accessed March 2018.

Mytesi® (crofelemer)
Prior Authorization Guidelines

Affected Medication(s)

- Mytesi delayed-release oral tablet

FDA Approved Indication(s)

- For symptomatic relief of non-infectious diarrhea in adult patients with HIV/AIDS on anti-retroviral therapy

Dosing

- 125 mg taken orally twice daily

Initial Authorization Criteria

1. Is the request for continuation of Mytesi (crofelemer) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member currently on anti-retroviral therapy and has had non-infectious diarrhea for longer than one month? (Provide list of relevant current medications and supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a trial with insufficient response, intolerance, or contraindication to one of the following alternative anti-diarrheal medications: loperamide, bismuth subsalicylate, or diphenoxylate/atropine? (Provide relevant past medication history and/or intolerance/contraindication)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required

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Effective Date: 1/1/19, 3/1/24

2. Does the member demonstrate a positive clinical response to therapy as defined as a decrease in the frequency and/or severity of diarrhea? (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Mytesi Prescribing Information. San Francisco, CA: Napo Pharmaceuticals, Inc. November 2020. Available at: www.mytesi.com.
2. Mangel AW, Chaturvedi P. Evaluation of crofelemer in the treatment of diarrhea-predominant irritable bowel syndrome patients. Digestion. 2008; 78(4): 180-186.
3. MacArthur, Rodger D., et al. "Efficacy and safety of crofelemer for noninfectious diarrhea in HIV-seropositive individuals (ADVENT trial): a randomized, double-blind, placebo-controlled, two-stage study." HIV clinical trials 14.6 (2013): 261-273.
4. Mytesi. Micromedex® Healthcare Series [Internet database]. Greenwood Village, Colo: Thomson Healthcare. Updated periodically.

Last Reviewed: 11/7/18, 11/18/20, 1/19/22, 1/20/23, 1/19/24
Effective Date: 1/1/19, 3/1/24

Northera® (droxidopa)
Prior Authorization Guidelines

Affected Medication(s)

- Northera oral capsule
- Droxidopa oral capsule

FDA Approved Indication(s)

- Treatment of orthostatic dizziness, lightheadedness, or the “feeling that you are about to black out” in adult patients with symptomatic neurogenic orthostatic hypotension (nOH) caused by primary autonomic failure (Parkinson's disease [PD], multiple system atrophy, and pure autonomic failure), dopamine beta-hydroxylase deficiency, and non-diabetic autonomic neuropathy

Dosing

- Starting dose: 100 mg, taken orally three times daily
- Titrate to symptomatic response, in increments of 100 mg three times daily every 24 to 48 hours up to a maximum dose of 600 mg three times daily

Initial Authorization Criteria

1. Is the request for continuation of Northera (droxidopa) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have documentation of primary autonomic failure (by Parkinson's disease [PD], multiple system atrophy, and pure autonomic failure), dopamine beta-hydroxylase deficiency, or non-diabetic autonomic neuropathy? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member 18 years of age or older?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is there documentation that the member has tried at least TWO non-pharmacological interventions to treat the conditions as outlined below? (Provide supporting documentation of non-pharmacological interventions tried)
    i. Elevation of the bed by 5-20 degrees
    ii. Use of compression stockings
    iii. Increased salt and water intake
    iv. Avoidance of precipitating factors including arising too quickly, alcohol consumption, hot baths, or hot environments

Last Reviewed: 11/21/18, 11/18/20, 3/17/21, 5/18/22, 7/21/23
Effective Date: 1/1/19, 12/15/20, 5/1/21, 7/15/22

v. Discontinuation of drugs which can cause orthostatic hypotension (i.e. diuretics, antihypertensive medications, nitrates, alpha-adrenergic antagonists, etc.)

a. If yes, continue to #6
b. If no, clinical review required

6. Does the member have a sufficient trial with inadequate response, intolerance, or contraindication to both midodrine AND fludrocortisone? (Provide supporting documentation)

a. If yes, continue to #7
b. If no, clinical review required

7. Is the treatment being prescribed by, or in consultation with, a neurologist, cardiologist, or nephrologist?

a. If yes, approve for 2 weeks
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

a. If yes, continue to #2
b. If no, clinical review required

2. Did the member have a positive clinical response to therapy as defined as less frequent episodes of orthostatic dizziness, lightheadedness, or the "feeling that you are about to black out"? (Provide supporting documentation of less frequent symptomatic episodes)

a. If yes, continue to #3
b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with, a neurologist, cardiologist, or nephrologist?

a. If yes, continue to #4
b. If no, clinical review required

4. Is clinical rationale provided for continued use beyond 2 weeks? (Provide supporting documentation)

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

Note:

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References:

1. Northera [prescribing information]. Deerfield, IL: Lundbeck. July 2019.

2. Kaufman H, Freeman R, Biagginoi I, et. al. Droxidopa for Neurogenic Orthostatic Hypotension. A Randomized, Placebo-Controlled, Phase 3Trial. Neurology. 2014;83:1-8.

Last Reviewed: 11/21/18, 11/18/20, 3/17/21, 5/18/22, 7/21/23
Effective Date: 1/1/19, 12/15/20, 5/1/21, 7/15/22

3. Kaufmann H, Malamut R, Norcliffe-Kaufmann L, Rosa K, Freeman R. The Orthostatic Hypotension Questionnaire (OHQ): validation of a novel symptom assessment scale. Clin Auton Res. 2012;22(2):79-80.

4. Freeman R. Clinical Practice. Neurogenic orthostatic hypotension. N Engl J Med. 2008;358(6);615-624.

Last Reviewed: 11/21/18, 11/18/20, 3/17/21, 5/18/22, 7/21/23
Effective Date: 1/1/19, 12/15/20, 5/1/21, 7/15/22

Nourianz® (istradefylline)
Prior Authorization Guidelines

Affected Medication(s)

- Nourianz oral tablet

FDA Approved Indication(s)

- For the intermittent treatment of "OFF" episodes in patients with Parkinson's disease treated with carbidopa/levodopa

Dosing

- 20 mg once daily
- Maximum dose is 40mg once daily

Initial Authorization Criteria

1. Is the request for continuation of Nourianz (istradefylline) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a diagnosis of advanced Parkinson's disease? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member 18 years of age or older?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member experience "OFF" episodes despite adjustment in carbidopa/levodopa dosing? (Examples include increasing levodopa dose or decreasing dose and increasing frequency) (Provide supporting documentation of dose adjustments trialed and response)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a previous trial with inadequate response or intolerance to at least 2 different classes of medications to help reduce "OFF" time as listed below OR a contraindication to all 3 classes? (Provide supporting documentation of agents trialed with inadequate response, intolerance, or contraindication)
    - Dopamine agonist (Examples include: ropinirole, rotigotine)
    - Catechol-O-methyl transferase (COMT) inhibitor (Examples include: entacapone, tolcapone)

Last Reviewed: 9/18/19, 5/1/20, 11/17/21, 3/17/23, 3/15/24
Effective Date: 10/15/19, 7/1/20, 1/1/22, 4/15/24

- Monoamine oxidase type B inhibitor (MAO-B) (Examples include: rasagiline, safinamide, selegiline)

a. If yes, continue to #7
b. If no, clinical review required

7. Is the treatment being prescribed by, or in consultation with, a neurologist?

a. If yes, approve for 6 months
b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?

a. If yes, continue to #2
b. If no, clinical review required

2. Were updated chart notes (within the past 12 months) with documentation of a positive response to therapy defined as a decrease in frequency of "OFF" episodes received? (Provide supporting documentation)

a. If yes, continue to #3
b. If no, clinical review required

3. Is the treatment being prescribed by or in consultation with a neurologist?

a. If yes, approve for 12 months reauthorization
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Nourianz [istradefylline] tablets. Kyowa Kirin Inc., Bedminster, NJ; November 2020
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 25 July. 2019].
3. Fox, Susan H., et al. "International Parkinson and movement disorder society evidence-based medicine review: Update on treatments for the motor symptoms of Parkinson's disease." Movement Disorders 33.8 (2018): 1248-1266.

Last Reviewed: 9/18/19, 5/20/20, 11/17/21, 3/17/23, 3/15/24
Effective Date: 10/15/19, 7/1/20, 1/1/22, 4/15/24

Nuplazid® (pimavanserin tartrate)
Prior Authorization Guidelines

Affected Medication(s)

- Nuplazid oral capsule
- Nuplazid oral tablet

FDA Approved Indication(s)

- Treatment of hallucinations and delusions associated with Parkinson's disease psychosis

Dosing

- 34 mg orally once daily

Initial Authorization Criteria

1. Is the request for continuation of Nuplazid (pimavanserin tartrate) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Have the member's hallucinations or delusions developed after the onset of Parkinson's disease? (Provide documentation of onset of hallucinations or delusions in relation to onset of Parkinson's Disease)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Was an attempt made to reduce doses or discontinue medications that may contribute to, or cause, hallucinations and/or delusions or has rationale for no dose reduction/discontinuation been received? (Examples of medications include: dopamine agonists, amantadine, monoamine oxidase B inhibitors, and anticholinergics) (Provide documentation of attempted dose reduction, discontinuation, or rationale for avoidance)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2

Last Reviewed: 11/21/18, 01/20/21, 5/19/21, 7/20/22, 7/21/23
Effective Date: 1/1/19

b. If no, clinical review required

2. Has the member experienced a positive clinical response to therapy as defined by a reduction in the frequency and/or severity of hallucinations or delusions? (Provide supporting documentation)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the treatment being prescribed by, or in consultation with, a neurologist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Nuplazid [prescribing information]. San Diego, CA: Acadia Pharmaceuticals, Inc; November 2020.
2. Cummings J, Isaacson S, Mills R, et al. Pimavanserin for patients with Parkinson's disease psychosis: a randomized, placebo-controlled phase 3 trial. The Lancet. 2014 Feb 8; 383:533-540.
3. Connolly B, Fox SH. Treatment of cognitive, psychiatric, and affective disorders associated with Parkinson's disease. Neurotherapeutics 2014; 11:78.
4. Rabey JM, Treves TA, Neufeld MY, et al. Low-dose clozapine in the treatment of levodopa-induced mental disturbances in Parkinson's disease. Neurology 1995; 45:432.
5. Fernandez HH, Friedman JH, Jacques C, Rosenfeld M. Quetiapine for the treatment of drug-induced psychosis in Parkinson's disease. Mov Disord 1999; 14:484.
6. Parkinson Study Group. Low-dose clozapine for the treatment of drug-induced psychosis in Parkinson's disease. N Engl J Med 1999; 340:757.

Ocaliva® (obeticholic acid)
Prior Authorization Guidelines

Affected Medication(s)

- Ocaliva oral tablet

FDA Approved Indication(s)

- For the treatment of primary biliary cholangitis (PBC) in combination with ursodeoxycholic acid (UDCA/ursodiol) in adults with an inadequate response to UDCA/ursodiol, or as monotherapy in adults unable to tolerate UDCA

Dosing

- Refer to package insert for specific dosing and titration based on Child-Pugh Class

Initial Authorization Criteria

1. Is the request for continuation of Ocaliva (obeticholic acid) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a trial with insufficient response defined as greater than ALP 1.67x ULN or total bilirubin greater than 1x ULN but less than 2x ULN with at least 12 months of ursodiol at a dose of ≥ 13 mg/kg/day?(Provide relevant medication history and response to therapy)
    a. If yes, continue to #6
    b. If no, continue to #5
5. Does the member have an intolerance or contraindication to ursodiol? (Provide supporting documentation of intolerance or contraindication)
    a. If yes, continue to #7
    b. If no, clinical review required
6. Will the member be using Ocaliva in combination with ursodiol? (Provide treatment plan)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the requested dosing appropriate for the members Child-Pugh class? (Provide Child Pugh class for review)
    a. If yes, continue to #8
    b. If no, clinical review required

Last Reviewed: 11/7/18, 1/20/21, 11/16/22, 11/17/23
Effective Date: 1/1/19

8. Is the treatment being prescribed by, or in consultation with, a hepatologist or gastrointestinal (GI) specialist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member having a positive clinical response to therapy as defined by a maintained reduction in alkaline phosphate (ALP) level from pretreatment level? (Provide updated ALP levels for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by, or in consultation with, a hepatologist or gastrointestinal (GI) specialist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Ocaliva Prescribing Information. New York, NY: Intercept Pharmaceuticals, Inc.; May 2022. Available at https://ocaliva.com/.
2. Lindor, KD, Gershwin ME, Poupon R et al. AASLD Practice Guidelines: Primary biliary cirrhosis. Hepatology. 2009; 50(1): 291-308.
3. European Association for the Study of the Liver (EASL). EASL clinical practice guidelines: the diagnosis and management of patients with primary biliary cholangitis. J Hepatology. 2017;67:145-72.
4. Lindor, Keith D., et al. "Primary biliary cholangitis: 2018 practice guidance from the American Association for the Study of Liver Diseases." Hepatology 69.1 (2019): 394-419.

Last Reviewed: 11/7/18, 1/20/21, 11/16/22, 11/17/23
Effective Date: 1/1/19

Octreotide Agents
Prior Authorization Guidelines

Affected Medication(s)

- Sandostatin injection solution
- Sandostatin LAR Depot intramuscular powder for suspension
- Octreotide acetate injection solution
- Mycapssa oral capsule

FDA Approved Indication(s)

- Sandostatin/octreotide injection:
  - To reduce blood levels of growth hormone and IGF-I (somatomedin C) in acromegaly patients who have had inadequate response to or cannot be treated with surgical resection, pituitary irradiation, and bromocriptine mesylate at maximally tolerated doses
  - For the symptomatic treatment of patients with metastatic carcinoid tumors where it suppresses or inhibits the severe diarrhea and flushing episodes associated with the disease
  - For the treatment of the profuse watery diarrhea associated with VIP-secreting tumors. Sandostatin studies were not designed to show an effect on the size, rate of growth or development of metastases
- Mycapssa
  - Long-term maintenance treatment in acromegaly patients who have responded to and tolerated treatment with octreotide or lanreotide

Dosing

- Refer to corresponding package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of Sandostatin (octreotide), Sandostatin LAR Depot (octreotide), octreotide acetate, or Mycapssa (octreotide) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member 18 years of age or older?
   a. If yes, continue to #4
   b. If no, clinical review required
4. What diagnosis is the medication being requested for? (Provide supporting documentation)
   a. Carcinoid tumors/Neuroendocrine tumors (e.g. GI tract, lung, thymus, pancreas, adrenal), continue to corresponding criteria

Last Reviewed: 12/19/18, 1/20/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 3/15/23

b. Diarrhea associated with vasoactive intestinal peptide tumors (VIPomas) [pancreatic, neuroendocrine (islet cell) tumor, insulinoma, glucagonoma, somatostatinoma, and gastrinoma], continue to corresponding criteria
c. Acromegaly, continue to corresponding criteria
d. Other oncology indication, continue to corresponding criteria
e. Other non-oncology indication(s), continue to corresponding criteria

Carcinoid tumors/Neuroendocrine tumors (e.g. GI tract, lung, thymus, pancreas, adrenal)

1. Does the member meet one or more of the following conditions? (Provide supporting documentation)

    - Severe diarrhea/flushing episodes (carcinoid syndrome)
    - Requested use is to treat symptoms related to hormone hypersecretion in pancreatic tumors
    - Requested use is for primary treatment of unresected primary gastrinoma
    - Requested use is for management of locoregional advanced or metastatic disease of the
    - bronchopulmonary, thymic, gastrointestinal tract
    - Requested use is for tumor control of unresectable and/or metastatic tumors of the pancreas
    - Requested use is for management of pheochromocytoma or paraganglioma with unresectable or distant metastases

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Diarrhea associated with vasoactive intestinal peptide tumors (VIPomas) [pancreatic, neuroendocrine (islet cell) tumor, insulinoma, glucagonoma, somatostatinoma, and gastrinoma]

1. Does the member have profuse watery diarrhea? (Provide supporting documentation)

    a. If yes, approve for 6 months
    b. If no, clinical review required

Acromegaly

1. Was baseline growth hormone (GH) and IGF-1 blood levels received? (Provide GH and IGF-1 serum levels for review)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Did the member have an inadequate response to surgery and/or radiotherapy or is the member not a candidate for surgery and/or radiotherapy? (Provide documentation of inadequate response or rationale why member is not a candidate)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the request for Mycapssa (octreotide)?

    a. If yes, continue to #4
    b. If no, approve for 6 months

4. Does the member have trial with inadequate response, intolerance, or contraindication to both Sandostatin LAR (octreotide) injection and Somatuline Depot (lanreotide) injection? (Provide supporting documentation)

    a. If yes, approve for 6 months
    b. If no, clinical review required

Oncology Indication

1. Is the medication being requested for an indication supported by National Comprehensive Cancer Network (NCCN) with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Other Non-Oncology Indication

1. Has the member tried and had an inadequate response OR does the member have a contraindication to all standard treatment options for the requested indication? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by or in consultation with an appropriate specialist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Does the member continue to meet the initial authorization criteria? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there documentation of any of the following side effects from the medication? (Examples of unacceptable toxicity include the following: biliary tract abnormalities, hypothyroidism, goiter, sinus bradycardia, cardiac arrhythmias, cardiac conduction abnormalities, pancreatitis, etc.) (Provide supporting documentation of absence of unacceptable toxicities)
    a. If yes, clinical review required
    b. If no, continue to #3
3. Does the member have a positive clinical response to therapy defined by one of the following? (Provide supporting documentation)
    - Improvement in symptoms including reduction in symptomatic episodes (such as diarrhea, rapid gastric dumping, flushing, bleeding, etc.)
    - Stabilization of glucose levels

Last Reviewed: 12/19/18, 1/20/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 3/15/23

- Decrease or stabilization in tumor size
- For acromegaly only: Reduction of growth hormone (GH) and/or IGF-I blood levels from baseline
- For neuroendocrine tumors of the pancreas only: Member has had disease progression and therapy will be continued in member with functional tumors in combination with systemic therapy

a. If yes, continue to #4
b. If no, clinical review required

4. Is the treatment being prescribed by or in consultation with an appropriate specialist?

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Sandostatin LAR [package insert]. East Hanover, NJ; Novartis Pharmaceuticals Corporation; March 2021.
2. MYCAPSSA (octreotide) delayed-release capsules, [package insert]. Needham, MA: Chiasma, Inc.; 2022.
3. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 30 Nov. 2020].
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Last Reviewed: 12/19/18, 1/20/21, 1/20/23, 1/19/24
Effective Date: 1/1/19, 3/15/23

Ogsiveo (nirogacestat hydrobromide)
Prior Authorization Guidelines

Affected Medication(s)

- Ogsiveo oral tablet

Indication(s)

- Treatment of adult patients with progressing desmoid tumors who require systemic treatment

Dosing:

- 150 mg orally twice daily until disease progression or unacceptable toxicity

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same anti-cancer medication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the medication being requested to be used for an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #4
    b. If no, continue to #3
3. Is the medication being requested being used for an indication supported by the National Comprehensive Cancer Network (NCCN) with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a previous trial with inadequate response, intolerance or contraindication to at least TWO of the following: imatinib, pazopanib, or sorafenib? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 1/19/24
Effective Date: 3/1/24

Reauthorization Criteria

1. Is the documented indication approved by the FDA or supported by the NCCN recommendation with an evidence level of 2A or higher? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there clinical documentation confirming disease responsiveness to therapy provided? (Examples include reduction in tumor size, objective response, delay in progression, partial response, etc.) (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. OGSIVEO (nirogacestat) tablets, [package insert]. Stamford, CT: SpringWorks Therapeutics, Inc.; 2023.
2. Drugs@FDA: FDA Approved Drug Products. 2023. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 11 Dec. 2023].
3. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Soft Tissue Sarcoma. Version 3.2023 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf. Accessed December 14, 2023.
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Last Reviewed: 1/19/24
Effective Date: 3/1/24

Oncology Agents
Prior Authorization Guidelines

Affected Medication(s)

- abiraterone oral tablet
- Actimmune (interferon gamma-1b subcutaneous solution)
- Akeega (abiraterone-niraparib oral tablet)
- Afinitor (everolimus oral tablet)
- Afinitor Disperz (everolimus tablet for suspension)
- Alecensa (alectinib oral capsule)
- Alunbrig (brigatinib oral tablet)
- Augtyro (repotrectinib oral capsule)
- Ayvakit (avapritinib oral tablet)
- Balversa (erdafitinib oral tablet)
- bexarotene 1% gel
- bexarotene capsule
- Bosulif (bosutinib oral capsule and tablet)
- Braftovi (encorafenib capsule)
- Brukinsa (zanubrutinib oral capsule)
- Cabometyx (cabozantinib oral tablet)
- Calquence (acalabrutinib oral tablet)
- Caprelsa (vandetanib oral tablet)
- Cometriq (cabozantinib oral capsule)
- Copiktra (duvelisib capsule)
- Cotellic (cobimetinib tablet)
- Daurismo (glasdegib tablet)
- Demser (metyrosine capsule)
- Emcyt (estramustine phosphate sodium capsule)
- Erivedge (vismodegib capsule)
- Erleada (apalutamide tablet)
- erlotinib tablet
- etoposide capsule
- everolimus tablet and tablet for suspension
- Exkivity (mobocertinib capsule)
- Fareston (toremifene citrate tablet)
- Fruzaqla (fruquintinib oral capsule)
- Fotivda (tivozanib capsule)
- Gavreto (pralsetinib capsule)
- gefitinib tablet
- Gilotrif (afatinib dimaleate tablet)
- Myleran (busulfan tablet)
- Nerlynx (neratinib tablet)
- Nexavar (sorafenib tosylate tablet)
- Nilandron (nilutamide tablet)
- nilutamide tablet
- Ninlaro (ixazomib capsule)
- Nubeqa (darolutamide tablet)
- Odomzo (sonidegib capsule)
- Ojjaara (momelotinib tablet)
- Orserdu (elacestrant)
- pazopanib tablet
- Pemazyre (pemigatinib tablet)
- Piqray (alpelisib tablet)
- Pomalyst (pomalidomide capsule)
- Qinlock (ripretinib tablet)
- Retevmo (selpercatinib capsule)
- Revlimid (lenalidomide capsule)
- Rezlidhia (olutasidenib tablet)
- Rozlytrek (entrectinib capsule and oral pellets)
- Rubraca (rucaparib tablet)
- Rydapt (midostaurin capsule)
- sorafenib tablet
- Sprycel (dasatinib tablet)
- Stivarga (regorafenib tablet)
- sunitinib capsule
- Sutent (sunitinib malate capsule)
- Tabloid (thioguanine tablet)
- Tabrecta (capmatinib tablet)
- Tafinlar (dabrafenib mesylate capsule and tablet for suspension)
- Tagrisso (osimertinib tablet)
- Talzenna (talazoparib capsule)
- Tarceva (erlotinib hydrochloride tablet)
- Targretin (bexarotene capsule and topical gel)
- Tasigna (nilotinib hydrochloride capsule)
- Tazverik (tazemetostat tablet
- temozolomide capsule

Last Reviewed: 1/16/19, 5/15/19, 7/17/19, 9/18/19, 11/20/19, 3/18/20, 5/20/20, 7/15/20, 9/16/20, 11/18/20, 1/20/21, 3/17/21, 5/19/21, 7/21/21, 9/15/21, 11/17/21, 3/16/22, 7/20/22, 9/21/22, 1/20/23, 5/19/23, 7/21/23, 11/17/23, 1/19/24, 3/15/24
Effective Date: 2/1/19, 7/1/19, 9/1/19, 10/15/2019, 1/1/20, 5/1/20, 7/1/20,8/15/20, 11/15/20, 12/15/20, 3/1/20, 5/1/21, 6/15/21, 9/1/21, 11/1/21, 1/1/22, 4/15/22, 9/1/22, 11/15/22, 3/15/23, 7/15/23, 12/20/23, 4/15/24

- Gleostine (lomustine capsule)
- Gleevec (imatinib tablet)
- Hycamtin (topotecan capsule)
- Ibrance (palbociclib capsule and tablet)
- Iclusig (ponatinib hydrochloride tablet)
- Idhifa (enasidenib tablet)
- imatinib tablet
- Imbruvica (ibrutinib capsule, tablet, and suspension)
- Inlyta (axitinib tablet)
- Inrebic (fedratinib capsule)
- Iressa (gefitinib tablet)
- Jakafi (ruxolitinib tablet)
- Jaypirca (pirtobrutinib tablet)
- Kisqali (ribociclib tablet)
- Krazati (adagrasib tablet)
- lapatinib tablet
- lenalidomide capsule
- Lenvima (lenvatinib capsule)
- Leukeran (chlorambucil tablet)
- Lonsurf (trifluridine/tipiracil tablet)
- Lorbrena (lorlatinib tablet)
- Lumakras (sotorasib tablet)
- Lynparza (olaparib tablet)
- Lytgobi (futibatinib tablet)
- Matulane (procarbazine capsule)
- Mekinist (trametinib dimethyl sulfoxide tablet and oral solution)
- Mektovi (binimetinib tablet)
- melphalan tablet
- Tepmetko (tepotinib tablet)
- Thalomid (thalidomide oral capsule)
- Tibsovo (ivosidenib tablet)
- Trelstar (triptorelin pamoate powder for IM injection)
- Tukysa (tucatinib tablet)
- Turalio (pexidartinib capsules)
- Tykerb (lapatinib ditosylate tablet)
- Valchlor (mechlorethamine hydrochloride topical gel)
- Vanflyta (quizartinib oral tablet)
- Venclexta (venetoclax tablet)
- Verzenio (abemaciclib tablet)
- Vitrakvi (larotrectinib tablet and oral solution)
- Vizimpro (dacomitinib tablet)
- Vonjo (pacritinib capsule)
- Votrient (pazopanib hydrochloride tablet)
- Welireg (belzutifan tablet)
- Xalkori (crizotinib capsule and oral pellets)
- Xospata (gilteritinib tablet)
- Xpovio (selinexor tablet)
- Xtandi (enzalutamide capsule)
- Xgeva (denosumab vial)
- Yonsa (abiraterone acetate tablet)
- Zejula (niraparib tablet)
- Zelboraf (vemurafenib tablet)
- Zolinza (vorinostat capsule)
- Zydelig (idelalisib tablet)
- Zykadia (ceritinib tablet)
- Zytiga (abiraterone acetate tablet)

Indication(s)

- Refer to major compendia for supported use

Dosing

- Refer to indication specific compendia supported dosing

Initial Authorization Criteria

Last Reviewed: 1/16/19, 5/15/19, 7/17/19, 9/18/19, 11/20/19, 3/18/20, 5/20/20, 7/15/20, 9/16/20, 11/18/20, 1/20/21, 3/17/21, 5/19/21, 7/21/21, 9/15/21, 11/17/21, 3/16/22, 7/20/22, 9/21/22, 1/20/23, 3/17,2023, 5/19/23, 7/21/23, 11/17/23, 1/19/24, 3/15/24
Effective Date: 2/1/19, 7/1/19, 9/1/19, 10/15/2019, 1/1/20, 5/1/20, 7/1/20, 8/15/20, 11/15/20, 12/15/20, 3/1/21, 5/1/21, 6/15/21, 9/1/21, 11/1/21, 1/1/22, 4/15/22, 9/1/22, 11/15/22, 3/1/23, 7/15/23, 12/20/23, 4/15/24

1. Is the request for continuation of therapy with the same anti-cancer medication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the medication being requested to be used for an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #4
    b. If no, continue to #3
3. Is the medication being requested being used for an indication supported by the National Comprehensive Cancer Network (NCCN) with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA or supported by the NCCN recommendation with an evidence level of 2A or higher? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there clinical documentation confirming disease responsiveness to therapy provided? (Examples include reduction in tumor size, objective response, delay in progression, partial response, etc.) (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with an oncologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 1/16/19, 5/15/19, 7/17/19, 9/18/19, 11/20/19, 3/18/20, 5/20/20, 7/15/20, 9/16/20, 11/18/20, 1/20/21, 3/17/21, 5/19/21, 7/21/21, 9/15/21, 11/17/21, 3/16/22, 7/20/22, 9/21/22, 1/20/23, 3/17,2023, 5/19/23, 7/21/23, 11/17/23, 1/19/24, 3/15/24
Effective Date: 2/1/19, 7/1/19, 9/1/19, 10/15/2019, 1/1/20, 5/1/20, 7/1/20, 8/15/20, 11/15/20, 12/15/20, 3/1/21, 5/1/21, 6/15/21, 9/1/21, 11/1/21, 1/1/22, 4/15/22, 9/1/22, 11/15/22, 3/1/23, 7/15/23, 12/20/23, 4/15/24

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. National Comprehensive Cancer Network. NCCN – NCCN Guidelines. Available at: https://www.nccn.org.

Last Reviewed: 1/16/19, 5/15/19, 7/17/19, 9/18/19, 11/20/19, 3/18/20, 5/20/20, 7/15/20, 9/16/20, 11/18/20, 1/20/21, 3/17/21, 5/19/21, 7/21/21, 9/15/21, 11/17/21, 3/16/22, 7/20/22, 9/21/22, 1/20/23, 3/17,2023, 5/19/23, 7/21/23, 11/17/23, 1/19/24, 3/15/24

Effective Date: 2/1/19, 7/1/19, 9/1/19, 10/15/2019, 1/1/20, 5/1/20, 7/1/20, 8/15/20, 11/15/20, 12/15/20, 3/1/21, 5/1/21, 6/15/21, 9/1/21, 11/1/21, 1/1/22, 4/15/22, 9/1/22, 11/15/22, 3/1/23, 7/15/23, 12/20/23, 4/15/24

Oral and Nasal CGRP Antagonists
Prior Authorization Guidelines

Affected Medication(s)

- Ubrelvy (ubrogepant) oral tablet
- Nurtec (rimegepant) oral disintegrating tablet
- Qulipta (atogepant) oral tablet
- Zavzpret (zavegepant) nasal spray

FDA Approved Indication(s)

- Ubrelvy: Acute treatment of migraine with or without aura in adults
- Nurtec ODT:
  - Acute treatment of migraine with or without aura in adults
  - Preventive treatment of episodic migraine in adults
- Qulipta: Preventive treatment of episodic or chronic migraine in adults
- Zavzpret: Acute treatment of migraine with or without aura in adults

Dosing

- Refer to package insert for recommended dosing for corresponding diagnosis

Initial Authorization Criteria

1. Is the request for continuation of a CGRP antagonist agent?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member 18 years of age or older?
   a. If yes, continue to #4
   b. If no, clinical review required
4. What is the requested diagnosis?
   a. Management of acute migraine, continue to #5
   b. Prevention of chronic/episodic migraine, continue to #7
5. Does the member have a previous trial with inadequate response to TWO triptan drugs (i.e. sumatriptan, rizatriptan, naratriptan, zolmitriptan, etc.)?
   a. If yes, continue to #12
   b. If no, continue to #6
6. Does the member have a contraindication to all triptan therapies? (Example of contraindications include: history of coronary artery disease, cardiac accessory conduction pathway disorders, history of stroke, transient

ischemic attack, or hemiplegic or basilar migraine, peripheral vascular disease, ischemic bowel disease, uncontrolled hypertension, or severe hepatic impairment) (Provide supporting documentation)

   a. If yes, continue to #12
   b. If no, clinical review required

7. Has the member experienced 4 or more headache days per month? (Provide supporting documentation)

   a. If yes, continue to #8
   b. If no, clinical review required

8. Has the member had a two-month trial and failure or intolerance to (2) of the following alternative agents with differing mechanisms of action for migraine prophylaxis: topiramate, divalproex, metoprolol, propranolol, timolol, atenolol, nadolol. amitriptyline, nortriptyline, venlafaxine, duloxetine? (Provide supporting documentation)

   a. If yes, continue to #10
   b. If no, continue to #9

9. Does the member have contraindications to all of the following alternative agents used for migraine prophylaxis: topiramate, divalproex, metoprolol, propranolol, timolol, atenolol, nadolol, amitriptyline, nortriptyline, venlafaxine, duloxetine? (Provide supporting documentation)

   a. If yes, continue to #10
   b. If no, clinical review required

10. Does the member have a documented trial with insufficient response, intolerance, or contraindication to all injectable CGRP agents? (Provide supporting documentation)

    a. If yes, continue to #11
    b. If no, clinical review required

11. Will CGRP antagonist to be used in combination with Botox?

    a. If yes, clinical review required
    b. If no, continue to #12

12. Will the CGRP antagonist be used in combination with another CGRP antagonist?

    a. If yes, clinical review required
    b. If no, approve for 6 months unless otherwise specified

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?

   a. If yes, continue to #2
   b. If no, clinical review required

2. Has the member demonstrated a positive clinical response to therapy?

a. If yes, continue to #3
b. If no, clinical review required

3. Will the CGRP antagonist be used in combination with another CGRP antagonist?

a. If yes, clinical review required
b. If no, approve for 12 months unless otherwise specified

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Ubrelvy [prescribing information]. Allergan USA, Inc. Madison, NJ. March 2021.
2. Nurtec ODT [prescribing Information]. Biohaven Pharmaceuticals, Inc. New Haven, CT. December 2021.
3. Qulipta tablets [prescribing information]. Madison, NJ: AbbVie; September 2021.
4. Zavzpret nasal spray, [prescribing information]. New York, NY: Pfizer; 2023.
5. Ailani J, Burch RC, Robbins MS, on behalf of the Board of Directors of the American Headache Society. The American Headache Society Consensus Statement: Update on integrating new migraine treatments into clinical practice. Headache. 2021;61(7):1021-1039.

Oral Fentanyl Products
Prior Authorization Guidelines

Affected Medication(s)

- Fentora (fentanyl citrate) buccal tablet
- Fentanyl citrate OTFC
- Fentanyl citrate buccal tablet

Indication(s)

- **Fentanyl citrate OTFC**: Management of breakthrough pain in cancer patients 16 years of age and older who are already receiving and who are tolerant to around-the-clock opioid therapy for their underlying persistent cancer pain
- **Fentora**: Management of breakthrough pain in cancer patients 18 years of age and older who are already receiving and who are tolerant to around-the-clock opioid therapy for their underlying persistent cancer pain

Dosing

- Refer to corresponding package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for renewal of a previously approved oral fentanyl product prior authorization with the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of cancer diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member meet the appropriate age for the FDA approved use of the medication? (At least 16 years of age for Actiq and at least 18 years of age for all others)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member's cancer pain inadequately controlled despite adherence to around the clock opioid therapy? (Provide documentation of current medication regimen and inadequately controlled pain)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a trial with inadequate response to at least TWO other oral or parenteral short-acting narcotic products that are used for breakthrough pain in cancer patients? (examples: morphine, hydromorphone, and oxycodone)? (Provide documentation of medications tried and inadequate response)
    a. If yes, continue to #6
    b. If no, clinical review required

Last Reviewed: 11/21/18, 9/18/19, 7/21/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 10/15/19, 9/1/21, 5/1/22, 7/15/23, 8/1/24

6. Is the member considered opioid tolerant defined as those who are taking at least or an equivalent dose of another opioid for a week or longer? (Provide documentation of opioid tolerance)

    - 60 mg of oral morphine/day
    - 25 mcg of transdermal fentanyl/hour
    - 30 mg oral oxycodone/day
    - 8 mg oral hydromorphone/day
    - 60 mg hydrocodone/day

    a. If yes, continue to #7
    b. If no, clinical review required

7. Is the medication being prescribed by, or in consult with, an oncologist or palliative care provider?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member demonstrate a positive clinical response to therapy as documented by a greater control of break-through pain? (Provide supporting documentation)

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Fentora [package insert]. Salt Lake City, Utah: Cephalon, Inc.; September 2018.
2. Fentanyl Citrate OTFC [package insert]. Parsippany, NJ: Teva Pharmaceuticals; January 2024.

Last Reviewed: 11/21/18, 9/18/19, 7/21/21, 3/16/22, 5/19/23, 5/17/24
Effective Date: 1/1/19, 10/15/19, 9/1/21, 5/1/22, 7/15/23, 8/1/24

Orfadin®, Nityr® (nitisinone)
Prior Authorization Guidelines

Affected Medication(s)

- Orfadin oral capsule
- Nityr oral tablet
- nitisinone capsule
- Orfadin oral suspension

Indication(s)

- For the treatment of adult and pediatric patients with hereditary tyrosinemia type 1 (HT-1) in combination with dietary restriction of tyrosine and phenylalanine

Dosing:

- 0.5 mg/kg orally twice daily, maximum dose of 2 mg/kg daily

Initial Authorization Criteria

1. Is the request for continuation of nitisinone therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is nitisinone being requested for an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is documentation confirming diagnosis of hereditary tyrosinemia type 1 provided? (Provide documentation of biochemical testing, clinical presentation, and/or DNA testing result)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is baseline urine or plasma succinylacetone level provided? (Provide baseline urine or plasma succinylacetone level for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is documentation of adherence to nutritional therapy provided? (Provide supporting documentation of restriction of tyrosine and phenylalanine adherence)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the member's current weight provided? (Provide member's weight for review)
    a. If yes, continue to #7

Last Reviewed: 12/19/18, 7/21/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 9/1/21, 6/1/23

b. If no, provider outreach required

7. Is the request for Nityr tablets or Orfadin suspension?

    a. If yes, continue to #8
    b. If no, continue to #9

8. Does the member have an intolerance to nitisinone capsules or is clinical rationale supporting inability to take nitisinone provided? (Provide supporting documentation of intolerance or clinical rationale)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Is the treatment being prescribed by, or in consultation with, a provider that is specialized in treatment of hereditary tyrosinemia or related disorders?

    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is nitisinone being requested for an FDA approved or major compendia supported indication?

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is documentation of adherence to nutritional therapy received? (Provide supporting documentation of restriction of tyrosine and phenylalanine adherence)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Has documentation of significant clinical response to therapy been provided? (Provide supporting documentation of complete urine or plasma succinylacetone suppression confirmed by lab result)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is member's current weight provided? (Provide member's weight for review)

    a. If yes, continue to #5
    b. If no, provider outreach required

5. Is the treatment being prescribed by or in consultation with a provider that is specialized in treatment of hereditary tyrosinemia or related disorders?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as

medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Nitisinone (Nityr) oral tablet [package insert]. Cambridge, UK: Cycle Pharmaceuticals Ltd.; July 2022.
2. Nitisinone (Orfadin) oral capsule/suspension [package insert]. Waltham, MA: Sobi, Inc; June 2022.
3. Nityr. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed July 5, 2018.
4. Orfadin. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed July 30, 2018.
5. Grompe MD, Hahn MD, PhD, Rand MD. Disorders of tyrosine metabolism. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed July 5, 2018.
6. DynaMed Plus [Internet]. Ipswich (MA): EBSCO Information Services. 1995 - . Record No. 916953, Hereditary tyrosinemia; [updated 2017 Jan 31, cited July 5, 2018]. Available at: http://www.dynamed.com/login.aspx?direct=true&site=DynaMed&id=916953. Registration and login required.

Orgovyx® (relugolix)
Prior Authorization Guidelines

Affected Medication(s)

- Orgovyx (relugolix) oral tablet

FDA Approved Indication(s)

- Treatment of adults with advanced prostate cancer

Dosing

- Loading dose of 360mg on the first day of treatment followed by 120mg taken orally one time daily at approximately the same time each day

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the medication being requested for an FDA approved indication? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, continue to #3
3. Is the medication being requested for an indication supported by the National Comprehensive Cancer Network (NCCN) recommendation with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #5
    a. If no, clinical review required
5. Does the member have a previous trial with inadequate response, intolerance, or contraindication to leuprolide? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the medication being prescribed by, or in consultation with, an oncologist?
    a. If yes, approve for 4 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA or supported by NCCN recommendation with an evidence level of 2A or higher? (Provide supporting documentation)
    a. If yes, continue to #2

Last Reviewed: 3/17/21, 5/18/22, 7/21/23
Effective Date: 5/1/21, 7/15/22

b. If no, clinical review required

2. Is there clinical documentation confirming disease responsiveness to therapy provided? (Example includes testosterone levels < 50ng/dL) (Provide supporting documentation)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the medication being prescribed by or in consultation with an oncologist?

    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. ORGOVYX (relugolix) tablets, [package insert]. Brisbane, CA: Myovant Sciences, Inc.; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 26 Jan. 2021].
3. Shore, Neal D., et al. "Oral relugolix for androgen-deprivation therapy in advanced prostate cancer." New England Journal of Medicine 382.23 (2020): 2187-2196.
4. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Prostate Cancer. Version 3.2022 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/default.aspx. Accessed April 15, 2022.

Last Reviewed: 3/17/21, 5/18/22, 7/21/23
Effective Date: 5/1/21, 7/15/22

Oriahnn® (elagolix, estradiol, and norethindrone acetate), Myfembree® (relugolix, estradiol, and norethindrone acetate) Prior Authorization Guidelines

Affected Medication(s)

- Oriahnn (elagolix, estradiol, and norethindrone) capsules
- Myfembree (relugolix, estradiol, and norethindrone) tablets

FDA Approved Indication(s)

- Management of heavy menstrual bleeding associated with uterine leiomyomas (fibroids) in premenopausal women
- Management of moderate to severe pain associated with endometriosis in premenopausal patients (Myfembree)

Dosing

- Oriahnn:
  - One capsule (elagolix 300mg, estradiol 1mg, norethindrone acetate 0.5mg) by mouth in the morning and one capsule (elagolix 300mg) in the evening. Note: Use of Oriahnn should be limited to 24 months due to risk of continued bone loss, which may not be reversible
- Myfembree:
  - One tablet (relugolix 40mg, estradiol 1mg and norethindrone acetate 0.5mg) one time daily

Initial Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #2
   b. If no, clinical review required
2. Is the treatment prescribed by, or in consultation with, an obstetrics/gynecologist or an endocrinologist?
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member premenopausal?
   a. If yes, continue to #4
   b. If no, clinical review required
4. What is the requested diagnosis?
   a. Heavy menstrual bleeding associated with uterine fibroids, continue to #5
   b. Pain associated with endometriosis, continue to #7
5. Has the member previously had a 3-month trial with at least 2 different hormonal contraceptives? (May include oral combination, oral progestin only, and/or progestin-releasing IUD) (Provide supporting documentation)
   a. If yes, continue to #6
   b. If no, clinical review required

Last Reviewed: 11/18/20, 1/19/22, 3/17/23, 3/15/24
Effective Date: 12/15/20, 3/1/22, 6/1/23, 4/15/24

6. Does the member have a previous trial with inadequate response, intolerance, or contraindication to tranexamic acid? (Provide supporting documentation)
    a. If yes, approve for 24 months
    b. If no, clinical review required

7. Does the member have a previous trial with inadequate response, intolerance, or contraindication to at least two non-steroidal anti-inflammatory drugs (NSAID)? (Provide supporting documentation)
    a. If yes, continue to #8
    b. If no, clinical review required

8. Has the member previously had a 3-month trial with at least one hormonal contraceptive? (May include oral combination, oral progestin only, and/or progestin-releasing IUD) (Provide supporting documentation)
    a. If yes, approve for 24 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Drugs@FDA: FDA Approved Drug Products. 2020. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 14 July, 2020].
2. ORIAHNN (elagolix, estradiol, and norethindrone acetate capsules; elagolix capsules), [package insert]. North Chicago, IL: Abvvie, Inc.; 2020.
3. MYFEMBREE® (relugolix, estradiol, and norethindrone acetate) tablets, [package insert]. Bisband, CA: Myovant Sciences; 2022.
4. Schlaff, William D., et al. "Elagolix for heavy menstrual bleeding in women with uterine fibroids." New England Journal of Medicine (2020).
5. NG88, N. I. C. E. "Heavy Menstrual Bleeding: assessment and management National Institute for Health and Clinical Excellence (NICE); 2018."
6. American College of Obstetricians and Gynecologists. "ACOG committee opinion no. 557: management of acute abnormal uterine bleeding in nonpregnant reproductive-aged women." Obstetrics and gynecology 121.4 (2013): 891.
7. Neri, Manuela, et al. "Clinical Utility Of Elagolix As An Oral Treatment For Women With Uterine Fibroids: A Short Report On The Emerging Efficacy Data." International Journal of Women's Health 11 (2019): 535.
8. American College of Obstetricians and Gynecologists. ACOG Practice Bulletin. Management of Symptomatic Uterine Leiomyomas. June 2021. Available at: https://www.acog.org/clinical/clinical-guidance/practice-bulletin/articles/2021/06/management-of-symptomatic-uterine-leiomyomas. Accessed on December 9, 2022.

Last Reviewed: 11/18/20, 1/19/22, 3/17/23, 3/15/24
Effective Date: 12/15/20, 3/1/22, 6/1/23, 4/15/24

Falcone, Tommaso, and Rebecca Flyckt. "Clinical management of endometriosis." Obstetrics & Gynecology 131.3 (2018): 557-571.

Orkambi® (lumacaftor/ivacaftor)
Prior Authorization Guidelines

Affected Medication(s)

- Orkambi oral tablet
- Orkambi oral granules

FDA Approved Indication(s)

- Treatment of cystic fibrosis (CF) in patients age 1 year and older who are homozygous for the F508del mutation in the CFTR gene

Dosing

- For patients less than 2 years old weighing 7 to less than 9 kg: One packet of Orkambi® (lumacaftor 75mg/ivacaftor 94mg) granules every 12 hours with fat-containing food
- For patients less than 2 years old weighing 9 to less than 14 kg: One packet of Orkambi® (lumacaftor 100mg/ivacaftor 125mg) granules every 12 hours with fat-containing food
- For patients less than 2 years old weighing 14kg or greater: One packet of Orkambi® (lumacaftor 150mg/ivacaftor 188mg) granules every 12 hours with fat-containing food
- For patients 2-5 years old weighing less than 14 kg: One packet of Orkambi (lumacaftor 100mg/ivacaftor 125mg) granules every 12 hours with fat-containing food
- For patients 2-5 years old weighing 14 kg or greater: One packet of Orkambi (lumacaftor 150mg/ivacaftor 188mg) granules every 12 hours with fat-containing food
- For patients 6-11 years old: Two Orkambi (lumacaftor 100mg/ivacaftor 125mg) tablets every 12 hours with fat-containing food
- For patients 12 years and older: Two Orkambi (lumacaftor 200mg/ivacaftor 125mg) tablets every 12 hours with fat-containing food

Initial Authorization Criteria

1. Is the request for continuation of Orkambi (lumacaftor/ivacaftor) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is there documentation that the member has the homozygous F508del mutation by an FDA-cleared CF mutation test? (Provide report for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has documentation of pulmonary function (baseline FEV1) and liver function (ALT and AST) been provided and are the liver enzymes within normal range? (Provide documentation of pulmonary and liver tests for review)
    a. If yes, continue to #5

Last Reviewed: 10/3/18, 5/20/20, 5/19/21, 5/18/22, 11/16/22, 11/17/23
Effective Date: 1/1/19, 7/1/20, 1/1/23

b. If no, clinical review required

5. Is the member at least 1 year of age?

   a. If yes, continue to #6
   b. If no, clinical review required

6. Is Orkambi (lumacaftor/ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis member?

   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)

   a. If yes, continue to #2
   b. If no, clinical review required

2. Were updated chart notes (within past year) provided with documentation of clinical response to prior therapy received? (Provide documentation of improvement of FEV1 from baseline and/or a reduction in the number of pulmonary exacerbations)

   a. If yes, continue to #3
   b. If no, clinical review required

3. Has documentation been provided of liver function tests (ALT and AST) within the last year and are they within normal limits? (Provide ALT and AST levels for review)

   a. If yes, continue to #4
   b. If no, clinical review required

4. Is Orkambi (lumacaftor/ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis member?

   a. If yes, approve for 12 months unless otherwise specified
   b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Orkambi® (lumacaftor/ivacaftor) [Prescribing Information]. Boston, MA: Vertex Pharmaceuticals Inc. September 2022.
2. Orkambi®. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed October 14, 2023.

Last Reviewed: 10/3/18, 5/20/20, 5/19/21, 5/18/22, 11/16/22, 10/15/2023, 11/17/23
Effective Date: 1/1/19, 7/1/20, 1/1/23, 11/18/2023

3. Simon, MD. Cystic fibrosis: Overview of the treatment of lung disease. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed May 14, 2020.

Oxervate® (cenegermin-bkbj)
Prior Authorization Guidelines

Affected Medication(s)

- Oxervate ophthalmic solution

FDA Approved Indication(s)

- Treatment of neurotrophic keratitis

Dosing

- One drop in affected eye(s) 6 times per day for 8 weeks

Initial Authorization Criteria

1. Is the medication being requested for an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 2 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a diagnosis of neurotrophic keratitis (NK) stage 2 or stage 3? (Characterized as persistent corneal epithelial defect and/or corneal stroma involvement with presence corneal ulcer)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has the member trialed both preservative-free artificial tears and topical antibiotic eye drops with inadequate response? (Provide documentation of previous medication history)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member previously been treated with a course of Oxervate for the same eye? (Note: Re-treatment with Oxervate is not supported)
    a. If yes, clinical review required
    b. If no, continue to #6
6. Is the medication being prescribed by, or in consultation with, and ophthalmologist?
    a. If yes, approve for 8 weeks
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

Last Reviewed: 7/17/19, 7/21/21, 7/20/22, 7/21/23
Effective Date: 9/1/19

References:

1. Drugs@FDA: FDA Approved Drug Products. 2018. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 5 Dec. 2018].
2. OXERVATE (cenegermin-bkbj) ophthalmic solution [package insert]. Boston, MA: Dompe US, Inc.; 2017.
3. Sacchetti M, Lambiase A. Diagnosis and management of neurotrophic keratitis. Clin Ophthalmol. 2014;8:571–579. Published 2014 Mar 19. doi:10.2147/OPTH.S45921
4. Bonini S, Lambiase A, Rama P et al. Phase 2 randomized, double-masked, vehicle-controlled trial of recombinant human nerve growth factor for neurotrophic keratitis. Ophthalmology 2018;125:1332–1343.
5. Semeraro F, Forbice E, Romano V, et al. Neurotrophic Keratitis. Ophthalmologica 2014;231:191-197. doi: 10.1159/000354380
6. National Institute for Health and Care Excellence. Cenegermin for treating neurotrophic keratits. https://www.nice.org.uk/guidance/ta532. Published July, 2018. Accessed May 7, 2019.
7. Graham RH and Hendrix MA. Neurotrophic Keratitis Treatment and Management. Medscape. https://emedicine.medscape.com/article/1194889. Updated September 13, 2018. Accessed May 7, 2019.

Last Reviewed: 7/17/19, 7/21/21, 7/20/22, 7/21/23
Effective Date: 9/1/19

Oral Pulmonary Arterial Hypertension (PAH) Agents
Prior Authorization Guidelines

Affected Medication(s)

- Adcirca (tadalafil) oral tablet
- Adempas (riociguat) oral tablet
- Alyq (tadalafil) oral tablet
- Ambrisentan oral tablet
- Bosentan oral tablet
- Letairis (ambrisentan) oral tablet
- Opsumit (macitentan) oral tablet
- Opsynvi (macitentan-tadalafil) oral tablet
- Orenitram ER (treprostinil) oral tablet
- Revatio (sildenafil) oral tablet
- Revatio (sildenafil) oral powder for suspension
- Sildenafil oral tablet (20 mg tablet only)
- Sildenafil 10 mg/ml oral suspension
- Tadalafil oral tablet (20 mg tablet only)
- Tracleer (bosentan) oral tablet
- Tracleer (bosentan) oral tablet for suspension
- Tyvaso (treprostinil) inhalation solution
- Uptravi (selexipag) oral tablet
- Ventavis (iloprost) inhalation solution

FDA Approved Indication(s)

- **Adcirca / Alyq**
  - Treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve exercise ability
- **Adempas**
  - Treatment of adults with persistent/recurrent chronic thromboembolic pulmonary hypertension (CTEPH) (WHO Group 4) after surgical treatment, or inoperable CTEPH, to improve exercise capacity and WHO functional class
  - Treatment of adults with PAH (WHO Group 1), to improve exercise capacity, WHO functional class and to delay clinical worsening
- **Letairis**
  - Treatment of PAH (WHO Group 1):
    - To improve exercise ability and delay clinical worsening
    - In combination with tadalafil to reduce the risks of disease progression and hospitalization for worsening PAH, and to improve exercise ability
- **Opsumit**
  - Treatment of PAH (WHO Group I) to reduce the risk of disease progression and hospitalization for PAH
- **Opsynvi**
  - Treatment of adults with PAH, WHO Group I and WHO Functional Class (FC) II–III). Individually, macitentan reduces the risk of clinical worsening events and hospitalization, and tadalafil improves exercise ability.
- **Orenitram ER**
  - Treatment of PAH (WHO Group I) to delay disease progression and to improve exercise capacity

Last Reviewed: 12/19/18, 7/17/19, 5/18/22, 9/21/22, 9/15/23, 5/17/24
Effective Date: 1/1/19, 9/1/19, 7/15/22, 11/1/22, 11/20/23, 6/15/24

- **Revatio**
  - Treatment of PAH (WHO Group 1) in adults to improve exercise ability and delay clinical worsening
  - Treatment of PAH (WHO Group 1) in pediatric patients 1 to 17 years old to improve exercise ability and, in pediatric patients too young to perform standardized exercise testing, pulmonary hemodynamics thought to underlie improvements in exercise
- **Tracleer**
  - Treatment of PAH (WHO Group 1):
    - In adults to improve exercise ability and to decrease clinical worsening
    - In pediatric patients ages 3 years and older with idiopathic or congenital PAH to improve pulmonary vascular resistance (PVR), which is expected to result in an improvement in exercise ability
- **Tyvaso**
  Inhaled:
  - Treatment of PAH (WHO Group I) in adult patients to improve exercise ability
  - Treatment of pulmonary hypertension associated with interstitial lung disease (WHO Group 3) to improve exercise ability.
- **Uptravi**
  - Treatment of pulmonary arterial hypertension (WHO Group I) to delay disease progression and reduce the risk of hospitalization for PAH
- **Ventavis**
  - Treatment of pulmonary arterial hypertension (WHO group I) in adult patients to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of clinical deterioration

Dosing

- Refer to corresponding package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same oral pulmonary arterial hypertension agent?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the request for use to treat PAH World Health OrganizaAtion (WHO) Group 1? (Provide documentation of PAH, WHO Group 1)
   a. If yes, continue to #6
   b. If no, continue to #4
4. Is the request for use to treat persistent/recurrent chronic thromboembolic pulmonary hypertension (CTEPH), WHO Group 4? (Provide documentation of CTEPH WHO Group 4)
   a. If yes, continue to corresponding criteria

b. If no, continue to #5

5. Is the request for Tyvaso for management of pulmonary hypertension associated with interstitial lung disease (WHO Group 3)? (Provide supporting documentation)
    a. If yes, continue to corresponding criteria
    b. If no, continue to #6
6. Is the request for a major compendia supported indication?
    a. If yes, continue to #9
    b. If no, clinical review required
7. Has the diagnosis been confirmed by right heart catheterization demonstrating mPAP $\geq$ 20 mmHg, PVR $\geq$ 3 Wood units, and PCWP $\leq$15 mmHg (or confirmed by another recommended test such as echocardiograph if catheterization cannot be performed)? (Provide supporting documentation)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Does the member have WHO or New York Heart Association (NYHA) Functional Class II-IV symptoms? (Provide supporting documentation)
    a. If yes, continue to #9
    b. If no, clinical review required
9. Is the prescriber a relevant specialist (i.e. pulmonologist or cardiologist)?
    a. If yes, continue to criteria corresponding to diagnosis and requested agent
    b. If no, clinical review required

Chronic Thromboembolic Pulmonary Hypertension

1. Has the diagnosis been confirmed by ventilation/perfusion (V/Q) scan, right heart catheterization demonstrating mPAP $\geq$ 20 mmHg and PCWP $\leq$15 mmHg, and presumed to be caused by thromboembolic occlusion of the pulmonary vasculature? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member status post pulmonary thromboendarterectomy, status post balloon pulmonary angioplasty, ineligible for surgery, or bridging definitive surgery? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the request for a female member with reproductive potential?
    a. If yes, continue to #5
    b. If no, continue to #6

5. Has a pregnancy test been obtained within 30 days prior to start of treatment to exclude pregnancy? (Provide supporting documentation)

    a. If yes, continue to #6
    b. If no, clinical review required

6. Will Adempas (riociguat) be used concomitantly with a PDE5 inhibitor or another organic nitrate?

    a. If yes, clinical review required
    b. If no, continue to #7

7. Is the medication being prescribed by, or in consultation with an appropriate specialist (i.e. pulmonologist or cardiologist)?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

PAH: Phosphodiesterase-5 Enzyme (PDE5) Inhibitors: Adcirca (tadalafil), Alyq (tadalafil), tadalafil, Revatio (sildenafil), sildenafil

1. Does the member currently take other organic nitrates in any form, regularly or intermittently? (Examples include isosorbide dinitrate, isosorbide mononitrate, and nitroglycerin) (Provide medication list for review)

    a. If yes, clinical review required
    b. If no, continue to #2

2. Will the requested PDE5 inhibitor be used concomitantly with Adempas (riociguat)? (Provide documentation of treatment plan)

    a. If yes, clinical review required
    b. If no, continue to #3

3. Is the request for Revatio (sildenafil) suspension?

    a. If yes, continue to #4
    b. If no, approve for 6 months

4. Is there documentation for why member is unable to take solid dosage form? (Provide documentation with rationale why the member is unable to take solid dosage form)

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

PAH: Endothelin Receptor Antagonists/ERA-PDE5 Combinations: Letairis (ambrisentan), Opsumit (macitentan), Opsynvi (macitentan-tadalafil), Tracleer (bosentan)

1. Does the member have documentation of inadequate response, contraindication, or intolerance to a PDE5 inhibitor (e.g. sildenafil) OR will the requested medication be taken along with a PDE5 inhibitor? (Provide documentation of inadequate response, contraindication, or intolerance)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the member a female of reproductive potential?

a. If yes, continue to #3
b. If no, continue to #4

3. Has a pregnancy test been obtained within 30 days prior to start of treatment to exclude pregnancy? (Provide supporting documentation)

   a. If yes, continue to #4
   b. If no, clinical review required

4. Does the member have preexisting moderate or severe hepatic impairment? (Provide documentation of hepatic status)

   a. If yes, clinical review required
   b. If no, continue to #5

5. Is the request for Letairis (ambrisentan)?

   a. If yes, continue to #6
   b. If no, continue to #7

6. Is the member 18 years of age or older?

   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

7. Is the request for Tracleer (bosentan)?

   a. If yes, continue to #8
   b. If no, continue to #9

8. Will Tracleer (bosentan) be used concurrently with cyclosporine or glyburide? (Provide documentation of medication list)

   a. If yes, clinical review required
   b. If no, approve for 6 months unless otherwise specified

9. Is the request for Opsumit (macitentan) or Opsynvi (macitentan-tadalafil)?

   a. If yes, continue to #10
   b. If no, clinical review required

10. Is the member 18 years of age or older?

    a. If yes, continue to #11
    b. If no, clinical review required

11. Does the member have documentation of inadequate response, contraindication, or intolerance to ambrisentan or bosentan? (Provide documentation of inadequate response, contraindication, or intolerance)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

<u>PAH: Soluble Guanylate Cyclase Stimulator: Adempas (riociguat)</u>

1. Is the member 18 years of age or older?

   a. If yes, continue to #2

b. If no, clinical review required

2. Does the member have documentation of inadequate response, contraindication, or intolerance to a PDE5 inhibitor (e.g. sildenafil)? (Provide documentation of inadequate response, contraindication, or intolerance)

   a. If yes, continue to #3
   b. If no, clinical review required

3. Does the member have documentation of inadequate response, contraindication, or intolerance to an endothelin receptor antagonist (e.g. Tracleer, Opsumit, or Letairis)? (Provide documentation of inadequate response, contraindication, or intolerance)

   a. If yes, continue to #4
   b. If no, clinical review required

4. Is the member a female of reproductive potential?

   a. If yes, continue to #5
   b. If no, continue to #6

5. Has pregnancy test been obtained within 30 days prior to start of treatment to exclude pregnancy? (Provide supporting documentation)

   a. If yes, continue to #6
   b. If no, clinical review required

6. Will Adempas (riociguat) be used concomitantly with a PDE5 inhibitor or another organic nitrate? (Provide documentation of treatment plan)

   a. If yes, clinical review required
   b. If no, approve for 6 months unless otherwise specified

<u>PAH: Prostanoids/Prostacyclins: Orenitram ER (treprostinil), Uptravi (selexipag), Tyvaso (treprostinil), Ventavis (iloprost)</u>

1. Does the member have documentation of inadequate response, contraindication, or intolerance to at least **two** of the following: PDE5 inhibitor, endothelin receptor antagonist, or Adempas (riociguat)? (Provide documentation of inadequate response, contraindication, or intolerance)

   a. If yes, continue to #2
   b. If no, clinical review required

2. Will the member be taking the requested agent in combination with another prostanoid/prostacyclin (e.g. epoprostenol, iloprost)? (Provide documentation of treatment plan)

   a. If yes, clinical review required
   b. If no, continue to #3

3. Does the member have severe hepatic impairment (Child Pugh Class C)? (Provide supporting documentation of hepatic status)

   a. If yes, clinical review required
   b. If no, approve for 6 months unless otherwise specified

Pulmonary Hypertension Associated with Interstitial Lung Disease: Tyvaso (treprostinil)

1. Has a diagnosis of pulmonary hypertension associated with interstitial lung disease (WHO group 3) been confirmed by right heart catheterization? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has a diagnosis of interstitial lung disease been confirmed by a computed tomography of the chest? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by, or in consultation with, an appropriate specialist (i.e. pulmonologist, cardiologist, or rheumatologist)?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Other Indications

1. Has the member tried and had an inadequate response OR dose the member have a contradiction to ALL standard treatment options for the requested indication? (Provide all prior treatment history, contraindication if appropriate, and treatment plan)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a positive clinical response to therapy? (Examples include improvement in 6-minute walking distance and/or stabilization or improvement in WHO functional class) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the prescriber a relevant specialist (i.e. pulmonologist or cardiologist)?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Last Reviewed: 12/19/18, 7/17/19, 5/18/22, 9/21/22, 9/15/23, 5/17/24
Effective Date: 1/1/19, 9/1/19, 7/15/22, 11/1/22, 11/20/23, 6/15/24

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Attridge RL, Moote R, Levine DJ. Chapter 17. Pulmonary Arterial Hypertension. In: DiPiro JT, Talbert RL, Yee GC, Matzke GR, Wells BG, Posey L. eds. Pharmacotherapy: A Pathophysiologic Approach, 9e New York, NY: McGraw-Hill; 2014.
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5. The Criteria Committee of the New York Heart Association. Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Great Vessels. 9th ed. Boston, Mass: Little, Brown & Co; 1994:253-256.
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7. McNeil K., Dunning J. (2007). Chronic thromboembolic pulmonary hypertension (CTEPH). Heart 93, 1152–1158. 10.1136/hrt.2004.053603
8. Simonneau, Gérald, et al. "Haemodynamic definitions and updated clinical classification of pulmonary hypertension." European Respiratory Journal 53.1 (2019).
9. Klinger, James R., et al. "Therapy for pulmonary arterial hypertension in adults: update of the CHEST guideline and expert panel report." Chest 155.3 (2019): 565-586.
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11. Humbert, Marc, et al. "2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension." European Respiratory Journal (2022).
12. Adcirca (tadalafil) [package insert]. Indianapolis, IN: Eli Lilly and Company; 2023.
13. Adempas (riociguat) [package insert]. Whippany, NJ: Bayer HealthCare Pharmaceuticals Inc; 2023.
14. Letairis (ambrisentan) [package insert]. Foster City, CA: Gilead Sciences, Inc; 2020.
15. Opsumit (macitentan) [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US; 2023.
16. Opsynvi (macitentan) [package insert]. Titusville, NJ: Actelion Pharmaceuticals US; 2024.
17. Orenitram ER (treprostinil) [package insert]. Research Triangle Park, NC: United Therapeutics Corp; 2023.
18. Revatio (sildenafil) [package insert]. NY, NY: Pfizer Labs; 2023.
19. Tracleer (bosentan) [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US; 2023.
20. Uptravi (selexipag) [package insert]. South San Francisco, CA: Actelion Pharmaceuticals US; 2023.
21. Ventavis (iloprost) [package insert]. Titusville, NJ: Actelion Pharmaceuticals US, Inc.; 2022.

Last Reviewed: 12/19/18, 7/17/19, 5/18/22, 9/21/22, 9/15/23, 5/17/24
Effective Date: 1/1/19, 9/1/19, 7/15/22, 11/1/22, 11/20/23, 6/15/24

22. Tyvaso (treprostinil) [package insert]. Research Triangle Park, NC: United Therapeutics Corp.; 2023.

Palforzia® (peanut allergen powder)
Prior Authorization Guidelines

Affected Medication(s)

- Palforzia oral capsule
- Palforzia powder sachet

FDA Approved Indication(s)

- Mitigation of allergic reactions, including anaphylaxis that may occur with accidental exposure to peanuts. Approved for use in patients with a confirmed diagnosis of peanut allergy. To be used in conjunction with a peanut avoidant diet

Dosing

- Administered in 3 sequential phases: Initial Dose Escalation, Up-Dosing and Maintenance. Open capsule(s) or sachet and empty into a few spoonfuls of refrigerated or room temperature semisolid food, mix well. Maintenance dose is 300mg daily; package insert should be referenced for initial dose escalation and up-dosing schedule.

Initial Authorization Criteria

1. Is the request for continuation of Palforzia (peanut allergen powder) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member between 4 and 17 years of age at the start of therapy?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a documented peanut allergy confirmed by peanut allergen skin testing ≥ 3mm compared to control or peanut-specific serum IgE ≥ 0.35kUA/L completed within 12 months?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a history of a previous systemic allergic reaction to peanuts requiring the use of epinephrine or resulting in ER visit/hospitalization?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is medical rationale provided for why adhering to a peanut avoidant diet alone is not sufficient therapy?
    a. If yes, continue to #7

Last Reviewed: 7/15/20, 9/15/21, 9/21/22, 9/15/23
Effective Date: 8/15/20

b. If no, clinical review required

7. Will the medication be used in combination with peanut avoidance?

   a. If yes, continue to #8
   b. If no, clinical review required

8. Will the first dose of each new up-dosing be administered under the supervision of a health care professional?

   a. If yes, continue to #9
   b. If no, clinical review required

9. Does the member have any of the following contraindications to therapy?

   - Uncontrolled asthma
   - A history of eosinophilic esophagitis
   - Other eosinophilic gastrointestinal disease
   - Chronic, recurrent, or severe gastroesophageal reflux disease (GERD)
   - Symptoms of dysphasia
   - Recurrent gastrointestinal symptoms of undiagnosed etiology

   a. If yes, clinical review required
   b. If no, continue to #10

10. Is the medication being prescribed by, or in consultation with, an allergist or immunologist?

    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

   a. If yes, continue to #2
   b. If no, clinical review required

2. Does the member remain adherent to peanut avoidant diet?

   a. If yes, continue to #3
   b. If no, clinical review required

3. Has the member had a positive clinical response to therapy as defined by an improvement in quality of life? (Provide supporting documentation of positive clinical response)

   a. If yes, continue to #4
   b. If no, clinical review required)

4. For member's who have used epinephrine while on Palforzia treatment, has supporting documentation been provided that demonstrates benefits of continued therapy outweigh the risks?

   a. If yes, continue to #5
   b. If no, clinical review required

5. Is the treatment being prescribed by or in consultation with an allergist or immunologist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

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References:

1. Palforzia [peanut allergen powder] capsules. Aimmune Therapeutics, Inc. Brisbane, CA; March 2021.
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Palynziq® (pegvaliase-pqpz)
Prior Authorization Guidelines

Affected Medication(s)

- Palynziq (pegvaliase-pqpz) subcutaneous solution

FDA Approved Indication(s)

- To reduce blood phenylalanine concentrations in adult patients with phenylketonuria who have uncontrolled blood phenylalanine concentrations greater than 600 micromol/L on existing management

Dosing

- Initial recommended dose: 2.5mg subcutaneously once weekly for four weeks
- Titrate dosage in step-wise manner over at least five weeks to achieve a dosage of 20mg one time daily, based on tolerability (Maximum dose: 60 mg/day)

Initial Authorization Criteria

1. Is the request for continuation of Palynziq (pegvaliase-pqpz) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a blood phenylalanine concentration of 600 micromol/L or greater? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member had a trial with inadequate response to a phenylalanine-restricted diet and does the treatment plan include continuation of a phenylalanine-restricted diet in combination with Palynziq (pegvaliase-pqpz)? (i.e. foods with high protein such as meat, fish, eggs, and milk products should be avoided) (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Has the member had a previous trial with inadequate response (defined as continued increased blood phenylalanine concentration), intolerance, or contraindication to treatment with sapropterin (Kuvan)? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required

Last Reviewed: 1/16/19, 7/21/21, 7/20/22, 7/21/23
Effective Date: 2/1/19

7. Does the treatment plan include monitoring blood phenylalanine concentration at least every 4 weeks until a maintenance dose is established? (Provide supporting documentation)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Is the treatment being prescribed by or in consultation with a specialist experienced in treatment of hyperphenylalaninemia?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication FDA approved or supported by major compendia? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within 1 year) provided with documentation of significant clinical response to therapy defined as a reduction in the blood phenylalanine level of at least 20% from pretreatment baseline or a blood phenylalanine level of 600 micromol/L or less? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the treatment plan include continuation of a phenylalanine-restricted diet in combination with Palynziq (pegvaliase-pqpz)? (i.e. foods with high protein such as meat, fish, eggs, and milk products should be avoided) (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by or in consultation with a specialist experienced in treatment of hyperphenylalaninemia?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

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References:

1. Drugs@FDA: FDA Approved Drug Products. 2022. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 15 Jul. 2022].
2. Palynziq (pegvaliase-pqpz) Injection [package insert]. Novato, CA: BioMarin Pharma, Inc; 2020.
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Last Reviewed: 1/16/19, 7/21/21, 7/20/22, 7/21/23
Effective Date: 2/1/19, 9/1/21

Paroxysmal Nocturnal Hemoglobinuria (PNH) Agents
Prior Authorization Guidelines

Affected Medication(s)

- Empaveli (pegcetacoplan) subcuataneous solution
- Fabhalta (iptacopan) oral capsule
- Voydeya (danicopan) oral tablet

FDA Approved Indication(s)

- Empaveli, Fabhalta: Treatment of adults with paroxysmal nocturnal hemoglobinuria (PNH)
- Voydeya: Treatment of extravascular hemolysis (EVH) in adults with paroxysmal nocturnal hemoglobinuria (PNH) as add-on therapy to ravulizumab (Ultomiris) or eculizumab (Soliris)

Dosing

- Empaveli: 1,080 mg subcutaneously twice weekly administered via an infusion pump or Empaveli on-body injector
- Fabhalta: 200 mg by mouth twice daily
- Voydeya: 150 mg by mouth three times daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a documented diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) as confirmed by detection of PNH clones of at least 10% by flow cytometry and the presence of at least 2 different glycosylphosphatidylinositol protein deficiencies within at least 2 different cell lines? (Provide supporting documentation of diagnosis)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a laboratory evidence of significant intravascular hemolysis (i.e., LDH ≥ 1.5x upper limit of normal) with symptomatic disease and at least one other indication for therapy regardless of transfusion dependence? (Provide supporting documentation of diagnosis)
    - Patient has symptomatic anemia (i.e., hemoglobin < 7 g/dL or hemoglobin < 10 g/dL, in at least two independent measurements in a patient with cardiac symptoms)
    - Presence of a thrombotic event related to PNH

Last Reviewed: 5/17/24
Effective Date: 6/15/24

- Presence of organ damage due to chronic hemolysis (i.e. renal insufficiency, pulmonary insufficiency/hypertension)
- Patient is pregnant and potential benefit outweighs potential fetal risk
- Patient has disabling fatigue
- Patient has abdominal pain requiring admission or opioid analgesia, dysphagia, or erectile dysfunction

a. If yes, continue to #6
b. If no, clinical review required

6. Has the member previously trialed Soliris or Ultomiris for at least 12 weeks with inadequate response, intolerance, or contraindication? (Provide supporting documentation)

a. If yes, continue to #7
b. If no, clinical review required

7. What is the requested medication?

a. Fabhalta, continue to #8
b. Voydeya, continue to #9
c. Empaveli, continue to #10

8. Has the member previously trialed Empaveli for at least 12 weeks with inadequate response, intolerance, or contraindication? (Provide supporting documentation)

a. If yes, continue to #10
b. If no, clinical review required

9. Will Voydeya be used in combination with Ultomiris or Soliris? (Provide treatment plan for review)

a. If yes, continue to #11
b. If no, clinical review required

10. Will the requested medication to be used with other complement inhibitor therapy?

a. If yes, clinical review required
b. If no, continue to #11

11. Is the medication being prescribed by, or in consult with, a hematologist?

a. If yes, approve for 6 months
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)

a. If yes, continue to #2
b. If no, clinical review required

2. Has the member developed a severe bone marrow failure syndrome, experienced spontaneous disease remission, or received a curative allogeneic stem cell transplant?

a. If yes, clinical review required
b. If no, continue to #3

3. Were updated chart notes (within past year) provided with documentation of significant clinical response to therapy received? (ex. decrease in serum LDH from baseline, stabilization or improvement in hemoglobin from baseline, decrease in transfusion requirement for baseline, reduction in thromboembolic events) (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the request for Voydeya?
    a. If yes, continue to #6
    b. If no, continue to #5
5. Will the requested medication to be used with other complement inhibitor therapy?
    a. If yes, clinical review required
    b. If no, continue to #6
6. Is the medication being prescribed by, or in consult with, a hematologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

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References:

1. FABHALTA® (iptacopan) capsules, [package insert]. East Hanover, NJ: Novartis Pharmaceuticals; 2024.
2. EMPAVELI (pegcetacoplan) injection solution [package insert]. Waltham, MA: Apellis Pharmaceuticals Inc; February 2024.
3. VOYDEYA (danicopan) tablets [package insert]. Boston, MA: Alexion Pharmaceuticals; April 2024.
4. Drugs@FDA: FDA Approved Drug Products. 2022. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 17 Jan. 2024].
5. Sahin F, Akay OM, Ayer M, et al. Pesg PNH diagnosis, follow-up and treatment guidelines. Am J Blood Res. 2016;6(2):19-27. Published 2016 Aug 5.
6. Peffault de Latour R, Roeth A, Kulasekararaj A, et al. Oral Monotherapy with Iptacopan, a Proximal Complement Inhibitor of Factor B, Has Superior Efficacy to Intravenous Terminal Complement Inhibition with Standard of Care Eculizumab or Ravulizumab and Favorable Safety in Patients with Paroxysmal Nocturnal Hemoglobinuria and Residual Anemia: Results from the Randomized, Active-Comparator-Controlled, Open-Label, Multicenter, Phase III Apply-PNH Study. Blood. 2022;140(Supplement 2):LBA-2-LBA-2.

PCSK9 Inhibitors
Prior Authorization Guidelines

Affected Medication(s)

- Praluent subcutaneous solution
- Repatha subcutaneous solution

FDA Approved Indication(s)

- **Praluent**
  - As an adjunct to diet, alone or in combination with other low-density lipoprotein cholesterol (LDL-C)-lowering therapies, in adults with primary hyperlipidemia, including heterozygous familial hypercholesterolemia (HeFH), to reduce LDL-C ,
  - To reduce the risk of myocardial infarction, stroke, and unstable angina requiring hospitalization in adults with established cardiovascular disease
  - As an adjunct to diet and other LDL-lowering therapies (e.g., statins, ezetimibe, LDL apheresis) in adults with homozygous familial hypercholesterolemia (HoFH) to reduce LDL-C
- **Repatha**
  - As an adjunct to diet, alone or in combination with other LCL-C-lowering therapies in adults with primary hyperlipidemia, including heterozygous familial hypercholesterolemia (HeFH), to reduce LDL-C
  - To reduce the risk of myocardial infarction, stroke, and coronary revascularization in adults with established cardiovascular disease
  - As an adjunct to diet and other LDL-C-lowering therapies in pediatric patients aged 10 years and older with HeFH, to reduce LDL-C
  - As an adjunct to diet and other LDL-C-lowering therapies (e.g., statins, ezetimibe, LDL apheresis) in adults and pediatric patients aged 10 years and older with homozygous familial hypercholesterolemia (HoFH), to reduce LDL-C

Dosing

- **Praluent**: 75 mg to 150 mg every 2 weeks OR 300 mg once per month
- **Repatha**: 140 mg every 2 weeks OR 420 mg once per month

Initial Authorization Criteria

1. Is the request for continuation of PCSK9 therapy?

   a. If yes, continue to Reauthorization
   b. If no, continue to #2

2. Is the requested medication being used for an FDA-approved indication?

   a. If yes, continue to #3
   b. If no, clinical review required

3. Is all of the following documentation provided? (Provide supporting documentation)

   - Complete lipid panel performed within the last 3 months
   - Baseline LDL-C (untreated)
   - Documentation of dietary measures being undertaken to lower cholesterol

Last Reviewed: 9/19/2018, 7/17/19, 11/17/21, 7/20/22, 7/21/23
Effective Date: 1/1/2019, 9/1/19, 1/1/22, 9/15/22, 9/15/23

a. If yes, continue to #4
b. If no, clinical review required

4. What is the diagnosis that PCSK9 inhibitor is being requested for? (Provide documentation of diagnosis)

    a. Heterozygous or Homozygous familial hypercholesterolemia (HeFH/HoFH), continue to #5
    b. Hypercholesterolemia with history of clinical atherosclerotic cardiovascular disease (ASCVD), continue to #7
    c. Other indication, continue to #9

5. Is pre-treatment LDL-cholesterol received (within 3 months) with baseline LDL-C ≥100 mg/dL on a maximally tolerated lipid-lowering regimen?

    a. If yes, continue to #6
    b. If no, clinical review required

6. Does the member meet at least one of the following: (Provide supporting documentation)

    - Family History of myocardial infarction before age 60 years in first-degree relative
    - Family History of myocardial infarction before age 50 years in second-degree relative
    - Family History of LDL-C greater than 190 mg/dL in a first- or second-degree relative
    - Tendinous xanthomata and/or arcus cornealis in first-degree relative or documented during physical examination
    - Functional mutation of LDL receptor, apoB, OR PCSK9 gene confirmed by genetic testing

    a. If yes, continue to #9
    b. If no, clinical review required

7. Is pre-treatment LDL-cholesterol received (within 3 months) with baseline LDL-C ≥ 70mg/dL on a maximally tolerated lipid-lowering regimen?

    a. If yes, continue to #8
    b. If no, clinical review required

8. Does the member have atherosclerotic cardiovascular disease (ASCVD) confirmed by at least one of the following: (Provide documentation of past medical history)

    - Acute coronary syndromes
    - History of myocardial infarction
    - Stable or unstable angina
    - Coronary or other arterial revascularization
    - Stroke
    - Transient ischemic attack
    - Peripheral arterial disease presumed to be of atherosclerotic origin

    a. If yes, continue to #9
    b. If no, clinical review required

9. Is the member currently receiving high-intensity statin therapy for consecutive 3 months and will continue with high-intensity statin therapy? High-intensity statin therapy includes: atorvastatin 40-80 mg or rosuvastatin 20-40 mg (Document current statin regimen with initiation date)

    a. If yes, continue to #14

b. If no, continue to #10

10. What is the rationale provided for avoiding high-intensity statin therapy? (Provide supporting documentation for avoidance)

    a. Statin intolerance due to myalgia or myopathy, continue to # 11
    b. History of rhabdomyolysis with creatinine kinase (CK) levels greater than 10-times upper limit of normal (document date occurred), continue to #13
    c. Labeled contraindication to all statins, continue to #13
    d. All other rationale, clinical review required

11. Is the member currently receiving a maximally tolerated dose of a statin AND ezetimibe and will continue statin and ezetimibe with PCSK9?

    a. If yes, continue to #14
    b. If no, continue to #12

12. Is documentation of persistent myalgia or myopathy on 2 separate 8 week trials with pravastatin, rosuvastatin, or fluvastatin provided? (Provide documentation of intolerance)

    a. If yes, continue to #13
    b. If no, clinical review required

13. Has the member been on ezetimibe for 3 consecutive months and will continue concurrently with PCSK9?

    a. If yes, continue to #14
    b. If no, clinical review required

14. Is the requested medication Repatha (evolocumab)?

    a. If yes, continue to #15
    b. If no, continue to #16

15. Does the member have a trial with inadequate response to, intolerance, or contraindication to Praluent (alirocumab)? (Provide supporting documentation)

    a. If yes, continue to #16
    b. If no, clinical review required

16. Is the medication being prescribed by or in consultation with cardiologist, endocrinologist, or lipid specialist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is updated lipid panel received with confirmation of significant reduction in LDL defined as a decrease in LDL levels of at least 40% from pre-treatment levels OR is updated LDL-C less than 100mg/dL? (Provide updated lab results)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the medication being prescribed by or in consultation with cardiologist, endocrinologist, or lipid specialist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Praluent (alirocumab) [Prescribing Information]. Bridgewater, NJ: Sanofi-Aventis U.S. LLC. April 2021.
2. Repatha (evolocumab) [Prescribing Information]. Thousand Oaks, CA: Amgen Inc. October 2021.
3. Praluent. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed July 31, 2017.
4. Repatha. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed July 31, 2017.
5. Stone NJ, Robinson JG, Lichtenstein AH, et al. 2013 ACC/AHA guideline on the treatment of blood cholesterol to reduce atherosclerotic cardiovascular risk in adults: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation.* 2014; June 24;129(25 Suupl 2):S1-45. Accessed July 31, 2018.
6. Rosenson RS, Durrington P. Familial hypercholesterolemia in adults: Overview. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. Available at: http://www.uptodate.com. Accessed July 31, 2018.
7. Grundy SM, Stone NJ, et al. 2018 ACC/AHA guideline on the management of blood cholesterol: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Journal of the American College of Cardiology. 2019;73(24):e285-e350. Available at: https://www.onlinejacc.org/content/73/24/e285.

Last Reviewed: 9/19/2018, 7/17/19, 11/17/21, 7/20/22, 7/21/23
Effective Date: 1/1/2019, 9/1/19, 1/1/22, 9/15/22, 9/15/23

Penicillamine
Prior Authorization Guidelines

Affected Medication(s)

- Cuprimine (penicillamine) oral capsule
- Depen (penicillamine) oral titratab
- penicillamine oral capsule/tablet

FDA Approved Indication(s)

- Indicated in the treatment of Wilson's disease, cystinuria, and in patients with severe, active rheumatoid arthritis who have failed to respond to an adequate trial of conventional therapy

Dosing

- See package insert for detail dosing information

Initial Authorization Criteria

1. Is the treatment being prescribed by, or in consultation with an appropriate specialist (Examples: GI specialist, hepatologist, or nephrologist)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the request for continuation of penicillamine therapy for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #3
3. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #4
    b. If no, clinical review required
4. What is the requested drug being used to treat? (Provide documentation of diagnosis)
    a. Wilson's disease, continue to #5
    b. Cystinuria, continue to #7
    c. Other indication, clinical review required
5. Is the request for penicillamine (Cuprimine) capsules?
    a. If yes, continue to #6
    b. If no, approve for 6 months unless otherwise specified
6. Has the member have a trial with insufficient response, intolerance, or contraindication to penicillamine (Depen) tablets? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed:11/18/20, 3/16/22, 5/19/23, 5/19/24
Effective Date: 1/15/21, 5/1/22, 6/15/24

7. Has the member had a trial with insufficient response or is resistant to conservative therapy such as increased fluid intake, sodium and protein restrictions? (Provide supporting documentation)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Has the member have a trial with insufficient response, intolerance, or contraindication to potassium citrate or potassium bicarbonate AND tiopronin? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Has the member have a positive clinical response to therapy? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the request for continuation of penicillamine therapy for the treatment of Wilson's disease or cystinuria?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Cuprimine (penicillamine) [Prescribing Information]. Bridgewater, NJ: Bausch Health US, LLC. November 2019.
2. Roberts EA, Schilsky ML. Diagnosis and Treatment of Wilson Disease: An Update. Hepatology. 2008;47(6):2089-2111 Available at: https://www.aasld.org/sites/default/files/2019-06/Wilson-Disease2009.pdf
3. European Association for the Study of the Liver. EASL Clinical Practice Guidelines: Wilson's disease. Journal of Hepatology. 2012;56(3):671-685. Available at: https://www.sciencedirect.com/science/article/pii/S0168827811008129?via%3Dihub
4. Penicillamine. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed October 19, 2020.

Last Reviewed: 11/18/20, 3/16/22, 5/19/23, 5/19/24
Effective Date: 1/15/21, 5/1/22, 6/15/24

Prevymis® (letermovir)
Prior Authorization Guidelines

Affected Medication(s)

- Prevymis oral tablet

FDA Approved Indication(s)

- Prophylaxis of cytomegalovirus (CMV) infection and disease in adult CMV-seropositive recipients [R+] of an allogeneic hematopoietic stem cell transplant (HSCT)
- Prophylaxis of CMV disease in adult kidney transplant recipients at high risk (Donor CMV seropositive/Recipient CMV seronegative [D+/R-])

Dosing

- HSCT: 480 mg once daily through day 100 post-transplantation
- Kidney Transplant: 480 mg once daily through day 200 post-transplantation

Authorization Criteria

1. Is the request for continuation of Prevymis (letermovir) therapy?
    a. If yes, clinical review required
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the current medication list been reviewed by the care team confirming no major drug interaction with Prevymis? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by or in consultation with a hematologist/oncologist, transplant specialist, or infectious disease specialist?
    a. If yes, continue to #5
    b. If no, clinical review required
5. What indication is Prevymis (letermovir) being requested for?
    a. Prophylaxis of CMV in allogeneic HSCT recipients, continue to #6
    b. Prophylaxis of CMV in adult kidney transplant recipients, continue to #8
6. Is Prevymis (letermovir) being initiated within 100 days of transplant? (Provide documentation of transplant date)
    a. If yes, continue to #7
    b. If no, clinical review required

Last Reviewed: 10/3/18, 7/21/21, 11/16/22, 9/15/2023
Effective Date: 1/1/19, 9/1/21, 11/20/23

7. Does the member meet <u>one</u> of the following criteria? (Provide documentation of CMV status for recipient or donor if applicable)

    - CMV-seropositive recipient OR
    - CMV seronegative recipient receiving a graft from seropositive donor (CMV D+/R-) who received a T cell-depleted allograft, an HLA-1 mismatched allograft, an umbilical cord blood allograft, or alemtuzumab

    a. If yes, approve for 4 months
    b. If no, clinical review required

8. Has documentation with rationale for avoidance or contraindication to both ganciclovir and valganciclovir been received? (Provide supporting documentation)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Is Prevymis (letermovir) being initiated within 200 days of transplant? (Provide documentation of transplant date)

    a. If yes, continue to #10
    b. If no, clinical review required

10. Is the member CMV-seronegative and received a transplant from a CMV-positive donor?

    a. If yes, approve for 7 months or up to 200 days from date of transplant
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Letermovir (Prevymis) [package insert]. Whitehouse Station, NJ: Merck & Co., Inc.; June 2023.
2. Prevymis. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed July 5, 2018.
3. Wingard JR, Marr KA, Thorner AR. Prevention of viral infections in hematopoietic cell transplant recipients. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed June 17, 2021

Last Reviewed: 10/3/18, 7/21/21, 11/16/22, 9/15/2023
Effective Date: 1/1/19, 9/1/21, 11/20/23

Prolia® (denosumab)
Prior Authorization Guidelines

Affected Medication(s)

- Prolia subcutaneous solution

FDA Approved Indication(s)

- Treatment of postmenopausal women with osteoporosis at high risk for fracture, defined as a history of osteoporotic fracture, or multiple risk factors for fracture; or patients who have failed or are intolerant to other available osteoporosis therapy.
- Treatment to increase bone mass in men with osteoporosis at high risk for fracture, defined as a history of osteoporotic fracture, or multiple risk factors for fracture; or patients who have failed or are intolerant to other available osteoporosis therapy
- Treatment of glucocorticoid-induced osteoporosis in men and women at high risk of fracture who are either initiating or continuing systemic glucocorticoids in a daily dosage equivalent to 7.5 mg or greater of prednisone and expected to remain on glucocorticoids for at least 6 months. High risk of fracture is defined as a history of osteoporotic fracture, multiple risk factors for fracture, or patients who have failed or are intolerant to other available osteoporosis therapy
- Treatment to increase bone mass in men at high risk for fracture receiving androgen deprivation therapy for nonmetastatic prostate cancer
- Treatment to increase bone mass in women at high risk for fracture receiving adjuvant aromatase inhibitor therapy for breast cancer

Dosing

- 60 mg subcutaneously once every 6 months

Initial Authorization Criteria

1. Is the request for continuation of Prolia (denosumab)?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a documented diagnosis of osteoporosis as indicated by one or more of the following? (Provide supporting documentation including DXA report within 2 years)
    - Hip DXA (femoral neck or total hip) or lumbar spine T-score less than or equal to -2.5 and/or forearm DXA 33% (one-third) radius
    - T-score less than or equal to -1 or low bone mass AND a history of fragility fracture to the hip or spine
    - T-score between -1 and -2.5 with a FRAX 10-year probability for major fracture ≥20% or hip fracture ≥3%

    a. If yes, continue to #6
    b. If no, continue to #4

Last Reviewed: 11/7/18, 5/19/21, 7/20/22, 7/21/23
Effective Date: 1/1/19

4. Does the member have non-metastatic prostate cancer and is currently receiving androgen deprivation therapy with confirmed osteopenia? (Provide supporting documentation)
    a. If yes, continue to #13
    b. If no, continue to #5
5. Does the member have breast cancer and is receiving adjuvant aromatase inhibitor with confirmed osteopenia?
    a. If yes, continue to #13
    b. If no, clinical review required
6. Is the member currently on systemic glucocorticoid therapy with a daily dosage equivalent of prednisone 7.5 mg or greater?
    a. If yes, continue to #7
    b. If no, continue to #8
7. Will the member continue with systemic glucocorticoid therapy at a daily dosage equivalent to 7.5 mg or greater of prednisone for at least 6 months?
    a. If yes, continue to #11
    b. If no, continue to #8
8. Is member at high risk for fracture as defined by one or more of the following? (Provide supporting documentation)
    - History of an osteoporotic fracture as an adult
    - Parental history of hip fracture
    - Low BMI
    - Rheumatoid arthritis
    - Alcohol intake of 3 or more drinks per day
    - Current smoking
    - History of oral glucocorticoids $\geq$ 5 mg/day of prednisone (or equivalent) for > 3 months in lifetime
    - Early menopause

    a. If yes, continue to #9
    b. If no, clinical review required
9. Does the member have five years of continuous treatment with bisphosphonates? (Provide all prior therapy history)
    a. If yes, continue to #13
    b. If no, continue to #10
10. Does the member have a trial with insufficient response to at least 12 months of bisphosphonate therapy (oral or IV) as defined by a decrease in T-score from baseline or member had a fracture while on bisphosphonate therapy? (Provide past relevant medication list with documentation of response to therapy)
    a. If yes, continue to #13
    b. If no, continue to #11

11. Does the member have a contraindication or intolerance to oral bisphosphonates? (Provide supporting documentation)
    a. If yes, continue to #12
    b. If no, clinical review required
12. Does the member with a contraindication or intolerance to IV bisphosphonates? (Provide supporting documentation)
    a. If yes, continue to #13
    b. If no, clinical review required
13. Is the member currently supplementing with at least 1,000 mg of calcium and 400 IU of vitamin D daily that will be continued throughout therapy? (Provide list of current relevant medications)
    a. If yes, continue to #14
    b. If no, clinical review required
14. Does member have any of the following contraindications to treatment with Prolia (denosumab)? (Provide serum calcium level for review)
    - Hypersensitivity to any component of the product
    - Hypocalcemia
    - Pregnancy

    a. If yes, clinical review required
    b. If no, continue to #15
15. Will Prolia (denosumab) be used concurrently with bisphosphonates or a PTH analog?
    a. If yes, clinical review required
    b. If no, continue to #16
16. Is the treatment being prescribed by or in consultation with an endocrinologist, a rheumatologist, or oncologist? (oncologist consultation if the patient has prostate or breast cancer)
    a. If yes, approve for up to 2 years unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Does the member continue to meet initial authorization criteria? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member demonstrate positive clinical response to therapy as defined by absence of fractures and/or an increase in bone mineral density from pretreatment baseline? (Provide updated DXA report and other supporting documentation)
    a. If yes, approve for up to 2 years unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Prolia [package insert]. Thousand Oaks, CA; Amgen, Inc.; May 2018. Accessed September 2018.
2. WHO Scientific Group on the Prevention and Management of Osteoporosis. Prevention and management of osteoporosis: report of a WHO scientific group. (WHO technical report series; 921). Geneva, Switzerland: WHO; 2000.
3. Kanis JA on behalf of the World Health Organization Scientific Group (2007). Assessment of osteoporosis at the primary health care level. Technical Report. World Health Organization Collaborating Center for Metabolic Bone Diseases. University of Sheffield, UK; 2007.
4. National Osteoporosis Foundation. Clinician's Guide to Prevention and Treatment of Osteoporosis. Washington, DC: National Osteoporosis Foundation; 2014.
5. Camacho PM, Petak SM, Binkley N, et al. AMERICAN ASSOCIATION OF CLINICAL ENDOCRINOLOGISTS AND AMERICAN COLLEGE OF ENDOCRINOLOGY CLINICAL PRACTICE GUIDELINES FOR THE DIAGNOSIS AND TREATMENT OF POSTMENOPAUSAL OSTEOPOROSIS - 2016. Endocr Pract. 2016 Sep 2; 22(Suppl 4):1-42.
6. Gnant M, Pfeiler G, Dubsky PC,et al. Adjuvant denosumab in breast cancer (ABCSG-18): a multicentre, randomised, double-blind, placebo-controlled trial. Lancet. 2015 Aug 1;386(9992):433-43.
7. Qaseem A, Forciea MA, McLean RM, Denberg TD; Clinical Guidelines Committee of the American College of Physicians. Treatment of Low Bone Density or Osteoporosis to Prevent Fractures in Men and Women: A Clinical Practice Guideline Update from the American College of Physicians. Ann Intern Med. 2017 May 9. doi: 10.7326/M15-1361.
8. Jeremiah MP, Unwin BK, Greenawald MH, et al. Diagnosis and Management of Osteoporosis. Am Fam Physician. 2015 Aug 15;92(4):261-8.
9. Camacho, Pauline M., et al. "American Association of Clinical Endocrinologists/American College of Endocrinology clinical practice guidelines for the diagnosis and treatment of postmenopausal osteoporosis 2020 update." Endocrine Practice 26 (2020): 1-46.

Last Reviewed: 11/7/18, 5/19/21, 7/20/22, 7/21/23
Effective Date: 1/1/19

Promacta® (eltrombopag olamine), Alvaiz™ (eltrombopag choline) Prior Authorization Guidelines

Affected Medication(s)

- Alvaiz oral tablet
- Promacta oral tablet
- Promacta oral suspension packet

FDA Approved Indication(s)

- Alvaiz
  - For the treatment of thrombocytopenia in adult and pediatric patients 6 years and older with persistent or chronic immune thrombocytopenia (ITP) who have had an insufficient response to corticosteroids, immunoglobulins, or splenectomy
  - For the treatment of thrombocytopenia in patients with chronic hepatitis C to allow the initiation and maintenance of interferon-based therapy
  - For the treatment of adult patients with severe aplastic anemia who have had an insufficient response to immunosuppressive therapy
- Promacta:
  - For the treatment of thrombocytopenia in adult and pediatric patients 1 years and older with persistent or chronic immune (idiopathic) thrombocytopenia (ITP) who have had an insufficient response to corticosteroids, immunoglobulins, or splenectomy
  - For the treatment of thrombocytopenia in patients with chronic hepatitis C to allow the initiation and maintenance of interferon-based therapy
  - For the treatment of patients with severe aplastic anemia who have had an insufficient response to immunosuppressive therapy
  - For first-line treatment of severe aplastic anemia, in combination with standard immunosuppressive therapy, in pediatric patients 2 years and older

Dosing

- Refer to respective package insert for dosing information
  -

Initial Authorization Criteria

1. Is the request for continuation of the same therapy for the same condition??
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the medication being requested for an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. What is the diagnosis that the drug is being requested for?
   a. Persistent or chronic idiopathic thrombocytopenic purpura (ITP), continue to corresponding criteria
   b. Severe aplastic anemia, continue to corresponding criteria
   c. Chronic hepatitis C-associated thrombocytopenia, continue to corresponding criteria

Last Reviewed: 11/7/18, 5/20/20, 5/18/22, 7/21/23, 3/15/24
Effective Date: 1/1/19, 7/1/20, 7/15/22, 4/15/24

d. Other indication, clinical review required

Persistent or Chronic Idiopathic Thrombocytopenic Purpura

1. Is the member's platelet count less than 30 x 10^9/L (30,000/mm)? (Provide platelet count for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member had an inadequate response, intolerance, or contraindication to glucocorticoids AND splenectomy or rituximab or immunoglobulins for ITP (Inadequate response defined as platelet count fails to reach greater than or equal to 50 x 10^9/L (50,000/mm))? (Provide supporting documentation for prior treatment history)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with a hematologist?
    a. If yes, approve for 3 months unless otherwise specified
    b. If no, clinical review required

Severe Aplastic Anemia

1. Is the member's platelet count less than 30 x 10^9/L (30,000/mm)? (Provide CBC with differential and platelet count for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member had a trial with an inadequate response, an intolerance, or contraindication to at least one prior immunosuppressive therapy (Example: cyclosporine)? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with a hematologist?
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Chronic Hepatitis C-associated Thrombocytopenia

1. Is the patient's platelet count less than 75 x 109/L (75,000/mm)? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there documentation of compensated liver disease (Defined as Child-Pugh Class A)? (Provide supported lab for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with a hepatologist or ID specialist?

a. If yes, approve for 2 months
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within previous 6 months) and supporting labs received with documentation meeting one of the following? (Provide supporting documentation including lab results for review)
    - Confirmed diagnosis of idiopathic thrombocytopenic purpura with platelet count greater than or equal to 50 x 10^9/L (50,000/mm)
    - Confirmed diagnosis of severe aplastic anemia with platelet count increases to 20 x 10^9/L above baseline OR stable platelet counts without transfusion for 8 or more weeks or hemoglobin increases by > 1.5 g/dL OR ANC increases 100% or ANC increase > 0.5 x10^9/L
    - Confirmed diagnosis of chronic hepatitis C-associated thrombocytopenia with platelet count increase to greater than or equal to 90 x 10^9/L (90,000/mm))

    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being prescribed by or in consultation with an appropriate specialist for the indicated diagnosis?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Promacta® (eltrombopag) [Prescribing Information]. Research Triangle Park, NC: GlaxoSmithKline LLC. April 2020.
2. Alvaiz™ (eltrombopag) [Prescribing Information]. East Hanover, NJ: Novartis. January 2024.
3. Promacta®. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed May 14, 2020.
4. George, PhD, Arnold, MD. Immune thrombocytopenia (ITP) in adults: Second-line and subsequent therapies. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed May 14, 2020.

Last Reviewed: 11/7/18, 5/20/20, 5/18/22, 7/21/23, 3/15/24
Effective Date: 1/1/19, 7/1/20, 7/15/22, 4/15/24

5. Afdhal NH, Dusheiko GM, Giannini EG, et al. Eltrombopag increases platelet numbers in thrombocytopenic patients with HCV infection and cirrhosis, allowing for effective antiviral therapy. Gastroenterology. 2014 Feb;146(2):442-52.e1.

Parathyroid Hormone (PTH) Analog Agents
Prior Authorization Guidelines

Affected Medication(s)

- Forteo (teriparatide) subcutaneous solution
- teriparatide subcutaneous solution
- Tymlos (abaloparatide) subcutaneous solution

FDA Approved Indication(s)

- Treatment of postmenopausal women with osteoporosis at high risk for fracture, defined as a history of osteoporotic fracture, multiple risk factors for fracture, or patients who have failed or are intolerant to other available osteoporosis therapy
  - Forteo, Tymlos
- To increase bone mass in men with primary or hypogonadal osteoporosis at high risk for fracture, defined as a history of osteoporotic fracture, multiple risk factors for fracture, or patients who have failed or are intolerant to other available osteoporosis therapy
  - Forteo
- Treatment of men and women with osteoporosis associated with sustained systemic glucocorticoid therapy (daily dosage equivalent to 5 mg or greater of prednisone) at high risk for fracture, defined as a history of osteoporotic fracture, multiple risk factors for fracture, or patients who have failed or are intolerant to other available osteoporosis therapy
  - Forteo
- To increase bone density in men with osteoporosis at high risk for fracture, defined as a history of osteoporotic fracture or multiple risk factors for fracture, or patients who have failed or are intolerant to other available osteoporosis therapy.
  - Tymlos

Dosing

- Forteo: 20 mcg subcutaneously once daily
- Tymlos: 80 mcg subcutaneously once daily

NOTE: Parathyroid hormone analogs have a lifetime cumulative treatment duration of 2 years

Authorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #2
   b. If no, clinical review required
2. Does the member have a documented diagnosis of osteoporosis as indicated by one or more of the following? (Provide supporting documentation including DXA report within 2 years)
   - Hip DXA (femoral neck or total hip) or lumbar spine T-score less than or equal to -2.5 and/or forearm DXA 33% (one-third) radius
   - T-score less than or equal to -1 or low bone mass AND a history of fragility fracture to the hip or spine
   - T-score between -1 and -2.5 with a FRAX 10-year probability for major fracture ≥20% or hip fracture ≥3%

Last Reviewed: 11/7/18, 5/20/20, 5/18/22, 7/21/23
Effective Date: 1/1/19, 7/1/20, 7/15/22, 9/15/23

a. If yes, continue to #3
b. If no, clinical review required

3. Does the member have a high risk for fracture as defined by one or more of the following? (Provide supporting documentation)

    - History of an osteoporotic fracture as an adult
    - Parental history of hip fracture
    - Low BMI
    - Rheumatoid arthritis
    - Alcohol intake of 3 or more drinks per day
    - Current smoking
    - History of oral glucocorticoids ≥ 5 mg/day of prednisone (or equivalent) for > 3 months in lifetime
    - Early menopause

    a. If yes, continue to #4
    b. If no, clinical review required

4. Does the member have five years of continuous treatment with bisphosphonates? (Provide all prior treatment history)

    a. If yes, continue to #8
    b. If no, continue to #5

5. Does the member have a trial with insufficient response to at least 12 months of bisphosphonate therapy (oral or IV) as defined by a decrease in T-score from baseline or member had a fracture while on bisphosphonate therapy? (Provide past relevant medication list with documentation of response to therapy)

    a. If yes, continue to #8
    b. If no, continue to #6

6. Does the member have a contradiction or intolerance to oral bisphosphonates? (Provide supporting documentation)

    a. If yes, continue to #7
    b. If no, clinical review required

7. Does the member have a contraindication or intolerance to IV bisphosphonates? (Provide supporting documentation)

    a. If yes, continue to #8
    b. If no, clinical review required

8. Does the member have documentation of a trial with insufficient response, an intolerance, or a contraindication to Prolia as defined by a decrease in T-score from baseline or member had a fracture while on Prolia therapy? (Provide past relevant medication list with documentation of response to therapy)

    a. If yes, continue to #9
    b. If no, clinical review required

9. For treatment of postmenopausal women with osteoporosis, is Tymlos being requested?

    a. If yes, continue to #11

b. If no, continue to #10
c. If not applicable, continue to #11

10. Is clinical rationale for avoiding Tymlos received? (Provide supporting documentation)
    a. If yes, continue to #11
    b. If no, clinical review required
11. Is the member currently supplementing with at least 1,000 mg of calcium and 400 IU of vitamin D daily that will be continued throughout therapy? (Provide list of current relevant medications)
    a. If yes, continue to #12
    b. If no, clinical review required
12. Is the member's serum calcium within normal range? (Provide serum calcium level for review)
    a. If yes, continue to #13
    b. If no, clinical review required
13. Does the member have increased risk of osteosarcoma? (i.e. Paget's disease of bone, unexplained elevations of alkaline phosphatase, open epiphyses, bone metastases or skeletal malignancies, hereditary disorders predisposing to osteosarcoma, or prior external beam or implant radiation therapy involving the skeleton)
    a. If yes, clinical review required
    b. If no, continue to #14
14. Will the requested parathyroid hormone analog be used concurrently with Prolia (denosumab), bisphosphonates, or another PTH analog?
    a. If yes, clinical review required
    b. If no, continue to #15
15. Is the treatment being prescribed by or in consultation with an endocrinologist or a rheumatologist?
    a. If yes, approve for 2 years or unless otherwise specified (lifetime therapy limit of 24 months)
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Forteo [package insert]. Indianapolis, IN; Eli Lilly and Company; April 2021.
2. Tymlos [package insert]. Waltham, MA; Radius Health; December 2021.
3. WHO Scientific Group on the Prevention and Management of Osteoporosis. Prevention and management of osteoporosis: report of a WHO scientific group. (WHO technical report series; 921). Geneva, Switzerland: WHO; 2000.
4. Kanis JA on behalf of the World Health Organization Scientific Group (2007). Assessment of osteoporosis at the primary health care level. Technical Report. World Health Organization Collaborating Center for Metabolic Bone Diseases. University of Sheffield, UK; 2007.

Last Reviewed: 11/7/18, 5/20/20, 5/18/22, 7/21/23
Effective Date: 1/1/19, 7/1/20, 7/15/22, 9/15/23

5. National Osteoporosis Foundation. Clinician's Guide to Prevention and Treatment of Osteoporosis. Washington, DC: National Osteoporosis Foundation; 2014.
6. Camacho PM, Petak SM, Binkley N, et al. AMERICAN ASSOCIATION OF CLINICAL ENDOCRINOLOGISTS AND AMERICAN COLLEGE OF ENDOCRINOLOGY CLINICAL PRACTICE GUIDELINES FOR THE DIAGNOSIS AND TREATMENT OF POSTMENOPAUSAL OSTEOPOROSIS - 2016. Endocr Pract. 2016 Sep 2; 22(Suppl 4):1-42.
7. Qaseem A, Forciea MA, McLean RM, Denberg TD; Clinical Guidelines Committee of the American College of Physicians. Treatment of Low Bone Density or Osteoporosis to Prevent Fractures in Men and Women: A Clinical Practice Guideline Update from the American College of Physicians. Ann Intern Med. 2017 May 9. doi: 10.7326/M15-1361.
8. Jeremiah MP, Unwin BK, Greenawald MH, et al. Diagnosis and Management of Osteoporosis. Am Fam Physician. 2015 Aug 15;92(4):261-8.
9. Camacho, Pauline M., et al. "American Association of Clinical Endocrinologists/American College of Endocrinology clinical practice guidelines for the diagnosis and treatment of postmenopausal osteoporosis—2020 update." Endocrine Practice 26 (2020): 1-46.

Pulmozyme® (dornase alfa), Bronchitol® (mannitol)
Prior Authorization Guidelines

Affected Medication(s)

- Pulmozyme (dornase alfa) inhalation solution
- Bronchitol (mannitol) inhalation powder

FDA Approved Indication(s)

- Pulmozyme: For daily administration in conjunction with standard therapies for the management of cystic fibrosis (CF) patients to improve pulmonary function. In CF patients with an FVC ≥ 40% of predicted, daily administration of Pulmozyme has also been shown to reduce the risk of respiratory tract infections requiring parenteral antibiotics
- Bronchitol: Add-on maintenance therapy to improve pulmonary function in adult patients 18 years and older with Cystic Fibrosis

Dosing

- Pulmozyme: 2.5 mg inhaled once or twice daily
- Bronchitol: 400mg inhaled twice daily

Initial Authorization Criteria

1. Is the request for continuation of Pulmozyme or Bronchitol therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a confirmed diagnosis of cystic fibrosis? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, continue to #6
4. Is the request for Bronchitol?
    a. If yes, continue to #5
    b. If no, continue to #7
5. Does the member have documentation of a previous trial with Pulmozyme in combination with hypertonic saline?
    a. If yes continue to #7
    b. If no, clinical review required
6. Has the member tried and had an inadequate response OR does the member have a contraindication to ALL standard treatment options for the requested indication? (Provide all prior treatment history, contraindication if appropriate, and treatment plan)
    a. If yes, continue to #7

Last Reviewed: 5/19/21, 11/16/22, 11/17/23
Effective Date: 6/15/21, 12/20/23

b. If no, clinical review required

7. Is the treatment being prescribed by or in consult with a pulmonologist or cystic fibrosis specialist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by or in consultation with a pulmonologist or cystic fibrosis specialist?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is documentation provided that the member is experiencing successful response to therapy? (Provide updated clinical information for review such as reduction in CF exacerbations compared to baseline, improvement in CF symptoms, reduction in respiratory infections, improvement in FEV1, etc.)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. BRONCHITOL® (mannitol) inhalation powder, [package insert]. Cary, NC: Chiesi USA, Inc; 2021.
2. PULMOZYME® (dornase alfa) solution, [package insert]. San Francisco, CA: Genentech, Inc; 2020.
3. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 12 Jan. 2021].
4. Flume, Patrick A., et al. "Efficacy and safety of inhaled dry-powder mannitol in adults with cystic fibrosis: An international, randomized controlled study." Journal of Cystic Fibrosis (2021).
5. Bilton D, Robinson P, Cooper P, Gallagher CG, Kolbe J, Fox H, Jaques A, Charlton B; CF301 Study Investigators. Inhaled dry powder mannitol in cystic fibrosis: an efficacy and safety study. Eur Respir J. 2011 Nov;38(5):1071-80. doi: 10.1183/09031936.00187510. Epub 2011 Apr 8. PMID: 21478216.
6. Aitken, Moira L., et al. "Long-term inhaled dry powder mannitol in cystic fibrosis: an international randomized study." American journal of respiratory and critical care medicine 185.6 (2012): 645-652.
7. Bilton, Diana, et al. "Pooled analysis of two large randomised phase III inhaled mannitol studies in cystic fibrosis." Journal of Cystic Fibrosis 12.4 (2013): 367-376.
8. Flume, Patrick A., et al. "Cystic fibrosis pulmonary guidelines: chronic medications for maintenance of lung health." American journal of respiratory and critical care medicine 176.10 (2007): 957-969.

Last Reviewed: 5/19/21, 11/16/22, 11/17/23
Effective Date: 6/15/21, 12/20/23

9. Villanueva, Gemma, et al. "Diagnosis and management of cystic fibrosis: summary of NICE guidance." Bmj 359 (2017).

Pyrukynd® (mitapivat)
Prior Authorization Guidelines

Affected Medication(s)

- Pyrukynd (mitapivat) oral tablet

FDA Approved Indication(s)

- Treatment of hemolytic anemia in adults with pyruvate kinase (PK) deficiency

Dosing

- Starting dose of 5mg twice daily; can be titrated up to 50mg by mouth two times daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is Pyrukynd (mitapivat) being requested for an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the patient 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of pyruvate kinase deficiency confirmed by biochemical (reduced PK activity in RBCs) or genetic testing (identifying a pathogenic PKLR gene mutation)? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have documentation of at least 2 variant alleles in the PKLR gene, of which at least 1 was a missense variant? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the member homozygous for the c.1436G>A (p.R479H) variant or have 2 non-missense variants (without the presence of another missense variant) in the PKLR gene? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #7
7. Is the member's hemoglobin 10 g/dL or less? (Provide supporting documentation)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Has the member previously had 6 or more transfusions in the past year or has severe symptomatic anemia? (Provide supporting documentation)

Last Reviewed: 5/18/22, 5/19/23, 5/17/24
Effective Date: 7/15/22, 6/15/24

a. If yes, continue to #9
b. If no, clinical review required

9. Is Pyrukynd (mitapivat) being prescribed by, or in consult with, a hematologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within past year) provided with documentation of significant clinical response to prior therapy received? (i.e. improvement in hemoglobin by at least 1.5 g/dL from baseline or reduction in RBC transfusions from baseline) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is Pyrukynd (mitapivat) being prescribed by, or in consult with, a hematologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. PYRUKYND® (mitapivat) tablets, [package insert]. Cambridge, MA: Agios Pharmaceuticals, Inc; 2022.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 29 Mar. 2022].
3. van Beers, Eduard J., et al. "Mitapivat (AG-348) in Adults with Pyruvate Kinase Deficiency Who Are Not Regularly Transfused: A Phase 3, Randomized, Multicenter, Double-Blind, Placebo-Controlled Study (ACTIVATE) in Progress." Blood 134 (2019): 4791.
4. Lynch, Megan, et al. "Mitapivat (AG-348) in adults with Pyruvate Kinase deficiency who are regularly transfused: a phase 3, open-label, multicenter, study (ACTIVATE-T) in progress." Blood 134 (2019): 3526.

Last Reviewed: 5/18/22, 5/19/23, 5/17/24
Effective Date: 7/15/22, 6/15/24

Recorlev® (levoketoconazole)
Prior Authorization Guidelines

Affected Medication(s)

- Recorlev (levoketoconazole) oral tablet

FDA Approved Indication(s)

- Treatment of endogenous hypercortisolemia in adult patients with Cushing's syndrome for whom surgery is not an option or has not been curative

Dosing

- Initial 150mg orally twice daily, titrated up to a max of 600mg twice daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is Recorlev (levoketoconazole) being requested for an FDA approved or compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the patient 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of endogenous Cushing's syndrome with a mean Urinary Free Cortisol level (UFC) greater than or equal to 1.5x the upper limit of normal (normal range: 11 to 138 nmol/day or 4 to 50 µg/day)? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member previously undergone pituitary surgery that was not curative or is the member not a candidate for surgery? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Has the member previously trialed a maximum tolerated dose of ketoconazole for at least 8 weeks with treatment failure or is there a documented intolerance or contraindication to ketoconazole? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #7
7. Is Recorlev (levoketoconazole) being prescribed by, or in consult with, an endocrinologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Last Reviewed: 5/18/22, 5/19/23, 5/17/24
Effective Date: 7/15/22, 6/15/24

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within past year) provided with documentation of significant clinical response to prior therapy received? (i.e. decrease in mUFC from baseline that is maintained within normal range) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is Recorlev (levoketoconazole) being prescribed by, or in consult with, an endocrinologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. RECORLEV (levoketoconazole) tablets, [package insert]. Chicago, IL: Xeris Pharmaceuticals, Inc; 2022.
2. DailyMed – Recorlev- levoketoconazole tablet. 2022. U.S. National Library of Medicine. National Institutes of Health. [online]
3. Lynnette K. Nieman, Beverly M. K. Biller, James W. Findling, M. Hassan Murad, John Newell-Price, Martin O. Savage, Antoine Tabarin, Treatment of Cushing's Syndrome: An Endocrine Society Clinical Practice Guideline, The Journal of Clinical Endocrinology & Metabolism, Volume 100, Issue 8, 1 August 2015, Pages 2807–2831, https://doi.org/10.1210/jc.2015-1818
4. Fleseriu, Maria, et al. "Consensus on diagnosis and management of Cushing's disease: a guideline update." The Lancet Diabetes & Endocrinology 9.12 (2021): 847-875.
5. Fleseriu, Maria, et al. "Efficacy and safety of levoketoconazole in the treatment of endogenous Cushing's syndrome (SONICS): a phase 3, multicentre, open-label, single-arm trial." The Lancet Diabetes & Endocrinology 7.11 (2019): 855-865.
6. Zacharieva, Sabina Z., et al. "MON-332 Safety and Efficacy of Levoketoconazole in the Treatment of Endogenous Cushing's Syndrome (LOGICS): A Double-Blind, Placebo-Controlled, Withdrawal Study." Journal of the Endocrine Society 4. Supplement 1 (2020): MON-332.

Last Reviewed: 5/18/22, 5/19/23, 5/17/24
Effective Date: 7/15/22, 6/15/24

Relistor® (methylnaltrexone bromide)
Prior Authorization Guidelines

Affected Medication(s)

- Relistor (methylnaltrexone) oral tablet
- Relistor (methylnaltrexone) subcutaneous solution
- Relistor (methylnaltrexone) vial

FDA Approved Indication(s)

- For the treatment of opioid-induced constipation (OIC) in adult patients with chronic non-cancer pain, including patients with chronic pain related to prior cancer or its treatment who do not require frequent (e.g., weekly) opioid dosage escalation
  - Relistor oral tablet
- For the treatment of OIC in adult patients with advanced illness or pain caused by active cancer who require opioid dosage escalation for palliative care
  - Relistor subcutaneous solution, vial

Dosing

- Refer to appropriate package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same opioid-induced constipation agent?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have a clinical diagnosis of opioid-induced constipation (OIC) as defined as the following? (Provide supporting documentation)
   - Loose stools are rarely present without the use of laxatives AND
   - More than 25% of SBMs associated with two or more of the following:
     - less than 3 spontaneous bowel movements (SBM) per week
     - straining
     - hard or lumpy stools
     - sensation of incomplete evacuation or anorectal obstruction/blockage
     - manual maneuvers to facilitate defecation

   a. If yes, continue to #4
   b. If no, clinical review required
4. Has the member been taking opioids for at least 4 weeks duration? (Provide supporting documentation)
   a. If yes, continue to #5
   b. If no, clinical review required

Last Reviewed: 12/19/18, 7/17/19, 5/19/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 9/1/19, 6/15/21, 6/1/23, 4/15/24

5. Is the member taking opioids for non-cancer pain or pain associated with a prior cancer?
    a. If yes, continue to #7
    b. If no and request is for subcutaneous solution or vial, continue to #6
    c. If no and request is for oral tablet, clinical review required
6. Is the member taking opioids due to advanced illness or require opioid dose escalation due to active cancer pain that requiring palliative care? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Has the member tried laxatives (e.g. psyllium, methylcellulose) **AND** at least one of the following categories for a minimum of 2 weeks (administered on a regular schedule, not PRN) and had inadequate response? (Provide documentation of medications trialed with response)
    - Stool softener (e.g. docusate)
    - Osmotic laxative (e.g. polyethylene glycol, lactulose, magnesium citrate)
    - Stimulant laxative (e.g. senna, bisacodyl)
    - Lubricant (e.g. mineral oil)

    a. If yes, continue to #8
    b. If no, clinical review required
8. Will another opioid antagonist be coadministered with the requested medication?
    a. If yes, clinical review required
    b. If no, continue to #9
9. Does the member have known or suspected gastrointestinal obstruction or is at increased risk of recurrent obstruction?
    a. If yes, clinical review required
    b. If no, continue to #10
10. Has the member tried agents, Movantik (naloxegol) or Symproic (naldemedine), with inadequate response or has a contraindication to use of either? (Provide supporting documentation)
    a. If yes, approve for 4 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member still on chronic opioid therapy that is per the FDA-approved indications? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required

Last Reviewed: 12/19/18, 7/17/19, 5/19/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 9/1/19, 6/15/21, 6/1/23, 4/15/24

3. Is the indication for OIC associated with advanced illness OR active cancer pain and under palliative care? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #4
4. Is the indication for OIC associated with chronic non-cancer pain? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has beneficial response to requested medication (i.e. increased number of bowel movements from baseline) been documented by the prescriber? (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Relistor (methylnaltrexone) [package insert]. Tarrytown, NY: Progenics Pharmaceuticals, Inc.; 2020
2. Camilleri M, Lembo A, Katzka D. Opioids in Gastroenterology: Treating Adverse Effects and Creating Therapeutic Benefits. Clin Gastroenterol Hepatol. 2017;15(9):1338-1349. https://doi.org/10.1016/j.cgh.2017.05.014
3. Davies, Andrew, et al. "MASCC recommendations on the management of constipation in patients with advanced cancer." *Supportive Care in Cancer* 28.1 (2020): 23-33.

Last Reviewed: 12/19/18, 7/17/19, 5/19/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 9/1/19, 6/15/21, 6/1/23, 4/15/24

Rezurock® (belumosudil)
Prior Authorization Guidelines

Affected Medication(s)

- Rezurock oral tablet

FDA Approved Indication(s)

- Adult and pediatric patients 12 years and older with chronic graft-versus-host disease after failure of at least two prior lines of systemic therapy

Dosing

- 200mg orally once daily

Initial Authorization Criteria

1. Has the requested medication previously been approved by OHSU for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a diagnosis of chronic graft vs host disease? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member 12 years of age or older?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a documented trial with inadequate response to systemic steroids? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have documentation of an inadequate response, intolerance, or contraindication to at least TWO of the following: ruxolitinib, tacrolimus, cyclosporine, ibrutinib, imatinib, methotrexate, sirolimus, or mycophenolate mofetil? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the medication prescribed by, or in consultation with, a provider specializing in transplant or oncology?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Last Reviewed: 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/22, 3/1/24

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within 1 year) with documentation of stability or improvement in chronic graft vs host disease? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a provider specializing in transplant or an oncologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. REZUROCK (belumosudil) tablets. Warrendale, PA 15086; Kadmon Pharmaceuticals, LLC. 2021.
2. Drugs@FDA: FDA Approved Drug Products. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 24 August. 2021].
3. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Hematopoietic Cell Transplant. Version 3.2023 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/pdf/hct. Accessed November 28, 2023.
4. Cutler, Corey, et al. "Belumosudil for chronic graft-versus-host disease (cGVHD) after 2 or more prior lines of therapy: the ROCKstar Study." Blood (2021).

Last Reviewed: 11/17/21, 3/17/23, 1/19/24
Effective Date: 1/1/22, 3/1/24

Rivfloza™ (nedosiran)
Prior Authorization Guidelines

Affected Medication(s)

- Rivfloza (nedosiran) subcutaneous injection

FDA Approved Indication(s)

- To lower urinary oxalate levels in children 9 years of age and older and adults with primary hyperoxaluria type 1 (PH1) and relatively preserved kidney function

Dosing

- < 50 kg: 128 once monthly
- ≥ 50 kg: 160 mg once monthly

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a documented diagnosis of primary hyperoxaluria type 1 as confirmed by genetic testing or biopsy? (Provide supporting documentation of diagnosis)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member 9 years of age or older? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member had a liver transplant?
    a. If yes, clinical review required
    b. If no, continue to #6
6. Does the patient have documentation of estimated glomerular filtration rate (eGFR) of 30 mL/min/1.73m2 or greater? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is Rivfloza (nedosiran) being prescribed by, or in consult with, a specialist in genetics, nephrology, or urology?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Last Reviewed: 3/15/24
Effective Date: 4/15/24

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within past year) provided with documentation of significant clinical response to therapy received? (ex. decrease in urinary oxalate excretion from baseline, reduction in spot urinary oxalate:creatinine ratio from baseline, stabilization of GFR)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is Rivfloza (nedosiran) being prescribed by, or in consult with, a specialist in genetics, nephrology, or urology?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. RIVFLOZA (nedosiran) subcutaneous injection, [package insert]. Costa Mesa, CA: Pyramid Laboratories; 2024.
2. Drugs@FDA: FDA Approved Drug Products. 2024. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 6 Feb. 2024].
3. Groothoff JW, Metry E, Deesker L, et al. Clinical practice recommendations for primary hyperoxaluria: an expert consensus statement from ERKNet and OxalEurope. Nat Rev Nephrol. 2023;19(3):194-211.
4. Baum MA, Langman C, Cochat P, et al. PHYOX2: a pivotal randomized study of nedosiran in primary hyperoxaluria type 1 or 2. Kidney Int. 2023;103(1):207-217.

Last Reviewed: 3/15/24
Effective Date: 4/15/24

Sabril® (vigabatrin)
Prior Authorization Guidelines

Affected Medication(s)

- Sabril oral tablet
- Sabril oral packet
- Vigabatrin oral tablet
- Vigabatrin oral packet
- Vigadrone oral packet
- Vigpoder oral packet

FDA Approved Indication(s)

- Adjunctive therapy in patients 2 years of age or older with refectory complex partial seizures who had an inadequate response to several alternative treatments
- As monotherapy in infants 1 month to 2 years of age with infantile spasms for whom the potential benefits outweigh the potential risk of vision loss

Dosing

Refractory Complex Partial Seizures:

- Pediatric (≥2 years of age to adolescents ≤16 years):
  - 10 to 15kg: 175mg twice daily initially, maintenance dose 525mg twice daily
  - >15 to 20kg: 225mg twice daily initially, maintenance dose 650mg twice daily
  - >20 to 25kg: 250mg twice daily initially, maintenance dose 750mg twice daily
  - >25kg to 60kg: 250mg twice daily initially, maintenance dose 1000mg twice daily
- Adolescents ≤16 years and weighing >60 kg or Adolescents ≥17 years:
  - 500 mg twice daily initially, maintenance 1,500 mg twice daily

Infantile Spasms: maximum daily dose of 150 mg/kg

Initial Authorization Criteria

1. Is the request for continuation of vigabatrin therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Review submitted diagnosis and verify criteria below:
   a. Refractory complex partial seizures, continue to corresponding criteria
   b. Infantile Spasm, continue to corresponding criteria
   c. Other indication, clinical review required

Refractory Complex Partial Seizures

1. Did the member have inadequate seizure control with at least TWO of the following anticonvulsants in the past: felbamate, lamotrigine, levetiracetam, oxcarbazepine, gabapentin, topiramate, tiagabine, zonisamide, or lacosamide? (Provide history of seizure therapy)
   a. If yes, continue to #2
   b. If no, clinical review required

Last Reviewed: 10/3/18, 11/20/19, 7/20/22, 5/19/23, 3/15/24
Effective Date: 1/1/19, 1/1/20, 9/1/22, 7/1/23, 4/15/24

2. Is documentation provided that indicates potential benefits from treatment outweigh the risk of vision loss? (Documentation must confirm member was educated on the risk of vision loss)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Was a baseline vision assessment completed?

    a. If yes, continue #4
    b. If no, clinical review required

4. Is the medication being prescribed by or in consultation with a neurologist who is certified with the Vigabatrin REMS program?

    a. If yes, approve for 12 months, unless otherwise specified
    b. If no, clinical review required

Infantile Spasms

1. Does documentation indicate potential benefits from treatment outweigh the risk of vision loss? (Documentation must confirm that member's parent or guardian was educated on the risk of vision loss)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the medication being prescribed by or in consultation with a neurologist who is certified with the Vigabatrin REMS program?

    a. If yes, approve for 2 months, unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is vigabatrin being prescribed for an FDA-approved indication or major compendia supported use? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Were updated chart notes provided (within 1 year) of clinical response to therapy with documentation of a routine vision assessment performed every 3 months? (Provide documentation of reduction in seizures/infantile spasms)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the medication being prescribed by or in consultation with a neurologist who is certified with the Vigabatrin REMS program?

    a. If yes, approve for 12 months, unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as

medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Sabril (vigabatrin) [Prescribing Information]. Deerfield, IL: Lundbeck. October 2021.
2. Sabril. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com.
3. Vigadrone [package insert]. Maple Grove, MC: Upsher-Smith Laboratories, LLC; February 2020.
4. Glaze, MD. Management and prognosis of infantile spasms. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com.
5. National Institute for Health and Care Excellence (NICE): Epilepsies: diagnosis and management. National Institute for Health and Care Excellence (NICE). London, United Kingdom. Available at: https://www.nice.org.uk/guidance/cg137/resources/epilepsies-diagnosis-and-management-35109515407813.

Last Reviewed: 10/3/18, 11/20/19, 7/20/22, 5/19/23, 3/15/24
Effective Date: 1/1/19, 1/1/20, 9/1/22, 7/1/23, 4/15/24

Samsca® (tolvaptan)
Prior Authorization Guidelines

Affected Medication(s)

- Samsca oral tablet
- tolvaptan oral tablet

FDA Approved Indication(s)

- Treatment of clinically significant hypervolemic and euvolemic hyponatremia (serum sodium <125 mEq/L or less marked hyponatremia that is symptomatic and has resisted correction with fluid restriction), including patients with heart failure and Syndrome of Inappropriate Antidiuretic Hormone (SIADH)

Dosing

- Initially: 15mg once daily
- Titrate as needed to a maximum of 60 mg once daily
- Do not administer for more than 30 days to minimize risk of liver injury

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a serum sodium level <125 mEq/L? (Provide serum sodium level for review)
    a. If yes, continue to #5
    b. If no, continue to #4
4. Does the member have symptomatic hyponatremia despite fluid restriction of <1000mL/day? (i.e. lethargy, weakness, irritability) (Provide documentation of fluid restriction)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Are drugs known to potentially cause SIADH being reviewed or discontinued when appropriate? (i.e. chlorpropamide, SSRIs, TCAs, carbamazepine, vincristine, nicotine, NSAIDs, etc.) (Provide documentation of medication reconciliation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have any of the following contraindications to therapy with Samsca (tolvaptan)?
    - Autosomal Dominant Polycystic Kidney Disease (ADPKD)
    - Urgent Need to Raise Serum Sodium Acutely

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Effective Date: 1/1/19, 8/15/20, 3/1/24

- Inability to Sense or Appropriately Respond to Thirst
- Hypovolemic Hyponatremia
- Concomitant Use of Strong CYP 3A Inhibitors
- Anuria

a. If yes, clinical review required
b. If no, continue to #7

7. Is the treatment being prescribed by, or in consultation with, an endocrinologist or nephrologist?

a. If yes, approve for 1 month
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Samsca (tolvaptan) [package insert]. Rockville, MD: Otsuka America Pharmaceuticals; November 2022.
2. Verbalis JG, Goldsmith SR, Greenberg A, et al. Hyponatremia treatment guidelines 2007: expert panel recommendations. Am J Med 2007;120(11 Suppl 1):S1-S21.
3. Schrier RW, Gross P, Gheorghiade M, et al. Tolvaptan, a selective oral vasopressin V2-receptor antagonist, for hyponatremia. N Engl J Med 2006;355(20):2099-2112.
4. Verbalis, Joseph G., et al. "Diagnosis, evaluation, and treatment of hyponatremia: expert panel recommendations." The American journal of medicine 126.10 (2013): S1-S42.

Last Reviewed: 11/21/18, 7/15/20, 11/18/20, 1/19/22, 1/20/23, 1/19/24
Effective Date: 1/1/19, 8/15/20, 3/1/24

Scemblix® (asciminib)
Prior Authorization Guidelines

Affected Medication(s)

- Scemblix oral tablet

FDA Approved Indication(s)

- Treatment of adults with Philadelphia chromosome-positive chronic myeloid leukemia (Ph+CML) in chronic phase (CP) that meet one of the following:
  - Are previously treated with two or more tyrosine kinase inhibitors (TKIs)
  - Have the T315I mutation

Dosing

- For patients previously treated with two or more tyrosine kinase inhibitors (TKIs): 80mg by mouth once daily, or 40mg orally twice daily
- For patients with the T315I mutation: 200mg twice daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with Scemblix (asciminib) medication?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the medication being requested for an FDA approved indication? (Provide supporting documentation)
   a. If yes, continue to #4
   b. If no, continue to #3
3. Is the medication being requested for an indication supported by the National Comprehensive Cancer Network (NCCN) recommendation with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
   a. If yes, continue to #5
   b. If no, clinical review required
5. Does the member have T315I mutation? (Provide supporting documentation)
   a. If yes, continue to #6
   b. If no, continue to #8
6. Has the member previously trialed Iclusig with an inadequate response or clinically significant adverse effect? (Provide supporting documentation)
   a. If yes, continue to #8

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Effective Date: 3/1/22

b. If no, continue to #7

7. Does the member have a contraindication to treatment with Iclusig?

   a. If yes, continue to #8
   b. If no, clinical review required

8. Is the medication being prescribed by, or in consultation with, an oncologist?

   a. If yes, approve for 4 months
   b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA or supported by NCCN recommendation with an evidence level of 2A or higher? (Provide supporting documentation)

   a. If yes, continue to #2
   b. If no, clinical review required

2. Is there clinical documentation confirming disease responsiveness to therapy provided? (Example: BCR-ABL1 ≤ 0.1%) (Provide supporting documentation)

   a. If yes, continue to #3
   b. If no, clinical review required

3. Is the medication being prescribed by or in consultation with an oncologist?

   a. If yes, approve for 12 months
   a. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. SCEMBLIX (asciminib) tablets, [package insert]. East Hanover, NJ: Novartis Pharmaceuticals; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 9 Dec. 2021].
3. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Chronic Myeloid Leukemia. Version 2.2024 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/pdf/cml.pdf. Accessed February 12, 2024.

Sexual Dysfunction Agents
Prior Authorization Guidelines

Affected Medication(s)

- Addyi (flibanserin) oral tablet
- Caverject (alprostadil) powder for reconstitution and intracavernosal injection
- Cialis (tadalafil) oral tablet
- Tadalafil oral tablet
- Edex (alprostadil) injection
- Vardenafil oral tablet
- Muse (alprostadil) uretheral suppository
- Vardenafil orally disintegrating tablet
- Stendra (avanadil) oral tablet
- Viagra (sildenafil) oral tablet
- Sildenafil oral tablet
- Vyleesi (bremelanotide) subcutaneous solution

FDA Approved Indication(s)

- For the treatment of premenopausal women with acquired, generalized hypoactive sexual desire disorder (HSDD), as characterized by low sexual desire that causes marked distress or interpersonal difficulty and is NOT due to a co-existing medical or psychiatric condition, problems within the relationship, or the effects of a medication or other drug substance
  - Addyi, Vyleesi
- For the treatment of erectile dysfunction
  - Caverject, Edex, Cialis, Sildenafil, Muse, Stendra, Tadalafil, Vardenafil, Viagra
- Adjunct to other diagnostic tests in the diagnosis of erectile dysfunction (ED)
  - Caverject
- For the treatment of signs and symptoms of benign prostatic hyperplasia (BPH)
  - Cialis, Tadalafil
- Treatment of erectile dysfunction and the signs and symptoms of benign prostatic hyperplasia (ED/BPH)
  - Cialis, Tadalafil

Dosing

- Refer to corresponding package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same agent used for the same indication?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required

Last Reviewed: 12/19/18, 7/17/19, 3/18/20, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 5/1/20, 1/1/23, 12/20/23

3. Which diagnosis is the medication being requested for?
    a. Erectile disorder, continue to corresponding criteria
    b. Female sexual interest/arousal disorder, continue to corresponding criteria
    c. As an adjunct to other diagnostic tests for diagnosis of erectile dysfunction, approve for one dose
    d. Benign prostatic hyperplasia, continue to corresponding criteria
    e. Other indication, continue to corresponding criteria

Erectile Disorder

1. Does the member experience one or more of the following symptoms on at least 75% of sexual activity for a duration of 6 months or greater? (Provide supporting documentation)

    - Marked difficulty in obtaining an erection during sexual activity
    - Marked difficulty in maintaining an erection until the completion of sexual activity
    - Marked decrease in erectile rigidity

    a. If yes, continue to #2
    b. If no, clinical review required
2. Do the above symptoms cause the member clinically significant distress? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Have other causes of sexual dysfunction been ruled out such as nonsexual mental disorder, severe relationship distress, other significant stressor, substance/medication side effect, and/or other medical condition (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Female Sexual Interest/ Arousal Disorder

1. Does the member demonstrate a lack of, or significantly reduced, sexual interest/arousal as manifested by at least 3 of the following? (Provide supporting documentation)

    - Absent/reduced interest in sexual activity
    - Absent/reduced sexual/erotic thoughts or fantasies
    - No/reduced initiation of sexual activity, and typically unreceptive to a partner's attempts to initiate
    - Absent/reduced sexual excitement/pleasure during sexual activity in almost all or all (approximately 75%–100%) sexual encounters (in identified situational contexts or, if generalized, in all contexts)
    - Absent/reduced sexual interest/arousal in response to any internal or external sexual/erotic cues (e.g., written, verbal, visual)
    - Absent/reduced genital or non-genital sensations during sexual activity in almost all or all (approximately 75%–100%) sexual encounters (in identified situational contexts or, if generalized, in all contexts)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Have the above symptoms persisted for a duration of 6 months or longer? (Provide supporting documentation of symptom duration)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Do the member's sexual symptoms cause the member clinically significant distress? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Have other causes of sexual dysfunction been ruled out such as nonsexual mental disorder, severe relationship distress, other significant stressor, substance/medication side effect, and/or other medical condition? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Benign Prostatic Hyperplasia

1. Does the member have documentation of a trial with inadequate response or intolerance to at least one alpha-adrenergic blocker (tamsulosin, doxazosin, terazosin, alfuzosin) AND one 5-alpha reductase inhibitor (finasteride or dutasteride)? (Provide supporting documentation of trials with inadequate response, intolerances, or contraindications)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Other Indications

1. Has the member tried and had an inadequate response OR does the member have a contraindication to ALL standard treatment options for the requested indication? (Provide all prior treatment history, contraindication if appropriate, and treatment plan)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member demonstrate a positive clinical response from therapy as defined by one of the below? (Provide supporting documentation)
    - For erectile disorder: Improvement in obtaining and maintaining an erection
    - For female sexual interest/arousal disorder: Improvement in sexual interest and/or sexual arousal
    - For benign prostatic hyperplasia: Improvement in urinary frequency, urinary urgency, nocturia, and/or incomplete emptying

Last Reviewed: 12/19/18, 7/17/19, 3/18/20, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 5/1/20, 1/1/23, 12/20/23

- For other indications: Clinical documentation confirming disease responsiveness to therapy provided

a. If yes, approve for 12 months unless otherwise specified
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Addyi (flibanserin) [Prescribing Information]. Bridgewater, NJ: Sprout Pharmaceuticals, Inc. September 2021.
2. Caverject (alprostadil) [Prescribing Information]. New York, NY: Pharmacia and Upjohn Company LLC. March 2019.
3. Muse (alprostadil) [Prescribing Information]. Somerset, NJ: Meda Pharmaceuticals. April 2018.
4. Stendra (avanafil) [Prescribing Information]. Cranford, NJ: Mist Pharmaceuticals, LLC. November 2021.
5. Cialis (tadalafil) [Prescribing Information]. Indianapolis, IN CT: Eli Lilly and Company. April 2022.
6. Viagra (sildenafil) [Prescribing Information]. New York, NY: Pfizer Laboratories Div Pfizer Inc. July 2021.
7. Sexual Dysfunctions, Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition. May 2013
8. McVary KT, Roehrborn CG, Avins AL, et al. Update on AUA guideline on the management of benign prostatic hyperplasia. J Urol 2011; 185:1793.
9. Burnett AL, Nehra A, Breau RH et al: Erectile dysfunction: AUA guideline. J Urol 2018; 200: 633.

Last Reviewed: 12/19/18, 7/17/19, 3/18/20, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 5/1/20, 1/1/23, 12/20/23

Signifor® (pasireotide)
Prior Authorization Guidelines

Affected Medication(s)

- Signifor subcutaneous solution

FDA Approved Indication(s)

- For treatment of adult patients with Cushing's disease for whom pituitary surgery is not an option or has not been curative

Dosing

- 0.3 to 0.9 mg by subcutaneous injection twice a day

Initial Authorization Criteria

1. Is the request for continuation of Signifor (pasireotide) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Did the member either have pituitary surgery that was not curative or is the member not a candidate for surgery? (Provide supporting documentation confirming persistent hypercortisolism)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the requested medication being prescribed or in consultation with an endocrinologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member demonstrate a positive clinical response to therapy as defined by a reduction in 24-hour urinary free cortisol levels or improvement in signs and symptoms of Cushing's disease? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the requested medication being prescribed or in consultation with an endocrinologist?
    a. If yes, approve for 12 months unless otherwise specified

Last Reviewed: 11/7/18, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19

b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Signifor [package insert]. East Hanover, NJ: Novartis Pharmaceuticals Corporation; September 2020.
2. Biller BMK, Grossman AB, Steward PM, et al. Treatment of adrenocorticotropin-dependent Cushing's syndrome: a consensus statement. J Clin Endocrinol Met. 2008;93(7):2454-2462.
3. Nieman LK, Biller BMK, Findling JW, et al. The diagnosis of Cushing's syndrome: An Endocrine Society Practice Guideline. J Clin Endocrinol Metab. 2008;93(5):1526–1540.
4. Nieman LK, Biller BMK, Findling JW, et al. Treatment of Cushing's syndrome: An Endocrine Society Clinical Practice Guideline. The J Clin Endocrinol Metab. 2015;100(8):2807-2831.

Last Reviewed: 11/7/18, 7/21/21, 11/16/22, 10/15/2023, 11/17/23
Effective Date: 1/1/19, 11/18/2023

Sirturo® (bedaquiline fumarate), Pretomanid
Prior Authorization Guidelines

Affected Medication(s)

- Sirturo (bedaquiline) oral tablet
- Pretomanid oral tablet

FDA Approved Indication(s)

- Sirturo: As part of combination therapy in the treatment of pediatric patients ≥5 years of age (weighing ≥15 kg) and adults with pulmonary multi-drug resistant tuberculosis (MDR-TB). Reserve for use when an effective treatment regimen cannot otherwise be provided
- Pretomanid: Part of a combination regimen with bedaquiline and linezolid for the treatment of adults with pulmonary extensively drug resistant (XDR), treatment-intolerant or nonresponsive multidrug-resistant (MDR) tuberculosis (TB)

Dosing

Sirturo

- Children ≥5 years and Adolescents:
  Weight 15 kg to <30 kg:
  Weeks 1 and 2: Oral: 200 mg once daily.
  Weeks 3 to 24: Oral: 100 mg 3 times weekly (Total weekly dose: 300 mg/week)

  Weight ≥30 kg:
  Weeks 1 and 2: Oral: 400 mg once daily.
  Weeks 3 to 24: Oral: 200 mg 3 times weekly (Total weekly dose: 600 mg/week)

- Adults: 400 mg orally once daily for the first two weeks, followed by 200 mg orally three times per week (with at least 48 hours between doses) for 22 weeks (total duration of 24 weeks)

Pretomanid

- 200 mg orally once daily for 26 weeks (in combination with bedaquiline and linezolid)

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #2
   b. If no, clinical review required
2. Does the patient have documentation of resistance to, intolerance to, or contraindication to quad therapy with isoniazid, rifampin, ethambutol, pyrazinamide? (Provide supporting documentation)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the request for one of the following treatment regimens? (Provide documentation of susceptible isolate and planned treatment regimen)

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Effective Date: 1/1/19,8/15/20, 11/1/21, 4/15/24

- Pretomanid + Sirturo + linezolid
- Sirturo + at least 3 additional antituberculotic agents active against member's *M. tuberculosis* isolate

a. If yes, continue to #4
b. If no, clinical review required

4. Is the treatment being prescribed by, or in consultation with, an infectious disease specialist or pulmonologist?

a. If yes, approve for up to 6 months
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Sirturo [package insert]. Titusville, NJ: Janssen Products LP.October 2021.
2. Pretomanid (pretomanid) oral tablet [package insert]. New York, NY: Mylan Laboratories; April 2020.
3. American Thoracic Society, Centers for Disease Control and Prevention, and Infectious Diseases Society of America. Treatment of Tuberculosis. MMWR Morb Mortal Wkly Rep 2003; 52(RR-11):1-80.
4. World Health Organization. WHO treatment guidelines for drug-resistant tuberculosis: 2016 update. URL: who.int/tb/areas-of-work/drug-resistant-tb/MDRTBguidelines2016.pdf. Available from Internet. Accessed 2018 November 5.
5. Nahid, Payam, et al. "Treatment of drug-resistant tuberculosis. An official ATS/CDC/ERS/IDSA clinical practice guideline." American journal of respiratory and critical care medicine 200.10 (2019): e93-e142.

Last Reviewed: 11/21/18, 7/15/20, 9/15/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 8/15/20, 11/1/21, 4/15/24

Skyclarys® (omaveloxolone)
Prior Authorization Guidelines

Affected Medication(s)

- Skyclarys (omaveloxolone) oral tablet

FDA Approved Indication(s)

- Treatment of Friedreich's ataxia (FA) in adults and adolescents aged 16 and older

Dosing

- 150 mg orally once daily

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is Skyclarys® (omaveloxolone) being requested for an FDA approved indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member between ages 16 and 40?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member Friedreich's ataxia as confirmed by genetic testing? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a stable modified Friedreich's Ataxia Rating Scale (mFARS) score between 20 and 80? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a left ventricular ejection fraction of at least 40%? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the treatment being prescribed by, or in consultation with, a clinical geneticist or neurologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA? (Provide supporting documentation)
    a. If yes, continue to #2

Last Reviewed: 5/19/23
Effective Date: 7/15/23

b. If no, clinical review required

2. Were updated chart notes (within past year) provided with documentation of clinical response to prior therapy received?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a clinical geneticist or neurologist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Skyclarys (omaveloxolone) capsules, [package insert]. Plano, TX: Reata Pharmaceuticals, Inc.; 2023.
2. Drugs@FDA: FDA Approved Drug Products. 2023. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 22 Mar. 2023].
3. Lynch, D. R., Chin, M. P., Delatycki, M. B., Subramony, S. H., Corti, M., Hoyle, J. C., Boesch, S., Nachbauer, W., Mariotti, C., Mathews, K. D., Giunti, P., Wilmot, G., Zesiewicz, T., Perlman, S., Goldsberry, A., O'Grady, M., & Meyer, C. J. (2021). Safety and Efficacy of Omaveloxolone in Friedreich Ataxia (MOXIe Study). Annals of neurology, 89(2), 212–225.
4. Lynch, D. R., Chin, M. P., Boesch, S., Delatycki, M. B., Giunti, P., Goldsberry, A., Hoyle, J. C., Mariotti, C., Mathews, K. D., Nachbauer, W., O'Grady, M., Perlman, S., Subramony, S. H., Wilmot, G., Zesiewicz, T., & Meyer, C. J. (2023). Efficacy of Omaveloxolone in Friedreich's Ataxia: Delayed-Start Analysis of the MOXIe Extension. Movement disorders : official journal of the Movement Disorder Society, 38(2), 313–320.
5. Corben, L. A., Collins, V., Milne, S., Farmer, J., Musheno, A., Lynch, D., Subramony, S., Pandolfo, M., Schulz, J. B., Lin, K., Delatycki, M. B., & Clinical Management Guidelines Writing Group (2022). Clinical management guidelines for Friedreich ataxia: best practice in rare diseases. Orphanet journal of rare diseases, 17(1), 415.

Sodium phenylbutyrate and Glycerol phenylbutyrate
Prior Authorization Guidelines

Affected Medication(s)

- Buphenyl oral powder
- Buphenyl oral tablet
- Sodium phenylbutyrate oral powder
- Sodium phenylbutyrate oral tablet
- Olpruva for oral suspension
- Pheburane oral pellets
- Ravicti oral solution

FDA Approved Indication(s)

Buphenyl:

- Adjunctive therapy in the chronic management of patients with urea cycle disorders involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS)
    - Indicated in all patients with neonatal-onset deficiency (complete enzymatic deficiency, presenting within the first 28 days of life)
    - Indicated in patients with late-onset disease (partial enzymatic deficiency, presenting after the first month of life) who have a history of hyperammonemic encephalopathy

Olpruva:

- Adjunctive therapy in the chronic management of adult and pediatric patients weighing 20 kg or greater with a BSA of 1.2 m^2 or greater, with urea cycle disorders involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS)

Pheburane:

- Adjunctive therapy in the chronic management of patients with urea cycle disorders involving deficiencies of carbamylphosphate synthetase (CPS), ornithine transcarbamylase (OTC), or argininosuccinic acid synthetase (AS)

Ravicti:

- Chronic management of patients 2 months of age and older with urea cycle disorders (UCDs) who cannot be managed by dietary protein restriction and/or amino acid supplementation alone
    - Must be used with dietary protein restriction and, in some cases, dietary supplements (e.g., essential amino acids, arginine, citrulline, protein-free calorie supplements)

Dosing

- Refer to drug specific package insert for dosing recommendations

Initial Authorization Criteria

a. Has the requested medication previously been approved by OHSU for the same indication?If yes, continue to Reauthorization
b. If no, continue to #2

Last Reviewed: 11/21/18, 1/20/21, 3/16/22, 3/15/23, 5/19/23, 7/21/23
Effective Date: 1/1/19, 3/1/21, 4/15/22, 9/15/23

2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the diagnosis confirmed by blood, enzyme, or genetic testing? (Provide lab result for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the baseline plasma ammonia level provided? (Provide lab result for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has the member tried a protein restrictive diet and has the member remained adherent to the protein restrictive diet? (Provide documentation of restrictive diet)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the requested medication Olpruva, Pheburane, or Ravicti (glycerol phenylbutyrate)?
    a. If yes, continue to #7
    b. If no, continue to #8
7. Does the member have a previous trial with inadequate response, intolerance, or contraindication to sodium phenylbutyrate? (Provide documentation of trial, intolerance, or contraindication)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Is the treatment being initiated by a provider that specializes in the treatment of inherited metabolic disorders? (Examples include a medical geneticist or an endocrinologist)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member adherent to a protein restrictive diet? (Provide documentation of restrictive diet)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member show a positive clinical response to therapy as defined by normalized plasma ammonia levels? (Provide documentation of normalized plasma ammonia levels)
    a. If yes, continue to #4

Last Reviewed: 11/21/18, 1/20/21, 3/16/22, 5/19/23, 7/21/23
Effective Date: 1/1/19, 3/1/21, 4/15/22, 9/15/23

b. If no, clinical review required

4. Is the treatment being initiated by a provider that specializes in the treatment of inherited metabolic disorders? (Examples include a medical geneticist or an endocrinologist)

   a. If yes, approve for 12 months unless otherwise specified
   b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Buphenyl (sodium phenylbutyrate) [Prescribing Information]. Lake Forest, IL: Horizon Pharma, Inc. May 2021.
2. Ravicti (glycerol phenylbutyrate) [Prescribing Information]. Lake Forest, IL: Horizon therapeutics, LLC. September 2021.
3. Olpruva (sodium phenylbutyrate kit) [Prescribing Information]. Newton, MA: Acer Therapeutics Inc. December 2022.
4. Pheburane (sodium phenybutyrate pellet) [Prescribing Information]. Bryn Mawr, PA: Medunik USA, Inc. September 2022.
5. Lee, B. Urea cycle disorder: clinical features and diagnosis. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed November 2018.
6. Lee, B. Urea cycle disorder: management. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed November 2018.

Last Reviewed: 11/21/18, 1/20/21, 3/16/22, 5/19/23, 7/21/23
Effective Date: 1/1/19, 3/1/21, 4/15/22, 9/15/23

Sohonos® (palovarotene)
Prior Authorization Guidelines

Affected Medication(s)

- Sohonos (palovarotene) oral capsule

FDA Approved Indication(s)

- For the reduction in volume of new heterotopic ossification (HO) in adults and pediatric patients aged 8 years and older for females and 10 years and older for males with fibrodysplasia ossificans progressiva (FOP)

Dosing

- Refer to package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation Sohonos (palovarotene) therapy for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #3
2. Is Sohonos® (palovorotene) being requested for an FDA approved indication? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
3. Is the member at least 8 years of age for females or at least 10 years of age for males?
    a. If yes, continue to #5
    b. If no, clinical review required
4. Does the member have documentation of FOP diagnosis, with the R206H ACVR1 mutation or other FOP variants reported to be associated with progressive heterotopic ossification? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
5. Is the treatment dose appropriate?
    a. If yes, continue to #7
    b. If no, clinical review required
6. Is the treatment being prescribed by, or in consultation with, an endocrinologist or appropriate specialist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required

Last Reviewed: 11/17/23
Effective Date: 12/20/23

2. Were updated chart notes (within past year) provided with documentation of clinical response to prior therapy received?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, an endocrinologist or appropriate specialist?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. SOHONOS (palovarotene) capsules [package insert]. Cambridge, MA: Ipsen Biopharmaceuticals, Inc; 2023.
2. Drugs@FDA: FDA Approved Drug Products. 2022. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 21 Sept. 2023].
3. Pignolo RJ, Hsiao EC, Al Mukaddam M, et al. Reduction of New Heterotopic Ossification (HO) in the Open-Label, Phase 3 MOVE Trial of Palovarotene for Fibrodysplasia Ossificans Progressiva (FOP). J Bone Miner Res. 2023;38(3):381-394.
4. Kaplan FS, et al. The medical management of fibrodysplasia ossificans progressiva: current treatment considerations. Proc Intl Clin Council FOP 2. 2022: 1-127.

Somavert® (pegvisomant)
Prior Authorization Guidelines

Affected Medication(s)

- Somavert subcutaneous powder for solution

FDA Approved Indication(s)

- Treatment of acromegaly in patients who have had an inadequate response to surgery or radiation therapy, or for whom these therapies are not appropriate

Dosing

- Loading Dose: 40 mg subcutaneously
- Maintenance Dose: 10 mg subcutaneously once daily, starting the day after loading dose administration

Initial Authorization Criteria

1. Is the request for continuation of Somavert (pegvisomant) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have an elevated insulin like growth factor-1(IGF-1) level for age and gender? (Provide baseline IGF-1 level for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a trial with inadequate response to, or documentation supporting they are not a candidate for either surgery or radiation therapy? (Provide documentation of inadequate response or rationale for avoiding therapy)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a trial with inadequate response to, intolerance, or contraindication to a first generation somatostatin receptor ligand (i.e. octreotide)? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a trial with inadequate response to, intolerance, or contraindication to cabergoline? (Note: cabergoline is only indicated if IGF1 <2.5 times the upper limit of normal) (Provide supporting documentation)
    a. If yes, continue to #7

Last Reviewed: 11/21/18, 7/21/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 4/15/24

b. If no, clinical review required

7. Is the treatment being prescribed by, or in consultation with, an endocrinologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member had a positive clinical response to therapy as defined as either a decrease in or normalization in insulin like growth factor for age and gender (IGF-1)? (Provide documentation of IGF-1 value for review)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, an endocrinologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Somavert (pegvisomant) Product Information, Pfizer. New York, NY. December 2017.
2. AACE Medical Guidelines for Clinical Practice for the Diagnosis and Treatment of Acromegaly, May/June.2004. http://www.aace.com/pub/pdf/guidelines/AcromegalyGuidelines2004.pdf.
3. Melmed S, Colao A, Molitch M, et al. Guidelines for Acromegaly Management: An Update. J Clin Endocrinol Metab. 2009;94:1509-1517.
4. Katznelson, Laurence, et al. "Acromegaly: an endocrine society clinical practice guideline." The Journal of Clinical Endocrinology & Metabolism 99.11 (2014): 3933-3951.
5. Melmed, Shlomo, et al. "A consensus statement on acromegaly therapeutic outcomes." Nature Reviews Endocrinology 14.9 (2018): 552-561.

Last Reviewed: 11/21/18, 7/21/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 4/15/24

Spravato® (esketamine)
Prior Authorization Guidelines

Affected Medication(s)

- Spravato nasal spray

FDA Approved Indication(s)

- For treatment-resistant depression (TRD) in adults in conjunction with an oral antidepressant
- Depressive symptoms in adults with major depressive disorder (MDD) with acute suicidal ideation or behavior

NOTE: The use of Spravato beyond 4 weeks has not been systematically evaluated in the treatment of depressive symptoms in patients with MDD with acute suicidal ideation or behavior

Dosing

Treatment-resistant depression (TRD):

- Induction: 56 mg twice weekly; may increase to 84 mg twice weekly based on efficacy and tolerability
- Maintenance: Starting week 5, continue previous administered dose (56 mg or 84 mg) and decrease dosing frequency to once weekly. Starting week 9 and after, continue established dose (56 mg or 84 mg) and adjust dosing to the least frequency to maintain remission/response; once weekly or once every 2 weeks

Major depressive disorder (MDD) with acute suicidal ideation or behavior:

- 84 mg twice weekly for 4 weeks, may be reduced to 56 mg twice per week based on tolerability. Use beyond 4 weeks has not been systematically evaluated in the treatment of depressive symptoms in patients with MDD with suicidal ideation or behavior.

NOTE: Spravato must be administered under the direct supervision of a healthcare provider

Initial Authorization Criteria

1. Is the request for continuation of Spravato (esketamine) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. What is the diagnosis that Spravato (esketamine) is being used for? (Provide documentation of diagnosis)
    a. Treatment-resistant depression (TRD), continue to #5
    b. Major depressive disorder (MDD) with acute suicidal ideation or behavior, continue to #7
    c. Other indication, clinical review required

Last Reviewed: 9/18/2019, 11/18/20, 1/19/22, 3/17/23, 9/15/2023
Effective Date: 10/15/2019, 1/15/21, 3/1/22, 11/20/23

5. Does the member have a previous trial with inadequate response to at least two antidepressants from at least two different classes when titrated up to the maximum indicated doses for a trial lasting a minimum of 8 weeks OR a contraindication to all classes? (Examples: SSRI, SNRI, TCA, bupropion) (Provide supporting documentation)

    a. If yes, continue to #6
    b. If no, clinical review required

6. Does the member have a previous trial with inadequate response, intolerance, or contraindication to at least two augmentation therapies with trials lasting a minimum of 4 weeks in conjunction with an oral antidepressant? Augmentation therapies include atypical antipsychotics, lithium, or a second antidepressant within an alternative class. (Provide supporting documentation)

    a. If yes, continue to #8
    b. If no, clinical review required

7. Does the member have active suicidal ideation and urgent symptom control is considered necessary? (Provider supporting documentation)

    a. If yes, continue to #8
    b. If no, clinical review required

8. Will Spravato be used in conjunction with an oral antidepressant? (Provide supporting documentation)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Is the treatment being prescribed by, or in consultation with, a mental health specialist and will Spravato be administered under the direct supervision of a healthcare provider?

    a. If yes, approve for the following:
        - Treatment-resistant depression: 6 months
        - Major depressive disorder (MDD) with acute suicidal ideation or behavior: 1 month
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?

    a. If yes, continue to #2
    b. If no, clinical review required

2. What is the diagnosis that Spravato (esketamine) is being used for? (Provide documentation of diagnosis)

    a. Treatment-resistant depression (TRD), continue to #3
    b. Major depressive disorder (MDD) with acute suicidal ideation or behavior, clinical review required
    c. Other indication, clinical review required

3. Were updated chart notes (within the past 12 months) with documentation of a positive response to therapy defined as a decrease in depressive symptoms provided? (Provide supporting documentation)

    a. If yes, continue to #4

b. If no, clinical review required

4. Will Spravato be used in conjunction with an oral antidepressant? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation, with a mental health specialist and will Spravato be administered under the direct supervision of a healthcare provider?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Esketamine. Drugs@FDA: FDA. Approved Drug Products 2019. Accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm?event=overview.process&varApplNo=211243. Accessed August 13, 2019.
2. Lexicomp Online [internet database]. Hudson, OH: Wolters Kluwer Clinical Drug Information, Inc. Updated periodically. Accessed August 13, 2019.
3. Esketamine nasal spray device (Spravato) [package insert]. Titusville, NJ. Janssen Pharmaceuticals, Inc.; July 2020.
4. Gelenberg AJ, Freeman MP, Markowitz JC, et al; American Psychiatric Association; Work Group on Major Depressive Disorder. Practice Guideline for the Treatment of Patients with Major Depressive Disorder, Third Edition. *Am J Psychiatry*. 2010.
5. Spravato (esketamine) REMS website. https://www.spravatorems.com/. Updated March 2019. Accessed August 14, 2019.
6. Daly EJ, Trivedi MH, Janik A, et al. Efficacy of Esketamine Nasal Spray Plus Oral Antidepressant Treatment for Relapse Prevention in Patients With Treatment-Resistant Depression: A Randomized Clinical Trial. *JAMA Psychiatry*. 2019. doi: 10.1001/jamapsychiatry.2019.1189.

Last Reviewed: 9/18/2019, 11/18/20, 1/19/22, 3/17/23, 9/15/2023
Effective Date: 10/15/2019, 1/15/21, 3/1/22, 11/20/23

Sucraid® (sacrosidase)
Prior Authorization Guidelines

Affected Medication(s)

- Sucraid (sacrosidase) oral solution

FDA Approved Indication(s)

- For treatment of genetically determined sucrase deficiency, which is part of congenital sucrase-isomaltase deficiency (CSID)

Dosing

- Patients ≤ 15 kg body weight: 1 mL (1 scoop or 28 drops) orally with each meal or snack
- Patients > 15 kg body weight: 2 mL (2 scoops or 56 drops) orally with each meal or snack

Initial Authorization Criteria

1. Is the request for continuation of Sucraid (sacrosidase) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 5 months of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a diagnosis of congenital sucrose-isomaltase deficiency (CSID) confirmed by one of the following? (Provide supporting documentation)
    - Genetic testing of the sucrase-isomaltase (SI) gene indicates a pathogenic mutation
    - Positive sucrose breath test
    - Small bowel biopsy showing low sucrose activity and normal amounts of other disaccharides
    - Meets all of the following criteria:
        - Stool pH< 6
        - Increase in breath hydrogen of >10ppm when challenged with sucrose after fasting
        - Negative lactose breath test

    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the requested medication being prescribed by, or in consultation with, a gastroenterologist or genetic specialist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 7/21/23
Effective Date: 9/15/23

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member demonstrate a positive clinical response to therapy such as fewer stools or lower total symptom score compared to baseline? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the requested medication being prescribed by, or in consultation with, a gastroenterologist or genetic specialist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Sucraid [package insert]. Vero Beach, FL: QOL Medical, LLC; January 2023.
2. Naim HY, Heine M, Zimmer KP. Congenital sucrose-isomaltase deficiency: Heterogeneity of inheritance, trafficking, and function of an intestinal enzyme complex. J Pediatr Gastroenterol Nutr. 2012;55:S13-S20.
3. Cohen SA. The clinical consequences of sucrose-isomaltase deficiency. Mol Cell Pediatr. 2016;3:5.
4. Gericke B, Amiri M, Scott CR, Naim HY. Molecular pathogenicity of novel sucrose-isomaltase mutations found in congenital sucrose-isomaltase deficiency patients. Biochim Biophys Acta Mol Basis Dis. 2017;1863:817-826.

Last Reviewed: 7/21/23
Effective Date: 9/15/23

Sunosi® (solriamfetol)
Prior Authorization Guidelines

Affected Medication(s)

- Sunosi® 75mg and 150mg oral tablets

FDA Approved Indication(s)

- Excessive Somnolence: Narcolepsy
- Excessive Somnolence: Obstructive sleep apnea

Dosing

- 37.5mg – 150mg once daily

Initial Authorization Criteria

1. Is the request for continuation of Sunosi® (solriamfetol) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment prescribed by or in consultation with a sleep specialist (e.g. neurology, pulmonology)?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has this member's diagnosis been confirmed by overnight polysomnogram, and for narcolepsy a multiple sleep latency test (MSLT)? Note: narcolepsy may be confirmed by low levels of orexin or hypocretin within cerebrospinal fluid (<110pg/mL or less than one third of the normative value of the lab)
    a. If yes, continue to #6
    b. If no, clinical review required
6. What is the underlying condition causing excessive daytime sleepiness?
    a. Narcolepsy, continue to #7
    b. Obstructive sleep apnea, continue to #8
    c. Other, clinical review required

Last Reviewed: 1/20/21, 5/18/22, 7/21/23
Effective Date: 3/1/21

7. Has this member had a documented trial with insufficient response, intolerance, or contraindication to at least one medication in each of the following groups?

    - Group 1: Modafinil or Armodafinil
    - Group 2: Stimulants (e.g. Methylphenidate, dextroamphetamine/amphetamine, etc)

    a. If yes, continue to #10
    b. If no, clinical review required

8. Does this member have evidence of current use or a history of intolerance to a primary treatment for obstructive sleep apnea (e.g. CPAP, mandibular device, surgical intervention)?

    a. If yes, continue to #9
    b. If no, clinical review required

9. Has this member had a documented trial with insufficient response, intolerance, or contraindication to modafinil or armodafinil?

    a. If yes, continue to #10
    b. If no, clinical review required

10. Have all other causes of excessive daytime sleepiness been ruled out or treated (e.g. restless leg syndrome, periodic limb movements, substance abuse, etc.)?

    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Were updated chart notes provided with documentation of significant clinical response to therapy (e.g. reduction in Epworth Sleepiness Scale [ESS])?

    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Morgenthaler TI, Kapur VK, Brown T, et al. Practice parameters for the treatment of narcolepsy and other hypersomnias of central origin. Sleep. 2007;30(12): 1705-1711.
2. Billiard M, Bassetti C, Dauvilliers Y, et al. EFNS guidelines on management of narcolepsy. Eur J Neurol. 2006;13(10):1035-1048.
3. Barateau L, Lopez R, Dauvilliers Y. Treatment options for narcolepsy. CNS Drugs. 2016;30:369-379.
4. Barateau L, Dauvilliers Y. Recent advances in treatment for narcolepsy. Ther Adv Neurol Disord. 2019;12:1-12.

Last Reviewed: 1/20/21, 5/18/22, 7/21/23
Effective Date: 3/1/21

5. Thorpy M, Bogan R. Update on the pharmacologic management of narcolepsy: mechanisms of action and clinical implications. Sleep Medicine. 2020;68:97-109.
6. Thorpy MJ, Shapiro, C, Mayer G, et al. A randomized study of solriamfetol for excessive sleepiness in narcolepsy. Ann Neurol. 2019;85:359-370.
7. Schweitzer P, Rosenburg R, Zammit G, et al. Solriamfetol for excessive sleepiness in obstructive sleep apnea (TONES 3): a randomized controlled trial. Am J Respir Crit Care Med. 2019;199(11): 1421 - 1431.
8. Malhotra A, Shapiro C, Pepin JL, et al. Long-term study of the safety and maintenance of efficacy of solriamfetol (JZP-110) in the treatment of excessive sleepiness in participants with narcolepsy aor obstructive sleep apnea. SleepJ. 2020;43(2):1-11.
9. Strollo PJ, Hedner J, Collop N, et al. Solriamfetol for the treatment of excessive sleepiness in OSA: a placebo-controlled randomized withdrawal study. Chest. 2019; 155(2):364-374.
10. Sunosi (solriamfetol) [Prescribing Information]. Palo Alto, CA: Jazz Pharmaceuticals, Inc. Oct 2020.

Symdeko® (tezacaftor/ivacaftor)
Prior Authorization Guidelines

Affected Medication(s)

- Symdeko oral tablet

FDA Approved Indication(s)

- Treatment of patients with cystic fibrosis (CF) aged 6 years and older who are homozygous for the F508del mutation or who have at least one mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene that is responsive to tezacaftor/ivacaftor based on in vitro data and/or clinical evidence

Dosing

- Refer to package insert for age and weight specific dosing

Initial Authorization Criteria

1. Is the request for continuation of Symdeko (tezacaftor/ivacaftor) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a documentation of homozygous F508del mutation by a FDA-cleared CF mutation test or at least one mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene that is responsive to tezacaftor/ivacaftor based on in vitro data and/or clinical data? (Provide report for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Has documentation of pulmonary function (baseline FEV1) and liver function (ALT and AST) been provided and are the liver enzymes within normal range? (Provide documentation of pulmonary and liver tests for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is Symdeko (tezacaftor/ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2

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b. If no, clinical review required

2. Were updated chart notes (within past year) provided with documentation of significant clinical response to prior therapy received? (Provide documentation of improvement of FEV1 from baseline)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Were updated chart notes (within past year) provided with documentation of follow up liver function tests? (Provide documentation of AST and ALT for review)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is Symdeko (tezacaftor/ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Symdeko® (tezacaftor/ivacaftor) [Prescribing Information]. Boston, MA: Vertex Pharmaceuticals Inc. December 2022.
2. Symdeko®. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com. Accessed May 14, 2020.
3. Simon, MD. Cystic fibrosis: Overview of the treatment of lung disease. Post TW, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com. Accessed May 14, 2020.

Last Reviewed: 10/3/18, 5/20/20, 5/19/21, 5/18/22, 7/21/23
Effective Date: 1/1/19, 7/1/20, 9/15/23

Symlinpen® (pramlintide acetate)
Prior Authorization Guidelines

Affected Medication(s)

- Symlinpen subcutaneous solution

FDA Approved Indication(s)

- An adjunctive treatment in patients with type 1 or type 2 diabetes who use mealtime insulin therapy and who have failed to achieve desired glucose control despite optimal insulin therapy

Dosing

- Type 2 diabetes:
  - Initiate 60 mcg before each meal
  - May titrate to 120 mcg before each meal
- Type 1 diabetes:
  - Initiate 15 mcg before each meal
  - May titrate up to 60 mcg before each meal

Initial Authorization Criteria

1. Is the request for continuation of Symlinpen (pramlintide acetate) therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member currently taking mealtime insulin and plans to remain on mealtime insulin throughout therapy with Symlinpen (pramlintide acetate)? (Provide documentation of relevant medication history and treatment plan)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Has the member failed to achieve glucose control despite insulin adherence and titration? (Provide documentation of insulin titration and response to dosing titration)
   a. If yes, continue to #5
   b. If no, clinical review required
5. Does the member have an HbA1c that is greater than 7% and less than or equal to 9%? (Provide HbA1C result for review)
   a. If yes, continue to #6
   b. If no, clinical review required

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Effective Date: 1/1/19

6. Does the member have a history of recurrent hypoglycemia requiring assistance in the past 6 months or a history of hypoglycemia unawareness? (Provide documentation of relevant past medical history including any hypoglycemia episodes requiring assistance)

    a. If yes, clinical review required
    b. If no, approve for 6 months unless otherwise specified

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the member currently taking insulin and plans to remain on insulin throughout therapy with Symlinpen (pramlintide acetate)? (Provide documentation of treatment plan)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Does the member show a positive clinical response to therapy defined as an HbA1c of less than or equal to 9%? (Provide documentation of HbA1c result for review)

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Symlin/SymlinPen [package insert]. San Diego, CA: Amylin Pharmaceuticals, Inc.; April 2016.
2. Standards of Medical Care in Diabetes-2018: American Diabetes Association (ADA). *Diabetes Care* January 2018;39(Supplement1).
3. American Diabetes Association; 9. Pharmacologic Approaches to Glycemic Treatment: Standards of Medical Care in Diabetes—2021. Diabetes Care 1 January 2021; 44 (Supplement_1): S111–S124.

Last Reviewed: 11/21/18, 5/19/21, 11/16/22, 11/17/23
Effective Date: 1/1/19

Synagis® (palivizumab)
Prior Authorization Guidelines

Affected Medication(s)

- Synagis intramuscular solution

FDA Approved Indication(s)

- Synagis is indicated for the prevention of serious lower respiratory tract disease caused by respiratory syncytial virus (RSV) in pediatric patients:
  - With a history of premature birth (less than or equal to 35 weeks gestational age) and who are 6 months of age or younger at the beginning of RSV season
  - With bronchopulmonary dysplasia (BPD) that required medical treatment within the previous 6 months and who are 24 months of age or younger at the beginning of RSV season
  - With hemodynamically significant congenital heart disease (CHD) and who are 24 months of age or younger at the beginning of RSV season

Dosing

- 15 mg per kg of body weight given monthly by intramuscular injection
  - The first dose of Synagis should be administered prior to commencement of the RSV season and the remaining doses should be administered monthly throughout the RSV season

Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #2
   b. If no, clinical review required
2. Is the member's weight provided for review? (Provide documentation of member weight)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have a history of hospitalization for RSV infection during the current RSV season?
   a. If yes, clinical review required
   b. If no, continue to #4
4. What indication is Synagis being requested for?
   a. Premature birth, continue to corresponding criteria
   b. Chronic lung disease of prematurity, continue to corresponding criteria
   c. Hemodynamically significant congenital heart disease, continue to corresponding criteria
   d. Anatomic pulmonary abnormalities or neuromuscular disorder, continue to corresponding criteria
   e. Immunocompromised, continue to corresponding criteria
   f. Cystic fibrosis, continue to corresponding criteria

<u>Premature Birth</u>

Last Reviewed: 12/19/18, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 9/1/21, 12/20/23

1. Does the member have a history of premature birth defined as less than 29 weeks gestation? (Provide documentation of gestation age for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member <12 months of age at the start of RSV season?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the treatment plan include 5 or less doses of Synagis? (Provide treatment plan for review)
    a. If yes, approve for up to 5 doses during RSV season
    b. If no, clinical review required

Chronic Lung Disease of Prematurity

1. Does the member have a gestational age of <32 weeks? (Provide documentation of gestational age for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a diagnosis of chronic lung disease as defined by a requirement for >21% oxygen for at least 28 days after birth? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member < 12 months old at the start of RSV season?
    a. If yes, approve for up to 5 doses
    b. If no, continue to #4
4. Is the member <24 months old at the start of RSV season?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a continued requirement for medical support including chronic corticosteroid therapy, diuretic therapy, or supplemental oxygen within 6 months of the start of RSV season? (Provide supporting documentation)
    a. If yes, approve for up to 5 doses during RSV season
    b. If no, clinical review required

Hemodynamically Significant Congenital Heart Disease

1. Is the member <12 months of age at onset of RSV season?
    a. If yes, continue to #2
    b. If no, continue to #5
2. Does the member have a diagnosis of acyanotic heart disease and is receiving medication to control congestive heart failure and will require cardiac surgical procedure? (Provide supporting documentation)
    a. If yes, approve for up to 6 doses
    b. If no, continue to #3

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3. Does the member have a diagnosis of moderate to severe pulmonary hypertension? (Provide supporting documentation)
    a. If yes, approve for up to 5 doses during the RSV season
    b. If no, continue to #4
4. Does the member have a diagnosis of cyanotic heart defect and RSV prophylaxis is recommended by a pediatric cardiologist? (Provide supporting documentation)
    a. If yes, approve up to 5 doses during RSV season
    b. If no, clinical review required
5. Is the member <24 months of age at onset of RSV season?
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have a history of cardiopulmonary bypass, ECMO, or cardiac transplant during the RSV season? (Provide documentation of cardiopulmonary bypass)
    a. If yes, approve up to 6 doses during RSV season
    b. If no, clinical review required

<u>Anatomic Pulmonary Abnormalities or Neuromuscular Disorder</u>

1. Is the member <12 months of age at the onset of RSV season?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a diagnosis of a neuromuscular disease or congenital anomaly that impairs the ability to clear secretions from the upper airway? (e.g. ineffective cough) (Provide supporting documentation)
    a. If yes, approve for up to 5 doses during RSV season
    b. If no, clinical review required

<u>Immunocompromised</u>

1. Is the member <24 months of age at the onset of RSV season?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Will the member continue to be profoundly immunocompromised during the RSV season? (Examples include: solid organ or hematopoietic stem cell transplantation, chemotherapy administration, or immunocompromising disease) (Provide supporting documentation)
    a. If yes, approve for up to 5 doses during RSV season
    b. If no, clinical review required

<u>Cystic Fibrosis</u>

1. Is the member <12 months of age at the onset of RSV season?
    a. If yes, continue to #2
    b. If no, continue to #3

2. Does the member have CLD of prematurity (defined as gestational age <32 weeks and a requirement for >21% oxygen for at least 28 days after birth) and/or nutritional compromise? (Provide supporting documentation)
    a. If yes, approve for up to 5 doses during RSV season
    b. If no, clinical review required
3. Is the member <24 months of age at the onset of RSV season?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have manifestations of severe lung disease as defined by previous hospitalization for pulmonary exacerbation in the first year of life or abnormalities on chest radiography/chest computed tomography that persist when member is not experiencing exacerbation? (Provide supporting documentation)
    a. If yes, approve for up to 5 doses during RSV season
    b. If no, continue to #5
5. Does the member have a weight for length that is < 10th percentile? (Provide documentation of weigh for length percentile for review)
    a. If yes, approve for up to 5 doses during RSV season
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Synagis (palivizumab) [Prescribing Information]. Gaithersburg, MD: MedImmune, LLC. May 2017.
2. Committee on Infectious Diseases. "Updated guidance for palivizumab prophylaxis among infants and young children at increased risk of hospitalization for respiratory syncytial virus infection." Pediatrics (2014): peds-2014. Available at: http://pediatrics.aappublications.org/content/pediatrics/134/2/415.full.pdf

Last Reviewed: 12/19/18, 7/21/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 9/1/21, 12/20/23

Tavneos® (avacopan)
Prior Authorization Guidelines

Affected Medication(s)

- Tavneos oral tablet

FDA Approved Indication(s)

- Adjunctive treatment of severe active antineutrophil cytoplasmic autoantibody-associated vasculitis (granulomatosis with polyangiitis and microscopic polyangiitis) in combination with standard therapy, including glucocorticoids, in adults

Dosing

- 30 mg orally twice daily

Initial Authorization Criteria

1. Is the request a renewal of a previously approved Tavneos (avacopan) prior authorization and provided indication is the same as previous approval?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a diagnosis of granulomatosis with polyangiitis or microscopic polyangiitis? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member 18 years of age or older?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a positive test for anti-PR3 or anti-MPO? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have at least 1 major item, or 3 non-major items, or the 2 renal items of proteinuria and hematuria on Birmingham Vasculitis Activity Score (BVAS)? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have a recent documented trial of at least 3 months with inadequate response to a maximally indicated dose of systemic steroids? (Provide supporting documentation)
    a. If yes, continue to #9

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b. If no, continue to #8

8. Is use of systemic steroids contraindicated or has the member had clinically significant adverse effects from a systemic steroid trial? (Provide supporting documentation)
    a. If yes, continue to #9
    b. If no, clinical review required
9. Is Tavneos prescribed in combination with either cyclophosphamide or rituximab?
    a. If yes, continue to #10
    b. If no, clinical review required
10. Is the medication prescribed by, or in consultation with, a rheumatologist?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within 1 year) demonstrating at least a 50% reduction in BVAS from baseline or remission? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a rheumatologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. TAVNEOS (avacopan) capsules, [package insert]. Cincinnati, OH: ChemoCentyx, Inc; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 23 Nov. 2021].
3. UpToDate [internet database]. Hudson, OH: Wolters Kluwer. Updated periodically. Accessed December 31,2021.
4. Lexi-Drugs [internet database]. Hudson, OH: Lexicomp, Inc. Wolter Kluwer. Updated periodically. Accessed December 31,2021.
5. Yates, Max, et al. "EULAR/ERA-EDTA recommendations for the management of ANCA-associated vasculitis." Annals of the rheumatic diseases 75.9 (2016): 1583-1594.

Last Reviewed: 1/19/22, 3/17/23, 1/19/24
Effective Date: 3/1/22, 3/1/24

Tegsedi® (inotersen sodium), Wainua™ (eplontersen)
Prior Authorization Guidelines

Affected Medication(s)

- Tegsedi subcutaneous solution
- Wainua subcutaneous solution

FDA Approved Indication(s)

- Treatment of polyneuropathy of hereditary transthyretin-mediated amyloidosis in adults

Dosing

- Tegsedi: 284 mg subcutaneously once weekly
- Wainua: 45 mg subcutaneously once monthly

Initial Authorization Criteria

1. Is the request for continuation of a previously approved agent?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation confirming the presence of a transthyretin (TTR) mutation? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have documentation of a biopsy that was found to be positive for amyloid deposits? (Provide documentation of biopsy)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member meet the diagnosis and clinical requirements for at least one of the following below? (Provide supporting documentation)
    - Subjective patient symptoms are suggestive of neuropathy
    - Abnormal nerve conduction studies are consistent with polyneuropathy
    - Abnormal neurological examination is suggestive of neuropathy

    a. If yes, continue to #7
    b. If no, clinical review required

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7. Is the treatment being prescribed by, or in consultation with, a neurologist, geneticist, or provider who specializes in the management of amyloidosis?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Has the member demonstrated a positive clinical response to therapy defined as an improvement in neuropathy symptoms? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a neurologist, geneticist, or provider who specializes in the management of amyloidosis?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Tegsedi (inotersen sodium) subcutaneous injection [package insert]. Boston, MA: Akcea Therpeutics, Inc; June 2022.
2. WAINUA™ (eplontersen) subcutaneous injection [package insert]. Wilmington, DE: AstraZeneca; December 2023.
3. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 8 Jan, 2019].
4. Ando Y, Coelho T, Berk JL, Cruz MW, Ericzon BG, Ikeda SI, et al. Guideline of transthyretin-related hereditary amyloidosis for clinicians. Orphanet Journal of Rare Diseases. 2013;8(1):1-18.
5. Hawkins PN, Ando Y, Dispenzeri A, Gonzalez-Duarte A, Adams D, Suhr OB. Evolving landscape in the management of transthyretin amyloidosis. Annals of Medicine. 2015;47(8):625-38.
6. Ando Y, Adams D, Benson MD, et al. Guidelines and new directions in the therapy and monitoring of ATTRv amyloidosis. Amyloid. 2022;29(3):143-155.

Last Reviewed: 3/20/19, 7/21/21, 3/17/23, 3/15/24
Effective Date: 5/1/19, 6/1/23, 4/15/24

Testosterone Products
Prior Authorization Guidelines

Affected Medication(s)

- Androderm (testosterone) transdermal patch
- Androgel (testosterone) 1% packet (25mg/2.5g)
- Androgel 1.62% gel packet
- Fortesta (testosterone) topical gel
- Jatenzo (testosterone undecanoate) oral capsule
- Testosterone 1% packet (25mg/2.5g)
- Testosterone 1.62% packet
- Testosterone 10 mg gel pump
- Testosterone 30mg/1.5ml pump
- Tlando (testosterone undecanoate) oral capsule
- Xyosted (testosterone) subcutaneous solution

FDA Approved Indication(s)

- Primary hypogonadism (congenital or acquired): Testicular failure due to conditions such as cryptorchidism, bilateral torsion, orchitis, vanishing testis syndrome, orchiectomy, Klinefelter's syndrome, chemotherapy, or toxic damage from alcohol or heavy metals
- Hypogonadotropic hypogonadism (congenital or acquired): Gonadotropin or luteinizing hormone-releasing hormone (LHRH) deficiency or pituitary-hypothalamic injury from tumors, trauma, or radiation

Dosing

- Refer to corresponding package insert for dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same testosterone therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a trial with insufficient response, intolerance, or contraindication to generic intramuscular testosterone? (Provide documentation of trial with insufficient response or intolerance)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a trial with insufficient response, intolerance, or contraindication to at least one of the following products?
    - Testosterone 50mg/5g gel
    - Testosterone 1% packet (50mg/5g)
    - Testosterone 12.5mg/1.25g gel pump

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Effective Date: 1/1/19, 5/1/19, 7/1/20, 11/1/21, 9/15/22, 12/20/23

- Testosterone 1.62% gel pump

a. If yes, continue to #5
b. If no, clinical review required

5. What indication is the testosterone medication being requested for?

a. Hypogonadism, continue to corresponding criteria
b. Gender affirming treatment, continue to corresponding criteria

Hypogonadism

1. Is the member currently taking testosterone replacement therapy? (Provide documentation of testosterone replacement therapy history)

a. If yes, continue to #2
b. If no, continue to #3

2. Does the member have a testosterone level that was taken in the morning that is either within normal range or below normal (below normal range: total testosterone < 300 ng/dL or free testosterone <5 ng/dL)? (Provide documentation of testosterone level)

a. If yes, continue to #4
b. If no, clinical review required

3. Does the member have documentation of TWO baseline testosterone levels, taken in the mornings of different days, that are below normal range (total testosterone< 300 ng/dL or free testosterone <5ng/dL)? (Provide documentation of testosterone levels)

a. If yes, continue to #4
b. If no, clinical review required

4. Does the member have at least TWO signs/symptoms of hypogonadism (If member is currently on testosterone therapy, did they experience at least TWO signs/symptoms before initiation of therapy)? (Examples include sleep disturbances, gynecomastia, decreased lean body mass, visceral obesity, hot flashes, changes in mood, cognitive impairment, insulin resistance, anemia, and low bone mineral density) (Provide supporting documentation of signs/symptoms)

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

Gender Affirming Treatment

1. Does the member have a diagnosis of gender identity disorder by a qualified mental health professional? (Provide supporting documentation)

a. If yes, approve for 6 months unless otherwise specified
b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

a. If yes, continue to #2

b. If no, clinical review required

2. Is the medication being used for gender affirming care?
   a. If yes, approve x 12 months
   b. If no, continue to #3

3. Does the member have a positive clinical response to therapy as defined by a total serum testosterone level that is within normal range? (Provide documentation of testosterone levels)
   a. If yes, approve for 12 months unless otherwise specified
   b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Androderm [prescribing information] Actavis Pharma, Inc. October 2016.
2. AndroGel 1% [prescribing information] AbbVie Inc. October 2016.
3. Fortesta [prescribing information] Endo Pharma, Inc. October 2016.
4. Jatenzo [testosterone undecanoate] capsules. Clarus Therapeutics, Inc. Northbrook, IL; 2019
5. Xyosted [prescribing information] Antares Pharma, Inc. September 2018.
6. Bhasin S, Brito JP, Cunningham GR, et al. Testosterone Therapy in Men With Hypogonadism: An Endocrine Society Clinical Practice Guideline. J Clin Endocrinol Metab 2018; 103:1715.
7. Olson-Kennedy J, Rosenthal SM, Hastings J, et al. UCSF Transgender Care & Treatment Guidelines: Health consideration for gender non-forming children and transgender adolescents. 2016 Jun. Available at: https://transcare.ucsf.edu/guidelines/youth

Last Reviewed: 12/19/18, 3/20/19, 5/20/20, 9/15/21, 7/20/22, 11/17/23
Effective Date: 1/1/19, 5/1/19, 7/1/20,11/1/21, 9/15/22, 12/20/23

Therapeutic Immunomodulators
Prior Authorization Guidelines

Affected Medication(s)

- Abrilada (adalimumab-afzb)
- Adalimumab-aaty
- Adalimumab-adaz
- Adalimumab-adbm
- Adalimumab-fkip
- Adalimumab-ryvk
- Actemra (tocilizumab)
- Actemra Actpen (tocilizumab)
- Amjevita (adalimumab-atto)
- Bimzelx (bimekizumab)
- Cimzia (certolizumab pegol)
- Cosentyx (secukinumab)
- Cyltezo (adalimumab-adbm)
- Enbrel (etanercept)
- Entyvio (vedolizmab)
- Hadlima (adalimumab-bwwd)
- Hulio (adalimumab-fkjp)
- Humira (adalimumab)
- Hyrimoz (adalimumab-adaz)
- Idacio (adalimumab-aacf)
- Ilumya (tildrakizumab)
- Kevzara (sarilumab)
- Kineret (anakinra)
- Omvoh (mirikuzumab-mrkz)
- Orencia (abatacept)
- Otezla (apremilast) oral tablet
- Siliq (brodalumab)
- Simlandi (adalimumab-ryvk)
- Simponi (golimumab)
- Skyrizi (risankizumab)
- Sotyktu (deucravacitinib) oral tablet
- Stelara (ustekinumab)
- Taltz (ixekizumab)
- Tremfya (guselkumab)
- Vesipity (etrasimod)
- Yuflyma (adalimumab-aaty)
- Yusimry (adalimumab-aqvh)
- Zymfentra (infliximab-dyyb)

FDA Approved Indication(s)

- Drug Compendia supported indications may be covered

Drug Name	RA	JIA	PsA	AS	SpA	Crohn	UC	Ps	HS	Uveitis	Other
Cimzia	X		X	X	X	X		X			
Adalimumab-adaz	X	X	X	X		X	X	X	X	X	
Adalimumab-adbm	X	X	X	X		X	X	X	X	X	
Hyrimoz	X	X	X	X		X	X	X	X	X	
Cyltezo	X	X	X	X		X	X	X	X	X	
Simponi	X		X	X			X				
Skyrizi			X			X		X			
Stelara			X			X	X	X			
Kevzara	X										
Taltz			X	X	X			X			
Bimzelx								X			
Cosentyx			X	X	X			X	X		X
Enbrel	X	X	X	X				X			
Entyvio						X	X				
Non-preferred adalimumab biosimilars	X	X	X	X		X	X	X	X	X	
Humira	X	X	X	X		X	X	X	X	X	

Last Reviewed: 11/21/18, 5/15/19, 9/18/19, 1/21/20, 7/15/20, 9/16/20, 11/18/20, 11/16/22, 3/17/23, 11/17/23, 1/19/24, 3/15/24, 5/17/24
Effective Date: 1/1/19, 7/1/19, 10/15/19, 3/15/20, 8/15/20, 11/15/20, 12/15/20, 1/1/23, 6/1/23, 12/20/23, 6/15/24

Actemra	X	X									X
Ilumya								X			
Kineret	X										X
Omvoh							X				
Orencia	X	X	X								X
Otezla			X					X			X
Siliq								X			
Sotyktu								X			
Tremfya			X					X			
Velsipity							X				
Zymfentra						X	X				

Dosing

- Refer to corresponding package insert for information

Initial Authorization Criteria

1. Is the request for continuation of therapeutic immunomodulatory therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication or a major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Will the requested medication be used concurrently with any other biologic therapy? (Examples: Enbrel, Actemra, Cimzia, Simponi, Orencia, Taltz, Cosentyx, Otezla, etc)
    a. If yes, clinical review required
    b. If no, continue to #4
4. What is the diagnosis that the medication is being requested for?
    a. Rheumatoid arthritis, continue to corresponding criteria
    b. Juvenile idiopathic arthritis, continue to corresponding criteria
    c. Ankylosing spondylitis/nr-axSpA, continue to corresponding criteria
    d. Psoriatic arthritis, continue to corresponding criteria
    e. Crohn's disease, continue to corresponding criteria
    f. Ulcerative colitis, continue to corresponding criteria
    g. Plaque psoriasis, continue to corresponding criteria
    h. Hidradenitis suppurativa, continue to corresponding criteria
    i. Uveitis, continue to corresponding criteria
    j. Other indication not listed, continue to corresponding criteria

Rheumatoid Arthritis (RA)

1. Is the diagnosis of rheumatoid arthritis (RA) confirmed by ACR/EULAR classification criteria AND has the diagnosis been documented for greater for 6 months? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member have moderate to severe active RA confirmed by one of the tests below and despite the current RA management regimen? (Provide test result for review and provide current RA regimen)

    - Patient Activity Scale (PAS) or PASII of 3.7 or higher
    - Routine Assessment of Patient Index Data 3 (RAPID3) of 2.0 or higher
    - Clinical Disease Activity Index (CDAI) of 10 or higher
    - Disease Activity Score (DAS) 28 erythrocyte sedimentation rate (ESR) of 3.2 or higher
    - Simplified Disease Activity Index (SDAI) of 11 or higher

    a. If yes, continue to #3
    b. If no, clinical review required

3. Did the member have an inadequate response to a 12 week trial of methotrexate? (Provide documentation of inadequate response to methotrexate)

    a. If yes, continue to #6
    b. If no, continue to #4

4. Does the member have a contraindication or history of intolerance to methotrexate? (Provide documentation of contraindication and/or intolerance. Note: 1. Alcohol consumption is not considered a contraindication 2. Nausea to oral formulation is not considered an intolerance)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Did the member have a contraindication to all OR an inadequate response to one 12 week trial with the following disease-modifying antirheumatic drugs: leflunomide, sulfasalazine, or hydroxychloroquine? (Provide documentation of contraindication or inadequate response to therapy)

    a. If yes, continue to #6
    b. If no, clinical review required

6. Is the request for a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), or Simponi (golimumab)?

    a. If yes, continue to #10
    b. If no, continue to #7

7. Does the member have a documented inadequate response, contraindication, or intolerance to TWO of the following agents: preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Rinvoq (upadacitinib), Simponi (golimumab), or Xeljanz/Xeljanz XR (tofacitinib citrate)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #8
    b. If no, clinical review required

8. Is the request for Kevzara (sarilumab)?

    a. If yes, continue to #10
    b. If no, continue to #9

9. Does the member have a documented inadequate response, contraindication, or intolerance to Kevzara (sarilumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #10
    b. If no, clinical review required

10. Is the medication being prescribed by or in consultation with a rheumatologist?

a. If yes, approve 6 months unless otherwise specified
b. If no, clinical review required

Juvenile Idiopathic Arthritis (JIA/PJIA)

1. Does the member have moderate to severe active polyarticular JIA defined as greater or equal to 5 swollen joints and at least 3 joints with limitation in motion? (Provide documentation of affected joints and current treatment regimen)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Did the member have an inadequate response to a 12 week trial of methotrexate? (Provide documentation of trial with inadequate response)

    a. If yes, continue to #5
    b. If no, continue to #3

3. Does the member have a contraindication or history of intolerance to methotrexate? (Provide documentation of contraindication and/or intolerance. Note: 1. Alcohol consumption is not considered a contraindication 2. Nausea to oral formulation is not considered an intolerance)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Did the member have a contraindication or history of intolerance to leflunomide? (Provide documentation of contraindication and/or intolerance)

    a. If yes, continue to #5
    b. If no, deny. Clinical criteria not met

5. Is the request for a preferred adalimumab biosimilar?

    a. If yes, continue to #7
    b. If no, continue to #6

6. Does the member have documentation of an inadequate response, intolerance, or contraindication to preferred adalimumab biosimilar? (Provide documentation of inadequate response, contraindication, and/or intolerance)

    a. If yes, continue to #7
    b. If no, clinical review required

7. Is the medication being prescribed by or in consultation with a rheumatologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Ankylosing Spondylitis (AS)/nr-axSpA

1. Does the member currently have active AS despite a current treatment regimen as defined by the below? (Provide supporting documentation)
    - Bath ankylosing spondylitis disease activity index (BASDAI) greater or equal to 4 OR
    - Ankylosing Spondylitis Disease Activity Score (ASDAS) greater or equal to 2.1 AND
    - Elevated CRP, positive MRI, or Radiographic sacroiliitis

    a. If yes, continue to #2
    b. If no, clinical review required

2. Did the member have an inadequate response or intolerance to TWO separate 4 week trials of prescription strength oral nonsteroidal anti-inflammatory drugs (NSAIDs)? (Provide documentation of NSAIDs tried, examples: ibuprofen, naproxen, diclofenac, meloxicam, etc.)

    a. If yes, continue to #4
    b. If no, continue to #3

3. Does the member have a contraindication to oral NSAIDs? (Provide documentation of contraindication)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Does the member have isolated sacroiliitis, or enthesitis disease? (Provide supporting documentation)

    a. If yes, continue to #5
    b. If no, continue to #7

5. Did the member have an inadequate response to a parenteral glucocorticoid injection? (Provide documentation of trial with inadequate response)

    a. If yes, continue to #9
    b. If no, continue to #6

6. Does the member have a contraindication to a parenteral glucocorticoid injection? (Provide documentation of contraindication)

    a. If yes, continue to #9
    b. If no, clinical review required

7. Did the member have an inadequate response to a 12 week trial with sulfasalazine? (Provide documentation of trial with inadequate response)

    a. If yes, continue to #9
    b. If no, continue to #8

8. Does the member have a contraindication or history of intolerance to sulfasalazine? (Provide documentation of contraindication and/or intolerance)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Is the request for a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), or Simponi (golimumab)?

    a. If yes, continue to #15
    b. If no, continue to #10

10. Does the member have a documented inadequate response, intolerance, or contraindication to TWO of the following agents: a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), or Simponi (golimumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #11
    b. If no, clinical review required

11. Is the request for Taltz (ixekizumab)?

    a. If yes, continue to #15
    b. If no, continue to #12

12. Does the member have a documented inadequate response, intolerance, or contraindication to Taltz (ixekizumab)? (Provide documentation of inadequate response, contraindication, and/or intolerance)

a. If yes, continue to #13
b. If no, clinical review required

13. Is the requested medication Cosentyx (secukinumab)?

    a. If yes, continue to #14
    b. If no, continue to #15

14. Does the member have a contraindication to intravenous Cosentyx (secukinumab)? (Provide documentation of contraindication. Note: Intravenous Cosentyx approved for use in adults only)

    a. If yes, continue to #15
    b. If no, clinical review required

15. Is the medication being prescribed by or in consultation with a rheumatologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Psoriatic Arthritis (PsA)

1. Does the member currently have active PsA defined as greater or equal to 3 swollen joints AND greater or equal to 3 tender or painful joints despite the current treatment regimen? (Provide documentation of affected joints and current treatment regimen)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Did the member have a contraindication or an inadequate response to a 12 week trial with one of the following: methotrexate, leflunomide, cyclosporine, sulfasalazine? (Provide documentation of trial with inadequate response or contraindication)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the request for a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Simponi (golimumab), Skyrizi (risankizumab) or Stelara (ustekinumab)?

    a. If yes, continue to #9
    b. If no, continue to #4

4. Does the member have a documented inadequate response, contraindication, or intolerance to TWO of the following agents: a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Simponi (golimumab), Skyrizi (risankizumab) or Stelara (ustekinumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #5
    b. If no, clinical review required

5. Is the request for Taltz (ixekizumab)?

    a. If yes, continue to #9
    b. If no, continue to #6

6. Does the member have a documented inadequate response, contraindication, or intolerance to BOTH of the following agents: Taltz (ixekizumab) and Xeljanz (tofacitinib)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #7

b. If no, clinical review required

7. Is the requested medication Cosentyx (secukinumab)?

    a. If yes, continue to #8
    b. If no, continue to #9

8. Does the member have a contraindication to intravenous Cosentyx (secukinumab)? (Provide documentation of contraindication. Note: Intravenous Cosentyx approved for use in adults only)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Is the medication being prescribed by or in consultation with a rheumatologist or dermatologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Crohn's Disease (CD)

1. Does the member currently have active CD defined as a Crohn's Disease Activity Index (CDAI) greater than 220 despite the current treatment regimen? (Provide documentation of CDAI and current treatment regimen)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Did the member have an inadequate response to TWO of the following oral agents for a minimum trial of 12 weeks each: 6-mercaptopurine, azathioprine, corticosteroid, methotrexate, mesalamine? (Provide documentation of 12 week trials with inadequate responses)

    a. If yes, continue to #4
    b. If no, continue to #3

3. Does the member have a contraindication or history of intolerance to at least TWO of the following oral agents: 6-mercaptopurine, azathioprine, corticosteroids, methotrexate, mesalamine, sulfasalazine? (Provide documentation of contraindications and/or intolerances)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is the medication being requested a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Skyrizi (risankizumab) or Stelara (ustekinumab)?

    a. If yes, continue to #9
    b. If no, continue to #5

5. Does the member have a documented inadequate response, contraindication, or intolerance to TWO of the following agents: a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Skyrizi (risankizumab) or Stelara (ustekinumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)

    a. If yes, continue to #6
    b. If no, clinical review required

6. Is the requested medication Zymfentra (infliximab-dyyb)?

    a. If yes, continue to #7
    b. If no, continue to #9

7. Does the member have a contraindication to maintenance dosing of intravenous infliximab or an infliximab biosimilar? (Provide documentation of contraindication)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Has the member received three intravenous induction doses of an infliximab product prior to initiation and does the member have a positive response to treatment? (Provide supporting documentation)
    a. If yes, continue to #9
    b. If no, clinical review required
9. Is the medication being prescribed by or in consultation with a gastroenterologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Ulcerative Colitis (UC)

1. Does the member currently have active Ulcerative Colitis? (Provide documentation of diagnosis confirmed by endoscopy, colonoscopy, or sigmoidoscopy with Mayo score of greater than 2)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the medication being requested a preferred adalimumab biosimilar, Simponi (golimumab), or Stelara (ustekinumab)?
    a. If yes, continue to #7
    b. If no, continue to #3
3. Does the member have a documented inadequate response, intolerance, or contraindication to TWO of the following: a preferred adalimumab biosimilar, Simponi (golimumab), or Stelara (ustekinumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the requested medication Zymfentra (infliximab-dyyb)?
    a. If yes, continue to #5
    b. If no, continue to #7
5. Does the member have a contraindication to maintenance dosing of intravenous infliximab or an infliximab biosimilar? (Provide documentation of contraindication)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Has the member received three intravenous induction doses of an infliximab product prior to initiation and does the member have a positive response to treatment? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the medication being prescribed by, or in consultation with, a gastroenterologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 11/21/18, 5/15/19, 9/18/19, 1/21/20, 7/15/20, 9/16/20, 11/18/20, 11/16/22, 3/17/23, 11/17/23, 1/19/24, 3/15/24, 5/17/24
Effective Date: 1/1/19, 7/1/19, 10/15/19, 3/15/2020, 8/15/20, 11/15/20, 12/15/20, 1/1/23, 6/1/23, 12/20/23, 06/15/24

Plaque Psoriasis (Ps)

1. Does the member currently have moderate to severe chronic Ps defined as having functional impairment (e.g. inability to use hands or feet or activities of daily living, or significant facial involvement preventing normal social interaction) AND one or more of the following: 1. At least 10% body surface area involvement AND/OR 2. Hand, foot or mucous membrane involvement? (Provide documentation of functional impairment and body area involvement)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Did the member have an inadequate response to TWO separate 12 week trials with TWO of the following systemic therapies: methotrexate, cyclosporine, phototherapy? (Provide documentation of 12 week trials with inadequate responses)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the medication being requested Cimzia (certolizumab pegol), a preferred adalimumab biosimilar, Skyrizi (risankizumab), or Stelara (ustekinumab)
    a. If yes, continue to #8
    b. If no, continue to #4
4. Is the request for Taltz (ixekizumab)?
    a. If yes, continue to #5
    b. If no, continue to #6
5. Does the member have documentation of an inadequate response, intolerance, or contraindication to ONE of the following agents: a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Skyrizi, (risankizumab), or Stelara (ustekinumab)? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #8
    b. If no, clinical review required
6. Does the member have documentation of an inadequate response, intolerance, or contraindication to TWO of the following agents:a preferred adalimumab biosimilar, Cimzia (certolizumab pegol), Skyrizi (risankizumab), or Stelara (ustekinumab)? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have documentation of an inadequate response, intolerance, or contraindication to Taltz (ixekizumab)? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #8
    b. If no, clinical review required
8. Is the treatment being prescribed by or in consultation with a dermatologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Hidradenitis Suppurativa

1. Does the member have Hurley stage II or III Hidradenitis Suppurativa? (Provide documentation of Hurley stage)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Did the member have a previous inadequate response to oral antibiotics? (Provide documentation of oral antibiotic regimen trialed and inadequate response)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the request for a preferred adalimumab biosimilar?
    a. If yes, continue to #5
    b. If no, continue to #4
4. Does the member have documentation of inadequate response, intolerance, or contraindication to a preferred adalimumab biosimilar? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by or in consultation with a dermatologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

<u>Uveitis</u>

1. Does the member have a diagnosis of non-infectious intermediate, posterior uveitis or panuveitis?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have an inadequate response, intolerance, or contraindication to at least TWO of the following: cyclosporine, systemic glucocorticoids, and/or an antimetabolite (mycophenolate, methotrexate, or azathioprine)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the request for a preferred adalimumab biosimilar?
    a. If yes, continue to #5
    b. If no, continue to #4
4. Does the member have documentation of an inadequate response, intolerance, or contraindication to a preferred adalimumab biosimilar? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by or in consultation with an ophthalmologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Other Indications

1. Is the request for an FDA approved indication?
    a. If yes, continue to #4
    b. If no, continue to #2
2. Is the requested use supported by major compendia not otherwise excluded by plan design?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member tried and had an inadequate response OR does the member have a contraindication to ALL standard treatment options for the requested indication? (Provide documentation of inadequate responses, contraindications, and/or intolerances)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment being prescribed by or in consultation with an appropriate specialist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication FDA-approved or supported by major compendia? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (dated within 1 year) provided with documentation of significant clinical response to therapy? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the request for Humira or a non-preferred adalimumab biosimilar?
    a. If yes, continue to #4
    b. If no, continue to #5
4. Does the member have documentation of an inadequate response, intolerance, or contraindication to two different preferred adalimumab biosimilars? (Provide documentation of inadequate response, contraindication, and/or intolerance)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Will the requested medication be used with other biologic therapy? (Examples: Enbrel, Actemra, Cimzia, Simponi, Orencia, Taltz, Cosentyx, Otezla, etc)
    a. If yes, clinical review required
    b. If no, continue to #6
6. Is the treatment being prescribed by or in consultation with an appropriate specialist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 11/21/18, 5/15/19, 9/18/19, 1/21/20, 7/15/20, 9/16/20, 11/18/20, 11/16/22, 3/17/23, 11/17/23, 1/19/24, 3/15/24, 5/17/24
Effective Date: 1/1/19, 7/1/19, 10/15/19, 3/15/2020, 8/15/20, 11/15/20, 12/15/20, 1/1/23, 6/1/23, 12/20/23, 06/15/24

Note:
Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Abrilada (adalimumab-afzb) [Prescribing Information]. New York, NY: Pfizer Inc. Oct 2023.
2. Adalimumab-adaz [Prescribing Information]. Princeton, NJ: Sandoz. Mar 2023.
3. Actemra (tocilizumab) [Prescribing Information]. South San Francisco, CA: Genentech. June 2022.
4. Amjevita (adalimumab-atto) [Prescribing Information]. Thousand Oaks, CA: Amgen Inc. Aug 2023.
5. Bimzelx (bimekizumab-bkzx) [Prescribing Information]. Smyrna, GA: UCB, Inc.; 2023.
6. Cimzia (certolizumab pegol) [Prescribing Information]. Smyrna, GA: UCB Inc. March 2021.
7. Cosentyx (secukinumab) [Prescribing Information]. East Hanover, NJ: Novartis Pharmaceuticals Corporation. December 2021.
8. Cyltezo (adalimumab-adbm) [Prescribing Information]. Ridgefield, CT: Boehringer Ingelheim Pharmaceuticals, Inc. Sep 2023.
9. Enbrel (etanercept) [Prescribing Information]. Thousand Oaks, CA: Amgen. June 2022.
10. Hadlima (adalimumab-bwwd) [Prescribing Information]. Jersey City, NJ: Organon LLC. Jul 2023
11. Hulio (adalimumab-fkjp) [Prescribing Information]. Morgantown, WV: Mylan Specialty L.P. Aug 2023
12. Humira (adalimumab) [Prescribing Information]. North Chicago, IL: AbbVie Inc. February 2021.
13. Hyrimoz (adalimumab-adaz) [Prescribing Information]. Princeton, NJ: Sandoz. Jun 2023.
14. Idacio (adalimumab-aacf) [Prescribing Information]. Lake Zurich, IL: Fresenius Kabi USA, LLC. Mar 2023.
15. Ilumya (tildrakizumab-asmn) [Prescribing Information]. Cranbury, NJ: Sun Pharma Global FZE. April 2022.
16. Kevzara (sarilumab) [Prescribing Information]. Bridgewater, NJ: Sanofi-aventis U.S. LLC. April 2018.
17. Kineret (anakinra) [Prescribing Information]. Stockholm, Sweden: Swedish Orphan Biovitrum AB. December 2020.
18. Omvoh (mirikizumab-mrkz) injection for Intravenous or subcutaneous use [Prescribing Information]. Indianapolis, IN: Eli Lilly and Company; 2023.
19. Orencia (abatacept) [Prescribing Information]. Princeton, NJ: E.R. Squibb & Sons L.L.C. December 2021.
20. Otezla (apremilast) [Prescribing Information]. Summit, NJ: Celgene. February 2021.
21. Siliq (brodalumab) [Prescribing Information]. Bridgewater, NJ: Valeant Pharmaceuticals North America LLC. June 2020.
22. Simlandi (adalimumab-ryvk) [Prescribing Information]. Leesburg, VA: Alvotech USA. February 2023.
23. Simponi (golimumab) [Prescribing Information]. Horsham, PA: Janssen Biotech Inc. September 2019.
24. Stelara (ustekinumab) [Prescribing Information]. Horsham, PA: Janssen Biotech Inc. August 2022.
25. Skyrizi (risankizumab-rzaa) [Prescribing Information]. North Chicago, IL: AbbVie Inc. June 2022.

Last Reviewed: 11/21/18, 5/15/19, 9/18/19, 1/21/20, 7/15/20, 9/16/20, 11/18/20, 11/16/22, 3/17/23, 11/17/23, 1/19/24, 3/15/24, 5/17/24
Effective Date: 1/1/19, 7/1/19, 10/15/19, 3/15/2020, 8/15/20, 11/15/20, 12/15/20, 1/1/23, 6/1/23, 12/20/23, 06/15/24

26. Taltz (ixekizumab) [Prescribing Information]. Indianapolis, IN: Eli Lilly and Company. July 2022.
27. Tremfya (guselkumab) [Prescribing Information]. Horsham, PA: Janssen Biotech Inc. July 2020.
28. Vesipity (etrasimod) [Prescribing Information]. New York, NY: Pfizer; October 2023.
29. Yuflyma (adalimumab-aaty) [Prescribing Information]. Jersey City, NJ: Celltrion USA. Oct 2023.
30. Yusimry (adalimumab-aqvh) [Prescribing Information]. Redwood City, CA: Coherus BioSciences Inc. Sep 2023.
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Tobi Podhaler® (tobramycin)
Prior Authorization Guidelines

Affected Medication(s)

- Tobi Podhaler inhalation capsule

FDA Approved Indication(s)

- For the management of cystic fibrosis patients with *Pseudomonas aeruginosa*

Dosing

- Tobi Podhaler
  - For adults and children 6 years of age and older: Inhale contents of four 28 mg capsules twice daily via podhaler device in cycles of 28 days on drug, 28 days off of drug

Initial Authorization Criteria

1. Is the request for continuation of Tobi Podhaler therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Does the member have a diagnosis of cystic fibrosis and a positive culture demonstrating infection with *Pseudomonas aeruginosa*? (Provide supporting documentation of diagnosis and positive culture for *Pseudomonas aeruginosa*)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Does the member have baseline FEV1 greater than or equal to 25%? (Provide baseline FEV1 for review)
   a. If yes, continue to #5
   b. If no, clinical review required
5. Has the member had a medical rationale for avoiding therapy with generic tobramycin inhaled solution? (Provide documentation of medical rationale for avoidance)
   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #2

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b. If no, clinical review required

2. Does the member have a positive response to therapy as defined by stability in their disease state? (Provide supporting documentation for review)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

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References:

1. Tobi Podhaler (tobramycin) [Prescribing Information]. East Hanover, NJ: Novartis Pharmaceuticals Corporation. July 2020.
2. Mogayzel Jr, Peter J., et al. "Cystic Fibrosis Foundation pulmonary guideline. Pharmacologic approaches to prevention and eradication of initial Pseudomonas aeruginosa infection." *Annals of the American Thoracic Society* 11.10 (2014): 1640-1650.

Last Reviewed: 12/19/18, 11/18/20, 9/15/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 1/15/21, 4/15/24

Trikafta® (elexacaftor/tezacaftor/ivacaftor)
Prior Authorization Guidelines

Affected Medication(s)

- Trikafta oral tablet
- Trikafta oral granules

FDA Approved Indication(s)

- Treatment of patients with cystic fibrosis (CF) ages 2 years and older who have at least one *F508del* mutation or other responsive mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene

Dosing

- Refer to package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of Trikafta (elexacaftor/tezacaftor/ivacaftor) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the patient have a documentation of at least one *F508del* mutation or other responsive mutation in the CFTR gene confirmed by an FDA-cleared CF mutation test? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member at least 2 years of age?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has documentation of pulmonary function (baseline FEV1), liver function (ALT and AST), and bilirubin been provided and are the liver enzymes within normal range? (Provide documentation of pulmonary and liver tests for review)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is Trikafta (elexacaftor/tezacaftor/ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis member?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Last Reviewed: 3/18/20, 7/21/21, 11/16/22, 5/19/23, 7/21/23
Effective Date: 5/1/20, 9/15/23

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within the past year) provided with documentation of clinical response to prior therapy received? (Provide documentation of improvement of FEV1 from baseline and/or a reduction in the number of pulmonary exacerbations)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has documentation been provided of liver function tests (ALT and AST) within the last year and are they within normal limits? (Provide ALT and AST levels for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is Trikafta (elexacaftor/tezacaftor/ivacaftor) being prescribed by, or in consult with, a pulmonologist or a specialist experienced in treating cystic fibrosis member?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Trikafta™ (elexacaftor/tezacaftor, and ivacaftor) [Prescribing Information]. Boston, MA: Vertex Pharmaceuticals Inc. October 2021.
2. Trikafta™. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com.
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Last Reviewed: 03/18/20, 7/21/21, 11/16/22, 5/19/23, 7/21/23
Effective Date: 5/1/20, 9/15/23

Truqap (capivastertib)
Prior Authorization Guidelines

Affected Medication(s)

- Truqap oral tablet

Indication(s)

- Treatment of adult patients with hormone receptor (HR)-positive, human epidermal growth factor receptor 2 (HER2)-negative, locally advanced or metastatic breast cancer with one or more PIK3CA/AKT1/PTEN-alterations as detected by an FDA-approved test following progression on at least one endocrine-based regimen in the metastatic setting or recurrence on or within 12 months of completing adjuvant therapy

Dosing:

- 400 mg orally twice daily, with or without food, for 4 days followed by 3 days off until disease progression or unacceptable toxicity

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same anti-cancer medication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the medication being requested to be used for an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #4
    b. If no, continue to #3
3. Is the medication being requested being used for an indication supported by the National Comprehensive Cancer Network (NCCN) with an evidence level of 2A or higher? (Provide disease staging, all prior treatment history, pathology report, and anticipated treatment plan for review)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have Karnofsky Performance Status greater or equal to 50% OR Eastern Cooperative Oncology Group (ECOG) performance status of 0-2? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have an identified alteration in PIK3CA only?
    a. If yes, continue to #6
    b. If no, continue to #7
6. Does the member have a previous trial with inadequate response, intolerance or contraindication to Piqray (alpelisib)? (Provide supporting documentation)
    a. If yes, continue to #7

Last Reviewed: 1/19/24
Effective Date: 3/1/24

b. If no, clinical review required

7. Is the medication being prescribed by or in consultation with an oncologist?

   a. If yes, approve for 4 months unless otherwise specified
   b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication approved by the FDA or supported by the NCCN recommendation with an evidence level of 2A or higher? (Provide documentation of diagnosis)

   a. If yes, continue to #2
   b. If no, clinical review required

2. Is there clinical documentation confirming disease responsiveness to therapy provided? (Examples include reduction in tumor size, objective response, delay in progression, partial response, etc.) (Provide supporting documentation)

   a. If yes, continue to #2
   b. If no, clinical review required

3. Is the medication being prescribed by or in consultation with an oncologist?

   a. If yes, approve for 12 months unless otherwise specified
   b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Truqap (capivasertib) tablets, [package insert]. Wilmington, DE: AstraZeneca Pharmaceuticals LP; 2023.
2. Drugs@FDA: FDA Approved Drug Products. 2023. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 11 Dec. 2023].
3. Clinical Practice Guidelines in Oncology (NCCN Guidelines): Breast Cancer. Version 5.2023 National Comprehensive Cancer Network website. Available from https://www.nccn.org/professionals/physician_gls/pdf/breast.pdf. Accessed December 12, 2023.
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Upneeq® (oxymetazoline hydrochloride)
Prior Authorization Guidelines

Affected Medication(s)

- Upneeq ophthalmic solution

FDA Approved Indication(s)

- Treatment of acquired blepharoptosis in adults

Dosing

- Instill one drop into affected eye(s) once daily

Initial Authorization Criteria

1. Is the request for continuation of Upneeq (oxymetazoline ophthalmic solution) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have blepharoptosis that is caused by any of the following?
        - Congenital ptosis
        - Horner syndrome
        - Myasthenia gravis
        - Mechanical cause ptosis
    a. If yes, clinical review required
    b. If no, continue to #5
5. Does the member's blepharoptosis cause functional impairment? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member meet one of the following? (Provide supporting documentation)
        - Marginal reflex distance-1 (MRD-1) ≤ 2mm
        - Inability to detect ≤ 8 of 17 points in the top 2 rows on the Leicester Peripheral Field Test (LPFT)
    a. If yes, continue to #7
    b. If no, clinical review required

Last Reviewed: 7/21/21, 3/17/23, 3/15/24
Effective Date: 9/1/21, 4/15/24

7. Is the treatment being initiated by, or in consultation with, an ophthalmologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member show a positive clinical response to therapy as defined by an improvement in visual field deficit? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by or in consultation with an ophthalmologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. UPNEEQ (oxymetazoline hydrochloride ophthalmic solution), [package insert]. Bridgewater, NJ: RVL Pharmaceuticals, Inc.; 2020.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 05 Oct. 2020].
3. Finsterer, Josef. "Ptosis: causes, presentation, and management." Aesthetic plastic surgery 27.3 (2003): 193-204.
4. Kersten, Robert C., Carlo de Conciliis, and Dwight R. Kulwin. "Acquired ptosis in the young and middle-aged adult population." Ophthalmology 102.6 (1995): 924-928.
5. Slonim, Charles B., et al. "Association of oxymetazoline hydrochloride, 0.1%, solution administration with visual field in acquired ptosis: a pooled analysis of 2 randomized clinical trials." JAMA ophthalmology 138.11 (2020): 1168-1175.

Last Reviewed: 7/21/21, 3/17/23, 3/15/24
Effective Date: 9/1/21, 4/15/24

Vascepa® (icosapent ethyl)
Prior Authorization Guidelines

Affected Medication(s)

- Vascepa oral capsule
- Icosapent ethyl capsule

FDA Approved Indication(s)

- As an adjunct to maximally tolerated statin therapy to reduce the risk of myocardial infarction, stroke, coronary revascularization, and unstable angina requiring hospitalization in adult patients with elevated triglyceride (TG) levels (≥ 150 mg/dL) and
  - Established cardiovascular disease OR
  - Diabetes mellitus and 2 or more additional risk factors for cardiovascular disease
- As an adjunct to diet to reduce TG levels in adult patients with severe (≥ 500 mg/dL) hypertriglyceridemia

Dosing

- Twice daily of a total 4 gram per day taken with food

Initial Authorization Criteria

1. Has Vascepa (icosapent ethyl) previously been approved by OHSU?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. What is the requested drug being used for? (Provide documentation of diagnosis)
   a. Hypertriglyceridemia, continue to #3
   b. Atherosclerotic cardiovascular disease prevention, continue to #5
   c. Other indication, clinical review required
3. Does the member have a triglyceride level of greater than 500 mg/dL confirmed by labs within 6 months? (Provide lab for review)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Did the member have a trial with inadequate response, intolerance, or a contraindication to fibrate, niacin or statin therapy (minimum 12 week trial)? (Provide supporting documentation)
   a. If yes, approve for 24 months unless otherwise specified
   b. If no, clinical review required
5. Is the member 45 years of age or older and have established cardiovascular disease confirmed by at least one of the following? (Provide supporting documentation)
   - Documented coronary artery disease defined as ≥50% stenosis in at least two major epicardial coronary arteries, prior myocardial infarction, OR prior hospitalization for high-risk non-ST-segment elevation acute coronary syndrome

Last Reviewed: 11/18/20, 1/19/22, 7/20/22, 7/21/23
Effective Date: 12/15/20, 3/1/22, 9/1/22

- Documented cerebrovascular or carotid disease defined as prior ischemic stroke, symptomatic carotid artery disease with ≥50% carotid arterial stenosis, asymptomatic carotid artery disease with ≥70% carotid arterial stenosis, OR history of carotid revascularization
- Documented peripheral arterial disease defined as ankle-brachial index (ABI) <0.9 with symptoms of intermittent claudication, or history of aorto-iliac or peripheral arterial intervention

a. If yes, continue to #8
b. If no, continue to #6

6. Is the member 50 years of age or older and have diabetes?

a. If yes, continue to #7
b. If no, clinical review required

7. Does the member have at least two of the following risk factors? (Provide supporting documentation)

- Men 55 years of age or older; women 65 years of age or older
- Cigarette smoker
- With Hypertension or on antihypertensive medication
- HDL-C ≤40 mg/dL for men or ≤50 mg/dL for women
- Hs-CRP >3.00 mg/L (0.3 mg/dL)
- Renal dysfunction: (CrCL>30mL/min and <60mL/min)
- Retinopathy defined as: non-proliferative retinopathy, pre-proliferative retinopathy, proliferative retinopathy, maculopathy, advanced diabetic eye disease or a history of photocoagulation
- Microalbuminuria or macroalbuminuria
- ABI <0.9 without symptoms of intermittent claudication

a. If yes, continue to #8
b. If no, clinical review required

8. Does the member have a triglyceride level between 135 mg/dL and 500 mg/dL within 6 months? (Provide lab for review)

a. If yes, continue to #9
b. If no, clinical review required

9. Is the member currently receiving maximally tolerated statin therapy and ezetimibe for four (4) consecutive weeks and will continue with therapy or have a documented intolerance or contraindication to the use of these agents? (Provide supporting documentation)

a. If yes, approve for 24 months unless otherwise specified
b. If no, clinical review required

Reauthorization Criteria

1. Has this member been seen within the past 12 months for treatment of hypertriglyceridemia or prevention of atherosclerotic cardiovascular disease?

a. If yes, approve for 24 months unless otherwise specified
b. If no, clinical review required

Note:

Last Reviewed: 11/18/20, 1/19/22, 7/20/22, 7/21/23
Effective Date: 12/15/20, 3/1/22, 9/1/22

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Vascepa (icosapent ethyl) [Prescribing Information]. Bridgewater, NJ: Amarin Pharma, Inc. July 2020.
2. Bhatt DL, Steg PG, Miler M, et al. Cardiovascular Risk Reduction with Icosapent Ethyl for Hypertriglyceridemia. New England Journal of Medicine. 2019;380(1):11-22.
3. Orringer, CE, Jacobson, TA, Maki, KC. National Lipid Association Scientific Statement on the use of icosapent ethyl in statin-treated patients with elevated triglycerides and high or very-high ASCVD risk. J Clin Lipidol. 2019;13(6):860-72.

Last Reviewed: 11/18/20, 1/19/22, 7/20/22, 7/21/23
Effective Date: 12/15/20, 3/1/22, 9/1/22

Vecamyl® (mecamylamine hydrochloride)
Prior Authorization Guidelines

Affected Medication(s)

- Vecamyl oral tablet

FDA Approved Indication(s)

- Management of moderately severe to severe essential hypertension and in uncomplicated cases of malignant hypertension

Dosing

- Refer to package insert for specific dosing recommendations

Initial Authorization Criteria

1. Is the request for continuation of Vecamyl (mecamylamine hydrochloride) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a trial with insufficient response, intolerance, or contraindication to at least 3 formulary anti-hypertensives from 3 different therapeutic classes at maximum tolerated doses (i.e. ACE-inhibitors, ARBs, thiazides, calcium channel blockers, beta-blockers, alpha-blockers)? (Provide relevant past treatment history)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member demonstrate a positive response to therapy as defined by a decrease in blood pressure from baseline? (Provide supporting documentation)
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

Last Reviewed: 11/7/18, 11/20/19, 7/21/21, 11/16/22, 9/15/2023
Effective Date: 1/1/19

References:

1. Vecamyl Prescribing Information. Colorado Springs, Co: Nexgen Pharma; April 2015. Available at: www.vecamyl.com.
2. James PA, Oparil S, Carter BL et al. 2014 evidence-based guideline for the management of high blood pressure in adults: report from the panel members appointed to the Eighth Joint National Committee (JNC 8). JAMA. 2014 Feb 5;311(5):507-20. doi: 10.1001/jama.2013.284427.
3. Chobanian AV, Bakris GL, Black HR et al. Seventh report of the Joint National Committee on Prevention, Detection, Evaluation, and Treatment of High Blood Pressure. Hypertension. 2003 Dec;42(6):1206-52. Epub 2003 Dec 1.
4. Gradman, AH. Rationale for triple-combination therapy for management of high blood pressure. J Clin Hypertens 2010; 12:869-878. doi: 10.1111/j.1751-7176.2010.00360.x

Last Reviewed: 11/7/18, 11/20/19, 7/21/21, 11/16/22, 9/15/23
Effective Date: 1/1/19

Veltassa® (patiromer)
Prior Authorization Guidelines

Affected Medication(s)

- Veltassa powder for suspension

FDA Approved Indication(s)

- Treatment of hyperkalemia (should not be used as an emergency treatment for life-threatening hyperkalemia)

Dosing

- Initially 8.4 g once daily
- Titrate as needed to maximum of 25.2 g once daily to reach desired serum potassium concentration

Initial Authorization Criteria

1. Is the request for continuation of Veltassa (patiromer) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the baseline potassium level received? (Provide documentation of lab)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Have potentially hyperkalemia contributing medications such as NSAIDs, ACEI, ARB, or aldosterone antagonists been reduced to lowest effective dose or discontinued if clinically appropriate? (Provide documentation of dose reduction/discontinuation or rationale why patient is not a candidate to do so)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member follow a low potassium diet? (Provide documentation of diet with 3 or less grams of potassium per day)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have a trial with inadequate response, intolerance, or contraindication to treatment with sodium polystyrene sulfonate or Lokelma? (Provide documentation of trial, intolerance, or contraindication)
    a. If yes, continue to #8
    b. If no, clinical review required

Last Reviewed: 11/21/18, 5/19/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 6/15/21

8. Is the treatment being prescribed by, or in consultation with, a nephrologist or cardiologist?
    a. If yes, approve for 6 months unless otherwise indicated
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a positive response to therapy as defined as a decrease in serum potassium? (Provide documentation of decrease in serum potassium compared to pre-treatment)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a nephrologist or cardiologist?
    a. If yes, approve for 12 months unless otherwise indicated
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Veltassa [Prescribing Information] Redwood City, CA: Relypsa, Inc; December 2021. Available at: www.veltassa.com.
2. Bakris GL, Pitt B, Weir MR, et al. Effect of patiromer on serum potassium levels in patients with hyperkalemia and diabetic kidney disease: The AMETHYST-DN randomized clinical trial. JAMA. 2015; 314(2):151-161.
3. Weir MR, Bakris GL, Bushinsky DA, et al; for the OPAL-HK Investigators. Patiromer in patients with kidney disease and hyperkalemia receiving RAAS inhibitors. N Engl J Med. 2015; 372(3):211-221.
4. Clase, Catherine M., et al. "Potassium homeostasis and management of dyskalemia in kidney diseases: conclusions from a Kidney Disease: Improving Global Outcomes (KDIGO) Controversies Conference." Kidney international 97.1 (2020): 42-61.

Last Reviewed: 11/21/18, 5/19/21, 11/16/22, 11/17/23
Effective Date: 1/1/19, 6/15/21

Verquvo® (vericiguat)
Prior Authorization Guidelines

Affected Medication(s)

- Verquvo oral tablet

FDA Approved Indication(s)

- Reduce risk of cardiovascular death and heart failure (HF) hospitalization following a hospitalization for heart failure or need for outpatient IV diuretics in adults with symptomatic chronic HF and ejection fraction less than 45%

Dosing

- Initial: 2.5 mg once daily with meals
- Target maintenance dose: 10 mg once daily

Initial Authorization Criteria

1. Is the request for continuation of Verquvo (vericiguat) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member aged 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have NYHA class II-IV heart failure with a left ventricular ejection fraction of 45% or less? (Provide lab for review)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a worsening heart failure event defined as one of the following?
    - History of a previous heart failure hospitalization within the last 6 months
    - Use of outpatient IV diuretics for heart failure within the last 3 months

    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the member currently taking a drug from each of the following classes at the maximum tolerated dose unless contraindicated? (Provide supporting documentation)
    - Beta Blocker: carvedilol 25mg twice daily, metoprolol succinate 200mg/day
    - ACE inhibitor/ARB/ARNI: captopril 50mg three times daily, enalapril 10mg twice daily, lisinopril 20-40mg/day, ramipril 5mg twice dialy, losartan 150mg/day, Entresto sacubitril 97mg/valsartan 103mg twice daily
    - Mineralocorticoid receptor agonist: spironolactone 25mg/day

Last Reviewed: 5/19/21, 7/20/22, 7/21/23
Effective Date: 6/15/21, 9/1/22

a. If yes, continue to #7
b. If no, clinical review required

7. Does the member have documentation of an inadequate response, intolerance, or contraindication to Jardiance (empagliflozin) or Farxiga (dapagliflozin)?

    a. If yes, continue to #8
    b. If no, clinical review required

8. Is the treatment being prescribed by or in consult with a cardiologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Is the treatment being prescribed by or in consultation with a cardiologist?

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is documentation provided that the member is experiencing successful response to Verquvo? (Provide updated clinical information for review such as reduction in HF hospitalizations compared to baseline, improvement in HF symptoms, reduction in need for IV diuretics etc.)

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. VERQUVO (vericiguat) tablets, [package insert]. Whithouse Station, NJ: Merck Sharp Dohme.; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 22 MAR. 2021].
3. Armstrong PW, Roessig L, Patel MJ, Anstrom KJ, Butler J, Voors AA, Lam CSP, Ponikowski P, Temple T, Pieske B, Ezekowitz J, Hernandez AF, Koglin J, O'Connor CM. A Multicenter, Randomized, Double-Blind, Placebo-Controlled Trial of the Efficacy and Safety of the Oral Soluble Guanylate Cyclase Stimulator: The VICTORIA Trial. JACC Heart Fail. 2018 Feb;6(2):96-104. doi: 10.1016/j.jchf.2017.08.013. Epub 2017 Oct 11. PMID: 29032136.
4. Yancy CW, Jessup M, Bozkurt B, Butler J, Casey DE Jr, Drazner MH, Fonarow GC, Geraci SA, Horwich T, Januzzi JL, Johnson MR, Kasper EK, Levy WC, Masoudi FA, McBride PE, McMurray JJ, Mitchell JE, Peterson PN, Riegel B, Sam F, Stevenson LW, Tang WH, Tsai EJ, Wilkoff BL; American College of Cardiology Foundation; American Heart Association Task Force on Practice Guidelines. 2013 ACCF/AHA guideline for the management

Last Reviewed: 5/19/21, 7/20/22, 7/21/23
Effective Date: 6/15/21, 9/1/22

of heart failure: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2013 Oct 15;62(16):e147-239. doi: 10.1016/j.jacc.2013.05.019. Epub 2013 Jun 5. PMID: 23747642.
5. Yancy, Clyde W., et al. "2017 ACC/AHA/HFSA focused update of the 2013 ACCF/AHA guideline for the management of heart failure: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America." Journal of the American College of Cardiology 70.6 (2017): 776-803.
6. McDonagh, Theresa A., et al. "2021 ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure: Developed by the Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC) With the special contribution of the Heart Failure Association (HFA) of the ESC." European heart journal 42.36 (2021): 3599-3726.

Vijoice® (alpelisib)
Prior Authorization Guidelines

Affected Medication(s)

- Vijoice (alpelisib) oral tablet

FDA Approved Indication(s)

- Treatment of adult and pediatric patients 2 years of age and older with severe manifestations of PIK3CA-Related Overgrowth Spectrum (PROS)

Dosing

- Adult: 250mg once daily until disease progression or unacceptable toxicity
- Pediatric: Initial dose of 50mg once daily, with a potential to increase to 125mg once daily after 24 weeks in patients 6-17 years old

Initial Authorization Criteria

1. Is the request for continuation of Vijoice (alpelisib) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member aged 2 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation of PIK3CA-Related Overgrowth Spectrum (PROS) with confirmed PIK3CA gene mutation? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have severe clinical manifestations resulting from a lesion associated with PROS and is the lesion both inoperable and causing functional impairment? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Has the member previously trialed sirolimus for at least 6 months with inadequate response or does the member have a documented intolerance or contraindication to sirolimus? (Note: inadequate response defined as continuing to have severe clinical manifestations resulting from the lesion with the lesion being inoperable and causing functional impairment despite current treatment)
    a. If yes, continue to #7
    b. If no, clinical review required

Last Reviewed: 7/20/22, 7/21/23
Effective Date: 9/1/22

7. Is Vijoice (alpelisib) being prescribed by, or in consult with, a specialist with experience in the treatment of PROS?
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the treatment being prescribed by, or in consultation with, a specialist with experience in the treatment of PROS?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Were updated chart notes (within the past year) provided with documentation of significant clinical response to therapy defined by the following? (Provide supporting documentation)
    - ≥20 % reduction from baseline in the sum of measurable target lesion volume confirmed by imaging
    - Absence of a ≥20% increase from baseline in any target lesion, progression of non-target lesions, or appearance of new lesion

    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. VIJOICE® (alpelisib) tablets, [package insert]. East Hanover, NJ: Novartis Pharmaceuticals; 2022.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 26 May. 2022].
3. Canaud, G., et al. "LBA23 EPIK-P1: Retrospective chart review study of patients (pts) with PIK3CA-related Overgrowth Spectrum (PROS) who have received alpelisib (ALP) as part of a compassionate use programme." Annals of Oncology 32 (2021): S1297.
4. Douzou S, Rawson M, Faivre L, et al. A standard of care for individuals with PIK3CA related disorders: an international expert consensus statement. Clinical Genetics. 2022; 101:32-47.
5. Canuad G, Hammill AM, Adams D, Vikkula M, and Keppler-Noreuil KM. A review of mechanisms of disease across PIK3CA-related disorders with vascular manifestations. Orphanet J Rare Dis. 2021;16:306.
6. Parker, Victoria ER, et al. "Safety and efficacy of low-dose sirolimus in the PIK3CA-related overgrowth spectrum." Genetics in Medicine 21.5 (2019): 1189-1198.

Last Reviewed: 7/20/22, 7/21/23
Effective Date: 9/1/22

Vivjoa® (oteseconazole)
Prior Authorization Guidelines

Affected Medication(s)

- Vivjoa oral capsule

FDA Approved Indication(s)

- Reduce the incidence of recurrent vulvovaginal candidiasis (RVVC) in females with a history of RVCC who are not of reproductive potential

Dosing

- Vivjoa only regimen: 600mg on day 1, 450mg on day 2, then 150mg once weekly for 11 weeks starting on day 14
- Fluconazole/Vivjoa regimen: fluconazole 150mg on day 1, 4, and 7. Vivjoa 150mg once daily for 7 days on days 14-20. Then, Vivjoa 150mg once weekly for 11 weeks starting on day 28.

Initial Authorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a diagnosis of recurrent vulvovaginal candidiasis defined by 3 or more episodes of vulvovaginal candidiasis in a 12 month period? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member a female of reproductive potential?
    a. If yes, clinical review required
    b. If no, continue to #4
4. Has the member trialed a minimum of 6-months of fluconazole therapy with persistent recurrent vulvovaginal candidiasis? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. VIVJOA™ (oteseconazole) capsules, [package insert]. Dunham, NC: Mycovia Pharmaceuticals, Inc.; 2022.

Last Reviewed: 9/21/22, 9/15/23
Effective Date: 11/1/22

2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 3 Aug. 2022].
3. Martens, Mark G., et al. "Phase 3 Study of the Safety and Efficacy of Oteseconazole in Treatment of Recurrent Vulvovaginal Candidiasis and Efficacy vs Fluconazole in Treatment of Acute Vulvovaginal Candidiasis Infections." American Journal of Obstetrics and Gynecology (2022).
4. Pappas, Peter G., et al. "Clinical practice guideline for the management of candidiasis: 2016 update by the Infectious Diseases Society of America." Clinical Infectious Diseases 62.4 (2016): e1-e50.

VMAT2 Inhibitors
Prior Authorization Guidelines

Affected Medication(s)

- Austedo (deutetrabenazine) oral tablet
- Austedo XR (deutetrabenazine) oral tablet
- Tetrabenazine oral tablet
- Xenazine (tetrabenazine) oral tablet
- Ingrezza (valbenazine) oral capsule
- Ingrezza (valbenazine) sprinkle capsule

FDA Approved Indication(s)

- **Austedo, Austedo XR:**
  - For the treatment of chorea associated with Huntington's disease
  - For the treatment of tardive dyskinesia in adults
- **Xenazine (tetrabenazine):**
  - For the treatment of chorea associated with Huntington's disease
- **Ingrezza:**
  - For the treatment of adults with chorea associated with Huntington's disease
  - For the treatment of tardive dyskinesia in adults

Dosing

- Refer to package insert for recommended dosing

Initial Authorization Criteria

1. Is the request for continuation of vesicular monoamine transporter 2 (VMAT2) inhibitor therapy?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. What is the diagnosis the medication is being requested for?
   a. Chorea associated with Huntington's disease, continue to corresponding criteria
   b. Tardive dyskinesia, continue to corresponding criteria
   c. Other indication, continue to corresponding criteria

Chorea associated with Huntington's Disease

1. Does the member have a diagnosis of Huntington's disease as defined by ALL of the following? (Provide supporting documentation)
   - DNA testing showing CAG expansion of $\geq$ 36
   - Family history (if known)

Last Reviewed: 11/7/18, 7/21/21, 11/16/22, 11/17/23, 5/17/24
Effective Date: 1/1/19, 12/20/23, 6/15/24

- Classic presentation (choreiform movements, psychiatric problems, and dementia)

a. If yes, continue to #2
b. If no, clinical review required

2. Is the member's chorea causing functional impairment in activities of daily life? (Provide supporting documentation)

a. If yes, continue to #3
b. If no, clinical review required

3. Is Austedo (deutetrabenazine), Austedo XR, or Ingrezza (valbenazine) being requested?

a. If yes, continue to #4
b. If no, continue to #5

4. Does the member have a trial with insufficient response, intolerance, or contraindication to tetrabenazine? (Provide documentation of trial with inadequate response, intolerance, or contraindication)

a. If yes, continue to #5
b. If no, clinical review required

5. Does the member have any of the following contraindications to the requested treatment?

- Suicidal, or have untreated or inadequately treated depression
- Hepatic impairment
- Taking concurrently with monoamine oxidase inhibitors (MAOIs) or reserpine
- Taking concurrently with other VMAT2 inhibitors

a. If yes, clinical review required
b. If no, continue to #6

6. Is the treatment being prescribed by, or in consultation with, a neurologist?

a. If yes, approve for 3 months unless otherwise specified
b. If no, clinical review required

<u>Tardive dyskinesia</u>

1. Has the patient been taking a dopamine receptor blocking agent for at least 3 months that contributed to the diagnosis? (Provide current/past medication history)

a. If yes, continue to #2
b. If no, clinical review required

2. Does the member have a diagnosis of moderate to severe tardive dyskinesia as defined by an Abnormal Involuntary Movement Scale (AIMS) score of ≥8 or an Extrapyramidal Symptom Rating Scale (ESRS) score of ≥20? (Provide supporting documentation and movement scale score)

a. If yes, continue to #3
b. If no, clinical review required

3. Is Austedo (deutetrabenazine) or Austedo XR being requested?

a. If yes, continue to #4
b. If no, continue to #5

4. Does the member have a trial with insufficient response, intolerance, or contraindication to Ingrezza (valbenazine)? (Provide documentation of trial with inadequate response, intolerance, or contraindication)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have any of the following contraindications to the requested treatment?
    - Suicidal, or have untreated or inadequately treated depression
    - Hepatic impairment
    - Taking concurrently with monoamine oxidase inhibitors (MAOIs) or reserpine
    - Taking concurrently with other VMAT2 inhibitors

    a. If yes, clinical review required
    b. If no, continue to #6
6. Is the treatment being prescribed by, or in consult with, a psychiatrist or neurologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Other Indications

1. Has the member tried and had an inadequate response OR does the member have a contraindication to ALL standard treatment options for the requested indication? (Provide all prior treatment history, contraindication if appropriate, and treatment plan)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have any of the following contraindications to the requested treatment?
    - Suicidal, or have untreated or inadequately treated depression
    - Hepatic impairment
    - Taking concurrently with monoamine oxidase inhibitors (MAOIs) or reserpine
    - Taking concurrently with other VMAT2 inhibitors

    a. If yes, clinical review required
    b. If no, approve for 6 months unless otherwise specified

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required

2. Does the member demonstrate a positive clinical response to therapy as defined by a decrease in chorea causing functional impairment OR an improvement in AIMS score of 2 or more points or improvement in ESRI score of 4 or more points compared to baseline? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consult with, a neurologist or psychiatrist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Ingrezza Prescribing Information. San Diego, CA: Neurocrine Biosciences, Inc. August 2022. Available at http://www.ingrezza.com.
2. Smith HS, Cox LR, Smith BR. Dopamine receptor antagonists. Annals of Palliative Medicine. July 2012; 1(2). DOI: 10.3978/j.issn.2224-5820.2012.07.09.
3. Bhidayasiri R, Fahn S, Weiner WJ, et al. Evidence-based guideline: Treatment of tardive syndromes. Report of the Guideline Development Subcommittee of the American Academy of Neurology. Neurology. 2013; 31: 463-469.
4. Xenazine (tetrabenazine) [Prescribing Information]. Deerfield, IL: Lundbeck Pharmaceuticals LLC. September 2017.
5. Austedo (deutetrabenazine) [Prescribing Information]. North Wales, PA: Teva Neuroscience, Inc. May 2022.
6. Xenazine. Micromedex. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: http://www.micromedexsolutions.com.
7. Arya, D., Khan, T., Margolius, A., & Fernandez, H. (2019). Tardive Dyskinesia: Treatment Update. Current Neurology and Neuroscience Reports, 19(9). doi: 10.1007/s11910-019-0976-1

Last Reviewed: 11/7/18, 7/21/21, 11/16/22, 11/17/23, 5/17/24
Effective Date: 1/1/19, 12/20/23, 6/15/24

Vowst® (fecal microbiota spores, live-brpk)
Prior Authorization Guidelines

Affected Medication(s)

- Vowst (fecal microbiota spores, live-brpk) oral capsules

FDA Approved Indication(s)

- To prevent the recurrence of Clostridioides difficile infection (CDI) in individuals 18 years of age and older following antibiotic treatment for recurrent CDI (rCDI)

Dosing

- 4 capsules orally once daily for 3 consecutive days

Initial Authorization Criteria

1. Is Vowst (fecal microbiota spores, live-brpk) being requested for an FDA approved indication? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member had a previous treatment course of Vowst for recurrent CDI?
    a. If yes, clinical review required
    b. If no, continue to #4
4. Has the member had 3 or more episodes of CDI within the past 12 months that were treated with oral vancomycin and/or Dificid (fidaxomicin)? (CDI defined as diarrhea for 2+ days and a confirmatory C. difficile toxin assay) (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Will the member have completed antibiotic treatment for recurrent CDI within 2 to 4 days prior to starting Vowst with resolution of active CDI? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the treatment plan include the use of magnesium citrate on the day before and at least 8 hours prior to taking the first dose of Vowst?
    a. If yes, continue to #7
    b. If no, clinical review required
7. Is the treatment being prescribed by, or in consultation with, a gastroenterologist or infectious disease specialist?
    a. If yes, approve for 7 days
    b. If no, clinical review required

Last Reviewed: 7/21/23
Effective Date: 9/15/23

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. VOWST (fecal microbiota spores, live-brpk) capsules, [package insert]. Cambridge, MA: Seres Therapeutics, Inc; 2023
2. Drugs@FDA: FDA Approved Drug Products. 2023. https://www.fda.gov/vaccines-blood-biologics/vowst [online] Available at: https://www.fda.gov/vaccines-blood-biologics/vowst [Accessed 14 Jun. 2023].
3. Kelly CR, Fischer M, Allegretti JR, et al. ACG Clinical Guidelines: Prevention, Diagnosis, and Treatment of Clostridioides difficile Infections [published correction appears in Am J Gastroenterol. 2022 Feb 1;117(2):358]. Am J Gastroenterol. 2021;116(6):1124-1147.
4. McDonald LC, Gerding DN, Johnson S, et al. Clinical Practice Guidelines for Clostridium difficile Infection in Adults and Children: 2017 Update by the Infectious Diseases Society of America (IDSA) and Society for Healthcare Epidemiology of America (SHEA). Clin Infect Dis. 2018;66(7):e1-e48.
5. Johnson S, Lavergne V, Skinner AM, et al. Clinical Practice Guideline by the Infectious Diseases Society of America (IDSA) and Society for Healthcare Epidemiology of America (SHEA): 2021 Focused Update Guidelines on Management of Clostridioides difficile Infection in Adults. Clin Infect Dis. 2021;73(5):e1029-e1044.

Voxzogo® (vosoritide)
Prior Authorization Guidelines

Affected Medication(s)

- Voxzogo (vosoritide subcutaneous solution)

FDA Approved Indication(s)

- To increase linear growth in pediatric patients with achondroplasia who are 5 years of age and older with open epiphyses

Dosing

- Refer to package insert for weight based dosing

Initial Authorization Criteria

1. Is the request for continuation of a previously approved Voxzogo (vosoritide) prior authorization with the same indication as the previous approval?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member age 5 to 17 years old?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a diagnosis of achondroplasia confirmed by molecular testing of FGFR3 gene? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the member's baseline height and growth velocity provided and is the member's growth velocity at least 1.5cm/yr? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is there documentation of open epiphyses? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required

Last Reviewed: 3/16/22, 3/17/23, 3/15/24
Effective Date: 4/15/22, 4/15/24

7. Has the member previously had or planning to have limb lengthening surgery? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #8
8. Is the medication being prescribed by, or in consultation with, a pediatric endocrinologist, orthopedist, or other prescriber specialized in the treatment of achondroplasia?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is Voxzogo (vosoritide) being requested for an FDA approved or major compendia supported indication? (Provide supporting documentation)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there documentation of continued open epiphyses? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has there been an increase in growth velocity compared to baseline and does the member's growth velocity remain greater than 1.5cm/yr? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the medication being prescribed by, or in consultation with, a pediatric endocrinologist, orthopedist, or other prescriber specialized in the treatment of achondroplasia?
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. VOXZOGO (vosoritide) for injection, [package insert]. Novato, CA: BioMarin Pharmaceutical, Inc; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 09 Dec. 2021].
3. Pauli, Richard M. "Achondroplasia: a comprehensive clinical review." Orphanet Journal of Rare Diseases 14.1 (2019): 1-49.
4. Ornitz, David M., and Laurence Legeai-Mallet. "Achondroplasia: Development, pathogenesis, and therapy." Developmental dynamics 246.4 (2017): 291-309.

Last Reviewed: 3/16/22, 3/17/23, 3/15/24
Effective Date: 4/15/22, 4/15/24

Vyndaqel® (tafamidis meglumine), Vyndamax® (tafamidis)
Prior Authorization Guidelines

Affected Medication(s)

- Vyndaqel oral capsule
- Vyndamax oral capsule

FDA Approved Indication(s)

- Treatment of the cardiomyopathy of wild type or hereditary transthyretin-mediated amyloidosis (ATTR-CM) in adults to reduce cardiovascular mortality and cardiovascular-related hospitalization

Dosing

- Vyndaqel: 80mg (four 20-mg capsules) orally one time daily
- Vyndamax: 61mg (one capsule) orally one time daily
- Vyndamax and Vyndaqel are not substitutable on a per mg basis

Initial Authorization Criteria

1. Is the request for continuation of Vyndaqel or Vyndamax therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member 18 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have documentation confirming the presence of a transthyretin (TTR) mutation or TTR precursor protein? (Provide supporting documentation)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have documentation of a biopsy that was found to be positive for amyloid deposits? (Provide documentation of biopsy)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member have cardiomyopathy caused by transthyretin-mediated amyloidosis? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the member have NYHA Class III or IV heart failure?

Last Reviewed: 11/20/19, 7/21/21, 7/20/22, 7/21/23
Effective Date: 1/1/20, 9/1/21

a. If yes, clinical review required
b. If no, continue to #8

8. Has the member had a liver transplant?

    a. If yes, clinical review required
    b. If no, continue to #9

9. Is the requested medication being prescribed by, or in consultation with, a cardiologist?

    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member demonstrated a positive clinical response to therapy defined as an improvement or stabilization in cardiomyopathy symptoms? (Provide supporting documentation)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Is the requested medication being prescribed by, or in consultation with, a cardiologist?

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. VYNDAQEL (tafamidis meglumine) and VYNDAMAX (tafamidis) oral capsules [package insert]. New York, NY: Pfizer Labs; 2019.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 30 July. 2019].
3. DailyMed - VYNDAQEL (tafamidis meglumine) and VYNDAMAX (tafamidis) oral capsules capsule. 2019. U.S. National Library of Medicine. National Institutes of Health. [online]
4. Maurer MS, Schwartz JH, Gundapaneni B, Elliott PM, Merlini G, Waddington-Cruz M, Kristen AV, Grogan M, Witteles R, Damy T, Drachman BM, Shah SJ, Hanna M, Judge DP, Barsdorf AI, Huber P, Patterson TA, Riley S, Schumacher J, Stewart M, Sultan MB, Rapezzi C; ATTR-ACT Study Investigators. Tafamidis Treatment for

Patients with Transthyretin Amyloid Cardiomyopathy. N Engl J Med. 2018 Sep 13;379(11):1007-1016. doi: 10.1056/NEJMoa1805689. Epub 2018 Aug 27.

5. Ando, Yukio, et al. "Guideline of transthyretin-related hereditary amyloidosis for clinicians." Orphanet journal of rare diseases 8.1 (2013): 1-18.

Wake Promoting Agents
Prior Authorization Guidelines

Affected Medication(s)

- Lumryz (sodium oxybate) packets for oral suspension
- Wakix (pitolisant hydrochloride) oral tablets
- Xyrem® (sodium oxybate) 500 mg/mL oral solution
- Sodium oxybate 500 mg/mL solution
- Xywav® (sodium oxybate) 500 mg/mL oral solution

FDA Approved Indication(s)

- Excessive somnolence: Narcolepsy
- Cataplexy in narcolepsy
- Idiopathic hypersomnia in adults (Xywav only)

Dosing

- Lumryz: 4.5g to 9g once nightly at bedtime
- Wakix: 8.9mg – 35.6mg once daily
- Xyrem and Xywav: Total nightly dose provided over two doses: bedtime and 2.5 – 4 hours later. Total daily dose range: 4.5 to 9 grams.

Initial Authorization Criteria

1. Is the request for continuation of a previously approved agent?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the requested drug FDA approved for this patient's age? (Note: policy covered agents have different FDA approved ages)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the treatment prescribed by or in consultation with a sleep specialist (e.g. neurology, pulmonology)?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Has this member's diagnosis been confirmed by overnight polysomnogram and multiple sleep latency test (MSLT)? (Note: narcolepsy may be confirmed by low levels of orexin or hypocretin within cerebrospinal fluid (<110pg/mL or less than one third of the normative value of the lab))
    a. If yes, continue to #6

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b. If no, clinical review required

6. What is the predominant symptom causing this request?

    a. Excessive daytime somnolence, continue to #7
    b. Cataplexy, approve for 3 months
    c. Idiopathic hypersomnia, continue to #7
    d. Other, clinical review required

7. Has this member had a documented trial with insufficient response, intolerance, or contraindication to at least one medication in each of the following groups? (Note: Requests for idiopathic hypersomnia do not require trial of solriamfetol (Sunosi))

    - Group 1: Modafinil or Armodafinil
    - Group 2: Stimulants (e.g. Methylphenidate, dextroamphetamine/amphetamine, etc.)
    - Group 3: Solriamfetol (Sunosi®)

    a. If yes, continue to #8
    b. If no, clinical review required

8. Have all other causes of excessive daytime sleepiness been ruled out or treated (e.g. obstructive sleep apnea, restless leg syndrome, periodic limb movements, substance abuse, etc.)?

    a. If yes, approve for 3 months
    b. If no, clinical review required

Reauthorization Criteria

1. Were updated chart notes provided with documentation of significant clinical response to therapy (e.g. reduction in cataplexy events or reduction in Epworth Sleepiness Scale [ESS])?

    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Morgenthaler TI, Kapur VK, Brown T, et al. Practice parameters for the treatment of narcolepsy and other hypersomnias of central origin. Sleep. 2007;30(12): 1705-1711.
2. Billiard M, Bassetti C, Dauvilliers Y, et al. EFNS guidelines on management of narcolepsy. Eur J Neurol. 2006;13(10):1035-1048.
3. Barateau L, Lopez R, Dauvilliers Y. Treatment options for narcolepsy. CNS Drugs. 2016;30:369-379.
4. Barateau L, Dauvilliers Y. Recent advances in treatment for narcolepsy. Ther Adv Neurol Disord. 2019;12:1-12.

Last Reviewed: 1/20/21, 5/18/22, 3/17/23, 7/21/23
Effective Date: 3/1/21, 7/15/22, 6/1/23, 9/15/23

5. Thorpy M, Bogan R. Update on the pharmacologic management of narcolepsy: mechanisms of action and clinical implications. Sleep Medicine. 2020;68:97-109.
6. Bogan RK, Thorpy MJ, Dauvilliers Y, et al. Efficacy and safety of calcium, magnesium, potassium and sodium oxybates (lower-sodium oxybate [LXB]; JZP-258) in a placebo-controlled, double-blind, randomized withdrawal study in adults with narcolepsu with cataplexy. SleepJ. 2020:1-13.
7. Dauvilliers Y, Bassetti C, Lammars GJ, et al. Pitolisant versus placebo or modafinil in patients with narcolepsy: a double-blind, randomised trial. Lancet Neurol. 2013; 12:1068-1075.
8. Dauvilliers Y, Arnulf I, Szakacs Z, et al. Long-term use of pitolisant to treat patients with narcolepsy: Harmony III study. SleepJ. 2019; 42(11):1-11.
9. Szakacs A, Dauvilliers Y, Mikhaylov V, et al. Safety and efficacy of pitolisant on cataplexy in patients with narcolepsy: a randomised, double-blind, placebo-controlled trial. Lancet Neurol. 2017;16:200-207.
10. Wakix (pitolisant) [Prescribing Information]. Plymouth Meeting, PA. Harmony Biosciences, LLC. Oct 2020.
11. Xyrem (sodium oxybate) [Prescribing Information]. Palo Alto, CA. Jazz Pharmaceuticals, Inc. Sept 2020.
12. Xywav (calcium, magnesium, potassium and sodium oxybate) [Prescribing Information]. Palo Alto, CA. Jazz Pharmaceuticalsm Inc. March 2022.
13. Dauvilliers, Yves, et al. "Safety and efficacy of lower-sodium oxybate in adults with idiopathic hypersomnia: a phase 3, placebo-controlled, double-blind, randomised withdrawal study." The Lancet Neurology 21.1 (2022): 53-65.
14. Lumryz (sodium oxybate for suspension, extended release) [Prescribing Information]. Chesterfield, MO. Avadel CNS Pharmaceuticals, LLC. June 2023.

Wegovy® (semaglutide)
Prior Authorization Guidelines

Affected Medication(s)

- Wegovy subcutaneous solution

FDA Approved Indication(s)

- Risk reduction of major adverse cardiovascular events (cardiovascular death, nonfatal MI, nonfatal stroke) in adults with established cardiovascular disease and either obesity or overweight
- As an adjunct to a reduced-calorie diet and increased physical activity for chronic weight management in adults with an initial BMI of ≥30 kg/m2 (obesity), or ≥27 kg/m2 (overweight) in the presence of at least one weight-related comorbid condition (i.e. hypertension, type 2 diabetes mellitus, dyslipidemia) and pediatric patients ≥12 years of age with an initial BMI at the ≥95th percentile standardized for age and sex (obesity)

Dosing

- 2.4 mg subcutaneously once weekly

Initial Authorization Criteria

1. Is the request for continuation of Wegovy (semaglutide) therapy for the same indication?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. What indication is the medication being requested for?
    a. Weight Management, clinical review required
    b. Risk reduction of major adverse cardiovascular events, continue #4
4. Is the member 45 years of age or older?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a BMI of 27 kg/m^2 or greater AND established cardiovascular disease (history of myocardial infarction, stroke, or symptomatic peripheral arterial disease)? (Provide supporting documentation)
    a. If yes, continue to #6
    b. If no, clinical review required
6. Is the member's current cardiovascular-related drug regimen (i.e. statin, antiplatelet, ACE-I, etc.) optimized and is the member adherent to this therapy? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, clinical review required
7. Does the treatment plan include health behavior and lifestyle modifications including physical activity goals, nutritional education, and behavior change? (Provide supporting documentation)
    a. If yes, approve for 6 months

Last Reviewed: 5/17/24
Effective Date: 6/15/24

b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA-approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is there clinical documentation confirming positive response to therapy as defined as 5% weight loss or more from baseline? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the treatment plan include on-going, age-appropriate health behavior and lifestyle modifications? (Provide supporting documentation)
    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Wegovy (semaglutide) [Prescribing Information]. Plainsboro, NJ: Novo Nordisk, Inc. March 2024.
2. Lincoff, A. Michael, et al. "Semaglutide and cardiovascular outcomes in obesity without diabetes." New England Journal of Medicine 389.24 (2023): 2221-2232.

Xermelo® (telotristat ethyl)
Prior Authorization Guidelines

Affected Medication(s)

- Xermelo oral tablet

FDA Approved Indication(s)

- For the treatment of carcinoid syndrome diarrhea in combination with somatostatin analog (SSA) therapy in adults inadequately controlled by SSA therapy

Dosing

- 250 mg three times daily

Initial Authorization Criteria

1. Is the request for continuation of Xermelo (telotristat) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have at least a 1-month trial with insufficient response to a somatostatin analog (SSA) (i.e. octreotide, lanreotide) at the maximum indicated dose? (Provide documentation of relevant past medication history and insufficient response)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is Xermelo (telotristat) being prescribed in combination with a somatostatin analog (SSA)? (Provide documentation of treatment regimen)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
2. Does the member demonstrate a positive clinical response to therapy defined as a reduction in bowel movement frequency or a reduction in urinary 5-hydroxyindoleacetic acid (5-HIAA) levels? (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required

Last Reviewed: 11/7/18, 1/20/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 3/1/21, 6/1/23, 4/15/24

3. Is Xermelo (telotristat) being prescribed in combination with a somatostatin analog (SSA) unless an intolerance or contraindication is present? (Provide documentation of treatment regimen)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Xermelo Prescribing Information. The Woodlands, TX: Lexicon Pharmaceuticals, Inc; January 2021. Available at: www.xermelo.com.
2. Kulke MH, Horsch D, Caplin ME, et al. Telotristat ethyl, a tryptophan hydroxylase inhibitor for the treatment of carcinoid syndrome. J Clin Oncol. 2016; 25(1): 14-23.
3. National Comprehensive Cancer Network. Neuroendocrine Tumors Version 1.2022. Available at: https://www.nccn.org/professionals/physician_gls/pdf/neuroendocrine.pdf
4. Kunz PL, Reidy-Lagunes D, Anthony LB, et al. North American Neuroendocrine Tumor Society (NANETS) guidelines: consensus guidelines for the management and treatment of neuroendocrine tumors. Pancreas. 2013; 42: 557-577.

Last Reviewed: 11/7/18, 1/20/21, 3/17/23, 3/15/24
Effective Date: 1/1/19, 3/1/21, 6/1/23, 4/15/24

Xifaxan® (rifaximin)
Prior Authorization Guidelines

Affected Medication(s)

- Xifaxan oral tablet

FDA Approved Indication(s)

- For the treatment of travelers' diarrhea (TD) caused by noninvasive strains of *Escherichia coli* in adults and pediatric patients 12 years of age and older
- For reduction in risk of overt hepatic encephalopathy (HE) recurrence in adults
- For the treatment of irritable bowel syndrome with diarrhea (IBS-D) in adults

Dosing

- Travelers' diarrhea: 200mg by mouth three times daily for 3 days
- Hepatic encephalopathy: 550mg by mouth twice daily or 400mg by mouth three times daily
- Irritable bowel syndrome with diarrhea: 550mg by mouth three times daily for 14 days
- Small intestinal bacterial overgrowth: 550mg by mouth three times daily for 14 days

Initial Authorization Criteria

1. Is the request for continuation of Xifaxan (rifaximin) therapy for treatment of hepatic encephalopathy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. What is the diagnosis that Xifaxan is being prescribed for?
    a. Travelers' diarrhea, continue to corresponding criteria
    b. Hepatic encephalopathy, continue to corresponding criteria
    c. Irritable bowel syndrome with diarrhea, continue to corresponding criteria
    d. Small intestinal bacterial overgrowth (SIBO), continue to corresponding criteria

Travelers' diarrhea

1. Is the member 12 years of age or older?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a trial with insufficient response, contraindication, or intolerance to TWO of the following regimens for the treatment of travelers' diarrhea? (Provide relevant past medication history or documentation of contraindication/intolerance)
    - Azithromycin 500-1000mg once daily for 1-3 days
    - Ciprofloxacin 500mg twice daily for 1-3 days
    - Levofloxacin 500mg once daily for 1-3 days

Last Reviewed: 11/7/18, 7/21/21, 7/20/22, 7/21/23
Effective Date: 1/1/19, 9/1/21, 9/1/22, 9/15/23

- Ofloxacin 200mg twice daily for 1-3 days

a. If yes, approve for 3 days
b. If no, clinical review required

Hepatic encephalopathy

1. Is the member 18 years of age or older?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a trial with insufficient response to lactulose in the past 30 days up to the maximum indicated dose? (Insufficient response defined as continued altered mental status) (Provide relevant past medication history)
    a. If yes, continue to #3
    b. If no, continue to #4
3. Will the member continue to take lactulose concurrently with Xifaxan?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required
4. Does the member have an intolerance or contraindication to lactulose? (Provide documentation of contraindication/intolerance)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have altered mental status?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Irritable bowel syndrome with diarrhea

1. Is the member 18 years of age or older?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a trial with insufficient response, intolerance, or contraindication to loperamide at the maximal indicated dose? (Provide relevant past medication history or documentation of contraindication/intolerance)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a trial with insufficient response, intolerance, or contraindication to an antispasmodic agent at the maximal indicated dose? (i.e. dicyclomine) (Provide relevant past medication history or documentation of contraindication/intolerance)
    a. If yes, continue to #4
    b. If no, clinical review required

4. Has the member had 3 or more previous Xifaxan (rifaximin) treatment courses for irritable bowel syndrome with diarrhea? (Provide relevant past medication history)
    a. If yes, clinical review required
    b. If no, approve for 14 days

Small intestinal bacterial overgrowth (SIBO)

1. Does the member have a diagnosis of small intestinal bacterial overgrowth (SIBO) confirmed by a positive carbohydrate breath test or a jejunal aspirate culture? (Provide supporting documentation)?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Does the member have a trial with insufficient response, intolerance, or contraindication to ONE of the following: amoxicillin/clavulanate, ciprofloxacin, trimethoprim-sulfamethoxazole, metronidazole, doxycycline, or tetracycline? (Provide relevant past medication history or documentation of contraindication/intolerance)
    a. If yes, approve for 14 days
    b. If no, continue to #3
3. Does the member have methane-predominant bacterial overgrowth and Xifaxan will be used in combination with neomycin? (Provide supporting documentation)
    a. If yes, approve for 14 days
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is Xifaxan (rifaximin) being used concurrently with lactulose unless a contraindication or intolerance is present? (Provide current treatment regimen or documentation of contraindication/intolerance)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member responding positively to therapy as defined by a decrease in symptoms? (Provide supporting documentation)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

Last Reviewed: 11/7/18, 7/21/21, 7/20/22, 7/21/23
Effective Date: 1/1/19, 9/1/21, 9/1/22, 9/15/23

References:

1. Xifaxan Prescribing Information. Bridgewater, NJ: Salix Pharmaceuticals; January 2018. Available at https://www.xifaxan.com/.
2. Vilstrup H, Amodio P, Bajaj J, et al. Hepatic encephalopathy in chronic liver disease: 2014 practice guideline by AASLD-EASL. Hepatology. 2014; 60 (2): 715-735.
3. Weinberg DS, Smalley W, Heidelbaugh JJ, Shahnaz S. American Gastroenterological Association Institute guideline on the pharmacological management of irritable bowel syndrome. Gastroenterology. 2014; 147: 1146-1149.
4. Riddle M, Connor B, Beeching N, et al. Guidelines for the prevention and treatment of travelers' diarrhea: a graded expert panel report. J Travel Med. 2017 Apr 1;24(suppl_1):S57-S74.
5. Centers for Disease Control and Prevention (CDC. CDC Yellow Book 2020: health information for international travel. Oxford University Press, 2019.
6. Pimentel, Mark, et al. "ACG clinical guideline: small intestinal bacterial overgrowth." Official journal of the American College of Gastroenterology| ACG 115.2 (2020): 165-178.

Last Reviewed: 11/7/18, 7/21/21, 7/20/22, 7/21/23
Effective Date: 1/1/19, 9/1/21, 9/1/22, 9/15/23

Zelapar® (selegiline hydrochloride)
Prior Authorization Guidelines

Affected Medication(s)

- Zelapar oral disintegrating tablet

FDA Approved Indication(s)

- As an adjunct in the management of patients with Parkinson's disease being treated with levodopa/carbidopa who exhibit deterioration in the quality of their response to this therapy

Dosing

- Initially, 1.25 mg once daily for at least 6 weeks
- May increase dose to 2.5 mg once daily after initial 6 weeks

Initial Authorization Criteria

1. Is the request for continuation of Zelapar (selegiline hydrochloride) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member currently on levodopa/carbidopa therapy that will be continued concurrently with requested medication? (Provide relevant medication history and treatment plan)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Is the member experiencing a deterioration in the quality of their clinical response to levodopa/carbidopa? (Provide supporting documentation of a decline in response to levodopa/carbidopa demonstrated by at least 3 hours of "off" time per day)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have a trial with inadequate response to, or rationale for avoiding therapy with, BOTH generic selegiline oral tablets/capsules and generic rasagiline oral tablets? (Provide supporting documentation of trials with inadequate responses)
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)

Last Reviewed: 11/21/18, 1/20/21, 5/19/23, 5/17/24
Effective Date: 1/1/19, 3/1/21

a. If yes, continue to #2
b. If no, clinical review required

2. Is the member currently on levodopa/carbidopa therapy that will be continued concurrently with requested medication? (Provide relevant medication history and treatment plan)

    a. If yes, continue to #3
    b. If no, clinical review required

3. Does the member have a positive clinical response to therapy as defined by a decrease in frequency or duration of "off" episodes? (Provide supporting documentation of a decrease in frequency or duration of off episodes)

    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

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References:

1. Zelapar (selegiline) [prescribing information.] Bridgewater, NJ: Valeant Pharmaceuticals North America LLC; Aug 2016.

Last Reviewed: 11/21/18, 1/20/21, 5/19/23, 5/17/24
Effective Date: 1/1/19, 3/1/21, 6/15/24

Zeposia® (ozanimod)
Prior Authorization Guidelines

Affected Medication(s)

- Zeposia (ozanimod) capsule

FDA Approved Indication(s)

- For treatment of adult patients with relapsing forms of multiple sclerosis, to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease
- For treatment of adult patients with moderately to severely active ulcerative colitis (UC)

Dosing

- 0.92 mg orally once daily

Initial Authorization Criteria

1. Is the request for continuation therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Will the requested medication be used concurrently with other biologic or MS therapy? (examples: Enbrel, Dupixent, Cosentyx, Cimzia, Skyrizi, Otezla, Gilenya, etc)
    a. If yes, clinical review required
    b. If no, continue to #4
4. What is the diagnosis that the medication is being requested for?
    a. Multiple sclerosis, continue to corresponding criteria
    b. Ulcerative colitis, continue to corresponding criteria
    c. Other indication, clinical review required

Multiple Sclerosis (MS)

1. Is an MRI result consistent with multiple sclerosis received? (Provide MRI for review)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have a history of inadequate response, intolerance or contraindication to at least one drug from each of the following groups? (Provide supporting documentation)

Last Reviewed: 11/16/22, 7/21/23, 3/15/24
Effective Date: 1/1/23, 9/15/23, 4/15/24

- Glatiramer acetate
- Dimethyl fumarate or Teriflunomide
- Fingolimod

a. If yes, continue to #4
b. If no, clinical review required

4. Will the requested medication be used with other disease-modifying therapy for multiple sclerosis?
   a. If yes, clinical review required
   b. If no, continue to #5
5. Is the requested multiple sclerosis agent being prescribed by or in consultation with a neurologist?
   a. If yes, approve for 12 months unless otherwise specified
   b. If no, clinical review required

Ulcerative Colitis (UC)

1. Does the member currently have active Ulcerative Colitis? (Provide documentation of diagnosis confirmed by endoscopy, colonoscopy, or sigmoidoscopy with Mayo score of greater than 2)
   a. If yes, continue to #2
   b. If no, clinical review required
2. Does the member have a documented inadequate response, intolerance, or contraindication to TWO of the following: preferred adalimumab biosimilar, Simponi (golimumab), or Stelara (ustekinumab)? (Provide documentation of inadequate responses, contraindications, and/or intolerances)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the medication being prescribed by, or in consultation with, a gastroenterologist?
   a. If yes, approve for 6 months unless otherwise specified
   b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #2
   b. If no, clinical review required
2. Will the updated chart note (dated within 1 year) provided with documentation of significant clinical response to therapy? (Provide supporting documentation)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Will the requested medication be used concurrently with other biologic or MS therapy? (examples: Enbrel, Dupixent, Cosentyx, Cimzia, Skyrizi, Otezla, Gilenya, etc)

Last Reviewed: 11/16/22, 7/21/23, 3/15/24
Effective Date: 1/1/23, 9/15/23, 4/15/24

a. If yes, clinical review required
b. If no, continue to #4

4. Is the treatment being prescribed by or in consultation with an appropriate specialist?

a. If yes, approve for 12 months unless otherwise specified
b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Zeposia (ozanimod) [Prescribing Information]. Summit, NJ: Celgene Corporation. September 2022.
2. Rae-Grand A, Day GS, Marrie RA, et al. Practice guideline recommendations summary: Disease-modifying therapies for adults with multiple sclerosis. Neurology. 2018;90(17):777-788. Available at: http://n.neurology.org/content/neurology/90/17/777.full.pdf.
3. Feuerstein JD, Isaacs KL, Schneider Y, et al. AGA Clinical Practice Guidelines on the Management of Moderate to Severe Ulcerative Colitis. Gastroenterology 2020;158:1450-1461. Available at: https://www.gastrojournal.org/article/S0016-5085(20)30018-4/pdf
4. Harbord M, Eliakim R, Bettenworth D, et al. Third European Evidence-based Consensus on Diagnosis and Management of Ulcerative Colitis. Part 2: Current Management. Journal of Crohn's and Colitis. 2017;11(7):769-84. Available at: https://academic.oup.com/ecco-jcc/article/11/7/769/2962457.

Zilbrysq® (zilucoplan)
Prior Authorization Guidelines

Affected Medication(s)

- Zilbrysq (zilucoplan) subcutaneous injection

FDA Approved Indication(s)

- Treatment of generalized myasthenia gravis (gMG) in adult patients who are anti-acetylcholine receptor (AChR) antibody positive

Dosing

- Subcutaneous daily dose:
  - Less than 56 kg: 16.6 mg
  - 56 to 77 kg: 23 mg
  - 77 kg or greater: 32.4 mg

Initial Authorization Criteria

1. Is the request for continuation of therapy with the same medication for the same indication?
   a. If yes, continue to Reauthorization
   b. If no, continue to #2
2. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
   a. If yes, continue to #3
   b. If no, clinical review required
3. Is the member 18 years of age or older? (Provide supporting documentation)
   a. If yes, continue to #4
   b. If no, clinical review required
4. Does the member have Myasthenia Gravis Foundation of America (MGFA) Clinical Classification of Class II to IV disease? (Provide supporting documentation)
   a. If yes, continue to #5
   b. If no, clinical review required
5. Does the member have a positive serologic test for anti-acetylcholine receptor (AChR) antibodies? (Provide supporting documentation)
   a. If yes, continue to #6
   b. If no, clinical review required
6. Has the member had a thymectomy? (Note: Applicable only to patients with thymomas OR non-thymomatous patients who are 50 years of age or younger)
   a. If yes or N/A, continue to #7
   b. If no, clinical review required
7. Does the member have MG-Activities of Daily Living (MG-ADL) total score of ≥6? (Provide supporting documentation)

Last Reviewed: 3/15/24
Effective Date: 4/15/24

a. If yes, continue to #8
b. If no, clinical review required

8. Will the member avoid or use with medications known to worsen or exacerbate symptoms of MG (e.g., certain antibiotics, beta-blockers, botulinum toxins, hydroxychloroquine, etc.)? (Provide supporting documentation)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Has the member had an inadequate response after a minimum one-year trial with two (2) or more immunosuppressive therapies (e.g., corticosteroids plus an immunosuppressant such as azathioprine, cyclosporine, mycophenolate, etc.) or did the member require chronic treatment with plasmapheresis or plasma exchange (PE) or intravenous immunoglobulin (IVIG) in addition to immunosuppressant therapy? (Provide supporting documentation)

    a. If yes, continue to #10
    b. If no, clinical review required

10. Is Zilbrysq (zilucoplan) being prescribed by, or in consult with, a specialist experienced in treating myasthenia gravis?

    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported? (Provide supporting documentation)

    a. If yes, continue to #2
    b. If no, clinical review required

2. Has the member developed a severe bone marrow failure syndrome, experienced spontaneous disease remission, or received a curative allogeneic stem cell transplant?

    a. If yes, clinical review required
    b. If no, continue to #3

3. Were updated chart notes (within past year) provided with documentation of significant clinical response to therapy received? (Ex. improvement from baseline in Myasthenia Gravis-Specific Activities of Daily Living scale (MG-ADL) total score or Quantitative Myasthenia Gravis (QMG) total score) (Provide supporting documentation)

    a. If yes, continue to #4
    b. If no, clinical review required

4. Is Zilbrysq (zilucoplan) being prescribed by, or in consult with, a specialist experienced in treating myasthenia gravis?

    a. If yes, approve for 12 months
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as

medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. ZILBRYSQ (zilucoplan) subcutaneous injection, [package insert]. Smyrna, GA.: UCB, Inc; 2024.
2. Drugs@FDA: FDA Approved Drug Products. 2022. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 17 Jan. 2024].
3. Sanders DB, Wolfe GI, Benatar M, et al. International consensus guidance for management of myasthenia gravis: Executive summary. Neurology. 2016;87(4):419-425.
4. Narayanaswami P, Sanders DB, Wolfe G, et al. International Consensus Guidance for Management of Myasthenia Gravis: 2020 Update. Neurology. 2021;96(3):114-122.
5. Howard JF Jr, Bresch S, Genge A, et al. Safety and efficacy of zilucoplan in patients with generalised myasthenia gravis (RAISE): a randomised, double-blind, placebo-controlled, phase 3 study. Lancet Neurol. 2023;22(5):395-406.

Zokinvy® (lonafarnib)
Prior Authorization Guidelines

Affected Medication(s)

- Zokinvy oral capsule

FDA Approved Indication(s)

- Patients 12 months of age and older with a body surface area of 0.39m^2 and above:
  - To reduce risk of mortality in Hutchinson-Gilford Progeria Syndrome
  - For Treatment of processing-deficient Progeroid Laminopathies with either:
    - Heterozygous LMNA mutation with progerin-like protein accumulation.
    - Homozygous or compound heterozygous ZMPSTE24 mutations.

Dosing

- Start at 115 mg/m^2 twice daily with morning and evening meals. After 4 months, increase to 150 mg/m^2 twice daily.

Initial Authorization Criteria

1. Is the request for renewal of a previously approved Zokinvy (lonafarnib) prior authorization with the same indication?

   a. If yes, continue to Reauthorization
   b. If no, continue to #2

2. Is this being requested for an FDA or major compendia supported indication?

   a. If yes, continue to #3
   b. If no, clinical review required

3. Does the member have a confirmed diagnosis of one of the following? (Provide supporting documentation)
   - HGPS confirmed by G608G mutation in the lamin A gene
   - Processing-deficient Progeroid Laminopathy with either of the following:
     - Heterozygous LMNA mutation with progerin-like protein accumulation.
     - Homozygous or compound heterozygous ZMPSTE24 mutations.

   a. If yes, continue to #4
   b. If no, clinical review required

4. Is the member 12 months of age or older?

   a. If yes, continue to #5
   b. If no, clinical review required

5. Is the member's BSA (or height and weight) provided and is dosing consistent with FDA approved dosing? (Provide BSA for review)

   a. If yes, continue to #6
   b. If no, clinical review required

Last Reviewed: 7/21/21, 3/17/23, 1/19/24
Effective Date: 9/1/21

6. Is there documentation of baseline monitoring and planned ongoing monitoring of all of the following? (Provide baseline labs and monitoring plan for review)
    - Comprehensive metabolic panel
    - CBC
    - Ophthalmological evaluation
    - Blood pressure

    a. If yes, continue to #7
    b. If no, clinical review required

7. Does the member's baseline monitoring meet all of the following? (Provide baseline labs for review)
    - APC >1,000/ml
    - Platelets >75,000/ml (transfusion independent)
    - Hemoglobin >9g/dl
    - Creatinine ≤ 1.5 ULN for age or GFR >70ml/min/1.73m^2
    - Bilirubin ≤ 1.5 ULN for age
    - ALT and AST <5 x normal range for age

    a. If yes, continue to #8
    b. If no, clinical review required

8. Is there documentation of avoidance of strong CYP3A inhibitors/inducers, midazolam, lovastatin, simvastatin, or atorvastatin? (Provide supporting documentation)

    a. If yes, continue to #9
    b. If no, clinical review required

9. Is the member a female of reproductive potential?

    a. If yes, continue to #10
    b. If no, continue to #11

10. Does the member have documentation of a negative pregnancy test and documentation of contraceptive use throughout planned treatment? (Provide supporting documentation)

    a. If yes, continue to #11
    b. If no, clinical review required

11. Is the requested medication being prescribed by, or in consultation with, a specialist with experience in treating progeria and/or progeroid laminopathies?

    a. If yes, approve for 4 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is Zokinvy (lonafarnib) being requested for an FDA approved or major compendia supported indication? (Provide supporting documentation)

    a. If yes, continue to #2

b. If no, clinical review required

2. Is there documentation of disease stabilization compared to natural disease progression? (Provide supporting documentation)

   a. If yes, continue to #3
   b. If no, clinical review required

3. Is the requested medication being prescribed by, or in consultation with, a specialist with experience in treating progeria and/or progeroid laminopathies?

   a. If yes, approve for 12 months
   b. If no, clinical review required

Note:

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References:

1. ZOKINVY (lonafarnib) capsules, [package insert]. Palo Alto, CA: Eiger Biopharmaceuticals, Inc.; 2021.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 5 FEB. 2021].
3. Gordon, Leslie B., et al. "Impact of farnesylation inhibitors on survival in Hutchinson-Gilford progeria syndrome." Circulation 130.1 (2014): 27-34.
4. Gordon, Leslie B., et al. "Association of lonafarnib treatment vs no treatment with mortality rate in patients with Hutchinson-Gilford progeria syndrome." Jama 319.16 (2018): 1687-1695.
5. Gordon, Leslie B., et al. "Clinical trial of the protein farnesylation inhibitors lonafarnib, pravastatin, and zoledronic acid in children with Hutchinson-Gilford progeria syndrome." Circulation 134.2 (2016): 114-125.
6. Gordon, Leslie B., et al. "Clinical trial of a farnesyltransferase inhibitor in children with Hutchinson–Gilford progeria syndrome." Proceedings of the National Academy of Sciences 109.41 (2012): 16666-16671.

Last Reviewed: 7/21/21, 3/17/23, 1/19/24
Effective Date: 9/1/21

Zontivity® (vorapaxar)
Prior Authorization Guidelines

Affected Medication(s)

- Zontivity oral tablet

FDA Approved Indication(s)

- Reduction of thrombotic cardiovascular events in patients with a history of myocardial infarction (MI) or with peripheral arterial disease (PAD). ZONTIVITY has been shown to reduce the rate of a combined endpoint of cardiovascular death, MI, stroke, and urgent coronary revascularization

Dosing

- 2.08 mg orally once daily

Initial Authorization Criteria

1. Is the request for continuation of Zontivity (vorapaxar) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Does the member have documentation of a previous myocardial infarction or current peripheral artery disease? (Provide relevant medical history)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Will the member concurrently be taking aspirin and/or clopidogrel with the requested medication? (Provide documentation of treatment plan)
    a. If yes, continue to #5
    b. If no, clinical review required
5. Does the member have history of a stroke, transient ischemic attack (TIA), intracranial hemorrhage (ICH), or active pathological bleeding? (Provide supporting documentation)
    a. If yes, clinical review required
    b. If no, continue to #6
6. Is the treatment being prescribed by, or in consultation with, a cardiologist?
    a. If yes, approve for 6 months unless otherwise specified
    b. If no, clinical review required

Reauthorization Criteria

Last Reviewed: 11/21/18, 1/20/21, 5/18/22, 7/21/23
Effective Date: 1/1/19, 7/15/22

1. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Will the member concurrently be taking aspirin and/or clopidogrel with the requested medication? (Provide documentation of treatment plan)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member had a stroke, transient ischemic attack (TIA), or intracranial hemorrhage (ICH) while taking Zontivity?
    a. If yes, clinical review required
    b. If no, continue to #4
4. Does the member have an absence of serious adverse reactions from therapy (i.e. bleeding)?
    a. If yes, continue to #5
    b. If no, clinical review required
5. Is the treatment being prescribed by, or in consultation with, a cardiologist?
    a. If yes, approve for 12 months unless otherwise specified
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Zontivity [prescribing information]. Whitehouse Station, NJ: Merck & Co. Inc.; April 2021.
2. Morrow D, Braunwald E, Bonaca M, et. al. Vorapaxar in the Secondary Prevention of Atherothrombotic Events. New England Journal of Medicine 2012;366:1404-13.
3. Smith S, Benjamin E, Bonow R, et. al. AHA/ACCF Secondary Prevention and Risk Reduction Therapy for Patients with Coronary and Other Atherosclerotic Vascular Disease: 2011 Update: A Guidline From The American Heart Association and American College of Cardiology Foundation. Circulation 2011;124:2458-2473.
4. Morrow D, Scirica B, Fox K, et. al. Evaluation of a novel antiplatelet agent for secondary prevention in patients with a history of atherosclerotic disease: Design and rationale for the Thrombin-Receptor Antagonist in Secondary Prevention of Atherothrombotic Ischemic Events (TRA 2°P)-TIMI trial. American Heart Journal 2009;158:335-341.e.3.
5. Gerhard-Herman MD, Gornik HL, Shishehbor MH, et al. 2016 AHA/ACC guideline in the management of patients with lower extremity peripheral artery disease. J Am Coll Cardiol. 2017;69(11):e1465-1508.

Last Reviewed: 11/21/18, 1/20/21, 5/18/22, 7/21/23
Effective Date: 1/1/19, 7/15/22

Ztalmy® (ganaxolone)
Prior Authorization Guidelines

Affected Medication(s)

- Ztalmy oral suspension

FDA Approved Indication(s)

- Treatment of seizures associated with cyclin-dependent kinase-like 5 (CDKL5) deficiency disorder (CDD) in patients 2 years of age and older

Dosing

- 28kg or less: 6mg/kg orally three times daily through day 7; then 11 mg/kg three times daily through day 14; then 16 mg/kg three times daily through day 21; and 21 mg/kg three times daily thereafter
- Greater than 28kg: 150 mg/3 mL orally 3 times daily through day 7; then 300 mg/6 mL three times daily through day 14; then 450 mg/9 mL three times daily through day 21; and 600 mg/12 mL three times daily thereafter

Initial Authorization Criteria

1. Is the request for continuation of Ztalmy (ganaxolone) therapy?
    a. If yes, continue to Reauthorization
    b. If no, continue to #2
2. Is the request for use to treat an FDA approved indication? (Provide documentation of diagnosis)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the member currently 2 years of age or older?
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member currently have confirmed diagnosis of cyclin-dependent kinase-like 5 (CDKL5) deficiency disorder confirmed by genetic testing? (Provide supporting documentation)
    a. If yes, approve continue to #5
    b. If no, clinical review required
5. Has the member previously trialed at least 2 regimens containing two or more of the following used in combination? (Provide supporting documentation)
        i. Clobazam, valproate, topiramate, levetiracetam, vigabatrin
    a. If yes, continue to #6
    b. If no, clinical review required
6. Does the member continue to have uncontrolled seizures despite previous therapy? (Provide supporting documentation)
    a. If yes, continue to #7

Last Reviewed: 9/21/22, 9/15/23
Effective Date: 11/1/22

b. If no, clinical review required

7. Is the medication prescribed by, or in consultation with, a neurologist? (Provide supporting documentation)
    a. If yes, approve for 6 months
    b. If no, clinical review required

Reauthorization Criteria

1. Is the documented indication Food and Drug Administration (FDA) approved or supported by major compendia?
    a. If yes, continue to #2
    b. If no, clinical review required
2. Were updated chart notes (within 1 year) with documentation of significant clinical response to therapy received? (Significant clinical response is defined by a decrease in seizure frequency compared to pre-treatment baseline) (Provide supporting documentation)
    a. If yes, continue to #3
    b. If no, clinical review required
3. Is the treatment being prescribed by, or in consultation with, a neurologist?
    a. If yes, approve for 12 months reauthorization
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. ZTALMY® (ganaxolone) oral suspension, [package insert]. Radnor, PA: Marinus Pharmaceuticals, Inc.; 2022.
2. Drugs@FDA: FDA Approved Drug Products. 2019. accessdata.fda.gov. [online] Available at: https://www.accessdata.fda.gov/scripts/cder/daf/index.cfm [Accessed 3 Aug. 2022].
3. Knight, Elia M. Pestana, et al. "Safety and efficacy of ganaxolone in patients with CDKL5 deficiency disorder: results from the double-blind phase of a randomised, placebo-controlled, phase 3 trial." The Lancet Neurology 21.5 (2022): 417-427.
4. Leonard, Helen, et al. "CDKL5 deficiency disorder: clinical features, diagnosis, and management." The Lancet Neurology (2022).
5. Olson, Heather E., et al. "Current neurologic treatment and emerging therapies in CDKL5 deficiency disorder." Journal of neurodevelopmental disorders 13.1 (2021): 1-11.

Last Reviewed: 9/21/22, 9/15/23
Effective Date: 11/1/22

Zurzuvae™ (zuranolone)
Prior Authorization Guidelines

Affected Medication(s)

- Zurzuvae oral capsule

FDA Approved Indication(s)

- For treatment of postpartum depression (PPD) in adults

Dosing

- 50 mg orally once daily in the evening for 14 days
- The safety and effectiveness of Zurzuvae use beyond 14 days in a single treatment course have not been established.

Initial Authorization Criteria

1. Is the request for use to treat an FDA approved or major compendia supported indication? (Provide documentation of diagnosis)
    a. If yes, continue to #2
    b. If no, clinical review required
2. Is the member 18 years of age or older?
    a. If yes, continue to #3
    b. If no, clinical review required
3. Has the member experienced a major depressive episode with onset no earlier than the third trimester of pregnancy and no later than four weeks after delivery? (Provide supporting documentation)
    a. If yes, continue to #4
    b. If no, clinical review required
4. Does the member have a confirmed diagnosis of severe PPD documented by a standardized evidence-based depression rating scale that reliably measures depressive symptoms? (Provide test result for review)
    - Edinburgh postnatal Depression Scale (EPDS) ≥ 19
    - Patient Health Questionnaire-9 (PHQ-9) ≥ 20
    - Beck Depression Inventory (BDI-II) ≥ 29
    - Hamilton Rating Scale for Depression ≥ 19
    - Montgomery-Asberg Depression Rating Scale ≥ 35

    a. If yes, continue to #5
    b. If no, clinical review required
5. Is urgent symptom control necessary due to risk of harm to mother/baby? (Provide supporting documentation)
    a. If yes, continue to #7
    b. If no, continue to #6

Last Reviewed: 1/19/24
Effective Date: 3/1/24

6. Does the member have a previous trial with inadequate response to at least two antidepressants from at least two different classes for a minimum of 4 weeks OR a contraindication to all classes? (Examples: SSRI, SNRI, TCA, bupropion) (Provide supporting documentation)

    a. If yes, continue to #7
    b. If no, clinical review required

7. Is the treatment being prescribed by, or in consultation with, a mental health specialist?

    a. If yes, approve for 14 days
    b. If no, clinical review required

Note:

Medication prior authorization guidelines are developed by a team of health care professionals based on standards of practice at the time of approval. Guidelines are used as a basis for making coverage decisions and do not serve as medical advice. Coverage of medications is subject to plan provisions. Additional coverage restrictions not listed in the guidelines may apply.

References:

1. Zurzuvae (zuranolone) oral capsules [package insert]. Cambridge, MA: Sage Therapeutics, Inc.; 2023.
2. Treatment and Management of Mental Health Conditions During Pregnancy and Postpartum: ACOG Clinical Practice Guideline No. 5. Obstet Gynecol. 2023;141(6):1262-1288.
3. Screening and Diagnosis of Mental Health Conditions During Pregnancy and Postpartum: ACOG Clinical Practice Guideline No. 4. Obstet Gynecol. 2023;141(6):1232-1261.
4. Deligiannidis KM, Meltzer-Brody S, Maximos B, et al. Zuranolone for the Treatment of Postpartum Depression. Am J Psychiatry. 2023;180(9):668-675.
5. Deligiannidis KM, Meltzer-Brody S, Gunduz-Bruce H, et al. Effect of Zuranolone vs Placebo in Postpartum Depression: A Randomized Clinical Trial [published correction appears in JAMA Psychiatry. 2022 Jul 1;79(7):740] [published correction appears in JAMA Psychiatry. 2023 Feb 1;80(2):191]. JAMA Psychiatry. 2021;78(9):951-959.