PHP Medicare

Prior Authorization Criteria for

Medicare Part B Drugs

Last updated November 2, 2022

Contents

ACTEMRA (tocilizumab) J3262

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Actemra (tocilizumab) for **3 months (initial RA/SJIA)**, **4 months (initial PJIA)**, **6 months (initial GCA)** and **one year (reauthorization)**. **For diagnosis of chimeric antigen receptor (CAR) T cell-induced severe or life-threatening cytokine release syndrome (CRS), the plan may authorize coverage for one week**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Actemra (tocilizumab) is a recombinant humanized monoclonal antibody that inhibits interleukin-6 activity, decreasing T-cell activation. It is FDA approved for the treatment of: 1. Adults with moderately to severely active rheumatoid arthritis who have had an inadequate response to one or more DMARDs 2. Polyarticular juvenile idiopathic arthritis (PJIA) in patients 2 years of age and older 3. Systemic juvenile idiopathic arthritis (SJIA) in patients 2 years of age and older 4. Adults and pediatric patients 2 years of age and older with chimeric antigen receptor (CAR) T cell-induced severe or life-threatening cytokine release syndrome 5. Adults with giant cell arteritis. For RA, PJIA, and SJIA, Actemra can be used as monotherapy or concurrently with methotrexate or other non-biologic DMARDs.

Required Medical Information:

1. Rheumatoid arthritis (RA)
    a. Documentation of moderately to severely active RA, **AND**
    b. Previous trial of at least one DMARD (e.g. methotrexate, leflunomide, hydroxychloroquine, or sulfasalazine), **AND**
    c. Patient has been tested for latent TB prior to initiating Actemra therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    d. Prescribed by or in consultation with a rheumatologist, **AND**
    e. Patient is 18 years of age or older

2. Polyarticular juvenile idiopathic arthritis (PJIA)
    a. Diagnosis of PJIA, **AND**
    b. Previous trial of at least one DMARD (e.g. methotrexate, leflunomide, hydroxychloroquine, or sulfasalazine), **AND**
    c. Patient has been tested for latent TB prior to initiating Actemra therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    d. Prescribed by or in consultation with a rheumatologist, **AND**
    e. Patient is 2 years of age or older

3. Systemic juvenile idiopathic arthritis (SJIA)
    a. Diagnosis of SJIA, **AND**
    b. Previous trial of one other systemic agent for SJIA such as a corticosteroid (oral, IV), a conventional synthetic DMARD (e.g., MTX, leflunomide, sulfasalazine), or a biologic DMARD (e.g., Kineret, a TNF inhibitor such as Enbrel, Humira or Remicade, or Ilaris), or a 1-month trial of a nonsteroidal anti-inflammatory drug, **AND**
    c. Patient has been tested for latent TB prior to initiating Actemra therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    d. Prescribed by or in consultation with a rheumatologist, **AND**
    e. Patient is 2 years of age or older
4. Cytokine release syndrome (CRS)
    a. Diagnosis of chimeric antigen receptor (CAR) T cell-induced severe or life-threatening cytokine release syndrome (CRS), **AND**
    b. The patient is 2 years of age or older, **AND**
    c. Patient has been tested for latent TB prior to initiating Actemra therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval)
5. Giant cell arteritis (GCA)
    a. Diagnosis of giant cell arteritis, **AND**
    b. Therapy is prescribed by or in consultation with a rheumatologist, **AND**
    c. The patient is 18 years of age or older, **AND**
    d. Patient has been tested for latent TB prior to initiating Actemra therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval)

Reauthorization:

1. Patient has a diagnosis of giant cell arteritis (GCA), **OR**
2. Patient has a diagnosis of moderate to severe rheumatoid arthritis (RA), polyarticular juvenile idiopathic arthritis (PJIA), or systemic juvenile idiopathic arthritis (SJIA) AND physician attestation that patient continues to benefit from the medication

Exclusion Criteria:

1. Avoid use in combination with biological DMARDs, such as tumor necrosis factor (TNF) antagonists, interleukin 1 receptor (IL-1R) antagonists, anti-CD20 monoclonal antibodies, and selective costimulation modulators, because of the possibility of increased immunosuppression and increased risk of infection.
2. Actemra is contraindicated in patients with known hypersensitivity to Actemra
3. Avoid live vaccines with Actemra
4. Do not administer ACTEMRA during an active infection, including localized infections
5. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Actemra prescribing information

Version History

Last Reviewed Date	Updates / Revisions
3/1/16	None

12/18/17	Addition to FDA indications
2/12/18	Update to required information and reauthorization criteria; Removal of clinical note
8/20/18	Update to required information
3/5/19	Update to initial and reauthorization timeframe
2/10/20	None
2/17/21	None
3/23/22	Addition to FDA indications; Addition to exclusion criteria; Update to initial approval timeframe; Addition to required medical information; Update to initial criteria for SJIA; Update to reauthorization criteria

ADAKVEO (crizanlizumab-tmca) J0791 – *effective 9/1/2022*

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage for 12 months (initial and reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
ADAKVEO is a selectin blocker indicated to reduce the frequency of vasoocclusive crises in adults and pediatric patients aged 16 years and older with sickle cell disease.

Required Medical Information:

1. Patient has diagnosis of sickle cell disease, **AND**
2. Therapy is prescribed by or given in consultation with a hematologist, **AND**
3. Patient is at least 16 years of age or older
    a. For patients 18 years of age or older, they must also meet ONE of the following:
        i. The patient had at least 2 sickle cell crises in the past year (a sickle cell crises is defined as a visit to an emergency room/medical facility for sickle cell disease-related pain which was treated with a parenterally administered narcotic or parenterally administered ketorolac, the occurrence of acute chest syndrome, priapism, or splenic sequestration)
        ii. The patient is having sickle-cell associated symptoms (e.g., pain or anemia) which are interfering with activities of daily living
        iii. The patient has a history of or has recurrent acute chest syndrome (ACS)

Reauthorization:

1. Patient has a diagnosis of sickle cell disease AND has maintained or experienced a reduction in acute complications of sickle-cell disease (SCD) (e.g., number of sickle cell crises, hospitalizations, acute chest syndrome [ACS]).

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Adakveo prescribing information

Version History

Last Reviewed Date	Updates / Revisions
3/23/22	Addition to part B criteria with effective date 9/1/2022

ADUHELM (aducanumab-avwa) J0172

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage for 6 months (initial) and 12 Months (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
ADUHELM (aducanumab-avwa) is an amyloid beta-directed antibody indicated for the treatment of Alzheimer's disease. Treatment with ADUHELM should be initiated in patients with mild cognitive impairment or mild dementia stage of disease, the population in which treatment was initiated in clinical trials. • There are no safety or effectiveness data on initiating treatment at earlier or later stages of the disease than were studied. • This indication is approved under accelerated approval based on reduction in amyloid beta plaques observed in patients treated with ADUHELM. Continued approval for this indication may be contingent upon verification of clinical benefit in confirmatory trial(s).

Required Medical Information:

1. Alzheimer's disease
    a. The patient has mild cognitive impairment or mild dementia stage of disease, AND
    b. Obtain a recent (within one year) brain MRI prior to initiating treatment, AND
    c. Therapy is prescribed by or in consultation with a neurologist, geriatrician, or psychiatrist, AND
    d. Patient must be enrolled in an FDA or National Institutes of Health (NIH) approved trial for Aduhelm.

Reauthorization:

1. Patient has a diagnosis of mild cognitive impairment due to Alzheimer's disease or mild dementia stage of Alzheimer's disease and meet ALL of the following criteria:
    a. The patient's cognitive decline has slowed or stopped
    b. The patient is not experiencing any amyloid-related imaging abnormalities (ARIA)
    c. Patient must be enrolled in an FDA or National Institutes of Health (NIH) approved trial for Aduhelm.

Exclusion Criteria:

Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Aduhelm prescribing information

Version History

Last Reviewed Date	Updates / Revisions
8/10/21	Addition to part B criteria with effective date 3/1/2022

3/23/22	Update to Jcode; Update to approval timeframe; Addition to initial criteria; Update to reauthorization criteria
11/2/22	Update to initial and reauthorization criteria

ALDURAZYME (laronidase) J1931

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Aldurazyme (laronidase) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Aldurazyme (laronidase) is hydrolytic lysosomal glycosaminoglycan (GAG)-specific enzyme indicated for patients with Hurler and Hurler-Scheie forms of Mucopolysaccharidosis I (MPS I) and for patients with the Scheie form who have moderate to severe symptoms.

Required Medical Information:
1. Member must have a diagnosis of Hurler and Hurler-Scheie forms of Mucopolysaccharidosis I (MPS I), **OR** Member must have a diagnosis of Scheie form of Mucopolysaccharidosis I (MPS I) who have moderate to severe symptoms, **AND**
2. Therapy is prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders, **AND**
3. Patient has a laboratory test demonstrating deficient alphaL-iduronidase activity in leukocytes, fibroblasts, plasma, or serum OR has a molecular genetic test demonstrating alpha-L-iduronidase gene mutation

Reauthorization:
1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:
1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. The risks and benefits of treating mildly affected patients with the Scheie form have not been established.
3. ALDURAZYME has not been evaluated for effects on the central nervous system manifestations of the disorder.

References:
Aldurazyme prescribing information

Version History

Last Reviewed Date	Updates / Revisions
3/1/16	None

3/3/17	None
2/12/18	None
3/5/19	None
2/10/20	None
2/17/21	Addition of reauthorization criteria
3/23/22	Addition to exclusion criteria; Addition to initial criteria

BENLYSTA (belimumab) J0490

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Benlysta (belimumab) for **4 months (initial SLE), 6 months (initial Lupus Nephritis), and one year (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Benlysta (belimumab) is a B-lymphocyte stimulator (BLyS)-specific inhibitor indicated for the treatment of: • Patients aged 5 years and older with active, autoantibody-positive, systemic lupus erythematosus who are receiving standard therapy (i.e. corticosteroids, antimalarials, immunosuppressives and NSAIDs). • Adult patients with active lupus nephritis who are receiving standard therapy (i.e. corticosteroids, antimalarials, immunosuppressives and NSAIDs).

Required Medical Information:
1. Systemic lupus erythematosus (SLE)
    a. Documentation of active, autoantibody-positive, SLE diagnosis, **AND**
    b. Documentation that the patient is receiving standard therapy for SLE (e.g. corticosteroids, antimalarials, NSAIDS or immunosuppressives), **AND**
    c. Patient does NOT have severe active central nervous system lupus, **AND**
    d. Patient is NOT on concurrent therapy with other biologic products (i.e. Rituximab, Orencia, Enbrel, Remicade, or Humira), **AND**
    e. Patient must be 5 years old or older, **AND**
    f. Therapy is prescribed by or in consultation with a rheumatologist, clinical immunologist, nephrologist, neurologist, or dermatologist
2. Lupus Nephritis
    a. Documentation of active lupus nephritis diagnosis, **AND**
    b. Documentation that the patient is receiving standard therapy for SLE (e.g. corticosteroids, antimalarials, NSAIDS or immunosuppressives), **AND**
    c. Patient does NOT have severe active central nervous system lupus, **AND**
    d. Patient is NOT on concurrent therapy with other biologic products (i.e. Rituximab, Orencia, Enbrel, Remicade, or Humira), **AND**
    e. Patient must be 18 years old or older, **AND**
    f. Therapy is prescribed by or in consultation with a nephrologist or rheumatologist

Reauthorization:
1. Diagnosis of autoantibody positive systemic lupus erythematosus (SLE) OR lupus nephritis **AND** physician attestation of improvement

Exclusion Criteria:

1. The efficacy of BENLYSTA has not been evaluated in patients with severe active central nervous system lupus. BENLYSTA has not been studied in combination with other biologics. Use of BENLYSTA is not recommended in these situations.
2. Live vaccines should not be given for 30 days before or concurrently with Benlysta, as clinical safety has not been established.
3. Previous anaphylaxis to Benlysta
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Benlysta prescribing information

Version History

Last Reviewed Date	Updates / Revisions
3/1/16	None
3/3/17	Removal of clinical note – not relevant to approval/denial
2/12/18	Update to required information; Addition of reauthorization criteria; Update to approval timeframe
1/1/19	Updates to authorization period, required information, and reauthorization criteria to align with part D criteria
3/5/19	None
2/10/20	Update to approval timeframe (initial and reauthorization), Update to indication and required information
2/17/21	Addition of new indication; Update to exclusion criteria
3/23/22	Addition to exclusion criteria; Update to initial approval time frame; Addition to initial criteria

BERINERT (C1 esterase inhibitor, human) J0597

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage for 3 months (initial) and 12 months (Reauthorization).** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
BERINERT (C1 esterase inhibitor, human) is a plasma-derived C1 Esterase Inhibitor (Human) indicated for the treatment of acute abdominal, facial, or laryngeal hereditary angioedema (HAE) attacks in adult and pediatric patients.

Required Medical Information:

1. **Hereditary Angioedema (HAE)**
    a. The patient has a diagnosis of hereditary angioedema (HAE) as confirmed by complement testing, AND
    b. The medication is prescribed by or given in consultation with a hematologist, immunologist, or allergist, AND
    c. The medication is being used for acute attacks of hereditary angioedema

Reauthorization:

1. Use initial criteria as Berinert is approved for acute attacks only. **AND**
2. Patient must have responded to therapy as determined by the prescribing physician.

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. Do not use in patients with a history of life-threatening immediate hypersensitivity reactions, including anaphylaxis, to C1 esterase inhibitor preparations.
3. The safety and efficacy of BERINERT for prophylactic therapy have not been established.

References:

Berinert package insert

Version History:

Last Reviewed Date	Updates / Revisions
11/10/2021	Addition to part B criteria with effective date 1/1/2022
3/23/22	Addition to exclusion criteria

BONIVA INJECTION (ibandronate) J1740

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Boniva (ibandronate) for **one year.** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Boniva (ibandronate) is a bisphosphonate that is FDA approved for the treatment of osteoporosis in postmenopausal women.

Required Medical Information:

1. **Treatment of osteoporosis in postmenopausal women**
    a. Postmenopausal woman with T-score </= -2.5 or history of osteoporotic fracture, **OR** postmenopausal woman with T-score -1.0 to -2.5 plus at least one of the following risk factors for fracture:
        i. BMI<21
        ii. Current smoker
        iii. History of previous osteoporotic fracture
        iv. History of hip fracture in a parent
        v. Oral glucocorticoid ≥5 mg/d of prednisone for >3 months (ever)
        vi. Alcohol intake (3 or more drinks/day)
        vii. Rheumatoid arthritis
        viii. Secondary causes of osteoporosis: type 1 diabetes, osteogenesis imperfecta in adults, untreated long-standing hyperthyroidism, hypogonadism or premature menopause (<40 years), chronic malnutrition or malabsorption, or chronic liver disease, **AND**
    b. Documented trial and failure of an oral bisphosphonate for one of the following reasons:
        i. GI intolerance
        ii. Issues related to absorption (or inability to take anything by mouth), compliance, or dosing posture (inability to sit or stand for at least 60 minutes)
        iii. Decrease in bone mineral density (BMD) while on therapy

Reauthorization:

1. When an updated DEXA scan since last authorization is available, there has been no clinically significant change or there has been a significant increase in T-score compared to previous DEXA, OR
2. Previous DEXA was less than 2 years ago and has not been repeated
3. If previous DEXA was more than 2 years ago, request updated DEXA.

Exclusion Criteria:

1. Use of Boniva is contraindicated in hypocalcemia. Hypocalcemia must be corrected prior to initiation of therapy.
2. Hypersensitivity to BONIVA Injection
3. Do not administer BONIVA injection to patients with severe renal impairment (creatinine clearance less than 30 mL/min)
4. Concurrent use with other medications for Osteoporosis
5. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Boniva prescribing information
National Osteoporosis Foundation Guidelines for Prevention and Treatment of Osteoporosis

Version History

Last Reviewed Date	Updates / Revisions
3/1/16	Update to exclusion criteria
3/3/17	Update to risk factors for osteoporosis
2/12/18	Update to required information
3/5/19	None
2/10/20	None
2/17/21	None
3/23/22	Addition to exclusion criteria; Update to initial criteria; Addition to references

BOTULINUM TOXINS J0585, J0586, J0587, J0588

BOTOX (OnabotulinumtoxinA) J0585, DYSPORT (AbobotulinumtoxinA) J0586, MYOBLOC (RimabotulinumtoxinB) J0587, XEOMIN (IncobotulinumtoxinA) J0588

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for BOTOX (OnabotulinumtoxinA), DYSPORT (AbobotulinumtoxinA), MYOBLOC (RimabotulinumtoxinB), or XEOMIN (IncobotulinumtoxinA) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication (s):

BOTOX (OnabotulinumtoxinA) is an acetylcholine release inhibitor and a neuromuscular blocking agent indicated for:

1. Treatment of strabismus in patients ≥ 12 years of age
2. Treatment of blepharospasm associated with dystonia in patients ≥ 12 years of age
3. Treatment of cervical dystonia in adult patients, to reduce the severity of abnormal head position and neck pain
4. Treatment of severe axillary hyperhidrosis that is inadequately managed by topical agents in adult patients
5. Prophylaxis of headaches in adult patients with chronic migraine (≥15 days per month with headache lasting 4 hours a day or longer)
6. Treatment of urinary incontinence due to detrusor overactivity associated with a neurologic condition [e.g., spinal cord injury (SCI), multiple sclerosis (MS)] in adults who have an inadequate response to or are intolerant of an anticholinergic medication
7. Treatment of neurogenic detrusor overactivity (NDO) in pediatric patients 5 years of age and older who have an inadequate response to or are intolerant of anticholinergic medication.
8. Treatment of overactive bladder with symptoms of urge urinary incontinence, urgency, and frequency, in adults who have an inadequate response to or are intolerant of an anticholinergic medication
9. Treatment of spasticity in patients 2 years of age and older
    a. *Upper Limb*: Biceps Brachii, Brachioradialis, Brachialis, Pronator Teres, Pronator Quadratus, Flexor Carpi Radialis, Flexor Carpi Ulnaris, Flexor Digitorum Profundus, Flexor Digitorum Sublimis, Lumbricals/Interossei, Adductor Pollicis, Flexor Pollicis Longus, Flexor pollicis brevis/ Opponens pollicis.
    b. *Lower Limb*: gastrocnemius, soleus, tibialis posterior, flexor hallucis longus and flexor digitorum longus

DYSPORT (AbobotulinumtoxinA) is an acetylcholine release inhibitor and a neuromuscular blocking agent indicated for:

1. Treatment of adults with cervical dystonia
2. Treatment of spasticity in patients 2 years of age and older.

MYOBLOC (RimabotulinumtoxinB) is indicated for

1. Treatment of cervical dystonia to reduce the severity of abnormal head position and neck pain associated with cervical dystonia in adults
2. Treatment of chronic sialorrhea in adults

XEOMIN (IncobotulinumtoxinA) is an acetylcholine release inhibitor and neuromuscular blocking agent indicated for:

1. Treatment of adults with cervical dystonia in both botulinum toxin-naïve and previously treated patients.
2. Treatment of blepharospasm in adults
3. Treatment of upper limb spasticity in adults.
4. The treatment of Upper limb spasticity in pediatric patients 2 to 17 years of age, excluding spasticity caused by cerebral palsy
5. Treatment of chronic sialorrhea in patients 2 years of age and older

Required Medical Information:

1. **Blepharospasm (Botox, Xeomin)**
    a. Documented diagnosis of blepharospasm associated with dystonia, **AND**
    b. Patient must be ≥12 years old or older (Botox) or ≥18 years (Xeomin)

2. **Cervical dystonia (spasmodic torticollis) (Botox, Dysport, Myobloc, Xeomin)**
    a. Documented diagnosis of cervical dystonia associated with neck pain and/or abnormal head position, **AND**
    b. Patient must be ≥18 years of age (Botox, Dysport, Myobloc, Xeomin)

3. **Upper/Lower Limb Spasticity (Botox, Dysport)**
    a. Documented diagnosis of spasticity in one of the following muscles
        i. Upper Limb: Biceps Brachii, Brachioradialis, Brachialis, Pronator Teres, Pronator Quadratus, Flexor Carpi Radialis, Flexor Carpi Ulnaris, Flexor Digitorum Profundus, Flexor Digitorum Sublimis, Lumbricals/Interossei, Adductor Pollicis, Flexor Pollicis Longus, Flexor pollicis brevis/ Opponens pollicis.
        ii. Lower Limb: gastrocnemius, soleus, tibialis posterior, flexor hallucis longus and flexor digitorum longus, **AND**
    b. Patient must be 2 years of age or older

4. **Upper limb spasticity (Xeomin)**
    a. Documented diagnosis of Upper limb spasticity **AND**
    b. Patient must be 18 years old or older **OR**
    c. Patient is 2 to 17 years of age AND spasticity is NOT caused by cerebral palsy

5. **Prophylaxis of headaches in adult patients with chronic migraine (≥15 days per month with headache lasting 4 hours a day or longer) (Botox)**
    a. Documented diagnosis of migraine headaches, along with history of recurrent headaches, **AND**
    b. Migraine headache prevention-prescribed by, or after consultation with, a neurologist or HA specialist, **AND**
    c. Patient has tried at least TWO standard prophylactic pharmacologic therapies, each from a different pharmacologic class (e.g., beta-blocker, anticonvulsant, tricyclic antidepressant) and patient has had inadequate efficacy, **AND**
    d. Patient must be 18 years old or older

6. **Neurogenic detrusor overactivity (NDO) (Botox)**
    a. The patient is 5 years of age or older **AND**
    b. The patient had an inadequate response to or is intolerant of anticholinergic medications (e.g. oxybutynin IR or ER, tolterodine, tolterodine ER, Toviaz, trospium, trospium XR, solifenacin, Enablex, Vesicare, Oxytrol) OR an oral beta-3 adrenergic agonist drug for overactive bladder (e.g. Myrbetriq)

7. **Urinary incontinence due to detrusor overactivity associated with a neurologic condition [e.g., spinal cord injury (SCI), multiple sclerosis (MS)] (Botox)**
    a. Documented diagnosis of detrusor overactivity (overactive bladder) associated with a neurologic condition (e.g. spinal cord injury, multiple sclerosis), **AND**
    b. Documented inadequate response or intolerance to an anticholinergic drug for overactive bladder (e.g. oxybutynin IR or ER, tolterodine, tolterodine ER, Toviaz, trospium, trospium XR, solifenacin, Enablex, Vesicare, Oxytrol) OR an oral beta-3 adrenergic agonist drug for overactive bladder (e.g. Myrbetriq), **AND**
    c. Patient must be 18 years old or older

8. **Overactive bladder with symptoms of urge urinary incontinence, urgency, and frequency (Botox)**
    a. Documented diagnosis of overactive bladder symptoms, **AND**
    b. Documented inadequate response or intolerance to an anticholinergic drug for overactive bladder (e.g. oxybutynin IR or ER, tolterodine, tolterodine ER, Toviaz, trospium, trospium XR, solifenacin, Enablex, Vesicare, Oxytrol) OR oral beta-3 adrenergic agonist drug for overactive bladder (e.g. Myrbetriq), **AND**
    c. Patient must be 18 years or older

9. **Primary axillary hyperhidrosis (Botox)**
    a. Documented diagnosis of primary axillary hyperhidrosis, **AND**
    b. Documented inadequate response to topical agents, **AND**
    c. Patient must be 18 years old or older

10. **Strabismus (Botox)**
    a. Documented diagnosis of strabismus, **AND**
    b. Patient must be 12 years old or older

11. **Chronic sialorrhea (Myobloc, Xeomin)**
    a. Documented diagnosis of chronic sialorrhea, **AND**
    b. Patient must be $\geq$ 2 years old or older (Xeomin) or $\geq$18 years (Myobloc)

Reauthorization:

1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:

1. BOTOX is contraindicated in patients with hypersensitivity to any botulinum toxin preparation or to any of the components in the formulation and in patients with an infection at the proposed site(s) of injection.
2. DYSPORT is contraindicated in patients who have allergy to cow's milk protein, and patients who have had hypersensitivity to any botulinum toxin product or to any of its components, and in patients with an infection at the proposed site (s) of injection.
3. MYOBLOC is contraindicated in patients with known hypersensitivity to any botulinum toxin preparation or to any of its components, and in patients with infection at proposed site(s) of injection.
4. XEOMIN is contraindicated in patients with hypersensitivity to active ingredient botulinum neurotoxin type A or any of its components and in patients with an infection at the proposed site(s) of injection.

5. Botulinum toxins when used for cosmetic purposes are not covered under Medicare.
6. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

- Botox, Dysport, Myobloc, and Xeomin prescribing information
- Overactive bladder Guidelines: https://www.auanet.org/guidelines/overactive-bladder-(oab)-guideline
- Lower urinary tract dysfunction in patients with spinal cord injury guidelines: https://onlinelibrary.wiley.com/doi/full/10.1111/iju.14186

Version History

Last Reviewed Date	Updates / Revisions
3/1/16	Addition to FDA approved indications for Botox and Xeomin; Update to exclusion criteria
3/3/17	Update wording for FDA indications; Update to required information for indication of migraine headaches; Update to required information for indication of primary axillary hyperhidrosis
2/12/18	Update to FDA indications per product labeling
8/20/18	Update to FDA approved indications
3/5/19	Update to required information (removal of diagnoses)
12/10/19	Addition to FDA approved indication for Botox
2/10/20	Update to FDA approved indications and required information
2/17/21	Update to FDA approved indications and required information; Addition of reauthorization criteria
11/10/21	Update to FDA approved indications
3/23/22	Addition of new FDA approved indication and initial criteria. Update to required information

CEREZYME (imiglucerase for injection) J1786

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Cerezyme (imiglucerase) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Cerezyme (imiglucerase for injection) is a hydrolytic lysosomal glucocerebrosidase-specific enzyme indicated for treatment of adults and pediatric patients 2 years of age and older with Type 1 Gaucher disease that results in one or more of the following conditions: anemia, thrombocytopenia, bone disease, hepatomegaly or splenomegaly.

Required Medical Information:

1. Documented diagnosis of Type 1 Gaucher disease, AND one of the following:
    a. Has symptomatic manifestations of skeletal disease as confirmed by radiological assay, including: joint deterioration, pathological fracture, avascular necrosis, definite osteopenia, marrow infiltration, **OR**
    b. Presents with one or more of the following: anemia, thrombocytopenia, hepatomegaly, splenomegaly, AND
2. Prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders, **AND**
3. Demonstration of deficient beta-glucocerebrosidase activity in leukoctyes or fibroblasts OR molecular genetic testing documenting glucocerebrosidase gene mutation

Reauthorization:

1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Cerezyme prescribing information

Version History

Last Reviewed Date	Updates / Revisions
3/1/16	None
3/3/17	Update to required information
2/12/18	None

3/5/19	None
2/10/20	None
2/17/21	Update to required information to align with part D criteria; Addition of reauthorization criteria
3/23/22	None

CINQAIR (reslizumab) J2786

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met**,** the plan may authorize coverage for Cinqair (reslizumab) for **12 months (initial)** AND **(reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
CINQAIR (reslizumab) is an interleukin-5 antagonist monoclonal antibody (IgG4 kappa) indicated for add-on maintenance treatment of patients with severe asthma aged 18 years and older, and with an eosinophilic phenotype

Required Medical Information:
1. Diagnosis of severe eosinophilic asthma and meet ALL of the following:
    a. The patient is 18 years of age or older, **AND**
    b. Patient had prior therapy with a medium, high-dose, or maximally tolerated dose of an inhaled corticosteroid AND at least one other maintenance medication (e.g., long-acting inhaled beta2-agonist, long-acting muscarinic antagonist, leukotriene receptor antagonist, theophylline, oral corticosteroid), **AND**
    c. Patient has a blood eosinophil level greater than or equal to 150 cells/mcL within the past 12 months, **AND**
    d. Cinqair will be used as add-on maintenance treatment, **AND**
    e. Cinqair will NOT be used concurrently with Xolair (omalizumab), Dupixent (dupilumab) or other anti-interleukin-5 (IL-5) biologics (e.g., Nucala, Fasenra) when these are used for the treatment of asthma, **AND**
    f. Cinqair is prescribed by or given in consultation with a physician specializing in allergy or pulmonary medicine, **AND**
    g. The patient meets ONE of the following Criteria:
        i. The patient experienced at least ONE asthma exacerbation requiring systemic corticosteroid burst lasting 3 or more days within the past 12 months or at least ONE serious exacerbation requiring hospitalization or ER visit within the past 12 months **OR**
        ii. The patient has poor symptom control despite current therapy as evidenced by at least THREE of the following within the past 4 weeks: Daytime asthma symptoms more than twice per week, any night waking due to asthma, SABA reliever (ie: rescue inhaler like Albuterol) for symptoms more than twice per week, any activity limitation due to asthma

Reauthorization:
1. Patient has diagnosis of severe eosinophilic asthma and has shown a clinical response as evidenced by ONE of the following:
    a. Reduction in asthma exacerbation from baseline
    b. Decreased utilization of rescue medications

c. Reduction in severity or frequency of asthma-related symptoms (e.g., wheezing, shortness of breath, coughing, etc.)
d. Increase in percent predicted FEV1 from pretreatment baseline

Exclusion Criteria:

1. Cinqair is NOT indicated for treatment of other eosinophilic conditions or relief of acute bronchospasm or status asthmaticus.
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Cinqair prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/19/19	Addition to part B criteria with effective date 1/1/20
2/10/20	Update to required information, reauthorization criteria, and authorization period
2/17/21	Update to required information and reauthorization criteria
3/23/22	Update to required information

CINRYZE (C1 esterase inhibitor, human) J0598

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Cinryze (C1 esterase inhibitor) for **one year (Initial and Reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Cinryze is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years of age and older) with Hereditary Angioedema (HAE).

Required Medical Information:

1. **Hereditary Angioedema (HAE)**
    a. Documented diagnosis of HAE, **AND**
    b. Prescribed by or in consultation with an allergist/immunologist or a physician that specializes in the treatment of HAE or related disorders, **AND**
    c. the patient has Hereditary Angioedema (HAE) type I or type II confirmed by low levels of functional C1-INH protein (less than 50% of normal) at baseline and lower than normal serum C4 levels at baseline.

Reauthorization:

- Patient is currently taking Cinryze for prophylaxis and meets all of the following:
    - patient has a diagnosis of HAE type I or II, and
    - according to the prescriber, the patient has had a favorable clinical response since initiating Cinryze as prophylactic therapy compared with baseline.

Exclusion Criteria:

1. Cinryze is contraindicated in patients who develop or have known hypersensitivity to human C1 esterase inhibitors**,** and/ or any component of the product.
2. Cinryze is NOT FDA indicated for treatment of acute hereditary angioedema (HAE) attack.
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Cinryze prescribing information

Version History

Last Reviewed Date	Updates / Revisions
3/1/16	None

3/3/17	Update to required information
2/12/18	None
1/1/19	Update to authorization period and required information to align with part D criteria
3/5/19	None
2/10/20	Update to authorization period
2/17/21	None
3/23/22	Addition to required information and addition of reauthorization criteria

DIABETES TESTING SUPPLIES: Non-Preferred Blood Glucose Meters and Test Strips

Covered Uses:

The health plan covers LifeScan and Abbott glucose meters and strips through network pharmacies, including:

LifeScan	Abbott
- OneTouch Verio Flex - OneTouch Verio Reflect	- FreeStyle Freedom Lite - FreeStyle Lite

The health plan may make an exception to cover non-LifeScan or non-Abbott products through pharmacies if the criteria listed below are met.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for non-preferred meter and test strips for **Lifetime approval for visual impairment/voice meter and 1 year for all other meters and test strips**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

Required Medical Information:

1. Member has visual impairment and requires a voice meter, and there is no voice meter in the preferred suite of products, **OR**
2. Member requires a meter that communicates with an insulin pump, and there is no such meter in the preferred suite of products, **OR**
3. Requests for other reasons will be considered on a case-by-case basis.

Last Reviewed Date	Updates / Revisions
5/15/20	Addition to part B criteria
2/17/21	None
9/9/21	Update to available products
11/10/21	Update to approval time frame
3/23/22	None

DUOPA (carbidopa/levodopa enteral suspension) J7340

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, Essence may authorize coverage for Duopa (carbidopa/levodopa) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Duopa (carbidopa/levodopa) is a combination of carbidopa (an aromatic amino acid decarboxylation inhibitor) and levodopa (an aromatic amino acid) indicated for the treatment of motor fluctuations in patients with advanced Parkinson's disease (PD).

Required Medical Information:
1. Treatment of motor fluctuations in patients with advanced Parkinson's disease (PD), **AND**
2. The prescriber is a neurologist, **AND**
3. Documented idiopathic PD based on the presence of bradykinesia and at least one other cardinal PD feature (i.e. tremor, rigidity, postural instability), **AND**
4. Documented response to L-dopa (i.e. with clearly defined "On" periods), **AND**
5. Documented persistent motor complications with disabling "Off" periods for a minimum of 3 hours/day, despite adequate medical therapy with levodopa-carbidopa, and at least one other class of anti-PD therapy (i.e. COMT inhibitor or MAO-B inhibitor)

Reauthorization:
1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:
1. Atypical Parkinson's syndrome ("Parkinson's Plus" syndrome) or secondary Parkinson's
2. Non-levodopa responsive PD
3. Contraindication to percutaneous endoscopic gastro-jejunal (PEG-J) tube placement or long-term use of a PEG-J
4. Contraindicated in patients taking nonselective monoamine oxidase (MAO) inhibitors
5. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

Version History

Last Reviewed Date	Updates / Revisions
1/1/18	Addition to part B criteria with effective date 1/1/18

8/20/18	Update to exclusion criteria
3/5/19	None
2/10/20	None
2/17/21	Addition of reauthorization criteria
3/23/22	None

EMPAVELI (Pegcetacoplan) J3490

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, **the plan may authorize coverage for initial request – 4 months, Reauthorization – 12 months.** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
EMPAVELI (Pegcetacoplan) is a complement inhibitor indicated for the treatment of adult patients with paroxysmal nocturnal hemoglobinuria (PNH). EMPAVELI is available only through a restricted program under a Risk Evaluation and Mitigation Strategy (REMS). Under the EMPAVELI REMS, prescribers must enroll in the program.

Required Medical Information:
1. A diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) and meet **ALL** of the following criteria:
    a. The patient is 18 years of age or older
    b. Therapy is prescribed by or given in consultation with a hematologist
    c. The patient has documented confirmation of PNH by flow cytometry demonstrating **ALL** of the following:
        i. At least 2 different GPI-protein deficiencies (e.g., CD55, CD59) on at least 2 cell lineages (e.g., erythrocytes, granulocytes)
        ii. PNH granulocyte clone size of 10% or greater
    d. The patient is not using concurrent C5 complement inhibitor therapy (e.g., Soliris, Ultomiris)
    e. The patient has tried and failed Soliris or Ultomiris as evidenced by hemoglobin levels <10.5 g/dL directly following at least 3 months of stable dosing

Reauthorization:

1. A diagnosis of paroxysmal nocturnal hemoglobinuria (PNH) and meet the following criterion:
    a. The patient has had clinical benefit compared to baseline during treatment with Soliris or Ultomiris (e.g., reduction in number of blood transfusions, improvement/stabilization of lactate dehydrogenase (LDH) and hemoglobin levels).

Exclusion Criteria:
1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. Patients who are not currently vaccinated against certain encapsulated bacteria unless the risks of delaying EMPAVELI treatment outweigh the risks of developing a serious bacterial infection with an encapsulated organism.
3. Patients with unresolved serious infection caused by encapsulated bacteria.

References:
Empaveli package insert

Version History

Last Reviewed Date	Updates / Revisions
11/10/2021	Addition to part B criteria with effective date 1/1/2022
3/23/22	None

ENTYVIO (vedolizumab) J3380

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Entyvio (vedolizumab) for **14 weeks (initial) and 1 year (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
ENTYVIO is an integrin receptor antagonist indicated in adults for the treatment of: • moderately to severely active ulcerative colitis. • moderately to severely active Crohn's disease.

Required Medical Information:
1. Documented diagnosis of moderately to severely active Crohn's disease **OR** Ulcerative Colitis, **AND**
2. Prescribed by or given in consultation with a gastroenterologist, **AND**
3. Patient must be 18 years of age or older, **AND**
4. Documented inadequate response to at least one conventional therapy drug (e.g. azathioprine, 6-mercaptopurine, sulfasalazine, methotrexate, oral mesalamine [Asacol, Apriso, Delzicol, Lialda, Pentasa], Colazal, balsalazide, Dipentum), **OR**
5. Documented inadequate response, loss of response or inability to tolerate a tumor necrosis factor (TNF) blocker (e.g., Remicade®, Humira®), **OR**
6. Steroid-dependent (e.g. inability to taper or discontinue), or documented inadequate response or intolerance to corticosteroids

Reauthorization:
1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:
1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. Concurrent Use with Other Biologics or with Targeted Synthetic Disease-Modifying Antirheumatic Drugs (DMARDs) used for an Inflammatory Condition.
3. Contraindicated in patients with known serious or severe hypersensitivity reaction to Entyvio or any of its excipients.
4. Not recommended in patients with confirmed diagnosis of progressive multifocal leukoencephalopathy (PML).
5. Not recommended in patients with active, severe infections until the infections are controlled.

References:

Entyvio prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	None
5/24/17	Update to exclusion criteria
6/7/18	None
6/26/19	None
4/29/20	Update to authorization time frame
2/17/21	Addition of reauthorization criteria
5/12/21	Update to authorization time frame, exclusion criteria, and required information
5/25/22	Update to FDA indication; Update to reauthorization approval time frame; Addition to exclusion criteria

EPOPROSTENOL (Flolan, Veletri) J1325

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary. ***Coverage of brand name Flolan and Veletri will be considered for approval in patients who have documented inadequate response to OR are not able to tolerate generic epoprostenol.***

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Epoprostenol for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Epoprostenol (prostacyclin) is a peripheral vasodilator FDA approved for the treatment of pulmonary arterial hypertension (PAH) (World Health Organization [WHO] group 1) to improve exercise capacity. Studies establishing effectiveness included predominately patients with NYHA functional class 3 to 4 symptoms and etiologies of idiopathic or heritable PAH or PAH associated with connective tissue diseases

Required Medical Information:

1. Documented diagnosis of one of the following:
    - Documented diagnosis of primary pulmonary hypertension functional NYHA Class III or IV, **OR**
    - Documented diagnosis of pulmonary hypertension due to scleroderma NYHA Class III or IV, **OR**
    - Documented diagnosis of pulmonary hypertension secondary to one of the following conditions: connective tissue disease, thromboembolic disease of the pulmonary arteries, HIV infection, cirrhosis, diet drugs, congenital left to right shunts; **AND**
2. Documented confirmatory pulmonary arterial hypertension diagnosis based on right heart catheterization based on the following parameters:
    - Mean pulmonary arterial pressure (PAP) of > 20 mmHg
    - Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    - Pulmonary vascular resistance (PVR) $\geq$ 3 Wood units; **AND**
3. Pulmonary hypertension has progressed despite maximal medical and/or surgical treatment, **AND**
4. Mean pulmonary artery pressure is greater than 25mmHg at rest or greater than 30mmHg with exertion, **AND**
5. Significant symptoms from pulmonary hypertension are present (i.e. severe dyspnea on exertion, fatigue, angina, or syncope), **AND**
6. Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out, **AND**
7. Patient must be 18 years old or older, **AND**
8. Therapy is prescribed by or in consultation with a cardiologist or pulmonologist

Reauthorization:

1. Diagnosis of pulmonary arterial hypertension (PAH) (WHO Group 1) and shown improvement from baseline in the 6-minute walk distance. **OR**
2. Patient remained stable from baseline in the 6-minute walk distance and the patient's World Health Organization (WHO) functional class remained stable or has improved.

Exclusion Criteria:

1. Contraindicated in patients with congestive heart failure caused by severe left ventricular systolic dysfunction
2. Contraindicated for long-term use in patients who develop pulmonary edema during dose initiation
3. Contraindicated if there is a known hypersensitivity to the drug or to structurally related compounds
4. Coverage excluded if pulmonary hypertension is secondary to pulmonary venous hypertension (i.e. left sided atrial or ventricular disease, left sided valvular heart disease) or disorders of the respiratory system (i.e. COPD, interstitial lung disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders)
5. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Veletri prescribing information
Flolan prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	None
5/24/17	Update to required medical information and exclusion criteria
6/7/18	Update to required medical information
6/26/19	None
4/29/20	None
5/12/21	Update to covered uses and added reauthorization criteria
5/25/22	Update to required medical information

Erythropoiesis Stimulating Agents (ESAs)

Procrit or Epogen; Aranesp; Mircera (methoxy polyethylene glycol epoetin-beta), Retacrit

Epogen/Procrit non-ESRD - J0885; Epogen/Procrit ESRD – Q4081
Aranesp non-ESRD - J0881; Aranesp ESRD - J0882
Mircera non-ESRD - J0888; Mircera ESRD - J0887
Retacrit (epoetin alfa-epbx) non-ESRD – Q5106; Retacrit (epoetin alfa-epbx) ESRD – Q5105

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for ESAs for a period of:

1 month (High risk for perioperative transfusions)
12 months (All other indications)

For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

Epoetin alfa (Procrit, Epogen), Epoetin alfa-epbx, (Retacrit) are FDA approved for:

1. 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 transfusion (RBC)
2. 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
3. Anemia due to zidovudine administered at $\leq$4200mg/week in HIV infected patients with endogenous serum erythropoietin levels of $\leq$500mUnits/ml
4. To reduce the need for allogeneic RBC transfusions among patients with perioperative hemoglobin > 10 to $\leq$ 13 g/dL who are at high risk for perioperative blood loss from elective, noncardiac, nonvascular surgery.

Darbepoetin alfa (Aranesp) is FDA approved for:

1. 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 transfusion (RBC)
2. 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

Methoxy polyethylene glycol epoetin-beta (Mircera) is FDA approved for:

1. Treatment of anemia associated with chronic kidney disease (CKD) in adult patients on dialysis and patients not on dialysis.
2. Treatment of anemia associated with chronic kidney disease (CKD) in 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

Required Medical Information: **NOTE:** If patient recently had a transfusion, use the most recent hemoglobin value before the transfusion.

Indication/Drug	Initial Authorization	Re-authorization
Anemia associated with Chronic Kidney Disease (CKD) in patients **NOT** on dialysis **Procrit/Epogen, Retacrit, Aranesp, Mircera** NOTE: CKD = Scr ≥ 3, CrCl <60 ml/min, or GFR <60 mL/min/1.73 m2	1. Diagnosis of CKD, **AND** 2. Hb < 10 g/dl (for adults and less than or equal to 11 g/dL for children) within 30 days of ESA therapy **AND** 3. Transferrin saturation ≥ 20% and ferritin ≥ 100 mcg/L, OR patient is receiving supplemental iron therapy IF serum ferritin <100 mcg/L or when TSAT <20%, **AND** **4.** B-12 and Folate levels have been drawn prior to initiating ESA unless member is currently on supplements (i.e. Vitamin B complex, Folic acid, Multivitamin, etc.). If not on supplement and no documentation of B12 and Folate levels, levels will be recommended by health plan pharmacist (but not required prior to approval)	1. Hb ≤ 11.5 g/dL in adults or ≤ 12 g/dL in children within 30 days of ESA therapy, **AND** 2. Documented increase in Hb since ESA therapy was initiated (for first re-authorization after initial therapy treatment), **AND** 3. Documented improvement in clinical symptoms with ESA therapy NOTE: Target hemoglobin may be higher if there is medical documentation showing the need, such as severe angina or severe pulmonary distress
Anemia associated with CKD or ESRD in patients on dialysis **Procrit/Epogen, Retacrit, Aranesp, Mircera** **NOTE: CKD =** Scr ≥ 3, CrCl <60 ml/min, or GFR <60 mL/min/1.73 m2	1. Diagnosis of CKD, **AND** 2. Hb < 10 g/dl within 30 days of ESA therapy, **AND** 3. Transferrin saturation ≥ 20% and ferritin ≥ 100 mcg/L, OR patient is receiving supplemental iron therapy IF serum ferritin < 100 mcg/L or when TSAT <20%, **AND** 4. B-12 and Folate levels have been drawn prior to initiating ESA unless member is currently on supplements (i.e. Vitamin B complex, Folic acid, Multivitamin, etc.). If not on supplement and no documentation of B12 and Folate levels, levels will be recommended by health plan pharmacist (but not required prior to approval)	1. Hb level < 10 g/dl within 30 days of ESA therapy (If the Hb level approaches or exceeds 11g/dl, reduce or interrupt the dose), **AND** 2. Documented increase in Hb since ESA therapy was initiated (for first re-authorization after initial therapy treatment), **AND** 3. Documented improvement in clinical symptoms with ESA therapy
Anemia in Zidovudine treated HIV-infected patients **Procrit/Epogen, Retacrit**	1. Serum erythropoietin levels of ≤ 500 mUnits/mL, **AND** 2. HIV infected patient is receiving zidovudine administered at ≤ 4200 mg/week, **AND** 3. Hb is ≤ 10 g/dl within 30 days of initial ESA dose, **AND** 4. Transferrin saturation ≥ 20% and ferritin ≥ 100 mcg/L, serum iron level OR patient is	1. Hb ≤ 12 g/dl within 30 days of ESA therapy, **AND** 2. Documented increase in Hgb needed to avoid RBC transfusions during the first 8 weeks of therapy (for first re-authorization after initial therapy treatment), **AND** 3. Documented improvement in clinical

	receiving supplemental iron therapy IF serum ferritin < 100 mcg/L or when TSAT <20%, **AND** 5. B-12 and Folate levels have been drawn prior to initiating ESA unless member is currently on supplements (i.e. Vitamin B complex, Folic acid, Multivitamin, etc.). If not on supplement and no documentation of B12 and Folate levels, levels will be recommended by health plan pharmacist (but not required prior to approval)	symptoms with ESA therapy
Reduction of allogeneic RBC transfusions in patients undergoing elective, noncardiac, nonvascular surgery **Procrit/Epogen, Retacrit** **NOTE:** DVT prophylaxis recommended during ESA therapy	1. Patient is undergoing hip or knee surgery, AND 2. Hb is ≤ 13 g/dl (this indication requires a lead time of at least 3 weeks prior to surgery), AND 3. Patient is unwilling or unable to donate autologous blood prior to surgery, AND 4. Patient is expected to lose more than 2 units of blood (i.e. high risk for perioperative blood loss from elective, noncardiac, nonvascular surgery), AND 5. Patient has had a work-up so that anemia appears to be that of chronic disease 6. Weekly dosage regimen for 3 weeks prior to surgery (e.g., days 21, -14, -7) and on the day of surgery will be covered	n/a

****Anemia secondary to myelosuppressive chemotherapy and Anemia associated with myelodysplastic syndrome (MDS) to be reviewed by medical oncology vendor if plan is contracted with vendor****

Exclusion Criteria:

1. Erythropoiesis Stimulating Agents are contraindicated in:
    a. Patients with uncontrolled hypertension
    b. Pure red cell aplasia (PRCA) that begins after treatment with erythropoietin protein drugs
    c. History of any serious allergic reactions to any of the erythropoietin protein drugs.
2. Per CMS National Coverage Determination, the following diagnoses are excluded from coverage:
    a. Anemia in cancer or cancer treatment patients due to folate deficiency, B-12 deficiency, iron deficiency, hemolysis, bleeding, or bone marrow fibrosis
    b. Anemia associated with treatment of acute and chronic myelogenous leukemias (CML, AML), or erythroid cancers
    c. Anemia of cancer not related to cancer treatment
    d. Anemia associated only with radiotherapy
    e. Prophylactic use to prevent chemotherapy-induced anemia
    f. Prophylactic use to reduce tumor hypoxia
    g. Patients with erythropoietin-type resistance due to neutralizing antibodies
    h. Anemia due to cancer treatment if patients have uncontrolled hypertension
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

Clinical note:

- In controlled trials of patients with chronic kidney disease, patients experienced greater risks for death, serious adverse cardiovascular reactions, and stroke when administered erythropoiesis-stimulating agents (ESAs) to target a hemoglobin level of greater than 11 g/dl.
- KDIGO Guidelines suggest that ESAs not be used to maintain hemoglobin above 11.5 g/dL in adult patients with CKD (upper boundary of hemoglobin in the control group of major ESA RCTs usually did not exceed 11.5g/dL).

References:

Procrit prescribing information, Epogen prescribing information, Aranesp prescribing information, Mircera prescribing information, Retacrit prescribing information
CMS Pub 100-02 *Medicare Benefit Policy Manual*, Chapter 15 – Covered Medical and Other Health Services, Section - 50.5.2.2 - Medicare Coverage of Epoetin Alfa (Procrit) for Preoperative Use (Rev. 1, 10-01-03)
KDIGO Clinical Practice Guidelines for Anemia in Chronic Kidney Disease https://kdigo.org/wp-content/uploads/2016/10/KDIGO-2012-Anemia-Guideline-English.pdf

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	None
5/24/17	Update to re-authorization criteria
6/7/18	Update to exclusion criteria; Addition of clinical note (KDIGO); Update to re-authorization period
8/20/18	Update to FDA approved indication for Mircera
1/1/19	Update to authorization period and required information to align with part D criteria
6/26/19	Update to initial criteria for Anemia with MDS and Reduction of allogeneic RBC transfusions in surgery
2/10/20	Addition of biosimilar to Procrit/Epogen – Retacrit effective 2/10/20
4/29/20	Update to approval time frame and reauthorization criteria; Remove Aranesp for use in reductions of allogeneic RBC transfusions
7/28/20	Update to initial criteria B12 and Folate requirements
5/12/21	Update to initial and re-authorization criteria
3/23/22	Update to initial criteria B12 and Folate requirements
5/25/22	Update to FDA indication; Update to required medical information; Update to reauthorization criteria; Update to references

EVENITY (romosozumab-AQQG) J3111

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage** for Evenity (romosozumab-aqqg) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
EVENITY (romosozumab-aqqg) is a sclerostin inhibitor indicated for the 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.

Required Medical Information:

1. Documented diagnosis of postmenopausal osteoporosis along with **ONE** of the following:
    a. The patient is unable to use oral therapy (i.e., upper gastrointestinal [GI] problems - unable to tolerate oral medication, lower GI problems - unable to absorb oral medications, trouble remembering to take oral medications or coordinating an oral bisphosphonate with other oral medications or their daily routine) **OR**
    b. The patient has an adequate trial of, intolerance to, or a contraindication to bisphosphonates (e.g., Fosamax [alendronate], Boniva [ibandronate]) **OR**
    c. The patient is at high risk for fracture (defined as *ONE* of the following):
        i. History of osteoporotic (i.e., fragility, low trauma) fracture(s)
        ii. 2 or more risk factors for fracture (e.g., history of multiple recent low trauma fractures, bone marrow density [BMD] T-score less than or equal to -2.5, corticosteroid use, or use of gonadotropin-releasing hormone [GnRH] analogs such as nafarelin, etc.)
        iii. No prior treatment for osteoporosis AND FRAX score greater than or equal to 20% for any major fracture OR greater than or equal to 3% for hip fracture
2. Patient has NOT received a total of 12 months of Evenity therapy

Exclusion Criteria:

1. Limit duration of use to 12 monthly doses. If osteoporosis therapy remains warranted, continued therapy with an anti-resorptive agent should be considered.
2. Pre-existing hypocalcemia must be corrected prior to initiating therapy with EVENITY.
3. Should not be initiated in patients who have had a myocardial infarction or stroke within the preceding year.
4. A history of systemic hypersensitivity to romosozumab or to any component of the product formulation.
5. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Evenity prescribing information

Version History:

Last Reviewed Date	Updates / Revisions
6/19/19	Addition to part B criteria with effective date 1/1/20
1/13/20	J Code updated
4/29/20	None
5/12/21	None
5/25/22	Addition to exclusion criteria

EVKEEZA (evinacumab-dgnb) J1305

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, **the plan may authorize coverage for 12 Months**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
EVKEEZA is an ANGPTL3 (angiopoietin-like 3) inhibitor indicated as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of adult and pediatric patients, aged 12 years and older, with homozygous familial hypercholesterolemia (HoFH). Limitations of Use: • The safety and effectiveness of EVKEEZA have not been established in patients with other causes of hypercholesterolemia, including those with heterozygous familial hypercholesterolemia (HeFH). • The effects of EVKEEZA on cardiovascular morbidity and mortality have not been determined.

Required Medical Information:
1. Homozygous familial hypercholesterolemia (HoFH)
    a. Diagnosis of homozygous familial hypercholesterolemia (HoFH), AND
    b. The patient is 12 years of age or older, AND
    c. Evkeeza will be used as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies. (e.g., statins, PCSK9 inhibitors, ezetimibe, lomitapide, lipoprotein apheresis

Reauthorization:
1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:
1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. History of serious hypersensitivity reactions to evinacumab-dgnb or to any of the excipients in EVKEEZA.

References:
Evkeeza prescribing information

Version History

Last Reviewed Date	Updates / Revisions
11/10/21	Addition to part B criteria with effective date 1/1/2022
5/25/22	No updates

FABRAZYME (agalsidase beta) J0180

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Fabrazyme (agalsidase beta) for **1 year initial and reauthorization.** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Fabrazyme is a hydrolytic lysosomal neutral glycosphingolipid-specific enzyme indicated for the treatment of adult and pediatric patients 2 years of age and older with confirmed Fabry disease.

Required Medical Information:

1. Member must have a diagnosis of Fabry disease, **AND**
2. Prescribed by or in consultation with a geneticist, endocrinologist, a metabolic disorder sub-specialist, or a physician who specializes in the treatment of lysosomal storage disorders, **AND**
3. The patient has a laboratory test demonstrating deficient alpha-galactosidase A activity in leukocytes or fibroblasts OR has a molecular genetic test demonstrating mutations in the galactosidase alpha gene, **AND**
4. The patient is 2 years of age or older

Reauthorization:

1. Diagnosis of Fabry disease, **AND**
2. Physician attestation that the patient has demonstrated improvement or stabilization

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Fabrazyme prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	None
5/24/17	Update to FDA indication wording/removal of disease description
6/7/18	None
6/26/19	None

4/29/20	Update to authorization time frame, required information. Addition of reauthorization criteria.
5/12/21	Update to authorization time frame, wording of FDA indication, and required information
5/25/22	Update to required medical information

FACTOR PRODUCTS Factor VIIa –NovoSeven RT J7189, SevenFact J7212

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Factor VIIa product for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
NovoSeven RT: 1. Treatment of bleeding episodes and peri-operative management in adults and children with hemophilia A or B with inhibitors, congenital Factor VII (FVII) deficiency, and Glanzmann's thrombasthenia with refractoriness to platelet transfusions, with or without antibodies to platelets 2. Treatment of bleeding episodes and peri-operative management in adults with acquired hemophilia SevenFact (Coagulation Factor VIIa-jncw): 1. For the treatment and control of bleeding episodes in adult and adolescents 12 years or older with hemophilia A or B with inhibitors. 2. Limitation of Use - is not indicated to treat patients with congenital Factor VII deficiency

Required Medical Information:

1. **SEVENFACT** - Bleeding episode or surgical intervention/invasive procedure, AND diagnosis of Hemophilia A or B with inhibitors
2. **NOVOSEVEN RT** - Bleeding episode or surgical intervention/invasive procedure, **AND** one of the following:
    a. Hemophilia A or B with inhibitors, **OR**
    b. Congenital Factor VII deficiency **OR**
    c. Glanzmann's thrombasthenia with refractoriness to platelet transfusions, with or without antibodies to platelets, **OR**
    d. Acquired hemophilia (adults only)

Exclusion Criteria:

1. NovoSeven Coagulation Factor VIIa (Recombinant) should not be administered to patients with known hypersensitivity to NovoSeven or any of its components
2. SevenFact should not be administered to patients with known allergy to rabbits or rabbit proteins. Severe hypersensitivity reaction to SEVENFACT or any of its components
3. Administer with caution in patients with known hypersensitivity to mouse, hamster, or bovine proteins.
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:
NovoSeven RT prescribing information, SevenFact prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	None
5/24/17	Update to required information
6/7/18	None
6/26/19	None
4/29/20	None
5/12/21	Addition of SevenFact effective 1/1/22
5/25/22	None

FACTOR PRODUCTS Factor VIII

Factor VIII Advate J7192, Koate-DVI J7190, Hemofil M J7190, Eloctate (Factor VIII, Fc fusion protein) J7205, Kogenate FS/Kogenate FS with BIO-SET J7192, Recombinate J7192, Xyntha J7185, Novoeight J7182, Obizur J7188, Nuwiq J7209, Adynovate J7207, Alphanate/VWF complex J7186, Kovaltry J7211, Wilate J7183, Humate-P J7187, Afstyla J7210, Jivi J7208, Esperoct J7204

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Factor VIII product for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Human (Koate DVI, Hemofil M, Alphanate/VWF complex, Wilate, Humate-P **Recombinant products (Advate, Eloctate, Kogenate FS/Kogenate FS with BIO-SET, Recombinate, Xyntha, Novoeight, Obizur, Nuwiq, Adynovate, Kovaltry, Jivi, Esperoct** • Control of bleeding episodes in patients with hemophilia A (classical hemophilia) • Routine prophylaxis of bleeding episodes in patients with hemophilia A (classical hemophilia) • Surgical prophylaxis in patients with hemophilia A **Recombinant Factor not indicated for treatment of von Willebrand disease

Required Medical Information:

1. Documentation of bleeding episode or surgical intervention or need for short-term routine prophylaxis to reduce frequency of spontaneous bleeding episodes, **AND**
2. Hemophilia A with decreased activity of clotting factor VIII or acquired factor VIII deficiency

Exclusion Criteria:

1. Factor product VIII is contraindicated in patients with history of anaphylactic or severe systemic response to plasma-derived products, any ingredient in the formulation, or components of the container.
2. Factor VIII products that contain bovine, hamster or mouse protein are contraindicated in patients with bovine protein, hamster protein, AND/OR murine protein hypersensitivity.
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Factor products prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	Update to FDA indications
5/24/17	Update J codes; Addition of factor products to criteria
6/7/18	Update to Kovaltry J code
6/26/19	Addition of factor products to criteria
4/29/20	None
5/12/21	Addition of Jivi and Esperoct effective 1/1/22
5/25/22	Remove Monoclate-P and Helixate FS (discontinued items)

FACTOR PRODUCTS Factor IX

Factor IX (AlphaNine SD J7193, BeneFix J7195 , Mononine J7193, Rixubis J7200, Alprolix J7201, Ixinity J7195, Bebulin/Bebulin VH J7194, Profilnine/Profilnine SD J7194, Idelvion J7202, Rebinyn J7203)

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Factor IX product for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
• Prevention and control of bleeding in patients with factor IX deficiency (hemophilia B [Christmas disease]). • **BeneFix, Rixubis, Alprolix, Ixinity, Idelvion, Rebinyn** also indicated for perioperative management in patients with hemophilia B.

Required Medical Information:

1. Documentation of bleeding episode or need for prophylaxis, **OR**
2. Surgical intervention (for BeneFix, Rixubis, Alprolix, Ixinity, Idelvion), **AND**
3. Hemophilia B with factor IX deficiency

Exclusion Criteria:

1. Factor product IX is contraindicated in patients with known hypersensitivity to hamster or mouse protein (product specific).
2. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Factor products prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	Update to FDA indications
5/24/17	Update J codes; Addition of factor products to criteria
6/7/18	Updated wording for required information based on FDA approved indications
6/26/19	Addition of factor products to criteria
4/29/20	None

5/12/21	None
5/25/22	None

FACTOR PRODUCTS - MISCELLANEOUS

RiaSTAP J7178, Corifact J7180, Tretten J7181, Feiba/Feiba NF J7198, Hemophilia clotting factor, NOC J7199, Coagadex J7175, *Hemlibra J7170*

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for the specified factor product for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Corifact is a Factor XIII concentrate for adult and pediatric patients with congenital Factor XIII deficiency for: • Routine prophylactic treatment • Peri-operative management of surgical bleeding **Coagadex** (J7175) - Coagulation Factor X (Human), is a plasma-derived human blood coagulation factor indicated in adults and children with hereditary Factor X deficiency for: • Routine prophylaxis to reduce the frequency of bleeding episodes, • On-demand treatment and control of bleeding episodes • Perioperative management of bleeding in patients with mild and moderate hereditary Factor X deficiency **Tretten** is a coagulation Factor XIII A-subunit (recombinant) indicated for routine prophylaxis of bleeding in patients with congenital factor XIII A-subunit deficiency **RiaSTAP**, Fibrinogen Concentrate (human), is a human blood coagulation factor indicated for the treatment of acute bleeding episodes in patients with congenital fibrinogen deficiency, including afibrinogenemia and hypofibrinogenemia **Feiba/Feiba NH** is an Anti-Inhibitor Coagulant Complex indicated for use in hemophilia A and B patients with inhibitors for: • Control and prevention of bleeding episodes • Perioperative management • Routine prophylaxis to prevent or reduce the frequency of bleeding episodes **Hemlibra** is a bispecific factor IXa- and factor X-directed antibody indicated for routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adult and pediatric patients ages newborn and older with hemophilia A (congenital factor VIII deficiency) with or without factor VIII inhibitors

Required Medical Information:

1. Refer above for product specific FDA approved indications

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. Do not use in patients with known anaphylactic or severe systemic reactions to human plasma-derived products (Corifact, RiaSTAP)
3. Tretten is NOT indicated for use in patients with congenital factor XIII B-subunit deficiency
4. Feiba/Feiba NH contraindicated in patient with:
    a. History of anaphylactic or severe hypersensitivity reactions to Feiba or any of its components, including factors of the kinin generating system
    b. Disseminated intravascular coagulation
    c. Acute thrombosis or embolism (including myocardial infarction)
5. Feiba is NOT indicated for the treatment of bleeding episodes resulting from coagulation factor deficiencies in the absence of inhibitors to factor VIII or factor IX

References:

Factor products prescribing information

Version History

Last Reviewed Date	Updates / Revisions
5/24/17	Addition to Part B criteria with effective date 5/24/17
6/26/19	Addition of factor products to criteria
4/29/20	Removal of Kcentra
2/17/21	Addition of Hemlibra to Part B criteria effective 9/1/21
5/12/21	None
5/25/22	None

FASENRA (benralizumab) J0517

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Fasenra (benralizumab) for **6 months (initial) and 1 year (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Fasenra (benralizumab) is an interleukin-5 receptor alpha-directed cytolytic monoclonal antibody (IgG1, kappa) indicated for the add-on maintenance treatment of patients with severe asthma age 12 years and older, and with an eosinophilic phenotype. Fasenra should be administered by a healthcare professional.

****Initially review for B vs D coverage****

- Fasenra Pen (Autoinjector) – intended for administration by patients/caregivers after proper training and after the healthcare provider determines it is appropriate
    - Not covered under part B – needs to be routed to PBM to evaluate coverage under Part D
- Fasenra prefilled syringe – intended for administration by healthcare provider only
    - Covered under part B – health plan to review

Required Medical Information:

1. Diagnosis of severe asthma, **AND**
2. Fasenra will be used for add-on maintenance treatment, **AND**
3. Peripheral blood eosinophil count of greater than or equal to 150 cells per microliter within the previous 6 weeks (prior to treatment with any anti-interleukin (IL)-5 therapy, **AND**
4. Has received at least 3 consecutive months of combination therapy with an inhaled corticosteroid **AND** one of the following: inhaled LABA, inhaled long-acting muscarinic antagonist, leukotriene receptor antagonist, or theophylline (NOTE: An exception to the requirement for a trial of one additional asthma controller/maintenance medication can be made if the patient has already received anti-IL-5 therapy (e.g., Cinqair, Fasenra, Nucala) used concomitantly with an ICS for at least 3 consecutive months), **AND**
5. Patient's asthma is uncontrolled or was uncontrolled prior to starting any anti-IL therapy as defined by **ONE** of the following:
    a. Patient experienced one or more asthma exacerbations requiring treatment with systemic corticosteroids in the previous year, **OR**
    b. Patient experienced one or more asthma exacerbation requiring hospitalization or an Emergency Department (ED) visit in the previous year, **OR**
    c. Patient has a FEV1 less than 80 percent predicted, **OR**
    d. Patient has an FEV1/FVC less than 0.80, **OR**
    e. Patient's asthma worsens upon tapering of oral corticosteroid therapy, **AND**
6. Prescribed by or in consultation with an allergist, immunologist, or pulmonologist, **AND**

7. Patient is 12 years of age or older

Reauthorization:

1. Patient has responded to Fasenra therapy as determined by the prescribing physician (e.g., decreased asthma exacerbations, decreased asthma symptoms, decreased hospitalizations, emergency department (ED)/urgent care, or physician visits due to asthma, decreased requirement for oral corticosteroid therapy) **AND**
2. Patient continues to receive therapy with an inhaled corticosteroid

Exclusion Criteria:

1. Fasenra is not indicated for relief of acute bronchospasm or status asthmaticus.
2. Concurrent use with Xolair or another Anti-Interleukin (IL) Monoclonal Antibody
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Fasenra prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/18/17	Addition to part B criteria with effective date 3/1/18
6/7/18	Update to hospital outpatient C code
1/1/19	Updates to authorization period, required information, and exclusion criteria to align with part D criteria; Addition of reauthorization criteria to align with part D criteria
6/26/19	Update to J code, Removal of C code
12/10/19	Addition of BvsD determination
2/10/20	None
4/29/20	Update to required information
5/12/21	None
5/25/22	None

FERAHEME (ferumoxytol) Q0138 (non-ESRD); Q0139 (ESRD)

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Feraheme (ferumoxytol) for **2 months.** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Feraheme (ferumoxytol) is an iron replacement product indicated for the treatment of iron deficiency anemia (IDA) in adult patients who have intolerance to oral iron or have had unsatisfactory response to oral iron, OR who have chronic kidney disease

Required Medical Information:

1. Diagnosis of iron deficiency anemia in adult patients, **AND**
    a. CBC and iron studies obtained within the last month to confirm diagnosis, more specifically:
        i. Hgb <13 g/dL in males and <12 g/dL in females, **AND**
        ii. Ferritin <30 µg/L and/or TSAT <20%, **AND**
    b. Have intolerance to or unsatisfactory response to oral iron, **OR**
    c. Have chronic kidney disease

Exclusion Criteria:

1. History of allergic reaction to any intravenous iron product
2. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Feraheme prescribing information
Hematology Am Soc Hematol Educ Program (2019) 2019 (1): 315–322

Version History

Last Reviewed Date	Updates / Revisions
8/20/18	Addition to part B criteria with effective date 1/1/19
6/26/19	None
4/29/20	None
2/17/21	Update to required information to add laboratory parameters
5/12/21	None
5/25/22	None

HYALURONAN

Synvisc J7325, Synvisc-One J7325, Orthovisc J7324, Euflexxa J7323, Monovisc J7327, GenVisc 850 J7320, Supartz J7321, Hyalgan J7321, Visco-3 J7321, Gelsyn-3 J7328, Gel-One J7326, Hymovis J7322, Durolane J7318, Trivisc J7329, Triluron J7332, Synojoynt J7331

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for hyaluronan for **3 months.** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Purified natural hyaluronates have been approved by the FDA for the treatment of osteoarthritis of the knee in patients who have failed to respond to simple analgesics and conservative nonpharmacologic therapy.

Required Medical Information:

1. **Osteoarthritis of the knee**
    a. Diagnosis of osteoarthritis of the knee, **AND**
    b. Radiographical evidence to support the diagnosis of osteoarthritis, **AND**
    c. Review of Local Coverage Determination (LCD) for state in which patient resides to apply additional clinical criteria, **OR**
    d. If no LCD exists or LCD does not define a timeframe for past therapy tried, then use the following criteria: patients have failed to respond adequately to at least a three-month period of conservative nonpharmacologic therapy and simple analgesics (e.g., acetaminophen)

Reauthorization:

1. Significant improvement in knee pain and known improvement in functional capacity resulted from the previous series of injections which has been documented in the medical record, **AND**
2. At least six months have elapsed since the prior series of injections, **AND**
3. Review of Local Coverage Determination (LCD) for state in which patient resides to apply additional clinical criteria

Exclusion Criteria:

1. Infections in the area of the injection site
2. Skin diseases in the area of the injection site
3. Allergy to avian or avian-derived products, including eggs, feathers, or poultry (Orthovisc)
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Hyaluronan products prescribing information

Version History

Last Reviewed Date	Updates / Revisions
8/20/18	Addition to part B criteria with effective date 1/1/19
6/26/19	Addition of Trivisc (J7329) effective date 1/1/20
2/10/20	Addition of Triluron J7332 and Synojoynt J7331 effective 2/10/20
4/29/20	Update to exclusion criteria
5/12/21	None
10/8/21	Addition to required medical information
11/10/21	Addition to required medical information
5/25/22	None

ILARIS (canakinumab) J0638

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Ilaris (canakinumab) for **Still's- 3 months initial, 1 year reauthorization, CAPS/SJIA-3 months initial, 1 years reauthorization, FMF/HIDS/MKD/TRAPS-4 months initial, 3 years reauthorization.** For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Ilaris is an interleukin-1 β blocker indicated for the treatment of: Periodic Fever Syndromes: • Cryopyrin-Associated Periodic Syndromes (CAPS) in adults and children 4 years of age and older including Familial Cold Autoinflammatory Syndrome (FCAS) and Muckle-Wells Syndrome (MWS) • Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) in adult and pediatric patients • Hyperimmunoglobulin D Syndrome (HIDS)/Mevalonate Kinase Deficiency (MKD) in adult and pediatric patients • Familial Mediterranean Fever (FMF) in adult and pediatric patients Active Still's Disease: • Including Adult-Onset Still's Disease (AOSD) and Systemic Juvenile Idiopathic Arthritis (SJIA) in patients aged 2 years and older.

Required Medical Information:

1. Patient has been tested for latent TB prior to initiating Ilaris therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
2. Patient has one of the following documented diagnoses:
    a. Cryopyrin-Associated Periodic Syndromes (CAPS) such as Familial Cold Autoinflammatory Syndrome (FCAS) **OR** Muckle-Wells Syndrome (MWS):
        i. Are at least 4 years of age **AND**
        ii. Prescribed by or in consultation with a rheumatologist, geneticist, allergist/immunologist, or dermatologist, **OR**
    b. Tumor Necrosis Factor Receptor Associated Periodic Syndrome (TRAPS) **OR** Hyperimmunoglobulin D Syndrome (HIDS) **OR** Mevalonate Kinase Deficiency (MKD):
        i. Prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, oncologist, hematologist **OR**
    c. Familial Mediterranean Fever (FMF):
        i. Prescribed by or in consultation with a rheumatologist, nephrologist, geneticist, gastroenterologist, oncologist, hematologist **OR**
    d. Active Still's Disease – including Adult Onset and Systemic Juvenile Idiopathic Arthritis (SIJA):
        i. Are at least 2 years of age or older **AND**
        ii. Prescribed by or in consultation with a rheumatologist **AND**

iii. Patient has tried at least 2 other biologics (tocilizumab, abatacept, TNF antagonists (e.g. etanercept, adalimumab, infliximab) **OR**
iv. pt has features of poor prognosis (e.g. arthritis of the hip, radiographic damage, 6-month duration of significant active systemic disease, defined by fever, elevated inflammatory markers, or requirement for treatment with systemic glucocorticoids **OR**
v. Pt has features of Still's disease or SJIA with active systemic features with concerns of progression to macrophage activation syndrome (MAS) [as determined by the prescribing physician]

Reauthorization:

1. Patient is being treated CAPS/MWS/FCAS/SJIA/FMF/HIDS/MKD/TRAPS/Still's - approve if the pt had a response to therapy as determined by prescribing physician.

Exclusion Criteria:

1. Treatment with Ilaris should not be initiated in patients with active infection requiring medical intervention.
2. Live vaccines should not be given concurrently with Ilaris.
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
4. Coverage excluded when used in combination with concurrent biologic therapy (e.g.TNF antagonists, etanercept, adalimumab, certolizumab pegol, golimumab, infliximab), anakinra, or rilonacept.

References:

Ilaris prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	None
5/24/17	Additional FDA approved diagnoses added
6/7/18	None
8/20/18	Update to required information
6/26/19	None
4/29/20	None
2/17/21	Addition of reauthorization criteria
5/12/21	Addition of FDA approved diagnosis, Update initial criteria and time frames
5/25/22	Update timeframe, reauthorization criteria, add to exclusion criteria

IMMUNE GLOBULIN

Intravenous: Gammagard J1569, Gammagard S/D J1566, Gammaked J1561, Gamunex-C J1561, Privigen J1459, Bivigam J1556, Carimune NF J1566, Flebogamma/Flebogamma DIF J1572, Gammaplex J1557, Octagam J1568, Panzyga J1599, Asceniv J1554

Subcutaneous: Hizentra J1559, Hyqvia J1575, Cuvitru J1555, Cutaquig J1551, Xembify J1558

Intramuscular: GamaSTAN J1460 or GamaSTAN S/D J1560

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Immune Globulin for **one year.** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Immune globulin is a blood product prepared from the pooled plasma of human donors. It contains a broad range of antibodies. Refer to individual product inserts for FDA approved uses. Subcutaneous: • FDA approved for treatment of Primary Immunodeficiency • Hizentra: maintenance therapy in adults with chronic inflammatory demyelinating polyneuropathy (CIDP) Intramuscular: FDA approved for the following: hepatitis A prophylaxis, measles, post-exposure prophylaxis for rubella in pregnancy, varicella prophylaxis when varicella zoster immune globulin not available

Required Medical Information:

Initial authorization

1. **Determination for B v D coverage:**
   a. Covered under Part B if:
      i. Patient is receiving IVIG infusion at a physician's office, **OR**
      ii. Patient is receiving IVIG via an external infusion pump and IVIG is covered under the applicable Medicare coverage policies (National and/or Local Coverage Policies), **OR**
      iii. Patient is receiving IVIG at home and has a diagnosis of primary immune deficiency disease or for the treatment of one of the following diagnoses: D80.0, D80.5, D81.0, D81.1, D81.2, D81.6, D81.7, D81.89, D81.9, D82.0, D83.0, D83.1, D83.2, D83.8, D83.9
   b. Covered under Part D if:
      i. Patient is receiving IVIG at home and does NOT have a diagnosis of primary immune deficiency disease or one of the following diagnoses: D80.0, D80.5, D81.0, D81.1, D81.2, D81.6, D81.7, D81.89, D81.9, D82.0, D83.0, D83.1, D83.2, D83.8, D83.9

AND,

2. **Evaluation of coverage for requested indication using:**
   a. Product labeling (FDA indication), OR
   b. Medical compendia, OR
   c. NCD and/or LCD

   Possible covered indications (not an exhaustive list):
   - Primary humoral immunodeficiency (congenital agammaglobulinemia, common variable immunodeficiency, Wiskott-Aldrich syndrome, X-linked agammaglobulinemia, severe combined immunodeficiency)
   - Immune thrombocytopenic purpura (ITP)
   - Chronic lymphocytic leukemia with associated hypogammaglobulinemia
   - Chronic inflammatory demyelinating polyneuropathy (CIDP)
   - Guillain-Barre syndrome
   - Myasthenia gravis
   - Multifocal motor neuropathy (MMN)
   - Short-term treatment of autoimmune mucocutaneous blistering diseases: pemphigus vulgaris, pemphigus foliaceus, bullous pemphigoid, mucous membrane pemphigoid (cicatricial pemphigoid), and epidermolysis bullosa acquisita) – per NCD
   - Scleromyxedema
   - Hemolytic anemia
   - Polymyositis
   - Dermatomyositis
   - Other indications as specified in NCD, LCDs, compendia that support medical necessity

AND,

3. **Review of National Coverage Determination or Local Coverage Determination for state in which patient resides to apply appropriate clinical criteria**

Reauthorization:

1. Documentation of improvement in symptoms/clinical status, **AND**
2. Review of National Coverage Determination or Local Coverage Determination for state in which patient resides to apply appropriate documentation and clinical criteria

Exclusion Criteria:

1. IgA deficient patients with antibodies against IgA and a history of hypersensitivity
2. Privigen and Hizentra contraindicated in hyperprolinemia
3. Gammaplex contraindicated in patients with a hereditary intolerance to fructose
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Immune globulin products prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	Addition to exclusion criteria; Addition of disclaimer
5/24/17	Addition to indications for subcutaneous and intramuscular use; Addition to exclusion criteria
6/7/18	Update to required information; Update to exclusion criteria; Update to reauthorization criteria
1/1/19	Update to authorization period to align with part D criteria
6/26/19	Addition of Cuvitru and Cutaquig - effective 1/1/20
9/24/19	Addition of Xembify – effective 3/1/20
2/10/20	Addition of indication for subcutaneous Hizentra
4/29/20	Addition of Panzyga and Asceniv – effective 1/1/21; Addition to exclusion criteria
5/12/21	Update Asceniv JCode
8/10/21	Update Xembify JCode
7/14/22	Update Cutaquig Jcode
8/23/22	None

INJECTAFER (ferric carboxymaltose) J1439

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Injectafer (ferric carboxymaltose) for **2 months**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Injectafer (ferric carboxymaltose) is an iron replacement product indicated for the treatment of iron deficiency anemia (IDA) in: • Adults and pediatric patients 1 year of age and older who have either intolerance to oral iron or an unsatisfactory response to oral iron. • Adult patients who have non-dialysis dependent chronic kidney disease.

Required Medical Information:
1. Diagnosis of iron deficiency anemia in adult patients, **AND**
    a. CBC and iron studies obtained within the last month to confirm diagnosis, more specifically:
        i. Hgb <13 g/dL in males and <12 g/dL in females, **AND**
        ii. Ferritin <30 µg/L and/or TSAT <20%, **AND**
    b. Have intolerance to or unsatisfactory response to oral iron, **OR**
    c. Have non-dialysis chronic kidney disease

Exclusion Criteria:
1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. Hypersensitivity to Injectafer or any of its inactive components.

References:
Injectafer prescribing information
Hematology Am Soc Hematol Educ Program (2019) 2019 (1): 315–322

Version History

Last Reviewed Date	Updates / Revisions
8/20/18	Addition to part B criteria with effective date 1/1/19
6/26/19	None
4/29/20	None
2/17/21	Update to required information to add laboratory parameters
8/10/21	None
8/23/22	Update to FDA indication; Addition to exclusion criteria

KRYSTEXXA (pegloticase) J2507

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Krystexxa (pegloticase) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Krystexxa (pegloticase) is a PEGylated uric acid specific enzyme indicated for the treatment of chronic gout in adult patients refractory to conventional therapy. Gout refractory to conventional therapy occurs in patients who have failed to normalize serum uric acid and whose signs and symptoms are inadequately controlled with xanthine oxidase inhibitors at the maximum medically appropriate dose or for whom these drugs are contraindicated. Pegloticase is not recommended for the treatment of asymptomatic hyperuricemia.

Required Medical Information:

1. **Gout**
    a. Documentation of chronic symptomatic gout diagnosis, **AND**
    b. Documentation of adequate trial of or contraindication to conventional therapy with xanthine oxidase inhibitors (i.e. allopurinol, Uloric), **AND**
    c. Documentation that oral urate-lowering agents will be discontinued prior to initiation of Krystexxa, **AND**
    d. Uric acid level of greater than or equal to 6 mg/dl, **AND**
    e. Patient must be 18 years old or older

Reauthorization:

1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:

1. A history of glucose-6-phosphate dehydrogenase (G6PD) deficiency.
2. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
3. Patients with history of serious hypersensitivity reactions, including anaphylaxis, to KRYSTEXXA or any of its components.

References:

Krystexxa prescribing information

Version History

Last Reviewed Date	Updates / Revisions
6/7/16	None
5/24/17	Update to required information
8/20/18	None
9/24/19	None
7/28/20	None
2/17/21	Addition of reauthorization criteria
8/10/21	None
8/23/22	Addition to exclusion criteria

LEMTRADA (alemtuzumab) J0202

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Lemtrada (alemtuzumab) **12 months (initial and reauthorization).** For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Lemtrada (alemtuzumab) is a CD52-directed cytolytic monoclonal antibody indicated for the treatment of patients with relapsing forms of multiple sclerosis (MS), to include relapsing-remitting disease and active secondary progressive disease, in adults. Because of its safety profile, the use of Lemtrada should generally be reserved for patients who have had an inadequate response to two or more drugs indicated for the treatment of MS. Limitations of Use: LEMTRADA is not recommended for use in patients with clinically isolated syndrome (CIS) because of its safety profile

Required Medical Information:

1. Diagnosis of relapsing form of multiple sclerosis (MS), to include relapsing-remitting disease and active secondary progressive disease, who have not completed a course of Lemtrada (including pt who started but not completed Lemtrada tx) **AND**
2. Prescribed by or in consultation with a neurologist or a physician who specializes in the treatment of MS, **AND**
3. The patient is 17 years of age or older **AND**
4. The patient must have had an inadequate response or was unable to tolerate according to the prescribing physician TWO disease modifying agents used for MS (e.g. Tecfidera, Gilenya, Aubagio, Tysabri, Avonex, Rebif, Plegridy, Betaseron, Extavia, Copaxone, Novantrone) **OR**
5. The patient has a highly-active or aggressive multiple sclerosis by meeting one of the following:
    a. The patient has demonstrated rapidly-advancing deterioration(s) in physical functioning (e.g., loss of mobility/or lower levels of ambulation, severe changes in strength or coordination) **OR**
    b. Disabling relapse(s) with suboptimal response to systemic corticosteroids **OR**
    c. Magnetic resonance imaging (MRI) findings suggest highly-active or aggressive multiple sclerosis (e.g., new, enlarging, or a high burden of T2 lesions or gadolinium lesions) **OR**
    d. Manifestation of multiple sclerosis-related cognitive impairment.

Reauthorization:

1. Relapsing form of multiple sclerosis (MS), to include relapsing-remitting disease and active secondary progressive disease, in adults. **AND**
2. At least 12 months has elapsed since receiving the most recent course of Lemtrada, **AND**

3. Prescribed by or in consultation with a neurologist or a physician who specializes in the treatment of MS

Exclusion Criteria:

1. Known hypersensitivity or anaphylactic reactions to alemtuzumab or any of the excipients in LEMTRADA.
2. Contraindicated in patients who are infected with Human Immunodeficiency Virus (HIV) because Lemtrada causes prolonged reductions of CD4+ lymphocyte counts.
3. Contraindicated in patients with active infection.
4. Current Use of Lemtrada with Other Disease-Modifying Agents Used for Multiple Sclerosis (MS).
5. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

Clinical Note:

- Complete any necessary immunizations 6 weeks prior to the start of Lemtrada
- Determine history of varicella infection or vaccination for varicella zoster virus. Consider vaccination in patients that are anti-body negative to varicella zoster virus. Postpone initiation of Lemtrada for 6 weeks following vaccination for varicella zoster virus.
- Do not administer live viral vaccines following a course of LEMTRADA
- Measure the urine protein to creatinine ratio prior to initiation of treatment.
- Conduct the following laboratory tests at baseline and at periodic intervals until 48 months after the last treatment course of LEMTRADA in order to monitor for early signs of potentially serious adverse effects:
    - Complete blood count (CBC) with differential (prior to treatment initiation and at monthly intervals thereafter)
    - Serum creatinine levels (prior to treatment initiation and at monthly intervals thereafter)
    - Urinalysis with urine cell counts (prior to treatment initiation and at monthly intervals thereafter)
    - A test of thyroid function, such as thyroid stimulating hormone (TSH) level (prior to treatment initiation and every 3 months thereafter)
    - Serum transaminases (alanine aminotransferase [ALT] and aspartate aminotransferase [AST]) and total bilirubin levels (prior to treatment initiation and periodically thereafter)
- Conduct baseline and yearly skin exams to monitor for melanoma

References:

Lemtrada prescribing information

Version History

Last Reviewed Date	Updates / Revisions
9/13/16	Updated J code; Addition of clinical notes
8/22/17	Update to wording of FDA indications and contraindications
8/20/18	Update to required information; Update to re-authorization; Change in authorization period – effective 1/1/19
9/24/19	Update to re-authorization criteria; Change in authorization period
7/28/20	Update to indication; Update to required information; Update to re-authorization
8/10/21	Update to required information, reauthorization criteria, and exclusion criteria
8/23/22	Addition to exclusion criteria and clinical note

NPLATE (romiplostim) J2796

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Nplate (romiplostim) for **four months (initial ITP), 12 months (initial HSARS),** and **12 months (reauthorization**). For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
1. NPLATE (romiplostim) is a thrombopoietin receptor agonist indicated for the treatment of thrombocytopenia in patients with chronic immune (idiopathic) thrombocytopenic purpura (ITP) who have had an insufficient response to corticosteroids, immunoglobulins, or splenectomy. Romiplostim should be used only in patients with ITP whose degree of thrombocytopenia and clinical condition increases the risk for bleeding. Romiplostim should not be used in an attempt to normalize platelet counts. 2. Nplate is indicated to increase survival in adults and in pediatric patients (including term neonates) acutely exposed to myelosuppressive doses of radiation (Hematopoietic Syndrome of Acute Radiation Syndrome [HSARS]).

Required Medical Information:
1. Patient has a diagnosis of Hematopoietic Syndrome of Acute Radiation Syndrome (HSARS: The patient was acutely exposed to myelosuppressive doses of radiation), **OR**
2. Patient has a diagnosis of immune thrombocytopenia (ITP) and meet ALL of the following criteria:
    a. The patient is at least 1 year old or older
        i. Age 1-17: patient has diagnosis of ITP for at least 6 months
    b. The patient had a trial of or contraindication to corticosteroids or immunoglobulins, or had an insufficient response to splenectomy
    c. The medication is being prescribed by or given in consultation with a hematologist or immunologist.

Reauthorization:
1. Patient has a diagnosis of Hematopoietic Syndrome of Acute Radiation Syndrome (HSARS: The patient was acutely exposed to myelosuppressive doses of radiation), **OR**
2. Patient is being treated for chronic immune thrombocytopenia (ITP) **AND** physician attestation of a clinical response

Exclusion Criteria:
1. Not indicated for the treatment of thrombocytopenia due to myelodysplastic syndrome (MDS) or any cause of thrombocytopenia other than chronic ITP
2. Should not be used in attempt to normalize platelet counts
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:
Nplate prescribing information

Version History

Last Reviewed Date	Updates / Revisions
9/13/16	None
3/3/17	Removal of Nplate NEXUS program from required information
8/22/17	None
8/20/18	Update to wording of exclusion criteria; Update to authorization period – effective 1/1/19; Addition of reauthorization criteria (effective 1/1/19)
9/24/19	Update to required medical information, authorization period and reauthorization criteria
7/28/20	Update to required medical information
8/10/21	Addition to FDA indication; Update to required medical information, reauthorization criteria, and initial approval time frame
8/23/22	None

OCREVUS (ocrelizumab) J2350

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Ocrevus (ocrelizumab) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Ocrevus (ocrelizumab) is a CD20-directed cytolytic antibody indicated for the treatment of patients with Relapsing forms of multiple sclerosis (MS), to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults OR primary progressive forms of multiple sclerosis.

Required Medical Information:

1. **Primary progressive multiple sclerosis (PPMS)**
    a. Documented diagnosis of primary progressive multiple sclerosis, **AND**
    b. The patient is 18 years of age or older, **AND**
    c. Hepatitis B virus and quantitative serum immunoglobulin screening are required before the first dose. If no documentation of HBV or immunoglobulin test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    d. Prescribed by or in consultation with, a physician who specializes in the treatment of MS and/or a neurologist
2. **Relapsing multiple sclerosis (to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease)**
    a. Documented diagnosis of relapsing form of multiple sclerosis to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, **AND**
    b. Patient had a previous trial of TWO agents indicated for the treatment of MS (i.e. Aubagio, Avonex, Gilenya, Plegridy, Rebif, Tecfidera, or glatiramer), **AND**
    c. The patient is 18 years of age or older, **AND**
    d. Hepatitis B virus and quantitative serum immunoglobulin screening are required before the first dose. If no documentation of HBV or immunoglobulin test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    e. Prescribed by or in consultation with, a physician who specializes in the treatment of MS and/or a neurologist

Reauthorization:

1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:

1. Contraindicated in patients with active hepatitis B infection
2. Concurrent use with other Disease-Modifying Agents used for MS
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Ocrevus prescribing information

Version History

Last Reviewed Date	Updates / Revisions
5/24/17	Addition to part B criteria with effective date 1/1/18
8/20/18	None
9/24/19	Update to FDA Approved Indication and required medical information
7/28/20	Update to required medical information
2/17/21	Addition of reauthorization criteria
8/10/21	Update to required medical information and exclusion criteria
8/23/22	None

ONPATTRO (patisiran) J0222 – *effective 9/1/2022*

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage** for **6 months (Initial), 12 months (reauthorization).** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy**.**

FDA Approved Indication(s):
Onpattro (patisiran) contains a transthyretin-directed small interfering RNA and is indicated for the treatment of the polyneuropathy of hereditary transthyretin-mediated amyloidosis in adults.

Required Medical Information:

1. A diagnosis of hereditary transthyretin-mediated amyloidosis (hATTR) with polyneuropathy and meet **ALL** the following criteria:
   - The patient is 18 years of age or older
   - Therapy is prescribed by or in consultation with a neurologist, cardiologist, physician at an amyloidosis treatment center, or medical geneticist
   - The patient has familial amyloid polyneuropathy (FAP) Stage 1 or 2 OR up to polyneuropathy disability (PND) Stage 3b polyneuropathy
   - The patient's diagnosis is confirmed by ONE of the following:
     - Biopsy of tissue/organ to confirm amyloid presence AND chemical typing to confirm presence of TTR protein
     - DNA genetic sequencing to confirm hATTR mutation

Reauthorization:

1. A diagnosis of hereditary TTR amyloidosis (hATTR) with polyneuropathy **AND**
2. The patient has not progressed to familial amyloid polyneuropathy (FAP) Stage 3 OR polyneuropathy disability (PND) Stage 4 polyneuropathy as evidenced by functional decline (e.g., wheelchair-bound, bedridden)

Exclusion Criteria:

Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Onpattro package insert

Version History:

Last Reviewed Date	Updates / Revisions
03/23/2022	Addition to part B criteria with effective date 9/1/2022
8/23/22	Update to initial and reauthorization criteria

ORENCIA (abatacept) J0129

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Orencia (abatacept) for **12 months (initial - aGVHD), 6 months (initial - all other dx), and one year (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Orencia is a selective T-cell costimulation modulator indicated for: 1. Moderately to severely active rheumatoid arthritis (RA) in adults. Orencia may be used as monotherapy or concomitantly with disease-modifying antirheumatic drugs (DMARDs) other than JAK inhibitors or bDMARDs (e.g., TNF antagonists). 2. Moderately to severely active polyarticular juvenile idiopathic arthritis in pediatric patients 6 years of age or older. Orencia may be used as monotherapy or in combination with methotrexate. 3. Psoriatic arthritis (PsA) in adults. Orencia may be used with or without non-biologic DMARDs. 4. The prophylaxis of acute graft versus host disease (aGVHD), in combination with a calcineurin inhibitor and methotrexate, in adults and pediatric patients 2 years of age and older undergoing hematopoietic stem cell transplantation (HSCT) from a matched or 1 allele-mismatched unrelated donor.

Required Medical Information:

1. **Rheumatoid arthritis in adults**
    a. Documentation of moderately to severely active RA, **AND**
    b. Documentation of trial of at least one conventional synthetic DMARD for at least 3 months (e.g. methotrexate, leflunomide, hydroxychloroquine, sulfasalazine, minocycline, gold) or documented medical reason for not taking. Patients who have already had a 3 month trial of a biologic for RA are not required to try a conventional synthetic DMARD. **AND**
    c. Patient has been tested for latent TB and screened for HBV prior to initiating Orencia. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    d. Patient is not receiving concurrent treatment with a biologic DMARD (e.g. Humira, Enbrel, Remicade, Actemra, Simponi) or targeted synthetic DMARD (e.g. Xeljanz, Olumiant), **AND**
    e. Patient is 18 years of age or older, **AND**
    f. Prescribed by or given in consultation with a rheumatologist

2. **Juvenile idiopathic arthritis**
    a. Documentation of moderately to severely active juvenile idiopathic arthritis, **AND**

b. Patient has been tested for latent TB and screened for HBV prior to initiating Orencia. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
c. Patient is not receiving concurrent treatment with a biologic DMARD (e.g. Humira, Enbrel, Remicade, Actemra, Simponi) or targeted synthetic DMARD (e.g. Xeljanz, Olumiant), **AND**
d. Patient is 6 years of age or older, **AND**
e. Prescribed by or given in consultation with a rheumatologist, **AND**
f. Patient has tried one other agent (e.g. NSAIDs, corticosteroids, methotrexate, leflunomide, sulfasalazine, Humira, Enbrel) for this condition, **OR**
g. Patient will be starting on Orencia concurrently with methotrexate, sulfasalazine or leflunomide or the patient has an absolute contraindication to methotrexate, sulfasalazine or leflunomide, **OR**
h. Patient has aggressive disease as determined by the prescribing physician

3. Psoriatic arthritis
a. Documented diagnosis of psoriatic arthritis, **AND**
b. Patient is 18 years of age or older, **AND**
c. Patient has been tested for latent TB and screened for HBV prior to initiating Orencia. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
d. Patient is not receiving concurrent treatment with a biologic DMARD (e.g. Humira, Enbrel, Remicade, Actemra, Simponi) or targeted synthetic DMARD (e.g. Xeljanz, Olumiant), **AND**
e. Prescribed by, or given in consultation with a rheumatologist or dermatologist

4. Prophylaxis of Acute graft versus host disease (aGVHD)
a. The patient is 2 years of age or older, **AND**
b. The patient is undergoing hematopoietic stem cell transplantation (HSCT) from a matched OR one allele-mismatched unrelated-donor, **AND**
c. Orencia will be used in combination with a calcineurin inhibitor (e.g., cyclosporine, tacrolimus, pimecrolimus) and methotrexate

Reauthorization Criteria:

1. Diagnosis of moderate to severe rheumatoid arthritis, psoriatic arthritis, or moderate to severe polyarticular juvenile idiopathic arthritis, **AND** physician attestation that the patient continues to benefit from the medication.
2. For the diagnosis of acute graft versus host disease (aGVHD), please refer to the Initial Criteria section.

Exclusion Criteria:

1. Orencia should not be given concomitantly with a biologic DMARD or targeted synthetic DMARD.
2. Live vaccines should not be given concurrently or within 3 months after discontinuation of Orencia.
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Orencia prescribing information

Version History

Last Reviewed Date	Updates / Revisions

9/13/16	None
8/22/17	Update to FDA indications; Update to required medical information
8/20/18	Update to required medical information
11/5/18	Addition of reauthorization criteria
1/1/19	Updates to authorization period, required information, and exclusion criteria to align with part D criteria
9/24/19	None
7/28/20	Update to required medical information
8/10/21	Update to FDA indications and required medical information
8/23/22	Addition of new FDA indication, required medical information, reauthorization criteria, update to approval time frame

RADICAVA (edaravone) J1301 – *effective 9/1/2022*

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage for 6 Months (initial and reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
RADICAVA is indicated for the treatment of amyotrophic lateral sclerosis (ALS).

Required Medical Information:

1. Amyotrophic lateral sclerosis (ALS)
    a. According to the prescribing physician, the patient has a definite or probable diagnosis of ALS, **AND**
    b. Patient has a score of two points or more on each item of the ALS Functional Rating Scale – Revised (ALSFRS-R) [ie, has retained most or all activities of daily living], **AND**
    c. Patient has a FVC greater than or equal to 80% (ie, normal respiratory function), **AND**
    d. Patient has been diagnosed with ALS for less than or equal to 2 years, **AND**
    e. Therapy is prescribed by or in consultation with a neurologist, a neuromuscular disease specialist, or a physician specializing in the treatment of ALS

Reauthorization:

1. Patient has diagnosis of amyotrophic lateral sclerosis (ALS) AND meets ALL of the following:
    a. According to the prescribing physician, the patient continues to benefit from therapy
    b. The patient is not requiring invasive ventilation

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. History of hypersensitivity to edaravone or any of the inactive ingredients in RADICAVA.

References:

Radicava prescribing information

Version History

Last Reviewed Date	Updates / Revisions
3/23/22	Addition to part B criteria with effective date 9/1/2022
8/23/22	None

REMICADE (infliximab) J1745, INFLECTRA (infliximab-dyyb) Q5103, RENFLEXIS (infliximab-abda) Q5104, AVSOLA (infliximab-axxq) Q5121

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for **3 months (initial) and one year (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy. Physician reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

FDA Approved Indication(s):
Remicade (Infliximab) is a tumor necrosis factor (TNF) blocker indicated for: 1. Crohn's disease - Reducing signs and symptoms and inducing and maintaining clinical remission in adult patients and pediatric patients 6 years of age and older with moderately to severely active Crohn's disease who have had an inadequate response to conventional therapy. 2. Fistulizing Crohn's disease- Reducing the number of draining enterocutaneous and rectovaginal fistulas and maintaining fistula closure in adult patients with fistulizing Crohn's disease. 3. Ulcerative colitis- Reducing signs and symptoms, inducing and maintaining clinical remission and mucosal healing, and eliminating corticosteroid use in adult patients and pediatric patients 6 years of age and older with moderately to severely active ulcerative colitis who have had an inadequate response to conventional therapy. 4. Rheumatoid arthritis- In combination with methotrexate, for reducing signs and symptoms, inhibiting the progression of structural damage, and improving physical function in patients with moderately to severely active rheumatoid arthritis. 5. Psoriatic Arthritis- Reducing signs and symptoms of active arthritis, inhibiting the progression of structural damage, and improving physical function in patients with psoriatic arthritis. 6. Plaque Psoriasis- Treatment of adult patients with chronic severe (i.e., extensive and/or disabling) plaque psoriasis who are candidates for systemic therapy and when other systemic therapies are medically less appropriate. 7.Ankylosing Spondylitis- Reducing signs and symptoms in patients with active ankylosing spondylitis

Required Medical Information:

1. **Crohn's disease**
    a. Documented diagnosis of moderately to severely active Crohn's disease, **AND**
    b. Prescribed by or in consultation with a gastroenterologist, **AND**
    c. Patient is at least 6 years old, **AND**
    d. Patient has tried corticosteroid (CS) or if CSs contraindicated or if currently on CS or if the patient has tried one other agent for CD (e.g. azathioprine, mercaptopurine, methotrexate, mesalamine) or the patient has had ileocolonic resection, **AND**
    e. Patient has been tested for latent TB and screened for HBV prior to initiating therapy. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval).

2. **Fistulizing Crohn's Disease**
    a. Documented diagnosis of fistulizing Crohn's disease, **AND**
    b. Prescribed by or in consultation with a gastroenterologist, **AND**
    c. Patient is at least 18 years old, **AND**
    d. Patient has been tested for latent TB and screened for HBV prior to initiating therapy. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval).

3. Ulcerative Colitis
    a. Documented diagnosis of moderately to severely active ulcerative colitis, **AND**
    b. Prescribed by or in consultation with a gastroenterologist, **AND**
    c. Patient is at least 6 years old, **AND**
    d. Tried one systemic agent (e.g. corticosteroids, azathioprine, mercaptopurine, methotrexate, mesalamine) or was intolerant to one of these agents OR the patient has pouchitis AND has tried therapy with an antibiotic, probiotic, corticosteroid enema, or mesalamine enema. (Note-a previous trial of a biologic also counts as a trial of one systemic agent for UC), **AND**
    e. Patient has been tested for latent TB and screened for HBV prior to initiating therapy. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval).

4. Rheumatoid Arthritis
    a. Documented diagnosis of moderately to severely active rheumatoid arthritis, **AND**
    b. Prescribed by or given in consultation with a rheumatologist, **AND**
    c. Infliximab is used in combination with methotrexate, **AND**
    d. Patient has tried ONE conventional synthetic DMARD (e.g. methotrexate, leflunomide, sulfasalazine, azathioprine, hydroxychloroquine) for at least 3 months (note: patients who have already had a 3-month trial of a biologic for RA are not required to step back and try a conventional synthetic DMARD), **AND**
    e. Patient has been tested for latent TB and screened for HBV prior to initiating therapy. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval).

5. **Psoriatic Arthritis**
    a. Documented diagnosis of active psoriatic arthritis, **AND**
    b. Prescribed by or given in consultation with a rheumatologist or dermatologist, **AND**
    c. Patient has been tested for latent TB and screened for HBV prior to initiating therapy. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval)

6. **Plaque Psoriasis**
    a. Prescribed by or given in consultation with a dermatologist, **AND**
    b. Patient is 18 years of age or older and has psoriasis covering 3% or more of body surface area (BSA) OR psoriatic lesions affecting the hands, feet, genital area, or face, **AND**
    c. Patient is a candidate for systemic therapy**, AND**
    d. Patient has tried at least at least one traditional systemic agent (e.g. methotrexate, corticosteroids, cyclosporine) for psoriasis for at least 3 months, unless intolerant or the patient has a contraindication to methotrexate (MTX), as determined by the prescriber, **AND**
    e. Patient has been tested for latent TB and screened for HBV prior to initiating therapy. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval).

7. **Ankylosing Spondylitis**
    a. Documented diagnosis of active ankylosing spondylitis, **AND**
    b. Prescribed by or given in consultation with a rheumatologist, **AND**
    c. Patient has been tested for latent TB and screened for HBV prior to initiating therapy. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval).

Reauthorization Criteria:

1. For all FDA-approved indications: patient has had a response, as determined by the prescriber.

Exclusion Criteria:

1. Treatment with infliximab should not be initiated in patients with an active infection, including clinically important localized infections.
2. Live vaccines or therapeutic infectious agents should not be given with infliximab
3. Concurrent use with Biologic DMARD or Targeted Synthetic DMARD
4. Infliximab at doses >5mg/kg should not be administered to patients with moderate to severe heart failure (NYHA Functional Class III/IV).
5. Hypersensitivity to inactive components of the product or murine proteins
6. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

Clinical Note:

- Recommended to test for Hepatitis B virus (HBV) infection prior to initiation of Remicade. Monitor HBV carriers throughout and after Remicade therapy. Remicade should be discontinued if reactivation occurs.

References:

1. Remicade prescribing information
2. Coates LC, Kavanaugh A, Mease PJ, et al. Group for Research and Assessment of Psoriasis and Psoriatic Arthritis 2015 Treatment Recommendations for Psoriatic Arthritis. Arthritis & Rheumatology. 2016; 68:1060-1071.
3. Gottlieb A, Korman NJ, Gordon KB, Feldman SR, Lebwohl M, Koo JY, et al. Guidelines of care for the management of psoriasis and psoriatic arthritis: Section 2. Psoriatic arthritis: overview and guidelines of care for treatment with an emphasis on the biologics. J Am Acad Dermatol. 2008 May;58(5):851-64.
4. Menter A, Korman NJ, Elmets CA, et al. Guidelines of care for the management of psoriasis and psoriatic arthritis. J Am Acad Dermatol. 2011 July 65:137-13.

Version History

Last Reviewed Date	Updates / Revisions
9/13/16	Addition of clinical note; Addition to exclusion criteria
8/22/17	Update to required information for psoriatic arthritis indication; Update to required information for plaque psoriasis indication; Addition of biosimilar Renflexis (infliximab-abda) effective 1/1/18
6/7/18	Updated Q codes for Renflexis and Inflectra
9/24/19	Addition of biosimilar, Ixifi; Addition of reauthorization criteria; Update to authorization period
7/28/20	Removal of Ixifi; Updates to required information; Update to reauthorization criteria
8/10/21	Addition of biosimilar, Avsola; Update to initial approval time frame, required medical information, reauthorization criteria, and exclusion criteria
8/23/22	Update to required medical information for Psoriatic arthritis and Plaque psoriasis

REMODULIN (treprostinil) J3285

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Remodulin (treprostinil) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Remodulin (treprostinil) is a prostacyclin vasodilator 1. For the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to diminish symptoms associated with exercise. Studies establishing effectiveness included patients with NYHA Functional Class II-IV symptoms and etiologies of idiopathic or heritable PAH (58%), PAH associated with congenital systemic-to-pulmonary shunts (23%), or PAH associated with connective tissue diseases (19%) 2. Patients who require transition from epoprostenol, to reduce the rate of clinical deterioration. The risks and benefits of each drug should be carefully considered prior to transition.

Required Medical Information:

1. Documented diagnosis of PAH WHO Group 1 AND one of the following:
    i. Documented diagnosis of primary arterial pulmonary hypertension NYHA functional Class III to IV, **OR**
    ii. Documented diagnosis of primary arterial pulmonary hypertension NYHA functional Class II and must have tried or is currently receiving one oral agent for PAH or patient has tried one inhaled or parenteral prostacyclin product for PAH **OR**
    iii. Patient has idiopathic PAH and meets one of the following:
        - Had an acute response to vasodilator testing that occurred during the right heart cath (defined as decrease in mPAP of at least 10 mm Hg to an absolute mPAP of less than 40 mm Hg without a decrease in cardiac output) AND has tried an oral CCB or
        - Pt did not have an acute response to vasodilator testing or
        - Cannot undergo vasodilator test or cannot take CCB due to extreme right HF (e.g. hypotension, cardiac index less than 1.5, or right atrial pressure greater than 20, or
        - Has tried a CCB without vasodilator testing, **OR**
    iv. Documented diagnosis of pulmonary hypertension secondary to one of the following conditions: connective tissue disease, thromboembolic disease of the pulmonary arteries, HIV infection, cirrhosis, diet drugs, congenital left to right shunts, **AND**
2. Documented confirmatory pulmonary arterial hypertension diagnosis based on right heart catheterization based on the following parameters:
    i. Mean pulmonary arterial pressure (PAP) of > 20 mmHg
    ii. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    iii. Pulmonary vascular resistance (PVR) > 3 Wood units; **AND**

3. Pulmonary hypertension has progressed despite maximal medical and/or surgical treatment, **AND**
4. Significant symptoms from pulmonary hypertension are present (i.e. severe dyspnea on exertion, fatigue, angina, or syncope), **AND**
5. Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out, **AND**
6. Patient must be 18 years old or older, **AND**
7. Prescribed by or in consultation with a pulmonologist OR a cardiologist

Reauthorization Criteria:

1. Diagnosis of pulmonary arterial hypertension (PAH; WHO Group 1) and meets one of the following:
    a. Patient has shown improvement from baseline in the 6-minute walk distance, **OR**
    b. Patient has remained stable from baseline in the 6-minute walk distance **AND** patient's World Health Organization (WHO) functional class symptoms remained stable or have improved.

Exclusion Criteria:

1. Coverage excluded if pulmonary hypertension is secondary to pulmonary venous hypertension (i.e. left sided atrial or ventricular disease, left sided valvular heart disease) or disorders of the respiratory system (i.e. COPD, interstitial lung disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders)
2. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Remodulin prescribing information

Version History

Last Reviewed Date	Updates / Revisions
9/13/16	None
8/22/17	Update to required medical information and exclusion criteria
8/20/18	Update to required information
9/24/19	Addition of reauthorization criteria
7/28/20	Update to required medical information
8/10/21	Update to required medical information and reauthorization criteria
8/23/22	Update to required medical information

REVATIO INJECTION (sildenafil) J3490

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Revatio injection (sildenafil) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
REVATIO is a phosphodiesterase-5 (PDE-5) inhibitor indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group I) in adults to improve exercise ability and delay clinical worsening. Studies establishing effectiveness were short-term (12 to 16 weeks) and included predominately patients with NYHA Functional Class II–III symptoms. Etiologies were idiopathic (71%) or associated with connective tissue disease (25%). (1) Limitation of Use: Adding sildenafil to bosentan therapy does not result in any beneficial effect on exercise capacity

Required Medical Information:

1. **Pulmonary arterial hypertension**
    a. Documented diagnosis of pulmonary arterial hypertension (WHO Group 1) based on right heart catheterization with the following parameters:
        i. Mean pulmonary artery pressure (PAP) of > 20 mmHg
        ii. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
        iii. Pulmonary vascular resistance (PVR) ≥ 3 Wood units; **AND**
    b. Patient has NYHA-WHO Functional Class II to IV symptoms, **AND**
    c. Documented medical reason for not taking oral sildenafil, **AND**
    d. Patient is NOT taking oral erectile dysfunction agents or any organic nitrates in any form, **AND**
    e. Patient is not taking guanylate cyclase (GC) stimulators, such as Adempas, **AND**
    f. Patient must be 18 years old or older, **AND**
    g. Prescribed by or given in consultation with a pulmonologist or a cardiologist

Reauthorization Criteria:

1. The patient has a diagnosis of pulmonary arterial hypertension (PAH) (World Health Organization Group 1), **AND**
2. Has shown improvement from baseline in the 6-minute walk distance, **OR**
3. Has remained stable from baseline in the 6-minute walk distance with a stable or improved WHO functional class.

Exclusion Criteria:

1. Revatio is contraindicated in patients with a known hypersensitivity to sildenafil or any component of the tablet or injection.
2. Revatio is contraindicated in patients taking organic nitrates in any form, either regularly or intermittently.
3. Revatio is contraindicated in patients taking riociguat (Adempas)
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Revatio prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	Addition to exclusion criteria
8/22/17	Update to required information
8/20/18	Update to required information
9/24/19	Addition of reauthorization criteria
7/28/20	None
11/10/21	None
11/2/22	Update to required information

SAPHNELO (Anifrolumab-Fnia) J0491

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage for 6 months (Initial) and 12 months (reauthorization).** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Saphnelo (Anifrolumab-Fnia) - a type I interferon (IFN) receptor antagonist indicated for the treatment of adult patients with moderate to severe systemic lupus erythematosus (SLE), who are receiving standard therapy. Limitations of Use: The efficacy of SAPHNELO has not been evaluated in patients with severe active lupus nephritis or severe active central nervous system lupus. Use of SAPHNELO is not recommended in these situations.

Required Medical Information:

1. Diagnosis of moderate to severe systemic lupus erythematosus (SLE) and meet **ALL** of the following criteria:
    a. The patient is 18 years of age or older
    b. Therapy is prescribed by or given in consultation with a rheumatologist
    c. The patient is receiving standard SLE therapy (e.g., oral corticosteroids, antimalarials, or immunosuppressants)

Reauthorization:

3. Diagnosis moderate to severe systemic lupus erythematosus (SLE) **AND** the patient has had clinical improvement while on Saphnelo.

Exclusion Criteria:

4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Saphnelo package insert

Version History:

Last Reviewed Date	Updates / Revisions
11/10/2021	Addition to part B criteria with effective date 1/1/2022
4/12/2022	Updated J Code
11/2/22	None

SIMPONI ARIA (golimumab) J1602

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Simponi Aria (golimumab) for **6 months (initial) and one year (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
SIMPONI ARIA (golimumab) is a tumor necrosis factor (TNF) blocker indicated for the treatment of: 1. Adult patients with moderately to severely active Rheumatoid Arthritis (RA) in combination with methotrexate 2. Active Psoriatic Arthritis (PsA) in patients 2 years of age and older 3. Adult patients with active Ankylosing Spondylitis (AS) 4. Active polyarticular Juvenile Idiopathic Arthritis (pJIA) in patients 2 years of age and older

Required Medical Information:

1. **Rheumatoid arthritis**
    a. Documentation of moderately to severely active RA**, AND**
    b. Being used in combination with methotrexate (unless contraindicated), **AND**
    c. Previous trial of or contraindication to at least one DMARD (disease-modifying antirheumatic drug), such as methotrexate, leflunomide, hydroxychloroquine, or sulfasalazine **AND**
    d. Patient has been tested for latent TB and screened for HBV prior to initiating Simponi Aria. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    e. Patient is 18 years of age or older, **AND**
    f. Prescribed by or given in consultation with a rheumatologist

2. **Psoriatic arthritis**
    a. Diagnosis of psoriatic arthritis, **AND**
    b. Patient has been tested for latent TB and screened for HBV prior to initiating Simponi Aria. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
    c. Prescribed by or given in consultation with a rheumatologist or dermatologist, **AND**
    d. Patient is 2 to 17 years of age, **OR**
    e. Patient is 18 years of age or older AND had previous trial of or contraindication to at least one DMARD (disease-modifying antirheumatic drug), such as methotrexate, leflunomide, hydroxychloroquine, or sulfasalazine

3. **Ankylosing spondylitis**
    a. Diagnosis of moderate to severe ankylosing spondylitis, **AND**

b. Patient has been tested for latent TB and screened for HBV prior to initiating Simponi Aria. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
c. Patient is 18 years of age or older, **AND**
d. Prescribed by or given in consultation with a rheumatologist, **AND**
e. Previous trial of or contraindication to NSAIDs (ie: diclofenac, Ibuprofen, naproxen, celecoxib, meloxicam, etc.)

4. **Polyarticular juvenile idiopathic arthritis**
   a. Diagnosis of polyarticular juvenile idiopathic arthritis, **AND**
   b. Patient is 2 years of age or older, **AND**
   c. Therapy is prescribed by or given in consultation with a rheumatologist, **AND**
   d. Patient has been tested for latent TB and screened for HBV prior to initiating Simponi Aria. If no documentation of TB and HBV test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
   e. Patient has tried one other medication for this condition OR patient has aggressive disease, as determined by the prescriber

Reauthorization:

1. Patient has diagnosis of moderate to severe rheumatoid arthritis, psoriatic arthritis, ankylosing spondylitis, or polyarticular juvenile idiopathic arthritis **AND**
2. The patient continues to benefit from the medication

Exclusion Criteria:

1. Concurrent use with a Biologic DMARD or Targeted Synthetic DMARD.
2. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

Clinical Note:

1. Monitoring of TB is recommended periodically during therapy, including patients who tested negative prior to treatment initiation or who previously received treatment for latent or active TB.
2. Recommended to evaluate for Hepatitis B virus (HBV) infection and carrier status prior to therapy. Additional monitor of labs and clinical signs of HBV in carriers is recommended throughout therapy and for several months following discontinuation.

References:

Simponi Aria prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	Addition of clinical notes regarding TB and HBV
8/22/17	None

12/18/17	Addition to FDA indications
8/20/18	Update to required information; Update to authorization period – effective 1/1/19
9/24/19	None
7/28/20	Update to required information
11/10/21	Update and addition to required information and FDA indications; Update to reauthorization criteria, exclusion criteria, and initial approval time frame
11/2/22	Update to required information and initial approval timeframe

SOLIRIS (eculizumab) J1300

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Soliris (eculizumab) for **6 months (initial - myasthenia gravis and paroxysmal nocturnal hemoglobinuria), one year (initial - all other indications) and one year (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
SOLIRIS (eculizumab) is a complement inhibitor indicated for: 1. The treatment of patients with paroxysmal nocturnal hemoglobinuria (PNH) to reduce hemolysis 2. The treatment of patients with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy 3. The treatment of adult patients with generalized Myasthenia Gravis (gMG) who are anti-acetylcholine receptor (AchR) antibody positive 4. The treatment of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive Limitation of Use: Soliris is not indicated for the treatment of patients with Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS).

Required Medical Information:

1. **Paroxysmal nocturnal hemoglobinuria**
    a. Documented diagnosis of Paroxysmal nocturnal hemoglobinuria, **AND**
    b. Documentation provided that the patient has been vaccinated with meningococcal vaccine at least two weeks prior to initiation of Soliris therapy (if not previously vaccinated), **AND**
    c. Patient must be 18 years old or older, **AND**
    d. The requested medication is prescribed by or in consultation with a hematologist, **AND**
    e. The patient is not using concurrent complement inhibitor therapy (e.g., Ultomiris, Empaveli), **AND**
    f. The patient has confirmed PNH as demonstrated by ALL of the following via flow cytometry:
        1 At least 2 different GPI-protein deficiencies (e.g., CD55, CD59) on at least 2 cell lineages (e.g., erythrocytes, granulocytes)
        2 PNH granulocyte clone size of 10% or higher, **AND**

    g. The patient meets one of the following:
        1 The patient is transitioning from an alternative complement inhibitor therapy (e.g., Ultomiris), **OR**
        2 The patient has evidence of intravascular hemolysis (e.g., lactate dehydrogenase [LDH] level greater than or equal to 1.5 X ULN, hemoglobinuria) AND presence of at least one PNH-related sign or symptom (e.g., history of blood transfusion due to PNH, symptoms of anemia, history of major adverse vascular event from thromboembolism)

2. **Atypical hemolytic uremic syndrome**
    a. Documented diagnosis of atypical hemolytic uremic syndrome, **AND**
    b. Documentation provided that the patient has been vaccinated with meningococcal vaccine at least two weeks prior to initiation of Soliris therapy (if not previously vaccinated)

3. **Myasthenia Gravis**
    a. Documented diagnosis of anti-acetylcholine receptor (AchR) antibody positive generalized myasthenia gravis (gMG), **AND**
    b. Documentation provided that the patient has been vaccinated with meningococcal vaccine at least two weeks prior to initiation of Soliris therapy (if not previously vaccinated), **AND**
    c. Patient must be 18 years old or older, **AND**
    d. The requested medication is prescribed by or in consultation with a neurologist, **AND**
    e. Patient is Myasthenia Gravis Foundation of America class II, III, or IV

4. **Neuromyelitis optica spectrum disorder (NMOSD)**
    a. Documented diagnosis of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive , **AND**
    b. Documentation provided that the patient has been vaccinated with meningococcal vaccine at least two weeks prior to initiation of Soliris therapy (if not previously vaccinated), **AND**
    c. Patient must be 18 years old or older **AND**
    d. The patient has at least ONE of the following core clinical characteristic: Optic neuritis, Acute myelitis, Area postrema syndrome, Acute brainstem syndrome, Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions, Symptomatic cerebral syndrome with NMOSD-typical brain lesions, **AND**
    e. The patient will NOT use rituximab, inebilizumab, or satrilizumab concurrently with Soliris, **AND**
    f. The requested medication is prescribed by or in consultation with a neurologist

Reauthorization:

1. Documented diagnosis of Atypical hemolytic uremic syndrome, **OR**
2. Documented diagnosis of Paroxysmal nocturnal hemoglobinuria, Myasthenia gravis, or Neuromyelitis optica spectrum disorder (NMOSD) AND physician attestation of clinical benefit compared to baseline.

Exclusion Criteria:

1. Soliris is contraindicated in patients with unresolved serious *Neisseria meningitidis* and in patients who are not currently vaccinated against *Neisseria meningitidis*.
2. Soliris is not indicated for the treatment of patients with Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS).
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Soliris prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	None

12/18/17	Addition to FDA indication; Addition to exclusion criteria
11/5/18	Addition to required information
12/10/19	Addition to FDA approved indication
11/10/20	Addition to required information; Addition of reauthorization criteria
2/17/21	Update to required information for PNH
11/10/21	Update to required information for AHUS and NMOSD
11/2/22	Update to required information for PNH

STELARA (ustekinumab) J3358 (IV)

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Stelara (ustekinumab) for **2 months (IV induction dose only)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
STELARA (ustekinumab) is a human interleukin-12 and -23 antagonist FDA approved for the treatment of 1. Adult and adolescent patients (6 years or older) with moderate to severe plaque psoriasis who are candidates for phototherapy or systemic therapy **(subQ)** 2. Adult and adolescent patients (6 years or older) with active psoriatic arthritis **(subQ)** 3. Adult patients with moderately to severely active Crohn's disease **(IV induction followed by subQ)** 4. Adult patients with moderately to severely active ulcerative colitis **(IV induction followed by subQ)**

Initially review for B vs D coverage

Per the Medicare Benefit Policy manual chapter 15:

50.2 - Determining Self-Administration of Drug or Biological (Rev. 157, Issued: 06-08-12, Effective: 07-01-12, Implementation: 07-02-12) The Medicare program provides limited benefits for outpatient prescription drugs. The program covers drugs that are furnished "incident to" a physician's service provided that the drugs are not usually self-administered by the patients who take them.

50.5 - Self-Administered Drugs and Biologicals (Rev. 1, 10-01-03) B3-2049.5 Medicare Part B does not cover drugs that are usually self-administered by the patient unless the statute provides for such coverage. The statute explicitly provides coverage, for blood clotting factors, drugs used in immunosuppressive therapy, erythropoietin for dialysis patients, certain oral anti-cancer drugs and anti-emetics used in certain situations.

Required Medical Information:

1. **Plaque psoriasis – Covered under Part D**

2. **Active psoriatic arthritis – Covered under Part D**

3. **Crohn's disease – Induction dose covered under Part B**
    a. Documented diagnosis of moderately to severely active Crohn's disease and meets one of the following:

i. Patient has tried or is currently taking corticosteroids, or corticosteroids are contraindicated, **OR**
ii. Patient has tried one other agent for CD (eg, azathioprine, 6-MP, MTX, certolizumab, vedolizumab, adalimumab, infliximab), **AND**

b. Patient must be 18 years old or older, **AND**
c. Patient has been tested for latent TB prior to initiating Stelara therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval), **AND**
d. Prescriber must be a gastroenterologist, **AND**
e. The patient has one of the following:
i. Patient has enterocutaneous (perianal or abdominal) or rectovaginal fistulas **OR**
ii. patient had ileocolonic resection (to reduce the chance of Crohn's disease recurrence)

4. Ulcerative colitis – Induction dose covered under part B

a. The patient is 18 years of age or older, **AND**
b. Therapy is prescribed by or given in consultation with a gastroenterologist, **AND**
c. The patient had a previous trial of or contraindication to at least ONE conventional therapy such as corticosteroids (i.e., budesonide, methylprednisolone), azathioprine, mercaptopurine, methotrexate, or mesalamine, **AND**
d. Patient has been tested for latent TB prior to initiating Stelara therapy. If no documentation of TB test, will be recommended by health plan pharmacist (but not required prior to approval)

Exclusion Criteria:

1. Stelara should not be used concomitantly with a biologic DMARD or targeted synthetic DMARD
2. Stelara should not be administered during any clinically important active infection. If a serious infection develops, stop Stelara until the infection resolves.
3. Stelara may increase risk of malignancy. The safety of Stelara in patients with a history of or a known malignancy has not been evaluated
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Stelara prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	Addition to FDA approved indications; Addition to required medical information for additional diagnoses
8/22/17	Removal of J code; Addition of Q code
12/18/17	Update to FDA indications; Addition of B vs D coverage information
6/7/18	Update to authorization period; Update to IV J code; Removal of required information for plaque psoriasis and psoriatic arthritis – covered under part D

8/20/18	Update to required information
11/5/18	None
1/1/19	Update to exclusion criteria to align with part D criteria
12/10/19	Update to FDA indications
11/10/20	Update to FDA indication and Addition of required information for UC
11/10/21	Update to FDA indication and required information for CD
11/2/22	Addition to required information for CD

TYSABRI (natalizumab) J2323

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Tysabri (natalizumab) for **one year** for the treatment of multiple sclerosis, and for **three months (initial) and one year (reauthorization)** for the treatment of Crohn's disease. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
TYSABRI (natalizumab) is an integrin receptor antagonist indicated for treatment of: 1. **Multiple Sclerosis**- As monotherapy for the treatment of relapsing forms of multiple sclerosis, to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, in adults. TYSABRI increases the risk of PML (progressive multifocal leukoencephalopathy). When initiating and continuing treatment with TYSABRI, physicians should consider whether the expected benefit of TYSABRI is sufficient to offset this risk. 2. **Crohn's Disease**- For Inducing and maintaining clinical response and remission in adult patients with moderately to severely active Crohn's disease with evidence of inflammation who have had an inadequate response to, or are unable to tolerate, conventional CD therapies and inhibitors of TNF-α Important Limitations: In Crohn's Disease, TYSABRI should not be used in combination with immunosuppressants or inhibitors of TNF-α. Because of the risk of PML, natalizumab is available only through a special restricted distribution program called the TOUCH prescribing program. Under the TOUCH prescribing program, only prescribers, infusion centers, and pharmacies associated with infusion centers registered with the program are able to prescribe, distribute, or infuse the product. In addition, natalizumab must be administered only to patients who are enrolled in and meet all the conditions of the TOUCH prescribing program.

Required Medical Information:

Indication	Initial authorization: Multiple sclerosis: 1 year Crohn's disease: 3 months	Re-authorization: Multiple sclerosis and Crohn's disease: 1 year
Multiple sclerosis	1. Documentation of relapsing form of Multiple Sclerosis to include clinically isolated syndrome, relapsing-remitting disease, and active secondary progressive disease, **AND** 2. Tysabri is being used as monotherapy, **AND** 3. Patient must be 18 years or older, **AND** 4. Prescribed by, or in consultation with, a neurologist or physician who specializes in the treatment of MS, **AND** 5. Patient has had an inadequate response to, or is unable to tolerate, one disease modifying agent used for MS (eg, interferon beta-1a (Avonex, Rebif), interferon beta-1b (Betaseron, Extavia), glatiramer acetate (Copaxone/Glatopa), Plegridy, fingolimod (Gilenya), Tecfidera, Lemtrada, daclizumab (Zinbryta), Aubagio), **OR** 6. The patient has highly active or aggressive disease according to the prescribing physician by meeting one of the following: A. The patient has demonstrated rapidly-advancing deterioration(s) in physical functioning (e.g., loss of mobility/or lower levels of ambulation, severe changes in strength or coordination), OR B. Disabling relapse(s) with suboptimal response to systemic corticosteroids, OR C. Magnetic resonance imaging (MRI) findings suggest highly-active or aggressive multiple sclerosis (e.g., new, enlarging, or a high burden of T2 lesions or gadolinium lesions), OR	Documented improvement in symptoms

	D. Manifestation of multiple sclerosis-related cognitive impairment	
Crohn's disease	1. Patient has moderately to severely active CD with evidence of inflammation (e.g., elevated C-reactive protein), **AND** 2. Patient has tried two of the following agents for CD for at least 2 months each - adalimumab, certolizumab pegol, infliximab, vedolizumab, ustekinzumab, **OR** Patient has had an inadequate response or was intolerant to these agents. **AND** 3. Patient is not on concurrent immunosuppressant therapy or TNF-alpha inhibitor (patient can be on an aminosalicylate), **AND** 4. Patient must be 18 years old or older, **AND** 5. Therapy initiated by or in consultation with a gastroenterologist	Patient has had a response to Tysabri, as determined by the prescribing physician.

Exclusion Criteria:

1. Tysabri is contraindicated in patients who have or have had PML, and who have had a hypersensitivity reaction to Tysabri.
2. Concurrent use of other disease-modifying agents used for MS. Concurrent use with immunosuppressants (eg, 6-mercaptopurine, azathioprine, cyclosporine, methotrexate) in Crohn's disease (CD) patients.
3. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Tysabri prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	None
12/18/17	Update to required information for Crohn's disease
11/5/18	Update to required information for Crohn's disease
12/10/19	Update to required information (to align with part D criteria)
11/10/20	Update to FDA approved indication for Multiple Sclerosis, Update to required information for Crohn's disease

11/10/21	Addition to exclusion criteria; Update to initial approval time frame for CD; Update to required information; Update to reauthorization criteria for CD
11/2/22	Update to initial criteria and approval timeframe for CD

TYVASO (treprostinil inhalation) J7686

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage for 6 months (initial PH-ILD) and 12 months (PAH and reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Tyvaso is a prostacyclin mimetic indicated for the treatment of: • Pulmonary arterial hypertension (PAH; WHO Group 1) to improve exercise ability. Studies establishing effectiveness predominately included patients with NYHA Functional Class III symptoms and etiologies of idiopathic or heritable PAH (56%) or PAH associated with connective tissue diseases (33%). • Pulmonary hypertension associated with interstitial lung disease (PH-ILD; WHO Group 3) to improve exercise ability. The study establishing effectiveness predominately included patients with etiologies of idiopathic interstitial pneumonia (IIP) (45%) inclusive of idiopathic pulmonary fibrosis (IPF), combined pulmonary fibrosis and emphysema (CPFE) (25%), and WHO Group 3 connective tissue disease (22%).

Required Medical Information:

1. **Pulmonary arterial hypertension (PAH)**
    a. Diagnosis of pulmonary arterial hypertension (PAH) World Health Organization (WHO) Group 1, AND
    b. Patient has primary pulmonary hypertension or pulmonary hypertension which is secondary to one of the following conditions: connective tissue disease, human immunodeficiency virus (HIV) infection, cirrhosis, anorexigens or congenital left to right shunts, AND
    c. Therapy is prescribed by or given in consultation with a cardiologist or pulmonologist, AND
    d. The patient has a confirmatory PAH diagnosis based on right heart catheterization with the following parameters:
        I. Pulmonary vascular resistance (PVR) $\geq$ 3 Wood units (WU)
        II. Mean pulmonary arterial pressure (mPAP) > 20 mmHg
        III. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg, AND
    e. The patient has New York Heart Association-World Health Organization (NYHA-WHO) Functional Class III to IV symptoms and meets ONE of the following:
        I. Patients with WHO Functional Class III symptoms must have had a trial of or contraindication to TWO of the following agents from different drug classes:
            1. An oral endothelin receptor antagonist (e.g., ambrisentan, bosentan, or macitentan)
            2. An oral phosphodiesterase-5 inhibitor (e.g., sildenafil or tadalafil)
            3. An oral cGMP inhibitor (e.g., riociguat), OR
        II. Patients with WHO Functional Class III symptoms with evidence of rapid progression or poor prognosis, OR WHO Functional Class IV symptoms must have had trial of or contraindication to ONE of the following:
            1. An IV/SQ prostacyclin (e.g., epoprostenol or treprostinil), AND
    f. The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition, AND

g. The beneficiary has significant symptoms from the pulmonary hypertension (i.e., severe dyspnea on exertion, and either fatigability, angina, or syncope), AND
h. Treatment with oral calcium channel blocking agents has been tried and failed, or has been considered and ruled out

2. Pulmonary Hypertension associated with interstitial lung disease (PH-ILD)

a. Diagnosis of pulmonary hypertension associated with interstitial lung disease (PH-ILD; WHO Group 3), AND
b. The presence of interstitial lung disease has been confirmed by a high-resolution CT scan of the chest, AND
c. The beneficiary has significant symptoms of pulmonary hypertension (e.g., dyspnea on exertion, fatigability), AND
d. Therapy is prescribed by or given in consultation with a cardiologist or pulmonologist, AND
e. The patient has a confirmatory PH diagnosis based on right heart catheterization with the following parameters:
    I. Pulmonary vascular resistance (PVR) of ≥ 3 Wood units (WU)
    II. Mean pulmonary arterial pressure (mPAP) of > 20 mmHg
    III. Pulmonary capillary wedge pressure (PCWP) of ≤ 15 mmHg

Reauthorization:

1. Diagnosis of pulmonary arterial hypertension (PAH; WHO Group 1) and meets one of the following:
    a. The patient has shown improvement from baseline in the 6-minute walk distance test, OR
    b. The patient has remained stable from baseline in the 6-minute walk distance test AND the patient's World Health Organization (WHO) functional class has improved or remained stable
2. Diagnosis of pulmonary hypertension associated with interstitial lung disease (PH-ILD; WHO Group 3) and meets one of the following:
    a. The patient has shown improvement from baseline in the 6-minute walk distance test, OR
    b. The patient has a stable 6-minute walk distance test

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. The pulmonary hypertension is not secondary to pulmonary venous hypertension (e.g., left sided atrial or ventricular disease, left sided valvular heart disease) or disorders of the respiratory system other than interstitial lung disease (e.g., chronic obstructive pulmonary disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders).

References:

Tyvaso prescribing information
World Symposium on Pulmonary Hypertension Guidelines

Version History

Last Reviewed Date	Updates / Revisions
11/10/21	Addition to part B criteria with effective date 1/1/2022
11/2/22	Addition to exclusion criteria and references; update to initial criteria

ULTOMIRIS (ravulizumab-cwvz) J1303

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, **the plan may authorize coverage** for Ultomiris (ravulizumab-cwvz) for **6 months (initial – PNH, gMG) and 12 months (initial and reauthorization – aHUS and reauthorization – PNH, gMG).** For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
Ultomiris (ravulizumab-cwvz) is a complement inhibitor indicated for: • The treatment of adult and pediatric patients one month of age and older with paroxysmal nocturnal hemoglobinuria (PNH) • Treatment of adults and pediatric patients one month of age and older with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy (TMA) • Treatment of adult patients with generalized myasthenia gravis (gMG) who are anti-acetylcholine receptor (AChR) antibody-positive

Required Medical Information:

1. **Paroxysmal nocturnal hemoglobinuria**
    a. Patient has confirmed PNH demonstrated by peripheral blood flow cytometry showing the absence or deficiency of GPI-anchored proteins (i.e. CD55, CD59) on at least 2 cell lineages (i.e. erythrocytes, granulocytes), **AND**
    b. Prescribed by or in consultation with a hematologist, **AND**
    c. Patient is one month of age or older, **AND**
    d. Prescriber must be enrolled in the ULTOMIRIS REMS program, **AND**
    e. Patient must be immunized with meningococcal vaccines at least 2 weeks prior to administering the first dose of ULTOMIRIS, unless the risks of delaying ULTOMIRIS therapy outweigh the risks of developing a meningococcal infection.
2. **Atypical hemolytic uremic syndrome**
    a. Prescribed by or in consultation with a nephrologist, **AND**
    b. Patient is one month of age or older, **AND**
    c. Patient does not have Shiga toxin E. coli related hemolytic uremic syndrome, **AND**
    d. Prescriber must be enrolled in the ULTOMIRIS REMS program, **AND**
    e. Patient must be immunized with meningococcal vaccines at least 2 weeks prior to administering the first dose of ULTOMIRIS, unless the risks of delaying ULTOMIRIS therapy outweigh the risks of developing a meningococcal infection.
3. **Generalized myasthenia gravis (gMG)**
    a. Patient is anti-acetylcholine receptor (AChR) antibody-positive, **AND**
    b. Patient is 18 years of age or older, **AND**
    c. Therapy is prescribed by or in consultation with a neurologist, **AND**

d. Patient is currently receiving or has tried and has contraindications, intolerance, or failed pyridostigmine, **AND**
e. Patient has evidence of unresolved symptoms of generalized Myasthenia Gravis (gMG), such as difficulty swallowing, difficulty breathing, or a functional disability resulting in the discontinuation of physical activity (e.g., double vision, talking, impairment of mobility), **AND**
f. Patient has myasthenia gravis foundation of America classification of II to IV and myasthenia gravis activities of daily living (MG-ADL) score of greater than or equal to 5, **AND**
g. Prescriber must be enrolled in the ULTOMIRIS REMS program, **AND**
h. Patient must be immunized with meningococcal vaccines at least 2 weeks prior to administering the first dose of ULTOMIRIS, unless the risks of delaying ULTOMIRIS therapy outweigh the risks of developing a meningococcal infection.

Reauthorization:

1. **Paroxysmal nocturnal hemoglobinuria**
    a. Diagnosis of PNH AND physician attestation of benefit (i.e. reduction in number of blood transfusions, improvement/stabilization of lactate dehydrogenase (LDH) and hemoglobin levels) compared to baseline
2. **Atypical hemolytic uremic syndrome**
    a. Diagnosis of aHUS AND physician attestation of benefit
3. **Generalized myasthenia gravis (gMG)**
    a. Diagnosis of gMG and physician attestation of benefit (e.g., reductions in exacerbations of myasthenia gravis, improvements in speech, swallowing, mobility, and respiratory function)

Exclusion Criteria:

- Ultomiris is contraindicated in patients with unresolved *Neisseria meningitidis* infection and in patients who are not currently vaccinated against *Neisseria meningitidis*, unless the risks of delaying treatment outweigh the risks of developing a meningococcal infection.
- Ultomiris is not indicated for the treatment of patients with Shiga toxin E. coli related hemolytic uremic syndrome (STEC-HUS).
- Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Ultomiris prescribing information

Version History

Last Reviewed Date	Updates / Revisions
2/17/2021	Addition to part B criteria with effective date 9/1/2021
11/10/21	Update to FDA approved indication and required medical information
8/23/22	Update to FDA approved indication and approval time frame
11/2/22	Update to initial authorization time frame for gMG; Update to initial and reauthorization criteria

VENTAVIS (iloprost) Q4074

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Ventavis (iloprost) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
VENTAVIS (iloprost) is a prostacyclin mimetic indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of deterioration. Studies establishing effectiveness included predominately patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH (65%) or PAH associated with connective tissue diseases (23%).

Required Medical Information:
1. Diagnosis of pulmonary arterial hypertension (PAH) (WHO Group 1), **AND**
2. The patient has primary pulmonary hypertension or pulmonary hypertension which is secondary to one of the following conditions: connective tissue disease, human immunodeficiency virus (HIV) infection, cirrhosis, anorexigens or congenital left to right shunts, **AND**
3. Documented confirmatory of pulmonary arterial hypertension (PAH) diagnosis based on right heart catheterization with the following parameters:
    a. Mean pulmonary artery pressure (PAP) of > 20 mmHg
    b. Pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg
    c. Pulmonary vascular resistance (PVR) of ≥ 3 Wood units, **AND**
4. Patient has New York Heart Association-World Health Group (NYHA-WHO) Functional Class III-IV symptoms and meets ONE of the following:
    a. Patients with WHO Functional Class III symptoms must have had a trial of or contraindication to TWO of the following agents from different drug classes:
        i. An oral endothelin receptor antagonist (e.g., ambrisentan, bosentan, or macitentan)
        ii. An oral phosphodiesterase-5 inhibitor (e.g., sildenafil or tadalafil)
        iii. An oral cGMP inhibitor (e.g., riociguat), **OR**
    b. Patients with WHO Functional Class III symptoms with evidence of rapid progression or poor prognosis, OR WHO Functional Class IV symptoms must have had trial of or contraindication to ONE of the following:
        i. An IV/SQ prostacyclin (e.g., epoprostenol or treprostinil), **AND**
5. Prescribed by or given in consultation with a cardiologist or pulmonologist, **AND**
6. The pulmonary hypertension has progressed despite maximal medical and/or surgical treatment of the identified condition, **AND**
7. The beneficiary has significant symptoms from the pulmonary hypertension (i.e., severe dyspnea on exertion, and either fatigability, angina, or syncope), **AND**
8. Treatment with oral calcium channel blocking agents has been tried and failed or has been considered and ruled out.

Reauthorization:

- Diagnosis of pulmonary arterial hypertension (PAH) (WHO Group 1), **AND** one of the following:
    - Patient has shown improvement from baseline in the 6-minute walk distance, **OR**
    - Patient has remained stable from baseline in the 6-minute walk distance, **AND** Patient's World Health Group (WHO) functional class remained stable or has improved

Exclusion Criteria:

1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.
2. The pulmonary hypertension is not secondary to pulmonary venous hypertension (e.g., left sided atrial or ventricular disease, left sided valvular heart disease) or disorders of the respiratory system (e.g., chronic obstructive pulmonary disease, interstitial lung disease, obstructive sleep apnea or other sleep disordered breathing, alveolar hypoventilation disorders)

References:

Ventavis prescribing information
World Symposium on Pulmonary Hypertension Guidelines

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	None
12/18/17	Update to required medical information
11/5/18	Addition of reauthorization criteria
12/10/19	None
11/10/20	None
11/10/21	None
11/2/22	Addition to exclusion criteria and references; update to initial criteria

VPRIV (velaglucerase) J3385

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for Vpriv (velaglucerase) for **one year**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
VPRIV (velaglucerase alfa for injection) is a hydrolytic lysosomal glucocerebroside-specific enzyme indicated for long-term enzyme replacement therapy (ERT) for pediatric and adult patients with type 1 Gaucher disease.

Required Medical Information:
1. Documented diagnosis of Type 1 Gaucher disease, **AND**
2. Systemic manifestations including ONE of the following:
    a. Skeletal disease (joint deterioration, pathological fracture, avascular necrosis, definite osteopenia, marrow infiltration), OR
    b. Anemia, OR
    c. Thrombocytopenia, OR
    d. Hepatomegaly, OR
    e. Splenomegaly

Reauthorization:
1. Patient is being treated for an FDA approved indication, or indication supported by NCD, LCD, or medical compendia **AND** physician attestation of improvement or stabilization.

Exclusion Criteria:
1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:
Vpriv prescribing information
National Gaucher Foundation http://www.gaucherdisease.org/

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	None
12/18/17	Update to required information

11/5/18	None
12/10/19	None
11/10/20	None
2/17/21	Addition of reauthorization criteria
11/10/21	None
11/2/22	None

XIAFLEX (collagenase clostridium histolyticum) J0775

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Xiaflex (collagenase clostridium histolyticum) for **3 months (Dupuytren's Contracture) and 6 months (Peyronie's Disease).** For this policy, the term "inadequate response" means lack of therapeutic effect, inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
1. Treatment of adult patients with Dupuytren's contracture with a palpable cord 2. Treatment of adult men with Peyronie's disease with a palpable plaque and curvature deformity of at least 30 degrees at the start of therapy. Because of the risks of corporal rupture or other serious penile injury, Xiaflex is available only through a restricted program under a REMS called the Xiaflex REMS Program. Prescribers and health care sites must be certified with the program.

Required Medical Information:

1. **Dupuytren's contracture**
    a. Documented diagnosis of Dupuytren's contracture with palpable cord, **AND**
    b. Provider must be a healthcare provider who is experienced with injection procedures of the hand and in treatment of Dupuytrens contracture (i.e. orthopedic physician/hand specialist), **AND**
    c. at baseline (prior to initial injection of Xiaflex), the patient had contracture of a metacarpophalangeal (MP) or proximal interphalangeal (PIP) joint of at least 20 degrees, **AND**
    d. the patient will not be treated with more than a total of three injections (maximum) per affected cord, **AND**
    e. Patient must be 18 years old or older, **AND**
    f. The prescriber has completed Xiaflex training as provided by the Risk Evaluation and Mitigation Strategy (REMS) program

2. **Peyronie's disease in men**
    a. Palpable plaque, **AND**
    b. Patient meets ONE of the following:
        - at baseline (prior to use of Xiaflex), the patient has a penile curvature deformity of at least 30 degrees **OR**
        - in a patient who has received prior treatment with Xiaflex, the patient has a penile curvature deformity of at least 15 degrees AND the patient has not previously been treated with a complete course (8 injections) of Xiaflex for Peyronie's disease, **AND**
    c. Administered by a healthcare provider experienced in the treatment of male urological diseases, **AND**
    d. Patient must be 18 years old or older, **AND**
    e. The prescriber has completed Xiaflex training as provided by the Risk Evaluation and Mitigation Strategy (REMS) program

Exclusion Criteria:

1. Treatment of Peyronie's plaques that involve the penile urethra
2. History of hypersensitivity to Xiaflex or to collagenase used in any other therapeutic applications
3. Retreatment (i.e., treatment beyond three injections per affected cord for those with Dupuytren's Contracture or beyond eight injections for Peyronie's Disease).
4. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Xiaflex prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	None
12/18/17	Update to required information for Peyronie's Disease
11/5/18	None
12/10/19	None
11/10/20	None
11/10/21	Addition to required medical information and exclusion criteria, Update to approval timeframe
11/2/22	Update to required medical information

XOLAIR (omalizumab) J2357

Covered Uses:

FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:

If all conditions are met, the plan may authorize coverage for Xolair (omalizumab) for **4 months (initial – Asthma and Chronic idiopathic/spontaneous urticaria), 6 months (initial – Nasal Polyps), and 12 months (reauthorization)**. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):
XOLAIR (omalizumab) is an anti-IgE antibody indicated for: 1. Adults and adolescents 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. 2. Chronic spontaneous urticaria [CSU; previously referred to as chronic idiopathic urticaria (CIU)] in adults and adolescents 12 years of age and older who remain symptomatic despite H1 antihistamine treatment. 3. Nasal polyps in adult patients 18 years of age and older with inadequate response to nasal corticosteroids, as add-on maintenance treatment. * Xolair has been approved for self-administration. Initiate therapy in a healthcare setting and once therapy has been safely established, the healthcare provider may determine whether self-administration of XOLAIR prefilled syringe by the patient or caregiver is appropriate, based on careful assessment of risk for anaphylaxis and mitigation strategies.

Initially review for B vs D coverage

- Xolair prefilled syringe – Initiate therapy in a healthcare setting and once therapy has been safely established, the healthcare provider may determine whether self-administration by the patient or caregiver is appropriate.
    - Determine if being administered by healthcare provider or patient/caregiver.
    - Self-administered by patient/caregiver is not covered under Part B.

- Xolair lyophilized powder – should only be prepared and injected by a healthcare provider.
    - Covered under part B – health plan to review

Required Medical Information:

Asthma

Initial Authorization	Reauthorization
1. Diagnosis of moderate to severe persistent asthma, **AND** 2. Positive skin test or in vitro testing (ie, a blood test	1. Patient has responded to therapy as determined by the prescribing physician, **AND** 2. Continues to receive therapy with one inhaled

for allergen-specific IgE antibodies such as an enzyme-linked immunoabsorbant assay (eg, immunoCAP, ELISA) or the RAST) for 1 or more perennial aeroallergens (eg, house dust mite, animal dander, cockroach, feathers, mold spores) and/or for 1 or more seasonal aeroallergens (grass, pollen, weeds), **AND** 3. Patient has received at least 3 months of combination therapy with an inhaled corticosteroid and at least one the following: long-acting beta-agonist (LABA), long-acting muscarinic antagonist (LAMA), leukotriene receptor antagonist, or theophylline. An exception can be made if the patient has already received anti-IL-4/13 (Dupixent) concomitantly with an ICS for at least 3 consecutive months, **AND** 4. Patient's asthma is uncontrolled or was uncontrolled prior to receiving any Xolair or anti-IL-4/13 therapy (Dupixent) as defined by ONE of the following: a. Patient experienced two or more asthma exacerbations requiring treatment with systemic corticosteroids in previous year, **OR** b. Patient experienced one or more asthma exacerbations requiring hospitalization or ED visit in the previous year, **OR** c. Patient has forced expiratory volume in 1 second (FEV1) less than 80% predicted, **OR** d. Patient has FEV1/forced vital capacity (FVC) less than 0.80, **OR** e. Patient's asthma worsens upon tapering of oral corticosteroid therapy, **AND** 5. Baseline IgE serum level is greater than or equal to 30 IU/ml, **AND** 6. Patient is 6 years of age or older, **AND** 7. Prescribed by or given in consultation with an allergist, immunologist, or pulmonologist	corticosteroid or inhaled corticosteroid containing combination product

Chronic idiopathic/spontaneous urticaria

Initial Authorization	Re-authorization
1. Diagnosis of chronic idiopathic urticaria (CIU; also called chronic spontaneous urticaria [CSU]), **AND** 2. Must have urticaria for more than 6 weeks, with symptoms present more than 3 days/wk despite daily non-sedating H1-antihistamine	Patient must have responded to therapy as determined by the prescribing physician.

therapy (e.g., cetirizine, desloratadine, fexofenadine, levocetirizine, loratadine), **AND** 3. Patient is 12 years or age or older, **AND** 4. Prescribed by or given in consultation with an allergist, immunologist, or dermatologist	

Nasal Polyps

Initial Authorization	Re-authorization
1. The patient has a baseline IgE level greater than or equal to 30 IU/ml, **AND** **2.** Patient is experiencing significant rhinosinusitis symptoms such as nasal obstruction, rhinorrhea, or reduction/loss of smell, **AND** 3. Patient is currently receiving therapy with an intranasal corticosteroid, **AND** 4. Prescribed by or in consult with an allergist, immunologist, or otolaryngologist, **AND** 5. Patient is 18 years and older	The patient continues to receive therapy with an intranasal corticosteroid and has responded to therapy

Exclusion Criteria:

1. Xolair is not indicated for other allergic conditions or other forms of urticaria
2. Xolair is not indicated for acute bronchospasm or status asthmaticus
3. Xolair should not be used in combination with an IL antagonist monoclonal antibody
4. Severe hypersensitivity reaction to XOLAIR or any ingredient of XOLAIR
5. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Xolair prescribing information

Version History

Last Reviewed Date	Updates / Revisions
12/6/16	Change in reauthorization from 6 months to 12 months; Change to asthma indication from 12 years of age and older to 6 years of age and older
12/18/17	Update to initial authorization criteria; Update to re-authorization criteria; Update to exclusion criteria
11/5/18	Update to required information; Update to authorization period
1/1/19	Update to authorization period, required information, and exclusion criteria to align with part D criteria
12/10/19	Update to CIU required information and Update to Asthma re-authorization criteria to align with Part D – Effective 1/1/2020
11/10/20	Update to asthma required information and reauthorization criteria

5/12/21	Addition of new FDA approved indication with initial and reauthorization criteria
11/10/21	Update to nasal polyps required information and Update to CIU reauthorization criteria
11/2/22	Update to FDA indication and initial criteria for urticaria; Addition to exclusion criteria

Template Drug J code

Covered Uses:
FDA-approved indications and off-label indications as specified in NCD or LCD, or supported in the medical compendia. Clinical reviewer must override criteria when, in his/her professional judgment, the requested item is medically necessary.

Coverage Duration:
If all conditions are met, the plan may authorize coverage for <XXX> for <one year>. For this policy, the term "inadequate response" means lack of therapeutic effect, and/or inability to tolerate due to adverse effects, or contraindication to therapy.

FDA Approved Indication(s):

Required Medical Information:

Exclusion Criteria:
1. Coverage excluded for any indications that are not supported in FDA labeling, NCD, LCD, or medical compendia.

References:

Version History

Last Reviewed Date	Updates / Revisions