State Fiscal Year 2021 Print Annual Reviews Quarter 1

Count	Category/Medication
1.	Aldurazyme® (Laronidase)/Naglazyme® (Galsulfase)
2.	Brineura® (Cerliponase Alfa)
3.	Cholbam® (Cholic Acid)
4.	Defitelio® (Defibrotide)
5.	Elaprase® (Idursulfase)
6.	Gamifant® (Emapalumab-lzsg)
7.	Gattex® [Teduglutide (rDNA Origin)]
8.	H.P. Acthar® Gel (Repository Corticotropin Injection)
9.	Jynarque® (Tolvaptan)
10.	Kanuma® (Sebelipase Alfa)
11.	Keveyis® (Dichlorphenamide)
12.	Lidocaine Topical Products
13.	Luxturna® (Voretigene Neparvovec-rzyl)
14.	Mepsevii® (Vestronidase Alfa-vjbk)
15.	Myalept® (Metreleptin)
16.	Mytesi® (Crofelemer)
17.	Qualaquin® (Quinine Sulfate)
18.	Sylvant® (Siltuximab)
19.	Symlin® (Pramlintide)
20.	Vimizim® (Elosulfase Alfa)
21.	Xuriden® (Uridine Triacetate)
22.	Zinplava™ (Bezlotoxumab)

Fiscal Year 2021 = July 1, 2020 – June 30, 2021

NOTE: An analysis of the atypical [Aged, Blind, and Disabled (ABD)] patient subgroup of the Oklahoma Medicaid population has been performed pertaining to all recommendations included in this DUR Board print annual review packet to ensure fair and knowledgeable deliberation of the potential impact of the recommendations on this patient population.

Fiscal Year 2021 Annual Review of Aldurazyme® (Laronidase) and Naglazyme® (Galsulfase)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Aldurazyme® (Laronidase) Approval Criteria:

1. An FDA approved diagnosis of Hurler, Hurler-Scheie, or Scheie syndrome (mucopolysaccharidosis type I; MPS 1) confirmed by:
   a. Enzyme assay demonstrating a deficiency of alpha-L-iduronidase (IDUA) enzyme activity; or
   b. Molecular genetic testing to confirm pathogenic mutations in the *IDUA* gene; and
2. For Scheie syndrome, the provider must document that the member has moderate-to-severe symptoms; and
3. Aldurazyme® must be administered by a health care professional prepared to manage anaphylaxis; and
4. Initial approvals will be for the duration of 6 months. Reauthorization may be granted if the prescriber documents the member is responding well to treatment; and
5. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to package labeling.

Naglazyme® (Galsulfase) Approval Criteria:

1. An FDA approved diagnosis of Maroteaux-Lamy syndrome (mucopolysaccharidosis type VI; MPS VI) confirmed by:
   a. Enzyme assay demonstrating a deficiency of arylsulfatase B (ASB) enzyme activity; or
   b. Genetic testing to confirm diagnosis of MPS VI; and
2. Naglazyme® must be administered by a health care professional prepared to manage anaphylaxis; and
3. Initial approvals will be for the duration of 6 months. Reauthorization may be granted if the prescriber documents the member is responding well to treatment; and
4. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to package labeling.

Utilization of Aldurazyme® (Laronidase) and Naglazyme® (Galsulfase): Fiscal Year 2021

There was no SoonerCare utilization, including pharmacy and medical claims, of Aldurazyme® (laronidase) or Naglazyme® (galsulfase) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Aldurazyme® (Laronidase) and Naglazyme® (Galsulfase)

There were no prior authorization requests submitted for Aldurazyme® (laronidase) or Naglazyme® (galsulfase) during fiscal year 2021.

Recommendations

The College of Pharmacy does not recommend any changes to the current Aldurazyme® (laronidase) and Naglazyme® (galsulfase) prior authorization criteria at this time.

Fiscal Year 2021 Annual Review of Brineura® (Cerliponase Alfa)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Brineura® (Cerliponase Alfa) Approval Criteria:

1. An FDA approved diagnosis of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) also known as tripeptidyl peptidase-1 (TPP-1) deficiency; and
2. Member must have confirmed TPP-1 enzymatic deficiency via enzyme assay, confirmed by molecular analysis; and
3. Member must be 3 years of age or older; and
4. Brineura® must be prescribed by a specialist with expertise in the treatment of CLN2 (or an advanced care practitioner with a supervising physician who is a specialist with expertise in treating CLN2); and
5. Brineura® must be administered in a health care facility by a prescriber who is knowledgeable in intraventricular administration; and
6. Member must not have ventriculoperitoneal shunts or acute intraventricular access device-related complications; and
7. Member must not have documented generalized status epilepticus within 4 weeks of initiating treatment; and
8. Prescriber must verify member's blood pressure and heart rate will be monitored prior to each infusion, during infusion, and post-infusion; and
9. Prescriber must be willing to perform regular 12-lead electrocardiogram (ECG) evaluation at baseline and at least every 6 months and verify that they are acceptable to the prescriber; and
10. A baseline assessment must be performed to assess the Motor plus Language CLN2 score; and
11. Initial authorizations will be for the duration of 6 months, at which time compliance will be required for continued approval. After 12 months of utilization, the prescriber must verify the member is responding to the medication as demonstrated by a 2 point or less decline in Motor plus Language CLN2 score from baseline; and
12. Approval quantity will be based on Brineura® *Prescribing Information* and FDA approved dosing regimen.

Utilization of Brineura® (Cerliponase Alfa): Fiscal Year 2021

There was no SoonerCare pharmacy utilization of Brineura® (cerliponase alfa) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Fiscal Year 2021 Utilization: Medical Claims

Fiscal Year	*Total Members	†Total Claims	Total Cost	Cost/ Claim	Total Units
2021	1	8	$14,400	$1,800	2,400

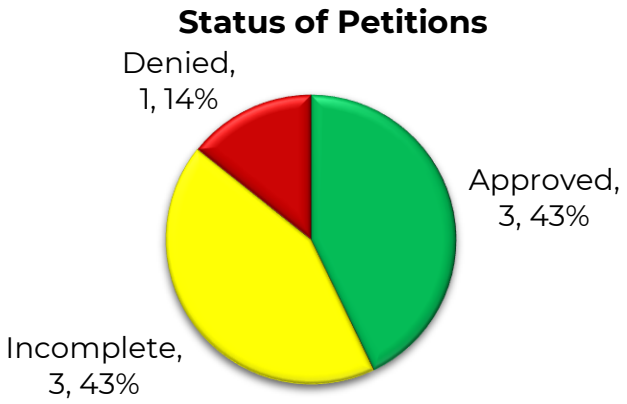
*Total number of unduplicated utilizing members.
†Total number of unduplicated claims.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021
Please note: There was no SoonerCare utilization of Brineura® (cerliponase alfa) during fiscal year 2020 (07/01/2019 to 06/30/2020) to allow for a fiscal year comparison.

Demographics of Members Utilizing Brineura® (Cerliponase Alfa)

- Due to the limited number of members utilizing Brineura® (cerliponase alfa) during fiscal year 2021, detailed demographic information could not be provided.

Prior Authorization of Brineura® (Cerliponase Alfa)

There were 7 prior authorization requests submitted for Brineura® (cerliponase alfa) for 1 unique member during fiscal year 2021. The following chart shows the status of the submitted petitions for fiscal year 2021.



Market News and Updates

Pipeline:

- **RGX-181:** Late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) is a rare genetic disorder caused by a mutation in the gene that makes tripeptidyl peptidase-1 (TPP-1). RGX-181 is a novel gene therapy that will use the adeno-associated virus 9 (AAV9) vector to deliver the TPP-1 gene directly into the central nervous system for patients with CLN2.

REGENXBIO is currently in the preclinical phase of development and plans to provide an update regarding the development program for RGX-181 in the second half of 2021.[1,2]

- **RGX-381:** REGENXBIO is also in the preclinical phase of development for RGX-381, a novel gene therapy for the treatment of ocular manifestations of CLN2. RGX-381 uses the AAV9 vector to deliver the TPP-1 gene directly to the retina. Currently there is no treatment available for the vision loss associated with disease progression in CLN2, which typically begins at the age of 4 years. RGX-381 is administered 1 time to the retina and may prevent future vision loss. More information regarding the development program for RGX-381 is expected in the second half of 2021.[3]

Recommendations

The College of Pharmacy does not recommend any changes to the current Brineura® (cerliponase alfa) prior authorization criteria at this time.

[1] REGENXBIO, Inc. Patients & Families: CLN2. Available online at: https://www.regenxbio.com/cln2/. Last accessed 09/15/2021.
[2] REGENXBIO, Inc. Therapeutic Programs: RGX-181. Available online at: https://regenxbio.com/rgx-181/. Last accessed 09/22/2021.
[3] REGENXBIO, Inc. Therapeutic Programs: RGX-381. Available online at: https://www.regenxbio.com/rgx-381/. Last accessed 09/20/2021.

Fiscal Year 2021 Annual Review of Cholbam® (Cholic Acid)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Cholbam® (Cholic Acid) Approval Criteria:

1. An FDA approved indication of 1 of the following:
   a. Treatment of bile acid disorders due to single enzyme defects (SEDs); or
   b. Adjunctive treatment of peroxisomal disorders (PDs) including Zellweger spectrum disorders in members who exhibit manifestations of liver disease, steatorrhea, or complications from decreased fat-soluble vitamin absorption; and
2. Treatment with Cholbam® should be initiated and monitored by a hematologist or pediatric gastroenterologist; and
3. The prescriber must verify that AST, ALT, GGT, alkaline phosphatase, bilirubin, and INR will be monitored every month for the first 3 months, every 3 months for the next 9 months, every 6 months during the next 3 years, and annually thereafter; and
4. Cholbam® should be discontinued if liver function does not improve within 3 months of starting treatment, if complete biliary obstruction develops, or if there are persistent clinical or laboratory indicators of worsening liver function or cholestasis; and
5. Initial approval will be for 3 months to monitor for compliance and liver function tests; and
6. Continuation approvals will be granted for the duration of 1 year; and
7. A quantity limit of 120 capsules per 30 days will apply. Quantity limit requests will be based on member's recent weight taken within the last 30 days.

Utilization of Cholbam® (Cholic Acid): Fiscal Year 2021

Fiscal Year 2021 Utilization: Pharmacy Claims

Fiscal Year	*Total Members	Total Claims	Total Cost	Cost/ Claim	Cost/ Day	Total Units	Total Days
2021	2	5	$41,432.05	$8,286.41	$276.21	150	150

*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021
Please note: There was no SoonerCare utilization of Cholbam® (cholic acid) during fiscal year 2020 (07/01/2019 to 06/30/2020) to allow for a fiscal year comparison.

Demographics of Members Utilizing Cholbam® (Cholic Acid)

- Due to the limited number of members utilizing Cholbam® (cholic acid) during fiscal year 2021, detailed demographic information could not be provided.

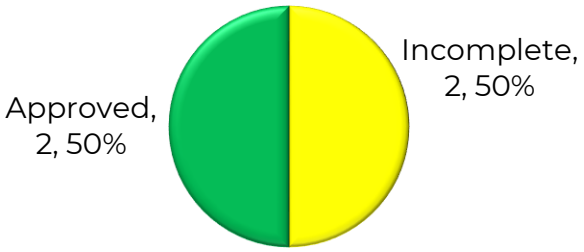
Top Prescriber Specialties of Cholbam® (Cholic Acid) by Number of Claims

- The only prescriber specialty listed on paid claims for Cholbam® (cholic acid) during fiscal year 2021 was pediatric gastroenterology.

Prior Authorization of Cholbam® (Cholic Acid)

There were 4 prior authorization requests submitted for Cholbam® (cholic acid) during fiscal year 2021. The following chart shows the status of the submitted petitions for fiscal year 2021.

Status of Petitions



Market News and Updates

Anticipated Patent Expiration(s):[4]

- Cholbam® (cholic acid): March 2022

Recommendations

The College of Pharmacy does not recommend any changes to the current Cholbam® (cholic acid) prior authorization criteria at this time.

Utilization Details of Cholbam® (Cholic Acid): Fiscal Year 2021

PRODUCT UTILIZED	TOTAL CLAIMS	TOTAL MEMBERS	TOTAL COST	COST/ CLAIM	CLAIMS/ MEMBER	% COST
CHOLBAM CAP 50MG	5	2	$41,432.05	$8,286.41	2.5	100%
TOTAL	**5**	**2***	**$41,432.05**	**$8,286.41**	**2.5**	**100%**

CAP = capsule
*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021

[4] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available on https://www.accessdata.fda.gov/scripts/cder/ob/patent_info.cfm. Last revised 09/2021. Last accessed 09/15/2021.

Fiscal Year 2021 Annual Review of Defitelio® (Defibrotide)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Defitelio® (Defibrotide) Approval Criteria:

1. An FDA approved diagnosis of hepatic veno-occlusive disease (VOD), also known as sinusoidal obstruction syndrome (SOS), with renal or pulmonary dysfunction following hematopoietic stem-cell transplantation (HSCT); and
2. Initial approvals will be for 1 month of therapy. An additional month of therapy (maximum total duration of 60 days of therapy) may be granted if the physician documents the continued need for therapy.

Utilization of Defitelio® (Defibrotide): Fiscal Year 2021

There was no SoonerCare utilization of Defitelio® (defibrotide) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Defitelio® (Defibrotide)

There were no prior authorization requests submitted for Defitelio® (defibrotide) during fiscal year 2021.

Market News and Updates

Anticipated Exclusivity Expiration(s):[5]

- Defitelio® (defibrotide): March 2023

Recommendations

The College of Pharmacy does not recommend any changes to the current Defitelio® (defibrotide) prior authorization criteria at this time.

[5] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available online at: https://www.accessdata.fda.gov/scripts/cder/ob/index.cfm. Last revised 09/2021. Last accessed 9/15/2021.

Fiscal Year 2021 Annual Review of Elaprase® (Idursulfase)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Elaprase® (Idursulfase) Approval Criteria:

1. An FDA approved diagnosis of Hunter syndrome (mucopolysaccharidosis type II; MPS II) confirmed by:
   a. Enzyme assay demonstrating a deficiency of iduronate-2-sulfatase (IDS) enzyme activity; or
   b. Molecular genetic testing confirming a hemizygous pathogenic variant in the *IDS* gene; and
2. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to the Elaprase® *Prescribing Information*.

Utilization of Elaprase® (Idursulfase): Fiscal Year 2021

Fiscal Year 2021 Utilization: Pharmacy Claims

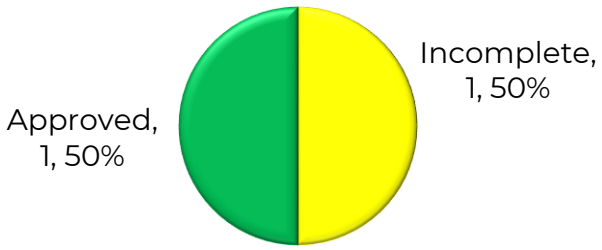
Fiscal Year	*Total Members	Total Claims	Total Cost	Cost/ Claim	Cost/ Day	Total Units	Total Days
2021	1	5	$188,206.85	$37,641.37	$1,344.33	180	140

*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021
Please note: There was no SoonerCare utilization of Elaprase® (idursulfase) during fiscal year 2020 (07/01/2019 to 06/30/2020) to allow for a fiscal year comparison.

Prior Authorization of Elaprase® (Idursulfase)

There were 2 prior authorization requests submitted for Elaprase® (idursulfase) during fiscal year 2021. The following chart shows the status of the submitted petitions for fiscal year 2021.

Status of Petitions



Market News and Updates

Pipeline:

- **DNL310:** Denali Therapeutics is developing DNL310 for the treatment of Hunter syndrome (mucopolysaccharidosis II or MPS II). DNL310 is a recombinant iduronate 2-sulfatase (IDS) enzyme engineered to cross the blood-brain-barrier (BBB) to replace the IDS enzyme and treat neuropathic and systemic forms of MPS II. DNL310 is currently in the early clinical stage of development. In November 2020, Denali announced the first human biomarker proof of concept data for its proprietary Transport Vehicle (TV) technology from a Phase 1/2 study of DNL310 in 5 pediatric patients with MPS II. The TV technology is designed to effectively deliver large therapeutic molecules, such as antibodies, enzymes, proteins, and oligonucleotides, across the BBB after intravenous administration. In July 2021, Denali announced positive interim data from the Phase 1/2 study, which demonstrated a safety profile consistent with enzyme replacement therapy and demonstrated improvements in clinical presentation. Based on these results, Denali plans to initiate a pivotal Phase 2/3 study in the first half of 2022.[6,7,8]
- **RGX-121:** REGENXBIO is developing RGX-121 for the treatment of MPS II. RGX-121 is a one-time gene therapy that uses the adeno-associated virus serotype 9 (AAV9) vector to deliver the *IDS* gene to the central nervous system (CNS) in an attempt to address the neurological manifestations of disease and prevent or stabilize cognitive decline. RGX-121 is administered intracisternally into the ventricles of the brain which could provide a permanent source of secreted IDS beyond the BBB. RGX-121 has received Orphan Drug, Rare Pediatric Disease, and Fast Track designations from the U.S. Food and Drug Administration (FDA). In May 2021, REGENXBIO announced positive interim data from the ongoing Phase 1/2 study in pediatric patients up to 5 years of age with MPS II, showing that patients treated with RGX-121 continue to have biomarker data indicating the IDS enzyme is active in the CNS. Additionally, continued cognitive development has been observed in the majority of patients who have been followed for more than 6 months. Patient

[6] Denali Therapeutics, Inc. Denali Pipeline. Available online at: https://www.denalitherapeutics.com/pipeline. Last accessed 09/15/2021.
[7] Denali Therapeutics, Inc. Denali Therapeutics Announces First Human Biomarker Proof of Concept for Its Transport Vehicle (TV) Technology Achieved in Phase 1/2 Study of ETV:IDS (DNL310) in Hunter Syndrome (MPS II). *Globe Newswire*. Available online at: https://www.globenewswire.com/news-release/2020/11/10/2123675/0/en/Denali-Therapeutics-Announces-First-Human-Biomarker-Proof-of-Concept-for-Its-Transport-Vehicle-TV-Technology-Achieved-in-Phase-1-2-Study-of-ETV-IDS-DNL310-in-Hunter-Syndrome-MPS-II.html. Issued 11/10/2020. Last accessed 09/22/2021.
[8] Denali Therapeutics, Inc. Denali Therapeutics Announces Positive Interim Data from Phase 1/2 Study with ETV:IDS (DNL130) in Patients with Lysosomal Storage Disease Hunter Syndrome (MPSII). Available online at: https://www.denalitherapeutics.com/investors/press-release?id=8321&type=api. Issued 07/25/2021. Last accessed 09/22/2021.

dosing has begun in a third cohort, and REGENXBIO plans to provide additional updates later in 2021.[9,10]

Recommendations

The College of Pharmacy does not recommend any changes to the current Elaprase® (idursulfase) prior authorization criteria at this time.

Utilization Details of Elaprase® (Idursulfase): Fiscal Year 2021

PRODUCT UTILIZED	TOTAL CLAIMS	TOTAL MEMBERS	TOTAL COST	COST/ CLAIM	CLAIMS/ MEMBER	% COST
ELAPRASE INJ 6MG/3ML	5	1	$188,206.85	$37,641.37	5	100%
TOTAL	**5**	**1***	**$188,206.85**	**$37,641.37**	**5**	**100%**

INJ = injection
*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021

[9] REGENXBIO, Inc. REGENXBIO Pipeline: RGX-121. Available online at: https://regenxbio.com/rgx-121/. Last accessed 09/22/2021.
[10] REGENXBIO, Inc. REGENXBIO Presents Additional Positive Interim Data from Phase I/II Trial of RGX-121 for the Treatment of MPS II (Hunter Syndrome) at American Society of Gene and Cell Therapy's 24th Annual Meeting. Available online at: https://regenxbio.gcs-web.com/news-releases/news-release-details/regenxbio-presents-additional-positive-interim-data-phase-iii-0. Issued 05/14/2021. Last accessed 09/22/2021.

Fiscal Year 2021 Annual Review of Gamifant® (Emapalumab-lzsg)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Gamifant® (Emapalumab-lzsg) Approval Criteria:

1. An FDA approved indication for the treatment of adult and pediatric members with primary hemophagocytic lymphohistiocytosis (HLH) with refractory, recurrent, or progressive disease or who are intolerant to conventional HLH therapy; and
2. Diagnosis of primary HLH must be confirmed by 1 of the following:
    a. Genetic testing confirming mutation of a gene known to cause primary HLH (e.g., *PRF*, *UNC13D*, *STX11*); or
    b. Family history consistent with primary HLH; or
    c. Member meets 5 of the following 8 diagnostic criteria:
        i. Fever; or
        ii. Splenomegaly; or
        iii. Cytopenias affecting at least 2 of 3 lineages in the peripheral blood (hemoglobin <9, platelets <100 x 10^9/L, neutrophils <1 x 10^9/L); or
        iv. Hypertriglyceridemia (fasting triglycerides >3mmol/L or ≥265mg/dL) and/or hypofibrinogenemia (≤1.5g/L); or
        v. Hemophagocytosis in bone marrow, spleen, or lymph nodes with no evidence of malignancy; or
        vi. Low or absent natural killer (NK)-cell activity; or
        vii. Hyperferritinemia (ferritin ≥500mcg/L); or
        viii. High levels of soluble interleukin-2 receptor (soluble CD25 ≥2,400U/mL); and
3. Gamifant® must be prescribed by, or in consultation with, a physician who specializes in the treatment of immune deficiency disorders; and
4. Member must have at least 1 of the following:
    a. Failure of at least 1 conventional HLH treatment (e.g., etoposide, dexamethasone, cyclosporine); or
    b. Documentation of progressive disease despite conventional HLH treatment; or
    c. A patient-specific, clinically significant reason why conventional HLH treatment is not appropriate for the member must be provided; and
5. Prescriber must verify dexamethasone dosed at least 5mg/m^2/day will be used concomitantly with Gamifant®; and

6. Prescriber must verify member has received or will receive prophylaxis for herpes zoster, *Pneumocystis jirovecii,* and fungal infection(s); and
7. Prescriber must verify member will be monitored for tuberculosis (TB), adenovirus, Epstein-Barr virus (EBV), and cytomegalovirus (CMV) every 2 weeks and as clinically indicated; and
8. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to the Gamifant® *Prescribing Information*; and
9. Approvals will be for the duration of 6 months with reauthorization granted if the prescriber documents the member is responding well to treatment, no unacceptable toxicity has occurred, and the member has not received hematopoietic stem cell transplantation (HSCT).

Utilization of Gamifant® (Emapalumab-lzsg): Fiscal Year 2021

There was no SoonerCare utilization, including pharmacy and medical claims, of Gamifant® (emapalumab-lzsg) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Gamifant® (Emapalumab-lzsg)

There were no prior authorization requests submitted for Gamifant® (emapalumab-lzsg) during fiscal year 2021.

Recommendations

The College of Pharmacy does not recommend any changes to the current Gamifant® (emapalumab-lzsg) prior authorization criteria at this time.

Fiscal Year 2021 Annual Review of Gattex® [Teduglutide (rDNA Origin)]

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Gattex® [Teduglutide (rDNA Origin)] Approval Criteria:

1. An FDA approved diagnosis of severe short bowel syndrome; and
2. Member must have required parenteral nutrition at least 3 times per week, every week, for the past 12 months; and
3. Documentation of all of the following:
    a. Prior use of supportive therapies (e.g., anti-motility agents, proton pump inhibitors, bile acid sequestrants, octreotide); and
    b. Colonoscopy within the previous 6 months, with removal of polyps if present; and
    c. Gastro-intestinal malignancy has been ruled out; and
4. Approval will be for the duration of 3 months, after which time, prescriber must verify benefit of medication by documented reduction of at least 20% in parenteral support. Subsequent approvals will be for the duration of 1 year.

Utilization of Gattex® [Teduglutide (rDNA Origin)]: Fiscal Year 202

Fiscal Year	*Total Members	Total Claims	Total Cost	Cost/ Claim	Cost/ Day	Total Units	Total Days
2021	2	14	$577,381.80	$41,241.56	$1,374.72	14	420

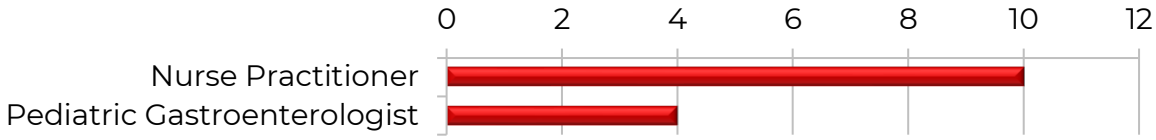
*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021
Please note: There was no SoonerCare utilization of Gattex® [teduglutide (rDNA origin)] during fiscal year 2020 (07/01/2019 to 06/30/2020) to allow for a fiscal year comparison.

Prior Authorization of Gattex® [Teduglutide (rDNA Origin)]

There were 16 prior authorization requests submitted for Gattex® [teduglutide (rDNA origin)] during fiscal year 2021. The following chart shows the status of the submitted petitions for fiscal year 2021.

Status of Petitions

Top Prescriber Specialties of Gattex® [Teduglutide (rDNA Origin)] by Number of Claims



Market News and Updates

Anticipated Exclusivity Expiration(s):[11]

- Gattex® [teduglutide (rDNA origin)]: May 2026

Recommendations

The College of Pharmacy does not recommend any changes to the current Gattex® [teduglutide (rDNA origin)] prior authorization criteria at this time.

Utilization Details of Gattex® [Teduglutide (rDNA Origin)]: Fiscal Year 2021

PRODUCT UTILIZED	TOTAL CLAIMS	TOTAL MEMBERS	TOTAL COST	COST/ CLAIM	CLAIMS/ MEMBER	% COST
GATTEX KIT 5MG	14	2	$577,381.80	$41,241.56	7	100%
TOTAL	**14**	**2***	**$577,381.80**	**$41,241.56**	**7**	**100%**

*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021

[11] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available online at: https://www.accessdata.fda.gov/scripts/cder/ob/index.cfm. Last revised 09/2021. Last accessed 09/23/2021.

Fiscal Year 2021 Annual Review of H.P. Acthar® Gel (Repository Corticotropin Injection)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

H.P. Acthar® Gel (Repository Corticotropin Injection) Approval Criteria:

1. An FDA approved diagnosis of infantile spasms; and
    a. Member must be 2 years of age or younger; and
    b. Must be prescribed by, or in consultation with, a neurologist or an advanced care practitioner with a supervising prescriber that is a neurologist; or
2. An FDA approved diagnosis of multiple sclerosis (MS); and
    a. Member is experiencing an acute exacerbation; and
    b. Must be prescribed by, or in consultation with, a neurologist or an advanced care practitioner with a supervising prescriber that is a neurologist or a prescriber that specializes in MS; and
    c. Prescriber must rule out pseudo-exacerbation from precipitating factors (e.g., pain, stress, infection, premenstrual syndrome); and
    d. Symptoms of acute exacerbation last at least 24 hours; and
    e. Member must be currently stable within the last 30 days on an immunomodulator agent, unless contraindicated; and
    f. A patient-specific, clinically significant reason why the member cannot use alternative corticosteroid therapy [e.g., intravenous (IV) methylprednisolone, IV dexamethasone, oral prednisone] must be provided; and
    g. A quantity limit of daily doses of up to 120 units for up to 3 weeks for acute exacerbation will apply; or
3. An FDA approved diagnosis of nephrotic syndrome without uremia of the idiopathic type or that is due to lupus erythematosus to induce a diuresis or a remission of proteinuria; and
    a. Must be prescribed by, or in consultation with, a nephrologist or an advanced care practitioner with a supervising prescriber that is a nephrologist; and
    b. A patient-specific, clinically significant reason why the member cannot use alternative corticosteroid therapy (e.g., prednisone) must be provided; or
4. An FDA approved diagnosis of the following disorders or diseases: rheumatic; collagen; dermatologic; allergic states; ophthalmic; respiratory; or edematous states; and
    a. A patient-specific, clinically significant reason why the member cannot use alternative corticosteroid therapy must be provided.

Utilization of H.P. Acthar® Gel (Repository Corticotropin Injection): Fiscal Year 2021

Comparison of Fiscal Years: Pharmacy Claims

Fiscal Year	*Total Members	Total Claims	Total Cost	Cost/ Claim	Cost/ Day	Total Units	Total Days
2020	5	11	$623,556.17	$56,686.92	$3,542.93	80	176
2021	1	4	$159,501.64	$39,875.41	$7,975.08	20	20
% Change	**-80.0%**	**-63.6%**	**-74.4%**	**-29.7%**	**125.1%**	**-75.0%**	**-88.6%**
Change	**-4**	**-7**	**-$464,054.53**	**-$16,811.51**	**$4,432.15**	**-60**	**-156**

*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2020 = 07/01/2019 to 06/30/2020; Fiscal Year 2021 = 07/01/2020 to 06/30/2021

Demographics of Members Utilizing H.P. Acthar® Gel (Repository Corticotropin Injection)

- Due to the limited number of members utilizing H.P. Acthar® Gel during fiscal year 2021, detailed demographic information could not be provided.

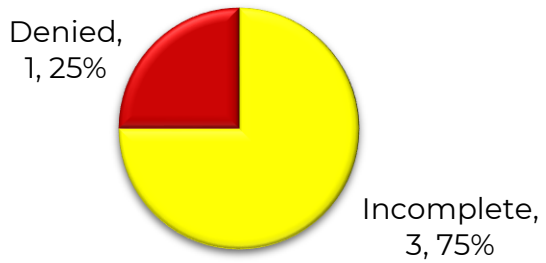
Top Prescriber Specialties of H.P. Acthar® Gel (Repository Corticotropin Injection) by Number of Claims

- The only prescriber specialty listed on paid pharmacy claims for H.P. Acthar® Gel was neurologist.

Prior Authorization of H.P. Acthar® Gel (Repository Corticotropin Injection)

There were 4 prior authorization requests submitted for 2 unique members for H.P. Acthar® Gel during fiscal year 2021. The following chart shows the status of the submitted petitions for fiscal year 2021.

Status of Petitions



Market News and Updates

News:

- **August 2021:** ANI Pharmaceuticals announced the U.S. Food and Drug Administration (FDA) has accepted their supplemental New Drug

Application (sNDA) for purified Cortrophin® Gel for review for use in multiple indications, including multiple sclerosis, rheumatoid arthritis, and nephrotic syndrome. The FDA set a Prescription Drug User Fee Act (PDUFA) action date of October 29, 2021.[12]

Recommendations

The College of Pharmacy does not recommend any changes to the current H.P. Acthar® Gel (repository corticotropin injection) prior authorization criteria at this time.

Utilization Details of H.P. Acthar® Gel (Repository Corticotropin Injection): Fiscal Year 2021

PRODUCT UTILIZED	TOTAL CLAIMS	*TOTAL MEMBERS	TOTAL COST	COST/ CLAIM	CLAIMS/ MEMBER
ACTHAR INJ 80 UNIT	4	1	$159,501.64	$39,875.41	4
TOTAL	**4**	**1**	**$159,501.64**	**$39,875.41**	**4**

INJ = Injection
*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021

[12] ANI Pharmaceuticals, Inc. ANI Announces FDA Acceptance of Purified Cortrophin® Gel Supplemental New Drug application for Multiple Indications Including Multiple Sclerosis, Rheumatoid Arthritis, and Nephrotic syndrome. Available online at: https://investor.anipharmaceuticals.com/news/news-details/2021/ANI-Pharmaceuticals-Announces-FDA-Acceptance-of-Purified-Cortrophin-Gel-Supplemental-New-Drug-Application-for-Multiple-Indications-Including-Multiple-Sclerosis-Rheumatoid-Arthritis-and-Nephrotic-Syndrome/default.aspx. Issued 08/31/2021. Last Accessed 09/15/2021.

Fiscal Year 2021 Annual Review of Jynarque® (Tolvaptan)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Jynarque® (Tolvaptan) Approval Criteria:

1. An FDA approved indication to slow kidney function decline in adults at risk of rapidly progressing autosomal dominant polycystic kidney disease (ADPKD); and
2. Member must be 18 years of age or older; and
3. Member must not have any contraindications to taking Jynarque® including the following:
    a. Taking any concomitant strong CYP3A inhibitors (e.g., ketoconazole, itraconazole, lopinavir/ritonavir, indinavir/ritonavir, ritonavir, conivaptan); and
    b. History of signs or symptoms of significant liver impairment or injury (does not include uncomplicated polycystic liver disease); and
    c. Uncorrected abnormal blood sodium concentrations; and
    d. Unable to sense or respond to thirst; and
    e. Hypovolemia; and
    f. Hypersensitivity to tolvaptan or any of its components; and
    g. Uncorrected urinary outflow obstruction; and
    h. Anuria; and
4. Member must not be taking any of the following medications concomitantly with Jynarque®:
    a. Strong CYP3A inhibitors (e.g., ketoconazole, itraconazole, lopinavir/ritonavir, indinavir/ritonavir, ritonavir, conivaptan); and
    b. Strong CYP3A inducers (e.g., rifampin); and
    c. OATP1B1/3 and OAT3 transporter substrates (e.g., statins, bosentan, glyburide, nateglinide, repaglinide, methotrexate, furosemide); and
    d. BCRP transporter substrates (e.g., rosuvastatin); and
    e. V_2-receptor agonists (e.g., desmopressin); and
5. Jynarque® must be prescribed by a nephrologist or specialist with expertise in the treatment of ADPKD (or an advanced care practitioner with a supervising physician who is a nephrologist or specialist with expertise in the treatment of ADPKD); and
6. Prescriber must agree to assess ALT, AST, and bilirubin prior to initiation of Jynarque®, at 2 weeks and 4 weeks after initiation, then monthly for 18 months, and every 3 months thereafter; and

7. Female members must not be pregnant and must have a negative pregnancy test prior to therapy initiation; and
8. Prescriber, pharmacy, and member must be enrolled in the Jynarque® Risk Evaluation and Mitigation Strategy (REMS) program and maintain enrollment throughout therapy.

Utilization of Jynarque® (Tolvaptan): Fiscal Year 2021

Comparison of Fiscal Years: Pharmacy Claims

Fiscal Year	*Total Members	Total Claims	Total Cost	Cost/ Claim	Cost/ Day	Total Units	Total Days
2020	2	18	$262,780.89	$14,598.94	$521.39	1,008	504
2021	2	13	$200,725.69	$15,440.44	$551.44	728	364
% Change	**0.00%**	**-27.8%**	**-23.6%**	**5.80%**	**5.80%**	**-27.8%**	**-27.8%**
Change	**0**	**-5**	**-$62,055.20**	**$841.50**	**$30.05**	**-280**	**-140**

*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2020 = 07/01/2019 to 06/30/2020; Fiscal Year 2021 = 07/01/2020 to 06/30/2021

Demographics of Members Utilizing Jynarque® (Tolvaptan)

- Due to the limited number of members utilizing Jynarque® (tolvaptan) during fiscal year 2021, detailed demographic information could not be provided.

Top Prescriber Specialties Jynarque® (Tolvaptan) of by Number of Claims

- The only prescriber specialty listed on paid pharmacy claims for Jynarque® (tolvaptan) during fiscal year 2021 was nephrologist.

Prior Authorization of Jynarque® (Tolvaptan)

There were 4 prior authorization requests submitted for Jynarque® (tolvaptan) during fiscal year 2021. The following chart shows the status of the submitted petitions for fiscal year 2021.

Status of Petitions

Market News and Updates

Anticipated Patent Expiration(s):[13]

- Jynarque® (tolvaptan): April 2030

Recommendations

The College of Pharmacy does not recommend any changes to the current Jynarque® (tolvaptan) prior authorization criteria at this time.

Utilization Details of Jynarque® (Tolvaptan): Fiscal Year 2021

PRODUCT UTILIZED	TOTAL CLAIMS	TOTAL MEMBERS	TOTAL COST	COST/ DAY	COST/ CLAIM	% COST
JYNARQUE PAK 90-30MG	12	1	$185,693.46	$552.66	$15,474.46	92.51%
JYNARQUE PAK 45-15MG	1	1	$15,032.23	$536.87	$15,032.23	7.49%
TOTAL	**13**	**2***	**$200,725.69**	**$551.44**	**$15,440.44**	**100%**

PAK = pack
*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021

[13] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available online at: https://www.accessdata.fda.gov/scripts/cder/ob/index.cfm. Last revised 09/2021. Last accessed 09/23/2021.

Fiscal Year 2021 Annual Review of Kanuma® (Sebelipase Alfa)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Kanuma® (Sebelipase Alfa) Approval Criteria:

1. An FDA approved diagnosis of Lysosomal Acid Lipase (LAL) deficiency; and
2. Kanuma® (sebelipase alfa) must be administered in a health care setting by a health care professional prepared to manage anaphylaxis; and
3. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to the Kanuma® *Prescribing Information*.

Utilization of Kanuma® (Sebelipase Alfa): Fiscal Year 2021

There was no SoonerCare utilization of Kanuma® (sebelipase alfa) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Kanuma® (Sebelipase Alfa)

There were no prior authorization requests submitted for Kanuma® (sebelipase alfa) during fiscal year 2021.

Recommendations

The College of Pharmacy does not recommend any changes to the current Kanuma® (sebelipase alfa) prior authorization criteria at this time.

Fiscal Year 2021 Annual Review of Keveyis® (Dichlorphenamide)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Keveyis® (Dichlorphenamide) Approval Criteria:

1. An FDA approved indication for the treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, or related variants; and
2. Prescriber documentation that all non-pharmacological treatments failed including the following:
    a. Hyperkalemic periodic paralysis:
        i. Acute attacks can be aborted with sugar or mild exercise; and
        ii. Avoiding foods rich in potassium; and
        iii. Avoiding fasting; and
        iv. High-carbohydrate diet; and
        v. Avoiding strenuous activity; and
        vi. Avoiding prolonged cold exposure; or
    b. Hypokalemic periodic paralysis:
        i. Low-carbohydrate diet (avoiding carbohydrate loading); and
        ii. Avoiding vigorous exercise (some mild attacks can be aborted by low level exercise); and
3. Prescriber documentation of frequent and severe attacks requiring pharmacological treatment (at least 1 attack per week but no more than 3 attacks per day); and
4. A 4-week trial within the last 90 days of acetazolamide in combination with:
    a. Hydrochlorothiazide in hyperkalemic periodic paralysis; or
    b. Spironolactone or triamterene in hypokalemic periodic paralysis; and
5. A quantity limit of 4 tablets per day will apply; and
6. Initial approvals will be for the duration of 3 months after which time compliance will be required for continued approval. Additionally, for continuation the prescriber must include information regarding reduced frequency or severity of attacks.

Utilization of Keveyis® (Dichlorphenamide): Fiscal Year 2021

There has been no SoonerCare utilization of Keveyis® (dichlorphenamide) since it was approved by the U.S. Food and Drug Administration (FDA) in

August 2015 through fiscal year 2021 (fiscal year 2021 = 07/01/2020 to 06/30/2021).

Prior Authorization of Keveyis® (Dichlorphenamide)

There have been no prior authorization requests submitted for Keveyis® (dichlorphenamide) since it was FDA approved in August 2015 through fiscal year 2021.

Market News and Updates

Anticipated Exclusivity Expiration(s):[14]

- Keveyis® (dichlorphenamide): August 2022

New(s):

- **July 2021:** Strongbridge Biopharma announced the publication of long-term efficacy and safety data for Keveyis® for the treatment of primary periodic paralysis in the journal *Muscle & Nerve,* reporting that the safety and efficacy of Keveyis® is sustained long-term. According to post hoc analyses of the HYPHOP study, Keveyis® remained efficacious over the entire 61-week study with no evidence of waning over time. Safety parameters were maintained with no additional adverse safety signals reported during the last 52 weeks in comparison to the first 9 weeks of the comprehensive study. Paresthesia and cognitive impairment were the most commonly reported adverse effects, which resolved following dose reduction.[15]

Recommendations

The College of Pharmacy does not recommend any changes to the current Keveyis® (dichlorphenamide) prior authorization criteria at this time.

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[14] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available online at: https://www.accessdata.fda.gov/scripts/cder/ob/index.cfm. Last revised 09/2021. Last accessed 09/23/2021.

[15] Strongbridge Biopharma Plc. Strongbridge Biopharma Plc Announces Publication of Long-Term Efficacy and Safety Results for Keveyis® (Dichlorphenamide) for the Treatment of Primary Periodic Paralysis in Muscle & Nerve. *BioSpace*. Available online at: https://www.biospace.com/article/releases/strongbridge-biopharma-plc-announces-publication-of-long-term-efficacy-and-safety-results-for-keveyis-dichlorphenamide-for-the-treatment-of-primary-periodic-paralysis-in-muscle-and-nerve/. Issued 07/13/2021. Last accessed 09/23/2021.

Fiscal Year 2021 Annual Review of Lidocaine Topical Products

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Lidotral® (Lidocaine 3.88% Topical Cream) Approval Criteria:

1. A patient-specific, clinically significant reason why the member cannot use other formulations of lidocaine including lidocaine 3% topical cream, which is available without prior authorization, must be provided.

Synera® (Lidocaine/Tetracaine Patch) Approval Criteria:

1. Member must be 3 years of age or older; and
2. Member must have an FDA approved need for local dermal analgesia for superficial venous access or superficial dermatological procedures; and
3. A patient-specific, clinically significant reason why the member cannot use EMLA® (lidocaine/prilocaine) cream, which is available without a prior authorization, must be provided; and
4. The total number of procedures must be provided on the prior authorization request; and
5. A quantity limit of 2 patches per day will apply.

ZTlido® (Lidocaine 1.8% Topical System) Approval Criteria:

1. An FDA approved diagnosis of pain due to postherpetic neuralgia (PHN); and
2. Documented treatment attempts, at recommended dosing, of at least 1 agent from 2 of following drug classes that failed to provide adequate relief or contraindication(s) to all of the following classes:
   a. Tricyclic antidepressants; or
   b. Anticonvulsants; or
   c. Topical or oral analgesics; and
3. A patient-specific, clinically significant reason why the member cannot use lidocaine 5% topical patches, which are available without prior authorization, must be provided; and
4. A quantity limit of 3 patches per day with a maximum of 90 patches per 30 days will apply.

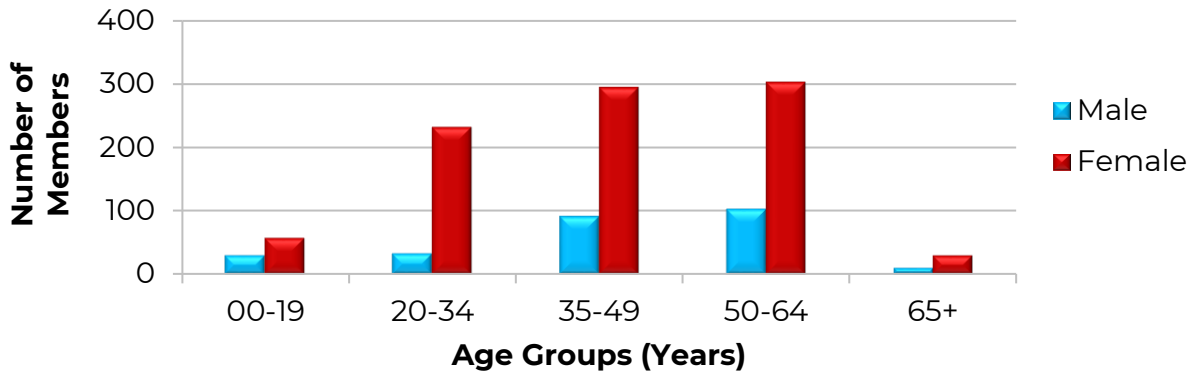
Utilization of Lidocaine Topical Products: Fiscal Year 2021

Comparison of Fiscal Years: Pharmacy Claims

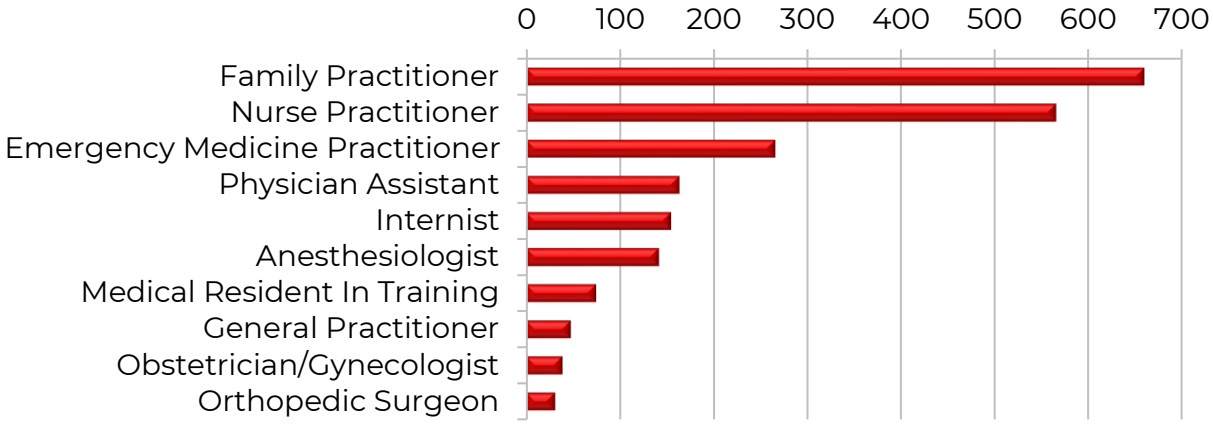
Fiscal Year	*Total Members	Total Claims	Total Cost	Cost/ Claim	Cost/ Day	Total Units	Total Days
2020	983	1,859	$167,425.70	$90.06	$3.40	56,779	49,225
2021	1,180	2,305	$187,691.11	$81.43	$3.06	72,810	61,386
% Change	**20.0%**	**24.0%**	**12.1%**	**-9.6%**	**-10.0%**	**28.2%**	**24.7%**
Change	**197**	**446**	**$20,265.41**	**-$8.63**	**-$0.34**	**16,031**	**12,161**

*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2020 = 07/01/2019 to 06/30/2020; Fiscal Year 2021 = 07/01/2020 to 06/30/2021

Demographics of Members Utilizing Lidocaine Topical Products



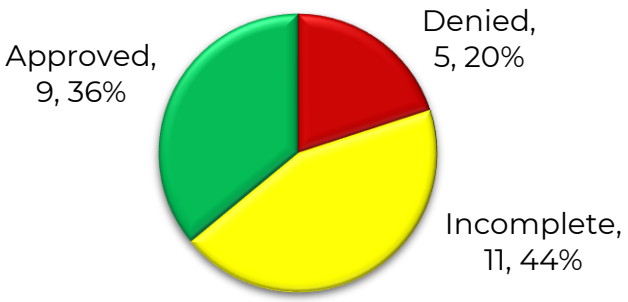
Top Prescriber Specialties of Lidocaine Topical Products by Number of Claims



Prior Authorization of Lidocaine Topical Products

There were 25 prior authorization requests submitted for lidocaine topical products during fiscal year 2021. The following chart shows the status of the submitted petitions.

Status of Petitions



Market News and Updates

Anticipated Patent Expiration(s):[16]

- ZTlido® (lidocaine 1.8% topical system): May 2031

New U.S. Food and Drug Administration Approval(s):

- **August 2020:** The FDA approved Amneal Pharmaceuticals' Abbreviated New Drug Application (ANDA) for their generic lidocaine 5% patch for the treatment of post-herpetic neuralgia. The lidocaine 5% patch is the first commercialized hydrogel product produced by Amneal.[17]

Recommendations

The College of Pharmacy does not recommend any changes to the current lidocaine topical products prior authorization criteria at this time.

Utilization Details of Lidocaine Topical Products: Fiscal Year 2020

PRODUCT UTILIZED	TOTAL CLAIMS	TOTAL MEMBERS	TOTAL COST	COST/ CLAIM	CLAIMS/ MEMBER	% COST
LIDOCAINE PRODUCTS						
LIDOCAINE PAD 5%	2,305	1,180	$187,691.11	$81.43	1.95	100%
TOTAL	**2,350**	**1,180***	**$187,691.11**	**$81.43**	**1.95**	**100%**

PAD = patch
*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021

[16] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available online at: https://www.accessdata.fda.gov/scripts/cder/ob/index.cfm. Last revised 09/2021. Last accessed 09/23/2021.
[17] Amneal Pharmaceuticals, Inc. Amneal Receives Abbreviated New Drug Application Approval for Lidocaine Patch, 5%. Available online at: https://investors.amneal.com/news/press-releases/press-release-details/2020/Amneal-Receives-Abbreviated-New-Drug-Application-Approval-for-Lidocaine-Patch-5/default.aspx. Issued 08/25/2020. Last accessed 09/23/2021.

Fiscal Year 2021 Annual Review of Luxturna® (Voretigene Neparvovec-rzyl)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Luxturna® (Voretigene Neparvovec-rzyl) Approval Criteria:

1. An FDA approved diagnosis of biallelic *RPE65* mutation-associated retinal dystrophy; and
    a. Diagnosis must be confirmed by genetic testing; and
2. Member must have sufficient viable retinal cells in both eyes as determined by the treating physician(s); and
3. Member must have best corrected visual acuity of 20/60 or worse in both eyes and/or visual field <20 degrees in any meridian in both eyes; and
4. Member must be 4 years of age or older; and
5. Member must not have participated in a previous *RPE65* gene therapy study or have previously received treatment with Luxturna®; and
6. Member must not have had intraocular surgery in the past 6 months; and
7. Female members of child bearing age must not be pregnant and must have a negative pregnancy test immediately prior to administration of Luxturna®; and
8. Male and female members of child bearing age must be willing to use effective contraception during treatment with Luxturna® and for at least 4 months after administration of Luxturna®; and
9. Member must take the recommended systemic oral corticosteroid regimen, starting 3 days prior to administration of Luxturna® to each eye, and continuing after administration of Luxturna®, as per the Luxturna® *Prescribing Information*; and
10. Luxturna® must be prescribed and administered by a retinal surgeon with expertise in the treatment of biallelic *RPE65* mutation-associated retinal dystrophy and in the administration of Luxturna® at an Ocular Gene Therapy Treatment Center; and
    a. Luxturna® must be shipped via cold chain supply shipping and delivery to the Ocular Gene Therapy Treatment Center where the member is scheduled to receive treatment; and
    b. Luxturna® must be stored frozen prior to preparation for administration (Luxturna® should be administered within 4 hours of preparation); and

    c. The receiving facility must have a mechanism in place to track patient-specific Luxturna® from receipt to storage to administration; and
11. Luxturna® must be administered subretinally to each eye on separate days within a close interval, but no fewer than 6 days apart; and
    a. The scheduled procedure date for each eye must be provided; and
12. Only 1 single-dose vial per eye will be approved per member per lifetime; and
    a. Each single-dose vial of Luxturna® is to be dispensed immediately prior to the scheduled procedure for the specific eye; or
13. A prior authorization request with patient-specific information may be submitted for consideration of Luxturna® for members not meeting all of the current prior authorization criteria requirements.

Utilization of Luxturna® (Voretigene Neparvovec-rzyl): Fiscal Year 2021

There was no SoonerCare utilization of Luxturna® (voretigene neparvovec-rzyl) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Luxturna® (Voretigene Neparvovec-rzyl)

There were no prior authorization requests submitted for Luxturna® (voretigene neparvovec-rzyl) during fiscal year 2021.

Market News and Updates

Pipeline:

- **GS030:** GenSight is developing GS030, a novel gene therapy for the treatment of retinitis pigmentosa (RP). GS030 uses optogenetics, a biologic technique that involves the transfer of a gene that encodes for a light-sensitive protein, which in turn causes neuronal cells to respond to light stimulation. GS030 consists of 2 complementary components, a gene therapy product encoding a photoactivatable protein, delivered via a modified adeno-associated virus serotype 2 (AAV2) vector, and biomimetic goggles that stimulate the engineered retinal cells (images are projected onto the retina by a light source that uses a specific wavelength). GenSight is currently conducting the Phase 1/2 PIONEER dose-escalation study to evaluate the safety and tolerability of GS030 in patients with RP. In May 2021, GenSight announced the first peer-reviewed publication of a case report demonstrating partial visual function recovery in a blind patient after treatment with GS030 optogenetic therapy. The investigational, injectable optogenetic vector GS030 in conjunction with light-stimulating goggles were utilized. The combination intervention resulted in partially restored visual perception and object localization. The findings of this case study describe the first

ever instance of visual restoration in a patient with neurodegenerative vision loss treated with optogenetic therapy.[18,19,20]

Recommendations

The College of Pharmacy does not recommend any changes to the current Luxturna® (voretigene neparvovec-rzyl) prior authorization criteria at this time.

[18] GenSight Biologics. GenSight Pipeline: GS030 for Retinitis Pigmentosa. Available online at: https://www.gensight-biologics.com/product/gs030-for-retinitis-pigmentosa/. Last accessed 09/23/2021.
[19] GenSight Biologics. GenSight Biologics Announces Nature Medicine Case Report Showing Visual Recovery After GS030 Optogenetic Treatment. Available online at: https://www.gensight-biologics.com/2021/05/25/gensight-biologics-announces-nature-medicine-case-report-showing-visual-recovery-after-gs030-optogenetic-treatment/. Issued 05/25/2021. Last accessed 09/23/2021.
[20] Sahel JA, Boulanger-Scemama E, Pagot C, et al. Partial Recovery of Visual Function in a Blind Patient After Optogenetic Therapy. *Nat Med* 2021; 27(7):1223-1229. doi: 10.1038/s41591-021-01351-4.

Fiscal Year 2021 Annual Review of Mepsevii® (Vestronidase Alfa-vjbk)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Mepsevii® (Vestronidase Alfa-vjbk) Approval Criteria:

1. An FDA approved diagnosis of Sly syndrome (mucopolysaccharidosis VII; MPS VII) confirmed by:
   a. Enzyme analysis demonstrating a deficiency of beta-glucuronidase activity; or
   b. Genetic testing to confirm diagnosis of MPS VII; and
2. Mepsevii® must be administered by a health care professional prepared to manage anaphylaxis; and
3. Initial approvals will be for the duration of 12 months. Reauthorization may be granted if the prescriber documents the member is responding well to treatment; and
4. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to the Mepsevii® *Prescribing Information.*

Utilization of Mepsevii® (Vestronidase Alfa-vjbk): Fiscal Year 2021

There was no SoonerCare utilization of Mepsevii® (vestronidase alfa-vjbk) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Mepsevii® (Vestronidase Alfa-vjbk)

There were no prior authorization requests submitted for Mepsevii® (vestronidase alfa-vjbk) during fiscal year 2021.

Recommendations

The College of Pharmacy does not recommend any changes to the current Mepsevii® (vestronidase alfa-vjbk) prior authorization criteria at this time.

Fiscal Year 2021 Annual Review of Myalept® (Metreleptin)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Myalept® (Metreleptin) Approval Criteria:

1. An FDA approved diagnosis of leptin deficiency in members with congenital or acquired generalized lipodystrophy; and
2. Approvals will not be granted for the following diagnoses:
   a. Metabolic disease without current evidence of generalized lipodystrophy; or
   b. HIV-related lipodystrophy; or
   c. General obesity not associated with congenital leptin deficiency; and
3. Myalept® must be prescribed by an endocrinologist; and
4. Prescriber must agree to test for neutralizing antibodies in patients who experience severe infections or if they suspect Myalept® is no longer effective; and
   a. Baseline hemoglobin A1c (HbA1c), fasting glucose, and fasting triglycerides must be included on prior authorization request; and
   b. Re-approvals will require recent lab values (HbA1c, fasting glucose, and fasting triglycerides) to ensure neutralizing antibodies have not developed; and
5. Prescriber and pharmacy must be enrolled in the Myalept® Risk Evaluation and Mitigation Strategies (REMS) program; and
6. Approvals will be for the duration of 3 months to evaluate compliance and ensure the prescriber is assessing continued efficacy; and
7. A quantity limit of 1 vial per day will apply.

Utilization of Myalept® (Metreleptin): Fiscal Year 2021

Comparison of Fiscal Years

Fiscal Year	*Total Members	Total Claims	Total Cost	Cost/ Claim	Cost/ Day	Total Units	Total Days
2020	1	12	$583,703.70	$48,641.97	$1,621.40	120	360
2021	1	12	$585,053.66	$48,754.47	$1,725.82	113	339
% Change	**0.0%**	**0.0%**	**0.2%**	**0.2%**	**6.4%**	**-5.8%**	**-5.8%**
Change	**0**	**0**	**$1,349.96**	**$112.50**	**$104.42**	**-7**	**-21**

*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2020 = 07/01/2019 to 06/30/2020; Fiscal Year 2021 = 07/01/2020 to 06/30/2021

Demographics of Members Utilizing Myalept® (Metreleptin)

- Due to the limited number of members utilizing Myalept® (metreleptin), detailed demographic information could not be provided.

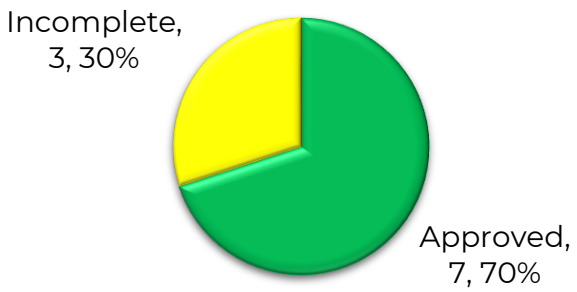
Top Prescriber Specialties of Myalept® (Metreleptin) by Number of Claims

- The only prescriber specialty listed on paid claims for Myalept® (metreleptin) during fiscal year 2021 was pediatric endocrinologist.

Prior Authorization of Myalept® (Metreleptin)

There were 10 prior authorization requests submitted for Myalept® (metreleptin) during fiscal year 2021. The following chart shows the status of the submitted petitions for fiscal year 2021.

Status of Petitions



Market News and Updates

News:

- **March 2021:** Amryt Pharma received positive feedback from the U.S. Food and Drug Administration (FDA) pertaining to the path towards approval of Myalept® for a new indication for patients with partial lipodystrophy (PL). Amryt was informed that the FDA will consider an efficacy supplement for Myalept® based on efficacy and safety data from a randomized, placebo-controlled study in PL patients, with further safety data to be collected over the span of 12 months. Amryt plans to enroll approximately 80 patients with PL with severe metabolic consequences of their disease as reflected by blood glucose control and/or triglyceride levels on optimal background treatment. The Phase 3 study is expected to begin by the end of 2021.[21]

[21] Amryt Pharma Plc. Amryt Receives Positive Feedback from the FDA on the Path Forward for Myalept® (Metreleptin) Indication in Partial Lipodystrophy. *Globe Newswire*. Available online at: https://www.globenewswire.com/en/news-release/2021/03/23/2197342/0/en/Amryt-Receives-Positive-Feedback-from-the-FDA-on-the-Path-Forward-for-Myalept-metreleptin-Indication-in-Partial-Lipodystrophy.html. Issued 03/23/2021. Last accessed 09/23/2021.

Recommendations

The College of Pharmacy does not recommend any changes to the current Myalept® (metreleptin) prior authorization criteria at this time.

Utilization Details of Myalept® (Metreleptin): Fiscal Year 2021

PRODUCT UTILIZED	TOTAL CLAIMS	TOTAL MEMBERS	TOTAL COST	COST/ CLAIM	CLAIMS/ MEMBER	% COST
MYALEPT INJ 11.3MG	12	1	$585,053.66	$48,754.47	12	100%
TOTAL	**12**	**1***	**$585,053.66**	**$48,754.47**	**12**	**100%**

INJ = injection
*Total number of unduplicated utilizing members.
Costs do not reflect rebated prices or net costs.
Fiscal Year 2021 = 07/01/2020 to 06/30/2021

Fiscal Year 2021 Annual Review of Mytesi® (Crofelemer)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Mytesi® (Crofelemer) Approval Criteria:

1. An FDA approved diagnosis of non-infectious diarrhea in adult members with human immunodeficiency virus (HIV)/acquired immunodeficiency syndrome (AIDS) currently on anti-retroviral therapy; and
2. Duration of diarrhea has been ≥4 weeks; and
3. Dietary modifications have failed; and
4. Prescribers must verify that infectious diarrhea has been ruled out via confirmation of all of the following:
    a. CD4 count has been measured and possible opportunistic infections have been ruled out; and
    b. Member does not have fever; and
    c. Stool studies for pathogens are negative including:
        i. Bacterial cultures; and
        ii. Ova, parasite, cryptosporidium and/or giardia; and
        iii. *Clostridium difficile* (*Clostridium difficile* testing should include a glutamate dehydrogenase screen and if positive, followed by a confirmatory test or nucleic acid amplification test in members with documented diarrhea; a toxin enzyme immunoassay should not be used as a stand-alone test); and
5. If stool study results are negative and the member has severe symptoms, particularly in the case of advanced immunodeficiency, an endoscopy with biopsy is recommended, at the prescriber's discretion, to rule out inflammatory bowel disease, cancer, cytomegalovirus (CMV) infection, microsporidium, or mycobacterium avium complex (MAC); and
6. A quantity limit of 60 tablets per 30 days will apply. Initial approvals will be for 4 weeks of therapy. An additional 6-month approval may be granted if the prescriber documents member is responding well to treatment.

Utilization of Mytesi® (Crofelemer): Fiscal Year 2021

There was no SoonerCare utilization of Mytesi® (crofelemer) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Mytesi® (Crofelemer)

There were no prior authorization requests submitted for Mytesi® (crofelemer) during fiscal year 2021.

Market News and Updates

Anticipated Patent Expiration(s):[22]

- Mytesi® (crofelemer): October 2031

Recommendations

The College of Pharmacy does not recommend any changes to the current Mytesi® (crofelemer) prior authorization criteria at this time.

[22] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available online at: https://www.accessdata.fda.gov/scripts/cder/ob/index.cfm. Last revised 09/2021. Last accessed 09/23/2021.

Fiscal Year 2021 Annual Review of Qualaquin® (Quinine Sulfate)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Qualaquin® (Quinine Sulfate) Approval Criteria:

1. An FDA approved diagnosis of malaria; and
2. Off-label use for the prevention/treatment of leg cramps and other related conditions will not be covered.

Utilization of Qualaquin® (Quinine Sulfate): Fiscal Year 2021

There was no SoonerCare utilization of Qualaquin® (quinine sulfate) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Qualaquin® (Quinine Sulfate)

There were 5 prior authorization requests submitted for Qualaquin® (quinine sulfate) during fiscal year 2021, all of which were denied.

Recommendations

The College of Pharmacy does not recommend any changes to the current Qualaquin® (quinine sulfate) prior authorization criteria at this time.

Fiscal Year 2021 Annual Review of Sylvant® (Siltuximab)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Sylvant® (Siltuximab) Approval Criteria:

1. An FDA approved diagnosis of Multicentric Castleman's Disease (also known as giant lymph node hyperplasia); and
2. Member must be Human Immunodeficiency Virus (HIV) and Human Herpesvirus-8 (HHV-8) negative; and
3. Member must be 18 years of age or older; and
4. The following FDA approved dosing restrictions will apply:
    a. 11mg/kg via intravenous (IV) infusion every 3 weeks until treatment failure (defined as disease progression based on increase in symptoms, radiologic progression, or deterioration in performance status); and
5. Sylvant® must be administered in a clinical setting able to provide resuscitation equipment, medications, and trained personnel; and
6. The prescriber must verify that a complete blood count (CBC) will be done prior to each dose for the first 12 months and for an additional 3 doses thereafter; and
7. Approvals will be for the duration of 6 months.

Utilization of Sylvant® (Siltuximab): Fiscal Year 2021

There was no SoonerCare utilization, including pharmacy and medical claims, of Sylvant® (siltuximab) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Sylvant® (Siltuximab)

There were no prior authorization requests submitted for Sylvant® (siltuximab) during fiscal year 2021.

Market News and Updates

Pipeline:

- **Sirolimus:** A Phase 2, single-arm, open-label, multi-center study of sirolimus in previously treated idiopathic Multicentric Castleman's Disease (iMCD) is currently being conducted. Blockade of interleukin 6 (IL-6) signaling with siltuximab or tocilizumab stops symptoms and improves lymphadenopathy in a portion of patients with iMCD; however, 66% of patients in the siltuximab Phase 2 clinical study did not meet response criteria, and recent studies found that IL-6 is not

significantly elevated in many iMCD patients. Recent research has suggested a key role for the phosphoinositide 3-kinase (PI3K)/Akt/mechanistic target of rapamycin (mTOR) pathway in iMCD pathogenesis and off-label administration of sirolimus, an mTOR inhibitor, has shown clinical activity. Based on these experiences, the current study is evaluating the efficacy of sirolimus as a therapy for iMCD patients who are either unable to tolerate IL-6 blockade therapy (siltuximab or tocilizumab), or who fail, relapse, or are refractory to such treatment. The estimated study enrollment is 24 male or female adults 18 to 80 years of age. Patients with iMCD who have failed previous therapy will take daily oral sirolimus (loading dose of 7.5mg/m^2 on day 1 and 2.5mg/m^2/day starting on day 2) for 12 months. The primary outcome measure is the proportion of patients achieving a positive clinical benefit response after 12 months.[23]

Recommendations

The College of Pharmacy does not recommend any changes to the current Sylvant® (siltuximab) prior authorization criteria at this time.

[23] Sirolimus in Previously Treated Idiopathic Multicentric Castleman Disease. *Clinicaltrials.gov*. Available online at: https://clinicaltrials.gov/ct2/show/study/NCT03933904. Last revised 07/29/2021. Last accessed 09/23/2021.

Fiscal Year 2021 Annual Review of Symlin® (Pramlintide)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Symlin® (Pramlintide) Approval Criteria:

1. An FDA approved diagnosis of type 1 or type 2 diabetes; and
2. Member must be using a basal-bolus insulin regimen; and
3. Member must have failed to achieve adequate glycemic control on basal-bolus insulin regimen or are gaining excessive weight on basal-bolus insulin regimen; and
4. Member must be receiving ongoing care under the guidance of a health care professional; and
5. Members meeting any of the following criteria should not be considered for Symlin® (pramlintide) therapy:
    a. Poor compliance with insulin regimen; or
    b. Poor compliance with self-blood glucose monitoring; or
    c. Hemoglobin A1C (HbA1c) >9%; or
    d. Recurrent severe hypoglycemia requiring assistance in the past 6 months; or
    e. Presence of hypoglycemia unawareness; or
    f. Diagnosis of gastroparesis; or
    g. Required use of medications that stimulate gastrointestinal motility; or
    h. Pediatric members 15 years of age or younger.

Utilization of Symlin® (Pramlintide): Fiscal Year 2021

There was no SoonerCare utilization of Symlin® (pramlintide) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Symlin® (Pramlintide)

There were no prior authorization requests submitted for Symlin® (pramlintide) during fiscal year 2021.

Recommendations

The College of Pharmacy does not recommend any changes to the current Symlin® (pramlintide) prior authorization criteria at this time.

Fiscal Year 2021 Annual Review of Vimizim® (Elosulfase Alfa)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Vimizim® (Elosulfase Alfa) Approval Criteria:

1. An FDA approved diagnosis of Morquio A syndrome (mucopolysaccharidosis type IVA; MPS IVA) confirmed by:
   a. Enzyme assay demonstrating a deficiency of N-acetylgalactosamine-6-sulfatase (GALNS) enzyme activity; or
   b. Molecular genetic testing to confirm biallelic pathogenic variants in GALNS; and
2. Vimizim® must be administered by a health care professional prepared to manage anaphylaxis; and
3. Initial approvals will be for the duration of 12 months. Reauthorization may be granted if the prescriber documents the member is responding well to treatment; and
4. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to package labeling.

Utilization of Vimizim® (Elosulfase Alfa): Fiscal Year 2021

There was no SoonerCare utilization, including pharmacy and medical claims, of Vimizim® (elosulfase alfa) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Vimizim® (Elosulfase Alfa)

There were no prior authorization requests submitted for Vimizim® (elosulfase alfa) during fiscal year 2021.

Recommendations

The College of Pharmacy does not recommend any changes to the current Vimizim® (elosulfase alfa) prior authorization criteria at this time.

Fiscal Year 2021 Annual Review of Xuriden® (Uridine Triacetate)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Xuriden® (Uridine Triacetate) Approval Criteria:

1. An FDA approved diagnosis of hereditary orotic aciduria defined by at least 1 of the following:
    a. Assay of the orotate phosphoribosyltransferase and orotidylic acid decarboxylase enzymes in the member's erythrocytes showing deficiency in both enzymes or deficiency in orotidylic acid decarboxylase alone; or
    b. Evidence of megaloblastic anemia; and
        i. Normal serum folate and vitamin B12 levels and no evidence of transcobalamine II deficiency; or
    c. Orotic acid crystals visualized in the urine via microscopy; and
2. The member's current weight must be provided on the prior authorization request; and
    a. Weights should be reassessed every 6 months to ensure proper dosing and effectiveness; or
    b. Prescriber can indicate urine orotic acid levels are within normal ranges and dosing remains appropriate; and
3. The prescriber must verify the member or caregiver is able to properly measure and administer medication; and
4. A quantity limit of 4 packets per day will apply.

Utilization of Xuriden® (Uridine Triacetate): Fiscal Year 2021

There was no SoonerCare utilization of Xuriden® (uridine triacetate) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Xuriden® (Uridine Triacetate)

There were no prior authorization requests submitted for Xuriden® (uridine triacetate) during fiscal year 2021.

Market News and Updates

Anticipated Patent Expiration(s):[24]

- Xuriden® (uridine triacetate): July 2023

Recommendations

The College of Pharmacy does not recommend any changes to the current Xuriden® (uridine triacetate) prior authorization criteria at this time.

[24] U.S. Food and Drug Administration (FDA). Orange Book: Approved Drug Products with Therapeutic Equivalence Evaluations. Available online at: https://www.accessdata.fda.gov/scripts/cder/ob/index.cfm. Last revised 09/2021. Last accessed 09/23/2021.

Fiscal Year 2021 Annual Review of Zinplava™ (Bezlotoxumab)

Oklahoma Health Care Authority
Fiscal Year 2021 Print Report

Current Prior Authorization Criteria

Zinplava™ (Bezlotoxumab) Approval Criteria:

1. An FDA approved diagnosis of *Clostridium difficile* infection (CDI) in members 18 years of age or older who are receiving antibacterial drug treatment of CDI and are at a high risk for CDI recurrence; and
    a. Prescriber must document the member has ≥1 of the following risk factor(s) for high risk of CDI recurrence:
        i. Age 65 years or older; or
        ii. ≥1 episode(s) of CDI within the 6 months prior to the episode under treatment; or
        iii. Need for ongoing therapy with concomitant antibiotics during treatment for CDI; or
        iv. Severe underlying medical disorders; or
        v. Immunocompromised; or
        vi. Clinically severe CDI (Zar score ≥2); and
2. Current or planned antibacterial drug for CDI must be provided on the prior authorization request to ensure medication is within standard of care; and
3. Prescriber must document that Zinplava™ (bezlotoxumab) will be administered while the member is receiving antibacterial drug treatment of CDI; and
4. The member's recent weight must be provided on the prior authorization request in order to authorize the appropriate amount of drug required according to package labeling.

Utilization of Zinplava™ (Bezlotoxumab): Fiscal Year 2021

There was no SoonerCare utilization of Zinplava™ (bezlotoxumab) during fiscal year 2021 (07/01/2020 to 06/30/2021).

Prior Authorization of Zinplava™ (Bezlotoxumab)

There were no prior authorization requests submitted for Zinplava™ (bezlotoxumab) during fiscal year 2021.

Market News and Updates

Pipeline:

- **LMN-Cdiff01**: In September 2021, Lumen Bioscience announced the launch of a Phase 1 study of LMN-Cdiff01, an oral biologic drug aimed at the prevention and treatment of recurrent *Clostridium difficile* infection (CDI). The exploratory study will investigate the delivery of LMN-Cdiff01 via enteric capsules which will release their contents at or before the terminal ileum. Study participant recruitment will draw from healthy individuals with mature ileostomies. Preliminary results of the study will show if capsule content release proceeds as expected. Results are forecasted for December 2021.[25]

Recommendations

The College of Pharmacy does not recommend any changes to the current Zinplava™ (bezlotoxumab) prior authorization criteria at this time.

[25] Lumen Bioscience. Lumen Bioscience Launches Phase 1 Trial of Oral Biologic Drug for Preventing C. difficile Infection Recurrence. *BioSpace*. https://www.biospace.com/article/releases/lumen-bioscience-launches-phase-1-trial-of-oral-biologic-drug-for-preventing-c-difficile-infection-recurrence/?keywords=c+difficile. Issued 09/01/2021. Last accessed 09/24/2021.