TX CLINICAL CRITERIA & PROCEDURE

CRITERIA NAME: Enzyme Replacement Therapy (Fabrazyme, Lumizyme, Nexviazyme, Vimizim, Naglazyme, Elaprase, Cerezyme, Aldurazyme, Ceprotin, Kanuma, Elelyso, VPRIV, Xenpozyme, Elfabrio, Adzynma)	**CRITERIA ID:** TX.CC.PHAR.33
BUSINESS UNIT: Superior HealthPlan	**FUNCTIONAL AREA:** Pharmacy, Medical Directors
EFFECTIVE DATE: 01/24/2022	**PRODUCT(S):** STAR, STAR Health, STAR Kids, Star Plus, CHIP, CHIP Perinate
REVIEWED/REVISED DATE: 01/24/2022, 01/24/2023, 03/28/2023, 04/28/2023, 4/3/2024, 4/15/2024, 8/1/2024	
REGULATOR MOST RECENT APPROVAL DATE(S): N/A	

CRITERIA STATEMENT:
The purpose of this clinical criteria is to provide a guide to medical necessity reviews for Enzyme Replacement Therapy (Fabrazyme, Lumizyme, Nexviazyme, Vimizim, Naglazyme, Elaprase, Cerezyme, Aldurazyme, Ceprotin, Kanuma, Elelyso, VPRIV, Xenpozyme, Elfabrio, Adzynma).

PURPOSE:
Consistent with the regulation at 42 CFR Section 438.210 and 42 CFR Section 457.1230(d), services covered under managed care contracts, including clinician-administered drugs, must be furnished in an amount, duration, and scope that is no less than the amount, duration, and scope for the same services specified in the state plan. While MCOs may place appropriate limits on drugs, MCOs may not use a standard for determining medical necessity that is more restrictive than what is used in the state plan, i.e., developed by the Vendor Drug Program. For example, if a member is denied a clinician administered drug in managed care because of the MCO's prior authorization criteria, but would have received the drug under the criteria specified in the state plan, then the MCO's prior authorization criteria would violate the amount, duration, and scope requirements cited above. HHSC intends to amend the Managed Care Contracts at the next opportunity to include this requirement. This same standard applies to CHIP formulary and CAD coverage.

Refer to the Outpatient Drug Services Handbook of the Texas Medicaid Provider Procedure Manual for more details on the clinical criteria and prior authorization requirements.

SCOPE:
This criteria applies to all directors, officers, and employees of Centene Corporation, its affiliates, health plans, and subsidiary companies (collectively, the "Company").

DEFINITIONS: N/A

POLICY:
It is the policy of Superior HealthPlan (SHP) and Centene Pharmacy Services (CPS) to follow state guidance for medical necessity review of Enzyme Replacement Therapy (Fabrazyme, Lumizyme, Nexviazyme, Vimizim, Naglazyme, Elaprase, Cerezyme, Aldurazyme, Ceprotin, Kanuma, Elelyso, Vpriv, Xenpozyme, Elfabrio, Adzynma).

PROCEDURE:
Provider must submit documentation (which may include office chart notes and lab results) supporting that member has met all approval criteria.

I. Initial Approval Criteria

A. FDA Approved Indications

1. Prior authorization approval for any of the enzyme replacement therapy listed below will be considered when the following criteria are met (a and b):
    a. A request for the specific enzyme replacement therapy
    b. The laboratory evidence of the enzyme deficiency. See Appendix A with for examples of applicable diagnostic confirming labs.
2. Specific for olipudase alfa-rpcp (Xenpozyme) requests, in addition to 1.a. and b. (above), client must also meet the following criteria:

a. Client has a diagnosis of non-central nervous system manifestations of acid sphingomyelinase deficiency (ASMD) in adult and pediatric clients - (diagnosis code: E75.241 and E75.244)
b. Verification of pregnancy status for female clients of reproductive potential prior to therapy initiation
c. Documentation of client's baseline transaminase level
d. Prescriber attestation to check alanine transaminase (ALT) and aspartate aminotransferase (AST) levels within one month prior to initiation of Xenpozyme; within 72 hours prior to any infusion during dose escalation, or prior to the next scheduled Xenpozyme infusion upon resuming treatment following a missed dose.

3. Listed below are the FDA approved indications, age restrictions and diagnosis codes (as applicable):
    a. Agalsidase beta (Fabrazyme) is indicated in clients 2 years of age and older with Fabry disease - (diagnosis code: E75.21).
    b. Alglucosidase alfa (Lumizyme) is indicated for clients with Pompe disease (GAA deficiency) – (diagnosis code: E74.02).
    c. Avalglucosidase alfa-ngpt (Nexviazyme) is indicated for clients 1 year of age and older with late onset Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency) – (diagnosis code: E74.02).
    d. Elosulfase alfa (Vimizim) is a hydrolytic lysosomal glycosaminoglycan (GAG) specific enzyme indicated for clients 5 years of age and older with mucopolysaccharidosis type IVA – (diagnosis code: E76.210).
    e. Galsulfase (Naglazyme) is a hydrolytic lysosomal glycosaminoglycan (GAG) specific enzyme indicated for clients with mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome) – (diagnosis codes: E76.29).
    f. Idursulfase (Elaprase) is a hydrolytic lysosomal glycosaminoglycan (GAG) specific enzyme indicated for clients with Hunter syndrome (mucopolysaccharidosis II, MPS II) – (diagnosis code: E76.1).
    g. Imiglucerase (Cerezyme) is indicated for long-term enzyme replacement therapy for clients 2 years of age and older with a confirmed diagnosis of Type 1 Gaucher disease - (diagnosis code: E75.22) that results in one or more of the following conditions:
        i. Anemia
        ii. Thrombocytopenia
        iii. Bone disease
        iv. Hepatomegaly or splenomegaly
    h. Laronidase (Aldurazyme) is indicated in clients with Hurler and Hurler-Scheie forms of mucopolysaccharidosis I (MPS I) and clients with the Scheie form who have moderate to severe symptoms – (diagnosis codes: E76.01, E76.02, and E76.03).
    i. Pegunigalsidase alfa-iwxj (Elfabrio) is indicated for the treatment of adult clients with confirmed Fabry disease – (diagnosis code: E75.21).
    j. Protein C concentrate, human (Ceprotin) is indicated in pediatric and adult clients with severe congenital protein C deficiency for the prevention and treatment of venous thrombosis and purpura fulminans – (diagnosis code: D68.59).
    k. Sebelipase alfa (Kanuma) is indicated for the treatment of pediatric and adult clients with a diagnosis of Lysosomal Acid Lipase (LAL) deficiency – (diagnosis code: E75.5).
    l. Taliglucerace alfa (Elelyso) is indicated for long-term enzyme replacement therapy for adult clients with a diagnosis of Type 1 Gaucher disease – (diagnosis code: E75.22).
    m. Velaglucerase alfa (VPRIV) is indicated for long-term enzyme replacement therapy for pediatric and adult clients with Type 1 Gaucher disease – (diagnosis code: E75.22).
    n. Olipudase alfa-rpcp (Xenpozyme) is indicated to treat non-central nervous system manifestations of acid sphingomyelinase deficiency (ASMD) in adult and pediatric clients - (diagnosis codes: E75.241 and E75.244).
    o. Apadamtase alfa (Adzynma) is indicated for prophylactic or on-demand enzyme replacement treatment for congenital thrombotic thrombocytopenic purpura (cTTP) in adult and pediatric clients – (diagnosis code: D69.42).

Approval duration: 6 months

Appendix A.

Enzyme replacement therapy	Indication	Diagnostic Lab Examples
Agalsidase beta (Fabrazyme)	Fabry disease	Enzyme assay demonstrating a deficiency of alpha-galactosidase activity OR DNA testing
Alglucosidase alfa (Lumizyme)	Pompe disease	Enzyme assay confirming low GAA activity OR DNA testing
Avalglucosidase alfa-ngpt (Nexviazyme)	Pompe disease	Enzyme assay confirming low GAA activity OR DNA testing
Elosulfase alfa (Vimizim)	Mucopolysaccharidosis type IVA	Enzyme assay demonstrating a deficiency of N-acetylgalactosamine-6-sulfatase activity OR DNA testing
Galsulfase (Naglazyme)	Mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome)	Enzyme assay demonstrating a deficiency in N-acetylgalactosamine 4-sulfatase (arylsulfatase B) activity OR DNA testing
Idursulfase (Elaprase)	Hunter syndrome (Mucopolysaccharidosis II, MPS II)	Enzyme assay demonstrating a deficiency of iduronate 2-sulfatase activity OR DNA testing
Imiglucerase (Cerezyme)	Type 1 Gaucher disease	Enzyme assay demonstrating a deficiency of beta-glucocerebrosidase (glucosidase) activity OR DNA testing
Laronidase (Aldurazyme)	Mucopolysaccharidosis I: Hurler, Hurler-Scheie, and Scheie Forms	Enzyme assay demonstrating deficiency of alpha-L-iduronidase activity OR DNA testing
Protein C Concentrate, human (Ceprotin)	congenital Protein C deficiency	Lab result confirming low protein C activity (due to low protein C levels or function or both)
Sebelipase alfa (Kanuma)	Lysosomal Acid Lipase (LAL) deficiency	Enzyme assay demonstrating a deficiency of LAL activity OR Lipase A - lysosomal acid type (LIPA) gene mutation
Taliglucerace alfa (Elelyso)	Type 1 Gaucher disease	Enzyme assay demonstrating a deficiency of beta-glucocerebrosidase (glucosidase) activity OR DNA testing
Velaglucerase alfa (VPRIV)	Type 1 Gaucher disease	Enzyme assay demonstrating a deficiency of beta-glucocerebrosidase (glucosidase) activity OR DNA testing
Olipudase alfa-rpcp (Xenpozyme)	Acid Sphingomyelinase Deficiency (ASMD)	Enzyme assay confirming decreased acid sphingomyelinase (ASM) enzyme activity OR DNA testing
Pegunigalsidase alfa-iwxj (Elfabrio)	Fabry Disease	Enzyme assay demonstrating a deficiency of alpha-galactosidase activity OR DNA testing
Apadamtase alfa (Adzynma)	Congenital thrombotic thrombocytopenic purpura (cTTP)	ADAMTS13 activity assay to measure activity of ADAMTS13 enzyme OR ADAMTS13 antigen test to measure amount of ADAMTS13 protein

REFERENCES:
Texas Medicaid Provider Procedure Manual: Outpatient Drug Services Handbook

ATTACHMENTS: N/A

REVISION LOG

REVISION TYPE	REVISION SUMMARY	DATE APPROVED & PUBLISHED
New Policy		01/24/2022
Ad Hoc Review	Annual Review: Corrected chemical drug name: Avalglucosidase alfa-ngpt (Nexviazyme). Verbiage	01/24/2023

	change: ‘age “xxx” years and older’ for consistency throughout policy.	
Ad Hoc Review	Formatted to new template. Per HHSC guidance, added prior auth requirements for olipudase alfa-rpcp (Xenpozyme): The client has a diagnosis of acid sphingomyelinase deficiency (diagnosis code E75.241 and E75.244); Verification of pregnancy status for female clients of reproductive potential prior to therapy initiation; Documentation of baseline transaminase assessment prior to treatment with Xenpozyme; Prescriber attestation to check alanine transaminase (ALT) and aspartate aminotransferase (AST) levels before initiation of therapy, during any dose escalations of therapy and before any missed doses.	03/28/2023
Ad Hoc Review	Updated verbiage in Section I. A. 2.a. and 2.d. to align with verbiage in CAD Manual	04/28/2023
Ad Hoc Review	Per HHSC guidance, added prior auth requirements for Velmanase alfa-tycv (Lamzede): Pregnancy status of female clients of reproductive potential must be verified prior to initiating treatment. Updated criteria for I.A.4 to include Lamzede and Elfabrio Updated to TX.CC.PHAR format template Added Centene copyright statement Removed Brineura from the criteria	4/3/2024
Ad Hoc Review	Per HHSC guidance, added prior auth requirements for cipaglucosidase alfa-atga (Pombiliti) Updated criteria step I.A.4 to I.A.5 Updated criteria for I.A.5 to include Adzynma and Pombiliti	4/15/2024
Ad hoc review	Removal and conversion of Lamzede and Pombiliti to their individual criteria	8/1/2024