Texas Prior Authorization Program
Clinical Criteria

Drug/Drug Class

Enzymes

- **Aldurazyme**
- **Ceprotin**
- **Elaprase**
- **Fabrazyme**
- **Galafold**
- **Naglazyme**
- **Nityr / Orfadin**
- **Revcovi**
- **Strensiq**
- **Vimizim**

Note: Click the hyperlink to navigate directly to that section.

Clinical Criteria Information Included in this Document

- **Drugs requiring prior authorization**: the list of drugs requiring prior authorization for this clinical criteria
- **Prior authorization criteria logic**: a description of how the prior authorization request will be evaluated against the clinical criteria rules
- **Logic diagram**: a visual depiction of the clinical criteria logic
- **Supporting tables**: a collection of information associated with the steps within the criteria (diagnosis codes, procedure codes, and therapy codes); provided when applicable
- **References**: clinical publications and sources relevant to this clinical criteria

Revision Notes

Annual review by staff

Removed criteria for Lumizyme – no longer on formulary

Updated references

Enzymes
Aldurazyme
Drugs Requiring Prior Authorization

The listed GCNS may not be an indication of TX Medicaid Formulary coverage. To learn the current formulary coverage, visit TxVendorDrug.com/formulary/formulary-search.

Drugs Requiring Prior Authorization	
Label Name	**GCN**
ALDURAZYME 2.9MG/5ML VIAL	19585

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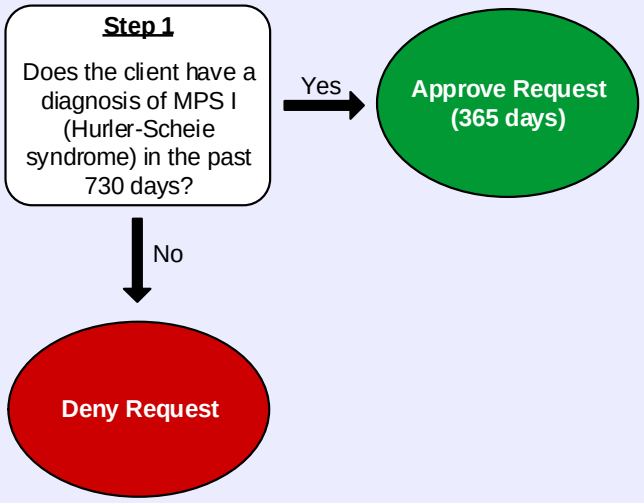
Enzymes
Aldurazyme
Clinical Criteria Logic

1. Does the client have a **diagnosis of mucopolysaccharidosis I** (also called MPS I and/or Hurler-Scheie syndrome) in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Aldurazyme
Clinical Criteria Logic Diagram



Enzymes
Ceprotin
Drugs Requiring Prior Authorization

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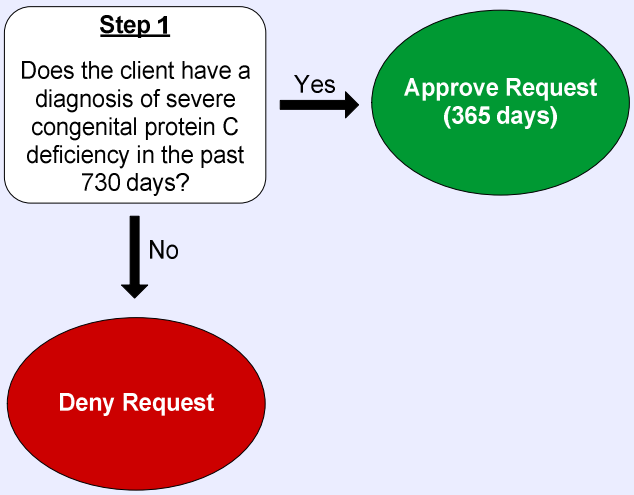
Drugs Requiring Prior Authorization	
Label Name	**GCN**
CEPROTIN 8002-1,200 UNITS VIAL	15483

Enzymes
Ceprotin
Clinical Criteria Logic

1. Does the client have a **diagnosis of severe congenital protein C deficiency** in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Ceprotin
Clinical Criteria Logic Diagram



Enzymes
Elaprase
Drugs Requiring Prior Authorization

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Drugs Requiring Prior Authorization	
Label Name	**GCN**
ELAPRASE 6MG/3ML VIAL	97047

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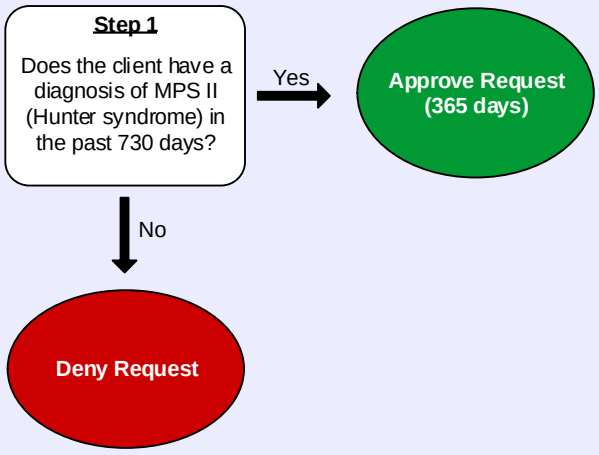
Enzymes
Elaprase
Clinical Criteria Logic

1. Does the client have a **diagnosis of mucopolysaccharidosis II** (Hunter syndrome) in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Elaprase
Clinical Criteria Logic Diagram



Enzymes
Fabrazyme
Drugs Requiring Prior Authorization

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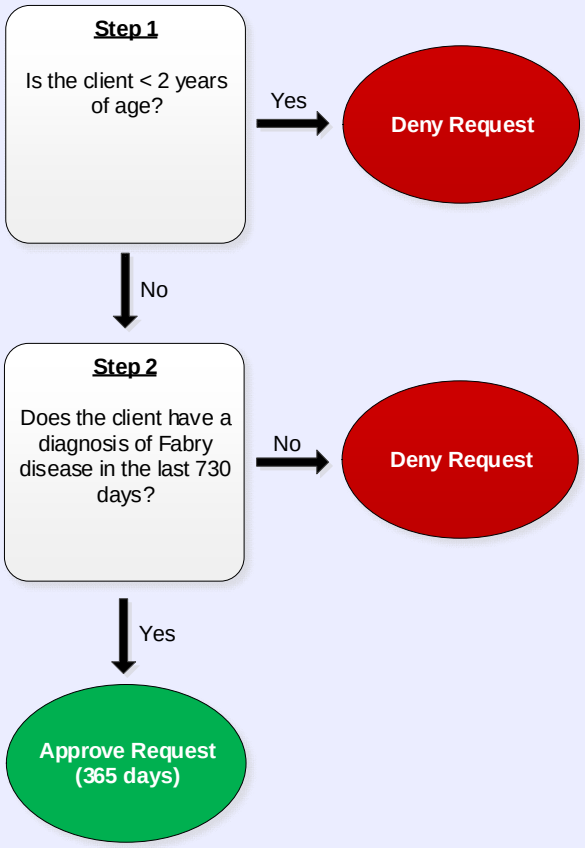
Drugs Requiring Prior Authorization	
Label Name	**GCN**
FABRAZYME 5MG VIAL	22348
FABRAZYME 35MG VIAL	18997

Enzymes
Fabrazyme
Clinical Criteria Logic

1. Is the client less than (<) 2 years of age?
   [ ] Yes (Deny)
   [ ] No (Go to #2)

2. Does the client have a **diagnosis of Fabry disease** in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Fabrazyme
Clinical Criteria Logic Diagram



Enzymes
Galafold
Drugs Requiring Prior Authorization

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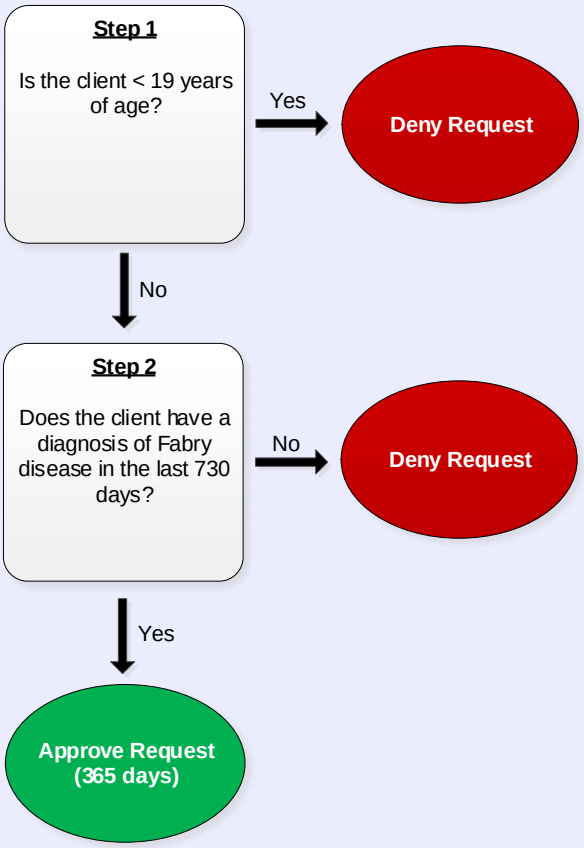
Drugs Requiring Prior Authorization	
Label Name	**GCN**
GALAFOLD 123 MG CAPSULE	43641

Enzymes
Galafold
Clinical Criteria Logic

1. Is the client less than (<) 19 years of age?
   [ ] Yes (Deny)
   [ ] No (Go to #2)

2. Does the client have a **diagnosis of Fabry disease** in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Galafold
Clinical Criteria Logic Diagram



Enzymes
Naglazyme
Drugs Requiring Prior Authorization

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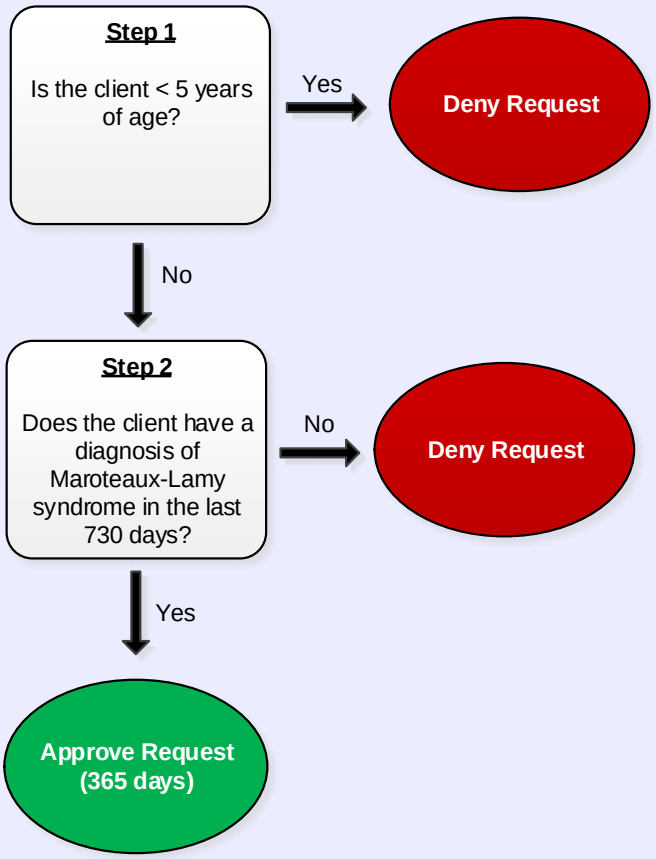
Drugs Requiring Prior Authorization	
Label Name	**GCN**
NAGLAZYME 5MG/5ML VIAL	24744

Enzymes
Naglazyme
Clinical Criteria Logic

1. Is the client less than (<) 5 years of age?
   [ ] Yes (Deny)
   [ ] No (Go to #2)

2. Does the client have a **diagnosis of mucopolysaccharidosis VI** (MPS VI, Maroteaux-Lamy syndrome) in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Naglazyme
Clinical Criteria Logic Diagram



Enzymes
Nityr / Orfadin
Drugs Requiring Prior Authorization

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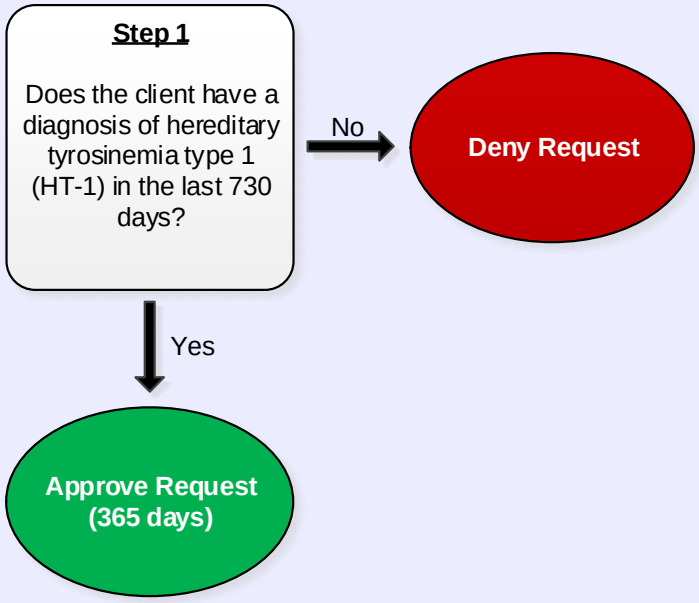
Drugs Requiring Prior Authorization	
Label Name	**GCN**
NITYR 2 MG TABLET	43007
NITYR 5 MG TABLET	43006
NITYR 10 MG TABLET	43008
ORFADIN 2 MG CAPSULE	15662
ORFADIN 5 MG CAPSULE	15663
ORFADIN 10 MG CAPSULE	15664
ORFADIN 20 MG CAPSULE	39031
ORFADIN 4 MG/ML SUSPENSION	41268

Enzymes
Nityr / Orfadin
Clinical Criteria Logic

1. Does the client have a **diagnosis of hereditary tyrosinemia type 1** (HT-1) in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Nityr / Orfadin
Clinical Criteria Logic Diagram



Enzymes
Revcovi
Drugs Requiring Prior Authorization

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Drugs Requiring Prior Authorization	
Label Name	**GCN**
REVCOVI 2.4 MG/1.5 ML VIAL	45525

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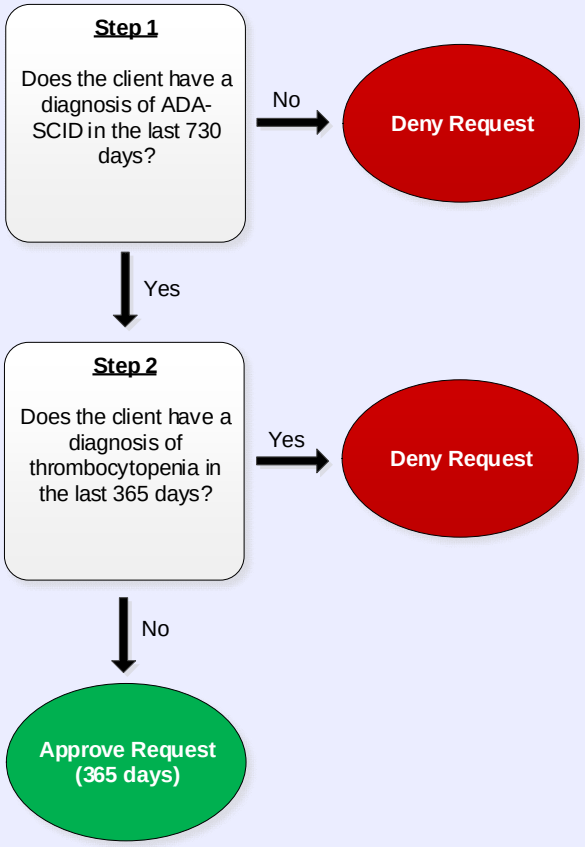
Enzymes
Revcovi

Clinical Criteria Logic

1. Does the client have a **diagnosis of adenosine deaminase severe combined immunodeficiency disease (ADA-SCID)** in the last 730 days?
   [ ] Yes (Go to #2)
   [ ] No (Deny)

2. Does the client have a diagnosis of **thrombocytopenia** in the last 365 days?
   [ ] Yes (Deny)
   [ ] No (Approve – 365 days)

Enzymes
Revcovi
Clinical Criteria Logic Diagram



Enzymes
Strensiq
Drugs Requiring Prior Authorization

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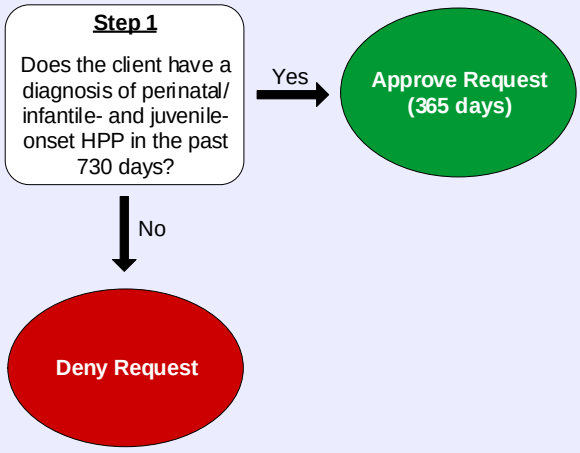
Drugs Requiring Prior Authorization	
Label Name	**GCN**
STRENSIQ 18 MG/0.45 ML VIAL	39938
STRENSIQ 28 MG/0.7 ML VIAL	39939
STRENSIQ 40 MG/ML VIAL	39857
STRENSIQ 80 MG/0.8 ML VIAL	39858

Enzymes
Strensiq
Clinical Criteria Logic

1. Does the client have a **diagnosis of perinatal/infantile- and juvenile-onset hypophosphatasia (HPP)** in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Strensiq
Clinical Criteria Logic Diagram



Enzymes
Vimizim
Drugs Requiring Prior Authorization

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Drugs Requiring Prior Authorization	
Label Name	**GCN**
VIMIZIM 5 MG/5 ML VIAL	36083

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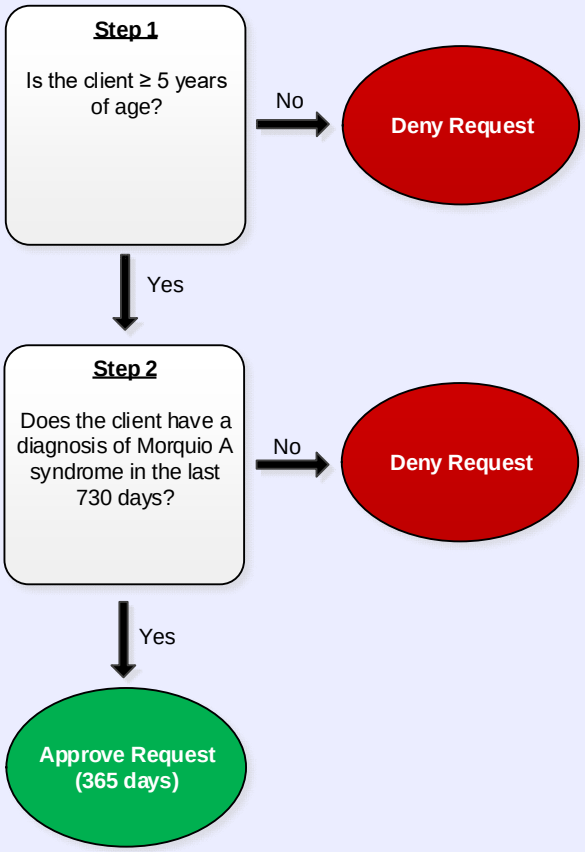
Enzymes
Vimizim
Clinical Criteria Logic

1. Is the client greater than or equal to (≥) 5 years of age?
   [ ] Yes (Go to #2)
   [ ] No (Deny)

2. Does the client have a **diagnosis of mucopolysaccharidosis IVA** (also called Morquio A syndrome) in the past 730 days?
   [ ] Yes (Approve – 365 days)
   [ ] No (Deny)

Enzymes
Vimizim
Clinical Criteria Logic Diagram



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Clinical Criteria Supporting Tables

Diagnosis of mucopolysaccharidosis I Required diagnosis: *1* Look back timeframe: *730 days*	
ICD-10 Code	**Description**
E7601	HURLER'S SYNDROME
E7602	HURLER-SCHEIE SYNDROME
E7603	SCHEIE'S SYNDROME

Diagnosis of adenosine deaminase severe combined immunodeficiency disease Required diagnosis: *1* Look back timeframe: *730 days*	
ICD-10 Code	**Description**
D8131	SEVERE COMBINED IMMUNODEFICIENCY DUE TO ADENOSINE DEAMINASE

Diagnosis of severe congenital protein C deficiency Required diagnosis: *1* Look back timeframe: *730 days*	
ICD-10 Code	**Description**
D6859	OTHER PRIMARY THROMBOPHILIA

Diagnosis of mucopolysaccharidosis II (Hunter Syndrome) Required diagnosis: *1* Look back timeframe: *730 days*	
ICD-10 Code	**Description**
E761	MUCOPOLYSACCHARIDOSIS, TYPE II (HUNTER SYNDROME)

Diagnosis of Fabry disease **Required diagnosis:** *1* **Look back timeframe:** *730 days*	
ICD-10 Code	**Description**
E7521	FABRY (-ANDERSON) DISEASE

Diagnosis of Pompe disease **Required diagnosis:** *1* **Look back timeframe:** *730 days*	
ICD-10 Code	**Description**
E7402	POMPE DISEASE

Diagnosis of perinatal/infantile- and juvenile-onset hypophosphatasia (HPP) **Required diagnosis:** *1* **Look back timeframe:** *730 days*	
ICD-10 Code	**Description**
E8331	FAMILIAL HYPOPHOSPHATEMIA
E8339	OTHER DISORDERS OF PHOSPHOROUS METABOLISM

Diagnosis of Maroteaux-Lamy Syndrome **Required diagnosis:** *1* **Look back timeframe:** *730 days*	
ICD-10 Code	**Description**
E7629	OTHER MUCOPOLYSACCHARIDOSES

Diagnosis of hereditary tyrosinemia type 1 (HT-1) **Required diagnosis:** *1* **Look back timeframe:** *730 days*	
ICD-10 Code	**Description**
E7021	TYROSINEMIA

Diagnosis of thrombocytopenia **Required diagnosis:** *1* **Look back timeframe:** *365 days*	
ICD-10 Code	**Description**
D693	IMMUNE THROMBOCYTOPENIC PURPURA

Diagnosis of thrombocytopenia **Required diagnosis:** *1* **Look back timeframe:** *365 days*	
ICD-10 Code	**Description**
D694	OTHER PRIMARY THROMBOCYTOPENIA
D6941	EVANS SYNDROME
D6942	CONGENITAL AND HEREDITARY THROMBOCYTOPENIA PURPURA
D6949	OTHER PRIMARY THROMBOCYTOPENIA
D695	SECONDARY THROMBOCYTOPENIA
D6951	POSTTRANSFUSION PURPURA
D6959	OTHER SECONDARY THROMBOCYTOPENIA
D696	THROMBOCYTOPENIA, UNSPECIFIED
D698	OTHER SPECIFIED HEMORRHAGIC CONDITIONS

Diagnosis of mucopolysaccharidosis IVA (Morquio A syndrome) **Required diagnosis:** *1* **Look back timeframe:** *730 days*	
ICD-10 Code	**Description**
E76210	MORQUIO A MUCOPOLYSACCHARIDOSES

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Enzymes

Clinical Criteria References

1. Aldurazyme prescribing information. Cambridge, MA. Genzyme Corporation. December 2019.
2. Ceprotin Prescribing Information. Lexington, MA. Baxalta US Inc. August 2021.
3. Elaprase Prescribing Information. Lexington, MA. Takeda Pharmaceuticals USA, Inc. September 2021.
4. Fabrazyme prescribing information. Cambridge, MA. Genzyme Corporation. March 2021.
5. Lumizyme Prescribing Information. Cambridge, MA. Genzyme Corporation. March 2021.
6. Naglazyme prescribing information. Novato, CA. BioMarin Pharmaceutical Inc. December 2019.
7. Nityr prescribing information. Cambridge, United Kingdom. Cycle Pharmaceuticals Ltd. June 2021.
8. Orfadin prescribing information. Waltham, MA. Swedish Orphan Biovitrum. May 2019.
9. Vimizim prescribing information. Novato, CA. BioMarin Pharmaceutical Inc. December 2019.
10. 2014 ICD-10-CM Diagnosis Codes, Volume 1. 2014. Available at **http://icd10data.com**. Accessed on March 26, 2014.
11. Clinical Pharmacology [online database]. Tampa, FL: Elsevier/Gold Standard, Inc.; 2022. Available at www.clinicalpharmacology.com. Accessed on July 13, 2022.
12. Micromedex [online database]. Available at www.micromedexsolutions.com. Accessed on July 13, 2022.
13. Revcovi Prescribing Information. Cary, NC. Chiesi USA, Inc. April 2021.
14. Galafold Prescribing Information. Philadelphia, PA. Amicus Therapeutics US, LLC. December 2021.

Publication History

The Publication History records the publication iterations and revisions to this document. Notes for the *most current revision* are also provided in the **Revision Notes** on the first page of this document.

Publication Date	Notes
05/16/2014	Initial publication and posting to website
08/11/2017	Annual review by staff Added criteria logic and diagram for Carbaglu, pages 10 and 11 Added criteria logic and diagram for Ravicti, pages 28 and 29 Added criteria logic and diagram for Vimizim, pages 31 and 32 Added diagnoses codes for hyperammonemia due to NAGS deficiency, page 33 Added diagnoses codes for urea cycle disorders and Morquio A syndrome, page 36 Updated references, page 37
11/22/2017	Added criteria logic and diagram for Orfadin, pages 27-29 Updated references, page 40
12/06/2017	Updated criteria logic and logic diagram for Fabrazyme, pages 19-20 Added age check to criteria logic and logic diagram for Vimizim, pages 34-35
12/27/2017	Updated criteria logic for Elaprase, page 16
03/30/2018	Added GCNs for Nityr to 'Drugs Requiring PA', page 27 Updated references, page 40
03/28/2019	Updated to include formulary statement (The listed GCNS may not be an indication of TX Medicaid Formulary coverage. To learn the current formulary coverage, visit TxVendorDrug.com/formulary/formulary-search.) on each 'Drug Requiring PA' table Removed criteria for Carbaglu and Ravicti – these agents are now included in the Urea Cycle Disorder criteria
09/11/2019	Added GCN for Revcovi to existing Adagen criteria Removed age requirement for Adagen criteria
04/05/2021	Removed Adagen GCN (33971) – product has been discontinued

Publication Date	Notes
	Updated ICD-10 code for ADA-SCID for Revcovi Removed GCN for Ceprotin 400-600 units vial (15482) – not on formulary Updated age to ≥ 2 years for Fabrazyme Updated references
04/15/2021	Annual review by staff Added criteria for Galafold (43641) Added criteria for Strensiq (39938, 39939, 39857, 39858) Updated references
07/13/2022	Annual review by staff Removed criteria for Lumizyme – no longer on formulary Updated references

