

## Texas Prior Authorization Program Clinical Criteria

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### 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

Added GCNs for nitisinone (15664, 15662, 39031, 15663) to drugs requiring PA (Nityr/Orfadin Table)

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](https://TxVendorDrug.com/formulary/formulary-search).

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

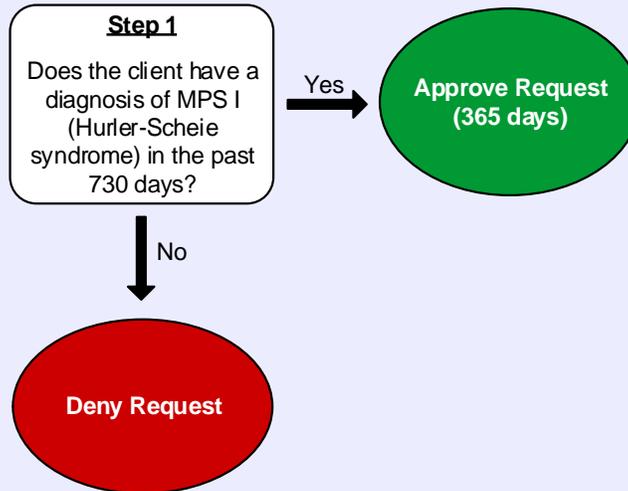


**Enzymes**  
**Aldurazyme**  
**Clinical Criteria Logic**

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

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](http://TxVendorDrug.com/formulary/formulary-search).

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



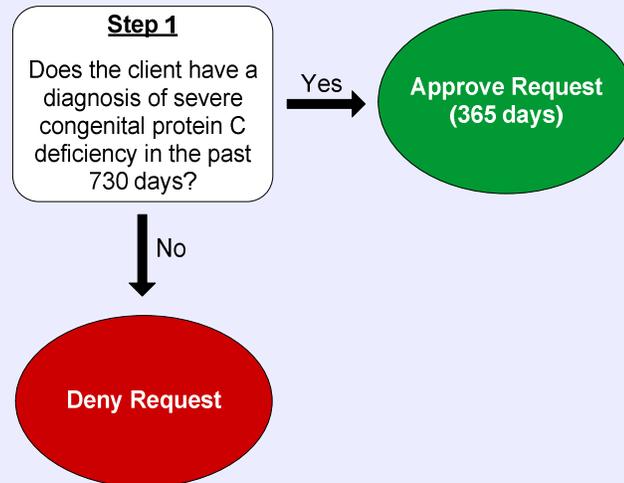
**Enzymes**  
**Ceprotin**  
**Clinical Criteria Logic**

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

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](http://TxVendorDrug.com/formulary/formulary-search).

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



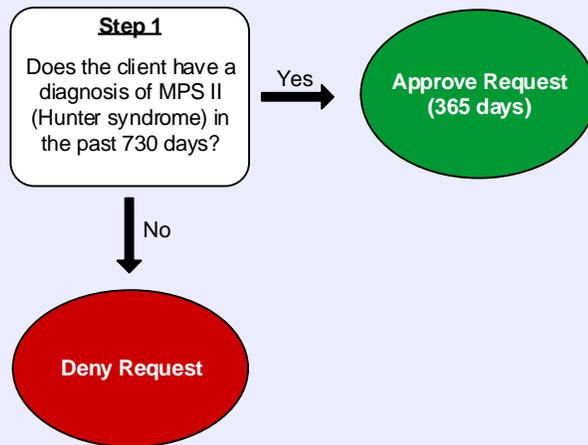
**Enzymes**  
**Elaprase**  
**Clinical Criteria Logic**

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

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](https://TxVendorDrug.com/formulary/formulary-search).

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

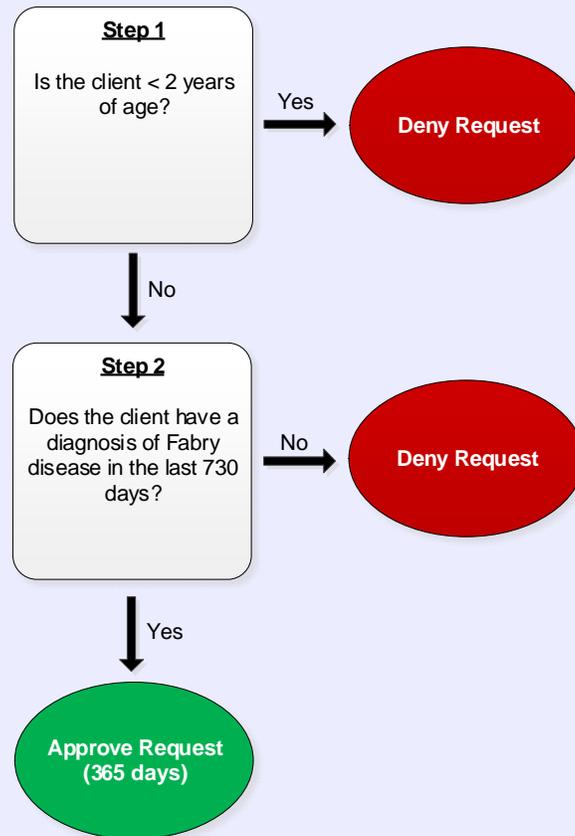


**Enzymes**  
**Fabrazyme**  
**Clinical Criteria Logic**

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

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](https://TxVendorDrug.com/formulary/formulary-search).

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



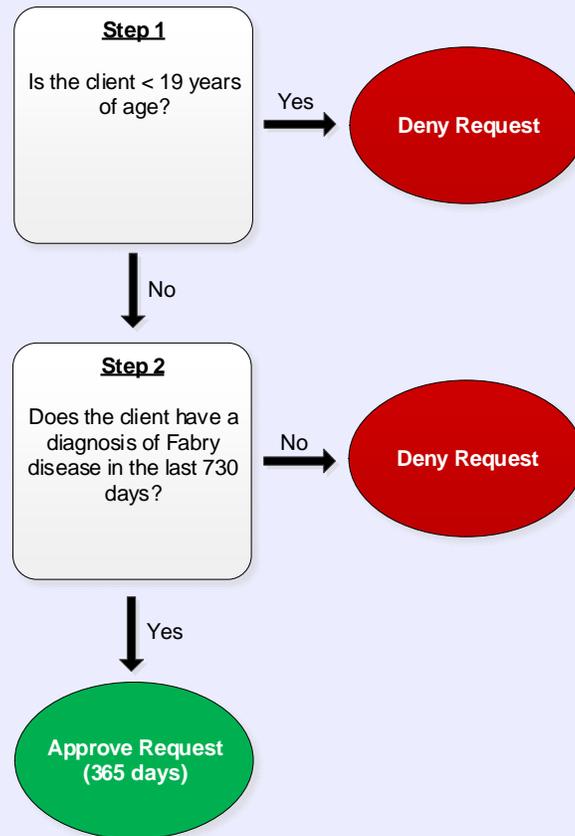
**Enzymes**  
**Galafold**  
**Clinical Criteria Logic**

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

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](http://TxVendorDrug.com/formulary/formulary-search).

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



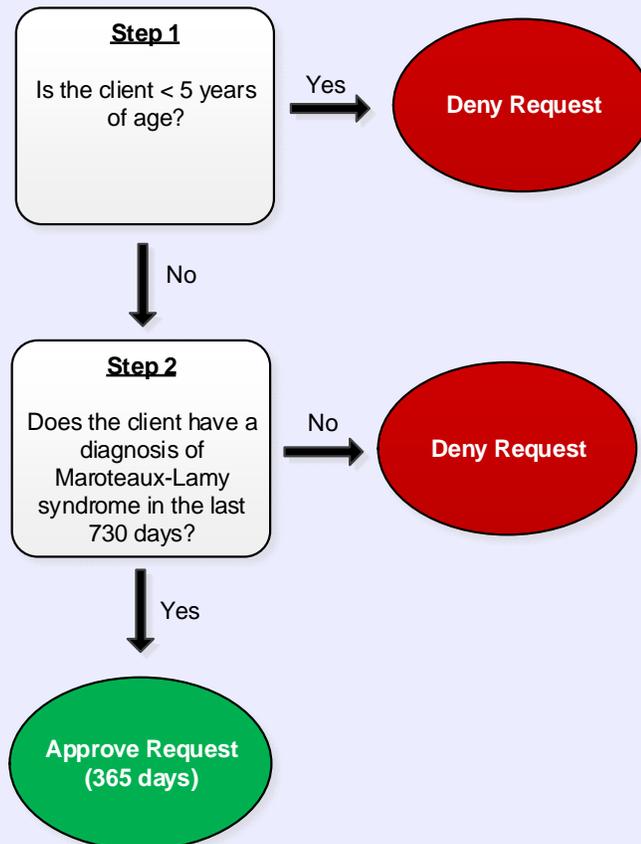
**Enzymes**  
**Naglazyme**  
**Clinical Criteria Logic**

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

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](http://TxVendorDrug.com/formulary/formulary-search).

Drugs Requiring Prior Authorization	
Label Name	GCN
NITYR 2 MG TABLET	43007
NITYR 5 MG TABLET	43006
NITYR 10 MG TABLET	43008
NITISINONE 10 MG CAPSULE	15664
NITISINONE 2 MG CAPSULE	15662
NITISINONE 20 MG CAPSULE	39031
NITISINONE 5 MG CAPSULE	15663
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

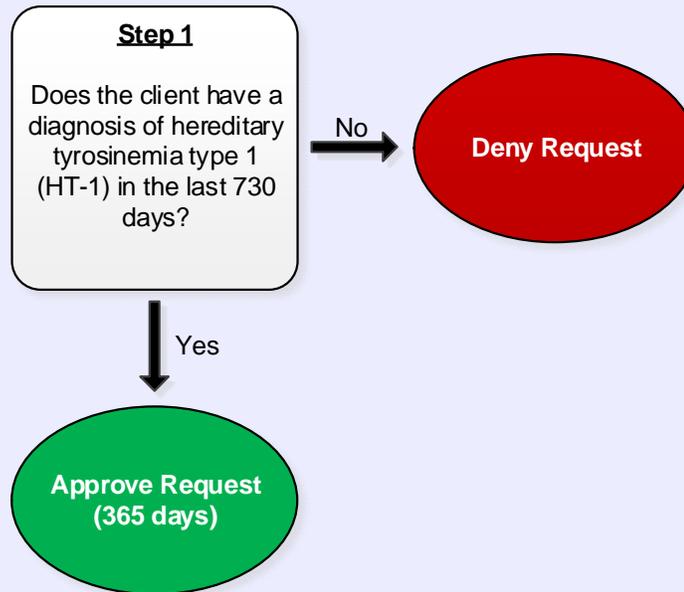


**Enzymes**  
**Nityr / Orfadin**  
**Clinical Criteria Logic**

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**

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](https://TxVendorDrug.com/formulary/formulary-search).

<b>Drugs Requiring Prior Authorization</b>	
<b>Label Name</b>	<b>GCN</b>
REVCOVI 2.4 MG/1.5 ML VIAL	45525



## Enzymes

## Revcovi

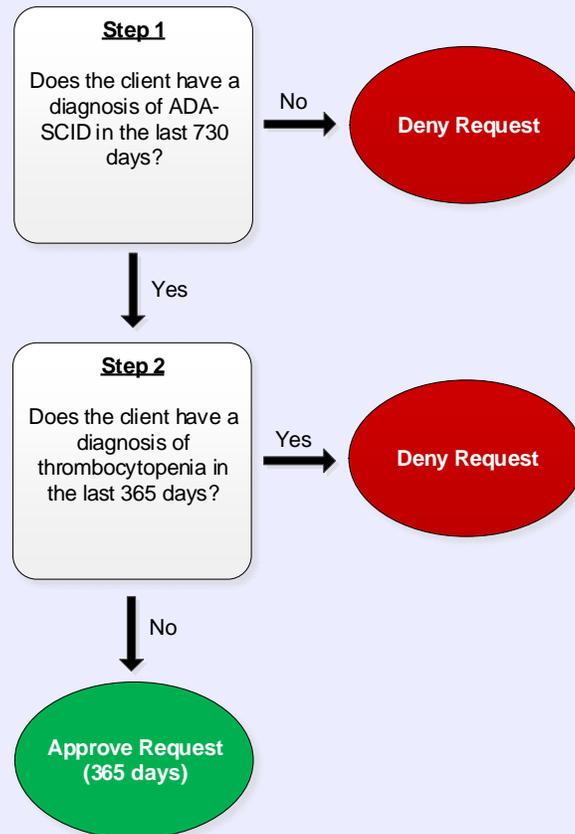
### Clinical Criteria Logic

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**

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](https://TxVendorDrug.com/formulary/formulary-search).

<b>Drugs Requiring Prior Authorization</b>	
<b>Label Name</b>	<b>GCN</b>
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



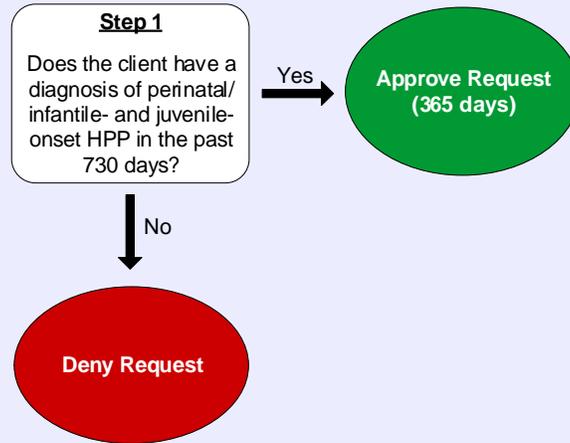
**Enzymes**  
**Strensiq**  
**Clinical Criteria Logic**

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**

*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](https://TxVendorDrug.com/formulary/formulary-search).*

<b>Drugs Requiring Prior Authorization</b>	
<b>Label Name</b>	<b>GCN</b>
VIMIZIM 5 MG/5 ML VIAL	36083



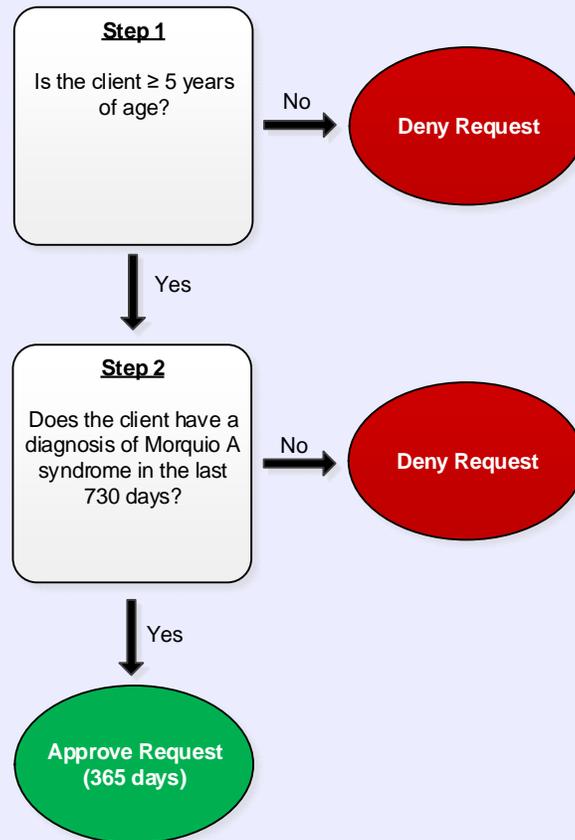
**Enzymes**  
**Vimizim**  
**Clinical Criteria Logic**

1. Is the client greater than or equal to ( $\geq$ ) 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





## Enzymes

### Clinical Criteria Supporting Tables

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

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

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

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

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

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

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

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

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

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

<b>Diagnosis of thrombocytopenia</b> <b>Required diagnosis: 1</b> <b>Look back timeframe: 365 days</b>	
<b>ICD-10 Code</b>	<b>Description</b>
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

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



## Enzymes

### Clinical Criteria References

1. Clinical Pharmacology [online database]. Tampa, FL: Elsevier/Gold Standard, Inc.; 2023. Available at [www.clinicalpharmacology.com](http://www.clinicalpharmacology.com). Accessed on August 18, 2023.
2. Micromedex [online database]. Available at [www.micromedexsolutions.com](http://www.micromedexsolutions.com). Accessed on August 18, 2023.
3. Aldurazyme Prescribing Information. Cambridge, MA. Genzyme Corporation. December 2019.
4. Ceprotin Prescribing Information. Lexington, MA. Takeda Pharmaceuticals USA, Inc. March 2023.
5. Elaprase Prescribing Information. Lexington, MA. Takeda Pharmaceuticals USA, Inc. September 2021.
6. Fabrazyme Prescribing Information. Cambridge, MA. Genzyme Corporation. March 2023.
7. Naglazyme Prescribing Information. Novato, CA. BioMarin Pharmaceutical Inc. December 2019.
8. Nityr Prescribing Information. Cambridge, United Kingdom. Cycle Pharmaceuticals Ltd. June 2021.
9. Orfadin Prescribing Information. Waltham, MA. Sobi, Inc. November 2021.
10. Revcovi Prescribing Information. Cary, NC. Chiesi USA, Inc. April 2021.
11. Vimizim Prescribing Information. Novato, CA. BioMarin Pharmaceutical Inc. December 2019.
12. Galafold Prescribing Information. Philadelphia, PA. Amicus Therapeutics US, Inc. June 2023.
13. Strensiq Prescribing Information. Boston, MA. Alexion Pharmaceuticals, Inc. June 2020.

## 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 <a href="http://TxVendorDrug.com/formulary/formulary-search">TxVendorDrug.com/formulary/formulary-search</a> .) 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 Ceptrotin 400-600 units vial (15482) – not on formulary Updated age to $\geq 2$ 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
05/08/2024	Annual review by staff Added GCNs for nitisinone (15664, 15662, 39031, 15663) to drugs requiring PA (Nityr/Orfadin Table) Updated references