

## Texas Prior Authorization Program Clinical Criteria

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### Drug/Drug Class

## Enzymes

- [Aldurazyme](#)
- [Adagen](#)
- [Ceprotin](#)
- [Elaprase](#)
- [Fabrazyme](#)
- [Lumizyme](#)
- [Naglazyme](#)
- [Nityr / Orfadin](#)
- [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

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](http://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



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

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

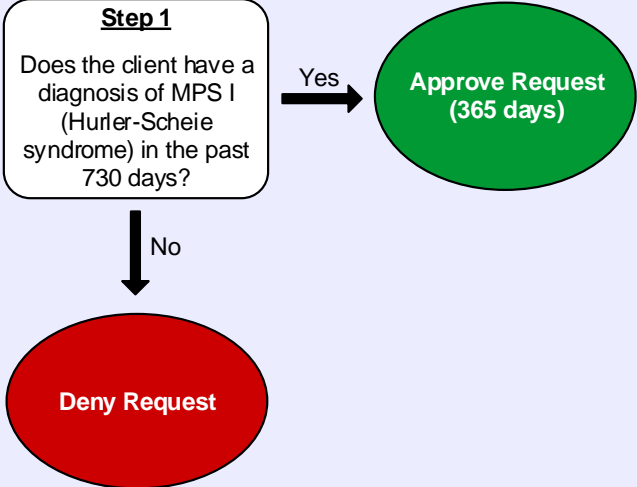


**Enzymes**  
**Aldurazyme**  
**Clinical Criteria Logic**

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 Adagen

### 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
ADAGEN 250 UNITS/ML VIAL	33971

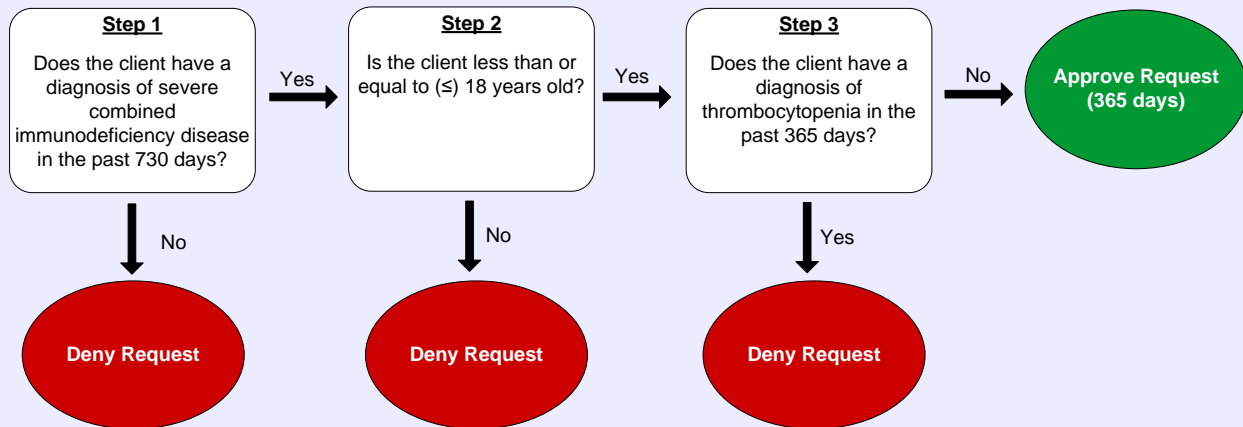
**Enzymes****Adagen****Clinical Criteria Logic**

1. Does the client have a **diagnosis of severe combined immunodeficiency disease** in the past 730 days?  
 Yes (Go to #2)  
 No (Deny)
2. Is the client less than or equal to ( $\leq$ ) 18 years of age?  
 Yes (Go to #3)  
 No (Deny)
3. Does the client have a diagnosis of **thrombocytopenia** in the past 365 days?  
 Yes (Deny)  
 No (Approve – 365 days)



# Enzymes Adagen

## 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).*

<b>Drugs Requiring Prior Authorization</b>	
<b>Label Name</b>	<b>GCN</b>
CEPROTIN 400-600 UNITS VIAL	15482
CEPROTIN 8002-1,200 UNITS VIAL	15483



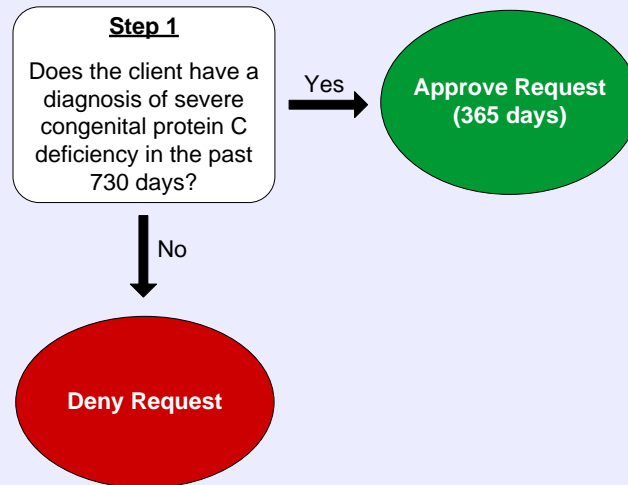
**Enzymes**  
**Ceprotin**  
**Clinical Criteria Logic**

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).*

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



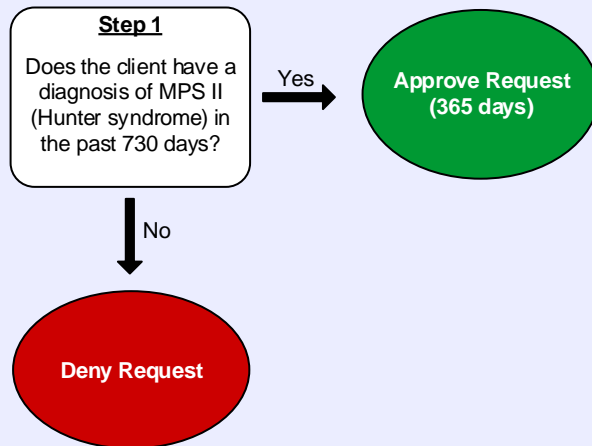
**Enzymes**  
**Elaprase**  
**Clinical Criteria Logic**

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

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



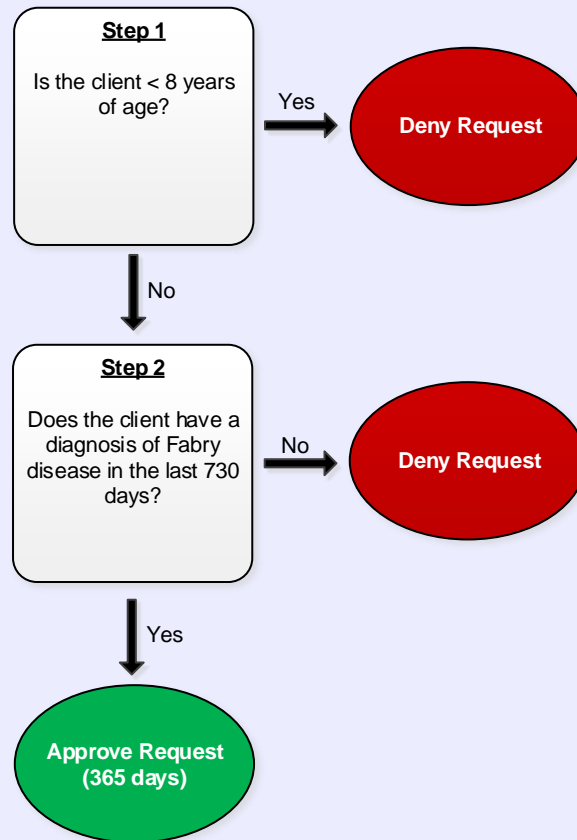
**Enzymes**  
**Fabrazyme**  
**Clinical Criteria Logic**

1. Is the client less than (<) 8 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 Lumizyme

### 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
LUMIZYME 50MG VIAL	26866

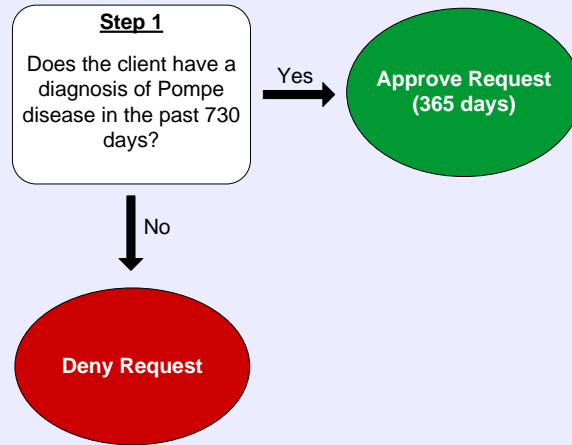


**Enzymes**  
**Lumizyme**  
**Clinical Criteria Logic**

1. Does the client have a **diagnosis of Pompe disease** in the past 730 days?  
 Yes (Approve – 365 days)  
 No (Deny)



# Enzymes Lumizyme 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



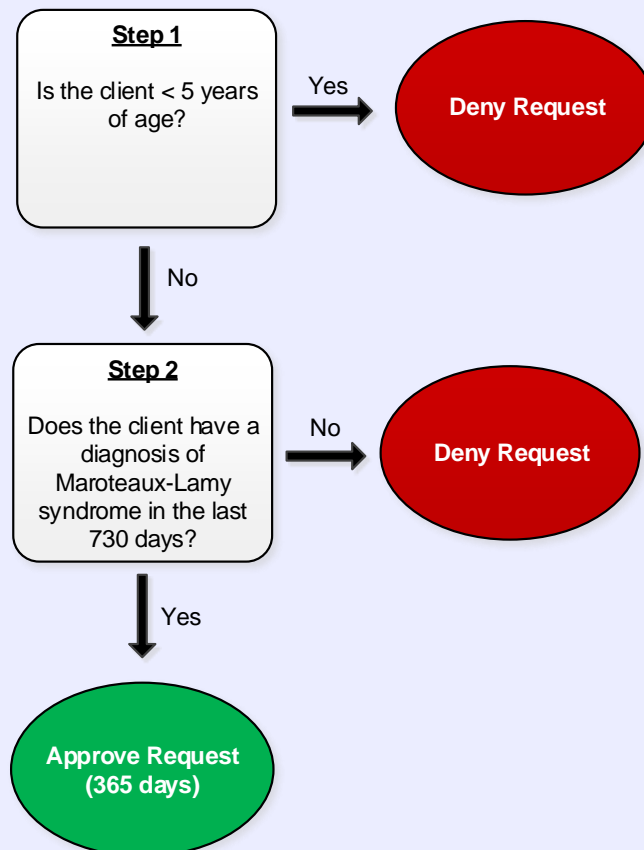
**Enzymes**  
**Naglazyme**  
**Clinical Criteria Logic**

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
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



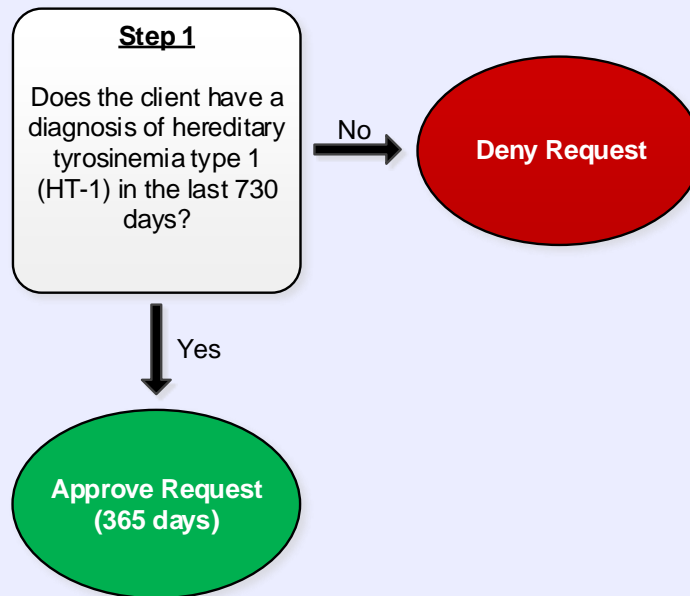


**Enzymes**  
**Nityr / Orfadin**  
**Clinical Criteria Logic**

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  
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](http://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



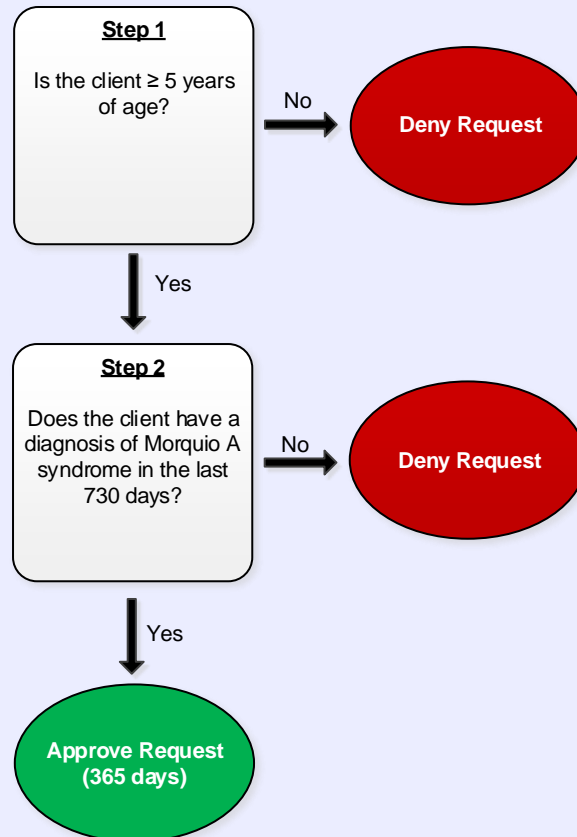
**Enzymes**  
**Vimizim**  
**Clinical Criteria Logic**

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 severe combined immunodeficiency disease</b> <b>Required diagnosis: 1</b> <b>Look back timeframe: 730 days</b>	
ICD-10 Code	Description
D810	SEVERE COMBINED IMMUNODEFICIENCY (SCID) WITH RETICULAR DYSGENESIS
D811	SEVERE COMBINED IMMUNODEFICIENCY (SCID) WITH LOW T- AND B-CELL NUMBERS
D812	SEVERE COMBINED IMMUNODEFICIENCY (SCID) WITH LOW OR NORMAL B-CELL NUMBERS
D8189	OTHER COMBINED IMMUNODEFICIENCIES
D819	COMBINED IMMUNODEFICIENCY, UNSPECIFIED

<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 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
D694	OTHER PRIMARY THROMBOCYTOPENIA
D6941	EVANS SYNDROME
D6942	CONGENITAL AND HEREDITARY THROMBOCYTOPENIA PURPURA
D6949	OTHER PRIMARY THROMBOCYTOPENIA
D695	SECONDARY THROMBOCYTOPENIA
D6951	POSTTRANSFUSION 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>
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. Aldurazyme prescribing information. Cambridge, MA. Genzyme Corporation. April 2013.
2. Adagen prescribing information. Gaithersburg, MD. Leadiant Biosciences, Inc. November 2017
3. Ceprotin prescribing information. Westlake Village, CA. Baxter Healthcare Corporation. September 2015.
4. Elaprase prescribing information. Lexington, MA. Shire Human Genetic Therapies, Inc. June 2013.
5. Fabrazyme prescribing information. Cambridge, MA. Genzyme Corporation. July 2010.
6. Lumizyme prescribing information. Cambridge, MA. Genzyme Corporation. August 2014.
7. Naglazyme prescribing information. Novato, CA. BioMarin Pharmaceutical Inc. March 2013.
8. Nityr prescribing information. Cambridge, United Kingdom. Cycle Pharmaceuticals Ltd. July 2017.
9. Orfadin prescribing information. Waltham, MA. Swedish Orphan Biovitrum. September 2017.
10. Vimizim prescribing information. Novato, CA. BioMarin Pharmaceutical Inc. February 2014.
11. 2014 ICD-9-CM Diagnosis Codes, Volume 1. 2014. Available at <http://www.icd9data.com>. Accessed on March 26, 2014.
12. 2014 ICD-10-CM Diagnosis Codes, Volume 1. 2014. Available at <http://icd10data.com>. Accessed on March 26, 2014.
13. Clinical Pharmacology [online database]. Tampa, FL: Elsevier/Gold Standard, Inc.; 2018. Available at [www.clinicalpharmacology.com](http://www.clinicalpharmacology.com). Accessed on March 30, 2018.
14. Micromedex [online database]. Available at [www.micromedexsolutions.com](http://www.micromedexsolutions.com). Accessed on March 30, 2018.

## 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