Aldurazyme® (laronidase)
(Intravenous)

Document Number: IC-0006

Last Review Date: 02/04/2019
Date of Origin: 11/28/2011
Dates Reviewed: 12/2011, 02/2013, 02/2014, 12/2014, 10/2015, 10/2016, 10/2017, 10/2018, 02/2019

I. Length of Authorization

Coverage will be provided for 12 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

- Aldurazyme 2.9 mg vial: 92 vials every 28 days

B. Max Units (per dose and over time) [Medical Benefit]:

- 667 billable units every 7 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Mucopolysaccharidosis I (MPS I) †

- Patient has a definitive diagnosis of MPS I confirmed by one of the following:
  - Detection of pathogenic mutations in the *IDUA* gene by molecular genetic testing; **OR**
  - Detection of deficient activity of the lysosomal enzyme α-L-iduronidase (IDUA); **AND**
- Diagnosis of Hurler (severe) or Hurler-Scheie (attenuated) forms of disease **OR**
- Diagnosis of Scheie (attenuated) form of disease with moderate to severe symptoms; **AND**
- Patient is 6 months of age or older; **AND**
- Patient has absence of severe cognitive impairment; **AND**
- Documented baseline value for urinary glycosaminoglycan (uGAG) ; **AND**
- Documented baseline values for one or more of the following:
  - Patients 6 years or greater: percent predicted forced vital capacity (FVC), 6-minute walk test, joint range of motion, left ventricular hypertrophy, growth, quality of life (CHAQ/HAQ/MPS HAQ); **OR**

- Patients 6 months to less than 6 years: cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC, and/or 6-minute walk test

† FDA approved indication(s)

IV. Renewal Criteria

Authorizations can be renewed based on the following criteria:

- Patient continues to meet the criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity reactions, acute respiratory complications, acute cardiorespiratory failure, severe infusion reactions, etc.; **AND**
- Patient does not have progressive/irreversible severe cognitive impairment; **AND**
- Patient has a documented reduction in uGAG levels; **AND**
- Patient has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following:
  - Patients 6 years or greater: stability or improvement in percent predicted FVC and/or 6-minute walk test, increased joint range of motion, decreased left ventricular hypertrophy, improved growth, improved quality of life (clinically meaningful change in the CHAQ/HAQ/MPS HAQ disability index); **OR**
  - Patients 6 months to less than 6 years: stability or improvement in cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC and/or 6-minute walk test

V. Dosage/Administration

Indication	Dose
Mucopolysaccharidosis I (MPS I)	0.58 mg/kg of body weight administered once weekly, as an intravenous infusion, over 3-4 hours.

VI. Billing Code/Availability Information

Jcode:

J1931 – Injection, laronidase, 0.1 mg; 1 billable unit = 0.1 mg

NDC:

Aldurazyme 2.9 mg/5 mL single-dose vial: 58468-0070-xx

VII. References

1. Aldurazyme [package insert]. Cambridge, MA; Genzyme Corporation.; April 2013. Accessed January 2019.

2. Clark LA. Mucopolysaccharidosis Type I. GeneReviews. www.ncbi.nlm.nih.gov/books/NBK1162/ (Accessed on August 24, 2018).
3. Muenzer J, Wraith JE, Clarke LA; International Consensus Panel on Management and Treatment of Mucopolysaccharidosis I. Mucopolysaccharidosis I: management and treatment guidelines. Pediatrics. 2009 Jan; 123(1):19-29. doi: 10.1542/peds.2008-0416.
4. Martins AM, Dualibi AP, Norato D, et al. 1.Guidelines for the Management of Mucopolysaccharidosis Type I. JPeds. 155 (4): S32 - S46.
5. Wisconsin Physicians Service Insurance Corporation. Local Coverage Determination (LCD): Drugs and Biologics (L34741). Centers for Medicare & Medicaid Services, Inc. Updated on 05/24/2018 with effective date 6/1/2018. Accessed January 2019.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E76.01	Hurler's syndrome
E76.02	Hurler-Scheie syndrome
E76.03	Scheie's syndrome

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD):

Jurisdiction(s): 5,8	**NCD/LCD Document (s):** L34741
https://www.cms.gov/medicare-coverage-database/search/lcd-date-search.aspx?DocID=L34741&bc=gAAAAAAAAAAAAA==	

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC