Aldurazyme® (laronidase)
(Intravenous)

Document Number: IC-0006

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I. Length of Authorization

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

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Aldurazyme 2.9 mg/5 mL vial: 92 vials every 28 days

B. Max Units (per dose and over time) [HCPCS Unit]:

- 667 billable units every 7 days

III. Initial Approval Criteria [1-6]

Coverage is provided in the following conditions:

- Patient is at least 6 months of age; **AND**
- Documented baseline value for urinary glycosaminoglycan (uGAG) has been obtained; **AND**
- Documented baseline age-appropriate values for one or more of the following have been obtained:
  - Patients 6 years of age or greater: percent predicted forced vital capacity (FVC), 6-minute walk test (6-MWT), joint range of motion, left ventricular hypertrophy, growth, and/or quality of life (CHAQ/HAQ/MPS HAQ); **OR**
  - Patients 6 months to less than 6 years of age: cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC, and/or 6-MWT; **AND**

***NOTE:** For very young patients in which FVC or 6-MWT are not suitable for measuring, requests will be reviewed on a case-by case basis.*

Universal Criteria [1]

- Patient does not have severe cognitive impairment; **AND**

Mucopolysaccharidosis I (MPS I) † Φ [1,2]

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- Patient has a definitive diagnosis of MPS I as confirmed by one of the following:
  - Detection of biallelic pathogenic mutations in the *IDUA* gene by molecular genetic testing; **OR**
  - Detection of deficient activity of the lysosomal enzyme α-L-iduronidase (IDUA); **AND**
- Patient has one of the following diagnoses:
  - Hurler (severe) or Hurler-Scheie (attenuated) forms of disease; **OR**
  - Scheie (attenuated) form of disease with moderate to severe symptoms

† FDA approved indication(s); ‡ Compendia recommended indication(s); Φ Orphan Drug

IV. Renewal Criteria [1,2,5,6]

Coverage may be renewed based on the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: anaphylaxis and severe hypersensitivity reactions, acute respiratory complications, acute cardiorespiratory failure, severe infusion reactions, etc.; **AND**
- Patient has a documented reduction in uGAG levels compared to pretreatment baseline; **AND**
- Patient has demonstrated a beneficial response to therapy compared to pretreatment age-appropriate baseline values in one or more of the following:
  - Patients 6 years of age or greater: stability or improvement in percent predicted FVC and/or 6-MWT, increased joint range of motion, decreased left ventricular hypertrophy, improved growth, and/or improved quality of life (clinically meaningful change in the CHAQ/HAQ/MPS HAQ disability index); **OR**
  - Patients 6 months to less than 6 years of age: stability or improvement in cardiac status, upper airway obstruction during sleep, growth velocity, mental development, FVC and/or 6-MWT

V. Dosage/Administration [1]

Indication	Dose
Mucopolysaccharidosis I (MPS I)	0.58 mg/kg of body weight administered once weekly as an intravenous infusion

VI. Billing Code/Availability Information

HCPCS Code:

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

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NDC:

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

VII. References

1. Aldurazyme [package insert]. Cambridge, MA; Genzyme Corporation.; December 2019. Accessed December 2022.
2. Clark LA. Mucopolysaccharidosis Type I. GeneReviews®. www.ncbi.nlm.nih.gov/books/NBK1162/. Initial Posting: October 31, 2002; Last Update: February 25, 2021. Accessed on January 11, 2023.
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. Clarke LA, Wraith JE, Beck M, et al. Long-term efficacy and safety of laronidase in the treatment of mucopolysaccharidosis I. Pediatrics. 2009 Jan;123(1):229-40. doi: 10.1542/peds.2007-3847.
6. Wraith JE, Beck M, Lane R, et al. Enzyme replacement therapy in patients who have mucopolysaccharidosis I and are younger than 5 years: results of a multinational study of recombinant human alpha-L-iduronidase (laronidase). Pediatrics. 2007 Jul;120(1):e37-46. Epub 2007 Jun 4.

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), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/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/LCA): N/A

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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.
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

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PO Box 1062
Minneapolis, MN 55440
Phone: 1.866.631.5404 (TTY: 711)
Fax: 763.847.4010
Email: customerservice@aspirushealthplan.com

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U.S. Department of Health and Human Services
200 Independence Avenue, SW
Room 509F, HHH Building
Washington, D.C. 20201
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Complaint forms are available at http://www.hhs.gov/ocr/office/file/index.html.

Language Assistance Services

Albanian: KUJDES: Nëse flitni shqip, për ju ka në dispozicion shërbime të asistencës gjuhësore, pa pagesë.Telefononi në 1.866.631.5404 (TTY: 711).

Arabic تنبيه: إذا كنت تتحدث اللغة العربية، فإن خدمات المساعدة اللغوية متاحة لك مجاناً . اتصل بن اعلى رقم الهاتف 1.866.631.5404(رقم هاتف الصم والبك : 711)

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Hindi: _यान द_: य_द आप ि◌हंदी बोलते ह_ तो आपके ि◌लए मु_त म_ भाषा सहायता सेवाएं उपल_ध ह_। 1.866.631.5404 (TTY: 711) पर कॉल कर_।

Hmong: LUS CEEV: Yog tias koj hais lus Hmoob, cov kev pab txog lus, muaj kev pab dawb rau koj. Hu rau 1.866.631.5404 (TTY: 711).

Korean: 주의: 한국어를 사용하시는 경우, 언어 지원 서비스를 무료로 이용하실 수 있습니다. 1.866.631.5404 (TTY: 711)번으로 전화해 주십시오.

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Spanish: ATENCIÓN: si habla español, tiene a su disposición servicios gratuitos de asistencia lingüística. Llame al 1.866.631.5404 (TTY: 711).

Tagalog: PAUNAWA: Kung nagsasalita ka ng Tagalog, maaari kang gumamit ng mga serbisyo ng tulong sa wika nangwalang bayad. Tumawag sa 1.866.631.5404 (TTY: 711)

Traditional Chinese: 注意：如果您使用繁體中文，您可以免費獲得語言援助服務。請 致電 1.866.631.5404 (TTY:711).

Vietnamese: CHÚ Ý: Nếu bạn nói Tiếng Việt, có các dịch vụ hỗ trợ ngôn ngữ miễn phí dành cho bạn. Gọi số 1.866.631.5404 (TTY: 711).

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Lao: ໂປດຊາບ: ຖ້າວ່າ ທ່ານເວົ້າພາສາ ລາວ, ການບໍລິການຊ່ວຍເຫຼືອດ້ານພາສາ,ໂດຍບໍ່ເສັ້ງຄ່າ, ແມ່ນມີພ້ອມໃຫ້ທ່ານ. ໂທຣ 1.866.631.5404 (TTY:711).

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