Clinical Policy Title:	agalsidase beta
Policy Number:	RxA.117
Drug(s) Applied:	Fabrazyme®
Original Policy Date:	02/07/2020
Last Review Date:	10/19/2023
Line of Business Policy Applies to:	All Line of Business (except Medicare)

Criteria

I. Initial Approval Criteria

A. Fabry Disease (must meet all):

1. Diagnosis of Fabry disease confirmed by one of the following (a or b):
   a. Enzyme assay demonstrating a deficiency of alpha-galactosidase activity;
   b. DNA testing;
2. Age ≥ 2 years;
3. Prescribed by or in consultation with a clinical geneticist, cardiologist, nephrologist, neurologist, lysosomal disease specialist, or Fabry disease specialist;
4. Fabrazyme is not prescribed concurrently with Galafold®;
5. Dose does not exceed 1 mg/kg body weight every 2 weeks.

Approval Duration
Commercial: 6 months
Medicaid: 6 months

II. Continued Therapy Approval

A. Fabry Disease (must meet all)**:**

1. Member is currently receiving medication that has been authorized by RxAdvance or the member has met initial approval criteria listed in this policy;
2. Member is responding positively to therapy as evidenced by improvement in the individual member's Fabry disease manifestation profile;
3. If request is for a dose increase, new dose does not exceed 1 mg/kg body weight every 2 weeks.

Approval Duration
Commercial: 6 months
Medicaid: 6 months

References

1. Desnick RJ, Brady R, Barranger J, et al. Fabry disease, an under-recognized multisystemic disorder: expert recommendations for diagnosis, management, and enzyme replacement therapy. Ann Intern Med. 2003; 138(4): 338-346. Available at: https://pubmed.ncbi.nlm.nih.gov/12585833/. Accessed October 13, 2022.
2. Ortiz A, Germain DP, Desnick RJ, Politei J, Mauer M, Burlina A, Eng C, Hopkin RJ, Laney D, Linhart A, Waldek S, Wallace E, Weidemann F, Wilcox WR. Fabry disease revisited: Management and treatment recommendations for adult patients. Mol Genet Metab. 2018 Apr;123(4):416-427. doi: 10.1016/j.ymgme.2018.02.014. Epub 2018 Feb 28. Review. PubMed PMID: 29530533. Available at: https://pubmed.ncbi.nlm.nih.gov/29530533/. Accessed October 13,

2022.

Review/Revision History	Review/Revised Date	P&T Approval Date
Policy was established	01/2020	02/07/2020
Updated references	04/30/2020	05/20/2020
Policy was reviewed: 1. Clinical policy title table was updated. 2. Drug(s) applied was updated. 3. Line of Business Policy Applies to was update to all lines of business. 4. Continued Therapy criteria II.A.1 was rephrased to "Currently receiving medication that has been authorized by RxAdvance..." 5. Initial Approval criteria: Commercial and Medicaid approval duration were updated from length of benefit to 6 months. 6. Continued Approval criteria: Commercial and Medicaid approval duration were updated from length of benefit to 6 months. 7. References were reviewed and updated. 8. Updated dosing regimen to add body weight. 9. Updated Initial and Continued Therapy to include body weight.	01/20/2021	03/09/2021
Policy was reviewed: 1. Initial Approval Criteria, I.A.2: Updated age criteria from Age 8 years of age or older to Age 2 years of age or older. 2. References were reviewed and updated.	11/24/2021	01/17/2022
Policy was reviewed: 1. Initial Approval Criteria, I.A.3: Updated to include new prescriber criteria Prescribed by or in consultation with a clinical geneticist, cardiologist, nephrologist, neurologist,	10/13/2022	01/17/2023

lysosomal disease specialist, or Fabry disease specialist. 2. Initial Approval Criteria, I.A.4: Updated to include new prescribing criteria Fabrazyme is not prescribed concurrently with Galafold®. 3. References were reviewed and updated.		
Policy was reviewed.	10/19/2023	10/19/2023