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Policy Number: C4865-A

## Fabrazyme (agalsidase beta)

### PRODUCTS AFFECTED

Fabrazyme (agalsidase beta)

### COVERAGE POLICY

Coverage for services, procedures, medical devices and drugs are dependent upon benefit eligibility as outlined in the member's specific benefit plan. This Coverage Guideline must be read in its entirety to determine coverage eligibility, if any. This Coverage Guideline provides information related to coverage determinations only and does not imply that a service or treatment is clinically appropriate or inappropriate. The provider and the member are responsible for all decisions regarding the appropriateness of care. Providers should provide Molina Healthcare complete medical rationale when requesting any exceptions to these guidelines.

#### **Documentation Requirements:**

Molina Healthcare reserves the right to require that additional documentation be made available as part of its coverage determination; quality improvement; and fraud; waste and abuse prevention processes. Documentation required may include, but is not limited to, patient records, test results and credentials of the provider ordering or performing a drug or service. Molina Healthcare may deny reimbursement or take additional appropriate action if the documentation provided does not support the initial determination that the drugs or services were medically necessary, not investigational or experimental, and otherwise within the scope of benefits afforded to the member, and/or the documentation demonstrates a pattern of billing or other practice that is inappropriate or excessive.

#### **DIAGNOSIS:**

Fabry disease

#### **REQUIRED MEDICAL INFORMATION:**

This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. If a drug within this policy receives an updated FDA label within the last 180 days, medical necessity for the member will be reviewed using the updated FDA label information along with state and federal requirements, benefit being administered and formulary preferencing. Coverage will be determined on a case-by case basis until the criteria can be updated through Molina Healthcare, Inc. clinical governance. Additional information may be required on a case-by-case basis to allow for adequate review. When the requested drug product for coverage is dosed by weight, body surface area or other member specific measurement, this data element is required as part of the medical necessity review. The Pharmacy and Therapeutics Committee has determined that the drug benefit shall be a mandatory generic and that generic drugs will be dispensed whenever available.

#### **A. FABRY DISEASE:**

1. (a) Documented diagnosis of classic Fabry disease with typical clinical manifestations confirmed by documented deficient  $\alpha$ -galactosidase A ( $\alpha$ -Gal A) enzyme activity in plasma, isolated leukocytes, and/or cultured cells using alpha galactosidase A enzyme assay (Males with classic Fabry disease

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have less than 1%  $\alpha$ -Gal A enzyme activity, Males with atypical Fabry disease have residual enzyme activity that is greater than 1% of normal) OR Molecular genetic testing that identifies a GLA mutation providing additional confirmation of the diagnosis [DOCUMENTATION REQUIRED]  
OR

(b) Diagnosed as a carrier of Fabry disease with significant clinical manifestations, confirmed by documented decrease  $\alpha$ -Gal A enzyme activity in plasma and/or isolated leukocytes confirms the carrier state in a female OR documented molecular genetic testing to clarify genetic status [DOCUMENTATION REQUIRED]  
AND

2. (a) Documentation of baseline status by Mainz Severity Score Index (MSSI) or FOS Mainz Severity Score Index

OR

(b) Documentation of objective/subjective clinical information, including signs/symptoms, with sufficient clinical manifestations to justify treatment and supported by at least one of the following: pain in the extremities, hypohidrosis, corneal opacities, kidney dysfunction, cardiac dysfunction, cerebrovascular disorders

OR

(c) Documentation of baseline plasma globotriaosylceramide (GL3 or Gb3) level

AND

3. Documentation of treatment plan that includes goals of therapy

AND

4. Prescriber attestation that member will NOT receive concurrent therapy with Galafold (migalastat) or other enzyme replacement therapy for Fabry disease [pegunigalsidase alfa (Elfabrio)]

### CONTINUATION OF THERAPY:

#### A. FABRY DISEASE:

1. Documentation of ANY of the following:

- a) Improvement or stabilization of Mainz Severity Score Index (MSSI) or FOS Mainz Severity Score Index

OR

- b) A marked clinical improvement in or a stabilization in disease progression from baseline disease manifestations (e.g., kidney dysfunction, cardiac dysfunction, etc.)

OR

- c) Disease response as evidenced by a reduction in plasma GL-3 and/or GL-3 inclusions or Gb3 compared to pre-treatment baseline

AND

2. Prescriber attests or clinical reviewer has found member is not receiving concurrent therapy with Galafold or other enzyme replacement therapy for Fabry disease [pegunigalsidase alfa (Elfabrio)]

AND

3. Prescriber attests to or clinical reviewer has found no evidence of intolerable adverse effects or drug toxicity (e.g., severe hypersensitivity reactions, severe infusion site reactions, compromised cardiac function, etc.)

### DURATION OF APPROVAL:

Initial authorization: 12 months, Continuation of therapy: 12 months

### PRESCRIBER REQUIREMENTS:

Prescribed by, or in consultation with, a board-certified Nephrologist, Cardiologist, Neurologist, Endocrinologist, Clinical Geneticist, Clinical Biochemical Geneticist, or physician experienced in the management of Fabry disease. [If prescribed in consultation, consultation notes must be submitted with initial request and reauthorization requests]

### AGE RESTRICTIONS:

2 years of age and older

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### QUANTITY:

1 mg/kg body weight given every two weeks as an IV infusion

**Maximum Quantity Limits** – To limit wastage, the use of 35mg vials should be restricted for patients weighing 31kg or greater. The use of a second 35mg vial should be for patients weighing 66kg or greater, and a third for patients weighing 101kg or greater.

### PLACE OF ADMINISTRATION:

The recommendation is that infused medications in this policy will be for pharmacy or medical benefit coverage administered in a place of service that is a non-hospital facility-based location as per the Molina Health Care Site of Care program.

Note: Site of Care Utilization Management Policy applies for Fabrazyme (agalsidase beta). For information on site of care, see [Specialty Medication Administration Site of Care Coverage Criteria \(molinamarketplace.com\)](https://molinamarketplace.com)

## DRUG INFORMATION

### ROUTE OF ADMINISTRATION:

Intravenous

### DRUG CLASS:

Fabry Disease - Agents

### FDA-APPROVED USES:

Indicated for treatment of adult and pediatric patients 2 years of age and older with confirmed Fabry disease.

E75.21 Fabry-Anderson disease

### COMPENDIAL APPROVED OFF-LABELED USES:

None

## APPENDIX

### APPENDIX:

None

## BACKGROUND AND OTHER CONSIDERATIONS

### BACKGROUND:

Agalsidase beta (Fabrazyme®) is the first FDA-approved therapy for Fabry disease. Agalsidase beta is designated an orphan drug by the US Food and Drug Administration for use in this condition. Agalsidase beta is a biosynthetic (recombinant DNA origin) form of human  $\alpha$ -galactosidase, an enzyme that metabolizes glycosphingolipids, including globotriaosylceramide (GL-3). This agent is prepared from cultures of genetically modified mammalian (Chinese hamster ovary) cells by recombinant DNA technology. Patients with Fabry disease have a deficiency of the lysosomal enzyme, ceramidetrihexosidase or  $\alpha$ -galactosidase A, which breaks down glycosphingolipids (predominantly globotriaosylceramide or GL-3). Glycosphingolipids accumulate in the lining of blood vessels in the heart, kidney, and other organs in patients without an adequate presence of  $\alpha$ -galactosidase A. Crises of severe pain in the extremities (acroparesthesias), hypohidrosis, corneal opacities and dysfunction of several organs (kidney, brain, heart) are the primary manifestations, and patients often have decreased life expectancy and experience renal failure, cardiomyopathy and cerebrovascular accidents. Progressive renal insufficiency and cardiovascular disease are causes of significant morbidity and mortality in Fabry

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disease, however nearly any organ may be affected.

Replacement of the deficient enzyme may reduce levels of the accumulated glycolipids in various tissues and attenuate disease symptoms.

Fabry disease is a rare X-linked metabolic disorder caused by the partial or complete deficiency of a lysosomal enzyme  $\alpha$ -galactosidase A. As a result of this enzyme deficiency, neutral sphingolipids with terminal  $\alpha$ -galactosyl residues [predominantly globotriaosylceramide (Gb3)] accumulate in the lysosomes of different tissues and fluids (epithelial cell of glomeruli and tubules of the kidneys; cardiac myocytes; ganglion cells of the autonomic system; cornea; endothelial, perithelial, and smooth muscle cells of blood vessels; and histiocytic and reticular cells of connective tissue). The mechanism by which  $\alpha$ -galactosidase A deficiency and glycolipid accumulation cause such a wide variety of complications is not well understood. Based on the pathology of Fabry disease, the chronic accumulation of  $\alpha$ -D-galactosyl moieties, particularly of Gb3, appears to be a chronic toxicity state. Fabry disease involves to a large extent a systemic vascular disorder causing cardiovascular complications and stroke. The clinical onset is generally characterized by severe acroparesthesias, angiokeratoma, corneal and lenticular opacities, and hypohidrosis. Over time, microvascular disease of the kidneys, heart, and brain progresses, leading to death.

Fabry disease is the second most prevalent lysosomal storage disorder, after Gaucher disease, with an estimated incidence ranging between 1:40,000 to 1:170,000 persons. It manifests primarily in affected hemizygous men and, to some extent, in heterozygous females. Clinical onset is variable; the disease usually appears during childhood but may not appear until the second or third decade. Pain, kidney failure, cardiomyopathy, and cerebrovascular events are the complications that are mainly responsible for morbidity and mortality of this disorder.

GLA is the only gene known to be associated with Fabry disease. Nearly 100% of affected males have an identifiable GLA mutation. However, there are cases where an identifiable mutation is not identified or where molecular testing is not available. Therefore, the most efficient and reliable method for the diagnosis of Fabry disease in affected males is the demonstration of deficient  $\alpha$ -galactosidase A ( $\alpha$ -Gal A) enzyme activity in plasma, isolated leukocytes, and/or cultured cells.

In females, measurement of  $\alpha$ -Gal A enzyme activity is unreliable; although demonstration of decreased  $\alpha$ -Gal A enzyme activity is diagnostic of the carrier state, many carrier females have normal  $\alpha$ -Gal A enzyme activity. Molecular genetic testing is the most reliable method for the diagnosis of carrier females. Prior to the availability of enzyme replacement therapy, treatment for Fabry disease consisted primarily of symptomatic care and non-specific corrective measures to treat complications, for example, analgesia, stroke prophylaxis, cardiac interventions including pacemaker, dialysis, and kidney transplantation. Pain, one of the most disturbing and early symptoms, is typically treated with relatively low doses of anti-epileptic medication such as carbamazepine, Neurontin and lamotrigine (Horowitz, 2007; Ries et al., 2003; Tremont-Lukats et al., 2000). It has been noted that phenytoin and carbamazepine diminish the chronic and episodic acroparesthesia (acroparesthesia: severe pain in the extremities, type of sphingolipidosis). Given the chronic nature of the disease and the high risk of addiction, narcotics are generally avoided as treatment of pain symptoms in Fabry disease. Non-steroidal anti-inflammatory drugs are also avoided, as they are ineffective and contraindicated due to potential effect on renal function. The kidney dysfunction in Fabry disease responds well to angiotensin converting enzyme inhibitors (ACEI) and angiotensin receptor blockers (ARB).

Normalization of blood pressure is crucial to preserve kidney function and prevent other vascular events. For Fabry patients who reach end-stage renal disease, renal transplantation is considered. For the primary or secondary prevention of stroke and other vascular pathologies, such as retinal artery occlusion, anti-platelet and anticoagulant therapy may be given. Metoclopramide and pancreatic enzymes are helpful in preventing post-prandial diarrhea and in reducing gastrointestinal symptoms. Effective anti-platelet agents, such as clopidogrel and aspirin/long-acting dipyridamole, have been administered to prevent strokes in all patients with Fabry disease, and particularly in those with a family history of stroke.

While the palliative therapies may prolong life in patients with Fabry disease, their efficiency is limited because these modalities do not treat the enzyme deficiency (and consequent substrate storage), which is the underlying cause of the disease.

## Enzyme Replacement Therapy (ERT)

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ERT is the first specific therapy for Fabry disease. It has been available since 2001 and definitive conclusions as to whether this therapy can modify the natural history of Fabry disease remains to be determined; however it has been shown to be effective in several clinical trials in reducing the amount of stored tissue glycolipids. ERT reduces pain episodes, stabilizes or improves renal function in patients with a mild renal disease, and reduces excess cardiac mass, gastrointestinal symptoms, and hypohidrosis. Gb3 deposition in the skin has also been reduced by the ERT, suggesting that periodic ultrastructural examination of skin might be useful to monitor response to therapy. Whether or not ERT will positively impact on mortality is still unknown. When to begin ERT remains controversial, in particular among female and patients with milder variants. Experts recommend that ERT should be provided in the second decade in male patients or as soon as symptoms are observed.

Two forms of  $\alpha$ -galactosidase A for ERT exist: agalsidase alfa (Replagal, 0.2 mg/kg per infusion) and agalsidase beta (Fabrazyme, 1 mg/kg per infusion). Both are approved in Europe and many other countries, however in the US the FDA approved only agalsidase beta. Both forms of the enzyme are usually administered every two weeks. No clear difference in clinical effect was demonstrated between the two enzyme preparations in a randomized controlled prospective study using either the same or the approved dose. The latter study was not randomized and has a number of flaws.

### Diagnosis and Monitoring:

Adverse events relating to ERT reported thus far have only been infusion related (hives, pruritus, fever, and rigors) responding to premedication and a decrease in infusion rate.

Initial assessment-- An initial evaluation should consist of the following:

- Detailed past medical history and review of systems. Clinical symptoms or signs such as neuropathic pain; heat intolerance (usually associated with exercise intolerance and avoidance of outdoors in summer months); decreased production of sweat, tears, or saliva; diarrhea; abdominal pain; angiokeratomas; and foamy urine should be carefully documented at baseline. Any history of transient ischemic attacks or ischemic strokes (particularly involving the posterior circulation) and myocardial disease should be thoroughly explored.
- Detailed family history that focuses on relatives with unexplained neurologic disease, kidney failure, or heart disease that was transmitted as an X-linked trait. In the National Institutes of Health (NIH) series, family history contributed to the diagnosis in 46 percent of patients.
- Careful physical examination, looking for angiokeratomas, telangiectasias, hypo- or anhydrosis, corneal opacities, edema or lymphedema, abnormal cardiac examination (evidence of LVH, arrhythmia).
- Examination of urine sediment and measurement of renal function. With renal involvement, there may be oval fat bodies (degenerating tubular epithelial cells with lipid inclusions) with a lamellar structure and a Maltese cross pattern under polarized microscopy; this is similar to what may be seen with nephrotic-range proteinuria of any cause.
- Electrocardiogram to evaluate for LVH and conduction defects. In females, LVH may be absent and cardiac magnetic resonance imaging (MRI) may be required to identify fibrotic lesions of Fabry disease, which can precede the onset of LVH.
- In addition, some clinicians obtain a baseline MRI of the brain as part of the initial evaluation, particularly in patients who present with neurologic manifestations. Findings on MRI may be normal or show evidence of an old ischemic lesion, white matter abnormalities on T2-weighted or fluid-attenuated inversion recovery (FLAIR) images, or pulvinar lesions on T1-weighted images.

### Establishing the diagnosis

Diagnosis in males-- In all male patients suspected of having Fabry disease, we measure leukocyte alpha-galactosidase A (alpha-Gal A) activity as the initial diagnostic assay. All patients (male and female) require mutation analysis to confirm the genetic variant in that individual. Our subsequent approach is as follows:

- In males with virtually undetectable (<3 percent) alpha-Gal A leukocyte activity, a diagnosis of classic Fabry disease can be established. Genetic testing should then be performed. Genetic

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testing in this setting facilitates diagnosis and genetic counseling in the patient's family (particularly in females) and establishes the patient's amenability to treatment with chaperone therapy (i.e., migalastat).

- In males with decreased but detectable (3 to 35 percent) alpha-Gal A leukocyte activity, a diagnosis of Fabry disease is worth strong consideration since, as mentioned above, some hemizygous males and males with atypical variants may have alpha-Gal A leukocyte activity within this range. In such patients, genetic testing to search for a disease-causing mutation in the GLA gene should be performed to confirm the diagnosis for genetic counseling and to establish the patient's amenability to treatment with chaperone therapy. If no disease-causing mutation is identified, a diagnosis of Fabry disease can be ruled out. If the patient is found to have a genetic variant of unknown significance, and alpha-Gal A leukocyte activity is not clearly below 30 percent, biopsy of an affected tissue or organ (e.g., the kidney) with demonstration of elevated globotriaosylceramide (Gb3) by mass spectroscopy may be helpful in confirming the diagnosis.
- In males with alpha-Gal A leukocyte activity >35 percent, a diagnosis of Fabry disease cannot be established.

**Diagnosis in females--** In all female patients suspected of having Fabry disease, we perform genetic mutational analysis of the GLA gene as the initial diagnostic assay. Measurement of alpha-Gal A activity in heterozygous females is unreliable because heterozygotes have variable levels of alpha-Gal A activity that can overlap with levels found in healthy controls. Thus, genetic testing is required to make the diagnosis of Fabry disease among females.

If no disease-causing mutation is identified, a diagnosis of Fabry disease can be ruled out. If the patient is found to have a genetic variant of unknown significance, biopsy of an affected tissue or organ (e.g., the kidney) with demonstration of elevated Gb3 by mass spectroscopy may be helpful in confirming the diagnosis.

**Tests used in the diagnosis of Fabry disease--** The diagnosis of Fabry disease is typically established with a combination of biochemical and molecular genetic testing and/or by family history, although incidental findings on renal or cardiac biopsies may also lead to the diagnosis. **Enzymatic assay for alpha-Gal A activity--** After a thorough clinical evaluation, measurement of alpha-galactosidase A (alpha-Gal A) activity is the first step in the laboratory diagnosis of male patients suspected of having Fabry disease. Alpha-Gal A activity can be measured in leukocytes, plasma, fibroblasts, or dried blood spots (DBS), although measurement of leukocyte alpha-Gal A activity is the standard enzymatic test at most laboratories. The sensitivity and specificity of the alpha-Gal A assay using leukocytes approaches 100 percent in males, but the assay will identify less than 50 percent of female carriers. Analysis of plasma alpha-galactosidase may be less sensitive than assay of enzyme activity in leukocytes. Based upon the available knowledge, neither end-stage renal disease (ESRD) nor dialysis affects the enzyme assay.

Although different methods have been used to describe the results, enzymatic activity level is most often expressed as the percent of normal. The enzymatic level can vary by population tested:

- Alpha-Gal A activity in leukocytes is undetectable in over 50 percent of hemizygous males and is usually less than 4 percent of normal control levels in the remainder.
- Levels in female carriers range from normal to very low.

**Cardiac variants,** a form of atypical disease, have 1 to 30 percent of normal activity levels. **Genetic testing--** Mutational analysis of the alpha-Gal A (galactosidase alpha [GLA]) gene is the gold-standard assay to confirm the diagnosis of Fabry disease in males or females. Routine analysis, which consists of sequencing the coding region and exon-intron boundaries of the GLA gene, can detect a sequence variant in more than 97 percent of males and females with abnormal alpha-Gal A activity. A small number of mutations are not detected by routine analysis and require additional procedures such as gene-targeted deletion/duplication analysis for identification.

Genotyping is recommended for all Fabry families since this knowledge may be particularly relevant for identifying other affected members of the family and for future therapies utilizing synthetic

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chaperones. Since more than 800 distinct mutations have thus far been identified, identification of a mutation in a new family requires essentially complete resequencing of the gene. Genetic analysis is only done at selected laboratories.

Gb3 and lysoGb3 levels-- Globotriaosylceramide (Gb3) and globotriaosylsphingosine (lysoGb3), a degradation product of Gb3, can be detected in the plasma and urine of patients with Fabry disease and have been proposed as potential biomarkers for diagnosis and monitoring disease activity [82-84]. We do not routinely measure Gb3 or lysoGb3 levels in patients with suspected or confirmed Fabry disease, given their uncertain utility in the diagnosis and monitoring of patients with Fabry disease. The role of these markers in the diagnosis of patients with Fabry disease remains limited for the following reasons:

- Plasma levels of Gb3 are elevated in hemizygous males but are normal or only slightly elevated in heterozygous females
- Some, but not all, studies have shown a relationship between urine Gb3 levels and Fabry disease severity and response to treatment.
- Plasma levels of lysoGb3 are elevated in hemizygous males and, to a lesser extent, in heterozygous females with classic Fabry disease symptoms. However, plasma lysoGb3 levels do not correlate with disease manifestations in males and correlate only weakly with manifestations in females.
- Plasma lysoGb3 is elevated to a similar range in other lysosomal storage disorders such as Gaucher disease.

However, plasma and urine lysoGb3 levels, if elevated, may help to confirm the diagnosis of Fabry disease among patients who are found to have a GLA gene variant of uncertain significance. In one study of 124 patients with alpha-Gal A mutations, those with a novel GLA variant and organ involvement consistent with Fabry disease had plasma lysoGb3 levels  $\geq 2.7$  ng/mL. By contrast, those with a novel variant and no organ involvement had plasma lysoGb3 levels  $< 2.7$  ng/mL.

Plasma and urine lysoGb3 and Gb3 levels may be useful as pharmacodynamic markers for monitoring the effects of enzyme replacement therapy (ERT) or pharmacologic chaperone therapy (e.g., migalastat). As an example, in patients receiving ERT, these biomarkers may progressively increase when such patients develop neutralizing antibodies to the recombinant enzyme. In patients receiving a pharmacologic chaperone, monitoring plasma and urine lysoGb3 and Gb3 levels may be important to make sure that the chaperone does not excessively inhibit alpha-Gal Activity. Tissue Biopsy-- In some cases, biopsy of skin or, more rarely, culture of skin fibroblasts may be helpful in establishing the diagnosis but is usually done only if no other means of diagnosis are available. Skin biopsy can demonstrate the characteristic glycolipid deposits in a relatively noninvasive way but is not a reliable approach for confirming the diagnosis of significant organ involvement in Fabry disease. Kidney biopsy may be helpful in establishing the diagnosis. The diagnosis of Fabry disease is sometimes made serendipitously when a kidney biopsy is obtained to diagnose the cause of proteinuria and/or decreased kidney function. A kidney biopsy may be of particular use when patients have nephrotic syndrome, gross hematuria, or other symptoms that require exclusion of other diagnoses.

Endomyocardial biopsies are routinely performed in some centers where cardiac variants are the dominant form of Fabry disease. As previously mentioned, the renal or cardiac biopsy may be essential in confirming the diagnosis of Fabry disease in patients who have newly described GLA variants with unknown significance.

Follow-up assessment: Once diagnosed, patients with Fabry disease, or asymptomatic heterozygote females, should be followed closely using an interdisciplinary approach that involves routine care by nephrology, cardiology, and neurology, with input from dermatology, ophthalmology, and psychiatry as required

- Annual reevaluation with documentation of any clinical symptoms or signs. The annual exams should also include routine hematology and chemistry profiles, urinalysis, urinary protein-to-creatinine ratio or albumin-to-creatinine ratio, and an estimation of

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renal function such as estimated glomerular filtration rate.

- Echocardiography and electrocardiography to detect or monitor cardiac abnormalities at least every two years. Cardiac MRI may be required to assess fibrotic lesions in patients with or without overt LVH, especially in younger females for whom myocardial fibrosis may precede the development of LVH.
- Asymptomatic female carriers should also have a complete baseline evaluation as above and should be reevaluated every three to five years, with increasing frequency with age. Atypical males with Fabry disease should be evaluated and monitored annually similar to those classically affected.
- Asymptomatic female carriers should also have a complete baseline evaluation as above and should be reevaluated every three to five years, with increasing frequency with age. Atypical males with Fabry disease should be evaluated and monitored annually similar to those classically affected.
- Asymptomatic female carriers should also have a complete baseline evaluation as above and should be reevaluated every three to five years, with increasing frequency with age. Atypical males with Fabry disease should be evaluated and monitored annually similar to those classically affected.
- Asymptomatic female carriers should also have a complete baseline evaluation as above and should be reevaluated every three to five years, with increasing frequency with age. Atypical males with Fabry disease should be evaluated and monitored annually similar to those classically affected.

### CONTRAINDICATIONS/EXCLUSIONS/DISCONTINUATION:

All other uses of Fabrazyme (agalsidase beta) are considered experimental/investigational and therefore, will follow Molina's Off- Label policy. Contraindications to Fabrazyme (agalsidase beta) include: No labeled contraindications.

### OTHER SPECIAL CONSIDERATIONS:

Patients who have had a positive skin test to Fabrazyme or who have tested positive for anti-Fabrazyme IgE may be successfully rechallenged with Fabrazyme (Bodensteiner et al., 2008).

## CODING/BILLING INFORMATION

*Note: 1) This list of codes may not be all-inclusive. 2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement*

| HCPCS CODE | DESCRIPTION                     |
|------------|---------------------------------|
| J0180      | Injection, agalsidase beta, 1mg |

### AVAILABLE DOSAGE FORMS:

Fabrazyme SOLR 35MG single-dose vial  
Fabrazyme SOLR 5MG single-dose vial

## REFERENCES

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| SUMMARY OF REVIEW/REVISIONS  | DATE                       |
|--|----------------------------|
| REVISION- Notable revisions:<br>Continuation of Therapy<br>References  | Q3 2024                    |
| REVISION- Notable revisions:<br>Required Medical Information<br>Continuation of Therapy<br>Quantity<br>Available Dosage Forms<br>References  | Q3 2023                    |
| REVISION- Notable revisions:<br>Required Medical Information<br>Continuation of Therapy<br>Prescriber Requirements<br>Contraindications/Exclusions/Discontinuation<br>Other Special Considerations<br>References | Q3 2022                    |
| Q2 2022 Established tracking in new format   | Historical changes on file |