AvMed

MEDICAL PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-877-535-1391**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If information provided is not complete, correct, or legible, authorization can be delayed**.

For Medicare Members: 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: https://www.cms.gov/medicare-coverage-database/overview-and-quick-search.aspx. Additional indications may be covered at the discretion of the health plan.

Drug Requested: Fabrazyme® (agalsidase beta) **(IV INFUSION ONLY) (J0180) (Medical)**

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member AvMed #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Name/Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight (if applicable): _____ **Date weight obtained:** _____

❑ Standard Review. In checking this box, the timeframe does not jeopardize the life or health of the member or the member's ability to regain maximum function and would not subject the member to severe pain.

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Auhorization Approval: 6 months
MAXIMUM approved dose will be 1mg/kg infused every 2 weeks.

❑ **Member is ≥ 2 years of age**

❑ Provider is a specialist in genetics or metabolic disorders, a cardiologist or a nephrologist

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- ❑ Member has a diagnosis of Fabry disease **(also referred to as Anderson-Fabry disease)**
- ❑ Diagnosis of Fabry disease has been confirmed by **one** of the following:
  - ❑ **For males**: α-GAL A enzyme activity <1.5nmol/hr/mL in plasma or <4nmol/hr/mL in isolated leukocytes **AND** documentation of disease-causing mutation in GLA gene located on Xq22.1 (labs must be submitted)
  - ❑ **For females**: documentation of disease-causing mutation in GLA gene located on Xq22.1 (lab must be submitted) **AND** documentation of clinically significant organ involvement (i.e., symptomatic cardiac disease, renal impairment, TIA or stroke history) must be submitted; symptoms must not be attributable to any other causes
- ❑ Baseline plasma globotriaosylceramide (GL-3) level must be submitted
- ❑ Baseline plasma or urinary sediment lyso-Gb3 level must be submitted
- ❑ Member must be taking appropriate prophylaxis/treatment medications for the following:
  - ❑ **RENAL:**
    - ❑ Current pharmacy claims for ACE inhibitor or angiotensin receptor blocker (ARB) therapy must be noted for members with proteinuria
  - ❑ **NEUROLOGICAL:**
    - ❑ Members with history of TIA or thrombotic stroke must have current pharmacy claims for antiplatelet therapy (i.e. clopidogrel, aspirin, prasugrel; etc.)
  - ❑ **CARDIAC:**
    - ❑ Pharmacy claims for ACE-I, calcium channel blocker, ARB, or antiplatelet therapy must be noted if member has documented valvular insufficiency, shortened PR interval, diastolic dysfunction, resting bradycardia or <EF
    - ❑ Current pharmacy claims for statin or other hyperlipidemia therapy must be noted for treatment of elevated lipids
  - ❑ **PULMONARY:**
    - ❑ Pharmacy claims for bronchodilator therapy must be noted for members with pulmonary symptoms
  - ❑ **ACROPARESTHESIA Monitoring:**
    - ❑ Pharmacy claims for gabapentin, carbamazepine, topiramate, oxcarbazepine, phenytoin or other anticonvulsant therapy must be noted for acroparesthesia treatment

Exclusion criteria: Well characterized benign GLS polymorphisms; absence of demonstrable Fabry disease-related tissue pathology or clinical symptoms; development of ESRD, without an option for renal transplantation, in combination with advanced heart failure (New York Heart Association class IV); current hemodialysis therapy; history of renal transplantation; persistent life-threatening or severe infusion reactions that do not respond to prophylaxis **(e.g., anaphylaxis)**; end-stage Fabry disease or other comorbidities with a life expectancy of <1 year

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Reauthorization Approval: 6 months. All criteria must be checked for approval. To support each line checked, all documentation (lab results, diagnostics, and/or chart notes) must be provided or request may be denied. **MAXIMUM approved dose will be 1mg/kg infused every 2 weeks.**

- ❑ Provider is a specialist in genetics or metabolic disorders, a cardiologist or a nephrologist
- ❑ Current plasma globotriaosylceramide (GL-3) level must be submitted and must have decreased from baseline level
- ❑ Current plasma or urinary sediment lyso-Gb3 level must be submitted and must have decreased from baseline level
- ❑ Current IgG anti-agalsidase antibody titer must be submitted
- ❑ Chart notes and labs for all criteria listed must be submitted to document clinical improvement or stabilization in member's renal, cardiac, cerebrovascular, pulmonary function and pain levels from baseline
- ❑ Member must be taking appropriate prophylaxis/treatment medications for member's renal, cardiac, cerebrovascular, pulmonary function and pain levels if applicable from baseline

Exclusion criteria: Well characterized benign GLS polymorphisms; absence of demonstrable Fabry disease-related tissue pathology or clinical symptoms; development of ESRD, without an option for renal transplantation, in combination with advanced heart failure (New York Heart Association class IV); current hemodialysis therapy; history of renal transplantation; persistent life-threatening or severe infusion reactions that do not respond to prophylaxis **(e.g., anaphylaxis)**; end-stage Fabry disease or other comorbidities with a life expectancy of <1 year.

Medication being provided by (check below that applies) - Limited Distribution Drug

- ❑ **Location/site of drug administration:** _____

  NPI or DEA # of administering location: _____

  OR
- ❑ **Specialty Pharmacy - PropriumRx**

  OR
- ❑ **Specialty Pharmacy**: _____

For urgent reviews: Practitioner should call AvMed Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. AvMed's definition of urgent is a lack of treatment that could seriously jeopardize the life or health of the member or the member's ability to regain maximum function.

Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.*

Previous therapies will be verified through pharmacy paid claims or submitted chart notes.*

***Approved by Pharmacy and Therapeutics Committee:** 4/3/2019
REVISED/UPDATED/REFORMATTED: ~~5/10/2019~~; ~~7/6/2019~~; ~~10/7/2019~~; **11/8/2021**;