Geisinger

POLICIES AND PROCEDURE MANUAL

Policy: MBP 248.0

Section: Medical Benefit Pharmaceutical Policy

Subject: Nexviazyme (avalglucosidase alfa-ngpt)

I. Policy:
Nexviazyme (avalglucosidase alfa-ngpt)

II. Purpose/Objective:
To provide a policy of coverage regarding Nexviazyme (avalglucosidase alfa-ngpt).

III. Responsibility:
A. Medical Directors
B. Medical Management
C. Pharmacy Department

IV. Required Definitions
1. Attachment – a supporting document that is developed and maintained by the policy writer or department requiring/authoring the policy.
2. Exhibit – a supporting document developed and maintained in a department other than
3. the department requiring/authoring the policy.
4. Devised – the date the policy was implemented.
5. Revised – the date of every revision to the policy, including typographical and grammatical changes.
6. Reviewed – the date documenting the annual review if the policy has no revisions necessary.

V. Additional Definitions
Medical Necessity or Medically Necessary means Covered Services rendered by a Health Care Provider that the Plan determines are:

a. appropriate for the symptoms and diagnosis or treatment of the Member's condition, illness, disease or injury;
b. provided for the diagnosis and the direct care and treatment of the Member's condition, illness disease or injury;
c. in accordance with current standards good medical treatment practiced by the general medical community;
d. not primarily for the convenience of the Member, or the Member's Health Care Provider; and
e. the most appropriate source or level of service that can safely be provided to the Member. When applied to hospitalization, this further means that the Member requires acute care as an inpatient due to the nature of the services rendered or the Member's condition, and the Member cannot receive safe or adequate care as an outpatient

Medicaid Business Segment
Medically Necessary — A service, item, procedure, or level of care compensable under the Medical Assistance program that is necessary for the proper treatment or management of an illness, injury, or disability is one that:

i. Will, or is reasonably expected to, prevent the onset of an illness, condition, injury or disability.
ii. Will, or is reasonably expected to, reduce or ameliorate the physical, mental or developmental effects of an illness, condition, injury or disability.
iii. Will assist the Member to achieve or maintain maximum functional capacity in performing daily activities, taking into account both the functional capacity of the Member and those functional capacities that are appropriate for Members of the same age.

DESCRIPTION:
Nexviazyme (Avalglucosidase alfa-ngpt) is an exogenous source of the enzyme acid alpha-glucosidase (GAA), which is required for glycogen cleavage. Due to an inherited GAA deficiency or absence, glycogen accumulates in the tissues of patients with Pompe disease. Mannose-6-phosphate (M6P) on avalglucosidase alfa mediates binding to M6P receptors on the cell surface with high affinity. After binding, it is internalized and transported to lysosomes where it is activated for increased enzymatic glycogen cleavage.

CRITERIA FOR USE: Requires Prior Authorization by Medical Director or Designee

Nexviazyme (avalglucosidase alfa-ngpt) will be considered medically necessary for all lines of business when all of the following criteria are met:

- Medical record documentation of a diagnosis of late-onset Pompe disease supported by:
    - Acid alpha-glucosidase (GAA) assay performed on dried blood spots, skin fibroblasts or muscle biopsy **AND**
    - Genetic testing showing a mutation in the GAA gene

  AND
- Medical record documentation of a consultation with a metabolic specialist and/or biochemical geneticist **AND**
- Medical record documentation of age greater than or equal to 1 year **AND**
- Medical record documentation of baseline percent-predicted forced vital capacity (% FVC) and 6-minute walk test (6MWT), if age appropriate **AND**
- Medical record documentation that the member is receiving an appropriate dose* based on patient's weight **AND**
- Medical record documentation that Nexviazyme will not be used in combination with other enzyme replacement therapy (e.g. Lumizyme)

AUTHORIZATION DURATION: Initial approval will be for 12 months or less if the reviewing provider feels it is medically appropriate. Subsequent approvals will be for an additional 12 months or less if the reviewing provider feels it is medically appropriate and will require the following:

- Medical record documentation of improvement or stabilization in percent-predicted forced vital capacity (% FVC) and/or 6-minute walk test (6MWT) **AND**
- Medical record documentation that the member is receiving an appropriate dose* based on patient's weight **AND**
- Medical record documentation that Nexviazyme will not be used in combination with other enzyme replacement therapy (e.g. Lumizyme)

*Note to reviewing pharmacist: For patients weighing ≥30 kg, the recommended dosage is 20 mg/kg (of actual body weight) every two weeks. For patients weighing < 30 kg, the recommended dosage is 40 mg/kg (of actual body weight) every two weeks.

Note: For Medicaid (GHP Family), any requests for services that do not meet criteria set in the PARP will be evaluated on a case-by-case basis.

LINE OF BUSINESS:
Eligibility and contract specific benefit limitations and/or exclusions will apply. Coverage statements found in the line of business specific benefit document will supersede this policy.

This policy will be revised as necessary and reviewed no less than annually**.**

Devised: 1/18/22

Revised: 1/17/23 (LOB carve out, Medicaid PARP statement, Medicaid business segment)

Reviewed: