Medical Policy Bulletin

Title:

Alglucosidase alfa (e.g., Lumizyme®), Avalglucosidase alfa-ngpt (Nexviazyme®), Cipaglucosidase alfa-atga (Pombiliti™)

Policy #:

MA08.036g

The Company makes decisions on coverage based on the Centers for Medicare and Medicaid Services (CMS) regulations and guidance, benefit plan documents and contracts, and the member's medical history and condition. If CMS does not have a position addressing a service, the Company makes decisions based on Company Policy Bulletins. Benefits may vary based on contract, and individual member benefits must be verified. The Company determines medical necessity only if the benefit exists and no contract exclusions are applicable. Although the Medicare Advantage Policy Bulletin is consistent with Medicare's regulations and guidance, the Company's payment methodology may differ from Medicare.

When services can be administered in various settings, the Company reserves the right to reimburse only those services that are furnished in the most appropriate and cost-effective setting that is appropriate to the member's medical needs and condition. This decision is based on the member's current medical condition and any required monitoring or additional services that may coincide with the delivery of this service.

This Policy Bulletin document describes the status of CMS coverage, medical terminology, and/or benefit plan documents and contracts at the time the document was developed. This Policy Bulletin will be reviewed regularly and be updated as Medicare changes their regulations and guidance, scientific and medical literature becomes available, and/or the benefit plan documents and/or contracts are changed.

Policy

Coverage is subject to the terms, conditions, and limitations of the member's Evidence of Coverage.

MEDICALLY NECESSARY

ALGLUCOSIDASE ALFA (LUMIZYME®)
Alglucosidase alfa (Lumizyme®) is considered medically necessary and, therefore, covered for the treatment of symptomatic individuals 1 month of age and older with infantile-onset or juvenile/adult-onset Pompe disease (lysosomal acid alpha-glucosidase [GAA] deficiency) when all of the following criteria are met, including dosing and frequency:

- Diagnosis is confirmed by either of the following:
  - Deficiency of GAA in leukocytes or skin fibroblasts or muscles
  - Confirmation of biallelic pathogenic variant(s) in the GAA gene
- Dosing and frequency: infants 1 month of age and older, children, adolescents and adult dosing IV: 20 mg/kg every 2 weeks

AVALGLUCOSIDASE ALFA-NGPT (NEXVIAZYME®)
Avalglucosidase alfa-ngpt (Nexviazyme®) is considered medically necessary and, therefore, covered for the treatment of individuals 1 year of age and older with late-onset Pompe disease (LOPD)(lysosomal GAA deficiency) when all of the following criteria are met, including dosing and frequency:

- The participant has confirmed GAA enzyme deficiency from any tissue source (e.g., skin, fibroblasts, muscle, or blood) and/or two confirmed GAA gene mutations.
- Dosing and frequency:
  - ≥30 kg, the recommended dosage is 20 mg/kg (of actual body weight) every 2 weeks.
  - <30 kg, the recommended dosage is 40 mg/kg (of actual body weight) every 2 weeks.

CIPAGLUCOSIDASE ALFA-ATGA (POMBILITI)
Cipaglucosidase alfa-atga (Pombiliti) is considered medically necessary and, therefore, covered for the treatment of adult individuals with diagnosis of LOPD when all of the following criteria are met, including dosing and frequency:

- Diagnosis of LOPD (e.g., presence of clinical signs and symptoms of the disease respiratory distress, skeletal muscle weakness) confirmed by either of the following:

    - Deficiency of GAA enzyme from any tissue sources (e.g., leukocytes or skin fibroblasts or muscles)
    - Confirmation of biallelic pathogenic variant(s) in the GAA gene
- Individual's weight is greater than or equal to 40 kg
- Inadequate response or inability to tolerate an enzyme replacement therapy for Pompe disease (PD) (e.g., alglucosidase alfa or avalglucosidase alfa-ngpt)
- Medication must not be used in combination with other miglustat products (i.e., Zavesca, Yargesa)
- Dosing and frequency: 20 mg/kg (of actual body weight) administered every other week as an intravenous infusion

Note: medication must be used in combination with oral miglustat (Opfolda).

EXPERIMENTAL/INVESTIGATIONAL

All other uses of alglucosidase alfa (Lumizyme®) and avalglucosidase alfa-ngpt (Nexviazyme®) are considered experimental/investigational and, therefore, not covered unless the indication is supported as an accepted off-label use, as defined in the Company medical policy on off-label coverage for prescription drugs and biologics.

DOSING AND FREQUENCY REQUIREMENTS

The Company reserves the right to modify the Dosing and Frequency Requirements listed in this policy to ensure consistency with the most recently published recommendations for the use of alglucosidase alfa (Lumizyme®) and avalglucosidase alfa-ngpt (Nexviazyme®). Changes to these guidelines are based on a consensus of information obtained from resources such as, but not limited to: the US Food and Drug Administration (FDA); Company-recognized authoritative pharmacology compendia; or published peer-reviewed clinical research. The professional provider must supply supporting documentation (i.e., published peer-reviewed literature) in order to request coverage for an amount of alglucosidase alfa (Lumizyme®) and avalglucosidase alfa-ngpt (Nexviazyme®) outside of the Dosing and Frequency Requirements listed in this policy. For a list of Company-recognized pharmacology compendia, view our policy on off-label coverage for prescription drugs and biologics.

Accurate member information is necessary for the Company to approve the requested dose and frequency of this drug. If the member’s dose, frequency, or regimen changes (based on factors such as changes in member weight or incomplete therapeutic response), the provider must submit those changes to the Company for a new approval based on those changes as part of the utilization management activities. The Company reserves the right to conduct postpayment review and audit procedures for any claims submitted for alglucosidase alfa (Lumizyme®) and avalglucosidase alfa-ngpt (Nexviazyme®).

REQUIRED DOCUMENTATION

The individual's medical record must reflect the medical necessity for the care provided. These medical records may include, but are not limited to: records from the professional provider's office, hospital, nursing home, home health agencies, therapies, and test reports.

The Company may conduct reviews and audits of services to our members, regardless of the participation status of the provider. All documentation is to be available to the Company upon request. Failure to produce the requested information may result in a denial for the drug.

When coverage of alglucosidase alfa (Lumizyme®) or avalglucosidase alfa-ngpt (Nexviazyme®) is requested outside of the Dosing and Frequency Requirements listed in this policy, the prescribing professional provider must supply documentation (i.e., published peer-reviewed literature) to the company that supports this request.

Guidelines

Per the US Food and Drug Administration (FDA)–approved labeling, the recommended dosing and frequency of alglucosidase alfa (Lumizyme®) is 20 mg/kg weight administered by intravenous (IV) infusion every 2 weeks.

Per the FDA-approved labeling, the recommended dosing and frequency of (Nexviazyme) avalglucosidase alfa-ngpt is 1 mg/kg of body weight administered once weekly as an intravenous infusion.

Per the FDA-approved labeling, the recommended dosing and frequency of cipaglucosidase alfa-atga (Pombiliti) is 20 mg/kg (of actual body weight) administered every other week as an intravenous infusion.

BLACK BOX WARNING

Refer to the specific manufacturer's prescribing information for any applicable Black Box Warnings.

BENEFIT APPLICATION

Subject to the terms and conditions of the applicable benefit contract, alglucosidase alfa (Lumizyme®), avalglucosidase alfa-ngpt (Nexviazyme®), and cipaglucosidase alfa-atga (Pombiliti) are covered under the medical benefits of the Company's products when the medical necessity criteria including dosing and frequency requirements listed in this medical policy are met.

US FOOD AND DRUG ADMINISTRATION (FDA) STATUS

On May 10, 2010, under a priority review, the FDA approved alglucosidase alfa (Lumizyme®) as an orphan drug. On August 26, 2021, under a priority review, the FDA approved avalglucosidase alfa-ngpt (Nexviazyme) as an orphan drug. On September 28, 2023, FDA approved cipaglucosidase alfa-atga (Pombiliti) + Opfolda (miglustat) 65 mg capsules as two-component therapy.

PEDIATRIC USE

The safety and effectiveness of alglucosidase alfa (Lumizyme) have not been established in individuals younger than 1 month of age. The safety and effectiveness of avalglucosidase alfa-ngpt (Nexviazyme) have not been established in individuals younger than 1 year of age. The safety and effectiveness of cipaglucosidase alfa-atga (Pombiliti) have not been established in individuals younger than 18 years of age.

Description

Pompe disease is a rare genetic disorder of glycogen metabolism that is caused by the absence or marked deficiency of the lysosomal enzyme acid alpha-glucosidase (GAA). This disease is also known as glycogen storage disease type II, GSD II, glycogenosis type II, or acid maltase deficiency. GAA is necessary for proper muscle functioning and is used by the heart and muscle cells to convert a form of sugar called glycogen into energy. Without the GAA enzyme action, glycogen builds up in the cells of the heart, skeletal muscles, and hepatic tissues. Ultimately, these body organs are weakened by the intralysosomal accumulation of glycogen. Pompe disease encompasses a range of phenotypes, each including myopathy, but with significant variability in the age of onset, organ involvement, and clinical severity.

Infantile-onset Pompe disease occurs in an estimated one in every 40,000 to 300,000 births. Symptoms begin in the first months of life, with feeding problems, poor weight gain, muscle weakness, floppiness, and head lag. The primary symptom is heart and skeletal muscle weakness, which leads to the development of cardiomyopathy, progressing respiratory weakness, and death, usually from respiratory failure. Younger individuals generally have a much more aggressive form of the disease.

Juvenile/adult-onset Pompe disease results in intralysosomal accumulation of glycogen that is limited primarily to skeletal muscle, resulting in progressive muscle weakness. The onset can be as early as the first decade of childhood or as late as the sixth decade of adulthood. The primary symptom is muscle weakness progressing to respiratory weakness and death from respiratory failure over several years. The heart is usually spared.

Enzyme-replacement therapy has been shown to decrease heart size, maintain normal heart function, improve muscle function, tone, and strength, and reduce glycogen accumulation. Enzyme replacement therapy is currently not recommended for individuals with no symptoms or objective signs (proximal muscle weakness or reduced forced vital capacity [FVC] in either upright or supine position) of Pompe disease.

Alglucosidase alfa (Lumizyme®) is FDA-approved for individuals with Pompe disease (GAA deficiency). The safety and efficacy was assessed in 57 treatment-naïve individuals with infantile-onset Pompe disease, aged 0.2 months to 3.5 years at first infusion, in three separate clinical trials. In all three trials, ventilator-free survival improved significantly compared with an untreated historical control. The safety and efficacy of alglucosidase alfa (Lumizyme®) was also assessed in 90 individuals with juvenile/adult-onset Pompe disease in a randomized, double-blinded, placebo-controlled trial. Alglucosidase alfa (Lumizyme®) was shown to have a significant increase in FVC and the

distance an individual with juvenile/adult-onset Pompe disease can walk within 6 minutes (6-minute walk test).

Avalglucosidase alfa-ngpt (Nexviazyme®) is a hydrolytic lysosomal glycosaminoglycan (GAG)-specific enzyme that is FDA approved for individuals with mucopolysaccharidosis VI (MPS VI; Maroteaux-Lamy syndrome).

The safety analysis was pooled from four clinical trials (mean exposure of 26 months, up to 85 months of treatment) that included 141 avalglucosidase alfa-ngpt (Nexviazyme®)-treated individuals (118 adult and 23 pediatric individuals). Serious adverse reactions reported in two or more individuals treated with avalglucosidase alfa-ngpt (Nexviazyme®) were: respiratory distress, chills, and pyrexia. Serious adverse events were similar across both adult and pediatric populations. Five avalglucosidase alfa-ngpt (Nexviazyme®)-treated individuals in clinical trials permanently discontinued the medication due to adverse reactions, including two of these individuals who discontinued the treatment because of a serious adverse reaction. The most frequently reported adverse reactions (>5%) in the pooled safety population were: headache, diarrhea, nausea, fatigue, arthralgia, myalgia, dizziness, rash, vomiting, pyrexia, abdominal pain, pruritus, erythema, abdominal pain upper, chills, cough, urticaria, dyspnea, hypertension, and hypotension.

The safety and efficacy of cipaglucosidase alfa-atga (Pombiliti) was investigated in a randomized, double-blind PROPEL study (ATB200-03; NCT03729362) that compared investigational enzyme replacement therapy (ERT) cipaglucosidase alfa+miglustat (cipa+mig) with alglucosidase alfa+placebo (alg) in adult individuals with late-onset Pompe disease (LOPD). ERT-experienced individuals that remained on alg (n=30) showed worsening or stability. Individuals who switched to cipa+mig (n=65) showed improvement or stability. Individuals who remained on alg demonstrated statistically significant within-group worsening for sitting and supine forced vital capacity; slow vital capacity; maximal expiratory pressure; and creatine kinase (CK) and hexose tetrasaccharide (Hex4) levels, and no significant improvements for any outcomes. Individuals that switched to cipa+mig did not demonstrate significant within-group worsening for any outcomes and showed significant improvements for 6-minute walk distance (absolute and % predicted); upper, lower and overall manual muscle test; PROMIS fatigue; physician and subject global impression of change (five of eight subdomains); and CK and Hex4 levels.

There may be additional indications contained in the Policy section of this document due to evaluation of criteria highlighted in the Company's off-label policy, and/or review of clinical guidelines issued by leading professional organizations and government entities.

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Coding

Inclusion of a code in this table does not imply reimbursement. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

The codes listed below are updated on a regular basis, in accordance with nationally accepted coding guidelines. Therefore, this policy applies to any and all future applicable coding changes, revisions, or updates.

In order to ensure optimal reimbursement, all health care services, devices, and pharmaceuticals should be reported using the billing codes and modifiers that most accurately represent the services rendered, unless otherwise directed by the Company.

The Coding Table lists any CPT, ICD-10, and HCPCS billing codes related only to the specific policy in which they appear.

CPT Procedure Code Number(s)
N/A

ICD - 10 Procedure Code Number(s)
N/A

ICD - 10 Diagnosis Code Number(s)
E74.02 Pompe disease

HCPCS Level II Code Number(s)
G0138 Intravenous infusion of cipaglucosidase alfa-atga, including provider/supplier acquisition and clinical supervision of oral administration of miglustat in preparation of receipt of cipaglucosidase alfa-atga
J0219 Injection, avalglucosidase alfa-ngpt, 4 mg
J0221 Injection, avalglucosidase alfa-ngpt, 4 mg
J1203 Injection, cipaglucosidase alfa-atga, 5 mg

NOT ELIGIBLE FOR REIMBURSEMENT

J0220 Injection, alglucosidase alfa, 10 mg, not otherwise specified

Revenue Code Number(s)
N/A

Policy History

Revisions From MA08.036g:

12/15/2025	This policy has been reissued in accordance with the Company's annual review process.
03/28/2025	The policy has been reviewed and reissued to communicate the Company’s continuing position on Alglucosidase alfa (e.g., Lumizyme®), Avalglucosidase alfa-ngpt (Nexviazyme®), Cipaglucosidase alfa-atga (Pombiliti™)

04/22/2024	This policy has been reissued in accordance with the Company's annual review process. This version of the policy will become effective 04/22/2024 **The title was changed FROM**: Alglucosidase alfa (e.g., Lumizyme®), Avalglucosidase alfa-ngpt (Nexviazyme®) **TO:** Alglucosidase alfa (e.g., Lumizyme®), Avalglucosidase alfa-ngpt (Nexviazyme®), Cipaglucosidase alfa-atga (Pombiliti™) This policy has been updated to communicate dosing and frequency requirements for new FDA-approved avalglucosidase Cipaglucosidase alfa-atga (Pombiliti™) THE FOLLOWING HCPCS CODE IS ADDED TO REPRESENT Cipaglucosidase alfa-atga (Pombiliti™): J1203 Injection, cipaglucosidase alfa-atga, 5 mg G0138 Intravenous infusion of cipaglucosidase alfa-atga including provider/supplier acquisition and clinical supervision of oral administration of miglustat in preparation of receipt of cipaglucosidase alfta-atga

Revisions from MA08.036f

09/05/2023	This policy has been reissued in accordance with the Company's annual review process.
05/04/2022	This policy has been reissued in accordance with the Company's annual review process.
04/01/2022	This version of the policy will become effective 04/01/2022. The following HCPCS codes have been **termed** from this policy: C9085 Injection, avalglucosidase alfa-ngpt, 4 mg J3590 Unclassified biologics The following HCPCS code has been **added** in this policy: J0219 Injection, avalglucosidase alfa-ngpt, 4 mg

Revisions from MA08.036e

01/01/2022	This policy has been identified for the HCPCS code update, effective 01/01/2022. The following HCPCS codes have been **termed** from this policy: C9399 Unclassified drugs or biologicals The following HCPCS code has been **added** to this policy: C9085 Injection, avalglucosidase alfa-ngpt, 4 mg

Revisions From MA08.036d:

09/27/2021	This policy has been reissued in accordance with the Company's annual review process. This version of the policy will become effective 09/27/2021 **The title was changed FROM**: Alglucosidase alfa (e.g., Lumizyme®) **TO:** Alglucosidase alfa (e.g., Lumizyme®), Avalglucosidase alfa-ngpt (Nexviazyme®) This policy has been updated to communicate dosing and frequency requirements for new FDA-approved avalglucosidase alfa-ngpt (Nexviazyme®) and requirements for laboratory and/or genetic testing consistent with the US Food and Drug Administration (FDA) labeling. THE FOLLOWING CODES ARE ADDED TO REPRESENT AVALGLUCOSIDASE ALFA-NGPT (NEXVIAZYME®): C9399 Unclassified drugs or biologicals

	J3590 Unclassified biologics

Revisions From MA08.036c:

04/08/2020	This policy has been reissued in accordance with the Company's annual review process.
06/03/2019	This version of the policy will become effective 06/03/2019. This policy has been updated to communicate dosing and frequency requirements for alglucosidase alfa (e.g., Lumizyme®) and requirements for laboratory and/or genetic testing consistent with the US Food and Drug Administration (FDA) labeling.

Revisions From MA08.036b:

06/06/2018	This policy has been reissued in accordance with the Company's annual review process.
01/22/2018	This version of the policy will become effective 01/22/2018. This policy has been updated to be consistent with the US Food and Drug Administration (FDA) labeling and NCCN compendia. This policy was updated to remove alglucosidase alfa (Myozyme®), which was discontinued in 2013. The title was also changed to remove "Myozyme".

Revisions From MA08.036a:

06/07/2017	The policy has been reviewed and reissued to communicate the Company’s continuing position on Alglucosidase alfa (e.g., Myozyme®, Lumizyme®).
03/16/2016	The policy has been reviewed and reissued to communicate the Company’s continuing position on Alglucosidase alfa (e.g., Myozyme®, Lumizyme®).
02/11/2015	This policy has been updated to be consistent with the US Food and Drug Administration (FDA) labeling and Drug Compendia. The criteria for alglucosidase alfa (Lumizyme®) was updated to include individuals with infantile-onset or juvenile/adult-onset Pompe disease under the age of eight years.

Revisions From MA08.036:

01/01/2015	This is a new policy.

Version Effective Date:
12/15/2025
Version Issued Date:
12/15/2025
Version Reissued Date:
N/A