EVH Clinical Guideline 5008.CC for Duchenne Muscular Dystrophy Drug Therapies

Guideline Number: EVH_CG_5008.CC	**Applicable Codes**	
"Evolent" refers to Evolent Health LLC and Evolent Specialty Services, Inc. 		
Original Date: January 2024	**Last Revised Date:** January 2026	**Implementation Date:** March 2026

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STATEMENT

General Information

- *It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity, and approval by the Medical Policy Committee.*
- *If the established criteria are not met, the request is referred to a Medical Director for review, if required for the plan and level of request.*

Purpose

The purpose of this guideline is to define the prior authorization process for the following drug(s): Amondys 45 (casimersen), Elevidys (delandistrogene moxeparvovec-rokl), and Vyondys 53 (golodirsen).

Scope

This guideline applies to all practitioners who are involved in providing the requested drug. This guideline is specific to the Health Plan's medical benefit.

INITIAL REVIEW CRITERIA

The request must meet all of the criteria listed under the drug-specific sections below.

Amondys 45 & Vyondys 53

- Must have a diagnosis of Duchenne Muscular Dystrophy (DMD) with a confirmed mutation of a DMD gene that is amenable to:
  - Exon 45 skipping (Amondys 45)
  - Exon 53 skipping (Vyondys 53)
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
- Must be prescribed by, or in consultation with, a provider board certified in ONE of the following specialties:
  - Neurology
  - Orthopedics
  - Physical medicine and rehabilitation
  - Neuromuscular medicine
  - Neurodevelopmental disabilities

- Must have documentation of ALL the following:
  - Current weight
  - 6-Minute Walk Test (6MWT) if ambulatory
  - Brooke Upper Extremity (BUE) Function score of ≤5
  - Stable pulmonary function with Forced Vital Capacity (FVC) ≥30% predicted
  - Urinalysis showing absence of proteinuria
  - Blood Urea Nitrogen (BUN)
  - Serum Creatinine (SCr)
- Must have tried (and been adherent) to standard corticosteroid therapy for a minimum of 6 months OR must have justification for discontinuation of standard therapy
- Must have documentation of the goals of therapy

Elevidys

- Must be at least 4 years of age
- Must have a diagnosis of Duchenne Muscular Dystrophy (DMD) with documentation of the following:
  - A mutation of the DMD gene
  - The mutation is NOT a deletion in exon 8 or exon 9
- Must be prescribed by, or in consultation with, a provider board certified in ONE of the following specialties:
  - Neurology
  - Orthopedics
  - Physical medicine and rehabilitation
  - Neuromuscular medicine
  - Neurodevelopmental disabilities
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
- Must have a baseline anti-AAVrh74 total binding antibody titer of < 1:400
- Must have attestation from the provider that:
  - The member will initiate a corticosteroid regimen prior to infusion and that treatment will continue for a minimum of 60 days after infusion
  - The member will not be receiving exon-skipping therapies (e.g., Amondys 45, Exondys 51) for DMD concomitantly or following Elevidys treatment
  - The member has not received Elevidys previously

REAUTHORIZATION CRITERIA

ELEVIDYS is not eligible for reauthorization as only a single course is allowed per member's lifetime.

AMONDYS 45 & VYONDYS 53 ***ONLY:*** All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. The request must meet all of the criteria listed below.

- Must continue to be adherent to the requested medication – infusion records are required to support adherence
- Must have a clinical response to therapy, based upon the prescriber's assessment while on therapy
- Must submit current:
  - Weight
  - 6MWT if ambulatory
  - BUE Function score of ≤5
  - FVC ≥30% predicted
  - Urinalysis showing absence of proteinuria
  - BUN
  - SCr
- Must have continued standard therapy previously initiated (if appropriate/tolerated)

APPROVAL DURATIONS

Initial Authorization	**Amondys 45 & Vyondys 53:** Up to 1 year **Elevidys:** Up to 6 months
Reauthorization	**Amondys 45 & Vyondys 53:** Same as initial **Elevidys:** N/A

CODING AND STANDARDS

Codes

Code	Brand	Description
J1413	Elevidys	Injection, delandistrogene moxeparvovec-rokl, per therapeutic dose
J1426	Amondys 45	Injection, casimersen, 10mg
J1429	Vyondys 53	Injection, golodirsen, 10mg

Applicable Lines of Business

☐	CHIP (Children's Health Insurance Program)
☐	Commercial
☐	Exchange/Marketplace
☒	Medicaid
☐	Medicare Advantage

BACKGROUND

Amondys 45 (casimersen) is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 45 skipping.

Vyondys 53 (golodirsen) is indicated for the treatment of Duchenne muscular dystrophy (DMD) in patients who have a confirmed mutation of the DMD gene that is amenable to exon 53 skipping.

Elevidys (delandistrogene moxparvovec-rokl) is indicated for the treatment of pediatric patients at least 4 years of age with Duchenne muscular dystrophy (DMD) with a confirmed mutation in the DMD gene. NOTE: The indication specifically in the non-ambulatory population is approved under accelerated approval based on expression of ELEVIDYS microdystrophin in skeletal muscle observed in patients treated with ELEVIDYS. Continued approval for this indication may be contingent upon verification and description of clinical benefit in a confirmatory trial(s).

For Exondys 51 (eteplirsen) – refer to the HFS criteria at **https://hfs.illinois.gov/content/dam/soi/en/web/hfs/sitecollectiondocuments/exondys51webcriteria.pdf**

Definitions

Duchenne muscular dystrophy (DMD) - is a rare, X-linked, recessive, life-threatening, degenerative neuromuscular disease affecting males. It is attributed to mutations in the DMD gene (chromosome Xp21), which is responsible for producing the protein dystrophin. Dystrophin is needed for proper muscle functioning and provides mechanical stability to muscle fibers during muscle contraction. The absence of, or defect in, this protein leads to progressive muscle degeneration with loss of independent ambulation, as well as respiratory and cardiac complications.

POLICY HISTORY

Date	Summary
January 2026	• Updated Elevidys approval duration to 6 months
October 2024	• Updated Elevidys age & ambulatory requirements
April 2024	• Added Elevidys
January 2024	• New Guidelines

LEGAL AND COMPLIANCE

Guideline Approval

Committee

Reviewed / Approved by Evolent Administrative Services Medical Policy Committee

Disclaimer

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REFERENCES

1. Vyondys 53 [prescribing information]. Sarepta Therapeutics, Cambridge, MA, June 2024.
2. Amondys 45 [prescribing information]. Sarepta Therapeutics, Cambridge, MA, July 2024.
3. Elevidys (delandistrogene moxeparvovec) [prescribing information]. Cambridge, MA: Sarepta Therapeutics Inc; September 2024.
4. Mendell JR, Shieh PB, McDonald CM, et al. Expression of SRP-9001 dystrophin and stabilization of motor function up to 2 years post-treatment with delandistrogene moxeparvovec gene therapy in individuals with Duchenne muscular dystrophy. Front Cell Dev Biol. 2023;11:1167762. doi:10.3389/fcell.2023.1167762 [PubMed 37497476]
5. Zaidman CM, Proud CM, McDonald CM, et al. Delandistrogene moxeparvovec gene therapy in ambulatory patients (aged ≥4 to <8 years) with duchenne muscular dystrophy: 1-year interim results from study SRP-9001-103 (ENDEAVOR). Ann Neurol. 2023;94(5):955-968. doi:10.1002/ana.26755 [PubMed 37539981]