EVH Clinical Guideline 5105.CC for Lenmeldy

Guideline Number:	Applicable Codes	
EVH_CG_5105.CC		
"Evolent" refers to Evolent Health LLC and Evolent Specialty Services, Inc. 		
Original Date:	**Last Revised Date:**	**Implementation Date:**
September 2025	June 2025	September 2025

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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 Lenmeldy (atidarsagene autotemcel) for the diagnosis of metachromatic leukodystrophy (MLD).

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 General Criteria ***and*** diagnosis-specific sections below.

General Criteria

- Must be prescribed by a hematologist, a neurologist, genetics specialist, or a stem cell transplant specialist
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling, based on the member's MLD disease subtype
- Must have documentation of ALL the following to confirm the diagnosis of metachromatic leukodystrophy (MCLD):
    - Genetic testing results that show bi-allelic mutations in the *ARSA* gene
    - Elevated urinary sulfatide levels in a 24-hour collection
    - ARSA enzyme activity below normal range in leukocytes
- Must submit laboratory screening/results or imaging documentation of ALL the following, **collected within the last 3 months**:
    - Negative human immunodeficiency virus 1 & 2 (HIV-1/HIV-2)
    - Negative Hepatitis B virus (HBV)

- Negative Hepatitis C virus (HCV)
- Negative Human T-lymphotropic virus 1 & 2 (HTLV-1/HTLV-2)
- Negative Cytomegalovirus (CMV)
- Negative Mycoplasma
- Negative pregnancy test is required in females of childbearing potential

- Must have documentation or an attestation from the prescriber on all the following:
  - Males capable of fathering a child and females of childbearing age should use an effective method of contraception from start of mobilization through at least 6 months after administration of LENMELDY
  - Must not have undergone allogeneic hematopoietic stem cell transplantation in the previous six months
  - Must not have myelodysplastic syndrome (MDS), acute myeloid leukemia (AML), or any other neoplastic disease
  - Must have not received Lenmeldy or any other gene therapy previously

Pre-Symptomatic Late Infantile (PSLI) MLD

- Must have disease onset ≤ 30 months of age
- Must have documentation that the member has an arylsulfatase A (ARSA) genotype consistent with presymptomatic late infantile MLD
- Must have documentation confirming absence of neurological signs and symptoms of MLD

Pre-Symptomatic Early Juvenile (PSEJ) MLD

- Must have disease onset was between > 30 months and < 7 years of age
- Must have documentation that the member has an arylsulfatase A (ARSA) genotype consistent with presymptomatic early juvenile MLD
- Must have documentation showing ONE of the following (which defines pre-symptomatic status in an early juvenile):
  - Absence of neurological signs and symptoms of MLD **OR**
  - Physical exam findings limited to abnormal reflexes and/or clonus

Early Symptomatic Early Juvenile (ESEJ) MLD

- Must have disease onset was between > 30 months and < 7 years of age
- Must have documentation that the member has an arylsulfatase A (ARSA) genotype consistent with early symptomatic early juvenile MLD
- Must have documentation of both of the following (which defines early symptomatic status):
  - Walking independently as defined as being at gross motor function classification for

metachromatic leukodystrophy [GMFC-MCLD] Level 0 (with or without ataxia) or GMFC-MCLD Level 1

- Member intelligence quotient (IQ) ≥ 85

APPROVAL DURATIONS

Initial Authorization	6 months
Reauthorization	N/A (Only single lifetime course allowed)

CODING AND STANDARDS

Codes

Code	Brand	Description
J3391	Lenmeldy	Injection, atidarsagene autotemcel, per treatment

Applicable Lines of Business

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

POLICY HISTORY

Date	Summary
September 2025	• New Guideline

LEGAL AND COMPLIANCE

Guideline Approval

Committee

Reviewed / Approved by Evolent Administrative Services Medical Policy Committee

Disclaimer

Evolent Clinical Guidelines do not constitute medical advice. Treating health care professionals are solely responsible for diagnosis, treatment, and medical advice. Evolent uses Clinical Guidelines in accordance with its contractual obligations to provide utilization management. Coverage for services varies for individual members according to the terms of their health care coverage or government program. Individual members' health care coverage may not utilize some Evolent Clinical Guidelines. Evolent clinical guidelines contain guidance that requires prior authorization and service limitations. A list of procedure codes, services or drugs may not be all inclusive and does not imply that a service or drug is a covered or non-covered service or drug. Evolent reserves the right to review and update this Clinical Guideline in its sole discretion. Notice of any changes shall be provided as required by applicable provider agreements and laws or regulations. Members should contact their Plan customer service representative for specific coverage information.

Evolent Clinical Guidelines are comprehensive and inclusive of various procedural applications for each service type. Our guidelines may be used to supplement Medicare criteria when such criteria is not fully established. When Medicare criteria is determined to not be fully established, we only reference the relevant portion of the *corresponding Evolent Clinical Guideline that is applicable to the specific service or item requested in order to determine medical necessity.*

REFERENCES

1. Lenmeldy (atidarsagene autotemcel) [prescribing information]. Boston, MA: Orchard Therapeutics North America; March 2024.
2. Bonkowsky, J. Metachromatic Leukodystrophy. In: Firth H. UpToDate. UpToDate; 2025. Accessed April 29, 2025. www.uptodate.com