

Hunter Syndrome Enzyme Replacement

Goal(s):

- Ensure medically appropriate use of enzyme replacement therapies that are FDA-approved for management of Hunter syndrome (also known as mucopolysaccharidosis [MPS] type II).

Length of Authorization:

- Up to 12 months

Requires PA:

- Elaprase® (idursulfase) and Avlayah™ (tvidenofusp alfa-eknm) for provider administered claims.

Covered Populations: FFS and CCO patients beginning 1/1/26 for idursulfase and 5/1/2026 for tvidenofusp alfa

Covered Alternatives:

- Current PMPDP preferred drug list per OAR 410-121-0030 at www.orpdl.org
- Searchable site for Oregon FFS Drug Class listed at www.orpdl.org/drugs/

Approval Criteria		
1. What diagnosis is being treated?	Record ICD10 code.	
2. Is this an FDA approved age and indication?	Yes: Go to #3	No: Pass to RPh. Deny; medical appropriateness
3. Is the request for continuation of therapy in a patient previously approved under fee-for-service (FFS) criteria?	Yes: Go to Renewal Criteria	No: Go to #4
4. Is the drug prescribed by or in consultation with a provider with expertise in managing lysosomal storage disorders (i.e., metabolic disorder specialist, endocrinologist, geneticist).	Yes: Go to #5	No: Pass to RPh. Deny; medical appropriateness
5. Is the patient currently on an enzyme replacement therapy for Hunter syndrome? Note: Use of dual enzyme replacement therapy in Hunter syndrome has not been evaluated.	Yes: Go to #6	No: Go to #7
6. Is the prescriber intending to switch from idursulfase to tvidenofusp alfa?	Yes: Go to #7	No: Pass to RPh. Deny; medical appropriateness

Approval Criteria

7. Has the diagnosis of Hunter syndrome (or MPS type II) been confirmed by either: - genetic testing OR - an enzyme assay showing deficient iduronate-2-sulfatase activity in leukocytes, fibroblasts, serum, or plasma?	Yes: Go to #8 Document date and test results_____	No: Pass to RPh. Deny; medical appropriateness
8. Is the request for idursulfase?	Yes: Go to #9	No: Go to #12
9. Is the patient aged 16 months or older? Note: The safety and efficacy of idursulfase in patients aged less than 16 months has not been established.	Yes: Go to #10	No: Pass to RPh. Deny; medical appropriateness
10. Is there documentation of one of the following symptoms of MPS type II: - Hepatosplenomegaly or - Skeletal deformities or - Neurocognitive deficits or - Cardiovascular disorders or - Impaired respiratory function?	Yes: Go to #11	No: Pass to RPh. Deny; medical appropriateness
11. Have one of the following tests to document baseline disease severity been documented: - Spleen volume or - Liver volume or - Forced vital capacity (FVC) or 6-minute walk test (6MWT)? - Echocardiogram - Sleep study	Yes: Pass to RPh. Pend; Refer to DMAP for secondary review. Approve for 1 year.	No: Pass to RPh. Deny; medical appropriateness
12. Is the request for tividenufusp alfa?	Yes: Go to #13	No: Pass to RPh. Deny; medical appropriateness
13. Is there documentation of neurologic manifestations consistent with Hunter syndrome or does the genotype predict a neurologic form of Hunter syndrome (e.g., cognitive impairment, developmental delay, seizures, or structural brain abnormalities on MRI)?	Yes: Go to #14	No: Pass to RPh. Deny; medical appropriateness
14. Has the provider documented goals of therapy for this treatment?	Yes: Go to #15	No: Pass to RPh. Deny; medical appropriateness

Approval Criteria

15. Have baseline hemoglobin, serum creatinine and urinary protein to creatinine levels been obtained?

Note: Due to risk of anemia, baseline hemoglobin should be obtained before starting therapy and reassessed 3 months afterwards. Because of the risk of membranous nephropathy, creatinine and urinary protein to creatinine ratio should be monitored if membranous nephropathy is suspected.

Yes: Pass to RPh. Pend; Refer to DMAP for secondary review.

Approve for 1 year.

No: Pass to RPh. Deny; medical appropriateness

Renewal Criteria

1. Is the request to renew idursulfase therapy?

Yes: Go to #2

No: Go to # 3

2. Has the patient's disease severity improved based on one of the following assessments

- Spleen volume or
- Liver volume or
- Forced vital capacity (FVC) or
- 6-minute walk test (6MWT)?

Yes: Pass to RPh. Pend; Refer to DMAP for secondary review.

Approve for 12 months

No: Pass to RPh. Deny; medical appropriateness

3. Is this request to renew tivenofusp alfa therapy?

Yes: Go to # 4

No: Pass to RPh. Deny; medical appropriateness

4. Have safety assessments (hemoglobin, serum creatinine, and urinary protein) been obtained?

Yes: Go to #5

No: Pass to RPh. Deny; medical appropriateness

5. Does provider attest that patient is responding to treatment (i.e., maintenance or improvement in neurologic manifestations consistent with Hunter syndrome) based upon goals outlined in initial request?

Yes: Pass to RPh. Pend; Refer to DMAP for secondary review.

Approve for 12 months

No: Pass to RPh. Deny; medical appropriateness