Louisiana Medicaid
Enzyme Replacement Therapy (ERT)
For Treatment of Mucopolysaccharidosis (MPS)

The *Louisiana Uniform Prescription Drug Prior Authorization Form* should be utilized to request clinical authorization for the following enzyme replacement therapy (ERT) agents used for the treatment of mucopolysaccharidosis (MPS).

*These agents have **Black Box Warnings**. Please refer to individual prescribing information for details.*

Approval criteria for specific agents:

Laronidase (Aldurazyme™)

- The recipient is 6 months of age or older on the date of the request; **AND**
- The recipient has a diagnosis of mucopolysaccharidosis I (MPS I) with one of the following phenotypes:
    - Hurler form (severe MPS I); **OR**
    - Hurler-Scheie form (attenuated MPS I); **OR**
    - Scheie form (attenuated MPS I) with moderate to severe symptoms; **AND**
- The diagnosis of MPS I is confirmed by both of the following [Include dates and results on request]:
    - Elevated urine glycosaminoglycans (GAGs); **AND**
    - Deficiency in the alpha-L-iduronidase enzyme; **AND**
- The medication is being prescribed by, or the requests states that the medication is being prescribed in consultation with, a provider experienced in the treatment of lysosomal storage diseases (LSDs), such as geneticist or endocrinologist; **AND**
- The recipient has at least **ONE** clinical manifestation known to respond to ERT (e.g., pulmonary, cardiac or musculoskeletal dysfunction); **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
    - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
    - The recipient has no concomitant drug therapies or disease states that limit the use of the requested agent; **AND**
    - The medication will not be used in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria for laronidase (Aldurazyme™)

- The recipient continues to meet all initial approval criteria; **AND**

- The recipient demonstrates a positive clinical response from baseline (i.e., improvement in respiratory function, cardiac function, walking ability or quality of life) and details of improvement are **stated on the request**.

Duration of initial approval: 6 months
Duration of reauthorization approval: 12 months

Idursulfase (Elaprase®)

- The recipient is 16 months of age or older on the date of the request; **AND**
- The recipient has a diagnosis of Hunter syndrome (Mucopolysaccharidosis II, MPS II) confirmed by both of the following [Include dates and results on request]:
    - Elevated urine glycosaminoglycans (GAGs); **AND**
    - Deficiency in the iduronate 2-sulfatase enzyme; **AND**
- The medication is being prescribed by or in consultation with a provider experienced in the treatment of lysosomal storage diseases (LSDs); **AND**
- The recipient has at least **ONE** clinical manifestation known to respond to ERT (e.g., pulmonary, cardiac or musculoskeletal dysfunction); **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
    - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
    - The recipient has no concomitant drug therapies or disease states that limit the use of the requested agent; **AND**
    - The medication will not be used in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria for idursulfase (Elaprase®)

- The recipient continues to meet all initial approval criteria; **AND**
- The recipient demonstrates a positive clinical response from baseline (i.e., improvement in respiratory function, cardiac function, walking ability or quality of life) and details of improvement are **stated on the request**.

Duration of initial approval: 6 months
Duration of reauthorization approval: 12 months

Elosulfase Alfa (Vimizim®)

- The recipient is 5 years of age or older on the date of the request; **AND**
- The recipient has a diagnosis of Morquio A syndrome (Mucopolysaccharidosis IVA, MPS IVA) confirmed by both of the following [Include dates and results on request]:

    - Elevated urine glycosaminoglycans (GAGs); **AND**
    - Deficiency in the N-acetyel-galactosamine-6-sulfate sulfatase (GALNS) enzyme; **AND**
- The medication is being prescribed by or in consultation with a provider experienced in the treatment of lysosomal storage diseases (LSDs); **AND**
- The recipient has at least **ONE** clinical manifestation known to respond to ERT (e.g., pulmonary, cardiac or musculoskeletal dysfunction); **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
    - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
    - The recipient has no concomitant drug therapies or disease states that limit the use of the requested agent; **AND**
    - The medication will not be used in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria for elosulfase alfa (Vimizim®)

- The recipient continues to meet all initial approval criteria; **AND**
- The recipient demonstrates a positive clinical response from baseline (i.e., improvement in respiratory function, cardiac function, walking ability or quality of life) and details of improvement are **stated on the request**.

Duration of initial approval: 6 months
Duration of reauthorization approval: 12 months

Galsulfase (Naglazyme®)

- The recipient is 5 years of age or older on the date of the request; **AND**
- The recipient has a diagnosis of Maroteaux-Lamy syndrome (Mucopolysaccharidosis VI, MPS VI) confirmed by both of the following [Include dates and results on request]:
    - Elevated urine glycosaminoglycans (GAGs); **AND**
    - Deficiency in the N-acetylgalactosamine 4-sulfatase (arylsulfatase B) enzyme; **AND**
- The medication is being prescribed by or in consultation with a provider experienced in the treatment of lysosomal storage diseases (LSDs); **AND**
- The recipient has at least **ONE** clinical manifestation known to respond to ERT (e.g., pulmonary, cardiac or musculoskeletal dysfunction); **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**

- All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
- The recipient has no concomitant drug therapies or disease states that limit the use of the requested agent; **AND**
- The medication will not be used in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria for galsulfase (Naglazyme®)

- The recipient continues to meet all initial approval criteria; **AND**
- The recipient demonstrates a positive clinical response from baseline (i.e., improvement in respiratory function, cardiac function, walking ability or quality of life) and details of improvement are **stated on the request**.

Duration of initial approval: 6 months
Duration of reauthorization approval: 12 months

Vestronidase Alfa-vjbk (Mepsevii™)

- The recipient is 5 months of age or older on the date of the request; **AND**
- The recipient has a diagnosis of Sly syndrome (Mucopolysaccharidosis VII, MPS VII) confirmed by both of the following [Include dates and results on request]:
  - Elevated urine glycosaminoglycans (GAGs); **AND**
  - Deficiency in the β-glucuronidase (GUS) enzyme; **AND**
- The medication is being prescribed by or in consultation with a provider experienced in the treatment of lysosomal storage diseases (LSDs); **AND**
- The recipient has at least **ONE** clinical manifestation known to respond to ERT (e.g., pulmonary, cardiac or musculoskeletal dysfunction); **AND**
- By submitting the authorization request, the prescriber attests to the following:
  - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
  - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
  - The recipient has no concomitant drug therapies or disease states that limit the use of the requested agent; **AND**
  - The medication will not be used in combination with any medication that is contraindicated or not recommended per FDA labeling.

Reauthorization Criteria for vestronidase alfa-vjbk (Mepsevii™)

- The recipient continues to meet all initial approval criteria; **AND**

- The recipient demonstrates a positive clinical response from baseline (i.e., improvement in respiratory function, cardiac function, walking ability or quality of life) and details of improvement are **stated on the request**.

Duration of initial approval: 6 months
Duration of reauthorization approval: 12 months

Additional edits may apply at Point-of-Sale (POS). Override options may be available. For more information, refer to the Louisiana Department of Health Pharmacy Benefits Management Services Manual at www.lamedicaid.com/provweb1/Providermanuals/manuals/PHARMACY/PHARMACY.pdf

References:

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