Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	X

Elaprase (idursulfase)

Override(s)	Approval Duration
Prior Authorization	1 year

Medications	Dosing Limit
Elaprase (idursulfase) 6 mg vial	0.5 mg/kg once per week

APPROVAL CRITERIA

Requests for Elaprase (idursulfase) may be approved if the following criteria are met:

I. Individual has a diagnosis of Mucopolysaccharidosis II (MPS II, Hunter syndrome) confirmed by (Scarpa 2011, Wraith 2008):
   A. Documented deficiency in iduronate 2-sulfatase enzyme activity as measured in fibroblasts or leukocytes combined with normal enzyme activity level of another sulfatase; **OR**
   B. Documented pathologic iduronate 2-sulfatase gene mutation;

AND

II. The individual has symptoms attributable to MPS II including (Muenzer 2012, Wraith 2008):
   A. Developmental delay or cognitive impairment; **OR**
   B. Frequent infections; **OR**
   C. Hearing loss; **OR**
   D. Hepatosplenomegaly; **OR**
   E. Hernias; **OR**
   F. Impaired respiratory function; **OR**
   G. Joint pain; **OR**
   H. Skeletal deformities; **OR**
   I. Sleep apnea; **OR**
   J. Valvular heart disease.

Elaprase (idursulfase) is may not be approved when the criteria above are not met and for all other indications.

Note:

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	X

Elaprase has a black box warning for risk of anaphylaxis. Life-threatening anaphylactic reactions have occurred and up to 24 hours after Elaprase infusions. Appropriate medical support should be available during Elaprase administration, and individuals should be educated on the signs and symptoms of anaphylaxis and to seek immediate medical care should they occur. Individuals with compromised respiratory function or acute respiratory disease may be at risk of serious acute exacerbation of their respiratory compromise due to hypersensitivity reactions, and require additional monitoring.

State Specific Mandates		
State name N/A	Date effective N/A	Mandate details (including specific bill if applicable) N/A

Key References:

1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: August 31, 2019.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
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4. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
5. Muenzer J, Bodamer O, Burton B, et al. The role of enzyme replacement therapy in severe Hunter syndrome-an expert panel consensus. *Eur J Pediatr*. 2012; 171(1):181-188.
6. Scarpa M, Almássy Z, Beck M, et al. Mucopolysaccharidosis type II: European recommendations for the diagnosis and multidisciplinary management of a rare disease. *Orphanet J Rare Dis*. 2011; 6:72.
7. Wang RY, Bodamer OA, Watson MS, Wilcox WR. American College of Medical Genetics (ACMG) Work Group on Diagnostic Confirmation of Lysosomal Storage Diseases. Lysosomal storage diseases: diagnostic confirmation and management of presymptomatic individuals. *Genet Med*. 2011; 13(5):457-484.
8. Wraith JE, Scarpa M, Beck M, et al. Mucopolysaccharidosis type II (Hunter syndrome): a clinical review and recommendations for treatment in the era of enzyme replacement therapy. *Eur J Pediatr*. 2008; 167(3):267-277.

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