Elaprase® (idursulfase)

When requesting Elaprase® (idursulfase), the individual requiring treatment must be diagnosed with the following FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved indication[1]

- Elaprase® (idursulfase) is indicated for the treatment of individuals with mucopolysaccharidosis type II (Hunter syndrome).

Approved Off-label Compendial use[2]

- No off-label uses meet evidence standards

Coverage Guidelines

Mucopolysaccharidosis Type II (Hunter syndrome)
Has a diagnosis of mucopolysaccharidosis type II (Hunter syndrome) confirmed by enzyme assay demonstrating a deficiency of iduronate sulfatase enzyme activity or by genetic testing[1,3]

Approval duration: 12 months

Additional Information for Prescribers[1]

Our prescribers are alerted to the following safety concerns as educational points/reminders:

- Life-threatening anaphylactic reactions, presenting as respiratory distress, hypoxia, hypotension, urticaria and/or angioedema of throat or tongue have occurred in patients during and up to 24 hours after Elaprase® (idursulfase) infusion. Closely observe individuals during and after Elaprase® (idursulfase) administration and be prepared to manage anaphylaxis.
- Late-emergent symptoms and signs of anaphylactic reactions have occurred up to 24 hours after initial treatment and recovery from an initial anaphylactic reaction. In addition, individuals experienced repeated anaphylaxis over a two- to four-month period, up to several years after initiating Elaprase® (idursulfase) treatment.
- Hunter syndrome individuals aged 7 years and younger with complete gene deletion, large gene rearrangements, nonsense, frameshift or splice site mutations experienced a higher incidence of hypersensitivity reactions, serious adverse reactions and anti-idursulfase antibody development.

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- Individuals with compromised respiratory function or acute febrile or respiratory illness may be at higher risk of life-threatening complications from hypersensitivity reactions. Careful consideration should be given to the individual's clinical status prior to administration of Elaprase® (idursulfase).
- The incidence of hypersensitivity reactions has been found to be higher in individuals who test positive for anti-idursulfase IgG antibodies than in those who test negative.

References

1. Elaprase® prescribing information. Lexington, MA: Shire Human Genetic Therapies, Inc.: Revised June 2013. http://pi.shirecontent.com/PI/PDFs/Elaprase_USA_ENG.pdf. Accessed February 14, 2017.
2. Klasco RK (Ed): DRUGDEX® system. Thomson Micromedex, Greenwood Village, CO. Accessed February 14, 2017. https://www.micromedexsolutions.com
3. Burton BK, Giugliani R. Diagnosing Hunter syndrome in pediatric practice: practical considerations and common pitfalls. *Eur J Pediatr*. 2012;171:631-639.Accessed February 14, 2017. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3306562/pdf/431_2012_Article_1703.pdf

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