BlueCross BlueShield
Federal Employee Program

CINRYZE
(C1 esterase inhibitor [human])

RATIONALE FOR INCLUSION IN PA PROGRAM

Background

Cinryze is a C1-esterase inhibitor used for routine prophylaxis against angioedema attacks in patients with hereditary angioedema (HAE). Hereditary angioedema is caused by having insufficient amounts of a plasma protein called C1-esterase inhibitor. People with HAE can develop rapid swelling of the hands, feet, limbs, face, intestinal tract, or airway. These acute attacks of swelling can occur spontaneously, or can be triggered by stress, surgery or infection. Swelling of the airway is potentially fatal without immediate treatment. Cinryze is intended to restore the level of functional C1-esterase inhibitor in a patient's plasma, thereby preventing the acute attack of swelling (1-4).

Regulatory Status

FDA-approved indication: Cinryze is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years of age and older) with Hereditary Angioedema (HAE) (2).

Hypersensitivity reactions may occur. Epinephrine should be immediately available to treat any acute severe hypersensitivity reactions following discontinuation of administration (2).

Thrombotic events have been reported at the recommended dose of C1 Esterase Inhibitor (human) products, including Cinryze, following treatment of HAE. Monitor closely patients with known risk factors for thrombotic events (2).

Cinryze is made from human plasma and may contain infectious agents, e.g., viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent (2).

Summary

Cinryze is a C1-esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adolescent and adult patients with Hereditary Angioedema (HAE). HAE symptoms include episodes of edema (swelling) in various body parts including the hands, feet, face, and airway. HAE is caused by mutations to C1-esterase-inhibitor (C1-INH). Serious arterial and venous thromboembolic (VTE) events have been reported at the recommended dose of plasma derived C1

esterase inhibitor products in patients with risk factors. The safety and efficacy of Cinryze in children less than 6 years of age has not been established. Persons who experience frequent and/or severe episodes may be candidates for prophylactic treatment (1-4).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Cinryze while maintaining optimal therapeutic outcomes.

References

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3. Zuraw BL, Banerji A, Bernstein JA, et al. US Hereditary Angioedema Association Medical Advisory Board 2013 recommendations for the management of hereditary angioedema due to C1 inhibitor deficiency. J Allergy Clin Immunol: In Practice. 2013; 1(5): 458-467.
4. Wintenberger C, Boccon-Gibod I, Launay D, et al. Tranexamic acid as maintenance treatment for non-histaminergic angioedema: analysis of efficacy and safety in 37 patients. Clin Exp Immunol 2014; 178:112.