

Hereditary Angioedema: Facilitating Diagnosis and Optimizing Clinical and Economic Outcomes

This supplement to *The American Journal of Managed Care* reviews the pathophysiology, symptoms and clinical course, and diagnostic challenges of hereditary angioedema (HAE). The current therapeutic landscape for HAE is discussed with a focus on treatment for acute angioedema attacks, short- and long-term HAE prophylaxis, and home-based therapy. The supplement also describes the economic burdens of HAE, both direct and indirect, and how changes in treatment practices, such as the wider adoption of home-based therapy, may improve patient outcomes and reduce costs.

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Type of activity: Knowledge

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Intended Audience

Medical directors, pharmacy directors, specialty pharmacists, and other managed care professionals who oversee the care of patients with hereditary angioedema.

Statement of Educational Need

Characterized by recurrent episodes of extravascular fluid accumulation and swelling of the extremities, abdomen, or larynx, hereditary angioedema (HAE) is a rare autosomal dominant disorder resulting from C1 inhibitor deficiency. While laryngeal involvement is less common, findings indicate that over 50% of affected patients experience at least 1 episode during their lifetime, potentially resulting in asphyxiation and death. HAE is a chronic disease that typically presents early in life. However, because the condition is uncommon and symptoms closely resemble other ailments (eg, allergic reactions, gastrointestinal tract obstructions), diagnostic delays may exceed 10 years.

Appropriate disease management may minimize morbidity and mortality, reduce emergency department visits, improve quality of life, and enable patients to work. Management generally encompasses treatment of acute episodes and prophylactic therapy to prevent exacerbations and reduce the frequency and severity of recurrent attacks. One major managed care issue related to treatment of HAE is third-party reimbursement for the newer, disease-specific therapies. Many of these newer agents have not yet been added to formularies, and as such, claims are automatically rejected, or are associated with high copays. As a result, patients may be treated with older, less expensive therapies (eg, androgens), and beyond their associated adverse effects, these agents are not optimally effective in preventing breakthrough attacks. To provide greater access to newer therapies, it is vital to develop an infrastructure that provides patients with copay assistance.

This activity provides an overview of the clinical presentation and diagnostic criteria of HAE, and examines the managed care implications of HAE, including direct and indirect costs, resource utilization, and reimbursement issues. The discussion also centers on conventionally used versus recently approved medications, and addresses selection of candidates who qualify for Cinryze therapy.

Educational Objectives

Upon completion of the educational activity, the participants should be able to:

- Review the clinical presentation of HAE and review diagnostic criteria
- Compare and contrast conventionally used versus recently approved medications, and discuss the safety, efficacy, and place in therapy of each
- Select candidates who qualify for Cinryze therapy and discuss appropriate training techniques
- Examine managed care implications of HAE, including direct and indirect costs, resource utilization, and reimbursement issues

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