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POSTER #14

Hereditary angioedema C1-inhibitor replacement therapy and coexisting autoimmune disorders: findings from a claims database

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ABSTRACT

Background: Autoimmune diseases are a leading cause of morbidity and mortality in the US (estimated prevalence: 4.5%) and often associated with dysregulation of the complement system (innate and adaptive immune response). The classic complement pathway is regulated by the C1-inhibitor (C1-INH), which binds to C1 to prevent its activation. Hereditary angioedema with C1-INH deficiency (C1-INH-HAE) may be linked to increased autoimmunity due to secondary deficiency of C1r, C1s, and other components.

Aims: It was hypothesized that increased regulation of the complement system via C1-INH replacement therapy may reduce autoimmunity in patients with C1-INH-HAE. The coexisting autoimmune disease claims frequency was compared between C1-INH-HAE patients treated with plasma-derived (pd) C1-INH vs "other (non-C1-INH)" treatments.

Methods: C1-INH-HAE patients were identified in the IMS Health PharMetrics Plus claims database between January 2012 and December 2015 by International Classification of Diseases 9/10 diagnosis code, and classified based on the use of pdC1-INH or "other (non-C1-INH)" treatments for HAE. Index date was the first claim for HAE treatment. For patients using pdC1-INH, the first fill was the index date, even if other HAE medications were used previously. Frequency of visit claims for autoimmune conditions was identified by diagnostic codes (primary or secondary). Mean visits per patient per year by treatment group, gender, and age ($<$50 vs $\geq$50 years) were summarized for autoimmune conditions.

Results: Of 589 patients with HAE identified (69% female, 38% aged $\geq$50 years), 276 (729 patient-years) received pdC1-INH and 313 (860 patient-years) received "other (non-C1-INH)" treatments. In this cohort, 12.9% of patients had $\geq$1 visit associated with a coexisting autoimmune disorder – the most common were lupus, alopecia, rheumatoid arthritis, sicca (Sjogren) syndrome, and connective tissue disorders. The mean (95% CI) number of visits for autoimmune diagnoses per patient per year was numerically lower for patients treated with pdC1-INH compared to those receiving "other (non-C1-INH)" treatments (1.37 [0.56–2.19] vs 2.28 [0.83–3.73]).

Conclusions: Based on these findings, it is concluded that treatment of C1-INH-HAE with pdC1-INH may have a positive impact on coexisting autoimmune conditions by normalizing complement. Further research is needed on this important issue. There may be implications for healthcare resource utilization among patients with HAE and coexisting autoimmune disorders.

KEYWORDS
Angioedema; C1-inhibitor; autoimmune; complement

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