

## **Quality of life and measures of well being of Canadian patients with HAE based on data from the 2020 national survey**

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**Introduction:** Hereditary angioedema (HAE) is a genetic disorder characterized by severe, acute skin and mucosal episodes of angioedema due to bradykinin-induced increases in vascular permeability. Most patients have deficient or dysfunctional C1 inhibitor (HAE C1INH), but a significant percentage have other mutations causing similar episodes of angioedema with normal C1INH (HAEnC1INH).

**Methods:** Data was acquired from an online survey sent to all members of HAE Canada who self-reported HAE C1INH or Acquired Angioedema (AAE) and HAEnC1INH. Responses related to burden of illness were collated and expressed as a percent of respondents.

**Results:** Forty-five respondents reported having HAEnC1INH, 106 reported having HAE C1INH and 4 AAE. HAE C1INH and AAE were combined for analysis. Age of onset (mean, range) was higher for HAEnC1INH (25, 1-58) years than for HAE C1INH (18, 0.25-77) years. In the prior year, those with HAEnC1INH were more likely have had attacks (90% vs 78%) and more likely to have >12 attacks per year (50% vs 27%) than those with HAE C1INH. More HAEnC1INH patients reported a high impact on personal (54% vs 26%) and financial (50% vs 17%) well-being and ability to work full-time (50% vs 20%) than those with HAE C1INH. Overall, those with HAEnC1INH had more concerns; specifically, a higher percentage were worried about interference with relationships (40% vs 21%) and social isolation (38% vs 11%). Depression was common (46% vs 41%) for both but more of those with HAEnC1INH felt powerless (72 vs 45%) with a lack of control of their disease (72 vs 42%).

**Discussion:** Although genotyping is available for some types of HAEnC1INH, there are no biomarkers available to measure disease activity in HAEnC1INH. Diagnosis is difficult and access to treatment is limited since there are no drugs approved for HAEnC1INH. Compared to the combined responses of HAE C1INH and AAE patients surveyed, HAEnC1INH patients suffered more frequent attacks with a significant impact on their quality of life and measures of wellbeing.

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