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 from 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 angioedema episodes with normal C1INH (HAEnC1INH). Some patients have acquired angioedema due to either autoimmune or lymphoproliferative disorders which affects C1INH.

Methods

Data was acquired from an online survey sent to all members of HAE Canada in 2020. Analysis was segregated by 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

	HAE C1INH (Type I/II) (n=106)	HAEnC1INH (n=45)	Acquired (n=4)
Age (years) mean (range)	52 (22-90)	49 (23-83)	55 (35-72)
Gender n (%)			
Female	81 (76%)	38 (84%)	2 (50%)
Male	24 (23%)	7 (16%)	2 (50%)
Medication to treat HAE n (%)			
Yes	94 (89%)	39 (87%)	
No	12 (11%)	6 (13%)	
Age of Onset; mean (range)	18 (0.25 -77)	25 (1-58)	25 (6-40)
Age of Diagnosis; mean (range)	32 (1-84)	39 (16-68)	42 (26-59)

Table 1.
Demographics and baseline characteristics. Most survey respondents had HAE C1INH (Type I/II). Only 4 had Acquired HAE. A majority of Type I/II HAE and HAEnC1INH patients were female and taking medication to treat HAE. For HAEnC1INH and Acquired HAE, onset and diagnosis were at an older age. Since few patients reported Acquired HAE, going forward they were combined with Type I/II HAE.

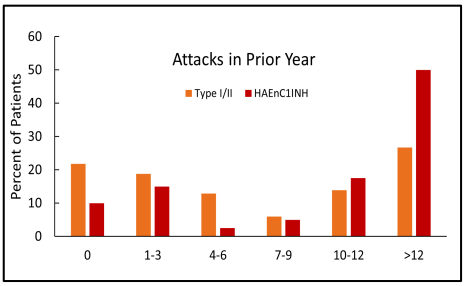

Figure 1. Despite a comparable proportion of respondents using HAE treatments; a higher percentage of those with HAEnC1INH had frequent attacks and only 10% had no attacks in the prior year.

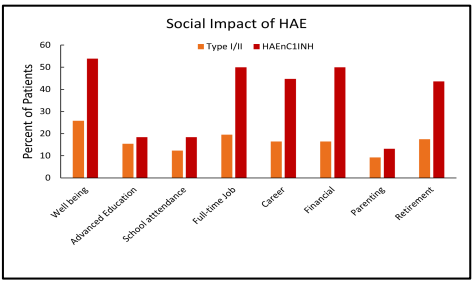

Figure 2. The social impact of HAE was greater for HAEnC1INH patients. A greater percentage reported a high impact on well being (54% vs 26%); ability to work full-time (50% vs 20%) or advance in their career (45% vs 17%) with the result that finances (50% vs 17%) and retirement (44% vs 18%) were also more affected.

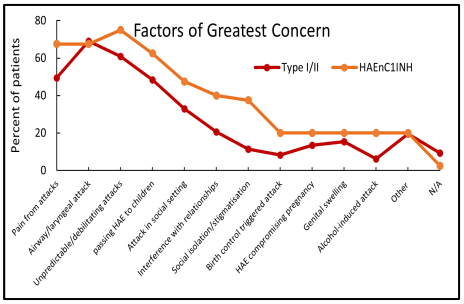

Figure 3. Respondents were asked to select all factors of greatest concern from a prepared list. For most factors, a greater proportion of HAEnC1INH patients expressed concern. The main exception was airway/laryngeal attacks which were of concern to nearly 70% of all patients.

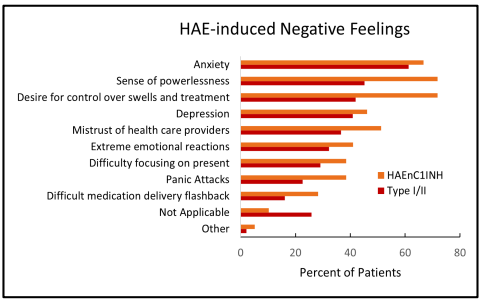

Figure 4. Anxiety (67% vs 61%) and depression (46% vs 41%) affected similar proportions of Type I/II and HAEnC1INH patients. However, significantly more of those with HAEnC1INH felt powerless (72% vs 45%) and with a lack of control of their disease (72% vs 42%). Overall, negative feelings affected a higher percentage of HAEnC1INH patients.

Conclusions

- Although genotyping is available for some types of HAEnC1NH, there are no biomarkers available to measure disease activity in HAEnC1INH.
- Diagnosis can be difficult and access to treatment can be 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 social functioning, quality of life and measures of wellbeing.

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