Real World Data of Canadian Adults Living with Hereditary Angioedema: Part 1 - Demographics

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Abstract

Background
Hereditary angioedema (HAE) is an unpredictable and serious genetic disorder affecting approximately 1:10,000 to 1:50,000. It is an autosomal dominant disorder due to C1 inhibitor deficiency. Clinically, it is manifested by painful, unpredictable edema of the face, larynx, abdomen, genitals and extremities. It can be debilitating and if left untreated, may be fatal. We sought to better understand the demographic profiles of patients living with HAE in Canada.

Methods
In 2017, a comprehensive survey was sent out to all HAE Canada members by email to gather information on HAE in Canada. Data from respondents have been collected and analyzed using percentage of total surveys to provide data on demographics of these patients.

Results
The demographic location of HAE patients living in Canada includes Ontario, Alberta, Manitoba, British Columbia, Nova Scotia, Quebec, Saskatchewan and Newfoundland and Labrador. 140respondents indicated their relationship to HAE as; 81% are adults living with HAE, 10% are caregivers of an adult living with HAE who lives with them, 2% are caregivers of an adult living with HAE who does not live with them, 2% are adults awaiting a diagnosis, and 4% are other or unknown. 109 respondents indicated 79% are female and 21% are male. When respondents were asked about their HAE type, 60% were found to have type 1/2 C1-inhibitor protein deficiency, 26% have HAE with normal C1-inhibitor, 10% unsure, and 4% have acquired angioedema.

Conclusions
This survey helps to better understand the current demographic profile of patients living with HAE and is the first national HAE survey done in Canada. However, data interpretation is limited due to uncertainty of necessary sample size required to be representative of the true population. Overall, our results demonstrate that HAE patients can be found across Canada and that the majority of patients in this survey are aware of their diagnosis.

Introduction

- HAE is a rare autosomal dominant disorder involving deficiencies in C1 inhibitor antigenic protein (C1-INH).
- It is clinically manifested by painful and unpredictable swelling that occurs on various parts of the body and is estimated to affect between 1:10,000 and 1:50,000.
- Some attacks may be life-threatening such as edema of the larynx, which may block an individual's ability to breathe.
- HAE attacks differ from other conditions in that they produce recurrent swelling without the presence of urticaria (hives)[1].
- The most commonly diagnosed is Type 1 HAE and Type 2 HAE, with Type 1 HAE involving low serum C1-INH levels and Type 2 HAE involving normal levels of C1-INH that are dysfunctional[1,2].
- HAE with normal C1-INH, formerly know as Type 3, is clinically similar to the others, however, remains partially unexplained due to normal levels and functioning of C1-INH[1,2].
- Indications for testing patients include positive family history or any clinical suspicion[1].
- There are various triggers that cause episodes of HAE, including stress, anxiety, surgeries, injuries and certain types of medications[2].
- The attacks tend to develop over a period of 24 hours and then begin to diminish over a period of 24 to 72 hours, however, some may last even longer.
- Management of attacks includes either "on-demand treatment", short-term prophylaxis or long-prophylaxis, depending on the severity and the availability of treatment options[1].
- HAE Canada has set out to better understand the demographic profiles of patients and their caregivers in order to help optimize health and well-being.
- Factors such as availability of specialists and availability of treatment options may determine this.

Methods

Starting in January 2017, all members of HAE Canada were emailed an extensive survey to gather data on patients living with HAE and their caretakers. This was the first HAE national survey done in Canada, being referred to as the "Report Card on HAE in Canada". Data was gathered and analyzed in October 2017 by using the percentage of total number of surveys answered for each question.

Results

The Demographic Location Of HAE Patients Living In Canada

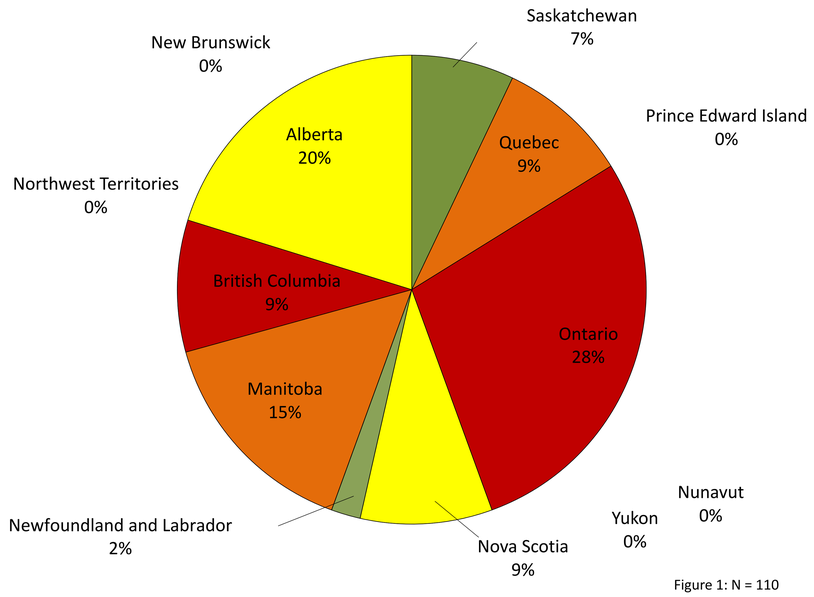

Figure 1: N = 110

Participant Response When Asked About Relationship To HAE In Canada

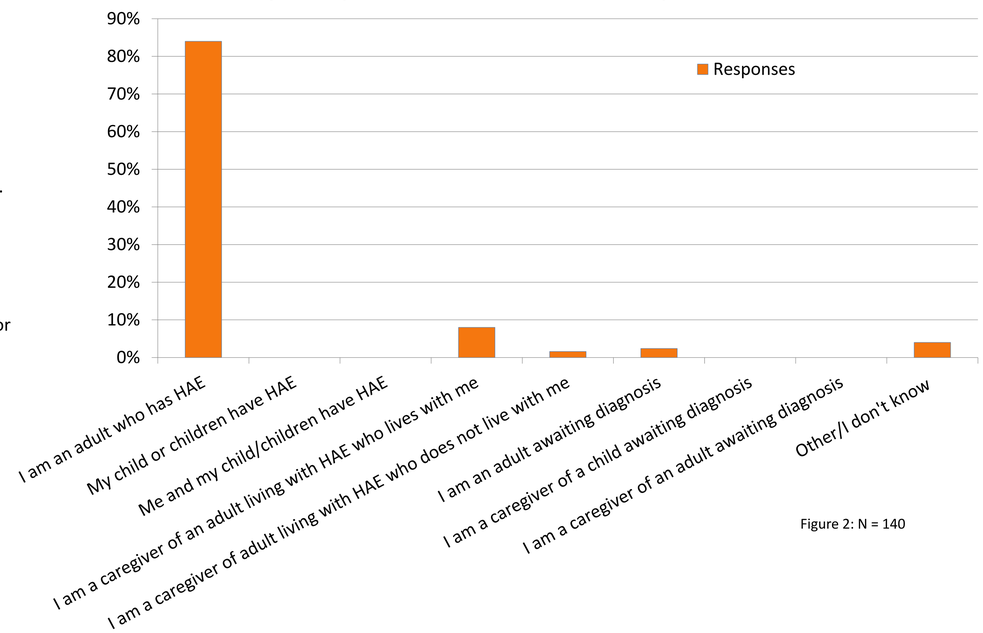

Figure 2: N = 140

Gender Differences Of HAE Patients In Canada

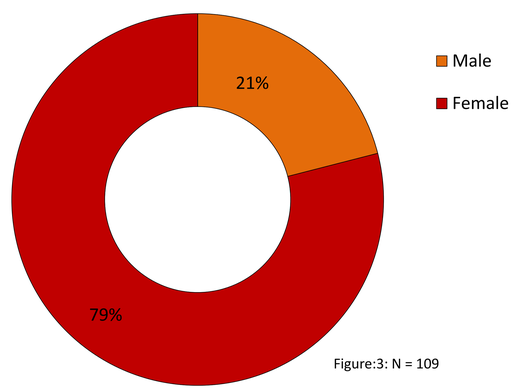

Figure:3: N = 109

Distribution Of HAE Type For Patients In Canada

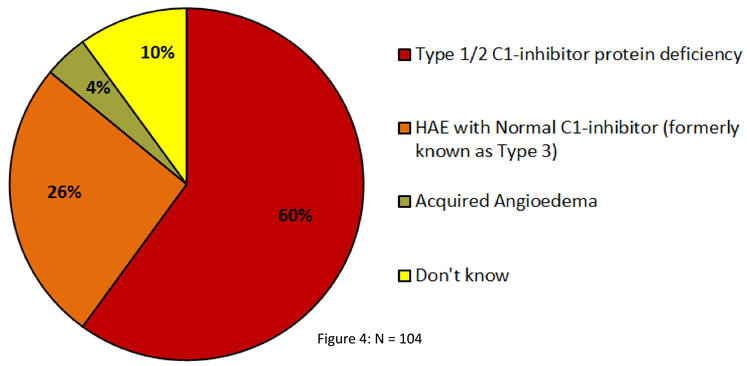

Figure 4: N = 104

Conclusion

The demographic profiles of both patients living with HAE and their caregivers have been gathered in order to better understand the distributions in Canada. This was the first national HAE survey done in Canada. Thus, data interpretation remains limited due to small sample sizes of 100-140 respondents per question.
Future studies will look to better establish the true HAE patient population by gathering a larger number of respondents.

References

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