Real World Data of Canadian Adults Living with Hereditary Angioedema: Part 2 - Attack Profile

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Abstract

Hereditary Angioedema (HAE) is a rare genetic disorder that is characterized by episodes of unpredictable painful swelling in different body parts involving the face, larynx, peripheral limbs, abdomen and genitals. In Canada, there are approximately 400-600 HAE subjects. To better understand the challenges of Canadians living with HAE we conducted the first web survey among our HAE Canada members, the objective was to gather real world data that will provide insight into the attack profiles of a HAE patient.

Method

In 2017, data was collected through voluntary online surveys of children, youth, and adults who live with HAE and their caregivers in Canada. The following data was based solely on adult participants.

Results

Among 104 participants with HAE they reported a diagnosis of: Type 1 or 2 C1-inhibitor protein deficiency (60%), HAE with normal C1-inhibitor (26%), acquired angioedema (4%), and unsure of diagnosis (10%). In the last year, 78% were symptomatic, 11% were asymptomatic, and 11% were unsure. Regarding the frequency of attacks, 61% had 7 or more attacks, 22% had 1-6 attacks, 6% had no attacks, and 10% were unsure. Identifiable attack triggers vary from stress (87%), typing/writing (78%), trauma (70%), illness (61%), medical procedures (61%), anxiety (55%), and Ace inhibitors (6%). Other factors that increase HAE frequency of attacks: 61% had 7 or more attacks, 22% had 1-6 attacks, 6% had no attacks, and 10% were unsure. Regarding the cause of attacks, 84% use an agent, compared to 16% who do not. The most common treatment agent used was C1-esterase inhibitor (Berinert IV).

Conclusion

Our findings demonstrate the majority of participants are knowledgeable in identifying their triggers and managing their attacks. Results show improvements are necessary for proper diagnosis and awareness of the disease. Since the number of people living with HAE is estimated, our data is limited to the respondents and may not represent the broader Canadian HAE population.

Introduction

Hereditary Angioedema (HAE) is a rare genetic disorder that is characterized by spontaneous and recurrent episodes of swelling of the skin in various parts of the body, as well as in the airways, and internal organs.

HAE is classified into 3 types: type I and type II are caused by a defect in the gene responsible for producing the protein C1 esterase inhibitor (C1-INH), while type III is caused by a defect in the gene responsible for producing the C1 INH-related protein (C1R).

Management of HAE can be divided into two approaches: prophylaxis and treatment of attacks.

Methods

In October 2017, the first National Canadian HAE survey was electronically distributed to all HAE Canada members for the purpose of gaining better insight on: HAE type, symptoms, frequency of attacks, identifiable triggers, and attack treatment.

Results

Type Of HAE

- Type 1 or 2 C1-inhibitor protein deficiency: 60%
- HAE with normal C1-inhibitor: 26%
- Acquired angioedema: 4%
- Unsure of diagnosis: 10%

Number Of Attacks In The Last Year

- 0: 6%
- 1-3: 20%
- 4-6: 20%
- 7-9: 10%
- 10-12: 10%
- 12 or more: 41%

Symptoms In The Last Year

- Symptomatic: 78%
- Asymptomatic: 11%
- Don’t know: 11%

Agents Used To Stop Attacks

- Icatibant: 47%
- Cinryze: 28%
- Berinert: 25%
- No impact on HAE attacks: 28%

Impact On Attacks

- No impact: 79%
- Increase HAE attacks: 20%
- Decrease HAE attacks: 1%

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