TP0728 | Hereditary angioedema in a real clinical practice

Viktorovich Emelyanov A; Sergeeva G; Leshenkova E

North-Western State Medical University named after I.I.Mechnikov, Saint-Petersburg, Russia

Background: Hereditary angioedema (HAE) is a rare disorder with recurrent attacks of severe swelling commonly affecting the extremities, face, intestinal tract, and airways. Little is known about patients with HAE in Russia. The aim of this study was to assess clinical characteristics of patients with HAE in real clinical practice.
Method: We examined 18 adult outpatients (55% males) aged 18-59 yrs (mean age 37 yrs) referred to our secondary care center by general practitioners. C1-esterase inhibitor quantity and activity in blood were evaluated by immunoenzyme analysis.
Results: All 18 patients had HAE type 1 with mean C1-inhibitor concentration and functional activity 31% and 30% of normal respectively. Eighty nine percent of patients had a positive family history. First symptoms of HAE appeared at age 1-26 yrs (mean age 11 yrs, 50% before 10 yrs, 78% before 20 yrs). HAE was diagnosed in 1-54 yrs (mean 23 yrs) after onset of the disease. Triggers of swelling were trauma - 78% and physical activity - 67%, stress — 22%, injections and insect stings — 11%, flu - 6%, dental manipulations - 6%, overnutrition — 6%. Frequency of HAE attacks varied 1-20 each year per patient (mean 7 per year/patient). Locations of swelling were extremities — 63%, face -50%, intestinal tract — 72%, larynx - 67%. One patient underwent tracheotomy for 6 times, other patient — for 2 times. One of the patients has 2 laparotomies due to HAE attacks during lifetime. Maintenance therapy in most patients included tranexamic acid, 2 patients were treated by danazol for many years. Icatibant was used in 14 patients and C1-concentrate — in 1 patient with acute attacks of HAE.
Conclusion: HAE may be a debilitated disorder. There is a significant delay (more than 20 yrs) between the onset and diagnosis of HAE. Most common therapy was tranexamic acid for maintenance and icatibant for acute attacks of swelling.

TP0729 | Real world data of Canadians living with hereditary Angioedema: Part 4- Treatment satisfaction

Brosz K[1]; Baidou J[1]; Howlett L[1]; Rowe A[1]; Steele K[1]; Falbo J[2]; Santucci S[2]; Valois J[2]; Yang WH[2,3]

[1]HAE Canada, Ottawa, Canada; [2]Ottawa Allergy Research Corporation, Ottawa, Canada; [3]University Of Ottawa Medical School, Ottawa, Canada

Background: Hereditary angioedema (HAE) is a chronic spontaneous life-threatening disease. Due to the unpredictable nature associated with the disease it can have a significant impact on a patient's quality of life. We sought to better understand the overall satisfaction of treatments from a patient's perspective.
Method: In 2017-2018, data were collected through voluntary online surveys of children, youth, and adults who live with HAE and their caregivers in Canada. The following data were based solely on adult participants.
Results: Once a proper diagnosis was obtained following patient navigation and treatments were established the annual number of days missed from work or school decreased by an average of 48%, the amount of phone calls to doctor's offices decreased 60%, the occurrence of unscheduled visits to health care professionals decreased 75%, the frequency of emergency room visits decreased 50%, and the number of hospitalizations decreased 67%. Most patients reported they were satisfied with the frequency they must use their HAE treatments (31%) and satisfied with the effectiveness of their current treatments to prevent attacks (40%). Overall patients were satisfied (39%) and very satisfied (24%) with their current HAE treatment.
Conclusion: Results show patients are generally satisfied with the treatments they are currently receiving; however, there are still necessary improvements that can enhance a patient's quality of life. All results are limited to the respondents and may not represent the broader Canadian HAE population.

TP0730 | Real world data of Canadian's living with hereditary angioedema: Part 1- demographics

Badiou J[1]; Howlett L[1]; Rowe A[1]; Steele K[1]; Falbo J[2]; Santucci S[2]; Valois J[2]; Yang WH[2,3]

[1]HAE Canada, Ottawa, Canada; [2]Ottawa Allergy Research Corporation, Ottawa, Canada; [3]University of Ottawa Medical School, Ottawa, Canada

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.
Method: In 2017-2018, the first National Canadian HAE survey was electronically sent to all HAE Canada members. Data from respondents was collected and analyzed using percentage of total surveys.
Results: The demographic location of HAE patients living in Canada includes Ontario, Alberta, Manitoba, British Columbia, Nova Scotia, Quebec, Saskatchewan and Newfoundland and Labrador. 140 respondents 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. Among, 109 respondents 79% indicated they are female and 21% are male. When 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.