

HAE attacks in Canadian patients with HAE: Triggers and treatment based on data from the 2020 national survey

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Introduction: Hereditary angioedema (HAE) is a genetic disorder leading to unpredictable, and painful episodes of angioedema due to bradykinin-induced increases in vascular permeability. Most patients have deficient or dysfunctional C1 inhibitor (HAEC1INH) (Type I/II) but a significant percentage have normal C1INH (HAEnC1INH) with angioedema.

Methods: Data from an online survey sent to all members of HAE Canada was sorted by self-reported HAEC1INH and HAEnC1INH. Responses related to attacks, their treatments and their triggers were collated and expressed as a percent of respondents.

Results: HAEC1INH was reported by 106 respondents and HAEnC1INH by 45. Reported age (mean, range) was similar: HAEC1INH (52, 22-90); HAEnC1INH (49, 23-83). In the prior year, those with HAEC1INH were more likely to be attack free (22% vs 10%) and less likely to have >12 attacks per year (27% vs 50%) than those with HAEnC1INH. They were also less likely to have laryngeal attacks (HAEC1INH: 24%; HAEnC1INH 43%). Most reported that some or all of their attacks had an identifiable trigger (HAEC1INH 82%; HAEnC1INH 78%) with stress (HAEC1INH 77%; HAEnC1INH 74%) and anxiety (HAEC1INH 51%; HAEnC1INH 60%) being the most common. Women further identified menstruation (HAEC1INH 42%; HAEnC1INH 53%) and estrogen contraceptives (HAEC1INH 18%; HAEnC1INH 67%) as triggers. The most common treatments for attacks were plasma-derived (PD) C1INH (HAEC1INH 58%; HAEnC1INH 39%) and icatibant (HAEC1INH 23%; HAEnC1INH 30%). One treatment resolved the attack for HAEC1INH 55%; HAEnC1INH 50%) and 2 treatments for 18% and 28%, respectively.

Discussion: Our data confirms that patients report stress is an important trigger for HAE attacks and demonstrates that for women, menstruation and contraception used in Canada are additional attack triggers. Despite treatment, patients with HAEnC1INH have more frequent attacks. A better understanding of the underlying mechanisms leading to angioedema and more targeted treatments for patients with HAEnC1INH may reduce attacks and improve their quality of life.