HAE with normal C1 esterase inhibitor (HAEnC1INH): Diagnosis, treatment and attack frequency from the 2020 Canadian patient survey

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Rationale: There are no diagnostic tests or approved medications for hereditary angioedema with normal C1INH (HAEnC1INH). As a result, diagnosis is difficult and often delayed. The treatments used are the same as for those with HAE with inadequate C1INH.

Methods: An online survey was sent to Canadian HAE patients in 2020 to better understand their health burden and treatments used. We extracted responses from patients who self-reported having HAEnC1INH and analyzed the data to better understand diagnosis, attack frequency and medication use.

Results: HAEnC1INH was self-reported by 45/167 participants (27%). They were mainly female (84%) and 54 (23-91) [mean (range)] years of age. There was a 14-year difference between the age of symptom onset at 26 (1-58) and of diagnosis at 39 (16-68) years of age. In the prior year, 50% of respondents reported having >12 attacks and 43% reported having a laryngeal attack despite 87% of these patients reporting using either prophylactic or on demand medication to treat their HAEnC1INH. Most respondents used pdC1INH to treat acute attacks (IV 30%, SQ 11%) and for prophylaxis IV33%, SQ 24%). Icatibant was used by 30% to treat acute attacks. Access to treatment was provided by a government plan (74%) or by private insurance (21%).

Discussion: There is still a long delay between the onset of symptoms and diagnosis. While most participants have access to treatment, most still have frequent attacks. There is a need for more awareness as well as targeted treatment for those with HAEnC1INH.

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