

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

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Objective

- To use recent survey data to compare attacks, triggers and treatment of HAE-C1INH and HAEnC1INH patients

Conclusions

- Patients with HAEnC1INH have more frequent attacks than those with HAE-C1INH. All HAE patients treat attacks mainly with plasma-derived (pd) C1-INH and icatibant.
- Stress is an important trigger for HAE attacks. For women, menstruation and contraception are additional attack triggers.
- More targeted treatments for patients with HAEnC1INH may reduce attacks and improve their quality of life.

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 (HAE-C1INH) (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 HAE-C1INH and HAEnC1INH. Responses related to attacks, their treatments and their triggers were collated and expressed as a percent of respondents.

Results

HAE-C1INH was reported by 106 respondents and HAEnC1INH by 45. Reported age (mean, range) was similar: HAE-C1INH (52, 22-90); HAEnC1INH (49, 23-83). A higher proportion of HAEnC1INH patients were female (87 vs 79%).

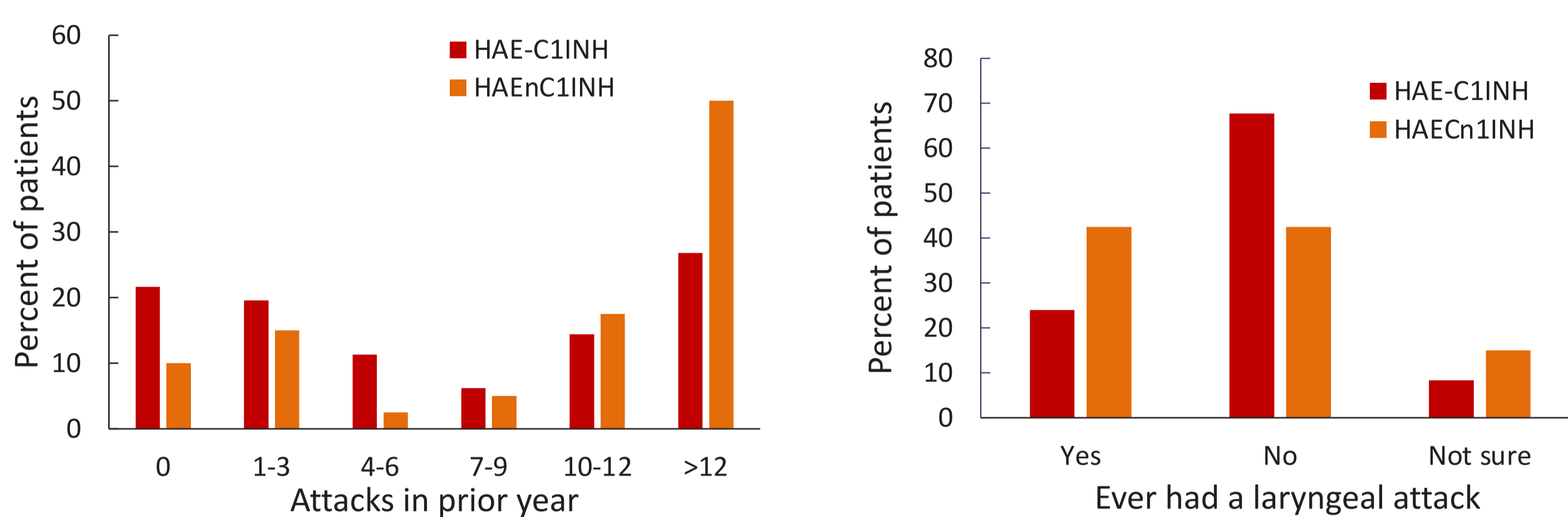


Figure 1. In the prior year, those with HAE-C1INH were more likely to be attack free (22% vs 10%) and less likely to have >12 attacks per year (27% vs 50%) than those with HAEnC1INH. They were also less likely to have laryngeal attacks (HAE-C1INH: 24%; HAEnC1INH 43%).

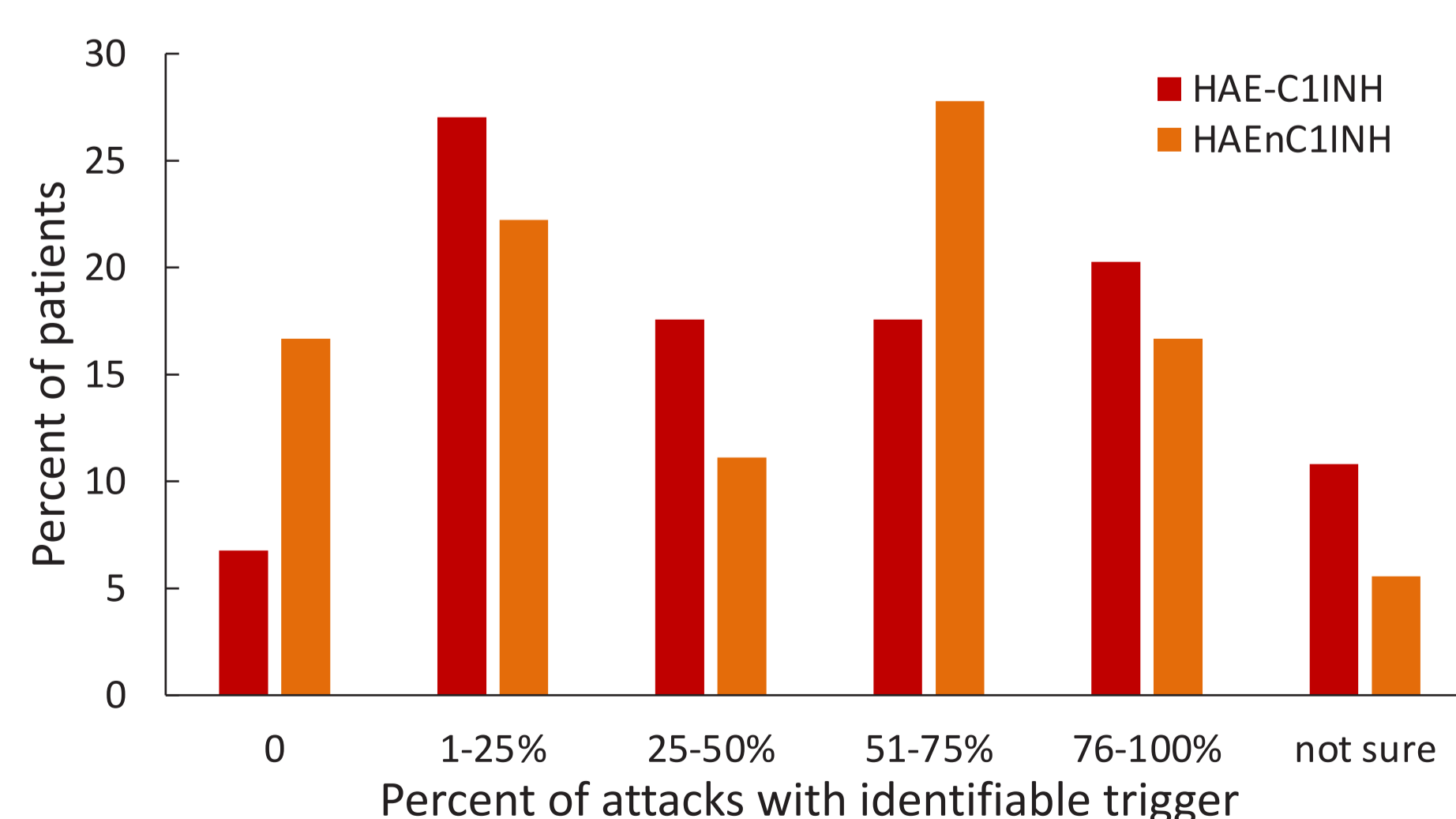


Figure 2 Most patients reported that some or all of their attacks had an identifiable trigger (HAE-C1INH 82%; HAEnC1INH 78%). A higher proportion of patients with HAE-C1INH had no identifiable triggers (HAE-C1INH 17%; HAEnC1INH 7%).

Results (continued)

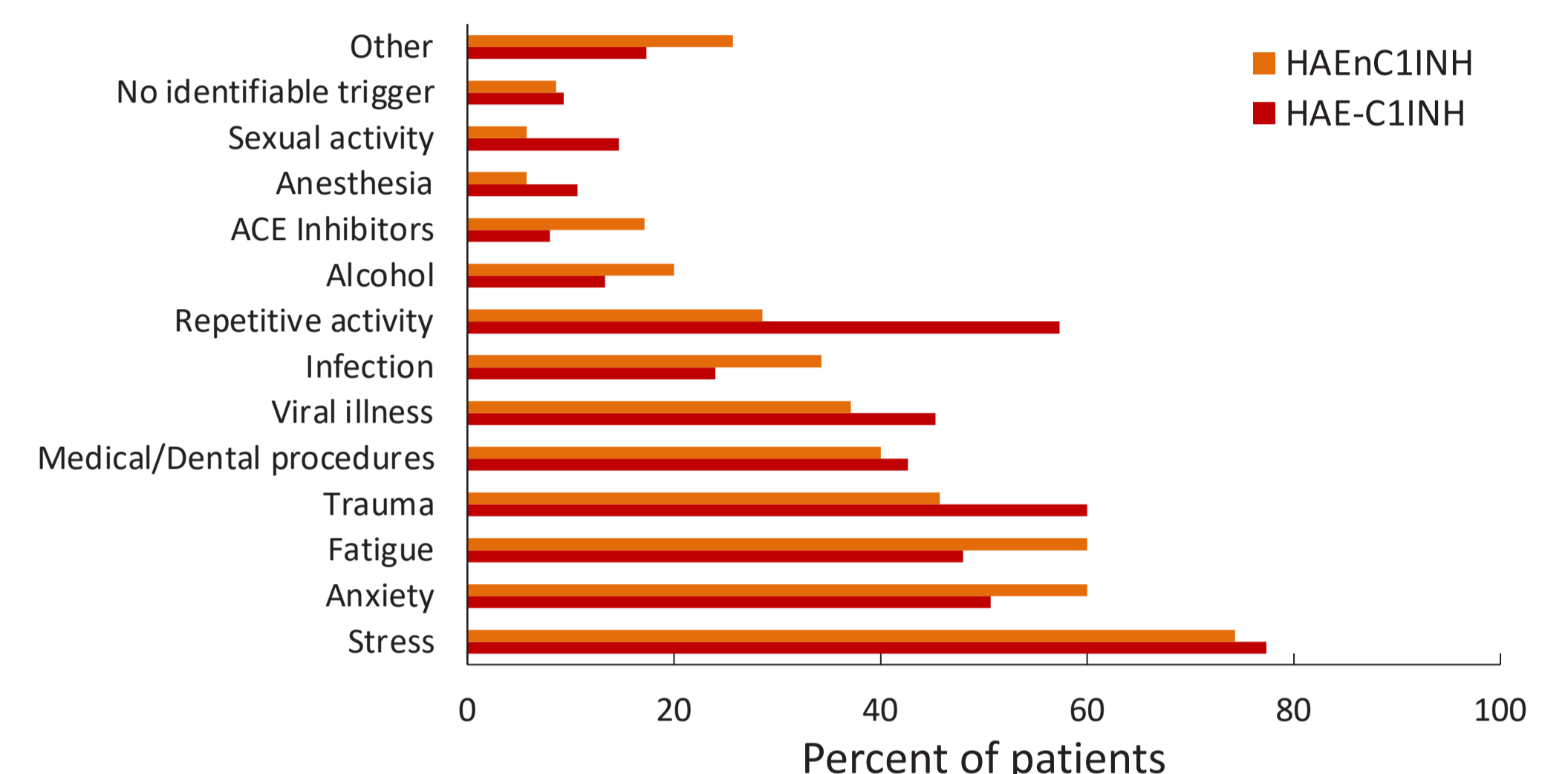


Figure 3. Stress (HAE-C1INH 77%; HAEnC1INH 74%) and anxiety (HAE-C1INH 51%; HAEnC1INH 60%) were the most common triggers. Repetitive activity affected more patients with HAE-C1INH (HAE-C1INH 57%; HAEnC1INH 29%).

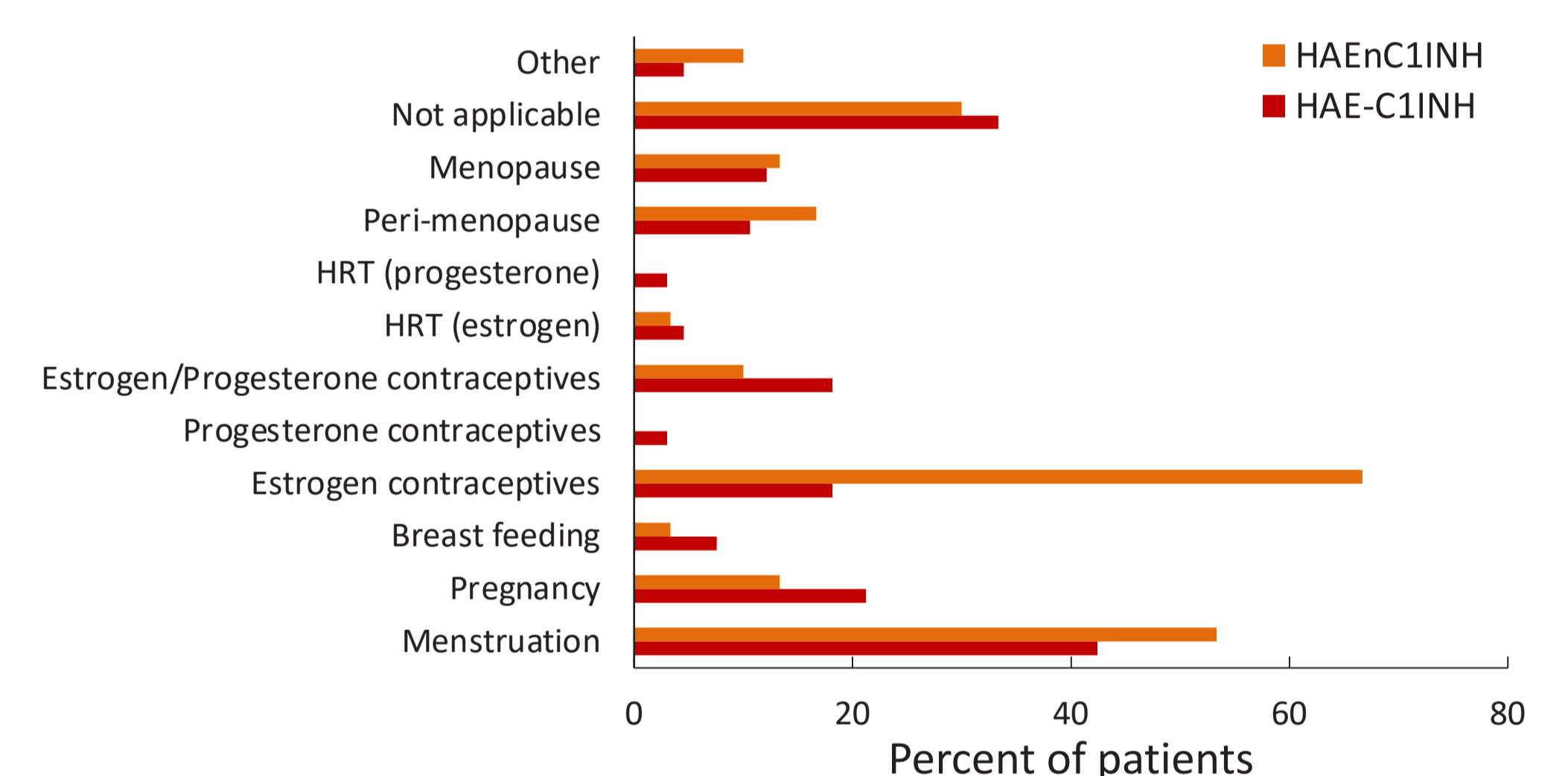


Figure 4. Women further identified menstruation (HAE-C1INH 42%; HAEnC1INH 53%) and estrogen contraceptives (HAE-C1INH 18%; HAEnC1INH 67%) as triggers.

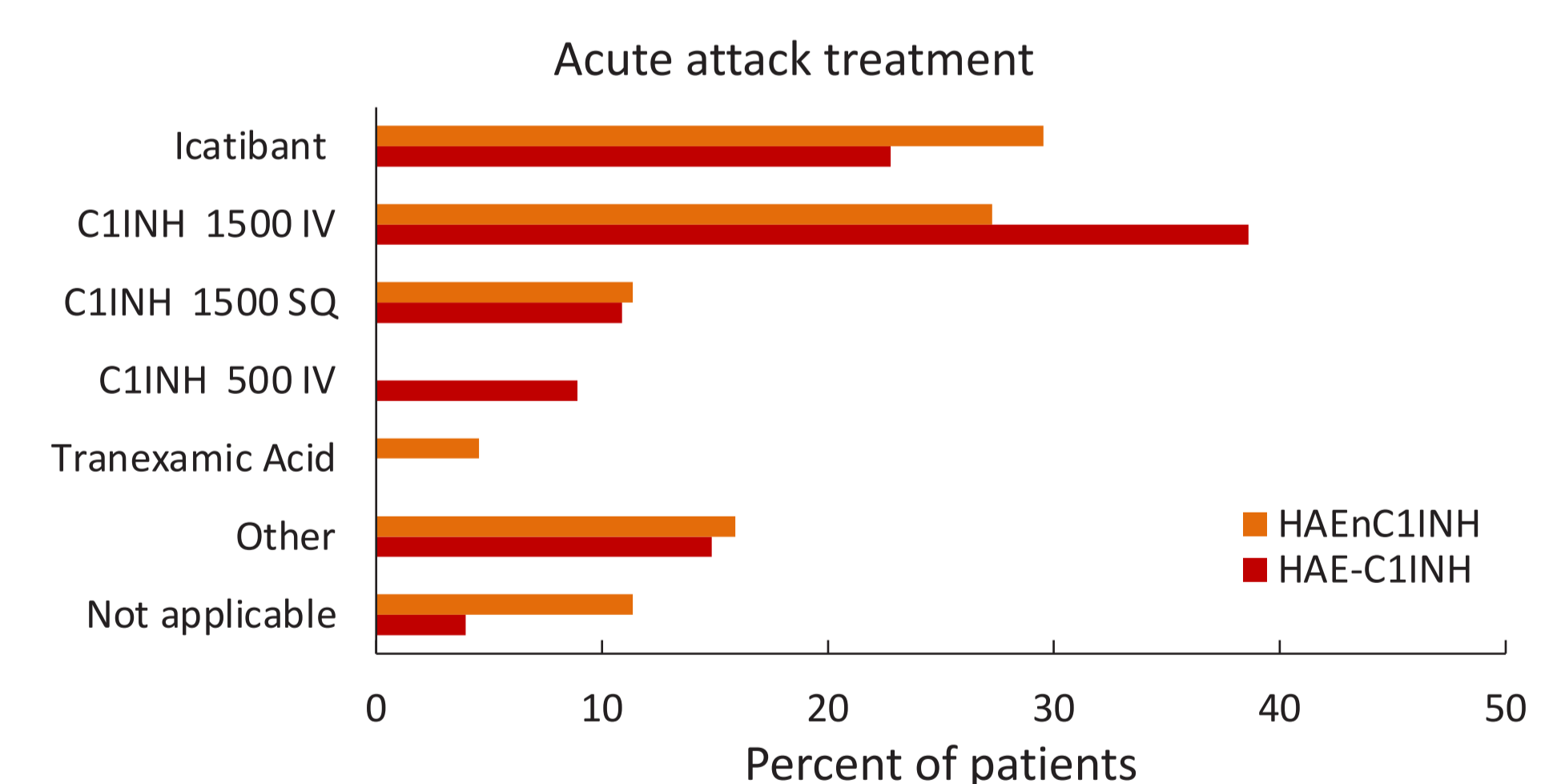


Figure 5. The most common treatments for acute attacks were pdC1INH (HAE-C1INH 58%; HAEnC1INH 39%) and icatibant (HAE-C1INH 23%; HAEnC1INH 30%). Other = prednisone, adrenaline, antihistamine, pain medication, herbal therapy

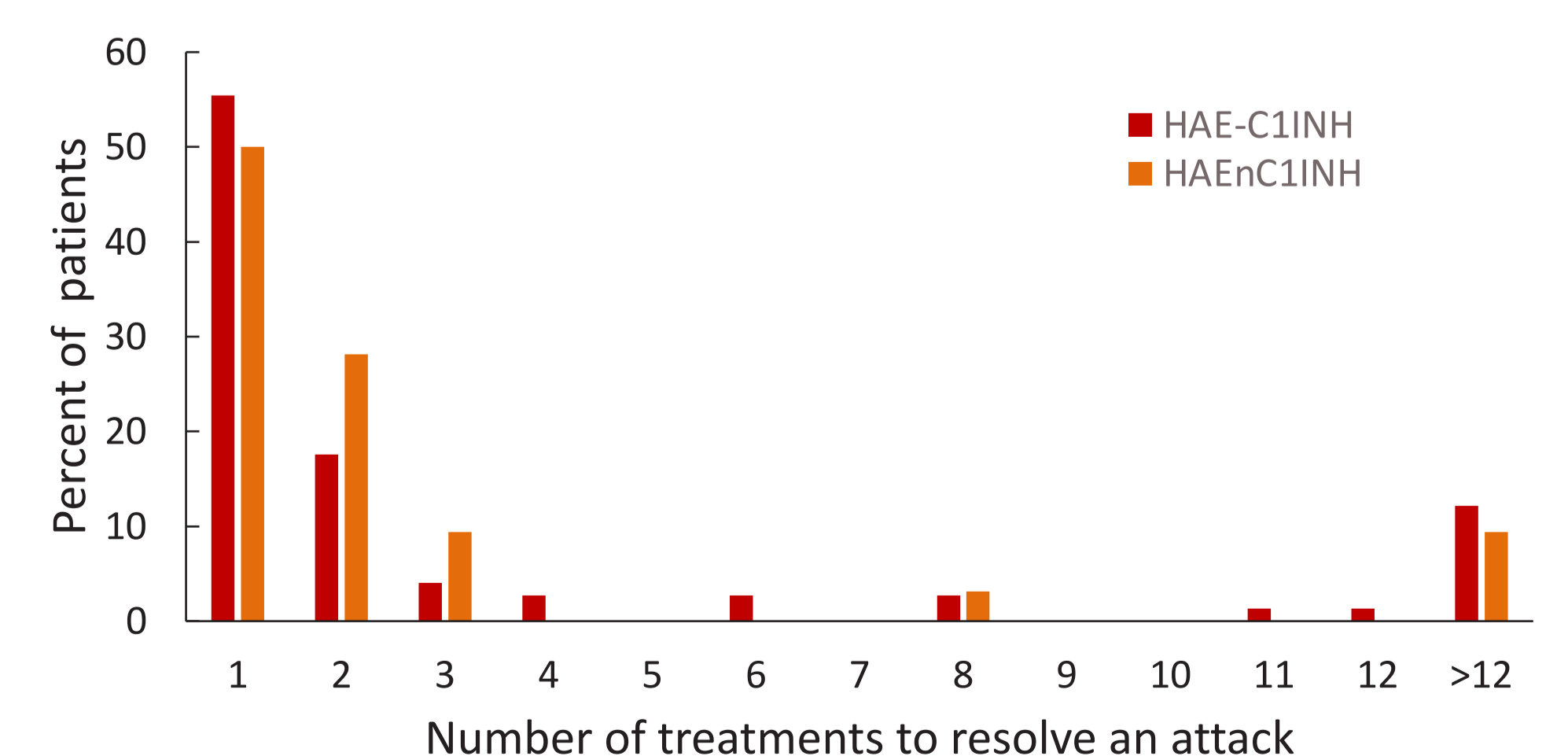


Figure 6. One treatment resolved the attack for 55% HAE-C1INH and 50% HAEnC1INH patients and 2 treatments for 18% and 28%, respectively.