

J. Badiou¹, M. Cooper¹, D. Dumbrille¹, S. M. Kelly², P. K. Keith³, G. Lacuesta⁴, A. Kanani⁵, C. Kalicinsky⁶, W. H. Yang^{2,7}

¹Hereditary Angioedema Canada; ²Red Maple Trials Inc., Ottawa, ON; ³McMaster University, Hamilton, ON; ⁴Dalhousie University, Halifax, NS; ⁵University of British Columbia, Vancouver, BC; ⁶University of Manitoba, Winnipeg, MB; ⁷University of Ottawa, Ottawa, ON.

Abstract

Introduction: Hereditary angioedema (HAE) is a genetic disorder resulting in low or dysfunctional C1 inhibitor (C1-INH): Type 1/2 HAE and/or dysregulation of bradykinin production: HAEnC1-INH (HAE with normal C1INH). HAE is characterized by recurring, painful swelling occurring in any part of the body. Laryngeal swelling can be fatal and abdominal swelling is very painful. Consequently, HAE patients often require emergency room (ER) visits.

Methods: In 2020, an online survey was sent to all members of HAE Canada. Survey results were collated, sorted by self-reported Type 1/2 HAE and HAEnC1-INH and expressed as percent of respondents.

Results: 106 (76% female) patients with Type 1/2 HAE and 45 with HAEnC1-INH (84% female) responded to the survey. In the prior year, most did not attend the ER for treatment of an angioedema attack (Type 1/2 HAE: 73%; HAEnC1-INH: 69%). For those who did, throat and facial swelling (39% vs 25%) and problems breathing (25% vs 11%) were more frequent for patients with HAEnC1-INH compared to Type 1/2 HAE.

Prior to treatment access, 40% of Type 1/2 and 51% of HAEnC1-INH patients had >6 ER visits in the prior year and (77.5% Type 1/2; 79.31% HAEnC1-INH had at least 1 visit, respectively). After access to treatment, 10% of Type 1/2 and 15% of HAEnC1-INH patients had >6 visits to the ER in the prior year while 44.59% vs 57.69% had at least 1 visit.

Discussion: These results suggest that providing treatment to patients that can be taken at home reduces ER visits and possibly health care costs.

Introduction

Hereditary angioedema (HAE) is a genetic disorder resulting in low or dysfunctional C1 inhibitor (C1-INH) termed Type 1/2 HAE and/or dysregulation of bradykinin production termed HAEnC1-INH (HAE with normal C1INH). HAE is characterized by recurring, painful swelling occurring in any part of the body. Laryngeal swelling can be fatal and abdominal swelling is very painful. Consequently, HAE patients often require emergency room (ER) visits. At-home treatment options (e.g., C1INH, icatibant, lanadelumab) are now available for some patients.

Objective

We compared Emergency Room (ER) use by HAE patients before and after availability of home treatment.

Methods

In 2020, an online survey was sent to all members of HAE Canada. Survey results were collated, sorted by self-reported HAE C1INH and HAEnC1-INH and expressed as percent of respondents. Responses to questions on emergency room use and symptoms requiring emergency treatment were analysed for this report.

Results

The survey collected data from 209 HAE patients and caregivers (child/youth). Of these 209 respondents, 106 adults (76% female, average age 52) had Type I or Type II HAE and 45 adults (84% female, average age 49) had HAE with normal C1INH.

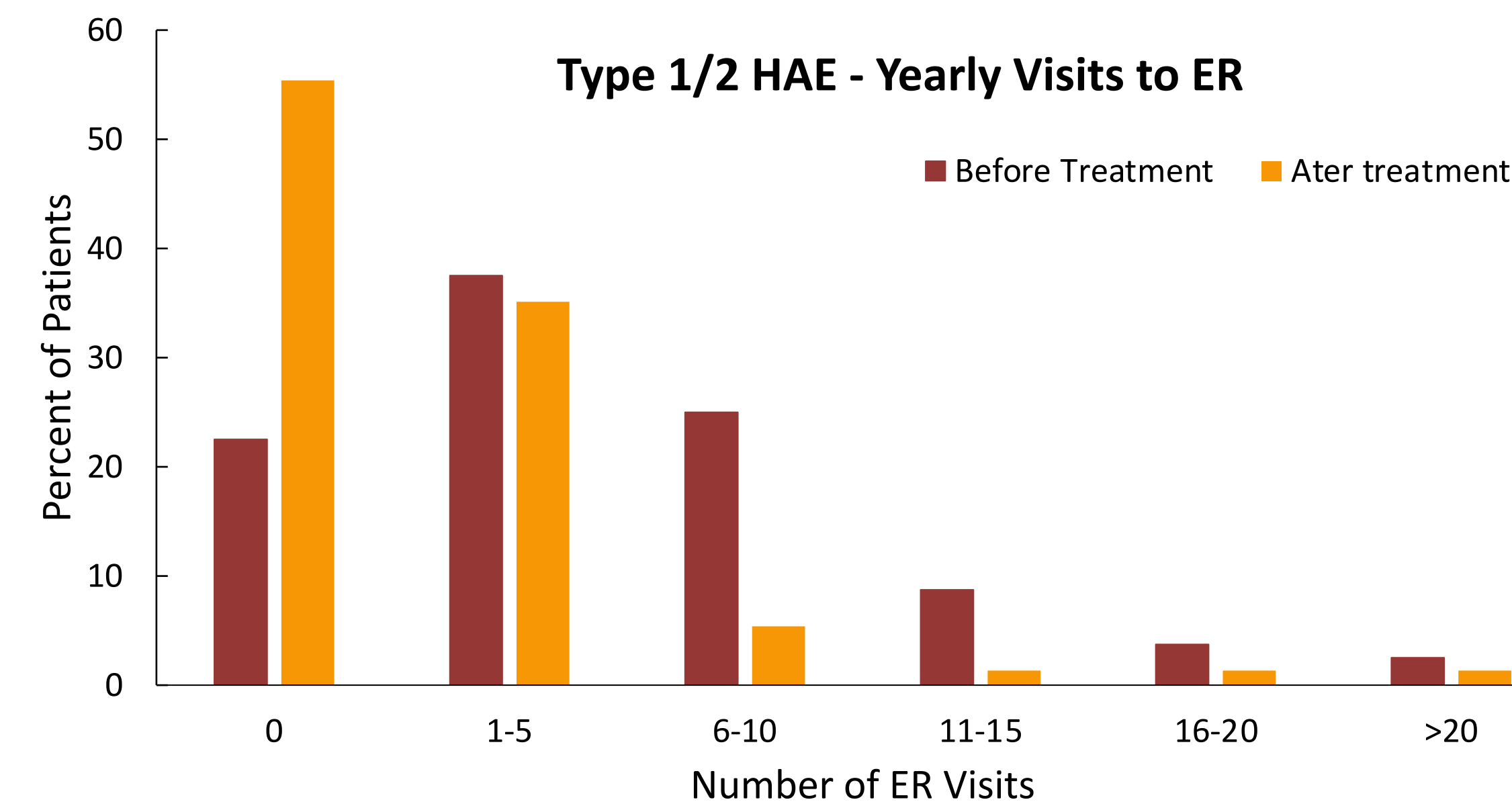


Figure 1. Before access to home treatment, most patients with Type 1/2 HAE (n=80); had 1 to 10 visits to the ER per year. A small number had more frequent (>10) events and 23% did not go to the ER. After access to home treatment (n=75), there was a shift to fewer ER visits. More than half had no visits and many fewer had 6 or more visits.

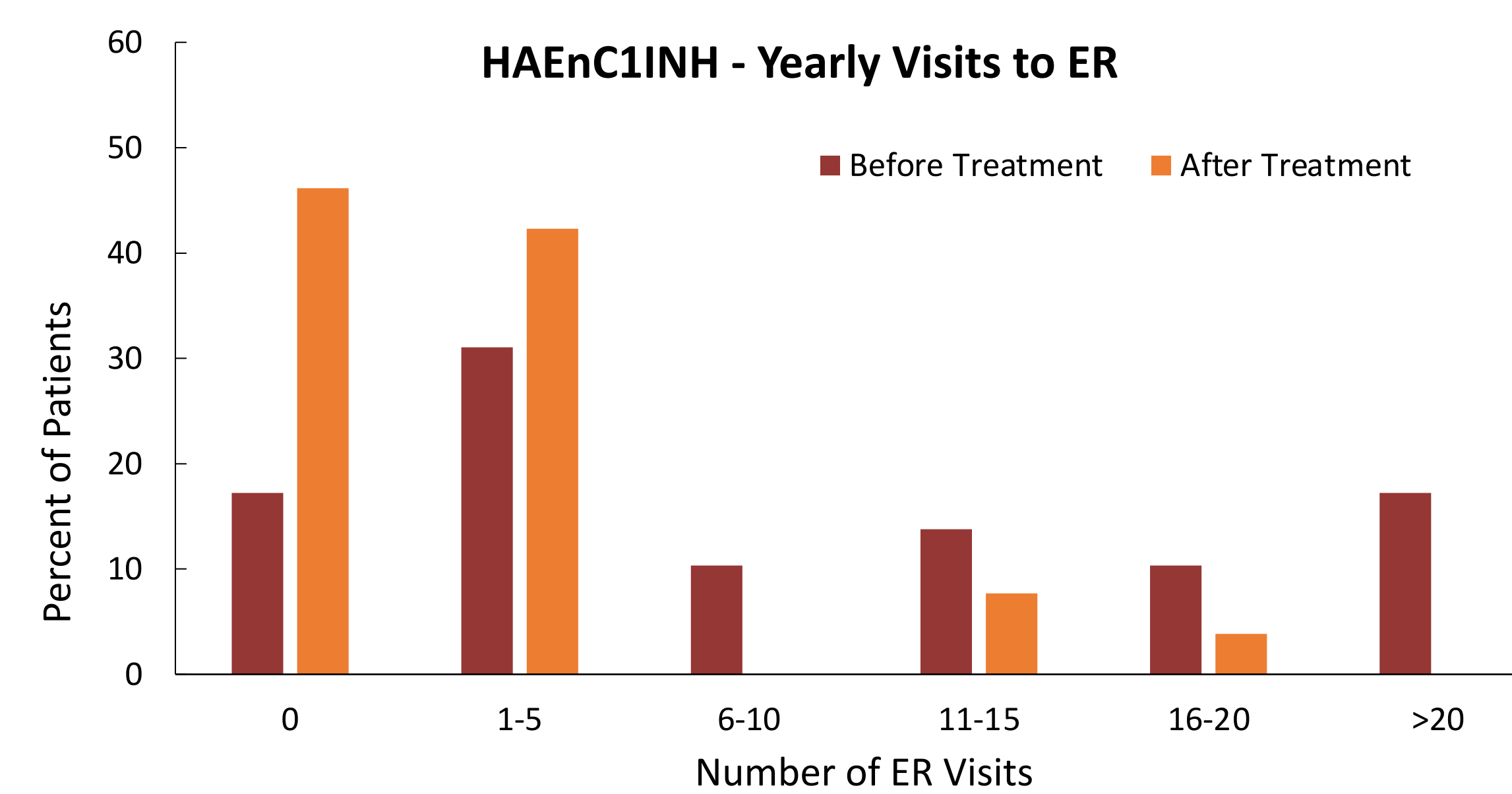


Figure 2. Patients with HAEnC1INH had frequent visits to the ER prior to home treatment access (n=29). Fifty-two percent had more than six ER visits per year. After access to home treatment (n=26), there was a large increase in those having no ER visits and a consequent decrease in more frequent visits.

ER Visits	Before Treatment				After Treatment			
	0	1-5	6-10	>10	0	1-5	6-10	>10
Type 1/2 HAE	23%	37%	25%	15%	55%	35%	5%	4%
HAEnC1INH	17%	31%	10%	41%	46%	42%	0%	12%

Table 1. Before access to home treatment, significantly more HAEnC1INH than Type 1/2 HAE patients had frequent (>10/year) visits to the ER. After access to home treatment, about half the patients had no ER visits and HAEnC1INH ER visit frequency was similar to that of those with Type 1/2 HAE.

Results (continued)

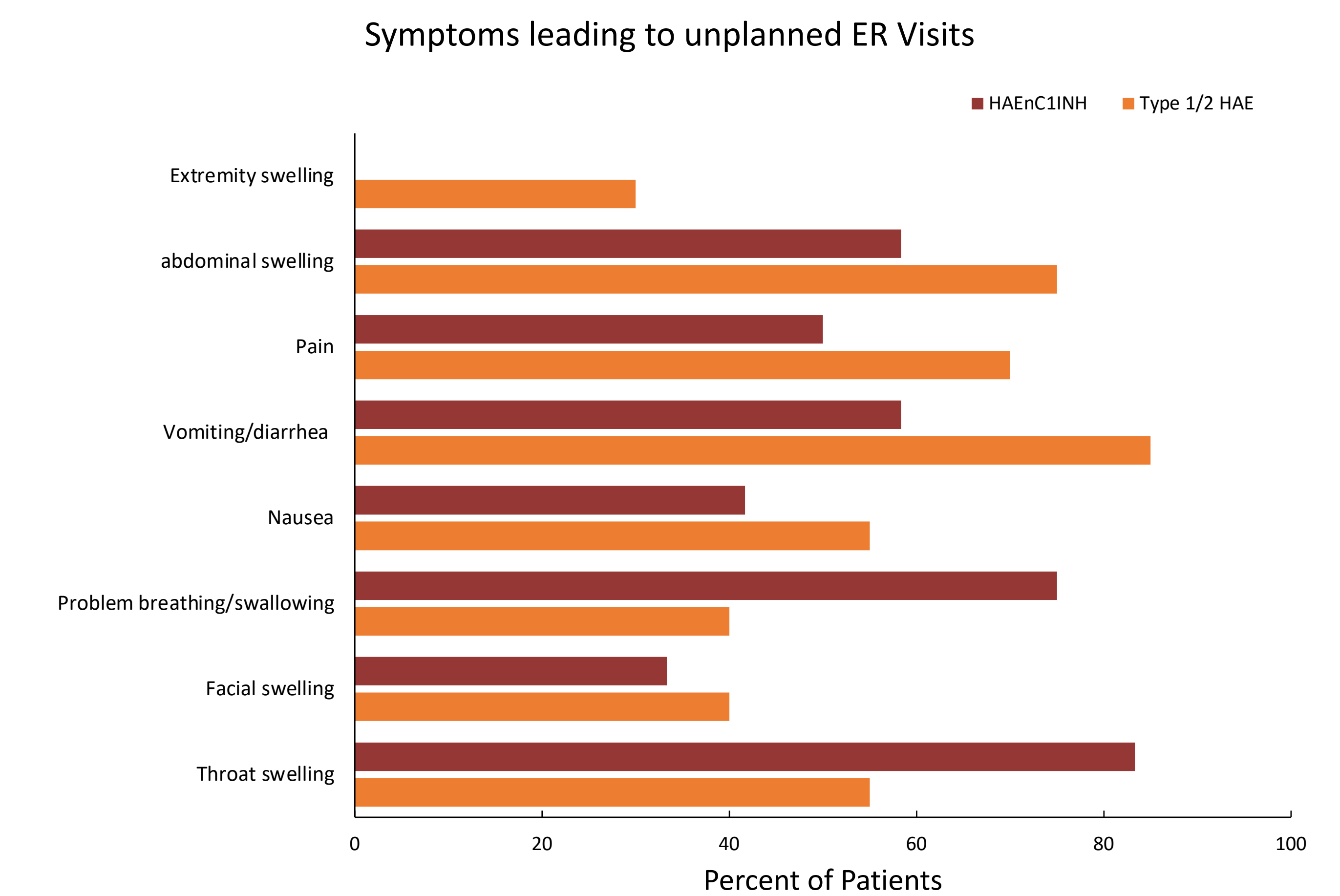


Figure 3 Most Type 1/2 (73%) and HAEnC1-INH (75%) patients did not have unplanned Emergency Room (ER) visits in the prior year. For Type 1/2 HAE patients who did, the most common symptoms were: vomiting/diarrhea, abdominal swelling and pain. For HAEnC1INH patients who did, the most common symptoms were: problems breathing/swallowing and throat swelling. Note that extremity swelling did not lead to ER visits for HAEnC1INH patients.

Conclusions

- Our results indicate that access to treatment and home management reduced the need for Emergency Room visits for patients with Type 1/2 HAE as well as for those with HAEnC1INH
- Prior to home treatment access, a much higher proportion of patients with HAEnC1INH had more than 10 Emergency Room visits per year. In fact, 17% had more than 20 Emergency Room visits.
- The symptoms driving Emergency Room visits differed for Type 1/2 HAE and HAEnC1INH patients. Problems breathing/swallowing and throat swelling were more common for HAEnC1INH patients while gastrointestinal symptoms and pain were more common for Type 1/2 HAE patients.
- ER physicians should be aware of HAE so that prompt referral for diagnosis, treatment and home management strategies can reduce ER visits

Acknowledgements

We are grateful to the HAE Canada Board of Directors and the Advocacy Committee, all volunteers, and especially members who supported and completed this survey questionnaire.