

Characterizing the Negative Impact of Delayed On-Demand Treatment of HAE Attacks

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Background

- Hereditary angioedema (HAE) is a rare genetic disease resulting in deficiency (type I) or dysfunction (type II) in the C1-inhibitor protein and subsequent uncontrolled activation of the kallikrein-kinin system
- People living with HAE experience unpredictable, painful and debilitating attacks of tissue swelling in various locations of the body that can be life-threatening depending on the location affected
- WAO/EAACI 2021 updated guidelines recommend that all HAE patients¹
 - consider treating all attacks as early as possible
 - have access to sufficient medication to treat two attacks
 - carry on-demand treatment at all times, regardless of prophylactic therapy
- Although self-administration of on-demand parenteral treatments has enhanced overall HAE attack management,² it is known that people living with HAE may delay on-demand treatment administration^{3,4}

Objective

- The objective of this analysis was to characterize the impact of delaying treatment of an HAE attack

Methods

- The US Hereditary Angioedema Association (HAEA) recruited people living with HAE to complete an online survey
 - Recruitment was stratified to include 50% of patients taking on-demand therapy only and 50% receiving long-term prophylaxis + on-demand therapy
- The survey was self-reported and took respondents approximately 20 minutes to complete
- The survey was completed by 107 individuals between September 6 and October 19, 2022; response rate 69% (107/155)
- Respondents provided consent for their data to be used anonymously or in aggregate
- Analysis was performed using descriptive statistics

Results

- Respondents included 107 HAE patients, 80.4% female, mean age 41 years (range 16-83); 50% using on-demand therapy only, 50% using prophylaxis + on-demand therapy (**Table 1**)

Table 1. Respondent characteristics

Characteristic	Total (N=107)
Age, mean (range), years	41 (16-83)
Gender, n (%)	
Female	86 (80.4)
Male	21 (19.6)
Type of therapy, n (%)	
On-demand only	53 (49.5)
Prophylaxis and on-demand	54 (50.5)
On-demand treatments used	
Icatibant	84 (78.5)
C1 esterase inhibitor (recombinant)	13 (12.1)
C1 esterase inhibitor (human)	9 (8.4)
Ecallantide	1 (0.9)
Prophylactic treatments used, n (%) of those using prophylaxis (n=54)	
Lanadelumab	31 (57.4)
Bertralstat	7 (13.0)
C1 esterase inhibitor (subcutaneous)	7 (13.0)
Androgens/steroids	5 (9.3)
C1 esterase inhibitor (intravenous)	4 (7.4)

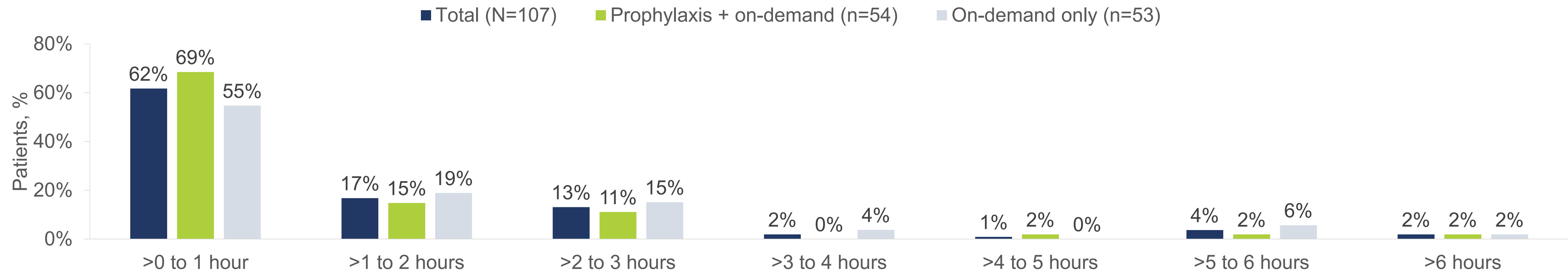
- HAE patients reported a mean time of 2.4 hours (median: 1 hour) before initiation of on-demand treatment after the first recognition of an attack (2.0 hours reported by those using prophylaxis + on-demand therapy; 2.7 hours reported by those using on-demand therapy only)
- The mean time to feel “in control” of their attack was 2.2 hours (median: 1 hour) following use of on-demand treatment, with only 1.4 hours when the attack was treated in 1 hour or less compared with 2.9 hours for those who waited ≥1 hour
- Most (62%) reported feeling “in control” of their attack in 1 hour or less (69% using prophylaxis + on-demand therapy; 55% using on-demand therapy only; **Figure 1**)

Disclosures

This study was sponsored by KalVista Pharmaceuticals. All authors met the ICMJE authorship criteria and had full access to relevant data. The authors had full editorial control of the data presented and provided final approval of all content. Neither honoraria nor payments were made for authorship.

Princess Ogbogu - Advisory Board/Consultant/Speaker/Grant support: AstraZeneca; GlaxoSmithKline; Sanofi
Hilary Longhurst - Honoraria/Travel grants and/or Speaker Bureau and/or Consultant/Clinical Research: BioCryst, CSL Behring, Intellia, Pharming, Shire/Takeda, and KalVista Pharmaceuticals
Sally van Kooten and Markus Heckmann - Employees of KalVista Pharmaceuticals.
Neil Malloy - Consultant fees from KalVista Pharmaceuticals.
Julie Ulloa - Consultant fees from KalVista Pharmaceuticals.
Ricardo Zwiener - Speaker: Takeda, CSL Behring, Novartis, Sanofi, Pint Pharma; Advisor: Takeda, CSL Behring, AbbVie, KalVista, Pint Pharma; Researcher: Takeda, Sanofi

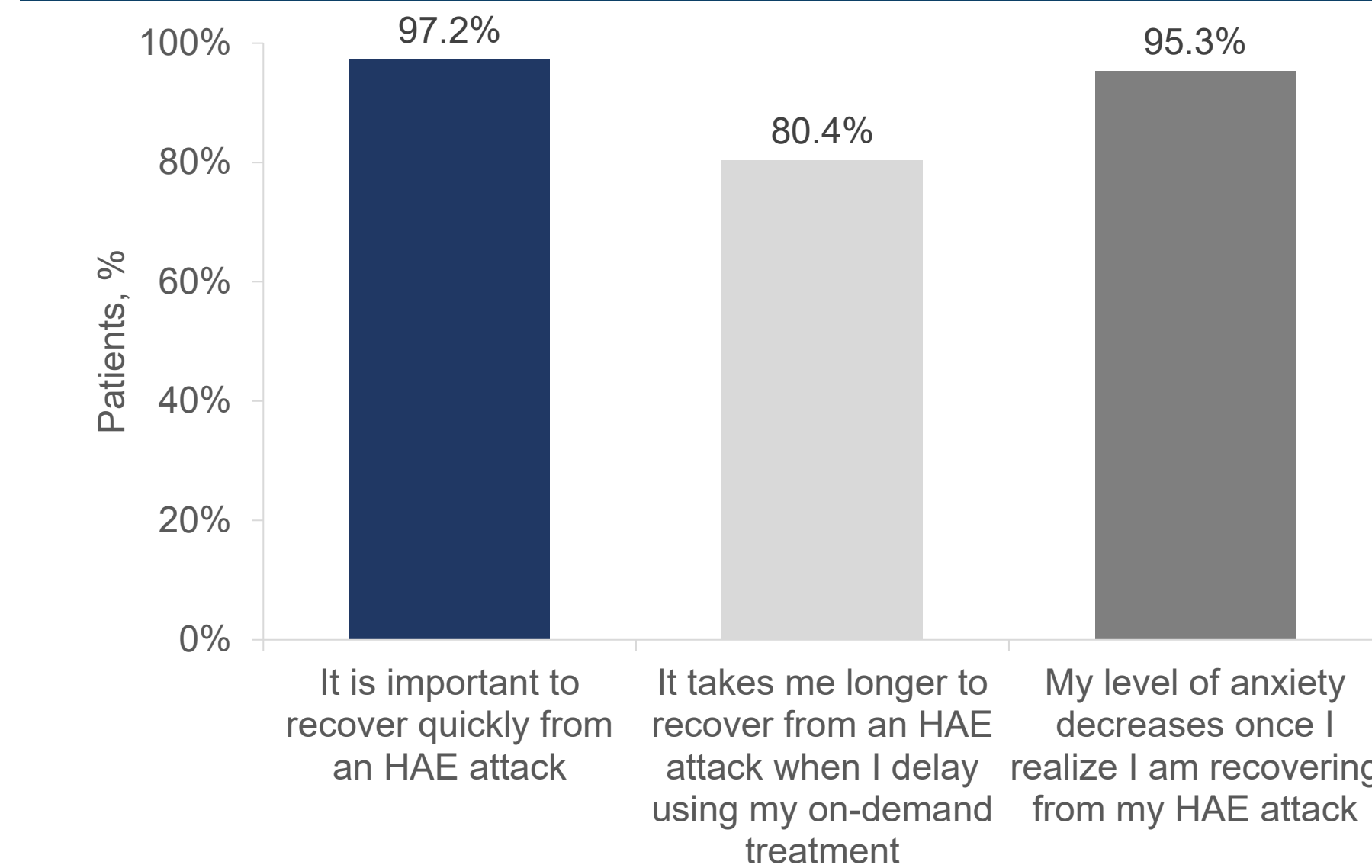
Figure 1. Time to feel “in control” of an HAE attack following use of on-demand treatment



Survey participants were asked how long (in hours) it takes for them to feel “in control” of their attack following use of on-demand treatment.

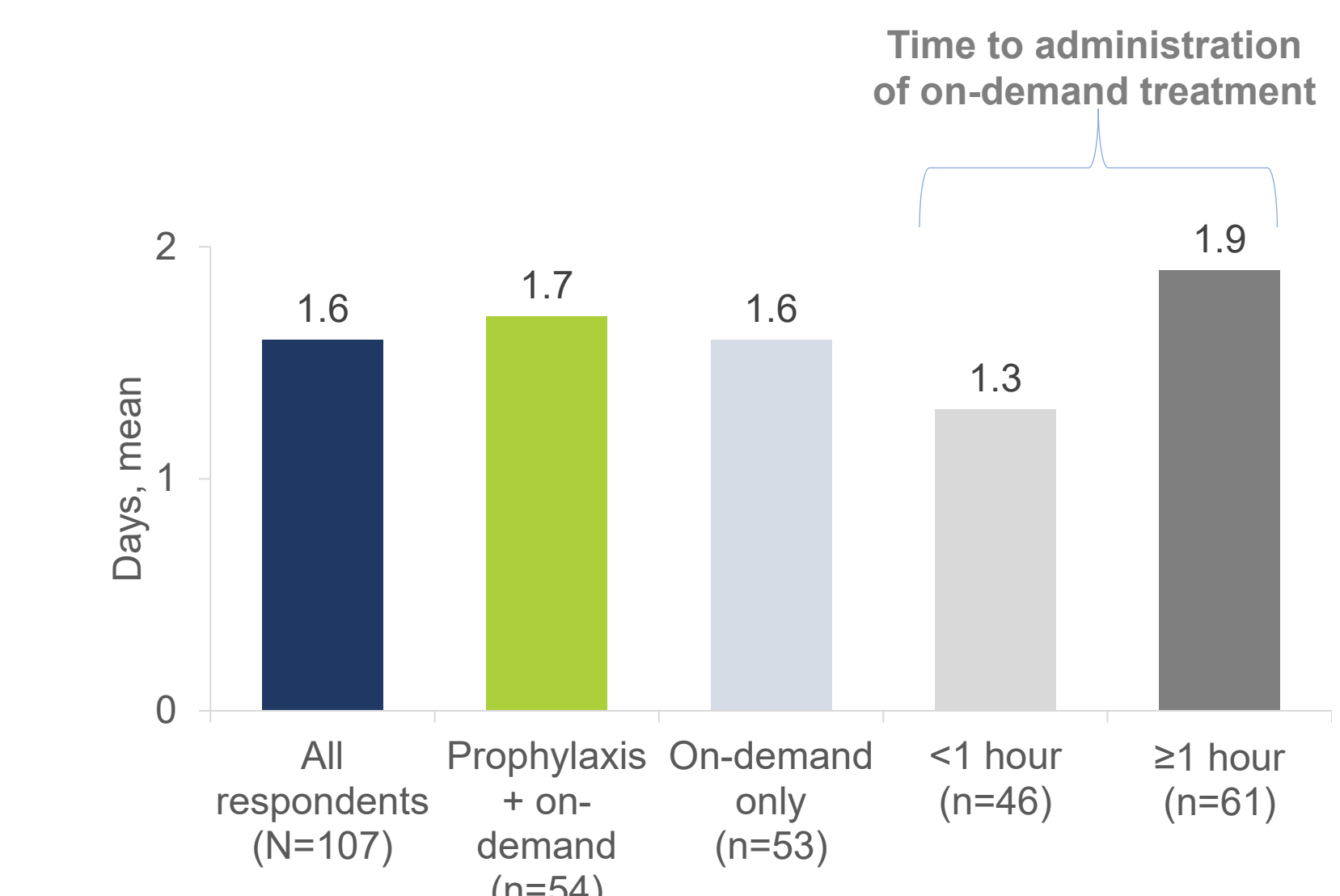
- Nearly all (97.2%) agreed that it is important to recover quickly from an HAE attack (**Figure 2**)
- The majority (80.4%) agreed that attack resolution takes longer when on-demand treatment is delayed
- Most (95.3%) reported that their anxiety decreases once they realize they are recovering from the attack
- Overall, full attack resolution took a mean of 1.6 days (median: 2 days); longer (1.9 days) when treatment was delayed for ≥1 hour. (**Figure 3**)
- Many (65.2%) experienced attack resolution in <1 day when the attack was treated in less than 1 hour compared to 36.1% of those who waited ≥1 hour

Figure 2. Survey statements and proportion of agreement



Survey participants were asked if they agreed with the presented statements and selected ‘Yes’ or ‘No’.

Figure 3. Time to feel “fully recovered” from an HAE attack following use of on-demand treatment



Survey participants were asked how long (in days) it takes for them to fully recover from their attack following use of on-demand treatment.

Conclusions

- Results highlight that delayed treatment of HAE attacks negatively impacts the time to feeling in control of an HAE attack and time to feeling fully recovered**
- Both the time to feeling in control of an HAE attack and time to feeling fully recovered were shorter for patients who treated HAE attacks in <1 hour compared with those who waited ≥1 hour**
- Survey responses indicated that people living with HAE understand the importance of treating early in their HAE attack journey and recognize that earlier treatment translates to quicker recovery and resolution**
- Patients also recognized that their level of anxiety decreases once they realize that they are recovering from an attack**

Presented at:

American Academy of Allergy, Asthma & Immunology (AAAAI) Annual Meeting - February 23-26, 2024, Washington, DC

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