

Recognizing the Importance of Early On-Demand Treatment in the Hereditary Angioedema (HAE) Attack Journey

Douglas H. Jones,¹ Princess Ogbogu,² Sally van Kooten,³ Markus Heckmann,³ Sherry Danese,⁴ Ledia Goga,^{3a} Markus Magerl⁵

¹Rocky Mountain Allergy at Tanner Clinic; Layton, Utah, United States; ²Pediatric Allergy, Immunology, and Rheumatology, University Hospitals Rainbow Babies and Children's Hospital, Cleveland, Ohio, United States; ³KalVista Pharmaceuticals, Inc.; Cambridge, Massachusetts, United States; ⁴Outcomes Insights; Agoura Hills, California, United States; ⁵Charité, Universitätsmedizin Berlin, Institute of Allergology, Berlin, Germany.

^aEmployee of KalVista Pharmaceuticals at the time the study was conducted

Background

- Hereditary angioedema (HAE) is a genetic disease resulting in deficiency (type I) or dysfunction (type II) in the complement-1 esterase inhibitor (C1-INH) protein and subsequent uncontrolled activation of the kallikrein kinin system (KKS).
- People living with HAE experience painful and debilitating attacks of tissue swelling in various locations of the body that can be life-threatening depending on the location affected.
- Symptoms of an HAE attack can potentially interfere with the ability to conduct daily activities (work, school, social activities)¹
- WAO/EAACI 2022 updated guidelines recommend that all HAE patients:²
 - 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,³ it is known that people living with HAE may delay on-demand treatment administration, missing the opportunity for optimal outcomes^{4,5}

Objective

- The objective of this survey was to characterize key aspects of HAE attacks (HAE attack journey), including on-demand treatment patterns, drivers of motivation to treat and goals for recovery, as reported by individuals living with HAE

Methods

- The US Hereditary Angioedema Association (HAEA) recruited people living with HAE to complete an online survey
- 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

References

- Bygum A. Hereditary angioedema—consequences of a new treatment paradigm in Denmark. Acta Derm Venereol. 2014;94(4):436–41.
- Maurer M., et al. The international WAO/EAACI guideline for the management of hereditary angioedema - The 2021 revision and update. World Allergy Organ J. 2022 Apr 7;15(3):100627.
- Longhurst HJ., et al. HAE international home therapy consensus document. Allergy Asthma Clin Immunol. 2010 Jul 28;6(1):22.
- Hernández Fernandez de Rojas D., et al. Treatment of HAE attacks in the Icatibant Outcome Survey: an analysis of icatibant self-administration versus administration by health care professionals. Int Arch Allergy Immunol 2015; 167: 21–28.
- Zanichelli A., et al. Safety, effectiveness, and impact on quality of life of self-administration with plasma-derived nanofiltered C1 inhibitor (Berinert) in patients with hereditary angioedema: the SABHA study. Orphanet J Rare Dis. 2018; 13: 51.

Results

Table 1. Respondent Characteristics

Characteristic	Total (n=107)
Age, mean	41yrs (Range 16-83 yrs)
Age category, n (%)	
24yrs and younger	14 (13.1%)
25yrs to 44yrs	51 (47.7%)
45yrs and older	42 (39.3%)
Gender, n (%)	
Female	86 (80.4%)
Male	21 (19.6%)
Type of therapy, n (%)	
On-demand only	53 (49.5%)
Prophylaxis with on-demand	54 (50.5%)
Level of anxiety when anticipating on-demand treatment*	
High anxiety (8-10 on scale of 1 to 10)	26 (24.3%)
Moderate anxiety (4-7 on scale of 1 to 10)	34 (31.8%)
Low anxiety (2-3 on scale of 1 to 10)	28 (26.2%)
No anxiety (1 on scale of 1 to 10)	19 (17.8%)
Time to HAE attack treatment	
Immediately treat all attacks	15 (14.0%)
Do not immediately treat all attacks	92 (86.0%)

*Anxiety scale 1 "not anxious" to 10 "extremely anxious"

Figure 1. On-demand Treatments Used

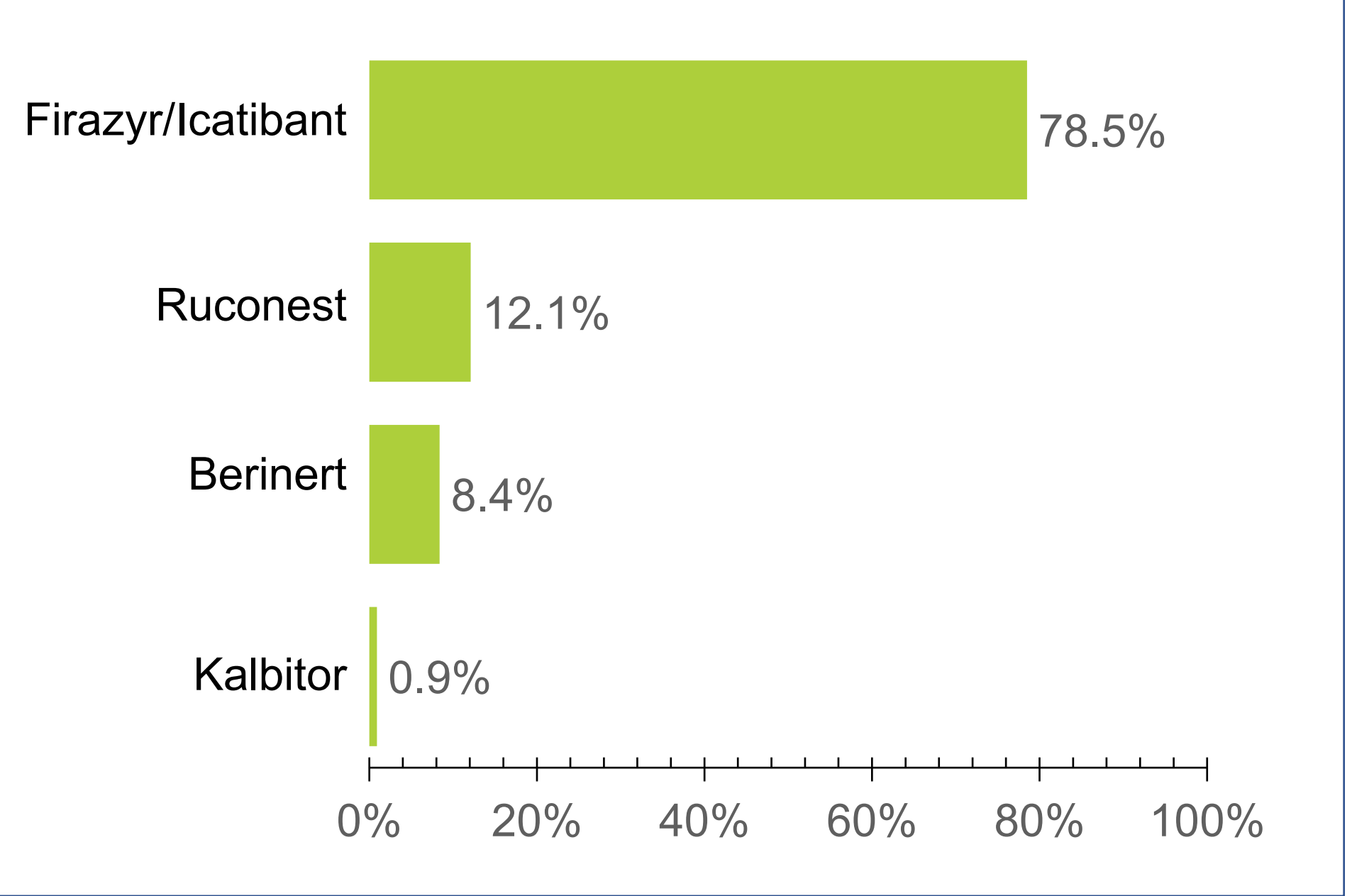
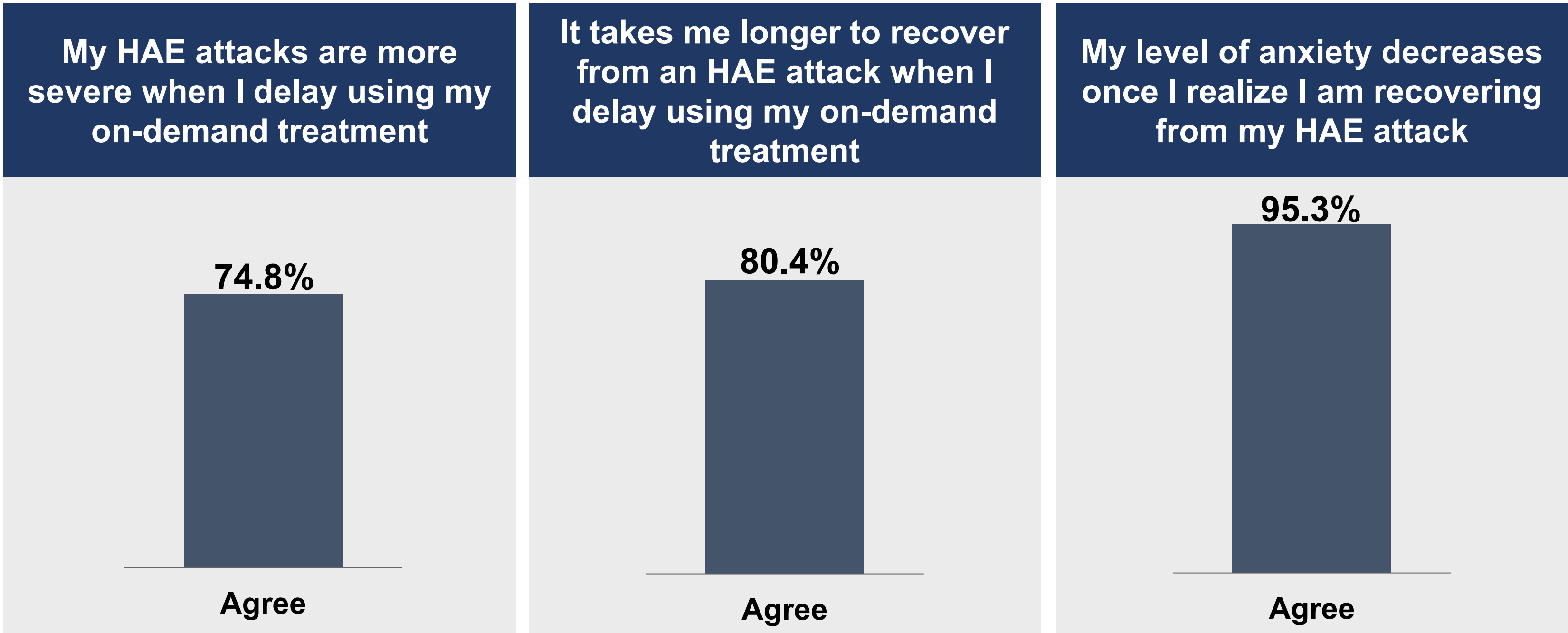


Figure 2. Survey Statements and Proportion of Agreement

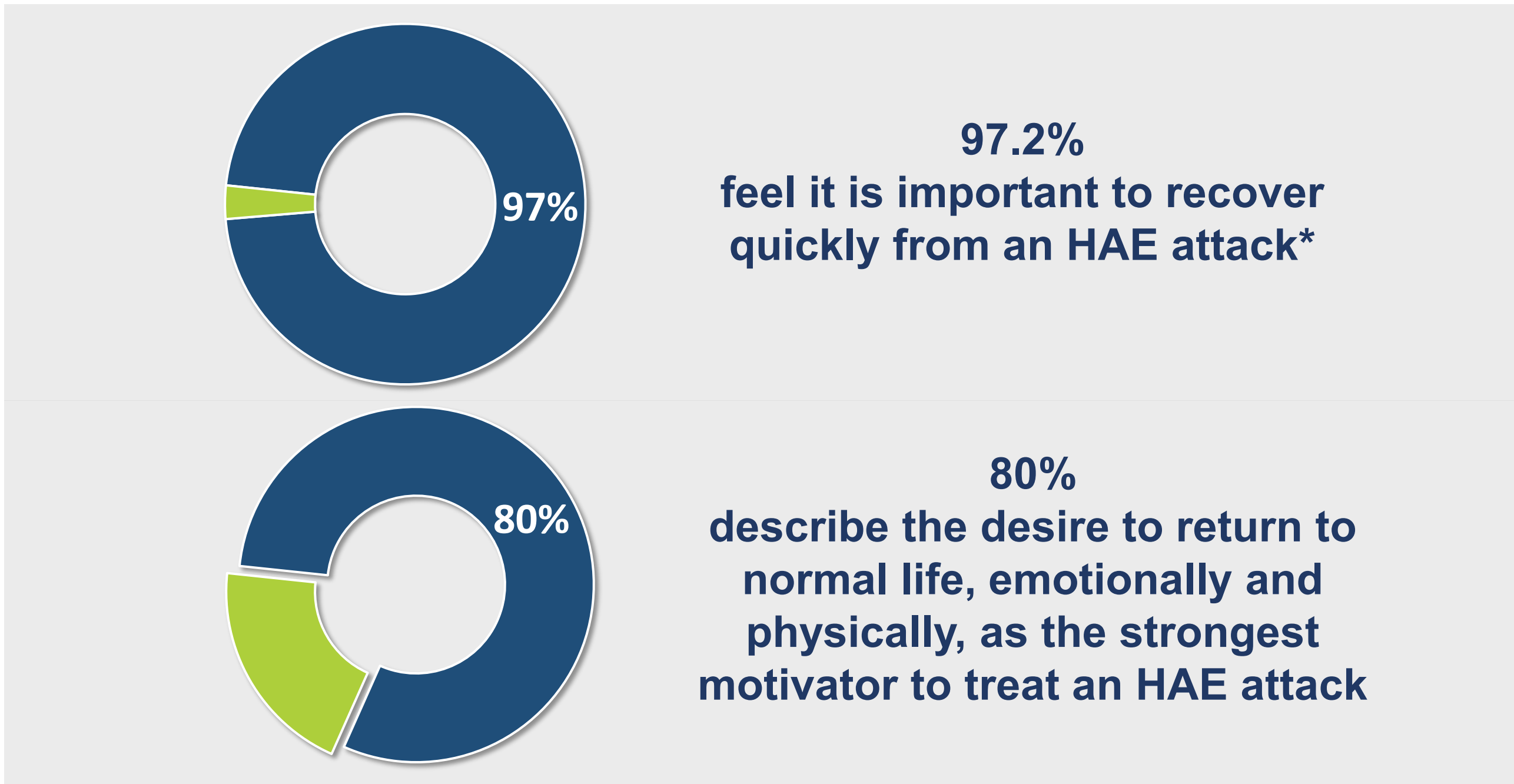


Survey participants were asked if they agreed with the presented statements and selected 'Yes' or 'No'.

Table 2. Statements Used by Respondents to Describe the Urgency to Recover Quickly from an HAE Attack

"I treat quickly because I don't want to feel miserable all day and deal with it becoming worse or spreading to another part of my body."	"It's important that I recover because attacks delay my day-to-day activities like going to work, my social life, my time with my family. Attacks bring my spirits down. It hurts me to see my loved ones observing my pain and suffering."
"Treating an attack early allows me to return to normal activities right after treatment. I may have slight residual swelling for several more hours, but that never hampers my daily activities."	"I've got things to do and it's annoying to either have to adjust to an attack or change plans because of an attack."
"The faster I recover, the faster I am able to feel like myself and just go back to regular life."	"Attacks immensely impact my quality of life. I am in a leadership role and not everyone at work understands HAE, so I want to be able to treat and get back to work as quickly as possible. I am also a very social person and don't want HAE to hold me back from living my life."
"I want to be able to live life to the fullest every day and not have to worry about missing out."	"I want to live a pain-free life that is not interrupted. I want to attend events, activities, and work as planned."
"I would initiate to help the attack resolve more quickly. The sooner I treat, the better. If I wait a longer amount of time, this will mean a longer time I am dealing with the attack. In the case of a stomach swelling, I could be dealing with a few more days until I am back to normal if I waited too long, depending on how severe the attack is."	"Attacks can quickly escalate, and then put you out of commission for a day or two. I would rather treat immediately and prevent that from happening."

Figure 3. Survey Statements and Proportion of Agreement



*Survey participants were asked if it was important to them to recover quickly from an HAE attack and selected 'Yes' or 'No'.

Conclusions

- Survey results highlighted patients' understanding of the importance of treating early in their HAE attack journey and underscored the implications early treatment has on their ability to return to normal activities
- People with HAE recognize that earlier treatment translates to quicker resolution, recovery, and return to normalcy
- Nearly all patients agree that it is important to quickly recover from their HAE attack
- Most describe the desire to return to normal life, emotionally and physically, as the strongest motivator to treat an HAE attack
- Additional studies are needed to explore if these findings align with patient behaviors when treating attacks with on-demand therapy

Disclosures

This study was sponsored by KalVista Pharmaceuticals, Inc. 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. Douglas H. Jones - Advisory Board/Consultant/Speaker: Amerimmune Allergy Testing, Inc., KalVista Pharmaceuticals, Inc., Pharvaris, Astra-Zeneca, BioCryst Pharmaceuticals, Regeneron/Sanofi, Zurvita Corporation, Pharming, and Shire/Takeda Princess Ogbogu - Advisory Board/Consultant/Speaker/Grant support: AstraZeneca; GlaxoSmithKline; Sanofi Sally van Kooten, Markus Heckmann and *Ledia Goga - Employees of KalVista Pharmaceuticals, Inc. (*at the time of the publication development) Sherry Danese - Consultant fees from KalVista Pharmaceuticals, Inc. Markus Magerl - Personal fees/Non-financial support: Shire Takeda, CSL Behring, Pharming, Biocryst, Kalvista, and Octapharma

Presented: 2023 US HAEA National Summit, July 20 - 23, 2023 in Orlando, Florida.

Please scan this QR code to view the poster after the congress.

