Recent Advances in Hereditary Angioedema Self-Administration Treatment: Summary of an International Hereditary Angioedema Expert Meeting

Timothy J. Craig

College of Medicine, Penn State University, Hershey Medical Center, Hershey, Pa., USA

A significant amount of progress has been made in recent years in the understanding and treatment of hereditary angioedema (HAE), with more products being licensed for use worldwide (table 1). With the recent registration and introduction of pasteurised-nanofiltered C1 esterase inhibitor for self-administration, patients have new treatment opportunities and an increasing number of healthcare providers are offering more patients the option of self-administration therapy. However, the international HAE expert meeting demonstrated slow uptake of this treatment option. In general, we need to increase awareness of the benefits of self-administration and encourage patients to enter training programmes linked to self-injection of treatment.

As discussed in the article by Cicardi et al. [1], the guidelines for HAE diagnosis and treatment have progressed over the years, with recent guidelines often reflecting current practice in the majority of areas, including for self-administration. As identified through both the guidelines and the international HAE expert meeting, training and follow-up is very important both for healthcare staff and patients, especially because self-administration is an option that is increasingly being adopted.

The results of the survey reported by Caballero et al. [2] identified a range of experience worldwide and demonstrated that some healthcare staff request specialist training from personnel in other departments, such as

haemophilia and immunology departments. The sharing of expertise is vital to ensure patients are fully trained and are competent in self-administration.

On the practical side of self-administration, as discussed by Symons et al. [3], patients appear to be learning skills quicker than expected and in fewer sessions than guidelines recommend. Nevertheless, the ability to visit patients' homes is restricted in some countries, highlighting the need for potential investment in home care nursing packages. Since home and self-care have evolved differently in different countries, the authors hope that highlighting these differences can stimulate discussion and sharing of ideas leading to improved techniques and education worldwide.

Boysen et al. [4] reported some of the challenges faced by HAE patients and how they have been resolved. One of the main barriers faced by patients being treated in hospital is convincing the treating physician that HAE requires different therapy to the 'usual angioedema'. Moving treatment into the home, especially with self-infusion, removes this barrier and also ensures early therapy. As previously shown, rapid treatment of an HAE attack can markedly reduce time to recovery and morbidity, improves absenteeism and may also be life-saving. More needs to be done to raise awareness of this rare disease and the life-saving implications of rapid treatment.

Table 1. Summary of approved treatments^a

Drug	Country	Date	Indicated for
Berinert	Germany/Hungary	1979	Acute attacks
	23 EU countries	2008	Acute attacks
	USA	2009	Acute attacks
	Canada	2010	Acute attacks
	23 EU countries	2011	Self-administration for acute attacks
	USA	2012	Self-administration for acute attacks
	23 EU countries	2013	Short-term prophylaxis, including self-administration
Cinryze	USA	2008	Prophylaxis
	USA	2009	Self-administration for prophylaxis
	EU	2011	Acute attacks, routine prevention and short-term prophylaxis, including self-administration
Ruconest	EU	2010	Acute attacks
Icatibant	EU	2008	Acute attacks
	EU	2011	Self-administration for acute attacks
	USA	2011	Self-administration for acute attacks
Ecallantide	USA	2009	Acute attacks

^a Approvals in countries that participated in the survey.

To aid rapid treatment of an HAE attack, self-administration is a viable treatment option. To make further progress, physicians need to follow their country guidelines more closely, modify them as needed and establish support networks for patients where they are able to seek guidance on care, including self-administration training, effective use of medications, managing their therapy and reporting adverse events.

Self-administration is universally recommended in recent guidelines and in clinical practice. As awareness increases so will the uptake of this treatment option. Although patients are being offered self-administration, not all patients are choosing it and more needs to be done to encourage uptake. The goal of many HAE patients is to be able to manage their own disease and self-administration of therapy can make this desire a reality.

Once again, I would like to express my thanks to all the participants of the international HAE expert meeting for their valuable contributions, and to CSL Behring for their support in the production of this supplement.

Acknowledgements

Editorial assistance was provided by Meridian HealthComms, Plumley, UK, funded by CSL Behring. The survey, international HAE expert meeting and preparation of the supplement were supported by an unrestricted educational grant from CSL Behring.

Notes

Berinert is a registered trademark of CSL Ltd. Group of Companies.

Cinryze is a registered trademark of ViroPharma Biologics Inc.

Firazyr is a registered trademark of Shire and its affiliates.

Kalbitor is a registered trademark of Dyax Corp.

Cetor is a registered trademark of Stichting Sanquin Bloedvoorziening.

Rhucin is a registered trademark of Pharming Group NV.

All trademarks and registered trademarks are the property of their respective owners.

Disclosure Statement

Dr. Craig speaks for CSL Behring, ViroPharma, Shire and Dyax. He does research for Pharming, CSL Behring, ViroPharma, Shire and Dyax. He is a consultant for CSL Behring and Dyax and has received educational grants from Dyax, ViroPharma and CSL Behring.

References

- 1 Cicardi M, Craig TJ, Martinez-Saguer I, Hébert J, Longhurst HJ: Review of recent guidelines and consensus statements on hereditary angioedema therapy with focus on self-administration. *Int Arch Allergy Immunol* 2013;161(suppl 1):3–9.
- 2 Caballero T, Sala-Cunill A, Cancian M, Craig TJ, Neri S, Keith PK, Boccon-Gibod I, Bethune C, Bork K: Current status of implementation of self-administration training in various regions of Europe, Canada and the USA in the management of hereditary angioedema. *Int Arch Allergy Immunol* 2013;161(suppl 1):10–16.
- 3 Symons C, Rossi O, Magerl M, Andritschke K: Practical approach to self-administration of intravenous C1-INH concentrate: a nursing perspective. *Int Arch Allergy Immunol* 2013;161(suppl 1):17–20.
- 4 Boysen HB, Bouillet L, Aygören-Pürsün E: Challenges of C1-inhibitor concentrate self-administration. *Int Arch Allergy Immunol* 2013;161(suppl 1):21–25.