

# WAO/EAACI HAE TREATMENT GUIDELINE - OVERVIEW OF 2017 REVISIONS

## Fact Sheet

The 2012 World Allergy Organization (WAO)<sup>1</sup> and the 2017 WAO/European Academy of Allergy and Clinical Immunology (EAACI)<sup>2</sup> guidelines were generated by consensus panels of experts to help guide and inform clinical decision-makers treating type 1 and type 2 Hereditary Angioedema (HAE)

Experts nominated to the panels fulfilled at least 1 of the following criteria<sup>1,2</sup>:



- Extensive clinical experience in the treatment of HAE
- Relevant publications in the field of HAE
- Relevant experience in evidence-based medicine

To ensure a global expertise and consensus, representative panels of experts were selected from around the world<sup>1,2</sup>

The table below highlights key points of evolution since the original publication of the guidelines in 2012<sup>1,2</sup>

This is not meant to be a complete review of the guidelines, and readers are strongly advised to consult the original publications for complete details

Analysis of changes between 2012 and 2017 guidelines is offered in light blue text, while black text is quoted directly from respective guidelines

Topic	Original 2012 <sup>1</sup>	Revised 2017 <sup>2</sup>
<b>Androgens</b>	Long-term prophylaxis with androgen derivatives is effective but must be regarded critically, especially on account of their androgenic and anabolic effects	<p><b>Recognition of the prevalence of adverse effects of androgens</b></p> <p>Androgens must be regarded critically, especially in light of their adverse androgenic and anabolic effects, drug interactions, and contraindications. Guideline authors suggest to use androgens as a second-line long-term prophylaxis. Side effects are numerous and involve the majority of patients</p>
<b>Diagnosis of children</b>	Screening children for HAE type 1 and type 2 should be deferred until the age of 12 months, and all offspring of an affected parent should be tested	<p><b>Greater emphasis on testing</b></p> <p>Children from HAE-affected families should be tested as soon as possible, and all offspring of an affected parent should be tested. All early complement testing performed in offspring of type 1 and type 2 patients should be repeated after the age of 1 year. Until a full investigation for HAE type 1 and type 2 is complete, all offspring should be considered to have HAE type 1 or type 2</p>

Topic	Original 2012 <sup>1</sup>	Revised 2017 <sup>2</sup>
<b>Long-term prophylaxis (patient consideration)</b>	Long-term prophylaxis should be considered in all severely symptomatic HAE type 1 and type 2 patients, taking into consideration the severity of disease, frequency of attacks, patient's quality of life, availability of resources, and failure to achieve adequate control by appropriate on-demand therapy	<p><b>Greater focus on the activity of the disease and routine assessment when deciding if long-term prophylaxis is appropriate</b></p> <p>Long-term prophylaxis should be individualized and considered in all severely symptomatic HAE type 1 and type 2 patients, taking into consideration the activity of the disease, frequency of attacks, patient's quality of life, availability of healthcare resources, and failure to achieve adequate control by appropriate on-demand therapy. It is recommended that prophylaxis be considered for patients who face events in life that are associated with increased disease activity. Successful long-term prophylaxis requires a high degree of compliance; therefore, the patient's preferences should be taken into consideration. Guideline authors currently recommend C1-INH as first-line long-term prophylaxis</p>
<b>Long-term prophylaxis (patient assessment)</b>		<p><b>More importance placed on the routine assessment for long-term prophylaxis</b></p> <p>It is recommended that patients be evaluated for long-term prophylaxis at every visit. Disease burden and patient preference should be taken into consideration</p>
<b>Management considerations</b>	All patients should have an action plan and product available to treat an attack of HAE	<p><b>Greater emphasis on integrated care</b></p> <p>It is suggested that HAE-specific, comprehensive, integrated care is available for all patients. All patients should have an action plan</p>
<b>On-demand treatment of attacks</b>	<p>All attacks that result in debilitation/dysfunction and/or involve the face, the neck, or the abdomen should be considered for on-demand treatment. Treatment of attacks affecting the upper airways is mandatory. Attacks should be treated as early as possible</p> <p>It is recommended that all patients should have on-demand treatment for 2 attacks and should carry on-demand treatment at all times</p>	<p><b>Increased recognition that all HAE attacks should be considered for on-demand therapy</b></p> <p>It is recommended that all attacks are considered for on-demand treatment. It is recommended that any attack affecting or potentially affecting the upper airway is treated. Attacks should be treated as early as possible</p> <p>It is recommended that all patients should have on-demand treatment for 2 attacks and should carry on-demand treatment at all times</p> <p>Early treatment is crucial in cases of upper airway involvement (tongue, uvula, larynx). Patients should self-administer treatment while awaiting transfer to hospital. It is extremely important to encourage all patients to seek further care immediately after administration of therapy. Upper airway swelling may progress or rebound, and repeat dosing may be necessary. Seeking emergency care after therapy is essential to reduce the risk of suffocation</p>
<b>Pre-procedural short-term prophylaxis</b>	The decision to give prophylaxis before a procedure depends on the patient's personal history and the likely risk associated with the procedure. The administration of short-term prophylaxis should be considered before surgeries, especially dental/intraoral surgery, where endotracheal intubation is required, where upper airway or pharynx is manipulated, and before bronchoscopy/endoscopy	<p><b>Shift from considering short-term prophylaxis to recommending short-term prophylaxis</b></p> <p>Despite the perceived benefits of pre-procedural prophylaxis with C1-INH concentrate, evidence for its efficacy is scarce. Case reports and series suggest that despite prophylaxis, swellings may occur even after relatively minor procedures.<sup>3,4</sup> However, several reports document a reduction in the incidence of swelling for both adults and children with pre-procedural prophylaxis, and the response appears to be dose-related.<sup>3,5,7</sup> Pre-procedural prophylaxis with C1-INH concentrate is therefore recommended for all medical, surgical, and dental procedures associated with any mechanical impact to the upper aerodigestive tract</p>

**References:** 1. Craig T, et al. *World Allergy Organ J.* 2012;5:182-199. 2. Maurer M, et al. *Allergy.* 2018; doi: 10.1111/all.13384. 3. Farkas H, et al. *Allergy.* 2012;67(12):1586-1593. 4. Aygören-Pürsün E, et al. *Allergy.* 2013;68(8):1034-1039. 5. Bork K, et al. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2011;112(1):58-64. 6. Farkas H, et al. *J Oral Maxillofac Surg.* 1999;57(4):404-408. 7. Magerl M, et al. *Ann Allergy Asthma Immunol.* 2017;118(1):110-112.

