



# Hereditary Angioedema (HAE)

## Recommendations for emergency treatment

Patient's name:

Country of residence:

Personal identity number:

Physician's name & tel no:

Hospital:

Date

Signature

**This patient is suffering from hereditary angioedema (HAE).**

C1 inhibitor (C1-INH, a complement factor) deficiency, which may cause local swellings and abdominal pain lasting several days, has been detected. Should edema in the region of the head and throat occur (risk of laryngeal edema!), immediate treatment with C1-INH concentrate is necessary. The patient normally carries his medication when travelling.

**Procedure for emergency treatment:**

1. Carefully dissolve 1 vial of dry substance (500 U\*) C1-INH in the solvent supplied (10 ml).  
**To avoid the formation of foam, do not shake the bottle!**
2. For both adults and children, slowly inject 20 U C1-INH per kg bodyweight intravenously. The remaining solution could also be administered. Repeat the dose if no effect is noticeable within 1-2 hours.
3. If C1-INH concentrate is not available, use 500-1000 ml fresh frozen plasma. As a final alternative, fresh plasma can be used.
4. In the case of laryngeal edema, intubation or even a tracheotomy may be necessary.
5. If laryngeal edema is suspected, send the patient directly to hospital.

**Warning!** Antihistamines have no effect in this form of edema, but corticosteroids and epinephrine may have some effect.

*\*1 unit is equivalent to the C1-INH activity in 1 ml fresh citrated plasma from healthy donors.*

**See [www.allaboutthae.com](http://www.allaboutthae.com) for general information on HAE.**