

Voluntary association for the study, therapy and fight against hereditary angioedema A.A.E.E. - ONLUS

The association was founded on 29th March 1980, and registered in Milan on 24th September 1980. Public licence n. A/4 21864 series H by deed of the notary Doct. Francesca Testa.

The association has the following aims:

- to spread the knowledge of the disease to allow a correct diagnosis.
- To get complete availability and high quality medical care to support all the patients with an adequate treatment.
- To issue a health identification document, officially recognised, and describing the disease the emergency therapy as well as the addresses of doctors to call for further information.
- To promote social-scientific meetings, congresses at a national and international level.
- To encourage meetings and exchanges of information among patients, patients and doctors and among doctors themselves.

This is a non-profit making voluntary association.

TREASURER : Sig.ra HUGUETTE LIMONTA SALA -
☎039/2014579

SECRETARIAT : Sig.ra GRAZIELLA BIGOTTO ☎
02/5472671

PUBLIC RELATION OFFICER :
VINCENZO PENNA

Frazione Quarto Inferiore, 192 - 14030 ASTI

☎/FAX 0141/299274

e-mail: info@angioedemaereditario.org

FOREIGN RELATIONS : NILLA CIAIRANO

☎ 011/9586940

e-mail: nillaci@fastweb.it

Angioedema due to the lack of C1-inhibitor :

what it is, what the symptoms are and how to treat it

The deficiency of the inhibitor of the first component of the Complement System, C1-inhibitor (C1-INH), can be caused by a genetic defect (hereditary angioedema) or by an increased consumption (acquired angioedema).

Symptoms are recurring swellings lasting 2 to 5 days. When the skin is involved, the affected area appears swollen, pale, not pitting and usually without a clear demarcation between affected and healthy parts. In the case of mucous involvement particularly dangerous is the glottis edema that can cause death for asphyxia. Frequently it can also occur in the bowel causing abdominal colic very similar to an «acute appendicitis».

It is not a disease caused by allergies and therefore can not be treated with anti allergic drugs. Trauma even minimal, and psychological stress can facilitate attacks. For the same reason operations in the mouth, such as dental surgery, endoscopic examinations, etc., can cause edema of the glottis.

The diagnosis of the disease is relatively simple and consists on the quantitative and functional determination of C1-INH.

The correct therapy, in case of an attack of the glottis, is the infusion of an adequate amount of C1-INH plasma concentrate, or the synthetic product Icatibant / acetate, an inhibitor of bradykinin available in Italy. Prevention of attacks, in adult people, is based on chronic administration of attenuated androgens. The administration of cortisone and antihistamines is useless.

BIBLIOGRAPHY : Agostoni A. - Cicardi M.
Medicine - Baltimore
71 :206-215, 1992

DOCTOR AVAILABLE IN CASE OF EMERGENCY:

In case of emergency for further medical information contact the following telephonenumber :

333/4004422

(active 24 hours a day)

In case of lack of BERINERT (C1-INH), call :

**CSL Behring S.p.A. P.le Stefano Turr, n. 5
20149 - MILANO - ITALY
tel. 02.34964.207 / 208 - fax 02.34964.264**

In case of lack of

**Firazyr® (Icatibant / acetate), call :
SHIRE ITALIA SpA - Piazza della Nunziata,
5/4 - 16124 - GENOVA - ITALY - Tel: +39
010 2758377 - Fax: +39 010 2745341**

The disease reveals itself with :

- subcutaneous edemas
- abdominal pain like colic caused by edema of gastroenteric mucosae
- dyspnea with asphyxiation caused by laryngeal edema
- symptoms last 1 to 3 day
- The correct therapy, in case of an attack of the glottis, is the infusion of an adequate amount of C1-INH plasma concentrate, or the synthetic product Icatibant / acetate, an inhibitor of bradykinin
- Failing this products, fresh frozen plasma can be administered.

CORTISONES AND ANTIHISTAMINICS ARE INEFFECTIVE.
