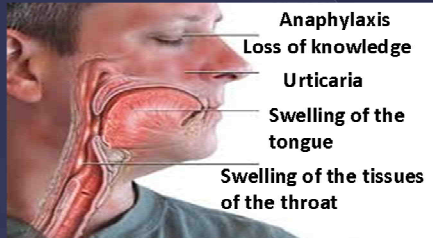


THE TREATMENT OF HEREDITARY ANGIOEDEMA: A REPORT OF TWO CASES

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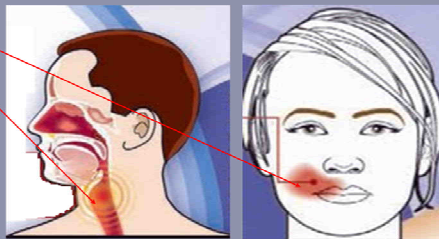
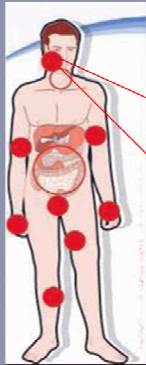
BACKGROUND:

Hereditary angioedema (HAE) type I is a rare genetic disorder, autosomal dominant, caused by a deficiency of the enzyme C1 inhibitor (C1INH).

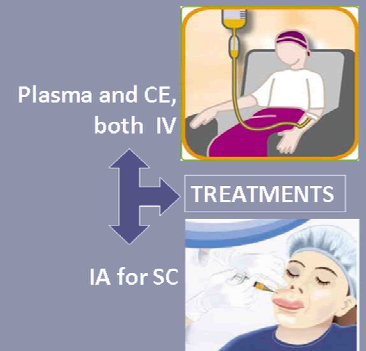
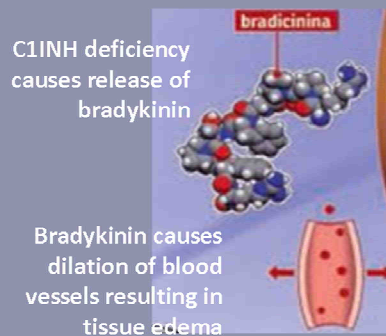


It is characterized by recurrent and unpredictable episodes of edema primarily affecting the extremities, face, throat and intestinal wall and may jeopardize the patient's life if not treated properly.

Available treatments for acute attacks are icatibant acetate (IA) for subcutaneous (SC), fresh plasma and C1INH esterase (CE) (foreign drug), both intravenously (IV).



Edema primarily affecting the extremities, face, throat and intestinal wall



OBJECTIVE:

To describe our experience in the treatment of acute episodes of HAE in two patients (brothers) from our health area.

METHODS:

Design: Retrospective observational study.

Period: January 2008-September 2011.

Data source: medical history, Sinfhos[®] software.

Data:

- age and sex
- prophylactic treatment
- number of crises
- accident and emergency (A&E) visits
- acute treatment
- efficiency, safety and cost.

RESULTS:

- Two siblings (male and female, 33 and 31 years respectively) with no prophylactic treatment for poorly controlled symptoms and poor tolerance.
- Between the two came to the A&E 19 times for presenting outbreaks with laryngeal edema, 11 were treated with CE, in 3 cases needed a second dose and in another one 4 doses.
- Already available in the hospital IA, the last 4 outbreaks were treated with a single dose of the same with a complete resolution of symptoms.
- There were no adverse effects with either drug.

CONCLUSIONS:

- In all episodes treated with IA, the outbreak was resolved with a single dose.
- The sc route of administration of the IA allows self-administration in crisis without the presence of laryngeal edema.
- The hematology safety profile, storage at room temperature and that it isn't a foreign drug represent additional advantages of IA against hematological derivatives.
- The average price of treatment with CE was € 1060.78 and with IA € 1692.29.