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Laryngeal edema is common in HAE and demands preventive measures

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Background
The aim of this study was to show the clinical characteristics and treatment of Hereditary Angioedema (HAE) in a tertiary center from Curitiba, south of Brazil.

Methods
A cross-sectional study reviewing records of patients in the Hospital de Clínicas, Federal University of Paraná. We analised the clinical characteristics, laboratory tests and treatment of patients with HAE.

Results
Forty two patients, male (45%), age mean 27.1±16.9 years. Symptoms started at median age 14 years (range 1 to 58ys.). Thirty seven (86%) had familial history of HAE. They had a median of 2 episodes/mo (range 0.2 to 30) lasting 3±1.7 days/episode. Edema of limbs, face, genital and laryngeal, abdominal pain, diarrhea and vomiting were seen in 73.8%, 52.4%, 23.8%, 17%, 81%, 24% and 38.1%, respectively. Twenty four (57%) had low C4 level, mean serum level of C4=12.7±10.6 mg/dL; 26 (62%) had low levels of C1 esterase inhibitor, 4 (9.5%) had low functional C1 esterase inhibitor and 12 (28.6%) had C1q deficiency and 3 (7.1%) had HAE type III confirmed. Fifteen (28.9%) were treated with Danazol 175±134 mg/day, 15 (28.9%) were using tranexamic acid as needed. No patient had Icatibant access.

Conclusions
HAE is a difficult to control disease. Patients needs to have a facilitated access to newer therapies to control their symptoms and reduce the adverse events of Danazol.

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