**Hereditary angioedema laryngeal attacks: report from the Czech national registry**

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BACKGROUND: Hereditary angioedema (HAE) is a rare genetic disorder caused by deficiency of C1 inhibitor (C1-INH) function. Laryngeal attacks are potentially life threatening, the patients are at risk for suffocation during the attack.

The goal of this study was to analyse laryngeal attacks from the Czech national registry of primary immunodeficiencies.

METHODS: We present data collected between March 2012 and December 2014. Data were collected in 4 centres intended for the diagnosis and treatment of HAE.

RESULTS: The data of 139 HAE patients (77 females, 62 males, 120 patients (86.3%) with type I HAE; 19 patients (13.7%) with type II HAE) were available.109 laryngeal attacks in 30 patients (19 females, 11 males; 24 patients (80%) with HAE type I patients, 6 patients (20%) with HAE type II) were recorded. The triggering factors described by the patients included stress in 8 (7.3%) attacks, infection in 7 (6.4%) attacks and injury in 2 (1.8%) attacks. However, in 80 attacks (73.4%), the triggering factor was not identified. 81(74.3%) of laryngeal attacks were treated with icatibant, 21 (19.3%) with recombinant C1-INH, 3 (2.7%) with plasma derived C1-INH, 1 (0.9%) with fresh frozen plasma, 2 (1.8%) by increase in androgens dose. Treatment had to be repeated in 11 attacks (10.1%). The drug was self-administered in 76 (69.7%) of the laryngeal attacks, 22 (20.2%) attacks was treated in the local hospital and only 3 cases (2.8%) were treated in HAE centre. Hospitalization in intensive care unit was necessary in 2 attacks (1.8%), Emergency medical service (EMS) was used in 2 attacks (1.8%).

CONCLUSIONS: Laryngeal oedema is the most serious manifestation of HAE. In our experience it comprises more than 10% of HAE attacks. The analysis of laryngeal attacks gives further insight into their course. To prevent fatal outcome, it is essential to instruct patient how to identify the first symptoms of laryngeal attack and how to use appropriate emergency procedures.