**Hereditary angioedema: Report from the Czech registry**

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BACKGROUND: Hereditary angioedema (HAE) is a rare genetic disorder caused by a deficiency in functional C1 inhibitor (C1-INH) activity. In patients with HAE, plasma proteolytic cascades normally regulated by C1-INH becomes activated, resulting in episodic, recurrent attacks of angioedema. These attacks are characterized by subcutaneous and submucosal oedema in various anatomical locations.

The goal of this study is to present the results of the acute HAE attacks treatment obtained 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 therapy of HAE.

RESULTS: The registry contains data of 139 HAE patients (women: 77, men: 62; HAE type I 86.3%, HAE type II 13.7%), showing HAE prevalence 1.32 per 100 000 inhabitants. 1084 attacks in 104 patients were recorded. The factors described by the patients as being associated with triggering HAE episodes included stress (10.4%), trauma (9.2%) and infection (3.6%). However, in most attacks triggering factor was not identified (68.8%). The most frequent were abdominal (47.7 %) followed by peripheral oedema (38.5%). Laryngeal oedema was presented in 10.1% of attacks. Prodromal symptoms (most often erythema marginatum, weakness or nausea) were reported by 13.1% of patients. 874 attacks (80.6 %) were actively treated (64.2% icatibant, 22.9% recombinant C1-INH, 2.6% plasma derivate C1-INH, 0.3% fresh frozen plasma, 6.8%increase in androgens dosage, 3.1% increase in tranexamic acid dosage). Treatment had to be repeated in 102 attacks (11.7%). Hospitalization was necessary in 7 attacks (0.9%), Emergency medical service (EMS) was used in 5 attacks (0.6%).

CONCLUSIONS: Our results show marked clinical variability in HAE patients. The fact that in more than 10% of patients required repeated treatment of single attack shows that although various therapeutic approaches are available, it is still difficult to choose the best therapeutic approach for a concrete patient.