P-2
Hereditary angioedema: Report from the Czech registry

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Background: Hereditary angioedema (HAE) is a rare, autosomal dominant disorder characterized by recurrent attacks of subcutaneous or sub-mucosal oedema. Symptoms are extremely variable in frequency, localization and severity. Laryngeal attacks are potentially life threatening, the patients are at risk of suffocation during the attack.

Methods: The goal of this study was to analyse HAE attacks in the Czech Republic (CR) between March 2012 to December 2016. Data were collected from the Czech National Registry of Primary Immunodeficiencies.

Results: The registry contains data of 150 HAE patients (females 81, males: 69; HAE type I 86.7%, HAE type II 13.3%), showing HAE prevalence 1.42 per 100 000 inhabitants. 2515 attacks in 119 (females: 65, males: 54; HAE type I 86.6%, HAE type II 13.4%) patients were recorded. The potential triggering factors for HAE attacks included stress (9.2%), trauma (7.1%) and infection (4.7%). However, in most attacks triggering factor was not identified (71.9%). The most frequent were abdominal attacks (54.7 %) followed by peripheral oedema (33.8%). Laryngeal oedema was presented in 10.6 % of attacks. 20.9 % attacks were combined. Prodromal symptoms (most often erythema marginatum, weakness or nausea) were reported by 13.6% of attacks. 2060 attacks (81.9 %) were actively treated (65.8% icatibant, 21.5% recombinant C1-INH, 5.9% plasma derived, highly purified, nanofiltered C1-inhibitor (pnfC1-INH), 0.5% plasma derived, nanofiltered pdC1-INH (nfC1-INH), 0.1% fresh frozen plasma, 4.2% increase in androgens dosage, 1.8% increase in tranexamic acid dosage). Treatment had to be repeated in 312 attacks (15.1%). Hospitalization was necessary in 25 attacks (1.2 %). Emergency medical service (EMS) was used in 11 attacks (0.5%).

Conclusions: The analysis of HAE attacks gives further insight into their course. Our results show marked clinical variability in HAE patients. The fact that in more than 15% of attacks 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.