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Hereditary angioedema

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1. Short Communication

In September, HAEi South Eastern Europe Workshop was held in the Republic of Macedonia, which was attended by experts from the US HAEA Angioedema Center at UC San Diego, patient organizations and patient group from Bulgaria, Macedonia, Montenegro, Romania, Serbia and Turkey. Bruce Zurraw and Sandra Christiansen talked about HAE fundamentals and currently available treatments. HAEi SEE countries regional patient advocate talked about patients' mental wellbeing and advocating for quality of life.

Hereditary angioedema is a genetic disease caused by a structural and functional disadvantage of C1 inhibitors, glycoproteins that inhibit numerous plasma proteases that are the subcomponent of the first fraction of the components (C1r, C1s), kallikrein, plasmin, and XI and XII coagulation factor. Deficiency of C1 inhibitors determines edematous subcutaneous injuries, of non-inflammatory character, with recursive movements [1, 2].

The disease can be rarely hereditary or more often disease has gain character. The present form can be caused by local anesthetic, drugs, food, infections, emotional stress, etc. The hereditary form is an inherited autosomal dominant disease [3, 4]. In the great percentage the angioedema is with idiopathic nature, without a known trigger.

The localization of the change may be on the oral mucosa (lips, tongue, soft palate, buccal mucosa), skin, face, hands, palms, legs, genitals and gastrointestinal and respiratory system [5, 6]. The clinical picture is characterized by the acute occurrence of a painless solitary or multiple swelling of soft tissues, with a size of several centimeters. The swelling is smooth and shiny and is withdrawn for 24-72 hours. Swelling in the throat is the most dangerous symptom. The edema of the epiglottis (Quincke's *edema*) is a serious complication because it can lead to suffocation and death. Therapy consists of an urgent application of antihistamines and systemic administration of corticosteroids. In more severe cases, intramuscular administration of epinephrine is required [7].

Dental treatment. The hereditary missing of a C1 inhibitor is a very rare disease and the performance of any dental intervention, without prior preparation (infusion of a inhibitor C1), can be very risky [8]. Due to the possibility of occurrence of Quincke's *edema*, dental care requires a complex approach through mandatory team work (dentist, pediatrician, internist, anesthesiologist).

After compulsory general medical preparation, all dental interventions should be performed using anesthesia (local or general), and in the presence of the specialist doctor who leads the patient. After

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the intervention is completed, it is necessary to observe the patient for at least another hour. Dental interventions in these patients should be undertaken only in dental clinics [9].

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