Vesna Grivcheva-Panovska, J Clin Exp Dermatol Res 2016, 7:4(Suppl)
http://dx.doi.org/10.4172/2155-9554.C1.029

6th International Conference on
Clinical and Experimental Dermatology
May 04-06, 2016 Chicago, USA

Innovative therapeutic approach of hereditary angioedema

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Hereditary angioedema (HAE) is a rare genetic, life-threatening disease. Patients affected by HAE most of the time are without visible signs of the disease, besides the attacks, which can be brutal, painful and lethal. Several therapeutic approaches have been developed and used with various success, recombinant human C1INH (rhC1INH) being the most innovative current approach. Recombinant human C1INH is a purified derivative from rabbit milk which expresses the gene that encodes the synthesis of C1INH. The amino acid sequence of the recombinant form is identical to the human C1INH. C1INH is single-chain plasma glycoprotein with a molecular mass of 73,650 belonging to the super-family of serine protease inhibitors in plasma. C1INH is the only known inhibitor of activated plasma subcomponents C1s and C1r of the complementary component 1 of the complementary cascade classical pathway. Furthermore, C1INH inhibits the Manan-associated serine protease 2 (MASP2) of the lectin pathway of the complement. Additionally, it is the main inhibitor of the activated factor XII, factor XI and kallikrein of the plasma contact system. The rhC1INH was primarily developed for treatment of acute angioedema attacks in patients with HAE due to C1INH activity deficiency. The rhC1INH inhibitory potential of target proteases C1s, kallikrein, factor XIa and factor XIIa is highly comparable to the endogenous human C1 esterase inhibitory potential *in vitro*. Additional data of the efficacy are obtained by multiple analysis of primary efficacy sensitivity end point as well as the results of secondary and explorative efficacy.

Biography

Vesna Grivcheva-Panovska has received her Tenured Professorship in Dermatology in 2013 at University Saints Cyril and Methodius in Skopje, Macedonia. She is the Head of the Unit of Allergy and Clinical Immunology at the University Clinic of Dermatology and she is in-charge of Clinical Diagnostics. She has been working in the field of HAE since 1984, publishing numerous papers in international journals and took active participation in six international scientific projects.

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