

Annual Congress on

# Rare Diseases & Orphan Drugs

October 26-27, 2016 Chicago, USA

## Evaluation of adherence to treatment in patients with hereditary angioedema due to C1 inhibitor deficiency: A comparison of three alternative strategies

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**Introduction:** Hereditary Angioedema with C1 Inhibitor Deficiency (C1-INH-HAE) is a rare autosomal genetic disorder presenting with recurrent episodes of swelling of the skin, gastrointestinal tract and upper airways that resolve within 48 to 96 hours. Multiple therapeutic options are available to treat acute attacks of C1-INH-HAE: Plasma derived C1-inhibitor (pdC1-INH), recombinant C1-inhibitor (rhC1-INH) and antagonist of bradykinin B2 receptor (icatibant). The administration route is a subcutaneous injection for icatibant, while pdC1-INH and rhC1-INH are administered endovenously. International guidelines suggest to treat all the attacks regardless of location and severity and to administer therapy as soon as possible. Moreover experts support the use of home therapy that appears to be a viable and effective strategy with a favorable impact on costs. We hypothesized that the use of different drugs and the self-administration strategy may influence the adherence to therapy.

**Aims:** The aim of this study is to evaluate and compare the compliance to the different therapeutic strategies used in our center.

**Methods:** 62 of 68 C1-INH-HAE patients followed in our Department were enrolled in this study (6 patients dropped out). The patients were divided in three groups: Group A: 25 patients (14 F, 11 M; median age 32 y; age range 8-76) using home therapy with pdC1-INH; Group B: 12 patients (8 F, 4 M; median age 37 y; age range 17-56) self-administering icatibant; Group C: 19 patients (12 F, 7 M; median age 41 y; age range 5-87) treated with pdC1-INH in healthcare setting. The patients were interviewed monthly from August to January 2015. The interviews allowed the collecting of data about the total number of attacks, the number of treated attacks, as well as the time from symptom onset to drug administration. Compliance was evaluated by means of the percentage of treated attacks and the time from symptom onset to treatment administration.

**Results:** During the observation period there were 918 attacks, of which 544 were treated (59.2%). The analysis reveals that the number of attacks of angioedema of the patients that chose self-administration strategy (Groups A and B) was significantly higher than the patients that performed health professional strategy (Group C) ( $p=0.0019$ ). Moreover, adherence to therapy of the patients of group A and B was significantly higher than the patients of group C ( $p=0.003$ ). Notably, in the home therapy group, the compliance was higher in patients that administered pdC1-INH (71.2%) than the patients using icatibant (44%). These data show a significantly better adherence to therapy in patients that choose self-administration than those performing health professional strategies, both for the percentage of treated attacks and for the time to infusion.

**Conclusion:** In conclusion home therapy improves the efficiency of treatment of C1-INH-HAE, and in our opinion this strategy has to be implemented in all centers treating C1-INH-HAE patients.

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