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Case Report

Background: Hereditary angioedema (HAE) is a rare and lifethreatening disorder characterised by attacks of angioedema due to deficient or dysfunctional C1-esterase inhibitor (C1-INH). Subcutaneous (SC) replacement of C1-INH with a plasma-derived equivalent is an approved therapy for patients with HAE. We report the efficacy and tolerability of C1-INH (SC) in one of the first patients with HAE in Germany to be treated throughout pregnancy and breastfeeding.

Methods and Results: The patient is a 29-year-old Hungarian female living in Germany who first experienced symptoms aged 14 years after starting hormone therapy due to acne. She experienced 4–6 attacks/year, typically lasting 5 days, affecting the hands, feet, face and stomach/intestines; attacks were mostly triggered by infections. The patient was treated with increasing doses of oral antihistamines and intravenous (IV) cortisone without response. In December 2018, she was diagnosed with HAE due to her family history and symptoms at the University Medical Center Mainz, and prescribed icatibant.

In August 2019, she was admitted to hospital for 3 days due to an acute facial attack resulting from otitis. As the patient was 2 months pregnant, she was treated with 1500 IU C1-INH (IV). Post hospitalisation, she was initially prescribed 2×1000 IU/week C1-INH (IV). However, in September 2019, due to an attack frequency during pregnancy of >1/month plus unsuccessful assisted or selfadministration attempts at home, an inability to drive to the medical centre and a language barrier, she was prescribed 2×3000 IU/week C1-INH (SC).

After starting C1-INH (SC) therapy, she only experienced two mild attacks: one in her hand on her wedding day and one suspected attack in her abdomen from her baby kicking. In March 2020, she gave birth to a healthy baby by caesarean section. She received a dose of C1-INH (SC) the day prior and no additional prophylaxis was required for the surgery. She continued to receive 2×3000 IU/week C1-INH (SC) while breastfeeding. In October 2020, her menstrual cycle resumed and she experienced an attack requiring 1000 IU C1-INH (IV). Since then, she has not experienced any further attacks and is satisfied that her HAE is well controlled. **Conclusion:** C1-INH (SC) therapy effectively controlled attacks in a patient with HAE throughout pregnancy and breastfeeding. Treatment with C1-INH (SC) was not associated with any adverse events for the mother or baby. Written informed consent was provided.

382 | Methodological perspectives for validating the involvement of paracetamol in the development of allergic diseases.

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Background: We suspect that paracetamol contributes to the development of allergic diseases. This suspicion is based on a set of observations : a) publications showing that taking paracetamol in young children would lead to the future onset of an allergic disease ; b) the communication of a personal case which involves the molecule in the development of a food allergy in adult ; c) the parallelism between the growth in paracetamol worldwide sales and the growth of allergic diseases, particularly food allergies.

Method: As its consumption has become so commonplace, it is difficult to confirm this suspicion. Here are four methodological approaches: (1) Prepare a retrospective drug questionnaire for each allergic patient; (2) A long-term prospective study of two cohorts: paracetamol authorised or strictly forbidden; (3) The development of allergic pathologies once paracetamol use is discontinued ; (4) Compare the development of paracetamol consumption and the allergy prevalence between countries.

Results: (1) would be unreliable. (2) is not feasible, for practical and ethical reasons. However (2) could be simplified by the simple observation of individuals presenting a bad reaction to the molecule; however the problem is that they are rare. (3) and (4) seem to be the most realistic ones. For (4) it would be interesting to resume the study of E. Von Mutius by focusing on the evolutionary differences of the paracetamol consumption between East and West Germany. In any case, studies are expected to be multi-centric and international, requiring cooperation between several hospital research units. An call is therefore launched.

Conclusion: There is an urgent need to find an explanation for the worldwide increase of allergic diseases. It is not enough to rely on the hygienic hypothesis. Nor on the « pollution » argument. Could it be that, in the same way as in the E. Poe's short story " The Purloined Letter ", paracetamol escapes suspicion because of its high profile?