Positive Response to 5HT-2 Antagonists in a Family Affected by Epidermolysis bullosa Dowling-Meara Type

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A young woman, aged 32, and her child, 8 months old, were referred to our Department in October 1990. At this time the mother had a few bullae limited to her face and hands, while the baby suffered from a severe eruption of serohemorrhagic bullous lesions on his face, palms, soles, trunk and in the diaper area, distributed in a herpetiform manner and causing severe discomfort. This eruption was first seen immediately after birth, with a period of remission in springtime and a relapse in September 1990. Cutaneous biopsies obtained from lesional areas of the forearm revealed intraepidermal bullae. Ultrastructural evaluation (TEM) showed cytolysis and clumping of the tonofilaments in the basal keratinocytes. Based on clinical characteristics and TEM findings, the diagnosis of epidermolysis bullosa simplex herpeti-formis Dowling-Meara type (EBS-DM) was made [1]. Pushed by the severity of the child's disease, we began the treatment with pipamperone (5-hydroxy-tryptamine-2 antagonist), a neuroleptic drug of the buthriophenone family, which was demonstrated to be useful in the treatment of EBS-DM as reported by Bonnetblanc and Bouquier [2]. We began at the dose of 10 mg/day for the child and 80 mg/day for the mother. After an initial worsening, the bullae dramatically decreased in severity and number in both patients in a period of 2-3 weeks. A complete clearing of the lesions was obtained at the doses of 20 and 120 mg/day respectively. During the 2 months of treatment, the mother began to suffer from initial 'drowsiness', while the child progressively showed torpidity and laziness with a slowed neuropsychomotor development compared to his age standards. Two months' withdrawal from pipamperone resulted in a progressive reappearing of the bullous lesions at trauma sites (hands, feet, groins). The clinical picture 'before therapy' was restored, and the developmental parameters assessed by the pediatricians returned to normal stage.

In order to avoid the heavy side effects of pipamperone, we tried to further assess another 5HT-2 antagonist, such as cyproheptadine, known to be devoid of important side effects in pediatric age [3]. In April 1991, we introduced cyproheptadine at the dose of 4 mg/day for the child and 8 mg/day for the mother. Clinical improvement was evident, and it was appreciated by the patients. Compared with pipamperone, the improvement was less dramatic, and we observed a longer period of induction. No side effects were referred in the successive follow-up.

Looking at this data, we can state that 5HT-2 antagonists really seem to be effective in the treatment of this bullous disease in a dose-dependent way. We confirm the efficacy of pipamperone in the treatment of EBS-DM. Moreover, cyproheptadine is an effective and useful drug, with incomparably less severe side effects than pipamperone. Though the role of serotonin receptor-mediated processes in the pathogenesis of EBS-DM and the ability of its antagonists to eventually restore the integrity of the tonofilaments in the basal layer keratinocytes remains unknown [4], we suggest the use of 5HT-2 antagonists in the treatment of EBS-DM, especially for those patients affected by severe forms of this disease.

References

Blanchet-Bardon C, Nazzaro V, Raynaud F, Hubert S, Mimos C: Epidermolyse bulleuse dominante de Dowling-Méara. Une epidermolyse bulleuse intraépidermique qui cache bien son pronostic. *Ann Dermatol Vénéréol* 1987; 114:341-348.

Bonnetblanc JM, Bouquier JJ: Response to pipamperone in case of epidermolysis bullosa herpetiformis. *Lancet* 1986;1327-1328.

Klein GL, Galant SP: A comparison of the antipruritic efficacy of hydroxyzine and cyproheptadine in children with atopic dermatitis. *Ann Allergy* 1980;44:142-145.

Hoffman M: A layer by layer look at skin blister disease. *Science* 1991;254:1111-1112.