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Response

B. Montegudo, M. Cabanillas, C. de las Heras, and J.M. Cacharrón

Servicio de Dermatología, Complejo Hospitalario Arquitecto Marcide-Novoa Santos, Ferrol, La Coruña, Spain

To the Editor:

We thank Professor Camacho and Professor Moreno for their comments on our article. In fact, in line with articles of a similar sort,^{1,2} and even those by these authors themselves,³ ours did not aim to be a systematic review of the different varieties of congenital localized hypertrichosis (as was stated in the article).⁴ Hence we had simplified the classification to four of the primary localized symmetrical clinical forms described in the specialist literature: cubital hypertrichosis, anterior cervical hypertrichosis, posterior cervical hypertrichosis and hypertrichosis of the lumbosacral area⁵ (identical to the division made by Vashi et al⁶ in the introduction to their article, not only on analysis of their series, as is suggested). This is inevitably involved leaving other conditions aside,⁷⁻⁹ including those covered by professors Camacho and Moreno in other publications.^{10,11} This list could also include hypertrichosis associated to congenital smooth muscle hamartoma,¹²⁻¹⁵ tufted angioma,^{12,13,16} congenital plaque-like glomangioma,¹⁷ the hair collar sign (that can surround aplasia cutis congenita, be a skin marker for cervical spinal dysraphism, or present with no associated malformations),^{5,13,18} ectopic eyelashes,¹² some syndromes with congenital localized hypertrichosis¹⁹ such as H syndrome,²⁰ pigmentary mosaicism of the Ito type,^{9,14,21} (the “twin spotting” phenomenon)²¹ and segmental odontomaxillary dysplasia,²² and even distichiasis,¹² or “physiological” processes such as scrotal skin^{12,13}—a benign, self-limiting process with no associated endocrinological abnormalities that occurs in the first few

months of life^{23,24}—it is not apparent at birth and resolves spontaneously in a manner similar to many cases of cubital hypertrichosis.²⁵

Anterior cervical hypertrichosis is a localized congenital variety of hypertrichosis that we have been able to see in 2 patients over a period of 3 months.^{4,26} As a result of our publication other members of the faculty have shared a further 2 sporadic cases with us, 1 of them associated with neurological abnormalities; this fact indicates that the syndrome is not as uncommon as previously thought, and that the small number of cases described in the literature could be due to underdiagnosis or the tendency to publish only familial cases or those with associated abnormalities.

It is true that, in 1989, Reed et al published a case of familial cervical hypertrichosis associated with scoliosis; all their patients presented increased hair growth on the posterior aspect of the neck and were therefore included in the posterior cervical hypertrichosis group. Those authors considered this to be the first familial case of posterior cervical hypertrichosis of autosomal dominant inheritance associated with scoliosis, but they indicated in the discussion that they had previously encountered cases of this form of hypertrichosis.²⁷

In most publications in English,³ including several reviews,^{1,29-31} Trattner et al²⁸ are cited as the first to describe ACH. We consider it correct to point out that there are earlier descriptions of the syndrome and of this particular rare variety in the non-English literature,³² which is more difficult to access, and we are pleased to find that the author of the first article on this subject was in fact

a dermatologist and member of the Spanish Academy of Dermatology and Venereology (AEDV).

The comments provided by professors Camacho and Moreno, serve not only to enrich our article, but also to highlight the issue of ACH.

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