

LETTERS TO THE EDITOR

Sporadic Anterior Cervical Hypertrichosis

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In issue one of the Centenary of our journal *Actas Dermo-Sifiliográficas* a case report appeared authored by Monteagudo et al¹ on “Isolated anterior cervical hypertrichosis” upon which we would like to make some comments.

Firstly, the classification of “localized familial hypertrichosis” (Table) is not entirely rigorous as this is a reference to 4 forms of localized hypertrichosis encountered in 11 patients by Vashi et al.² There are many classifications available in dermatology and trichology books that group localized congenital hypertrichosis

into several groups, one of which is “nevoid localized hypertrichosis”—a group that includes anterior cervical hypertrichosis (ACH). There is no doubt that simpler classifications can be used to ease the strain on those new to hypertrichosis, classifications such as those we used ourselves when we wrote the chapter on hypertrichosis for the Spanish Academy of Dermatology and Venereology (AEDV) *Clínicas Dermatológicas*.³ The version we used was taken from Olsen,⁴ a current authority on trichology and expert on hypertrichosis. When he wrote on this subject in the 2008 edition of “Fitzpatrick’s Dermatology in General Medicine” with Paus and Messenger,⁵ he broadened the category of nevoid hypertrichosis to include hypertrichosis of the eyebrows and auricles of the ears. Even so, consideration is still needed of “polythelia pilosa,” “hypertrichosis of the tip of the nose” and “eyelash trichomegaly,” as one of the present authors, Dr Camacho, noted in *Monografías de Dermatología*,^{6,7} in books on trichology by Camacho and Montagna⁸ and Blume-Peytavi et al,⁹ and in the chapter on “Hypertrichosis and hirsutism” in “Dermatology” by Bologna et al.^{10,11}

Leaving the issue of classification aside, we would like to comment on the case of the 27-year-old patient with ACH. The diagnosis is acceptable considering the image provided, and—as the condition is neither familial nor associated to any other abnormality—it is reasonable to assume this is sporadic nonsyndromal ACH. We also agree with the authors that this condition is possibly under diagnosed, and must be recognized to exclude associations and avoid requests for unnecessary studies that entail high personal and social costs.

But we do not agree with the authors that Trattner et al described ACH in 1991.¹² What Trattner et al actually discovered was the association of ACH with peripheral sensorimotor neuropathy; although the confusion of Monteagudo et al¹ is justified given that Trattner et al stated in the discussion section that “they had found no publications of sporadic or familial cases of ACH.”

“Goat’s beard hypertrichosis,” as it is also known, is characterized by the appearance of a small area of terminal hair just above the laryngeal prominence. This can be a sporadic finding, as in the case of Monteagudo et al,¹ or it can be the marker of a peripheral neuropathy, hallux valgus, thalassemia minor, or ocular albinism.¹³ Although a hereditary pattern has not been established, it seems

Table. Congenital circumscribed hypertrichosis

Congenital pigmented nevi
Becker melanosis
Nevoid hypertrichosis
Primary nevoid hypertrichosis
Cubital hypertrichosis (hairy elbow syndrome)
Hypertrichosis of the eyebrows
Hypertrichosis of the external ear
Hypertrichosis of the tip of the nose
Eyelash trichomegaly
Polythelia pilosa
Hairy cutaneous malformations of palms and soles
Anterior cervical hypertrichosis
Posterior cervical hypertrichosis
Secondary nevoid hypertrichosis
Hypertrichosis associated with neurofibroma
Hemihypertrophy associated with muscular problems
Associated to the underlying hemodynamic system
Hypertrichosis with lipodystrophy
Hypertrichosis with scoliosis
Spinal dysraphism

Adapted from references: 3-11.

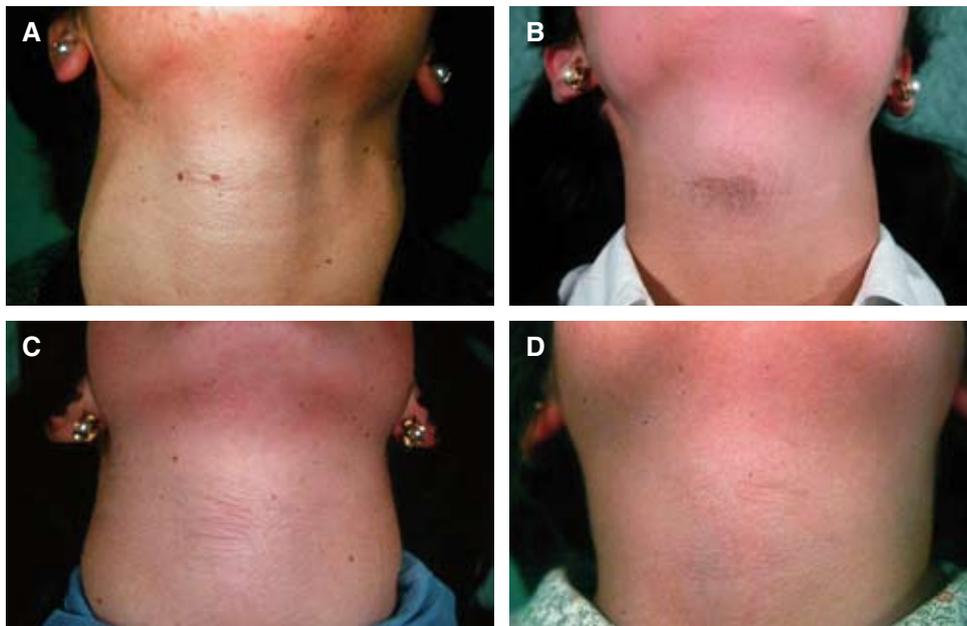


Figure 1. Four cases of familial anterior cervical hypertrichosis. A, Woman aged 42, mother of the other three patients. B, Woman of 18. C, Girl of 16. D, Girl of 11.

that sporadic familial ACH has autosomal dominant transmission or is X-linked dominant,¹⁴ while recessive forms are associated with neuropathies.

We accept that when writing an article authors do not generally refer to textbooks or encyclopedias, but in the article in *Monografías de Dermatología*⁷ and in the chapter on hypertrichosis in the AEDV *Clínicas Dermatológicas*³ it states the following regarding ACH: “we have decided to consider the this form of hypertrichosis, which we described many years ago,¹⁵ within the category of nevoid hypertrichosis, because although we then related it with underlying pigmentary cellular nevi—although this was not proven by biopsy as the patients did not agree to the procedure—we did not consider the reduced level of touch and pain sensation detected in a woman and three of her daughters to be important.” This reference to the description provided by one of us reads: “Congenital nevoid hypertrichosis. Represented by pigmentary cellular nevi, Becker nevus, faun tail nevus associated with spina bifida, hairy elbow syndrome, hypertrichosis of the outer ear or eyebrows, and other less common familial forms, with tufts of ectopic hair possibly related to a nevus (Figures 7.2–7.5).” In the figure legend to which we refer it said: “Familial tuft of hair in the anterior cervical zone. All those affected are women.” In fact, the woman’s 2 sons showed no signs of ACH. And even if Trattner et al¹² did not read this book, in 1989 Reed et al¹⁶ published an article on familial cervical hypertrichosis associated with scoliosis and also claimed to have discovered this syndrome. More recently, Tsukahara and Hajji¹⁴ and Lee et al¹⁷ have published other cases of familial ACH.

In any case, we are grateful to Dr Monteagudo et al for their publication, as this is clearly the first sporadic case in Spain, and it has also given us the opportunity to update the classification of localized hypertrichosis and to once more present the case of the 4 women from Granada, a mother (Figure 1 A) and her three daughters (Figures 1B, 1C and 1D), who all had familial ACH.

References

1. Monteagudo B, Cabanillas M, de las Heras C, Cacharrón JM. Hipertrichosis cervical anterior esporádica. *Actas Dermosifiliogr.* 2009;100:61-4.
2. Vashi RA, Manzini AJ, Paller AS. Primary generalized and localized hypertrichosis in children. *Arch Dermatol.* 2001;137:877-84.
3. Moreno-Giménez JC, Camacho FM. Hipertrichosis. In: Camacho FM, ed. *Enfermedades de los anejos. Clínicas Dermatológicas de la Academia Española de Dermatología y Venereología. Educación Médica Continuada. Vol. 3, núm. 1.* Madrid: PRODUG; 2006. p. 11-28.
4. Olsen EA. Hypertrichosis. In: Olsen EA, ed. *Disorders of hair growth: diagnosis and treatment.* New York: McGraw-Hill; 1993. p. 315-36.
5. Paus R, Olsen EA, Messenger AG. Hair growth disorders. In: Wolff K, Goldsmith LA, Katz SI, Gilchrist BA, Paller AS, Leffell DJ, eds. *Fitzpatrick’s Dermatology in General Medicine.* 7th ed. New York: McGraw-Hill; 2008. p. 753-77.
6. Pérez Bernal AM, Camacho F. Hipertrichosis. Concepto y clasificación general. *Monogr Dermatol.* 1992;5:315-27.
7. Camacho F, Rodríguez-Pichardo A, García-Bravo B. Hipertrichosis congénitas circunscritas. *Monogr Dermatol.* 1992;5:334-41.

8. Camacho F. Hipertrichosis. In: Camacho F, Montagna W, eds. *Tricología. Enfermedades del folículo pilosebáceo*. Madrid: Aula Médica; 1995. p. 123-38.
9. Camacho FM. Hypertrichosis. In: Blume-Peytavi U, Tosti A, Whiting DA, Trüeb RM, eds. *Hair growth and disorders*. Berlin: Springer Verlag; 2008. p. 333-56.
10. Camacho F. Hypertrichosis and hirsutism. In: Bologna JL, Jorizzo JL, Rapini RP, Horn TD, Mascaró JM, Mancini AJ, et al, eds. *Dermatology*. London: Mosby; 2003. p. 1051-9.
11. Camacho-Martínez FM. Hypertrichosis and hirsutism. In: Bologna JL, Jorizzo JL, Rapini RP, Callen JP, Horn TD, Mancini AJ, et al, eds. *Dermatology*. 2nd ed. London: Mosby Elsevier; 2008. p. 1007-18.
12. Trattner A, Hodak E, Sagie-Lerman T, David M, Nitzan M, Garty BZ. Familial congenital anterior cervical hypertrichosis associated with peripheral sensory and motor neuropathy – A new syndrome? *J Am Acad Dermatol*. 1991;25:767-70.
13. Braddock SR, Jones KL, Bird LM, Villegas I, Jones MC. Anterior cervical hypertrichosis: a dominantly inherited isolated defect. *Am J Med Genet*. 1995;55:498-9.
14. Tsukahara M, Kajii T. Hairy throat: a dominant trait affecting seen members of a family. *Clin Dysmorphol*. 1992;1:165-7.
15. Camacho F. Hipertrichosis e hirsutismo. In: Camacho F, Montagna W, eds. *Tricología. Tricology. Trichologie*. Madrid: Garsi; 1982. p. 123-38.
16. Reed OM, Mellette JR, Fitzpatrick JE. Familial cervical hypertrichosis with underlying kyphoscoliosis. *J Am Acad Dermatol*. 1989;20:1069-72.
17. Lee HW, Lee MW, Choi JH, Moon KC, Kob JK. Familial anterior cervical hypertrichosis. *J Am Acad Dermatol*. 2005;53:530-2.

Response

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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.

References

1. Heitink MV, Quaedvlieg PJ, van Neer FJ, Frank J. Sporadic nonsyndromal anterior cervical hypertrichosis: case report and review of the literature. *Int J Dermatol.* 2007;46 Suppl 3:9-12.
2. Lee HW, Lee MW, Choi JH, Moon KC, Koh JK. Familial anterior cervical hypertrichosis. *J Am Acad Dermatol.* 2005;53:530-2.
3. García-Hernández MJ, Ortega-Resinas M, Camacho FM. Primary multifocal localized hypertrichosis. *Eur J Dermatol.* 2001;11:35-7.
4. Monteagudo B, Cabanillas M, de las Heras C, Cacharrón JM. Hipertrichosis cervical anterior esporádica. *Actas Dermosifiliogr.* 2009;100:61-4.
5. Trüeb RM. Causes and management of hypertrichosis. *Am J Clin Dermatol.* 2002;3:617-27.
6. Vashi RA, Manzini AJ, Paller AS. Primary generalized and localized hypertrichosis in children. *Arch Dermatol.* 2001;137:877-84.
7. León-Muiños E, Monteagudo B, Carballeira I, Corrales A. Nevo de Becker en la cara y el cuello. *Piel.* 2009;24:164.
8. Monteagudo B, Labandeira J, Peteiro C, de las Heras C, Cacharrón JM. Nevo melanocítico congénito gigante asociado a angioliopomas. *Med Cutan Iber Lat Am.* 2009;37:55-7.
9. Monteagudo Sánchez B, León Muiños E, Cabanillas González M, Martínez Meléndez G. Hipertrichosis localizada y mosaicismos pigmentario tipo Ito (hipomelanosis de Ito). *An Pediatr (Barc).* 2008;68:307-9.
10. Camacho FM. Hypertrichosis. In: Blume-Peytavi U, Tosti A, Whiting DA, Trüeb RM, editors. *Hair growth and disorders.* Berlin: Springer Verlag; 2008. p. 333-56.
11. Camacho-Martínez FM. Hypertrichosis and hirsutism. In: Bologna JL, Jorizzo JL, Rapini RP, Callen JP, Horn TD, Mancini AJ, et al, editors. *Dermatology.* 2nd ed. London: Mosby Elsevier; 2008. p. 1007-18.
12. Rogers M, Wong LC. Hair disorders. In: Eichenfield LF, Frieden IJ, Esterly NB, editors. *Neonatal Dermatology.* 2nd ed. Philadelphia: Elsevier; 2008. p. 517-35.
13. Rogers M, Tay YW. Hair disorders. In: Schachner LA, Hansen RC, editors. *Pediatric Dermatology.* 3rd ed. Edinburgh: Mosby; 2003. p. 525-59.
14. Wendelin DS, Pope DN, Mallory SB. Hypertrichosis. *J Am Acad Dermatol.* 2003;48:161-79.
15. Holland KE, Galbraith SS. Generalized congenital smooth muscle hamartoma presenting with hypertrichosis, excess skin folds, and follicular dimpling. *Pediatr Dermatol.* 2008;25:236-9.
16. Herron MD, Coffin CM, Vanderhooft SL. Tufted angiomas: variability of the clinical morphology. *Pediatr Dermatol.* 2002;19:394-401.
17. Rodríguez-Martín M, Sánchez R, Sáez-Rodríguez M, García-Bustinduy M, Sidro M, Pérez N, et al. Congenital plaque-like glomangioma associated with superficial hypertrichosis. *J Am Acad Dermatol.* 2008;58 5 Suppl 1:S92-3.
18. Roche-Gamón E, Febrer-Bosch I, Alegre de Miguel V. Signo del collar de pelo asociado a aplasia cutis congénita del cuero cabelludo. *Actas Dermosifiliogr.* 2007;98:442-3.
19. García-Cruz D, Figuera LE, Cantu JM. Inherited hypertrichoses. *Clin Genet.* 2002;61:321-9.
20. Molho-Pessach V, Agha Z, Aamar S, Glaser B, Doviner V, Hiller N, et al. The H syndrome: a genodermatosis characterized by indurated, hyperpigmented, and hypertrichotic skin with systemic manifestations. *J Am Acad Dermatol.* 2008;59:79-85.
21. López-Barrantes O, Torreló A, Mediero IG, Zambrano A, Happle R. Nevoid hypertrichosis and hypomelanosis. *Eur J Dermatol.* 2002;12:583-5.
22. Koenig LJ, Lynch DP, Yancey KB. Segmental odontomaxillary dysplasia presenting with facial hypertrichosis, commissural lip clefting, and hyperlinear palms. *Pediatr Dermatol.* 2008;25:491-2.
23. Leung AKC, Hedge HR, Stephure DK. Scrotal hair in identical twin infants. *Int J Dermatol.* 2005;44:1042-4.
24. Sentchordi Montane L, Quintanar Rioja A, Ayala Bernardo de Quirós L, Martínez Granero MA, Bonet Serra B. Vello escrotal en lactantes. *An Pediatr (Barc).* 2008;68:146-8.
25. Yuste-Chaves M, Zafra-Cobo MI, Martínez de Salinas A, Bravo-Piris J. Hipertrichosis cubital. Descripción de dos casos. *Actas Dermosifiliogr.* 2007;98:719-21.
26. Monteagudo Sánchez B, León Muiños E, Cabanillas González M, Martínez Meléndez G. Hipertrichosis cervical anterior. *An Pediatr (Barc).* 2008;69:591-2.
27. Reed OM, Mellette JR Jr, Fitzpatrick JE. Familial cervical hypertrichosis with underlying kyphoscoliosis. *J Am Acad Dermatol.* 1989;20:1069-72.
28. Trattner A, Hodak E, Sagie-Lerman T, David M, Nitzan M, Garty BZ. Familial congenital anterior cervical hypertrichosis associated with peripheral sensory and motor neuropathy— a new syndrome? *J Am Acad Dermatol.* 1991;25:767-70.
29. Corona-Rivera JR, González-Abarca S, Hernández-Rocha J, García-Cruz D, Corona-Rivera A. Mental retardation in a boy with anterior cervical hypertrichosis. *Am J Med Genet A.* 2005;135:69-71.
30. Thienpont B, Vermeesch J, Devriendt K. Anterior cervical hypertrichosis and mental retardation. *Clin Dysmorphol.* 2006;15:189-90.
31. Nanda A, Al-Arabi I, Ali MT, Alsaleh QA. Anterior cervical hypertrichosis (hairy throat): is it a sign to worry about? *Clin Exp Dermatol.* 2007;32:112-4.
32. Camacho F. Hipertrichosis e hirsutismo. In: Camacho F, Montagna W, editors. *Tricología. Tricology. Trichologie.* Madrid: Garsi; 1982. p. 123-38.