Do you know this syndrome?*
*Você conhece esta síndrome?**

Gisela Nascimento Pereira[1]
Laura de Freitas Xavier[2]
Neusa Yuriko Sakai Valente[3]
Jackson Machado-Pinto[4]

CASE REPORT

A two-year-old girl, leucodermal, was brought in by her parents, who reported she was born with straight black scalp hair (Figure 1), but, in the sixth month of age, hair started showing textural changes, becoming more curly, and changing color from black to brown and later to blond. The parents also reported loss of hair. Since then, the child presented blond hair that seemed to be uncombed in certain areas, particularly in the occipital region (Figures 2 and 3). There was no associated abnormality. There was also no report of chemical coumpound use, nor a family history of a similar condition.

At optical microscopy, the hair seemed normal.

The electronic microscopy showed hairs with a shallow and discontinued longitudinal ridge, with elliptical cross sections, with or without chamfer or bevel (reniform) (Figures 4 and 5). Some of the hairs also presented loss of normal cuticular pattern, with eroded areas.

WHAT IS THIS SYNDROME?
Uncombable hair syndrome

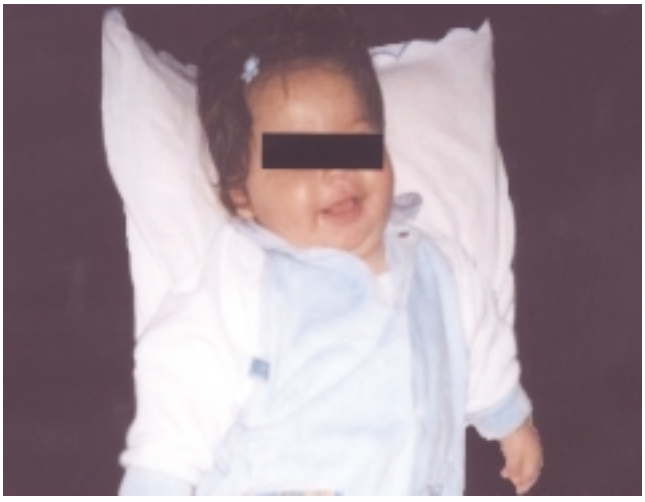

FIGURE 1: Young baby with dark straight hair

FIGURE 2: Child at two years of age, with blond, dry, unkempt hair

Received on January 02, 2006.
Approved by the Consultive Council and accepted for publication on January 10, 2006.
* Work done at Santa Casa de Misericórdia, Belo Horizonte / Faculdade de Ciências Médicas de Minas Gerais - Belo Horizonte (MG), Brazil.
Conflict of interest: None.

[1] Internal Medicine, at present specializing in Dermatology at the Santa Casa de Misericóridia de Belo Horizonte - Faculdade de Ciências Médicas de Minas Gerais - Belo Horizonte (MG), Brazil.
[2] M.D., Specialist in Dermatology.
[3] Ph.D. at the Department of Dermatology, Medical School, Universidade de São Paulo - USP - São Paulo (SP), Brazil.
[4] .D., M.Sc. in Dermatology, Ph.D. in Medicine, Head of the Dermatology Clinic, Santa Casa de Belo Horizonte and Faculdade de Ciências Médicas de Minas Gerais - Belo Horizonte (MG), Brazil.

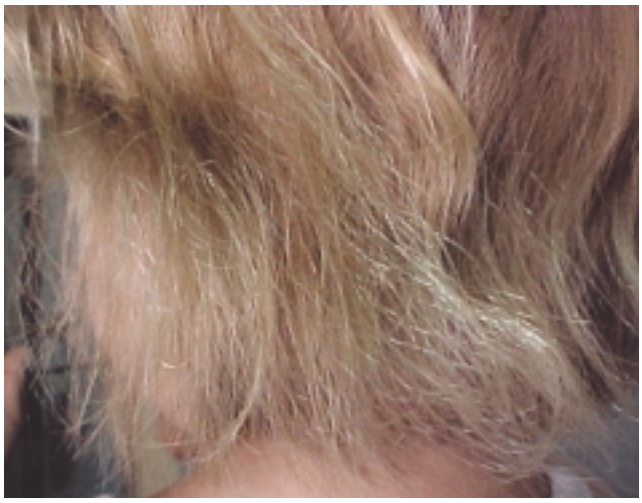

Figure 3: Child with tousled hair

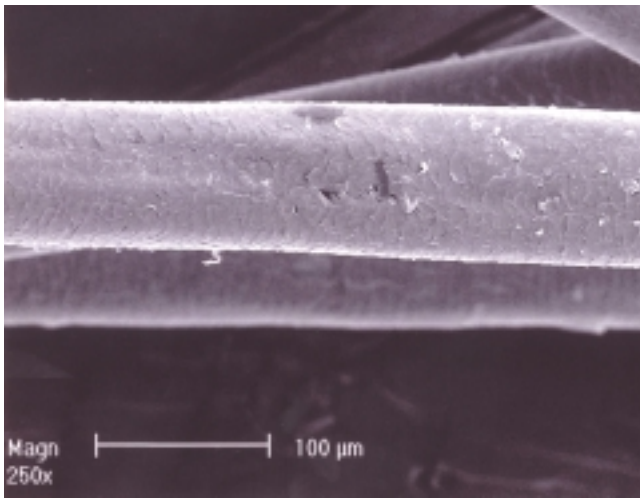

Figure 5: Electronic microscopy showing longitudinal ridge. Cuticle with areas with loss of normal pattern and erosion

The Uncombable Hair Syndrome may begin at any time during childhood, since the third month of age until adolescence. This is slow-growing silver-blonde, generally tousled, hair which does not change when combed or brushed. Even though it is dry and hard at the touch, it is not frail as a matter of course. The condition may be sporadic or of dominant autosomic inheritance, generally without any associated abnormalities.[1-4]

At optical microscopy, the hair seems normal or it has a longitudinal ridge. Differently from most hair shaft defects, which can be readily seen at simple microscopy, this does not occur with this syndrome.[2]

Electronic microscopy is diagnostic. It confirms the longitudinal ridge, and at cross section, 50% of the hairs are triangle-shaped, reniform, flattened, or simply irregular. The triangular shape and the ridge provide the basis for the name *pili trianguli et canaliculi*. The cuticle is normal but this case showed to behave otherwise. This may be explained by the wearing-out effect of the attempt to comb them by means of vigorous brushing. The longitudinal ridge is not specific to this syndrome. It may be present in normal hair and in other conditions, such as Marie-Unna Syndrome and many types of ectodermal dysplasia.[2-11]

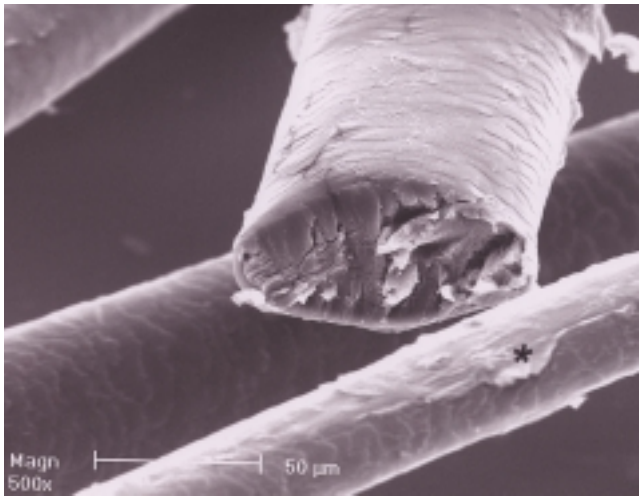

Figure 4: Electronic microscopy showing hair with elliptical cross section and loss of cuticle pattern. Hair(*) highlighted with cuticle erosion

As for pathogenesis, the hairshaft defect may be secondary to an abnormal configuration of the internal root sheath, which becomes prematurely keratinized in the hair follicle. Becoming stiff, it leads to the formation of the ridge on the malleable shaft that crosses it.[9]

Differential diagnosis involves the following entities:

- moniletrix: a rare disease of dominant autosomic inheritance with variable penetrance, which can occur in both sexes. Variations can be found in shaft thickness, with small nodes similar to necklace beads, producing partial alopecia. The velus hair is replaced by defective non-growing hair. Eyebrows, axillary, pubic and body hair may also be compromised and there may be association with keratosis pilaris and koilonychia;[12]

- Woolen hair: disease characterized by a diffuse or localized compromising of hair, which is curly, wool-looking, frail and, on occasion, lighter than the rest. The dominant or recessive autosomic inheritance can be seen, or the manifestation may be congenital, without genetic determinant. In the latter case, occurrence is not diffuse, compromising a specific area of the scalp. In this case, it is called woolen-hair nevus. According to the authors, this is the main

differential diagnosis of the case at hand, for the cross sections of woolen hairs are elliptical and ridges are not described on the shafts of such hairs;[12]

- trichorrhexis nodosa: congenital condition that may accompany other ectodermal defects. The hair cuticle undergoes rupture and the hair presents grayish nodules similar to interwoven paintbrushes.[12]

- *tricorrexis invaginata*: shows bamboo hair by hair intususception. It is a characteristic of Netherton Syndrome and some ichthyoses. The compromised hairs are dry, sparse, and frail, reflecting light in an irregular way;[12]

- *pili torti*: disease of dominant autosomic inheritance, more common in blonde girls. The hair is twisted along its axis. It becomes apparent from the second or third year of life, showing hair with an irregular shine and it is associable to other defects, such as neurosensorial deafness.[12]

As for treatment, there is none that is regularly effective. Nevertheless, there is a report of clinical improvement after four months supplementation with biotin (dosage of 0.9 mg / day VO).[5] Conditioners may be of some help. Spontaneous improvement occurs in some years.[3,5,9,10] ❑

Abstract: We report the case of a 2 year old girl, born with black and straight scalp hair. By the time she was 6 months old, hair color and texture changed to silver blond and became uncombable. The uncombable hair syndrome is a rare entity, in which scalp hair becomes dry and tousled after being previously normal in early childhood. Under light microscopy, hair may appear normal. With scanning electron microscopy the diagnosis can be reliably established. There's no effective treatment.
Keywords: Ectodermal dysplasia; Hair; Hair diseases

Resumo: *Os autores apresentam caso de criança de dois anos, com história de cabelos negros e lisos ao nascimento que, aos seis meses de idade, desenvolveram alterações da textura e da cor, tornando-se mais claros e impenteáveis. A rara síndrome dos cabelos impenteáveis é caracterizada pelo surgimento de cabelos de crescimento lento, loiro-prateados, desordenados, em indivíduos com cabelos previamente normais. A microscopia óptica é normal e a microscopia eletrônica é diagnóstica. Não há tratamento efetivo.*
Palavras-chave: Cabelo; Displasia ectodérmica; Doenças do cabelo

REFERENCES

1. Zanca A, Zanca A. Ancient observations of "Uncombable hair syndrome". Int J Dermatol. 1993;32:707.
2. Zegpi M, Roa I. The Uncombable hair syndrome. Arch Pathol Lab Med. 1987;111:754-5.
3. Ang P, Tay YK. What Syndrome is this? Uncombable hair syndrome (Pili trianguli et canaliculi). Pediatr Dermatol. 1998;15:475-6.
4. Ferrando J, Gratacós MR, Fontarnau R, Rodellas AC. Síndrome de los cabellos "impeinables". Med Cut ILA. 1977;1:39-46.
5. Shelley WB, Shelley ED. Uncombable hair syndrome: Observations on response to biotin and occurrence in siblings with ectodermal dysplasia. J Am Acad Dermatol. 1985;13:97-102.
6. Dupré A, Bonafé JL, Litoux F, Victor M. Le syndrome des cheveux incoiffables. Ann Dermatol Venereol. 1978;105:627-30.
7. Ferrando J, Fontarnau R, Gratacos MR, Mascaro JM. Pili canaliculi ("Cheveux incoiffables" ou "Cheveux en fibre de verre"). Ann Dermatol. Venereol. 1980;107:243-8.
8. Itin PH, Bühler U, Büchner SA, Guggenheim R. Pili trianguli et canaliculi: a distinctive hair shaft defect leading to uncombable hair. Dermatology. 1993;187:296-8.
9. Harper J, Oranje AP, Prose NS. Textbook of pediatric dermatology. Oxford: Balckwell Science Ltd; 2000. 1475 p.
10. Boyer JD, Cobb MW, Sperling LC, Rushin JM. Loose anagen hair syndrome mimicking the uncombable hair syndrome. Cutis. 1966;57:111-2.
11. Laurent R, Yulzari M, Makki S, Agache P. Syndrome des cheveux incoiffables. Ann Dermatol Venereol. 1978;105:633-5.
12. Lowy G, Alonso FJF, Cestari SCP, Cestari TF, Oliveira ZNP. Atlas de dermatologia pediátrica. Rio de Janeiro: Medsi; 2000. p.96-7.

MAILING ADDRESS:
Gisela Nascimento Pereira
Rua Washington Luiz,175 - Santa Tereza
36201-066 - Barbacena - MG
Tel.: +55 31 3377-1307 / +55 32 3331-2375
E-mail: giselamf@uol.com.br