

Denouement and Discussion

Uncombable Hair (*Pili Trianguli et Canaliculi*)

The clinical features and dermoscopy and electron microscopy findings lead to the diagnosis of *pili trianguli et canaliculi*, or uncombable hair syndrome. The (unworried) patient and his family were reassured that the condition is harmless and would improve over time, as had been observed with the patient's sister. Uncombable hair syndrome was first described in 1973 by Stroud and Mehregan¹ and by Dupré and colleagues.² This syndrome is a rare, genetically determined hair shaft abnormality transmitted in an autosomal dominant fashion with variable penetrance. As an underlying pathologic condition, a keratinization disorder of the hair shaft is suspected. The clinical presentation is highly distinct, with silvery-blond, dry, frizzy hair that stands out from the scalp and is resistant to combing.³ The phenotype is the result of the longitudinal grooving of the hair shaft.⁴ The anomaly is limited to the scalp hair.⁵ Occasionally, *pili trianguli et canaliculi* can be found in children with normal-appearing hair. In contrast to uncombable hair syndrome, the hair shaft abnormality is only present in a limited number of hairs, and with absence of the characteristic phenotype, this can be considered a variation from the norm. In individuals with uncombable hair syndrome as manifested in our patient, however, the vast majority of the scalp hair (>50%) demonstrates grooving and triangular shape, so by observing this feature, together with the clinical appearance of unruly, frizzy hair, the diagnosis can be made.⁶

This hair phenotype usually occurs as an isolated condition and without any associated symptoms. Nevertheless, uncombable hair has been reported together with ectodermal dysplasia, hypotrichosis of Marie Unna, retinal dysplasia, pigmentary dystrophy, juvenile cataract, digit abnormalities, tooth enamel anomalies, oligodontia, and angel-shaped phalangeoepiphyseal dysplasia.^{7,8} However, in cases of ectodermal dysplasia, a grooving and twisting of the hair shaft (*pili torti et canaliculi*) can be observed.⁹ Although Shelley and Shelley¹⁰ report a positive effect of biotin on the hair shaft structure, no specific therapy is recommended in patients with *pili trianguli et canaliculi* because the condition usually improves spontaneously with age. Using conditioner can increase hair strength to a certain extent and may improve its appearance owing to the moisturizing effect. Most important, affected individuals and their family members should be informed about the benign and self-limiting nature of

this condition and its autosomal-dominant inheritance pattern.

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REFERENCES

1. Stroud JD, Mehregan AH. *The First Human Hair Symposium*. New York, NY: Medcom Press; 1973.
2. Dupré A, Rochiccioli P, Bonafé JL. «Cheveux incoiffables»: anomalie congénitale des cheveux. *Bull Soc Fr Dermatol Syphiligr*. 1973;80:111-112.
3. Rieubland C, de Viragh PA, Addor MC. Uncombable hair syndrome: a clinical report. *Eur J Med Genet*. 2007;50(4):309-314.
4. Happle R. *Hair Anomalies and Syndrome Recognition*. London, England: Martin Dunitz Ltd; 2000.
5. Blume-Peytavi U, Mandt N. *Hair Shaft Abnormalities*. Philadelphia, PA: Churchill Livingstone; 2000.
6. Hicks J, Metry DW, Barrish J, Levy M. Uncombable hair (cheveux incoiffables, *pili trianguli et canaliculi*) syndrome: brief review and role of scanning electron microscopy in diagnosis. *Ultrastruct Pathol*. 2001;25(2):99-103.
7. Whiting DA. *Hair Shaft Defects*. Barcelona, Spain: McGraw-Hill; 2003.
8. Fritz TM, Trüeb RM. Uncombable hair syndrome with angel-shaped phalangeoepiphyseal dysplasia. *Pediatr Dermatol*. 2000;17(1):21-24.
9. Trüeb RM, Spycher MA, Schumacher F, Burg G. *Pili torti et canaliculi* in ectodermal dysplasia [in German]. *Hautarzt*. 1994;45(6):372-377.
10. 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(1):97-102.

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