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

Gustavo Novaes Martins Ferreira[1]
Neusa Yuriko Sakai Valente[2]
Ricardo Romiti[3]

CASE REPORT

Two-year-old white male patient with blond and slightly curled hair, who had been presenting hair rarefaction over the past six months in the vertex region, as well as in the occipital portion of scalp. Mother had noticed spontaneous massive detachment of hairs in the affected regions since the beginning of the condition. She denied any previous diseases affecting hairs or even scalp.

Upon dermatological examination, extensive hair rarefaction was noticed in the vertex region (Figure 1), associated to thin and sparse hairs, which could be easily extracted upon mild traction, with no pain. In the occipital region, hairs formed curls that the mother had difficulty for combing. He presented no cutaneous lesions, nor dental or ungueal alterations.

Trichogram showed 100% of anagen hairs, none of them exhibiting inner or outer involving sheaths (Figures 2 and 3). In many of them, cuticles were ruffled, in a "loose sock" aspect (Figure 4), and irregular longitudinal depressions occurred in the proximal portions of the hair shafts, next to the bulb. As to hair caliber, all were of the vellus type, or intermediate.

He was the single child of non-related parents, and did not present any other personal or familial affection. Laboratorial tests were within normality.

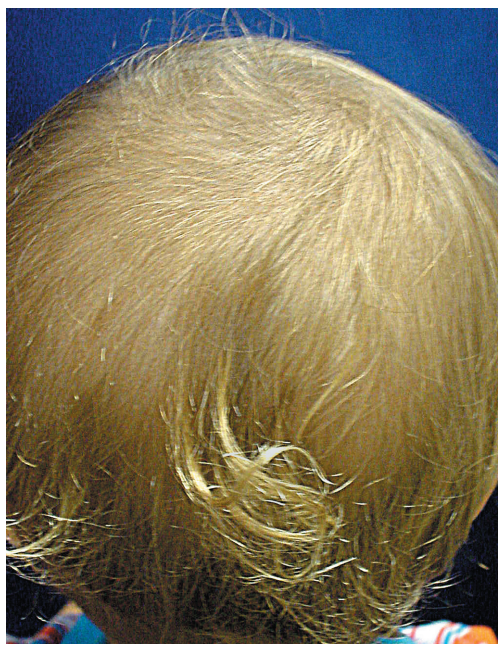

FIGURE 1: Thin and sparse hairs in the vertex region

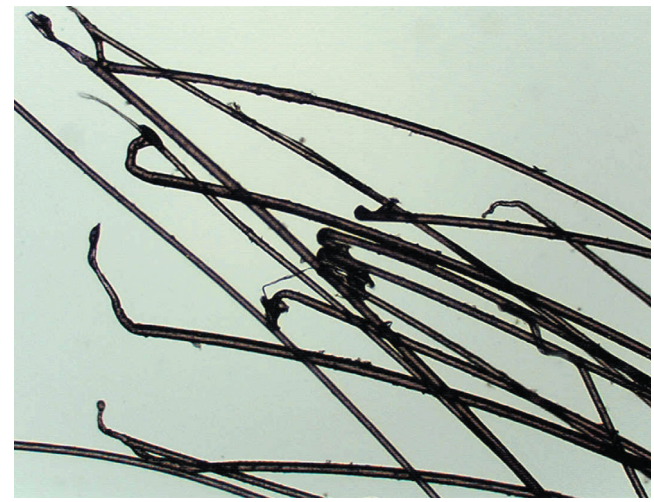

FIGURE 2: Trichogram showing anagen hairs with absence of involving sheaths (100x)

Received on March 22, 2006.
Approved by the Consultive Council and accepted for publication on June 23, 2006.
* Work done at Dermatology Department of the Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo - FMUSP - São Paulo (SP), Brazil.
Conflict of interests: None

[1] Resident Doctor in Dermatology at the Department of Dermatology, Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo - FMUSP São Paulo (SP), Brazil.
[2] PhD, Professor at the Department of Dermatology, Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo - FMUSP - São Paulo (SP), Brazil.
[3] PhD, Professor at the Department of Dermatology, Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo - FMUSP - São Paulo (SP), Brazil.

FIGURE 3: Greater detail of a loose anagen hair (200x)

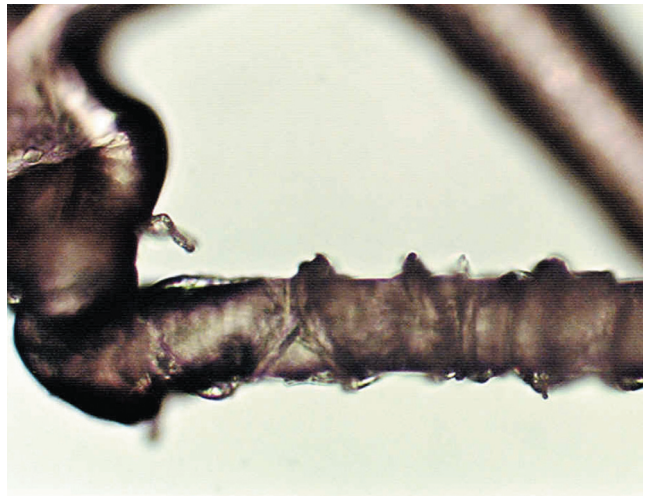

FIGURE 4: Ruffled aspect of hair cuticle (400x)

WHAT IS THIS SYNDROME?
Loose anagen hair syndrome

History

Loose anagen hair syndrome (LAHS) was first described in 1984 by Zaun,[1] under the denomination of *Syndrome of loosely attached hair in childhood*, and then by Nödl,[2] in 1986. Current name (LAHS) was introduced in 1989, simultaneously by Hamm[3] and Price.[4]

Epidemiology and clinical picture

The syndrome affects mainly children of the female gender, with onset of manifestations between two and six years of age. The patients usually have blond or light-brown hair. Individuals with darker hair have also been reported.[5]

LAHS tends to sporadic; however, familial cases with features of dominant autosomic inheritance have been reported.[6,7] Familial occurrence associated to systemic alterations, including colobomas and macular dystrophy, has been rarely observed.[8,9]

Clinically, it is characterized by rarefaction and reduction of hair length; hairs detach easily from scalp, and are extracted with no pain upon mild traction. Hairs are thinner than normal, and do not need frequent haircuts, for they grow slowly. This fact is more often noted in girls, which generally have longer hairs. Presence of curls in the occipital region, often rebel to combing,[3-7] as observed in the present case.

Diagnosis

LAHS diagnosis is based on clinical picture and presence of loose anagen hairs (LAH), defined on microscopic examination with anagen hairs without inner and outer sheaths, with bulbs of irregular shape, presenting in their proximal portion a ruffled cuticle with a "loose sock" aspect. However, LAHS diagnosis depends on the number and percentage of LAH in the mild traction test and in trichogram. In order to understand the meaning of the results of these to LAHS diagnostic tests, it can be useful to review a few characteristics of the hairs of normal people.

Most hairs in the human scalp are anagen (around 85%), and almost all the rest is of telogen hairs (15% in average). Mild traction can result in the detachment of a few telogen hairs, but not of anagen. In order to detach anagen hairs from a normal scalp, forced traction by means of the use of an instrument, such as a hemostat, is needed. Hairs pulled by forced traction are usually removed along with their inner and outer sheaths. In trichogram, 70% or more of hairs should be LAH for a diagnosis that is consistent with LAHS.[5]

Pathogenesis

LAHS is posited to be the result of premature keratinization of the inner hair sheath of the hairs, leading to deficient adhesion between cuticles of the hair and inner sheath.[10,11] Chapalain et al.[10] described mutations on gene K6HF, which has been identified as responsible for the production of keratin in of the hair sheath, detected in members of three out of nine analyzed LAHS families.

Prognosis and treatament

In adolescence, affected patients usually start presenting faster hair growth, with hairs progressively becoming longer, denser and darker. Most patients continue presenting a small percentage of LAH on trichogram and mild traction test, which demonstrates the permanence of the defect in hair anchoring, albeit

less intense. This fact also explains the rarity of diagnosis of the syndrome in adults.[5,12]

There is no specific therapy for LAHS. Further elucidation of its physiopathological mechanisms may perhaps make future therapeutic interventions possible for the more severe cases.

Abstract: Loose anagen hair syndrome is characterized by thin and sparse hairs that can be easily extracted upon mild traction. The disorder affects predominantly children. Trichogram shows at least 70% of loose anagen hairs devoid of inner and outer hair sheaths, presenting a ruffled cuticle at the proximal portion. Although benign and self-limited, the disorder frequently concerns parents and should be further differentiated from telogen effluvium and trichotillomania.
Keywords: Alopecia; Hairs; Hypotricosis

Resumo: *A síndrome dos cabelos anágenos frouxos caracteriza-se por cabelos finos e rarefeitos que podem ser facilmente extraídos, de forma indolor, mediante leve tração. Acomete principalmente crianças. O tricograma evidencia 70% ou mais de pêlos anágenos desprovidos das bainhas radiculares interna e externa, apresentando cutícula enrugada em sua porção proximal. Apesar de benigno e auto-limitado, o distúrbio freqüentemente aflige os pais e deve ser diferenciado do eflúvio telógeno e da tricotilomania.*
Palavras-chave: Alopecia; Cabelos; Hipotricose

REFERENCES

1. Zaun H. Syndrome of loosely attached hair in childhood. Pediatric dermatology: advances in diagnosis and treatment. New York: Springer Verlag NY Inc; 1984. p.64-5.
2. Nödl F, Zaun H, Zinn KH. Gesteigerte epilierbarkeit von anagenhaaren bei kindern als folge eines rei fungsdefects der follikel mit gestörter verhafftung von haarschaft und wurzelscheiden. Aktuelle Derm. 1986;12:55-7.
3. Hamm H, Traupe H. Loose anagen hair of childhood: the phenomenon of easily pluckable hair. J Am Acad Dermatol. 1989;20:242-8.
4. Price VH, Gummer CL. Loose anagen syndrome. J Am Acad Dermatol. 1989;20:249-56.
5. Tosti A. Loose anagen hair syndrome and loose anagen hair. Arch Dermatol. 2002;138:521-2.
6. Tosti A, Peluso AM, Misciali C, Venturo N, Patrizi A, Fanti PA. Loose anagen hair. Arch Dermatol. 1997;133:1089-93.
7. Baden HP, Kvedar JC, Magro CM. Loose anagen hair as a cause of hereditary hair loss in children. Arch Dermatol. 1992;128:1349-53.
8. Murphy MF, McGinnity FG, Allen GE. New familial association between coloboma and loose anagen syndrome. Clin Genet. 1995;47:214-6.
9. Sato MT, Brenner FM, Marzagão R. Síndrome dos cabelos anágenos frouxos associada à distrofia macular – descrição de uma família. An Bras Dermatol. 2004;79:725-31.
10. Chapalain V, Winter H, Langbein L, Le Roy JM, Labreze C, Nikolic M, et al. Is the loose anagen hair syndrome a keratin disorder? A clinical and molecular study. Arch Dermatol. 2002;138:501-6.
11. Lalevic-Vasic B, Polic DJ, Milinkovic R. Le syndrome des cheveux anagènes caducs. Ann Dermatol Venereol. 1990;117:701-7.
12. O'Donnel BP, Sperling LC, James WD. Loose anagen hair syndrome. Int J Dermatol. 1992;31:107-9.

MAILING ADDRESS:
Ricardo Romiti
Rua Dr. Neto de Araújo, 320 - cj. 1004 – Vila Mariana
04111-001 - São Paulo – SP - Brazil
E-mail: rromiti@hotmail.com