Short anagen syndrome

Richard J. Antaya, MD,a,b Eirini Sideridou, MD,c and Elise A. Olsen, MDd

New Haven, Connecticut, and Durham, North Carolina

Short anagen syndrome is an uncommon, probably underreported, condition whose clinical characteristics are poorly recognized and whose incidence is poorly documented in the medical literature. We describe the clinicopathologic features of a child with short anagen syndrome and propose methods for diagnosing this entity by clinical examination, trichogram, light microscopic examination of the hair shaft, scalp biopsy, and measurement of scalp hair growth rate. (J Am Acad Dermatol 2005;53:S130-4.)

Hair that “doesn’t grow long” can be psychologically traumatic for parents of young children with this problem, perplexing for physicians, and full of potential multisystem ramifications for the affected child. The differential diagnosis includes conditions, most commonly genetic, that cause the hair to break off or genetic abnormalities in hair production, most commonly associated with other abnormalities. Fortunately, short anagen syndrome is a condition that is unassociated with either scalp hair breakage, total hair loss, or usually with any other serious association. There are very few reports, however, of this condition in the literature,1-3 possibly secondary to the relatively banal presentation and the usual reliance on history versus physical findings to make the diagnosis.

We report here a case of a 10-year-old child whose short, fine hair since birth is best captured under the term short anagen syndrome. The means by which to definitely make this diagnosis by clinical and microscopic examination of hairs are discussed and defined.

CASE REPORT

A healthy, 10-year-old white girl was referred to the Yale Pediatric Dermatology Clinic with very short, fine, blond scalp hair that had not been cut since birth. She was the firstborn of healthy, non-consanguineous parents. The pregnancy and perinatal period had been uneventful. At birth, she was said to have scanty, blond hair, which did not seem grossly abnormal. Otherwise, she was healthy, and her growth and development had been normal. She and her parents denied any problems with her teeth, nails, or sweating. Neither her parents nor her younger sister had any hair abnormalities. Two paternal aunts, however, were reported to have had very short hair until 3 years of age.

Physical examination revealed short scalp hair that was uniform in quality, fine in texture, and distributed in a normal pattern with mildly decreased overall density (Fig 1). Her eyelashes, eyebrows, and hair elsewhere on the body were normal. The rest of the physical examination was normal, without any abnormalities of the skin, teeth or nails.

HAIR EVALUATION METHODS

The following tests were performed after the purpose of the evaluation was explained to the patient and her parents, and consent obtained:

• Maximum scalp hair length: This value was determined by direct physical examination.

Fig 1. Girl with sparse, fine, short, blond scalp hair at age 11 years.
• Microscopic examination of the hair shafts: Hairs were cut, and the distal and mid shafts examined under light microscopy.
• Hair pull test: Gentle hair pull was performed from several areas of the scalp, and microscopic examination of the hair roots was performed.
• Anagen to telogen ratio: Approximately 40 hairs were plucked from the vertex region, and the roots were examined at 10 × magnification to establish anagen to telogen ratio.
• Histology: A 4-mm punch biopsy of the scalp skin was taken from the occipital region. The specimen was sectioned vertically, stained with hematoxylin-eosin, and evaluated under light microscopy.
• Mean growth rate of the hair: This value was derived by close shaving the hair in an area of approximately 4 cm² at the vertex of the scalp and, 4 weeks later, reshaving the same area, and collecting and measuring the length of the shaved hair.
• Scanning electron microscopic examination: Approximately 4 plucked and 4 shaved hairs were examined under the scanning electron microscope for potential hair shaft abnormalities, and measurement of hair diameter and description of the cuticular structure were performed.

RESULTS
Maximal scalp hair length was 35 mm. Examination of the patient's hair shafts by light microscopy showed normal anatomy and tapered tips. There was an increased number of telogen hairs extracted in all areas of the scalp with the hair pull test. The anagen to telogen ratio (8/25) derived from the hair pluck was significantly reduced with only 24% of hairs in anagen. The scalp biopsy revealed both terminal as well as vellus hair follicles. Because vertical instead of horizontal sections were prepared, no quantitative numbers of either could be determined. There was a mild lymphocytic infiltrate around a few follicles, and a spotty lymphohistiocytic infiltrate in the upper and mid dermis but no peribulbar inflammation or fibrosis. Measurement of shaved hairs provided a mean scalp hair growth rate of 0.3 mm per day. Scanning electron microscopic examination showed hair shafts of normal shape but small caliber (average diameter, 33 μm), with minimal widening of the cuticular spaces. Less overlap between successive cuticular cell edges (widened spaces) has been seen on electron microscopic evaluation of the hair shafts in patients with tricho-dental syndrome and short anagen hairs.

DISCUSSION
Our patient had a congenital anomaly of abnormally short scalp hair. There are many syndromes with this phenotype that involve hair fragility and breakage, but few in which the hair density is fairly normal and the hairs are otherwise normal except for length (Table I). Support for the diagnosis of short anagen syndrome includes the normal hair shafts with a short growth phase as shown by the maximum length of 35 mm when one might expect up to 10 times that length, decreased number of hairs in anagen (24% vs normal of 90%+), and confirmation that hairs were not breaking off (tapered tips of normal anagen hairs). Other causes of a decrease in anagen hairs, such as telogen effluvium or female pattern hair loss, are excluded on the basis of the patient’s history (congenital onset) and/or clinical features (diffuse loss). Loose anagen syndrome is ruled out by the absence of loose anagen hairs on hair pull and the absence of any unruly sections of hair.

The major problem in short anagen syndrome and the one that leads to all the other clinical findings is decreased anagen duration. The duration of anagen, which determines ultimate hair length, appears to be under genetic control but is modified by age, sex, and probably other factors, including hormones. Eyebrows and eyelashes are shorter than scalp hair because they have a shorter anagen phase. Duration of anagen may be grossly calculated by dividing the overall length of an uncut hair by the average daily scalp hair growth rate for that patient. In our patient, the growth rate was normal or 0.3 mm/day. As the maximal length of the hair was 3.5 cm in our patient, the duration of anagen in her scalp hair is approximately 4 months, far shorter than the normal of up to 36 months in adults. Whether all patients with short anagen syndrome will have normal scalp hair growth rate is yet to be determined.

Headington in his 1993 review on telogen effluvium speculated that some individuals may experience increased shedding related to idiopathic shortening of anagen. Kersey in 1987 was the first to provide documentation that the short scalp hair in a patient with tricho-dental syndrome was caused by a short anagen phase of the hair cycle. In his review of congenital hypotrichosis, de Berker points out that shortened anagen phase provides an explanation for hair that does not appear to grow. Whitmore in 1999 reported a 19-year-old woman with a lifelong history of sparse and short scalp hair growth with tapered ends of scalp hair, negative hair pull, and increased telogen percentage on hair pluck, all consistent with the short anagen syndrome. This latter patient also had linear scleroderma of the left
leg and had had surgical repair of an occluded lacrimal duct. Particularly germane to our case, Barraud-Klenovsek and Trüeb recently reported on 2 cases of congenital hypotrichosis related to short anagen without any associated abnormalities. Although there is scanty literature on this problem, according to Elise A. Olsen, MD (oral communication, 2002), short anagen syndrome (ie, short hair with variable decreased hair density and without associated hair shaft fragility or hair unruliness in an otherwise healthy child) appears to be an uncommon but not rare condition in children.

In the United States and most countries, 5% topical minoxidil is licensed for use in androgenetic alopecia in men although it does not have any specific antiandrogen effect. Many clinicians use it also for other than its licensed indications, for example in alopecia areata, congenital hypotrichosis, and loose anagen syndrome. The precise mechanism by which minoxidil stimulates hair growth is still not established, but clinically minoxidil stimulates telogen hair follicles to enter into anagen and prolongs the duration of anagen. Thus, topical minoxidil would seem to be an appropriate treatment for this condition. Unfortunately, our teenage patient, who has worn a wig successfully for years, was non-compliant and did not remain on the medication long enough to fully assess its efficacy.

Short anagen syndrome is a relatively recently recognized entity, which is poorly documented in the literature. However, it can be readily diagnosed by simple clinical means: a history of congenitally short hair without evidence of increased fragility or breakage or underlying scalp inflammation; a physical examination of the scalp characterized by uniformly and abnormally short scalp hair with decreased density (but not frank alopecia); a normal light microscopic examination of the hair shafts including tapered tips of hairs that have not been clipped or cut; and a hair pluck revealing a significant increase in the telogen to anagen ratio. Evaluation of the mean hair growth rate by close shaving the scalp hair and reevaluating several weeks later rules out a growth disturbance. This “hair window” is noninvasive, inexpensive, and, when combined with the other findings, helps cinch the diagnosis. Skin biopsy and electron microscopic examination of the hair shaft add little to the evaluation, and we do not recommend these for the routine evaluation of this condition.

Table I. Hair disorders characterized by “hair that won’t grow long” in otherwise healthy-appearing children

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Genetics</th>
<th>Scalp hair: clinical findings</th>
<th>Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td>Short anagen syndrome</td>
<td>?</td>
<td>Diffuse short hair with decreased density, but no baldness</td>
<td>Birth to early childhood</td>
</tr>
<tr>
<td>Tricho-dental dysplasia</td>
<td>AD</td>
<td>Short, fine hair</td>
<td>Infancy</td>
</tr>
<tr>
<td>Hereditary hypotrichosis</td>
<td>AR, AD, X-linked recessive</td>
<td>Sparse, thin, and short scalp hair</td>
<td>Birth to infancy</td>
</tr>
<tr>
<td>Loose anagen syndrome</td>
<td>AD or sporadic</td>
<td>Three major phenotypes: (1) diffuse decrease in density, slow growing short hair; (2) patchy unruly hair; (3) easily extractable, otherwise normal hair</td>
<td>~Age 2 y</td>
</tr>
<tr>
<td>Juvenile macular dystrophy and congenital hypokeratosis</td>
<td>AR</td>
<td>Sparse hair that never requires cutting; fusiform beading of hair shaft or pili torti</td>
<td>Birth</td>
</tr>
<tr>
<td>Monilethrix</td>
<td>AD</td>
<td>Fragile, beaded hair and baldness</td>
<td>Birth to adolescence</td>
</tr>
<tr>
<td>Pili torti (not as part of syndrome)</td>
<td>AD, AR, or sporadic</td>
<td>Brittle, spangled hairs, especially in the occipital and temporal regions</td>
<td>Birth to age 2 y</td>
</tr>
<tr>
<td>Trichothiodystrophy</td>
<td>AR</td>
<td>Alopecia of scalp with extremely brittle, short, flattened hairs</td>
<td>Early infancy</td>
</tr>
</tbody>
</table>

A. Anagen; AD, autosomal dominant; AR, autosomal recessive; T, telogen; MR, mental retardation.
At this point, we can only speculate about the eventual outcome in our patient or in others with this problem. Barraud-Klenovsek and Trüb report that their first patient with congenital hypotrichosis/short anagen syndrome had three family members with a similar disorder that appeared to resolve spontaneously during puberty and adulthood.

The authors thank Dr David Whiting for generously and skillfully reviewing the scalp biopsy of our patient.

REFERENCES