



Hairy elbows

A case study

Pablo Fernandez-Crehuet

Ricardo Ruiz-Villaverde

Jose Luis Fernandez-Crehuet Serrano

Case study

A boy aged 6 years was referred to our clinic for evaluation of the presence of fair, thin hair on both of his elbows. This condition had first been observed when he was 2 years of age and the hair had subsequently increased in length and thickness (*Figure 1*). He had a history of asthma and was being treated by a paediatrician. His family medical record was otherwise unremarkable.

This unusual hairiness was symmetrically distributed on the extensor surfaces of both proximal forearms and distal arms. The underlying skin showed no abnormalities. No hypertrichosis was found elsewhere and examination of teeth, skeleton and fingernails was also normal. No other morphological changes were noted. In addition, his height was appropriate for his age. No developmental, mental or physical impairment was observed.

The blood cell count and general biochemistry, as well as thyroid and sexual hormonal profiles were all normal. Radiological examination, which was performed on the parents' request, was normal.

The boy was diagnosed with hypertrichosis cubiti (HC), and shaving of the areas was recommended.

Question 1

What is a primary nevoid hypertrichosis?



Figure 1. Bilateral hairy elbows

Question 2

Do you need to perform any tests to diagnose it?

Question 3

What is an appropriate therapeutic approach to this condition?

Answer 1

Nevoid hypertrichosis is a localised type of hypertrichosis characterised by its low incidence. It is defined by the growth of hair on a localised area of the body surface at birth or during the first years of life. It usually involves an isolated region but, occasionally, several areas may be involved.¹

Nevoid hypertrichoses can be subdivided into primary or secondary. HC, or hairy elbows syndrome (*Table 1*), is included in the first group. It was first described by Beighton² in 1970. This circumscribed hypertrichosis clinically presents with a significant amount of lanuginous, thin hair on skin otherwise normal in texture and morphology. The distribution is symmetrical and bilateral on the extensor surfaces of the distal third of the arms and proximal forearms. These hairs progressively become longer and more pigmented.

This condition frequently arises in early childhood (1–3 years) and reaches its greatest severity at the age of 4–5 years.³ The inheritance pattern is variable. There have been reports of autosomal dominant and recessive cases, as well as sporadic ones.

Table 1. Primary nevoid hypertrichosis

- Hairy elbow syndrome
- Eyebrow hypertrichosis
- Hypertrichosis of the auricle
- Hairy polythelia
- Posterior cervical hypertrichosis
- Anterior cervical hypertrichosis
- Eyelashes hypertrichosis
- Nose edge hypertrichosis
- Hairy palms and soles malformation

Answer 2

As far as the hairy elbows syndrome is concerned, two types of patients may be considered. On the one hand, there are sporadic cases that do not present with any radiological, morphological or hormonal changes. On the other hand, some cases of HC form part of a more complex syndrome. Several anomalies and disorders have been reported in the literature, such as short stature (usually restricted to cases of patients with Weill-Marchesani syndrome), facial or limb deformities, developmental disorders, bone disorders and ocular and neurological abnormalities. Therefore, the need to perform a workup to rule out an associated disorder will be determined by the findings of a properly conducted medical history. Awareness of this condition is important in avoiding unnecessary tests.

A skin biopsy of the affected area does not usually provide any significant data. A trichogram may show a normal proportion of hair in all three phases of their cycle, or else increased hair in the

anagen phase, which would explain their length (eg. 90% anagen, 9% telogen, 1% catagen).

Answer 3

It is important to emphasise to parents the benign nature of HC, so that expenditure on expensive and useless treatments can be avoided. Depigmentation or shaving of the area may be initially recommended. More definitive therapies, such as laser hair removal, should be delayed until adolescence for those rare cases that have not spontaneously regressed by then.⁴ Additionally, the psychosocial needs of the child should be considered.

The actual prevalence of HC, as well as its strength of association with other disorders, remains unknown. This may be the result of misdiagnosis or miscommunication in sporadic cases such as the one reported.

Authors

Pablo Fernández-Crehuet Serrano MD, PhD, Consultant, Department of Dermatology, Hospital

Alto Guadalquivir, Andújar, Jaén, Spain. pablocrehuet@hotmail.com

Ricardo Ruiz-Villaverde MD, PhD, Consultant, Department of Dermatology, Complejo Hospitalario de Jaén, Spain

Jose Luis Fernández-Crehuet MD, PhD, Consultant, Department of Dermatology, Hospital Alto Guadalquivir, Andújar, Jaén, Spain

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correspondence afp@racgp.org.au