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Congenital hypertrichosis lanuginosa: neonatal shaving

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SUMMARY A newborn baby presented with overgrowth of lanugo hair and a precocious tooth. She was shaved infrequently until aged 9 months, when spontaneous loss of body hair began. Pyloric stenosis at 6 weeks was surgically corrected. There was a family history of hypertrichosis and dental anomalies.

Congenital hypertrichosis lanuginosa is a rare disorder that causes persistent, excessive lanugo and dental anomalies. Neonatal and dermatological textbooks do not provide guidance on immediate management: this report describes one approach, shaving.

Case report

The baby girl, the first child of unrelated parents,

Fig. 1 A case of congenital hypertrichosis lanuginosa at presentation at 4 days of age.

was covered at birth with long, curly blond hair, which spared only her hands, feet, and face (Fig. 1). It was soft and silky, up to 2.5 cm long, encroached onto the forehead, cheeks, pinnae, and inside the external auditory meati, and extended to the margins of the labia minora and anal verge. The scalp hair was the same length and texture, but darker and thicker. There was a partially erupted upper central incisor tooth but no other abnormality: she weighed 3350 g, with length 49.5 cm and head circumference 35 cm.

The mother, a hairdresser, denied using any hair promoting applications at work. The father, an airforce technician, admitted that members of his family were, or had been, excessively hairy. He himself had been very hairy at birth, with a precocious tooth. His excess hair had gradually disappeared, though he had needed dentistry for supernumerary secondary teeth. Affected members of his family were all born with a single tooth. There was sparse but unusually long hair on his forearms and the dorsal surface of his hands: his hair distribution elsewhere was normal.

Dermatology textbooks provided the diagnosis but no help with immediate management, and the parents wanted 'something to be done'. Mother and baby were nursed in a single room, for privacy, but she did not express any feelings of alienation about her daughter, whose thick curls were not unattractive. Indeed, when she went to a routine demonstration of baby bathing for new mothers, she returned disconcerted: 'that baby was *completely bald*'.

Embarrassment, and the difficulty in keeping the profusely hairy nappy area clean, persuaded the parents and myself that shaving was necessary. We used a conventional razor on the fourth day, but after an hour, and with only half the baby shaved, this method was abandoned. The following day an electric 'Ladyshave' razor, used dry, completed the shave in half an hour, easily and painlessly. They went home the same day: the baby was breast fed.

She was admitted aged 5 weeks with pyloric stenosis, which was treated successfully by pyloromyotomy. There was no family history of this. She was shaved all over every two weeks or so until 3 months of age, when arms and legs only were shaved, monthly. Her mother stopped shaving her by 9 months, when body hair was spontaneously being shed. At 14 months there was only excess hair around her ears, wrists, buttocks, and ankles (Fig. 2). Her eyelashes were very long, and her scalp hair needed cutting four times in the first year. When last seen, at the age of 19 months, she was having trimmings to the facial and aural hair every three to four months.

The precocious upper incisor was shed in the first

Fig. 2 The case at 14 months of age, after no shaving for five months, showing spontaneous hair loss.

week. Two lower incisors appeared at 9 months, four more by 14 months, with a gap at the position of the neonatal tooth, where a tooth finally erupted at 17 months. The primary incisors appeared unusually small (Mr D R Purnell, Consultant Oral Surgeon).

Discussion

Excess hair in childhood may be due to either *hirsutism*—that is, androgen induced, with an adult sexual distribution of coarse 'terminal' hair (for example, the adrenogenital syndrome)—or *hypertrichosis*—that is, an excess of vellus or lanugo hair, drug induced (for example, phenytoin or diazoxide) or accompanying a syndrome (for example, de Lange's or Hurler's syndromes).

Congenital hypertrichosis lanuginosa is almost invariably transmitted as an autosomal dominant with varying expressivity.^{1 2} In some families the hairiness is present at birth and persists and in some it diminishes; other case reports describe little excess hair at birth, but increasing hair growth into adult life. Anomalous dental eruption is common and sometimes there are ear malformations: pyloric stenosis is not reported.^{1 3}

Management includes dental care, counselling, shaving, bleaching the hair, and chemical and electrical epilation. A 4 year old boy's hypertrichosis was controlled by shaving and epilatory creams, but at birth only his facial hair was excessive, and generalised hypertrichosis developed during infancy and diminished in later childhood.² No treatment is mentioned of two children aged 3 and 16 months whose similarly affected mother shaved and used chemical and electrical epilation, and neonatal shaving is not described in the accompanying review

of other case reports.³ It is reassuring that shaving does not increase the profusion or rate of regrowth of hair,^{1 4} despite what old wives' tales may warn, or young shavers hope, to the contrary.

Congenital hypertrichosis lanuginosa is a very rare condition, with about 40 case reports.¹ Beighton quotes an incidence of one in 1000 million.² When presented with a newborn baby affected by a rare syndrome it may be difficult not to fudge or bluff: here the first reaction was to say that it was an extreme instance of the profuse lanugo of some ethnic groups and many preterm babies. Others in this family had been similarly misinformed and did not subsequently seek medical advice. The best paediatric policy is to admit ignorance but to correct it promptly.

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