one might postulate an autosomal dominant inheritance. We have been unable to find any reference to the mode of inheritance of the anomaly, nor are there any other reports of more than one case within a family.

We thank Dr A J Keay and Dr J Syme for referring these cases, Dr M J Godman who performed angiographic studies, and Mr W H Bisset who carried out surgery.

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Rapp-Hodgkin ectodermal dysplasia syndrome

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SUMMARY We report a mother and daughter with Rapp-Hodgkin ectodermal dysplasia syndrome. Features of the previously described cases were confirmed, although no growth deficiencies were observed. Autosomal dominant inheritance was probable although X-linked transmission could not be excluded.

The Rapp-Hodgkin syndrome is a dominantly inherited syndrome characterised by hypohidrotic ectodermal dysplasia associated with a narrow nose, cleft palate or lip, or both, a small mouth, short stature, and hypospadias in the male. It is a distinct syndrome and differs from other well-known hypohidrotic and hidrotic ectodermal dysplasias. The first family (mother, son, and daughter) affected by the syndrome was reported in 1968 by Rapp and Hodgkin.¹ In 1971 Summitt and Hiatt² reported a single case with the same symptoms. This paper describes a second family (mother and daughter) with a Rapp-Hodgkin syndrome.

Case report

The 4½-year-old girl was admitted to our hospital with suspected ingestion of a foreign body. She had been born at 38 weeks' gestation (birthweight 2650 g; length 48 cm). The mother had not taken any unusual drugs during the pregnancy. At the time of conception the mother was 30 and the father 32; the

parents were not blood-relatives; the girl is an only child. At birth bilateral cleft palate and bifid uvula were noted. She was operated on at age 3 years after having suffered from otitis media, probably owing to the cleft palate.

On examination we observed peculiar craniofacies (Fig. 1), abundant, very thick, reddish, curly hair which was not easily combed. The hair appeared structurally normal when observed through a scanning microscope; its stress-strain features were not tested. The girl had a small face with maxillary hypoplasia and a small triangular mouth, lateral hypoplasia of the eyebrows; she had lost her eyelashes and suffered from epiphora caused by aplasia of the lacrimal canaliculi. Teeth were normal as far as number and age of eruption were concerned, but appeared small and grey (enamel hypoplasia). The nose was narrow with hypoplastic alae nasi and a bulbous tip. Finger and toenails were small, narrow, and dysplastic. Both feet had partial cutaneous syndactyly between second and third toes and clinodactyly of the third toes. No finger-tip ridges were visible; the dermatoglyphic examination showed generalised flattening of the crests which were also partially dissociated. A^c hypothenar patterns in both hands and a high axial triradius (t' left, t'' right) were also present. According to our evaluation the number of sweat pores on the surface of epidermal ridges was normal (c. 20/cm) although the features described made it difficult to perform an accurate count with a stereomicroscope.³ (According

Fig. 1 Affected mother and daughter; note the small mouth, narrow nose, lateral hypoplasia of the eyebrows, and the peculiar hair of the daughter (the mother has no hair and is wearing a wig).

to the patient's case history, sweating had always been normal.)

Growth was normal and followed the 20th centile of the Tanner-Whitehouse charts. Since the mother's height was 150 cm (3rd centile) and the father's 166 cm (10th centile), the girl was at the upper limit of her genetic potential. Skeletal maturity was evaluated according to the Tanner and Whitehouse method and corresponded to chronological age. Motor and mental development were normal, although when first examined (at 4½ years) she was found to have some delay in language development for which she received speech therapy. She appears to be intellectually normal.

The mother is now 36 and presents similar facial characteristics: high forehead, narrow nose, and small mouth. She too had a cleft palate and, although it has been operated on, she has retained a nasal voice. At age 16 years she lost her hair; in Fig. 1 she is wearing a wig. At present she is suffering from an extensive form of alopecia of the scalp with early signs of skin atrophy; follicles are no longer clearly visible and a residual crown of fine, dry, brittle hair can be observed in the temporo-occipital area. She also has sparse eyebrows and eyelashes. Her skin appears to be slightly atrophic with thin telangiectases on the areas not generally covered by clothes. Finger and toenails are dystrophic, affected by

Fig. 2 The mother's feet: note the partial cutaneous syndactyly (2nd-3rd toes) and the dystrophic nails.

pachyonychia and pterygium (Fig. 2). The characteristics of the dermatoglyphics are comparable with those of her daughter, although the flattening of epidermal ridges is more marked so that they are not always recognisable, which makes evaluation impossible. Hypodontia is also present.

Discussion

This mother and daughter closely resemble other reported cases, although the evidence does not suggest hypohidrosis; therefore if present it must be in a very light form, for it has never troubled them.

We do not think it possible to confirm the association of this syndrome as reported by Wannarachue *et al.*⁴ with growth deficiency. Although the mother is only 150 cm (3rd centile), she comes from a short but otherwise healthy family; the daughter's growth pattern, instead, follows the 20th centile and is at the upper limit of her genetic potential. The aetiology of the Rapp-Hodgkin syndrome is unknown. The mother is apparently the first case from unaffected parentage although it was not possible for us to examine any other members of the family. The expression of the syndrome seems to be the same in both sexes.⁴ Our report supports the hypothesis of autosomal dominant transmission, although X-linked inheritance cannot be excluded.

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Infantile aortic aneurysm complicating umbilical arterial catheterisation

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SUMMARY A true aneurysm of the lower thoracic aorta in a preterm infant dying of septicaemia at 14½ weeks is described. This is a rare complication of infection associated with umbilical arterial catheterisation and the first case in which *Pseudomonas aeruginosa* was the likely causative organism.

Mycotic aortic aneurysm in childhood was a well-known clinical entity in the days when bacterial endocarditis was more common than it is today.¹ Aortic aneurysms which present within the first 6 months of life are an even rarer phenomenon, but since 1976 five such cases have been reported each bearing a striking similarity to each other.²⁻⁵ All have occurred in preterm infants who have had umbilical arterial catheterisation complicated by a staphylococcal septicaemia. The present case differs from these in that a pseudomonas septicaemia appears to have produced a true rather than a false aortic aneurysm.

Case report

A girl was born at 26 weeks' gestation by normal vertex vaginal delivery after rupture of the membranes 2 weeks previously. She cried at birth but required intubation at 4 minutes for respiratory distress. An infection screen showed no evidence of intrauterine infection and prophylactic gentamicin and penicillin were given for 11 days. From day 1 to

26 an umbilical arterial catheter was *in situ* with the tip checked radiographically to be at the level of the 8th thoracic vertebra. Respiratory distress syndrome on the first day of life was complicated by interstitial emphysema secondary to high artificial ventilatory pressures. Pneumothorax was followed on day 4 by pneumonia, a haemothorax, and disseminated intravascular coagulopathy. Jaundice necessitated an exchange transfusion via an umbilical venous catheter which was in place from day 3 to 8. On day 5 the infant's general condition had worsened and *Pseudomonas* sp. and a coagulase-negative *Staphylococcus* sp. were cultured from the tip of the endotracheal suction catheter but not from blood. These organisms were again found on day 8 on the endotracheal tube and the venous catheter tip respectively. The pneumonia improved by day 9 when ticarcillin was added to the antibiotic regimen. The baby was extubated on day 11 but the pneumonia worsened. Chloramphenicol was given with some success but anaemia, apnoeic attacks, and episodes of bradycardia persisted. By day 21 the baby became septicaemic and was reventilated for a further 6 days. *Pseudomonas* sp., *Enterobacter* sp., and *Monilia* sp. were cultured from the blood on several occasions during the next few days, and *Pseudomonas* sp. was also found on the umbilical arterial tip and the endotracheal tube. The septicaemia was treated with ticarcillin, gentamicin, and an exchange transfusion. By day 26 there had been some improvement, the infant was extubated and the umbilical arterial