the ease with which glucose can diffuse into the pads of the test strips.

These observations show that if the PCV is greatly abnormal, the Dextrostix/Eyetone system (and to a lesser extent the Reflotest/Reflomat system) will give misleading results. This is mainly of relevance in polycythaemic patients, particularly in small-for-dates neonates⁴ who are routinely screened for hypoglycaemia and in whom a false diagnosis may be made if glucose concentrations are assessed only by a reagent strip method. Neonates with rhesus isoimmunisation are also screened for hypoglycaemia⁵ and since they can be severely anaemic a false diagnosis of normoglycaemia may be made.

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Tracheal compression by an anomalous innominate artery A report of 2 cases in a family

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SUMMARY The case histories of 2 brothers who presented with stridor soon after birth are described. In each, tracheal compression resulted from an anomalous innominate artery. We know of no previous familial reports of this condition.

The clinical entity of an anomalous innominate artery compressing the trachea and causing respiratory symptoms was first recognised by Gross and Neuhauser in 1948.¹ However, scepticism exists as to the clinical significance of this anomaly, as it can be an incidental finding in asymptomatic subjects. The 2 brothers presented here had had severe stridor since birth caused by compression of the trachea by an anomalous innominate artery, requiring surgery for its relief.

Case reports

Case 1. The patient, a boy, was born after a normal pregnancy to healthy and unrelated parents. Delivery was by assisted breech, and he weighed 3.35 kg at birth. He was asphyxiated with an Apgar score of 1 at one minute. Endo-tracheal intubation was carried out without any difficulty, and he was

given intermittent positive pressure ventilation for 5 minutes. Thereafter he breathed normally and was transferred to the nursery.

At 6 hours, he developed inspiratory stridor with cyanosis and subcostal recession. Laryngoscopy showed that the uvula, epiglottis, and vocal cords were red and slightly oedematous. This was attributed to previous intubation and he was allowed to settle in oxygen. Within hours, his condition deteriorated and endo-tracheal intubation was repeated to relieve respiratory distress. During the next 10 days, several trials of extubation resulted in recurrence of the symptoms. He was then transferred to the Royal Hospital for Sick Children in Edinburgh for further investigations. Clinical examination showed a small, scrawny infant with an endo-tracheal tube *in situ*. No other abnormalities were noted. Serum sodium, potassium, calcium and urea, blood gas tensions and pH, and a full blood count were normal. Bacteriological cultures from throat, umbilicus, and blood were negative. Routine chest x-ray film was normal, but lateral views of the neck during a barium swallow showed anterior indentation of the air-filled trachea at the level of T3 without associated compression of the oesophagus. Angiographic studies subsequently confirmed an anomalous origin of the innominate

artery, close to the origin of the left common carotid. This aberrant vessel passed anteriorly to the trachea and was thought to be responsible for the baby's symptoms. Because he required endo-tracheal intubation for relief of stridor and respiratory distress, a decision was made to relieve the compression surgically. At operation, the angiographic findings were confirmed. The aortic arch was approximated to the posterior aspect of the sternum, thereby lifting the innominate artery off the trachea. After this, the baby was extubated without recurrence of respiratory distress. However, he remained slightly stridulous especially during upper respiratory tract infections. Stridor finally disappeared after age 2 and at present, aged 6 years, his growth and development are normal.

Case 2. His brother was born 3 years later by spontaneous vertex delivery after an uneventful pregnancy. He weighed 3.22 kg and breathed spontaneously at birth. Stridor was obvious immediately and within hours he became cyanosed with sternal recession. Endo-tracheal intubation resulted in a striking relief of symptoms and he was transferred to this hospital for further management.

On examination he was a normal boy who breathed quietly through an oro-tracheal tube. Chest x-ray film, lateral neck x-ray film (with oro-tracheal tube *in situ*), and barium swallow were normal. A tracheogram (with Dionosil) showed extrinsic indentation of the right antero-lateral aspect of the trachea just above the carina. Angiography showed the same anatomy as in his brother, with the innominate artery arising to the left of mid-line, and coursing anteriorly to the trachea. At tracheoscopy, a pulsatile extraluminal mass was demonstrated on the anterior wall of the trachea. Pressure on this area with the bronchoscope occluded

the right brachial pulse, and provided further confirmation of the diagnosis.²

The same operative procedure was carried out but relief of symptoms was not as striking as in the brother. He remained mildly stridulous during sleep and was admitted to hospital on three occasions when symptoms were exacerbated by upper respiratory tract infections. Now aged 3, he has an occasional barking cough, but is developing and thriving satisfactorily.

Discussion

The innominate artery normally arises from the aortic arch directly over the trachea, about 1 cm above the carina. If it arises too far to the left or too posteriorly, it has to pass the anterior surface of the trachea as it comes upwards to the apex of the right hemithorax. A long and flabby vessel may not cause symptoms, but compression of the anterior trachea will occur if the vessel is short and tight.

The incidence of innominate artery compression of the trachea is not known. In a review of 104 cases of vascular anomalies causing tracheal compression, Fearon and Shortreed² found that 66% of them were due to an anomalous innominate artery. Mustard *et al.*³ have reported on 16 years' experience of tracheal compression by vascular anomalies in children and concluded that an anomalous innominate artery is 'the commonest of all the vascular anomalies causing tracheal obstruction'. Symptoms resulting from the anomaly are non-specific, and because of other more common causes of stridor in infancy, the diagnosis of an anomalous innominate artery comes low on the list. In her review of vascular rings causing tracheobronchial compression in infants, Keith⁴ stated that newborn babies may not be severely symptomatic, and that symptoms became worse in the first 2 months of life. The condition should also be considered in infants with recurrent or chronic respiratory problems, and in one study of 100 infants less than 18 months old with bronchopneumonia, no fewer than 6% were found to have this anomaly.⁵

Both children described above were treated surgically for relief of respiratory distress, and at operation anatomical findings were identical. It is interesting to note that since early infancy, their father had been noted to make 'squeaky noises' when asleep. He was said to have been chronically unwell as a child with recurrent bronchitis and fever. Now, aged 32, he has occasional inspiratory stridor with upper respiratory tract infections. In view of the mild nature of his symptoms he was not investigated fully but chest and lateral neck x-ray films were normal. Were he shown to have the same condition,

Figure Diagram of the anatomy in the 2 brothers.

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.

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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