anxiety generated among medical attendants and parents.

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Dr Leonard Arthur died on Christmas Day 1983.

Factor V deficiency and antenatal intraventricular haemorrhage

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SUMMARY Serial obstetric ultrasound showed the development of asymmetrical ventricular dilatation between 28 and 32 weeks' gestation. After delivery at 38 weeks, progressive ventricular dilatation required a ventriculo-atrial shunt. Investigation of postoperative bleeding into the cerebral ventricles consistently showed factor V values of only two per cent.

Periventricular haemorrhage commonly occurs postnatally between 26 and 30 weeks' gestation but is much less common antenatally. Although respiratory failure and its complications seem to be the most important causes of postnatal periventricular haemorrhage, circumstances leading to antenatal haemorrhage have been difficult to analyse. Thrombocytopenia (due to maternal platelet antibodies) has been reported,¹ and we have observed hydrocephalus after maternal warfarin treatment. Inherited coagulation disorders have not hitherto been associated with antenatal haemorrhage, although they have been associated with this postnatally.^{2,3} The two most serious consequences of periventricular haemorrhage are parenchymal extension with liquefaction of cerebral tissue and hydrocephalus from obstruction in the fourth ventricle or basal cisterns.⁴ We report a girl with congenital factor V deficiency who showed evidence of antenatal periventricular haemorrhage with subsequent hydrocephalus and recurrent intraventricular bleeding.

Case report

This girl is the second child of non-consanguineous Sikh parents who have a healthy 2 year old son. There is no family history of neurological or haematological disease. The pregnancy was monitored with ultrasound. Fetal head growth was near the 50th centile until 32 weeks' gestation, when acceleration in the biparietal diameter above the 90th centile and right ventricular dilatation were noted (Fig. 1). Spontaneous labour began at 37 weeks' gestation and caesarean section was carried out because of breech presentation. The girl weighed 2.6 kg (above 10th centile) and her head circumference was 34.3 cm (above 90th centile).

After a short seizure at the age of 10 days, she was admitted to Hammersmith Hospital. Head control was impaired and visual following poor. Investigation showed a normal haemoglobin concentration and platelet count and negative serology for cytomegalovirus and toxoplasmosis. Ultrasound scan confirmed the gross right ventricular dilatation, with mild dilatation of the left ventricle. Right ventricular puncture showed xanthochromic fluid with a resting pressure of 15 mm Hg, sterile on culture. Her head circumference continued to grow along the 90th centile for the next three weeks but at the age of 6 weeks growth accelerated, she became drowsy, fed poorly, and a ventriculo-atrial drainage system was inserted (Mr Pal Singh). Excessive haemorrhage was noted during surgery and over the following week the scalp wound developed a 20 ml

Fig. 1 Antenatal ultrasound scan at 32 weeks' gestation (axial view) showing dilated right ventricle.

haematoma. The ventricular catheter became blocked with blood and had to be replaced after 10 days. Head control improved but the patient failed to fix visually. At the age of four months she was readmitted to hospital with drowsiness, vomiting, and a bulging fontanelle. At operation the ventricular catheter and the valve were found to be blocked with blood clot. Two days after operation she had pallor and tachycardia. Her haemoglobin concentration had fallen from 11.3 g/dl to 4.5 g/dl, with a raised platelet count ($516 \times 10^9/l$).

Prothrombin time was prolonged at 61 seconds (normal 13 seconds); partial thromboplastin time was also prolonged at 178 seconds (normal <40 seconds) but thrombin time was within the normal range at 16 seconds and plasma fibrinogen was also normal at 2.34 g/l. She was transfused with packed red cells and 20 ml/kg of fresh frozen plasma. Cranial ultrasound scan two days after operation showed some dilatation of the posterior horn of the right lateral ventricle with a large blood clot in the lumen of the ventricle (Fig. 2).

Repeated sampling has confirmed very low factor V concentrations of two per cent.⁵ The child's parents are probably heterozygous with variable penetrance (maternal factor V concentration 52%, paternal 78%) while her brother is apparently normal with a factor V value of 100%. Our patient was treated with daily fresh frozen plasma for two weeks after operation. Unfortunately, one month after the second surgical revision she presented with the ventricular catheter blocked once again by blood

Fig. 2 Right parasagittal ultrasound scan at 4 months postnatal age, two days after shunt revision showing dilatation of the occipital horn and a round echogenic blood clot in the lumen of the ventricle.

clot and has required another revision. She is now being treated with fresh frozen plasma twice a week.

Discussion

The timing of the fetal head enlargement and appearance of right ventricular dilatation and subsequent hydrocephalus all suggest a periventricular haemorrhage at between 28 and 30 weeks' gestation with involvement of the right parenchyma and subsequent cerebral liquefaction. The asymmetrical ventricular dilatation subsequently found is identical to that we have observed many times with postnatal periventricular haemorrhage. In factor V deficiency clotting occurs extremely slowly but does eventually form.⁵ Thus, if a small haemorrhage started in the germinal matrix, factor V deficiency would allow the bleeding to continue for longer than usual. Having spread through the ventricular system, the blood would eventually clot, hence the hydrocephalus. After shunt surgery, a similar problem occurs with the slightest movement of the ventricular catheter encouraging bleeding which continues but then eventually clots in the catheter, blocking the shunt system. It is hoped that regular fresh frozen plasma will reduce the risk of both bleeding and the resultant blockages.

Factor V deficiency is a rare disorder inherited as an autosomal recessive gene. Clinical bleeding problems have been reported as less severe than in

many haemophiliacs and are largely confined to perioperative bleeding.⁶ Our patient is the first reported case of factor V deficiency with antenatal intracranial haemorrhage and hydrocephalus. In view of this and the reports of perinatal intracranial haemorrhage in factor VII³ and factor VIII deficiencies,² it is suggested that in infants with evidence of antenatal periventricular haemorrhage and hydrocephalus or postnatal periventricular haemorrhage without respiratory failure prothrombin time, partial thromboplastin time, thrombin time, and platelet count should be measured to identify any potentially treatable underlying disorders.

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Correction

Archives of Disease in Childhood 1984;**59**:525. *Enterochromaffin cells in coeliac disease*

In the **Results** section: Line 10—'lower' should read 'higher'. Line 14—'higher' should read 'lower'