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IMAGES IN NEONATAL MEDICINE.....

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Unusual presentation of neonatal haemophilia A

Figure 1 Widespread purple discoloration in a baby with severe haemophilia A. Parental/guardian informed consent was obtained for publication of this figure.

A second male twin was born at 36 weeks' gestation by emergency caesarian section owing to failure to progress following spontaneous onset of labour. He was difficult to deliver because of a transverse lie. He developed respiratory distress shortly after birth, requiring oxygen and antibiotics, and became lethargic, hypothermic (temperature 36.1°C) and tachycardic (174 beats/min) at 48 h of age. A large purple lesion was noted on his back (fig 1). His activated partial thromboplastin time was 186 s, reducing to 30.3 s with a 50% mix of normal plasma. Factor studies confirmed severe haemophilia A with a factor VIII:c level of <0.05 IU/ml. Haemoglobin dropped from 196 g/l to 97 g/l 48 h after birth. He required transfusion, factor VIII and phototherapy for jaundice. Cranial and abdominal ultrasound scans excluded other haemorrhages. His twin brother remained well, but screening revealed he was also affected.

In the absence of a family history, diagnosis of VIII deficiency is usually made following a bleeding episode, 18–54% of which occur within the first month of life.^{1–3} Bleeding sites include cephalohaematomas (particularly after ventouse deliveries), puncture sites from venesection or intramuscular drug administration, after circumcision, umbilical stump bleeding, intracranial haemorrhages and bleeds into the gastrointestinal tract or other major organs.^{1–3} Massive intradermal haemorrhage in the immediate newborn period has not been previously reported, although extensive bruises are a common method of presentation in older children. We assume this was related to the baby's difficult extraction at caesarean section.

Anthony R Hart, C Mae Wong, Alan T Gibson
Neonatal Unit, Sheffield Teaching Hospital Trust, Sheffield, UK

Correspondence to: Dr Anthony Hart, Neonatal Intensive Care Unit, Jessop Wing, Sheffield Teaching Hospitals NHS Foundation Trust, Tree Root Walk, Sheffield S10 2SF, UK; t.hart@doctors.org.uk

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