Intra-Hepatic Haemorrhage and Shock during Post-Natal Period, in Two Brothers with Haemophilia

Abstract:
Sir
Haemophilia rarely presents with severe visceral haemorrhage in the first week of life. A 15 hour old boy presented with haemorrhagic shock and coagulopathy. He was diagnosed with liver haemorrhage and severe haemophilia A. His younger brother, diagnosed with haemophilia at birth, presented with shock due to liver haemorrhage at 79 hours.

Case Reports
A boy, born at term by vaginal delivery, developed coagulopathy and hypovolaemic shock at 15 hours. pH was 6.66, lactate 21 mmol/l, haemoglobin 8.9 g/dl, platelets 178x10⁹/l, APTT 245 seconds, PT 21 seconds, fibrinogen 0.9 g/l. He was ventilated and received O-negative blood, plasma, vitamin K, antibiotics and inotropic support. He responded to treatment and was extubated after eight hours. Coagulation normalised but his required further blood and platelet transfusions. He developed abdominal distention with hepatomegaly. Ultrasound images suggested a hepatic tumour with haemorrhage into the tumour and the peritoneal cavity (Figure 1). He required further blood and platelets transfusions. By day thirteen, the ultrasound appearances were consistent with a resolving hematoma. Repeated coagulation screens showed prolonged APTT. Factor VIII level of <0.01 IU/ml confirmed a diagnosis of haemophilia A.

Two years later a brother was born at term by vaginal delivery. Factor VIII level was <0.01 IU/ml at birth. At 79 hours, he developed abdominal distention, hepatomegaly and haemorrhagic shock with haemoglobin 6.8 g/dl, pH 7.08, lactate 18 mmol/l and an unrecordable APTT. He was ventilated, and received O-negative blood, recombinant factor VIII and intravenous vitamin K. Ultrasound showed a hematoma in the right lobe of the liver with intraperitoneal haemorrhage. He responded to treatment and was extubated after five hours. Neither infant had evidence of intracranial bleeding.

Discussion
Coagulation factors are synthesised in the foetus from 10 weeks gestation. While levels of some coagulation factors are low at birth, factor VIII level is within the adult range. Sepsis or ischaemia can derange coagulation. Nevertheless, factor VIII or IX deficiency should be suspected in any newborn with an isolated or disproportionately prolonged APTT. Haemophilia rarely presents with severe haemorrhage in neonates. When the family history is known, the diagnosis is made on cord blood. Between 37% and 68% of patients are diagnosed in the first month of life, with the higher presentation rate associated with circumcision. The commonest presentation is intracranial or extracranial haemorrhage (41%). The remainder of cases present with bleeding from puncture sites, umbilical cord, or following circumcision. Only 2.5% of cases present with haemorrhages into visceral organs, usually the spleen. In the three described cases of liver haemorrhage in neonates, symptoms developed 11 to 72 hours after vaginal delivery at term. We report the first known cases of haemophilia presenting as intrahepatic haemorrhage in the neonatal period in brothers.

The rapid correction of the coagulopathy, deranged liver function indices and the imaging suggestive of a liver tumour delayed diagnosis in the first case. Hepatic haematomas can be difficult to differentiate from other lesions with ultrasound and CT. Our experience demonstrated the importance of considering alternative diagnoses in infants presenting with unusual clinical and radiological findings.

T Nabialek¹, R Pinnamaneni¹, MS Saleemi¹, B Nolan², JD Corcoran¹
¹Department of Paediatrics, Rotunda Hospital, Parnell Street, Dublin 1
²Department of Haematology, Our Lady’s Children’s Hospital, Crumlin, Dublin 12
Email: tomasz.nabialek@gmail.com

References