CASE REPORT

Recognising serious umbilical cord anomalies

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SUMMARY

Umbilical vessel catheterisation is a common intervention in neonatal care. Many complications are recognised, some of which are life-threatening. We report the case of a term neonate who was compromised at birth following antepartum haemorrhage with evidence of multiorgan ischaemic injury. Following resuscitation and umbilical vessel catheterisation, she developed pneumoperitoneum. At laparotomy, a patent vitellointestinal duct was identified and resected. Intestinal perforation was found in the duct wall, most plausibly explained by the unintentional catheterisation of the duct via the umbilicus. Learning to recognise umbilical cord anomalies, such as patent vitellointestinal duct, can be simple and could prevent potentially serious complications.

BACKGROUND

Vascular access is frequently gained via the umbilicus in neonates, usually on neonatal units and in paediatric and emergency departments. Indeed, learning to insert umbilical catheters is deemed an essential part of junior paediatric/neonatal training, as well as that of the Advanced Paediatric Life Support (APLS) course, which is undertaken by clinicians from many disciplines, including anaesthetics and paediatric surgery.^{1 2} Umbilical cord *examination* is usually performed by midwifery and obstetric teams.³ The case presented here is not only a reminder of the serious complications of umbilical catheterisation, but also highlights the importance of the opportunity to examine umbilical cord anatomy during the procedure. Neither of these lessons is specifically covered by the standard Royal College of Paediatrics and Child Health (RCPCH) or APLS training.

CASE PRESENTATION

A full-term baby girl was born by spontaneous vaginal delivery (birth weight 2600 g). She had asymmetrical growth restriction of unknown cause. Pregnancy had been complicated by the intrauterine death of a twin fetus at 12 weeks' gestation. During labour there was an unexpected fetal bradycardia. Forty minutes later, the baby was delivered with signs of antepartum haemorrhage: a rush of blood, and a very short umbilical cord which appeared to be detaching from the placenta. She was floppy and pale. Her heart rate was initially 60 bpm, and she required resuscitation with inflation and ventilation breaths until the heart rate rose above 100 bpm by 3 min of age. Her respiratory effort improved and she was transferred to the neonatal unit with continuous positive airway pressure

(CPAP) respiratory support. Apgar scores were 1, 4 and 5 at 1, 5 and 10 min, respectively. Blood could not be obtained from the umbilical cord for blood gas analysis.

On admission to the neonatal unit, the infant remained pale and a peripheral venous gas demonstrated anaemia (haemoglobin 5.4 g/dL), almost certainly due to the antepartum haemorrhage, with a severe, mixed acidosis (pH 6.9). Non-invasive blood pressure measurements were low (mean arterial pressure 24–34 mm Hg), but responded to boluses of saline and blood, via a peripheral cannula. Clinical criteria for therapeutic cooling were not met, as a good suck and spontaneous limb movements were observed, with no clinical seizure activity. During the initial peripheral blood transfusion, insertion of an umbilical arterial catheter (UAC) and an umbilical venous catheter (UVC) was attempted for secure access and monitoring. When the cord was cut near the base, a 'pop' was heard and a small cleft was found between the vein and arteries. This was carefully examined but not identified as abnormal anatomy. Umbilical arterial access could not be obtained, despite several attempts, and the procedure was abandoned. The UVC was easily inserted on the first pass, at approximately 2 h of age. Maintenance fluids were administered peripherally. Cefotaxime was given. Orogastric tube aspirates contained altered blood, presumed to have been swallowed at birth. Nasal CPAP was continued for 6 h before the infant could self-ventilate in air with a normal CO₂.

Eight hours after umbilical catheterisation, the abdomen became distended, with sparse bowel sounds, but was neither tense nor tender. Meconium had been passed. No enteral feed had been given, though a nasogastric tube was in situ. Pneumoperitoneum was suspected and the UVC, assumed to be responsible, was removed immediately.

INVESTIGATIONS

Immediately after placement, correct positioning of the UVC had been confirmed radiologically (figure 1). Chest X-ray showed mild changes of respiratory distress syndrome. Cranial ultrasound scan was normal. Blood results demonstrated renal impairment (creatinine 151 µmol/L on day 0).

Once pneumoperitoneum was suspected, an abdominal X-ray showed extensive free air (figure 2) and a review of the initial radiograph (figure 1) revealed that a small pneumoperitoneum had previously been present.

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Figure 1 Plain X-ray to confirm the position of umbilical venous catheter. Rigler's (double wall) sign was later noted in the left iliac fossa (arrow).

DIFFERENTIAL DIAGNOSIS

Possible reasons for the pneumoperitoneum were considered at the time, including spontaneous perforation due to ischaemic bowel/necrotising enterocolitis, iatrogenic bowel perforation via the nasogastric tube or perforation through the wall of the umbilical vein into the peritoneal cavity (another recognised complication of umbilical catheterisation), which prompted the urgent removal of the UVC. Spontaneous perforation would be unusual in a term infant. However, this baby's suspected ischaemic injury would have increased the odds of both spontaneous as well as iatrogenic perforation. Although further imaging could have helped differentiate the cause, a laparotomy would likely be needed for treatment. Therefore a diagnostic and

Figure 2 Eight hours after umbilical catheterisation, significant pneumoperitoneum was present, including air visible under the diaphragm (arrow).

potentially therapeutic laparotomy was the more efficient option.

The initial unusual appearance of the cord stump was recalled at this point, but again not recognised as abnormal anatomy which might have predisposed to the true pathology as found in the denouement of this case.

TREATMENT

The baby began vomiting altered blood and the nasogastric tube was placed on free drainage. Metronidazole was given and the baby was transferred at 14 h of age to the nearest tertiary neonatal surgical centre. At laparotomy 3 h later, macroscopically normal bowel was found, with a patent vitellointestinal duct (VID) attached to the umbilicus. A discrete perforation was noted in the wall of the duct, but no other associated anomaly. The duct and 3 cm of ileum were resected with end-to-end ileal anastomosis. Histology of the perforated duct and ileum demonstrated flattened mucosa with loss of villi and normal glands, as well as some subepithelial fibrin with haemorrhage. This is consistent with ischaemic injury antecedent to the perforation.

OUTCOME AND FOLLOW-UP

Postoperatively, our patient was admitted to the paediatric intensive care unit. She required mechanical ventilatory support for 6 days following surgery. Deranged renal function and electrolyte imbalances were conservatively managed with appropriate fluid and electrolyte supplements. Serum creatinine normalised by day 7 of life.

On days 3 and 4 of life, the baby had worsening abdominal distension, large nasogastric aspirates and radiological evidence of necrotising enterocolitis (figure 3). This was managed conservatively with intravenous antibiotics and total parental nutrition. Enteral feeds were introduced on day 13, and gradually increased until full enteral feeding was attained by day 17 of life. The infant was discharged home on day 22 following full recovery. She has been followed up until 12 months of age, and there were initial concerns about constipation and gastro-

Figure 3 On day 5, mottled changes were seen in the right upper quadrant (arrow), suggestive of necrotising enterocolitis.

oesophageal reflux. Her developmental progress now appears satisfactory apart from a tendency to abduct her left thumb, for which she is receiving ongoing physiotherapy.

DISCUSSION

In this case, we propose that the perinatal combination of anaemia, hypoxia and hypotension caused ischaemic bowel, friable enough to be perforated by a UAC misplaced down the anomalous patent VID during attempted insertion.

The VID connects the yolk sac to the gut in the embryo, providing nutrition until the placenta is established. This connection normally involutes from the terminal part of the ileum during the 5th–9th weeks of intrauterine life. Incomplete involution leads to a variety of anomalies, occurring in 2–3% of children.⁴ Meckel's diverticulum is the commonest variant, while patent VID is the rarest, representing approximately 5% of all VID remnants.⁵ Others include a diverticulum associated with a persistent cord to the umbilicus, a solid cord or a cord with a central cyst. VID remnants are usually asymptomatic but can present with gastrointestinal bleeding, intussusception, pain, perforation, obstruction, volvulus, umbilical discharge or a polyp.⁴ A completely patent VID is a fistulous connection between the ileum and umbilicus. It can regress spontaneously but usually presents in the neonatal period with persistent umbilical discharge of intestinal contents.

Umbilical catheterisation can be complicated by infection, ischaemic and thromboembolic events (affecting various organs), haemorrhage, hypoglycaemia, extravasation, peritonitis, hepatic laceration, cardiac tamponade, arrhythmias or pulmonary infarction.⁶ Reported in the literature are several cases of umbilical catheterisation causing direct peritoneal perforation or urachal/bladder perforation/injury. However, we found only one case similar to that which we report in which a UVC perforated a Meckel's diverticulum via an anomalous umbilical connection (a VID cord-type remnant).⁷

Before umbilical catheterisation, careful consideration of risks and benefits is fundamental. Parental consent for this procedure is usually deemed implicit but this “does not obviate the need for the clinician to explain its purpose and if appropriate to explain any risk,” either preinsertion or postinsertion.⁸ All healthcare professionals performing umbilical catheterisation should be trained to recognise abnormal umbilical cord anatomy, such as patent urachus and VID.⁹ This could be incorporated into newborn resuscitation training (such as the Advanced Life Support Group's (ALSG's) Newborn Life Support course). If an anomalous umbilical duct is identified, accidental catheterisation and potential perforation may be prevented, as well as the complications of the duct itself. Additional diagnostic information may be obtained by examining the

‘placental end’ of the umbilical cord. Ultrasonography and/or contrast studies may then characterise the umbilical defect prior to timely intervention to minimise complications and avoid unnecessary patient morbidity.

Learning points

- ▶ When inserting umbilical vessel catheters, the potential risks can be life-threatening, and may sometimes outweigh the potential benefits.
- ▶ Umbilical cords should be examined at birth and during catheterisation.
- ▶ Those healthcare professionals performing these tasks should recognise and understand the significance of umbilical cord anomalies which can be identified before complications arise.
- ▶ Those teaching others to insert umbilical catheters should reinforce the above messages as they are not covered by other standard training.

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