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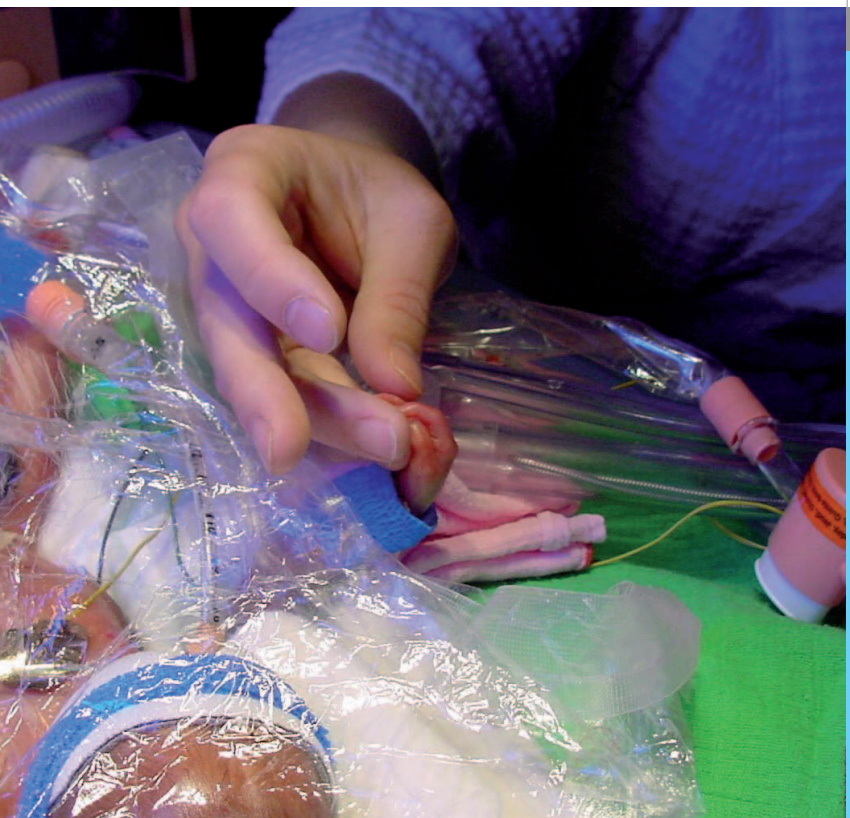
Catastrophic gastrointestinal event in a 10-week-old extremely growth restricted preterm infant



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The mother of this female infant was transferred to our institution at 25 5/7 weeks of gestation because of intrauterine growth restriction after an otherwise unremarkable pregnancy. The healthy 22-year-old mother had no risk factors for a placental insufficiency or preeclampsia, apart from smoking 6-8 cigarettes per day. Amniocentesis revealed a normal female karyotype (46, XX).

Following induction of fetal lung maturation, the girl was born by cesarean section at 26 1/7 weeks of gestation because of non-reassuring fetal monitoring and pathological Doppler flow studies of the middle cerebral artery as well as the umbilical vessels. The infant adapted reasonably well with Apgar scores of 4 and 6 at 1 and 5 minutes, respectively. The arterial umbilical cord pH was 7.21. Her body weight was 360 g (160 g below the 3rd percentile). Because of respiratory distress she was intubated at the age of 9 minutes. After surfactant administration (Curosurf®) her oxygen saturation rose from 68% to 98% and the FiO_2 could be reduced to 0.4.

Initial hemodynamic instability resolved within 24 hours and her respiratory status stabilized. Severe persistent neutropenia (neutrophil count of 0.1 G/l) was successfully treated with three doses of GCSF (10 mcg/kg/day). Enteral nutrition could only be introduced very slowly because of ongoing feeding intolerance. She needed multiple rectal irrigations un-

til she passed her first meconium. At two weeks of age, she received 70 ml/kg/day of enteral nutrition and it took another two weeks for her to be advanced to full enteral feeds (160 ml/kg/day.) Full fortification was achieved two days later. At that time (30 2/7 weeks corrected age), her weight was 600 g.

On day of life (DOL) 10, she had an episode of a catheter-associated bacteremia with coagulase-negative staphylococci which was treated with a 7-day-course of vancomycin. On DOL 19, she failed a first extubation attempt after only 8 hours because of recurrent apnea. A second attempt on DOL 30 was successful. Cerebral ultrasound examinations remained normal.

At the corrected age of 35 weeks she was transferred to our intermediate care unit. At that time, she was on room air and required CPAP support for a few hours per day. She was on full enteral feeds and started to drink from the bottle (video). Gentle abdominal massage was used intermittently to facilitate bowel movements.

One week later (DOL 72), there was a sudden dramatic deterioration with vomiting, apnea, and an increased oxygen requirement. She was rapidly intubated. Her blood pressure was initially not measurable and increased to a mean of only 20 mmHg after a first bolus of normal saline. Two hours after the first symptoms had been noted, she had a pH 6.7, a pCO_2 of 6 kPa, a bi-

carbonate concentration of 5 mmol/l and a BE of -25 mmol/l. The serum lactate level was 25 mmol/l. The hemoglobin had fallen to 55 g/l (one week earlier her hemoglobin concentration had been 96 g/l). Her platelet count was normal. She was started on dopamine (20 mcg/kg/min) and epinephrine (0.25 mcg/kg/min) and she was transfused with packed red blood cells (20 ml/kg). Despite these measures, her blood pressure remained low. Echocardiography showed good biventricular function but there were signs of hypovolemia. Cerebral ultrasound was normal. Her abdomen became progressively distended (Fig. 1). On abdominal X-ray, there were enlarged bowel loops but no pneumatosis and no evidence of free air was seen (Fig. 2). On ultrasound, the bowel walls appeared thickened and there was very poor intestinal perfusion with a resistance index > 1 (Fig. 3).

Exploratory laparotomy was not considered to be a reasonable option in the presence of refractory hypotension, severe metabolic acidosis and profound coagulopathy. The multidisciplinary team informed the parents of the futility of further live prolonging therapies, and redirection of care was agreed upon. She died in the arms of her parents a few minutes after extubation.

At autopsy, there was extensive hemorrhagic infarction of the bowel involving the jejunum (2.5 cm distal of the ligament of Treitz), the ileum and the ascen-

ding colon (Fig. 4). On histopathology there was severe acute congestion with hemorrhagic infarction of the inner layers of the intestine (Fig. 5, 6) without evidence of inflammation or peritonitis. Histology of liver and heart were normal but the kidneys showed acute tubular necrosis (Fig. 7). These findings were felt to be consistent with hemorrhagic infarction in the territory of the superior mesenteric vein and artery. Unfortunately, no obvious cause could be identified: there was no evidence vascular thrombosis, no volvulus, no intussusception and no evidence of right heart failure.

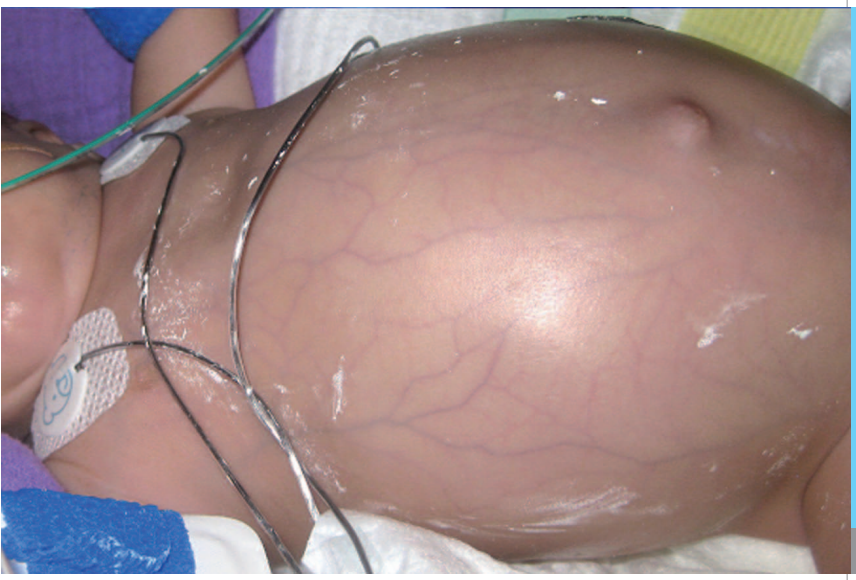

Fig. 1

During the patient's clinical deterioration, the abdomen rapidly became very distended.

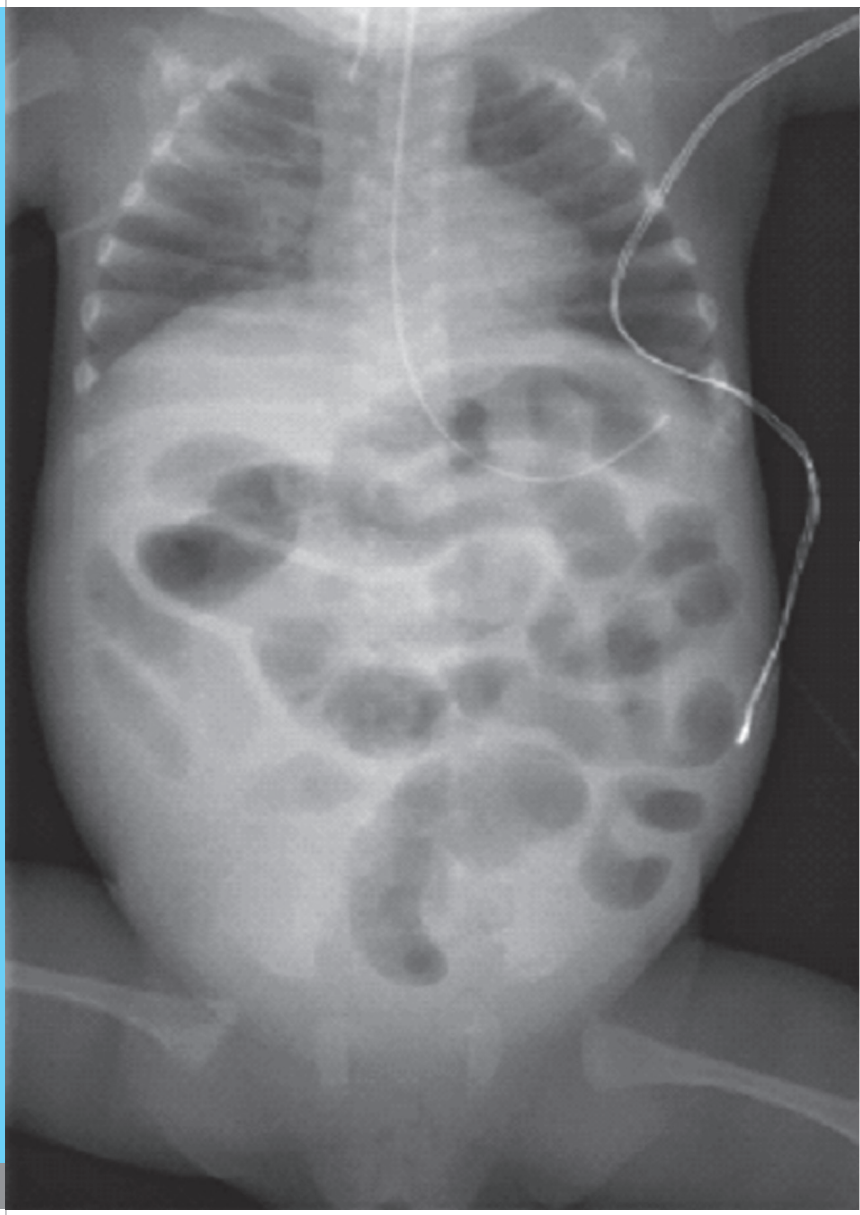

Fig. 2

Babygram showing distended and thickened bowel loops without signs of pneumatosis or bowel perforation.

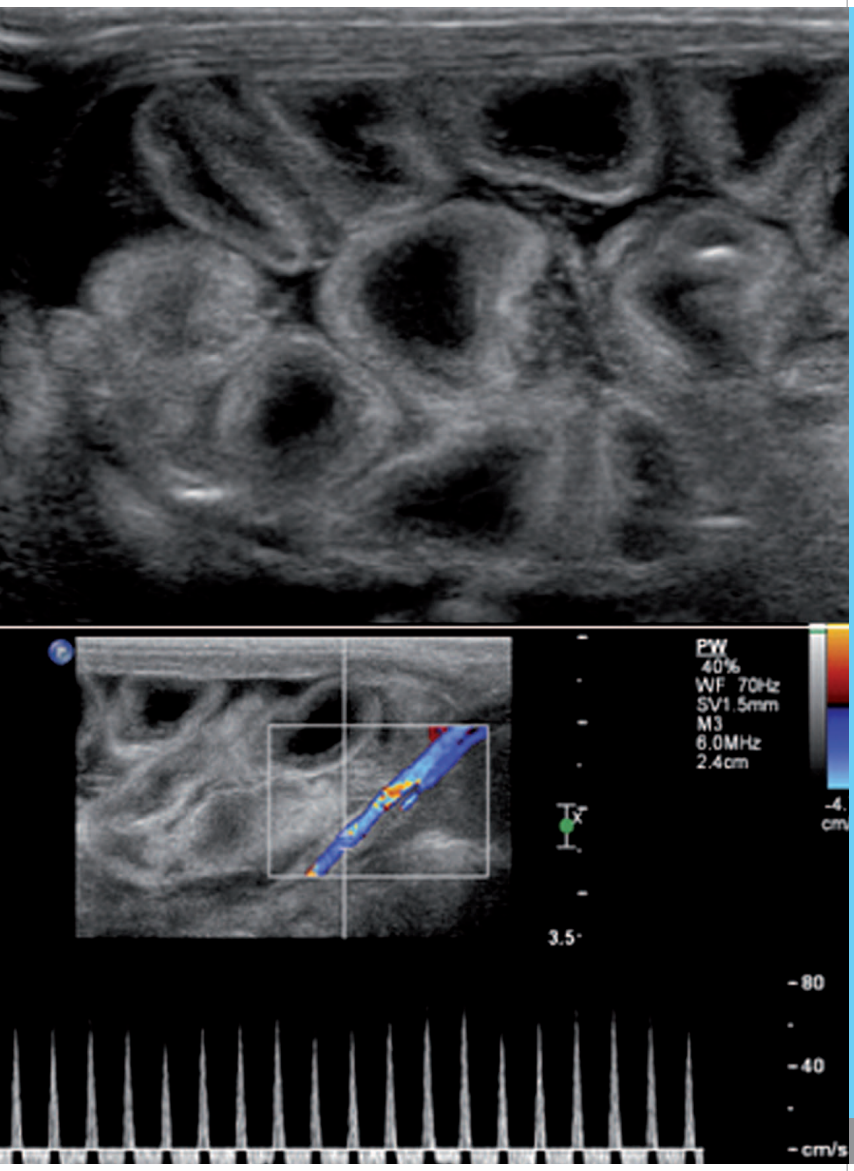

Fig. 3

Abdominal ultrasound examination demonstrating thickened bowel loops (upper panel) with poor perfusion (note reverse diastolic blood flow, bottom panel).

Fig. 4

Abdominal situs at autopsy: dilated and congested bowel loops.

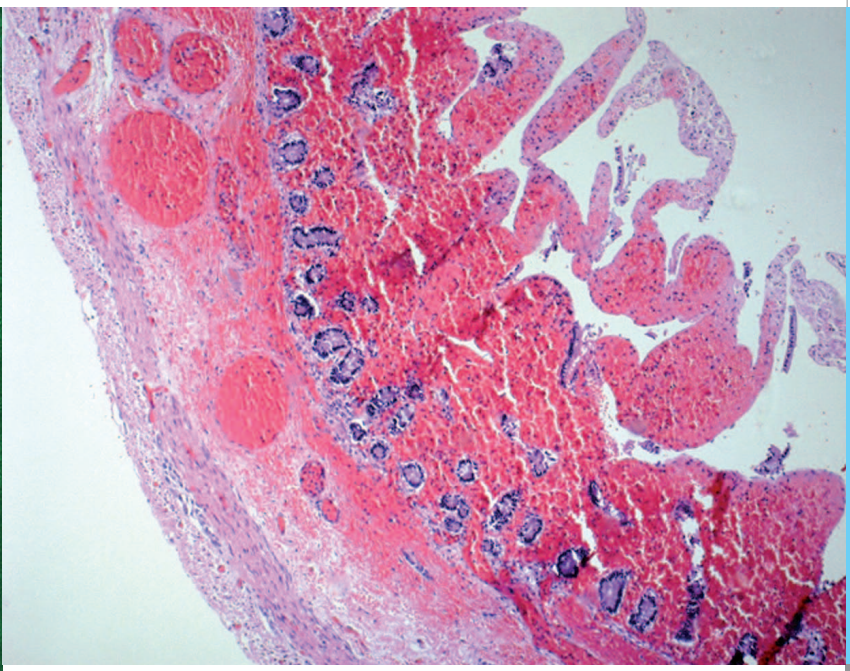

Fig. 5

Histopathology (low power view): severe acute congestion and hemorrhagic infarction of the jejunum, ileum and ascending colon.

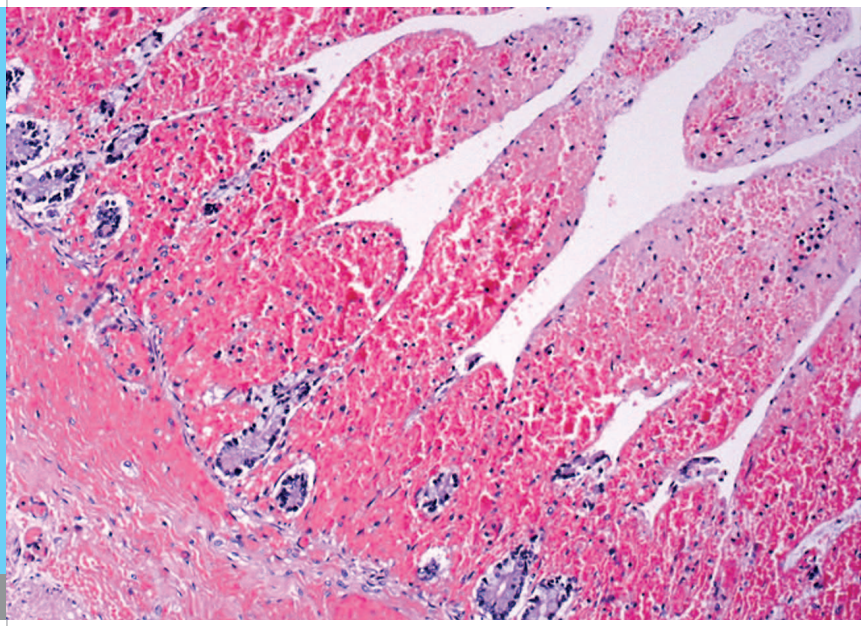

Fig. 6

Histopathology (high power view): severe acute congestion and hemorrhagic infarction of the jejunum, ileum and ascending colon.

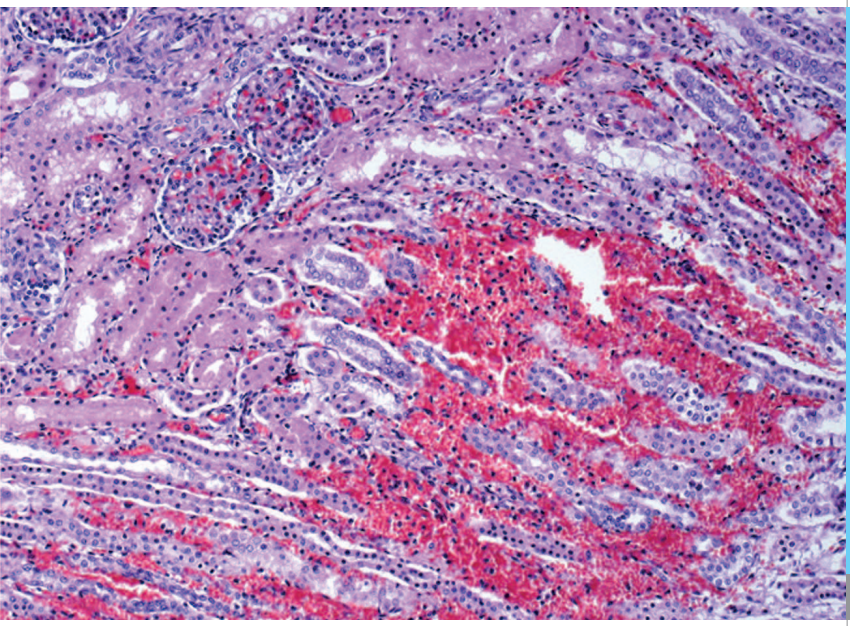

Fig. 7

Histopathology: kidney with signs of acute tubular necrosis.

DISCUSSION

At 24 weeks of gestation, it was felt unreasonable to deliver this patient because of her extreme growth restriction and the possibility of intrauterine fetal demise was accepted. However, when she reached 26 weeks of gestation, active intervention was agreed upon despite her extremely low birth weight. She had very few cardiorespiratory problems and ongoing intensive care measures seemed ethically justified.

Feeding intolerance is not uncommon in growth restricted preterm infants. In this group of patients, higher rates of necrotizing enterocolitis (NEC) (1, 2) and focal intestinal perforations (FIP) (3) have been described. Because of the increased risk of intestinal complications, her enteral nutrition with mother's milk was advanced only very slowly and she eventually tolerated fully fortified feedings at the age of one month. At that point, the risk for NEC or FIP seemed to have become minimal.

Six weeks later, a different catastrophic gastrointestinal event led to her death: massive hemorrhagic infarction of the small bowel and the ascending colon. Even after autopsy, the pathophysiological mechanism which led to intestinal hemorrhagic infarction in the territory of the superior mesenteric vein remained unexplained. There was no evidence of a thrombotic or a thromboembolic event. We therefore speculate that a spontaneously resolving volvulus may have caused transient occlusion of the superior mesenteric vein.

The clinical findings of the abdominal Doppler study (Fig. 3), echocardiography (evidence of hypovolemia) and rapidly developing lactic acidosis would be consistent with this assumption. Abdominal massage has previously been described as a risk factor for intestinal volvulus in preterm infants (4) and a small case series has been presented in the December 2006 COTM (5). This practice should probably be abandoned in the NICU.

ACKNOWLEDGEMET

We are grateful to the parents for allowing us to use a video tape of the patient.

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