

Umbilical hernia associated with extracelomic intestinal atresia and perforation of the ileum in a newborn

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The umbilicus is the site of various embryological defects involving the midgut (herniation), abdominal wall, vessels and urachus. Hernia of the umbilical cord has been recognized as a distinct entity occurring at a specific embryological step.¹ Occasionally it is associated with patent vitello-intestinal duct (PVID) anomaly.² We report the first case of umbilical hernia in a neonate associated with ileal atresia complicated by intestinal perforation, meconium staining of liquor and meconium aspiration syndrome.

CASE

A male baby weighing 1750 g at 33 weeks gestation was born to a 28-year-old mother (G3P2) with meconium-stained liquor. The baby had meconium aspiration syndrome for which he received ventilatory support in the level 3 nursery at Maternity and Children's Hospital. The baby was found to have a discharge of meconium from an opening in the gut protruding from the umbilical ring into the cord. There were black-colored tubular structures present within the cord close to the umbilical clamp (Figure 1). The baby was active and vitals were maintained. No other gross congenital anomalies were detected. Considering possible iatrogenic gut injury due to umbilical clamp, an urgent exploration was carried out under general anesthesia. Intraoperatively, a type 3A atresia of the distal ileum (Figure 2) was found in the extracelomic gut (in the umbilical cord). Approximately 20 centimeters of proximal ileum was grossly dilated with perforation at the end. The distal segment was hypoplastic with a gap in the mesentery. Saline irrigation into the distal segment revealed normal patency of the rest of the gut and strings of white mucus. The black, shriveled tubular structures between the two ends of the atresia were excised and sent for histopathological examination. The dilated segment of the ileum was excised and an ileo-ileal anastomosis was done. The post-

operative course was uneventful. At 5 months follow up the baby was doing well. Histopathology reported the black tubular tissues as atrophic intestine.

DISCUSSION

A varied number of anomalies affect the umbilicus. Umbilical cord hernia is a distinct but poorly defined anomaly where the midgut is found herniating into the substance of the umbilical cord. Unlike omphalocele and gastroschisis, there is no deficiency of the anterior abdominal wall. Also there is an absence of association with congenital anomalies, namely cardiac, chromosomal anomalies (in omphalocele) and intestinal atresias (gastroschisis) in patients with umbilical cord hernia. It originates at an early stage of embryogenesis and therefore may be detectable by fetal ultrasonography as early as the second trimester.¹ Interestingly, this early embryopathy persists throughout the rest of gestation as a stable umbilical mass.¹ At birth, it is mostly misdiagnosed as a 'small omphalocele' by many or brought to the attention of the pediatric surgeon after accidental injury to the gut by an umbilical clamp. Usually the contents are bowel, the hernia is reducible and anatomically the umbilical ring is complete, but may be somewhat wide. Neonatal correction is easy and cures the problem. Otherwise spontaneous epithelialisation of the coverings of the hernia would form a 'cutis navel'. In addition to this benign presentation of umbilical hernia, there have been reports of associated PVID producing meconium discharge in the perinatal or commonly in the postnatal period.²

Animal as well as autopsy studies on human fetuses have established the etiogenesis of ileal and jejunal atresias.³⁻⁶ Frequent clinical instances of intestinal atresias as a result of intrauterine mesenteric vascular insults such as volvulus, intussusception, internal hernia and constriction of the mesentery in a tight gastroschisis

or omphalocele defect have been observed. Atresia due to bowel incarceration in omphalocele⁷⁻⁹ or umbilical clamping of an occult omphalocele¹⁰ have been reported in the literature. Resorption of the isolated loop of the gut following vascular accident (intracelomic) leads to atresia.⁹

In our case, the initial appearance (Figure 1) was akin to umbilical hernia with PVID with meconium discharge from the cord. The black-colored loop-like structures in the cord and closely applied umbilical clamp seemed to indicate vascular injury to the gut and gangrene. This prompted an early exploration. Intraoperative findings suggested a 'type 3A' ileal atresia (Figure 2) with perforation of the dilated proximal segment of the gut. The distal segment was hypoplastic. The absence of meconium in the distal segment and the presence of whitish mucus strings favored an early fetal vascular accident involving the mesentery and small gut.

We hypothesized that this early vascular accident could be the causative factor for persistence of extracelomic herniation of the midgut and umbilical cord herniation. The blackish loops in the cord were found to be atrophic intestinal loops on histopathology. These might represent the partially resorbed portion of de-vascularised fetal intestines. This association is a rare occurrence.

Umbilical hernia is a distinct anomaly occurring at a different embryological stage as compared to omphalocele and gastroschisis. Vascular accidents may occur in the extracelomic gut causing formation of intestinal atresia. This may in turn cause persistence of hernia in postnatal life. Delivery room personnel should be aware of this anomaly and apply the clamp at a safe distance to avoid catastrophe. Any thickening of the base of the cord or a small opening on the lateral aspect of the cord should warn about this entity. Babies usually lack other congenital anomalies and surgical correction is simple at birth unless associated with PVID or intestinal atresia.



Figure 1. Hernia of the umbilical cord with perforation of the gut and black tubular structures in the cord.

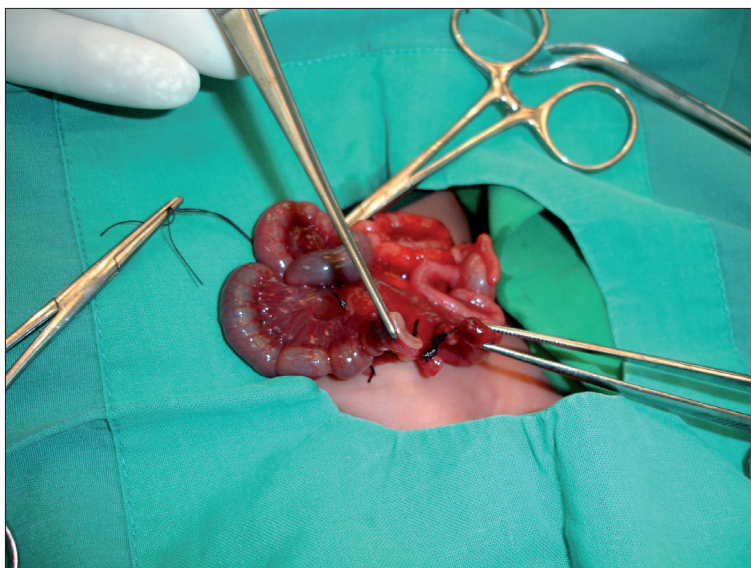


Figure 2. Type 3A ileal atresia.

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