Pitfalls in the umbilical pit: giant hernia of the umbilical cord

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DESCRIPTION
A full term male infant was admitted to hospital with evisceration of the abdominal contents through the ventral abdominal wall (ﬁgure 1). Examination revealed a skin-lined umbilical ring with eviscerated loops of bowel present within a membrane. The umbilical cord was inserted normally at the umbilical ring. Insertion of the rectus abdominis muscle was also normal. The echocardiograph was normal. No cardiac or other systemic abnormalities were found. Although a chromosomal analysis was not carried out, the neonate had no morphological features suggestive of Down’s syndrome.

On exploration almost all of the small intestine, appendix, caecum and part of the ascending colon was found in the sac. There was no evidence of atresia, obstruction or volvulus. The bowel was replaced in the peritoneal cavity and an umbilicoplasty performed.

Hernia of the umbilical cord is usually confused with omphalocele, but the conditions differ as regards embryological origin, morphological appearance and prognosis.

Embryologically, omphalocele occurs at 3–5 weeks of gestation due to primary failure of the body’s lateral ventral fold. Hernia of the umbilical cord occurs at 12–14 weeks of gestation due to failure of the return of the midgut from the umbilical coelom into the peritoneal cavity.

Morphologically, omphaloceles are more than 4 cm in diameter, the umbilical cord is inserted at the apex of the herniated sac and the herniated organs consist of the bowel, liver and, infrequently, the gallbladder. There is broad and lateral insertion of the rectus abdominis muscle on the costal margin. However, in hernia of the umbilical cord only the midgut is herniated and insertion of the umbilical cord and rectus abdominis muscle is normal. Malrotation is always present.1 2

Learning points
▸ Omphalocele and umbilical hernia both present with evisceration of the abdominal contents through the umbilicus, but they differ in embryological origin, morphological appearance and prognosis.
▸ Omphalocele is associated with chromosomal abnormalities, and cardiac and other systemic defects and therefore carries a poorer prognosis.
▸ Hernia of the umbilical cord is rare and has an excellent prognosis after surgery as there are no associated abnormalities.

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REFERENCES

Figure 1 A skin-lined umbilical ring with eviscerated loops of bowel present within a membrane.