

An umbilical defect

During their caesarean delivery, one of three triplet fetuses, aged 31 weeks and 2 days, was noted to have a clear shiny sac protruding from the base of the umbilical cord. The defect was immediately covered with moist dressings, which upon removal had some green-yellow staining. Physical examination showed an intact membranous sac containing what seemed to be several loops of bowel. Upon closer inspection of the sac, a small stoma was discovered (figure).

Questions

- (1) What is your diagnosis?
- (2) What additional diagnostic investigation could be useful in this case? Is it necessary?
- (3) What is the treatment of choice?

Answers

(1) There is a protrusion of abdominal contents through a mid-line defect that involves the umbilicus and a surrounding sac. It is the presence of the surrounding sac that classifies this defect as an omphalocele rather than gastroschisis (see discussion).

The stoma is the probable source of the fluid drainage and likely represents a patent persistent omphalomesenteric duct. A ruptured omphalocele can be ruled out by the presence of an otherwise intact sac.

(2) Water soluble contrast material could be injected via a small catheter into the stoma. Upon abdominal radiography, if the contrast material is found to have entered the lumen of the small bowel, then the presence of a persistent patent omphalomesenteric duct is confirmed. This test would not be necessary in this case, however, as the diagnosis can be made clinically.

(3) Surgery is the only treatment option. The operation

would be two-fold, requiring both resection of the duct and surrounding bowel as well as repair of the omphalocele.

Discussion

This case involves two congenital umbilical disorders occurring simultaneously: omphalocele and persistent omphalomesenteric, or vitelline, duct.

During normal embryological development, the primitive gut is derived from the intracoelomic portion, or roof, of the yolk sac. As the gut grows and develops, the wide communication between the mid-gut and the yolk sac narrows substantially to a small connection on the future ileum, known as the omphalomesenteric or vitelline duct.¹ This duct continues to shrink until around the fifth week of gestation, when it breaks off completely.² Meanwhile, the mid-gut elongates to form the U shaped mid-gut loop, which projects ventrally into the extraembryonic coelom in the umbilical cord. This physiological herniation occurs by the sixth week of gestation,³ and is thought to be due to a lack of space in the abdominal cavity subsequent to rapid growth of the kidneys and liver. By the 10th week, the omphalomesenteric duct should have completely obliterated, and by week 11, the mid-gut loop returned to the abdominal wall.

The most common congenital anomaly of the small intestine is persistence of all or part of the omphalomesenteric duct. Complete persistence of the omphalomesenteric duct is rare, occurring in less than 6% of omphalomesenteric duct anomalies,² and results in a patent tract between the ileal lumen and the outside of the body via the umbilicus. More commonly, in about 2% of the population and more than 98% of

omphalomesenteric duct defects, the duct may partially obliterate leaving a blind pouch on the antimesenteric surface of the ileum, known as Meckel's diverticulum.

Failure of the physiological hernia to reduce results in persistence of abdominal contents in the umbilical cord.³ This is known as an omphalocele, and typically consists of loops of bowel and rarely stomach, liver, or heart contained within a sac composed of peritoneum, amnion, and Wharton jelly lying outside the body through a defect in the abdominal wall just below the umbilicus.⁴ This differs from gastroschisis, which is a full thickness abdominal wall defect through which intestines can eviscerate into the amniotic cavity rather than the embryonic coelom.¹ The umbilical cord is not involved, and thus the defect is not midline but rather usually occurs on the right side.³ Unlike gastroschisis, omphalocele is associated with other chromosomal or anatomical anomalies.^{1,3,4}

Treatment of both patent omphalomesenteric duct and omphalocele is surgical. The portion of ileum containing the duct must be excised and the surrounding bowel reattached. Repair of the omphalocele depends on size. In this case, the omphalocele was considered small (less than 4 cm) and thus repair involved returning the bowels to the abdomen and closing the skin and fascia. Repair of a medium sized omphalocele (4-6 cm) involves skin closure only, leaving a ventral hernia to be repaired later after stretching of the abdominal wall. Large omphaloceles (7-10 cm) can be repaired by construction of a silo or prosthesis to cover the hernia and applying continuous pressure to enlarge the inadequate abdominal wall.⁴ Conservative treatment for unruptured omphaloceles of this size has been proposed and involves allowing the sac to harden and epithelialise, during which time the abdomen can grow.²

Survival after surgical repair for defects of all sizes is about 15%-60% and improves with increased birth weight and immediacy of repair.⁴

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- 1 O'Rahilly R, Muller F. *Human embryology and teratology*. 2nd ed. New York: Wiley-Liss, 1996: 59.
- 2 Skandalkis JE, Gray SW, Ricketts RR, Skandalkis JL. Anterior body wall. In: Skandalkis JE, Gray SW. *Embryology for surgeons*. 2nd ed. Baltimore: Williams & Wilkins, 1994: 540-94.
- 3 Moore KL, Persaud TVN. *The developing human: clinically oriented embryology*. Philadelphia: Saunders, 1998: 283-92.
- 4 Skandalkis JE, Gray SW, Ricketts RR, Richardson DD. Small intestines. In: Skandalkis JE, Gray SW. *Embryology for surgeons*. 2nd ed. Baltimore: Williams & Wilkins, 1994: 184-242.

