

A giant omphalocele in a preterm infant: the conservative approach

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DESCRIPTION

A routine antenatal ultrasound scan in the first trimester revealed a large omphalocele mainly occupied by liver in a female infant. Owing to preterm labour she was delivered by caesarean section at 29⁺⁴ weeks of gestation with a birth weight of 1450 g. No other malformations were present and her karyotype was normal.

The neonatal period was complicated by severe respiratory distress with pulmonary hypertension requiring prolonged high-frequency ventilation and inhaled nitric oxide. The infant developed bilateral grade 2 intraventricular haemorrhages. A haemodynamically significant ductus arteriosus was closed surgically. Owing to these complications, primary closure of the 4×4 cm large omphalocele (figure 1) was initially not feasible. Palliative care was considered, but was not an acceptable option to the parents. Hence conservative management was chosen aiming for delayed closure when the patient was stable enough.

Initial topical treatment with hydrofibre dressing with silver was stopped after 2 weeks, as concerns relating to silver toxicity have been described in the literature.¹ Our patient had no clinical signs of silver toxicity and epithelialisation continued to progress well with hydrofibre dressings without silver. Complete epithelialisation was achieved at 4 months of age (figure 2). Final surgical repair 1 month later was uneventful and the patient could be extubated on the second postoperative day.



Figure 2 Omphalocele completely epithelialised at 4 months of age.

An abdominal wall midline defect with a diameter greater than 4–6 cm and containing liver is usually considered to be a giant omphalocele (GO). However, no universal consensus exists on the definition. GO is associated with pulmonary hypoplasia and carries a high mortality risk. Delayed closure of a GO is a well-known alternative management technique in neonates who are either too unstable or with a defect considered too large for primary closure.² There is a paucity of reports describing a successful outcome with a delayed approach in very preterm infants. Active treatment of GOs in this population is often deemed futile as chances of survival without major morbidity are low.³

This case demonstrates the possibility of a satisfactory outcome in a very low birthweight infant with a GO undergoing delayed closure (figure 3). However, it must be emphasised that the clinical course was highly complicated and hospitalisation prolonged. Following surgical repair, there were



Figure 1 Omphalocele 4 weeks after birth.



Figure 3 The result a few months after secondary closure of the omphalocele. A transabdominal jejunal catheter in place due to feeding difficulties.



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Learning points

- ▶ In very low birthweight infants, primary closure of even a small-sized to mid-sized omphalocele might not be feasible in the first few weeks of life due to complications of prematurity.
- ▶ Conservative treatment with secondary delayed closure can be a successful alternative to primary closure or staged repair in these infants.

repeated episodes of severe cardiopulmonary compromise with readmissions to the intensive care unit.

Our patient was discharged at a corrected age of 10 months. At this time she showed severe delays in gross motor and moderate delays in fine motor development. The follow-up

examination at a corrected age of 15 months revealed only mild neurodevelopmental delay owing to great improvements in gross and fine motor development, with cognitive test results within normal limits for the infant's age.

Contributors FD and BR wrote the manuscript. FD, BR and the patient's parents provided the pictures. AM revised the manuscript. AM and WK were the surgeons in charge of the case.

Competing interests None.

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