Giant exomphalos—conservative or operative treatment?

DAVID C. S. GOUGH AND ALEX W. AULDIST

Royal Children's Hospital, Parkville, Victoria, Australia

SUMMARY The rate of survival for infants with intact giant exomphalos has much improved during the last 20 years; this is partly due to better respiratory and nutritional support. The use of a staged operative closure using a sialon prosthesis has been advocated for 12 years, but our data do not show this to be superior to nonoperative management.

Children with intact exomphalos pose considerable problems in management. The first question is whether an operative or a nonoperative method should be used when simple closure is not possible either because of the size of the defect or because of the contents of the sac.

Nonoperative treatment using drying agents—such as mercurochrome or alcohol—to encourage cicatrization, first advocated in 1899 by Ahlfeld, still has support, and if it is successfully used the child is left with a soundly healed abdominal wall or one that requires little subsequent surgery (Grob, 1963; Soave, 1963).

The skin mobilisation method of surgical treatment (Olshaussen, 1887; Gross, 1948) usually leaves an enormous hernia and there are significant technical problems in later repair of the abdominal wall. Subsequent surgery for this defect carries a mortality rate of 27% (Shuster, 1967).

Survival rates of patients with large exomphalos have been poor; in Melbourne between 1952 and 1962 the mortality rate was 75% for infants with defects of 5 cm or more across (Jones, 1963).

The arrival of a new concept in treatment, that of staged reduction by exterior pressure applied with a sac sutured to the edge of the defect (Shuster, 1967) was greeted by many surgeons as the answer to this problem. The aim of this treatment was to reduce the defect as quickly as possible with early discharge of the patient from hospital. However, our experience with this method of treatment has not been entirely satisfactory, so we were prompted to review our patients.

Royal Manchester Children's Hospital, Manchester

DAVID C. S. GOUGH, senior surgical registrar

Royal Children's Hospital, Melbourne

ALEX W. AULDIST, consultant paediatric surgeon

Materials and methods

Between 1967 and 1977, 87 patients were admitted to the neonatal unit of this hospital with intact exomphalos. 17 of these had defects measuring >5 cm diameter at the base, and they were retrospectively reviewed. Primary closure of muscle and skin was possible in 3 of them, 6 were selected according to the preference of the responsible surgeon for treatment by the Shuster technique using a sialon pouch, and 8 were similarly chosen for nonoperative treatment.

Results

There were 3 patients in whom primary closure was possible with a survival rate of 100%. One child has had two exchange transfusions for jaundice and at review had a pronounced hearing loss. There were no other medical or surgical problems in this group and their stay in hospital was about 3 weeks.

Two of the 6 patients treated with sialon prosthesis died. None had any other major malformation and both deaths were in the first 2 weeks of life. The cause of death was Gram-negative sepsis while on appropriate chemotherapy. Two of the survivors needed major reconstructive surgery of the abdominal wall and this was completed successfully. The patients treated by this staged repair stayed in hospital for between 4 and 7 weeks, but we noted that during the period that the prosthesis was *in situ*, no patient could be successfully established on alimentary feeding, so total parenteral nutrition was undertaken routinely in this group.

Eight patients were given conservative treatment and 3 of them died. Two deaths were due to associated malformations; one death at 24 hours was due

to Potter's syndrome and the other, from cardiac failure at age one week, was due to Fallot's tetralogy. Both diagnoses were confirmed at necropsy. In the remaining 6 patients the intact sac was painted with 2% aqueous mercurochrome. There was one death of a child in this group caused by achromobacter sepsis while he was being given inappropriate treatment with antibiotics, and 4 of the survivors have had, or will need, minor reparatory surgery to their umbilical defects. One child in this group was diagnosed as having mercury intoxication due to mercurochrome applications. He had a 9 cm defect (Fig. 1) and after initial sepsis and respiratory failure requiring ventilation failed to thrive at age 8 weeks (Fig. 2). Mercurochrome 2% had been

applied daily to wet areas. At this stage we proceeded with pure alcohol solutions as the only application and he then showed satisfactory weight gain (Fig. 3). One other patient in the series is mentally retarded with microcephaly. We believe this may be a case of undiagnosed mercury intoxication. Patients treated conservatively stayed in hospital for a time varying between 6 and 16 weeks.

Discussion

The object of treatment of these children should be primarily to save life, but an important consideration is to achieve a sound abdominal wall with as short a stay in hospital as possible. In this hospital there has

Fig. 1 Case 1. Mercurochrome treatment aged one day.

Fig. 2 Case 1. Mercurochrome treatment aged 8 weeks.

Fig. 3 Case 1. Mercurochrome treatment aged 11 months.

been an overall improvement in results: in 1952–62 there were 3 survivors out of 13 patients (Jones, 1963), in 1967–77 there were 12 survivors out of 17 patients. Improvements in the supportive care, particularly better respiratory support and nutritional techniques, seem primarily responsible for the higher rates of survival.

Several reports have confirmed the safety of non-operative treatment (Grob, 1957, 1963; Soave, 1963; Firor, 1971; Ryan, 1973). Even among the writings of surgeons who favour an aggressive surgical approach there are reports which show 28 survivors from 34 patients treated conservatively (Kim, 1976). These figures are mirrored closely by current results.

We are not aware of any substantial series of patients treated by the Shuster method that has better results. In fact Shuster's original series of 11 neonates with intact exomphalos treated by his own method, remains the largest single series and is still the most successful, with 9 patients surviving.

More recent and smaller series have shown mortality rates to be between 33 and 50% (Gilbert *et al.*, 1968; Ryan, 1973). A combination of staged reduction with skin or sialon prosthesis achieved good results in all 4 patients operated on by Mahour *et al.* (1973). Some arguments support operative management and emphasise that other anomalies may be present—such as gut atresia or diaphragmatic hernia (Mahour *et al.*, 1973). In our experience these anomalies can be diagnosed by clinical and radiological examination and, if present, may dictate a purely surgical approach, although their surgical correction may not of course prevent mortality.

The long-term aim of a soundly healed abdominal wall can be achieved in most patients by either technique. Secondary surgery will be necessary in

many patients, as shown in this series and confirmed by others (Shuster, 1967; Gilbert *et al.*, 1968), but the extent of operation is usually less in those who have been treated conservatively initially.

Sepsis is now the overriding problem in all methods of treatment. These patients stay a long time in intensive care units, and the use of systemic antibiotics and local antiseptics does not always prevent severe and sometimes life-threatening infections. This complication was responsible for the 3 preventable deaths in our series. Our experience has been that appropriate treatment with antibiotics controls sepsis more effectively in the absence of a foreign body prosthesis.

The risk of mercury poisoning with the use of mercurochrome has been emphasised (Fagan *et al.*, 1977). We believe this can be overcome by using mercurochrome without the impurity of free mercury and by using it only for a short period. We have also experimented with the use of a drying agent—such as alcohol not containing mercurochrome.

Other complications of conservative treatment—such as volvulus (Grob, 1963)—have been seen with the use of sialon prosthesis, but we did not encounter them. Since completing this series we have had one patient in whom the exomphalos sac ruptured while undergoing conservative treatment; this was successfully treated with a sialon prosthesis.

As long periods of stay in hospital are necessary in all forms of treatment, we encourage the parents to participate in the treatment of the child (Fig. 4).

Conclusions

At the Royal Children's Hospital, Melbourne, the survival of infants with intact giant exomphalos has improved significantly since the reported

Fig. 4 Case 2. Mercurochrome treatment aged 2 weeks, breast feeding.

mortality for the period 1952–62 of 75%, to 12 survivors in 17 patients in the current series. Improved respiratory support, intravenous nutrition, and new antibiotics have all contributed.

The results now reported indicate that if primary operative closure is not possible there is little difference in mortality between nonoperative management and staged operative closure using a sialon prosthesis.

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Correspondence to Mr D. C. S. Gough, Royal Manchester Children's Hospital, Pendlebury, Manchester M27 1HA.

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