CONSERVATIVE TREATMENT OF GIANT OMPHALOCELE*

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Omphalocele is a congenital malformation characterized by the failure of the abdominal walls to join together on the middle line as a consequence of a developmental defect.

The diameter of the resulting opening may range from a few centimetres to the almost total absence of the abdominal wall.

The diameter of the defect usually ranges from 3 to 8 cm. Naturally, the wider the opening, the more completely do the intestine and liver, covered only by the thin amniotic sac and peritoneum, protrude from the abdomen.

The sac is so transparent that the abdominal organs contained in it can easily be recognized.

Incidence

Omphalocele is a rare condition, with an incidence of one case in every 6,000 births. The obstetrician must avoid rupturing the amniotic sac, as occurs in about 20% of cases, because this is almost invariably followed by secondary infection of the peritoneum.

The hernia should be disinfected with alcohol and covered with a sterile dressing. The newborn infant should then be transferred as soon as possible to the Department of Paediatric Surgery because the sooner the severity of the defect is investigated and the most suitable treatment selected, the less danger is there of contaminating the thin avascular membrane.

There can be no doubt that this represents an emergency, because the air that the infant swallows as soon as it starts breathing fills the intestine and gradually distends it and it therefore becomes increasingly difficult to reduce it into the abdominal cavity. Furthermore, unless a dressing is immediately placed over the sac, it may become infected, giving rise to mechanical ileus and peritonitis.

Unless other malformations incompatible with life are present, every possible attempt must be made to cure the malformation.

Associated Anomalies. In our experience, associated malformations are present in at least 55% of

cases. Many, such as harelip, palatoschisis, extrophia of the bladder, meningocele, inguinal hernia and talipes, do not require immediate treatment.

On the other hand, it is of the utmost importance that the presence of more severe anomalies such as atresia of the intestine, duplication, caecal intussusception, malrotation, patent omphalomesenteric ductus, or trans-diaphragmatic hernia is investigated.

These malformations may be associated with omphaloceles of any size. Whatever the size of the hernia, there is always a disproportion between the volume of the organs contained in it and the capacity of the abdominal cavity into which this mass must be replaced, thus giving rise to a difficult surgical problem.

Size of Omphalocele and Indications for Treatment

Prognosis. In mild cases of omphalocele in which the diameter of the defect in the abdominal wall is less than 3 to 4 cm. and only a small part of the liver and part of the intestine protrude into the sac, the surgical problem is not severe and the malformation can be corrected surgically.

When the defect is small, the only problem is to join together the fasciae of the recti abdominis muscles and the abdominal wall solidly after having excised the amniotic sac.

Two-step Treatment. Cases of omphalocele of larger size, in which the diameter of the defect is 4 to 6 cm. or more, usually cannot be treated in one step because the fasciae of the recti abdominis cannot be brought together. In such cases the two-step method is used. With this technique the amniotic tissue is preserved in order to prevent the formation of adhesions between the abdominal organs and skin.

The two-step method was described by Williams in 1930 and first successfully applied by Gross in 1948.

This technique has since been universally adopted. It has many advantages, but also a number of drawbacks, and cannot always be applied. In fact, the amount of cutaneous tissue available is not always abundant and, in such cases, the suture

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is under considerable tension, with consequent considerable increase in endo-abdominal pressure caused by the partly reduced abdominal organs, distended by the intestinal gases; these displace the diaphragm upwards, thus obstructing the return blood flow to the heart in the vena cava inferior and vena porta, with the consequent occurrence of cyanosis, dyspnoea, and even irreversible shock.

It should be remembered that compression of the intestine can cause perforation and peritonitis. These complications must be carefully considered in newborn infants with a large omphalocele containing most of the liver. In such cases, provided the sac has not been injured, the writer prefers the conservative method, with which excellent anatomical and clinical results have been obtained in four cases (Figs. 5 and 6).

Conservative Method

When a newborn infant with omphalocele in which the diameter of the abdominal defect is more than 4 cm. (Fig. 1) is transferred to our department, surgery is discarded in favour of the following procedure: the umbilical cord is clamped and cut as short as possible; the entire surface of the amniotic sac and the abdominal wall are disinfected with alcohol and are then swabbed with a 2% mercurochrome solution; a sterile protective dressing is applied. The patient is then placed in a warm bed under a protective arch which prevents the blankets from contaminating the sac. This antiseptic treatment is continued for a few days until the thin amniotic tissue necroses and becomes covered with a thick squamous crust which protects the sac from the risk of possible infections or perforation.

The base of the omphalocele is then protected by means of a ring-shaped wad of cotton wool wrapped with a bandage, in order to fix it in the required position and to prevent sudden movements. Appropriate antibiotic treatment must naturally be instituted upon admittance. Granulation tissue is gradually formed under the crust, from the borders of the sac inwards (see Fig. 2), and is then substituted by epithelial tissue as the dry layer drops off (see Fig. 3).

Sometimes a strong-smelling corpusculated serous secretion oozes out from under the crust.

As epithelization proceeds from the borders towards the centre of the defect, cicatrization and subsequent retraction occur in the tissues over the defect, and this is associated with the gradual replacement of the intestine and liver in the abdominal cavity, without risk of respiratory or cardiac complications, or shock.

Thus, the process which failed to take place, on account of a developmental defect of the walls of the abdomen during the tenth to twelfth weeks of foetal life, occurs partly and incompletely.

During this process of healing by second intention, lasting from two to three months (according to the size of the omphalocele), the abdominal cavity begins to form and to enclose the organs which it had formerly failed to contain.

Thus, also the liver (which almost always protrudes, to a variable extent, into the amniotic sac) forms its own site, and no inflammatory processes due to compression and causing perivisceritis (as observed with the two-step surgical method) occur. At about the end of the third month, the whole surface of the abdomen is covered with skin (see Fig. 4), and the child must be wrapped with a soft elastic bandage of suitable tension.

Our Clinical Experience. Our experience of conservative treatment of omphalocele, proposed by Ahlfeld in 1899 and successfully applied by Grob in 1957 and Cunningham in 1956, began in 1956. This technique was adopted after the Gross method had failed in three cases of large-sized omphalocele; these cases died during the first 24 to 48 post-operative hours of shock caused by compression of the intestine and thoracic organs because only an inadequate amount of skin tissue could be mobilized to cover the uninjured amniotic sac and consequently the suture was always excessively taut.

In our experience, the amniotic sac disinfected and treated with 2% mercurochrome does not become contaminated and does not perforate; the gradual process of epithelization from the borders inward favours cicatricial retraction of the skin and this covers the defect over a period of some weeks without danger of compression and shock; this is associated with the gradual replacement of the abdominal organs into the abdominal cavity, until they can be replaced surgically with plastic surgery of the abdominal walls as soon as this seems feasible.

As indicated in the Table, 25 newborn infants with omphalocele of different sizes were transferred to our department from Liguria and other Regions during the period 1951-1960. Of these 25 patients, five were not treated or operated upon because of the presence of other associated malformations and of the severe condition of the babies, due to rupture of the sac (*in utero*). All five died.

Eight infants with small omphaloceles (2 to 4 cm.) were operated on immediately. Three died during the post-operative period.

FIG. 1.

FIG. 2.

FIG. 4.

FIG. 3.

FIG. 5.

FIG. 1.—A 15-hour-old female child with large omphalocele (9 cm.).
 FIG. 2.—Condition of the omphalocele after 26 days of treatment.
 FIG. 3.—Condition of the omphalocele after 39 days of treatment.
 FIG. 4.—After about three months the omphalocele has been completely covered by the skin.

FIG. 5.—The same child at about 18 months of age. Plastic surgery of the abdomen was performed at the age of 12 months. The abdomen is not globose; the walls are tonic. In this case mesenterium commune was present but did not require surgical correction. Appendicectomy was performed.

TABLE
 INCIDENCE AND MANAGEMENT OF OMPHALOCELE
 1951-1961 (25 CASES)

No. of Cases	Diameter of Neck of Sac (cm.)	Untreated	Treatment			Results	
			One-stage	Two-stage	Conservative	Patients Living	Deaths
5	2-8	5	8				5
8	2-4						3
5	3-5			3	2	4	1
3	6-9			3			3
4	7-9				4	4	
25			8	6	6	13	12

Associated anomalies were as follows: malrotation, 12 cases; congenital heart disease, 2; Meckel's diverticulum, 1; atresia small bowel, 1; and polydactyly, 1 case.

FIG. 6.—This child was born prematurely (at 7 months) on September 14, 1958, with a large omphalocele. Weight at birth was 1.375 kg. Omphalocele was treated conservatively. The course of the healing process was identical to that in the previously illustrated case.

FIG. 7.—This figure shows the same patient as in Fig. 6 after plastic surgery of the abdominal walls; this was performed without difficulty and with excellent results at about 1 year of age. Appendicectomy was performed.

Five other cases had larger omphaloceles (diameter, from 3 to 5 cm.). Of these, three were treated with the two-step surgical method, with one fatality due to respiratory changes a few hours after the operation, and two were treated with the conservative method with excellent results.

Of the seven cases with giant omphalocele (diameter, more than 7 cm.), three were immediately treated by mobilizing the skin and covering the sac; in all cases the suture was very taut at the end of the operation and the patients died after 24, 36, and 52 hours, respectively, on account of respiratory and cardiocirculatory complications. The other four cases, in which the diameter of the defect was 8 and 9 cm. respectively, were treated with the conservative method, and are now in good health after subsequent plastic surgery (see Figs. 5 and 6). We feel sure that in these four especially severe cases, such good results would not have been obtainable with the two-step surgical method.

Mesenterium commune was found at surgery or at autopsy in seven cases, and defective joining of the mesenteries in five cases in this series of 25 cases of omphalocele. In no case did the malformation cause complications requiring immediate surgery. This is another factor in favour of the conservative treatment of omphalocele, which is associated with the above-described malformations in a high proportion of cases.

Naturally, surgery can be performed during the conservative stage of treatment should signs of intestinal obstruction appear (direct radiographs are valuable in this connexion); or should the sac rupture.

A noteworthy feature is that no evidence of perivisceritis, such as is inevitably found between the intestinal loops in cases with the two-step method with extensive mobilization of the skin, was observed in infants treated with the conservative method during the subsequent operation for plastic reconstruction of the abdominal walls.

We always perform appendicectomy, after examining the intestine, before suturing the abdomen.

Conclusions

The results obtained in our patients indicate that the prognosis for omphalocele can be improved further if conservative treatment is adopted by surgeons with more confidence.

This optimistic outlook also holds good for cases of giant omphalocele containing most of the liver, such as our four patients; in these severer cases, the 15% incidence of favourable results reported by Gross may well increase to 40 or 45% in the absence of other associated malformations incompatible with life.

The use of skin grafts has been proposed in cases of large omphalocele, but the results have been uncertain. Naturally, less severe cases of omphalocele can also be treated conservatively (Fig. 7).

The only objection to the conservative method is that it requires prolonged hospitalization, but this objection cannot be accepted in view of the aims of neonatal surgery, i.e. to ensure treatment of the patients' malformation by the safest, even if costliest, method.

REFERENCES

- Ahlfeld, F. (1899). Der Alkohol bei der Behandlung inoperabler Bauchbrüche. *Mschr. Geburtsh. Gynäk.*, **10**, 124.
- Cunningham, A. A. (1956). Exomphalos. *Arch. Dis. Childh.*, **31**, 144.
- Grob, M. (1957). *Lehrbuch Der Kinderchirurgie*, p. 311. Thieme, Stuttgart.
- Gross, R. E. (1948). A new method for surgical treatment of large omphaloceles. *Surgery*, **24**, 277.
- Williams, C. (1930). Congenital defects of the anterior abdominal wall. *Surg. Clin. N. Amer.*, **10**, 805.