EXOMPHALOS (syn. OMPHALOCELE)

A REVIEW OF 45 CASES

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In rare anomalies personal experience of a few patients can lead to biased or incorrect impressions unless analysis of a larger series is applied as a corrective. The result should be a better appreciation of the difficulties to be met and overcome in the future.

A congenital hernia into the base of the umbilical cord is known as an exomphalos in British usage and an omphalocele in American usage. Smaller examples are readily repaired but the largest constitute one of the greatest challenges in paediatric surgery. Other serious malformations are often present, frequently cause complications and may vitiate the results of surgery.

The earliest description of an exomphalos is found in the writings of Ambroise Paré (1510-1590) who recognized its serious nature and poor prognosis (1634). Mery (1716) wrote '... a yet more extensive degree... in which the very nature of the case seems to preclude all hope of assistance from the art of surgery. The dissection of such cases has shown the liver, stomach, spleen, omentum, large and small intestines lying in the umbilical hernia'. The first reports of successful surgical treatment are those of Hey (1803) and Hamilton (1806). Scarpa (1812) drew attention to the association of exomphalos with other malformations and concluded that the condition was almost always fatal because of them or because the hernia was too large to be reduced. Ahlfeld (1899) used alcohol dressings on the sac in a patient successfully treated without operation.

Anatomy and Natural History

Return of the gut from the extra-embryonic coelome is normally completed in the tenth week of foetal life, by which time the coelomic cavity is large enough to accommodate the still lengthening alimentary canal. Frazer (1931) suggested that this return might be partly due to a sudden relative decrease in the size of the liver, failure of which could explain its presence in the sac in the larger

types of exomphalos. The true cause of persistence of a foetal type of anatomy is unknown.

At birth the sac is a shiny pellucid membrane formed by a layer of amnion with peritoneum beneath it. Between these is stratum of pale yellow embryonic tissue (Wharton's jelly) of varying thickness and with occasional bulbous excrescences. At the periphery of the sac there are some blood vessels beneath the peritoneum (Fig. 1), but they are apparently inadequate, for death of the sac commences at the moment of birth. Other factors such as the drying effect of contact with the air and perhaps increase in intra-abdominal pressure may contribute to necrosis of the sac. Within six to 12 hours it becomes cloudy, yellow and opaque, and the surface becomes granular, sticky and malodorous. With longer exposure it becomes an inelastic eschar liable to rupture with evisceration.

When rupture occurs before birth, the gut spills into the amniotic cavity where vernix caseosa and sometimes meconium produce distinctive and formidable changes. The bowel becomes thick and woody (Fig. 2), and appears to be much shortened. The vessels of the serosal surface are deeply congested and loops of bowel become cemented together over nodules of vernix. These changes, resembling meconium peritonitis, make surgical repair difficult and sometimes impossible.

When rupture occurs during birth it also carries the risk of peritonitis, but when repair is effected without undue delay or additional contamination the outlook is much less grave than when prepartum rupture has occurred.

The Patients

Between 1952 and 1962, 45 infants with exomphalos were admitted to the Royal Children's Hospital, Melbourne, and, as a result of a retrospective study of their case notes, they have been classified according to whether the sac was intact or ruptured. Those in whom the sac was intact

FIG. 1a.—Blood vessels in the periphery of exomphalos sac: section taken at 1.5 mm. from the edge of the sac shows vessels grouped near the peritoneal surface. ($\times 75$.)

FIG. 1b.—Section taken at 2.5 mm. from edge of the sac showing diminishing number of vessels as the central area of the sac is approached (periphery to the right). ($\times 25$.)

have been further subdivided into three groups according to the size of the exomphalos, with a separate category for those in whom it formed part of a complex malformation: vesico-intestinal fissure (Fig. 3).

Two dimensions of an exomphalos are each important: (i) *the cubic capacity of the sac*, which

determines the volume of the contents; as a practical matter this cannot be subjected to measurement and is indicated here by an approximate maximum diameter; (ii) *the diameter of the neck*, or defect in the abdominal wall, which partly determines the ease or difficulty of repair of the musculo-fascial layers.

FIG. 2a.

FIG. 2a.—Normal neonatal bowel. ($\times 40$.)

FIG. 2b.

FIG. 2b.—Pathological changes in the bowel wall after prepartum rupture of the exomphalos sac. The serosal layer is greatly thickened and with higher magnification squames of vernix caseosa surrounded by a zone of small round cells can be identified. The picture closely resembles that of meconium peritonitis. ($\times 40$.)

FIG. 3.—Classification of exomphalos in 44 cases. One additional infant who survived has not been included in the classification because of insufficient information in the case notes on the size of the exomphalos.

A classification based on these measurements was applied to the 32 cases in this series in which the sac was intact. The case notes of one infant who survived contained insufficient information on the size of the exomphalos for allocation to a specific group.

Group I: This comprised three infants with small intact exomphalos, who were born at term; none had any other malformations and all survived.

Group II: This comprised 16 infants with medium-size intact exomphalos; in only one was part of the liver herniated and the majority survived.

Group III: This consisted of 13 infants with large intact exomphalos. Part of the liver was present in the sac in 11 of the infants, more than half of whom were born prematurely. The incidence of associated anomalies was high and the mortality in this group as a whole was also high.

Group IV: Vesico-intestinal fissure occurred in six patients. Malformations in five of them conformed to the standard pattern and the only survivor presented a minor degree of the anomaly, as will be described later.

Group V: This consisted of six infants in whom the sac had ruptured at the time of admission to hospital, and in five the changes in the bowel wall were consistent with rupture *in utero*. In the sixth patient rupture probably occurred during delivery.

Treatment

Repair of the abdominal wall was achieved in one stage in each of the 19 patients in Groups I and II, and 16 survived to leave hospital.

In 10 infants in Group III repair in two stages was planned, but eight died after the first operation. In four instances the sac was covered with mobilized skin and in the other four skin alone was closed after excision of the sac. The second stage was successfully completed in two infants at 1 year and 2½ years respectively, and in each the sac had been excised, closing skin alone, at the first operation. Complete repair in one stage was carried out on three infants in this group and one of them survived.

Surgical correction of vesico-intestinal fissure in one stage was successful in one infant in Group IV with a minor degree of the anomaly. In three of the remaining five some attempt at correction was made but none was successful.

One-stage repair was performed in three infants in Group V and one survived. Closure of the skin alone was achieved in two infants; one survived and now awaits the second stage which has been deferred indefinitely for there is no evidence of increased abdominal capacity. In one moribund infant no operation was attempted.

Of the 45 infants with exomphalos, 22 (approximately 46%) left hospital after surgical treatment, and three of these died before reaching 1 year of age. There are thus 19 children (approximately 40%) now alive; in two of these repair was completed in two stages and two are awaiting the second stage. The number of survivors in each group is shown in Fig. 3.

Vesico-intestinal Fissure

Several authors have put forward an alternative name for the condition, but vesico-intestinal fissure, suggested by Schwalbe (1909), best indicates the lack of separation of the urogenital sinus and the alimentary canal. Johnston (1913) analysed several possible embryological theories in a thorough study of the condition.

In this complex anomaly there is an exomphalos which, though of large extent, is of little capacity, for the surface does not bulge but retains the contour of the abdomen. The typical form of the malformation is shown in Fig. 4. Beneath the exomphalos there is an ectopia vesicae with a central area of ectopic intestinal mucosa; through this passes the terminal ileum which is frequently prolapsed. Beneath this again there opens a segment of colon which ends blindly in the pelvis; the anus is imperforate. Bladder mucosa is present on each side of the intestinal area, and the penis (or clitoris) is

split and duplicated. Malformations of the internal and external genitalia are frequent and variable.

The abnormalities in five typical cases are shown in Table 1. The sixth and only surviving infant displayed a relatively minor degree of the defect in which the alimentary canal was intact and the ectopia vesicae was covered with a thin layer of unsupported skin. The exomphalos was repaired in one operation.

Experience in the five infants who died yielded no constructive suggestions for the treatment of this condition. Rickham (1960) has reported the first and, it seems, the only survivor in whom the complete anomaly was present.

Methods of Treatment in Patients with Intact Sac

I. Primary Repair. Closure of each layer of the anterior abdominal wall is the surgical ideal and an exomphalos of small or medium size can usually be repaired in this way.

II. Two-stage Repair. When the sac is large, and especially when it contains part of the liver, the abdominal cavity may be too small to permit complete reduction and closure. Gross (1948) devised a method in which the intact sac is retained, skin is freed by undermining and drawn over the sac to provide coverage. Complete repair is deferred until the capacity of the abdomen has increased sufficiently and this may require one year or even longer. This method requires a sac of acceptable viability, and the first stage should be performed as soon after birth as possible, preferably within six hours. Retaining the sac beneath the skin diminishes the formation of adhesions between the bowel and the parietes. Adhesions may be

FIG. 4.—Diagram showing the standard pattern of the malformations in vesico-intestinal fissure: E., exomphalos; C., umbilical cord; E.I.M., ectopic intestinal mucosa; I., prolapsed ileum; D.C., distal colon; E.B.C., extrophied bladder mucosa; U., ureteric orifices; I.A., imperforate anus.

numerous when only skin is closed, but in two infants whose treatment was conducted personally the adhesions were diaphanous and not insurmountable.

A disadvantage of closing skin over the intact sac is that intra-abdominal exploration is precluded and a remediable second lesion cannot be detected, as the following personal experiences illustrate.

Case 1. A full-term infant was operated on 12 hours after birth for an exomphalos 7.5 cm. in diameter in

TABLE 1
VESICO-INTESTINAL FISSURE

	Case 19*	Case 30	Case 39	Case 43	Case 38	Case 25
Sex ..	F	F	M	M	M	M
Premature ..	+	+	+	+	+	+
Exomphalos ..	+	+	+	+	+	+
Ectopia vesicae	Partial	+	+	+	Partial	+
Ileum ..	Normal	Prolapsed	Prolapsed	Prolapsed	Prolapsed	Atretic
Appendix ..	Normal	Duplicated	Absent	Duplicated	Absent	Duplicated
Colon ..	Normal	Agensis	Blind end loop	Blind end loop	Blind end loop	Blind end loop
Anus ..	Ectopic	Imperforate	Imperforate	Imperforate	Imperforate	Imperforate
Ureters ..	L. megaureter	Megaureters	Normal	R. absent L. normal	Megaureters	Normal
Urethra ..	Normal	Absent	Duplicated	Absent	Absent	Absent
Penis ..	—	—	Duplicated	?	Duplicated	Absent
Testes ..	—	—	Intra-abdominal	Undescended	Undescended	Undescended
Vagina ..	Pinhole orifice	Absent	—	—	—	—
Uterus ..	?	Duplicated	—	—	—	—
Kidneys ..	R. hydro-nephrosis	Normal	Normal	Absent on right	Bilateral hydro-nephrosis	Normal
Other anomalies ..	Cleft lip; facial cleft; absent fibula	Nil	Hemivertebrae	Talipes	No muscle in abdominal wall	Malrotation; malfixation

* Survivor.

which the liver was present. A pre-operative radiograph of the chest showed no abnormality. Skin was drawn over the intact sac and closed with a vertical line of silk mattress sutures. Severe dyspnoea developed after operation, respirations rose to 80 per minute and death occurred suddenly 36 hours later. Autopsy disclosed a large left posterior (Bochdalek) diaphragmatic defect through which much of the bowel had passed.

Because of this experience, the sac, though viable, was excised in the next patient.

Case 2. A male Maltese infant weighing 2.5 kg., was operated on when 2 hours old for a large exomphalos 12 cm. in diameter (neck: 10 cm. in diameter), which contained a large portion of the liver and coils of small bowel. No other abnormalities had been detected before operation. The sac was excised except for an area 2 cm. in diameter where the third 'middle' lobe of the liver, which measured 8 cm. $\times$ 8 cm. $\times$ 6 cm., was adherent. The splenic flexure of the colon was situated in the left pleural cavity having passed through a defect 5 cm. in diameter in the anterior part of the left cupola of the diaphragm. The left end of the transverse mesocolon formed a sliding hernial sac which was reflected downwards to display the edge of the defect. It was bounded anteriorly by the costal cartilage and was closed with interrupted silk mattress sutures, the most anterior stitch passing through perichondrium. The caecum was subhepatic and incompletely rotated; there was no fixation of the mesentery of the small bowel and no duodenal obstruction. The liver was pressed back into the peritoneal cavity with difficulty and skin alone brought over the coils of bowel after extensive dissection in the subcutaneous plane. At the end of the operation a large left indirect hernia appeared and was welcomed as a 'safety valve'. Radiographs showed that the diaphragm was raised with partial collapse of the right lower lobe of the lung. Intermittent vomiting due to gastro-oesophageal reflux was troublesome, but subsided in the following four weeks in response to thickened feeds and nursing in the erect position. The baby left hospital at 3 months of age weighing 4 kg.

On three occasions during the first year, bouts of vomiting with dehydration led to readmission to hospital. Barium fluoroscopy showed persistent gastro-oesophageal reflux and rotation of the pyloric antrum. Either of these factors could have caused the vomiting, but partial intestinal obstruction due to intraperitoneal adhesions could not be completely excluded. In hospital the symptoms soon subsided and laparotomy was not necessary. When the baby was 22 months of age it became possible for the first time to reduce manually the content of the ventral hernia into the peritoneal cavity; however, before the second operation could be arranged he contracted pulmonary tuberculosis from his father, who was infected while working as a steward in a migrants' club. Within three months the patient's small apical infiltration and enlarged hilar nodes responded to antituberculous drugs and a sanatorium régime.

At the age of 2 years 6 months, when the patient

weighed 11.7 kg., repair of the hernia was performed. There were filmy adhesions between all the coils of small bowel and the inner surface of the skin, but these could be separated easily without leaving bare or bleeding surfaces. The defect measured 10 cm. vertically and 8 cm. transversely and closure presented considerable difficulty. Flaps of anterior rectus sheath were raised and reflected medially, peritoneum and fascial flaps being reconstructed as a single layer, reinforced in its lower third by bringing together the lateral margins of the anterior rectus sheath. At the end of the operation the left inguinal hernia was very large and tense—to such a degree that compression of the scrotum produced an excursion in the anaesthetist's inflation bag. Despite this there was no respiratory embarrassment after operation. The child was nursed in an oxygen tent and his respiration rate did not exceed 30 per minute. Convalescence was rapid and he left hospital 12 days after operation.

Three months later the inguinal hernia sac was excised, the weakened canal extensively repaired, and an undescended testis placed in the scrotum. When seen six months later the patient was very well.

The following factors were important in this boy's survival: (i) absence of prematurity and associated malformations; (ii) discovery and repair of the diaphragmatic hernia; (iii) a two-stage repair; and (iv) development of an inguinal hernia as a 'safety valve'.

III. Non-operative Treatment. (a) Torsion and strapping has been suggested as the treatment for a small exomphalos sac, but the presence of congenital adhesions in nine of 32 cases with an intact sac indicates a possible danger. In practice patients with this type of exomphalos tolerate surgical repair well.

(b) Grob (1957) described the use of an aqueous solution of mercurochrome to preserve the sac until the surrounding skin had grown in to cover the exomphalos. Wollenweber and Coe (1959) also used a modification of this method successfully. The stay in hospital may be long, perhaps four to six months, and lingering fears of rupture remain despite reports of success. Nevertheless, the high mortality in Group III might justify a trial of non-operative methods.

Causes of Death

Group II (16 cases, three deaths). At autopsy extensive pulmonary atelectasis was found in two infants and in one of these the inhalation of a considerable amount of liquor amnii and cellular debris was attributed to foetal distress during delivery. The third infant was cyanosed from birth until sudden death on the fourth day after operation. A cardiac anomaly was strongly suspected but unconfirmed as permission for autopsy was refused.

Three infants survived complete repair and left hospital but died within 12 months. One death was probably due to an overwhelming virus infection for a large pericardial effusion was the only significant post-mortem finding. The second infant had a truncus arteriosus, was cyanosed throughout life and died of bronchopneumonia. The third patient had a partial eventration of the left side of the diaphragm, which was successfully repaired at the same time as the exomphalos. Death from pulmonary suppuration occurred at the age of 2 months and at autopsy a partial eventration of the right half of the diaphragm was found. A pulmonary abscess was present in the basal portion of the right lung and was thought to have originated in a sequestered lobe.

In two of these three patients death was the direct but delayed effect of a congenital defect.

TABLE 2
CAUSES OF DEATH IN GROUP III

Causes of Death	No. of Cases
<i>Cardiac anomalies</i>	6
Coarctation + patent ductus arteriosus	2*
Ventricular septal defect	2*
Coarctation, ventricular septal defect, anomalous pulmonary venous drainage	1*
Coarctation (Turner's syndrome)	1*
<i>Haemorrhage</i>	2
Cerebral (tentorial tear)	1
Suprarenal	1
<i>Respiratory insufficiency</i>	2
Atelectasis, collapse	1
Compression, diaphragmatic hernia	1*
Total	10

* Fatal malformation.

Group III (13 patients, 10 deaths). Analysis of the deaths in this group (Table 2) shows that six infants had severe cardiac malformations, two had extensive haemorrhages and two died from respiratory embarrassment. In one of the last-mentioned there was a diaphragmatic hernia and the other, a premature infant, probably died from the effects of high intra-abdominal pressure, although only skin was closed at operation. Autopsy showed neither atelectasis nor any other malformation.

A study of the deaths in this group has yielded some information that might help to save more babies. One example is the infant whose diaphragmatic hernia might have been detected if the abdomen had been explored, for there was no evidence of it in the pre-operative radiograph. The presence of this anomaly in cases of exomphalos is not common and it raises the question whether it is justifiable to excise the sac, which could be preserved

with advantage in a two-stage repair, in order to discover an additional unsuspected but remediable defect. The answer remains in doubt despite the findings described in Case 2 where survival was largely due to repair of a diaphragmatic hernia.

Surgical correction of the cardiac defects was theoretically possible in each of the six infants in which they occurred, but operation is not undertaken in the neonatal period being deferred for months or years to a more favourable time. The defects are usually compatible with life provided no extra burdens, such as anaesthesia and operation for exomphalos, are added.

The natural eagerness to operate as an emergency on an infant with an exomphalos should be restrained, for it is reasonable to spend some time in a search for cardiac and other malformations especially in infants in whom the neck of the exomphalos exceeds 5 cm. and part of the liver is herniated. Prematurity and pulmonary atelectasis should also be taken into account, for when they occur in patients of this type (Group III), the results of operation are so poor that recourse to non-operative treatment might be justified.

Factors Influencing Survival

Size of Exomphalos. It is evident that the smaller the sac and its capacity, the better the chance of survival. There also seems to be some correlation between the large defect, herniation of the liver and prematurity.

Prematurity. Infants with a large exomphalos sac or associated developmental defects tend to be born prematurely (at least six of the 13 in Group III, all of the six in Group IV). Prematurity brings its own risks and one, atelectasis, is particularly pertinent, for it may be aggravated by pulmonary collapse due to the elevation of the diaphragm by a raised intra-abdominal pressure after operation. The size of an exomphalos should strictly be related to the size of the patient, for a sac 5 cm. in diameter is relatively much larger in an infant weighing 2 kg. than a sac of the same size in one weighing 4 kg.

Post-operative Intra-abdominal Pressure. Some rise in intra-abdominal pressure after operation is almost inevitable in all cases of exomphalos, even when only skin is drawn over an intact sac. Respiratory distress and cyanosis are usually attributed to an elevation of the diaphragm limiting pulmonary aeration. Oedema of the lower extremities may also be due to a raised intra-abdominal pressure, or to compression of the inferior vena cava by a forcibly reduced 'third lobe' of the liver.

TABLE 3A
COEXISTENT MAJOR MALFORMATIONS

Malformation	No. of Cases	Deaths
Cardiac		
Coarctation alone	3	3
Coarctation and patent ductus	2	2
Ventricular septal defect	2	2
Anomalous pulmonary venous drainage ..	1	1
Truncus arteriosus	1	1
Total	9	9
Genito-urinary		
Vesico-intestinal fissure	6	5
Horseshoe kidney	2	1
Hydronephrosis	1	1
Megaureters	3	2
Triad syndrome*	1	1
Total	13	10
Alimentary		
Malrotation and malfixation	8	0
Cystic liver	1	0
Diaphragmatic		
Anterior defect	1	0
Posterior defect	1	1
Eventration (bilateral)	1	0
Pulmonary		
Sequestered lobe	1	1
Total	35	21

* Absent anterior abdominal wall muscle; megaureter and hydronephrosis; undescended testes.

TABLE 3B
COEXISTENT MINOR MALFORMATIONS

Malformation	No. of Cases
Talipes	2
Absent fibula	1
Hemivertebrae	2
Micrognathos	2
Cleft lip	2
Cleft palate	2
Facial cleft	1
Hypospadias	1
Finger anomaly	1
Total	14

There were 12 infants in this series who had dyspnoea and a respiratory rate of 80-100 per minute after operation; nine of these infants died and seven were found at autopsy to have a cause other than pulmonary compression for their respiratory distress: severe cardiac malformation in four, hyaline membrane in two and cerebral haemorrhage in one. It is reasonably certain that in the remaining two infants respiratory distress and death were due to raised intra-abdominal pressure, for no alternative cause was found at autopsy.

Three infants with severe dyspnoea recovered;

all were born at term and two showed post-operative radiographic evidence of pulmonary collapse which was temporary.

The conclusion is that in the majority of patients with respiratory distress after operation, the cause is probably other than high intra-abdominal pressure alone, which may be less important than at first appears.

Herniation of Liver. There is no doubt that herniation of the liver into the sac is an ominous finding, for it tends to occur when the abdominal defect is large, and in premature infants with other malformations. Of 14 patients in whom herniation of the liver occurred only three survived.

The portion of the liver in the sac is demarcated by an additional deep fissure which marks the right margin of a third or 'middle' lobe which is bounded on the left by the fissure of the ligamentum venosum. In some cases continuity with the rest of the liver is lost, and the extra lobe has a 'pedicle' of its own which contains a third hepatic duct. The gall-bladder in such cases is usually found in a shallow depression on the under surface of the middle lobe. In one of my patients a large middle lobe of this type weighed 85 g. and was successfully excised after peeling from it the gall-bladder and the hepatic ducts. A two-stage repair was completed at the age of 1 year and the child is now 8 years old and thriving.

Coexistent Malformations. In 45 patients there were 10 in whom exomphalos was the only malformation present and all of these survived. Each of the other 35 infants had at least one other serious developmental anomaly (Tables 3a and b) and in 21 of the 27 deaths these malformations played a significant part. Malrotation with malfixation was reported at operation or at autopsy in only eight infants. This is a low estimate of its incidence and it was probably undetected in others. However, there was not one instance in which mid-gut volvulus or duodenal obstruction occurred at any time.

Rupture of the Sac. Pre-partum rupture of the sac inevitably complicates surgical treatment and in all but the smallest defects a two-stage repair is probably indicated. Peritonitis and intestinal obstruction due to adhesions are also to be expected even when skin coverage and repair can be effected.

Early Operation. The short life of the sac is important, for operation should be carried out as early as possible after birth if it is to be retained in a two-stage repair. Theoretically there is an additional advantage in early operation, for aeration

of the bowel is incomplete, its cubic capacity is less and the rise in intra-abdominal pressure after operation should also be smaller.

Summary and Conclusions

A retrospective study of the case notes of 45 infants with exomphalos has led to the arrangement of those in which the sac was intact into three groups according to the size of the exomphalos. Two additional groups were distinguished by reason of rupture of the sac or because the exomphalos formed part of a complex anomaly: vesico-intestinal fissure.

The survival rate was best when the exomphalos was small or medium-sized, and when the liver was not present in the sac. In such infants a primary repair of the abdominal wall was usually possible and the result satisfactory.

Additional serious malformations, chiefly cardiac, were present in 35 of the 45 infants, and they were the direct or indirect cause of death in 21 (approximately 43%). Less serious associated malformations were found in 14 infants.

Malrotation and malfixation were recorded in only eight infants, but mid-gut volvulus and/or duodenal obstruction did not occur in any patient.

A diaphragmatic hernia or eventration was found in three infants and the question is raised whether it is advisable to explore the abdomen for remediable conditions rather than close skin over the intact sac, which is desirable when a two-stage repair is contemplated.

There were only three survivors in a group of 13 infants in whom the abdominal defect exceeded 5 cm. in diameter; in 11 instances the sac contained part of the liver. In two of the three survivors

the skin alone was closed at the first operation and repair was satisfactorily completed in two stages.

A study of the 10 deaths in this group showed that cardiac malformations were responsible for six, and in retrospect there seems to be a place for a non-operative method of treatment in this type of patient, especially when born prematurely and when pulmonary atelectasis is present.

Factors considered to determine the outcome in an infant with exomphalos are (1) the size of sac and the defect in the abdominal wall, and the presence of the liver within it; (2) pre-partum rupture of the sac; (3) coexistent malformations; (4) prematurity; and (5) early operation.

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REFERENCES

- Ahlfeld, F. (1899). Der Alkohol bei der Behandlung inoperabler Bauchbrüche. *Mschr. Geburtsh. Gynäk.*, 10, 124.
- Frazer, J. E. (1931). *A Manual of Embryology*. Baillière, Tindall and Cox, London.
- Grob, M. (1957). *Lehrbuch der Kinderchirurgie*. Thieme, Stuttgart.
- Gross, R. E. (1948). A new method for surgical treatment of large omphaloceles. *Surgery*, 24, 277.
- Hamilton, J. (1806). In Cooper, A. (1807). *The Anatomy and Surgical Treatment of Crural and Umbilical Hernia*, part 2, p. 56. Longman, London.
- Hey, W. (1803). *Practical Observations in Surgery*, p. 226. Cadell and Davies, London.
- Johnston, T. B. (1913). Extroversion of the bladder, complicated by the presence of intestinal openings on the surface of the extroverted area. *J. Anat. Physiol. (Lond.)*, 48, 89.
- Mery, J. (1716). Description de deux exomphales monstreuses. *Mém. Acad. roy. Sci.*, p. 136.
- Paré, Ambroise (1634). *The works of that famous Chirurgion*, Book 24, chap. 66, p. 959. Th. Cotes and R. Young, London.
- Rickham, P. P. (1960). Vesico-intestinal fissure. *Arch. Dis. Childh.*, 35, 97.
- Scarpa, A. (1812). *Traite pratique des Hernies*. Gabon, Paris.
- Schwalbe, E. (1909). *Die Morphologie der Missbildungen des Menschen und der Tiere*. Fischer, Jena.
- Wollenweber, E. J. and Coe, H. E. (1959). Conservative management of eventration in the newborn, with survival. *Amer. J. Surg.*, 97, 769.