RUPTURE OF EXOMPHALOS AND GASTROSCHISIS*

BY

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Exomphalos is a rare condition. Jarcho (1937) reports an incidence of one in 6,600 live births. During the past nine years over 1,400 cases have been admitted to the Alder Hey Neonatal Surgical Unit and only 48 of them were infants with an uncomplicated exomphalos. During this period about 400,000 children were born in the area served by the Unit and at the same time over 120 infants with oesophageal atresia were admitted (Table 1).

Postnatal Rupture of Exomphalos

In the past it has frequently been asserted that in a considerable percentage of children born with an exomphalos, rupture of the protecting membrane occurs either during or shortly after birth. In the Boston Children's Hospital series (Gross, 1953) rupture of the sac occurred in 20% and in the Birmingham series studied by McKeown, MacMahon and Record (1953) over a third of the cases suffered from this complication. There was a mortality rate of 64% in the Boston and of 74% in the Birmingham series respectively.

Both the incidence and the mortality figures for postnatal rupture of the exomphalos were published 10 years ago and are somewhat out of date. With improved technique in delivery, rupture of the sac should become a rarity and we have only twice seen this condition during the past nine years (Fig. 1). Both these cases were operated upon soon after the rupture occurred and both recovered. If the exomphalos is repaired soon after rupture the mortality should today be hardly higher than that following operation for uncomplicated exomphalos.

That recovery can occur even under the most unfavourable circumstances can be seen from the remarkable description by Reed (1913), of one of the earliest successful cases that I could find in the published material.

'I was called to attend Senora Y.A., a Mexican woman in confinement on March 14th, 1913. I found that the head of the infant was already free and with the next pain a moment later the trunk was expelled. I was

astonished at finding that the whole intestine, both small and large, was outside the abdominal cavity. Examination showed that the bowel had passed along inside the cord for about 2 inches at which point the walls of the cord had ruptured allowing the bowel to escape laterally.

'No preparation for the confinement had been made; the bed was filthy dirty and the mass of intestine was thickly sprinkled with bits of straw, feathers, crumbs of food and faecal matter from the mother.

'... I hurriedly ligated the cord, delivered the placenta and wrapping the baby in the cleanest thing I could find, I called my colleague Dr. T. B. Smith and we went together to see the disemboweled infant and took it at once to the Arizona Copper Company's Hospital. It was placed on the operating table two hours after birth. By this time the bowels were matted together with fibrous adhesions which included many of the particles of debris mentioned above. They were cleaned gently with sponges and warm salt solution, but the cleaning was not very thorough of course.

'The umbilical opening admitted the tip of two fingers. It was enlarged for half an inch upwards and downwards and the cord bearing edges were trimmed off. The intestines were then replaced and a hurried closure was made. . . . The child made an uneventful recovery save for one small stitch abscess.'

Antenatal Rupture of Exomphalos: Gastroschisis

Perusal of the literature seems to suggest that antenatal rupture of exomphalos is very rare. In 1957 Kiesewetter could find only 10 cases in the literature, but he overlooked a considerable number (see Johns, 1946; Hollenberg, 1948; Parkkulainen, 1959); as far as I can ascertain only two further cases have been published since (Parkkulainen, 1959; Leroux and Leroux, 1961). The condition

TABLE 1
INCIDENCE OF EXOMPHALOS

	Number
Admissions: nine years Neonatal Surgical Unit (total number of births about 400,000)	1,400
Uncomplicated exomphalos	48
Postnatal rupture	2
Antenatal rupture	13
Total	63

* A paper read at a meeting of the British Association of Paediatric Surgeons in London, September 1962.

FIG. 1.—Large exomphalos which ruptured during labour.

FIG. 1.

FIG. 2.—Gastroschisis. Case 2: The stomach and the whole of the small intestine have prolapsed. The length of the small intestine was only 50 cm.

FIG. 2.

is undoubtedly much more common than is indicated by these figures. It is likely that as many of these babies are stillborn, grossly deformed or premature (Johns, 1946); they are only rarely operated upon and hardly ever recover; they are, therefore, only rarely reported in the medical journals. As all infants with severe congenital malformations born in our region are admitted to our Unit regardless of their birth weight, multiplicity of deformities or general condition, we have seen a number of these almost hopeless cases. In addition to the 48 cases of uncomplicated exomphalos and the two cases of postnatal rupture, we have admitted, during the past nine years, 13 infants with antenatal rupture of the exomphalos.

Embryology and Pathology. This condition is usually called gastroschisis in the American journals and has been defined by Moore and Stokes (1953) as a hernial abnormality of the anterior abdominal wall, which occurs in an extra-umbilical position and where there is no membranous covering of the intestine. The opening in the abdominal wall is always lateral to the umbilical cord which appears to have a normal attachment. In some cases there is a strip of apparently normal skin between the opening and the attachment of the umbilical cord. In some infants there are remnants of the exomphalos sac attached to part or the whole of the circumference of the abdominal opening; there were three such cases in our series. These cases were otherwise identical with those where there was no remnant of the sac present. It is striking that in all our cases and in most of the cases reported in the literature the opening was to the right of the umbilicus. It is suggested that the exomphalos sac ruptures during intrauterine life. The position of the umbilical vein and ligamentum teres usually determines the site of the prolapse of the intestine to the right of the

umbilical cord. In all our cases the whole of the small and large gut from the stomach to the sigmoid colon was eviscerated. There was complete non-rotation, the intestine being suspended from a common primitive dorsal mesentery. In all cases the gut was dilated, the walls being enormously thickened and oedematous and of the consistency of rubber. There was usually gross shortening of the small intestine which, in spite of its large diameter, was only between 25 and 75 cm. in length (Fig. 2). The intestines were always closely adherent to each other and were frequently surrounded by oedematous, gelatinous material. The intestinal circulation was always impaired and the colour of the intestines was dark purple or blue with subserous haemorrhages. In two of our cases the gut was frankly gangrenous. The caecum was usually conical in shape and the appendix arose from its apex and was short and wide. A greenish-yellow or haemorrhagic membrane was often closely attached to part of the intestine. This 'membrane' was not attached to the abdominal opening and on histology showed no cellular structure. In no case could intestinal peristalsis be observed during operation.

Of our 13 cases, 10 were premature infants. In two infants there were gross additional malformations. One child had a meconium ileus, the other, a mongol with a gross cardiac malformation, had valves in the posterior urethra and cerebral angiomas.

Treatment

Treatment is, as yet, most unsatisfactory and the mortality is very high (Table 2). About a dozen survivals have been reported (Watkins, 1943; Hardaway, 1954; Hollenberg, 1948; Parkkulainen, 1959).

TABLE 2

	Number
Died before operation	1
Prolonged resuscitation	1
Operation within a few hours of birth	11
Resection of part of gut	1 (fatal)
Enterostomies	4 (all fatal)
Suture of abdominal skin only	1 (fatal)

The prolapsed intestine should be covered immediately with sterile moist dressings, a catheter passed into the stomach and gastro-intestinal suction started. Unfortunately, in our cases this did not decrease the bulk of the prolapsed intestinal mass to any significant degree, as this was not due to intestinal distension but to the thickness of the intestinal wall.

At operation the abdominal opening was usually enlarged. The intestine was cleaned and the gelatinous matrix and pseudo membrane removed as far as possible unless this procedure caused profuse haemorrhage. An attempt was then made to

FIG. 3.—Gastroschisis. The abdominal skin was mobilized and closed over the prolapsed intestine.

TABLE 3
RESULTS

	Number
Surviving cases	2
Fatal cases	11
Died before operation	1
Died on table	2
Died within three hours after operation	2
Died within eight to 36 hours after operation	6

TABLE 4
FINDINGS AT AUTOPSY (10 CASES)

Findings	Number
Lungs not expanded	10
Thrombosis left pulmonary artery	1
Sclerema neonatorum	4
Peritonitis	3
Intraperitoneal haemorrhage	3
Thrombosis superior mesenteric vein	1
Intracranial haemorrhage	2

return the intestine into the abdominal cavity. The enormous bulk of the intestine and the small size of the abdominal cavity made this a very difficult and dangerous procedure. Closure of the abdomen was always under considerable tension and the respiratory movements of the diaphragm were often impeded. Resection of part of the gut in order to reduce the bulk is usually inadvisable because the intestine is already too short. It was unsuccessfully attempted in one of our cases. In many cases the only procedure that can be done is undermining of the abdominal skin and closure of the skin over the prolapsed bowel. Some of the successful cases reported were treated in this way and so was one of our cases (Fig. 3). Enterostomies in order to deflate the gut were unsuccessfully attempted in four cases.

It is not surprising that deaths from shock or post-operative respiratory embarrassment are very common. In some of the more recent cases we have attempted to combat post-operative respiratory embarrassment by connecting the infant to a respirator, but, although this prolonged life, it was not permanently successful. Even if shock or respiratory troubles are not fatal, death may be caused by prolonged ileus following the operation. The thickened oedematous gut is not capable of peristalsis for many days and the patient will need intestinal suction and intravenous therapy over long periods. If the patient survives this complication, one is frequently left with an infant with a very short length of intestine unable to absorb sufficient nourishment from the gut. This is illustrated by the case reported by Kiesewetter (1957), which, although feeding started 48 hours after operation, succumbed at 6 weeks of age from malnutrition and septicaemia.

Results

In our series of 13 cases (Table 3), one child, with multiple abnormalities was not fit for operation and died two hours after admission; 12 were operated upon. Two cases died on the table, two within a few hours and six between eight and 36 hours after operation. With the exception of one case (the child with meconium ileus) autopsy was carried out in all infants (Table 4). The most striking finding was that in all cases the lungs had hardly expanded at all and in a number of infants the pathologists were surprised that the child was able to breathe at all. Sclerema neonatorum occurred in four of the very premature infants. Death was, therefore, primarily due to pulmonary complications.

Two children have survived. The first patient, operated upon by my colleague, Miss I. Forshall,

FIG. 4.—Case 1: Gastroschisis; the patient at 5½ years.

was a boy weighing 6 lb. 1 oz. (2,740 g.). The prolapsed intestine was moderately thickened and oedematous and could be replaced with difficulty into the abdominal cavity. The abdomen was sutured under considerable tension. There was a prolonged ileus lasting seven days, necessitating gastric suction and intravenous therapy. Feeding was commenced on the eighth day and the boy then made an uninterrupted recovery. He is now 5½ years old (Fig. 4).

The second patient, a premature girl, weighing 4 lb. (1,800 g.) on admission, had some sclerema, a temperature of 94° F. (34.4° C.) and Cheyne-Stokes respiration. The prolapsed gut was about a third of the normal length. There were patches of doubtful viability in the walls of the intestine. The gut was returned with difficulty into the abdominal cavity and the abdomen was sutured under tension. Ileus persisted for eight days. A faecal fistula developed on the tenth day. The child suffered from intestinal

FIG. 5.—Case 2 at 10 months of age. A premature infant with gastroschisis.

hurry and malabsorption for the first three weeks and would not have survived but for continuous intravenous therapy and especially the daily administration of intravenous fat emulsion over a period of two weeks. The faecal fistula healed spontaneously when the baby was 3½ weeks old. The child then made an uninterrupted recovery. She weighed 12 lb. 3 oz. (5,500 g.) when 4 months old and is now 10 months old weighing 19 lb. (8,550 g.) (Fig. 5).

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