EXOMPHALOS

BY

A. A. CUNNINGHAM

From the Paediatric Unit, Kingston Hospital, Kingston-upon-Thames

(RECEIVED FOR PUBLICATION DECEMBER 22, 1955)

Exomphalos is a rare condition in which there is a congenital malformation of the umbilical cord, alone or combined with a defect of the supra-umbilical region of the abdomen, and through which the viscera herniate into a sac covered by amniotic membrane and peritoneum. The sac may vary in size in its largest diameter to as much as 15 cm. and, according to size, will contain varying amounts of intestine, stomach and liver. The neck of the sac is surrounded by skin with a clear line of demarcation, and occasionally the skin extends a short way up the side of the sac. Exomphalos includes conditions described under the following names: omphalocoele, amniocele, amniotic hernia and hernia into the umbilical cord.

Embryological Aspect

The condition at birth resembles the appearance in embryos before the tenth week of foetal life with the coelomic cavity as a forward extension into the base of the cord, the pocket containing intestine and other abdominal viscera. Normally, after the tenth week, the organs are withdrawn into the abdominal cavity. There have been a number of theories in regard to aetiology:

1. It is due to retardation in the development of the abdominal cavity about the third month of foetal life, causing the abdominal organs to remain in the cord pocket because of insufficient room in the abdomen.

2. It is due to a disparity between the size of the abdominal cavity and the viscera, which has resulted from retarded development of the abdominal parietes.

3. The following theory is put forward by Margulies (1945), after study of the work of Pernkopf (1925), Politzer and Sternberg (1930), who each studied the formation of the anterior abdominal wall of human embryos in the early weeks of pregnancy. According to Margulies the combined umbilical and supra-umbilical defect follows a failure of development about the third week of embryo life, whereas a purely umbilical ring defect originates at the eighth to tenth week. In the latter case there is a normal anatomy of the upper abdominal aponeurosis. The supra-umbilical defect is due either to failure of the amniotic covering to become closely adherent to the transverse septum, or failure of the connective tissue of the transverse septum to proliferate and thus to push the umbilical covering downwards towards the umbilical pedicle. An umbilical ring defect could be caused by failure in the proliferation of the adventitial connective tissue of the umbilical vessels.

Those who accept Margulies' view differentiate hernia into the umbilical cord from the combined umbilical and supra-umbilical defect, which they classify as 'omphalocoele'. In hernia into the umbilical cord there is generally only intestine outside the umbilical ring, the neck of the defect having a diameter of about 4 cm. or less, the sac when unruptured being covered by peritoneum and amniotic membrane. In omphalocoele the sac is also covered by peritoneum and amniotic membrane, but the neck of the sac is generally larger than 4 cm. in diameter, and the sac may contain not only small and large intestine, but other organs such as stomach and liver, thus presenting a much more difficult problem in treatment. Although there may be justification at times in separating the conditions from the embryological concept, it is simplest in reviewing the literature to retain the inclusive term 'exomphalos'.

The Literature

An extensive review by Jarcho (1937), covering about 450 cases, did not show any hereditary tendency. On the other hand in a short series of seven cases, Benson, Penberthy and Hill (1949) included a brother and sister, each showing a hernia into the umbilical cord. Paucot and Gellé (1936) described a similar condition in three successive pregnancies in a young woman.

Associated congenital abnormalities occur with more than coincidental frequency. In Ladd and
EXOMPHALOS

Gross's series of 88 cases 59% had some other abnormality (Ladd and Gross, 1941), in most cases of minor degree. Twenty-eight per cent. had malrotation of the intestine with intestinal obstruction, requiring operation at the time of repair or subsequently.

Since the latter half of the nineteenth century various efforts have been made to treat the condition surgically. A review of the literature, however, gives a biased impression, and one must assume that with large examples of exomphalos there has been a tendency to report only the successful or near-successful results. Earliest successes were in cases which we would now classify as 'hernia into the umbilical cord'. The earliest was published by Visick in 1873. Aribat (1901) and Walravens (1902) between them collected about 200 cases up to 1900. Altpeter (1931) collected 109 cases between 1900 and 1929, adding three of his own. Jarcho (1937) collected 46 cases between 1929 and 1937, adding one of his own. Of these 47 cases, 36 were submitted to radical surgical operations, and in these there were only five deaths. Massabauu and Guibal (1933) collected 23 cases, mostly from the European literature and added one of their own. Operations had been performed in 12 cases where the abdominal defect was 4 cm. or less in diameter, and there were nine recoveries. Jarcho, in his analysis, reported that of 159 cases since 1900 the liver was in the sac in 46 cases, and of these 12 recovered after operation, giving a recovery rate of about 25%.

A truer picture, however, is given by Gross (1953), who states that in patients treated in a series from 1940 to 1950 inclusive, the mortality rate was 34% for those accepted for surgery. Of the 20 deaths in the series, four were due to congenital heart disease, and one to an infected meningocele. In six of the children who survived there was a subsequent operation for the treatment of intestinal obstruction due to malrotation of the intestine.

Many of the deaths in the early cases were due to injudicious crowding of the viscera into the abdomen.

---

**Table: Recovery with Surgical Treatment**

<table>
<thead>
<tr>
<th>Weight</th>
<th>Diameter of Abdominal Defect (cm.)</th>
<th>Contents of Sac</th>
<th>Age at Operation (hr.)</th>
<th>Type of Operation</th>
<th>Result</th>
<th>Author</th>
</tr>
</thead>
<tbody>
<tr>
<td>7 lb. (3,175 g.)</td>
<td>11 1/25 × 6 25</td>
<td>Whole liver, most of intestine</td>
<td>12</td>
<td>Sac excised, one-stage closure</td>
<td>Ventral hernia</td>
<td>Hipsley (1929)</td>
</tr>
<tr>
<td>7 lb. (3,402 g.)</td>
<td>Hernia 'size of child's head'</td>
<td>Entire liver, two-thirds stomach, pancreas and most of intestine</td>
<td>—</td>
<td>Sac excised, one-stage closure</td>
<td>Good result</td>
<td>Dott (1932)</td>
</tr>
<tr>
<td>6 lb. 14 oz. (3,118 g.)</td>
<td>—</td>
<td>Whole liver</td>
<td>12</td>
<td>One-stage closure</td>
<td>Strong scar when 3 years old</td>
<td>Jarcho (1937)</td>
</tr>
<tr>
<td>6 lb. 10 oz. (3,005 g.)</td>
<td>7</td>
<td>Liver, stomach, small intestine</td>
<td>8</td>
<td>First-stage closure</td>
<td>Second-stage operation 22 mth., good result</td>
<td>Benson <em>et al.</em> (1949)</td>
</tr>
<tr>
<td>—</td>
<td>Size of tennis ball</td>
<td>Larger part liver, large and small intestine</td>
<td>36</td>
<td>One-stage repair</td>
<td>Ventral hernia followed with more or less spontaneous cure by 1954</td>
<td>Nash (1950). (Surgeon, C. Donald. Subsequent report, H. H. Nixon)</td>
</tr>
<tr>
<td>Full term</td>
<td>10</td>
<td>Liver, small intestine</td>
<td>48 approximately</td>
<td>Sac excised, attempt at one-stage repair</td>
<td>Resuture of wound 8 days after first operation and 8 days after second. Awaiting repair of two incisional hernias</td>
<td>Fox (1951)</td>
</tr>
<tr>
<td>Full term</td>
<td>11</td>
<td>Almost entire liver, part of small intestine</td>
<td>10</td>
<td>Sac excised, diaphragmatic hernia present, skin closure only</td>
<td>Recovery from first operation. Two subsequent attempts at repair, but large ventral hernia still present at 3 yr.</td>
<td>Welch (1951)</td>
</tr>
<tr>
<td>5 lb. 8 oz. (2,495 g.)</td>
<td>10</td>
<td>Most of liver, spleen and large and small intestine</td>
<td>58</td>
<td>Sac not opened, first-stage closure with difficulty</td>
<td>Skin flap necrosed 6 days after operation and treated by skin graft. Second repair operation due at 14 months</td>
<td>Welch (1951)</td>
</tr>
</tbody>
</table>
in a one-stage operation, which caused heart failure due to pressure on the inferior vena cava by the forcibly replaced liver. Gross, in 1948, advised not opening the amniotic sac at the first operation, but instead freeing, undercutting and approximating the skin margins over the sac, thus preventing adhesion of the abdominal viscera to the subcutaneous tissue. A second-stage repair operation could be delayed until the child was 6 to 12 months of age. Other surgeons, however, while accepting the advance of the two-stage operation, have remained of the opinion that the sac should be opened at the initial operation, because of the danger of associated intra-abdominal congenital malformations.

The Table lists certain cases, in which the exomphalos was relatively large, and are some of the best examples of recovery with surgical treatment.

I have been privileged to see some notes and photographs of a patient treated in The Hospital for Sick Children, Great Ormond Street, London, by D. L. B. Farley (1955), who operated on a baby boy under the care of G. H. MacNab in 1953. There was a large exomphalos containing the liver and most of the intestine, with measurements at the base 6 cm. by 5 cm. The sac was removed at the first operation, and a one-stage repair attempted. The wound broke down, and as closure was impossible, a large patch of amnion was sewn in place overlapping the defect approximately one inch all round. The skin again broke down, but the amnion remained intact, preventing evisceration. The wound was re-sutured, and the child recovered. In November, 1955, he was reported as fit and well, but with an enormous ventral hernia, and with the lower ribs growing inwards, tending to bisect the liver.

Before 1940 rupture of the sac before operation was mostly fatal, but since then the advent of chemotherapy and the wide selection of antibiotics have improved the outlook. If the rupture has taken place after birth, the prognosis is better than where it has occurred during intra-uterine life and the intestines have become oedematous and matted together.

Conservative Treatment

Only brief mention of this is to be found in the literature. One of the early methods of treatment was to apply a protective bandage, without any attempt to reduce the hernia. Arribat (1901) lists some cases of this type. In 1899 Ahlfeld described a method whereby the hernia was reduced as far as possible under light narcosis after careful cleansing of the sac and surrounding skin, followed by alcohol compresses. In the first case he treated the liver was in the sac, and complete reduction was impossible, but the child thrived without operation, and one year later the scar was excised and the wound closed. In Jarcho's review of 46 cases, reported between 1929 and 1937, there was one recovery with Ahlfeld's method, but the child was accidentally killed at the age of 7 months. Ahlfeld's method, however, never found favour. Gross (1953) stated that in one of their personal cases the skin of the abdominal wall had started to grow up over the margins of the sac, and they were encouraged to think this would continue, so operation was deferred, but unfortunately infection set in and the infant died. Ellison Nash (1950) reported a case of an exomphalos the size of a large orange at birth. Cure was spontaneous, leaving an umbilical hernia with a ring 2.5 cm. in diameter. The dimensions of the neck of the sac were not given. Nash recommended a dressing of penicillin and sulphonamide cream. Denis Browne (1955) has successfully treated two cases of the hernia into the cord type by simply twisting the cord so as to squeeze the contents back into the abdomen, and then strapping down as for an umbilical hernia.

In view of the poor results experienced in the surgical treatment of cases of large exomphalos, it was decided in 1948, without knowledge of Ahlfeld's work, to try conservative treatment using absolute alcohol dressings in conjunction with an antibiotic, penicillin. Two cases were treated with results as follows:

Case Reports

Case 1. R.H., a boy, was born normally on December 5, 1948 (weight 5 lb. 10 oz. 2,551 g.). Exomphalos with unruptured sac contained intestine, stomach and part of the liver. The sac was 6 cm. in diameter at the neck. Absolute alcohol dressings were applied twice daily.


December 13. Sac moist, black and necrotic in appearance. Swabs showed non-haemolytic streptococci only. Penicillin treatment begun, 30,000 units, four hourly, by injection.

December 23. Feeding satisfactory. No vomiting. Exomphalos area shrinking. Less discharge on dressings. Baby's weight 5 lb. 11 oz. (2,580 g.).

December 29. Sac wall hard, dry and black. Small area of moist granulation tissue at the edge. Haemoglobin 90%.

December 30. Progress maintained. Swab from exomphalos showed Staphylococcus aureus sensitive to penicillin. Penicillin injections discontinued, but penicillin continued by mouth.
January 4, 1949. The granulating area uncovered by skin had contracted to 5 cm. in diameter.


January 11. Exomphalos area more moist. Penicillin again started by mouth. Alcohol discontinued. Eusol dressings applied. Weight 7 lb. (3,175 g.).


February 4. General progress satisfactory. The granulating area of the exomphalos had contracted to about 1 cm. in diameter. Eusol dressings discontinued. Scarlet red ointment applied. Weight 8 lb. 4 oz. (3,742 g.).

February 13. Exomphalos healed. Left inguinal hernia noted for the first time. Penicillin discontinued. The child was discharged from hospital, 70 days after birth. Weight 9 lb. (4,082 g.).

Follow-up. The baby was seen again on May 30 when he was 6 months old. Weight 15 lb. (6,804 g.). Normal development. There was an abdominal herniation 5 cm. wide, 6½ cm. long, with puckered skin 2 cm. in diameter in the central area. A large left inguinal hernia present.

July 13. Developed ischio-rectal abscess. Treated surgically. Quick recovery. Bilateral inguinal hernias noted, left much larger than the right.

On January 22, 1950, when the child was 1½ years old, recurrent vomiting was reported. Inguinal hernias present. The child had stood up for the first time at between 10 and 11 months of age, and was walking quite well when seen. He started to talk soon after 1 year of age.

On January 28 he was admitted for left inguinal herniotomy.

On April 13, 1951, he was admitted with complaint of attacks of abdominal pain with screaming and occasional vomiting. The child settled quickly in hospital. Radiological investigation was negative.

On November 30, 1953, the child weighed 37 lb. (16.4 kg.), was very well apart from a central abdominal defect with herniation, extending from the xiphisternum to the umbilicus. The bulge on standing was 5 cm. by 9 cm.

On October 13, 1954, the mother again gave a history of intermittent abdominal pain with some abdominal distension. The herniation on standing was 7·5 cm. by 10 cm.

On March 5, 1955, the child was admitted with the history of frequent attacks of abdominal pain and vomiting. Radiological examination was again negative. It was decided to repair the abdominal hernia defect. On March 16 operation was performed by Mr. R. H. Franklin. A transverse incision was made and the puckered area in the middle of the scar was excised. The peritoneum was opened showing numerous adhesions between coils of intestine. Several adhesions were divided. The appendix was removed. The hernia was closed with a Mayo type repair using catgut. The child made an uneventful recovery after operation, which produced a very satisfactory cosmetic result (Fig. 2).

On May 2, there was again a history of recurrent abdominal pain and occasional vomiting, but conservative measures were adopted. The symptoms were probably due to adhesions.

Case 2. S.C., a boy, was born on November 15, 1949. Birth weight was 6 lb. 15 oz. (3,147 g.). A large exomphalos was present, the sac unruptured, 8 cm. at the neck (Fig. 3). It contained liver, stomach and the greater part of the intestines. Absolute alcohol dressings were applied twice daily. Penicillin, 60,000 units three hourly by mouth, was given.

November 18. The child's condition had deteriorated considerably since birth. He was very ill with poor colour and rapid respirations and intermittent vomiting. There were signs of dehydration. Penicillin was changed to the intramuscular route, and 100 ml. of 4·3% glucose in 1·5 normal saline given intramuscularly.

November 19. Vomiting frequently, coffee ground in type. Well marked dehydration, and 150 ml. normal saline given intravenously followed by slow drip using 4·3% glucose in 1·5 normal saline.

November 21. Condition remained very poor.
Vomiting less marked. One large stool with excess mucus. Feeding by pipette and oesophageal tube, using expressed breast milk in small quantities, retained.

November 22. Very weak—grey. Generally about one large vomit in 24 hours. Intravenous drip changed to intramuscular drip.

November 23. Still extremely ill. Vomited twice in the previous 24 hours. The abdomen was distended, but the motions were normal in appearance. Intramuscular fluid therapy continued.

November 24. Abdomen distended. The exomphalos area was black and gangrenous in appearance with offensive discharge. Absolute alcohol dressings continued.

November 25. There was slight improvement. The vomiting continued, but small feeds up to 6 drachms E.B.M. every one-and-a-half hours were for the most part retained. Penicillin continued. Absolute alcohol dressings continued.

November 28. The abdomen became more distended. The respiration rate was raised, and there were recurrent attacks of cyanosis. The exomphalos area was moist with offensive discharge, increasing in amount, but the skin was growing in satisfactorily from the edge. Absolute alcohol dressings alternated with 'eusol' dressings.

November 29. The child was weak and dehydrated. Respirations rapid and distressed. Abdomen distended. Vomiting more frequently. Stools normal. The child responded to intramuscular fluid therapy. Weight 6 lb. 2 oz. (2,778 g.).

December 1. Improved. Temperature still raised to 100° F. Intramuscular drip continued.

December 3. Temperature raised. General condition improved. There was an offensive discharge from the exomphalos area. By this time the child was taking breast milk, 1½ oz. two hourly by bottle, without vomiting.

December 6. Temperature between 100° F. and 102° F. Vomiting had begun again, so feeds were reduced in amount. Motions normal. Weight 6 lb. 6 oz. (2,892 g.).

December 8. Temperature normal. Small feeds retained well. General condition improved.

December 10. Covering of the exomphalos area thick and greenish-black with a raw area 8 cm. in diameter. The skin was growing in at the edge, and the lateral bulging of the swelling was less marked.

December 12. Vomiting re-started with rapid respiration and recurrent cyanotic attacks, probably following an aspirated vomit. Intramuscular saline again given.

December 13. Vomiting in small amounts continued. Intramuscular glucose saline continued. Weight 6 lb. 7 oz. (2,920 g.).


December 22. General condition improved, but occasional vomiting interrupted progress. The surface of the exomphalos still had a black area of necrosis, but skin edges were cleaner in appearance, and about 7½ cm. apart. The tumour swelling was 6½ cm. above the general level of the abdomen, and the widest diameter was 8 cm.

December 24. The child was taking 2½ oz. National
dried milk three hourly. No vomiting. Absolute alcohol and 'eusoil' dressings continued alternately.

**December 29.** Good progress maintained. The child reported as seeming hungry for the first time. The skin healing continued, and the discharge from the exomphalos was less offensive.

**January 1, 1950.** Penicillin, which had been given continuously from birth, was discontinued. Haemoglobin 68%.

**January 13.** Weight 8 lb. 1 oz. (3,657 g.). The raw exomphalos area was reduced to 5½ cm. in diameter.

**January 24.** Blood transfusion, 50 ml. of fresh blood given.

**February 2.** Good progress maintained. The raw area had decreased to between 4 cm. and 5 cm. in diameter. Scarlet red ointment only applied.

**February 7.** Area of granulation tissue had contracted to a circle about 2½ cm. in diameter. Weight 9 lb. 8 oz. (4,309 g.).

**February 28.** Again recurrence of vomiting, but without abdominal distension. There were several septic skin spots. Penicillin again given intramuscularly, 30,000 units four hourly.

**March 3.** Vomiting had ceased when the sepsis was brought under control. The raw area was contracting quickly.

**March 5.** Penicillin discontinued.

**March 23.** Exomphalos almost healed. There was an area of unhealthy central scar tissue with puckered skin, which would break down intermittently, producing a raw area with serous discharge, which would then crust and break down again.

On March 6 the patient was discharged from hospital (weight 12 lb. 8 oz., 5,670 g.) after a period of treatment extending over 143 days.

He was followed up as an out-patient. On March 25 there was still a small, unhealthy crust less than 1 cm. in diameter in the central part of the exomphalos area. The child's general condition was very good.

By June 20 the wound had closed completely. There was, however, a considerable central abdominal herniation with a muscle gap of about 8 cm.

By August, 1950, good progress was maintained, and development was generally normal. A belt was ordered to control the large abdominal herniation.

By October 10 the baby could sit up well, and was beginning to crawl. The mother herself, with considerable ingenuity, had made a binder which was much more satisfactory than the official truss provided.

When seen on January 9, 1951, the child was beginning to speak. He could crawl well. The muscle gap was well controlled by the home-made belt, although the central herniation scar was delicate and tissue-like in consistency.

When he was 16½ months old, on April 3, he had been walking for one month. The mother's only complaint was of occasional constipation requiring medicine from time to time.

When seen on November 13 the child was very well. He had a large herniation extending from the ensiform cartilage to the umbilicus with wide divarication of the recti.

His weight on September 3, 1952, was 29 lb. (13·1 kg.) and height 89 cm. His general progress was good, and he was reported to be of above average intelligence. The abdominal defect was less noticeable, and the skin over the exomphalos more healthy in appearance.

On November 3, 1953 (weight 32 lb., 14·5 kg.; height 98 cm.), he had a central abdominal defect 7·5 cm. by 15 cm. when lying down and 6·25 cm. by 11 cm. when standing. There was also considerable foreshortening of the sternum. Generally, however, there was good posture apart from slight valgus knee deformity.

On January 25, 1955, when 5 yr. 3 mth. (weight 37 lb., 16·4 kg.; height 113 cm.), the child's general health was good apart from attacks of vomiting brought on by articles of diet which he disliked. There was little complaint of abdominal pain. His general progress at school was very good, and he was rarely absent.

On examination there was a herniation defect when standing 7·5 cm. by 9 cm.; when lying flat with the head raised 12·5 cm. by 6·5 cm.; but when lying quietly at ease only a slight bulge 6·5 cm. vertically was noticed. There was a small central, puckered scar 4 cm. in diameter. There was little disability, so it was decided to defer any question of a repair operation until a later date.

**Discussion**

There is a general statement to the effect that exomphalos occurs about once in every 5,000 births. The figures for three large maternity units in this area are of interest:

<table>
<thead>
<tr>
<th>Unit 1</th>
<th>Yearly number of live deliveries approximately</th>
<th>2,000</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Births over five years</td>
<td>10,000</td>
</tr>
<tr>
<td></td>
<td>Exomphalos</td>
<td>small 2, large 1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unit 2</td>
<td>Yearly number of live deliveries approximately</td>
<td>1,500</td>
</tr>
<tr>
<td></td>
<td>Births over five years</td>
<td>7,500</td>
</tr>
<tr>
<td></td>
<td>Exomphalos</td>
<td>small 1, large 2</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unit 3</td>
<td>Yearly number of live deliveries approximately</td>
<td>1,300</td>
</tr>
<tr>
<td></td>
<td>Total over five years</td>
<td>6,500</td>
</tr>
<tr>
<td></td>
<td>Exomphalos</td>
<td>small 1</td>
</tr>
</tbody>
</table>
ARCHIVES OF DISEASE IN CHILDHOOD

Table 3

DEATHS FROM CONGENITAL EXOMPHALOS IN 1951 AND 1952

<table>
<thead>
<tr>
<th></th>
<th>Sex</th>
<th>1 Week</th>
<th>Under 30 min</th>
<th>Over 30 min. and under One Hour</th>
<th>Over One Hour and under One Day</th>
<th>1 Day</th>
<th>2 Days</th>
<th>3 Days</th>
<th>4 Days</th>
<th>5 Days</th>
<th>6 Days</th>
<th>7 Days</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mention of other congenital malformations</td>
<td>M</td>
<td>3</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No other congenital malformations</td>
<td>M</td>
<td>12</td>
<td>1</td>
<td></td>
<td></td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>11</td>
<td>1</td>
<td></td>
<td></td>
<td>2</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mention of operation</td>
<td>M</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No operation</td>
<td>M</td>
<td>13</td>
<td></td>
<td></td>
<td></td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>11</td>
<td></td>
<td></td>
<td></td>
<td>1</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total, 1951</td>
<td>M</td>
<td>15</td>
<td>1</td>
<td></td>
<td></td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>13</td>
<td>1</td>
<td></td>
<td></td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mention of other congenital malformation</td>
<td>M</td>
<td>7</td>
<td>1</td>
<td></td>
<td></td>
<td>3</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td>1</td>
<td>4</td>
<td>1</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No other congenital malformations</td>
<td>M</td>
<td>12</td>
<td>1</td>
<td></td>
<td></td>
<td>3</td>
<td>3</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>9</td>
<td>1</td>
<td></td>
<td></td>
<td>1</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mention of operation</td>
<td>M</td>
<td>3</td>
<td></td>
<td></td>
<td></td>
<td>1</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>5</td>
<td></td>
<td></td>
<td></td>
<td>1</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No operation</td>
<td>M</td>
<td>16</td>
<td></td>
<td></td>
<td></td>
<td>3</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>6</td>
<td></td>
<td></td>
<td></td>
<td>1</td>
<td>3</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total, 1952</td>
<td>M</td>
<td>19</td>
<td>1</td>
<td></td>
<td></td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>11</td>
<td>1</td>
<td></td>
<td></td>
<td>3</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Thus out of a total of 24,000 deliveries there were in all seven exomphalos cases, of which three were large. It is interesting to note, however, that in this hospital, where we have on the average 1,500 deliveries a year, we have not had a child with a large exomphalos since November, 1949.

If we take statistics for England and Wales according to the birth rate, the figures for live births would be as follows:

1951 ... ... 607,529
1952 ... ... 673,735

Working on an average incidence of 1/5,000 births, this would give a total figure of approximately 120 cases a year, of which the majority would be small and easily treated surgically.

Thanks to the cooperation of the Registrar General, who supplied me with details of deaths from congenital exomphalos in the years 1951 and 1952, we have the figures in Table 3. In some, other congenital abnormalities were mentioned in association. In 1951 there were 28 deaths and in 1952 30 deaths. If we accept the figure of 120 cases a year as an average, this would mean that a quarter of the cases died each year. It is very probable that these deaths occurred mostly in exomphalos cases with large sacs, and from the statistics it would appear that operations were only attempted in a small proportion of the fatal cases. In 1951, for example, there was mention of operation in only four out of 28 cases, and in 1952 in eight out of 30 cases. The others apparently died without treatment, unless of course the mention of operation was omitted from the death certificate. All deaths occurred within one week of birth.

Summary

The literature on the treatment of exomphalos is reviewed.

Two cases of exomphalos with large, unruptured sacs treated conservatively with quite successful results are reported, proving that conservative treatment is worthy of trial. Alcohol and 'eusol' dressings were applied and penicillin administered by mouth and by injection.

I wish to acknowledge the valuable help of my assistants, Dr. E. G. A. Crawshaw and Dr. Vivian Usborne, Sister E. Hall and Sister J. V. Cooper, and the various members of the nursing staff who were responsible for the detailed treatment of the babies. I wish also to thank the Librarian at the Royal Society of Medicine for help with references, and also my secretary, Miss J. Osmond, for assistance in the preparation of this article.

The cost of the investigation by the Registrar General was covered by a grant from the Research Fund, South West Metropolitan Regional Hospital Board.

References

EXOMPHALOS
