

Case Report

A simple technique for the repair of a complex case of exomphalos major

Nandini K. Bedi*, Alka G. Grewal

Department of Paediatric Surgery, Christian Medical College & Hospital, Ludhiana, Punjab, India

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*Correspondence:

Dr. Nandini K. Bedi,

E-mail: nandinikbedi@gmail.com, nandinikbedi@yahoo.co.in

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ABSTRACT

Various techniques have been tried and described by various surgeons to repair an exomphalos major. Our patient was a case of giant exomphalos, also called a hepato-omphalocele, presenting at 22 months of age with a major exodus of abdominal viscera. Our technique of staged repair was simple, innovative, without the use of prosthesis or elaborate equipment, did not require assisted ventilation or prolonged hospital stay and consequently cut on the cost of treatment. There were no post-operative complications and the result was very satisfactory. We wish to call it the 'Kaul-Bedi Technique' for repair of complex omphaloceles.

Keywords: Exomphalos major, Giant omphalocele, Hepato-omphalocele, Complex omphalocele, Kaul-Bedi technique

INTRODUCTION

Exomphalos is a congenital abdominal wall defect which may be varying in severity from a herniation into the umbilical cord, to a membrane covered abdominal wall defect extending from the umbilicus to the epigastric region. If the defect is up to 5 cm in diameter it is considered an exomphalos minor. A diameter exceeding 5 cm is a big defect, called exomphalos major.

The incidence of exomphalos in the west is about 1 in 10000 births¹ and in Europe about 0.77 in 10000 births.² The incidence of exomphalos major varies between 1 in 4000 to 5000 births.³ It can be diagnosed in utero by an antenatal ultrasound, which may lead to an elective caesarean section so as to avoid rupture of the covering membrane. It is important that the obstetrician is aware of the condition so that the cord is not ligated too close to the base as a small exomphalos can be mistaken for the cord itself and gut gangrene from exomphalos ligation is known to occur.

Exomphalos minor is treated by primary closure of the defect. This may be delayed if the sac is infected. Exomphalos major is a challenge for the surgeon. The aim is to return the contents to the abdominal cavity and close the anterior abdominal wall. The problems to overcome are, a lack of space in the abdominal cavity, viscera in the sac containing part or whole of the liver, which is not compressible and cannot be fitted into the small, undeveloped space available, and lastly, inability to close the abdominal wall which is deficient. An attempt at repair may lead to an abdominal compartment syndrome with difficulty in respiration/ventilation and obstructed venous flow in the abdomen. The condition is challenging and life threatening.

Various techniques have been tried and described by various surgeons. Our patient was a case of giant exomphalos also called a hepato-omphalocele presenting at 22 months with a major exodus of abdominal viscera. Our technique of staged repair was simple, innovative, without the use of prosthesis or elaborate equipment did

not require assisted ventilation or prolonged hospital stay and consequently also cut on the cost of treatment. There were no post-operative complications and the result was more than satisfactory.

CASE REPORT

Our patient was a 1 year 10 month old girl, weighing 8 kg, presenting with an exomphalos major, for definitive surgery. She had a giant exomphalos which was completely covered with skin, giving a 'pedunculated' appearance (Figure 1). The defect was approximately 12 cm in the midline and 10 cm from side to side. In supine position the exomphalos would lie 'beside the child' as seen on the plain X-ray of the child (Figure 2). On examination, the entire liver and spleen were palpated in the lesion. Looking at the X-ray, the abdominal cavity, aside from the pelvis, was extremely reduced in capacity. The lower ribs were caving in on an empty space. There was very little bowel gas seen in the abdomen, mostly the proximal and distal most bowel only. Almost the entire gut was seen in the exomphalos lesion. Ultrasonography with Doppler showed anteriorly pushed Inferior Vena Cava (IVC) with superficially placed hepatic veins, the right hepatic vein was 4.8 cm in length. Accommodating the contents of the exomphalos in the abdominal cavity was a major challenge.



Figure 1: Patient in supine position with a giant omphalocele beside her.



Figure 2: Plain radiograph of the patient's chest, abdomen and omphalocele showing crowding of the lower ribs over an empty space and almost the entire bowel gas shadow is in the omphalocele. Abdominal space can be assessed.

Staged repair was planned. The exomphalos was opened in the midline. There were adhesions of the distal ileal loop to the expected umbilical site. The expected bare surface of the liver and part of the anterior surface were adherent to the exomphalos covering. There were multiple bowel loop adhesions. The exomphalos contained the entire liver, which was globose in shape, the spleen, the stomach, the duodenum, the pancreas, large part of small bowel and major part of colon (Figure 3A). All dimensions of the liver were larger than those of the defect. The exomphalos covering consisted of skin with scarring, a thick and tight connective tissue and peritoneum lining the inner surface.

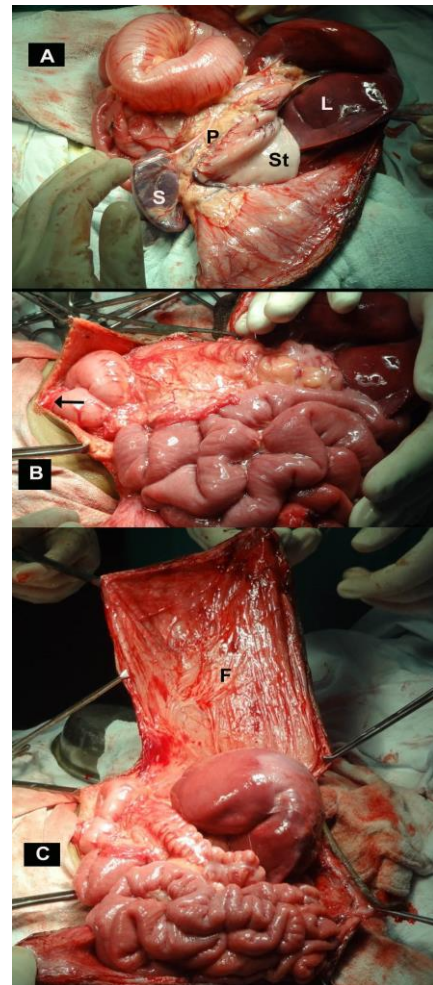


Figure 3: A) Contents of the omphalocele seen at laparotomy. Seen are L-liver, St-stomach, P-pancreas, S-spleen and also seen are duodenum, colon and loops of small bowel. B) The mid line abdominal incision is extended inferiorly up to the pubis as shown by a black arrow. C) Shown is F-the flap over the exomphalos with its peritoneal layer intact. The skin alone is repaired in the region where the incision is extended. In the region of the exomphalos the layers are not separated from the muscle, simply trimmed and repaired in the mid line, hence maintaining a tug on the muscles while applying pressure on the contents.

We decided first to widen the defect so that the liver and the other organs would be able to fall back easily. The liver being a solid organ could not be compressed hence space had to be made for it. The length of the hepatic veins, and their anterior position can lead to fatal mishaps during dissection. Keeping the above in mind we extended the mid line incision from the xiphisternum to the pubic symphysis (Figure 3B), splitting the recti and abdominal muscles in the midline. We gained about 2-3cm above and about 10-12 cm length below the defect. The skin and sub-cutaneous layer was lifted off the muscle sheath where the length was gained so as to come together in the midline freely and easily, leaving a ventral hernia underneath. The exomphalos covering was kept with the peritoneal layer intact and was trimmed freely, discarding about 7-10 cm from each side (Figure 3C), so that when pulled together in the mid line it created traction on the lateral muscles reducing the gap. Closure was done in the midline, with a little traction, using non-absorbable, interrupted sutures placed close together. It was hoped that as the peritoneal layer was left intact, there would be fewer adhesions at the time of the next procedure. The post-operative appearance is shown in Figure 4A. The child reversed from anaesthesia in the operating room and did not require ventilation or cardiovascular support.

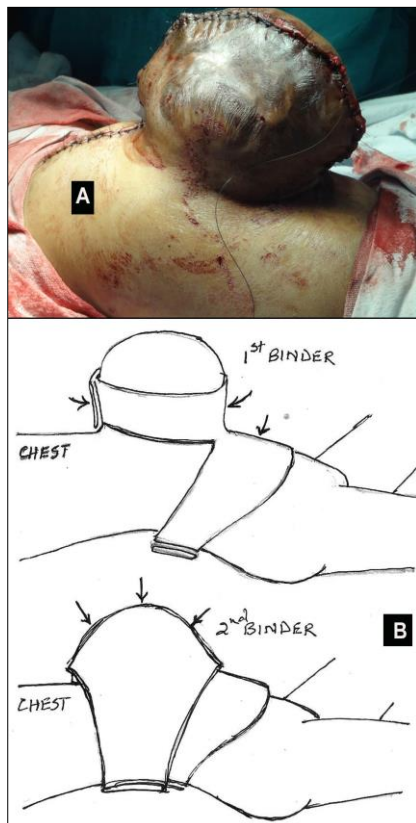


Figure 4: A) Appearance after stage I repair. B) Made to order binders and their application. The 1st binder encircles the omphalocele and the 2nd binder applies pressure from the top (arrows show direction of application of pressure).

In view of the fact that a complete ventral hernia had been created and, that the liver in its extra-abdominal situation had no triangular ligaments and was hanging precariously from its venous and biliary attachments, supporting binders were provided. These were made to order (Figure 4B). The first binder was placed inferior to the lesion with a segment encircling the base of the lesion. The aim was that compression is provided by the second binder directly from the top, pushing the contents intra-abdominally and not outside the defect. The binders were designed to provide support to the abdominal organs while ambulating as they were only covered by skin anteriorly.

By the sixth month we planned the second stage repair. Though we found some adhesions to the skin and flap, the liver was comfortably situated within the abdominal cavity (Figure 5A). Compressible bowel loops were present in the prominence of the ventral hernia. At this point stay sutures were taken all round the defect (Figure 5A) and pulled up. Few sutures were taken superiorly and inferiorly to reduce the defect without causing difficulty in ventilation. A prolene mesh, size 20 × 15 cm PRO-VISC 3D™, dual side mesh with polyurethane visceral and polyester parietal lining (LOTUS™, Lotus Surgicals Pvt Ltd, Mumbai, India) was sutured across the defect to the muscle edges with interrupted prolene sutures after trimming it to approximately 15 cm vertically and 11 cm from side to side, maximum width (Figure 5B). Another 5 cm or so of skin was trimmed from either side and closed in the midline (Figure 6C).

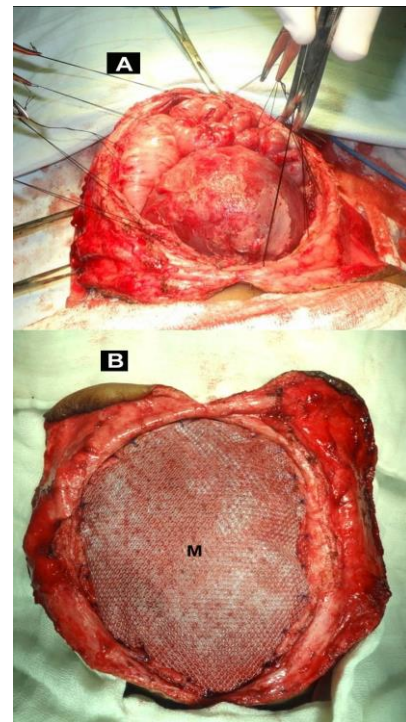


Figure 5: A) Re-exploration shows liver staying comfortably within the abdominal domain. Sutures taken all round the muscular margins for placement of mesh. B) Abdominal defect with M-mesh in place.



Figure 6: A comparative study of A) preoperative appearance, B) result after 1st surgery and C) result after 2nd surgery.

DISCUSSION

Exomphalos major has as many ways of management as the number of surgeons treating it. No single method is suitable for all patients. No single case is identical to another. There are variations in the size of the defect classifying it as major or minor. There are variations in the shape of the sac which may be conical with a wide base and small top, or globular with a large sac hanging on a relatively small diameter base with a variably small abdominal cavity. The latter is probably closest to the presentation of our patient. Then there are variations in the content of the hernia sac; the extent of gut, presence or absence of solid organs, mainly liver. There are variations in the size of the abdominal cavity and what it can accommodate. There are variations in the associated anomalies, whether syndromic or non-syndromic. There is also the issue of membrane coverage, intact or ruptured. Add to this the major evil, 'lack of awareness' which leads to delayed presentation as seen in our case and lastly, the experience of the operating surgeon. With so many variables there is not likely to be a single procedure which can satisfactorily deal with the problem. By and large, if the exomphalos cannot be repaired primarily then alternate methods have to be thought of.

If primary repair is not possible then the general consensus is to speed up the epithelisation process of the membrane covering the exomphalos.⁴ Many chemicals are known to do so and have been used, for example, mercurochrome, povidone iodine,^{5,6} alcohol, silver sulphadiazine,⁷ silver nitrate, gentian violet,⁸ aqueous eosin⁹ and Manuka honey.¹⁰ Silastic sheet application has also been done to encourage granulation underneath it.¹¹ When epithelisation of the sac has been achieved a ventral hernia is created which is closed in a second stage surgery.

Primary closure has also been done by using the 'silo' technique¹²⁻¹⁴ where gradual tightening of the silo bag increases the intra-abdominal space and primary closure can be done in weeks. It has also been attempted with 'vacuum assistance'.¹⁵ This is possible up to a point, where sufficient abdominal wall is present to close the defect without causing an abdominal compartment syndrome, or, if caused, appropriate intensive care management is available. Staged reduction is also done with the use of tissue expanders¹⁶ and air injection.³ Definitive closure with an absorbable mesh⁶ or alloderm graft² over the fascial defect is reported as a primary repair, provided it can be covered with skin. Traction compression closure using the exomphalos sac itself like a silo is also reported.¹⁷ Double breasting of the sheath has also been used with some success.⁵ A technique comprising of all the above techniques described for primary closure done together as a single procedure is also reported.¹

All these procedures which result in definitive closure of the exomphalos major require either prolonged admission with gradual expansion of the abdominal cavity or compromised closure of the abdomen with respiratory and vascular support. They may also require staged surgery with multiple admissions and some with a prolonged stay, or the use of expensive prosthesis. All the above procedures due to their multiplicity, prolonged stay, type of support required, use of prostheses and occurrence of complications can be quite expensive and have significant morbidity and mortality.

Our patient was a 22 month girl with a giant exomphalos (Figure 1). We do not classify it by the dimensions of the defect as the child was almost 2 years old and the dimensions hold good for neonates only. We classify it by its contents and its overall size. The exomphalos contained the entire liver, spleen, pancreas, stomach, duodenum, large part of small intestine as well as large part of colon. The defect measured 12 cm vertically and 10 cm from side to side and all dimensions of the liver were larger than the defect. The sac had a pedunculated appearance and was larger in body than base. The child surprisingly, was not hampered by it. She was as active as any other toddler her age. She ran around, holding the sac in front of her with both hands. She was in a habit of sitting cross legged with the sac in her lap and she slept in lateral position with the sac lying in front of her. It is

quite a miracle that the hepatic veins did not get avulsed during this period.¹¹

We planned a staged repair, anticipating at the least two stages or more before a mesh could be placed. In the first stage we widened the defect and extended it from xiphisternum to pubic symphysis by splitting the sheath and peritoneum in the mid line keeping the recti and abdominal muscles intact. The skin, connective tissue and peritoneum of the sac were kept intact. The incision which was extended above and below the defect had the skin dissected off the rectus sheath and abdominal muscle sheath so that only skin closure was done in the midline. The sac-covering which was now in the central part of the incision was not separated from its attachment with the muscle and was trimmed comfortably in the midline so as to maintain adequate traction on the muscles laterally without compromising the respiration and venous drainage of the abdomen upon closure in the midline. The superior extension of the wound also helped in avoiding kinking of the hepatic veins which were subcutaneous earlier and compressed behind the liver after the first stage repair. The post-operative appearance was of a ventral hernia in the protruding region only but was actually a complete ventral hernia. Feeding was started on the 4th post-operative day and the child was discharged on the 7th post-operative day. Ventilation and cardiovascular supports were not required.

The binders were in view of the shape and hanging nature of the liver, so as to provide support and gentle compression at the same time. The actual peripheral traction and generalised compression were provided by the method of dissection and wound closure itself. The appearance after 3 months was the desired result which we were looking for and the first encircling binder was discarded. The second binder was continued as a support and protection till the parents brought the child for the second stage of the procedure.

In the second stage surgery some adhesions were found which had to be separated. The adhesions were dense probably because we operated after a 6 months gap. An earlier surgery may have avoided the operative difficulty.

The muscle margin with sheath was defined and stay-sutures taken all around the defect. All the stays were held up and pulled towards the centre. This manoeuvre helped us in assessing the size of the defect and also in deciding how much could be repaired directly. Direct sheath and muscle repair was done with prolene sutures at the two ends of the defect and the remaining defect was closed with a PRO-VISC 3DTM dual mesh, as described, tailored and sutured to the sheath and muscle with interrupted prolene sutures. The skin was again reduced and closed in the mid line. The final appearance is shown in Figure 6C. The patient did not require any specialized care or support and went home on the 7th post-op day.

Our procedure, though staged, required only 1 week of hospitalization during each surgery. Specialized care, ventilator support, cardiovascular support and prostheses were not required. One prolene mesh was required. Post-operatively parenteral nutrition was not required as the child was kept nil orally for only 4 days and started feeding thereafter. All these procedures helped in cutting the cost of treatment. They also helped in avoiding ICU related complications and morbidity. This technique, which has been derived by us, has not been described before in literature. We recommend that it be called 'the Kaul Bedi technique' of omphalocele repair, wherein we have essentially 'widened the base to reduce the pressure'. We confidently recommend our procedure for the repair of complex cases of giant omphaloceles.

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