

Case Report

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

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ABSTRACT

In 2014 we published a procedure, the Kaul-Bedi technique, by which we used the epithelized covering of a giant exomphalos major, or hepato-omphalocele, to reduce the herniated organs and replace them into the abdominal cavity in stages. We published our technique when we were able to cover the defect with a dual PROVISC prolene mesh. We anticipated a long period of time before we would be able to bring about a primary closure of the abdominal wall defect. Contrary to our expectations, we were able to achieve a primary, complete, muscular closure of the defect within the next three years with removal of the mesh. We are presenting the stages of repair, discussing the complications encountered and the result achieved. We also discuss the advantages and the drawbacks of this procedure.

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

INTRODUCTION

Exomphalos is a congenital abdominal wall defect which varies in severity and its incidence is 1 in 10000 births in the west and about 0.77 in 10000 births in Europe. If the defect is up to 5 cm in diameter it is considered an exomphalos minor, whereas a defect more than 5 cm in diameter is exomphalos major. The incidence of exomphalos major is about 1 in 4000 to 5000 births and it is diagnosed antenatally by foetal ultrasonography.¹

The patient we presented in our previous publication was a 22-month old girl with a giant omphalocele having a defect with the vertical dimension of 12 cm and transverse dimension of 10 cm. The contents of this giant omphalocele were the entire liver, spleen, pancreas, part of stomach and duodenum, most of the small bowel, ascending, transverse and part of the descending colon. The abdominal contents consisted of the adrenals,

urogenital system and the proximal and distal-most parts of the gastro-intestinal tract. The abdominal cavity was extremely small with caving in of the lower rib cage. The liver was globose and suspended on its blood supply.¹

This child underwent staged repair, the first of which was done by the Kaul-Bedi technique, as described in our previous publication.¹ The second stage surgery was done after 8 months to further reduce the contents into the expanding abdominal cavity. Contrary to our expectations, we were able to reduce the contents completely and close the defect with a PROVISC 3D™ dual sided Prolene mesh (LOTUS™, Lotus Surgical Pvt Ltd, Mumbai, India).¹

We then planned to reduce the size of the mesh after a few years, gradually, in stages as the child would grow, to achieve a primary, muscular closure of the abdominal wall defect.

CASE REPORT

The patient was a 2 ½ year old girl who, at the age of 22 months had undergone the first surgery for a multi staged repair of a giant hepato-omphalocele. The procedure was described in a previous publication, as the Kaul-Bedi technique.¹ After 8 months she was able to undergo the second stage operation, wherein the defect could now be covered by a dual prolene mesh.¹ The organs had been reduced into the abdominal cavity. The plan now was to reduce the defect gradually by reducing the size of the mesh till such time as when the muscles along the margins of the defect could be brought together in the midline without the help of a mesh.

The first attempt at reducing the size of the mesh was taken after one year when the child was 3 ½ years old. Those of you who have used a mesh to repair hernias would know how a mesh adheres to its adjoining tissue layers by stimulating the laying of fibrous tissue. We had used a 'dual' sided mesh which had polyurethane on the visceral side and polyester on the parietal side. We anticipated that it would be very difficult to lift the epithelized exomphalos flap which was covering the mesh, entirely, remove the mesh and replace it with a smaller one. We also felt that the new mesh may not be significantly small enough to justify the procedure with its accompanying trauma and blood loss in this small child. Keeping this in mind and after multiple discussions with the manufacturers we decided to do another new procedure, which the manufacturers assured us, had not been attempted before. We could not find any literature similar to what we were intending to do. We planned a double breasting of the mesh after cutting it in its entire length in the mid line.

As anticipated, we found dense adherence of the flap to the mesh. What we did not anticipate, were the dense adhesions of the liver and small bowel to the visceral side of the mesh. This required painstaking dissection and led to one or two small serosal tears in the small bowel which were repaired. We were able to reduce the mesh from side to side by about 5-6 cm. We excised the excess tissue flap and closed in the midline. The child was accepting feeds after 24 hours and was discharged after 5 days.



Figure 1: The mesh has been dissected and cut in the midline.



Figure 2: Double breasted repair of the mesh in the midline.

At the first follow up, after a week, there was a serous discharge from the main wound in the epigastric region. We sent cultures and started a course of antibiotics. The child remained well and did not show any systemic signs of infection. The cultures did not grow anything but the discharge continued occasionally scabbing over, discharging altered blood, causing skin excoriation and not responding to dressings or curettage. We finally decided to remove the mesh and replace with a smaller one. The danger of the second mesh getting infected was there so at the time of surgery, after painting and draping, we placed Betadine soaked gauze over the discharging sinus and sutured it to the skin with about 2 cm to spare all around the discharging site.

We then excised the scar and the covered area together. The child was 4 ½ years old now and we used a Prolene PROVISC 3D™, 15 cm by 7.5 cm, dual sided mesh (LOTUS™, Lotus Surgicals Pvt Ltd, Mumbai, India). It was reduced to 10 cm vertically and 5 cm from side to side. The mesh that was removed was sent for culture and grew *Klebsiella*, sensitive to ceftriaxone and amikacin (amongst others), which we used for 10 days. The child remained well.

For the next year the child came regularly in follow up and was progressing well. At this point we decided to remove the mesh and replace it with a smaller one. During surgery we faced, as before, dense adherence to both sides of the mesh. After careful dissection we removed the mesh and discovered that we were now able to bring the muscular edges of the defect together in the midline without causing any signs of respiratory distress or abdominal compartment syndrome. She was able to feed after 24 hours and went home after 5 days.

She is now about 3 years in follow up and doing well. However, about 6 months after the last surgery she came in with sudden onset of vomiting and knowing she had malrotation, we immediately admitted and investigated her. She turned out to have gastroenteritis which soon followed with diarrhoea. Abdominal X-rays did not show any signs of intestinal obstruction and she settled the next day and went home.



Figure 3: Final appearance of the anterior abdominal wall showing midline scar, over a relatively normal looking abdomen with absence of umbilicus.

DISCUSSION

Our patient who initially presented at the age of 1 year and 10 months with a giant hepato-omphalocele underwent five major staged operations which resulted in a primary and complete repair of the abdominal wall at age 5 years and 6 months. We have essentially used a new staged procedure [1] for a giant omphalocele repair using minimal prosthetic support, avoiding mechanical ventilation and obtaining complete abdominal wall closure. There was no threat of abdominal compartment syndrome (ACS) nor prolonged stay in hospital. Many compounds have been used over the anterior abdominal wall defect to cause epithelization over the membrane to improve its stability and strength, creating a ventral hernia, which is then later repaired.²⁻⁵ As our patient presented at 1 year and 10 months of age with epithelization over the defect, we planned a staged closure of a ‘giant ventral hernia’. Many authors have published various techniques of delayed primary as well as staged secondary closure, each technique having its pros and cons. We presented our technique in a previous paper to which this paper is a sequel.¹ The proponents of delayed primary closure have to deal with prolonged hospital stay, with mechanical ventilation.^{6,7} The main problems faced are impending ACS and its sequelae, catheter and ventilator related infections and subsequent morbidity as well as mortalities. Added to this is the expense incurred.

Staged repair of giant omphalocele involves increasing the space in the abdominal cavity and completing abdominal wall closure. The proponents of delayed primary abdominal wall closure depend on the fact that delaying the repair gradually increases the intra-abdominal domain, which makes a delayed primary repair possible.^{6,7} Others have used techniques like progressive pneumoperitoneum, intra-abdominal tissue expanders used for varying periods of time, followed by secondary abdominal wall repair which may or may not be staged.⁸⁻¹⁰ Pneumoperitoneum is difficult to maintain and requires prolonged monitoring. It creates discomfort and may not be well tolerated. It can have life threatening complications of bowel injury or air embolization.

Tissue expanders have reported good results but have also reported complications like rupture of the prosthesis leading to intestinal obstruction, poor tolerance of prosthesis and ACS like situation. Intra-abdominal infections are also reported, significantly increasing the morbidity and occasionally causing mortality. They may also require repeated changes and are expensive.

Without increasing the intra-abdominal domain, the procedures for abdominal wall repair are the component separation technique (CST) described by Ramirez in 1990.¹¹⁻¹³ It has reported complications like seroma and haematoma formation, wound infections, skin necrosis and recurrence of hernia. The patient may require ventilation for a variable period and is exposed to its sequelae. Another procedure is the Lazaro da Silva’s technique of abdominal wall repair which requires the creation of rectus sheath flaps and skin flaps.¹⁴ It is a fairly extensive procedure which would require a variable period of post-operative ventilation.

We performed a technique of staged repair after epithelization of the sac wherein we utilized the epithelized sac itself as a flap to tighten the hernia and push the contents towards the abdominal domain; while widening the defect to reduce the pressure within the abdomen. To achieve this, we did not separate the layers within the sac itself nor did we separate it from the Recti muscles around the rim of the defect. We were able to achieve pressure on the contents of the sac, pushing them towards the abdominal cavity. Secondly, by widening the defect, that is incising the muscles and sheath in the midline above till the xiphisternum, and below till the pubic symphysis, and closing the skin and subcutaneous tissues only, in this region, we reduced intra-abdominal pressure, protecting the child from ACS and its complications. The contents of the sac were now acting as ‘tissue expanders’, helping in increasing the abdominal domain. Also, by not separating the skin and subcutaneous tissues in the flaps, the pull on the muscles of the abdominal wall helped in bringing the recti together in the midline.

After double breasting of the mesh was done, there was a wound discharge which went on to become a seroma with a discharging sinus. This was a setback in our staged repair specially when it grew *Klebsiella* for which antibiotics were required. As the child had an Exomphalos Major with almost the entire gut lying in the epithelised sac, she also had malrotation of the gut. This leaves open the possibility of a volvulus at a later time. She has the chances of an adhesion obstruction as much as any patient after a laparotomy. At each successive laparotomy we noticed adherence to the mesh as well as to the abdominal wall. The gut was also stuck to its own loops. None of this was causing obstruction and, in our opinion, would actually help in avoiding a volvulus. We did not attempt to separate these loops during the surgeries as they were not dilated and we did not want to cause iatrogenic adhesions which might lead to intestinal obstruction. Another problem may

arise when she grows up and decides to have a child. How would pregnancy affect the surgical scar, is a pressing question but more pressing than that is the question of whether, when the time comes, she should be allowed to go into normal labour or undergo planned caesarean section. Though the parents have been informed of the above possibilities, we believe that the problems will best be dealt with as and when they arise. The even more pressing and immediate problem, which we feel will have to be dealt with much sooner is the creation of an umbilicus. We feel that we should address that problem as soon as the child is old enough to realize the fact that she does not have an umbilicus and asks for the procedure. Though superfluous to the underlying condition we strongly feel that such a procedure is mandatory if the child is having psychological effects due to the absence of an umbilicus.

CONCLUSION

Due to the Kaul-Bedi technique, which we described in our earlier publication, we were able to correct this hepato-omphalocele within approximately three and a half years and bring the abdominal muscles together in the midline. We were able to completely avoid mechanical ventilation, ACS and their associated complications. Though we used a prolene dual mesh in the second procedure, many authors have left the patient with a mesh in place. We, on the other hand, were able to get the abdominal wall muscles together in the midline without making flaps or doing elaborate plastic procedures on the abdominal wall. The hospital stay of our patient was never more than 7 days and feeding was started by 2nd or 3rd post-operated day. The procedures were simple and did not incur huge costs. Mesh infection which occurred after double breasting the mesh was a complication which, having occurred should have been treated with mesh removal. We also feel that the procedures probably could have been done at shorter intervals. However, we feel confident in recommending this procedure for patients with giant omphaloceles.

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