

The Rare Congenital Anomaly Omphalocele: A Review of 25 years of Management

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Abstract

Objective: The objective of this study was to:

1. Evaluate the various techniques chosen for management of small versus giant omphalocele & the complications that arose for each
2. Compare the above outcomes with current published literature

Study Design: Cases of omphalocele that underwent evaluation between January 1991 and December 2016 at Health Sciences Centre (HSC) Children's Hospital were identified. Inclusion criteria was restricted to live births with a plan for postnatal repair and patients who had their initial treatment for the defect at HSC hospital.

Results: Among the 28 cases, 14 patients were categorized as having a small omphalocele. Of these 14 patients, 11 underwent primary fascial closure, 1 had a staged closure, and 2 were treated with a delayed closure technique. No complications occurred among the small omphalocele patients. Fourteen patients were categorized as having a giant omphalocele. Of these 14 patients, 2 underwent primary fascial closure, 8 had a staged closure, and 4 were treated with a delayed closure technique. Of the 2 patients who underwent primary fascial closure, 1 developed an infection at the surgical site and respiratory failure. Three of the 8 patients treated with staged closure developed various combinations of inferior vena cava thrombosis, metabolic acidosis, acute renal failure, respiratory failure and infection at the surgical site. Delayed closure did not result in any complications.

Conclusion: Primary fascial closure is the preferred technique for small omphalocele repair at HSC Children's Hospital and in current published literature. Primary fascial closure is an excellent technique for small omphalocele as it requires only one procedure and few complications arise. Staged closure is the preferred technique for giant omphalocele reduction at HSC Children's Hospital and in current published literature. This technique is often chosen as it produces less complications than primary fascial closure in giant omphalocele. Delayed closure was less frequently employed for giant omphalocele despite having the lowest complication rates of all three techniques.

Key Words: Omphalocele, small, giant, primary, staged, delayed, silo, mesh, skin flap

Limitations: Omphalocele documentation - description versus measurement. Giant omphalocele definition. Unknown eviscerated contents. Small sample size.

Purpose: The purpose of this study was to relay to physician assistants current information about the treatment of omphalocele patients treated in Manitoba, and to provide a comparison to treatment in current literature. The results of this study will help physician assistants educate their patients about what to expect in regards to epidemiology, embryology, diagnostics, co-morbidities, post-natal management, treatments and potential complications.

Introduction

EPIDEMIOLOGY AND EMBRYOLOGY

Omphalocele is a rare congenital anomaly that occurs approximately once in every 5,000 births (1,2). Omphalocele is defined by herniation of abdominal contents into the base of the umbilical cord (1,2). The midline defect results from the failed progression of the anterolateral abdominal walls during weeks 6 to 10 of embryonic development leaving the herniated viscera covered by a peritoneal sac (2,3). The embryo begins as a flat oval disk composed of three layers: the dorsal ectoderm, responsible for formation of the central nervous system, skin and sensory organs; the ventral endoderm, which results in the gut and its appendages; and the intermediary mesoderm, which evolves into the muscular skeletal system and connective tissue (3–5). At week 4 of gestation, the edges of the embryonic disc begin to fold over to form the ventral wall (3,4). By week 6 of gestation, the gastrointestinal system begins to develop so rapidly that the embryonic abdomen cannot accommodate the fast growing contents (4,5). As a consequence, the majority of the midgut is temporarily herniated outside of the abdomen such that the intestine loops into the umbilical cord (2,3). Failure of the herniated contents to return to the abdominal cavity by week 10 results in an omphalocele (3,4). Although there are many theories proposed, the reason for this failure is still unknown (3).

COMPOSITION AND CLASSIFICATION

The omphalocele sac is composed of dual membranes, one from the amnion and one the peritoneum, both of which sandwich an inner fluid called Wharton's jelly (5). Omphalocele defects are variable in the diameter of the defect and in the viscera content found within the sac: small bowel, colon, liver, spleen or stomach (6).

Omphalocele often is categorized as 'small' versus 'giant', in addition to, whether the sac is ruptured (7). The definitions of small and giant omphalocele vary greatly in the literature (7–9). Some make the distinction based on the diameter of the defect, while others base it on whether the liver is eviscerated into the sac (7,9). Most recent literature has defined giant as including both characteristics: a defect greater than or equal to 5 cm and containing liver (8,10,11). The position of the liver (rather than any other organ) is important because complications can result from overzealous reduction of the liver into the underdeveloped abdominal cavity causing occlusion of the suprahepatic inferior vena cava (9,12). These complications include: hepatic vein thrombosis, liver infarction, increased venous pressure, hepatic necrosis, and liver failure (9,12).

DIAGNOSTICS

Omphalocele may be suspected in the fetus of a pregnant mother who has an elevated serum alpha-fetoprotein (2,3). Diagnosis can be made prenatally with ultrasound (2,3). Amniocentesis is offered to mothers to identify chromosomal abnormalities because omphalocele is frequently associated with other congenital and chromosomal anomalies (2,3).

CO-MORBIDITIES

Omphalocele is often associated with chromosomal abnormalities, syndromes and other congenital anomalies (13). Pulmonary associations include: respiratory hypoplasia, pulmonary hypertension and diaphragmatic defects (7,10,13). Cardiac defects include: patent ductus arteriosus, patent foramen ovale, atrial and/or ventricular defects (7,10). Gastrointestinal defects include malrotation, intestinal atresias and duplications (7,10,13). Renal and neural tube defects also occur (7,10,13). Additionally, some of the common genetic anomalies and syndromes associated with omphalocele include: Pentalogy of Cantrell, Beckwith-Wiedemann syndrome, Pallister-Killian syndrome, Ritscher-Schinzel syndrome, CHARGE, Trisomy 13 and Trisomy 18 (7,13).

POSTNATAL MANAGEMENT

The mode of delivering a fetus with known omphalocele is controversial. While caesarean section may be preferred by some physicians, the literature has not proven that caesarean improves outcomes for infants (10,14).

Immediate postnatal management of omphalocele is important to improve outcomes and mortality rates (10,14). Goals of management included temperature control, hydration, cardiorespiratory stability, vascular access and care of the defect (10,14). In the case of respiratory depression from an associated anomaly, intubation for airway stability may be required (15). To prevent intestinal dilation and to decompress the abdomen, insertion of an orogastric or nasogastric tube is mandatory (10,15). To prevent hypovolemic shock due to the evaporative loss, establishment of peripheral intravenous access is required (15). The omphalocele must be covered with saline-soaked gauze and plastic wrap to prevent fluid and heat loss and to reduce infection risk (10,14,15). Use of a heat lamp can also help to prevent hypothermia (15). Lastly, routine neonatal blood work is important to evaluate electrolytes and glucose (10,14). Vitamin K and antibiotic administration, especially in ruptured omphalocele, may reduce the risk of bleeding and infection (10,14). As cardiac co-morbidities are common, a fetal echocardiogram should be considered (10).

TREATMENT

Treatment options for omphalocele are typically based on the diameter of the abdominal wall defect and the presence of liver within the sac (10). The main treatment options are: (i) primary fascial closure, (ii) staged closure with silo, skin flap or mesh, and (iii) delayed closure (8).

PRIMARY FACIAL CLOSURE

Primary fascial closure involves resection of the peritoneal sac, reduction of the eviscerated organs into the abdominal cavity, and closure of the abdominal wall layers, the fascia and the skin (14). The fascial layer may be closed in anatomic layers or as a mass closure (16). The layered technique involves closure of consecutive abdominal wall layers individually (16). The mass closure involves closing all the layers of the abdominal wall as a single layer (16). Layered or mass closure is done as one procedure shortly after birth once the baby has been stabilized (10). Primary fascial closure is done as one procedure and typically renders better results with small omphalocele (8).

STAGED CLOSURE

Staged closure involves the gradual reduction of the eviscerated organs into the abdominal cavity (14). Gradual reduction reduces the chance of abdominal compartment syndrome which can result from overcrowding of the abdominal cavity (14). Abdominal compartment syndrome can cause ischemic bowel from compression of mesenteric vessels, renal failure from reduced perfusion, impaired pulmonary function, and circulatory compromise due to inferior vena cava compression (7,9,12,14).

The skin flap technique was first described in 1948 by Gross *et al.* The skin of the abdominal wall is undermined circumferentially away from the defect toward the anterior axillary lines (14). The skin edges are then pulled together medially for closure with minimal tension (14). This technique leaves the patient with a ventral hernia, thus subsequent surgery to close the hernia is required (10). This hernia closure may be performed 6 – 24 months after the skin closure (4).

The silo approach was first described in 1967 by Schuster *et al.* This technique involves suturing a sac-like non-adhesive material to the edges of the fascial layer and covering the evisceration within the sac creating a chimney (14). The silo pouch is covered in sterile gauze and is suspended from the top of the incubator to keep the intestine in anatomic alignment (10). The eviscerated contents are then gradually reduced over several days to weeks by gravity or manually (14). When viscera are sufficiently reduced, fascial, mesh or skin flap closure can be attempted (10).

The last staged technique uses mesh. Synthetic or biologic mesh can be used to contain the evisceration within the abdominal region and aids in restoring abdominal wall stability (6,17). Synthetic mesh materials which are permanent in nature are typically composed of polypropylene,

polyester or polytetrafluoroethylene materials (18). Synthetic mesh materials containing Dexon or Vicryl are designed to completely degrade over time (18). Biological mesh materials are created from biological decellularized human or bovine skin, used to create a collagen framework to support collagen deposition from the mesh recipient (18).

Mesh can be used in a versatile manner with respect to ventral defect repair. In some situations a patient may undergo a staged closure where permanent synthetic mesh material is used to secure the fascial edges under tension to act as a bridge (19). Serial excision of the central aspect of the mesh material to be completed every few days (19). Eventually, the final procedure would result in mesh excision followed by primary fascial closure alone, or primary fascial closure reinforced with a biological mesh (19). Alternatively, insertion of mesh may be followed by coverage with skin flaps or topical therapy to promote epithelialization (6,8,11,17). In some cases the mesh may be removed when the child is bigger, where the fascial edges are pulled closed. (6,17). Mesh can also be used as an independent technique for omphalocele closure (8,11). A permanent mesh material may be used to reduce the tension that would occur if primary fascial closure were attempted (11,20). In this case, no subsequent surgeries are planned to remove the mesh (8,11).

DELAYED CLOSURE

Delayed closure involves the use of topical agents to promote granulation and epithelization over the membranous sac (10). This technique is also known as the “paint and wait” method (21). A bacteriostatic cream or ointment is applied to the sac daily and then covered with a dressing (5,10,16). Epithelization can take several months to a year to complete (1). Eventually, a closure procedure is performed to close the remaining ventral herniation (7). Agents currently used include silver sulfadiazine and betadine (7,10). Silver sulfadiazine is a topical antibiotic (5). It is a synthetic sulfonamide and can be used to prevent and treat infections in patients (5). Risks of silver sulfadiazine use include allergic reaction and hemolysis in patients with G6PD deficiency (5,8). Silver sulfadiazine should be used with caution in individuals with hepatic or renal impairment (5,8). Betadine is an iodine based antiseptic used for its antimicrobial properties (5). Use of betadine comes with risk of iodine-induced thyroid dysfunction (5,8).

Study Design

This was a retrospective chart review of patients with omphalocele that underwent treatment at HSC Children’s Hospital between January 1991 and December 2016. Forty infants were identified, 11 of whom died shortly after birth. These 11 patients were not considered for postnatal repair and not included in this review. One patient was treated for omphalocele at a different hospital but subsequently assessed at HSC; this patient was also excluded from this study. Of the remaining 28 patients, cases were separated into two categories: small versus giant. Fourteen patients met the published criteria defining giant omphalocele as a defect diameter greater

than or equal to 5 cm and containing liver. Fourteen patients remained in the small omphalocele category. Only 1 patient with ruptured omphalocele met the inclusion criteria. All initial surgical repairs, primary fascial closures or staged closures, were done within the first 7 days of life.

Results

SMALL OMPHALOCELE

Of the 14 small omphalocele cases, 78.6% (n = 11) underwent primary fascial closure, 7.1% (n = 1) underwent staged closure via the skin flap technique, and 14.3% (n = 2) were allowed to epithelialize (Table 1). One patient underwent epithelialization by default; the omphalocele was so small that the little sac granulated over before the planned primary closure was to be completed. No complications occurred in the 14 small omphalocele repairs (Table 1)

GIANT OMPHALOCELE

Of the 14 giant omphalocele cases, 14.3% (n = 2) underwent primary fascial closure. One of these patients (50%) had a surgical site infection and respiratory failure due to abdominal compartment syndrome (Table 1). Eight patients (57.1%) underwent staged closure (Table 1). Of the 8 staged closure cases, 62.5% (n = 5) were managed with mesh closure (Table 2). Two of these 5 patients (40%) had complications (Table 2). One developed an inferior vena cava thrombosis, metabolic acidosis, and acute renal failure. The second case developed respiratory failure due to abdominal compartment syndrome. Two patients (25%) from the staged closure cases were reduced with a silo; none of these cases had a complication (Table 2). One patient (12.5%) from the staged closure cases was reduced with the skin flap technique (Table 2). This patient developed a surgical site infection (Table 2). Thus, of the 8 patients that underwent staged closure, 37.5% (n = 3) had a complication (Table 1). None of the 4 patients (28.6%) that had a delayed closure had a complication (Table 1).

RUPTURED

Only 3 of the 40 cases had ruptured omphalocele. Two of which were among the mortalities. The 1 case of ruptured omphalocele that was treated was giant. This patient underwent a staged closure with use of a silo. No complications resulted with this treatment.

Discussion

This study provides an overview of the different treatment modalities for patients both born with omphalocele and treated at the HSC Children's Hospital during the last 25 years. The main goal of this study was to investigate the treatment options used for omphalocele at the HSC Children's Hospital. Specifically, to identify the preferred methods for treatment based on small versus giant omphalocele, to review the complications that arose from each situation, and lastly, to compare the overall results with the current published literature.

MORBIDITY AND MORTALITY

Of the 40 cases of omphalocele reviewed over the last 25 years, 11 (27.5%) patients did not survive to have a planned procedure. Of these 11 patients, the size of defect varied from very small (2 cm) to giant (12 cm). Various organs were present within the sacs. Some sacs had small bowel or liver only. Other non-survivors had varying combinations of liver, small bowel, colon, spleen, pancreas and kidney within the sacs. Causes of death also varied, including: respiratory failure, cardiac defects and failure to thrive. Historically, mortality in omphalocele patients was believed to be due to five factors: promptness of repair, defect size and content, associated congenital anomalies, prematurity, and sac rupture (15). However, current literature reports that the greatest influence on mortality in omphalocele patients is the presence co-morbidities (10). As morbidity and mortality were not part of the objectives included in this document, further data analysis into this area was not completed.

SMALL OMPHALOCELE

Small omphalocele was present in 50% of the 28 patients who met the inclusion criteria for this study. The majority of eviscerated contents within the small omphaloceles was small bowel only, but a few patients had liver only. All of the small defects were less than 5 cm or were described as small by the surgeon. The most frequently used technique for closure of the small omphalocele at HSC Children's Hospital was primary fascial closure. Delayed closure and staged closure were rarely used for small omphalocele repair. No complications were seen with any of the small omphalocele repairs at HSC.

Based on current literature, primary closure is the preferred technique for small defects (10,14). Small omphalocele is a topic less frequently discussed in the current literature compared to giant omphalocele. This is likely due to the fact that small omphalocele closure tends to be more straightforward and leads to fewer complications. A paper published in 2004 by Heider *et al.* which investigated the management of omphalocele closures over a 23 year period, identified several cases of small omphalocele (22). This article reported that the majority of small omphalocele were repaired via primary closure (88% of the cases), with a smaller portion repaired using delayed closure (12% of the cases) (22). When these values are compared to those found

over the 25 year study at HSC Children's Hospital: primary fascial closure was used in 78.6% of the cases at HSC and 88% in the literature; delayed closure was used in 14.3% of the cases at HSC and 12% in the current literature; finally, staged closure was recorded for 8.1% of the cases of HSC and none for current literature. Overall, it appears as though the approach to small omphalocele repair at HSC is similar to that described in the current literature.

GIANT OMPHALOCELE

Giant omphalocele was present in 50% of the 28 patients who met the inclusion criteria for this study. The contents within the giant omphaloceles varied more than within the small omphaloceles. All giant omphalocele contained liver and had a defect of at least 5 cm, or were described as giant with herniated liver. Similar to the small omphalocele, all three types of repairs were used to manage giant omphalocele: primary fascial closure, staged, and delayed. The preferred method of closure of giant omphalocele at HSC was staged closure. Mesh was the most common technique, then silo, and then skin flap. Of the staged procedures, skin flap had the highest rate of complication, followed by mesh closure, silos had none. Delayed closure was used less often than staged closure, it however resulted in no complications. Primary fascial closure, used the least for giant omphalocele repair was found to have the highest rate of complications.

Based on current literature, giant omphalocele reduction tends to be much more of a challenge than small omphalocele. This challenge lies in the various complications that can result when the evisceration is reduced too quickly. When considering which closure technique is appropriate, several variables need to be considered. A systematic review published in 2010 investigated the management of giant omphalocele closures over a 42 year period; 21 papers were identified (11). Among the identified papers, the majority of giant omphalocele were repaired via staged closure (52.4% of the cases), followed by delayed closure (28.6% of the cases), with the least smallest portion repaired using primary closure (19% of the cases) (11). When these values are compared to those found over the 25 year study at HSC Children's Hospital, the results are as follows: primary fascial closure was used 14.3% of the time at HSC and 19% in the literature; staged closure was used in 57.1% of the cases at HSC and 52.4% in the literature; finally, delayed closure was recorded for 28.6% of the cases at HSC and 28.6% in the literature. The HSC experience with giant omphalocele is remarkably similar to that described in the current literature.

Complications were noted in each study cited in the systematic review published in 2010, however, the rates of complications within each article were not reported (7). Overall, the authors who chose primary fascial closure for giant omphalocele found that over half of the cases resulted in herniation and that a smaller portion of the cases resulted in mortality due to complications (11). This differs from the complications seen at HSC for primary closure where surgical site infection and respiratory failure were primarily seen. Similar to HSC, the authors who chose staged reduction as a surgical technique for giant omphalocele noted local skin infection and prolonged mechanical ventilation due to respiratory failure as a complication (11). Though some HSC and the literature shared similar complications for staged closure, HSC patients experienced inferior vena cava thrombosis, metabolic acidosis, and acute renal failure. The patients described in the

literature experienced muscle paralysis, skin necrosis, sutures pulling away from the skin/amnion junction, sac tearing, sepsis and hernia (11). Lastly, the article identified that delayed closure resulted in such complications as ischemic small bowel, infection and sepsis, and partial skin necrosis, which differs entirely from the experience at HSC where no complications were seen with delayed closure (11).

PRIMARY FASCIAL CLOSURE

Primary fascial closure appears to be a technique that is typically limited to smaller defects. Current literature describes this technique as being beneficial because both the reduction and ventral defect closure are completed as a single procedure (7). Additionally, this procedure has been shown to lead to less risk of local infection and increased chance of early enteral feeding (8). Problems arise with this technique when attempting to close an omphalocele that is too large in size (7,14). Attempting closure when the abdominal cavity is unable to accommodate the volume of evisceration should lead one to consider a prosthetic mesh; if one fails to recognize the need for a staged closure then primary closure results in complications (7,14). The addition of a prosthetic mesh to the procedure may result in the need for subsequent surgery to remove the foreign mesh material negating the benefit of primary closure being a single procedure (7). In addition, forced reduction of a large omphalocele leads to the increased risk of abdominal compartment syndrome (9,12).

The findings from the 25 year analysis of the HSC data are analogous to those found in the literature. Primary closure of small omphalocele yielded fewer complications than primary closure of giant omphalocele repair.

STAGED CLOSURE: SKIN FLAP

The skin flap technique as a staged closure is useful for larger defects (14). Current literature describes this technique as being beneficial since abdominal compartment syndrome is uncommon. The skin flap technique allows for gentle, gradual reduction of the viscera (7). However, because this technique only closes skin, it leaves a ventral hernia which requires another planned procedure to close the fascia (8,14). Additionally, if a defect is too large there may not be enough skin to cover the herniate viscera; thus, additional material would be required to close the gap (7). Other disadvantages include such complications as local infection and skin necrosis (10).

The findings from the 25 year analysis of the HSC data is comparable to that found in the literature. Of the HSC patients, 2 patients underwent the skin flap procedure and one had a complication.

STAGED CLOSURE: MESH

The mesh closure, which allows for a closure and stabilizes the abdominal wall, is useful for larger defects and ruptured omphalocele (23). This method is advantageous for the same reasons as the skin flap since it reduces the risk of abdominal compartment syndrome as it allows for gradual reduction of eviscerated content into the abdominal cavity (7). Disadvantages of mesh closure technique are cost and the high rate of subsequent herniation (7,11). One paper reported that although this technique had a low infection rate, a number of patients required subsequent silo reductions because the mesh closure was inadequate (23).

The results from the 25 year study at HSC are not quite analogous to those seen in the current literature. Forty percent of HSC patients who underwent the mesh procedure had complications. Although current literature states this technique should prevent abdominal compartment syndrome, it occurred at a high rate at HSC.

STAGED CLOSURE: SILO

The silo approach is excellent for giant and ruptured omphalocele (14). Current literature reports that this technique is beneficial for the same reason as seen with skin flap and mesh closure; it minimizes the risk of abdominal compartment syndrome (7). In addition, the silo can help to protect ruptured omphalocele contents in a controlled manner (7,14). However, current literature reports some risks of this procedure, such as wound infection, loss of fascial margin, fistula formation, dehiscence, and sepsis (7,23). Additionally, this technique also requires multiple procedures (11,14,23)

Although complications are described in the literature, patients who underwent the silo procedure at HSC experienced no complications. At HSC, this technique appears to have prevented abdominal compartment syndrome and was used when treating the one ruptured omphalocele.

STAGED CLOSURE

Overall staged closure seems to be the preferred method for closure of giant omphalocele at HSC. However, it was also used for small omphalocele. This technique resulted in fewer complications for giant omphalocele repair versus using the primary fascial closure technique. Of the three types of staged closure repair, the preferred technique was mesh closure, then silo, and finally skin flap. Though mesh closure was the preferred method of the staged closure techniques, it yielded a higher complication rate than that seen with the silo method, a technique used less frequently but yielding less complication.

DELAYED CLOSURE

Delayed closure is a conservative technique that is often used for giant omphalocele and involves non-operative management with late closure (23). This technique is advantageous as it avoids surgery in a newborn, uses topical agents to help prevent infection, is economical, and may require only one surgery to close the ventral hernia (7,23). There are, however, disadvantages to delayed closure. Delayed closure through epithelization can take several months and the surgery to close the ventral defect may not take place for years (7,23). Additionally, anomalies of the gastrointestinal tract may not be identified as the abdominal cavity is not explored as it would with a more invasive technique (7,23).

The findings from the 25 year analysis of the HSC data is comparable to those found in the literature. This technique resulted in the fewest number of complications overall for giant omphalocele. Similarly, small omphalocele patients at HSC managed with the delayed technique had no complications.

OVERALL CONCLUSIONS

Primary fascial closure is the preferred technique for small omphalocele repair at HSC Children's Hospital and elsewhere. With small omphalocele, primary closure is a less complex procedure which results in few complications.

Based on the findings in the HSC cases and elsewhere, management of giant omphalocele remains a challenge. All closure techniques, primary, staged and delayed, are used for giant omphalocele repair. When deciding on which technique to use, the surgeon and family must compare both the pros and cons of each method. The delayed method is often a good choice as it has been found to be associated with few complications. However, this technique has the longest treatment plan. Primary closure, although it has the shortest treatment plan, has the highest complication rate. Primary closure may be a technique that should be reserved for small omphalocele. The staged method has proven effective at HSC and other centres. The staged technique has a medium duration treatment plan and has a moderate rate of complications. Among the types of staged techniques, the silo resulted in the fewest complications; the mesh and skin flap staged techniques resulted in the most. Essentially, when choosing a method for giant omphalocele closure, one needs to weigh the risks and benefits of a long treatment plan with a low complication rate with a short treatment plan with a high complication rate.

LIMITATIONS OF STUDY

Description versus measurement. There is no consensus among surgeons about how omphalocele should be documented. In some cases, a numerical measurement of the defect was documented. At other times, a descriptor, such as 'small' or 'giant' was reported. This inconsistency made it difficult to categorize cases and, as a result, may have affected the comparative analysis.

The definition of giant omphalocele. This definition varies throughout the literature. Although most recent literature tends to use some similar features to define giant (greater than or equal to 5 cm and liver involved) past literature is much more variable.

Eviscerated contents. One of the initial goals of this study was to compare eviscerated contents between omphaloceles to see how this affected outcome. However, not all cases listed the various contents found within the sac. Additionally, the current literature often only discusses the presence of liver within the sac.

The number of omphalocele patients born at HSC within the timeline chosen and who survived to be treated was only 28 cases. This is a small cohort which makes it difficult to compare data and have statistically significant results. Thus, a statistical analysis of the data could not be done for this study.

FUTURE PROSPECTS

This project, which reviewed the presentation, investigation, and management of patients with omphalocele treated at Children's Hospital Health Sciences Centre in the last 25 years, is part of a larger project currently in progress. The larger project, the Surgical Congenital Diseases Long-term Follow-up Study, reviews the presentation, investigation, and management of nine congenital anomalies managed with surgery. The eventual plan for this project will be to link this data with the Manitoba Centre for Health Policy database with the intention of identifying factors that affect long term socioeconomic outcomes in order to establish a long-term follow-up clinic in Winnipeg for children with these anomalies. The clinic will follow each patient in a standardized manner to identify complications early, to provide on-going patient and family support, and to improve long-term outcomes.

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Table 1: Comparative analysis of surgical technique and complications in small versus giant omphalocele cases. Total cases of evaluation n = 28.

	Primary Closure	Complications	Staged Closure	Complications	Delayed Closure	Complications
Small (n = 14)	78.6% (n = 11)	0	7.1% (n = 1)	0	14.3% (n = 2)	0
Giant (n = 14)	14.3% (n = 2)	50.0% (n = 1)	57.1% (n = 8)	37.5% (n = 3)	28.6% (n = 4)	0

Table 2: Comparative analysis of the three methods of staged repair and associated complications in giant omphalocele cases. Total cases with staged repair for giant omphalocele n = 8.

Silo	Mesh	Skin Flap
25.0% (n = 2)	62.5% (n = 5)	12.5% (n = 1)
0	40% (n = 2)	100% (n = 1)