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European Paediatric Surgeons' Association Consensus Statement on the Management of Giant Omphalocele

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Abstract

Keywords

- consensus statement
- giant omphalocele
- surgical management
- conservative treatment
- outcomes

Introduction Giant omphalocele management had not reached a consensus from the pediatric surgical perspective regarding conservative treatments, surgical approaches, and clinical outcomes. This topic was therefore selected for the 2023 Consensus Session of the European Paediatric Surgeons' Association (EUPSA).

Methods Literature review was conducted by seven EUPSA members, guided by a set of predefined areas relating to the management of giant omphalocele: (1) conservative management, (2) surgical management, and (3) outcomes. Members were assigned to specific topics, with discrepancies resolved through structured group discussion and further literature review. Consensus was reached through unanimous agreement among the contributing members. Each topic was presented with available evidence to congress participants. Comments from participants were accounted to formulate the final consensus statement.

Results Giant omphalocele is appropriately defined as viscerio-abdominal disproportion preventing primary closure. Regarding (1) *conservative management*, the “paint and wait” approach is recommended when anatomical constraints or high surgical risk preclude primary closure. Common painting agents include povidone-iodine and silver sulfadiazine, with Manuka honey gaining interest, though consensus on dosing, duration, and complications remains unclear. With regards to (2) *surgical management*, early closure favors biological meshes, while delayed closure appears most effective using native tissues. High mortality in delayed patch closure likely reflects a population with the most severe defects. Finally, (3) *outcomes* highlights key prognostic factors

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including chromosomal abnormalities, cardiac defects, and low birth weight, which may guide counseling, screening, and treatment. Limited data on ruptured omphalocele indicates towards increased mortality, necessitating prompt intervention. Complications following management are relatively rare and typically intervention-related, underscoring the need for long-term, multidisciplinary follow-up.

Conclusion A consensus statement on the management of giant omphalocele was developed based on current evidence and peer practice, though limitations relating to a scarcity of high-level evidence and significant heterogeneity across studies should be acknowledged. Despite these constraints, this consensus statement provides evidence-based guidance to support pediatric surgeons in informed decision-making for this pathology.

Introduction

Omphaloceles are congenital defects of the umbilical ring arising from arrested embryonic development of the lateral abdominal wall folds, though cephalic and caudal folding defects may also occur, contributing to increased morbidity.^{1–3} Subsequently, viscera situate outside of the abdominal cavity, enclosed by a three-layered membrane comprising external amnion, Wharton's jelly, and inner peritoneum. Extracorporeal situation of abdominal contents impairs expansion of the abdominal cavity during gestation.^{1,4}

Omphaloceles occur in approximately 1 to 3.8 per 10,000 live births, though true incidence is likely higher when accounting for elective abortions and fetal demise.^{5,6}

Omphaloceles can be classified according to the size, contents, and integrity of the sac, with defects ranging from approximately 2 to 15 cm.^{7,8} Giant omphalocele is associated with particularly high morbidity and mortality, with lethal outcomes reported in around one-fifth of neonates.⁸ However, there is significant variation in the criteria used to define giant omphalocele, which include defect size >5 cm, liver herniation, degree of abdominopelvic disproportion, and inability to achieve primary closure.^{3,5,8–10} This variability contributes to inconsistent incidence rates within the literature, ranging from 14 to 57% of cases.^{11–13}

Despite advances in neonatal care and surgical techniques, management of giant omphalocele remains controversial, with no consensus as to the optimal approach.

Two major strategies employed in the management of giant omphalocele are (1) nonoperative delayed closure and (2) staged surgical closure.^{3,14} Nonoperative delayed closure is achieved through application of topical medications onto the omphalocele membrane which promote the formation of eschar, followed by degranulation and epithelialization, allowing subsequent interval repair of the remaining ventral hernia.^{3,15} Surgical management aims to reduce the herniated viscera, close the fascia and skin, and reconstruct a stable abdominal wall while avoiding excessive intraabdominal pressure.¹⁴ Staged closure achieves tissue closure after multiple operations. Several surgical techniques exist, with a lack of consensus as to the optimal approach. Choice of technique

varies with defect size and type, newborn size, and associated conditions.^{3,14,16} Closure can be achieved primarily, or utilizing amnion-inversion, prosthetic silos, vacuum-assisted closure, tissue expanders, synthetic mesh, component separation, or skin flap creation.¹⁴

Given the absence of standardized, evidence-based guidance, clinical decision-making is often inconsistent and dependent on individual surgeon preference and institutional experience. To address this gap, the European Paediatric Surgeons' Association (EUPSA) initiated a structured consensus process aimed at evaluating the literature and current peer practices, with a focus on conservative management, surgical strategies, and clinical outcomes. This process aimed to provide clarity and consistency in the management of giant omphalocele, culminating in the development of a peer-reviewed consensus statement intended to support pediatric surgeons in informed decision-making for this pathology.

Methods

Seven pediatric surgeons, appointed as panelists by the EUPSA Consensus Committee, were tasked with reviewing the literature to synthesize evidence relating to giant omphalocele, with the aim of developing a consensus statement focused on postnatal management and outcomes.

Three subtopics were initially determined based on current evidence gaps within the literature: (1) conservative management (definition of giant omphalocele; painting substances—types, outcomes, and toxicity; painting to surgery timing), (2) surgical management (classification of techniques, median number of procedures, approach to intervention), (3) outcomes of omphalocele (factors influencing outcomes, ruptured omphalocele outcomes, complications in management).

Systematic search across Cochrane, Medline, and EMBASE was conducted for each subtopic until April 2023 using search terms “Omphalocele,” “Exomphalos,” “Giant,” and “Closure” or “Outcome.” Truncation, wildcards, and Boolean operators were applied to include all relevant papers. Abstract screens were conducted for each subtopic, followed by full-text screens. The management approach was agreed by

attendees and alternative approaches accounted to formulate the consensus statement.

Panelists were assigned to specific subtopics, with discrepancies resolved through structured group discussion and further literature review. Consensus was reached through unanimous agreement among the contributing members.

Each subtopic was presented with available evidence to congress participants at the 2023 EUPSA Annual Congress. Specific questions were posed by the chair (A.K.S.). Panelists and participants were invited to share their perspectives and alternative viewpoints based on clinical practice; all questions raised were incorporated into this statement. Discussion was focused on postnatal management, which is most relevant to neonatal and pediatric surgical practice.

Results

Definition of Giant Omphalocele

Definitions of giant omphalocele vary substantially within the literature, though they can be broadly categorized into anatomical definitions and those based on the inability to achieve primary closure.

Anatomical definitions include a defect diameter greater than 5 cm, a large sac diameter (without specific measurement), a sac containing part or all of the liver, and viscerio-abdominal disproportion.⁷ The second category, inability to perform primary closure, serves as a broader term not solely dependent on anatomical characteristics, but also on increased risks associated with neonatal comorbidities, such as prematurity, respiratory distress, and cardiac failure.¹⁷ The term “viscero-abdominal disproportion” effectively encompasses both definitions, by evaluating the size of the sac relative to the neonate, to guide decisions on primary closure versus conservative management. This definition accounts for variability in neonatal weight and size, which may not be adequately addressed in definitions based solely on sac characteristics. The prenatal omphalocele circumference/abdominal circumference ratio, determined using two-dimensional ultrasound at three gestational intervals, has been validated as a prognostic tool for predicting the type of surgical closure and survival outcomes.¹⁸ Prospective studies to standardize measurement techniques and define ratios for giant omphalocele would be of value.

Consensus on the Definition of Giant Omphalocele

Giant omphalocele is defined as omphalocele that cannot be closed primarily due to large viscerio-abdominal disproportion or due to the presence of significant comorbidities that delay primary closure beyond a month after birth.

Conservative Management of Omphalocele

Indications for “Paint and Wait” Conservative Management in Giant Omphalocele

“Paint and wait” is the application of escharotic agents onto the omphalocele sac to encourage epithelialization, with subsequent delayed surgical closure.¹⁹ Indications include (1) defects that cannot be closed anatomically, often those with significant viscerio-abdominal disproportion, (2) neonates

with significant comorbidities, increasing anesthetic/surgical risks, (3) under-resourced environments where advanced surgical, anesthetic, and NICU care standards are not met.^{19,20}

Materials Used for Painting the Omphalocele Sac

Povidone-iodine, silver sulfadiazine, dissodic 2% aqueous eosin, *Acacia nilotica*, Gentian violet, and Manuka honey have been described as painting agents across case reports and series, with mixed findings regarding toxicity. Povidone-iodine and silver sulfadiazine are the most frequently reported agents.²¹ Manuka honey, initially used in the treatment of adult burns, has been introduced into omphalocele management with promising outcomes and minimal complications reported thus far.²² Discussion at the Consensus Statement Session highlighted the use of bismuth-based preparations and dressing the sac without any agents, though there is a lack of clear evidence in the literature to support these approaches.

Dose, Amount, and Duration of Painting Agents

Dosage, quantity, and duration of use varied between painting agents and preparations. Topical povidone-iodine, silver sulfadiazine, dissodic aqueous eosin, Gentian violet, and *Acacia nilotica* required daily application, while silver sulfadiazine and Manuka honey impregnated dressings were changed two to three times per week (→Table 1).^{7,20–26} Of note, reported dosages are derived primarily from observational studies and case reports; thus, they should be regarded as descriptive summaries, rather than evidence-based recommendations.

Complications and Side Effects of Painting Agents

Alcohol and mercurochrome, once widely utilized in omphalocele management, were largely abandoned following reports of mercury poisoning associated with mercurochrome and neurological and cardiac complications linked to alcohol.^{27,28} Complications of modern painting agents have been inconsistently reported, with variability between studies. In high concentrations, povidone-iodine has been associated with thyroid dysfunction,²⁹ while silver sulfadiazine has been linked to neuropathies, seizures, neurological complications, and rarely sepsis and bowel perforation.¹⁷ Similarly, the use of dissodic 2% aqueous eosin has been implicated in sepsis and bowel obstruction, with a proposed etiopathogenic mechanism involving intestinal inflammation leading to functional bowel disorder and subsequent occlusion, potentially exacerbated by unrecognized anatomical anomalies such as intestinal malrotation.³⁰ Manuka honey has been associated with botulism, hyperglycemia, and allergy, though sparsely reported²² (→Table 2).

Time Between Start of Conservative Management to Surgical Closure

The interval between initial application of painting agents and surgical closure varied across different agents and studies. One study for povidone-iodine found an average time to surgical closure of just under 15 months, while another reported 10 months.^{7,21} Silver sulfadiazine had a similar range at around 4 to 16 months²⁰ compared with an

Table 1 Dose, amount, and duration of use for painting agents used in the conservative management of giant omphalocele

Painting agent	Dose, amount, and duration
Povidone-iodine	5% daily + antibiotic powder twice daily 1–2.5% diluted iodine 1:4–1:10 every 1–2 days
Silver sulfadiazine	Topical products: i.e., 20 g cream daily for 3–4 weeks Silver-impregnated hydro-fiber dressing, changed every 3–7 days
Dissodic aqueous eosin	2%, twice daily
Gentian violet	1%, 1–2 mL 1–2 times daily
<i>Acacia nilotica</i>	Combined with Gentian violet—twice daily
Manuka honey	Ointment + impregnated gauze, changed every 2–3 days Mixed with 1% Manuka oil—2–3 times weekly

Table 2 Complications and side effects of painting agents used in the conservative management of giant omphalocele

Painting agent	Complications and side effects
Povidone-iodine	Hypothyroidism
Silver sulfadiazine	Neutropenia Nephrotic syndrome Peripheral neuropathy Seizures Sepsis and bowel perforation (rare)
Dissodic 2% aqueous eosin	Bowel obstruction Sepsis
Mercurochrome	Mercury poisoning
Alcohol	Neurological and cardiac complications
Manuka honey ³⁶	Botulism Hyperglycemia Allergy

average of 17 months for Manuka honey, though based on relatively few reports²⁵ (►Table 3).

Time to Full Epithelialization

The concept and definition of full epithelialization remains controversial, with debate over the degree to which an objective measurement can be made. Generally, full epithelialization is considered the point at which the sac appears entirely covered by skin. This was typically achieved after approximately 2 to 4 months, with some variation between reports.^{7,20,22,24,31} For silver sulfadiazine, the timeframe

Table 3 Time from initial application to surgical closure for painting agents used in the conservative management of giant omphalocele

Painting agent	Time to surgical closure (months)
Povidone-iodine	14.78 ± 3.24 (6–18) 10 (9.2–12)
Silver sulfadiazine-impregnated dressings	10 (4–16)
Manuka honey	17 (11–38)

Note: Aggregated data reported as average (range).

Table 4 Time from initial application to full epithelialization for painting agents used in the conservative management of giant omphalocele

Painting agent	Time to full epithelialization
Povidone-iodine	7.83 ± 4.82 weeks 4 months (3–6)
Silver sulfadiazine	Cream: 5 months (1–12) Dressings: 2.7 months (0.9–4.4)
Manuka honey	2 months (1.6–4) 2.4 months (0.9–6.6)
<i>Acacia nilotica</i>	7.42 ± 6.59 weeks
Dissodic 2% aqueous eosin	2.3 ± 0.23 months

Note: Aggregated data reported as average (range).

varied by method of administration, with cream achieving epithelialization after 5 months, compared with just under 3 months for impregnated dressings.^{20,31} Manuka honey, in contrast, demonstrated epithelialization at around 2 months²² (►Table 4).

Average Length of Hospital Stay

Length of hospital stay ranged from around 1 to 3 months for povidone-iodine, silver sulfadiazine, and Manuka honey.^{3,22} Of note, hospitals in Africa reduced inpatient durations by training parents to paint the omphalocele sac at home, with regular follow-up to monitor progression³⁰ (►Table 5). Although no studies specifically investigated or compared the costs of different painting agents, shortened hospital stay was considered an important factor in reducing financial burdens, though contingent upon appropriate education and support.

Consensus on Conservative Management of Giant Omphalocele

Conservative “paint and wait” management is recommended when anatomical constraints or high surgical risk preclude primary closure (►Fig. 1). Povidone-iodine and silver sulfadiazine are the most frequently reported painting agents, with Manuka honey gaining interest and increasingly used across various institutions.

Table 5 Time from initial application to surgical closure for painting agents used in the conservative management of giant omphalocele

Painting agent	Time to surgical closure (days)
Povidone-iodine	27 (14–34)
Silver sulfadiazine	40 (20–78)
Manuka honey	66 (21–121)
Dissodic 2% aqueous eosin	21 $\pm$ 6 Parents painting at home: 11 (1–60)

Note: Aggregated data reported as average (range).

There is no clear recommendation regarding the optimal dose, duration, interval between application and closure, or complications for different painting agents. As such, randomized controlled trials are required to establish an evidence-based consensus on outcomes and to determine the most effective agent for use.

Surgical Management of Giant Omphalocele

For consensus on surgical management of giant omphalocele, literature search identified 378 articles, of which 36 met inclusion criteria (►Table 6). These articles comprised 8 case reports, 9 case series, and 19 retrospective case note reviews, analyzing a total of 356 patients with giant omphalocele.

Intervention Classification

Surgical techniques were grouped into early and delayed methods. Early interventions, defined by definitive closure of the fascia and skin prior to discharge, were subdivided into (1) primary fascial closure with sutures and (2) closure using a biological or synthetic patch. Delayed methods, defined as definitive closure after discharge, were similarly classified based on patch use. Delayed closure without a patch included (1) simple primary closure with sutures and (2) composite closure employing adjunctive techniques such as (i) flap and component separation methods, (ii) botulinum toxin injection with progressive pneumoperitoneum, and (iii) use of tissue expanders.

Early Intervention

The early intervention group comprised 18 articles (5 case reports, 5 case series, 8 retrospective case note reviews).^{32–47} Three studies reported both early and delayed closure, with subgroup analysis enabling evaluation of outcomes for each technique. A total of 170 patients were analyzed; just over half ($n=96$, 56%) of the patients were managed without the use of a patch. Among patch repairs ($n=74$, 44%), 58% ($n=43$) utilized Goretex, while 42% employed biological meshes including bovine ($n=1$), human acellular dermal matrix and small intestinal submucosa ($n=24$), and Bovine Pericardium Gen P ($n=6$).

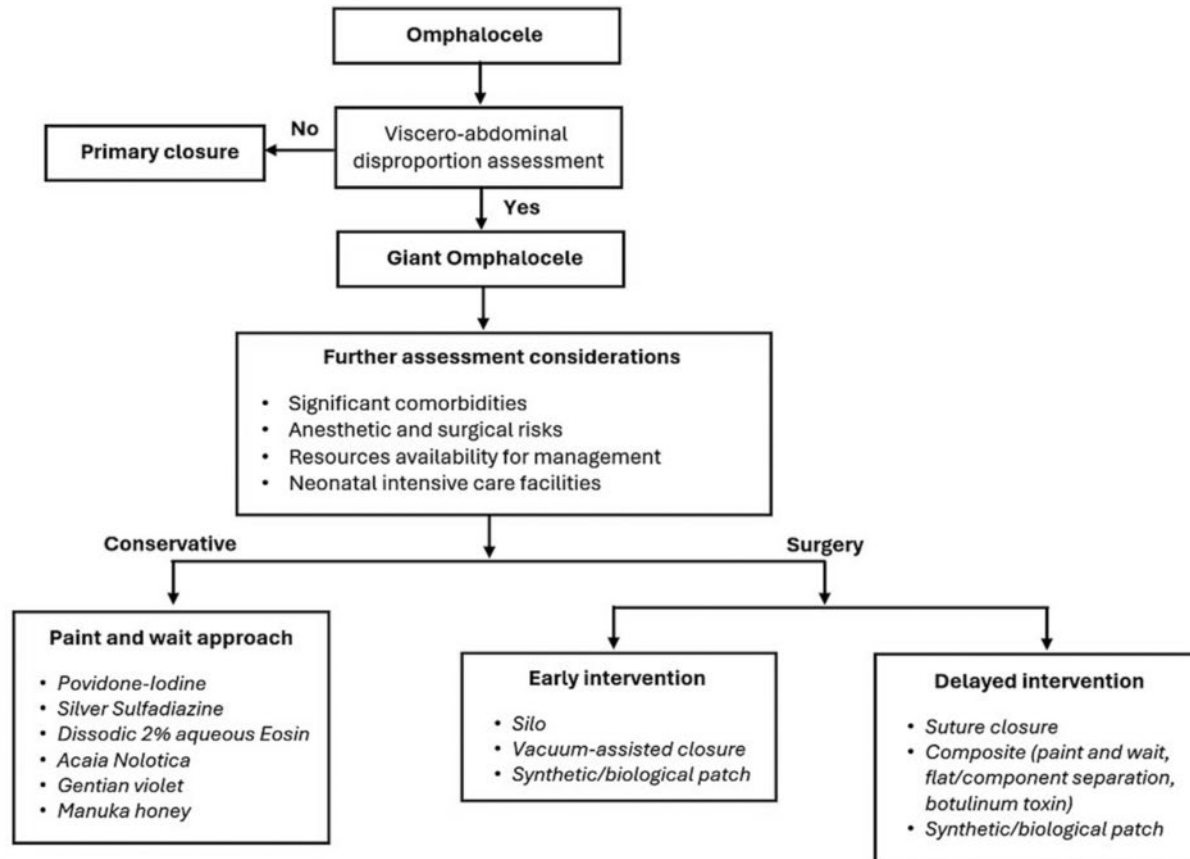


Fig. 1 Management options and pathway for giant omphalocele.

Table 6 Inclusion and exclusion criteria for consensus on surgical management of giant omphalocele

Inclusion	- Articles mentioning “Giant Omphalocele” - Discussion of surgical technique used - Discussion and follow-up of patients to the point of fascial and skin closure - Analysis of outcomes grouped by treatment method
Exclusion	- Omphalocele only - Initial nonoperative approach only

Various techniques were reported to reduce viscerobdominal disproportion. Seven studies surgically converted the sac into a silo, four reported serial sac ligation, and four used dressings, including a novel vacuum-assisted closure (VAC) dressing to address patch-related morbidity such as infection, facilitating wound healing and skin closure.³⁸ In three papers, the technique was not recorded. Procedures to convert the sac into a silo included inpatient application of a neoprene binder, designed by the hospital's orthotic department, which allowed for gradual reduction of the viscera until an early method of closure could be achieved.⁴⁸ In addition, the Abello technique described the use of Duoderm to reinforce the sac, followed by gradual reduction using tongue depressors as clamps in the apex of the silo. Once the Duoderm could be inverted, the omphalocele was deemed ready for closure.¹⁶ Median number of procedures for early intervention was poorly documented, and unreported in seven articles. Where recorded, the median number of procedures was two, with a range of one to six.

Morbidity in the early closure group was high at 54% ($n=92$), reflecting the complexity of giant omphalocele patients who often present with substantial comorbidities requiring numerous invasive interventions. Mortality in this group was 8% ($n=13$). Subgroup analysis of primary closure revealed morbidity of 49% and mortality of 6%. The synthetic patch group exhibited a higher morbidity (74%) and mortality (14%). Within the biological patch group however, morbidity was the lowest of the three methods at 42%, and mortality fell to 3%.

In evaluating causes of delayed surgery, one article identified respiratory insufficiency as a primary factor. In this study, patients with respiratory insufficiency underwent closure at around 76 days, while those without respiratory insufficiency achieved closure on day 1.⁴⁶

Consensus on Early Intervention for Giant Omphalocele

Among early interventions, biological patch closure utilizing bovine pericardium, human acellular dermal matrix, or small intestinal submucosa achieved the most favorable outcomes. In contrast, synthetic patch closure was associated with the worst outcomes, characterized by higher rates of morbidity and mortality.

Delayed Intervention

Delayed closure was described in 18 articles (3 case reports, 5 case series, 10 retrospective studies).^{7,20,31,46,49–62} A total of 186 patients were reported, with only 17% requiring a patch,

compared with 42% in the early closure group. This was likely attributable to reduced viscerobdominal disproportion observed in older children with access to techniques including component separation, botulinum toxin injections, and tissue expanders which were not available at birth.

For delayed closures without a patch ($n=170$), 78% were simple suture closures while 22% were composite. Composite methods included skin flaps and component separation ($n=32$, 17%), botulinum toxin with progressive pneumoperitoneum ($n=1$, 1%), and tissue expanders ($n=7$, 4%).

For delayed patch closure ($n=16$), there was an equal distribution between biologic meshes (Surgisis, Tutopatch, Alloderm) and synthetic patches (Goretex, Prolene) ($n=4$ in each group). Patch type was not recorded for eight patients.

In contrast to early closure, initial management for delayed closure typically involved a “paint and wait” approach, rather than techniques aimed at serial reduction over days.

Delayed Closure Without Patch—Suture Closure

For delayed simple suture closure ($n=146$), 72% were initially managed with topical treatments including dressings, silver sulfadiazine, and povidone-iodine. Conversion of the sac into a silo was described in 11 cases (8%), while initial management was not reported in 30 cases (21%).

The median number of procedures was unreliably reported, with a significant number unrecorded (100 patients across 4 articles). Notably, some incisions or reductions in early closure methods were performed under general anesthesia; however, the number of procedures was not specified.

Delayed simple closure had a morbidity of 9% (9/105) including episodes of infection and sepsis, though a significant proportion were not recorded. Mortality for this approach was 18% (27/146).

Delayed Closure Without Patch—Composite

For delayed composite closures ($n=40$), an initial “paint and wait” approach was reported in 78%. Initial management of the sac was not documented in 9 cases (22%).

Several flap and component separation techniques have been described, including the San Martin technique, originally developed for large midline incisional hernias, and the Lazarro da Silva technique.^{52,53} Other methods include component separation, as well as triple- and double-layered flap techniques.^{54,55} The median number of procedures was one; however, this parameter was not recorded for 10 out of 32 patients. Morbidity, where reported, was 28% (5/18), primarily related to sepsis and surgical complications including bleeding, wound infection, and skin necrosis. No mortality was reported for these techniques.

For tissue expanders, procedure counts were inaccurately recorded, ranging from one to “multiple.”^{56–58} Typically, tissue expanders were inserted under general anesthesia, with subsequent weekly outpatient visits to incrementally increase volume. Morbidity and mortality were often unreported, as papers highlighted technique rather than outcomes.

One case report utilized botulinum toxin to relax the abdominal wall with progressive pneumoperitoneum to achieve defect closure, with no associated morbidity or mortality.⁶⁰

Delayed Closure with Patch

For delayed patch closure, initial management was recorded in 8 of 16 cases, with 7 utilizing topical treatment and 1 employing a biological patch. The number of procedures was greater compared with closure without a patch, ranging from three to multiple (though unrecorded in two studies).

Morbidity for delayed patch closure was 25%, including patch infection and recurrent hernia. Notably, mortality was significantly higher at 63%, related to associated anomalies in 9 out of 10 cases. This likely reflects a self-selected older population with the most challenging defects to close, alongside additional factors not discussed in the literature.

Consensus on Delayed Intervention for Giant Omphalocele

For delayed closure techniques, utilizing native tissues yielded favorable outcomes. Flap and component separation techniques required the fewest procedures (median 1), with morbidity of 28% and no associated mortality. Simple suture closure had the lowest morbidity at 9%, though mortality was higher at 18%, with a median of three procedures. Delayed patch closure had the highest morbidity (25%) and mortality (63%).

It is important to acknowledge the inherent bias in morbidity and mortality rates. Subgroup analysis differentiating isolated cases of omphalocele from those involving complex comorbidities may provide more reliable results. Additionally, establishing a registry via a Delphi consensus could offer valuable insights.

Consensus on Surgical Management of Giant Omphalocele

Owing to the large number of techniques and insufficient information available for each, a consensus on the optimal approach to surgical management could not be established.

In early closure, biological patches are likely to offer improved outcomes owing to their integration with native tissues and potential for resorption in cases of infection. Conversely, for delayed closure methods utilizing the patient's own tissues appear most effective. Delayed methods appear more conducive to primary closure, with reduced reliance on patches and greater flexibility in the techniques that can be employed. For patients requiring patches for delayed closure, mortality is high, likely reflecting a population with the most severe defects. A summary and comparison of early and delayed closure are provided in [Table 7](#) and [Fig. 2](#).

In summary, no single treatment modality is suitable for all giant omphaloceles. Each center operates within a unique

surgical environment with diverse experiences. The presence of comorbidities in omphalocele patients plays a crucial role in determining appropriate management strategies. Employing an index that incorporates these factors may be beneficial in guiding treatment decisions.

Outcomes in Omphalocele

Literature search focused on outcomes in omphalocele aimed at identifying high-level evidence, including systematic reviews, meta-analyses, randomized controlled studies, comparative cohort studies, and large cohort studies involving more than 100 live births. The primary outcome was mortality, with associated anomalies considered a factor of special interest.

Factors Influencing the Outcome of Patients with Omphalocele

Two major reviews evaluated prognostic factors, one incorporating 23 studies comprising 396 giant omphaloceles,⁸ the other with 88 studies, primarily focused on cardiac anomalies.⁶³ Additionally, eight multicenter or registry studies, with cohorts ranging from 123 to 2,308 patients, were identified.^{64–71}

The largest registry study, utilizing data from the 1995–2005 National Birth Defects Prevention Network, found that among 2,308 cases of omphalocele, only 22% were isolated; the majority were associated with other birth defects, chromosomal abnormalities, central nervous system disorders, or congenital heart defects.⁶⁶ This study reported key prognostic factors including defect type (isolated or with associated anomalies) and birth weight. Another key predictor of

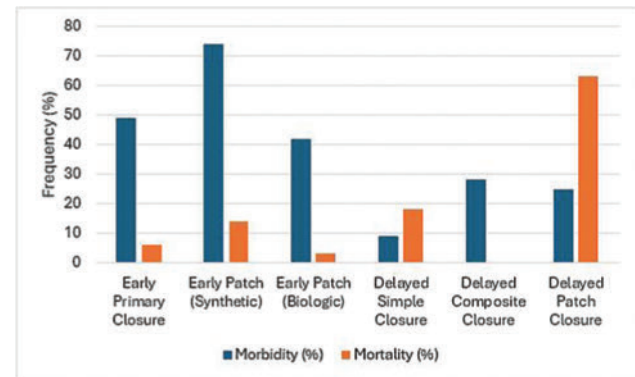


Fig. 2 Comparison of morbidity and mortality for early and delayed closure techniques employed in the management of giant omphalocele.

Table 7 Summary and comparison of early versus delayed closure in the management of giant omphalocele

Factor	Early closure	Delayed closure
Number of patients	170	186
Patch use	44%	17%
Morbidity	54% overall	Simple: 9%; Composite: 28%; Patch: 25%
Mortality	8% overall	Simple: 18%; Composite: 0%; Patch: 63%
Median number of procedures	2 (poorly documented)	1 (composite), 3 (simple), multiple (patch)

mortality was respiratory insufficiency, with prematurity and ruptured sacs included in this group.⁸ Independent predictors of mortality included chromosomal anomalies, congenital heart defects, central nervous system defects, female sex, lung hypoplasia, defect size, and birth weight.^{64–71} Factors with no influence were gestational age, necrotizing enterocolitis, maternal diabetes, obesity, smoking, hypertension, and mode of delivery.^{64,65,68}

Consensus on Factors Influencing the Outcome of Patients with Omphalocele

Based on data from large cohort studies and registries, key prognostic factors for patients with omphalocele were identified, though contradicting results have been published. Commonly cited factors include associated chromosomal and cardiac anomalies, as well as low birth weight.

Recommendations

Factors influencing postnatal management outcomes and mortality should be acknowledged when counseling parents. Active screening for these factors is essential for all omphalocele patients and should be taken into consideration when determining treatment strategies, provided the clinical situation permits.

Outcomes of Patients with Ruptured Omphalocele

Limited evidence exists regarding outcomes in ruptured omphalocele, primarily derived from case reports and series with less than 10 patients. One large review of 23 studies identified only 22 cases of ruptured giant omphalocele.⁸ Additional cohort studies included 11 to 18 patients, with mortality rates ranging from 32 to 50%.^{72–74}

There is significant heterogeneity in techniques used to manage ruptured omphalocele. Acute care typically involves placing the child in a sterile bag and obtaining intravenous access to initiate fluid resuscitation and antibiotics. Ongoing management approaches may include closure of the rupture with initiation of topical agents, the use of a silo for gradual reduction, or direct defect closure, with or without the use of a patch.

Consensus on Outcomes in Patients with Ruptured Omphalocele

Data regarding treatment outcomes for ruptured omphalocele are scarce and predominantly based on case reports and series with small patient cohorts. Evidence suggests that children with ruptured omphalocele have an increased risk of mortality.

Recommendation

An international study is necessary to evaluate treatment outcomes in larger populations of children with ruptured omphalocele. Rupture of omphalocele should be prevented if possible. When rupture occurs, prompt surgical intervention combined with intravenous antibiotics is recommended to prevent sepsis.

Complications That Can Occur in Omphalocele Management

Limited evidence is available relating to complications that occur in the management of omphalocele. Articles identified

included one systematic review, one narrative review, and one cohort study of 170 patients.^{14,75,76}

Complications were divided into two groups: (1) inherent to omphalocele and (2) associated with interventions. Inherent complications included mortality (approximately 10% in isolated cases; 25–40% in complex omphalocele), feeding difficulties, respiratory insufficiency, malrotation and volvulus, and neurodevelopmental issues. Intervention-related complications included infection, small bowel obstruction, and abdominal compartment syndrome. Of note, kinking of the portal vein and hepatic vessels was believed to be underreported in the literature.

Consensus on Complications That Can Occur in Omphalocele Management

Complications following management of omphalocele are relatively rare and typically intervention-related. Consequently, most complications arise within the first few years of life.

Recommendation

Information regarding intervention-related complications should be discussed when counseling parents. Long-term multidisciplinary follow-up is essential to enable early identification and management of potential complications.

Limitations

There is a scarcity of high-level evidence within the literature regarding the management of giant omphalocele, with the vast majority of data available from case reports, series, and small retrospective reviews. As such, recommendations should be applied in the context of individual patient characteristics, clinical setting, and institutional expertise.

Due to significant heterogeneity within the existing literature, it was not feasible to conduct a meta-analysis or perform meaningful statistical comparisons. Definitions of giant omphalocele vary across studies, resulting in differing patient populations and clinical presentations. Additionally, comorbid conditions, which are key determinants of outcomes, were inconsistently reported. These limitations precluded standardization and reduced the reliability of any pooled statistical analysis.

Conclusion

A peer-reviewed consensus statement on the management of giant omphalocele has been developed based on current evidence and peer practice.

Conservative “paint and wait” management is recommended when primary closure is not feasible as a result of significant viscero-abdominal disproportion or when comorbidities increase surgical risk. For early surgical closure, biological patches are associated with lower rates of morbidity and mortality and are thus preferred over synthetic materials. For delayed closure, techniques utilizing native tissues, including simple suture and composite methods such as component separation, are associated with the most favorable outcomes. Poor prognostic indicators include

chromosomal and cardiac anomalies, as well as low birth weight.

Future research should prioritize the development of standardized definitions and outcome measures, with the establishment of multicenter registries and collaborative studies to strengthen the current evidence base. This will enable the refinement of management strategies, promoting more consistent care and optimizing outcomes for patients with giant omphalocele.

Conflict of Interest

None declared.

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