A Past Anomaly, a Present Crisis: Appendicitis in a Teenager with a Background of Exomphalos Repair

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Abstract

Exomphalos (omphalocele) is a congenital abdominal wall defect characterized by herniation of abdominal viscera through a central defect at the umbilicus, covered by peritoneum and amnion. It is frequently associated with other congenital anomalies, particularly of the cardiovascular, gastrointestinal, genitourinary, and musculoskeletal systems. Surgical repair in infancy can result in dense intra-abdominal adhesions, altered organ positioning, and abdominal wall deformities, which complicate subsequent abdominal procedures. While laparoscopic appendectomy is widely regarded as the standard treatment for uncomplicated appendicitis due to its benefits of reduced postoperative pain, wound infection, and shorter hospital stay, prior major abdominal surgery often mandates open access to avoid iatrogenic injury. We report a rare case of acute subhepatic appendicitis in a 16-year-old male with a complex history of congenital and surgical anomalies following exomphalos repair.

Introduction

Exomphalos (omphalocele) occurs in approximately 1 in 4,000 live births .[1] The condition involves failure of the midgut to return to the abdominal cavity during embryologic development, resulting in herniation of abdominal contents into the umbilical cord base, covered by a thin membrane of peritoneum and amnion [1]. Up to 60% of infants with exomphalos have associated anomalies, most commonly cardiac defects (35–40%), gastrointestinal malformations, and chromosomal abnormalities such as trisomy 13 or 18 [1]. Advances in neonatal surgery and intensive care have markedly improved survival, but postoperative complications—especially adhesive bowel disease and altered organ positions—can persist lifelong. [2,3] The following case highlights how these anatomical and surgical factors can influence surgical decision-making when managing acute appendicitis later in life.

Case Report

A 16-year-old male presented to the Emergency Department at the Royal Hospital, Oman, with a three-day history of right flank pain associated with nausea, reduced appetite, and diarrhea. There was no vomiting, fever, or urinary symptoms.

Past medical history included antenatally diagnosed exomphalos (omphalocele), double outlet right ventricle with left pulmonary artery stenosis, left-sided cerebrovascular accident in infancy resulting in right-sided hemiplegia, and chronic lung disease related to prolonged ventilatory support.

Past surgical history of exomphalos repair, fundoplication and gastrostomy tube placement, aortopexy for tracheomalacia, and surgical correction of double outlet right ventricle in early childhood.

On examination, the patient was hemodynamically stable and afebrile. Inspection revealed well-healed midline sternotomy and laparotomy scars with a mild upper abdominal wall deformity [Figure 1]. The

abdomen was soft, with localized tenderness over the right flank and subcostal region, but without guarding or rigidity. No palpable masses or organomegaly were detected.



Figure 1: Preoperative examination showing multiple well healed laparotomy scars, midline sternotomy scar.

Routine laboratory studies were unremarkable except for C-reactive protein (CRP) of 39 mg/L, indicating mild inflammation. White blood cell counts and liver function tests were within normal limits.

Contrast-enhanced CT abdomen showed an inflamed, non-perforated appendix located subhepatically, consistent with acute subhepatic appendicitis [Figure 2]. There was no abscess or free intraperitoneal air.



Figure 2: Ct scan images: red arrows pointing at acute non-complicated appendicitis in the subhepatic region measuring 10mm in diameter with surrounding fat stranding, with peritoneal thickening. no free fluid, no pneumoperitoneum.

Given the patient's extensive surgical history and expected dense adhesions, a laparoscopic approach was deemed high-risk, and open exploratory laparotomy was planned after detailed discussion and informed consent from the patient and family.

Intraoperatively, previous midline incision was explored, dense adhesions were noted between the anterior abdominal wall, small bowel loops, and the omentum. The cecum and appendix were located in a subhepatic position, adherent to the inferior surface of the right lobe of the liver [Figure 3]. The appendix

was inflamed but not perforated careful adhesiolysis was performed to expose the appendix, which was then ligated and removed conventionally [Figure 4].

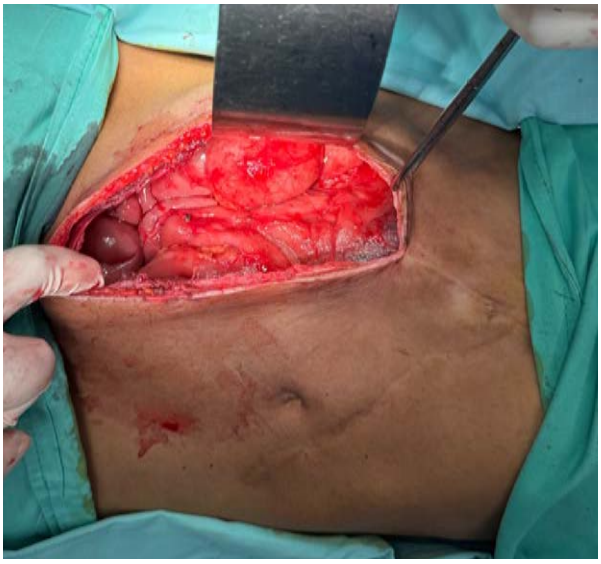


Figure 3: Intraoperative picture showing the appendix after adhesiolysis.



Figure 4: Excised appendix.

No intraoperative complications occurred. The abdominal cavity was irrigated, and the incision was closed in layers after confirming hemostasis.

The postoperative course was uneventful. Oral intake was resumed on postoperative day two, and the patient was discharged on day three. At two-week follow-up, he remained asymptomatic with well-healed wounds

Discussion

This case underscores the unique challenges of managing acute appendicitis in patients with repaired exomphalos and multiple prior abdominal surgeries.

In the context of this case, the decision to proceed with appendectomy rather than conservative antibiotic management warrants discussion. Our patient was a 16-year-old with exomphalos and a history of multiple abdominal surgeries, placing them in a high-risk category for non-operative management. Although the CODA trial demonstrated that antibiotic therapy can be non-inferior to surgery in the short term, it also reported a higher rate of appendectomy within 90 days and an increased long-term need for surgery among patients initially treated with antibiotics.[4] In patients with complex congenital abdominal anatomy and prior operative intervention, failure of conservative treatment may result in recurrent symptoms, repeated hospital admissions, and prolonged antibiotic exposure. Furthermore, delayed appendectomy in such patients may be technically more challenging due to adhesions and altered anatomy, potentially increasing operative risk. In this setting, early surgical intervention provided definitive treatment, reduced the risk of recurrence, and offered a more predictable and durable outcome, supporting the decision for appendectomy in our case.

Laparoscopic appendectomy offers several advantages in the general population, but in patients with extensive intra-abdominal adhesions, distorted anatomy, or prior congenital anomaly repair, the risk of visceral or vascular injury during trocar insertion is significantly higher.

In exomphalos survivors, studies have shown:

- Adhesive small bowel obstruction rates of 8–10% during long-term follow-up.
- Abnormal organ positions, including malrotation and subhepatic or retrocecal appendix, increasing diagnostic difficulty.
- Abdominal wall defects or prosthetic repairs that complicate safe laparoscopic access.

In this patient, the subhepatic location of the appendix likely resulted from altered cecal positioning due to congenital or postoperative adhesions. CT imaging was crucial for accurate preoperative localization, which guided the surgical approach.

Given these anatomical complexities, an open approach for appendectomy allowed controlled adhesiolysis, direct visualization, and safe dissection in a high-risk abdomen. The decision to avoid laparoscopy was justified by both patient safety and the high probability of conversion. Finally, the case raises the consideration of prophylactic appendectomy in patients with repaired exomphalos undergoing future laparotomies, as this may prevent future diagnostic dilemmas or emergency reoperations in complex abdomens.

Conclusion

Exomphalos repair in infancy often leads to lifelong anatomical and surgical challenges. In such patients presenting with acute abdominal conditions, careful preoperative imaging, individualized planning, and intraoperative flexibility are essential. While laparoscopic surgery remains the gold standard for appendicitis, an open approach remains safer in selected patients with extensive adhesions and distorted anatomy. Prophylactic appendectomy may be justified during future planned laparotomies in exomphalos survivors. The following practical considerations summarize key learning points from this case: Repaired exomphalos is frequently associated with dense adhesions and abnormal organ positioning. Laparoscopic appendectomy should be cautiously considered in patients with prior complex abdominal surgeries. Preoperative CT imaging is essential for anatomical mapping and surgical planning. Careful planning for abdominal access is essential in anticipation of difficult entry and abnormal anatomy. Open surgery remains an appropriate and safe approach in such complex cases. Prophylactic appendectomy may be justified during future planned laparotomies in exomphalos survivors.

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