Abstract:
Surgical disease has gained increased attention in recent years as contributing to the substantial global burden of disease. Congenital anomalies and surgically correctable ailments of newborns, such as those affecting the chest and abdomen, often require initial intervention with potential for long-term, disease-free survival. The true prevalence of these conditions, however, and the available resources for their management in low- and middle-income countries are unclear. This chapter provides an overview of congenital abdominal anomalies within the context of low- and middle-income countries, and a practical guide to recognition and initial management of those who present for care.

Keywords:
pediatric surgery; global surgery; congenital malformations; LMICs

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Abdominal Congenital Malformations in Low- and Middle-Income Countries: An Update on Management

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Congenital anomalies, and other surgical conditions specific to pediatric patients, contribute to the substantial global burden of disease attributed to surgically correctable ailments.[1] The actual incidence of congenital anomalies in low- and middle-income countries (LMICs) remains unknown, with estimates ranging from 12 per 1000 live births to 26-35% of neonates seeking health care in select LMICs.[2-4]

When considering the impact of congenital anomalies on the health of children in LMICs and the desire to save their lives, the deficit in the availability of pediatric surgeons must also be acknowledged. Compared to the United States, with 1 pediatric surgeon for every 108 305 children,[5] many LMICs suffer severe shortages of trained surgeons, from 0.49 surgeon per 100 000

population and only 1 pediatric surgeon for 5.6 million children in Rwanda[6] to 0.18 surgeon per 100 000 people in Sierra Leone where there are no practicing pediatric surgeons.[7] Recognition and appropriate care of children with a congenital anomaly that requires surgery involves identification of the appropriate and available provider. This imposes the need for greater vigilance of emergency care providers and a keen understanding of the means for access to definitive surgical care. Identification of the local surgical and anesthetic resources, including those for neonates, should occur prior to the presentation of a patient for emergent care to facilitate timely and appropriate definitive care.

The most critical time in the care of a newborn with a congenital anomaly occurs before the surgeon enters the picture. The initial resuscitation and timely identification of congenital anomalies are essential to survival beyond the newborn period.[8] Once the patient has survived the initial resuscitation and the anomaly in question has been recognized, activation of appropriate referral patterns (ideally already established) is key. Although less robust than in high-income countries (HICs), successful referral systems, such as those described in the neurosurgical literature for follow-up of patients with spina bifida in Uganda, do exist in LMICs.[9] In the adult literature, surveillance programs have been established in hopes of creating sustainable avenues for referral and transfer of surgical patients to highest-level hospitals.[10,11] Although not directly translatable to the pediatric population where timely (and, if possible, prenatal) diagnosis is essential for survival, valuable lessons can be deduced from existing screening and treatment paradigms in LMICs.

Although this chapter cannot fully unpack the nuances of a referral program, the following recommendations are provided to aide in the recognition and initial management of neonates with potentially survivable conditions that the emergency medicine clinician may encounter in LMICs.

INITIAL MANAGEMENT FOR THE FRONTLINE PROVIDER

Abdominal congenital malformations present in a limited number of ways and generally do not require complex investigations beyond examination and radiography. Similarly, initial management is not overly complex. Basic resuscitation, avoiding hypothermia (present in up to 85% of neonates presenting for surgical care in one study[8]), management of hypoglycemia, decompressing obstructed bowel, and prompt referral and transport to definitive care will allow many babies to survive to surgery who otherwise currently do not. Intravenous access with administration of saline boluses (20 mL/kg) titrated to clinical signs of adequate hydration should be included in the initial management of any neonate with bilious vomiting.[12] To avoid life-threatening and neurologically devastating hypoglycemia, maintenance intravenous (IV) fluids must include dextrose, ideally in a 10% solution for fasting neonates. Hypothermia is also a risk for neonates, particularly during resuscitation and transport. Kangaroo care, skin-to-skin with a parent or guardian, is the most straightforward way to avoid this. Other methods include wrapping the baby with cotton and/or blankets and placing warmed bean bags, or gloves filled with hot water around the child while taking care to avoid burn injuries. For babies with signs of a bowel obstruction, nasogastric (NG) decompression should be done, aspirating frequently or draining into a glove, to prevent emesis and aspiration. These infants should not be fed until definitive care is determined.

Although most congenital anomalies of the abdomen are diagnosed prenatally by ultrasound in HICs, most babies born with abdominal anomalies in LMICs have not had antenatal imaging or diagnosis and thus usually present within hours or days of birth with visible anomalies or signs of intestinal obstruction such as feeding intolerance/bilious emesis and/or abdominal distension.

Visible Anomalies: Omphalocele, Gastroschisis, and Sacrococcygeal Teratoma

Visible anomalies of the abdomen commonly include abdominal wall defects, omphalocele and gastroschisis, and sacrococcygeal teratomas (Figure 1). Of these, gastroschisis is the most immediately life-threatening, and the initial care has the greatest potential impact on survival. Mortality for gastroschisis in LMICs remains high, ranging from 50 to 100% in sub-Saharan Africa[13-16] compared to mortality of 5% in HICs.[17] After the defect is recognized, bowel coverage with clean plastic that will protect but not adhere to the bowel is recommended to prevent hypothermia, bowel injury, and insensible fluid losses. Placing the lower half of the baby in a sterile bag, tied under the arms, is common practice in HICs for transfer or management until surgery. A clean plastic shopping bag or plastic food wrap can be similarly effective at preventing heat and fluid losses in LMICs. Kangaroo care or skin-to-skin contact with the mother is encouraged to prevent hypothermia, ideally with the child positioned on their side with the bowel supported to prevent kinking of the mesenteric vessels as they come through the abdominal wall defect, and subsequent bowel ischemia. Intravenous access and administration of resuscitative

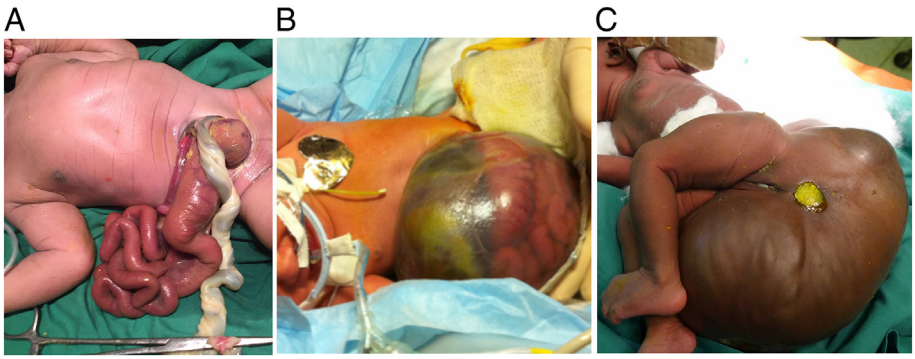

Figure 1. Externally visible congenital anomalies: (A) gastroschisis, (B) omphalocele, and (C) sacrococcygeal teratoma.

fluids with dextrose to prevent hypoglycemia, IV antibiotics for empiric neonatal sepsis treatment, NG decompression, and prompt transfer to a surgical center should follow.[18] These babies should not be breast-fed before surgical care is initiated. Time is critical for these children, and efforts should be made to transfer babies within hours of recognition of the defect. Surgical care emphasizes reducing the bowel into the abdomen and then closing the defect, either primarily if it will not put the baby at risk of abdominal compartment syndrome or, more commonly, gradually with a "silo"—premade if available or as improvised in some LMICs from a sterile urine or IV fluid bag which is sewn to the fascia under local anesthesia.[19,20]

Omphalocele becomes an emergency if the sac is ruptured, and if ruptured, it should be treated the same as gastroschisis. If the sac is intact, it is initially quite fragile, and management should focus on protecting and preserving it as an ideal covering for the bowel and promoting neoepithelialization. A variety of topical agents have been used successfully,[21] but generally, twice-a-day wrapping of the omphalocele sac with gauze moistened with a mild topical antibacterial solution allows these babies to go home with parental application of zinc oxide, dissodic 2% aqueous eosin, Sofra-Tulle gauze, or similar solution to the sac within 1-2 weeks of birth.[22,23] This allows skin to grow over the omphalocele sac, and then it becomes an elective hernia repair. Unlike gastroschisis, these babies usually have normal intestinal function and should be encouraged to breast-feed from birth. Babies with omphalocele have a high incidence of associated anomalies, in particular, chromosomal defects in 30-40% of cases and cardiac anomalies in 14-47% of cases.[24] A thorough physical examination for signs of these will help direct patients to appropriate care.

Sacrococcygeal teratomas (Figure 1) are often externally visible as a mass attached to the coccyx, and as long as the baby is passing urine and stool and does not have obvious bleeding, it is not an emergent concern but rather an urgent problem. Untreated, these masses go on to malignant degeneration over the first 2 years of life but require emergent neonatal management if they are complicated by obstruction of the genitourinary and/or gastrointestinal (GI) tracts or bleeding. In the case of a bleeding vessel, simple pressure would be the first step in management, with suture ligation of the vessel if it does not stop bleeding with prompt referral to a pediatric surgeon familiar with the problem.

Neonatal Intestinal Obstruction

Neonatal intestinal obstruction typically presents as bilious vomiting, abdominal distension, or failure or delayed passage of meconium. These are the most common presentations of newborns needing surgery for abdominal congenital anomalies. Any baby with bilious vomiting should be assumed to have a GI obstruction until proven otherwise. The most time sensitive is malrotation with midgut volvulus, which can occur at any age, presenting in the first week of life in 50% of cases and under 1 year of age in 90%.[25] Twisting of the mesentery causes obstruction of both the intestinal lumen and the blood supply to the intestine which can lead to bowel necrosis within 8 hours. Affected babies are typically healthy and are initially tolerating feeds and then have sudden onset of pain, bilious vomiting, and eventual lethargy and will progress to death if untreated. If this is suspected, an emergent surgical consult is warranted. Plain abdominal radiographs are rarely diagnostic, but an upper GI contrast study demonstrating an abnormal position of the duodenojejunal junction, to the right of the vertebral body, lower than the duodenal bulb, or anterior on lateral views, is diagnostic of the malrotation and should prompt immediate surgical evaluation.[25] If accessing pediatric surgical care would delay care by hours, the

closest general surgeon should be asked to provide care to save the child's life and bowel. In the interim, NG decompression, IV resuscitation with saline boluses and 10% dextrose in saline for maintenance fluids, warming the baby, and getting blood cross-matched should be initiated.

The more common presentation of neonatal intestinal obstruction includes gradual onset of abdominal distension and bilious vomiting over the first week of life. Thorough examination is critical to rule out an anorectal malformation and must include a digital rectal examination to ensure patency. Plain radiographs, with 2 views of the abdomen (Figure 2),[26] can show dilated loops of intestine and free air that might indicate a perforation. When the obstruction results in a "double bubble" on radiographs, duodenal atresia and/or annular pancreas is usually the problem. In the setting of an obvious, more distal, obstruction on radiographs, a contrast enema can be very helpful in determining the etiology, particularly differentiating intestinal atresias (microcolon, obstruction) from Hirschsprung disease (HD) (dilated bowel proximally transitioning to narrow caliber, patent intestine) (Figure 3). Similar to other anomalies, infants with suspected atresia, anorectal malformations (ARMs) or HD should have NG decompression, IV resuscitation and maintenance fluids, and warming to prevent hypothermia. Babies with neonatal intestinal obstruction require surgical care, which generally includes laparotomy and anastomosis for intestinal atresia, and emergent diverting colostomy for babies with ARMs and HD. Depending on center capabilities and experience, definitive reconstructive surgery in the neonatal period may be performed.[27,28] Delays in care can lead to perforation and life-threatening complications.

Anorectal Malformations

ARMs include a spectrum of congenital defects involving the most distal gastrointestinal tract, the anorectum, with variable involvement of the urogenital system.[29] Although a true birth incidence is difficult to ascertain, population-based estimates for the incidence of ARM report 1 in 5000 live births.[30] These malformations range from skin-level defects and perineal fistulas to complex anomalies such as cloacal. Akin to the anatomy, the identification and management of these anomalies are similarly varied. Although often presenting as part of a syndrome or complex series of anomalies, there is a lower incidence of associated malformations in most African literature likely due to the high neonatal mortality from undetected defects.[31,32] Identification of ARMs depends on a thorough physical examination to evaluate positioning of the anal opening with respect to the urogenital tract. If not identified at birth, ARMs are likely to present with intestinal obstruction, accounting for an estimated 57-67% of neonatal intestinal obstructions.[30]

Hirschsprung Disease

Increased awareness and technological advances in HICs have resulted in earlier and more efficient diagnosis of HD, with 90% of patients identified during the neonatal period.[33] In contrast, as in

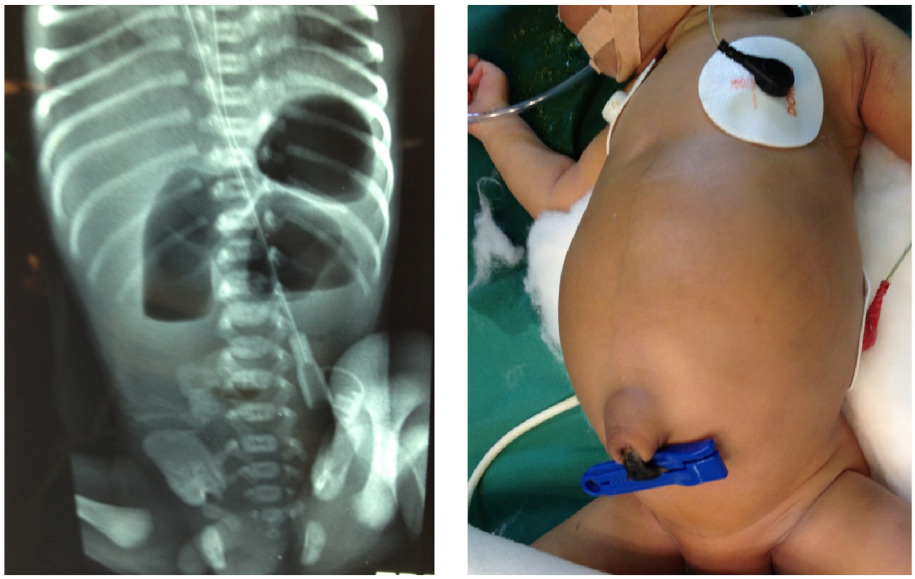

Figure 2. Intestinal atresia, triple bubble, and abdominal distention.

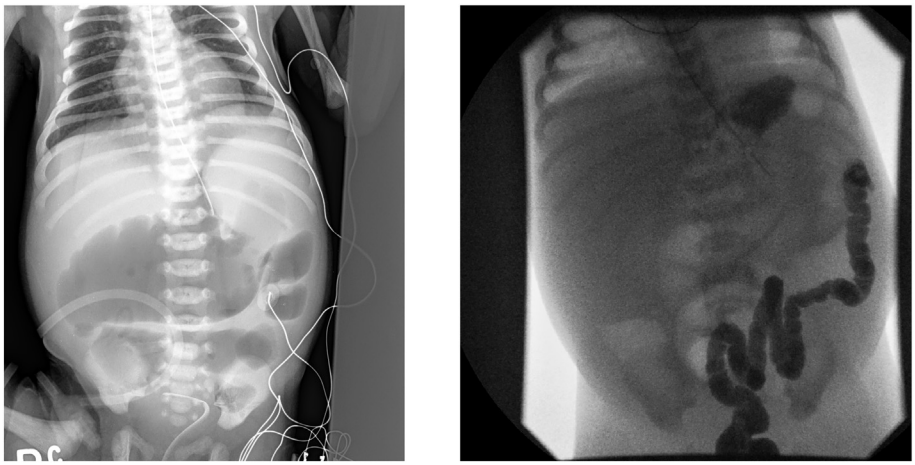

Figure 3. Intestinal atresia: plain radiograph and contrast enema showing microcolon.

many conditions without outwardly apparent anomalies, less than 45% of patients with HD present in the neonatal period in sub-Saharan Africa.[34,35] When evaluating a neonate or older child for abdominal distension, intestinal obstruction, feeding intolerance, or difficulty stooling, a detailed clinical history is needed to identify defining features of HD in contrast to other conditions. Notably, a history of failure to pass meconium within the first 24-48 hours of life and longstanding constipation, often with failure to thrive, is suggestive of HD and thus should prompt further investigation.[36] More severe presentations may include bowel perforation or Hirschsprung-associated enterocolitis in 5-10% of patients who present with a delayed diagnosis of HD.[37,38] These patients may present with sepsis, and for those or others who present with symptoms of abdominal sepsis, assurance of adequate airway, breathing, and circulation should be followed by aggressive administration of intravenous fluids, broad-spectrum antibiotics, and surgical consultation. Further management is likely to include rectal irrigations and creation of colostomy[39] prior to surgical pull-through for definitive management.

OPTIMAL RESOURCES FOR SURGICAL CARE

The Global Initiative for Children's Surgery (GICS) is a consortium of allies, institutions, and health care providers from both HICs and LMICs.[40] This initiative includes representation from 13 pediatric specialties. The GICS works to promote the inclusion of children's surgical care in existing health initiatives, provides guidance in strategic planning among surgical subspecialties, ensuring universal health coverage, and aids in upscaling of the global health workforce.[41] To accomplish these goals and to better serve the 1.7 billion children and adolescents without access to surgical care,[42] GICS published the Optimal Resources for Surgical Care project to provide guidelines for essential surgical care for children in LMICs.[43,44] Although these guidelines cannot address all aspects of patient care or nuances specific to geographic regions, they are meant to delineate the resources and personnel required to provide surgical services to children at various resource levels. With broad dissemination of these guidelines, governments and ministries of health will be pressed to meet the minimum requirements and help direct transport of complex patients to the hospitals best suited for their needs.[40,45]

SUMMARY

Given the common delay in diagnosis for patients with congenital abdominal anomalies in LMICs, the greatest pitfall to avoid is further delays in recognition and management. For the emergency care provider, having a high index of suspicion in patients who present with external signs of anomalies, vomiting, abnormal stooling, or abdominal distension should prompt initiation of resuscitation, and transport to surgical care is essential.

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