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A Case Study Examining the Current Evidence and Controversies in the Management of High-Risk Neonates With Malrotation

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A B S T R A C T

Introduction: Heterotaxy syndrome (HS) is a loss of the normal position of abdominal and thoracic organs and can be associated with severe cardiac disease and high mortality rates. Frequently associated intestinal malrotation further increases complications. Because of the high morbidity and mortality in this population, ongoing controversies exist regarding appropriate screening and management of these neonates.

Case: A neonate born with HS and cardiac disease underwent complex cardiac surgery on first day of life. He is later found to have malrotation with midgut volvulus and undergoes Ladd procedure with intestinal detorsion. He subsequently presents with small bowel obstruction necessitating a second abdominal surgical intervention.

Conclusion: Although surgery is necessary here, this case demonstrates why current controversy exists regarding initial screening and subsequent offering of prophylactic Ladd procedure in asymptomatic neonates with HS because of the risk of challenging complications.

Introduction

Heterotaxy syndrome (HS) is a complex diagnosis in which there is loss of the normal asymmetrical relation of thoracic and abdominal organs leading to varied and potentially serious hemodynamic and physiologic consequences (Mishra, 2015). Complex congenital heart disease is commonly associated with HS. Although surgical and medical managements have advanced remarkably over the past several decades, the 1-year mortality rate of neonates with HS and complex congenital heart disease remains high, ranging from 50% up to 85% (Kim, 2011).

Extracardiac findings in HS include the lungs and bronchial tree, liver, spleen, pancreas, gall bladder, and gut. These malformations lead to potential pathophysiologic consequences, such as inability to fight infection, ciliary clearance problems, platelet dysfunction, biliary atresia, and gut ischemia (Alsoufi, 2016; Lampl, 2009). Although typically overshadowed by the challenging cardiac anomalies, malrotation has been reported in as high as 90% of neonates with HS (Landisch, 2015; Salavitabar, 2015). Malrotation refers to abnormal rotation and fixation of the intestines in the abdominal cavity occurring during embryologic development. Many neonates with malrotation will remain asymptomatic, whereas others may be symptomatic with bilious emesis or associated with other anomalies such as midgut volvulus (Morris, 2016).

The general incidence of HS is 1 per 100,000 to 200,000 live births (Mishra, 2015). The incidence of malrotation in the general population is estimated by the Centers for Disease Control at 3.9 per 10,000 live births but is seen in 40% to 90% of neonates with HS (Graziano, 2015). The combination of cardiac anomalies and malrotation carries a high mortality leading to much debate among experts regarding the appropriate management strategies.

Here, a case report highlighting the short- and long-term outcomes of a neonate born with HS and the associated cardiac and intestinal complications is presented.

Case report

A full-term (39 and 2/7 week) male with prenatally diagnosed complex congenital heart disease and HS was born after planned induction of labor at a tertiary care center. A postnatal echocardiogram confirmed the diagnosis of an unbalanced atrioventricular canal with a left-sided dominant single ventricle and pulmonary atresia. Umbilical lines were placed by the neonatology team, and a prostaglandin infusion was initiated to secure ductal-dependent blood flow. However, he developed progressive hypoxemia despite adequate pulmonary blood flow and was taken for a cardiac

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computerized tomography scan, which diagnosed obstructed pulmonary venous return, a true cardiac surgical emergency. He was then immediately taken to the operating room and underwent reimplantation of his pulmonary veins and placement of a 3.5-mm modified Blalock-Taussig shunt. His initial postoperative cardiac course was unremarkable, and he maintained hemodynamic stability. Before initiating enteral feedings on postoperative day 6, he underwent screening for extracardiac findings in HS. These included an abdominal ultrasound and upper gastrointestinal (UGI) series with small bowel follow through. The abdominal ultrasound was limited as cardiac lines, tubes, and dressings overlaid most of the field. Findings from the ultrasound included a midline liver and asplenia. The UGI (Figure 1C, D) demonstrated the duodenum coursing anteriorly and then posteriorly with the duodenojejunal junction in a low position, findings consistent with malrotation. Thin duodenal loops were also noted, suggestive of a midgut volvulus. Because of these findings, he was taken to the operating room where direct visualization verified midgut volvulus. He underwent surgical repair with Ladd's procedure, which involves division of the mesenteric or Ladd's bands and placement of the large intestines to the left side of the abdominal cavity and small intestines to the right (Graziano, 2015). He was extubated 4 days after his surgery and underwent successful feeding advancement. He had a prolonged hospitalization and rehabilitation needs because of other comorbidities not associated with HS and eventually transitioned to home around 7 months of life.

At 13 months of age, he presented to the emergency department with multiple episodes of bilious emesis and decreased bowel movements. An abdominal radiograph was obtained (Figure 2), which demonstrated large dilated loops of bowel, suggesting small bowel obstruction (SBO). Because of his clinical presentation and the abdominal film, the pediatric surgery team proceeded with exploratory laparotomy and which confirmed extensive abdominal adhesions causing SBO. He then underwent adhesiolysis and detorsion of bowel. After an appropriate course of gut rest, he was resumed on his home enteral feeding regimen without further complication and discharged home 9 days after his presentation in the emergency department.

Discussion

The definitive diagnostic gold standard for malrotation is an UGI series. In normal anatomy, the dudodenojejunal junction will be seen at the level of the ligament of Treitz to the left side of the spine (Sizemore, 2008). An intestinal rotational disorder is diagnosed when the duodenojejunal junction is not seen at that location (Morris, 2016). The sensitivity in diagnosing malrotation in UGI is 96%, enforcing its prominent diagnostic role (Carroll, 2016).

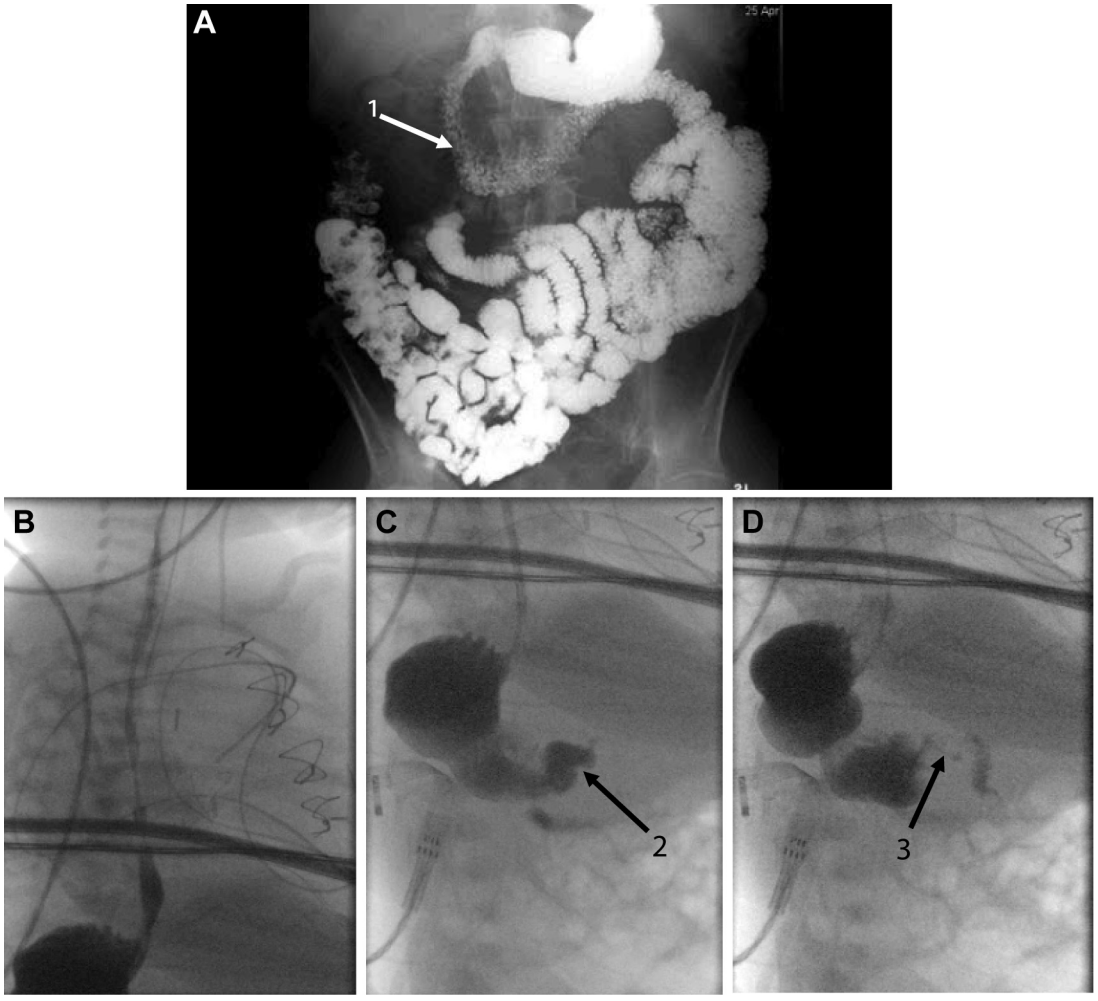

Figure 1. (A) UGI with small bowel follow through demonstrating normal anatomy. Note the left-sided stomach and C-loop of the duodenum (1) extending from the pylorus to the ligament of Treitz. (B) UGI demonstrating right-sided stomach consistent with heterotaxy. (C, D) UGI demonstrating telescoping of bowel (2) with malrotation and concern for midgut volvulus (3). UGI = Upper gastrointestinal.