Concurrent dextrogastria, reverse midgut rotation and intestinal atresia in a neonate

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ABSTRACT
A case of dextrogastria associated with multiple jejunal atresia and inverse rotation of the bowel in a neonate is presented. The management of this rare condition is highlighted with a review of the literature.

KEY WORDS: Dextrogastria, intestinal atresia, malrotation

INTRODUCTION
Neonatal intestinal obstruction is a relatively common anomaly in pediatric surgical practice. The common etiologic factors in our region are anorectal malformations, Hirschsprung’s disease and jejunoileal atresias.[1-3] Situs inversus occurs in approximately 1 in 8,500 people; however, the incidence of situs inversus abdominus (SIA) varies between 1 in 4,000 and 1 in 20,000 live births and may present a diagnostic and management challenge to the clinician and radiologist.[4] Situs ambiguous or heterotaxy implies a disordered organ arrangement in the chest or abdomen, whereas complete situs inversus implies that the organs of the chest and abdomen are arranged in opposite positions (the heart is on the right (dextrocardia) as is the two-lobed lung, while the liver, spleen and three-lobed lung are on the left). We report a case of neonatal intestinal obstruction from multiple intestinal atresias and the inverse rotation of midgut in a patient with dextrogastria and a review of recent literature.

CASE HISTORY
Y.T. presented at 22 h of life with bilious vomiting, associated with progressive abdominal distension. He was at 33 weeks gestation. Mother was 36 years of age and para 5⁺0 (3 alive). There was history of polyhydramnios during pregnancy. There was no family history of congenital anomalies. During presentation, he was an active neonate, pink in room air, anicteric and afebrile. The fontanelles were patent and normotensive. He was tachypneic, but the lung fields were clear. Heart sounds were normal. Abdomen was grossly distended with visible bowel loops and hyperactive bowel sounds. There was no evidence of peritonitis. The anus was normally sited and patent. Other systems were essentially normal. Parents were pauper and could not afford required investigations and prescribed drugs.

Packed cell volume was 54% and electrolytes and urea were normal. Abdominal X-ray [Figure 1] showed centrally placed dilated bowel loops with multiple air-fluid levels. The liver shadow was displaced to the left. Gastric fundus gas was absent in the left hypochondrium and there was no gas in the rectum. The radiologist had an impression of small intestinal obstruction due to atresia; dextrogastria was not observed and reported by them.

The infant was stabilized and the clinical status was optimized. At exploratory laparotomy [Figures 2 and 3], the stomach was located on the right and grossly dilated up to the proximal jejunum (20 cm from the...
duodenojejunal junction). The right lobe of liver was in the left hypochondrium. Spleen was absent. There were multiple jejunal atresia (type IV) and reverse rotation of the midgut; the transverse colon was behind the duodenum and the root of the mesentery. Post-atretic part of the bowel was stenosed but patent. The atretic parts of the bowel and about 8 cm of the distal part of bulbous jejunum were resected. End-to-side jejunoileal anastomosis was also performed. The immediate postoperative period was uneventful. Patient passed greenish mucoid stool on the fifth postoperative day and normal stool from the sixth day. However, he developed a high-grade fever and deep cholestatic jaundice (conjugated bilirubin, 45% of total) on the seventh day. Fever was continuous and jaundice worsened; however, there was no evidence of peritonitis. The infant still passed stool 2-3 times daily. Blood culture yielded the heavy growth of *Klebsiella* spp., which was resistant to the cefotaxime hitherto used, but sensitive to ceftazidime. Despite the change in antibiotics, the condition of the infant deteriorated and he died on the tenth postoperative day. Post mortem showed clean peritoneal cavity with intact anastomotic site.

**DISCUSSION**

Situs anomalies are rare conditions and can present with diagnostic and management challenges. There have been reports on the overlapping spectrum of situs anomalies isolating organs or encompassing both the thoracic and abdominal viscera. From our literature, the occurrence of dextrogastria, multiple jejunal atresia and inverse rotation of the intestine with levocardia in a neonate have not been reported in Africa, although there are reports of isolated intestinal malrotation in children of Nigeria. Budhiraja reported the fourth case of isolated levocardia associated with SIA and malrotation with volvulus. Earlier reports have shown occurrence of isolated levocardia with SIA and another report has shown that dextrogastria can also occur in isolation.

Embryologically, the mechanism responsible for the normal rotation of the loops of the heart and stomach to the left is not fully understood. The rotational direction of the stomach is owing to the forces exerted on it and the mesentery by the adjacent organs. Genetic and familial theories have been adduced for the etiology; however, mostly, isolated and accidental events are reported. The presence of multiple intestinal atresias could be explained by a vascular insult from the associated reverse rotation of the midgut in this patient.

SIA has a spectrum of presentation, which could be overlapping, and the intestinal obstruction is considered to be common. The occurrence of accompanying intestinal atresia that is observed in this patient is very rare and has led to early presentation. To the uninitiated, situs anomalies present a diagnostic challenge and can be confusing for the experienced practitioner, particularly in the face of intestinal obstruction. Plain and contrast enhanced X-rays and abdominal ultrasound can assist in preoperative diagnosis. Computerized tomography and radionuclear studies may enhance the diagnosis of the spectrum of anomalies where these facilities are available. The gross intestinal obstruction did not give room for a contrast GI series in this patient and dextrogastria was not suspected in this patient on the plain film.

Malrotation of the bowel is frequently observed in heterotaxy syndrome, which could be an asplenic or polysplenic syndrome. An asplenic heterotaxy syndrome leads to an immune deficiency state and the newborn surgical patients are further susceptible to overwhelming nosocomial infection, as observed in this case.

In conclusion, although the combination of dextrogastria, intestinal atresia and malrotation are rare and poses a
diagnostic challenge, the careful evaluation of patients may assist in facing this. Early presentation and appropriate surgical management may offer a chance of survival. Efforts should be made for proper aseptic procedures and nursing care of these patients. The early isolation of offending organisms and proper selection of appropriate antibiotics would reduce morbidity and prevent mortality.

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REFERENCES


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