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Case Report
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Congenital Segmental Duodenal Dilatation: An Unusual Cause of Upper Gastrointestinal Obstruction in a Newborn

Zulu S, Mashaba N, Milford K and Grieve AD*

Department of Pediatric Surgery, Nelson Mandela Children's Hospital, University of the Witwatersrand, South Africa

Abstract

Segmental dilatation of the intestine is an unusual malformation with a poorly understood etiology. It occurs most commonly in the ileum, followed by the colon and jejunum. Congenital Segmental Duodenal Dilatation (CSDD), defined as a localized dilatation of the duodenum without a distal obstruction, is extremely rare, with only 5 cases previously reported in the literature, with none of these originating from Africa. Herein, we describe a case of CSDD presenting to a tertiary pediatric hospital in South Africa.

Case Presentation

Baby X was born *via* Caesarean section at 31 weeks' gestation (birth weight: 1900 g) and required ventilation at birth for neonatal Respiratory Distress Syndrome (RDS). A post-intubation plain chest radiograph noted an abdominal right upper quadrant large dilated air-filled structure in the right upper quadrant (Figure 1). As background, the infant was one of a pair of twins. The sibling, with a birth weight of 2000 g, had no reported perinatal concerns.

The patient was referred to our service at a tertiary pediatric hospital for further investigation and management. A right upper quadrant mass was evident on inspection of the abdomen. This mass was difficult to define on palpation but was noted to be of soft consistency with a smooth surface. It was noted to be mobile, but examiners could not get 'above' or 'below' it. Auscultation revealed normal bowel sounds without any bruits. Generally, the abdomen was not distended, soft and non-tender. Plain anteroposterior abdominal radiography (Figure 1) demonstrated a large dilated air-filled structure in the right upper quadrant, with a relative paucity of distal gas. Pneumoperitoneum was not evident, and the tip of the nasogastric tube was in the expected position of the stomach, to the left of the spine. The heart was in the normal position. A plain lateral abdominal radiograph (Figure 2) demonstrated no proximal intestinal obstruction, and the air-filled structure could not be differentiated from the gastric bubble.

Abdominal ultrasound demonstrated a sizeable gas-filled cystic mass in the RUQ that was

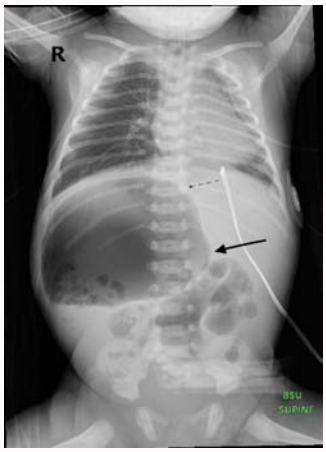

Figure 1: Antero-posterior chest and abdomen radiograph: Chest, normal positioned heart, unremarkable. Abdomen, black arrow shows a large dilated air-filled loop of bowel in the right upper quadrant. Paucity of gas in the rectum. Dotted arrow shows a nasogastric tube tip in the left upper quadrant. No features of obstruction in the abdomen.

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Correspondence:
Sphamandla Zulu, Department of Pediatric Surgery, Nelson Mandela Children's Hospital, University of the Witwatersrand, Johannesburg, South Africa,
E-mail: sphamandlazulu@hotmail.com

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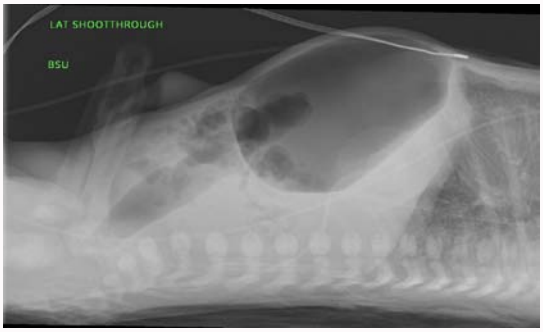

Figure 2: Lateral shoot through abdominal radiograph: No pneumoperitoneum. No nasogastric tube in the visible. Dilated air-filled loop not differentiatable from the gastric bubble.

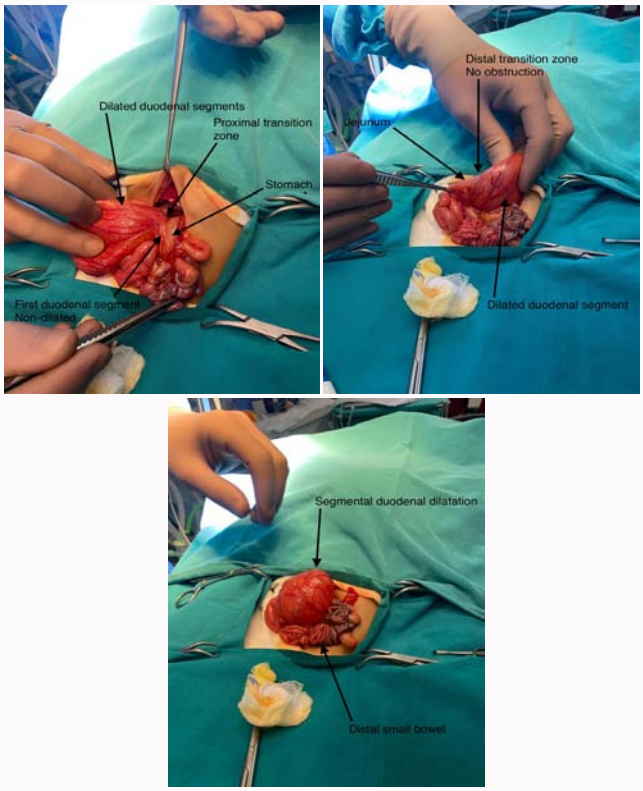

Figure 3: Intra-operative findings: Normal stomach, pylorus and first part of the duodenum. Segmental duodenal dilatation with proximal obstructive web between second part of the duodenum and the rest of the segments.

discrete from the liver. The mass could not be further described due to an extensive ring-down artifact in the RUQ. The stomach was demonstrated in the left upper quadrant and the nasogastric tube was confirmed to be within its lumen. All other solid organs were unremarkable. Differentials included an enteric duplication cyst, a peri-hepatic cyst, or a variation of duodenal atresia.

Exploratory laparotomy was performed *via* a supra-umbilical celiotomy. On identification of the duodenum the second, third and fourth segments were found to be massively dilated (Figure 3). Upon opening the cystic dilatation, the downstream intestine was easily cannulated with no evidence of mechanical obstruction. Retrograde cannulation of the stomach with a feeding tube was not possible. Further exploration revealed a web with a pinhole defect immediately distal to the Ampulla of Vater. The stomach and the

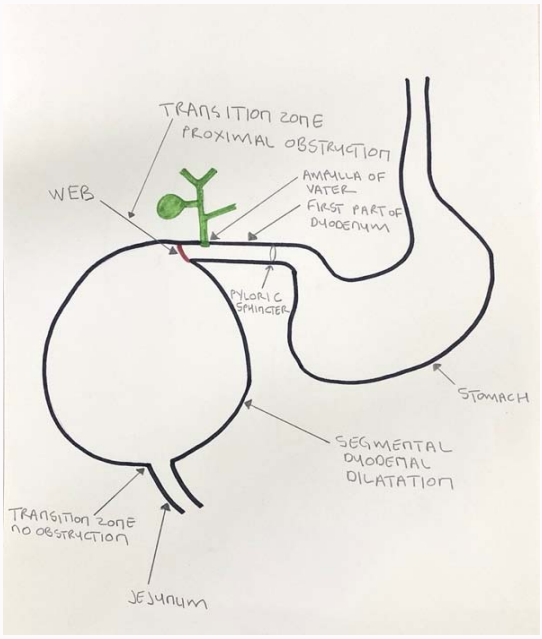

Figure 4: Sketch illustration of the intra-operative findings.

Table 1: Swenson and Rathauser criteria for congenital dilatation of the intestine.

Swenson and Rathauser criteria for congenital dilatation of the intestine.
1. Limited bowel dilatation with 3- to 4-fold increased size.
2. Abrupt transition between dilated and normal bowel.
3. No intrinsic or extrinsic barrier distal to the dilatation
4. Clinical picture of intestinal occlusion or sub-occlusion
5. Normality of neuronal plexus.
6. Complete recovery after resectioning the affected segment.

pylorus were both normal (Figure 4). Proximally, resection of the web with stricturoplasty was performed to preserve the Ampulla of Vater. A tapered enteroplasty was performed to reduce the duodenum to a normal caliber.

The patient's postoperative course was complicated by a single episode of nosocomial sepsis which responded well to targeted antibiotic therapy. There was persistent respiratory distress related to prematurity. Enteral feeding was advanced by building volume in a stepwise fashion over time. The patient was on full-volume bolus feeds at discharge from the hospital on day 19 of life. The patient was reviewed as an out-patient on day 41 of life and, at this point, was tolerating breastfeeding and gaining weight. Subsequent abdominal radiographs revealed persistent duodenal dilatation without evidence of upper gastrointestinal obstruction.

Discussion

Congenital Segmental Dilatation of the Duodenum (CSDD) is a neonatal condition without a clear etiology. To the best of our knowledge this is the 6th described report and the first in Africa. CSDD is characterized by a localized dilatation of the duodenum in the absence of distal obstruction. Diagnostic criteria for congenital dilatation of the intestine were first proposed by Swenson and Rathauser in 1959 (Table 1) [1-5].

Proposed etiologies include extrinsic intrauterine intestinal compression, vascular insult, and primitive neuromuscular

dysfunction. Katsura described the associated absence of intestinal cells of Cajal in their case which may point to an interesting pathophysiological process [6], but the etiology remains unclear. More than 50% CSDDs present during the neonatal period A cystic abdominal lesion is a non-specific finding on prenatal ultrasound [7] and postnatally ultrasound and plain abdominal radiographs may demonstrate an air-filled lesion of unclear origin [6]. The differential diagnosis for CSDD includes intestinal duplication cyst, giant neonatal choledochal cyst, hepatic cysts, and duodenal stenosis or web [8]. Invariably, CSDD is confirmed only during operative exploration as preoperative diagnosis remains a challenge due to a lack of specific radiologic signs.

The management of segmental intestinal dilatation is traditionally resection of the lesion with primary anastomosis. However, the duodenum presents unique challenges due to its critical anatomic junctures with structures entering from the pancreas and liver. These make complete resection an unfavorable option and currently, there is no consensus on the ideal surgical management for CSDD [6].

In a case reported by Katsura et al., the Ampulla of Vater was identified in the dilated segment and the duodenum was partially resected and then tapered to preserve the ampulla [6]. Our patient had a proximal stricture between D1 and D2 but no distal occlusions. We opted for a stricturoplasty of the proximal web and a tapered duodenoplasty.

Conclusion

We present the first case described in the African continent of congenital segmental dilatation of the duodenum. Careful consideration must be given to the local anatomy and surrounding critical structure when planning reconstruction. Concurrent anatomical abnormalities, such as a proximal web, need to be identified and addressed.

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