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Rare anomaly unveiled: Duodenum inversum, case report with review of literature

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Abstract

Aim of the study: To emphasize the diagnosis and management of Duodenum Inversum (DI), a rare congenital anomaly in duodenal rotation causing non-specific gastrointestinal symptoms in children.

Case Descriptions:

Case 1: A 2-day-old full-term male presented with feeding reluctance and bilious vomiting. Imaging ruled out malrotation but revealed an atypical Duodenal configuration (DI). Nasogastric feeding led to full tolerance and weight gain at 3- and 6-months follow-up.

Case 2: An 11-month-old girl had persistent non-bilious vomiting misdiagnosed as refractory gastroesophageal reflux. Imaging later confirmed DI. Nasojejunal feeding with hydrolyzed formula improved her symptoms and weight gain.

Conclusions: DI is a rare anomaly often missed due to subtle symptoms and normal duodenojejunal junction positioning. Conservative management usually suffices, though surgery may be needed for severe cases. Recognizing DI's imaging features is crucial to avoid unnecessary interventions. More studies are required to improve understanding and treatment outcomes.

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Introduction

Duodenum Inversum (DI), also known as duodenum reflexum or inverted duodenum, is a rare anomaly of duodenal rotation and fixation leading to abnormal configuration of the duodenum [4].

In this condition, the usual curvature of the third and fourth segments of the duodenum follows an atypical path, as the third part of the duodenum extends upward to the right of the midline before crossing over to the left, while the Duodenojejunal Junction (DJJ) remains in its normal position [14]. This differs from normal anatomy, in which the third portion of the duodenum travels horizontally across the midline before the fourth portion ascends to create the DJJ. This manuscript was prepared following the CARE guidelines (https://www.care-statement.org).

Aim: To shed light on this rare congenital anomaly to better understand the pathology and its management.

Cases presentations

Case 1

A 2-day-old full-term male infant who was born in good general condition but showed reluctance to feed, leading to the insertion of a Nasogastric (NG) tube to assist with feeding. The infant experienced two episodes of bilious vomiting, prompting the NG tube to be placed on free drainage, and an upper Gastrointestinal (GI) contrast series was performed to rule out midgut malrotation. The contrast study revealed that the duodenum did not exhibit the typical C-shaped configuration. Although the duodenum initially passed normally into the second part (D2), it then took a sharp cranial course behind D2 before

crossing to the left of the midline and entering the left-sided jejunal loops, while this appearance could suggest a redundant duodenum with the Duodenojejunal Junction (DJJ) positioned left of the midline (Figure 1), DI was ultimately diagnosed as a normal variant. The infant resumed NG feeding, which was well-tolerated, and achieved full feeds without further episodes of vomiting and appropriate weight gain in follow up at 3 and 6 months.

Case 2

An 11-month-old girl presented with persistent non-bilious vomiting since the age of 2 months, diagnosed with Gastro-esophageal Reflux (GOR) but showed no improvement with anti-reflux formula, thickener (Gaviscon), or Proton Pump Inhibitors (PPIs). She was failing to thrive, with a body weight of 5.6 kg. An abdominal ultrasound and upper Gastrointestinal (GI) contrast study suggested gastric outlet obstruction, prompting a referral for potential surgery. Upon admission, she was placed on intravenous fluids and started on PPIs. A repeat upper GI contrast study revealed free flow of contrast from the stomach into a normal-calibre duodenum and proximal jejunum, with normal gastric emptying; however, the duodenal configuration was abnormal, exhibiting a large loop to the right of the spine before the contrast moved to the left at the pylorus, raising concerns for possible malrotation, although no gastroesophageal reflux was noted (Figure 2). She subsequently began Masojejunal (NJ) tube feeding with a hydrolysed formula, which was well tolerated, leading to gradual achievement of full feeds and subsequent weight gain.

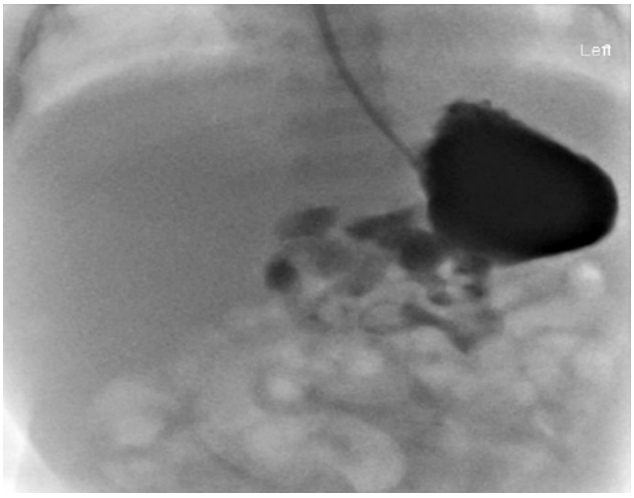

Figure 1: Illustrates the contrast medium passing from the stomach (S) to the pylorus (P), then into the first part of the duodenum (D1), followed by the second part (D2). It then flows upward through the third part (D3) in front of D2 before the fourth part (D4) crosses the midline, ultimately reaching the normal duodenojejunal junction (DJJ).

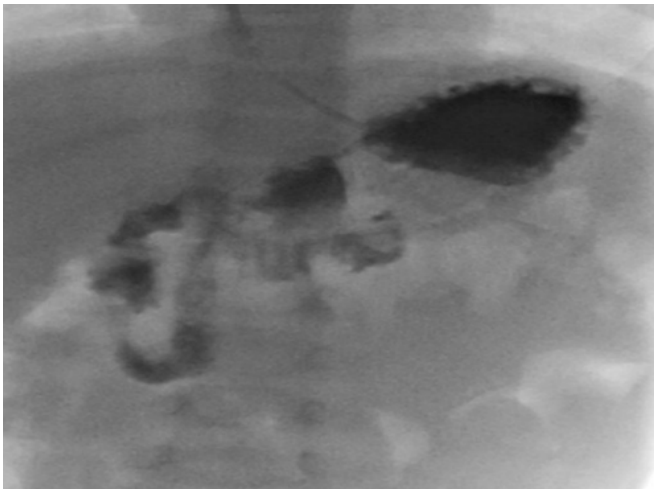

Figure 2: Illustrates the contrast medium passing from the stomach (S) to the pylorus (P), then into the first part of the duodenum (D1), followed by the second part (D2). It then flOne ows upward through the third part (D3) medial to D2 to the right of the spine before the fourth part (D4) crosses the midline, ultimately reaching the normal duodenojejunal junction (DJJ)

Review of literature

Table 1: Summarises the reported cases of paediatric DI.

	Age, Gender	Presenting symptom	Duration of Symptoms	Investigations	Management	Outcomes at 6 months follow up	Remarks
Long et al [13]	16 years, F	Nausea, vomiting, crampy abdominal pain,?? SMAS	Chronic	Upper GI series	NJ feeding, ranitidine, omeprazole	No improvement	Laparotomy + duodenum was mobilized, partial Ladd's procedure
Kim et al [10]	One month, M	Gastro-oesophageal reflux	Chronic	Upper GI series	Diagnostic laparoscopy at 1 year for intra-abdominal testes without symptoms	Improved	Preterm, T21, Abdominal testes
Dogan et al [5]	12 years, F	Nausea and vomiting	4-5 years	Upper GI series + endoscopy	Omeprazole	Improved	
Menchise et al [14]	10 years, F	Intermittent abdominal pain and bilious emesis	2 weeks	Upper GI series + endoscopy	NJ feeding, antacid	Improved	
Patel et al [15]	Two months, M	Stiffening, arching back, staring eye, bubbles from the mouth and nose	4 weeks	Upper GI series + PH monitoring	Ranitidine for 2 weeks	No improvement	Nissen's fundoplication
	Two months, M	Difficulty breathing, intermittent stiffening, gasping and spitting up	3 weeks	Upper Gi series + PH monitoring	Lansoprazole	Improved	
Sharma, et al [18]	16 years, F	Non-bloody, non-bilious vomiting	5 days	Upper Gi series + CT scan of the abdomen	NJ feeding	Improved	Occasional retching

Discussion

Duodenum Inversum (DI) is a rare gastrointestinal anomaly where the proximal segment of the duodenum folds back on itself, crossing behind the pancreas in a more cephalad position than usual, before becoming fixed in a nearly normal location giving the impression of Square Root sign (√) [4].

Despite the abnormal configuration, the DJJ often appears to be in a normal position on upper GI imaging, leading to this condition frequently being overlooked. While DI typically does not result in morbidity or mortality, it remains an important diagnostic consideration, as it can present with symptoms of proximal gastrointestinal obstruction due to the duodenal configuration [13].

It could be considered a variant of duodenal rotation or fixation, but without the associated risk of potential midgut volvulus that is typically seen with a shortened mesentery base [6].

The embryologic origin of DI is unclear and is considered a torsion anomaly; although it has been reported to cause delayed gastric emptying in some patients, it is not associated with midgut volvulus [13].

It is thought to result from the redundancy of the dorsal mesentery with a mobile duodenum and is frequently associated with anomalies in the fixation or position of the right kidney, pancreas, and transverse mesocolon, that's why it is thought to predispose to cholecystitis, pancreatitis and peptic ulcer disease [9-16].

Four types of DI were reported by Feldman and Morrison; Type 1 is characterized by a complete inversion of the duodenum with the absence of the duodenal curve, while Type 2 retains the presence of the duodenal curve. Type 3 involves a duodenal curve with significant redundancy of the duodenum, and Type 4 is defined as DI associated with malrotation. However, the practical value of this classification is limited. It is often asymptomatic, with many symptoms arising due to stasis within the duodenal loop [1,7].

The reported cases of DI in both adult and paediatric literature are limited. Table 1 provides a summary of the published paediatric cases. Patient ages ranged from a few days old to 16 years, with symptom duration varying widely. Patients with DI are frequently asymptomatic or may have non-specific symptoms included chronic nausea, vomiting (both bilious and non-bilious), abdominal pain and other Gastroesophageal Reflux (GOR) manifestations, while in adult, pancreatitis, gallbladder disease and peptic ulcer disease were reported [11,19].

It has been suggested that many symptoms linked to DI result from stasis in the duodenal loop. Patients with this abnormality show a notable reduction in gastroesophageal sphincter pressure while eating, along with increased pyloric regurgitation and decreased motility in the antrum and delayed gastric emptying; therefore, all these factors contribute to symptoms of GOR [8,17].

DI has been linked to other abnormalities, such as annular pancreas and incomplete bowel rotation [2,4,12].

Upper Gastrointestinal (GI) contrast series is the main diagnostic test which revealed the characteristic duodenal configuration and PH monitoring in those who experienced reflux symptoms. Medical management is the mainstay of therapy in absence of obstruction with Nasojejunal tube (NJ) tube feeding and antacid medications, such as H2 blockers or proton pump inhibitors to control GOR and treatment of duodenitis.

Surgical intervention was required in three cases. The first case involved a diagnostic laparoscopy performed at one year of age for mainly intra-abdominal testes, without GI symptoms but concerns about possible malrotation. This patient, who had Down syndrome with undescended testes (which necessitated laparoscopy), was found to have an anterior band over the second part of the Duodenum (D2), distinct from typical Ladd's bands. This band was divided, confirming normal positioning of the DJJ [10]. The other two cases did not respond to conservative treatment laparotomy in one case whose presentation was mimicking Superior Mesenteric Artery Syndrome (SMAS), where the duodenum was mobilized, and a partial Ladd's procedure was performed [12]. The other patient underwent a Nissen's fundoplication to manage GOR symptoms that did not respond to maximal medical therapy, which effectively resolved their issues [15].

This manuscript enriches the limited literature on duodenum Inversum in paediatrics, adding valuable case-based insights into this rare anomaly. It provides thorough case details, including specific imaging findings and successful conservative management, which can guide clinicians in identifying and managing similar cases. The well-structured content, adherence to CARE guidelines, and a relevant literature review strengthen the manuscript's clarity and reliability.

With only two cases, the report's findings may have limited generalizability and lack long-term outcome data, which could otherwise provide more comprehensive insight. The figures and embryologic explanations are brief, which reduces clarity in distinguishing DI from other similar conditions and understanding its developmental basis. Additionally, minimal discussion on differential diagnoses, pathophysiology, and comparisons of surgical versus non-surgical outcomes may limit the manuscript's practical utility for broader clinical decision-making.

Conclusion

Duodenum Inversum (DI) is a rare congenital anomaly that can present with vague gastrointestinal symptoms, often leading to delayed diagnosis. It is usually managed medically with nasojejunal feeding and acid suppression, but surgery may be needed in persistent or complicated cases. Early recognition through imaging and identifying the Square Root sign is key to appropriate management and avoiding unnecessary surgery. Long term follow up is recommended and further studies are needed to better understand the long-term outcomes and refine treatment strategies.

Declarations

Informed consent: Informed consent was obtained from the parents of the patients.

Authorship: All authors attest that they meet the current ICMJE criteria for Authorship.

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