Journal of Paediatric Surgeons of Bangladesh (2022) Vol. 13 (1 & 2)
Official Journal of the Association of Paediatric Surgeons of Bangladesh
DOI: https://doi.org/10.3329/jpsb.v.13i1.87417

Journal of Paediatric Surgeons of Bangladesh

Case Report

Neonatal Duodenal Obstruction in Situs Inversus: Surgical Implications of a Pre-Duodenal Portal Vein

Mona NJ[1]*, Alam MR[2], Ferdous KNM[3]

Abstract

Background:
Pre-duodenal portal vein (PDPV) is a rare congenital vascular anomaly resulting from abnormal embryological development of the vitelline venous system. Although often asymptomatic, PDPV may be associated with duodenal obstruction, particularly in neonates with additional congenital anomalies such as situs inversus, intestinal malrotation, and duodenal atresia.

Case Presentation:
We report a four-day-old male neonate presenting with bilious vomiting and features of proximal intestinal obstruction. Imaging revealed duodenal obstruction with situs inversusabdominus. Laparotomy demonstrated a pre-duodenal portal vein crossing the second part of the duodenum with an associated intraluminal duodenal membrane. Membrane excision and gastro-duodenostomy were performed to bypass the obstruction while avoiding injury to the anomalous portal vein. The postoperative recovery was uneventful.

Conclusion:
The coexistence of PDPV, duodenal obstruction, and situs inversus is exceedingly rare. Intraoperative recognition of anomalous vascular anatomy and tailored surgical planning are critical to prevent catastrophic vascular injury and ensure favorable outcomes.

Keywords: Pre-duodenal portal vein; Situs inversus; Duodenal obstruction; Neonate; Congenital anomaly

1. Nusrat Jahan Mona
Consultant, Department of Pediatric Surgery
Al-Manar Specialized Hospital
Dhaka, Bangladesh

2. Muhammad Rashedul Alam
Associate Professor, Division of Pediatric Surgery
Bangladesh Shishu Hospital & Institute
Dhaka, Bangladesh

3. Kazi Md Noor-ul Ferdous
Professor, Division of Pediatric Surgery
Bangladesh Shishu Hospital & Institute
Dhaka, Bangladesh

Correspondence to: Nusrat Jahan Mona
Consultant, Department of Pediatric Surgery
Al-Manar Specialized Hospital
Dhaka, Bangladesh

Introduction:

Pre-duodenal portal vein (PDPV) is an uncommon congenital anomaly in which the portal vein passes anterior to the duodenum rather than posteriorly, as a result of abnormal regression of embryonic vitelline veins [1]. PDPV itself is rarely the sole cause of duodenal obstruction but is frequently associated with other congenital abnormalities, including situs inversus, intestinal malrotation, annular pancreas, duodenal atresia, polysplenia, and congenital heart disease [2–4].

The combination of PDPV with situs inversus and duodenal obstruction is exceptionally rare, with only sporadic cases reported in the literature [3,5]. Recognition of this anomaly is essential, as inadvertent injury to the portal vein during surgery can result in catastrophic hemorrhage. We report a rare case of neonatal duodenal obstruction in association with PDPV and situs inversus and discuss its embryology, clinical significance, and surgical management.

Case Presentation

A four-day-old male neonate weighing 2 kg presented with bilious, non-projectile vomiting since birth, progressive dehydration, tachycardia, and decreased urine output. Antenatal history was unremarkable. On physical examination, the abdomen was soft and non-tender with epigastric fullness. The perineum and anal opening were normal.

Plain abdominal radiograph demonstrated features suggestive of proximal intestinal obstruction. Ultrasonography of the abdomen revealed situs inversusabdominus with a dilated duodenum.

Exploratory laparotomy revealed the liver on the left side and the stomach on the right, confirming situs inversus. The duodenum was markedly dilated up to its third part, and the duodeno-jejunal junction was located on the right side with a floating caecum. A pre-duodenal portal vein was identified crossing anterior to the second part of the duodenum. An intraluminal duodenal membrane was found and excised via enterotomy in the third part of the duodenum.

Due to the presence and position of the PDPV, duodeno-duodenostomy was not feasible. Therefore, a gastro-duodenostomy was performed to bypass the obstruction while avoiding portal vein injury and potential future compression. The postoperative course was uneventful, and the patient was discharged on the 10th postoperative day.

Discussion

Embryological Basis

The portal vein develops from paired vitelline veins during the fourth to tenth weeks of gestation. PDPV results from persistence of the anterior vitelline venous channel with regression of the posterior channel, leading to an anomalous pre-duodenal course of the portal vein [2,4].

Association with Situs Inversus & Other Anomalies

PDPV is rarely isolated and is most commonly associated with other congenital anomalies. Situs inversus and heterotaxy syndromes are frequently reported associations, reflecting the shared embryological disturbances affecting left-right axis determination [3,4]. Congenital cardiac anomalies occur in approximately 5–10% of patients with situs inversus, underscoring the importance of thorough preoperative evaluation [4].

Mechanism of Duodenal Obstruction

Although PDPV may cause extrinsic compression of the duodenum, most cases of obstruction are due to intrinsic lesions such as duodenal atresia or webs, with PDPV acting as an associated rather than causative factor [1,5]. Therefore, when PDPV is identified intraoperatively, surgeons must actively search for intrinsic causes of obstruction.

Surgical Considerations

Surgical management must be individualized. Duodeno-duodenostomy or duodeno-jejunostomy is generally preferred when anatomy permits [2]. However, in cases where the PDPV precludes safe duodenal anastomosis, gastro-duodenostomy or gastro-jejunostomy offers a viable alternative [5]. Meticulous awareness of mirror-image anatomy in situs inversus is essential to avoid vascular injury [3].

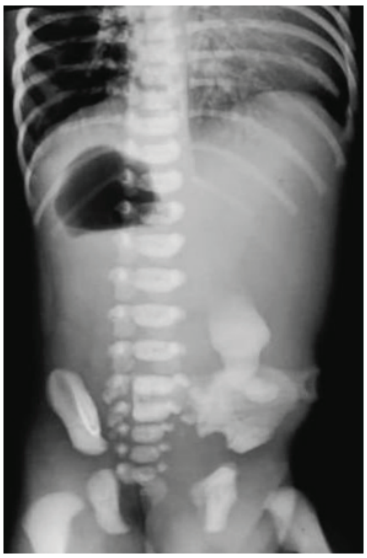

Figure 1: X-ray: showing fundic gas on the right side

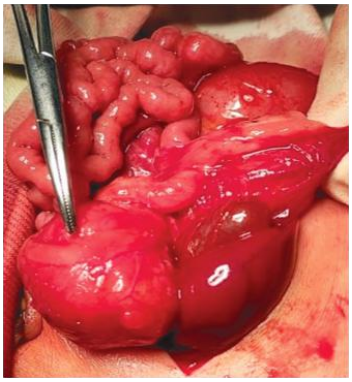

Fig: 2: Liver & gall bladder on the Left, Stomach on the Right side

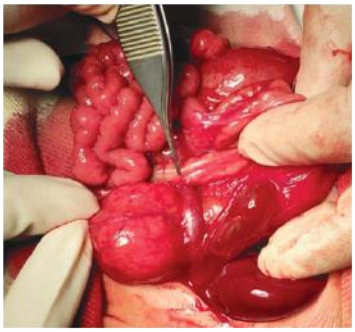

Fig: 3: Pre duodenal portal vein anterior to second part of duodenum

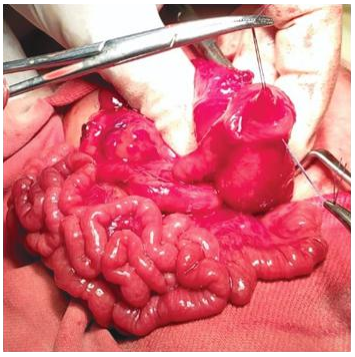

Fig: 4: Enterotomy& excision of internal membrane

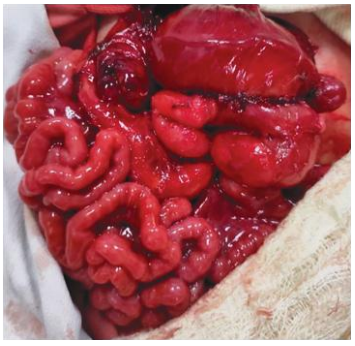

Fig: 5: Gastro-duodenostomy

Conclusion

Duodenal obstruction associated with pre-duodenal portal vein and situs inversus is exceedingly rare. Successful management requires high intraoperative vigilance, recognition of anomalous vascular anatomy, and flexibility in surgical strategy. Familiarity with mirror anatomy and embryological variations is crucial to prevent iatrogenic injury and achieve optimal outcomes.

References

1. Srivastava P, Shaikh M, Mirza B, Jaiman R, Arshad M. Preduodenal portal vein associated with duodenal obstruction of other etiology: a case series. J Neonatal Surg. 2020;5(4):54.
2. Arafa A, Nasreldin HM, Ahmed AH. Laparoscopic duodenoduodenostomy in cases of neonatal duodenal obstruction owing to preduodenal portal vein. Egypt J Surg. 2022;41(3).
3. Ziv Y, Lombrozo R, Dintsman M. Preduodenal portal vein with situs inversus and duodenal atresia. AustPaediatr J. 1986;22:69–70.
4. Yi SQ, Tanaka S, Tanaka A, et al. An extremely rare inversion of preduodenal portal vein and common bile duct associated with multiple malformations. AnatEmbryol (Berl). 2004;208(2):87–96.
5. Nakagawa Y, Sumida W, Makita S, et al. Laparoscopic gastrojejunostomy for duodenal atresia with situs inversus and preduodenal portal vein: a report of two cases. Front Pediatr. 2023;11:1220393.