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CASE REPORT

Situs Inversus Totalis with Type I Duodenal Atresia and Preduodenal Portal Vein in a Preterm Neonate: A Case Report

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Abstract

Background: Situs inversus totalis is a rare congenital laterality disorder characterised by mirror-image transposition of the thoracic and abdominal viscera. It may coexist with congenital cardiac, gastrointestinal and vascular anomalies, including duodenal atresia and preduodenal portal veinboth independently recognised causes of neonatal duodenal obstruction. Simultaneous occurrence of all three anomalies is exceptionally rare and poses distinct diagnostic and operative challenges.

Case Presentation: A male neonate born at 35 weeks' gestation, weighing 2.1 kg, presented on day 3 of life with bilious vomiting, feeding intolerance and failure to pass meconium. Examination revealed a scaphoid abdomen and a right-sided cardiac apex. Chest and abdominal radiography demonstrated dextrocardia and a right-sided gastric bubble; abdominal ultrasonography subsequently confirmed mirror-image visceral arrangement, consistent with situs inversus totalis. Echocardiography identified a secundum atrial septal defect and a small patent ductus arteriosus. Following resuscitation and preoperative optimisation, the infant underwent exploratory laparotomy through a left upper transverse incision, which confirmed complete situs inversus totalis and revealed a dilated proximal duodenum obstructed by an intraluminal mucosal web (Type I duodenal atresia). A preduodenal portal vein was also identified, crossing anterior to the first part of the duodenum immediately adjacent to the site of intrinsic obstruction.

Intervention and Outcome: The duodenal web was excised through a longitudinal duodenotomy. Because the preduodenal portal vein lay close to the obstructed segment, a diamond-shaped duodenoduodenostomy (Kimura technique) was performed to achieve a wide, tension-free bypass while preserving the anomalous vein. The postoperative course was uneventful; enteral feeding commenced on postoperative day 6 and was advanced without complication. The infant was discharged on postoperative day 10 in good clinical condition.

Conclusion: In neonates with situs abnormalities and congenital duodenal obstruction, surgeons should anticipate coexisting vascular anomalies. Careful intraoperative identification of a preduodenal portal vein is essential to avoid vascular injury and diamond-shaped duodenoduodenostomy can safely address combined intrinsic and extrinsic obstruction while preserving the anomalous vessel.

Keywords: Situs Inversus, Intestinal Atresia, Duodenal Obstruction, Portal Vein, Infant, Newborn, Infant, Premature.

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1. Introduction

Situs inversus totalis (SIT) is a rare congenital laterality disorder characterised by complete mirror-image transposition of the thoracic and abdominal viscera, with a reported incidence of approximately 1 in 8,000 to 1 in 25,000 live births (1,2). Although isolated SIT is generally compatible with a normal lifespan, its clinical significance lies in its recognised association with congenital cardiovascular, gastrointestinal, hepatobiliary and splenic anomalies, which can substantially influence diagnostic evaluation, operative planning, and perioperative management (1–3).Normal left–right asymmetry is established during early embryogenesis through coordinated ciliary motion at the embryonic node and tightly regulated molecular signalling involving genes such as *NODAL*, *LEFTY*, *PITX2*, and *ZIC3*. Disruption of these pathways produces a spectrum of laterality defects, ranging from isolated dextrocardia to complete situs inversus and heterotaxy syndromes (2,4). Abnormal ciliary function also underlies primary ciliary dyskinesia (PCD), in which approximately 50% of affected patients demonstrate situs inversus as part of Kartagener syndrome, reflecting the shared dependence of nodal ciliary flow on normal ciliary structure and motility (4,5).Congenital duodenal atresia is among the most common causes of neonatal intestinal obstruction, occurring in approximately 1 in 5,000–10,000 live births (6). It has classically been attributed to failure of recanalisation of the solid embryonic duodenal cord during the sixth to tenth weeks of gestation; however, this model does not fully account for the variability of atresia phenotypes observed clinically and current evidence increasingly implicates disrupted genetic signalling pathways alongside the well-established association between duodenal atresia and trisomy 21 (6,7). Type I duodenal atresia, the most frequent subtype, is characterised by an intraluminal mucosal diaphragm or web with preservation of bowel continuity (7). Affected neonates typically present within the first days of life with bilious vomiting and feeding intolerance, while antenatal ultrasonography may demonstrate polyhydramnios and the characteristic double-bubble sign (6,8).Preduodenal portal vein (PDPV) is an uncommon developmental anomaly in which the portal vein courses anterior to the duodenum, rather than posteriorly, owing to abnormal persistence and regression of the embryonic vitelline venous system (9,10). Although PDPV is frequently an incidental intraoperative finding, it may cause clinically significant extrinsic duodenal compression and has been reported in association with intestinal malrotation, annular pancreas, biliary atresia, polysplenia, congenital heart disease and laterality disorders, including situs inversus (10–12). Its principal clinical hazard, however, is not obstruction but the risk of catastrophic portal vein injury if the anomaly is not recognised during surgery.The coexistence of SIT, duodenal atresia, and PDPV is exceptionally rare, with only isolated case reports available in the literature (11–14). This combination presents unique diagnostic and technical challenges: mirror-image anatomy can complicate radiological interpretation and operative orientation, while the close anatomical relationship between the obstructing lesion and the anomalous portal vein demands meticulous surgical planning to achieve adequate decompression without vascular injury.We report a preterm neonate with situs inversus totalis associated with Type I duodenal atresia, preduodenal portal vein, atrial septal defect and patent ductus arteriosus. This case adds to a small body of literature describing this specific anomaly cluster and illustrates the importance of comprehensive preoperative assessment, careful intraoperative recognition of associated vascular anomalies and individualised surgical decision-making in neonates with complex congenital malformations.

2. Case Presentation

2.1 Patient Demographics and Antenatal/Birth History

A male neonate was born at 35 weeks' gestation by spontaneous vaginal delivery to a healthy 26-year-old primigravida following an uncomplicated pregnancy. There was no history of maternal diabetes mellitus, teratogenic drug exposure or consanguinity and no family history of congenital anomalies was elicited. Antenatal ultrasonography was not done, and polyhydramnios was not reported. Birth weight was 2.1 kg.

2.2 Presenting Complaint

The infant developed repeated episodes of non-projectile bilious vomiting following the initiation of feeds, progressively increasing in frequency with associated feeding intolerance. He had not passed meconium since birth. He was referred to our neonatal surgical unit on day 3 of life for further evaluation.

2.3 Clinical Findings

On admission, the neonate was mildly dehydrated but haemodynamically stable. Physical examination revealed a scaphoid, non-distended abdomen without

visible bowel loops or palpable masses with normal bowel sounds. Cardiovascular examination revealed the cardiac apex and heart sounds predominantly on the right hemithorax, raising suspicion of dextrocardia. No dysmorphic features suggestive of trisomy 21 or other syndromic disorders were identified.

2.4 Laboratory Investigations

Baseline complete blood count and serum electrolytes were within acceptable limits following correction of dehydration.

2.5 Imaging

Chest radiography demonstrated dextrocardia; abdominal radiography showed a right-sided gastric bubble with paucity of distal bowel gas, suggesting congenital proximal intestinal obstruction in the context of situs inversus totalis (Figure 1). Abdominal ultrasonography confirmed mirror-image visceral orientation with the liver occupying predominantly the left upper quadrant and the stomach and spleen located on the right. Transthoracic echocardiography confirmed dextrocardia and demonstrated a secundum atrial septal defect and a small patent ductus arteriosus, without evidence of complex cyanotic congenital heart disease.

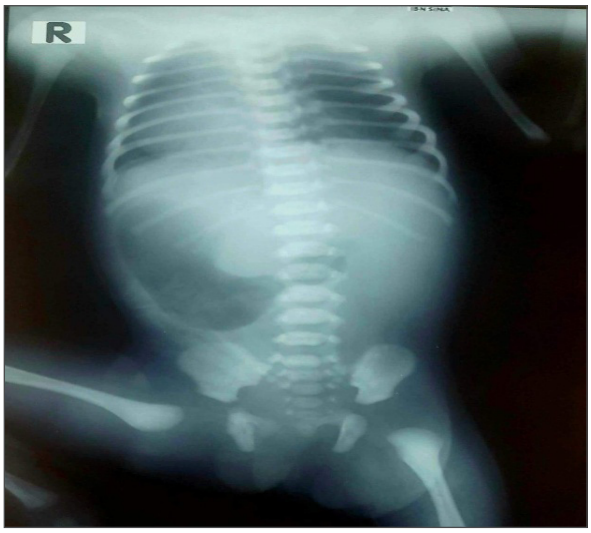

Figure 1. *Chest and abdominal radiograph showing dextrocardia, right-sided gastric bubble and mirror-image anatomy.*

2.6 Differential Diagnosis

The combination of bilious vomiting, delayed meconium passage and radiologically confirmed situs inversus was considered consistent with congenital duodenal obstruction, with intestinal malrotation with volvulus, annular pancreas, jejunal atresia, and Hirschsprung disease considered as alternative or coexisting causes of neonatal obstruction. The absence of a "double-bubble" transition to a completely gasless abdomen argued against complete distal atresia, and the clinical stability of the infant made volvulus with vascular compromise less likely, although this could not be excluded on imaging alone and was ultimately excluded at laparotomy.

2.7 Final Diagnosis

Congenital duodenal obstruction associated with situs inversus totalis, confirmed intraoperatively as Type I duodenal atresia with a coexisting preduodenal portal vein.

2.8 Multidisciplinary Preoperative Management

Initial management included strict nil-by-mouth status, nasogastric decompression, intravenous fluid resuscitation, electrolyte correction and broad-spectrum antibiotics per institutional neonatal surgical protocol. Following multidisciplinary evaluation by the neonatal, paediatric surgical, anaesthetic and cardiology teams and after adequate preoperative optimisation the infant underwent exploratory laparotomy.

2.9 Operative Findings

A left upper transverse abdominal incision was selected to facilitate exposure of the mirror-image anatomy. Complete situs inversus totalis was confirmed intraoperatively: the liver was situated predominantly on the left and the stomach and spleen on the right. The proximal duodenum was markedly dilated, with a collapsed distal small bowel. Exploration identified a Type I duodenal atresia caused by an obstructing intraluminal mucosal diaphragm (duodenal web). An unexpected finding was a preduodenal portal vein crossing anterior to the first part of the duodenum, immediately adjacent to the site of intrinsic obstruction (Figure 2).

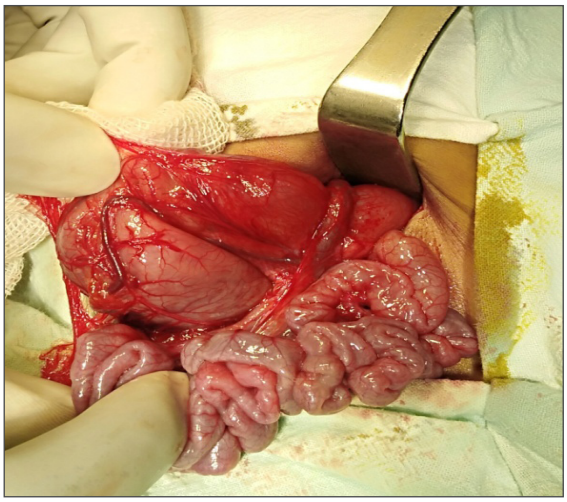

Figure 2. *Intraoperative photograph demonstrating the preduodenal portal vein crossing anterior to the proximal duodenum.*

2.10 Surgical Procedure and Intraoperative Challenges

A longitudinal duodenotomy was performed over the dilated proximal duodenum, and the obstructing mucosal diaphragm was completely excised (Figure 3). Because the preduodenal portal vein crossed directly anterior to the obstructed segment web excision alone risked leaving residual extrinsic narrowing once the duodenum decompressed. To address this, a diamond-shaped duodenoduodenostomy (Kimura technique) was constructed to bypass the compressed segment while preserving the anomalous vein, avoiding any direct dissection or traction on the vessel. The anastomosis was tension-free and well vascularised, and no intraoperative complications occurred.

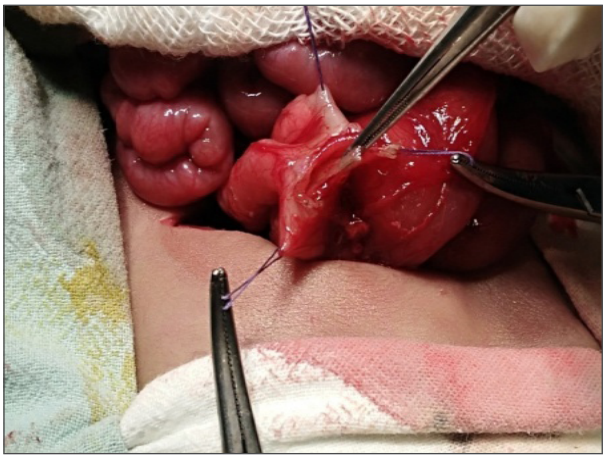

Figure 3. *Completed diamond-shaped duodenoduodenostomy (Kimura technique)*.

2.11 Postoperative Management

The infant was managed in the neonatal intensive care unit with nasogastric decompression, intravenous fluids, parenteral nutritional support and prophylactic antibiotics. Nasogastric drainage gradually decreased and enteral feeding commenced on postoperative day 6 following return of bowel function. Feeding was progressively advanced without vomiting or abdominal distension. The infant passed stools normally, tolerated full enteral feeds and demonstrated satisfactory weight gain and was discharged home on postoperative day 10 in good clinical condition.

2.12 Histopathology

Not performed

2.13 Follow-up and Current Status

At 3-month follow-up, the infant remained clinically well, with normal feeding tolerance, appropriate weight gain and no clinical evidence of recurrent obstruction or postoperative complication.

3. Discussion

3.1 Epidemiology and Clinical Significance

Situs inversus totalis (SIT) is a rare congenital laterality disorder, with a reported incidence of 1 in 8,000–25,000 live births (1,2). Although isolated SIT is compatible with a normal lifespan, its clinical importance lies in its association with congenital cardiovascular, gastrointestinal, and hepatobiliary anomalies (2,3). The present case is notable for

combining SIT with Type I duodenal atresia, a preduodenal portal vein (PDPV), atrial septal defect and patent ductus arteriosus in a single preterm neonate a constellation reported only sporadically in the literature, almost exclusively as isolated case reports rather than case series (11–14,19,20).

3.2 Embryology and Pathophysiology

SIT arises from disordered left–right axis determination during gastrulation, driven by abnormal nodal ciliary flow and downstream signalling through *NODAL*, *LEFTY*, *PITX2*, and *ZIC3* (2). Congenital duodenal atresia, by contrast, has classically been attributed to failure of recanalisation of the solid embryonic duodenal cord the Tandler hypothesis though this model does not fully explain the range of atresia phenotypes observed clinically and current evidence increasingly implicates disrupted genetic signalling pathways alongside the well-established association with trisomy 21 (6,7). These two processes are temporally and mechanistically distinct and no unifying genetic defect has been demonstrated to link them; their coexistence, here and in prior reports is most plausibly explained by a shared embryonic vulnerability to disruption across multiple independent developmental pathways during early gestation rather than a single common cause (7–10).PDPV adds a further layer of embryological complexity, arising from abnormal persistence and regression of the paired embryonic vitelline veins such that the portal vein comes to lie anterior, rather than posterior, to the duodenum (9,10). It may remain asymptomatic throughout life or produce varying degrees of extrinsic duodenal compression, and has been reported in association with malrotation, annular pancreas, biliary atresia, polysplenia, and laterality disorders reflecting a shared window of vulnerability in vitelline venous and gut-rotation development (9–12).

3.3 Comparison with Published Literature

Most existing reports of SIT with congenital duodenal obstruction describe duodenal atresia or annular pancreas as isolated coexisting lesions without a PDPV (13,14,15,16). Reports describing PDPV coexisting with situs anomalies more often feature malrotation as the associated intestinal pathology rather than duodenal atresia (11,12,20). The combination reported here intrinsic duodenal atresia and PDPV occurring together in the setting of SIThas correspondingly few directly comparable published cases (11,12), which is what makes this report a genuine rather than incremental addition to the literature.

3.4 Diagnostic Challenges and Differential Diagnosis

Diagnosis in this case was complicated by the mirror-image anatomy itself. Antenatal ultrasonography did not detect SIT, duodenal atresia or PDPVconsistent with reports that laterality anomalies and PDPV are frequentlymissedantenatallyandmoreoftendiagnosed postnatally on plain radiography, echocardiography or at laparotomy (6,8). Reversed anatomical landmarks can disorient interpretation of the double-bubble sign and gastric bubble position which appear on the right rather than the left in SIT; recognising dextrocardia clinically and radiologically was therefore an important early step that appropriately redirected image interpretation in this case. As outlined in the Case Presentation, malrotation with volvulus, annular pancreas, jejunal atresia and Hirschsprung disease were considered as alternative or coexisting causes of obstruction; confirmation of the diagnosis ultimately required direct laparotomy rather than imaging alone underscoring that imaging in SIT should be regarded as suggestive rather than definitive.

3.5 Operative Decision-Making and Alternative Surgical Approaches

Operative planning was shaped principally by the unexpected finding of the PDPV directly overlying the site of intrinsic obstruction. This relationship created a dual technical problem: the web required excision to relieve true mechanical obstruction, while the overlying vein precluded any reconstruction placing tension or traction on the anterior duodenal wall given the risk of catastrophic venous injury haemorrhage or portal vein thrombosis (9,17). Web excision alone risked residual extrinsic narrowing once the duodenum decompressed; conversely, bypass without web excision would have left the intrinsic obstruction untreated recognition of both lesions was essential, consistent with prior reports of coexisting intrinsic and extrinsic obstruction (11,12).

Diamond-shaped duodenoduodenostomy, first described by Kimura and colleagues in 1977, remains the standard reconstruction for congenital duodenal obstruction, providing a wide, tension-free anastomosis with good long-term functional outcomes (17,18). However, where a PDPV lies close to the natural anastomotic site, some authors have instead selected duodenojejunostomy or in recently reported cases, laparoscopic gastrojejunostomy, specifically to route the anastomosis away from the anomalous vessel (19). Laparoscopic duodenoduodenostomy has also been shown in systematic review to achieve

outcomes equivalent to open repair in experienced hands (19). In this patient, an open diamond-shaped duodenoduodenostomy was chosen given prematuritylow birth weight, and the need for direct tactile control around the PDPV; this provided the widest tension-free lumen while keeping dissection clear of the veinconsistent with the rationale in comparable published cases (11,20). The principal advantage of this approach is avoidance of vascular injury with preserved physiological continuity; its main disadvantage relative to duodenojejunostomy or gastrojejunostomy is that it requires more extensive proximal duodenal mobilisation in immediate proximity to the anomalous vessel, demanding meticulous technique.

3.6 Postoperative outcome and prognosis

Enteral feeding commenced on postoperative day 6, with discharge on day 10 without complication. Congenital cardiac anomalies occur more frequently in SIT than in the general population and should be actively investigated before surgery (3,13); the atrial septal defect and patent ductus arteriosus in this patient were haemodynamically insignificant and did not affect perioperative management, consistent with the generally favourable prognosis reported when associated cardiac lesions are simple rather than complex (3,13). With a single patient and short reported follow-up this favourable early course should be interpreted cautiously rather than generalised.

3.7 Lessons learned and clinical implications

Mirror-image anatomy should prompt active evaluation for other coexisting congenital anomalies both radiologically before operation and directly at laparotomy. PDPV should specifically be anticipated whenever congenital duodenal obstruction is encountered alongside situs abnormalities, malrotation, annular pancreas or polysplenia, since an unrecognised PDPV carries a real risk of catastrophic intraoperative vascular injury (9,17,20). Where intrinsic and extrinsic obstruction coexist, both lesions must be addressed, and the anastomotic technique individualised favouring, where anatomically feasible a wide bypass that avoids direct manipulation of the anomalous vessel with duodenojejunostomy or gastrojejunostomy reserved as alternatives when anatomy precludes a safe duodenoduodenal anastomosis (19,20,21).

Limitations

This report describes a single patient with a short-reported follow-up interval precluding assessment of longer-term outcomes such as anastomotic stricture or duodenogastric reflux. No genetic or ciliary function testing was performed to evaluate for an underlying ciliopathy. Associated-anomaly screening was limited to echocardiography without systematic documentation of renal, vertebral, or biliary tract assessment. As with any single case report, causal or mechanistic conclusions linking the three anomalies cannot be drawn.

4. Conclusion

Situs inversus totalis associated with Type I duodenal atresia and a preduodenal portal vein is an exceptionally rare combination that presents distinct diagnostic and operative challenges. This case underscores the importance of thorough preoperative evaluation and meticulous intraoperative exploration to identify associated vascular anomalies capable of altering surgical strategy. Recognition of a preduodenal portal vein before definitive reconstruction is essential to prevent inadvertent vascular injury during correction of congenital duodenal obstruction. In this patient excision of the duodenal web followed by diamond-shaped duodenoduodenostomy achieved effective relief of both intrinsic and extrinsic obstruction while preserving portal venous integrity with a favourable early postoperative course. Longer-term follow-up and further reported cases will be needed to confirm whether this outcome is generalisable.

Declarations

Ethics Approval and Consent to Participate

This case report describes a single patient and did not require formal institutional ethics committee approval consistent with institutional policy for anonymised single-patient case reports.

Consent for Publication

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying clinical images (Figures 1–3), including acknowledgement that, despite efforts to remove identifying features anonymity cannot be absolutely guaranteed. A copy of the consent form is available for review by the Editor-in-Chief upon request.

Conflict of Interest

The authors declare no conflict of interest.

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Author Contributions

Author 1, 2, 3 performed the surgery and conceived the report. Author 1 collected clinical data and drafted the manuscript. Author 3 supervised the case and critically revised the manuscript. All authors read and approved the final manuscript."

Data Availability Statement

Data supporting this case report are included within the article. Further clinical details are available from the corresponding author upon reasonable request subject to patient confidentiality restrictions.

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