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RARE DELAYED PRESENTATION OF DUODENAL ATRESIA WITH ANNULAR PANCREAS: A CASE REPORT

DIVYA PRIYA S S[1]
JAI DURAIRAJ P[2]
KEERTHIKA M[3]
[1]POSTGRADUATE, DEPARTMENT OF GENERAL SURGERY, SAVEETHA MEDICAL COLLEGE AND HOSPITAL, CHENNAI
[2]PROFESSOR, SAVEETHA MEDICAL COLLEGE AND HOSPITAL, CHENNAI

Abstract

Background: Duodenal atresia is a congenital gastrointestinal obstruction caused by failure of fetal duodenal recanalization. Although it commonly presents in the neonatal period with bilious vomiting, atypical delayed presentations are increasingly recognized, particularly in Type 1 atresia (duodenal web).

Case Presentation: We describe an 11-month-old infant with a 6-month history of recurrent non-bilious vomiting, intolerance to solids, and failure to thrive. Imaging suggested proximal duodenal obstruction, while intraoperative findings revealed Type 1 duodenal atresia due to a duodenal web, compounded by annular pancreas. Surgical correction was achieved via Kimura's diamond-shaped duodenoduodenostomy.

Outcome: The infant had an uneventful recovery, with resolution of vomiting, tolerance of regular diet, and significant catch-up growth.

Conclusion: This case highlights the importance of maintaining suspicion for congenital duodenal obstruction beyond the neonatal period. Annular pancreas in conjunction with a duodenal web adds diagnostic and operative complexity, but timely recognition and Kimura's duodenoduodenostomy provide excellent outcomes.

Background

Congenital duodenal obstruction is a leading cause of neonatal intestinal blockage, with an incidence of roughly 1 in 5,000–10,000 live births [1,2]. The embryologic basis is failed duodenal recanalisation during weeks 8–10 of gestation, producing intrinsic obstruction (atresia/stenosis/web) or, less commonly, extrinsic compression [3]. Duodenal atresia is classically divided into three subtypes—Type 1 (web/diaphragm with intact muscularis), Type 2 (fibrous cord), and Type 3 (complete separation)—with Type 1 accounting for the great majority [1,4].

Typical neonatal presentation is early bilious vomiting and the radiographic "double-bubble" sign due to gastric and proximal duodenal distension [5,6]. However, fenestrated duodenal webs may permit partial transit, leading to delayed or insidious symptoms—intermittent non-bilious vomiting, early satiety, poor weight gain, and failure to thrive—well beyond the newborn period [7–9].

Duodenal atresia frequently coexists with congenital anomalies, notably trisomy 21, cardiac defects, malrotation, and annular pancreas; the latter forms a pancreatic ring around the second part of the duodenum and can add an extrinsic component to intrinsic obstruction [1,2,10,11]. Annular pancreas itself is uncommon, but presents earlier and more severely in children than in adults, especially when combined with intrinsic lesions such as a web or stenosis [6,11].

Diagnosis in delayed cases can be challenging. Contrast studies may reveal the "windsock" deformity—ballooning of a fenestrated web into the distal lumen—while upper GI endoscopy can directly visualise a mucosal diaphragm and exclude acquired strictures [7,12].

Surgery remains definitive. Kimura's diamond-shaped duodenoduodenostomy restores physiologic flow with low leak rates and excellent functional outcomes, and is widely regarded as the procedure of choice for intrinsic duodenal obstruction [8,13]. Alternatives (web excision/duodenotomy or gastrojejunostomy) are reserved for select anatomy or low-birth-weight contexts but carry trade-offs in long-term physiology and complication profiles [4,14].

Case Presentation

An 11-month-old male infant was referred to our surgical unit with a 6-month history of recurrent non-bilious vomiting, predominantly after introduction of solid foods. The vomiting was intermittent but

progressive, occasionally projectile, and associated with feeding intolerance. Parents reported failure to thrive, poor weight gain, irritability, and recurrent hospital visits for dehydration.

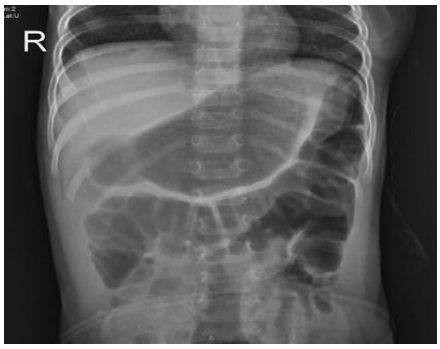


Figure 1. Radiograph of abdomen showing single bubble sign

On examination, the child was malnourished, weighing 5.2 kg (<3rd centile for age), with visible wasting. Mild epigastric fullness was noted, more prominent post-prandially, but without tenderness or palpable mass. Bowel sounds were normal, and systemic examination was unremarkable.

Investigations:

Plain abdominal X-ray: Markedly dilated gastric shadow with paucity of distal bowel gas.

Ultrasound abdomen: Normal, with no evidence of malrotation or volvulus.

Barium meal follow-through: Demonstrated a grossly distended stomach and proximal duodenum with delayed passage of contrast into distal bowel—suggestive of incomplete duodenal obstruction.

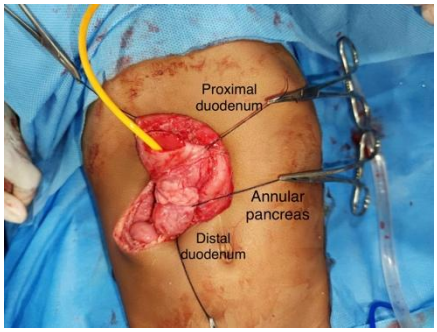
Given the persistence of symptoms, poor growth, and radiological evidence of obstruction, **s**urgical exploration was indicated.

Intraoperative findings:

A right supraumbilical transverse incision was made. Intraoperatively, the second part of the duodenum was encircled by pancreatic tissue—confirming annular pancreas. On introducing a Foley's catheter into the proximal duodenum and inflating the bulb, a duodenal web distal to the ampulla was identified (Type 1 duodenal atresia).

Figure 2. Intraoperative picture of duodenum

A Kimura's diamond-shaped duodenoduodenostomy was performed, creating a side-to-side bypass of the obstructed segment while preserving duodenal continuity and avoiding ampullary injury.



Postoperative course: The infant had an uneventful recovery. Oral feeding was initiated gradually, and the child tolerated solids without vomiting. At 3-month follow-up, **catch-up growth** was evident (weight gain of 2.1 kg), with resolution of symptoms.

Differential Diagnosis

In an infant with **chronic non-bilious vomiting and growth failure**, the experienced clinician must adopt a broad differential, systematically excluding both acquired and congenital causes:

- **Gastroesophageal reflux disease (GERD):** Common in infancy, but typically associated with regurgitation, irritability, and esophagitis rather than progressive gastric outlet obstruction. Our patient's barium study demonstrated obstruction rather than reflux.
- **Hypertrophic pyloric stenosis:** Presents with non-bilious projectile vomiting, but occurs between 2–8 weeks of age. At 11 months, pyloric stenosis is highly unlikely, and ultrasound excluded it.
- **Malrotation with intermittent volvulus:** A critical differential in persistent vomiting. Doppler ultrasound excluded malrotation in this case.

- **Peptic stricture / acquired duodenal stenosis:** Possible in older children, but rare in infancy, and imaging pointed toward congenital obstruction.
- **Congenital duodenal obstruction (web/atresia):** Strongly suggested by radiographs and contrast study. In this case, partial passage of contrast confirmed incomplete obstruction, fitting Type 1 duodenal web.

Thus, the diagnosis of **delayed presentation of Type 1 duodenal atresia with associated annular pancreas** was established intraoperatively

Outcome and Follow-Up

Postoperative recovery was uneventful. Oral feeding was resumed gradually; the infant tolerated solids, gained weight steadily, and was discharged in good condition. At follow-up, growth velocity had improved significantly, with complete resolution of vomiting.

DISCUSSION

Although most cases of duodenal atresia are identified neonatally, delayed presentation is well documented in fenestrated Type 1 webs, where partial patency allows liquids to pass while solids precipitate symptoms—explaining recurrent vomiting, feeding intolerance, and growth faltering over months to years [7–9]. In a landmark series, children operated as late as 72 months commonly had failure to thrive, anaemia, and reflux-related complications at diagnosis [8]. Individual reports echo this heterogeneity—for example, a 6-year-old with cyclical vomiting ultimately proven to have a duodenal web on contrast study and endoscopy [9].

Annular pancreas compounds the pathophysiology by adding extrinsic compression to intrinsic narrowing, often intensifying the obstructive physiology and complicating exposure at operation [6,11]. This interplay mandates high clinical suspicion in infants and toddlers with persistent vomiting and poor weight gain, even when early imaging is non-diagnostic [1,10,11].

Imaging is pivotal but nuanced: while the double-bubble suggests complete atresia in neonates, the windsock sign is more typical of a fenestrated web in delayed presentations; endoscopy, when available, increases diagnostic confidence and helps exclude acquired peptic strictures [7,12].

Operative strategy has evolved toward Kimura's diamond duodenoduodenostomy given its physiologic streamlining, wide anastomosis, and low morbidity, with consistently excellent long-term outcomes and catch-up growth in contemporary series [5,13]. Longitudinal data show high survival and restoration of growth trajectories after timely repair, whereas diagnostic delay is associated with malnutrition, reflux oesophagitis, strictures, and aspiration-related morbidity [5,8]. Alternatives—web excision or bypass procedures—have roles in select scenarios but are generally less favoured due to technical variability and non-physiologic flow, respectively [4,14].

Our case, an 11-month-old with a Type 1 web plus annular pancreas, fits the delayed-presentation phenotype: tolerance of liquids, failure with solids, progressive growth faltering, and ultimately definitive correction with Kimura's duodenoduodenostomy—followed by rapid symptom resolution and catch-up growth [5,8,13]. The key message is pragmatic: in infants with persistent vomiting and failure to thrive, congenital duodenal obstruction must remain on the differential—even beyond the neonatal window—and early operative correction is both safe and transformative when anatomy is clarified [1,6,8,13,14].

Learning Points

- Type 1 duodenal atresia (web) may present beyond the neonatal period with vague symptoms such as recurrent vomiting and poor weight gain.
- Annular pancreas may coexist with duodenal webs, amplifying obstruction severity and complicating operative management.
- Barium studies and endoscopy remain pivotal when delayed congenital obstruction is suspected.
- Kimura's duodenoduodenostomy offers reliable physiological repair with excellent outcomes, even in complex cases.
- A high index of suspicion is essential to avoid prolonged morbidity in infants with unexplained vomiting and failure to thrive.

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