

Co-Occurrence of Esophageal Ring and Peri-Pyloric Pancreatic Rest in a Child with Down Syndrome: A Case Report

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Abstract

Background: Down syndrome (DS) is commonly associated with gastrointestinal (GI) anomalies—esophageal abnormalities, though rare, present significant concerns, especially in children with DS. **Case Presentation:** A 3-year-old male with DS presented with persistent vomiting. Diagnostic investigations revealed a foreign body lodged above a lower esophageal stricture, likely an esophageal ring, as seen in barium studies. Subsequent upper endoscopy confirmed these findings and incidentally identified a peri-pyloric pancreatic rest. The child underwent successful endoscopic intervention, leading to symptom resolution and clinical improvement. **Conclusion:** This case highlights the rare co-occurrence of an esophageal ring and peri-pyloric pancreatic rest in a child with DS. Early diagnosis and tailored management are crucial for alleviating symptoms and enhancing the quality of life in DS patients with gastrointestinal anomalies.

Keywords

Down Syndrome, Esophageal Ring, Pancreatic Rest, Gastrointestinal Anomalies, Endoscopic Intervention

1. Introduction

Down syndrome (DS), caused by trisomy 21, is the most common chromosomal abnormality worldwide [1]. It encompasses a spectrum of medical complexities, including gastrointestinal anomalies that occur in about 6% of the patients with DS [2]. Among these, esophageal abnormalities pose a rare yet significant con-

cern, particularly in children with DS, affecting their feeding patterns and overall well-being [3]. While esophageal webs or rings represent an infrequent manifestation within this population, their coexistence with other gastrointestinal anomalies remains scarcely documented.

In this report, we describe a 3-year-old child diagnosed with DS who exhibited a rare co-occurrence of an esophageal ring and peri-pyloric pancreatic rest. To the best of the author's knowledge, this represents the first documented case of Down syndrome presenting with this combined occurrence. Our findings underscore the imperative consideration of esophageal anomalies in the diagnostic spectrum of children with DS, especially those exhibiting persistent vomiting and feeding difficulties.

This case also stresses the importance of a thorough evaluation in children with DS to identify potential co-existing abnormalities. Timely diagnosis and tailored management strategies are pivotal in relieving symptoms and enhancing the quality of life for these children, emphasizing the significance of early intervention and targeted care.

2. Case Presentation

A 3-year-old male diagnosed with Down syndrome presented to the emergency room with persistent vomiting. He was born to non-consanguineous parents and exhibited dysmorphic features at birth, with karyotyping confirming trisomy 21. Initial cardiac echocardiogram revealed a minor ventricular septal defect (VSD), while routine thyroid screenings yielded the expected results. The child had no respiratory issues in the neonatal period and had received appropriate vaccinations.

Upon arrival at the emergency department, the child displayed no signs of respiratory distress or airway obstruction, maintaining stable vitals. There was no salivation, drooling, stridor, or wheeze then. Upon initial assessment, the patient's anthropometric measurements were documented as follows: a weight of 13 kg, falling within the 25-50th percentile range on the DS growth chart; a height of 88 cm, positioned at the 25th percentile; and a head circumference (HC) of 47 cm, corresponding to the 50th percentile on relevant growth charts.

Initial chest X-rays were inconclusive, prompting a cautious approach given the child's association with DS and a concurrent cardiac anomaly. Consequently, a non-invasive approach involving upper gastrointestinal (GI) series using barium was pursued with parental consent. The test revealed a foreign body, a constriction in the lower esophagus, proximal dilation, and an esophageal ring or web (**Figure 1**). Differential diagnoses, including foreign body ingestion, stricture, esophageal web, achalasia, general gastrointestinal symptoms in Down syndrome, duodenal atresia or stenosis, celiac disease, hypotonia, and neurological disorders or metabolic diseases were systematically evaluated and ruled out through a comprehensive assessment of patient history, thorough physical examination, and pertinent laboratory investigations. The rationale for excluding diagnoses other than a foreign body in the esophagus: stricture is typically associated with severe esophagitis

History of previous surgery like TEF; the ring's appearance and absence of significant esophageal damage argue against a stricture; the esophageal web is located higher in the esophagus (post-cricoid region) and commonly associated with Plummer-Vinson syndrome, which does not align with this presentation achalasia is characterized by esophageal dilation and absence of peristalsis, which were evident in imaging studies; which were not seen here; duodenal atresia or stenosis are congenital anomalies typically present neonatally with bilious vomiting and the classic "double bubble" sign on imaging, not seen here; celiac disease is common in down syndrome but primarily presents with diarrhea, malabsorption, and positive serological markers (negative here); Hypotonia in Down syndrome contributes to dysphagia but does not explain anatomical abnormalities like the esophageal ring or pancreatic rest; neurological disorders or metabolic diseases were ruled out due to regular developmental evaluation (for this population) and absence of systemic signs like seizures or metabolic decomposition.



Figure 1. Barium swallow showing significant focal stenosis at the distal esophagus associated with proximal dilatation with filling defect likely impacted foreign body and delayed clearance.

Diagnostic and therapeutic upper GI endoscopy was deemed necessary to remove the foreign body to confirm the diagnosis of the esophageal ring and assess the severity of the obstruction considering the absence of prior surgeries or corrosive/battery ingestion history. Given the complexity of the child's cardiac history, an upper GI endoscopy was considered following consultations with cardiology and neurology services and with the family's consent. Subsequently, on Feb 13, 2023, the child underwent an esophagogastroduodenoscopy (EGD) under general anesthesia. The patient was intubated, ventilated, and kept in his left lateral position; a regular scope was used (outer diameter 9.8 cm). The esophagus was dilated with food particles and fungal white patches; as we didn't have a brush

to take a sample, we tried to flush it, which was flushable. We advanced until we reached 20 cm from the upper incisors, where we faced a foreign body (Beads) that was successfully retrieved by Roth net; the foreign body was lying on a concentric ring-like stricture that we couldn't bypass with regular scope due to difficulty negotiating the stricture, a controlled radial expansion (CRE) balloon was used for dilation, sequentially increasing the diameter from 10 mm to 11 mm, and then to 12 mm. Which was easily dilated, and mild mucosal tearing noticed. Afterward, we were able to bypass the stricture and enter the Stomach. The Stomach exhibited inflammation, erosions, and gastritis. An incidental finding of a peri-pyloric pancreatic rest was also noted (**Figure 2**). The duodenum was normal.



Figure 2. Endoscopic appearances of the esophagus and Stomach. (A) Bead-like foreign body (FB) in the lower part of the esophagus; (B) Concentric esophageal ring; (C) FB retrieved by a Roth net; (D) CRE Balloon dilatation; and (E) and (F) peri-pyloric pancreatic rest with evidence of gastritis and erosions.

The child's clinical condition notably improved post-intervention, demonstrating considerable tolerance to feeding. Subsequent follow-ups highlighted a favorable response and resolution of the presenting symptoms. After three weeks (short-term follow-up), the patient's weight was 13 Kg, height was 88 cm, and HC was 47 cm. After three months, the weight was 16 kg, height was 89cm, and HC was 47 cm. After 1 year (long-term follow-up), the weight was 19 kg, height was 90 cm, and HC was 50 cm—Indicating a favorable outcome post intervention.

3. Discussion

Gastrointestinal (GI) anomalies are the second most common associated anomalies in individuals with DS, spanning both pediatric and adult populations. These include esophageal, duodenal, and small bowel structural defects, encompassing atresia, stenosis, annular pancreas, imperforate anus, and Hirschsprung disease [2] [4]. Antenatal scans commonly detect such anomalies, aiding in early intervention planning [5]. However, our patient's delayed presentation at the age of three years hints at the incomplete nature of the obstruction, challenging antenatal detection methodologies in some instances. Esophageal rings are concentric, smooth, thin extensions of normal mucosal tissues located in the distal esophagus and can cause partial obstruction. They are categorized into three types based on location and epithelial lining.

Type A rings are located above the esophagogastric junction (EGJ), are composed of muscular tissue (muscularis propria), and are lined by squamous epithelium. They are usually asymptomatic but can cause functional narrowing during swallowing. Type B or Schatzki rings (at the junction, mixed epithelium) located at the squamocolumnar junction, typically at the EGJ, composed of mucosa and submucosa without muscularis propria; the upper surface is covered by squamous epithelium, and the lower surface by columnar epithelium. It's considered the most common type of mucosal tissue, associated with intermittent dysphagia, especially to solids, termed "steakhouse syndrome" and Type C (distal, columnar epithelium) is rare and located below the EGJ, represent diaphragmatic impressions and are of little clinical significance [6]. While the direct association between DS and esophageal rings remains less understood, various factors might contribute to their formation, including gastroesophageal reflux disease (GERD) or hiatal hernias often associated with this population [7].

Esophageal rings are thought to arise from congenital anomalies, chronic inflammation, or gastroesophageal reflux disease (GERD). Schatzki rings are commonly linked to GERD, as reflux can induce mucosal injury and healing, leading to scarring and ring formation. The role of motility disorders and genetic predispositions in their development remains unclear [8].

The successful management of the esophageal ring through endoscopic dilation (e.g., Savary dilatation or balloon dilation), as observed in this case, underscores the effectiveness of timely intervention in relieving symptoms and improving the child's quality of life. Our patient's case epitomizes the challenges posed by esophageal ring-associated complications in DS. The diagnostic journey, involving upper GI series and subsequent endoscopic confirmation, unveiled an esophageal ring alongside a constrictive lesion. The following intervention, featuring endoscopic dilation, relieved the obstruction and addressed an incidental finding of peri-pyloric pancreatic rest, alleviating symptoms and promoting substantial clinical improvement.

Furthermore, our case sheds light on an incidental discovery—a peri-pyloric pancreatic rest—within the context of DS. This anomaly is characterized by pan-

creatic tissue developing without typical anatomical or vascular connections and termed a pancreatic rest, aberrant pancreas, heterotopic pancreas, or ectopic pancreas [9]. Pancreatic rest occurrence is not uncommon and can be detected in approximately 1% of pediatric upper gastrointestinal endoscopies. This finding is notably more prevalent in patients with esophageal atresia and may also be associated with trisomy 21, as observed in our patient [10].

Pancreatic rests (ectopic pancreas) are congenital anomalies where pancreatic tissue is outside its usual anatomical location. Peri-pyloric rests are found near the pylorus and can sometimes mimic tumors or cause gastric outlet obstruction—thought to arise from improper migration of pancreatic tissue during embryogenesis. Ectopic pancreatic tissue can retain exocrine and endocrine functions, potentially causing inflammation, cyst formation, or obstruction [11].

Pancreatic rests have been noted in association with Down syndrome, possibly due to increased congenital anomalies affecting gastrointestinal development. GERD and hypotonia may exacerbate symptoms in these patients. It is often asymptomatic but may cause epigastric pain, nausea, vomiting, gastric outlet obstruction, or bleeding [12].

Endoscopy may show a submucosal lesion, often near the pylorus. Imaging (CT, MRI, or endoscopic ultrasound) helps confirm the diagnosis. A biopsy is rarely performed but may show pancreatic acini and ducts [13].

Asymptomatic lesions usually require no intervention. Symptomatic lesions (e.g., causing obstruction or bleeding) may require resection. Depending on the lesion's size and location, endoscopic resection or laparoscopic surgery can be used. Special Considerations in Down Syndrome: Due to increased susceptibility to GERD, gastric motility issues, and potential feeding difficulties, patients with Down syndrome require careful evaluation. Multidisciplinary management, including gastroenterologists and pediatricians, is crucial [14].

Both esophageal rings and peri-pyloric pancreatic rests are essential considerations in patients with Down syndrome. While esophageal rings are primarily managed through dilation and GERD treatment, pancreatic rests may require surgical intervention if symptomatic. Early diagnosis and targeted management can improve outcomes in these conditions.

The importance of comprehensive evaluations in children with Down syndrome cannot be overstated. Thorough assessments are essential for identifying potential coexisting abnormalities, which influence management strategies and prognostic outcomes. Tailored interventions and early diagnosis remain pivotal in improving the overall prognosis for these individuals.

Future research should determine the prevalence and associations between esophageal and pancreatic anomalies in DS individuals. Long-term follow-ups are imperative to grasp these concurrent anomalies' clinical implications and develop improved diagnostic and therapeutic strategies. Unraveling shared genetic factors or developmental pathways could offer valuable insights, facilitating tailored man-

agement strategies for DS patients with similar gastrointestinal anomalies in the future.

In conclusion, our case presents a rare yet intriguing co-occurrence of an esophageal ring and peri-pyloric pancreatic rest in a child with Down syndrome. This case emphasizes the significance of vigilance and comprehensive evaluations in diagnosing and managing gastrointestinal anomalies in individuals with DS, mainly when presenting with symptoms such as persistent vomiting and feeding difficulties. The successful intervention addressed the esophageal ring and incidental peri-pyloric pancreatic rest, highlighting the importance of early, comprehensive management strategies for improved clinical outcomes.

Consent for Publication

Written informed consent was obtained from the patients and their parents to publish this case series according to the journal's patient consent policy.

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Availability of Data and Material

The data used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Conflicts of Interest

The authors have no conflicts of interest to declare. All co-authors have seen and agreed with the manuscript's contents, and there is no financial interest to report. We certify that the submission is original work and is not under review at any other journal.

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