

The Uncommon Diagnosis of Windsock Deformity for a Common Presentation

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We report a two-year-old Indonesian female who presented with vomiting and weight loss for more than one month and was ultimately diagnosed with duodenal obstruction due to a windsock deformity. This is a rare and intrinsic congenital anomaly of the duodenum. The diagnosis as well as immediate and conclusive surgical management is discussed.

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Vomiting is common in children. The causes range from simple to complex and possibly fatal. The clinician would face multiple common and benign cases related to overfeeding, gastroesophageal reflux and gastrointestinal infections¹. However, diagnostic difficulty could occur when the underlying cause is an intracranial pathology, a metabolic abnormality or a structural anomaly of the pylorus or duodenum.

Intestinal obstruction is a congenital phenomenon which occasionally presents in association with other congenital anomalies, and could be either intrinsic or extrinsic². Extrinsic causes include malrotation, Ladd's bands, and annular pancreas, while intrinsic include duodenal atresia, stenosis or web².

The aim of this presentation is to report a rare and uncommon presentation of a delayed duodenal obstruction caused by a windsock deformity.

THE CASE

A two-year-old Indonesian female presented with a history of recurrent vomiting during the last 6 weeks. The vomiting had increased in frequency over the previous five days, and the child was unable to tolerate any oral diet. The vomiting was described as large in amount and bilious and was associated with 2 kg loss of weight in two weeks. She had not responded to antiemetic medication and fluid rehydration therapy. History revealed recent travel to Indonesia where her symptoms began, but no further relevant details were relevant. The child had an uneventful peripartum period followed by a normal delivery; no vomiting or any other medical symptoms in the neonatal period were revealed.

On examination, the child was in a mild state of shock with fluid deficit of five percent causing hypoactivity and mild tachycardia. Blood pressure, temperature and capillary refill time were within normal limits. Her weight for age was four standard deviations below the normal limits. Inspection of the abdomen showed active and visible peristaltic movement; however, palpation revealed no organomegaly or tenderness.

Bolus intravenous fluids were administered to resuscitate the patient, and an initial blood gas analysis showed a mixed respiratory and metabolic alkalosis, hypokalemia and mild hyponatremia. Abdominal plain film radiograph showed a markedly distended stomach with an air-fluid level; however, air was seen tracking all the way to the rectum and no distended bowel loops were noted, see figures 1 and 2. An abdominal ultrasound showed a dilated common bile duct with minimal intrahepatic biliary dilatation.

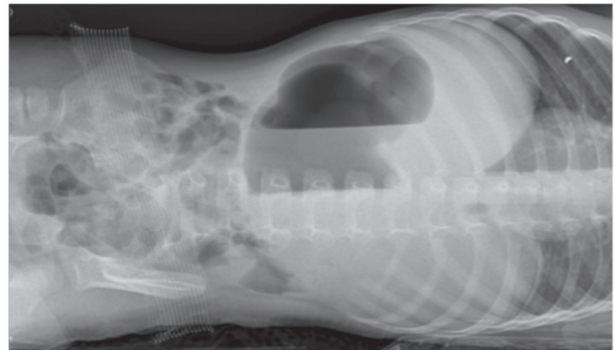


Figure 1: Preoperative Lateral Abdominal X-ray Showing Distended Stomach

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Figure 2: Abdominal Plain X-ray Showing a Distended Stomach

Antibiotic was administered and a nasogastric tube inserted; it was regularly aspirated and produced bilious fluid. The child remained nil by mouth, and total parenteral nutrition was provided. A Gastrografin study on the third day of admission confirmed the diagnosis of a duodenal obstruction by showing the stomach, duodenal bulb, and second part of the duodenum to be significantly dilated with markedly narrowed third part, see figure 3.

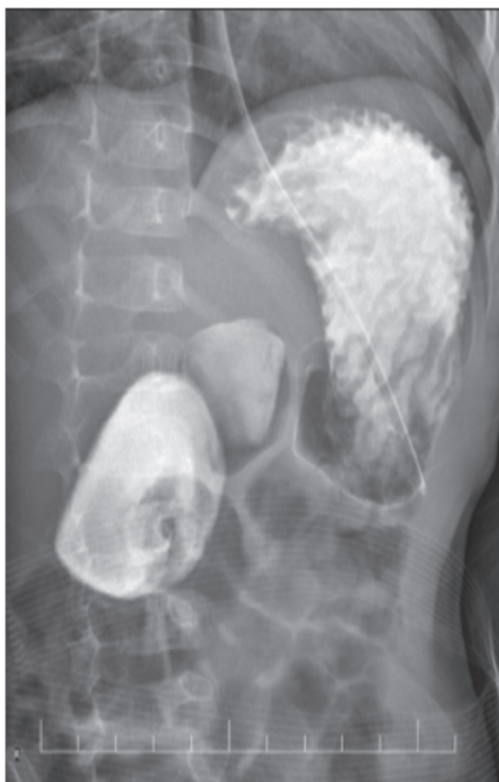


Figure 3: Gastrografin Study Showing the Stomach, Duodenal Bulb and Second Part of the Duodenum, Significantly Dilated with Markedly Narrowed Third Part

Laparotomy with excision of the duodenal membrane was completed successfully on the sixth day of admission. The lateral wall of the second part of the duodenum was opened and a membrane occluding the entire lumen was found to be causing near-total obstruction. The membrane was excised with caution not to injure the neighboring ampulla, and the duodenum was closed transversely with interrupted sutures.

Milk feeds were started and on the fifth day and the child was discharged home. At one month postoperative follow-up, the patient was doing well with a clean and well-healed surgical wound and an 800g increase in weight. Four months postoperatively, she had improved markedly, and her weight was recorded between the second and ninth growth centiles.

DISCUSSION

This report aims to highlight the management of a delayed presentation of duodenal obstruction. Melek et al reported less than 100 cases of windsock deformity until 2008, and a further search revealed only nine more cases between 2009 and 2013³.

Prevalence of an intrinsic pathology, such as duodenal atresia, stenosis or web, occurs in 1/6000 children and stenosis rarely occurs in 10%^{1,2}. A windsock deformity is usually an isolated anomaly, most frequently located between the first and second part of the duodenum in 85%-90% of cases^{1,3}. Only 10% of cases were combined with congenital disorders such as Down’s syndrome, Situs Inversus, congenital heart disease, annular pancreas or intestinal malrotation^{4,5}. Such pathology presenting beyond the age of infancy has rarely been reported⁶. The eldest patient with a duodenal web found in the medical literature was 82 years old at the time of presentation; the authors speculated that her use of oral potassium supplements was the cause rather than congenital abnormality⁶.

The duodenal web could be imperforate, also known as atresia, intraluminal imperforate popularly termed as the windsock deformity. The web could be perforated with central or eccentric apertures, see figure 3. A windsock deformity is formed due to flawed embryological development in the seventh week, when there is “failure of normal recanalization of the duodenum after epithelial cell occlusion of the foregut lumen. Over time, the web or diaphragm passively elongates, as a result of continual peristalsis against it”⁷.

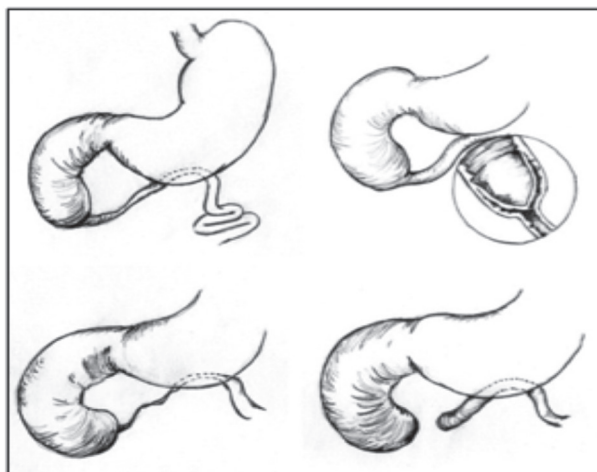


Figure 3: Diagram of Windsock Deformity Variants

A windsock deformity is a radiological diagnosis, so termed due to the pathognomonic sign seen in an upper gastrointestinal radiological series. A windsock appearance is created by obstruction to the contrast flow by the web or membrane resulting in a "contrast-filled sac seen entirely within the lumen of the duodenum and a visualized narrow radiolucent line"⁷. The characteristic appearance is created due to constant peristalsis causing the membrane to prolapse distally⁴. This study is diagnostic when the barium retention is noted for at least 6 hours³. Other imagings such as abdominal ultrasound and flexible upper gastrointestinal endoscopy have been used as a diagnostic tools⁴. Antenatal, small bowel gas observed distal to a 'double bubble,' would raise the suspicion of duodenal stenosis, web or malrotation with midgut volvulus².

The majority of cases are asymptomatic. The symptoms include nausea, vomiting, abdominal pain and weight loss; however, it is noteworthy to mention that the deformity has caused recurrent pancreatitis and gastrointestinal bleeding in some patients^{3,4}. The pain could be in the epigastric, right-upper or in umbilical area and is worsened by eating; vomiting or certain postures could bring relief³. While a case of atresia would usually present in the immediate post-natal period, an incomplete duodenal obstruction presents later in childhood, as highlighted in our case². It should be considered in any child with a history of failure to thrive and bile-stained vomiting.

The initial step in management is fluid resuscitation. Definitive surgical management includes the classic open approach, which was introduced by Kimura in 1977 as a technique to create a side-to-side duodenoduodenostomy through a diamond shaped incision². He later developed it further to form the double layer anastomosis method without the need for a stoma allowing for faster recovery time and return to normal functionality².

Recently, laparoscopic approach has proved to be a safe and effective option^{8,9}. However, such a minimally invasive approach prevents proper exploration of all bowel segments and also requires appropriate laparoscopic surgical skills^{8,9}. Endoscopic intervention using laser therapy, sphincterotome incision, high-frequency-wave snare/cutter, and hot biopsy forceps with dilatation have all been documented but the experience is limited, and their use requires expertise¹⁰.

CONCLUSION

The windsock deformity is a rare cause of small intestinal obstruction and usually presents in the early neonatal period. This case emphasized the importance of considering congenital pathologies as the cause of vomiting and failure to thrive even in older children.

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