

Case Report

Windsock Deformity in Jejunal Obstruction

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ABSTRACT

Introduction: Windsock deformity (WD) is a rare case with incidence of 1 in 6000 to 10000 live births. It is commonly seen in duodenum but rare in jejunum.

Presentation of case: A 2.5yr female child came with complain of abdominal distention, vomiting and incomplete defecation. Patient looked malnourished. Barium study showed dilated stomach and duodenum upto proximal jejunum.

Discussion: WD is a rare occurrence causing intestinal obstruction. More common in second part of duodenum. In jejunum and ileum it is uncommon. 'Windsock' sign on X-ray is pathognomonic. Surgery is the treatment of choice.

Conclusion: WD is rare but early diagnosis and treatment can save the patient. Diagnosis can be done preoperatively. Even intra op diagnosis can be followed with successful surgery. Surgical resection is the treatment widely used.

Keywords: Windsock deformity, jejunal web, intestinal obstruction, surgical resection.

INTRODUCTION

Windsock deformity (WD) is a rare cause for intestinal obstruction. More often it occurs in the 2nd part of duodenum. Rarely it may occur in jejunoileal segment.

Nelson first described the entity in 1947. [1] These intraluminal diverticula are believed to arise from an improper luminal recanalization of the foregut in the 7th week of embryogenesis. A residual tissue diaphragm may span the entire circumference of the duodenum and only allow passage of enteric contents through fenestrations. [2] The presence of these rare diverticulae can be

seen on upper gastrointestinal series and MDCT scans demonstrating the pathognomonic "windsock" sign.

PRESENTATION OF CASE

A 2.5 yr female child came to the outpatient department with complain of abdominal distention, frequent vomiting and incomplete defecation since 1 month. Patient appeared malnourished. Barium study showed dilated stomach and duodenum upto proximal part of jejunum with little barium shadow seen beyond jejunum upto the rectum [Figure 1]. Intra op bowel showed dilatation upto

10cm from DJ flexure [Figure 2]. On opening the stricture at jejunum showed a mucosal web with a small central opening [Figure 3]. Mucosal web resection

done and stricturoplasty completed [Figure4]. 3 month follow up of the patient showed improvement in the weight and no post op complications.



Figure 1. Barium study showing Windsock Deformity.



Figure 2. Jejunum Obstruction.

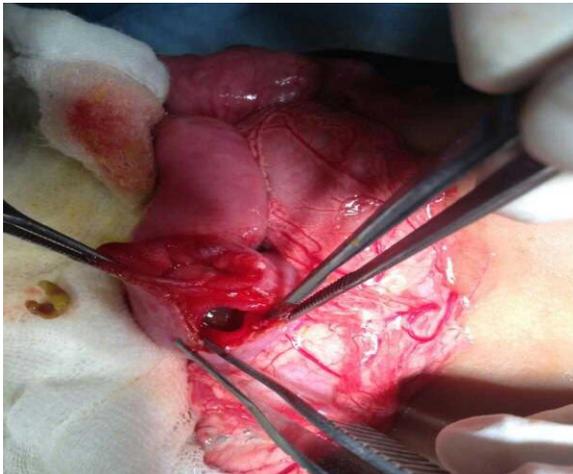


Figure 3. Resection.



Figure 4. Stricturoplasty

Table 1 - Differentiating characteristics between jejunal and

Characteristic	Jejunal Atresia	Ileal Atresia
Gestational age	Lower than that of ileal atresia	Low
Birth weight	Lower than that of ileal atresia	Low
Atresias	May be multiple	Simple
Antenatal perforation	Uncommon	Common
Associated malformations	Some	Rare
Postoperative course	Prolonged	Short
Mortality	Higher than that of ileal atresia	Low

ileal atresia.

DISCUSSION

Windsock deformity (WD) is a rare cause for intestinal obstruction. More often it occurs in the 2nd part of duodenum. Rarely it may occur in jejunoileal segment.

This windsock appearance is most commonly located in the second portion of the duodenum and consists of the barium-filled diverticulum that lies entirely within the duodenum.

The windsock appearance is formed by passive elongation of the intraluminal diverticulum due to continual peristalsis of the duodenum. [2,3]

Small intestinal atresia/stenosis most frequently affects the duodenum (~50%), followed by the jejunum (~35%); the ileum (~15%) is least likely to be affected. [4,5] There are several different types of jejuna and ileal atresia: they can range from having a small area of blockage or web to missing large sections of the intestines.

It historically has been categorized into Type I (mucosal web), Type II (atretic fibrous cord), Type IIIa (V shaped mesenteric defect), Type IIIb (apple peel atresia) and type IV (multiple atresias). [6] [Figure 5]

A severe form of duodenal atresia/stenosis is described as “apple-peel” deformity. This name is derived from the appearance of the intestine as it spirals around the blood supply and resembles an apple peel. [7]

The “apple-peel” or “Christmas-tree” deformity (type IIIb) occurs in approximately 10% of cases and is associated with atresia near the ligament of Treitz, lack of a dorsal mesentery, and recirculatory, retrograde blood supply from the ileocolic, middle colic, or right colic arterial distribution to the distal bowel. [8]

Clinical presentation of patients with jejunoileal atresia is as follows:

- Common characteristics
 - Polyhydramnios on prenatal ultrasound (28%) [9]
 - Prematurity (35%) [10]
 - Low birth weight (25-50%) [11]
- Classic signs
 - Bilious emesis that warrants emergent surgical evaluation (most patients)

- Abdominal distention (in distal atresias)
- Jaundice (32%)
- Failure to pass meconium in the first 24 hours (Rule out Hirschsprung disease. Passage of meconium does not rule out intestinal atresia.)
 - Signs of continuous fluid loss
- Dehydration, manifested by sunken fontanel and dry membranes
- Decreased urine output (best clinical indication of tissue perfusion)
- Tachycardia
- Decreased pulse pressure
- Low-grade fever
- Neurological involvement, manifested by irritability, lethargy, or coma

There are certain differentiating characteristics between jejunal and ileal atresia [Table 1]. [10-14]

Video capsule endoscopy: Although in use for more than 10 years in specific GI conditions in adults, this has been recently introduced used in the diagnosis of small intestinal atresias in neonates. [15]

Treatment usually involves surgical resection of the area of obstruction although newer options have become available in recent years such as endoscopic balloon dilatation. [16] In recent years, however, several authors [17,18] have reported the use of resection and primary anastomosis as a reasonable treatment option regardless of the location of the colon atresia.

CONCLUSION

Windsock deformity (WD) in jejunoileal segment of the intestine is uncommon as compared to duodenum. The treatment for jejunoileal web

remains surgical excision even though variable approaches may be used. (*Endoscopic laser therapy has been successfully tried for duodenal web. Simultaneous laparotomy with endoscopy has also proved useful.* [19]) Our patient underwent resection of the mucosal web and stricturoplasty.

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