

## A Rare Disease in a Neonate: Segmental Intestinal Volvulus

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### Abstract

Primary segmental intestinal volvulus (PSIV) is a rare disease but shows an aggressive clinical course. Early diagnosis and prompt management is paramount in these children otherwise life threatening necrosis and perforation may occur. A 1-day-old newborn girl with PSIV is presented in this study for whom an urgent surgical intervention including resection of ischemic ileal segment and ileo-ileal anastomosis solved the problem. It is also aimed in this report to emphasize the presentation, imaging findings and management of PSIV with a brief literature review.

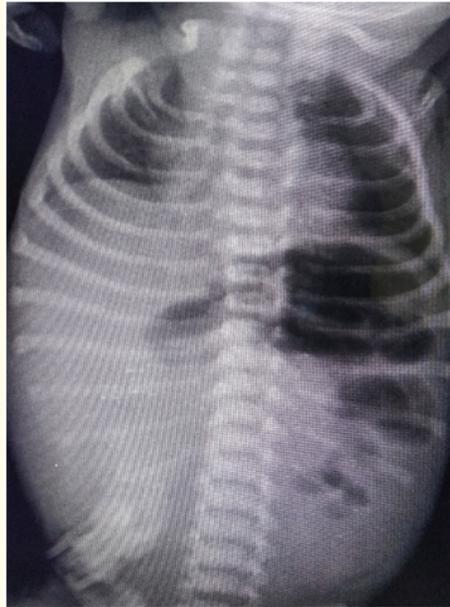
**Keywords:** Primary Segmental Intestinal Volvulus; Children

### Introduction

Primary segmental intestinal volvulus (PSIV) is a rare disease but shows an aggressive clinical course. In order to prevent necrosis and perforation, early diagnosis and prompt management is paramount. Here we present a 1-day-old newborn girl with PSIV whose clinical features and radiologic findings appeared to be intestinal atresia. An urgent surgical intervention including resection of ischemic ileal segment and ileo-ileal anastomosis solved the problem. It is also aimed in this report to review the presentation, imaging findings and management of PSIV under the light of relevant literature.

### Case Report

A 1-day-old female weighing 2110 g at 35 weeks gestation was born via caesarean section to a 34-year-old mother (G2P2). The patient was admitted to our department with abdominal distention, bilious emesis and failure to pass meconium on the day of birth. Antenatal history was unremarkable. The Apgar scores were 7 at 1 minute and 8 at 5 minutes. Plain abdominal radiograph was suggestive of neonatal intestinal obstruction including abdominal distention with displacement of intestinal gasses towards left (Figure 1). A water soluble enema study was found to be unremarkable. A nasogastric tube drainage failed to alleviate the obstruction. Initial medical management included restoration of adequate body temperature, hydration and electrolyte balance together with empiric antibiotic coverage including ampicillin, gentamicin and metronidazole. The neonate underwent an urgent exploratory laparotomy with a preoperative diagnosis of intestinal atresia. Mid-ileal volvulus was encountered with ischemic changes of the 15 cm of the involved ileum (Figure 2). There was no evidence of obvious pathologies responsible for volvulus like malrotation, intestinal atresia or congenital bands. The involved ileal segment was resected and end-to-end ileoileal anastomosis was performed (Figure 3). Histopathologic examination revealed a transmural ischemic infarct of resected ileal segment. The postoperative course was uneventful. Bowel activity returned 5 days following surgery and she was discharged home 7 days following surgical treatment. The baby is gaining weight and doing well and is under our follow-up.



**Figure 1:** X-ray showing a rather gasless distended abdomen with dilated intestinal loops displaced to the left of the abdominal cavity.



**Figure 2:** Operative view showing volvulated and ischemic ileal segment.



**Figure 3:** Operative view at the end of surgical intervention. Note the ileo-ileal anastomosis finished and large mesenteric defect closed with absorbable sutures.

### Discussion

Anomalies of the gut development including a narrow intestinal mesenteric root has long be considered as the major cause of volvulus with catastrophic end result like intestinal necrosis [1]. If whole intestine including ascending colon is involved, massive intestinal necrosis leading to short bowel disease may occur as an unwanted end result. It has been reported that of the intestinal volvulus, 80% of cases present during the first year of life and of these 60% are diagnosed at the neonatal period [2].

In children, small bowel volvulus is usually secondary to congenital or postoperative bands, duplication cyst, internal herniation and Meckel’s diverticulum [3-7]. PSIV is the torsion of a segment of the small intestine without any other abnormalities. It has been reported that volvulus without malrotation occurs in 19 to 26% of small bowel volvulus and PSIV effecting ileum during the neonatal period is extremely rare and most of these effected babies have prematurity [5,8].

The exact cause of PSIV is not clear. Some possible mechanisms of PSIV reported previously include stasis of the bowel content, long, narrow, band-like mesentery, changes in the intraabdominal pressure and hyper-peristalsis [9,10]. Other postulated reasons for PSIV include insufficient fixation of the intestines, immoderate initiation of feedings at an early stage of life and abdominal nursing including abdominal wall massage [11,12]. Our patient did not reveal any of these predisposing factors and an ischemic volvulated ileal segment was safely resected and ileo-ileal anastomosis reconstituted the continuity of the gastrointestinal tract. The differential diagnosis of PSIV includes NEC, spontaneous intestinal perforation, meconium plug syndrome and ileal atresia [13-15]. Our preoperative diagnosis was intestinal atresia which turned out to be primary segmental ileal volvulus postoperatively.

Majority of the previous reports of children with PSIV include preterm neonates and the occurrence of this entity in the neonatal period is extremely rare [5,7,15]. Although there are no specific clinical findings revealing PSIV, clinical course of PSIV may involve catastrophic results including massive rectal bleeding causing intractable shock state [16]. We did not observe rectal bleeding in our case but there was clinical evidence of sepsis including instability in thermoregulation, tachycardia and tachypnea. In a previous report it has been reported that, of the children with intestinal volvulus, ischemic changes of the affected bowel were seen in 90% of the cases without malrotation compared to the 18% incidence in the cases with malrotation [5]. It has been suggested that colon has the role of a cushion and a fixed cecum results in a tight volvulus while a mobile cecum results in a flexible volvulus and the end result is less severe ischemia and delayed necrosis [9,16]. It should also be kept in mind that extensive intestinal necrosis that may occur in volvulus is one of the three common causes of short bowel syndrome together with necrotizing enterocolitis and intestinal atresia [17,18]. Hopefully a limited segment of ileum measuring 15 cm was found to be ischemic in our patient. Although abdominal cavity was not found to be contaminated due to absence of intestinal perforation, resection of volvulated ischemic intestinal segment with ileo-ileal anastomosis was found to be inevitable in our case.

## Conclusion

In conclusion, diagnosis of PSIV is challenging due to the lack of specific clinical and radiologic findings and confirmation of this disease entity is only possible at laparotomy. PSIV should be considered as an urgent surgical disease and a prompt laparotomy is indicated for the preservation of gastrointestinal tract. The health providers dealing with these children should keep this anomaly in mind and a prompt pediatric surgical consultation is recommended and the patient should be treated accordingly.

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