

transplantation can prevent the recurrence of ALF [2] and should be offered in presence of INR >4 and total bilirubin >17.6 mg/dL, irrespective of hepatic encephalopathy [5]. Many metabolic/genetic disorders are associated with recurrent ALF in pediatric age group [4,6]. Recurrent ALF associated with mild infections or febrile episodes is extremely rare and can occur in mitochondrial defects, neuroblastoma amplification sequence deficiency and Wolcott-Rallison syndrome [4]. It is recommended that any child of consanguineous parents presenting with diabetes within the first 6 months of life should be tested for *EIF2AK3* mutations [1,2]. Patients should be carefully managed during every febrile episode and early referral to a liver transplant centre should be done, if liver function deteriorates. Recurrent ALF in pediatric age group should raise the possibility of autoimmune, metabolic or genetic diseases even if other obvious etiologies are present.

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JOSEPH J VALAMPARAMPIL^{1*}, NARESH SHANMUGAM¹
AND MOHAMED RELA^{1,2}

From Department of Paediatric Hepatology,
¹Institute of Liver Disease and Transplantation
Dr Rela Institute and Medical Centre, Chennai, India; and
²Kings College Hospital, London, United Kingdom.
*josephvalam@yahoo.co.in

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Primary Segmental Intestinal Volvulus in a Neonate

Primary segmental intestinal volvulus is a rare disease with an aggressive clinical course. Early diagnosis and prompt management prevents life-threatening necrosis and perforation. A 1-day-old newborn girl with this disorder is reported to emphasize the presentation, imaging findings and management.

Keywords: *Necrosis, Perforation, Surgical emergency.*

P rimary segmental intestinal volvulus (PSIV) is a rare disease with an aggressive clinical course. To prevent necrosis and perforation, early diagnosis and prompt management is paramount. A 1-day-old newborn girl with PSIV whose clinical features and radiologic findings appeared to be intestinal atresia is presented.

A 1-day-old female weighing 2110 g at 35 weeks gestation was born *via* caesarean section to a 34-year-old mother (para 2). The patient was admitted to our department with abdominal distention, bilious emesis and

failure to pass meconium. Plain abdominal radiograph was suggestive of neonatal intestinal obstruction. Initial management included restoration of adequate body temperature, hydration and electrolyte balance. The neonate underwent an urgent exploratory laparotomy. Mid-ileal volvulus was encountered with ischemic changes of the 15 cm of the involved ileum. 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. Histopathologic examination revealed an ischemic infarct of resected ileal segment. The postoperative course was uneventful. The baby is gaining weight and doing well and is under our follow-up.

Anomalies of the gut development including a narrow intestinal mesenteric root has long been considered as the major cause of volvulus with catastrophic end result like intestinal necrosis [1]. If whole intestine is involved, massive intestinal necrosis leading to short bowel disease may occur. Of the intestinal volvulus, 80% of cases present during the first year of life and of these 60% are

diagnosed in the neonatal period [2,3]. PSIV is the torsion of a segment of small intestine without any other abnormalities. Volvulus without malrotation occurs in 19 to 26% of small bowel volvulus, and PSIV affecting ileum during the neonatal period is extremely rare and usually occurs in preterms [4].

The exact cause of PSIV is not clear. Some possible mechanisms include stasis of the bowel content, long, narrow, band-like mesentery, changes in the intraabdominal pressure and hyper-peristalsis, insufficient fixation of the intestines, immoderate initiation of feedings at an early stage of life and abdominal nursing including abdominal wall massage [5,6]. Our patient did not reveal any of these predisposing factors. The differential diagnosis of PSIV includes NEC, spontaneous intestinal perforation, meconium plug syndrome and ileal atresia.

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]. Although there no specific clinical findings revealing PSIV, clinical course of PSIV may involve catastrophic results including massive rectal bleeding causing intractable shock state. We did not observe rectal bleeding in our case but there was clinical evidence of sepsis. In a previous report, of the children with intestinal volvulus, ischemic changes of the affected bowel were seen in 90% of the cases without malrotation as compared to 18% incidence in the cases with malrotation [5]. 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. The end result is less severe ischemia and delayed necrosis. It should 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. A limited segment of ileum was ischemic in our patient. Abdominal cavity was not contaminated and resection of volvulated ischemic intestinal segment with ileo-ileal anastomosis was required in our case.

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.

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VOLKAN SARPER ERIKCI
Saglik Bilimleri University,
Department of Pediatric Surgery,
Tepecik Training Hospital, Izmir, Turkey.
verikci@yahoo.com

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Colonic Perforation in a Term Newborn with Hereditary Protein C Deficiency

We describe a term infant who experienced recurrent apnea associated with intracranial hemorrhage and later, developed colonic perforation. Plasma protein C activity was below detectable limits and a heterozygous *PROC* mutation was identified. Neonatal colonic perforation is rare, and this case report highlights the importance of considering congenital Protein C deficiency.

Inherited protein C deficiency is a prothrombotic condition caused by homozygous or compound heterozygous defects in the *PROC* gene (2q13-q14). Purpura fulminans, intracranial hemorrhage (ICH), and blindness are the major complications in affected patients. Although heterozygous protein C deficiency is usually of mild severity, it can also cause severe symptoms, especially during the neonatal period.

A 1-day-old female newborn was transferred to our hospital because of recurrent apnea (**Fig. 1**). The baby