The Turkish Journal of Pediatrics 1997; 39: 551-555

BUDD – CHIARI SYNDROME IN A CHILD SECONDARY TO MEMBRANOUS OBSTRUCTION OF THE HEPATIC VEIN TREATED BY PERCUTANEOUS TRANSLUMINAL ANGIOPLASTY*

Report of A Case

*Buket Altuntaş MD***, Neşe Yaralı MD***, Semanur Kuyucu MD****

*Zafer Arslan MD*****, Ülker Ertan MD***, Ayşe Erden MD******

*Turan Cumhur MD*******, Tahsin Teziç MD********

SUMMARY: Altuntaş B, Yaralı N, Kuyucu S, Arslan Z, Ertan Ü, Erden A, Cumhur T, Teziç T. (Dr. Sami Ulus Children's Hospital and Department of Radiology, Yüksek İhtisas Hospital, Ankara, Turkey). Budd-Chiari syndrome in a child secondary to membranous obstruction of the hepatic vein treated by percutaneus transluminal angioplasty. Turk J Pediatr 1997; 39: 551-555.

Budd-Chiari syndrome (BCS) due to membranous obstruction of the hepatic vein and the inferior vena cava is rare in children. We report a child with BCS that had a membranous obstruction at the level of the hepatic veins. The web was successfully dilated percutaneously by balloon catheters. Symptoms and signs of obstruction improved without any complication. As percutaneous catheterization is an effective, safe and repatable procedure, we recommend this technique for treatment of children and adults with BCS due to membranous obstruction of the hepatic veins. *Key words: membranous obstruction, hepatic vein, children, Budd-Chiari syndrome, percutaneous transluminal angioplasty.*

Budd-Chiari syndrome (BCS) is a relatively uncommon group of disorders caused by occlusion of the major hepatic veins and/or the inferior vena cava (IVC), at or near the level of the hepatic vein ostia. Patients with BCS present with ascites, abdominal pain, hepatomegaly, edema, and occasionally jaundice. Depending on the rapidity and the extent of the obstruction of hepatic vein outflow, the course of the syndrome may be rapid or chronic. The majority of the cases are due to an obstruction of the hepatic veins or IVC caused by thrombi or tumors[1]. Very infrequently, BCS develops secondary to a membranous obstruction of the hepatic vein[2].

Interventional radiological procedures have gained wide acceptance in the treatment of patients with BCS. One such procedure is percutaneous transluminal angioplasty of occlusion or stenoses of the hepatic vein or IVC[3].

---

* From Dr. Sami Ulus Children's Hospital, and Department of Radiology, Yüksek İhtisas Hospital, Ankara.

** Pediatrician and Pediatric Gastroenterologist, Dr. Sami Ulus Children's Hospital.

*** Pediatrician, Dr. Sami Ulus Children's Hospital.

**** Associate Professor of Pediatrics and Pediatric Allergist, Dr Sami Ulus Children's Hospital.

***** Radiologist, Yüksek İhtisas Hospital.

****** Associate Professor of Radiology, Yüksek İhtisas Hospital.

******* Professor of Pediatrics and Pediatric Endocrinologist, Dr. Sami Ulus Children's Hospital.

In this case report, we report a child with BCS due to membranous obstruction of the hepatic vein who was treated by percutaneous hepatic vein angioplasty.

Case Report

A nine-year-old boy presented with a three-week history of abdominal distention and hematemesis. Physical examination revealed a thin boy, without jaundice, with a normal chest and heart. Slight dilatation of the superficial veins of the abdomen was noted. The liver edge was palpated 2 cm below the right costal margin and the spleen 2 cm below the left costal margin. There were marked ascites and edema of the lower extremities and scrotum. Paracentesis yielded a transudate with an albumin of 0.67 g/dl. Results of liver function tests were mildly elevated. Hemoglobin was 10.4 g/dl. Hypochrom microcytic anemia compatible with iron deficiency was present as demonstrated by low serum transferrin saturation. Serum ceruloplasmin, serum and urinary copper, α-1- antitrypsin and sweat electrolyte levels were normal. Hepatitis B surface antigen and anti hepatitis C antibody were negative. Portal system duplex Doppler ultrasonography demonstrated patent splenic and portal veins. There was an anastomosis between hepatic veins, and the caudate lobe was hypertrophic (Fig. 1). These findings are typical of Budd-Chiari syndrome. To assess the cause of thrombosis, serum levels of abnormal inhibitors of a hemostatic system, such as antithrombin III, protein C and protein S, were confirmed as normal, and antiphospholipid antibodies were negative. Patency of IVC was demonstrated by transfemoral inferior cavography. Membranous web between the hepatic vein and IVC was demonstrated by percutaneous transhepatic angiography (Fig. 2). Following transhepatic balloon dilatation of the membrane, ascites resolved completely and did not recur. An early follow-up ultrasonographic examination revealed a patent hepatic vein. Unfortunately, micronodular cirrhosis was diagnosed by liver biopsy (Fig. 3), and endoscopic examination of the esophagus revealed grade III varices.

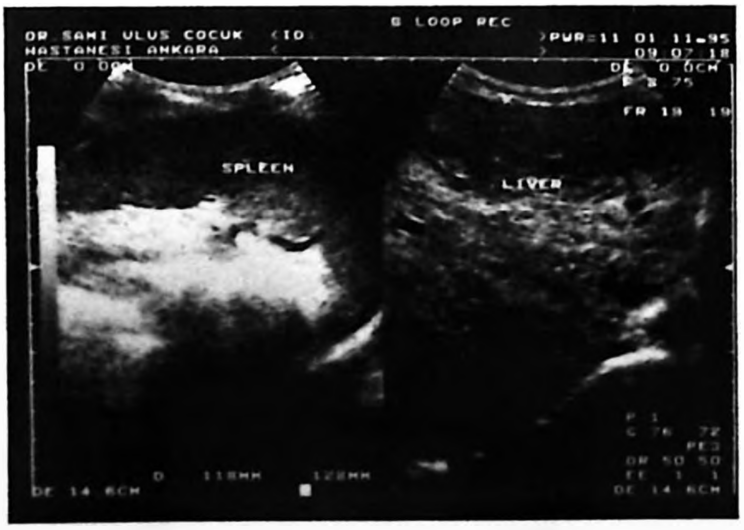

Fig. 1: Ultrasonography shows coarseness of the liver echogenicity. The hepatic veins are thin and irregular. Spleen is homogeneously enlarged.

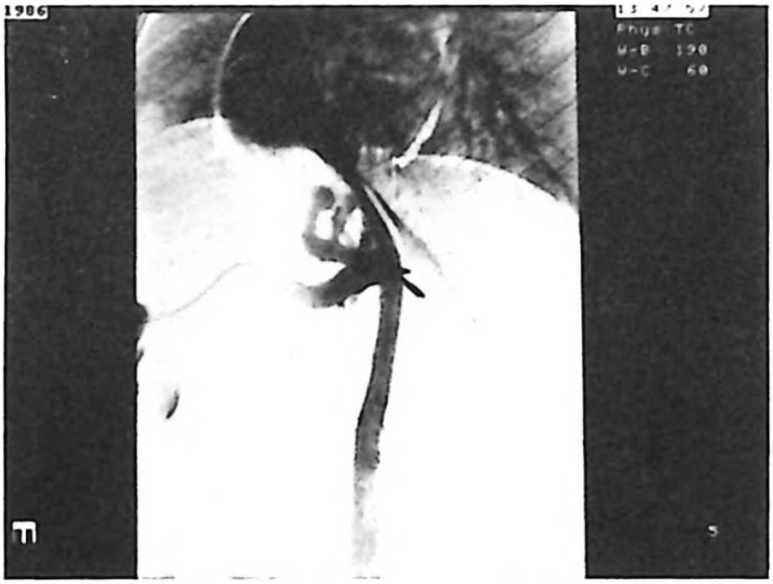

Fig. 2: Patent IVC at transfemoral inferior cavography; membranous web between the hepatic vein and IVC demonstrated by percutaneous transhepatic angiography.

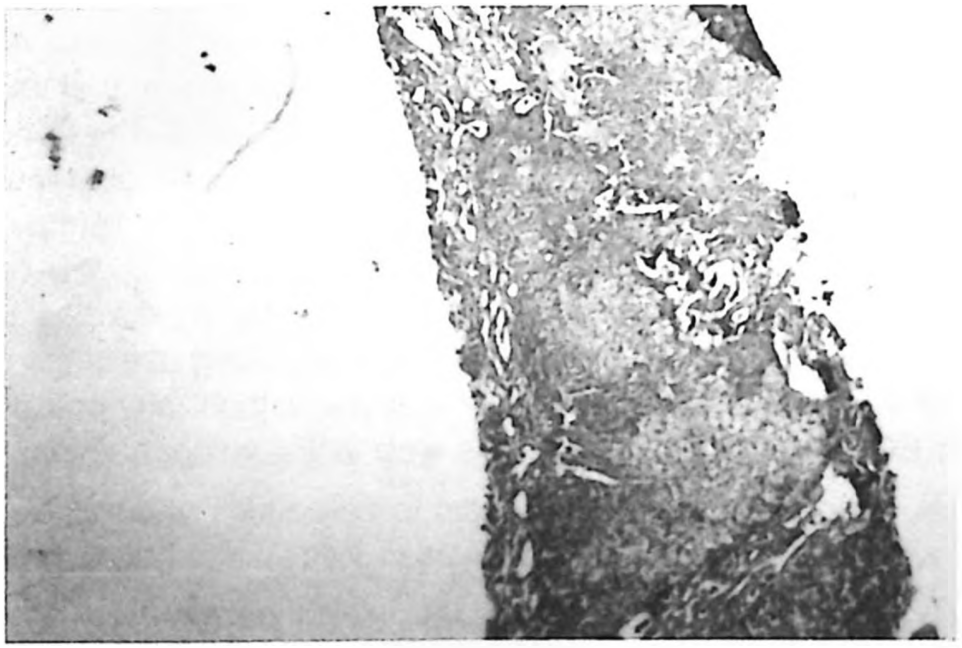

Fig. 3: Liver: Micronodular cirrhosis (HE x 400).

Discussion

Budd-Chiari syndrome is caused by an obstruction of the hepatic vein outflow. Possible causes of this syndrome are myeloproliferative disorders, thrombotic conditions, malignant neoplasm, and membranous obstruction of the hepatic vein and IVC. The latter is particularly infrequent, except in Orients, where it is a major cause of suprahepatic portal hypertension[4-6]. Chronic by nature, BCS can progress to liver cirrhosis if left untreated, resulting in a poor prognosis as in this case report[1].

In Turkish literature, we were unable to find information regarding the frequency of various causes of BCS. Okten et al.[7] reported four cases of membranous obstruction of IVC in adults. There is some controversy surrounding the etiology and pathogenesis of membranous obstruction. Some outhors accept the theory of thrombosis and organization, but others accept a congenital origin based on non-union or malunion during the developmental stages of the hepatic veins and IVC[8,9]. In our case, as the symptoms and the signs appeared at a very young age; the levels of antithrombin III, protein C and protein S (potential causes of thrombosis) were normal; and antiphospholipid antibodies were negative, we suggest that the membrane in the hepatic vein was congenital.

Treatment options include conservative treatment, interventional radiological techniques, a variety of surgical vascular by-passes, decompression procedures, and liver transplantation. Treatment method should be individualized according to the site of obstruction and the extent of liver failure. The commonly used surgical procedures include finger fracture, transcardiac membranotomy or bypass grafting. However, postoperative mortality rates with such procedures are high: 20 percent following transcardiac finger fracture and 40 percent following bypass grafting or inferior cavotomy[4]. Interventional radiological procedures have gained popularity in the treatment of BCS. These procedures include percutaneous transluminal angioplasty of occlusions or stenoses of the hepatic vein or IVC. Percutaneous transluminal angioplasty is a simple and safe procedure. It may be repeated if the initial attempt fails or the obstruction recurs[7,8]. Although there have been several reports on the use of balloon angioplasty in adults with BCS, there is little data regarding use of the procedure in children. We successfully used this procedure, without any complication, in a child with BCS due to a membranous web in the hepatic vein.

In conclusion, balloon angioplasty appears to be a safe and effective treatment for children with BCS due to membranous obstruction of the hepatic veins or IVC.

REFERENCES

1. Mitchell MC, Boitnott JK, Kaufman S, Cameron JL, Maddrey WC. Budd-Chiari syndrome: etiology, diagnosis and management. Medicine 1982; 61: 199-218.
2. Hoffman HP, Stockland B, Heyden U. Membranous obstruction of the inferior vena cava with Budd-Chiari syndrome in children: a report of nine cases. J Pediatr Gastroenterol Nutr 1987; 6: 878-884.
3. Martin LG, Henderson JM, Millikan WJ Jr, Casaerella WJ, Kaufman SL. Angioplasty for long-term treatment of patients with Budd-Chiari syndrome. Am J Roentgenol 1990; 154: 1007-1010.
4. Hirooka M, Kimura C. Membranous obstruction of the hepatic portion of the inferior vena cava. Arch Surg 1976; 100: 656-663.
5. Kimura C, Matsuda S, Koie H, Hirooka M. Membranous obstruction of the inferior vena cava: clinical study of nine cases. Surgery 1971; 74: 551-559.

6. Takeuchi J, Takada A, Hosumara Y, Matsuda Y, Ikegami F. Budd-Chiari syndrome with obstruction of the inferior vena cava. Am J Med 1971; 51: 11-20.
7. Okten A, Besisik F, Boztas G, et al. Vena cava inferiorun membranoz obstruksiyonu ve Budd-Chiari sendromu. Turk J Gastrohepatol 1993; 4: 295-298.
8. Terabayashi H, Okuda K, Nomura F, Ohnishi K, Wong P. Transformation of inferior vena caval thrombosis to membranous obstruction in a patient with the lupus anticoagulant. Gastroenterol 1986; 91: 219-224.
9. Kage M, Arakawa M, Kojiro M, Okuda K. Histopathology of membranous obstruction of the inferior vena cava in the Budd-Chiari syndrome. Gastroenterol 1992; 102: 2081-2090.
10. Sawada S, Oshima T, Kato T, Kubota Y, Tanaka Y. Budd-Chiari syndrome treated by percutaneous transluminal angioplasty. Acta Radiol Diagnosis 1985; 26: 25-27.
11. Yamada R, Sato M, Kawabata M, Nakatsuka H, Nakamura K, Kobayashi N. Segmental obstruction of the hepatic inferior vena cava treated by transluminal angioplasty. Radiology 1983; 149: 91-96.