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# Budd-Chiari Syndrome in Childhood: Presentation of 13 Cases*

**Nurten Koçak, M.D.** / Şinasi Özsoylu, M.D.*** Safiye Göğüş, M.D.******

Occlusion of the hepatic veins (Budd-Chiari Syndrome) is rare in childhood.[1] Intra or perihepatic neoplastic processes thrombosis, polycythemia vera, inflammatory bowel disease, visceral thrombophlebitis migrans, and congenital web or hypoplasia of the hepatic veins all produce occlusion of the hepatic veins (Budd-Chiari Syndrome).[2,3] Rapidly developing ascites, hepatomegaly, edema of the legs, and mild icterus are the classical findings.[1] Diagnosis of this entity is possible by histopathological appearance of centrilobular hemorrhagic necrosis[4] and by hepatic venography showing the occlusion of the hepatic veins.[5] The prognosis in this disease is poor. Because of the rarity of this entity, we would like to present 13 cases of Budd-Chiari Syndrome.

## *Material and Methods*

Thirteen cases of Budd-Chiari Syndrome were diagnosed at Hacettepe Children's Hospital since 1968. The patients' ages ranged from 13 months to 15 years; five were females and 8 males. Diagnosis was verified by incision biopsy in one, by postmortem biopsy in one, by autopsy in one, and by liver needle biobsy in ten. In additon, on 6 of the patients inferior venacavography was performed.

## *Case Reports*

Two of the case reports are given in detail. The clinical and laboratory findings of all cases are summarized in Table I and II.

* From Hacettepe University, Children's Hospital, Department of Pediatrics,
**Associate Professor of Pediatrics.
*** Professor of Pediatrics, Hematologist, Head of the Pediatric Hematology and Hepatology Sections.
**** Associate Professor of Pediatrics, Pediatric Pathologist.

TABLE I

THE CLINICAL FINDINGS OF THE PATIENTS

| Patients' number | 1 | 2 | 3 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 | 12 | 13 |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Age | 13/12 | 2 | 2 | 2 | 2.5 | 2.5 | 3 | 3 | 3 | 3 | 3.5 | 5 | 15 |
| Sex | M | F | F | M | F | M | M | M | M | F | F | M | M |
| Duration of ascites (in days) | 10 | 15 | 60 | 15 | 10 | 60 | 30 | 60 | 7 | 10 | 90 | 60 | 10 |
| Pretibial edema | + | + | − | + | − | + | + | − | − | − | − | + | − |
| Hepatomegaly (in cm) | 5 | 8 | 12 | 6 | 7 | 8 | 6 | 6 | 8 | 8 | 4 | 9 | 8 |
| Splenomegaly (in cm) | − | − | − | − | − | − | − | − | − | − | − | 3 | − |
| Icterus | − | − | − | − | − | − | − | − | + | − | − | − | + |
| Abdominal pain | − | − | − | − | + | − | − | − | − | − | − | − | − |
| Diarrhea | − | − | − | + | − | + | + | − | + | − | − | + | − |
| Vomiting | − | − | − | − | − | + | − | − | − | − | − | − | − |

## TABLE II

### THE LABORATORY FINDINGS OF THE PATIENTS

| Patients' number | 1 | 1 | 2 | 4 | 5 | 6 | 7 | 8 | 9 | 10 | 11 | 12 | 13 |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Hb g % | 11.4 | 7.98 | 10.16 | 9.15 | 9 | 7.50 | 4.5 | 6.73 | 10.87 | 8.30 | 11.70 | 13.26 | 12.82 |
| WBC | 13600 | 10000 | 6000 | 8000 | 8000 | 7000 | 5600 | 11000 | 11000 | 17000 | 24000 | 13100 | 9000 |
| Bilirubin (mg/dl) | — | — | — | — | — | — | — | — | 2.5 | — | 3.8 | — | — |
| SGOT (KU) | 110 | 53 | 105 | 26 | 51 | 27 | 37 | 280 | 142 | 240 | 45 | 87 | 240 |
| SGPT (KU) | 84 | 30 | 32 | 8 | 9 | 12 | 12 | 140 | 47 | 64 | 20 | 22 | 200 |
| CCF | + | + | ++ | ++ | ++ | ++ | ++ | ++ | | +++ | ++ | +++ | |
| Thymol (U) | — | 4.5 | 9 | 9.7 | 5 | 8.9 | 7 | 1.5 | — | 17.4 | 5.8 | 3 | 4.6 |
| Ascites: | | | | | | | | | | | | | |
| Protein content (mg/dl) | 1680 | 3300 | 3240 | 2700 | — | 960 | 1080 | 2240 | 1800 | 960 | 1560 | 1500 | 2680 |
| Rivalta | + | — | + | + | — | — | — | + | + | — | | | |
| Specific gravity | 1012 | 1013 | 1010 | 1014 | — | 1012 | 1011 | 1011 | 1014 | 1006 | 1016 | 1014 | 1017 |
| Cells | RE | — | — | — | RE | | — | — | RE | — | RL | RL,RE | RE |
| Bone marrow | N | N | N | N | N | N | N | N | N | N | N | N | — |
| Peripheral venous pressure | ND | 22 cm | ND | ND | ND | 12 cm | 16 cm | 23 cm | 7 cm | ND | ND | ND | ND |
| Esophagogram | N | N | N | N | ND | ND | N | N | ND | N | ND | N | N |
| Hepatic venogram | occ | ND | occ | ND | ND | ND | occ | ND | occ | ND | occ | occ | ND |

| | |
|---|---|
| — | Negative |
| N | Normal |
| ND | Not done |
| RE | Rare erythrocytes |
| RL | Rare leukocytes |
| occ | Occlusion |

**Case Number 1:** M. Y., (464054) a 13 month old boy who was admitted to Hacettepe Children's Hospital because of extrophya vesicalis, epispadias, and abdominal swelling for 10 days. The abdomen was slightly distended and free fluid was detected. The liver was 5 cm palpable by ballotment. The character of the ascitic fluid was transudate; an I. V. P., and esophagogram were found to be normal. Microscopic examination by liver needle biopsy revealed intensive hemorrhagic necrosis of the central areas of the lobules with sparing of the portal areas. Hepatic venogram showed complete occlusion of the main vena hepatica. Diuretics were started. He was last seen in 1978 at 5 years of age; although his general condition and development were good, he still had ascites and pretibial edema. Unfortunately, further studies could not be performed.

**Case Number 6:** Y. G. was 2.5 year old boy (891265), whose abdominal swelling had started apruptly 2 months previously, following a short period of diarrhea. No history of icterus, hematemesis or melena was obtained. The abdomen was grossly distended due to ascites and the liver was enlarged, it was palpable 8 cm below the right costal margin by ballotment. The ascitic fluid was transudate. He had pretibial edema and a nonspecific lung infection. Bone marrow aspiration and an I. V. P. did not reveal any abnormalities. In spite of diuretics, antibiotics, and vigorous supportive treatment he died 2 days after the admission.

In the post-mortem examination, the liver was found to be enlarged (880 gr normal for his age being 458 g) and thrombi were detected in the hepatic veins and its large branches. The parenchyma was firm and congested with hemorrhage and necrosis producing a mottled, dark, red-brown appearance. Coronal sections of the liver showed several large hepatic veins to be occluded by thrombi. On microscopic examination, these thrombi were in different ages (recent, early organization, partial recanalization). There were severe hemorrhagic necrosis involving the central and mid-lobular zones of the liver. (Figures 1-4).

*Discussion*

Although portal hypertension is commonly observed in Budd-Chiari syndrome, in Turkish children it is more frequently observed in liver cirrhosis, idiopathic hypertension, and extrahepatic portal obstruction.[6] It very rarely occurs due to Cruveilhier-Baumgarten disease, however, we observed this in one of our previously.[7] Sudden onset of ascites is the most characteristic clinical feature of hepatic vein occlusion. Although ascitic fluid is generally transudate, the protein content may be high in some cases, and occasionally, the fluid may be hemorrhagic.[8] In all our cases, ascites developed rapidly and in 2 cases the protein content was

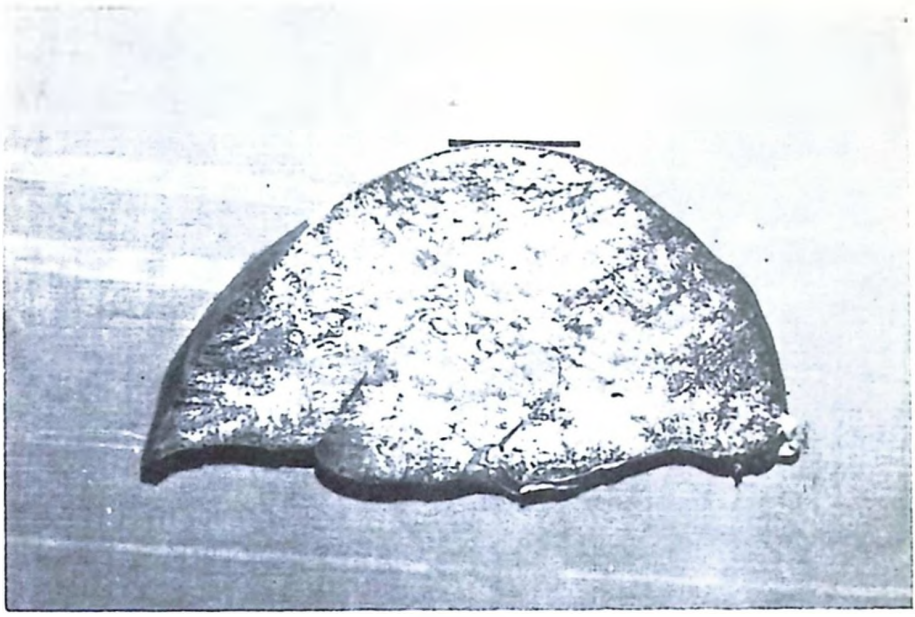

**Figure 1**
Several large hepatic veins are occluded (arrows) by organizing thrombus on coronal section of the liver

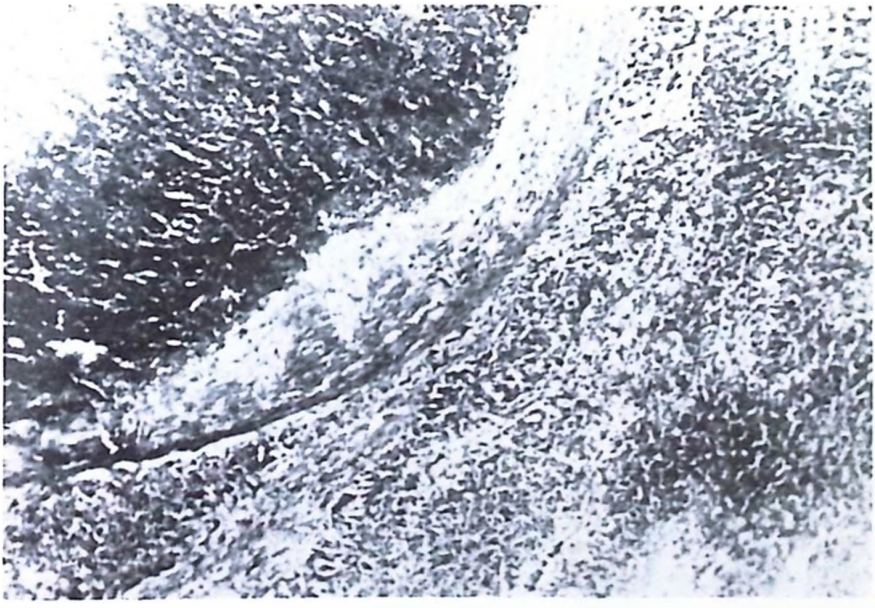

**Figure 2**
Recent thrombus in a large hepatic vein and severe passive congestion of the liver. H&E x 25

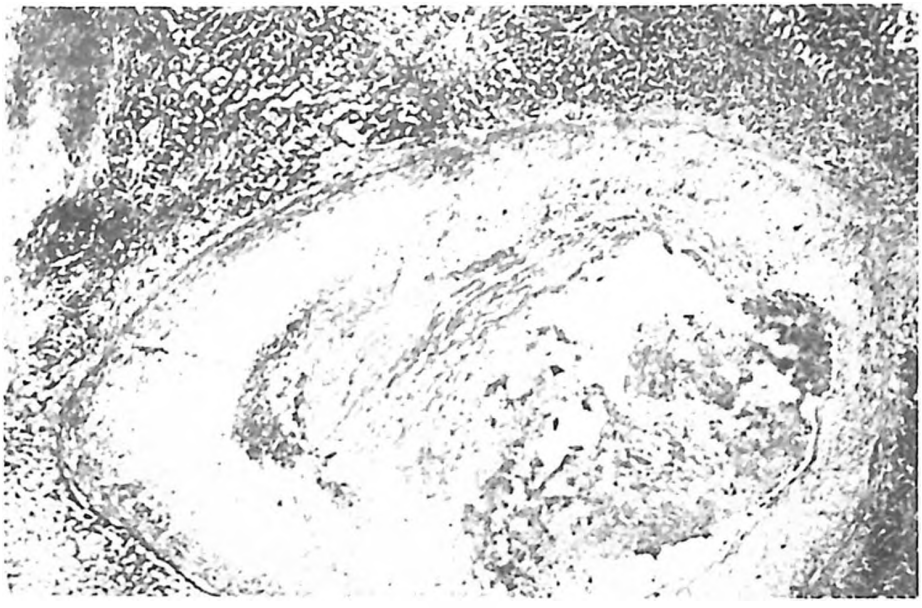

**Figure 3**
Early organization of thrombus in a large hepatic vein. H&E x 25

**Figure 4**
Severe passive congestion of liver with centrilobular hemorrhagic necrosis. H&E x 25

above 3 g/dl. Hepatomegaly is due to elevated intrahepatic venous pressure which may cause excessive bleeding following liver needle biobsy; however, this was not observed in any of our patients. Abdominal pain is a less constant symptom. Mild icterus in 2 of our patients and splenomegaly in one, may be found in up to 25 % of the cases[1] in chronic form of the disease. If the inferior vena cava is blocked, edema of the legs develops as was observed in 6 of our cases. In long-standing cases, esophageal varices due to portal hypertension are sometimes present, but was not seen any of our cases. In the acute type of the hepatic vein occlusion, inflammatory bowel disease, such as ulcerative colitis and Crohn's the clinical symptoms of which are diarrhea and vomiting, may be an etiological factor,[9] as it was in 5 of our cases.

Primary and secondary thrombi are some of the most important etiological factors in this syndrome in children.[9, 10] In one of our cases thrombosis of the hepatic veins was shown; however, the cause of thrombosis is unknown in this case. Intrahepatic or intra-abdominal tumors and trauma were also ruled out in our cases. Venocavagraphy was carried out in 6 of our patients as is necessary for the diagnosis of Budd-Chiari Syndrome.

In Budd-Chiari Syndrome central venous congestion of the liver and centrilobular hemorrhagic necrosis are the characteristic histopathological findings, and these changes were shown in all of our cases. These changes may also be observed in the veno-occlusive disease which is generally related to senesio alkaloids and crotalaria,[8, 11] but in this disease only the medium and small hepatic veins are involved. Although liver function test changes may occur, these were not found to be characteristic in our patients.

Porto-caval shunt may be successful when there is a patent inferior vena cava and absence of marked venous collaterals; however, the surgical procedures are very difficult in small children.[1] Paracentesis is not helpful and leads to bleeding or electrolyte imbalance. Therefore, treatment is generally symptomatic. Diuretics are recommended. Hepatic venous congestion progresses rapidly to hepatic failure, coma and death. Hepatic failure was suspected in one of our patients (Number 13). All the patients were put on diuretics, 2 of them died, 1 has ascites still and 10 did not come for follow-up.

*Summary*

13 cases of Budd-Chiari syndrome are presented, etiological, clinical, histopathological and laboratory findings are discussed and related literature is reviewed.

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