BUDD – CHIARI SYNDROME

*Şinasi Özsoylu MD**

To the Editor

I read with enthusiasm Altuntaş et al.'s interesting case presentation entitled "Budd-Chiari Syndrome in a Child Secondary to Membranous Obstruction of the Hepatic Vein Treated by Percutaneous Transluminal Angiography" in the recent issue of the Journal (1997; 39: 551-555).

Since the authors could not find information on the subject in Turkish literature, I would like to bring to their attention that, a study involving one of the largest groups with Budd-Chiari syndrome was published by our group[1].

There were some interesting findings in the authors' case. Despite the most likely congenital nature of the membranous web between the hepatic vein and IVC, symptoms of the disease were noticed three weeks prior to the admission of this nine-year-old boy. The liver is usually enlarged in this syndrome but was found almost normal-sized, possibly due to the well-developed cirrhosis. Although intensive central venous congestion and hemorrhagic necrosis of the liver are generally expected in this syndrome, it was not found in the present case, again probably related to the late clinical presentation of the patient.

As a follow-up to the authors' case, it would also be interesting to observe whether the morphologic changes of the liver and the esophageal varices would improve in time, since the cause has been successfully eliminated.

REFERENCES

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REPLY

*Buket Altuntaş MD**

To the Editor

It was interesting to see the comments of Dr. Şinasi Özsoylu to our article on treatment of hepatic vein obstruction by balloon dilatation (Turk J Pediatr 1997; 39: 551-555). Dr. Özsoylu has drawn attention to his publication about Budd-Chiari Syndrome (BCS) in childhood. The noted article was not shown in the references of our article, as the main aim of our article was to treat BCS due to membranous obstruction of the hepatic vein by balloon angioplasty. We could not find any information on the treatment of BCS in childhood by balloon dilatation in the Turkish literature.

Although it is generally believed that membranous obstruction of the hepatic vein and inferior vena cava is caused by congenital malformation, the congenital anomaly theory cannot explain the late onset of the disease, thus inviting other theories (i.e. thrombosis and its sequelae)[1]. As the lesion is rarely observed symptomatically in children[2] and no thrombotic conditions were noticed in our patient, membranous obstruction was thought to be of congenital origin. It can be concluded that our patient may be asymptomatic until the collateral circulation becomes inadequate and decompensation takes place.

Ascites disappeared after the second day of the treatment and variceal size was reduced on follow-up. We did not need to plan a second liver biopsy because cirrhosis was well developed on the first biopsy.

REFERENCES

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