RESEARCH PAPER

Pediatric Budd-Chiari Syndrome: A Case Series

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Objective: To study the diagnostic methods and treatment outcomes in children with Budd-Chiari syndrome. **Methods**: Case records of 25 patients with Budd-Chiari syndrome were evaluated retrospectively. These patients were investigated with imaging techniques and underwent balloon angioplasty or surgical management. **Results**: 21 patients underwent balloon angioplasty, of which 17 had good medium- to long-term results, while only one out of four patients who underwent a portocaval shunt survived.**Conclusion**: The balloon angioplasty has satisfactory outcome in the treatment of acute Budd-Chiari syndrome. In failed cases, the surgical therapy may be attempted, but the outcomes do not appear rewarding.

Keywords: *Balloon angioplasty, Outcome, Treatment, Venous thrombosis.*

Budd-Chiari syndrome (BCS) is a heterogeneous group of clinical conditions presenting with hepatic venous outflow obstruction from the level of the hepatic veins to the junction of the inferior vena cava (IVC) with the right atrium [1]. The causes of venous obstruction in children are many [1-3], with hypercoagulable state being the most common. Clinical manifestations in many cases are nonspecific, and high index of suspicion with imaging may be critical for early diagnosis of hepatic venous obstruction and accurate assessment of the extent of disease. The data regarding pediatric BCS are scarce, and the treatment strategies are not well defined. We describe our experience with diagnostic methods and outcome of the endovascular and surgical therapies in children with BCS managed at our center.

METHODS

The case records of children (age ≤12 y) diagnosed as BCS during January 2000 to January 2016 at our center were retrospectively reviewed. All these patients were diagnosed based on their clinical features, radiological investigations and angiographic findings. After relevant clinical examination, patients were further evaluated with laboratory and radiological investigations. Blood tests were aimed at finding the etiological factor, evaluating the extent of hepatic decompensation and ascertaining the fitness for the therapeutic procedure. Blood investigations were; complete blood count, liver function test, renal function test, coagulation profile and investigation for causative factors. A baseline abdominal ultrasonography along with color Doppler ultrasound using 'M mode' was performed in most patients. If color Doppler was suggestive of BCS, diagnosis was confirmed with computed tomography (CT) or magnetic resonance (MR) hepatic venous angiogram. Conventional angiography was performed under general anesthesia.

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The initial therapeutic approach was transjugular in all cases *via* internal jugular puncture or by railroading over IVC catheter that was introduced till the right internal jugular vein through femoral puncture. In selected cases where this approach failed, further attempts were made *via* a transhepatic route with insertion of a percutaneous transhepatic guidewire with railroading of catheter through transjugular route. With transjugular or rarely transhepatic wire in place, the balloon angioplasty was performed to achieve the patency of hepatic veins and IVC. Liver biopsy was performed at the end of angiography procedure under same anesthesia using trucut biopsy gun. Angioplasty was considered to be successful if restoration of or clear improvement in hepatic venous outflow was shown by contrast angiography, and symptomatic improvement. Post-procedure, patients were treated with intravenous heparin, which was tapered gradually. Patency and improvement was assessed with Doppler ultrasound and liver function tests. Patients were started on oral warfarin or subcutaneous low molecular weight heparin injections simultaneously when intravenous heparin was tapered.

WHAT THIS STUDY ADDS?

- Budd-Chiari syndrome may have good outcome if treated early with hepatic/ inferior vena cava venoplasty.

Adequacy of anticoagulation therapy was monitored with measurement of prothrombin time and activated partial thromboplastin time. Oral anticoagulation was continued for at least one year. If patency was not demonstrated on Doppler ultrasound, the patients were posted for porto-caval shunt surgery if liver biopsy showed non-cirrhotic change, and were referred to liver transplantation surgery, if liver was cirrhotic.

RESULTS

We evaluated records of 25 children (14 boys). Abdominal distension was the most common presenting symptom seen in 20 patients followed by associated or preceding loose motions in 8 patients. Other symptoms seen were poor feeding (*n*=2), limb swelling/edema (*n*=3), fever (*n*=3), and inguino-scrotal swelling, abdominal pain, lower limb pain, vomiting and respiratory distress in one patient each. Color Doppler was performed in 18 patients and could diagnose hepatic vein obstruction in 15 patients. In 3 patients, diagnosis of hepatic venous obstruction was missed on color Doppler and was diagnosed by MR angiography (*n*=1) or conventional hepatic/IVC angiography (*n*=2). CT scan with CT hepatic venous angiography was performed in 11 patients and MR angiography was done in 9 patients, of which 3 patients had both CT scan and MR angiography. In 20 patients, the site of obstruction was convincingly diagnosed by radiological investigations (Doppler ultrasound or CT angiography or MR angiography), whereas these investigations failed to delineate site of obstruction in five patients. In these five patients, hepatic/ IVC angiography convincingly diagnosed a hepatic venous/IVC obstruction.

Hepatic-IVC angiogram was performed in 21 patients and antegrade flow was established successfully in 20 patients. In all the patients one or more hepatic veins were obstructed, and in five patients IVC was obstructed in addition to hepatic veins. In one of these patients, transjugular approach failed and hence transhepatic route was used, while in one other patient jugular as well as transhepatic route failed. Of the four patients who did not undergo hepatic/IVC angiography, two showed spontaneous resolution of symptoms and two patients died. Four patients underwent portocaval shunting with H-portocaval jump graft; one survived while three died. Three patients out of 20 patients in whom flow was established after successful angioplasty, developed recurrence of symptoms after few months. All these three patients underwent portovenous shunt surgery, of which two died in late postoperative period. Overall, out of 25 patients, 17 patients recovered and survived while eight patients died. Two died before any intervention could be done, one died (post-shunt surgery) after failed attempt at angioplasty, and five died due to other reasons (two post-shunt surgery, one each with sepsis following lower respiratory tract infection and pulmonary hemorrhage, and cause was not known in one patient as patient did not follow-up) following successful angioplasty. Of the 15 available liver biopsy reports, four patients showed changes of cirrhosis and were referred for liver transplantation and the remaining 11 patients who showed no changes of liver cirrhosis, 7 responded well to venoplasty treatment and were continued on anticoagulation. Four patients who showed noncirrhotic changes and did not respond to venoplasty intervention eventually underwent portosystemic shunt surgery.

DISCUSSION

In this hospital record review of 25 patients of BCS, we could achieve flow in atleast one hepatic vein in 20 (95%) out of 21 procedures and were able to maintain patency in 17 (80%) of these patients. Overall there were 8 deaths, out of which 5 were in those in whom flow in hepatic veins was achieved.

The major limitation in our study is an uncontrolled design and lack of comparison of hepatic balloon angioplasty with venous stents. We believe that the hepatic venous stents may be difficult to place in young infants and children; they have risk of migration and are prone for blockage requiring prolonged life-long anticoagulation. Similarly, our policy was to proceed with either portovenous surgical shunt or refer to liver transplant in failed balloon angioplasties depending on the liver histology. Hence we lack comparison with transhepatic portacaval shunt (TIPPS) or direct intrahepatic portacaval shunt (DIPS). Moreover, a hypercoagulable state is expected in patients with BCS, but we could not perform relevant work-up in our series.

Interventional radiological procedures for BCS are described in adults but literature on this modality in pediatric age group is scarce [4,5]. Miller, *et al*. [6] studied imaging with pathological findings in 21 patients and concluded that each imaging modality offers certain

values and limitations in the assessment of vascular or parenchymal findings in BCS. In this study, status of hepatic veins was correctly assessed and correlated with pathology in 13 of 20 patients who had sonograms, in 12 of 15 patients who had MRI, and in nine of 18 patients with contrast-enhanced CT scans [6]. In our study also, all three modalities along with hepatic angiography were used for diagnosis.

The primary goal of treatment is the resolution of hepatic congestion in order to improve liver perfusion and preserve functioning hepatocytes. If medical therapy fails, then valuable time to salvage the functioning liver may be lost. Hence we used aggressive early use of interventional radiology. Recanalization of stenotic or occluded hepatic veins to restore venous outflow is the initial procedure of choice [5]. Various approaches may be used to access the hepatic veins. Transjugular catheterization by internal jugular vein puncture or railroading over IVC catheter through femoral puncture is attempted first. If the stenotic or occluded hepatic vein cannot be crossed by using a transjugular approach, an ultrasound-guided transhepatic needle puncture of a patent segment of the hepatic vein is performed to obtain access [6-8]. In one of our patient, transhepatic access was used successfully for hepatic venous angiography, while even this approached failed in one patient. Balloon angioplasty had good results in our series. In patients with failed endovascular approach, surgical therapy of portocaval shunting was attempted if liver was congested and noncirrhotic. Our results of surgical therapy were not rewarding as only one out of four such patients survived.

To conclude, early aggressive use of interventional radiological procedure may achieve successful diagnosis and recanalization in most children with BCS. In failed cases, the surgical therapy may be attempted, but the outcomes do not seem to be rewarding. If the access for balloon angioplasty is difficult through IJV, it can be aided by transfemoral or transhepatic approach aggressively.

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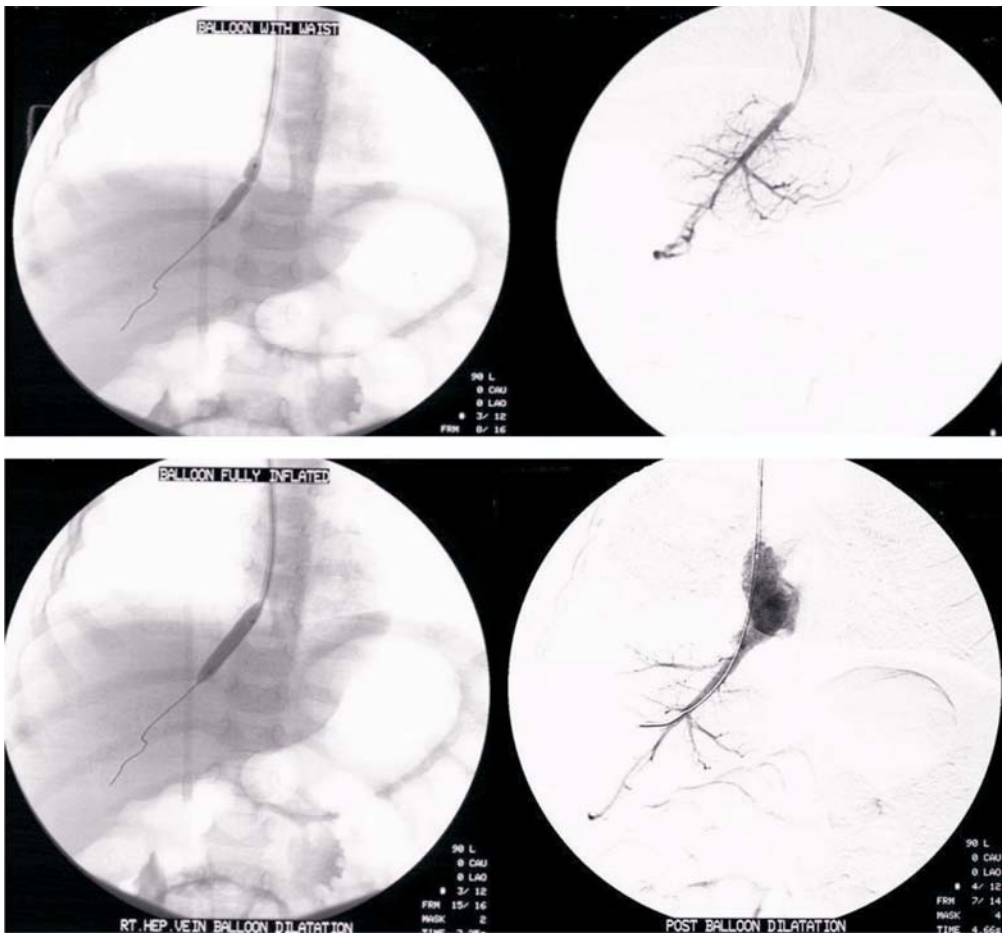

WEB FIG. 1 *Pre dilatation (upper half) showing waist like narrowing with absence of free flow towards heart, whereas post dilatation (lower half), the narrowing has disappeared and there is free flow of contrast to heart.*