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*Original article*

# Alternate pathways in hepatic venous outflow obstruction by color doppler imaging

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## SUMMARY

**Introduction: Primary Budd – Chiari syndrome is a curable form of portal hypertension. Doppler ultrasonography is an excellent tool in the primary diagnosis of Budd Chiari syndrome and in the follow up of patients after definitive therapy. Methodology: Ninety-seven patients with clinically suspected Budd Chiari syndrome underwent color Doppler imaging to identify the site and cause for obstruction, flow across the obstructed segment of the hepatic veins and / or inferior vena cava and to identify the collateral pathways of hepatic venous drainage. Sixty-two patients were subjected to angiography and Doppler – angiography correlation was also studied. Results: Seventy one out of the ninety seven patients screened had Doppler evidence of Budd Chiari syndrome. 23.94%, 18.30%, 57.74% had type I, type II and type III Budd chiari syndrome respectively. Intrahepatic "comma" collaterals and inferior right hepatic vein were the major collaterals identified in patients with type I and type III disease. Doppler correlated with angiogram in 56 out of 62 patients (90.3%). Conclusion: Color Doppler imaging is an excellent technique for qualitative data on flow direction and pattern, thereby contributing significantly to the diagnosis. It is the choice of investigation in the initial evaluation of patients with suspected primary Budd Chiari Syndrome. It is also recommended as a prime imaging procedure to determine alternative venous pathways, stent and vessel patency following intervention.**


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## INTRODUCTION

Primary Budd – Chiari syndrome is a curable form of portal hypertension. In India, it is etiopathologically distinct from the west[1-4] and constitutes 7 to 8 % of all cases of portal hypertension Early diagnosis and prompt intervention can prevent long-term complications of cirrhosis and hepatocellular carcinoma[5]

Chronic obstruction of hepatic venous outflow results in opening of potential collateral channels within the liver and along side the vena cava. New collateral veins drain the obstructed inferior vena cava into the superior vena caval territory. Portal vein and its tributaries may dilate and in severe cases there may be a reversal of flow, which would then serve as an outflow rather than an inflow tract.

In an earlier study, we had reported the role of Real Time (B mode) ultra sonogram (RT US) in pre-operative diagnosis and postoperative follow-up for graft patency in Coarctation of Inferior Vena Cava (CIVC).[6] B-mode ultrasound though ideal for screening neither provides information on the hemodynamics, noror the pressure gradient across the obstructed segment, the latter information being crucial for management. Decompression of an obstructed segment, showing low or no venous flow would be unproductive.

Doppler ultrasonography is more informative than B-Mode US in Budd-Chiari syndrome. It provides additional circulatory hemodynamic information within the hepatic vein, portal vein and also the vena cava[7] The IVC and hepatic veins normally show phasic flow towards the heart. The phasic variations with respiration and right atrial events are reflected in the hepatic venous flow, which are responsible for the characteristic "signature" pattern. When the IVC is obstructed adjacent to the opening of the hepatic veins, the phasic oscillations fail to be reflected, resulting in a flat waveform in the hepatic veins, a major criterion for diagnosis of BCS. In the absence of a pressure

gradient, the flow across the obstructed segment is continuous. In partial obstruction, the blood flow is turbulent.

The aim of the study was to evaluate color-flow Doppler imaging in primary BCS, to identify characteristic vascular patterns with an emphasis on the alternative pathways that predominate in various types of BCS.

### *Methodology:*

Ninety-seven patients with clinical features of Budd Chiari Syndrome (primary and secondary) underwent color Doppler imaging using Core vision pro with inbuilt facility for analyzing the Doppler indices. The liver size, echo texture, size of the caudate lobe and spleen, dimensions of the portal vein, patency of the three major hepatic veins at their junction with IVC and site of entry of HV in relation to the obstructed segment of IVC were recorded. The collaterals (alternative pathways) were classified as intrahepatic comma shaped, angular, sub hepatic and porto systemic. The IVC was traced up to its entry into the right atrium and evaluated both in longitudinal and transverse sections. The suprahepatic segment of IVC was screened for the site, the type (membranous/ stenotic) and length of obstruction (long/ short segment) and for stasis thrombus proximal to the obstruction.

Doppler waveforms were obtained from within the portal vein, retro hepatic and suprahepatic IVC, major hepatic veins, communicating vessels between the hepatic veins and intrahepatic collaterals. Phasic variations with respiration, and type of flow: turbulent, flat/ no wave or reversal of flow[8] in the major HV and IVC were recorded. Doppler study was correlated with angiography in 62 patients. This included 17 patients with type I BCS, 4 with type II BCS and 41 patients with type III BCS.

## RESULTS

Seventy one (73.19 %) of the 97 patients with BCS had isolated obstruction of the major hepatic vein and or obstruction of the suprahepatic segment of the IVC. The remaining 26 (26.80 %) on Doppler had secondary compression (mild to severe) of the HV or the IVC by a cirrhotic nodule, tense ascites or intrinsic obstruction due to an invading hepatoma and were excluded from the analysis.

Based on the site of obstruction, patients were categorized as

Type I – Obstruction to the major hepatic veins alone – 17 (23.94 %) (Fig 1)

Type II – Obstruction to the suprahepatic IVC alone – 13 (18.30%) (Fig 2)

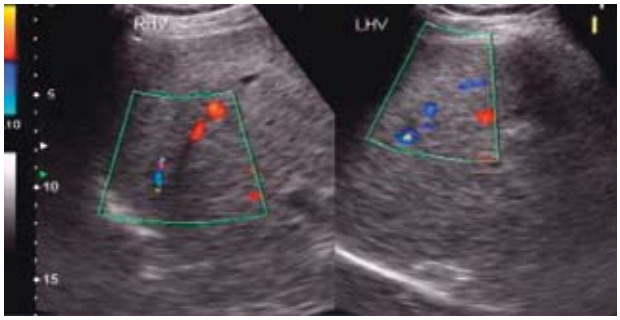


Fig 1. Occlusion of right and left hepatic veins on color Doppler

Type III – Combination Obstruction to the major hepatic veins and IVC – 41 (57.74%)

Liver was enlarged uniformly in 61% in patients with hepatic vein involvement either in isolation or in combination with suprahepatic IVC obstruction; 56.3% had caudate lobe hypertrophy. One fifth of the patients had a shrunken liver. The dimensions of splenoportal axis was normal in more than three fourths of patients with BCS and the flow was hepatopetal in all but ten (14.1) patients. Doppler correlated with angiogram in 56 out of 62 patients (90.3%). Doppler characteristics in the three types of BCS are given in Table 1.

### *Type I BCS*

Of the 17 patients with Type I BCS, all the three major hepatic veins were occluded in 7 (47.2%); isolated obstruction to the right, middle or the left HV in 3(17.6%), 1(5.9%) and 2 (11.8%) patients respectively. Four had combination obstructions of more than one major HV (23.5%).

Color Doppler imaging with pulsed wave Doppler study defined an abnormal waveform in one or more HVns in all these patients (Fig). The spectral waveforms were flat and

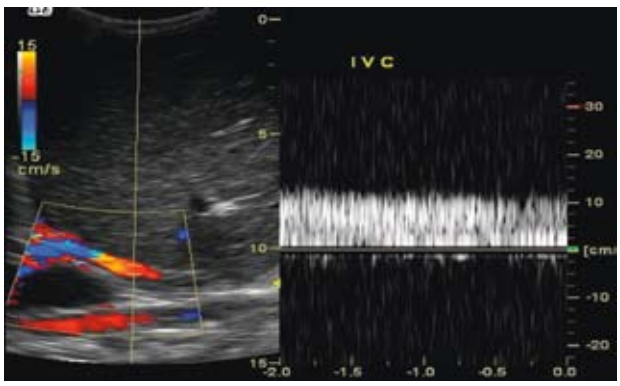


Fig 2. Inferior vena cava obstruction on color Doppler

**Table 1**. Doppler characteristics in the three types of BCS (Numbers in parentheses indicate percentages)

| Feature | Type I (17) | Type II (13) | Type III (41) |
|---|---|---|---|
| Liver | | | |
| Normal | 3 (17.6) | 7 (53.8) | 4 (9.8) |
| Enlarged | 12 (70.6) | 2 (15.4) | 29 (70.7) |
| Shrunken | 2 (11.6%) | 4 (30.8) | 8 (19.5) |
| Caudate lobe hypertrophy | 11 (64.7%) | 3 (23.1) | 26 (63.4) |
| Hepatic vein occlusion | | | |
| All three veins | 7 (47.1) | - | 25 (61) |
| 2 veins | 4 (23.5) | - | 7 (17.1) |
| 1 vein | 6 (35.2) | - | 9 (21.6) |
| IVC occlusion | | | |
| Web | - | 8 (61.5) | 6 (14.6) |
| Thrombus | - | 1 (7.7) | 0 |
| Stenosis/ coarctation | - | 4 (30.8) | 35 (85.4) |
| Normal splenoportal axis | 13 (76.5) | 9 (69.2) | 32 (78) |
| Flow pattern across occluded HV/ IVC | | | |
| Turbulence | 0 | 6 (46.2) | 31 (75.6) |
| Uniphasic | 0 | 4 (30.8) | 0 |
| Aphasic | 16 (94.1) | 1 (7.7) | 3 (7.3) |
| Reversal of flow | 1 (5.9) | 2 (15.4) | 7 (17.1) |
| Alternate pathways (collaterals) | | | |
| Intrahepatic “comma” collaterals | 11 964.7) | 1 (7.7) | 31 (75.6) |
| Inferior right hepatic vein | 4 (23.5) | 1 (7.7) | 9 (21.9) |
| HV to subcapsular veins | 1 (5.9) | 0 | 0 |
| No intrahepatic collaterals | 1 (5.9) | 11 (84.6) | 1 (2.4) |
| Correlation with angiography | 14/17 (82.35) | 3/4 (75) | 39/41 (95.1) |

essentially aphasic in 16 (94.11%) and were reversed in one. In 11 patients (64.7%) there was comma - shaped collaterals (Fig 3) between the adjacent hepatic veins. In one patient a communication was seen between the obstructed HVns and the sub capsular systemic veins. The right inferior hepatic vein (Fig 4) was prominent in 4 patients (23.52%).

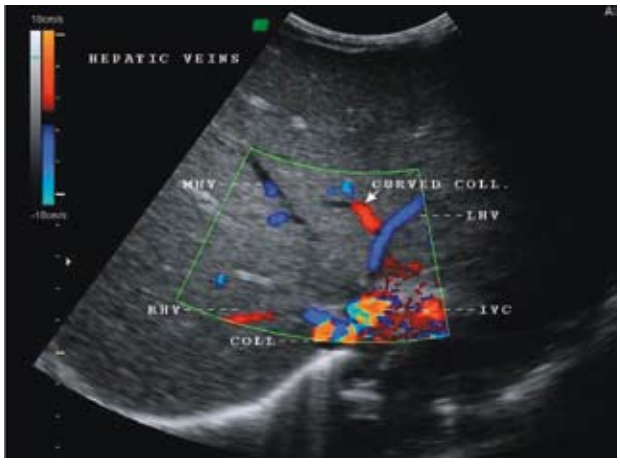


Fig. 3 Comma shaped intrahepatic curved collaterals

Doppler findings correlated with angiography in 14 patients (82.4%). In 3 patients (17.6%) with occluded Hepatic Veins on Doppler, angiography revealed patency of these veins. At 6-months follow-up after angioplasty/ stent placement the Hepatic Vein remained patent in 8 (47.05%) and occluded in 3 (17.64%). Six patients (42.9%) were lost to follow-up.

### *Type II BCS*

Of the 13 patients with Type II BCS, a web or membrane was responsible for the obstruction in 8 (61.53%); the lumen was narrowed or stenotic in 4(30.8%) and one patient had a long segment thrombus. There was a stasis thrombus proximal to the site of obstruction in one (7.96%).

Doppler study showed no flow across the obstructed segment in one, turbulent in 6 (46.15%), slow uniphasic in 4 (30.76%) and reversal of flow in 2 (15.4). IRHV was prominent in one patient. One patient had prominent intrahepatic collaterals (7.69%). The blood flow in the portal vein was hepatopetal in all (84.6%) but two patients.

Angiogram was done in 4 patients. Doppler findings correlated with angiogram in all cases except one (7.6%).

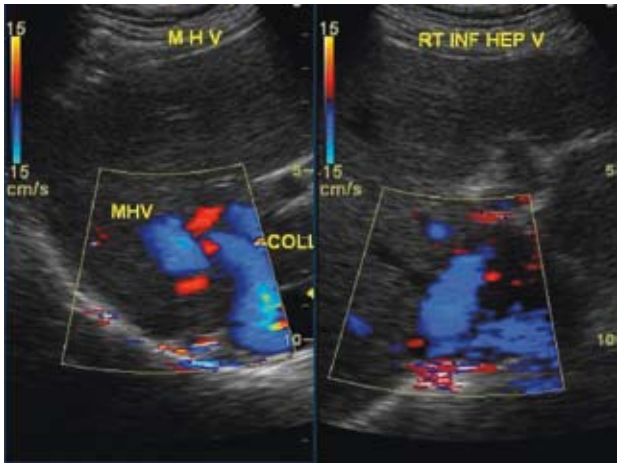


Fig 4.Collateral drainage through the right inferior hepatic vein

Luminal narrowing on Doppler was found to be due to an extrinsic compression by a cirrhotic nodule in this patient. At six months following intervention the IVC remained patent on Doppler in the remaining three patients.

### *Type III BCS*

Majority of the patients i.e. 41 patients (57.4%) had Type III BCS. All the three major HV were involved in 25 (61%), in isolation in 7 (17.1%) and in combination in 9 (21.95%). The suprahepatic IVC was stenotic in 26 (63.41%), membrane obstruction was present in 6 (14.63%) and a coarctated segment was noticeable in 9 (22%).

Doppler showed no flow across the obstructed segment in 3 (7.31%), flow was turbulent in 31(75.60%) and was reversed in 7 (17.1%). The flow in the portal vein was hepatopetal in 34 (82.9%).

All patients underwent an angiogram and the findings at angiography correlated with the Doppler in all but two patients, who were found to have a cirrhotic nodule compressing the HV/ IVC. Twenty-six patients had HV and IVC angioplasty with or without stent placement. The remaining 13 had a cavoatrial bypass. One patient with an occluded Cavo atrial PTFE graft had an IVC angioplasty. Doppler screening for graft patency at 6 monthly intervals for one year revealed a patent hepatic vein and IVC lumen in 34 patients. In 3 the stent had occluded and re-stenting was done. Two patients were lost to follow up.

## DISCUSSION

The diagnosis of BCS with major hepatic vein obstruction is clinically difficult and often mistaken for cirrhosis liver with refractory ascites. In those with inferior vena cava obstruction, venous congestion in the lower limbs is mistaken for a filarial limb and it is not uncommon to come across patients who have undergone repeated venous stripping for varicose veins. [9]

The combination of B mode ultra sonogram, pulsed wave Doppler and color Doppler imaging has revolutionized the understanding of the vascular changes in the hepatic venous outflow tract (HVOT) as well as in the portal vein in Budd Chiari syndrome.[10] Diagnosis of BCS based on waveform at Doppler imaging have been popularized by Ohta et al.[8] In the present study, we were able to classify the HVOT obstruction into three types based on the obstruction to the major HV and / or suprahepatic IVC obstruction.

Duplex Doppler sonography improved the diagnostic yield of obstruction of the major hepatic veins by 40% in the series reported by Chawla et al.[7] The authors reported abnormal waveform in one or more HV. No flow and uniphasic flow was noted based on the nature of blocked vein.[7] In the present series 17 (23.94 %) patients had major HV occlusion. Prominent IRHV was a major intrahepatic vessel in our series with Type I and Type III BCS and was inconspicuous in Type II obstruction.

Color Doppler imaging is a useful investigation in the initial diagnosis of veno-venous shunts associated with Budd-Chiari Syndrome. [11] It is sensitive in identifying irregular, compressed, and otherwise abnormal hepatic veins. These include intrahepatic collaterals referred to as 'comma shaped collaterals' a common alternative pathway in primary HV block.[11] These vessels are easily detected by color Doppler imaging and appear as sine en quo for diagnosis of BCS. [12] Venous spectral Doppler wave forms in the veno venous shunts are flattened and was aphasic in all cases. A slow or absent flow was seen in the obstructed veins as well as in the collaterals. Similar observations were made by Steingruber et al.[11]

Unlike the west, majority of patients in South Asian countries and South Africa have either membrane obstruction of the suprahepatic segment of the IVC or a coarcted obstructed segment. While Real Time Ultra sonogram has been used to initially screen patients with primary BCS, Doppler provides added information on the direction of blood flow across the obstructed segment, providing an insight to successful outcome after decompression. Doppler in addition, in our study as well in other series[13] has proved to be a simple noninvasive investigative tool for follow-up for vessel patency following intervention.

Angiogram correlations with Doppler study have been widely reported. Intrahepatic collaterals, direction of the blood flow in the hepatic vein was better brought out on Doppler compared to angiogram.[13] Angiogram seldom gives information on direction of blood flow; however, pressure gradient across the obstructed segment can predetermine the efficiency of balloon dilation and is a recommended procedure before intervention; greater the pressure gradient, better the outcome. The outcome was poor when the gradient was absent on reversed. Color Doppler sonography, in our series was useful in demonstrating the direction of blood flow through interconnecting vessels and in evaluating the patency of the HV and IVC after treatment. Invasive angiogram can thus be circumvented in follow-up of patients. Millner et al[14] and Lee et al[13] made similar observations. The alternative pathways of veno venous shunt, prominent IRHV and the direction of blood flow in the portal vein were better outlined using Doppler, wherein venogram was less informative.

In conclusion, color Doppler imaging is an excellent technique for qualitative data on flow direction and pattern, thereby contributing significantly to the diagnosis. It is the choice of investigation in the initial evaluation of patients with suspected primary Budd Chiari Syndrome. It is also recommended as a prime imaging procedure to determine alternative venous pathways, stent and vessel patency following intervention.

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