Ultrasonic Evaluation of Spontaneous Portosystemic Shunts in Patients with Budd-Chiari Syndrome

Ji-Chen Wang1, Shi-Feng Cai2, Chen Su1, Hui-Li Fan3, Yong-Hao Gai1,*, Guang-Chuan Wang4 and Li Chen1

1Department of Ultrasound, Shandong Provincial Hospital Affiliated to Shandong University, Jinan, China
2Department of Radiology, Shandong Provincial Hospital Affiliated to Shandong University, Jinan, China
3Department of Ultrasound, Heze Municipal Hospital of Shandong Province, Heze, China
4Department of Gastroenterology, Shandong Provincial Hospital Affiliated to Shandong University, Jinan, China

*Corresponding author: Department of Ultrasound, Shandong Provincial Hospital Affiliated to Shandong University, 324 Jingwu Road, Jinan, China. Fax: 86-53168773689, Email: gailaoshitougao@163.com

Received 2020 April 12; Revised 2020 October 25; Accepted 2020 October 27.

Abstract

Background: Spontaneous portosystemic shunts (SPSS) are one of the hallmarks of Budd-Chiari syndrome (BCS). Ultrasound can accurately show the location and type of portosystemic collaterals.

Objectives: To study the sonographic feature of SPSS in patients with BCS and to evaluate differences in the main portal vein diameter among multiple types of portosystemic shunts.

Patients and Methods: Ultrasonographies of 44 patients with SPSS among 352 BCS patients between June 2000 and November 2015 were reviewed retrospectively. The SPSS in 44 BCS patients were first detected by ultrasound and then confirmed via digital subtraction angiography (DSA), computed tomography angiography (CTA) or magnetic resonance venography (MRV). The location, course, diameter and hemodynamics of the spontaneous portosystemic shunts were observed by ultrasound. In addition, one-way analysis of variance (ANOVA) was performed to evaluate the difference in the main portal vein diameter between the different shunt types.

Results: The blood drainage patterns of SPSS in 44 of 352 patients with BCS were classified as the following five types: portal-umbilical shunts (15/44), portal-hepatic shunts (11/44), portal-accessory hepatic shunts (6/44) (the accessory hepatic veins included the inferior right hepatic vein and the caudate lobe vein), splenorenal shunts (8/44) and main portal vein-inferior vena cava shunts (4/44). The corresponding hemodynamics of the five types mentioned above were obtained. Main portal vein-inferior vena cava shunts had a significantly larger mean portal trunk diameter compared with all other types (P < 0.05 for all comparisons). In addition, the mean portal trunk diameters in portal-umbilical shunts and portal-hepatic shunts were obviously larger than that of splenorenal shunts (P < 0.05), while there were no statistically significant differences between the other types.

Conclusion: Spontaneous portosystemic shunts are not rare in patients with BCS. Ultrasound is a reliable means for their diagnosis and it offers substantial information for use in clinical treatment.

Keywords: Portosystemic Shunts, Ultrasound, Budd-Chiari Syndrome

1. Background

Budd-Chiari syndrome (BCS) is a group of disorders that are characterized as hepatic venous outflow tract obstruction, regardless of the mechanism of obstruction, which can be located at the level of the hepatic venules, the large hepatic veins and the inferior vena cava or the right atrium (1-3). Obstruction caused by cardiac and pericardial diseases and sinusoidal obstruction syndrome is not considered BCS (3, 4). The impaired hepatic venous flow caused by thrombosis or structural compression will lead to dramatically increased sinusoidal pressure, which results in hepatic venous congestion, ischemic injury, and eventually liver cirrhosis (1). Depending on the duration and severity of the disease, BCS can be categorized as fulminant (5%), acute (20%), and subacute or chronic BCS (60%) (2). In patients with fulminant or acute liver disease, venous collaterals have not yet been established, whereas several types of collateral circulation are seen in subacute or chronic forms in an attempt to decompress the hepatic sinusoidal pressure (1). The collateral circulation can be classified as intrahepatic, extrahepatic and portosystemic pathways (5). Currently, spontaneous portosystemic...
shunts (SPSS) in patients with BCS have not been sufficiently investigated by ultrasound. In this study, the ultrasonic features of spontaneous portosystemic shunts in patients with BCS were investigated.

2. Objectives

In this study, we aimed to assess the sonographic feature of SPSS in patients with BCS and to evaluate the differences in the main portal vein diameter among multiple types of portosystemic shunts.

3. Patients and Methods

3.1. Patients

All procedures were performed in accordance with the ethical standards of the responsible committee on human experimentation and with the 1964 Declaration of Helsinki and later versions. Written informed consent from the patients was waived. Ultrasonograms of 352 chronic BCS patients from June 2000 to November 2015 were retrospectively studied. Diagnoses of BCS were confirmed via digital subtraction angiography (DSA) and ultrasonography (US). The only criterion for inclusion in this study was adequate visualization of spontaneous portosystemic shunts on ultrasonogram. However, some types of SPSS (such as coronary venous collaterals) that may not be adequately visualized by ultrasound due to excessive intestinal gas, ascites and technical difficulty were excluded from this study. All sonograms were acquired by an expert with more than 20 years of experience in ultrasound investigation and further evaluated by two expert sonographers with 7 and 10 years of experience with consensus agreement. Of these 352 patients, 44 patients were found to have SPSS, which were first detected by ultrasound and then confirmed via DSA, computed tomography angiography (CTA) or magnetic resonance venography (MRV) after 1 to 2 weeks.

3.2. Ultrasonic Examination

Ultrasonography was performed using an HDI 3500 (ATL-Philips, Bothell, WA, USA), an Envisor HD system (Philips Healthcare, Best, The Netherlands), or a Logiq E9 (GE Healthcare, Waukesha, WI, USA) with 3 to 6 MHz convex transducers. All examinations were performed early in the morning after patients had fasted more than 8 hours and all patients were scanned in the supine or left recumbent position. First, the courses of portal veins and splenic veins were carefully observed to determine whether there was collateral circulation (portosystemic venous shunts) between the portal veins and the inferior vena cava, hepatic veins, and accessory hepatic veins or between the splenic veins and the left renal veins. In the presence of SPSS, the diameter of the corresponding vessels was measured during normal respiration, and the venous shunts were classified. Doppler sonography was used to observe and record the blood flow direction and velocity within the portal veins. Doppler gain and wall filtering were optimized for the lumen without overpainting or aliasing. The Doppler angle of incidence was less than 60°, and the sample volume was adjusted to include at least one third of the lumen’s diameter. The mean velocity was calculated by integrating the time-averaged value based on the Doppler spectrum. The maximum diameter of the portal trunk and the mean portal flow velocity were measured three times, with the average values used for analysis.

3.3. Statistical Analysis

Continuous quantitative variables are expressed as the mean ± standard deviation or the median (first quartile-third quartile); whereas categorical variables are summarized as counts (percentages). Normality was tested using the Shapiro-Wilk test. One-way analysis of variance (ANOVA) was performed to evaluate the difference in portal vein diameter, and multiple comparisons between the groups were performed using the LSD-t-test. P values < 0.05 were considered statistically significant. The statistical values were assessed using SPSS version 23.0 (IBM, Chicago, IL, USA).

4. Results

4.1. Clinical Information of 44 BCS Patients with SPSS

The forty-four patients (18 male, 26 female) had a mean age of 46.3 ± 9.4 years, ranging from 27 to 65 years. Among all patients, the median time from the first clinical symptom to diagnosis was 36 months (first quartile-third quartile, 8 - 91 months). The main clinical symptoms were right upper abdominal distension, varicose veins on the abdominal wall and edema of the lower extremities. Five patients had a history of upper gastrointestinal bleeding, and 2 patients without definite symptoms were found during the health examination in our hospital. Clinical and demographic characteristics of the study patients are shown in Table 1.
The classifications of Spontaneous Portosystemic Shunts

SPSS were detected in 44 of the 352 BCS patients by ultrasound and confirmed by DSA, CTA, or MRV. According to the blood drainage patterns of SPSS, the 44 cases were classified as the following five types:

1) Portal-Umbilical Shunts (15 cases): The ultrasonic feature was that the umbilical veins were recanalized. Blood from the left sagittal branch of the portal vein flowed to the umbilical veins and then to the superficial periumbilical epigastric veins. The superficial periumbilical epigastric veins were tortuous and dilated (Figure 1). The mean diameter of 15 recanalized umbilical veins was 0.88 ± 0.18 cm, and the average velocity was 18.50 ± 3.21 cm/s; the average diameter of the main portal veins was 1.36 ± 0.16 cm. Among 15 patients, 14 exhibited hepatopetal blood flow in the main portal veins, with an average velocity of 12.71 ± 2.94 cm/s, and the rest exhibited hepatofugal blood flow, with a velocity of 6.22 cm/s.

2) Portal-hepatic shunts (11 cases, including six cases with portal vein-left hepatic vein shunts, 4 cases with portal vein-right hepatic vein shunts, and 1 case with portal vein-middle hepatic vein shunts): Sonographic findings revealed that the portal venous blood flowed through single or multiple communications into the hepatic veins mentioned above or onward to the other intercommunicating hepatic vein, and finally into the inferior vena cava (IVC) (Figure 2); the average diameter of the main portal veins was 1.29 ± 0.15 cm. Among 11 patients, 9 exhibited hepatopetal blood in the main portal veins, with an average velocity of 19.30 ± 2.57 cm/s; 1 exhibited hepatofugal blood, with a velocity of 7.13 cm/s; and the remaining cases exhibited hepatopetal and hepatofugal blood simultaneously.

3) Portal-accessory hepatic shunts (the accessory hepatic veins include the inferior right hepatic vein and caudate lobe vein) (six cases, including three cases with portal vein-inferior right hepatic vein shunts, two cases with portal vein-caudate lobe vein shunts and one case with portal vein-inferior right hepatic vein + caudate lobe vein shunts): Sonographic features were roughly consistent with those of portal-hepatic venous shunts. Portal venous blood flowed through the collateral circulation into the accessory hepatic veins and then into the inferior vena cava (Figure 3). The average diameter of the main portal veins was 1.23 ± 0.12 cm. All patients had hepatopetal blood flow in the main portal veins, with an average velocity of 16.30 ± 2.42 cm/s.

4) Splenorenal Shunts (Eight Cases): The average diameter of the main portal veins was 1.12 ± 0.14 cm. The splenic veins at the splenic hilus were dilated and tortuous, with an average diameter of 1.41 ± 0.16 cm. Hepatofugal main portal venous blood flow was found in all eight patients, with an average velocity of 7.0 ± 1.27 cm/s. Retrograde blood flow was also found in the splenic vein posterior to the pancreas. Multiple-section scanning revealed that the branches of the splenic veins anastomosed with the left renal veins through communicating branches (Figure 4). Color Doppler showed that the blood within the main portal veins reversed to the splenic veins and then continued to the left renal veins via communicating branches.

5) Main Portal Vein-Inferior Vena Cava Shunts (Four Cases): Abnormal directional flow was detected in all patients. Among them, hepatofugal blood in the main portal veins was found in three patients, and bidirectional (hepatopetal and hepatofugal) blood flow was found in one patient. Hepatofugal blood in the main portal veins flowed into the inferior vena cava through the collateral circulation (Figure 5). The main portal veins were obviously dilated, with an average diameter of 1.60 ± 0.16 cm. The portal hemodynamic characteristics of the 44 BCS patients are shown in Table 2.

No combined shunts were detected in the 44 BCS patients. Dilated hepatic arteries with accelerated blood flow were detected by ultrasound in all three patients with hepatofugal or bidirectional blood flow of the main portal veins in the first two types. Main portal vein-inferior vena cava shunts had a significantly larger mean portal trunk diameter compared with all other shunt types (P < 0.05 for all comparisons). In addition, the mean portal trunk diameters in portal-umbilical shunts and portal-hepatic shunts were obviously larger than that of splenorenal shunts (P < 0.05), while there were no statistically significant differences between the other types. Statistical analysis results are reported in Table 2.

| Table 1. Patient Characteristics (N = 44)* |
|-----------------|-----------------|
| Variables       | Values          |
| Age, y          | 46.3 ± 9.4 (27 - 65) |
| Sex             |                 |
| Male            | 18 (40.9)       |
| Female          | 26 (59.1)       |
| The obstruction level in BCS cases |                |
| HVs             | 4 (9.1)         |
| IVC             | 0 (0)           |
| HVs and IVC co-existed | 40 (90.9) |

Abbreviations: BCS, Budd-Chiari syndrome; HVs, hepatic veins; IVC, inferior vena cava; SD, standard deviation; y, year.

*Values are expressed as the No. (%) or mean ± SD (Range).
4.3. The Other Major Sonographic Findings in 44 Patients with Spontaneous Portosystemic Shunts

Among the 132 total hepatic veins of 44 patients in this group, there were only 13 hepatic veins with unobstructed lumens, among which there were five hepatic veins with tortuous lumens caused by the liver parenchyma extrusion that remained. There were 119 abnormal hepatic veins.
Figure 2. Portal-hepatic shunt. A, Sonograms show that the blood of the main portal vein (MPV) flows to the communicating vessels (CVs) via the shunt. The diameter of the shunt is 0.53 cm, and the mean flow velocity is 30.5 cm/s; B, sonogram shows the intrahepatic CVs from the shunt to the left hepatic vein (LHV); C, sonogram shows the inlet of the LHV, with an accelerated mean flow velocity of 171.3 cm/s. Membranous stenosis is also found in the inlet (arrow); D, magnetic resonance venography (MRV) shows that blood from the portal vein (PV) flows to the CVs via the shunt; E, blood from the CVs drained into the inferior vena cava (arrow) via the LHV; F, venography following placement of the guidewire in the LHV illustrates the presence of a portal-hepatic venous shunt and shows complete occlusion of the retrohepatic segment of the inferior vena cava (arrow).

(including stenosis and occlusion of the lumen), accounting for 90.2% (119/132) of all hepatic veins, with an average of 2.7 (119/44) per patient. Except for the 58 lumens that were stenosed to different degrees, there were 58 lumens with complete occlusion and 3 lumens that had disappeared due to thrombosis, accounting for 51.3% of the veins (61/119). Among 44 patients, there were only 4 with a patent inferior vena cava (IVC), while the remaining 40 patients had mixed BCS, in which obstructed HVs and an obstructed IVC co-existed in the same patient. Collateral pathways of the inferior vena cava were also detected in each patient with an obstructed inferior vena cava by DSA, CTA or MRV. The blood from the obstructed inferior vena cava was drained into the superior vena cava via the ascending lumbar vein, left renal vein or left inferior phrenic vein, as previously reported (5, 6).

5. Discussion

In this group of patients with chronic BCS, portal-umbilical shunts caused by the reopened umbilical vein were the most common type and the incidence of patent...
Figure 3. Portal-accessory hepatic venous shunt. A, Color Doppler sonogram shows that the blood from the portal vein (PV) flows to the left hepatic vein (LHV) through the communicating vessel (CV) (right hand image) and onward to the middle hepatic vein (MHV) (left hand image) via communicating branches (arrow); B, color Doppler sonograms show that blood drains into the inferior vena cava (IVC) by the dilated inferior right hepatic vein (IRHV) (left hand image) and caudate lobe vein (CLV) (right hand image); C, diagram of a portal-accessory hepatic shunt shows segmental occlusion of the LHV, MHV, and IVC, and complete occlusion of the right hepatic vein (RHV). Blood from the left portal vein (LPV) flows into the LHV and MHV through the CVs and then drains into the IVC through the IRHV and CLV.

paraumbilical vein was 4.3% (15/352). Sonograms showed that the recanalized umbilical vein runs from the umbilical part of the left branch of the portal vein to the liver margin, and drains into the anterior abdominal veins. Clini-
cally, this route is characterized by a "Medusa’s head" appearance, i.e., a network of dilated veins around the umbilicus (7). Because the course of the umbilical vein is roughly towards the transducer, maintaining a low angle (< 60°), this shunt can be accurately diagnosed with a transabdominal probe.

Previous studies have confirmed that few communicating branches exist between portal veins and hepatic veins in normal individuals (8). Among patients with chronic BCS, portal hypertension due to hepatic venous outflow tract obstruction is found in almost all patients. The increasing pressure in the portal vein makes the small communicating branches gradually dilate and eventually develop into enlarged collaterals, namely, portosystemic shunts. Therefore, a large amount of portal vein blood can be drained through the intrahepatic portosystemic venous shunts rather than the hepatic sinusoid, directly into the hepatic vein and accessory hepatic vein (the caudate lobe vein or inferior right hepatic vein), and then enter the systemic circulation. The spontaneous shunts between the portal vein and hepatic vein, as well as the accessory hepatic vein, are similar in terms of hemodynamics and ultrasonography; i.e., the left or the right branch of the portal vein connects the hepatic vein or accessory hepatic vein via communicating branches to the inferior vena cava. In accordance with our previous report regarding the high incidence of hepatic vein obstruction in BCS patients, dilated collateral vessels between the hepatic vein and hepatic vein or accessory hepatic vein were observed in most patients (9, 10). Therefore, portal-hepatic and portal-accessory hepatic portosystemic shunts would easily be mistaken as the collateral vessels mentioned above. Finding the communication vessel directly connecting the branches of the portal vein to the hepatic vein or accessory hepatic vein is the key to the diagnosis of the two types.

By observing multiple real-time sections, we found
that the communicating branches of the splenic vein anastomose with the left renal vein in the retroperitoneal space, which is the direct sign needed to diagnose spontaneous splenorenal shunts. However, there are numerous dilated and tortuous vessels in the region of the splenic and left renal hilum, and they are sometimes difficult to distinguish from the splenorenal shunt. Observation of the hepatofugal blood flow of the portal vein and splenic
which may be seen as a result of portal venous compromise had a dilated hepatic artery with accelerated blood flow, three patients with the first two types. All of these patients gal or bidirectional portal venous flow was exhibited in the splenic vein and left renal vein. In addition, hepatofu-
municating branches were simultaneously found between tal vein of the fourth type had hepatofugal flow, large com-
flow did not decrease obviously. Although all the main por-
branches, but the velocity of hepatopetal portal venous
had the presence of large portosystemic communicating
fifth type. In addition, the former three types not only
four types mentioned above was smaller than that of the
this group.

dilation of the main portal veins among the former
types was mentioned above was smaller than that of the
fifth type. In addition, the former three types not only
had the presence of large portosystemic communicating
branches, but the velocity of hepatopetal portal venous
flow did not decrease obviously. Although all the main por-
venous branches were simultaneously found between
the splenic vein and left renal vein. In addition, hepatofu-
gal or bidirectional portal venous flow was exhibited in
three patients with the first two types. All of these patients
had a dilated hepatic artery with accelerated blood flow,
which may be seen as a result of portal venous compromise
in portal hypertension. The above ultrasonic features illustrated that the amount of shunted blood in the former four types was larger. Therefore, for the first four types of portosystemic shunts, we can mainly treat the lesions of the hepatic vein and inferior vena cava, whereas the last type requires a transjugular intrahepatic portosystemic shunt (TIPS) due to inability to provide sufficient hepatic decompression (12).

In summary, sonographic examination is a reliable way to observe and record the course, location and hemodynamics of the portosystemic venous shunts, which can be helpful for establishing therapeutic plans.

Footnotes

Authors’ Contributions: Yong-Hao Gai and Shi-Feng Cai conceived and supervised the study. Ji-Chen Wang and Chen Su analyzed the patient data. Ji-Chen Wang and Li Chen drafted the manuscript. Hui-Li Fan and Guang-Chuan Wang prepared figures.

Conflict of Interests: The authors declare that there are no conflicts of interest.

Ethical Approval: All procedures were performed in accordance with the ethical standards of the Responsible Committee on human experimentation and with the 1964 Declaration of Helsinki and later versions.

Funding/Support: This study was supported by the Shandong Provincial Science and Technology Development Project Foundation of China (nos. 2013GSF11827 and 2018GSF118105018), the Health Care and Family Planning Technology Development Plans of Shandong Province (nos. 2013WS0108) and the Key R and D Program of Shandong Province (nos. 2018GSF118178).

Informed Consent: Written informed consent from the patients was waived.

References

1. Menon KV, Shah V, Kamath PS. The Budd-Chiari syndrome. N Engl J Med. 2004;350(6):578-85. doi: 10.1056/NEJMoa020282. [PubMed: 14762185].
2. Senzolo M, Cholongitas EC, Patch D, Burroughs AK. Update on the classification, assessment of prognosis and therapy of Budd-Chiari syndrome. Nat Clin Pract Gastroenterol Hepatol. 2005;2(4):182-90. doi: 10.1038/ncpgasthep0041. [PubMed: 16265183].
3. Janssen HL, Garcia-Pagan JC, Elias E, Menitha G, Hadengue A, Valla DC, et al. Budd-Chiari syndrome: a review by an expert panel. J Hepato-
2002;30(3):364-71. doi: 10.1016/s0168-8278(02)00434-8. [PubMed: 12586305].
4. DeLeve LD, Valla DC, Garcia-Tsao G. American Association for the Study of Liver Diseases. Vascular disorders of the liver. *Hepatology*. 2009;49(5):1729–64. doi: 10.1002/hep.22772. [PubMed: 19399912]. [PubMed Central: PMC6697263].

5. Cai SF, Gai YH, Liu QW. Computed tomography angiography manifestations of collateral circulations in Budd-Chiari syndrome. *Exp Ther Med*. 2015;9(2):399–404. doi: 10.3892/etm.2014.2125. [PubMed: 25574205]. [PubMed Central: PMC4280981].

6. Cho OK, Koo JH, Kim YS, Rhim HC, Koh BH, Seo HS. Collateral pathways in Budd-Chiari syndrome: CT and venographic correlation. *AJR Am J Roentgenol*. 1996;167(5):1163–7. doi: 10.2214/ajr.167.5.8911174. [PubMed: 8911174].

7. Morin C, Lafortune M, Pomier G, Robin M, Breton G. Patent parausbilical vein: anatomic and hemodynamic variants and their clinical importance. *Radiology*. 1992;185(1):253–6. doi: 10.1148/radiology.185.1.1523319. [PubMed: 1523319].

8. Guo Z, He S, Wu Y. Observation of the anastomoses of intrahepatic veins in normal men. *Hua Xi Yi Ke Da Xue Xue Bao*. 1996;27(3):270–4. [PubMed: 9189060].

9. Gai YH, Cai SF, Guo WR, Zhang CQ, Liang B, Jia T, et al. Sonographic classification of draining pathways of obstructed hepatic veins in Budd-Chiari syndrome. *J Clin Ultrasound*. 2014;42(3):334–42. doi: 10.1002/jcu.22107. [PubMed: 24166504].

10. Song RX, Cai SF, Ma S, Liu ZL, Gai YH, Zhang CQ, et al. Magnetic resonance venography findings of obstructed hepatic veins and the inferior vena cava in patients with Budd-Chiari syndrome. *Korean J Radiol*. 2018;19(3):381–8. doi: 10.3348/kjr.2018.19.3.381. [PubMed: 2973215]. [PubMed Central: PMC5904464].

11. Bandali MF, Mirakhur A, Lee EW, Ferris MC, Sadler DJ, Gray RR, et al. Portal hypertension: Imaging of portosystemic collateral pathways and associated image-guided therapy. *World J Gastroenterol*. 2017;23(30):4735–46. doi: 10.3748/wjg.v23.i30.4735. [PubMed: 28348478]. [PubMed Central: PMC5352913].

12. Bargallo X, Gilabert R, Nicolau C, Garcia-Pagan JC, Ayuso JR, Bru C. Sonography of Budd-Chiari syndrome. *AJR Am J Roentgenol*. 2006;187(3):W33–41. doi: 10.2214/AJR.04.0918. [PubMed: 16794137].