IMAGING PATTERNS OF DOPPLER SONOGRAPHY FOR EVALUATION OF BUDD CHIARI SYNDROME IN CHILDREN

Aysha Anjum,[1] Eisha Tahir,[2] Mamoona Chiragh[3]

ABSTRACT

Background: Current literature on Budd Chiari Syndrome (BCS) in children is limited. **Objective:** To evaluate Ultrasound Doppler imaging as non-invasive and non-ionizing method of detecting children with clinical suspicion of Budd Chiari Syndrome. **Methodology:** This was a cross-sectional study conducted from 1st February 2016 to 31st December 2017. Seventeen patients with clinically suspected BCS of age < 12 years, both gender, were registered from outpatient department of tertiary care unit, over a period of one year. Ultrasound and Doppler sonography was performed in all patients. Visualization of hepatic veins and IVC were noted along with flow and spectral waveform patterns in IVC, hepatic veins and portal veins. Intra-hepatic collaterals, caudate lobe hypertrophy, hepatosplenomegaly and ascites were also documented. The data was entered and analyzed by SPSS version 20. **Results:** The study included 11 (64.70%) female and 6 (35.29%) male patients with a mean age of 1.01±0.55 years. All patients had clinical suspicion of BCS (yellow discoloration of skin or sclera 52.94%, epistaxis 5.88%, abdominal pain 94.11%, distended abdomen 88.23%, palpable liver 64.70%, ascites 88.23%). Ultrasonography showed right, middle and left hepatic veins occlusion 52.94%, 29.41% and 41.17% patients, respectively. Hepatic veins flow was reversed in 29.41% and absent in 52.94% cases. Intrahepatic collaterals, caudate lobe hypertrophy, hepatomegaly, portal hypertension, portal vein thrombosis and partial thrombosis of IVC were seen in 58.82%, 64.70%, 76.47%, 41.17%, 5.88% and 11.76% patients, respectively. Portal vein flow was hepatopetal in 52.94% and hepatofugal in 23.52% cases. **Conclusion:** Doppler sonography is a useful, non-invasive and non-ionizing imaging modality to diagnose and follow-up of patients with Budd Chiari Syndrome.

Keywords: Budd Chiari Syndrome, Non-invasive, Doppler ultrasonography, Hepatopetal, Hepatofugal.

INTRODUCTION

Blockade of the venous vasculature of the liver is denoted as Budd–Chiari syndrome (BCS).[1] Closing off of hepatic veins results in amplified venous pressures that is foundation of ascites development and esophageal, gastric and rectal varices establishment.[2] Obstruction is also responsible of centrilobular necrosis due to ischemia. If this condition persists, hepatomegaly will develop. It is evident as triad of abdominal pain, ascites, and hepatomegaly. It can be fulminant, acute, chronic, or asymptomatic.[1] When obstacle is mainly due to venous process; it is Primary Budd-Chiari syndrome. Membranous obstruction of inferior vena cava (IVC) is the commonest etiology of it in pediatrics.[2] This syndrome is sporadic in children. So, misdiagnoses and postponements in its identification are plentiful. Its prevalence is not well stated in the literature. Existing data are inadequate.[3,4] Imaging execute a vital fragment in the early recognition and assessment of the magnitude of disease in BCS. Timely analysis and interference to alleviate liver congestion is crucial to renovate liver task and lessen portal hypertension. Interventional radiology obliges a significant part in the management of these patients.[5] The confident conclusion of BCS generally necessitates a liver biopsy, proving venous congestion with centrilobular necrosis, or a hepatic venogram displaying block of the hepatic veins. Venography can clearly delineate the nature and severity of an obstruction. However, before ensuing these investigations, a high index of suspicion is obligatory. Most of the times, deranged clotting profile prohibit these conclusive invasive diagnostic procedures.[6] Angiography, right now, is primary diagnostic modality for BCS. However, ultrasonography/Doppler is a valuable way for early evaluation and follow up of these patients. Ultrasonography provides diagnostic proof for BCS in many cases. It is relatively low cost, widely available and noninvasive.[7] The study was conducted to evaluate Ultrasound/Doppler imaging as non-invasive and non-ionizing method of diagnosing children with clinical suspicion of Budd Chiari Syndrome.

METHODOLOGY

This descriptive cross sectional study, included 17 cases of clinically suspected Budd Chiari Syndrome of age less than 12 years, both gender, registered from outpatient department of tertiary care unit of Children Hospital and the Institute of Child Health, Lahore and was carried from 1st February 2016 to 31st January 2017.

1. Department of Radiology, Children Hospital, Lahore, Pakistan.
2. Department of Radiology, Shalamar Hospital, Lahore, Pakistan.
3. Nawaz Sharif Social Security Hospital, Lahore, Pakistan.

Correspondence: Dr. Ayesha Anjum, Assistant Professor of Radiology, Children Hospital, Lahore, Pakistan

E-mail: anjumf@gmail.com **Received:** 15-11-2017 **Accepted:** 10-01-2018

After obtaining written informed consent and clarifying the technique of ultrasonongraphy, Ultrasound and Doppler sonography was performed in 17 patients with clinically suspected Budd Chiari Syndrome by fellow of radiology. Visualization of hepatic veins and IVC were noted along with flow and spectral waveform patterns in IVC, hepatic veins and portal veins. Intra-hepatic collaterals, caudate lobe hypertrophy, hepatosplenomegaly and ascites were also documented. Demographic features like age and sex were noted and statistical analysis was done using SPSS version 20. Ethical approval was sought from hospital ethical committee.

RESULTS

Out of 17 patients, 11 (64.70%) were female and 6 (35.29%) were male with a mean age of 1.01±0.55 years. All patients had clinical suspicion of Budd Chiari Syndrome (history of yellow discoloration of skin or sclera, gastrointestinal or mucosal bleeding, abdominal pain distended abdomen, palpable liver, ascites). Duration of symptoms was from one month to 3 years. Clinical characteristics of patients are shown in table I.

Table I: Clinical characteristics of patients

Clinical characteristics	No. of patients	Percentage
Yellow discoloration of skin or sclera	9	52.94
Pain in abdomen	16	94.11
Abdominal distension	15	88.23
Bleeding (epistaxis)	1	5.88
Palpable liver	11	64.70
Ascites	15	88.23

Table II: Sonographic features in Budd Chiari syndrome

Parameters		No (%)
Narrowing/occlusion of veins	Right hepatic vein	9 (52.94)
	Middle hepatic vein	5 (29.41)
	Left hepatic vein	7 (41.17)
Flow in hepatic veins	Reverse	5 (29.41)
	Absent	9 (52.94)
Intrahepatic collaterals		10 (58.82)
Caudate lobe hypertrophy		11 (64.70)
Hepatomegaly		13 (76.47)
Ascites		15 (88.23)
Portal vein changes	Hepatopetal flow	9 (52.94)
	Hepatofugal flow	4 (23.52)
Portal hypertension		7 (41.17)
Portal vein thrombosis		1 (5.88)
Partial thrombosis of Inferior vena ceva		2 (11.76)

Ultrasonography was performed in all patients. Hepatic veins, portal vein and IVC abnormalities on Grey scale and Doppler sonography along with liver changes are shown in table II.

DISCUSSION

Ultrasound and Doppler signify essential imaging modalities for detecting BCS in children. The imaging results are subject to obstruction level, duration of obstruction and secondary decompensation. Caudate lobe hypertrophy is a striking feature.[8] Obstruction may be manifested by constricting, echogenic thrombus/web or membranes, and transformed flow configurations. Collaterals that cultivate may be intrahepatic or extra hepatic.[9] Signs of hepatic failure are typically perceived in late stages.[10] Recognizing the clinical appearance and imaging findings can aid in attaining the precise diagnosis because an early diagnosis of BCS will influence patient management.[8] The mean age of patients in our study was 1.01±0.55 years while in a study by Nagral A et al, the median age was 22 months.[9] In our study, 64.70%were females and 35.29% were male patients. The female predominance was observed in our study. However, male predominance (62.5%) was reported in a study by Nagral A et al.[9]

In another study by Kumar S et al, 75% were male patients.[10] Millener P et al, documented 61.9% female patients.[11] In a study by Grant EG et al, all patients were female.[12] The duration of symptoms in our study was from one month to three years while in a study by Nagral A et al, the median duration was 13 weeks.[9] In our study, 94.11% patients were presented with abdominal pain while in a study by Cheng D et al, abdominal pain was found in 21% patients.[13] The other symptoms presented in our study were yellow discoloration of skin or sclera in 52.94%, abdominal distension in 88.23% and bleeding (epistaxis) in 5.88% patients. However, Nagral A et al, reported jaundice in 12.5%, abdominal distension in 81% and bleeding (hematemesis, malena, epistaxis) in 25% patients.[9]

In our study, liver was palpable in 64.70% and ascites was clinically detectable in 88.23% cases. Jaffe R et al, reported a case of male infant with ascites present at birth.[14] Hepatomegaly and ascites were detected on ultrasonography in 76.47% and 88.23% cases respectively, in our study. While Nagral A et al, demonstrated hepatic enlargement and ascites in abdomen in 69% and 75% patients respectively. They also reported splenomegaly in 44% cases

which was not found in any of case in our study.[9] In our study caudate lobe hypertrophy was found in 64.70% cases. In a study by Boozari B et al, who evaluated diagnostic and prognostic role of ultrasound in budd chairi syndrome, reported caudate lobe hypertrophy in 67% cases.[15] Baert AL et al, reported caudate lobe hypertrophy in 60% cases.[16] While in a study by Gupta S et al, caudate lobe enlargement was seen in all patients on ultrasound examination.[17] Intrahepatic collaterals were detected on ultrasonography in 58.82% patients in our study while these collaterals were reported in 73% cases by Boozari B et al, 94.6% cases by Chawla Y et al, and 72.72% cases by Kane R et al.[15,18,19]

In our study, occlusion of right, middle and left hepatic veins were found in 52.94%, 29.41% and 41.17% cases, respectively (Figure 2). In a study by Chawla Y et al, who performed Duplex Doppler sonography in 37 consecutive angiographically proven patients with Budd–Chiari syndrome, they reported abnormalities of right, middle and left hepatic veins in 56.75%, 40.54% and 48.64% patients, respectively.[18] In a study by Kane R et al, Doppler sonographic scans identified occlusion of three hepatic veins in 18.18%, two hepatic veins in 36.36% and one hepatic vein in 45.45% cases.[19]In a study by Ralls PW et al, narrowing of hepatic veins were seen in 80% patients.[20]

Reverse flow in hepatic veins was noted in 29.41 % and flow was absent in 52.94% cases in our study. In a study by Ralls PW et al, reverse flow was seen in 40% cases.[20] In a study by Bolondi L et al, Doppler ultrasonographic investigation showed flow in the hepatic veins was completely absent in 25% cases and reversed in 25% cases.[21]

In our study, portal vein hepatopetal flow was detected in 52.94% and hepatofugal flow in 23.52 % cases. While in a study by Chawla Y et al, hepatopetal flow was found in the portal vein of 91.3% patients and flow was hepatofugal in 4.34% patients.[18] Bolondi L et al, showed hepatopetal flow in 87.5% cases.[21] In our study, the portal vein thrombosis was detected in 5.88% patients while it was found in 4.34% cases by Chawla Y et al and 12.5% cases by Bolondi L et al.[18,21] Portal vein hypertension was noted in 41.17% cases and partial thrombosis of IVC was seen in 11.76% patients in our study.

CONCLUSION

Diagnosis is necessary for timely intervention of Budd-Chiari syndrome. So, an efficacious problem-solving approach is of dynamic significance. Ultrasound/Doppler Sonography is a noninvasive, useful and real aid for diagnosis of Budd chiari syndrome.

REFERENCES

1. Menon KN, Shah V, Kamath PS. The Budd–Chiari syndrome. N Engl J Med 2004;350(6):578-85.
2. Janssen HL, Garcia-Pagan JC, Elias E, Mentha G, Hadengue A, Valla DC. Budd–Chiari syndrome: a review by an expert panel. J Hepatol 2003;38(3):364-71.
3. Tripathi D, Sunderraj L, Vemala V, Mehrzad H, Zia Z, Mangat K, West R, Chen F, Elias E, Olliff SP. Long-term outcomes following percutaneous hepatic vein recanalization for Budd–Chiari syndrome. Liver Int 2017;37(1):111-20.
4. Nobre S, Khanna R, Bab N, Kyrana E, Height S, Karani J, Kane P, Heaton N, Dhawan A. Primary Budd-Chiari Syndrome in Children: King's College Hospital Experience. J Pediatr Gastro Nutr 2017;65(1):93-6.
5. Stanley P. Budd-Chiari syndrome. Radiol 1989;170(3):625-7.
6. Bargalló X, Gilabert R, Nicolau C, García-Pagán JC, Bosch J, Brú C. Sonography of the caudate vein: value in diagnosing Budd-Chiari syndrome. AJR 2003;181(6):1641-5.
7. Copelan A, Remer EM, Sands M, Nghiem H, Kapoor B. Diagnosis and management of Budd Chiari syndrome: an update. J Vasc Interv Radiol 2015;38(1):1-4.
8. Parekh J, Matei VM, Canas Coto A, Friedman D, Lee WM. Budd chiari syndrome causing acute liver failure: A multicenter case series. Liver Transpl 2017;23(2):135-42.
9. Nagral A, Hasija RP, Marar S, Nabi F. Budd-Chiari syndrome in children: experience with therapeutic radiological intervention. J Pediatr Gastroenterol Nutr 2010;50(1):74-8.
10. Kumar S, et al. Percutaneous interventional strategies for management of Budd-Chiari syndrome (BCS) in pediatric patients. JVIR 2013;24(4):29-35
11. Millener P, Grant EG, Rose S, Duerinckx A, Schiller VL, Tessler FN, Perrella RR, Ragavendra NA. Color Doppler imaging findings in patients with Budd-Chiari syndrome: correlation with venographic findings. AJR 1993;161(2):307-12.
12. Grant EG, Perrella R, Tessler FN, Lois J, Busuttil R. Budd-Chiari syndrome: the results of duplex and color

Doppler imaging. AJR 1989;152(2):377-81.
13. Cheng D, Xu H, Lu ZJ, Hua R, Qiu H, Du H, Xu X, Zhang J. Clinical features and etiology of Budd–Chiari syndrome in Chinese patients: A single-center study. J Gastroenterol Hepatol 2013;28(6):1061-7.
14. Jaffe R, Yunis EJ. Congenital Budd-Chiari syndrome. Pediatr Pathol 1983;1(2):187-92.
15. Boozari B, Bahr MJ, Kubicka S, Klempnauer J, Ultrasonography in patients with Budd–Chiari syndrome–Diagnostic signs and prognostic implications. J Hepatol 2008;49(4):572-80.
16. Baert AL, Fevery J, Marchal G, Goddeeris P, Wilms G, Ponette E, De Groote J. Early diagnosis of Budd-Chiari syndrome by computed tomography and ultrasonography: report of five cases. Gastroenterol 1983;84(3):587-95.
17. Gupta S, Barter S, Phillips GW, Gibson RN, Hodgson HJ. Comparison of ultrasonography, computed tomography and 99mTc liver scan in diagnosis of Budd-Chiari syndrome. Gut 1987;28(3):242-7.
18. Chawla Y, Kumar S, Dhiman RK, Suri S, Dilawari JB. Duplex Doppler sonography in patients with Budd–Chiari syndrome. J Gastroenterol Hepatol 1999;14(9):904-7.
19. Kane R, Eustace S. Diagnosis of Budd-Chiari syndrome: comparison between sonography and MR angiography. Radiol 1995;195(1):117-21.
20. Ralls PW, Johnson MB, Radin DR, Boswell Jr WD, Lee KP, Halls JM. Budd-Chiari syndrome: detection with color Doppler sonography. AJR 1992;159(1):113-6.
21. Bolondi L, Gaiani S, Bassi SL, Zironi G, Bonino F, Brunetto M, Barbara L. Diagnosis of Budd—Chiari syndrome by pulsed Doppler ultrasound. Gastroenterol 1991;100(5):1324-31.

Article Citation: Anjum A, Tahir E, Chiragh M. Imaging patterns of doppler songraphy for evaluation of budd chiari syndrome in children. JSZMC 2018;9(1):1324-1327.