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Intrahepatic venous collaterals

H. Naganuma,[1] **H. Ishida,**[1] **K. Konno,**[1] **T. Komatsuda,**[1] **Y. Hamashima,**[1] **J. Ishida,**[2] **O. Masamune**[1]

[1]First Department of Internal Medicine, Akita University School of Medicine, 1-1-1 Hondo, Akita, Japan
[2]Department of Radiology, Tokyo Medical College, 6-7-1 Nishishinjuku, Shinjuku-ku, Tokyo, Japan

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Abstract
Background: The aim of this study was to reevaluate the causes and sites of intrahepatic venous collaterals and to determine the role of color Doppler sonography in the diagnosis of this relatively rare vascular abnormality.
Methods: Real-time color Doppler sonography was used to study 21 patients with intrahepatic venous collaterals. The cause, distribution, and clinical manifestations of collaterals were determined, and Doppler waveforms obtained from the collaterals were also analyzed.
Results: First, the causes of intrahepatic venous collaterals were divided roughly into two groups according to the presence or absence of veno-occlusions. The former group included liver tumors (six cases), primary Budd-Chiari syndrome (five cases), and metastatic adrenal tumors invading the inferior vena cava (two cases). The latter group consisted of diaphragmatic hernia (three cases), Osler-Weber-Rendu disease (two cases), and congestive liver (one case). The cause was not determined in two cases. Second, venous collaterals were distributed throughout the entire liver in primary Budd-Chiari syndrome but localized in the other cases. Third, Doppler waveforms of the collaterals were divided into two patterns: flat flow and multiphasic flow. Flat flow pattern was seen in patients with veno-occlusive diseases, and multiphasic flow pattern was seen in patients without veno-occlusive disease.
Conclusion: The relationship between intrahepatic venous collaterals and veno-occlusive diseases has been emphasized in the literature, but the results of our series showed that they occurred under a wide variety of conditions, even without veno-occlusive diseases, including diaphragmatic hernia and Osler-Weber-Rendu disease. The analysis of the Doppler waveforms of the collaterals was useful in differentiating those due to veno-occlusive diseases and those not.

Key words: Ultrasound—Doppler—Hepatic vein—Collaterals—Budd-Chiari syndrome—Liver tumor—Adrenal tumor—Diaphragmatic hernia.

Correspondence to: H. Naganuma

Intrahepatic venous collaterals have been reported as an important finding indicating hepatic venous or inferior vena cava (IVC) obstruction [1, 2], but those associated with other conditions rarely have been reported [3, 4].

Regarding the diagnosis of intrahepatic venous collaterals, hepatic venography was the sole tool, but it has been replaced gradually by noninvasive medical imaging techniques (computed tomography, magnetic resonance imaging, and color Doppler sonography). In fact, with color Doppler sonography, we can easily detect venous collaterals [5–7].

The aim of this paper is, through a retrospective analysis of the results of 21 patients with intrahepatic venous collaterals, to reinvestigate the causes, color Doppler findings, and clinical manifestations of these relatively rare vascular changes and to determine the role of color Doppler sonography in the diagnosis.

Subjects and methods

Twenty-one patients (11 men, 10 women; mean age = 39.6 years, range = 1–83 years) with intrahepatic venous collaterals were studied retrospectively. The final diagnosis of intrahepatic venous collaterals was based on hepatic venographic findings in 14 patients and on typical color Doppler findings in seven patients in whom the communication between the detected collaterals and the neighboring hepatic vein was confirmed by color Doppler sonography.

The collaterals were not known before the sonographic examination in any of the patients.

Table 1. Summary of 21 cases of intrahepatic venous collaterals

Cases	Number of patients	Sex (M:F)	Age range (mean)	Obstructive site[a]	Distribution of collaterals[a]	Doppler signals of collaterals
Primary B-C syndrome	5	4:1	25–57 (39.8)	Suprahepatic IVC (4) MHV (1)	Both lobes (5)	Flat flow
Liver tumor	6	3:3	2–47 (33.2)	Tumor (4) Tumor thrombus (2)	Right lobe (6)	Flat flow
Diaphragmatic hernia	3	0:3	1–10 (4.0)	(—)	Right lobe (3)	Multiphasic flow
Adrenal tumor	2	1:1	57–62 (59.9)	IVC	Right lobe-IVC	Multiphasic flow
Osler disease	2	1:1	47–75 (61.0)	(—)	Right lobe (1) Left lobe (1)	Multiphasic flow
Congestive liver	1	1:0	47	(—)	Right lobe	Multiphasic flow
Unknown	2	1:1	51–83 (67.0)	(—)	Right lobe	Multiphasic flow

[a] Number of cases indicated within parentheses
B-C, Budd Chiari; MHV, middle hepatic vein

We precisely examined each case from the following points of view:

1. clinical findings: symptoms, causes, and presence or absence of diffuse hepatic disease
2. presence or absence of other anomalies in the intrahepatic and extrahepatic vascular system
3. color Doppler findings: distribution of venous collaterals and Doppler waveforms of the collaterals

The subjects fasted for 14–17 h before the sonographic examination.

A Toshiba SSA-270A or SSA 380 A (Tokyo, Japan) color Doppler unit with a 2.5-MHz electronic sector and a 3.75-MHz convex transducer was used. A pulse repetition frequency of 3–6 kHz and a filter of 50 or 100 Hz were used.

The Doppler waveforms of the collaterals were divided roughly into two flow patterns: flat and multiphasic.

Results (Table 1)

The causes of intrahepatic venous collaterals were divided roughly into two groups according to the presence or absence of veno-occlusions. The former group included liver tumors (six cases), primary Budd-Chiari syndrome (five cases), and metastatic adrenal tumors invading the IVC (two cases). The latter group consisted of diaphragmatic hernia (three cases), Osler-Weber-Rendu disease (two cases), and congestive liver (one case). The cause was not determined in two cases.

The six liver tumor cases consisted of four hemangiomas, a hepatocellular carcinoma, and a hepatoblastoma. All tumors were located in the right lobe, ranging in size from 5 to 12 cm, with a mean of 7. 8 cm. In the hemangiomas, venous collaterals developed by compression of tumor between the right and middle hepatic veins. In the hepatocellular carcinoma, the right hepatic vein was invaded by a tumor thrombus; in the hepatoblastoma, the middle hepatic vein was invaded by a tumor thrombus. Venous collaterals developed around the thrombosed hepatic vein.

The five primary Budd-Chiari syndrome cases consisted of four cases of suprahepatic IVC membranous obstruction and a case of middle hepatic vein obstruction. The intrahepatic vasculature in the cases with suprahepatic IVC obstruction appeared abnormal, with failure to visualize any of the normal right, middle, and left hepatic veins. Venous collaterals were distributed throughout the entire liver (Fig. 1). In the case with middle hepatic vein obstruction, the middle hepatic vein was thrombosed at its junction with the IVC. The right and left hepatic veins communicated with the IVC, but the middle hepatic vein was thrombosed just before the entrance to the IVC. Fine venous collaterals between the right hepatic vein and the middle hepatic vein and between the middle hepatic vein and the left hepatic vein provided drainage by passing the obstruction.

The two adrenal metastasis cases, one from an esophageal carcinoma and one from a lung carcinoma, showed direct invasion into the IVC. Transhepatic venous collaterals developed from the IVC just below the obstruction to the right hepatic vein (Fig. 2).

In the diaphragmatic hernia cases, the right hepatic lobe was markedly elevated, and venous collaterals developed between the right and the middle hepatic veins (Fig. 3). There was no vascular abnormality in the hepatic left lobe.

In the case with Osler-Weber-Rendu disease, venous collaterals occurred between the right and middle hepatic veins.

Doppler waveforms of the collaterals were divided into two flow patterns: flat and multiphasic. The flat flow pattern was seen in patients with veno-occlusive disease, and the multiphasic flow pattern was seen in patients without veno-occlusive disease (Figs. 1, 3).

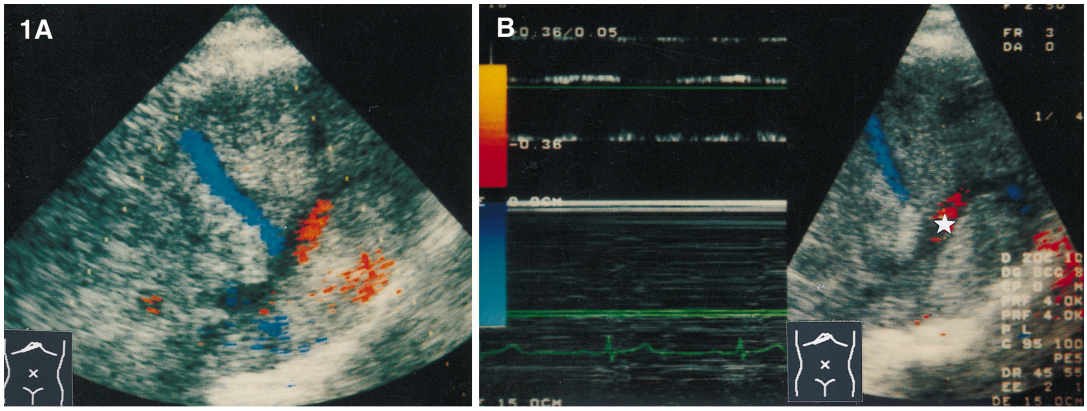

Fig. 1. Primary Budd-Chiari syndrome with venous collaterals. **A** Color Doppler imaging shows tortuous vessels. **B** Flat Doppler waveform obtained from the collateral (*star*, sampling point). **C** Angiogram shows interruption of the IVC (*V*) and development of many fine collaterals.

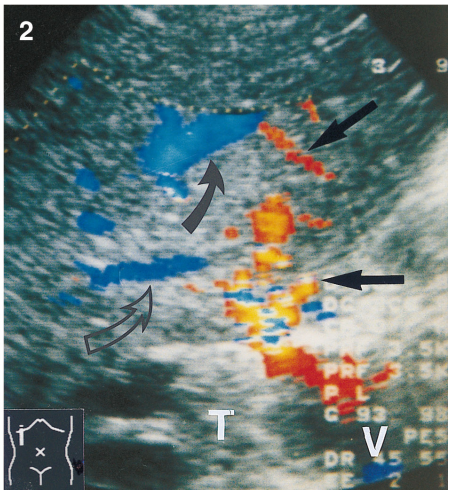

Fig. 2. Adrenal metastasis with transhepatic venous collaterals. Color Doppler sonogram shows many fine collaterals (*arrow*) connecting the IVC and the right hepatic vein (*T*, adrenal metastasis; *V*, IVC; *curved arrow*, right hepatic vein; *curved open arrow*, inferior right hepatic vein).

Other intrahepatic vascular abnormalities included portal thrombus in the case with middle hepatic vein obstruction, portal tumor thrombus in two cases with liver tumors (hepatoblastoma and hepatocellular carcinoma), and arterioportal shunt and portovenous shunt in the patients with Osler-Weber-Rendu disease.

The only extrahepatic vascular anomaly was cavernous transformation of the portal vein in the patient with middle hepatic vein obstruction. No other extrahepatic vascular abnormalities were found by sonography in the other patients.

Clinical manifestations included edema and jaundice in two patients with primary Budd-Chiari syndrome and esophageal varices in all four cases with suprahepatic IVC obstruction. Three tumor (two adrenal metastasis and a hepatocellular carcinoma) patients complained of general fatigue and anorexia. Two of three patients with diaphragmatic hernia developed increasing lassitude and recurrent low-grade fevers due to repetitive respiratory infection. The two cases with Osler-Weber-Rendu disease showed occasional gastrointestinal bleeding from mucosal telangiectasis.

A patient with primary Budd-Chiari syndrome died 1 month after the operation from surgical complications. The two patients with adrenal metastasis died of cachexia within 3 months of diagnosis. The other patients are alive.

Discussion

Intrahepatic venous collaterals were considered to be relatively rare but have been reported in the literature with increasing frequency and are not so rare as once believed [5–7]. They have been reported mainly in patients with hepatic venous or IVC obstruction (veno-occlusive diasease), and their relationship has been em-

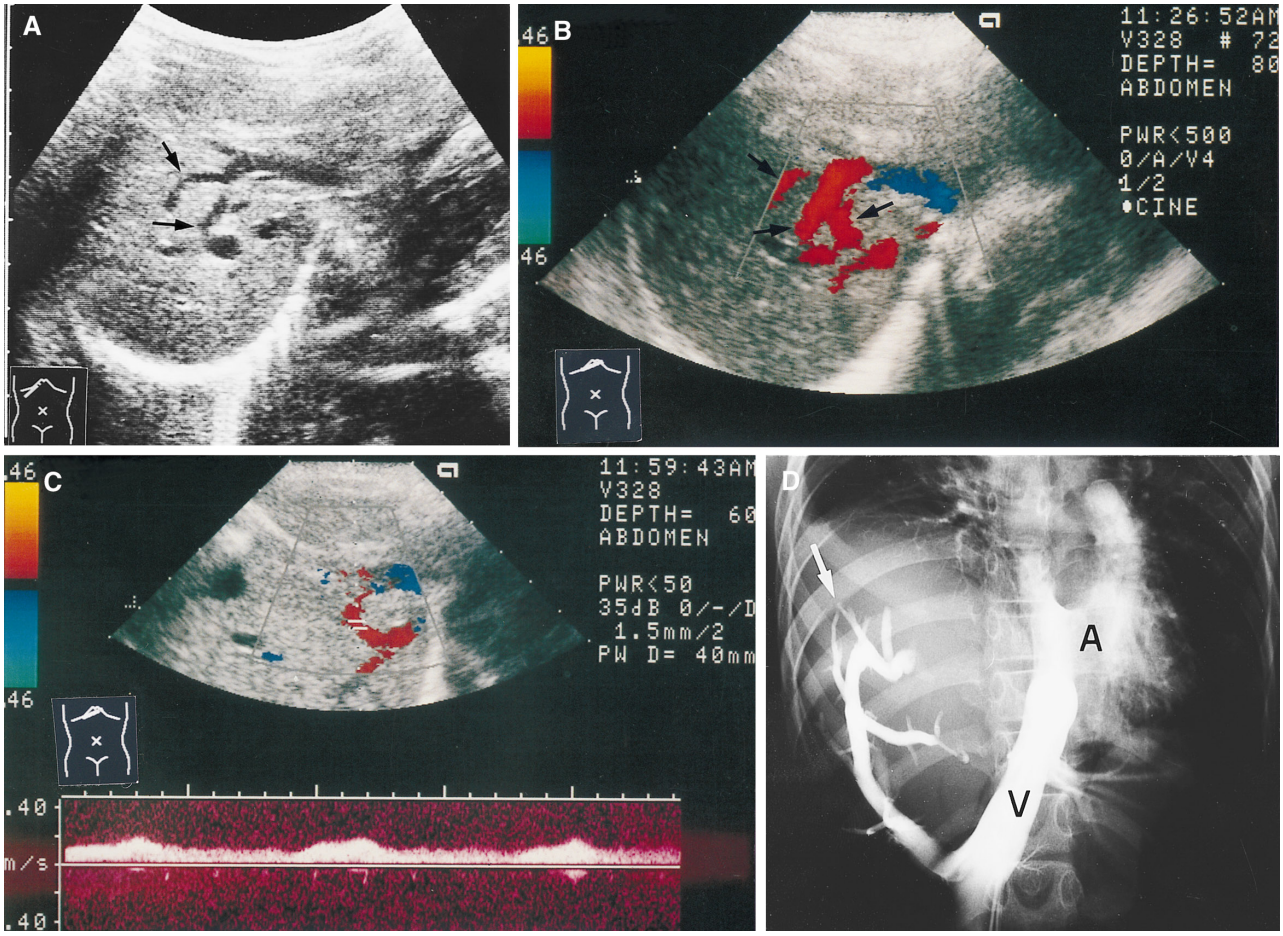

Fig. 3. Diaphragmatic hernia with venous collaterals in the right hepatic lobe. **A** Transverse sonogram of the liver shows tortuous venous collaterals (*arrows*) between the right and middle hepatic veins. **B** Color Doppler imaging confirms the diagnosis (*arrows*, collaterals). **C** Phasic Doppler waveform obtained from collateral. **D** Venogram shows venous collaterals (*arrow*) in the elevated right hepatic lobe (*V*, IVC; *A*, right atrium).

phasized [1, 2, 5–7]. The most common causal disease of intrahepatic venous collaterals reported in the literature is the membranous obstruction of the suprahepatic IVC or that of hepatic veins, the so-called primary Budd-Chiari syndrome [1, 2]. Our series included five such cases, consisting of four cases of membranous obstruction of the IVC and one case of middle hepatic vein obstruction. Of interest is the case of middle hepatic vein obstruction. Unlike the cases with IVC obstruction, the nonthrombosed right and left hepatic veins had their normal course, providing many collaterals as a direct route of drainage from the middle hepatic vein to the right hepatic vein or to the left hepatic vein.

Liver tumors may obstruct or compress major hepatic veins or the IVC directly or in company with a tumor thrombus [8]. In fact, in our series, liver tumors were the most frequent cause, and venous collaterals occurred as a result of tumor compression in four cases and secondary to tumor thrombus in two cases. Of interest is the fact that three of the four cases in the former group were hepatic hemangiomas. It shows that benign tumors can distort the hepatic anatomy to affect changes in hepatic venous flow. It also indicates that the presence of venous collaterals is not a reliable sign for differentiating benign from malignant tumors.

Our series also included two patients with metastatic adrenal tumors invading the IVC with formation of transhepatic fine venous collaterals. Such multiple fine draining vessels originating from the IVC just below the obstruction and providing a relatively direct route of drainage from the IVC to the right atrium via the right hepatic vein have not been reported until now, and this finding is worth noting when one considers the mode of development of draining vessels in patients with IVC obstruction secondary to adrenal tumors.

The precise definition of Budd-Chiari syndrome differs depending on the investigator, but hepatic venous

or IVC obstruction due to liver, renal, or adrenal tumors is usually categorized as secondary Budd-Chiari syndrome [1, 2]. Thus, in the broadest sense of the term, Budd-Chiari syndrome represents 61.9% (13/21 cases) of all the cases in our series. Several studies have documented that the development of intrahepatic venous collaterals follows an obstruction to hepatic venous outflow, regardless of the cause, and it may contribute to the diversion of blood away from obstructed hepatic veins to patent hepatic veins, which helps in draining and decompressing hepatic venous blood [1, 2]. In our series, the four cases with obstruction of the suprahepatic IVC showed venous collaterals occurred throughout the entire liver. Venous collaterals were localized in the other cases. Venous collaterals occurred around the tumor or the tumor thrombus in tumor patients. This observation suggests that the distribution of venous collaterals helps in assuming the obstructive site, and the presence of a tumor should be considered when localized venous collaterals are visible on color Doppler sonography. Although the tumor itself is usually visible on gray-scale sonogram, instances may occur in which an isoechoic tumor is difficult to identify.

More interesting is the fact that our series included eight cases without hepatic venous flow obstruction. If we eliminate two cases of unknown cause, venous collaterals can be encountered as a result of diaphragmatic hernia (three cases) or as an associated finding of Osler-Weber-Rendu disease (two cases) or congestive liver (one case).

Underlying diseases reported in the literature that predispose a patient to intrahepatic venous collaterals without hepatic venous flow obstruction include congestive liver [3] and Osler-Weber-Rendu disease [4], but these are very rare.

The mechanism of formation of intrahepatic venous collaterals without hepatic venous or IVC obstruction is not completely understood, but the angiographic and Doppler findings of the patients with diaphragmatic hernias led us to speculate that a small degree of hepatic outflow disturbance secondary to distortion of the hepatic anatomy, undetectable by angiography, is the likely mechanism of intrahepatic venous collaterals in these patients. It would be of interest to observe the whole liver in patients with diaphragmatic hernias.

The association between Osler-Weber-Rendu disease and intrahepatic venous collaterals is also worth noting. Although the skin and the mucosal membranes are most frequently affected, Osler-Weber-Rendu disease can involve any part of the body [9]. Hepatic involvements include arteriovenous shunts, arterioportal shunts, portovenous shunts, and, less frequently, venovenous shunts (venous collaterals) [4, 10]. In fact, both patients with Osler-Weber-Rendu disease in our series showed not only venous collaterals but also another shunt, an arterioportal shunt in one patient and a portovenous shunt in the other. This finding suggests that, when detecting different kinds of shunts within the liver, Osler-Weber-Rendu diease must be included in the differential diagnosis.

Middleton and Middleton reported a case of congestive heart failure with intrahepatic venous collaterals between the right and middle hepatic veins [3]. They did not comment on the possible mechanism of the collateral formation without hepatic venous flow obstruction. Our series included a similar case having collaterals between the right and middle hepatic veins. Although the mechanism of intrahepatic venous collaterals in our case was not determined, a high pressure in the hepatic venous system secondary to hepatic venous congestion was thought to be the most probable cause.

Although the visualization of collaterals by color Doppler sonography depends on the size of the vessel, recognition of the possibility of the occurrence of venous collaterals is essential when examining patients with Osler-Weber-Rendu disease, diaphragmatic hernia, or congestive liver by color Doppler sonography.

The diagnosis of intrahepatic venous collaterals was made solely by hepatic venography, but color Doppler sonography with high Doppler sensitivity can now be used to confirm the communication between the hepatic veins and collaterals within and around the liver, leading us to establish immediately the final diagnosis of intrahepatic venous collaterals. In our series, we performed hepatic venography successively after color Doppler sonography in the first 14 cases to confirm the diagnosis, but because there were no discrepancies between the hepatic venographic and color Doppler findings, we now believe that the diagnosis by color Doppler sonography alone is sufficient. With the increasing utilization of sonography and color Doppler sonography, the fortuitous detection of intrahepatic venous collaterals may increase.

The Doppler waveforms of the collaterals pose another interesting problem. The waveforms in the hepatic veins are very complex, varying with the cardiac cycle [11]. Normal hepatic venous flow in the liver is phasic, reflecting right atrial and right ventricular diastole, followed by right ventricular systole [11]. In veno-occlusive disease, which prevents pressure changes from being transmitted to the intrahepatic veins, Doppler waveforms can appear flat, with a loss of the normal phasic pattern [11, 12]. Thus, it seems to be very logical that one would observe a phasic flow in the venous collaterals in patients without veno-occlusive disease, instead of a flat flow usually observed in the venous collaterals due to veno-occlu-

sive disease. A lot of care must be taken to observe the entire length of the detected collaterals by color Doppler imaging combined with spectral analysis to assume the cause of collaterals [13].

Clinical manifestations of our cases were thought to differ greatly according to the underlying disease, as were typically seen in patients with Osler-Weber-Rendu disease or those with tumor. The presence of venous collaterals had no prognostic value. According to the literature, the clinical manifestations of primary Budd-Chiari syndrome include abdominal pain, ascites, hepatomegaly, and edema [1, 2]. In our series, of five cases of primary Budd-Chiari syndrome, only two cases showed clinical manifestations: jaundice and edema. The other three cases were almost symptom-free. In these cases, the obstruction of the hepatic veins was presumably gradual, and collaterals arose that were sufficient to prevent the occurrence of clinical symptoms.

In conclusion, we report 21 cases with intrahepatic venous collaterals. The majority of intrahepatic venous collaterals encountered in a clinical setting have been believed to be those associated with Budd-Chiari syndrome, but our observations show that many other diseases, even without hepatic venous or IVC obstruction, predispose patients to intrahepatic venous collaterals, including congestive liver, diaphragmatic hernia, and Osler-Weber-Rendu disease.

Color Doppler sonography clearly visualized the distribution of venous collaterals, which helped in assuming the obstructive site. The addition of analysis of Doppler waveforms has proved very helpful in differentiating venous collaterals with venous obstruction from those without. Thus, color Doppler sonography seems to be a very suitable technique for the diagnosis of intrahepatic venous collaterals.

References

1. Reynolds TR. Budd-Chiari syndrome. In: Schiff L, Schiff ER, eds. *Diseases of the liver*. Philadelphia: JB Lippincott, 1993:1091–1098
2. Sherlock S, Dooley J. The hepatic artery and hepatic veins: the liver in circulatory failure. In: Sherlock S, Dooley J, eds. *Diseases of the liver and biliary system.* London: Blackwell Scientific Publications, 1993:179–198
3. Middleton MA, Middleton WD. Intrahepatic venous collaterals in a patient with congestive heart failure. *J Ultrasound Med* 1994;13:479–481
4. Naganuma H, Ishida H, Niizawa M, et al. Hepatic involvement in Osler-Weber-Rendu disease: findings on pulsed and color Doppler sonography. *AJR* 1995;165:1421–1425
5. Grant E, Parrella R, Tessler FN, et al. Budd-Chiari syndrome: the results of duplex and color Doppler imaging. *AJR* 1989;152:377–381
6. Ralls PW, Johnson MB, Radin DR, et al. Budd-Chiari syndrome: detection with color Doppler sonography. *AJR* 1992;159:113–116
7. Grant EG, Schiller VL, Millener P, et al. Color Doppler imaging of the hepatic vasculature. *AJR* 1992;159:943–950
8. Okuda K, Kojiro M, Okuda H. Neoplasms of the liver. In: Schiff L, Schiff ER, eds. *Diseases of the liver*. Philadelphia: JB Lippincott, 1993:1236–1296
9. Reilly PJ, Nostrant TT. Clinical manifestations of hereditary hemorrhagic telangiectasis. *Am J Gastroenterol* 1984;79:363–367
10. Buscarini E, Buscarini L, Civardi G, et al. Hepatic vascular malformations in hereditary hemorrhagic telagiectasia: imaging findings. *AJR* 1994:163:1105–1110
11. Merritt CRB. Abdomen. In: Merritt CRB, ed. *Doppler color imaging.* New York: Churchill Livingstone, 1992:141–194
12. Hosoki T, Kuroda C, Tokunaga K, et al. Hepatic venous outflow obstruction: evaluation with pulsed duplex sonography. *Radiology* 1989;170:733–737
13. Bolondi L, Gaiani S, Li Bassi G, et al. Diagnosis of Budd-Chiari syndrome by pulsed Doppler ultrasound. *Gastroenterology* 1991;100:1324–1331