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CT angiography and Doppler ultrasound evaluation of congenital portosystemic shunts

Konjenital portosistemik şantların BT anjiyografi ve doppler ultrason ile değerlendirilmesi

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ABSTRACT

Aim: The aim of the study was to describe the Doppler ultrasonography and computed tomography findings that should be considered in the diagnosis and treatment of congenital portosystemic shunts.
Methods: Archive retrospectively scanned. In consideration of shunts: communication type and aneurysm were defined. Additional imaging modalities were utilized.
Results: 11 patients were included in the study. The ages ranged from 0 to 158 months. There were two patients with shunt connecting segment-4 portal vein - middle hepatic vein, two patients with segment-3 portal vein - left hepatic vein, two patients with left portal vein – middle hepatic vein, two patients with portal vein - left renal vein, two patients with portal vein -inferior vena cava, and one patient with portal vein - perirectal venous plexus.
Conclusion: Some classifications used in congenital portosystemic shunts are insufficient in guiding treatment. Elaborate definition of the imaging findings including the involved vessels, type of communication, and presence of aneurysm or dilated vessels is of the prime importance for tailoring clinical management of the patients.

Keywords: Congenital portosystemic shunt; Computed tomography; Doppler ultrasonography,

ÖZ

Amaç: Konjenital portosistemik şantların tanısında ve tedavinin yönlendirilmesinde dikkat edilmesi gereken Doppler ultrasonografi ve Bilgisayarlı tomografi bulgularını tanımlamaktır.
Metod: Arşiv geriye dönük olarak taranmıştır. Şantlar göz önüne alındığında: bağlantı tipi ve anevrizma varlığı tanımlandı. Ek görüntüleme yöntemleri varsa not edildi.
Bulgular: Çalışmaya 11 hasta dahil edildi. Yaşlar 0 ile 158 ay arasında değişiyordu. Şant bağlantısı segment-4 portal ven - orta hepatik ven olan iki hasta, segment-3 portal ven - sol hepatik ven olan iki hasta, sol portal ven - orta hepatik ven olan iki hasta, portal ven - sol renal ven olan iki hasta, portal ven - inferior vena kava olan iki hasta ve portal ven - perirektal venöz pleksuslu bir hasta vardı.
Sonuç: Konjenital portosistemik şantlarda kullanılan bazı sınıflamalar tedavi yönlendirmesinde yetersiz kalmaktadır. İlgili damarlar, iletişim tipi ve anevrizma veya dilate damarların varlığı dahil olmak üzere görüntüleme bulgularının ayrıntılı tanımı, hastaların klinik yönetiminin özelleştirilmesi için birincil öneme sahiptir.

Anahtar Kelimeler: Konjenital Portosistemik Şant; Bilgisayarlı Tomografi; Doppler Ultrasonografi

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Introduction

Congenital portosystemic shunt (CPSS) is a rare disease seen in about one in thirty thousand children. CPSS is an abnormal connection between the portal and venous vessels. The exact etiology is still unknown. Previous studies suggest complex genetic origin, congenital malformative processes developing in response to portal hypertension, congenital liver hemangioma and others [1-4]. As a result of the diversion of portal blood to the systemic circulation, CPSS may result in serious complications, both in intrauterine life and after birth, such as intrauterine growth retardation (IUGR), galactosemia, hepatopulmonary syndrome, hepatic encephalopathy and liver tumors. However, complications such as ascites and portal hypertension are rarely seen as opposed to cirrhosis and portal vein thrombosis [5-7]

The Doppler ultrasound (d-US) is the imaging modality of choice in diagnosis, follow-up and post-treatment monitoring. Antenatal d-US screening may provide early diagnosis. Although the d-US is radiation-free, easy to access and inexpensive, it might not provide elaborate information related to shunt anatomy, intrahepatic portal vasculature and some of the complications [2-4]. With d-US imaging we can analyze vascular flow velocity and flow direction. Computed Tomography Angiography (CTA) is usually performed when the d-US is inadequate in diagnosis, during treatment planning for further analysis of shunt and proper anatomic definition, and probable complication evaluation. Magnetic Resonance Imaging (MRI) is utilized in the suspicion of liver tumor and evaluation of the brain, in the presence of neurological symptoms. Direct Subtraction Angiography (DSA) on the other hand is usually considered for treatment planning or evaluation of CPSS, when the portal system is not visualized via other imaging modalities [8-12].

Due to the aforementioned complications, all of the shunts are required to be closed as early as possible for proper intellectual and physiological growth, with the exceptions of small intrahepatic shunts that might close spontaneously within the first one to two years of life. Invasive treatment might be postponed for the children with galactosemia, which may be controlled with dietary modifications and neonatal cholestasis, and which may resolve spontaneously [1, 3, 5]. The aim of this study was to describe the sonographic and CTA findings of CPSS.

Material and Method

This study was approved by the local Ethics Committee and complies with the Helsinki Declaration (2019-292). We retrospectively reviewed clinical information, imaging findings, disease complications of pediatric age group patients diagnosed with CPSS, who were admitted between February 2016 and June 2020. We included all patients diagnosed with CPSS via CTA and d-US. The exclusion criteria for the study were non-diagnostic imaging due to patient incoordination and patients lost to follow-up. In consideration of shunts: shunt quantity, communication type (side-to-side, end-to-site), and presence or absence of aneurysm were defined. In this study, we sub-grouped CPSS according to flow dynamics. When all the portal blood flows to systemic vessels it is referred to as 'end-to-site shunt'; if only part of the blood is diverted from the portal system to the venous system it is referred to as 'side-to-side shunt. Additional imaging modalities were utilized, such as cranial MRI for evaluation encephalopathy, and thorax CTA for assessment of cardiovascular complications. Patients' age, gender, clinical findings and concurrent pathologies, were recorded.

CTA exams were performed using the Siemens Somatom force (Siemens Healthcare GmbH, Erlangen, Germany). CTA parameters were as follows: 2×192×0.6-mm slice collimation using z-axis flying focal spot technique; 0.25 second gantry rotation time. Automated tube voltage was used according to the patient's size. A dose of 1.5-2.0 mL/kg of iodinated contrast medium (Iohexol, iodine content 350 mg/mL; Omnipaque TM, GE Healthcare) was intravenously administered via the peripheral vein. Images were acquired at arterial, portal, and venous phases.

Statistical analyses were performed via the SPSS (Statistical Package for the Social Sciences) v.22 package program. Mean and standard deviation values were used for descriptive statistics.

Results

Eleven patients diagnosed with CPSS were included in the study. Five patients were female and the remaining six patients were male. Ages ranged from 0 to 158 months. The mean age was 68.7 ±57.78 months. Demographic findings, vascular communication types and additional pathologies are summarized in Table 1.

Table 1: Demographic findings, involved vessels, type of communication and additional pathologies

	Gender	Age (month)	Involved vessels	Type of communication	Additional pathologies and notes
1	M	87	PV-Left renal vein	End-to-side	-
2	F	145	Segment 4 PV-Middle HV	With aneurysm	-
3	M	101	PV-Perirectal venous plexus	End-to-side	Pulmonary hypertension
4	M	78	PV-IVC	Side-to-side	Pulmonary AVF Globus pallidus T1 hyperintensity
5	F	7	Segment 3 PV-Left HV	With aneurysm	
6	F	49	PV-Left renal vein	End-to-side	Pulmonary hypertension
7	M	128	PV-IVC	Side-to-side	Pulmonary hypertension
8	M	158	Segment 4 PV-Middle HV	With aneurysm	-
9	M	0	Segment 3 PV-Left HV	Subcapsular shunt	Shunt disappeared at 3-month-old follow-up
10	F	1	Left PV-Left HV	With aneurysm	-
11	F	2	Left PV-Left HV	Subcapsular shunt	-

(PV: portal vein; HV: hepatic vein; IVC: inferior vena cava; AVF: arteriovenous fistula)

In our study, six patients had intrahepatic and five had extrahepatic CPSS. None of the patients had mixed CPSS. Two patients had CPSS between segment-4 portal vein (PV) branch and middle hepatic vein (HV), two patients between the left PV and middle HV, two patients between segment 3 branches of the PV and left HV, two patients between the main PV and left renal vein, two patients between the PV and inferior vena cava, and one patient had a dilated vascular structure connecting perirectal venous plexus.

One patient had an intrahepatic CPSS. The diagnosis was made during antenatal ultrasound screening and postnatal d-US confirmed diagnosis of CPSS between segment 3 PV and left HV with an aneurysmatic vein, in the anterior subcapsular area. CTA was utilized for evaluation of probable complications and further treatment planning when he was 1-months-old. CTA depicted a decreased size of the CPSS diameter compared to the US. The patient was booked for follow up and d-US was scheduled for 3 months later. During the follow up the patient was asymptomatic and the shunt could not be visualized during d-US, suggestive of spontaneous resolution.

In all intrahepatic CPSS patients, d-US and CTA imaging depicted asymmetric dilatation of the hepatic vein to which portal flow diverted when compared to uninvolved hepatic veins (Figure 1).

Aneurysmatic dilatation of the shunt was observed in four patients (two had shunt between PV segment 4 branch and middle HV, one had between PV segment 3 branch and left HV, and one had between left PV and left HV) (Figure 1).

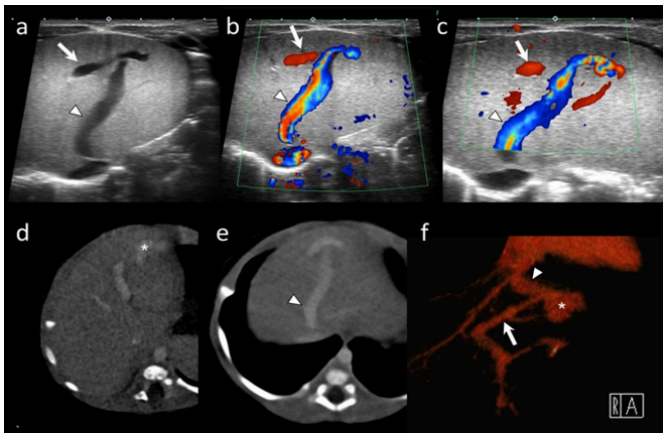

Figure 1. One-month-old female patient. B-mode US (a), Doppler US (b, c), axial CTA (d, e), and volume rendering technique (VRT) image (f) of aneurysmatic (*) CPSS between left portal vein (arrow) and middle hepatic vein (arrowhead). Note that the asymmetric dilatation of the middle hepatic vein (arrowhead) due to diverted blood flow from the portal system

Four patients had pulmonary vascular pathologies. Two patients with pulmonary hypertension had end-to-side type shunts one terminating at the left renal vein (Figure 2) and the other one at the perirectal venous plexus (Figure 3). Intrahepatic portal venous branches of these two patients could not be observed via d-US. The other two patients had side-to-side shunts between the portal vein and inferior vena cava (IVC) (Figure 4). One of the patients with pulmonary hypertension and the other with pulmonary arteriovenous fistula (AVF) diagnosis. These two patients had a hypoplastic intrahepatic portal venous system due to the diversion of portal blood flow to the systemic circulation.

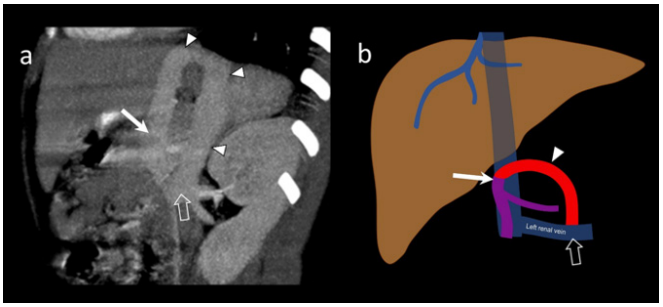

Figure 2. A 7-year-old male patient. Coronal oblique maximum intensity projection (MIP) CT images (a) and schematic drawing (b) of CPSS between the portal vein and left renal vein. CPSS (arrowhead) originating from portal confluence (arrow) ascending till the level of the diaphragm, then makes a 180-degree turn and coursing caudally towards the pelvis and drains into the left renal vein (blank arrow)

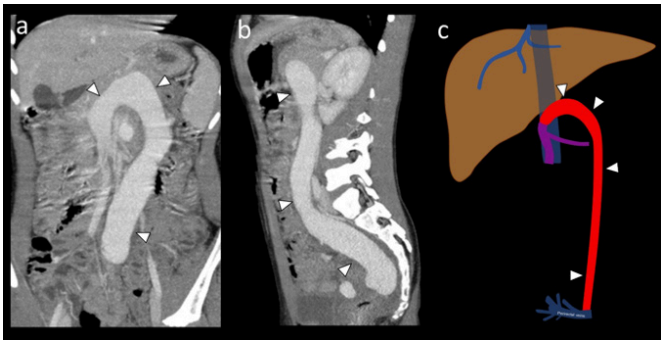

Figure 3: 8 years old male patient with CPSS between the portal vein and perirectal venous plexus. Coronal and sagittal MIP images (a, b) and the schematic drawing (c) of the dilated vessel (arrowhead). Shunt caliber reaches up to 2,5 cm diameter.

Six patients had cranial MRI due to abnormal neurological examination. Two patients had increased T1 intensity in globus pallidus, suggestive of hepatic encephalopathy (Figure 4). One of them had an end-to-sides type shunt between the portal vein and perirectal venous plexus and the other one had side-to-sides type shunt between the PV and IVC.

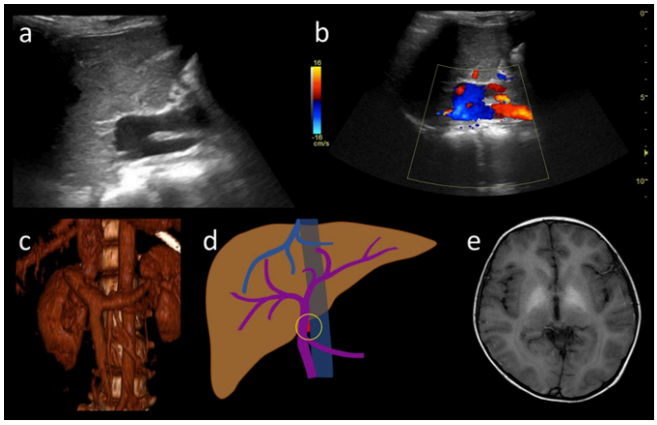

Figure 4: 6 years old male patient. Previously follow up for pulmonary arteriovenous fistula (AVF). Sagittal ultrasound image (a) depicts a side-to-side connection between the portal vein (PV) and inferior vena cava (IVC). Sagittal Doppler US (b) demonstrates the flow portal vein to IVC. Volume rendering technique (VRT) (c) image and schematic drawing (d) of the shunt. Cranial MRI (e) of the same patient, T1 weighted sequence image shows increased intensity in bilateral globus pallidus.

Discussion

In the literature, there are several classification systems for CPSS, which is essentially divided into two groups, namely intrahepatic and extrahepatic. Extrahepatic CPSS are further divided into two subgroups that are type 1 -congenital absence of an intrahepatic portal system, and type 2 -hypoplasia of the main trunk. Kobayashi et al. sub-grouped the extrahepatic shunts according to drainage vein. If the shunt is drained to IVC it is referred to as type A, to the renal vein as type B, and to the iliac vein as type C [13]. For intrahepatic shunts, the 'Park classification' is most commonly used, in which intrahepatic shunts are sub-grouped into 4 types: Type 1: A single vessel communication between the main branch of the portal vein and IVC; Type 2: Peripheral location in one segment; Type 3: Peripheral location in one segment through an aneurysm; Type 4: Multiple small communications distributed diffusely in both lobes [1,4,5,14,15] (Table 2).

Another concern is for the 'Abernethy Classification'. Type 1 CPSS is defined as the absence of an intrahepatic portal system and liver transplantation is considered solely as a treatment modality. However, advances in imaging revealed that the hypoplastic intrahepatic portal system might not be able to be visualized with routine imaging modalities due to small

caliber or diminished blood flow. In that sense, the term 'absence' might not be 'the grim truth'. Kanazawa et al. sub-grouped intrahepatic portal system in a series of eighteen patients using shunt occlusion test based on the severity of hypoplasia (mild, moderate, severe). In that study, liver transplantation was required only for two patients [10]. A balloon occlusion test is utilized during transcatheter angiography and is useful for the patients with severe hypoplastic portal veins in whom closure of the shunt may cause colon necrosis. Treatment modality for these patients is staged interventional or surgical closure. Liver transplantation is taken into consideration as the last option [3-5].

Table 2: Mostly used classifications for intra and extrahepatic CPSS of Kobayashi et al and Park et al (11, 12).

Most Commonly Used Classifications	Types	Shunt from	Drainage vein
Extrahepatic Shunt (Kobayashi et al)	Type A	PV	IVC
	Type B	PV	Renal vein
	Type C	PV	Iliac vein
Intrahepatic Shunts (Park et al)	Type 1	PV	IVC
	Type 2	Peripheral sides of PV	Peripheral sides of HV
	Type 3	Peripheral sides of PV with aneurysm	Peripheral sides of HV
	Type 4	Multiple peripheral sides of PV	Multiple peripheral sides of HV

PV: portal vein; IVC: inferior vena cava; HV: hepatic vein.

Bernard et al. found that previously used CPSS classification systems might not be sufficient for treatment planning. Probability of the presence of both intra and extrahepatic shunts, the presence of ductus venosus, and plasticity of intrahepatic portal system is also important for prognosis and clinical management, but not taken into consideration in any of aforementioned classification systems [1]. In our study, we did not use any of the classification systems in the literature. We believe that an elaborate definition of the shunt is of prime importance for clinical patient management. Detailed definition of the involved vessels, type of communication (side to side -partial diversion of blood flow-, end to side – a total diversion of blood flow), and the presence of aneurysm or dilated vessels, are essential for treatment planning.

Once the diagnosis is made, the patients should also be evaluated for probable concurrent congenital abnormalities. Cardiovascular system abnormalities (atrial septal defect, ventricular septal defect, etc.) are the most common, followed by spleen (polysplenia, asplenia) and other vascular anomalies (splenic artery aneurysm, primitive hypoglossal artery, etc.) among the congenital abnormalities [5]. Hence, vice versa, patients with the aforementioned abnormalities should be investigated for CPSS. In our study, two patients who had pulmonary hypertension and pulmonary AVF were incidentally diagnosed with CPSS.

Clinical management depends on the type of the CPSS and the age of the patient. Follow-ups might be sufficient, in particular for asymptomatic patients with small intrahepatic shunts which may spontaneously disappear within the first two years of life. In one patient the shunt that we detected in the neonatal period disappeared at a 3-month follow-up, suggestive of spontaneous resolution. In symptomatic patients or over two years of age, treatment of choice for the closure of the CPSS is interventional radiological procedures due to the lower invasiveness. Surgery is not taken into consideration unless interventional radiological procedures are failed [3-5,10]. Patients with portosystemic shunts might suffer from hepatic encephalopathy due to hyperammonemia. In our study, six patients had cranial MRI due to abnormal neurological examination and two patients had T1 hyperintensity in globus pallidus bilaterally, suggestive for hepatic encephalopathy [1, 4, 5, 10].

Limitations: The major limitation was the limited number of patients. This is mainly due to the rarity of the disease. Additionally, we excluded some cases lost to follow up, lack of CTA imaging and non-diagnostic CTA examinations.

Conclusion

Abnormalities in liver function tests, galactosemia, IUGR, and some congenital abnormalities might be related to CPSS. Dedicated detailed imaging of the portal system not limited to main branches, portal trunk, superior mesenteric vein, and splenic vein, but also including tiny intraparenchymal end branches, aids in the diagnosis of CPSS.

Radiologists should be aware of clues leading to the diagnosis, such as asymmetric enlargement of the involved hepatic veins which might be related to portosystemic shunt due to asymmetric drainage. Current classification systems of CPSS might not be adequate for clinical management. Detailed definition of involved vessels, type of the shunt and presence of aneurysm/dilatation, should be reported.

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