Original Article

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Congenital Portosystemic Shunt: An Under-Recognized Cause of Pulmonary Arterial Hypertension

Gopinath Periaswamy[1], Pudhiavan A[2], Navya Christopher[3], Kartheek Guthikonda[4], Venkatesh Kasi[5], Mathew Cherian[6]

[1]Resident, Kovai Medical Center and Hospital, Coimbatore, Tamil Nadu, India. Email: gperiaswamy@gmail.com, ORCID ID: 0000-0001-7079-8546

[2]Lead Consultant Cardiothoracic Radiologist, Kovai Medical Center and Hospital, Tamil Nadu, India.

Email: drpudhiavan@gmail.com, ORCID ID: 0000-0003-2324-3489

[3]Resident, Kovai Medical Center and Hospital, Tamil Nadu, India. Email: navyachr7780@gmail.com, ORCID ID: 0000-0003-1616-9110

[4]Resident, Kovai Medical Center and Hospital, Tamil Nadu, India. Email: kartheek.guthikonda@gmail.com, ORCID ID: 0000-0002-5317-3787

[5]Lead Consultant abdominal Radiologist, Kovai Medical Center and Hospital, Tamil Nadu, India.

Email: drkasivenkatesh@yahoo.co.in, ORCID ID: 0000-0003-3894-1722

[6]Head, Kovai Medical Center and Hospital, Tamil Nadu, India. Email: drmathewacademics@gmail.com. ORCID ID: 0000-0003-3993-6562

Abstract

Background: Congenital portosystemic shunt (CPSS) is an uncommon vascular malformation which causes partial or complete shunting of the portal blood into systemic circulation. The purpose of this study is to retrospectively analyze the clinical features, imaging features and therapeutic outcomes of the cases of CPSS diagnosed in our institution. **Subjects and Methods:** A retrospective analysis of the clinical features, imaging features and therapeutic outcomes of all 8 patients of CPSS in our institution between 2017 and 2021 was conducted in this study. Park et al classification was used for intrahepatic portosystemic shunts and Morgan & Superina classification for extrahepatic portosystemic shunts. **Results:** Of the total eight cases of congenital portosystemic shunts, four cases had extrahepatic portosystemic shunts and four cases had intrahepatic portosystemic shunts. Five cases presented with pulmonary arterial hypertension (PAH). Associated features were present in four cases among which two cases had PDA with ASD (Fig 4), one case had only PDA and one case had pulmonary arterio venous fistula, double SVC and focal nodular hyperplasia in the liver. However, no statistical correlation was found between the type of shunt or associated features with portosystemic shunts and development of PAH. **Conclusion:** In our retrospective study we observed that unexplained pulmonary arterial hypertension was present in the majority of cases of CPSS and the development of PAH could not be explained by the presence of associated conditions with it. CPSS could be a more common cause of undiagnosed PAH in children than previously recognised. Hence, the children with unexplained PAH should be screened for CPSS also.

Keywords: Congenital portosystemic shunt, Pulmonary arterial hypertension.

Corresponding Author: Dr. Gopinath Periaswamy, Resident, Kovai Medical Center and Hospital, Coimbatore, Tamil Nadu, India.
Email: gperiaswamy@gmail.com

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Introduction

Congenital portosystemic shunt (CPSS) is an uncommon vascular malformation which causes partial or complete shunting of the portal blood into systemic circulation.[1,2]
With advancement in imaging methods, CPSS has been diagnosed more frequently and is being increasingly detected in children with unexplained pulmonary arterial hypertension (PAH).[3,4] However, the frequency of CPSS and the role of interventional radiology in CPSS-related PAH are still unclear.
A retrospective analysis of the clinical features, imaging features and therapeutic outcomes of all 8 patients of CPSS in our institution between 2017 and 2021 was conducted in this study. According to the study, CPSS appears to be a common cause of unexplained PAH in children. For the majority of CPSS patients, shunt closure or liver transplantation may avoid or even improve PAH progression.

Subjects and Methods

A total of 8 patients of CPSS between 2017 and 2021 were included in this retrospective study. They had been subjected to echocardiography to diagnose PAH and the diagnosis of CPSS was confirmed using CT angiography and MRI. CT was performed using a multidetector third generation dual source system and MRI was performed using a 3T MRI system. The types of congenital porto systemic shunt were classified as per Park et al,[5] classification for intrahepatic portosystemic shunts and

Morgan & Superina classification for extrahepatic portosystemic shunts.[6] This was a retrospective study using previous images and data available so institutional ethical board approval was not required.

Continuous variables are presented as the mean ± standard deviation. Categorical variables are presented as numbers (percentages) and Chi square test as appropriate. All statistical tests were performed using SPSS version 22.0 (SPSS, Chicago, IL, USA).

Results

Among the 8 patients included in this study, 6 were males and 2 were females. The age group varied from seven days of life to seven years of life. The clinical presentations among the study population are given in table 1 among which respiratory distress with pulmonary arterial hypertension was the leading presentation [Table 1].

Table 1: Frequency of the clinical features at first presentation.

Clinical features at first presentation	Number of patients
Respiratory distress	5
Jaundice	1
Seizures	1
Antenatally detected liver cyst	1

Four cases had extrahepatic portosystemic shunts and four cases had intrahepatic portosystemic shunts. Two cases had type 1 [Figure 1] and one each had type 2 and type 3 intrahepatic portosystemic shunts. Two cases had type 1 b [Figure 2] and two cases had type 2 [Figure 3] extrahepatic portosystemic shunts.

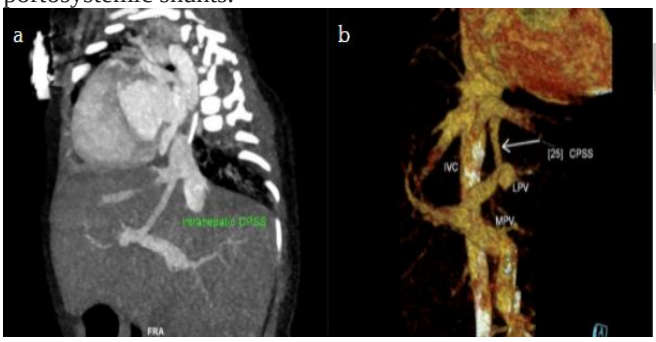

Fig 1: Coronal CT reformat (a) and VRT images (b) shows Intrahepatic shunt between left portal vein and IVC - s/o Type 1 intrahepatic porto- systemic shunt.

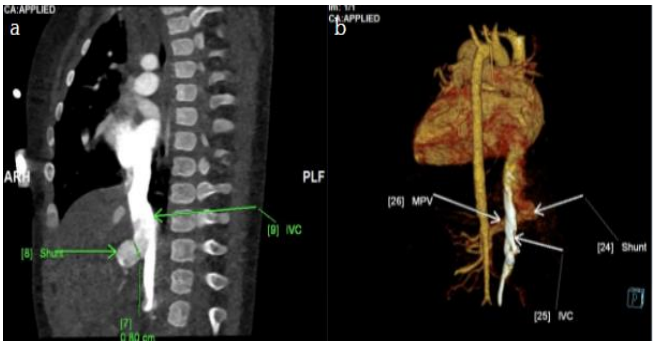

Fig 2: Sagittal CT reformat (a) and VRT images (b) shows large extrahepatic portosystemic shunt between the MPV and IVC - S/o Type Ib Extrahepatic portosystemic shunt.

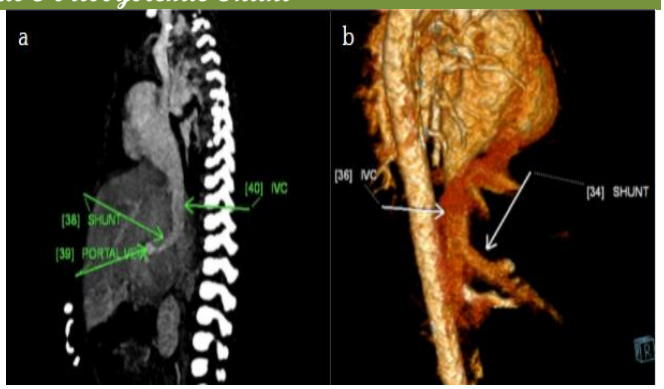

Figure 3: Sagittal CT reformat (a) and VRT images (b) shows side to side shunt between the MPV and IVC - S/o Type II Extrahepatic portosystemic shunt.

Of the total eight cases of congenital portosystemic shunts, five cases presented with PAH and of the five cases, three were extrahepatic portosystemic shunts and two were intrahepatic portosystemic shunts. Statistical analysis of any relevance in development of PAH in EPS or IPS groups [Table 2] shows statistical insignificance (Fisher exact test - 1).

Table 2: Comparison of congenital portosystemic shunt cases with presence of PAH.

	PAH	Non-PAH
CEPS	3	1
CIPS	2	2

Associated features were present in four cases among which, two cases had PDA with ASD [Figure 4], one case had only PDA and one case had pulmonary arterio venous fistula [Figure 5], double SVC and focal nodular hyperplasia in the liver [Figure 5]. However there was no significant association between the presence of associated features and development of PAH [Table 3] in these cases (Fisher exact test - 1).

Table 3: Comparison of presence of associated features with development of PAH.

	PAH present	PAH absent
Associated features present	3	1
Associated features absent	2	2

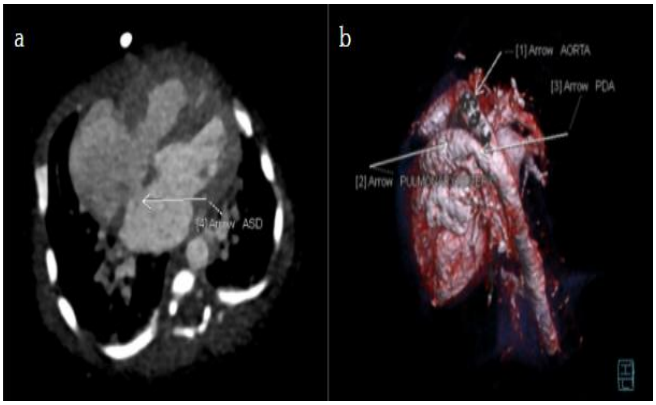

Figure 4: Axial CT image at the heart level (a) and VRT image of the heart (b) shows a PDA and small ASD in Type 2 intrahepatic portosystemic shunt.

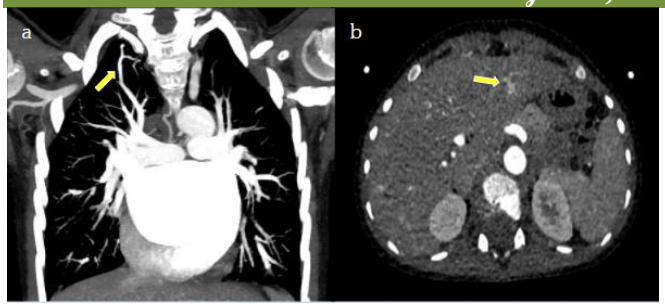

Figure 5: Coronal CT image of thorax (a) and Axial CT image (b) of liver shows right apical pulmonary AV fistula and Focal nodular hyperplasia in left lobe of liver in Type Ib Extrahepatic portosystemic shunt.

Four children were treated with portosystemic shunt embolisation interventionally using vascular plugs [Figure 6]; three children died before any interventional procedure could be performed and one child had spontaneous closure of intrahepatic portosystemic shunt within one year. The treated four children are on regular follow-up and are doing better without any progression of the PAH.

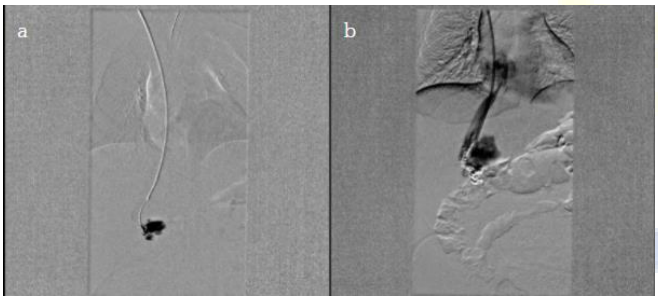

Figure 6: Angiographic images pre (a) and post embolisation (b) shows aneurysmally dilated vascular sac adjacent to GB fossa embolised with vascular plugs.

Discussion

CPSS is a rare portosystemic vascular anomaly with varied clinical presentation, which can range from incidental detection to hepatic encephalopathy or liver failure, with common presentations being hypergalactosemia, hyperammonemia and hyperglycemia.[7]

In our study during the period of 2017 to 2021, we retrospectively analyzed 8 cases of congenital portosystemic shunt diagnosed using imaging in our institution and found that among the 8 patients, 5 patients presented with PAH. Though PAH is not regarded as a major clinical presentation, it turned out to be the most common manifestation in our study. Four cases had extrahepatic shunts and four cases had intrahepatic shunts and there was no association between the type of shunt and development of PAH.

Congenital portosystemic shunts are classified according to the location as intrahepatic or extrahepatic portosystemic shunts. Intrahepatic portosystemic shunts are further classified according to Park et al classification as,[5]

Type 1 - Single connection between right portal vein and IVC.

Type 2 - Multiple connections between portal and hepatic veins within a hepatic segment.

Type 3 - Type 2 with an intervening aneurysm.

Type 4 - Multiple and distributed throughout the liver.

In addition to the above four types, a fifth type has been suggested by Gallego et al.[6]

Type 5 - Persistent ductus venosus.

Extrahepatic portosystemic shunts are further classified according to Morgan et al classification as.[7]

Type 1 - Absent intrahepatic portal branches

1a - Splenic vein and superior mesenteric vein drain separately into a systemic vein

1b - Splenic vein and superior mesenteric vein drain together via a common trunk

Type 2 - Partial shunt with preserved hepatic flow.

CPSS is associated with multiple anomalies which include heterotaxy, congenital heart disease, hepatopulmonary syndrome and liver lesions.[8] Among the congenital heart diseases, ASD was most common and the majority of the cases progressed to heart failure without any treatment.[9]

In our study, associated features were present in four cases, among which two cases had PDA with ASD, one case had only PDA and one had pulmonary arterio venous fistula, double SVC and focal nodular hyperplasia in the liver. However there was no significant association between the presence of associated features and development of PAH in these cases.[10]

The treatment for CPSS includes medical management, interventional radiological management and surgical management. Intrahepatic shunts detected during antenatal or infant period may not always require treatment, since many will close spontaneously by the age of one year with relief of symptoms, in contrast to extrahepatic shunts and patent ductus venosus, which are unlikely to close.[10] Interventional radiological procedures can be used to embolize the portosystemic shunts.[11] Surgical ligation can also be performed in some cases. Extrahepatic or large intrahepatic shunts not responsive to embolization, or with unsuccessful radiological intervention, or with the development of HCC or hepatoblastoma can be treated by liver resection or transplantation.[12]

CPSS is not a widely sought out cause of pulmonary arterial hypertension. The exact mechanism of development of PAH in patients with CPSS is not clear, but various mechanisms have been postulated. A possible hypothesis is that the vasoactive substances present in the gastrointestinal circulation (e.g., serotonin, histamine, estrogen, glucagon) can bypass the hepatic parenchyma without being metabolized, pass through the shunts and reach the pulmonary parenchyma, causing pulmonary vasoconstriction and increased pulmonary vascular resistance.[13] Another possible explanation is that the early phase of PAH may be due to microembolic arteriopathy, and advanced PAH may be due to plexogenic arteriopathy.[14]

In our retrospective study we observed that unexplained pulmonary arterial hypertension was present in the majority of cases of CPSS and the development of PAH could not be explained by the presence of associated conditions with it. Hence CPSS should be regarded as a cause of unexplained

PAH in pediatric cases.

Limitations:

It is a retrospective study and the overall cohort sizes were small. PAH was not confirmed by cardiac catheterization and PAH cases with other causes were not compared and the incidence of CPSS in all PAH cases was not calculated.

Conclusion

CPSS could be a more common cause of undiagnosed PAH in children than previously recognized. These data suggest that children with unexplained PAH should be screened for CPSS. In the majority of CPSS patients, shunt closure or liver transplantation can prevent PAH progression.

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