Case Report

# Hepatic Lesions in Children Related to Congenital Intrahepatic Portal Venous Shunts

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## Abstract

We present two patients with hepatic lesions associated with congenital portosystemic shunts (CPSS), a rare vascular malformation, in order to emphasize the variable clinical presentation of this condition and different management strategies in each case. CPSS can give rise to several complications such as hepatic encephalopathy, portopulmonary hypertension and hepatic tumors. These hepatic lesions though most commonly benign in nature have an increased risk of malignant transformation. Therefore, we underline the necessity of considering CPSS in the differential diagnoses for all cases of space-occupying hepatic lesions in pediatric patients.

**Keywords:** Congenital portosystemic shunts, hepatic lesions

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**Abbreviations:**

CPSS: Congenital portosystemic shunts

IVC: Inferior vena cava

FNH: Focal nodular hyperplasia

## Introduction

Congenital portosystemic shunts (CPSS) are rare vascular malformations creating abnormal connections between the portal and the systemic venous systems, diverting the blood flow away from the liver and into the systemic circulation. This is probably due to incomplete or lacking involution of the primordial abdominal embryonic veins (Franchi-Abella et al.,2018). CPSS are often associated with other malformations; involving the cardiovascular system most commonly and genetic disorders such as Down and Turner syndromes. The most common members of the systemic venous circulation involved are the inferior vena cava (IVC) and the right atrium. CPSSs can be single or multiple, intrahepatic or extrahepatic and can vary in size from case to case. Extrahepatic CPSS are also known as Abernethy malformations, named after Dr. J. Abernethy who first described a malformation of this kind. CPSS may also be due to a persistent or patent ductus venosus which many authors categorize on its own, as it is due to the persistence of the fetal circulation pattern (Bernard et al.,2012). Imaging modalities remain the mainstay tools for diagnosis of CPSS, with ultrasound Doppler imaging being the gold standard for both diagnosis, as well as follow up before, during and after treatment (Timpanaro et al.,2015).

CPSS can lead to a wide variety of complications and has an equally large range of clinical presentations making its diagnosis particularly difficult. The main and most clinically important complications include: hepatic encephalopathy with hyperammonemia and delayed mental development or learning difficulties, liver tumors, portopulmonary hypertension and its consequences and pulmonary arteriovenous shunts formation. Some CPSS may remain asymptomatic for decades but carry the risk of hepatic lesions development, as seen in the following two cases (Bernard et al.,2012).

### Case 1:

A six year old male patient (patient A) presented at our clinic with intermittent abdominal pain for more than six months, located in the periumbilical region and no other gastrointestinal symptoms. The parents reported a history of periodic atypical abdominal pain episodes for the past two years. The pain had no association with the timing of food intake or specific foods. During the clinical and laboratory investigation, elevated liver transaminases were noted and ultrasound and MRI imaging revealed three hepatic lesions rapidly increasing in size, as well as a wide shunt between the right portal vein branch and the inferior vena cava (Figures 1 and 2). An open biopsy of the lesions was performed, a tissue sample sized 1.5cm was taken and sent for histopathologic examination, which revealed an atypical hyperplastic nodular lesion; consistent with focal nodular hyperplasia (FNH) of the liver. The postoperative period was uneventful and the patient was discharged three days postoperatively. Since no major complications were present it was decided that the best option at the time was to continue conservatively. We are monitoring the patient closely and we are in the process of designing an appropriate plan for closure of the shunt to avoid portal hypertension and possible

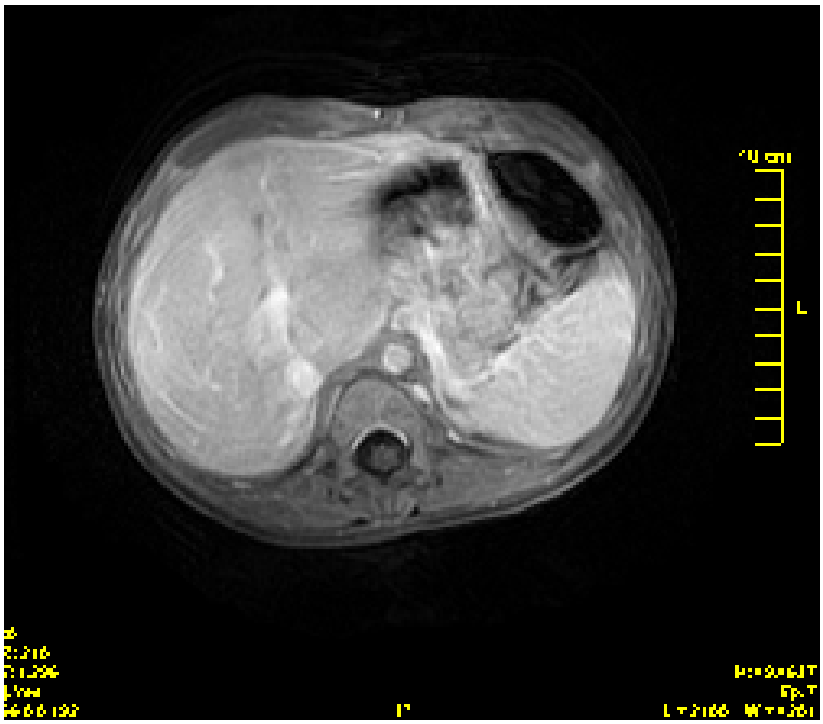

**Figure 1:** Patient A. MRI visualizing communication between the right portal brunch and the inferior vena cava (IVC).

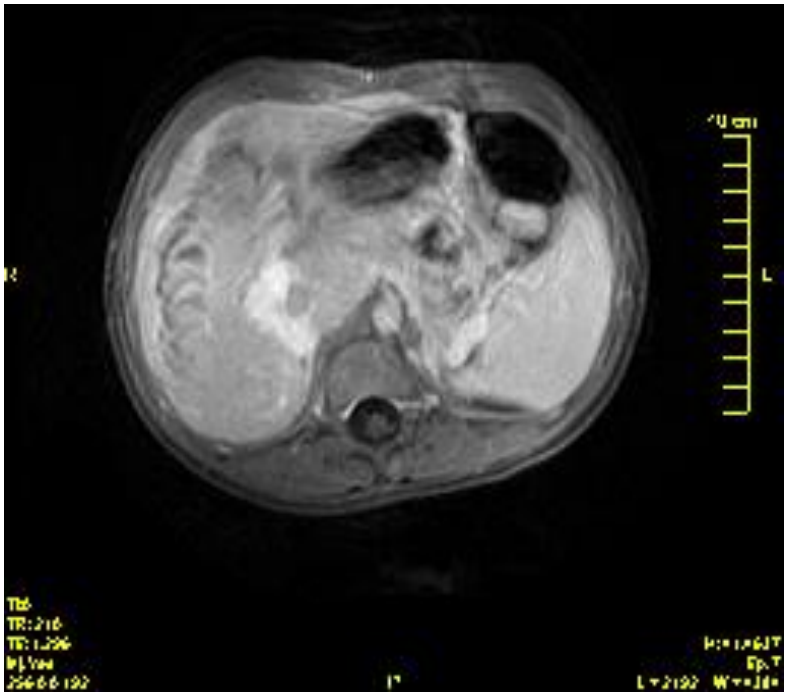

**Figure 2:** Patient A. MRI presenting a wide shunt between the right branch of the portal vein and the IVC.

hepatic failure. Until now, the patient's status remains unchanged and no further actions have been taken.

**Case 2:**

A seven year old male (patient B) presented at the pediatric clinic of our hospital with gastrointestinal disturbances. Gastroscopy was performed which revealed active gastritis. In the patient's medical history galactosemia (not clinically proven) was reported, as well as, hyperbetalipoproteinemia, behavioral changes and hyperactivity. Blood testing revealed elevated liver enzymes and coagulation disturbances. The patient was put on medical therapy with vitamins K, H and A, as well as lactulose. Ultrasound imaging showed an intrahepatic shunt between the IVC and the right branch of the portal vein and 3-4 hyperechoic lesions in the right hepatic lobe (Figures 3-5). MRI investigation revealed multiple hepatic lesions with increased MR sign in the T1 sequences; (Differential diagnosis: regenerative lesions, multifocal nodular hyperplasia) and shunting between the right portal vein branch and the right hepatic vein. Biopsy of the lesions showed an atypical form of focal nodular hyperplasia with no signs of associated malignancy. The patient was discharged on day 6 postoperatively and during follow up his condition remained unchanged 1 month later. Due to the altered hepatic function and coexisting coagulation disturbances seen on presentation, the patient was considered a good candidate for liver transplantation and was put on the national transplantation register.

**Discussion**

The incidence of 1 in 30 000 births reported in neonates becomes 1 in 50 000 in patients over one year old, due to a great percentage of spontaneous closure of the shunt seen within the first year of life. In children not diagnosed during prenatal ultrasound examinations, a CPSS may be diagnosed as an incidental finding during ultrasound or other imaging examination for an unrelated condition or due to complications.

The increasing diagnostic sensitivity of ultrasound imaging has led to earlier diagnosis of CPSS and better planning of subsequent treatment. Once a CPSS has been confirmed additional information about its location and structures involved, as well as investigation for the existence of any hepatic lesions, can be given by a CT and/or MRI scan (Franchi-Abella et al.,2018). Angiography can be used to perform an occlusion test to reveal possible development of portal hypertension after closure of the shunt or for permanent occlusion of the shunt if deemed appropriate (Timpanaro et al.,2015).

These lesions can be single or multiple and are seen with all anatomic types of CPSS. Histologically these lesions include benign tumors such as, adenomas, focal nodular dyplasias and nodular regeneration hyperplasias, as well as malignancies like hepatocellular carcinomas, hepatoblastomas and sarcomas. It has been reported that even benign lesions found in the liver of patients with CPSS carry a risk of malignant transformation which leads to an increasing need for accurate diagnosis and appropriate management of these

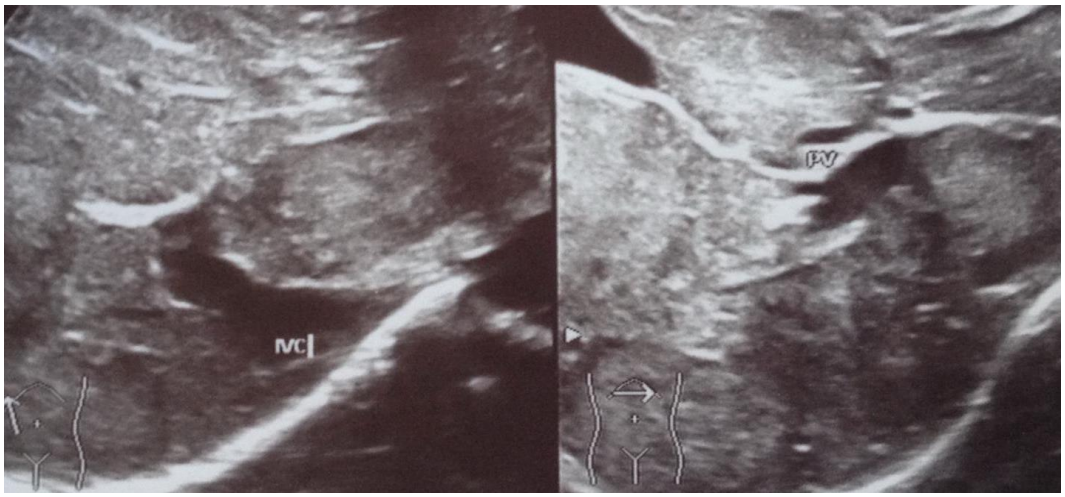


**Figure 3:** Patient B. U/S showing enlargement of the portal vein (PV) due to shunt with the IVC.

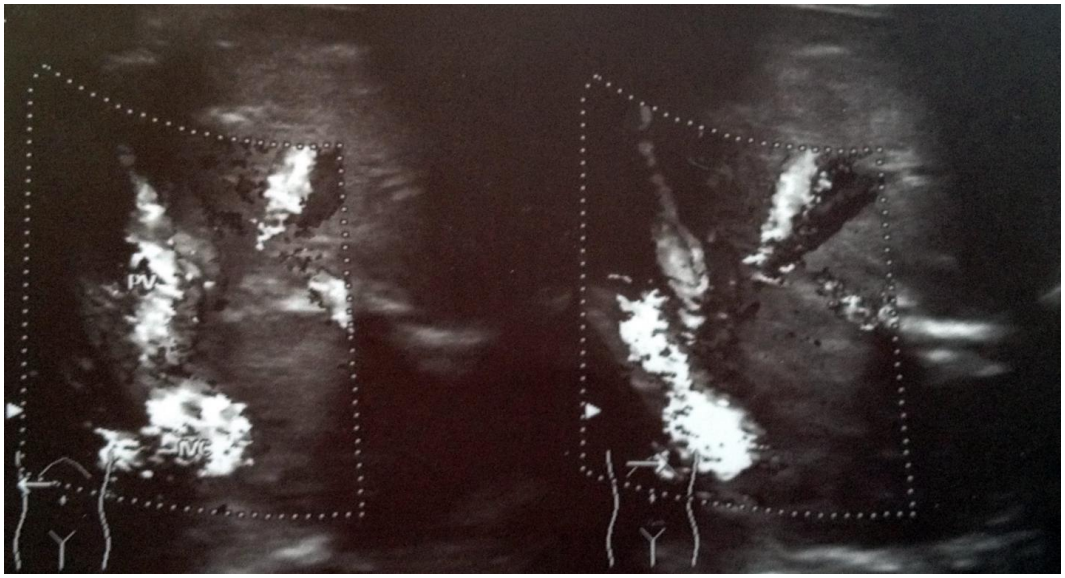


**Figure 4:** Patient B. U/S with Doppler imaging showing portosystemic shunt.

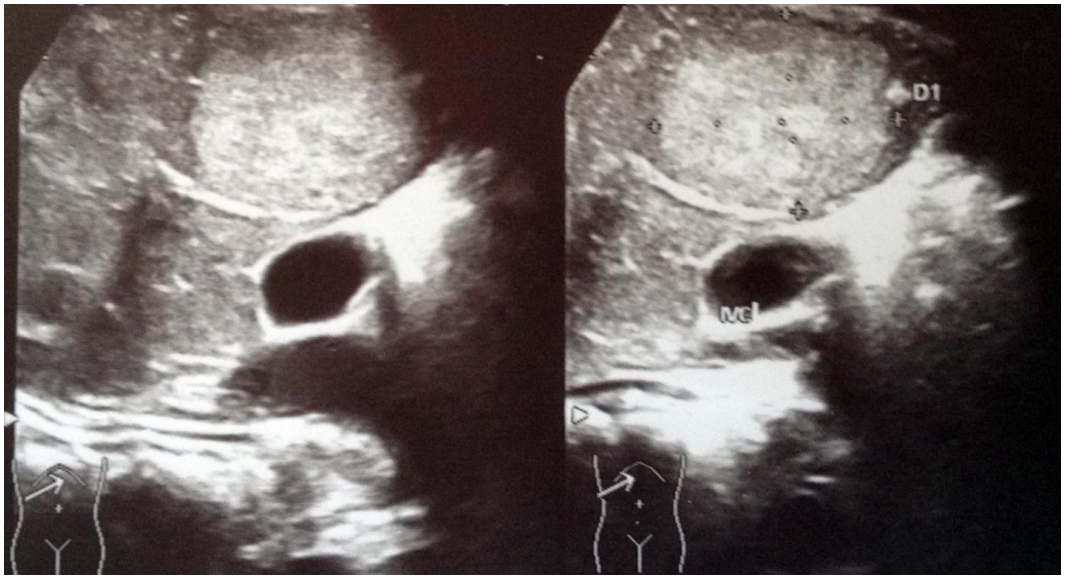


**Figure 5:** Patient B. U/S presenting a mass in proximity with the IVC, histologically found to be focal nodular hyperplasia.

patients (Franchi-Abella S. et al.,2018).

It has been proposed that CPSS leads to poor portal flow, the reduced mesenteric blood return to the liver reduces the levels of insulin and glucagon passing through the liver parenchyma and the regenerative abilities of the liver. This leaves the liver vulnerable to both benign and malignant tumor formation (Lautz T.B. et al., 2016). All of the above are supported by the findings of benign nodules regression, even disappearance, after successful closure of the portosystemic shunt and establishment of the physiologic portal blood flow (Bernard et al. 2012).

Bernard et.al (2012) in their review of 265 children diagnosed with CPSS have recorded 64 patients with associated hepatic tumors of which 26 were single and 38 multiple, histologic examinations showed mainly benign lesions such as FNH and nodular regenerative hyperplasia, while 7 cases were found to have malignant tumors. Two of these were found during follow up of previously known adenomas or nodular regenerative hyperplastic lesions and all malignancies were associated with extrahepatic CPSS. In 21 of these cases were the shunt was closed without tumor resection, the masses either significantly regressed or disappeared after closure of the CPSS.

Lautz et al. (2016) reported two cases with concurrent CPSS and hepatoblastomas, the patients presented with normal liver enzymes, rapidly enlarging hepatic mases and increased levels of αFP. Similar cases were also reported by Marois et al (1979), Barton and Keller (1989), Kawano et al. (2007) and Loomba et al. (2012) which further emphasizes the possibility of malignant tumor development in patients with CPSS, especially extrahepatic CPSS. Small liver masses with normal αFP levels can be safely monitored but enlarging masses must be biopsied (Lautz et al.,2016).

The management of patients diagnosed with CPSS, varies according to their age at presentation and the presence of complications as well as their severity. For asymptomatic infants under the age of one, a conservative approach of serial follow ups is proposed as there is an increased chance of spontaneous regression within the first year of life (Chocarro et al., 2016). Symptomatic patients especially those with hyperammonemia and pulmonary hypertension closure of the shunt either surgical or percutaneous in a one- or two-stage procedure is recommended as soon as possible to ensure minimal progression of their condition (Franchi-Abella et al.,2010). In the case of patients who present with hepatic lesions, benign lesions have been shown to regress considerably or even disappear after closure of the CPSS (Blanc et al.,2014). Hepatic malignancies associated with CPSS, must be resected when possible followed by adjuvant chemotherapy or if deemed unresectable, neoadjuvant treatment can be given followed by tumor resection if successful. Liver transplantation may also be a viable choice for patients presenting with malignant hepatic lesions and CPSS as it resolves at once both the malignancy and the CPSS issue, however lifelong immunosuppression is an issue in this case and it must be taken into consideration (Lautz et al.,2016).

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