Original research article

Pre and postnatal diagnosis of congenital portosystemic shunt: Impact of interventional therapy

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ABSTRACT

Introduction: Congenital portosystemic shunts (CPSS) are rare vascular malformations that can lead to severe complications. With advanced imaging techniques, diagnosis is becoming more feasible occurring in fetal life. Different approaches have been adopted to manage these cases, with an increased utilization of interventional therapy recently. This cohort aims to describe the course of children diagnosed with CPSS and the impact of interventional therapy on the outcome.
Methods: Retrospective chart review was done for all patients who were diagnosed with CPSS in our institution between January 2006 and December 2015.
Results: Six patients were diagnosed with CPSS. During this period, 8,680 mothers carrying 9548 fetuses underwent fetal ultrasound examinations. Three patients were diagnosed antenatally at a median [IQ] gestational age of 33 [26–33] weeks, and three patients were diagnosed postnatally at 0, 2, and 43 months, respectively. At a median follow-up of 87 [74–110] months, 5 patients are alive; 4 of whom had received transcatheter closure for different indications, and one who had spontaneous resolution of her CPSS. One infant died at the age of 6 weeks secondary to sepsis.
Conclusion: CPSS can result in significant complications in children. Interventional therapy is feasible at any age group, but long-term follow-up is warranted.

1. Introduction

Congenital portosystemic shunts (CPSS) are rare vascular malformations that shunt blood between the portal system and any vein of the inferior vena cava system [1–3]. They were originally reported by Abernethy in 1793, who described a postmortem examination of a 10-month-old girl that revealed termination of the portal vein (PV) in the inferior vena cava (IVC) at the level of the renal veins [4]. These rare fistulae are best classified into intrahepatic and extrahepatic shunts [1–3]. In the former, one or more abnormal intrahepatic connections form between branches of the PV and the hepatic veins or IVC. While in extrahepatic shunts, the portomesenteric blood drains into a systemic vein, bypassing the liver through a complete or partial shunt in the absence or presence of the portal vein respectively [1–3]. Recently, Blanc et al. proposed a new classification correlated with surgical strategy rather than the portal origin of the shunt, which may help in planning conservative surgery in selected cases [5].

In this cohort study we describe the clinical course of six patients who were diagnosed with CPSS, and we attempt to evaluate the role of interventional therapy using AMPLATZER® Vascular Plugs, in managing this condition and its impact on the outcome.

2. Patients and methods

Between January 2006 and December 2015, six patients were diagnosed with CPSS at our institution; three of them were diagnosed antenatally. During the same period, 8680 mothers carrying 9548 fetuses underwent fetal ultrasound (US) examinations.

Proof of the shunt was provided by fetal US, abdomen US, or combination of both. Four patients were managed by transcatheter closure of the shunt using AMPLATZER® Vascular Plugs. Follow up for all alive patients consisted of echocardiography, ultrasonographic monitoring of the shunt with clinical and biochemical assessment on

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Abbreviations

ADV	Absent Ductus Venosus
AVM	Arteriovenous Malformation
CHF	Congestive heart failure
CPSS	Congenital Portosystemic Shunt
CT	Computed Tomography
GA	Gestational Age
IQ	Interquartile range
IUGR	Intrauterine Growth Restriction
IVC	Inferior vena cava
MR	Magnetic Resonance
PSS	Portosystemic Shunt
PV	Portal vein
US	Ultrasound
VSD	Ventricular septal defect
3-D	3-Dimensional
4-D	4-Dimensional

indicated in 2 patients: in Case 1, a successful closure of extrahepatic PSS was done on the second day of life (Fig. 1), leading to complete resolution of congestive heart failure (CHF) symptoms. The patient continued to grow well, and her cutaneous hemangiomas completely regressed.

In Case 2, a successful transcatheter closure of extrahepatic PSS was done at one week of age. The course was complicated by plug dislocation and thrombosis of the IVC and infra-renal veins. The plug was extracted later, and the patient continued to grow well with minimal residual shunt presents.

In Case 3, the patient developed CHF on 3rd day of life due to large ventricular septal defect (VSD). Post cardiac surgery she developed pericardial tamponade and necrotizing enterocolitis and died at the age of 6 weeks due to overwhelming sepsis.
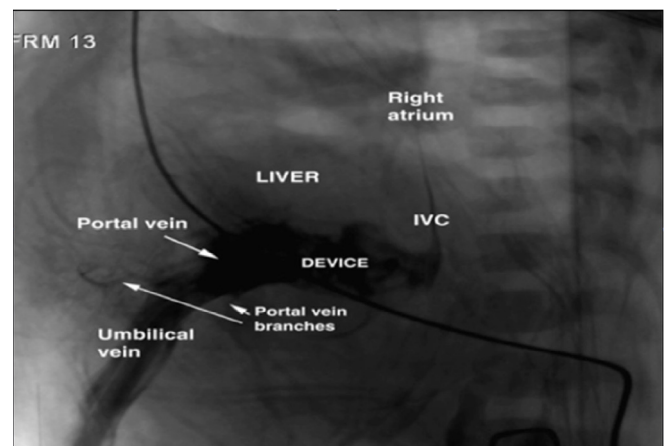
regular basis at 1, 6 and 12 months post shunt closure, and annually after. The median [IQ] follow-up duration for our patients is 87 [74–110] months.

3. Results

Table 1 summarizes the six patients, their findings, management and follow up status. Three patients were diagnosed with intrahepatic PSS and three with extrahepatic PSS.

3.1. Patients diagnosed antenatally

All 3 patients were born prematurely at 35–36 week gestational age (GA). Fetal US findings are summarized in Table 1.

Transcatheter closure using AMPLATZER® Vascular Plugs was

Fig. 1. Closure of extrahepatic shunt using vascular plug in case (1). (Abbreviation: IVC: Inferior vena cava)

Table 1
Summary of all patients with congenital portosystemic shunts.

Case	Dx Method	Age at Dx	Type of shunt	Fetal US findings	Biometry/ hemodynamics	Clinical presentation	Biochemical findings at Dx	Associated anomalies/findings	Mngx	FU in mo	Outcome
1	AN	33 w (GA)	EPSS	ADV UV-IVC Shunt	LGA Polyhydramnios	CHF at age 2 days	Hypoglycemia High GGT Indirect-Hyperbilirubinemia	Cutaneous Hemangioma	Transcatheter closure	115	Alive & well
2	AN	26 w (GA)	EPSS	ADV Right portal vein-IVC Shunt	IUGR Oligohydraminos	Asymptomatic	High LFT's	Static Communicating Hydrocephalus	Transcatheter closure (removed later)	92	Alive & well. Minimal residual shunt
3	AN	33 w (GA)	IPSS	Large DV with unusal course Single UA	Normal	CHF, melena & low platelets at age 3 days	High LFTs Indirect-Hyperbilirubinemia High ammonia	Dandy-Walker malformation, VSD, ASD	No treatment	1.5	Died due to sepsis
4	PN	4 d	IPSS	NA	Normal	None (incidental)	Normal	None	Self - resolved	82	Alive & well
5	PN	7 w	IPSS	NA	IUGR	Jaundice, FTT, & galactosuria at age 4 wks	Hypoglycemia, High LFT's Direct-Hyperbilirubinemia High ammonia	Hepatic Hemangioma	Transcatheter closure	110	Alive & well
6	PN	3.5 y	EPSS	NA	Normal	Cyanosis, clubbing & FTT at age 2.5 y	Positive reducing substances in urine	Pulmonary AVM	Transcatheter closure	74	Alive & well

Abbreviations:AN: Antenatal, ASD: Atrial septal defect, AVM: Arteriovenous malformation, CHF: Congestive heart failure, D: day, DV: Ductus venosus, Dx: Diagnosis, EPSS:Extrahepatic Portosystemic shunt, FTT: Failure to thrive, FU: Follow up, GGT: Gamma-glutamyl transpeptidase, IPSS: Intrahepatic Portosystemic shunt, IUGR: Intrauterine growth restriction, IVC: Inferior vena cava, LGA: Large for gestational age, LFT: Liver function test, MO: Month, NA: Not applicable, PN: Postnatal, UA: Umbilical artery, US: Ultrasound, UV: Umbilical vein, VSD: Ventricular septal defect, W: week, Y: year.

3.2. Patients diagnosed postnatally

All 3 patients were born at term (>37 week GA).

In Case 4, intrahepatic PSS was diagnosed incidentally at the age of 4 days of life. The patient was managed conservatively with spontaneous resolution of the shunt at 6 months of follow up.

In Case 5, liver US revealed large intrahepatic PSS with closed ductus venosus and a small hemangioma in the right hepatic lobe. Urine chromatography was positive for galactose, but the activity assay for serum galactose-1-phosphate uridyl transferase enzyme was normal. The patient underwent successful transcatheter closure of the shunt; intracatheter findings are illustrated in (Figs. 2 and 3). Follow-up liver US one month after the procedure showed complete resolution of the PSS, and partial regression of the hepatic hemanigoma with normalization of the metabolic abnormalities. The hepatic hemangioma resolved completely at 6 months follow up.

The last patient in our cohort (Case 6) presented with a history of long standing hypoxemia secondary to significant intrapulmonary shunting that was detected across an arteriovenous malformation (AVM) within the pulmonary bed, with no significant intracardiac shunt or pulmonary hypertention. Intracatheter findings showed hypoplasia of the right portal vein and a venous connection between the IVC and PV (Figs. 4 and 5). Transcatheter closure resulted in complete resolution of the shunt, improvement of saturation to 100% on room air, and growth of hypolastic portal branches.

All three patients are growing and developing normally.

4. Discussion

CPSS is a rare disorder occurring 1 in 30,000 births [6]. Nonetheless, more cases in fetuses and children are being diagnosed in the last 3 decades as result of advances in imaging techniques [6—9].

Prenatal detection of these shunts has become more feasible since fetal US was first utilized in the early 1980's [10]. With further development of high resolution sonography combined with 3-dimensional (3-D) and (4-D) applications, a more detailed analysis of this abnormality and its variants became available, even as early as 14 weeks GA [7,11].

Several fetal US findings can raise the suspicion of CPSS [12—14]. Nevertheless, agenesis of ductus venosus (ADV) [7,9,11,13] and intrauterine growth restriction (IUGR) [6,8,9,14,15] are among the most frequently reported findings with CPSS. Two of our patients who were diagnosed prenatally had ADV on their fetal US (Case 1 & 2). A further 2 patients had IUGR (Case 2 & 5). None of those patients were dysmorphic, syndromic or had any significant associated malformations.
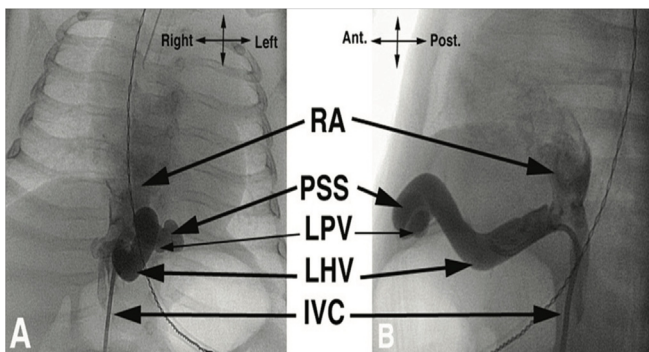

Fig. 3. Postero-anterior (A), and lateral (B) projections of left portal venogram using the transhepatic sheath after deployment of the two vascular plugs (white arrow) in Case (5). There is faint visualization of the LHV indicating a small residual shunt. The LPV is better visualized than in Fig. 2. (Abbreviations: Ant.: Anterior, LHV: Left hepatic vein, LPV: Left portal vein, Post.: Posterior).

The development of IUGR and its correlation with ductus venosus shunting was described in details in a study by Kiserud et. Al [16]. Umbilical circulation compromise was found to be a major determinant of the shunt flow especially in the presence of large ductus venosus diameter. The high shunt flow implies less perfusion
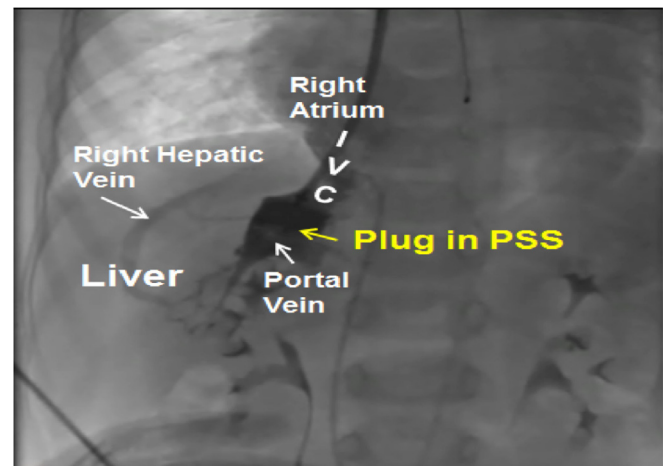
Fig. 4. Injection into the portosystemic shunt just before release of the closure device plug in Case (6). (Abbrevation: PSS: Portosystemic Shunt)

Fig. 2. Postero-anterior (A), and lateral (B) projections of left hepatic venogram in Case (5). It shows an intrahepatic portosystemic shunt, between the left hepatic and left portal veins. (Abbreviations: Ant.: Anterior, IVC: Inferior vena cava, LHV: Left hepatic vein, LPV: Left portal vein, PSS: Portosystemic shunt, Post.: Posterior, RA: Right atrium).
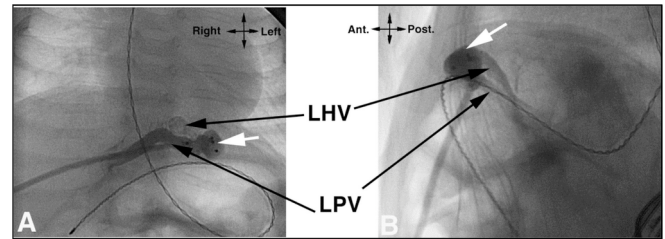

Fig. 5. Pulmonary artery angiogram. Extensive pulmonary arteriovenous malformation in Case (6).

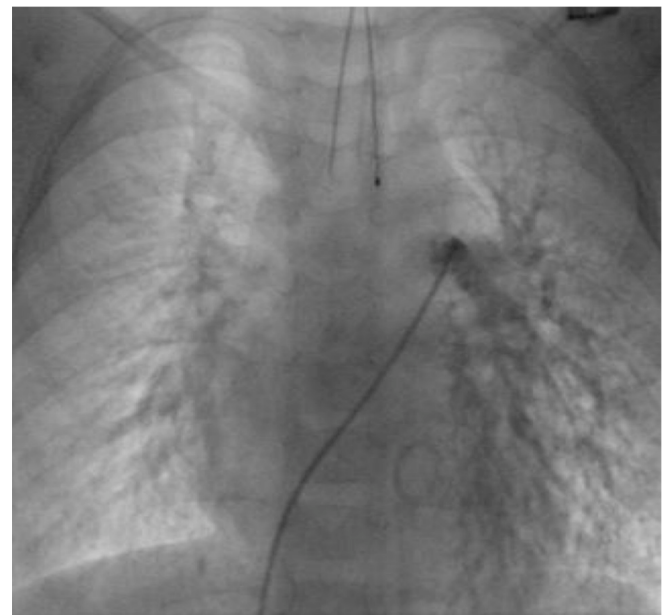to the liver resulting in IUGR even without the presence of hypoxia. This is supported by the fact that the liver plays a major role in fetal growth via the production of the necessary growth factors.

Results of the study by Delle Chiaie et al. were consistent with the previous findings. They also proposed that an isolated ADV or a finding of IUGR without an evident cause should raise suspicion for CPSS [15]. In addition, they emphasized that when ADV is detected, close follow up of fetus' hemodynamics is critical to identify CHF which may develop shortly after delivery [9,11,15]. A complication that was encountered in Case (1) in our cohort. In practice, one should recognize these vascular anomalies before hemodynamic distress or in case of unexplained hemodynamic changes [9].

Diagnosing CPSS postnatally can be more difficult as the presentation is extremely variable. It may range from being asymptomatic (Case 4), to delayed presentation for weeks after birth or even several years as in (Cases 5 & 6) respectively [1,2,6,17]. It is also noteworthy to mention that this rare vascular malformation mostly presents with manifestations of more common diseases, thus making its diagnosis challenging to most physicians. Examples of such manifestations are metabolic abnormalities, liver disease, and neurological disorders including learning disabilities, attention-deficit-hyperactivity disorder, and developmental delay [6].

Among the commonly encountered metabolic abnormalities with CPSS is neonatal galactosemia in the presence of normal enzyme assay for galactose metabolism. This is quite characteristic for CPSS and may be found incidentally during newborn screening [18—20]. Transient neonatal cholestasis may also complicate both intra- & extrahepatic CPSS [6,8,14,17,21], and may co-exist with other more common causes of neonatal cholestasis such as biliary atresia [6,22]. Interestingly, both neonatal galactosemia and cholestasis were found in our patient (Case 5). Other metabolic abnormalities that were detected in our patients are hyperammonemia, elevated liver enzymes, coagulopathy, hyperglycemia, hypoglycemia, and liver and skin hemangiomas; all of which were previously reported with CPSS [6,17,23,24].

CPSS may also be associated with serious complications such as hepatic failure, hepatopulmonary syndrome, portosystemic encephalopathy, and liver tumors [2,6,17,20,25—27]. These can be the presenting problem or may complicate an existing diagnosis of CPSS. Our last patient in this cohort (Case 6) had a delayed severe presentation, possibly because of the rarity of this entity and the limited knowledge about it. The child was managed as a case of asthma with persistent hypoxemia for almost a year before he was referred to us. His examination was significant for cyanosis secondary to worsening hepatopulmonary syndrome without pulmonary hypertension. Franchi et al. [6] postulated an explanation for hypoxemia in such cases. They suggested that PSS results in diversion of vasoactive mediators into the systemic circulation leading to dilatation of the intrapulmonary vessels and the subsequent development of ventilation-perfusion mismatch and hypoxemia.

Investigations for cases with suspected CPSS are typically initiated with Doppler US. It is the key imaging modality for diagnosis of the shunt, monitoring during the therapeutic procedure and future follow-up [6,17]. Computed tomography (CT) and Magnetic resonance (MR) imaging are used at a later stage for confirmation of the diagnosis as well as for delineation of the course of the shunt and its branches [1,2,17]. Work up should also include biochemical assessment and a search for malformations and syndromes, for its well-known high association [8,9,28,29], particularly congenital cardiac defects [29], and chromosomal abnormalities especially Down syndrome [24,28,30]. In our study, 5 out of 6 patients had other malformations involving different systems (Neurological, Cardiac, Gastrointestinal, Skin and Pulmonary - Table 1). The neurological and cardiac malformations identified in Case 3 appeared to indicate a poor prognosis.

Management of CPSS cases (conservative vs. interventional) should take into account several factors, mainly the shunt ratio, type of the shunt and the presence of complications [1—3,26]. Conservative management is usually effective in those cases that present with mild metabolic abnormalities and in some intrahepatic shunts that are asymptomatic and may regress spontaneously before the age of 2 years [1,6,15,20]. We opted for this approach in our asymptomatic patient (Case 4) and the prognosis was satisfactory. However, with large communications involving the extrahepatic portal veins and ductus venosus and in those with shunt ratio >30% that may persist throughout life and carry risks of complications, closure of the shunt is mandated [1,2,15,22]. Both surgical and endovascular embolization have been described as therapeutic options for shunt closure [6,13,14,20,27,30—38].

We performed transcatheter closure using AMPLATZER® Vascular Plugs in 4 of our patients. Indications for shunt closure in those patients were acute heart failure (Case 1), hypoxemia (Case 6), and a high shunt ratio of 70% and 40% in (Cases 2 & 5) respectively.

We think interventional catheterization using vascular plugs carries several advantages over the surgical option. It is minimally invasive, associated with less morbidity and the patient can be discharged home on the same day of the procedure. Furthermore, it can be carried out at any age in symptomatic patients, and with prenatal diagnosis early intervention is becoming more feasible [13,14,31]. The procedure was carried out on the second and seventh day of life in Case 1 and 2 respectively.

Transcather closure of the shunt provided effective treatment of CPSS complications in our patients. It has resulted in regression of associated skin and liver hemangiomas (Case 1 & 5), portal vessel growth in cases of portal hypoplasia (Case 6) [33], and normal growth and development of these children. After closure of the shunt, all metabolic abnormalities shown by abnormal liver enzymes and coagulation studies, elevated bile acids and hyperammonemia graduallly returned to normal levels within few days to weeks. These results are consistent with what has been previously reported [17,32,34,37,38].

Complications of interventional catheterization occur infrequently including plug dislocation and vein thrombosis which we experienced with Case (2). However in contrast to embolization coils [30,35,36], vascular plugs can be repositioned and removed if necessary [1]. The device place can also be precisely confirmed by US before release.

It is worth mentioning that some complicated cases in literature were managed by liver transplantation [39], and it may be necessary in the event where intrahepatic portal branches fail to form after shunt closure [6,40]. However, the experience gained over the past 15 years indicates that there is very little place left, if any, for liver transplantation in the management of congenital portosystemic shunts in children [17].

Based on our experience we conclude that interventional catheterization is an excellent therapeutic option for the treatment of selected cases of CPSS resulting in improved patient outcomes, and it is feasible at any age group. Long term follow up for several years is necessary in complicated cases, or in case of persistent complications such as pulmonary hypertension, and to detect the emergence of additional shunt that may require closure [17].

Conflicts of interest

Authors declare that they have no conflict of interest.

Ethical approval

This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent

For this Cohort study, no informed consent was required. Data reporting was anonymous. Patients' confidentiality was protected during data collection, and manuscript preparation.

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Compliance with ethical standards

The study was approved by Al Ain Medical District Human Research Ethics Committee — Protocol No. 13/76 (CRD 275/13).

Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijpam.2019.02.009.

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