Accepted: 7 December 2017

DOI: 10.1111/petr.13111

CASE REPORT

WILEY

Unusual spontaneous porto-systemic shunt: The importance of diagnosing non-anatomical porto-systemic shunts to improve portal flow in pediatric living–related liver transplantation. Case report

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Abstract

Collateral circulation secondary to liver cirrhosis may cause the development of large PSSs that may steal flow from the main portal circulation. It is important to identify these shunts prior to, or during the transplant surgery because they might cause an insufficient portal flow to the implanted graft. There are few reports of "steal flow syndrome" cases in pediatrics, even in biliary atresia patients that may have portal hypoplasia as an associated malformation. We present a 12-month-old female who received an uneventful LDLT from her mother, and the GRWR was 4.8. During the early post-operative period, she became hemodynamically unstable, developed ascites, and altered LFT. The post-operative ultrasound identified reversed portal flow, finding a non-anatomical PSS. A 3D CT scan confirmed the presence of a mesocaval shunt through the territory of the right gonadal vein, draining into the right iliac vein, with no portal inflow into the liver. The patient was re-operated, and the shunt was ligated. An intraoperative Doppler ultrasound showed adequate portal inflow after the procedure; the patient evolved satisfactorily and was discharged home on day number 49. The aim was to report a case of post-operative steal syndrome in a pediatric recipient due to a mesocaval shunt not diagnosed during the pretransplant evaluation.

KEYWORDS
living donor liver transplantation, portal thrombosis, porto-systemic shunt

1 | INTRODUCTION

The development of large collateral circulation is part of the pathophysiology of portal hypertension secondary to cirrhosis.[1] These collaterals may evolve into big spontaneous anatomical or non-anatomical PSSs that generate reversal portal flow into the systemic circulation, reducing the portal inflow. The degree of flow diversion may vary from a mild decrease to a complete absence of portal inflow, with the consequent portal thrombosis and reversed flow. The importance of identifying those dominant collaterals during the pretransplant evaluation has been very well described in adults,[2,3] as well as the subsequent need for post-transplant occlusion, but there is scattered information in this regard in pediatrics. In most cases, after a successful liver transplant, these large shunts tend to disappear due the low portal resistance present in the new liver; nevertheless, the spontaneous closure

Abbreviations: GRWR, graft recipient weight ratio; IMV, inferior mesenteric vein; INR, international normalized ratio; IVC, inferior vena cava; LDLT, living donor liver transplantation; LFT, liver function tests; LT, liver transplantation; PSS, porto-systemic shunt; SMV, superior mesenteric vein; US, ultrasound.

might be slow, delayed, or might not occur.[4,5] In such extreme cases, the persistence of the spontaneous shunts can cause "steal flow syndrome" into the new liver. Therefore, an early diagnosis of main portal flow reduction is mandatory to ligate or embolize the shunts, even in the operating room at the end of the case.[2] Mesocaval shunts are rare spontaneous porto-systemic collateral vessels between the superior and the IVC,[1] and more rarely, they involve the territory of the gonadal vein. Although the Doppler US is the gold standard test for preoperative portal flow evaluation,[6] mesocaval shunts may be undetected and could be only diagnosed in the post-transplantation period using Doppler US or angiotomography. The aim of this manuscript was to describe a case of a spontaneous right mesocaval shunt that required treatment after a pediatric LDLT in order to normalize the portal flow.

2 | PATIENTS AND METHODS

This is a 12-month-old female with biliary atresia and a failed Kasai. She presented with repeated episodes of cholangitis, ascites, and severe malnutrition (weight: 5550 g, height: 62 cm, Z-scores: −5.518 and −4.045, respectively). Pretransplant LFT are shown in Table 1. The patient underwent the pretransplant evaluation, and a LDLT was proposed. The donor was her mother, a 29-year-old woman with non-significant prior medical history. An uneventful orthotopic liver transplant was performed using the piggyback technique; the GRWR was 4.8. During the preanhepatic phase, a portal hypoplasia associated with biliary atresia was observed. For this reason, a venous interposition graft was built at the level of the SMV-PV junction using an isogroup cadaveric iliac vein graft. The donor left lateral segment was engrafted, and at the end of the procedure, arterial and venous flows were found to be normal clinically and checked with pulsatile and US Doppler. The surgical aspects are shown in Table 2. On the first post-operative day, the Doppler ultrasound reported absence of portal flow and inverted flow in the SMV. The patient was brought to the operating room, the vascular graft was removed due to partial thrombosis, secondary to the reversed portal flow, and flow was recovered at the end of the case. During the second post-operative day, the patient developed ascites, edema, and elevated liver enzymes (Table 1). A new Doppler ultrasound reported absence of portal flow with increased arterial peak flow and again inverted flow in the SMV, but found a vessel with reversed flow that brought the suspicion of a non-anatomical PSS. A CT scan was performed, confirming the presence of a non-anatomical meso-cava shunt through the territory of the right gonadal vein draining into the right main iliac vein (Figure 1), together with complete absence of portal flow. An exploratory laparotomy was then performed; the right colon was mobilized to expose the retro-peritoneum, identifying the mesocaval shunt. The shunt was encircled with silk sutures (Figure 2) and was progressively reduced the size of the shunt up to obtain a normal portal flow. A DUS and flowmeter proved the recovery of normal portal flow and velocity into the liver (Figure 3). Finally, the non-anatomical shunt was tied off and transected. At the end of the case, and daily during the following

TABLE 1 Recipient liver function test

Recipient LFT	Pre-op	POD1	POD2	After shunt closure
AST (IU/dL)	1076	674	889	348
ALT (IU/dL)	998	848	1009	489
TB (mg/dL)	32.5	6.5	2.8	2.5
AP (IU/dL)	143	125	74	50
GGT (IU/dL)	19	-	15	16
INR	1.5	2.31	2.8	2.1
Na (mEq/dL)	132	142	145	148
Creatinine (mg/dL)	0.5	0.4	0.3	0.3

TB, total bilirubin; AP, alkaline phosphatase; GGT, gamma-glutamyl-transpeptidase; INR, international normalized ratio; Na, sodium.

TABLE 2 Surgical aspects

Graft volume (mL)	316
CIT (minutes)	90
WIT (minutes)	45
TIT (minutes)	135
RBC transfusion (mL)	200
FFP transfusion (mL)	80
Piggyback	Yes
Artery reconstruction	End-to-end
Venous reconstruction	Cadaveric graft
Biliary reconstruction	Roux-en-Y
Immunosuppression	methylprednisolone
Others	Insulin

CIT, cold ischemia time; WIT, warm ischemia time; TIT, total ischemia time; RBC, red blood cells; FFP, fresh-frozen plasma.

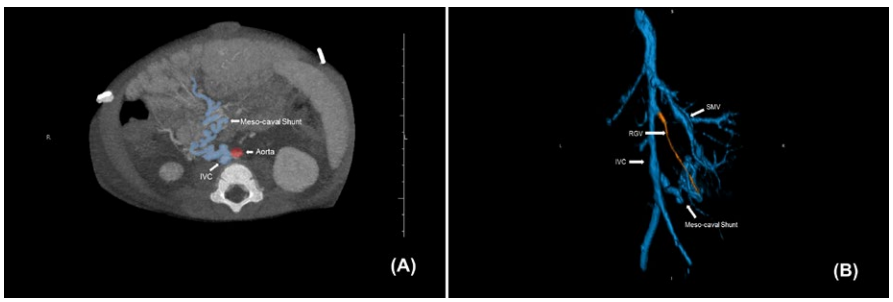

FIGURE 1 A, CT scan shows a mesocaval shunt. B, 3D reconstruction, RGV in orange. IVC, inferior vena cava; RGV, right gonadal vein; SMV, superior mesenteric vein

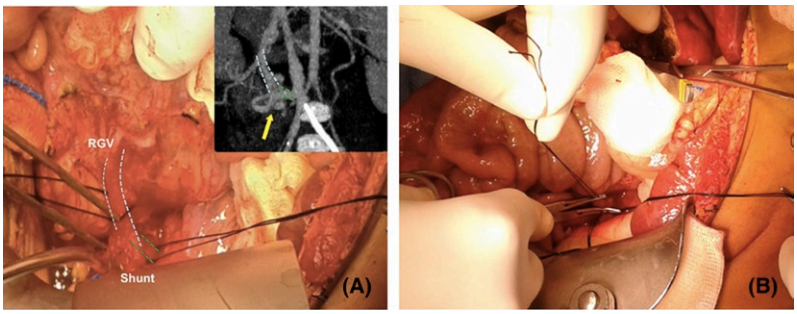

FIGURE 2 A. The RGV is mark with white dotted lines (in the CT and in the surgical picture); the shunt is marked with green dotted lines in both pictures. B. Picture of the RGV that is encircled with silk suture. The shunt is encircled with silk suture as well.

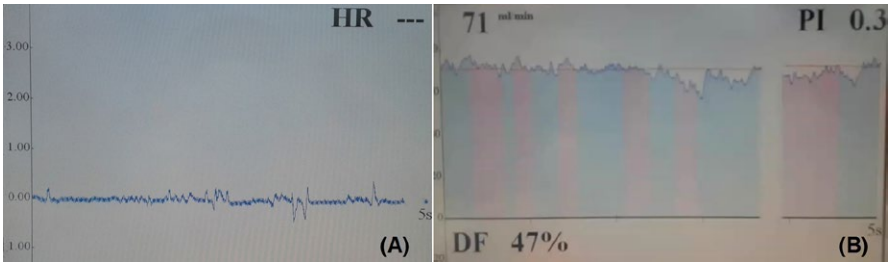

FIGURE 3 A. Flowmeter shows absence of portal flow with increased arterial peak flow before ligating the shunt. B. After the shunt was ligated, the flowmeter shows a normal portal inflow.

3 days, the portal venous graft was monitored by Doppler ultrasound and remained with normal flow. The patient normalized the liver functions and was sent home. For a follow-up period of 21 months, the patient showed catch-up growth and was able to maintain normal liver function and vascular flows.

3 | DISCUSSION

PSSs are reported to occur in approximately 18% of the patients evaluated for liver transplant.[7,8] Scattered reports describe this situation in the pediatric literature. When PSSs occur, the stealing flow syndrome causes reversion of the portal flow, increasing the risk of portal vein thrombosis. Unfortunately, shunts cannot always be recognized during the pretransplant evaluation, or even during the LT procedure in spite of the increasing quality of the image studies. The chances of diagnosis decrease when paired with the age of the patients due to the size of the vessels and the minimal flow that the method used could detect. Many patients with BA have hypoplasia or anatomical malformations of the portal vein; this has been reported as a risk factor for post-transplant complications.[9] Portal vein thrombosis due to portal hypoplasia is the most frequent complication, with a reported incidence between 4% and 16%.[10-12]

The presence of a low portal flow at the end of the engraftment should trigger suspicion of an existing shunt that might require treatment in the short or medium term. A triple-phase CT angiography with vascular reconstruction can identify the shunt; if it is performed prior to LT, it would allow a planned search to identify and ligate the shunt after or during the engraftment. Routine intraoperative portal vein flow measurements can provide the opportunity of identifying venous anomalies and to act accordingly during the transplant surgery.[4] Unfortunately, in some instances, these shunts are not identified preoperatively and cannot be assessed during surgery, or do not become clinically evident until the post-transplant period, as seen in the case reported here. Cases like this might indicate a change in our current evaluation guidelines, and maybe will be mandatory to include the use of three-phasic CT scan.

So far, there is no established algorithm to treat PSS before, during, or after liver transplant; the evidence on the efficacy of different treatment plans is largely limited to case reports; and there is no consensus on what or when the intervention is best for treating large or persistent anatomical or non-anatomical shunts.[5] Shunt ligation may be considered dangerous, in particular following living donor LT,[6] when an excessive portal vein flow could be found, but this is an extrapolation of the adult knowledge to pediatrics, and it might be wrong considering that most of the pediatric patients received a graft with a GRWR greater than 2, like this patient, who received a graft with a GRWR of 4.8. When the flowmeter is used to examine the flow, if the flow does not get back to normal after reperfusion, we should look for possible shunting. In this case, we use the flowmeter during the second relaparotomy, and we could prove the correct portal inflow.

Within the described therapeutic options are (i) preoperative TIPS to decompress the shunting; (ii) intraoperative shunt ligation; (iii) intraoperative assessment of portal flow with the intention of intervening if there is evidence of inadequate flow to the allograft; (iv) close monitoring of the shunt and functional status of the transplanted liver without any intervention for a small PSS; (v) creation of porto-renal anastomosis in case of portal vein thrombosis or post-operative percutaneous interventions such as shunt embolization.[5] Most of those recommendations are for the adult population, and they might or cannot be extrapolated to pediatrics.[13-15]

Therefore, the strategy we propose is to search for the shunts in the pretransplant period, to measure flows after the engraftment and act accordingly, and, finally, in the early post-operative period, to give careful attention to the Doppler US in order to identify possible shunts, and to act accordingly in order to improve long-term graft and patient survival.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

AUTHORS' CONTRIBUTIONS

Juan S. Rubio, Pablo A. Farinelli, Diego A. Ramisch, Hugo Paladini, Pablo D´Angelo, and Nicolás Aguirre: Contributed substantially; Juan S. Rubio, Carolina Rumbo, and Pablo Barros Schelotto: Drafted and revised the manuscript; Carolina Rumbo, Pablo Barros Schelotto, and Gabriel E. Gondolesi: Revised and approved the manuscript.

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REFERENCES

1. Boixadera H, Tomasello A, Quiroga S. Successful embolization of a spontaneous mesocaval shunt using the Amplatzer vascular plug II. *Cardiovasc Intervent Radiol*. 2010;33:1044-1048.
2. Kim S-J, Kim D-G, Park J-H, et al. Clinical analysis of living donor liver transplantation in patients with portal vein thrombosis. *Clin Transplant*. 2010;25:111-118.
3. Hwang S, Kim DY, Ahn CS, et al. Computational simulation-based vessel interposition reconstruction technique for portal vein hypoplasia in pediatric liver transplantation. *Transplant Proc*. 2013;45:255-258.
4. Chmurowicz T, Zasada-cedro K, Wojcicki M. Cavoportal hemitransposition for unrecognized spontaneous mesocaval shunt after liver transplantation: a case report. *Transpl Int*. 2013;26:1-4.
5. Awad N, Horrow MM, Parsikia A, et al. Perioperative management of spontaneous splenorenal shunts in orthotopic liver transplant patients. *Exp Clin Transplant*. 2012;5:475-481.
6. Aucejo FN, Hashimoto K, Quintini C, et al. Triple-phase computed tomography and intraoperative flow measurements improve the management of portosystemic shunts during liver transplantation. *Liver Transpl*. 2008;14:96-99.
7. Cherqui D, Panis Y, Gheung P, et al. Spontaneous portosystemic shunts in cirrhotics: implications for orthotopic liver transplantation. *Transplant Proc*. 1993;25:1120-1121.
8. Ploeg RJ, D'Alessandro AM, Stegall MD, et al. Effect of surgical and spontaneous portasystemic shunts on liver transplantation. *Transplant Proc*. 1993;25:1946-1948.
9. Gurevich M, Guy-Viterbo V, Janssen M, et al. Living donor liver transplantation in children. *Ann Surg*. 2015;262:1141-1149.
10. de Magnée C, Bourdeaux C, De Dobbeleer F, et al. Impact of pretransplant liver hemodynamics and portal reconstruction techniques on post-transplant portal vein complications in pediatric liver transplantation: a retrospective analysis in 197 recipients. *Ann Surg*. 2011;254:55-61.
11. Tannuri ACA, Gibelli NEM, Ricardi LRS, et al. Orthotopic liver transplantation in biliary atresia: a single-center experience. *Transplant Proc*. 2011;43:181-183.
12. Matsumoto R, Uchida K, Nishida S, et al. Extra-anatomical mesoportal venous jump graft repair for early portal vein thrombosis after liver transplant in an infant with a hypoplastic portal vein: a case report. *Transplant Proc*. 2016;48:3186-3190.
13. Sato Y, Yamamoto S, Takeishi T, et al. Management of major portosystemic shunting in small-for-size adult living-related donor liver transplantation with a left-sided. *Surg Today*. 2006;36:354-360.
14. Ikegami T, Shirabe K, Nakagawara H, et al. Obstructing spontaneous major shunt vessels is mandatory to keep adequate portal inflow in living-donor liver transplantation. *Transplantation*. 2013;95:1270-1277.
15. Abujudeh HH, Samanta AK, Cho KC, Klein KM. Spontaneous portosystemic shunting several years following liver transplantation: successful treatment via percutaneous embolization. *Liver Transpl*. 2004;10:324-326.

How to cite this article: Rubio JS, Rumbo C, Farinelli PA, et al. Unusual spontaneous porto-systemic shunt: The importance of diagnosing non-anatomical porto-systemic shunts to improve portal flow in pediatric living–related liver transplantation. Case report. *Pediatr Transplant*. 2018;22:e13111. https://doi.org/10.1111/petr.13111