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CASE REPORT

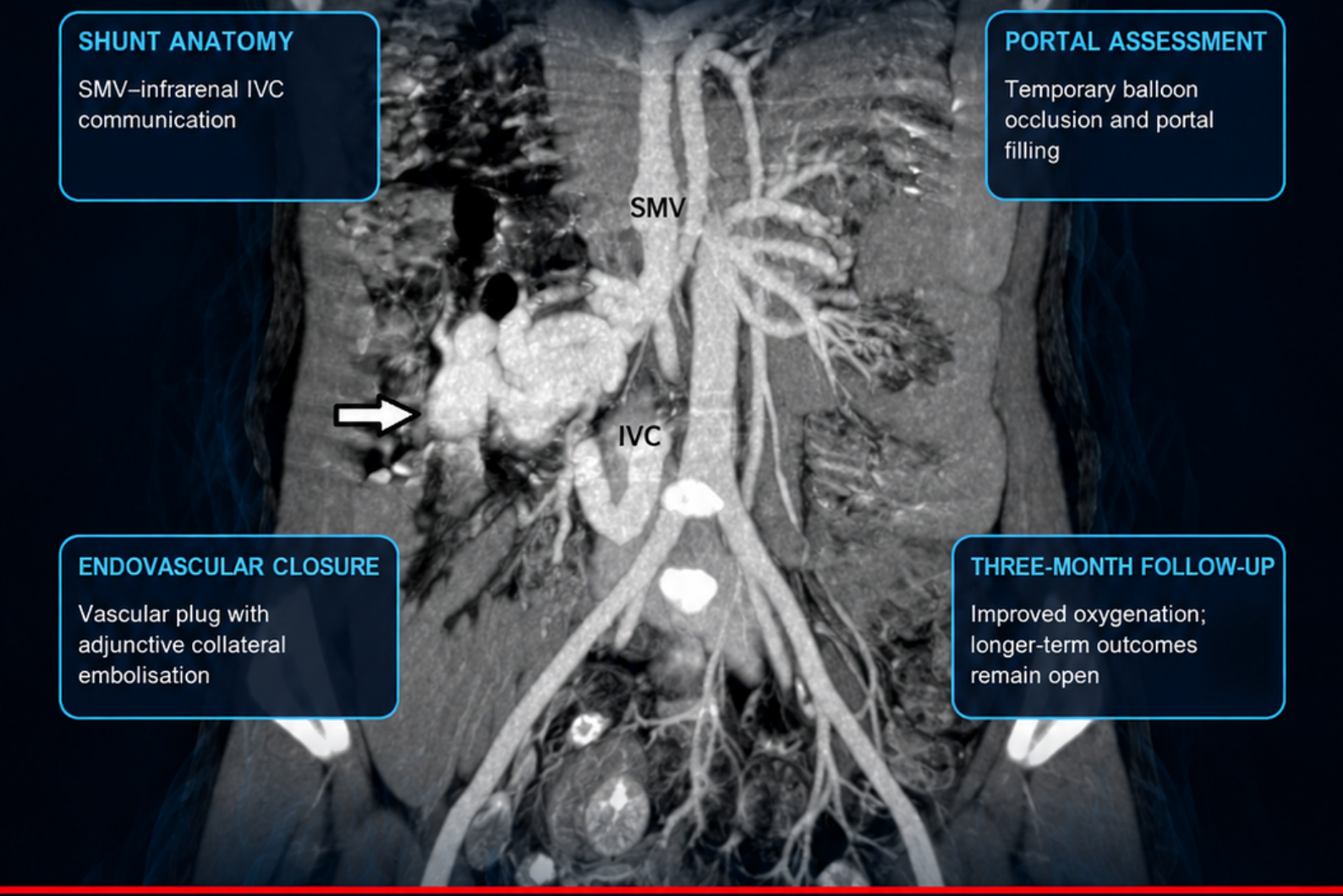
# TYPE II ABERNETHY MALFORMATION WITH HEPATOPULMONARY SYNDROME

## SINGLE-STAGE ENDOVASCULAR SHUNT OCCLUSION

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CHENNAI, INDIA

**SHUNT ANATOMY**
SMV–infrarenal IVC communication

**PORTAL ASSESSMENT**
Temporary balloon occlusion and portal filling




**ENDOVASCULAR CLOSURE**
Vascular plug with adjunctive collateral embolisation

**THREE-MONTH FOLLOW-UP**
Improved oxygenation; longer-term outcomes remain open

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# Type II Abernethy Malformation with Hepatopulmonary Syndrome: Single-Stage Endovascular Occlusion of an SMV–IVC Communication — A Case Report

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## Abstract

*Background: Congenital extrahepatic portosystemic shunts (Abernethy malformations) can be associated with hyperammonemia and hepatopulmonary syndrome (HPS). Endovascular closure requires careful assessment of shunt anatomy and portal venous drainage.*

*Case presentation: A 12-year-old girl presented with progressive exertional dyspnea, central cyanosis, clubbing, and room-air $SpO_2$ of 74%. Serum ammonia was 103 µmol/L, while liver biochemistry was within the reference ranges. Echocardiography showed no structural cardiac cause for the cyanosis and agitated saline bubble echocardiography showed delayed appearance (6 cardiac cycles) of bubbles in the left atrium. CT pulmonary angiography demonstrated distal pulmonary vascular dilatation and subpleural telangiectatic vessels, consistent with HPS. The preprocedural ABG on room air showed pH 7.45, $PaCO_2$ 31 mmHg, $PaO_2$ 62 mmHg, $HCO_3^-$ 22 mEq/L, and A–a oxygen gradient of 49 mmHg; the follow-up ABG normalized. The baseline $SpO_2$/$PaO_2$ discrepancy remained unresolved. Abdominal CT angiography showed a side-to-side communication between the superior mesenteric vein (SMV) and infrarenal inferior vena cava (IVC), with tortuous collateral channels and normal, patent intrahepatic portal branches. During temporary balloon occlusion, portal pressure rose from 7 to 17 mmHg, and contrast opacified the intrahepatic portal branches and hepatic parenchyma. The primary communication was occluded with a 12-mm Amplatzer Vascular Plug II, with adjunctive gelfoam embolisation of collateral channels.*

*Outcome: Doppler ultrasound demonstrated hepatopetal portal flow during early follow-up. At 3 months, room-air $SpO_2$ was 94%, central cyanosis had resolved, serum ammonia had decreased to 46.8 µmol/L, and the follow-up ABG normalized. Conclusion: This case highlights the role of cross-sectional vascular mapping and temporary balloon occlusion assessment before single-stage endovascular closure of a congenital portosystemic shunt.*

**Keywords: Abernethy malformation; hepatopulmonary syndrome; plug embolisation**

## 1. Introduction & Historical Context

Abernethy malformation, also termed congenital portosystemic shunt (CPSS), is a rare vascular anomaly in which portal venous blood is diverted partly or completely into the systemic circulation. Clinical manifestations vary and may include hyperammonemia, hepatic nodular lesions, hepatopulmonary syndrome (HPS), and portopulmonary hypertension. We report a child with a side-to-side superior mesenteric vein (SMV)–infrarenal inferior vena cava (IVC) communication, marked hypoxemia, and imaging findings consistent with HPS, treated by single-stage endovascular occlusion. [1–4]

## 1.1 Historical Context & Classification Evolution

Abernethy malformation was described by John Abernethy in 1793 [5]. The Morgan–Superina classification distinguishes Type I shunts, in which intrahepatic portal venous branches are absent, from Type II shunts, in which a portosystemic communication coexists with a patent intrahepatic portal venous system. This distinction is clinically important because the presence and adequacy of intrahepatic portal branches influence whether definitive shunt closure is feasible. [6]

Type I shunts are characterised by absent intrahepatic portal venous branches and complete diversion of portal flow. Type II shunts retain intrahepatic portal branches, with a variable proportion of portal blood diverted through an abnormal communication. Classification and assessment of portal venous anatomy should be based on cross-sectional imaging and, when closure is contemplated, catheter angiography and temporary occlusion assessment. [6–10]

The anatomical configuration and systemic drainage site vary among patients. In this case, cross-sectional imaging demonstrated a side-to-side communication between the SMV and infrarenal IVC, with associated tortuous collateral channels. The intrahepatic portal branches were normal.

Management depends on shunt anatomy, intrahepatic portal venous development, associated complications, and hemodynamic response to temporary occlusion. The management of Type I shunts depends on the actual portal anatomy and clinical context, and liver transplantation may be considered in selected patients. Selected Type II shunts can be treated by endovascular device closure or surgery; staged closure may be considered when temporary occlusion suggests inadequate portal venous capacity or a concerning hemodynamic response. [2, 8–10, 12]

## 1.2 Therapeutic Mandate & Interventional Principles

Accurate delineation of the shunt and intrahepatic portal venous anatomy is central to treatment planning. Cross-sectional imaging defines the anatomy and associated abnormalities, while catheter venography and temporary balloon

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occlusion can demonstrate portal filling and provide hemodynamic information before permanent closure. Treatment should be individualized by a multidisciplinary team. [1, 2, 6–10]

## 2. Case Presentation

### 2.1 Clinical Presentation & Baseline Evaluation

A 12-year-old girl presented with a 6-month history of progressive exertional dyspnoea (NYHA functional class II), fatigue, and bluish discoloration of the lips, fingers, and toes. She weighed about 29 kg. Examination showed central cyanosis and grade III clubbing of the fingers and toes. Room-air oxygen saturation ($SpO_2$) was 74% in the supine position and the preprocedural ABG was obtained at the same time. Oxygen saturation improved to 97% with supplemental oxygen. Scleral icterus and clinical stigmata of chronic liver disease were absent (Figure 1).

Laboratory testing showed haemoglobin of 15.2 g/dL and haematocrit of 46%. Serum ammonia was elevated at 103 µmol/L. Liver biochemistry was within the reference ranges (AST 34 U/L, ALT 24 U/L, total bilirubin 0.9 mg/dL, albumin 4.2 g/dL, and alkaline phosphatase 142 U/L). INR was 1.25 and APTT was 28 seconds.

### 2.2 Diagnostic Workup & Imaging Delineation

Because of central cyanosis, clubbing, and marked hypoxemia, cardiac, pulmonary, and abdominal vascular imaging was performed to investigate the cause.

1. Transthoracic Echocardiography (TTE): Transthoracic echocardiography demonstrated a structurally and functionally normal heart with normal intracardiac anatomy, normal biventricular systolic performance, intact atrial and ventricular septa, and no evidence of cyanotic congenital heart disease (ruling out atrial or ventricular septal defects, patent ductus arteriosus, Tetralogy of Fallot, transposition of great arteries, or total anomalous pulmonary venous connection). Establishing a structurally normal heart is essential in pediatric patients presenting with cyanosis and clubbing to exclude primary cardiac defects that frequently masquerade as the primary diagnosis. This normal cardiac finding redirected the diagnostic evaluation toward an extra-cardiac etiology and transthoracic echocardiography was performed following intravenous administration of agitated saline contrast, with assessment of microbubble passage through the right and left cardiac chambers. Microbubbles were observed in the left atrium after approximately 6 cardiac cycles following right atrial opacification suggesting an intrapulmonary right-to-left shunt.

2. ABG: The preprocedural arterial blood gas analysis obtained on room air, in the supine position at the same time as the $SpO_2$ measurement showed pH 7.45, $PaCO_2$ 31 mmHg, $PaO_2$ 62 mmHg, $HCO_3^-$ 22 mEq/L and an A–a oxygen gradient of 49 mmHg (assuming $FiO_2$: 0.21; atmospheric pressure ($P_{atm}$): 760 mmHg; water vapour pressure (PH2O): 47 mmHg; respiratory quotient (R): 0.8). This showed hypoxemia, hypocapnia and an elevated A–a oxygen gradient which indicated poor oxygenation and possibility of hepatopulmonary syndrome.

3. Thoracic CT Pulmonary Angiography (CTPA) and Catheter pulmonary angiogram: Thoracic CTPA was performed to evaluate the pulmonary vascular tree. CTPA manifested as distal vascular dilatation with subpleural telangiectasia predominantly affecting the subpleural lung bases, without evidence of pulmonary thromboembolism, macro-arteriovenous malformations, or parenchymal lung disease. Catheter pulmonary angiogram under fluoroscopy was done during the evaluation showed similar distal vascular dilatations. This characteristic morphological pattern represents the classic imaging hallmark of intrapulmonary vascular dilations (IPVDs) in Hepatopulmonary Syndrome (HPS) (Figure 2).

4. Abdominal Color Doppler Ultrasound & CTA: Doppler ultrasound demonstrated a patent superior mesenteric vein (SMV) and splenic vein, normal intrahepatic portal branches within the liver hilum. It showed few tortuous venous channels in RIF. Multidetector Computed Tomography Angiography (CTA) with 3D volume-rendered reconstruction established the detailed anatomy of a Type II Abernethy malformation. Imaging revealed a direct side-to-side extrahepatic shunt originating from the SMV and draining directly into the infrarenal inferior vena cava (IVC), accompanied by tortuous venous collateral channels. Right and left intrahepatic portal venous branches appeared normal and patent. No focal hepatic masses (FNH, adenoma, HCC) or signs of portal hypertension (varices, splenomegaly, ascites) were present (Figures 3 and 4).

### 2.3 Angiographic Workup & Temporary Balloon Occlusion Test

Under general anaesthesia, venous access was obtained through the right internal jugular vein. The IVC was catheterised and a long sheath positioned in the IVC. The shunt was selectively catheterised from the infrarenal IVC, and venography was performed to delineate the communication and collateral channels (Figure 5).

Temporary balloon occlusion was performed using a compliant balloon to assess intrahepatic portal venous filling and hemodynamic response before definitive closure.

During temporary balloon occlusion, contrast injection demonstrated opacification of SMV and main portal vein with flow through the right and left intrahepatic portal branches and opacification of the hepatic parenchyma. This confirmed visible intrahepatic portal venous drainage during occlusion (Figure 6).

Portal pressure was measured through the central lumen of the occlusion balloon connected to a pressure transducer. The baseline portal pressure was 7 mmHg and the pressure during temporary balloon occlusion was 17 mmHg, a rise of 10 mmHg. On the basis of the absolute occlusion pressure, patent intrahepatic portal branches and angiographically demonstrated portal flow, the treating team proceeded with single-stage closure during the same procedure. Systemic venous pressure was not recorded during the procedure.

## 2.4 Single-Stage Plug Embolisation & Post-Procedural Recovery

The shunt measured 8 mm; a 12-mm vascular plug was selected to provide 50% oversizing. A 12-mm Amplatzer Vascular Plug II (Abbott, Abbott Park, IL, USA) was advanced transjugularly and deployed across the primary SMV – infrarenal IVC communication near its IVC end (Figure 7).

To prevent residual shunting through adjacent tortuous peri-shunt venous channels, supplementary gelfoam embolisation of the collateral vessels was performed through the catheter positioned para-axially distally in the shunt from the IVC through another IJV access. Completion angiography demonstrated no residual filling of the primary shunt. Post procedure CT confirmed the appropriate position of the vascular plug (Figures 8 and 9).

The post-procedural course was uneventful. The 29-kg patient received prophylactic anticoagulation according to our institutional protocol, with enoxaparin 40 mg subcutaneously once daily for 3 days. Apixaban 2.5 mg twice daily was started on post-operative day 3, overlapping with enoxaparin on that day, and continued for 3 months from post-operative day 3 to prevent SMV or portal vein thrombosis. Dosing followed our institutional protocol (fixed doses). Monitoring consisted of Doppler ultrasound on post-procedure days 1 and 3 which showed stable device position, shunt occlusion, and hepatopetal intrahepatic portal flow. Clinical surveillance focused on bleeding manifestations. The patient and her parents were counselled to seek medical review if bleeding occurred. No laboratory assessment of anticoagulant intensity is documented, and further details of the dosing rationale are unavailable in the records (Figure 10).

At 3-month follow-up, room-air $SpO_2$ was 94% and central cyanosis had resolved. The ABG obtained simultaneously with the $SpO_2$ measurement had normalised (pH 7.40, $PaCO_2$ 38 mmHg, $PaO_2$ 92 mmHg, $HCO_3^-$ 23 mEq/L, estimated A–a gradient approximately 10.2 mmHg on room air using same assumptions made during pre-procedure ABG). Serum ammonia decreased to 46.8 µmol/L. No bleeding manifestations were reported during the three-month follow-up.

At the 3-month follow-up, the parent reported that the child was attending school and carrying out her usual activities without substantial difficulty.

Longer-term outcomes beyond the documented follow-up are not yet available.

Table 1. Diagnostic summary, hemodynamic measurements and clinical outcomes.

| Parameter / Modality | Pre-Intervention Baseline | Post-Embolisation Outcome |
|---|---|---|
| **Shunt Anatomy (CTA)** | Side-to-side SMV-to-infrarenal IVC shunt; tortuous venous collaterals; normal intrahepatic portal veins | Complete plug occlusion of shunt & gelfoam embolization of tortuous channels. |
| **Pulmonary & Oxygenation** | Room air $SpO_2$ 74% (corrects to 97% on oxygen); TTE shows normal heart; CTPA shows distal vascular dilatation & subpleural telangiectasia; cyanosis & clubbing present | Room air $SpO_2$ 94%; complete regression of central cyanosis over 3 months. |
| **Serum Ammonia** | Elevated: 103 µmol/L (normal reference 7–30 µmol/L) | Reduced to 46.8 µmol/L post-procedure at 3-month follow-up. |
| **Portal Hemodynamics** | Baseline portal pressure 7 mmHg; balloon occlusion portal pressure 17 mmHg (Pressure rise 10 mmHg) | Doppler on days 1 and 3 showed good hepatopetal flow; no clinical features of portal hypertension at 3 months. Post-treatment portal pressure was not measured, and long-term exclusion of portal hypertension is not possible. |
| **Liver Function & Masses** | AST 34 U/L, ALT 24 U/L, albumin 4.2 g/dL, INR 1.25; zero focal liver masses on US/CTA | Follow-up liver biochemistry values are not reported in the available case description. |

Table 2. Clinical timeline.

| Time point | Clinical events and findings |
|---|---|
| **Approximately 6 months before presentation** | Progressive exertional dyspnoea (NYHA class II), fatigue, and bluish discoloration of lips, fingers, and toes. |
| **Initial presentation** | 12-year-old girl with central cyanosis, grade III clubbing, and room-air $SpO_2$ 74%; saturation improved to 97% with supplemental oxygen. Serum ammonia 103 µmol/L; liver biochemistry within reference ranges. |
| **Diagnostic evaluation** | Echocardiography showed normal cardiac anatomy and delayed microbubble passage into the left atrium after approximately 6 cycles, suggesting an intrapulmonary shunt. CTPA and catheter pulmonary angiogram showed distal pulmonary vascular dilatation and subpleural telangiectasia. ABG and abdominal Doppler/CTA supported HPS associated with a Type II extrahepatic portosystemic shunt. |
| **Angiographic assessment / temporary occlusion** | Transjugular venous access and shunt venography delineated the SMV–infrarenal IVC communication and collateral channels. Temporary balloon occlusion demonstrated filling of right and left intrahepatic portal branches and hepatic parenchyma. Portal pressure rose from 7 to 17 mmHg. |
| **Definitive intervention** | During the same procedure after angiographic assessment / temporary occlusion a 12-mm Amplatzer Vascular Plug II was deployed across the primary communication; adjunctive gelfoam embolisation was performed for collateral channels. Completion angiography showed no residual filling of the primary shunt; post-procedure CT confirmed device position. |
| **Post-procedure days 1 to 3** | Doppler ultrasound showed stable device position, shunt occlusion, and hepatopetal intrahepatic portal flow. An uneventful post-procedural course and prophylactic anticoagulation. |
| **Three-month follow-up** | Room-air $SpO_2$ 94%; central cyanosis resolved; ABG reported as normalised ($PaO_2$ 92 mmHg); serum ammonia decreased to 46.8 µmol/L. |
| **Beyond 3 months** | Longer-term outcomes are not yet available |

## Imaging Findings



Figure 1. Clinical picture of the patient's hand showing pandigital clubbing.

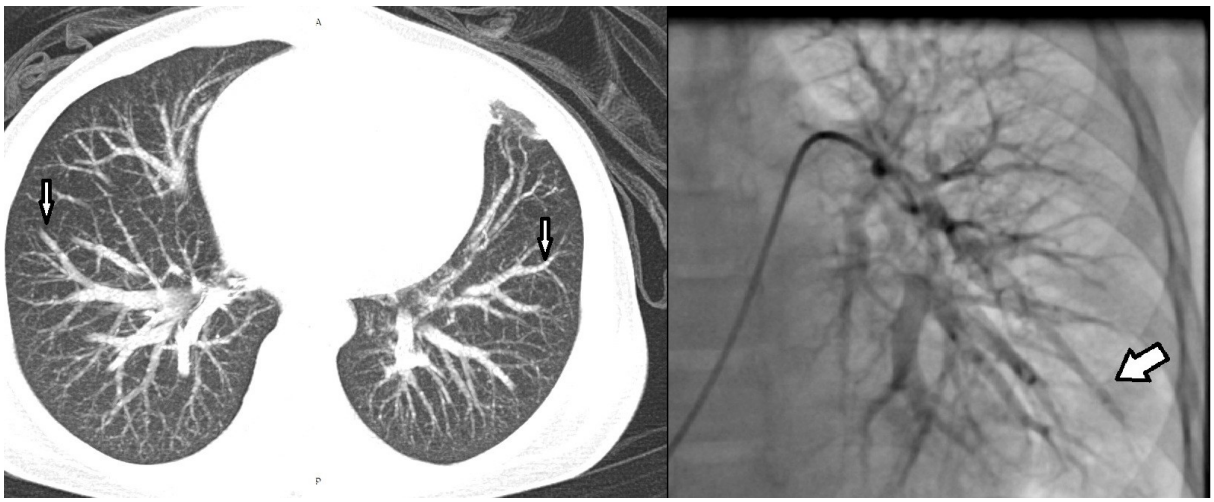


Figure 2. Axial MIP image in lung window showing pulmonary vascular dilatation up to the periphery (arrows) and catheter angiogram of left pulmonary artery showing the same (arrow).

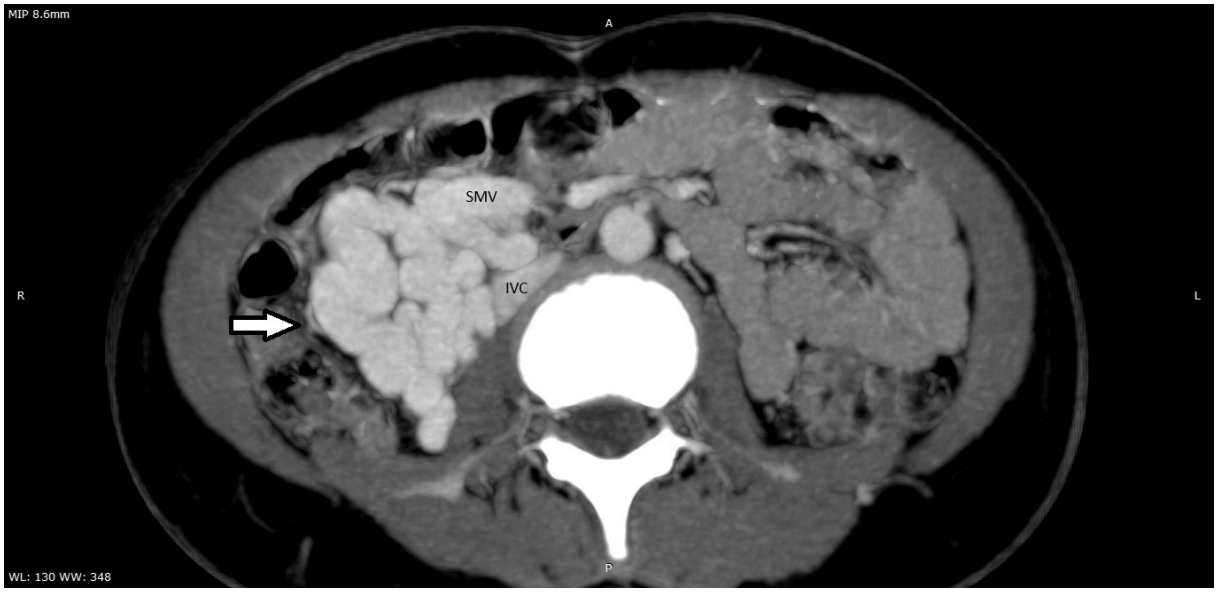



Figure 3. Axial MIP contrast-enhanced CT image showing the SMV and the tortuous shunt (arrow) draining into the IVC.




Figure 4. Coronal MIP contrast-enhanced CT image showing the tortuous shunt (arrow) draining into the IVC.

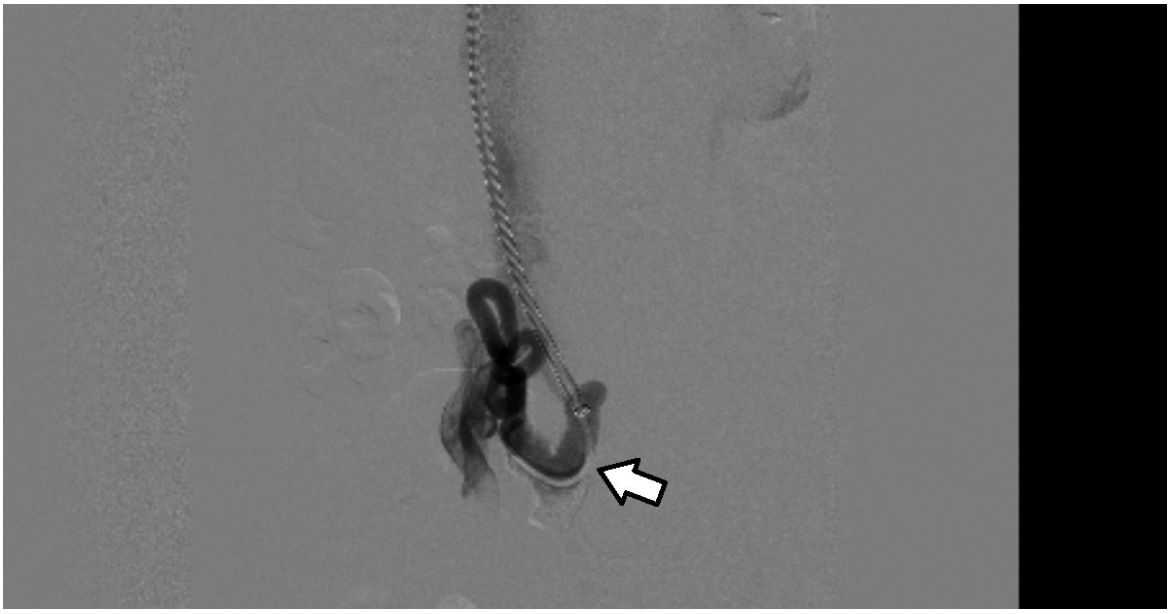


Figure 5. Cannulation of the shunt from IVC side and venogram showing the shunt (arrow)



Figure 6. Balloon occlusion venogram using a balloon catheter (black arrow) showing good flow through portal vein (blue arrow) and parenchymal blush.

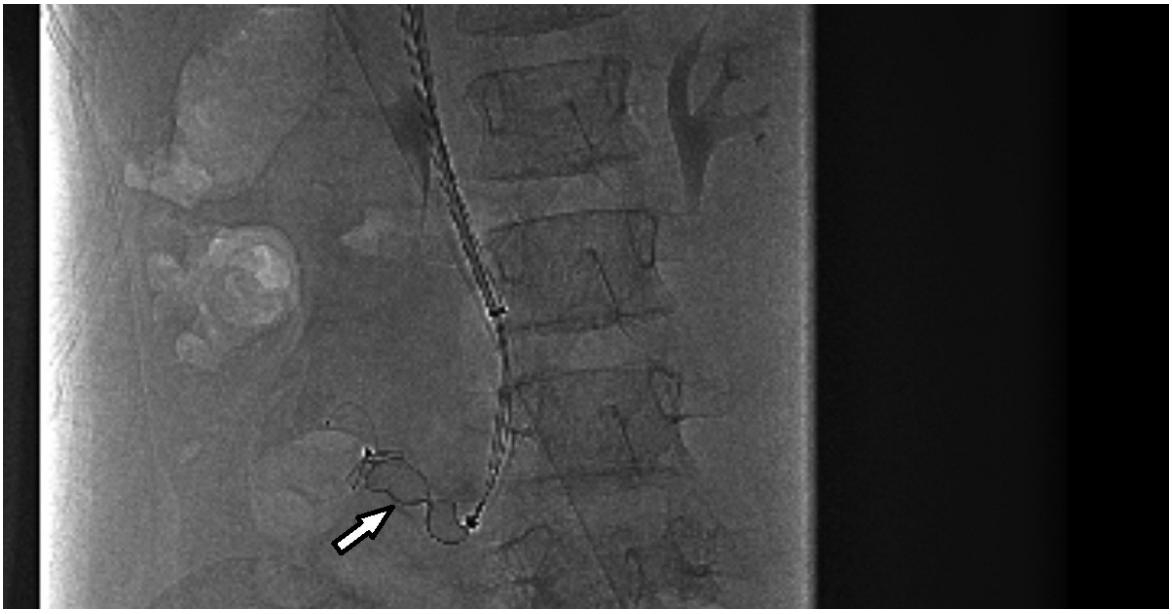


Figure 7. AVP II plug (arrow) deployed in the shunt close to IVC.

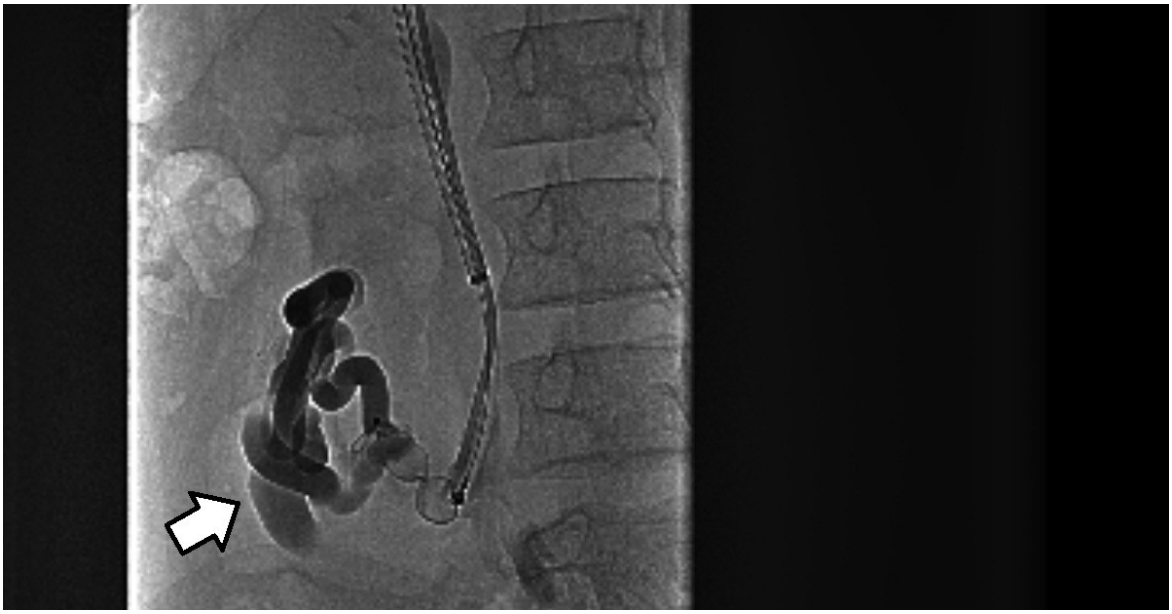


Figure 8. Angiographic image of the tortuous collateral vessels (arrow), treated with adjunctive Gelfoam embolisation.

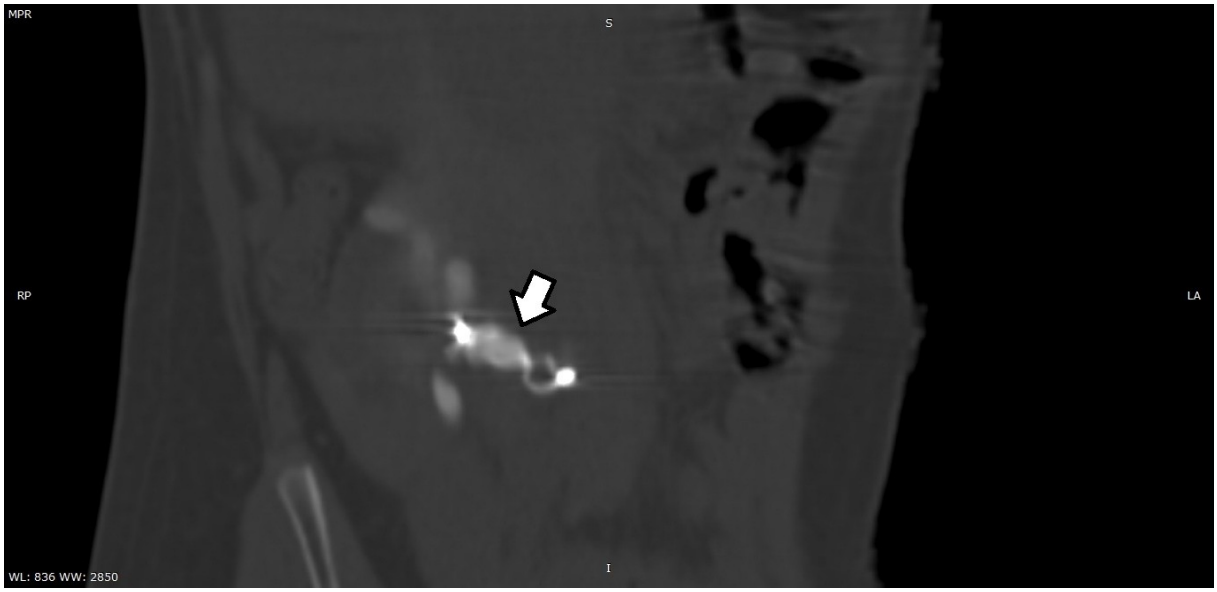



Figure 9. Post-procedure CT confirming the appropriate position of AVP II plug (arrow) in the shunt.

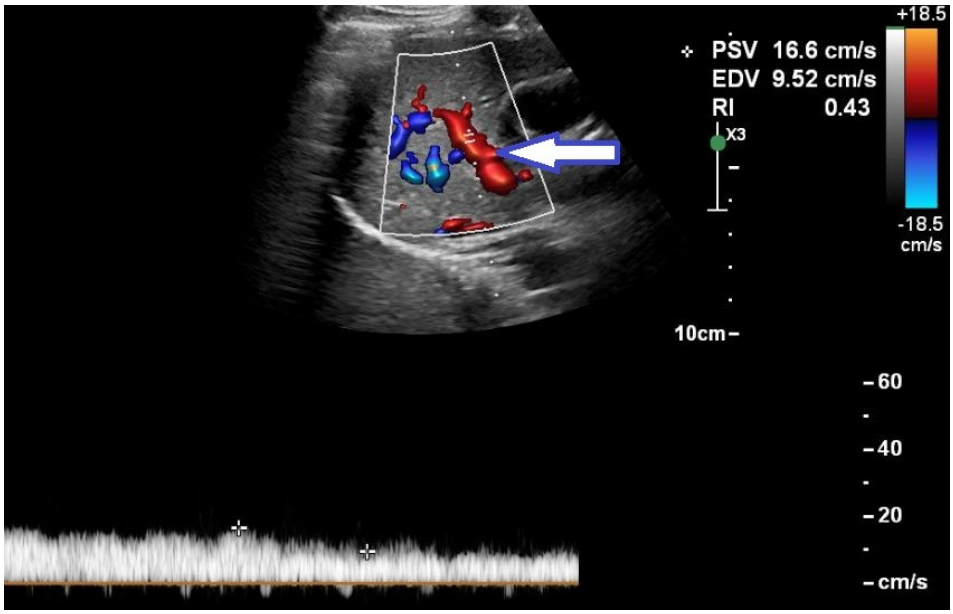



Figure 10. Post-procedure (day 3) portal vein Doppler showing the main portal vein (arrow) with hepatopetal flow.

## 3. Discussion

### 3.1 Embryological Mechanisms & SMV-Infrarenal IVC Shunt Anatomy

Congenital portosystemic shunts are thought to arise from abnormal development and remodelling of the embryonic venous system. The resulting communication allows some or all splanchnic venous blood to bypass hepatic first-pass circulation. [3]

In this patient, the side-to-side SMV–infrarenal IVC communication coexisted with normal, patent intrahepatic portal branches on imaging and during temporary balloon occlusion. Demonstration of portal filling during occlusion was an important procedural finding supporting the feasibility of shunt closure.

### 3.2 Diagnostic Trap: Differentiating Abernethy Malformation from Cyanotic Heart Disease

Cyanosis, clubbing, and marked hypoxemia in a child prompt evaluation for congenital heart disease as well as extracardiac causes. In this case, echocardiography did not identify a structural cardiac cause, prompting evaluation for extracardiac causes of hypoxemia. Agitated saline echocardiography showed delayed appearance of bubbles in the left atrium after 6 cycles indicating pulmonary right to left shunting. [1, 11]

The preprocedural ABG obtained on room air and supine position showed pH 7.45, $PaCO_2$ 31 mmHg, $PaO_2$ 62 mmHg, $HCO_3^-$ 22 mEq/L, and an A–a oxygen gradient of 49 mmHg. At 3 months, the follow-up ABG normalized which is consistent with CPSS-related shunting as the mechanism of hypoxemia, although causation cannot be established from a single case.

The simultaneous baseline $SpO_2$ of 74% and $PaO_2$ of 62 mmHg were rechecked against the available records and were correctly transcribed. The discrepancy could not be resolved from those records and remains a diagnostic limitation when interpreting baseline oxygenation and its subsequent improvement.

### 3.3 Pathophysiology of Hepatopulmonary Syndrome in Portosystemic Shunting

HPS is characterised by impaired arterial oxygenation and intrapulmonary vascular dilatation in the setting of liver disease or portal-systemic shunting. In this case, the preprocedural ABG showed $PaO_2$ 62 mmHg and A–a oxygen gradient of 49 mmHg which showed the impaired arterial oxygenation. The intrapulmonary vascular dilatation was confirmed by CTPA and contrast bubble echocardiography.

Portosystemic shunting may contribute to intrapulmonary vascular dilatation through altered handling of vasoactive substances and inflammatory signalling. Increased pulmonary nitric oxide production, endothelial signalling and intravascular macrophage-related pathways have been implicated in HPS. The resulting ventilation–perfusion mismatch, diffusion limitation and intrapulmonary shunting can impair arterial oxygenation. These mechanisms were not directly investigated in this patient. [3, 4]

Following shunt closure, room-air $SpO_2$ improved from 74% at presentation to 94% at 3 months, and central cyanosis resolved. Serum ammonia also decreased and ABG normalized. These observations are consistent with clinical improvement after restoration of portal flow through the liver, although the duration of follow-up does not establish long-term resolution of all pulmonary vascular changes.

### 3.4 Role of Balloon Occlusion Testing and Interventional Device Selection

CT or MR angiography helps define shunt anatomy, but catheter angiography with temporary balloon occlusion can provide additional information about portal venous filling and the hemodynamic response to closure. The test is particularly useful when deciding between single-stage and staged treatment. [1, 2, 12]

Occlusion-test pressures are context-dependent guidance, not universal safety cut-offs. Bruckheimer et al. used an occlusion pressure above 18 mmHg as one reason to consider staged treatment when portal vessels were diminutive. McLin et al. recommend measuring the portosystemic pressure gradient during the occlusion test, as it is considered more reliable than absolute portal pressure. They note that an absolute mesenteric pressure >30 mmHg or a gradient >20 mmHg is considered by some centres an indication for two-stage closure, that either approach may be considered for gradients of 10–20 mmHg depending on anatomical and clinical features, and that the final decision is made during the procedure. In our patient, pressure measured through the occlusion balloon rose from 7 to 17 mmHg, a rise of 10 mmHg. Systemic venous pressure was not recorded, so this is not a portosystemic gradient. Single-stage closure was chosen on the basis of the absolute pressure, the portal anatomy and the good portal flow seen during occlusion, and this is not presented as a general rule. [2, 12]

Endovascular occlusion with a vascular plug or other occlusion device is one treatment option for selected congenital portosystemic shunts. A range of device-based approaches has been described in published reports. [13–15] Device choice depends on shunt diameter, length, configuration, landing zone, and flow. In this case, a 12-mm Amplatzer Vascular Plug II was used for the primary communication, with adjunctive gelfoam embolisation of tortuous collateral channels. [2, 8, 13, 14]

### 3.5 Anticoagulation and Follow-up

Thrombosis upstream of the occlusion is a recognised concern after shunt closure, and expert guidance recommends prophylactic anticoagulation according to local protocols. The regimen used in this patient is described in Section 2.4; the available records do not permit independent assessment of its dosing rationale. Ongoing clinical and Doppler surveillance is important. [2]

Eliminating portosystemic bypass re-establishes hepatic clearance of gut-derived toxins and vasoactive substances. Closure is recommended for CPSS with systemic complications [2], and published series report improvement of CPSS-related manifestations after closure [8]. Consistent with this, our patient showed improvement in hypoxemia at 3 months.

## 4. Conclusion

Type II Abernethy malformation featuring a direct side-to-side anastomosis between the superior mesenteric vein and the infrarenal inferior vena cava is a rare but correctable vascular anomaly that can present with Hepatopulmonary Syndrome and hyperammonemia. This case illustrates the value of detailed vascular mapping and hemodynamic assessment when planning endovascular closure. Establishing a normal cardiac anatomical baseline on echocardiography is vital to avoid misdiagnosing cyanotic heart disease. Single-stage endovascular plug embolisation supplemented by gelfoam embolisation of tortuous collaterals provides a minimally invasive treatment that, in this patient, was followed by improved portal perfusion, a reduction in ammonia from 103 to 46.8 µmol/L (still above the reference range) and improved oxygenation at 3 months. These findings are limited to this patient and the three-month follow-up. Longer-term pulmonary vascular outcomes require further follow-up.

## Author Contributions

Rajakumar Ramar: Conceptualisation, clinical investigation, data collection and manuscript preparation.

Shivashankar Kumaresan: Methodology.

Kirubakaran Balu, Benin Reejan and Sibi Elango: Critical revision of the manuscript.

Babu Peter Sathyanathan: Supervision.

## Declarations

**Consent for publication:** Written informed consent for publication of clinical details and images was obtained from the patient's parent.

**Ethics committee approval:** Ethics committee approval was not required for this case report according to the institutional protocol at Madras Medical College.

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**Competing interests:** The authors declare no competing interests.

**Author approval:** All authors reviewed and approved the submitted revised manuscript and agree to be accountable for the work.

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