# Abernethy Syndrome Complicated by Severe Porto-Pulmonary Hypertension and Giant Pulmonary Artery Ectasia with Chronic *in Situ* Thrombosis: A Longitudinal Case Report

## Síndrome de Abernethy Complicada de Hipertensão Porto-pulmonar Severa e Ectasia Gigante da Artéria Pulmonar com Trombose Crónica *In Situ*: Um Relato Longitudinal de Caso

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## Abstract:

Abernethy syndrome is a rare congenital portosystemic vascular anomaly in which portal venous blood partially or completely bypasses the liver. Porto-pulmonary hypertension is one of its most severe complications and may determine long-term prognosis. We report a patient with Abernethy syndrome characterized by complete agenesis of the intrahepatic and common portal vein and a large splenorenal portosystemic shunt, complicated by severe porto-pulmonary hypertension. Given the absence of intrahepatic portal venous branches, shunt closure was not feasible; and liver transplantation is not possible as long as severe pulmonary hypertension persists. Serial multimodality imaging demonstrated progressive giant dilatation of the main pulmonary artery, reaching 7 cm in diameter, associated with chronic in situ thrombosis. Despite advanced pulmonary vascular disease, right ventricular systolic function and functional pulmonary capacity remained preserved under targeted pulmonary hypertension therapy, delaying the need for lung and subsequent liver transplantation. This case underscores the importance of individualized, multidisciplinary management in high-risk patients with Abernethy syndrome, an underrecognized condition that may be associated with severe complications.

**Keywords:** Hypertension, Portal; Hypertension, Pulmonary; Portal Vein/abnormalities; Vascular Malformations.

## Resumo:

A síndrome de Abernethy é uma rara anomalia vascular portossistémica congénita na qual o sangue venoso portal ultrapassa parcial ou completamente o fígado. A hipertensão porto-pulmonar constitui uma das suas complicações mais graves e pode determinar o prognóstico a longo prazo. Relatamos o caso de um doente com síndrome de Abernethy caracterizada por agenesia completa da veia porta intra-hepática e comum, associada a um importante shunt portossistémico esplenorrenal, complicado por hipertensão porto-pulmonar grave. Dada a ausência de ramos portais intra-hepáticos, o encerramento do shunt não foi exequível, assim como a transplantação hepática não foi possível dada a persistência de hipertensão pulmonar grave. A avaliação seriada através de imagiologia multimodal demonstrou dilatação gigante progressiva da artéria pulmonar principal, atingindo 7 cm de diâmetro, associada a trombose crónica in situ. Apesar da doença vascular pulmonar avançada, a função sistólica do ventrículo direito e a capacidade funcional pulmonar mantiveram-se preservadas sob terapêutica dirigida para a hipertensão pulmonar, permitindo adiar a necessidade de transplantação pulmonar e, subsequentemente, também a de fígado. Este caso enfatiza a importância de uma abordagem individualizada e multidisciplinar em doentes de alto risco com síndrome de Abernethy, uma entidade subdiagnosticada e que se pode associar a complicações graves.

**Palavras-chave:** Hipertensão Portal; Hipertensão Pulmonar; Malformações Vasculares; Veia Porta/anomalias congénitas.

### Learning Points

1. Abernethy syndrome can lead to severe porto-pulmonary hypertension, underscoring the importance of considering congenital portosystemic shunts in young patients presenting with otherwise unexplained pre-capillary pulmonary hypertension.
2. Chronic diversion of portal venous blood in Abernethy syndrome promotes progressive vascular remodelling.
3. Giant pulmonary artery ectasia complicated by chronic in situ thrombosis represents a rare but severe manifestation of porto-pulmonary hypertension.
4. Therapeutic decision-making in advanced Abernethy syndrome requires multidisciplinary reassessment.

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https://doi.org/10.60591/crspmi.564

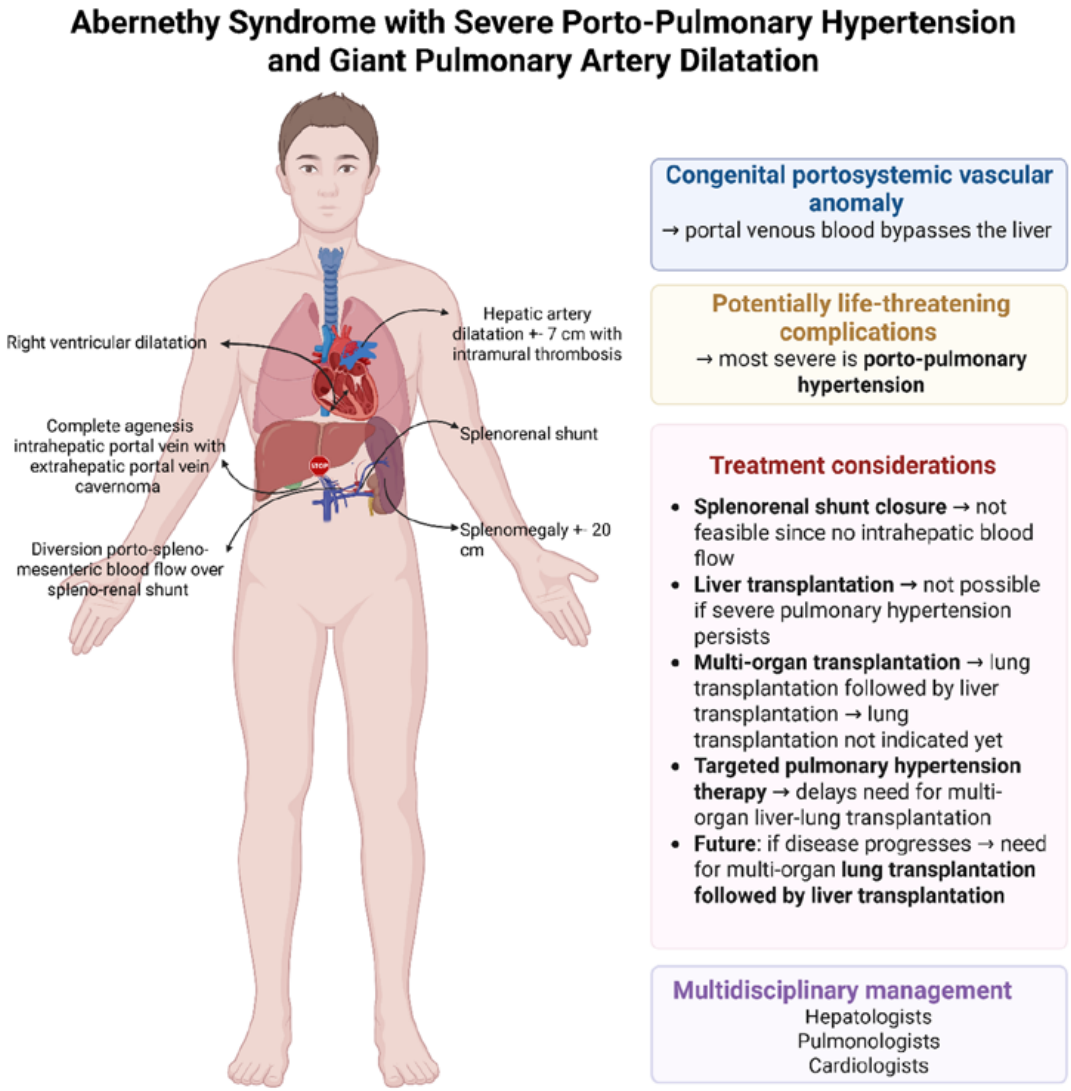



**Figure 1**: Infographic: case summary.

## Introduction

Abernethy syndrome is a rare congenital vascular anomaly in which portal venous blood partially or completely bypasses the liver and drains directly into the systemic circulation.[1,2] It is traditionally classified into type I, characterized by a complete portosystemic shunt with absence of intrahepatic portal veins, and type II, in which a partial shunt is present with hypoplastic but preserved intrahepatic portal branches.[1-3] Given the lack of intra-hepatic blood flow and filtration of portal blood, affected patients may develop a broad spectrum of complications, including hepatic encephalopathy, hepatic nodular lesions (including focal nodular hyperplasia, hepatocellular adenoma, and hepatocellular carcinoma), hepatopulmonary syndrome, and porto-pulmonary hypertension.[1,2,4]

Porto-pulmonary hypertension represents one of the most severe and life-limiting complications of Abernethy syndrome, occurring in approximately 15%, and often determining long-term prognosis as well as eligibility for liver transplantation.[1] While pulmonary artery enlargement is commonly observed in advanced pulmonary hypertension, extreme dilatation reaching aneurysmal dimensions and complicated by chronic in situ thrombosis is exceedingly rare in Abernethy syndrome.[5,6] A structured literature search was performed in PubMed and Embase, until January 2026, using combinations of the following keywords: 'Abernethy syndrome', 'congenital portosystemic shunt', 'porto-pulmonary hypertension', 'pulmonary artery aneurysm', and 'pulmonary artery ectasia'. Based on this search, no previously reported cases describing giant pulmonary artery ectasia with chronic in situ thrombosis in the context of Abernethy syndrome were identified.

We report a complex case of Abernethy syndrome complicated by severe porto-pulmonary hypertension and giant pulmonary artery dilatation with chronic in situ thrombosis, underscoring the diagnostic challenges, therapeutic dilemmas, and the critical importance of long-term multidisciplinary follow-up.

## Case Report

### INITIAL PRESENTATION AND WORK-UP

A 25-year-old patient was first evaluated in 2016 after presenting with exertional dyspnoea. At initial presentation, the patient had no relevant medical comorbidities and reported a healthy lifestyle, with no history of tobacco or alcohol use. Initial cardiological assessment included transthoracic echocardiography, which demonstrated findings suggestive of pulmonary hypertension in the absence of overt right ventricular dysfunction. This prompted further evaluation by right heart catheterization. Haemodynamic assessment revealed pre-capillary pulmonary hypertension with a systolic pulmonary artery pressure of 45 mmHg and a mean pulmonary artery pressure (mPAP) of 28 mmHg. Pulmonary vascular resistance was elevated at 3.8 Wood units. Cardiac output was 4.8 L/min, right atrial pressure was 6 mmHg, and pulmonary capillary wedge pressure was normal at 10 mmHg. Subsequent diagnostic work-up with contrast-enhanced computed tomography (CT) and CT angiography demonstrated congenital absence of the intrahepatic portal vein with complete agenesis of the intrahepatic portal venous branches, marked splenomegaly measuring up to 21 cm, and a large congenital splenorenal shunt measuring 2.5 cm (Fig. 2 panel A). No biochemical or radiological features of liver dysfunction or cirrhosis were identified. These findings were consistent with a diagnosis of Abernethy syndrome with complete intrahepatic portal vein agenesis, resulting in diversion of portal venous flow through the large congenital splenorenal shunt (Fig. 2 panel A). Severe porto-pulmonary hypertension was identified as the predominant clinical complication.

### INITIAL THERAPEUTIC APPROACH

Given the hemodynamically compensated state, targeted medical therapy for pulmonary arterial hypertension was initiated. Treatment consisted of bosentan, a dual endothelin receptor antagonist (125 mg twice daily), combined with ramipril, an angiotensin-converting enzyme inhibition (5 mg once daily). At that time, the patient was discussed for shunt closure; however, this was deemed not feasible due to complete agenesis of the intrahepatic portal vein and absence of intrahepatic portal venous branches. Liver transplantation was considered among the therapeutic options; however, persistently elevated pulmonary artery pressures (45 mmHg), despite medical therapy, constituted a contraindication.[7,8] Given the patient's stable cardiorespiratory status, lung transplantation was not pursued at that stage.

### FOLLOW-UP AND OUTCOME

Between 2016 and 2023, the patient underwent structured multidisciplinary follow-up. The patient remained clinically relatively stable, reporting exertional dyspnoea during strenuous physical activity and mild peripheral malleolar oedema, without significant functional decline. Serial echocardiographic assessments demonstrated a gradual increase in pulmonary artery systolic pressure, progressive right ventricular dilatation, and progressive dilatation of the pulmonary artery, culminating in a sudden worsening of echocardiographic parameters in 2023. Transthoracic echocardiography performed in April 2023 demonstrated a left ventricle of borderline increased dimensions with preserved systolic function (left ventricular ejection fraction 55%-60%). Interventricular septal motion

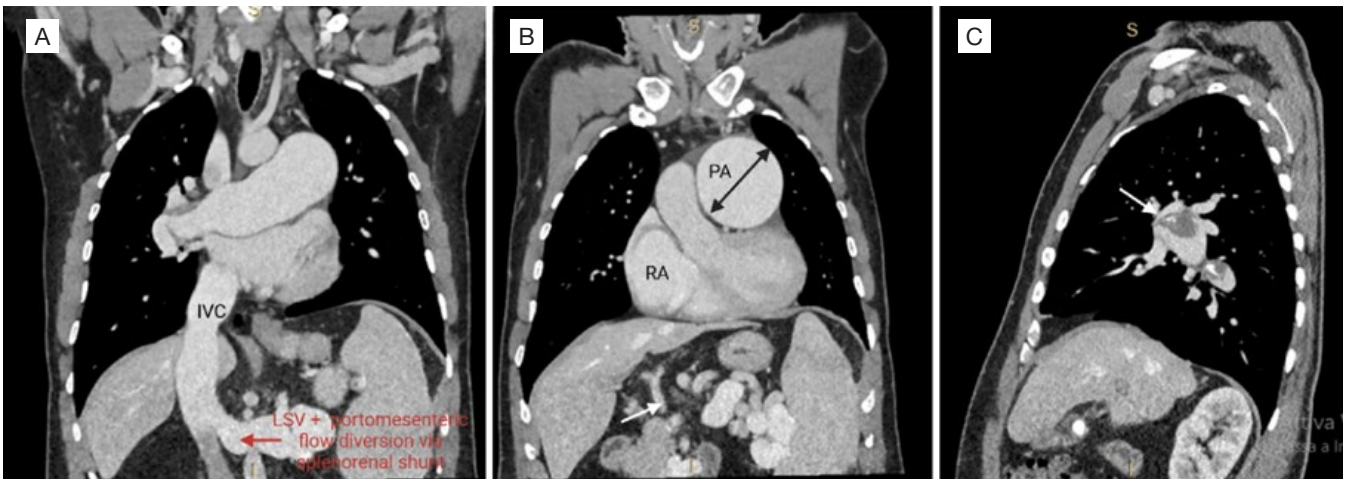



**Figure 2**: Contrast-enhanced CT images of the thorax and abdomen.
**Panel A**: Coronal portal venous phase CT image of the abdomen demonstrating the left renal vein with a splenorenal shunt, resulting in drainage/diversion of the portomesenteric blood flow into the inferior vena cava. **Panel B**: Coronal portal venous phase CT image demonstrating progressive dilatation of the pulmonary arteries. The main pulmonary artery measures approximately 7 cm in diameter (black arrow), while the right and left pulmonary artery branches measure 3-4 cm. Progressive right atrial (RA) dilatation is also observed. The white arrow indicates a hypertrophic hepatic artery associated with complete agenesis of the common and intrahepatic portal veins. **Panel C**: Sagittal contrast-enhanced CT image of the chest showing proximal intraluminal thrombotic defects measuring 2-3 cm with central calcifications, consistent with chronic in situ thrombosis.

was abnormal, consistent with right ventricular pressure overload. The right ventricle was severely dilated, although systolic function remained preserved, with a tricuspid annular plane systolic excursion (TAPSE) of 18 mm. A striking finding was severe dilatation of the pulmonary artery, with the main trunk measuring 60 mm, representing a marked increase compared with previous measurements (maximum 40 mm). Estimated systolic pulmonary artery pressure was approximately 60-62 mmHg. Serial hepatological evaluations demonstrated relatively preserved liver function with mild hyperbilirubinemia and preserved synthetic function, reflected by near-normal serum albumin levels and INR. Repeated abdominal ultrasonography showed a liver of normal size, a patent extrahepatic portal vein, and persistent splenomegaly of 19 cm. During this period, the patient was repeatedly discussed in multidisciplinary meetings due to progressive cardiopulmonary disease despite clinical stability. Given the clinically stable condition, the limited median lung survival following lung transplantation (approximately 6.7 years[9]), and the patient's young age, lung transplantation was not indicated at that stage. Consideration of liver transplantation was deferred, as isolated liver transplantation in the absence of lung transplantation was not considered feasible.

In January 2025, at the age of 34 years, contrast-enhanced CT of the chest demonstrated further massive ectasia of the pulmonary arteries. The main pulmonary artery measured approximately 7 cm in diameter, while the right and left pulmonary artery branches measured 3-4 cm (Fig. 2 panel B). Proximal intraluminal thrombotic defects measuring 2-3 cm, with central calcifications, were identified, consistent with chronic in situ thrombosis (Fig. 2 panel C). No acute segmental or subsegmental pulmonary emboli were detected. The case was subsequently discussed in a multidisciplinary meeting involving cardiologists, thoracic surgeons, pulmonologists, and hepatobiliary specialists. The pulmonary artery thrombotic lesions were interpreted as secondary to vascular dilatation and blood flow stasis rather than embolic disease. Given the diffuse involvement of the pulmonary arterial tree, the absence of clearly defined surgical targets, and the high operative risk, isolated surgical correction of the pulmonary artery aneurysm was not recommended and lung transplantation remained unjustified at that stage, favouring conservative treatment with close follow-up.

At the most recent combined hepatological, cardiological, and pulmonological evaluation in September 2025, the patient remained hemodynamically compensated under medical therapy with mild restrictive lung disease with otherwise preserved pulmonary function. Targeted pulmonary arterial hypertension therapy, to delay the need for lung and subsequent liver transplantation, consisted of sildenafil, a phosphodiesterase-5 inhibitor (20 mg three times daily), and macitentan, a dual endothelin receptor antagonist (10 mg once daily), in combination with low-dose loop diuretics. Close cardiopulmonary and hepatological follow-up was planned, with repeat imaging and continued multidisciplinary reassessment.

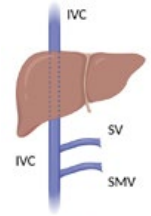
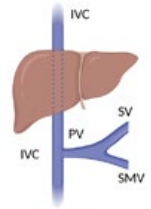
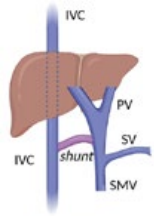
## Discussion

Abernethy syndrome, also known as a congenital extrahepatic portosystemic shunt, exemplifies how congenital abnormalities of the portal venous system can have profound systemic consequences.[2] In the present case, Abernethy syndrome constituted the primary pathogenic driver of portal hypertension with hypersplenism and the development of severe porto-pulmonary hypertension, ultimately culminating in extreme pulmonary arterial remodelling complicated by chronic in situ thrombosis.

The hallmark of Abernethy syndrome is the partial or complete diversion of portal venous blood flow. Congenital portosystemic shunts are classified into two main types according to the extent of portal venous perfusion. Type I shunts (Ia and Ib) are defined by a complete absence of portal blood flow to the liver, with total diversion of portal venous blood into the systemic circulation. In type Ia, the splenic and superior mesenteric veins drain separately, whereas in type Ib they converge before entering the systemic circulation.[2] Type I shunts are rare, with an estimated prevalence of 1:30 000-1:50 000, and are associated with a high burden of complications, including hepatic encephalopathy in approximately 28% of patients, pulmonary hypertension in 15%, hepatopulmonary syndrome in 3%, liver adenomas in 15%, and hepatocellular carcinoma in 12%.[2,4] Type II shunts involve partial diversion of portal blood, allowing residual hepatic perfusion, and are more common and often diagnosed later in life, with a generally milder clinical course (Table 1).[2] In our case, imaging demonstrated complete intrahepatic portal vein agenesis with diversion of portal and mesenteric venous blood flow through a large congenital splenorenal shunt draining into the inferior vena cava. Given the complete absence of intrahepatic portal perfusion, this anatomy is most consistent with a type I portosystemic shunt, albeit with a more specific and atypical configuration that does not conform to the classical Type Ia or Ib subtypes.

The complete diversion of portal venous blood flow into the systemic circulation results in impaired hepatic clearance of vasoactive, pro-inflammatory, and pro-proliferative mediators, including endothelin-1, serotonin, glucagon, and various cytokines.[10] Sustained systemic exposure to these mediators is thought to promote pulmonary vasoconstriction, endothelial dysfunction, and smooth muscle cell proliferation.[10] In addition, chronic high-flow conditions, elevated pulmonary arterial pressures, and abnormal shear stress likely contribute to progressive pulmonary arterial wall remodelling.[2,10] The long-term preservation of right ventricular systolic function observed in this patient suggests effective adaptive remodelling, delaying the onset of right-heart failure and allowing the gradual development of marked pulmonary artery dilatation. Although pulmonary artery enlargement is not uncommon in advanced pulmonary hypertension, extreme dilatation exceeding 6-7 cm is rare and raises concerns regarding rupture, dissection, or

**Table 1**: Anatomical classification, prevalence, and clinical complications of Abernethy malformation.

| Type | Anatomy | Prevalence | Main Complications | Complication Rates |
|---|---|---|---|---|
| **Type I (Ia, Ib)** | Complete absence of portal venous perfusion; portal blood is entirely diverted into the systemic circulation. | Rare; estimated prevalence 1:30 000-1:50 000 | Hepatic encephalopathy, pulmonary hypertension, hepatopulmonary syndrome, liver tumours (adenomas, hepatocellular carcinoma), hyperammonaemia | Hepatic encephalopathy: 28% (10-year: 13%, 20-year: 24%, 30-year: 28%); Pulmonary hypertension: 15%; Hepatopulmonary syndrome: 3%; Liver adenomas: 15%; Hepatocellular carcinoma: 12% |
|  Type 1A | **Ia:** Splenic and superior mesenteric veins drain separately into the systemic circulation. | - | - | - |
|  Type 1B | **Ib:** Splenic and superior mesenteric veins merge before systemic drainage. | - | - | - |
| **Type II**  Type 2 | Partial shunt; a portion of portal blood perfuses the liver while the remainder is diverted into the systemic circulation | More common than Type I; often diagnosed later in life | Hepatic encephalopathy, pulmonary hypertension, hepatopulmonary syndrome, liver tumours, hyperammonaemia (generally less severe than Type I) | Complications similar to Type I but typically milder; pulmonary complications mainly observed in symptomatic patients |

compression of adjacent mediastinal structures.[5,6] After systematic literature research, this case represents one of the first reported cases of Abernethy syndrome complicated by giant pulmonary artery ectasia with chronic in situ thrombosis.

Management of Abernethy syndrome–associated porto-pulmonary hypertension remains particularly challenging and requires an individualized, multidisciplinary approach. Targeted pulmonary arterial hypertension therapy, including endothelin receptor antagonists and phosphodiesterase-5 inhibitors, has been shown to improve haemodynamics and functional status and, in this case, contributed to prolonged clinical stability despite ongoing vascular remodelling.[11,12] Definitive treatment options for Abernethy syndrome include shunt closure or liver transplantation, depending on shunt anatomy and the severity of associated complications.[2,7,13] However, complete agenesis of the intrahepatic portal venous system precluded shunt closure in this patient. Moreover, severe porto-pulmonary hypertension is a well-recognized contraindication to liver transplantation because of unacceptably high perioperative mortality.[8,14-16] In the present case, the coexistence of advanced pulmonary vascular disease and young patient age further complicated transplant decision-making.[9] Lung transplantation was repeatedly considered but deferred due to sustained clinical stability, limited post-transplant lung survival expectancy (approximately 6.7 years), and the potential subsequent need for liver transplantation.[9] With future disease progression, the most likely therapeutic option would be combined multi-organ lung and liver transplantation. Contemporary combined lung–liver transplantation is an exceedingly rare procedure, typically reserved for patients with concomitant end-stage pulmonary and hepatic disease. Although published experience is limited, available case series suggest outcomes comparable to those of single-organ transplantation

when performed in highly experienced centres.[17] This case therefore illustrates the therapeutic impasse that may arise in advanced Abernethy syndrome and underscores the necessity of ongoing longitudinal reassessment within a multidisciplinary framework at specialized centres.

The occurrence of chronic intraluminal pulmonary artery thrombosis adds further complexity to this case. In this setting, thrombosis is most plausibly explained by blood flow stasis within massively dilated pulmonary arteries in combination with endothelial dysfunction, rather than by an underlying systemic procoagulant state. The presence of chronic intraluminal thrombosis posed a significant therapeutic dilemma. Anticoagulation is considered on a case-by-case decision in pulmonary hypertension, particularly in the presence of thrombotic lesions.[18] However, given the coexistence of portal hypertension, severe thrombocytopenia, and extensive portosystemic collateral circulation, anticoagulation was considered high risk and therefore not initiated. Surgical repair of pulmonary artery aneurysms is generally reserved for symptomatic patients, rapidly expanding aneurysms, or those complicated by rupture or compression of adjacent structures. In the present case, the diffuse involvement of the pulmonary arterial tree and the absence of clearly defined surgical targets led the multidisciplinary team to favour conservative management.

This report has several limitations. Firstly, it describes a single case, which limits the generalizability of the observations. Secondly, histopathological confirmation was not available, as no surgical or biopsy specimens were obtained. Thirdly, genetic testing was not performed and therefore potential underlying genetic contributors could not be evaluated. Finally, although the patient has been followed longitudinally, very long-term outcomes remain unknown. Despite these limitations, the detailed and extended clinical follow-up provide valuable insights into the natural history and complex management of severe porto-pulmonary complications in Abernethy syndrome.

In conclusion, this case highlights the severity of pulmonary vascular complications associated with Abernethy syndrome and underscores the critical importance of the portal-pulmonary interaction in determining long-term outcomes. Long-term multidisciplinary follow-up remains essential to navigate the complex therapeutic challenges posed by this rare but severe condition. ■

### Acknowledgement

We would like to thank the patient for his consent and contribution to this case.

### Contributorship Statement

KF and EP - Manuscript writing
AZ – Manuscript review
MS – Attending physician and senior supervisor of the manuscript
All authors approved the final version to be published.

### Declaração de Contribuição

EP, KF – Redação do manuscrito
AZ, UC – Redação e revisão do manuscrito, supervisores séniores do manuscrito
MS – Redação e revisão do manuscrito, médico assistente, supervisor sénior do manuscrito
Todos os autores aprovaram a versão final a ser publicada.

### Ethical Disclosures

Conflicts of Interest: The authors have no conflicts of interest to declare.
Financing Support: This work has not received any contribution, grant or scholarship.
Confidentiality of Data: The authors declare that they have followed the protocols of their work center on the publication of patient data.
Patient Consent: Ethical approval was not required for this case report according to institutional policy. Written informed consent was obtained from the patient for publication of this case report and the accompanying images.
Provenance and Peer Review: Not commissioned; externally peer-reviewed

### Responsabilidades Éticas

Conflitos de Interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.
Fontes de Financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.
Confidencialidade dos Dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.
Consentimento: De acordo com a política institucional, não foi necessária aprovação ética para este relato de caso. Foi obtido o consentimento informado por escrito do paciente para a publicação deste relato de caso e das imagens que o acompanham.
Proveniência e Revisão por Pares: Não comissionado; revisão externa por pares.



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Received / Recebido: 11/02/2026
Accepted / Aceite: 09/03/2026
Published online / Publicado online: 15/04/2026
Published / Publicado: 15/04/2026

**REFERENCES**

1. Venkatesan P. WHO world malaria report 2024. Lancet Microbe. De GotMcLin VA, Franchi-Abella S, Brütsch T, Bahadori A, Casotti V, de Ville de Goyet J, et al. Expert management of congenital portosystemic shunts and their complications. JHEP Rep. 2024;6:100933.

2. Baiges A, Turon F, Simón-Talero M, Tasayco S, Bueno J, Zekrini K, et al. Congenital extrahepatic portosystemic shunts (Abernethy malformation): an international observational study. Hepatology. 2020;71:658–69.
3. Northup PG, Garcia-Pagan JC, Garcia-Tsao G, Intagliata NM, Superina RA, Roberts LN, et al. Vascular liver disorders, portal vein thrombosis, and procedural bleeding in patients with liver disease: 2020 practice guidance by the American Association for the Study of Liver Diseases. Hepatology. 2021;73:366–413.
4. Sanada Y, Mizuta K, Niki T, Tashiro M, Hirata Y, Okada N, et al. Hepatocellular nodules resulting from congenital extrahepatic portosystemic shunts can differentiate into potentially malignant hepatocellular adenomas. J Hepatobiliary Pancreat Sci. 2015;22:746–56.
5. Schwarz S, Benazzo A, Prosch H, Jaksch P, Klepetko W, Hoetzenecker K, et al. Lung transplantation for pulmonary hypertension with giant pulmonary artery aneurysm. J Thorac Cardiovasc Surg. 2020;159:2543–50.
6. Doi A, Gajera J, Niewodowski D, Gangahanumaiah S, Whitford H, Snell G, et al. Surgical management of giant pulmonary artery aneurysms in patients with severe pulmonary arterial hypertension. J Card Surg. 2022;37:1019–25.
7. Uchida H, Sakamoto S, Kasahara M, Kudo H, Okajima H, Nio M, et al. Long-term outcome of liver transplantation for congenital extrahepatic portosystemic shunt. Liver Transpl. 2021;27:236–47.
8. Samuel D, De Martin E, Berg T, Berenguer M, Burra P, Fondevila C, et al. EASL Clinical Practice Guidelines on liver transplantation. J Hepatol. 2024;81:1040–86.
9. Christie JD, Van Raemdonck D, Fisher AJ. Lung transplantation. N Engl J Med. 2024;391:1822–36.
10. Hoeper MM, Krowka MJ, Strassburg CP. Portopulmonary hypertension and hepatopulmonary syndrome. Lancet. 2004;363:1461–8.
11. Angeli P, Bernardi M, Villanueva C, Francoz C, Mookerjee RP, Trebicka J, et al. EASL Clinical Practice Guidelines for the management of patients with decompensated cirrhosis. J Hepatol. 2018;69:406–60.
12. Ruopp NF, Cockrill BA. Diagnosis and treatment of pulmonary arterial hypertension. JAMA. 2022;327:1379.
13. Squires RH, Ng V, Romero R, Ekong U, Hardikar W, Emre S, et al. Evaluation of the pediatric patient for liver transplantation: 2014 practice guideline by the American Association for the Study of Liver Diseases, American Society of Transplantation and the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Hepatology. 2014;60:362–98.
14. Houlihan DD, Holt A, Elliot C, Ferguson JW. Review article: liver transplantation for the pulmonary disorders of portal hypertension. Aliment Pharmacol Ther. 2013;37:183–94.
15. Savale L, Guimas M, Ebstein N, Fertin M, Jevnikar M, Renard S, et al. Portopulmonary hypertension in the current era of pulmonary hypertension management. J Hepatol. 2020;73:130–9.
16. Martin P, DiMartini A, Feng S, Brown R, Fallon M. Evaluation for liver transplantation in adults: 2013 practice guideline by the American Association for the Study of Liver Diseases and the American Society of Transplantation. Hepatology. 2014;59:1144–65.
17. Raevens S, Boret M, De Pauw M, Fallon MB, Van Vlierberghe H. Pulmonary abnormalities in liver disease: relevance to transplantation and outcome. Hepatology. 2021;74:1674–86.
18. Bertoletti L, Escal J, Boucly A, Turquier S, Jevnikar M, Lamblin N, et al. Association between anticoagulant therapy and survival in pulmonary arterial hypertension. J Am Coll Cardiol. 2025;86:982–95.