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Case Report

Re-Emergent Cyanosis Post-Fontan: Unraveling the Differential Diagnosis and Optimizing Targeted Management

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Abstract

Background: The Fontan procedure represents the definitive palliative surgical intervention for patients with single ventricle physiology, establishing a total cavopulmonary connection that redirects systemic venous return directly to the pulmonary arteries. While this procedure aims to normalize systemic arterial oxygen saturation, numerous patients experience persistent or recurrent cyanosis, which significantly impacts long-term morbidity and mortality. The major causes of desaturation include surgically created fenestrations, baffle leaks, pulmonary arteriovenous malformations, and Veno-Venous Collaterals (VVC) draining into the pulmonary veins or left atrium. VVC develop in approximately 20-25% of patients undergoing catheterization following Fontan operations, believed to result from chronically elevated central venous pressure. These collaterals function as right-to-left shunts, leading to systemic desaturation and increasing the risk of paradoxical embolism.

Case Illustration: We present a 34-year-old female with tricuspid atresia, pulmonary atresia, ventricular septal defect, atrial septal defect, and hypoplastic right ventricle who underwent a fenestrated Fontan procedure at age three. She presented with a 9-year history of progressive fatigue, dyspnea, palpitations, and chest pain, worsening over 2 years with persistent cyanosis requiring supplemental oxygen. Physical examination revealed oxygen saturation of 81%, digital clubbing, and a grade 3 out of 6 pansystolic murmur. Cardiac magnetic resonance imaging identified a 12-millimeter fistula between the left innominate vein and left upper pulmonary vein. Catheter-based embolization was performed using a 16/18 millimeter patent ductus arteriosus occluder device via a transjugular approach. Post-procedural angiography confirmed successful fistula closure with immediate improvement in oxygen saturation from 81% to 94%. The patient was discharged in stable condition with sustained clinical improvement at follow-up.

Conclusions: VVCs represent an important etiology of persistent cyanosis in post-Fontan patients. Advanced cardiac imaging, particularly cardiac magnetic resonance, is critical for identifying these abnormal vascular connections. Transcatheter embolization using appropriately sized occluder devices can provide effective treatment with immediate clinical benefit. However, patient selection remains crucial as closure may negatively impact survival in certain subgroups, particularly those with elevated Fontan pressures. Comprehensive hemodynamic assessment and careful consideration of risks versus benefits are essential before intervention.

Keywords: Fontan procedure, Cyanosis, Veno-venous collateral, Transcatheter embolization, Patent ductus arteriosus occluder

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Introduction

The Fontan procedure represents a milestone in the surgical management of patients with single ventricle physiology, establishing a total cavopulmonary connection that separates the pulmonary and systemic circulations.[1] First described by Fontan and colleagues in 1968 for patients with tricuspid atresia, this palliative operation has evolved significantly, with contemporary techniques involving staged surgical palliation culminating in the creation of an extracardiac conduit connecting the inferior vena cava to the pulmonary arteries.[16-17] The global population of individuals living with Fontan circulation continues to expand, with estimates suggesting approximately 66 persons per million in 2020, projected to increase to 79 persons per million by 2030, with the demographic progressively shifting toward adult predominance.[2] Despite improvements in surgical technique and perioperative management, long-term survival and quality of life remain compromised by various complications inherent to the unique Fontan circulation physiology.[3,5,20]

The absence of a subpulmonary ventricle in Fontan circulation results in chronically elevated central venous pressure, non-pulsatile pulmonary blood flow, and at least mild reduction in systemic cardiac output.[3,16] While successful Fontan palliation allows many patients to lead relatively normal lives through early adulthood, the unique hemodynamic characteristics of this circulation predispose patients to numerous complications, including restrictive lung disease, arrhythmias, ventricular dysfunction, protein-losing enteropathy, plastic bronchitis, hepatic dysfunction, and persistent or recurrent cyanosis.[3,11,15,19] Among these complications, cyanosis represents a particularly significant concern, as it serves as a surrogate marker for unfavorable Fontan hemodynamics and has been identified as the strongest independent predictor of late mortality following Fontan operation, with hazard ratios exceeding 12 in multivariable analyses.[6] Understanding the etiology and optimal management of cyanosis in this population is therefore critical to improving long-term outcomes.[1,5]

The major causes of cyanosis following Fontan operation include surgically created fenestrations or baffle leaks, drainage of the coronary sinus into the left atrium, pulmonary arteriovenous malformations, and Veno-Venous Collaterals (VVC) draining into the pulmonary veins or left atrium.[3,6,12] VVC develop in approximately 20 to 25 percent of patients undergoing cardiac catheterization following Fontan or Glenn operations, representing aberrant connections between the systemic venous circulation and the pulmonary venous system or left atrium.[10,14] These collaterals are believed to arise from chronically elevated central venous pressure, either at rest or during physical activity, and may represent persistent embryological venous channels that fail to regress or enlarge due to hemodynamic stress.[10,14] The innominate vein represents the most frequent site for collateral formation, likely due to its anatomical proximity to pulmonary veins and atrial structures.[14] While transcatheter occlusion of these collaterals has been proposed as a therapeutic intervention, recent studies have presented conflicting findings regarding safety and efficacy, with some reports suggesting potential harm from intervention, particularly in patients with elevated Fontan pressures or atriopulmonary-type connections.[8-9]

In this report, we present a rare case of a large veno-venous fistula connecting the left innominate vein to the left upper pulmonary vein in a young adult female patient with prior Fontan fenestration procedure, resulting in significant arterial desaturation and clinical symptoms. We describe the diagnostic approach utilizing advanced cardiac imaging, the catheter-based intervention performed to close the fistula, and the immediate clinical improvement achieved. Additionally, we provide a comprehensive review of the relevant literature regarding the pathophysiology, diagnosis, and management of VVC in the post-Fontan population, highlighting areas of ongoing controversy and the need for individualized treatment decisions based on careful hemodynamic assessment and consideration of patient-specific risk factors.

Case Illustration

A 34-year-old female patient presented to our congenital heart disease clinic with a 9-year history of progressive symptoms including chronic fatigue, exertional dyspnea, palpitations, and intermittent chest pain. Her symptoms had significantly worsened over the preceding 2 years, with dyspnea occurring with minimal daily activities and development of visible cyanosis requiring supplemental oxygen therapy. Her current medical regimen included bosentan 2 mg once daily, sildenafil 30 mg three times daily, and ramipril 2.5 mg once daily. The patient described intermittent chest pain without progressive worsening, and palpitations accompanying dyspneic episodes. She

maintained a supine sleeping position without difficulty and denied presyncope, syncope, fever, cough, or gastrointestinal complaints.

The patient was born as the 3rd child of a twin pregnancy, delivered at 34 weeks gestational age following a complicated pregnancy with three episodes of vaginal spotting. Birth weight was 2 kg with length 49 cm. The neonate cried immediately after delivery but developed cyanosis, requiring incubator care for 10 days. Prenatal care was provided regularly by an obstetrician throughout pregnancy. Both parents were 27 years old at conception. Family history was negative for congenital heart disease or other significant hereditary conditions. At age 3 years, comprehensive cardiac evaluation revealed complex congenital heart disease including tricuspid atresia, pulmonary atresia, large ventricular septal defect, atrial septal defect, hypoplastic right ventricle, and pulmonary hypertension.

Surgical correction was performed via a fenestrated Fontan procedure, involving creation of a total cavopulmonary connection with anastomosis of the superior vena cava and inferior vena cava to the right pulmonary artery, with intentional fenestration to the right atrium. The postoperative course was uncomplicated, and the patient remained clinically stable with regular outpatient cardiology follow-up through early adulthood. However, beginning approximately 9 years prior to current presentation, she noted gradual onset of exercise intolerance and fatigue, which progressively worsened over subsequent years. The symptoms eventually led to her current presentation requiring comprehensive evaluation for persistent cyanosis.

On presentation, the patient appeared in no acute distress but demonstrated visible central cyanosis. Vital signs revealed blood pressure of 120/75 mmHg, heart rate of 69 bpm, respiratory rate of 18 breaths per minute, and oxygen saturation of 81% on room air. Cardiovascular examination revealed a regular rhythm with a grade 3/6 pansystolic murmur best heard along the upper left sternal border. Prominent digital clubbing was present in all extremities. Respiratory examination demonstrated clear breath sounds bilaterally without adventitious sounds. Abdominal examination was unremarkable without hepatosplenomegaly or ascites. There was no peripheral edema, and peripheral pulses were palpable and symmetric.

Laboratory evaluation revealed elevated hemoglobin of 16.4 g/dL and hematocrit of 53.8%, consistent with chronic hypoxemia-induced erythrocytosis. Mild hypokalemia was noted with serum potassium of 3.2 mEq/L. 12-lead electrocardiography demonstrated sinus rhythm with voltage criteria for left ventricular hypertrophy (Figure 1). Chest radiography showed a cardiothoracic ratio of 42% with pulmonary plethora (Figure 2). Transthoracic echocardiography confirmed tricuspid atresia, pulmonary atresia, large atrial septal defect with right-to-left shunting, and severely hypoplastic right ventricle. The Fontan conduit remained patent with laminar flow and no thrombus formation (Figure 3).

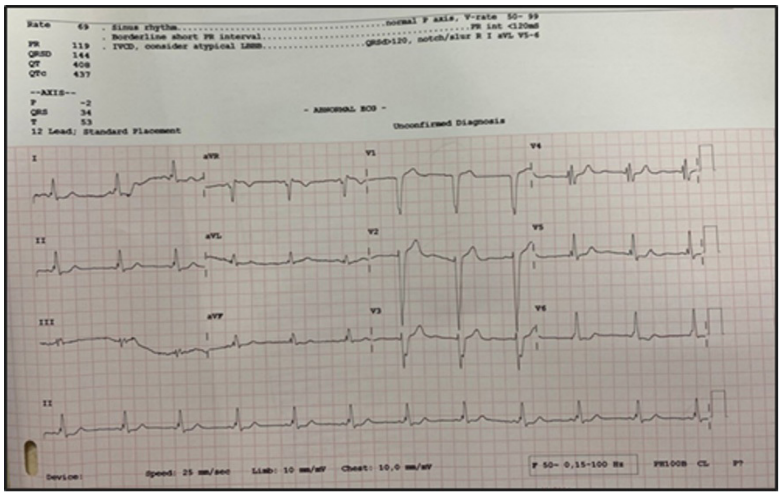


Figure 1. Twelve-lead electrocardiogram demonstrating sinus rhythm with heart rate of sixty-nine beats per minute, normal axis (QRS axis thirty-four degrees, P wave axis negative two degrees), and voltage criteria for left ventricular hypertrophy. QRS duration measures one hundred fourteen milliseconds. No evidence of right ventricular hypertrophy, pathological Q waves, ST segment abnormalities, or T wave inversions.

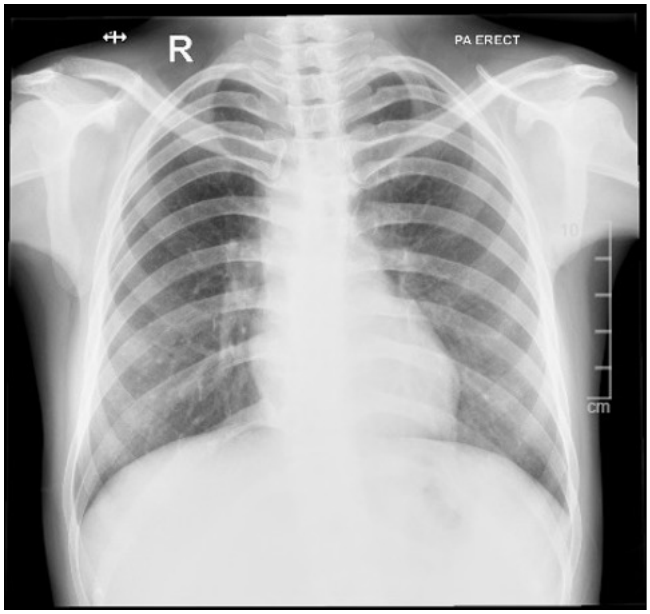


Figure 2. Chest radiograph in posteroanterior projection demonstrating cardiothoracic ratio of forty-two percent with normal aortic and pulmonary artery segment contours. The cardiac silhouette shows flattening of the left heart border with downward displacement of the apex. Pulmonary vascular markings extend beyond two-thirds of bilateral lung fields, consistent with pulmonary plethora. No pleural effusions or parenchymal infiltrates are present.

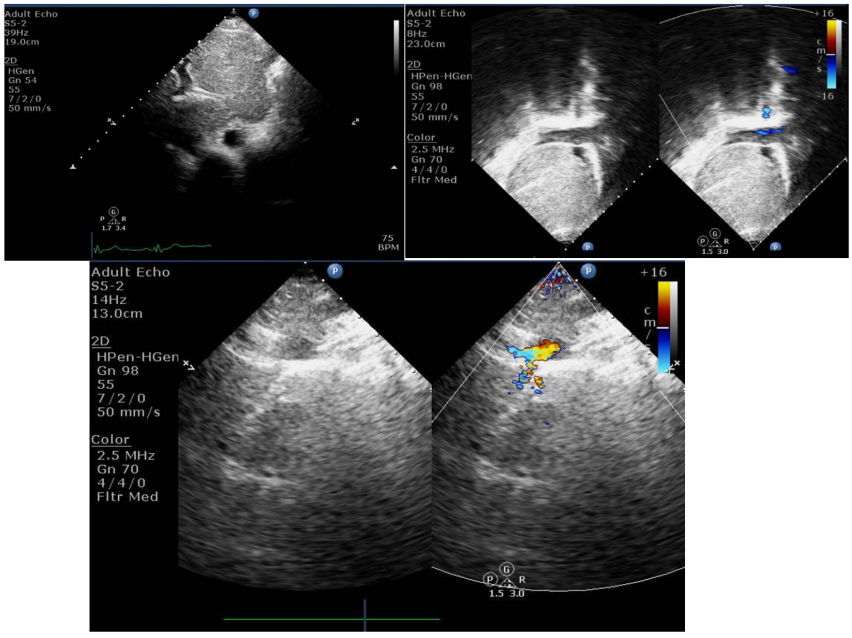


Figure 3. Transthoracic echocardiography demonstrating patent Fontan extracardiac conduit connecting the inferior vena cava to the pulmonary arteries with laminar flow pattern and no evidence of thrombus formation. The anastomosis between the superior vena cava and right pulmonary artery from prior bidirectional cavopulmonary shunt also demonstrates patent connection with minimal pressure gradient.

Cardiac magnetic resonance imaging provided comprehensive anatomical assessment. The study confirmed there is a connection between the innominate vein and the upper left pulmonary vein with obstruction, with suspicion of a veno-venous fistula with obstruction. The diameter of the fistula drainage (at the innominate vein) is 12 mm. 4D flow assessment demonstrated significant right-to-left shunting through this connection. The aortic arch was right-sided without coarctation. Mid-apical left ventricular fibrosis was noted on late gadolinium enhancement imaging. No thrombi were identified within the Fontan circuit (Figure 4).

Based on comprehensive evaluation, the patient was diagnosed with a large veno-venous fistula connecting the left innominate vein to the left upper pulmonary vein, complicated by persistent cyanosis and secondary erythrocytosis. Transcatheter embolization was performed under general anesthesia. Pre-procedural assessment demonstrated favorable pulmonary vascular hemodynamics, with a Pulmonary Vascular Resistance (PVR) of 1.16 WU, indexed PVR (PVRi) of 1.82 WU·m², and a PVR-to-Systemic Vascular Resistance (SVR) ratio of 0.11. Selective innominate vein angiography confirmed the large fistula draining into the left upper pulmonary vein. A sixteen-by-eighteen-millimeter patent ductus arteriosus occluder device was successfully deployed, achieving complete fistula closure (Figure 5). Following device deployment, repeat hemodynamic evaluation showed a PVR of 1.30 WU, PVRi of 0.82 WU·m², and a PVR/SVR ratio of 0.07. No clinically significant increase in Fontan pathway pressure was observed following device closure, supporting the hemodynamic tolerability of the intervention. Importantly, post-intervention oxygen saturation improved dramatically to 94.3%. The patient was discharged on day 3 with sustained clinical improvement. Follow-up echocardiography demonstrated a patent Fontan pathway with preserved ventricular function (Figure 6).

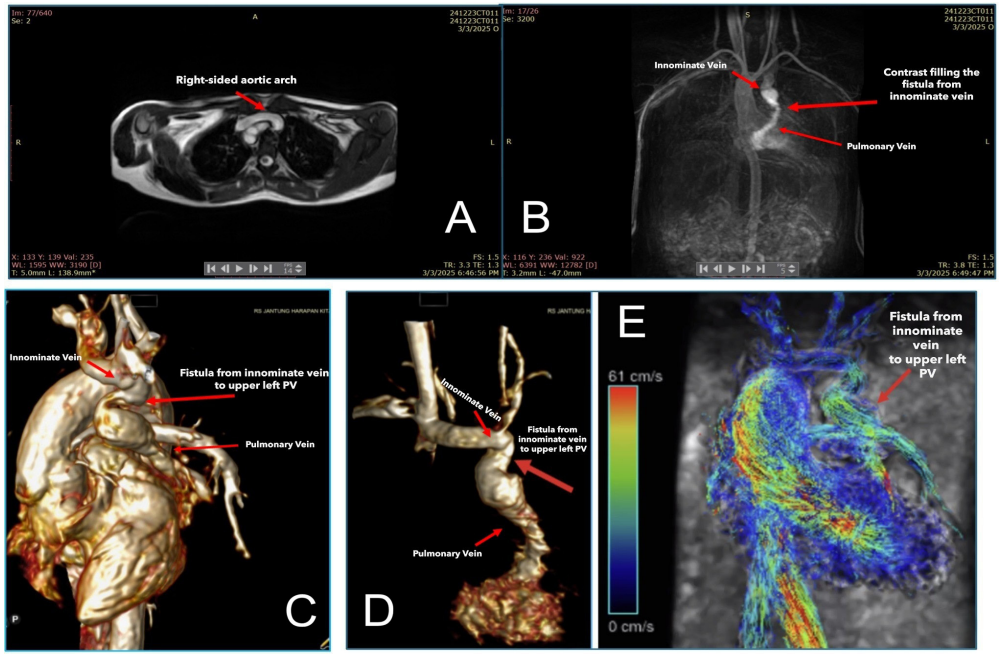


Figure 4. Cardiac magnetic resonance imaging. Panel A shows axial view demonstrating right-sided aortic arch. Panel B demonstrates maximum intensity projection magnetic resonance angiography with contrast medium filling the large veno-venous fistula originating from the innominate vein. Panels C and D show three-dimensional volume rendering technique images revealing the abnormal connection between the left innominate vein and left upper pulmonary vein, with the fistula measuring approximately twelve millimeters in diameter at its origin. Panel E demonstrates four-dimensional flow imaging confirming significant right-to-left shunting through the veno-venous collateral with flow directed from the systemic venous circulation into the pulmonary venous system.

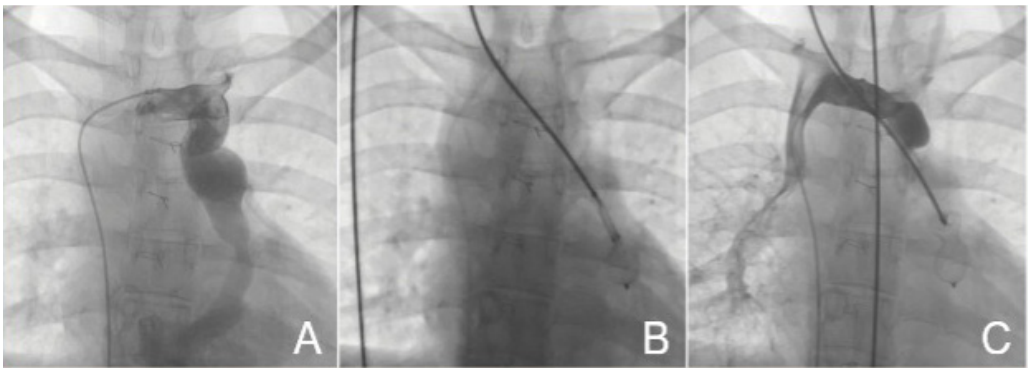


Figure 5. Cardiac catheterization angiography images. Panel A demonstrates initial selective angiography of the left innominate vein in anteroposterior projection, showing rapid contrast filling of the large veno-venous fistula with immediate drainage into the left upper pulmonary vein and subsequently into the left atrium, creating a significant right-to-left shunt. Panel B shows successful deployment and positioning of the sixteen by eighteen millimeter patent ductus arteriosus occluder device (Lifetech) across the fistula orifice at the innominate vein origin. Panel C demonstrates repeat angiography following device deployment, confirming complete cessation of flow through the veno-venous collateral with no residual shunting observed.

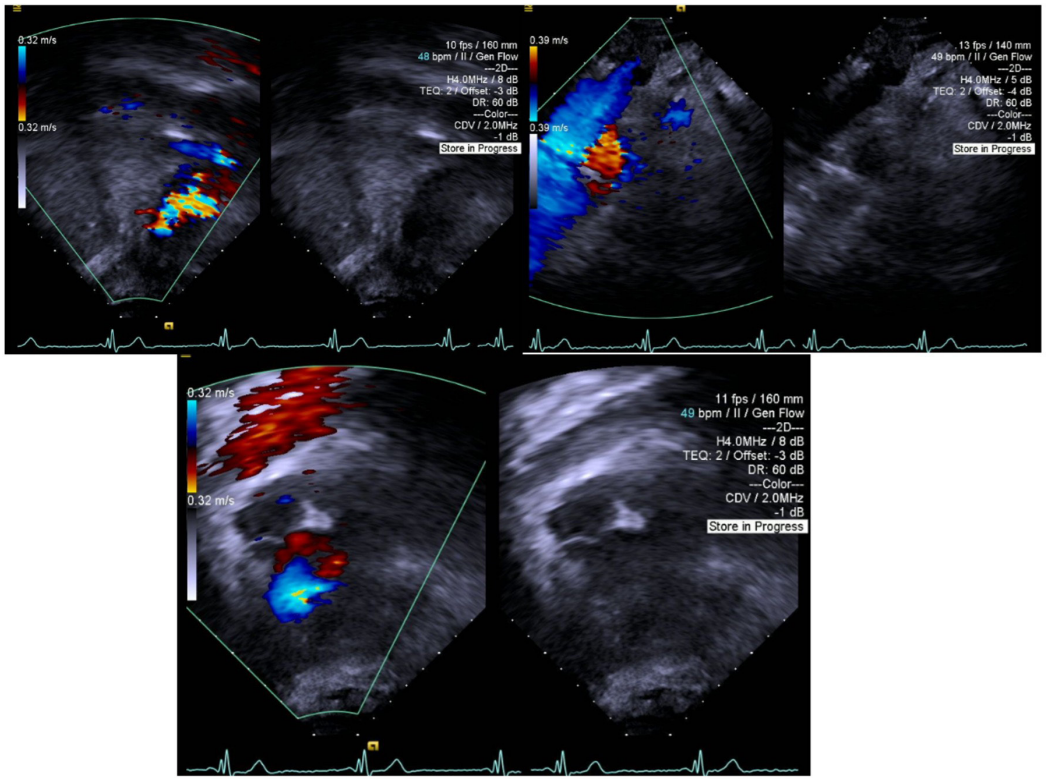


Figure 6. Follow-up transthoracic echocardiography performed approximately one month after transcatheter embolization, demonstrating preserved left ventricular systolic function with good contractility. The Fontan extracardiac conduit remains widely patent with normal laminar flow patterns and no evidence of thrombus formation. The anastomosis between the inferior vena cava and conduit shows unobstructed flow. The anastomosis between the right superior vena cava and right pulmonary artery from the prior bidirectional cavopulmonary shunt also remains patent. No evidence of device migration or embolization is observed.

Discussion

This case illustrates an uncommon but clinically significant complication of Fontan circulation, specifically the development of a large veno-venous collateral connecting the systemic venous circulation to the pulmonary venous system, resulting in persistent cyanosis and exercise intolerance three decades after initial surgical palliation.[14] The successful transcatheter closure using a patent ductus arteriosus occluder device resulted in immediate and sustained clinical improvement, highlighting the importance of accurate diagnosis and timely intervention in appropriately selected patients.[9,18] However, this case also underscores the complex considerations necessary when evaluating patients for potential collateral closure, given emerging evidence suggesting potential harm from such interventions in certain patient subgroups.[8] Our patient, with relatively preserved Fontan hemodynamics and low central venous pressures, represented an ideal candidate for intervention, demonstrating dramatic improvement in oxygen saturation from 81 to 94% post-intervention.

The Fontan procedure has undergone substantial evolution since its initial description in 1968, with contemporary approaches following a staged surgical palliation strategy.[1,16-17] Our patient underwent a fenestrated Fontan procedure, which incorporates an intentional small atrial septal defect serving as a pop-off valve between the systemic venous system and left atrium.[30] The unique Fontan hemodynamics, characterized by the absence of a subpulmonary ventricle and chronically elevated central venous pressure, predispose patients to numerous long-term complications, including progressive PVR elevation, declining cardiac output, and circulatory failure.[3,11,16] The fenestration provides theoretical advantages by maintaining consistent systemic ventricular preload and potentially reducing postoperative pleural effusions and hospital length of stay.[30] However, fenestration may cause mild arterial desaturation through right-to-left shunting, but persistent or recurrent cyanosis after Fontan should prompt evaluation for alternative causes, particularly VVC.[23,30]

Despite successful Fontan operation aiming to normalize systemic arterial oxygen saturation, many patients experience persistent or recurrent cyanosis, with prevalence reaching approximately 50% at hospital discharge and 39% at long-term follow-up.[6] Among patients undergoing catheterization for clinical indications, cyanosis prevalence reaches 72%.[6] The presence of cyanosis carries significant prognostic implications, identified as the strongest independent predictor of late mortality following Fontan operation, with a hazard ratio exceeding 12.[6] VVC, either alone or in combination with patent fenestration, accounts for more than half of cases of persistent cyanosis.[6,10] These collaterals develop through multiple pathophysiological mechanisms, including chronically elevated central venous pressure creating hemodynamic stress that promotes dilation of residual embryological venous channels.[10,14] The innominate vein represents the most frequent anatomical site for collateral formation.[14]

Advanced cardiac imaging plays a critical role in identifying the anatomical substrate underlying cyanosis in post-Fontan patients.[13,29] While transthoracic echocardiography serves as the primary non-invasive surveillance modality, it has significant limitations in adult patients with complex congenital heart disease, including suboptimal acoustic windows and inability to consistently visualize extracardiac vascular structures.[13] Cardiac magnetic resonance imaging provides comprehensive anatomical and functional assessment ideally suited for evaluation of adult Fontan patients, offering excellent spatial resolution for visualization of extracardiac vascular structures and collateral vessels.[13,29] Contrast-enhanced magnetic resonance angiography delineates vascular anatomy with high precision, while 4D flow imaging enables quantification of blood flow through abnormal pathways.[13] In our patient, cardiac magnetic resonance definitively identified the 12 mm veno-venous collateral, with flow assessment demonstrating significant shunting accounting for arterial desaturation.

Cardiac catheterization with selective angiography remains the gold standard for definitive anatomical delineation of VVC, while simultaneously enabling comprehensive hemodynamic assessment and therapeutic intervention.[18] Pre-intervention hemodynamic measurements provide critical information regarding Fontan circuit pressures, transpulmonary gradient, and pulmonary vascular resistance, which inform risk stratification and intervention decision-making.[1,18] Transcatheter occlusion has emerged as the primary therapeutic approach for symptomatic patients with suitable anatomy.[9,18] Various device types have been employed, including coils, vascular plugs, and septal occluders, with device selection depending on collateral size, length, and tortuosity.[18] In our patient, the 16/18 mm patent ductus arteriosus occluder provided secure anchoring and complete

flow occlusion, with immediate improvement in oxygen saturation and resolution of symptoms.

Despite apparent success of collateral embolization in improving oxygen saturation, recent evidence has raised important concerns regarding safety and long-term outcomes.[8] A landmark study by Poterucha and colleagues reported 5-year survival of 74% in patients who underwent embolization compared to 92% in those who did not, with embolization failing to demonstrate significant improvement in hemoglobin levels or exercise capacity.[8] These findings suggest VVC may serve adaptive functions in maintaining cardiac output by decompressing the elevated central venous system, and their closure may paradoxically worsen outcomes in certain subgroups.[8] Patients with atriopulmonary-type Fontan connections, heterotaxy syndrome, or Fontan pressures >18 mmHg demonstrate poorer outcomes following intervention.[8] In contrast, other reports documented clinical benefit from embolization.[9] This discrepancy likely reflects differences in patient selection.

Our patient, with relatively low Fontan pressures (mean 10-11 mmHg pre-intervention), preserved ventricular function with an ejection fraction of 56%, and low PVR of 1.16 WU, represented an ideal candidate for intervention based on current understanding of risk factors.[4,8-9] Her dramatic clinical improvement and sustained benefit at follow-up 3 days later in the outpatient department support the appropriateness of intervention in carefully selected patients with favorable hemodynamics and symptomatic hypoxemia primarily attributable to collateral shunting rather than underlying circulatory failure.[9,22] Long-term outcomes following collateral embolization require ongoing surveillance for potential complications, including collateral recurrence, device-related complications, and progression of underlying Fontan circulatory failure.[11,20] Regular follow-up with clinical monitoring, functional assessment, and periodic imaging remains essential to detect these complications and ensure sustained benefit from intervention.[1,13]

This case underscores several important clinical principles in managing adult patients with Fontan circulation presenting with cyanosis.1 First, systematic diagnostic evaluation utilizing advanced imaging modalities, particularly cardiac magnetic resonance, enables accurate identification of the anatomical substrate underlying desaturation.[13,29] Second, comprehensive hemodynamic assessment via cardiac catheterization provides critical information facilitating risk stratification and intervention planning.[18] Third, patient selection for transcatheter collateral closure should incorporate multiple factors including symptom severity, degree of desaturation, Fontan pressures, ventricular function, and presence of other complications indicating advanced Fontan failure.[1,8] Fourth, patients with favorable hemodynamics and symptomatic hypoxemia primarily attributable to collateral shunting represent the most appropriate candidates for intervention.[9] Fifth, long-term follow-up remains essential to detect potential recurrence, device complications, and progression of underlying Fontan failure.[11,20]

Conclusion

We present the successful management of a rare but clinically significant complication of Fontan circulation, specifically a large veno-venous fistula connecting the innominate vein to the pulmonary venous system resulting in persistent cyanosis three decades after initial surgical palliation. Advanced cardiac imaging, particularly cardiac magnetic resonance imaging with four-dimensional flow assessment, enabled definitive anatomical diagnosis and intervention planning. Transcatheter embolization using a patent ductus arteriosus occluder device achieved immediate and sustained clinical improvement with a dramatic increase in oxygen saturation and resolution of symptoms. However, patient selection for such interventions requires careful consideration of multiple factors including hemodynamic assessment, symptoms, and overall Fontan circulatory status, given emerging evidence suggesting potential harm from collateral closure in certain patient subgroups. This case highlights the importance of comprehensive evaluation, individualized decision-making, and ongoing surveillance in the management of adult patients with complex congenital heart disease and Fontan circulation.

List of Abbreviations

PVR	Pulmonary Vascular Resistance
PVRi	Indexed Pulmonary Vascular Resistance
SVR	Systemic Vascular Resistance
VVC	Veno-Venous Collaterals

Ethical Clearance

Not applicable.

Publication Approval

The authors consent to the publication of this manuscript.

Authors' Contributions

OL, AAS, TAP: conception and design, or analysis and interpretation of data. OL, AAS, TAP: drafting the article or revising it critically for important intellectual content. OL: final approval of the version to be published.

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Conflict of Interest

None.

Availability of Data and Materials

Not applicable.

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Generative AI and AI-Assisted Technologies in the Writing Process

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References

1. Rychik J, Atz AM, Celermajer DS, Deal BJ, Gatzoulis MA, Gewillig MH, et al. Evaluation and management of the child and adult with fontan circulation: a scientific statement from the American Heart Association. Circulation. 2019;140:e234-84.
2. Plappert L, Edwards S, Senatore A, De Martini A. The epidemiology of persons living with Fontan in 2020 and projections for 2030: development of an epidemiology model providing multinational estimates. Adv Ther. 2022;39:1004-15.
3. Mazza G, Gribaudo E, Agnoletti G. The pathophysiology and complications of Fontan circulation. Acta Biomed. 2021;92:e2021260.
4. Goldberg DJ, Avitabile CM, McBride MG, Paridon SM. Exercise capacity in the Fontan circulation. Cardiol Young. 2013;23:824-30.
5. Khairy P, Fernandes SM, Mayer JE Jr, Triedman JK, Walsh EP, Lock JE, et al. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. Circulation. 2008;117:85-92.
6. Schafstedde M, Nordmeyer S, Schleiger A, Nordmeyer J, Berger F, Kramer P, et al. Persisting and reoccurring cyanosis after Fontan operation is associated with increased late mortality. Eur J Cardiothorac Surg. 2022;61:54-61.
7. Duncan BW, Desai S. Pulmonary arteriovenous malformations after cavopulmonary anastomosis. Ann Thorac Surg. 2003;76:1759-66.
8. Poterucha JT, Johnson JN, Taggart NW, Cabalka AK, Hagler DJ, Driscoll DJ, et al. Embolization of veno-venous collaterals after the Fontan operation is associated with decreased survival. Congenit Heart Dis. 2015;10:E230-6.
9. Tsoutsinos A, Kakava F, Kanakis M, Samanidis G, Despotopoulos S, Vagenakis G, et al. Desaturation after Fontan procedure due to systemic venous collateral and improvement after closure with embolization. Clin Case Rep. 2024;12:e9374.
10. Heinemann M, Breuer J, Steger V, Steil E, Sieverding L, Ziemer G. Incidence and impact of systemic venous collateral development after Glenn and Fontan procedures. Thorac Cardiovasc Surg. 2001;49:172-8.
11. Deal BJ, Jacobs ML. Management of the failing Fontan circulation. Heart. 2012;98:1098-104.
12. Ohuchi H, Mori A, Nakai M, Fujimoto K, Iwasa T, Sakaguchi H, et al. Pulmonary arteriovenous fistulae after Fontan operation: incidence,

clinical characteristics, and impact on all-cause mortality. Front Pediatr. 2022;10:914433.
13. Hong SH, Kim YM, Lee CH, Park SJ, Kim SH. CT and MRI evaluation of the Fontan pathway: pearls and pitfalls. Cardiovasc Imaging Asia. 2017;1:133-43.
14. Schaffner D, Perez MH, Duran R, Pretre R, Di Bernardo S. Transcatheter interventional procedure to innominate vein turn-down procedure for failing Fontan circulation: a case report. Front Pediatr. 2024;12:1328758.
15. Abdulkarim A, Shaji S, Elfituri M, Gunsaulus M, Zafar MA, Zaidi AN, et al. Pulmonary complications in patients with Fontan circulation. J Am Coll Cardiol. 2023;81:2434-44.
16. Gewillig M, Brown SC. The Fontan circulation after 45 years: update in physiology. Heart. 2016;102:1081-6.
17. Trusty PM, Slesnick TC, Wei ZA, Rossignac J, Kanter KR, Fogel MA, et al. Fontan surgical planning: previous accomplishments, current challenges, and future directions. J Cardiovasc Transl Res. 2018;11:133-44.
18. Goldstein BH, Holzer RJ. Interventional catheterization in adult congenital heart disease. Expert Rev Cardiovasc Ther. 2015;13:691-706.
19. Kiesewetter CH, Sheron N, Vettukattil JJ, Hacking N, Stedman B, Millward-Sadler H, et al. Hepatic changes in the failing Fontan circulation. Heart. 2007;93:579-584.
20. Downing TE, Allen KY, Glatz AC, Rogers LS, Ravishankar C, Rychik J, et al. Long-term survival after the Fontan operation: twenty years of experience at a single center. J Thorac Cardiovasc Surg. 2017;154:243-53.
21. Kim SJ, Kim WH, Lim HG, Lee JY. Outcome of 200 patients after an extracardiac Fontan procedure. J Thorac Cardiovasc Surg. 2008;136:108-16.
22. Pike NA, Evangelista LS, Doering LV, Eastwood JA, Lewis AB, Child JS. Quality of life, health status, and depression: comparison between adolescents and adults after the Fontan procedure with healthy counterparts. J Cardiovasc Nurs. 2012;27:539-46.
23. Varma C, Warr MR, Hendler AL, Paul NS, Webb GD, Therrien J. Prevalence of silent pulmonary emboli in adults after the Fontan operation. J Am Coll Cardiol. 2003;41:2252-8.
24. McElhinney DB, Kreutzer J, Lang P, Mayer JE Jr, del Nido PJ, Lock JE. Incorporation of the hepatic veins into the cavopulmonary circulation in patients with heterotaxy and pulmonary arteriovenous malformations after a Kawashima procedure. Ann Thorac Surg. 2005;80:1597-603.
25. Meadows J, Gauvreau K, Jenkins K. Hepatic function and exercise performance in patients with fontan circulation. Pediatr Cardiol. 2017;38:1443-50.
26. Balling G, Vogt M, Kaemmerer H, Eicken A, Meisner H, Hess J. Intracardiac thrombus formation after the Fontan operation. J Thorac Cardiovasc Surg. 2000;119:745-52.
27. Egbe AC, Connolly HM, Khan AR, Niaz T, Said SM, Schaff HV, et al. Outcomes in adult Fontan patients with atrial tachyarrhythmias. Am Heart J. 2017;186:12-20.
28. Cordina R, O'Meagher S, Gould H, Rae C, Kemp G, Pasco JA, et al. Skeletal muscle abnormalities and exercise capacity in adults with a Fontan circulation. Heart. 2013;99:1530-4.
29. Averin K, Hirsch JC, Seckeler MD, Whiteside W, Beekman RH 3rd, Goldstein BH. Magnetic resonance imaging and computed tomography in the evaluation of patients with single ventricle anatomy before bidirectional superior cavopulmonary connection. Congenit Heart Dis. 2014;9:314-25.
30. Salazar JD, Zafar F, Siddiqui K, Coleman RD, Morales DL. Fenestration closure in patients with fontan circulation: indications and outcomes. Congenit Heart Dis. 2010;5:225-30.