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CASE REPORT
Adult congenital heart disease

Percutaneous rheolytic thrombectomy and cerebral embolic protection in a massive thrombosis of a fenestrated Fontan conduit: a case report

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Background Clinical thromboembolism in Fontan patients is often a catastrophic event resulting in death and adverse long-term outcomes. The treatment of acute thromboembolic complications in these patients is very controversial.

Case summary We describe the use of rheolytic thrombectomy in a Fontan patient with life-threatening pulmonary embolism, employing a cerebral protection system to reduce the risk of stroke through the fenestration.

Discussion Rheolytic thrombectomy may be a successful alternative to systemic thrombolytic therapy and open surgical resection for the treatment of acute high-risk pulmonary embolism in the Fontan population. Embolic protection device to capture and remove thrombus/debris may be an innovative tool to reduce the risk of stroke through the fenestration while performing a percutaneous procedure in fenestrated Fontan patient.

Keywords Pulmonary embolism • Fontan circulation • Percutaneous thrombectomy • Cerebral protection system • Case report

ESC Curriculum 7.1 Haemodynamic instability • 9.7 Adult congenital heart disease • 9.5 Pulmonary thromboembolism

Learning points

- Thromboembolic complications following the Fontan palliation are common and are associated with high mortality and morbidity.
- Rheolytic thrombectomy for the treatment of acute high-risk pulmonary embolism is a safe and successful alternative to systemic thrombolytic therapy and open surgical resection in Fontan population.
- A cerebral embolic protection device is an innovative tool that may reduce the risk of stroke through the fenestration by capturing and removing thrombus/debris while performing percutaneous procedures in fenestrated Fontan patients.

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Introduction

The Fontan procedure is the final stage of surgical palliation of single ventricle physiology. The goal of this procedure is to separate systemic and pulmonary blood flow by directing systemic venous return through Fontan connection to the pulmonary arteries, excluding the pulsatile subpulmonary ventricle contribution.

The haemodynamic impact of Fontan circulation results in long-term multi-organ deleterious effects potentiating the pathogenesis of thrombosis.[1] Thrombotic diseases post-Fontan status range between 1 and 33% and represent the principal cause of mortality and morbidity in these patients.[2]

We describe a case of acute bilateral massive pulmonary embolism (PE) due to thrombosis of Fontan conduit successfully treated with percutaneous rheolytic thrombectomy aspiration supported by cerebral embolic protection to prevent the risk of stroke through the fenestration.

Timeline

At birth (7 days)	Blalock–Thomas–Taussig shunt
3 years	Bidirectional cavopulmonary shunt
9 years	Fontan completion with extra cardiac fenestrated conduit (18 mm)
22 years	Life-threatening PE treated with percutaneous rheolytic thrombectomy after placement of a cerebral protection system
23 years	Stable condition at 1 year of outpatient follow-up. Good-adherence to warfarin therapy. No recurrence of PE.

Case presentation

We reported the case of a 22-year-old man with undifferentiated single ventricle, common atrioventricular valve, D-malposition, and pulmonary stenosis in right isomerism surgically palliated with extra-cardiac fenestrated Fontan technique at the age of 9 years. He refused anticoagulant prophylaxis with Warfarin and he consumed aspirin inconstantly. Patient was admitted to our emergency department for acute respiratory failure. At presentation, blood pressure and oxygen saturation were undetectable. Electrocardiogram revealed a sinus tachycardia at 110 beats/min with tall P waves and complete right bundle branch block. Glasgow score was 15. Lung auscultation was remarkable for decreased breath sounds and cardiac examination showed a Grade III/VI holosystolic murmur along the left sternal border. Patient showed hepatomegaly and lower bilateral oedema without evidence of venous phlebitis. Arterial pulses were diminished. Arterial blood gas analysis demonstrated pH 7.24 (interquartile range: 7.35–7.45), pCO_2 51 mmHg (n.v.: 35–48 mmHg), pO_2 68 mmHg (n.v.: 83–108 mmHg), HCO_3 20 mmol/L (n.v.: 21–28 mmol/L), lactate 4.1 mmol/L (n.v.: <1.3 mmol/L), and P/F 68. Patient underwent endotracheal intubation and vasopressor support with noradrenaline. Laboratory values revealed both elevated D-dimer (6432 ng/mL: n.v. until 500 ng/mL) and high-sensitivity Troponin I (0.841 ng/mL; n.v. until 0.055 ng/mL). Bedside echocardiography documented a dilated and dysfunctional single ventricle (ejection fraction 35–40%) with a moderate common atrioventricular valve regurgitation; transpulmonary pressure gradient was 10 mmHg. Inferior vena cava appeared dilated without respiratory modulation pattern at Doppler interrogation. No flow was detected in pulmonary arteries.

Helical total body computed tomography scan demonstrated a large expansive filling defect extending from the right common iliac vein to cavopulmonary anastomosis and almost complete obstruction of both pulmonary arteries (*Figure 1*). In addition, multiple aorto-pulmonary collateral vessels and early focal ischaemic lesions of the left renal cortex were detected. Cerebrovascular lesions were ruled out.

Due to high bleeding risk associated to systemic thrombolysis as well as to surgical embolectomy, the multidisciplinary heart team referred the patient to the cardiac catheterization laboratory for rheolytic thrombectomy as alternative management option.

A cerebral embolic protection device (Sentinel™, Boston Scientific, Santa Rosa, CA, USA) was placed through a 6 Fr sheath from right radial artery over a 0.014-inch guidewire. The two filters of the system were delivered respectively in the brachiocephalic and the left common carotid arteries (*Figure 2*).

After cannulation of right femoral vein with an 8 Fr sheat, a pigtail was advanced in the pulmonary trunk on an angiographic 0.035-inch guidewire. The pulmonary trunk pressure was 15 mmHg. Injection of contrast medium confirmed a huge occlusive thrombus in the Fontan conduit. Hence, through a JR 4.0 guiding catheter, a 0.035-inch guidewire was placed distally in the pulmonary artery, and activated thrombectomy catheter (AngioJet PE™ System) was advanced though the thrombus at ~1 cm every 3 s. Multiple passages of the catheter were performed, until vessels were patent and thrombus burden considerably reduced (*Figure 3* and Supplementary material online, *Videos S1–S3*).

Successful catheter embolectomy rapidly restored normal haemodynamic parameters and gas exchanges. Patient was subsequently transferred to the ICU. Infusion of unfractionated heparin was begun followed tby oral administration of Warfarin once the INR achieved therapeutic range. Inotropic support was weaned at 72 h after recovery of systolic function of single ventricle. Invasive ventilation was stopped at 5 days and the patient was treated with low flow oxygen therapy until the stabilization of oxygen saturation above 90% in room air. Acute worsening of renal function due to paradoxical systemic embolization across the Fontan fenestration prolonged the hospitalization. He was discharged on 18 days in a stable condition persisting after 1 years of outpatient follow-up. The patient adhered to warfarin therapy that is titrated to a target INR range between 2.0 and 2.5. Low-dose diuretic therapy was stopped after 6 months and no additional pharmacological treatments were prescribed except for anticoagulation drugs.

Discussion

Pulmonary embolism is a life-threatening condition for Fontan population. The increase of pulmonary vascular resistance reduces acutely cardiac preload and compromises the cardiac output of single ventricle.

The management of high-risk PE in Fontan patient is very challenging, requiring a multidisciplinary evaluation to choose the optimal treatment.

The lack of a consensus document impacts heavily on the difficulty to choose the strategy of treatment. It should be evaluated case by case, based on localization of thrombi, Fontan function, and comorbidities. Although systemic thrombolysis has been attempted for the treatment of thromboembolic complications of Fontan, administration of thrombolytic drugs remains controversial due to high bleeding risk. The use of fibrinolytic agents is contraindicated in Fontan patients, who are commonly affected by cirrhosis, thrombocytopenia, and anaemia. Surgical embolectomy represents a valid alternative, but it is a high-stake manoeuvres carrying the added risk of anaesthesia.[3]

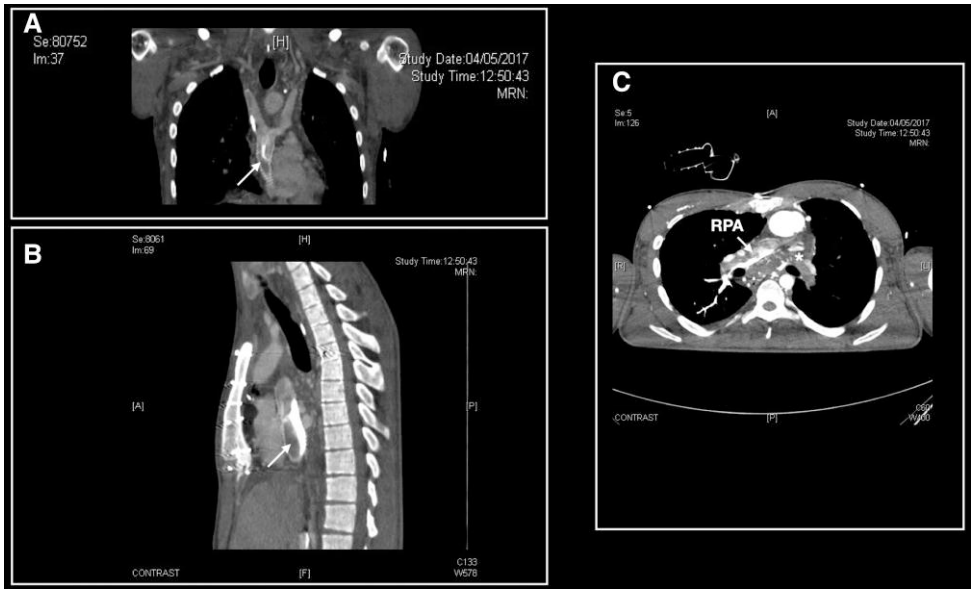

Figure 1 Axial delayed-phase computed tomography pulmonary angiogram shows the filling defect of extra-cardiac Fontan conduit (arrows) indicating the presence of thrombosis (*A* and *B*). Unopacified left pulmonary artery (*) and irregular endoluminal contour of the right pulmonary artery (arrow) are suggestive of pulmonary emboli (*C*).

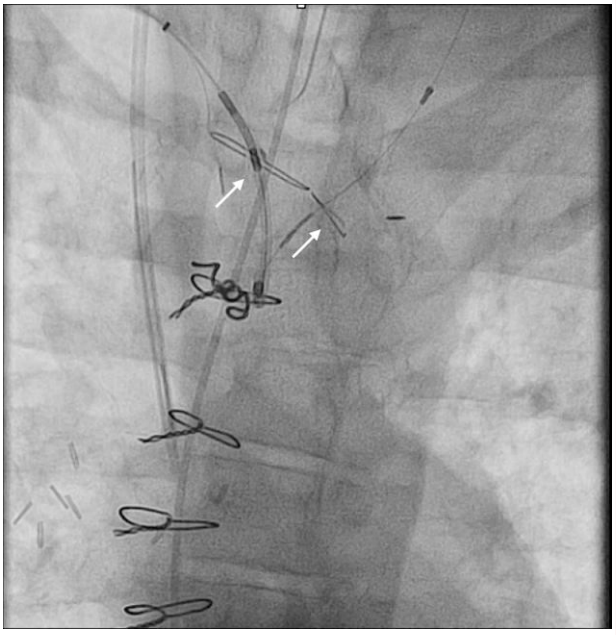

Figure 2 Prophylactic cerebral protection using Sentinel™ (Boston Scientific, Santa Rosa, CA, USA) device to prevent embolic stroke. The filters of device (arrows) are deployed into the brachiocephalic artery (Proximal Filter), and into the left common carotid artery (Distal Filter) according to the medical guidance.

Recent advances in endovascular techniques suggest that percutaneous recanalization is feasible and effective in patient with contraindications to thrombolysis and surgery.[4] Endovascular options may include catheter-directed thrombolysis, mechanical thrombectomy, suction thrombectomy, or a combination of these techniques.

The role of percutaneous thrombectomy for the treatment of massive PE in single ventricle palliation status is not yet elucidated. Anecdotal reports documented, similarly to our case, the efficacy of rheolytic thrombectomy using hydrodynamic fragmentation and aspiration of clot by AngioJet system.[5,6] The low profile of catheter provides excellent trackability and crossability, also in cavopulmonary circuit.

Unlike previous reports, we employed a cerebral protection system to minimize ideally both the risk of silent cerebral ischaemic injury and the severity of clinically evident stroke related to manipulation manoeuvres. In fact, patients with fenestrated Fontan have a major risk of ischaemic events during peripheral rheolytic thrombectomy in respect to the general population, which is represented only by anecdotical cases of paradoxical embolism. These events are due to the presence of the fenestration between cavo-pulmonary conduit and the single ventricle, which allows a direct communication between venous and arterial circulation. This may lead to the migration of the thrombus located in the conduit or even of the debris generated by the thrombectomy system and provocate a cerebral ischaemic injury.To our knowledge, this is the first report documenting the use of cerebral embolic protection devices for endovascular procedure in Fontan patients. Alongside the pathophysiological rationale supporting our procedure, we recognize the limits of this technique. In fact, the cerebral embolic protection device does not provide complete cerebral protection because of the left vertebral artery remains uncovered. Complex anatomy of the aortic arc and vascular tortuosity of the supra-aortic vessels may be make difficult the placement of device and thus predispose to failure. Finally, arterial manipulation may promote injury to the vessel, resulting in dissections, arterial thrombosis, and embolism.

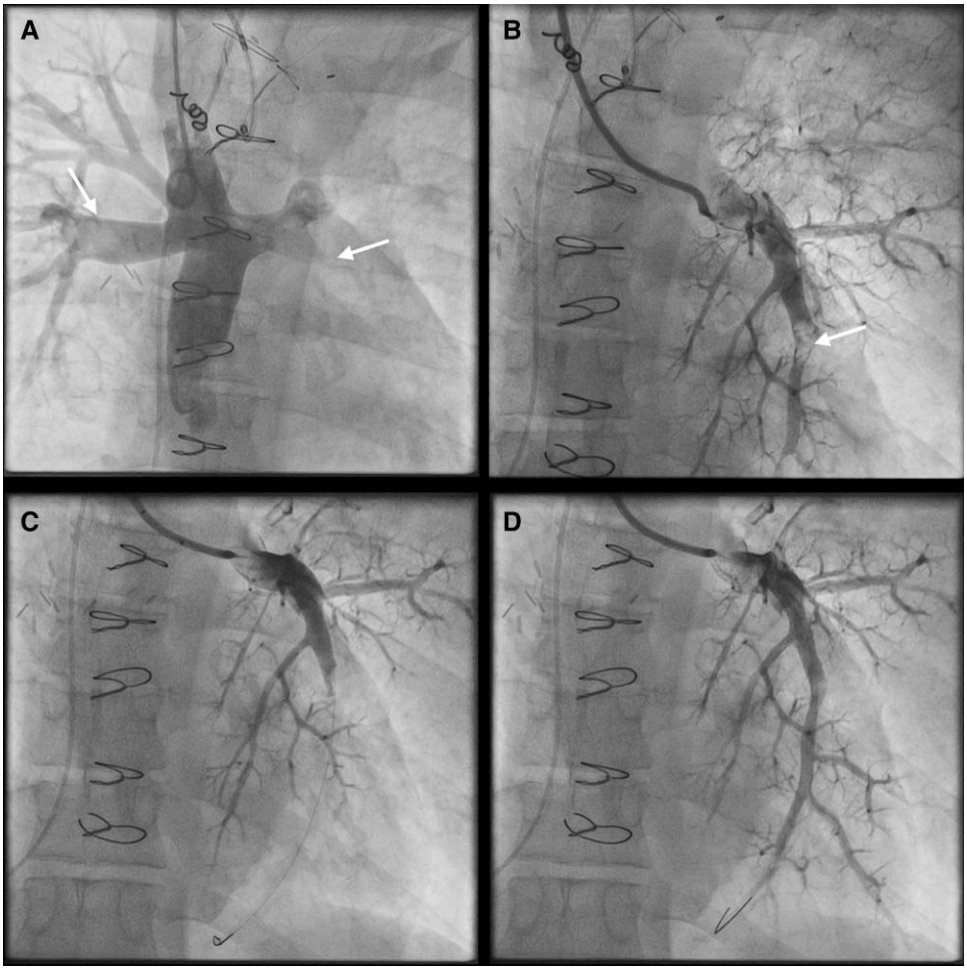

Figure 3 Pulmonary angiography shows massive bilateral embolism (arrows) resulting in the reduction of flow to both right and left lobes (*A*, *B*). Initial angiography of left pulmonary artery (*B* and *C*) and final result of the trombectomy after multiple passages of the catheter of the AngioJet system directed at inferior branch of the left pulmonary artery (*D*).

These observations make the search for preventive strategies for young patients with fenestrated Fontan circulation experiencing severe thromboembolic complications related to complex congenital heart diseases.

Conclusion

Rheolytic thrombectomy for the treatment of acute high-risk PE is a safe and successful alternative to systemic thrombolytic therapy and open surgical resection in Fontan patients. By minimal invasive approach, it permits the early restoration of pulmonary blood flow assuring the prompt reduction of pulmonary artery pressure and resistances and the recovery of single ventricle function. Further evaluation in a larger cohort of patients is warranted to assess whether this treatment may offer an alternative or complement to thrombolysis or surgical thrombectomy.

The use of cerebral embolic protection during endovascular procedures in fenestrated Fontan circulation may be an attractive tool to mitigate the risk of ischaemic cerebral injury in specific high-risk percutaneous scenarios. Further studies will be needed to assess the additional value of the embolic protection device in comparison to rheolytic thrombectomy without the device.

Lead author biography

Raffaella Marzullo, MD, works as a consultant paediatric cardiologist at Monaldi's Hospital of Naples in Italy. Her interests are focused on interventional cardiology, gender medicine, and pregnancy-related diseases in patients with congenital heart defects.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The patient provided informed consent for publication in accordance with COPE guidelines.

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Funding: None declared.

Data availability

The data underlying this article are available in the article and in its online supplementary material.

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