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CASE REPORT
Adult congenital heart disease

Transcatheter thrombectomy of acute pulmonary embolism in an adult Fontan patient: a case report

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Background The Fontan operation is a well-described palliative procedure for functional single-ventricle patients. This population has an increased risk of thrombo-embolic events. Adequate imaging, and therefore diagnosis, requires an understanding of the unique anatomy and physiology of a Fontan. Optimal strategies for the prevention and treatment of thrombo-embolic complications in the Fontan population are poorly defined.

Case summary A 28-year-old female with a history of Ebstein anomaly of the tricuspid valve status post-Fontan presented with chest pain and acute hypoxia. Computed tomographic angiography (CTA) reported a submassive pulmonary embolism (PE). She was initiated on a heparin drip. Catheterization demonstrated elevated Fontan pressures and a large thrombus in the right lower pulmonary artery that was removed with an aspiration device. The patient was transitioned to a direct oral anticoagulant (DOAC) following the procedure.

Discussion Thrombotic complications are common in the adult Fontan population. Given the morbidity and mortality associated with this complication, the use of proper imaging techniques is imperative. Traditional CTA imaging for PE in Fontan patients often has contrast filling defects related to their anatomy and physiology. Utilization of adequate imaging techniques helps decrease cost and additional radiation exposure. In addition, it avoids inappropriate hospitalization, need for anticoagulation, and potential need for catheterization toconfirm or exclude the presence of PE. For Fontan patients with a thrombus, prior event, and no contraindications, current guidelines recommend oral anticoagulation with a vitamin K antagonist; however, there are increasing data on the use of DOACs in this population.

Keywords Ebstein anomaly • Fontan operation • Thrombus • Right-sided catheterization • Anticoagulation • Case report

ESC curriculum 2.1 Imaging modalities • 2.4 Cardiac computed tomography • 8.6 Secondary prevention • 9.7 Adult congenital heart disease • 9.5 Pulmonary thrombo-embolism

Learning points

- Imaging of Fontan patients can be technically challenging and requires knowledge of the anatomy and physiology of this unique population.
- The standard of care for anticoagulation in adult patients with Fontan physiology remains vitamin K antagonists; however, research has demonstrated the safety and efficacy of direct oral anticoagulants.
- Catheter-based embolectomy devices can be safely and effectively utilized in the treatment of pulmonary emboli in the Fontan population.

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Introduction

The Fontan operation is a well-described palliative procedure for functional single-ventricle individuals, typically performed during their childhood. As these patients age, long-term complications of this operation become more prevalent. In the adult Fontan population, approximately one-quarter of patients experience an embolic or thrombotic complication,[1] with close to half of those patients experiencing clinically silent pulmonary emboli.[2] The increased risk of thrombo-embolic events can in part be attributed to the intrinsic haemodynamics of the Fontan circuit. Diagnosis remains challenging, as adequate imaging requires an understanding of the unique anatomy and physiology of this population.[3] Additionally, optimal strategies for the prevention and treatment of thrombo-embolic complications in the Fontan population are poorly defined.[4]

Summary figure

Timeline	Event
Birth	Born with severe Ebstein anomaly of the tricuspid valve
Age 5 years	Tricuspid valve replacement, superior cavopulmonary anastomosis, and atrial septal defect closure
Age 12 years	Removal of tricuspid bioprosthesis with patch closure of native tricuspid valve orifices, Fontan completion, and atrial septal defect creation
Age 25 years	Device closure of Fontan fenestration
Age 28 (day of event)	Presents with acute-onset chest pain, dyspnoea, and worsened hypoxia. Chest computed tomographic angiography interpreted as submassive pulmonary embolism. Started on an unfractionated heparin drip
Day 1 post-event	Transferred to an expert Adult Congenital Heart Disease Center
Day 2 post-event	Cardiac catheterization with transcatheter embolectomy of a large acute thrombus in the right lower pulmonary artery. Immediate improvement in oxygen saturations
Day 3 post-event	Transition to direct oral anticoagulants with a plan for life-long anticoagulation. Discharged home
1 year post-event	Doing well in clinic follow-up. No further thrombo-embolic events. Tolerating full-dose anticoagulation without bleeding complication

Case presentation

We present the case of a 28-year-old Hispanic female born with severe Ebstein anomaly of the tricuspid valve. She was initially palliated with right ventricular plication, tricuspid valve repair, and atrial septal defect closure at 5 years of age. She subsequently underwent placement of a 25 mm Hancock™ valve in the tricuspid position and creation of a superior cavopulmonary anastomosis. She developed a high-grade heart block for which an epicardial dual-chamber pacemaker was placed, also at 5 years of age. There was no history of atrial arrhythmia. Her surgical intervention culminated in tricuspid valve removal with patch occlusion of the right ventricle and a fenestrated extracardiac Fontan at 12 years of age. Due to hypoxia, her fenestration was occluded at 25 years of age with a 6 mm Amplatzer™ Septal Occluder via cardiac catheterization, with resulting improvement in oxygen saturations by ~15% (*Figure 1*).

The patient presented to an outside emergency department with 5 days of chest pain, progressive dyspnoea, and fatigue. On the day of presentation, she had become short of breath with minimal exertion. She denied recent illnesses or travel. The patient was hypoxic on arrival at the outside facility with an oxygen saturation of 78%, which improved to 88% on 4 L of oxygen by nasal cannulation. On physical exam, she was tachypnoeic but comfortable appearing without accessory muscle use. She had a well-healed median sternotomy scar. Pulmonary auscultation demonstrates clear lung fields with good airflow. Her cardiac exam was notable for a single S1 and S2, and non-pulsatile jugular venous distension consistent with her known Fontan physiology. The remainder of her exam was benign. An electrocardiogram (ECG) demonstrated sinus tachycardia at a rate of 117 b.p.m. and a right bundle branch block that was pre-existing (*Figure 2*). Her laboratory evaluation demonstrated a haemoglobin of 14.8 g/dL (normal range, 11.5–16.0), haematocrit of 45.3% (37–47), and platelets of 203 K/μL (130–400). There was no evidence of infectious processes. Her international normalized ratio (INR) was 1.15 (≤1). Brain natriuretic peptide and troponin were not elevated. A helical chest computed tomographic angiography (CTA) was obtained and interpreted as showing a submassive pulmonary embolism (PE) with no contrast filling the proximal branch pulmonary arteries.

Her non-cardiac history included hypothyroidism and polycystic ovarian syndrome, for which she took levothyroxine and metformin,

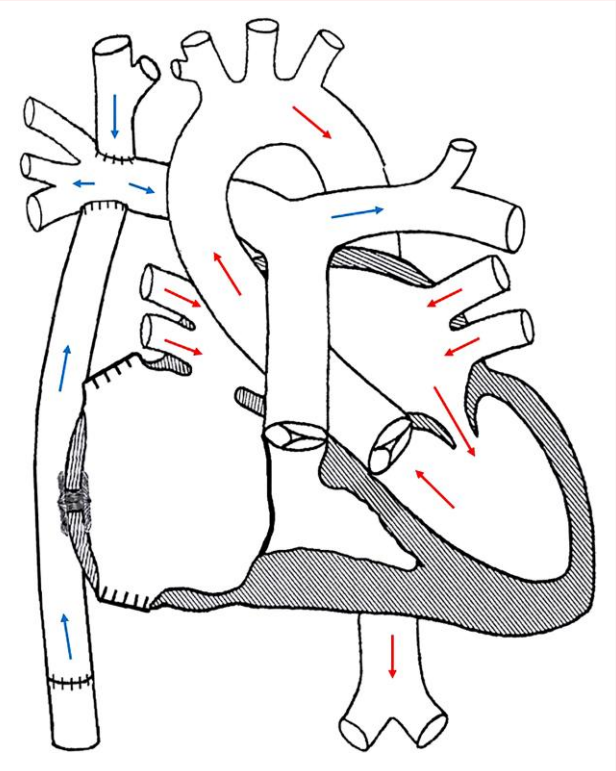

Figure 1 Heart diagram. Ebstein anomaly of the tricuspid valve status post-patch exclusion of the right ventricle, superior cavopulmonary anastomosis, fenestrated Fontan, and subsequent device occlusion of the fenestration.

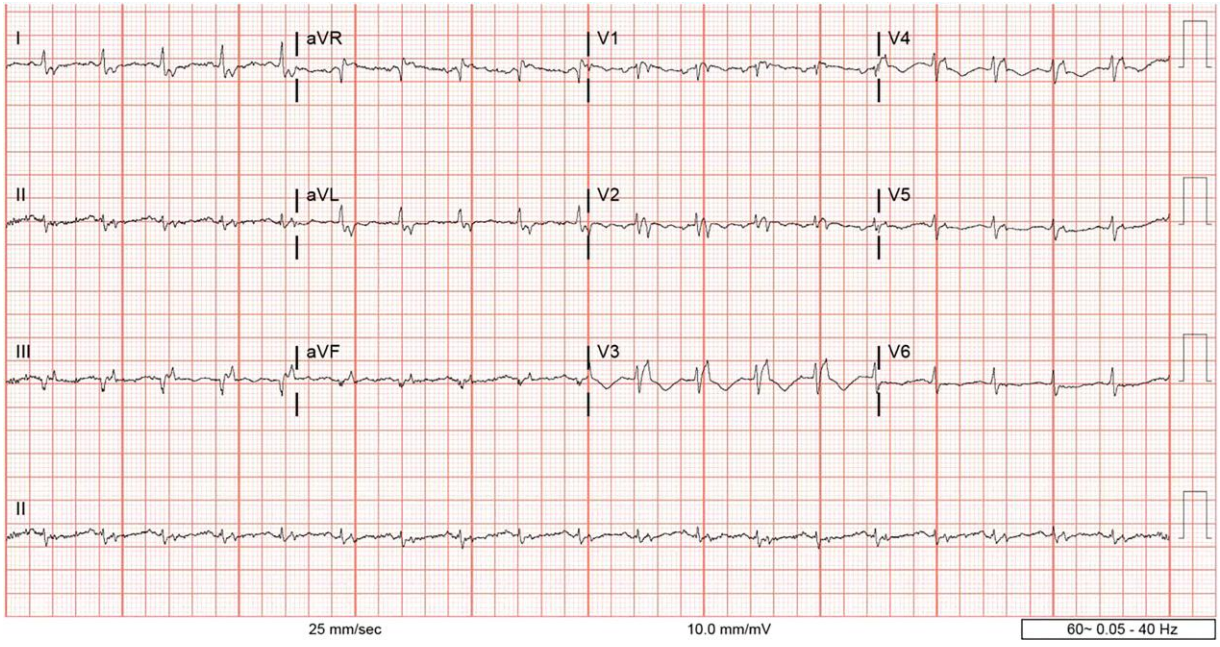

Figure 2 Electrocardiogram. Sinus tachycardia at a rate of 117 b.p.m. and a right bundle branch block that was pre-existing.

respectively. She also had a history of COVID-19 infection, 11 months prior to her presentation, which did not require treatment. The patient had a history of miscarriages with unknown aetiology. She was on rivaroxaban, a direct oral anticoagulant (DOAC) for chronic thromboprophylaxis in the setting of her Fontan physiology. She had recently become pregnant but unfortunately experienced another miscarriage; in this setting, she had discontinued and not reinitiated her anticoagulation.

The differential diagnosis for hypoxia in a patient with a Fontan includes the presence of a Fontan fenestration, venovenous collateral vessels, pulmonary arteriovenous malformations, PE, acute pulmonary processes such as infection, and anaemia.

The patient was haemodynamically stable and was started on an infusion of unfractionated heparin and transferred to our facility for further management. On exam, she was well-appearing and remained short of breath with hypoxia, though she was able to speak in full sentences. Her lungs were clear bilaterally. Her cardiac examination demonstrated a regular rate and rhythm, a single S1 and single S2, and no murmurs or gallop. Her capillary refill was <3 s, and she had no oedema. A D-dimer was obtained and was elevated at 8.48 μg/mL fibrinogen equvalent units (FEU). We reviewed the CTA (*Figure 3*). She underwent cardiac catheterization to better elucidate the cause of her shortness of breath and desaturation. Her Fontan pressure was noted to be elevated at 21 mmHg (previously 14 mmHg 2 years prior). Angiography demonstrated a large thrombus in the right lower pulmonary artery branch, with additional thrombi in the subsegmental branches (*Figure 4A* and *B*). The proximal right pulmonary artery and the entire left pulmonary artery were free of thrombi.

Following confirmation of the right lower pulmonary artery emboli in the setting of elevated Fontan pressures and significant symptomatic hypoxia, it was decided that the patient would benefit from embolectomy. Using the transcatheter Penumbra Indigo™ Aspiration System, the larger right lower pulmonary artery branch segment and three additional subsegmental branches were addressed. A significant clot was removed (*Figure 5*), and post-aspiration angiography demonstrated resolution of the filling defect (*Figure 4C* and *D*).

The patient tolerated the procedure well. Following the procedure, she was restarted on rivaroxaban and her heparin drip was discontinued. She was weaned off supplemental oxygen and discharged home 2 days post-procedure with an oxygen saturation of 92% on room air.

The patient remains well in follow-up. An outpatient thrombophilia workup was obtained and was negative for antiphospholipid syndrome, factor V Leiden, rheumatologic pathology, or other known causes. Given her Fontan physiology and PE, she was instructed to continue life-long anticoagulation and has remained free of further thromboembolic events since admission. She continues to be followed by adult congenital cardiology. In follow-up, at 1 year after the event, she has remained well. She remained on full-dose anticoagulation, which has been well tolerated without bleeding complications or recurrent thromboembolic events.

Discussion

Thrombotic complications are common in the adult Fontan population, with one retrospective study finding that 25% of adult Fontan patients experienced a thrombotic or embolic complication[1] and another noting that 17% of patients experienced clinically silent pulmonary emboli.[2] The risk of thrombo-embolism is present even in the most optimal Fontan patient. Given the morbidity and mortality associated with this complication, the use of proper imaging techniques is imperative. As there is no ventricle pumping to the pulmonary circulation, tests to evaluate for cardiac strain (ECG, troponin, and echocardiogram) have little utility in the Fontan patient. Traditional CTA imaging for PE in Fontan patients often has contrast filling defects due to a combination of the low phasic flow of venous blood through the Fontan circuit into the pulmonary arteries and the streaming phenomenon created by the surgical anastomoses. These two factors result in inadequate

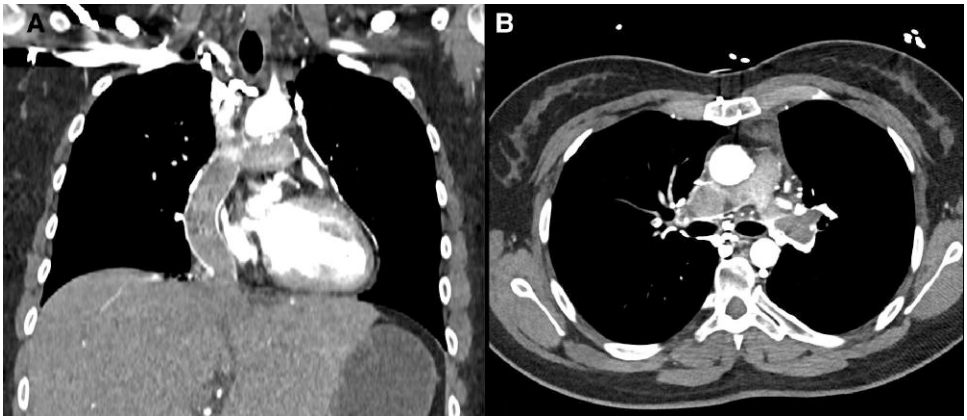

Figure 3 Computed tomography scan. (*A*) Coronal imaging demonstrating the superior and inferior caval anastomoses with a lack of contrast filling and (*B*) axial imaging demonstrating the branch pulmonary arteries with a lack of contrast filling.

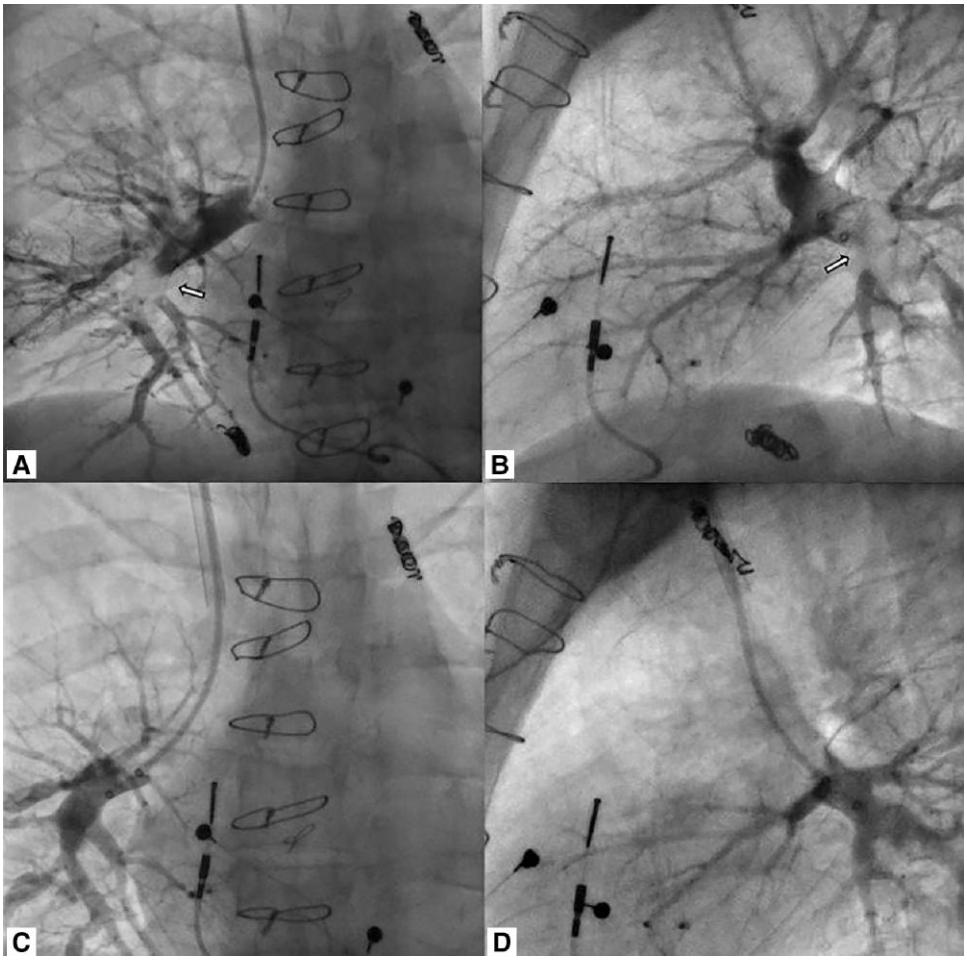

Figure 4 Pre-intervention and post-intervention angiography. The right lower pulmonary artery branch demonstrates a filling defect on antero-posterior (*A*) and lateral angiographic imaging (*B*). Arrows indicate the thrombus. There is resolution of the right lower pulmonary artery branch filling defect on antero-posterior (*C*) and lateral angiographic imaging (*D*) following catheter-based embolectomy.

Figure 5 Large thrombus. The large thrombus removed utilizing a transcatheter aspiration system.

contrast mixing and therefore the presence of both opacified and unopacified blood, which may be interpreted as a thrombus by persons not familiar with the patient's anatomy and physiology. The superior caval (Glenn) anastomosis flow is often preferentially to one lung, and when CT contrast is administered through a single upper extremity, only the ipsilateral lung may demonstrate contrast filling. For optimal contrast filling, it is recommended that a dual-contrast injection into the upper and lower extremities is employed, allowing for opacification of the Glenn anastomosis, Fontan baffle, and bilateral branch pulmonary arteries. Additionally, using a biphasic or dual-phase technique allows for imaging to be obtained immediately following contrast injection as well as at a 60–90 s delay, providing a venous phase and appropriate Fontan conduit opacification.[3] Employing adequate imaging techniques will help to decrease costs and additional radiation exposure associated with the need for repeat imaging, as well as avoid inappropriate hospitalization, need for anticoagulation, and potential need for catheterization to confirm or exclude the presence of PE.

For the acute treatment of haemodynamically significant PE in Fontan patients, clinical care options include systemic parenteral thrombolysis, catheter-directed heparin, or mechanical aspiration. Given the unreliability of imaging in Fontan patients, the risk of bleeding with systemic thrombolysis outweighs the benefits of its use in comparison to other interventions. If there is a concern for true PE, the patient should proceed to cardiac catheterization for confirmation and appropriate treatment (local thrombolysis vs. thrombectomy). If the anatomy and haemodynamics are favourable, we illustrate how endovascular thrombectomy via transcatheter aspiration is a reasonable therapeutic option and provides for immediate improvement in symptoms and oxygen saturation.

For Fontan patients with known or suspected thrombi, prior thrombo-embolic events, and no contraindications to anticoagulation, current guidelines recommend oral anticoagulation with a vitamin K antagonist (i.e. warfarin). Similar anticoagulation may be considered in Fontan patients without known or suspected thrombi or thrombo-embolic events.[4,5] However, warfarin is challenging for patients given its many drug and dietary interventions and cumbersome monitoring requirements. A recent meta-analysis of thromboprophylactic strategies in the Fontan population suggested that DOACs were superior to warfarin in preventing thrombo-embolic events without a significant increase in bleeding risk.[6]

Conclusions

Thrombo-embolic events, including pulmonary emboli, are a relatively common occurrence in the Fontan population that can lead to short- and long-term complications. Given the importance of accurate diagnosis and timely treatment of pulmonary emboli, it is prudent that physicians caring for this population understand Fontan anatomy and physiology and the accompanying diagnostic imaging nuances. We have demonstrated that catheter-based thrombectomy is feasible in Fontan anatomy and that this may provide improved acute benefits with the immediate mechanical removal of the thrombus. Life-long anticoagulation is indicated for any patient with a Fontan and prior thrombotic or embolic event. Warfarin is recommended, but there are recent data supporting the use of DOACs in this population.

Lead author biography

Dr Christina Benjamin, DO, received her medical degree from Des Moines University in 2017. She completed her paediatric residency at the University of Oklahoma—Tulsa School of Community Medicine in 2020. Dr Benjamin recently completed her paediatric cardiology fellowship at Phoenix Children's in AZ, USA, and will remain on as faculty at Phoenix Children's.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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Data availability

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

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