

Rudimentary Right Ventricle Thrombus Leading to Multifocal Stroke in Fontan Circulation

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Abstract

Presentation

Patients with Fontan circulation are at increased risk of thromboembolism. We report a case of multifocal stroke caused by thrombus in a rudimentary right ventricle and highlight the need to consider stroke in Fontan patients with neurological symptoms.

Diagnosis

A CT brain which presented normal. MRI/MRA brain performed demonstrating multifocal shower of infarcts consistent with an acute embolic cerebrovascular event. Transthoracic echo was normal. Transoesophageal echo located a thrombus within the trabeculations of the rudimentary right ventricle, confirming a diagnosis of thromboembolic stroke secondary to RV thrombus.

Treatment

Multidisciplinary care including cardiology, neurology, and haematology. Commenced on anticoagulation with an INR target range of 2-3. Possible future catheter procedure to occlude right ventricle.

Discussion

This case highlights the persistent risk of thromboembolic events in Fontan patients in whom residual cardiac structures such as a rudimentary ventricle can serve as a source of thrombus. Multimodal imaging is critical for diagnosis. Optimal thromboprophylaxis remains uncertain.

Introduction

The Fontan procedure is the final stage of surgical palliation for patients with single-ventricle physiology. Its unique haemodynamics predispose patients to lifelong thromboembolic risk

from venous stasis, endothelial dysfunction, and prothrombotic coagulation abnormalities¹⁻³. Atrial arrhythmias further increase the risk⁴. Both overt and silent cerebral infarctions contribute to morbidity². No consensus exists on post-Fontan antithrombotic therapy. We report a case of multifocal cerebral infarction in a stable Fontan patient.

Case Report

An 11-year-old boy, AR, with a background of pulmonary atresia with intact ventricular septum and completed Fontan circulation was admitted to Cork University Hospital after transient right-sided hemiplegia. He was treated with Warfarin for 12 months post-Fontan, then maintained on daily aspirin with good compliance.

On the evening of admission, AR developed right arm, leg, and facial weakness. There was no associated headache, seizure activity, or loss of consciousness. His symptoms began resolving within 25 minutes and he had complete resolution of symptoms approximately 2 hours later. Neurological examination was normal, with no residual deficits, and an initial CT brain showed no acute abnormality.

Considering his underlying Fontan physiology, a transient ischaemic attack (TIA) was suspected. MRI/MRA brain was performed revealing acute infarcts in the left basal ganglia, left parietal lobe, left anterior insula, and superior right frontal lobe, consistent with a multifocal embolic pattern. The major branches of the Circle of Willis were patent.

In view of these findings, AR was transferred to CHI Crumlin for further cardiac evaluation. Transoesophageal echocardiography demonstrated thrombus within the trabeculations of his rudimentary right ventricle. Otherwise, his echocardiography demonstrated a patent Fontan circuit with no fenestration and preserved single ventricular function.

Therapeutic anticoagulation was initiated with enoxaparin and subsequently transitioned to Warfarin with a target INR of 2-3. Lifelong therapy is likely in view of his persistent risk factors. Consideration is being made for cardiac catheterisation to occlude flow into the rudimentary RV via the regurgitant tricuspid valve.

Given his excellent early recovery, a favourable long-term neurological outcome is anticipated.

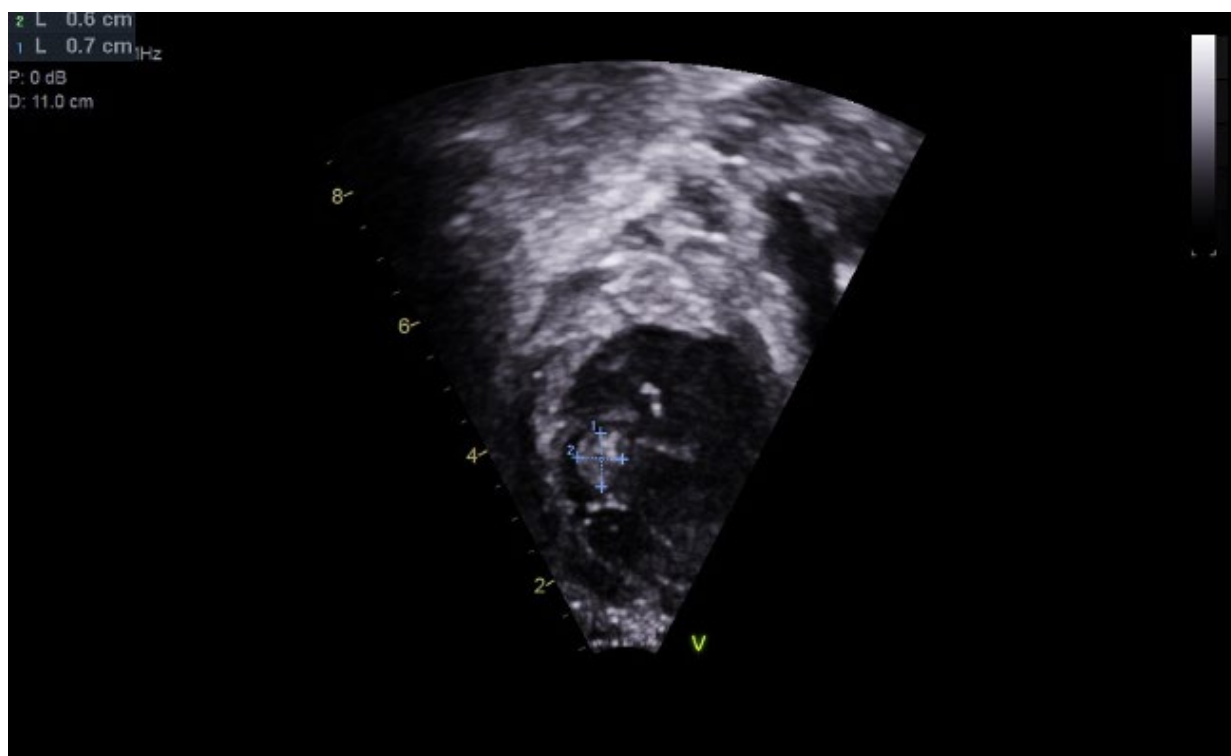


Figure 1: 0.6 x 0.7cm thrombus within trabeculations of rudimentary right ventricle.

Discussion

This case highlights the persistent risk of thromboembolic events in Fontan patients. Thromboembolic events are well documented in this patient population, with venous thrombosis reported in 4–19%, stroke in 3–19%, and intracardiac thrombosis in 17–33% of cases^{1,2}. Silent cerebral infarctions are common with many events only identified by surveillance imaging⁵. MRI studies show up to 39% prevalence of subclinical stroke².

Residual cardiac structures, such as the rudimentary ventricle seen in this case, can serve as a reservoir for thrombus formation despite a well-functioning Fontan circulation⁶. Interventional strategies, including catheter-based occlusion of such chambers, may be considered in high-risk cases to reduce thromboembolic risk, although evidence guiding patient selection and long-term outcomes remains limited.

In paediatric embolic arterial stroke, multiple small emboli may result in low-volume infarcts. Preserved collateral circulation, including via the Circle of Willis, can maintain perfusion to tissues. In addition, paediatric neuroplasticity may facilitate rapid functional recovery, resulting in minimal or transient neurological deficits despite radiological evidence of infarction⁷, as observed in this case.

Aspirin is frequently used for thromboprophylaxis, but resistance occurs in up to 52% of patients⁸. Warfarin provides more robust protection but is difficult to manage in children, with up to 28% of paediatric patients remaining subtherapeutic more than 70% of the time³. Direct oral anticoagulants (DOACs) are emerging options. The UNIVERSE trial demonstrated the feasibility of direct oral anticoagulation with rivaroxaban in paediatric patients following Fontan completion, although long-term safety and efficacy data remain limited⁹. Current guidelines from the American Heart Association and European Society of Cardiology recommend vitamin K antagonists or low molecular weight heparin for 3–12 months post-Fontan, with aspirin continued long-term unless additional risk factors justify ongoing anticoagulation⁸.

This case highlights the persistent risk of thromboembolic events in Fontan patients, and that multimodal imaging (MRI, TOE) is critical for diagnosis. The evidence base guiding decisions around prophylactic and therapeutic anticoagulation consists mainly of small trials and observational studies, with variable clinical practice. Individualised decisions regarding lifelong anticoagulation should consider factors such as thrombus occurrence, presence or absence of fenestration and flow dynamics. With increasing Fontan survival into adulthood, proactive prevention, surveillance imaging, and multidisciplinary care are essential to optimise outcomes.

Declarations of Conflicts of Interest:

None declared.

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References:

1. Van Den Helm S, Sparks CN, Ignjatovic V, Monagle P, Attard C. Increased risk for thromboembolism after Fontan surgery: considerations for thromboprophylaxis. *frontiersin.org* S Van Den Helm, CN Sparks, V Ignjatovic, P Monagle, C Attard *Frontiers in Pediatrics*, 2022 • *frontiersin.org* [Internet]. 2022 Mar 28 [cited 2025 Aug 4];10. Available from: <https://www.frontiersin.org/articles/10.3389/fped.2022.803408/full>
2. Van den Eynde J, Possner M, Alahdab F, Veldtman G, Goldstein BH, Rathod RH, et al. Thromboprophylaxis in patients with Fontan circulation. *jacc.org* J Van den Eynde, M Possner, F Alahdab, G Veldtman, BH Goldstein, RH Rathod *Journal of the American College of Cardiology*, 2023 • *jacc.org* [Internet]. 2023 Jan 31 [cited 2025 Aug 4];81(4):374–89. Available from: <https://www.jacc.org/doi/abs/10.1016/j.jacc.2022.10.037>
3. McCrindle BW, Manlhiot C, Cochrane A, Roberts R, Hughes M, Szechtman B, et al. Factors associated with thrombotic complications after the Fontan procedure: a secondary analysis of a multicenter, randomized trial of primary thromboprophylaxis for. *jacc.org* [Internet]. 2013 Jan 22 [cited 2025 Aug 4];61(3):346–53. Available from: <https://www.jacc.org/doi/abs/10.1016/j.jacc.2012.08.1023>
4. Alsaied T, Alsidawi S, Allen C, Heart JF, 2015 undefined. Strategies for thromboprophylaxis in Fontan circulation: a meta-analysis. *heart.bmj.com* T Alsaied, S Alsidawi, CC Allen, J Faircloth, JS Palumbo, GR Veldtman *Heart*, 2015 • *heart.bmj.com* [Internet]. [cited 2025 Aug 4]; Available from: <https://heart.bmj.com/content/101/21/1731.short>
5. Al-Jazairi AS, Al Alshaykh HA, Di Salvo G, De Vol EB, Alhalees ZY. Assessment of Late Thromboembolic Complications Post-Fontan Procedure in Relation to Different Antithrombotic Regimens: 30-Years' Follow-up Experience. *journals.sagepub.com* AS Al-Jazairi, HA Al Alshaykh, G Di Salvo, EB De Vol, ZY Alhalees *Annals of Pharmacotherapy*, 2019 • *journals.sagepub.com* [Internet]. 2019 Aug 1 [cited 2025 Aug 4];53(8):786–93. Available from: <https://journals.sagepub.com/doi/abs/10.1177/1060028019829860>
6. Egbe A, Connolly H, Niaz T, ... VYA heart, 2017 undefined. Prevalence and outcome of thrombotic and embolic complications in adults after Fontan operation. *ElsevierAC* Egbe, HM Connolly, T Niaz, V Yogeswaran, NW Taggart, MY Qureshi, JT Poterucha *American heart journal*, 2017 • *Elsevier* [Internet]. [cited 2025 Aug 4]; Available from: <https://www.sciencedirect.com/science/article/pii/S0002870316302101>

7. Blackburn E, Darco | Felice, Devito | Andrea, Ioppolo R, Lorio | Sara, Quirk | Bernadine, et al. DEVELOPMENTAL MEDICINE & CHILD NEUROLOGY Predictors of motor outcome after childhood arterial ischemic stroke. 2021 [cited 2026 Jan 22]; Available from: <https://www.>
8. Giglia TM, Massicotte MP, Tweddell JS, Barst RJ, Bauman M, Erickson CC, et al. Prevention and treatment of thrombosis in pediatric and congenital heart disease: a scientific statement from the American Heart Association. ahajournals.org TM Giglia, MP Massicotte, JS Tweddell, RJ Barst, M Bauman, CC Erickson, TF Feltes *Circulation*, 2013 • ahajournals.org [Internet]. 2013 Dec 17 [cited 2025 Aug 4];128(24):2622–703. Available from: <https://www.ahajournals.org/doi/abs/10.1161/01.cir.0000436140.77832.7a>
9. Zhu P, Willmann S, Zhou W, Yang H, Michelson AD, McCrindle BW, et al. Dosing Regimen Prediction and Confirmation With Rivaroxaban for Thromboprophylaxis in Children After the Fontan Procedure: Insights From the Phase III UNIVERSE Study. *J Clin Pharmacol* [Internet]. 2022 Feb 1 [cited 2026 Jan 22];62(2):220. Available from: <https://pmc.ncbi.nlm.nih.gov/articles/PMC9303933/>