Acute ST-elevation myocardial infarction with Guillain-Barre syndrome in Fontan circulation

Rohit Sunil Walse, Bhagwati Pant, Arun Gopalakrishnan, Bijulal Sasidharan
Department of Cardiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram, Kerala, India

ABSTRACT

Total cavopulmonary connection, commonly referred to as the Fontan procedure, is the established destination therapy for univentricular hearts. While the procedure permits a longer survival of these patients, this circulation involves several compromises from normal human circulation and poses several challenges with increasing age after surgery. We present an instance of acute ST-elevation myocardial infarction with Guillain-Barre syndrome in an adult after Fontan palliation and discuss the challenges in management.

Keywords: Adult congenital heart disease, grown up with congenital heart disease, percutaneous coronary intervention, total cavopulmonary connection

INTRODUCTION

Total cavopulmonary connection, commonly referred to as the Fontan procedure, is the established destination therapy for complex univentricular hearts. The Fontan pathway is a milestone in the palliation of children born with complex univentricular heart diseases, 90% of whom would not have survived beyond their first birthday without intervention. While the procedure permits a longer survival of these patients, this circulation involves several compromises from normal human circulation and poses several challenges with increasing age after surgery. We present an instance of acute ST-elevation myocardial infarction in an adult after Fontan palliation and discuss the challenges in management.

CASE REPORT

The index patient is a 33-year-old female with complex univentricular congenital heart disease who had been palliated with lateral tunnel Fontan procedure. She had been diagnosed with tricuspid atresia, ventricular septal defect, and pulmonary atresia in early infancy. After initial palliation with a right modified Blalock-Taussig shunt (BTS), she had undergone single-stage lateral tunnel Fontan completion with interruption of the right BTS at 5 years of life. The postoperative period was uneventful, and she was on regular follow-up in the Fontan clinic. She had been diagnosed with paroxysmal atrial tachycardia at 31 years of life and was instituted on warfarin with a target international normalized ratio (INR) of 2.0–3.0.

The patient had presented with acute-onset bilateral lower limb and upper limb weakness with drooping of eyelids. On admission under neurology care, her vital signs were normal, and her higher functions were noted to be appropriate. Neurological examination showed bilateral ptosis with facial droop. Her muscle strength was 1/5 in both the lower limbs and 3/5 in the upper limbs. She was in sinus rhythm, and the blood pressure was recorded as 118/70 mmHg. Precordial examination

Video available on: https://journals.lww.com/aopc

Access this article online

Quick Response Code:

Website: https://journals.lww.com/aopc

DOI: 10.4103/apc.apc_134_22

How to cite this article: Walse RS, Pant B, Gopalakrishnan A, Sasidharan B. Acute ST-elevation myocardial infarction with Guillain-Barre syndrome in Fontan circulation. Ann Pediatr Card 2023;16:141-3.

Address for correspondence: Dr. Arun Gopalakrishnan, Department of Cardiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Thiruvananthapuram - 695 011, Kerala, India.
E-mail: arungopalakrishnan99@gmail.com

Submitted: 27-Oct-2022 **Revised:** 04-Dec-2022 **Accepted:** 07-Feb-2023 **Published:** 16-Aug-2023

revealed normal cardiac size, normal first heart sound, single second heart sound, and no cardiac murmurs. Anti-GQ1b antibody titer was noted to be elevated. She was diagnosed with acute inflammatory demyelinating polyneuropathy (Guillain-Barre syndrome) and was treated with plasmapheresis.

In the intensive care, frequent atrial tachyarrhythmias were noted (atrial tachycardia with varying block and atrial flutter). A successful pharmacological cardioversion was done with amiodarone infusion, followed by oral maintenance therapy. Echocardiography showed normal ventricular function, no atrioventricular valve regurgitation, and normal flow in the cavopulmonary circuits. During the hospital stay, oral anticoagulation was withheld temporarily before plasmapheresis therapy. Following the third cycle of plasmapheresis, she developed acute retrosternal chest pain with ventricular fibrillation within 2 min [Figure 1] and required multiple defibrillation shocks. She had to be mechanically ventilated and was put on inotropic support. The cardiac troponin-T was elevated at 230 ng/ml. A 12-lead electrocardiogram showed ST-segment elevation in leads I, augmented vector left with corresponding ST-segment depression in II, III, augmented vector foot (aVF), V2-V6. She was then rushed to the cardiac catheterization laboratory.

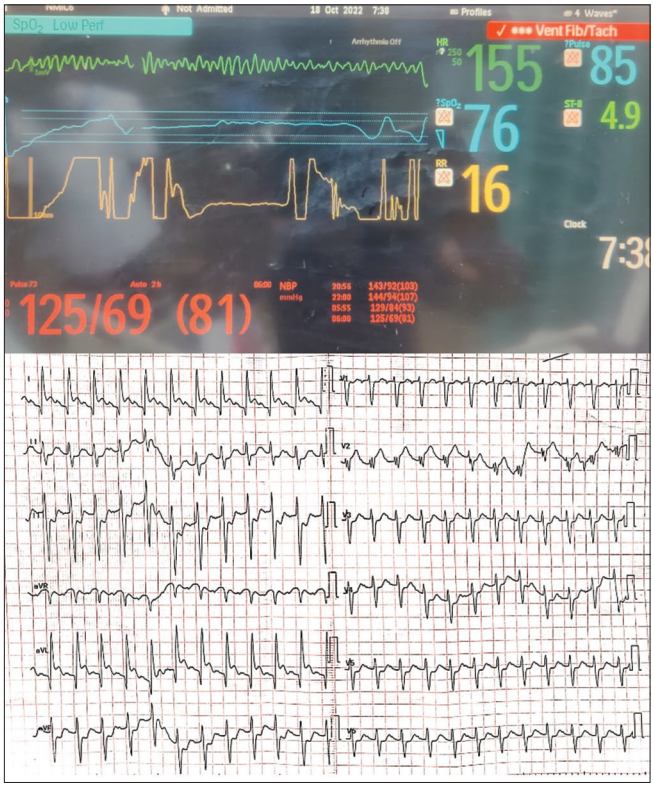

Figure 1: Upper panel shows monitor electrocardiographic trace with ventricular fibrillation; lower panel shows 12-lead electrocardiogram with ST-segment elevation in I, augmented vector left and reciprocal ST-segment depression in II, III, augmented vector foot (aVF), V2-V6

Coronary angiogram showed complete thrombotic occlusion of the left anterior descending (LAD) artery [Figure 2]. A stable position distal to the lesion was achieved with a 0.014-inch coronary guidewire through a 6F extra-backup guide catheter. Serial aspiration thrombectomy runs were performed using an Export AP aspiration catheter (Medtronic, Minneapolis, MN); however, due to a small residual thrombus, balloon angioplasty was performed. Postangioplasty angiography showed migration of the thrombus to diagonal branches which also was aspirated. Final angiogram showed complete thrombus resolution, and the LAD flow improved significantly with thrombolysis in myocardial infarction 3 flow. The right coronary angiography revealed normal right coronary artery with coronary cameral fistula to the coronary sinus [Video 1]. She was transferred to the intensive care unit on aspirin, clopidogrel, and heparin infusion in stable condition with no further recurrence of arrhythmic episodes. She was extubated to room air the next day, and her vitals remained stable thereafter. Neurologically, there was a complete recovery of power to Grade 5 in all limbs over the next 2 days. The patient was discharged on warfarin, aspirin, and clopidogrel.

DISCUSSION

The Fontan circulation is prone to thromboembolic events due to the sluggish nature of the revised systemic venous circuit, acquired procoagulant state due to altered hepatic synthetic function, endothelial dysfunction, and the absence of the right ventricular pump. While 29% of atriopulmonary Fontan patients are noted to have systemic thromboembolic events by the end of the third decade, this figure is lower in lateral tunnel Fontan (15%) and least in extracardiac (8%).[1] Atrial arrhythmia is a

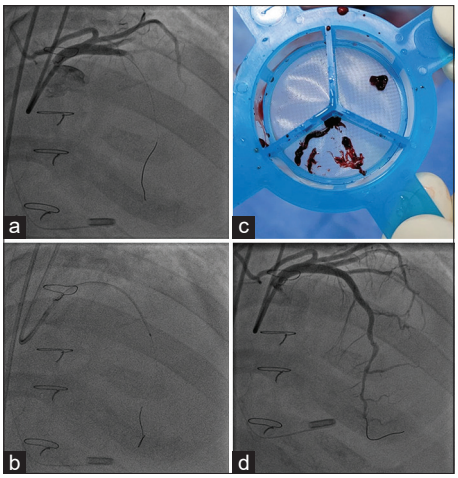

Figure 2: (a) Left coronary angiogram showing thrombotic occlusion of the left anterior descending artery. (b) Thrombectomy with export catheter. (c) Large aspirated thrombus. (d) Final angiogram showing complete recanalization of the left anterior descending artery

definite predisposing factor for thromboembolic events post-Fontan palliation.

However, there is no uniform consensus for indefinite long-term thromboprophylaxis in lateral tunnel and extracardiac Fontan. The highest thrombotic risk is in the initial months following Fontan completion due to the altered hemodynamics, endothelial dysfunction, perioperative stress, and hepatic dysfunction, due to which anticoagulation with Vitamin K antagonists (VKA) is almost universally followed.[2] The period from 2 to 10 years after Fontan constitutes a plateau phase with low thromboembolic incidence. Thromboprophylaxis with VKA was no better than aspirin during this period, while the former was associated with higher bleeding.[3] Thereafter, the thrombotic risk increases in adulthood and tends to parallel the increase in arrhythmic burden.[4] While the European and American guidelines recommend VKA for patients with Fontan and a history of atrial thrombus, atrial arrhythmia, or prior thromboembolic events, the committee did not recommend it for all Fontan patients.[5,6]

The effect of plasmapheresis on anticoagulant function in patients receiving therapeutic anticoagulation has been a matter of debate. While several clotting factors are removed during plasmapheresis, the effects of the procedure on the INR and activated partial thromboplastin time and anti-Xa levels have been variably reported in the literature.[7] The event in the index case could be related to the thrombotic milieu due to atrial arrhythmia and altered coagulation profile during plasmapheresis leading to embolic coronary artery occlusion.

CONCLUSIONS

Adults with Fontan circulation are at increased risk of thromboembolic events, including embolic myocardial infarction. Mechanical thrombectomy is a potentially safe and feasible therapy for coronary thromboembolism in these patients. Oral anticoagulation with VKAs should never be stopped in patients with Fontan circulation and atrial arrhythmia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Egbe AC, Connolly HM, Niaz T, Yogeswaran V, Taggart NW, Qureshi MY, *et al.* Prevalence and outcome of thrombotic and embolic complications in adults after Fontan operation. Am Heart J 2017;183:10-7.
2. Marrone C, Galasso G, Piccolo R, de Leva F, Paladini R, Piscione F, *et al.* Antiplatelet versus anticoagulation therapy after extracardiac conduit Fontan: A systematic review and meta-analysis. Pediatr Cardiol 2011;32:32-9.
3. Iyengar AJ, Winlaw DS, Galati JC, Wheaton GR, Gentles TL, Grigg LE, *et al.* No difference between aspirin and warfarin after extracardiac Fontan in a propensity score analysis of 475 patients. Eur J Cardiothorac Surg 2016;50:980-7.
4. Heidendael JF, Engele LJ, Bouma BJ, Dipchand AI, Thorne SA, McCrindle BW, *et al.* Coagulation and anticoagulation in Fontan patients. Can J Cardiol 2022;38:1024-35.
5. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, *et al.* ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010;31:2915-57.
6. 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.
7. Hodulik KL, Root AG, Ledbetter LS, Onwuemene OA. Effects of therapeutic plasma exchange on anticoagulants in patients receiving therapeutic anticoagulation: A systematic review. Transfusion 2019;59:1870-9.