



Percutaneous coronary intervention following Fontan procedure

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ABSTRACT

Fontan procedure is known to increase the risk of thromboembolic events. However, coronary artery thrombotic occlusion is rarely reported in patients with Fontan procedure. We present a case of a 10-year-old boy with hypoplastic left heart syndrome palliated with a Fontan procedure who presented with myocardial infarction secondary to thrombotic occlusion of the left circumflex coronary artery. He underwent successful percutaneous coronary intervention with thrombus aspiration, balloon angioplasty, and stent placement, highlighting the necessity of collaboration between congenital and adult cardiologists to treat acute coronary syndrome among this challenging young population.

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A 10 year old boy born with a history of hypoplastic left heart syndrome (mitral stenosis and aortic stenosis), with past history of stage I hybrid palliation (bilateral pulmonary arteries were banded and ducts arteriosus stent was then placed) followed by comprehensive stage II bilateral Glenn anastomosis, arch reconstruction, atrial septectomy, removal of the ducts arteriosus stent and removal of bilateral pulmonary artery bands. His postoperative course was complicated and needed placement of bilateral pulmonary artery stents after the second surgery. Finally, he underwent fenestrated extracardiac Fontan completion at age 3 with fenestration closure through cardiac catheterization at age 9 due to increasing cyanosis. He was on furosemide, enalapril, aspirin, and regular follow-up in the pediatric cardiology clinic.

He presented with sudden onset, acute chest pain. Troponin was elevated at 33 ng/milliliter (ng/ml). Electrocardiogram (ECG) confirmed a new V2 to V4 ST-segment depression. He was taken to the catheterization lab. Coronary angiogram showed complete occlusion of the proximal left circumflex coronary artery (LCX) by thrombus embolization (Fig. 1). An adult interventional cardiologist was called to assist in coronary intervention. A stable position distal to the lesion was achieved with a 0.014-inch coronary guide-wire through a 6-Fr left Judkins guide catheter. Serial aspiration thrombectomy was performed using an Export AP aspiration catheter (Medtronic, Minneapolis MN); however, due to a small residual thrombus, balloon angioplasty was performed. Post-angioplasty angiography showed complete thrombus resolution,

and the left circumflex flow improved significantly with Thrombolysis in Myocardial Infarction (TIMI) 3 flow (Fig. 2). He was transferred to the intensive care unit (ICU) on aspirin, clopidogrel, and heparin drip in stable condition.

However, 24 h later, the patient continued to complain of chest pain and nausea. A repeat troponin increased to 46.65 ng/ml. ECG confirmed lateral V5 and V6 ST-segment elevation. Trans-thoracic echocardiography showed new deterioration in the right ventricular systolic function, with apical, and interventricular septum hypokinesia. His clinical picture was concerning for coronary re-embolization or post-intervention complications (dissection, hematoma, or subacute recoil). He was emergently taken to the cath lab, where a significant clot burden in the left anterior descending (LAD) and LCX was found. Subsequently, mechanical aspiration thrombectomy was performed using the CATRX Indigo System (Penumbra, Inc, Alameda, California) with clots aspirated from both LAD and LCX. Following aspiration, a local injection of nitroglycerin was performed in both vessels. However, significant stenosis, which wasn't present at initial catheterization, was noted in the proximal LCX (Fig. 3). Nevertheless, there was no evidence of dissection. The persistent stenosis seen in the proximal LCX could be related to residual thrombus or post-angioplasty trauma on the vessel wall that may be produced a hematoma of the vessel wall or a subacute recoil of the vessel. Given the persistent stenosis, and the high risk of reocclusion of the LCX, a drug-eluting Onyx 2.5 m m × 15 mm resolute stent (Medtronic, Minneapolis MN) was placed in the proximal LCX. Final angiography showed significant improvement of flow in the LCX (Fig. 4). He was transferred to the intensive care unit (ICU) on aspirin, clopidogrel, and heparin

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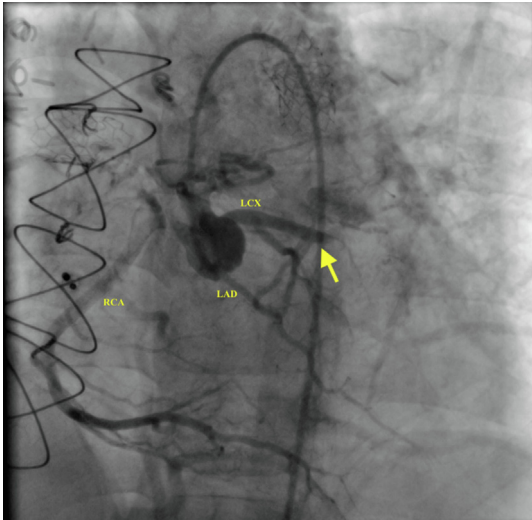


Fig. 1. Coronary angiogram showing occlusion of proximal LCX coronary artery (yellow arrow). (LAD: left anterior descending artery. LCX: left circumflex artery. RCA: right coronary artery).

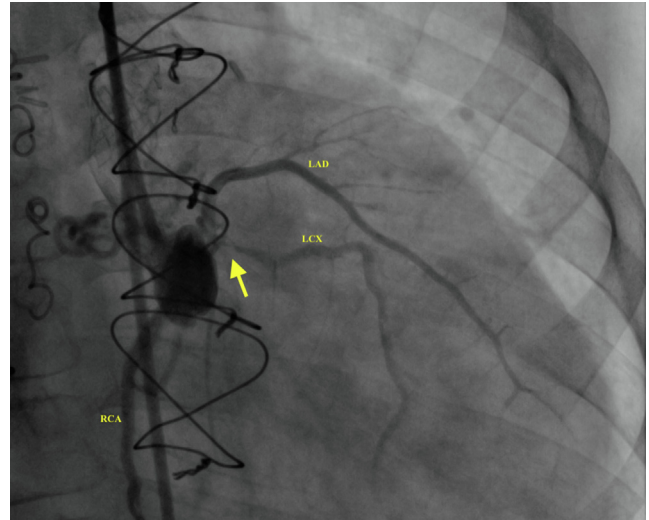


Fig. 3. Coronary angiogram showing significant stenosis in the proximal LCX coronary artery (yellow arrow). (LAD: left anterior descending artery. LCX: left circumflex artery. RCA: right coronary artery).

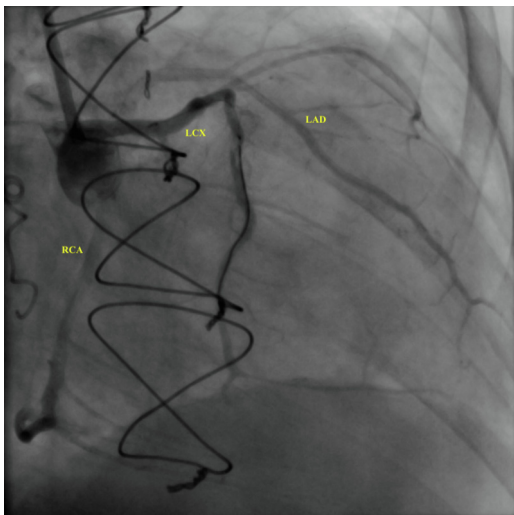


Fig. 2. Post-aspiration angiography showing complete thrombus resolution, the LCX flow improved significantly. (LAD: left anterior descending artery. LCX: left circumflex artery. RCA: right coronary artery).

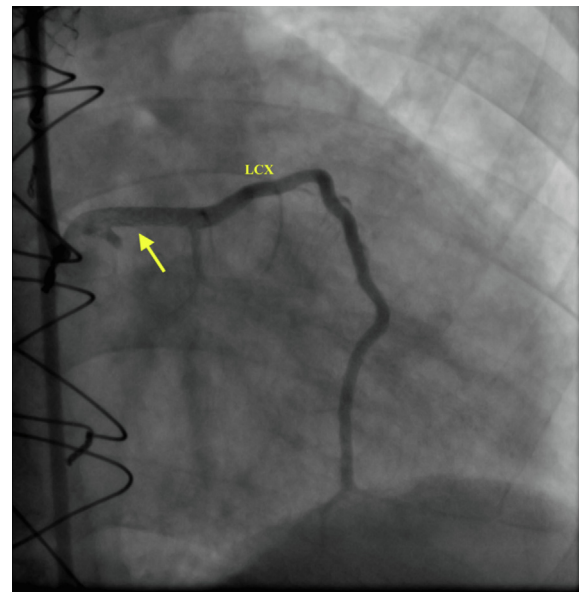


Fig. 4. Final angiography showing significant improvement of flow in the LCX with proximal drug-eluting Onyx 2.5 mm x 15 mm resolute stent (yellow arrow).

drip in stable condition. Overnight he did very well with complete resolution of his symptoms. Seven days after the second procedure, there was a significant improvement in cardiac function. The patient was discharged home on metoprolol, aspirin, clopidogrel, and warfarin, in addition to his home medications. The patient has been seen by the transplant surgery team and is currently being worked up for a cardiac transplant.

The Fontan procedure is a palliative procedure performed in patients with many forms of single ventricle including, hypoplastic left heart syndrome (HLHS) [1]. It aims to decrease the workload of the right ventricle, which supports both the pulmonary and systemic circulations [1]. Fontan procedure is known to increase the risk of systemic or pulmonary thromboembolic events [2,3]. Fontan patient has all the risk factors described by Virchow's triad [4], including stasis due to sluggish intracardiac blood flow or non-detected arrhythmia, the procoagulant state with deficiencies in protein C, protein S, and antithrombin III due to congestive hepatopathy, and endothelial dysfunction with multiple intravascular

prosthetic materials. The prevalence of thromboembolic events among Fontan patients is about 25 percent [2]. Although coronary artery thrombotic occlusion is uncommon in Fontan patients [2], however, it has been described in the literature [5–10]. In the retrospective review of 387 adults with a previous Fontan operation by Egbe et.al. no patient had a myocardial infarction [2]. Systemic thromboembolic events occur with increased frequency in Fontan patients with atrial arrhythmias or fenestration communication between the right atrium/Fontan connection and the left atrium. Furthermore, intracardiac thrombus formation in adults with Fontan circulation, carries a significant risk of death, especially in hemodynamically unstable patients [11]. We present a case of myocardial infarction (MI) secondary to thrombotic occlusion in a 10-year-old patient with a Fontan circulation, who underwent successful percutaneous coronary intervention (PCI) with thrombus aspiration and stent placement. The present case may be the

first reported case of using a percutaneous stent as a supplemental strategy in addition to thrombus aspiration for treating a thromboembolic MI in an HLHS Fontan patient. Performing PCI in patients with HLHS and Fontan circulation is a challenging task, especially in the setting of acute myocardial infarction where time is critical.

Initially, we attempted to reestablish coronary blood flow through thrombus aspiration. However, stenting was used in the second procedure in view of the newly discovered proximal LCX stenosis, which wasn't present at initial catheterization. The persistent stenosis in the proximal LCX could be related to residual thrombus or post-angioplasty trauma on the vessel wall that may be produced a hematoma of the vessel wall or a subacute recoil of the vessel. Given the persistent stenosis and the high risk of reocclusion of the LCX, a drug-eluting stent was placed in the proximal LCX. The adult interventional cardiology team faced a technical challenge while engaging the coronary ostia via the hypoplastic aortic root to place the stent. However, the assistance of the pediatrics interventional team allowed successful engagement. Given the initial complete occlusion of the LCX and the LAD coronary arteries by a thrombus on cardiac catheterization, the most likely diagnosis was a coronary artery embolism. The plausible source of embolism was in either the aortic root or the cardiac chambers. Although, no obvious culprit was identified. Our patient hypercoagulable workup was positive for factor V Leiden thrombophilia. There is no clear consensus regarding the primary prevention of thromboembolic events after Fontan procedure [3,12]. Current evidence suggests that it is reasonable for all patients with Fontan circulation to receive aspirin [3]. However, warfarin is usually used in Fontan patients with documented thromboembolic events for secondary prevention [1,3]. Given the unusual presentation, the recent coronary stenting and suspected embolic event, treatment with warfarin, clopidogrel, and aspirin were initiated.

In conclusion, there is scarce literature that provides evidence-based guidance to optimal treatment of coronary thromboembolism in the setting of Fontan circulation. Therefore, this case demonstrates that thrombectomy and stent placement is a potentially feasible and lifesaving therapy that can be considered in this challenging young population. It also highlights the benefit of shared experience between congenital and adult cardiologists.

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None of the authors have any source of funding to declare.

Declaration of Competing Interest

None of the authors have any conflicts of interest to declare.

Appendix A. Supplementary material

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ijcha.2020.100511>.

References

- [1] C.A. Warnes, R.G. Williams, T.M. Bashore, J.S. Child, H.M. Connolly, J.A. Dearani, P. Del Nido, J.W. Fasules, T.P. Graham, Jr., Z.M. Hijazi, S.A. Hunt, M.E. King, M.J. Landzberg, P.D. Miner, M.J. Radford, E.P. Walsh, G.D. Webb, ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons, *J. Am. Coll. Cardiol.* 52(23) (2008) e143–e263.
- [2] A.C. Egbe, H.M. Connolly, T. Niaz, V. Yogeswaran, N.W. Taggart, M.Y. Qureshi, J. T. Poterucha, A.R. Khan, D.J. Driscoll, Prevalence and outcome of thrombotic and embolic complications in adults after Fontan operation, *Am. Heart J.* 183 (2017) 10–17.
- [3] J. Rychik, A.M. Atz, D.S. Celermajer, B.J. Deal, M.A. Gatzoulis, M.H. Gewillig, T.-Y. Hsia, D.T. Hsu, A.H. Kovacs, B.W. McCrindle, J.W. Newburger, N.A. Pike, M. Rodefeld, D.N. Rosenthal, K.R. Schumacher, B.S. Marino, K. Stout, G. Veldtman, A.K. Younoszai, Y. d'Udekem, Evaluation and management of the child and adult with Fontan circulation: a scientific statement from the American heart association, *Circulation* 140 (6) (2019) e234–e284.
- [4] P.R. Ermis, D.L. Morales, The adult Fontan patient: update for 2011, *Methodist. Debakey Cardiovasc. J.* 7 (2) (2011) 3–8.
- [5] E.M. Graham, H. Shakir, A.M. Atz, J.M. Ringewald, S.M. Bradley, Neo-aortic root modification for late thrombosis after Norwood palliation, *Ann. Thorac. Surg.* 82 (5) (2006) e29–e30.
- [6] G.S. Morchi, R.R. Wolfe, J.D. Kay, Antiphospholipid syndrome leading to myocardial infarction in a Fontan patient 17 years after Norwood palliation, *Congenit. Heart Dis.* 4 (1) (2009) 54–58.
- [7] R.S. Hastings, D.B. McElhinney, M. Saric, C. Ngai, A.H. Skolnick, Embolic myocardial infarction in a patient with a Fontan circulation, *World J. Pediatr. Congenit. Heart Surg.* 5 (4) (2014) 631–634.
- [8] A.R. Keraliya, D.J. Murphy, M.L. Steigner, R. Blankstein, Thrombus in hypoplastic aorta: an uncommon cause of acute myocardial infarction, *J. Cardiovasc. Comput. Tomogr.* 10 (3) (2016) 263–264.
- [9] S.R. Deshpande, A. Dalal, D.W. Kim, V. Babaliaros, M. Jokhadar, Acute embolic myocardial infarction and heart failure in a Fontan patient: recovery with impella device and successful transplantation, *ASAIO J.* 62 (6) (2016) e52–e54.
- [10] R. Shamoony, H. Habib, U. Rampal, A. Hamdan, M. Bikkina, F. Shamoony, A rare case of embolic ST-elevation myocardial infarction in an adult patient with repaired hypoplastic left heart syndrome, *World J. Pediatr. Congenit. Heart Surg.* 8 (4) (2017) 543–549.
- [11] W. Tsang, B. Johansson, O. Salehian, J. Holm, G. Webb, M.A. Gatzoulis, J. Therrien, Intracardiac thrombus in adults with the Fontan circulation, *Cardiol. Young.* 17 (6) (2007) 646–651.
- [12] P. Monagle, A. Cochrane, R. Roberts, C. Manlhiot, R. Weintraub, B. Szechtman, M. Hughes, M. Andrew, B.W. McCrindle, Fontan Anticoagulation Study G. A multicenter, randomized trial comparing heparin/warfarin and acetylsalicylic acid as primary thromboprophylaxis for 2 years after the Fontan procedure in children, *J. Am. Coll. Cardiol.* 58 (6) (2011) 645–651.