Annals of Cardiology and Cardiovascular Medicine

Case Series
Published: 14 Jan, 2023

Challenging Cases of Failing Fontan Fenestration Occlusion

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Abstract

The Fontan operation is the current surgical procedure to treat single-ventricle congenital heart disease, by splitting the systemic and pulmonary circulations and thus permitting lifespan to adulthood for the majority of pediatric patients. In case of cyanotic patients due to failing Fontan, it is necessary to identify and occasionally treat the existence of hemodynamically significant shunts. We describe two patients with a fenestrated Fontan circulation, who presented to our department with severe desaturation and benefited from transcatheter Fontan fenestration occlusion.

Keywords: Fontan procedure; Fontan fenestration; Fenestration occlusion; Single ventricle

Introduction

The Fontan circulation is a palliative procedure attempting to create a physiologic balance between the circulations, achieved through a series of challenging but widespread surgical procedures during the first years of life. This procedure is applicable typically for patients with single ventricle physiology. The Fontan circulation consists of a single working ventricle pumping blood to the systemic circulation, while venous blood drains *via* surgical connection of the vena cave to the pulmonary arteries [1].

In high and intermediate risk patients, during the procedure, a connection is created between the systemic veins and the right atrium called a fenestration in order to prevent complications from the increased pulmonary resistance and to improve cardiac output at the cost of systemic blood saturation [2]. It has been shown to improve early postoperative outcomes, including decreased duration and quantity of chest tube drainage, shorter duration of mechanical ventilation, and shorter postoperative length of stay. Although a benefit in the early postoperative period, Fontan fenestration has theoretical long-term risks including cyanosis and systemic thromboembolic events due to a persistent right to left shunt combined with an increased risk of thrombus formation due to venous stasis and hypercoagulability. Alternatively, a persistent fenestration may be a benefit because lower central venous pressure may decrease the risk of exercise intolerance, protein losing enteropathy, plastic bronchitis, and brady arrhythmias. The question of if and when to intentionally close a fenestration remains unanswered [3-7].

Apart from Fontan fenestration, in case of cyanotic patients due to failing Fontan it is necessary to identify and occasionally treat the existence of other possible hemodynamically significant shunts (veno-venous, aorto-pulmonary or systemic-pulmonary venous collaterals) [8,9].

In this paper, we present two cyanotic patients with failing Fontan circulation and an open fenestration.

Case Series

Case 1

A 10-year-old male with a history of tricuspid, hypoplastic right ventricle atresia and L-transposition of the great arteries. This patient had the Glenn procedure in 2014 and a fenestrated Fontan completion in 2018. One year later, he was admitted to our department due to episodes of severe cyanosis, paroxysmal nocturnal dyspnea, severe drop in exercise ability and episodes of syncope. In a series of right/left cardiac catheterizations in the span of 2 years, we observed a progressively worse blood oxygen saturation and a significant right to left shunt through the fenestration, located close to the right appendage. After optimal medical treatment, a satisfactory

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Received Date: *19 Dec 2022*
Accepted Date: *10 Jan 2023*
Published Date: *14 Jan 2023*

Citation:
Chatziantoniou A, Despotopoulos S Vagenakis G, Kanakis MA, Apostolopoulou SC, Papagiannis J, et al. Challenging Cases of Failing Fontan Fenestration Occlusion. Ann Cardiol Cardiovasc Med. 2023; 6(1): 1049.*

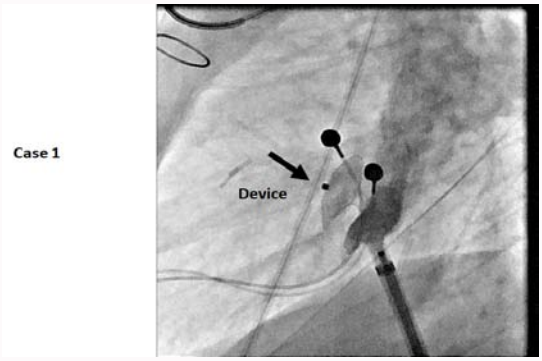

Figure 1: Progressively worse blood oxygen saturation and a significant right to left shunt through the fenestration.

baseline of Fontan circulation mean pulmonary pressure was achieved and thus, the fenestration occlusion was decided after a well-tolerated test occlusion (Figure 1).

Case 2

A 36-year-old female with a history of pulmonary valve atresia and hypoplastic right ventricle. She had a lateral tunnel procedure with a fenestration between the Fontan graft and the right atrium in 1999 and a Fontan fenestration occlusion in 2017 abroad. In 2021 she was admitted in our department with severely reduced exercise capacity and significant desaturation (SpO_2 78% to 80%). Cardiopulmonary treadmill stress test (CPET) was performed with significant reduction in exercise capacity (VO_2max 50% to 55% of predicted for BSA, gender and age). During right heart catheterization, a residual significant shunt cranially to the first occlusion device was revealed-at the base of the right atrial appendage. No other significant collateral (veno-venous or arterio-venous connections) (Figure 2).

In both cases, a temporary balloon occlusion test was performed without increase in the mean systemic venous pressure or decrease in cardiac output (Case 1 mean SVC/Fontan circuit/PA 12 mmHg to 13 mmHg, SpO_2 85% to 97%, Case 2 mean SVC/Fontan circuit/PA 8-9 mmHg to10 mmHg, vessels were identified from the imaging of the pulmonary artery, ascending and descending aorta SpO_2 80% to 92%). After a well-tolerated occlusion test and the fact that no other cause of cyanosis was shown, the decision for permanent fenestration closure was taken.

Given that the fenestration was located next to the subaortic single atrium in the first case, and that there was a second fenestration cranially to the occluded one in the second case, operating in such vulnerable areas was considered as high risk. After careful imaging and sizing balloon, atrial septal defect closure devices with a diameter of 4 mm and 12 mm respectively were implanted and the fenestrations were successfully occluded, without short term complications. In both cases, there was an initial relief of symptoms where the post operative blood oxygen saturation was 97% and 92% respectively.

Discussion

There are trials that have shown that Fontan fenestration even in standard improves early outcomes without affecting long term prognosis [5]. Other series though, suggest that Fontan fenestration brings no long-term benefits without affecting event free survival or Fontan failure [10]. So, the dilemma to fenestrate or not to fenestrate remains.

Of great importance for their prognosis, is the optimal medical treatment of the failing Fontan patients. The pharmacologic therapy includes the careful use of diuretics, inotropic agents, anticoagulants, beta blockers, Angiotensin-Converting Enzyme Inhibitors (ACEIs), Angiotensin Receptor Blockers (ARBs), Mineralocorticoid Receptor Antagonists (MRAs) or digoxin. The benefit of the new era heart failure drugs, such as Angiotensin Receptor-Neprilysin Inhibitors (ARNIs), sodium-Glucose Cotransporter-2 Inhibitors (SGLT2Is), I(f) inhibitors or soluble guanylate cyclase stimulators, is not proved yet, especially when the single ventricle is of right morphology. Moreover, treatment with endothelin-1 receptor antagonists could theoretically improve cardiopulmonary hemodynamics and exercise capacity of those patients [11].

As the patients with Fontan circulation are getting older there are more and more cases where a fenestrated Fontan physiology may prove harmful to the patient and the closure or non-closure of the fenestration is becoming increasingly relevant. Thus, fenestration

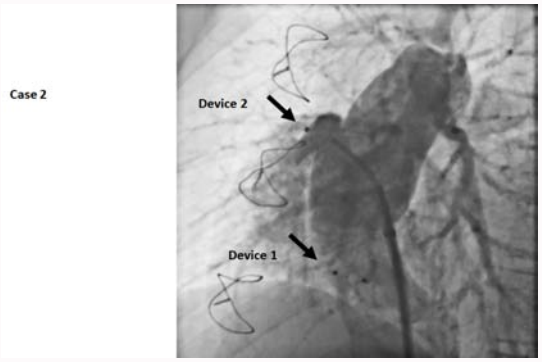

Figure 2: Vessels were identified from the imaging of the pulmonary artery, ascending and descending aorta.

closure is something that needs clear criteria at baseline and at balloon occlusion testing for doctors to follow. So far different criteria have been proposed for closure. According to some investigators fenestration closure is not recommended if cardiac output drops more than 30% with greater than a 4 mmHg increase in mean systemic venous pressure [12]. Other authors suggest that a Fontan fenestration should not be occluded in patients with a high venous pressure greater than 16 mmHg or 20 mmHg. Small series have proposed a few criteria with good results [13] but more research is needed on that field. Cardiac catheterization remains the gold standard for the evaluation of any pathology that can cause decreased cardiac output during temporary occlusion, such as distorted pulmonary arteries or aortopulmonary collaterals [14].

Another matter worthy of discussion is the type of device to be used. Different alternatives have been proposed, such as atrial septal defect closure devices and vascular plug devices [15] in the intracardiac Fontan procedure or covered stent grafts in total cavopulmonary connection [16], highlighting the lack of a dedicated Fontan closure device, especially in anatomically challenging cases as the aforementioned. According to another interesting theory, a more suitable device for fenestration closure could be a bioabsorbable device, since it is possible to re-fenestrate them in the case of a rare situation such as protein-losing enteropathy and ascites after fenestration closure [17]. Despite the limited number of those cases, which make prospective double-blind studies difficult, there is a great need for specialized fenestration occlusion devices produced by medical industry.

What is highlighted in cases as the ones we presented, is that a specialized care heart team, that includes interventional cardiologists, cardiothoracic surgeons, imaging specialists, anesthesiologists, cardiac Cath lab staff, and cardiologists should be the cornerstone of decision making and work in tandem. That would provide not only the benefit of optimal candidate patient choice for fenestration, but also the selection of the optimal area to fenestrate. This multidisciplinary team approach determines patients' treatment plan short term but long term too. For example, the heart team could pre-determine the 'ideal' site of fenestration by the time of Fontan creation in the childhood for potential future occlusion, providing an easy and safe interventional access.

Moreover, due to the proximity of the fenestration to the right atrial appendage there may be benefit in surgical closure during the operation, as performed in other operations in patients with congenital heart disease, though more studies are needed in this area.

Conclusion

To sum up patients with Fontan circulation are very complex and demanding cases. Major role plays the single ventricle morphology (right or left), the existence or not of atrioventricular regurgitation and pulmonary artery resistance. The "correct" management of this group is challenging. In case of cyanotic patients due to failing Fontan, it is necessary to identify and occasionally treat the existence of possible hemodynamically significant shunts and simultaneously estimate the benefit or not of fenestration closure. In the future, thorough research is needed on this field, and multidisciplinary approach in combination with optimal medical device management and evolution is recommended.

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