Case Report

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Obstructed Extracardiac Fontan Conduit in an 18-Year-Old with Dextrocardia: A Unique Solution to A Unique Anatomy

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Abstract

The extracardiac Fontan procedure is the final common pathway for palliation of patients with univentricular physiology. The procedure is typically performed on cardiopulmonary bypass with or without fenestration using an adequate-sized polytetrafluoroethylene graft as the extracardiac conduit. On a few occasions, some patients may develop narrowing of their conduit with or without calcifications, which is often managed by various transcatheter techniques. Rarely, some of these conduits must be replaced surgically. In the current report, we present an alternative to standard surgical replacement of the calcified Fontan conduit in a patient with dextrocardia.

Keywords: Fontan, dextrocardia, off-pump, obstructed fontan, single ventricle

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Introduction

Fontan and Baudet[1] described the original atriopulmonary Fontan in 1971 for a patient with tricuspid atresia. Since then, the procedure has undergone multiple modifications, with the extracardiac conduit Fontan (ECF) becoming more or less the current standard[2]. A variety of conduit materials have been used, ranging from homografts to polytetrafluoroethylene (PTFE), with the latter being preferred due to the lower incidence of calcifications and the ease of transcatheter interventions if needed. In addition, homografts tend to have a more diffuse pattern of calcifications and significant stenosis at either the inferior vena cava or the pulmonary end of the conduit[3]. Despite the lower rate of transcatheter interventions on PTFE conduits, a small number of patients may require complete surgical replacement of the conduit, which is often performed using cardiopulmonary bypass (CPB) and is associated with higher risks due to the marginal nature of many of these patients at the time they present with obstructed Fontan conduit. We present the case of an 18-year-old patient with dextrocardia and univentricular physiology who developed diffuse calcifications and obstruction in his Fontan PTFE conduit that was not amenable to transcatheter therapy and underwent repeat surgery with a novel solution to his obstructed conduit.

Case Presentation

Patient Presentation

An 18-year-old male with known dextrocardia and univentricular physiology secondary to unbalanced complete atrioventricular septal defect and pulmonary atresia with transposition of the great arteries underwent all three-stage palliation for his SV, with his last procedure being a non-fenestrated ECF (16 mm graft). He presented with exertional fatigue and deranged liver function. Computed tomography (CT) scan, and cardiac catheterization showed diffusely calcified conduit with stenosis of the inferior vena caval/conduit anastomotic site (Figure 1A and B). Because of the diffusely calcified nature of the conduit combined with anastomotic stenosis, the decision was made to replace the conduit surgically.

Surgical Technique

Repeat median sternotomy was performed, and initial dissection revealed a diffusely calcified conduit with obvious narrowing at the inferior vena cava anastomotic site (Figure 2). The conduit was quite stuck to the left phrenic nerve and was connected directly opposite to the left superior vena cavopulmonary anastomotic site. We decided to create an alternate conduit pathway to the right pulmonary artery (RPA) and perform this without the use

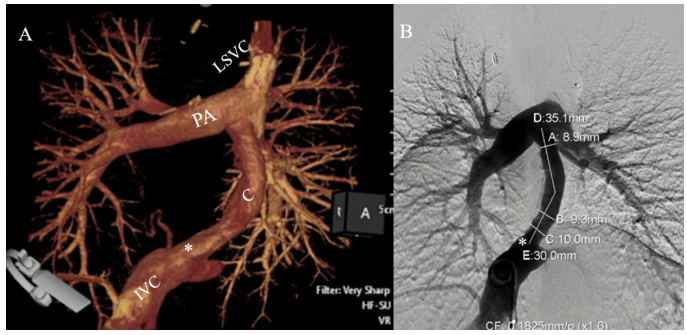

Figure 1. Preoperative computed tomography scan **(A)**, and cardiac catheterization **(B)** showing diffuse calcifications in the extracardiac Fontan conduit and significant narrowing at the inferior vena caval/conduit anastomotic site (asterisk). Notice the significant reduction (almost 50%) in the actual conduit diameter from the original 16 mm as measured on cardiac catheterization which is attributed to most likely to an ongoing intimal proliferation
IVC: Inferior vena cava, C: Conduit, LSVC: Left superior vena cava, PA: Pulmonary artery

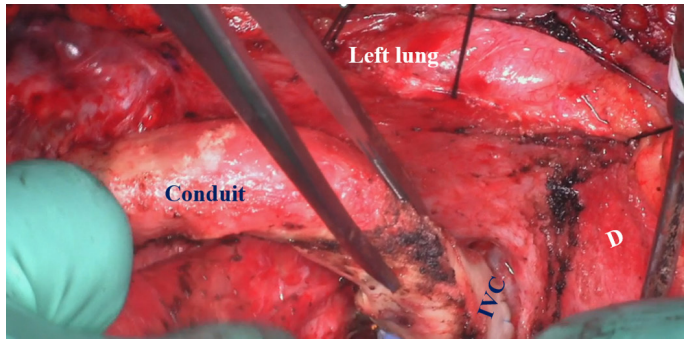

Figure 2. Intraoperative photo after a repeat median sternotomy and initial dissection showing the previously calcified extracardiac conduit and the narrowed inferior vena caval/conduit anastomosis
IVC: Inferior vena cava, D: Diaphragm

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of CPB. Thorough dissection of the heart was performed, and the RPA was completely mobilized and isolated.

We dissected the inferior vena cava well below the anastomotic site. It was clear that a new conduit could be placed behind the heart to the RPA. Heparin was administered systemically, and a side-biting clamp was applied on the inferior vena cava/conduit connection without completely occluding the conduit to maintain hemodynamics. A 20-mm PTFE externally reinforced graft was then connected in an end-to-side fashion to the inferior vena cava/conduit using a 5/0 polypropylene suture (Figure 3A). The anastomosis was then de-aired, and the graft was brought behind the ventricle to the RPA, where it was connected in a similar fashion after adjusting its length (Figure 3B). The conduit position was satisfactory (Figure 4), and the rest of the procedure and chest closure were performed in the standard fashion.

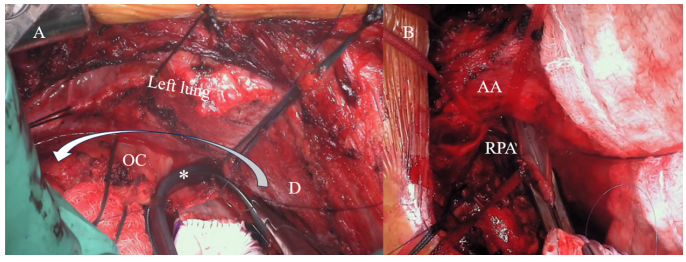

Figure 3. Intraoperative photos showing **(A)** the anastomosis of the new conduit (ringed PTFE) to the inferior vena caval/previous conduit with a side-biting clamp (white asterisk), partially occluding the old conduit to ensure hemodynamic stability during performance of the anastomosis by maintaining venous return to the lung (white arrow), and **(B)** the new conduit to the right pulmonary artery anastomosis is being completed
OC: Old conduit; D: Diaphragm; RPA: Right pulmonary artery; AA: Ascending aorta

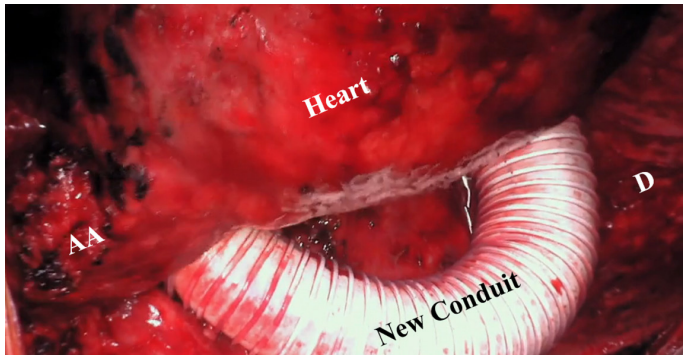

Figure 4. Intraoperative photo showing the completion of the new conduit which is placed to the right side behind the heart
D: Diaphragm, AA: Ascending aorta

Outcome

The postoperative course was uneventful. He received no transfusion, was extubated in the operating room, and was discharged 9 days later. Pre-discharge echocardiogram and CT scan showed a widely patent new Fontan conduit (Figure 5A, B) and cardiac function remained unchanged. He continued to perform well during his 2-year follow-up with no concerns related to either Fontan conduit.

Discussion

ECF continues to be the preferred final palliation for patients with univentricular physiology. The most commonly used conduit is the PTFE conduit because of its favorable characteristics. However, some patients continued to require either transcatheter or less commonly repeat operation because of the development of long-term anastomotic stenosis and/or conduit obstruction.

The majority of these patients, when present with an obstructed conduit, have deranged multisystem organ function, especially the liver and/or kidney. Therefore, combined organ transplantation has become the preferred treatment option due to the risks involved in reoperating on these patients. With the obvious limitations of transplantation, the reality of the need for repeat surgery

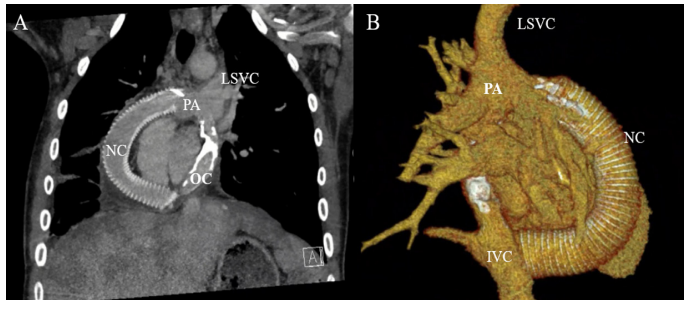

Figure 5. Follow-up computed tomography scan: **(A)** coronal cut, and **(B)** 3-dimentional reconstruction (posterior view) showing widely patent new conduit connections and the diffusely calcified old conduit *in situ*
NC: New conduit, OC: Old conduit, PA: Pulmonary artery, LSVC: Left superior vena cava, IVC: Inferior vena cava

cannot be ignored. Findings alternate surgical solutions that can minimize the need for blood transfusions and the ongoing development of immunological sensitization, which may compromise the patient's opportunity for future transplantation, in addition to the other known drawbacks of CPB[4].

In the current report, we present a unique solution to an obstructed ECF conduit without the use of CPB. The advantages of this solution are clear; the patient received no transfusion in the perioperative period, avoidance of CPB minimized/avoided further derangements in other organ systems, and allowed maintenance of the same Fontan hemodynamics. Placing the new conduit away from the left superior vena cavopulmonary connection optimizes hemodynamics, minimizes energy losses[5], and allows better distribution of venous return, particularly the hepatic venous blood, to both lungs, as shown in many previous publications[6]. Leaving the old conduit *in situ* avoided injury to the left phrenic nerve and simplified the procedure.

Conclusion

Although reoperation on obstructed Fontan conduits is rare, apart from the need for transplantation, finding alternate solutions to avoid the use of CPB should be strongly considered. Off-pump ECF is feasible in many patients, even those with positional anomalies, but it requires adequate planning and unique surgical strategies.

Ethics

Informed Consent: Permission was obtained from the patient to publish this case report.

Authorship Contributions

Surgical and Medical Practices: Said SM, Concept: Mashadi AH, Said SM, Design: Mashadi AH, Said SM, Data Collection and/or Processing: Mashadi AH, Said SM, Analysis and/or Interpretation: Mashadi AH, Said SM, Literature Search: Mashadi AH, Said SM, Writing: Mashadi AH, Said SM.

Conflict of Interest: The authors declare no conflicts of interest concerning the authorship or publication of this article.

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