

CONGENITAL HEART DISEASE

CASE REPORT: CLINICAL CASE

Fontan Conversion in an Adult With Hypoplastic Left Heart Syndrome

A 38-Year-Old Norwood Success



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ABSTRACT

A 38-year-old man with a functional single ventricle secondary to hypoplastic left heart syndrome presented with exertional fatigue. His last palliation was an intra-atrial conduit Fontan procedure. Comprehensive evaluation showed elevated liver enzyme values and a small, calcified conduit. Successful conversion to a nonfenestrated extracardiac conduit Fontan was performed with normalization of his liver enzyme values. (JACC Case Rep. 2024;29:102495)
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HISTORY OF PRESENTATION

A 38-year-old man who was born with a functional single ventricle secondary to hypoplastic left heart syndrome (HLHS) presented with exertional fatigue

TAKE-HOME MESSAGES

- This case highlights the importance of close and lifelong follow-up of functional single ventricle patients.
- Exertional symptoms in patients with previous Fontan palliation can be multifactorial.
- Replacement of small Fontan conduits and Fontan conversion may be considered to delay the progression of liver dysfunction and improve Fontan hemodynamics which may delay the need for transplantation.

and shortness of breath. After his Fontan palliation, he did quite well over his late childhood and early adolescent years. However, later he received a diagnosis of exercise intolerance, attaining only 68% of his target heart rate on stress testing. His evaluation at that time showed a widely patent Fontan pathway but significant venovenous collateral vessels (VVCs) that correlated with a drop in his oxygen saturation to 91%. Additional testing showed evidence of cardiac cirrhosis and long-term liver enzyme elevation. His medical regimen included angiotensin-converting enzyme-inhibitors and antiplatelet therapy, and he was treated with antibiotics for recurrent pneumonia. He also underwent transcatheter device closure of VVCs.

PAST MEDICAL/SURGICAL HISTORY

The patient was born with HLHS secondary to aortic and mitral atresia. He underwent all 3 stages of

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**ABBREVIATIONS
AND ACRONYMS****ECF** = extracardiac Fontan**HLHS** = hypoplastic left heart syndrome**VVC** = venovenous collateral (vessels)

palliation in infancy, with a final completion intra-atrial conduit Fontan procedure using a 12-mm graft. Other associated medical problems included Tourette syndrome, cardiac cirrhosis, anxiety/depression, previous stent placement for recurrent coarctation, and previous transcatheter closure of VVCs.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included heart failure, Fontan failure, obstructed Fontan conduit, arrhythmias/chronotropic insufficiency, pulmonary arteriovenous malformations, and VVCs.

INVESTIGATIONS

Laboratory tests showed elevated liver enzymes. An electrocardiogram showed sinus rhythm, but on Holter monitoring, there were a few beats of supra-ventricular tachyarrhythmias. A transthoracic echocardiogram revealed trivial systemic tricuspid valve regurgitation and mild reduction in systemic right ventricular function.

There was a moderate obstructive pattern on his pulmonary function tests (Figure 1), in addition to mild hypoxemia on room air with a resting oxygen saturation of 91% to 94%. This was maintained during his 6-minute walk test.

Cross-sectional imaging showed evidence of liver cirrhosis with hepatosplenomegaly, but no biopsy was performed. It was noticed that the previous Fontan conduit was calcified.

Hemodynamic data from cardiac catheterization showed Fontan pressure of 14 to 15 mm Hg (Figure 2), as well as a small, calcified intra-atrial Fontan conduit measuring 10.5 mm in its smallest dimensions (Figure 3, Video 1). Attempts at conduit dilation/stenting were not successful because of the calcification.

MANAGEMENT

After extensive multidisciplinary team discussion, the decision was made to proceed with Fontan conversion,¹ given the patient's small size conduit and the difficulty of transcatheter stenting of such calcified small conduits.²

A fourth sternotomy was performed. Normothermic cardiopulmonary bypass was initiated through aortic and bicaval cannulation, and cardioplegic arrest was achieved with del Nido antegrade cardioplegia (Video 2). The previous Fontan conduit was completely calcified (Figure 4A), and it was excised in its entirety (Figure 4B). After closure of the common atrial incision, a new 20-mm polytetrafluoroethylene conduit was then used to complete the

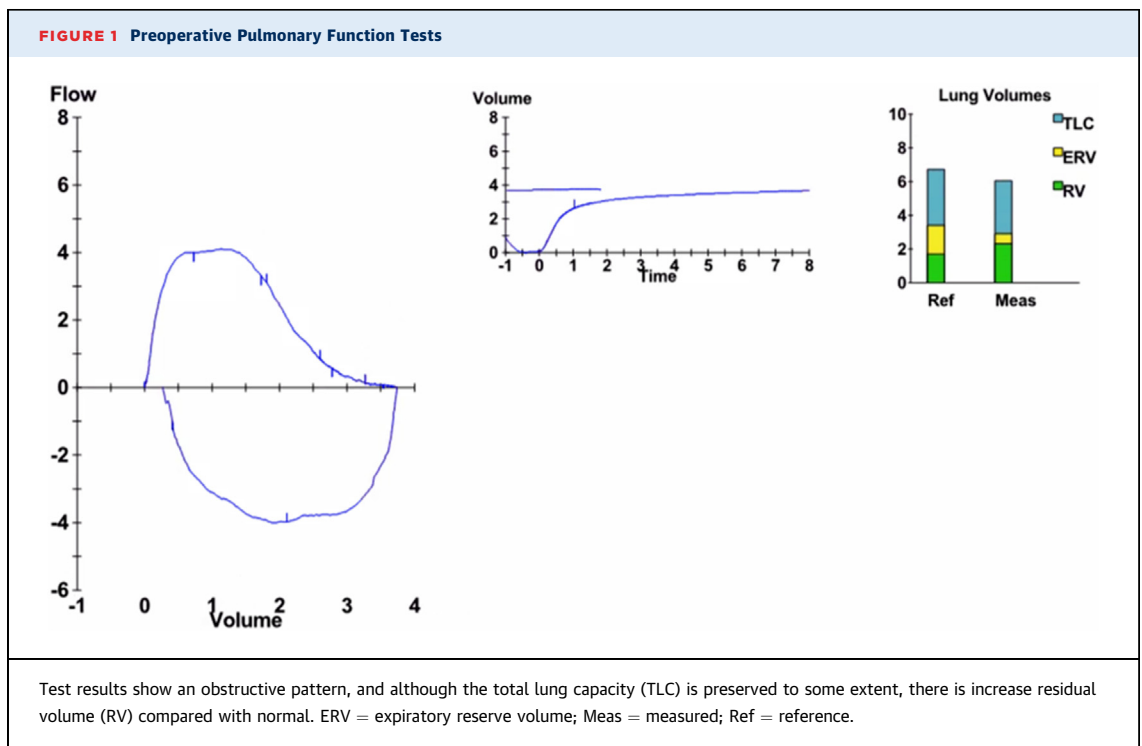
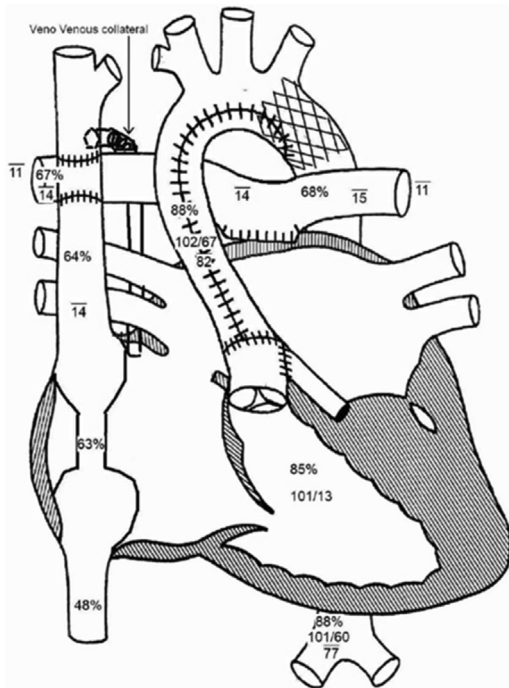
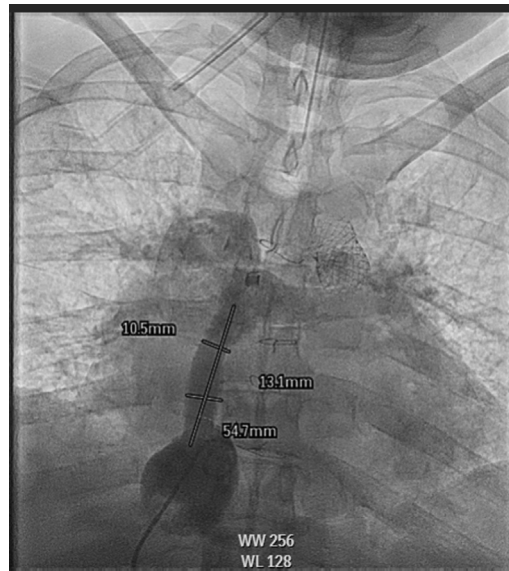


FIGURE 2 Hemodynamic Data From Preoperative Cardiac Catheterization



Findings show slightly elevated systemic ventricular end-diastolic pressure, satisfactory pulmonary artery pressure, and no systemic outflow tract obstruction.

FIGURE 3 Preoperative Cardiac Catheterization



Findings show a small, calcified, extracardiac Fontan conduit with a minimal diameter of 10.5 mm. A dilated inferior vena cava is seen.

total cavopulmonary connection in an extracardiac fashion (Figures 4C and 4D). No fenestration was needed because of the patient's good preoperative hemodynamics. The patient tolerated the procedure well. He was extubated in the operating room and was discharged 7 days later. His liver enzyme levels normalized before his hospital discharge.

FOLLOW-UP

The patient continued to be followed up, and his repeat cardiac catheterization showed a widely patent extracardiac Fontan (ECF) conduit and satisfactory hemodynamics (Video 3). His systemic ventricular function remained stable, with no reported arrhythmias. At 1 year postoperatively, he remained off the list for cardiac/liver transplantation.

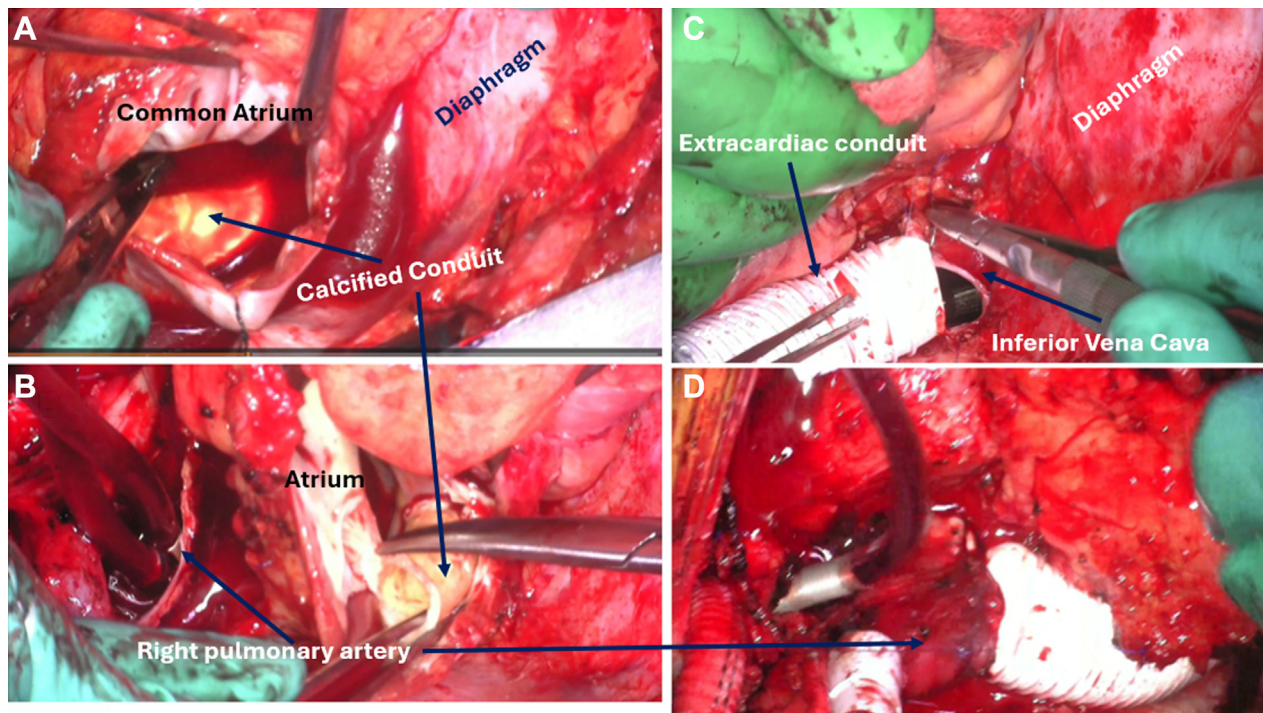
DISCUSSION

To our knowledge, this is one of the oldest transplant-free surviving patients with HLHS who had a Norwood procedure as a neonate and completed all

his surgical palliation, ending with the Fontan procedure. ECF is the most common variant of the Fontan procedure currently performed, and the recommended optimum conduit size is 18 to 20 mm, on the basis of studies of flow dynamics in various conduits sizes.³ Although replacing old Fontan conduits is not a frequently performed procedure, it is still a consideration in some patients to delay or avoid the need for transplantation, especially in the presence of-somewhat preserved systemic ventricular function and acceptable Fontan hemodynamics.

Rigorous follow-up of Fontan-treated patients is mandatory to detect abnormalities in the Fontan circulation that may require immediate transcatheter/surgical interventions before the development of irreversible end-organ damage.⁴

One could argue to consider the current patient for heart transplantation or combined heart/liver transplantation. However, in the presence of satisfactory hemodynamics and an identifiable anatomic problem, a standard surgical option with acceptable risk, if present, should be considered. In a patient who is otherwise functioning and nonhospitalized, the waiting list for transplantation will be quite long, and changing the Fontan conduit in the current case was the immediately feasible option to prevent further liver deterioration. Considering the natural history of

FIGURE 4 Intraoperative Findings

Intraoperative photographs showing (A) the calcified small intra-atrial conduit through the opened common atrium, (B) excision of the conduit from its atrial connection, (C) connection of the new extracardiac Fontan conduit to the inferior vena cava, and (D) the final completed extracardiac Fontan connection.

Fontan physiology, the current patient may eventually require transplantation, and this will be determined during his future follow-up.

CONCLUSIONS

Calcified Fontan conduits can be safely replaced, especially if the conduits are small relative to the patient's size and in the absence of reliable transcatheter options. The short-term outcomes are excellent in carefully selected patients. This procedure may increase freedom from heart transplantation and may prevent further or rapid deterioration in liver function.

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KEY WORDS Fontan, Fontan conversion, hypoplastic left heart syndrome, Norwood, single ventricle

APPENDIX For supplemental videos, please see the online version of this paper.