Upsizing the extracardiac Fontan conduit—the fourth staged procedure for the single-ventricle palliation?

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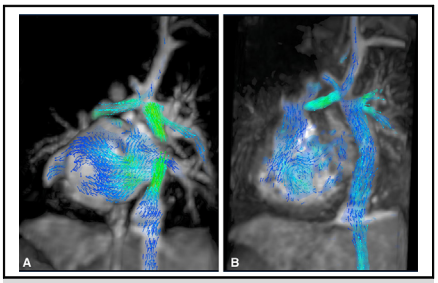

Cardiac MRI with 4-D flow pre- and post-Fontan conduit-upsize procedure.

Patients with functional single-ventricle hearts are palliated with a series of surgical procedures culminating in the Fontan. The most widely performed Fontan technique today is the extracardiac conduit (ECC), with some surgeons still performing the lateral tunnel. In those receiving the ECC, the lack of growth of the conduit is a concern as patients grow into adulthood. We present a case series of patients with ECC with Fontan failure and small conduits who underwent surgical augmentation of their circulation with a Fontan-upsize procedure.

CENTRAL MESSAGE

Extracardiac conduits placed in childhood may be inadequate for adults with the Fontan circulation. We present a case series of patients with Fontan failure and extracardiac conduit-upsize procedure.

METHODS

We conducted a retrospective review of patients receiving Fontan-upsize procedures at our institution from 2021 to 2023. The institutional review board approved the study protocol and publication of data (institutional review board: PR00015566, approved July 21, 2021). Patient consent for the study and publication of the study data was waived due to minimal risk.

RESULTS

We performed 6 Fontan-upsize procedures. Clinical and procedural characteristics are shown in Table 1. The median age was 17.5 years. Five patients had an ECC and one an intra-extracardiac Fontan, with conduit diameter sizes ranging from 14 to 16 mm. The conduit size was unknown in one patient but measured <14 mm by angiography. Preoperative median Fontan pressure was 18.5 mm Hg (interquartile range [IQR], 17.25-19 mm Hg). Preoperative hemodynamic and qualitative ventricular function are shown in Tables E1 and E2. Four patients exhibited significant hepatic fibrosis. All were anticoagulated with aspirin, except for one with history of recurrent gastrointestinal bleeding.

The main surgical indication for patients was Fontan failure with elevated Fontan pressure. Most patients underwent the Fontan-upsize procedure with other concomitant cardiac procedures. The lack of existing evidence for the potential benefits of the Fontan-upsize procedure was well explained to the families and patients as part of informed consent. All underwent Fontan upsize with either Gore-Tex or exGraft (PECA Labs) with conduit sizes ranging from 18 to 24 mm. exGraft conduits were favored because of their potential to be progressively dilated up to 200% of their diameter for future interventions. All upsized conduits were nonfenestrated except for one patient, who underwent a concurrent pulmonary arterioplasty. The extracted conduits were notable for their relatively small sizes, but no remarkable distortion or thrombi were seen. Figure 1 is a cardiac magnetic resonance imaging representation of the Fontan conduit narrowing preoperatively and wide patency of the conduit postoperatively (patient 4).

Postoperative clinical data are shown in Tables E2 and E3. There were no notable difficulties or complications during surgery. Recovery of the patients was uneventful with a median intensive care unit and hospital stay of 4.5 days (IQR, 4, 9.5) and 12.5 days (IQR, 9, 19.7), respectively. Two patients had transient tachyarrhythmia during the postoperative period. One patient had bleeding intraoperatively and thus underwent delayed sternal closure on postoperative day 1. All patients

TABLE 1. Clinical and procedural characteristics of patients with Fontan circulation undergoing conduit-upsize procedure

Patient no.	Age at time of Fontan upsize, y	Sex	Weight, kg	Anatomic diagnosis	Age at first Fontan procedure, y	Fontan (size/type)	Surgical indication	Fontan revision	Concomitant surgical procedure
1	20	F	57.7	Heterotaxy, situs inversus, mitral atresia, pulmonary atresia, hypoplastic left ventricle, status post bioprosthetic tricuspid valve	4	14-mm extracardiac, nonfenestrated	Fontan failure Severe bioprosthetic valve regurgitation	18-mm extracardiac with Gore-Tex, non-fenestrated	33-mm Perimount Mangal Mitral Ease Valve Implantation
2	9	M	33.5	Double-outlet right ventricle, unbalanced AVC, hypoplastic left ventricle, pulmonary atresia, bilateral SVC	2.8	16-mm intra-extracardiac with ring supported conduit 4-mm fenestration	Fontan failure Severe AV regurgitation Sinus-node dysfunction	18-mm extracardiac with Gore-Tex, fenestrated	Tricuspid valve repair Pulmonary arterioplasty Epicardial pacemaker
3	15	M	84	Heterotaxy, HLHS	1.6	14-mm intra-extracardiac 4-mm fenestration	Fontan failure Grade 3-4 hepatic fibrosis Sinus-node dysfunction	20-mm intra-extracardiac with exGraft non-fenestrated	Atrial pacing lead implantation
4	20	M	47	Heterotaxy, DORV, Pulmonary atresia, mitral atresia, hypoplastic LV	3.5	14-mm extracardiac 4-mm fenestration	Fontan failure Severe hepatic fibrosis/ recurrent GI bleeding due to portal hypertension Severe tricuspid valve regurgitation	20-mm extracardiac with exGraft, non-fenestrated	Tricuspid valvuloplasty
5	15	M	85.5	Heterotaxy, unbalanced AVC, hypoplastic LV	1.5	14-mm intra-extracardiac, 4-mm fenestration	Fontan failure Grade 3-4 hepatic fibrosis	24-mm extracardiac with Gore-Tex, nonfenestrated	
6	29	M	111	L-TGA, DILV, aortic coarctation	3	Size unknown (measured <14 mm on by catheterization) extracardiac	Fontan failure/ conduit stenosis with decompressing veins Liver cirrhosis EOL pacemaker + dysfunctional ventricular leads	24-mm extracardiac with exGraft nonfenestrated	Epicardial pacemaker revision

F, Female; *M*, male; *AVC*, atrioventricular canal; *SVC*, superior vena cava; *AV*, atrioventricular; *HLHS*, hypoplastic left heart syndrome; *DORV*, double-outlet right ventricle; *LV*, left ventricle; *GI*, gastrointestinal; *L-TGA*, L-transposition of the great arteries; *DILV*, double-inlet left ventricle; *EOL*, end of life.

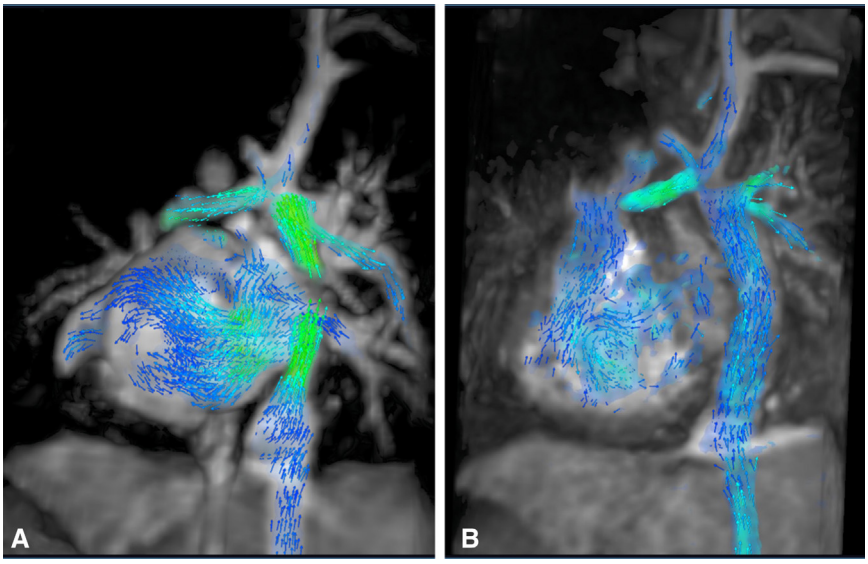

FIGURE 1. Cardiac MRI of the extracardiac Fontan pathway pre- and postconduit upsize procedure. Cardiac MRI with time-resolved 3-dimensional velocity encoding by phase contrast (4D flow) of patient 4 in the sagittal view. A, Preoperative MRI demonstrating a small extracardiac conduit Fontan, measuring 10 × 13 mm at its narrowest dimension, with discrete flow acceleration at the Fontan anastomosis with the inferior vena cava. There is artifact in the middle of the Fontan conduit secondary to fenestration device closure. B, Cardiac MRI 1-month post-Fontan revision, demonstrating widely patent Fontan conduit with low flow velocity. *MRI*, Magnetic resonance imaging.

were discharged home and were asymptomatic after a median time of 8.5 months (3-26 months). Most notably, patient 4, who underwent tricuspid valve repair and Fontan upsize from 14- to 20-mm conduit, had years' long history of liver fibrosis, portal hypertension, and recurrent gastrointestinal bleeding but had no recurrence of bleeding within 6 months. Postoperative hemodynamic data were not available in this short-term follow-up.

DISCUSSION

After 3 decades, there still does not seem to be much difference in outcomes between the lateral tunnel and the ECC.[1] However, the lack of growth of the ECC conduit is worrisome as patients grow. Furthermore, cross-sectional studies of the ECC show as much as 25% reduction over time.[2] Many are doubting that the sizes of 14- and 16-mm diameter will be large enough for adults with a Fontan circulation. There is growing concern that even patients with conduits of 18 and 20 mm may be exposed to a greater risk of liver fibrosis, and it has been suggested that optimal exercise capacity would require a conduit size comprising between 22 and 26 mm in adults.[3,4] Interventional catheterization procedures have been described to dilate obstructed ECC,[5] but catheterization intervention is often limited by the size of the ECC because these conduits tend to calcify. Over the years, our center has implanted at least 185 16-mm ring-reinforced conduits and 25 14-mm ECC. Many of these ECCs, particularly the ring-reinforced conduits, would not be amenable to noninvasive dilation and will require surgery to upsize. So far, we have successfully upsized the Fontan conduits in 6 of our patients and demonstrated the relative safety of this procedure. Longitudinal follow-up is critical to evaluate the long-term impact. We believe that today, the Fontan procedure performed in early childhood can no longer be considered as a final palliation and that many of our patients will require additional procedures.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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TABLE E1. Preoperative hemodynamic data

Patient no.	Fontan pressure, mm Hg	Cardiac index (Qs), L/m^2/min	Qp:Qs	Ventricular end-diastolic pressure, mm Hg	Pulmonary vascular resistance, Woods unit m^2
1	19	2.4	0.7:1	6	2.8
2	19	3.3	0.9:1	8	2.9
3	22	5.8	0.6:1	14	1.9
4	17	3.4	1:1	10	1.5
5	15	3.3	1:1	7	1.2
6	18	1.7	Unable to calculate	12	Unable to calculate
Median (IQR)	18.5 (17.25-19)	3.3 (2.63-3.38)	0.9:1 (0.7:111)	9 (7.25-11.5)	1.9 (1.5-2.8)

Qp:Qs, Pulmonary to systemic flow ratio; *IQR*, interquartile range.

TABLE E2. Qualitative systemic ventricular function and systemic saturation pre- and post-Fontan-upsize procedure

Patient no.	Preoperative ventricular function (by echocardiography)	Postoperative ventricular function (by echocardiography)	Preoperative systemic saturation	Postoperative systemic saturation
1	Lower limit of normal RV function	Mild RV dysfunction	90%	89%*
2	Normal RV function	Normal RV function	95%	93%
3	Normal RV function	Normal RV function	89%	90%
4	Normal RV function	Normal RV function	95%	96%
5	Normal RV function	Normal RV function	95%	96%
6	Moderate LV dysfunction	Moderate LV dysfunction	98%	92%

RV, Right ventricle; *LV*, left ventricle. *With presence of venovenous collaterals and pulmonary arteriovenous malformations.

TABLE E3. Operative and postoperative characteristics of patients undergoing the Fontan-upsize procedure

Patient no.	Cardiopulmonary bypass, Min	Aortic crossclamp, Min	ICU LOS, days	Hospital LOS, days	Mechanical ventilation, days	Chest tube duration, days	Complications
1	241	90	4	9	1	6	None
2	304	89	13	16	1	15	None
3	203	63	4	8	1	6	None
4	184	96	3	30	0.5	6	GI bleeding*/JET
5	105	65	5	9	1	4	JET/SVT
6	234	None	11	21	1	6	Bleeding, delayed sternal closure POD 1
Median (IQR)	218 (188, 239)	89 (65, 90)	4.5 (4, 9.5)	12.5 (9, 19.7)	1 (1, 1)	6 (6, 6)	

ICU, Intensive care unit; *LOS*, length of stay; *GI*, gastrointestinal; *JET*, junctional ectopic tachycardia; *SVT*, supraventricular tachycardia; *POD*, postoperative day; *IQR*, interquartile range. *Patient with history of coagulation disorder and history of previous GI bleeding.