Biventricular Fontan conversion in criss-cross, superior-inferior ventricles using arterial switch and extra-anatomical left ventricle to aorta prosthesis

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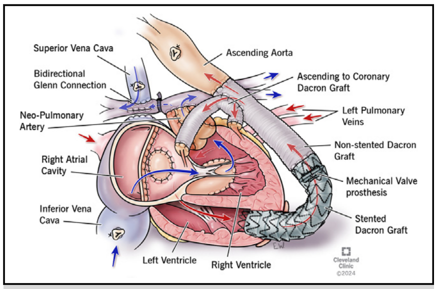


Anatomy of biventricular Fontan conversion using extra-anatomical prosthesis.

CENTRAL MESSAGE

We describe a biventricular conversion in an adult with double outlet right ventricle, superior-inferior ventricles, criss-cross connection status post Fontan presenting with subaortic obstruction.

Video clip is available online.

To view the AATS Annual Meeting Webcast, see the URL next to the webcast thumbnail.

We present a 22-year-old female patient born with double-outlet right ventricle, criss-cross orientation of the atrioventricular valves with superior inferior relation of ventricles, right-handed ventricular topology, large perimembranous inlet ventricular septal defect with a straddling mitral valve, and bilateral superior vena cavae (but thrombosed left superior vena cava from previous interventions). The patient had previous pulmonary artery banding, atrial septectomy, bidirectional cavopulmonary anastomosis, and lateral tunnel nonfenestrated Fontan (Figure 1 and Figure 2, *A*).

She developed venovenous collaterals requiring multiple coil embolization procedures, as well as progressive subaortic right ventricular outflow obstruction (peak gradient 90 mm Hg) with liver and kidney dysfunction. Her preoperative diagnostic catheter showed a cardiac index of 2.4 L/min/m^2 with a Qp:Qs of 1, normal Fontan pressure of 9 mm Hg, normal systemic right ventricular end-diastolic pressure, normal transpulmonary gradient with a calculated pulmonary vascular resistance index of 1.2 WU /m^2, and no significant aortopulmonary or venovenous collaterals.

The patient felt strongly that avoiding a transplant at her time would be the preferred goal; therefore, using shared decision-making between the patient, the patient's family, the patient's cardiologist, and the surgeon, it was decided to proceed with a biventricular repair. After advanced imaging, it was evident that the right ventricle was of reasonable size to be incorporated into the subpulmonary circulation. The morphologic right ventricle was moderately hypoplastic with normal systolic function. The tricuspid valve (TV) had no stenosis and trivial regurgitation with an annulus of 2.42 cm. The morphologic left ventricle (LV) was mildly dilated with normal systolic function. There was a straddling mitral valve with no stenosis and mild regurgitation. The option of routing the full systemic venous return through the posterior inlet ventricular septal defect (VSD) was discussed but the right ventricle and TV were deemed to be too small to accommodate the baffle and the full systemic venous flow. Therefore, we proceeded with relief of the right ventricular outlet obstruction and performed a definitive LV-based one and half biventricular conversion.

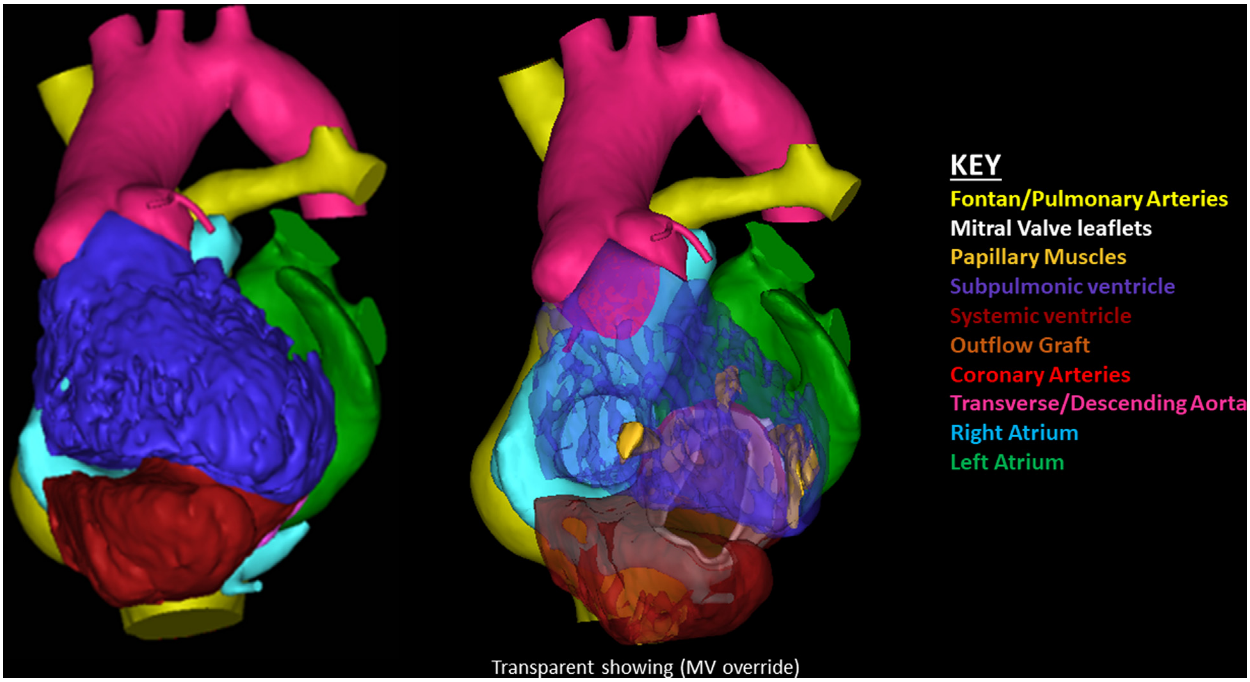


FIGURE 1. Preoperative anatomy showing double-outlet right ventricle, criss-cross orientation of the atrioventricular valves with superior inferior−oriented ventricles, large inlet ventricular septal defect with a straddling mitral valve, and lateral tunnel Fontan.

The institutional review board or equivalent ethics committee of the Cleveland Clinic did not approve this study because it is a case report. The subject provided informed written consent for the publication of the entire study.

OPERATIVE STEPS

The intention of the procedure was to create an LV-based biventricular conversion. Therefore, on the back table, before beginning the surgery, a 28-mm non-bare metal stent conduit (RelayPro NBS; Terumo Aortic) was prepared by suturing a 23-mm mechanical On-X valve (Artivion) into the stented end of the conduit. On the opposite end, a skirt of bovine pericardium was sutured 1 cm from the flaring edge to allow for apical insertion in the LV. A 28-mm Hemashield graft (Getinge) was sutured to the end of the stented graft to provide additional length (Figure 2, *B*). All sutures were reinforced with bioglue and a felted cuff was created for later use to buttress the apical cannulation.

The procedure was carried out on cardiopulmonary bypass with aortic crossclamping and intermittent blood cardioplegia (Video 1). The lateral tunnel was excised widely (Figure 2, *C*). Through the TV, subaortic muscle bundles were resected. Because of the smaller diameter of the TV making access to all muscle bundles difficult (Figure 2, *D*) an aortotomy was created and additional resection performed. On evaluation of the straddling mitral valve, there was a papillary muscle that landed on the crest of the septum before reaching the free wall of the right ventricle. The attachment to the free wall was divided leaving the attachment to the crest of the septum (Figure 2, *E*) The VSD was closed with a bovine pericardial patch and a running 4-0 polypropylene suture both through the TV and the aortotomy (Figure 2, *F*).

To construct the LV apical to aortic connection, the apex was elevated with sponges and a 20-mm coring circular knife was used to create a hole in the apex of the LV. Twelve circumferential full-thickness sutures were placed in the apex using the circular felt ring. The previously prepared conduit device was oriented to keep the S-shaped (kink prevention) wire outward and inserted into the apex of the LV using the bovine pericardial skirt to secure the device to the apex. After securing the 12 Ti-Cron sutures (Medtronic), a 4-0 polypropylene suture was run around the sewing ring to ensure a tight seal between the graft and the left ventricle. The heart was placed back into the chest and the graft was curved back around keeping the S wire built into the graft on the outer aspect of the graft to avoid kinking (Figure 2, *G*).

Coronary buttons were harvested, and the aortic defects were patched with bovine pericardium. The aorta was closed at the sinotubular junction. An 8-mm Hemashield graft was sutured end-to-end to both coronary buttons (Cabrol technique). The aorta was transected high and anastomosed to the left pulmonary artery.

To reconstruct the LV outflow, the distal end of the conduit graft was anastomosed to the distal ascending aorta

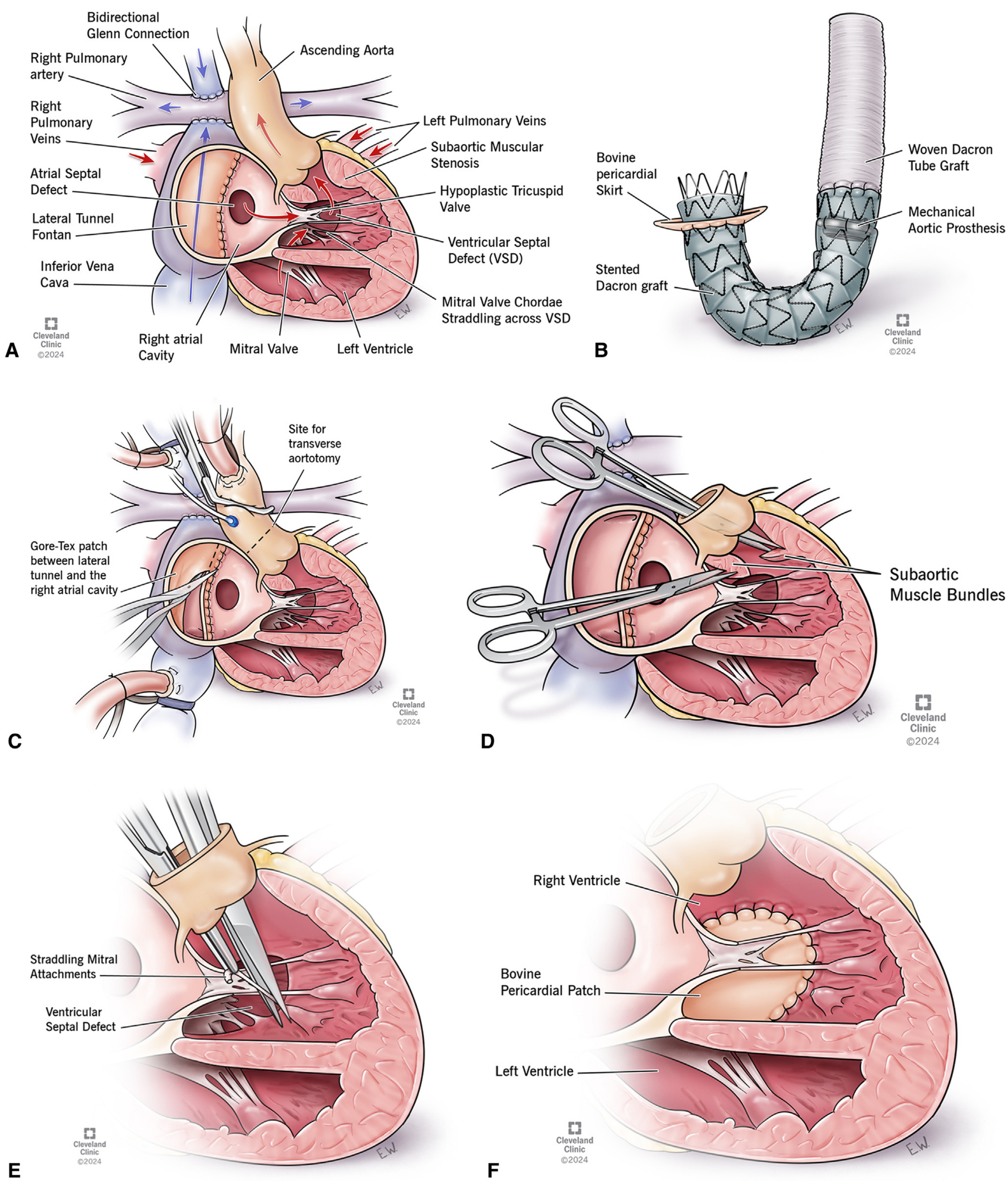


FIGURE 2. A, Initial anatomy showing double-outlet right ventricle, criss-cross orientation of the atrioventricular valves with superior inferior oriented ventricles, right-handed ventricular topology, large perimembranous inlet ventricular septal defect with a straddling mitral valve, status post-lateral tunnel Fontan. B, A 28-mm RelayPro non-bare metal stent conduit was prepared by suturing a 23-mm mechanical On-X valve into the stented end of the conduit. On the opposite end, a skirt of bovine pericardium was sutured 1 cm from the flaring edge. A 28-mm Hemashield graft was sutured to the end of the stented graft. C, Takedown of lateral tunnel. D, Resection of subaortic muscle bundles. E, Division of attachment of the mitral papillary muscle to the free wall of the right ventricle. F, Closure of ventricular septal defect with a bovine pericardial patch. G, Insertion of conduit into the left ventricular apex. The conduit device

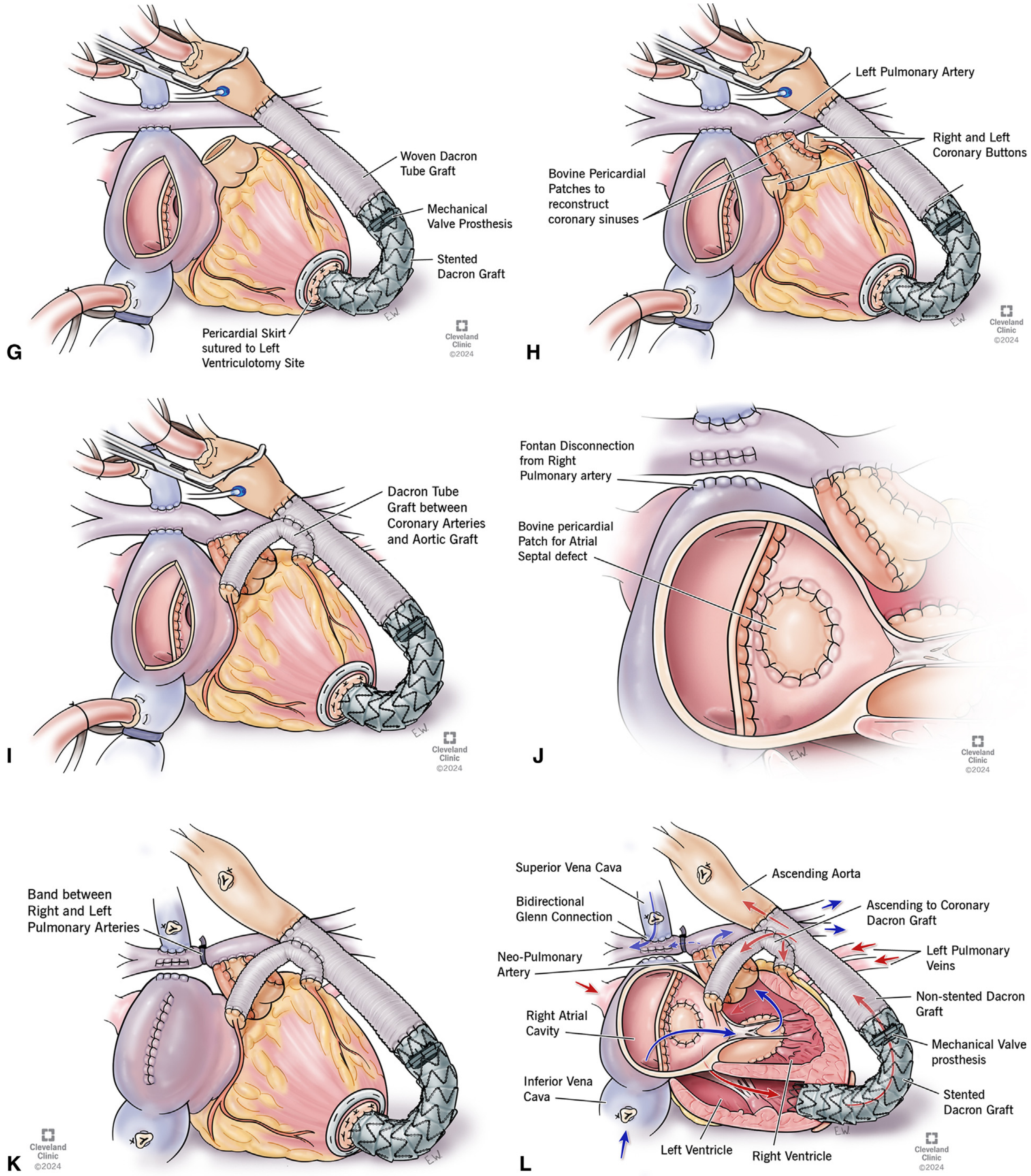
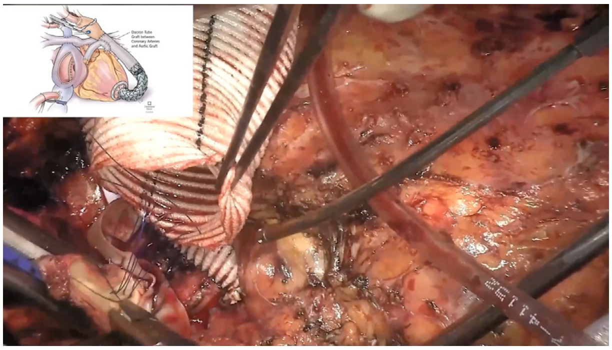


FIGURE 2. (*continued*).

was oriented to keep the S-shaped (kink prevention) built-in wire outward and inserted into the apex of the left ventricle using the bovine pericardial skirt to secure the device to the apex. H, Harvest of coronary buttons and reconstruction of aortic root with bovine pericardium. I, Cabral technique of coronary button re-implantation using an 8-mm Hemashield graft sutured end-to-end to both coronary buttons. Anastomosis of the proximal aorta to the left pulmonary artery. Anastomosis of the left ventricular outflow conduit graft to the distal ascending aorta. Side-to-side anastomosis between the 8-mm coronary graft and the LVOT composite conduit. J, Takedown of right atrial connection to the right pulmonary artery. Closure of atrial septal defect with a bovine pericardial patch. K, Insertion of pulmonary artery band between the right and left pulmonary arteries. L, Final reconstruction. *LVOT*, Left ventricular outflow tract.



VIDEO 1. Operative steps showing novel biventricular Fontan conversion in criss-cross heart and superior-inferior ventricles using extra-anatomical left ventricle apex to ascending aortic composite prosthesis. Video available at: https://www.jtcvs.org/article/S2666-2507(25)00046-X/fulltext.

(Figure 2, *H*). This was followed by a wide side-to-side anastomosis between the 8-mm coronary graft and the composite conduit. The final configuration can be visualized in Figure 2, *I*.

Lastly, the right atrial connection to the right pulmonary artery was taken down and oversewn. The atrial septal defect was closed with a bovine pericardial patch (Figure 2, *J*). After weaning off bypass a pulmonary artery band was created between the right and left pulmonary arteries to prevent a pulsatile cavopulmonary anastomosis (Figure 2, *K*). The final one and half biventricular conversion is shown (Figure 2, *L*).

Early after surgery, there was diastolic dysfunction that needed to be managed with diuretics and afterload reduction. Currently, 1 year after operation, she is clinically asymptomatic and back to working out at the gym.

DISCUSSION

Long-term survival for children with single-ventricle disease is improving. Transplant-free survival at 15 years after a Fontan procedure has been found to be 85% for those with a systemic right ventricle, 93% for those with a systemic left ventricle, and 89% for those with a nonclassifiable ventricle.[1] For many years before death, patients who have undergone a Fontan procedure experience long-term morbidities including thromboembolic disease, venovenous collaterals, arrythmias, protein-losing enteropathy, liver and kidney dysfunction, and plastic bronchitis.[2] Survival to heart transplant is typically combined with liver transplant, which are associated with long postoperative hospital stays, significant morbidities, and 1-, 3-, and 5-year survival rate of only 87%, 80%, and 78%, respectively.[3] The premise of biventricular conversion is to lower the atrial venous pressure to circumvent the chronic effects of the Fontan circulation.

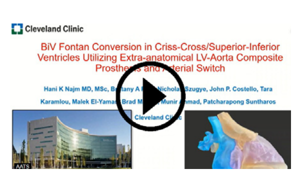
When creating novel operative techniques, 3-dimensional printed and digital models using virtual reality provide essential information to the surgeon to assess feasibility, refine sequence, and anticipate problems.[4-6] The thought process as we envisioned this procedure was aided by the fact the apicoaortic conduits has been used successfully with good results in cases of severe left ventricular outflow tract obstruction.[7] In addition, data are available for the long-term results of the Cabrol technique for coronary anastomosis.[8] Hence, we were reasonably comfortable in predicting the functionality of the proposed reconstruction. In addition, experience with apical cannulation for LV assist devices helped this creative procedure.[9] The novelty of this procedure is in diverting the entirety of the LV outflow from the VSD to the apex and the addition of distant prosthetic valve and an arterial switch; a proof of concept that aortic valve and coronaries need not be on the immediate outlet of the LV. In patients who are clinically well but with signs of early Fontan-related complications, biventricular conversion should be considered whenever possible before overt organ failure.

CONCLUSIONS

This case demonstrates several novel approaches to perform a biventricular conversion in a complex adult congenital cardiac patient who had been directed down a single-ventricle pathway. The ability to use an LV apical to ascending aortic valved graft is demonstrated, including an arterial switch to resolve an anatomically challenging configuration with superior-inferior relation of ventricles.

Webcast

You can watch a Webcast of this AATS meeting presentation by going to: https://www.aats.org/resources/novel-biventricular-fontan-con-7336.



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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