literature, this has been performed by closure of both ends of the tunnel by transaortic and transventricular access. However, closure of just the ventricular end is an option to maintain coronary perfusion [6].

In conclusion, we present a neonate with a rare aorta-right ventricular tunnel connected to the apex of the right ventricle. Surgical closure of both ends of the ARVT was successfully performed.

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References

Tailored Strategies for Staged Management of Complex Congenital Cardiac Lesions
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Although many complex congenital cardiac lesions can be primarily repaired, there remain circumstances where a staged approach may carry lower risk than a primary repair. We present 4 clinical cases where stable intermediate stages allowed somatic growth and facilitated successful biventricular repair or univentricular palliation.

The choice of complete primary repair for complex congenital cardiac disease versus a staging strategy is usually straightforward; most biventricular lesions are now repaired primarily. In some cases, subtleties of lesion complexity such as hypoplastic pulmonary arteries or concomitant noncardiac lesions such as tracheal stenosis increase perioperative risk [1, 2]. A staged approach that addresses immediate physiologic and anatomic concerns to allow a period of growth and stability is more appropriate in these cases and allows subsequent complete repair at a more acceptable risk. Univentricular lesions with issues such as transposition streaming and high pulmonary vascular resistance need optimal conditions for pulmonary vascular resistance to decrease. The selection and application of staging strategies is case specific. We present 4 cases where intermati palliative strategies were designed to enable somatic growth to mitigate unfavorable anatomy and physiology.

Case Reports
Patient 1
A male infant presented with the following univentricular anatomy: d-transposition of the great arteries (d-TGA), right ventricular hypoplasia, straddling right atrioventricular valve, nonrestrictive ventricular septal defect (VSD), and excess pulmonary blood flow. He underwent neonatal balloon atrial sepsostomy and pulmonary artery banding at age 3 months in another institution. Cyanosis limited band tightening due to transposition streaming; consequently, pulmonary blood flow remained excessive. At age 8 months, his pulmonary vascular resistance index was 10 Wood units m² excluding Fontan candidacy. He was then referred to our unit. We opted to perform atrial septectomy, palliative arterial switch operation to correct the transposition streaming, and restricted pulmonary blood flow by placing a 6-mm polytetrafluoroethylene interposition graft between the neo-pulmonary root and the distal pulmonary artery as a modified banding technique. The postoperative course was uncomplicated and systemic saturations were 75% to 85% during the intervening months. Six months later his pulmonary vascular resistance index was 2.7 Wood unit m². He proceeded to creation of a bidirectional superior cavopulmonary anastomosis at age 20 months and subsequently completed his total cavopulmonary connection at age 3 years. He remains asymptomatic at 3 years follow-up.

Patient 2
A 3-kg female presented with d-TGA and multiple VSDs, including a large outflow malalignment defect, 2 apical muscular defects, and a large mid-muscular defect. As the left ventricle was at systemic pressure we performed an elective arterial switch operation at age 6 weeks but considered the risks of heart block, residual VSDs, and ventricular dysfunction to be substantial if VSD closure was attempted at this stage. We therefore
interposed a short segment of 5-mm polytetrafluoroethylene graft between the reconstructed neopulmonary root and the distal main pulmonary artery as a modified pulmonary artery band. The baby was discharged on postoperative day 8. At age 15 months and weighing 8 kg she underwent pulmonary artery “debanding” and closure of the VSDs. The outlet and mid-muscular defects were closed transatrially while the apical muscular defect was closed through a limited apical right ventriculotomy. Debanding involved removal of the interposition graft, augmentation of the posterior (facing) pulmonary sinuses and main pulmonary artery with bovine pericardium, and anastomosis of the reconstructed pulmonary root to the distal pulmonary confluence. The postoperative course was uneventful and at 6 weeks follow-up the child is well without evidence of residual lesions.

**Patient 3**

A 2.3-kg neonate presented with the following anatomy: a right atrial isomerism, pulmonary atresia or VSD with d-TGA, severely hypoplastic pulmonary arteries, supracardiac total anomalous pulmonary venous connection (TAPVC), tracheal stenosis, and low birth weight. The unobstructed TAPVC connected to a left superior vena cava draining to an unroofed coronary sinus. The immediate priority was to establish a reliable source of pulmonary blood flow and promote growth of the 2-mm diameter pulmonary arteries. A 3.5-mm left modified Blalock-Taussig (BT) shunt was constructed with ligation of the ductus arteriosus on day 3 of life. Systemic oxygenation (SaO₂ 75%) and cardiac output were satisfactory. Mid-tracheal stenosis due to complete tracheal rings was identified at age 6 weeks and supported non-invasively with continuous positive airway pressure. At age 4 months and weighing 3.8 kg we proceeded with tracheal resection and sliding tracheoplasty on cardiopulmonary bypass. The supracardiac TAPVC was addressed by suturing the pulmonary venous connection to the posterior left-sided atrium using partial occlusion clamps with the heart beating. The central pulmonary arteries remained hypoplastic (4 mm) so the modified BT shunt was upsized to 5 mm. Subsequently, at 8 kg and aged 16 months with 6.1-mm central pulmonary arteries she underwent a Rastelli repair with a 12-mm pulmonary homograft, shunt takedown, and atrial septation. Initial right ventricle pressure was 85% of systemic with good hemodynamics. She was discharged home 3 weeks postoperatively on nocturnal continuous positive airway pressure at 7 cm water. Her pulmonary hypertension continues to improve with right ventricle pressures half systemic at 5-months follow-up.

**Patient 4**

A 2.6-kg male presented with truncus arteriosus type A3 (van Praagh) with hypoplastic (2 mm) branch pulmonary arteries. The ductus arteriosus supplied the left pulmonary artery (LPA) and the right pulmonary artery (RPA) arose from the left aspect of the common arterial trunk. This anatomy was high risk for primary repair due to the pulmonary arterial hypoplasia and discontinuity. Consequently, at age 3½ weeks the central pulmonary arteries were reconstructed on cardiopulmonary bypass and pulmonary blood flow was established using a 3.5-mm modified BT shunt. The postoperative course was uneventful; the baby was discharged with systemic saturations of 80% to 85%. At age 16 months and weighing 8.8 kg with an RPA of 7.1 mm and LPA 2.9 mm, he underwent closure of the subtruncal VSD, takedown of the shunt, and creation of right ventricle to pulmonary artery continuity using a 14-mm pulmonary homograft. Early right ventricle pressure was 60% of systemic. At most recent follow-up 2½ years after repair, he is asymptomatic and fully saturated with mild LPA origin stenosis.

**Comment**

Although primary repair of complex cardiac lesions is preferable in neonates, there remain situations where a staged approach may reduce total risk. Interim somatic growth is used to optimize anatomy and physiology for a later successful repair. Hypoplastic pulmonary arteries are a substantial impediment to successful neonatal biventricular repair. Early reconnection of disconnected pulmonary arteries and use of systemic-to-pulmonary artery shunts can promote growth of hypoplastic pulmonary arteries. Moreover, early high resistance limits unfavorable diastolic run-off until pulmonary artery growth occurs and resistance decreases, thereby producing a robust interim physiologic state. Cyanosis due to transposition streaming may limit effective pulmonary artery banding in single ventricle lesions. The arterial switch operation corrects streaming and the “banded” situation, with an outflow valve on the pulmonary side, is a robust preparation providing a balanced circulation.

Although palliative procedures are associated with an interstage mortality risk [3], intensive monitoring and early intervention can dramatically improve outcomes for these patients in the current era [4]. Moreover, ventricular dysfunction, conduction block, and baffle pathway obstructions have been avoided in the cases presented, likely due to the larger cardiac chamber sizes at the time of final intracardiac repair. Major airway interventions such as slide tracheoplasty can be incorporated at a stage where repair risk is minimized. Importantly, staging should be planned so that the interim physiologic and anatomic state is hemodynamically robust, allows for a significant degree of growth, and carries a discernibly lower risk than primary complete correction. Using these concepts, one can broaden therapeutic strategies and introduce alternate concepts for managing complex lesions.

**References**

Successful Use of the Total Artificial Heart in the Failing Fontan Circulation

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Typical left ventricular assist devices are often ineffective for the failing Fontan circulation. We report the first successful use of a total artificial heart as a bridge to transplant in a patient who had previously undergone a Fontan operation.


The Fontan operation is the final planned palliative operation for children with functional single ventricles (SVs). This circulation is usually tolerated initially, but eventually fails secondary to chronic low cardiac output and elevated venous pressure. In the failing Fontan circulation, placement of a ventricular assist device (VAD) may not address the underlying circulatory impairments. Implantation of a total artificial heart (TAH) restores a “normal biventricular circulation” to potentially reverse systemic manifestations of Fontan failure and improve patient candidacy for transplantation.

A 13-year-old male with pulmonary atresia and intact ventricular septum developed severe, refractory circulatory failure 10 years after an extracardiac Fontan operation. He was hospitalized at an outside institution with severely depressed left ventricular systolic function, respiratory failure, hepatic insufficiency with coagulopathy, and plastic bronchitis.

On transfer to our hospital, the patient was in extremis with multiorgan failure and respiratory decompensation. Echocardiography and catheterization confirmed severely depressed ventricular function with elevated Fontan pressures. With informed consent from the family, a 70-cc SynCardia TAH (SynCardia Systems, Inc, Tucson, AZ) was implanted (Fig 1).

Cardiopulmonary bypass was initiated through the femoral vessels. Because of the extracardiac conduit (ECC) Fontan pathway the remaining native right atrial (RA) cavity was too small to receive superior vena cava (SVC) and inferior vena cava (IVC) flow (Fig 2A). Therefore, in order to insert the TAH, it was necessary to construct a “neo-RA.” After aortic cross-clamping, the ECC and SVC were separated from the right pulmonary artery, which was reconstructed with a homograft patch. Because the existing ECC was too small to serve as the neo-RA, a 24-mm Gore-Tex graft (W.L. Gore and Associates, Flagstaff, AZ) was inserted as an interposition graft between the IVC and the SVC to serve as a neo-RA. The left atricle was excised, preserving the mitral annulus and a cuff of ventricular muscle. The rudimentary right ventricle was resected. The remaining free wall of the RA was used to close the atrial communication, creating a pathway for drainage of the right pulmonary veins to the left atrium.

An atrial connector for the TAH was sutured to the mitral annulus and cuff of left ventricular muscle. An ellipse of the Gore-Tex graft was excised and an atrial connector sutured to the neo-RA. The pulmonary artery bifurcation was opened and the pulmonary artery outflow graft anastomosed in an end-to-side fashion. The aortic outflow graft was anastomosed in an end-to-end fashion to the aorta. The drive lines for the artificial ventricles were tunneled to the left upper quadrant. The artificial ventricles were filled with saline and attached to the atrial connectors and the outflow grafts (Fig 2B).

The postoperative course was complicated by significant bleeding and external compression of the neo-RA and reconstructed IVC that required reexploration and eventual stent placement. Additional complications included rhabdomyolysis and renal failure necessitating renal replacement therapy. Therapy with inhaled tissue plasminogen activator and rigid bronchoscopy were necessary to clear the airway of the casts resulting from plastic bronchitis. Cardiac output was well preserved with the VAD rates between 120 and 160, percent systole at 50% to 55%, and a cardiac output ranging from 5.5 to 75 L/minute. Over several weeks, the patient demonstrated recovery of all end-organ function, including resolution of plastic bronchitis. The patient underwent rehabilitation therapy and was able to ambulate without assistance. On postoperative day 61, a suitable donor heart became available and the patient underwent orthotopic heart transplantation.

The patient suffered a cardiac arrest on posttransplant day 5 related to severe graft dysfunction from combined acute antibody mediated and cellular rejection. The