malperfusion of the celiac trunk, superior mesenteric artery, renal arteries, and left external iliac artery.

Emergency endovascular stent grafting was performed 6 hours after the initial operation. Two aortic self-expandable, noncovered stents (E-XL, 32 mm × 130 mm and 28 mm × 100 mm; Jotec GmbH) were placed in the dissected thoracoabdominal aorta to expand the true lumen and increase perfusion of the abdominal branches. Persistent superior mesenteric artery malperfusion was treated with 2 balloon-expandable Herculink stents (6.5 mm × 18 mm; Guidant, Indianapolis, Ind), and 2 self-expandable nitinol SMART iliac stents (8 mm × 60 mm and 7 mm × 40 mm; Cordis Corporation, Bridgewater, NJ) were deployed into the left external iliac artery. Postprocedure angiography showed sufficient perfusion of all abdominal aortic branches and the arteries of the left leg (Figure 1).

The patient was successfully weaned from the ventilator 8 weeks postoperatively after a prolonged clinical course. Eleven weeks after the initial operation, she was mobilizing fully without any permanent neurologic deficits.

DISCUSSION

The frozen elephant trunk technique allows for single-stage repair of the ascending aorta and transverse aortic arch after antegrade endovascular treatment of the DA.^{1,2} In patients with acute type A dissection, coverage of additional intimal tears and improved true lumen perfusion may lead to false lumen exclusion and obliteration after the frozen elephant trunk procedure.³ Although midterm results for this technique are promising,

controversy exists as to whether a hybrid or a conventional approach should be applied for type A aortic dissection.^{3,4}

The case described herein reveals an unexpected intraoperative complication requiring switching from a hybrid to a conventional surgical repair with extra-anatomic reconstruction of the thoracic aorta, followed by emergency endovascular treatment of the remaining aorta owing to significant malperfusion. Our case illustrates the complex and multidisciplinary approach that is occasionally required for patients with acute aortic dissection.

The precise cause of the technical device failure during our frozen elephant trunk operation is difficult to determine. We can postulate, however, that stent-graft deployment in the very small true lumen might have resulted in trapping of the device's nose cone and inability to extract the prosthetic portion of the hybrid device. A newer version of the E-vita stent graft (E-vita open plus; Jotec GmbH) now has a much lower profile nose cone to prevent this problem from occurring.

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Long-term systemic right ventricular support in transposition and dextrocardia

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For patients with complex congenital heart disease (CHD) who are in end-stage heart failure, a significant disparity in the use of mechanical circulatory support persists.¹ Recent reports have shown the feasibility and initial success of supporting patients with systemic morphologic right ventricles (RVs) with continuous-flow ventricular assist devices (VADs)²; however, long-term outcome data are lacking. We present our successful experience with a HeartMate II left ventricular assist system (Thoratec Corporation; Pleasanton, Calif) in a mirror image position to support the

FIGURE 1. Intraoperative transesophageal echocardiography. A, The systemic right ventricular apex was indented by the surgeon to locate the appropriate location for the ventricular assist device inflow cannula. B, Appropriate positioning of the inflow cannula was confirmed after the procedure.

systemic morphologic RV in a patient with congenitally corrected transposition of the great arteries and situs inversus with dextrocardia.

CLINICAL SUMMARY

A 56-year-old man with a history of situs inversus, dextrocardia, atrioventricular discordance, and ventriculoarterial discordance (levotransposition of the great arteries) with a ventricular septal defect, atrial septal defect, and pulmonary stenosis was seen with progressive heart failure symptoms, systemic RV failure (ejection fraction <20%), and maximal oxygen consumption of 10.7 mL/(kg · min), despite both optimal medical therapy and biventricular pacing. This patient had undergone 2 previous sternotomies for atrial and ventricular septal defect repair and pulmonary valvotomy at 13 and 31 years of age, respectively. He was referred for a heart transplant; however, he was found to have renal cell carcinoma, which was treated by nephrectomy. VAD implantation was therefore recommended for long-term support to ensure cancer-free survival.

Preoperative imaging for the implantation of a HeartMate II VAD confirmed the systemic ventricle to be a morphologic RV that was located posterolaterally to the morphologic left ventricle in the right hemithorax. A reoperative median sternotomy was performed. Exposure of the patient's aorta was limited by its rightward displacement and the large overlying pulmonary artery. Cardiopulmonary bypass was instituted from the left femoral vessels. The VAD outflow graft was running to the left (medial) of the heart and was anastomosed to the anterior ascending aorta.

The surface of the RV apex was indented and observed on intraoperative transesophageal echocardiography to select an appropriate cannulation site (Figure 1, A). Prominent trabeculae were encountered and were aggressively resected in the area of the ventriculotomy to prevent obstruction of the inflow cannula. The VAD inflow cannula was then implanted in the standard fashion, VAD support was initiated, and the patient was weaned from cardiopulmonary bypass.

Transesophageal echocardiography was also used to confirm appropriate orientation of the VAD inflow cannula

toward the atrioventricular valve (Figure 1, B). Because of the posterolateral displacement of the systemic RV and the posterior direction of the apex, the VAD pump and inflow cannula needed to be aligned in a nearly anteroposterior direction to maintain the proper orientation (Figure 2).

The patient had an uncomplicated postoperative course and was discharged 13 days after the operation. He remains free of symptoms and has not required readmission to the hospital in more than 1 year.

DISCUSSION

Because of advances in pediatric cardiovascular care, most children with complex CHD survive to adulthood. Among adults with CHD, ventricular dysfunction and heart failure are especially common in those with single or

FIGURE 2. Postoperative sagittal computed tomographic scan reconstruction demonstrating the near anteroposterior orientation of the ventricular assist device pump and inflow cannula within the right hemithorax. A, Anterior; P, posterior.

systemic RV, and such patients represent a growing subset of the population being listed for heart transplants. The need for mechanical circulatory support for this patient population is therefore growing.

Familiarity with CHD anatomy, morphology, and variations in the hemodynamic physiology is important when implantation of a VAD is being contemplated. For example, the RV is more triangular and has prominent trabeculations and the moderator band near the apex. All these characteristics may affect flow into the VAD inflow cannula and lead to low flow, suction events, or VAD obstruction and thrombosis. Attention to the potential need for resection of the trabeculae and the moderator band is important. There have been reports of the use of both pulsatile and continuous-flow VADs² in morphologic RVs to support the systemic circulation. Preoperative and intraoperative imaging is critical in achieving optimal VAD inflow cannula orientation.

Although the HeartMate II VAD is not designed for patients with dextrocardia, it can be adapted to them.³ When

the device is rotated 180°, its orientation is reversed, making it appropriate for implantation into the right hemithorax. Next-generation VADs with smaller pump designs will likely minimize this issue.

This case illustrates how patients with complex CHD lesions can be supported with VADs with good long-term outcome as long as proper attention is paid to anatomic details and their potential impact on device implantation and function. Further studies are necessary to confirm the applicability of this technology across the spectrum of CHD.

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