There are several options to address RV failure in patients after atrial switch operations. Some groups advocate the conversion from the atrial to an arterial switch operation,[1] and others support orthotopic HTx.[2]

Although we are generally in favor of HTx, we also face the general problem of a lack of donors for patients awaiting HTx.[4] A bridge to transplantation with different VADs has been one answer for patients whose condition deteriorates while on the waiting list, but using VADs in patients with ''abnormal'' anatomy presents a surgical challenge, especially after the Senning operation.

As patients with TGA rarely show left ventricular (LV) dysfunction, the use of a univentricular VAD should be adequate for most, if not all, patients. To the best of our knowledge, there is no single report in the literature describing biventricular mechanical circulatory support in patients with ventricular failure after atrial switch operation. Installing an RV assist device is easy, because the enlarged and thickened left atrium is an optimal structure for the inflow conduit. The right ventricle may be another option for the LV assist device inflow conduit, but it requires dissection and exposure of the right ventricle and ventricular fibrillation, or even cardioplegic cardiac arrest as described by George and colleagues.[5] However, if biventricular support should be required, installing an LV assist device might be done by cannulating the superior vena cava as the right atrium, which is ''in the middle'' of the heart and completely hidden by the left atrium. Another option might be the apex of the LV. Sewing a graft to the pulmonary artery may be more difficult than normal, because the main pulmonary artery in patients with TGA is often located behind the ascending aorta.

CONCLUSIONS

Despite these difficulties, implantable VADs in failing ventricles have the potential to allow hemodynamic, metabolic, and pulmonary recovery. The implantation of VADs in children and young adults with congenital heart disease requires modifications of the perioperative management and standard surgical techniques as described.

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Ventricular septal defect repair in an infant with severe pulmonary hypertension and preoperatively diagnosed left ventricular noncompaction

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Intracardiac repair for congenital heart disease complicated by left ventricular noncompaction (LVNC) has been rarely described. This is the first report of ventricular septal defect (VSD) repair in an infant with severe pulmonary hypertension (PH) and preoperatively diagnosed LVNC.

CLINICAL SUMMARY

A 2-month-old infant with a VSD was referred to the Toho University Omori Medical Center Department of

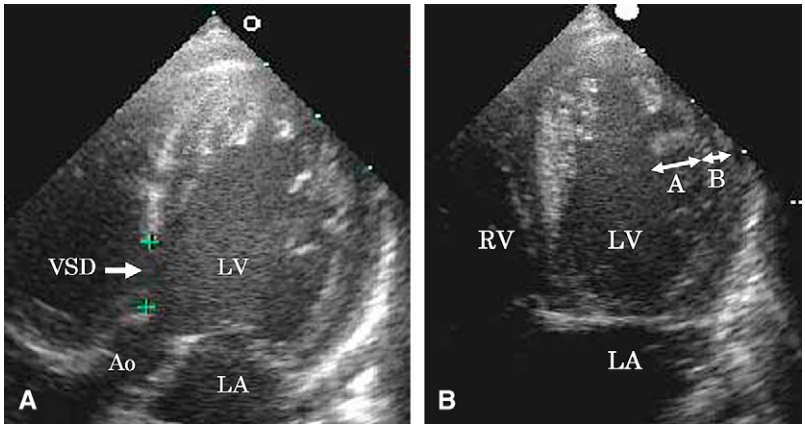

FIGURE 1. A, Four-chamber view of echocardiogram shows a large perimembranous inlet VSD. Distinct trabeculations are recognized in the apical and mid-lateral segments inside the left ventricle. B, Another 4-chamber view of echocardiogram. There are prominent trabeculations and deep intertrabecular recesses in the apex and mid-lateral of left ventricle. *Arrow A* shows the distance from the peak of prominent trabeculations to intertrabecular recesses: the noncompacted layer. *Arrow B* shows the distance from the epicardium to the intertrabecular recesses: the compacted layer. This case satisfies the diagnostic criterion for LVNC that the length of *A* must be greater than double the length of *B*.[1] *Ao,* Aorta; *LA,* left atrium; *LV*, left ventricle; *VSD,* ventricular septal defect; *RV,* right ventricle.

Pediatrics. An echocardiogram revealed that the VSD was located at the perimembranous inlet and was greater than 10 mm in diameter (Figure 1, *A*); severe PH was also noted. LVNC was later diagnosed because of the presence of prominent trabeculations and deep intertrabecular recesses in the left ventricle (Figure 1, *B*). Color Doppler echocardiography showed the presence of blood flow into the deep intertrabecular recesses in the left ventricle.

After admission, the patient's condition gradually deteriorated and he had congestive heart failure. Left ventricular percent fractional shortening decreased by 20.6%, and left ventricular end-diastolic diameter was 35.0 mm on echocardiography. Progression of cardiomegaly and lung congestion were obvious on a chest x-ray film taken at age 4 months.

Then cardiac catheterization was performed. Right ventricular pressure was almost equal to left ventricular pressure. Aortic pressure was 85/48 mm Hg, right ventricular pressure was 79/5/10 mm Hg, and pulmonary artery pressure was 77/30/53 mm Hg; the massive left-to-right shunt was also evaluated (Qp/Qs = 4.9, Rp = 3.1 WU/m^2, L-R shunt ratio = 77%).

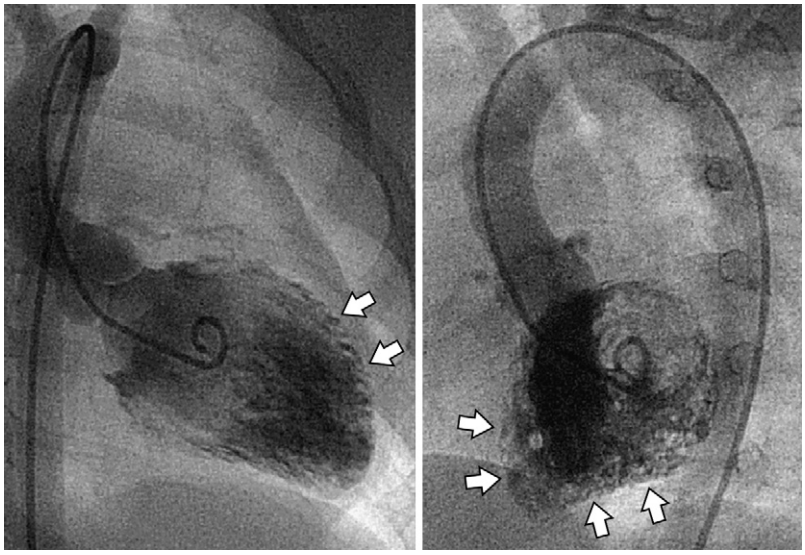

FIGURE 2. Left ventriculography at diastolic phase 10 months after the repair. Right anterior oblique position view and left anterior oblique position view revealing many characteristic filling defects (*arrows*) in the LVNC.

To relieve cardiac volume overload and progressive PH, intracardiac repair of the VSD patch closure was indicated despite the patient's low cardiac output. During surgery, the VSD was closed with a patch, but low cardiac output syndrome developed. Decreased urine output required peritoneal dialysis; PH crisis occurred and was treated with nitric oxide inhalation. Intensive treatment improved the patient's hemodynamic and respiratory status, and he was weaned from peritoneal dialysis. Nitric oxide inhalation therapy was stopped, and sildenafil citrate was given via nasogastric tube to prevent PH crisis.

On the 33rd day after surgery, an echocardiogram showed improvements in percent fractional shortening of 41% and left ventricular end-diastolic diameter of 28.2 mm. The patient was discharged, and home oxygen therapy was continued for residual PH. Ten months later, cardiac catheterization was done. Left ventriculography demonstrated characteristic findings of LVNC (Figure 2). However, PH was clearly improved: Pulmonary artery pressure was reduced to 26/10/16 mm Hg, and aortic pressure was 88/50 mm Hg. At present, 3 years after the repair, he is doing well without home oxygen therapy or sildenafil administration.

DISCUSSION

LVNC is categorized as an unclassified cardiomyopathy in the World Health Organization classification of cardiomyopathy.[1] The characteristic pathologic findings of LVNC include excessively prominent trabeculations and deep intertrabecular recesses.[1-3]

Patients with LVNC are candidates for serious heart failure, arrhythmia, and embolism.[1,2] Patients with LVNC have a poor prognosis because of the absence of established therapies. In serious cases, heart transplantation may be indicated.[2] However, in Japan heart transplantation remains a difficult option, particularly for neonate and infant patients. Unfortunately, with the exception of transplantation, the operative indications for congenital heart disease with LVNC and severe PH remain controversial.

Pignatelli and coworkers[3] reported a large series of pediatric LVNC cases. In that series, LVNC was associated with congenital heart disease in only 5 of 36 patients. Two of the 5 patients underwent open surgery (VSD repair and Norwood procedure for hypoplastic left heart syndrome), although LVNC was diagnosed only after surgery in both patients. Remarkably, both patients survived and were alive at the time of the report. These unexpectedly favorable outcomes encouraged us to pursue surgical treatment in the present case.

However, another case report described a neonate who underwent end-to-end anastomosis as an arch repair for the complex of aortic coarctation and VSD with LVNC that had been diagnosed before surgery.[4] The unfortunate and sudden death might have been caused by PH induced by surgical intervention in the neonatal period and a remaining large VSD.

Recent improvements in both the technology of echocardiography and our understanding of LVNC should make diagnosis of LVNC easier. In one report, an adult patient with isolated LVNC underwent successful left ventricular restoration and valve plasties.[5] In Japan, the range of surgical interventions other than heart transplantation is expected to be extended for patients with LVNC.

CONCLUSIONS

To our knowledge, this is the first successful VSD repair in an infant with severe pulmonary hypertension and preoperatively diagnosed LVNC. Because our patient continues to be at risk for heart failure, arrhythmic attack, and thromboembolism, careful monitoring will continue.

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