number of advantages including versatility, ease of implementation, and familiarity, this modality has a number of disadvantages, including immobilization, requirement for intensive care monitoring, and risks of bleeding, thrombosis, infection, and multiorgan failure. These attendant disadvantages, which increase over time, hamper the use of ECMO for long-term support.

Experience with ventricular assist devices in children is increasing, as smaller devices are becoming available. Hetzer and colleagues [9] reported results of the Berlin Heart ventricular assist device (a pneumatic paracorporeal assist device) in a critically ill group of pediatric patients ranging in age from 6 days to 16 years. The authors reported an overall survival of 40%. The most frequently observed complication was postoperative bleeding.

Recently, the MDBVAD has been demonstrated to be a valid option for bridge-to-transplantation in the adult population [3, 4]. This device is an implantable electromagnetically-actuated axial flow pump. The initial multicenter experience has demonstrated a post-implantation 30-day survival of 81%; also, nearly one-third of the patients undergoing implantation have undergone a successful transplant after a median time of support of 74.5 days [3]. Major advantages of this device include ease of implantation, reduced size, ready patient mobilization post-implant, and the possibility for discharge home while waiting for transplant. No infections were reported after implant. The incidence of post-implant bleeding was the major complication.

The ventricular assist devices currently available in the United States for small pediatric patients (BSA < 1.4) have been essentially limited to the Thoratec, Abiomed, and centrifugal pump systems. We believe that the MDBVAD offered distinct advantages with respect to implantability, maintenance of patient mobility, and possibly a lower thromboembolic risk.

This report describes the first pediatric implantation of the MDBVAD in the United States. This patient also represented the youngest implant to date worldwide. Our experience has confirmed the reported encouraging results. The patient's physiologic and psychological status improved dramatically after LVAD implantation. LVAD function did not precipitate right ventricular dysfunction, although overall cardiac output was clearly right ventricular dependent. Postoperative mortality after MDBVAD has been strongly related to preoperative morbidity [3, 4]. Early timing of LVAD implant prior to development of multisystem organ failure is likely to be critical for success.

Despite its promise, the MDBVAD has a number of limitations. First, as with all LVADs, its application is limited to patients with normal pulmonary function. Second, although it is much smaller than existing pulsatile LVAD designs, it is still much too large for use in neonates and small children. In general, the MDVAD is limited to patients with a body surface area of 1.2 m^2 or greater, although a modified version of the pump (DeBakey VAD Child System) has subsequently been introduced for use in patients 5 to 16 years of age (body surface area, > 0.7 m^2). In this patient, significant hemolysis became evident toward the end of support. This was believed to be secondary to pump thrombus. Finally, like many other LVADs, it requires full anticoagulation. Future technological improvement will likely provide surgeons with miniaturized pumps that can be accommodated by smaller children and infants.

In conclusion, we report the feasibility of MDBVAD as a device for ventricular support in children. Early implantation is recommended to preserve organ function for a successful bridge to transplantation.

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FEATURE ARTICLES

Double Orifice Tricuspid Valve in an Infant With Tetralogy of Fallot

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Atrioventricular valve duplication, also known as double orifice valve, is an excitingly rare anomaly that can involve either the mitral or more uncommonly the tricuspid valve. Herein we describe a case of a double orifice

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tricuspid valve that prejudiced the complete repair in a symptomatic infant with tetralogy of Fallot.

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This extremely rare malformation first reported by Greenfild [1] in 1876, is characterized by the presence of a double orifice in either the tricuspid or mitral valve. Each orifice is provided with a fully developed subvalvular tensor apparatus. It can either occur isolated or in association with other cardiac malformations, especially with atrioventricular septal defect.

The term "duplication" was first used by Hartman in 1937 who classified the double orifice mitral valve into three anatomic variants: (1) L = with the accessory orifice eccentric into the commissure; (2) B = with two symmetric central orifices sharing one tensor apparatus; (3) S = with two symmetric central orifices, each orifice with an independent set of chordae and papillary muscles [2]. Cascos and colleagues elaborated on Hartman's classification defining three variants of tricuspid valve duplication: (1) commissural variant, Hartman's type L, with the accessory ostium lying into the commissure; (2) central variant or bridge type, Hartman's type B and S, in which a fibrous bridge of tissue was identified connecting the leaflets dividing the orifice in two; (3) hole variant, which the secondary hole lays within a leaflet, but is still provided with a tensor apparatus [3].

The double orifice tricuspid valve is much rarer than the double orifice mitral valve and has been generally described in association with atrioventricular septal defect or as isolated [2–5]. To the best of our knowledge, we believe it has never been described in association with tetralogy of Fallot.

A 2.9 kilogram full-term newborn of an uncomplicated pregnancy was presented at birth with cyanosis. An echocardiogram showed tetralogy of Fallot and the baby was discharged home in good condition.

One month later the baby was readmitted because of severe cyanosis and one anoxic spell. An echocardiogram repeated at the time of the admission showed tetralogy of Fallot with a hypoplastic pulmonary annulus and a well developed pulmonary trunk and branches. The tricuspid valve appeared to be abnormal with redundant leaflets. The patient was taken to the operating room and surgery was carried out through a median sternotomy on conventional cardiopulmonary bypass with bi-caval and aortic cannulation. Body temperature was lowered to 28°C. Opening the chest disclosed a left anterior descending coronary artery originating from the right coronary artery and crossing the infundibulum 6 to 7 millimetres under the pulmonary valve annulus. A trans-atrial complete repair was planned and after cross clamping the aorta blood cold cardioplegia was administered into the aortic root.

The right atrium opening showed a mildly hypoplastic tricuspid valve orifice with dysplastic redundant leaflets and the presence of two symmetric centrally located tricuspid orifices (bridge type) (Fig 1).

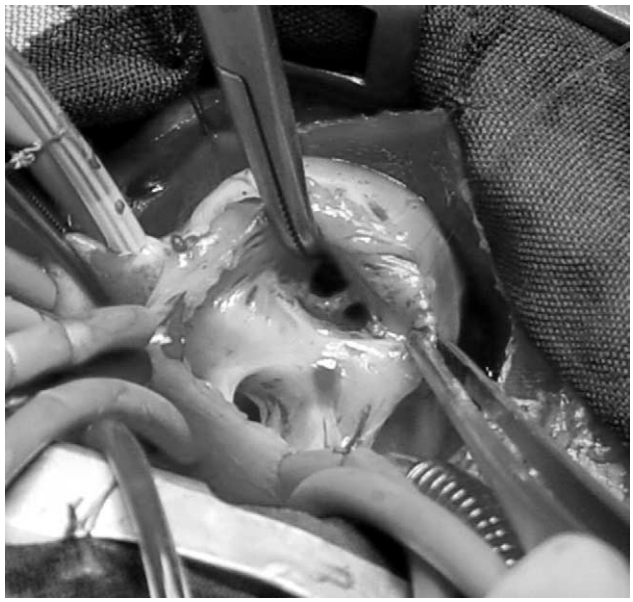

Fig 1. Intraoperative view; surgical orientation. Double orifice tricuspid valve visualized through the right atriotomy.

By saline injection the competence of both orifices was proved, but it was absolutely impossible to visualize either the ventricular septal defect or the right ventricular outflow tract through the tricuspid valve.

An extensive infundibulotomy was not feasible because of the presence of the left anterior descending coronary artery originating from the right coronary artery. A short incision was then made in the pulmonary artery trunk through the pulmonary valve annulus and was carried a few millimeters into the free wall of the infundibulum. Through the incision, a small amount of hypertrophic right ventricular muscle could be excised. The incision was than closed with a bovine pericardial patch. A patent foramen ovale was partially suture closed. The baby was easily weaned from the cardiopulmonary bypass with moderate doses of inotropic support. Pressure in the pulmonary artery was 20 to 10 mm Hg and arterial saturation was 95%. The sternum was closed and the baby was taken to the postoperative intensive care unit where he spent 3 days without any major complications.

On the postoperative day 2, the baby was weaned from the ventilator, and the day after he was transferred to the ward where he spent an unremarkable 10-day recovery period.

The baby is now 5 months old and doing well, waiting for tetralogy of Fallot complete repair. The echocardiogram shows a perfectly competent double orifice tricuspid valve (Fig 2) with no transvalvular gradient and no more than moderate right ventricular outflow tract obstruction.

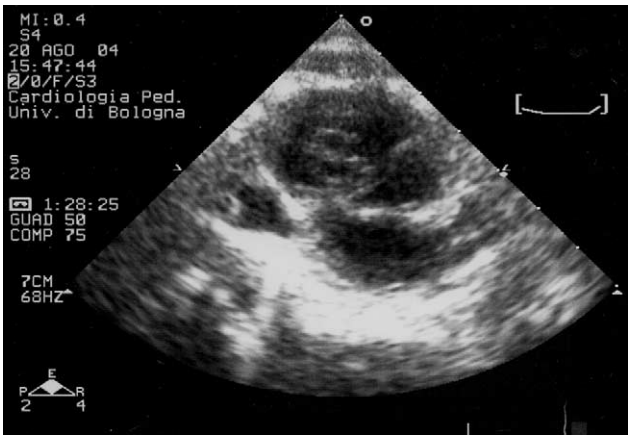

Fig 2. Two-dimensional echocardiogram in parasternal short axis view shows the two orifices of the tricuspid valve during diastole.

Comment

Double orifice tricuspid or mitral valve formation has been explained as an imperfect formation of the atrioventricular valve leaflets by the endocardial cushion growth and fusion, thus explaining the frequent association with partial or complete atrioventricular canal [4, 5]. Nevertheless, both mitral and tricuspid variants can also occur not in association with atrioventricular canal [6]. Usually, if not associated with other cardiac malformations, they are considered benign with no important hemodynamic consequences.

To the best of our knowledge this is the first case of double orifice tricuspid valve that has been reported in association with tetralogy of Fallot. On the other hand, tricuspid valve anomalies other than atrioventricular canal, such as straddling tricuspid valve, Ebstein's malformation, accessory leaflet or leaflets prolapse, have been described, although infrequently, in association with tetralogy of Fallot [7, 8]; nevertheless they had negligible technical impact on the surgical correction of tetralogy of Fallot.

In the patient we reported, the tricuspid anomaly described had a fundamental impact on the surgical strategy, not enabling us to approach the ventricular septal defect and the right ventricular outflow tract through the tricuspid valve. Such a rare occurring malformation associated with the coronary artery anomaly can seriously challenge tetralogy of Fallot complete repair.

At the time of surgery we decided for palliation because of the small body size of the patient. In a larger baby, either a trans-atrial or a trans-ventricular approach may be planned. If a larger tricuspid orifice is found, the ventricular septal defect can be patched through the right atrium and the infundibulum can be enlarged from inside. In this case, to achieve a fair visualization, a partial detachment of the tricuspid valve anterior and septal leaflets from the annulus should be done. However, if the infundibular obstruction appears to be severe, a right ventriculotomy, just below the level of the anomalous anterior descending coronary artery, may be done, closing the ventricular septal defect through it and relieving the infundibular obstruction by positioning a right ventricle to pulmonary artery conduit as an accessory outlet.

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FEATURE ARTICLES

Persistent Sinus Venosus Valve Mimicking Pulmonary Stenosis and Atrial Tumor

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A case of giant remnant of the right sinus venosus valve simulating first a pulmonary stenosis and afterward a left atrial tumor is described. We believe that this is the first reported case in which a correct diagnosis was performed before surgery.

(Ann Thorac Surg 2006;81:1123–6)

Remnants of the embryologic sinus venosus valves are not uncommon. However, the persistence of a sinus venosus valve sufficiently large enough to protrude into the pulmonary artery and obstruct the blood flow through the pulmonary valve is extremely rare. We believe that only three cases have been reported to date [1–3]. We have diagnosed and successfully operated on a

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