CASE REPORT

Correction of Tetralogy of Fallot with Absent Pulmonary Valve Syndrome in a Young Infant Using a Bicuspid Equine Pericardial Tube

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Absent pulmonary valve syndrome (APVS) is an uncommon variant of tetralogy of Fallot (TOF), which manifests morphologically as vestigial pulmonary valve cusps at the right ventricle–pulmonary trunk junction. The aneurysmally dilated pulmonary arteries may compress the tracheobronchial tree and cause severe respiratory distress in the neonatal or infant stage. Early surgical correction in these patients is necessary despite the high operative mortality rate. A 1-day-old male neonate suffered from progressive shortness of breath after birth. Echocardiography confirmed the diagnosis of TOF with APVS. The marked dilatation of pulmonary arteries resulted in airway compression in addition to heart failure. Total surgical correction was performed at 40 days of age, using a homemade bicuspid equine pericardial tube for right ventricular outflow reconstruction. The short-term follow-up echocardiogram demonstrated good motility of the pericardial leaflet. However, patients receiving this type of valved conduit require meticulous long-term follow-up. [*J Formos Med Assoc* 2006;105(4):329–333]

Key Words: absent pulmonary valve, bicuspid, equine pericardium, syndrome, tetralogy of Fallot

The combination of tetralogy of Fallot (TOF) and agenesis of the pulmonary valve is referred to as absent pulmonary valve syndrome (APVS), which was first described by Chevers in 1846.[1] The reported incidence of this condition ranged from 3.6% to 6% among patients with TOF.[2,3] Embryologically, it is similar to classic TOF, except that the ductus arteriosus is usually absent, which is possibly responsible for the dilatation of the pulmonary arteries. Other proposed causative factors of pulmonary aneurysm include the orientation of the infundibulum,[4] the degree of valvular stenosis,[5] and the abnormal fibrotic component of the tracheobronchial tree. Patients suffer from airway compression by the dilated pulmonary trunk and its branches. Some cases present with severe respiratory problems shortly after birth, and require intubation with ventilatory support. Urgent surgical treatment in the neonatal stage is indicated even though operative mortality has been reported to be as high as 30–50%.[1,6,7]

Here, we report successful surgical correction of a case of TOF-APVS at the age of 40 days. The patient's right ventricular outflow tract (RVOT) was reconstructed with a homemade bileaflet equine pericardial tube conduit, which has not been previously reported in the treatment of this syndrome.

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Received: February 23, 2005
Revised: April 7, 2005
Accepted: June 7, 2005

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Case Report

A male neonate was referred to our institution 6 hours after delivery because of tachypnea with cyanosis. He was intubated and given mechanical ventilatory support. On physical examination, his weight was 2890 g, and a Grade III/VI to-and-fro heart murmur in the left upper sternal border was audible on auscultation. Chest X-ray showed prominent bilateral pulmonary arteries and cardiomegaly with a cardiothoracic ratio of 0.7. Color Doppler echocardiography revealed a large ventricular septal defect, overriding aorta, a hypoplastic pulmonary annulus with a rudimentary pulmonary valve, and severely dilated main pulmonary trunk anterior to its branches (Figure 1). TOF with APVS was diagnosed. Cytogenetic analysis revealed deletion of chromosome 22q11. Although the patient was weaned from the ventilator and discharged 15 days later, he was readmitted, re-intubated and given mechanical ventilation due to progressive dyspnea and cyanosis at the age of 1 month. Repeated arterial blood gas data showed persistently high carbon dioxide level and unstable hemodynamic condition despite high doses of inotropic agents. Surgical intervention was considered to be the only chance for survival. The patient underwent surgery at the age of 40 days.

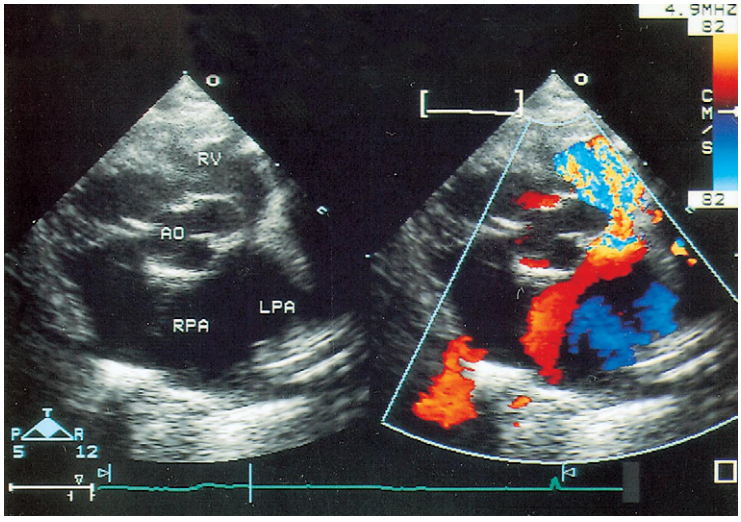

Figure 1. Preoperative echocardiography (parasternal short axial view) shows a large ventricular septal defect, a hypoplastic pulmonary annulus with a rudimentary pulmonary valve, and markedly dilated main pulmonary trunk and its branches. The severe pulmonary regurgitant flow at the right ventricular outflow tract could be identified from the color Doppler images. RV = right ventricle; AO = aorta; RPA = right pulmonary artery; LPA = left pulmonary artery.

The surgical procedure was as follows: after median sternotomy, no thymus was found. Hypothermic cardiopulmonary bypass was conducted with bicaval cannulation after lowering of temperature to a rectal reading of 20° C; the aorta was then cross-clamped, and blood cardioplegic solution was injected. A longitudinal right ventriculotomy was performed. The pulmonary valve was hypoplastic with merely remnant tissue noted. An unruptured sinus valsalva aneurysm was noted incidentally between the ventricular septal defect and the remnant pulmonary valve. The parietal band was excised, and a polytetrafluoroethylene patch was used to close the ventricular septal defect as well as the sinus valsalva aneurysm. The pulmonary arteries were completely dissected to the hilum, and the main pulmonary artery was transected at the level above the pulmonary annuli. The right and left pulmonary arteries were plicated by excising an elliptical portion of the confluent bilateral arteries. A homemade 12-mm bicuspid pericardial tube conduit made from equine pericardium (equine pericardial patch; Edwards Lifesciences, Santa Ana, CA, USA) was constructed according to the method of Kreutzer et al.[8] Briefly, the equine pericardium was trimmed into two patches with different geometric forms: a rectangle (the future conduit) and a trapezoid (the future bicuspid valve). The trapezoid was superimposed on the rectangle, with its longer base placed 5 mm below the distal end of the rectangle. The longest base of the trapezoidal patch was 10% larger than the rectangular one, which was first fixed to the rectangular patch at three points (middle and bilateral ends) with 6-0 polypropylene sutures. These sutures were connected in a curvilinear way to form two valvular cusps. The remnant pericardium on the trapezoidal patch was trimmed along the suture curve. A 12-mm Hegar dilator was placed over the pericardial rectangle with the cuspids. The lateral edges of the rectangle were folded over, trimmed and connected by the double running sutures with 6-0 poly-

propylene. A bicuspid pericardial tube conduit was constructed, and was then used for connection between the right ventricle and distal pulmonary artery.

Due to cardiac swelling, sternal wound closure was delayed. The peak inflating pressure of the respirator was kept above 25 mmHg due to suspicion of peripheral airway compression distal to the pulmonary hilum. Progressive hypercarbia developed on the 2nd postoperative day. Diffuse wheezing was found on auscultation. The respiratory rate was adjusted from 35 to 40 times/min, with the inspiratory/expiratory ratio at more than 1:2 to prevent air trapping. The positive end expiratory pressure was kept at 5 mmHg.

The sternal wound was closed 7 days after the operation, and the baby was weaned from the ventilator 3 days later. Fungemia (*Candida parapsilosis*) and *Pseudomonas* sepsis complicated the postoperative course. These infections were cured with medical treatment. Color Doppler echocardiogram 3 months after surgery showed normal-sized pulmonary arteries. The bicuspid pulmonary leaflet was functioning without regurgitation of flow, and only mild pulmonary stenosis across the RVOT was noted (Figure 2).

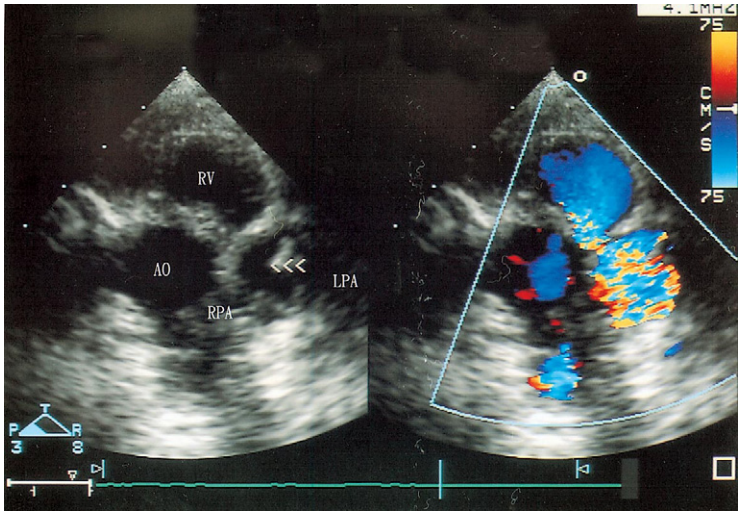

Figure 2. Postoperative echocardiography demonstrates mobile bicuspid leaflets (arrowheads) with mild pulmonary stenosis and regurgitation. RV = right ventricle; AO = aorta; RPA = right pulmonary artery; LPA = left pulmonary artery.

Discussion

APVS is an uncommon disease entity, the precise etiology of which remains to be established. A specific genetic and embryologic mechanism involving the interaction of the neural crest and the primitive aortic arches due to deletions in chromosome 22 is suspected.[9] Cytogenetic investigation in our patient also showed a 22q11 deletion. Embryologically, the only difference between APVS and classic TOF is the absence of the ductus arteriosus in the fetus. It has been speculated that the presence of ductus arteriosus in this lesion is incompatible with fetal survival to birth, because the majority of left ventricular output flows retrograde through the ductus into the right ventricle and across the ventricular septal defect back into the left ventricle.[10] This finally results in heart failure, and only those patients without ductus arteriosus can survive to birth. The agenesis of the arteriosus is also thought to cause dilatation of the pulmonary arterial trunk because of the to-and-fro flow between the pulmonary artery and right ventricle *in utero*. The main clinical presentations of this syndrome include airway compression by the dilated pulmonary trunk and its branches, and right heart failure resulting from severe pulmonary valve insufficiency. However, variations in clinical manifestations may occur according to the onset and severity of the compromised airway. In patients with mild respiratory and cardiac symptoms, the management is similar to that of TOF. For those patients with severe respiratory distress and/or right heart failure in the neonatal or infant stage, urgent surgical treatment is necessary.

The goal of surgical repair is to correct TOF and relieve the central airway compression. Several surgical techniques have been described to accomplish this goal, such as plication of the pulmonary arteries and a valve insertion,[1] homograft interpositions,[2] resection of the main pulmonary artery and anterior portion of the main branches with or without homograft interposition,[11] and outflow reconstruction without valve insertion.[12] Godart et al reported an excellent result after surgical treatment with angioplasty only, and they concluded that there is no need for pulmonary valve inser-

tion in such patients.[13] However, Hew et al reported that aggressive homograft replacement of the pulmonary arteries was associated with improved survival in patients with APVS and respiratory distress.[14] Our patient had severe respiratory distress, and use of a valve within the conduit was considered to be necessary due to concern that high pulmonary vascular resistance during the neonatal stage had led to residual pulmonary valve regurgitation that would complicate the postoperative course. It was therefore considered logical to attempt to normalize the cardiovascular status by anatomic repair with an artificial pulmonary valve. Kreutzer et al reported the outcome of RVOT reconstruction with the autologous bicuspid pericardial tube conduit for D-transposition of great arteries, TOF, truncus arteriosus, pulmonary atresia with ventricular septal defect and double-outlet ventricle.[8] They reported freedom from conduit-related reintervention at 5 and 10 years in 90% and 81% of patients, respectively.[8] However, few of their patients were infants, and the treatment of our patient also differed in that commercially available equine pericardium was used instead of autologous pericardium for reconstruction of the valved conduit. Although short-term (3 months) follow-up echocardiogram demonstrated good motility of the pericardial leaflet in this case, the long-term outcome remains uncertain; close, long-term observation is necessary.

Surgical procedures may not completely resolve the problems associated with TOF-APVS, where the pathology of airway compromise extends beyond the proximal pulmonary artery into the arterioles and related bronchioles. In autopsy cases, tufts of arteries that entwine and compress the intrapulmonary bronchi have been found, which is especially common in young infant patients with severe respiratory symptoms.[15] Additional intrapulmonary bronchovascular anomalies may contribute to severe respiratory problems, even after correction.[16] In our patient, the lung biopsy pathology did not show any evidence of the aforementioned anomalies. Delayed closure of the sternotomy wound with temporary coverage using a prosthetic patch has also been reported to provide decompression of the tracheobronchial tree.[16,17] Thus, in this case, prolonged mechanical ventilation, adequate pulmonary toilet and, possibly, delayed sternal closure were favorable for postoperative recovery.

In conclusion, this case suggests that a homemade pericardial bileaflet conduit might provide a suitable replacement of the homograft for RVOT reconstruction in cases of TOF-APVS with respiratory distress. The long-term results using such a valved conduit need to be followed meticulously.

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