Repair of congenital pulmonary vein stenosis

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Congenital pulmonary vein stenosis (CPVS) is a rare fatal congenital cardiovascular defect and its operative mortality is very high. A 6-year-old boy presented with bilateral congenital pulmonary vein stenosis complicated by severe pulmonary hypertension and mitral insufficiency. A successful angioplasty in combination with mitral valvuloplasty was performed for pulmonary veins with living autologous atrial tissue and sutureless in situ pericardium.

Key words: pulmonary vein stenosis; congenital heart disease; surgery

Introduction
Congenital pulmonary vein stenosis (CPVS) is a rare malformation with a high mortality, and no successful treatment of this disease was reported in China. We report a child with bilateral CPVS complicated by severe pulmonary hypertension and mitral insufficiency, who underwent a successful angioplasty for pulmonary veins with living autologous atrial tissue and sutureless in situ pericardium in combination with mitral valvuloplasty at our pediatric center of cardiovascular surgery.

Case report
A 6-year-old boy felt fatigue, palpitation and dyspnea on exertion in recent 6 months and had lost consciousness while playing games repeatedly in the last 4 months. No characteristic manifestations but pulmonary hypertension were found on physical examination, electrocardiogram and chest film. Findings of echocardiography with color Doppler examination helped to diagnose his disease as CPVS complicated by pulmonary hypertension and mitral insufficiency. Stenosis of the junctional areas was observed between the 4 pulmonary veins and the left atrium. The diameter of the pulmonary veins was about 4 mm with a venous flow velocity of 2.2 m/s, and a tricuspid valve regurgitation jet with a velocity of 4.7 m/s (i.e., an estimated right ventricular pressure of 95 mmHg), and moderate mitral regurgitation was noted. Further investigation of ultra fast computed tomography (UFCT) confirmed the diagnosis (Fig.).

Operation was performed on June 1, 2005. The pulmonary artery was infused with 4ºC protective solution during cardiopulmonary bypass (CPB). After the interatrial septum was incised, the left atrium and mitral valve were explored to probe the orifices and adjacent extrapulmonary venous segments of the pulmonary veins were probed. There were severe tubiform stenosis in the orifices of the left superior, left inferior and right inferior pulmonary veins and membranous stenosis in the extrapulmonary segment of the left superior pulmonary veins, which was about 2 cm away from the orifice, without any abnormality in the vessel wall. Anular dilatation was the main cause of mitral regurgitation. The apex of the heart was retracted anteriorly and rightward to expose the left atrial appendage and the left pulmonary veins. A V-shaped incision was made with one limb through the anterior wall of the left superior pulmonary vein and the other through the left inferior vein. The tip of the left atrial appendage was opened by a fish-mouth incision and sutured to widen the opening in the pulmonary veins. This provided a double pathway for left pulmonary venous return. The stenotic membrane of the left superior pulmonary vein was excised. The right inferior pulmonary vein was incised from its ostium, all the way up to the pericardial reflection, to reach normal segment. Then a flap of the native pericardium was anastomosed to the right atrial wall to create a pouch of the in situ pericardium that passively drains the right inferior pulmonary vein into the wide-open left atrium. The dilated annulus was repaired by anuloplasty.
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Case report

Fig. UFCT 3D computer-generated picture of narrowed pulmonary veins. Arrows: the areas of stenosis.

Postrepair cardiac function was good, and after discontinuation of the cardiopulmonary bypass, the mean pulmonary arterial pressure was 34 mmHg (52 mmHg before operation).

The postoperative course was uneventful and echocardiography showed that the stenosis in the pulmonary veins disappeared and the venous flow velocity was normal. The patient was discharged from the hospital 7 days after the operation.

Discussion

CPVS, a rare fatal congenital cardiovascular defect with a very high operative mortality, is estimated to occur in 0.5% of pediatric cardiac necropsies.\textsuperscript{[1-5]} In most patients, CPVS can be repaired by open-heart angioplasty but percutaneous balloon catheter angioplasty may cause restenosis significantly. In surgical methods, living autologous atrial tissue repair and sutureless \textit{in situ} pericardium repair have a better long-term result and a lower restenotic rate. They have been increasingly accepted but the traditional operation, artificial patch repair, was no longer employed in recent years because of its high rate of restenosis. Repair of CPVS with living autologous atrial tissue was reported first by Pacifico and his colleagues in 1984.\textsuperscript{[6]} This method widens right pulmonary venous stenosis with a flap of interatrial septum and left pulmonary venous stenosis with left atrial appendage. Because of the growth potential of autologous atrial tissue, this method will provide lasting relief of obstruction. Sutureless \textit{in situ} pericardium repair was reported first by Lacour-Gayet F and his colleagues\textsuperscript{[7]} in 1995 to repair acquired right pulmonary venous obstruction after correction of total anomalous pulmonary venous connection (TAPVC) and was used in congenital right pulmonary venous stenosis in 2005.\textsuperscript{[8]} Because of its "no-touch" technique, sutureless repair with \textit{in situ} pericardium reduces the risk of subsequent restenosis. In our patient, living autologous atrial tissue repair and sutureless \textit{in situ} pericardium repair were employed in combination to relieve bilateral pulmonary venous stenosis, and the short-term result was excellent.

Perfusion of the pulmonary artery with hypothermic protective solution during CPB was proved to be an effective method for lung protection, especially for patients with severe pulmonary hypertension.\textsuperscript{[9,10]} The postoperative course of this patient was uneventful. Based on previous studies we believed that the result was at least partly associated with the employment of lung protection.

In short, CPVS is a rare malformation with very poor operative results. The child we report is the first patient who underwent a successful bilateral pulmonary venous angioplasty with living autologous atrial tissue repair combined with sutureless \textit{in situ} pericardium repair in China.

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References


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