Surgical Treatment of Double Outlet Right Ventricle with Absent Pulmonary Valve and Bronchiarctia

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To the Editor: We report a successful treatment of a case diagnosed with double outlet right ventricle (DORV) with absent pulmonary valve (APV) and bronchiarctia.

The patient is a female of 14-year-old with body weight 44 kg, who was referred to our hospital for cardiac murmur and history of exercise intolerance and recurrent respiratory tract infections. She presented cyanotic. Electrocardiogram showed right ventricle hypertrophy and right bundle branch block. The chest X-ray showed right pulmonary artery dilation, pulmonary blood vessels decreased, heart shape enlargement and cardio/thoracic ratio at 0.52. Echocardiography and cardiac computerized tomography (CT) scan supported the diagnosis. The CT scan also showed severe left bronchial stenosis [Figure 1a]. The diagnosis was confirmed in the operation. The operation of total correction was performed under cardiopulmonary bypass with hypothermia. Operative findings included right ventricle enlargement, the aorta overriding the ventricle septal defect (VSD) more than 90%, aorta/main pulmonary artery ratio at 1:1 in diameter, right ventricular outflow tract, and the pulmonary valve annulus stenosis. The absence of pulmonary valve is presented in Figure 1b. After clamping the aorta, the cardioplegia solution was given through the aortic root. Oblique incision was made in the right ventricle outlet tract, abnormal muscles were resected, and the VSD was repaired with autologous pericardial. The right ventricular outflow tract and the main pulmonary artery were widened transannulusly using autologous pericardial with three leaflets. Inotropic drugs were used to maintain cardiac function postoperatively. The patient recovered well during 1-year follow-up.

To our knowledge, DORV with VSD, right ventricle outlet and pulmonary artery annulus stenosis, APV, and bronchiarctia has not been reported in literature. DORV with VSD and pulmonary artery or valve stenosis is a kind of congenital heart disease. The surgical treatment result is not satisfactory.[1] However, the girl needed to undergo the surgical treatment due to complex pathological changes. During the operation, we reconstructed the main pulmonary valve, repaired the VSD, and reconstructed the left and right ventricle outflow tract. We repaired the VSD with autologous pericardial to make the left ventricle outflow tract elastic, considering patient's condition, growth, and development. The autologous pericardium with three leaflets was used to widen the right ventricle outflow tract to avoid the stenosis and reduce the pulmonary valve

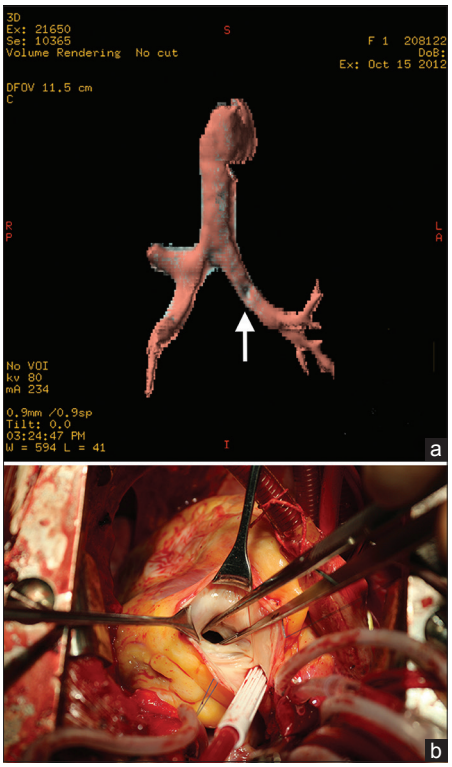

Figure 1: (a) Severe left bronchial stenosis (white arrow) (b) absence of pulmonary valve.

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regurgitation.[2,3] Residual shunt should also be avoided. It is quite difficult for patients with bronchiarctia to recover, but most of them can recover uneventfully by strengthening the airway care, taking physical therapy, and preventing and managing infection postoperatively.[4] The girl's recovery showed that the bronchiarctia is not the surgical contraindication but will affect the period of the recovery. The patient needs to be followed up for cardiac function, and the recurrence of left and right ventricle outflow tract stenosis as well as pulmonary valve regurgitation increases.

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Conflicts of interest

There are no conflicts of interest.

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