Surgical Repair Of Pulmonary Arterial Sling Associated With Tetralogy Of Fallot

Pulmonary artery sling associated with tetralogy of Fallot was successfully repaired in 2 patients. In 1 patient, extensive reconstruction was needed for severe hypoplasia in the left pulmonary artery, followed by definitive repair. In the other patient, surgical repair was achieved in a single-stage fashion.

(Key words: tetralogy of Fallot, pulmonary artery sling, pulmonary stenosis, tracheal stenosis, tracheal bronchus)

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Pulmonary artery (PA) sling may coexist with tetralogy of Fallot. This rare combination of malformations was treated surgically in 2 patients.

Cases

Patient 1. A male baby had shown cyanosis immediately after birth, diagnosed as having tetralogy of Fallot. Since 6 months old, the patient had presented stridor. At the age of 8 months, a left systemic-to-pulmonary shunt was constructed via a left thoracotomy at another hospital. The patient was subsequently referred to our institution at 2 years old, and angiography demonstrated that the left PA was severely hypoplastic originating from the right PA and coursing behind the trachea. Bronchography illustrated tracheal stenosis, as well as an anomalous origin of the right upper bronchus directly from the trachea (Fig. 1A). At the age of 2 years and 11 months, the left PA was divided at its origin, and no portion of the PA was left behind the trachea. The hypoplastic left PA was extraanatomically reconstructed at the level of the intrapulmonary PA using a 10 mm-diameter heterologous pericardial roll tube through a re-thoracotomy in the left chest.

For blood supply to the left lung, a 4 mm-diameter polytetrafluoroethylene tube was interposed between the prosthesis and the descending aorta. At 3 months later, repair of the tetralogy of Fallot was carried out, anastomosing the prosthetic tube for the left PA to the pulmonary trunk (Fig. 2A). Postoperative catheterization at 14 months after repair found the systolic right ventricular pressure to be 48 mmHg, with no obvious pulmonary stenosis. This patient is currently doing very well at 77 months after the repair without major complaint in respiration.

Patient 2. A female baby was born with cardiac murmur, and diagnosed as having tetralogy of Fallot, on echocardiography. At the age of 8 months, the patient was referred to our institution. The left PA was found to course abnormally on angiography, and a diagnosis of PA sling was confirmed using electron-beam computed tomography scanning (Fig. 1B). Tracheal bronchus was coexisting. No respiratory symptoms had been noted. Primary repair of the cardiovascular malformations was carried out at 1 year and 9 months old. The left PA was extensively dissected behind the trachea, divided at its origin, translocated anteriorly, and anastomosed to the pulmonary trunk in front of the left main bronchus (Fig. 2B). The tetralogy of Fallot was repaired using a transannular patch. Postoperative catheterization at 14 months after the repair found no obstruction at the translocated left PA, and the systolic right ventricular pressure was 34 mmHg. The patient is doing very well at 17 months after the procedure with...
Fig. 1. Preoperative imaging.
Tracheal stenosis, as well as abnormal branching in the right upper bronchus, was clearly illustrated by bronchography in Patient 1 (A). In Patient 2, electron-beam computed tomography scanning confirmed the slender left pulmonary artery originating from the right pulmonary artery and coursing behind the trachea (B). In this patient, tracheal bronchus was also present, and the left superior caval vein was persistent.

Fig. 2. Schema of the operative procedure for reconstruction of the left pulmonary artery and the right ventricular outflow tract. In Patient 1, the heterologous pericardial roll tube, emplaced for the reconstruction of the left pulmonary artery, was directly anastomosed to the pulmonary trunk. The right ventricular outflow tract was augmented using a pedicled autologous pericardial patch (A). In Patient 2, the left PA was extensively dissected behind the trachea, divided at its origin, translocated anteriorly, and anastomosed to the pulmonary trunk in front of the left main bronchus with no use of prosthetic material. The right ventricular outflow tract was augmented using a transannular patch (B).