Simultaneous complex single ventricle palliation and tracheoplasty for heterotaxy syndrome

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Abstract
Although isomerism of the bronchial tree is an integral part of heterotaxy syndrome, the association of congenital tracheal stenosis is rare in this group of disorders, and it has not yet been thoroughly described in the literature. This condition is potentially life-threatening and precludes single ventricle palliation. This report presents the case of a 5-month-old infant with symptomatic congenital tracheal stenosis, functionally univentricular heart and extracardiac total anomalous pulmonary venous connection (TAPVC). The condition was successfully treated with bidirectional Glenn anastomosis, central pulmonary artery plasty, repair of TAPVC and slide tracheoplasty.

Keywords: CHD heterotaxy • Tracheal surgery • Paediatric

INTRODUCTION
The surgical management of infants with congenital tracheal stenosis and a complex univentricular heart is very challenging. Airway obstruction has a significant impact on the Fontan circulation. Extracardiac total anomalous pulmonary venous connection (TAPVC) is very common in patients with right atrial isomerism, and it is also a key factor in their poor survival.

CASE REPORT
An infant was born by Caesarean section at 41 weeks gestational age and weighed 2320 g. Prenatal echocardiography disclosed heterotaxy syndrome, which was confirmed postnatally; showing a single atrium, single right ventricle, pulmonary atresia, patent ductus arteriosus (PDA), pulmonary coarctation, supracardiac TAPVC and a bilateral superior vena cava (SVC). The PDA was kept open by continuous infusion of prostaglandin E-1.

Three-dimensional computed tomography done at 2 months of age detected discrete tracheal stenosis (Fig.1). She was intubated due to respiratory distress at 4 months of age. Bronchoscopy confirmed tracheal stenosis with a complete cartilaginous tracheal ring. Cardiac catheterization and angiography showed an acceptable mean pulmonary artery pressure of 13 mmHg, a pulmonary artery resistance of 1.2 Wood units, a pulmonary artery (Nakata) index of 253 mm²/m², a ventricular end-diastolic volume of 282% of normal and a ventricular ejection fraction of 41%.

The patient (weight 4.5 kg) underwent bidirectional Glenn (BDG) anastomosis, central pulmonary artery plasty, repair of TAPVC and slide tracheoplasty under cardiopulmonary bypass (CPB) 6 days after the cardiac catheterization. The central pulmonary artery was divided at the insertion of the PDA, and the ductal tissue was completely resected. The confluence of the central pulmonary artery was reconstructed by end-to-end anastomosis, and was enlarged with left SVC as BDG anastomosis. The posterior pericardium was opened just above the common pulmonary vein during the cooling phase. The common pulmonary vein connected to the right superior vena cava (RSVC) 5 mm proximal to the RSVC-atrial junction, and lay directly behind the single atrium. The target core temperature of 20°C was achieved, and the heart was arrested by cold blood cardioplegia and the circulation was stopped. The posterior wall of the atrium was incised from the pulmonary venous orifice in the RSVC through a right atriotomy, and was extended parallel to the common pulmonary vein. A longitudinal incision on the common pulmonary vein was extended to the junction of the left pulmonary veins. The divided edge of the posterior atrial wall was anastomosed to the opening in the common pulmonary venous confluence using continuous 7-0 polypropylene sutures (Prolene; Ethicon Inc. Somerville, NJ, USA). Then the aorta was declamped, and the patient was rewarmed. The proximal portion of the RSVC was ligated because of its small diameter.

Subsequently, the anterior trachea was exposed. The midpoint of the stenosis was divided, and longitudinal incisions were created over the stenotic segment anteriorly and posteriorly on the distal and proximal tracheal stumps, respectively. Sliding oblique anastomosis was performed with interrupted 6-0 polydioxanone sutures (PDS; Ethicon Inc. Somerville, NJ, USA). The patient was successfully weaned from CPB with systemic saturation of 80% and pulmonary artery pressure of 15 mmHg. The total bypass time was 351 min, aortic cross-clamp time was 35...
min, and deep hypothermic circulatory arrest time was 19 min. The patient was extubated on postoperative day 10. Two weeks later, she presented with symptoms of high airway obstruction. Rigid bronchoscopy was performed on postoperative day 36, which confirmed the short-segment of restenosis with tracheomalacia. A tracheostomy was performed to stent the stenosed segment on postoperative day 43. At 12 months of follow-up, she weighed 7 kg, and still needed a tracheostomy tube. However, her usual respiratory condition was stable without ventilator support. Her oxygen saturation was 86%. The follow-up echocardiography showed unobstructed flow within the BDG and no signs of pulmonary venous obstruction. She was waiting for Fontan completion with or without additional tracheal procedure.

**DISCUSSION**

Congenital tracheal stenosis is rarely associated with a functionally univentricular heart, and it may have a significant impact on the outcome [1, 2]. Our institutional policy for congenital tracheal stenosis and cardiac anomalies is to surgically correct both conditions concomitantly to obtain a better outcome, and to avoid using prosthetic material, which carries a significant risk of infection after tracheal surgery [3]. PDA stent may be an attractive option as the first stage palliation. However, PDA stent is not available in routine practice in Japan. Therefore, concomitant BDG anastomosis and slide tracheoplasty were performed in the current case. Successful Glenn circulation is highly dependent on an unobstructed pulmonary venous pathway as well as the respiratory conditions. TAPVC associated with heterotaxy syndrome has remained a challenge with high mortality rate. Yun and associates reported the efficacy of the primary sutureless repair technique for TAPVC associated with right atrial isomerism [4]. We also have adopted the sutureless repair as a primary procedure for TAPVC with right atrial isomerism since 2006 [5]. However, tracheoplasty requires dissection of the posterior mediastinal space around the bronchi, which poses the risk of massive bleeding after primary sutureless repair. Hence, an intra-atrial direct anastomosis for TAPVC was performed as a workaround to avoid torsion and rotation of the pulmonary veins. Although, the patient underwent the most aggressive surgical therapy, her postoperative hemodynamic and respiratory condition was stable.

In conclusion, concomitant tracheoplasty and complex single ventricle palliation are thus considered to be indicated when adequate pulmonary arteries and pulmonary vascular resistance are confirmed by cardiac catheterization.

**Conflict of interest:** none declared.

**REFERENCES**


*Figure 1:* Three-dimensional computed tomography shows discrete tracheal stenosis (white arrow) and the joining of the pulmonary venous confluence (*) with the RSVC (yellow arrow). (LSVC: left superior vena cava; PDA: patent ductus arteriosus).