Concomitant Slide Tracheoplasty and Cardiac Operation for Congenital Tracheal Stenosis Associated With VACTERL

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The association of congenital tracheal stenosis and tracheoesophageal (TE) fistula is rare. Here, we report 2 patients with tracheobronchial stenosis (complete cartilage ring) involving the lower trachea and right bronchus. Both patients had associated VACTERL (vertebral anomalies, anal atresia, cardiovascular anomalies, TE, renal, and limb defects) congenital cardiac defects and tracheal diverticula after repair of the TE fistula in type C esophageal atresia.

Concomitant repair of congenital cardiac defects and a slide tracheoplasty with elimination of the diverticula were performed successfully.


Tracheoesophageal (TE) fistula and esophageal atresia are developmental defects of the primitive embryonic foregut [1]. Tracheomalacia and tracheal diverticula are commonly observed after surgical repair of a TE fistula, which relates to the redundant membranous portion of the trachea [2]. However, the association between tracheal stenosis and TE fistula has not been well described [3]. Here, we report 2 infants with congenital heart defects and TE fistulas, with complete cartilage rings in the lower trachea and proximal right bronchus, which implies incorrect separation of the foregut, resulting in a stenotic trachea and distal TE fistula.

Technique

Patient 1

A male infant was referred to our hospital because of difficulties with weaning him from a ventilator at age 2 months. He received TE fistula repair for type C esophageal atresia when he was 1 day old. A chest roentgenogram and echocardiography revealed left lung agenesis, a ventricular septal defect, and agenesis of the left pulmonary artery. Several episodes of severe desaturation and CO2 retention occurred. Computed tomography revealed stenosis of the lower trachea and that the endotracheal tube slipped into a blind pouch.

Surgical intervention was performed through a median sternotomy with standard cardiopulmonary bypass. The trachea became a diverticula and a stenotic lower tracheal segment at the midportion. The diverticula extended to near the carina and the suture. Clips from a previous operation were found at the distal end, which confirmed that it was the residual segment of a previous TE fistula. Complete cartilage rings were disclosed when slide tracheoplasty was performed from the lower tue to the right bronchus (Fig 1).

His ventricular septal defect, atrial septal defect, and infundibular stenosis were corrected simultaneously. The aorta clamping time was 72 minutes, and the duration of the cardiopulmonary bypass was 174 minutes. The patient was separated from the cardiopulmonary bypass smoothly. The wound was primarily closed without an irrigation system, and broad-spectrum antibiotics were prescribed for prophylaxis after operation.

The infant was gradually weaned from the ventilator and successfully extubated 3 weeks after the operation. He was discharged 5 weeks after the operation. He has been alive and well for 2 years and 8 months, without supplemental oxygen or mechanical ventilation.

Patient 2

A male infant was born in our hospital with a prenatal diagnosis of congenital heart disease. After birth, esophageal atresia with distal TE fistula was disclosed. Right ventricle hypoplasia, a ventricular septal defect, and pulmonary stenosis were also revealed. He received TE fistula repair. The fistula rose from the carina, and the gap from the proximal atrial esophageal end to the TE fistula

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was approximately 1 cm. He was extubated 2 weeks after this operation.

However, he was intubated again when he was 6 months old because of respiratory failure and a para-influenza virus infection. Computed tomography and bronchoscopy showed the stenotic lower trachea and his right bronchus orifice (Fig 2). An operation was arranged because the infant could not be weaned off the ventilator after 1 month of treatment.

During the operation, a blind pouch and complete tracheal rings from the lower trachea to the right bronchus were confirmed. Slide tracheoplasty and a bidirectional Glenn shunt were performed smoothly. The aorta cross-clamp time was 12 minutes, and the total duration of the cardiopulmonary bypass was 172 minutes. Extubation was successful 5 days after the operation. Postoperative bronchoscopy was performed 2 weeks after the operation and confirmed a patent right main bronchus. He was then transferred to the pediatric gastrointestinal section because of cholangitis.

The patient was followed up at an outpatient clinic, without ventilator support. Computed tomography 7 months after the operation showed a widely patent lower trachea and bronchus. However, a subglottic stenosis related to previous intubation was also found.

Comment

Esophageal atresia and TE fistula are relatively common birth defects (approximately 1 in 3,000 live births). Of the several malformation associations involving TE fistula, the most clearly described is the VACTERL association, which includes vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb malformations [1].

Tracheal stenosis in patients with TE fistula has rarely been reported [3]. Here, we show that tracheal stenosis should be carefully evaluated when difficult extubation occurs in patients with previous surgical repair of TE fistula in addition to the relatively common tracheomalacia. The TE fistulas in our 2 patients were related to the VACTERL association, and we did not find tracheal stenosis in our patients with an isolated TE fistula. These findings suggest that tracheal stenosis should be more vigorously investigated in TE fistula related to VACTERL association.

Type C esophageal atresia usually occurs when the distal TE fistula rises near the carina portion. The operation revealed that the stenotic segment starts at the orifice of the tracheal diverticula (blind pouch). It mimics the cartilage and membranous portion of the lower trachea and assumes 2 separate circular configurations (Fig 2B). Le Bret and colleagues [3] reported a patient with tracheal stenosis and TE fistula that was also associated with a residual tracheal pouch.

In conjunction with the findings of their reported patient, we assert that tracheal stenosis tends to occur in the lower trachea, below the origin of the TE fistula, which becomes a pouch after being repaired. The presence of a long pouch might be a sign of a lower tracheal stenosis. We hypothesized that the stenotic trachea and the upper part of the fistula would be difficult to distinguish in a “normal trachea” during an esophageal atresia operation. Thus, the TE fistula was ligated at the usual position near the carina instead of at the true origin of the fistula. Furthermore, if the endotracheal tube enters the pouch, then effective ventilation would be severely compromised. The intensivist should be aware of this lethal complication and promptly treat it. In our

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Fig 1. (A) Intraoperative photograph shows the upper trachea separating into a stenotic trachea (yellow loop) and diverticula (blue loop). (B) The transection of the stenotic lower trachea shows a complete cartilage ring with a lumen of approximately 2 mm.
experience, computed tomography is a noninvasive and helpful way to confirm the diagnosis.

Although resection and end-to-end anastomosis are feasible for treating a short-segment tracheal stenosis [4], we adopted a slide tracheoplasty as our procedure of choice, which also compensates for the presence of a right bronchial stenosis [5]. In the operations we outlined, the proximal trachea was opened posteriorly, and the distal trachea was opened anteriorly. The tracheal pouch was opened, and the orifice was incorporated into the new trachea to avoid formation of a residual pouch postoperatively. The proximal tracheal flap was advanced to cross the stenotic orifice of the right bronchus. Because the trachea is a potential source of contamination, we performed a slide tracheoplasty during a partial cardiopulmonary bypass and avoided using cardiotomy suction when the airway was opened. After the trachea was repaired, the intraoperative bronchoscope was used to confirm the patency and ventilation pressure to ensure that no air was leaking.

The heart was then opened, and the congenital heart defect was repaired in a standard manner. Although the operation in the first patient was complicated by left lung agenesis and the second patient required a bidirectional Glenn shunt as a single-ventricle palliation, simultaneous slide tracheobronchial plasty and congenital heart operations were performed successfully.

In conclusion, although rarely reported, tracheal stenosis is found in patients with a TE fistula and a VACTERL association. Simultaneous slide tracheoplasty and congenital heart repair are effective treatments in this situation.

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References


