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Complex surgical treatment of congenital tracheal stenosis with associated unilateral lung agenesis

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ABSTRACT

Background: The combination of congenital tracheal stenosis (CTS) and unilateral pulmonary agenesis is rare and fatal. Especially, right lung agenesis is associated with dextrocardia and often causes tracheal alignment disorder. The mortality is estimated with 12.5–65%. Slide Tracheoplasty (ST) has been improved the prognosis, however, the postoperative course of CTS with single lung remains unclear. Here, we describe the postoperative challenging situation with bronchial intervention and other devices.

Case presentation: A 9-month-old girl with CTS, right lung agenesis, dextrocardia, and bilateral superior vena cava underwent ST anterior to the aortic arch. On postoperative day 50, tracheostomy was performed for long-term airway pressure support. Six months after the ST, she showed severe bronchomalacia at the bifurcation of the lobar bronchi, which was sandwiched between the descending aorta and left pulmonary artery. Direct bronchopexy and mobilization of left pulmonary artery resolved the severe bronchomalacia. Additionally, the kinked trachea caused a tracheal ulcer and frequent accidental decannulations. The ulcer was made above the aortic arch by contact with the tracheal cannula. The customized Montgomery T-tube solved both problems and enabled the stable management for airway. Her condition eventually improved, and she was discharged home after 14 months of hospitalization.

Conclusion: This case presented a very rare and critical condition. This is the first report to address the severe condition after ST anterior to the aortic arch. Surgical interventions or other devices for treatment are thought to be modified according to each condition.

1. Background

The combination of congenital tracheal stenosis (CTS) and unilateral pulmonary agenesis is rare, but these disorders are often associated with each other [1–5]. A patient with this combination sometimes grows without any symptoms, but airway infection or other triggers can suddenly worsen the patient's condition, and this can be fatal. Slide tracheoplasty (ST) is a standard surgical approach for this combination, and it has been shown to improve mortality. In the combination of CTS and right lung agenesis, tracheal kinking and the complex of major vessels can result in a poor prognosis. Therefore, ST has been modified recently [6]. We experienced a rare case of postoperative bronchomalacia with right lung agenesis, dextrocardia, and bilateral superior vena cava. Additionally, the kinked trachea caused a critical problem with airway management. This was a challenging situation after modified ST for CTS, and we describe the patient's clinical course.

2. Case presentation

An infant girl was born at the gestational age of 41 weeks, and her weight was 3500 g. Her right lung was found to be absent just after birth (Fig. 1A). However, initially, she had no symptoms, and her growth process was normal. At 9 months of age, an airway infection suddenly induced life-threatening respiratory deficiency. Computed tomography (CT) identified dextrocardia and bilateral superior vena cava (Fig. 1B). She was intubated and transferred to our institution. On arrival at our institution, her arterial pH was 6.84 and pCO2 was 105 mmHg, indicating severe obstructive pulmonary disorder. Emergency extracorporeal membrane oxygenation (ECMO) was performed, and her life was saved. Under ECMO, tracheoscopy revealed congenital tracheal stenosis with complete circular cartilaginous rings. On the

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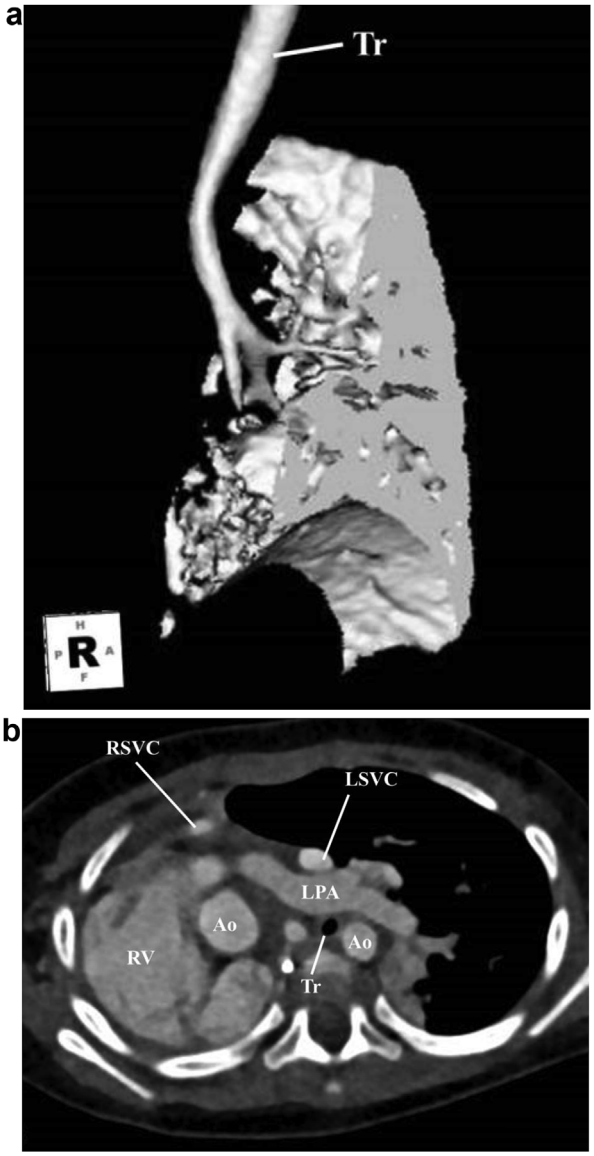

Fig. 1. A. 3D multidetedtor computed tomography at birth. (Tr = trachea.). B. Contrast enhanced computed tomography just before slide tracheoplasty. (Tr = trachea, Ao = aorta, RSVC = right superior vena cava, LSVC = left superior vena cava, LPA = left pulmonary artery, RV = right ventricle.)

following day, ST was performed. Operative findings showed that the length of stenosis was 28 mm (from the middle of the trachea to near the bifurcation of the upper and lower lobar bronchi). The trachea was bent posteriorly, and this was considered to be associated with her dextrocardia. Thus, tracheal anastomosis was performed anterior to the aortic arch. An endotracheal tube (4 mm) was inserted through the anastomotic site, and ECMO was weaned during the surgery. The anatomical narrow opening of the lobar bronchus required postoperative airway pressure support, and it did not allow extubation. On postoperative day 50, tracheostomy was performed.

At 6 months after the surgery, bronchomalacia at the bifurcation of the lobar bronchus gradually worsened and required high airway pressure (over 20 cmH2O) (Fig. 2A). CT revealed that the bifurcation of the lobar bronchi was sandwiched between the descending aorta and left pulmonary artery (LPA) (Fig. 2B). At that time, direct bronchopexy was performed with nonabsorbable sutures (4-0 PRONOVA®, Ethicon, USA), and stitch sutures were placed in the upper and lower lobar bronchi and the bronchial bifurcation. To prevent LPA damage by the string of bronchopexy, mobilization of the LPA with suspension by nonabsorbable sutures (4-0 PRONOVA®) was simultaneously performed (Fig. 2C). The bronchus was suspended anteriorly, and the LPA was suspended caudally. Besides, the left superior vena cava (LSVC) and upper lobe of the left lung were anterior to the trachea, and the sutures caused LSVC kink and blood pressure decrease. The sutures were passed through the sternum and were tied under bronchoscopy. For the suture space, we collapsed a part of the covering upper lobe into the left pleural cavity. Postoperative ventilation-perfusion mismatch was noted; however, the condition gradually improved in a few weeks. Bronchomalacia at the bifurcation considerably resolved, and continuous airway pressure with ventilation decreased to 6 cmH2O (Fig. 2D). Mild bronchomalacia remained, and it required positive airway pressure support (6 cmH2O).

A few days after tracheostomy, the tip of tracheostomy tube contacted with the tracheal wall near the aortic arch, and made ulcer (Fig. 3A). Tracheal cannula was changed to length-flexible tube (Tracheoflex®, Rusch, Germany) to avoid the contact ulcer. But the length of the tube in the trachea was very short, about 20 mm, and that easily led to accidental-decannulation. Taken together, we planned to customize Montgomery T-tube and prevent the tracheal contact and the decannulation. The shape was the upper limb of the tube was closed, and the posterior side of the lower limb was cut off to avoid contact with the tracheal wall above the aorta (Fig. 3B). The 1st design of the T-tube did not provide efficient positive airway pressure, probably because of the dead space in the oral side of the T-tube. As such, the T tube was modified some times, and the upper limb was filled with silicone for eliminating dead space (Fig. 3C). It finally provided the stable maintenance of airway, no contact with aortic arch (Fig. 3D), and efficient positive pressure lung ventilation. She survived without any brain damage, and she was discharged home after 14 months of hospitalization.

3. Discussion

The combination of CTS and unilateral pulmonary agenesis is critical and has a high mortality rate (12.5–65%) [1–5]. For this combination, ST has been shown to be a feasible procedure. In contrast, ST for single lung patients, especially those with right lung agenesis, has been found to be highly associated with postoperative tracheobronchomalacia. CTS with a single lung often involves anomaly of the heart or large vessels in the mediastinal space. The cardiovascular complexity affects the trachea through compression and results in tracheal kinking. In CTS with a single lung, a previous study reported the effectiveness of ST anterior to the aorta with concomitant tracheopexy [6]. However, there are no previous reports which refer the postoperative complications in detail after ST in CTS with a single lung.

Several studies have recently addressed the efficacy of bronchopexy for severe bronchomalacia, and the method of bronchopexy varies depending on the underlying disease [7,8]. Most reports have mentioned acquired bronchomalacia, which is associated with extrinsic compression and repositioning. The most popular method of bronchopexy is suspension of the target bronchus at nearby tissues. The other surgical options for bronchomalacia include resection of the malacic part and placement of an internal or external stent.

In the present case, the congenital kinked trachea became almost straight after ST, and thus, we did not simultaneously perform tracheopexy. The bifurcation of the lobar bronchi was already sandwiched between the descending aorta and PA before ST, but it appeared to be gradually compressed by those vessels. This is probably because the positon of the bronchi was changed by suspension in ST. Therefore, bronchomalacia worsened in a few months. In the second operation, tracheopexy was not effective for bronchomalacia. Additionally, the target bronchus was anatomically narrow, and there was limited space around the bifurcation of the bronchus. Thus, resection of the malacic part or stent placement appeared to be quite difficult. Furthermore, the LPA was located immediately above the target bronchus and the LSVC was near the target site. To prevent damage to these vessels from