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CASE REPORT

A long-term survival case of tracheal agenesis: management for tracheoesophageal fistula and esophageal reconstruction

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Abstract Tracheal agenesis is a very rare disorder which leads to severe respiratory disorders immediately after birth. Reports are very limited on long-term survival cases. We report here a long-term survival case with Floyd's type I tracheal agenesis. During the neonatal stage, the patient underwent abdominal esophageal banding to substitute esophagus for trachea and transection at the cervical esophagus with esophagostomy. Subsequently, airway management was difficult due to a fragile tracheoesophageal fistula, but the fistula was conservatively treated and stabilized with the patient's growth. This patient is a very rare case in whom oral feeding was achieved after esophageal reconstruction using a gastric tube. For this case, we describe mainly (1) the management method of the tracheoesophageal fistula and (2) esophageal reconstruction without thoracotomy.

Keywords Tracheal agenesis · Tracheoesophageal fistula · Esophageal reconstruction · Floyd's type I · Long-term survival

Abbreviations

PEEP	Positive end expiratory pressure
IMV	Intermittent mandatory ventilation
CPAP	Continuous positive airway pressure

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Introduction

Tracheal agenesis is rare among neonatal disorders. Its prognosis is very poor because its clinical symptoms are usually severe, and many cases die soon after birth [1–3]. We report herein a long-term survival case of Floyd's type I tracheal agenesis. The patient's esophagus was used as a tracheal substitute. Then he was provided strict conservative respiratory management for the fragile tracheoesophageal fistula and underwent esophageal reconstruction with a gastric tube. He remains alive at age 4 years 6 months.

Case report

An infant boy with a birth weight of 2,550 g was born by Cesarean section at 37 weeks and 4 days of gestation. Antenatal polyhydramnios was noted. The baby had no audible cry after birth. Respiratory stability was not achieved by mask ventilation, so he was resuscitated by intubation (later determined to be esophageal intubation and not endotracheal intubation). A transverse colostomy was performed at age 0 day for imperforate anus. When injection of milk was initiated through the gastric tube at age 4 days, the patient's respiratory condition deteriorated. At age 5 days, esophagography showed the trachea and bronchi (Fig. 1a). Bronchoscopy revealed the epiglottis but no glottis, and the distal segment in the direction of the trachea ended blindly (Fig. 1b). A fistula was observed extending from the esophagus to the trachea, and the patient was diagnosed with tracheal agenesis. Endoscopy and 3D-CT showed a tracheoesophageal fistula 5 cm distal to the esophageal inlet, and the tracheal bifurcation was approximately 1.5 cm from the fistula. The patient was diagnosed with Floyd's type I tracheal agenesis. The right

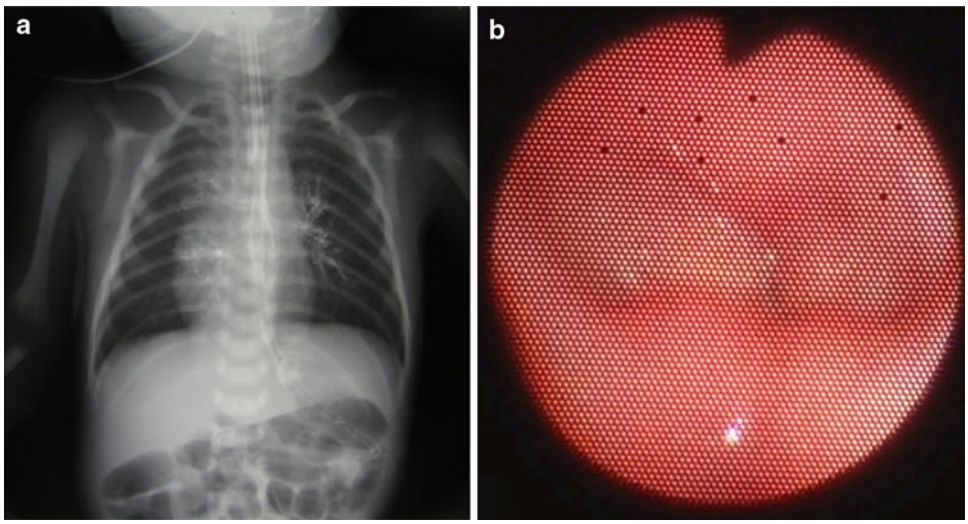

Fig. 1 **a** Esophagography at age 5 days. Contrast enhanced imaging of the esophagus showed trachea and bronchi. **b** Bronchoscopy showed the epiglottis but the distal portion ended blindly with no trachea

upper lobe bronchus arose in an area 0.8 cm from the fistula and formed a tracheal bronchus (Fig. 2). The tracheoesophageal fistula was narrow. Bronchoscopy revealed a tracheoesophageal fistula originating from the anterior esophageal wall, and the diameter of the fistula changed markedly with respiration. The fistula was a pinball size during expiration (Fig. 3a).

The patient had other malformations such as an imperforate anus, atrial septal defect, hypospadias, and polydactyly. At age 13 days, he underwent laparotomy and abdominal esophageal banding at the upper gastroesophageal junction with gastrostomy. Bronchoscopy showed that the tracheoesophageal fistula was tracheomalacia-like. In such a case, the fistula can narrow easily and tracheal secretion clearance can be impaired. Thus, expiratory impairment can frequently develop due to salivary drainage. Therefore, resection of the cervical esophagus was performed at age 53 days, and a salivary fistula was created with the cranial segment of the esophagus. A tracheal cannula was inserted into the caudal portion of the esophagus where a tracheal fistula was made. Thereafter the patient continued to have respiratory problems but his respiratory condition improved gradually. The mode of mechanical ventilation was changed from intermittent mandatory ventilation (IMV) to continuous positive airway pressure (CPAP). However, he had repeated bronchitis, air trapping developed due to swelling around the tracheoesophageal fistula, and his lungs showed emphysema-like changes. The patient had cyanotic attacks and bradycardia, and expiratory rib cage compression was used to promote exhalation.

Subsequently, positive end expiratory pressure (PEEP) was slowly lowered. The patient was able to manage without the ventilator for progressively longer periods beginning at age 18 months. He initially needed PEEP because of the collapsing tracheoesophageal fistula. Other

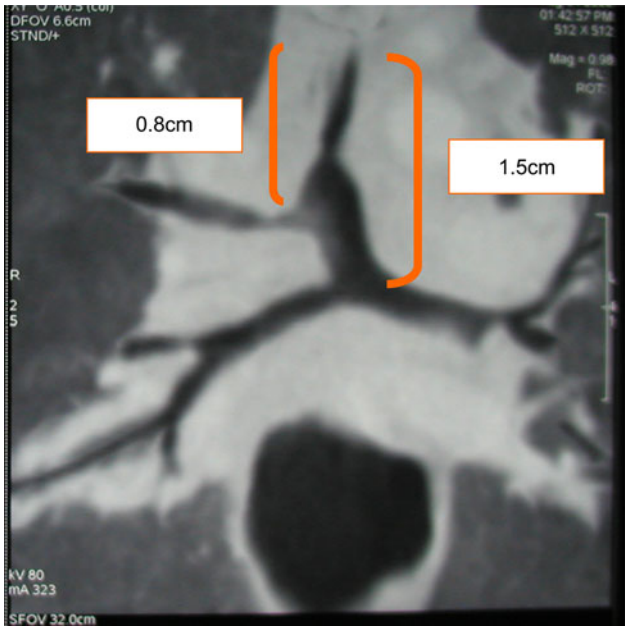

Fig. 2 3D-CT findings. Tracheal branching from the esophagus was narrowed at the superior portion. The tracheal bifurcation was 1.5 cm from the fistula. The right upper lobe bronchus arose in an area 0.8 cm from the fistula and formed a tracheal bronchus

procedures were also considered such as resection of stenosis of the fistula, end-to-end anastomosis, and placement of an external stent [4, 5]. The esophagus, tracheal fistula, and trachea were periodically examined by endoscopy. Occlusion of the fistula during straining and coughing gradually improved due to maturing tracheal cartilage and enlarging fistula diameter as the patient grew (Fig. 3b). The patient was in a stable respiratory condition and successfully being weaned from ventilator.

His respiratory condition was improving. Therefore, at age 3 years 2 months, the esophagus was reconstructed using a whole gastric tube by the retrosternal route to

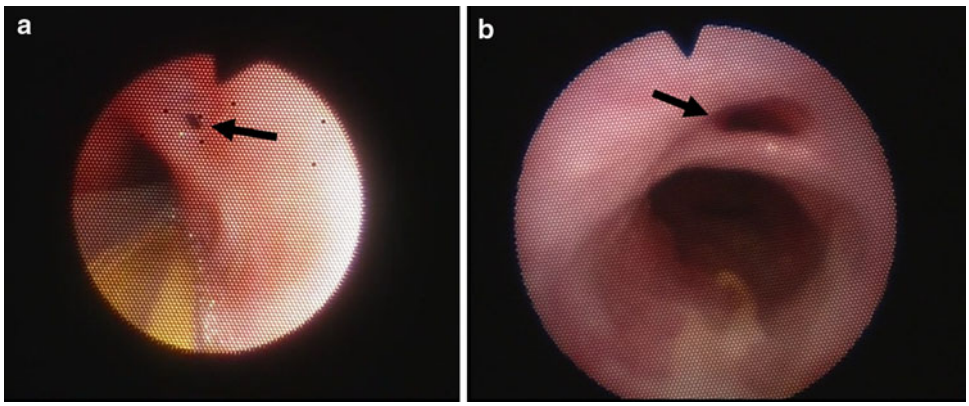

Fig. 3 Bronchoscopic findings. **a** At age 13 days, bronchoscopy revealed a tracheoesophageal fistula originating form the anterior esophageal wall, and the diameter of the fistula changed markedly with respiration. The image shows a very small diameter during expiration (*arrow*). **b** At age 2 years 10 months, the tissue had become sound around the opening of the tracheoesophageal fistula and changes in diameter were minimal during respiration (*arrow*)

enable oral feeding (Fig. 4a, b). Presently, the patient is receiving training in oral feeding. He has normal growth in height and weight and very good intellectual development. He uses the mechanical ventilator only at night, but he can manage without it most of the time.

Discussion

Tracheal agenesis was first reported by Payne et al. in 1900. Since then, over 140 cases have been reported in the literature. The majority of the cases died at birth or shortly thereafter. Most cases do not survive long term because of difficult airway management and serious associated disorders. In our search of the literature, we have found only four other long-term survival cases to date [2, 4–6]. Our patient had Floyd's type I tracheal agenesis, and airway management was difficult due to a fragile tracheoesophageal fistula. Conservative treatment was performed for the tracheoesophageal fistula, and the fistula became stable with the patient's physical growth. This patient is a very rare case in whom oral feeding was achieved after tracheal reconstruction using a gastric tube [6].

Floyd's classification of tracheal agenesis is the most commonly used system [3]. It divides the condition into three main types according to the form of the fistula, tracheoesophageal or bronchoesophageal fistula: type I (11% of all published tracheal agenesis cases), type II (61%), and

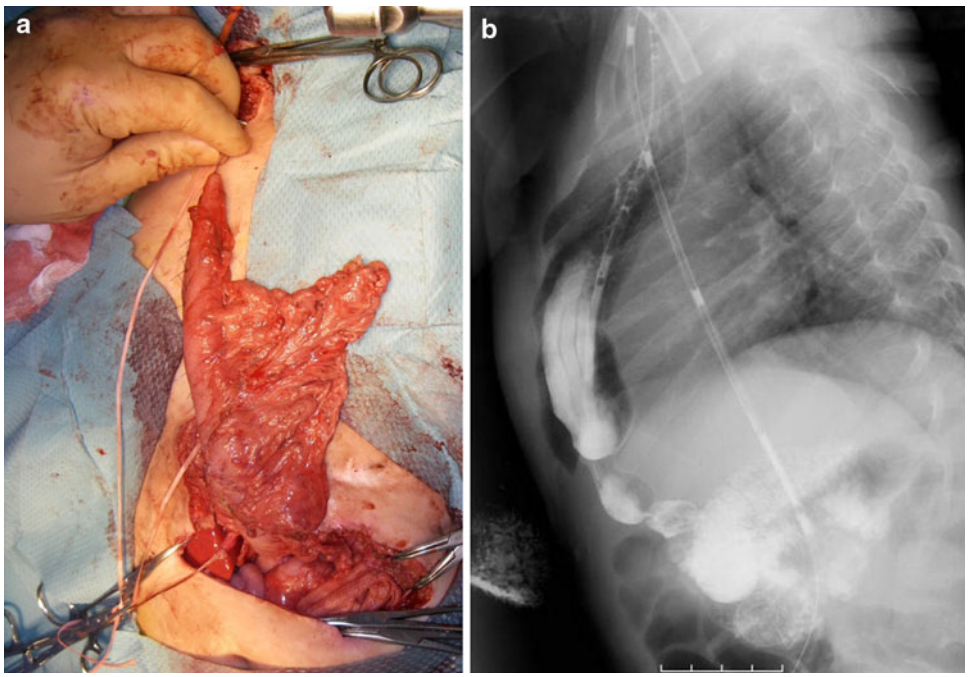

Fig. 4 **a** Esophageal reconstruction. A whole gastric tube was used for the esophageal reconstruction. Blood supply was from the gastroepiploic artery. **b** Oral contrast enhanced image taken after esophageal reconstruction

type III (23%) [7]. Types II and III have bronchi which directly formed a fistula with the esophagus, so the airway can easily occlude. In these types, tube insertion is more difficult into the tracheoesophageal or bronchoesophageal fistula and airway maintenance is also more difficult compared to type I. Most type I patients also die in the early neonatal period because of ventilatory failure due to esophageal collapse or accumulation of secretion.

In general, tracheal agenesis was suspected because of a history of antenatal polyhydramnios, absence of the first cry at birth, possibility of mask ventilation, and impossibility of endotracheal intubation, and was diagnosed by esophagography depicting the trachea, bronchoscopy showing tracheal atresia. If mask ventilation is possible and the infant can be successfully resuscitated immediately after birth, the next steps are to perform (1) abdominal esophageal banding for palliative surgery to substitute esophagus for trachea, (2) transection at the cervical esophagus with esophagostomy, and (3) gastrostomy for enteral feeding [8–11].

After palliative surgery is performed for substituting esophagus for trachea, the tracheoesophageal fistula should be treated in one of three major ways depending on the position and size of the fistula: (1) conservative management by follow-up, (2) direct insertion of the tip of an endotracheal tube into the fistula, or (3) surgical treatment of external stent placement after resection of the stenotic area and anastomosis. In the patient of the present report, intubation of the fistula was not selected because difficulties in its management were expected, including accidental extubation and sacrifice of the right tracheal bronchus. The tracheoesophageal fistula could easily collapse (Fig. 3a). In our case, ventilation improved with gradual mechanical stability of the tracheoesophageal fistula and with the patient's physical growth (Fig. 3b). Thus, his respiratory condition also stabilized.

Hiyama et al. placed an endotracheal tube in a fistula for a type II case and had successful respiratory management by CPAP ventilation. They performed esophageal reconstruction with colonic interposition between the residual esophagus and stomach [6]. We performed esophageal reconstruction on our patient at age 3 years 2 months to enable oral feeding. To our best knowledge, there have been only two cases, including our patient, who underwent esophageal reconstruction for tracheal agenesis. If small intestine or colon interposition for esophageal substitution is used in the reconstruction, there could be an ulcer on the intestinal side with a high risk for ischemia. Therefore, a gastric tube was used in our patient (Fig. 4a, b). In our route of reconstruction, intrathoracic anastomosis was not selected because respiratory failure was expected from swelling and collapse of the fistula due to surgical manipulation around it. Since a gastric tube was used, the suturing in the antethoracic route was thought to be impossible because of the distance. Therefore, the retrosternal route was selected. This route made it unnecessary to do any manipulation near the tracheoesophageal fistula during the surgery, and esophageal reconstruction was possible without thoracotomy. However, extracorporeal membrane oxygenation (ECMO) was set up so that it could be initiated any time an unexpected situation developed during the surgery.

Presently, our patient is being trained in oral feeding, and he has been gradually learning to swallow saliva. He had normal physical growth in height and weight due to enteral feeding and good intellectual and mental development. We await future developments in laryngotracheal transplant or regenerative medicine for treating his speech.

A written informed consent was obtained from the parents for submitting the manuscript. This study has been performed with the ethical standards laid down in the 1964 Declaration of Helsinki.

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