Surgical management of tracheal agenesis

Tracheal agenesis is a rare congenital malformation that usually is fatal. This report describes our experience in two such cases. In both cases, the diagnosis was suspected at birth, because the patients had respiratory distress without an audible cry and were difficult to intubate. A gastrostomy and banding of the abdominal esophagus provided effective initial stabilization, in conjunction with respiratory management. The first patient also had complex cardiac malformations, and the infant died of cardiac failure 1 week after birth. The second infant, who had tracheal agenesis with a proximal tracheoesophageal fistula and a bronchoesophageal fistula, was managed successfully. At 9 months of age, a tracheotomy was performed, a long T tube was inserted to maintain the airway patency beyond the proximal tracheoesophageal fistula, and the patient was discharged. At 3 years of age, esophageal reconstruction was performed with a colonic interposition graft. The patient is thriving and developing normally at 4 years of age. Diagnosis at birth and maintenance of airway patency are essential for successful management of tracheal agenesis. Initial surgical interventions are palliative but lifesaving. Subsequent management focuses on improving the quality of life and allowing swallowing and speech.

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Tracheal agenesis is a rare malformation that has been reported to be uniformly fatal. This disorder was first described in 1900 by Payne. The majority of patients survived for only a few hours, and the diagnosis was established post mortem. Recently, several surgical approaches have been suggested, although definitive treatment has not been established and long-term survivals are few. This article describes our experience with two cases of tracheal agenesis.

Case reports

CASE 1. A male infant was born with a birth weight of 2826 gm. Cyanosis and absence of crying were noted. Attempted endotracheal intubation with a 3 mm diameter tube was unsuccessful because the tube would not advance beyond the vocal cords. Ventilation with 100% oxygen administered with a bag and a mask produced a slight improvement in heart rate and color, but stridor and respiratory distress continued. The infant was transferred immediately to our hospital. The infant was intubated with a 2.5 mm tube, which was difficult to advance beyond the distal subglottic area, after which the clinical condition improved substantially. A suction tube introduced into the stomach through the intubation tube aspirated gastric contents (Fig. 1, A). Furthermore, positive-pressure ventilation produced abdominal distention. These findings strongly suggested an esophagotracheal communication. During the initial surgical procedure, the abdominal esophagus above the cardia was bound by simple wrapping with a nonsplitting 6 mm diameter artificial prosthesis of expanded polytetrafluoroethylene mesh. Then a gastrostomy was created with a No. 12 mushroom tube. The infant was supported with mechanical ventilation but died 7 days after the operation.

Autopsy disclosed multiple congenital anomalies (Fig. 1, B). The upper part of the esophagus ended in a blind pouch, and the larynx was hypoplastic just below the vocal cords and communicated with the distal esophagus. The trachea was absent between the subglossal lesion and the carina, and the bronchi arose separately from the esophagus (type III by Floyd's classification). Additional findings included a single atrium and single ventricle, patent ductus arteriosus, preductal coarctation of the aorta, bicuspid pulmonary valve, bilobular right lung, and aspiration pneumonia.

CASE 2. A male infant with a birth weight of 2185 gm and a history of hydramnios was born by spontaneous vaginal delivery at 35 weeks of gestation. Cyanosis and absence of crying were noted after birth. Respiratory distress developed immediately after birth but improved after endotracheal intubation with a 3 mm diameter tube. During the next 24 hours, the clinical condition remained satisfactory, but excessive oral secre-
Fig. 1. Chest roentgenogram (A) and postmortem photograph (B) of a neonate with tracheal agenesis who died at 1 week of age (case 1). Chest roentgenogram shows that a suction tube introduced through the intubation tube led into the stomach. The upper part of the esophagus ended blindly, and the hypoplastic larynx communicated with the distal esophagus. The bronchi arose independently from the esophagus 4 cm above the gastric cardia. The trachea was absent between the subglossal lesion and the carina.

Insertions were noted. Insertion of a nasogastric tube was difficult but finally was achieved by inserting the tube along the endotracheal tube. At this point, little doubt existed that the patient had a tracheoesophageal fistula and esophageal atresia. Forty-eight hours after birth, respiratory distress developed after positive-pressure ventilation produced abdominal distention.

The infant was transferred to our hospital. The same abdominal esophageal banding as in case 1 was performed and a gastrostomy was created. The patient's condition immediately improved. Laryngoscopy showed a normal larynx without a tracheoesophageal cleft. Esophagography revealed a cervical tracheoesophageal fistula (Fig. 2, A). Bronchography through the endotracheal tube demonstrated tracheal atresia between the cervical fistula and the carina and bronchi that arose from the esophagus (type II by Floyd's classification). To maintain airway patency, we inserted an endotracheal tube into the esophagus with its tip just above the bronchoesophageal fistula. The infant remained on assisted ventilation with a continuous positive airway pressure of 3 cm H$_2$O and received tube feedings through the gastrostomy for approximately 9 months. At 10 months of age, a tracheotomy was created 1 cm below the vocal cords in the intact upper part of the trachea, and a long T tube (5 cm) was inserted through the tracheotomy. The patient was now able to maintain adequate ventilation while spontaneously breathing room air. When he was 2 years of age, we divided the esophagus just above the tracheal entrance and exteriorized the proximal orifice. When he was 3 years of age, we reconstructed the esophagus by means of a colonic interposition graft. Now, at 4 years of age, the patient's development is normal both neurologically and physically. He can speak several words when he closes the orifice of the T tube, and he can eat a small amount.

Discussion

The etiology of tracheal agenesis is unknown. However, because the esophagus and the respiratory tract have a common origin, congenital abnormalities of the trachea usually have associated anomalies of the esophagus. Tracheal atresia is often associated with esophageal communication with the trachea or main stem bronchi, and aeration of the lung becomes possible by ventilation through the esophagus. However, most patients with tracheal atresia die within minutes or hours of birth because of the difficulties in diagnosing and treating this unusual anomaly. Tracheal agenesis should be suspected in any
Fig. 2. Esophagogram (A) and schema of operative procedures (B) in a patient with tracheal agenesis (case 2). Esophagogram shows a cervical tracheoesophageal fistula (TEF), tracheal agenesis between the cervical fistula and the carina, and bronchi arising from the esophagus.

neonate with a history of hydramnios, absent crying, respiratory distress, and difficulty in intubation.\(^4\)\(^8\) Mask ventilation can assist in resuscitation, and the administration of oxygen via esophageal intubation can be lifesaving in patients in whom tracheal agenesis is suspected. In our patients, esophageal intubation through the communication between the proximal trachea and the cervical esophagus provided temporary access to the lungs and permitted time for a definitive diagnosis, which usually can be made by chest roentgenography or esophagography. So that regurgitation of gastric fluid and abdominal distention owing to positive-pressure mechanical ventilation can be prevented, gastrostomy and esophageal banding in the abdominal portion should be performed. Fonkalsrud, Martelle, and Maloney\(^7\) have proposed isolating the esophagus below the fistula to prevent aspiration of gastric fluid through the bronchoesophageal fistula, whereas Altman, Randolph, and Shearin\(^8\) have recommended gastric division to avoid entering the chest. Esophageal banding and gastrostomy are equally effective, technically more simple, and also avoid entering the chest. Cervical esophagostomy is not necessary in critically ill infants, because saliva does not cause severe aspiration pneumonia in patients with esophageal atresia.\(^11\) However, the pharyngeal sump tube is necessary to prevent aspiration pneumonia until cervical esophagostomy is performed.

Our first patient died of cardiac failure resulting from uncorrectable cardiac anomalies. Patients with tracheal agenesis frequently have other congenital malformations including cardiac, genitourinary, respiratory, gastrointestinal, and other anomalies.\(^12\)\(^13\) These anomalies often are the major causes of death in neonates with tracheal agenesis.

Once the emergency therapy has been completed and the patient is in stable condition, the long-term management provides an even more challenging problem. Although Sankaran and colleagues\(^14\) have reported on two patients with a short segment of tracheal agenesis who underwent direct tracheal anastomosis, no patient
with a long atretic segment, impossible to anastomose directly, has survived. Mansfield\textsuperscript{15} reported that a direct anastomosis of the trachea and bronchi is possible so long as the tracheal stenosis does not exceed half the total tracheal length, but the lengths of tracheal agenesis in our cases were more than half the total tracheal length. When the esophagus is used as a part of the airway without internal or external support, distal pneumonia, mucous plugging of the fistula, or air trapping results, usually with death in short order. Fortunately, our second patient had isolated tracheal agenesis. In this case, the tip of the esophageal T tube was positioned near the bronchoesophageal fistula and continuous positive airway pressure was used in early infancy. A distal tracheostomy, which is impractical in most cases, increases susceptibility to respiratory infection because the air conduit is short.\textsuperscript{16} A tracheostomy in the intact proximal trachea and a long T tube in the esophagus greatly facilitate airway maintenance. This is the first patient with a long atretic segment of trachea who has survived and achieved normal physiologic and neurologic development. This technique should be considered for patients who have an intact segment of proximal trachea and a tracheoesophageal fistula. This procedure permits speech when the orifice of the T tube is closed.

A feeding route is necessary for long-term survival in patients with tracheal agenesis. Feeding has to be exclusively through the gastrostomy until the respiratory tract has been divided completely from the digestive tract. Gastrointestinal continuity can be restored via esophageal reconstruction, as in case 2.

A variety of tracheal prostheses have been developed for the patients who require tracheal replacement. For example, Beall and coworkers\textsuperscript{16} have reported extensive animal studies, but limited clinical experience, using Marlex mesh (Bard Implants, Billerica, Mass.), and Borrie, Redshow, and Dobbinson\textsuperscript{17} have reported extensive experience using silicone rubber combined with sub-terminal Dacron suture cuffs. Because the procedure is performed on young children, growth and development of the trachea are problematic, and homologous tracheal replacement is preferable. Bechara, Tabek, and Berman\textsuperscript{18} have attempted to create a new trachea from a main left bronchus and have one case of 48-hour survival. At present, no form of tracheal replacement is satisfactory. Although definitive repair will be difficult, we believe that homologous tissue offers the best hope for a satisfactory tracheal prosthesis. A flexible, noncollapsible airway is unlikely to allow growth and development.

REFERENCES