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Right Pneumonectomy Syndrome: Report of Two Cases

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Abstract: We report herein the cases of two infants who developed right pneumonectomy syndrome, both of whom were born with gross C-type esophageal atresia (EA/TEF), and a hypoplastic right lung arising from the lower esophagus, being a bronchopulmonary foregut malformation (BPFM). Appropriate and well-timed treatments for a variety of sequelae primarily caused by the mediastinal shift must be considered after right pneumonectomy in early childhood.

Key Words: right pneumonectomy syndrome, bronchopulmonary foregut malformation (BPFM), mediastinal shift

Introduction

Pneumonectomy is rarely performed in early childhood but it is usually followed by many late sequelae caused by the mediastinal shift, especially after right pneumonectomy. In 1988, Stolar et al. reported a fatal complication after right pneumonectomy in a newborn baby with a congenital cystic adenomatoid malformation of the lung (CCAM) and called special attention to the right pneumonectomy syndrome.[1] We report herein the cases of two infants who developed right pneumonectomy syndrome following surgery for gross C-type esophageal atresia (EA/TEF) with bronchopulmonary foregut malformation (BPFM).

Case Reports

Case 1

A 1804-g male infant born at 37 weeks of gestation was transferred to our hospital on the day of birth on July 1, 1985, with a diagnosis of EA/TEF. A primary repair of the EA/TEF and a gastrostomy were performed the same day, but 1 week later a hypoplastic right lung arising from the lower esophagus was confirmed by esophagogram, angiography, and pulmonary scintigram (Fig. 1). Thus, division of the bronchoesophageal fistula (BEF) followed by a right pneumonectomy was performed on the 22nd day of life and the patient was discharged from hospital 1 month postoperatively.

He was re-admitted 3 months later due to severe respiratory distress, and was treated in the ICU on a ventilator. An aortopexy for tracheomalacia, a celiotomy for intestinal volvulus, a regastrostomy for tube feeding, and an antireflux procedure for gastroesophageal reflux (GER) were also subsequently performed. However, due to frequent bouts of severe pneumonia, he was kept intubated and ventilated in the ICU for 15 out of 20 months, and finally died of sepsis and DIC at the age of 2 years.

At autopsy, in addition to pneumonia, stenosis of the trachea by compression and scarring, a long remnant of TEF, and a blind pouch of the rudimental right bronchus were confirmed (Fig. 2).

Case 2

A 2578-g female infant born at 39 weeks of gestation by normal vaginal delivery after an uneventful pregnancy was transferred to our hospital on the day of birth on August 1, 1988 with a diagnosis of EA/TEF. A gastrostomy and primary repair of the EA/TEF were performed on the first and second days of life respectively. Again, a hypoplastic right lung arising from the distal esophagus was confirmed by esophagography, CT, angiography, and pulmonary scintigram (Fig. 3). A right pneumonectomy was performed 2 months after birth and at the same time the pleural cavity was replaced with a $3 \times 5 \times 1.5$ cm silicon block

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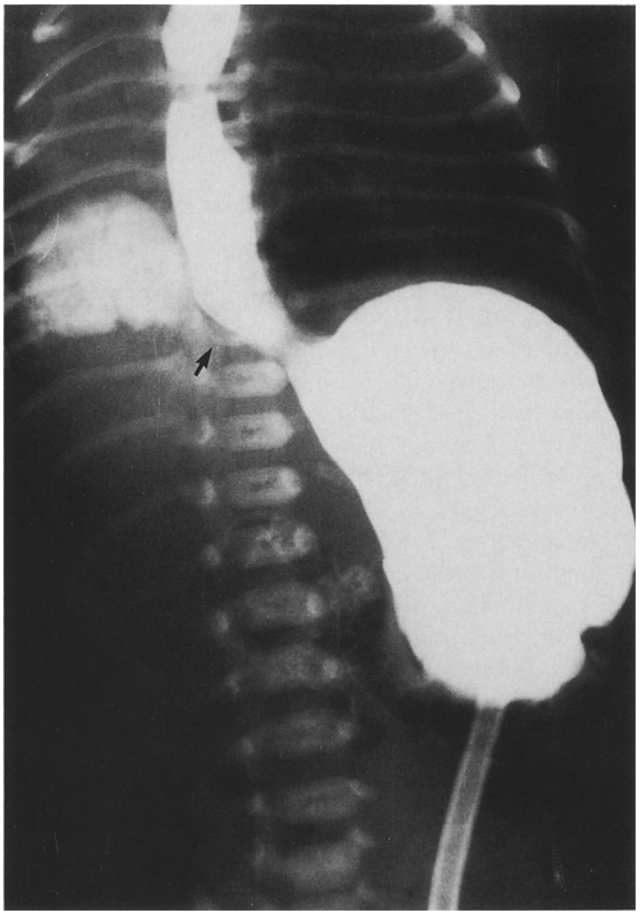


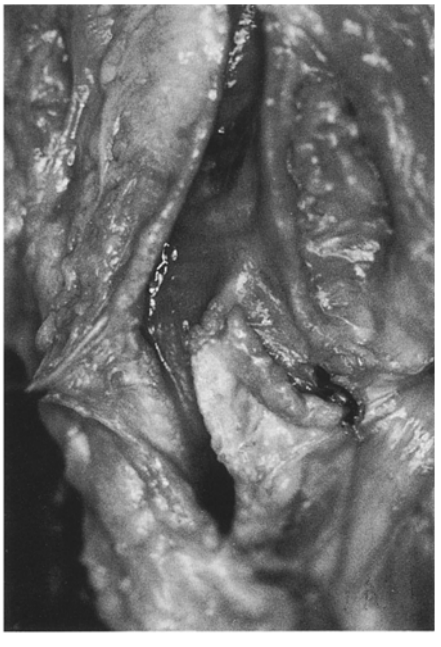
Fig. 1. The esophagogram of case 1 taken after primary repair of a gross C-type esophageal atresia (EA/TEF). A hypoplastic right lung arising from the lower esophagus can be seen. The *black arrow* shows the bronchoesophageal fistula (BEF)

to prevent mediastinal shift. Nevertheless, 2 months later, she was readmitted due to severe respiratory distress and ventilated in the ICU. A celiotomy for adhesive ileus and a gastrostomy were performed at 6 months of age, and an aortopexy with a pleuroplasty to relieve tracheal stenosis was done 2 weeks later. Thereafter, the condition of the infant gradually improved and she was successfully weaned from controlled ventilation to continuous positive airway pressure (CPAP). At the age of 8 months, tracheostomy was performed for long term CPAP, after which she underwent three more operations for ileus. She was finally discharged from hospital at the age of 1 year and 3 months.

On January 11th, 1990, at the age of 17 months, she was readmitted due to severe respiratory distress and was again ventilated in the ICU. However, this time her recovery was more rapid and she was discharged from the ICU 2 weeks later. Thereafter, lysis of intestinal adhesions, an antireflux procedure with pyloroplasty, and a CV catheter replacement for venous access were performed. During the following 3 years she suffered from frequent episodes of bronchial asthma-like upper respiratory infections which were successfully treated by medical therapy.

Discussion

Schaffer reported a higher mortality rate with right lung agenesis than with left lung agenesis and attributed this difference to the greater movement and rotation of the heart and mediastinal contents.[2] During early childhood, the mediastinal shift which occurs after



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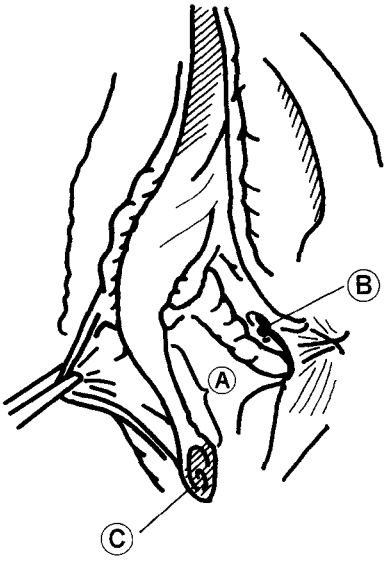

b

Fig. 2. **a** Photograph and **b** schema of the respiratory tract from the autopsy of case 1 (posterior view). *A*, stenotic part of the trachea; *B*, remnant of the TEF; *C*, a blind pouch in the right bronchus

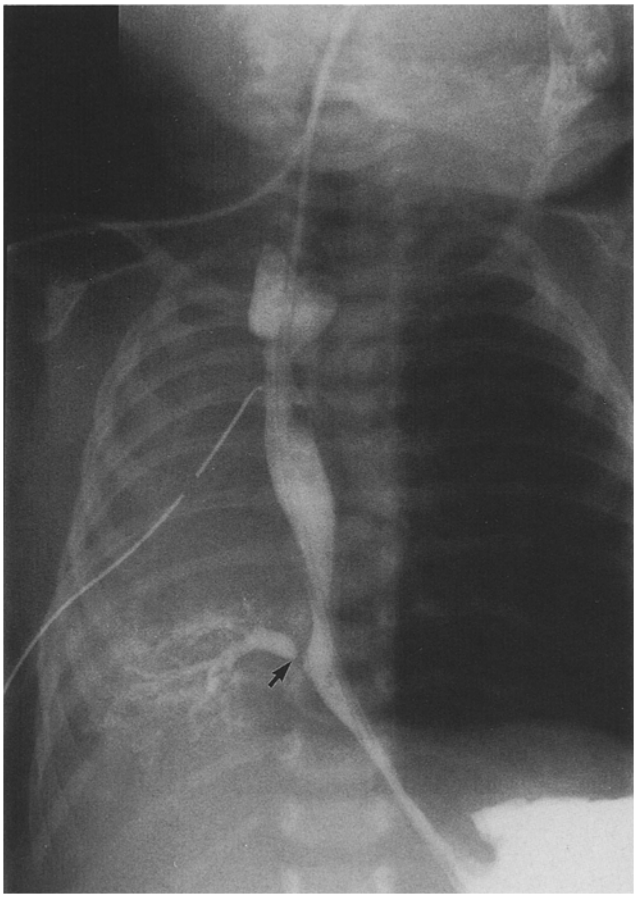

Fig. 3. The esophagogram of case 2 taken after repair of the EA/TEF. Again a hypoplastic right lung arising from lower esophagus can be seen. The *black arrow* shows the bronchoesophageal fistula

right pneumonectomy may cause severe respiratory distress due to tracheomalacia. Furthermore, deviation and angulation of the esophagus will cause esophageal dilatation and abdominal distension, which eventually compress the trachea and suppress respiratory movement. The negative pressure of the pneumonectomized thorax may also cause GER.

According to various reports, aortopexy, vascular bypass formation, and tracheoplasty have been performed to relieve these symptoms, but these treatments when performed alone have failed to relieve the symptoms in most cases.[3–5] Johnson et al.[6] and Powell et al.[7] reported the successful use of a prosthesis after pneumonectomy to prevent mediastinal shift and its related complications in children.

Right pneumonectomy syndrome has also been reported in adult patients, and therefore thoracic surgeons have now started using injections of non-absorbable gas such as SF-6 after lobectomy or pneumonectomy.[8,9] Moreover, Kosloske et al. recently reported successful treatment for right pneumonectomy syndrome utilizing tissue expander replacement.[10] We believe that some kind of prosthesis should definitely replace the excised lung to prevent mediastinal shift and overinflation of the contralateral lung and to prevent deformity of the affected thorax. This, in turn, would enhance the effects of other surgical interventions.

Our experience of these two patients led us to conclude:

1. Pneumonectomy in early childhood may cause fatal complications, especially right pneumonectomy.
2. At the time of right pneumonectomy, replacement with some type of prosthesis or an injection of non-absorbable gas should be seriously considered to prevent mediastinal shift, overinflation of the contralateral lung, and deformity of the affected thorax.
3. Aortopexy for tracheomalacia, tracheostomy for long-term intubation, gastrostomy for the prevention of abdominal distension and feeding, and CV cannulation to provide venous access for long-term medications should be performed at the most appropriate age and in the right order.
4. If the above procedures should fail to relieve the symptoms, major surgery such as vascular bypass or tracheo-bronchoplastic surgery should be considered.

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