

Isolated Congenital Tracheobiliary Fistula

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Background: Congenital tracheobiliary fistula is a rare developmental anomaly with a persistent communication between the biliary system and the trachea. **Characteristics:** A 7-day-old baby with severe respiratory distress and aspiration pneumonia. **Outcome:** Tracheobiliary fistula identified on bronchoscopy. Open surgical excision of fistula was followed by improvement. **Message:** This condition should be considered in the differential diagnosis of intractable aspiration pneumonia.

Keywords: Aspiration pneumonia, Bronchoscopy, Neonate, Respiratory distress.

Congenital tracheobiliary or bronchobiliary fistula is a rare anomalous communication between the carina or the main bronchus and the biliary system [1]. To date, only few cases have been reported in the literature.

CASE REPORT

A 7-day-old full term male baby, born to a primi-gravida, by normal vaginal delivery and with immediate cry, had an episode of vomiting immediately after a breastfeed given at an about 4-5 hours of birth. This was followed by increasing respiratory distress which was managed initially at a local hospital, and then referred to our center on day 2 of life. At admission, the baby was in severe respiratory distress with heart rate of 160-170/min, respiratory rate of 80/min with retractions and saturation of 60-70% on room air; arterial blood gas showed mixed

acidosis and the baby was mechanically ventilated. Chest X-ray showed left upper lobe opacity, and intravenous antibiotics were started in view of neutrophilic leucocytosis and positive C-reactive protein. The baby improved gradually and weaned off from invasive to noninvasive mode of ventilation; however, the baby continued to have intractable coughing, persistent respiratory distress, chronic respiratory acidosis, persistent opacity on the right side and high FiO₂ requirement. Computed tomography chest done showed consolidation in the right upper lobe and apico-posterior segment of left upper and lower lobes, and gastrograffin study showed no upper GI congenital anomaly. The baby was then referred for further management to a pediatric surgeon who performed bronchoscopy, which showed a third opening (*Fig. 1*) at the level of the carina. The opening was cannulated with an X-ray positive catheter,

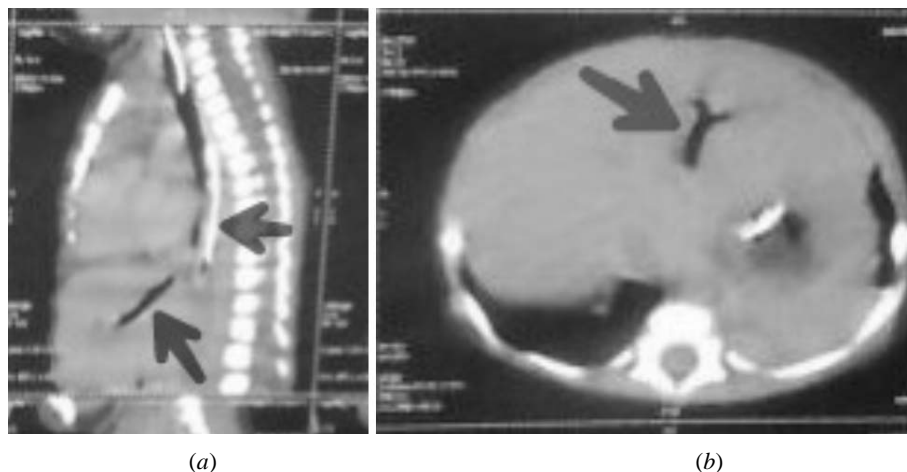


Fig. 1 CECT chest reveals evidence of an airfilled fistulous tract (arrows) between the carina and left intrahepaticbiliary tract.

and a fistula crossing the diaphragm and communicating with the left hepatic duct was found. The diagnosis of hepatobiliary fistula was made on bronchoscopy. A right posterolateral thoracotomy was done, fistula was identified, and ligated at both the ends and divided. Tracheobiliary fistula was about 6 cm long arising from the carina between right and left ostium of main bronchi passed through esophageal hiatus and inserted at the left side of bile duct. The histopathology showed a tubular structure lined by squamous epithelium at places by columnar epithelium, muscular wall shows few mucous glands and cartilages with few mononuclear cells infiltrates. Small muscular ventricular septal defect with Persistent Pulmonary Hypertension was identified on color Doppler evaluation. The baby improved after surgery and discharged home after 10 days of hospitalization. The baby is now gaining weight, taking direct breastfeeding well at 6 months of age.

DISCUSSION

Previous reports suggest that majority of tracheobiliary fistula originate either from the right main bronchus (42%) or carina (42%). In our patient, it was arising from the carina between right and left ostium of main bronchi, passed through esophageal hiatus, and inserted at the left side of bile duct. In our case, the specimen obtained only from the proximal portion of the fistula, and histological examination corresponded with previous reports.

The underlying pathogenesis of congenital fistula is still not clear. However, two possible embryological mechanisms have been suggested: (a) fusion of an anomalous bronchial bud with an anomalous bile duct; (b) duplication of the upper gastrointestinal tract [1].

The fistula was diagnosed at an early age (median age, 32 d; range, 12 h to 6 y) in most cases (87%) [1-3]; only four cases were diagnosed in adults [4-7]. Recurrent episodes of aspiration, two episodes of extubation failure due to aspiration, and bilious secretion present in the endotracheal tube gave us a clinical clue. The most common presenting symptom in previously reported cases was respiratory, including cough, dyspnea, cyanosis, bilious sputum, and pulmonary infection.

However, these symptoms are nonspecific, and other pathologic conditions (including tracheoesophageal fistula, gastroesophageal reflux, gastrointestinal obstruction, and aspiration pneumonia) should be included in the differential diagnosis [8].

Bronchoscopy has been the most commonly used (52%) method for diagnosis, followed by hepatobiliary scintigraphy (Tc99m HIDA scan) and bronchography. Recently, two cases using multidetector CT with multiplanar and 3D reformations to delineate the fistula have been reported by Gunlemez, *et al.* [9]

The possibility of congenital anomalies of respiratory tract should always be considered when evaluating a neonate with persistent pneumonia and/or recurrent aspirations.

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