CHAPTER 68

CONGENITAL DISEASES OF THE LUNG

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Congenital anomalies of the lower respiratory tract include anomalies of the intrathoracic airways, including the trachea, bronchi, and lower airways. In this section, we will examine congenital lower respiratory tract diseases in detail.

TRACHEA ANOMALIES

Tracheal Agenesis and Atresia

Agenesis of trachea is a rare and mortal congenital anomaly characterized by interrupted or absent development of trachea[1]. The prevalence of tracheal anesthesia is less than 1: 50,000 and the male / female ratio is 2: 1. Overall, 52% of cases are associated with preterm labor and about half of cases are associated with polyhydramnios[2].

Anatomically examined in 3 subtypes[3]:

Type I; proximal agenesis and presence of distal tracheo-esophageal fistula,

Type II; complete absence and presence of normal bifurcated bronchi,

Type III; The two main bronchi occur independently of the esophagus.

Congenital Tracheal Stenosis

Congenital stenosis of trachea is a rare anomaly characterized by complete cartilage rings and the absence of membranous trachea[4]. These circular "O" rings can be elongated for a variable length of the trachea. Short segment stenosis covers <50% of the trachea length; long segment stenosis may extend for the entire trachea (and sometimes beyond the carina)[5, 6].

Etiology and pathogenesis are unknown, but they are the result of a developmental abnormality. It may be associated with other tracheobronchial anomalies such as tracheal bronchus, trifurcation of carina, pulmonary agenesis or hypoplasia. The majority of patients are associated with left pulmonary sliding.

Symptoms may begin at birth with shortness of breath, need for intubation and resistance to mechanical ventilation. Lower respiratory tract infection can expose the condition and cause severe symptoms. Symptoms are: stridor or noisy breathing, intercostal and supraclavicular retractions, episodes of cyanosis and recurrent infections. Endoscopic evaluation may increase symptoms in some patient. In infants with tracheal stenosis, respiratory symptoms can be particularly severe; Mechanical ventilation attempts may not be sufficient for ventilation as the stenosis may involve the entire trachea or bronchi. Tracheostomy is not helpful in these situations. If mechanical ventilation cannot be applied, the patient can only be rescued by extra corporeal membrane oxygenation (ECMO) and surgical treatment must be urgently performed. On the contrary, some patients do not compromise their daily living activities and may only show symptoms during exercise[7].

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For asymptomatic patients who do not have any of the above features in advanced imaging, elective surgical resection or observation is a reasonable option. The decision should be shared and decided in detail with the family about the potential benefits and risks of each approach.

Surgery must be evaluated in three aspects: First, surgery is curative and is not usually associated with significant complications. Second, individuals with BPS will have a moderate risk of developing infections later in life, especially if the lesion is an ILS. When infection or respiratory symptoms develop, surgery becomes urgent and an increased risk of postoperative complications (eg air leak, effusion or pneumonia) occurs compared to elective surgery performed in asymptomatic patients. Third, imaging cannot always distinguish between BPS and CPAM or hybrid lesions, and there will be a risk of developing complications (infection or malignant degeneration) if CPAM or hybrid lesions are left in place. [48]

Bronchopulmonary sequestration (BPS) is the differential diagnosis of CPAM. On prenatal ultrasound, BPS appears as a well-defined, homogeneous, echodeneous mass. BPS is generally not connected to the tracheobronchial system, apart from CPAM, and its blood supply is provided by an abnormal systemic artery. Systemic artery source can be demonstrated with color flow Doppler. [49]

In addition to BPS, the differential diagnosis of CPAM includes congenital diaphragmatic hernia, bronchogenic cyst, congenital lobar emphysema, and pneumatoceles that can be sequelae of localized pulmonary interstitial emphysema, especially bacterial pneumonia, which may occur in patients on mechanical ventilation. These disorders can generally be distinguished from CPAM based on the radiographic appearance and clinical history.

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