Pulmonary Malformation

22

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Introduction

From a surgical viewpoint, congenital pulmonary adenomatous malformation (CPAM) and bronchopulmonary sequestration (BPS) are the most relevant inborn pulmonary malformations. However, in rare cases, bronchogenic cysts and congenital lobar emphysema also need surgical intervention. CPAM (Fig. 22.1) is considered to represent a hamartomatous change in the tertiary bronchioles, with an incidence of 1 in 25,000–35,000 live births. BPS has an estimated incidence of 0.15–1.7 % in the general population and is defined as a region of lung parenchyma that lacks a normal connection to the tracheobronchial tree and possesses an anomalous systemic blood supply (Fig. 22.2). There is a high proportion of mixed-type lesions, suggesting that CPAM and BPS may have the same developmental ancestry.

Other indications for thoracic procedures in newborns are inborn aberrations such as esophageal atresia, cardiac malformations, and hernia diaphragmatica.

The technical issues related to posterior lateral thoracotomy and lobar or segmental resection are the same as those in adults. Thoracoscopic treatment (Fig. 22.3) is feasible even in newborns and is well established, particularly for extralobar sequestration and lobectomy.

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Figure 22.1

Congenital pulmonary adenomatous malformation

Figure 22.2

Bronchopulmonary sequestration

Figure 22.1

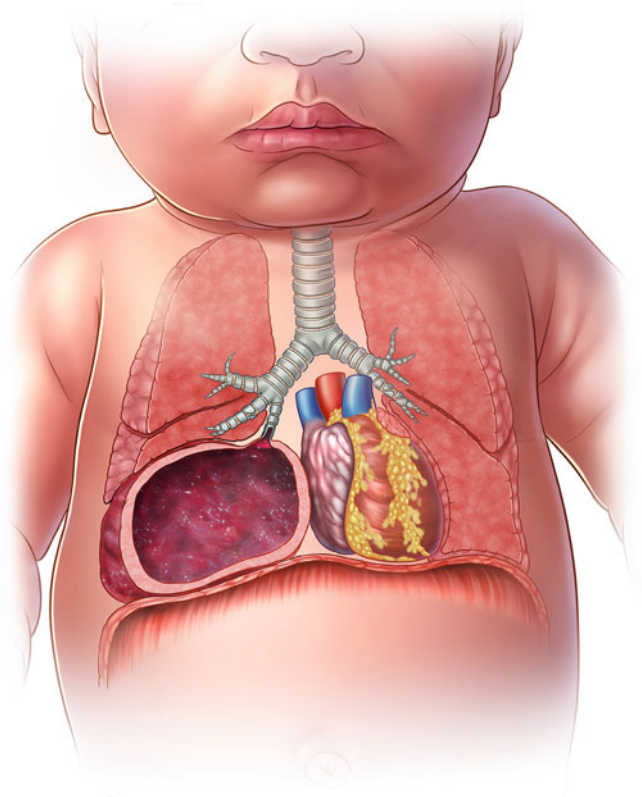


Figure 22.2

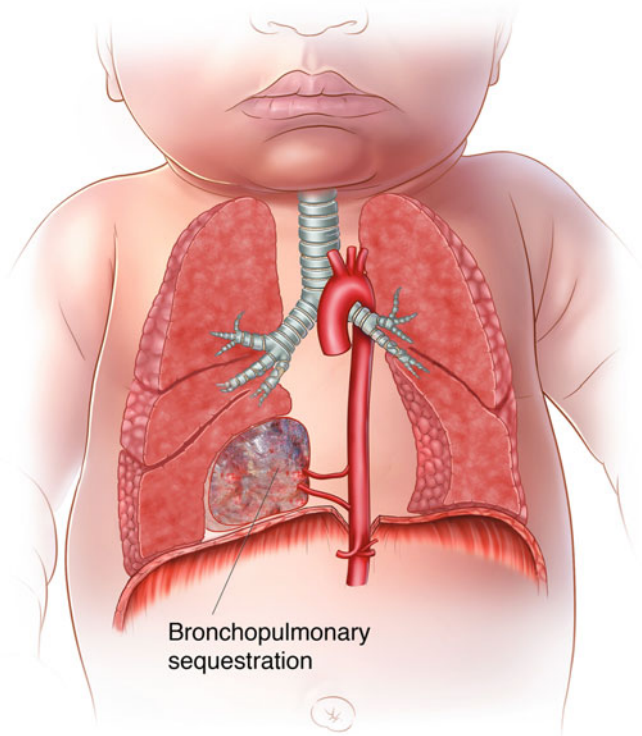
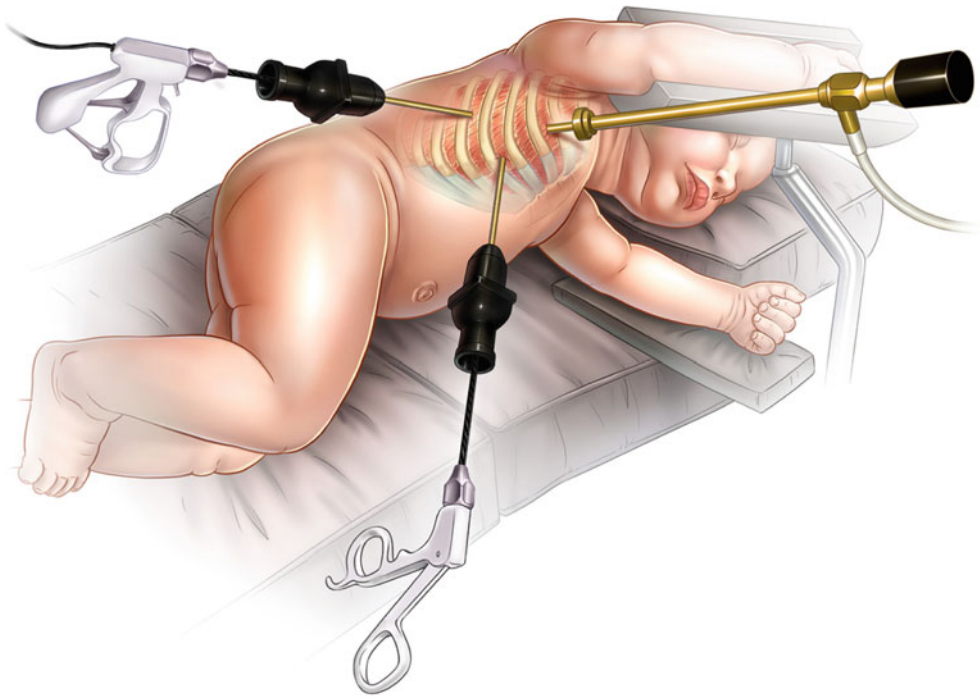


Figure 22.3

Thoracoscopy in infants

Figure 22.3



History

The first description in the English language literature of CPAM—also called CCAM (congenital cystic adenomatous malformation)—was written by Ch'in and Tang in 1949.

Since 1977, the histopathologic classification penned by JT Stocker has been widely quoted, although, by itself, it does not affect treatment decisions, has no prognostic value, and is not even sufficient to provide an accurate diagnosis.

Pulmonary sequestrations were first described in 1861 by the Austrian anatomist Rokitansky as *accessory pulmonary lobes*, but were renamed *pulmonary sequestrations* by Pryce in 1946. The first parenchymal lung lesion to be detected antenatally was reported in 1975 in Australia. Subsequently, large series reporting early outcomes were published in the 1990s.

Indications

There is no doubt that surgical intervention is necessary for postnatally symptomatic cystic or parenchymal pulmonary lesions; however, in the unborn fetus, a range of therapeutic options is available. Ultrasonically defined macrocystic disease may be aspirated or shunted using a thoracoamniotic shunt, or even excised with fetal surgery or intranatal techniques, such as the EXIT (ex utero intrapartum treatment) maneuver. Early thoracotomy to excise expanding cystic lesions is safe, is nearly free of complications, and allows appropriate growth of the normal compressed lung.

Most macrocystic lesions, stable lesions, and lesions with a fetal mediastinal shift come to surgery. Sometimes, prenatally observed cystic lesions become smaller, and this spontaneous shrinkage is prognostically favorable. However, true resolution of antenatally diagnosed lesions is the exception; therefore, they must be evaluated by frequent CT or MRI scans postnatally.

In 2009, Stanton et al. performed a systematic review and meta-analysis of 41 reports describing 1,070 patients. They found that 505 neonates survived into infancy without surgery and only 16 of them (3.2 %) became symptomatic. The authors showed that for all ages, elective surgery was associated with significantly fewer complications than emergency surgery. The onset of symptoms occurred within the first 7 months of life. Therefore, resection must be planned before 10 months of age; most authors advise elective resection at the age of 6 months. Because lung tissue develops in the first year of life, this period of maturation should be utilized for the expected compensatory capacity of the rest of the lung tissue after resection.

When considering surgical intervention, one must weigh the risk of postoperative complications against the lifelong risk of leaving lesions in situ. There are many cases describing a correlation between CPAM and malignancy (rhabdomyosarcoma, adenocarcinoma, and bronchoalveolar carcinoma); however, there were no reports of malignancy in the series collected by Stanton et al.

So far, there is no identifiable prognostic feature in postnatal life suggesting the need for intervention, although there have been attempts to correlate size with outcome at the antenatal stage. Furthermore, there is no consensus regarding the indications for resecting small symptom-free lesions. Davenport et al. summarized: *We continue to advocate excision of all "significant" lesions, even if asymptomatic, although this can be difficult to define. . .we believe that the accumulated hazards of pulmonary infection and a risk of malignancy later in life outweigh the risks of early, elective excisional surgery*.

This statement reflects the perception of most authors over the past years.

Surgical Principles

The anatomic guidelines are the same in infancy and in adulthood. General principles of thoracotomy and pulmonary resection apply to both children and adults, except that diminutive size, associated lesions, and unique pathologic entities require special consideration.

The first 2 weeks after birth are essential for the newborn to adapt to life outside the uterus. During this short period, the newborn will stabilize and become more resistant. However, early surgical resection for symptomatic CPAM or BPS within 1 month of age is proven to be safe. For an elective resectional procedure, the age of 5–7 months may be ideal, because such an early resection allows compensatory lung growth before lung maturation.

Posterolateral Thoracotomy

The patient is placed in the lateral decubitus position with the upper arm extended and placed over the head (Fig. 22.4). Because of the vulnerable conditions in the small child, stretching of the brachial plexus must be avoided. Rolled towels and other positioning devices may be placed to optimize stabilization and exposure of the operative field.

Usually, a gently curved thoracotomy incision is made. For wide exposure of the entire chest, the incision begins below the level of the breast and is carried out posteriorly in a straight line (which differs from the approach used in adults). To limit postoperative morbidity, it is desirable and usually possible to use a muscle-sparing approach. This affords adequate exposure, yet avoids division of the anterior serratus muscle and the chest wall musculature other than the latissimus dorsi. In most infants, the second is the highest palpable rib.

Ordinarily, lobectomy in the right or left chest and any pulmonary resection may be performed through the fifth intercostal space. At times, the fourth or sixth intercostal space should be used if a particular anomaly dictates this approach. The basic principle is adequate exposure with good visualization of the pulmonary arterial and venous branches leading into the involved lobe, as well as the lobar bronchi. Gentle lateral and inferior traction on the lobe exposes the hilum. The visceral pleura is carefully incised circumferentially, exposing the hilar structures (Fig. 22.5). The inferior pulmonary ligament should be divided at this time to facilitate expansion of the lower lobe. In most instances, it is favorable to dissect and divide the pulmonary arterial branches first, the venous drainage second, and the bronchus last.

The main arterial branches are individually encircled, ligated, and divided, typically with nonresorbable sutures and double proximal ligatures. The same approach is used for the venous drainage of the lobe. The bronchus is clamped and divided. Commercially available stapler systems are not fine enough for infants; a simple running sewn closure is best (Fig. 22.6). Air leaks may be identified by filling the chest with warm saline and by controlled inflation of the residual lung.

In case of lobar emphysema, the left upper lobe most commonly is involved, followed by the right upper and middle lobes. The appropriate treatment is emergency lobectomy. In BPS, the most common form of sequestration is intralobar. Extralobar sequestrations occur frequently with posterolateral diaphragmatic hernias, congenital heart disease, and duplication cysts. In CPAM, one or multiple pulmonary cysts may occur; the lower lobes frequently are involved.

Figure 22.4

Lateral decubitus position

Figure 22.5

Exposure of the hilar structures

Figure 22.4

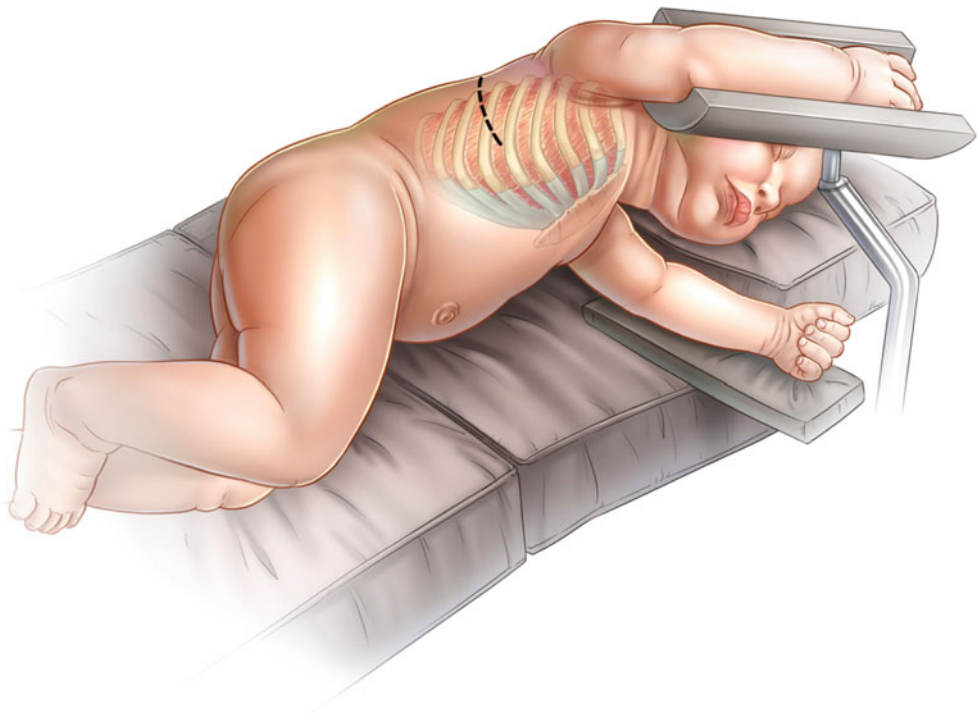


Figure 22.5

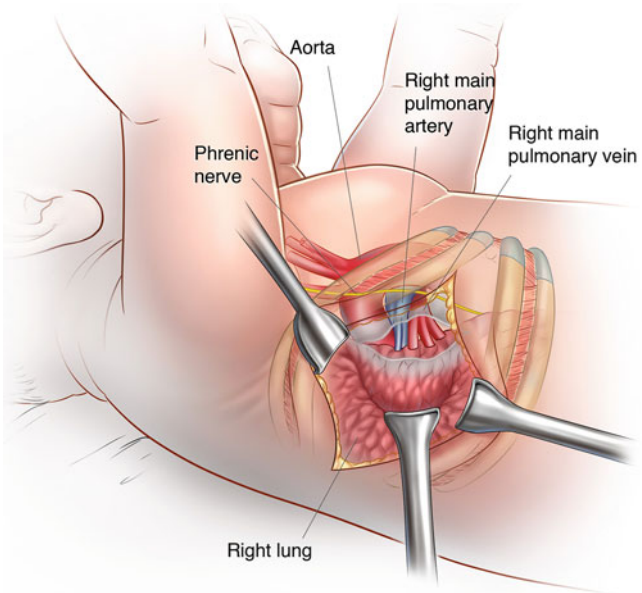
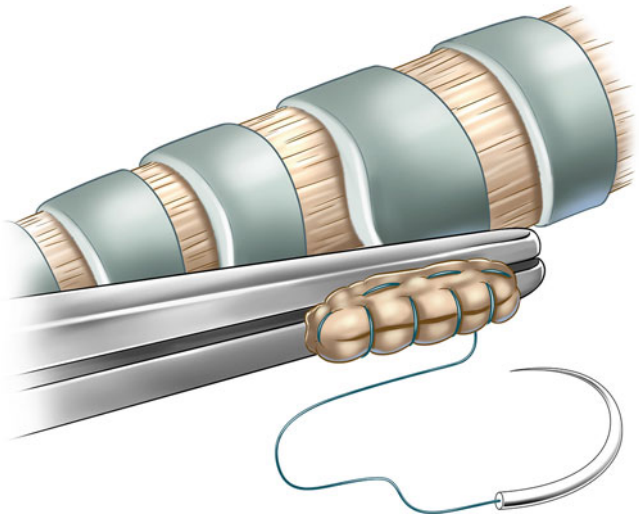


Figure 22.6

Running suture for closing the bronchus

Figure 22.6



Special Aspects

Thoracoscopic lung resection is a safe and effective technique. The procedure is performed with the patient in the lateral decubitus position and with single-lung ventilation. Three to five endoscopic ports (3–12 mm) are necessary (Fig. 22.3). The Endo GIA stapler (Covidien, Mansfield, MA) is useful in older patients to complete the fissure and take in the main pulmonary vessels and bronchus. In smaller patients (<15 kg), endoclips and ligatures are used. The LigaSure (Valleylab/Covidien, Mansfield, MA), a bipolar sealing device with a 5-mm curved dissector design, is the primary mode of vessel ligation. It also proves useful in sealing the lung and completing the fissure.

The chest initially is inflated with low-flow, low-pressure carbon dioxide to help complete lung collapse. A flow of 1 L/min and pressure of 4–6 mmHg are recommended.

Conclusion

A few inborn diseases, such as CPAM and BPS, must be observed and the treatment strategy determined by an interdisciplinary team of neonatologists, obstetricians, and pediatric surgeons. The removal techniques basically are the same as the approaches used in adults.

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