Treatment options and anesthetic considerations for postnatal management of congenital lung malformation

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ABSTRACT

Congenital lung malformations or CLM have resulted in an apparent increase in their frequency with the advances in fetal ultrasound technology and more widely available investigations in prenatal and postnatal periods. The advantage of an early diagnosis came together with the difficulty of the treatment decision, in special in the postnatal management of asymptomatic CLM. In this article, we summarized findings from current literature, with a particular emphasis on the therapeutic strategies.

Management of infants for thoracic surgery represents a challenge not only for pediatric surgeon but also for anesthesiologists because of the anatomic and physiologic variations particularities. There are two main important parts that must be obtained: one lung ventilation and specific techniques for optimal pain management, distinguishing pediatric thoracic anesthesia from other pediatric anesthetic disciplines.

A comprehensive multimodal approach is necessarily followed by a long-term assessment of respiratory function of children who underwent surgery for lung resection.

Keywords: congenital lung malformations, postnatal management, surgery, anesthesia, children

INTRODUCTION

Congenital lung abnormalities (CLA), also termed bronchopulmonary malformations (BPM) are isolated structural birth defects characterized by focal disruption of normal lung development with unknown aetiology and pathogenesis.

There are no many data on their frequency, the estimated incidence is 1 in 3000 to 1 in 6000 live births [1,2].

Although uncommon overall, advances in ultrasound technology have resulted in an apparent increase in their frequency [1,2,3]. A large percentage of these will remain asymptomatic in the neonatal period and later. The rising incidence is likely due to the widespread availability of prenatal ultrasound combined with the improved resolution of ultrasound technology that enables the detection of smaller lung lesions along with supplementary fetal magnetic resonance imaging (MRI) [4].
THE CLASSIFICATION AT PRESENTATION. CLINICAL APPROACH

The nomenclature of congenital lung disease was often confusing with terms that often overlap and used inconsistently before and after birth. A clear dialogue among various professionals (obstetricians, perinatologists, pediatric surgeons, pathologists, pediatricians, radiologists) became important and prompted calls for reappraisal of the diagnosis, investigation and management of congenital lung disease [5,6,7,8,9,10,11].

It has been suggested by Bush and Co [6], that few principles should be followed:

• keep clinical and pathologic description separate: clinical descriptions should use clear terms and not include assumptions of pathology.
• antenatal ultrasound abnormalities should be described using such terms as increased echogenicity with large, small, or multiple cysts rather than as “Congenital pulmonary airway malformation or CPAM,” which is, and remains, a histological diagnosis.
• a radiographic abnormality should be described as solid or cystic and their size should be recorded. Only if the lesion has been excised is it relevant to make a pathologic diagnosis (for example various histologic types of CPAM) [6,7].
• the descriptions should be made without indulgence in embryologic speculation, specific antenatal diagnoses often have to be revised after postnatal excision of the lesion [6,12].

The need for a simple, easy and clinically relevant classification system of congenital lung malformation comes from the fact that often present to the pediatric surgeon during infancy or early childhood and a surgical decision cannot be based on a system that relies on histology taken after surgery [5].

Michael Seear et al [5] propose a simplified classification of congenital lung malformation based on the results of a prenatal ultrasound plus a postnatal chest radiograph:

Group 1 – congenital solid/cystic lung abnormality
• Congenital pulmonary airway malformation
• Intra and extra lobar sequestrations
• Bronchial atresia
• Bronchogenic cyst
• Mixed malformations

Group 2 – congenital hyperlucent lobe
• Congenital lobar emphysema
• Polyalveolar lobe

Group 3 – congenital small lung
• Lung/lobar agenesis
• Pulmonary hypoplasia associations

This classification is based on the information immediately available to the clinician and is also intended for areas where these investigations are easily available. It also provides guidance for extra investigations specific to each group and has clinical value especially for surgeon at the time of initial presentation [5].

EMBRYOLOGY AND POSSIBLE COMPLICATIONS

Congenital lung malformation arises during the embryonic development of the lungs, either due to interruption of the normal organogenesis or a mismatch of the epithelial-mesenchymal interaction caused by dysregulation of cellular signalling [13].

Another theory of causation is that these lesions are related to airway obstruction with secondary pulmonary dysplastic changes [14].

Two different theories have been put forward to explain the pathophysiological mechanisms [15]. The environmental hypothesis states that a persistent expression of early lung development markers caused by potential genetics defects would lead to a focal and temporary interruption of the lung morphogenesis.

More recently, histological studies of lung malformations have suggested an obstructive hypothesis: focal obstruction of the air tree, either functional (peristalsis abnormality) or organic (bronchial stenosis) [15,16]. The modes and timing of obstructive events are currently poorly understood [14].

Antenatal complications can appear: fetal hydrops, pleural effusion, or polyhydramnios secondary to failure of normal fetal swallowing because of esophageal compression.

Postnatal complications include: infections (bacterial and also fungal and mycobacterial), bleeding (which may lead to hemothorax), air embolism, high-output cardiac failure due to shunting through systemic collaterals, pneumothorax and malignant changes [10].

A congenital lung abnormality is often found in conjunction with other abnormalities: congenital heart defect, gastrointestinal defects, genetic syndromes, vascular, genital, urinary, limbs, respiratory anomalies.

THERAPEUTIC STRATEGIES

Fetal management

Fetal interventions play an important role in large congenital lung lesions associated with hydrops. Maternal steroids are the mainstay of fetal management of congenital lung malformations [17]. A study performed at the Children’s Hospital of Philadelphia showed that although the mechanism for the steroid effect is not fully understood, the need
for fetal procedures dropped and a rise in survival rates was observed after this strategy was implemented. In cases where foetuses fail to respond to maternal steroids trials other fetal interventions may be used [18].

Thoracoamniotic shunts have a clear role in large lesions containing a dominant macrocyst associated with hydrops and non-responders to steroids treatment [17,19].

Fetal lung resection was the mainstay procedure in hydropic foetuses until the introduction of steroids. Currently, fetal lung resection is reserved for borderline viable hydropic foetuses with microcystic CPAMs or bronchopulmonary sequestrations (BPS) that are non-responders to maternal steroids [17,19].

The EXIT (ex-utero intrapartum treatment) procedure remains somewhat controversial, proponents arguing that it provides a hemodynamically stable environment for the fetus, maximizing the chance for survival in viable hydropic foetuses [17,19].

Postnatal management

The management of congenital lung malformations depends on the type of lesion and, first, on the presence or absence of symptoms. In the presence of respiratory distress, plain films of the chest are the best initial study to determine the degree of mediastinal shift and to rule out pneumothorax. A preoperative CT scan is ideal, providing information on the underlying lesion, location and the existence of a systemic feeding vessel [17,19]. In symptomatic patients the urgency of intervention is clear. Surgery consists of lobectomy for CPAMs, congenital lobar emphysema (CLE) or intralobar BPS and resection of the lesion for extralobar BPS or bronchogenic cysts. These can be accomplished through open or thoracoscopic surgery.

The management of asymptomatic lesions is still controversial. The rationale for preemptive resection of CPAMs and BPS is based on the desire to avoid complications, such as recurrent pneumonias, pneumothorax, hemoptysis and malignant transformation [17,20]. The controversy is related to the belief that many lesions regress and disappear. Currently, the mainstay of treatment for asymptomatic congenital lung lesions is early resection, at 6-10 weeks of age [21]. The advantages of performing a lobectomy at this age are the low rate of complications in experienced centers and the negligible impact on long-term pulmonary function associated with infant lobectomy, the potential for compensatory lung growth being higher in younger infants.

Outcomes

The long term outcome is determined by a number of factors including development of symptoms, requirement or selection for surgical resection, extent of resection, type of lesion. The overall survival rates are now around 95% [17,22].

Total lung capacity (TLC) is generally preserved at long-term follow-up after lobectomy for congenital lung lesions. Studies suggest that overexpansion of residual lung may be compensating for loss of tissue and contributing to TLC rather than true lung growth [22,21].

The extent of resection plays an important role in the long term outcome. In the extreme situation of a pneumonectomy the long term sequelae include scoliosis and postpneumonectomy syndrome (rotation of the heart and great vessels and bronchial compression). Implantation of prostheses into the empty hemithorax appears to be effective in children. Interestingly, the respiratory function is generally reasonably well preserved in children who went through pneumonectomy before the age of 5. Recently, performing segmentectomies in place of lobectomies for congenital lung lesions has been proposed, when anatomically possible. This should be balanced against the risk of incomplete resection and recurrence [19,22].

Chest wall deformities are a complication of open lobectomy through thoracotomy. 7% of children undergoing surgery for congenital lung lesions were identified to have chest wall deformities after the age of 5. With increasing use of thoracoscopic techniques these complications will be reduced [23].

Anesthetic Consideration

Management of infants for thoracic surgery represents a challenge for anesthesiologists because of the anatomic and physiologic variations particularities. The most important one remains the size of the patient with all the technical difficulties involved ranking one of the niche practices and that makes it different from adults. The preoperative evaluation plays an important part providing important information that can change the approach and it should include the neonatal history as this may indicate comorbid pulmonary and cardiac problems along with syndromes which must be investigated [24].

There are two main important parts that must be obtained: one lung ventilation and specific techniques for optimal pain management, distinguishing pediatric thoracic anesthesia from other pediatric anesthetic disciplines.

There are numerous techniques described to achieve lung isolation, must emphasize that there is no single technique suitable [25,26]. Non-cardiac thoracic surgery in pediatric patients requires different strategies adapted to their age, size and device to achieve one lung ventilation. The risk of hypoxic episodes during surgical procedures is more present in pediatric patients compared to adults on
one hand the pressure gradient in lateral position is smaller between the non-dependent and dependent lung parts and that leads to a more equal distribution of pulmonary blood flow and on the other hand the chest wall elasticity is higher, these two can further lead to overall ventilation/perfusion mismatch. Techniques and device choice for lung isolation includes the use of balloon-tipped bronchial blockers, endobronchial intubation (is the most common approach with the help of flexible bronchoscope, fluoroscopy or blind advancement) and double lumen tube (ideally, but limited for 8 years old patients) [24,25,26]. However, based on the level of training and abilities of the clinician the techniques and devices used must always be adapted to the size and age of the patient. The clinician must be aware of the advantages and disadvantages when considering a device or technique and to weight the risks and benefits.

Pain management represents an essential part for all children undergoing thoracic surgery. Pain assessment in these patients remains a challenge giving their size and age. Further, effective pain management reduces respiratory complications. A comprehensive multimodal approach involves: neuraxial techniques (epidural analgesia), loco-regional analgesia (such as intercostal nerve block, erector spinae block, serratus anterior plane block, thoracic paravertebral block), and analgesics such as non-steroidal anti-inflammatory drugs, paracetamol and opioids should be employed to ensure adequate pain relief [24,25,26]. Unlike in adults, regional blocks in children are performed under general anesthesia to improve compliance and reduce patient movement during block placement. Infants require higher relative doses of local anesthetic than adults to achieve nerve blockade and being at higher risk of neurologic or cardiac toxicity due to low plasma protein binding and decreased intrinsic clearance.

All approaches to single lung ventilation require some degree of practice to be instituted safely and effectively [24,25,26].

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Concerning the long-term assessment of respiratory function of children who underwent surgery for lung resection, the literature is rare with opposite results, some studies has shown that a restrictive syndrome can develop if the resection is not made soon enough to benefit from the lung compensatory growth [15].

A recent study comparing the respiratory outcome of patients operated on by thoracotomy for congenital lung malformations to control patients operated on for inguinal hernia showed that patients operated on for lung lesions had a higher prevalence of wheezing, lower respiratory tract infections, and needed more inhaled bronchodilators and systemic steroids in their first years of life, but these sequelae tended to disappear after 4 years of age [15,20].

Complications in adulthood are thoracic malformations (thorax growth asymmetry, scoliosis, pectus excavatum) related to the size of the malformation. Abnormalities of the mammary gland can also be seen [15,27].

**CONCLUSION**

With the increased access and application of prenatal ultrasound and magnetic resonance imaging (MRI) most lung abnormalities are detected prior to birth [5,6]. Many congenital lung abnormalities may regressed significantly by the time of birth [5,28,29] and for others the natural history is unknown [6].

Although, the management of asymptomatic lesions is still controversial, in symptomatic patients the surgery is necessary and can be accomplished through open or thoracoscopic surgery. Regarding the anesthesia, there are different techniques and device choices. In the multidisciplinary team, the clinicians should implement a multimodal approach with each patient.
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