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Congenital lung lesions: Postnatal management and outcome

Dakshesh H. Parikh, MBBS, MS, FRCS (Peds), MD[a,*],
Shree Vishna Rasiah, BM, BS, MRCPCH[b]

[a] Department of Paediatric Surgery, Birmingham Children's Hospital NHS Foundation Trust, Steelhouse Lane, Birmingham B4 6NH, UK
[b] Southern West Midlands Newborn Network and Birmingham Women's Health Care NHS Trust, Mindelsohn Way, Edgbaston Birmingham, B15 2TG UK

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ABSTRACT

Antenatal diagnosis of lung lesion has become more accurate resulting in dilemma and controversies of its antenatal and postnatal management. Majority of antenatally diagnosed congenital lung lesions are asymptomatic in the neonatal age group. Large lung lesions cause respiratory compromise and inevitably require urgent investigations and surgery. The congenital lung lesion presenting with hydrops requires careful postnatal management of lung hypoplasia and persistent pulmonary hypertension. Preoperative stabilization with gentle ventilation with permissive hypercapnia and delayed surgery similar to congenital diaphragmatic hernia management has been shown to result in good outcome. The diagnostic investigations and surgical management of the asymptomatic lung lesions remain controversial. Postnatal management and outcome of congenital cystic lung lesions are discussed.

Introduction

Prenatal ultrasound scan in most instances accurately diagnoses the congenital lung lesion in the western world. Congenital cystic lung lesions include a wide spectrum of pathological abnormalities: congenital cystic adenomatoid malformation (CCAM), pulmonary intra and extralobar sequestrations, bronchial atresia, foregut cysts, and congenital lobar emphysema. Although many sonographers antenatally diagnose pathological conditions, such as CCAM, the reality is that congenital lung lesions on antenatal ultrasound can only be microcystic or macrocystic.[1,2] The sequential scanning can then diagnose increasing size of the lesion, mediastinal shift, polyhydramnios, hydrops, and associated cardiac anomaly/dysfunction by fetal echo. Large lung lesions with hydrops can result in fetal mortality.[3] The progress and persistence of large lesions on serial antenatal ultrasound at 32–34 weeks of pregnancy governs the location of the delivery in UK.

Varying degrees of respiratory distress is a presentation feature in approximately 25–30% of infants in immediate postnatal period. However, subsequent presentations and symptoms in infancy and childhood are related to infections and occasionally to respiratory distress related to sudden increase in size, compression of airway, or rarely with spontaneous pneumothorax.[4–6] However, many may remain asymptomatic and require either follow-up or elective resection depending on the evidence-based and best-informed strategy employed by the surgical unit.[7–10] In order to manage the variable presentations of congenital cystic lung lesions, it is important to have a good understanding of the cardio-respiratory neonatal physiology and the impact of these lesions on the development of the intra-thoracic organs and their subsequent function. This will help us understand how these lesions might present postnatally and guide their respective management.

Neonatal cardio-respiratory adaptation

Survival following birth depends upon a neonate undergoing several adoptive and coordinated cardio-respiratory changes. This is one of the most critical periods of adaptation that ensures that a dependent fetus makes a smooth transition into its new environment and breathes air for the first time. As the newborn inflates the lung with air, the pulmonary arterial oxygen levels increase and carbon dioxide levels decrease. This results in reduction in pulmonary vascular resistance and increases the cardiac output into the neonatal lungs. As the umbilical blood flow is severed and systemic vascular resistance increases, the fetal circulation closes, depending on ductus venosus, foramen ovale, and ductus arteriosus.[11] When this transition does not occur, the neonate remains in a persistent fetal circulation with continuing persistent pulmonary resistance and pulmonary hypertension of the newborn (PPHN).[12,13]

Continuous gas exchange and breathing require adequate surfactant at the alveolar level to maintain and reduce the work of breathing. Surfactant deficiency results in respiratory distress syndrome especially in the extremely preterm neonates. The

The study was performed in Birmingham Children's Hospital NHS Foundation Trust, Southern West Midlands Newborn Network Lead, and Birmingham Women's Health Care NHS Trust, Birmingham.

* Corresponding author.
E-mail address: dakshesh.parikh@bch.nhs.uk (D.H. Parikh).

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surfactant deficiency should be anticipated and managed by antenatal maternal administration of steroids (bethamethasone) and failing that by the exogenous administration of surfactant via the endotracheal tube postnatally. Maternal administration of antenatal steroids promotes surfactant production and maturation of the fetal lung.[14,15] Exogenous administration of the surfactant has been proven effective in prophylactic and rescue treatment of respiratory distress syndrome in premature infants.

Physiological and physical factors responsible for lung growth

Embrylogically, fetal lung normally develops in 5 stages: embryonic, pseudoglandular, canalicular, saccular, and alveolar.[11] There are important physiological and physical factors, which ensure normal lung growth in utero. Of these, fetal lung fluid and mechanical distension is paramount for the development of the lung. This fluid is produced in the lungs and moves toward the oropharynx, where it is either swallowed or mixed in the amniotic fluid. It increases in volume and keeps the lungs distended in order to stimulate parenchymal growth.[16,17] Therefore, any reduction or lack of fetal lung fluid can result in pulmonary hypoplasia especially if it occurs in the early stages of embryological development.

Physically, the lung tissue needs to grow and expand within the intra-thoracic cavity. Anything that affects its growth and expansion in utero can result in pulmonary hypoplasia of the affected side especially earlier on in the embryological stage. This is also observed in congenital diaphragmatic hernias associated with pulmonary hypoplasia. Large space occupying congenital thoracic lesions can have a similar effect on the lung development, resulting in pulmonary hypoplasia.

Postnatal cardio-respiratory pathophysiology in congenital lung abnormalities

The important functions of cardio-respiratory system are to adequately uptake oxygen from inspired air and deliver it to the vital organs and other parts of the body. This vital respiratory function is dependent on number of factors, namely, partial pressure of oxygen in inspired air, ventilation and perfusion matching, hemoglobin concentration in blood, cardiac output, and blood volume.[18] The continuing gas exchange depends upon adequate alveolar ventilation and pulmonary blood flow. Congenital anomalies such as congenital diaphragmatic hernia and congenital cystic lung lesions are associated with varying degrees of lung hypoplasia, thickened pulmonary arterial branches, and pulmonary hypertension. Furthermore, large congenital cystic lung abnormalities, lobar emphysema, and neonatal intra-thoracic tumors can cause a significant mass effect with mediastinal deviation compressing heart and mediastinal structures postnatally.

Providing oxygen to vital organs in the presence of pulmonary hypoplasia with PPHN poses a significant challenge in postnatal period. In this situation, right ventricle works harder to maintain cardiac output by shunting across the patent foramen ovale and patent ductus arteriosus. The management of PPHN and oxygen delivery to vital organs becomes difficult in the presence of cardiac anomalies, chest infections, and prematurity. Management of PPHN in the postnatal period is directed toward reducing the pulmonary arterial resistance, supporting cardiac function while preventing parenchymal injury during assisted ventilation. The combination of PPHN in association with prematurity, cardiac defect, and large space occupying congenital lung lesion can be lethal. The outcome in this situation is dependent on tertiary neonatal and/or pediatric intensive care expertise.

Optimum oxygen delivery with assisted ventilation

Significant advances in neonatal intensive care medicine allow us to support neonates with assisted ventilation to optimize oxygen delivery while monitoring the effectiveness of the delivery. Broadly, 2 modes of assisted ventilation strategy are employed to support a neonate in respiratory distress.[19] The conventional ventilation traditionally delivers tidal ventilation based on the pressure settings set by the clinicians and stops when pressure is reached. While the volume guarantees ventilation, which sets the volume, the ventilator generates the necessary parameters to deliver the set volume. The other mode of ventilation is called high-frequency oscillation. This is aimed at delivering smaller gas volumes at rapid rates.

The current practice in neonatology is to electively ventilate with high-frequency oscillation in antenatally diagnosed congenital diaphragmatic hernia and those with severe hydrops fetalis in order to reduce barotrauma to hypoplastic lung parenchyma. The “gentle ventilation” strategy with permissive hypercapnia has improved the outcomes in these babies.[20] Similar strategies can be employed to manage large space occupying congenital intra-thoracic lesions associated with pulmonary hypoplasia and/or hydrops.

Where there is evidence of PPHN clinically (pre- and post-ductal saturation difference of more than 10%), serial echocardiogram assessment is vital in reviewing progress and guiding cardio-vascular and inotropic support. Echocardiogram demonstrates the right to left shunting across the patent foramen ovale and/or detects the patency of ductus arteriosus, right ventricular hypertrophy, and any congenital heart defects. In those patients with tricuspid regurgitation (TR) measurable with continuous wave Doppler, the modified Bernoulli equation for right ventricular–atrial pressure difference provides an estimate of right ventricular systolic pressure and in the absence of right ventricular outflow tract obstruction, pulmonary artery systolic pressure (peak pressure difference $= 4 \times$ peak TR velocity^2).[12] In patients with significant PPHN, the use of inhaled nitric oxide (iNO) is an important pulmonary vasodilatator in reducing the pulmonary vascular resistance, improving cardiac function, and ultimately oxygenation. In resistant cases of pulmonary hypertension despite full cardio-respiratory support on HFOV, iNO, and inotropes, extracorporeal membrane oxygenation (ECMO) should be considered early. The indications for considering ECMO would be a rising oxygenation index of more than 20, despite all the above measures to manage the resistant PPHN.

Congenital lung lesions with hydrops

Congenital cystic lung lesions causing polyhydramnios leading to fetal hydrops are fortunately rare. The mortality associated with hydrops both in utero and postnatally is reported as between 68% and 89%.[21–23] Lung hypoplasia is thought to be the cause of mortality in majority of these cases. The pathological spectrum and natural history of the cystic lung lesions and fetal hydrops are variable.[24–30] Antenatal management of cystic lung lesions is covered by an article within this journal and is thus not elaborated in this article.

Delayed surgery following preoperative stabilization

Lung hypoplasia associated with poor pulmonary compliance and varying degrees of pulmonary vascular resistance and resultant persistent pulmonary hypertension is observed in neonates with congenital diaphragmatic hernia (CDH). It is well-recognised fact in the management of CDH that immediate intervention