Chapter 28: Neonatal pulmonary disorders

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The establishment of breathing and the adaptation of the neonatal circulation to extrauterine life are two of the most important events which occur at the time of birth. These physiological changes are mediated by a number of factors acting both systemically and locally, but the major control of these events is via the respiratory centres in the brainstem. It is thus vital that these are present in an intact and functional state if the adaptation to extrauterine life is to occur uneventfully. Most of the diseases seen in the neonatal period are a consequence of adverse factors affecting these normal physiological processes, such as immaturity, infection, damage at the time of birth, or congenital malformation.

Onset of breathing

The onset of breathing is brought about by the responses to a number of complex reflexes which stimulate the respiratory centres. These include thermal stimulation such as cold, external sensory stimulation, particularly tactile, and intrinsic reflexes within the rib cage as the chest changes in shape secondary to the pressure applied during and immediately after the birth process itself. Biochemical changes, such as hypoxaemia and hypercarbia also have an important stimulatory effect on the central nervous system, as do the acute pressure changes within the cardiovascular system which occur secondary to clamping of the umbilical cord. The inspiratory pressures required to open the lungs are thought to be relatively low, of the order of 10-20 cmH₂O, which is sufficient to overcome the resistive forces of the lungs, the residual fluid within them and the chest wall (Milner and Vyas, 1982). A functional residual capacity (FRC) is established within the first few breaths and a positive expiratory force is exerted during the later part of each breath in order to aid the clearance of lung liquid. The normal functional residual capacity of the newborn is approximately 30 mL/kg and this lung volume is stabilized during the first 30-60 minutes of life (Klaus et al, 1962).

The lungs are normally filled with surfactant rich fluid \textit{in utero} at the same volume as the subsequent functional residual capacity after birth. This liquid is emptied from the lungs during and after the birth process, partly by external compression of the chest as it passes through the vagina, or the uterine wall in the case of the caesarean section. This accounts for approximately one-third of the lung liquid, the remaining fluid is absorbed directly across the pulmonary lymphatics or into the pulmonary capillaries during the first minutes after birth (Strang, 1977).

These changes in lung volume and compliance are matched by major circulatory adaptation which occurs simultaneously. Before birth only about 10% of the cardiac output passes through the lungs because of the high pulmonary vascular resistance and the fact that the lungs are not being used to sustain oxygenation. After birth the aeration of the lungs produces a rapid reduction in pulmonary vascular resistance and this is accompanied by a rise in P\textsubscript{aO₂} from the intrauterine levels of 4 kPa (30 mmHg) to the postnatal level of 8-12 kPa (50-90 mmHg); these changes stimulate the closure of the ductus arteriosus and foramen ovale, thus closing off the large right to left intracardial shunts which are present before birth. These fetal channels are not, however, irreversibly closed at this stage and may re-open during periods of stress and hypoxia.
Central control of breathing is vital to the maintenance of life and the respiratory centres must be maintained intact throughout this period. They are particularly sensitive to asphyxial insult, including hypoxia and hypercarbia associated with respiratory and metabolic acidosis. They may be damaged by trauma or by localized areas of haemorrhage during a difficult or protracted delivery. They are also sensitive to the effects of drugs such as pethidine given to the mother before birth for sedation or analgesia. Any interruption to these normal physiological processes will result in birth asphyxia.

**Birth asphyxia**

Asphyxia at birth may be acute or chronic. Some babies suffer from acute lack of oxygen during the birth process, for example as a consequence of cord prolapse, while others may have had chronic intrauterine asphyxia as a result of postmaturity or placental dysfunction secondary to hypertensive changes.

The major groups of conditions leading to birth asphyxia may be divided into three categories, maternal, placental/cord related and fetal. Maternal predisposing factors include hypertension, diabetes, underlying cardiac or renal disease, abuse of drugs or alcohol, hypotension, or anaesthetic complications such as aspiration. Other causes include multiple births, abnormal presentation, cephalopelvic disproportion and prolonged labour. Placental problems will include abruption, placenta praevia, early separation and cord prolapse. Fetal conditions include pulmonary hypoplasia, anaemia secondary to Rhesus disease, intrauterine meconium aspiration and congenital malformations, particularly of the lungs or heart.

The events which occur during an asphyxial episode have been well described by Dawes et al (1963). The asphyxiated infant gasps initially and this is followed by a period of primary apnoea during which there is a decrease in heart rate, but maintenance of blood pressure and peripheral circulation; hypoxaemia results in profound cyanosis. Resuscitation during this period will be effective by the use of external stimulation and the application of oxygen which will be inhaled during the next phase of breathing, beginning 1-2 minutes later. This secondary gasping period lasts for a further 4-5 minutes following which terminal apnoea occurs. At this stage there is profound hypoxia, hypercarbia, acidosis and circulatory collapse. After 7-8 minutes of significant hypoxia cerebral damage will begin. Resuscitation at this stage requires positive pressure ventilation, usually following endotracheal intubation utilizing pressures of up to 20-30 cm with an initial breath of 3-5 seconds (Vyas et al, 1981). Bag and mask ventilation may also be effective in the absence of personnel skilled in intubation. Circulatory support by external cardiac massage is essential in those with profound bradycardia or cardiac arrest. Drugs such as glucose, sodium bicarbonate and calcium gluconate may also be required. Adrenaline may be given via the endotracheal tube in those with profound circulatory depression (Greenberg, Roberts and Baskin, 1981). Reversal of maternal sedative drugs such as pethidine, which prolongs the period of primary apnoea, may also be achieved by the use of parenteral naloxone.

After acute resuscitation severely asphyxiated infants may require extensive support including ventilation, maintenance of blood pressure and stimulation of cardiac output, control of renal failure and acid-base balance. Neurological outcome also depends on the control of seizures and cerebral oedema in the perinatal period. The outcome is extremely variable,
although a very significant proportion of those who survive the initial neonatal period may turn out to be normal (Thompson, Searle, and Russell, 1977).

**Hyaline membrane disease**

Hyaline membrane disease (HMD) is the commonest cause of respiratory difficulty in this age group and remains the major cause of mortality and morbidity in the neonatal period. Its incidence is inversely proportional to birth weight and it is particularly prevalent in babies who weigh less than 1.5 kg at birth and whose gestation is less than 32 weeks. The illness is caused primarily by surfactant deficiency (Avery and Mead, 1959). Other predisposing factors include birth asphyxia, caesarean section without labour, maternal haemorrhage, maternal diabetes and multiple pregnancy.

The infant typically develops signs of respiratory difficulty within the first 2 hours after birth. These include indrawing of respiratory muscles, rapid respiratory rate, sternal recession and the classical grunt heard on expiration - a mechanism used to maintain oxygen levels (Harrison, Heese and Klein, 1968). The chest X-ray typically shows a ground glass appearance secondary to alveolar hypoventilation and air bronchogram due to maintenance of large airway patency of the cartilage content. The differential diagnosis must include group B streptococcal pneumonia (Pyati et al, 1981), persistent fetal circulation (Fox and Duara, 1983), and underlying cardiac abnormalities such as transposition of the great arteries.

**Treatment**

Treatment of hyaline membrane disease is complex. Prevention of preterm delivery is desirable wherever possible, antenatal steroids may stimulate lung maturity in selected patients (Liggins and Howie, 1972). More recently, the replacement of surfactant artificially has begun to achieve some success, although the benefits remain short lived (Morley, 1984). Standard treatment for hyaline membrane disease is shown in *Table 28.1*.

**Table 28.1 Treatment of hyaline membrane disease**

<table>
<thead>
<tr>
<th>Neutral thermal environment</th>
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<tbody>
<tr>
<td>Maintenance of fluid balance</td>
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<tr>
<td>Oxygen $\text{PaO}_2$ 7-12 kPa (70-90 mmHg)</td>
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<tr>
<td>Correction of acid-base imbalance</td>
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<tr>
<td>Continuous positive airway pressure (CPAP)</td>
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<td>Artificial ventilation</td>
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<td>Paralysis</td>
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<td>Pulmonary vasodilators (tolazoline)</td>
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<td>Antibiotics for other causes (group B streptococci).</td>
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The use of mechanical ventilation in conjunction with positive end expiratory pressure (PEEP) has been the mainstay of respiratory support for infants with hyaline membrane disease over the last 20 years. However, this treatment carries a significant risk of side-effects such as induction of pneumothorax or subsequent bronchopulmonary dysplasia and should therefore not be instituted unless significant respiratory failure is present. Indications for its introduction include significant apnoea or impending exhaustion of the infant, inability to
maintain PaO₂ above 8 kPa (60 mmHg) in 60% oxygen or PaCO₂ below 8 kPa (60 mmHg). Most neonatal ventilators available at present are pressure limited, time cycled, constant flow generators, which produce a square wave input and which are applied at peak inspiratory pressures of less than 20 cmH₂O, initially at rates of 30-35 breaths per minute, inspiratory time 0.3-0.5 seconds and with positive end expiratory pressure of 3-4 cmH₂O. More recently, rapid rate ventilation of 60-120 breaths per minute has been under investigation (Field, Milner and Hopkin, 1984). High frequency ventilation, utilizing jet ventilators or an oscillator pump circuit, has also been evaluated in this condition (Frantz, Werthammer and Stark, 1983). Treatment is not without hazard to the infant, and a number of important complications are commonly seen including pneumothorax (Greenhough et al, 1984) - this may be prevented by selective paralysis - subglottic stenosis from prolonged endotracheal intubation, secondary infection and subsequent bronchopulmonary dysplasia (Northway, Rosan and Porter, 1967). Nowadays, with adequate support, 95% of infants suffering from significant hyaline membrane disease may expect to survive (Greenhough and Robertson, 1985), although the mortality rate remains significantly greater in those infants weighing less than 1 kg (Yu, Zhao and Bajuk, 1984). The ultimate prognosis for lung function is good providing that there has not been significant associated bronchopulmonary dysplasia.

**Transient tachypnoea of the newborn**

This condition is seen in a number of infants who show signs of respiratory distress, including tachypnoea, indrawing and cyanosis, lasting from 12 to 24 hours and sometimes longer. The chest X-ray shows mild bilateral haziness secondary to persistent interstitial lung fluid. These infants have a good prognosis and require basic supportive care and oxygen therapy with adequate control of blood gases. Very few require ventilation. The recovery is usually fairly rapid and the long-term outlook is excellent (Yu, 1986a). This disease is more common in infants born after caesarean section, probably because less fluid is squeezed from the lungs during the birth process itself so some of these infants may have a lower initial functional residual capacity (Milner, Saunders and Hopkin, 1978).

**Patent ductus arteriosus**

Patent ductus arteriosus is uncommon among normal term infants. It is, however, extremely common in infants with hyaline membrane disease, reaching levels of over 40% in those under 1 kg and 20% in those between 1.0 and 1.5 kg at birth (Ellison et al, 1983). It is more common in those who have higher fluid intakes (Bell et al, 1980).

Many preterm infants, especially those with hyaline membrane disease, develop classical signs of patent ductus arteriosus during the first 3 or 4 days of life, these include tachycardia, bounding peripheral pulses, a murmur below the midclavicular region, which gradually becomes continuous, and a palpable systolic thrill over the precordium. During the latter part of the first week of life the shunt in these babies becomes increasingly left to right in nature and this leads to pulmonary oedema and decreased lung compliance. The infant commonly has an associated increase in ventilatory requirements as this process occurs and often develops secondary infection in the lungs at this stage. The liver is also frequently enlarged and this indicates the onset of cardiac failure. Diagnosis may be confirmed utilizing echocardiography which will also exclude other underlying cardiac lesions. Chest X-ray shows
cardiomegaly and plethoric lung fields in addition to the background pulmonary shadowing present from the hyaline membrane disease.

Management initially includes restriction of fluid intake to 60-70% of daily requirement, diuretic therapy and increased end expiratory pressure to reduce pulmonary oedema. If these measures fail to produce significant reduction in ductal shunting within 24-48 hours then indomethacin, a prostaglandin synthetase inhibitor, is given in a dose of 0.2 mg/kg per day in two divided doses over 2-3 days. This has been shown to produce a satisfactory response in most infants in a large multicentric trial (Gersony et al, 1983). A significant proportion will re-open subsequently but, in many cases, a satisfactory response is produced which allows weaning of the infant from the ventilator. If these measures fail then surgical ligation, preferably undertaken in the neonatal unit, is performed.

**Persistent pulmonary hypertension (persistent fetal circulation)**

Persistent fetal circulation occurs when the normal reduction in pulmonary vascular resistance fails to occur after birth. The normal stimuli for this process include the increased oxygen levels after breathing starts and the changes in circulatory pressures which occur with clamping of the cord. Increased tone of the pulmonary arterioles may be brought about by persistent hypoxia, especially in the presence of acidosis, and this results in an attempt by the newborn to revert to the fetal circulatory pattern. This complication may occur in babies with hyaline membrane disease, but is particularly likely to happen in those who have had intrauterine asphyxia. It is also seen in infants with meconium aspiration syndrome (Fox et al, 1977) and in those with group B streptococcal septicaemia. It is difficult in some cases to distinguish this from cyanotic congenital heart disease and echocardiography is required to exclude underlying cardiac lesions.

Treatment consists of ventilatory support and an attempt to increase the pH to more than 7.48. At this level pulmonary vasodilatation is more likely to occur. Hyperventilation reducing the PaCO₂ to 3.5-4 kPa (25-30 mmHg) may assist this process (Drummond et al, 1981). Administration of sodium bicarbonate may also improve the pH. If hyperventilation fails to produce an adequate response infusion of tolazoline in a dose of 1-2 mg/kg may produce an improvement in PaO₂ and this should be followed with a continuous infusion at the same dosage/kg per hour if a satisfactory response occurs.

Tolazoline may also produce systemic hypotension and in this situation the baby will need to be supported with colloids in the form of plasma or blood transfusion and some may require dopamine in addition to maintain blood pressure.

**Meconium aspiration syndrome**

The passage of meconium before birth may be present in 8-15% of deliveries. In a study of 1000 consecutive births Gregory et al (1974) demonstrated the wide variability of X-ray changes and disease, with an overall incidence of 1.6% of neonates producing symptoms. The disease is most commonly seen in term or postmature infants and is much less common in preterm infants (Ting and Brady, 19750. The most dangerous consequence of the passage of meconium is its aspiration into the lower respiratory tract. Since it is usually a response to intrauterine asphyxia it is not uncommonly associated with the presence of fetal
gasping and meconium may readily be aspirated into the trachea or even into the peripheral parts of the lung before or during birth itself. The results of this process may produce profound effects on the respiratory system. Large plugs can obstruct the main airway, leading to acute asphyxia, smaller amounts aspirated into the peripheral airways will cause partial obstruction secondary to chemical pneumonitis and lead to significant overinflation, or collapse and atelectasis of adjacent lung tissue. This results in a high functional residual capacity with generalized overinflation and areas of marked ventilation-perfusion abnormality. Air leak with pneumothorax and pneumomediastinum is common and may be seen in as many as 40-50% of severely affected cases. There is also frequently an associated persistent pulmonary hypertension secondary to the asphyxial insult and this further contributes to the profound hypoxia which is present in those with the worst disease (Fox et al, 1977).

The disease is preventable with adequate antenatal care and the rapid delivery of infants who show signs of intrauterine distress. At birth it is vital to clear the airway by adequate suction of the trachea (Ting and Brady, 1975). After this procedure the infant should be observed for signs of respiratory distress. A chest X-ray should be performed in all who show any signs of respiratory difficulty. Treatment thereafter is supportive and includes correction of acid-base imbalance, antibiotics because of possible secondary bacterial infection and ventilatory support when indicated. Those who fail to respond to ventilation by the demonstration of persistent hypoxia have a very poor prognosis (Vidyasagar et al, 1975). Paralysis may be helpful (Runkle and Bancalari, 1984) and tolazoline should be given to those who have evidence of significant pulmonary hypertension. This condition continues to carry a significant mortality and a number of survivors have chronic lung disease secondary to the pressures required to ventilate them, these result in bronchopulmonary dysplasia. The long-term prognosis, in the vast majority of cases, is however, for normal lung function.

Infections

**Neonatal pneumonia**

Pneumonia in the neonatal period is extremely common particularly in babies who have other respiratory problems. It may be acquired before, during or after birth. The causative organisms may be bacterial or viral. The most serious bacterial pneumonia at present is due to the group B beta-haemolytic streptococcus. Up to 15% of mothers may harbour this organism in the birth canal and the infants will be in contact with this during normal birth. Less than 1% of such infants become significantly ill (Pyati et al, 1981), but the mortality in this group is extremely high. Predisposing factors include preterm delivery, prolonged rupture of the membranes and birth asphyxia. Presenting features are identical to hyaline membrane disease, with tachypnoea, indrawing, respiratory distress and cyanosis. Chest X-ray shows a ground glass appearance with bilateral generalized opacity present indistinguishable from hyaline membrane disease. In these cases the condition may be rapidly fatal unless early and appropriate antibiotics are given. Most babies at this stage are septicaemic and may show other complicating features such as disseminated intravascular coagulation.

Treatment consists of circulatory support and antibiotics, usually penicillin and gentamicin, or a third generation cephalosporin such as cefotaxime or ceftazidime. Other management consists of full intensive care including ventilation, maintenance of
cardiovascular stability and the prevention of hypoxia and acidosis. Despite early and aggressive therapy the mortality for those with rapidly progressive disease remains extremely high and is of the order of 60%.

A number of other bacteria may cause neonatal pneumonia including staphylococci. *Staphylococcus aureus* or *Staph. epidermidis* may be found in respiratory secretions among babies who are being ventilated, also *Pneumococcus* and *Listeria monocytogenes*. Gram-negative organisms, such as *Escherichia coli*, *Klebsiella* and particularly *Pseudomonas aeruginosa* are more commonly found in infants who have chronic lung disease and are often acquired via the ventilator and its humidification system.

**Chlamydia and fungi**

*Chlamydia trachomatis* has recently been shown to cause pneumonia in neonates and should be sought in those who have persistent symptoms or those who do not respond to usual antibiotic therapy (Frommell et al, 1979). Fungi, including *Candida albicans*, have recently been seen more frequently, particularly among debilitated neonates who have required repeated courses of antibacterial agents to control chronic chest infections. Extended treatment with appropriate parenteral antifungal agents is necessary.

**Viral pneumonia**

Viral pneumonia, particularly due to organisms such as cytomegalovirus, is also being increasingly recognized in the neonatal period. Recent surveys have shown that as many as 0.3-1% of neonates have passively acquitted cytomegalovirus in the upper airway at the time of birth (Peckham et al, 1983). These infants may develop persistent pneumonitis, which can lead to recurrent episodes of wheezing and clinically evident respiratory infection. The organism is also readily cultured from the urine in such cases. Another important virus is rubella which may cause interstitial pneumonitis (Boner et al, 1983). Herpes simplex types 1 and 2 are other causes of generalized infection and fatal lung disease in a small number of neonates, although this may be amenable to antiviral agents such as acyclovir.

**Bronchopulmonary dysplasia**

Bronchopulmonary dysplasia (BPD) is a chronic lung condition which has become increasingly frequent as more small babies have survived after intensive support with extended periods of oxygen therapy and mechanical ventilation. The incidence of the condition varies between 6 and 25% and is more common among those under 1 kg (Yu et al, 1983a). The aetiology is multifactorial and the classical changes were originally described by Northway, Rosan and Porter in 1967. Contributory factors include inhibition of mucociliary clearance secondary to endotracheal intubation, barotrauma from extended ventilatory support especially at high pressures, oxygen toxicity, chronic infection of the lower respiratory tract and hypersecretion secondary to the chronic respiratory disease. Other contributory factors may include pulmonary oedema and patent ductus arteriosus (O’Brodovich and Mellins, 1985). Vitamin deficiencies, including vitamins A and D and phosphate deficiency resulting in rickets and vitamin E deficiency have also been proposed as contributory factors. Infants developing bronchopulmonary dysplasia remain ventilator dependent after the first 3-4 weeks.
of life. In the most severe cases they may be ventilator dependent and subsequently oxygen dependent for many months.

Chest X-ray shows widespread overinflation, particularly in the lower zones, with fibrosis and loss of lung volume present throughout the lungs, but particularly in the upper zones. In the most severe cases these changes worsen with time and with intercurrent infections cardiac failure supervenes. Feeding, weight gain and growth may be extremely difficult to achieve in those who remain persistently hypoxic despite intensive support (Yu et al, 1983b).

Treatment consists of adequate oxygenation, diuretics, theophylline and other bronchodilators where indicated, adequate vitamins including vitamins A, D and E and antibiotics for intercurrent infections. Steroids may be helpful (Mammell et al, 1983) and their role is currently under further investigation. Those who survive show gradual improvement in lung function. There is also a considerably higher incidence of sudden infant death syndrome in this group. Those with milder disease may have virtually normal lung function after recovery and particularly after the first 2 years of life, during which time respiratory infections appear to be especially common.

Recurrent apnoea

This is an extremely common condition of preterm babies and is a variation from the normal pattern of periodic breathing which is seen at this age. Significant apnoea may be defined as episodes of cessation of breathing lasting 20 seconds or more, or less than 20 seconds if accompanied by bradycardia of significance (American Academy of Pediatrics, 1978). They occur for a variety of reasons including infections, for example pneumonia, meningitis or septicaemia, metabolic acidosis, hypoglycaemia, secondary to seizures or intracranial haemorrhage, respiratory depression from drugs and as a result of exhaustion in relation to respiratory or cardiac failure. They may be of central origin indicating immaturity of the respiratory centres. Upper airway obstruction such as cleft palate, choanal atresia or micrognathia will also contribute to such symptoms. Prior to treatment babies should receive full investigation for underlying disorders. These should be corrected whenever possible.

Those for whom no underlying pathology is found, except for immaturity of the respiratory centres, may respond to the administration of theophylline which is more effective than continuous positive airway pressure (Jones, 1982). It is important to ensure that the baby is not anaemic or chronically hypoxic or acidic, as correction of these abnormalities will reduce the incidence of the apnoic attacks. Those who have severe apnoea may require ventilation until such time as the respiratory system has matured and they are able to sustain an adequate breathing pattern spontaneously.

Congenital anomalies of the respiratory tract

These include abnormalities of the nose, palate, pharynx, larynx, trachea and major bronchi, lungs, ribcage and neuromuscular disorders. The range of lesions seen is shown in Table 28.2.
### Table 28.2 Congenital anomalies of the respiratory tract

**Upper respiratory**
- Choanal atresia
- Cleft lip and/or palate
- Pierre Robin syndrome

**Larynx**
- Laryngomalacia
- Stenosis
- Cleft
- Web
- Vocal cord paralysis
- Atresia

**Obstructive lesions of larynx**
- Haemangioma
- Cysts
- Papilloma

**Tracheobronchial lesions**
- Tracheo-oesophageal fistula with or without oesophageal atresia
- Tracheal pouch
- Tracheomalacia
- Tracheal stenosis
- Tracheal agenesis
- Bronchial stenosis
- Bronchial agenesis

**Extrinsic lesions**
- Vascular ring
- Aberrant innominate artery
- Duplication cyst

**Intrapulmonary lesions**
- Congenital lobar emphysema
- Congenital lung cysts
- Cystic adenomatoid malformation
- Pulmonary sequestration
- Pulmonary hypoplasia
- Pulmonary agenesis

**Diaphragm**
- Diaphragmatic hernia
- Eventration of diaphragm
- Paralysis of diaphragm

**Rib cage**
- Asphyxiating thoracic dystrophy
- Captomelic dwarfism
- Hypophosphatasia
- Osteogenesis imperfecta

**Neuromuscular disease**
- Congenital spinal muscular atrophy (Werdnig-Hoffman)
- Neonatal myotonic dystrophy
- Congenital myasthenia gravis.
Diseases of the upper respiratory tract and larynx are considered elsewhere in this volume and will not be discussed further at this stage. Disorders of the lower respiratory tree, below the larynx, are discussed in the following sections.

**Trachea**

Tracheal stenosis may be congenital or acquired. These infants present with biphasic stridor and respiratory difficulty, which is not always present at birth but which may become evident during an intercurrent respiratory infection. Wheezing is another common presenting feature due to retention of lung secretions below the level of the obstruction. Tracheal stenosis is more commonly caused by scarring secondary to prolonged endotracheal intubation. Direct tracheal surgery is an extremely high risk procedure and most children will be treated with a tracheostomy until subsequent tracheal enlargement, either spontaneous or surgically induced, occurs as age increases. Tracheal atresia is an extremely rare lesion and not usually compatible with life. It is commonly associated with congenital malformations of the oesophagus since the two structures have a common embryological origin. Tracheomalacia is due to weakening of the trachea wall, either spontaneously or in association with an extrinsic compressive lesion such as an aberrant blood vessel or vascular ring. It is also commonly found at the site of a tracheo-oesophageal fistula. Infants with tracheomalacia may suffer from respiratory difficulties, particularly during intercurrent infection, and during expiration when positive expiratory pressure in the lungs compresses the tracheal wall and causes collapse of the airway. If major apnoeic attacks are occurring as a consequence of this then aortopexy may be necessary (Filler, Rossello and Lebnowitz, 1976).

**Extrinsic lesions**

Lesions outside the trachea or major bronchi may produce respiratory obstruction by pressure on the airway, these include aberrant blood vessels (Westaby et al, 1984), hyperdynamic arteries in association with a cardiac lesion causing large left to right shunt, and bronchogenic cysts. Cystic hygroma may also produce tracheal compression if it lies in the upper mediastinum and particularly if there is sudden haemorrhage into it which can occasionally occur.

Vascular rings occur in various types and may produce localized pressure on the trachea and oesophagus. This may result in secondary tracheomalacia, which may be persistent postoperatively (Roesler et al, 1983). Vascular rings take various forms including double aortic arch 54%, right aortic arch and left ligamentum arteriosum 16%, anomalous subclavian artery 12%, pulmonary artery sing 12%, anomalous innominate artery 6%. Treatment is operative to re-route the aberrant vessels and to allow natural growth of the trachea and recovery of the tracheomalacia.

**Lung abnormalities**

**Congenital lung cysts**

Congenital lung cysts may be found incidentally on the chest X-ray when it is taken for other reasons, but are more commonly noticed when they become infected. They may be difficult to differentiate from pneumatoceles, particularly in staphylococcal infection. After
the acute phase they are persistent whereas infective pneumatoceles usually disappear with
time. They may also be seen in a sequestrated lobe of the lung which has become infected
and this too must be excluded by subsequent investigation using ventilation perfusion lung
scan. Treatment is by surgical removal since there is a high risk of recurrent infection for
these children in the future.

**Cystic adenomatoid malformation**

This abnormality is often found in the neonatal period when it may be associated with
inability to establish normal breathing. It is also seen as a persistent shadowing on the chest
X-ray, either as an area of apparent non-aeration or with multicystic lesions present. These
are most commonly found in the right middle lobe, although the upper lobes may also be
affected (Yu, 1986b). Treatment is surgical in all cases after resolution of any underlying
infection. The prognosis after the removal of the cyst is usually extremely good and the rest
of the lung compensates well for the area which has been removed.

**Congenital lobar emphysema**

This condition is thought to be due to an abnormality of the bronchial or mucosal
lining of the associated airway. It is most frequently due to abnormal development of the
cartilage within the airway, but may be secondary to extrinsic airway compression by a cyst,
tumour or a blood vessel. The lobe progressively overinflates and presses on the surrounding
lung. This will result in respiratory distress, wheezing and reduced air entry over the affected
lobe. Most cases present in infancy, but a few are found incidentally in later childhood and
are relatively asymptomatic at this time. The lobes are affected with the following frequency,
left upper, right middle, right upper, and rarely the lower lobes. A significant proportion of
patients have associated cardiac disease and this should be excluded by echocardiography.

Treatment consists of lobectomy in those who have significant symptoms, particularly
in the younger child. Those with few symptoms may be treated conservatively and a number
will improve with natural growth and development. The long-term lung function after surgery
is good (McBride et al, 1980).

**Lobar sequestration**

These are areas of non-functioning lung tissue derived from abnormal embryonic
development. They have aberrant connections to the tracheobronchial tree and an abnormal
blood supply. They may be divided into two major groups - extralobar and intralobar. The
extralobar type, which has its own separate pleura and an arterial blood supply usually from
the aorta, may communicate with the trachea or bronchi or occasionally with the gut.
Intralobar sequestrations lie within the visceral pleura and are intimately associated with
normal lung tissue. They may have tracheobronchial communication, but usually ventilate
extremely poorly, they also have a systemic blood supply. Repeated infection in these lobes
is not uncommon. On dynamic imaging they show extremely poor ventilation and no
perfusion because they are not supplied from the pulmonary vascular bed. Treatment is by
surgical removal, since repeated infections commonly occur in these lesions.
Pulmonary agenesis

Unilateral pulmonary agenesis may be asymptomatic and found on incidental chest X-ray. There is, however, a significant incidence of related congenital malformations particularly affecting the vertebral column and the cardiovascular system. Chest X-rays show an opaque hemithorax with the heart shifted to the affected side. There is no specific treatment and the prognosis for lung function is good since the unaffected lung usually compensates to a very significant degree.

Bilateral pulmonary hypoplasia occurs in relation to a number of neonatal problems including renal agenesis (Potter's syndrome). It is also seen after prolonged amniotic fluid leak, intrauterine hydrops and skeletal abnormalities such as asphyxiating thoracic dystrophy. The outlook in this condition depends to a great extent on the degree of hypoplasia that is present. Severe cases are incompatible with life. Unilateral pulmonary hypoplasia occurs in association with diaphragmatic hernia.

Rib cage abnormalities

Abnormalities of the rib cage occur in a number of neonatal syndromes, including asphyxiating thoracic dystrophy, captomelic dwarfism, hypophosphatasia and osteogenesis imperfecta. Many of these are lethal in the neonatal period, although a number of cases have survived beyond this time with intensive respiratory support and surgery to enlarge the chest cavity.

Neuromuscular disease

A number of infants are born with congenital neuromuscular disease which may be present at birth. Some infants have similar symptoms following severe birth asphyxia. The most common disease of this type is congenital spinal muscular atrophy (Werdnig-Hoffman disease), which presents with weakness, poor respiratory effort, feeding problems and often a history of poor intrauterine fetal movements. Most infants die of respiratory failure secondary to associated swallowing difficulties and recurrent aspiration. Other conditions producing similar problems include neonatal myotonic dystrophy and congenital myasthenia gravis.