Paediatric
Respiratory Reviews

The neonatal chest X-ray

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Summary   The chest X-ray is the most valuable imaging modality in the assessment of the neonate with respiratory distress. Whilst many of the radiological appearances are relatively non-specific, integration of the clinical features with the X-ray appearances will help the clinician arrive at the correct diagnosis in most cases. In a minority of infants, particularly those with a congenital malformation of the chest or airways other imaging modalities may be required. This paper will describe the radiological appearances of the most important causes of neonatal respiratory distress and highlight those situations where more complex investigations are necessary. © 2001 Harcourt Publishers Ltd

INTRODUCTION

The causes of respiratory distress in the neonatal period can be classified into abnormalities primarily affecting aeration, circulation or development of the thorax.[1] Conditions of the lungs and cardiovascular system account for the majority but abnormalities of the tracheobronchial tree, chest wall, diaphragm and a variety of neuromuscular diseases must also be considered in the differential diagnosis. Chest radiography is the most valuable imaging modality in the investigation of neonatal respiratory disorders, although ultrasound scanning may be useful to confirm the presence of pleural fluid and to evaluate a suspected 'mass' lesion lying adjacent to the chest wall or diaphragm. Other modalities including fluoroscopy, computed tomography (CT) and magnetic resonance imaging (MRI), nuclear medicine studies, bronchography and angiography may occasionally be invaluable.

Radiographic technique

Radiographic examination of the critically ill newborn infant can be a daunting task for any radiographer. Close cooperation between the radiographic, nursing and medical staff with mutual understanding of the needs of the baby

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and the demands for high quality radiography should help to overcome many of these difficulties. It is important to keep the exposure to ionising radiation to a minimum to reduce the small but important risk of malignancy associated with X-ray examinations.[2] However, the risk must be kept in perspective and balanced against the risk of missing an important diagnosis or complication of treatment by failing to perform an X-ray or CT examination.[3]

The major role of the chest X-ray in these infants is not only to confirm or exclude a suspected diagnosis but also to check for the position of the various tubes and catheters used in intensive care. The chest X-ray is also used to monitor pulmonary inflation, particularly in infants who are being ventilated by high frequency oscillatory ventilation and to check for the development of any complications, e.g. the development of air leaks or collapse of the right upper lobe and left lung when the endotracheal tube is too long.

In general, the standard AP supine projection of the chest is sufficient, although the upper two-thirds of the abdomen should be included if the positions of the umbilical venous and arterial catheters are to be checked. The umbilical arterial catheter tip should be above the coeliac axis at the level of D10 or 11 or be lower than the origin of the renal arteries just below L3. The optimum position for an umbilical venous catheter is at D8–9. Prolonged venous access is often maintained by percutaneous (PICC) long lines. These lines should not cross the right atrium and the tips should terminate in the IVC just below the right atrium or in the distal SVC. Malposition
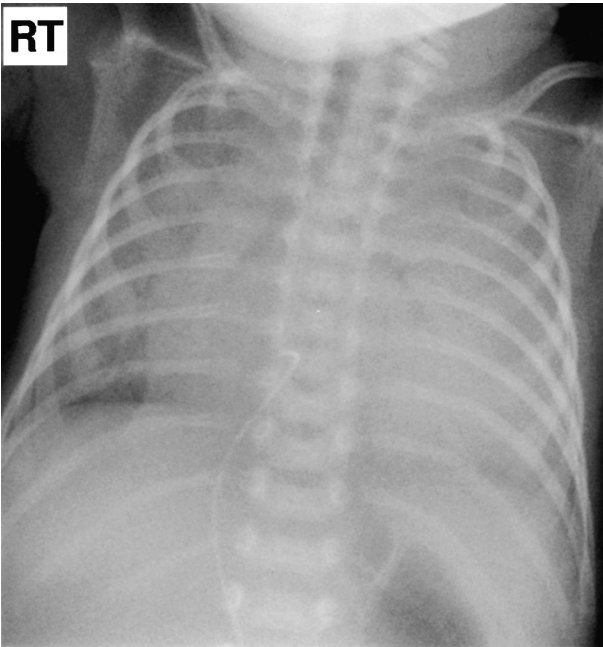
Figure 1 Cardiomegaly due to a large pericardial effusion caused by perforation of the right atrial wall by the angulated tip of central venous catheter.

Figure 2 Transient tachypnoea of the newborn with over-inflated lungs and increased interstitial streaky markings.

of catheters can cause cardiac arrhythmias or pleural and pericardial effusions[4] (Figs. 1, 12).

Radiographic technique, particularly the supine projection, must be taken into consideration when interpreting the neonatal X-ray.[5] Rotation of the baby may erroneously suggest that there is cardiomegaly, mediastinal shift or an increase in translucency of the lung. A number of artefacts may also give rise to confusion, e.g. the hole in the Perspex top of the incubator when projected over the lung fields may simulate a bulla and skinfolds may mimic pneumothoraces (Fig. 8).

The normal neonatal chest X-ray

Aeration of the normal neonatal lung is virtually complete within two or three respiratory cycles after birth and the lung fields should appear symmetrically aerated on the initial X-ray with the diaphragms lying at the level of the 8th ribs posteriorly and 6th ribs anteriorly. The heart size is often difficult to evaluate because of the supine AP projection and thymic shadow. The transverse cardiothoracic ratio should be <60%. An absent thymic shadow in the sick neonate is more likely to be related to involution of the thymus because of stress rather than thymic aplasia associated with Di George syndrome.

Transient tachypnoea of the newborn

Within the first 3 h of life the normal neonate may show variable degrees of retention of fetal fluid in the lungs. Delayed clearance of this fluid may be associated with an increased respiratory rate and a minor reduction of oxygen saturation. Transient tachypnoea of the newborn, or 'wet lung disease', is more commonly seen following caesarian section birth and is associated with maternal diabetes. The chest X-ray shows normally or slightly over-inflated lung fields with increased interstitial streaky shadowing extending into the peripheral lung fields in association with a normal or mildly enlarged heart and occasionally small pleural effusions. The radiographic appearances may be difficult to distinguish from those caused by infection or cardiac failure but the clinical course is benign, gradually improving by 24–48 h of age (Figs 2, 16).

DISORDERS OF AERATION

The majority of diseases affecting aeration of the neonatal lung are associated with abnormal pulmonary opacities on the chest X-ray (Tables 1 and 2). However, conditions primarily affecting the mechanics of ventilation, e.g. neuromuscular disorders, may show essentially normal lung markings (Table 3).

Table 1 Causes of bilateral increase in pulmonary opacity.

Poor inspiration
Transient tachypnoea of the newborn
Idiopathic respiratory distress syndrome
Infection-Group B streptococcus
Meconium aspiration syndrome
Pulmonary haemorrhage/oedema
Aspiration
Pulmonary lymphangectasia
Alveolar proteinosis
Bilateral pleural effusions

Table 2 Causes of unilateral increase in pulmonary opacity.

Artefact, rotation
Infection
Aspiration
Pulmonary collapse
IRDS post surfactant therapy
Pleural effusion
Mass lesion, e.g. CCAM, diaphragmatic hernia
Pulmonary agenesis/hypoplasia
Gross cardiomegaly

Table 3 Causes of respiratory distress with 'normal' lungs.

Choanal atresia
Tracheal obstruction, e.g. vascular ring
Persistent fetal circulation
Neuromuscular
Skeletal dysplasia
Abdominal distension-splinting diaphragms
Metabolic acidosis
Drug related

Idiopathic respiratory distress syndrome (IRDS)

IRDS or hyaline membrane disease remains the most important cause of neonatal respiratory distress although the incidence and severity has reduced following the routine administration of steroids to mothers threatening premature delivery. The disorder arises from deficiency of pulmonary surfactant resulting in atelectasis of the alveoli, dilatation of the terminal bronchioles and poor gas exchange. The baby with IRDS develops a grunting respiratory pattern in association with intercostal recession and sternal retraction, which if not immediately obvious or birth may develop over the first 4–6 h. The condition primarily occurs in premature infants of <32 weeks gestation but is occasionally seen in term infants following caesarean section, in infants of diabetic mothers and in infants exposed to perinatal asphyxia.

The X-ray appearances depend on the severity of the disorder, with poorly inflated lungs being the cardinal feature. In mild disease the lungs show fine homogeneous ground glass shadowing (grade1) but when more severe, widespread air bronchograms become visible (grade 2) followed by the development of confluent alveolar shadowing (grade 3) leading to a complete white-out of the lung fields with obscuring of the cardiac border in the most severe cases (grade 4) (Fig. 3).[6,7] The appearances are modified by the degree of ventilatory support, the phase of respiration at the time of radiography and various therapeutic measures. Whilst IRDS classically shows a homogeneous and symmetrical distribution of pulmonary shadowing, the administration of pulmonary surfactant via the ET tube may give rise to a more patchy and heteroge-

neous pattern which is more suggestive of infection rather than IRDS (Fig. 4).[8] A number of complications may occur including air leaks, pulmonary haemorrhages together with non-respiratory complications such as intracranial haemorrhage and hypoxic ischaemic brain damage.

Air leaks

Air leaks are most commonly associated with positive pressure ventilation; they may occur spontaneously or develop following resuscitation. Following alveolar rupture the air tracks along the interstitial spaces and

Figure 3 Severe idiopathic respiratory distress syndrome (IRDS) with confluent alveolar shadowing, air bronchograms (arrow) and an almost complete loss of the cardiac outline. Note the ET tube is too long, with the tip at the carina.
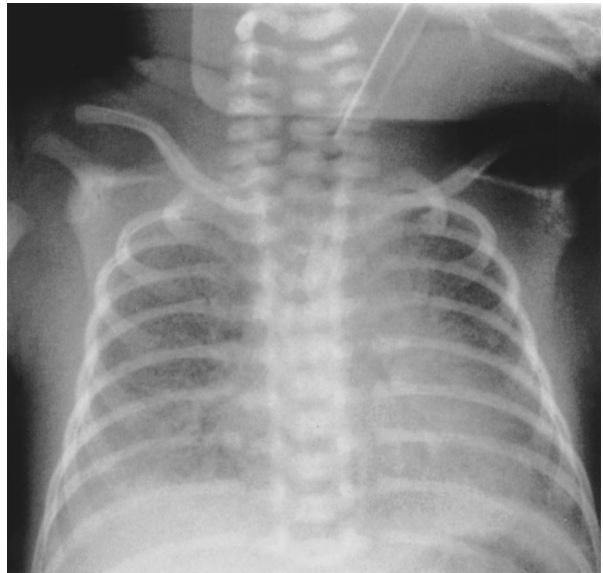

Figure 4 IRDS—incomplete clearing of the lungs following surfactant therapy. More confluent consolidation is present at the right base.

lymphatics and may be seen as pulmonary interstitial emphysema (PIE) prior to the development of a pneumothorax, pneumomediastinum, pneumoperitoneum or pneumopericardium (Figs 5–8). Rarely, intravascular air embolism may occur but this is universally fatal.
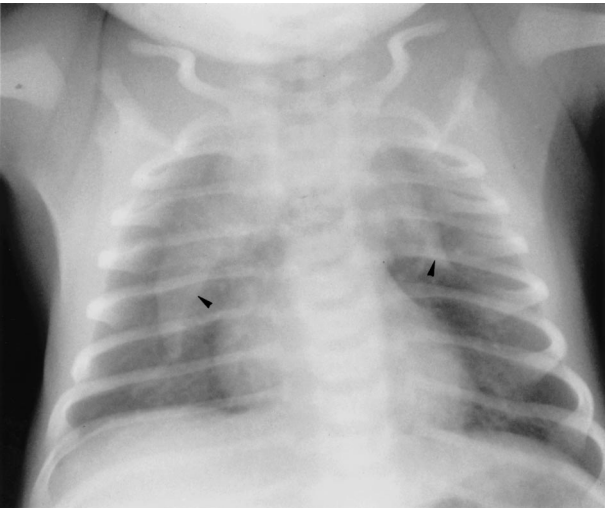

Figure 5   Spontaneous pneumomediastinum. Note the thymus gland outlined by air giving an 'angel's wings' appearance (arrowheads).
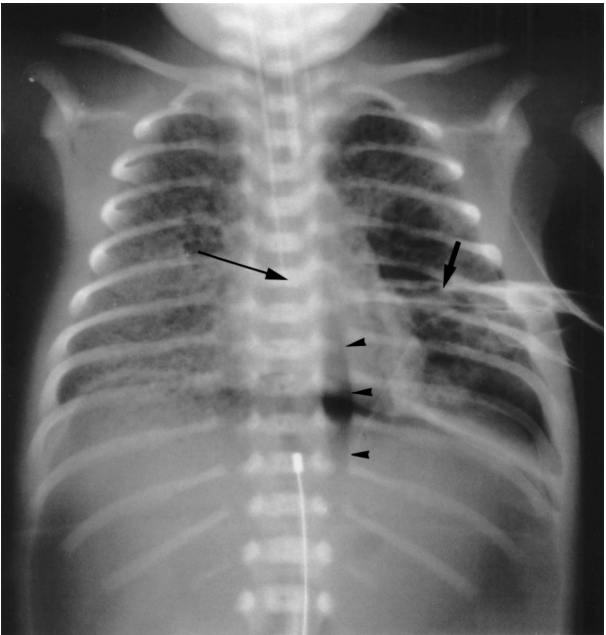

Figure 6   Severe bilateral pulmonary interstitial emphysema, following drainage of left pneumothorax by intercostal drain (short arrow). Note mediastinal air to the left of the spine tracking down to the peritoneal cavity (arrowheads). The diaphragm is visible throughout its length and there is increased translucency over the upper abdomen indicating a large pneumoperitoneum. Note the nasogastric tube is too short with the tip in the mid oesophagus (long arrow).
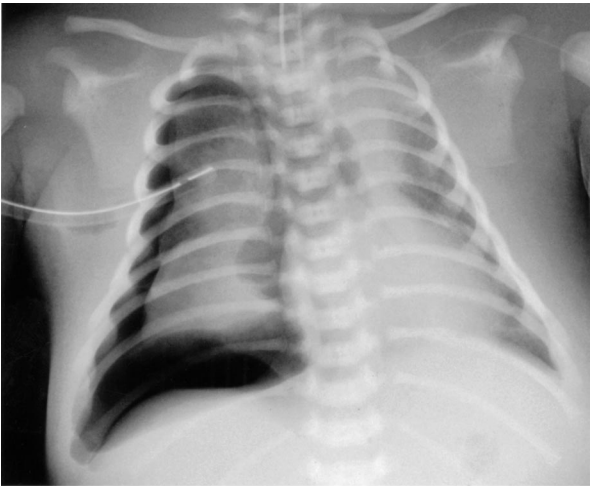
(a)
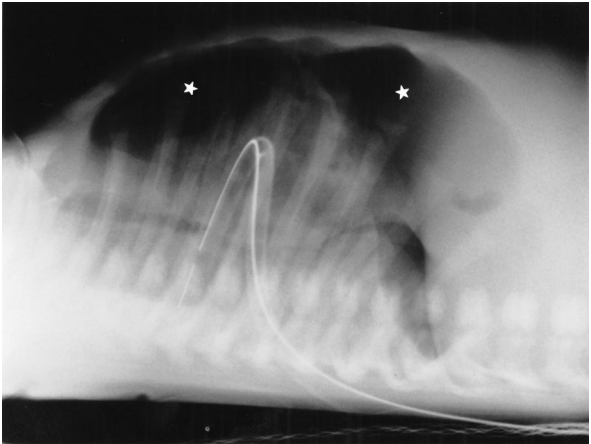
(b)

Figure 7   (a) Right sided tension pneumothorax incompletely drained by the intercostal drain. There is flattening of the diaphragm and shift of the midline with compression of the left lung. (b) Lateral view shows that the chest drain is angulated posteriorly and is not ideally sited to drain the anterior and subpulmonary collection of air (asterisk).
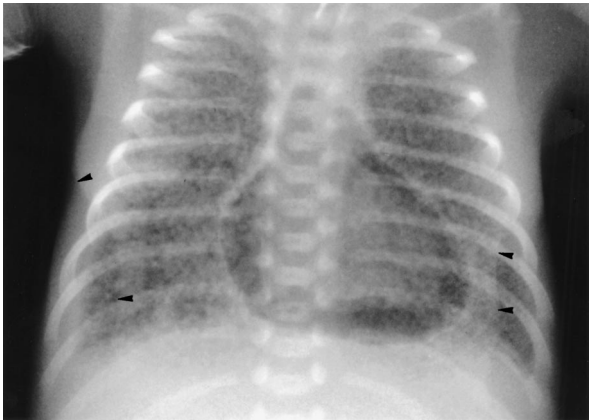

Figure 8   Pneumopericardium has developed in association with bilateral pulmonary interstitial emphysema. Note bilateral skinfolds at bases simulating a pneumothorax (arrowheads).

Pulmonary interstitial emphysema appears as linear or tiny cystic translucencies extending from the hilum to the periphery of the lungs. When more severe, the lungs become hyper-expanded and stiff, compressing the heart and mediastinal structures and reducing venous return.

PIE generally affects both lungs symmetrically but unilateral and even a lobar distribution may be seen. Mediastinal air collects anteriorly and surrounds the thymus displacing the pleura laterally. Air may also collect behind the heart and in the inferior pulmonary ligament and may subsequently track down into the peritoneal cavity. A pneumomediastinum is easily visualised when air outlines the thymus, or causes a radiolucent halo over the cardiac outline (Fig. 5). However, it may be difficult to distinguish from a pneumothorax lying anteriorly. A lateral shoot or decubitus view may help to differentiate the two since air in the mediastinum may surround the thymus whereas a loculated pneumothorax may displace the thymus upwards. Pneumothoraces often develop rapidly resulting in marked tension displacing the mediastinum, particularly when they occur in association with lungs stiffened because of PIE.

Pneumothoraces are generally easy to detect radiographically as the lung edge becomes visible but they may be more subtle when found in a subpulmonary position the diaphragm or causing depression of loculated anteriorly.[7] A lateral shoot-through X-ray can be particularly useful to check the position of where the chest drain in relation to the collection of air, when the pneumothorax persists following the insertion of an intercostal drain (Fig. 7).[7]

Pulmonary haemorrhage

Pulmonary haemorrhage is a severe form of pulmonary oedema which most frequently accounts for a sudden deterioration in an infant with idiopathic respiratory distress syndrome. The diagnosis is usually made clinically as the result of pink frothy haemorrhagic fluid being aspirated from the ET tube. The chest X-ray appearances are variable and range from symmetrical homogeneous to patchy heterogeneous shadowing and in severe cases may show a 'white-out'.

Meconium aspiration syndrome

Meconium aspiration syndrome most commonly occurs in term or post-mature babies following inhalation of meconium-stained liquor during delivery. Meconium is viscid and causes patchy partial or complete bronchial occlusion which may be associated with the development of a severe inflammatory response and secondary surfactant deficiency. The chest X-ray shows widespread patchy infiltrates associated with peripheral hyperinflation
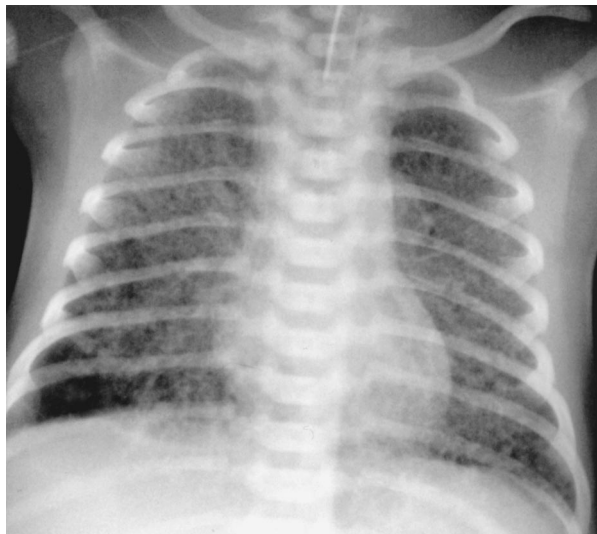

Figure 9   Meconium aspiration syndrome with heterogeneous pulmonary infiltrates in association with hyperinflated lungs. Note the area of focal hyperinflation in the right costophrenic angle.

sometimes in association with a pleural effusion (Fig. 9). Ventilatory support is often required and air leaks, pulmonary haemorrhage and persistent pulmonary hypertension may develop.

Neonatal pneumonia

Neonatal pneumonia occurs following infection with a number of organisms either acquired transplacentally, following inhalation of infected amniotic fluid or following contamination from the birth canal during delivery. Commonly encountered organisms include group B streptococci, *Escherichia coli* and *Staphylococcus aureus*, although viral and fungal infections also occur. The chest X-ray may show patchy or confluent pulmonary infiltrates typical of infection or increased interstitial shadowing (Fig. 10). It may be impossible to differentiate between IRDS, TTNB and cardiac failure on the basis of the chest X-ray, particularly in infants with Group B streptococcal infection. However, infants with pneumonia generally show marked systemic illness in contrast to infants with IRDS and TTNB.

Congenital alveolar proteinosis

Congenital alveolar proteinosis is most commonly related to a deficiency of surfactant protein-B which may be inherited as an autosomal recessive condition. Although rare, this condition should be considered in term infants where there is failure of the respiratory distress to improve after 1 week, particularly when infection and cardiac disease have been excluded. The chest X-ray shows a diffuse granular pattern with air bronchograms as in
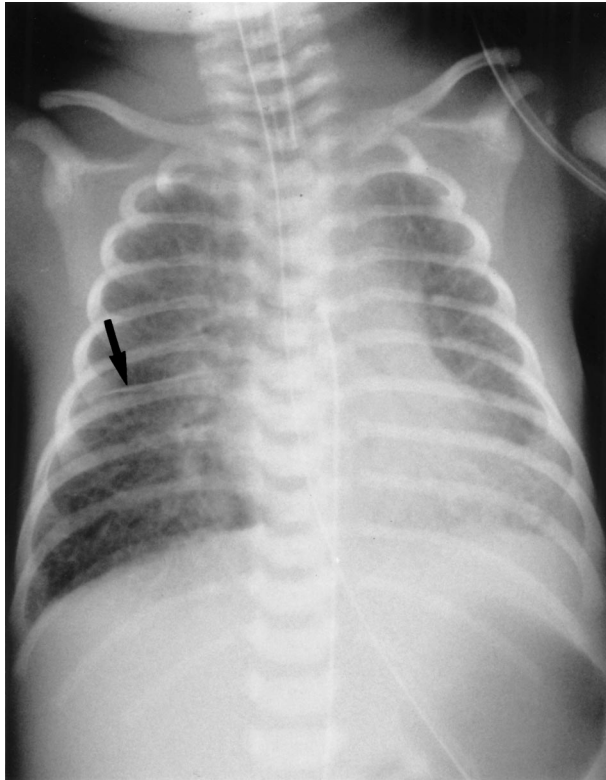
Figure 10 Group B streptococcal pneumonia. Widespread interstitial shadowing with more confluent alveolar consolidation at the left bases and fluid in the horizontal fissure (arrow).

classical IRDS. High resolution CT demonstrates diffuse ground glass shadowing with prominence of the interlobular septa typically seen in alveolar proteinosis in older patients.[9,10] Diagnosis can be confirmed by lung biopsy but in infants with an appropriate clinical history and typical X-ray appearances, analysis of alveolar lavage and peripheral blood samples for surfactant protein B, combined with the demonstration of typical appearances on HRCT may be sufficient to clinch the diagnosis. It may be possible to provide respiratory support until lung transplantation becomes possible but the results have not been universally successful and compassionate withdrawal of care should also be considered.

Pleural effusions

Pleural effusions compress the lung tissue and prevent normal ventilation and aeration. The aetiology of many pleural effusions remains unresolved but infective, lymphatic and cardiac causes should be considered. A pleural effusion may manifest as a generalised increase in opacification throughout the hemithorax, although when sufficiently large, pleural fluid may be seen to displace the lung edge away from the costal margin (Fig. 11). Unilateral effusions may cause mediastinal shift to the contralateral side. The presence and size of a pleural effusion can be confirmed by an ultrasound examination, at which
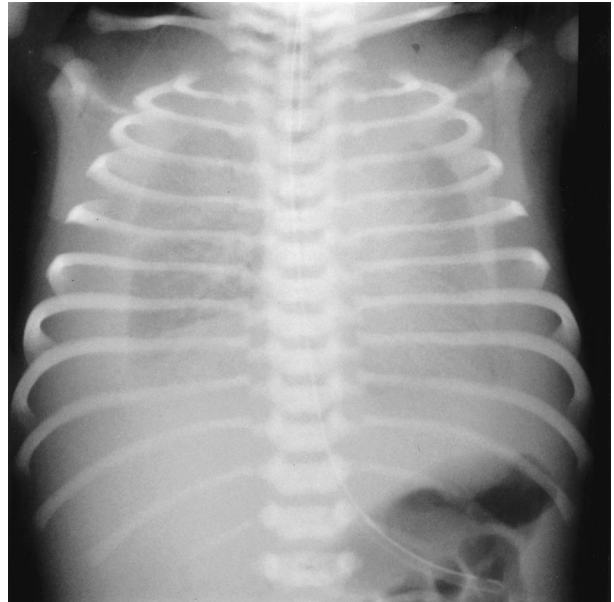

Figure 11 Large bilateral pleural effusions of unknown aetiology, which resolved spontaneously. Note the lung edge is displaced away from the chest wall.

time a suitable site for drainage can be identified. Chylous effusions may also occur in association with pulmonary lymphangiectasia, a rare disorder which may be associated with congenital heart disease or generalised lymphatic abnormalities, e.g. in Turner's and Noonan's syndrome. Pulmonary lymphangiectasia is characterised by diffuse coarsely nodular or reticular pulmonary shadowing.[7,11]

Neuromuscular disorders

Neuromuscular disorders give rise to reduced ventilation due to poor respiratory drive or weakness of the thoracic cage muscles. The chest X-ray may appear normal or show areas of collapse which may recur. Recurrent aspiration may also result in repeated episodes of consolidation. Pointers on a chest radiograph to an underlying neuromuscular aetiology include a bell-shaped thorax, thin ribs, a scoliosis and eventration of the diaphragm (Fig. 12). MRI scanning of the brain and brainstem should be considered in those infants thought to have a central neurological cause for hypoventilation.

Chronic lung disease

Chronic lung disease is defined as any infant requiring oxygen supplementation beyond 28 days of age or 36 weeks post-conceptual age in which the chest X-ray is persistently abnormal. A number of disorders may be categorized under the term chronic lung disease, including broncho-pulmonary dysplasia (BPD), chronic pulmonary
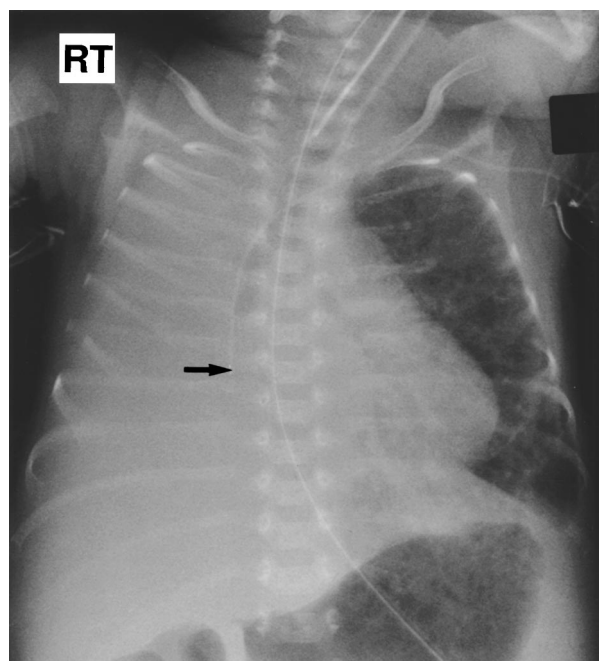
Figure 12 Congenital myotonic dystrophy. Note collapse of the right lung. The left lung is over-inflated showing increased reticular markings. The ribs are exceptionally slender indicating probable neuro-muscular aetiology for the infant's respiratory distress. PICC line is too long terminating in the right atrium and should be withdrawn 1–2 cm into the SVC (arrow).

Figure 13 Broncho-pulmonary dysplasia—age 4 weeks. There is an increase in reticular markings (linear/streaky), which have gradually developed following IRDS in the early neonatal period. The lungs are mildly over-inflated.

DISORDERS OF CIRCULATION

Persistent fetal circulation (PFC) or the presence of congenital heart disease must be considered in those infants in whom high levels of inspired oxygen fail to raise the arterial oxygen saturation or in whom there is clinical evidence of cardiac failure.

Persistent fetal circulation

Persistent fetal circulation, i.e. persistent pulmonary hypertension, occurs in association with a number of underlying causes including birth asphyxia, neonatal pneumonia, meconium aspiration syndrome, congenital diaphragmatic hernia and sepsis. The normal levels of pulmonary arterial pressures seen *in utero* fail to fall postnatally, resulting in a right to left shunt at atrial level through the persistent foramen ovale. In PFC the chest X-ray is often unremarkable, in contrast to the severe degree of hypoxaemia present, although the lung fields often show decreased pulmonary vascularity. Echocardiography should be performed to exclude congenital heart disease.

Congenital heart disease

A discussion of neonatal presentation of congenital cardiac disorders is outside the scope of this article. The chest X-ray appearances of infants with congenital heart disease are variable depending on the nature of the defect. Alteration in heart size and contour, the presence of increased or decreased pulmonary vascularity and evidence of pulmonary oedema should be looked for (Figs 14–16). The situs of the heart and abdominal viscera should also be noted.[13] Although the majority of

insufficiency syndrome of prematurity (CPIP) and Wilson-Mickity syndrome. The distinction between the various entities is not always clear, although CPIP and Wilson-Mickity disease generally commence towards the end of the first week of life in infants who have not been previously ventilated. BPD follows treatment with mechanical ventilation, particularly in infants with hyaline membrane disease. Aetiology is multifactorial but barotrauma, oxygen toxicity and infection are thought to be the most significant factors. The classical appearance of BPD, as first described by Northway, is characterised by overinflated lungs with the development of a marked cystic appearance due to focal areas of hyperinflation interspersed with adjacent areas of atelectasis.

Nowadays these 'cystic' appearances are less frequently seen and more commonly infants with BPD develop patchy streaky shadowing with scattered areas of emphysema in association with mild hyperinflation (Fig. 13). The radiographic appearances may gradually improve, but infants with chronic lung disease there is a high morbidity, develop recurrent episodes of infection, gastro-oesophageal reflux and bronchospasm and an increased risk of sudden death syndrome.[6,12] These infants have high nutritional requirements and may develop metabolic bone disease of prematurity. The development of rib fractures may contribute to respiratory failure.
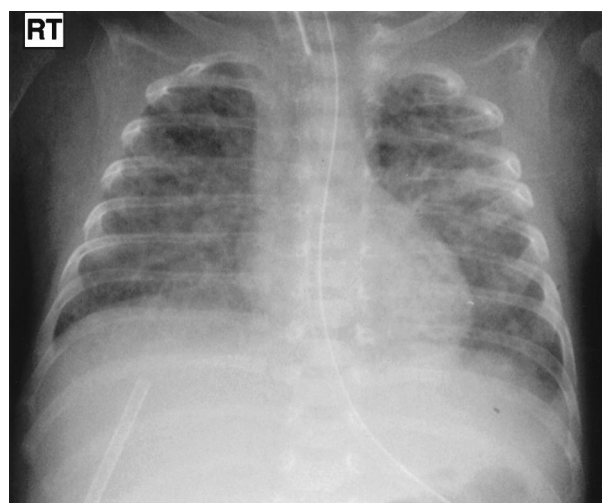
Figure 14 Pulmonary atresia. The lung fields are hyperinflated and show a marked decrease in vascularity (pulmonary oligaemia). The heart is not enlarged.
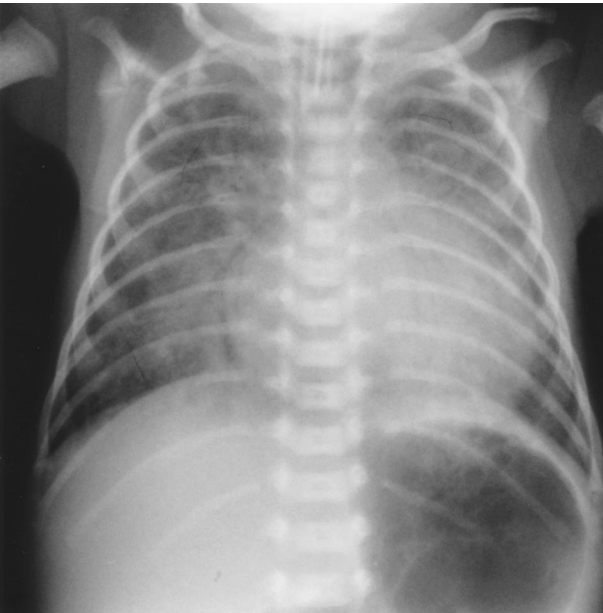

Figure 15 Patent ductus arteriosus. The heart is enlarged and the lung fields show an increase in vascularity (pulmonary plethora). More confluent consolidation is noted centrally due to the development of pulmonary oedema.

infants have a patent ductus arteriosus during the first few days of life only one third of premature infants will develop a significant left to right shunt to result in cardiac failure. The chest X-ray may show an increasing heart size, pulmonary plethora and possibly the development of increased haziness due to pulmonary oedema. Once
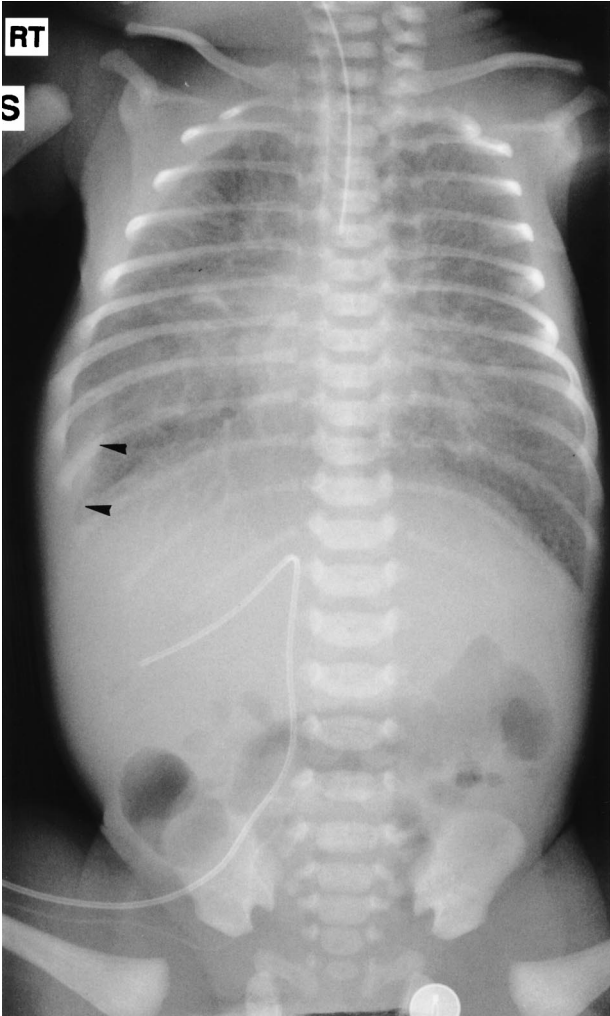

Figure 16 Total anomalous pulmonary venous drainage should always be considered where a small heart is seen with increased interstitial markings of pulmonary oedema. There is a small right sided pleural effusion (arrowheads). Note the misplaced umbilical venous catheter, which has failed to cross the ductus venosus into the IVC.

suspected clinically the diagnosis should be confirmed by echocardiography.

DEVELOPMENTAL ABNORMALITY

Prenatal diagnosis of congenital malformations by ultrasound screening has altered the natural history of many congenital thoracic malformations. Pregnancies may be terminated following the diagnosis of a serious malformation, or infants may present to the paediatric service at an early stage with respiratory distress and signs consistent with the prenatal diagnosis. The radiological presentation of some of the more important congenital malformations will be described below. As a general rule, if a baby presents with significant respiratory distress due to a congenital thoracic

abnormality the chest radiograph shows an obvious abnormality.

Tracheo-oesophageal fistula (TOF) and oesophageal atresia (OA)

TOF and OA present early with excessive secretions or with episodes of intermittent cyanosis. A diagnosis is made by the failure to pass a naso-gastric tube and confirmed by AP and lateral radiography. The chest X-ray may show together a dilated upper oesophageal pouch with right upper lobe consolidation following episodes of recurrent aspiration. Babies with a TOF are likely to demonstrate excessive bowel gas. Infants with OA and TOF may have associated anomalies such as the Vacteral association, including vertebral, rib and radial anomalies, congenital heart disease, renal anomalies and ano-rectal atresia.

Congenital diaphragmatic hernia

CDH most commonly occurs through a postero-lateral defect (Bochdalek foramen) in the diaphragm. CDH occurs more frequently on the left. Typical radiographic findings are of 'cystic' radiolucencies, due to the presence of air in herniated bowel with contralateral deviation of the mediastinum (Fig. 17). If the X-ray is taken very early before air has replaced fluid in the bowel the hemithorax will appear opaque. Some right-sided hernias may also present as dense opacity in the lower right hemithorax,
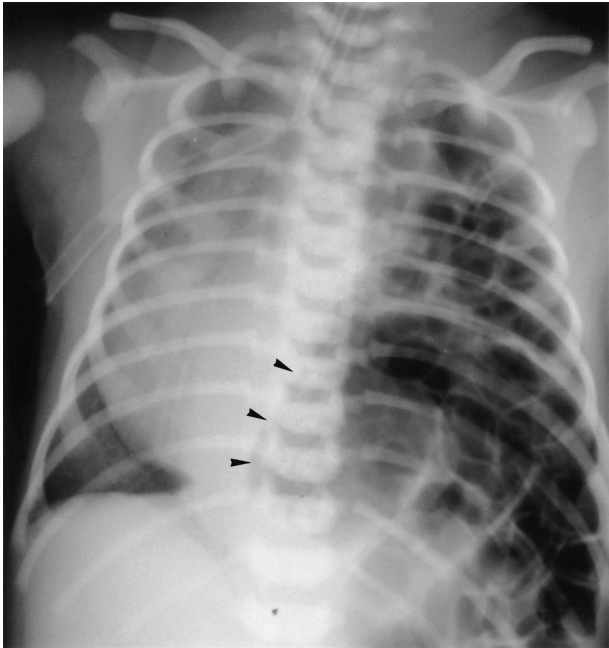

Figure 17 Congenital diaphragmatic hernia. The bowel in the left hemithorax is displacing the mediastinum to the right and compressing the right lung. The stomach is intrathoracic (arrowheads) A right sided pneumothorax has been drained.

when the liver and kidney rather than bowel is contained in the hernia. The presence of the stomach in the chest is said to be associated with an early intrauterine herniation and poorer prognosis but this is not invariably the case. These appearances must be distinguished from those of cystic adenomatoid malformation of the lung. The abnormal orientation of the bowel loops and even size of the 'cysts' due to the herniated bowel suggest the correct diagnosis.[7,11] Where there is continuing doubt, an ultrasound examination of the chest and upper GI contrast studies can be helpful to confirm the presence of bowel loops in the chest and in difficult cases MRI scanning may also contribute. CDH is often associated with severe pulmonary hypoplasia and persistent fetal circulation, which may ultimately determine the outcome. The presence of visible aerated lung on the ipsilateral side to the hernia is associated with a good outcome, whereas the finding of a pneumothorax on the contralateral side is associated with a grave prognosis (Fig. 17).[14] Intensive care, possibly with ECMO, is required until the infant has stabilised after which time surgical repair can be undertaken.

Congenital cystic adenomatoid malformation of the lung (CCAM)

In the majority of infants diagnosed prenatally the lesions may regress prior to birth.[6,11] The infants are often asymptomatic and a postnatal chest X-ray may be normal although CT scanning generally demonstrates a small residual lesion. The need for surgery to prevent any long-term consequences, e.g. risk of tumour development or severe infection, is a matter of debate. Infants with larger lesions may present with early signs and symptoms of respiratory distress. CCAM commonly affects a single lobe, but several lobes or the whole lung may be involved and when severe may be associated with pulmonary hypoplasia. The chest X-ray will show a mass with cysts of varying sizes and density depending on the relative fluid content of the cysts. In severe cases there may be mediastinal shift. Treatment is surgical in symptomatic infants.

Pulmonary sequestration

Pulmonary sequestration[6,11] is a foregut malformation where a segment of lung parenchyma develops in isolation from the rest of the lung, lacking connection with the tracheo-bronchial tree and generally having a systemic blood supply. Infants may present with respiratory distress due to a large left to right shunt or with a pleural effusion; some diagnosed prenatally may remain asymptomatic. The chest X-ray may be normal or show increased basal opacity due to the sequestrated segment which if large enough will result in a degree of mediastinal shift (Fig. 18a, b and c). An echogenic mass in association with an abnormal arterial supply and venous drainage can
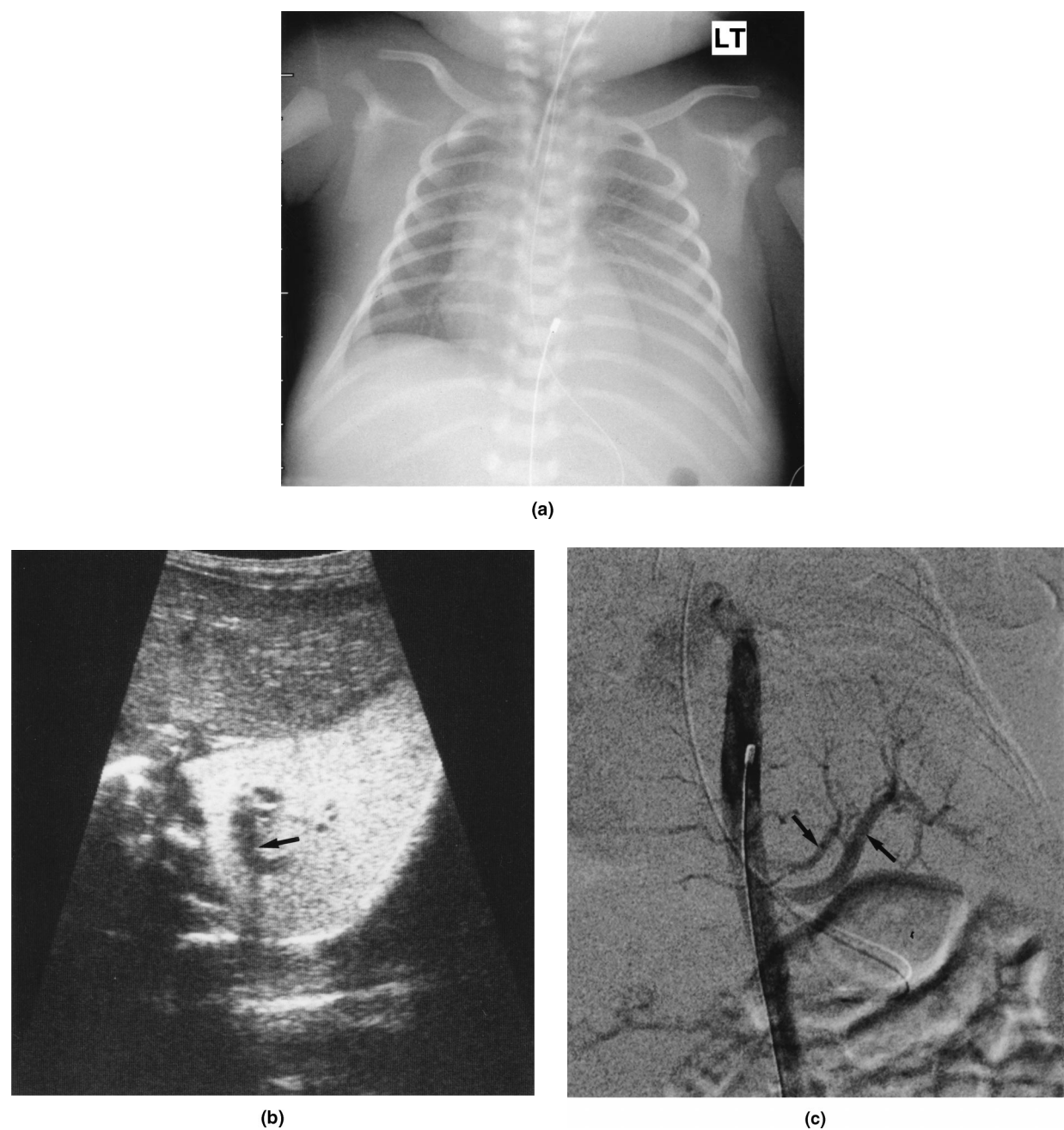
(a)

(b)

(c)

Figure 18   Extralobar pulmonary sequestration in a premature thought clinically to have IRDS. (a) There is increased density at the left base with mild displacement of the mediastinum to the right, in addition to more generalised ground glass shadowing of IRDS. (b) Transverse section of an ultrasound examination through the upper abdomen and lung base shows an echogenic mass at the left base with abnormal vessels (arrow) arising from the aorta and draining to the azygos system. (c) Aortogram showing two arteries arising from the aorta to supply the pulmonary sequestration (arrows).

sometimes be demonstrated by ultrasound scanning. Although CT or MRI can be useful for further delineation of the lesion some cardiothoracic surgeons still request angiography accurately to demonstrate the vascular connections.

Congenital lobar emphysema

Congenital lobar emphysema[6,11] most frequently involves the left upper, the right upper or the right middle lobe.

The aetiology of lobar emphysema is sometimes due to polyalveolar lungs and at other times to an intrinsic abnormality of the tracheo-bronchial tree such as bronchomalacia or a vascular ring compressing the main bronchus. Chest X-ray appearances are usually diagnostic. The affected lobe is often markedly over-aerated compressing the adjacent normal lung (Fig. 19). The abnormal lobe may appear as an opaque mass at birth until retained fluid has been replaced by air. Ventilation perfusion isotope scans can be useful to demonstrate obstructive
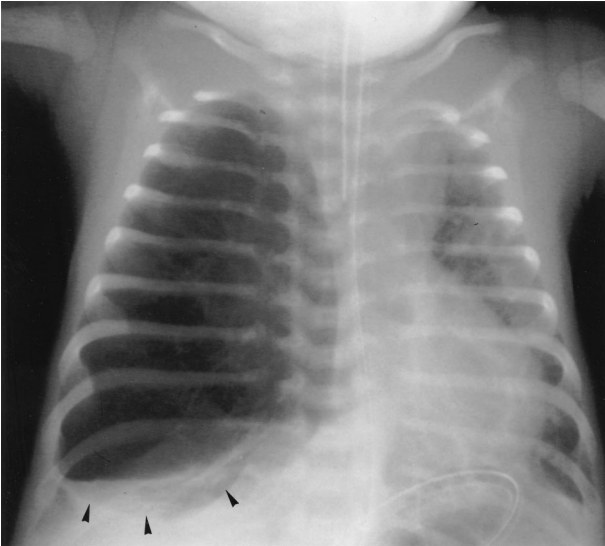
Figure 19 Right upper lobe congenital lobar emphysema. There is gross over-expansion with marked decrease in vascularity of the right upper lobe. The right lower and middle lobes (arrowheads) are compressed and the mediastinum is shifted to the left.

emphysema where there is diagnostic doubt and CT scanning may be useful to exclude the possibility of a vascular ring or duplication cyst compressing the affected bronchus.
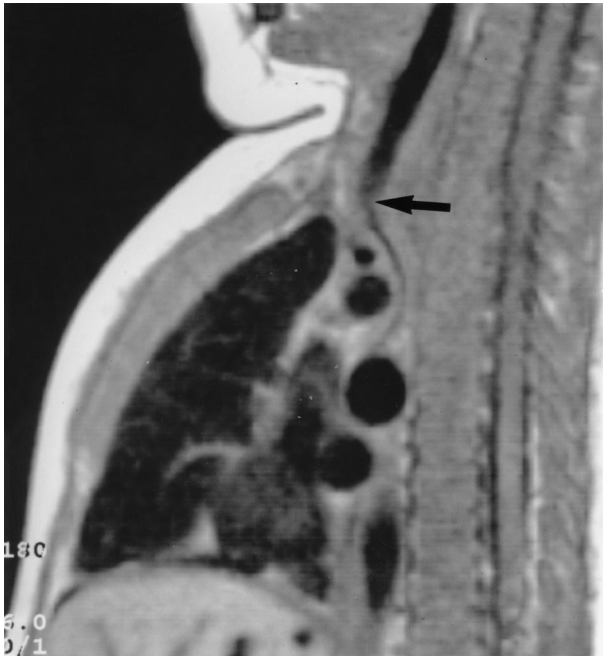
Pulmonary agenesis and hypoplasia

Unilateral pulmonary agenesis[7,11] is associated with other congenital anomalies particularly of the cardiovascular system and tracheo-bronchial tree (Fig. 20b). The characteristic appearances are of gross shift of the mediastinum with an opaque hemi-thorax and ipsilateral rib crowding. It is important to confirm the diagnosis by demonstrating absence of the main bronchus and pulmonary artery, since rarely complete collapse of the lung due to mucus plug obstruction can mimic pulmonary agenesis. A number of strategies can be used to confirm the diagnosis depending on local expertise. These include bronchoscopy, echocardiography, bronchography, isotope studies, CT and MRI (Fig. 20a and b).

Unilateral pulmonary hypoplasia may occur in isolation or be associated with anomalies such as pulmonary sequestration and congenital diaphragmatic hernia. The plain radiograph demonstrates a small lung with reduced vascularity and displacement of the heart to the hypoplastic side. Fluoroscopy or nuclear medicine studies may be useful to exclude air trapping in the normal lung, which will show compensatory overinflation. Bilateral pulmonary hypoplasia is generally associated with severe oligohydramnios, e.g. due to renal agenesis and often presents with early respiratory distress. The chest X-ray may demonstrate a bell-shaped thorax. The lungs may show increased pulmonary shadowing; pneumothoraces and pneumomediastinum commonly occur.
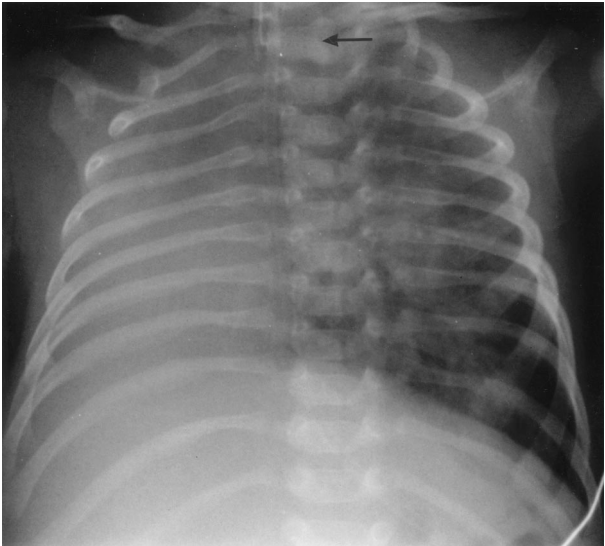
(a)

(b)

Figure 20 Right pulmonary agenesis in an infant presenting with stridor due to associated tracheal stenosis and complex congenital heart disease. (a) The right lung is opaque and the heart has shifted into the right hemithorax. The left lung shows compensatory hyperinflation. There is a segmentation anomaly of the second dorsal vertebrae (arrow). (b) MRI scan in the sagittal plane demonstrates a severe tracheal stenosis commencing at the thoracic inlet (arrow). (from Dr M. McCarty)

Upper airway obstruction

This[15] may uncommonly occur due to tracheal stenosis (Fig. 20b) or compression, caused by a congenital vascular ring or an intraluminal abnormality within the trachea such as a haemangioma. Upper airway obstruction results in progressive overinflation of the lungs which at times
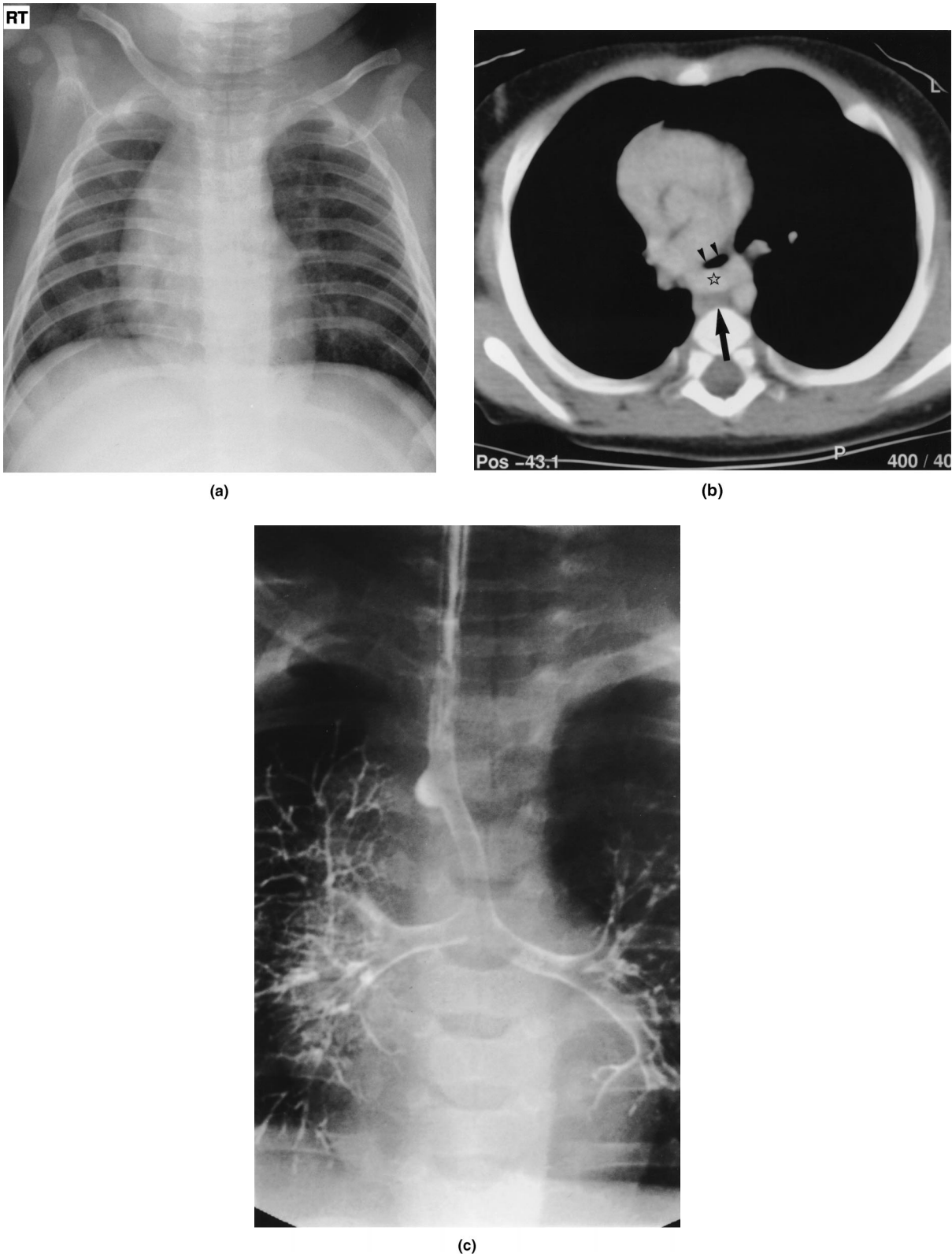
(a)

(b)

(c)

Figure 21   Pulmonary artery sling and tracheal stenosis. (a) Chest X-ray demonstrates hyperinflation of the left hemithorax with slight shift of the mediastinum to the right. (b) CT scan shows the left pulmonary artery arising from the right pulmonary artery (asterisk) passing posteriorly to the left between the trachea (arrowhead) and oesophagus (arrow) before passing over the left main bronchus. (c) Bronchogram. Note the diameter of the trachea is less than the main bronchi. The carina is widened, a feature associated with tracheo-bronchial anomalies. There is a right tracheal diverticulum. Despite the slight overinflation of the left lung there is no obvious compression of the left main bronchus.

can give rise to severe respiratory distress. Urgent diagnosis is essential. This may be achieved by a combination of bronchoscopy, barium swallow, or CT/MRI scanning (Fig. 21). Angiography may be required for pre-operative assessment of vascular rings.

Skeletal anomalies

Although rare, disorders of the skeleton may give rise to respiratory distress. Bone dysplasias, particularly those associated with short ribs and narrow thorax, e.g. asphyxiating thoracic dystrophy, may be associated with pulmonary hypoplasia and difficulty in ventilation. Infants with costo-vertebral anomalies or severe arthrogryphosis may also have reduced respiratory excursion giving rise to respiratory distress.

PRACTICE POINTS

- High quality chest radiography is essential.
- Consider radiographic techniques and artefacts when interpreting the chest X-ray.
- Check the position of all catheters, tubes and wires and record on the film if the position is subsequently corrected.
- Integration of clinical and radiological features is the key to making the correct diagnosis.
- Consider less common causes of respiratory distress when the chest X-ray is normal or findings are atypical.

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