Congenital abnormalities of the neonatal thorax: a pictorial review.

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Objectives

Objectives:

We present a pictorial review covering a range of congenital thoracic abnormalities seen at our large tertiary referral centre including diaphragmatic hernias, atresias and various cystic abnormalities but excluding cardiovascular abnormalities. The anatomy and embryology are illustrated where relevant and we include examples of images from modalities including plain film, ultrasound, CT scans and MRI scans including antenatal images. The radiological findings are described and the pertinent radiological features are highlighted to increase confidence in reporting of such conditions.
Body

Background:

There are a number of congenital abnormalities that can affect the neonatal thorax involving the respiratory, gastrointestinal and cardiovascular systems. These abnormalities are often extremely difficult to diagnose clinically and therefore the role of imaging remains vital. They are important causes of morbidity in newborns and with the advancement of neonatal surgery, imaging both antenatally and postnatally can assist in earlier accurate diagnoses to allow the evaluation of therapeutic options and planning of surgery. We review a range of congenital thoracic pathologies from our current experience in a large tertiary referral centre.

Congenital pathologies:

LUNG AGENESIS

Agenesis of the lung is a congenital absence of one or both lungs. Lung agenesis is less common than aplasia: these are differentiated by the complete absence of the main bronchus, pulmonary vessels and lung tissue in agenesis and presence of a rudimentary bronchus in aplasia. Lung agenesis is more common on the left side and survival rate is better in these cases, since the right lung is the largest.

It can be associated with other congenital abnormalities, known by the acronym VACTERL (vertebral abnormalities, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal abnormalities and limb buds). Radiologically, there is mediastinal shift to the affected side, therefore, right lung agenesis may be confused with dextrocardia.

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FOREGUT DUPLICATION CYSTS

Foregut duplication cysts are a group of well defined congenital malformations which contain fluid and are lined by an epithelium and have a layer of smooth muscle. They
include bronchogenic, oesophageal duplication, neuroenteric, pericardial and thymic cysts as well as mature cystic teratomas, lymphangiomas and meningocoeles.

Bronchogenic cysts

These are lined by respiratory epithelium and have smooth muscle, cartilage and mucous gland tissue. They most commonly occur at the carina. Most are asymptomatic but can present due to compression of adjacent structures. Surgical removal is usually performed at our institution.

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Oesophageal duplication cysts

They are attached to but do not communicate with a portion of the gastrointestinal tract. They most commonly occur in the small bowel or distal oesophagus. Presentation may be incidental or related to a palpable mass or compression of adjacent structures, bowel obstruction, ulceration or perforation. Management is usually by surgical removal.

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CONGENITAL DIAPHRAGMATIC HERNIAS (CDH)

CDH is a malformation due to congenital defects in the diaphragm, more commonly seen posteriorly (Bochdalek hernia) and left sided. Depending on the hernia contents, the radiographic appearances can vary between complete opacification of the hemithorax to an air-containing cystic mass. It is associated with lung aplasia/hypoplasia and babies present with respiratory distress at birth. In our institution these are dealt with surgically at an early opportunity depending on the medical stability of the baby. Close long term follow up is then necessary to observe for possible recurrences and complications.

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OESOPHAGEAL ATRESIA AND TRACHEOESOPHAGEAL FISTULA
In oesophageal atresia, the continuity of the oesophageal wall is interrupted, resulting in an atretic segment; the oesophagus is divided into two blind pouches, upper and lower, and may or may not communicate with the tracheobronchial tree, through tracheoesophageal fistulae. The fistula can be distal (more common), H shaped, proximal or, rarely, both proximal and distal.

**Fig. 23**: Anatomical classification of anomalies of the oesophagus and trachea, with approximate incidences. (From: Gross RE. Surgery of infancy and childhood. Philadelphia, PA: WB Saunders, 1953)


Radiologically, there is an air filled pharyngeal pouch (possibly with an indwelling tube) and, in the presence of a fistula, distal bowel gas is seen. Oesophageal atresia is associated with other congenital abnormalities, known by the acronym VACTERL (vertebral abnormalities, anal atresia, cardiac abnormalities, tracheoesophageal fistula, renal abnormalities and limb buds).

**CONGENITAL CYSTIC ADENOMATOID MALFORMATION (CCAM)**

CCAM is an abnormal adenomatoid proliferation which replaces normal alveoli; usually it involves only one lobe. Radiologically, CCAM can have multiple appearances, depending on the histological sub-type.
Fig. 30: Histological classification of Congenital Cystic Adenomatoid Malformations by Stocker et al 1977. Type 1 have one or more large cysts; Type 2 have multiple small cysts of uniform size; Type 3 appear solid but have microscopic cysts.


Type 1 have one or more large cysts; type 2 have multiple small cysts of uniform size; type 3 appear solid but have microscopic cysts. CCAM's although originally at birth may be filled with fluid they communicate with the bronchial tree, and therefore can fill with air giving an air /fluid level. It can be detected antenatally or can present at birth with respiratory distress. The lesions can sometimes dissappear after birth but in our institution
they are usually electively surgically removed within the first year of life when diagnosed antenatally.

**SEQUESTRATION**

Pulmonary sequestration is an area of abnormal pulmonary tissue which does not have a bronchial communication and shows anomalous arterial supply from a systemic artery arising from the aorta; the more frequent location is the left lower lobe. Extralobar sequestrations have a separate pleural lining and are associated with other abnormalities, whereas intralobar sequestrations, which are more common, do not. Babies can present with recurrent chest infections. Radiographically sequestrations appear as radiopaque masses and, as seen also in cases of infection, they may contain gas, resulting in multiloculated cystic masses. In our institution, these are surgically removed usually within the first year when diagnosed antenatally.

**BRONCHIAL ATRESIA**

Bronchial atresia is a congenital abnormality of unknown aetiology where a segmental bronchus does not communicate with a central airway. It can co-exist with a pulmonary sequestration or a bronchogenic cyst and most are assymptomatic. It most commonly occurs in the apical posterior segment of the left upper lobe and secondly most common in the left lower lobe.

On imaging there is focal interruption of a lobar segmental or subsegmental bronchus with peripheral mucous impaction causing a bronchocoele or mucocoele with associated hyperinflation of the obstructed lung segment.

Treatment can be conservative although surgical resection is offered in our institution and often performed if complicated with infection or respiratory compromise.
Relevant References:


Fig. 1: The chest x ray of a day old newborn shows complete opacification of the right hemithorax, the left lung is hyperexpanded and there is mediastinal shift to the right. The diagnosis was found to be a right lung agenesis.
Fig. 2: An axial image of an enhanced chest CT of the same baby viewed on lung windows reveal no definable right lung or right main bronchus. The right hemithorax is small and occupied by the heart and great vessels (the whole mediastinum being shifted to the right). There is overexpansion of the left lung. The findings are that of a right lung agenesis.

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Fig. 3: An axial image of an enhanced chest CT of the same baby viewed on mediastinal windows show a single left pulmonary artery and no definable right pulmonary artery or right main bronchus. The heart and mediastinum is completely shifted to the right side of the thorax. The findings are those of a right lung agenesis.

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Fig. 4: A coronal reformat image of the enhanced chest CT scan of the same baby viewed on mediastinal windows. It demonstrates the single left hilum and the mediastinal shift can be better appreciated. The findings are that of a right lung agenesis.

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**Fig. 5:** A coronal reformat image of the enhanced chest CT scan of the same baby viewed on mediastinal windows. It demonstrates the single left main bronchus and no right main bronchus together with medastinal shift to the right. The findings are that of a right lung agenesis.

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**Fig. 6:** A CXR of a newborn baby showing a posterior mediastinal mass seen behind the heart and mediastinum on the left side of the vertebral column. There is also hyperlucency of the left lung noted. The baby was found to have a foregut duplication cyst (bronchogenic cyst) on further imaging.

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Fig. 7: An axial image of a contrast enhanced CT Scan of the chest of the same baby viewed on lung windows. It demonstrates a posterior mediastinal mass compressing the left main bronchus causing left lung hyperlucency and airtrapping. On resection, pathology confirmed a bronchogenic cyst.

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Fig. 8: An axial image of a contrast enhanced CT Scan of the chest of the same baby viewed on soft tissue windows. It shows a low density, well defined oval mass in the posterior mediastinum compressing the left main bronchus. On resection, pathology confirmed a bronchogenic cyst.

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Fig. 9: A coronal reformat of an enhanced CT scan of the chest of the same baby viewed on soft tissue windows. It demonstrates a low density cystic mass surrounding and compressing the left main bronchus. On resection, pathology confirmed a bronchogenic cyst.

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**Fig. 10:** A transverse ultrasound image of the lower thorax of an asymptomatic baby with an antenatally diagnosed abdominal cystic structure. It shows a well defined anechoic structure displaying increased through transmission seen both above and below the diaphragm. Appearances confirm the cystic nature of the lesion seen at the level of the diaphragm. On resection in the first year of age, pathology confirmed an oesophageal duplication cyst.

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Fig. 11: A longitudinal ultrasound image of the lower thorax of an asymptomatic baby with an antenatally diagnosed abdominal cystic structure. Appearances confirm the well defined anechoic cystic nature of the lesion seen both above and below the level of the diaphragm. The cyst has a "bowel wall signature", with its wall demonstrating alternated hypoechoic and hyperechoic layers, corresponding to mucosa (hyper-) and muscularis propria (hypo-). On resection in the first year of age, pathology confirmed an oesophageal duplication cyst.

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Fig. 12: An axial T2 weighted MRI image of the same baby displays a well defined multiloculated cystic structure in the posterior mediastinum. It extends down within the mediastinum through the hiatus of the diaphragm, interposing between the left lobe of the liver and the stomach. The structure shows again trilaminar walls, appearances are highly suggestive of an oesophageal duplication cyst. The lack of air within the cyst makes a connection with the bronchial tree less likely. On resection in the first year of age, pathology confirmed an oesophageal duplication cyst.

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Fig. 13: A coronal T2 weighted MRI image of the same baby displaying a well defined multiloculated cystic structure in the posterior mediastinum seen crossing the midline. It extends down within the mediastinum through the hiatus of the diaphragm, interposing between the left lobe of the liver and the stomach. The structure shows again trilaminar walls, appearances are highly suggestive of an oesophageal duplication cyst. The lack of air within the cyst makes a connection with the bronchial tree less likely. On resection, pathology confirmed an oesophageal duplication cyst.

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Fig. 14: A sagittal T2 weighted MRI image of the same baby displaying the well defined multiloculated cystic structure in the posterior mediastinum. It extends down within the mediastinum through the hiatus of the diaphragm, interposing between the left lobe of the liver and the stomach. The structure shows again trilaminar walls, appearances are highly suggestive of an oesophageal duplication cyst. The lack of air within the cyst makes a connection with the bronchial tree less likely. On resection in the first year of age, pathology confirmed an oesophageal duplication cyst.

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Fig. 15: Lateral image of a barium swallow investigation in the same baby, showing displacement of the oesophagus anteriorly and with no communication between the oesophagus to the cystic structure. On resection in the first year of age, pathology confirmed an oesophageal duplication cyst.

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Fig. 16: Anteroposterior image of the barium swallow investigation in the same baby. It shows displacement of the oesophagus to the left and no communication between the oesophagus to the cystic structure. On resection in the first year of age, pathology confirmed an oesophageal duplication cyst.

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Fig. 17: A antenatal MRI scan showing a T2 weighted MRI image displaying the fluid filled stomach (bright signal) and small bowel loops in the left side of the chest. Findings are in keeping with a left sided congenital diaphragmatic hernia.

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Fig. 18: A CXR taken on Day 1 on the child with an antenatal diagnosis of a left congenital diaphragmatic hernia. There is opacification of the left lower chest with poor definition of the diaphragm. There are also multiple lucencies seen within it. There is minimal mediastinal shift. The NG tube tip projects above the level where the diaphragm is expected.

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Fig. 19: An axial image of a contrast enhanced CT scan of the same baby visualised on lung windows demonstrating fluid and bowel loops within the left hemithorax displacing lung. The scan was only performed due to deteriorating clinical condition. CT scans are not usually performed in our institution in babies with Congenital diaphragmatic hernias for diagnostic purposes.

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**Fig. 20:** An axial enhanced CT scan image of the chest of the same baby viewed on soft tissue windows. There is both solid and fluid seen within the left hemithorax consistent with the known left congenital diaphragmatic hernia. There is minimal mediastinal shift. The scan was only performed due to deteriorating clinical condition. CT scans are not usually performed in our institution in babies with Congenital diaphragmatic hernias for diagnostic purposes.

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Fig. 21: An axial enhanced CT image of the chest of the same baby viewed on soft tissue windows. There is fluid filled bowel with a mesenteric vessel seen extending into the left hemithorax consistent with the known left congenital diaphragmatic hernia. The scan was only performed due to deteriorating clinical condition. CT scans are not usually performed in our institution in babies with Congenital diaphragmatic hernias for diagnostic purposes.

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Fig. 22: A coronal reformat enhanced CT image of the chest of the same baby viewed on soft tissue windows. There is fluid filled bowel with a mesenteric vessel as well as the spleen seen extending into the left hemithorax consistent with the known left congenital diaphragmatic hernia. The scan was only performed due to deteriorating clinical condition. CT scans are not usually performed in our institution in babies with Congenital diaphragmatic hernias for diagnostic purposes.

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Fig. 23: Anatomical classification of anomalies of the oesophagus and trachea, with approximate incidences. (From: Gross RE. Surgery of infancy and childhood. Philadelphia, PA: WB Saunders, 1953)

Fig. 24: A CXR of a day old baby revealing a coiled up Replogle suction catheter in the proximal oesophageal pouch of an oesophageal atresia. Gas in the stomach and proximal bowel implies the presence of a distal tracheooesophageal fistula.

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Fig. 25: A CXR of a newborn with a known oesophageal atresia and a distal tracheooesophageal fisula. The baby has a replogle suction catheter in the oesophageal pouch. The baby is noted to have a a butterfly vertebra at T5 as part of the VACTERL association.

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Fig. 26: A CXR showing a day old baby with a known oesophageal atresia and a distal tracheo-oesophageal fistula. A replege suction tube is in a proximal oesophageal pouch. Multiple vertebral anomalies including a butterfly vertebra at T9 and a left bifid 10th rib are seen as part of the VACTERL association. A fractured right clavicle is also noted.

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**Fig. 27:** A CXR of a day old baby. The baby is intubated and has a suction replogle tube in situ in the proximal oesophageal pouch. There is distal gas seen in the distended stomach and duodenum indicating a distal tracheooesophageal fistula. The child also has an associated duodenal atresia.

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Fig. 28: A pelvic ultrasound of the same baby with an oesophageal atresia and a distal tracheo-oesophageal fistula. He is noted to have a left pelvic kidney as part of the VACTERL association.

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Fig. 29: A barium swallow image in another baby revealing an H-type tracheooesophageal fistula between the oesophagus and trachea in a baby with multiple chest infections. The baby also had a duodenal atresia.

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Fig. 30: Histological classification of Congenital Cystic Adenomatoid Malformations by Stocker et al 1977. Type 1 have one or more large cysts; Type 2 have multiple small cysts of uniform size; Type 3 appear solid but have microscopic cysts.

Fig. 31: An antenatal MRI showing a coronal T2 weighted image (lying in the head down position) with a multiloculated high signal (fluid containing) lesion lying in the left lung base with the diaphragm intact. The lesion corresponded to a left lower lobe Congenital Cystic Adenomatoid malformation.

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Fig. 32: A CXR taken on day 1 of the same neonate from the above antenatal MRI scan. This shows an air filled multicystic lesion in the left lower lobe causing some mediastinal shift. A thoraco-amniotic shunt which was sited antenatally now lies within the lesion. The child had a left lower lobe Congenital Cystic Adenomatoid Malformation (CCAM) which was resected within the first year of life.

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**Fig. 33:** An axial image from an enhanced CT scan of the chest of the same neonate viewed on lung windows. The image shows multicystic lesion in the left lower lobe with air fluid levels within them. There is also a small left sided pneumothorax present. The child had a left lower lobe Congenital Cystic Adenomatoid Malformation (CCAM) which was resected within the first year of life.

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Fig. 34: An axial image from an enhanced CT scan of the chest of the same neonate viewed on soft tissue windows. The image shows fluid within the multicystic lesion in the left lower lobe. A thoraco-amniotic shunt placed in utero is identified within the lesion. No systemic feeding vessel was identified. The child had a left lower lobe Congenital Cystic Adenomatoid Malformation (CCAM) which was resected within the first year of life.

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**Fig. 35:** A coronal image from an enhanced CT scan of the chest of the same neonate viewed on soft tissue windows. The image shows fluid within the multicystic lesion in the left lower lobe. No systemic feeding vessel was identified. The child had a left lower lobe Congenital Cystic Adenomatoid Malformation (CCAM) which was resected within the first year of life.

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Fig. 36: A CXR from a newborn showing increased density in the left lower lobe. There had been an antenatal history of a congenital lung anomaly. The lesion later correlated with a lower lobe sequestration which was resected within the first year of life.

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**Fig. 37:** An axial image from an enhanced CT scan of the same baby above viewed on lung windows. There is a solid left lower lobe enhancing mass with large feeding vessels seen within the mass. The lesion corresponded to a sequestration which was resected in the first year of life.

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Fig. 38: An axial image from an enhanced CT scan of the same baby viewed on soft tissue windows. There is a solid left lower lobe enhancing mass with a large feeding vessel (red Arrow) seen extending from the aorta to the mass. The lesion corresponded to a sequestration which was resected in the first year of life.

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Fig. 39: A coronal image from an enhanced CT scan of the same baby above viewed on soft tissue windows. There is a solid left lower lobe enhancing mass with two large feeding vessels seen extending from the aorta to the mass. The lesion corresponded to a sequestration which was surgically removed within the first year of life.

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Fig. 40: A CXR from a newborn with an antenatal diagnosis of a left lung anomaly. There is a lucent cystic lesion in the left lower lobe causing mediastinal shift. The baby was tachypnoeic. The lesion was diagnosed as a sequestration on subsequent imaging. The sequestration was surgically removed in the first year of life.

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Fig. 41: An axial image from an enhanced CT scan of the chest of the same baby viewed on lung windows. It reveals an air filled multicystic mass in the left lower lobe with no real solid component. The mass was found to be a sequestration which was surgically removed within the first year of life.

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**Fig. 42:** An axial image from an enhanced CT scan of the chest of the same baby viewed on soft tissue windows reveals a large systemic vessel extending from the aorta towards the lesion. The mass was found to be a sequestration which was surgically removed in the first year of life.

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**Fig. 43:** A coronal image from an enhanced CT scan of the chest of the same baby viewed on lung windows. It reveals an area of hyperlucency with cystic air spaces in the left lower lobe with a large systemic vessel extending from the aorta towards the lesion. The mass was found to be a sequestration which was surgically removed in the first year of life.

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**Fig. 44:** An axial image from an enhanced CT scan of the chest of a newborn performed for an antenatally diagnosed lung lesion, viewed on lung windows. The baby was assymtomatic. The CT scan shows a tubular non enhancing lesion in the apicoposterior segment of the left upper lobe with associated hyperlucent lung and air trapping distally. The appearances are consistent with a bronchial atresia.

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Fig. 45: An axial image from the enhanced CT scan of the chest on the same baby viewed on soft tissue windows. The CT scan shows a tubular non enhancing lesion in the apicoposterior segment of the left upper lobe. The appearances are consistent with a bronchial atresia.

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**Fig. 46:** A coronal image from the enhanced CT scan of the chest of the same baby viewed on lung windows. The CT scan shows a tubular non enhancing lesion in the apicoposterior segment of the left upper lobe with associated hyperlucent lung and air trapping distally. The appearances are consistent with a bronchial atresia.

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Conclusions

We have attempted to provide a broad overview of the congenital abnormalities that we see in the neonatal thorax at our institution. We have included Xrays, MRI scans and CT scans where appropriate to illustrate key diagnostic findings for these conditions. Where helpful we have included hisological and anatomical classifications and also briefly covered management options in some cases.

We feel that this pictorial review will serve to increase confidence in recognising and reporting of these abnormalities leading to more rapid and appropriate management of this vulnerable group of patients.