

INTRODUCTION

Congenital lung anomalies vary widely in their clinical manifestation and imaging appearance, multidetector computer tomography (CT) is frequently required for confirmation of diagnosis, further characterization, and preoperative evaluation in the case of surgical lesions. Recently, with the development and widespread availability of multidetector CT scanners, CT has assumed a greater role in the noninvasive evaluation of congenital lung anomalies. The combination of fast speed, high spatial resolution, and enhanced quality of multiplanar reformation and three-dimensional reconstructions makes multidetector CT an ideal noninvasive method for evaluating congenital lung anomalies (*Lee et al., 2008*).

Multidetector CT has the ability to produce high-quality thin-section CT images, recently developed postprocessing methods, including Multiplaner and 3D volume rendering, are now routinely used to evaluate congenital lung anomalies (*Siegel and Lee, 2007*).

Congenital lung anomalies can be divided into nonvascular (parenchymal) lesions, vascular lesions, and a combination of both (*Zylak et al., 2002*).

The spectrum of congenital bronchopulmonary malformation includes congenital cystic adenomatoid malformation, congenital

lobar emphysema and bronchogenic cyst, they have unique manifestation often mimic thoracic pathology and can present acutely and necessitate emergent evaluation and management (*Shanmugam et al., 2005*).

CT angiography is particularly useful for evaluation of the vascular component of congenital lung anomalies (*Ohtsuki et al., 2005*).

Vascular lesions includes anomalies of pulmonary artery, anomalies of pulmonary vein, combined anomalies of pulmonary artery and vein pulmonary arteriovenous fistula, the detection, characterization, and follow-up of pulmonary AVMs are best accomplished by using CT specifically multidetector CT, owing to its ability to accurately display complex AVM angioarchitecture. CTA angiography protocol, when combined with 3D reconstruction, is particularly useful in depicting the angioarchitecture of the feeding arteries and draining veins and in estimating the size of the pulmonary AVM (*Daltro et al., 2004*).

Combined abnormal parenchymal and abnormal vasculature includes Hypogenetic lung syndrome, also known as congenital pulmonary venolobar syndrome or scimitar syndrome. CT angiography with 3D reconstruction is particularly helpful in evaluating the entire course of the anomalous scimitar vein and its eventual drainage site, which is most commonly the inferior vena cava (*Berrocal et al., 2003*).

AIM OF THE WORK

The aim of work is to review the role of multidetector CT in evaluation of congenital lung anomalies.

Chapter (1)

ANATOMY OF THE LUNG AND PULMONARY VESSELS

THE LUNGS

The lungs are described as having costal, mediastinal, apical and diaphragmatic surfaces. The right lung has three lobes and the left has two, with the lingula of the left upper lobe corresponding to the right middle lobe.

One terminal bronchiole with lung tissue forms an acinus which, together with vessels, lymphatics and nerves, forms the primary lobule. Three to five primary lobules form a secondary lobule (*Ryan et al., 2011*)

The subsegmental structures of the lung and the secondary pulmonary lobule Three units of lung structure have been described at the subsegmental level of the lung

- a) The primary pulmonary lobule.*
- b) The acinus*
- c) The secondary pulmonary lobule*

a) Primary Pulmonary Lobule

In **1947 Miller originally** described the primary pulmonary lobule and defined it as the lung unit distal to the respiratory bronchioles (*Kim et al., 2002*).

The primary pulmonary lobule consists of alveolar ducts, alveolar sacs and alveoli. Approximately 30–50 primary pulmonary lobules can be found in one secondary pulmonary lobule (*Matsuoka et al., 2005*).

It cannot be demonstrated by CT in either normal or abnormal states, this unit is of no practical radiological significance (*Verschakelen and De Wever, 2007*).

b) Acinus

The acinus is considered as the anatomic and functional unit of the lung. Its anatomic definition describes the acinus as the portion of lung distal to a terminal bronchiole and supplied by a first-order respiratory bronchiole or bronchioles so, it is the the largest unit in which all airways participate in gas exchange (*Webb, 2006*).

c) Secondary Pulmonary Lobule

A good understanding of the lung anatomy in general and of the anatomy of the secondary pulmonary lobule in particular is extremely useful in understanding the pathology and pathogenesis of most pulmonary disease states (*Webb, 2006*).

The secondary pulmonary lobule is defined as the smallest unit of lung structure margined by connective tissue septa (*Verschakelen and De Wever, 2007*).

It is supplied by a group of terminal bronchioles, is irregularly polyhedral in shape and is approximately 1-2.5 cm on each side (*Devakonda et al., 2010*).

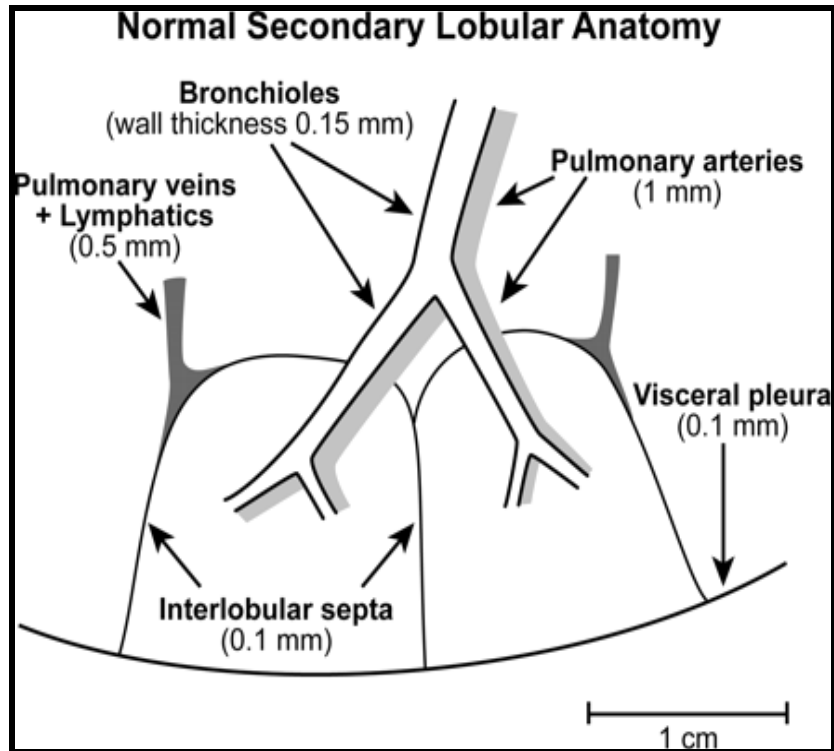


Figure (1): Secondary pulmonary lobule, as shown by (*Kim et al., 2002*). Diagram shows secondary lobular anatomy. Secondary lobules represent fundamental anatomic units of the lung and are defined by centrilobular structures, including pulmonary arteries/arterioles and their accompanying bronchi/bronchioles, and peripheral structures, including the pulmonary veins and lymphatics within the interlobular septae (*Devakonda et al., 2010*).

The secondary pulmonary lobules are demarcated from each other by interlobular connective tissue septa. From these interlobular septa, fibrous strands penetrate into the lobule between the acini (*Kim et al., 2002*).

The interlobular septa are also continuous peripherally with the pleura (**Webb, 2006**). The septa in the upper lobes tend to be longer and more randomly arranged, where as in the lower lung fields they appear to be shorter and more horizontally oriented perpendicular to the pleural surfaces (**Devakonda et al., 2010**).

The lobular bronchiole is distributed with the accompanying artery, which has the same irregular dichotomous branching into the central portion of the lobule (**Verschakelen and De Wever, 2007**).

Bronchopulmonary segments

Each lobe is subdivided into bronchpulmonariy segments, each of which is supplied by a segmental bronchus artery and vein. Each segment takes its title from that of its supplying bronchus (**Ryan et al., 2011**).

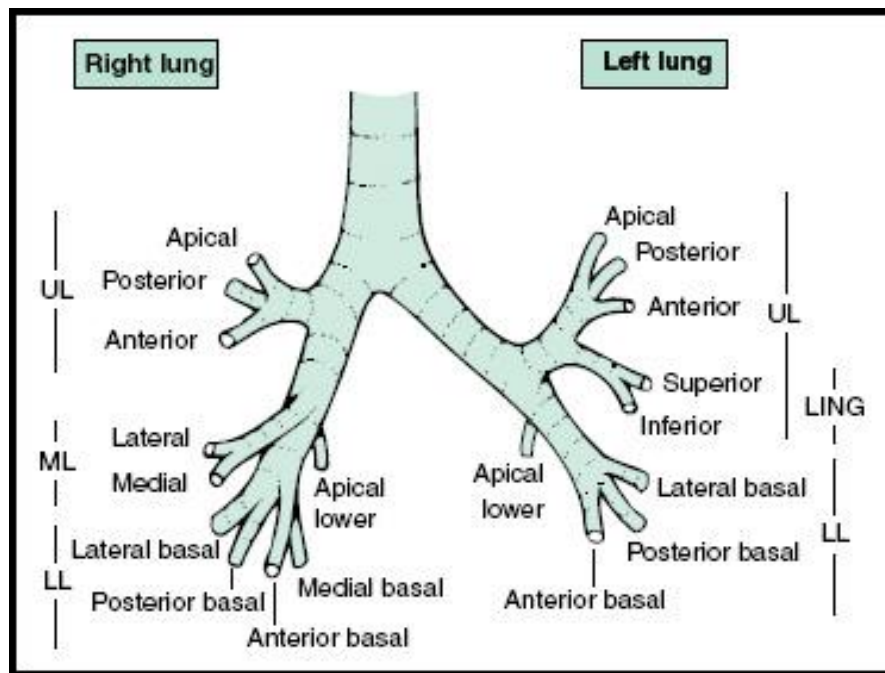


Figure (2): Bronchopulmonary segments. LING, lingula; LL, lower lobe; ML, middle lobe; UL, upper lobe (*Bourke and Burns, 2011*)

Anatomy of small airways

Airways divide by dichotomous branching with approximately 23 generations of branches identifiable from the trachea to the alveoli; finally the terminal bronchioles are reached (*Matsuoka et al., 2005*).

The bronchioles include two categories: *the membranous bronchioles* (lobular and terminal) and *the respiratory bronchioles*. The term “small airways” is often also used to describe these bronchioles and small airway disease is then defined as the pathological condition in which these bronchioles are affected (*Verschakelen and De Wever, 2007*).

The **lobular bronchioles** enter the core of the secondary pulmonary lobule and divide into a number of **terminal bronchioles** according to the size of the lobule. These terminal bronchioles represent the most distal purely conducting portion of the tracheo-bronchial tree; that is they conduct air without being involved in gas exchange. The terminal bronchioles give rise to the **respiratory bronchioles, which** are so designated because alveoli bud directly from their walls and are also involved in gas exchange. The respiratory bronchioles give rise to alveolar ducts. The alveolar ducts finally lead into the alveolar sacs containing several alveoli (*Webb, 2006*).

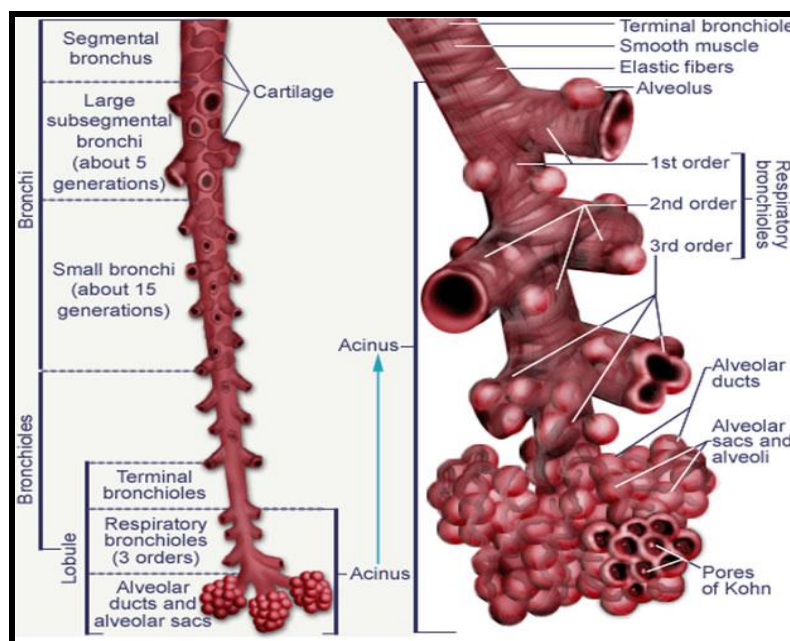


Figure (3): Anatomic organization of the tracheo-bronchial tree; the lobular bronchioles enter the core of the secondary pulmonary lobule and divide into a number of terminal bronchioles according to the size of the lobule (*Devakonda et al., 2010*).

The pulmonary Artery

The **pulmonary trunk** leaves the fibrous pericardium and bifurcates almost at once in the concavity of the aortic arch anterior to the left main bronchus.

The **right pulmonary artery** is longer than the left. It passes across the Mid`line below The carina and comes to lie anterior to the right main bronchus It bifurcates while still in the hilum of the right lung. An artery for the right upper lobe passes anterior to the right upper lobe bronchus The **interlobar arter** to the right middle and lower lobes passes with bronchus intermedius (*Ryan et al., 2011*).

The **left pulmonary artery** spirals over the superior aspect of the left main bronchus to reach its posterior surface. It is attached to the concavity of the aortic arch by the ligamentum arteriosum.

The pulmonary veins

These do not follow the bronchial pattern but tend to run in intersegmental septa. Two veins pass to each hilum – from lung tissue above and below each oblique fissure .These enter the mediastinum slightly below and anterior to the pulmonary arteries. On the right side the veins from the lobes may remain separate, so that three veins leave the right lung and enter the left atrium. On the left side the two pulmonary veins may unite and enter the left atrium as a single vessel (*Ryan et al., 2011*).

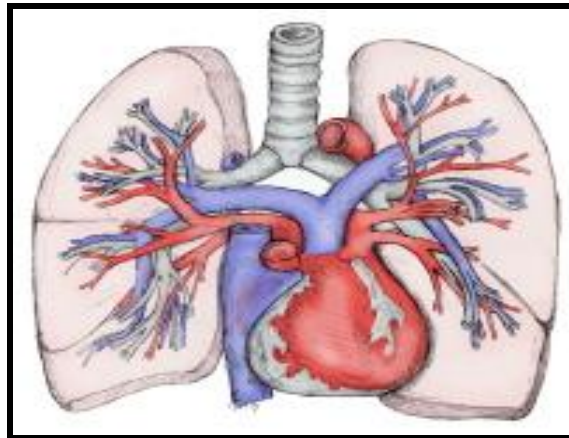


Figure (4): Pulmonary artery and vein in relation to airways and lungs (*Mendoza et al., 2013*).

Anatomic organization of the lymphatics

There are two intercommunicating networks of lymph flow. First there is a rich subpleural plexus, which is connected to and drained by the septal lymphatic channels. These channels follow interlobular septa and progress into axial connective tissue sheaths around veins as they progress centrally (*Webb, 2006*). Another system of lymphatic channels (peri broncho vascular) is found in the axial connective tissue around arteries, bronchi and bronchioles (*Webb, 2006*).

There are no lymphatics found in alveolar walls (*Verschakelen and De Wever, 2007*). Lymphatic fluid that is escaping from the capillaries has to migrate towards the pulmonary lymphatics, which are located in the peribronchiolar and the perivascular spaces, the inter lobular septa and the pleural network; A disease process following perilymphatic

distribution would be understood by recognition of the lymphatic distribution throughout lungs (fig. 1) (*Devakonda et al., 2010*).

Multidetector Computed Tomography (MDCT) Features of the normal lung

Secondary Pulmonary Lobule

Although the identification of secondary pulmonary lobules in normal patients may be difficult with CT, some features that help to identify this anatomical structure are often present. A few septa can be visible in the lung periphery in normal subjects, mostly anteriorly and along the mediastinal pleural surfaces (*Webb, 2006*).

The location of the interlobular septa can also often be inferred by locating septal pulmonary vein branches. They present as linear, arcuate or branching structures about 5-10 mm from the centrilobular arteriole (*Stern et al., 2009*).

This centrilobular arteriole presents as a dot-like, linear or branching opacity within the centre of the lobule or for lobules abutting the pleura at about 1cm from the pleural surface (*Verschakelen and De Wever, 2007*).

Some smaller intralobular vascular branches may be visible between the septa and the centrilobular arteriole, again

presenting as small dots or branching lines, but this time, about 3-5mm from the septa (**Webb, 2006**).

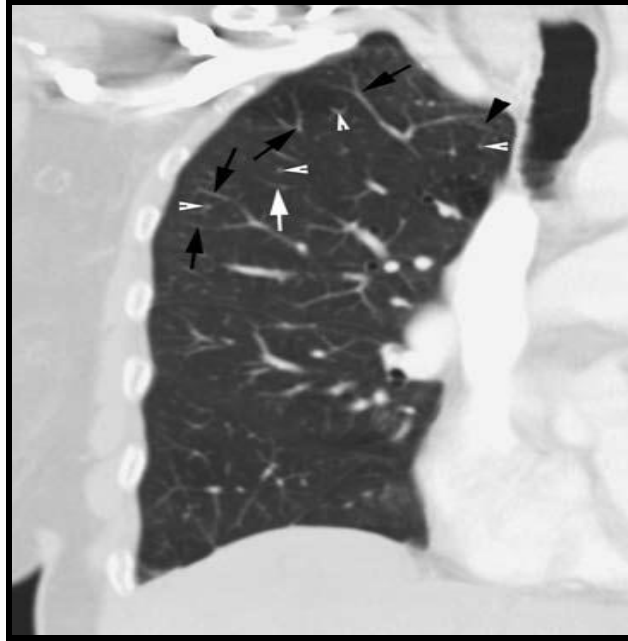


Figure (5): CT of the normal lung (coronal reconstruction). Interlobular septa can sometimes be recognised anteriorly and along the mediastinal surfaces (*arrowhead*) but can more often be inferred by locating septal pulmonary vein branches (*arrows*) presenting as linear, arcuate or branching structures approximately 5-10 mm from the centrilobar arteriole (*white arrowheads*) (**Webb, 2006**).

Lung Parenchyma

The density of the lung parenchyma should be of greater density than tracheal air. This density is determined by three components: *lung tissue*, *blood* in small vessels beyond the resolution of CT and *air* (**Stern et al., 2000**).

Lung density decreases when lung volume is increased (*Sundaram et al., 2008*).

Fissures

On conventional CT fissures are less visible than on plain radiographs.

They are seen as regions of relative avascularity on the outer cortex of the lobe, where tapering vessels are less visible. Discrete lines are only seen if the vertical axis of the fissure is perpendicular to plane of the CT slice, which sometimes occurs in parts of the oblique fissure but not in the transverse fissure. On high-resolution CT fissures are seen as sharp lines (*Ryan et al., 2011*).

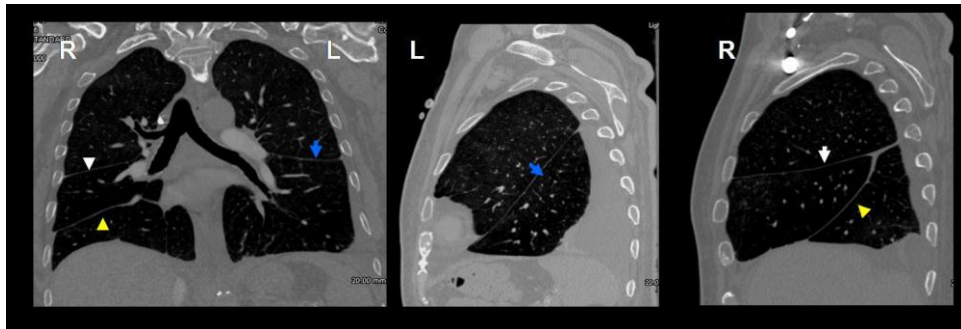


Figure (6): The coronal CT image on the left shows the right minor (horizontal) fissure (white arrow), right major fissure (yellow arrow), and left major (oblique) fissure. These structures are also seen on the right lateral sagittal CT image on the right. The minor fissure separates the right superior lobe from the right middle lobe. The right major fissure separates the right upper and middle lobes from the inferior lobe. The left major fissure (middle CT image) separates the left upper and lower lobes (*Joseph, 2010*).

On CT the minor fissure position is represented by an oval area of reduced vascularity at the level of the bronchus intermedius. The normal minor fissure is not seen as a line on axial CT imaging but is apparent on multiplanar reformats (*Adam and Dixon, 2008*).

TRACHEA:

The trachea extending from the lower part of the larynx, on a level with the sixth cervical vertebra, to the upper border of the fifth thoracic vertebra, where it divides into the two bronchi, one for each lung. The trachea is nearly but not quite cylindrical, in adult it measures about **11 cm.** in length. Its diameter in normal human adult male; from side to side; is from 1.3 to 2.5 cm in the coronal plane, in normal human adult female; from side to side; is from 1 to 2.1 cm in the coronal plane, being always greater in the male than in the female. Tracheal length is dynamic, changing with respiration and neck flexion and extension. For example, the carina can change in position up to 3cm between inspiration and expiration (*Stern et al., 1993*).

The intrathoracic length of the trachea is 6-9 cm. The trachea scaffolding is comprised of C-shaped cartilage rings completed by the pars-membranacea posteriorly. In the trachea and mainstem bronchi; the cartilage is present as "C" shaped rings in their outer wall open at the back. The carina resides approximately at the level of the fifth thoracic vertebral body,