Evaluation of the Lungs and Airways

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The following points should be considered in evaluating the lungs and airways:

1. Congenital airway anomalies are more common in patients with congenital heart disease. Congenital stenosis and bilateral left- or right-sidedness is common in patients with asplenia or polysplenia-type heterotaxies.
2. Specifically, look for a right upper lobe (pig) bronchus, especially in patients with chronic right upper lobe collapse.
3. Tracheobronchomalacia is common in congenital heart disease patients. A narrowed horseshoe-like appearance of the airway may be seen on CT.
4. Extrinsic airway compression is very common in patients with complex congenital heart disease. Look for vascular rings, pulmonary sling, and dilated structures compressing the airway.

Normal Anatomy

Normally, the right mainstem bronchus courses posterior and slightly superior to the right main pulmonary artery (eparterial bronchus), with the right upper lobe bronchus arising more proximal than the left. The left mainstem bronchus courses under the left main pulmonary artery (hyparterial bronchus).

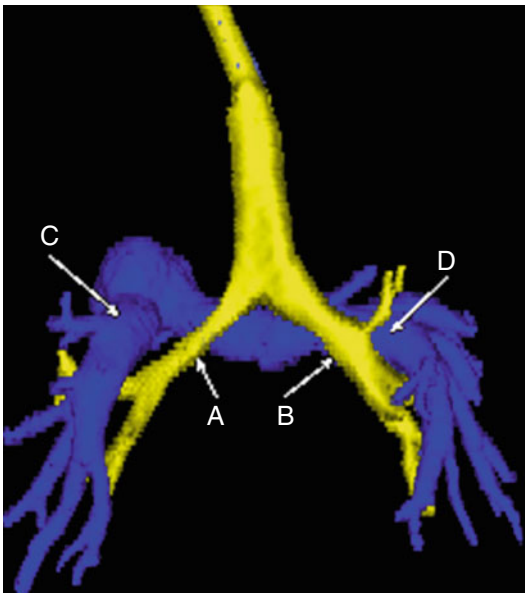

Fig. 9.1 Posterior projection from a cardiac CTA image showing a typical left bronchus (*A*), which is elongated and is known as a hyparterial bronchus because the left main pulmonary artery (*C*) is draped over the proximal left bronchus (*A*). The right main bronchus (*B*) has a shorter course before giving off the upper lobe bronchus more proximally. Notice that the left main pulmonary artery (*D*) is not draped over the bronchus (*B*). This is known as an *eparterial bronchus*

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Tracheal (Pig) Bronchus

A tracheal, or pig, bronchus is an anatomic variant (typically right) upper lobe bronchus originating from the trachea above the carina. Two types exist: displaced bronchus and supernumerary bronchus. This condition may cause persistent or recurrent right upper lobe pneumonias.

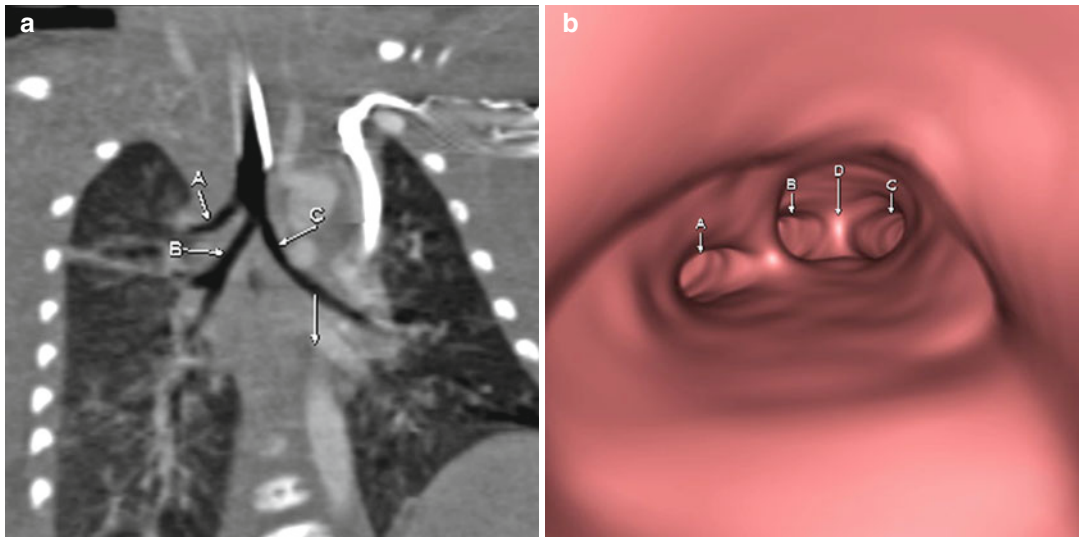

Fig. 9.2 Coronal (**a**) and virtual bronchoscopy. Descending aorta (*arrow*) is shown. (**b**) Images demonstrating a right upper lobe accessory bronchus (*A*) arising from the trachea above the carina (*D*). The right bronchus (*B*) and left main bronchus (*C*) are shown

Airway Compression from Enlarged Pulmonary Arteries

Enlarged pulmonary arteries, when present, may be a frequent cause of airway compression in infants and small children with congenital heart disease. Enlarged pulmonary arteries frequently are seen in patients with congenital absence of the pulmonic valve, as observed in tetralogy of Fallot variants.

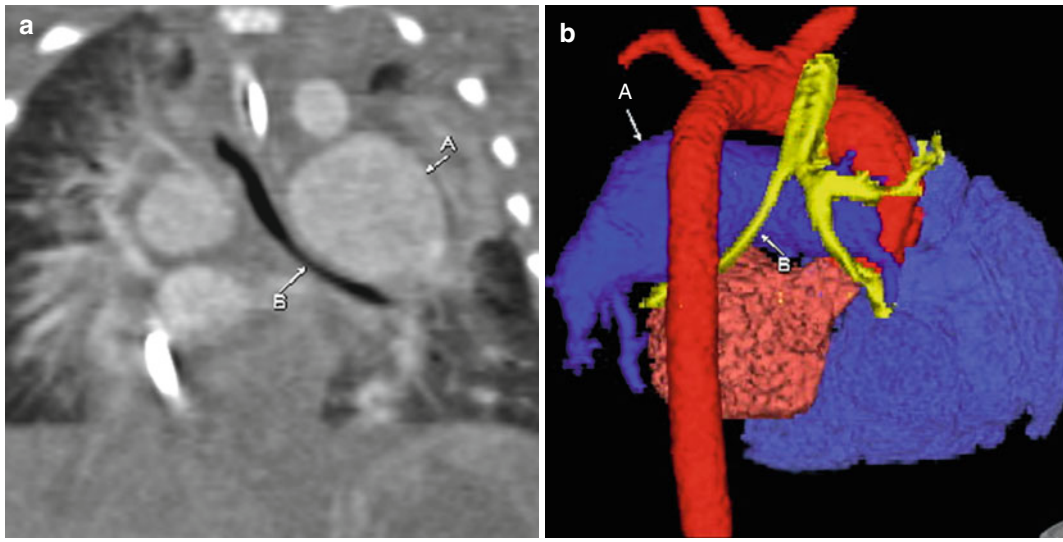

Fig. 9.3 Enhanced coronal (**a**) and posterior projection color-coded three-dimensional (3D; **b**) images showing an enlarged left main pulmonary artery (*A*) with mass effect, resulting in moderate narrowing of the adjacent left main bronchus (*B*)

Tracheobronchomalacia

Tracheobronchomalacia is a tracheobronchial cartilaginous abnormality resulting in abnormally increased pliability and intermittent airway collapse. Dynamic examination, such as fluoroscopy or endoscopy, shows characteristic changes in airway caliber. Tracheobronchomalacia may be primary or secondary.

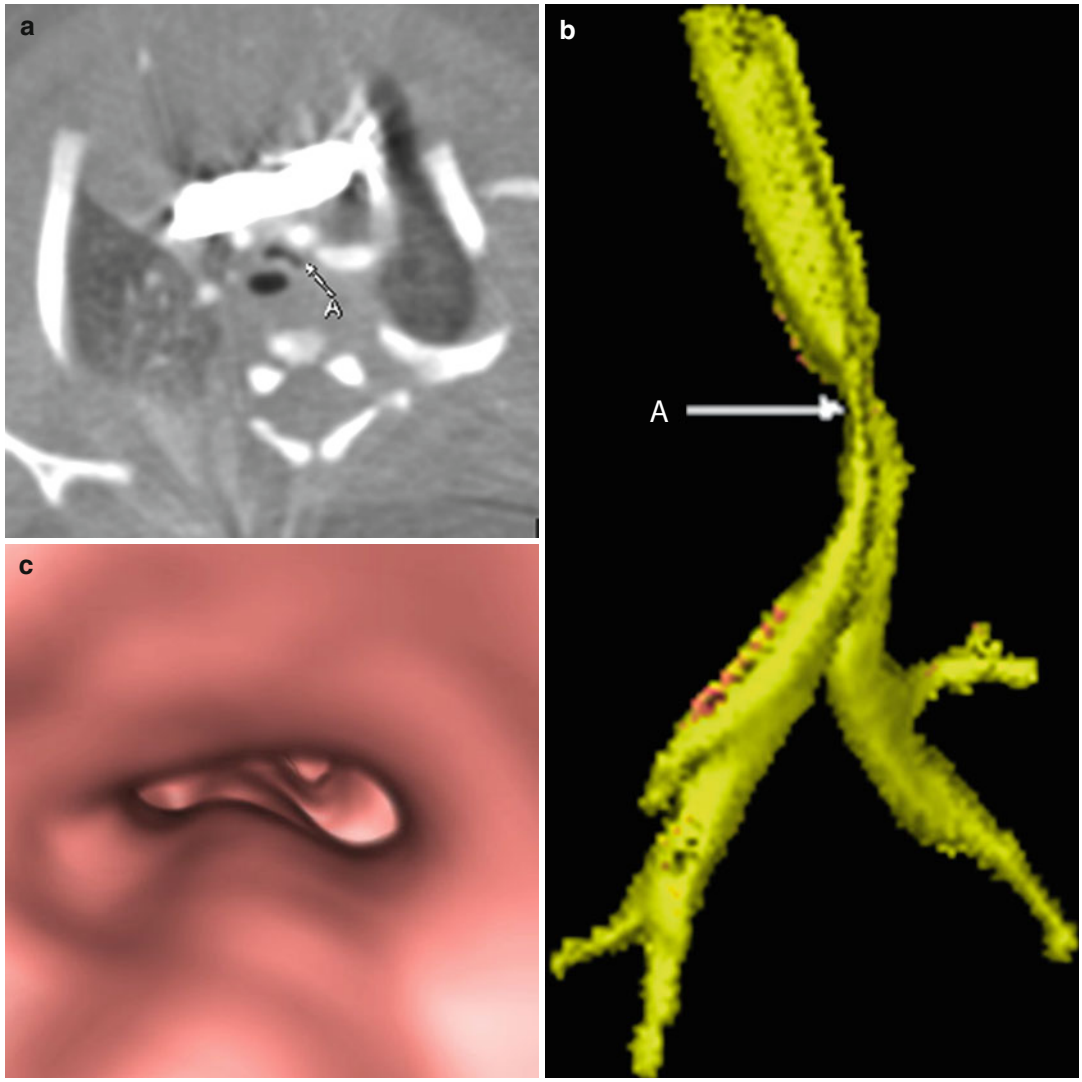

Fig. 9.4 Enhanced axial (**a**), lateral projection color-coded 3D (**b**), and virtual bronchoscopy (**c**) images show an area of mid-tracheal narrowing (*A*) consistent with tracheomalacia. Notice the characteristic horseshoe shape of the trachea (*A*) which is a typical CT finding in patients with tracheomalacia

Double Aortic Arch

Double aortic arch is the congenital persistence of both left and right fourth embryologic arches. Two arches surround and compress the trachea anteriorly and esophagus posteriorly. Surgical management is altered depending on whether the right or left arch is dominant; the right arch more commonly is dominant. Double aortic arch is the most common symptomatic vascular anomaly. It forms a true vascular ring.

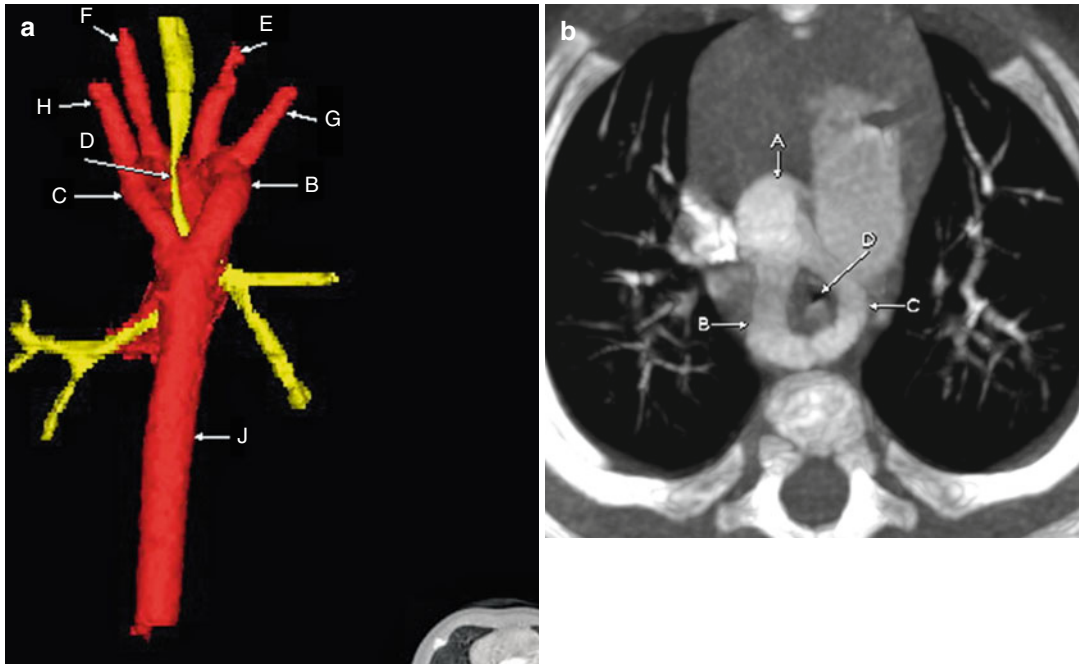

Fig. 9.5 Posterior projection color-coded 3D (**a**) and enhanced axial CT (**b**) images showing tracheal compression (*D*) by the surrounding vascular ring formed by the right (*B*) and left (*C*) aortic arches, which divide from the ascending aorta (*A*) and join again posteriorly to form the descending aorta (*J*). There is symmetric branching of the great vessels, with the right (*E*) and left (*F*) carotid and right (*G*) and left (*H*) subclavian arteries arising from the corresponding aortic arch.

Pulmonary Sling

Pulmonary sling is an anomaly in which the left pulmonary artery originates from the right pulmonary artery and then courses around the trachea and proximal mainstem bronchi, often resulting in severe airway compression. The anomalous left pulmonary artery courses between the trachea and esophagus as it crosses from the right to left hemithorax unlike the aberrant right and left subclavian arteries, which typically course posterior to the esophagus. Airway compression often is asymmetric and may result in unilateral hyperinflation from air trapping. On a lateral esophagram, indentation of the posterior trachea and anterior esophagus is characteristic. Pulmonary sling is not a true vascular ring.

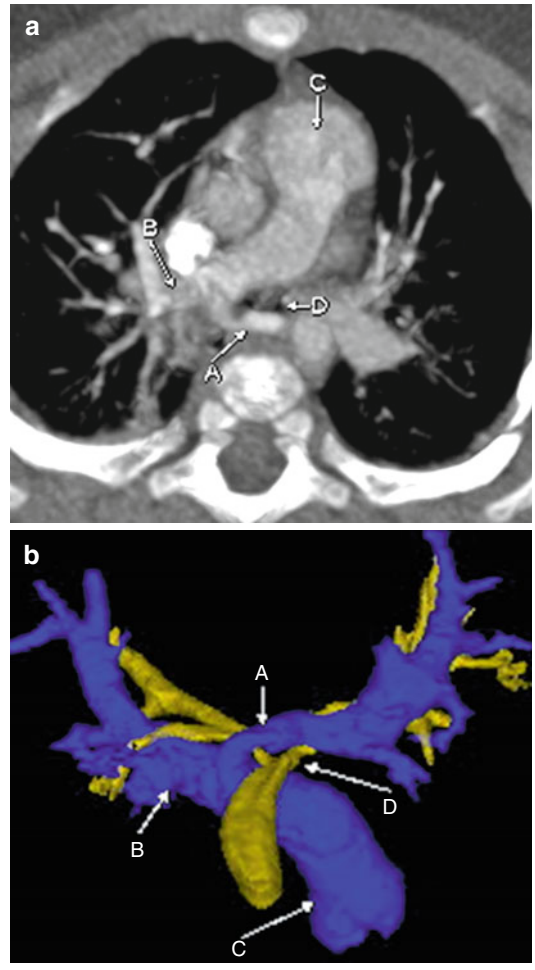

Fig. 9.6 Axial CT (**a**) with anterior (**b**) and superior (**c**) projection 3D color-coded images from a cardiac CTA scan showing severe airway compression (*D*) in a patient with pulmonary sling. Notice that the left pulmonary artery (*A*) arises from the right pulmonary artery (*B*) and then slings around the trachea to the left. The main pulmonary artery (*C*) is in the normal position

Right Aortic Arch with Aberrant Left Subclavian Artery

Right aortic arch with an aberrant left subclavian artery (RAA-ALSCA) is a true vascular ring when the patent ductus arteriosus (PDA) arises from the aberrant right subclavian artery. In this scenario, the right arch (right), aberrant subclavian (posterior), pulmonary artery (anterior), and PDA (left) form a complete vascular ring around the trachea and esophagus. Tracheal narrowing and stridor are common, but the patient may become increasingly symptomatic with time.

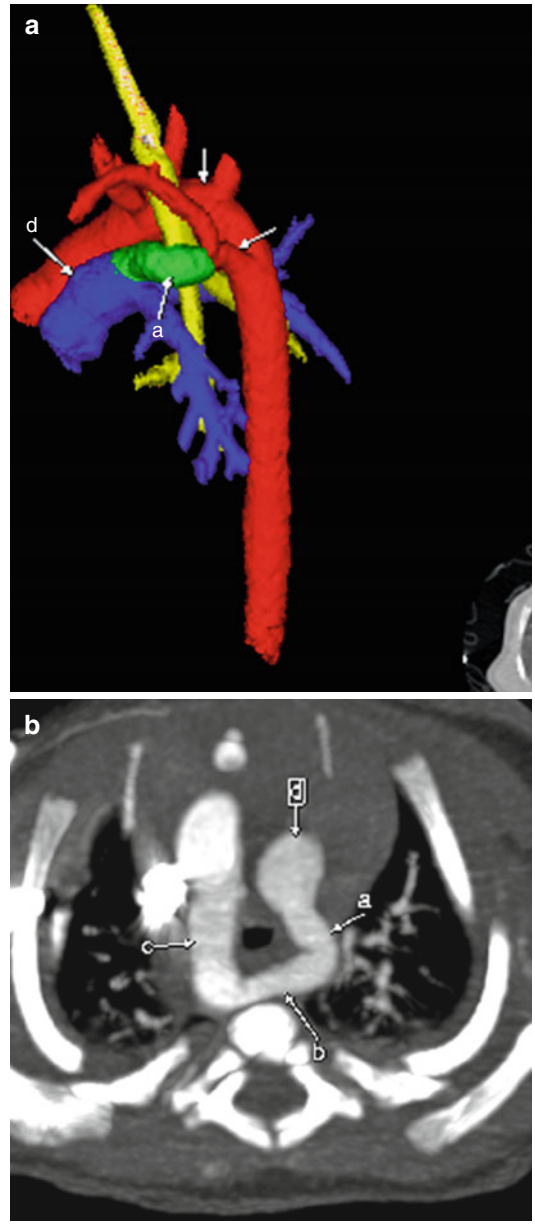

Fig. 9.7 Color-coded 3D (**a**) and axial CT (**b**) images from a cardiac CTA scan showing a right aortic arch (*c*) with an aberrant left subclavian artery (*b*). The PDA (*a*) arising from the main pulmonary artery (*d*) completes the vascular ring around the trachea (*yellow*) and esophagus

Right Aortic Arch with Aberrant Left Subclavian Artery with Kommerell Diverticulum

RAA-ALSCA often occurs with a Kommerell diverticulum, which is seen as a focal dilatation at the origin of the aberrant subclavian artery off the descending right aortic arch. A Kommerell diverticulum may be present in patients with a left aortic arch and an aberrant right subclavian artery but is much less common.

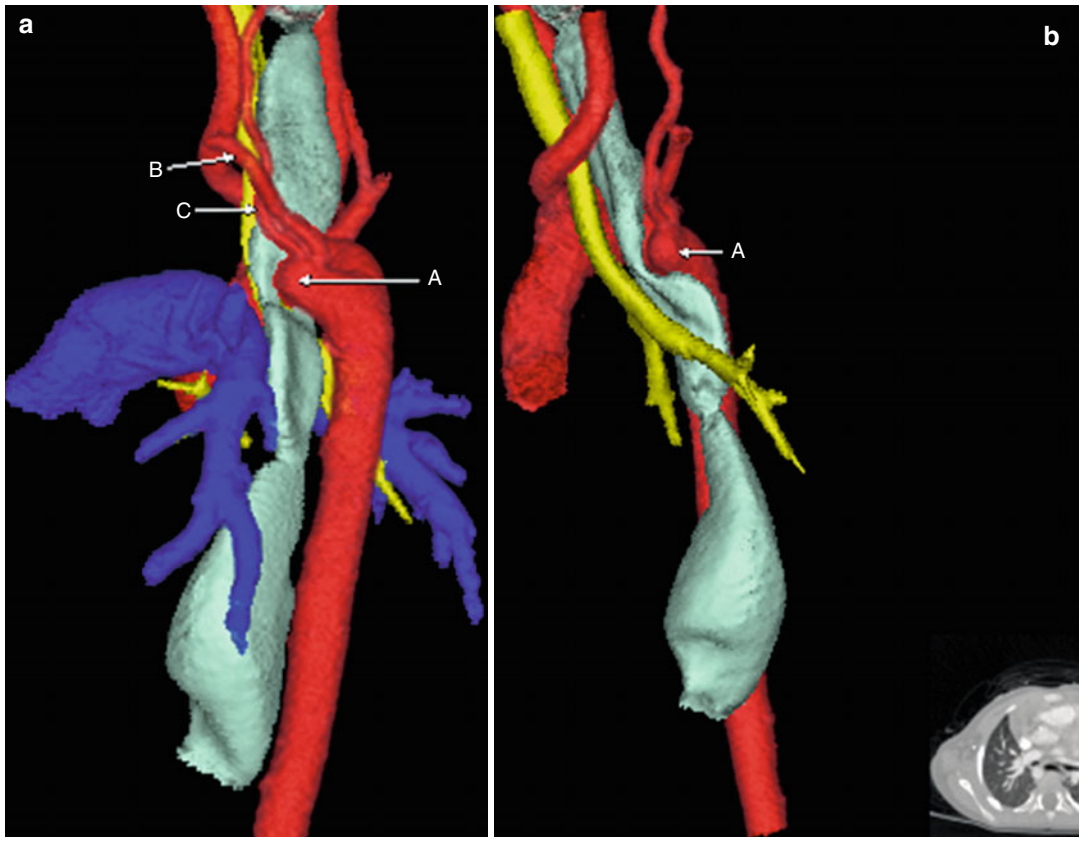

Fig. 9.8 (**a**, **b**) Color-coded 3D images from a cardiac CTA scan showing mild airway compression in a patient with a right arch and aberrant left subclavian (*B*) and vertebral (*C*) arteries. Notice the compression of the esophagus (*green*) and the contouring of the trachea (*yellow*) from the large diverticulum of Kommerell (*A*)