



# Left Pulmonary Artery Sling—Anatomy and Imaging

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Distinctive imaging findings are present when the left pulmonary artery (LPA) arises from the right pulmonary artery and forms a sling around the airway passing between airway and esophagus to reach the left lung. It is important to recognize the 2 distinct types of pulmonary sling. The less complex type I is associated with tracheobronchomalacia and is often managed successfully by LPA reimplantation. The more common and more complex type II is strongly associated with long segment tracheal stenosis. Appropriate management needs to address the airway abnormality in addition to the aberrant LPA. Both types, especially type II, are associated with cardiovascular, pulmonary, and other abnormalities that also need to be considered in therapeutic decisions. The role of imaging is to identify, define, and display the relevant anatomic relationships to facilitate appropriate management. Recent surgical advances have increased the likelihood of success of surgical correction of long segment airway stenosis. *Semin Ultrasound CT MRI 31:158-170 © 2010 Elsevier Inc. All rights reserved.*

Distal tracheal obstruction by an aberrant left pulmonary artery (LPA) was first described in 1897.<sup>1</sup> In this congenital anomaly the LPA originates from the posterior aspect of the right pulmonary artery (RPA) and courses over the right main bronchus and then posteriorly between the trachea and esophagus to reach the left lung, thereby forming a partial sling around the trachea (Figs. 1-10).<sup>1,2</sup> A patent ductus arteriosus (PDA) or ductal ligament may contribute to encirclement of the trachea.<sup>3</sup>

Descriptions of left pulmonary artery sling (LPAS) are typically somewhat of an afterthought, tacked on to the end of a discussion of vascular rings. Although somewhat uncommon, LPAS can stand on its own as a distinct and important abnormality. Prompt recognition, accompanied by an understanding of the spectrum of the anomalies seen, greatly assists appropriate management decisions.

It is theorized that LPAS occurs when the left postbranchial pulmonary arterial vessels cannot connect with the left sixth branchial arch (which usually forms the proximal LPA), and a secondary connection is acquired to the right sixth branchial arch through the embryonic peritracheal primitive mesenchymal vessels.<sup>2,4</sup> This connection is usually posterior, forming a pulmonary sling, and less commonly anterior (pseudo pulmonary sling, also known as criss-crossed pulmonary arteries).<sup>5-7</sup> A genetic influence on the LPAS anomaly is apparent by its occurrence in identical twins and trisomies 18 and 21.<sup>8,9</sup>

Approximately two-thirds of LPAS cases are associated with intrinsic tracheal stenosis with complete cartilaginous rings, often involving a long segment of the trachea (Figs. 4-10).<sup>2,3,10,11</sup> To highlight this association, Berdon<sup>1</sup> coined the term ring sling complex.

Pulmonary slings frequently present with respiratory symptoms in young infancy, the timing and severity of symptoms depends on the degree of accompanying airway abnormality and may be precipitated by an acute respiratory infection.<sup>2,12</sup> The presentation may be catastrophic and untreated pulmonary sling carries a high morbidity and mortality, most of which is due to the airway and other associated anomalies rather than the aberrant artery itself.<sup>13</sup> Feeding difficulty without overt respiratory symptoms may be another presenting symptom. Pulmonary sling can occasionally be found in a less symptomatic or even asymptomatic older child or adult (Figs. 2 and 6).<sup>12,14,15</sup> In some patients, symptoms from associated abnormalities may dominate the clinical presentation.<sup>2</sup>

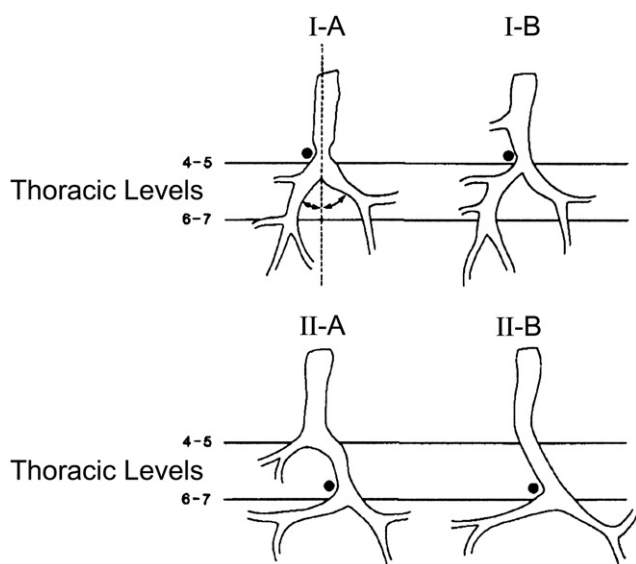
LPAS along with vascular ring is a diagnosis often missed on chest radiographs. The purpose of this review is to provide an overview of the imaging findings in this entity and emphasize the role of imaging in defining the anatomy and important associated anomalies.

## Classification, Anatomic Variations, and Anomalies Associated With LPAS

LPAS has been divided into 2 types (Fig. 1).<sup>6</sup> In type I, the LPAS is located just above the carina (T4-T5) abutting the distal trachea and proximal right main stem bronchus and typically compressing the adjacent airways, resulting in distal

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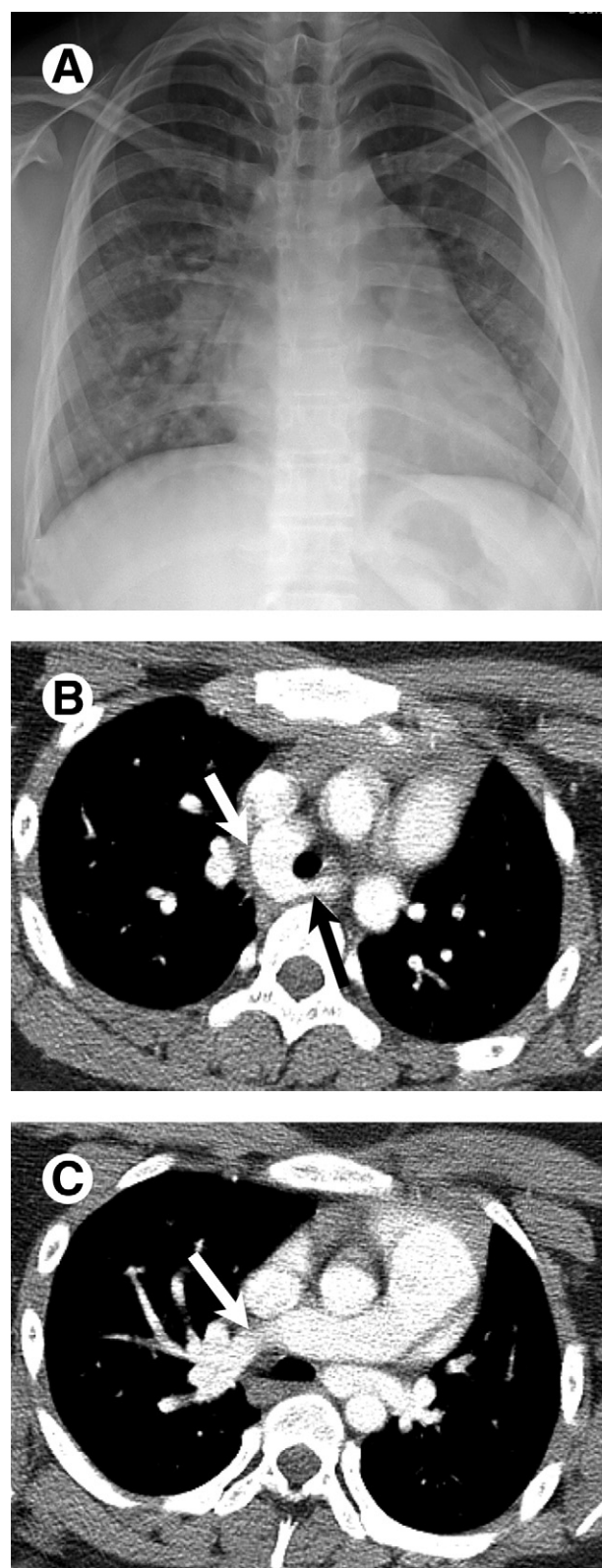
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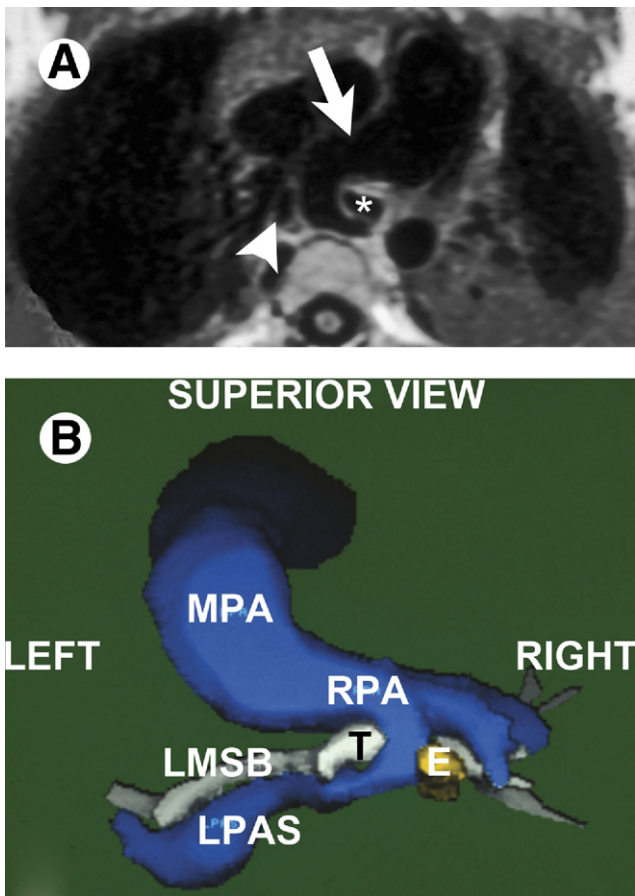
**Figure 1** Classification of anatomic types of pulmonary sling—solid circle denotes left pulmonary artery (LPA) origin. Type I—PA sling at T4-5 level just above carina. IA without and I B with right tracheal bronchus. Type II—PA sling at T6-7 level just above low horizontal carina. IIA right upper lobe “tracheal” bronchus at usual site of carina, distal airway stenosis, bridging right bronchus. II B—Same as A except no separate right upper lobe bronchus. Reprinted with permission from Wells TR, Gwinn JL, Landing BH, et al. *J Pediatr Surg* 23:892-898, 1988.<sup>6</sup>

tracheal and right-sided bronchomalacia that may cause air trapping and hyperinflation of the right lung.<sup>6,14,16</sup> The air trapping may be marked enough to resemble congenital lobar hyperinflation both pre- and postnatally.<sup>17,18</sup> A tracheal bronchus may be present but the trachea is usually otherwise normal, although there are occasional reports of tracheal stenosis associated with the type I LPAS.<sup>14</sup> Type I pulmonary sling has been divided into 2 subtypes depending on the absence (type IA) (Figs. 2 and 3) or presence (type IB) of a tracheal bronchus.<sup>6</sup>

The type II LPAS is more common, especially type IIB (Fig. 1).<sup>6,14</sup> The sling is more inferiorly located (T5-6) in the chest adjacent to a low T-shaped carina, somewhat leftward in location. There is usually abnormal bronchial branching, typically a bridging right bronchus (Figs. 6B, 7D, and 8B). There are 2 subtypes, type IIA (Figs. 4H, 5B, 6B, and 7D) with a supernumerary, usually right, tracheal type bronchus (or diverticulum) at the expected location of the normal carina, and type IIB (Figs. 8B, 9C, and 10E) with a long trachea and low carina with an increased angle of the bronchi (T-shaped carina and bridging right bronchus).<sup>6,14,16,19,20</sup> The most important abnormality associated with all variants of type II LPAS is long segment tracheal stenosis with complete cartilaginous rings (Figs. 4-10), typically extending from the usual level of the carina (location of tracheal bronchus, when present) to the abnormally low carina seen in this entity.<sup>6,21,22</sup> This segment has been variably termed trachea or intermediate left bronchus, the airway whereas veering slightly to the left remains in the mediastinum until it bifurcates at the inverted T carina. The association of congenital airway stenosis and LPAS is so strong that



**Figure 2** Type IA left pulmonary artery sling (LPAS)—15-year-old girl with seizure disorder and fever. A, Chest radiograph. The central pulmonary vasculature appears prominent (known ASD), the lungs are symmetrically aerated with no specific abnormality evident. B and C, axial cuts from a chest computed tomography (CT) obtained for possible pneumonia with incidental finding of type IA LPAS. Note high level of sling (black arrow) just below the aortic arch and normal trachea and carina. Right pulmonary artery (RPA—white arrows in B and C).



**Figure 3** Type IA LPAS. Five- and half-year-old F with cough and fever, no stridor. A, MR of chest obtained because of outside chest radiograph (not available) suggesting possible right aortic arch/vascular ring. Axial T1 weighted image demonstrates a normal left arch but a prominent RPA giving rise to the LPA (arrow) passing between the trachea at the carina (\*) and the esophagus (arrowhead). Note the normal caliber airway. B, Type 1 A LPAS, viewed from above. Note LPA arising from proximal RPA and passing between the normal caliber trachea (t) and esophagus (e) en route to the left lung. MPA = main pulmonary artery; LMSB = left main stem bronchus. Reprinted with kind permission from Springer Science + Business Media.<sup>21</sup> (Color version of figure is available online.)

when one is found, the other should be specifically sought,<sup>23</sup> although both long segment stenosis and bridging bronchus occur in the absence of LPAS.<sup>16</sup> The length and degree of stenosis is variable, it can involve the entire trachea or shorter airway segment and can extend into the proximal main stem bronchi (Fig. 8B), right more than left.<sup>23,24</sup>

A more complex variant of LPAS, a partial pulmonary sling has also been described, where there is a normal LPA supplying the left upper lobe and low type II LPAS supplying the lower left lung (Figs. 9C and D). As in other type II slings, this variant also is strongly associated with tracheal stenosis.<sup>16,25</sup> Like complete anomalous LPA, a partial anomalous LPA that passes anterior to the trachea can occur, this is a relatively harmless anatomic variant.<sup>7</sup> It is crucial that the anterior (benign) or posterior (LPAS) course of the LPA be assessed by imaging.<sup>7</sup>

Other lung abnormalities are associated with the type II LPAS and may confound the picture and even mask the pres-

ence of a pulmonary sling. These include right-sided pulmonary hypoplasia and agenesis (Figs. 4, 7, 9, and 10) (22% of type II LPAS) with a small or absent RPA.<sup>4,8,6,11,26</sup> Thus, both bilateral hyperinflation (Fig. 5) or decreased right lung volume (Figs. 4, 7, 9, and 10) are characteristic findings in type II LPAS. Occasional reports of LPAS with left lung hypoplasia have been published.<sup>6,27</sup>

Variation of tracheobronchomalacia or other vascular airway compression may also be a component of airway problems in patients with LPAS, especially in the presence of right lung hypoplasia or agenesis with secondary dextroposition of the heart and distortion of the great vessels with the horizontally oriented aortic arch passing immediately anterior to the trachea (Fig. 10).<sup>26,28</sup>

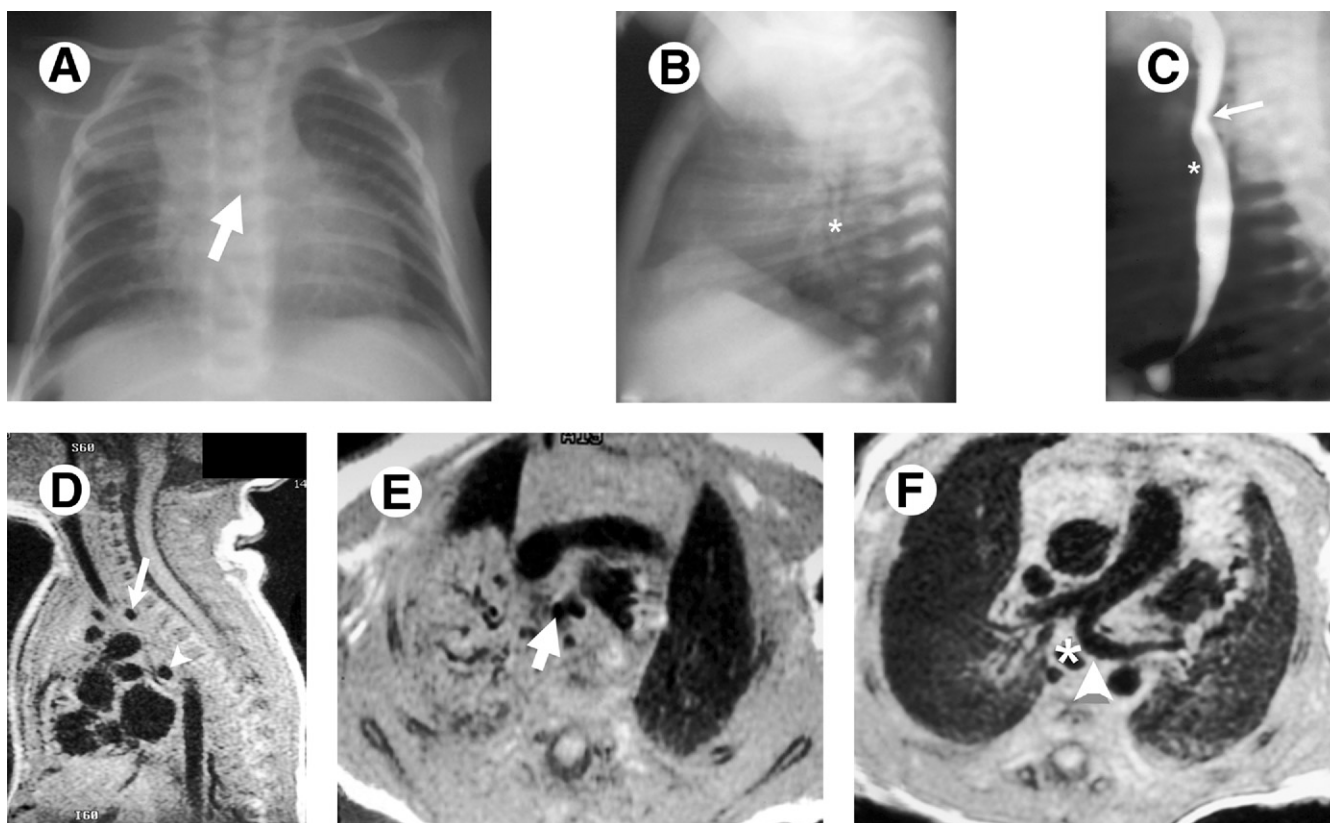
LPAS has been considered part of an expanded spectrum of bronchopulmonary malformations which include various elements of foregut malformations, airway anomalies, lung parenchymal abnormalities, and vascular anomalies.<sup>16</sup> Many of these features have been described in association with LPAS, especially type II, including the aforementioned airway and lung anomalies as well as Scimitar Syndrome and its features, hypoplastic right lung, and pulmonary artery; ipsilateral anomalous pulmonary venous return; pulmonary sequestration and horseshoe lung (Fig. 9).<sup>16,29</sup> There may be stenosis of the origin of the anomalous LPA, in addition peripheral LPA stenoses have been described in conjunction with left lung hypoplasia.<sup>30</sup>

Pulmonary slings are associated with numerous other organ anomalies, including those of the VACTERL spectrum (vertebral anomalies, imperforate anus, cardiac anomalies, tracheoesophageal fistula, renal and limb anomalies).<sup>3,20,24</sup> The most common associations are cardiovascular anomalies (40%-60%), including left superior vena cava (may be unroofed), aberrant right subclavian artery (Fig. 4), aortic coarctation, PDA, partial anomalous pulmonary venous return, atrial septal defect, ventricular septal defect, Tetralogy of Fallot (Fig. 6), and more complex anomalies with single ventricle physiology.<sup>2,8,11,13,14,20-24</sup> Even other vascular rings, such as double aortic arch can be associated with LPAS.<sup>29</sup> Gastrointestinal anomalies, including imperforate anus (Fig. 6), biliary atresia, absent gallbladder, Meckel's diverticulum, and Hirschsprung's disease also coexist with LPAS.<sup>6,14,24</sup>

## Imaging

### Chest Radiographs

There are several radiographic findings that, while not definitive, can suggest a possible pulmonary sling. In type I, a characteristic radiographic finding is right-sided hyperinflation due to partial obstruction and right bronchomalacia; occasionally left-sided hyperinflation is seen.<sup>1,2,16</sup> In the early newborn period, the abnormally ventilated right lung may be fluid filled and appear more solid due to prolonged retention of fetal lung fluid.<sup>1</sup> Occasionally, a right sided tracheal bronchus above the carina may be evident radiographically. The enlarged RPA may produce a rounded right sided soft tissue density indenting the carina and right main stem bronchus.



**Figure 4** Five-day-old M with severe stridor. Type IIA LPAS. A and B, PA and lateral radiographs of the chest demonstrate a smaller right lung with right upper lobe atelectasis. The intrathoracic trachea is barely visible on both views and there is a low T-shaped carina (arrow in A). There is a suggestion of a soft tissue structure between the trachea and esophagus on the lateral view (\*). C, Esophagram—lateral view. There is a large posterior impression on the esophagus (arrow), later shown by MR to be due to an aberrant right subclavian artery. The lower anterior impression between the trachea and esophagus from the LPAS (\*) was not initially appreciated. D, Oblique sagittal T1 weighted MR image demonstrates a markedly narrowed intrathoracic airway with 2 vascular impressions, the upper from the aberrant right subclavian artery (arrow) and the lower from the LPAS (arrowhead). E-G, Axial MR images. E, Demonstrates the right upper lobe bronchus origin (arrow). Note right upper lobe atelectasis. F, Low LPAS (arrowhead) between slit like trachea and esophagus (asterisk). G, Low horizontal carina with bridging right bronchus (arrows), left bronchus (arrowhead). H, 3D reconstruction of vessels and airway, posterior view. Note aberrant right subclavian artery (arrowhead), low level of LPAS (blue), right upper lobe bronchus (white) with long segment tracheal narrowing below the right upper lobe bronchus to the low horizontal carina. Appearance consistent with type IIA PA sling. I, echocardiogram—also identified LPA (arrow) arising from RPA (arrowhead). J, Subsequent angiogram (lateral view with injection in MPA) confirmed LPAS. RPA (arrowhead), LPA (arrow). C, D, F, H, reprinted with permission from Springer Science + Business Media.<sup>21</sup> (Color version of figure is available online.)

On the lateral chest radiograph, in both types, a small soft tissue mass can sometimes be appreciated between the mid trachea and esophagus (Fig. 4B), representing the LPA coursing between these 2 structures.<sup>2</sup> This finding is often better visualized on an esophagram (Figs. 4C and 7B).

Either bilateral hyperinflation (tracheal stenosis) or right-sided volume loss (lung abnormality) on a chest radiograph should raise the suspicion for a type II LPAS, especially when the trachea appears narrow or difficult to visualize and the carina is low and horizontal.<sup>1,6,16</sup>

## Esophagram

As is the case with vascular rings, an esophagram is of limited value in assessing a pulmonary sling. A small soft tissue mass may be appreciated between the trachea and esophagus

(Figs. 4C and 7B) as opposed to vascular rings where the posterior compressing vessel is posterior to both the trachea and esophagus.<sup>2</sup> However, this is not invariably present and other lesions, such as a bronchogenic cyst may occupy the same location.<sup>2</sup> Also, an anomalous right subclavian artery is one of the anomalies that may be associated with LPAS. This produces a posterior oblique impression on the esophagus and may confuse the picture (Fig. 4C). The esophagram and fluoroscopic visualization of the chest provides an additional opportunity to observe tracheobronchomalacia or tracheal stenosis. High kVp magnified technique provides the best detail of the airway (Fig. 10B).

If there is reasonable suspicion of a pulmonary sling on the chest radiograph, an esophagram is probably an unnecessary additional study. However, it is important to be able to rec-

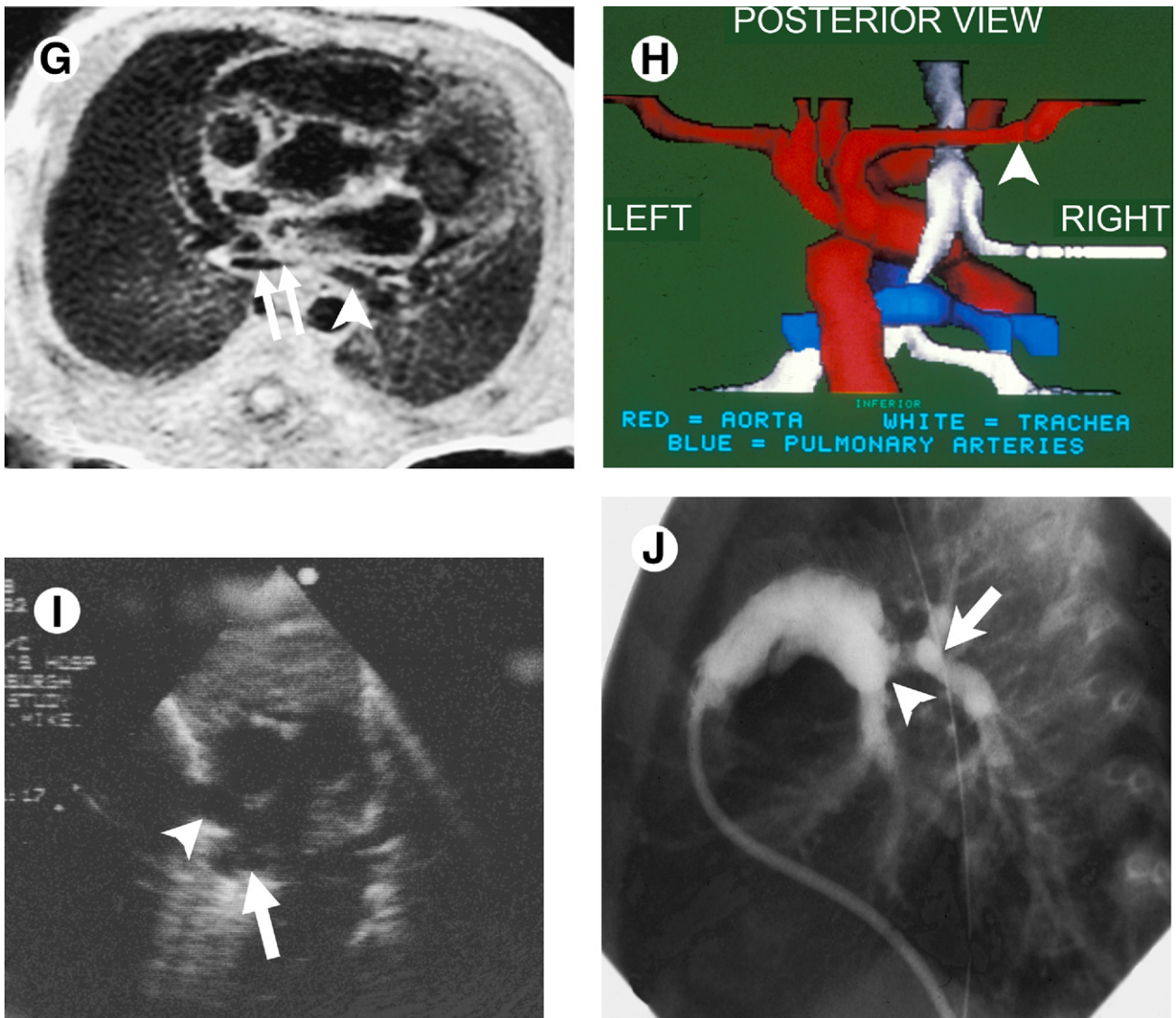


Figure 4 (continued)

ognize a possible pulmonary sling when an esophagram is obtained (Fig. 7B).

### Ultrasound

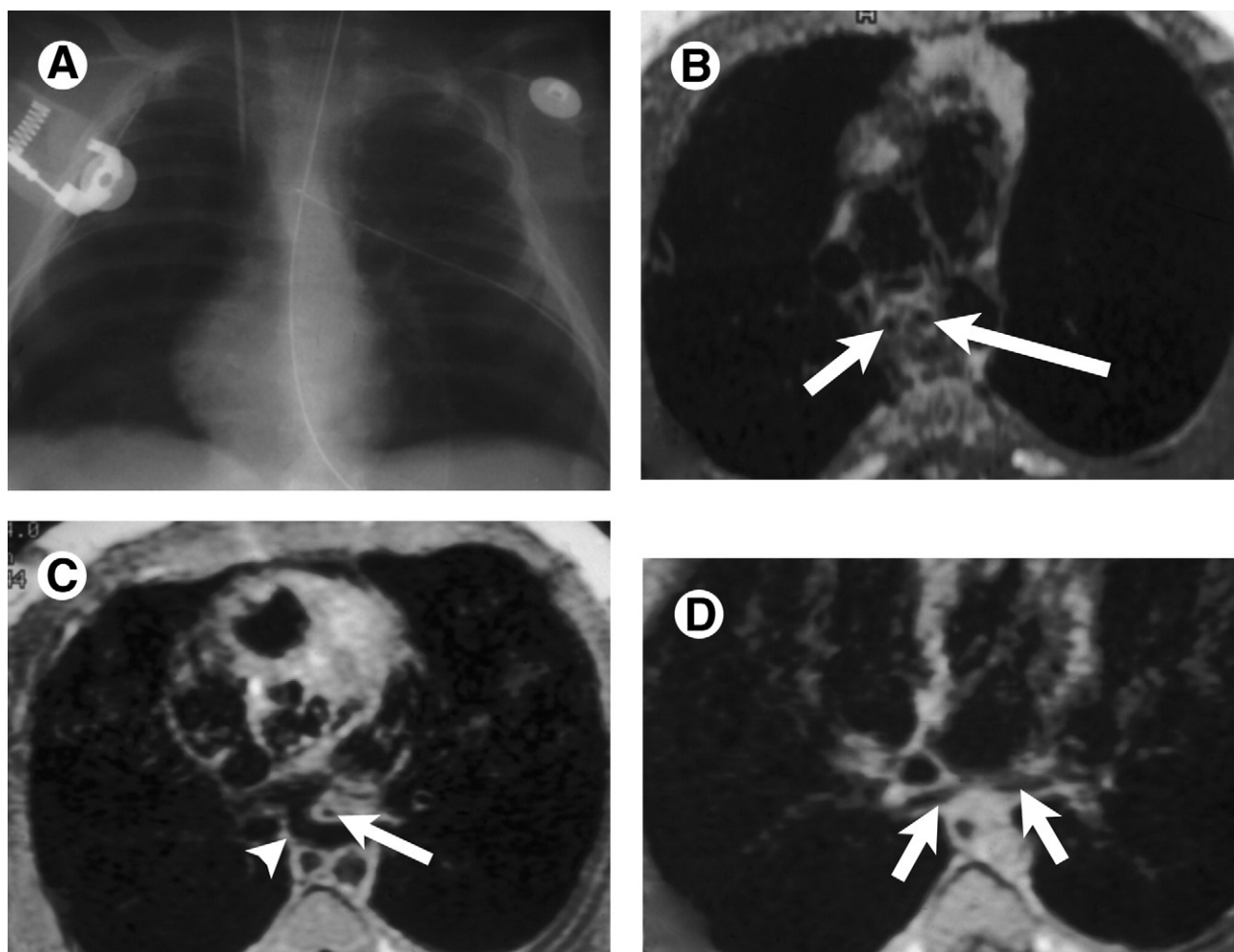
The thin body habitus of young children is especially suited to evaluation by ultrasound. Exquisite highly detailed images are obtained of the heart and central great vessels. More peripheral vessels and structures close to the lung are poorly seen or not visualized at all. If specifically evaluated, a pulmonary sling may be seen on ultrasound imaging (Fig. 4I); however, the diagnosis may be missed, especially when the anatomy is considerably distorted as in right lung agenesis.<sup>4</sup> Ultrasound does not image the airway well and the restricted field of view fails to optimally display the complex three-dimensional (3D) relationships between the lungs, heart, airways, and vascular structures.<sup>4</sup> Although associated intracardiac abnormalities such as atrial septal defect, ventricular septal defect, PDA, and Tetralogy of Fallot are thoroughly assessed on ultrasound, the spectrum of other anomalies as-

sociated with the pulmonary sling are incompletely evaluated.

Prenatal ultrasound imaging may be the first study to pick up a lung anomaly. LPAS should be added to the list of abnormalities that can cause lung asymmetry; the differential includes cystic pulmonary airway malformation, pulmonary sequestration, bronchial atresia, congenital lobar hyperinflation, and congenital diaphragmatic hernia.<sup>4,16,18</sup> In 1 published case, there was a persistently uniform enlarged echogenic right lung on ultrasound and uniform bright signal on prenatal magnetic resonance MR thought to be congenital lobar hyperinflation.<sup>18</sup> The LPAS with compression and distortion of the distal trachea and right bronchus was not identified until postnatal MR imaging (MRI) and computed tomography (CT) were obtained.

### Conventional Angiography

Prior to the availability of high quality CT and MR angiography (MRA), the diagnosis of pulmonary sling was made or



**Figure 5** Type IIA LPAS, newborn male with severe respiratory distress, difficult to ventilate. A, Anteroposterior chest radiograph. Left chest tube for prior pneumothorax. The lungs are hyperaerated and the trachea difficult to visualize. B, Axial T1 weighted MR images. There is a tracheal type right upper lobe bronchus (right arrow) at the normal carinal level with the airway continuing as a very stenotic intermediate left bronchus (left arrow, B, C) to the level of the LPAS. C, Pinpoint airway (arrow) at level of low LPAS (arrowhead). D, Low T-shaped carina with horizontal orientation of normal caliber left and bridging right bronchi (arrows). (C and D) reprinted with kind permission from Springer Science + Business Media.<sup>21</sup>

confirmed by catheter angiography (Fig. 4J). The pulmonary sling can often readily be demonstrated and any associated intracardiac anomalies fully assessed. The airway and lungs are visible but are relatively incompletely evaluated compared with CT and MRI.<sup>11</sup> With the severe distortion of normal anatomy that accompanies right lung agenesis, even the LPAS can be missed.<sup>31</sup> This technique is invasive (catheterization), often requires anesthesia, and is associated with high radiation dose and possibly large quantities of intravenous contrast.

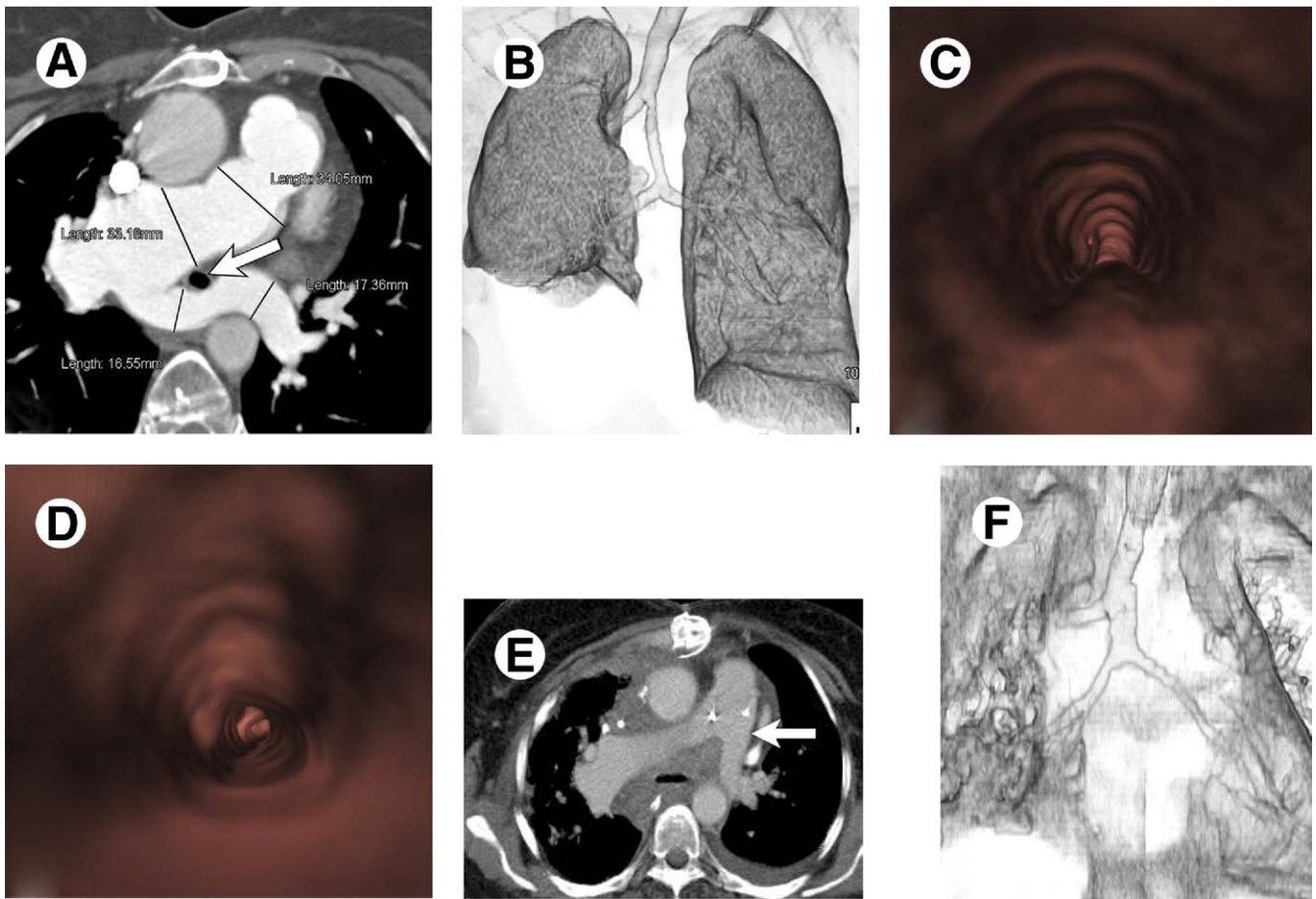
### Computed Tomography Imaging

Both CT and MR can be used to demonstrate most of the vascular, airway, and lung findings associated with LPAS (Figs. 2, 6, 7, and 8). Both can be reformatted in multiple planes with excellent two-dimensional and 3D display of anatomy that is very useful in surgical planning.<sup>10,14,19,32,33</sup> However, the diagnosis and important details can be over-

looked with poor quality imaging on both CT and MR (Fig. 9B).

CT has the advantage of much better visualization of lung parenchyma and airways, higher spatial resolution, and faster scanning with lower requirement for sedation or anesthesia.<sup>13</sup> CT angiographic technique is best for optimally evaluating LPAS. The disadvantage of CT is exposure to ionizing radiation, of especial concern in young children. Careful attention to CT parameters can produce high quality, relatively low dose studies. Electrocardiographically gated CT angiography (CTA), in particular, imparts a much higher dose of radiation (3-4X greater) than a nongated study. Gated CTA is usually not necessary in evaluating a potential pulmonary sling as the associated intracardiac anomalies are well evaluated by cardiac ultrasound and the extracardiac vascular and airway abnormalities are well-defined without cardiac gating.

In our department, we use 2 cc/kg of contrast (Omnipaque 350) injected intravenously either by hand bolus (in young



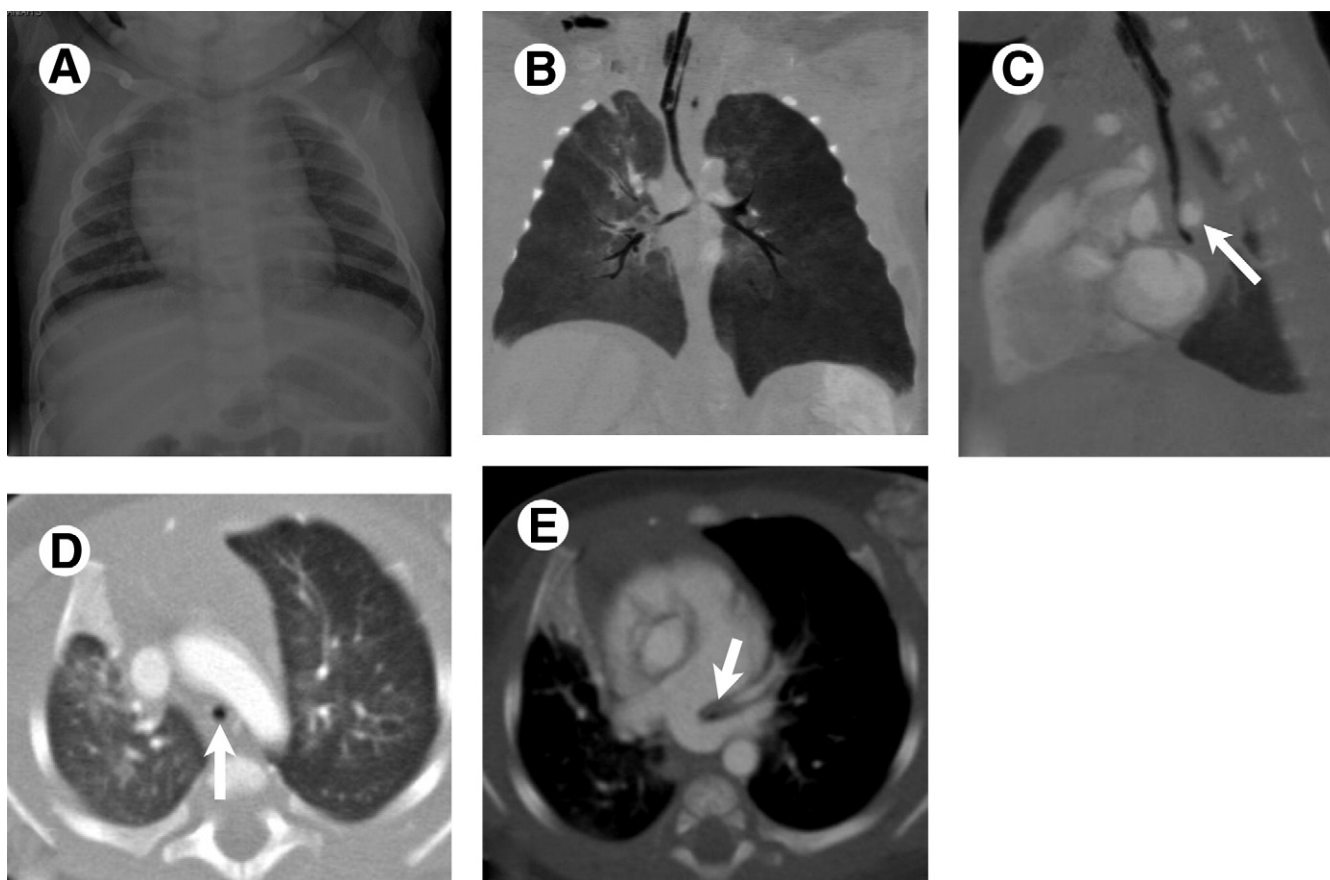
**Figure 6** Type IIA LPAS. Forty-nine-year-old F postrepair of Tetralogy of Fallot in childhood. Has had lifelong shortness of breath and exercise intolerance ascribed to tracheomalacia. A, Axial CT MIP image. The MPA and RPA are enlarged with a LPAS and rounded, narrowed trachea (arrow). B, Coronal CT volume rendered reconstruction of the chest. The upper trachea is normal in caliber, there is long segment airway stenosis extending from the right upper lobe bronchus origin to the lower T-shaped carinal bifurcation, typical anatomy of type IIA LPAS. Note: elevation of the right hemidiaphragm (known right paralysis). C and D, CT virtual bronchoscopy. C, Upper trachea. Note the large caliber of the airway and the posteriorly bulging membranous portion of the normal trachea. D, The more inferior airway has a much smaller caliber and complete posterior cartilagenous rings. E and F, Postoperative CT (a routine chest CT not a CTA) – 5 days status post LPA reimplantation, sliding tracheoplasty, right diaphragmatic plication, and pulmonary valve placement. E, Axial image shows patent reimplemented LPA (arrow) and decreased size of MPA and RPA. F, 3D reconstruction of airway demonstrates shortened larger diameter trachea and lower right diaphragm. Courtesy: Francis Chan MD, Stanford University Medical Center. (Color version of figure is available online.)

infants) or via injector if possible. The rate of intravenous injection varies with the size of the child and available IV site, from 1 to 4 cc/s. A useful rule of thumb is that the length of the contrast bolus should exceed the scan time by approximately 10 seconds to ensure that the pulmonary vessels are opacified throughout the scan. In very small infants it is often helpful to dilute the contrast by 1/3 or 1/2 with saline to obtain a longer contrast bolus. A chaser saline bolus at the same rate should immediately follow the contrast using a dual injector or double stopcock for hand injection. This ensures that the child receives all the intended contrast as a significant amount may otherwise remain in the tubing, and also continues the progression of the contrast bolus through the vascular system. We monitor the progression of the bolus with intermittent low dose CT images at the desired location, triggering the scan when the vessel in question, in this case

pulmonary artery, is opacified. There is a short delay (minimum 4 s) on our scanner (Siemens sensation 64) between triggering and the start of scanning.

We attempt to adhere to ALARA principles by paying close attention to technique. We use kVp settings of 80 kVp (<20 kg) or 100 kVp. CTA is well suited to lower kVp techniques due to the higher contrast resolution afforded despite some increased noise. We use the dose saving mAs automodulation technique available with the scanner. Reference mAs is usually set at 100-150 mA for CTA, gantry cycle time of 0.5 seconds, detector collimation 0.6 mm, and pitch of 1. We also routinely use a padded breast shield in all young girls after the scout image to further decrease breast radiation dose.

We usually obtain 0.6-1 mm axial helical slices using a soft tissue algorithm, these can be reconstructed with a 50% over-



**Figure 7** Nine-week-old F with marked respiratory distress. Type IIB LPAS. A, The intrathoracic trachea is poorly visualized on this chest radiograph. There is a low T-shaped carina with symmetrically aerated lungs. B and C, Coronal and sagittal minimum intensity projection, CT reconstructed images of the airway demonstrating anatomy suggestive of pulmonary sling type IIB. There is long segment lower tracheal and proximal bronchial narrowing with a low T-shaped carina. The LPA (arrow in C) is seen low behind the narrowed airway. D and E, Axial CT images. D, At the level of the aortic arch there is a rounded markedly stenotic trachea (arrow). E, The airway (arrow) continues to be markedly stenotic at the level of the low LPAS. Courtesy: Ron Cohen, MD, Oakland Children's Hospital.

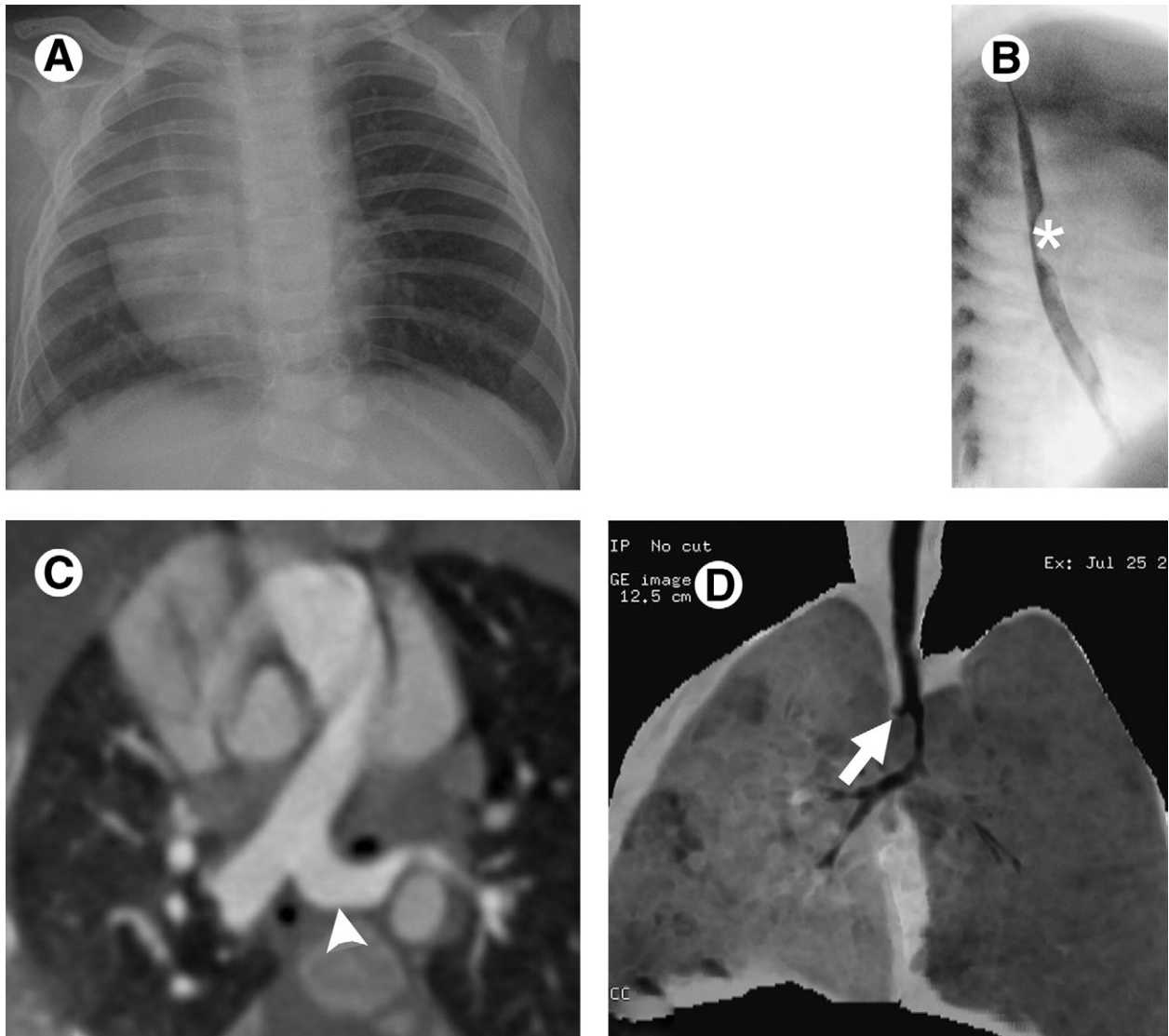
lap. In addition to the axial images we routinely reformat 2-mm axial slices with a lung algorithm and 2-mm soft tissue coronal and sagittal images. The images are also sent to an independent workstation or the 3D laboratory for 3D reconstructions. Direct interaction with the dataset with multiplanar volume rendered 3D, and maximum and minimum intensity projection reformats of the anatomy are often helpful in detailed evaluation and demonstration of the anatomic relationships (Figs. 3B, 4H, 6B, 7D, 8B and C, 10E and F).<sup>19</sup> Virtual bronchoscopic fly through of the airway is an additional useful reconstruction technique (Figs. 6C and D).<sup>10</sup>

### Magnetic Resonance Imaging

Lack of ionizing radiation exposure is the major advantage of MRI (Figs. 3-5, 9 and 10). The disadvantages are long scan times with greater need for sedation and anesthesia and decreased spatial resolution compared with CT as well as relatively suboptimal visualization of the airways and lungs.<sup>14,21</sup> Increased cost and less scanner availability of MR versus CT are additional considerations. The scanner we use is a 1.5 or 3 T GE Signa unit.

Both gated and respiratory compensated thin section black blood (spin echo or double inversion) and white blood (gradient echo) sequences can be obtained to evaluate the vascular and airway anatomy. Direct thin cut (3-4 mm) oblique T1 weighted images of the trachea along the airway long axis (posted off the scout images) in sagittal and coronal planes are helpful but time-consuming.<sup>14,16,34</sup> Breath hold MRA is a key sequence, multiple phases can readily be obtained if needed. The airway is only seen reasonably on later phases when the vascular opacification has faded somewhat. MRA is obtained dynamically following a bolus of intravenous contrast (gadolinium). We use a contrast dose of, 0.2 cc/lb<sup>-1</sup> (double dose) followed by a 10-30 cc saline bolus. In young infants, the contrast can be injected by hand with simultaneous triggering of the scan with the start of the contrast injection. In older children, we use a small (1-2 cc) test bolus injected at the same rate as the later contrast and followed by the same saline bolus. We calculate the timing of the MR scan in such a way that the optimal opacification of vessels with contrast occurs in the middle of the scan sequence, when the center of K space (which determines contrast resolution) is





**Figure 8** Six-month F with stridor and feeding difficulty. Type IIB LPAS. A, Chest radiograph. The right lung appears hypoplastic and the airway is poorly visualized. B, Esophagram with soft tissue mass (\*) between the trachea and esophagus suggesting LPAS. C, Axial CT shows corresponding LPAS (arrowhead) passing between the airway and esophagus. D, Coronal CT minimum intensity projection reconstruction, type IIB LPAS airway appearance. There is a blind diverticulum (arrow) on the right off the upper trachea and long segment narrowed airway distally to the low horizontal carina. The left main bronchus is poorly visualized due to fluid/mucous.

being filled in the sequence that we use. The formula used is as follows:

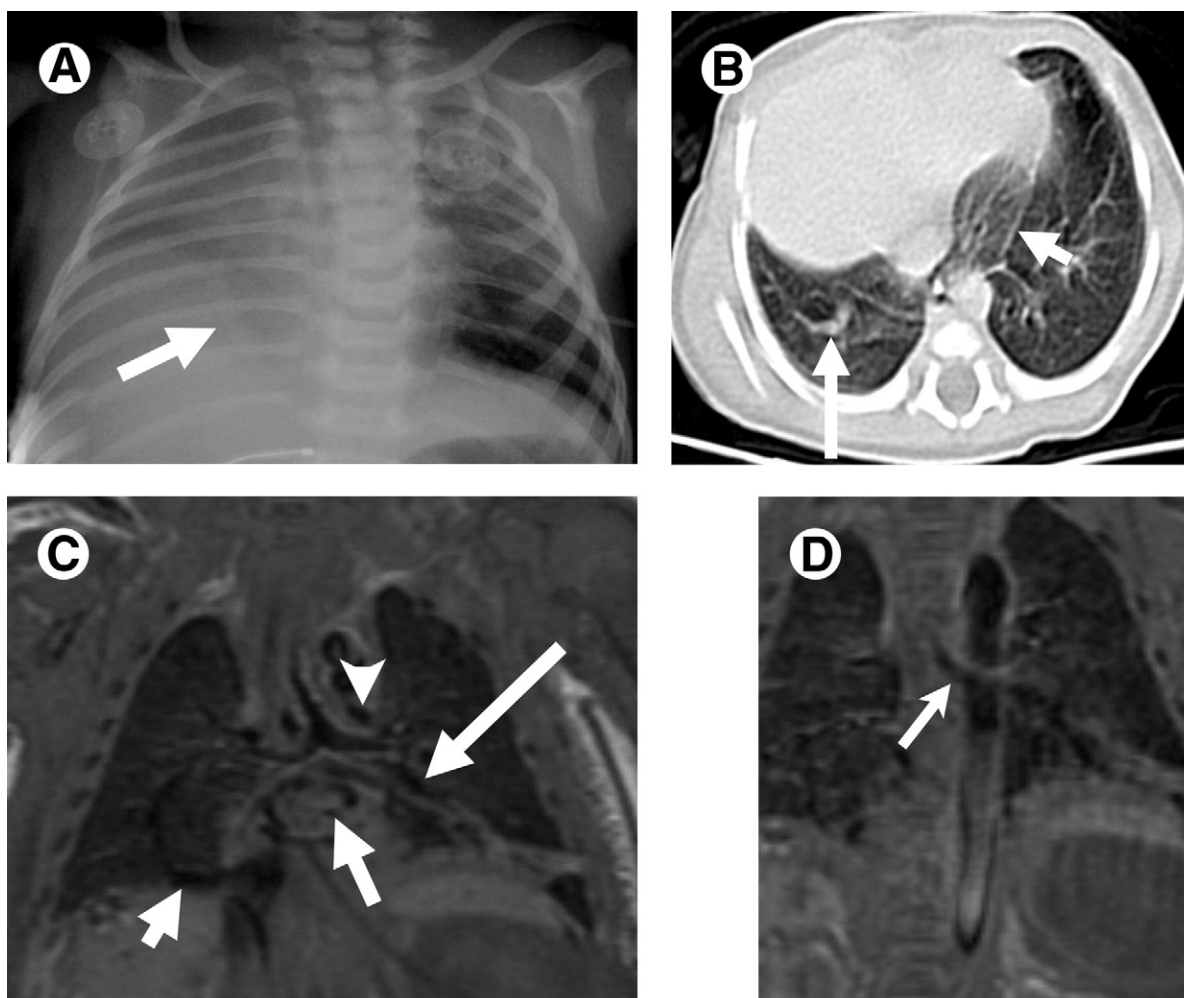
$$\text{scan delay} = \text{optimal time of test bolus visualization} \\ - \text{scan time}/2 + \text{injection time}/2.$$

For example, the best visualization of the desired vessel, in this case, pulmonary artery occurred at 10 seconds on the test bolus, scan time (1 phase) = 20 seconds, and injection time 10 cc (Ten cubic centimetres contrast at 1 cc/sec 9), the formula results in a scan delay of  $10 - 20/2 + 10/2 = 5$  seconds. Time resolved MRA has been described for evaluation of pulmonary sling,<sup>35</sup> we tend to prefer the higher resolution conventional multiphase MRA.

Additionally, as necessary MR can be used to evaluate intracardiac anatomy and biventricular function. Phase-contrast MR sequences can be used to assess QP/QS and shunting as well as valvular regurgitation and differential pulmonary flows.<sup>34</sup>

Patients with respiratory distress and possible LPAS are at high risk for respiratory decompensation and need to be carefully monitored while undergoing both CT and MR imaging, preferably by anesthesia or intensive care unit physicians. Oxygen saturation, pulse, blood pressure, and electrocardiogram tracings should be continuously assessed.

Prenatal MRI is becoming more commonplace in the evaluation and monitoring of fetal lung anomalies. LPAS should be an additional consideration when a bronchopulmonary malformation is suspected.<sup>18</sup>



**Figure 9** Ten-days-old M with persistent respiratory symptoms. Type IIB LPAS. A, chest radiograph—there is a small volume hazy right lung with ill-defined vertical linear shadow at the right lung base thought to possibly represent an anomalous vein as part of scimitar syndrome. B, Poor quality noncontrast CT. Pulmonary sling was not visible. Scimitar vein (arrow) and horseshoe lung (short arrow) were not appreciated. C and D, Partial LPAS and scimitar syndrome. T1 weighted coronal MR with typical narrow distal airway and T-carina associated with LPAS type IIB. In this child there is a partial sling, the upper lobe LPA arises normally (arrowhead) whereas the lower LPA arises from the RPA (C, long arrow, D, arrow). In addition, there is a right sided anomalous scimitar vein (right arrow in C) draining to the inferior vena cava and atelectatic sequestered horseshoe lung (middle arrow in C). A, C, D, reprinted with kind permission from Springer Science + Business Media.<sup>16</sup>

## Nuclear Medicine Imaging

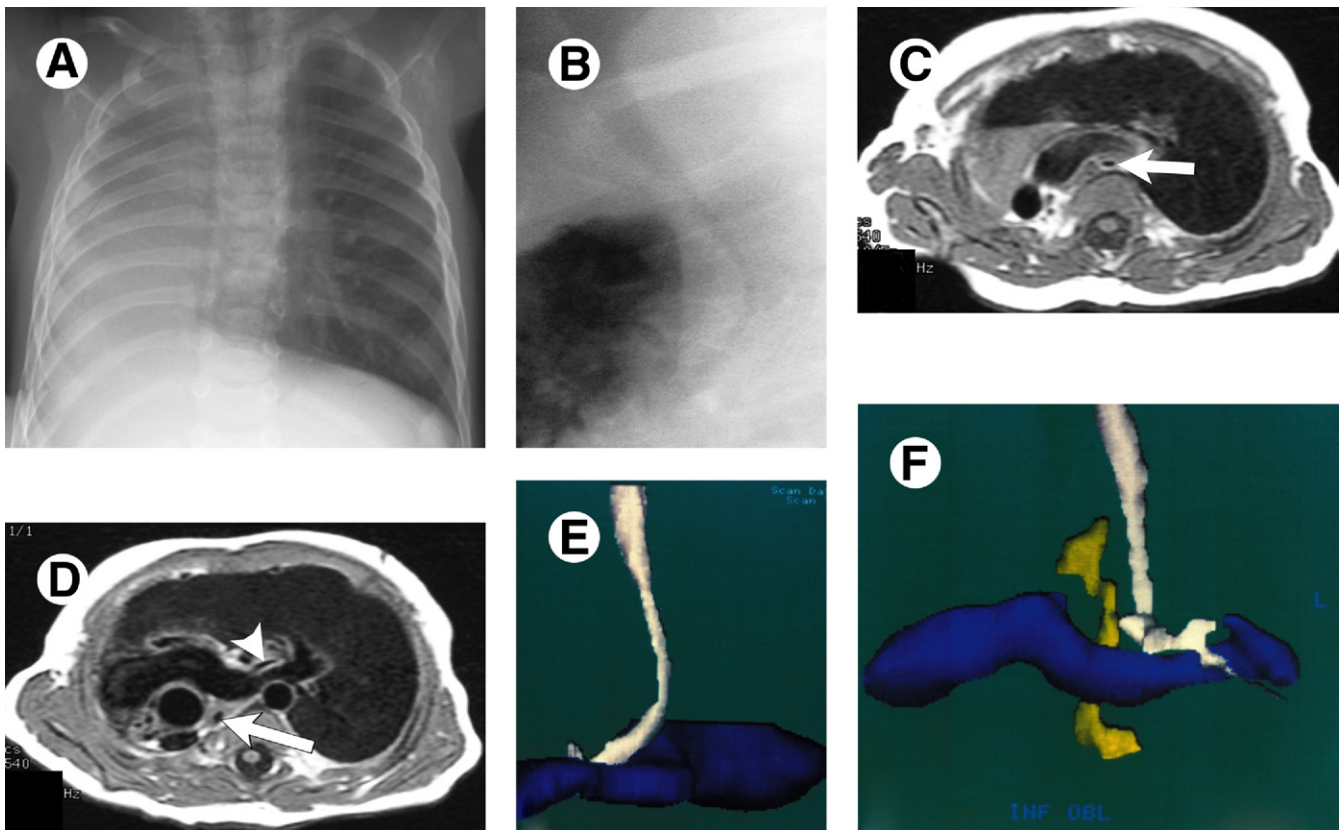
This has somewhat limited use in pulmonary sling, being most useful in assessing differential pulmonary perfusion (Technetium Macroaggregated Albumen) especially postoperatively. Occasionally, lung ventilation scan (Inhaled xenon wash-in and washout) may be helpful in differentiating airway from vascular pathology.

## Bronchoscopy

Bronchoscopy of patients with pulmonary sling may be fraught with difficulties both in performance and interpretation. It is especially potentially hazardous in the situation of severe tracheal stenosis and can precipitate an acute respiratory crisis.<sup>14,36</sup> The bronchoscope often cannot pass beyond the area of tracheal stenosis to evaluate the more distal air-

way, which is readily assessed by CT.<sup>10,14</sup> The unusual anatomy is easy to misinterpret: a case has been described in which the long segment airway narrowing was completely overlooked with the study being called normal because the tracheal bronchus was misinterpreted as the carina.<sup>21</sup> Bronchoscopy has the advantage of evaluating mucosal detail, as well as dynamic airway changes, such as tracheobronchomalacia and location of extrinsic pulsation.

Bronchoscopy is most useful in the operating room, where airway management is carefully controlled. The surgeon in the operating room often cannot accurately evaluate the extent of tracheal stenosis externally. Preoperative multidetector computed tomography may overestimate the length of airway stenosis, especially in the presence of intraluminal mucous.<sup>10,37</sup> Bronchoscopy, in conjunction



**Figure 10** Two-year-old M with longstanding respiratory symptoms. A, Chest radiograph—there is marked cardiomeastinal shift to the right with a small volume right hemithorax. Aeration in the right upper chest could be due to right upper lung aeration versus left lung herniation. Findings suggestive of right lung agenesis or marked hypoplasia. B, Magnified high KV lateral view of the airway with narrowed and posteriorly bowed intrathoracic trachea. C and D, Axial T1W MR images. C, There is absence of the right lung with left lung herniation to the right anteriorly. The trachea (arrow) is markedly narrowed as well as compressed posteriorly by the crossing aortic arch. D, There is pulmonary sling anatomy characterized by the LPA crossing to the left between the airway (left bronchus—arrowhead) and the esophagus (arrow). Normally, the LPA is anterior to the airway. E and F, 3D reconstructions of the airway in LPO and inferior oblique projections. The long segment tracheal narrowing consistent with type IIB sling is illustrated with the LPA passing between trachea (white) and esophagus (yellow) to reach the single left lung. B-D, reprinted with permission from Newman B, Gondor M. *Am J Roentgenol* 168:55-58, 1997.<sup>26</sup> (Color version of figure is available online.)

with surgical methods of marking the exposed trachea, identifies complete cartilaginous rings and accurately delineates the length of airway stenosis.<sup>3,32</sup> Post repair bronchoscopic assessment of the tracheal lumen and anastomoses is also commonly employed.<sup>3,24,38</sup>

### Bronchography

Although bronchography was useful in the past in defining the associated airway anomalies,<sup>2</sup> the high resolution reconstructed images now available with multidetector computed tomography imaging render bronchography obsolete and unnecessary in the imaging workup of the airway anomalies associated with LPAS.<sup>10,32</sup>

### Management of LPAS

Patients with LPAS are managed based on clinical symptoms and anatomy. Asymptomatic patients with a type I sling can

be followed clinically.<sup>12</sup> In those with respiratory symptoms, reimplantation of the LPA to the left is appropriate and often sufficient management. The PDA or ductal ligament are also usually ligated. Respiratory symptoms, usually mild airway compression and malacia abate postoperatively. Additional lesions, such as cardiac anomalies, are dealt with on their own merits.

It has been emphasized repeatedly in the literature that reimplantation of the LPA in the type II sling will not result in improvement of respiratory symptoms and that surgical management should address both the anomalous LPA (reimplantation or translocation anteriorly) and the airway stenosis.<sup>2,8</sup> Nonetheless, there are still relatively recent reports and anecdotes of children in whom the airway anomalies are not addressed in the hope that they will improve spontaneously.<sup>8,23</sup> Survival to adulthood without surgery is possible with less severe congenital tracheal stenosis (Fig. 6). Even in the presence of complete tracheal rings, it has been suggested

that some tracheal growth can occur, although patients often remain variably symptomatic.<sup>3,24,33</sup>

The airway component is the most difficult issue to deal with surgically, and effective long segment tracheal repair techniques have only been developed relatively recently. Earlier tracheal repair techniques included resection or patching of the stenotic segment with pericardium, costal cartilage, or tracheal autograft.<sup>3,24,38,39</sup> Patch techniques have been variably successful; granulation tissue formation, restenosis, and collapse or disruption of the patch have been troublesome complications.<sup>25,38,39</sup> Resection and direct reanastomosis entails extensive mobilization of the trachea and bronchi and is preferred when a shorter segment (<25%-40% of tracheal length) is involved but is difficult to accomplish when a long segment is stenotic.<sup>3,24,38,39</sup> Nonetheless there have been reports of long-term success with this method.<sup>21,23</sup>

The technique that seems to be most commonly employed currently for long segment stenosis is sliding tracheoplasty which has the advantage of limited dissection and using the trachea itself for the repair, so the airway is lined by normal mucosa and can grow with the child.<sup>24,25,37,39,40</sup> In this technique, the trachea is first transected at the identified midpoint of the stenotic segment. Following this long vertical anterior and corresponding posterior tracheal incisions are made in the proximal and distal abnormal airway, respectively. These are then trimmed, slid together, and sutured with resultant shortening of this segment of airway by 50% but doubling of the diameter of the lumen and quadrupling of the cross-sectional area of the trachea (Fig. 6E).<sup>3,24</sup> The incisions can be extended into the proximal bronchi if needed and the technique is also applicable when there is right lung agenesis or hypoplasia.<sup>37</sup> A modification of this technique using the right tracheal upper lobe bronchus in the repair has also been described.<sup>41</sup> Granulation tissue formation especially distally has been a postoperative concern.<sup>24,39</sup>

When right-sided lung hypoplasia or agenesis is present, an additional aortopexy may be beneficial to move the anteriorly crossing aorta away from the airway.<sup>28</sup> Management of other associated anomalies may also be necessary as dictated by clinical symptoms and anatomy.

Altering the course of the LPA so that it is anterior to the trachea is most commonly accomplished by transecting the vessel at its origin from the RPA and reimplanting it anteriorly with good resultant patency (Fig. 6F). Simple translocation of the LPA anteriorly without reimplantation has been advocated when the trachea is transected while being repaired at the same surgery. LPA translocation is most effective in the setting of hypoplasia or agenesis of the right lung.<sup>37</sup> In other situations, concern has been raised that translocation may cause tension, kinking, and narrowing of the proximal LPA and anterior compression of the trachea.<sup>3</sup>

Postoperative imaging depends on the clinical issues. In the early postoperative period, chest radiographs are relied on for tube and line position and gross assessment of the airways and lungs. Both MR and CT can be used as needed to evaluate the postoperative central airways and relocated LPA (Figs. 6E and F). CT, with its faster imaging and higher spatial resolution is often preferred and remains vastly superior in

evaluating lung parenchymal issues. Both MR phase-contrast imaging and nuclear lung perfusion can be used to obtain differential lung blood flow. Bronchoscopy is employed to evaluate the tracheobronchial repair and development of granulation tissue with concomitant balloon dilatation of the airway or resection of obstructing granulation tissue.

## Summary

This review attempts to illustrate and discuss the important imaging aspects of the LPAS and ring/sling complex. A thorough understanding of the spectrum of this complex anomaly on imaging studies as well as the current surgical approaches to this entity is necessary so that imagers provide the information needed to make appropriate patient management decisions.

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