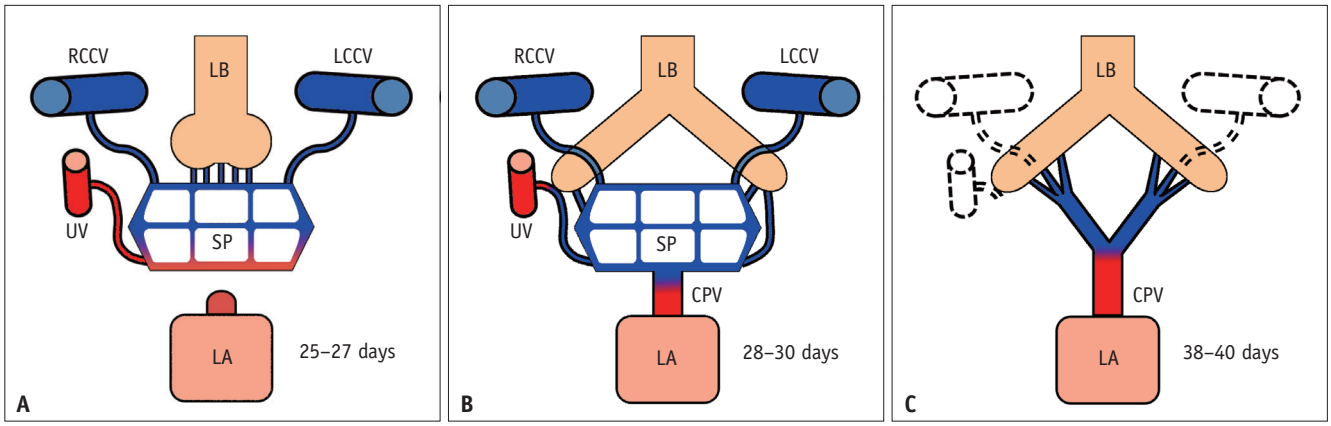
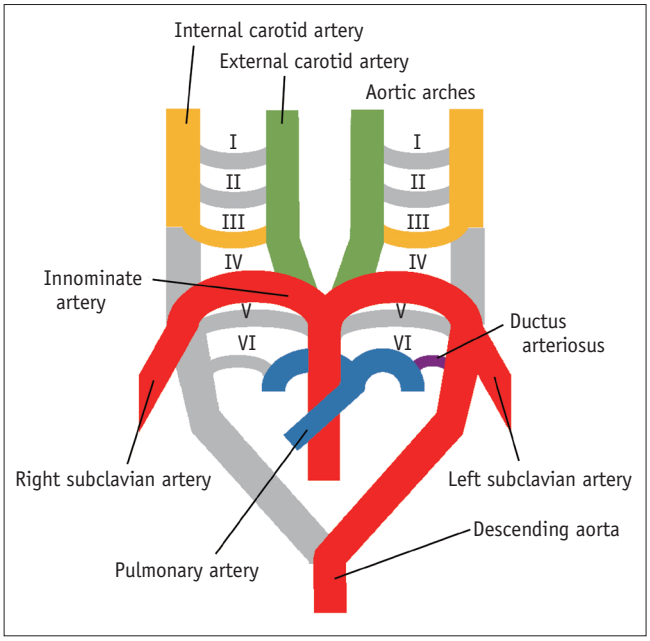
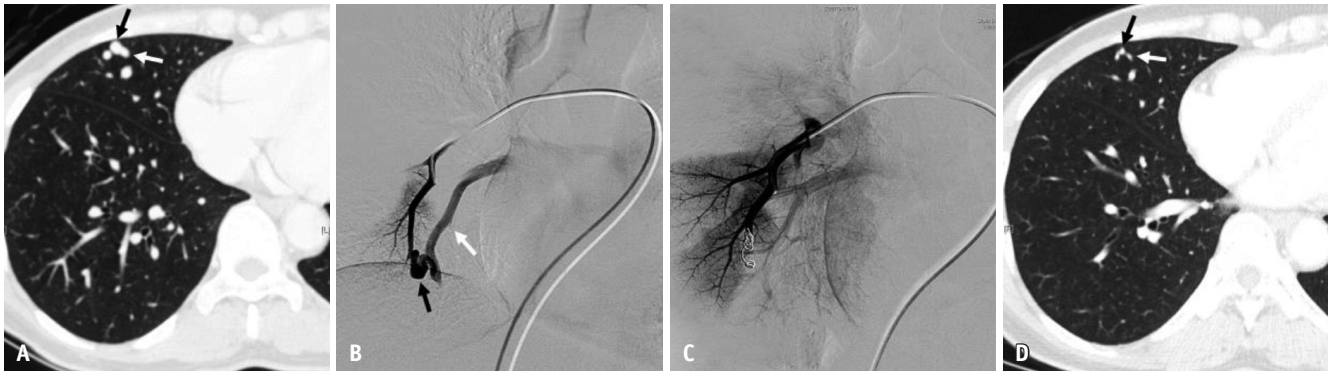
SUPPLEMENT



Supplementary Fig. 1. Illustration of the normal development of the pulmonary veins from the SP during the first month of embryogenesis (A-C). CPV = common pulmonary vein, LA = left atrium, LB = lung buds, LCCV = left common cardinal vein, RCCV = right common cardinal vein, SP = splanchnic plexus, UV = umbilicovitelline vein

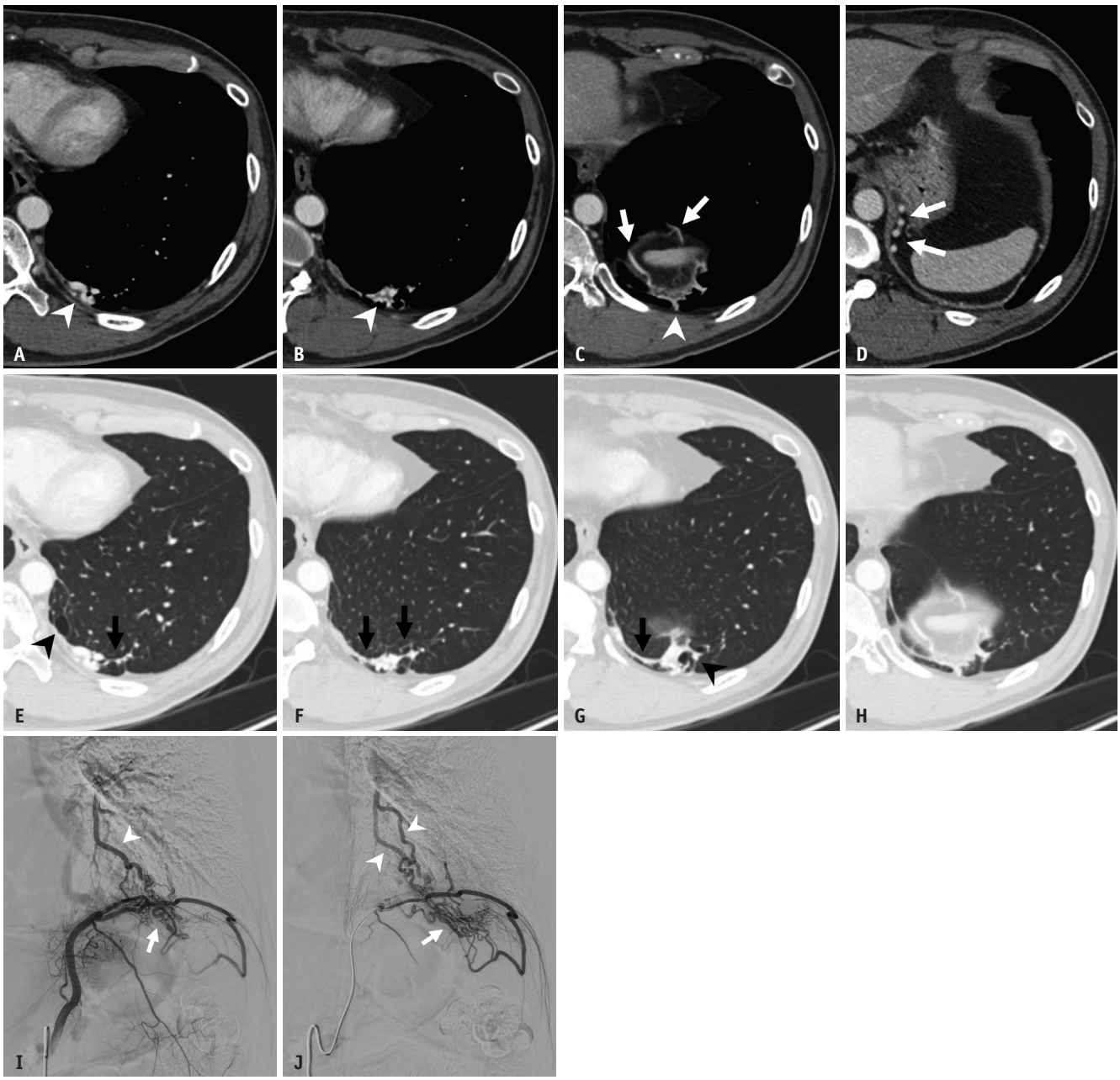


Supplementary Fig. 2. Outline of the embryological development of the pulmonary arteries from the 6th pharyngeal arch.



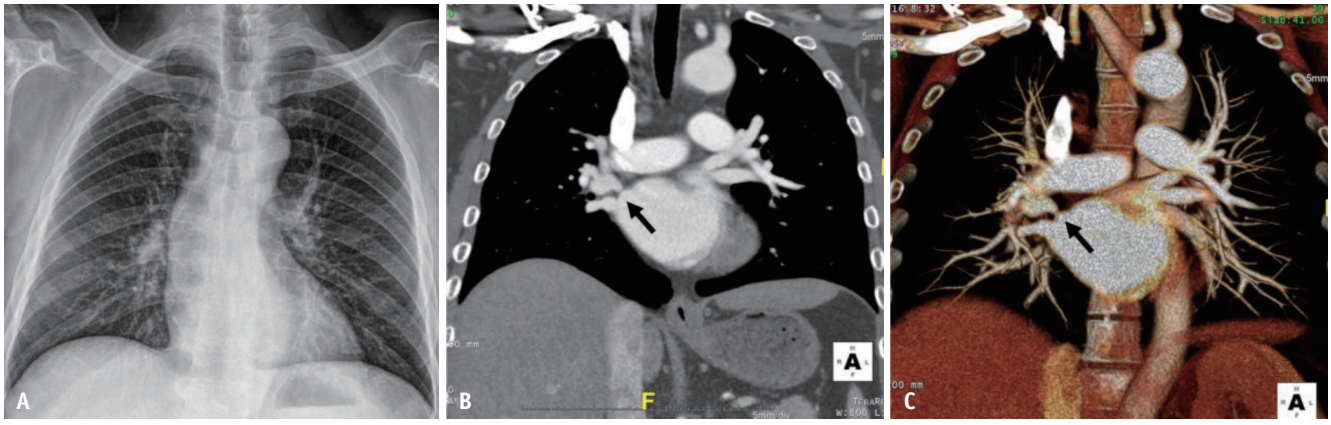
Supplementary Fig. 3. A 48-year-old female patient who underwent transcatheter embolization of a pulmonary arteriovenous malformation.

A, B. Axial chest CT scan and a selective pulmonary angiogram acquired before embolization shows a sac (black arrows), and a dilated drainage vein (white arrows) of pulmonary arteriovenous malformation in the right middle lobe. **C.** Pulmonary angiogram obtained after embolization demonstrates complete occlusion of the pulmonary arteriovenous malformation. **D.** Axial chest CT scan obtained 17 months after embolization demonstrates a decrease in diameter of the sac (black arrow) and the draining vein (white arrow).

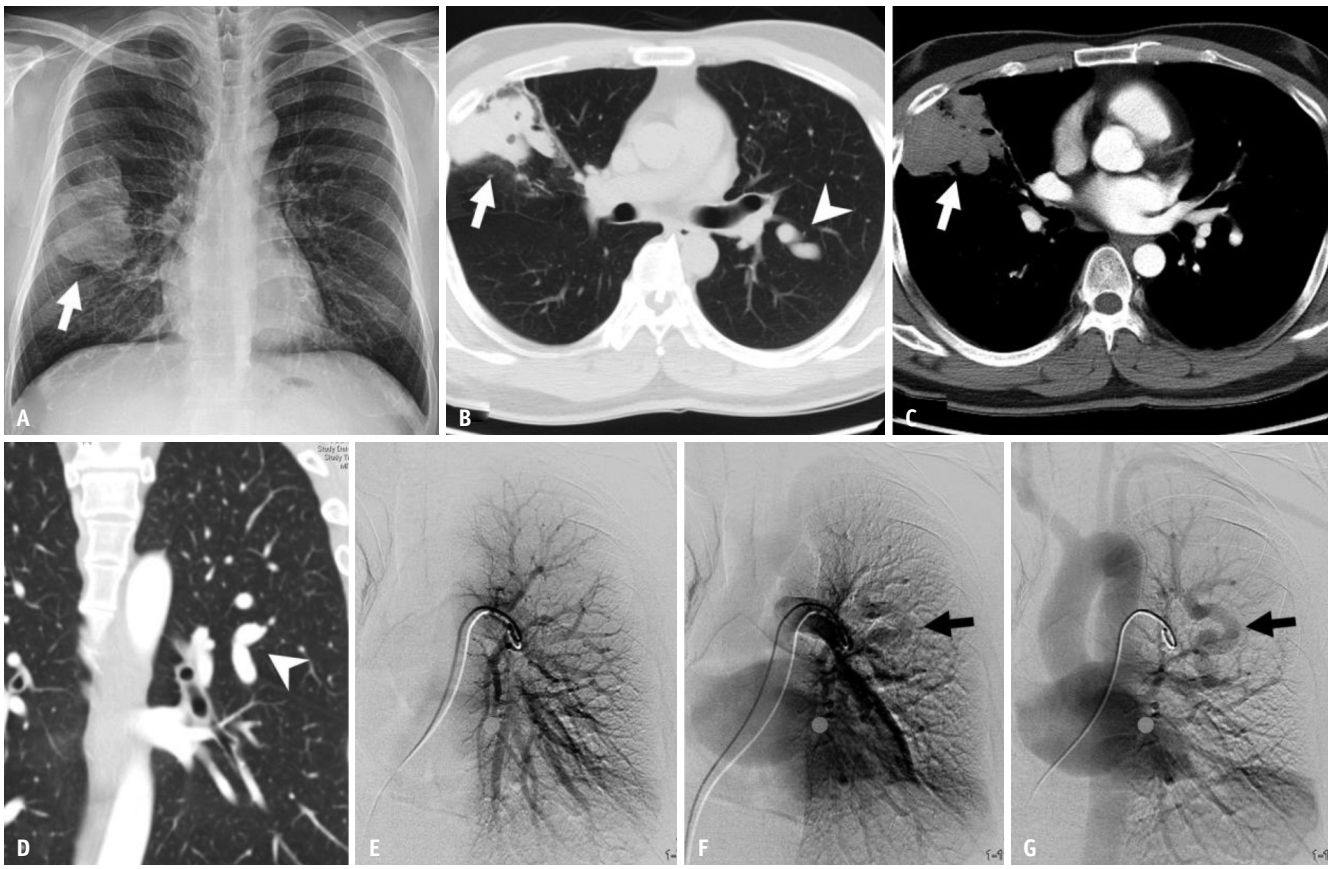


Supplementary Fig. 4. Systemic arterial supply to the lung in a 42-year-old male with exertional dyspnea.
A-D. Serial contrast-enhanced axial CT images with a mediastinal window setting show engorged vascular structures along the pleural surface (arrowheads), connected with branches from hypertrophied left inferior phrenic artery (arrows). **E-H.** Serial contrast-enhanced axial CT images with a lung window setting demonstrate architectural distortion of the lung parenchyma (arrows) and cystic airspaces (arrowheads) around engorged vascular structures. **I, J.** Selective left inferior phrenic angiograms show the hypertrophied left inferior phrenic artery and pulmonary arteries (arrowheads) opacified by the systemic-to-pulmonary artery shunt (arrows).

Supplementary Fig. 5. A 57-year-old female with an anomalous pulmonary vein at the subsegmental level.
A. Coronal CT images demonstrate an abnormal tortuous vascular structure (arrowheads) at the right upper lobe. **B.** The super-selective right pulmonary angiogram shows that parts of the right apical segment drain directly into the right superior pulmonary vein via a dilated tortuous collateral vein (arrows).

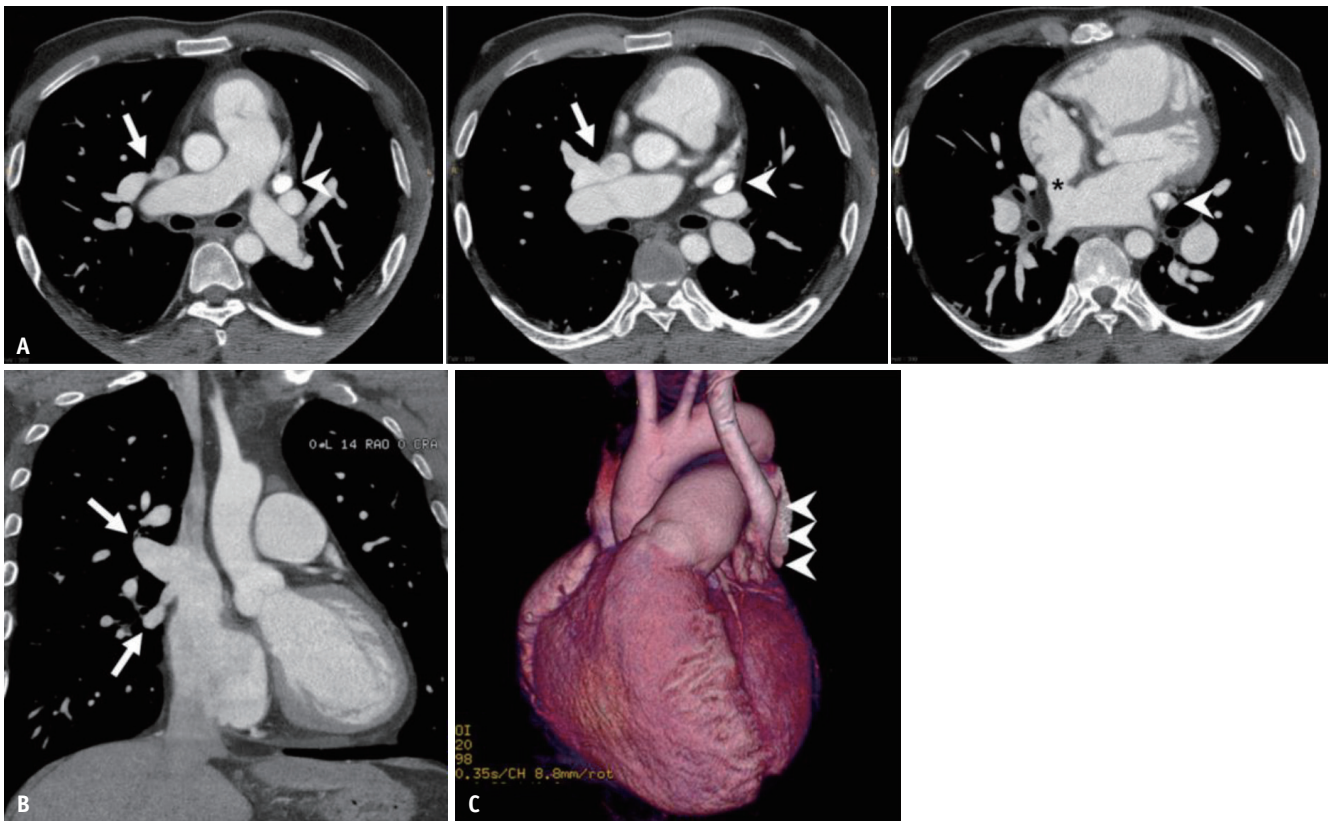


Supplementary Fig. 6. Incidentally found pulmonary vein stenosis in a 52-year-old male.
A. Chest radiograph shows no definite abnormality. **B, C.** Coronal CT and three-dimensional volume-rendering images demonstrate focal tight stenosis (arrows) involving the right superior pulmonary vein.



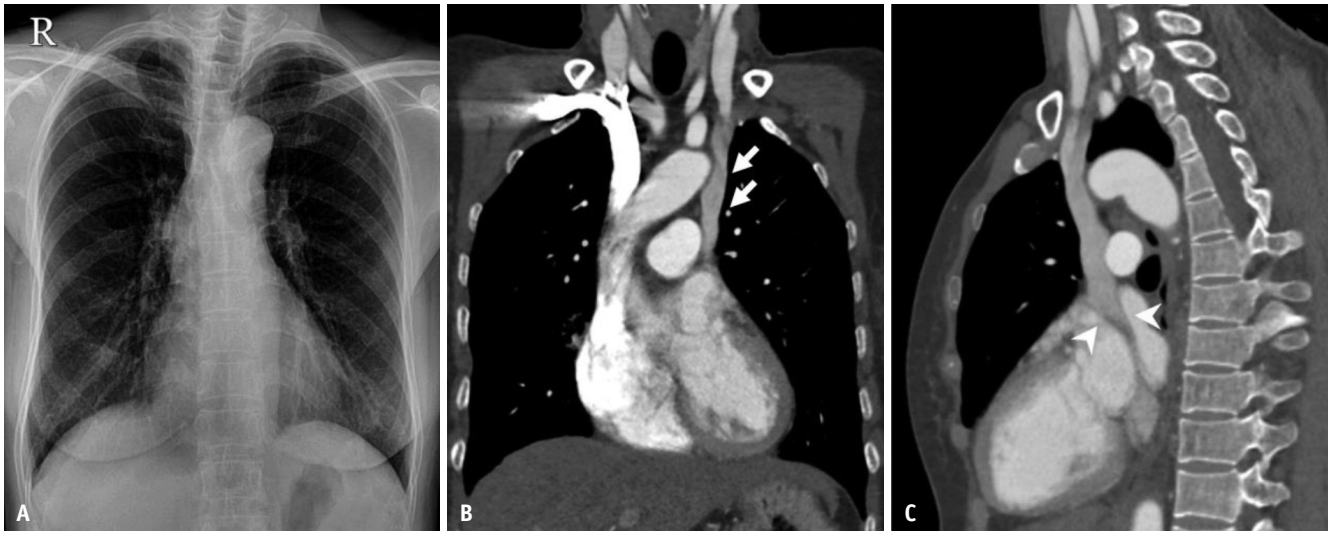
Supplementary Fig. 7. Pulmonary vein varix associated with congenital cystic adenomatoid malformation type 1 in a 39-year-old male.

A. Chest radiograph shows a mass-like increased opacity (arrow) in the right mid-zone. **B-D.** Contrast-enhanced CT images with lung and mediastinal window settings show a mixed solid and cystic mass (arrows) involving the right upper lobe consistent with a cystic adenomatoid malformation. An abnormal tortuous vascular dilatation (arrowheads) is also noted in the left parahilar area. **E.** The selective left pulmonary angiogram shows no evidence of arterial dilatation or arteriovenous fistula. **F, G.** Delayed pulmonary venous return from the left upper lobe drained directly into the proximal superior pulmonary vein via a dilated tortuous collateral vein, representing pulmonary varix (arrows).

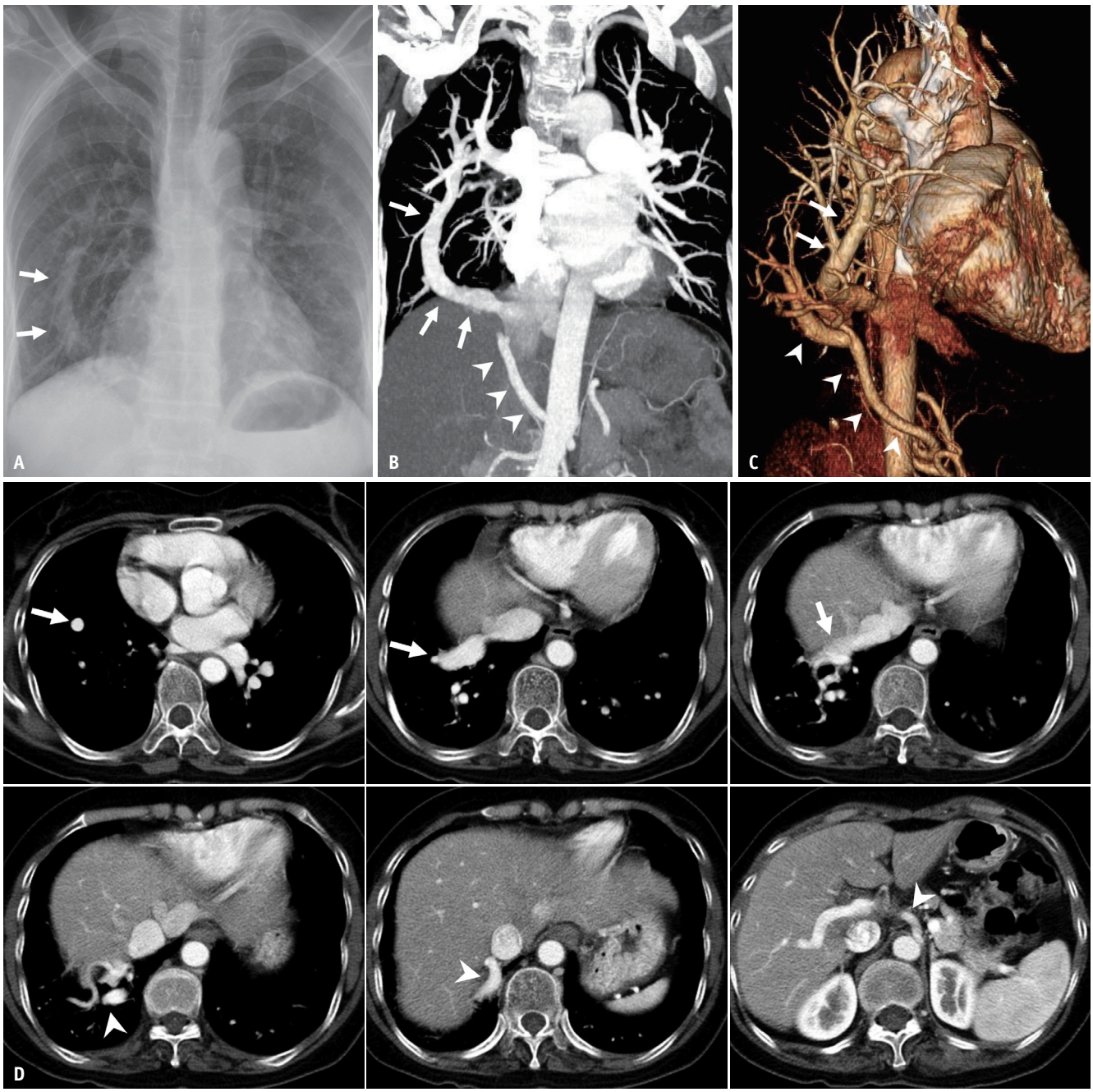


Supplementary Fig. 8. Partial anomalous pulmonary venous drainage involving the right superior pulmonary vein in a 40-year-old male.

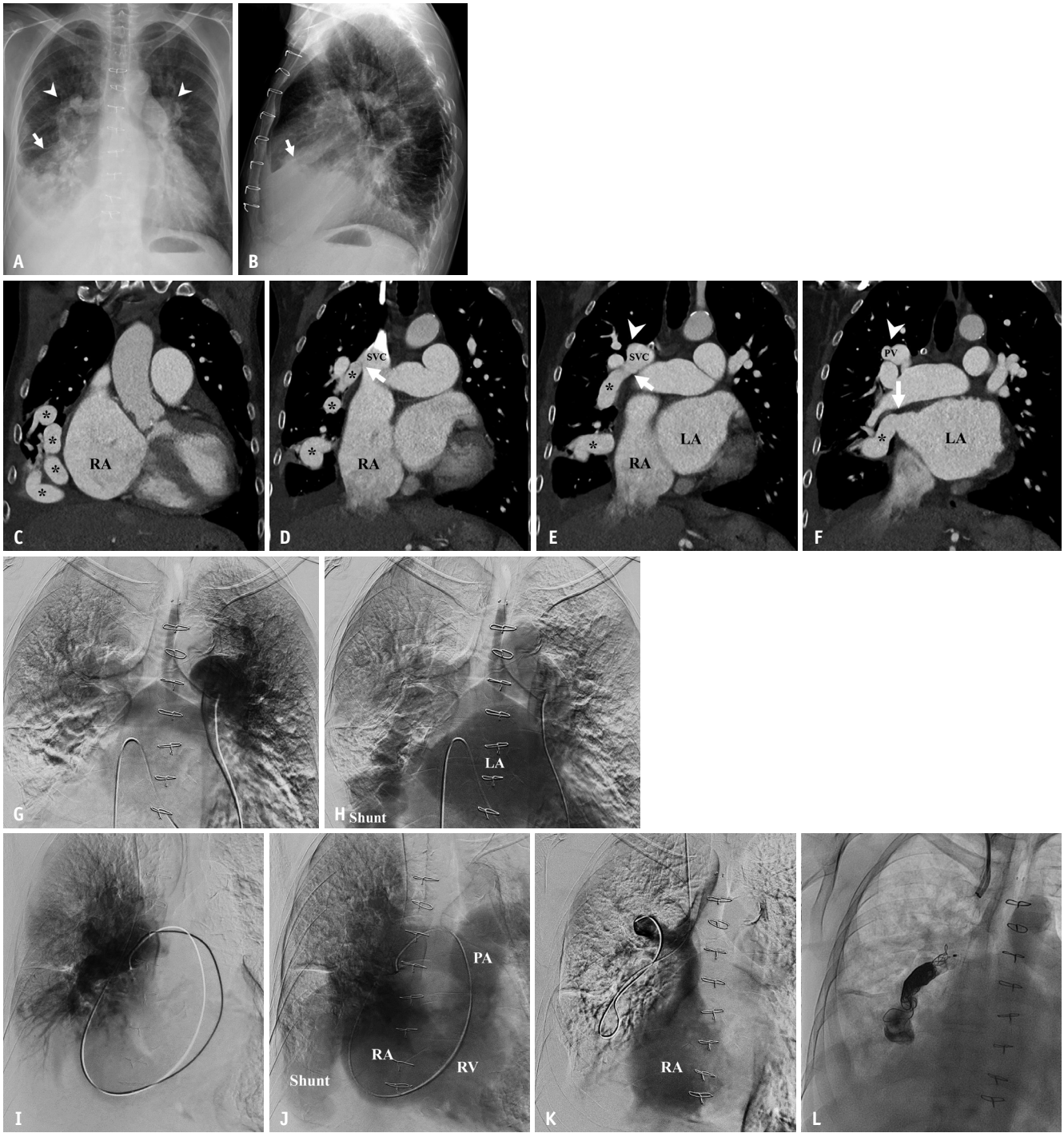
A. Serial axial CT images show that the right superior pulmonary vein drains into the superior vena cava (arrows). The right inferior pulmonary vein has a normal connection with the left atrium. A superior sinus venosus atrial septal defect (*) caused marked right ventricular dilatation. A persistent left superior vena cava (arrowheads) is also noted. **B.** Coronal CT image shows that right superior pulmonary veins (arrows) from the right upper and middle lobes joined the superior vena cava. **C.** Three-dimensional volume-rendering of the persistent left superior vena cava (arrowheads) draining into the coronary sinus.



Supplementary Fig. 9. Left superior vena cava in a 62-year-old female.
A. Chest radiograph shows no definite abnormality. **B, C.** The reformatted coronal and oblique sagittal CT images show left superior vena cava (arrows) coursing inferiorly to the left of the aortic arch and draining into the coronary sinus (arrowheads).



Supplementary Fig. 10. Scimitar syndrome in a 56-year-old female.
A. Chest radiographs show a curvilinear vascular marking (arrows) adjacent to the right heart border, as well as a reduced volume in the right lung field. **B, C.** Coronal CT image and volume-rendered image demonstrate the anomalous curvilinear pulmonary vein (arrows) draining into the infra-diaphragmatic inferior vena cava. Aberrant systemic arterial supply (arrowheads) from the abdominal aorta to the right lung is noted. **D.** Serial axial CT images show a tangled vascular structure in the right lower lobe consisting of an anomalous pulmonary vein (arrows) and aberrant systemic artery (arrowheads).



Supplementary Fig. 11. Partial anomalous pulmonary venous drainage in a 59-year-old male who underwent mitral and aortic valve repair eleven years earlier.

A, B. Posteroanterior and left lateral chest radiographs show markedly dilated central pulmonary arteries (arrowheads), cardiomegaly, and right pleural effusion. A large area of increased opacities (arrows) obscuring the right heart border and diaphragm is also noted. **C-F.** Serial coronal CT images show the right superior PV draining into the SVC (arrowheads), consistent with partial anomalous pulmonary venous drainage. There is a large tortuous abnormal vascular structure (*) connecting the LA and the SVC (arrows), resulting in another left-to-right shunt. **G, H.** The left common pulmonary angiogram demonstrates delayed opacification of the LA and shunt vessel. **I, J.** The right common pulmonary angiogram reveals pulmonary venous drainage into the SVC and subsequent recirculation into the pulmonary trunk. **K, L.** After catheterization of the shunt vessel, coil and glue embolization was performed to reduce the left-to-right shunt. LA = left atrium, PA = pulmonary artery, PV = pulmonary vein, RA = right atrium, RV = right ventricle, SVC = superior vena cava