

Partial Anomalous Pulmonary Venous Connection in Adults: Evaluation with MDCT

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1. Educational Presentation

Educational Objective

The educational objectives of this exhibit are to present the CT appearance of partial anomalous pulmonary venous connection (PAPVC) in adults and discuss related issues assessable with multidetector CT (MDCT).

Disclosure

The authors of this presentation do not have any affiliation with the manufacturer of any commercial product or services.

Background

- Partial anomalous pulmonary venous connection is a congenital abnormality in which one or more of the pulmonary veins connect to a systemic vein or directly to the right atrium potentially producing a left to right shunt.
- The most common forms encountered in adults are PAPVC of the left upper lobe pulmonary vein to the left brachiocephalic vein and PAPVC of the right upper lobe pulmonary vein to the superior vena cava (SVC).
- In this exhibit we show the appearances of various forms of PAPVC and discuss issues regarding the anomaly evaluable by MDCT.

Presentation Overview

- Discuss difficulty in discerning pulmonary venous connections to the SVC and strategies to improve visualization of these connections.
- Association of PAPVC with sinus venosus atrial septal defect.
- Evaluation of the status of the remainder of the pulmonary venous circulation, as this determines the extent of left to right shunting.
- Evaluation of the flow direction in the anomalous vessel. Under certain circumstances, flow in the anomalous pulmonary vein can be away from the systemic venous connection producing a right to left shunt. We demonstrate how flow direction can be determined on MDCT.
- Complications secondary to PAPVC after surgery on the contralateral lung.
- Post-operative appearance and complications after surgical repair of PAPVC.
- We illustrate the use of multiplanar image reformatting and volume rendering to best depict the imaging features of PAPVC.

PAPVC: DEFINITION AND INCIDENCE

Partial anomalous pulmonary venous connection (PAPVC) is present when one or more, but not all, of the pulmonary veins connect to a systemic vein, the right atrium, or the coronary sinus.

Left Lung Veins connect to:

- 1. left innominate vein
- 2. coronary sinus
- 3. hemiazygous vein
- 4. anomalous vein that drains into the innominate vein.

Right Lung Veins connect to:

- 1. Superior/inferior vena cava
- 2. azygous vein
- 3. right atrium
- 4. hepatic vein

Less common variations:

- Absence of the coronary sinus with pulmonary veins connecting to multiple systemic sites or to the left atrium
- Connection to the portal vein

Incidence: 0.4-0.7% with higher rates in females.

PAPVC OF THE LEFT UPPER LOBE PULMONARY VEIN

- On CT, this is seen as a vertically oriented anomalous vein to the left of the aortic arch.
- This is often mistaken for persistence of the left SVC.
- PAPVC of the left superior pulmonary vein drains to the left brachiocephalic vein.

Partial Anomalous Pulmonary Venous Connection (PAPVC) of the Left Upper Lobe Pulmonary Vein



35-year-old man with PAPVC of the left superior pulmonary vein. CT demonstrates anomalous vessel (arrow) adjacent to the aortic arch.

Partial Anomalous Pulmonary Venous Connection (PAPVC) of the Left Upper Lobe Pulmonary Vein

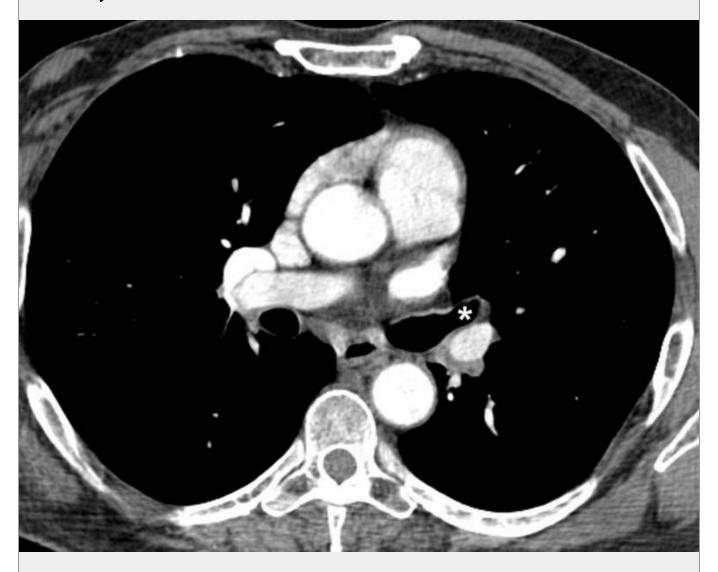
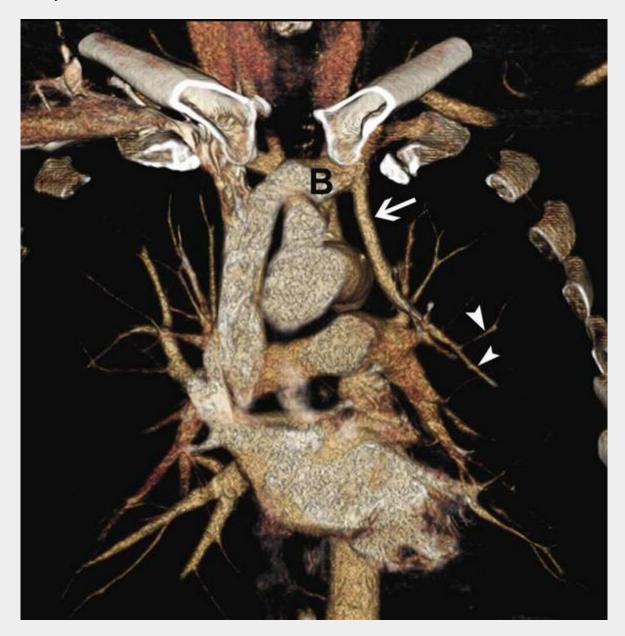


Image at a lower level demonstrates that the left superior pulmonary vein is not seen in its normal location, anterior to the left upper lobe bronchus (asterisk).

Partial Anomalous Pulmonary Venous Connection (PAPVC) of the Left Upper Lobe Pulmonary Vein

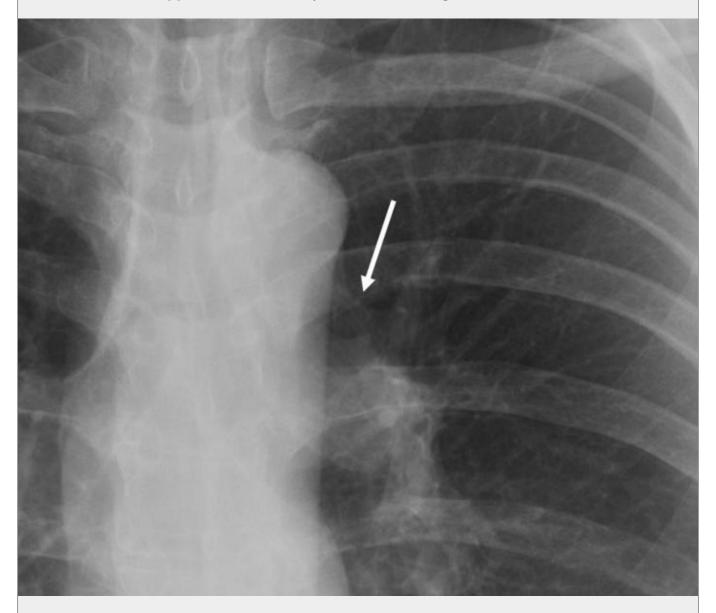


Volume rendered view from anterior perspective shows anomalous left superior pulmonary vein (arrow) joining with left brachiocephalic vein (B). Arrowheads = tributaries of left upper lobe pulmonary veins.

Clinical Implications:

- Catheter placements
- CHF from left heart failure spares the left upper lobe
- Right heart failure results in Left upper lobe pulmonary edema
- Obstruction of left brachiocephalic vein can result in right to left shunt (unoxygenated blood from left subclavian vein drains to anomalous left superior pulmonary vein to collateral channels to pulmonary veins to left atrium).[Levy JM, J Vasc Interv Radiol 2002; 13:423-425.]
- Right lung disease or right pneumonectomy results in significant left to right shunt.[Black MD. Ann Thor Surg 1992;53:689-91]

PAPVC of the Left Upper Lobe Pulmonary Vein: CXR Findings



PAPVC (arrow) of the Left Upper Lobe Pulmonary Vein on a chest radiology.

PAPVC of the Left Upper Lobe Pulmonary Vein



Volume rendering of PAPVC of the Left Upper Lobe Pulmonary Vein (arrowhead).

PAPVC Connection in Adults



PAPVC OF THE LEFT UPPER LOBE VS. PERSISTANT LEFT SUPERIOR VENA CAVA

Persistent Left SVC:

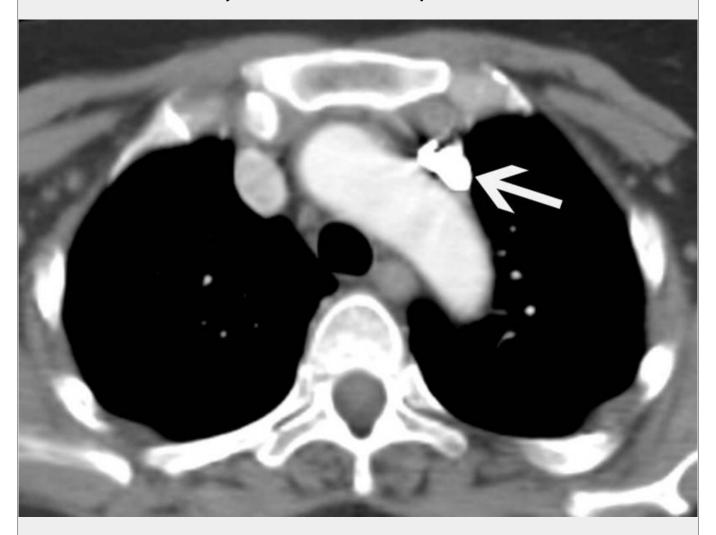
- Most common venous anomaly of the thorax.
- Incidence 0.5 -2.0 % in normal individuals and up to 10% with congenital heart disease.
- Results from failure of embryologic regression of a portion of the left common and anterior cardinal veins.
- Drains the left jugular and left subclavian vein and courses inferiorly anterior to the left hilum to drain into a dilated coronary sinus.
- Right superior vena cava may or may not be present, and may be small if present.
- In some cases, persistent left superior vena cava can connect to the left atrium producing a right to left shunt.

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



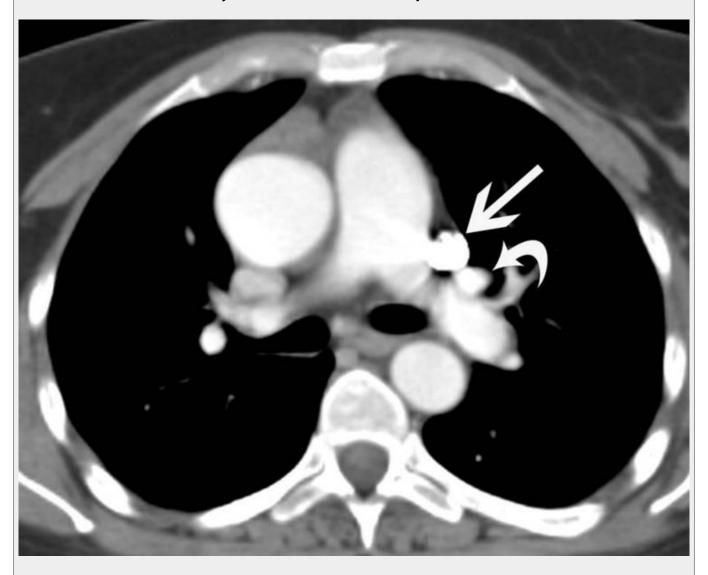
Chest radiograph shows a vascular catheter coursing along the left mediastinum in a persistent left superior vena cava.

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



CT shows persistent left superior vena cava (arrow) anterior to the aortic arch.

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



CT shows persistent left superior vena cava (straight arrow) anterior to left superior pulmonary vein (curved arrow).

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



Reformatted coronal MIP image shows persistent left superior vena cava (white arrow) draining to coronary sinus (black arrow).

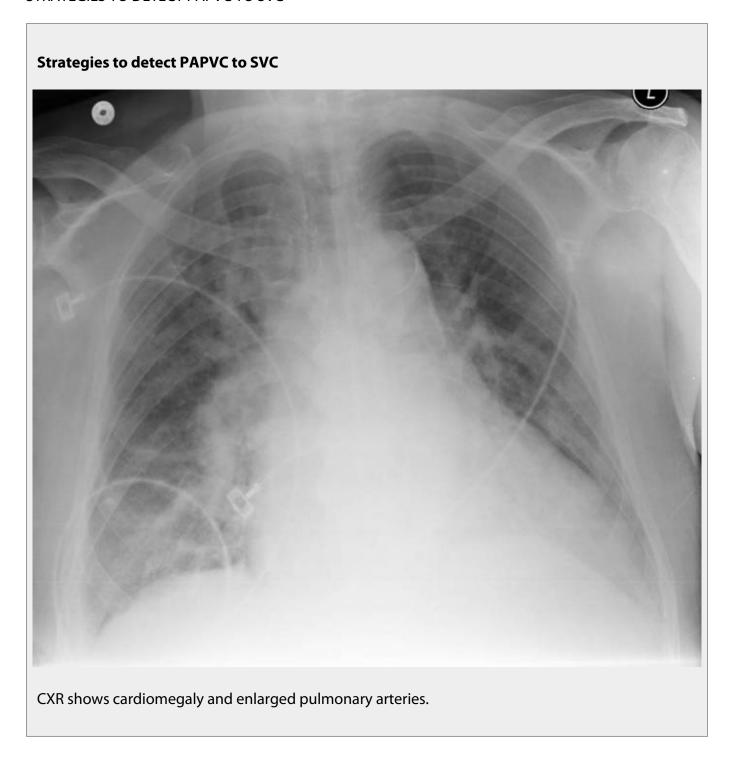
PAPVC - Left Upper Lobe Pulmonary Vein Left brachiocephalic vein is normal or enlarged No vessels anterior to the left main bronchus Left upper lobe pulmonary Veins enter aberrant vessel Normal Coronary Sinus Left upper Lobe Pulmonary Left brachiocephalic vein is small or absent 2 vessels anterior to Left main bronchus (Left superior pulmonary vein and left SVC) Left superior pulmonary vein has normal course Coronary sinus is enlarged

PAPVC - SHUNTING

With isolated PAPVC, CXR is usually normal since shunt is less than 2:1. With larger shunts findings are similar to ASD:

- pulmonary overcirculation
- enlarged RA and RV
- enlarged main and hilar pulmonary arteries with small aorta
- enlarged systemic vein at site of connection (SVC, azygos vein)
- need to see the course of the abnormal PV in the lung to make the diagnosis on CXR

STRATEGIES TO DETECT PAPVC TO SVC



Abrupt change in caliber of SVC:

Strategies to detect PAPVC to SVC: Abrupt change in caliber of SVC



CT shows dilated RA and RV, change in caliber SVC, multiple site of communication between pulmonary veins and SVC (arrows). Often with PAPVC to SVC there is more than one site of abnormal connection.

Evaluate SVC in wide windows in addition to soft tissue windows:

Strategies to detect PAPVC to SVC

Normal windows: width = 450 level = 50

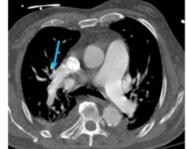


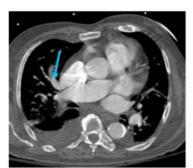




Wide windows: Width = 800 level = 100







Note that connections of anamalous vein to SVC (arrows) are more discernable with wider windows.

PAPVC AND ATRIAL SEPTAL DEFECTS

- Partial anomalous pulmonary venous connection (PAPVC) is associated with atrial septal defects (ASD).
- 15% of secundum ASD's and 85 100% of sinus venosus ASD's are associated with PAPVC.

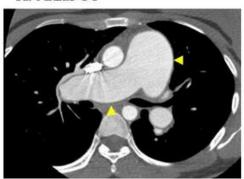
PAPVC and ASD



CXR shows enlarged pulmonary arteries and right ventricular enlargement on lateral view.

PAPVC and ASD

A. Axial CT



B. Coronal Oblique CT



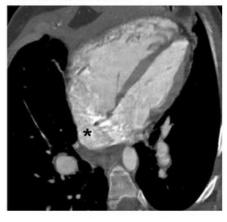
A. Enlarged PA (arrowhead).

B & C. Large sinus venosus ASD (asterisk)

D. Multiple sites of PAPVC (arrows)

Note: large ASD allows small amount of right to left shunting (contrast passes into LA through ASD)

C. Transverse CT



D. Volume Rendered Coronal



PAPVC and ASD

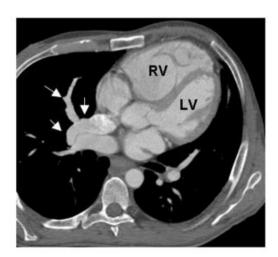
PAPVC and Atrial Septal Defects

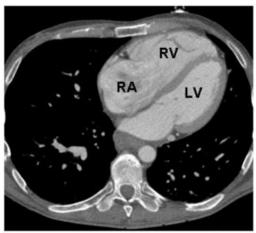
Small RUL pulmonary vein draining to SVC (arrows)





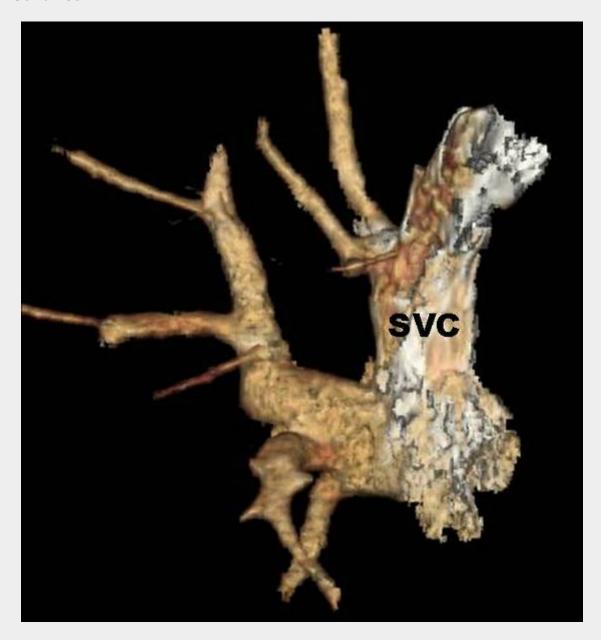
Large confluence of pulmonary veins draining to inferior aspect of SVC (arrowheads)





Small sinus venosus ASD with dilated RA and RV

PAPVC and ASD



Volume rendering demonstrates extensive connections of pulmonary veins from the right lung to the superior vena cava (SVC).

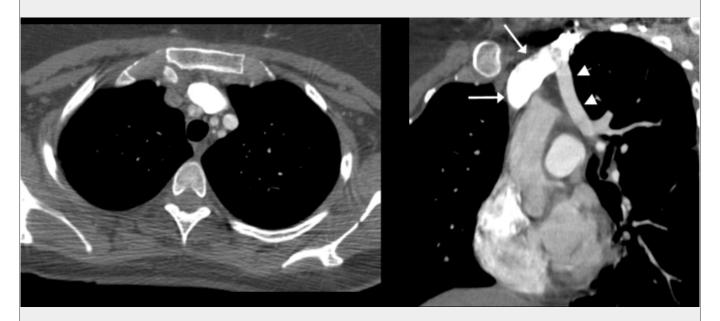
PAPVC FLOW DIRECTION

- Flow direction in anomalous pulmonary vein is usually towards systemic veins producing a left to right shunt.
- With PAPVC to SVC, the attenuation of the contrast enhanced anomalous veins is the same as the remainder of the pulmonary venous system and significantly less than that of the SVC. This indicates that flow in the anomalous vein is towards the SVC.



Note that attenuation of the anomalous pulmonary veins is less than that of the SVC (arrows).

• With PAPVC of left upper lobe pulmonary vein, flow direction in the anomalous vessel is apparent if contrast in injected into the left arm. The attenuation of the left brachiocephalic vein (LBCV) is very high due to undiluted contrast while the attenuation of the anomalous vessel near its junction with the brachiocephalic vein is significantly lower. A low attenuation flow artifact may even be present in the LBCV due to inflow from the anomalous pulmonary vein.

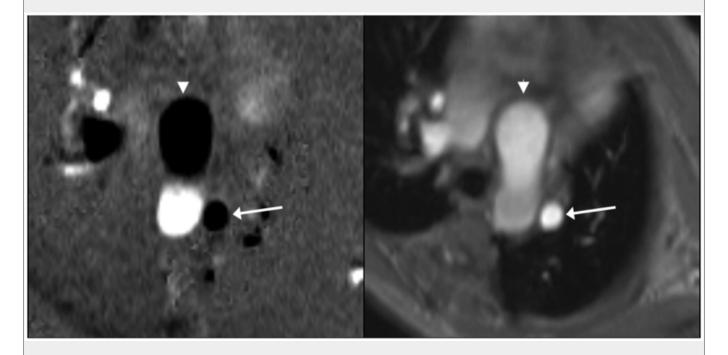


With PAPVC of left upper lobe pulmonary vein, flow direction in the anomalous vessel is apparent if contrast in injected into the left arm. The attenuation of the left brachiocephalic vein (LBCV) [arrows] is very high due to undiluted contrast while the attenuation of the anomalous vessel [arrowheads] near its junction with the brachiocephalic vein is significantly lower. A low attenuation flow artifact may even be present in the LBCV due to inflow from the anomalous pulmonary vein.

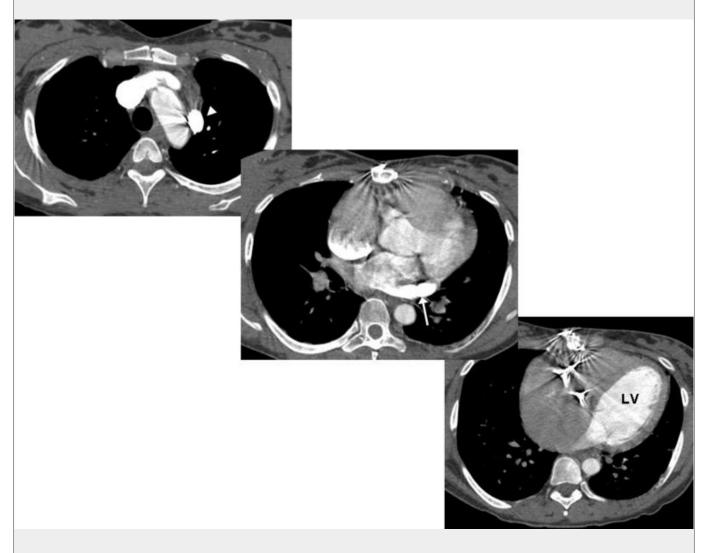
• In some cases flow can be retrograde in the anomalous pulmonary vein away from the LBCV.



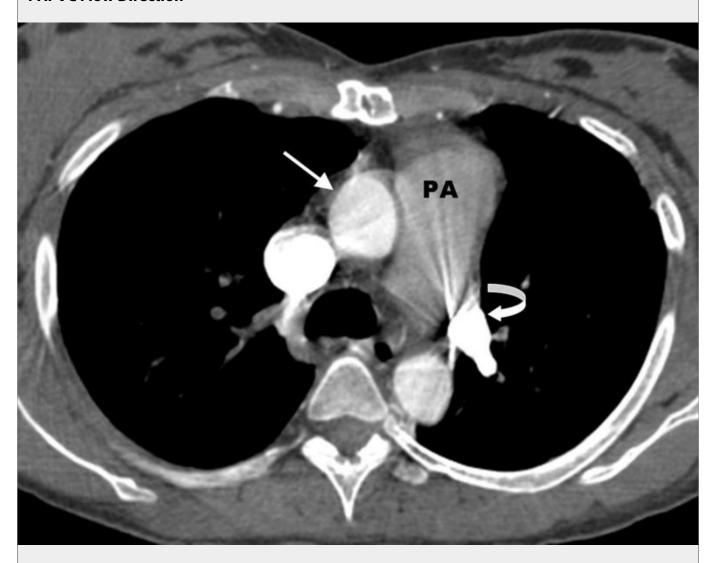
CT in patient with heart failure from cardiomyopathy shows PAPVC of left upper lobe pulmonary veins (LUL PV) [arrows] to the left brachiocephalic vein (LBCV) [arrowhead]. Images were obtained early after injection of IV contrast as study was performed to exclude pulmonary embolism. There is opacification of anomalous LUL PV before contrast has reached the remainder of the pulmonary veins and left sided cardiac chambers. (Prolonged circulation time from heart failure likely also contributes to lack of contrast in normal pulmonary veins.) Contrast must have entered anomalous pulmonary vein via retrograde flow from LBCV. We speculate that elevated systemic venous pressure from patient's cardiomyopathy also contributes to retrograde flow in anomalous pulmonary vein.



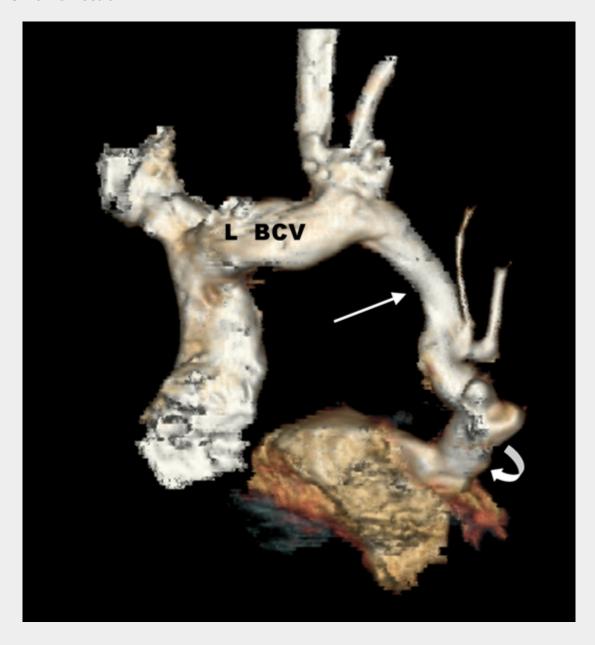
Oblique axial phase contrast MRI study on same patient performed after stabilization of heart failure shows direction of flow in anomalous vein [arrow] has normalized to cephalad, towards LBCV, in same direction as flow in ascending aorta [arrowhead] (phase image on left with magnitude image on right).



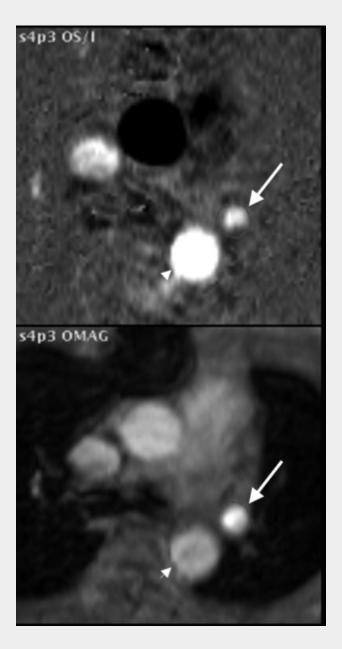
CT scan in patient with history of tricuspid valve replacement for tricuspid valve endocarditis shows PAPVC [arrowhead] that forms conduit from LBCV to left inferior pulmonary vein [arrow]. Flow direction in anomalous vessel is away from LBCV causing a right to left shunt. We speculate that elevated systemic venous pressure from right heart disease is significantly contributory to this phenomenon. Note opacification of left ventricle before opacification of the right cardiac chambers.



Axial image at level of main pulmonary artery shows early opacification of aorta (straight arrow) before opacification of pulmonary artery (PA) due to flow of contrast from LBCV to left atrium. Note the dense contrast in the anamalous pulmonary vein (curved arrow).



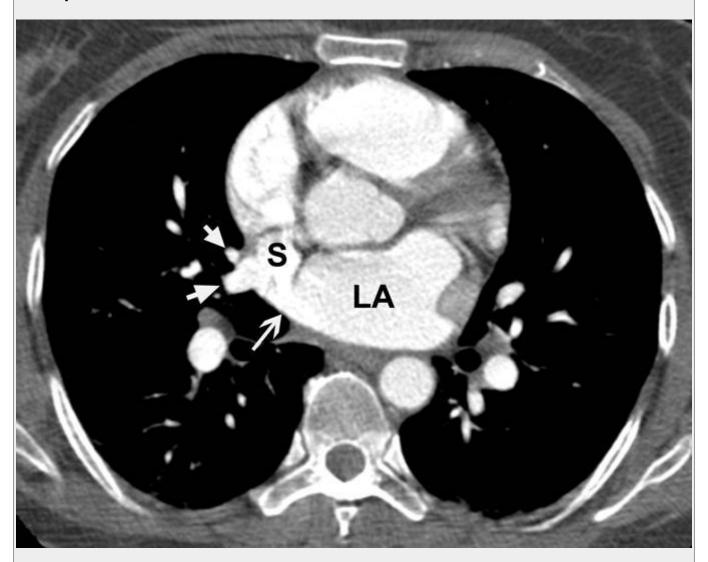
Volume rendered view from the anterior prospective demonstrates the left brachiocephalic vein (L BCV) to anomalous pulmonary vein connection (arrow).



Phase contrast image from MRI study confirms direction of flow in anomalous vessel [arrows] is caudad away from LBCV, same direction as flow in descending thoracic aorta [arrowheads] (phase contrast image on top with magnitude image on bottom).

PAPVC SURGICAL CORRECTION

Cardiac gated CT on 25 year-old woman with sinus venosus atrial septal defect and multiple anomalous pulmonary veins in the right lung draining to the superior vena cava.



CT shows sinus venosus atrial septal defect (long arrow) allowing communication of the left atrium (LA) and superior vena cava (S). Anomalous pulmonary veins (short arrows) are also seen draining to the superior vena cava.



At a slightly higher level additional anomalous pulmonary veins (short arrows) are seen draining to the superior vena cava (long arrow).



CT shows persistent left superior vena cava (arrow).

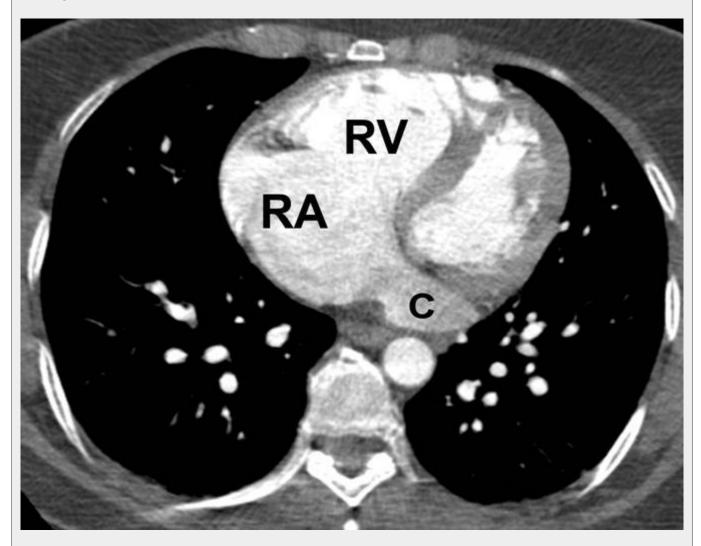
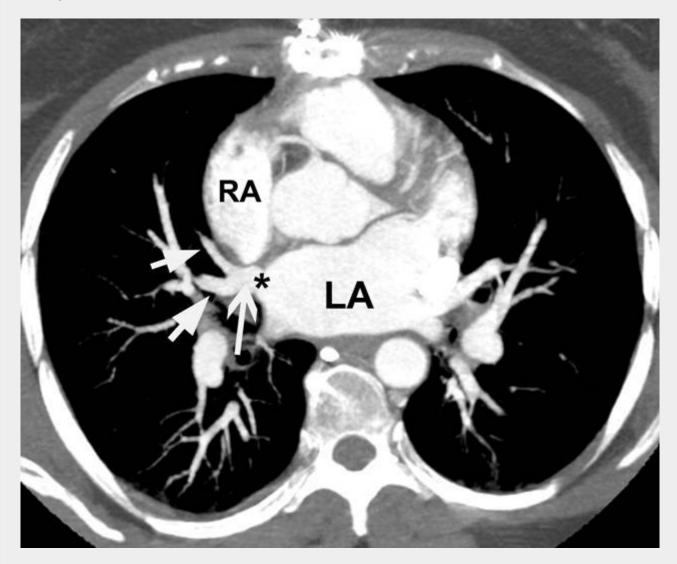
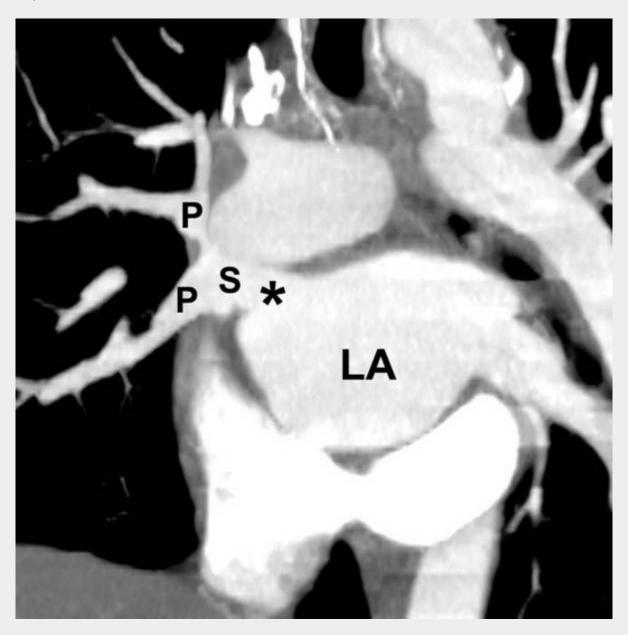


Image through cardiac chambers shows dilated right atrium (RA) and right ventricle (RV) consistent with left to right shunt. Coronary sinus (C) is also dilated due to persistent left superior vena cava.

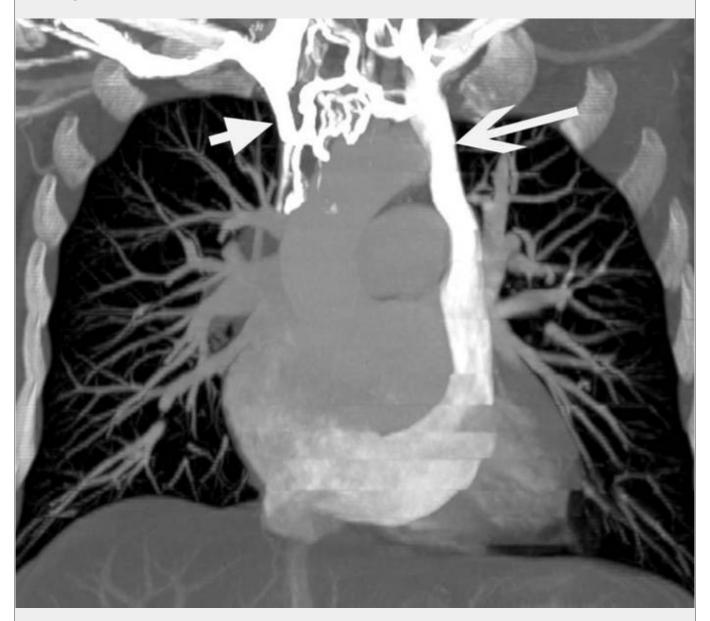
At surgery the right superior vena cava was ligated above the junction with the most superior anomalous right pulmonary vein and the communication between the inferior superior vena cava and the right atrium was closed. Thus blood return from the anomalous pulmonary veins now drains to the inferior portion of the right superior vena cava and to the right atrium via the sinus venosus atrial septal defect.



Post-operative CT scan shows anomalous pulmonary veins (short arrows) draining to inferior portion of superior vena cava (long arrow) to left atrium (LA) via sinus venosus atrial septal defect (asterisk).

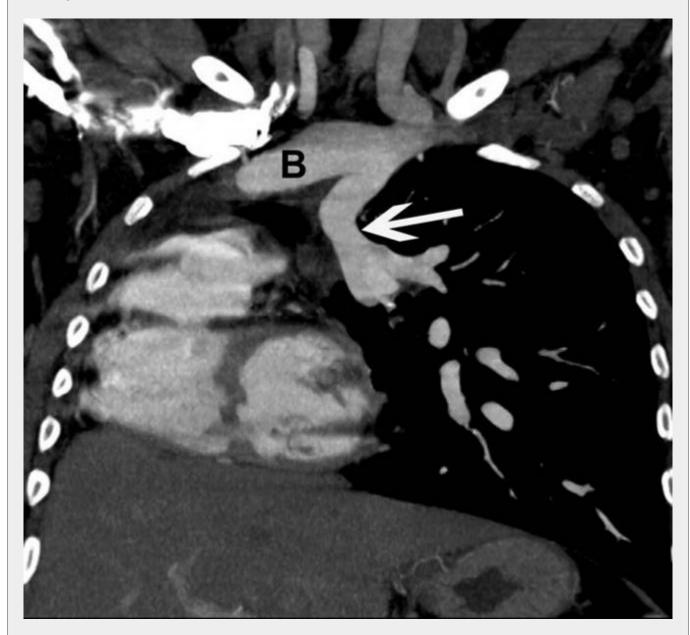


Reformatted coronal CT image shows anomalous pulmonary veins (P) draining to inferior segment of superior vena cava (S) to left atrium (LA) via sinus venosus atrial septal defect (asterisk).

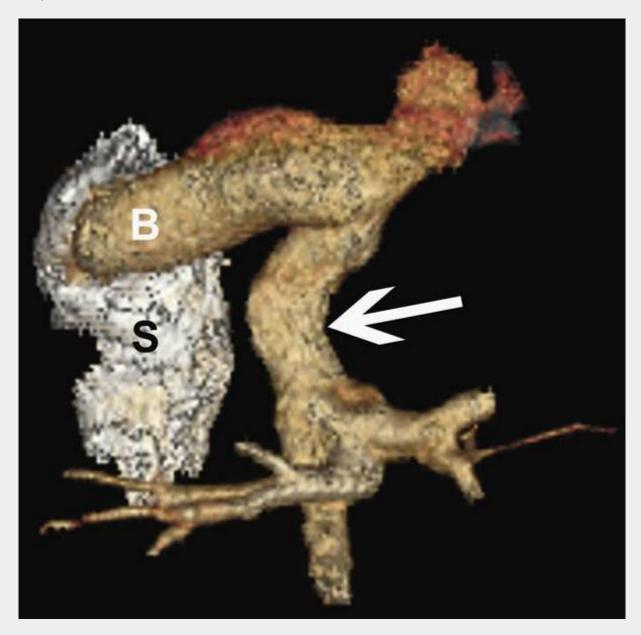


Reformatted coronal MIP image shows blood flow pattern above ligated portion of right superior vena cava. Superior portion of right superior vena cava (short arrow) communicates with left superior vena cava (long arrow) via collateral mediastinal veins.

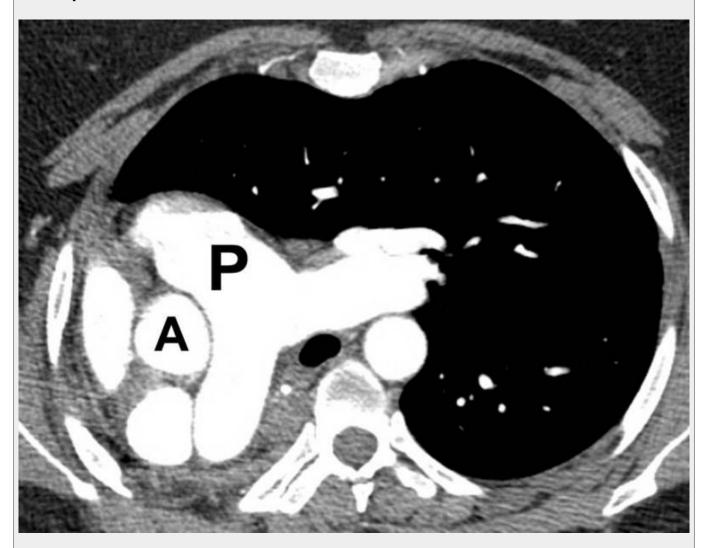
55-year-old woman with PAPVC of left upper lobe pulmonary vein who had right pneumonectomy for lung cancer:



Coronal reformatted MIP image shows anomalous left upper lobe pulmonary vein (arrow) draining to left brachiocephalic vein (B).



Volume rendered view of vasculature from anterior perspective shows anomalous left upper lobe pulmonary vein (arrow) draining to left brachiocephalic vein (B). S = superior vena cava.



CT Image shows that main pulmonary artery (P) is larger in diameter than aorta (A) consistent with increased pulmonary blood flow.

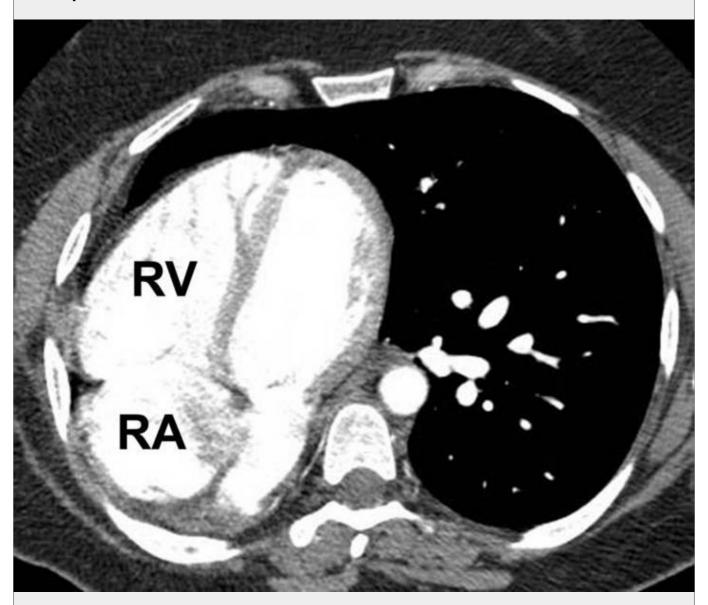
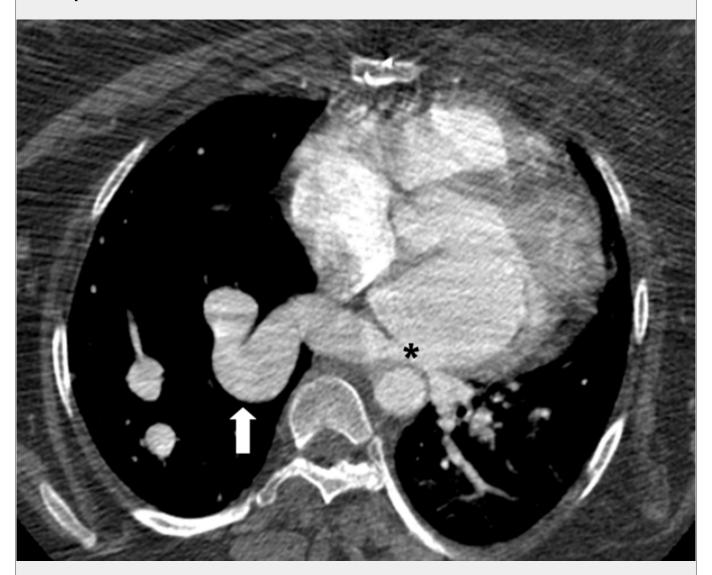


Image of heart shows that right atrium (RA) and right ventricle (RV) are dilated compared to left-sided chambers consistent with significant left to right shunt. After right pneumonectomy, a larger percentage of pulmonary venous blood shunts to the right circulation through the anomalous left superior pulmonary vein.



Patient with history of PAPVC and repair with anastamosis of anomalous vein to left atrium. Patient developed stenosis at junction [asterisk] of pulmonary vein and left atrium resulting in a large pulmonary varix [arrow].



Volume rendered from anterior perspective shows large pulmonary varix extending throughout right lung [arrows].

2. Conclusions

After reviewing this exhibit, the participant should be familiar with the MDCT evaluation of partial anomalous pulmonary venous connection in adults.

3. References

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4. Personal Information

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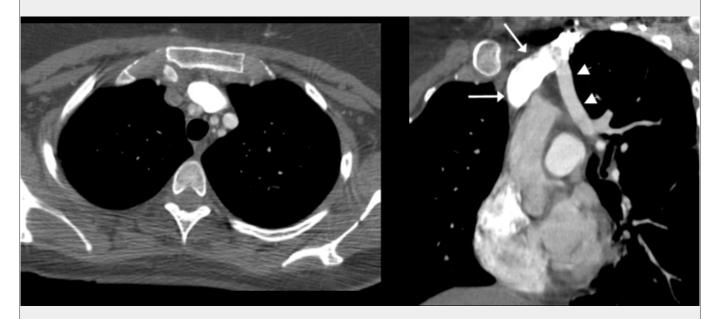
5. Mediafiles

PAPVC Connection in Adults

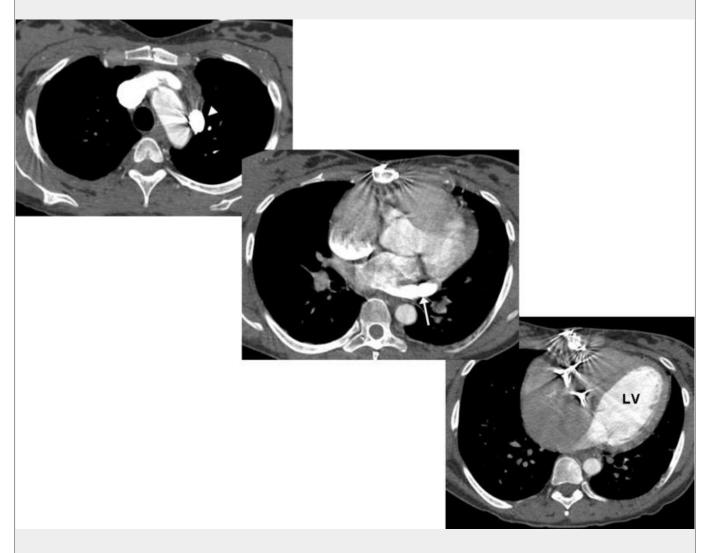




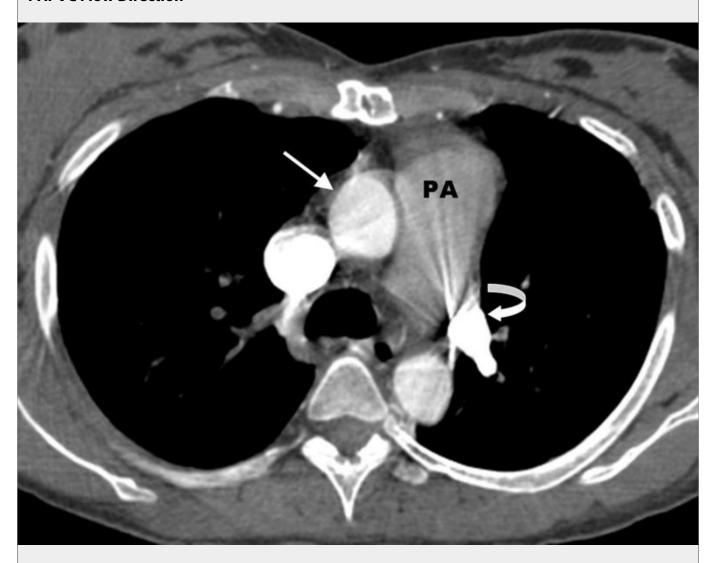
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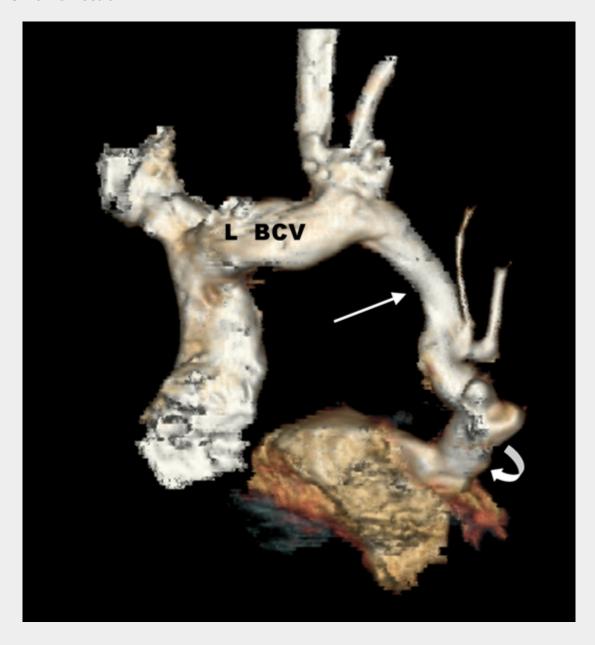
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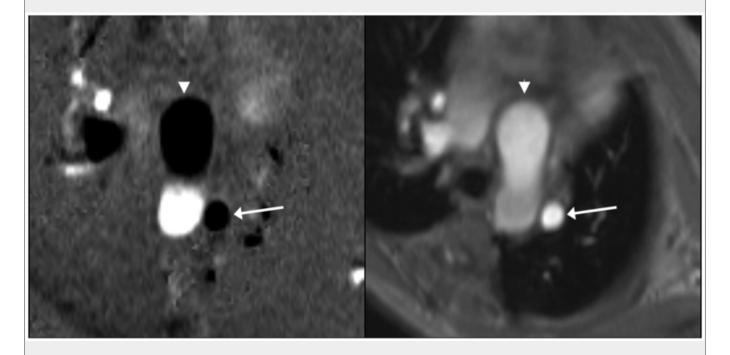
CT scan in patient with history of tricuspid valve replacement for tricuspid valve endocarditis shows PAPVC [arrowhead] that forms conduit from LBCV to left inferior pulmonary vein [arrow]. Flow direction in anomalous vessel is away from LBCV causing a right to left shunt. We speculate that elevated systemic venous pressure from right heart disease is significantly contributory to this phenomenon. Note opacification of left ventricle before opacification of the right cardiac chambers.



Axial image at level of main pulmonary artery shows early opacification of aorta (straight arrow) before opacification of pulmonary artery (PA) due to flow of contrast from LBCV to left atrium. Note the dense contrast in the anamalous pulmonary vein (curved arrow).



Volume rendered view from the anterior prospective demonstrates the left brachiocephalic vein (L BCV) to anomalous pulmonary vein connection (arrow).

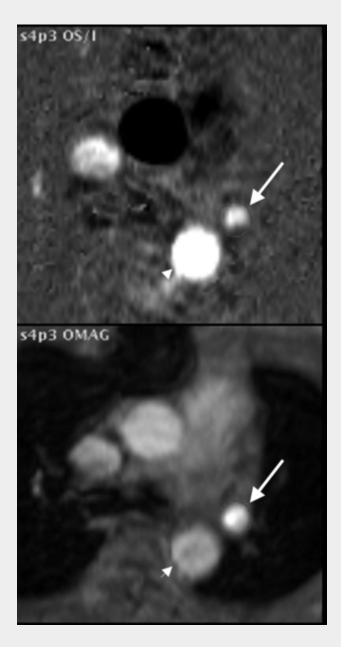


Oblique axial phase contrast MRI study on same patient performed after stabilization of heart failure shows direction of flow in anomalous vein [arrow] has normalized to cephalad, towards LBCV, in same direction as flow in ascending aorta [arrowhead] (phase image on left with magnitude image on right).

PAPVC Flow Direction

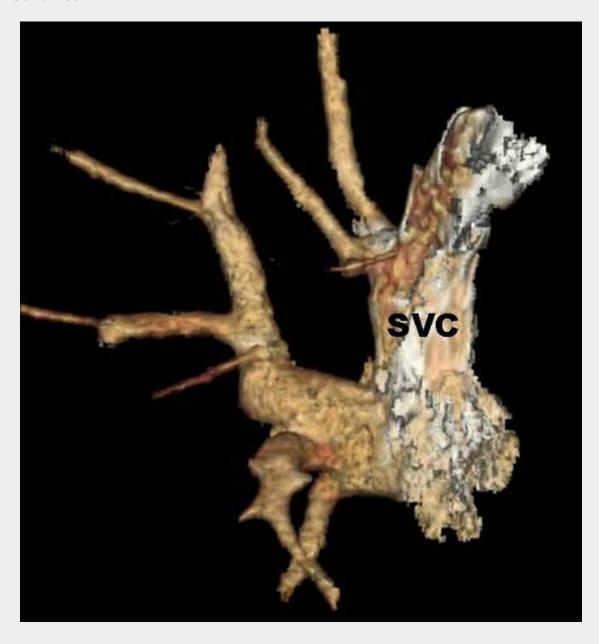


Note that attenuation of the anomalous pulmonary veins is less than that of the SVC (arrows).



Phase contrast image from MRI study confirms direction of flow in anomalous vessel [arrows] is caudad away from LBCV, same direction as flow in descending thoracic aorta [arrowheads] (phase contrast image on top with magnitude image on bottom).

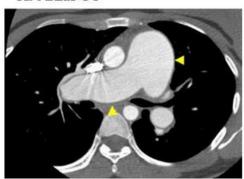
PAPVC and ASD



Volume rendering demonstrates extensive connections of pulmonary veins from the right lung to the superior vena cava (SVC).

PAPVC and ASD

A. Axial CT



B. Coronal Oblique CT



A. Enlarged PA (arrowhead).

B & C. Large sinus venosus ASD (asterisk)

D. Multiple sites of PAPVC (arrows)

Note: large ASD allows small amount of right to left shunting (contrast passes into LA through ASD)

C. Transverse CT



D. Volume Rendered Coronal



PAPVC and ASD

PAPVC and ASD



CXR shows enlarged pulmonary arteries and right ventricular enlargement on lateral view.

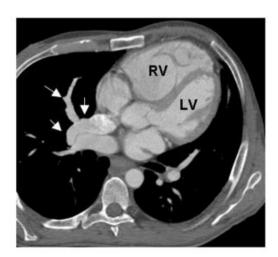
PAPVC and Atrial Septal Defects

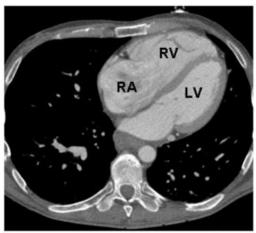
Small RUL pulmonary vein draining to SVC (arrows)





Large confluence of pulmonary veins draining to inferior aspect of SVC (arrowheads)





Small sinus venosus ASD with dilated RA and RV

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



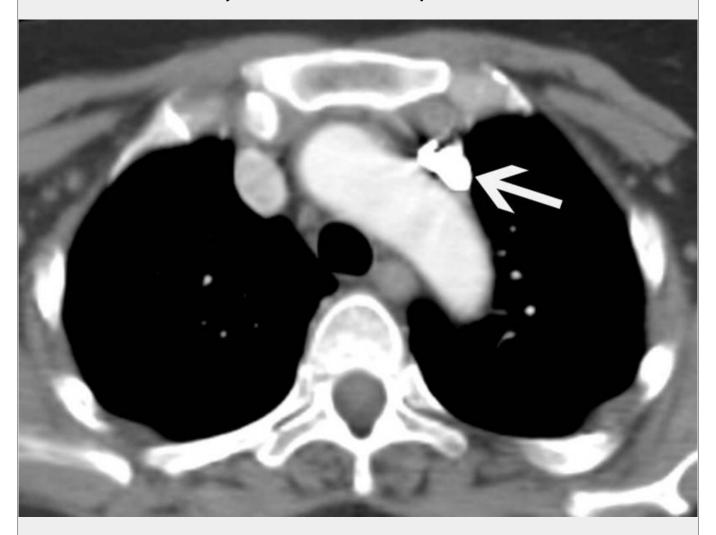
Chest radiograph shows a vascular catheter coursing along the left mediastinum in a persistent left superior vena cava.

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



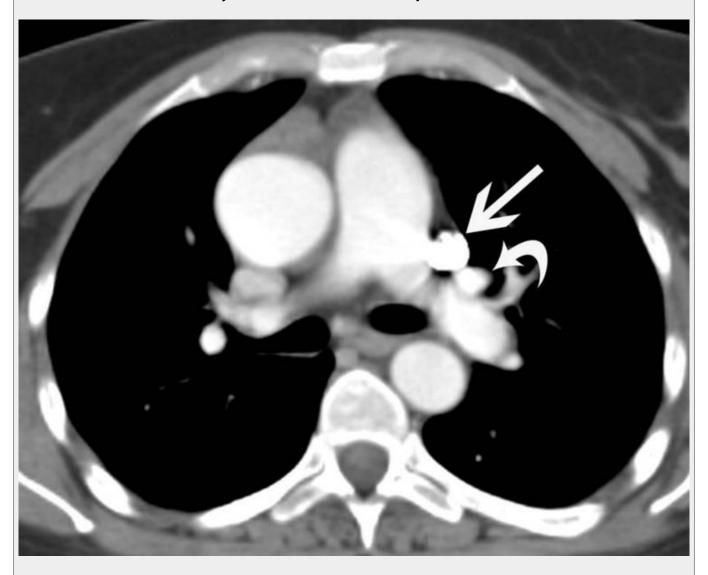
Reformatted coronal MIP image shows persistent left superior vena cava (white arrow) draining to coronary sinus (black arrow).

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



CT shows persistent left superior vena cava (arrow) anterior to the aortic arch.

PAPVC of the LUL Pulmonary Vein vs. Persistent Left Superior Vena Cava



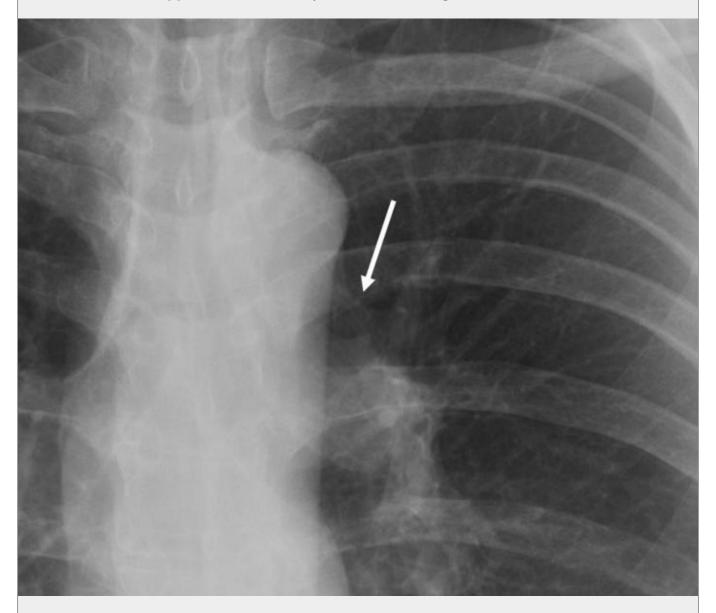
CT shows persistent left superior vena cava (straight arrow) anterior to left superior pulmonary vein (curved arrow).

PAPVC of the Left Upper Lobe Pulmonary Vein



Volume rendering of PAPVC of the Left Upper Lobe Pulmonary Vein (arrowhead).

PAPVC of the Left Upper Lobe Pulmonary Vein: CXR Findings



PAPVC (arrow) of the Left Upper Lobe Pulmonary Vein on a chest radiology.

Partial Anomalous Pulmonary Venous Connection (PAPVC) of the Left Upper Lobe Pulmonary Vein

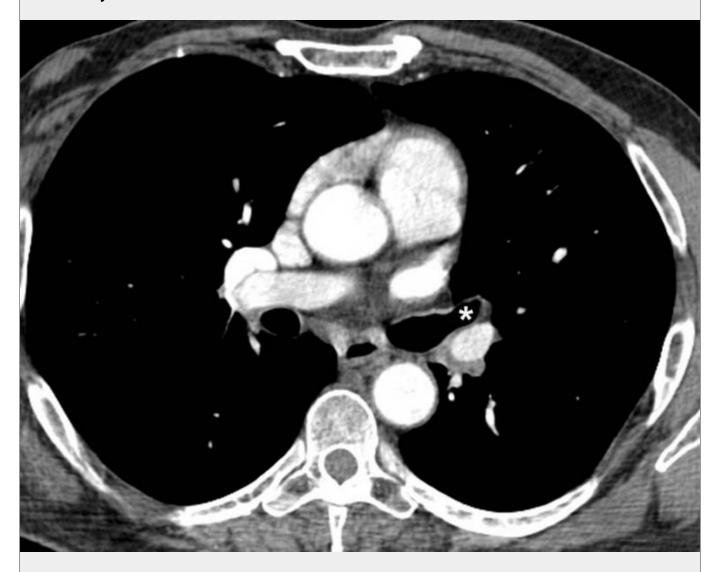
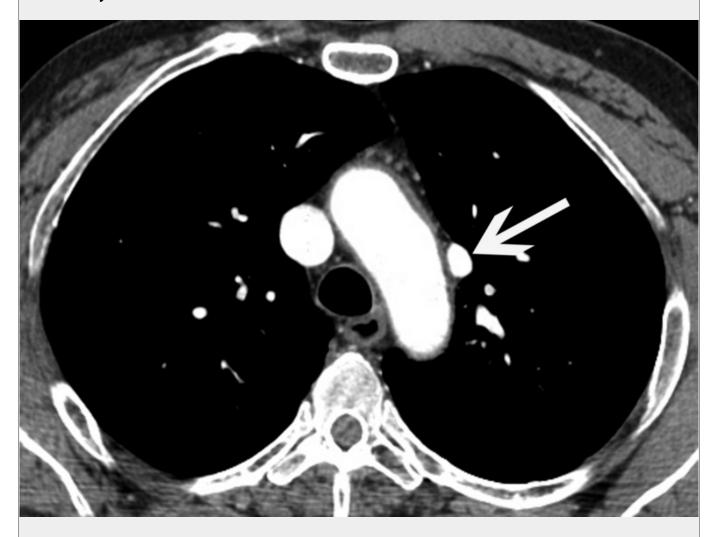


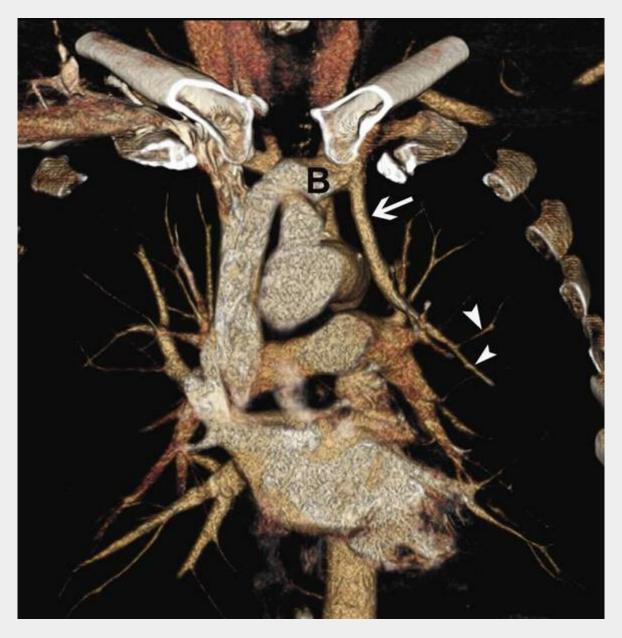
Image at a lower level demonstrates that the left superior pulmonary vein is not seen in its normal location, anterior to the left upper lobe bronchus (asterisk).

Partial Anomalous Pulmonary Venous Connection (PAPVC) of the Left Upper Lobe Pulmonary Vein

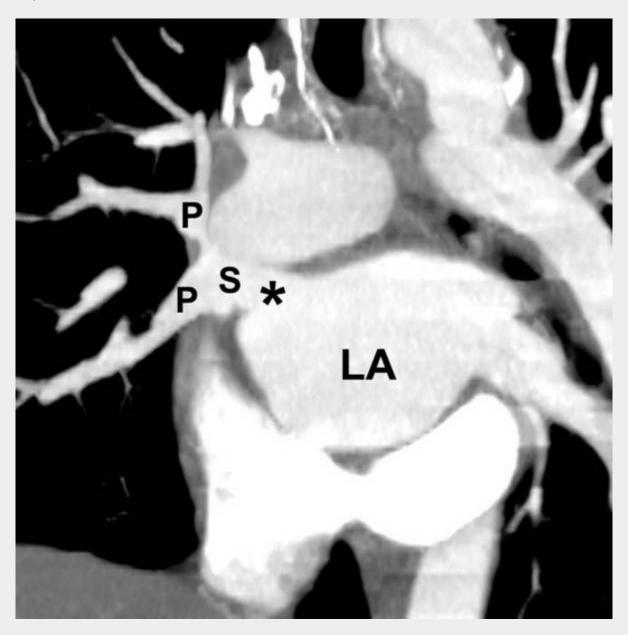


35-year-old man with PAPVC of the left superior pulmonary vein. CT demonstrates anomalous vessel (arrow) adjacent to the aortic arch.

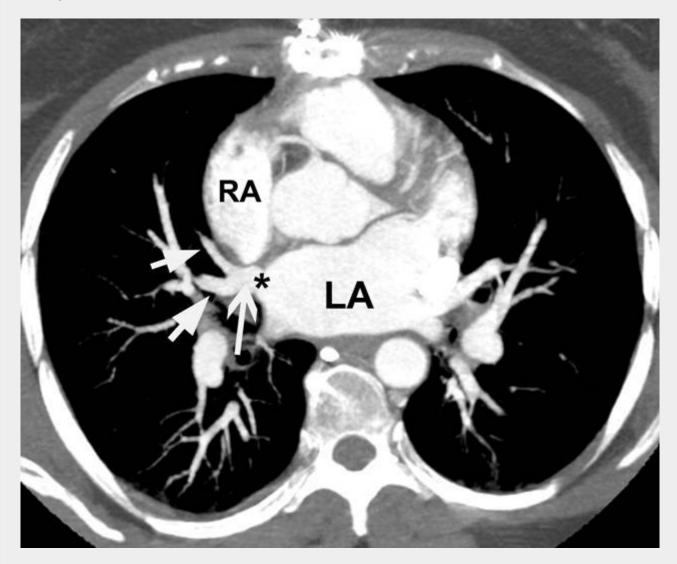
Partial Anomalous Pulmonary Venous Connection (PAPVC) of the Left Upper Lobe Pulmonary Vein



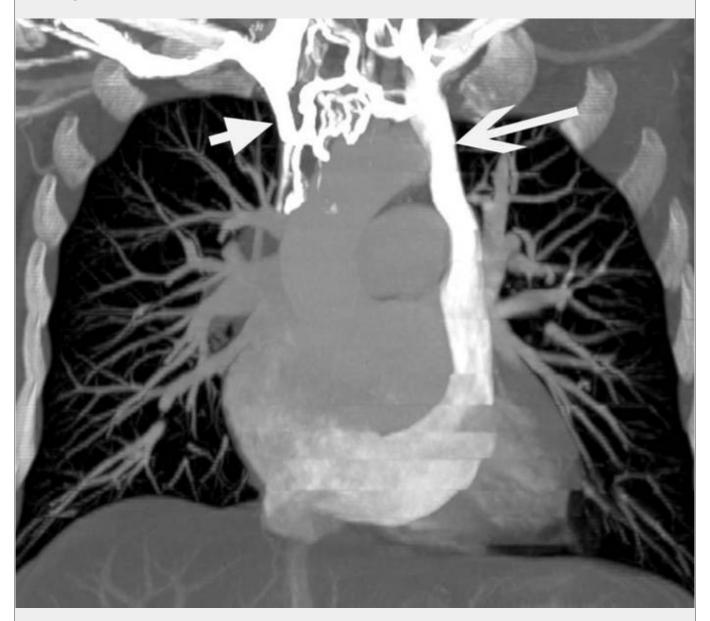
Volume rendered view from anterior perspective shows anomalous left superior pulmonary vein (arrow) joining with left brachiocephalic vein (B). Arrowheads = tributaries of left upper lobe pulmonary veins.



Reformatted coronal CT image shows anomalous pulmonary veins (P) draining to inferior segment of superior vena cava (S) to left atrium (LA) via sinus venosus atrial septal defect (asterisk).



Post-operative CT scan shows anomalous pulmonary veins (short arrows) draining to inferior portion of superior vena cava (long arrow) to left atrium (LA) via sinus venosus atrial septal defect (asterisk).



Reformatted coronal MIP image shows blood flow pattern above ligated portion of right superior vena cava. Superior portion of right superior vena cava (short arrow) communicates with left superior vena cava (long arrow) via collateral mediastinal veins.



Volume rendered from anterior perspective shows large pulmonary varix extending throughout right lung [arrows].

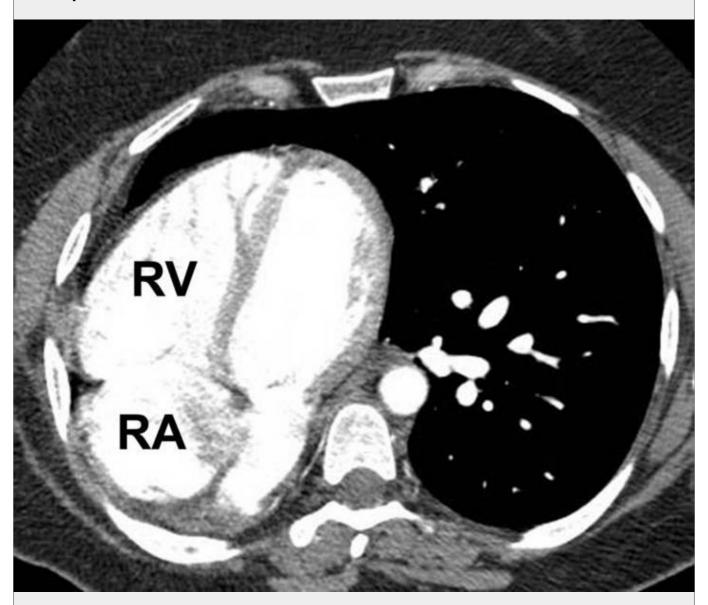
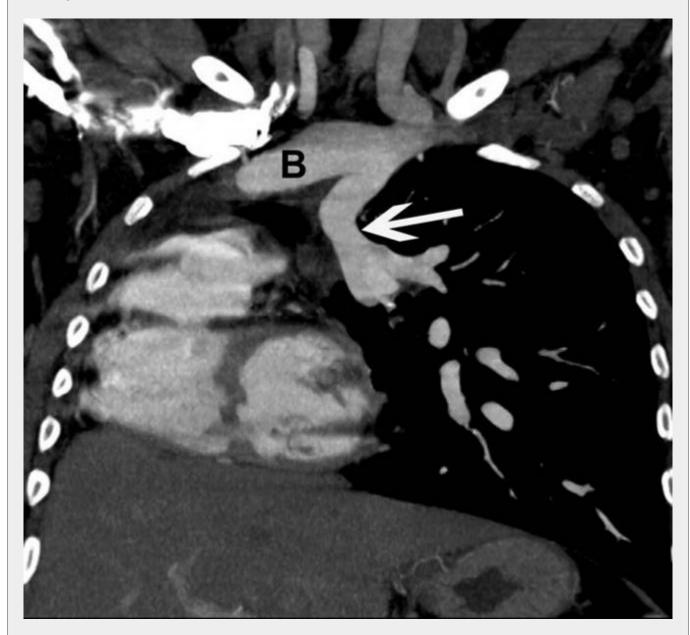
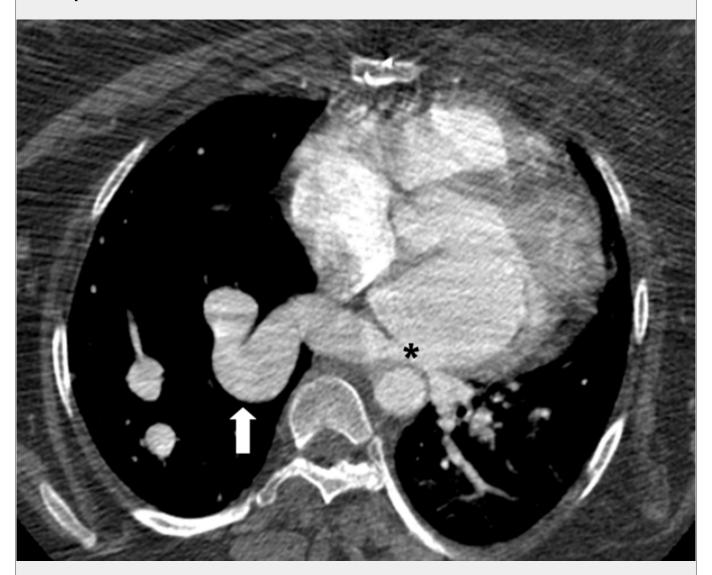


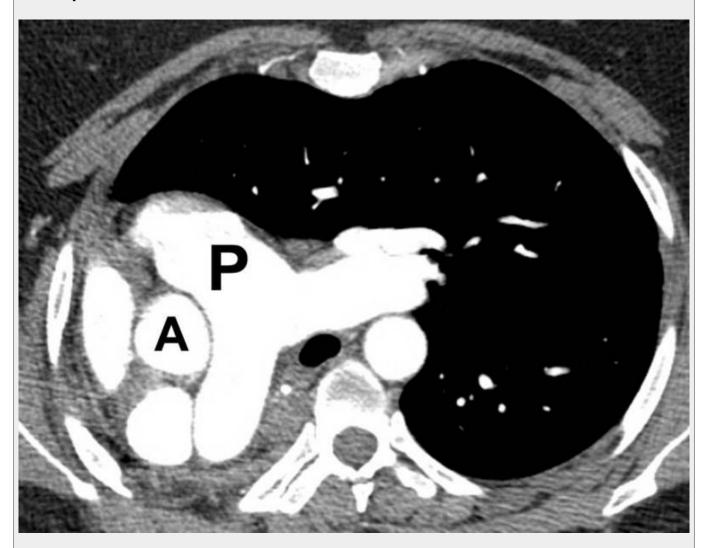
Image of heart shows that right atrium (RA) and right ventricle (RV) are dilated compared to left-sided chambers consistent with significant left to right shunt. After right pneumonectomy, a larger percentage of pulmonary venous blood shunts to the right circulation through the anomalous left superior pulmonary vein.



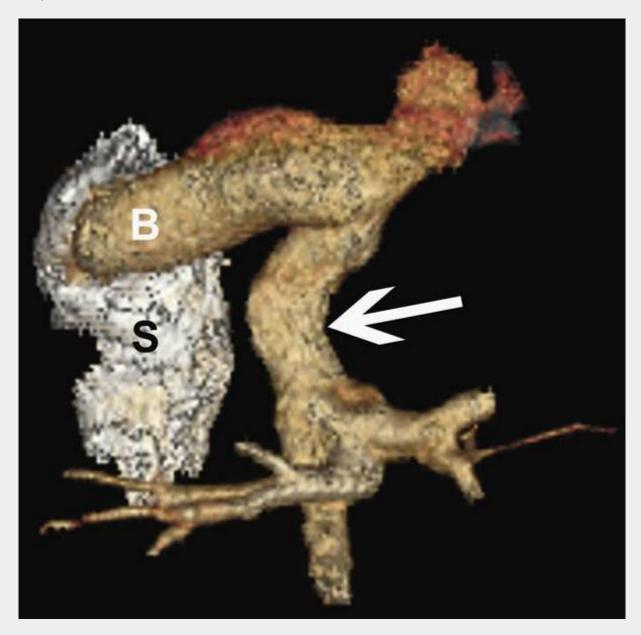
Coronal reformatted MIP image shows anomalous left upper lobe pulmonary vein (arrow) draining to left brachiocephalic vein (B).



Patient with history of PAPVC and repair with anastamosis of anomalous vein to left atrium. Patient developed stenosis at junction [asterisk] of pulmonary vein and left atrium resulting in a large pulmonary varix [arrow].



CT Image shows that main pulmonary artery (P) is larger in diameter than aorta (A) consistent with increased pulmonary blood flow.



Volume rendered view of vasculature from anterior perspective shows anomalous left upper lobe pulmonary vein (arrow) draining to left brachiocephalic vein (B). S = superior vena cava.

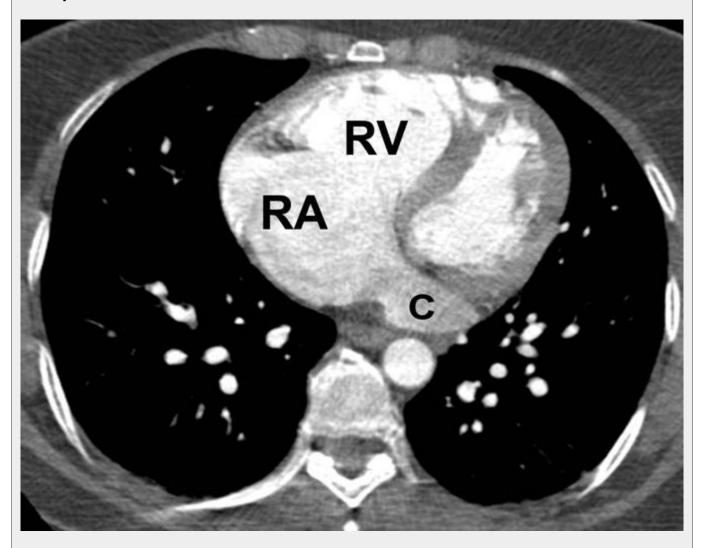
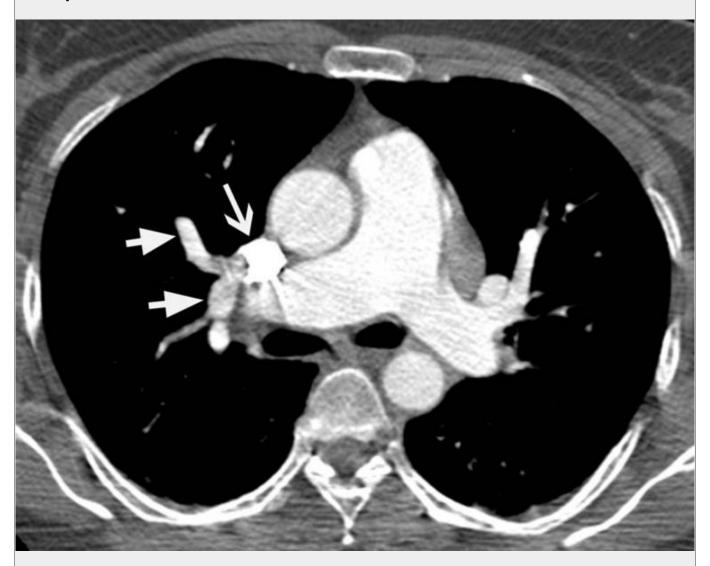


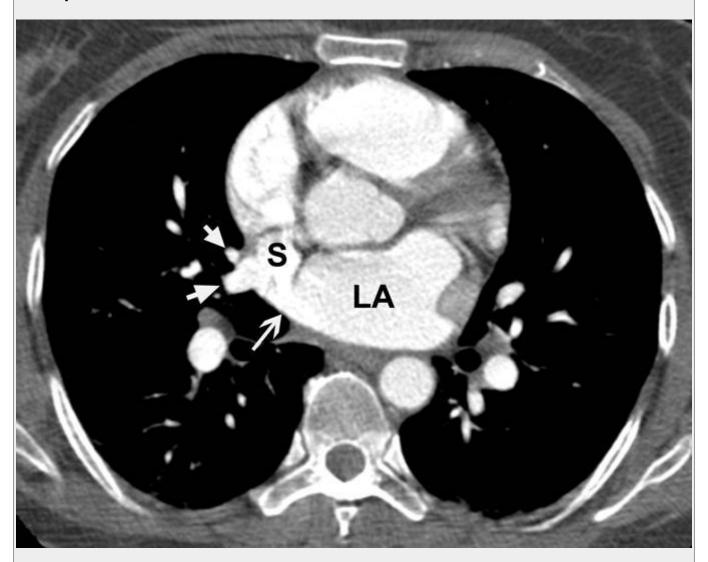
Image through cardiac chambers shows dilated right atrium (RA) and right ventricle (RV) consistent with left to right shunt. Coronary sinus (C) is also dilated due to persistent left superior vena cava.



At a slightly higher level additional anomalous pulmonary veins (short arrows) are seen draining to the superior vena cava (long arrow).



CT shows persistent left superior vena cava (arrow).



CT shows sinus venosus atrial septal defect (long arrow) allowing communication of the left atrium (LA) and superior vena cava (S). Anomalous pulmonary veins (short arrows) are also seen draining to the superior vena cava.

Strategies to detect PAPVC to SVC

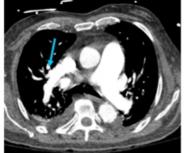


CXR shows cardiomegaly and enlarged pulmonary arteries.

Strategies to detect PAPVC to SVC

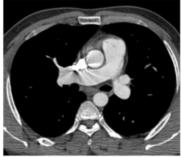
Normal windows: width = 450 level = 50

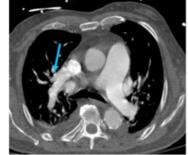






Wide windows:
Width = 800 level = 100







Note that connections of anamalous vein to SVC (arrows) are more discernable with wider windows.

Strategies to detect PAPVC to SVC: Abrupt change in caliber of SVC



CT shows dilated RA and RV, change in caliber SVC, multiple site of communication between pulmonary veins and SVC (arrows). Often with PAPVC to SVC there is more than one site of abnormal connection.