

# A CASE OF PARTIAL ANOMALOUS PULMONARY VENOUS CONNECTIONS

Comprehensive cardiovascular assessment with Multimodality Imaging and discussion of possible surgical versus conservative management.

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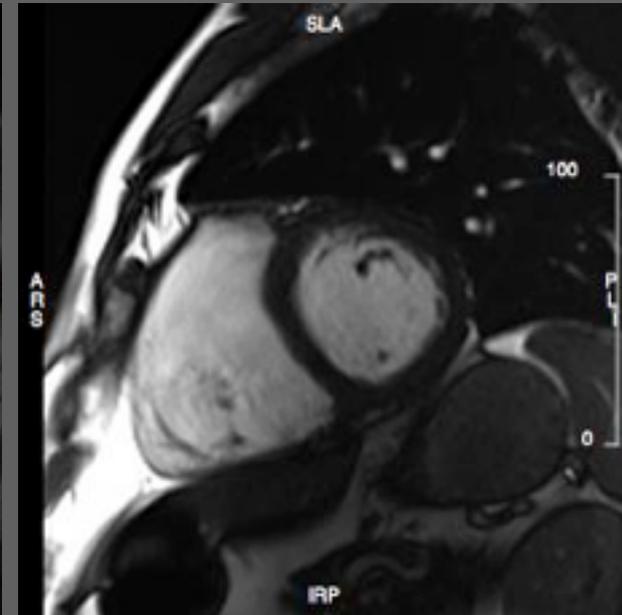
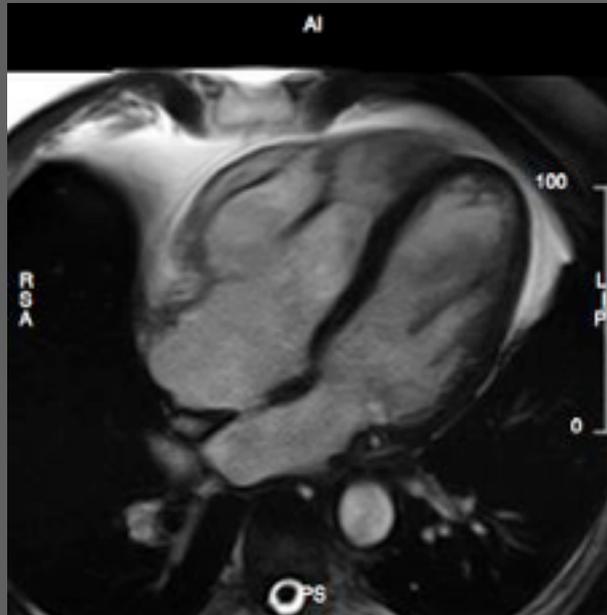
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# Clinical Presentation:

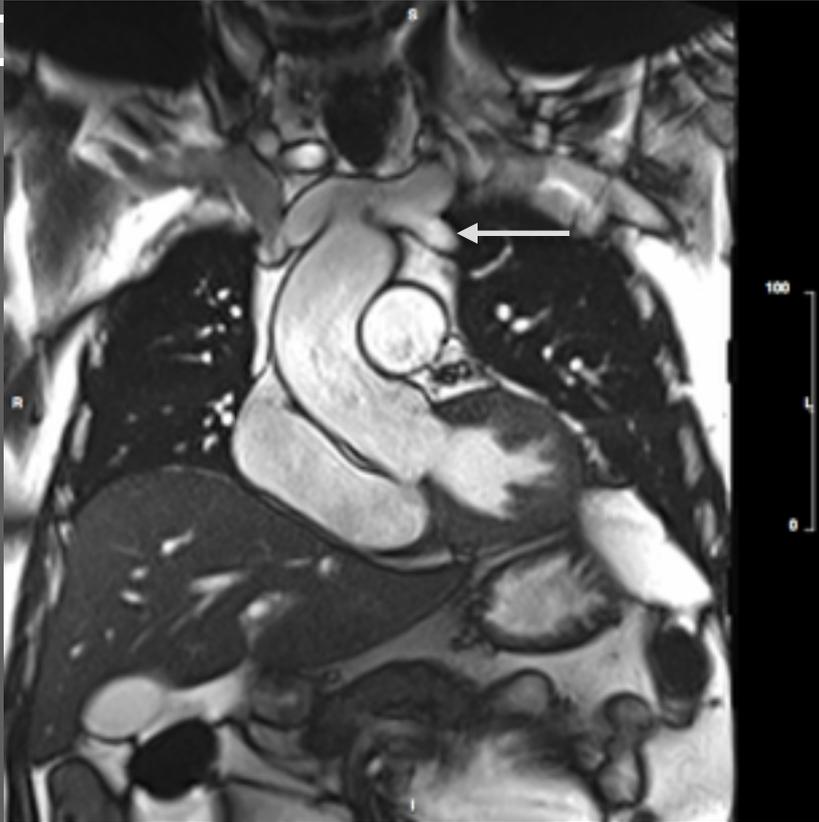
- A 35 year old caucasian male who has been active and well, presented with increasing frequency of palpitation, some breathlessness but
- He **managed 12 minutes** of Bruce protocol with no symptoms or significant ECG changes. Recent chest infection but otherwise asymptomatic.
- Clinical examination was unremarkable, Liver not enlarged, not tender, no haemoptysis, cyanosis or ankle oedema.
- biochemistry and haematology unremarkable.
- echocardiography suggests RV dilatation and mild tricuspid dilatation.
- No ASD, no areas of right ventricular aneurysmal dilatation or regional hypokinesia were seen, and the patient was referred for CMR to determine cause of significant RV dilatation.

# Cine SSFP

- ▣ Significant right ventricular dilatation
- ▣ Mild TR
- ▣ No ASD



# CMR



# CTPA



# CTPA

WL: 204 WW: 182



RA

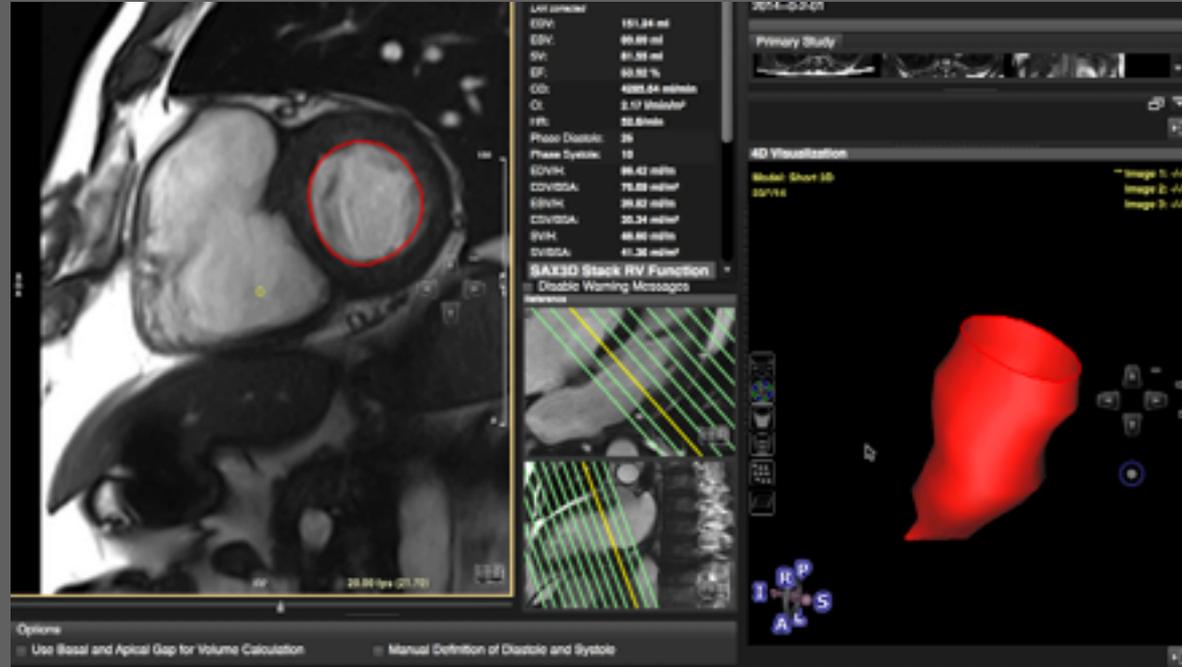
LP



RA

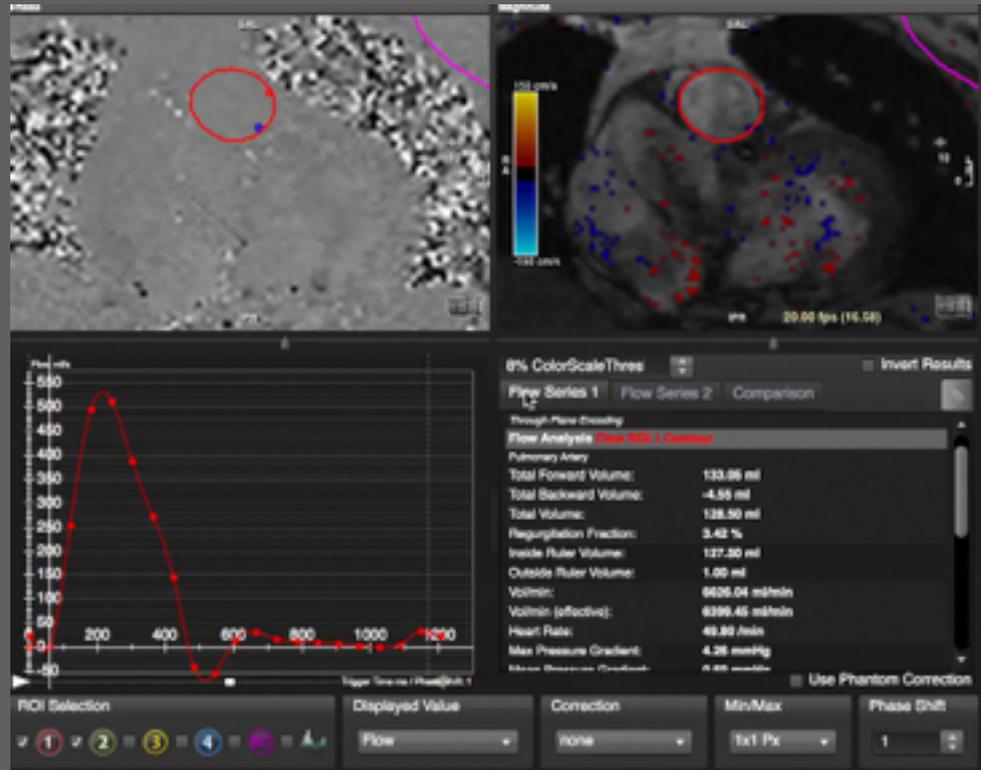
# 4D LV and RV Volumetric Analysis

- In addition to morphological assessment, it is important to get detailed quantitative information of volume loading of the left and right ventricle and shunt volumes.
- This can be calculated by choosing an end-diastolic and end-systolic frames of both ventricles of the short axis SSFP cine images.
- In normal conditions and in the absence of significant valve regurgitation, there is no significant difference between left and right ventricular stroke volumes.
- The stroke volume difference in this case was:
- $RVSV = 151 - LVSV = 125\text{ml} = 26\text{ml}$



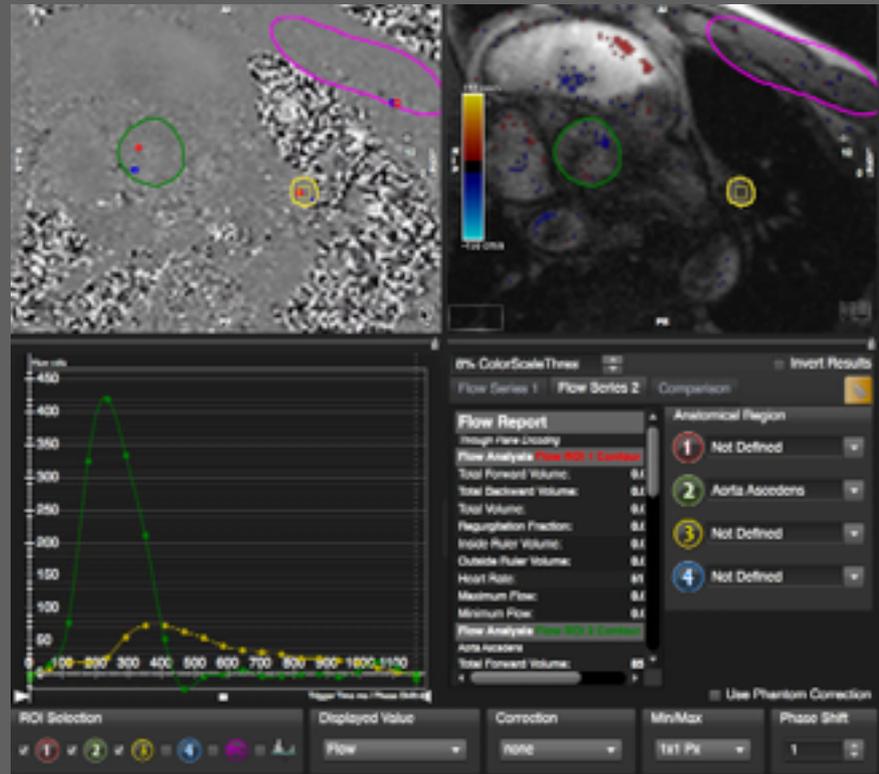
# Quantitative Assessment by phase shift velocity mapping

- Phase velocity mapping allows for accurate and reproducible measurements of blood flow and can be used to directly assess flow volume through blood vessels.
- A region of interest is drawn on the ascending aorta and pulmonary artery respectively, perpendicular to the flow direction around the pulmonary artery.
- Flow volume difference is assessed by comparison of flow differences between systemic and pulmonary blood flow ( $Q_p:Q_s$ )
- Any additional differences in flow are explained by tricuspid valve or mitral valve regurgitation.



# Aortic and Pulmonary Vein Flow Analysis

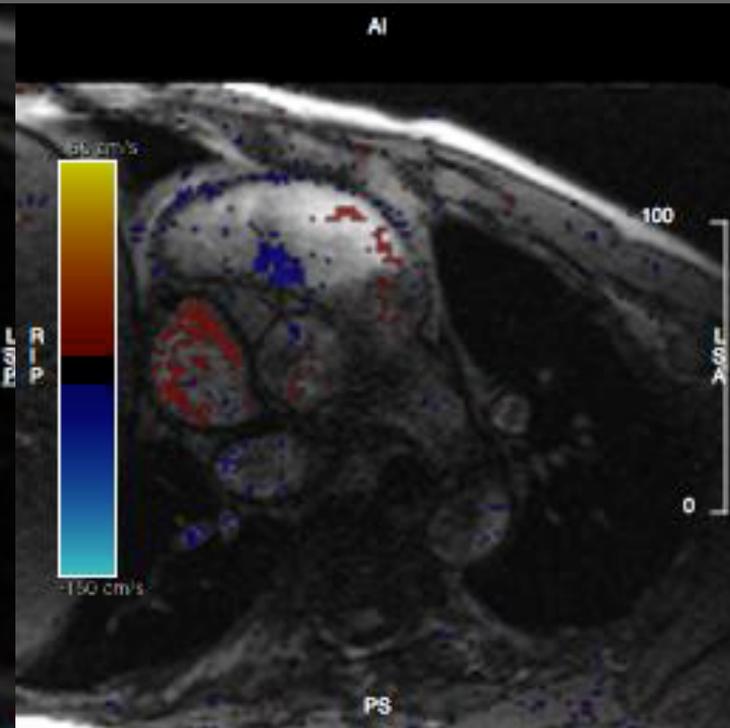
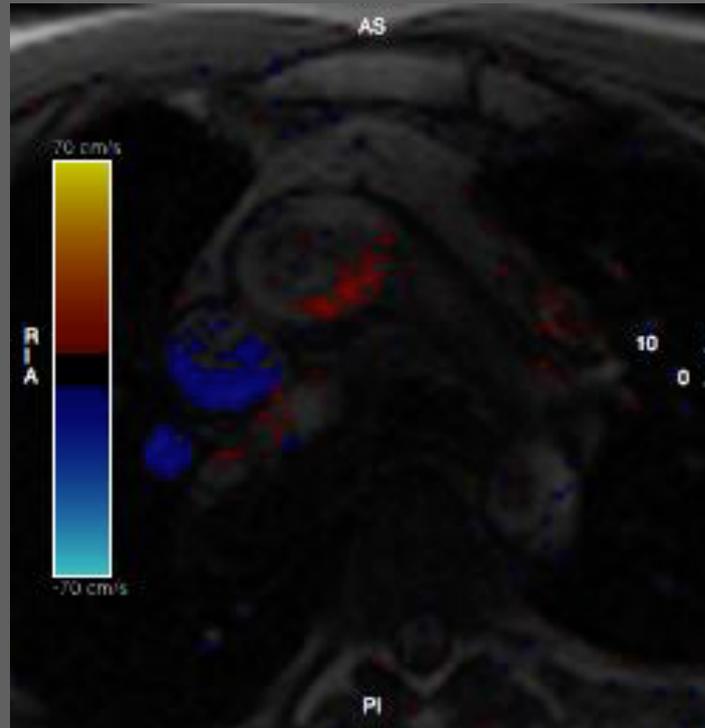
- PA flow=128ml
- Ascending Aorta flow = 83ml
- Stroke volume difference by phase velocity mapping = 45ml.
- Direct phase velocity encoded assessment of the left upper PV was 38ml.
- The difference between the two is 7ml and corresponds to mild tricuspid regurgitation.
- The ratio of Pulmonary flow : Systemic arterial flow ( $Q_p:Q_s$ ) is 128:83 or 1:1.54



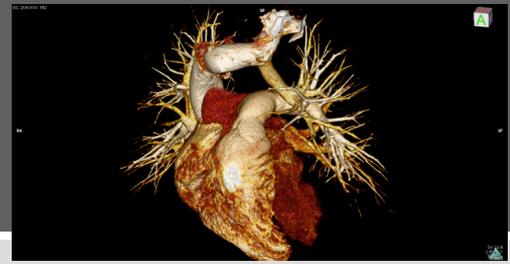
# Shunt assessment by Phase Shift Velocity Mapping- the importance of Perpendicular Alignment

Suboptimal (7% error)

Better alignment



# Types of anomalies



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- **PAPVC with ASD 80-90% of cases**

This is the most common type of PAPVC. The ASD is usually the sinus venosus type. The anomalous pulmonary vein, usually the right upper or middle pulmonary vein can either override the intra atrial septum (anomalous drainage) or can drain separately into the superior vena cava (true anomalous connection). Usually, the connection is unobstructed.

- **PAPVC with intact atrial septum (isolated PAPVC)** : Only 3% of patients had PAPVC from the left lung to the innominate vein.

This is a very rare finding and mostly involves the anomalous drainage of the right upper pulmonary vein into the superior vena cava.

- **PAPVC with complex congenital heart disease (heterotaxia)**

This is usually seen with heterotaxia syndromes (polysplenia). Left atrial isomerism with a common atrium is observed. Because of the abnormal positioning of the intra-atrial septum, the right-sided pulmonary veins anastomose to the anatomically right-sided atrium, which is the atrium that also receives the inferior vena cava. About half of the cases may involve some degree of obstruction, either due to narrowing of a discrete area or due to diffuse hypoplasia of that vein. This condition is associated with presence of ipsilateral pulmonary arterial hypoplasia.

- **Scimitar syndrome** (right pulmonary vein to inferior vena cava with lung sequestration)

This syndrome is also known as Halasz syndrome, mirror-image lung syndrome, hypogenetic lung syndrome, epibronchial right pulmonary artery syndrome, vena cava bronchovascular syndrome, or congenital pulmonary venolobar syndrome. It is more common in females and can be familial

# How common is it ? 0.1%

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J Thorac Imaging. 2009 May;24(2):89-95. doi: 10.1097/RTI.0b013e318194c942.

MDCT of partial anomalous pulmonary venous return (PAPVR) in adults. Ho ML1, Bhalla S, Bierhals A, Gutierrez F.

**PURPOSE:**to determine the lobar distribution and associated radiologic/clinical findings of partial anomalous pulmonary venous return (PAPVR) in the adult population using multidetector computed tomography (MDCT).

**RESULTS:**

Calculated disease prevalence was 0.1%, mean patient age of 58 years and a 58% female predominance. PAPVR was observed with 47% frequency in the left upper lobe, 38% right upper lobe (RUL), 13% right lower lobe, and 2% left lower lobe. Among cases of RUL PAPVR, 42% were associated with sinus venosus atrial septal defect (ASD). Other reported anomalies were right-sided volume overload (47%), isolated upper lobe PAPVR (29% left and 5% right), bilateral PAPVR (4%), scimitar syndrome (13%), persistent left superior vena cava (9%), and azygos continuation of the inferior vena cava (4%). Reported cardiopulmonary signs/symptoms and imaging modalities other than MDCT were neither sensitive nor specific for PAPVR. Surgical repair was performed in 21% of cases and included ASD patching, intracardiac baffle, anomalous vein anastomosis, systemic vein translocation, and Warden procedure.

# CTPA

WL: 204 WW: 182



#### References

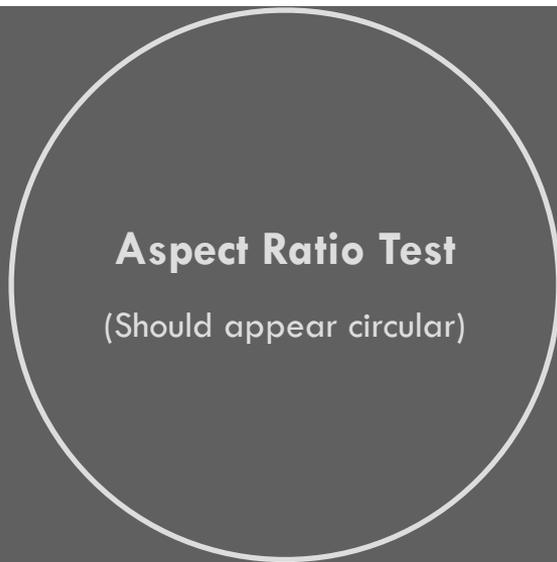
Myerson SG , Bellenger N, Pennell, D Assessment of Left Ventricular Mass by Cardiovascular Magnetic Resonance

Mohiaddin R.H, Makoto A, Kilner P.J, Pennell D.J, Manzara C, Longmore D.B; MR Phase-shift velocity mapping of mitral and pulmonary venous flow. J Comput Assist Tomogr. 15 1991:237-243.

Javangula K, Cole J, Cross M, Kay PH. An unusual manifestation of left partial anomalous pulmonary venous connection. Interact CardioVasc Thorac Surg 2010;11:846-848.

Al-Ahmari S, Chandrasekarank, BrilakasE, TahlilW, Dearani J, Malouf J, Gilman G, Seward JB, Tajik AJ. Isolated partial anomalous pulmonary venous

# Widescreen Test Pattern (16:9)



**Aspect Ratio Test**

(Should appear circular)

4x3

16x9