

# Sinus venosus atrial septal defect with persistent left SVC and partial anomalous pulmonary venous drainage.

## Clinical Case Portal

### Date of publication:

27 Sep 2006

**Topics:** Congenital Heart Disease

### Authors:

Claire Waterworth MSc, David Dawson MSc, Petros Nihoyannopoulos MD FRCP FACC FESC

Echocardiography Department, Hammersmith Hospital, Du Cane Road, London W12 0HS

e-mail: [petros@ic.ac.uk](mailto:petros@ic.ac.uk)

## Case Report

We present a 72 year old female complaining of breathlessness who was referred for assessment of her pulmonary hypertension. Transthoracic echocardiography with contrast revealed a sinus venosus atrial septal defect with persistent left SVC and partial anomalous pulmonary venous drainage.

Patient history prior to current observation :A 72 year old female presented to her general practitioner complaining of breathlessness. She was a long-term smoker, but gave up in 2003. The GP found her to be hypertensive, and a loud systolic murmur was heard. She was referred to her local hospital, where echocardiography showed normal left ventricular function, a dilated right ventricle and mild tricuspid regurgitation with an estimated pulmonary artery pressure of 75mmHg. She was subsequently referred to the National Pulmonary Hypertension service at Hammersmith Hospital for assessment of her pulmonary hypertension.

Clinical findings on admission, evolution and outcome :

**Clinical Examination:** Heart rate was 110bpm and regular with ectopic beats, blood pressure was 137/92mmHg, respiratory rate was 12 breaths per minute, oxygen saturation on air was 98% and JVP was elevated at 7cm above the sternal angle. Her apex beat was displaced laterally to the anterior axillary line and to the sixth intercostal space. She had a thrill in the pulmonary area as well as a harsh systolic murmur. S1 was audible and her pulmonary second sound was loud. She had mild ankle oedema and normal peripheral pulses. Respiratory and abdominal examinations were normal.

**Chest X-Ray** fig. 1 showed gross cardiac enlargement with an unfolded and ectatic aorta, large central pulmonary arteries and no focal lung lesions.

**ECG** fig. 2 showed sinus rhythm with right axis deviation, incomplete right bundle branch block and right ventricular hypertrophy.

**Transthoracic Echocardiography** showed a markedly dilated coronary sinus and a dilated, volume loaded right ventricle fig. 3. Left arm vein injection of agitated gelofusine confirmed the diagnosis of a persistent left SVC fig. 4, fig. 5. A sinus venosus atrial septal defect was identified directly on imaging fig. 6, with anomalous right upper pulmonary venous drainage to the origin of the SVC, straddling the atrial septal defect fig. 7. Peak TR velocity was 3.6m/s and Qp/Qs was 2.6:1.

**Reconstructed CT images** fig. 8 revealed that the right upper pulmonary vein drains directly into the right SVC.

**Cardiac Catheterisation** fig. 9 revealed tortuous coronaries with no flow limiting lesion. Mean PA pressure was 34mmHg, LVEDP 12mmHg with a cardiac index of 6.26. PVR was 177 and Qp/Qs was 1.3:1.

On review of these results, it was felt that her symptoms and mildly elevated PA pressure were mainly related to the atrial septal defect. She was referred back to her local cardiologist for potential closure of the defect.

## Discussion

In patients with a dilated coronary sinus, there are three differential diagnoses; pulmonary hypertension, a persistent left superior vena cava and partial anomalous pulmonary venous drainage.

A persistent left superior vena cava is the most common venous abnormality of the thorax, affecting 0.5-2% of the general population, and up to 10% of patients with congenital abnormalities of the heart<sup>1</sup>. It is of little clinical significance, and requires masterly inactivity since the systemic venous blood continues to return to the right atrium. However, if the patient is referred for cardiac surgery the surgeon needs to be made aware of the presence of a persistent left SVC to avoid blood flooding the field during cardiopulmonary bypass. Similarly if the patient requires diagnostic or therapeutic transcatheter procedures such as pacemaker implantation, the operator needs to be made aware to avoid complications during the procedure. The diagnosis is easily confirmed by transthoracic echo, with a left arm vein injection of agitated saline or gelofusine<sup>2</sup>. In the presence of a persistent left SVC, in the parasternal long axis view, the contrast will appear first in the coronary sinus and then in the RV fig. 4.

There are seven common variations of partial anomalous pulmonary venous drainage<sup>3</sup>:

- Right upper and middle lobe pulmonary vein to superior vena cava
- Right upper and middle lobe pulmonary vein to right atrium
- Right pulmonary veins to inferior vena cava
- Right lower pulmonary vein to inferior vena cava with anomalous arterial blood supply to right lower lobe and lung sequestration (Scimitar syndrome)
- Left upper pulmonary vein to innominate vein via and anomalous left vertical vein

- Left upper or lower pulmonary vein(s) to coronary sinus
- Left lower pulmonary vein to right atrium or inferior vena cava

The most common variant is a connection between the right upper and middle lobe pulmonary vein to the superior vena cava or the right atrium<sup>4</sup>. In this case the right lower lobe pulmonary vein is often connected normally to the left atrium. It is still a rare congenital lesion, affecting 0.4-0.7% of congenitally malformed hearts<sup>3</sup>. Patients with isolated partial anomalous pulmonary venous drainage are usually completely asymptomatic early in life, and those patients that do develop symptoms have more than one anomalously connected pulmonary vein, or associated lesions. Symptoms, if they do exist, relate to the magnitude of left-to-right shunting and resemble those of an atrial septal defect – exertional dyspnea, atrial arrhythmias and, later in life, right heart failure.

One of the most common lesions associated with partial anomalous pulmonary venous connection is a sinus venous atrial septal defect. A sinus venosus atrial septal defect occurs when there is a deficiency of infolding of the atrial wall in the region of the superior caval vein. It is found within the mouth of the superior caval vein, which has a biatrial connection, overriding the rim of the oval fossa so as to produce what is effectively an extracardiac but interatrial communication<sup>5</sup>. Again, it is a rare lesion, making up approximately 10% of all atrial septal defects<sup>6</sup>. It is often not seen on standard transthoracic echo, but should be considered when the right ventricle appears dilated and volume loaded with an apparently intact inter-atrial septum.

## Conclusion

This case demonstrates the importance of investigating all differential diagnoses and looking for associated lesions in congenital heart disease. Often, congenital abnormalities such as those seen in this case are not imaged directly, but there are indirect indicators, which should lead to further investigation. When a dilated coronary sinus is seen on transthoracic echo, a left arm vein injection of contrast (agitated saline or gelofusine) is readily diagnostic for persistent left SVC. Similarly, when the right ventricle appears dilated and volume loaded with an apparently intact atrial septum, non-conventional transthoracic echo views may be needed to look for a sinus venosus atrial septal defect.

## References

1. Edwards J, DuShane J. Thoracic venous anomalies. *Arch Pathol* 1950; 49: 514-37.
2. Foale R, Bourdillon PD, Somerville J, Rickards A. Anomalous systemic venous return: recognition by two-dimensional echocardiography. *Eur Heart J*. 1983; 4: 186-195.
3. Gatzoulis MA, Webb GD, Daubeney PEF. Diagnosis and management of adult congenital heart disease 2003 pp205.
4. Brody H. Drainage of the pulmonary veins into the right side of the heart. *Arch Pathol* 1942; 33: 221-229.
5. Al Zaghaf AM, Li J, Anderson RH, Lincoln C, Shore D, Rigby ML. Anatomical criteria for the diagnosis of sinus venosus atrial septal defects. *Heart* 1997; 78: 298-304.
6. Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. First of two parts. *N Engl J Med*. 1990 Dec 13; 323(24): 1645-50.

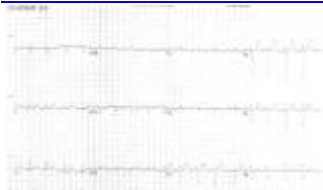
Fig. 1 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage\\_ Chest X-Ray](#)



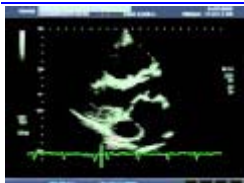
Fig. 2 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage\\_ 12 Lead ECG](#)



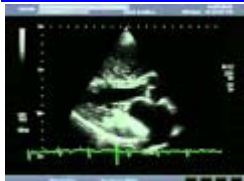
Video 1 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage\\_ TTEcho: Plax view](#)



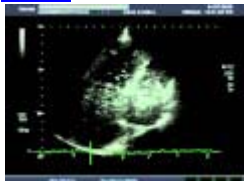
Video 2 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage\\_ Contrast Echo: PLAX](#)



Video 3 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage\\_ TTEcho: Subcostal View](#)



Video 4 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage\\_ TTEcho: Subcostal View](#)



Video 5 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage TTEcho: Ap4ch](#)

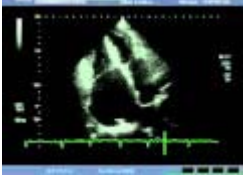


Fig. 3 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage Reconstructed CT - partial anomalous pulmonary venous drainage](#)



Fig. 4 :

[Sinus venosus ASD - left SVC and partial anomalous pulmonary venous drainage Cardiac Catheterisation](#)

