

# IMAGES

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### **Late presentation and successful treatment of classical scimitar syndrome**

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### **Abstract**

Scimitar syndrome is a form of partial anomalous pulmonary venous drainage that is dramatically visible on plain chest radiography (CXR). In these individuals the entire venous drainage from the right lung enters a single anomalous large vein that descends to the inferior vena cava. This descending vein is visible on CXR as a curvilinear density along the right heart border and resembles the curved Turkish sword that gives the condition its name. Scimitar syndrome forms part of the large spectrum of associated conditions known as venolobar syndrome. These include right lung hypoplasia or sequestered segments of right lung, congenital heart disease and various others. We report the case of a young woman who presented incidentally, with a murmur, at 16 years of age. Full investigation including angiography showed a large atrial septal defect with right heart dilation and scimitar syndrome. She underwent surgical correction with uneventful and complete correction by baffling of the scimitar vein from its entry into the inferior vena to the left atrium through the enlarged atrial septal defect.

**MeSH:** heart defects, congenital, scimitar syndrome

### **Introduction**

Scimitar syndrome is a form of anomalous pulmonary venous drainage (APVD). APVD implies partial or total failure of the pulmonary veins to reach the left atrium. Instead, pulmonary venous drainage is anomalously connected to systemic vein/s, typically to the superior or inferior vena cava (SVC or IVC) or directly to the right atrium (RA). APVD accounts for up to 2% of all congenital heart disease and is usually associated with left to right shunting at atrial level through some form of atrial septal defect (ASD). The haemodynamic effects of APVD are also those of left to right shunting at atrial level, thereby exacerbating right heart volume overload.

The most common form of partial AVPD is connection to the SVC or SVC-RA junction in association with a superior sinus venosus defect. Scimitar syndrome is

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rarer, with the right pulmonary vein/s draining to the inferior vena cava. This abnormal form of drainage is visible on plain chest radiography, and was first described in 1836.<sup>1</sup> The APVD is seen as a curvilinear density along the right heart border, similar in shape to the classic Turkish curved sword. The term 'scimitar syndrome' was coined by Neill in 1960 to describe the CXR appearance.<sup>2</sup> Scimitar syndrome must be differentiated from the pseudoscimitar syndrome in which an abnormal descending vein pursues an aberrant course in the right lung but drains normally into the left atrium, and from Kartagener's syndrome.

Scimitar syndrome is also known as Halasz's syndrome, mirror-image lung syndrome, hypogenetic lung syndrome, epibronchial right pulmonary artery syndrome and vena cava bronchovascular syndrome. It occurs more commonly in females and is occasionally familial.<sup>2</sup> The left lung is very rarely involved and the reason for this is unknown.<sup>3</sup> The true incidence of this condition is unknown since the syndrome may remain undetected in asymptomatic patients who do not undertake a CXR.

Scimitar syndrome overlaps with pulmonary sequestration and the term venolobar syndrome has been coined to include these associated pulmonary and vascular malformations.<sup>4</sup>

The condition is associated with:<sup>4</sup>

1. Partial agenesis or hypoplasia of the right lung with bronchial isomerism
2. Diverticulum or hypoplasia of the right bronchial system.
3. Hypoplasia or agenesis of the right pulmonary artery. This may cause mediastinal shift to the right side and the scimitar vein may be difficult to appreciate or even completely obscured.
4. Abnormal systemic blood supply to at least part of the right lung, most frequently the posterior basal segment of the lower lobe, usually arising from the infradiaphragmatic descending aorta.
5. Dextrocardia due to right lung hypoplasia with mediastinal shift.
6. Accessory diaphragm or eventration or partial absence of the diaphragm.
7. Phrenic cyst
8. Horseshoe lung
9. Oesophageal and gastric lung
10. Absence of the pericardium
11. Other congenital cardiac malformations (25% of cases) including ASD, ventricular septal defect, coarctation of the aorta, tetralogy of Fallot, pulmonary stenosis, absent inferior vena cava with azygos continuation to superior vena cava and persistent left superior vena cava.<sup>5</sup> Moreover, the APVD may also be partially obstructed, further contributing to pulmonary hypertension.<sup>6</sup>

The detection of any of these anomalies should lead to a search for other components. The clinical spectrum of scimitar syndrome ranges from severely ill infants to asymptomatic adults, and cases may present in one of three ways:<sup>7,8</sup>

In the neonatal period with respiratory and/or cardiac failure. This is most commonly caused by pulmonary hypertension due to cardiac and/or right lung anomalies. Treatment is surgical and outcome is dependant on the nature and severity of the anomalies.<sup>9</sup> Heart failure may also be caused because of a large arterial supply from the abdominal aorta to a sequestered lobe. In these situations, cardiac catheterisation may be used to embolise the aberrant pulmonary blood supply. This not only relieves heart failure but also makes the surgical field more bloodless.

At any stage in life due to recurrent chest infections, usually affecting the right lower lobe that often has an abnormal arterial blood supply and venous drainage. Severity and frequency of chest infections is related to the degree of pulmonary hypoplasia.

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Lobectomy or even right pneumonectomy may be required to deal with bronchiectasis and prevent further chest infections. Affected individuals may also present with haemoptysis due to pulmonary hypertension.<sup>10</sup>

At any stage in life as an incidental finding e.g. due to the detection of a murmur or due to the evident CXR abnormalities.<sup>11</sup>

Useful investigations include echocardiography, angiography and computerised chest tomography.<sup>12</sup> Magnetic resonance imaging also delineates abnormal vessels. APVD with significant left to right shunting is corrected surgically, in this case, by baffling anomalous pulmonary venous return to the left atrium through the inferior vena cava or by reimplantation of the scimitar vein to the left atrium. Potential complications include acute or chronic postoperative thrombosis at the anastomotic/baffle sites.

We present a case of scimitar syndrome that presented in the late teens with a murmur. The patient (female) was well and asymptomatic, with no cardiac or respiratory problems. Echocardiography showed a dilated right heart and left to right shunting across a moderate ASD. Transoesophageal echocardiography confirmed the defect and showed left pulmonary veins draining to left atrium, but failed to demonstrate the right pulmonary veins.

Cardiac catheterisation easily demonstrated both left pulmonary veins but failed to show the right pulmonary veins (figures 1,2).

Figure 1 Left upper pulmonary vein to left atrium (hand injection)

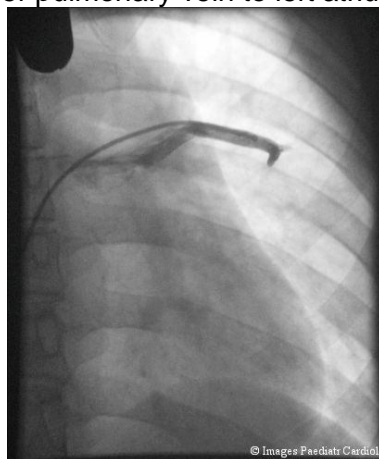


Figure 2 Left lower pulmonary vein to left atrium (hand injection)



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Right pulmonary artery angiogram showed classical scimitar syndrome, with right pulmonary veins joining to a descending (scimitar) vein that drained to the inferior vena cava (figures 3-5).

Figure 3 Right pulmonary artery angiogram - arterial phase.



Figure 4 Right pulmonary artery angiogram - venous phase.

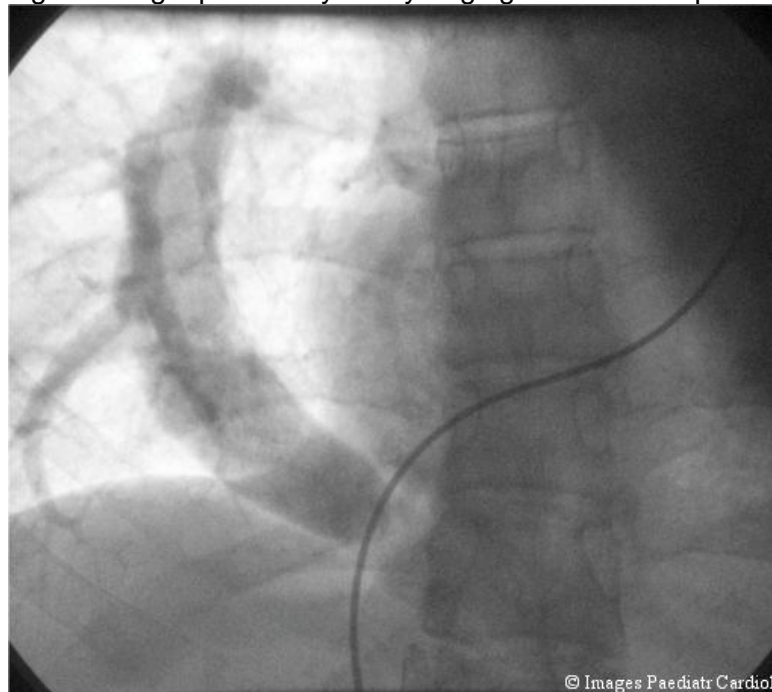


Figure 5 Right pulmonary artery angiogram - arterial and venous phases superimposed



In retrospect, the scimitar vein was also visible on direct fluoroscopy (figure 6). The entire right pulmonary artery angiogram is shown in figure 7.

Saturations confirmed left to right shunting with step-up at the level of the inferior vena cava (figure 8).

Plain CXR (figure 9) and lateral CXR (figure 10) also amply demonstrated the scimitar vein along with no other pulmonary pathology.

Chest computerised tomography was performed to exclude the possibility of pulmonary sequestration. Sequestration was not present, but the descending vein could be clearly seen within the right lung (figure 11), and inferiorly coursing medially towards the inferior vena cava (figure 12).

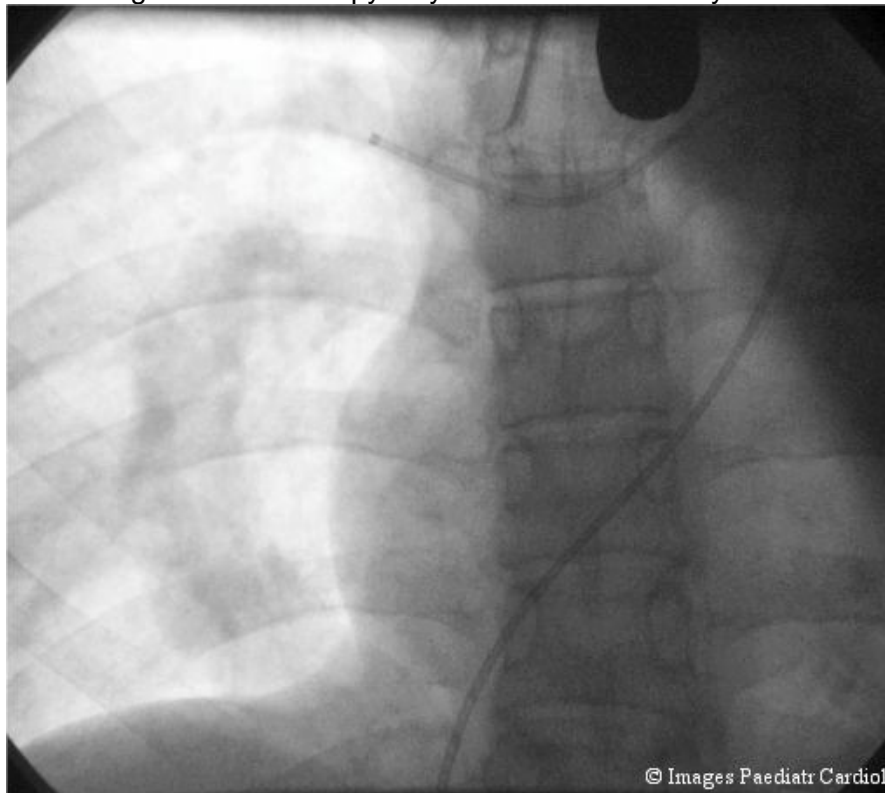
Surgical repair was successfully and uneventfully carried out by baffling the orifice of the scimitar vein at its entry into the inferior vena cava to the ASD. Autologous pericardium was used (figures 13-16). The patient was placed on bypass with bicaval drainage and aortic return and cooled to 18 degrees celcius. Due to the close proximity of the hepatic vein confluence and the inferior vena cava, the latter could not be snared in order to isolate the venous return to the right atrium. Consequently, the entire corrective procedure was performed under hypothermic circulatory arrest.

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Figure 7 Right pulmonary artery angiogram



Figure 6 Fluoroscopy only - scimitar vein clearly seen



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Figure 8 Saturations showing step-up at the level of the inferior vena cava

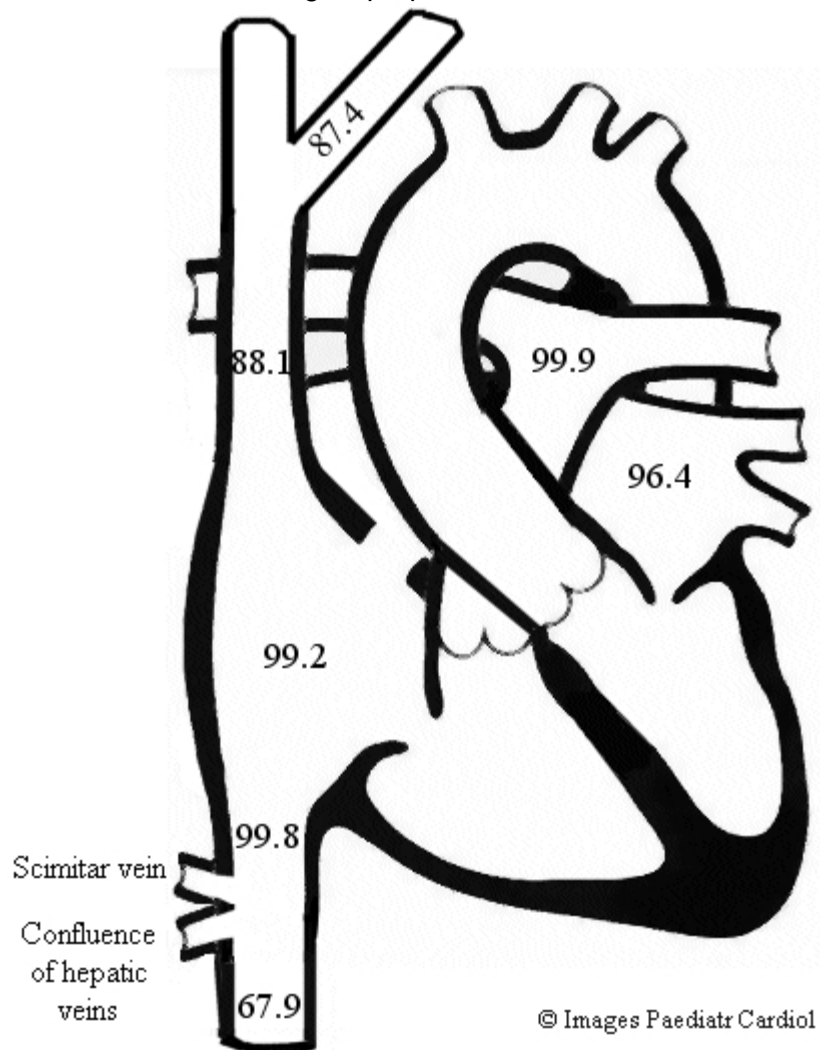
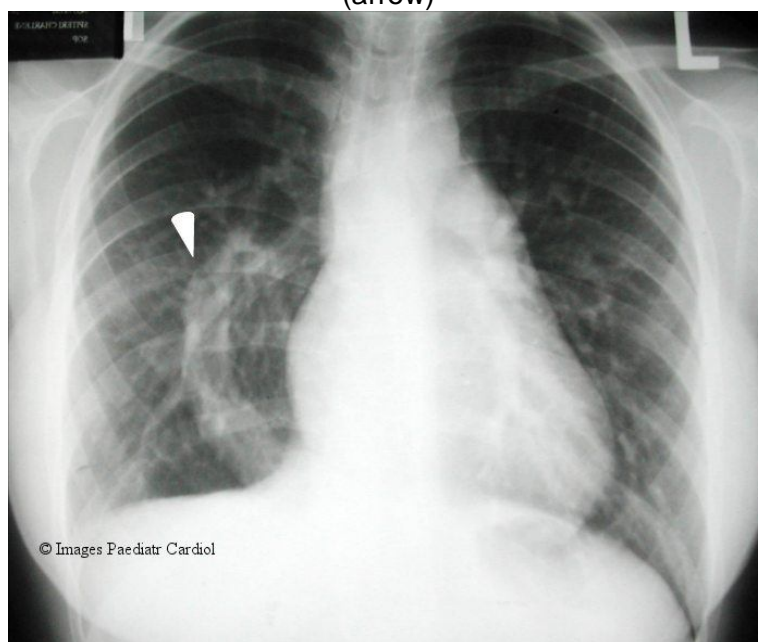


Figure 9 CXR - scimitar vein seen as a curvilinear density along the right heart border (arrow)





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Figure 10 Lateral CXR - scimitar vein seen as a curvilinear density (arrows)

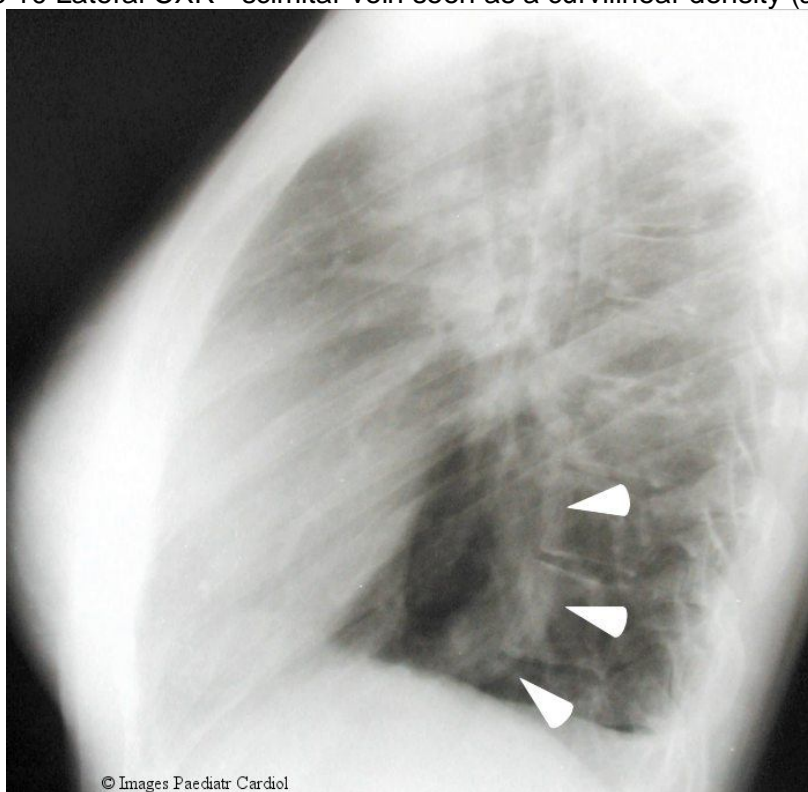


Figure 11 Chest CT slice showing descending vein in right lung field (arrow)





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Figure 12 Chest CT slice showing descending vein in right lung field coursing towards the inferior vena cava (arrow)

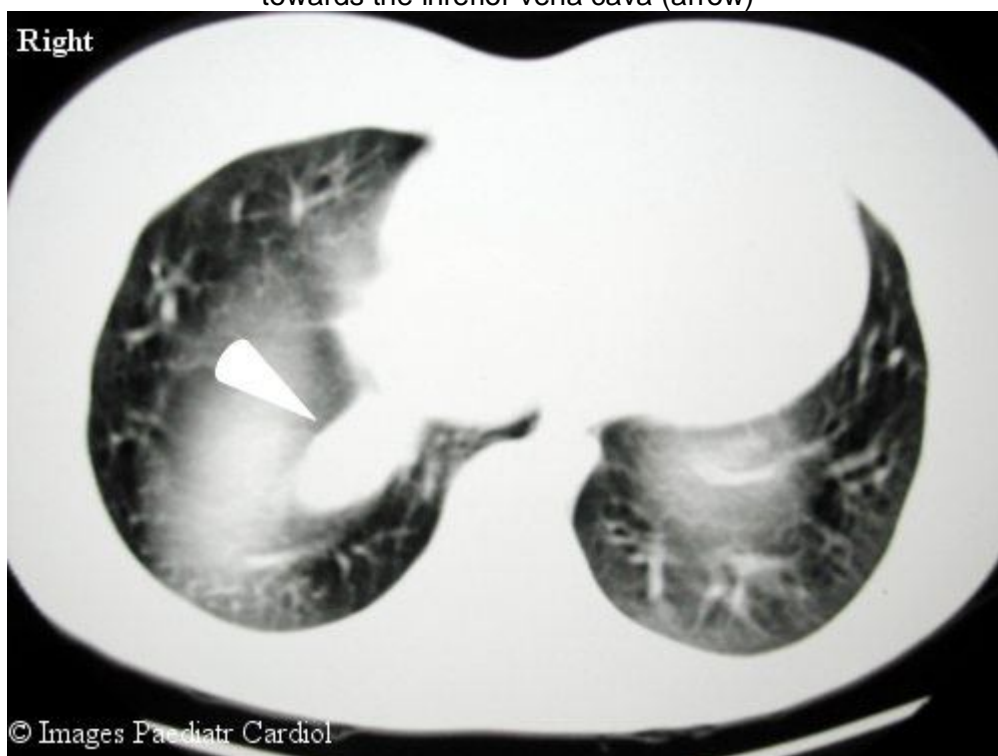
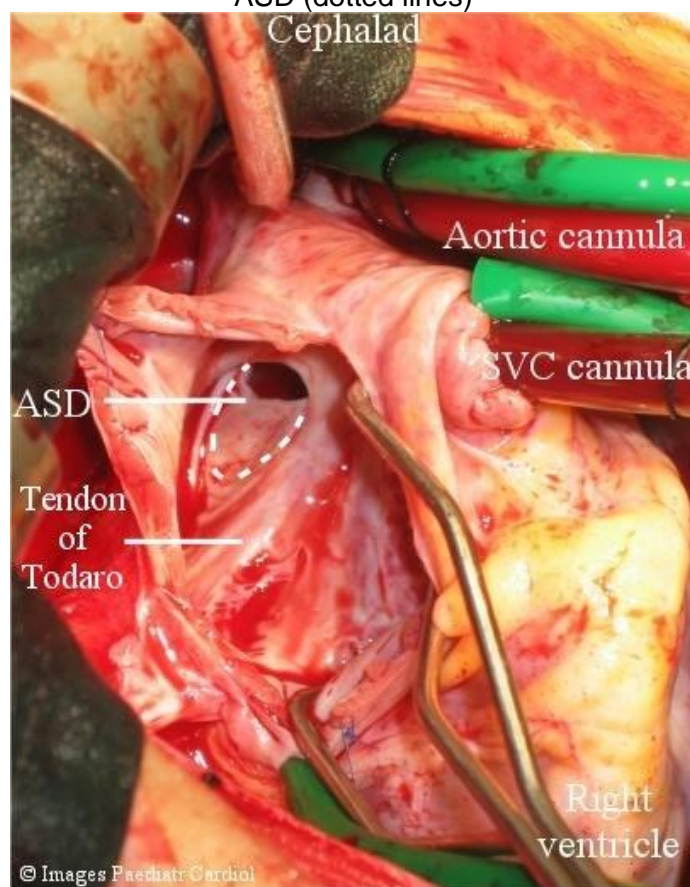


Figure 13 Right atriotomy showing atrial septal defect (ASD) and margins of enlarged ASD (dotted lines)



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Figure 14 Right atriotomy with more inferior exposure showing scimitar vein opening

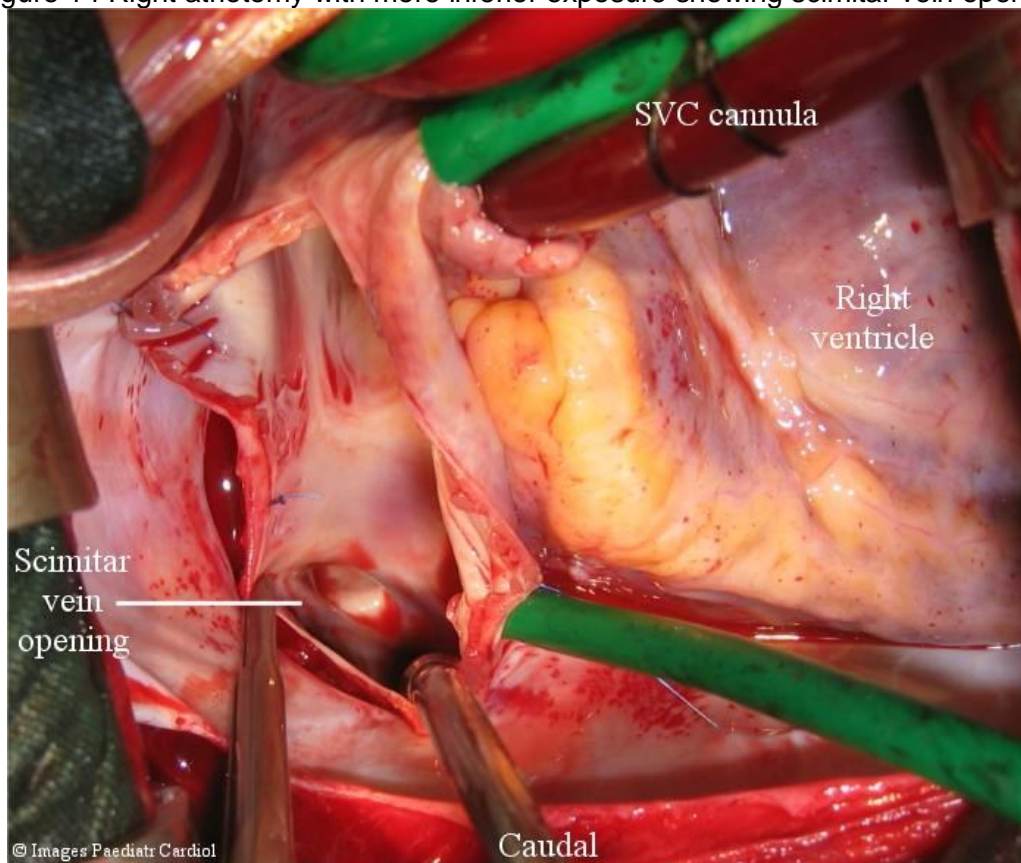


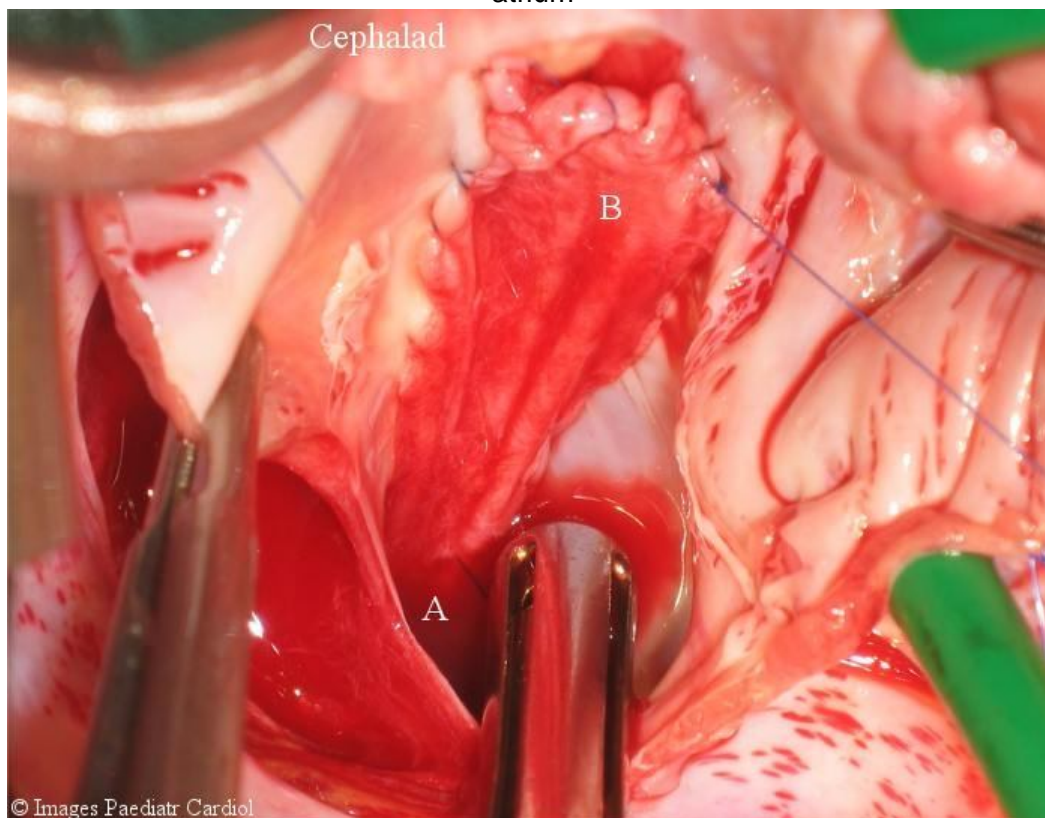
Figure 15 Freshly harvested autologous pericardial patch





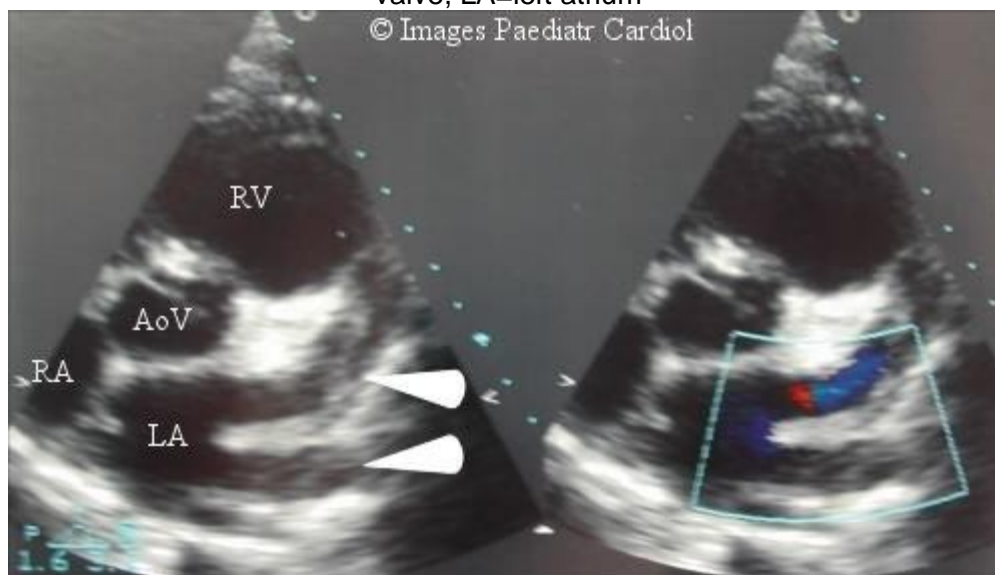
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Figure 16 Right atriotomy showing pericardial patch baffling pulmonary venous return from scimitar vein opening (A) to atrial septal defect (B) therefore into the left atrium



The patient had an uneventful postoperative course. The predischarge echocardiogram showed residual dilatation of the pulmonary veins and right ventricle and unobstructed baffle flow (figures 17-20).

Figure 17 Parasternal short axis view showing dilated left pulmonary veins (arrows). The right ventricle is also dilated. RA=right atrium, RV=right ventricle, AoV=aortic valve, LA=left atrium



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Figure 18 Apical four chamber view showing baffle on its way to ASD. B=baffle, RA=right atrium, TV=tricuspid valve, RV=right ventricle, CS=coronary sinus

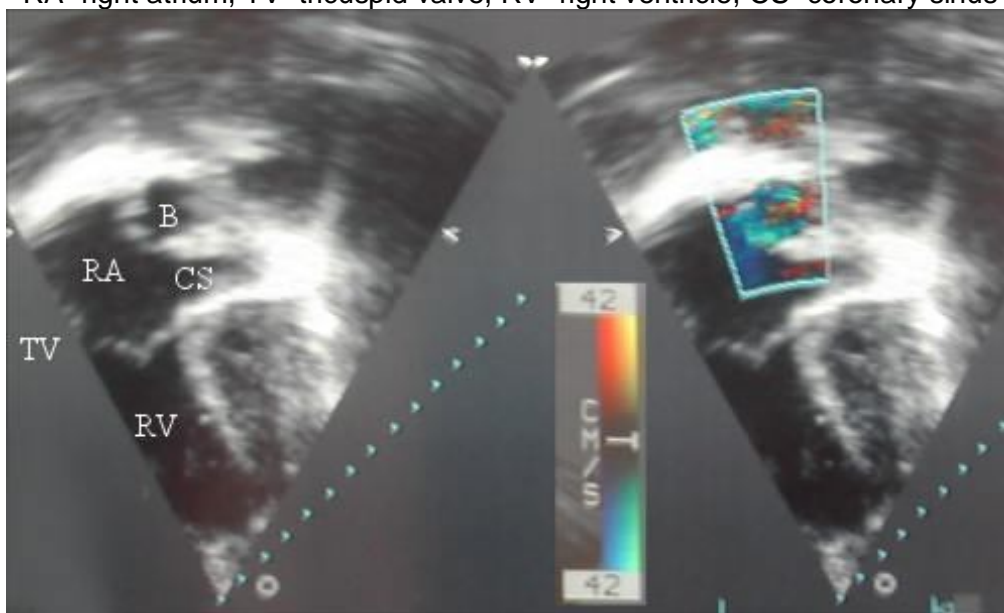
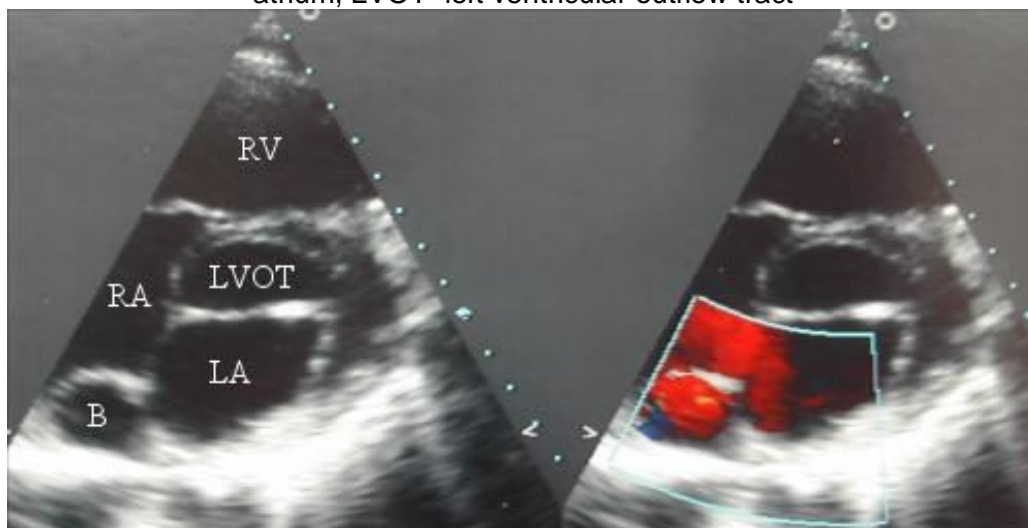
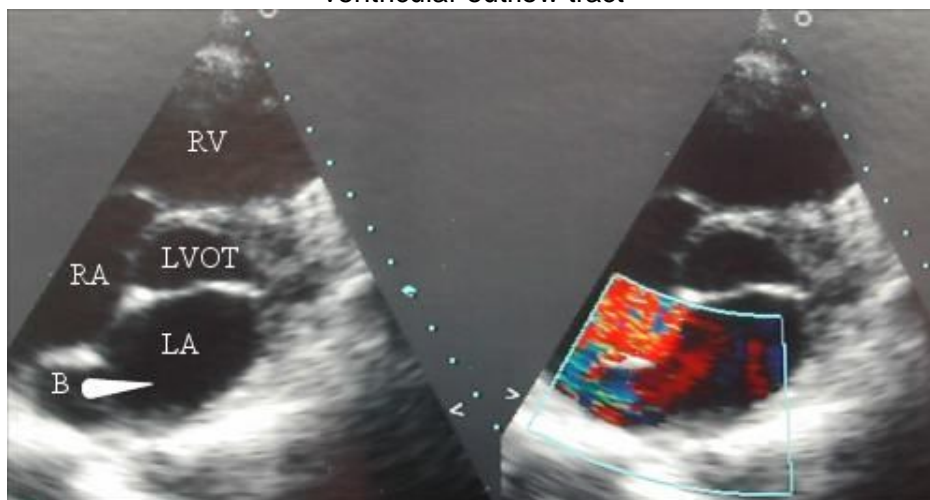


Figure 19 Parasternal short axis view showing baffle just below the level where it drains into the left atrium. B=baffle, RA=right atrium, RV=right ventricle, LA=left atrium, LVOT=left ventricular outflow tract



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Figure 20 Parasternal short axis view showing baffle draining into the left atrium (arrow). B=baffle, RA=right atrium, RV=right ventricle, LA=left atrium, LVOT=left ventricular outflow tract



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