

Case Report

Simple Radiography can Rescue from Dilemma

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Abstract:

24 year old young lady came to us with a diagnosis of atrial septal defect (ASD) secundum who lost her follow up since her 4 years of age. Later her history, physical findings and investigations change the diagnosis into supracardiac total anomalous pulmonary venous drainage. Typical 'snowman sign' in chest x-ray and transthoracic echo helped to review the case. Proper history taking and careful evaluation by investigation is needful for each individual.

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Key Words :

Anomalous pulmonary venous return, Snowman sign, Chest x-ray, Congenital heart disease.

Introduction:

Congenital heart disease has a wide range of expression that vary from patient to patient. Patient's own complaints associated with physical examination carry significant information and can add a diagnosis with current one.

Case presentation:

24 year old young lady came to us with the complaints of respiratory distress following heavy work. She was diagnosed as ASD secundum since her 4 years of age but was not properly advised about her disease, treatment and outcome and lost her follow up. When she came to us, we re-evaluate her clinically and few investigations were done. On physical examination we found clubbing with desaturation (SPO2 86%).

There was ejection systolic murmur over upper left sternal border and prominent S2. Her ECG showed

normal sinus rhythm, rate 75/min, incomplete right bundle branch block. Then a chest x-ray advised which turned the diagnosis into a new dimension. There was cardiomegaly with snowman sign or figure of 8 and dilated base of heart.

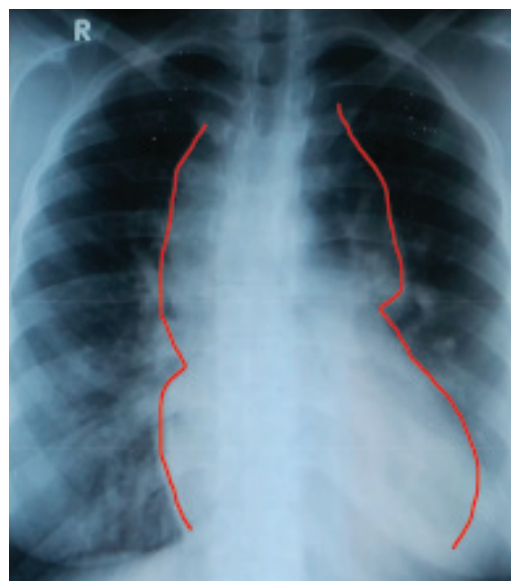


Fig 2: Frontal CXR depict a snowman sign or figure of 8 heart or cottage loaf heart. The dilated vertical vein on the left, brachiocephalic vein on top, and the superior vena cava on the right form the head of the snowman; the body of the snowman is formed by enlarged right atrium.



Fig.-1: Cyanosis & clubbing in upper nailbed.

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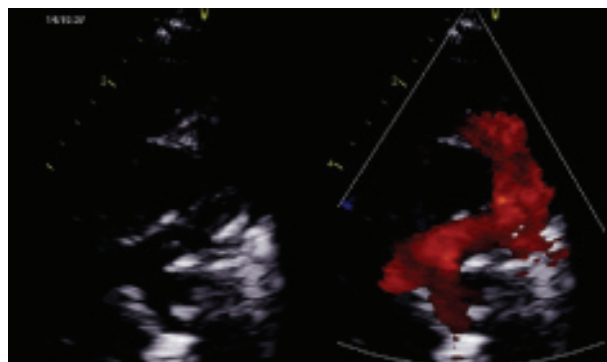


Fig.-3: *Supra sternal view showed all pulmonary veins not draining to left atrium rather forming a confluence & draining to vertical vein*

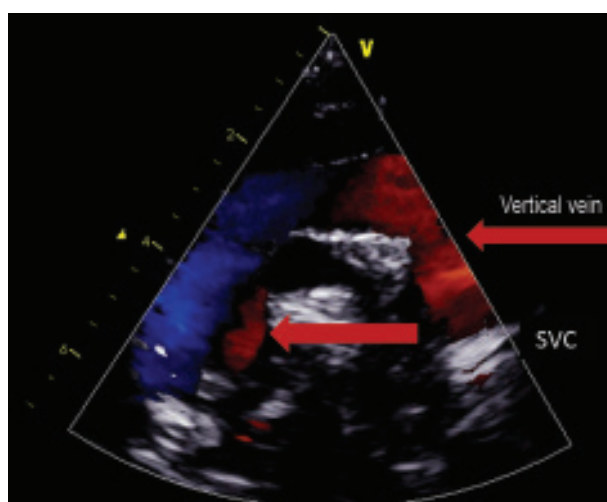


Fig.-4: *Supra sternal view showed vertical vein draining to Superior vena cava to right atrium.*

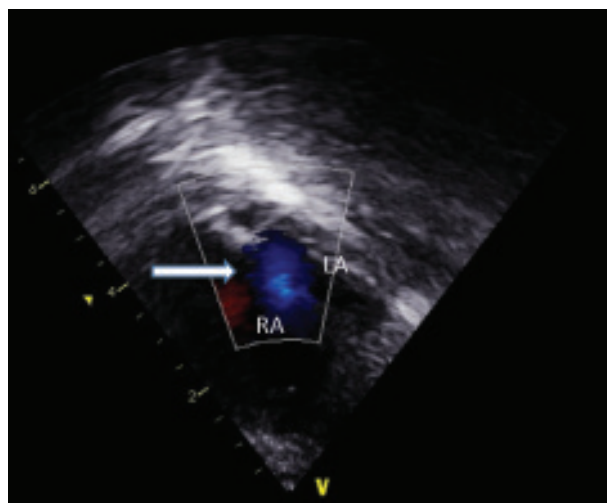


Fig.-5: *Sub costal bicaval view showed atrial septal defect with right to left shunt.*

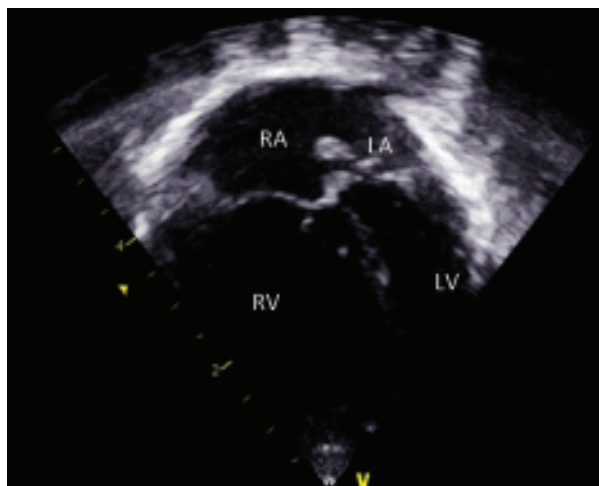


Fig.-6: *Apical four chamber view showed dilated right atrium & ventricle with small left atrium & ventricle.*

Then a repeat echocardiography done and found her all pulmonary vein forming a confluence draining into a vertical vein then through SVC draining into RA. Atrial septal defect at secundum location, (9X 10) mm with right to left shunt with moderate pulmonary hypertension & dilated right atrium, ventricle.

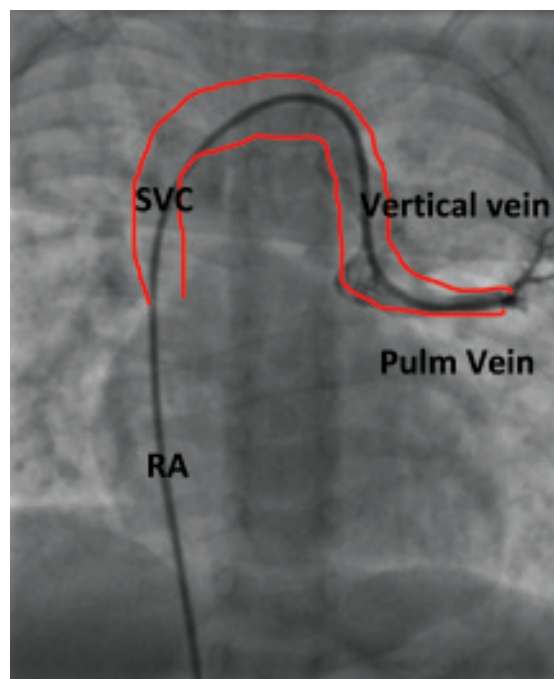


Fig.-7: *Cardiac catheterization showed all pulmonary veins draining to vertical vein to SVC to RA.*

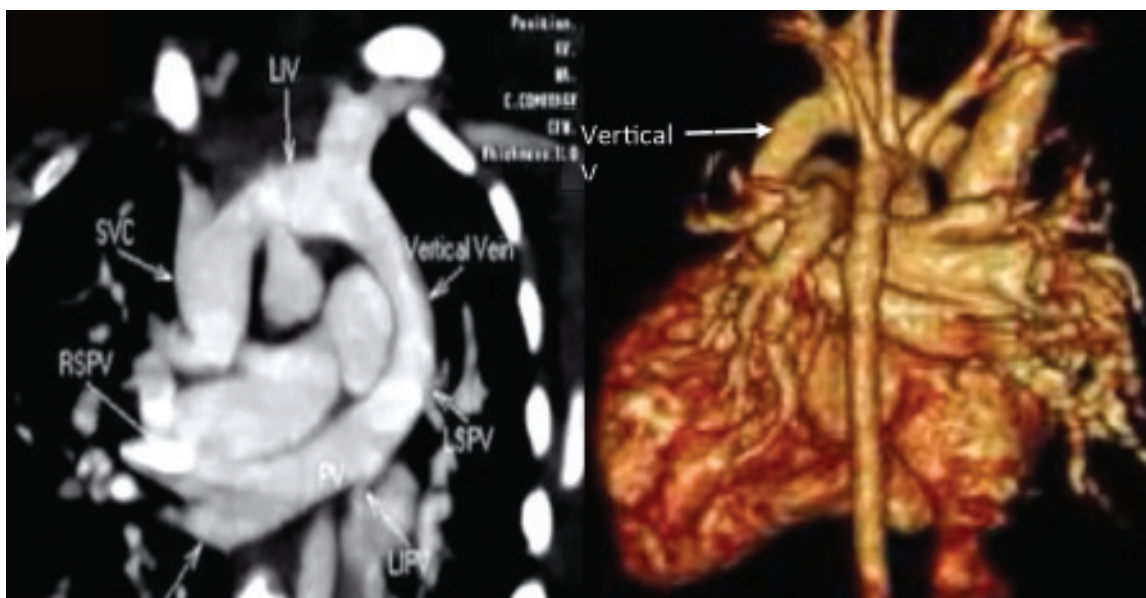


Fig 8: CT angiogram showed all pulmonary veins draining to vertical vein to SVC to RA.

Cardiac catheterization done to see pulmonary vascular resistance and to rule out other associated cardiac anomalies. Finally, she was diagnosed as Supracardiac Total anomalous pulmonary venous drainage (TAPVC) with moderate pulmonary hypertension with unobstructed pulmonary & vertical venous drainage and referred to cardiac surgeon for surgical correction.

Her all pulmonary veins rerouted to LA and ASD closed. Her postoperative period was uneventful and discharged with a good hemodynamics. She is now on regular follow up through our OPD and having a healthier life.

Discussion:

There are various types of anomalous pulmonary venous drainage like partial anomalous pulmonary venous drainage (PAPVC), scimitar syndrome, total anomalous pulmonary venous drainage. TAPVC means total anomalous pulmonary venous drainage. There are 4 types of TAPVC, a. supracardiac b. infracardiac c. cardiac and d. mixed.¹ Complex pulmonary venous malformation has an incidence of 1-3 per 100,000 and 2:1 female preponderance.²

TAPVC accounts for 1% of all congenital heart disease, and supracardiac variety is 50% of all kinds of TAPVC.¹ Our patient was initially diagnosed as ASD secundum with right to left shunt. As she had respiratory distress following heavy work and presented with clubbing, our suspicion was increased

pulmonary arterial pressure and reverse shunt. But her chest x-ray did not match with her initial diagnosis which showed snowman sign suggestive of total anomalous pulmonary venous return. Chest x-ray is simple, cheap, reproducible which provides diagnostic information.³ Posterior-anterior view chest x-ray with or without lateral view provide a valuable and inclusive single-image overview of heart, mediastinum, pulmonary vessel, lung fields and thoracic skeleton. All of which are potentially relevant to adult congenital heart disease.⁴ Radiographically a clue can be found about presence of abnormal pulmonary vein like scimitar syndrome (Turkish sword), supracardiac TAPVC (snowman).² Re-evaluated echo showed her all pulmonary veins forming a confluence draining into a vertical vein then through SVC to RA and an interatrial communication with right to left shunt. There was volume overload in RA and RV with pulmonary arterial hypertension. Sign of right ventricular volume overload is common features to all forms of TAPVC.⁵ In case of supracardiac TAPVC the pathway for pulmonary venous confluence can be imaged through suprasternal notch short axis view. The most common site for connection is the left SVC (i.e. left vertical vein), with subsequent drainage to the dilated left innominate vein and right SVC.¹ Diagnostic accuracy of TAPVC through transthoracic echocardiography (TTEs) is 92.86%. The individual PV should be imaged from multiple

windows, but the parasternal, subcostal, suprasternal notch view mostly are used⁶. TAPVC may remain undetected upto 60 years old with several TTEs performed at multiple institution. Literature confirms the difficulties in complete assessment of PVs in TTEs.⁷ Differential diagnosis of TAPVC includes ASD secundum, common atrium, PAPVC. In older patient mild cyanosis is present in case of TAPVC but this presentation is absent in case of ASD secundum and PAPVC unless there is associated pulmonary hypertension⁸. Depending on pulmonary venous obstruction there are two types of TAPVC. a. TAPVC with pulmonary venous obstruction and b. TAPVC without pulmonary venous obstruction.¹ Patient with venous obstruction present earlier and without treatment 50% patient died within 3 months and 80% patient died within 1 year. Without pulmonary venous obstruction two third patient died reaching 1st year. The patient who survive infancy may develop increase pulmonary resistance.⁸ TAPVC without pulmonary venous obstruction with a large interatrial shunt may present as a large ASD secundum that was presentation of our patient. Cardiac cath has role to calculate pulmonary vascular resistance and any obstruction in pulmonary vein and vertical vein. Our patient is 24 years old, so we did cardiac catheterization to see those things. We found low pulmonary pressure (systolic 30mmHg, diastolic 10 mmHg, mean pressure 16 mmHg) and low pulmonary vascular resistance (0.89 wood unit) which was exception for this age and disease.

All patients are not lucky enough like this patient as many of their life ended with a tragic event.

Conclusion:

Co-relation of patient's symptom and physical examination finding with investigation help to reach into a diagnosis with less deviation that also create better treatment option.

Conflict of Interest - None.

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