

CASE REPORT

Total Anomalous Pulmonary Venous Connection (TAPVC) with Severe Pulmonary Hypertension*Virendra C. Patil**Department of Medicine, Krishna Institute of Medical Sciences Deemed University (KIMSUDU),
Dhebewadi Road, Malkapur- 415539, Karad (Maharashtra) India***Abstract:**

Here we are reporting echocardiographic findings of 8 month child with Total Anomalous Pulmonary Venous Connection (TAPVC). All pulmonary veins are draining into a venous chamber (venous confluence) posterior to Left Atrium (LA). This chamber is draining in to persistent left superior vena cava (supradiaphragmatic type) with moderate (TR) and estimated (RV) systolic pressure was 88.2 mm of Hg suggestive of severe pulmonary hypertension.

Keywords: Total Anomalous Pulmonary Venous Connection

Echocardiogram and Doppler Report**Summary:**

Visceroatrial situs solitus with cardiac apex was towards left. Atrio-ventricular and ventriculo-arterial connections were concordant. All pulmonary veins were seen draining into a Venous Chamber (VC) or venous confluence posterior to left atrium (LA). This chamber was draining in to persistent left superior vena cava [supradiaphragmatic type] Normally draining systemic veins with two atria and two ventricular chambers were seen. Inter Atrial Septum (IAS) shows a small ASD with right to left flow. Interventricular Septum (IVS) was intact. Mitral valve and tricuspid valve were seen opening normally. Moderate Tricuspid Regurgitation (TR) was noticed, estimated Right Ventricular Systolic Pressure (RVSP) was 88.2 mm of Hg. There was

no Mitral Regurgitation (MR). Right Atrium (RA) and Right Ventricle (RV) were moderately dilated. Pulmonary Valve (PV) was normal with trivial PR. Main Pulmonary Artery (MPA) and its branches were dilated. There was no PDA flow. Aortic valve was normal. Aortic arch was left sided with no coarctation. There was no Pericardial Effusion (PE).

Impression:

Congenital cyanotic heart disease with supradiaphragmatic type: Total Anomalous Pulmonary Venous Connection (TAPVC) with severe pulmonary hypertension. [Abbreviations: PLAX: Parasternal Long Axis, PSAX: Parasternal Short Axis, CFM: Colour Flow Map, CW: Continuous Wave].

Fig.1A: Subcostal view showing CFM across ASD scundum type with all pulmonary veins are seen draining into a venous chamber posterior to LA [venous confluence].

Fig. 1B: Subcostal view showing ASD scundum type with pulmonary veins are seen draining into a venous chamber posterior to LA (venous confluence) with dilated RA and RV.

Fig. 1C: Subcostal view showing CFM with CW with moderate TR with PAP:88.2 mmHg.

Fig. 1D: PSAX at aortic level view showing CFM with CW across pulmonary valve and main pulmonary artery.

Fig. 1E: PSAX view showing dilated RV with flattened IVS with normal size LV.

Fig.1F: Suprasternal view- CFM & CW with Venous confluence draining in to left Superior

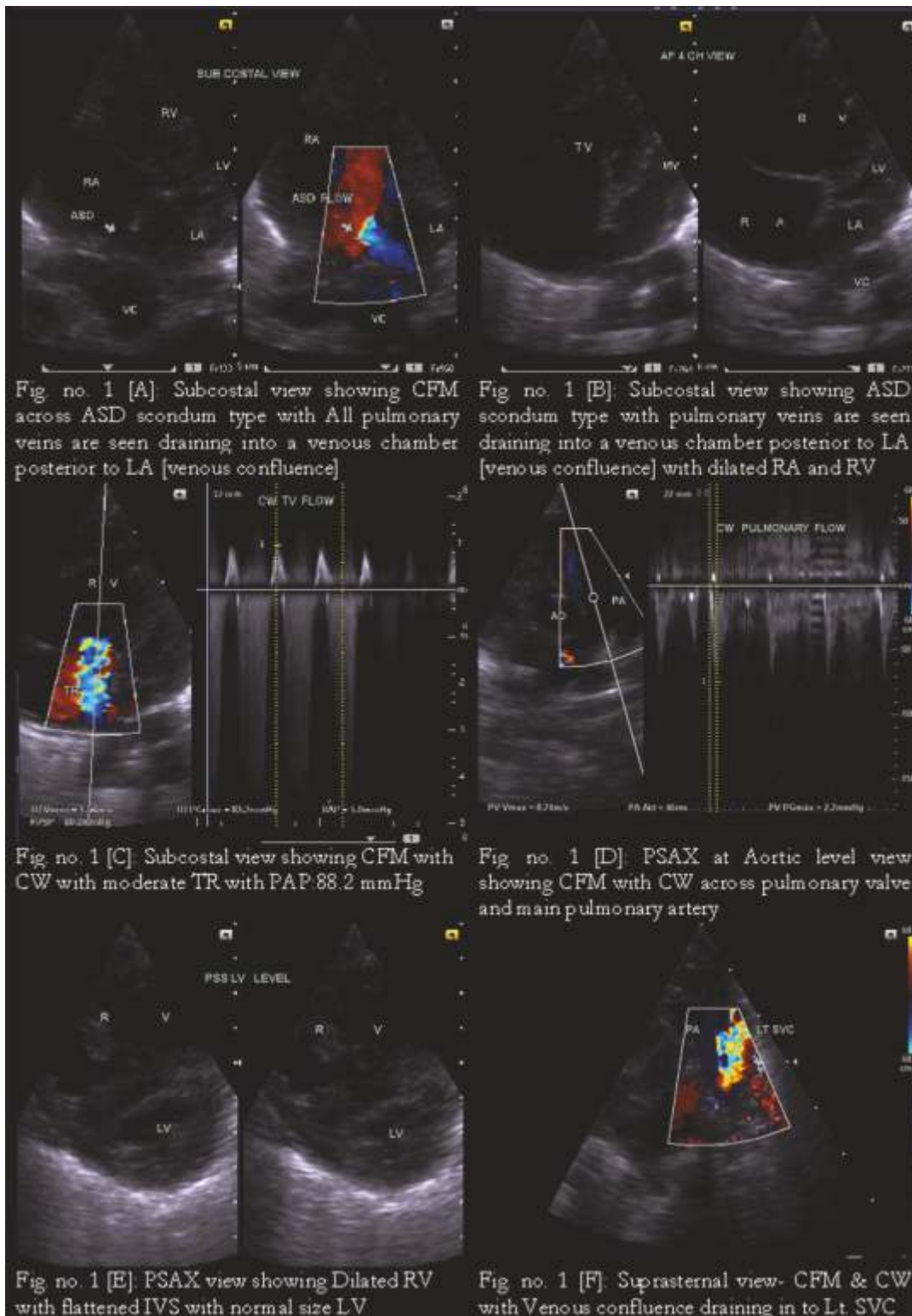


Fig. 1: Echocardiogram and Doppler Report of TAPVC

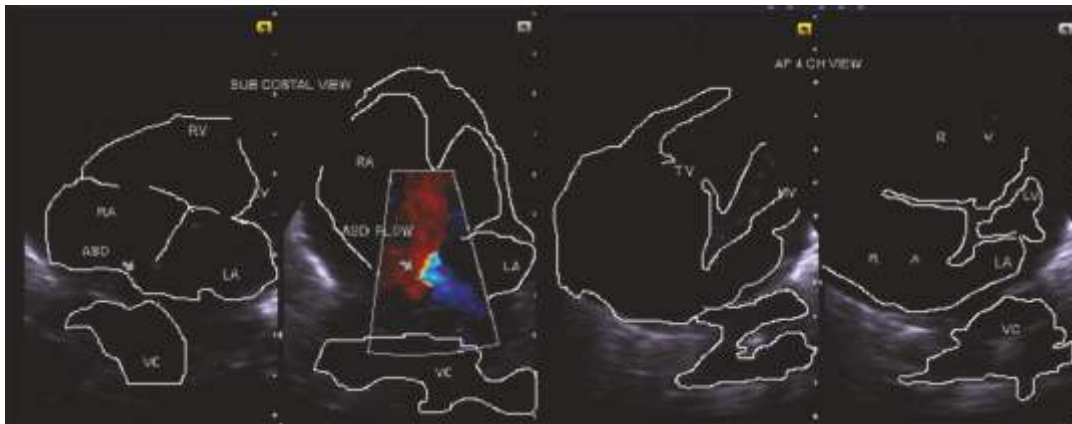


Fig. 2: Schematic Echocardiogram Image Presentation of TAPVC

Vena Cava (SVC).

Fig. 2: Schematic presentation of TAPVC showing ASD scndum type with All pulmonary veins are seen draining into a venous chamber posterior to LA [venous confluence] with no connection of VC to LA. [Echocardiogram and Doppler imaging was done with Siemens Accuson–X-300 with 4-8 MHz probe].

Introduction:

TAPVC or Total Anomalous Pulmonary Venous Return (TAPVR) is a congenital heart disease in which there is no connection between the pulmonary veins and the left atrium [1]. It is a relatively uncommon congenital defect representing approximately 2% of all congenital heart anomalies. It is found in approximately 7 persons in 100,000 populations. It is associated with atrial septal defect, making compatible to life.

Pathophysiology:

The diagnosis of TAPVC is made when all four pulmonary veins drain anomalously to the right atrium or to a tributary of the systemic veins [2]. An anomalous connection is established that allows the pulmonary veins to deliver pulmonary venous blood to the right side of the heart rather than to the left side. A patent foramen ovale or an

atrial septal defect develops in utero, allowing a right-to-left shunting of blood without which the infant would die [1].

Types of TAPVC:

It can be categorized by the site of drainage into the systemic circulation (supracardiac, 45%; infracardiac, 25%; cardiac, 25%; mixed, 5%) [2]. The clinical presentation is different if the pulmonary venous drainage is unobstructed (heart failure, mild cyanosis) or obstructed (respiratory failure, severe heart failure).

Diagnosis:

The diagnosis is usually established by two-dimensional echocardiography [3]. TAPVC on chest radiograph shows bulging of the superior mediastinum bilaterally, producing a 'snowman' contour' or 'figure of eight'. TAPVC on Electrocardiogram shows evidence of right axis deviation and right ventricular hypertrophy. Surgical management depends on the anatomic type.

Management:

Obstructed TAPVC requires urgent surgical intervention, whereas unobstructed TAPVC can be dealt with electively; although this is usually operated on once the diagnosis is made. Postoperative pulmonary artery hypertension can

be problematic. Recent surgical results with isolated TAPVC have improved, with operative mortality consistently at less than 10%. A particularly challenging group of patients are those with single ventricle physiology and TAPVC with high operative mortality and poor long-term survival [4]. The severity of symptoms in TAPVC depends primarily on the degree of Pulmonary Venous Obstruction (PVO). Patient with severe PVO, patients can present with severe

cyanosis, respiratory distress, and acidosis within the first few hours of life. Surgical repair of TAPVC has involved side-to-side anastomosis between the confluence of pulmonary veins and the left atrium by right side approach [5]. The operative methods of TAPVC depend on the cardiac deformation [6]. Prompt diagnosis proper surgery and timely treatment of the postoperative complications will achieve a satisfactory outcome.

References

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