Two Dimensional Sector Scan Echocardiographic Profile of Total Anomalous Pulmonary Venous Connections.

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SUMMARY :

Sixteen patients aged 3 days to 5 months are included in this study of Cardiac catheterization proven cases of TAPVC in order to validate 2-D Echocardiographic imaging in this condition. It is shown that 2-D Echocardiography provides detailed anatomic imaging of CPVC and its afferent and efferent connections in various types of TAPVC. The sensitivity of echo diagnosis largely depends upon the skill and knowledge of the echocardiographer.

Diagnostic M-mode echocardiographic features of total anomalous pulmonary venous connections (TAPVC) have been reported (1-2). The diagnosis is primarily based on demonstration of signs of right ventricular volume overload and visualization of anomalous linear echo, with its characteristic motion pattern, behind the left and right atrium representing common pulmonary venous chamber (CPVC) 1-2.. When Pulmonary veins do not drain normally into the left atrium, they form anomalous venous channels which then return to either supra cardiac or infra cardiac or rarely intra cardiac sites. Anatomically anomalous right and left pulmonary veins become confluent and form a common pulmonary venous chamber (CPVC) i.e. afferent pulmonary venous connections. The position of CPVC is usually postero-superior to the left atrium in supra cardiac connections (1) and directly posterior to the left atrium in coronary sinus type of connections (3). Uncommonly CPVC directly communicates with the right atrium or rarely pulmonary veins may enter the right atrium individually without a common venous confluence chamber. From the CPVC a channel called vertical vein either ascends left ward to join the left innominate vein and right superior vena cava or descends down ward and pass through the diaphragm and joins inferior vena caval system at the porta hepatis i.e. Efferent connections. Uncommonly the ascending vertical vein may join the right superior vena cava directly. These connections show consierable anatomic variation Pulmonary venous connections of the CPVC (afferent connections) and the cardiac connections of the CPVC i.v. site of drainage (efferent connections) can not be visualized by single crystal echocardiography. Two dimensional sector scan echocardiography allows for visualization of these additional anatomic structures (3-4-5-6). Echo imaging of the afferent pulmonary venous connections of CPVC and efferent connections via ventrical vein to the infradiaphragmatic sites have been reported (3-5-6). The afferent connections to the CPVC and efferent to the supra-cardiac site and to the coronary sinus have also been recently reported (3-4-5-6).

We present our experience with 16 patients who had total anomalous pulmonary venous connections (TAPVC), in whom prospective diagnosis was made by two dimensional sector scan echocardiography, with a view to discuss the diagnostic two dimensional echocardiographic features of TAPVC.

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Material and Methods :

Sixteen patients age 3 days to 5 months were included in the study. Fourteen patients were studied at the children's memorial Hospital Chicago, U.S.A. and two patients at the National Institute of Cardiovascular Diseases, Karachi, Pakistan.

Two dimensional sector scan studies were performed in supine positin using an advanced technology laboratories (ATL) sector scanner and Varian Associates Model 3000 phased array system. The cardiac imaging was done by using a 3.5 and 5.0 MHz transducers, positioned at the supra sternal, subxiphoid, apical and parasternal locations and by utilizing various planner projections (7). The diagnosis of TAPVC was prospectively made in all patients and confirmed by cardiac catheterization. In order to validate echo imaging, echo contrast studies were obtained during cardiac catheterization in two patients who had anomalous venous connections to the right superior vena cava.

Results :

The site of anomalous venous connections was determined by angiography in all 16 patients. In 9/16 the anomalous venous connections were into the innominate-superior vena cava system and in 4/6 were solely into the coronary sinus. In three remaining patients anomalous venous connections were partially into the superior vena cava and paritally into the coronary sinus. There was no instance of anomalous pulmonary venous connection below the diaphragm. Prospective 2D-Echocardiographic diagnosis of anomalous venous connections was made in 13/16 patients in whom these connections were soley to the superior vena cava or coronary sinus. In the remaining three patients although diag nosis of anomalous venous connections could be made but the exact site of anomalous venous connections could not be determined.

Imaging of CPVC :

Echocardiographically imaging of the CPVC is the diagnostic hall mark of anomalous venous connections. The CPVC was imaged directly posterior to the left atrium in 4/16 patients in whom anomalous connections were to the coronary sinus. It was located at the atrio ventricular Junction in the long axis left parasternal view. (Fig. 1). The superior extension of the CPVC behind the left atrium varied as did the concava posterior deformity of the left atrium. Confirmation of the position of the CPVC at the atrio ventricular junction could be obtained in four chamber apical and subxiphoid views as well as high left parasternal view (Fig. 2-3). No separate coronary sinus was visualised at the atrio ventricular junction in these patients. From the supra sternal location the enlarged coronary sinus was imaged in one patient. The transducer was placed in supra sternal notch with echo beam in the coronal plane. The aorta was sectioned in the short axis and was imaged as a circular echo and inferiorly the right pulmonary artery was sectioned in long axis. Superior vena cava was imaged in long axis on the right side of the aortic arch echo. The transducer was then angled slightly posteriorly. Both right and left pulmonary veins were imaged entering a pear shaped coronary sinus below the pulmonary artery (Fig. 4).

In 9/14 patients in whom angiographically the anomalous venous connections were into the left innominate-right superior vena cava system, the CPVC imaged postero-superior to the left atrium away from the atrio-ventricular Junction in the parasternal long axis view. In the high left

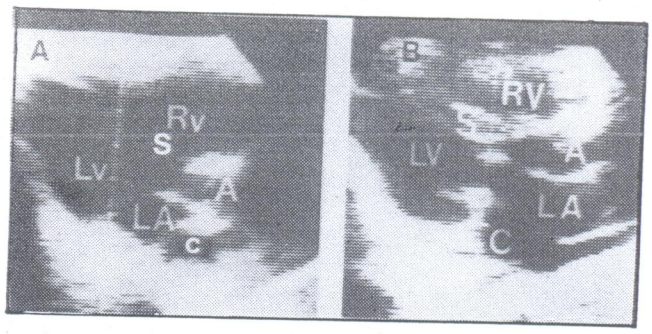

FIGURE NO. 1.

A. Two dimensional sector scan echocardiacgraphic long axis view obtained from left para sternal location in a patient with total anomalous pulmonary venous connections to innominate-superior vena Caval system. The Common pulmonary venous chamber (C) is located postero-superior to the left atrium (LA). A = Aorta, LV = Left Ventricle, RV = Right Ventricle, S = Septum.

B. Long axis echo view from left parasternal location of a patient with anamolous pulmonary venous connections to Coronary sinus. Common pulmonary venous chamber (C) is located at the atrio-ventricular junction and indents the left atrium posteriorly. This location suggests that C is an enlarged Coronary sinus.

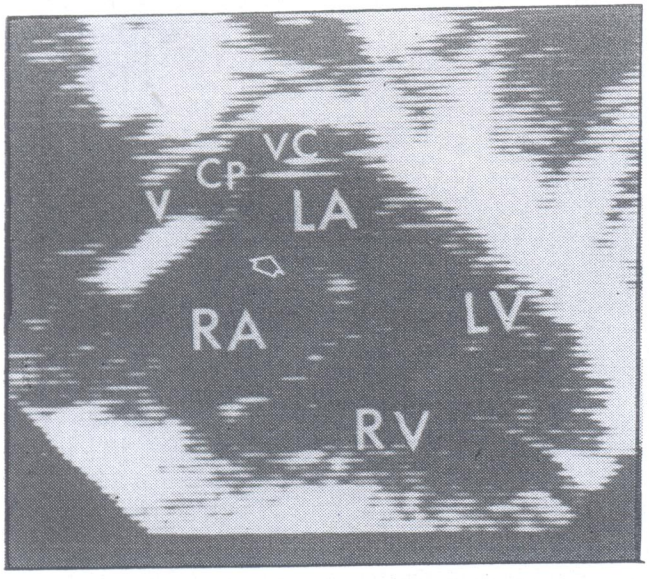

FIGURE NO. 2.

From subxiphoid location a four chamber view sector scan in a patient with anomalous pulmonary venous connections to superior vena cava. The common pulmonary venous chamber (CPVC) lies postero-superior to the left atrium (LA). A moderate size secundum atrial septal defect is present (arrow), and right ventricle (RV) is markedly enlarged. Right and left pulmonary veins are well seen. Right pulmonary vein (V) lies closer to the atrial septum and extends postriorly.

parasternal view the dominant superior location of the CPVC vis-a-vis left atrium was confirmed. (Fig. 3). Apical four chamber and subxiphoid for chambers views provided confirmation of the position of the CPVC.

The CPVC was also visulaised, in all 9/14 patients with supra cardiac anomalous conneections, from the supra sternal location with echo beam in the coronal plane. It was imaged as a transverse structure below the right pulmonary artery and was only occasionally clearly demarated from the left atrium in one section (Fig. 5).

Imaging of Afferent Pulmonary Venous Connections of the CPVC.

The search began by locating the CPVC and then searching for pulmonary veins. In apical and subxiphoid four chamber views right veins were seen close to the atrial septum and extended behind the right atrium. Left pulmonary veins were more posteriorly situated (Fig. 2). These connections could also be imaged from the

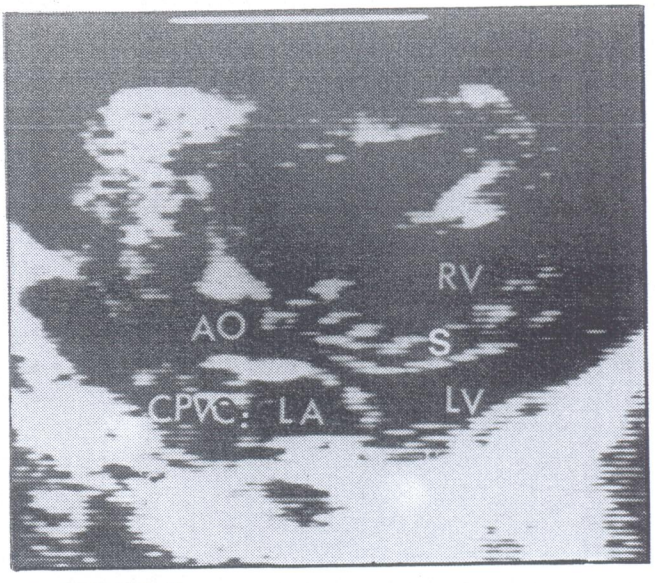

FIGURE NO. 3.

Sector scan long axis view, obtained from high left parasternal location in a patient with anomalous pulmonary venous connections to superior vena cava. Right ventricle (RV) is markedly enlarged while left atrium (LA) and left ventricle are small. The CPVC lies superior and slightly posterior to the left atrium (LA). AO = Aorta.

suprasternal location with echo beam in the coronal plane. These connection were imaged in all patients in whom anomalous connections were to the supra cardiac site and in one patient with anomalous connection into the coronary sinus (Fig. 4). From the supra sternal location the left pulmonary veins were less often imaged with certainity. It was considered imperative that afferent venous connections of the CPVC are imaged from any of the locations.

Imaging of efferent connections of CPVC :

The transducer was placed at the subxiphoid location and four chambers image was obtained, Superior vena cava was imaged by tilting the transducer superiorly and anteriorly in right ward direction of the patient. A dilated superior vena cava was imaged to the right of the ascending aorta and continued transversly into the innominate vein which at times was imaged as a continuous structure with the left vertical vein and CPVC. The right atrium — superior vena caval connections were imaged in all patients. The transducer was then rotated in sagittal plane and inferior vena cava, right atrium and abdominal aorta were imaged and search for descending anomalous

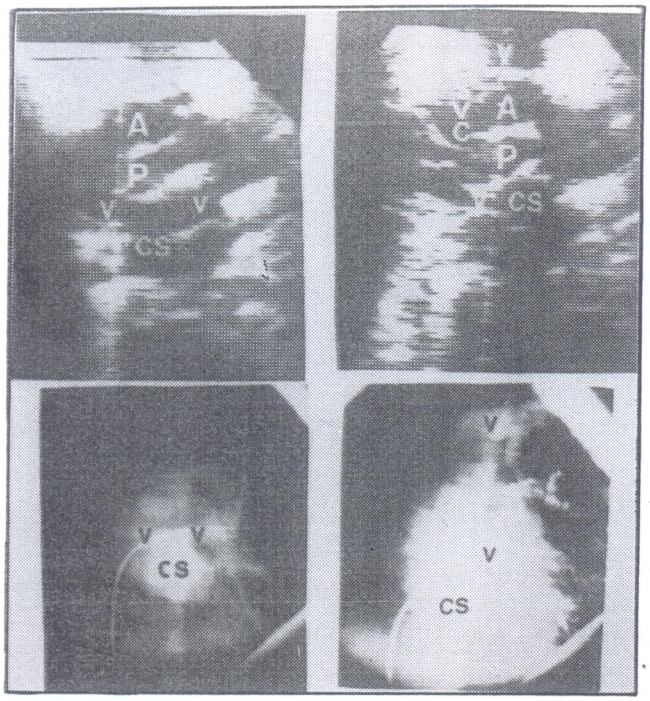

FIGURE NO. 4.

A-B. Sector scans from supra-sternal location with echo beam in Coronal body plane.

A. Is a cut at a more posterior plane than B and shows right and left pulmonary veins (V) entering a pear shaped enlarged Coronary sinus.

B. Aortic arch (A) is sectioned in short axis and a large transverse channel (V) is imaged superior to A which represents a dilated left innominate vein due to partial anomalous venous connections of left upper pulmonary veins. Enlarged superior vena Cava (SVC) is imaged in long axis on the right side of A. The right pulmonary artery (P) is below the A and is cut in its long axis. The section is at a slightly posterior plane to the LA and pulmonary veins (V) are seen entering a large coronary sinus (CS).

C. Correlative cine angiogram in AP view. All pulmonary viens from the right lung and lower lobe of left lung are seen entering the coronary sinus.

D. Partial anomalous venous connections of the left upper lobe pulmonary veins to the innominate vein are seen.

vertical vein was made in all cases. Complete supra cardiac efferent connections could not be imaged from subxiphoid location in all patients. From suprasternal location imaging of CPVC and its afferent and Efferent connections was

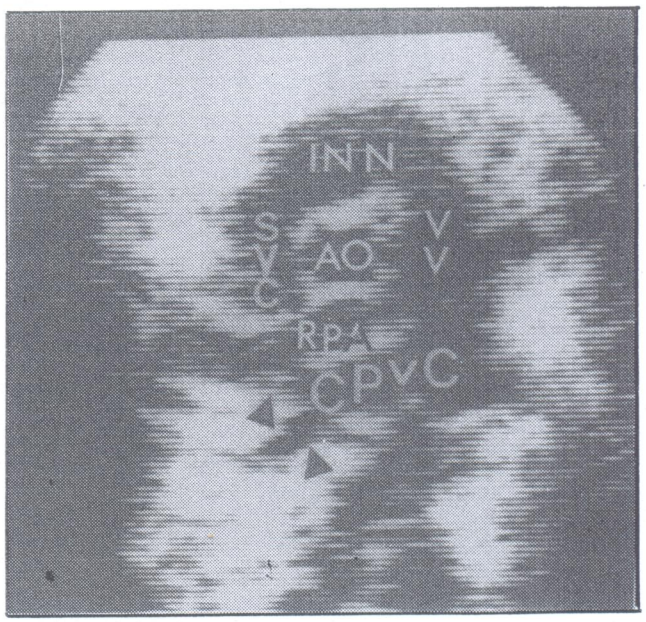

FIGURE NO. 5.

Sector scan obtained from supra sternal location with echo beam in the coronal body plane. CPVC and its afferent right pulmonary venous connections (arrows) are imaged below the right pulmonary artery (RPA). From the left side of the CPVC ascending. vertical vein (VV) innominate vein (Inn) and superior vena cava are imaged. AO = aortic arch.

obtained in all patients (9/14) with total anomalous venous connections to the superior vena caval system. The transducer was placed at the supra sternal notch with echo beam in the coronal body plane. The aortic arch in short axis, was echoed, as a circular image and right pulmonary in its long axis upto the right hilum. A dilated superior vena cava lay to the right of the aortic arch image and was connected to a markedly enlarged innominate vein. The vertical vein and CPVC were echoed as transverse structure below the right pulmonary artery. Minor adjustments of the transducer were required to image these structures; slightly posteriorly for imaging the CPVC, left ward for vertical vein — CPVC connections and right ward and anterior for SVC — right atrial conncetions. Rotation of the transducer from coronal to sagittal plane clearly distinguished superiorly situated innominate vein from inferiorly located aortic arch (Fig. 7). In this view innominate-SVC venous confluence was imaged as an arch convex antero-superiorly while aortic arch-descending aorta appeared as an arch convex postero-superiorly. Further

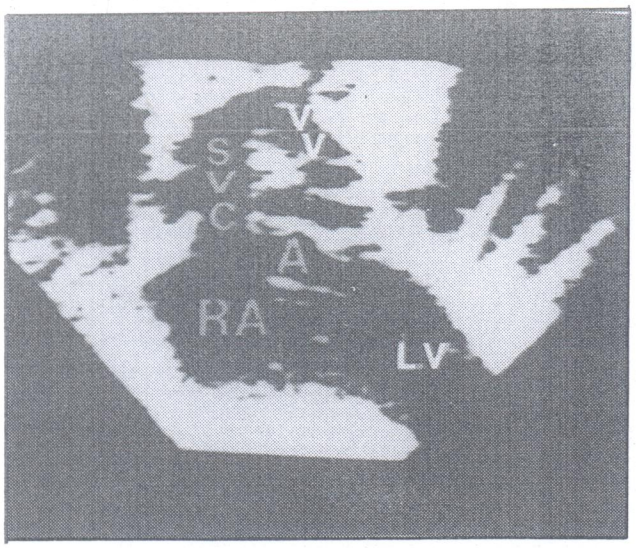

FIGURE NO. 6.

Long axis view obtained from subxiphoid location. The transducer is rotated right ward and superiorly from the four chamber position and the efferent connections of CPVC to the vertical vein (VV), innominate vein, superior vena cava (SVC) and right atrium (RA) are imaged. A = Aorta, LV = left ventricle.

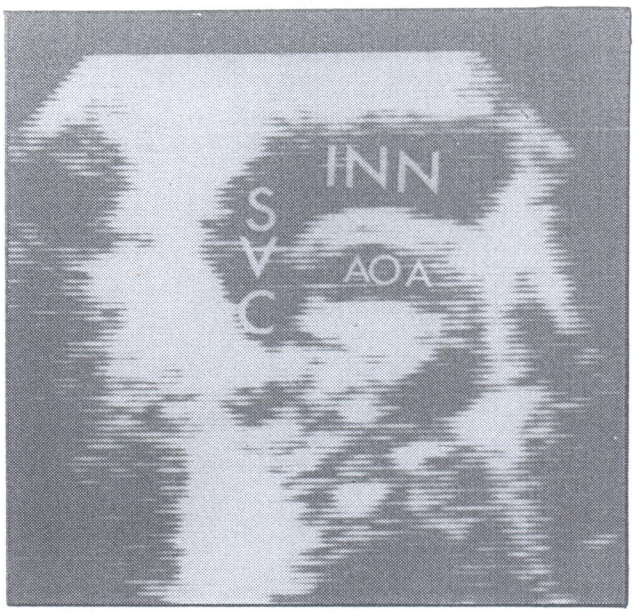

FIGURE NO. 7.

From the coronal plane in Fig. 5, the transducer is turned into Sagittal body plane. Superiorly markedly enlarged innominate vein and inferiorly, aortic arch and descending aorta are imaged. Aortic arch is convex postero-supriorly while innominate-SVC venous complex is convex antero-superiorly.

more arterial pulsations of the aortic arch image clearly distinguished it from the echoes of anomalous venous connections.

In one patient with split form of anomalous venous connections dilated innominate — superior vena cava complex was imaged along with both pulmonary veins which entered a pear shaped coronary sinus from (Fig. 4) superior direction.

Validation of Echo images

In order to validate the identification of echo images in two patients echo contrast studies were obtained. The transducer was placed in the subxiphoid location and right atrium superior vena cava-innominate connections were imaged. The catheter was placed in the innominate vein. Ten ml of 5% Dextrose was injected in the catheter and Echo opacification of the innominate vein SVC and right atrium was obtained thereby confirming the anatomic identification of echo images (Fig. 8).

Discussion :

This study documents two dimensional echocardiographic imaging of the CPVC and its afferent and efferent connections in the supra cardiac and coronary sinus types of total anomalous pulmonary venous connections. Prospective diagnosis of these two types could be made in all instances.

M-mode echocardiographic imaging of the common pulmonary venous chamber behind the left atrium, features of an enlarged and hypertrophied right ventricle and paradoxic ventricular septal motion are suggestive but not diagnostic of total anomalous venous connections. (1-2). Suprious echoes behind the left atrium can some times be seen in normal hearts and in heart with enlarged coronary sinus due to persistent left superior vena cava. Theefore on M-mode echocardiography distinction from total anomalous pulmonary venous connections may be difficult if not impossible (1). Two dimensional sector scan echocardiography however allows for imaging not only the CPVC but also its afferent and efferent connections.

Our study and the reported experience shows that the common pulmonary venous chamber can be imaged in long axis view from left parasternal and high left para sternal locations as well as in four chamber view from apical and subxiphoid

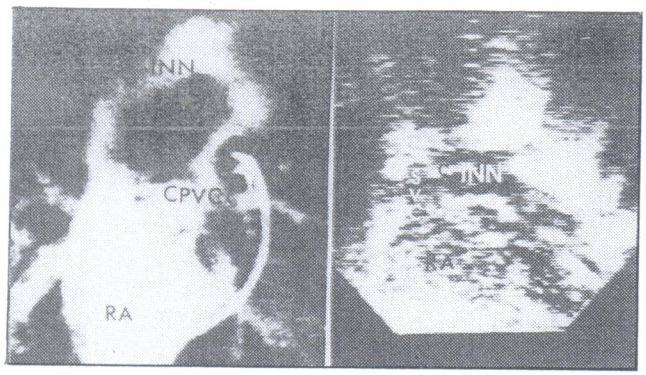

FIGURE NO. 8.

A. Pulmonary artery cine angiogram, AP view. On levophase CPVC, vertical vein and its connections to the innominate vein and superior vena cava are opacified.

B. Long axis view sector scan of the same patients as in panel A. The catheter (C) is imaged in the innominate vein (INN). Injection of 10 ml of 5% Dextrose in the innominate vein shows echo opacification of superior vena cava and right atrium thereby validating the anatomincal identification of echo images.

locations (3-6). By utilising all these images the position of CPVC, vis-a-vis left atrium, can be evaluated with precision. The CPVC is located postero-superior to the left atrium away from atrio-ventricular Junction in the supra cardiac type (1-3). If the Coronary sinus can be imaged in the atrio-ventricular groove, then the supra cardiac type of anomalous venous connections are most likely. In the Coronary sinus type of anomalous venous connections with CPVC is actually an enlarged coronary sinus and in the long axis or four chamber images it extends upto the atrio-ventricular groove. We have no experience of imaging a cor-triatrium heart but it seems likely that partitioning of the left atrium from anterior to posterior wall by the lower portin of pulmonary venous chamber in triatrium heart would distinguish it from the TPAVC where the CPVC lies posteriorly or postero superior to the posterior wall of left atrium, and produces an anteriorly convex deformity of the left atrium. It is therefore imperative that the anterior wall of the left atrium is shown to be free of the anterior wall of the CPVC echo image. The common pulmonary venous chamber can be imaged from supra sternal location with echo beam in the coronal plane (3). The transducer has then to be directed slightly posterior to the right pulmonary artery to image the CPVC. We were able to image an enlarged coronary sinus with its afferent pulmonary venous connections in one of our patients. How often this is possible remains to be determined.

Afferent connections of the CPVC.

In the supra and infra cardiac type of TAPVC the pulmonary veins enter the CPVC from right and left ward direction. Therefore echocardiographic images obtained from subxiphoid, apical and supra sternal location are most useful. From the supra sternal location echo beam has to be in the coronal plane. The CPVC then is seen as a horizontal chamber with left and right pulmonary veins entering it. The right pulmonary veins are most readily imaged compared to the left veins. Imaging of subxiphoid four chamber views not only establish the position of CPVC but also its venous connection in all cases.

Difficulties in imaging the pulmonary venous connections can be anticipated where all four pulmonry veins enter the right atrium directly without intervening common pulmonary venous chamber (4).

In the coronary sinus type we were able to image both right and left pulmonary venous connections as well as the enlarged coronary sinus from the supra sternal location. The sensitivity of this approach remains to be established.

Efferent connections of the CPVC.

In the supra cardiac form imaging of vertical vein-innominate vein-superior vena cava complex can be obtained in almost all cases from the supra sternal location (3).

Maredly enlarged innominate venous echoes lie superior to the circular aortic arch echoes with echo beam in coronal plane. The aortic arch echoes can be clearly distinguished because of their display of arterial pulsations. Furthermore the aortic arch echoes can be shown to continue into descending aorta image by rotating the transducer from Coronal to sagittal body plane. The enlarged innominate vein can be shown to the connected to superior vein Cava on the right and to vertical vein on the left. Although subxiphoid location can be utilised to image efferent connections of the CPVC, these are best imaged from supra sternal location (5).

We have not encountered a case of subdiaphragmatic type of TAPVC, however it is reported that CPVC and descending vertical vein can be imaged from supra sternal as well as subxiphoid

locations (3).

Inferior vena cava, oesophagus, aorta and vertical vein are closely related at the diaphragmatic site. Inter-structural relationship of vertical vein can very greatly. Usually Inferior vena cava lies to the right of Thoracic aorta and vertical vein is anterior to the aorta while oesophagus is the most posterior structure. Inferior vena cava connects to the right atrium superiorly while aorta is pulsatile. These features are helpful in identification of the abnormal vertical vein particularly when dopher flue studies can be combined with echoimaging (6).

Echo cardiographic diagnosis of TAPVC should be based on imaging of Common pulmonary venous chamber its afferent and efferent pulmonary venous Connections to the supra cardiac, intra cardiac and infra cardiac sites. Angiography is capable of demonsrating all these connections in one view, two domensional sector scan echo-cardiography, in our and reported experience (3), is capable of demonstrating these anamolous connections when various location and imaging planes are used. A step-wise approach is essential. Common pulmonary venous chamber is first visualised. Then its afferent venous connections are imaged from apical or subxiphoid sites, and as a third step supra cardiac efferent connections of the CPVC are searched from supra sternal location and confirmed from subxiphoid site. Infra cardiac efferent connections are looked for from subxiphoid and location and confirmed from supra cardiac site. The doppler technique has added another modality for distinguishing anomalous venous channel from various other normally present structures at the subxiphoid location (6). If this approach is strictly followed and all possible connections are searched, particularly when both afferent and efferent connections of one type have been imaged, then the prospective diagnosis of TAPVR Can be readily made. We belive that our and the reported failure to detect split forms of anaomalous efferent connections stems from failure to follow a systematic search.

In summary we conclude that two dimensional sector scan echocardiography provides detailed anatomic imaging of CPVC and its afferent and Efferent Connections in various types of TAPVC.

The sensitivity of prospective echo diagnosis largely depends upon the skill of the echocardiographer and his detailed knowledge of various types of anomalous pulmonary venous connections.

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