

Approach to a Dilated Right Ventricle (For the beginners)

S.K. Parashar *

The right ventricle (RV) is no longer considered as a neglected chamber, because numerous studies have shown its prognostic significance in various cardiovascular diseases like congenital lesions, pulmonary hypertension, myocardial infarction, left ventricular (LV) dysfunction etc.. The RV is anatomically placed beneath the sternum and anteriorly positioned to the left ventricle (LV). The muscle mass of RV is approximately one-sixth that of LV, as it pumps blood against approximately one-sixth the resistance of LV. As compared to LV, the RV is thin walled (approx. 3-4 mm). The normal RV is accustomed to a low pulmonary resistance and hence a low afterload. As such normal RV pressure is low and has a high compliance. The RV is therefore sensitive to changes in afterload. As such RV enlargement occurs in response to chronic pressure and / or volume overload and also any cause leading to RV failure like RV infarction or dysplasia. Echocardiographic assessment of RV is limited by (a) complex geometry of the chamber (b) pronounced trabeculations that compromise accurate endocardial delineation (a) anterior position that often limits echo image quality. Owing to incomplete visualization of the RV in a single 2-D echo view, all possible scan planes need to be projected for a comprehensive evaluation of RV. This paper is mainly focused for level 1 / 2 echocardiographers.

What constitutes an RV enlargement: This can be (a) quantitative (b) qualitative. Various workers have proposed quantitative parameters to assess RV enlargement. However the problem is that there is a lack of fixed reference points to ensure optimization of RV. Depending upon the cut planes there can be significant variation in measured dimensions and is mostly underestimated. As such it has been proposed to obtain a focused RV view to concentrate on the lateral wall of RV. In this view (**Fig. 1**), the 4-chamber view is readjusted to focus on the RV rather than LV. It should be ensured that RV is not foreshortened. If one is able to get the desired view then two important measurements of RV are (a) the maximal short axis dimension in basal 1/3 of the ventricular cavity (b) longitudinal dimension from tricuspid annulus to RV apex. Diameter more than 42 mm at base and > 86 mm longitudinal dimension indicate RV enlargement. However the RV dimensions can be distorted and falsely enlarged in patients with chest and thoracic spine deformities.

Qualitative: Based on the above limitations and from a day to day practical point of view, a qualitative assessment is usually performed. Qualitatively the RV is smaller, and no more than 2/3 of LV in 4 chamber view. If RV appears larger than LV in this view then it is significantly enlarged (**Fig. 2**). Furthermore as RV enlarges it may displace the LV and form the apex – ‘apex forming ventricle’. This may indicate moderate RV enlargement but this finding has not been validated quantitatively. Visually the RV / LV end diastolic diameter ratio is ≥ 1.0 in cases of RV enlargement.

* Consultant cardiologist & Director, Non Invasive Cardiac Laboratory. Metro Hospitals & Heart Institute, Delhi. E-mail: drparashar@yahoo.com
Mobile : 09810146231

Approach to RV dilatation: (a) history & clinical examination: Suffice to say that they are extremely important in evaluation and may reduce the differential diagnosis and will aid in reaching a final diagnosis (b) ECG and X-ray chest form an important supplementary aids in evaluation (c) Echocardiography, both transthoracic and if need be transesophageal (TEE). They form a vital link in evaluation of RV. All scan planes for RV should be meticulously studied as echo strongly contributes in reaching a final diagnosis.

Common causes of RV dilatation : The various causes can be broadly classified in three groups (A) Conditions of pressure overload (B) Conditions of volume overload (C) Myocardial disorders. There are a large number of conditions in these groups, but only few common ones are discussed as seen in daily clinical practice.

Table 1: Conditions of Pressure Overload

Pulmonary Arterial Hypertension	
Group 1.	Idiopathic, congenital lesions
Group 2.	Associated with left heart disease
Group 3.	Associated with lung disease / hypoxia
Group 4.	Chronic thrombotic / embolic disease

RVOT obstruction	
Subvalvular, valvular or supra-valvular level	

Approach: The hemodynamic diagnosis of pulmonary arterial hypertension (PAH) comprises of a mean PA pressure of > 25 mmHg and a average peak systolic PA pressure of > 35-38 mmHg. While assessing RV enlargement, a PSAX view can give a qualitative idea of volume or pressure overloading of RV and hence a clue to increased RV / PA pressure. In presence of a normal RV / PA pressure, the interventricular septum (IVS) assumes a convex shape towards RV because of an increased LV pressure in relation to RV. With increasing PA / RV pressure, this pressure gradient is reduced. As such the IVS becomes flat and with increasing pressure assumes a 'D' shape with IVS convexity towards LV. (**Fig 3**). This configuration of the IVS is dependent on the relative pressure gradient between LV and RV at each stage of cardiac cycle. This distortion is present both in pressure overload (distortion both in diastole and systole) and volume overload (distortion most marked in end diastole with relatively more normal geometry in end systole). As such 2-D echo gives a semi-quantitative idea of PAH. Moreover it is a guide for the echocardiographer to carefully assess the PA pressure. As is well known, a more quantitative idea is obtained by peak tricuspid regurgitation velocity using simplified Bernoulli equation i.e. $RVSP = 4 \times TRV^2 + RA \text{ pressure}$. This is based on the principle that any regurgitant jet velocity is based on the pressure difference between the transmitting and the receiving chamber. As such higher the RV pressure, the higher will be the TR jet velocity. In this method there should be no RVOT obstructive lesion which invalidates this formula.

There are several methods for evaluation of mean PA pressure. However two most widely used methods are (a) $0.61 \times \text{Peak SAP} + 1.95 \text{ mmHg}$ or simply $0.60 \times \text{Peak SAP}$ obtained by conventional TR jet velocity method .(b) mean TR jet velocity, obtained by planimetry of jet + RA pressure (**Fig4**). Both methods have been validated with invasive studies.

Once a PAH is assessed, then based on table1, the etiology can be determined. In all the groups, the history, clinical examination, ECG, peripheral venous contrast echo can give an important clue to the possible diagnosis. In all these conditions RV and RA will be enlarged. A careful examination of all scan planes would reveal any left sided valvular lesions including that of tricuspid valve leading to PAH. (**Fig 5**). Group 3 mainly includes cor pulmonale, the commonest cause of which is COPD, and will be evident by history, absence of any other cause for RV / RA enlargement and PAH. Group 4 can have a very variable history & clinical presentation and hence a high index of suspicion will aid in diagnosis. One of the commonest association of pulmonary thromboembolism is deep vein thrombosis. Pulmonary embolism occurs in about 50% cases of DVT. Besides clinical and biochemical parameters, echo could be helpful in diagnosis. Though echo has a low to moderate sensitivity in diagnosis, but is still an integral part of diagnosis, prognosis and guide to management. The diagnostic role of echo is (indirect) and (direct). The indirect evidences include RV enlargement, McConnells sign – regional RV dysfunction sparing the apex., TR, PAH, dilated IVC with decreased inspiratory collapse due to increased RA pressure. The direct evidences are floating right heart thrombus, including IVC (**Fig 6,7**). Thrombus in pulmonary arterial circulation is seen in < 20% cases, but TEE may be more diagnostic. RVOT obstruction can be isolated or more commonly part of a complex disease process. A complete echo including color flow mapping (CFM) & Doppler will provide diagnosis. CFM helps in localising the lesion by demonstrating the site of onset of turbulence i.e. infundibular, pulmonary valvular or supra valvular level. An attempt should be made to see for other associated lesions especially congenital. As such once a PAH is evident, it is reasonably easy to reach an etiological diagnosis by deductive echo and process of exclusion.

Table 2: Conditions of RV Volume Overload

Pre – Tricuspid Shunts : Atrial septal defect

Tricuspid valve lesions

Pulmonary regurgitation

Approach : Irrespective of age, any unexplained RV, RA enlargement needs exclusion of atrial septal defect which can be of any anatomical type like secundum, primum, sinus venosus etc. In case of any doubt, a transesophageal echo will be diagnostic. The first two anatomical types can be easily visualised from various scan planes, especially subcostal views as apical 4 chamber view may show a false positive secundum defect, unless CFM shows a classical L-R flow. A primum defect , has many synonyms like incomplete or partial endocardial cushion defect / partial

atrioventricular septal defect / ostium primum defect. It is usually a large defect in the inferior part of the septum and is often associated with cleft mitral valve. Unlike secundum ASD, it is often best imaged from apical four chamber view (**Fig 8**). Normally the septal leaflet of tricuspid valve originates more apically compared to anterior mitral leaflet, but in ostium primum ASD, both leaflets arise at the same level as a consequence of endocardial cushion defect. The cleft mitral valve, mainly of anterior leaflet, is visualized from short axis view. CFM evaluates the shunt flow, estimates presence and degree of MR, TR and excludes VSD. TR and PR Doppler gradients help in assessment of PA pressure. Sinus venosus ASD, whether of superior vena caval or inferior vena caval type, usually present a diagnostic problem, especially in adults and those having suboptimal subcostal window as this is the most helpful imaging window. It is important to image the SVC as it enters the RA. A CFM aids in diagnosis as it straddles the ASD. In any case, usually a TEE is performed to confirm the diagnosis (**Fig 9**).

The primary diseases of tricuspid valve leading to regurgitation are relatively uncommon because most of the TR are functional. However some relatively common lesions are endocarditis, pacing electrode damage of the valve, myxomatous degeneration, carcinoid tumors, Ebsteins anomaly which mainly leads to RA enlargement with marked displacement of septal and posterior tricuspid leaflet apically in relation to insertion of mitral valve and is more than 8 mm / m² or more than 15-20 mm in absolute values (**Fig 10**). The RA is hugely dilated. Because of displaced tricuspid leaflets, the RV is divided into two parts: the atrialized ventricle (**Fig 11**) and the functional ventricle. The atrialized part of RV contributes nothing towards RV stroke output. Though RA and atrialized portion of RV behave as venous receiving chamber they do not contract simultaneously as a unit.

Though PR can be congenital (absence of pulmonary valve, dilatation of pulmonary valve ring due to PAH or idiopathic dilatation of pulmonary artery), but in adult practice, it is mostly acquired due to post interventional and post surgical valvotomy like total correction of Tetralogy of Fallots or other congenital lesions requiring RVOT repair / reconstruction. As such in all post intervention or post operative situation, CFM must be utilized to interrogate the pulmonary valve. As a practical guide, on CFM when the PR jet (red as it is directed towards transducer) changes to a blue coloration distally (jet moving away from transducer) it indicates a moderately severe PR.

Table 3: Myocardial Disorders

RV cardiomyopathy
RV infarction
Uhl's anomaly
Terminal stage of cardiac remodelling
Miscellaneous

Approach : RV cardiomyopathy, also referred in literature as arrhythmogenic right ventricular dysplasia, is a non ischemic cardiomyopathy usually presenting with malignant ventricular arrhythmias, cardiac failure, syncope etc. In about 17% of cases it is a common cause of sudden cardiac death in young patients and athletes. One of the basic lesion in this condition is progressive diffuse or segmental replacement of RV myocardium by fibro fatty tissue. It is mainly characterized by RV dilatation and dysfunction, hyperreflective moderator, localized thinning and segmental systolic outpouching, localized RV aneurysm, excessive trabeculation (**fig 12-14**). A high index of suspicion is required for diagnosis in a patient presenting with ventricular tachycardia and or / syncope with RV enlargement on echo.

RV infarction (RVI) accompanies inferoposterior myocardial infarction in about 30-50% of cases. It is extremely important to recognize RVI as it is associated with significant immediate mortality and morbidity and has a well delineated set of priorities for its management. A clinical clue to RVI is present when there is hypotension, elevated jugular venous pressure and occasionally shock- all in the presence of clear lung fields. Other clue is provided by ST elevation of $> 0.1\text{mV}$ in V4R. Based on this presentation, echocardiography can be a helpful guide to diagnosis. Abnormal important findings include RV dilatation, RV regional wall abnormalities, reversed interventricular septal curvature towards LV due to increased RV end diastolic pressure. However the RWMA of RV may be correlated with whole clinical picture as it could be a relative non specific finding. There is significant RA enlargement. Doppler findings may further support diagnosis by detecting TR or a shunt flow across patent foramen ovale. Many of the echo findings of wall motion defect may be transient and not be visualized after 48 – 72 hours.

Uhl's anomaly is a such a rare condition that its exact incidence and prevalence is not known. The RV is hugely dilated with very thin (parchment like) walls. RA is dilated. However tricuspid leaflets show normal attachment and there is no significant TR. Due to its rarity, one is liable to miss diagnosis in adult clinical practice.

Finally RV, as a cardiac chamber, can be involved in terminal stage of any cardiac condition remodelling like dilated cardiomyopathy, myocardial infarction, heart failure, etc. As such besides RV,RA dilatation and RV dysfunction, there will be echo features of the primary cardiac condition.

Summary

- Right ventricular enlargement is not an uncommon situation.
 - A spectrum of diseases lead to increased RV pressure or volume overload leading to RV enlargement. The myocardial causes are relatively uncommon.
 - Only few common causes of RV enlargement are highlighted as seen in daily clinical practice
 - There is no need to get biased by the referral diagnosis
 - Form your own clinical impression by a good clinical history and clinical examination
 - The operator should never be content with one diagnosis as there could be multiple lesions
 - Examine every cardiac structure and use multiple imaging planes with proper sequence to explain etiology of RV enlargement. Follow the sequence of 2-D study followed by CFM and Doppler examination.
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Descriptive Legends

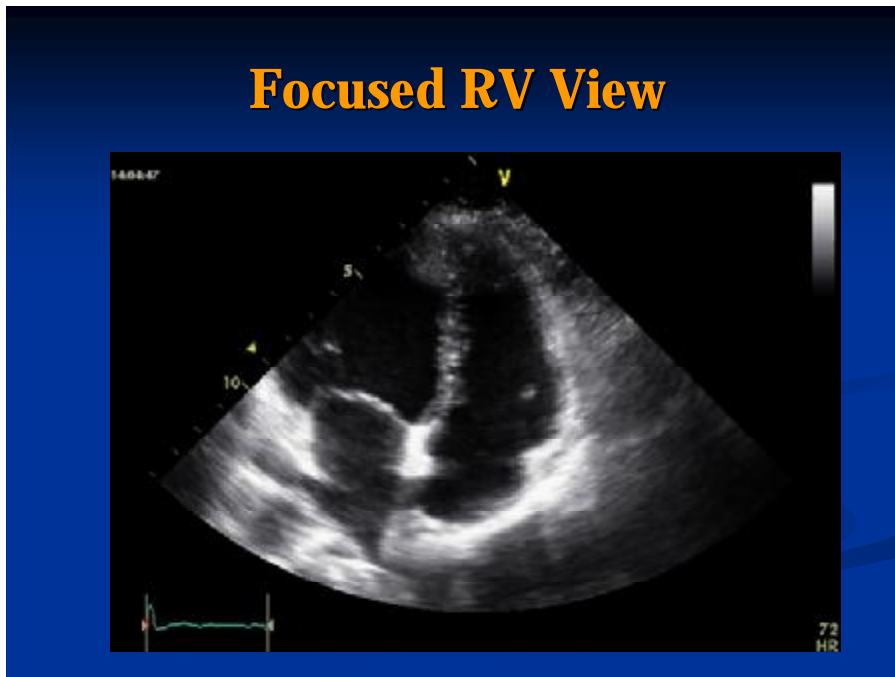


Fig 1: The RV is on the right side of the image which has been modified to bring the whole RV in the view and the lateral wall is well seen.

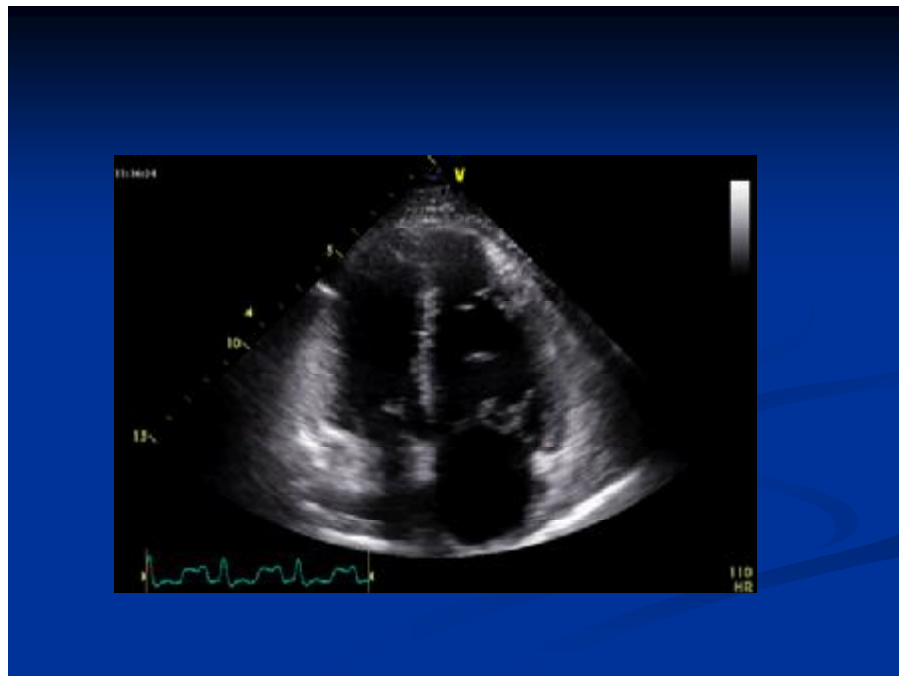


Fig 2: An apical 4 chamber view with right ventricle and right atrium on the right side and left sided chambers on left side. Qualitatively the RV is significantly enlarged as compared to left ventricle.

Increased RV Pressure

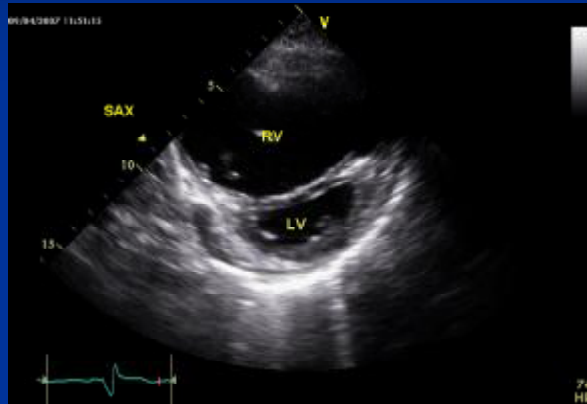


Fig 3: A parasternal short axis view showing a 'D' shaped ventricle with IVS showing convexity towards LV. Note marked enlargement of RV: SAX- short axis view, RV – right ventricle, LV-left ventricle

Mean PAP=33+10 mmHg.

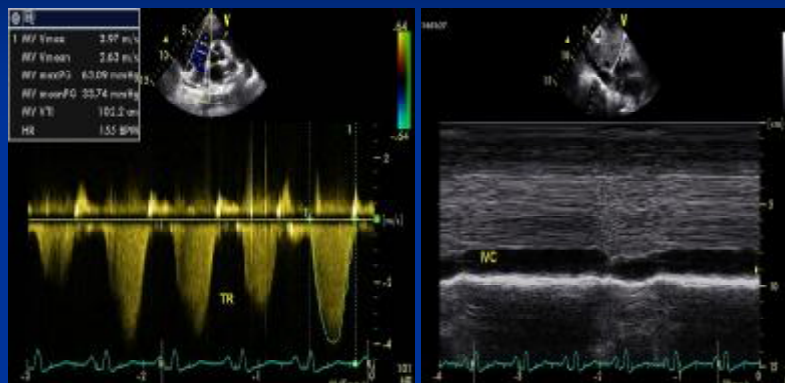


Fig 4: One of the proposed methods for calculation of mean PA pressure. The planimetered TR jet gives a mean gradient of 33 mmHg to which RA pressure is added based on RA size and collapsibility. The calculation square, though mentioned as MV, indicates TV calculation. In daily practice an average of at least three readings should be taken.

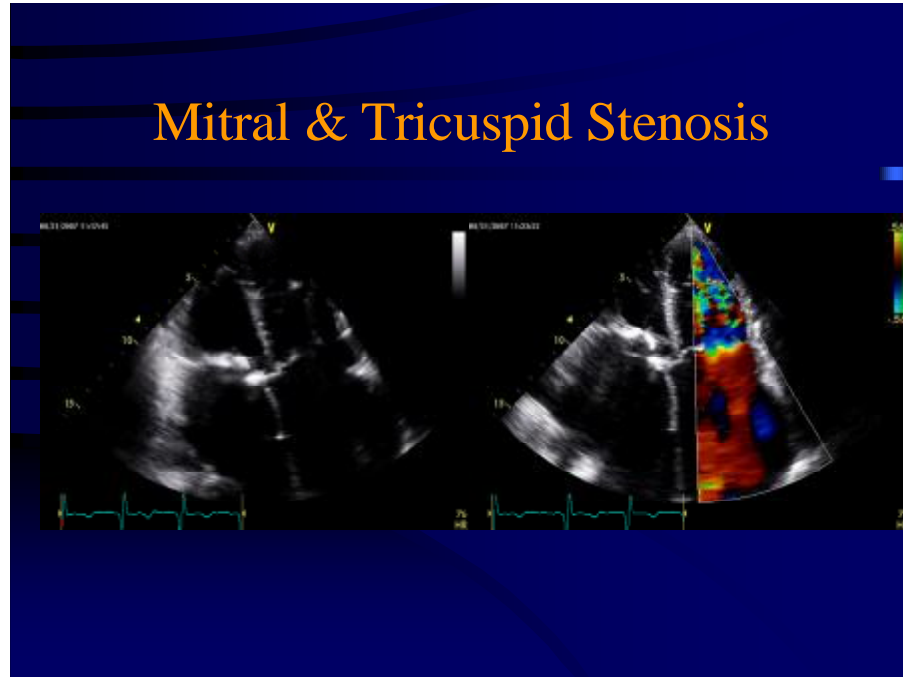


Fig 5: Showing stenosis of mitral valve (left side of screen) with tricuspid stenosis. Color flow mapping across tricuspid valve shows turbulence across the valve



Fig 6: A case of pulmonary embolism showing multiple thrombi in right atrium as seen from parasternal short axis view

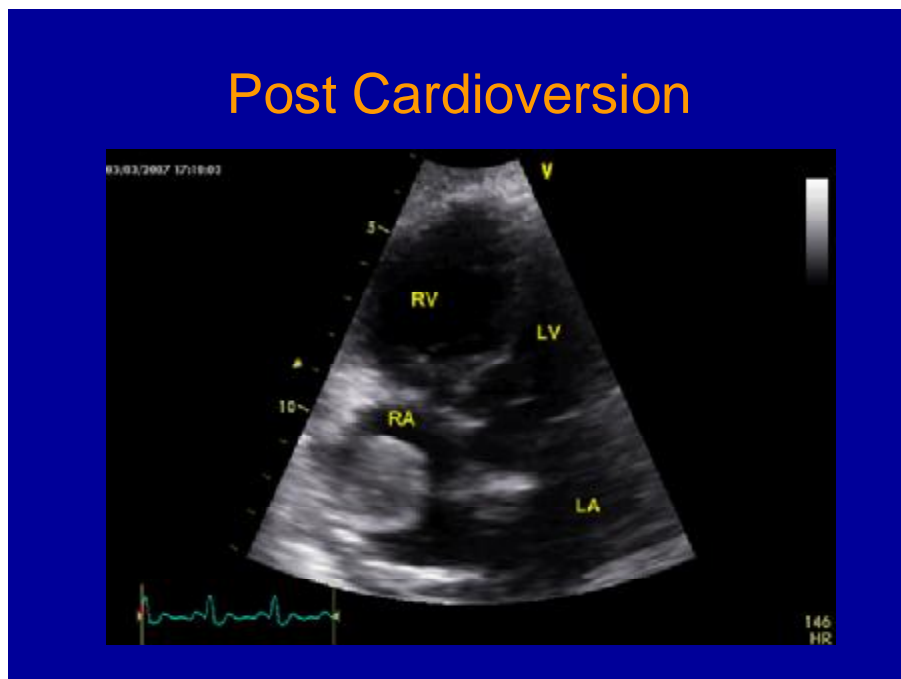


Fig 7: A case of constrictive pericarditis who was cardioverted for AF/ flutter. Shortly after cardioversion patient developed multiple biatrial thrombi possibly due to atrial fibrillation and atrial stunning. RA-right atrium. LA – left atrium, RV- right ventricle, LV left ventricle



Fig 8: An apical 4 chamber view showing ostium primum ASD. See text for details

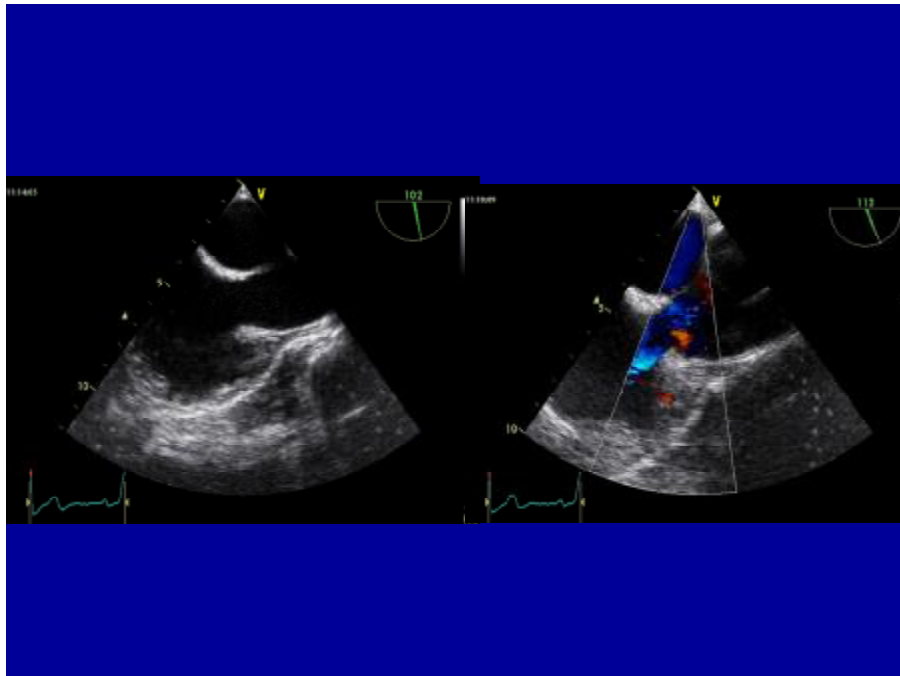


Fig 9: A transesophageal echo showing a sinus venosus defect in a bicaval view. CFM shows flow from LA (seen towards transducer) to RA

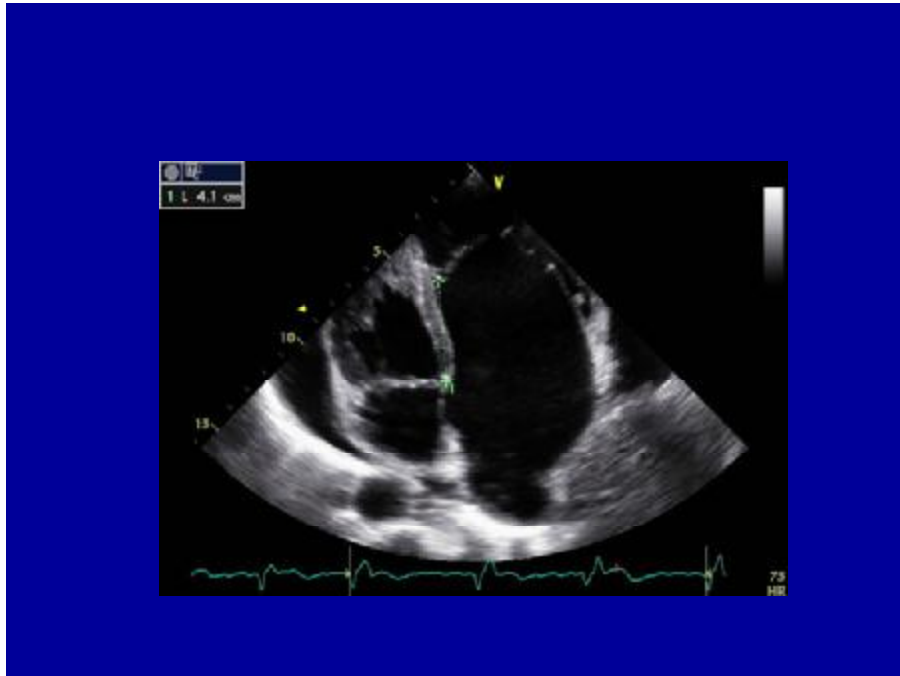


Fig 10: A case of Ebsteins anomaly showing marked apical displacement of septal leaflet of tricuspid valve (4.1 cms) as compared to mitral valve. The anterior tricuspid leaflet has large amplitude. The tricuspid annulus is enlarged (right side of image)

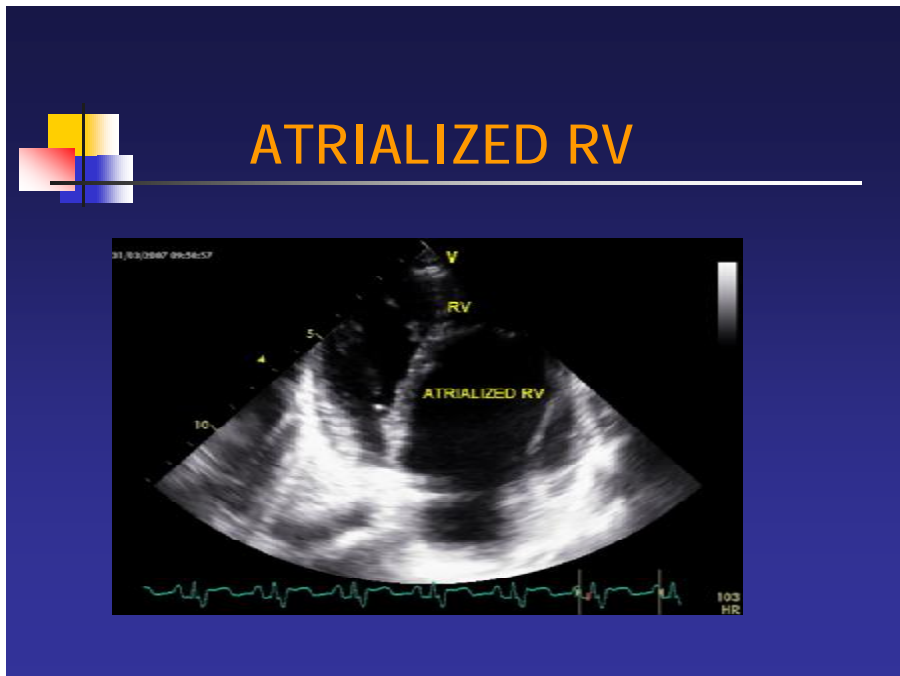


Fig 11: Another case of Ebsteins anomaly showing large atrialized right ventricle. The functional right ventricle (RV) is small and thin walled



Fig 12: A case of right sided cardiomyopathy. Apical 4 chamber view shows enlarged right ventricle , as seen on right side of the image, with hyperreflective moderator band extending from IVS to the lateral wall of RV.



Fig 13: Same patient as in Fig.12, showing localized systolic outpouching in the mid portion of lateral wall of right ventricle in a modified apical 4 chamber view



Fig 14: Same patient as in Figs 12,13. A subcostal view showing localized atrophy of RV in mid segment and an apical RV aneurysm.

Suggested Reading

1. Recommendations for chamber quantification. Eur J Echocardiogr 2006; 7: 79
2. The right ventricle: anatomy, physiology and clinical imaging. Heart 2008; 94: 1510
3. Echocardiography in the assessment of right heart function. Eur J Echocardiogr.2008;9:225
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7. Echocardiography in Congenital Heart diseases: A Practical Approach: 2010, Savitri Shrivastava et al. Siddharth Publications
8. Comprehensive Text Book of Echocardiography: 2013: Navin C Nanda, Jaypee Brothers, Medical Publishers (P) Ltd. Delhi