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Heart Failure in a Patient with Ebstein's Anomaly Precipitated by Arrhythmia: A Case Report

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ABSTRACT

Background Ebstein's anomaly (EA) is a rare cyanotic congenital defect of the tricuspid valve (TV) and the right ventricle (RV) in which the attachments of the septal and posterior valve leaflets are apically displaced. This case demonstrates the unknown cause of an adult onset Ebstein's anomaly in a female patient with Heart failure precipitated by arrhythmia along with polycythemia as the predominant manifestation. **Case Presentation** We report a 50-year-old female presented with severe dyspnea on exertion, productive cough, orthopnea, decreased appetite, fatigue and progressive tiredness along with increased palpitation. She developed a progressive dyspnea from grade II to grade IV. Her Echocardiogram (ECHO)examinations revealed Heart Failure (HF), arrhythmia, Ebstein's anomaly, severe tricuspid regurgitation, mild tricuspid stenosis, and normal Ejection Fraction (EF); her laboratory findings revealed polycythemia. **Conclusion:** Like many other heart conditions, there is no true cure for Heart Failure (HF) in cyanotic congenital heart disease. Mortality rate for heart failure in cyanotic congenital diseases is still higher than for the general population. In most of the cases in adults, though treatment can reduce or even eliminate symptoms, management of this heart failure with Ebstein's Anomaly condition is complex so individualized treatment is required.

BACKGROUND

Ebstein's Anomaly is a rare cyanotic congenital disorder that occurs in less than 1 per 2,00,000 live births. It involves primarily the apical displacement of the tricuspid valve and right ventricular myopathy [1]. Ebstein's anomaly is a spectrum of tricuspid valvular and right ventricular dysplasia. The exact cause is unknown.A small proportion of cases with moderate to severe abnormalities are present in adults, however most cases are diagnosed in infancy or childhood. Children, adolescents, and adults with Ebstein's anomaly are presented with exertional dyspnea, fatigue, cyanosis, and palpitations. Some patients might be asymptomatic. The most associated cardiac defects include atrial septal defect or patent foramen ovale (~80–90%) and right ventricular outflow tract obstruction that can occur secondary to structural abnormalities (pulmonary valve stenosis or pulmonary atresia), branch pulmonary artery stenosis or patent ductus arteriosus [2]. Ebstein's patients are also at risk of atrial arrhythmias. The contributing reason for this could be atrial enlargement and development of accessory pathways. This can sometimes precipitate heart failure symptoms in patients with cyanotic heart diseases.

Anatomical Description:

The tricuspid valve leaflets show a variable degree of failed delamination with fibrous and muscular attachments to the right ventricular myocardium. The displacement of the hinge point of the septal and posterior leaflet in the right ventricle towards the apex is the hallmark finding of Ebstein's Anomaly.

Etiology:

Studies suggest that genetic, reproductive, and environmental risk factors are commonly contributing to Ebstein's Anomaly. It is more common in first degree relatives (twins), patients with a history of congenital heart defects, and maternal exposure to benzodiazepines [3]. Mothers exposed to lithium therapy may also give birth to offspring with Ebstein's Anomaly.

Cardiac malformations Associated with Ebstein's Anomaly

Inter-atrial communication is the major defect noted in 80 to 94% of patients with Ebstein's Anomaly. Bicuspid or aortic valve stenosis, sub aortic stenosis, mitral valve prolapse, Ventricular septal defect (VSD), pulmonary stenosis is associated with Ebstein's Anomaly.

Tricuspid valve stenosis is found to be present in most of Ebstein's Anomaly cases. Left ventricular malformations, lesions, patent foramen ovale and atrial septal defects are commonly associated.

Here, we report a case of Ebstein's Anomaly of Tricuspid valve with tricuspid stenosis in a patient with underlying medical conditions such as severe tricuspid regurgitation, Bronchial Asthma and polycythemia that was managed therapeutically.

CASE PRESENTATION

A 50-year-old female patient from southern district in Tamil Nadu was admitted to the Cardiology department of a super specialty hospital. She was presented with the complaints of dyspnea on exertion for the past 6 months (Grade IV), associated with a productive cough with white sputum expulsion, orthopnea, and decreased appetite. The patient also experienced fatigue and tiredness of progressive type along with increased palpitation; intermittently for the past 6 months that lasted for 10 minutes. She experienced 2 episodes of voluminous vomiting and giddiness with no loss of consciousness. She also complained of decreased sleep for the past 6 months due to dyspnea as her Sleeping time was between 10pm and 1pm. She had a past medical history of Diabetic Mellitus type II, hemorrhoid procedure done in 2005, and postmenopausal status. But she was not on any medications.

On examination the ECG revealed atrial flutter with 2:1 AV Conduction with Right Bundle branch block [FIG]. Baseline blood investigations revealed hyperuricemia (Serum Uric acid-10.6 mg/dl), uncontrolled diabetes mellitus (HbA_1C-9.7%), subclinical hypothyroidism (TSH-5.28 microIU/ml), hyperkalemia (5.90 mEq/L), and polycythemia (Hb- 18.7 mg/dl). 2D ECHO showed Ebstein's Anomaly of Tricuspid Valve, STL Displacement measuring 40mm, Severe Low Pressure Tricuspid Regurgitation (RVSP-39 mmHg), Non Coaptating Tricuspid Leaflet, Right Ventricle Dysfunction Present (TAPSE - 9mm, S'-0.67m/s), Normal Left Ventricle Ejection Fraction, LV 33/22, and Ejection Fraction: 62% [FIG]. It has been found that her Renal Function tests (RFT) were found to be normal.

With the clinical features and echocardiographic characteristics fulfilling the diagnosis criteria, a clinical diagnosis of Ebstein's Anomaly of Tricuspid Valve was established. In this patient, following are the findings that were found to be associated with Ebstein's Anomaly such as Cyanotic congenital heart disease, Severe Tricuspid Regurgitation, Right Ventricle Dysfunction, Cyanotic Congenital Heart Disease - Right to Left shunt OS Atrial septal defect,

Atrial flutter with variable AV Conduction was done. She was treated with diuretics Injection Torsemide (Dytor) 10 mg (which was later changed to oral tablet) and tablet Digoxin (Lanoxin) 0.25 mg which are the standard therapeutic management in Ebstein's anomaly for volume overload related to right Heart Failure. Further she was on tablet spironolactone (Aldactone) 25 mg and tablet Amiodarone (Cardarone)200 mg. Prophylactically, she was on Inj. Low Molecular Weight Heparin 0.6 IU for thrombo-embolism.

On Pulmonology opinion, pulmonologist investigations revealed bronchial asthma, for which she was initiated on tablet Acebrophylline (AB Phylline). Other underlying medical conditions like Polycythemia were managed by venesection and hyperkalemia by K bind. Patient is better, stable, at present drug optimized and is being discharged at stable cardiac status. Further she is being followed up in the Cardiology outpatient department.

DISCUSSION

Ebstein's anomaly comprises approximately 1% of all congenital heart diseases [4]. If the deformity in the tricuspid valve is severe, the patient may experience severe symptomatic representation. In contrast if there is a mild degree of tricuspid malformation the patient may be asymptomatic.

A large spectrum of lesions and complications has been described in our patient. Our patient had complex abnormalities as well as tricuspid stenosis which are otherwise rare in Ebstein's anomaly. Also interestingly in this patient, there is coexistence of other illnesses such as Bronchial asthma, polycythemia, hyperthyroidism, hyperkalemia, and uncontrolled diabetes were noted. The potassium is eventually excreted in the urine after being exchanged for sodium and water by the ATP-ase pump. As a result, hyperkalemia may result from aldosterone antagonistic effects so a known side effect of spironolactone is hyperkalemia. But in this case, hyperkalemia is not found to be associated with spironolactone. Due to Amiodarone's strong lipid affinity, high tissue concentration, and a host of negative consequences, including thyroid dysfunction, amiodarone can impede the entry of T4 and T3 into peripheral tissues. Amiodarone and its main metabolite both have the potential to directly damage thyroid follicular cells, which can result in destructive thyroiditis. Due to amiodarone's relatively high iodine content, normal thyroid auto-regulation is lost [5], but in this case, subclinical hypothyroidism was present before she was administered with Amiodarone.

In adults, the most typical presenting symptom in Ebstein's patients is arrhythmia. These arrhythmias are generally not well tolerated. Despite the complexity of the morphological classification of congenital heart abnormalities, three major categories account for a significant portion of adult Cyanotic congenital heart disease patients who have arrhythmia due to their prevalence and higher incidence. Arrhythmia in adults with CHD can have a variety of clinical effects, from clinically concealed arrhythmia to abrupt death. A vicious cycle of clinical decompensation is frequently the outcome of persistent or recurrent arrhythmia, which can lead to progressive hemodynamic deterioration and vice versa. In contrast, the population of CHD are small, anatomically diverse, and have relatively low rates of sudden death, with annual mortality in even "high risk" groups hovering around 2%. Therefore, it is challenging to pinpoint congenital heart disease's risk factors and gauge how therapies affect survival.

Due to the size, irregular shape, and tricuspid regurgitation of the right ventricle, right heart failure affects the forward flow of blood through the heart. A repaired or replaced valve that degenerates over time can cause considerable tricuspid regurgitation, and the right ventricular chamber is typically small with varied degrees of right ventricular outflow tract blockage. The right ventricle may not be able to withstand volume loads or other hemodynamic stressors as well as a normal right ventricle, even when lower right ventricular size is considered. Additionally, the atrialized right ventricle and expanded right atrium serve as a reservoir which decreases right ventricular cardiac output thus causing Heart Failure due to Ebstein's Anomaly [6].

Management of Heart Failure patients with Ebstein's anomaly is usually determined by patient's individual characteristics like age, clinical presentation, sex etc. The major composition of management includes monitoring, therapeutic management, and surgical intervention [7]. All patients with Ebstein's anomaly should be carefully monitored irrespective of their severity. Only the time interval for monitoring varies according to the severity and complexity of the case. Usually the clinical presentations, signs for heart failure, arrhythmia, and cyanosis are monitored. Further medical management is required only in moderate to severe cases, which is further varied with respect to presence or absence of symptoms. Eventually most of the patients may require surgery. Patients with a New York Heart Association class I or II can be managed therapeutically, but further patients with a NYHA class III or IV may require surgical intervention [8]. Many physicians believe that

severe tricuspid regurgitation and right ventricular dysfunction are indications for surgery even in asymptomatic patients where a tricuspid valve repair is prioritized.

It should be highlighted that the Pharmacotherapeutic management-based approach in patients with Ebstein's anomaly may be questionable because operative indication for symptomatic patients has been clearly defined [9]. In our case the patient was symptomatic with complaints of dyspnea, orthopnea, severe tricuspid regurgitation, mild tricuspid stenosis, and increased palpitation, where surgery is usually indicated, but the patient denied and refused to undergo a surgery. And hence it was decided to compromise with therapeutic management instead of a heart transplant. Furthermore, since the patient was acyanotic, with a good ejection fraction of 60%, normal left ventricular function and acceptable right ventricular function she was symptomatically treated. She was therapeutically managed on diuretic therapy, anticoagulants, and inotropes.

CONCLUSION

Ebstein's anomaly is an uncommon cyanotic congenital anomaly complex with a broad spectrum of anatomic and clinical features. This case report demonstrates Heart Failure (HF) with Ebstein's Anomaly of Tricuspid valve in a female patient and polycythemia as the predominant manifestation. Initially, patients with Ebstein's anomaly are usually seen in mid to later adulthood life. Like many other heart conditions, there is no true cure for HF patients with Ebstein's anomaly. While mild to moderate heart failure can be treated medically, surgical intervention is necessary to treat increasing ventricular dysfunction and tricuspid valve regurgitation. The patient with the Ebstein's anomaly continues to face difficulties with poor Right Ventricle (RV) function and the requirement for reoperation on the tricuspid valve because of recurrent TR. Patients with Ebstein's continue to have mortality at a higher rate than the general population. More research is required to determine the best time to intervene, and surgical and medical strategies to address poor RV function and associated right-sided heart failure continue to evolve. With the precise knowledge, the clinical spectrum is variable and depends on the severity of defects associated with anatomical changes so management option is a kind of necessity. To avoid late RV failure and better short- and long-term results, surgical surgery is recommended prior to the development of significant right heart hypertrophy or malfunction. Thus, it is important that Heart Failure patients with Ebstein's anomaly should be evaluated regularly by a cardiologist. Considering the severity of the cases

and potential for fatal outcome, it is hoped that the survival rate of heart failure with Ebstein's anomaly of Tricuspid valve of all ages will continue to surpass.

LIST OF ABBREVIATIONS

EA: Ebstein's anomaly; RV: Right Ventricle; TV: Tricuspid Valve; TR: Tricuspid Regurgitation; VSD: Ventricular septal defect; TSH: Thyroid Stimulating Hormone; CHD: Cyanotic Congenital Heart Disease; HF: Heart Failure; Hb: Hemoglobin; HbA_1C: Glycosylated Hemoglobin; ECG: Electrocardiogram; ECHO: Echocardiogram; RFT: Renal Function Test; NYHA: New York Heart Association; OS ASD: Atrial septal defect; LMWH: Low Molecular Weight Heparin; RV: Right Ventricle; STL: Septal Tricuspid Leaflets.

DECLARATIONS

Ethics approval and consent to participate

The approval for this study and consent to participate was obtained from Institutional Human Ethics Committee (IHEC, PSG Institute of Medical Sciences & Research) and REF. NO: PSG/IHEC/2022/Appr/Exp/207.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Availability of data and materials

Not Applicable.

Competing interests

The authors declare that they have no competing interests to publish this case report.

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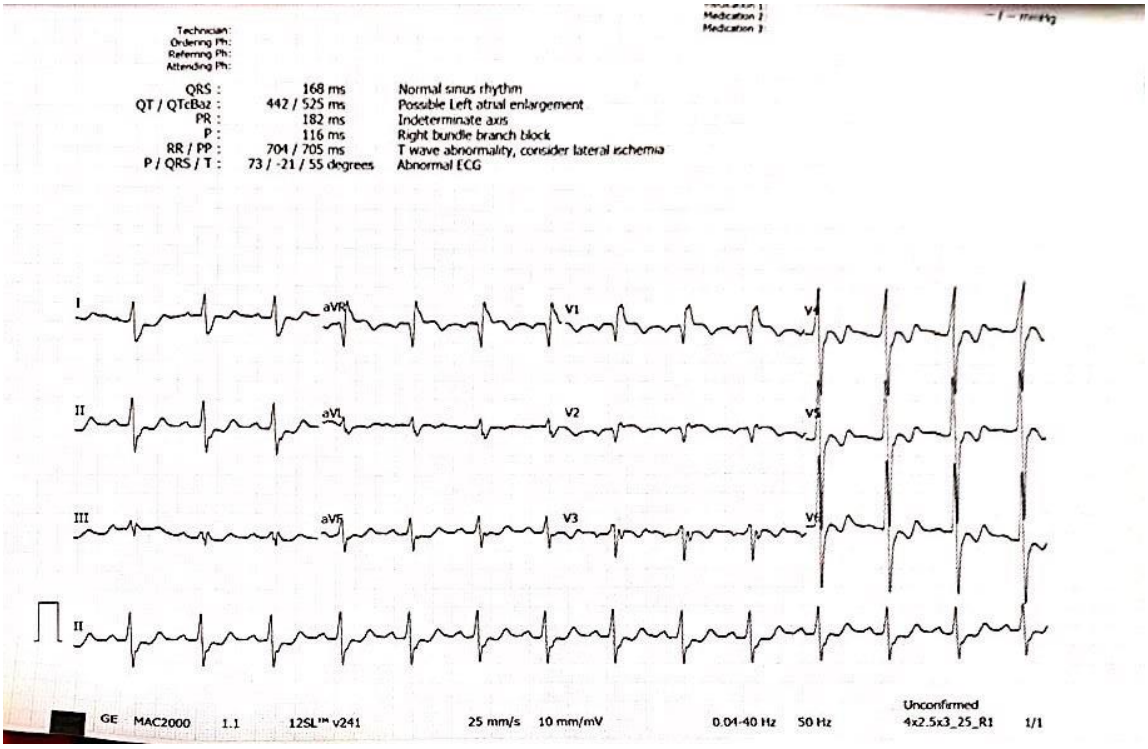

Figure 1: Normal sinus rhythm. 1:1 AV conduction, Right Axis deviation, Himalayan P waves, Bizarre QRS complex; Ebstein's Anamoly.

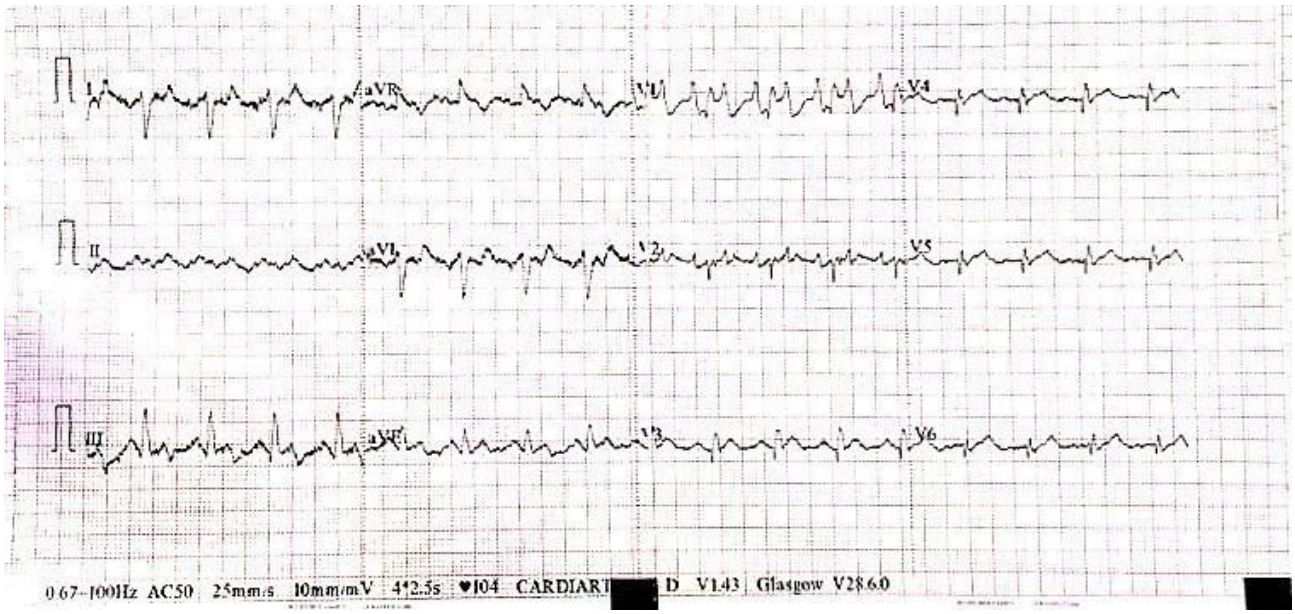

Figure 2: RBBB with secondary ST-T changes.

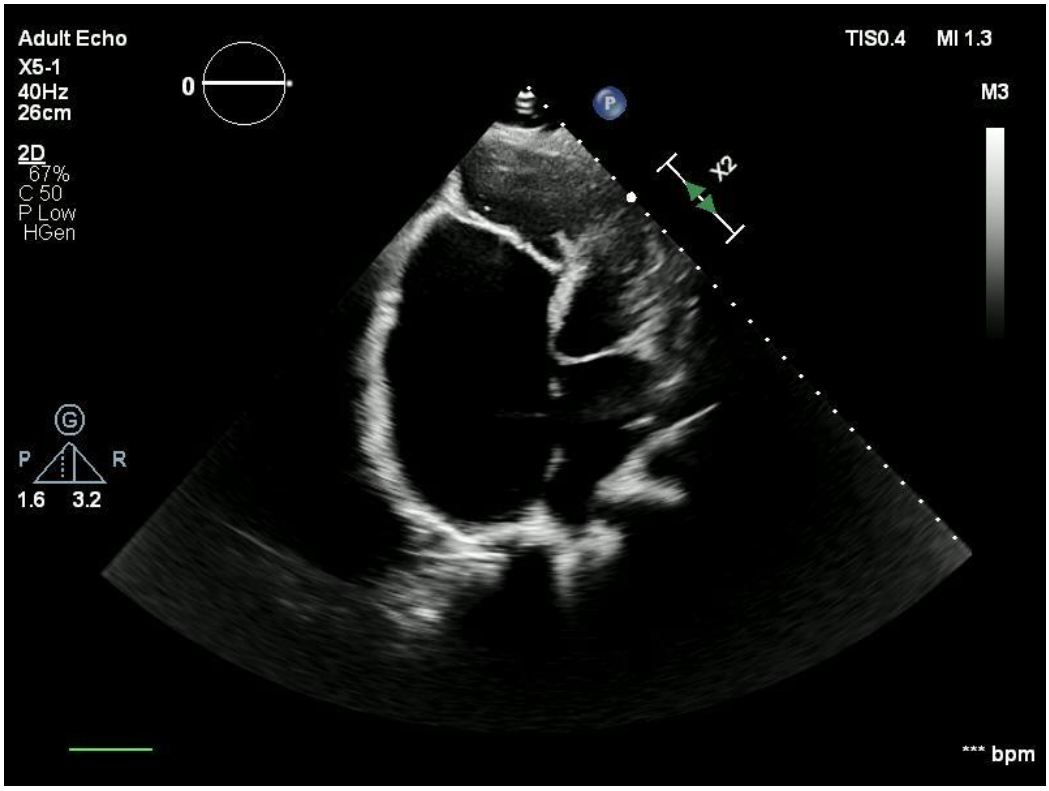

Figure 3: Four chamber view with trans dilated RA/RV with displacement of septal leaflet of TV with hypoplastic RV suggestive of severe Ebstein's Anomaly of TV.

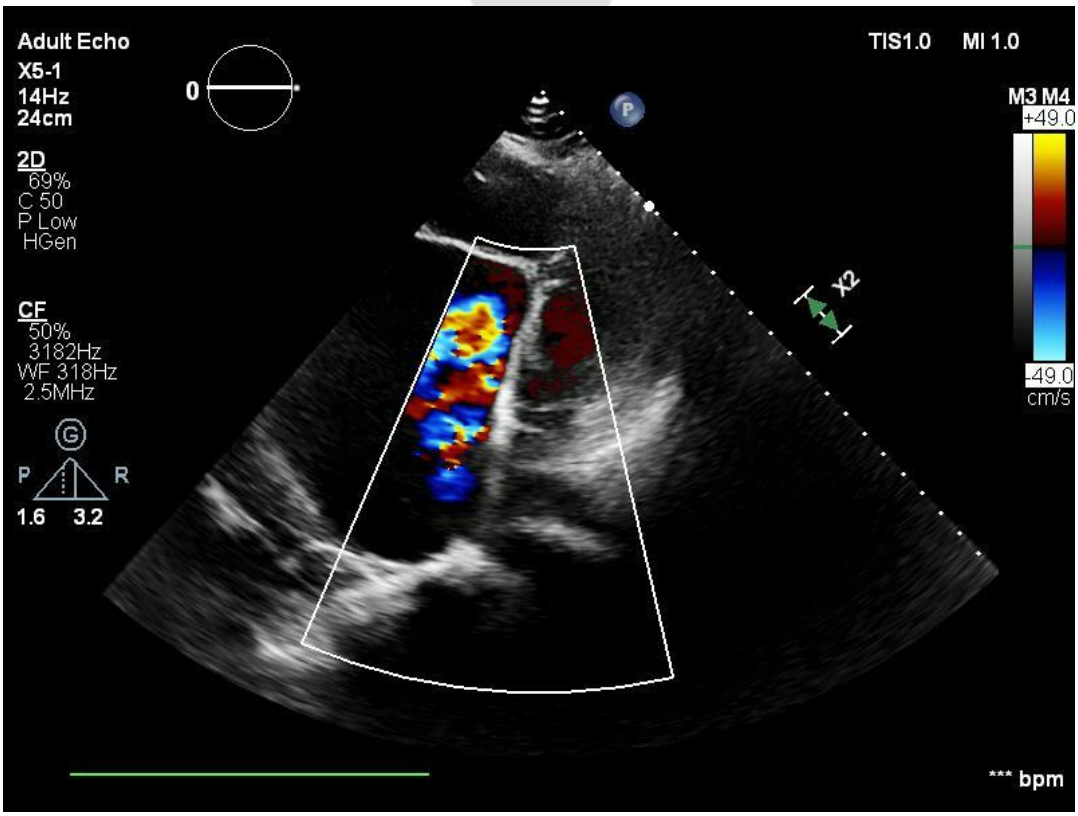

Figure 4: Colour transthoracic ECHO suggestive of severe TVR

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