World Journal of Biology Pharmacy and Health Sciences
eISSN: 2582-5542
Cross Ref DOI: 10.30574/wjbphs
Journal homepage: https://wjbphs.com/

(CASE REPORT)

Ebstein's anomaly in an infant: Case report and literature review

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World Journal of Biology Pharmacy and Health Sciences, 2025, 21(03), 259-273

Publication history: Received on 29 January 2025; revised on 07 March 2025; accepted on 10 March 2025

Article DOI: https://doi.org/10.30574/wjbphs.2025.21.3.0268

Abstract

Ebstein's anomaly is a rare congenital heart defect characterized by apical displacement of the tricuspid valve and reduced volume of the true functional right ventricle. In its most severe form, the disease presents as right heart failure and cyanosis in the infant. Although many of these infants can be managed conservatively, surgical intervention may become necessary in the sickest of them. Surgical intervention in this subgroup can be challenging and requires meticulous appreciation of the anatomy and physiology on a case-to-case basis. Multicenter studies are required to define a uniform algorithm and approach towards these patients.

Keywords: Ebstein's Anomaly; ASD; RV Dysfunction in Ebstein's Anomaly; Himalayan P Waves On ECG; Severe Tricuspid Regurgitation; DORV; Complete atrio-ventricular canal defect; CAVCD

1. Introduction

Ebstein's anomaly (EA) is a rare congenital cardiac defect with a prevalence of 2.4 per 10,000 live births and accounts for less than 1% of all newly diagnosed congenital disorders [1]. EA is characterized by apical displacement of the tricuspid valve and reduced volume of the true functional right ventricle [2] (Figure 1, 2).

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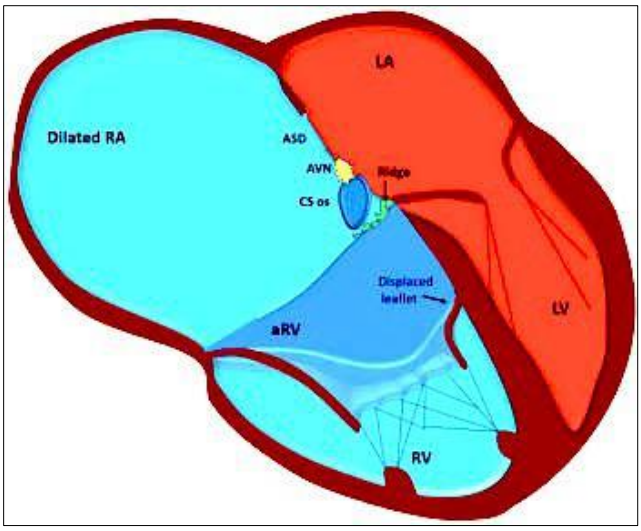


Figure 1 Diagrammatic illustration. Illustration of a heart with Ebstein's anomaly. The right atrium (RA) is dilated and so is the tricuspid annulus. The septal tricuspid leaflet is apically displaced, and there is a fibromuscular ridge instead (arrow). The coronary sinus (CS) ostium is dilated, and the AV node (AVN) is irregular, and its body can reach the upper border of the CS ostium. ASD, atrial septal defect; aRV, atrialised RV; LA, left atrium; LV, left ventricle; RV, right ventricle

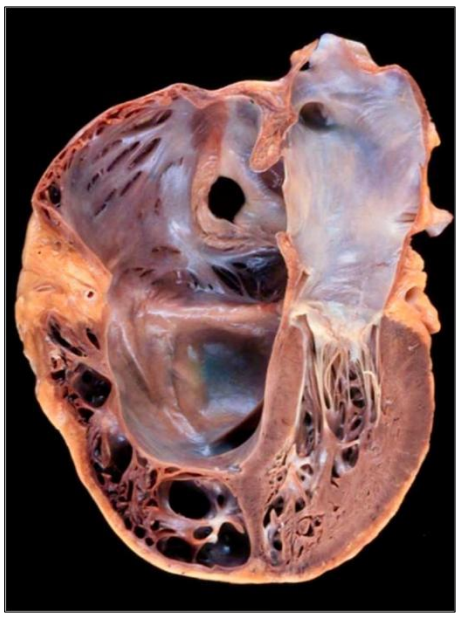


Figure 2 Pathologic specimen cut in the 4-chamber plane from a patient with Ebstein anomaly. The tricuspid valve is displaced markedly inferiorly, and the right ventricular wall is extremely thin

EA is not just a disorder of the tricuspid valve but also affects the right ventricle (RV) myocardium [3]. It encompasses a wide anatomical spectrum and the disorder can present itself either as cyanosis in the infant or exercise intolerance in the older adult [4, 5]. Symptomatic infants represent a distinct group of very sick patients with serious medical and surgical challenges [6].

Infants with symptomatic Ebstein's anomaly have a poor prognosis, with an expected mortality rate of 50% to 75% [7]. Previous attempts at palliative surgical treatment in this neonatal group have produced equally dismal results [7-9].

2. Classification of Ebstein anomaly

There are several classification systems for Ebstein anomaly. The most commonly used are the Carpentier classification (anatomical classification) (Figure 3) and the Celermajer index or the Great Ormond Street Echocardiography (GOSE) score for neonates.

These classification systems are crucial for determining the severity of the Ebstein anomaly.

2.1. Carpentier classification [4]

2.1.1. Type A: mild

- Mild apical displacement of the septal and posterior tricuspid valve leaflets
- Small "atrialized" ventricle
- Adequate function of the functional right ventricle (frv)

2.1.2. Type B: moderate

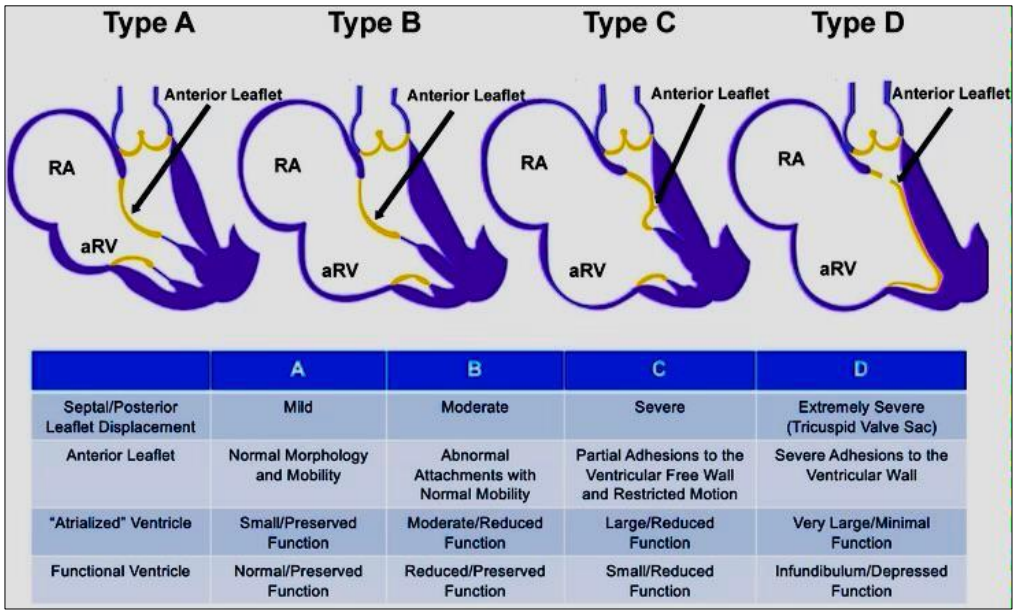
- Moderate apical displacement of the septal and posterior tricuspid valve leaflets
- Abnormal attachment of the otherwise freely mobile anterior leaflet
- Moderately sized "atrialized" ventricle with reduced function
- Reduced volume but adequate function of the FRV

2.1.3. Type C: severe

- Severe apical displacement of the septal and posterior tricuspid valve leaflets
- Abnormal attachments restricting the mobility of the anterior leaflet and potentially obstructing the right ventricular outflow tract
- Large "atrialized" ventricle with reduced function
- Small frv

2.1.4. ***type D:*** *extensive (tricuspid valve sac)*

- Complete non-delamination of the tricuspid valve leaflets
- Multiple abnormal attachments
- Almost complete "atrialization" of the right ventricle with the exception of a small insufficient infundibular component.



	A	B	C	D
Septal/Posterior Leaflet Displacement	Mild	Moderate	Severe	Extremely Severe (Tricuspid Valve Sac)
Anterior Leaflet	Normal Morphology and Mobility	Abnormal Attachments with Normal Mobility	Partial Adhesions to the Ventricular Free Wall and Restricted Motion	Severe Adhesions to the Ventricular Wall
"Atrialized" Ventricle	Small/Preserved Function	Moderate/Reduced Function	Large/Reduced Function	Very Large/Minimal Function
Functional Ventricle	Normal/Preserved Function	Reduced/Preserved Function	Small/Reduced Function	Infundibulum/Depressed Function

Figure 3 Carpentier classification of Ebsteins anomaly- Diagrammatic illustration.

2.2. GOSE scoring system

First reported by Celermajer et al [10], the Great Ormond Street Echocardiogram (GOSE) score has important prognostic value in stratifying risk of mortality (Figure 4).

2.3. Mortality prediction based on GOSE score.

GOSE score		
GOSE score	**Ratio**	**Mortality**
I	<0.5	8%
II	0.5-1.0	8%
III (acyanotic)	1.1-1.4	10% early, 45% late
III (cyanotic)	1.1-1.4	100%
IV	>1.5	100%

Figure 4 Gose Score.

2.4. Case Report

An 11 month severely breathness and cyanotic female infant was referred to our cardiology OPD for comprehensive color echocardiography and opinion regarding management of cyanotic congenital heart disease.

On clinical examination, the child was of average built highly irritable and continuously crying (Figure 5). There was significant central cyanosis present, which was increasing on crying.

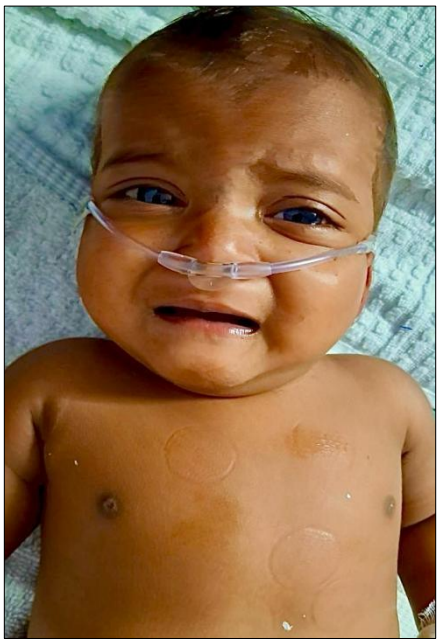


Figure 5 Facial appearance of our index patient

The infant's weight was 6.6 kg, height was 64 cm, BP was 80/60 mmHg, HR was 110/min, respiratory rate was 26/min and SPO2 was 77% at room air and 97 % on continuous oxygen. All the peripheral pulses were normally palpable without any radio-femoral delay. Cardiovascular examination revealed LVS_3 and LVS_4 gallop sounds over apex and multiple clicks during systole at left lower sternal edge. Systemic examination was normal.

Xray chest (PA) view demonstrated severe cardiomegaly with reduced pulmonary blood flow (Figure 6).

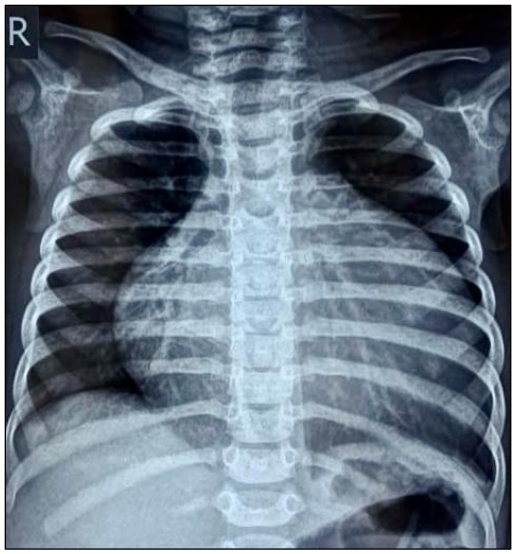


Figure 6 X-ray chest PA view depicting severe cardiomegaly with reduced pulmonary blood flow

2.4.1. Resting ECG exhibited the following features (Figure 7):

- Himalayan P waves in L_1, L_2, AVF, V_3-V_6
- QRS axis of + 120°.
- Sinus tachycardia which ventricular rate of 132/min.

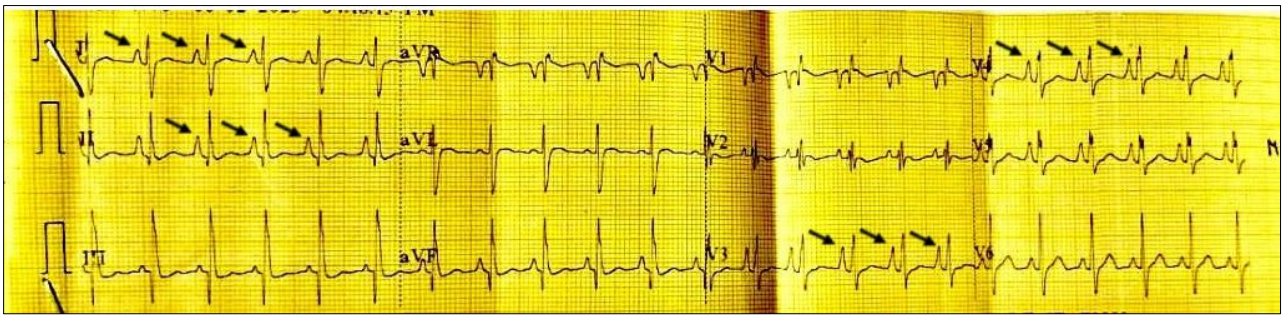


Figure 7 Resting ECG exhibited Himalayan P waves in leads L_1, L_2, AVF, V_3-V_6, QRS axis of +120°, and sinus tachycardia with ventricular rate of 132/min; Black arrows pointing towards Himalayan P waves

2.5. Transthoracic Echocardiography

All echocardiography evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using a pediatric probe equipped with harmonic variable frequency electronic single crystal array transducer while the subject was lying in supine and left lateral decubitus positions.

Conventional M-mode, two-dimensional, pulse wave doppler (PWD), continuous wave doppler (CWD) and sequential segmental transthoracic echocardiography was performed in the classical subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views.

2.6. M-mode Echocardiography

M-mode echocardiography of left and right ventricle was implemented for estimating various ventricular parameters (Table 1, Figure 8).

Table 1 Calculations of M-mode echocardiography.

Measurements	**RV**	**LV**
IVS d	5.5 mm	5.2 mm
ID d	18.1 mm	20.5 mm
PW d	6.4 mm	5.5 mm
IVS s	5.8 mm	7.0 mm
ID s	16.5 mm	12.2 mm
PW s	8.6 mm	13.2 mm
EF	20 %	74 %
%FS	8 %	40 %
EDV	9.8 ml	13.6 ml
ESV	7.8 ml	3.5 ml
SV	2.0 ml	10.0 ml
Mass	18 g	19 g
IVS, interventricular septum, ID, internal dimension; PW, posterior wall, d, diastole; s, systole; FS, fractional shortening; EDV, end-diastolic volume; ESV, end systolic volume; SV, stroke volume; EF, ejection fraction.		

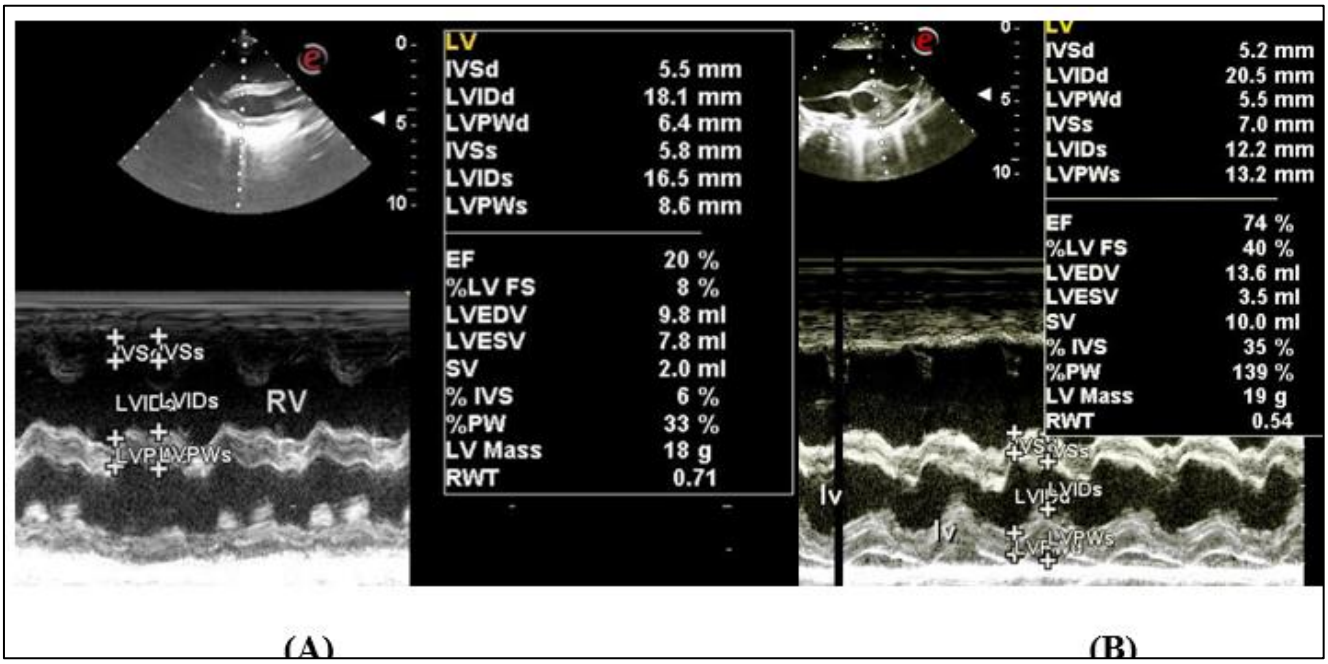


Figure 8 M-mode echocardiography (A) M-mode RV estimations; (B) M-mode LV estimations

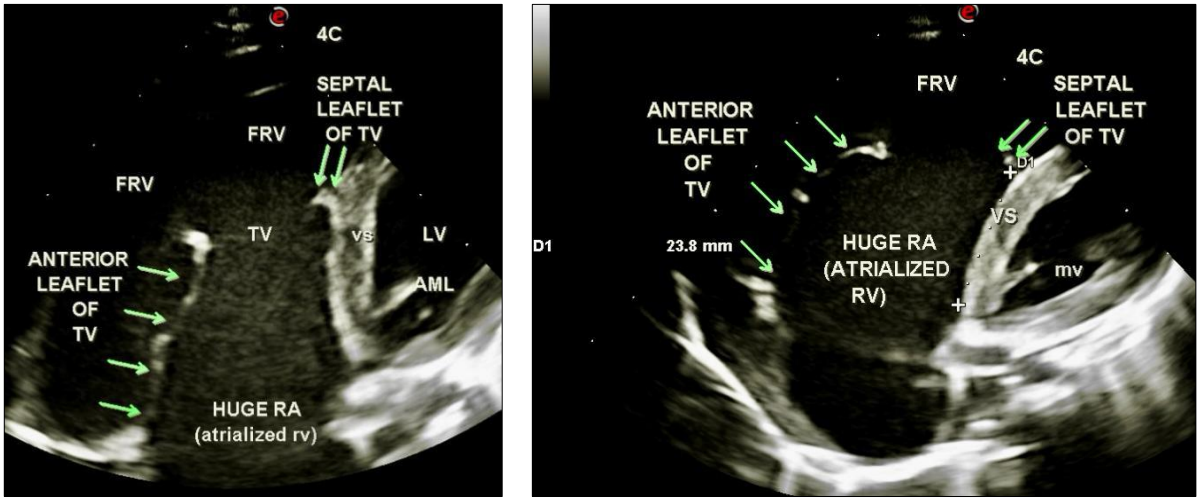
2.7. Summary of M-mode echocardiography

mode echocardiography depicted dilated FRV with severely reduced FRV EF (20 %). On the contrary the LV showed normal dimensions and LVEF (74 %)

3. 2-Dimensional-Transthoracic Echocardiography

2Dimensional transthoracic echocardiography (TTE) was conducted in explicit detail and it demonstrated typical features of EA:

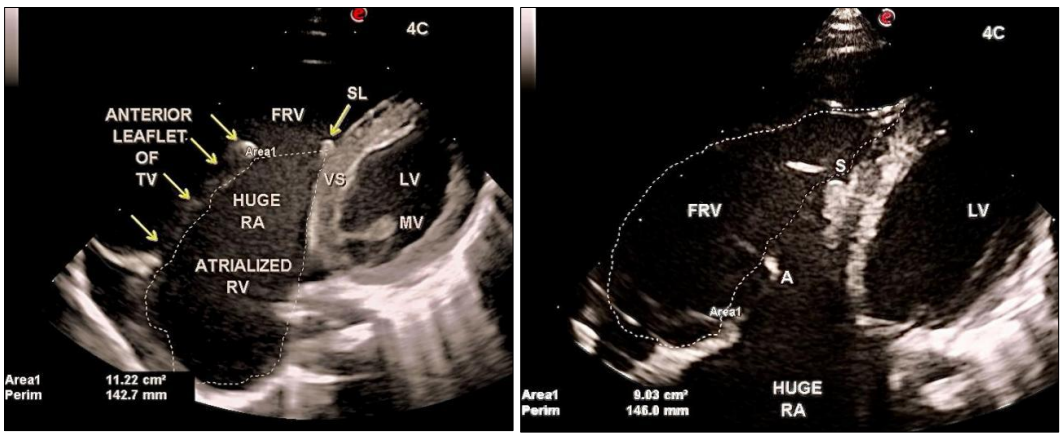
- Apical displacement of the TV (Figure 9).
- Apical displacement and dilation of TV annulus (Figure 9).
- Septal TV leaflet (STVL) was rudimentary and adhered to the ventricular septum (Figure 9).
- STVL was displaced by 27.2 mm from MV insertion (Figure 9).
- Anterior TV leaflet (ATVL) was large, sail like, redundant and attached to the AV junction (Figure 9).



(A) **(B)**

Figure 9 Transthoracic echocardiography. (A) and (B) demonstrates apically displaced rudimentary septal leaflet and large, sail like and redundant anterior leaflet of TV in apical 4C view. FRV, functional right ventricle; TV, tricuspid valve; AML, anterior mitral leaflet; LV, left ventricle; mv, mitral valve.

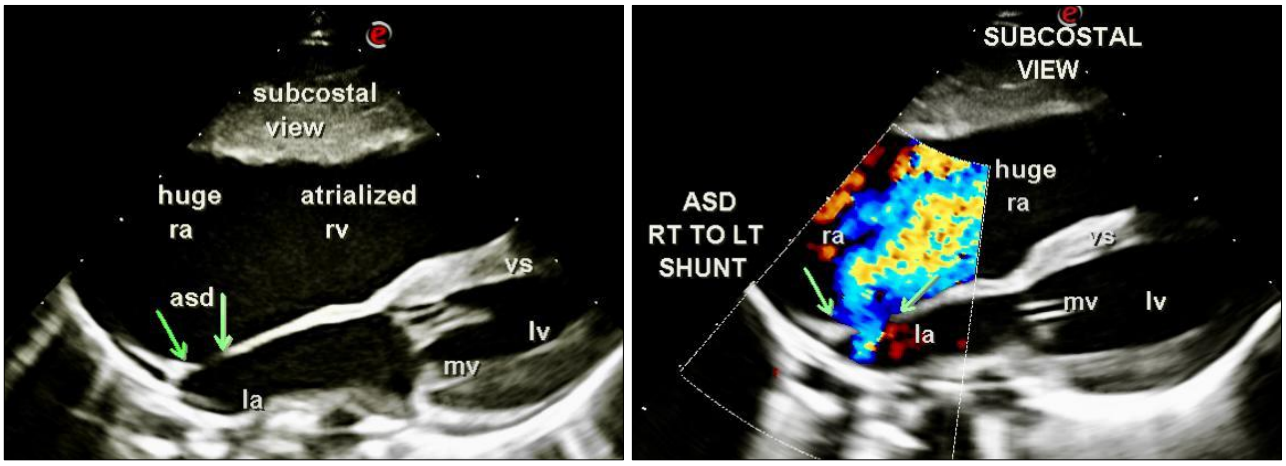
- Huge dilatation of RA (atrialized RV). The area of atrialized RV was 11.22 sqcm (Figure 10).
- Dilated FRV. The area of FRV was 9.03 sqcm (Figure 10).



(A) **(B)**

Figure 10 (A) 4C view showing huge RA (atrialized RV) with an area of 11.23 sqcm; (B) FRV is dilated having an area of 9.03 sqcm; TV, tricuspid valve; FRV, functional right ventricle; SL, septal leaflet; LV, left ventricle; MV, mitral valve; S, septal leaflet; A, anterior leaflet

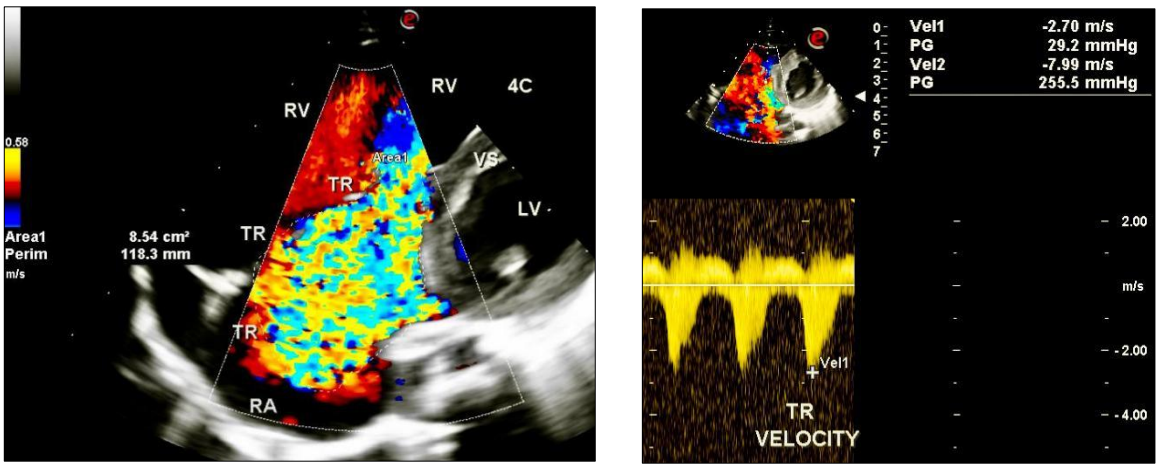
- Moderate ostium secundum ASD of size 4.5 mm with right to left shunt (Figure 11).



(A) (B)

Figure 11 (A) Moderate size ostium secundum ASD detected in the subcostal view; (B) Right to left shunt across ASD. ra, right atrium; rv, right ventricle; lv, left ventricle; mv, mitral valve; la, left atrium

- Severe TR with a jet area of 9.67 sqcm, occupying nearly whole of RA with a TR velocity of 2.6 m/sec (Figure 12).



(A) **(B)**

Figure 12 (A) On color flow mapping, a severe TR jet with an area of 8.54 sqcm is clearly demarcated in the apical 4C view; (B) On continuous wave doppler analysis across TV valve, a TR jet with a peak velocity of 2.70 m/sec (gradient 29.2 mmHg). TR, tricuspid regurgitation; RV, right ventricle; RA, right atrium; VS, ventricular septum; LV, left ventricle

3.1. Summary of Color echocardiography

On transthoracic color echocardiography, classical features of EA were displayed. According to the Carpentier's classification our index patient was Type C, even though the FRV was dilated with severely reduced RV systolic function (FRV EF 20 %).

Subsequently, the patient's attendants were advised immediate surgical correction of EA and the infant was referred to a tertiary care pediatric cardiovascular institute.

4. Discussion

EA is a rare congenital cardiac abnormality involving the tricuspid valve and the right ventricle (RV) [11]. Ebstein anomalies comprise < 1% of congenital heart defects [12]. The anomaly was initially described by the pathologist Wilhelm Ebstein in 1866 after performing an autopsy on a 19-year-old cyanotic male with exertional dyspnea and palpitations who died of a sudden cardiac arrest [13].

4.1. Associated Anomalies [2]

Abnormalities commonly associated with Ebstein anomaly include:

- Secundum atrial septal defect
- Variable degree of RV outflow tract obstruction.
- Functional or anatomical pulmonary atresia
- Ventricular septal defects, tetralogy of Fallot
- Transposition of the great arteries
- Atrioventricular canal defects.

In general, the clinical manifestations of Ebstein anomaly can range from asymptomatic to severe, depending on the degree of tricuspid valve displacement and severity of regurgitation, the effective right ventricular volume, and the associated malformations [14].

Atrial tachycardias, including atrial fibrillation, atrial flutter, or ectopic atrial tachycardia, can occur in 25% to 65% of patients with Ebstein anomaly. Additionally, 10% to 25% of patients have one or more accessory pathways, which increase the risk of protracted arrhythmias that can produce cardiac failure and sudden cardiac death [15, 16].

4.2. Etiopathogenesis

Ebstein anomaly is thought to be associated with chromosome 15q duplications during embryological cardiac development. There are also case reports suggesting a possible association of Ebstein anomaly with chromosome 11q rearrangements. Other reports indicate an association between Ebstein malformation and mutations in MYH7; MYH7 mutations are frequently associated with cardiomyopathies [17, 18].

Another known association of Ebstein anomaly is the teratogenic effect of lithium. Case-control and cohort studies of pregnant women taking lithium in the 1970s and 1980s demonstrated a risk of Ebstein anomaly in < 2% of their newborns [19]. Ebstein anomaly has also been associated with maternal exposure to benzodiazepines and varnishing materials [16, 20].

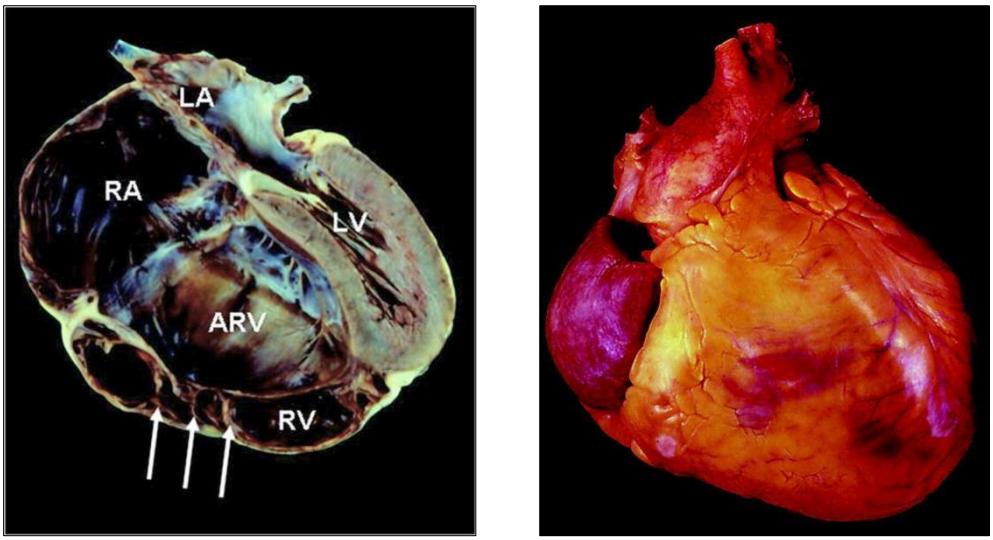
4.3. Epidemiology

Ebstein anomaly accounts for 0.3% to 0.6% of all congenital heart defects and is reported to occur in 0.2 to 0.7 per 10,000 live births [21]. Most cases of Ebstein anomaly are sporadic, with no identifiable etiology. Some studies suggest that there may be a familial inheritance, as has been observed in monozygotic twins [22]. There is a higher incidence of recurrence in the offspring of affected women (6%) than in the offspring of affected men (1%) [23].

4.4. Pathophysiology

4.4.1. Anatomic Abnormalities

The main pathophysiological abnormality of Ebstein anomaly is the failure of delamination of the tricuspid valve leaflets from the interventricular septum in utero [24]. The apical displacement mainly affects the posterior and septal leaflets, leading to the apical displacement of the tricuspid annulus and anteroapical displacement of the tricuspid orifice [3, 22] (Figure 13).



(A) (B)

Figure 13 Ebsteins Anomaly-Anatomical abnormalities. (A) Severe Ebstein's malformation of tricuspid valve (4-chamber view) showing marked downward displacement of shelf-like posterior leaflet with attachment to underlying free wall by numerous muscular stumps (arrows), markedly dilated atrialized portion of right ventricle (ARV), small functional portion of right ventricle (RV), leftward bowing of ventricular septum, and marked dilatation of right atrium (RA). (B) Pathological specimen depicting gross right sided chamber dilation. LA indicates left atrium; LV, left ventricle

This apical displacement has also been described as a rotational displacement of the tricuspid leaflets towards the RV outflow tract. The displacement is usually defined as > 8mm/m2 displacement of the septal leaflet from the anatomic tricuspid annulus [25]. The anterior leaflet of the tricuspid valve often has abnormal chordal attachments and becomes hypermobile, described as "sail-like." Alternately, there may be tethering of the anterior leaflet that causes restricted motion [3, 24]. There may also be fenestrations of the anterior leaflet [15].

The RV becomes split into two portions: the "atrialized" and the "functional" RV. The atrialized RV is where the tricuspid valvular inflow should normally be, extending from the undisplaced tricuspid annulus to the functional RV. The atrialized RV receives the regurgitant flow from the tricuspid regurgitation and becomes dilated along with the right atrium (RA). The FRV can be very small and may consist of just the RV outflow tract in cases of severe apical displacement of the septal and posterior leaflets of the tricuspid valve [12, 15].

More than 80% of patients with Ebstein anomaly also have a secundum atrial septal defect or a patent foramen ovale through which paradoxical emboli can travel [26]. Ventricular septal defects and pulmonary atresia have been concurrently found with Ebstein malformation [22]. One study of 539 patients at the Mayo Clinic demonstrated the incidences of the following cardiac abnormalities and Ebstein anomaly: atrial septal defect or patent foramen ovale in 84%, ventricular septal defect in 4.3%, pulmonary stenosis requiring surgical intervention in 6%, and accessory conduction pathways in 14% [16].

Ebstein anomaly associated with conduction system abnormalities is thought to be due to compression of the atrioventricular node by the septal leaflet, disrupting its function. Accessory pathways are common and often multiple [25]. Furthermore, 5% to 25% of patients with Ebstein anomaly have been reported to have Wolff-Parkinson-White (WPW) syndrome, making Ebstein anomaly the most commonly found congenital heart defect associated with WPW [27].

4.4.2. Functional Abnormalities

The faulty coaptation of the tricuspid leaflets due to their anatomic displacement and tethering abnormalities leads to tricuspid regurgitation. The degree of tricuspid regurgitation is dictated by the severity of the apical displacement of the posterior and septal leaflets, the abnormality of the anterior leaflet attachments, and the integrity of the leaflets themselves [22, 24]. The atrialized RV is often dyskinetic from fibrosis and myopathy, and the functional RV can become enlarged and dysfunctional from severe tricuspid regurgitation [22, 28] (Figure 14).

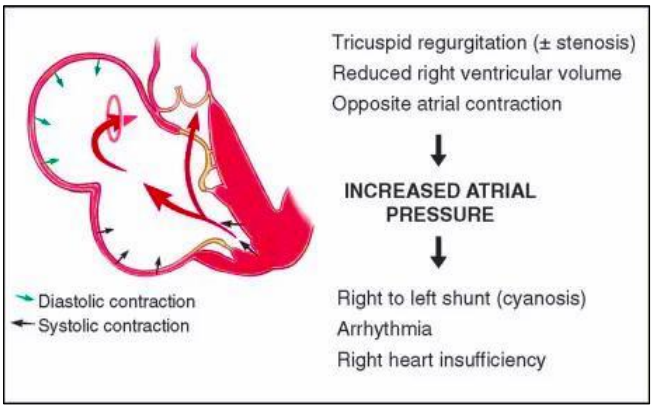


Figure 14 Pathophysiogy of Ebsteins anomaly. The hemodynamic consequence of Ebstein's malformation is predominantly tricuspid regurgitation Associated tricuspid stenosis is occasionally present. Tricuspid valve dysfunction in association with reduced right ventricular volume and opposing atrial contraction (i.e., diastolic in the atrium and systolic in the atrialized chamber) increase the pressure in the right atrium. The increased right atrial pressure, in association with the atrial septal defect, produces cyanosis, arrhythmias, and right heart insufficiency.

Right ventricular volume overload produces ventricular septal flattening with left ventricular dysfunction [22]. The frequently associated atrial septal defect or a stretched patent foramen ovale will permit right to left shunting from severe tricuspid regurgitation, resulting in hypoxemia and cyanosis [29].

4.5. Management

4.5.1. Medical Management

It is well known that in Ebstein anomaly, the mainstay of treatment is supportive :

- To reduce pulmonary vascular resistance
- To correct hypoxemia
- To improve the cyanosis
- Heart failure is treated with loop diuretics and guideline-directed medical therapy
- Patients with supraventricular tachyarrhythmias can receive rate-control medications such as beta blockers or calcium channel blockers. The other option is rhythm control with requisite medicine.
- Patients with intractable arrhythmias may require catheter ablation.

4.5.2. Surgical Management

Indications for surgery in neonates with Ebstein anomaly include:

- Right heart failure due to severe tricuspid regurgitation
- A cardiothoracic ratio > 80%
- Severe cyanosis with dependency on PGEI
- Mechanical ventilation dependency [30, 31].

In children and adults with Ebstein anomaly, indications for surgery include:

- Heart failure symptoms (New York Heart Association functional class III or IV)
- Progressive exercise intolerance
- Evidence of RV dysfunction or progressive RV dilatation
- Evidence of paradoxical emboli
- Arrhythmias refractory to medical and catheter ablation therapies
- Cyanosis with oxygen saturations < 90%
- Severe tricuspid regurgitation
- Significant concomitant lesions such as pulmonic stenosis or atrial or ventricular septal defects
- Progressive cardiomegaly
- Cardiothoracic ratio > 65%

- Reduced left ventricular function [30, 32, 33].

Surgical intervention comprises tricuspid valve repair and patch closure of the atrial septal defect [34]. Multiple surgical approaches have been described for the treatment of Ebstein anomaly. Cone reconstruction is the currently preferred surgical approach in young children and adults [35] (Figure 15).

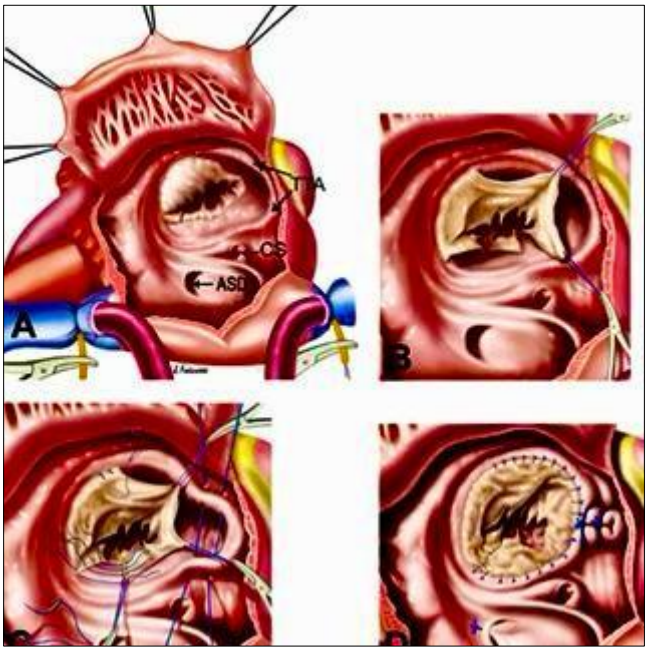


Figure 15 The Cone Reconstruction for Ebstein Anomaly. Tricuspid valve reconstruction with detachment and delamination of anterior, septal and posterior leaflet tissue, rotation and reattachment of the recruited leaflet tissue at the true atrioventricular junction/tricuspid valve annulus thereby creating a 360 degree leaflet "cone," plication of the tricuspid annulus to reduce its size, and internal plication of "atrialized" ventricle.

4.6. Prognosis

Decreased survival is seen in patients with Ebstein anomaly who do not undergo surgical repair, survival rates for these patients are 90% at 1 year, 75% at 10 years, 50% at 15 years, and 40% at 20 years [36]. Sudden cardiac death due to ventricular arrhythmias has been reported as an 8.6% in a 50-year cumulative incidence from birth [37].

Late postoperative outcomes in patients with Ebstein anomaly have shown an overall survival rate of 98% at 1 year, 94% at 4 years, 90% at 10 years, 86% at 15 years, and 76% at 20 years [26]. More specifically, after cone reconstruction, only 2% of patients needed reoperation at 6 years [38].

The Celermajer index or GOSE score [10] is an established score that uses echocardiography to predict mortality (Figure 16). It is measured as the ratio between the area of the RA and the atrialized RV to the combined area of the functional RV, the left atrium, and the left ventricle at end-diastole. Mortality increases with increasing grade.

- Grade 1: ratio less than 0.5; mortality of 0%.
- Grade 2: ratio of 0.5 to 0.99; mortality of 10%.
- Grade 3: ratio of 1.0 to 1.49; mortality of 4%.
- Grade 4: ratio greater than 1.5; 100% mortality.

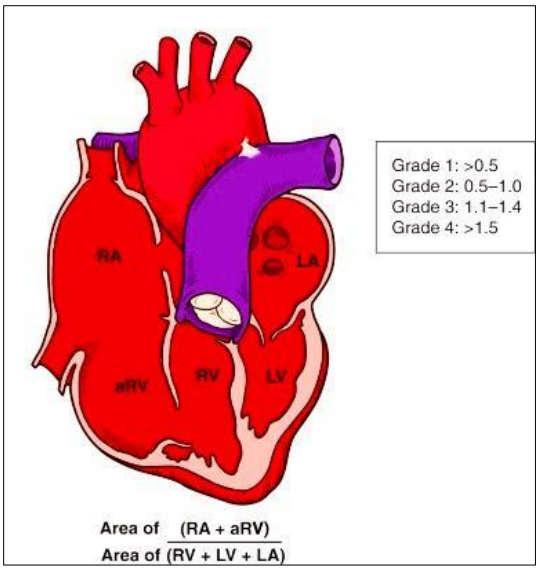


Figure 16 Celermajer index-figurative portrayal. RA, right atrial; aRV, atrialized right ventricle; RV, right ventricle; LA, left atrium; LV, left ventricle

4.7. Complications in Patients with Unrepaired Ebstein Anomaly

4.7.1. The most common complications in this subset of patients include:

- Heart failure resulting from right or biventricular dysfunction or tachyarrhythmias
- Intractable atrial or atrioventricular arrhythmias
- Paradoxical emboli resulting in myocardial infarction or cerebrovascular accident
- Sudden cardiac death due to tachyarrhythmias or heart failure [15].

4.7.2. Long-term predictors of death in patients with unrepaired Ebstein anomaly include:

- Severe tricuspid regurgitation
- Cardiothoracic ratio ≥ 65%
- Impaired pulmonary growth and function from cardiomegaly
- Impaired left ventricular function from RV dilatation with consequent compression and displacement of the interventricular septum
- NYHA functional class III or IV
- Cyanosis
- Early age at diagnosis [36]

5. Conclusion

Ebstein's anomaly is a complex birth defect with a broad anatomic and clinical spectrum. Management is complex and must be individualized. Accurate knowledge of the various anatomic and hemodynamic variables, associated malformations, and management options is essential. Thus, patients with Ebstein's anomaly must be regularly evaluated by a cardiologist who specializes in congenital heart disease. With improved management strategies, it is hoped that the survival of patients with this anomaly of all ages will continue to improve.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no conflict of interest.

Statement of ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional committee of Prakash Heart Station, Nirala nagar, Lucknow.

Statement of informed consent

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

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