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Ebstein's Anomaly: Epidemiology, Causes and Management Options

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ABSTRACT

Ebstein's abnormality is an intricate congenital deformity of the tricuspid valve. Newborn and infant health is an oftendiscussed topic. The condition exhibits a wide range of severity and medical characteristics, necessitating tailored treatment for each patient. This article provides an account of the inherent characteristics and progression of Ebstein's abnormalities in children and adolescents. This includes the indications and manifestations that appear at the time of diagnosis. This section describes the contemporary methods used to categorize Ebstein's abnormalities. We provide diagnostic techniques for assessing the severity of illnesses that have the potential to enhance decision-making in surgical procedures. Furthermore, we explore several surgical alternatives for neonates in severe conditions, as well as many surgical procedures following the operation. In order to effectively enhance both the duration and quality of life for this complex and diverse group of patients, it is essential to possess sufficient information and comprehension of the aforementioned factors. Effective management requires a nuanced and personalized approach. It is crucial to have an accurate understanding of the many anatomical and hemodynamic factors, related abnormalities, and available treatment choices.

Keywords- Tricuspid Valve, Ebstein Anomaly, Manifestations, Hemodynamic factors.

I. INTRODUCTION

In 1866, Wilhelm Ebstein first described the Ebstein anomaly (EA). It is a condition that shows septal plates and originates from the right ventricular myocardium below the tricuspid valve. EA, an uncommon congenital cardiac condition, affects approximately 2.4 to 10,000 live births. When seen through an embryological lens, EA reveals itself as a collection of symptoms brought on by different degrees of degeneration in the lower endocardium. Removing the tricuspid tract from its apex primarily damages the posterior portion. The myopathic right ventricle (RV) bifurcates into two sections: the bottom, known as the "active" section, and the upper, known as the "atrialized" section, which experiences distortion between the actual annulus and the hinge point of the apical displacement septal tract. Just a scrap of paper. The amount of leaf movement has a direct correlation to the active RV volume. EA symptoms can range from minor in older adults to quite severe in younger children, and even fatal in very young infants. Ectopic abortion causes irregular heartbeats and fluid retention in the developing baby. At EA Cervical Diagnostics, there is a 48% infant mortality rate. Significant medical and surgical obstacles contribute to a neonatal mortality rate of 17–56% [1-5].

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II. DISEASE MECHANISMS

The foetus lowers the right ventricular wall to the right as the tricuspid valve tracts and chordae grow. The atrioventricular (AV) level is the next stop in this progression. The deterioration process in Ebstein's anomaly stops short of reaching the level of contact with AVs. Furthermore, the reattached apical portions of the valve tissue are incompletely healed. As a result, the right ventricle atrializes, and the tricuspid valve passageways distort and move. A study of fifty abnormal hearts revealed the whole right ventricle to be morphologically defective. Ebstein's anomalies typically link to common congenital, structural, or systemic illnesses such as intracardiac shunts, venous ulcers, and circulatory problems (such as Wolf-Parkinson-White syndrome [WPW]) [6-10].

The arterialization of the right ventricle and the removal of the tricuspid tracts both affect the heart's blood flow because of the Ebstein abnormality. Disruption of traction triggers tricuspid re-entry. From gentle repositories with sluggish tricuspid tracts to huge displays and difficult retrieval, the difficulty of repositioning is proportional to the magnitude of the tract transfer. While it is an atomic component of the right atrium, the arterialized section of the right ventricle contracts and relaxes in tandem with the right ventricle. This discordant contraction causes the right atrium to clot. Figure 1 illustrates how the atrialized right ventricle connects to the full right ventricle during ventricular systole, aggravating tricuspid regurgitation and returning blood to the right atrium [10,15].

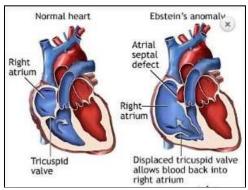


Figure 1: Normal Heart and Ebstein Anomaly Heart [3]

III. CAUSES

Many cases of Ebstein's abnormality, a congenital condition, go undetected. Some environmental variables that may have a role in etiology include the following maternal traits: the mother's history of preterm birth, the mother's exposure to alcohol, the mother's usage of benzodiazepines, and people. White people are more likely to have Ebstein's anomaly. Unlike other ethnic groups, Strips, or flaps, make up the three main components of the tricuspid valve. The right atrium (upper chamber) and the right ventricle (lower chamber) can open to allow blood to circulate while the heart is resting. They shut down to prevent blood from leaking from the right ventricle into the right atrium while the heart pumps. In healthy individuals, the tract typically sits above the right ventricle, but in those with Ebstein's abnormality, it sits below. Typically, the tube is bigger than normal. A malfunctioning valve can cause abnormal blood flow. When blood returns to the right atrium, it bypasses the lungs. Raising the heart rate and the body's water content are two side effects of promoting blood flow. It is possible for the pulmonary valve, which links to the lungs, to constrict. A problem with the valves leads to abnormal blood flow. When blood returns to the right atrium, it bypasses the lungs. Raising the heart rate and the body's water content are two side effects of promoting blood flow. It is possible for the pulmonary valve, which links to the lungs, to constrict [16-20].



Figure 2 Symptoms of Ebstein's Anomaly In adults [4]

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IV. EPIDEMIOLOGY

An Ebstein abnormality causes approximately 9.60% of all congenital cardiac abnormalities. Its exact distribution remains a mystery due to the lack of readily accessible soft forms. The extensive use of echocardiography has led to several instances being detected. Factors pertaining to gender, age, and race White women's offspring have a higher prevalence of the Ebstein abnormality. But there aren't any particular sexual inclinations. The following are some of the life stages in which an Ebstein anomaly may manifest: In pediatrics, an echocardiogram is the gold standard for identifying an Ebstein abnormality. Infants and children's health: When resistance to the pulmonary arteries is poor, symptoms in children typically appear; the Ebstein anomaly is a sign of cyanosis and/or severe heart failure. Fatigue, worsened dyspnea, cyanosis, tricuspid regurgitation, right heart failure, palpitations, and an irregular pulse are also symptoms of Ebstein in adults (Figure 3) [21-30].

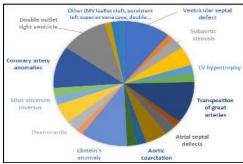


Figure 3: Prevalence of Ebstein Anomaly among all congenital heart diseases [5]]

V. PROGNOSIS

The level of predictability depends on how serious the condition is and what treatments are available. Women with Ebstein abnormalities seem to tolerate pregnancies well, assuming proper monitoring. The following are examples of negative predictive indicators: Male gender, Presentation history Chest radiographs showing a cardiothoracic score greater than 0.65, Septal leaflet attachment ratio (septal to distal) is the average distance from the AV ring to the distal attachment of the leaflet. The length of the septal tract should be greater than 0.45. Classification by the New York Heart Association (NYHA) is based on the following criteria: The New York Heart Association (NYHA) classifies patients based on the following criteria: elevated right ventricular area to active right ventricle (grade 1), below 0.5, up to grade 4, and over 1.5 (associated risk of 2.7 per grade increase). Some studies have shown a correlation between this and death rates (Table 1) [31-35].

Table 1: Classification of Ebstein Anomaly [6]

Carpentier Classification system	Celermajer Scoring system (GOSE) (Rightatrium atrialized RV)/(Functional RV teft atrium left ventricte) based on combined area
Type A: Adequate volume of the true RV	Grade <0.5
Type B: Large atrialized portion of the RV with freely mobile anterior leaflet	Grade 2: 0.5-0.99
Type C: Restrictive mobility of the anterior Leaflet	Grade 3: 1.0-1.49
Type D: Almost complete atrialization of theRV	Grade 1.5

Mortality: The disease's natural progression differs according to the magnitude of the tricuspid valve's displacement. In younger people, poor diagnoses and more severe diseases are common. The presentation's average age is in the middle years. Extant observational evidence indicates that around 5% of these individuals survive to be 50 years old or older. At 85 years old, the patient's age was the highest recorded. Paralysis, paradoxical embolism, brain tumors, heart failure, constipation, abrupt cardiac death, bacterial endocarditis, and acute ischemia damage are all potential consequences of Ebstein's disease [7].

Signs and symptoms: Patients may experience the hemodynamic repercussions of Ebstein's abnormality and other structural and surgical system disorders, as well as a wide range of symptoms. The most prevalent cause of cyanosis is extreme heart failure, or the right-to-left migration of the little finger. Here we are in the life cycle of a baby, which mirrors that of an adult. It might not show up until later in life. In adults, paroxysmal arrhythmias cause cyanosis, which can be

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temporarily severe and develop suddenly. When it shows up, cyanosis becomes more severe. When the left ventricle fails and the right ventricle fails as well, it leads to heart failure, which manifests as fatigue and dyspnoea. Paroxysmal supraventricular tachycardia (SVT) and Wolff-Parkinson-White syndrome (WPW) can lead to heart failure and sudden cardiac death in as many as one-third of individuals. Because these pathways are present, fatal ventricular arrhythmias can happen. Ankle edema and ascites are signs of right heart failure. The following are some other common, but minor, symptoms: These symptoms include bacterial endocarditis, paradoxical embolism, stroke, transient ischemic attack, right-to-left shunt, and brain tumour.

Diagnosis: In most cases, a heart murmur or other abnormal heart sound is not a reason for alarm. Nonetheless, in order to ascertain the root cause, your primary care physician or your child's physician would probably suggest that you consult a cardiologist [36].

Echocardiogram: This test uses sound waves to generate a thorough picture of your heart. It reveals how your heart's tricuspid valve developed and how blood flows to it. Every now and then, doctors perform a transesophageal echocardiogram. This examination involves threading a tiny transducer down a tube that travels from the neck (esophagus) to the stomach (gastrointestinal tract). Because the neck is close to the heart, this exam will provide a clearer picture of your heart Figures-4 (8).

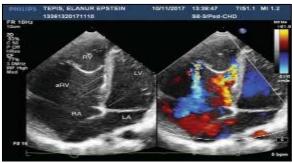


Figure 4: Echocardiogram of Ebstein Anomaly [8]

Electrocardiogram (**ECG**): Many people with Ebstein anomaly do not commonly have ECG abnormalities. Right atrial enlargement can manifest as elongated and broad P waves, along with either total or partial bundle branch obstruction. The amplitude of the R waves on lead V1 and V2 is diminished. The unusual structure of the QRS terminal pattern is caused by a disturbance in infra-Hisian conduction and aberrant activation of the atrialized right ventricle (figure 5) [38-45].



Figure 5: Electrocardiogram of Ebstein Anomaly [9]

Holter monitor: Holter monitor is a portable ECG device that you wear out of your doctor's office. It records the electrical activity of your heart as you do your normal activities for a day or two (figure 5)[46-50].

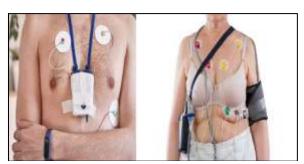


Figure 6: Holter Monitoring Male and Female

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Chest X-ray:

A chest X-ray is a picture of your heart, lungs and blood vessels. She can tell your doctor if your heart is enlarged (figure 6) [11].



Figure 7: Chest X-ray of Ebstein Anomaly

Cardiac MRI'':

Cardiac MRI uses magnetic fields and radio waves to create detailed images of your heart. This test will give your doctor a detailed view of your tricuspid valve. This allows your doctor to see the size of your heart chambers and how well they perform (figure 7) [51-55].



Figure 8: Cardiac MRI of Ebstein Anomaly

Pulse oximetry:

This test measures the amount of oxygen in your blood by attaching a sensor to your finger or toe [13].

Exercise stress test:

Whether you're pressing a button or riding a stationary bike, this test will keep tabs on your vitals. One way to find out how your heart handles exercise is to get a stress test. The doctor can use this information to establish a safe exercise level.

Electrophysiology study (EP)

The procedure involves mapping the heart's output by inserting a thin, flexible catheter into the circulation and connecting it to the inner cardiac electrodes. Additionally, with the use of electrodes, physicians can cause or halt arrhythmias, stimulate the heartbeat, and more. This will aid medical professionals in ascertaining the drug's efficacy in addressing arrhythmia.

Cardiac catheterization

Patients only require cardiac catheterization during preoperative coronary angiography to diagnose Ebstein's abnormality. People with the anomaly typically have normal pulmonary arteries and ventricular pressure, despite the possibility of increased ventricular pressure at the diastolic end. Even if there is a lot of tricuspid duplication, the right atrial pressure could be normal if the right atrium is quite open. Oximetry may show that the arterial system has ruptured prior to the right and left ventricles making contact with the atrium [56-60].

Medications

Medications can help people with cardiac rhythm disorders maintain a normal heart rate and manage their heart rate. Your doctor may prescribe medication, such as water tablets, to alleviate the symptoms and indications of heart

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failure. Your doctor may recommend blood thinners if they detect an irregular heartbeat or an atrial septal defect, which is a hole in the wall that separates the upper chambers of the heart. Certain children receive an inhalant known as nitric oxide to improve blood circulation to the lungs. Because of the effects on the heart caused by tricuspid atrialization of the right ventricle, valve repair, and septal abnormalities in Ebstein's abnormality, medication is needed. Antibiotic prophylaxis may be necessary for patients at risk of bacterial endocarditis. Radiofrequency ablation, as opposed to medication treatment, is the standard method for treating SVT. Digoxin, ACE inhibitors, and diuretics are among the medicines used to treat congestive heart failure [15].

Operations like surgery:

If your symptoms are causing your health problems, your doctor may recommend surgery. Your doctor may suggest surgery as a solution if your heart rate increases or decreases. In the event that surgical intervention is necessary, it is critical to select a surgeon with extensive training and expertise in the area. A number of methods can accomplish the surgical correction of Ebstein anomalies and associated malformations.

Tricuspid valve repair:

In order for the valve to function properly, surgeons narrow the hole and allow the valve leaflets to join. Wrapping a piece of tape around the valve ensures its stability. People often perform this operation once the valve tissue has sufficiently healed. Cone reconstruction is a relatively recent method for repairing the tricuspid valve. The surgeons remove the tricuspid valve leaflets from the heart muscle. After that, they coil up into a structure known as a "leaflet cone." Your valve can eventually require maintenance or replacement (Figure 8).

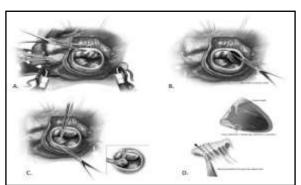


Figure 9: The Cone Reconstruction for Ebstein Anomaly [16]

Surgery to repair the tricuspid valve:

If restoration is not possible, your surgeon may remove the damaged valve and implant a bioprosthetic tissue or mechanical valve in its place. Mechanical valves replace tricuspid valves, which is a rare occurrence. Blood thinners are required to prevent blood clots in patients with mechanical valves. Preventing endocarditis, an inflammation of the heart lining, is necessary for patients with prosthetic valves (Figure 9) prior to dental treatments [61].

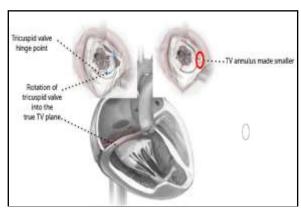


Figure 10: Tricuspid valve replacement in case of Ebstein Anomaly Closure of the atrial septal defect

If there is a hole between the upper chambers of the heart (atrial septal defect), your surgeon may repair or replace the defective valve. Your surgeon may also be able to correct some of your heart problems during this operation [62].

Maze procedure

If patients have a rapid heartbeat, their surgeon may perform a Maze procedure during valve repair or other surgery. In this procedure, your surgeon will make a maze scar or by making small holes in the upper chambers of your

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heart. Because red tissues do not absorb electricity, they interfere with the disappearance of heart symptoms, which can lead to other types of arrhythmias. Cold therapy (radiotherapy) or heat (radio frequency) energy can also be used to make scars.

Radiofrequency catheter ablation:

If the patient has a fast or abnormal heartbeat, the doctor may prescribe the procedure. The doctor connects one or more catheters to your heart through your blood vessels. The nerves at the end of the catheter use heat (radiofrequency ablation) to damage a small area of tissue in the heart. This will prevent the unusual symptoms that are causing your arrhythmia. Some people may need repeated procedures (figure 10) [63-70].

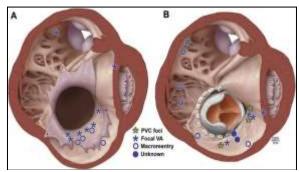


Figure 11: Sites for Catheter Ablation for Ventricular Arrythmia in Ebstein AnomalyPatients [20]

Heart transplantation

If the patient has severe Ebstein anomaly and poor heart function, a heart transplant might benecessary.

VI. EVALUATION

Individuals with an Ebstein abnormality should undergo regular screenings at a center that specializes in congenital heart disease (CHD). For adults with an Ebstein anomaly, the recommended diagnostic investigations are Class I electrocardiography (ECG), radiography, and echocardiography-Doppler ultrasonography. Class IIa: Diagnosing EbsteiRelaxation techniques and/or pulse oximetry may aid in the diagnosis of Ebstein anomalies in (Level of certification: C) If it is clinically feasible, an electrophysiologic study (EPS) can help identify aberrant Ebstein patients in the elderly who have a history of or suspicion of supraventricular arrhythmia. The procedure may be beneficial for men aged 35 and up, premenopausal women aged 35 and up with malignant coronary arteries, and postmenopausal women. (All evidence level is B) [21].

VII. MANAGEMENT

Patients who have a history of paradoxical embolus or atrial fibrillation and have an Ebstein abnormality should take warfarin for anticoagulation. (Class I, Category C Establishment Level). For adults presenting with an Ebstein anomaly, the following are the suggested procedures for catheter intervention: Only hospitals that specialize in catheterization and care for older patients with Ebstein abnormalities should insert catheters. EPS and pace problems (Class I, Level of Evidence: C) exist. In some cases, people with Ebstein abnormalities and recurrent supraventricular tachycardia may find relief from catheter ablation. (Phase IIa, level of evidence: B), Surgeons with training and expertise in congenital heart disease (CHD) should repair or replace the tricuspid valve while partially closing the atrial septal defect (ASD), if possible, for patients with an Ebstein anomaly who exhibit the following symptoms or degeneration of exercise volume: cyanosis and hypertension.

Adults diagnosed with Ebstein's abnormality who exhibit the following symptoms (degree of evidence: B, all) should have modified tricuspid valve surgery or replacement: Bioartificial tricuspid valve, atrial and/or ventricular arrhythmia development or progression, mixed recurrence, stenosis, weak resilience, or New York Cardiac Association (NYHA) III or IV Performance category; stronger tricuspid regurgitation (TR); continuous RV dilation, contraction, or contractile function; and bioartificial tricuspid valve We can anticipate early procedure effectiveness if the bioprosthetic stenosis is less severe, leading to less severe symptoms and increased exercise tolerance after valve correction. There are additional factors to consider during management. Pregnant women who are of reproductive age or who have Ebstein's abnormality should consult an adult physician with expertise in congenital heart disease (CHD) for counselling. (Class I, Category C Establishment Level). Keeping endocarditis at bay: Individuals who have had surgery for cyanotic anomalies or who have artificial heart valves should take antibiotic precautions before dental procedures involving gingival tissue or the region of teeth surrounding the apex of an oral piercing. (Standard of evidence: C, Class IIa) [20].

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VIII. DISCUSSION

Ebstein's anomalies in fixed valves are rare. The tricuspid valve separates the right ventricle, which pumps blood into the lungs, from the right atrium, which receives blood from the body. In Ebstein's anomaly, two tricuspid valve tips descend to the pump chamber's base. You may connect the third, lengthy sheet to the room's wall. Normal blood flow (from the right atrium to the right ventricle) becomes problematic in extremely rare instances when the valve becomes paralyzed. Squeezing the right ventricle typically results in the tricuspid valve leaking and returning to the right atrium. This causes the right atrium to expand. Serious cardiac failure can develop from tricuspid regurgitation if it is severe enough. Too much pressure builds up in the right atrium due to repetition. In a typical foot, the PFO, a closed hole, connects the right and left atriums. Typically, PFOs cease following delivery [21].

High right atrial pressure maintains an open PFO in Ebstein's anomaly. Bypassing the pulmonary system, this conduit enables the conveyance of non-oxygenated ("blue") blood from the right atrium to the left. As a result, blood oxygen levels will drop. The reason for this is Blue or "cyanotic" children with low oxygen saturation may be a symptom of Ebstein's abnormality. Heart conditions such as pulmonary valve stenosis, atresia, atrial septal defect, or ventricular septal defect can coexist with Ebstein's abnormality. Additionally, supraventricular tachycardia episodes can occur in many people with Ebstein's abnormality due to an additional cardiac conduction route, a disorder known as Wolff-Parkinson-White syndrome [22].

In the absence of Ebstein abnormalities, patients can be referred for percutaneous or surgical ASD closure, although the indication for intervention may change in their presence. We should approach patients undergoing right-to-left percutaneous ablation of the accessory tract or those with Ebstein abnormalities with caution when considering the combination of atrial fibrillation and shunt due to the potential for paradoxical embolism. If there are multiple access mechanisms, it is reasonable to suspect Ebstein anomalies. Even if they have obvious restrictions, patients with significant cardiomegaly and continence abnormalities may not complain of many symptoms. As part of their regular evaluations, these patients should undergo exercise testing to identify their performance limitations. Exercise can cause cyanosis, so it is important to check respiratory integrity during exercise testing. Patients recently diagnosed with Ebstein anomalies may also present with concurrent pulmonary arterial hypertension (PAH) when cyanosis and an elevated right heart rate are present. Since PAH is quite rare in Ebstein patients, this is frequently an incorrect diagnosis [71-82].

IX. CONCLUSION

The clinical and anatomical manifestations of the complicated birth abnormality known as Ebstein's anomaly are quite varied. Management requires a personalized approach due to its complexity. We must precisely understand the many anatomic and hemodynamic factors, related abnormalities, and treatment choices. Patients with Ebstein's anomaly should have routine evaluations by a cardiologist with expertise in congenital cardiac disease. It is believed that the prognosis for individuals of all ages affected by this defect may further improve as a result of more effective management techniques.

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