Ebstein Anomaly

Abstract

Background: Ebstein's anomaly is a complex congenital malformation of the tricuspid valve. Introduction to newborn and (baby) health is common. The severity and medical features of the disease vary widely and require appropriate treatment for the patient. This review describes the natural history of children and adolescents with Ebstein's anomalies. This includes the signs and symptoms that appear at the time of diagnosis. Explains current techniques for classifying Ebstein's anomalies. We report diagnostic methods for diagnosing the severity of diseases that may improve decisions during surgical intervention. In addition, we discuss various surgical options for critically ill newborns and several postoperative surgical interventions. Without adequate knowledge and understanding of the above, this complex and diverse group of patients can be properly treated to improve not only the duration but also the quality of life.

Conclusion: Management is complex and must be individualized. Precise knowledge about the different anatomic and hemodynamic variables, associated malformations, and management options is essential.

Keywords: *Arrhythmia; Congenital Heart Disease; Tricuspid Valve, Ebstein Anomaly.*

Introduction

Ebstein anomaly (EA) was first described by Wilhelm Ebstein in 1866, which identifies septal plates and occurs below the tricuspid valve from the right ventricular myocardium. EA is rare congenital heart disease with an incidence of 2.4 to 10,000 live births. From the embryological point of view, EA is the result of various degrees of degeneration from the lower endocardium, leading to several distinct features. There are various stages of apical removal of the tricuspid tract, then the posterior region is more affected. In addition, the right ventricle (RV) is myopathic and divided into two parts, the "atrialized" part deformed between the true annular and the hinge point of the apical displaced septal tract, while the RV is the "active" lower part. , A piece of paper. Active RV volume can be significantly reduced depending on the level of leaf movement. Clinical manifestations of EA vary widely from old age to mild forms to severe forms and high mortality in infant life. In the womb, EA can cause hydrops and arrhythmias. In addition, EA Cervical Diagnostics has an infant mortality

rate of 48%. Mortality at birth ranges from 17 to 56% and has serious medical and surgical challenges (1).

Review of literature

Ebstein anomaly makes up 9.60 % of all congenital heart disease cases. It's a type of right ventricular myopathy that causes significant regurgitation due to tricuspid valve delamination failure and extremely variable tricuspid valve morphology. It's a unique congenital heart defect that can manifest itself in a variety of ways, from a very symptomatic newborn to an asymptomatic adult. Surgery conducted from infancy and into adulthood has a high operative death rate, whereas operation performed after infancy and into adulthood has a low operating mortality rate. The majority of patients in all age groups have outstanding late survival and quality of life after being admitted to the hospital. The most prevalent late consequence is atrial tachyarrhythmias. More tricuspid repair approaches have been published in the literature than any other congenital or acquired cardiac defect. This is largely owing to the unlimited anatomical variety that this abnormality entails. The Ebstein anomaly cone reconstruction can produce a near-anatomic restoration of the tricuspid valve anatomy. These adjustments have yielded encouraging early and intermediate effects. Reduced right ventricular function, as well as the necessity for reoperation for recurrent tricuspid regurgitation, remains a difficulty for certain patients. The goal of this article is to explain creative ways for addressing poor right ventricular function and concomitant right-sided heart failure, as well as the current standard of care for diagnosis and treatment of the Ebstein anomaly (1).

Pathophysiology

Fetal development of the tricuspid valve tracts and chordae involves lowering the right ventricular wall to the right. This process progresses to the level of the atrioventricular (AV). In Ebstein's anomaly, this process of degradation is incomplete and does not reach the level of AV interaction. Also, the apical parts of the valve tissue, which are often reattached, do not heal completely. This causes the tricuspid valve passages to deform and shift, and the right ventricle to atrialize. In a study of 50 hearts with abnormalities, the entire right ventricle was found to be morphologically abnormal. Ebstein's anomalies are often associated with other congenital, structural, or systemic diseases, including intracardiac shunt, venous ulcers, and circulatory disorders (e.g., Wolf-Parkinson-White syndrome [WPW]) (2).

The hemodynamic effects of the Ebstein anomaly are caused by tricuspid tracts being removed and not functioning as well as atrialization of the right ventricle. Traction disruption leads to tricuspid re-entry. The difficulty of repositioning depends on the size of the tract transfer, from soft repositories with tricuspid tracts that move slowly to difficult retrieval and large displays. The atrialized portion of the right ventricle, although an atomic part of the right atrium, contracts and relaxes with the right ventricle. This discordant contraction leads to blood clotting in the right atrium. During the ventricular systole, the atrialized portion of the right ventricle attaches to the entire right ventricle, causing the blood to return to the right atrium, underscoring the effects of tricuspid regurgitation (figure 1) (3).

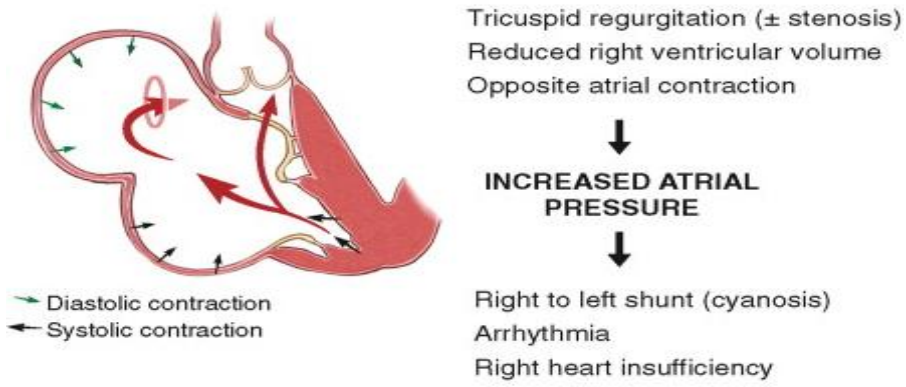

Figure 1 Pathophysiology of Ebstein Anomaly (4)

Causes

Ebstein's anomaly is a congenital disorder and is often undiagnosed. Environmental factors related to etiology include the following maternal characteristics: mother's lithium uptake in early pregnancy, mother's use of benzodiazepine, mother's exposure to vodka, mother's premature infant history, and humans. Whites are at increased risk of developing Ebstein's anomaly. Compared to other races. The tricuspid valve usually consists of three parts, called strips or flaps. While the heart is relaxing, the tracts open to allow blood to flow from the right atrium (upper chamber) to the right ventricle (lower chamber). When the heart is pumping, they close to prevent blood from flowing from the right ventricle to the right atrium. In people with Ebstein's anomaly, the tract is inserted at the depth of the right ventricle rather than in the normal position. The tube is usually larger than usual. Dysfunction often causes valve dysfunction, which can lead to abnormal blood flow. Instead of flowing into the lungs, blood flows back into the right atrium. Supporting blood flow can increase heart rate and water accumulation in the body. The valve that connects to the lungs (pulmonary valve) may narrow (5).

In many cases, people also have an opening in the wall that separates the two upper chambers of the heart (atrial septal defect), and the flow of blood through the opening can cause anoxic blood to enter the body. This can lead to cyanosis, a blue color of skin caused by a lack of oxygen. The Ebstein anomaly occurs when a baby grows in the womb. The reason is unknown. Taking certain medications (like lithium or benzodiazepines) during pregnancy can make a difference. The situation is unusual, and most common among whites (6).

Epidemiology

Ebstein anomaly accounts for about 9.60 % of all congenital heart defects. Its exact distribution is unknown because soft forms are generally not available. With the widespread use of echocardiography, many cases are detected. Race, gender, and age-related demographics The Ebstein anomaly is more common in children of white women. However, there are no specific sexual preferences. Ebstein anomaly can show up at different stages of life as follows: Child Health: Ebstein anomaly is usually diagnosed by echocardiography. Infant and child health: The Ebstein anomaly indicates cyanosis and/or severe heart failure; In

general, symptoms in childhood develop when resistance to the pulmonary arteries is low. Adult Health: Ebstein develops fatigue, aggravated dyspnea, cyanosis, tricuspid regurgitation and/or right heart failure, and palpitations; Frequently irregular heartbeat (figure 2) (7).

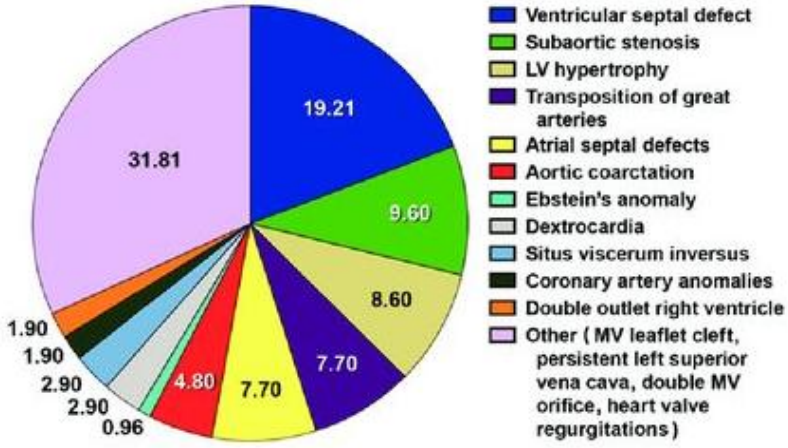

Figure 2 Prevalence of Ebstein Anomaly among all Congenital Heart Diseases (8)

Prognosis

Predictability depends on the severity of the disease and the treatment options available. Pregnancy in women with Ebstein anomaly appears to be well tolerated with adequate monitoring. Negative predictive indicators include the following: Male gender, Prior years of presentation, Cardiothoracic score above 0.65 on chest radiographs, Septal leaflet attachment ratio (i.e., the average distance between AV ring and distal attachment of the leaflet) (septal to septal tract length) more than 0.45, Increased joint of the right ventricular area and right ventricle equal to the active right ventricle (grade 1), less than 0.5, to grade 4, more than 1.5 (related risk of 2.7 related risk per grade increase), and New York Heart Association (NYHA) class: This is linked to mortality rates in some studies (table 1) (9)

Carpentier Classification system	Celermajer Scoring system (GOSE) (Right atrium + atrialized RV)/(Functional RV + left atrium + left ventricle) - 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 the RV	Grade 1.5

Table 1 Classification of Ebstein Anomaly (10)

Disease / Death: The natural course of the disease varies depending on the size of the movement of the tricuspid valve. Younger patients often develop more severe illnesses and poor diagnoses. The average age of the presentation is middle age. According to old observational data, about 5% of these patients live beyond 50 years. The oldest patient on

record was 85 years old. Ebstein's complications include heart failure, constipation, sudden cardiac death, bacterial endocarditis, brain tumors, paradoxical embolism, transient ischemic injury, and paralysis (11).

Symptoms and Signs

Patients may present with a variety of symptoms associated with the anatomical abnormalities of Ebstein's anomaly and their hemodynamic effects, or with diseases associated with the structure and surgical system. Cyanosis is very common and is usually caused by right-to-left movement of the little finger and/or severe heart failure. It's the time of a newborn's life, a repetition of an adult's life. It may first appear in adult life. Temporary onset/severity of cyanosis in adults is due to paroxysmal arrhythmias. When it appears, cyanosis gets worse. Fatigue and dyspnea are caused by heart failure secondary to right ventricular failure and decreased left ventricular output. Heart failure and sudden cardiac death can occur as a result of paroxysmal supraventricular tachycardia (SVT) or Wolff-Parkinson-White syndrome (WPW) in up to one-third of patients. Fatal ventricular arrhythmias can occur due to the presence of available pathways. Symptoms of right heart failure include ankle edema and ascites. Other common mild symptoms are Right-to-left shunt that causes brain tumor, bacterial endocarditis, paradoxical embolism, stroke, transient ischemic attack (12).

Diagnosis

Abnormal heart sounds, such as a heart murmur, don't usually cause concern. However, your doctor or your child's doctor will likely refer you to a doctor who specializes in treating heart conditions (cardiologist) to determine the cause (13).

Echocardiogram

This test uses sound waves to create detailed images of your heart. It shows the formation of the tricuspid valve and blood flow to your heart. Occasionally, a transesophageal echocardiogram is performed. This test uses a tube containing a small transducer inserted from your throat to your stomach (esophagus) to your gastrointestinal tract. Because your throat is close to your heart, this test will give you a more detailed picture of your heart (figure 3) (14).

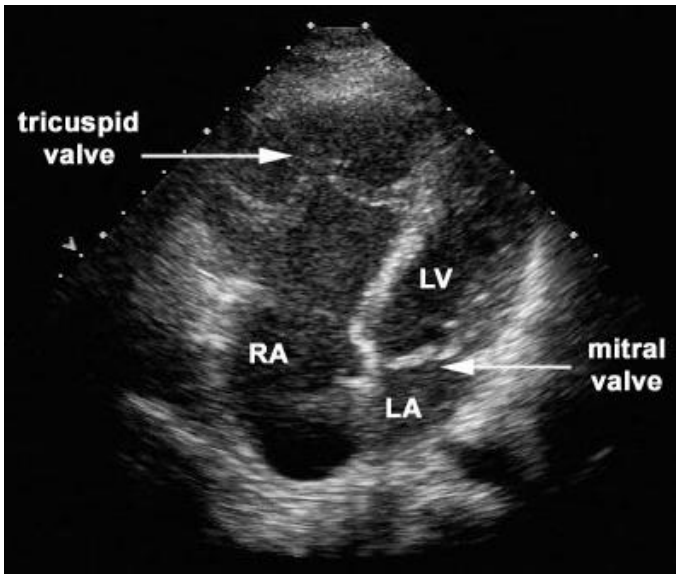

Figure 3 Echocardiogram of Ebstein Anomaly (15)

Electrocardiogram (ECG)

ECG is uncommon in many patients with Ebstein anomaly. It may show long and wide P waves as a result of right atrial enlargement, as well as complete or incomplete bulk branch block. R waves on lead V1 and V2 are small. The bizarre morphology of the QRS terminal pattern is the result of disruption of infra-Hisian conduction and abnormal activation of the atrialized right ventricle (figure 4) (16).

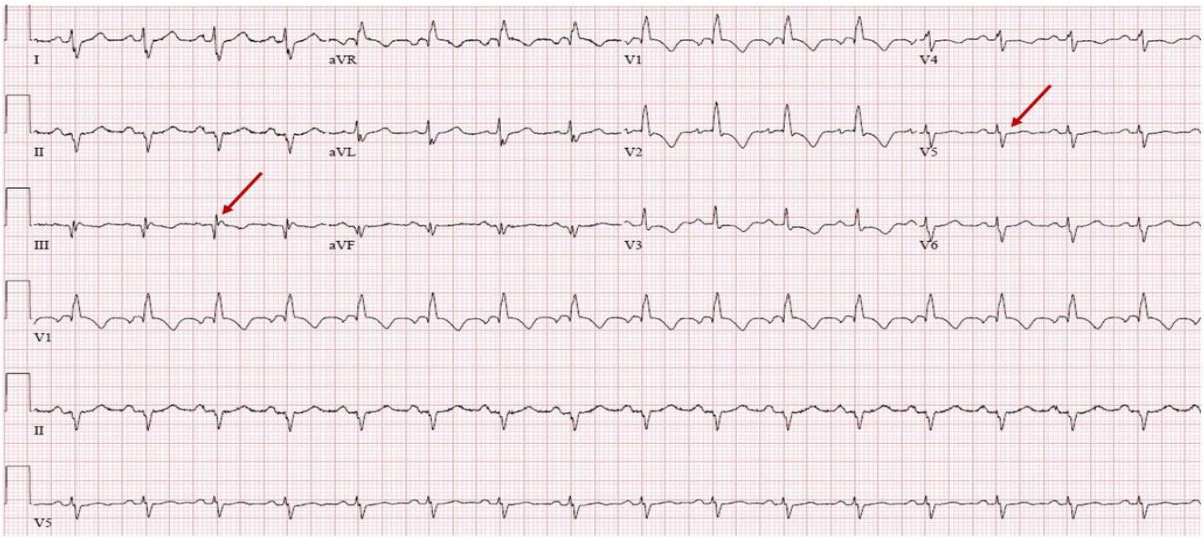

Figure 4 Electrocardiogram of Ebstein Anomaly (17)

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) (18).

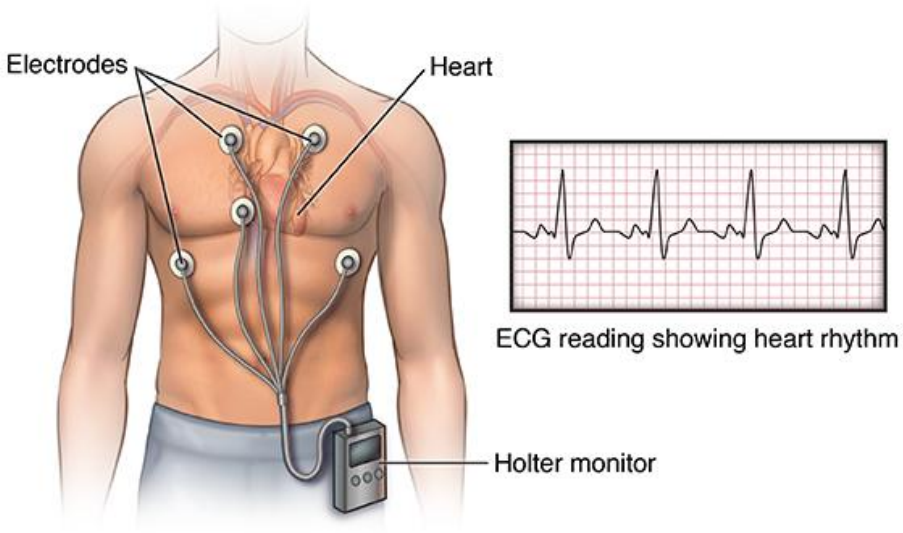

Figure 5 Holter Monitoring (19)

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) (20).

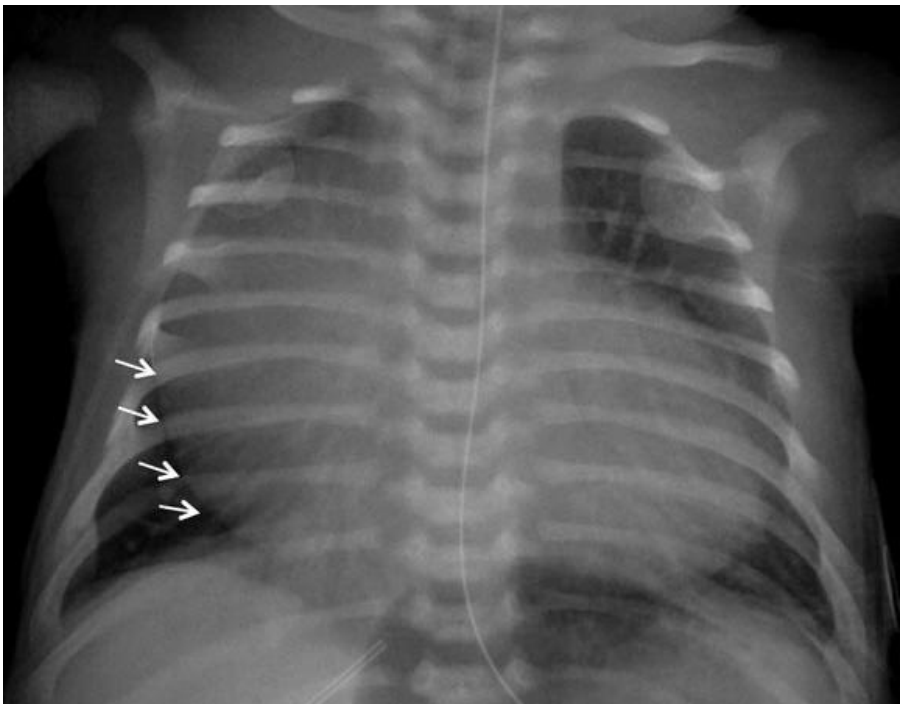

Figure 6 Chest X-ray of Ebstein Anomaly (21)

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) (22).

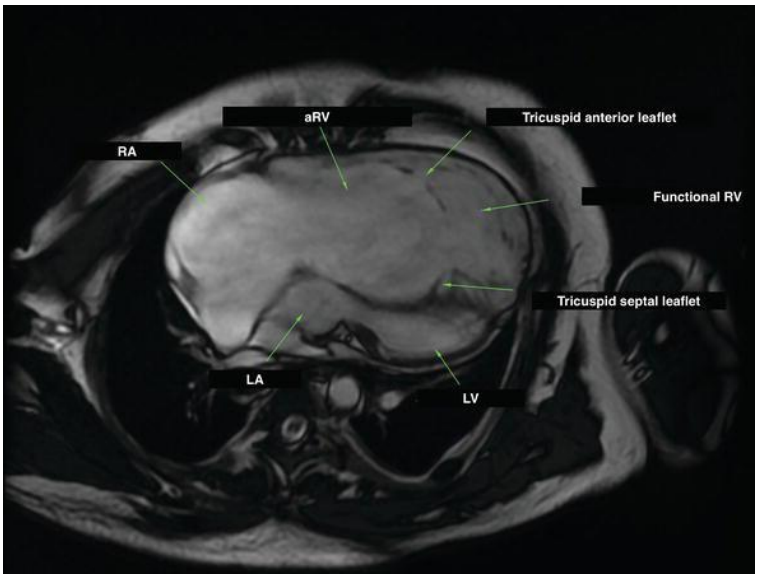

Figure 7 Cardiac MRI of Ebstein Anomaly (23)

Pulse oximetry

In this test, a sensor attached to your finger or toe measures the amount of oxygen in your blood (24).

Exercise stress test

This test monitors your blood pressure, heart rate, heart rate, and respiratory rate while you press or ride a stationary bike. Stress tests can show how your heart responds to exercise. This can help the doctor determine what level of exercise is safe (25).

Electrophysiology study (EP)

To perform this test, doctors attach a thin, flexible catheter to the inner electrodes of the heart in the bloodstream to map the heart's output. In addition, doctors can use electrodes to stimulate the heartbeat, induce or stop arrhythmias. This will help doctors determine if the drug is effective in treating arrhythmia (26).

Cardiac catheterization

In patients with Ebstein's anomaly, diagnosis of cardiac catheterization is rarely required, except for preoperative coronary angiography. Ventricular pressure and normal pulmonary artery are common in patients with the abnormality, although ventricular pressure may be elevated at the diastolic end. Right atrial pressure may be normal despite severe tricuspid duplication, especially if the right atrium is wide open. Oximetry may indicate a rupture of the arterial system before contact between the atrium and the right and left ventricles (27).

Treatment

Treatment for Ebstein's abnormalities depends on the severity of the symptom and your symptoms and signs. The goal of treatment is to reduce your symptoms and prevent future

complications, such as heart failure and arrhythmia. General Precautions: If you do not have any symptoms or signs of abnormal heartbeat, your doctor may recommend that you monitor your heart condition with regular checkups. Follow-up appointments usually include physical examinations and tests such as ECG, echocardiogram, Holter monitor test, and stress test (28).

Medications

If you have a heart rhythm disorder, medicine can help control your heart rate and keep your heart rate normal. Your doctor may also prescribe medications to reduce the symptoms and signs of heart failure, such as water pills. If you have problems with your heartbeat or the opening (atrial septal defect) between the upper chambers of your heart, your doctor may prescribe blood thinners. Some children are given an inhalant called nitric oxide to help improve blood flow to the lungs. Ebstein's anomaly requires pharmacological treatment due to the cardiovascular effects that result from tricuspid atrialization of the right ventricle, valve reconstruction, and septal defects. Patients may need antibiotic prophylaxis for bacterial endocarditis. Treatment of SVT is usually done by removal of radiofrequency ablation rather than drug therapy. CHF is treated with ACE inhibitors, diuretics, and digoxin (29).

Surgery or other procedures

Your doctor may recommend surgery if your symptoms affect your health. Surgery may also be recommended if your heart rate increases and your heart rate decreases. If you need surgery, it is important to choose a surgeon who is experienced in the field and has the training and experience to perform surgical procedures. A variety of procedures can be used for surgical treatment of Ebstein abnormalities and related defects (29).

Tricuspid valve repair

Surgeons reduce the size of the valve opening and allow the valve leaflets to come together for proper operation. Tape can be placed around the valve to hold it. This procedure is usually performed when there is sufficient tissue from the valve to heal. A new form of tricuspid valve repair is called cone reconstruction. Surgeons isolate the tricuspid valve leaflets from the myocardium. Then the leaves turn and rearrange, forming a "leaflet cone." Sometimes your valve may need to be repaired or replaced in the future (figure 8) (30).

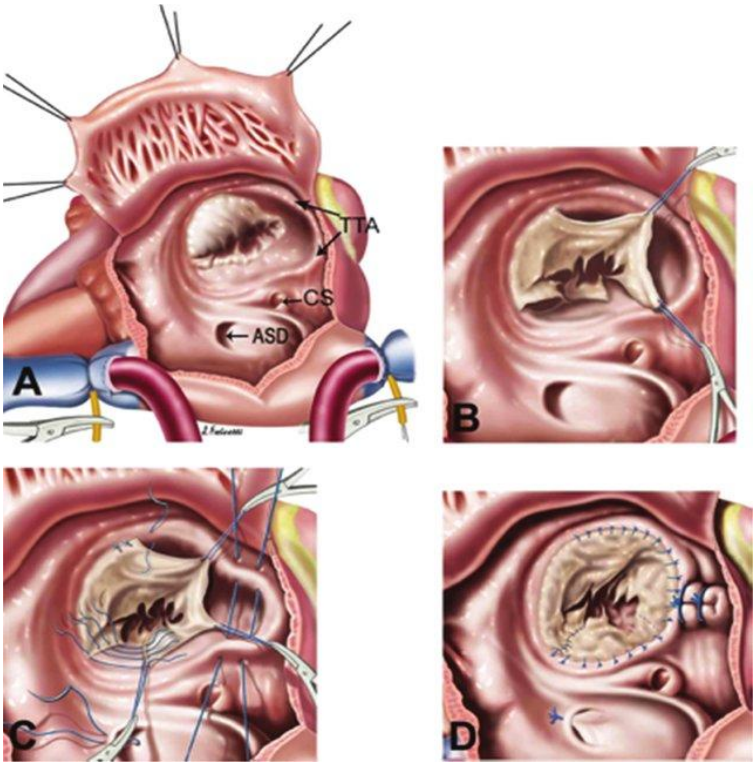

Figure 8 The Cone Reconstruction for Ebstein Anomaly (30)

Tricuspid valve replacement

If the valve cannot be repaired, your surgeon may remove it and replace it with bioprosthetic tissue or a mechanical valve. Mechanical valves are rarely used to replace tricuspid valves. If you have a mechanical valve, you will need to take blood thinners to prevent blood clots. If you have an artificial valve of any kind, you will need to take medicine to prevent inflammation of the lining of your heart (endocarditis) before dental procedures (figure 9) (30).

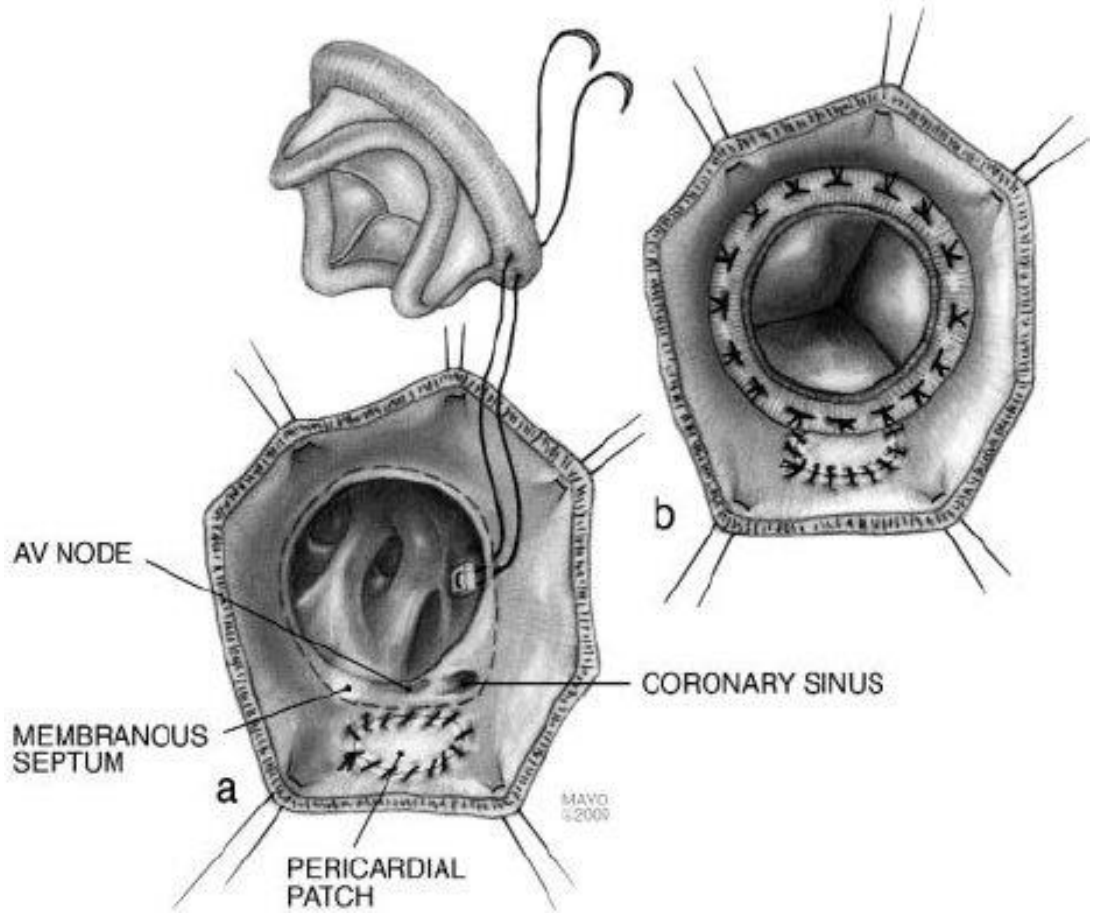

Figure 9 Tricuspid valve replacement in case of Ebstein Anomaly (30)

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 (30).

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 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 (30).

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) (31).

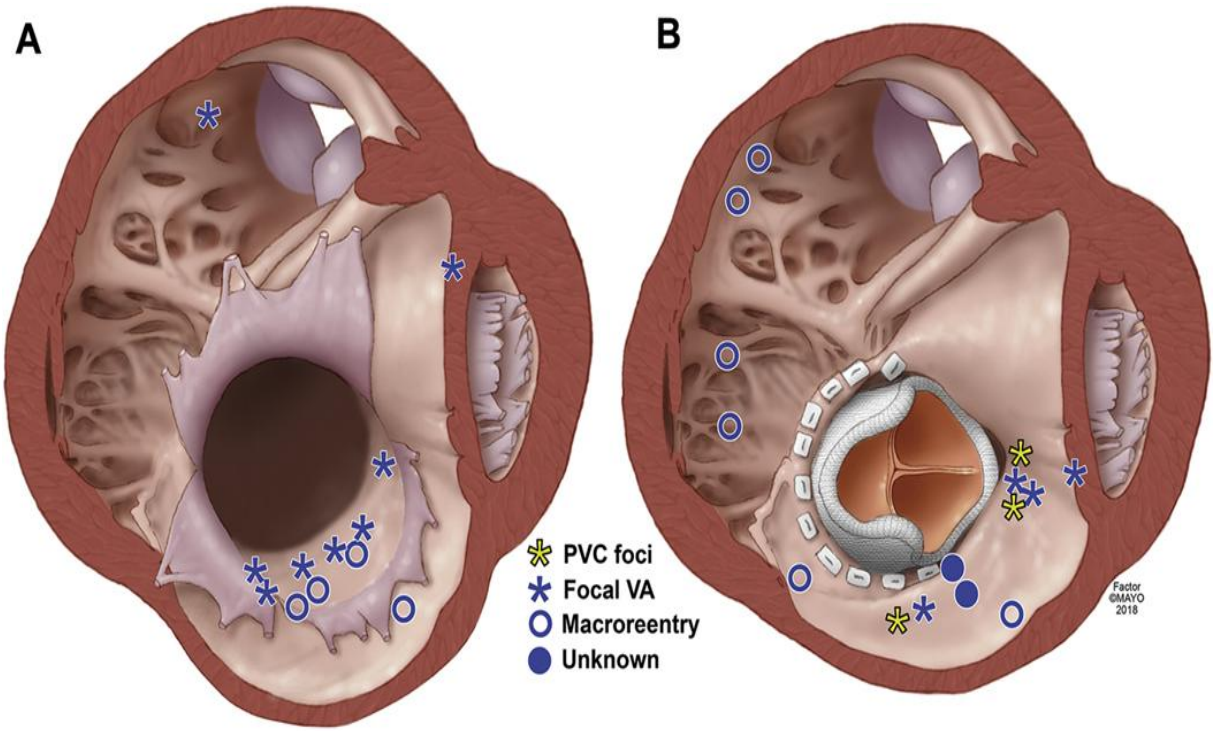

Figure 10 Sites for Catheter Ablation for Ventricular Arrythmia in Ebstein Anomaly Patients (31)

Heart transplantation

If the patient has severe Ebstein anomaly and poor heart function, a heart transplant might be necessary (32).

Guidelines

Evaluation

It is recommended that all patients with Ebstein anomaly be screened periodically in a specialized facility for adults with congenital heart disease (CHD). Class I: Electrocardiography (ECG), radiography, and echocardiography-Doppler ultrasonography are recommended diagnostic studies of Ebstein anomaly in adults. Class IIa: Relaxation and/or use of pulse oximetry may help diagnose Ebstein anomaly in older patients. (Level of evidence: C), Electrophysiologic examination (EPS) may be useful in diagnosing abnormal Ebstein patients in elderly patients with recorded or suspected supraventricular arrhythmia (consider removal of the next radiofrequency catheter, if clinically possible). (Level of evidence: C), as well as a comprehensive evaluation of Ebstein anomaly in elderly patients, Doppler transesophageal echocardiography (TEE) (if transthoracic imaging does not provide sufficient anatomic information); Holter monitoring; EPS history or ECG evidence of access methods; and/or coronary angiography (when planning surgical correction, if suspected coronary artery disease, and in men 35 years of age or older, premenstrual women 35 years or older with malignant coronary arteries, and women of postmenopausal) may be helpful. (Level of evidence: B, for all) (32).

Management

Anticoagulation with warfarin is recommended for patients with Ebstein anomaly with a history of a paradoxical embolus or atrial fibrillation. (Class I, Level of evidence: C). Recommendations for catheter intervention in adults with Ebstein anomaly are summarized below. Catheterization implants should be performed in facilities with specialized catheterization and management of elderly patients with Ebstein anomalies. (Class I, Level of evidence: C), EPS/pacing problems, Catheter ablation may provide benefits in the treatment of recurrent supraventricular, tachycardia in some patients with Ebstein anomaly. (Phase IIa, level of evidence: B), Surgical intervention (all recommendations of class I), surgeons with training and expertise in CHD should perform tricuspid valve repair or replacement with a partial closure of the atrial septal defect (ASD), if available, patients with Ebstein anomaly with the following indicators (level of evidence: B, all): Symptoms or degeneration of exercise volume, Cyanosis, and hypertension (33).

Modification of tricuspid valve surgery/replacement is recommended for adults with Ebstein's anomaly with the following indicators (level of evidence: B, all): symptoms, diminished resilience or New York Cardiac Association (NYHA) III or IV Performance category, stronger tricuspid regurgitation (TR) continuous RV dilation, contraction, RV contractile function or atrial and/or ventricular and/or ventricular arrhythmia development/progression, mixed recurrence, and stenosis, bioartificial tricuspid valve Dysfunction, after correction with a prosthetic valve greater than 12-15 mm Hg) and efficacy can be predicted early with a lesser degree of bioprosthetic stenosis, with reduced symptoms or exercise tolerance. Other management considerations: Women of childbearing potential, women with Ebstein's anomaly should seek pregnancy counseling with an adult doctor who has experience with CHD. (Class I, Level of evidence: C). Prevention of endocarditis: For postoperative patients with cyanotic abnormalities and artificial heart valves, antibiotic prevention is recommended before dental intervention with gingival tissue or the area of teeth around the apex of oral piercing. (Category IIa, Level of evidence: C)(33).

Discussion

Ebstein's anomalies are rare in fixed valves. The tricuspid valve separates the right atrium (the chamber that receives blood from the body) and the right ventricle (the chamber that pumps blood into the lungs). In Ebstein's anomaly, two tricuspid valve tips move to the bottom of the pump chamber. The third sheet is made long and can be attached to the wall of the room. In rare cases, the valve becomes paralyzed, making normal blood flow difficult (from the right atrium to the right ventricle). Typically, this destruction causes the tricuspid valve to bleed and return to the right atrium when the right ventricle is tightened (compressed). As a result, the right atrium grows. If tricuspid regurgitation is bad enough, it can lead to severe heart failure. Excessive repetition in the right atrium causes too much pressure in the right atrium. Footage usually has a closed-hole or connection or opening between the right and left atrium called PFO. PFOs usually stop after birth (34).

In Ebstein's anomaly, high right atrial pressure maintains an open PFO. This connection allows non-oxygenated ("blue") blood to flow from the right atrium to the left, bypassing the

lungs and entering the body. This will lower the oxygen level in the blood. This is why children with Ebstein's anomaly may be blue or "cyanotic" and have low oxygen saturation. Ebstein's anomaly can occur with other heart injuries, such as pulmonary valve stenosis or atresia, atrial septal defect, or ventricular septal defect. Also, many patients with Ebstein's anomaly have an extra conduction pathway in the heart, which can lead to episodes of an abnormally high heart rate called supraventricular tachycardia (a condition known as Wolff-Parkinson-White syndrome) (34).

Although patients with Ebstein anomalies may be referred for percutaneous or surgical ASD closure, the presence of Ebstein anomalies may alter the recommendation for intervention. Because of the risk of paradoxical embolism, caution should be exercised in the association between atrial fibrillation and shunt in patients with right-to-left percutaneous ablation of the accessory tract and Ebstein anomalies. The presence of multiple access methods should raise suspicion of Ebstein anomalies. Patients with continence anomalies and marked cardiomegaly may complain of few symptoms despite noticeable limitations. Exercise tests will show performance limits and should be included as part of routine testing for these patients. Exercise testing should also include monitoring of respiratory integrity, as exercise can lead to cyanosis. Newly diagnosed patients with Ebstein anomaly may be diagnosed with concomitant pulmonary arterial hypertension (PAH), particularly in the presence of cyanosis and increased right heart rate. This is often a misdiagnosis, as PAH is very uncommon in Ebstein patients (34).

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.

Conflict of interest

It is not applicable

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