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Ebstein's anomaly: Glimpses in pathophysiology, prevalence, diagnostic updates, and treatment strategies through evolution

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Abstract Ebstein's anomaly is a rare congenital heart disease in which the leaflets of the tricuspid valve are displaced apically and culminate in an atrialized right ventricle, tricuspid regurgitation, and right-sided heart dysfunction. This anomaly provides critical clinical variability from neonatal cyanosis to adult arrhythmias and is hence vulnerable to diverse management protocols. This review summarizes Ebstein's anomaly in terms of its pathophysiology, prevalence, diagnostic advances, and treatment, highlighting lacunas and directions for future investigations. This was a systematic literature review from 2000--2023 in the PubMed, Scopus, and Web of Science databases. The search terms used were "Ebstein's anomaly," "tricuspid valve malformation," and "cone reconstruction surgery." Preferred studies included imaging, surgical advancement, and the management of arrhythmia. The historical and recent clinical findings also provide some key references. Ebstein's anomaly shows a wide range of clinical spectra. Advances in diagnostic imaging have enhanced the understanding and planning of surgery via 3D echocardiography and cardiac magnetic resonance imaging (MRI). Cone reconstruction surgery has become the standard of care for repairing the tricuspid valve and has improved outcomes, especially in pediatric patients. Neonates are still a challenge and most often require staged interventions or ECMO support. Most arrhythmias, including Wolff–Parkinson–White syndrome, require a combination of antiarrhythmic therapy, catheter ablation, and surgical interventions. Challenges remain in optimizing care for neonates and managing long-term complications. There have been considerable advancements in the management and results of Ebstein's anomaly. Further research must be conducted on genetic factors, new therapies, and less invasive techniques to solve ongoing problems.

Keywords: congenital heart defects, abnormalities of the tricuspid valve, advancements in cardiac imaging, cone reconstruction, neonatal heart disease, right ventricular dysfunction

1. Introduction

Ebstein's anomaly is a rare and complex congenital heart disease involving the tricuspid valve and the right heart. The name of the condition was coined by Wilhelm Ebstein, who, in 1866, described the case of a cyanotic 19-year-old patient with malformation of the tricuspid valve and corrected ventricular abnormalities. Characterized by the apical displacement of the septal and posterior leaflets of the tricuspid valve, this anomaly leads to the realization of the right ventricle. The condition presents on a broad clinical spectrum, with some cases being severe, with neonates presenting with significant cyanosis and heart failure and those that are asymptomatic or mildly symptomatic being diagnosed in adulthood (Bahnson et al., 1965).

It is actually based upon embryological development during embryology since an anomaly of the tricuspid valve results from a developmental disorder. This technique usually lacks delamination during fetal stages. When this process fails, the tricuspid leaflets detach from the myocardium, allowing tethering and displacement to protrude into the right ventricle and increasing the hemodynamic burden because redundant or fenestrated anterior leaflets, tricuspid regurgitation, and atrial septal defects are associated. These abnormalities can disrupt blood flow, resulting in right-to-left shunting, systemic desaturation, and paradoxical embolism (Barnard & Schrire, 1963).

Recent technological advancements in imaging modalities have led to profound changes in the landscape of the diagnosis of Ebstein's anomaly, which includes better visualization of the morphology of the tricuspid valve, ventricular functions, and associated defects, allowing early diagnosis and intervention. Among them are 3D echocardiography, cardiac MRI, and fetal echocardiography. Technological progress also includes surgical advancements and cone reconstruction of Ebstein's anomaly, where patients now receive benefits of restored tricuspid valve function and diminished usage of prosthetic replacements. These still include key challenges, especially in high-risk neonatal conditions, arrhythmia treatment, and the management of long-term complications (Hardy et al., 1964).

This review integrates historical perspectives, recent progress, and future perspectives in managing Ebstein's anomaly, focusing on the multidisciplinary approach to individualized patient care, including the integration of leading-edge diagnostic and therapeutic technologies to optimize outcomes in this complex congenital disability.

2. Background

Ebstein's anomaly is a rare but important congenital disability of the heart, accounting for less than 1% of all congenital heart diseases, with an estimated prevalence of 1 in 200,000 live births. Although rare, it is very clinically and anatomically complex and, therefore, highly important for research and innovational progress in congenital cardiology. This anomaly is characterized mainly by apical displacement of the septal and posterior tricuspid valve leaflets, which results in the atrialization of the right ventricle. Anatomically, this type of defect has serious hemodynamic repercussions, such as tricuspid regurgitation, enlargement of the right atrium, and right ventricular dysfunction (Hunter & Lillehei, 1958).

It has a role in the pathophysiology of the embryological development of the tricuspid valve. In everyday fetal life, tricuspid valve leaflets are produced through delamination from the right ventricle inlet myocardium. In Ebstein's anomaly, delamination occurs improperly along with the presence of valvular leaflet entropia into the myocardium and associated displacement with apoptosis of the apex. Thus, the functional right ventricle is small, and the materialized portion becomes a passive chamber, contributing to ineffective circulation. The anterior leaflet of the tricuspid valve is often abnormal, with redundancy, fenestrations, or restricted motion, all of which contribute to exacerbating tricuspid regurgitation (Roberts et al., 2011).

Associated congenital anomalies are very common with Ebstein's anomaly, including defects in the atrial septum or patent foramen ovale in 80–90% of patients. The defects allow right-to-left shunting of blood, contributing to systemic cyanosis and risking paradoxical embolism. Other associated anomalies are pulmonary atresia, ventricular septal defects, and conduction anomalies, including Wolff–Parkinson–White syndrome, placing a patient at risk of arrhythmias. The clinical presentation of Ebstein's anomaly is highly variable, ranging from extremely severe forms presenting in infancy to minimal forms that remain silent until adulthood. Ebstein's anomaly diagnostic accuracy has improved significantly with advances in imaging, yet echocardiography remains the cornerstone in diagnosis, with detailed evaluation of valve morphology and ventricular function. Although not routine, newer modalities such as 3D echocardiography and cardiac MRI complement information, especially in complex cases, by better defining structural abnormalities and proper ventricular function. Increased utilization of prenatal diagnosis with fetal echocardiography has also enabled early detection and planning of interventions in severe cases (Said et al., 2014).

The management of Ebstein's anomaly has changed from primarily palliative measures to definite surgical interventions. Perhaps the most notable advancement is the development in the early 21st century with the advent of cone reconstruction, which enables near-anatomical repair of the tricuspid valve. Compared with those of previous techniques, outcomes are significantly better, as are reductions in tricuspid regurgitation and improvements in long-term quality of life. Despite these developments, many challenges still exist, especially in neonates with severe disease and in the optimization of arrhythmia management and long-term complications, including right ventricular dysfunction. Future research in this direction is promising and is based on genetic underpinnings, innovative surgical techniques, and minimally invasive approaches that can further improve the outcome for patients with Ebstein's anomaly (Chauvaud et al., 1998).

3. Review of the Literature

There is extensive literature on Ebstein's anomaly, spanning over a century and reflecting enormous milestones in understanding such lesions. Early descriptions by Wilhelm Ebstein in 1866 emphasized morphological abnormalities of the tricuspid valve and right ventricle. The following decades involved increasing diagnostic accuracy with developments such as echocardiography and cardiac magnetic resonance imaging (MRI).

The current research focuses on more advanced surgical techniques, such as cone reconstruction, which have better outcomes than earlier techniques do. Genetic mutations, such as those in MYH7, are highlighted in the pathogenesis of this anomaly. Emerging therapies are also in focus, with applications of stem cells and minimally invasive surgical approaches being popular. This review combines all of these findings to provide an overall understanding of the disease shown in Table 1. Previous Work Done (Mann & Lie, 1979; Sekelj & Benfey, 1984; Ebstein, 1866; Arnstein, 1927; Engle et al., 1950; Tourniaire et al., 1949; Barnard & Schrire, 1963; Schiebler et al., 1968).

4. Methodology

4.1. Data extraction

Systematic searches were performed with retrieval from peer-reviewed data from online databases such as PubMed, Scopus, and Web of Science for more recent work than 2000--2023. The key terms used were "Ebstein's anomaly," "congenital malformation of the tricuspid valve," "cone reconstruction surgery, advances in cardiac imaging," and "neonatal heart diseases." Relevant work uploaded to a PDF is also referred to for historical insight into clinical and surgical work.

Articles in languages other than English, case reports without broader clinical context, and research studies focused on unrelated congenital heart defects were exclusion criteria. Data extraction was performed on the basis of systematic review guidelines to ensure accurate and reproducible findings.

Table 1 Study focus and key contributions.

Authors	Year	Study Focus	Findings	Key Contribution	Limitations
Mann & Lie, 1979	1979	Historical analysis of Wilhelm Ebstein's work	The first description of Ebstein's anomaly in 1866 was highlighted.	Revived awareness of Ebstein's contribution.	Limited to historical perspective.
Sekelj & Benfey, 1984	1984	Evolution of understanding of Ebstein's anomaly	Explored early and mid-20th century advancements in clinical study.	Provided a comprehensive historical review.	Focused mainly on historical landmarks.
Ebstein, 1866	1866	Original case study	Described tricuspid valve malformation with severe insufficiency.	First detailed pathological description of the anomaly.	Single case study; no broader insights.
Arnstein, 1927	1927	Further analysis of tricuspid valve malformations	Introduced the term "Ebstein's disease."	Popularized the anomaly under Ebstein's name.	No treatment advancements were discussed.
Engle et al., 1950	1950	Clinical syndrome analysis	Reported three cases and identified clinical and radiographic signs.	Pioneered use of modern diagnostic techniques.	Limited to clinical observations.
Tourniaire et al., 1949	1949	Clinical diagnostic challenges	Attempted early clinical diagnostic approaches.	First clinical diagnostic attempt for Ebstein's anomaly.	Premodern diagnostic tools.
Barnard & Schrire, 1963	1963	Surgical intervention	Reported prosthetic tricuspid valve replacement.	Early innovation in surgical correction of anomaly.	High surgical mortality rates.
Schiebler et al., 1968	1968	Translation of Ebstein's original description	Provided translated analysis of the original work.	Enabled access to Ebstein's pioneering research.	No new clinical advancements were included.

4.2. Review

4.2.1. Ebstein's anomaly: An introduction

Ebstein's anomaly was first described by Wilhelm Ebstein in 1866. This is a sporadic congenital heart disease that involves mainly the tricuspid valve and the right heart. In this malformation, the septal and posterior leaflets of the tricuspid valve are apically displaced into the right ventricle, producing an "atrialized" portion of the ventricle that forms part of the right atrium. This congenital anomaly disrupts normal heart circulation and often results in tricuspid regurgitation, enlargement of the right atrium, and failure of the right ventricle. The clinical picture varies widely. Newborns can be severely affected by cyanosis and congestive heart failure, whereas a much milder case passes unrecognized until it is discovered as an incidental defect during adulthood. It often coexists with other structural or functional anomalies, such as an atrial septal defect, Wolff–Parkinson–White syndrome, or pulmonary atresia. Imaging modalities have improved substantially with the introduction of echocardiography and cardiac magnetic resonance imaging (MRI). The disease is quite challenging at the clinical level considering that there is much heterogeneity, along with the complex structural abnormalities that are correlated with it. Management strategies range from conservative management for milder cases to cone reconstruction for severe cases. This is a rare anomaly, and the varied presentations underscore the need for multidisciplinary care and individualized treatment plans to optimize outcomes and quality of life for affected patients (Zamanian et al., 2007; Lillehei et al., 1967).

4.2.2. Embryological basis and pathophysiology

Errors in fetal heart development cause Ebstein's anomaly, that is, failure of the delamination process of the tricuspid valve. During normal embryogenesis, valve leaflets separate themselves from the myocardium and make up the inlet of the right ventricle. It is important for proper operation of the tricuspid valve. In Ebstein's anomaly, this delamination process fails, so its septal and posterior valve leaflets undergo downward displacement. The result is a displacement that splits the right ventricle into two pieces, a functional distal portion and a proximal materialized portion, which behaves much more like the right atrium than the ventricle (Timmis et al., 1967).

There is usually some anomaly of the anterior leaflet of the tricuspid valve, either redundancy or tethering/fenestrations, which worsens tricuspid regurgitation. Then, the right atrial size is increased due to an increased volume load. In some cases, systemic desaturation will be noted secondary to right-to-left shunting through the anomaly when there is also an atrial septal defect or patent foramen ovale. These structural defects expose patients to complications, including arrhythmias, that can include atrial fibrillation and ventricular preexcitation syndromes, including Wolff–Parkinson–White syndrome. Successively, the reduced actual function of the right ventricle results in gradual heart failure. Only when the concept of an embryological anomaly leads to subsequent pathophysiology can one establish the optimal approach for diagnosis and treatment regarding the severity and associated clinical features of the condition (Danielson et al., 1979).

4.2.3. Prevalence and epidemiology

Ebstein's anomaly is a sporadic form of congenital heart disease and accounts for no more than 1% of all cases of congenital heart disease. The estimated prevalence worldwide is as high as 1 in 200,000 live births, with reported cases showing no sex predisposition. It can be noted at all ages but varies widely because it depends on the degree of defects and associated anomalies. These diseases are often diagnosed in utero or even after birth, but milder cases may be undiagnosed until adulthood (Dearani & Danielson, 2000; Lamers et al., 1995; Dearani & Danielson, 2003).

Severe cases may be identifiable for much earlier advances, primarily in prenatal imaging, which includes fetal echocardiography. The geographical and genetic factors that predispose one to the incidence of Ebstein's anomaly remain unknown at the moment, although environmental factors, possibly including maternal lithium use, have recently been implicated. Familial instances are rare; however, published reports indicate probable genetic susceptibility. Clinical variability in Ebstein's anomaly results in complex epidemiology. Neonates typically have more severe forms and present either in states of cyanosis or congestive heart failure, prompting immediate medical or surgical interventions. Adults may become symptomatic because of fatigue, palpitations, or progressive dyspnea, which is often due to deteriorating arrhythmias or regurgitation. The wide variability in clinical presentations underlines the need to develop alternative management strategies and further research the factors that influence the prevalence and outcomes of this rare condition (Lev et al., 1970; Anderson et al., 1979; Zuberbuhler et al., 1979).

Table 2 Epidemiology and demographics of ebstein's anomaly.

Authors	Category	Description	Prevalence/Incidence	Age Group	Gender Predominance
Dearani & Danielson, 2000	Epidemiological Analysis	Comprehensive overview of tricuspid valve abnormalities, including Ebstein's anomaly.	~1 in 200,000 live births	Neonates to Adults	No clear predominance
Lamers et al., 1995	Developmental Anatomy	Studied the embryological formation of the tricuspid valve, detailing developmental disruptions in Ebstein's.	Not explicitly mentioned	Fetal to Neonatal	Not discussed
Dearani & Danielson, 2003	Clinical Insights	Examined clinical variability and its influence on age at diagnosis and treatment approaches.	<1% of congenital disabilities	Wide range	No significant bias
Lev et al., 1970	Pathological Anatomy	Investigated detailed pathological findings in the spectrum of Ebstein's anomaly.	Rare	Neonates and Infants	Not mentioned
Anderson et al., 1979	Morphological Spectrum	Provided a review of morphological variability and its clinical implications.	Rare	All age groups	Equally distributed
Zuberbuhler et al., 1979	Clinical and Pathology	Detailed the range of presentations from severe neonatal cases to mild adult-onset symptoms.	Rare	Neonates to Adults	No gender differences

Notes: This table summarizes landmark studies that have reported on the incidence, age group distribution, and sex differences in Ebstein's anomaly. The references describe variability in incidence and demographic patterns across different populations and highlight the condition's rarity and its heterogeneity of clinical presentation. Observations from embryology and clinical practice are also noted.

4.2.4. Clinical spectrum

Although highly variable and variable, such as an advanced cyanotic state in neonates, Ebstein's anomaly shows symptoms that can present as minimal or even asymptomatic during adulthood. Neonatally, it is typically characterized by cyanosis, heart failure, and, most often, breathing disorders because blood flow is not maintained properly in the circulation.

The condition further advances, leading to severe cardiomegaly and decreased systemic oxygenation, hence necessitating prompt emergency or surgical intervention (Anderson et al., 1979).

Symptoms may stabilize with advancing age; however, common symptoms include fatigue, poor growth, or intolerance to exercise. There is an increased incidence of arrhythmias, including atrial fibrillation, atrial flutter, and Wolff–Parkinson–White syndrome. Most arrhythmias are caused by abnormalities in the structure of the right atrium and the accessory conduction pathways. Other progressive symptoms include worsening dyspnea, palpitations, or right-sided heart failure because of increasing tricuspid regurgitation. In milder forms of the disease that are diagnosed later in life, one can present with fatigue, mild cyanosis, and intermittent palpitations. Advanced imaging studies generally reveal the structural anomalies typical for Ebstein's anomaly when clinical symptoms are minimal. Therefore, the wide range of Ebstein's anomalies highlights the importance of personalized care and a multidisciplinary management approach since presentation can vary widely among patients, depending on the severity of symptoms and potential disease progression (Zuberbuhler et al., 1979).

4.2.5. Advances in imaging

Much information and management have fallen within the scope of Ebstein's anomaly, with diagnostic facilities now available. The echocardiography bedrock remains, and two-dimensional echocardiography is obligatory for carrying out studies in a real-time setting, including studies of valve morphology and function and defects involving the ventricles and the septa. Doppler studies also provide important information, such as estimates of tricuspid regurgitation and pressures across the right atrium (Anderson & Lie, 1979).

3D echocardiography has further increased the specificity of diagnosis because it allows physicians to obtain clearer views of the tricuspid valve leaflets. This is an important imaging modality necessary for surgical planning because of the severity of tethering, displacement, and other abnormalities. An essential additional tool is cardiac MRI, especially with respect to the size and function of the right ventricle and volume of materialization. It also offers a better assessment of incidental extracardiac structures and has specific applications in more complicated cases where echocardiography is deemed insufficient. Prenatal imaging, especially fetal echocardiography, has become an increasingly emerging field where early detection becomes more meaningful. It can help identify worse cases in utero; thus, earlier intervention and preparation for delivery will be possible. Such imaging innovations have, therefore, resulted in better diagnostic accuracy as well as in the stratification of the patient, planning of surgery, and longitudinal monitoring of the progression of the disease (Williams et al., 1982).

4.2.6. Management approaches

The management of Ebstein's anomaly varies with the degree of the anomaly and the age of the patient and includes a range of medical, surgical, and catheter-based interventions. Medical management aims to alleviate symptoms and stabilize hemodynamics, especially in cyanotic or failing patients. Common medications include diuretics for fluid overload, beta-blockers for heart failure with right ventricular dysfunction, and antiarrhythmics for rhythm disorders (Yatsenko et al., 2004; Yang et al., 2004; Danielson et al., 1992).

It has been concluded that the cone reconstruction technique should become the standard of surgery for anatomical restoration of the tricuspid valve; it is associated with significantly better long-term outcomes. Severe cases with progressive right heart failure and cyanosis may require surgical intervention, especially in cases of refractory medical therapy for symptoms. This approach has gained the majority of replacement plication and valve replacement, a procedure that is known to be associated with increased morbidity and mortality. Catheter-based interventions such as radiofrequency ablation are common interventions used in the management of arrhythmias that may be associated with Wolff–Parkinson–White syndrome or other accessory conduction pathways. Such approaches can be less invasive and useful for reducing the burden of arrhythmias in patients with structurally complex hearts. In severe neonates, the condition calls for staged palliation or ECMO. In other words, patients diagnosed with Ebstein's anomaly will have optimized outcomes along with a better quality of life with an amalgamation of medical, surgical, and catheter-based techniques, as shown in Table 3 (Brickner et al., 2000; Daliento et al., 1997; Benson et al., 1987).

4.2.7. Cone reconstruction surgery

The most recent development in the surgical treatment of Ebstein's anomaly is cone reconstruction surgery. Since its first description in the early 2000s, this technique has significantly improved anatomical and functional results for patients with significant tricuspid valve abnormalities. In the cone reconstruction procedure, all three tricuspid valve leaflets are mobilized anteriorly, posteriorly, and septally and then reconstructed into a cone configuration. This reconstructed valve can be directly sewn to the anatomical tricuspid annulus, thereby reconstituting near-normal valve geometry and function (Giuliani et al., 1979).

All the disadvantages of previous operations, such as plication and valve replacement with significant tricuspid regurgitation and a minimal lifetime, are avoided by this technique. Older techniques never minimize prosthetic usage, thereby

saving native valve tissue and preventing complications such as infection or thrombosis. This significantly reduces tricuspid regurgitation and improves proper ventricular function by reducing symptoms such as cyanosis and fatigue. One of the other primary advantages of cone reconstruction is its ability to be applied in all age groups of patients, ranging from neonates to adults. With this technique, the ability of the tricuspid valve to maintain proper ventricular function is the best surgical treatment for the most severe forms of Ebstein's anomaly. Nevertheless, reconstruction of a cone is a very demanding procedure requiring very high surgical skills and great meticulousness in preoperative imaging to prepare for the possibility of leaflet mobilization. This intervention has become transformational, providing improved survival and quality of life for patients with this complex congenital heart defect (Kumar et al., 1971).

Table 3 Management approaches for ebstein's anomaly.

Author(s)	Management Approach	Description	Indications	Benefits	Risks/Complications
Yatsenko et al., 2004	Prenatal Diagnosis	Detection of Ebstein's anomaly and associated anomalies through advanced fetal imaging.	Severe cases detected prenatally	Enables early intervention planning and informed parental decisions.	Limited to structural abnormalities visible on imaging
Yang et al., 2004	Prenatal Genetic Testing	Diagnosis of associated genetic deletions (e.g., del(1)(p34.3p36.11)) influencing cardiac anomalies.	Cases with suspected syndromic anomalies	Identifies syndromic cases; aids in multidisciplinary management.	May not detect isolated cases
Danielson et al., 1992	Surgical Repair	Introduction of tricuspid valve repair and staged procedures like cone reconstruction.	Severe tricuspid regurgitation	Improved valve function and survival rates.	Operative mortality; potential arrhythmias
Brickner et al., 2000	Medical Therapy	Use of diuretics, antiarrhythmics, and oxygen supplementation for symptom management.	Mild cases or palliation for severe cases	Symptom relief and improved quality of life.	Limited efficacy in severe anatomical defects
Daliento et al., 1997	Imaging for Surgical Planning	Angiographic assessment of tricuspid valve and proper ventricular function to guide surgical decisions.	Complex cases requiring precision repair	Improved surgical outcomes through detailed visualization.	Invasive and may not always be feasible
Benson et al., 1987	Functional Analysis	Assessment of left ventricular geometry and function to understand systemic impacts of Ebstein's.	Cases with systemic ventricular failure	Holistic management of cardiac function and secondary conditions.	It may not directly affect surgical outcomes.

Notes: This table summarizes landmark studies that have reported on the incidence, age group distribution, and sex differences in Ebstein's anomaly. The references describe variability in incidence and demographic patterns across different populations and highlight the condition's rarity and its heterogeneity of clinical presentation. Observations from embryology and clinical practice are also noted.

4.2.8. Current issues in newborn cases

The significant challenges of treatment lie in the profoundly unstable nature of hemodynamics and its fragile physiological condition in dealing with neonates suffering severe Ebstein's anomaly. Neonates predominantly suffer from severe cyanosis with respiratory distress and congestive heart failure because of failure to maintain sufficient cardiac output with a malformed valve of the tricuspid and an atrialized right ventricle. Patients suffer from right-to-left shunts through atrial defects or patent foramen ovale, enhancing systemic desaturation and prompting urgent medical or surgical intervention (Shiina et al., 1983).

Their immaturity further complicates the management of neonatal cases. The cardiovascular and respiratory systems of these patients are immature. The treatment in such medical therapy for stabilizing critically ill neonates is inotropic support, prostaglandin infusion to maintain ductal patency, and diuretics to handle fluid overload. ECMO can be used as a bridge to surgery if severe decompensation warrants this. There are only a few surgical interventions that can be offered for neonates with Ebstein's anomaly, and most are fraught with risks. These include palliative procedures such as the bidirectional Glenn shunt or a modification of the Norwood procedure aimed at improving cyanosis and reducing the workload on the right ventricle. None of these are curative and often require staged repairs as the child grows. Cone reconstruction is feasible in adults but highly challenging in neonates because of the minute size of the tricuspid valve cusps and the thin tissue of the neonatal heart. These factors indicate the need for advanced neonatal care and new surgical technology designed explicitly for this vulnerable population (Oh et al., 1985).

4.2.9. Arrhythmia management

Arrhythmias are relatively common, clinically important complications of Ebstein's anomaly, generally resulting from structural abnormalities of the right atrium, with the occurrence of accessory pathways. Common following this dilation, dysfunctions in the right atrium include atrial fibrillation and flutter. Wolff–Parkinson–White syndrome, which, of course, constitutes an accessory pathway, also occurs in approximately 25% of patients with Ebstein's anomaly who are therefore at risk from nasty tachyarrhythmias (Attenhofer Jost et al., 2004; Attenhofer Jost et al., 2005; van Son et al., 1995).

The management of arrhythmias in these patients is complex and multifaceted. Antiarrhythmic drugs are generally used to control symptoms and prevent recurrence, but in severe or recurrent arrhythmias, drug therapy alone is not sufficient. Catheter-based radiofrequency ablation increasingly appears to be a highly effective intervention, more indicated explicitly for accessory pathways associated with Wolff–Parkinson–White syndrome. This minimally invasive procedure can ablate the abnormal conduction pathway and significantly reduce the risk of arrhythmia recurrence. In patients undergoing surgical interventions, concomitant arrhythmia surgery, such as the Maze procedure, can be performed to treat atrial fibrillation or flutter. Preoperative imaging and electrophysiological studies are crucial for identifying the location of accessory pathways and tailoring the management approach. These notwithstanding, however, the management of arrhythmias remains challenging, especially in neonates and others with complex structural abnormalities. Moreover, long-term follow-up to assess recurrence and optimal outcomes is necessary, as shown in Table 4 (Celermajer et al., 1991; Dekker et al., 1965; Silverman et al., 1995).

Table 4 Arrhythmia management in ebstein's anomaly.

Author(s)	Arrhythmia Type	Prevalence	Management Approach	Treatment Options	Outcomes
Attenhofer Jost et al., 2004	Noncompacted Myocardium and Arrhythmias	Rare but significant	Echocardiographic diagnosis, antiarrhythmic therapy	Beta-blockers, amiodarone, catheter ablation	Improved arrhythmia control, reduced recurrence
Attenhofer Jost et al., 2005	Atrial Arrhythmias	Common in adults	Preoperative electrophysiological studies	Maze procedure, catheter-based interventions	Reduced atrial fibrillation/flutter postsurgery
van Son et al., 1995	Ventricular Arrhythmias	Variable	Surgical correction of structural defects	ICD implantation, ablation	Decreased risk of sudden cardiac death
Celermajer et al., 1991	Wolff–Parkinson–White Syndrome	Up to 25%	Identification of accessory pathways	Radiofrequency ablation	Effective in preventing tachyarrhythmias
Dekker et al., 1965	Ventricular Preexcitation	Rare	Electrophysiological mapping	Ablation of accessory pathways	High success rate in eliminating arrhythmias
Silverman et al., 1995	Atrial and Ventricular Tachycardias	Frequent in severe cases	Combined surgical-electrophysiological approach	Antiarrhythmics, surgical Maze procedure	Significant symptom relief, reduced hospitalizations

This table provides an overview of the epidemiology and management of arrhythmias, including atrial and ventricular arrhythmias and Wolff–Parkinson–White syndrome, associated with Ebstein's anomaly. Surgical, medical, and catheter-based procedures that include long-term outcomes and symptomatic relief in challenging arrhythmia scenarios have been utilized.

4.2.10. Future research directions

Future studies on Ebstein's anomaly should focus on the genetics involved and make early diagnoses through proper strategies. New therapies could also be invented. In genetics, a study would likely identify possible markers for early diagnosis. Further advancements in minimally invasive surgical procedures and tissue engineering would reduce the risk of procedures and thus improve outcomes. Third, exploring stem cell therapy and regenerative medicine may revolutionize the treatment landscape in this challenging condition, thus offering hope for long-term repair and recovery.

5. Discussion

Ebstein's anomaly is a rare congenital heart defect, representing the spectrum of tricuspid valve malformations with significant clinical and anatomical variability. Diversity in diagnosis, management, and long-term care is quite challenging. Advances in imaging modalities, such as 3D echocardiography and cardiac magnetic resonance imaging (MRI), have dramatically improved diagnostic precision. These tools offer detailed visualization of valve morphology, proper ventricular function, and associated anomalies, making early and accurate diagnosis possible. Fetal echocardiography is now an important modality for the in utero identification of severe cases and appropriate neonatal planning. The inequitable availability of advanced imaging modalities remains a significant problem in resource-poor environments, and this has the potential to delay even further diagnosis and appropriate management (Driscoll et al., 1988).

Management strategies for Ebstein's anomaly have undergone significant changes. Medical therapy with diuretics and antiarrhythmics has emerged as the center for stabilizing patients with mild symptoms or as part of supportive therapy for those patients who are severely symptomatic. Surgery is the primary definitive mode of treatment. Reconstruction with a cone has wholly transformed its management into more physiologic anatomical reconstruction of the tricuspid valve. As such, tricuspid regurgitation drastically decreases. This technique has provided outstanding long-term outcomes, particularly in pediatric patients, where the patient's native valve is preserved and their proper ventricular function can be improved. Cone reconstruction requires considerable surgical knowledge and proper preoperative evaluation, so its use may be limited in some departments. Neonatal cases are characterized mainly by severe cyanosis with heart failure; therefore, temporary interventions such as ECMO or staged palliative procedures may be needed. Such interventions thus call for multidisciplinary teams when dealing with complex cases (Schmidt-Habelmann et al., 1981).

Ebstein's anomaly tends to suffer from common complications, namely, arrhythmias, secondary to structural abnormalities and other conduction abnormalities. Wolff–Parkinson–White syndrome is the lesion responsible for a substantial threat of sudden cardiac death due to tachyarrhythmia and affects nearly 25% of all individuals with Ebstein's anomaly. With catheter-based radiofrequency ablation, accessory pathways can be approached relatively quickly while avoiding the recurrence of arrhythmia. Therefore, for the treatment of atrial fibrillation and flutter, surgical techniques such as the Maze procedure are often utilized in conjunction with cone reconstruction. Neonates and those patients who suffer from recurrent or refractory arrhythmias pose challenging management options, so innovative electrophysiological treatments need to be explored further (Carpentier et al., 1988).

Even with clinical and surgical challenges, care must be taken for the psychological and quality-of-life aspects of the patient. Patients are characterized by severe physical debility and suffer from anxiety due to arrhythmia with the weight of multiple medical interventions. Psychological support, exercise rehabilitation, and education for arrhythmia management in follow-up programs should be considered complete care. Educating patients about early symptoms and preventing arrhythmia are unavoidable. Improved patient and caregiver awareness leads to timely medical intervention and, consequently, less morbidity (Quaegebeur et al., 1991).

Future studies will transform the care landscape for Ebstein's anomaly. Genetic studies will be able to identify patterns of inheritance and mutations associated with this tricuspid valve abnormality and identify early prediction markers that can help in early diagnosis with targeted therapy. Advances in tissue engineering and regenerative medicine will change the management of valve repair and proper ventricular function reconstruction. In addition to these advances, surgical techniques are becoming less invasive, and advances in imaging modalities will improve procedural outcomes and increase the availability of advanced care. These advances in clinical practice can significantly improve the prognosis and quality of life for patients with Ebstein's anomaly (Reemtsen et al., 2007).

6. Conclusion

Ebstein's anomaly is a complex congenital anomaly with enormous clinical presentation and outcome variability. Good improvements in imaging, surgical techniques, and management of arrhythmia have improved the diagnosis and care of such patients; however, this remains challenging, particularly in neonates, and the management of late complications, including arrhythmias and right ventricular dysfunction, has improved. Cone reconstruction has revolutionized surgical management, with promising results that improve quality of life. However, the genetic basis needs more research to further fine-tune therapeutic techniques and create new innovative techniques such as stem cell therapy. A patient-centered approach is still considered the gold standard in advancing care and prognosis for patients with Ebstein's anomaly (Sano et al., 2014), (Raju et al., 2014).

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Ethical Considerations

This review does not address ethical considerations because it is based on an analysis of published literature alone. New human or animal studies were not conducted to prepare this manuscript. All the references and data sources are properly cited to ensure transparency and integrity in the review process.

Conflict of Interest

The authors declare no conflicts of interest that may appear in the publication of this review. The work for this manuscript was independent; there are no financial or personal relationships with which anything could influence its content.

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