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Review Article

Dealing with Ebstein's anomaly

Lianne M. Geerdink,[1] Livia Kapusta[2,3]

[1]*Department of Pediatrics, Radboud University Nijmegen Medical Centre, Nijmegen, The Netherlands;* [2]*Pediatric Cardiology Unit, E. Wolfson Medical Centre, Holon, Israel;* [3]*Children's Heart Centre, Radboud University Nijmegen Medical Centre, Nijmegen, The Netherlands*

Abstract Ebstein's anomaly is a complex congenital disorder of the tricuspid valve. Presentation in neonatal life and (early) childhood is common. Disease severity and clinical features vary widely and require a patient-tailored treatment. In this review, we describe the natural history of children and adolescents with Ebstein's anomaly, including symptoms and signs presenting at diagnosis. Current classification strategies of Ebstein's anomaly are discussed. We report on diagnostic methods for establishing the severity of disease that might enhance decision on the timing of surgical intervention. Furthermore, we describe different surgical options for severely ill neonates and multiple surgical interventions after infancy. Only with ample knowledge and understanding of the above, this complex and diverse group of patients can be correctly treated in order to improve not only duration, but also quality of life.

Keywords: Ebstein's anomaly; classification; diagnostics; management; surgery; outcome

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EBSTEIN'S ANOMALY IS A RARE CONGENITAL HEART disorder that occurs in about one to five per 200,000 live births, accounting for $<1\%$ of all congenital heart diseases.[1] The essence of the originally described Ebstein's anomaly is a displacement of the effective tricuspid valve orifice downward into the right ventricle, dividing the right ventricle into an atrialised and a functional portion. The severity of this malformation varies from mild, with few or no clinical findings, to severe defects incompatible with life. Presentation within the first 12 months of life is highly associated with poor prognosis, whereas in older children the long-term prognosis is superior. In this review, we summarise the current management related to the anatomical and electrophysiological comorbidities of Ebstein's anomaly, as well as surgical interventions and predictors of outcome in the first two decades of life.

Correspondence to: L. M. Geerdink, Department of Pediatrics, Radboud University Nijmegen Medical Centre, PO Box 9101, 6500 HB Nijmegen, The Netherlands. Tel: 0031-6-47470386; Fax: 0031-24-3619348; E-mail: lgeerdink@gmail.com

Pathological features

In the normal heart, the tricuspid valve consists of the anterior, posterior, and septal leaflet. In Ebstein's anomaly, an abnormal development of the valvular components is seen. The leaflet tissue of the valves extends from the ventricular walls into a process called "delamination". Owing to incomplete delamination, the hinge points of the septal and posterior leaflet are displaced. They are attached within the cavity of the ventricle itself, creating a functional tricuspid annulus below the true anatomic annulus. Furthermore, a rotation between 10° and 90° from the level of the atrioventricular junction often accompanies this apical displacement of the functional annulus[2] (Fig 1). The leaflets have irregular shapes, are dysplastic and are tethered by short chordae and papillary muscles or have direct myocardial insertions. In a subset of patients, complete fusion makes it impossible to distinguish one leaflet from another. The malformed tricuspid valve may be incompetent, stenotic, or, rarely, imperforated.

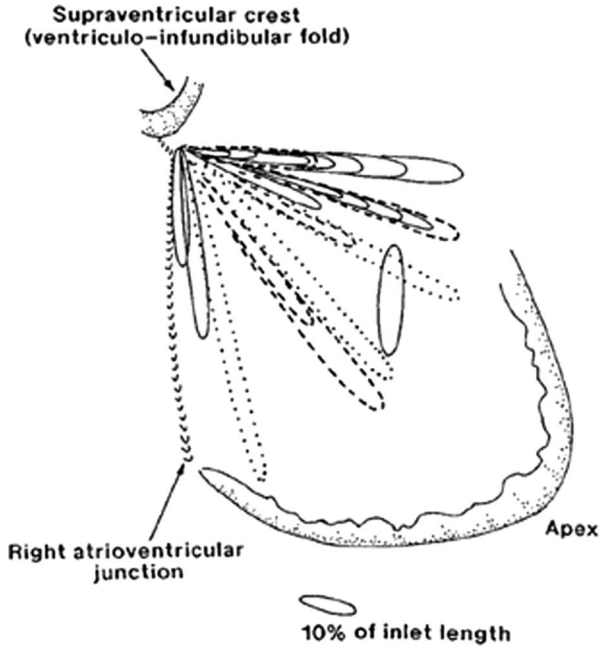

Figure 1.
Representation of the right ventricle. The ellipses demonstrate effective valvular orifices as estimated in 23 specimens by Schreiber et al. The solid lines represent cases with linear attachment; the ellipses with dotted lines and broken lines represent hearts with focal and hyphenated attachment of the anterosuperior leaflet, respectively. The vertical ellipse represents the solitary case with the origin parallel to the atrioventricular junction. The scale represents the maximal width of the orifice as a percentage of the distance between the atrioventricular junction and the apex of the right ventricle, measured along the acute margin. Source: Schreiber et al.[2]

The intraventricular displaced functional tricuspid annulus divides the right ventricle into two parts: the "atrialised" and the functional right ventricle. The atrialised right ventricle, which can be seen as a continuation of the true right atrium, shows variable degrees of thinning of the wall; it is grossly fibrotic and contains only sparse muscular fibres. The functional right ventricle may be massively dilated, including the infundibulum. The wall may be thinned and fibrotic.

Classification

Classifying Ebstein's anomaly is difficult because of its wide spectrum of abnormalities. The first attempt was made by Carpentier et al.,[3] proposing four grades of Ebstein's anomaly: type A, the volume of the true right ventricle is adequate; type B, there is a large atrialised component of the right ventricle but the anterior leaflet moves freely; type C, the anterior leaflet is severely restricted in its movement and may cause significant obstruction of the right ventricular outflow tract; and type D, almost complete atrialisation of the right ventricle with the exception of a small infundibular component. This classification, however, has its restrictions because the various components of the anomaly do not necessarily correlate with the functional severity of the disease, and a combination of types can be seen.

Celermajer et al.[4] described an echocardiographic grading score for neonates with Ebstein's anomaly; the Glasgow Outcome Score Extended Score grade 1–4. Severity of Ebstein's anomaly was graded by calculating the ratio of the combined area of the right atrium and the atrialised right ventricle to that of the combined area of the functional right ventricle, left atrium, and left ventricle in a four-chamber view at end diastole. The ratio was used to define four grades of increasing severity: grade 1, ratio <0.5; grade 2, 0.5–0.99; grade 3, 1–1.49, and grade 4 ≥ 1.5.

Dearani et al.[1] proposed a new classification according to two different approaches. The first is based on echocardiographic appearance. Ebstein's anomaly is categorised into mild, moderate, or severe, similar to the classification used for echocardiographic quantification of mitral regurgitation. However, they describe this classification as imprecise and subjective. Their second approach considers the exact anatomy of all three leaflets of the tricuspid valve, the right ventricle, and the right atrium, resulting in the classification of Ebstein's anomaly type I–IV. The latter limits classification of Ebstein's anomaly to direct surgical observation.

Associated cardiac malformations

An interatrial communication is commonly present, either as a patent foramen ovale or as a (secondary) atrial septal defect. This communication is usually associated with a right-to-left shunt, although left-to-right shunt may be present in some young patients with mild forms of Ebstein's anomaly. A ventricular septal defect is often present, usually leading to right-to-left shunting in the presence of neonatal anatomic or functional severe pulmonic stenosis and pulmonary atresia. Furthermore, Ebstein's anomaly is often associated with distal right ventricle anomalies including pulmonary valve stenosis or atresia. Miscellaneous cardiac defects are reported as well.

Attenhofer et al.[5] focused on the left side of the heart in Ebstein's anomaly patients, finding abnormalities involving the myocardium or valves in 39% of the patients, for example mitral valve prolapse or dysplasia, bicuspid aortic valve, and non-compaction. Therefore, Ebstein's anomaly should not be regarded as a disease of the right side of the heart alone.

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Clinical presentation

Physical examination

Physical signs on examination vary markedly, but they are present in the majority of the patients. Growth, development, and body build of the patients are generally normal, although Kapusta et al.[6] found children with Ebstein's anomaly to be slightly shorter than the population average in addition to the mean body mass index standard deviation score being slightly decreased. Cyanosis is seen in all degrees along with various degrees of digital clubbing. A prominent third or even a fourth heart sound is often heard. In the study by Celermajer et al.,[4] out of 88 neonates, 65 (74%) presented with cyanosis, nine presented with heart failure, eight neonates had an incidental murmur, five presented with arrhythmia, and one neonate was diagnosed after cardiac screening, performed because of the clinical suspicion of having trisomy 18. In the study of Arizmendi et al.,[7] 18 out of 21 (86%) neonates presented with cyanosis, six with heart failure, and six with a murmur. No neonates were seen with arrhythmia. Older children are often referred for further examination after a heart murmur is heard.[4,7] Celermajer et al. described a murmur as a presenting symptom in 33 out of 50 (66%) children aged up to 10 years; seven of the 50 patients had cyanosis, six had arrhythmias, and four had heart failure. In adolescence, arrhythmias, but also heart failure, murmurs, and cyanosis are reported as presenting symptoms.[4,7]

Electrophysiology

Atrial and ventricular arrhythmias are common in patients with Ebstein's anomaly. Owing to the fact that the anatomic tricuspid annulus represents an incomplete fibrous ring, it may allow direct muscular connections between the right atrium and right ventricle, providing the anatomic substrate for ventricular pre-excitation. Pre-excitation and Wolff–Parkinson–White syndrome are strongly associated with Ebstein's anomaly.

Delhaas et al.[8] studied 93 children with Ebstein's anomaly for pre-excitation or arrhythmic events. Rhythm disturbances were exhibited in 17%. The median age at diagnosis of this subgroup (n = 16) was 0 days and the median age at first electrocardiogram abnormality was 3 months. There were five patients who presented with supraventricular tachycardia, three showed pre-excitation on routine electrocardiogram at initial diagnosis of Ebstein's anomaly, and seven presented later in life with rhythm disturbances. In all, two patients died, one because of intractable supraventricular tachycardia and one probably due to an arrhythmic event. All 14 surviving patients showed pre-excitations, in four intermittently. In addition, atrial flutter, atrial fibrillation, and also ventricular tachyarrhythmia may occur in young patients with Ebstein's anomaly.

Laboratory assessment

Echocardiography

Two-dimensional echocardiography is the most important diagnostic tool for children with congenital heart disease. It is portable, non-invasive, has no radiation risk, and provides immediate high-resolution anatomical and physiological information.

Prenatal echocardiography

Foetal echocardiography is an accurate predictor of congenital heart disease from the mid-trimester of pregnancy.[9] Abnormalities of the tricuspid valve can be accurately defined prenatally, although there is much overlap between valvular dysplasia and Ebstein's malformation. Sharland et al.[9] diagnosed by echocardiography 38 foetuses with abnormalities of the tricuspid valve. In all, 14 were diagnosed with Ebstein's anomaly and diagnosis was confirmed in 10 cases; three proved to have valve dysplasia only and one was lost to follow-up. Nevertheless, foetuses with severe Ebstein's anomaly might suffer from early intrauterine demise before the first ultrasound examination is made, resulting in underestimation of the disease diagnosed prenatally.

Echocardiography in infants and children

In patients with Ebstein's anomaly, the anatomy and function of the tricuspid and pulmonary valve, the right atrium, and the atrialised portion of the right ventricle, as well as the size and function of both ventricles, can be carefully evaluated by echocardiography (Fig 2). An apical displacement of the septal tricuspid valve leaflet from the insertion of the anterior leaflet of the mitral valve by more than 0.8 cm/m^2 body surface area distinguishes Ebstein's anomaly from other malformations associated with tricuspid regurgitation.[10] However, the quality of images can be compromised in non-cooperative patients or when only poor acoustic windows are available.

Real-time three-dimensional echocardiography (3DE) is a feasible method in addition to conventional two-dimensional echocardiography to evaluate the anatomy and the dynamic function of the abnormal tricuspid valve in young Ebstein's anomaly patients. It also permits clear differentiation from tricuspid valve dysplasia.[11] It can show extension of

tricuspid regurgitation and adds information regarding septal leaflet insertion and right ventricle volumes. Furthermore, the size, location, and shapes of septal defects can be visualised clearly. Even foetal hearts can be examined by 3DE. However, this modality is not yet widely used as a bedside technique.

Electrocardiogram

In most patients with Ebstein's anomaly, electrocardiogram findings are abnormal, especially in older patients, but they are not diagnostic. The rhythm is usually a normal sinus rhythm. P waves can be increased in amplitude as a sign of right atrium enlargement. P waves and the PR interval are often prolonged because of a delayed conduction in the enlarged right atrium. Small initial q waves may be found, occasionally seen in leads as far as V4, and are also considered to be due to right atrial enlargement. Signs of ventricular pre-excitation may be present. The QRS might show right axis deviation. It is usually of low voltage and prolonged, and it might exhibit a right bundle branch block pattern of various types in the right precordial leads (Fig 3). The right bundle branch block might be caused by pressure of the atrialised portion of the right ventricle onto the right septal surface. Left bundle branch block and atrioventricular block are also common.

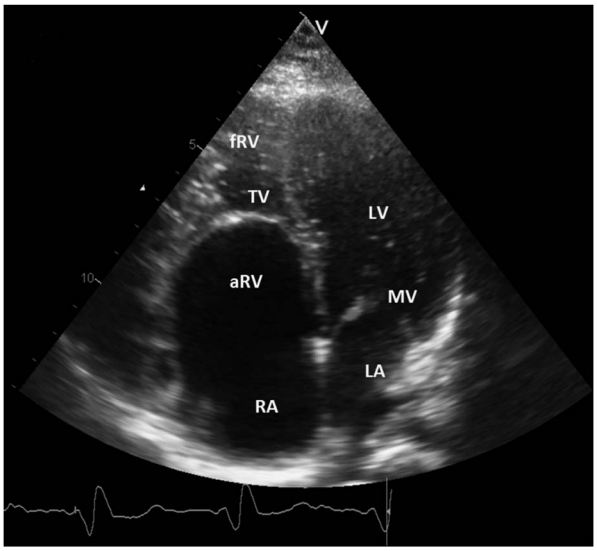

Figure 2.
Echocardiographic feature of a patient with Ebstein's anomaly demonstrating the apical displacement of the TV, dividing the right ventricle in an atrialised (aRV) and a functional portion (fRV). LA = left atrium; LV = left ventricle; MV = mitral valve; RA = right atrium; TV = tricuspid valve.

Exercise testing

Exercise testing may assist in determining the severity of the disease. Children, adolescents, and adults with Ebstein's anomaly may have limited exercise tolerance, defined as maximal oxygen consumption (VO_2) at peak exercise.[12–14] There is a strong negative correlation between blood oxygen levels and exercise tolerance. Observed heart rate at maximum exercise is lower than predicted.[13] The cardiac output at rest is within the lower range of normal for age. Driscoll et al.[12] showed that surgical repair improves exercise tolerance mostly because of the elimination of the right-to-left shunt in these patients. The exercise performance of patients without atrial septal defect was significantly greater than that of pre-operative patients with atrial septal defect, but was similar to that of patients who had atrial septal defect closure. This suggests that the presence and size of intracardiac right-to-left shunt might be a major contributing factor to exercise intolerance. The study of MacLellan-Tobert et al.[13] supports this. By shunting, only a portion of the systemic venous return reaches the lung for ventilator exchange of carbon dioxide. To maintain normal systemic arterial partial pressure of carbon dioxide, the patient must compensate by increasing ventilator exchange to allow for removal of carbon dioxide. Indeed, in Ebstein's anomaly patients minute

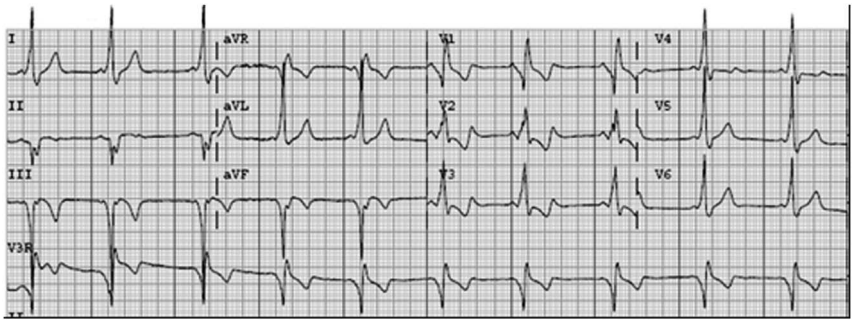

Figure 3.
A 12-lead electrocardiogram of an 8-year-old girl with Ebstein's anomaly. Signs of pre-excitation and repolarisation disorder are present.

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ventilation at rest and ventilatory equivalent for oxygen (VE/VO_2) at rest and during exercise are significantly higher before surgical repair than after.[12–14] In addition, improvements in tricuspid and consecutive right heart function might be another substrate for the improvement in exercise performance after surgery.[14]

However, it is difficult to establish from which age children can safely participate in exercise tests. Furthermore, young children might be more difficult to encourage to exercise to exhaustion. Interpretation of results should be performed carefully, whereas sex and body surface area influence outcomes.[15]

Chest radiography

The cardiac silhouette may vary from almost normal to a typical configuration of a box or balloon-shaped heart with a narrow waist. The greatly enlarged right atrium is mainly responsible, but also the dilated right ventricular outflow tract can be displaced outward and upward. The aorta cross-section is small, and the pulmonary trunk, which normally appears as a discrete convex bulge, is absent. The cardiothoracic ratio is an important factor influencing outcome of patients with Ebstein's anomaly. Pulmonary vascularity may be normal or decreased. When a newborn presents with systemic desaturation and the chest radiography shows cardiomegaly and decreased pulmonary vascularity, the diagnosis of Ebstein's anomaly should be considered.

Cardiac magnetic resonance imaging

Cardiac magnetic resonance imaging is a powerful tool providing anatomic and haemodynamic information that echocardiography alone cannot always provide. The extent of tricuspid valve regurgitation, the degree of right-to-left shunt, as well as the ventricular volumes and function can be well assessed. Tobler et al.[16] even conclude that the volume of the atrialised right ventricle is a novel cardiac magnetic resonance imaging measure, which may express severity of disease. To fully characterise right heart size and function, the atrialised right ventricle, functional right ventricle, and total right ventricle volumes need to be measured by adequate cine view of a four-chamber, axial and short-axis view. The axial view, however, appears to provide the most reproducible data.[17]

Cardiac magnetic resonance imaging is also particularly helpful in cases with associated cardiac malformation and allows accurate planning of the best surgical approach. Attenhofer et al.[18] compared the use of echocardiography and cardiac magnetic resonance imaging in patients with Ebstein's anomaly. They concluded that both provide complementary data. For appropriate risk stratification in Ebstein's anomaly, both examinations should be performed before surgery.

However, cardiac magnetic resonance imaging requires specific knowledge of anatomical variations and functional implications of Ebstein's anomaly, as well as high-resolution imaging, and extended patient compliance. General anaesthesia is commonly integrated into the examination protocol of younger children.

Medical management

Children with mild Ebstein's anomaly might not need any interventions during infancy, childhood, or adulthood. These patients, however, will need lifelong visits at the outpatient clinic. The frequency of these visits depends on their clinical condition. Others with moderate to severe types of Ebstein's anomaly – with or without associated malformations – might require medical treatment and/or intervention early in life.

Medical treatment

Little evidence is published about the use of medication in children with Ebstein's anomaly. In the large paediatric cohort of Kapusta et al.,[6] 41% of the children with Ebstein's anomaly ($n = 38$) required medication at presentation. Patients diagnosed during the first year of life received significantly more medication as compared with children diagnosed later in life. Prostaglandin was required in 23 of 93 patients, all in the neonatal period. Neonates often require short-term prostaglandin therapy in case of severe tricuspid regurgitation and severe cyanosis, due to the functional pulmonary valve atresia and the high pulmonary arteriolar resistance. Other required medication at presentation included diuretics ($n = 15$), digitalis ($n = 14$), and inotropics ($n = 5$). At the end of their study, 19% of their children were classified as New York Heart Association class II for which only a few patients required – a combination of – medication.[6] Medication included diuretics ($n = 5$), angiotensin-converting enzyme blocker ($n = 1$), digitalis ($n = 1$), beta-blocker ($n = 4$), and aspirin ($n = 2$).

Reemtsen et al.[19] reported on 16 pre-operative neonates of whom 88% were in need of prostaglandin and 56% required inotropics – dopamine, dobutamine, or epinephrine. Furthermore, inhaled nitric oxide may play an important therapeutic role in neonates with Ebstein's anomaly after discontinuation of prostaglandin.[20]

Rhythm anomalies, including re-entrant tachycardias, are comorbidities requiring medical intervention

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at any age in patients with Ebstein's anomaly. Supraventricular arrhythmias are the most common rhythm disturbance in children and tend to be recurrent and drug resistant. Currently, catheter ablation of accessory pathways is often performed and anti-arrhythmic drugs are less often prescribed. Delhaas et al.[8] found in 16 out of 93 patients of their paediatric population rhythm disturbances of which nine out of 14 of the surviving patients underwent ablation. Of these patients, four were still on antiarrhythmic drugs after a mean follow-up of 13 years and 3 months.

Radiofrequency ablation procedures

Up to one-third of the Ebstein's anomaly patients are presenting accessory pathways. These are usually right sided and may even be multiple.[8,21] Indications for radiofrequency ablation include apparent life-threatening arrhythmias and drug-refractory tachycardia. Radiofrequency ablation procedures in Ebstein's anomaly patients are challenging because of the anatomical and functional variations. The right atrium may be severely dilated, the atrioventricular junction might be less well demarcated, abnormal electrocardiograms can be recorded along the atrialised right ventricle, and catheter stability may be impaired.[21]

Acute success rates for patients with Ebstein's anomaly are similar to those of patients with other congenital heart diseases, but Ebstein's anomaly patients are more likely to have a recurrence.[22] Supraventricular tachyarrhythmias can also be successfully ablated at the time of operative repair.[23]

Surgical management

The customary objectives of surgical intervention in patients with Ebstein's anomaly are: (1) to reduce the severity and haemodynamic sequelae of tricuspid regurgitation and/or stenosis; (2) to improve hypoxaemia; and (3) to assist in controlling intractable tachyarrhythmias.

The indications for operation in the first two decades of life are multiple and vary widely. In neonates, they include overt heart failure, cyanosis, acidosis associated with tricuspid regurgitation, depressed right ventricle function, severe cardiomegaly, persistent need for inotropic support, ventilator dependency, and prostaglandin-dependent circulation.[19,24] After infancy, frequently reported indications for surgery include New York Heart Association class III or IV, progressive exercise intolerance, tachyarrhythmias refractory to drug treatment or not amenable to catheter-based intervention, recurrent palpitations or significant associated anomalies. Furthermore, patients with a cardiothoracic ratio >0.65 and persistent severe cyanosis may need surgical intervention.[25–28]

Neonates

Symptomatic neonates with Ebstein's anomaly can present with severe heart failure, cyanosis, and acidosis, and without surgical intervention most of them will die.[19,24,29] Surgical intervention includes tricuspid valve repair, right ventricle exclusion, or transplantation. The first essential surgical decision that needs to be made is whether a two-ventricle repair is feasible, valve repair, or not, right ventricle exclusion.[19,24] Right ventricle exclusion was first suggested by Starnes and associates in 1991,[29] but the technique has been modified over the last 10 years. If the functional portion of the right ventricle is inadequate or if there is right ventricle outflow tract obstruction, one might choose for this right ventricle exclusion. This is performed by patching the tricuspid valve, preferably with a fenestration of the patch (Figs 4 and 5).[19,30] A reduction right atrioplasty is performed and possibly also a right ventricle plication. Next, the right ventricle outflow tract is accessed. Finally, a Blalock–Taussig shunt provides pulmonary blood flow.

A two-ventricle approach may include modifications in the anterior leaflet creating functionally monocuspid tricuspid valve architecture. A reduction atrioplasty is performed with partial closure of the atrial septal defect, leaving behind a small fenestration. Finally, other cardiac defects are simultaneously repaired.[24]

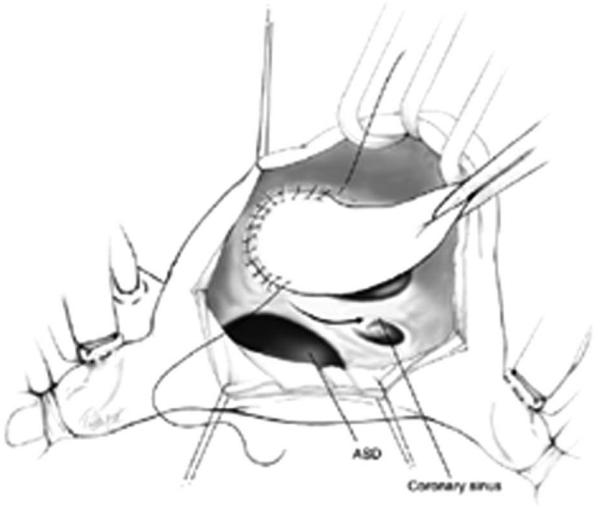

Figure 4.
A glutaraldehyde-fixed autologous pericardial patch is sewn at the "anatomic" level of the tricuspid valve annulus. ASD = atrial septal defect. Source: Reemtsen et al.[19]

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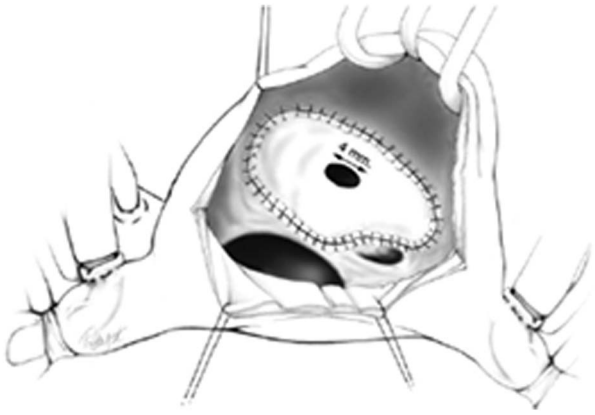

Figure 5.
A fenestration in the patch is created by using a 4-mm coronary punch. Source: Reemtsen et al.[19]

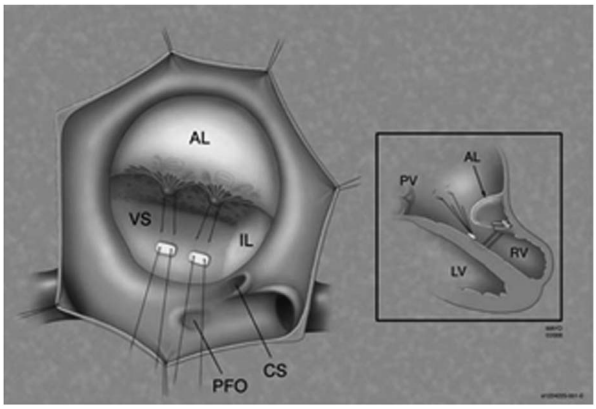

Figure 6.
The manoeuvres are designed to progressively bring the leading edge of the anterior leaflet (AL) closer to the ventricular septum (VS), or septal leaflet, in order to optimise leaflet coaptation and establish competence of the valve. The base of the intact major papillary muscle(s), which arises from the free wall of the right ventricle, is moved towards the ventricular septum at the appropriate level with pledgeted horizontal mattress sutures. Inset, coronal view of the right ventricle (RV) and the right atrium demonstrating a small dimple effect that occurs in the anterior free wall of the right ventricle after this manoeuvre is completed. CS = coronary sinus; IL = inferior leaflet; LV = left ventricle; PFO = patent foramen ovale; PV = pulmonary valve. Source: Dearani et al.[32]

The choice of surgical approach seems to be partially dependent on the clinic's experience. Reemtsen et al.[19] in 2006 described 16 critically ill neonates undergoing surgery. There were 12 patients who underwent right ventricle exclusion as initial palliation; in three patients, the tricuspid valve was repaired and one patient underwent heart transplantation. The overall survival was 69% with no late deaths reported at a mean follow-up of 27 months. Knott-Craig et al.[24] described in 2007 surgical intervention in 22 neonates and five young infants. In all but two patients, two-ventricle repair was performed. They reported a 74% survival rate with a median follow-up of 5.4 years. Boston et al. reported in 2011 the long-term results of 23 neonates and nine young infants. A biventricular repair was achieved in 91% of the patients. The early survival was 78% with one late death. The mean follow-up was 5.9 years.[31]

Nevertheless, some neonates and young infants with a primary biventricular repair will eventually need a bidirectional cavopulmonary shunt because the right ventricle fails to support the pulmonary circulation. This technique is called a "one and a half ventricle repair". To the best of our knowledge, there are no data available about the number of neonates who underwent a biventricular repair in the neonatal period, but were eventually in need of a bidirectional cavopulmonary shunt later in life. Surgical management of critically ill neonates and young infants remains challenging.

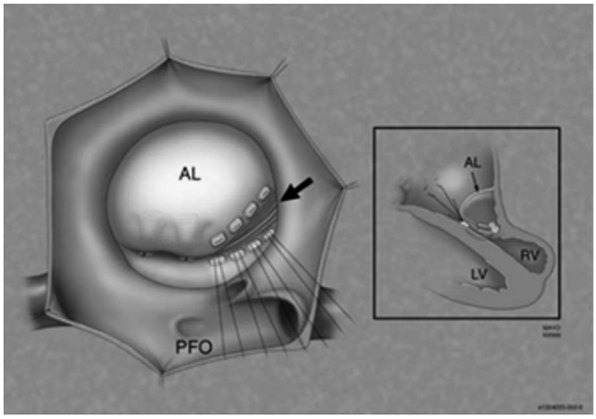

Figure 7.
The inferior angle of the tricuspid orifice is closed by bringing the right side of the anterior leaflet down to the septum and plicating the non-functional inferior leaflet in the process (arrow). Inset, after all of the mattress sutures are secured, improved proximity of the leading edge of the anterior leaflet with the ventricular septum is noted. Source: Dearani et al.[32]

Child, adolescent, and adult

Surgical treatment after infancy is much more promising. A biventricular repair is usually possible. Dearani et al.[32] described their experience in surgical treatment up to 2006. The repair was predominantly based on a satisfactory arrangement of the anterior leaflet (Figs 6–8). Some patients will have a large enough posterior leaflet permitting bicuspid reconstruction, and rarely a trifoliate repair is possible involving the septal leaflet as well. Various modifications in

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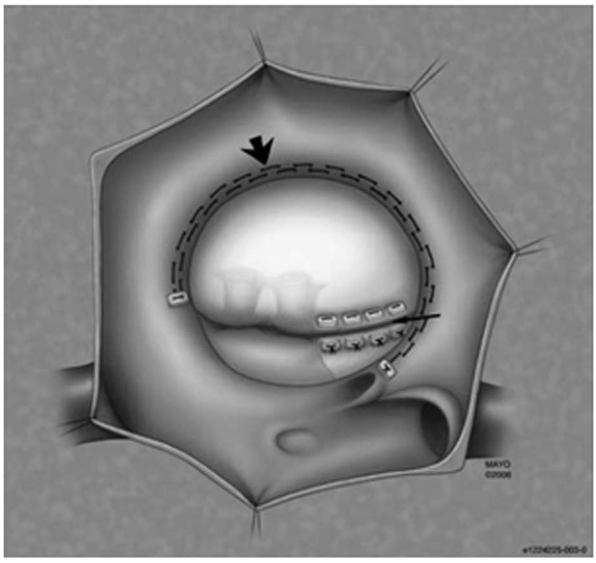

Figure 8.
Plication of the inferior angle of the annulus with pledgeted mattress sutures (arrow). An anterior purse-string annuloplasty (arrowhead) may be performed to further narrow the tricuspid annulus. This annuloplasty may begin at the anteroseptal commissure, anterior to the membranous septum, and ends beyond the inferoseptal commissure adjacent to the coronary sinus. Alternatively, the annuloplasty can be performed posterolaterally to reduce the size of the annulus, which also brings the free wall closer to the septum. Source: Dearani et al.[32]

the technique of repair have been advanced with growing experience.

In 2004, another technique for valve repair was described by Wu et al.[33] This repair restores to near normal anatomical and physiological functioning of the tricuspid valve. The displaced posterior and septal leaflets with some chordae tendinae and corresponding papillary muscle are detached from the annulus and ventricular wall. The leaflets are then reattached to the native posterior annulus with re-implantation of the papillary muscle. Most of the atrialised portion of the ventricular wall is excised and the tricuspid annulus is plicated. In case of a severely hypoplastic septal leaflet, autologous pericardial tissue is used to create a new leaflet.

In 2009, Malhotra et al.[34] described a technique with reconstruction performed at the functional rather than the anatomic orifice. Detachment and re-implantation of valve leaflets is avoided and only limited plication is performed at the level of the displaced valve.

Da Silva et al.[35] proposed a conic reconstruction of the tricuspid valve in 2004. This technique consists of (almost) total detachment of the anterior tricuspid megaleaflet from the ventricular wall and valvular ring. This leaflet is transformed into a cone rotating it clockwise. The vertex remains fixed in the right ventricular tip. Then the atrialised right ventricle is plicated to exclude its thin part. The true tricuspid annulus is plicated to make up the new annulus at the anatomically correct level. After its plication, the base of the cone is sutured to the tricuspid ring. The end result of this cone reconstruction includes 360° of leaflet tissue surrounding the right atrioventricular junction.

In general, the "one and a half ventricle repair" using a bidirectional cavopulmonary shunt is preserved for Ebstein's anomaly patients with a severely enlarged and poorly functioning right ventricle.[32,33] This shunt reduces the preload of the right ventricle but optimises the preload of the left ventricle. It therefore reduces both the risk of post-repair tricuspid stenosis and of potential progression of residual tricuspid regurgitation.

Valve reconstruction is preferable to replacement given the attendant risks of thromboembolism and the need for re-intervention with patient's growth. Tricuspid valve replacement in adulthood is also a risk factor for 30-day mortality and for complications.[36] However, in case of extreme hypoplasia of the anterior leaflet and severe right ventricular dysfunction, valve replacement may be the only option. The type of implanted prosthesis, mechanical or biological, seems not to affect the outcome.[37] When replacing the valve, the conduction tissue and the right coronary artery should be protected. All leaflet tissue should be removed to avoid obstruction.[32] In case of severe dysfunction of the left ventricle, cardiac transplantation should be considered.

Outcome of surgical intervention

Surgical outcomes vary with an early survival rate of 86–96% and a 10-year survival rate up to 96%.[25,26,28] Studies included different surgical procedures and a subset of included patients had prior surgical intervention. Although re-operations were associated with a trend towards higher mortality, this did not reach significance level.[26] No significant difference was found between valve repair and valve replacement groups. Commonly reported post-operative complications include atrial and ventricular arrhythmias, low cardiac output, and respiratory insufficiency.[25,26,28] In all, 77–82% are free from re-operation 10 years after surgery.[25,28]

Pre-operative risk factors for mortality include an increased pre-operative right ventricle size with decreased systolic function, pre-operative decreased left ventricle systolic function, cardiothoracic ratio >0.65, and variables associated with right ventricle outflow tract obstruction or narrow pulmonary arteries.[25] In addition, younger age and corresponding

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low bodyweight and body surface area $<0.5\,m^2$ is associated with higher mortality rates.[26] Pre-operative risk factors for re-operation are pulmonary valve stenosis, hypoplastic or stenotic pulmonary arteries, prior cavopulmonary shunt, and younger age at the time of operation.[25]

Factors associated with poor prognosis of Ebstein's anomaly

The prognosis of patients diagnosed in the neonatal period is generally poor. Survival rates vary from 56–83% after 1 month to 55–71% after 1 year.[7,38] In older children and adolescents, the outcome is superior. Multiple factors associated with poor outcome have been described over the last two decades. They include presentation in foetal or neonatal life, echocardiographic grade of severity 3 or 4 by Celermajer index, New York Heart Association class III or IV, associated anomalies, and a cardiothoracic ratio of ≥ 0.65.[4,7,39] Furthermore, male sex and the absence of Wolff–Parkinson–White syndrome are reported as negative predictors of long-term survival, in contrast to what might be expected in a larger cohort.[39]

Kapusta et al.[6] also found that young age at presentation (<12 months) is strongly associated with death. The mortality rate in these children is 7.9 times the risk in the group of children ≥ 12 months. In addition to hepatomegaly, the need for medication and mechanical ventilation at presentation were associated with death. Furthermore, patients with a patent arterial duct, ventricular septal defect, or pulmonary valve defects have a higher mortality rate.

Conclusion

Ebstein's anomaly is a rare and complex congenital disorder characterised by a rotational displacement of at least one of the leaflets of the tricuspid valve. Clinical features vary widely from mild symptoms presenting in adolescence to severe neonatal cyanosis and heart failure resulting in death. New diagnostic methodologies are available for establishing severity of disease. Many patients develop arrhythmias and may need recurrent radiofrequency ablation procedures. Severely cyanotic neonates on mechanical ventilation with persistent need for inotropic support often require immediate surgical treatment in order to survive, but these interventions remain extremely challenging. Therapeutic options after infancy are more promising. New surgical techniques have been developed over the last decade and they seem to alter both prognosis and quality of life in the first two decades of life.

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Conflicts of Interest

None.

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