Tricuspid Valve Abnormalities

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Abstract

Tricuspid valve abnormalities are relatively rare lesions in the spectrum of congenital heart disease which can affect the leaflets, chordal apparatus, and/or papillary muscles of the tricuspid valve. Ebstein anomaly is the most common form of congenital tricuspid valve disease. Although outcomes remain poor in neonates with important tricuspid valve disease, Ebstein anomaly can be managed successfully in children, adolescents, and young adults with recent surgical advances and current interventional techniques. Comprehensive cardiac imaging is essential to allow for accurate diagnosis and appropriate decision-making, and should always be performed by an advanced cardiac imaging specialist with expertise in congenital heart disease. In this chapter, anatomical features pertaining to the range of congenital heart defects which affect the tricuspid valve are reviewed, current imaging techniques are detailed, and contemporary approaches to management are discussed.

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1 Morphology of the Normal Tricuspid Valve

In the structurally normal heart, the tricuspid valve guards the inlet portion of the right ventricle. The three leaflets of the tricuspid valve are located in septal, anterior, and posterior (also called inferior or mural) positions. The papillary muscles (medial, anterior, inferior) provide support for the tricuspid valve leaflets. Unlike the mitral valve, however, chordal attachments from the septal leaflet of the tricuspid valve to the ventricular septum are present. The tricuspid valve septal attachments are more apically positioned compared to the anterior mitral leaflet (creating an "offset" between the right and left atrioventricular valve positions); a distance of up to 8 mm/m^2 between the attachment of the septal tricuspid leaflet and the anterior mitral leaflet is considered normal (Edwards 1993; Seward 1993).

2 Congenital Defects of the Tricuspid Valve

Isolated congenital malformations of the tricuspid valve are relatively rare lesions. Tricuspid valve disease can affect the leaflets, chordal apparatus, and/or papillary muscles. For the purposes of this discussion, tricuspid valve disease will be reviewed according to morphologic abnormalities pertaining to:

(a) Displacement of the valve leaflets from the atrioventricular junction towards the apex of the right ventricle (Ebstein anomaly)
(b) Hypoplasia of the valve annulus or thickening of the valve leaflets (tricuspid valve stenosis)
(c) Abnormal/incomplete leaflet development (tricuspid valve dysplasia) or
(d) Tricuspid valve disease present in the context of other forms of complex congenital heart disease

2.1 Ebstein Anomaly

Although a rare disease (occurring in approximately 3–5 per 100,000 live births and accounting for less than 1% of all forms of congenital heart disease) (Samanek and Voriskova 1999; Correa-Villasenor et al. 1994), Ebstein anomaly is one of the most striking abnormalities of the tricuspid valve. The malformation is often complex and typically involves the tricuspid valve leaflets, the chordal apparatus, and the myocardium of the right ventricle. Additionally, Ebstein anomaly is frequently associated with additional lesions affecting the right or left heart and can encompass an unusually wide clinical spectrum. Diagnostic criteria for Ebstein anomaly typically include (1) apical (downward) displacement of the tricuspid valve functional annulus (hinge point of the valve from the atrioventricular ring >8 mm/m^2), (2) adherence of the tricuspid valve leaflets to underlying myocardium (failure of delamination), (3) redundancy, fenestrations, and/or tethering of the anterior leaflet, (4) dilation of the true tricuspid valve annulus (right atrioventricular junction), and (5) dilation of the atrialized portion of the right ventricle (Attenhofer Jost et al. 2012a).

While Ebstein anomaly diagnosed in fetal or neonatal life continues to be associated with dismal outcomes (perinatal mortality estimated at 45%) even in contemporary series (Yetman et al. 1998; Wald et al. 2005; Bove et al. 2009; Freud et al. 2015), in some instances underlying Ebstein anomaly may remain undetected throughout adult life to be discovered only at the time of autopsy (Celermajer et al. 1994). The clinical presentation of symptomatic Ebstein anomaly in the adult may include fatigue, palpitations, decreased exercise tolerance, edema, and/or cyanosis. Tachyarrhythmias (atrial or ventricular), heart failure, paradoxical embolism, transient ischemic attacks or stroke, endocarditis, and sudden death are recognized sequelae of Ebstein anomaly (Celermajer et al. 1994). Contemporary guidelines suggest that symptomatic patients should be considered for operative repair or replacement of the tricuspid valve (Attenhofer Jost et al. 2012b; Warnes et al. 2008; Baumgartner et al. 2010). Younger age at diagnosis, male gender, cardiothoracic ratio ≥0.65, and the severity of tricuspid valve leaflet displacement on echocardiography have been identified as independent predictors of cardiac

mortality in patients with Ebstein anomaly (Attie et al. 2000). The functional integrity of the tricuspid valve in Ebstein anomaly generally depends on several factors which include the degree of displacement of the right atrioventricular junction, extent of dysplasia of septal and posterior leaflets, presence and size of fenestrations, size and restriction of the anterior leaflet, and dilatation of the "functional" tricuspid valve annulus.

Comprehensive cardiac imaging is crucial for accurate diagnosis and decision-making, and should always be performed by an advanced cardiac imaging specialist with expertise in congenital heart disease. Multi-modality imaging should be employed to characterize morphology of the tricuspid valve leaflets and underlying mechanism of their dysfunction (transthoracic ± transesophageal echocardiography); to quantify dimensions of the right atrium, atrialized right ventricle, and functional right ventricle (cardiac magnetic resonance imaging [CMR] or cardiac CT); and to delineate associated congenital heart disease lesions as well as structure of the left myocardium (echocardiography ± cardiac magnetic resonance imaging or cardiac CT).

2.1.1 Displacement of the Atrioventricular Junction

Ebstein anomaly arises from an arrest in fetal development which results in incomplete delamination of the tricuspid valve leaflets (persistent adherence of the septal ± posterior leaflets to the underlying myocardium) resulting in an apically displaced attachment of the leaflet(s) into the right ventricle in a spiral fashion such that the valve is displaced anteriorly and rightward. A distance of >8 mm/m^2 between the hinge points of the anterior mitral valve leaflet and tricuspid valve septal leaflet is considered diagnostic for Ebstein anomaly (Seward 1993). The apical displacement of the right atrioventricular junction results in two components of the right ventricle. The portion between the true tricuspid annulus and the functional annulus is termed the "atrialized" right ventricle and the remainder of the right ventricle is designated as the "functional" right ventricle (Fig. 1). The degree of adherence of the posterior and septal leaflets determines the spectrum of disease severity, ranging from mild forms with minimal displacement of the septal leaflet to complete failure of delamination of the leaflets resulting in an imperforate membrane (tricuspid sac).

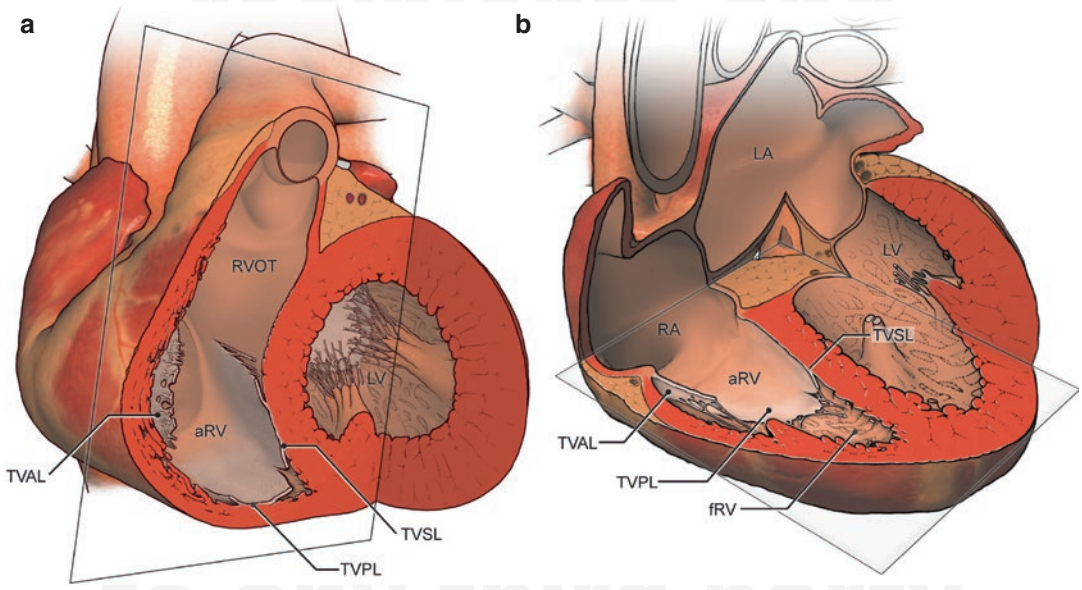

Fig. 1 Schematic demonstrating Ebstein anomaly in the short axis view (**a**) and axial view (**b**). *aRV* atrialized right ventricle, *fRV* functional right ventricle, *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RVOT* right ventricular outflow tract, *TVAL* tricuspid valve anterior leaflet, *TVPL* tricuspid valve posterior leaflet, *TVSL* tricuspid valve septal leaflet

2.1.2 Development of the Septal and Posterior Leaflets

In Ebstein anomaly, the displaced septal and posterior leaflets of the tricuspid valve can often appear thickened or dysplastic (Freedom 1997). The septal leaflet can assume a wide spectrum of anatomic malformations or may be altogether absent. Chordae may be few or even absent and are typically shortened and dysplastic with anomalous insertions into the right ventricular wall (Freedom 1997). Tricuspid regurgitation is commonly seen in Ebstein anomaly, often a result of leaflet malcoaptation, although fenestrations can provide an additional mechanism for valve insufficiency; tricuspid valve stenosis has been described although is infrequently observed (Fig. 2).

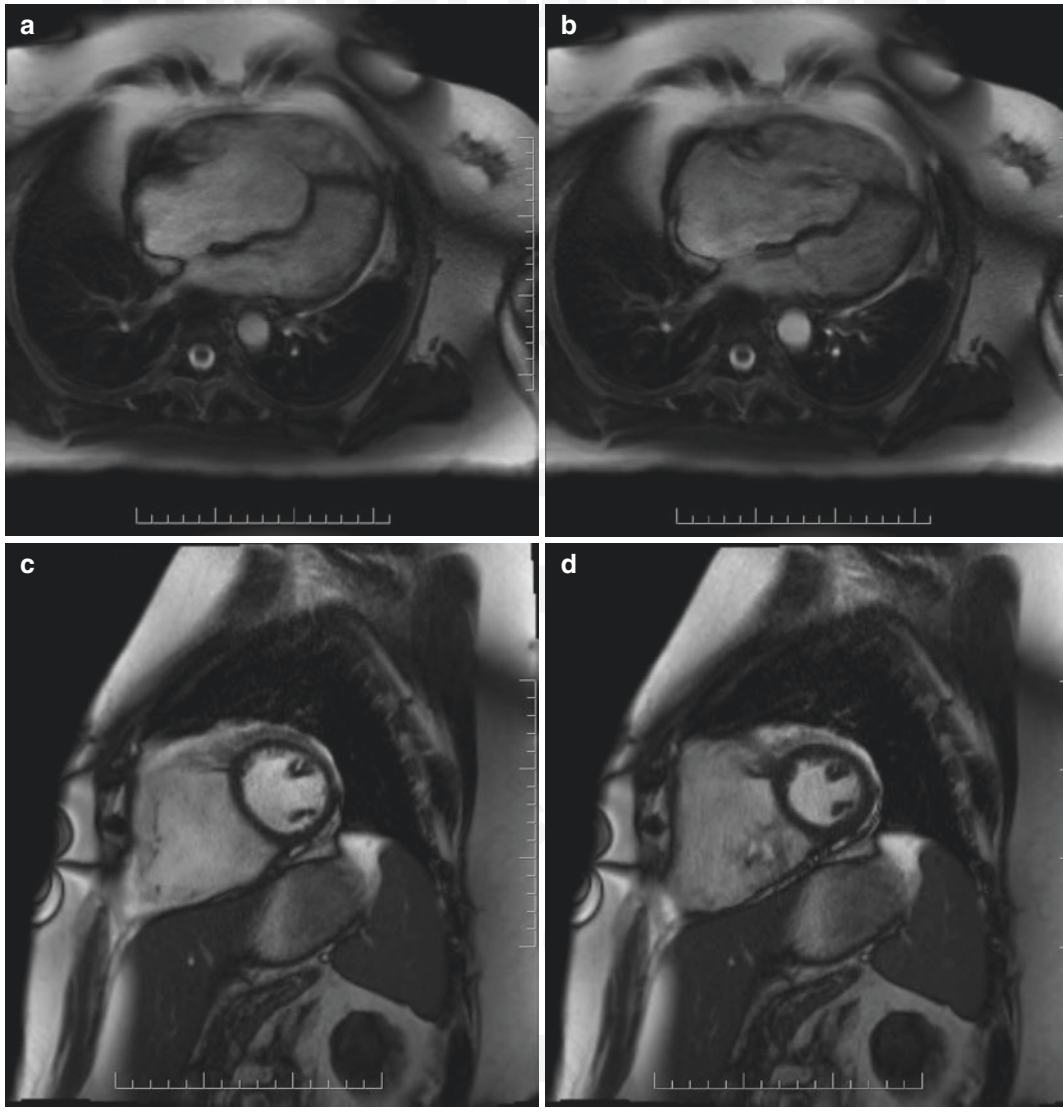

Fig. 2 Cardiac magnetic resonance imaging of severe tricuspid regurgitation in Ebstein anomaly; (**a**) four chamber view in diastole, (**b**) four chamber view in systole, (**c**) short axis view in diastole, (**d**) short axis view in systole

2.1.3 Morphology of the Anterior Leaflet

In contrast to the septal leaflet, the anterior leaflet of the tricuspid valve is rarely displaced. The anterior leaflet is characteristically elongated and, if sufficiently redundant, may contribute to obstruction of the right ventricular outflow tract. Fenestrations within the leaflet are commonly seen. Tethering of the anterior leaflet frequently arises from chordal attachments to the ventricular free wall and/or displaced papillary muscles. Size and mobility of the anterior leaflet are important determinants for tricuspid valve anatomic repair (Cone reconstruction) (Brown 2011), and therefore detailed description of leaflet morphology is critical when surgery referral is being considered. In rare situations, the anterior leaflet forms an atretic membrane (Ho et al. 2005).

2.1.4 Development of the Right Ventricle

The atrialized portion of the right ventricle is typically dilated and thinned in the patient with Ebstein anomaly. Size of the atrialized right ventricle has been associated with poor prognosis in both children (Celermajer et al. 1992) and adults (Celermajer et al. 1994; Attie et al. 2000). Morphologic features of the functional right ventricle apex often include increased trabeculations although the myocardium itself may be thinned or even aneurysmal. In the extreme form, replacement of the myocardium with fibrous tissue has been observed. Hypoplasia of the functional right ventricle in neonates and children is typically a result of marked apical displacement of the atrioventricular junction. In contrast, the functional portion of the right ventricle is often described as normal in size or even dilated in the adult (Yalonetsky et al. 2011; Lee et al. 2012; Fratz et al. 2012) and has been attributed to chronicity and severity of tricuspid valve regurgitation (Tobler et al. 2011).

2.1.5 Associated Cardiac Abnormalities

Numerous additional congenital heart lesions have been described in conjunction with Ebstein anomaly. An interatrial communication, such as a secundum atrial septal defect or a patent foramen ovale, is the most frequently reported association (80% of cases) (Danielson et al. 1992) (Fig. 3). Right-to-left shunting at atrial level may give rise to cyanosis or may allow for paradoxical emboli which may result in transient ischemic attacks or stroke. Ventricular septal defects are occasionally associated with Ebstein anomaly. Associated right heart lesions include pulmonary stenosis, pulmonary atresia, or tetralogy of Fallot; associated left heart lesions include bicuspid aortic valve, aortic stenosis, aortic atresia, coarctation of the aorta, and mitral valve defects. Noncompaction of the left ventricular myocardium is a well-described association (Attenhofer Jost et al. 2005) (Fig. 4). The apical displacement of the septal tricuspid valve may result in discontinuity of the central fibrous body and septal atrioventricular ring, thereby allowing for accessory pathway formation, as seen in Wolff–Parkinson–White syndrome (Khositseth et al. 2004), which provides a substrate for supraventricular tachycardia. Atrial enlargement may give rise to atrial fibrillation or atrial flutter. Ventricular arrhythmias, in the setting myocardial fibrosis or left ventricular noncompaction, have been described although are far less common than atrial arrhythmias.

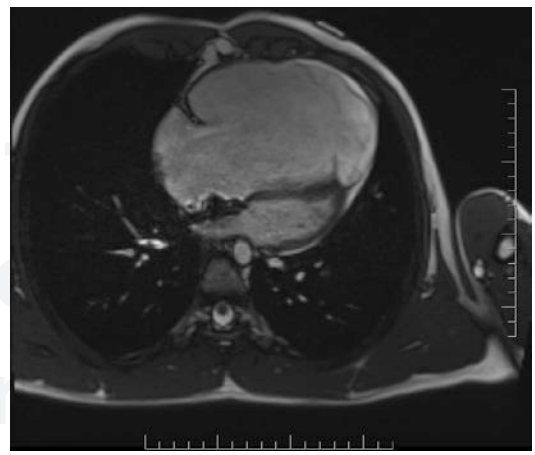

Fig. 3 Cardiac magnetic resonance imaging of an Amplatzer device closure of an atrial septal defect in a patient with Ebstein anomaly

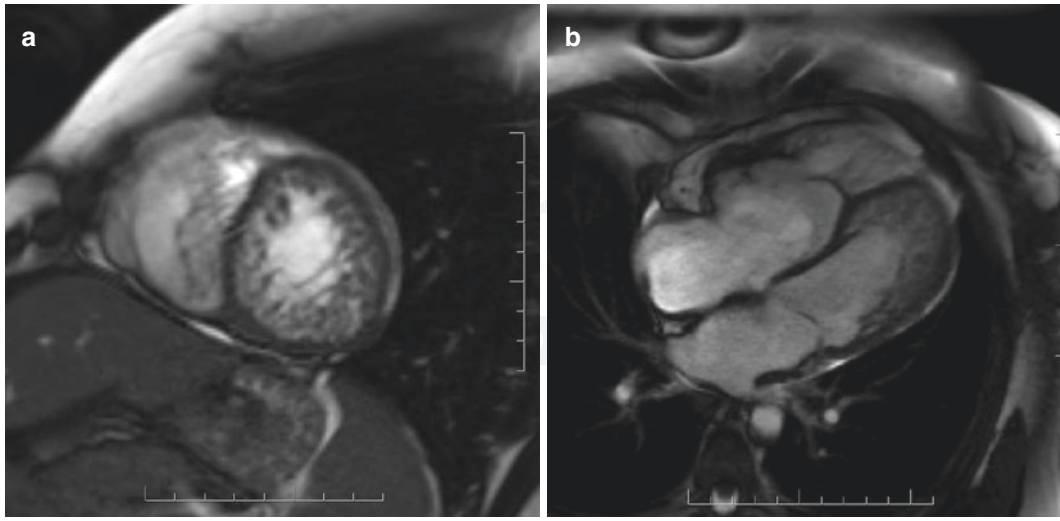

Fig. 4 Cardiac magnetic resonance imaging of left ventricular noncompaction in a patient with Ebstein anomaly in short axis (**a**) and four chamber views (**b**)

2.2 Tricuspid Stenosis

Isolated stenosis of the tricuspid valve stenosis is a rare lesion and is typically congenital. Stenosis is usually seen at valvular level due to hypoplasia or underdevelopment of the tricuspid annulus or thickening of the valve leaflets with reduced valve orifice. Occasionally, stenosis is supravalvular, and occurs as a result of a supravalvular ring or membrane at the level of the tricuspid annulus or at the midportion of the leaflets. In isolated valvular tricuspid stenosis the reduced valve orifice may result from thickened or hypoplastic tricuspid leaflets, underdeveloped or fused commissures, abnormal chordal attachments (i.e., parachute deformity) (Freedom 1997), and/or annular hypoplasia. Significant tricuspid stenosis can result in dilation and hypertrophy of the right atrium, and in some cases, hepatomegaly.

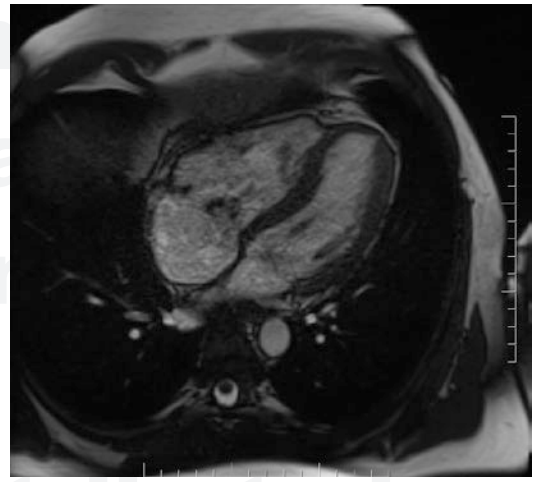

Fig. 5 Cardiac magnetic resonance imaging of a tricuspid valve dysplasia and severe TR

2.3 Tricuspid Valve Dysplasia

Tricuspid regurgitation is commonly a secondary finding in the presence of right ventricular disease (i.e., pressure/volume overload, systolic/diastolic dysfunction). Rarely, tricuspid regurgitation is a primary manifestation of tricuspid valve dysplasia. Tricuspid valve dysplasia is not a single entity but encompasses various congenital malformations such as malformation of the valve leaflets, hypoplasia of the papillary muscles, and/or abnormalities of tendinous chordae apparatus. Tricuspid valve leaflet hypoplasia and tethering, most commonly involving the septal leaflet, resulting in tricuspid regurgitation, is the most frequent expression of tricuspid valve dysplasia (Edwards 1993) (Fig. 5).

2.4 Tricuspid Valve Disease Associated with Other Congenital Lesions

The tricuspid valve may be involved in more complex forms of congenital heart disease. Congenital corrected transposition of the great arteries (cc-TGA) is often associated with an Ebstein-like malformation of the tricuspid valve (Fig. 6). In this condition, the tricuspid valve serves as a systemic atrioventricular valve and, therefore, is exposed to higher pressure than in typical Ebstein anomaly where the tricuspid valve is a subpulmonary atrioventricular valve. The tricuspid valve leaflets in this condition are often dysplastic and, as in usual Ebstein anomaly, apical displacement of the atrioventricular junction may be present. Unless bona fide Ebstein disease, the anterior leaflet is usually not elongated and the incomplete delamination of the septal and posterior leaflets are limited such that the atrialized portion of the right ventricle is not particularly enlarged (hence the designation Ebstein-like anomaly) (Allwork et al. 1976; Silverman et al. 1995; Anderson et al. 1978; Horvath et al. 1994). Annular hypoplasia and tricuspid stenosis have been described in various forms of complex congenital heart disease such as tetralogy of Fallot, pulmonary atresia with intact ventricular septum, transposition of the great arteries, and double-outlet right ventricle. A rare form of tricuspid valve disease is the so-called "double-orifice tricuspid valve" which has been seen in association with Ebstein anomaly, tricuspid valve dysplasia, tetralogy of Fallot, and atrioventricular septal defect (AVSD). Extreme forms of Ebstein anomaly or severe tricuspid stenosis/atresia will result in profound underdevelopment (hypoplasia) of the right ventricle resulting in "single ventricle physiology" of the left ventricular type.

3 Management

Much of the literature pertaining to management of the adult with tricuspid valve disease focuses on those with Ebstein anomaly given the rarity of the other tricuspid valve lesions (described above). With advancing age, survival in the adult with unrepaired Ebstein anomaly substantially decreases over time. In a cohort of 72 unoperated patients over the age of 25 years, Attie and colleagues demonstrated that survival was 76% at 10 years of follow-up with a marked decrease to 41% at 20 years (Attie et al. 2000). The American Heart Association states that surgery in the adult with Ebstein anomaly should only be performed by

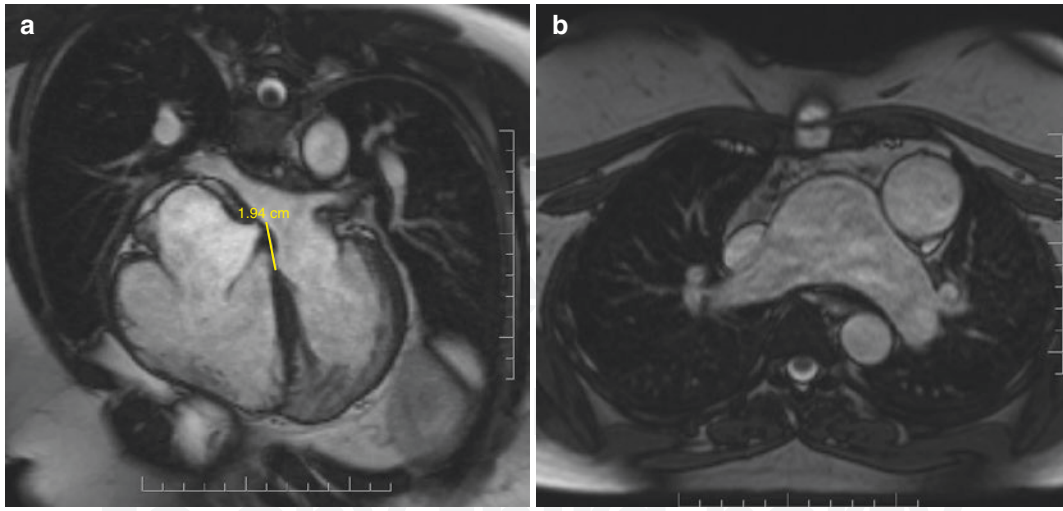

Fig. 6 Cardiac magnetic resonance imaging of congenitally corrected transposition of the great arteries with Ebstein-like anomaly of the tricuspid valve (systemic atrioventricular valve) (**a**) and demonstration of anterior and leftward position of the aorta relative to the pulmonary artery (**b**)

surgeons with training and expertise in adult congenital heart disease and outlines the following as class 1 indications for operative intervention: development of symptoms/decline in exercise testing, development of cyanosis, presence of paradoxical emboli, progressive enlargement of cardiothoracic silhouette on chest X-ray, and progressive enlargement or dysfunction of the right ventricle (grade of evidence B for all listed criteria) (Warnes et al. 2008). The European Society of Cardiology lists virtually identical criteria for surgical intervention (class 1 or IIa for factors listed above, grade of evidence C) (Baumgartner et al. 2010).

The goals of surgery typically include improvement in tricuspid valve function, closure of residual intracardiac shunts (atrial/ventricular septal defects), down-sizing of the right atrium through plications and/or arrhythmia surgery (typically the Maze procedure). Specific surgical strategies may include tricuspid valve repair (traditionally monocusp repair and more recently Cone repair), or tricuspid valve replacement, and in the event of extreme right ventricular hypoplasia insertion of a direct superior vena cava to pulmonary artery connection (i.e., a "Glenn" shunt to offload the right ventricle, or a one and a half ventricle repair) (Fig. 7). In rare occasions, severe Ebstein anomaly with a markedly attenuated right ventricle is best treated with a Fontan palliation (single ventricle strategy) (Fig. 8). Given the variability in right heart function and the challenges in assessment of systolic function in the presence of significant tricuspid valve regurgitation, surgery for Ebstein anomaly

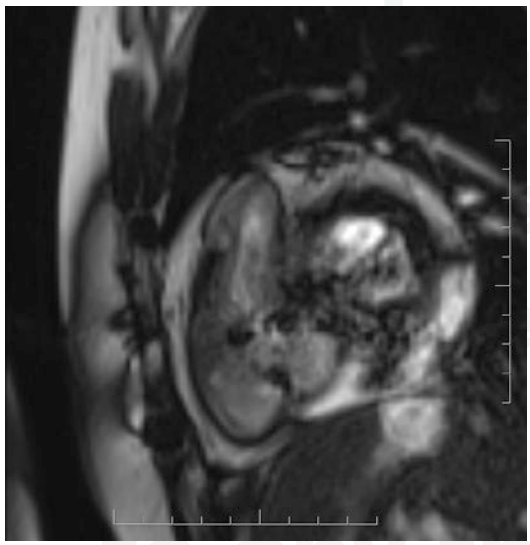

Fig. 7 Cardiac magnetic resonance imaging demonstrating prosthetic tricuspid valve replacement in a patient with Ebstein anomaly

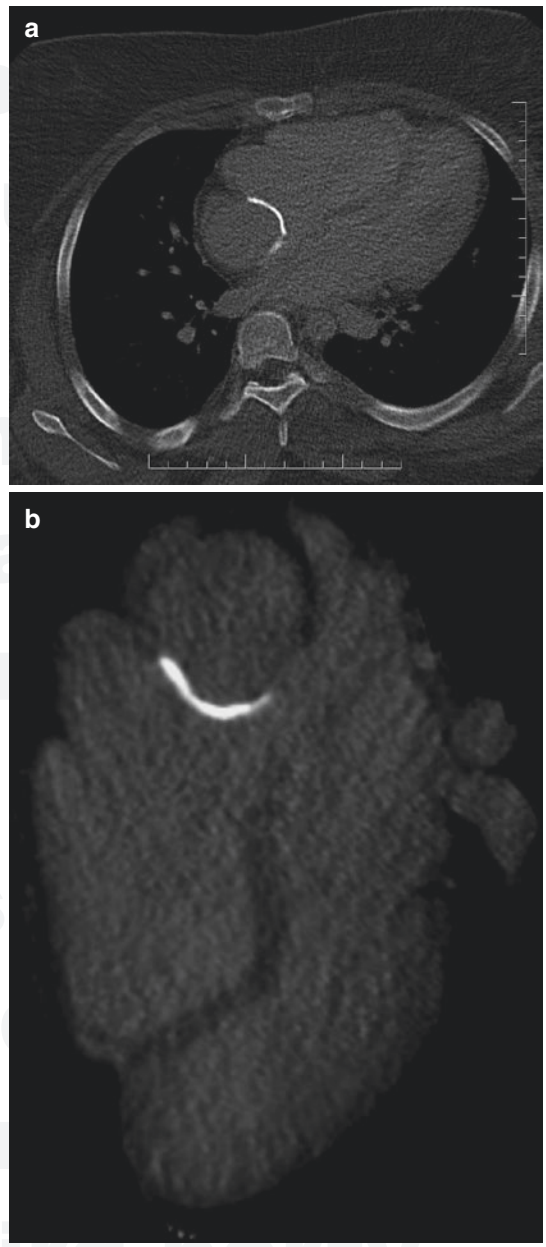

Fig. 8 Cardiac CT of a patient with underlying Ebstein anomaly palliated with a lateral tunnel Fontan, now calcified. Axial view (**a**) and reconstructed four chamber view (**b**)

can incur a perioperative mortality not typically seen in other forms of congenital heart disease. Therefore, the decision to proceed with surgery in a given individual should involve detailed discussion amongst a specialized team of adult congenital heart disease specialists, congenital heart disease surgeons, cardiac anesthesiologists, and in some cases, electrophysiologists. The cone surgical repair, first described by da Silva (da Silva et al. 2007), involving the delamination and rotation of the anterior septal leaflet with use of remnant septal and posterior leaflets to create a cone with bicuspid coaptation of valve leaflets, is now the considered the surgical repair of choice and is directed by the adequacy of the anterior leaflet size and morphology. Recent CMR studies have demonstrated that following Cone repair, right ventricular end-diastolic volumes decrease, right ventricular ejection fraction remains the same or decreases (Lange et al. 2015; Li et al. 2016), and pulmonary forward flow and left ventricular filling increases corresponding to an improvement in aerobic capacity on exercise testing (Ibrahim et al. 2015).

4 Imaging Techniques

4.1 Chest X-Ray

The (erect) chest X-ray can be normal or nearly normal in mild cases of Ebstein anomaly. The characteristic findings of a "globular" cardiac silhouette, described in patients with severe Ebstein anomaly, relates to marked enlargement of the right atrium. Cardiomegaly on X-ray is independently associated with adverse outcomes in patients with Ebstein anomaly (Attie et al. 2000). Contemporary management guidelines for adults with congenital heart disease suggest that enlarged or enlarging cardiothoracic ratio, as measured on chest X-ray, can be considered an indication for operative repair (Warnes et al. 2008; Baumgartner et al. 2010).

4.2 Echocardiography

Transthoracic echocardiography is considered the imaging modality of choice for the initial diagnosis of tricuspid valve disease. Characterization of valve morphology and function (i.e., stenosis or regurgitation) can be reliably achieved using transthoracic echocardiography primarily, and transesophageal echocardiography occasionally. Estimation of right ventricular pressure, cardiac chamber dimensions (right atrial, atrialized right ventricular, and ventricle sizes), and ventricular systolic function can be achieved; evaluation of associated congenital heart disease lesions (such as atrial or ventricular septal defects) can be accomplished. Following initial diagnosis, transthoracic echocardiography remains the imaging modality of choice for first-line surveillance of disease progression in the adult given its widespread availability. Detailed anatomic and functional review of the right heart, with specific focus on tricuspid valve morphology, as detailed above, is essential for patients referred for surgical repair. Severity of Ebstein anomaly, as determined by linear extent of apical displacement of the septal leaflet indexed to body surface area (Seward 1993) or a ratio of the areas of the right atrium and atrialized right ventricle as compared with the remainder of the heart can be readily accomplished by echocardiography (Celermajer et al. 1992).

4.3 Cardiovascular Magnetic Resonance Imaging (CMR)

Virtually all aspects of Ebstein anomaly are reliably characterized using CMR, with the exception of detailed characterization of some aspects of valve morphology. As such, CMR is an important, although emerging imaging modality in the preoperative and postoperative assessment of patients with tricuspid valve disease, such as Ebstein anomaly. CMR can provide morphologic and physiologic information not reliably obtained using echocardiography alone. Steady-state free-precession (SSFP) cine imaging can allow for characterization of tricuspid valve morphology in patients with poor acoustic windows, can provide quantification of right heart volumes (atrialized right ventricle and functional right ventricle) and ventricular systolic function (Yalonetsky et al. 2011). Several authors have reported lower interobserver variability in the axial plane as compared with the short axis view

(Yalonetsky et al. 2011; Fratz et al. 2012). Phase contrast flow analysis can allow for estimation of magnitude of an intracardiac shunt (using comparison of flow volumes through the main pulmonary artery and aorta) and measurement of tricuspid regurgitation (by comparison of main pulmonary artery flow and right ventricular stroke volume). Magnetic resonance angiography (MRA) should be used for assessment of associated vascular abnormalities, such as abnormalities in pulmonary arterial development, coarctation of the aorta, or patent ductus arteriosus.

The role of CMR for risk stratification and preoperative assessment of the patient with Ebstein anomaly is emerging, although as of yet is incompletely defined. Given the complexities of right ventricular anatomy in the patient with Ebstein anomaly it is not surprising that echocardiographic indices of right ventricular systolic function correlate poorly with CMR-derived assessment of right ventricular function (Kuhn et al. 2016). A recent prospective study of 16 patients with Ebstein anomaly at the Mayo clinic comparing information provided by echocardiography and CMR as compared with interoperative findings revealed that CMR is superior to echocardiography for assessment of some aspects of tricuspid valve anatomy (specifically posterior leaflet morphology and presence of valve fenestrations) and that quantification of right heart size and function was only possible with CMR; these authors concluded that it is their opinion that CMR be a "compulsory" component of preoperative screening for the patient with Ebstein anomaly (Attenhofer Jost et al. 2012a). That the functional right ventricle is not diminutive, as suggested by echocardiography, but is normal in size or, more commonly, is enlarged when indexed for body surface area (some reports suggest a median increase in right ventricular end-diastolic volume more than twice normal) is a relatively new observation afforded by CMR and is likely explained by the presence of chronic tricuspid regurgitation (Fratz et al. 2012; Tobler et al. 2011). A severity score for Ebstein anomaly was first described by Celermajer derived from chamber areas apparent in the four chamber view on echocardiography ([right atrium + atrialized right ventricle]/[functional right ventricle + left atrium + left ventricle]) and was related to adverse outcomes (Celermajer et al. 1992). The first CMR-derived index of severity of Ebstein anomaly was described by Tobler and colleagues, specifically the ratio of functional right ventricular end-diastolic volume/left ventricular end-diastolic volume ratio was inversely related to peak aerobic capacity on cardiopulmonary exercise testing; however volume of the atrialized right ventricle, another novel measure, was the only CMR variable found to be independently predictive of exercise capacity (Tobler et al. 2011). More recently, Hosch et al. proposed a simplified severity index relating CMR-derived volumes of all right heart to all left heart structures ([right atrium + atrialized right ventricle + functional right ventricle/left atrium + left ventricle]) which was found to be correlated to a range of clinical parameters of heart failure and functional capacity (Hosch et al. 2014).

Although CMR is widely cited as the imaging modality of choice to evaluate anatomy, physiology, and ventricular function in the patient with Ebstein anomaly, in certain situations CMR may not be performed successfully. Given relatively limited availability and cost, CMR is still considered a restricted imaging modality. In the presence of ongoing irregularities in heart rhythm (such as frequent ectopy or ongoing atrial arrhythmias) image quality may be significantly impaired due to suboptimal cardiac gating resulting in a non-diagnostic examination. CMR is generally contraindicated in pregnancy (particularly with regard to administration of gadolinium and if scanning occurs in the first trimester), in patients with in-dwelling devices (pacemakers and/or defibrillators) and in those who are claustrophobic. Anesthesia is generally required to complete a CMR study in younger children.

4.4 Cardiac Computerized Tomography

Cardiac CT is generally regarded as a partial alternative to CMR. Strengths include rapidity of data acquisition, ability to acquire isotropic data at high spatial resolution (allowing for subsequent off-line analysis), and ability to demonstrate intracardiac shunting following opacification of one side of the

heart (atrium and ventricle) with dense contrast. For the patient with Ebstein anomaly in particular, CT will allow for precise visualization of the tricuspid valve position, morphology of the chordae tendinae and papillary muscles, presence of an intracardiac shunt, and extent of myocardial trabeculation. Limitations in temporal resolution and acute administration of beta-blockade for heart rate modulation to optimize image acquisition may affect accuracy of assessment of ventricular function by cardiac CT. With contemporary dose reduction strategies, cardiac CT is achievable at relatively low radiation exposure (generally in the order of 1–2 mSv) (da Silva et al. 2007; Lange et al. 2015; Li et al. 2016; Ibrahim et al. 2015).

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