Curr Treat Options Cardio Med (2014) 16:338
DOI 10.1007/s11936-014-0338-x

Adult Congenital Heart Disease (A Bhatt and K Niwa, Section Editors)

Ebstein Anomaly: Assessment, Management, and Timing of Intervention

Puneeta Arya, MBBS[1]
Rebecca Beroukhim, MD[1,2,*]

Address
*,[1]Department of Pediatric/Congenital Cardiology, Massachusetts General Hospital, 175 Cambridge Street, 5th floor, Boston, MA 02114, USA
Email: rsberoukhim@mgh.harvard.edu
[2]Deparment of Cardiology, Boston Children's Hospital, Boston, MA, USA

Published online: 22 August 2014

This article is part of the Topical Collection on *Adult Congenital Heart Disease*

Keywords Ebstein · Tricuspid valve · Congenital heart disease · Cyanosis · Adult · Pediatric · Fetal · Arrhythmias · Left ventricular noncompaction · Patent foramen ovale · Atrial septal defect · Echocardiography · Cardiovascular magnetic resonance imaging · Pregnancy

Opinion statement

Ebstein anomaly is a developmental abnormality of the tricuspid valve and right ventricle that results in tricuspid regurgitation and right heart enlargement. Because of the variation in clinical severity and associated findings, patients require a detailed, well-tailored evaluation. For these reasons, management of adults with Ebstein anomaly should take place in a center with expertise in adult congenital heart disease. In many patients, the decision regarding if and when to perform surgery remains controversial, largely because of a lack of published data demonstrating improved postoperative symptoms and survival compared to the natural history of the disease. Because standard two-dimensional echocardiography and cardiovascular magnetic resonance imaging planes do not provide the necessary data to preoperatively manage patients, comprehensive echocardiography and cardiovascular magnetic resonance imaging protocols by experts trained in congenital heart disease are essential in the preoperative management of patients with Ebstein anomaly. As patients may be unaware of their exercise limitations, and for prognostic value, serial cardiopulmonary exercise stress testing is very useful in the evaluation of Ebstein anomaly patients. Surgical tricuspid valve repair historically has not been highly successful because of the marked distortion of tricuspid valve leaflets and right ventricular pathology. Over the last several years, reports of newer surgical techniques to repair the valve, with concurrent advances in arrhythmia management of patients hold promise for improved long term outcomes of patients with Ebstein anomaly. However, because Ebstein anomaly is rare and tricuspid valve repair remains technically challenging, the newer valve repair techniques have not yet gained widespread acceptance throughout the adult congenital heart disease community.

Introduction

In 1866, Dr. Wilhelm Ebstein published the case of a 19-year-old man who died of cyanotic heart disease secondary to a malformation of the tricuspid valve, which ultimately became known as Ebstein anomaly (EA) [1]. Since that time, advances have been made in the diagnosis and management of this unique congenital heart defect.

Epidemiology and genetics

The incidence of Ebstein anomaly is ~1 per 200,000 live births [2, 3]. Although most cases of Ebstein anomaly are sporadic, familial cases have been well described in the literature [4, 5]. Furthermore, mutations in several genes encoding sarcomeric proteins have been identified in association with Ebstein anomaly including cardiac myosin-binding protein C (MYBPC3), α-cardiac actin (ACTC1), cardiac troponin T (TNNT2), α-tropomyosin (TPM1), and cardiac troponin I (TNNI3)[6]. The genetic association with sarcomeric proteins lends credence to the assertion that Ebstein anomaly is a disease of the myocardium as well as valve tissue [7]. More recently, a mutation in the gene encoding β-myosin heavy chain (MYH7) has been described in several families with Ebstein anomaly and left ventricular noncompaction, a well-known association [6, 8]. Given its multifactorial inheritance, the morphology and presentation of this disease is variable and unique to each patient.

Anatomy

Ebstein anomaly is a congenital heart defect characterized by three major pathologic abnormalities of the tricuspid valve that result in tricuspid regurgitation: (1) adhesion of the posterior and septal leaflets to the underlying myocardium with rightward and anterior displacement of the functional annulus (by echocardiography, an exaggerated displacement of the septal leaflet into the ventricular cavity in the apical four-chamber view), (2) redundancy and fenestrations of the anterior tricuspid valve leaflet, and (3) dilatation of the anatomic tricuspid valve annulus at the true atrioventricular junction [9, 10]. The pathologic basis is a failure of 'delamination' of the leaflet tissue from the right ventricular myocardium, a process that involves the valve tissue, annulus, and subvalvular apparatus. The anterior leaflet retains a normal attachment to the native tricuspid annulus. The direction of functional annulus displacement has been previously described as 'downward' and 'apical,' but is more appropriately characterized as 'rightward and anterior,' as there is a rotational displacement toward the right ventricular outflow tract [10]. The anterior leaflet is classically large and irregularly shaped, with short chordal attachments or direct myocardial insertion [9]. There are a host of associated abnormalities, resulting in a broad spectrum of pathologic, echocardiographic, and clinical features (Table 1).

Clinical presentation and physical examination

There is a wide variation in clinical presentation from the fetus to the adult with Ebstein anomaly. The timing of presentation depends on the

Table 1. Morphologic features of Ebstein anomaly and associated abnormalities

Morphologic features of Ebstein anomaly	% of patients	Reference(s)
1. Adhesion of the posterior and septal leaflets to the underlying myocardium with displacement of the hinge point of the septal leaflet into the ventricular cavity, and atrialization of the right ventricle. [a]	100 %	[10]
[By echocardiography, exaggerated displacement of the septal leaflet into the ventricular cavity in the apical four-chamber view.]		
2. Dilatation of the tricuspid valve annulus[a]	100 %	[9]
3. Redundancy ± fenestrations of the anterior tricuspid valve leaflet [a]	100 %	[9]
4. Right ventricular dilatation	66 %	[7]
Associated echocardiographic abnormalities		
1. Varying degrees of tricuspid regurgitation	100 %	[11]
2. Atrial communication (patent foramen ovale or atrial septal defect)	50 % 89 %	[9, 11, 40••]
3. Ventricular septal defect	2 %	[40••]
4. Tricuspid stenosis		
5. Right ventricular outflow tract obstruction (functional or anatomic pulmonary atresia)	6 %	[40••]
6. Mitral valve prolapse	13 %	[9]
7. Bicuspid aortic valve	5 %	[9]
8. Left ventricular abnormalities (noncompaction, systolic and diastolic dysfunction)		
9. Partially anomalous pulmonary venous drainage	2 %	[40••]
Associated electrocardiographic abnormalities		
1. Right bundle branch block	58 % 68 %	[9, 11]
2. First degree heart block	31 %	[9]
3. Pre-excitation	18 % 44 %	[9, 11, 40••]
4. Nonspecific intraventricular conduction delay/block	15 %	[9]

[a] Required for diagnosis.

severity of tricuspid valve leaflet displacement, the quality of prenatal ultrasound screening, and the clinical severity of the disease. Generally speaking, more severe disease results in earlier presentation. Adults will present either with symptomatic arrhythmias, or with evidence of right- or left-sided heart failure. Occasionally, the initial presentation may also be that of sudden cardiac death, which has been attributed to atrial fibrillation with accelerated conduction through an accessory pathway, or from ventricular arrhythmias. On physical examination, patients may have a right ventricular lift and a murmur of tricuspid regurgitation. The first heart sound is widely split, and the tricuspid component is loud and delayed because of closure of the large anterior leaflet. Patients with severe right heart failure may have jugular venous distention. However, the V wave of tricuspid regurgitation seldom appears in the jugular pulse because of the damping effect of the large atrium and the thin walled atrialized right ventricle, even in Ebstein patients with severe tricuspid regurgitation. Forty seven percent of adult patients with Ebstein anomaly have clinical cyanosis because of a combination of

increased right atrial pressure from tricuspid regurgitation and a right-to-left atrial level shunt [11].

Electrocardiography

The electrocardiogram (ECG) is usually abnormal in patients with Ebstein anomaly. P waves are often quite tall and peaked (Himalayan P waves). A qR pattern can be seen in lead V1 to V4. There can be some intraventricular conduction delay with a widened QRS with or without a right bundle branch pattern. Pre-excitation, in the form of Wolff-Parkinson-White syndrome and manifest pre-excitation, may be present in 18 % 44 % of the cases with usually a right sided bypass tract in the right posterior or right posteroseptal region. A fractionated QRS, defined as a conduction delay presumably caused by myocardial scar, is associated with increased atrialized right ventricular volume, lower right ventricular ejection fraction, lower oxygen consumption, lower oxygen saturation, and an increased risk of arrhythmic events in Ebstein anomaly [12••, 13]. Pseudo normalization of the right bundle branch block through a slow accessory pathway can also be seen. Multiple accessory pathways may be present [3, 14–18].

Chest radiography

The chest x-ray can be nearly normal in mild cases. Right atrial enlargement with a globular cardiac contour can be seen severe cases. Cardiomegaly on chest x-ray has been shown to be an independent risk factor for adverse outcome [11].

Echocardiography

Transthoracic echocardiography is the modality of choice for the diagnosis and morphologic evaluation of Ebstein anomaly. The goals of echocardiography are (1) to identify the classic morphologic features of Ebstein anomaly as well as nonessential, but commonly associated features (see Table 1); (2) to evaluate the cardiac physiology including severity of tricuspid regurgitation, ventricular function, and presence or absence of right-to-left atrial level shunting; and (3) to exclude other causes of tricuspid regurgitation (for example dysplastic tricuspid valve, tricuspid valve prolapse, right ventricular dysplasia, endocarditis, and annular dilatation) [19]. The anatomic assessment that is particularly well suited to echocardiography is the evaluation for leaflet dysplasia, including position of valve leaflets, thickening, tethering, and fenestrations [20]. Misunderstanding about tricuspid valve morphology based on two-dimensional (2D) echocardiography is common, partly because only two of the three leaflets are typically seen in standard long axis views resulting in difficulty correctly identifying each leaflet. In addition, tricuspid valve leaflet commissures, coaptation orifice and the en face view of the valve are often difficult to see by 2D imaging [21]. Because the valve is not seen in its entirety in any standard 2D imaging plane, 2D

sweeps are essential to define valve morphology in the parasternal long axis, short axis, and apical views. Good quality 3D echocardiography can add useful information about valve morphology in patients with good imaging windows. In particular, the degree of rotation of the effective tricuspid valve annulus toward the right ventricular outflow tract is well seen by 3D imaging [22]. Paradoxical septal motion, typically seen at the base of the heart, may be related to pressure differences between atrialized right ventricle and adjacent left ventricle, and/or conduction abnormalities. Transesophageal echocardiography is useful for preoperative definition of tricuspid valve anatomy and the atrial septum in patients with poor transthoracic acoustic windows, and also intraoperatively during surgical repair. A proposed transthoracic echocardiography imaging protocol is show in Table 2.

Cardiovascular magnetic resonance imaging

Cardiovascular magnetic resonance (CMR) imaging is an important emerging modality in the pre- and postoperative evaluation of patients with Ebstein anomaly (Fig. 1). CMR adds morphologic and physiologic information that cannot be obtained by echocardiography alone, including evaluation of valve morphology in patients with difficult acoustic windows, accurate evaluation of right atrial and right ventricular volumes, quantification of tricuspid regurgitation fraction, and shunt quantification in patients with an atrial septal defect. The imaging protocol should address the same areas of interest as outlined in the transthoracic echocardiography imaging protocol (Table 2), with additional data on right atrial and ventricular volumes, right ventricular ejection fraction, tricuspid regurgitation fraction, and shunt quantification. CMR imaging also allows for assessment of left ventricular size and function, left ventricular noncompaction, and regions of fibrosis on myocardial delayed enhancement imaging. Chest computerized tomography is an alternative to CMR imaging but has a more limited application because of lower temporal resolution and inability to quantify flow volume.

Exercise stress testing

Patients with congenital heart disease often self-limit and are, therefore, unaware of their exercise limitations. Exercise stress testing offers a reliable tool for assessment of functional capacity. Deterioration in exercise capacity has important prognostic implications [23]. Peak oxygen consumption is significantly reduced even in relatively asymptomatic patients with Ebstein anomaly [24]. Peak VO2 has been found to be the strongest exercise marker of outcome in this population. A predicted peak VO2<60 % is associated with a higher risk of death, non-elective hospitalization and surgical repair [25]. A progressive decline in biventricular and tricuspid valve function, and chronotropic insufficiency are thought to contribute to the time-related decline in exercise function [26]. Right-to-left shunting across a patent foramen ovale or an

Table 2. Transthoracic echocardiographic imaging protocol for Ebstein anomaly of the tricuspid valve

Tricuspid valve morphology	
• Varying degrees of rotation of the valve orifice	Identify septal leaflet displacement in the apical four-chamber view (>8 mm/m^2). Depending on the severity of pathology, inflow may be oriented toward the apex of the RV, or tilted toward the RVOT. [a]
• Anterior leaflet pathology	Assess for elongation, tethering to the RV free wall, and fenestrations. [a]
• Septal and posterior leaflet pathology	Assess for displacement of hinge points or absence of leaflets. [a]
• En face views of the tricuspid valve	Cross-sectional view of valve coaptation and regurgitant orifice.[a]
• Native annular dilatation	Measure native tricuspid valve annulus in the apical four-chamber and parasternal long axis views.
Tricuspid valve function	
• Severity of tricuspid regurgitation	Measure vena contracta in long and short axis views of the valve. [a]
• Mechanism of regurgitation	Identify the location of valve regurgitation and adjacent leaflet/chordal pathology.
• Tricuspid stenosis	Measure mean tricuspid valve inflow gradient.
Right ventricle	
• RV size and function	Imaging of the RV from all standard views with assessment of size and function.
• RV systolic pressure	Measure peak tricuspid regurgitation velocity.
• RVOT	Exclude RVOT obstruction, evaluate for pulmonary valve stenosis and regurgitation.
Right atrium	Measure right atrial dimensions and volume, include atrialized RV.
Inferior vena cava	Assessment of inferior vena cava size and respiratory variation to estimate right atrial pressure.
Atrial septum	Assess for PFO or ASD from various imaging planes, including subxiphoid and right sternal border view (TEE may be indicated). Assess for direction of shunt.
Ventricular septum	Exclude ventricular septal defects with 2D and color Doppler sweeps of the ventricular septum from apical and parasternal short axis views. Assess for paradoxical septal motion of the basal ventricular septum. Assess systolic septal position to estimate RV pressure.
Other	
• Left ventricle	Exclude LV noncompaction, quantitative evaluation of LV size and function.
• Mitral valve	Evaluate for mitral valve prolapse and regurgitation.
• Aortic valve	Exclude bicuspid aortic valve.
• Pulmonary veins	If possible, identify pulmonary venous connections from parasternal or suprasternal views. Sweep the left innominate vein to exclude anomalous pulmonary vein connection.

ASD atrial septal defect, LV left ventricle, PFO patent foramen ovale, RV right ventricle, RVOT right ventricular outflow tract.
Imaging tips: optimize image depth and width, avoid over-zooming on the valve. Optimize color Doppler with particular attention to high frame rate (>20 Hz). Though wide color boxes may capture geographic extent of regurgitation, identification of vena contracta, flow direction and flow velocity becomes limited with low frame rates.
[a]The best 2D imaging of valve morphology and regurgitation may be in the parasternal long, parasternal short, or apical views depending on the severity of valve rotation. Two-dimensional sweeps of the valve are essential in each plane. Depending on acoustic limitations, evaluation may be better seen by transesophageal echocardiography and/or 3D imaging.

atrial septal defect can be exacerbated during exercise. The cyanosis contributes to exercise intolerance through a reduction in arterial oxygen content and increase in physiological dead space [25]. This can manifest as inefficient ventilation, an elevated VE/VCO2, as well as a decrease in peak VO2 [24, 25].

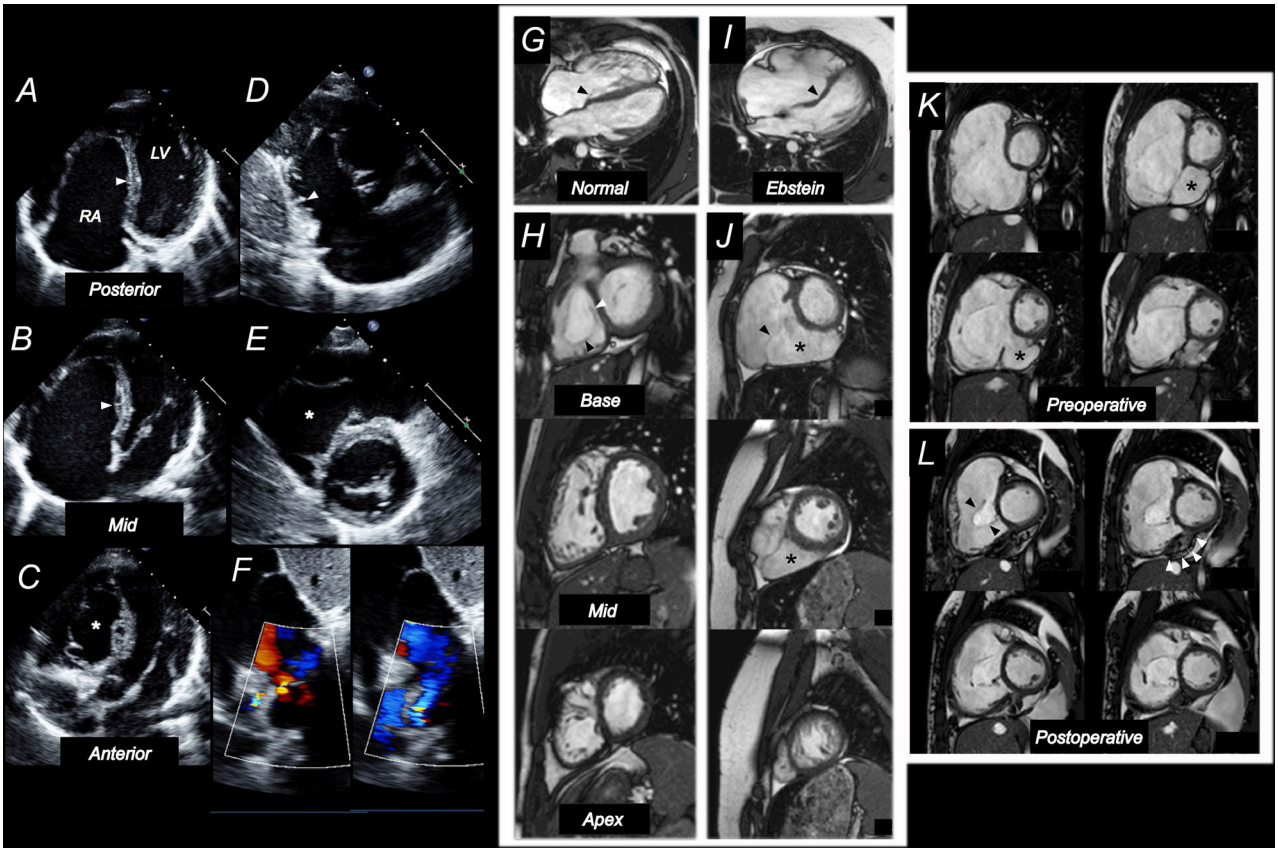

Figure 1. Echocardiogram (A-F) and cardiovascular magnetic resonance (G-L) images of Ebstein anomaly. (A) Apical four-chamber view tilted posteriorly, demonstrates severe tethering of the posterior leaflet of the tricuspid valve (*arrow*) with atrialization of the right ventricle; (B) apical four-chamber view at the midventricular level demonstrates severe tethering of the septal leaflet *(arrow)*; and (C) apical four-chamber view tilted anteriorly demonstrates an en face view of the tricuspid valve, which is oriented toward the right ventricular outflow tract (*); (D) parasternal long axis view tilted toward the right ventricle demonstrates severe tethering of the posterior leaflet (*arrow*); (E) parasternal short axis view demonstrates atrialization of the right ventricle (*) with rotation of the tricuspid valve orifice toward the right ventricular outflow tract; (F) subxiphoid color Doppler imaging of the atrial septum demonstrates bidirectional flow through a patent foramen ovale; (G) CMR of a normal heart in the four-chamber view demonstrates normal attachment of the septal leaflet of the tricuspid valve to the annulus (*arrow*); (H) normal heart in the short axis view at the base demonstrates the normal formation and orientation of the septal (*white arrow*) and posterior (*black arrow*) tricuspid valve leaflets at the base of the heart with inflow directed toward the apex of the heart; (I) CMR of a heart with Ebstein anomaly demonstrates tethering of the septal leaflet (*arrow*), and (J) an elongated anterior leaflet *(arrow)* with atrialization of the right ventricle (*asterisk*) at the basal and mid-ventricular level; (K) preoperative CMR images of a patient with Ebstein anomaly after dehiscence of a previous right ventricular plication (*); (L) postoperative CMR images after Cone reconstruction of the tricuspid valve, resulting in a bileaflet valve (*black arrows*) and tissue thickening along the inferior wall of the right ventricle from a repeat plication (*white arrows*).

Medical management

Medical management of Ebstein anomaly includes noninvasive imaging, evaluation and treatment of occult and symptomatic arrhythmias, and as-

sessment for cyanosis and right heart failure. Patients with progressive right ventricular enlargement and dysfunction, and progressive tricuspid regurgitation may develop signs of right heart failure with peripheral edema and cyanosis. Diuretics may result in reduction of peripheral edema but will not affect the fatigue and dyspnea related to low left sided cardiac output. Anticoagulation with warfarin is recommended for patients with Ebstein anomaly with a history of paradoxical embolus or atrial fibrillation [27]. Newer oral anticoagulants such as rivaroxaban, dabigatran, and apixaban may be considered as alternatives to warfarin, though these have not been directly studied in the Ebstein population [28]. Antibiotic prophylaxis before dental procedures that involve manipulation of gingival tissue or the periapical region of the teeth or perforation of the oral mucosa is reasonable in cyanotic patients with Ebstein anomaly and postoperative patients with a prosthetic cardiac valve. Endocarditis prophylaxis is usually unnecessary in acyanotic, unoperated patients.

Ultimately, all patients with Ebstein anomaly should have a periodic evaluation in a center with expertise in ACHD management [27]. The timing of follow-up at an expert ACHD center depends on the clinical status of the patient. For low risk patients who are asymptomatic and acyanotic with mild tricuspid regurgitation, we suggest follow-up every 2–3 years. For higher risk patients, we suggest follow-up every 6 12 months. The Seattle Heart Failure Model can be used by general cardiologists to identify Ebstein patients at highest risk for adverse outcome, who should be referred to tertiary care centers [29•].

Physical activity

Exercise recommendations vary based on the severity of the disease. Even mild cases may be associated with arrhythmias; and there is an increased risk of sudden death with exercise in severe cases. The exercise recommendations according to the Task Force I report on CHD include the following: (1) athletes with mild Ebstein anomaly, without cyanosis, and with a nearly normal heart size and no evidence of arrhythmia can participate in all sports; (2) athletes with tricuspid regurgitation of moderate severity can participate in low intensity competitive sports if there is no evidence of arrhythmia on ambulatory ECG monitoring; and (3) athletes with severe Ebstein anomaly are precluded from all sports participation. However, after surgical repair, low intensity competitive sports can be permitted if tricuspid regurgitation is absent or mild, heart size on chest radiograph is not substantially increased and arrhythmia is not present on ambulatory ECG monitoring and exercise test [30].

Arrhythmia management

Supraventricular tachycardia secondary to accessory pathways, primary atrial tachycardia and atrioventricular node reentrant tachycardia are often seen in Ebstein anomaly. A high percentage of patients have other arrhythmia substrates including atrial flutter, atriofascicular fibers, and ventricular tachycardia [31]. Management of the tachyarrhythmias can be challenging but

effective control is important for optimal patient outcome. Current data suggests that all Ebstein anomaly patients with symptomatic arrhythmias and those scheduled for surgical repair should undergo pre-operative electrophysiology study regardless of the presence of symptoms [16, 32, 33]. Localization of accessory pathways is often challenging due to a massively enlarged right heart, displaced tricuspid annulus, and distortion of anatomic landmarks, and because over 50 % of patients have multiple accessory pathways [16, 31]. The success rate for catheter ablation is lower in Ebstein anomaly (≤81 %) patients compared with patients with structurally normal hearts (≥95 %) [14, 15, 34]. Rarely, right coronary artery stenosis has been seen following catheter ablation as reported by Bertram et al in two case reports [35]. If catheter ablation is unsuccessful or deemed inappropriate, surgical interruption can be performed. For any patients with history of atrial flutter or fibrillation a Maze procedure can be incorporated into the surgery.

Cardiac catheterization

Adults with Ebstein anomaly have varying degrees of right-to-left shunting across an atrial level defect. Percutaneous closure of atrial septal defects in Ebstein anomaly is indicated in patients with paradoxical embolus (defined as stroke or transient ischemic attack, brain abscess, or myocardial infarction) [36]. Although there is paucity of data, percutaneous closure may reduce cyanosis and improve functional capacity in selected cases [37]. Cardiac catheterization for hemodynamic assessment only is rarely needed. Preoperative coronary artery angiography should be considered if there is a suspicion of coronary artery disease (CAD), in men ≥35 years, in premenopausal women ≥35 years who have CAD risk factors, and postmenopausal women [27].

Surgery

Surgeons with training and expertise in congenital heart disease should be chosen to operate on patients with Ebstein anomaly [27]. The goals of surgery are to improve functional status and reduce the risk of sudden death in patients with Ebstein anomaly. In neonates, surgery should be avoided if possible to allow time for pulmonary vascular resistance to decline and right ventricular output to improve, potentially obviating the need for early surgery. The American College of Cardiology/American Heart Association 2008 guidelines for surgical intervention include (1) symptoms or deteriorating exercise capacity; (2) cyanosis (oxygen saturation less than 90 %); (3) paradoxical embolism; (4) progressive cardiomegaly on chest x-ray; and (5) progressive right ventricular dilation or reduction in right ventricular systolic function [27]. Nevertheless, the decision about whether to perform surgery in individual patients with Ebstein anomaly is often debated because of unpredictable surgical outcomes. Preoperative decision making and planning should involve a team of cardiologists, cardiac surgeons and electrophysiologists when appropriate.

Surgical techniques are tailored to each patient with the goal of improving the severity of tricuspid regurgitation, reducing the arrhythmia burden, closing any inter- atrial communications, and pacemaker placement when necessary. When possible, tricuspid valve repair is the preferred surgical

method. Techniques for tricuspid valve repair have undergone an evolution over the last 30 years, and generally involve reducing the annulus size with or without an annuloplasty ring, repositioning tricuspid valve leaflets to allow better coaptation, reducing the size of the right atrium and atrialized right ventricle, and closing inter atrial communications. Although technically challenging with a steep learning curve, there have been promising results of the cone operation from various institutions worldwide [33, 38, 39, 40••]. The cone operation is a repair technique that aims to create a 'cone' shaped reconstructed leaflet that coapts as a bicuspid valve rather than a monocusp valve as was traditionally performed (Fig. 1) [38].

Surgical options include tricuspid valve repair (cone vs. monocusp repair), bioprosthetic valve replacement, and bidirectional cavopulmonary anastomosis. When possible, valve repair is the preferred surgical method. Factors that favor valve repair include younger age at operation, the absence of massive right ventricular or annular dilatation, the presence of a septal leaflet, and no history of surgical leaflet delamination during a previous procedure [39]. Even though the cone operation is preferable to a monocusp technique, patients with highly muscularized anterior leaflets or completely absent septal leaflets may not be candidates for the cone operation. Many factors in support of valve repair can be determined by preoperative assessment, including a detailed echocardiographic evaluation. However, because the degree of anterior leaflet muscularization and presence of septal leaflet tissue may be difficult to assess by preoperative echocardiography, the final decision regarding surgical technique is made in the operating room. Tricuspid valve replacement is recommended for older patients (>60 years old) and those with pulmonary hypertension [39, 41••]. Rarely, patients with severely enlarged right ventricles, severe right ventricular dysfunction, or low cardiac output syndrome may require bidirectional cavopulmonary anastomosis or the Fontan operation [42]. Risk factors for postoperative death include lack of postoperative improvement, older age at surgery, preoperative left ventricular ejection fraction <50 %, diabetes mellitus, and preoperative history of heart failure [41••].

Prognosis

The prognosis of Ebstein anomaly varies from one patient to another, depending on the morphology of the heart, burden of arrhythmias, and resultant clinical sequelae. In an adult cohort of 72 unoperated patients over the age of 25, survival was 89 % at 1 year, 76 % at 10 year, and 41 % at 20 years of follow-up. Risk factors for cardiac-related adverse outcome include earlier age at diagnosis, increasing degree of cardiomegaly on chest radiography, male gender, increasing severity of tricuspid valve leaflet displacement, increasing tricuspid regurgitation severity, New York Heart Association functional class III or IV, and reduced exercise capacity [11, 25]. In contrast, of a cohort of 81 adult patients aged 50–79 years with Ebstein anomaly who underwent surgical intervention, 20-year survival was 65 % vs 74 % for age- and sex-matched controls. Although postoperative outcomes have improved over time, a lack of published data persists on whether surgical management of adult patients with Ebstein anomaly improves the natural history and mortality of the disease.

Pregnancy

With physiologic changes of pregnancy including increased blood volume and increased blood flow, pregnant women with Ebstein anomaly are at risk for right heart failure, arrhythmias and occasionally sudden death [43, 44]. Following a review of 127 pregnancies in women with Ebstein anomaly, the risk of heart failure was 3.1 % with a 4 % risk of arrhythmias [44]. Thus women with Ebstein anomaly should undergo prepregnancy counseling with a multidisciplinary team with expertise in adult congenital heart disease [27]. Although most women with Ebstein anomaly can have a successful pregnancy with proper care, there is an increased risk of low birth weight and fetal loss if significant cyanosis is present. Asymptomatic women with New York Heart Association class I and no history of arrhythmias or WPW may plan for pregnancy without prior intervention. For patients with a history of arrhythmia, catheter ablation should be considered prior to pregnancy [45]. Surgical tricuspid valve repair or replacement with closure of atrial septal defect should be considered prior to pregnancy in higher risk patients, including those with exercise intolerance, cyanosis, significant tricuspid regurgitation or cardiomegaly on chest radiography [45]. Mechanical tricuspid valve replacement prior to pregnancy is not recommended because of the risk of bleeding during pregnancy [45]. The risk of congenital heart disease in offspring is ~6 % [27]. Therefore, all women with Ebstein anomaly should be offered fetal echocardiography screening in the 18th 22nd week of pregnancy.

Compliance with Ethics Guidelines

Conflict of Interest
Dr. Puneeta Arya and Dr. Rebecca Beroukhim each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent
This article does not contain any studies with human or animal subjects performed by any of the authors.

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- • Of importance
- •• Of outstanding importance

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