

EBSTEIN ANOMALY

Definition

Ebstein anomaly is a malformation of the tricuspid valve, with apical displacement of the septal and posterior leaflets from the atrioventricular ring, resulting in a reduction in size of the right ventricle.¹⁻³ There are no cords to suspend the leaflets, so they arise from the cavity of the ventricle and are attached to its wall. The anomaly results in right ventricular dysplasia with dilatation and dyskinesis of the atrialised right ventricle. Thus, the disease is characterized by:

- adherence of the septal and posterior leaflets to the underlying myocardium
- downward (apical) displacement of the functional annulus
- dilation of the “atrialized” portion of the right ventricle, with various degrees of hypertrophy and thinning of the wall
- redundancy, fenestrations, and tethering of the anterior leaflet
- dilation of the right atrioventricular junction (true tricuspid annulus).
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Epidemiology

It represents 1% of all congenital heart disease. Associated cardiac malformations are ASD (80%), left ventricular fibrosis, and ventricular non-compaction.¹

Presentation

Ebstein anomaly has an extremely variable natural history and prognosis. Adults usually present with exercise intolerance and varying degrees of dyspnoea and cyanosis and/or supraventricular tachycardia. Paradoxical embolization, brain abscess and sudden death may occur. Patients with Ebstein anomaly who reach late adolescence and adulthood often have a good outcome.⁴ Arrhythmias are very common and represent AVRT (6-30% of patients have right lateral or posteroseptal accessory pathways), intra-atrial reentry, atrial flutter, AF, or AVNRT.⁵ Wolff-Parkinson-White syndrome with overt ventricular preexcitation is seen in 60% of patients presenting with arrhythmias. Overall, accessory AV and atriofascicular pathways occur in up to 25% of all patients with Ebstein, and are more often right sided and multiple than in patients without the disorder.^{6,7} LBBB tachycardias can be due to antidromic AVRT or conduction over a bystander accessory pathway. Depending on the severity of the malformation and the arrhythmia, SVT can produce cyanosis and severe symptoms or sudden death due to rapid conduction to the ventricles during AF or atrial flutter when an accessory pathway is present. Ventricular tachycardia may also occur, and focal tachycardia substrates in the atrialized RV are more common than macro-re-entrant substrates.⁸

Hepatorenal dysfunction, as measured using the model for end-stage liver disease excluding international normalized ratio (the MELD-XI score), is a predictor of mortality that complements right atrial dysfunction, hypertension, and low stroke volume, which are markers of hemodynamic dysfunction in patients with Ebstein anomaly.⁹

Physical Examination

S₁ and **S₂** are widely split

S₃ or **S₄** usually present

Systolic murmur of TR

Hepatomegaly due to right heart failure may be present.

Investigations

The **ECG** shows enlarged P waves, prolonged PR interval, and complete or incomplete RBBB. The QRS duration is a marker of RV enlargement and dysfunction. QRS fractionation is associated with a greater atrialized RV volume. A preserved surface ECG identifies a subset of patients with Ebstein anomaly with mild morphological and functional abnormalities and better clinical profile.¹⁰ RBBB is usually present and, in the presence of a right-sided accessory pathway, ventricular pre-excitation can mask the ECG evidence of RBBB. In patients without preexcitation, the absence of RBBB is a strong predictor of accessory pathways.

Chest radiography reveals cardiomegaly in severe cases (Table 1). A cardiothoracic ratio >0.65 carries a poor prognosis.²

Two-dimensional echocardiography is essential to establish the diagnosis and guide management (Figure 1).

3-D echocardiography and magnetic resonance imaging also provide precise anatomical and functional assessment of the tricuspid valves and the right ventricle.

Therapy

Anticoagulation with warfarin is recommended for patients with Ebstein anomaly with a history of paradoxical embolus or AF. Surgery is recommended in symptomatic patients, or in the presence of cyanosis (O₂ saturation < 90%), progressive RV dilatation, severe TR, or paradoxical embolism (Tables 2 and 3).^{11,12} Surgical treatment is now feasible at specializing centers with biventricular or univentricular tricuspid repair or tricuspid valve replacement with a bioprosthesis. Mortality is now less than 2.5% with late death rate over 7 years of 7.6%.^{2,13} Due to the infinite anatomic variability encountered with this anomaly there have been more techniques of tricuspid repair reported in the literature than any other congenital or

acquired cardiac lesion. Reduced right ventricular function continues to be a challenge for some patients, as is the need for reoperation for recurrent tricuspid regurgitation.³ Catheter ablation of accessory pathways is difficult with a 80% success rate and up to 40% recurrence rate,^{5,14} and surgical ablation may be needed to ensure success and to avoid recurrent arrhythmias and instability in the perioperative period. Strategies that included epicardial lead placement, transmyocardial lead implantation, coronary sinus lead implantation, and exteriorization of preexisting transvenous leads, are useful to avoid having a ventricular lead traverse the tricuspid valve.¹⁵ Patients with Ebstein anomaly are at significant risk for sudden death throughout their lifetimes. Pulmonary stenosis, heart failure, and prior syncope or VT are strongly predictive of future SD events, supporting the notion of ventricular arrhythmias being the most likely trigger.¹⁶ In a recent Registry (43% of patients subjected to surgery), patients with a presumed mild Ebstein anatomy have a 35-year cumulative mortality of 11% (vs 4% for the matched control subjects), whereas patients with presumed severe anomaly demonstrated a cumulative mortality of 18% 35 years following diagnosis.¹⁷

Pregnancy

Acyanotic patients with good RV and without cyanosis tolerate pregnancy well although there is an increased risk of RV failure, arrhythmia and paradoxical embolism, premature birth and fetal loss.¹⁸ Symptomatic patients with Ebstein anomaly with saturations <85% and/or heart failure should be advised against pregnancy (Table 4).¹⁹

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