Ebstein Anomaly

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KEYWORDS

• Ebstein anomaly • Congenital heart disease • Arrhythmia • Radiofrequency catheter ablation

KEY POINTS

- Ebstein anomaly is a rare form of congenital heart disease with a uniquely high prevalence of arrhythmias.
- The most prevalent arrhythmia mechanisms are intrinsic to the underlying embryologic defects and may manifest at any stage.
- Current electrophysiological and surgical strategies are well equipped to address these arrhythmia mechanisms, yet despite available technology and a robust understanding of the mechanisms, these cases remain challenging.
- Surgical techniques that render arrhythmia substrates unreachable mandate comprehensive pre-surgical electrophysiological assessment and potential ablation.
- As in all forms of adult congenital heart disease, as the population ages the need to address atrial fibrillation management and risk stratification for sudden cardiac death becomes ever more pertinent.

INTRODUCTION

Ebstein anomaly is a rare variant of congenital heart disease, with a prevalence of 5.2 per 100,000 live births, representing only 1% of all congenital cardiac lesions.[1] Symptoms may occur at any stage from fetal life to old age: early presentation is often driven by more severe tricuspid valve insufficiency and associated structural anomalies, such as functional or anatomic right ventricular outflow tract obstruction,[2] whereas presentation in adult life is often arrhythmic. In a proportion of patients, it may be found incidentally. Historical population analyses found an infant mortality rate of 23.4%,[1] although improved perioperative care and surgical techniques have increased survival significantly.

Ebstein anomaly was eponymously described in 1866 in a 19-year-old patient admitted to Allerheiligen Hospital in Breslau, Germany (now Wroclaw, Poland), where Wilhelm Ebstein was in his early career as an assistant physician having recently qualified from medical school. The patient reported increasing dyspnea and palpitations since childhood, and on examination had "marked jugular venous pulsations synchronous with the heart beat," dullness to percussion across a large area of the chest, and systolic and diastolic murmurs. The patient died a few days later; at autopsy, Ebstein found the right atrium and right ventricle to be extremely dilated and the tricuspid valve profoundly abnormal.[3]

Ebstein anomaly arises from embryologic failure of the posterior and septal tricuspid valve leaflets to delaminate from the underlying myocardium in the inlet portion of the right ventricle (RV), to which they remain fused. This displaces the leaflet attachment (functional annulus) toward the RV apex to varying degrees, creating a thin-walled "atrialized" area within the RV. The posterior and anterior leaflets are typically fused such that they

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cannot be distinguished visually, whereas the septal leaflet is frequently underdeveloped, described by Ebstein as "discoid" or representing a "3 penny piece."[4] The anterior leaflet is enlarged and often fenestrated,[5] with thickened valve leaflets and abnormal chordae and papillary muscles. In severe cases, the posterior and septal leaflets are completely fused to the ventricular myocardium, and associated chordae and papillary muscles may be completely absent,[4] and redundant anterior leaflets prolapse into the RV outflow tract causing functional obstruction. Associated structural lesions may be present, most commonly pulmonary valve stenosis or atresia and atrial septal defects.[2]

The anatomic malformation of the tricuspid valve and the myocardial structural changes result in multiple important electrophysiologic alterations in Ebstein anomaly. The compact atrioventricular (AV) node is frequently displaced inferiorly toward the coronary sinus os, although the penetrating His bundle is normally located within the central fibrous body at the apex of the triangle of Koch.[4] The PR interval may be prolonged due to right atrial dilation and delayed interatrial conduction. Intrinsic AV nodal conduction may be prolonged in a minority.[6] Electrocardiographically, RV conduction delay is common, caused by delayed activation of the atrialized RV and by abnormalities of the His-Purkinje system. Histologically, the right bundle branch may be abnormal and underdeveloped or completely absent, potentially related to malformation of the septal leaflet and medial papillary muscle.[4] The QRS duration has been shown to reflect the degree of RV dilatation and dysfunction. The presence of fractionation, defined as a distinct low-amplitude wave in the terminal QRS or early ST segment, is a marker of a greater right atrialized portion of the ventricle.

PRACTICAL CONSIDERATIONS IN ELECTROPHYSIOLOGICAL PROCEDURES IN EBSTEIN ANOMALY

- Review prior operative reports detailing valve surgery and atrial incisions.
- Vascular occlusion from recurrent access in childhood is common in adult congenital heart disease; ultrasound confirmation of femoral venous patency can be helpful before the case.
- Determine ventricular function and anticipate potential hemodynamic complications to establish periprocedural management strategies.
- Right-to-left shunting across patent foramen ovale or atrial septal defect (ASD) is common and should be assessed before the procedure to determine optimal anticoagulation strategies. Fatal thromboembolic complications have been reported.[7]
- Angiography of the right atrium can provide fluoroscopic delineation of the true annulus, atrialized RV, and displaced valve leaflets (**Fig. 1**). The right coronary artery and the tricuspid valve fat pad also identify the position of the tricuspid annulus.

ARRHYTHMIA MECHANISMS

Accessory Pathway–Mediated Tachycardia

Structural and histologic abnormalities around the right atrioventricular junction precipitate a high prevalence of right-sided accessory pathways (APs). The vast majority of APs are located on the inferior tricuspid valve: posteroseptal approximating the coronary sinus os and posterolateral concordant with the posterior and septal leaflets. A minority of right-sided pathways is located in a

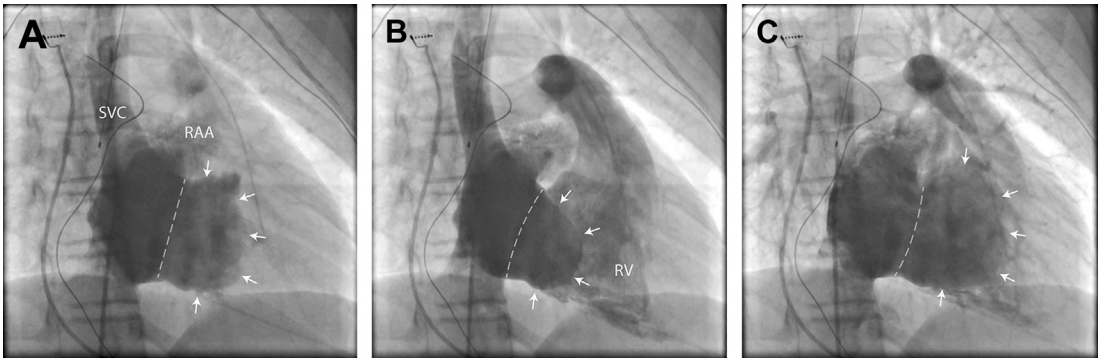

Fig. 1. Angiographic visualization of the tricuspid annulus and valve leaflets. Three fluoroscopic images of the right atrium (RA) and RV from the right anterior oblique view during angiographic injection in the RA. The images captured during ventricular diastole (*A*, *C*) and systole (*B*) demonstrate the position of the true tricuspid annulus (*dashed line*) and displaced valve leaflets (*white arrows*) separated by the atrialized right ventricle (ARV). RAA, right atrial appendage.

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