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Ebstein Anomaly: An Overview for Nursing

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Ebstein anomaly is a rare congenital heart defect. Many nurses have probably never encountered this anomaly, with very few able to accurately depict the pathological anatomy of the condition. As technology further develops, providers are better equipped to recognize and manage Ebstein anomaly. There are important considerations for nurses when caring for an individual with Ebstein anomaly. The aim of this article is to give an overview of the condition exploring the pathophysiology, how patients typically present, and how to effectively care for a patient with Ebstein anomaly regarding medical and surgical courses of treatment. It is important for nurses to have a resource to reference on Ebstein anomaly, and the majority of current literature is solely based for medical providers. Furthermore, Ebstein patients may be seen on a variety of units in the hospital beyond cardiology (i.e., pregnant patient with a diagnosis of Ebstein anomaly).

Overview

IN 1866, WILHELM Ebstein first diagnosed and described what would come to be called *Ebstein anomaly*: a rare congenital heart abnormality with displacement and malformation of the tricuspid valve accompanied by an enlarged right atrium and a dilated, thinned right ventricle. The variability of this congenital heart defect demands that nurses be able to recognize common signs and symptoms of Ebstein anomaly to appropriately anticipate interventions and coordinate care.

According to Agarwala, Hijazi, and Dearani (2012), the estimated risk of Ebstein anomaly is 1 in 20,000 live births and occurs equally in males and females. The risk is higher in infants of mothers who took lithium during early pregnancy. There are recent studies suggesting that there may be a genetic component to Ebstein anomaly, although this correlation is still premature and requires further inquiry and research. Each patient with Ebstein anomaly differs, with varied degrees of severity and symptomatology. The individual with Ebstein anomaly often has an enlarged and incompetent tricuspid valve, creating retrograde blood flow from the right ventricle into the right atrium. The tricuspid valve is misplaced extending downward into the right ventricle; therefore, resulting in a portion of the right ventricle becoming atrialized (Brown & Dearani, 2012). Ebstein anomaly can be classified as mild, moderate, or severe depending on the extent of displacement of the valve leaflets and the degree of tricuspid regurgitation and resultant right chamber enlargement and dysfunction. Other cardiac defects that can be associated with Ebstein anomaly are atrial septal defect (ASD), ventricular septal defect (VSD), pulmonary outflow obstruction, patent

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Table 1 Signs, symptoms and possible diagnoses.

Common signs and symptoms:	Common presentations by age
• Fatigue • Decreased activity tolerance • Shortness of breath with activity • Cyanosis • Palpitations (i.e., paroxysmal atrial arrhythmias, premature ventricular beats) • Heart murmurs • Peripheral edema *Brown and Dearani (2012)	• Neonates: Cyanosis • Infants: Heart failure • Children: Incidental murmurs • Adolescents/Adults: Arrhythmias *Negoi et al. (2013)

ductus arteriosus (PDA), coarctation of the aorta, mitral valve prolapse, bicuspid aortic valve and left ventricular noncompaction (Brown & Dearani, 2012). Arrhythmias are very common with Ebstein anomaly and are usually caused by the atrial dilation. About 15% of Ebstein patients will have additional accessory pathways, usually associated with Wolff–Parkinson–White syndrome. Furthermore, Ebstein anomaly patients in end-stage heart failure are at high risk for ventricular arrhythmias (Brown & Dearani, 2012) (Table 1).

Echocardiography is the standard for establishing a diagnosis of Ebstein anomaly, but chest x-ray is often used in adjunct during the diagnostic phase. Chest x-ray findings commonly include right-sided cardiomegaly. A frontal view chest x-ray may reveal a globular or box shaped heart due to right atrial enlargement and pulmonary trunk hypoplasia. The chest x-ray images will vary in accordance with the individual's severity of the disease (Ferguson, Krishnamurthy, & Oldham, 2007).

Ebstein anomaly is often diagnosed in the newborn period; however, based on the severity of the malformation this defect may not be discovered until adolescence or even adulthood. The Great Ormond Street Ebstein Score (GOSE score) can be calculated during echocardiography to determine the severity of the anomaly and to gather information to determine potential courses of treatment. The GOSE score is calculated in the four-chamber view of echocardiography, comparing the area of the right atrium and atrialized right ventricle to the functional right ventricle, left atrium, and left ventricle (Brown & Dearani, 2012). Another method of classification of Ebstein anomaly is based upon the Carpentier classification. The Carpentier classification system categorizes severity into types I–IV as illustrated in Table 2.

Patients with Ebstein anomaly typically will have an abnormal electrocardiogram with the possible presence of tall and wide P waves formed by the right atrial enlargement. Patients may also have a complete or incomplete right bundle–branch block. Although complete heart block is rare, there is a significant incidence of first-degree atrioventricular block in Ebstein patients again due to the right atrial enlargement and abnormalities in the atrioventricular conduction system (Attenhofer Jost, Connolly, Dearani, Edwards, & Danielson, 2007).

Nursing Interventions in a Medically Managed Patient

The parent or patient (pending age of the child) who is being managed medically should be instructed by the nurse to report signs and symptoms of heart failure such as a new onset of shortness of breath, increased swelling, weight gain, and/or decreased tolerance to activity. Special consideration should be paid during physical assessments to signs and symptoms of fluid overload including, but not limited to, weight gain, periorbital edema in morning or after waking up from naps, audible crackles with auscultation, jugular venous distention, extra heart sounds (S3, S4, murmur), bounding pulses, and restlessness. Nursing considerations for

Table 2 Carpentier classification of Ebstein anomaly.

- Type I (least severe): The anterior tricuspid leaflet is larger and mobile, but the posterior and septal leaflets are apically displaced, dysplastic, or absent. The atrialized ventricular chamber size varies from relatively small to large.
- Type II: The anterior, posterior, and often septal leaflets are present, but are relatively small and displaced in a spiral fashion toward the apex. The atrialized ventricular chamber is moderately large.
- Type III: The anterior leaflet has restricted motion with shortened, fused, and tethered chordae. Direct insertion of papillary muscles into the anterior leaflet is frequently present. The posterior and septal leaflets are displaced, dysplastic, and usually not reconstructable. The atrialized ventricular chamber is large.
- Type IV (most severe): The anterior leaflet is severely deformed and displaced into the right ventricular outflow tract. There may be few or no chordae, and direct insertions of the papillary muscles into the leading edge of the valve are common. The posterior leaflet is typically dysplastic or absent, and the septal leaflet is represented by a ridge of fibrous material descending apically from the membranous septum. Tricuspid valve tissue is displaced into the right ventricular outflow tract and may cause obstruction of blood flow (functional tricuspid stenosis). Nearly all of the right ventricular cavity is atrialized ventricle.

(Agarwala et al., 2012, Morphology section, para. 3)