



## EBSTEIN ANOMALY

### Background

Ebstein anomaly is characterized by malformation and dysplasia of the tricuspid valve. It typically consists of apical displacement of the septal and posterior leaflets of the tricuspid valve. There is also an excessively large antero-superior leaflet. Because of the valve malformations, there are variable degrees of tricuspid regurgitation. In addition, a portion of the ventricle becomes thin-walled and “atrialized”. In childhood, the amount of right ventricle present below the tricuspid valve (“functional” right ventricle” can be small, but sometimes increases in size as a response to chronic volume overload. Atrial septal defect or patent foramen ovale is seen in at least 1/3 of cases.

Ten to twenty-five percent of patients have associated accessory pathways (Wolff-Parkinson-White syndrome). Other associated anomalies include: right ventricular outflow tract obstruction including pulmonary stenosis and atresia, ventricular septal defect, congenitally corrected transposition of the great arteries, tetralogy of Fallot, and non-compaction of the left ventricle.

The clinical spectrum of Ebstein disease depends largely on the degree of leaflet malformation, the severity of tricuspid regurgitation, the size and function of the “functional” right ventricle, and associated lesions.

Clinical manifestations of Ebstein anomaly are variable. Some adults are completely asymptomatic, whereas those with severe forms of Ebstein anomaly may not survive into adulthood. Common cardiac complications consist of supraventricular arrhythmias, right heart failure, and cyanosis secondary to right to left shunting in the presence of an atrial septal defect.

### Effects of pregnancy-related hemodynamic changes

Pregnancy is associated with significant hemodynamic changes including an increase in cardiac output (see Cardiovascular Changes During Pregnancy). The ability of women with Ebstein anomaly to tolerate these changes depends on the right ventricular size and function and the degree of tricuspid regurgitation. In women with limited right heart reserve, the increased volume from pregnancy can result in right heart failure. The fall in peripheral vascular resistance that occurs during pregnancy can augment right-to-left shunting in women with atrial/ventricular septal defects, worsening maternal cyanosis and hypoxemia.

### Maternal complications

In the absence of significant cyanosis, arrhythmia, and/or right ventricular dysfunction, pregnancy is usually well tolerated. The most common adverse maternal cardiac events are supraventricular arrhythmia (4%) and heart failure (3%). More rarely, women experience worsening cyanosis and hypoxia (1-6). There are also other cardiac characteristics that can have an impact on outcomes (see General Considerations). Maternal deaths have not been described.

For women who have had valve repair or replacement (typically a prosthetic tricuspid valve), the risk associated with pregnancy will depend on the status of the prosthetic valve, the size and function of the “functional” right ventricle and the individual’s propensity to arrhythmias.

### **Fetal complications**

Premature delivery and small-for-gestational-age babies occur in 22% and 12% of pregnancy respectively. (6) Three fetal deaths have been described. Women with cyanosis are at increased risk of miscarriage/fetal loss and premature delivery.

### **Management strategies**

#### **Preconception counseling/Contraceptive methods**

Successful pregnancy can be achieved in many women with Ebstein disease, and the maternal complications both during pregnancy and peripartum are usually manageable. Women with cyanosis should be evaluated by a congenital heart specialist prior to pregnancy for the feasibility of repair.

Ideally, a comprehensive cardiovascular examination should be undertaken before embarking on pregnancy. This includes a careful history and physical examination with oxygen saturation measurement, an echocardiogram, and an electrocardiogram. The additional prognostic benefit of cardiopulmonary exercise testing has not been defined, but can be performed in order to assess the women’s functional status and her ability to increase heart rate during exercise. Cardiac magnetic resonance imaging may be useful for assessment of right ventricular function.

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5%, compared to a background risk of approximately 1% of having a baby with congenital heart disease.

A discussion about contraceptive methods is appropriate in all women with Ebstein anomaly. Estrogen-containing oral contraceptive methods are associated with an increased risk of thromboembolism and should be used with caution in women with atrial arrhythmias, atrial septal defects, or cyanosis. Progesterone-only forms of contraception are not associated with thromboembolic risk and can be suitable alternatives (see Contraception).

Medications use should be reviewed if a woman is contemplating pregnancy or is pregnant. The MOTHERISK website (<http://www.motherisk.org>) is an excellent resource.

#### **Ante-partum care**

Coordinated care between a congenital heart disease specialist and a high-risk obstetrician at a high-risk pregnancy center should be implemented. The frequency of follow up visits is dictated by women’s functional status and the residual lesions at the time of conception.

For women with preserved ventricular function, minimal tricuspid regurgitation, no history of arrhythmias, and no cyanosis, the risk of adverse events during pregnancy is low. For women with right ventricular dysfunction or cyanosis, close follow-up by a dedicated multidisciplinary team of experienced cardiologists, high-risk obstetricians, and anesthetists is important.

Treatment for symptomatic heart failure may be necessary in some women with significant right ventricular dysfunction. Volume overload at the time of labour and delivery should also be avoided in women with right ventricular dysfunction as it can result in heart failure.

Supraventricular arrhythmia can be treated medically or with DC cardioversion when pregnant women are unstable or unresponsive to medical therapy (see Arrhythmias).

All women should be offered dedicated fetal echocardiography at 20 weeks gestation.

### **Labour and delivery**

Labour and delivery should be planned carefully with a multidisciplinary team well in advance. It is important to communicate the delivery plan to the woman and to other physicians involved in her care. The best delivery plan is not useful if information is not readily available when needed.

Generally, vaginal deliveries are recommended unless there are obstetric indications for a cesarean delivery. Good pain management for labour and delivery is very important in order to minimize maternal cardiac stress. To decrease maternal expulsive efforts during the second stage of labour, forceps or vacuum delivery is often utilized. To decrease potential harmful complications from difficult mid-cavity assisted delivery, uterine contractions are often utilized to facilitate the initial descent of the presenting part.

The need for maternal monitoring at the time of labour and delivery is dictated by the women's functional status, the degree of ventricular dysfunction, and the presence of cyanosis. Most women do not require invasive monitoring. To detect potential arrhythmias early, continuous monitoring with electrocardiography may be helpful in some instances. Continuous oximetry should be employed in women with cyanosis.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with Ebstein anomaly. However, some experts continue to administer antibiotics because they feel that the risks of adverse reactions to antibiotics are small and the risk of developing endocarditis has major health consequences.

For women with residual interatrial (ASD or PFO) or interventricular shunts (VSD), air-particulate filters (bubble trap filters) are recommended for all intravenous lines at the time of labour and delivery.

### **Post-partum care**

The hemodynamic changes of pregnancy may take up to six months to normalize. Women should be seen early after pregnancy (usually within 6-8 weeks). The frequency of additional follow up visits should be dictated by the clinical status of the women.

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