



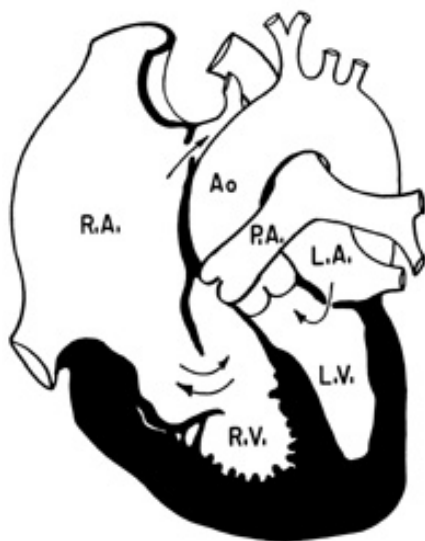
# HOLY C.O.W.!

## IT'S...

Clinical Question of the Week #24  
December 8th, 2008 through December  
15th, 2008

Please e-mail your answers to Kuo, Tim, Wendy, and Kevin ([klian@mednet.ucla.edu](mailto:klian@mednet.ucla.edu); [tprovias@mednet.ucla.edu](mailto:tprovias@mednet.ucla.edu); [wsimon@mednet.ucla.edu](mailto:wsimon@mednet.ucla.edu); [kbreger@mednet.ucla.edu](mailto:kbreger@mednet.ucla.edu)) by 0800 on Monday, December 15th, 2008. The resident or intern with the most correct answers at the end of each month will receive a prize!

**Case:** A 35-year-old woman presents to establish primary care in the Internal Medicine Suite. She has a history of bipolar disorder that has been well controlled on medication and otherwise has been healthy to her knowledge. Her other past medical history is significant only for appendectomy at the age 14. She is married and has one three-year-old child, who accompanies her today. Family history is notable for history of depression in her mother and uncle. Her physical exam is unremarkable. She asks if you could take a listen to her son, as he was recently noted to have an incidental murmur. He underwent some sort of ultrasound tests, but she didn't quite understand what all the pediatric doctors were talking about. She brings this picture that the specialist drew (below), and asks if you might be able to explain.



### Questions:

#### 1. What is the diagnosis?

Ebstein's anomaly is a congenital malformation of the right ventricle and tricuspid valve. It occurs in somewhere around 1:50,000-200,000 individuals, and has been associated with an increased risk among children born to mothers treated with lithium. The anomaly is classified into three grades of severity (mild, moderate, severe) based on the degree of valve leaflet displacement and subsequent tricuspid regurgitation resulting in right sided

chamber dilation and dysfunction. There is also a more detailed staging system (I-IV) based on anatomic findings at surgery.

Clinical presentation of the anomaly can be highly variable, ranging from asymptomatic with normal life expectancy to severe heart failure shortly after birth. Diagnosis may occur as a fetus (abnormal prenatal scan), neonate (cyanosis), infant (heart failure), children (incidental murmur), and adolescent-adult age (arrhythmia). Symptoms include those of early cyanosis and heart failure in severe cases to exertional dyspnea, fatigue, cyanosis, and palpitations/tachyarrhythmias. Physical examination is similarly variable but may reveal widely split S1 and S2 (representative of RBBB) as well as findings of tricuspid regurgitation (prominent v wave on neck exam, systolic murmur). (0.5)

\*\*The disorder is named after Wilhelm Ebstein (1836-1912), a noted German physician who was a professor at Göttingen University and a specialist in malassimilation and defective nutrition, who in his time pioneered a version of an extreme low-carb diet for patients with abnormal nutrition.

**2. Name the two primary abnormalities of this condition.**

The two primary anatomic abnormalities of Ebstein's anomaly include malformation of the tricuspid valve annulus or leaflets and atrialized right ventricle. In particular, the posterior and septal leaflets of the tricuspid valve may be small or absent, and are displaced downward into the body of the right ventricle. This results in a large portion of the right ventricle essentially residing in the right atrium and a small distal proper ventricular chamber which may only consist of the right ventricular outflow tract.

Other cardiac anomalies associated with Ebstein's anomaly include defects in the interatrial septum, VSD with or without pulmonary atresia, pulmonary outflow obstruction, patent ductus arteriosus, coarctation of the aorta, one or more accessory pathways resulting in increased risk of arrhythmia, and various left heart lesions (bicuspid aortic valve, systolic/diastolic dysfunction, MVP, left ventricular noncompaction). (1)

**3. Name two EKG findings associated with this condition.**

Electrocardiogram changes found in Ebstein's anomaly include right bundle branch block with right atrial anomaly, WPW pattern with left bundle branch pattern right prechordal predominant S waves, low right sided voltage on chest leads, other atrial or supraventricular tachycardia, and 1<sup>st</sup> degree AV block. (1)

**4. What are the indications for surgery? What surgical techniques are used?**

Medical management is as follows: 1) cyanotic newborns – supportive care is given until pulmonary vascular resistance drops; 2) infants and children with heart failure – inotropic agents, digoxin, lasix; 3) arrhythmias – antiarrhythmic drugs and/or catheter-based ablation.

Indications for surgery according to the ACC/AHA include severe tricuspid regurgitation and heart failure, decreasing exercise tolerance, the presences of TR with atrial fibrillation, and favorable anatomic orientation of the tricuspid valve. The Danielson repair includes plicating the atrialized portion of the ventricle with multiple commissuroplasty stitches. The Carpentier repair involves detaching the sail-like anterior leaflet from the tricuspid valve annulus and translocating it in order to create essentially a one-leaflet valve in addition to the plication of the right ventricle. In neonates with pulmonary atresia or RVOT obstruction, a Blalock-Taussig shunt is performed prior to the repair. Tricuspid bioprosthetic valve replacement is performed in cases where repair is not possible. (0.5)