RADY 403 Case Presentation

Justin Hills, MS3
September 2018

Ed. by John Lilly, MD

UNC SCHOOL OF MEDICINE Radiology

Focused patient history and workup

- Ex-32w1d F neonate delivered via C-Section due to reversal of umbilical artery end-diastolic flow (REDF)
- Cyanosis: peripheral > central
- Moderate chest retractions
- Holosystolic blowing murmur obscuring S1 and S2
- CPAP +5 was applied for respiratory support given retractions
- Transferred to NCCC for further management with plan to intubate depending on saturations and work of breathing

DDx

Cardiovascular

- Pulmonary atresia with intact ventricular septum PAIVS
- Ebstein's anomaly
- Truncus arteriosus
- Transposition of great vessels
- Tricuspid atresia
- Tetralogy of Fallot
- TAPVR
- Pericardial effusion

Respiratory

- Neonatal respiratory distress syndrome

Infectious

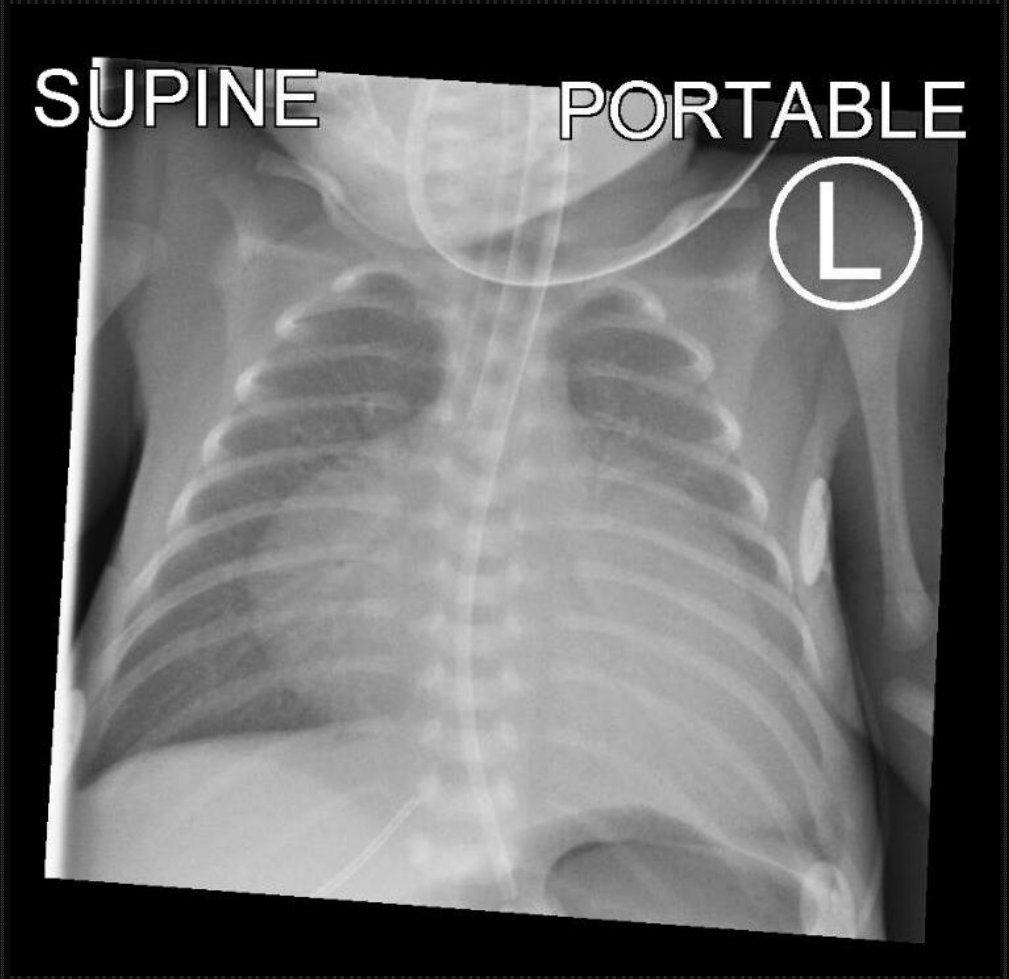
- Infectious myocarditis
- Congenital rubella

List of imaging studies

- XR Chest Portable
- Echocardiogram

XR Chest Portable

AIRWAYS
- Midline trachea
- Visible carina
- (R) w/o atelectasis
- (L) grossly obscured

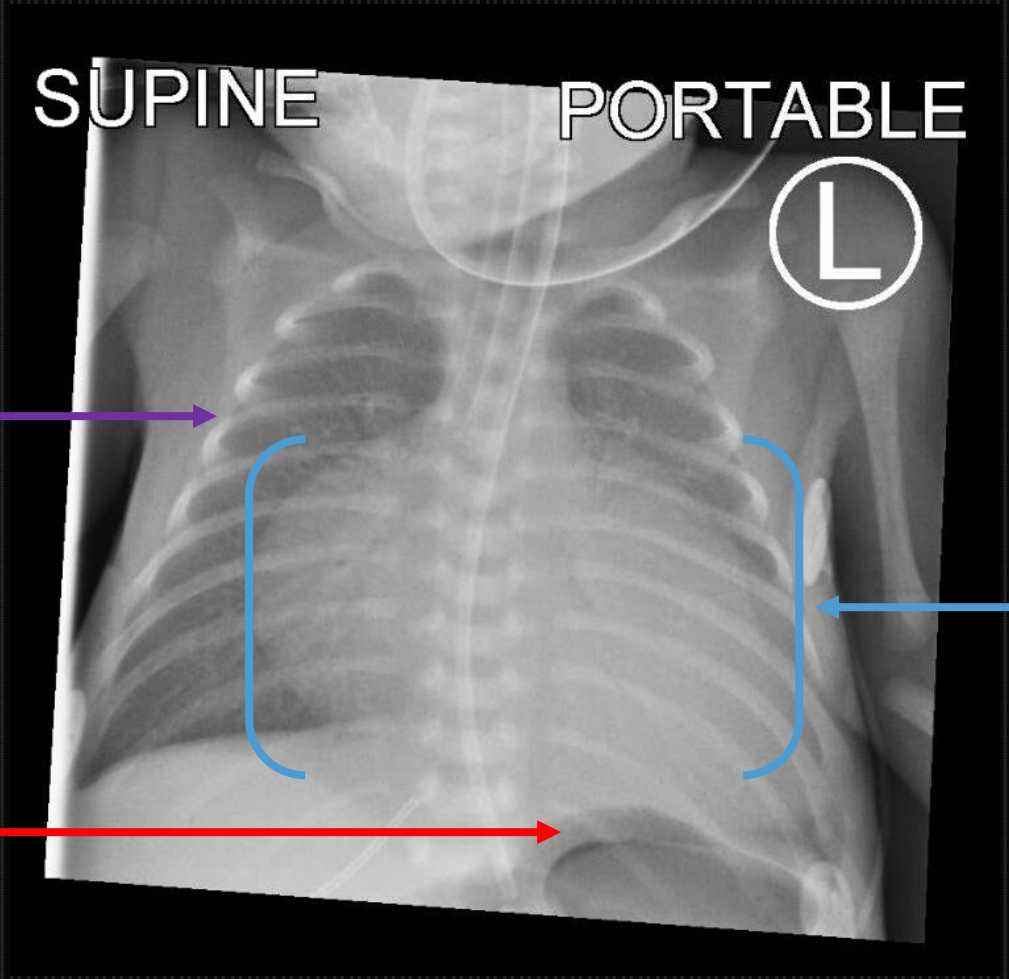


BONES
- No acute osseous abnormalities

XR Chest Portable

EFFUSIONS
- No effusions or pneumothoraces

DIAPHRAGM
- Mild lung hyperinflation
- Gastric bubble present

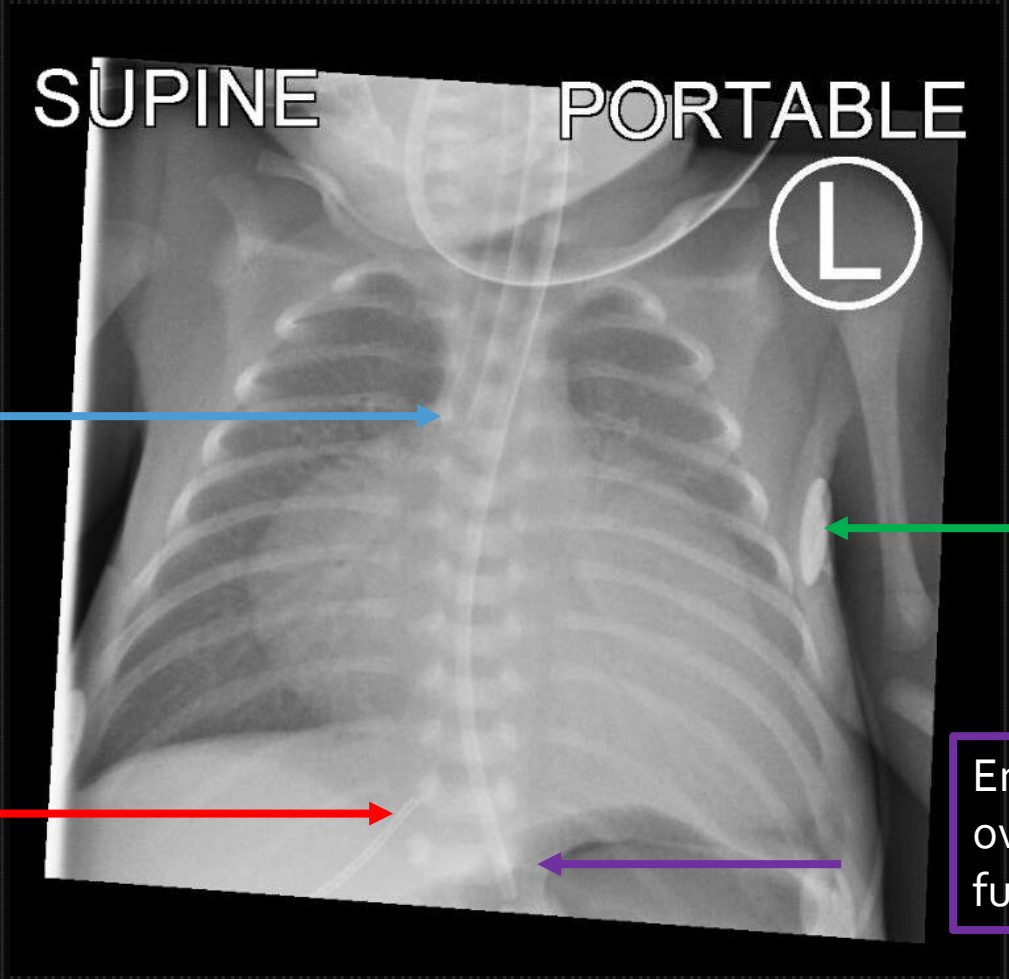


CARDIAC SILHOUETTE AND MEDIASTINUM
- Cardiomegaly
- "Wall-to-Wall" heart

XR Chest Portable

LUNG FIELDS
- No focal opacities
- Sparse vasculature

Endotracheal tube

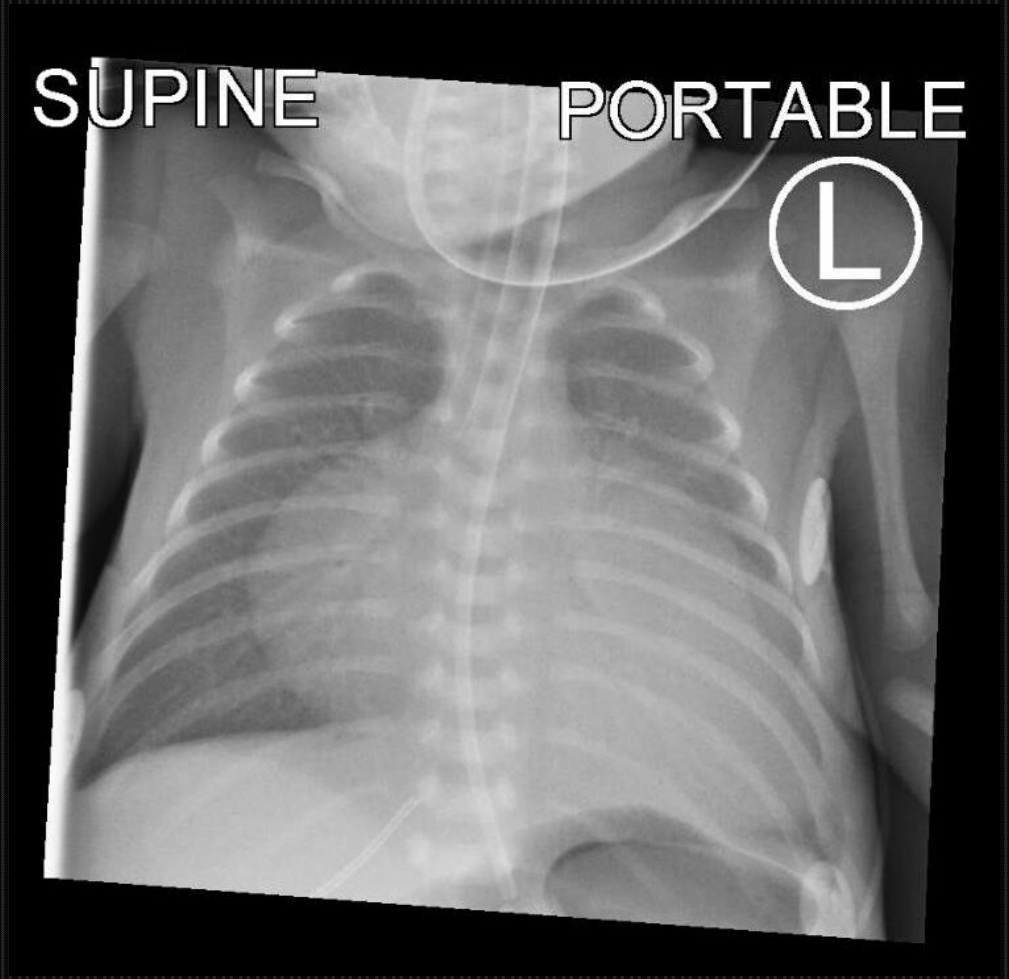


EKG leads

Umbilical Venous Catheter

Enteric tube with tip overlying the gastric fundus

XR Chest Portable



IMPRESSION:

1. Massive cardiomegaly
2. Endotracheal tube terminating approximately 0.5 cm above the carina, consider slight retraction

Echocardiogram

PATIENT FINDINGS

1. Severe dilation of right atrium
2. Severe apical displacement of the tricuspid valve leaflets into RV
3. Moderate to severe tricuspid valve regurgitation
4. "Atrialized," hypoplastic RV
5. Patent foramen ovale with a moderate-size right-to-left shunt
6. Tortuous patent ductus arteriosus, left to right shunt, large and unrestrictive

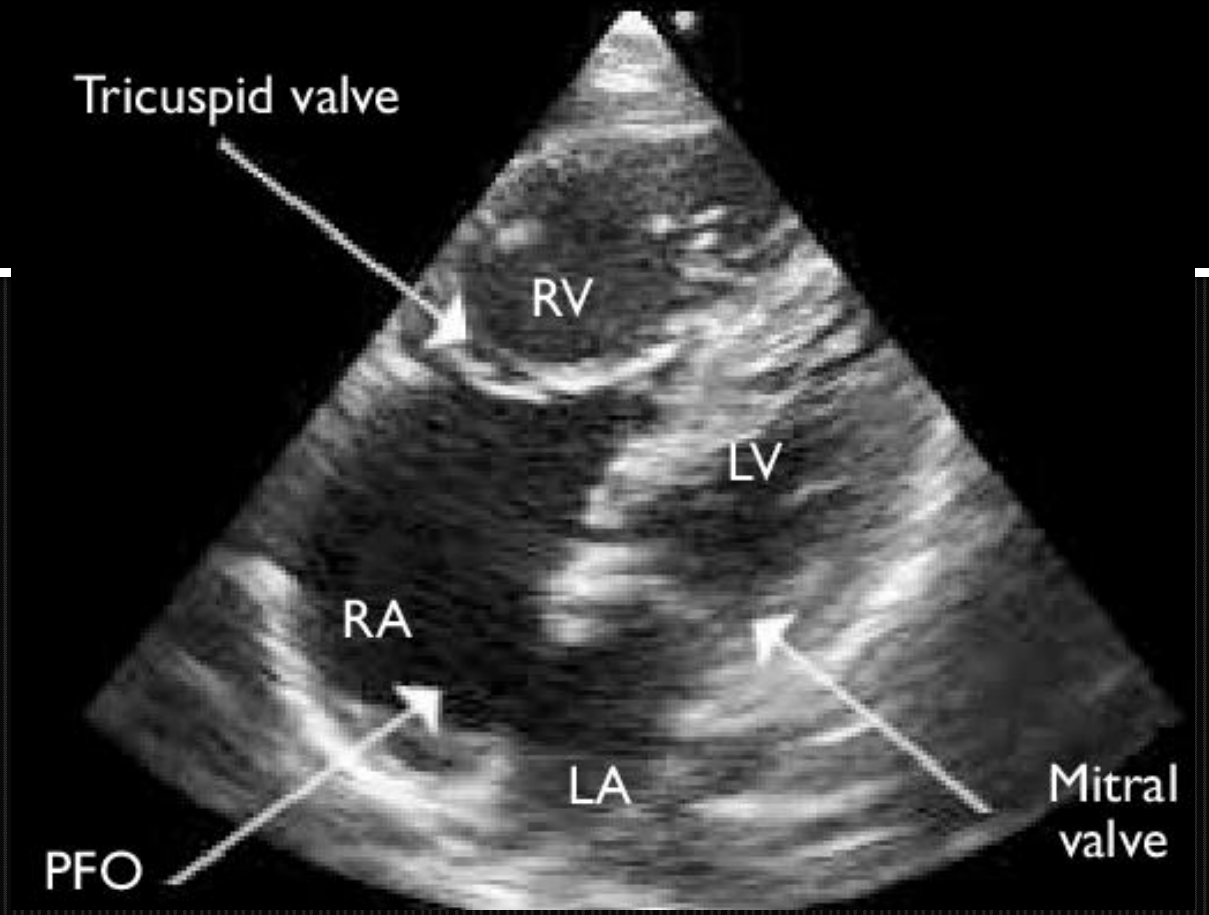


IMAGE CAPTION: Apical 4 chamber, 2 -dimensional echocardiogram shows Ebstein's anomaly of the tricuspid valve. There is displacement of the tricuspid valve toward the apex of the right ventricle (RV) and a patent foramen ovale (PFO). LA: left atrium; LV: left ventricle; RA: right atrium.

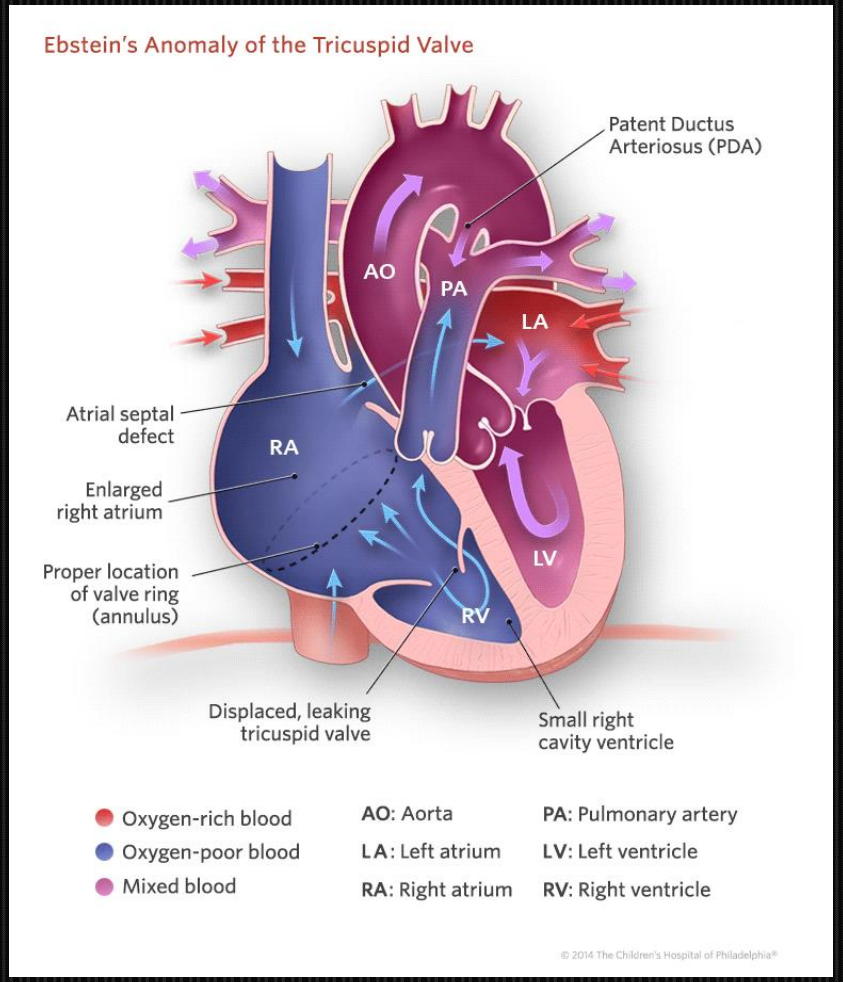
Athappan, G & Chengat, Vipindas & Unnikrishnan, A & Chandraprakasam, Satish & Kumar, S & Ganesh, Nanda. (2009). Aortic dissection presenting as posterior circulation stroke. Singapore medical journal. 50. e35-8.

Patient treatment and outcome

- Patient was intubated and remained in NCCC
- Developed bradycardia with poor perfusion and pulseless electrical activity
- CXR maintained baseline enlarged cardiac silhouette without pulmonary effusions or pneumothoraces
- Patient expired
- Primary Cause of Death: Ebstein's anomaly
- Secondary Cause of Death: Cardiac arrhythmia

Discussion: Ebstein's anomaly

- Congenital malformation
- Displacement of malformed tricuspid valve leaflets into the RV
- Enlarged RA
- "Atrialized" hypoplastic RV
- Moderate to severe tricuspid regurgitation



https://www.chop.edu/conditions-diseases/ebstein-s-anomaly-tricuspid-valve

Discussion: Ebstein's anomaly

- Unknown etiology
- Estimated risk in the general population is 1 in 20,000 live births with no gender predilection
- Associated with maternal lithium use during the first trimester of pregnancy
- Clinical presentation varies widely, ranging from critically ill neonates to asymptomatic adults

Discussion: Ebstein's anomaly

- Routine Imaging of Asymptomatic Children and Adults
  - ECG, Doppler transthoracic Echo, Cardiac MRI
- Medical Management of Symptomatic Children
  - **Alprostadil** (prostaglandin E1) to keep the ductus arteriosus open and improve oxygenation
  - **Nitric oxide** to decrease PVR and improve antegrade blood flow through the pulmonary artery
  - **Milrinone** for its inotropic and pulmonary vasodilator effects
- Surgical Intervention
  - Tricuspid valvuloplasty (widening/ballooning) or replacement, selective plication of the atrialized right ventricle (folding it back to expand the LV), closure of intracardiac shunts (i.e. ASD), right reduction atrioplasty, and any indicated arrhythmia procedures

Discussion: Ebstein's anomaly

- Arrhythmias are amongst the most feared complications for patients with Ebstein's anomaly
- In these patients, congenital and acquired substrates for arrhythmias coexist and can manifest at different disease stages
- Many arrhythmias are based on accessory pathways located along the anomalous atrioventricular valve (TV)
- Associated arrhythmias may include atrial ectopic tachycardia (AET), atrial flutter, atrial reentry tachycardia (ART), atrial fibrillation, WPW and ventricular tachyarrhythmia

UNC Top Three

1. Ebstein's anomaly is a congenital malformation characterized by malformed and displaced tricuspid valve leaflets and a "wall-to-wall" heart on chest radiograph
2. Although many patients with Ebstein's anomaly are asymptomatic, surgical and/or medical intervention are critical if symptoms develop
3. Cardiac arrhythmias are amongst the most feared complications for patients with Ebstein's anomaly

References

Aly, Hany. Respiratory Disorders in the Newborn. Pediatrics in Review Jun 2004, 25 (6) 201-208; doi: 10.1542/pir.25-6-201.

Athappan, G & Chengat, Vipindas & Unnikrishnan, A & Chandraprakasam, Satish & Kumar, S & Ganesh, Nanda. (2009). Aortic dissection presenting as posterior circulation stroke. Singapore medical journal. 50. e35-8.

Celermajer DS, Cullen S, Sullivan ID, et al. Outcome in neonates with Ebstein's anomaly. J Am Coll Cardiol 1992; 19:1041.

Correa-Villaseñor A, Ferencz C, Neill CA, et al. Ebstein's malformation of the tricuspid valve: genetic and environmental factors. The Baltimore-Washington Infant Study Group. Teratology 1994; 50:137.

Dearani JA, Mora BN, Nelson TJ, et al. Ebstein anomaly review: what's now, what's next? Expert Rev Cardiovasc Ther 2015; 13:1101.

Foresti S., Lupo P., Cappato R. (2014) Ebstein's Anomaly and Arrhythmia Management. In: Giamberti A., Chessa M. (eds) The Tricuspid Valve in Congenital Heart Disease. Springer, Milano

Knott-Craig CJ, Goldberg SP, Overholt ED, et al. Repair of neonates and young infants with Ebstein's anomaly and related disorders. Ann Thorac Surg 2007; 84:587.