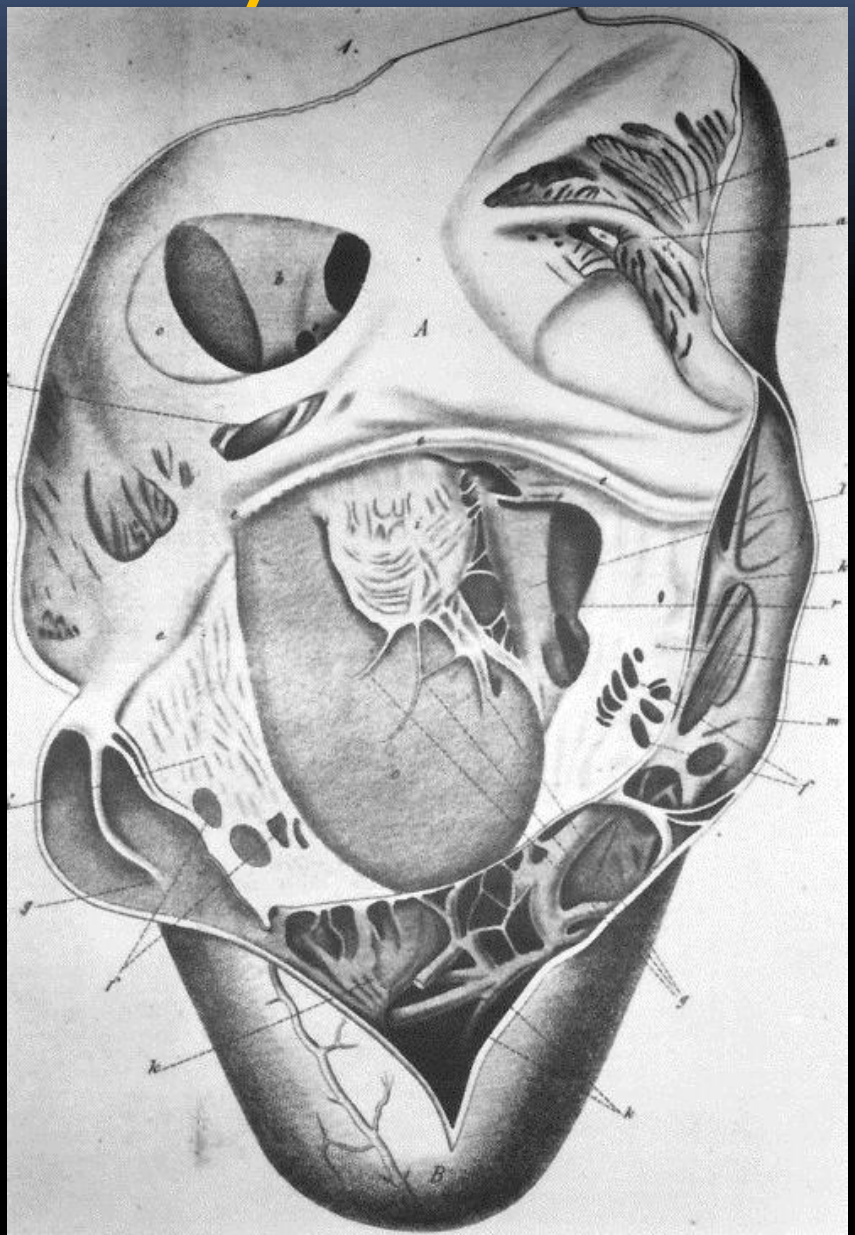
Ebstein Anomaly
Spectrum of Disease and the Role of Imaging Modalities

Department of Pediatrics
Chonnam National University Medical School
Young Kuk Cho

Overview

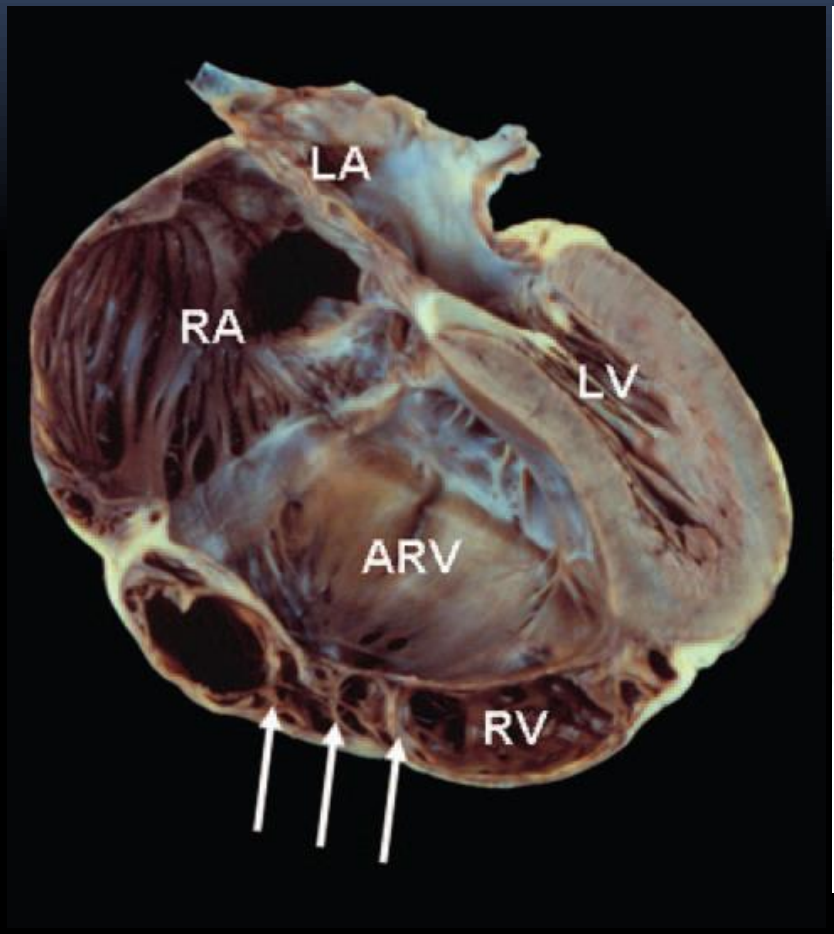
- Ebstein anomaly def.
- Classification
  - by Echo.
  - Carpentier
  - Celermajer index
  - anatomic findings at surgery
- Imaging modality
  - Chest X-ray
  - ECG
  - Echocardiography
  - Cardiac MRI

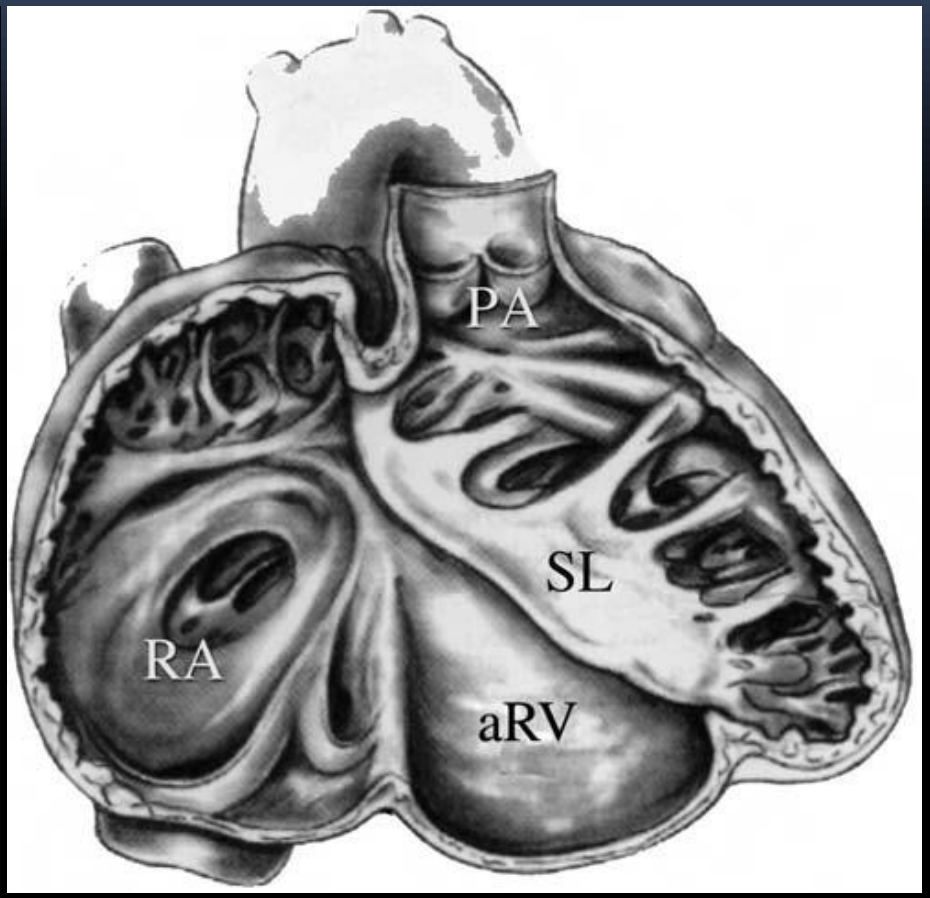
Ebstein anomaly



Mann RJ, Lie JT. The life story of Wilhelm Ebstein (1836 – 1912) and his almost overlooked description of a congenital heart disease. *Mayo Clin Proc. 1979;54:197–204.*

Ebstein anomaly





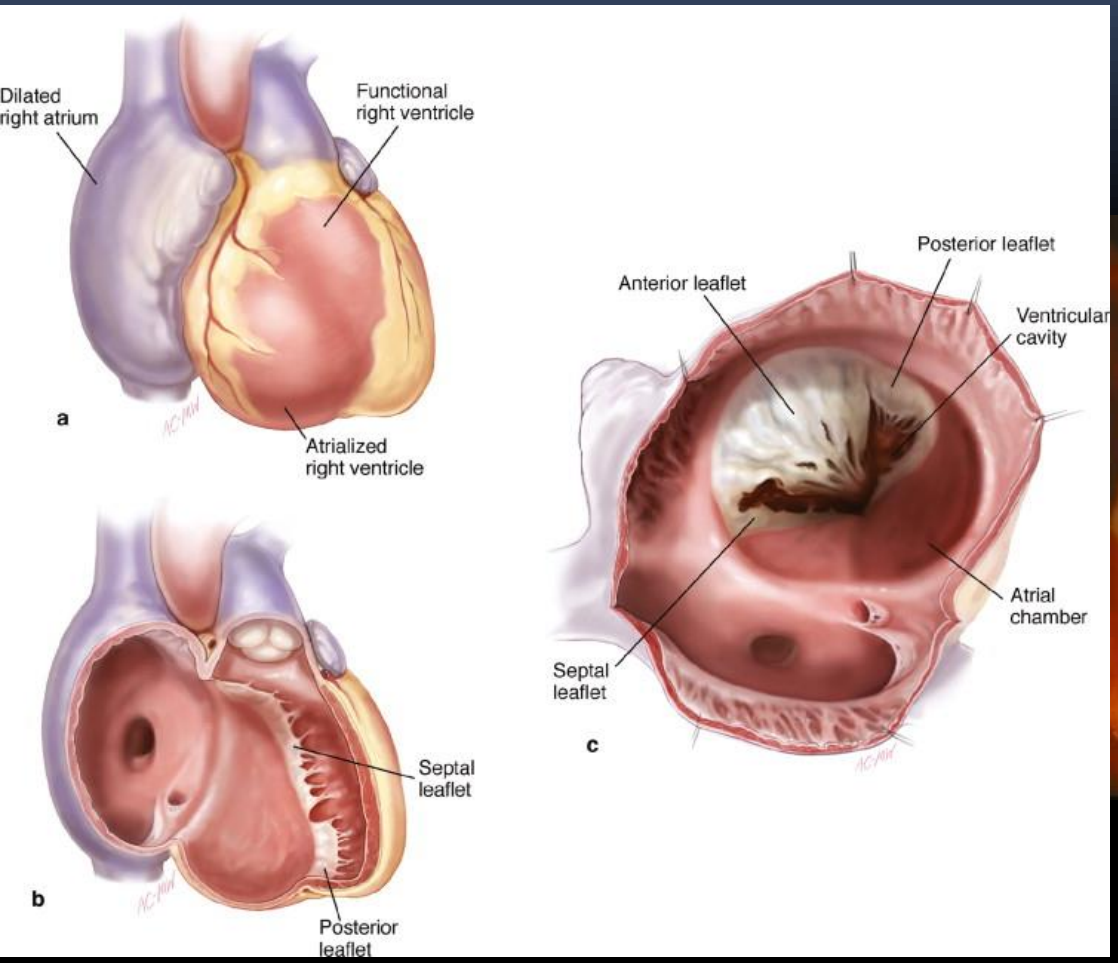
Attenhofer Jost, et al.. Circulation 2007; 115:277.

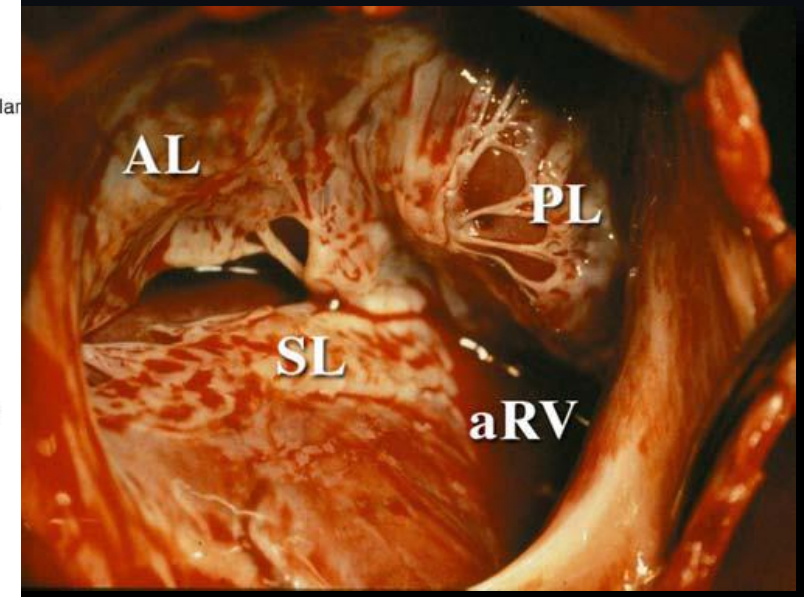
Chauvaud S, Carpentier A (2007). MMCTS. doi:10.1510/mmcts.2007.003038

Marked cardiomegaly

Attenhofer Jost, et al. Circulation 2007; 115:277.

Severe Ebstein's malformation



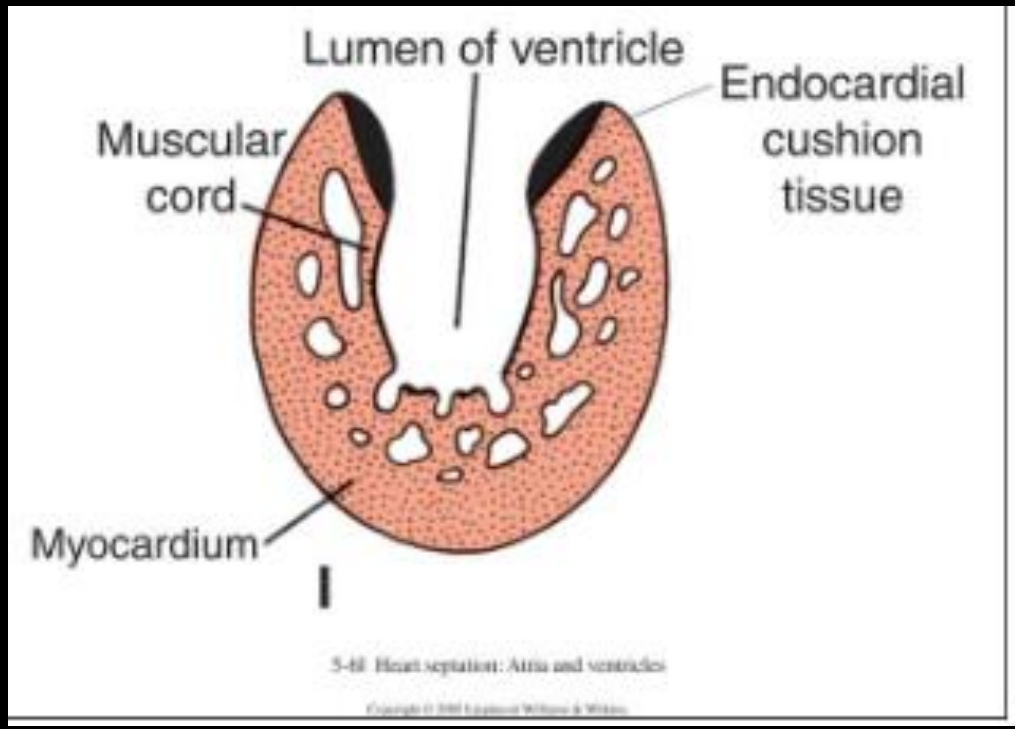


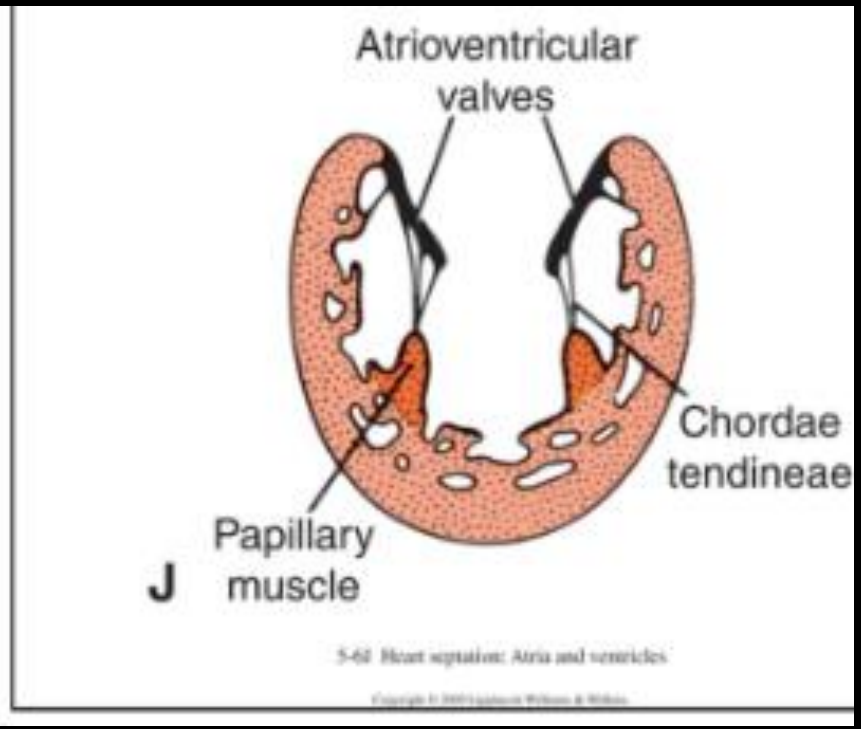
Alain Carpentier, et al. Carpentier's Reconstructive Valve Surgery: From Valve Analysis to Valve Reconstruction CHAPTER 24 , 247-257

Chauvaud S, Carpentier A (2007). MMCTS. doi:10.1510/mmcts.2007.003038

AV valve formation

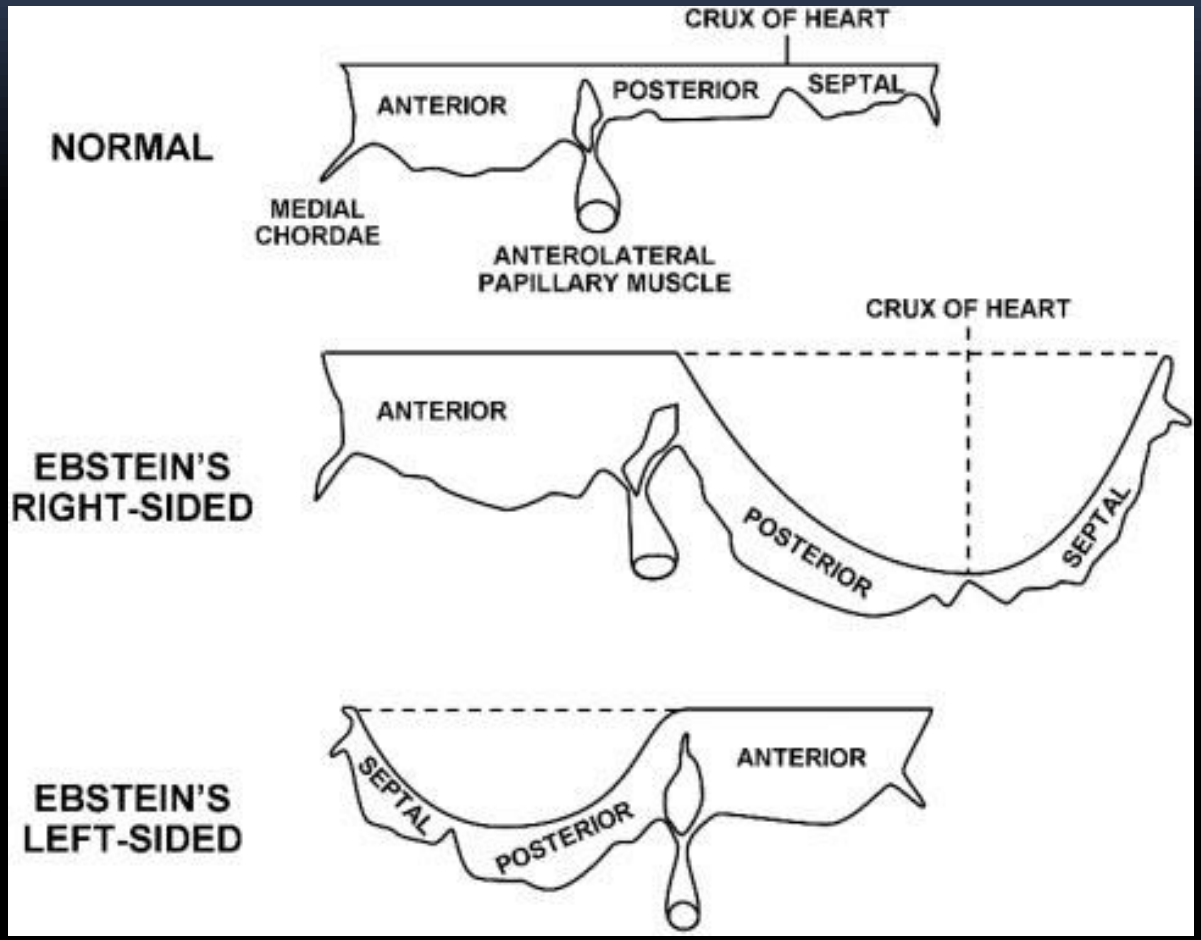
Failure of
delamination of endocardial cushion tissue





http://www.studyblue.com/notes/note/n/heart-embryo/deck/4384504

Apical displacement



Anderson KR, et al. Morphologic spectrum of Ebstein's anomaly of the heart: a review. Mayo Clin Proc. 1979;54:174 –180

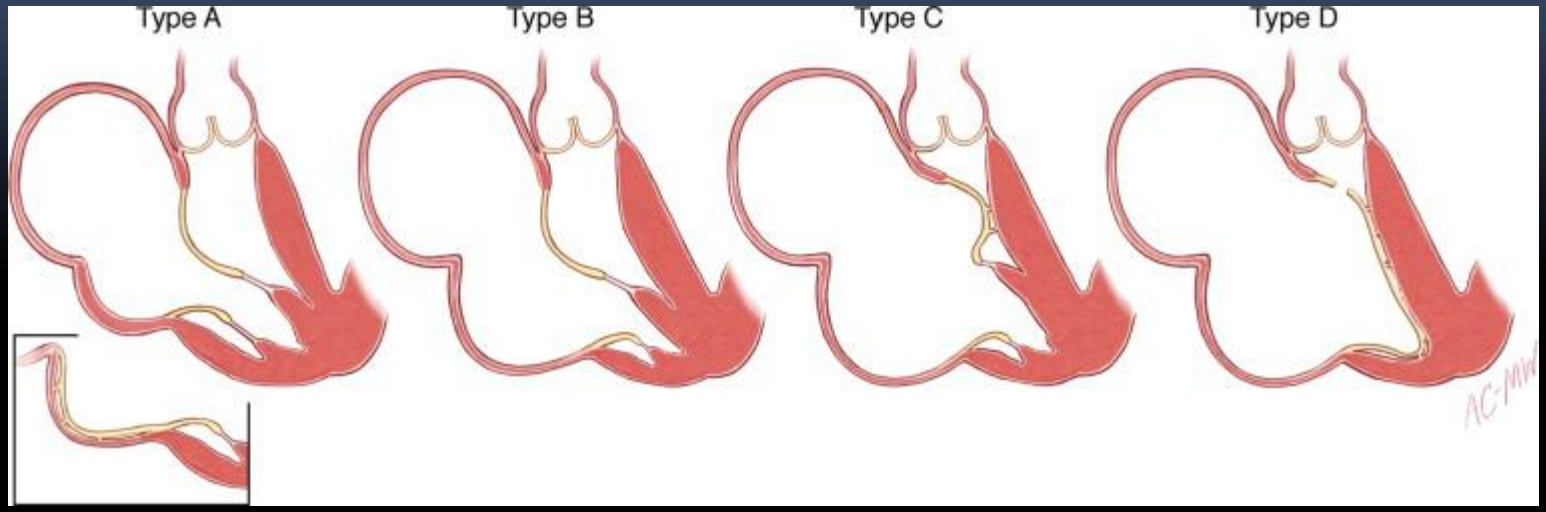
Class. by Echo.

Simple
  mild, moderate, or severe

- extent of apical displacement of vv leaflets
- degree of TR
- degree of RV dilatation & dysfunction

Attenhofer et al. Circulation 2007; 115:277.

Carpentier class.



SL, PL displace.	**+**	**+ +**	**+ + +**	"SAC"
AL				
Morphology	Normal	Abnormal chordae	Partial adhesions	Extensive adhesions
Mobility	Normal	Normal	Restricted	Absent
a RV				
Size	Small	Large	Large	SAC
Contractility	Preserved	Reduced	Minimal	Absent
RV				
Size	Normal	Reduced	Small	Infundibulum
Contractility	Preserved	Preserved /reduced	Reduced	Severely depressed

Class. by anatomic findings at surgery

Type	Anterior Leaflet: Size	Anterior Leaflet: Mobility	Posterior Leaflet	Septal Leaflet	Atrialized RV Chamber Size
I	Larger	Mobile	Apically displaced, dysplastic, or absent.		Varies from relatively small to large.
II	Relatively small and displaced in a spiral fashion toward the apex.				Moderately large.
III		Restricted motion Shortened, fused, and tethered chordae. Direct insertion of papillary muscles into the anterior leaflet is frequently present.	Displaced, dysplastic, and usually not reconstructible.		Large
IV		Severely deformed Few or no chordae. Direct insertion of the papillary muscles into the leading edge of the valve is common.	Typically dysplastic or absent	Represented by a ridge of fibrous material descending apically from the membranous septum.	Nearly the entire RV cavity is atrialized. TV tissue is displaced into the RVOT and may cause obstruction of blood flow (functional tricuspid stenosis).

Dearani, et al. Ann Thorac Surg 2000; 69:S106.
Moss & Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult , 8th edit. 889-912

Celermajer index score

In neonate.

$$\frac{\text{RA area + aRV area}}{\text{fRV area+ LA area + LV area}}$$

Grade	ratio
1	< 0.5
2	0.5 ~ 0.99
3	1 ~ 1.49
4	> 1.5

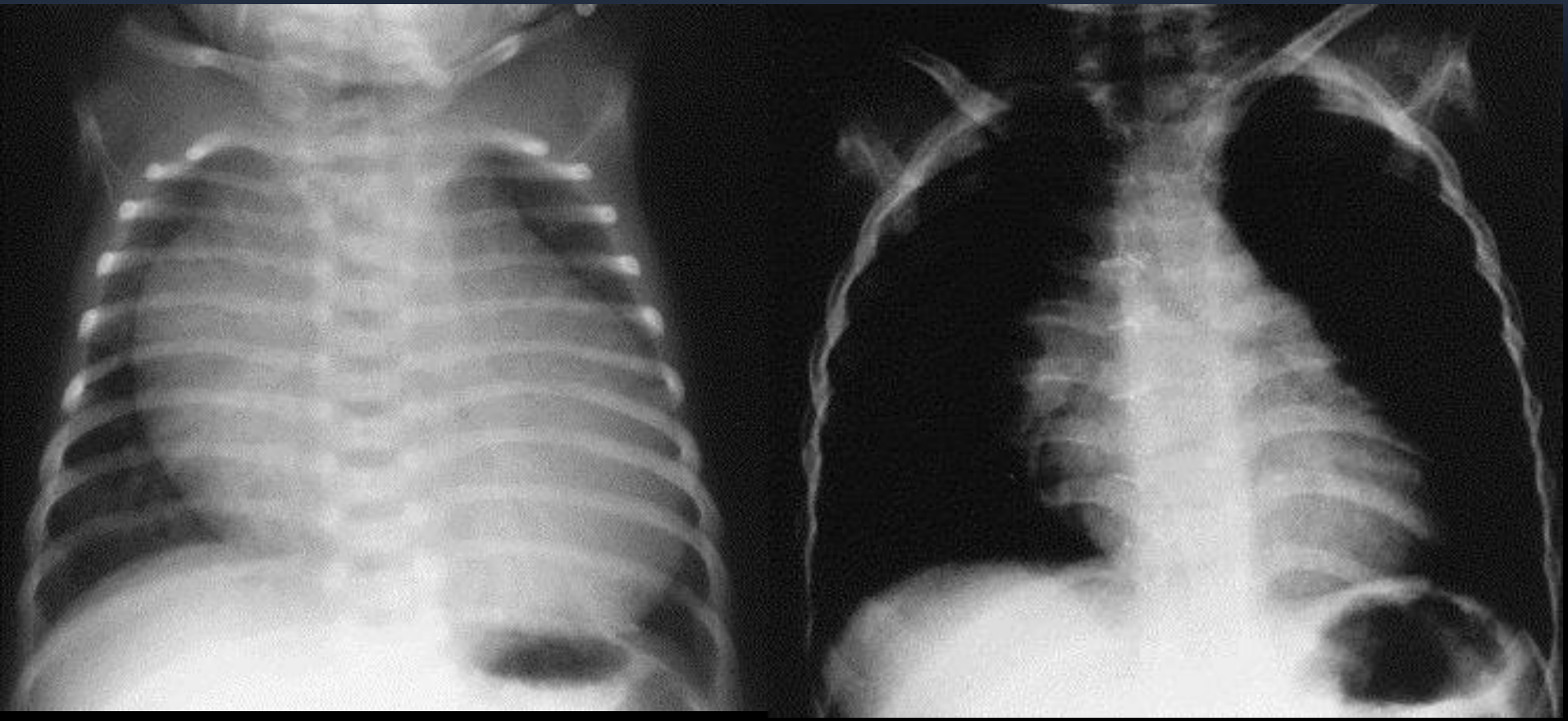
Celermajer et al. J Am Coll Cardiol. 1994;23:170 –176.

Great Ormond Street Echocardiography (GOSE) score

GOSE score	ratio	Mortality (%)
1-2	< 1.0	8
3 (acyanotic)	1.1 ~ 1.4	10 (early) 45 (late)
3 (cyanotic)	1.1 ~ 1.4	100
4	> 1.5	100

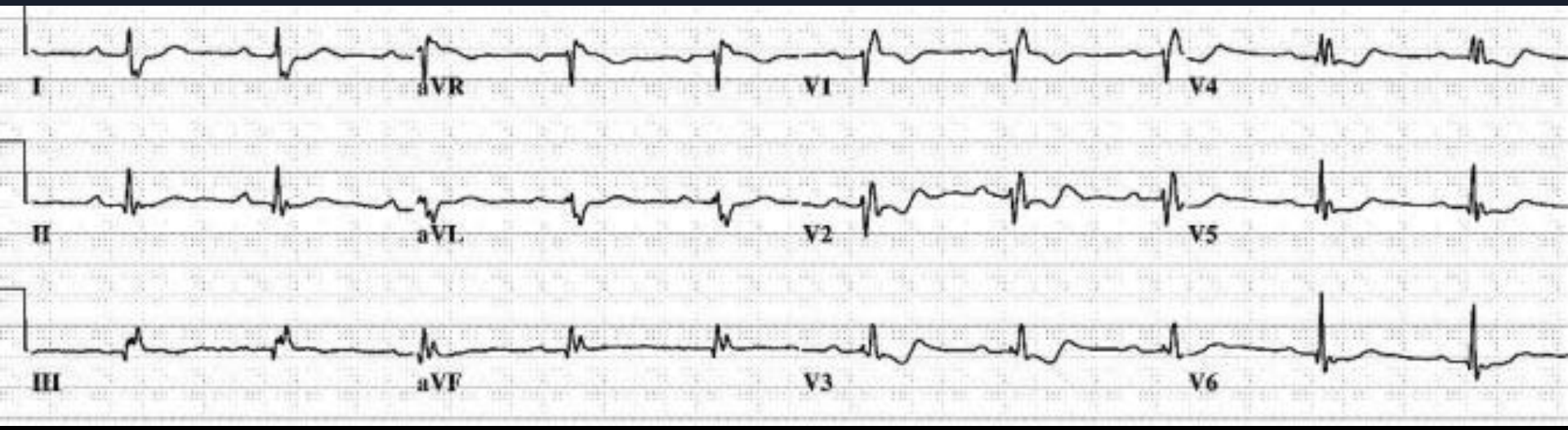
Celermajer et al. J Am Coll Cardiol. 1994;23:170 –176.

Chest X-ray



Knott-Craig et al. Ann Thorac Surg. 2000;69(5):1505-10.

Electrocardiography



Attenhofer Jost, CH, Connolly, HM, Dearani, JA, et al. Ebstein's anomaly. Circulation 2007; 115:277.

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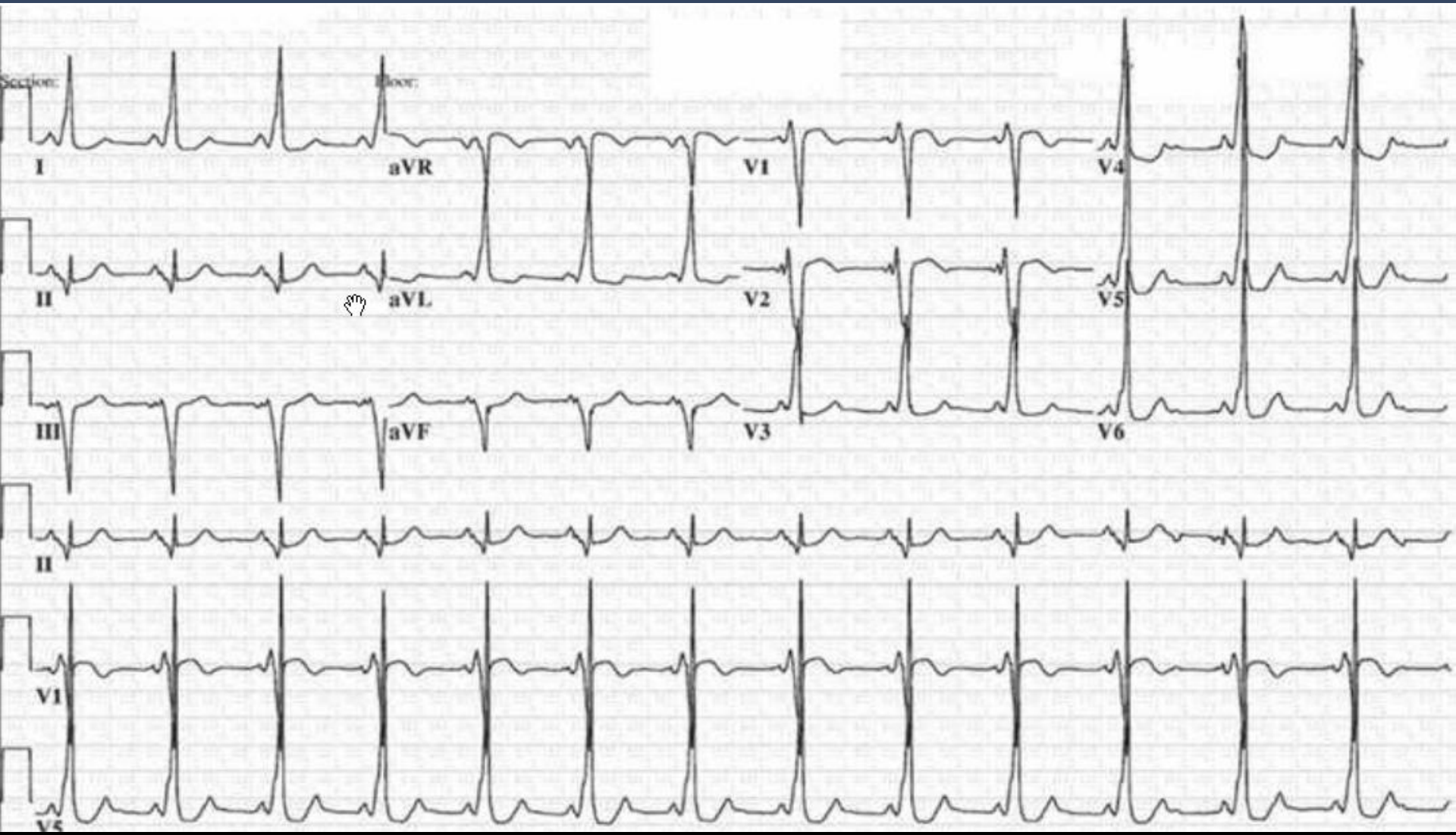
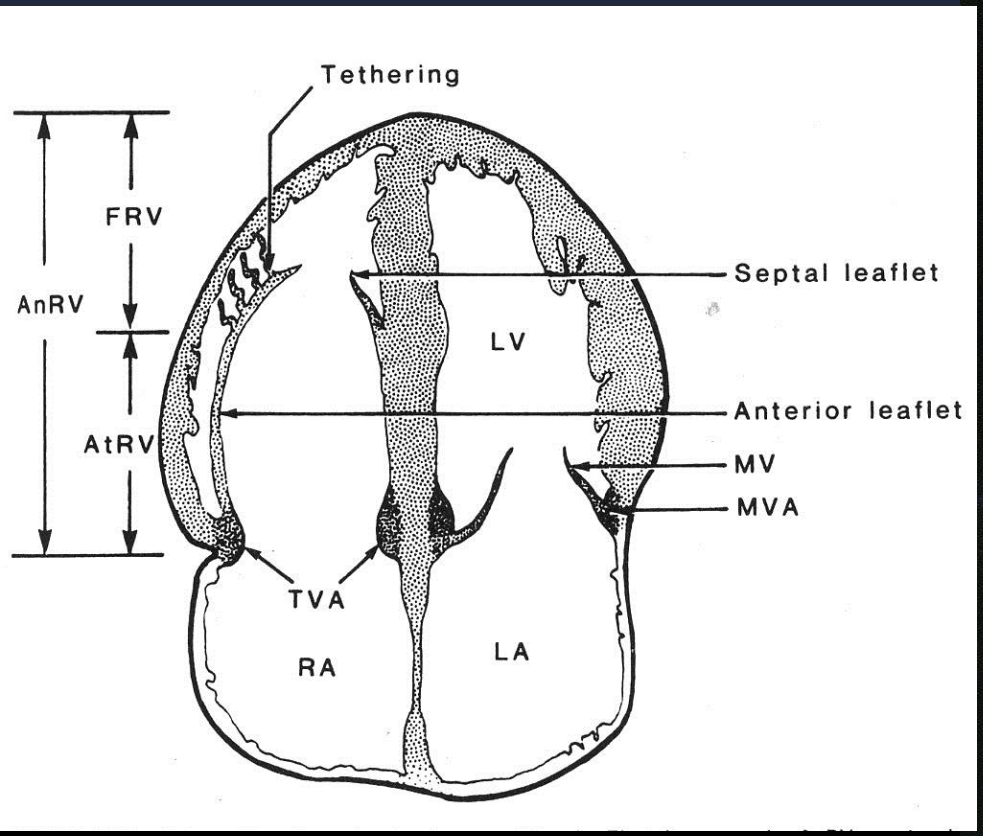
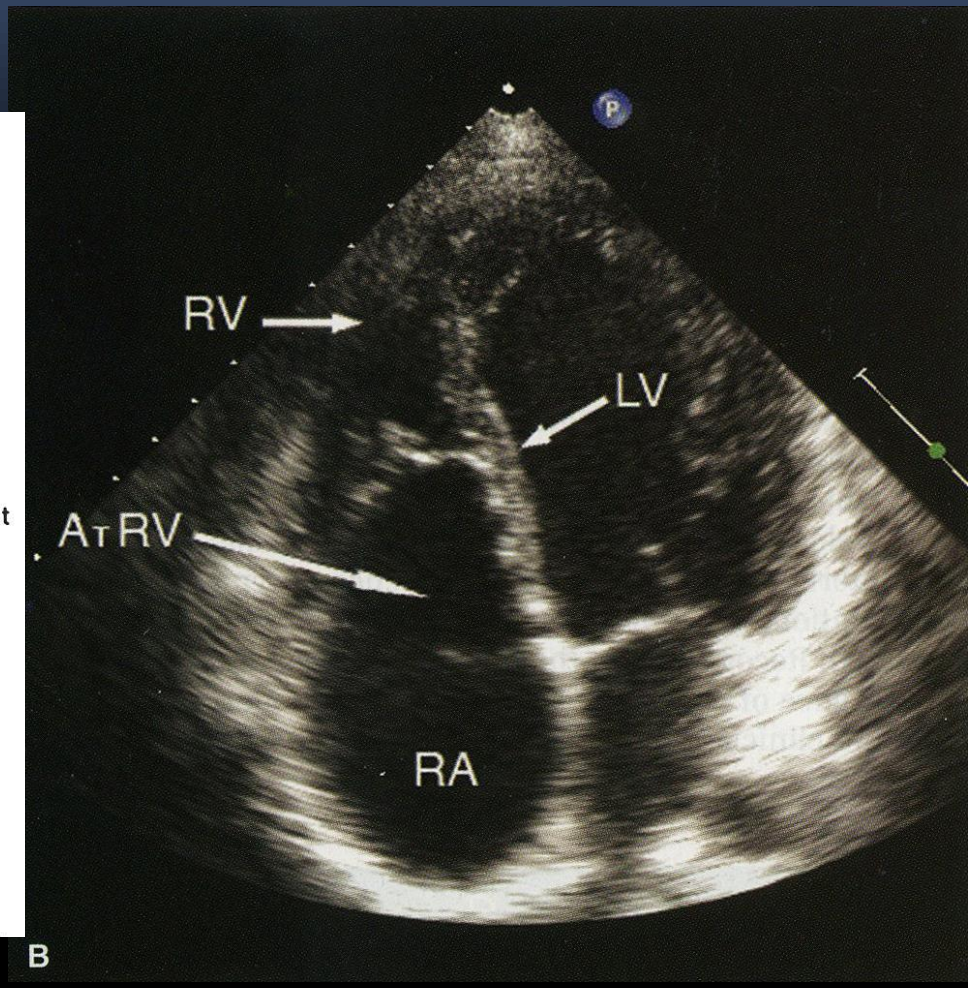


Moss & Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult , 8th edit. 889-912

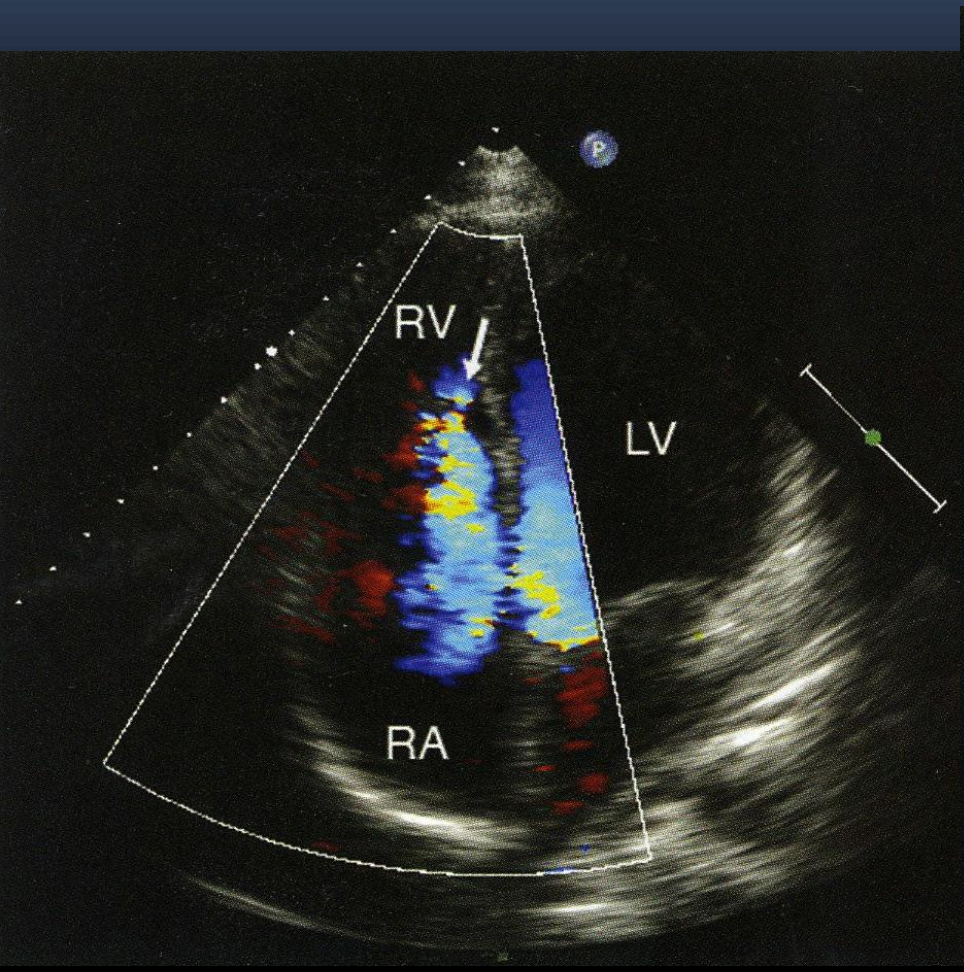
Echocardiography

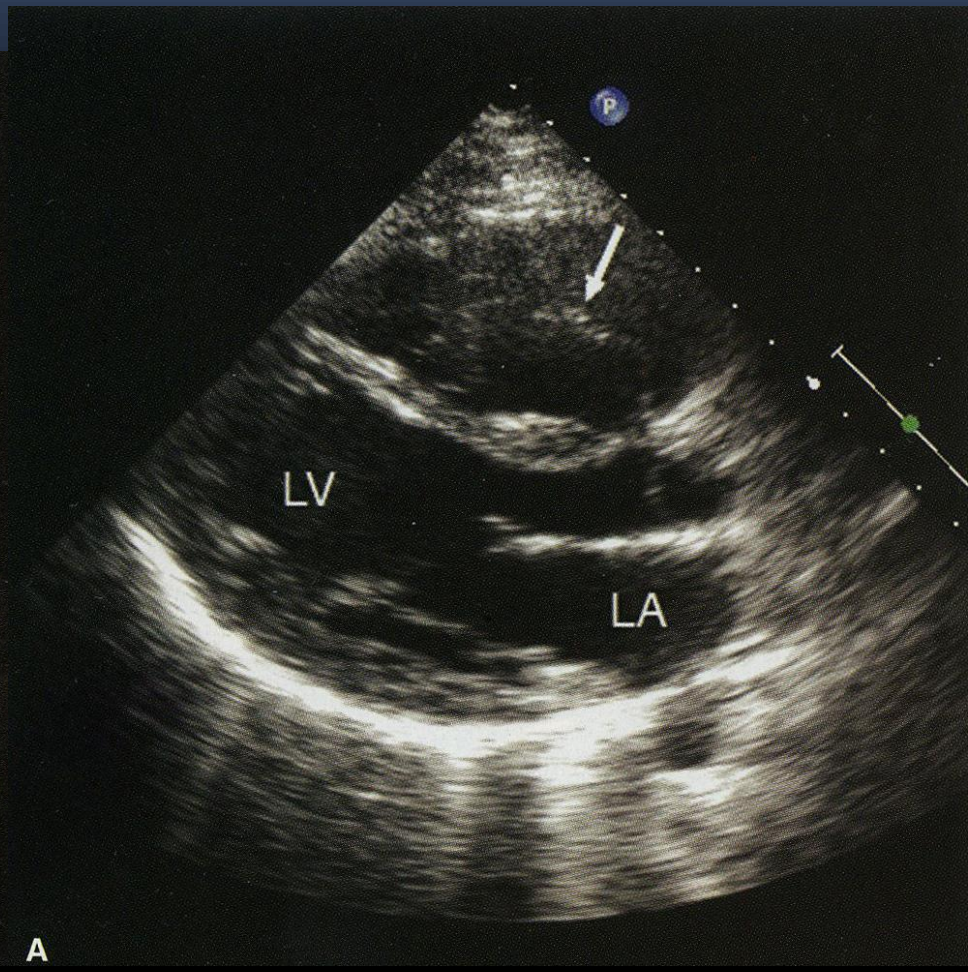




Armstrong et al, Feigenbaum's Echocardiography, Lippincott Williams & Wilkins

Echocardiography





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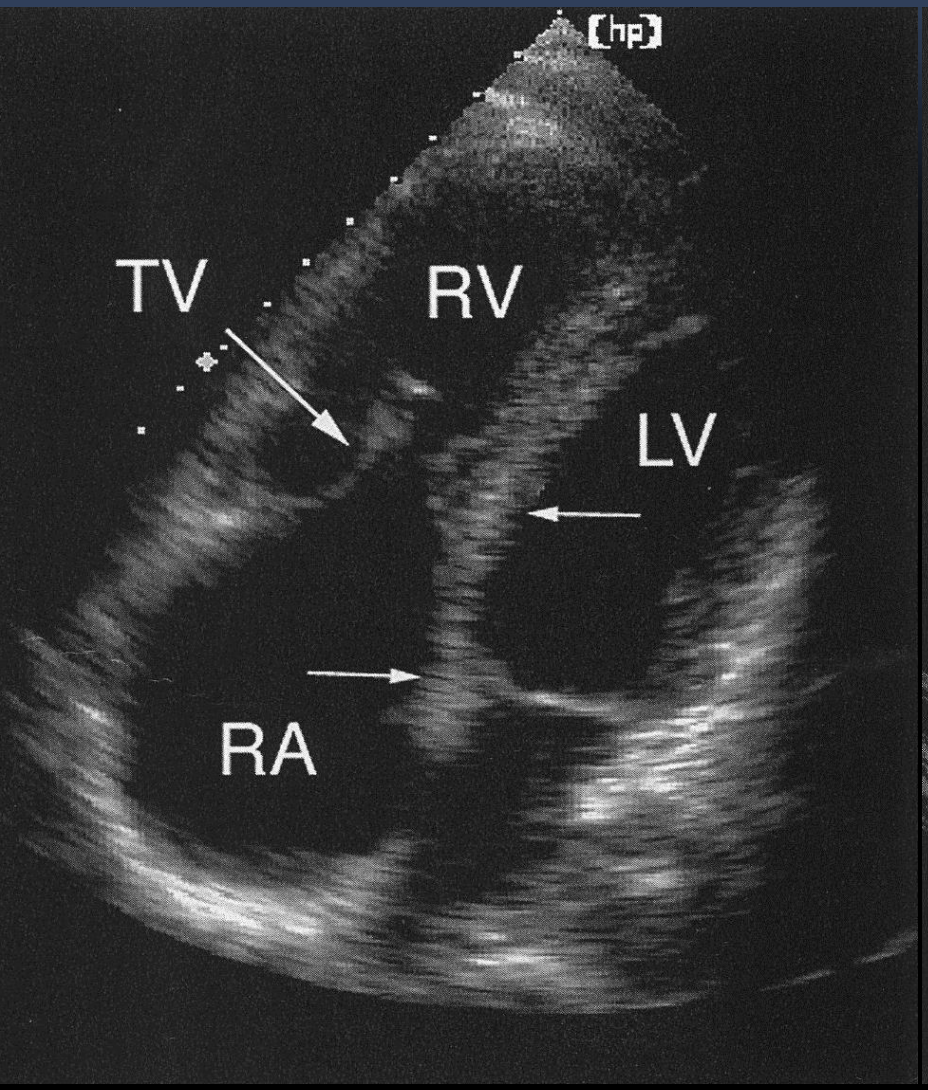

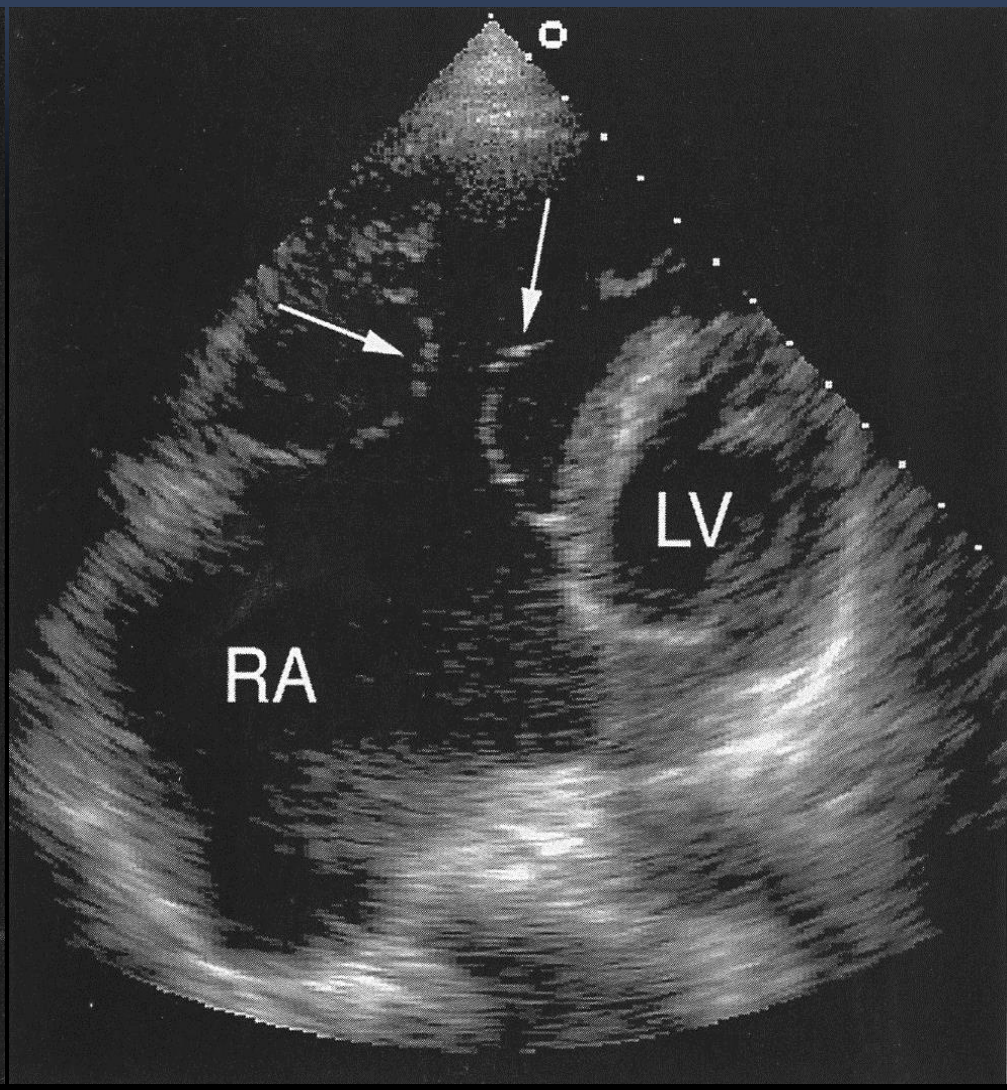

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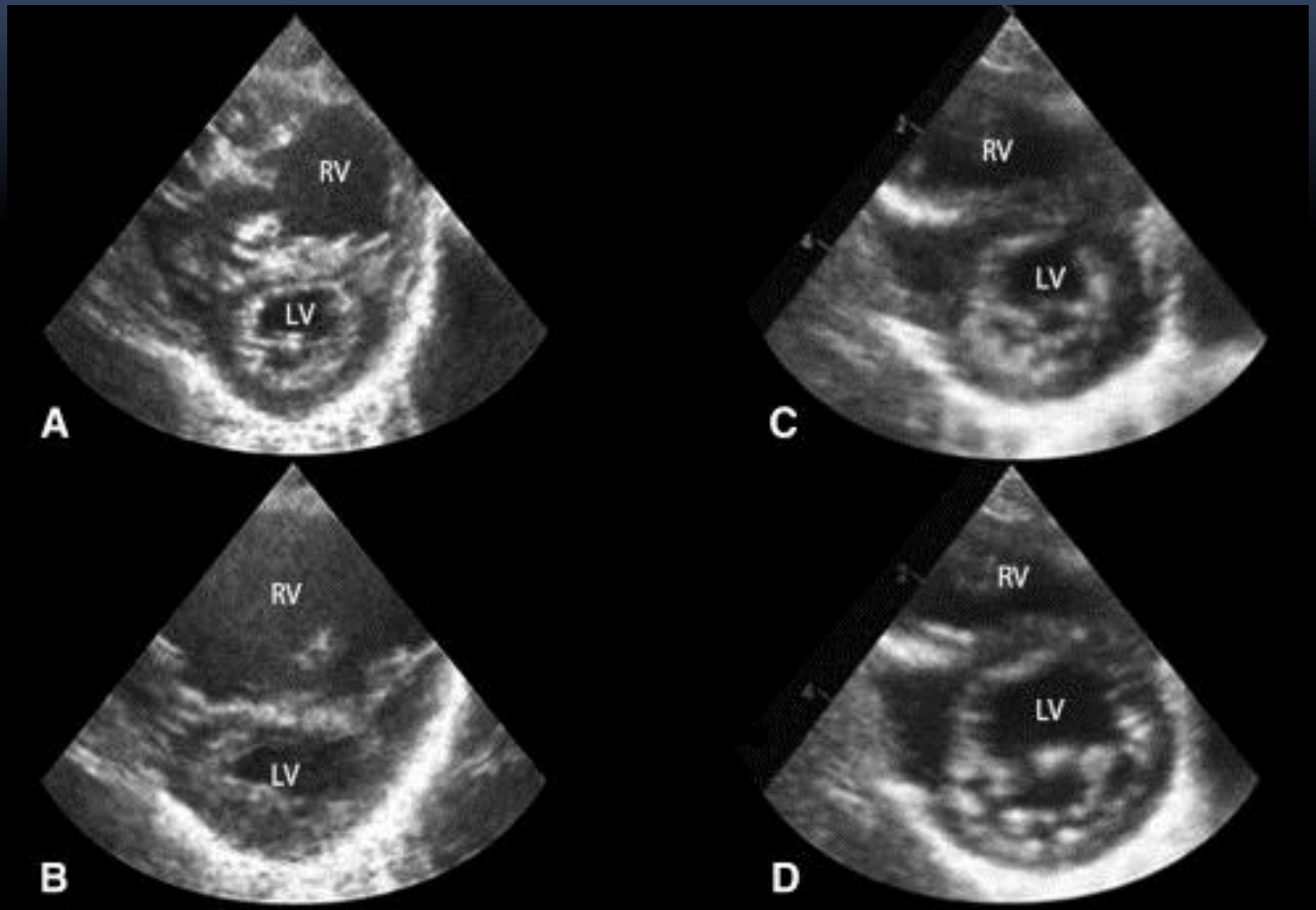
Echocardiography



Kawabata, et al Journal of Thoracic and Cardiovascular Surgery, 142, 1582-1584

Fetal Echocardiography

Szwast, Anita - Fetal Cardiovascular Imaging: A Disease-Based Approach, 287-296

Fetal Echocardiography

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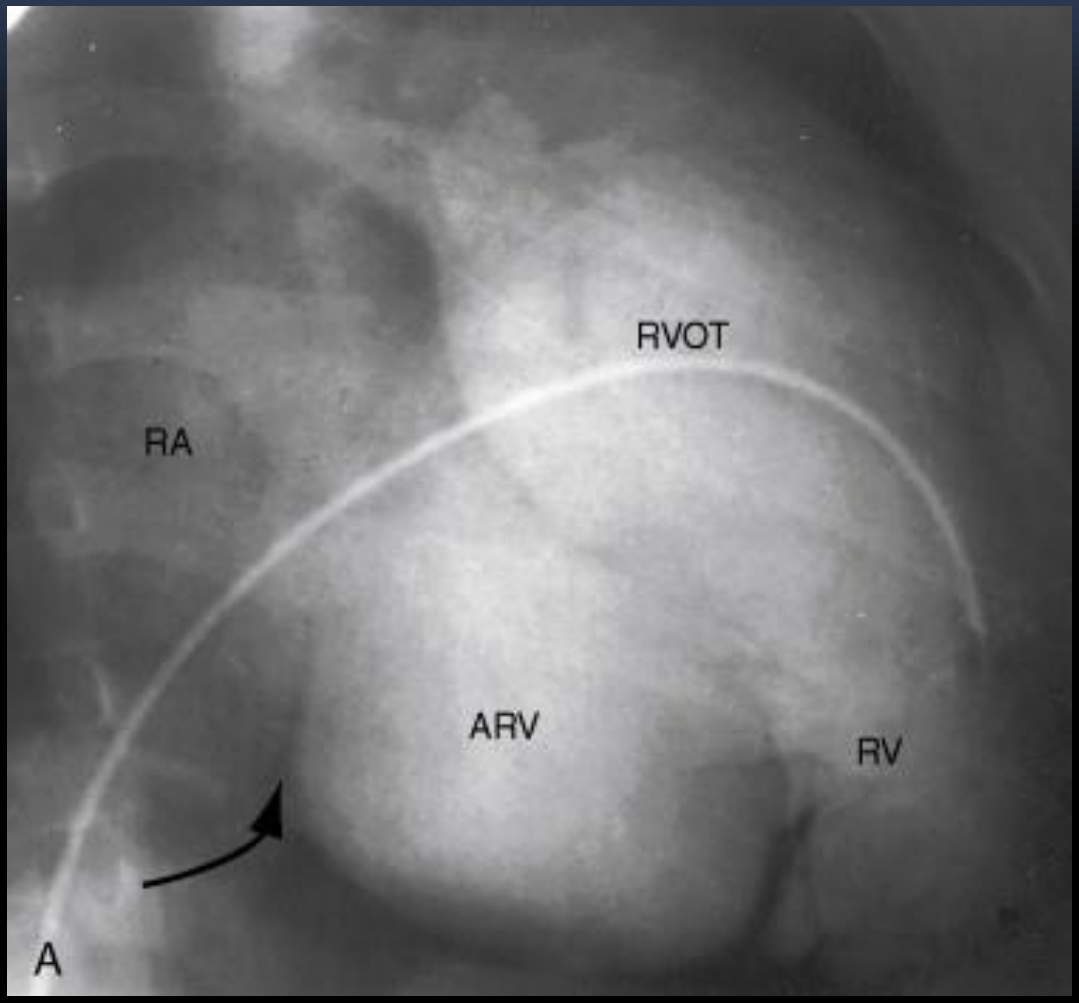
Angiocardiogram

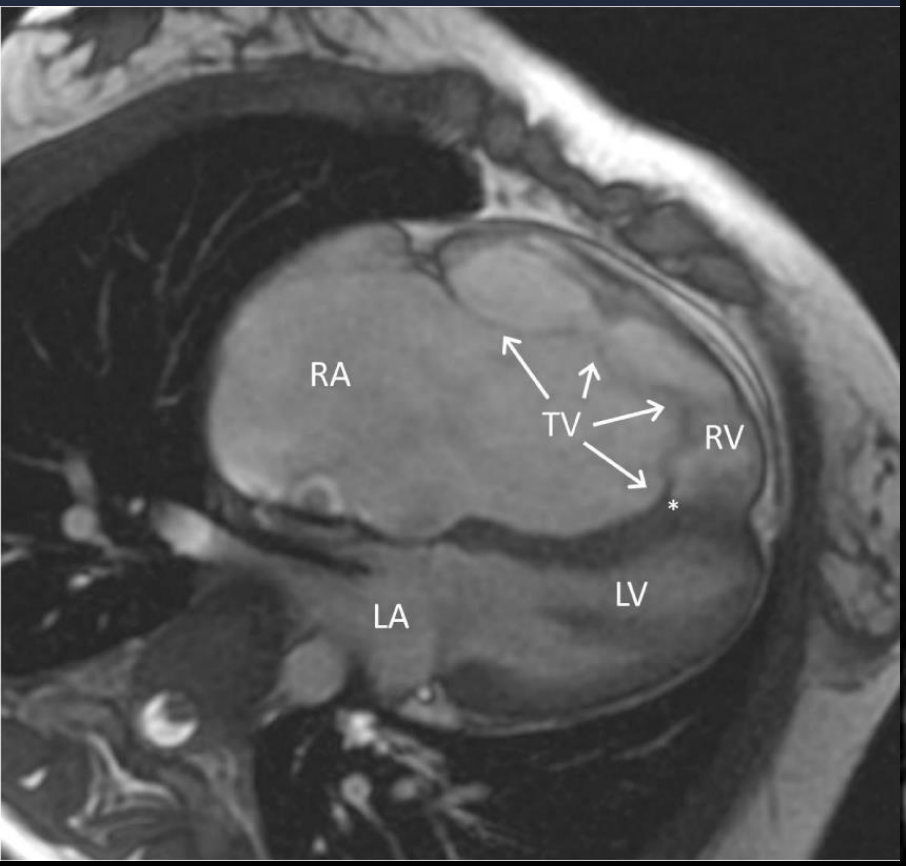


Perloff, Joseph K., MD - Clinical Recognition of Congenital Heart Disease, 176-195

Cardiac CT

Aggarwala, et al Journal of Cardiovascular Computed Tomography 1, Issue 3, 168-169

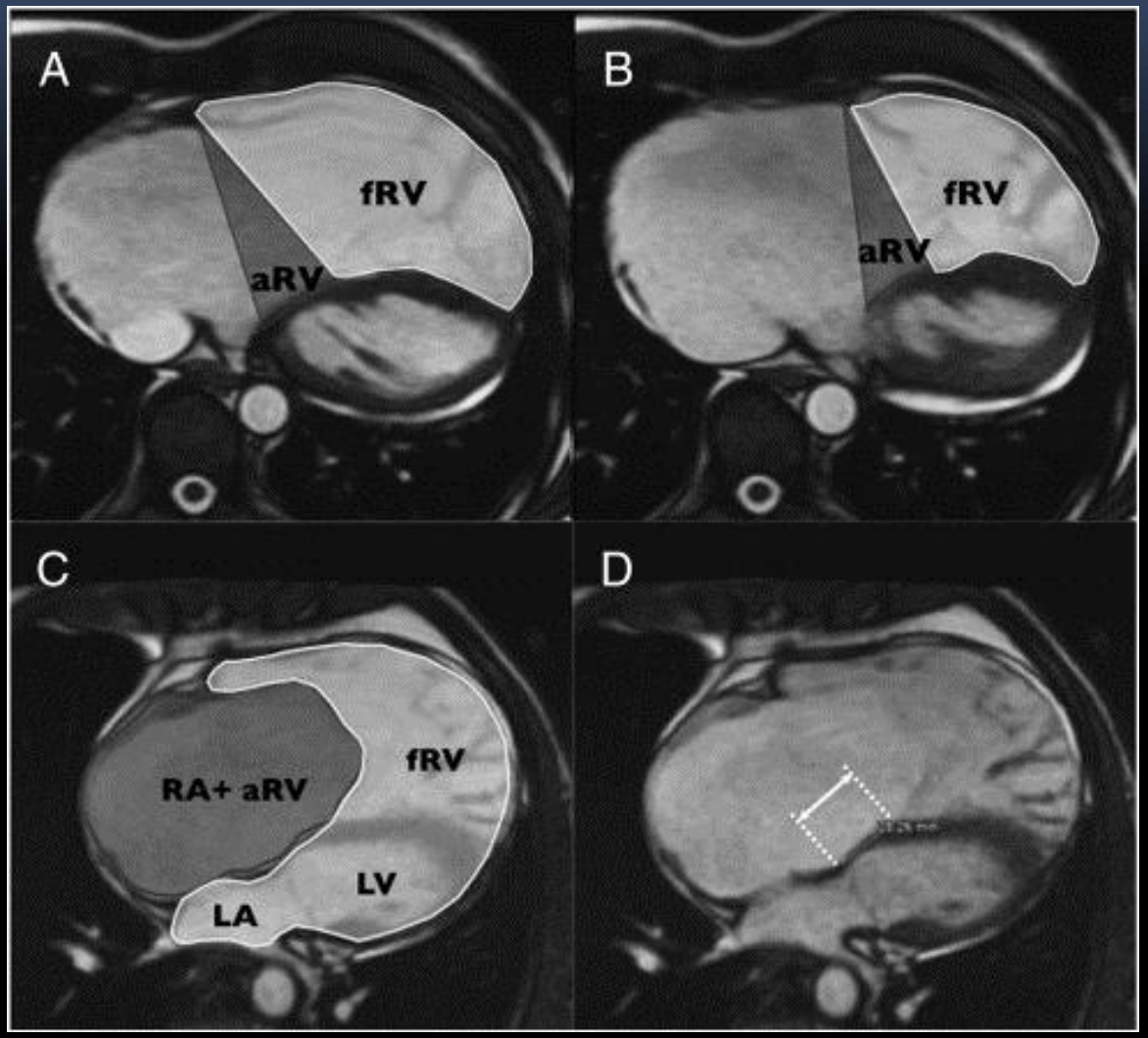
Cardiac MRI



Steinmetz et al (2012). J Clin Exp Cardiolog S8:008.

Dimopoulos, Konstantinos - Cases in Adult Congenital Heart Disease, 167-172

Cardiac MRI



Yalonetsky, Sergey, et al. Am J Cardiol 2011;107:767–773

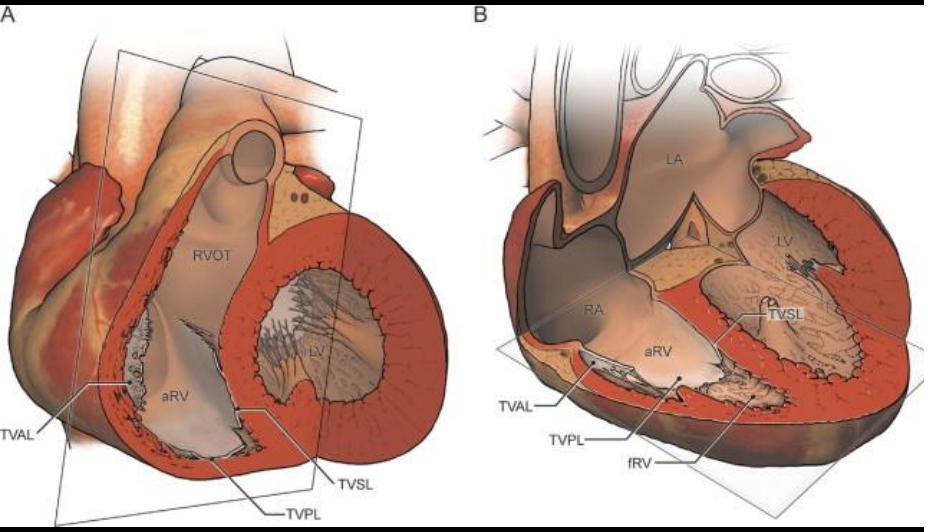
Cardiac MRI

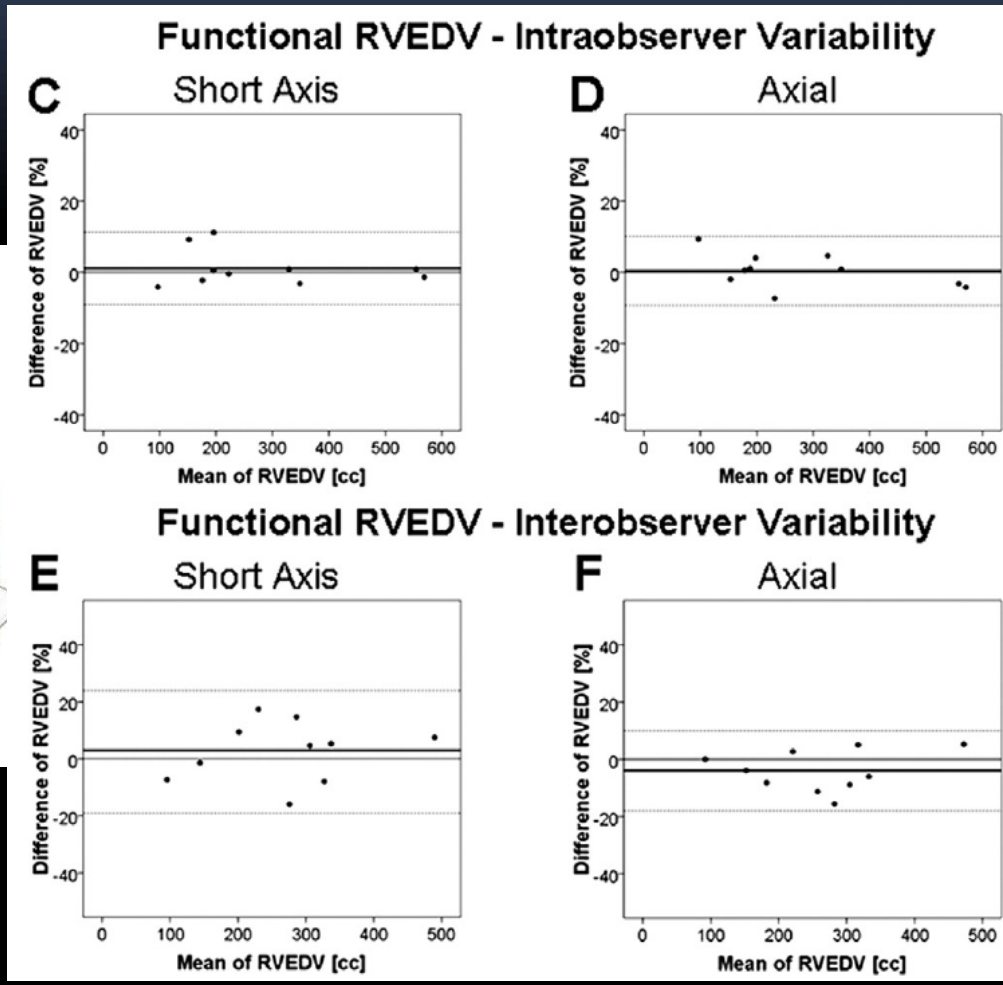
Cardiac Magnetic Resonance Imaging and the Assessment of Ebstein Anomaly in Adults

Sergey Yalonetsky, MD[a], Daniel Tobler, MD[a], Matthias Greutmann, MD[a], Andrew M. Crean, MD[a,b], Bernd J. Wintersperger, MD[b,c], Elsie T. Nguyen, MD[b], Erwin N. Oechslin, MD[a], Candice K. Silversides, MD[a], and Rachel M. Wald, MD[a,d,*]

No published studies have evaluated the role of cardiac magnetic resonance (CMR) imaging for the assessment of Ebstein anomaly. Our objective was to evaluate the right heart characteristics in adults with unrepaired Ebstein anomaly using contemporary CMR imaging techniques. Consecutive patients with unrepaired Ebstein anomaly and complete CMR studies from 2004 to 2009 were identified (n = 32). Volumetric measurements were obtained from the short-axis and axial views, including assessment of the functional right ventricular (RV) end-diastolic volume (EDV) and end-systolic volume. The volume of the atrialized portion of the right ventricle in end-diastole was calculated as the difference between the total RVEDV and the functional RVEDV. The reproducibility of the measurements in the axial and short-axis views was determined within and between observers. The median value derived from the short-axis and axial views was 136 ml/m^2 (range 59 to 347) and 136 ml/m^2 (range 63 to 342) for the functional RVEDV, 153 ml/m^2 (range 64 to 441) and 154 ml/m^2 (range 67 to 436) for the total RVEDV, 49% (range 32% to 46%) and 50% (range 40% to 64%) for the functional RV ejection fraction, respectively. The axial measurements demonstrated lower intraobserver and interobserver variability than the short-axis approach for all values, with the exception of the intraobserver functional RVEDV and interobserver total RVEDV for which the limits of agreement and variance were not significantly different between the 2 views. In conclusion, measurements of right heart size and systolic function in patients with Ebstein anomaly can be reliably achieved using CMR imaging. Axial imaging appeared to provide more reproducible data than that obtained from the short-axis views. (Am J Cardiol 2011;107:767–773)

Cardiac MRI





Yalonetsky, Sergey, et al. Am J Cardiol 2011;107:767–773

Summary

Ebstein anomaly
- SL & PL to the underlying myocardium
- Downward displacement of the functional annulus
  - > 8 mm/m2 BSA
- Atrialized RV dilation
- AL : redundancy, fenestrations, & tethering

Classification
- Echo.
- Carpentier
- Celermajer score (GOSE)
- Anatomic findings during surgery

Dx & Assessment
- mainly echocardiography
- Cardiac MRI