



Cleveland Clinic Children's

NOCIA Symposium

4/27/2019

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Pediatric Cardiology Fellow

I have no disclosures

Objectives

- **Case illustrations of ACHD**
- **Embryology**
- **Echocardiography**
- **Surgical Options**
- **Take home points for my imaging friends**

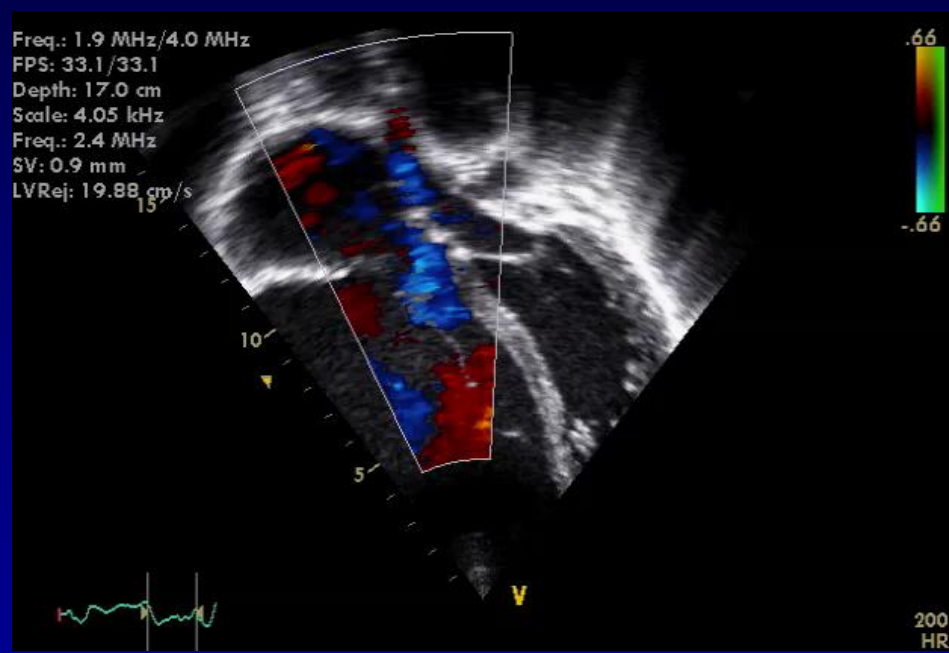
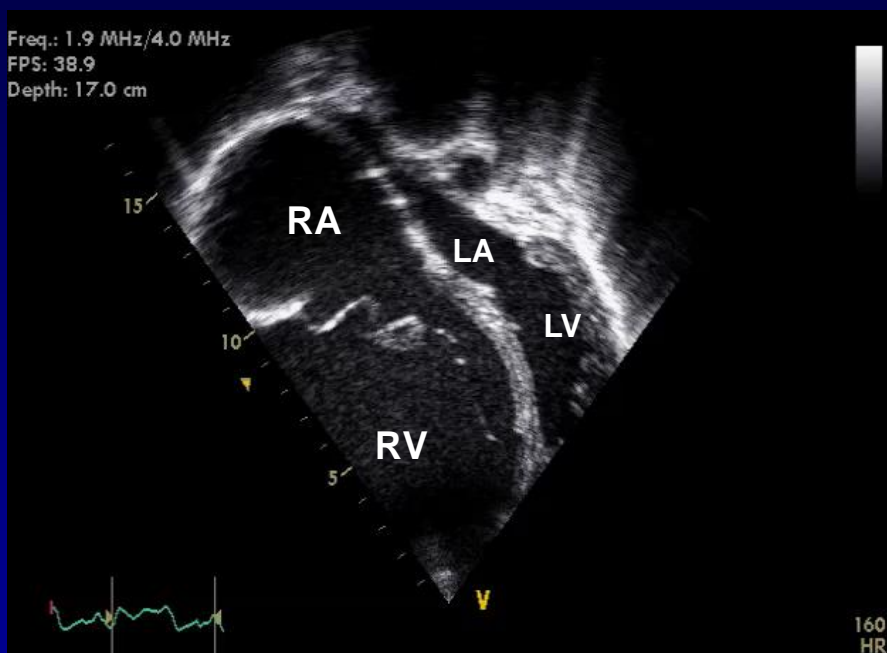
Case 1



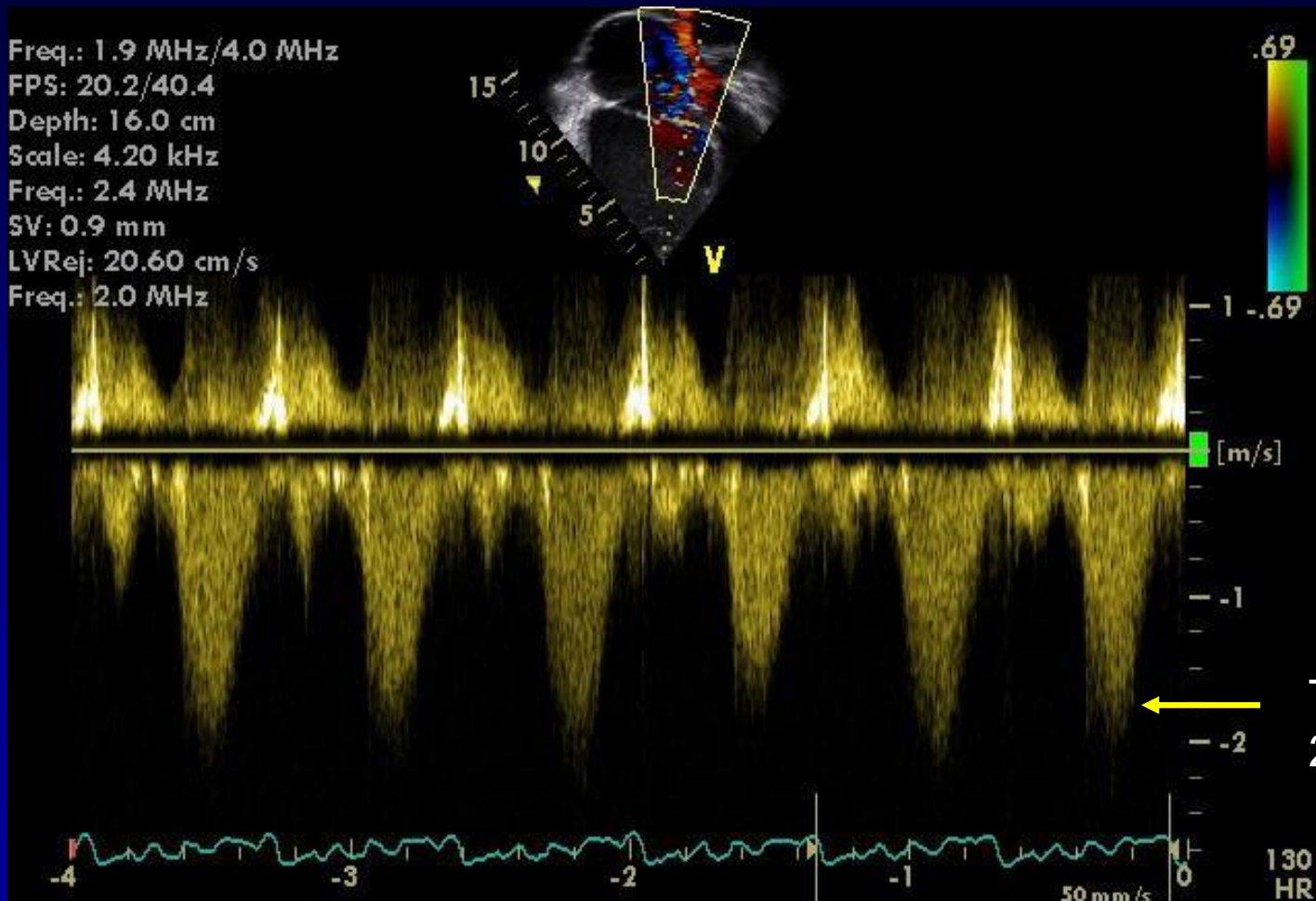
- **25 year old male with known congenital heart disease diagnosed at age 3**
- **He underwent initial surgery at 3 years of age (details unknown)**
- **He presented with impaired exercise tolerance and shortness of breath**
- **He also had history of atrial arrhythmias**

Transthoracic echo

Apical 4 chamber view



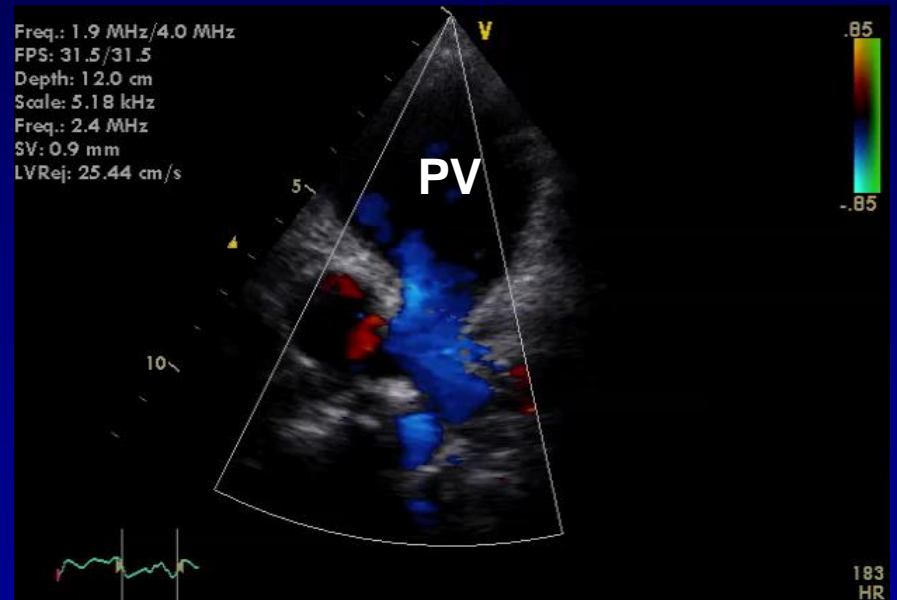
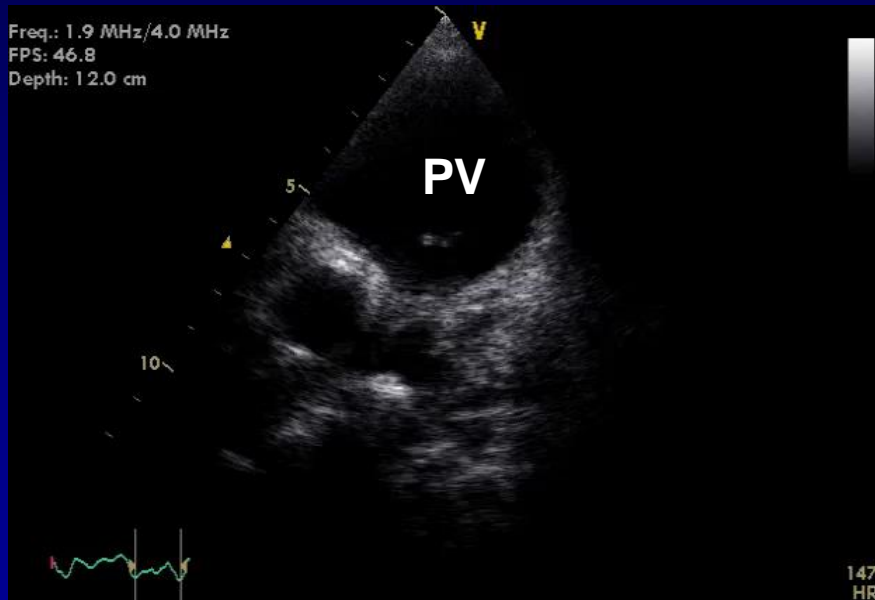
Severe TR through a very hyper mobile anterior leaflet



Velocity and severity are not the same

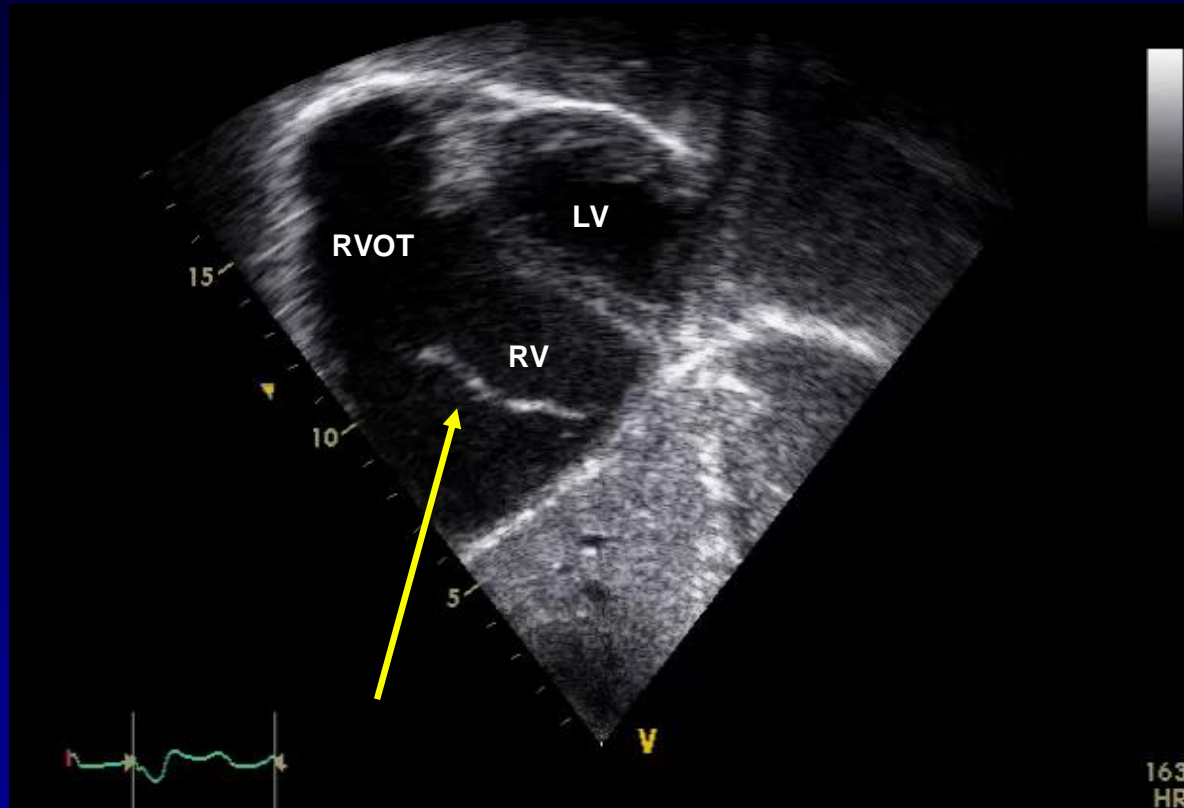
Transthoracic echo

PSAX looking at the Pulmonary Valve



No stenosis, mild- moderate PR

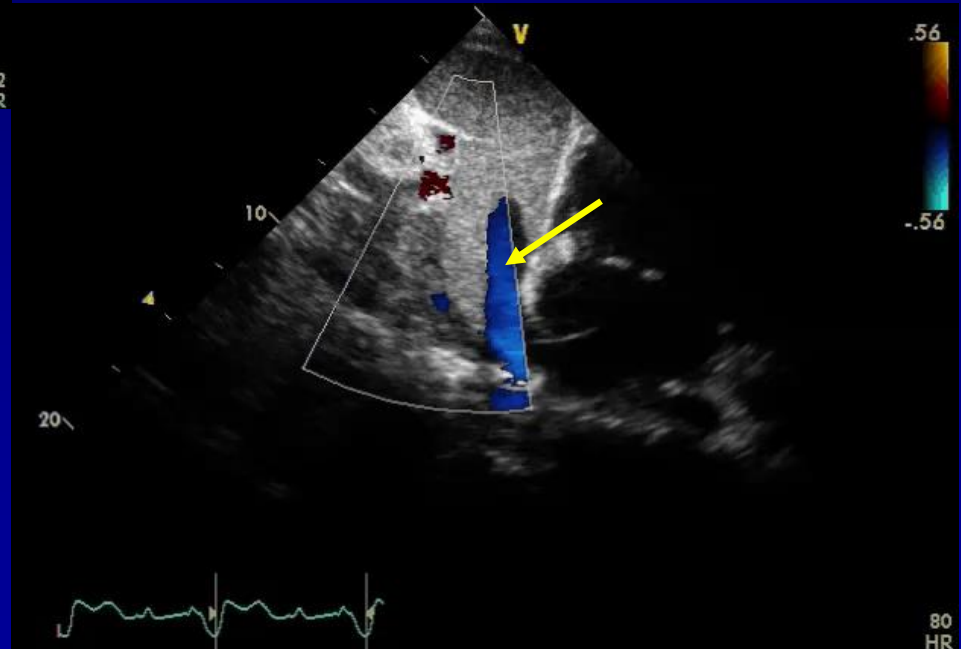
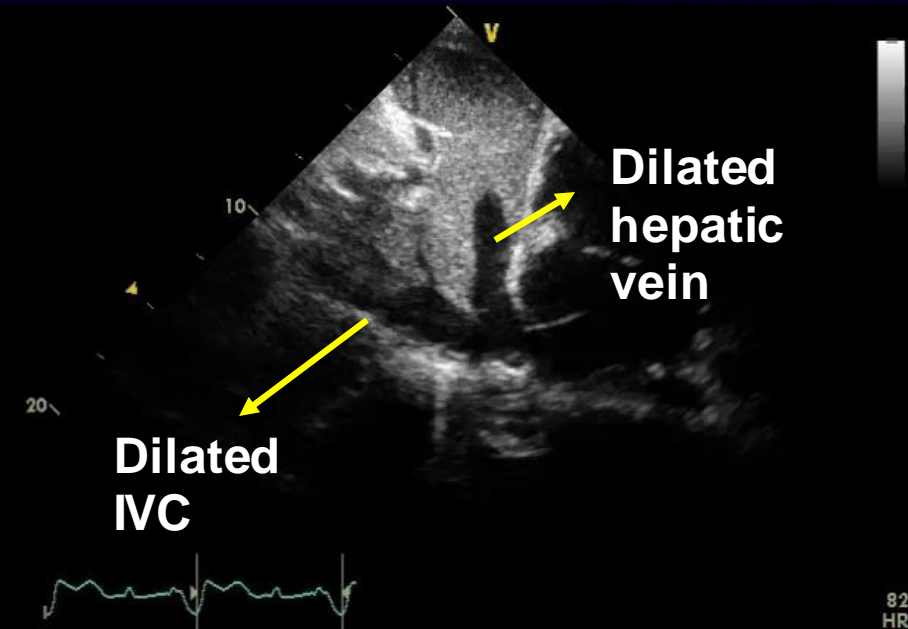
Subcostal Views



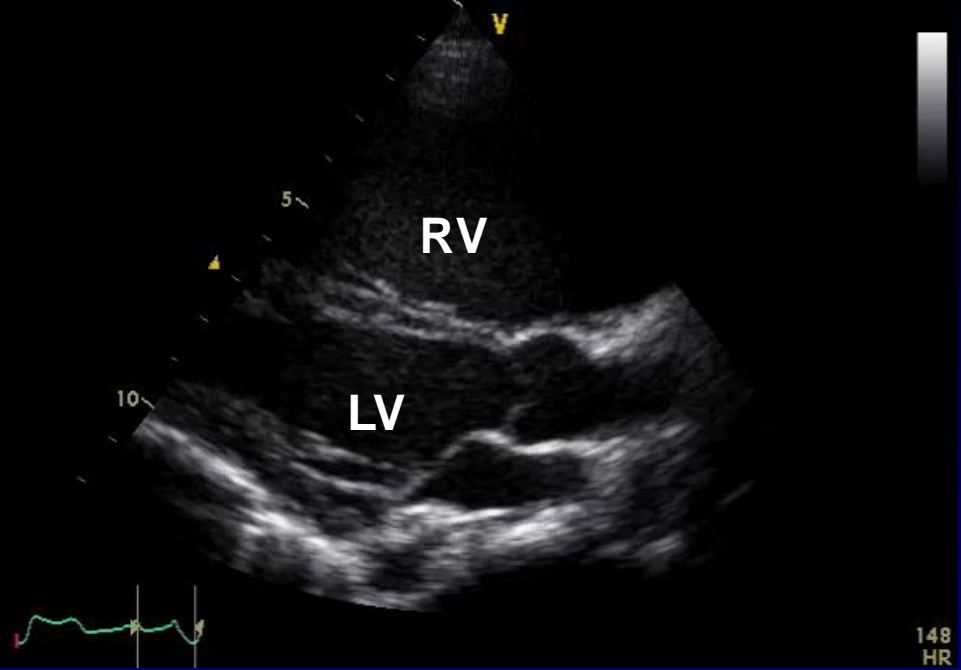
Subcostal short axis view showing the anterior leaflet in the RVOT

Subcostal IVC

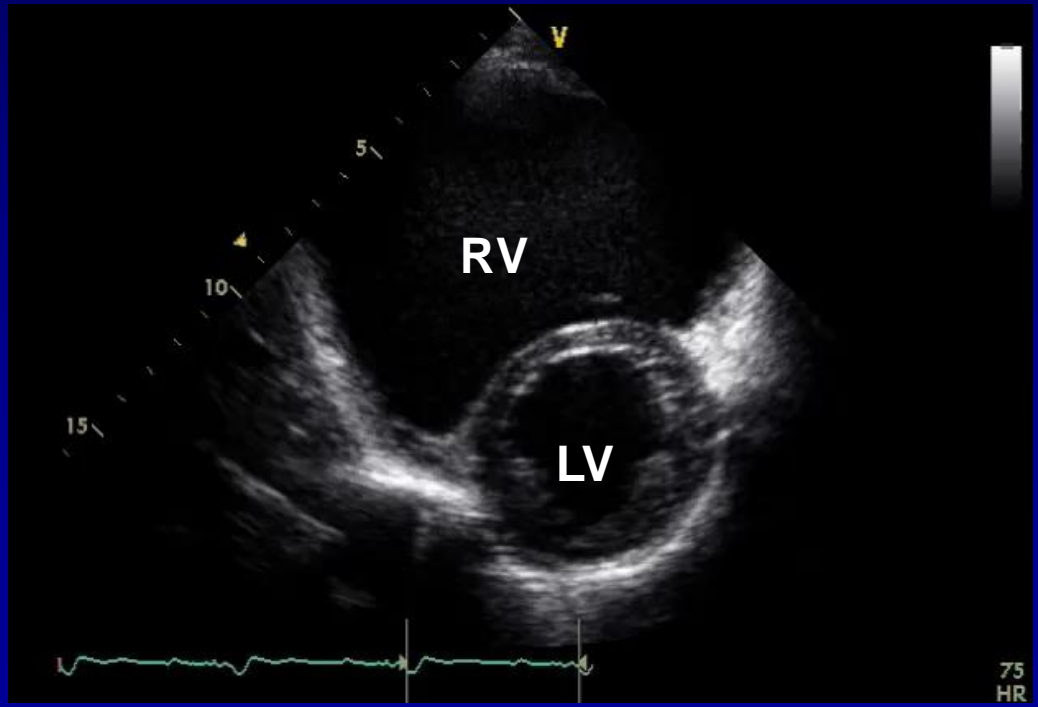
Provides an estimate of right atrial pressure



PLAX

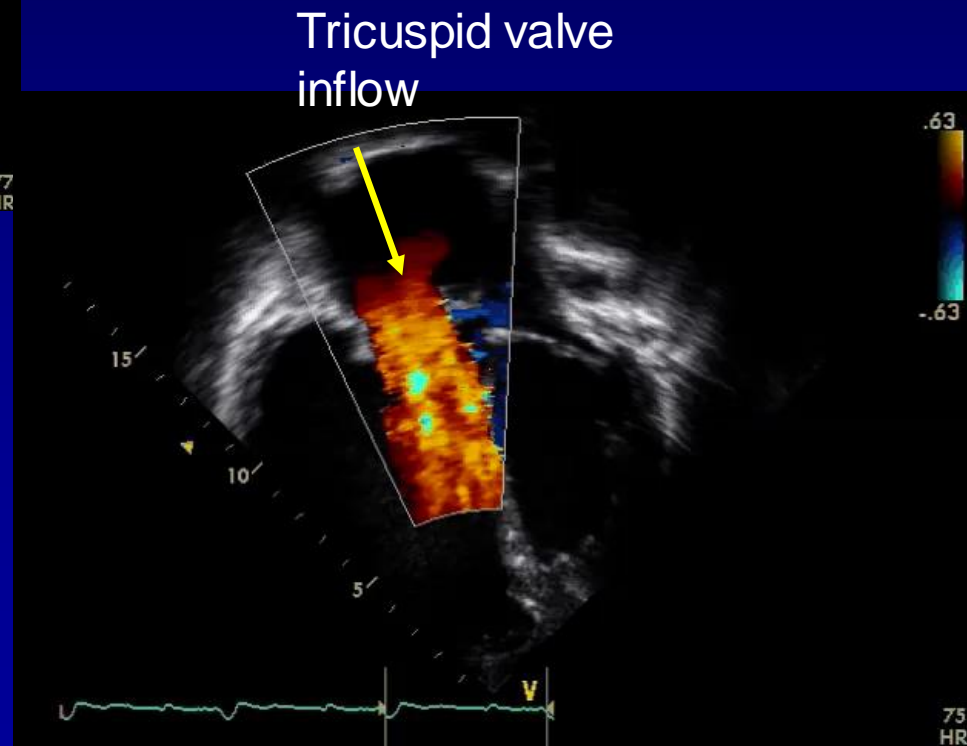
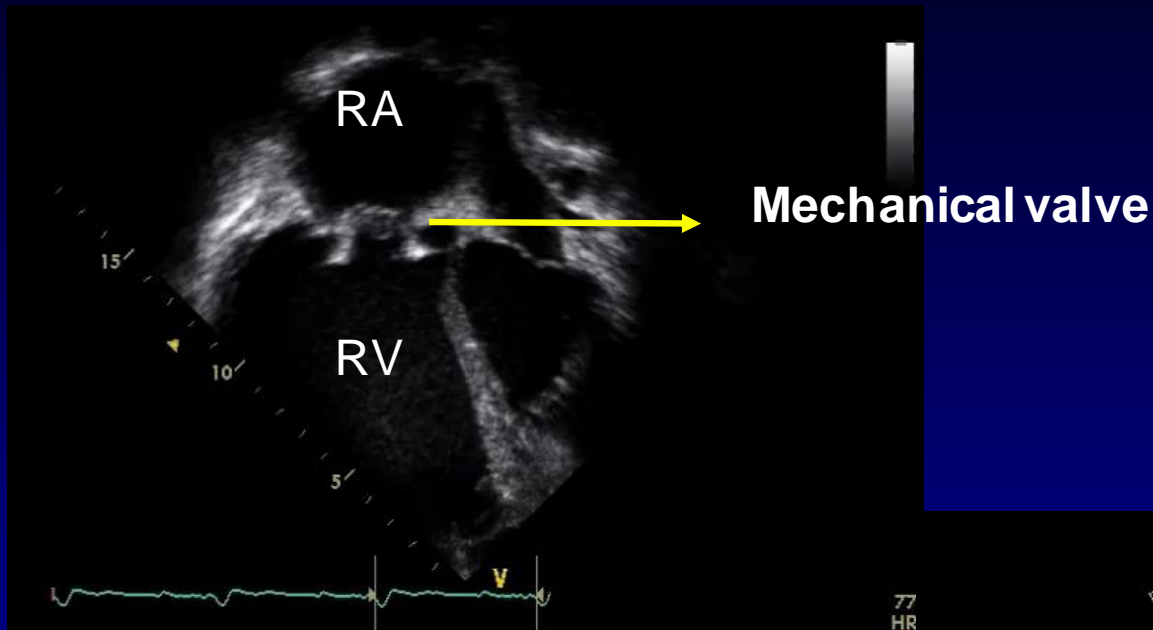


PSAX



**Patient underwent
Tricuspid valve replacement**

Tricuspid Valve Replacement



Ebstein's Anomaly

- **Dr. Wilhelm Ebstein**
- **Described this lesion in a 19-year-old laborer, Joseph Prescher on autopsy in 1864**
- **Cyanosis, cardiomegaly, palpitations and JVD**



Fig. 2. The Ernst August Hospital, University of Göttingen, was constructed in 1851. Wilhelm Ebstein was appointed as Professor of Medicine at this location in 1874.

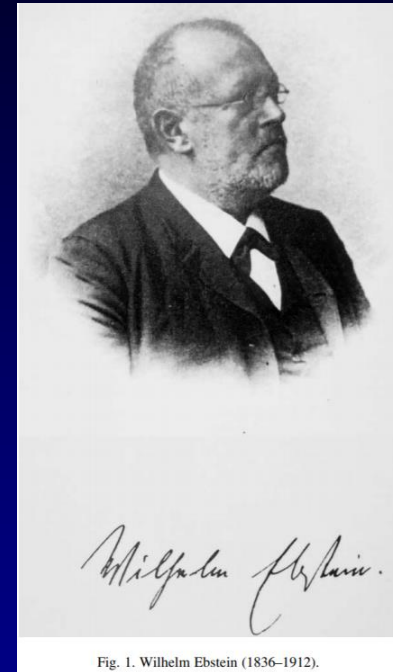


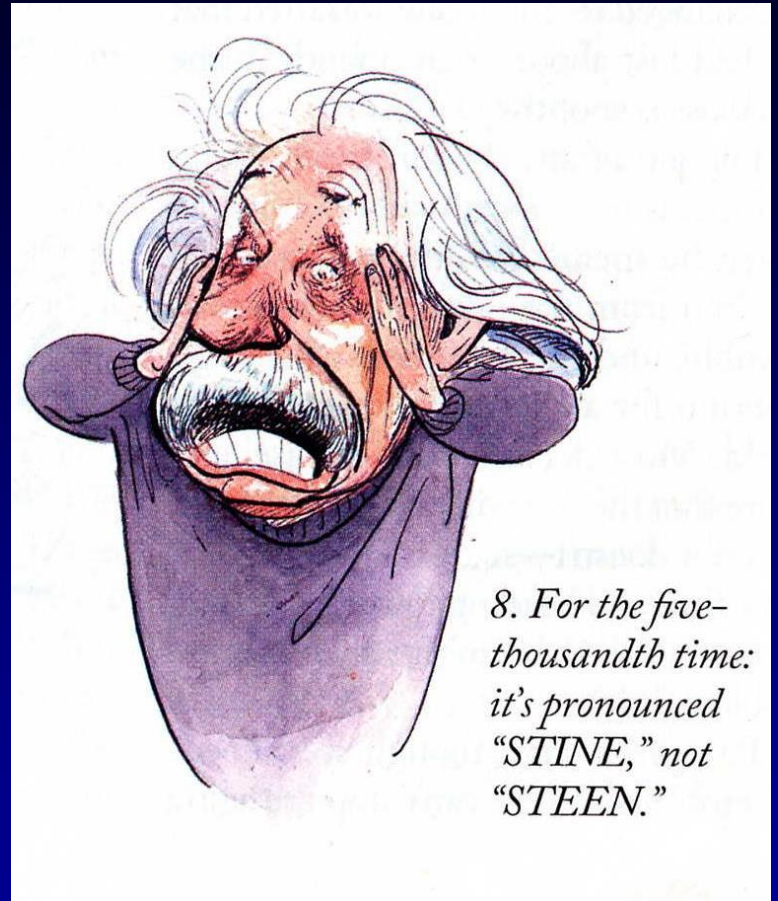
Fig. 1. Wilhelm Ebstein (1836–1912).

J.A.M. van Son et al. / European Journal of Cardio-thoracic Surgery 20 (2001) 1082–1085

WILHELM EBSTEIN.



Wilhelm Ebstein.



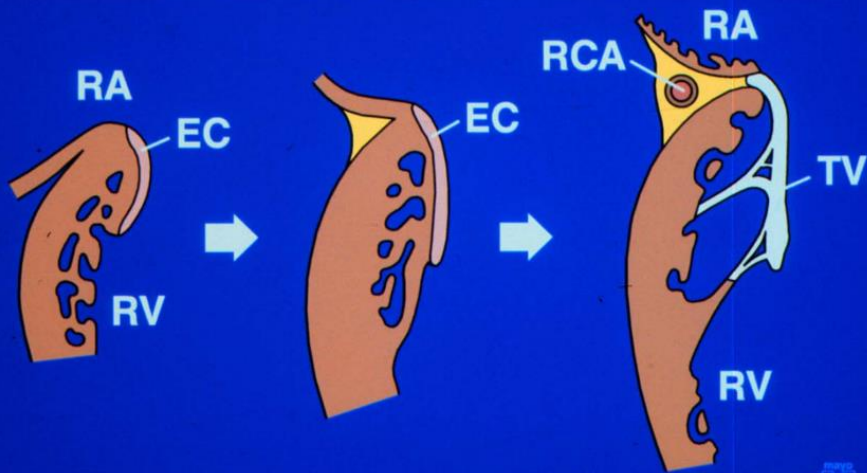
*8. For the five-thousandth time:
it's pronounced
"STINE," not
"STEEN."*

Definition

- **Malformation of Tricuspid Valve (TV) and Right Ventricle(RV)**
- **“Failure of delamination:” Adherence of the septal and posterior leaflets to the myocardium, sail like anterior leaflet**
- **Anterior and apical rotation of the functional orifice and valvar hinge points**
- **Dilated “atrialized” portion of the RV & true TV annulus (TR)**

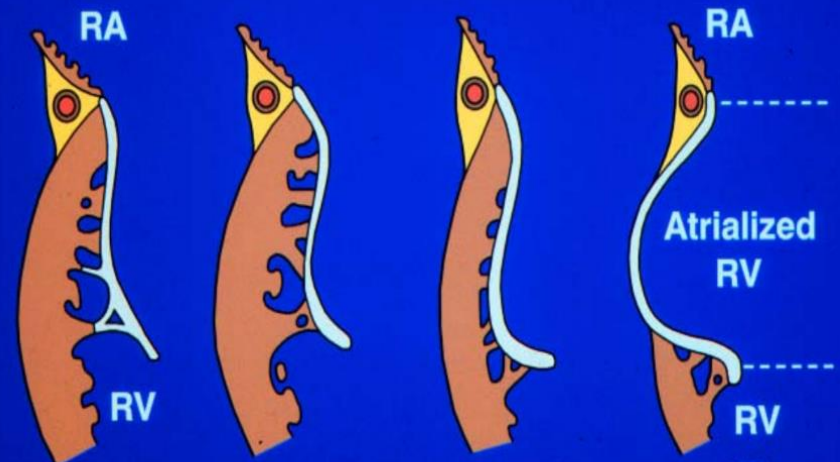
Embryology

Normal



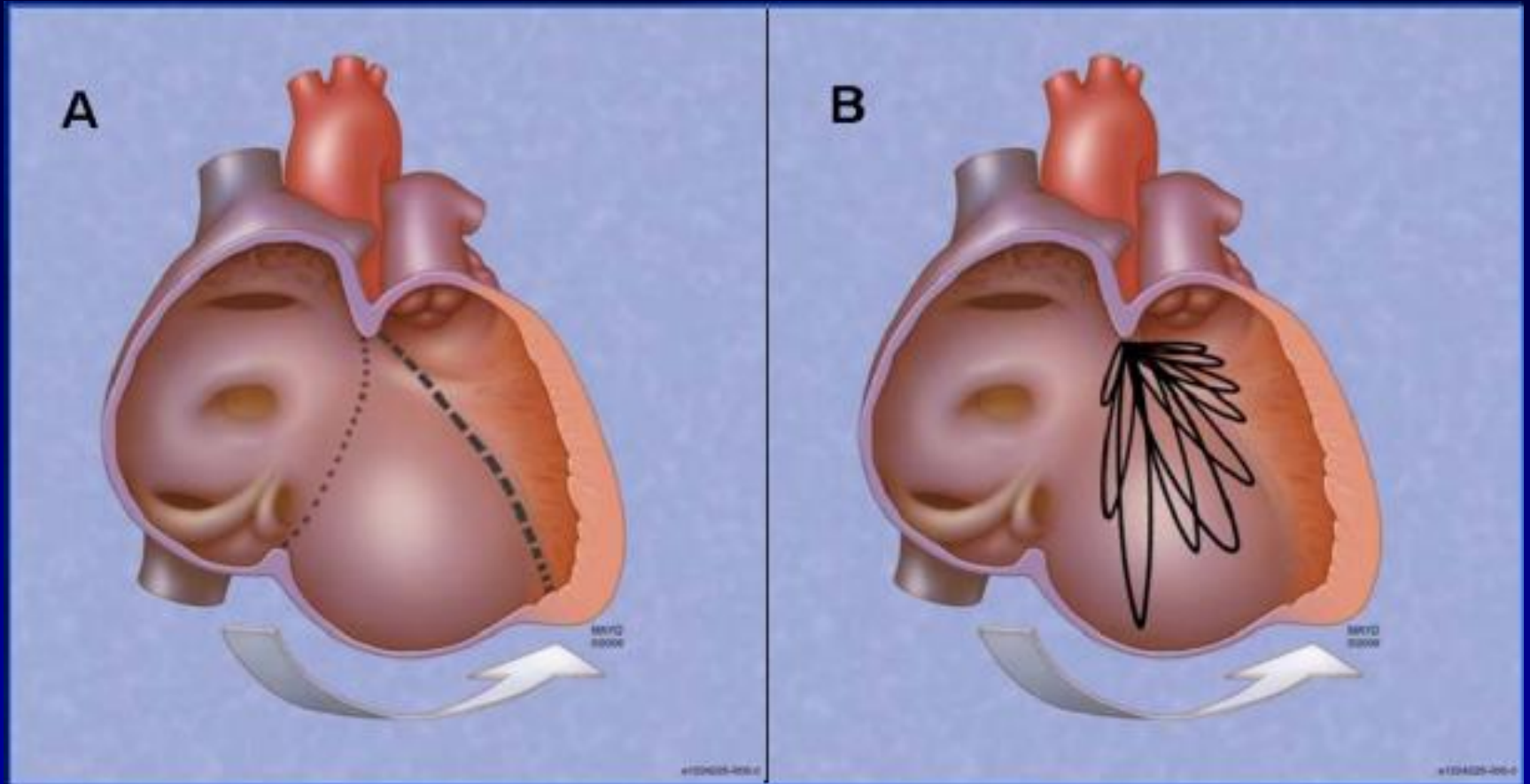
CG150308-01

Ebstein's: Failure of Delamination

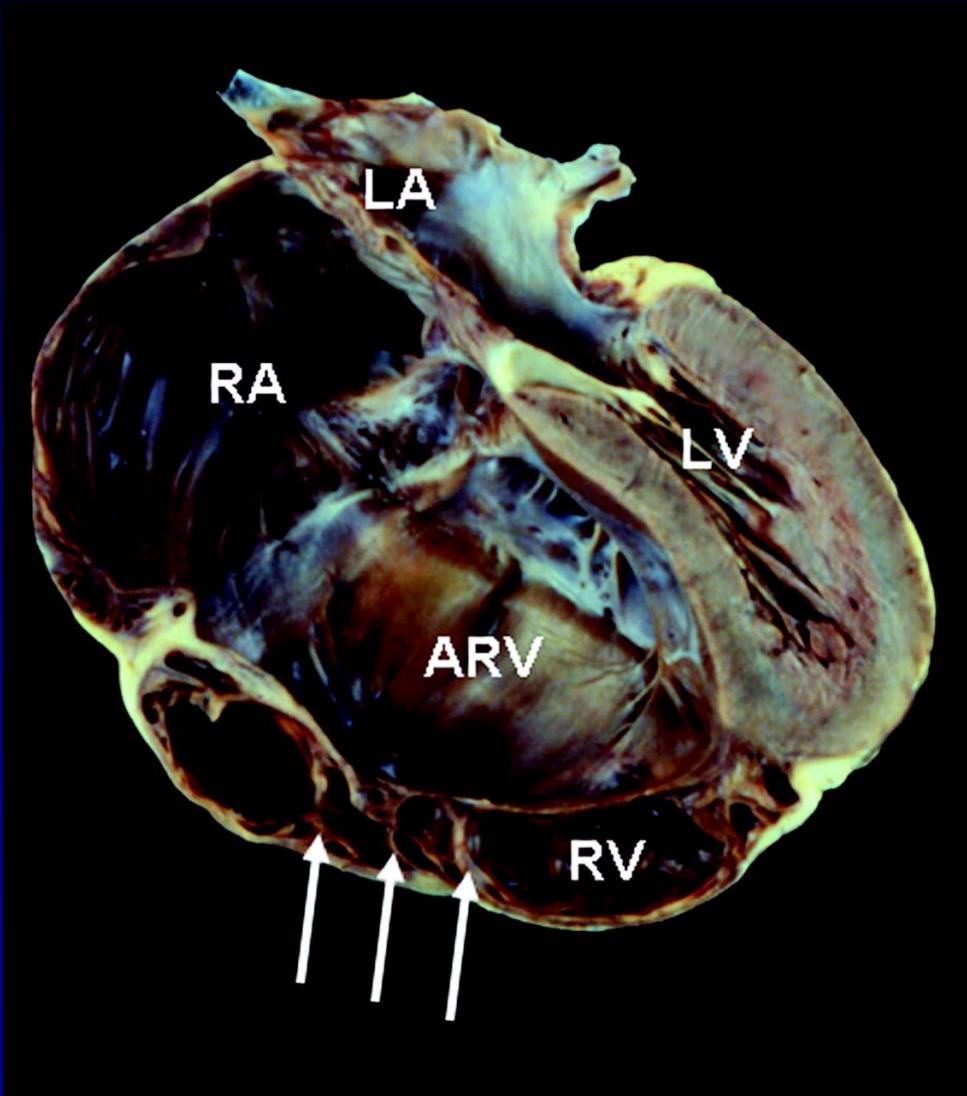


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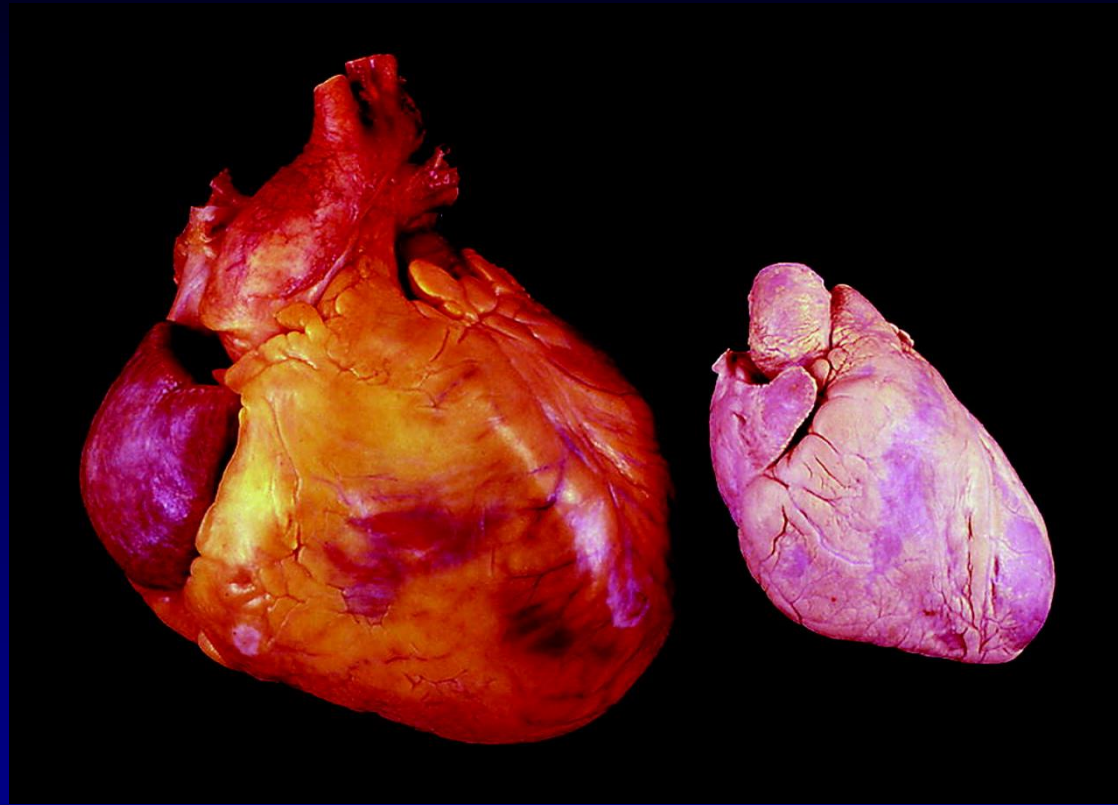
Rotational Displacement of the TV Functional Orifice and Hinges



Source: *Echocardiography in Pediatric and Congenital Heart Disease; From Fetus to Adult* Lai, Mertens, Cohen, Geva



Severe Ebstein's malformation of tricuspid valve (4-chamber view) with marked downward displacement of shelf-like posterior leaflet with attachment to underlying free wall by numerous muscular stumps (arrows), markedly dilated atrialized portion of right ventricle (ARV), small functional portion of right ventricle (RV), leftward bowing of ventricular septum, and marked dilatation of RA

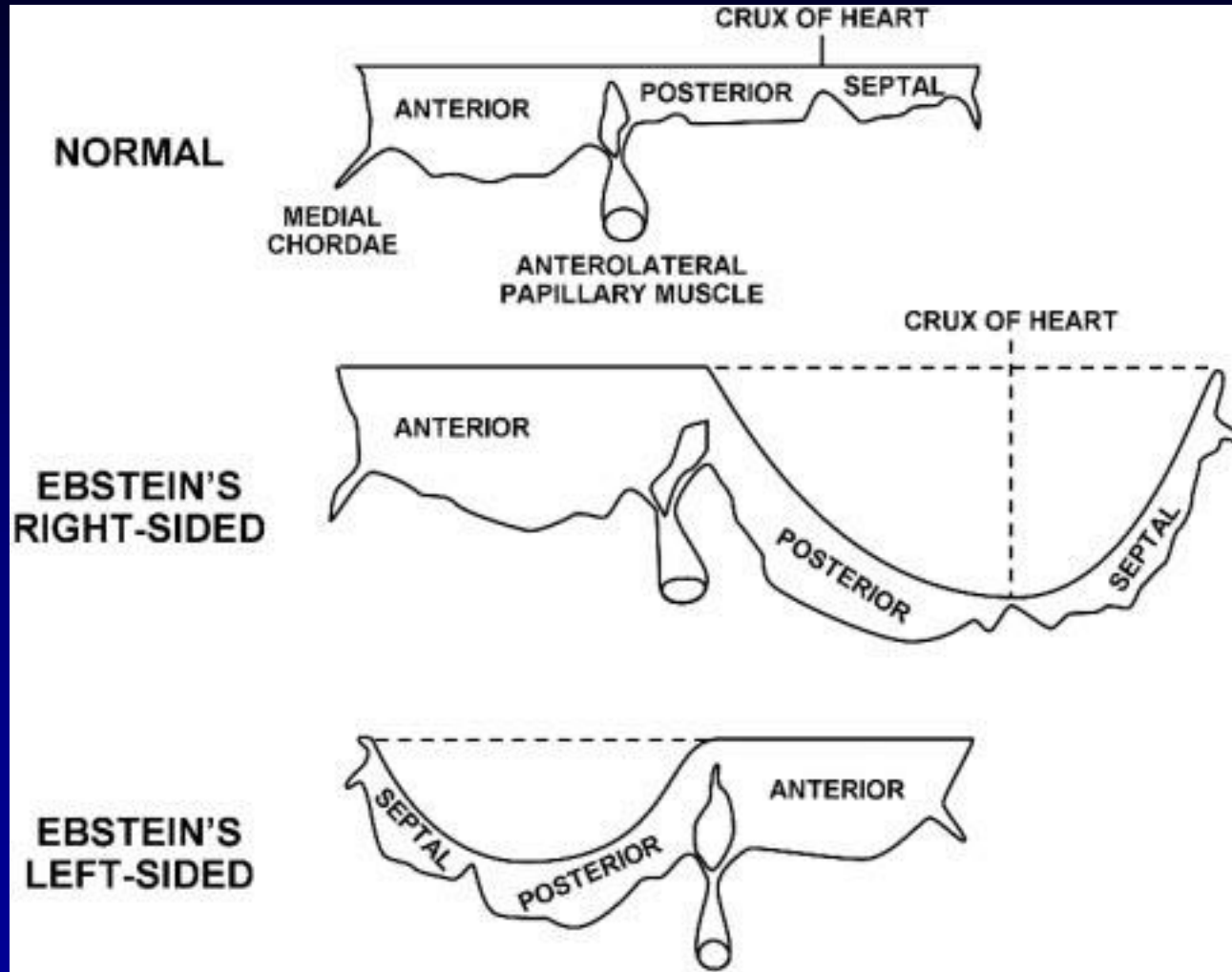


Marked cardiomegaly caused by right-sided chamber dilatation in a 67-year-old man with severe Ebstein's anomaly, with normal heart at right for comparison (anterior view)

Clinical Features in Adults

- When diagnosed in adulthood, symptoms are less severe and dramatic unlike neonatal Ebstein's
- Dyspnea on exertion
- Decrease exercise tolerance
- Arrhythmia
- Symptoms of right heart failure like JVD, pedal edema, hepatomegaly

Imaging the cardiac crux



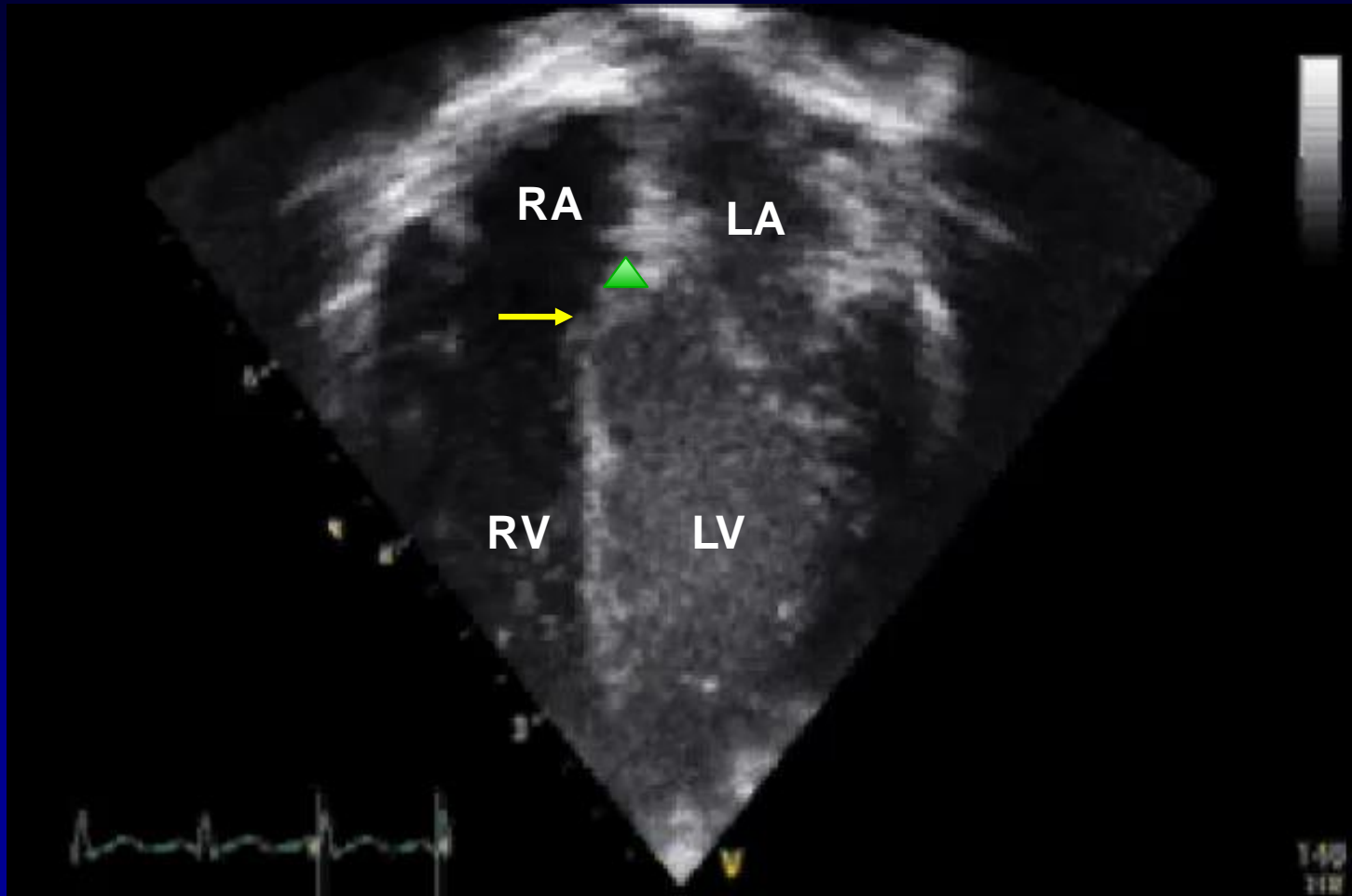
Attenhofer Jost et al

<https://doi.org/10.1161/CIRCULATIONAHA.106.619338> Circulation. 2007;115:277-285

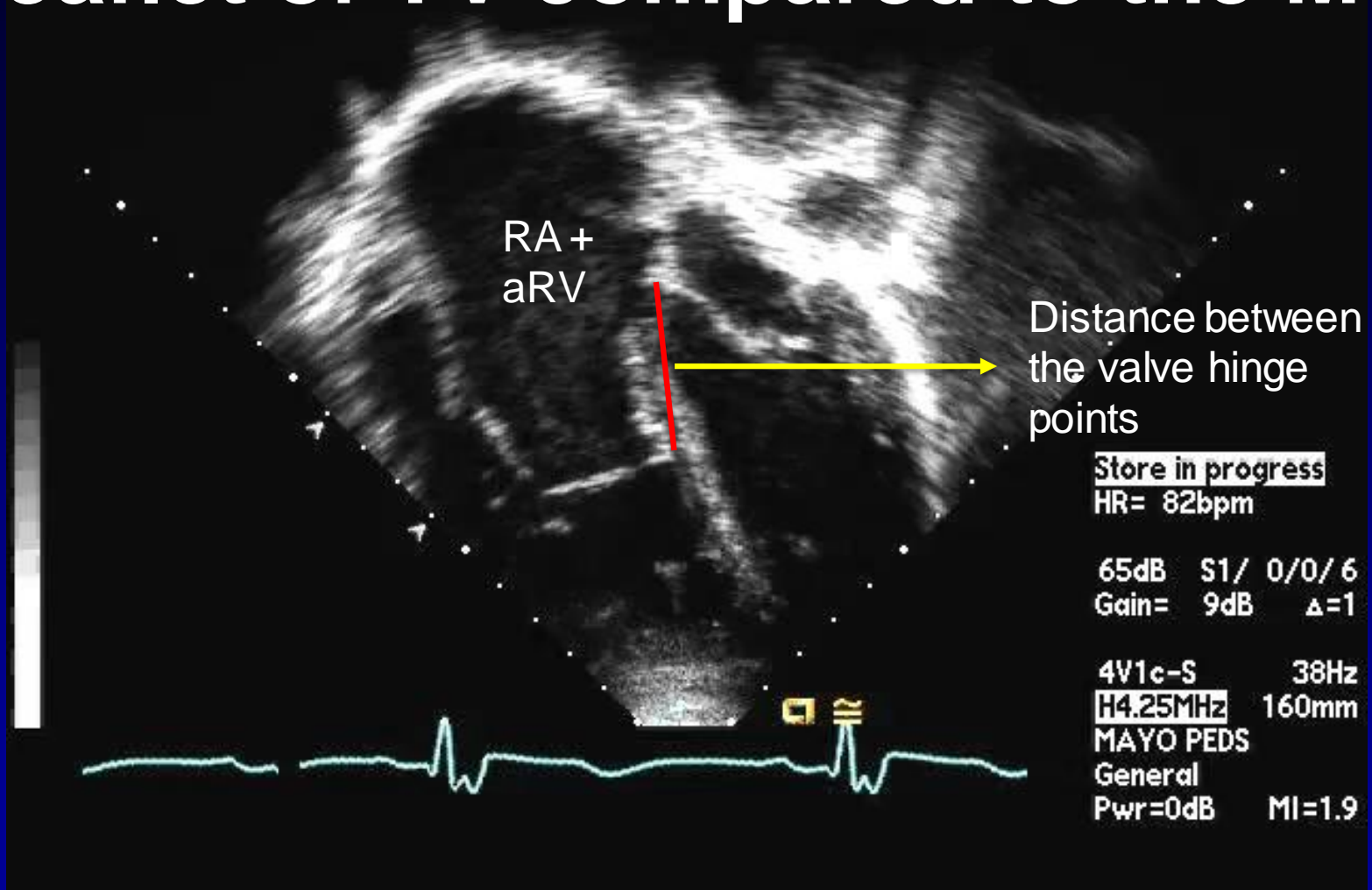
Echocardiographic Recognition of Ebstein's

- Imaging the internal cardiac crux
- Displacement Index= $\frac{\text{Distance b/n valvar hinge points}}{\text{BSA}}$
- Hearts with failed delamination have a delamination index of $> 8\text{mm/m}^2$

Normal Cardiac Crux



Apical displacement of the septal leaflet of TV compared to the MV

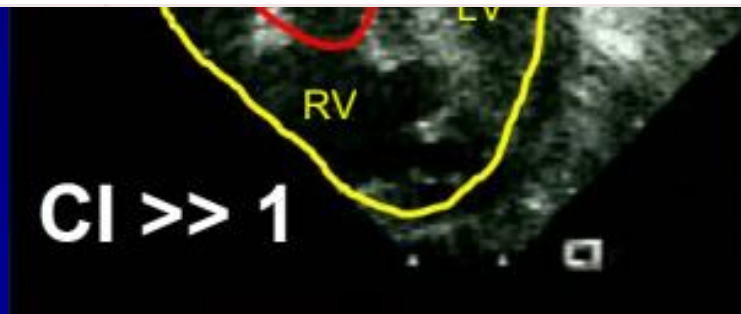
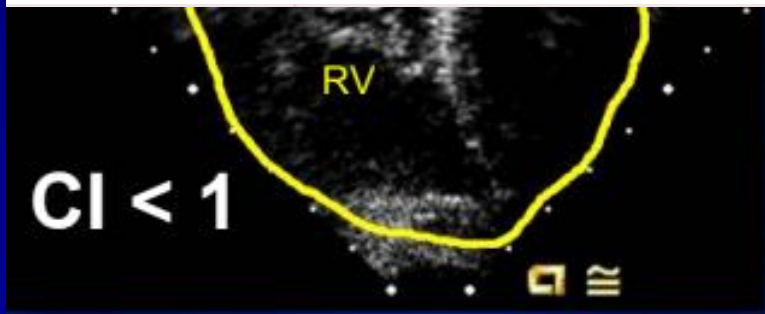


Source: Echocardiography in Pediatric and Congenital Heart Disease; From Fetus to Adult Lai, Mertens, Cohen, Geva

Celemajer Index in Ebstein's Anomaly

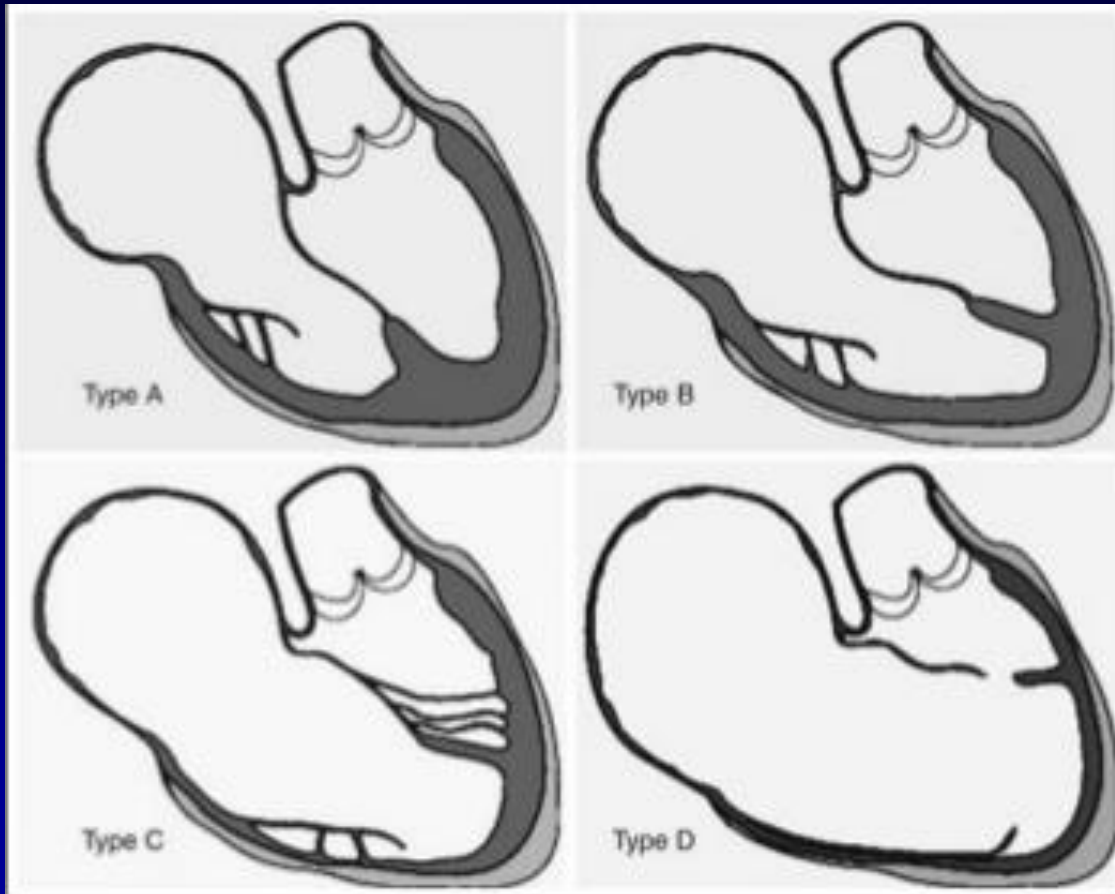


GOSE score	Index (RA + RV): (RV + LA + LV)	Risk of mortality (%)
Grade 1	Ratio < 0.5	0
Grade 2	Ratio of 0.5 to 0.99	10
Grade 3	Ratio of 1 to 1.49	44 - 100
Grade 4	Ratio \geq 1.5	100



Celemajer DS, Cullen S, Sullivan ID, Spiegelhalter DJ, Wyse RK, Deanfield JE. Outcome in neonates with Ebstein's anomaly. **J Am Coll Cardiol.** 1992; 19:1041

Carpentier et al 1988



Type	Description
Type A	RV vol adequate
Type B	RV atrialized, anterior leaflet moves freely
Type C	Severe restriction of anterior leaflet movement, RVOT obstruction
Type D	Near complete atrialization of ventricle

Associated Anomalies

ECHO

- **PFO/ASD (R-L shunt) (commonest)**
- **RV myopathy (sometimes LV can be involved)**
- **RVOT obstruction/ PS**
- **VSD**
- **BAV, MVP, LV abnormalities (non compaction (B myosin heavy chain)**

EKG

- **Accessory pathways**

Repairs

- 1. Monoleaflet repair**
- 2. Cone repair**
- 3. TV replacement**

Indications for Surgical Repair in Adults

- NYHA functional class III or IV
- NYHA functional class I or II with cardiomegaly & CT ratio of ≥ 0.65
- Significant cyanosis and polycythemia/
Paradoxical embolism
- Tachycardia and WPW
- Left ventricular dysfunction

Determining suitability of monoleaflet Repair

- **Mobility of the anterior leaflet**

(At least ½ of the leaflet mobile, free from any tethering)

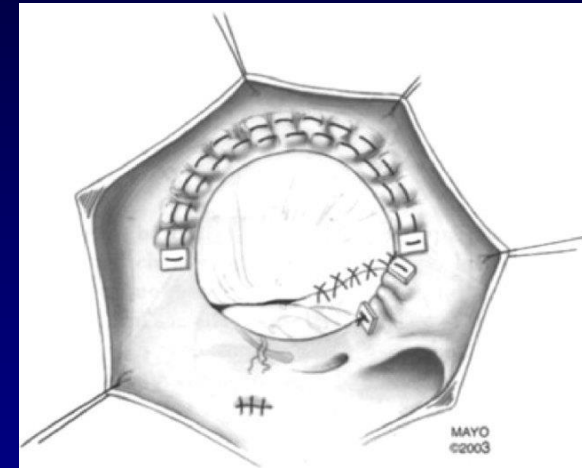
(Apical 4 chamber)

- **Single jet of regurgitation vs multiple**

- (fenestrated leaflets) not suitable (Apical 4, sax)

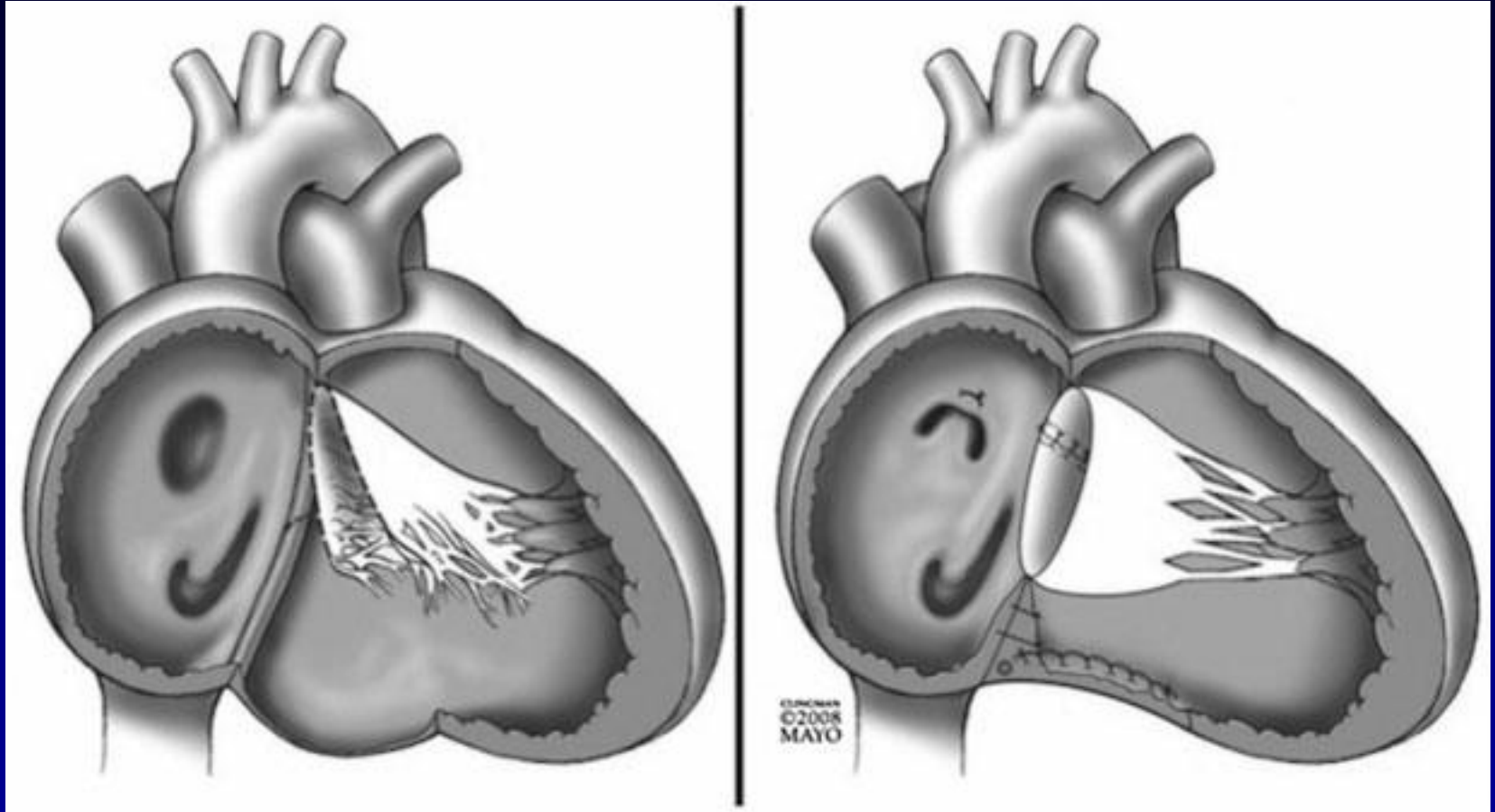
- **Presence of leaflet tissue adjacent to the pulmonary valve (severe displacement)**

(psax)



[Mayo Clin Proc.](#) 1979 Mar;54(3):185-92.
Surgical repair of Ebstein's anomaly.
[Danielson GK](#), [Maloney JD](#), [Devloo RA](#).

Cone Repair



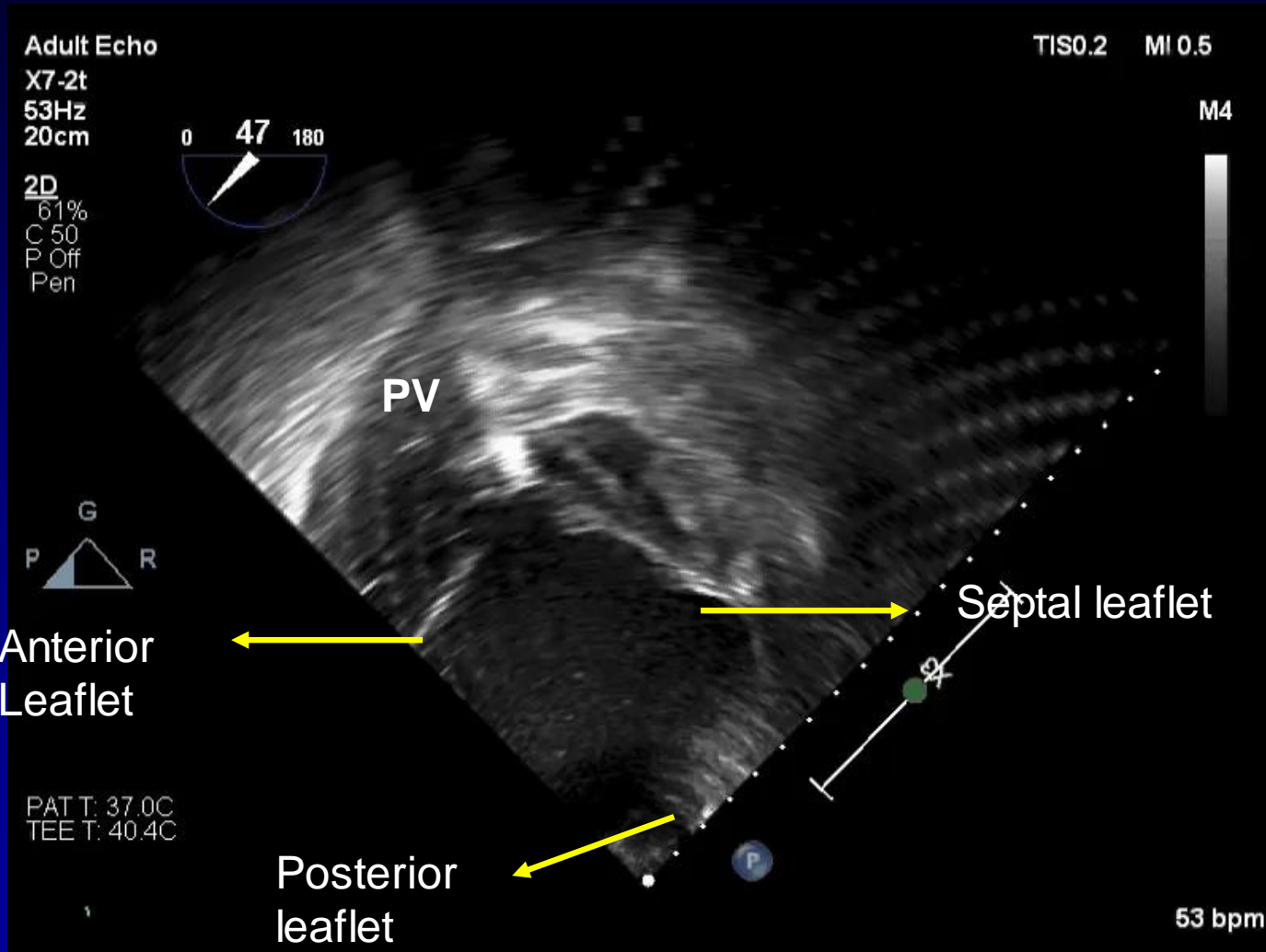
Congenit Heart Dis. 2014;9:266–271

360-degree leaflet tissue repair anchored at true annulus

Case 2

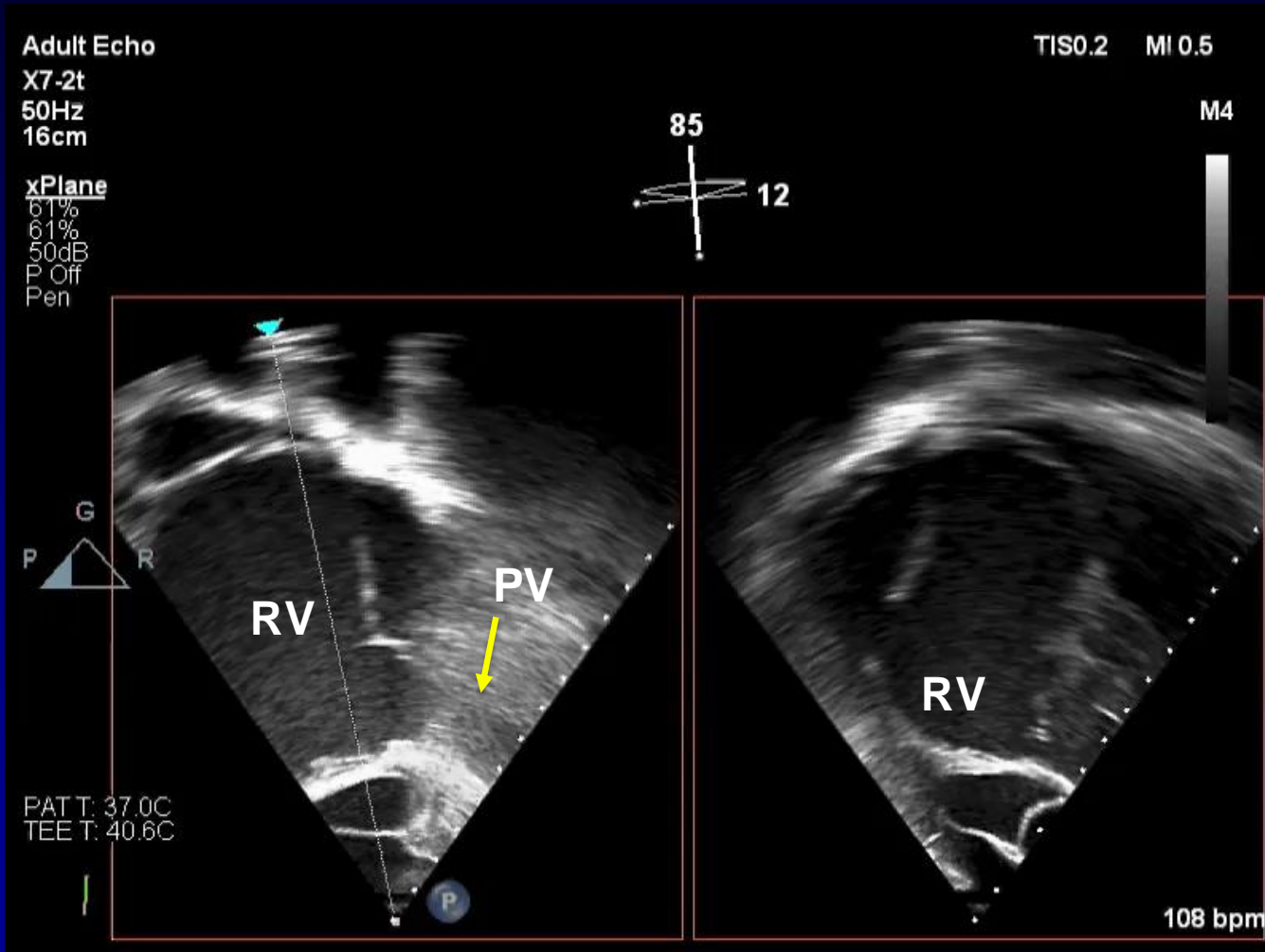
- 33 yo male with Ebstein's anomaly diagnosed as a Child
(very limited records on the severity of Ebstein's)
- Tolerated a secundum ASD Closure with no residual shunt by cath, (mean right atrial pressures of 20, LVEDP of 18 mmHg)
- Paroxysmal Atrial Fibrillation
- Symptomatic with exertional dyspnea and declining functional capacity

Transesophageal echo

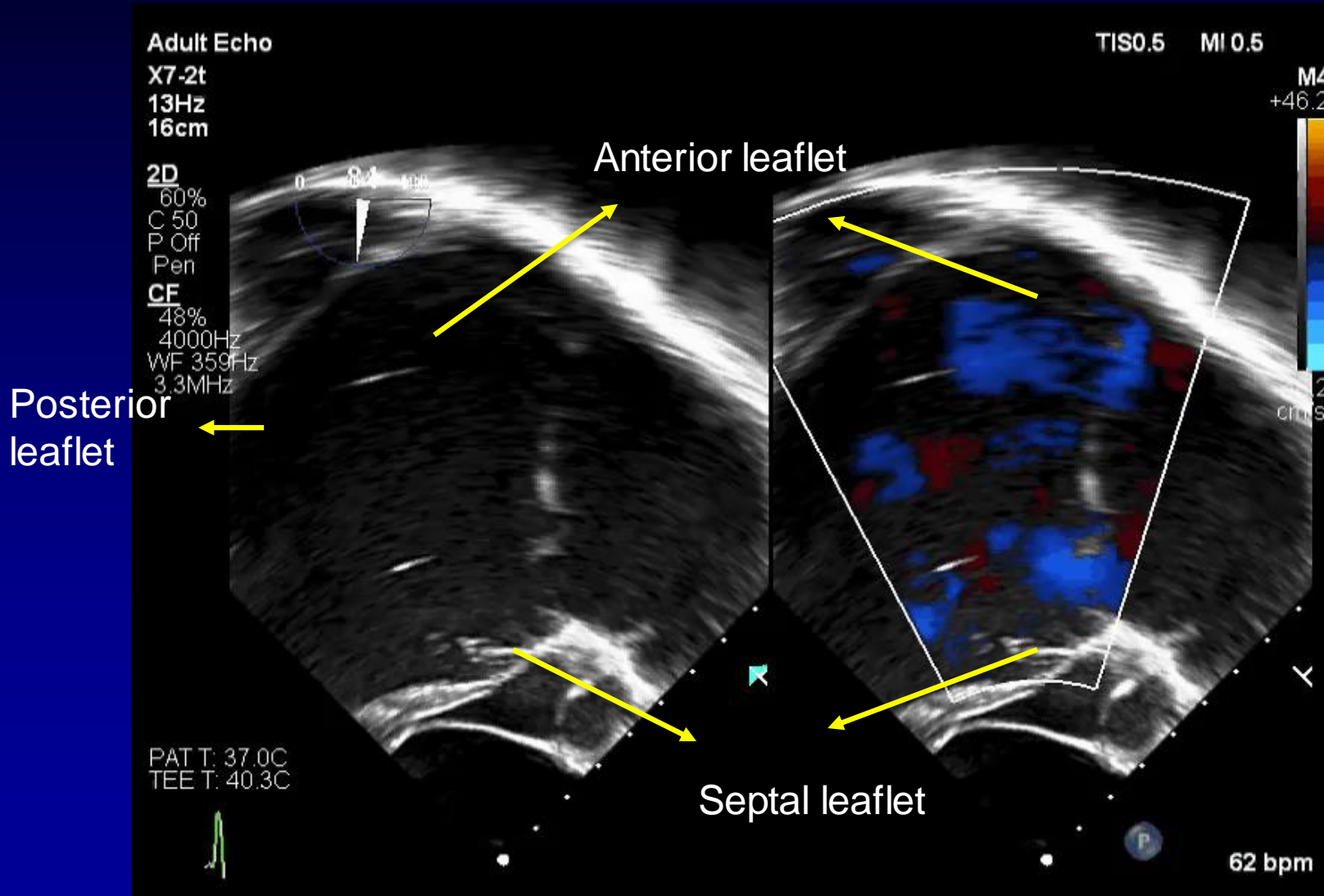


Transgastric RVOT view

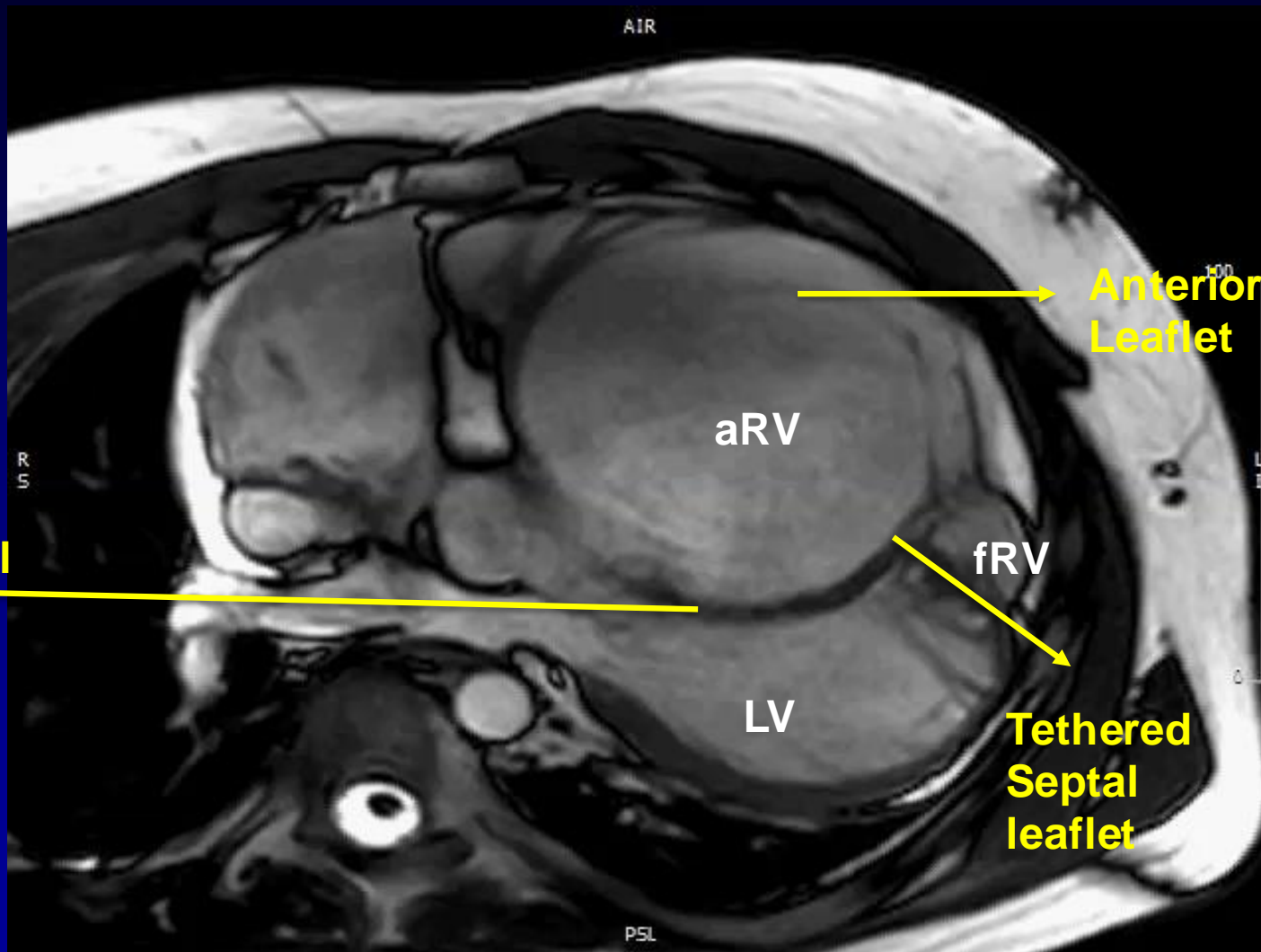
Transesophageal Echo



Transesophageal Echo

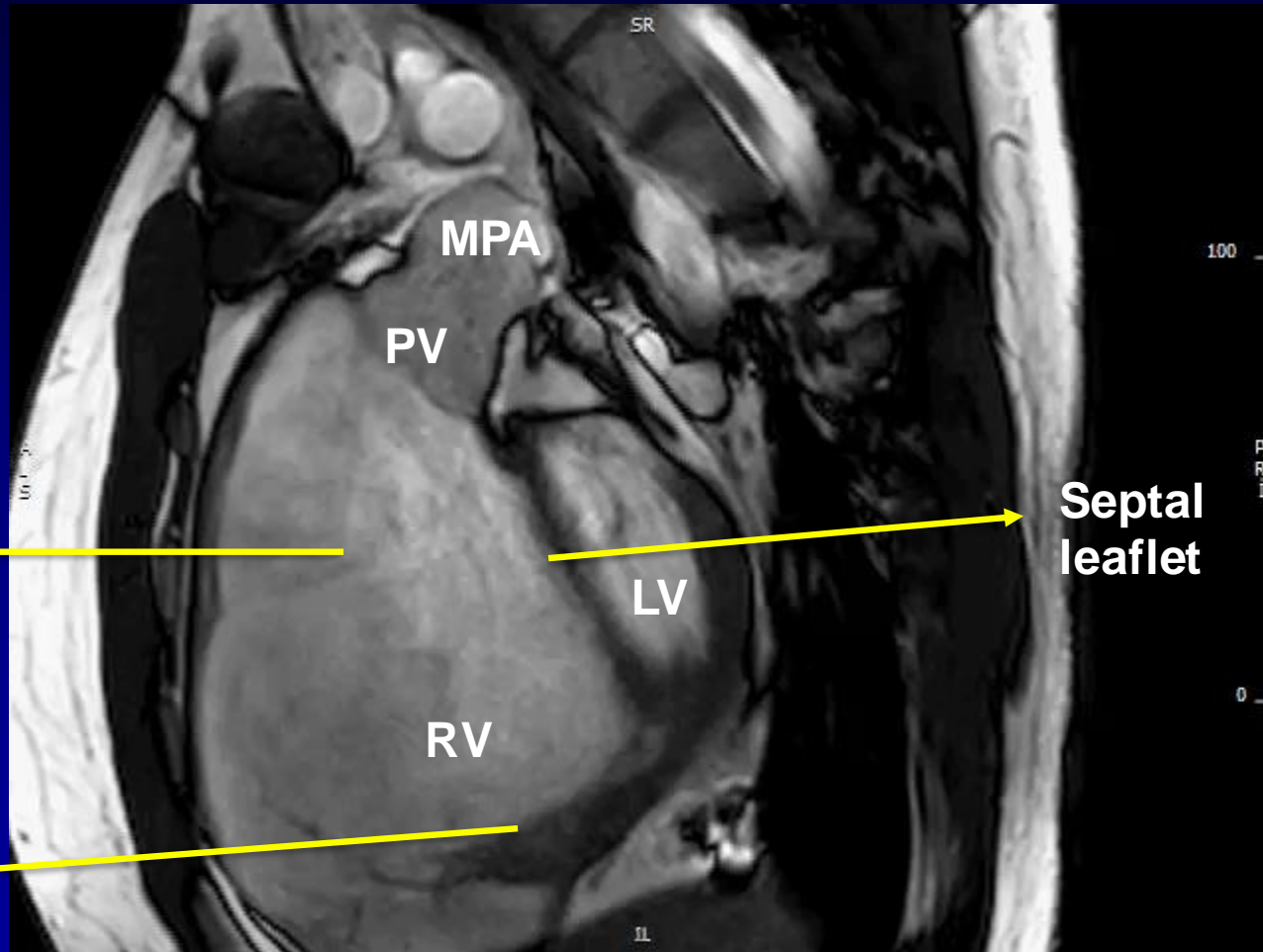


Cardiac MRI



Paradoxical
septal
motion

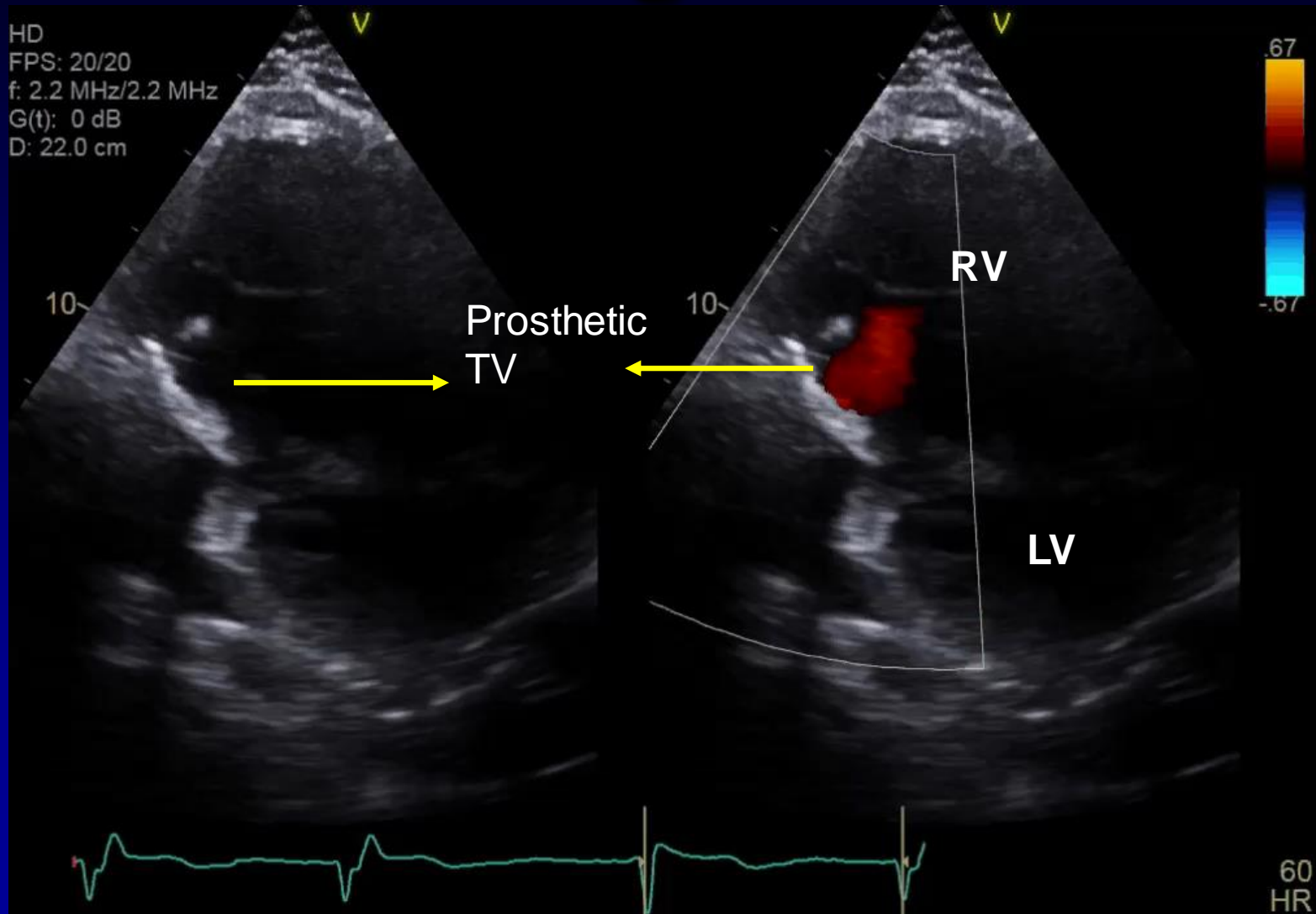
Cardiac MRI



Surgical Repair

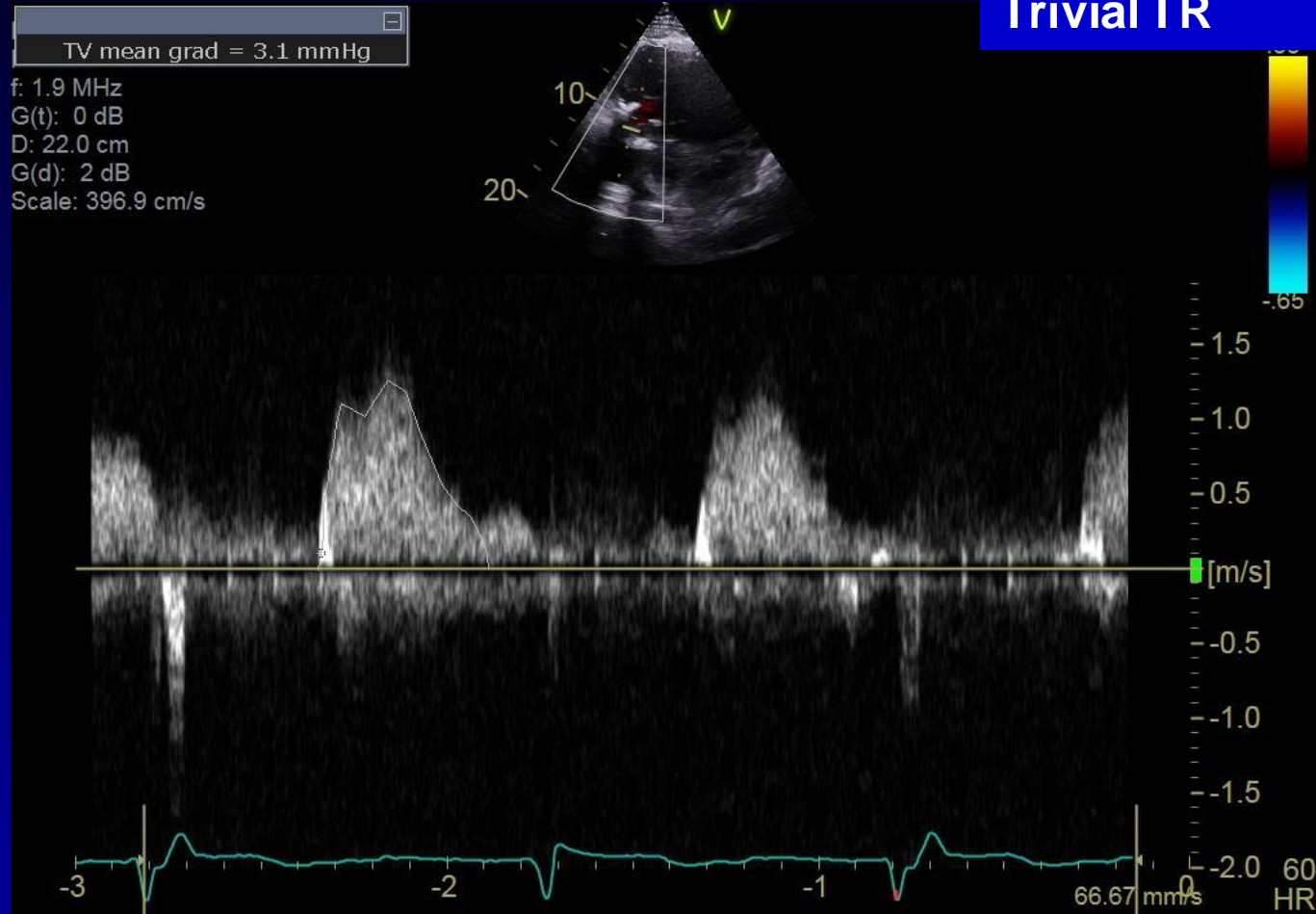
- TV replacement with a #33 Biocor valve
- Right atrial reduction
- Explant of the ASD device
- ASD closure with pericardial patch
- Biatrial cryo MAZE

Post Surgical Echo



No Inflow gradient

Trivial TR



Take home points

- **Adults can surprise you with late onset symptoms**
- **The tricuspid valve has to be imaged in multiple planes to assess each of the leaflets and true extent of TR**
- **Look for abnormal attachments and valvar dysplasia**
- **Look for other associated malformations like ASD and PS**

- **RV function**

Fractional area change, Tei index (MPI)

- **LV function**

M mode, EF



Cleveland Clinic

Every life deserves world class care.