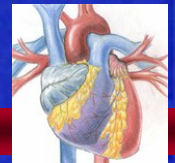
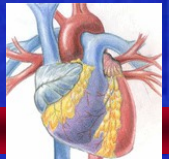
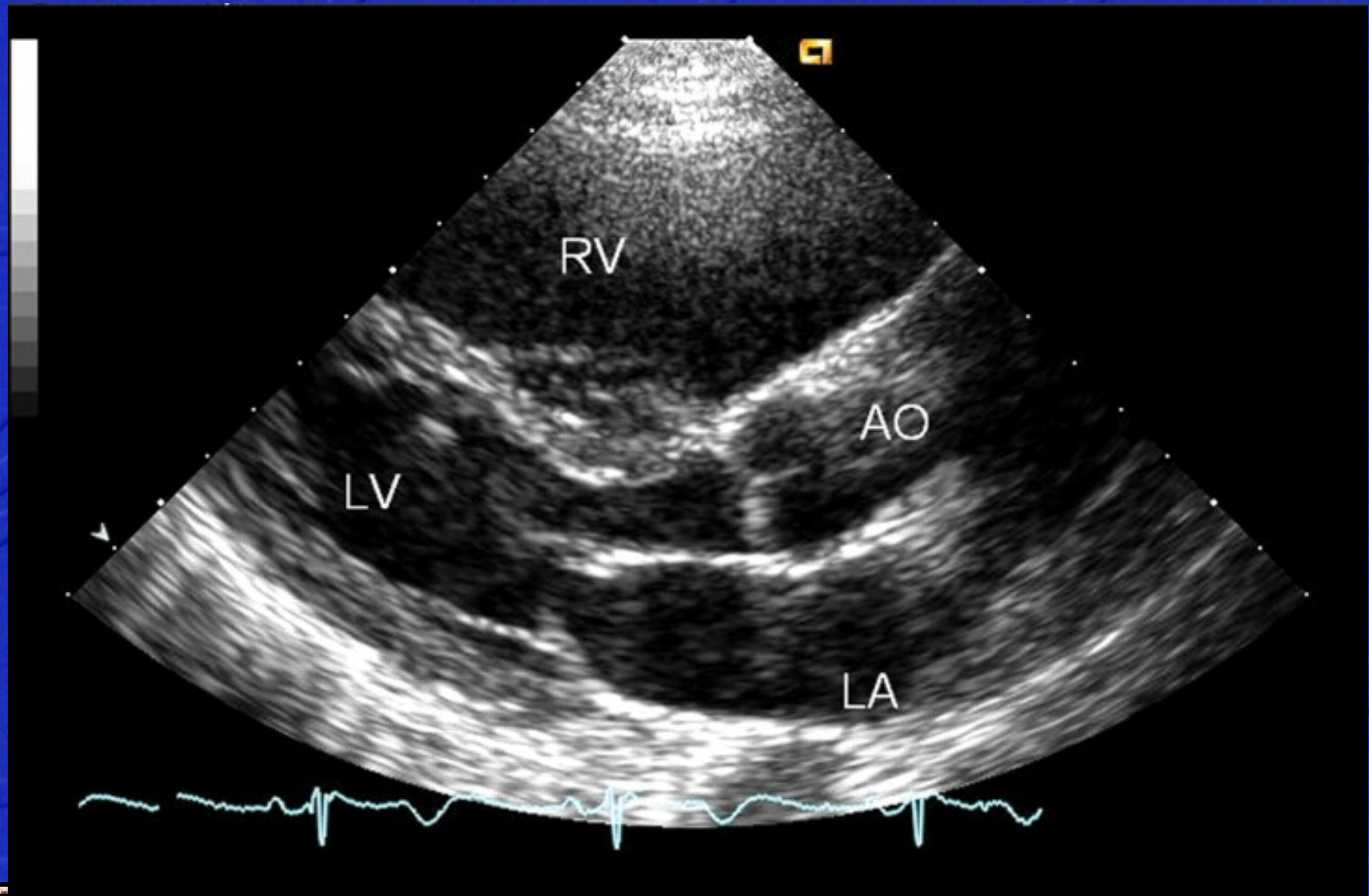


ACHD A Problem Oriented Approach...

P.D. CHD & Invasive Cardiology

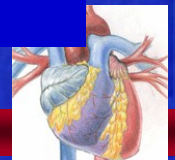




Causes of RV Dilatation

- **RV Volume overload**
RV Contracts well
- **RV Pressure overload**
RV does not contract
But has Hypertrophy!
- **Cardiomyopathy**
RV does not contract
- **RV infarction**
RV does not contract

Guidance™ and a
Motor JPEB II decompressor
is needed to see this picture.



RV Volume overload...

- **Tricuspid regurgitation**

Acquired (structural, functional)

Congenital (Ebstein's)

- **Pulmonary regurgitation**

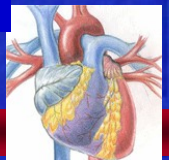
Acquired (structural, functional)

Congenital (Ebstein's)

- **ASD**

Secundum, Primum, Sinus Venosus

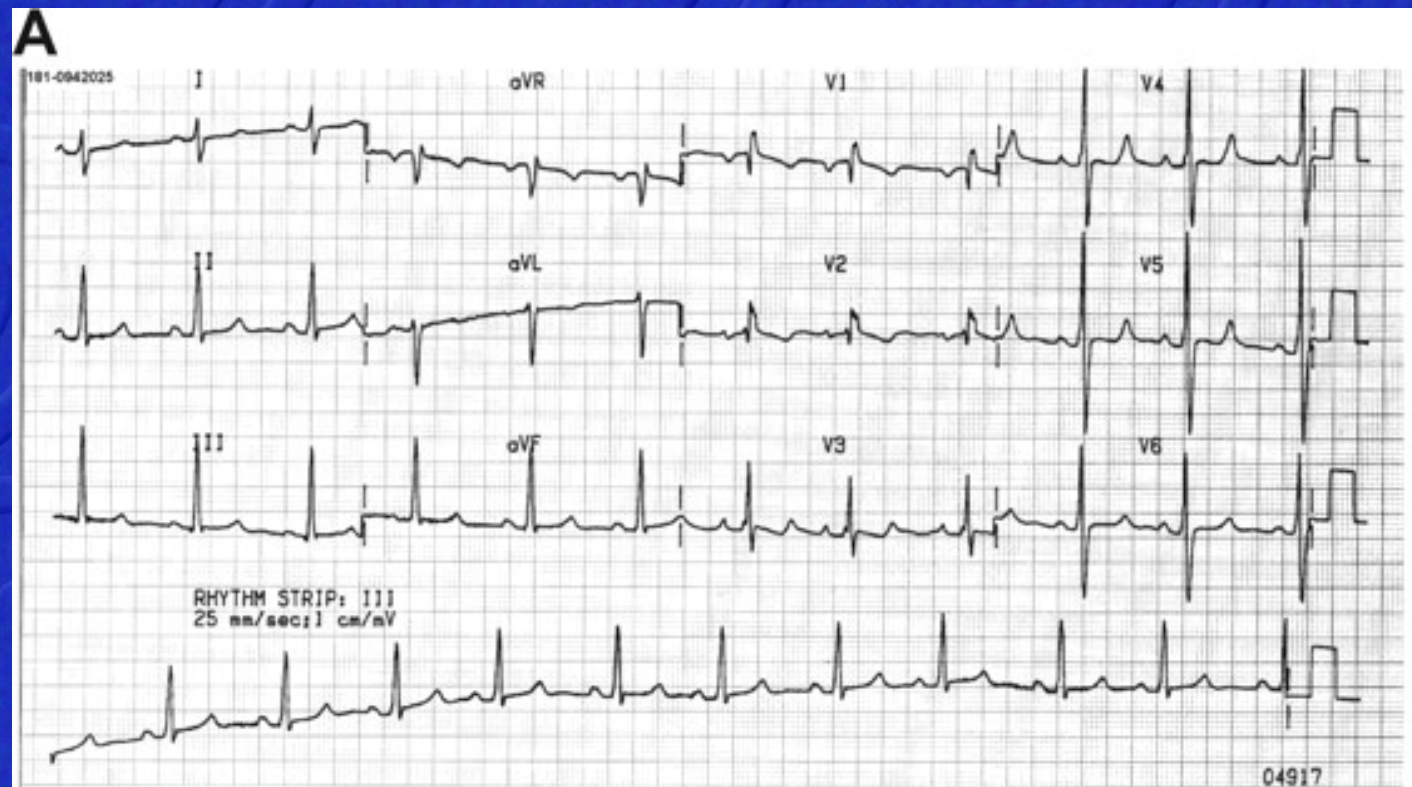
PAPVD



27 yrs old girl...

Mild right-axis deviation, voltage evidence of right ventricular hypertrophy

- Generally asymptomatic
- RV heave
- 2/6 systolic murmur 2nd LICS, fixed splitting S2...
- ECG:



Normal Heart Sounds

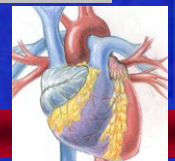
Valve Closure



- **S1** (closure of **Mitral** and **Tricuspid** valves)
- **S2** (closure of the **Aortic** and **Pulmonic** valves)
- High pitched: heard best with the Diaphragm
- Which is S1? S2? (location where best heard, rhythm, timing compared to the carotid pulse and JVP waves)

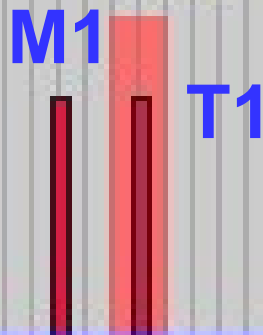


EMORY



LUB - S1

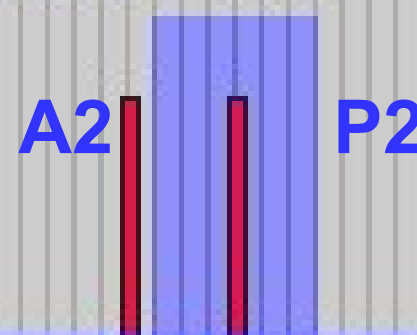
Normal Range
0.02 - 0.04



Tricuspid
Mitral

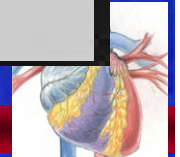
DUB - S2

Normal Range
0.01 - 0.07



Aortic
Pulmonic

0.01 Seconds Between Marks



LUB - S1

Normal Range

0.02 - 0.04

Seconds

M1

T1

Tricuspid

Mitral

LUB - S1

Minimum Audible

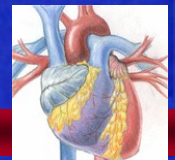
Split

0.2 Seconds

Tricuspid

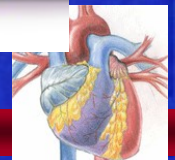
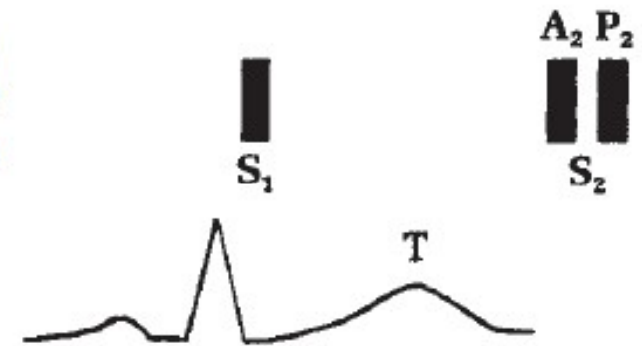
Mitral

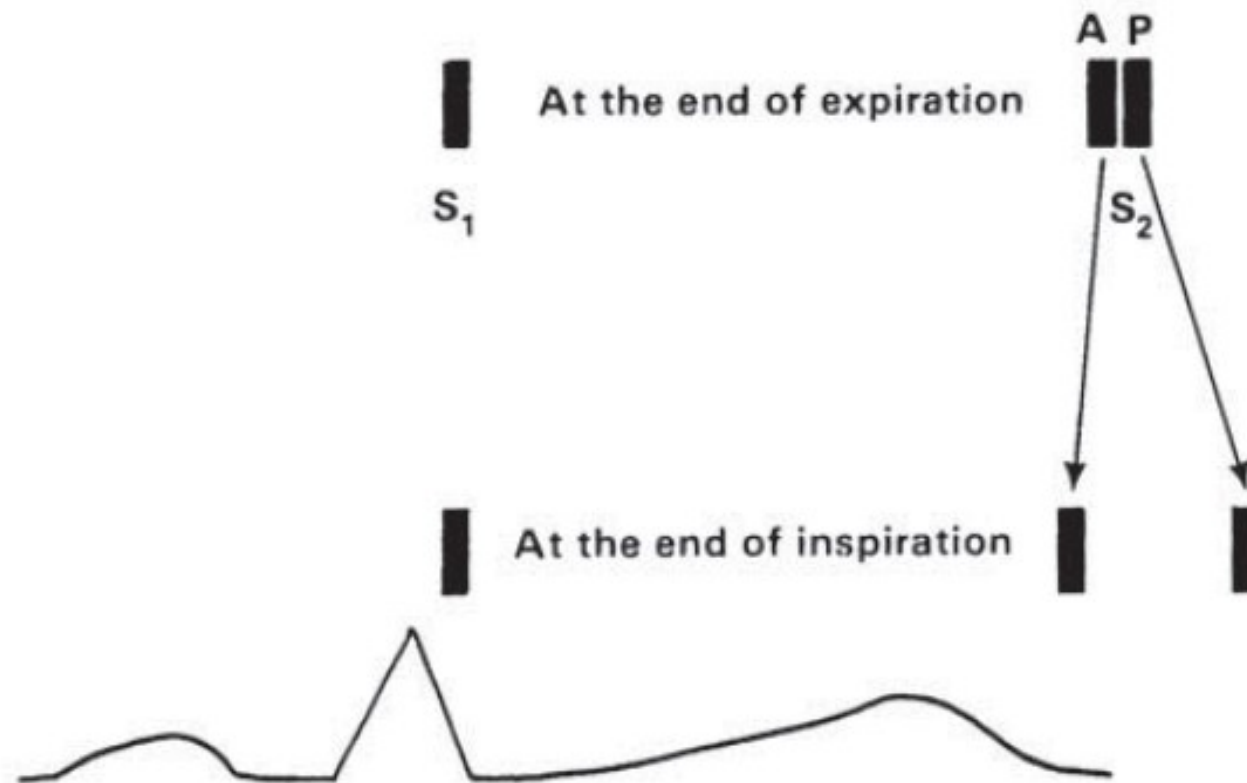
0.01 Seconds Between Marks



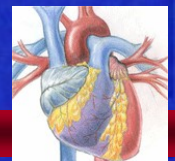
ΔΕΥΤΕΡΟΣ ΤΟΝΟΣ S2: A2-P2

A₂ is the aortic valve closure component of the S₂.
P₂ is the pulmonary valve closure component of the S₂. Note that the S₂ occurs near the end of the T wave of the ECG; i.e., the T wave is a systolic event.



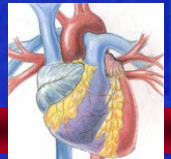


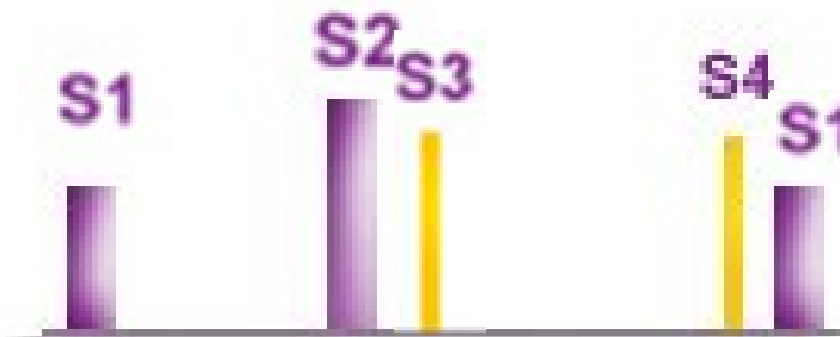
The P₂ outward movement contributes more to the inspiratory widening of the S₂ than does the inward movement of the A₂.



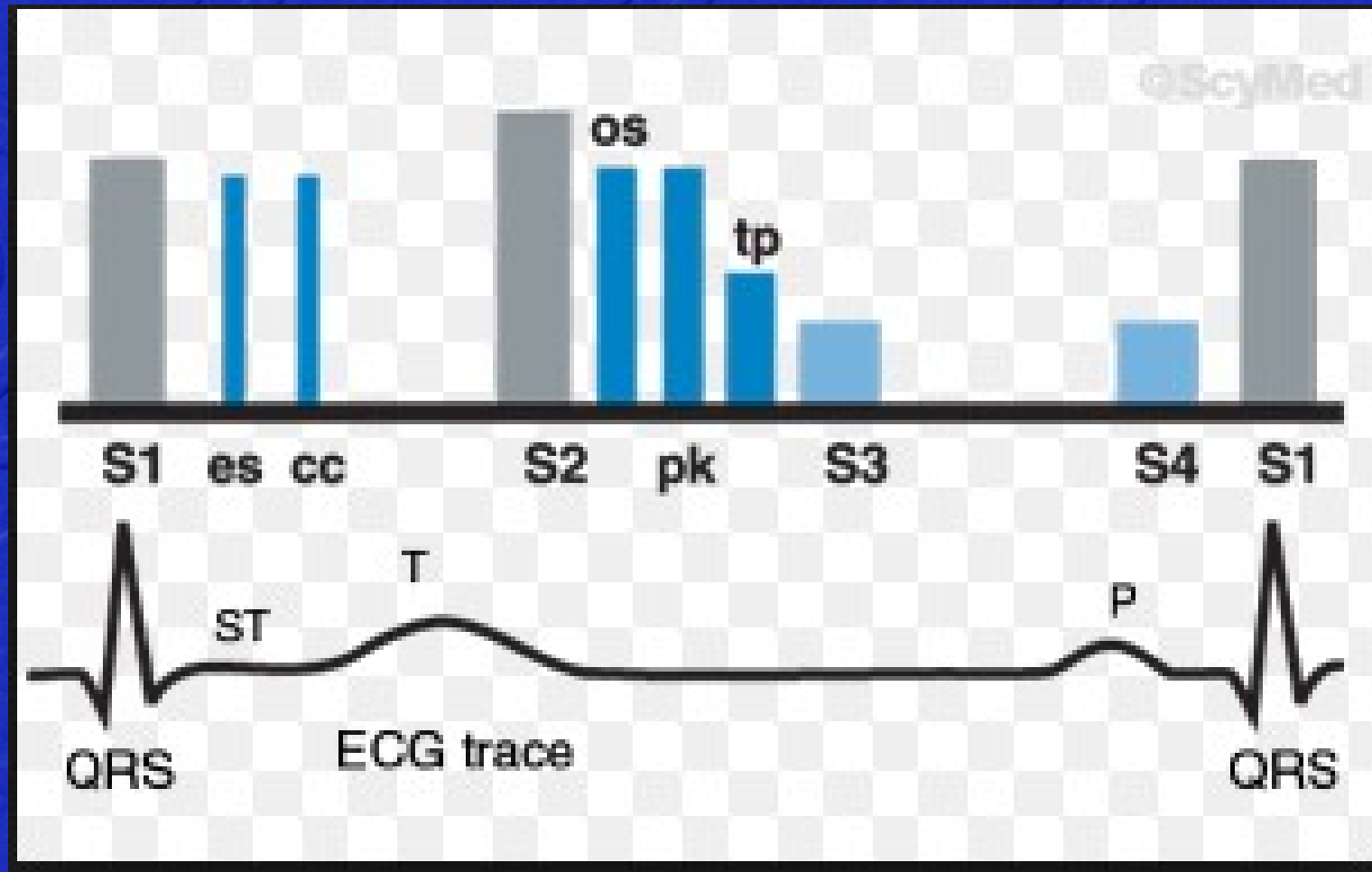
ΦΥΣΙΟΛΟΓΙΚΟΣ ΔΙΧΑΣΜΟΣ S_2

- A2 & P2, συμπίπτουν με την δικροτική εγκοπή της αντίστοιχης αρτηρίας
- Κατά την εισπνοή καθυστερεί ο P2 και ο S2 διχάζεται φυσιολογικά (μικρότερη αντίσταση στην πνευμονική αρτηρία αυξημένο hangout interval). 2ο LICs.





Πρόσθετοι τόνοι...



ES: ejection sound, CC: Click, OS: opening snap, PK: pericardial knock, TP: tumor plop



Διαταραχές S_2

- Άυξηση έντασης A_2 (υπέρταση, ατρησία πνευμονικής, πλήρης μετάθεση, διάταση αορτής)
- Άυξηση έντασης P_2 (πνευμονική υπέρταση, διάταση πνευμονικής όπως στο ASD)

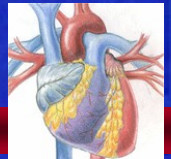
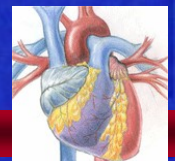


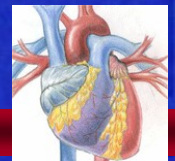
TABLE 31.3. Factors Affecting Intensity of First and Second Heart Sounds

	S_1	S_2	
		A_2	P_2
Increased	<ul style="list-style-type: none"> PR <160 ms Mitral stenosis with pliable valve Hyperdynamic states Holosystolic MVP Rapid heart rates 	<ul style="list-style-type: none"> Systemic HTN Hyperdynamic states Aortic dilation 	<ul style="list-style-type: none"> Pulmonary HTN Atrial septal defect
Decreased	<ul style="list-style-type: none"> PR >200 ms Poor LV systolic function Mitral stenosis with rigid valve LBBB Acute aortic regurgitation 	<ul style="list-style-type: none"> Calcific aortic stenosis Aortic regurgitation 	<ul style="list-style-type: none"> Pulmonic stenosis

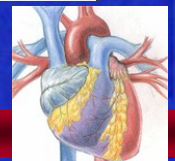
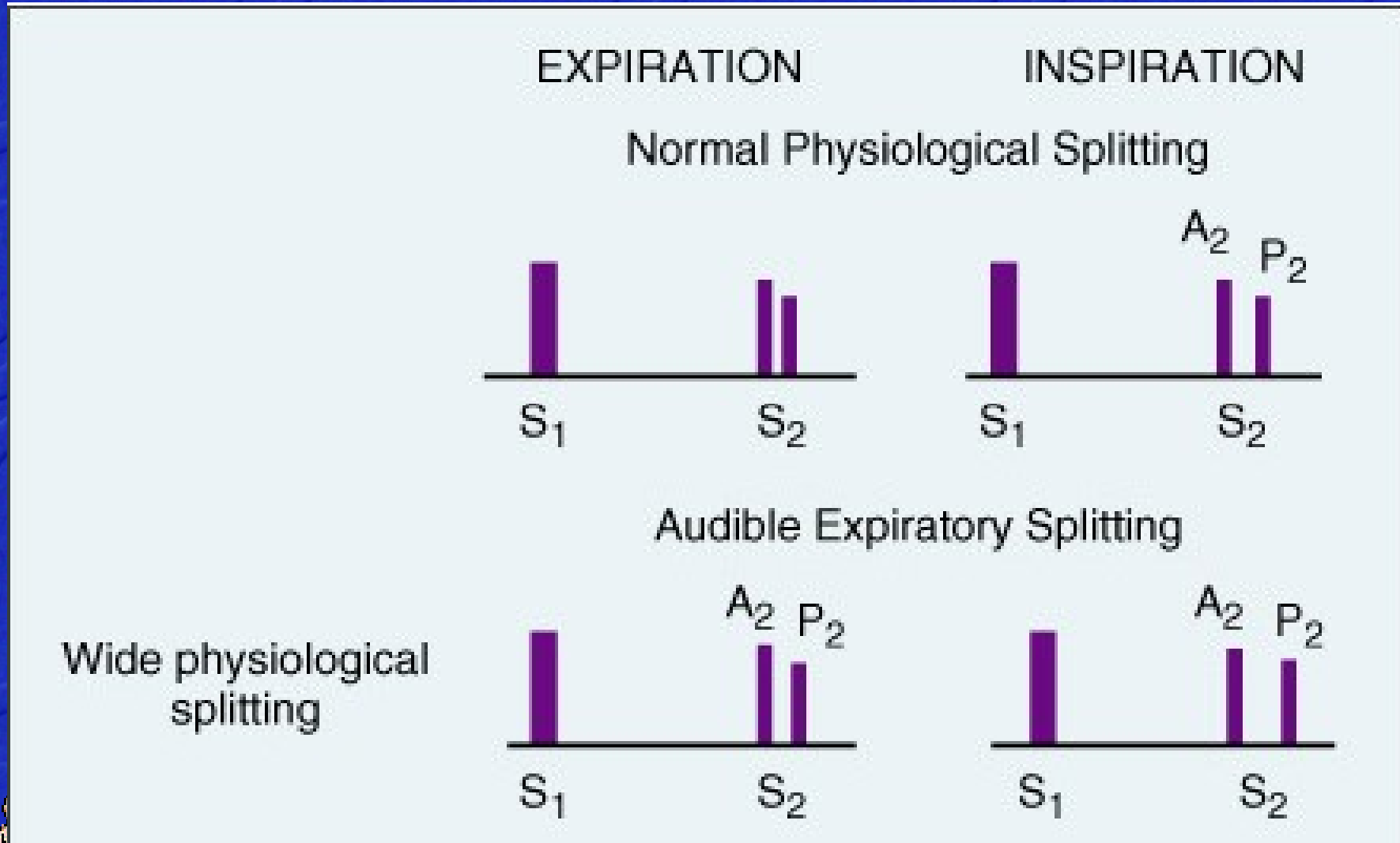


Διαταραχές S_2

- **Ανώμαλος διχασμός:**
 - Μόνιμα διχασμένος S_2 , αλλά με αναπνευστική διακύμανση: (RBBB, MR)
 - Fixed Splitting: (ASD... ATRIAL SEPTAL DEFECT)
 - Paradoxical Splitting: (LBBB, βηματοδότης)



Διαταραχές S_2



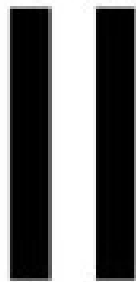
ΠΑΡΑΔΟΞΟΣ διχασμός S_2

EXPIRATION

S_1



S_2



P_2 A_2

INSPIRATION

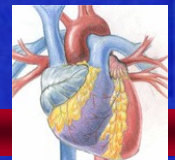
S_1



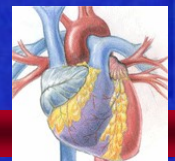
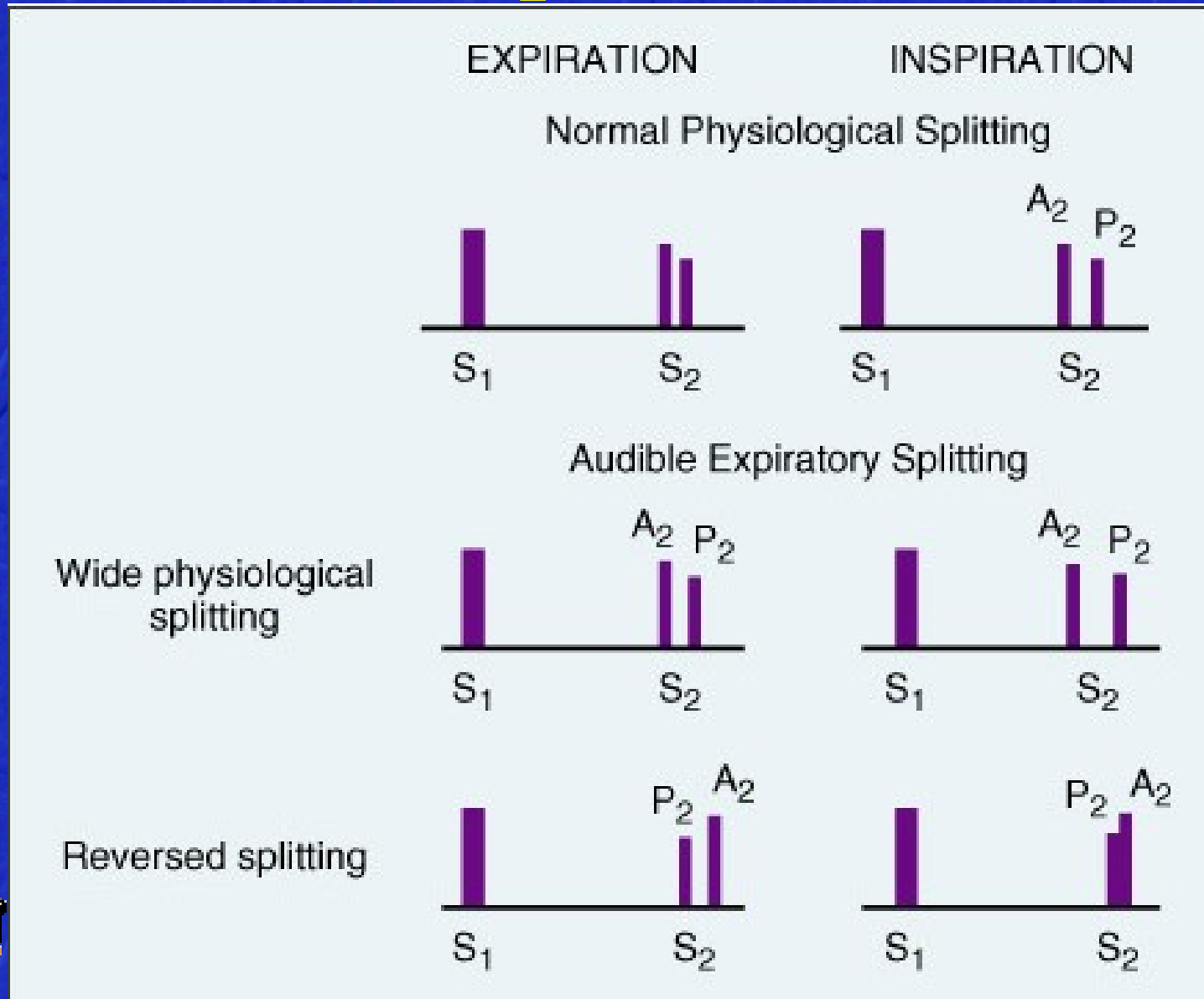
S_2



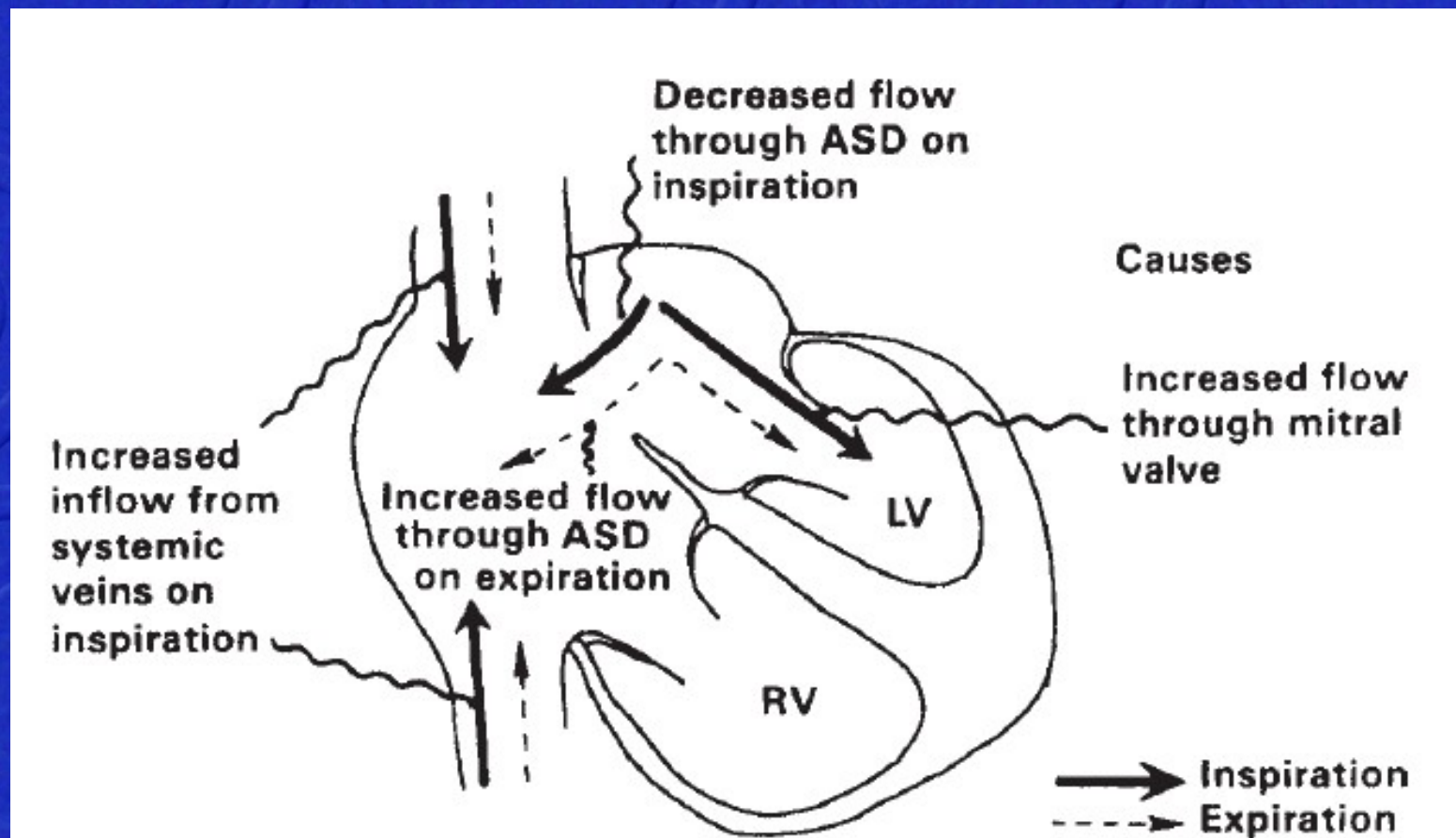
$A_2 - P_2$



Διαταραχές S_2

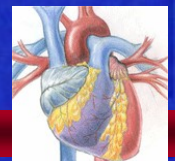
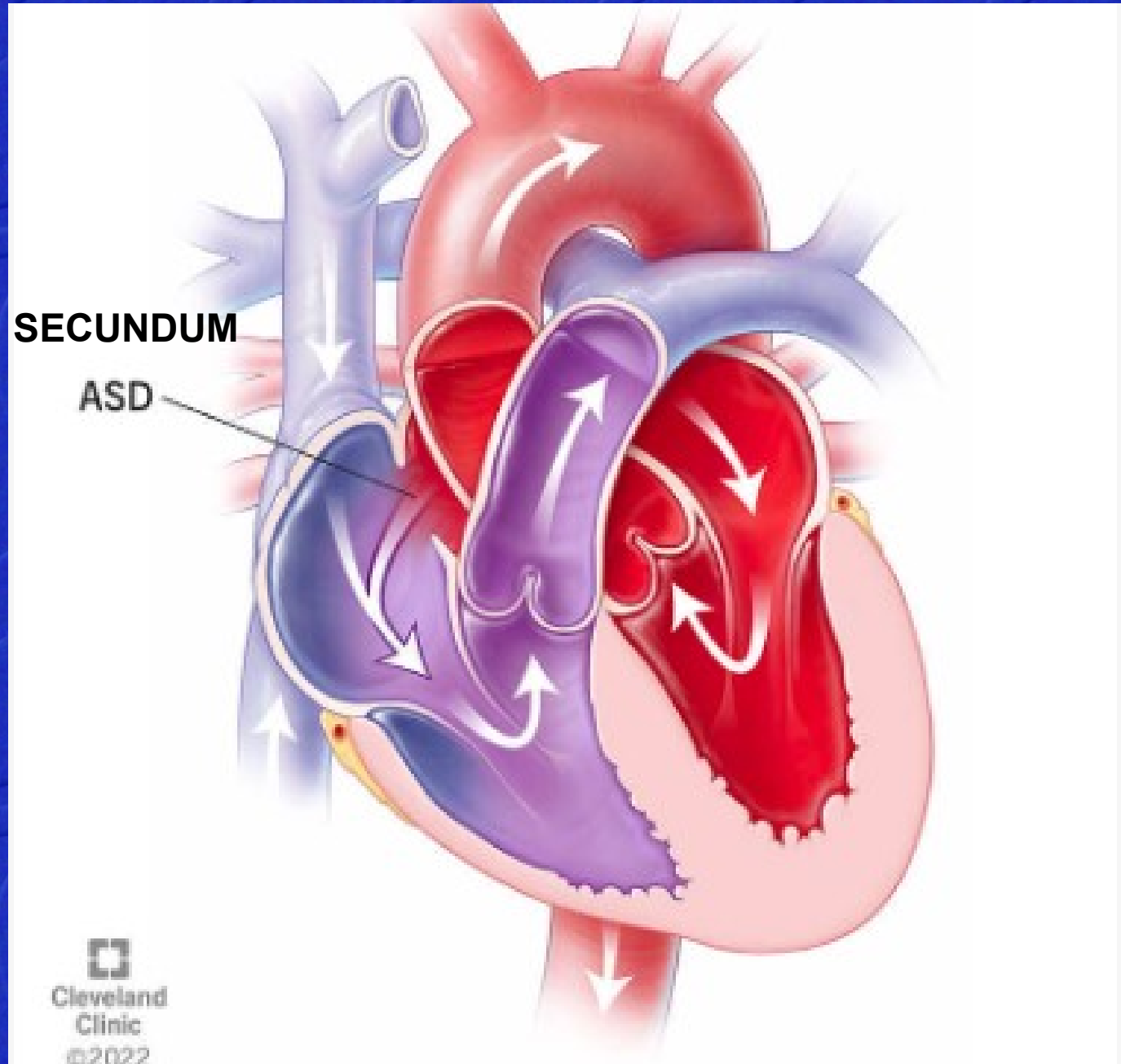


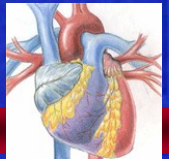
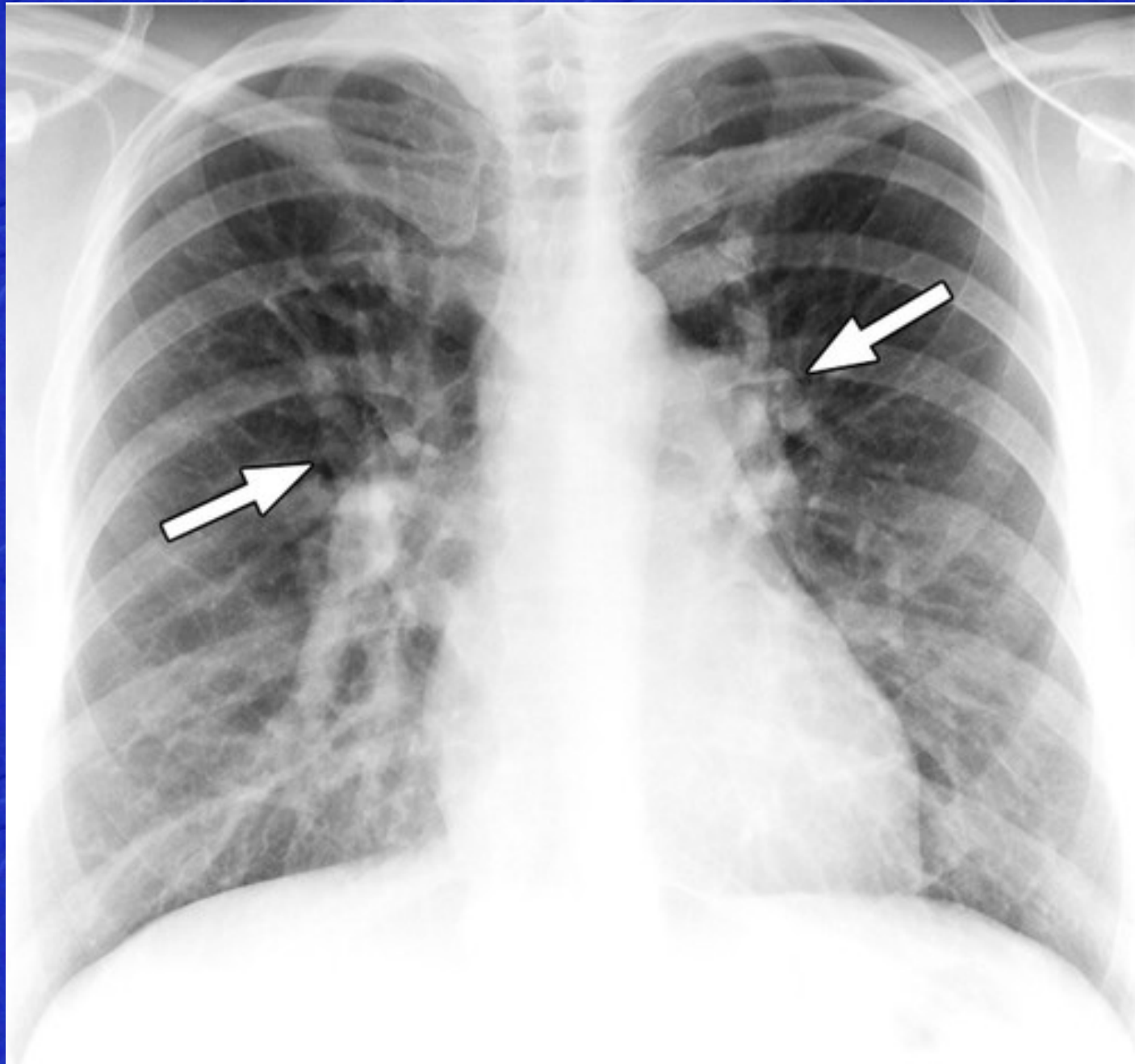
ASD: ΣΤΑΘΕΡΟΣ ΔΙΧΑΣΜΟΣ S₂



The increased inflow into the right atrium on inspiration (vertical solid arrows) causes a decreased flow through the ASD and thus increased flow through the mitral valve.

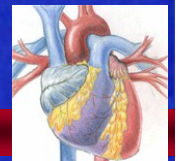






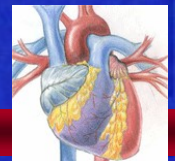
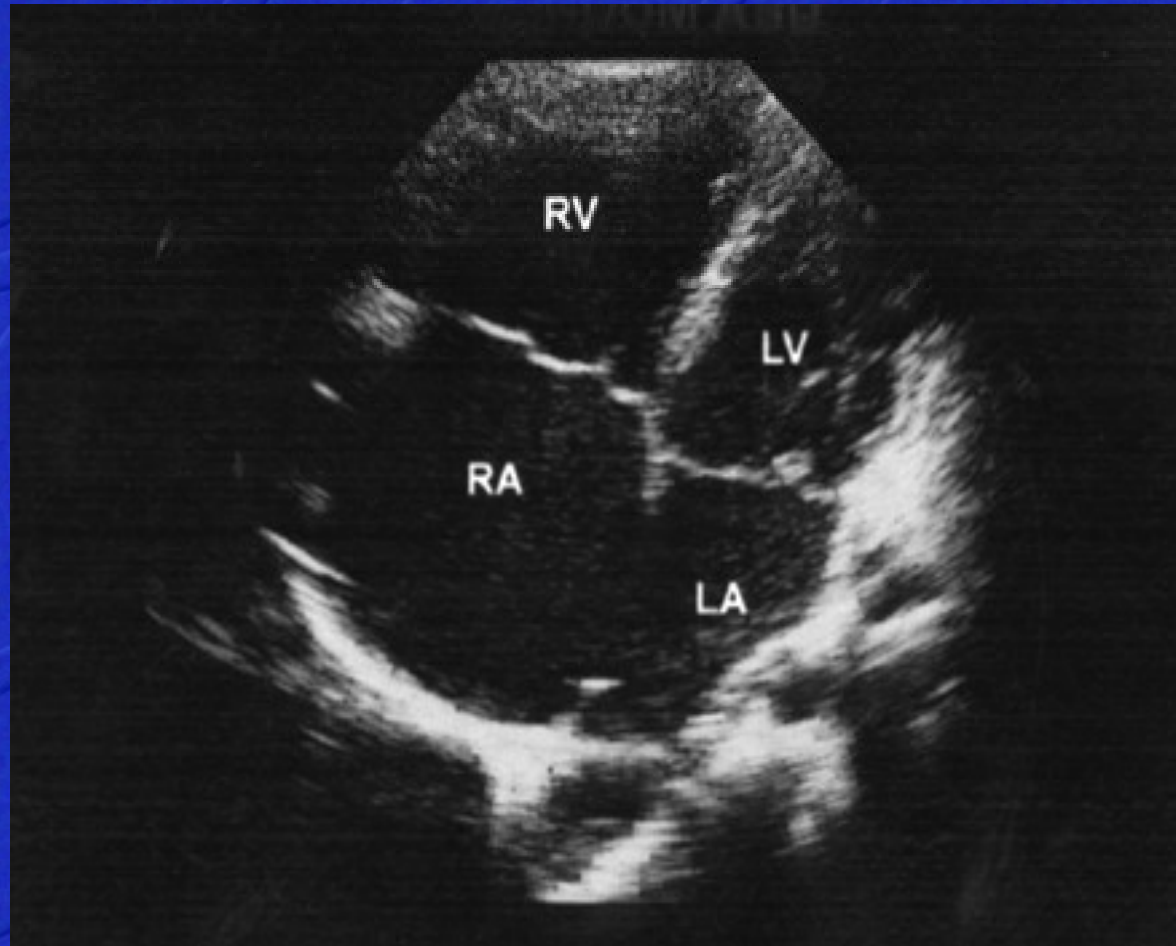


Patras University Hospital



Atrial Septal Defect

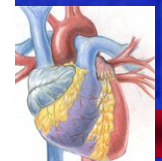
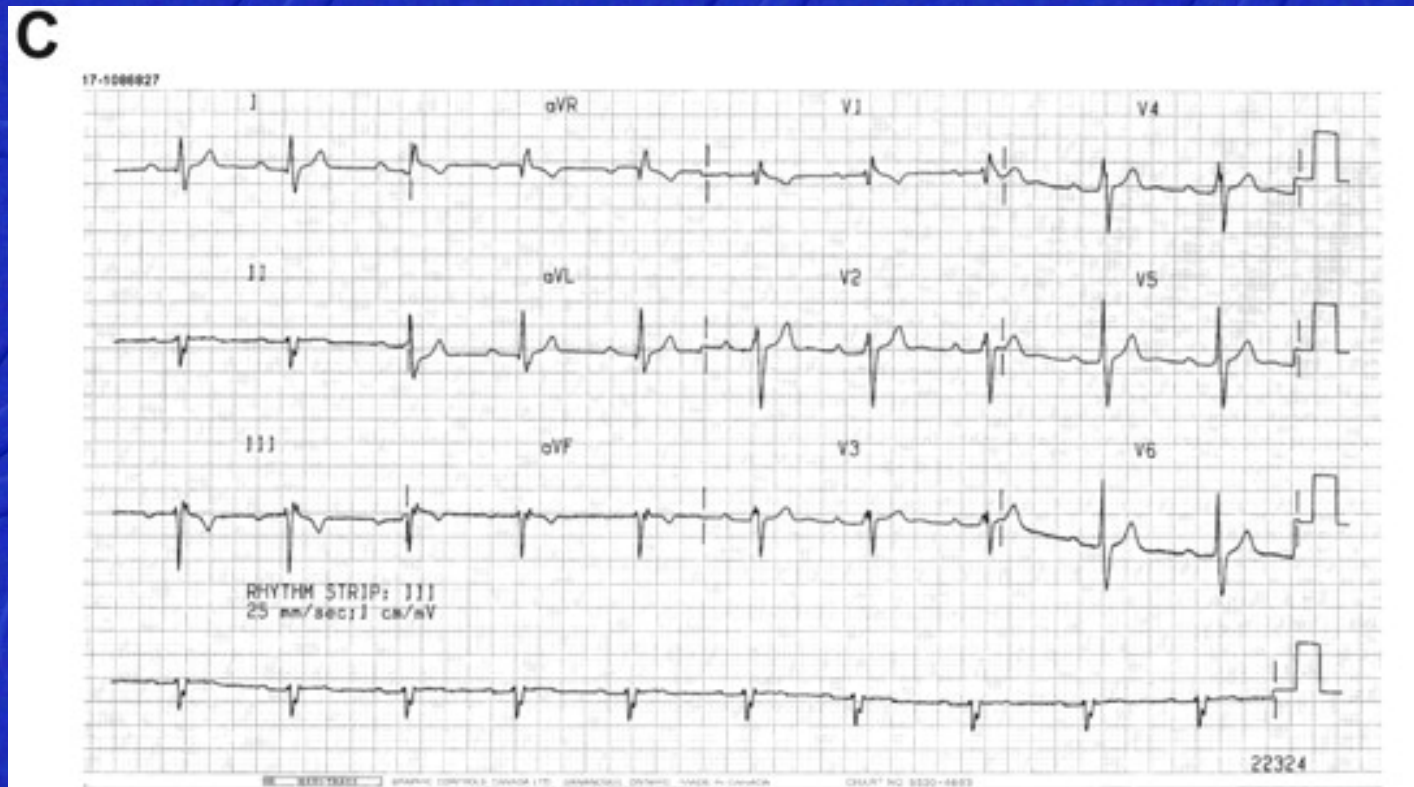
- 1/1500 live births
- Secundum
 - ▶ most common ACHD (6-10%)
 - ▶ ECG: RAD



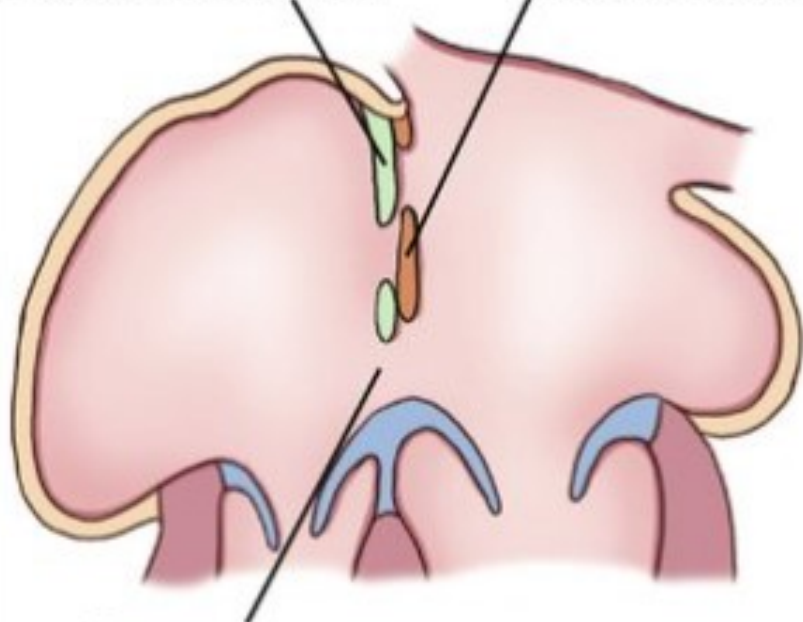
27 yrs old girl...

First-degree AV block, left-axis deviation, voltage evidence of right ventricular hypertrophy

- Generally asymptomatic
- RV heave
- 2/6 systolic murmur 2nd LICS and cardiac apex...
- ECG:



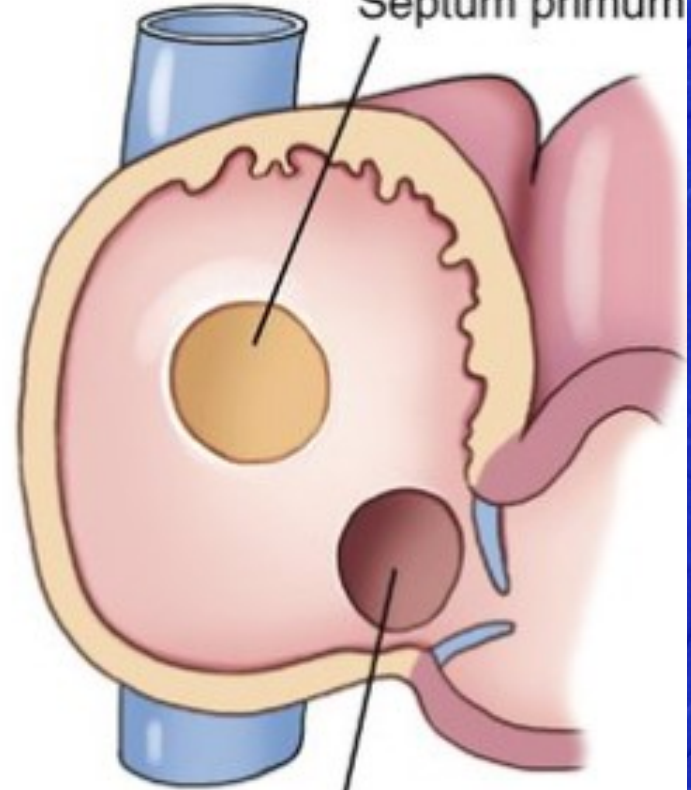
Septum secundum Septum primum



Patent ostium primum

A

Septum primum

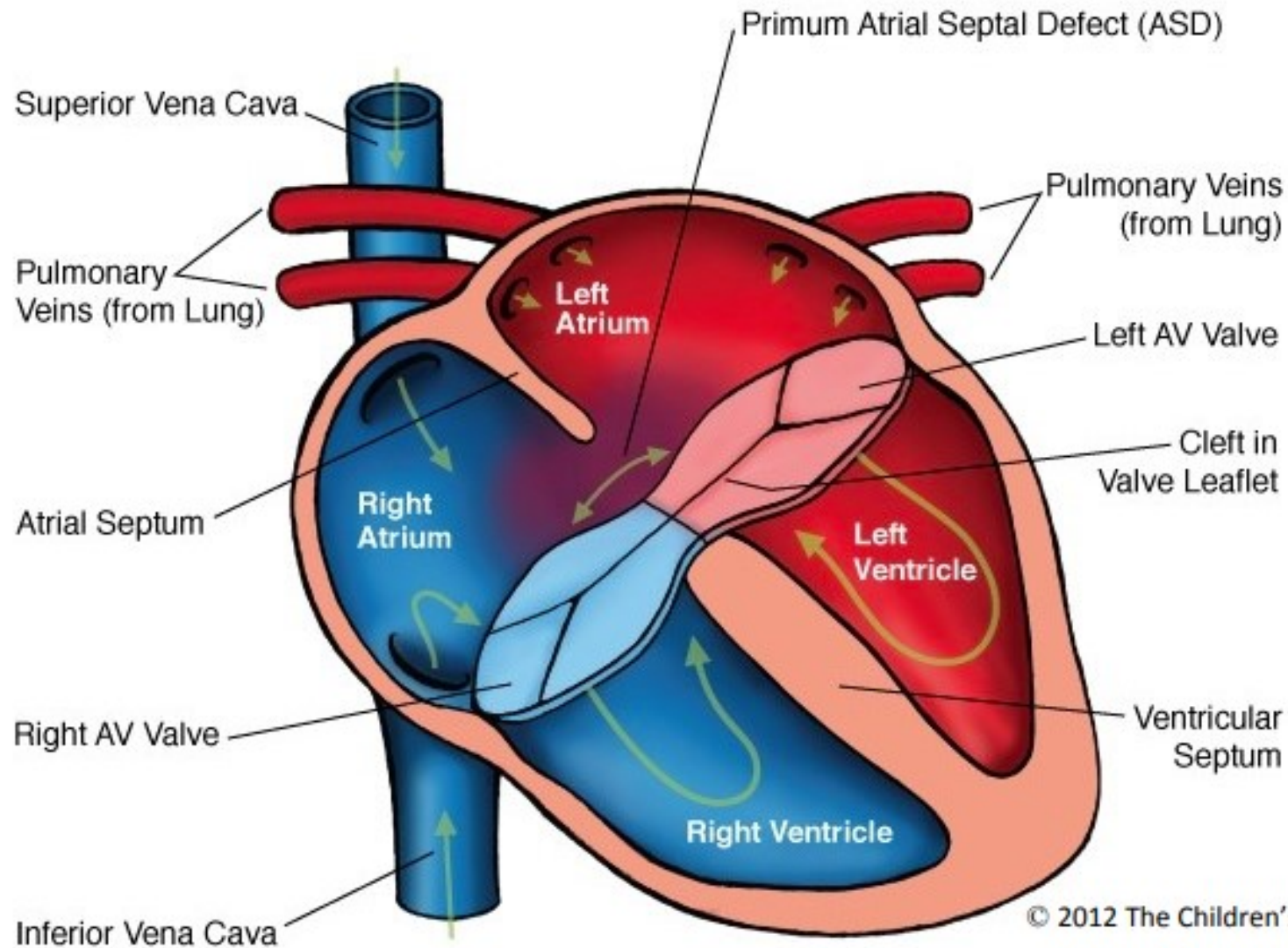


B

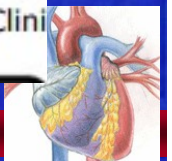
Ostium primum



Atrioventricular Septal Defect (AVSD) Partial



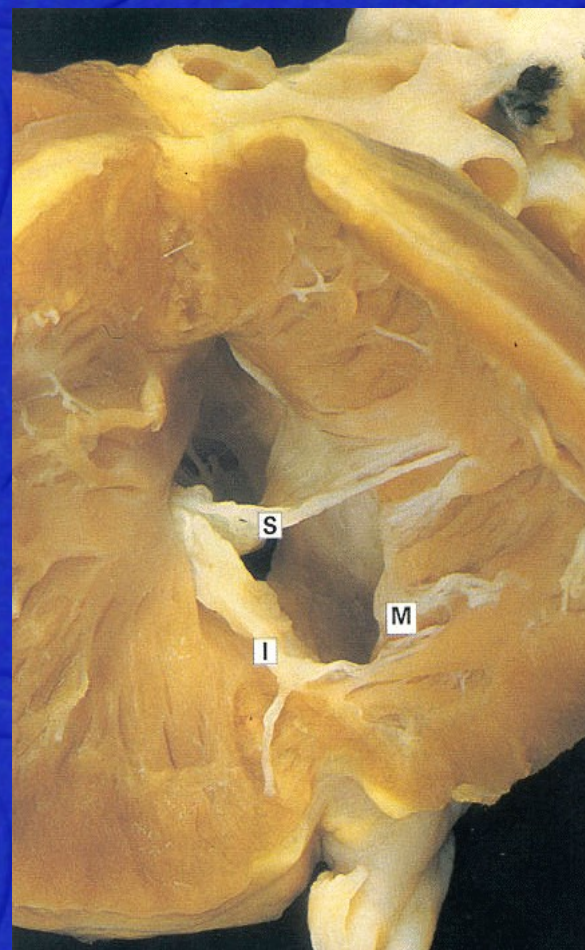
© 2012 The Children's Heart Clinic



Ostium Primum...

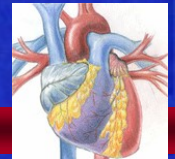


Cleft MV (partial AVSD)...

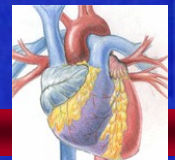
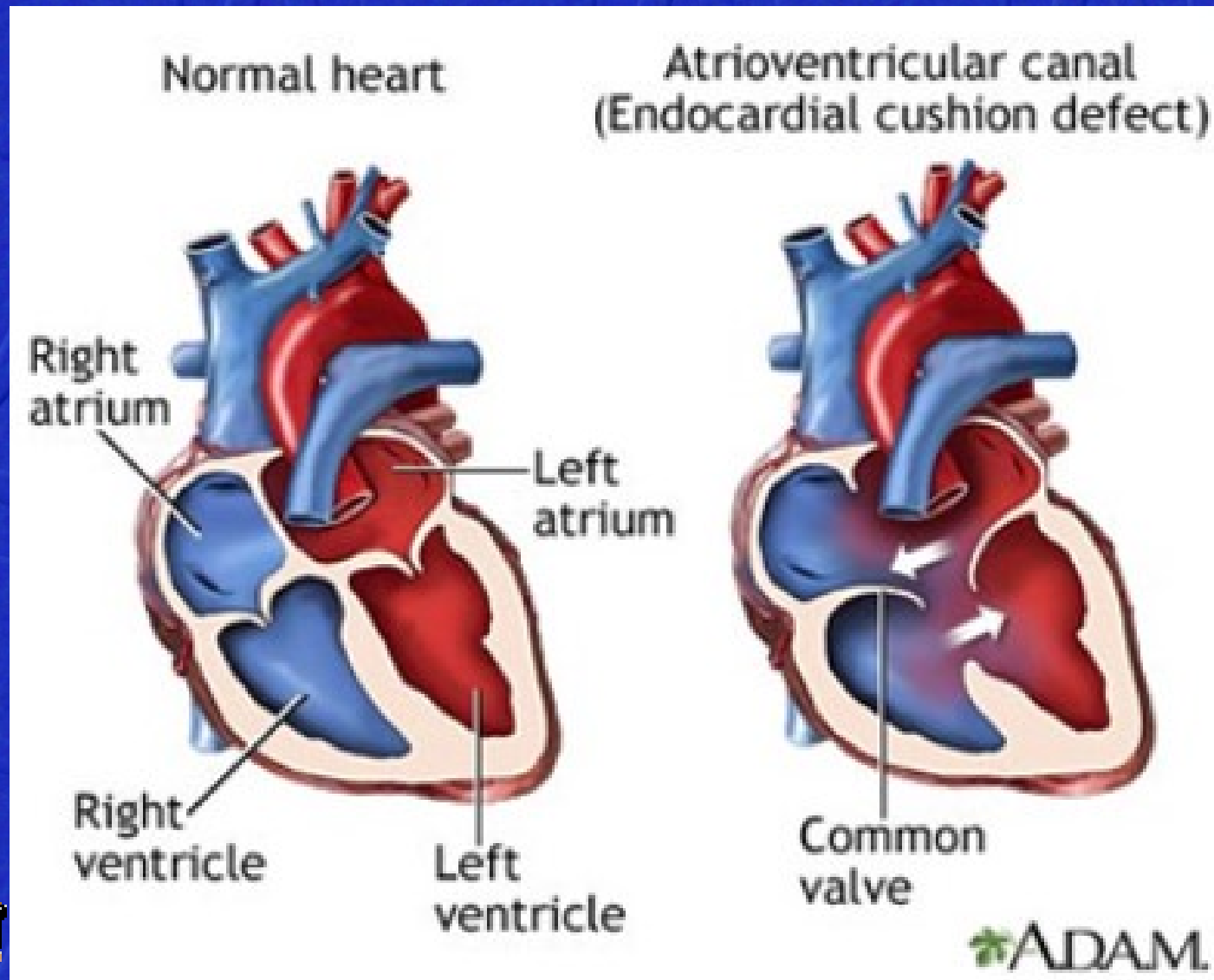


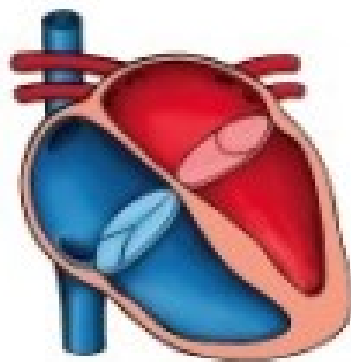
Atrial Septal Defect

- 1/1500 live births
- Secundum
 - ▶ most common ACHD (6-10%)
 - ▶ RAD
- Primum
 - ▶ associated with other endocardial cushion defects (cleft AV valves, inlet type VSD)
 - ▶ LAD

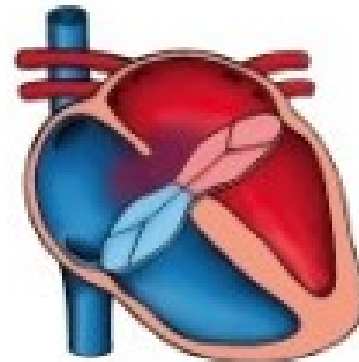


Atrio Ventricular Septal Defect (AVSD)...

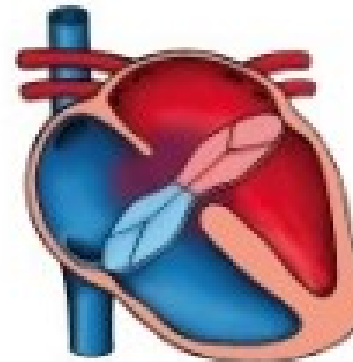




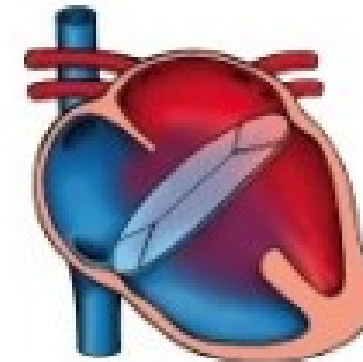
**Normal
heart**



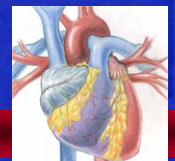
**Partial
AVSD**



**Transitional
AVSD**

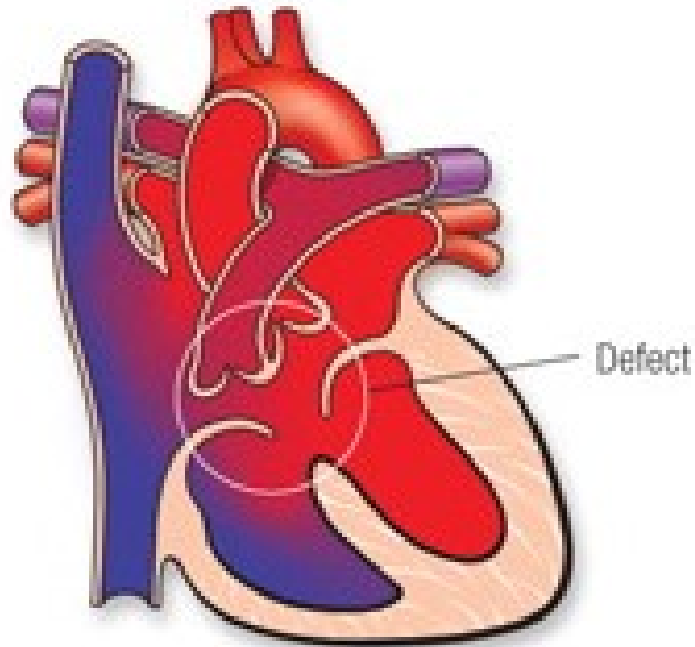


**Complete
AVSD**



Partial vs. Complete AV cannal...

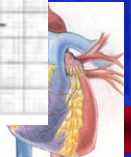
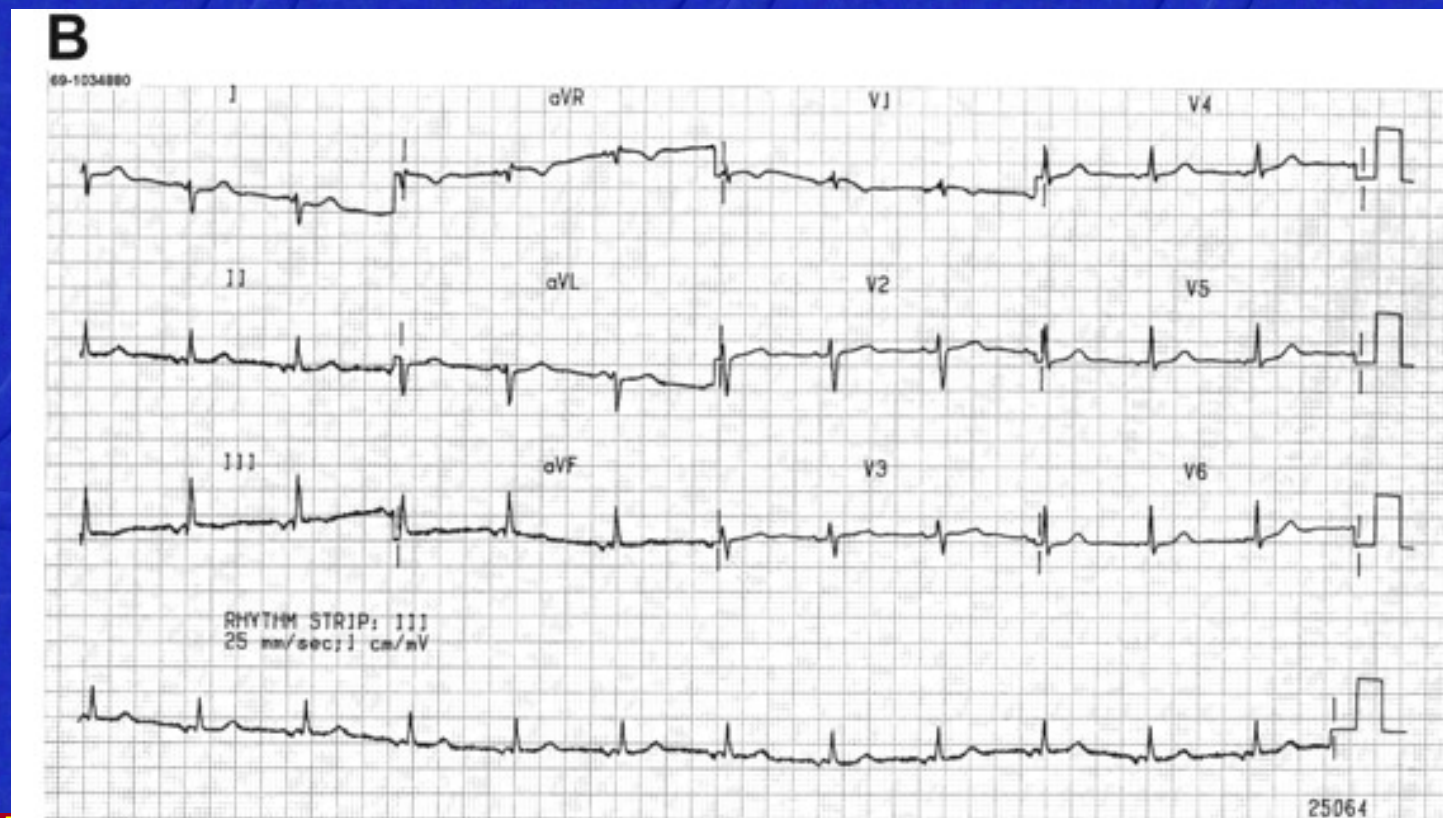
Atrioventricular Canal Defect

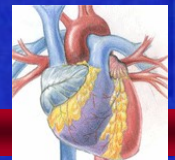
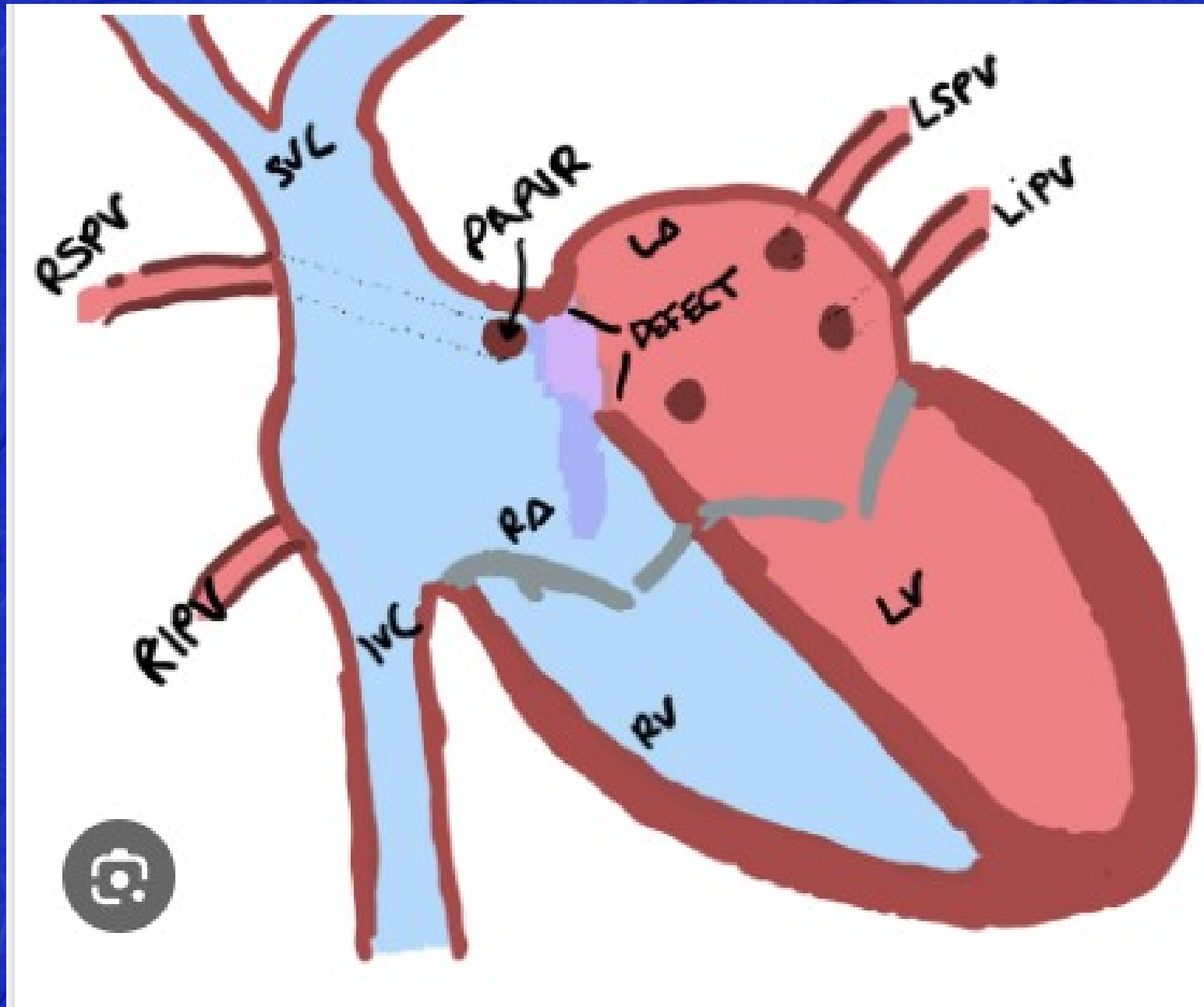


27 yrs old girl...

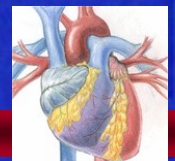
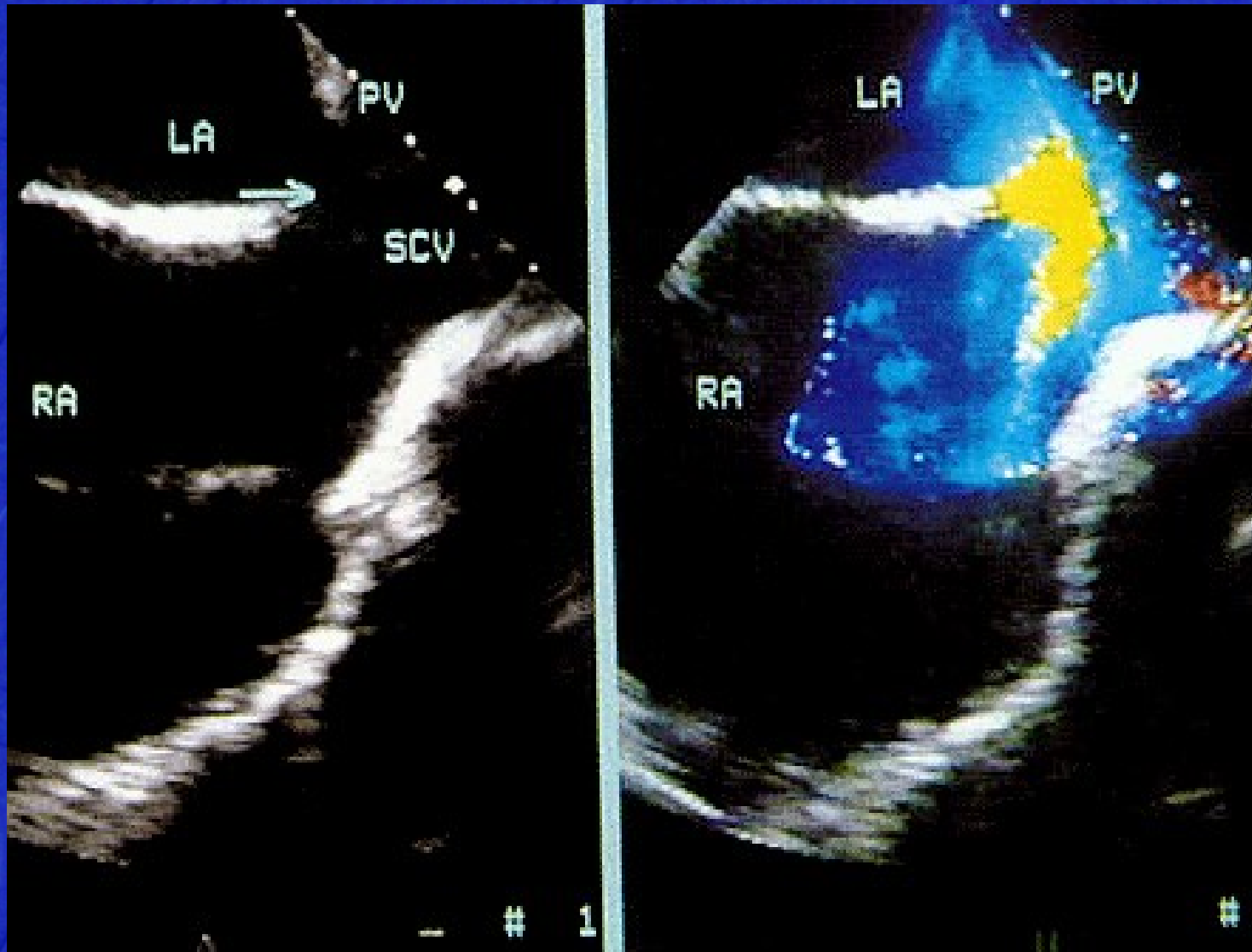
Inverted inferior P waves, right-axis deviation

- Generally asymptomatic
- RV heave
- 2/6 systolic murmur 2nd LICS
- ECG:



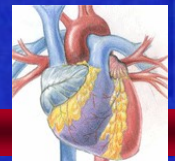


ASD: Sinus Venosus...



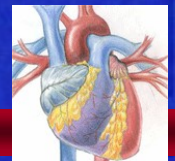
Atrial Septal Defect

- 1/1500 live births
- Secundum
 - ▶ most common ACHD (6-10%)
 - ▶ RAD
- Primum
 - ▶ associated with other endocardial cushion defects (cleft AV valves, inlet type VSD)
 - ▶ LAD
- Sinus Venosus
 - ▶ large, associated with anomalous pulmonary venous drainage (usually R superior PV)



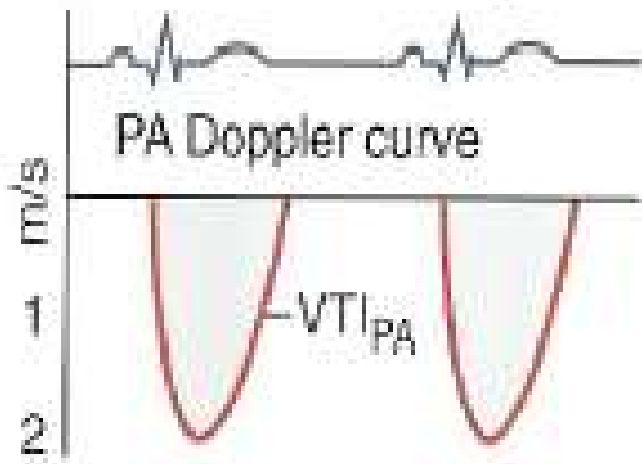
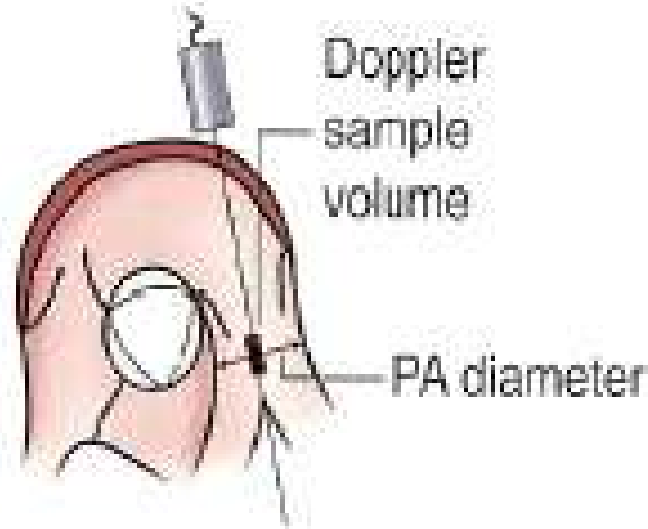
ASD - Clinical

- Majority repaired in childhood, but may present in adolescence/adulthood
- Asymptomatic
 - ▶ murmur, abnl ECG/CXR
- Symptomatic
 - ▶ dyspnea/CHF
 - ▶ CVA/emboli
 - ▶ Atrial Fibrillation
 - ▶ Respiratory Infections



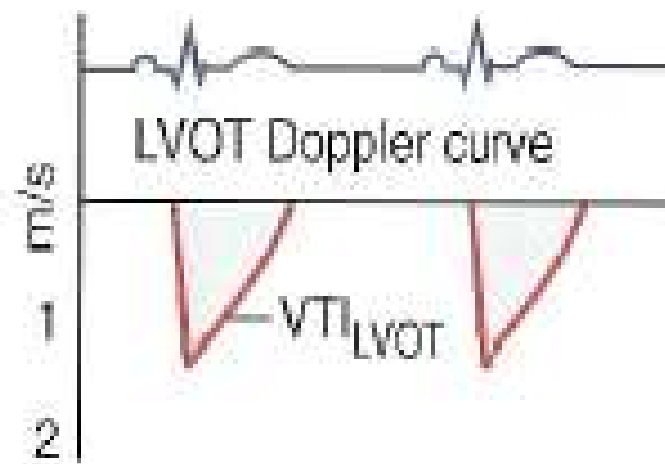
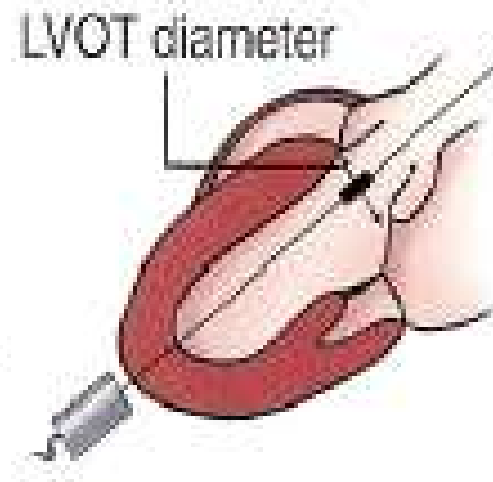
Shunt calculation (Q_p/Q_s)...

Pulmonary Flow (Q_p)



$$Q_p = CSA_{PA} \times VTI_{PA}$$

Systemic Flow (Q_s)



$$Q_s = CSA_{LVOT} \times VTI_{LVOT}$$



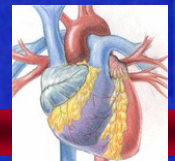
ASD: Therapy

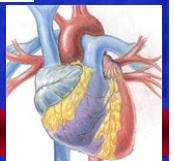
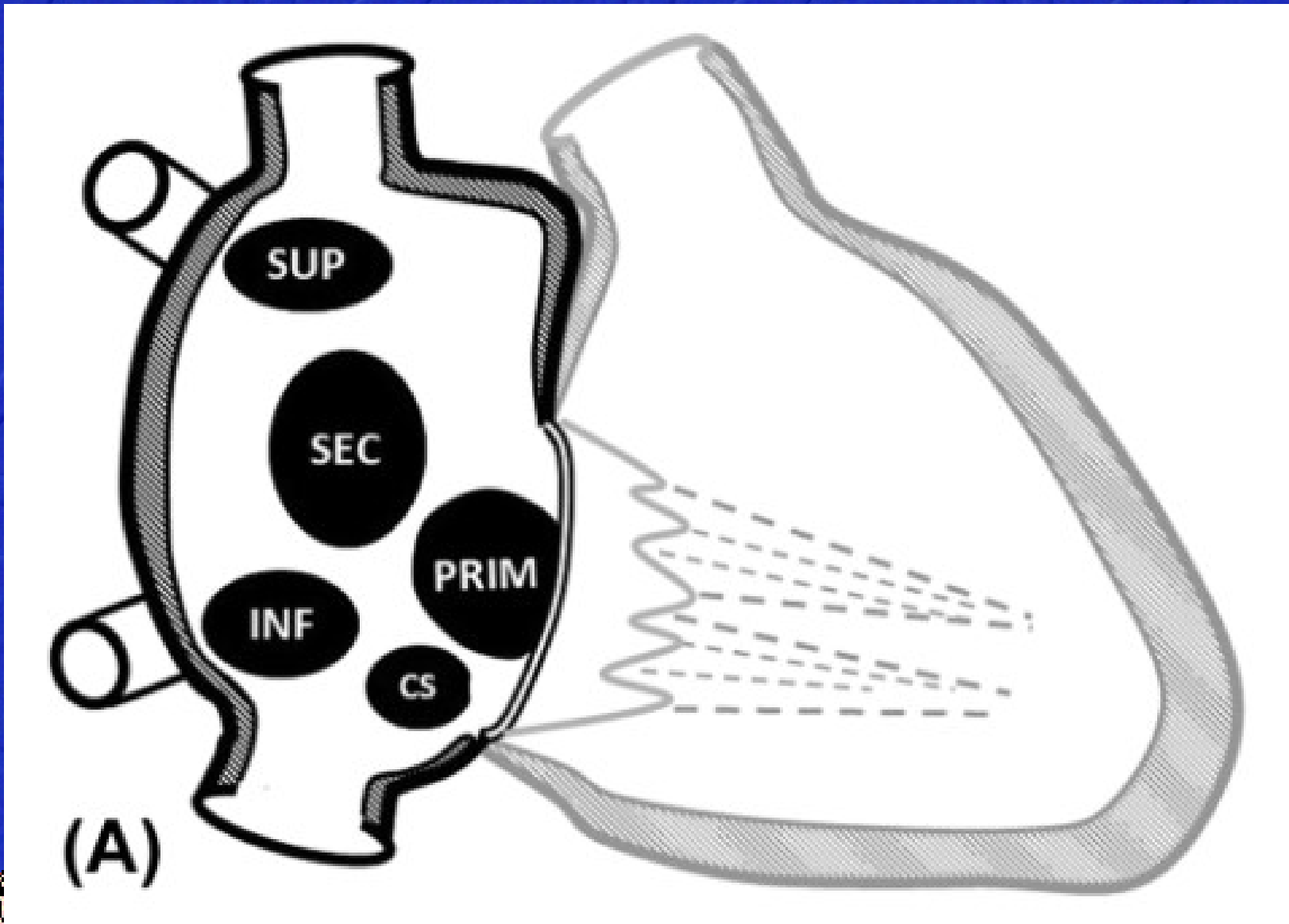
- **Percutaneous Closure**

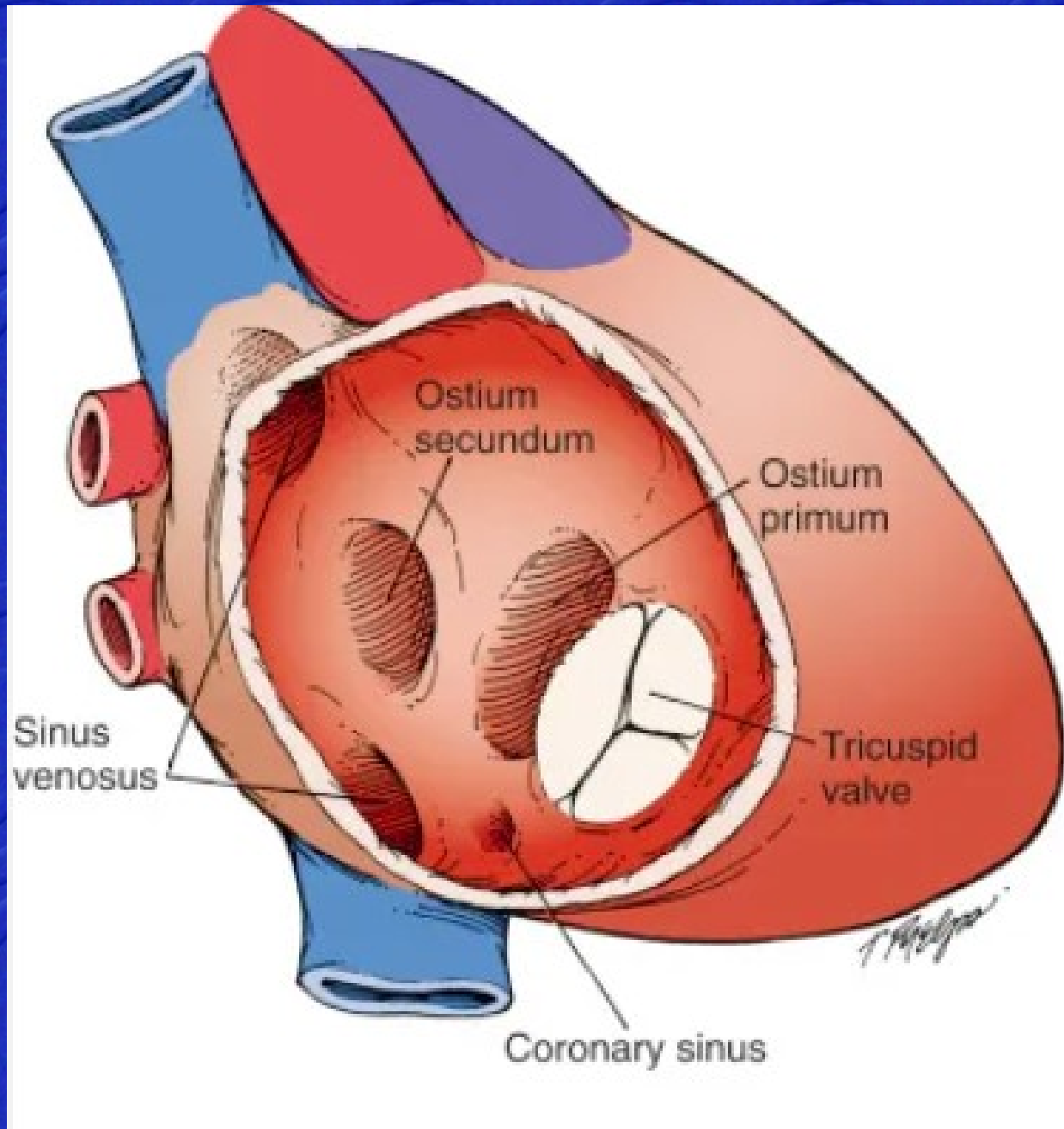
- ▶ only for secundum (contra in others)
- ▶ adequate superior/inferior rim around ASD
- ▶ no R-L shunting

- **Surgical Closure**

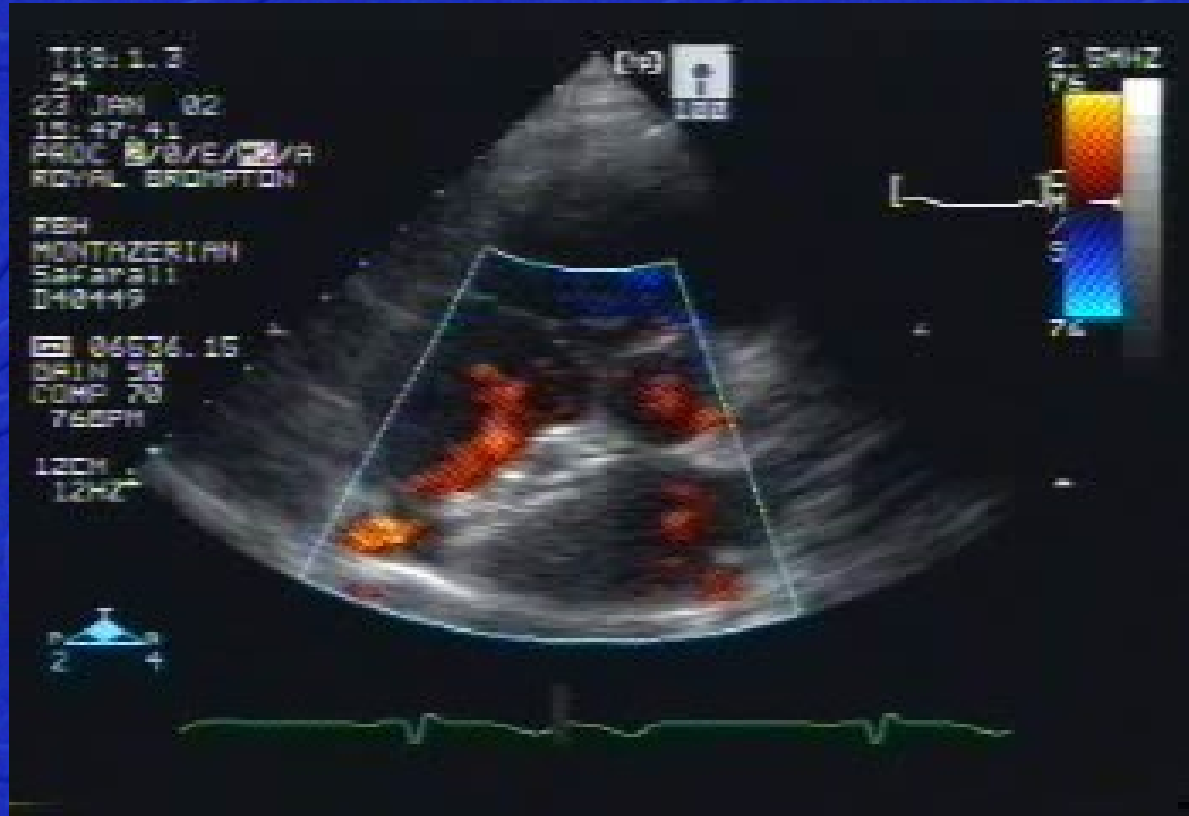
- ▶ Good prognosis:
 - closure age < 25, PA pressure <40
 - If >25 or PA>40, decreased survival due to CHF, stroke, and afib

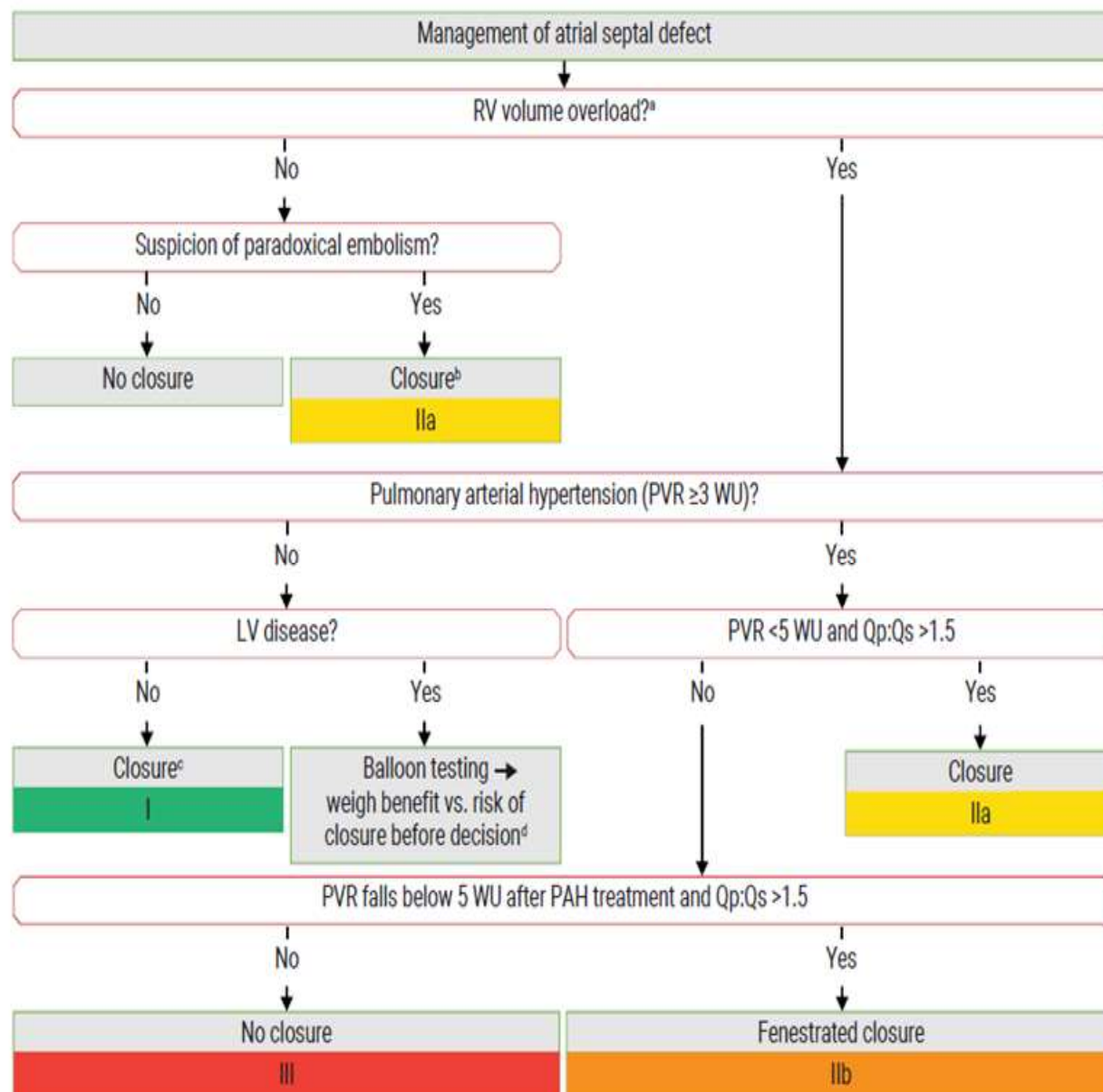






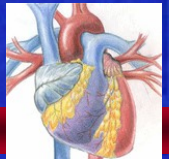
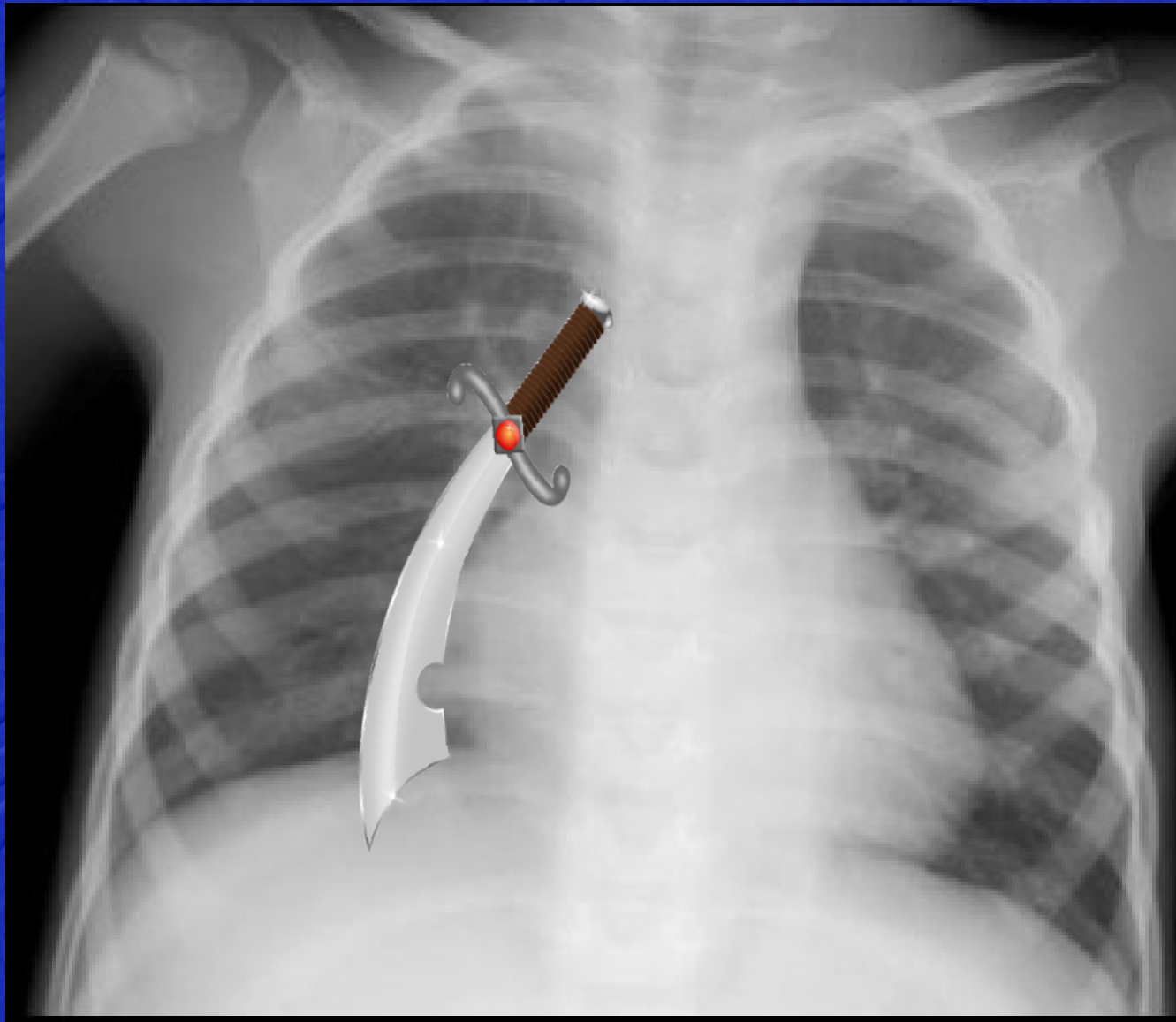
ASD: Transcatheter closure...



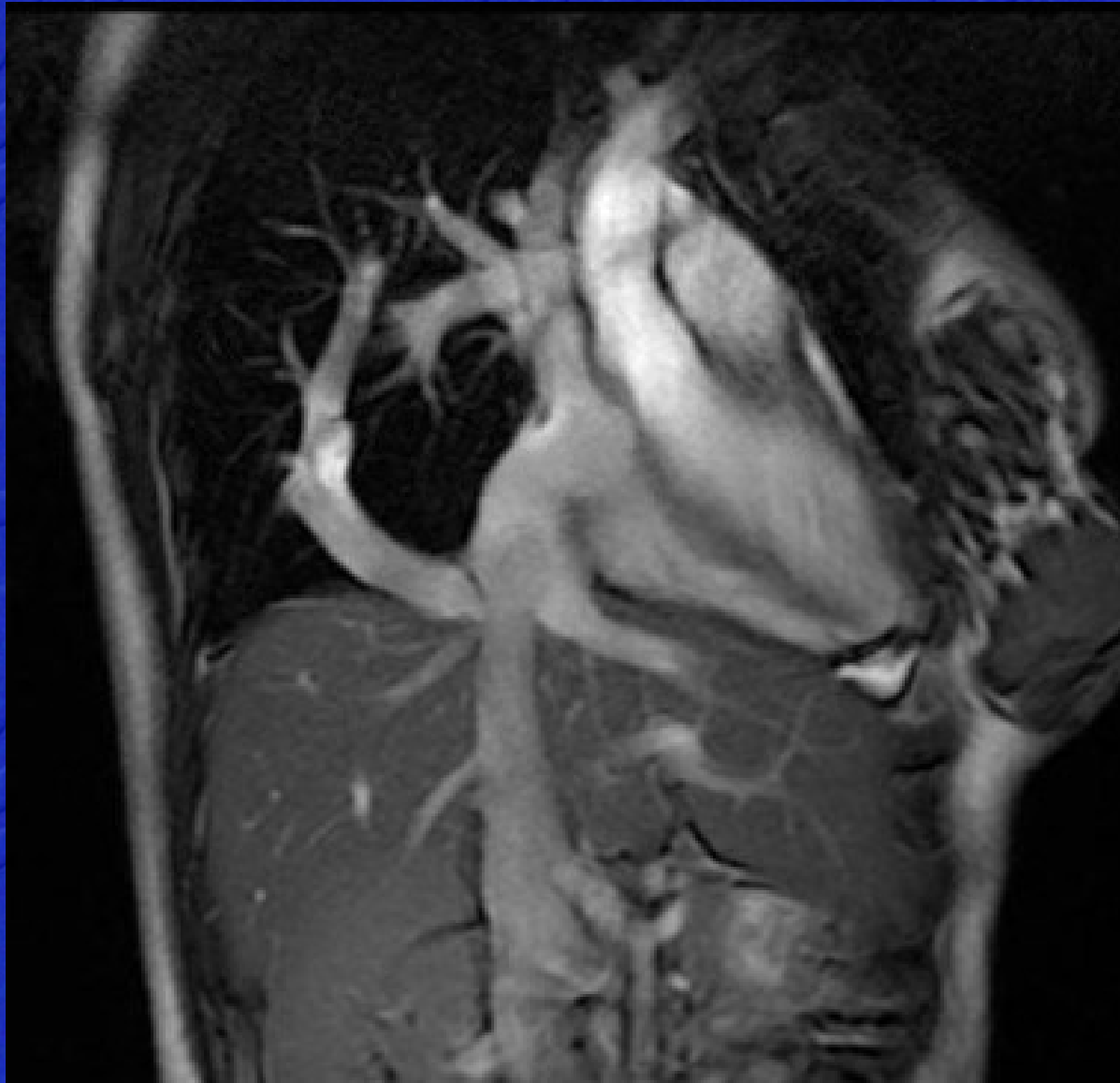


^aRV enlargement with increased stroke volume.
^bProviding there is no PAH or LV disease.
^cIn elderly patients not suitable for device closure, carefully weigh surgical risk vs. potential benefit of ASD closure.
^dCarefully weigh the benefit of eliminating L-R shunt against the potential negative impact of ASD closure on outcome due to an increase in filling pressure (taking closure, fenestrated closure, and no closure into consideration).

What is this???



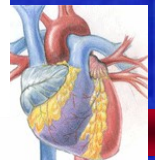
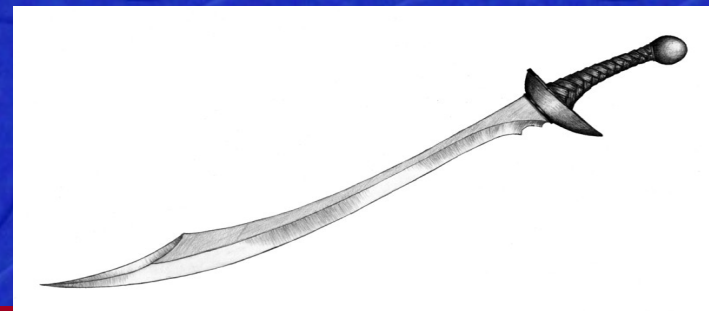
What is this???



What is this???

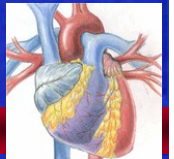
- An anomalous vein connects between the **pulmonary venous circulation** and **systemic venous circulation**.

This anomalous pulmonary venous return can be either partial (PAPVR) or total (TAPVR). The syndrome associated with **PAPVR** is more commonly known as "**Scimitar syndrome**"



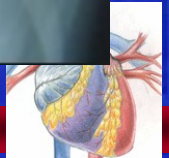
Scimitar Syndrome

- 1-3 per 1,00,000 births
- Hypogenetic lung syndrome /
Congenital pulmonary venolobar syndrome
- FELSON - coined the term CPVS
- WOODRING et al - extended the term CPVS to
cover a range of anomalies including pulmonary
hypoplasia & pulmonary sequestration

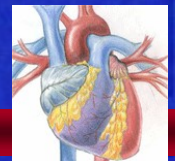
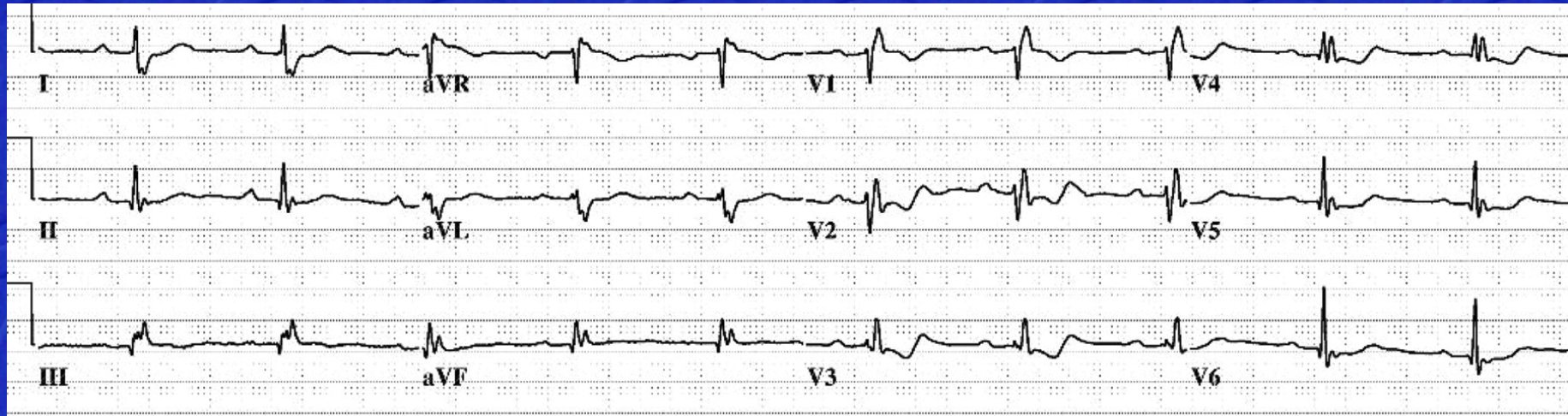


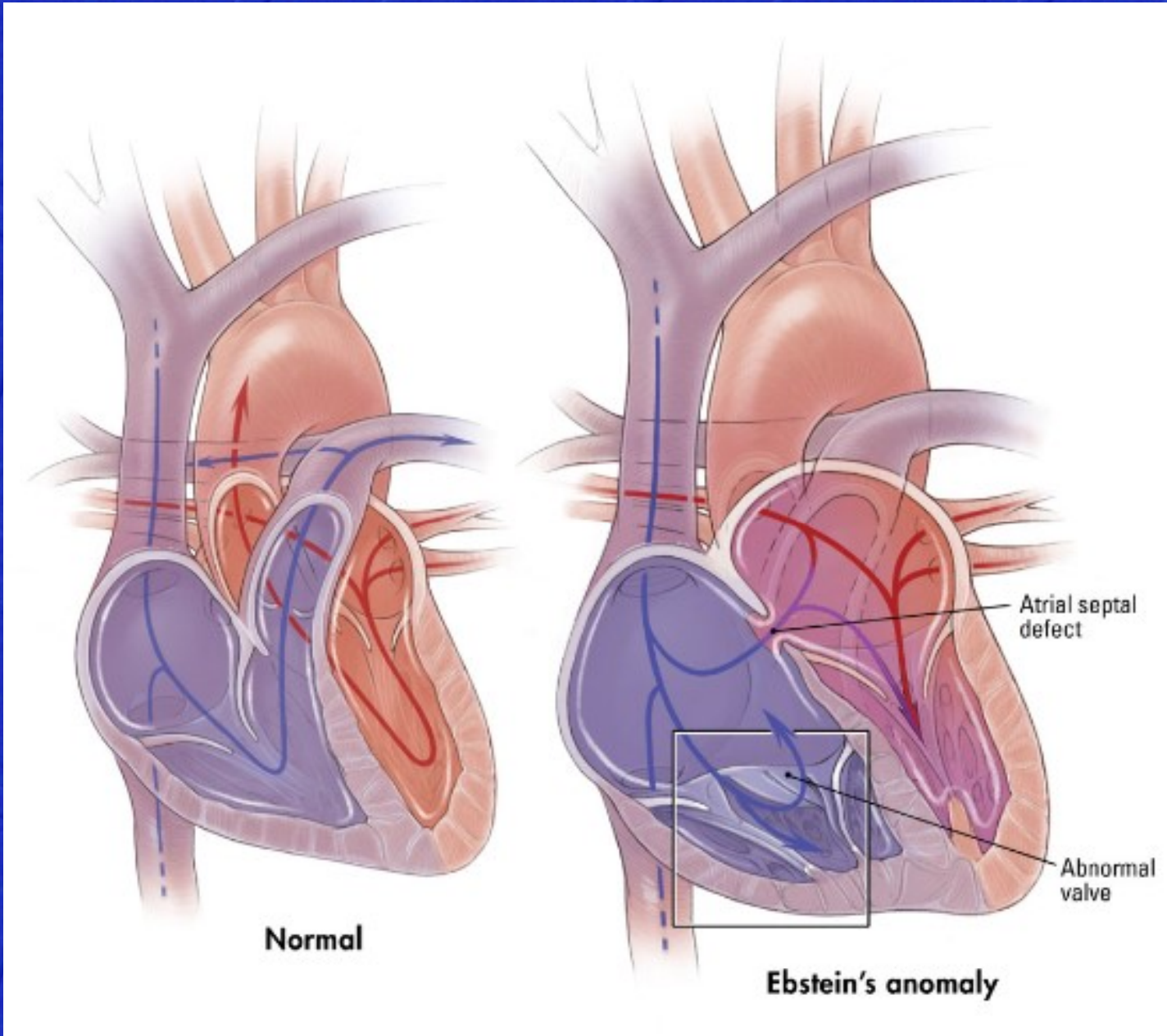
GUCH: Clinical oriented approach cont...

- 34 yrs old @ SOB...
- 3/6 systolic murmur
2nd LICS and 4th
LICS parasternal
- Ht 55 %, SAT 95%



Prolongation of the PR interval (226 ms), right bundle-branch block, and somewhat bizarre configuration of the QRS complex.





Normal

Ebstein's anomaly

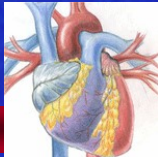


Figure 4. Severe Ebstein's malformation of tricuspid valve (4-chamber view) showing marked downward displacement of shelf-like posterior leaflet with attachment to underlying free wall by numerous muscular stumps (arrows), markedly dilated atrialized portion of right ventricle (ARV), small functional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

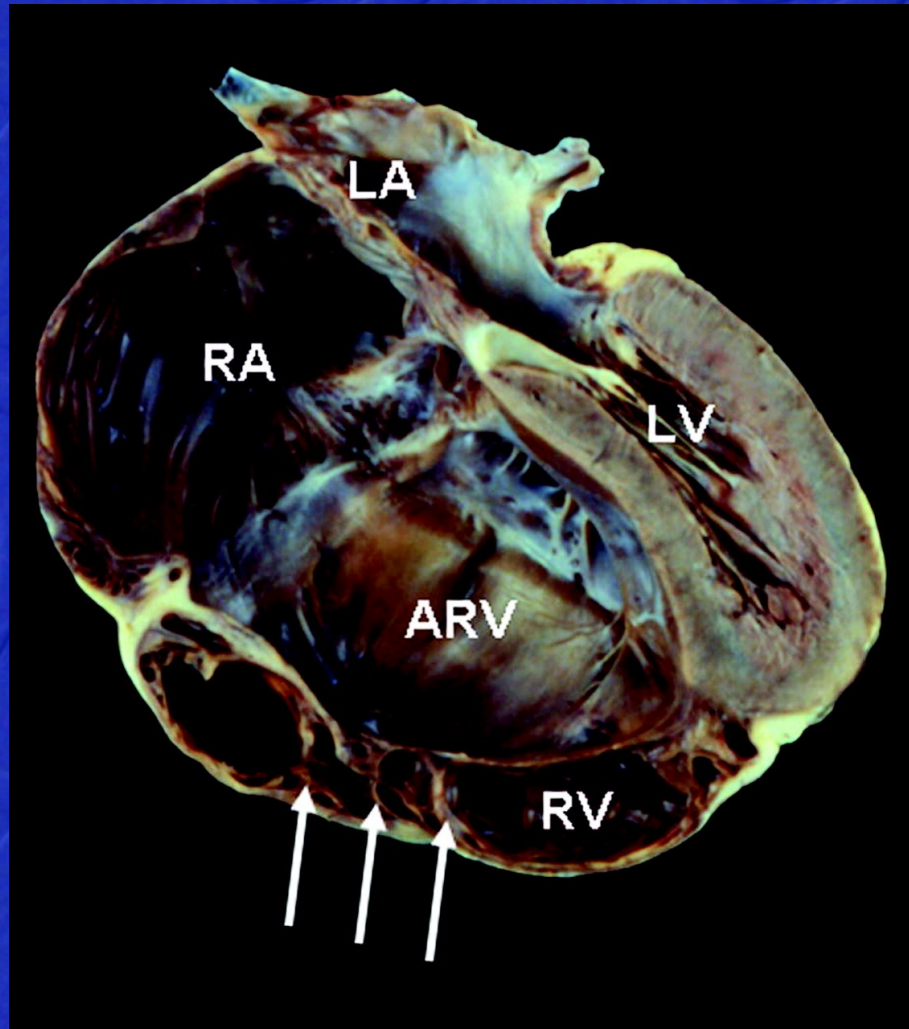
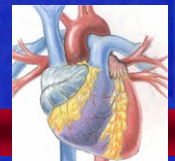
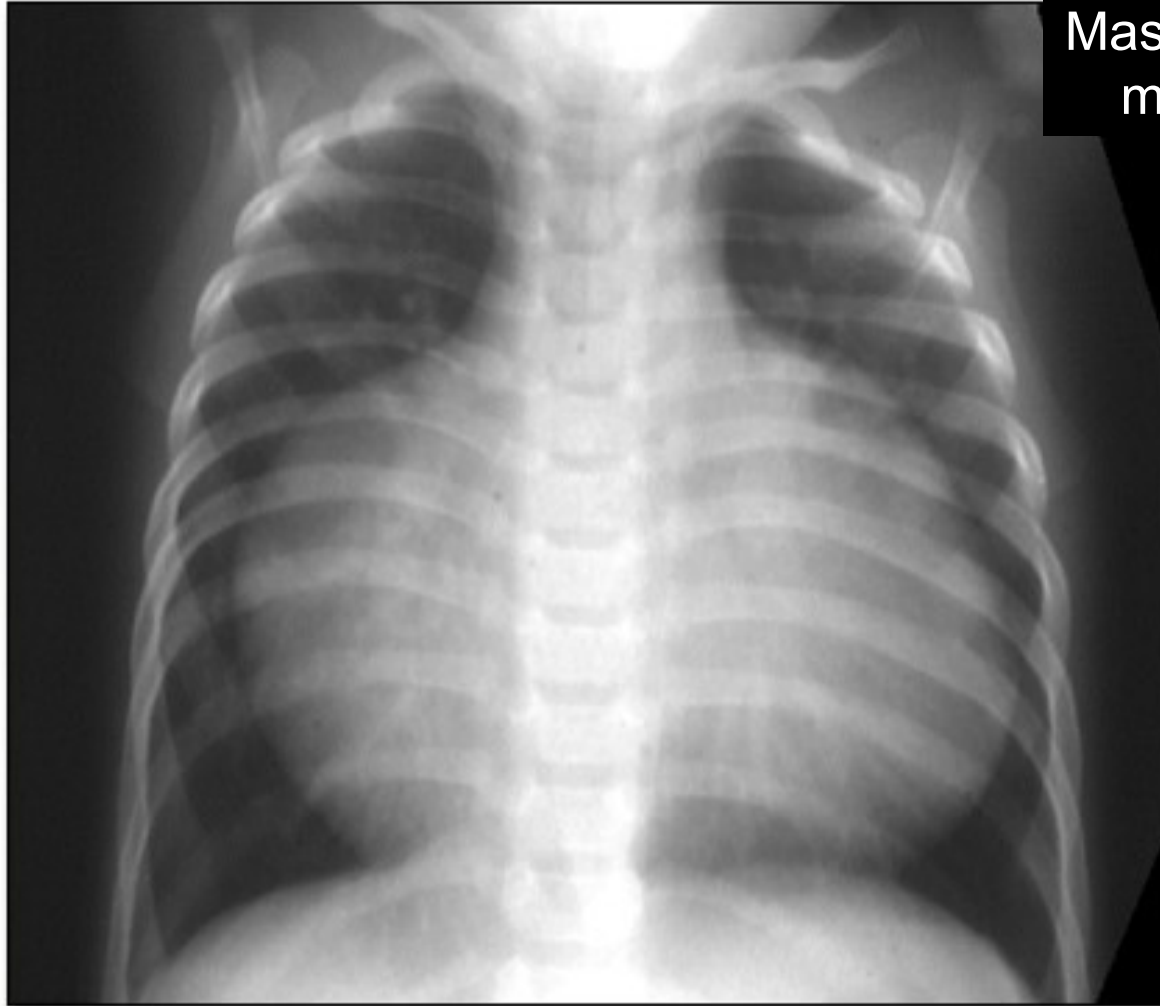


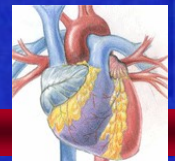
Figure 3. Marked cardiomegaly caused by right-sided chamber dilatation in a 67-year-old man with severe Ebstein's anomaly, with normal heart at right for comparison (anterior view).



CXR of a Patient With Ebstein Anomaly

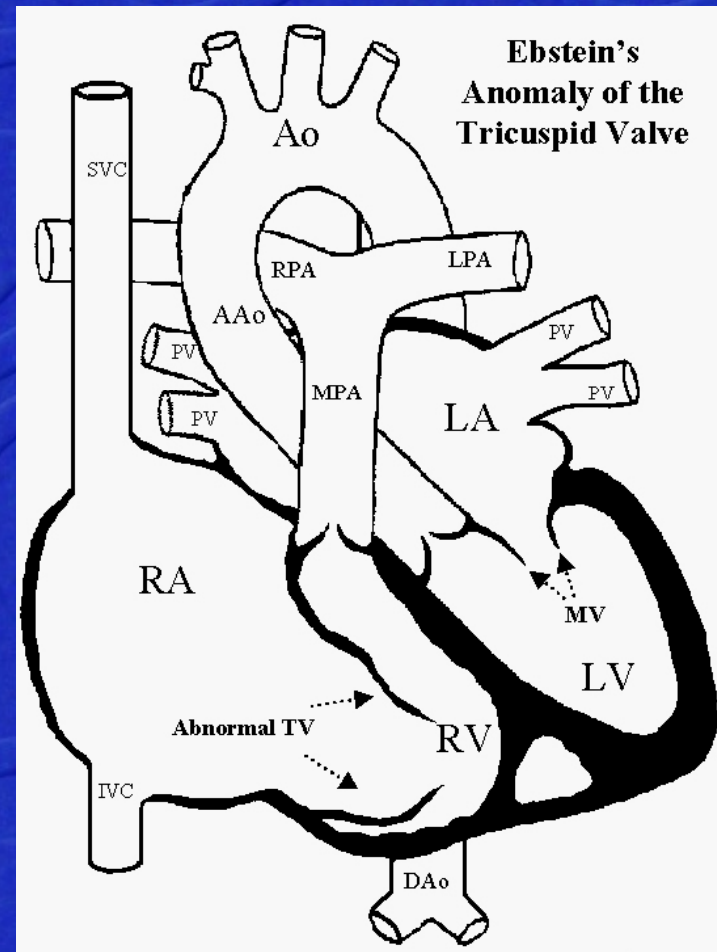


Massive cardiomegaly,
mainly due to RAE



Ebsteins Anomaly

- Atrialization of RV, sail-like TV, TR
- 50% ASD/PFO
- 50% ECG evidence of WPW
- Age at presentation varies from childhood→adulthood and depends on factors such as severity of TR, Pulm Vascular resistance in newborn, and associated abnormalities such as ASD



Ebsteins: Clinical Presentation

- Pediatric

- ▶ murmur

- Adult (unrepaired with ASD)

- ▶ atrial arrhythmias

- ▶ murmur

- ▶ cyanosis

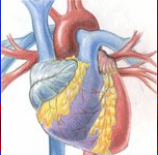
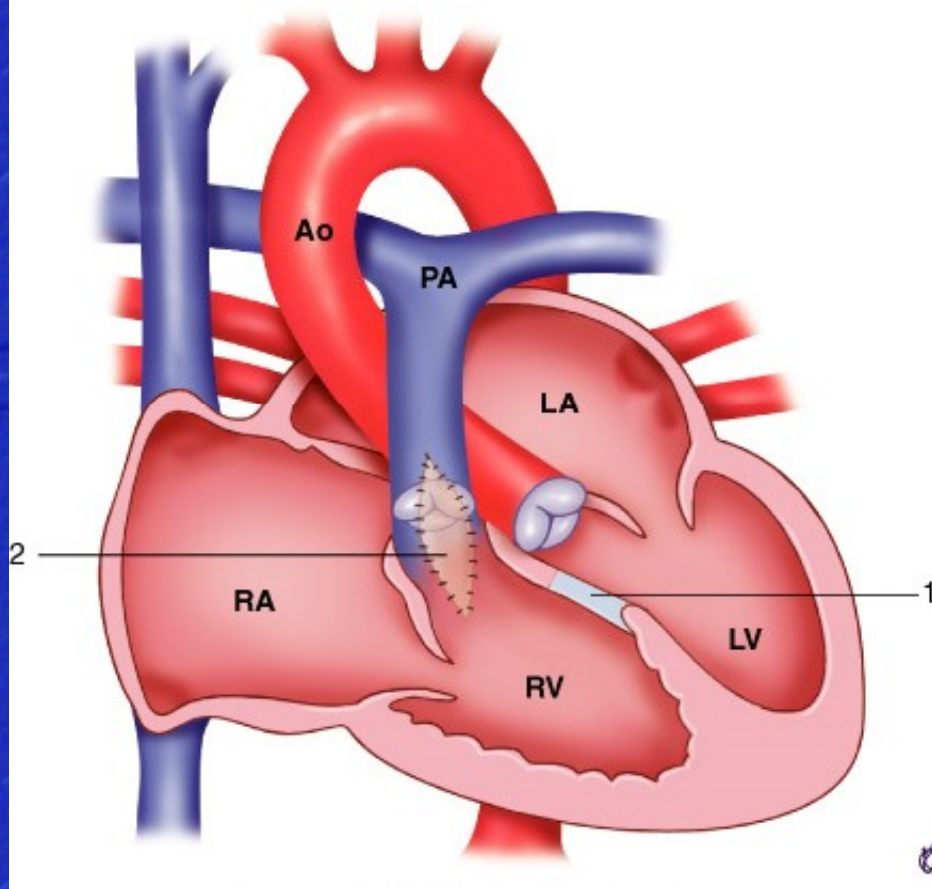
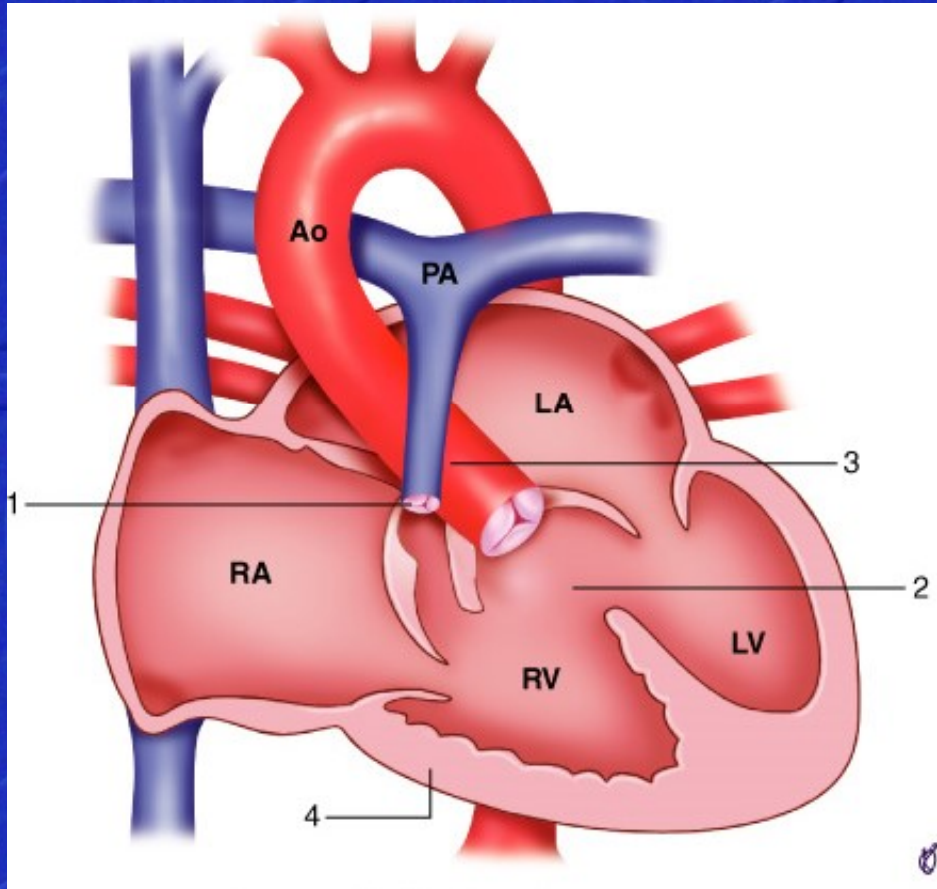
- R→L shunt NOT due to PulmHTN but TR jet directed across ASD

- ▶ exercise intolerance

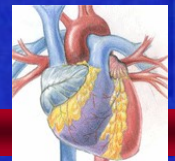
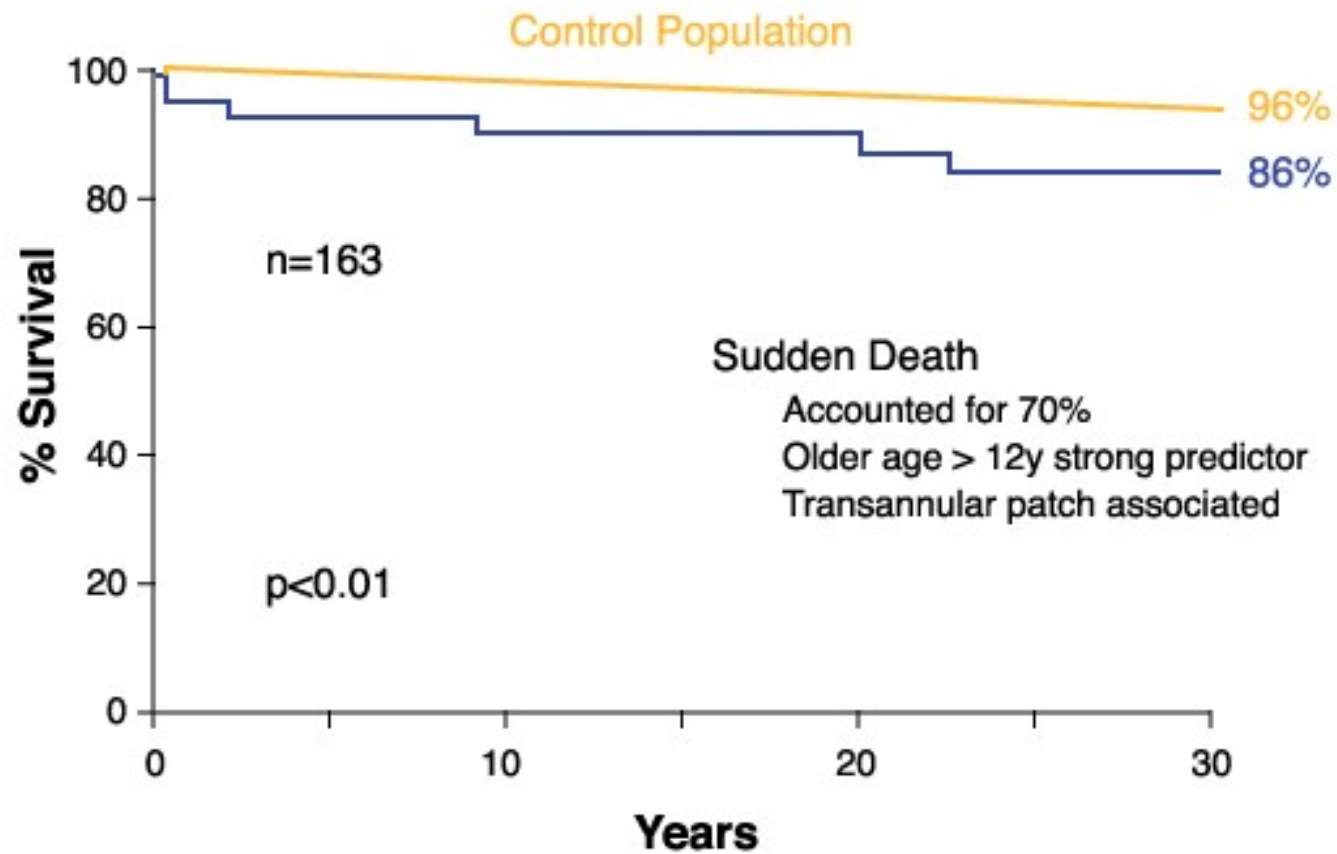
- Surgery in pts with significant TR/sxs



Repaired ToF (rToF)...

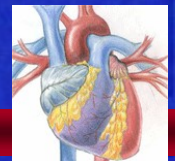
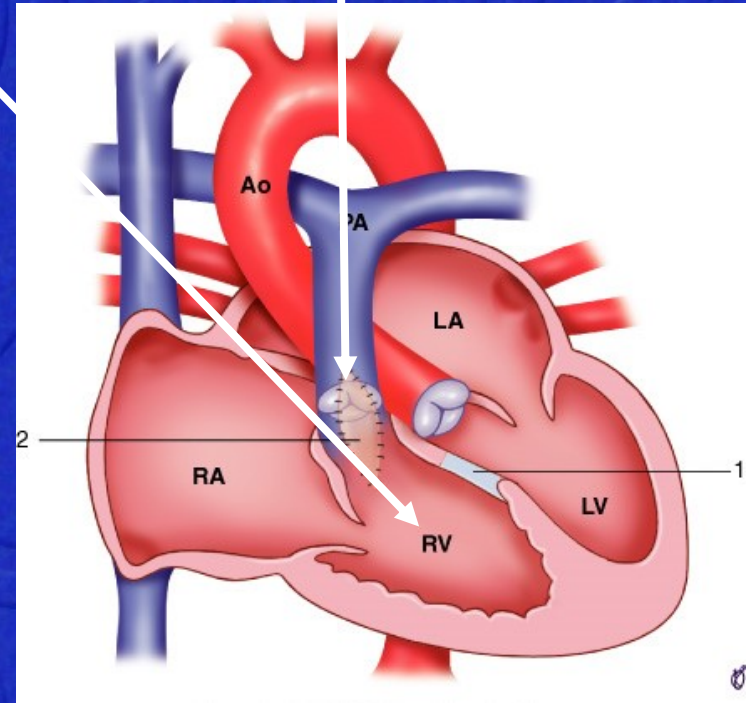


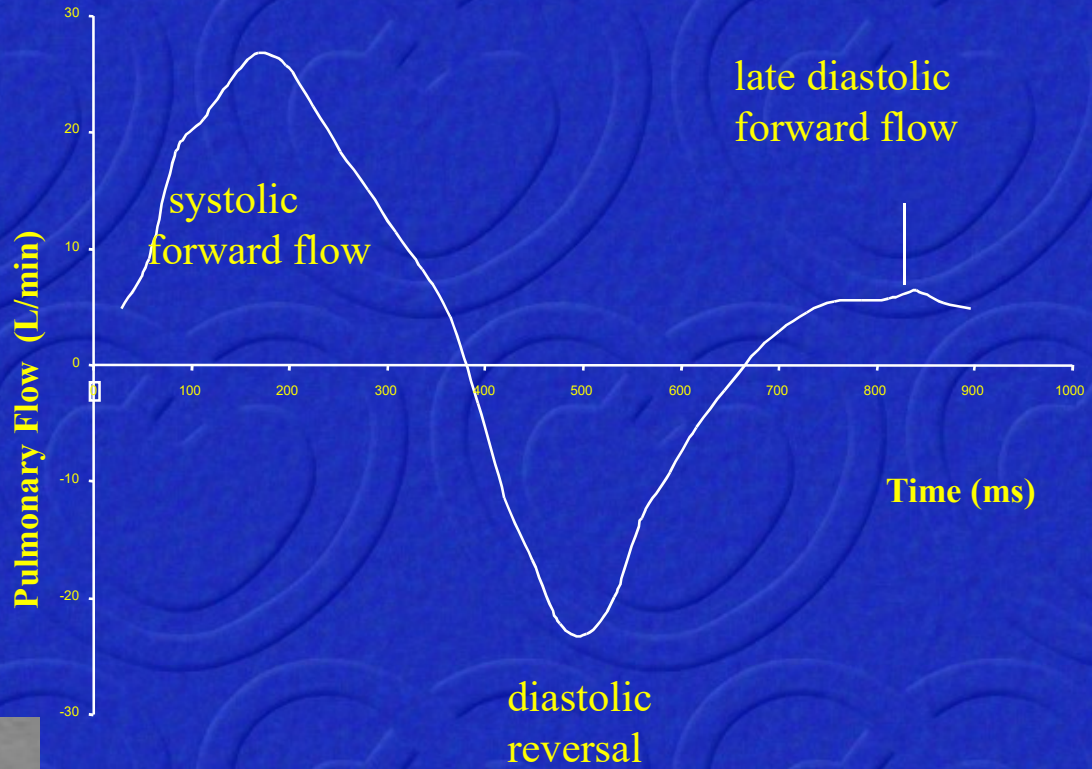
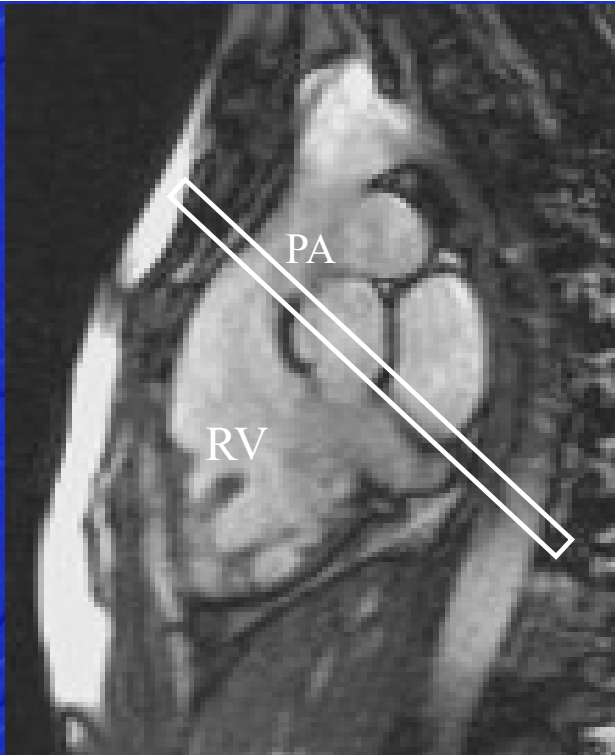
Survival Following Complete Repair of Tetralogy of Fallot



ToF: Correction or Repair;

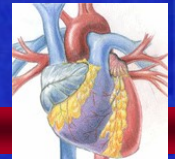
- Pulmonary Regurgitation
- RV dilatation/dysfunction
- RVOT, PV, PA stenosis
- VSD/MAPCAs
- TR & RA
- RVSP
- Aortic dilatation, AR ;



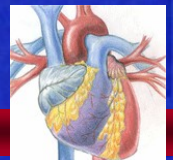
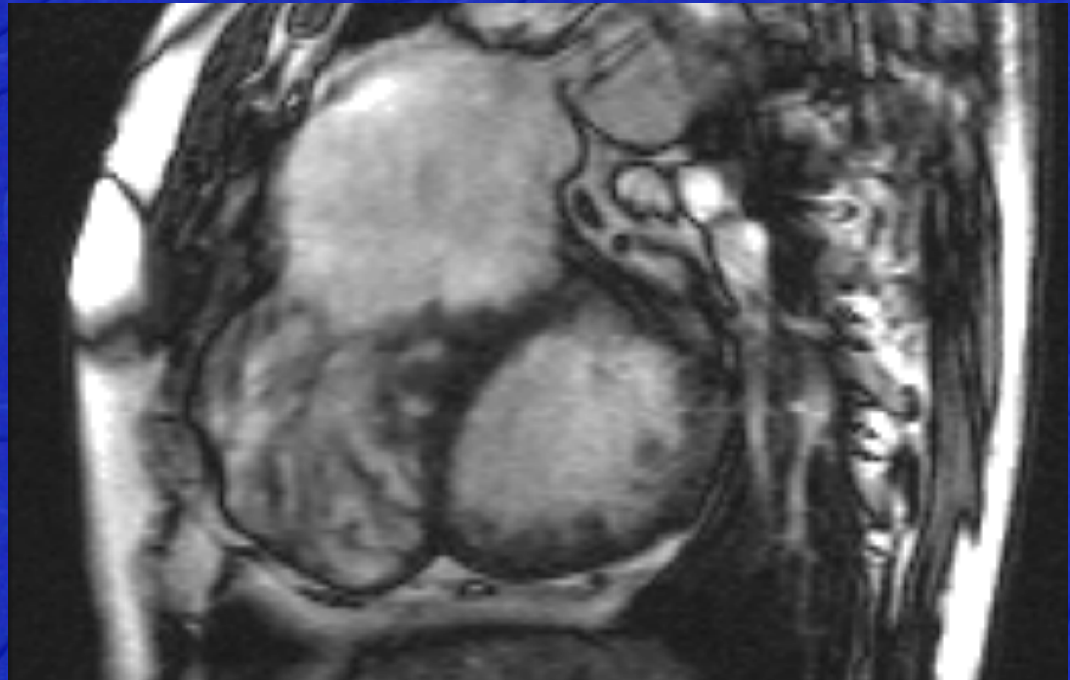


PRF range 0-55%

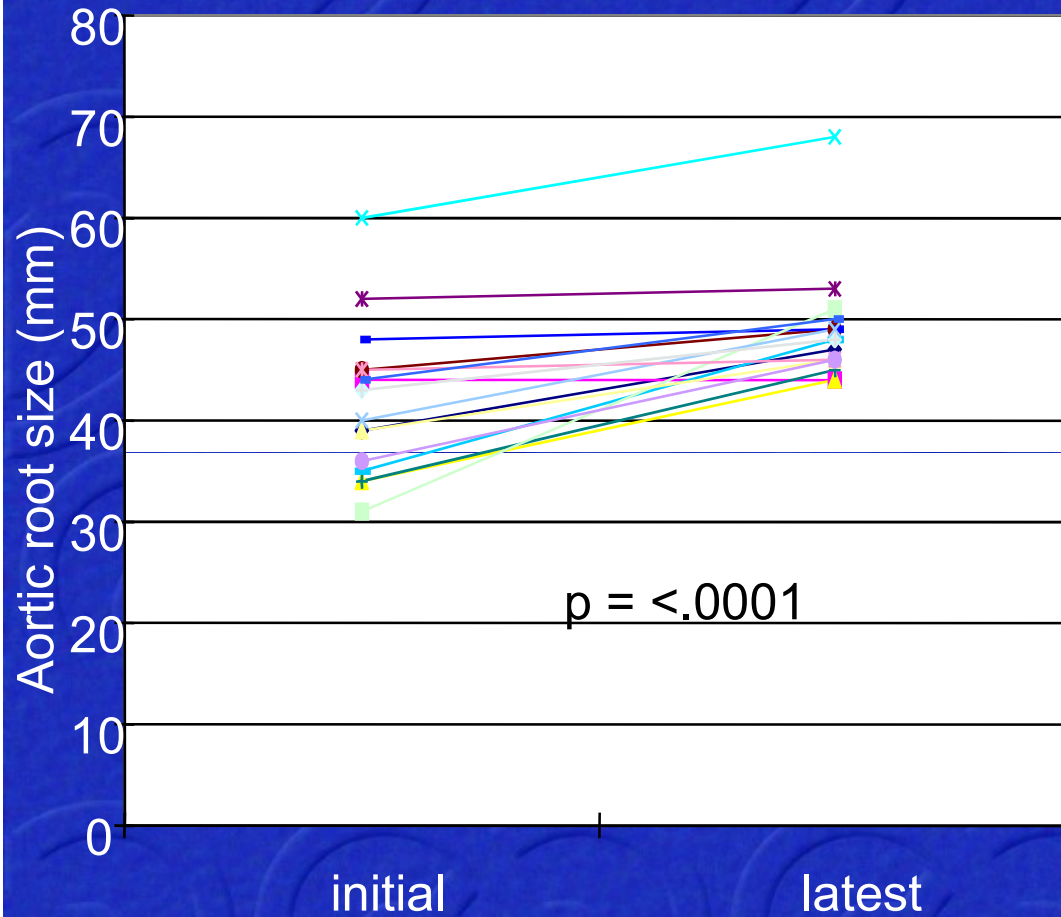
(n=101 consecutive adults with TET repair)



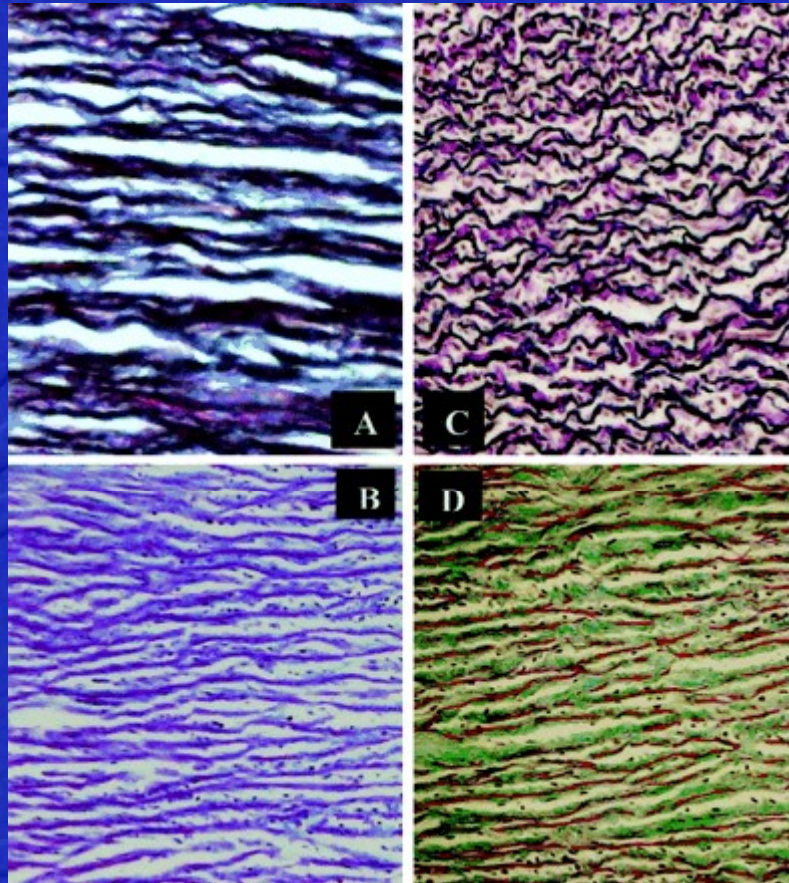
RVOT: akinesia & aneurysms...



Ao Root Dilatation



Ao Root Dilatation



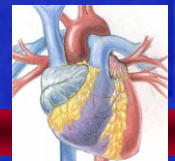
A and B are from a normal child.
C and D are from a case of TOF
aged 20 months.

C shows disruption of the elastic
lamellae

D shows increase in fibrous tissue
(green).

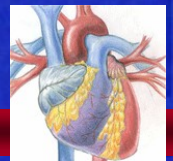
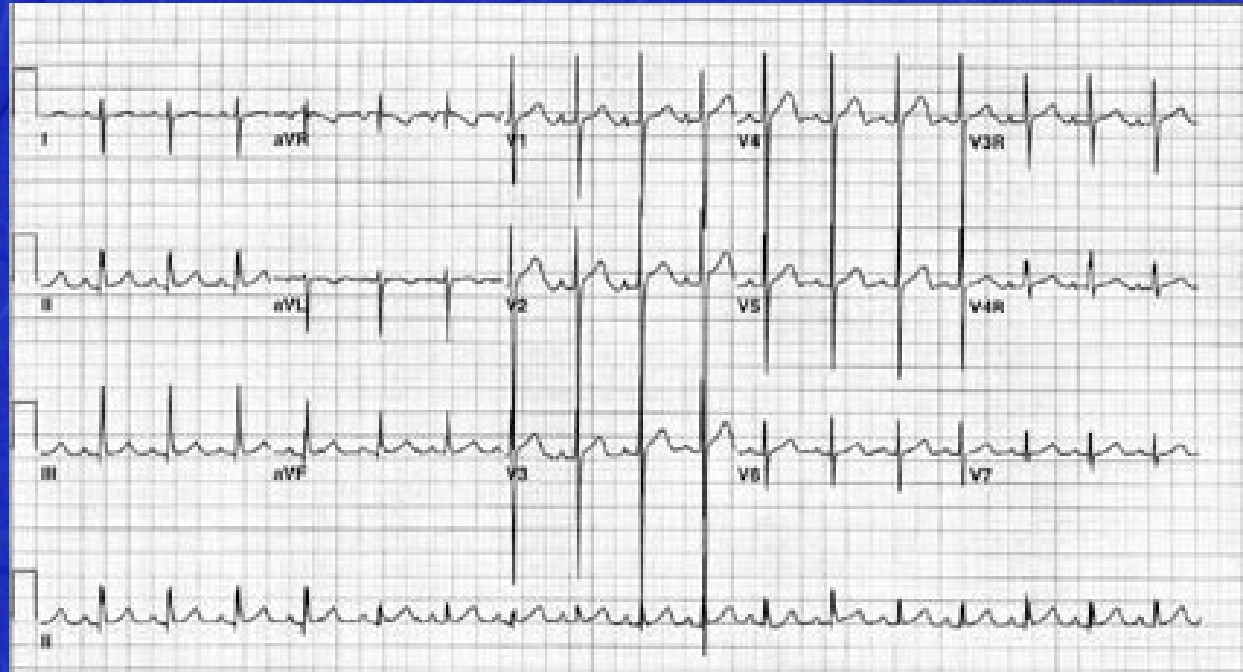
A and C are stained in elastic van
Gieson and B and D in trichrome.

Magnification x200

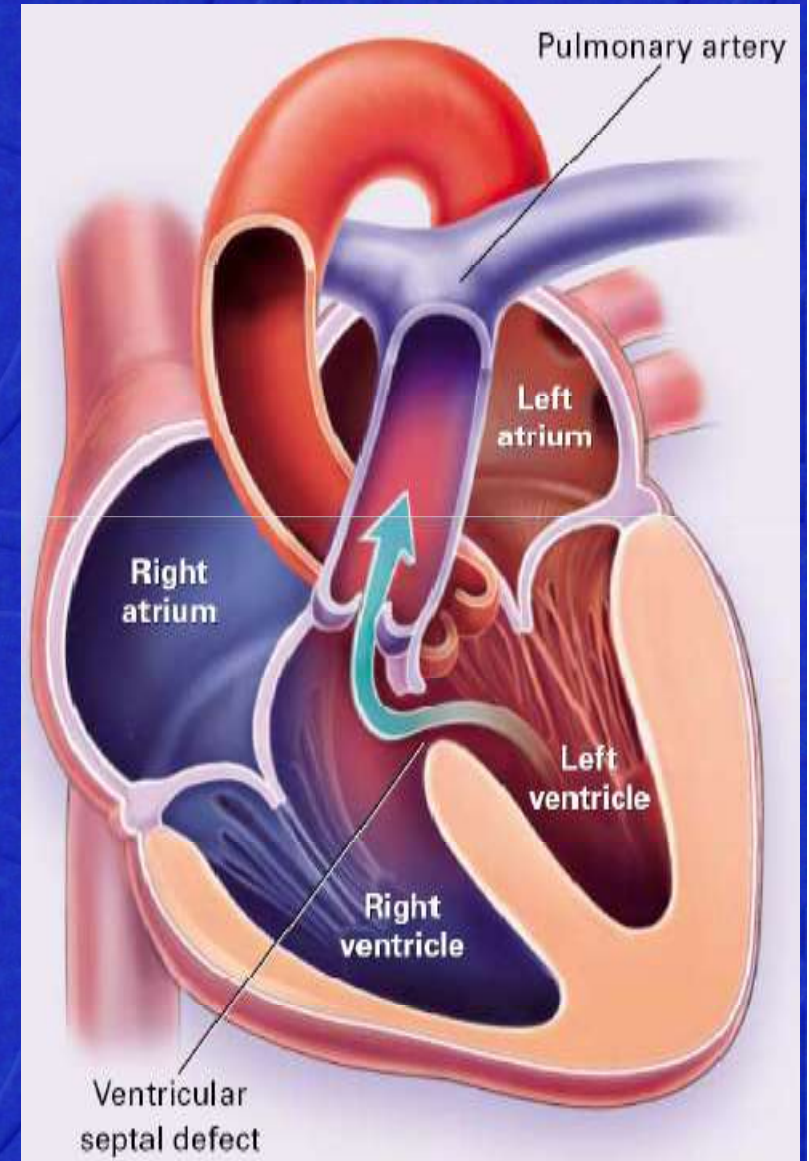
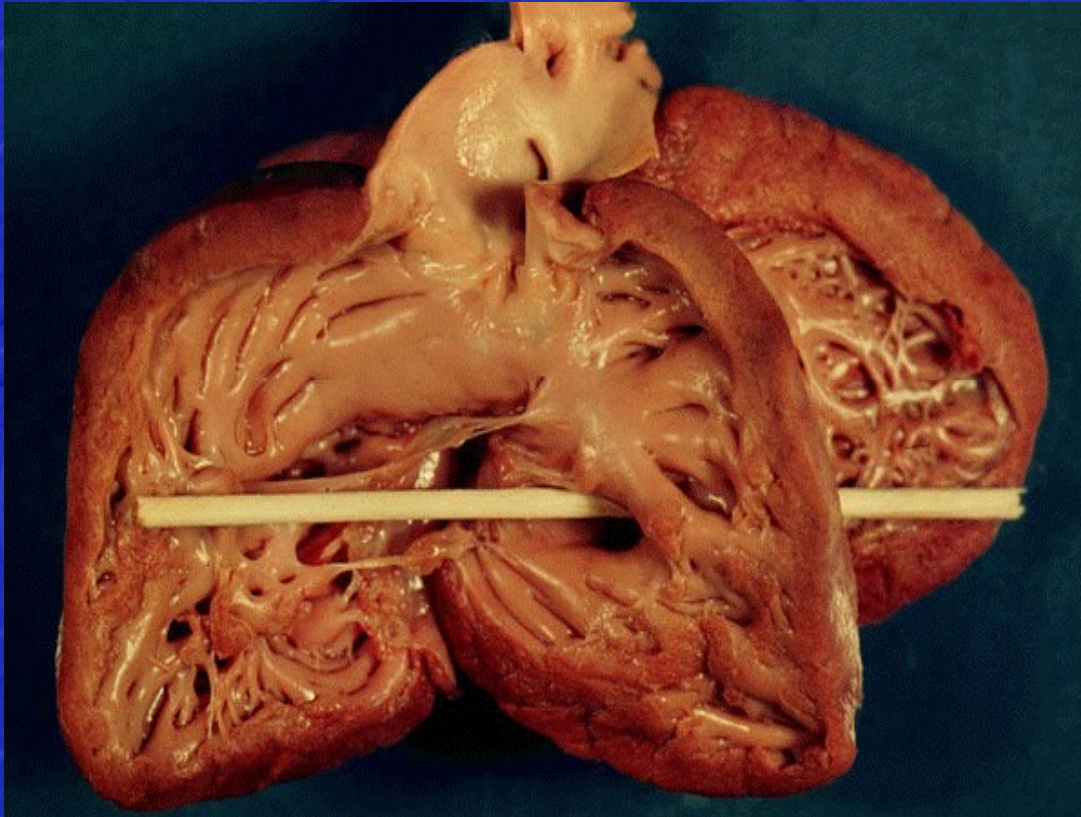


14 yrs old boy with spontaneously closed (age 1.5 yrs) membranous VSD...

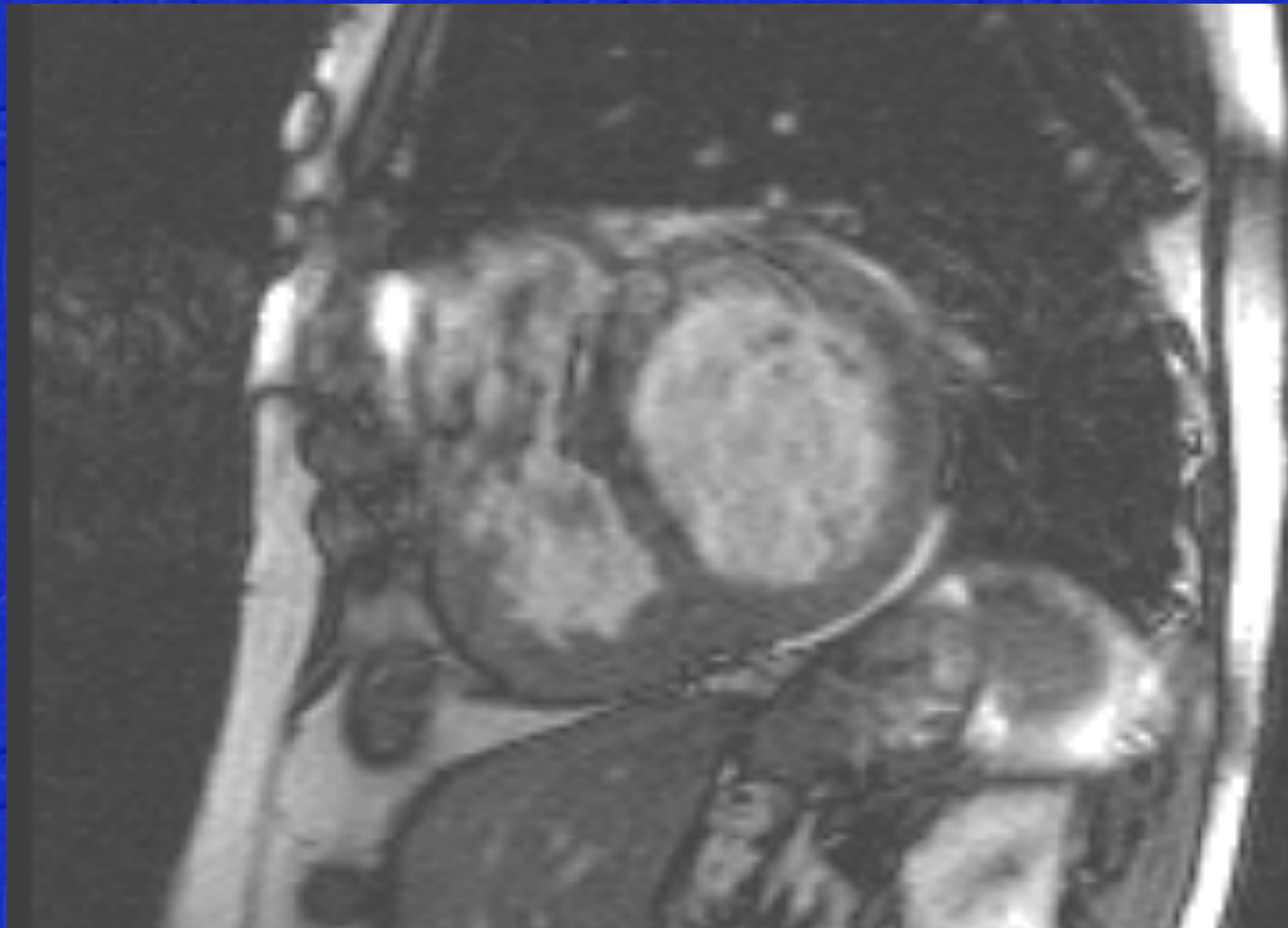
- Comes back with SOB and 4/6 murmur 2nd LICS
- CRX: ok...
- ECG: RAD, RVH
- SAT 96% Ht 43%



Ventricular Septal Defect (VSD)...



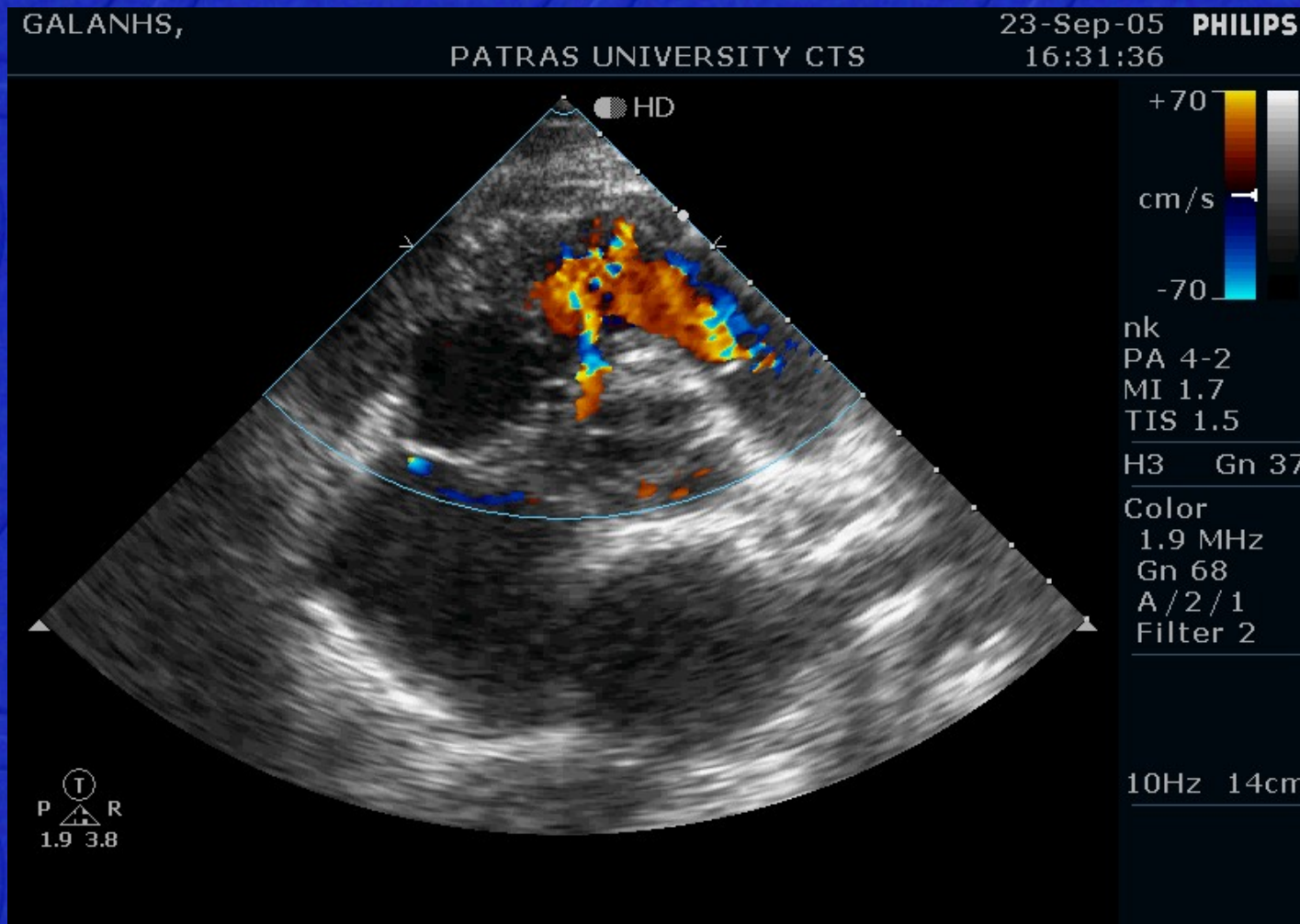
VSD: Complications...

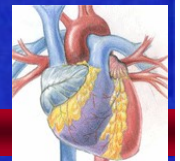
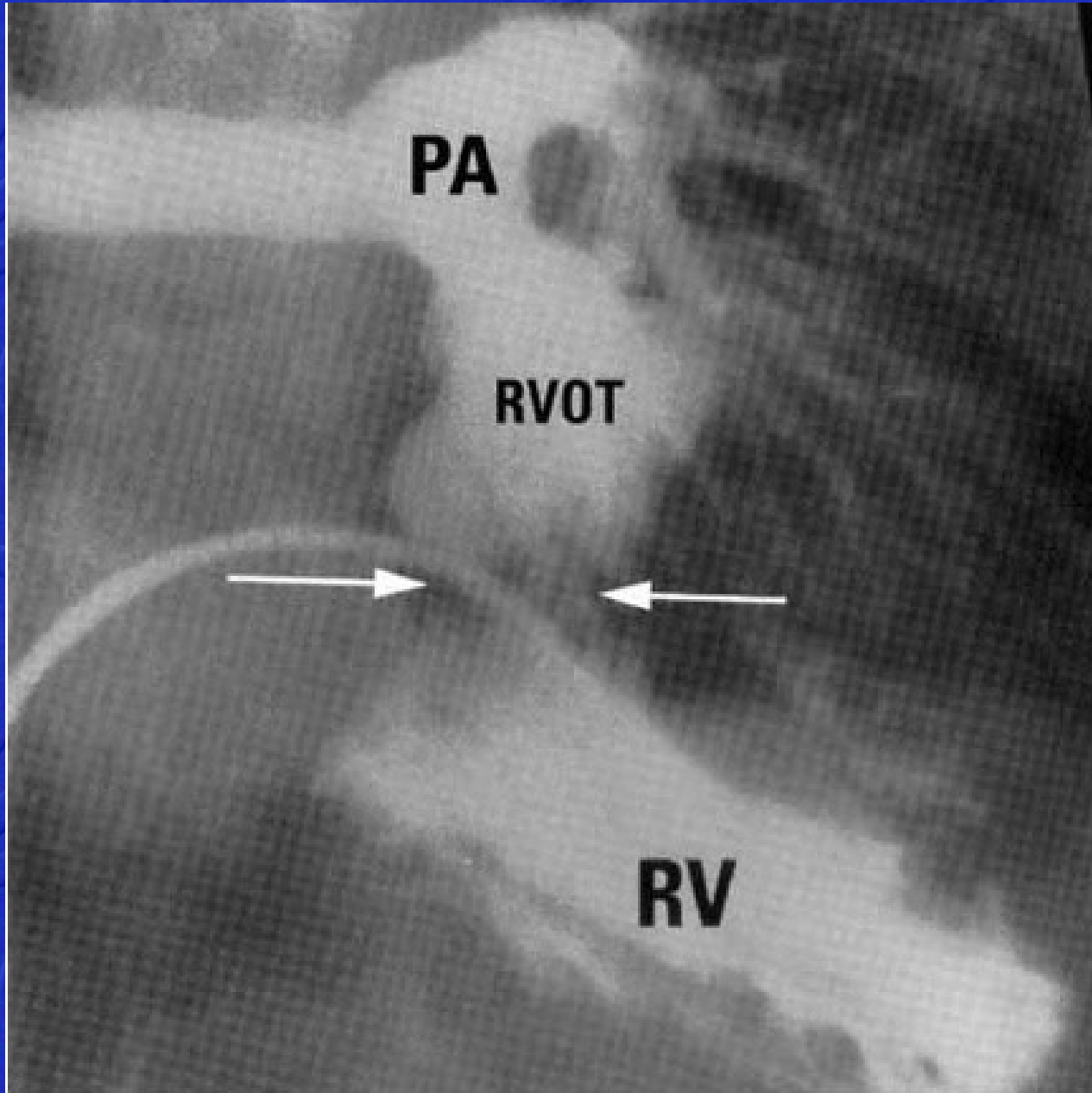


- RVOT obstruction (Double chamber RV)

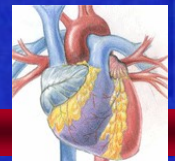
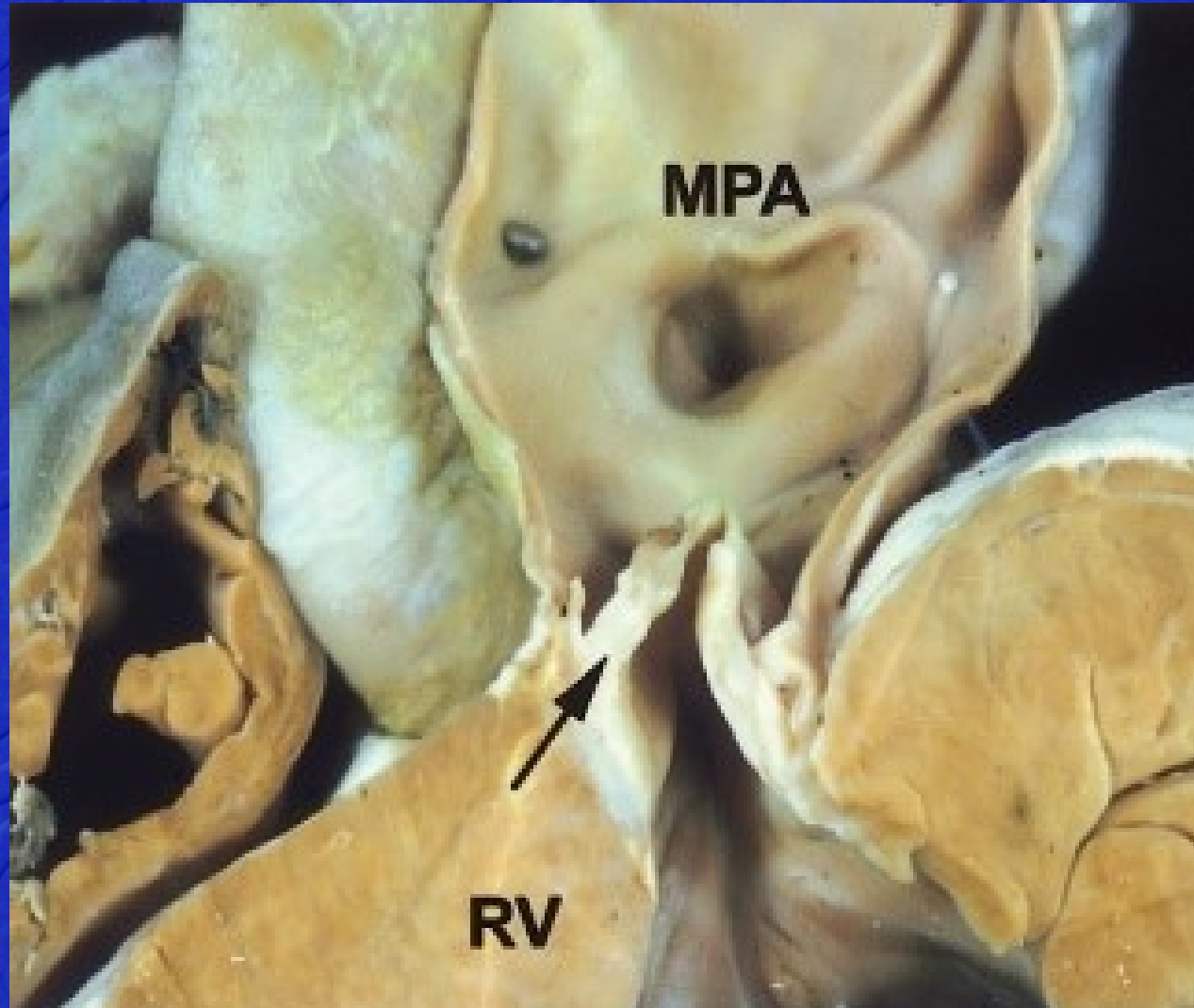


Double-chambered RV





Pulmonary Stenosis (PS)



Pulmonary Stenosis (PS)

Gyrosan NT Intera
CARDIAC-R9 T1W/BB-SAG

DELAPORTAS ' EVAGGELOS

M
CARDIAC

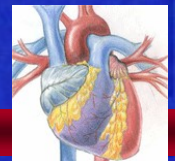
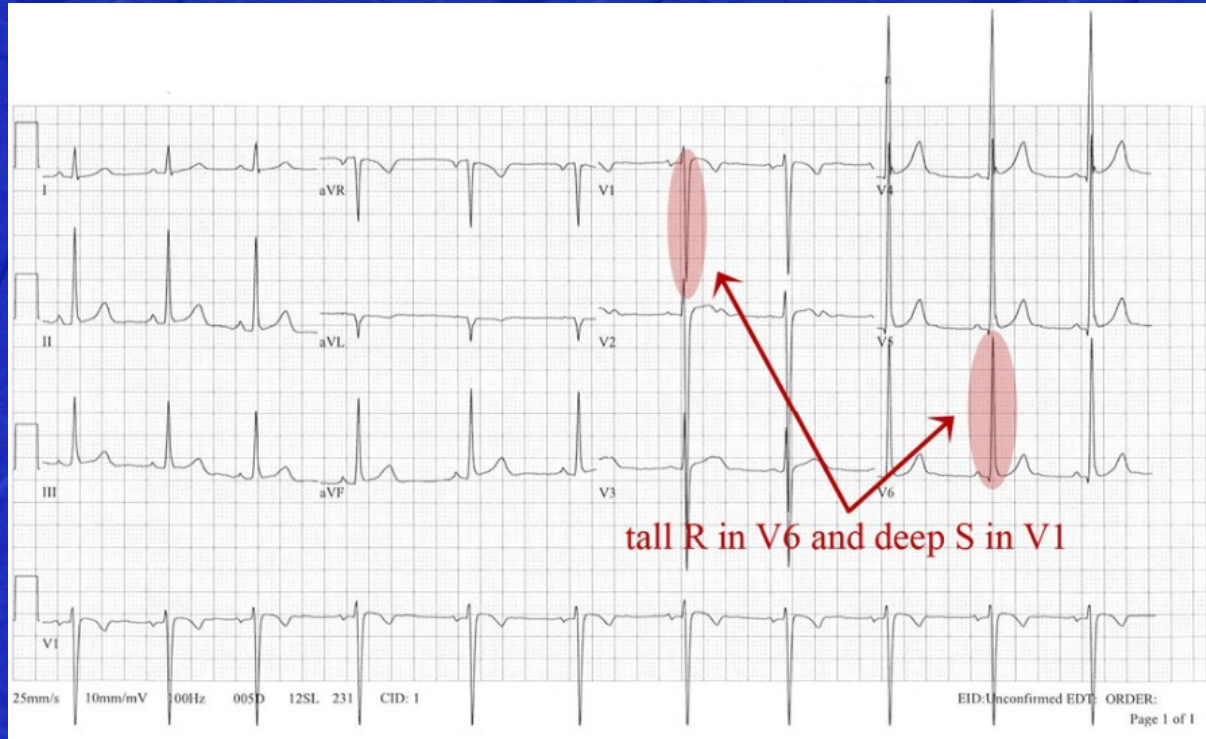


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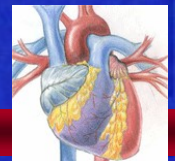
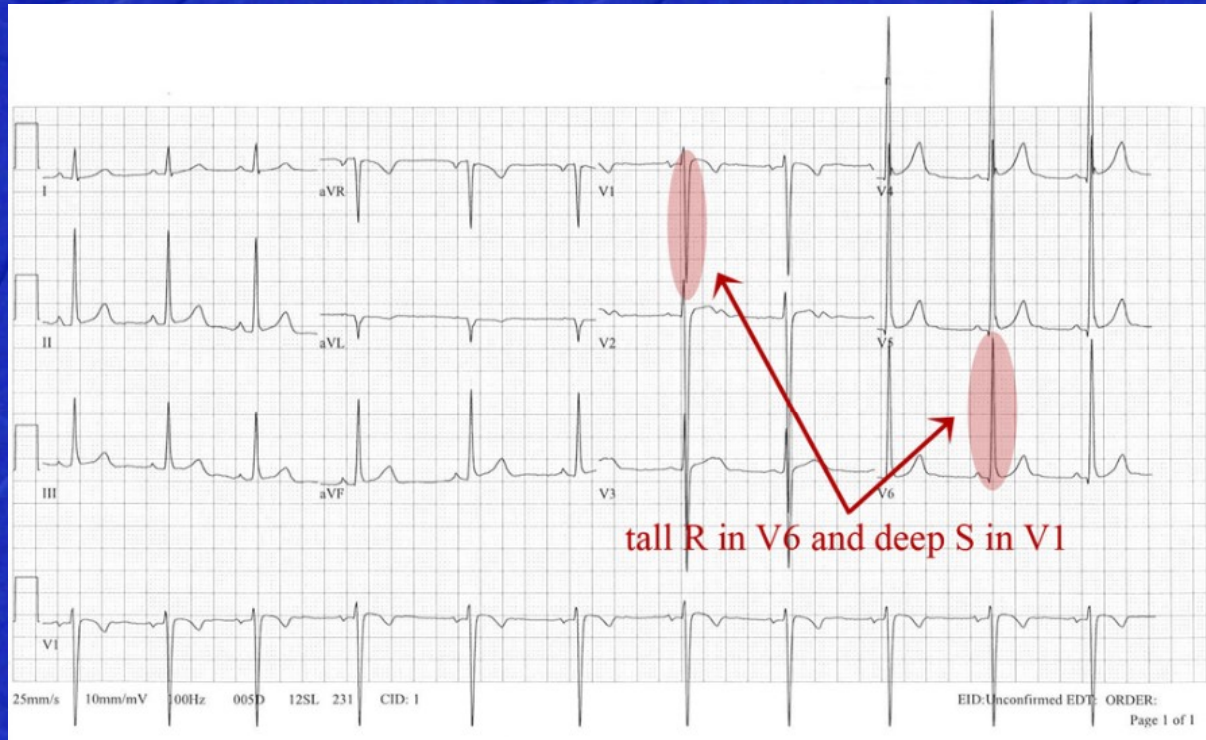
52 yrs old lady with HTN, Chol, FHx, angina, SOB and weakness...

- BP 178/90 mmHg
 - ECG =>
 - 2/6 systolic c-d
- 2nd LICS...

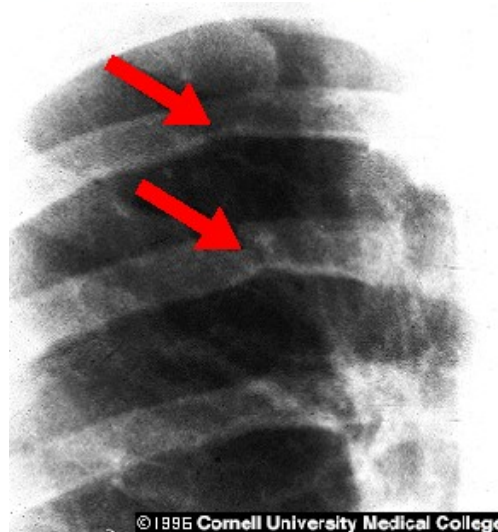
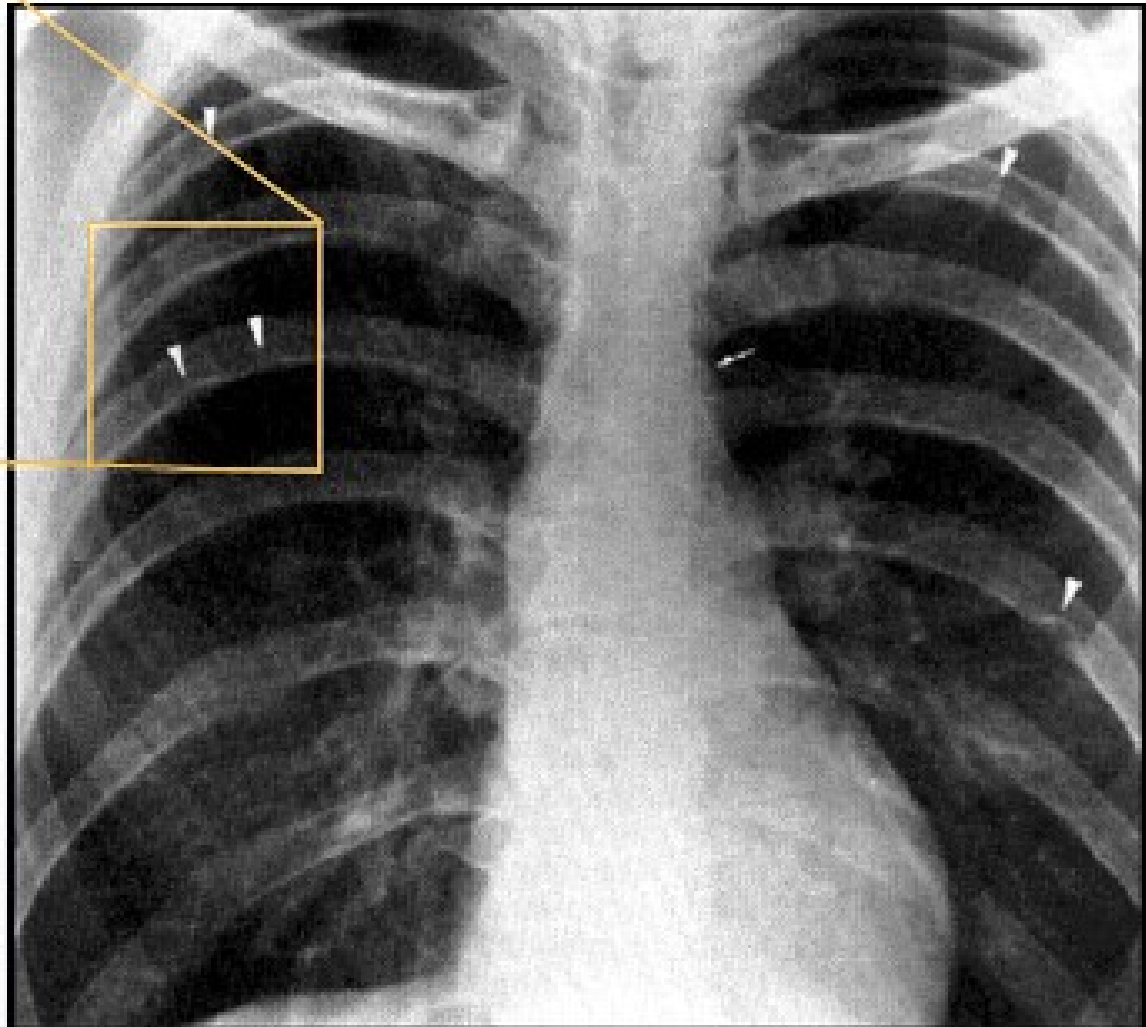
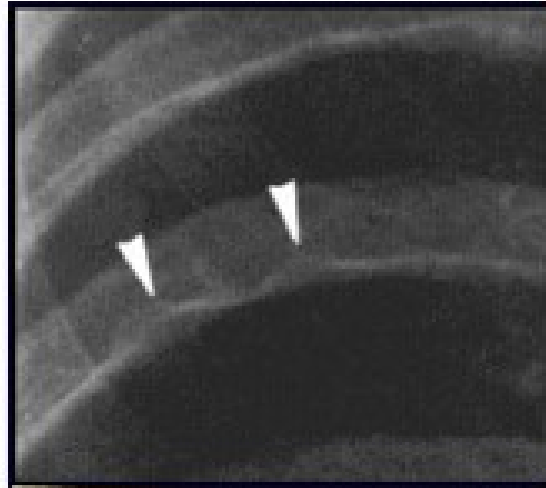


52 yrs old lady with HTN, Chol, FHx, angina, SOB and weakness...

- BP 178/90 mmHg
- ECG =>
- 2/6 systolic c-d
2nd LICS...
- Radio-femoral delay
- R-Leg BP 95/85mmHg



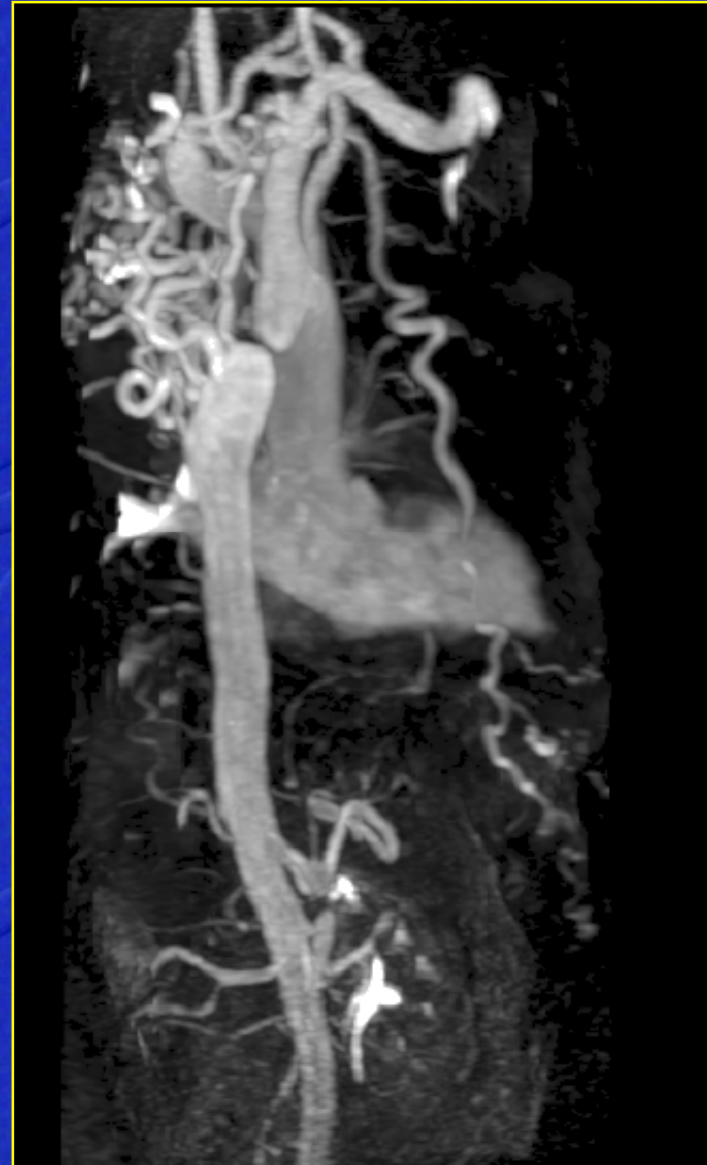
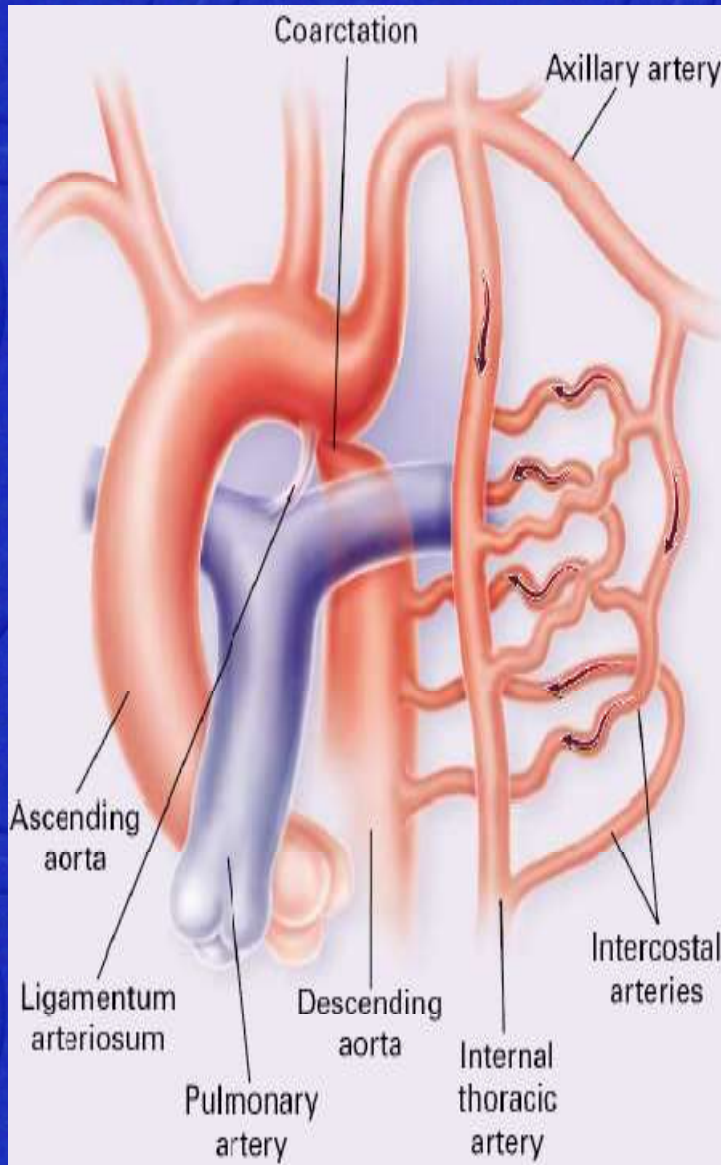
Rib notching



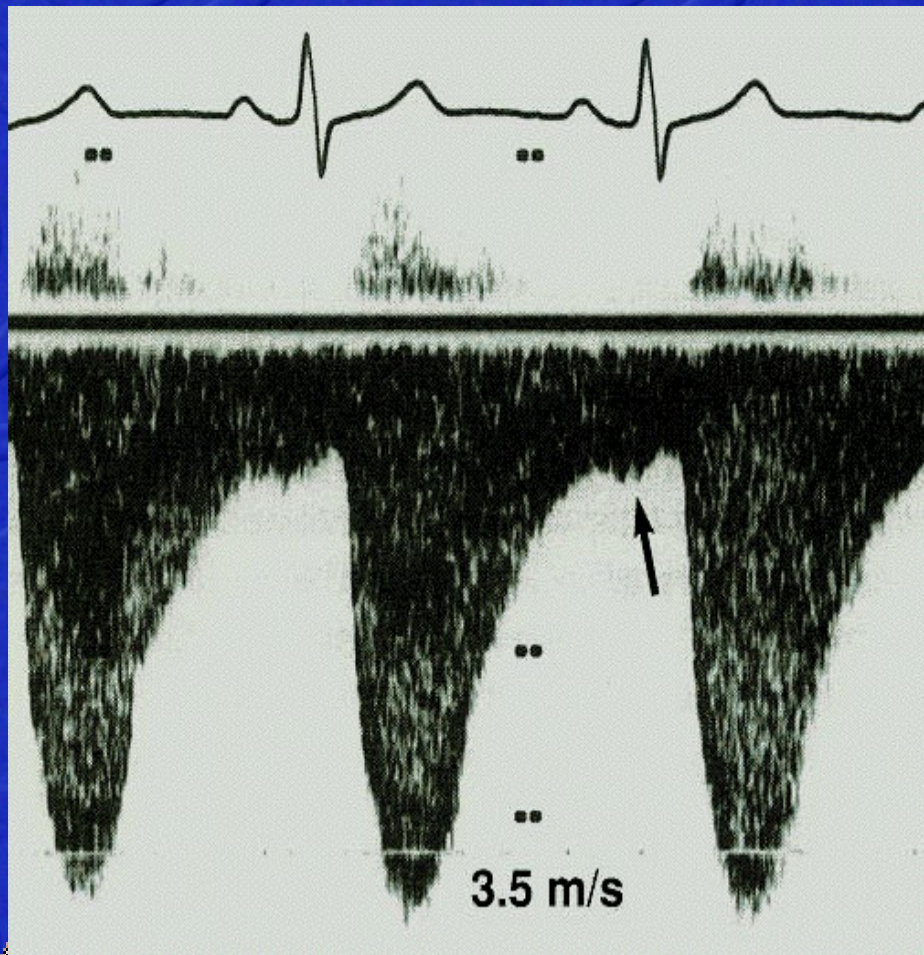
© 1995 Cornell University Medical College



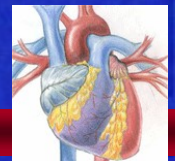
Coarctation of the Aorta (CoA)...



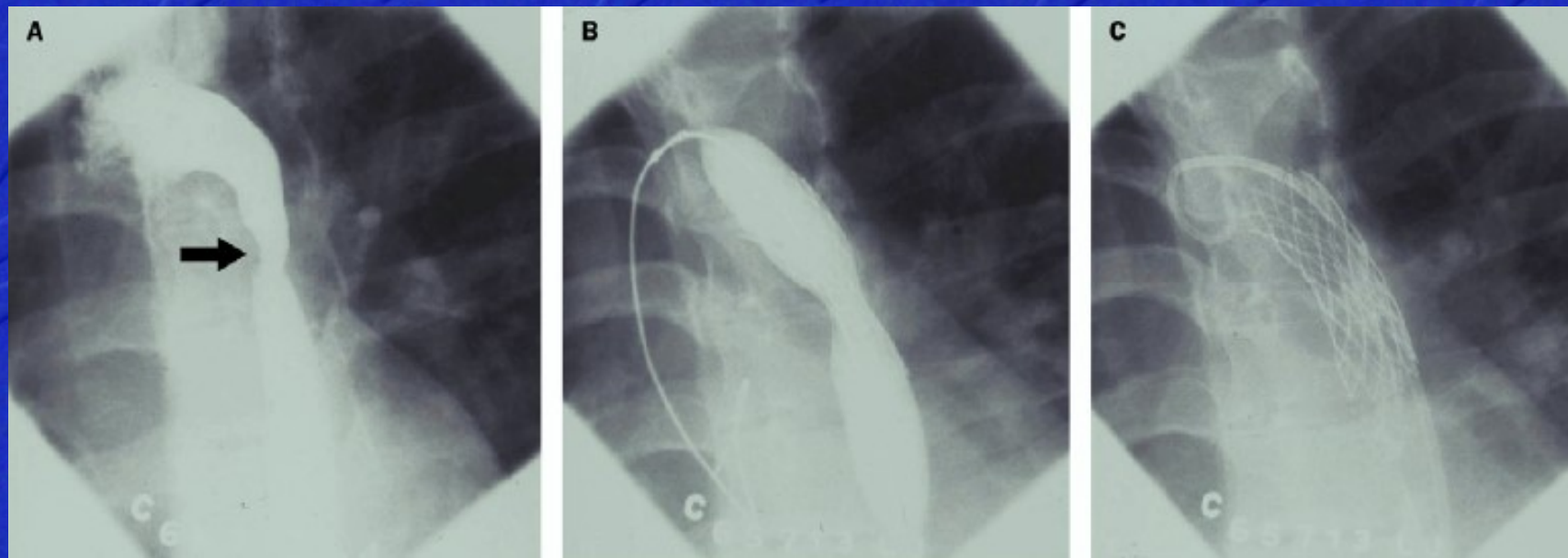
Coarctation of the Aorta (CoA)...



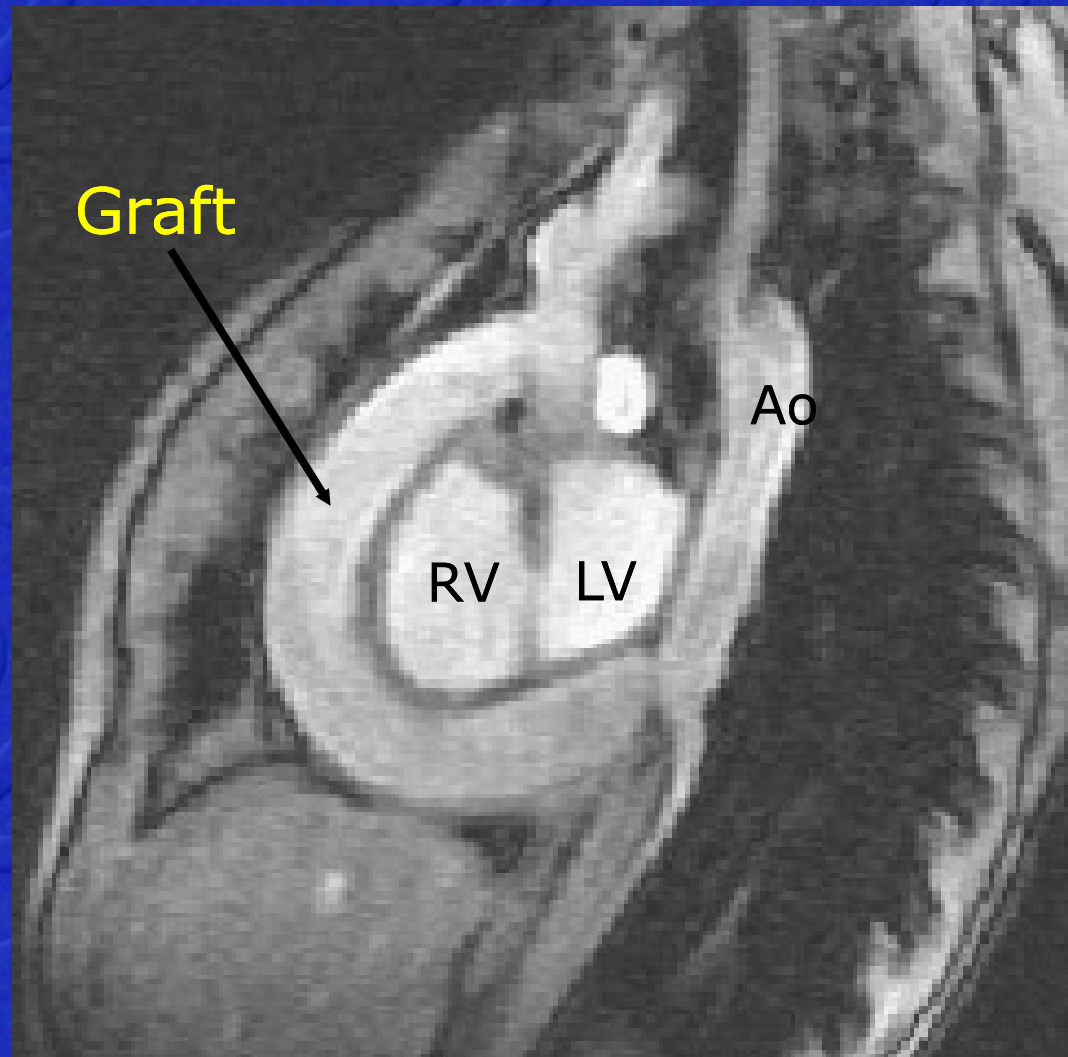
- HTN?
- RA-RL pressure ↓
- Collaterals
- LVH
- Function
- AV



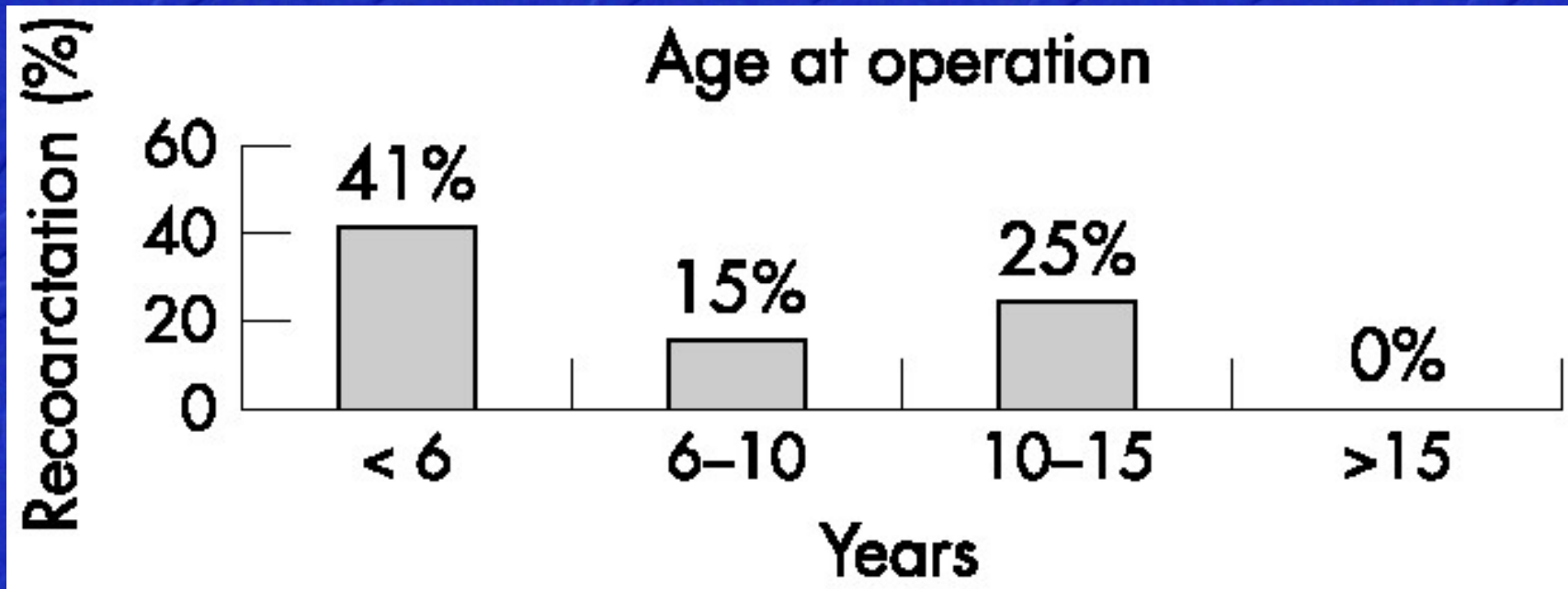
Coarctation of the Aorta (CoA)...



Coarctation of the Aorta (CoA)...



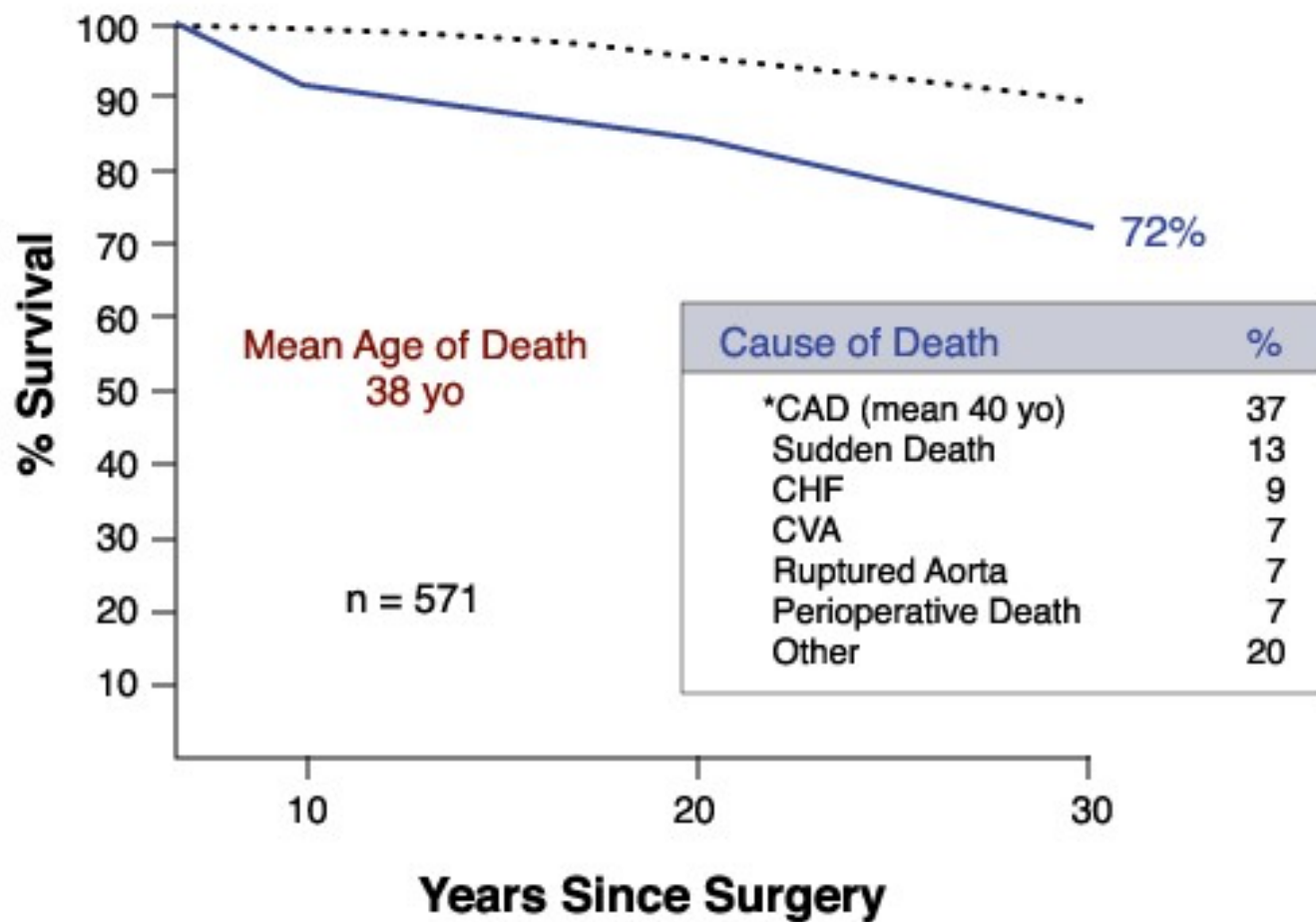
Coarctation of the Aorta (CoA)...



23% Re-CoA

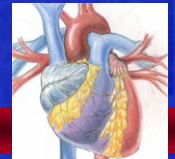


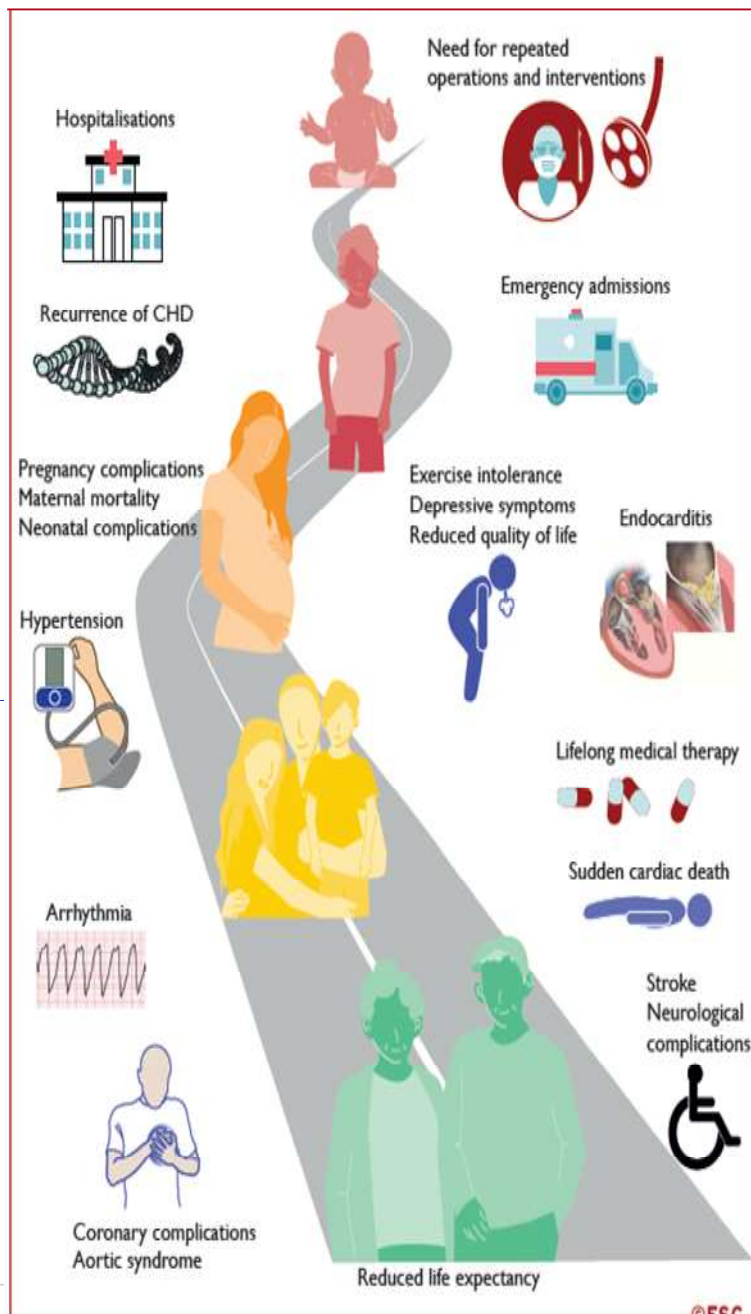
Survival After Coarctation Repair



Adult Congenital Heart Disease is an epidemic!

- Over 2 million in USA
- Most common birth defect
- Most (over 85%) now living to adulthood
- **Most common presenting in adulthood:** bicuspid aortic valve, coarctation of aorta, atrial septal defect
- **Most common post-op seen as adults:** tetralogy of Fallot, Transposition of Great Arteries (TGA)





Congenital heart disease A lifelong chronic condition



A lifetime of specialist care

Royal Brompton & Harefield **NHS**
NHS Foundation Trust

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▶ **Children's heart surgery review**

- See the [chronology](#) of the Safe and Sustainable review process
- Read about the IRP review



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▶ **Researchers find new way to predict risk of death in heart patients**

Health Minister describes incredibly important research

▶ **Highest numbers of staff recommend our hospitals**

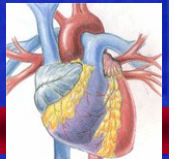
In a national survey published today, Royal Brompton & Harefield is identified as one of the most trusted NHS organisations in England.



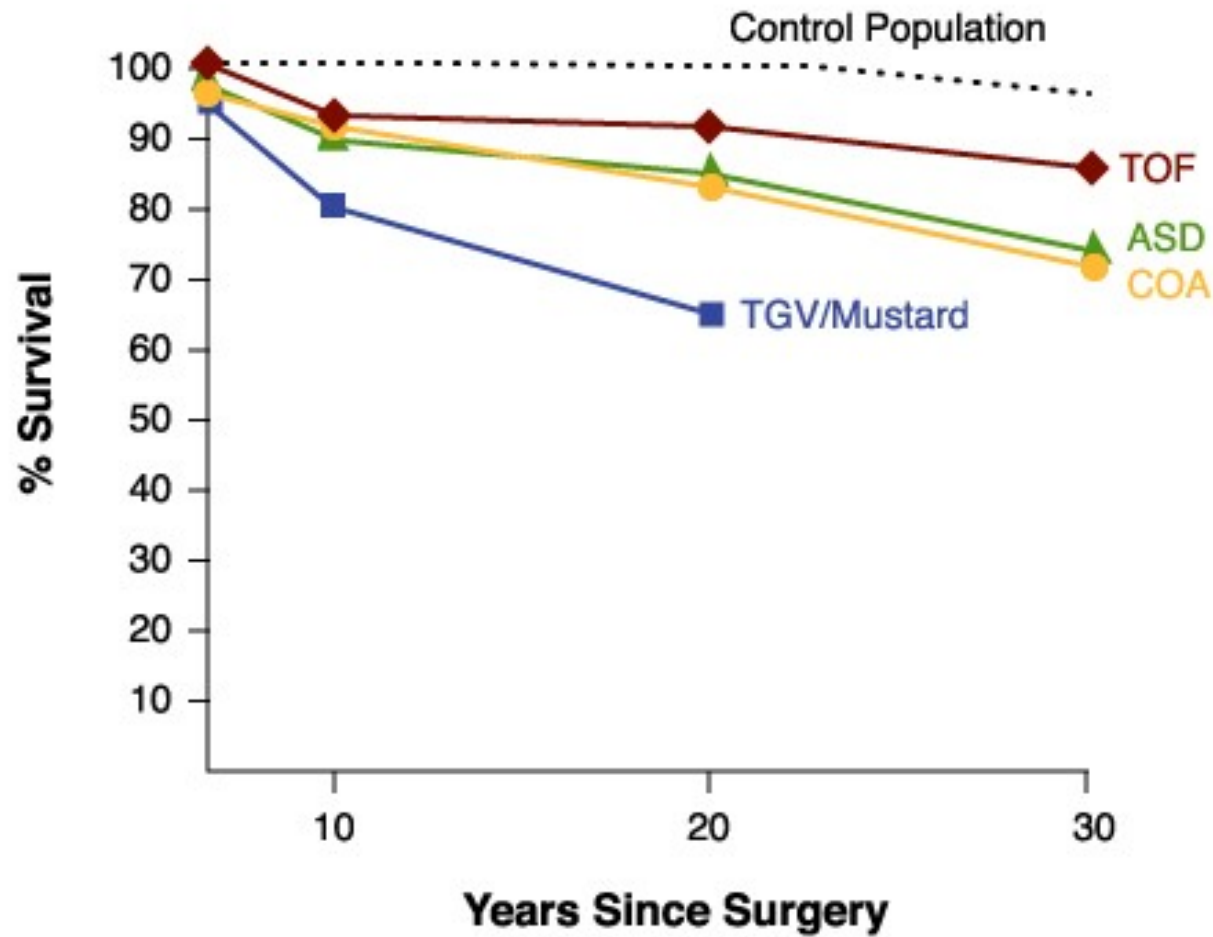
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Survival in ACHD

- Lesion specific
- Age at operation
- Type of operation
- Residual lesions
- Ventricular function
- Arrhythmias
- Follow up...

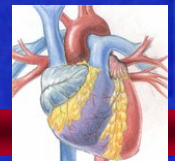


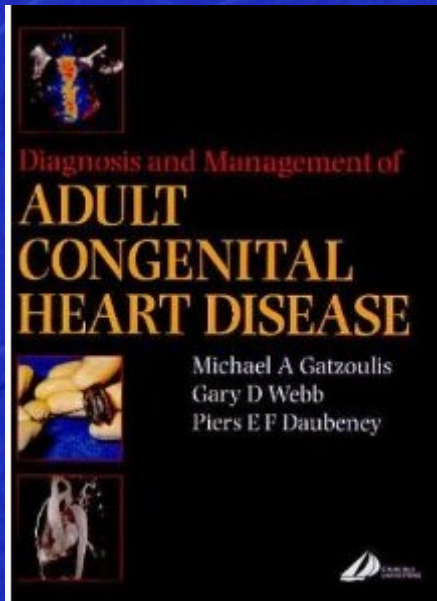
Survival Following CHD Surgery



Daniels, C.J. Congenital Heart Disease. ACCSAP V

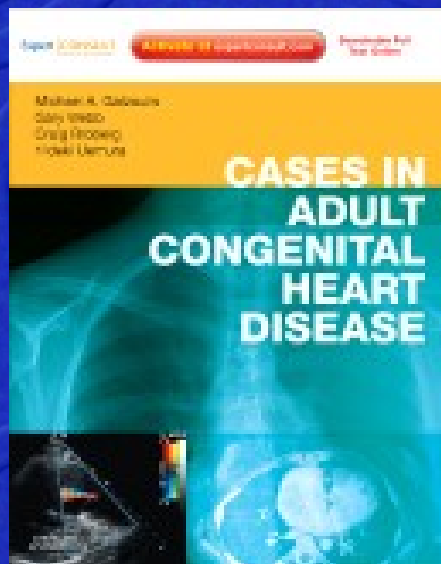
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Diagnosis and Management of Adult Congenital Heart Disease, Churchill Livingstone 2003

Michael A. Gatzoulis, Gary D. Webb, Piers E.F. Daubeny,



Cases in Adult Congenital Heart Disease - Expert Consult: Online and Print Atlas, Elsevier

By Michael A. Gatzoulis, MD, PhD, Gary D. Webb, MD, Craig Broberg, MD and Hideki Uemura, MD, FRCS



Patras University Hospital



Thomas-Blalock-Taussig Shunt



Vivien Thomas



Alfred Blalock

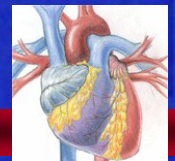


Helen Taussig



Something the Lord Made - Best Made-for-TV Movie, 2004

Patras University Hospital



Dr. Blalock does the Blalock
(Johns Hopkins)

