



ATRIOVENTRICULAR SEPTAL DEFECTS (AVSD)



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Definition:

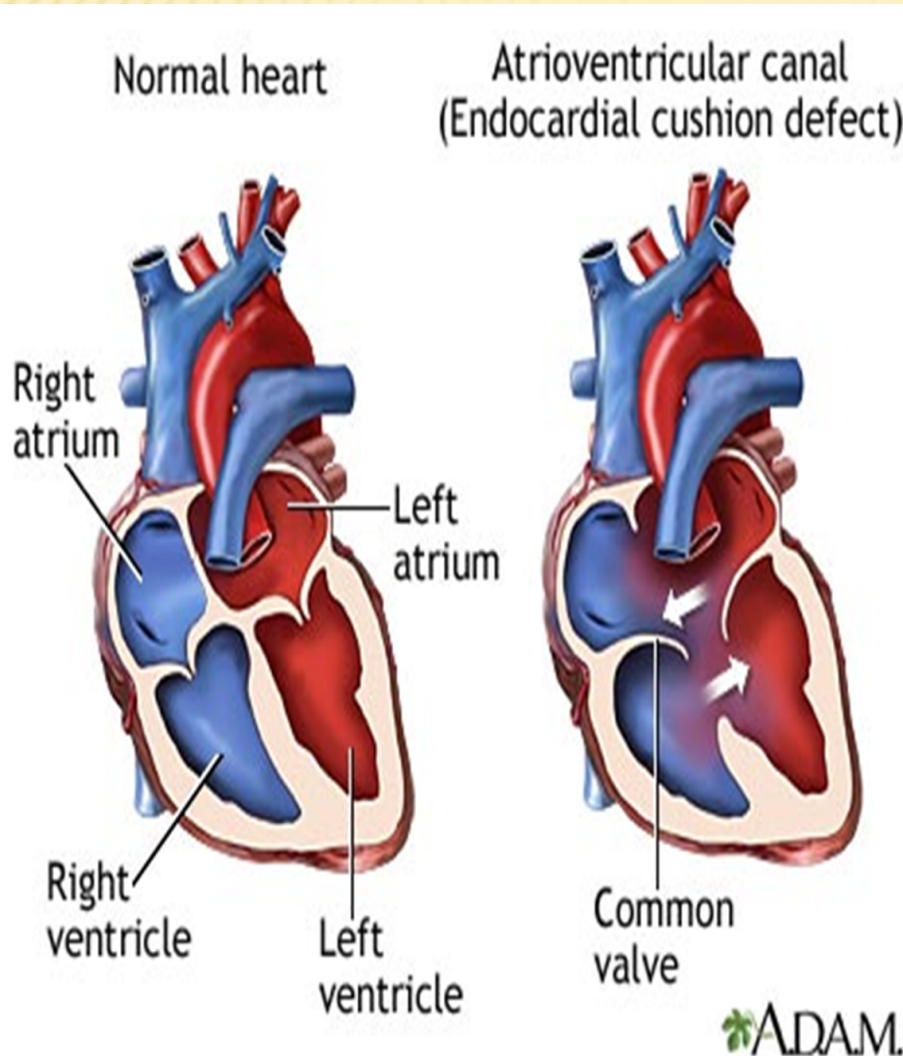
A DEFECT OF THE AV SEPTUM - ABNORMALITIES OF THE AV VALVES.

- Terms:

Atrioventricular Septal Defects (AVSD)

= Atrioventricular canal defects (AVC)

= Endocardial cushion defects (ECD)



Frequency

- 4-5% of CHD
 - 0.19 in 1000 live births
 - 18% of abnormal fetal hearts.
-

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- 40-50% Down syndrome have CHD - # 40% have an AVSD.
- The Male: Female ratio # 1
- AVSD Mother -> 9.6-14.3% children CHD

HISTORY

- Infants with cAVSD -> CHF

(*Berger : only 54% of patients born with cAVSD were alive at 6 month-age, 35% at 12m, 15% at 24m and 4% at 5y*)

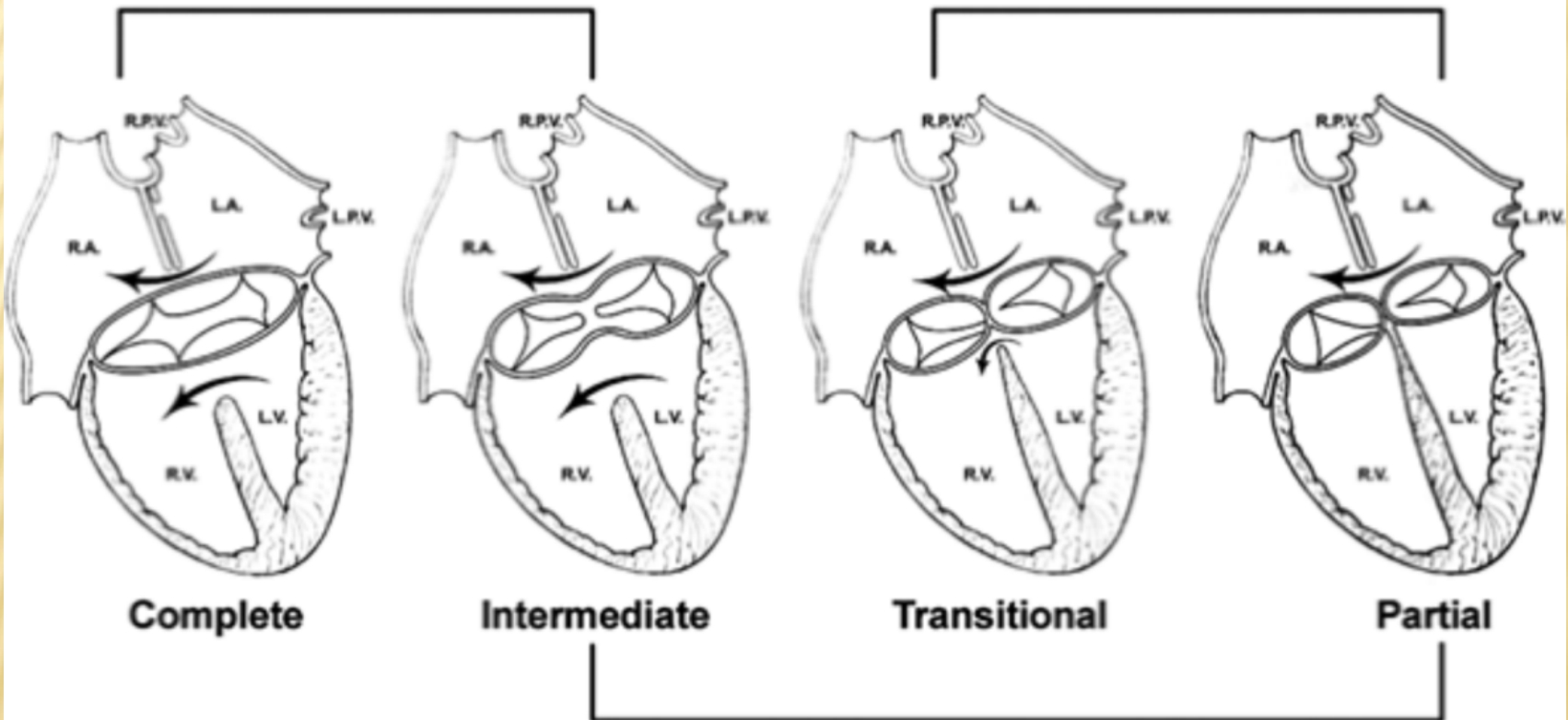
- Nonrestrictive o.p ASD -> Left AV valve regurgitation -> CHF , mortality rate of 33%/1st year.

CLASSIFICATION

AVSD Summary

Similar physiology – VSD & ASD

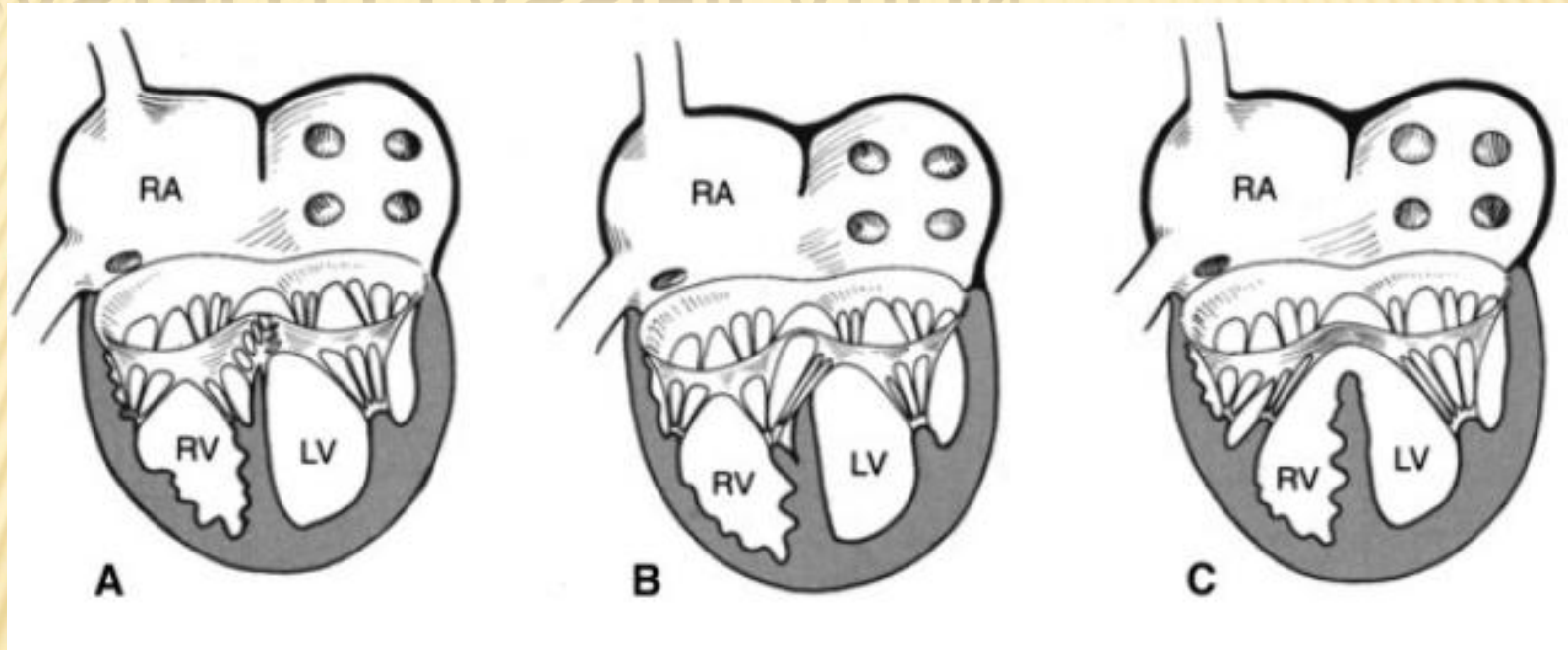
Similar physiology – ASD



Similar AV valve anatomy:

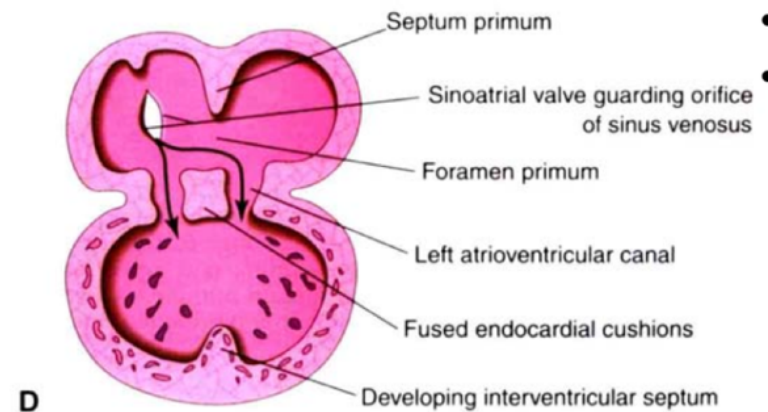
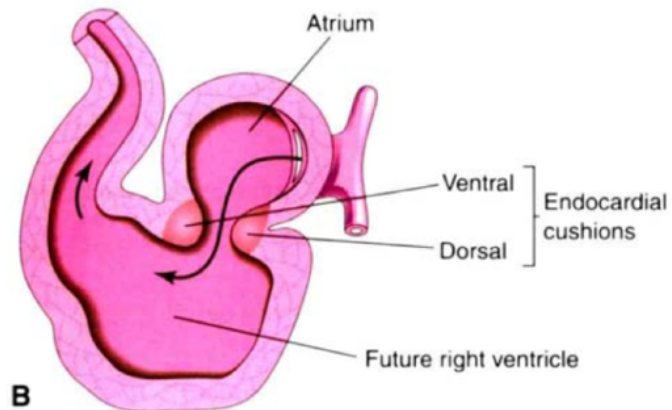
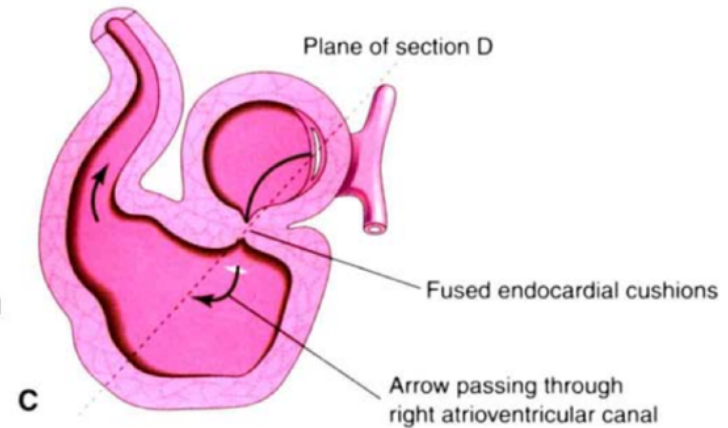
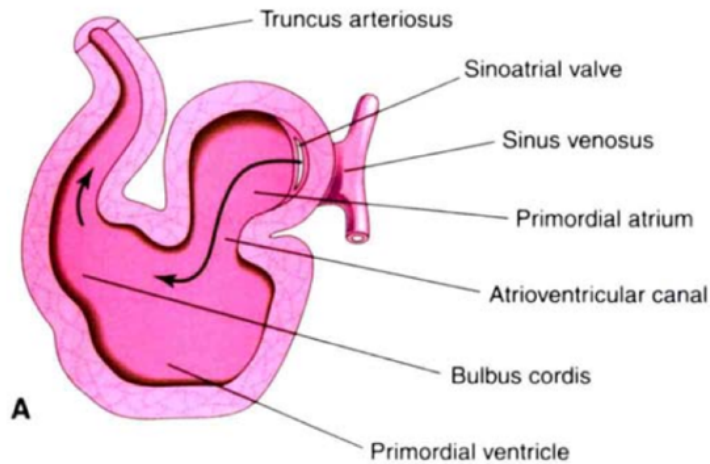
A tongue of tissue divides the common AV valve into a right and left component by connecting the anterior and posterior "bridging" leaflets centrally

RASTELLI CLASSIFICATION



3 types of complete AVSD based on the morphology of the anterior bridging leaflet, its degree of bridging and its chordal attachments

EMBRYOLOGY



Weeks 4-5

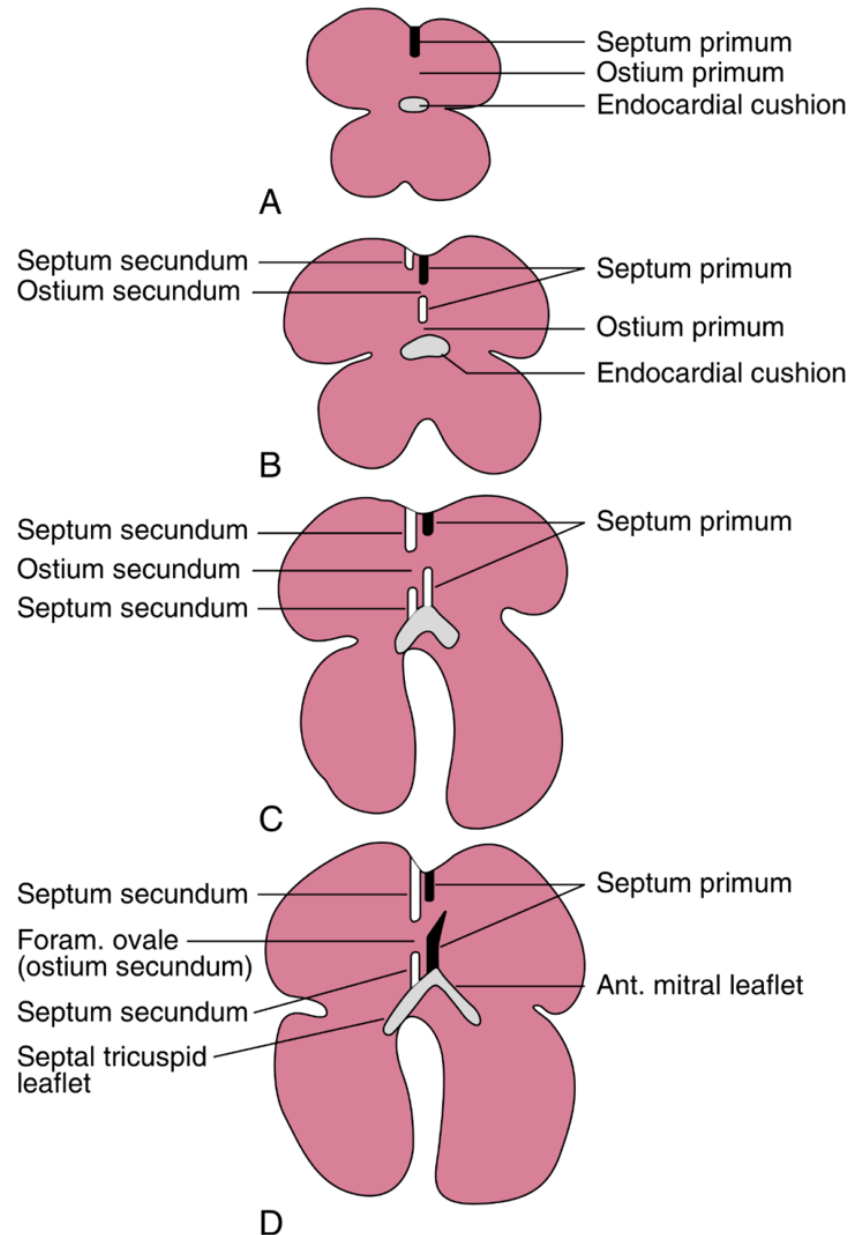
From Moore & Persaud 1998

EMBRYOLOGY

***Partial AVSDs,
incomplete fusion of the
superior and inferior
endocardial cushions**

***Complete AVSD is
associated with lack of
fusion**

Perloff 's Clinical Recognition of CHD
6th edition – Figure 15.1 – p213



CONOTRUNCAL ANOMALIES (BULBUS CORDIS)

- Associated with AVSD:
 - Tetralogy of Fallot (most common, # 6%)
 - Double outlet right ventricle
 - Transposition of the great arteries

MAIN FEATURES:

✗ **Partial AVSD:**

- A primum atrial septal defect (ASD)
- 2 distinct mitral and tricuspid valve annuli.
- Cleft of the mitral valve.

✗ **Complete AVSD:**

- A primum ASD – contiguous with an inlet VSD.
- Single annulus.

HEMODYNAMICS

- Excessive pulmonary blood flow (L-R shunt)
- No interventricular communication -> # large ASD -> increased RV stroke volume.
- cAVSD : large LR shunt -> RV- PA pressure# systemic pressure
 - > pulmonary hypertension from birth
 - > have rapid progression of pul.vascular disease

PHYSICAL APPEARANCE



Down syndrome – accelerated development of pulmonary hypertension.

CLINICAL FEATURES

Partial AVSD:

asymptomatic until adulthood

fatigue, tachypnea, and failure to thrive.

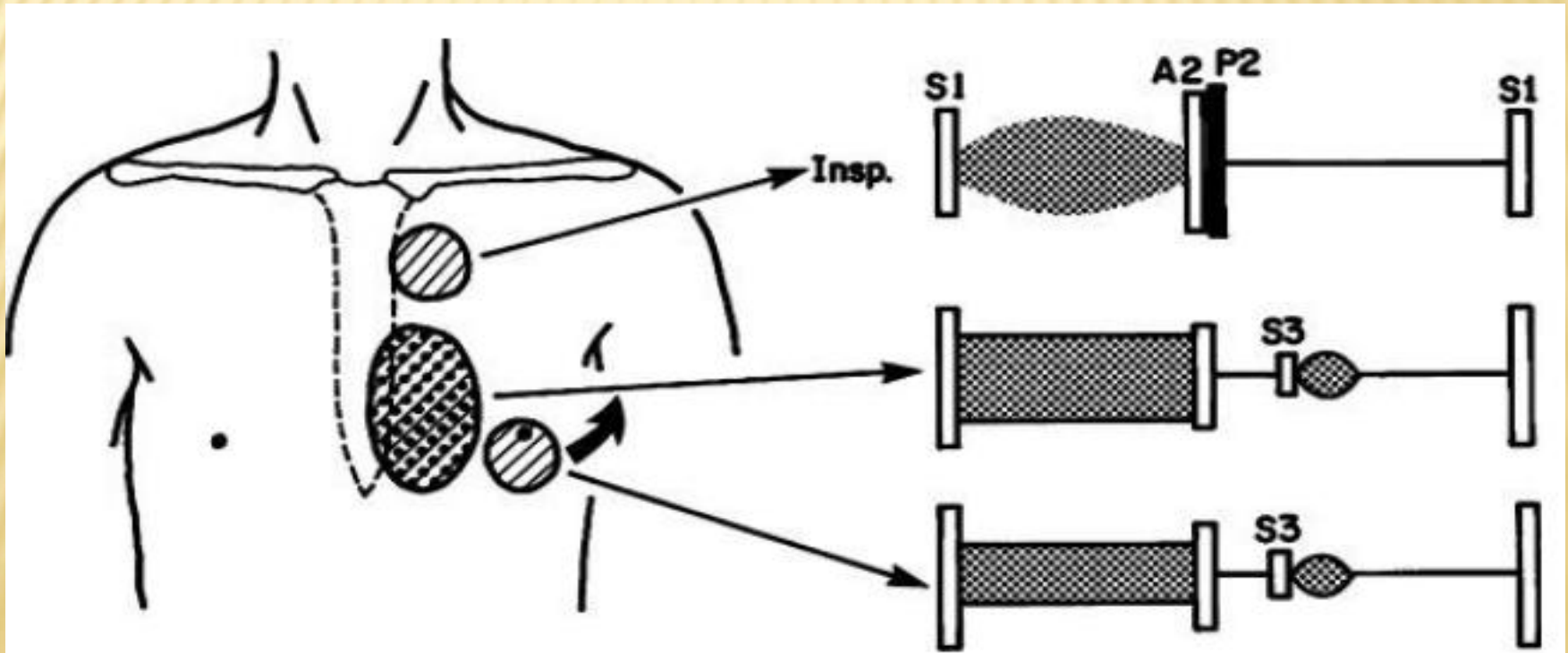
Complete AVSD:

CHF in infants

- Frequent upper respiratory tract infection
- Poor feeding – weight gain
- Sweating
- symptoms by 1 year-old

EXAMINATION:

- typical **systolic ejection** / the ULSB
- The widely split 2nd sound
- **holosystolic murmur at the apex.**
- Diastolic murmur / the LLSB



CHEST XRAY

- cardiomegaly
- pulmonary vascular markings.

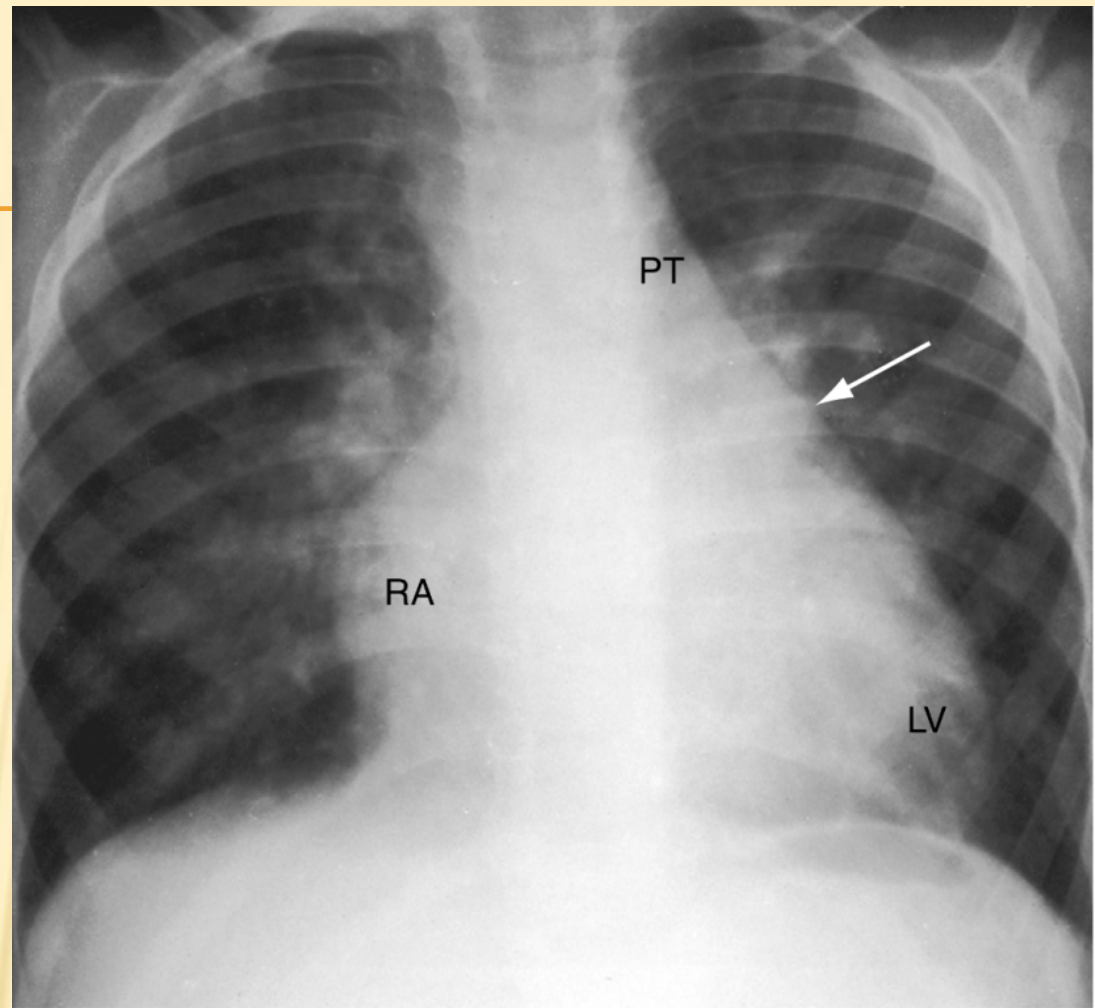


FIGURE 15-103 X-ray from a 19-month-old female with a non-restrictive ostium primum atrial septal defect, a restrictive inlet ventricular septal defect, a 2 to 1 left-to-right shunt, and moderate to marked regurgitation of the left atrioventricular valve. An enlarged left atrial appendage (arrow) straightened the left cardiac border. A prominent right atrium (RA) occupies the right lower cardiac border, and a dilated left ventricle (LV) occupies the apex. Pulmonary vascularity is increased.

ECG

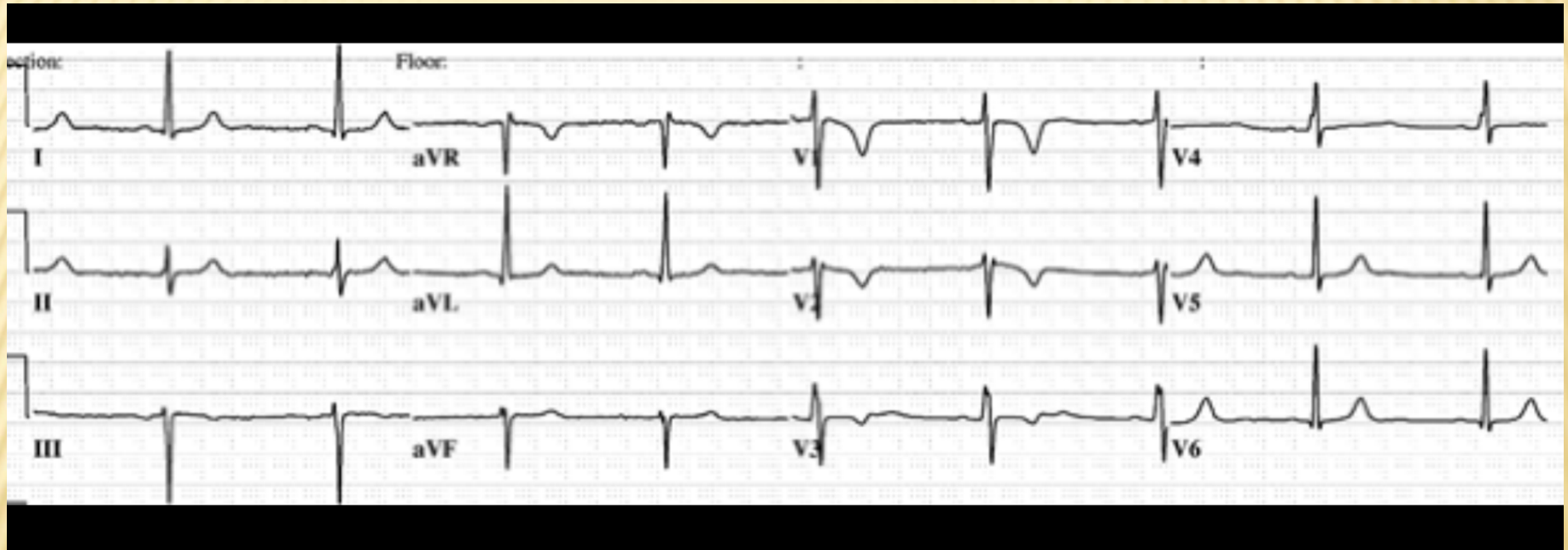
- The AV node is more posterior and inferior (near the coronary sinus ostium) than normal
 - The bundle of His usually courses along the inferior rim of the VSD
- > northwest or leftward axis, a prolonged PR interval, RBBB on electrocardiogram.

ECG



A 3 month-old with complete AVSD –
Left axis deviation and right ventricular hypertrophy.

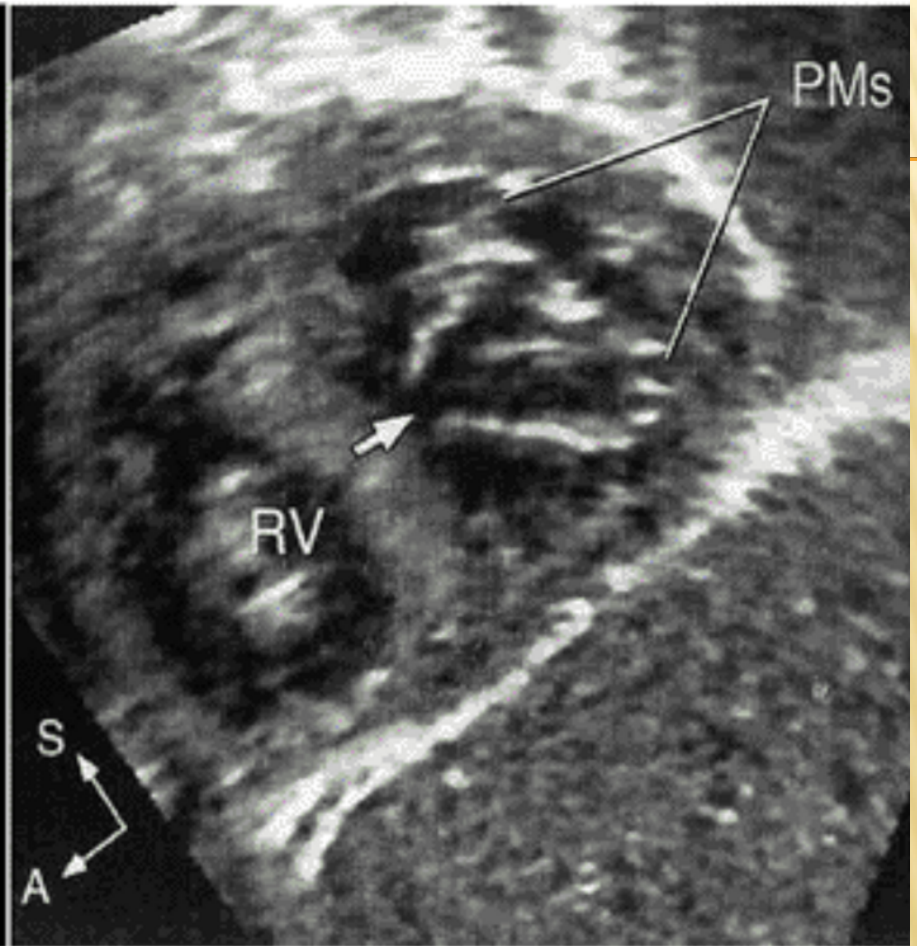
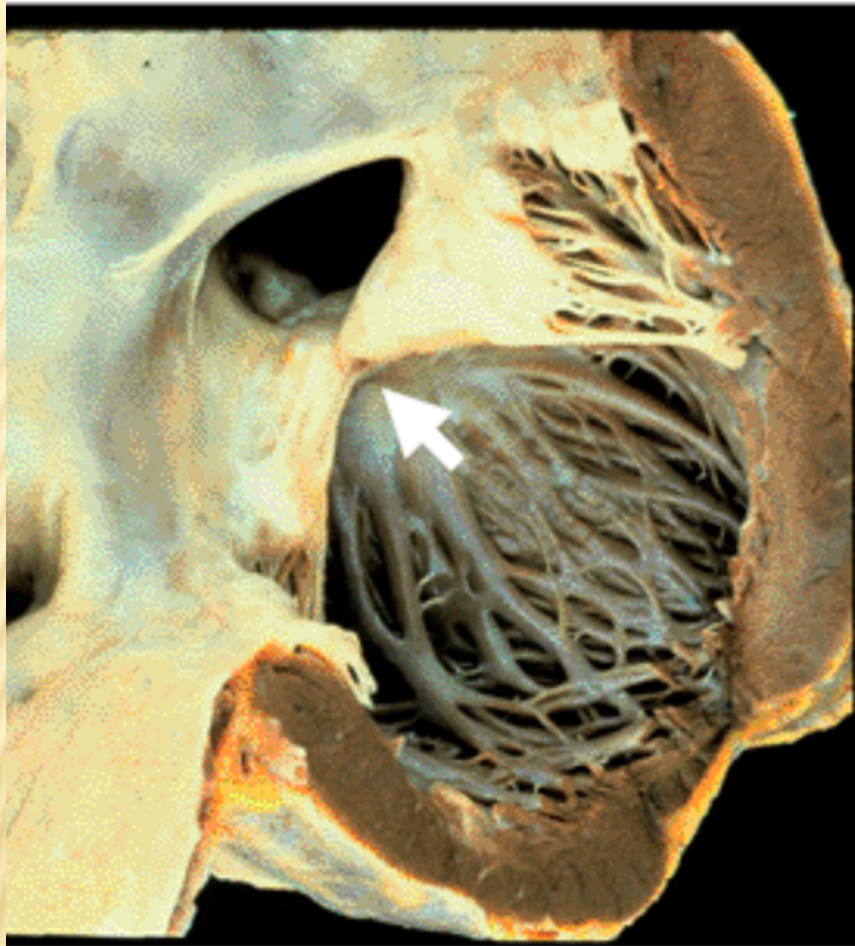
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A 17 year old had partial AVSD repair in childhood.
Left axis deviation and nonspecific T wave changes.

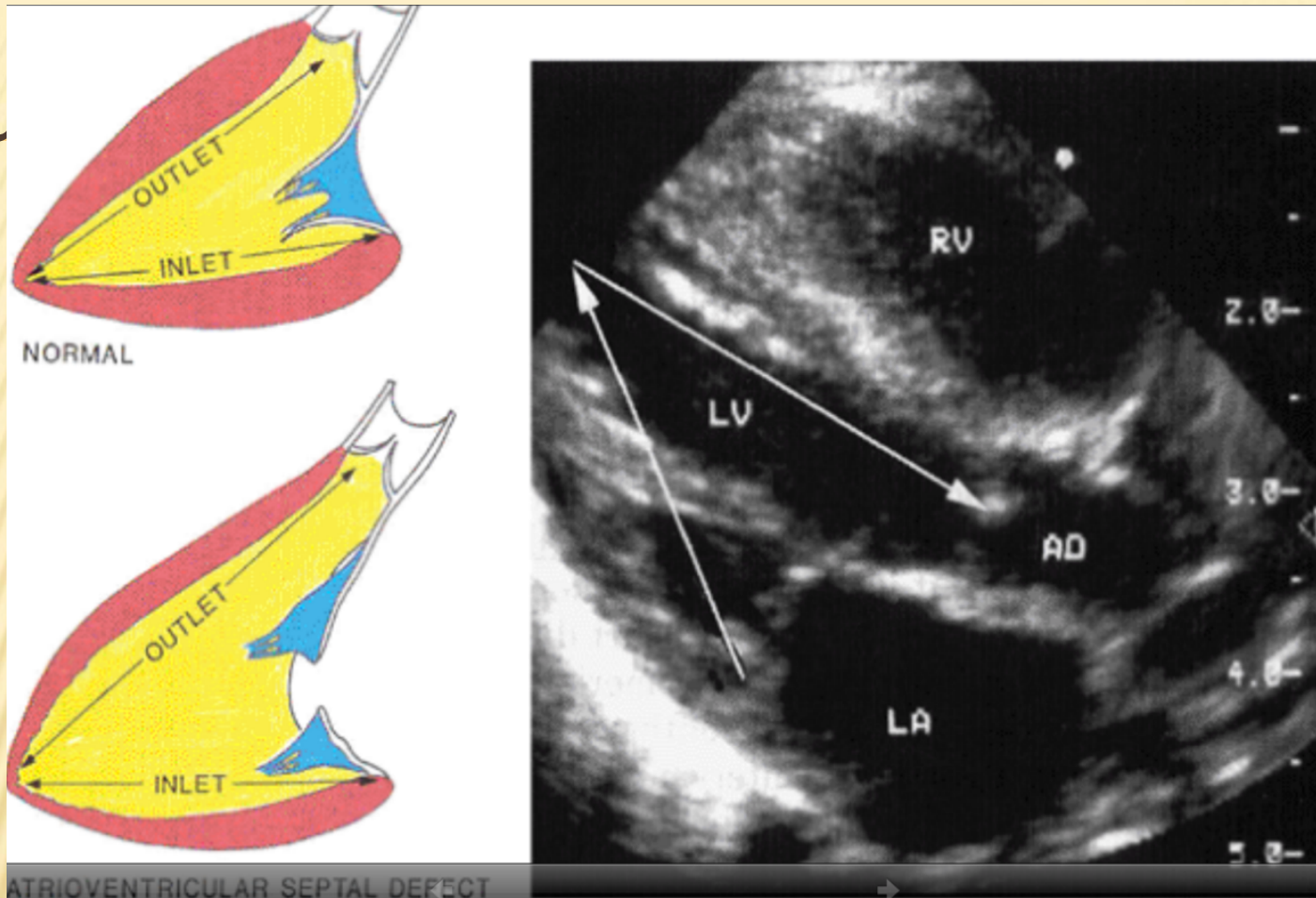
ECHOCARDIOGRAPHY

- Identified and characterize valvular abnormalities
- ASD and VSD morphology
- Associated anomalies.
- Degree of interatrial and interventricular shunting



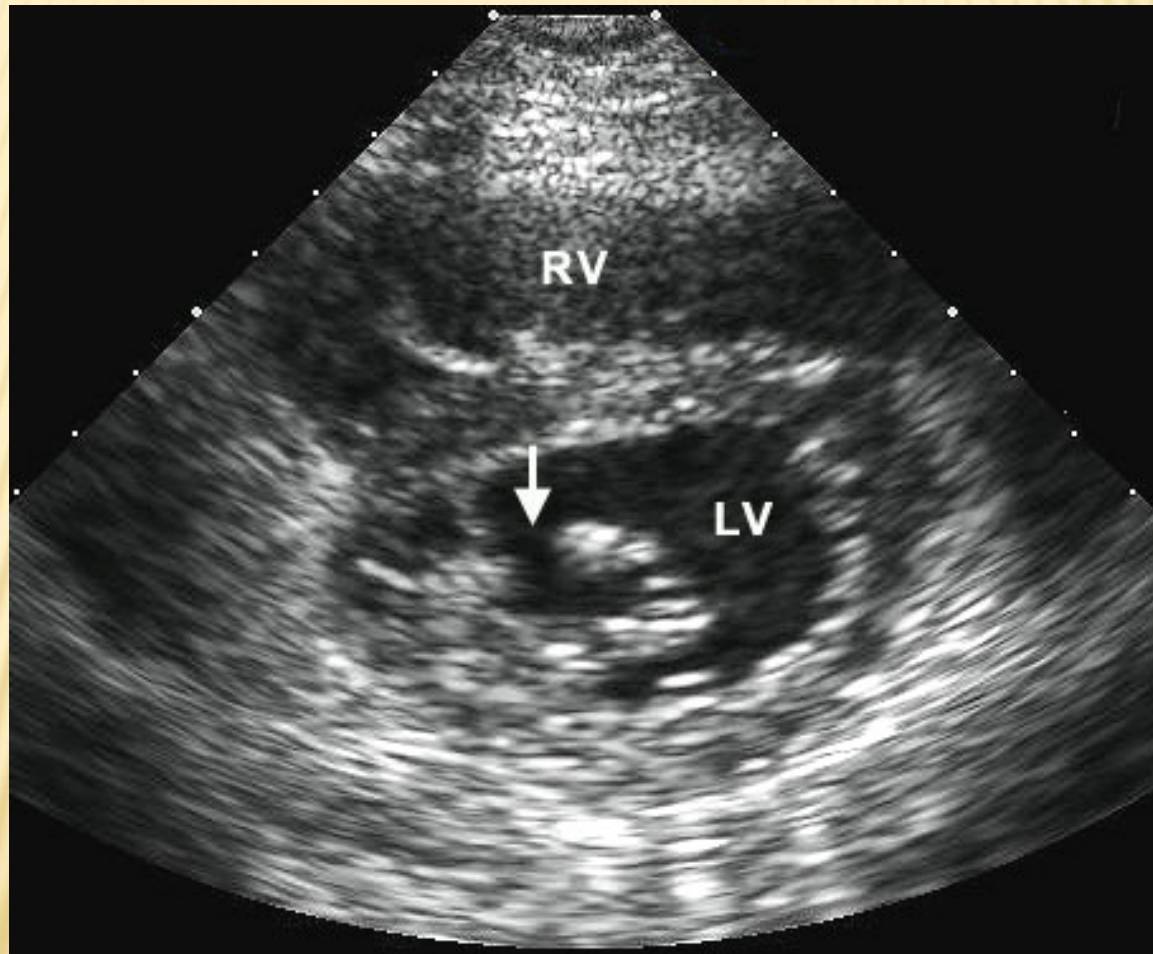
- In AVSD, the cleft in the anterior leaflet of the left atrioventricular valve is typically ***oriented toward the midportion of the ventricular septum*** (arrow) along the anterior-inferior rim of the septal defect.

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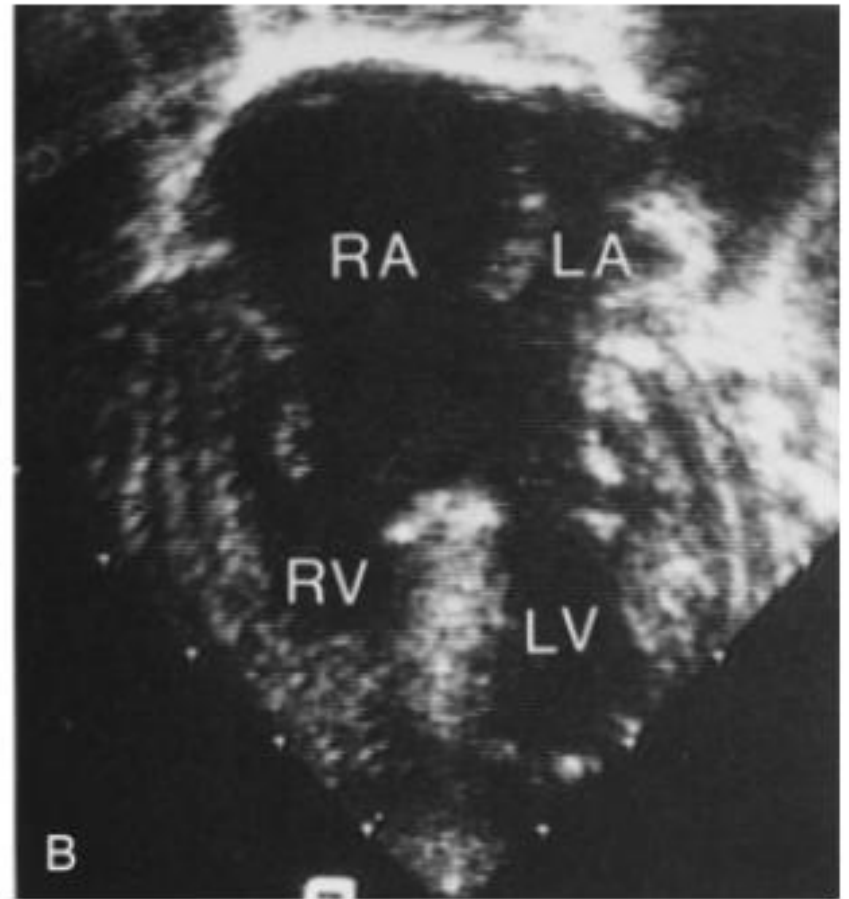
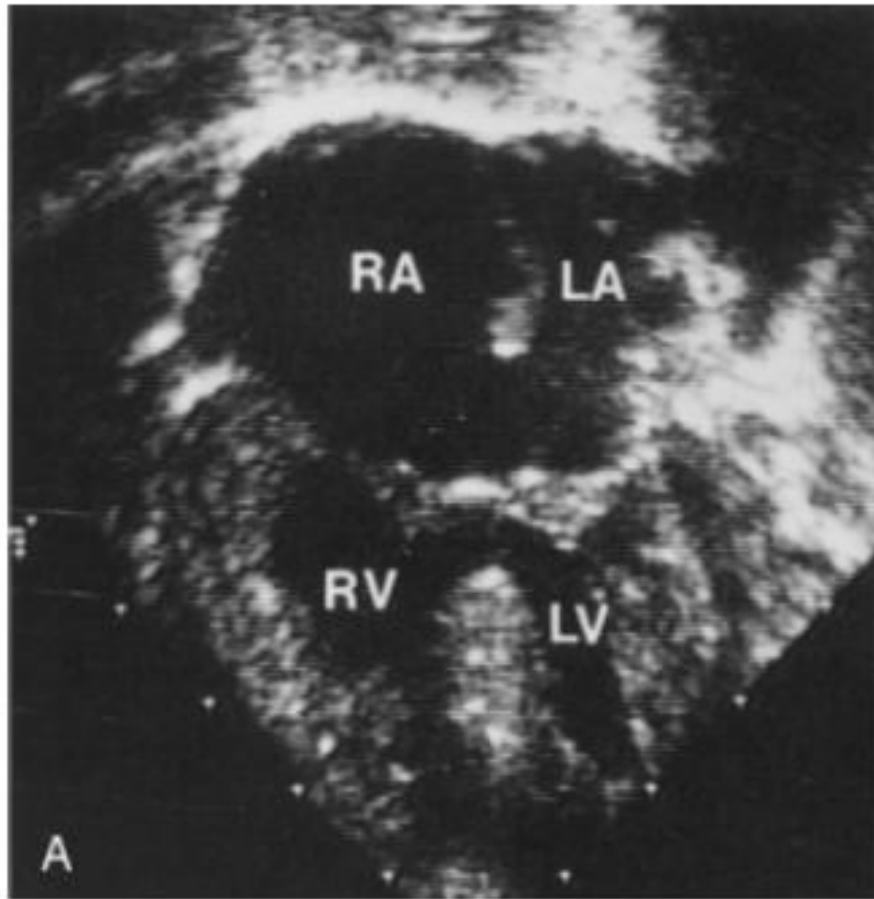


In the normal heart, the aortic valve is wedged between the mitral and tricuspid annuli. In AVSD the aortic valve is displaced or "oesprung" anteriorly (Fig. 31.11). This anterior displacement creates an elongate, so-called gooseneck deformity of the LVOT. (Fig. 31.12

CLEFT OF MITRAL VALVE



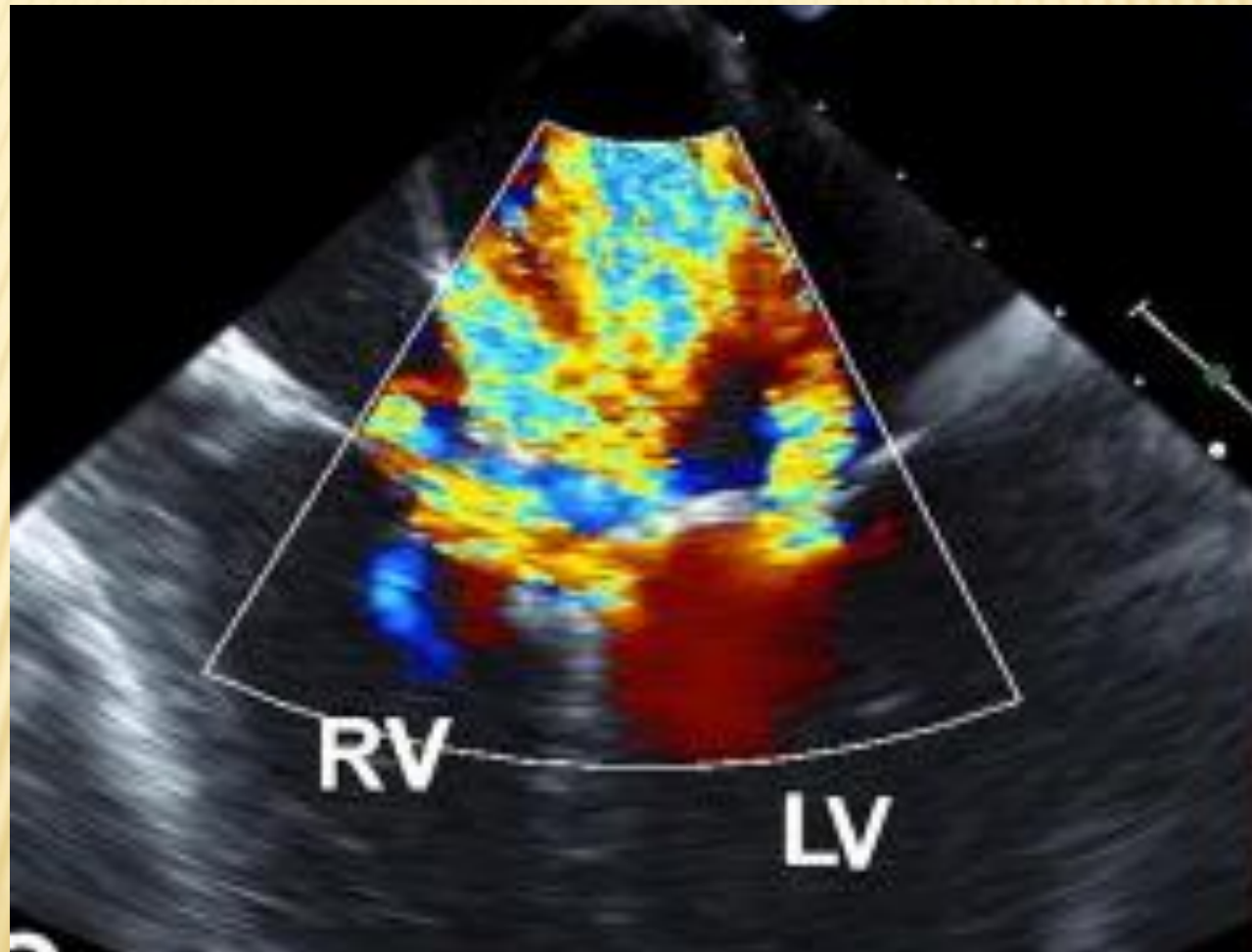
APICAL 4 CHAMBERS PLANE



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CARDIAC CATHETERIZATION

- rarely are required for diagnosis or management of patients with a partial AVSD.
- Frequently used to evaluate patient in complete AVSD
- Measure the pulmonary vascular resistance and evaluate the associated defects (TOF, DORV, TGV)

MEDICAL TREATMENT

- Treatment HF: Digoxin, Diuretics, ACEI...
- Manage the infections : antibiotics, nutrition.
- Prophylaxis bacterial endocarditis.

TIMING TO OPERATION

- A partial AVSD: 2-4 years of age
- A complete AVSD: 3-6 months of age.
- The intermediate AVSD depend on the size of the VSD – larger is earlier.

SURGICAL TREATMENT

- **Papillary procedure:** PA banding is no longer recommended unless other associated abnormalities make complete repair a high-risk procedure
- The mortality rate for PA banding may be as high as 15%.

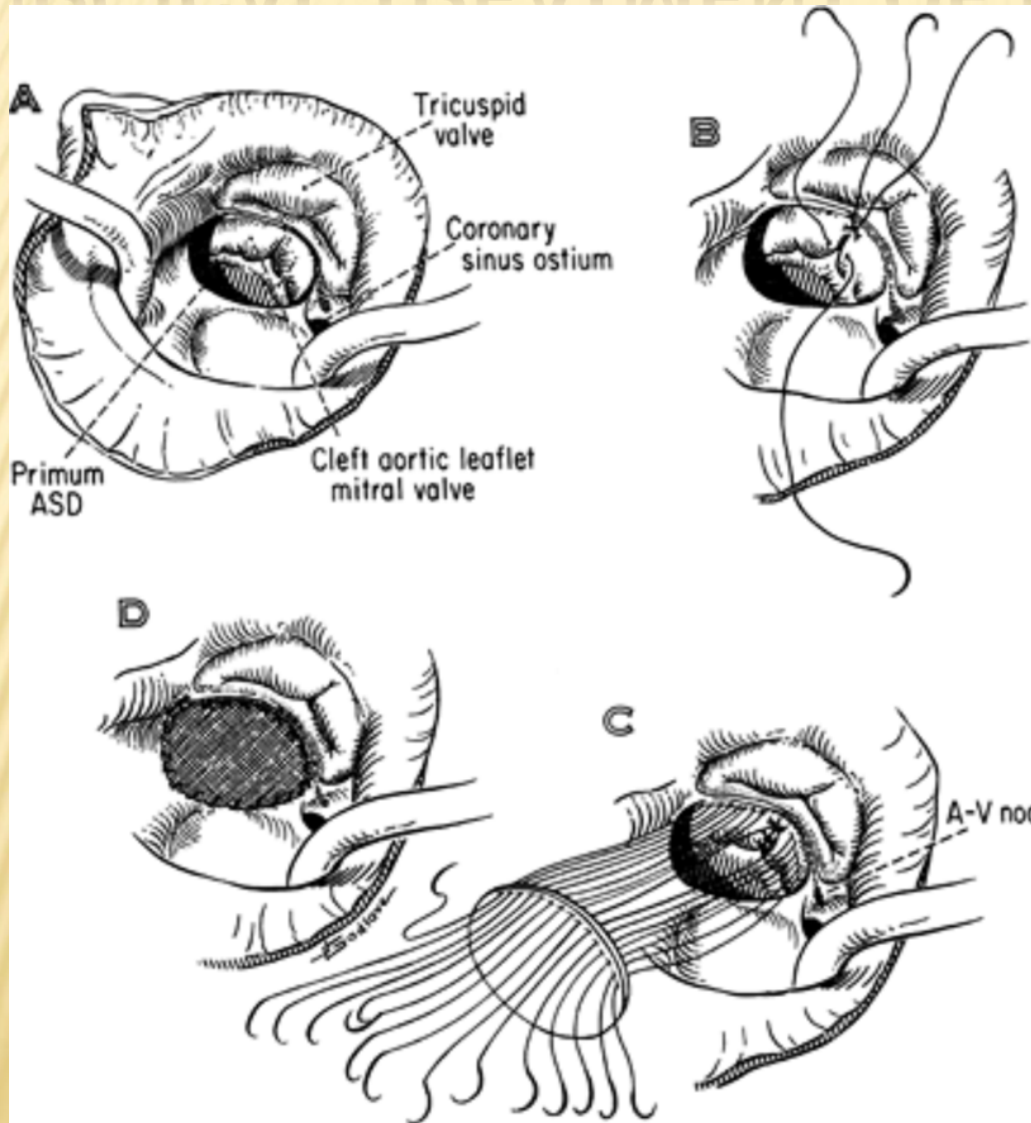
SURGICAL TREATMENT

Complete repair

□ Principles:

- closure of the ASD - VSD
- Competent AV valves
- Avoidance of damage to the AV node and bundle of His.

SURGICAL TREATMENT OF PARTIAL AVSD



- A. Surgical exposure
- B. Closure of the mitral valve cleft
- C. Prosthetic patch closure of an defect
- D. Repair completed

SURGICAL TREATMENT OF COMPLETE AVSD

- Repair of complete AVSD – using: single patch, two patch and modified single patch techniques.
- Patch : pericardium, polytetrafluoroethylene (PTFE), Dacron.

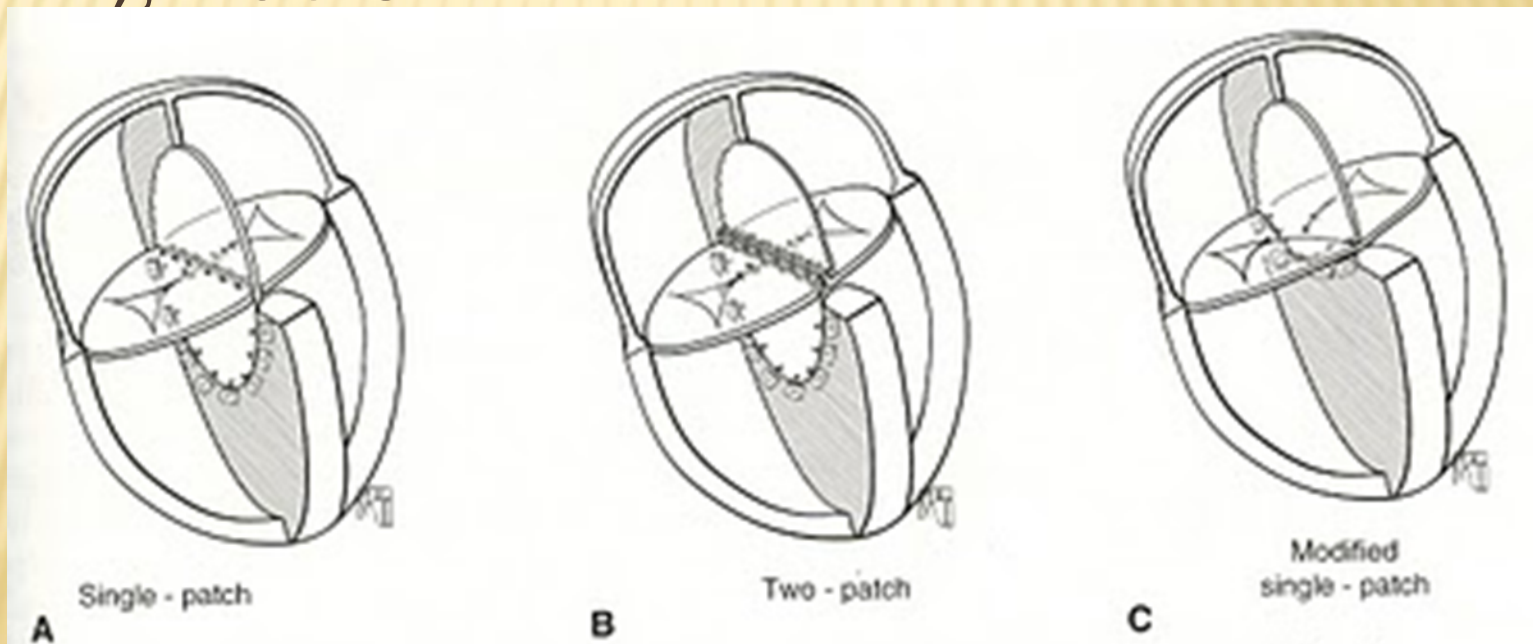
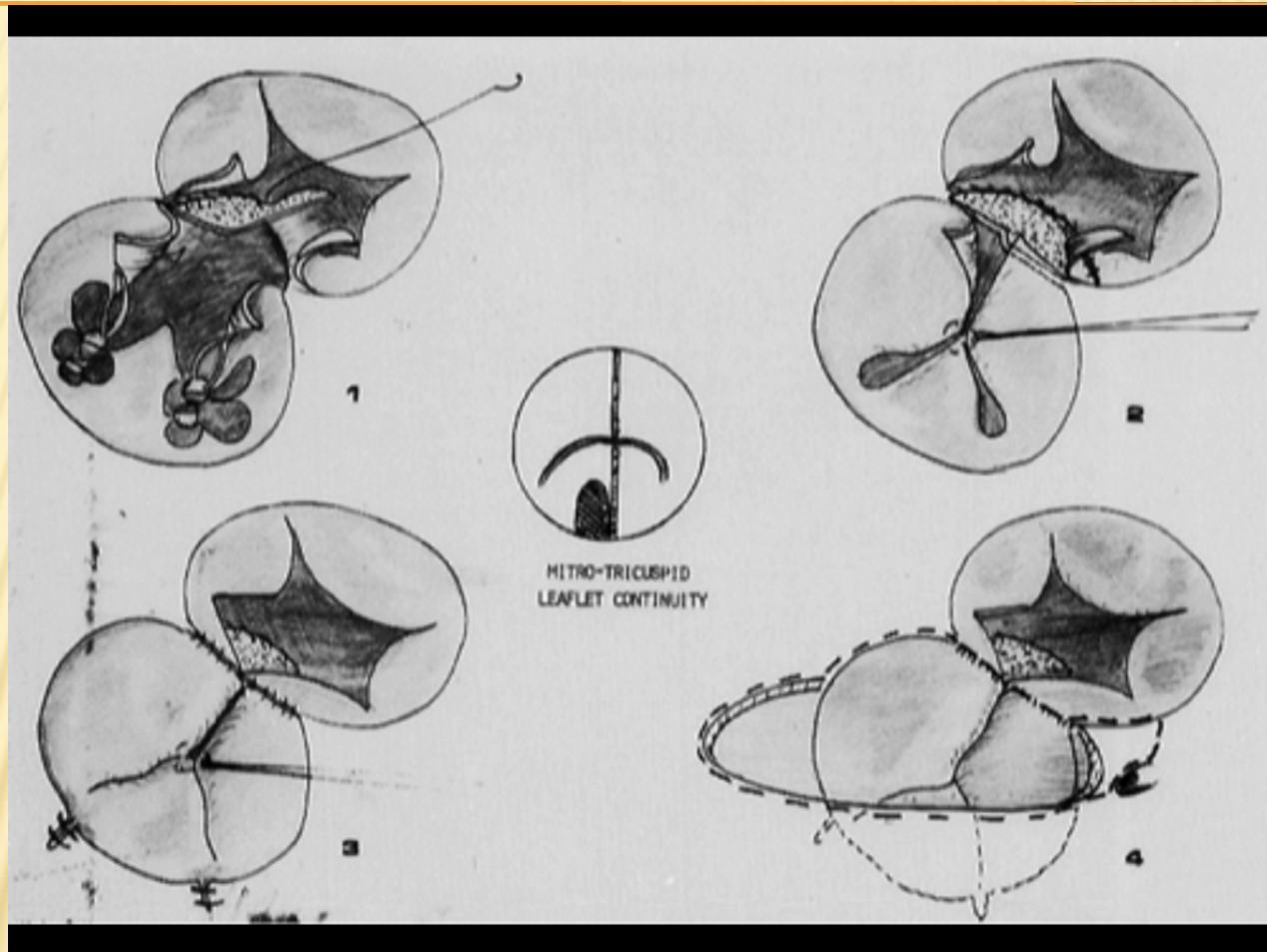


Fig. 18-5 Schematic three-dimensional reconstruction of the three different surgical techniques: single-patch (A), two-patch, (B), and modified single-patch (C).



Carpentier technique for repair of complete AVSD with the double patch technique. Concept of a trileaflet left atrioventricular valve - F 31.5 Moss Adam

RISK FACTORS

- Date of operation
- Age of operation
- Preoperative AV valve incompetence.
- Down syndrome

REOPERATION AFTER REPAIR

- regurgitation or stenosis of the left atrioventricular valve, subaortic stenosis, or residual -recurrent ASD.
- Mayo clinic 1995: 25% reoperation because of regurgitation and LOVT obstruction.

TAKE HOME MESSAGE

- AVSD represent a spectrum of intracardiac anomalies resulting from varied development of endocardial cushion structures.
- Range: partial AVSD(o.p ASD, cleft MV) – intermediate (transitional) AVSD – complete AVSD (ASD, VSD and common AV valve.)
- Partial AVSD- are treated like large ASD – can be operated on electively (2-4 years)
- Complete AVSD – should repair before 6 months of age.

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- Techniques : 1, 2 or modified- single patch.
- Post-operative management: follow up
 - Prevention of pulmonary hypertensive crises.
 - Residual of Recurrent left AV valve insufficiency
 - Permanent pacing
 - Left ventricular outflow tract obstruction

REFERENCES

- ❑ Moss and Adams' Heart Disease in Infants, Children and Adolescents, 7th edition 2008
- ❑ Perloff's Clinical Recognition of Congenital Heart Disease 6th 2012
- ❑ Adult Congenital Heart Disease 2009
- ❑ Pediatric Cardiac Surgery 2003
- ❑ Pediatric Cardiology by Park - 2008

THANK YOU FOR YOUR ATTENTION!

