Echocardiography Conference

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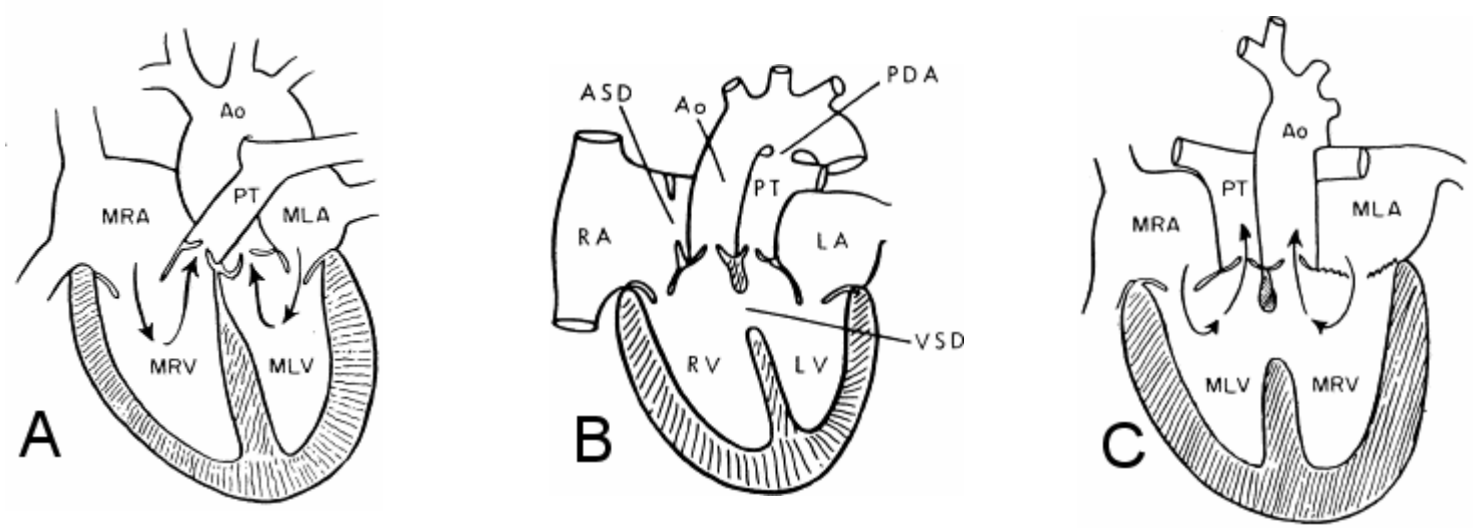
Goals for conference

- Better understanding of transposition of great vessels
  - D/L, ‘congenitally corrected’
- Review of ASD types and board pearls

Case 1

- 40 yo WM with hx Congenitally corrected Transposition of the Great Vessels presenting with increased DOE
- 1994 L Subclavian to L PA shunt
- Pulmonary Stenosis
  - Porcine valve replacement
- VSD
  - Patch repair
- Hx Left AV Regurgitation
  - Repaired 2000

Transposition of the Great Vessels



A – Normal Cardiac development
B – Complete transposition of the Great Vessels
Note VSD and PDA
C – Congenitally corrected transposition

Hornung et al.

Vocabulary

- Atrioventricular concordance vs. discordance
- Ventriculoarterial concordance vs. discordance
  - Tansposition features Ventriculoarterial discordance
- D-loop – Complete transposition; Aorta arises right of pulmonary artery
- L-loop – Congenitally corrected; Aorta arises left of pulmonary artery

The “Broderick” classification

- Atria are in their usual position
- The tricuspid valve attaches to the morphologic RV
- The mitral valve attaches to the morphologic LV
- The aortic valve is connected to the aorta
- The pulmonary valve connects to the pulmonary artery

Complete Transposition

- Most common
  - cyanotic congenital heart disease
  - presenting in infancy
- 5-7% of all congenital heart disease
- 20-30/100,000 live births
- 2:1 male:female

Hornung et al.

Complete Transposition D-Loop

- Aorta arises from RV
  - Coarses right and anterior to Pulmonary Artery
- Pulmonary artery arises from LV
- Coronaries typically from aorta but various origins

Hornung et al.

Associated lesions

- 50% have another heart defect
- 40-45% with VSD
- 25% with pulmonary outflow tract obstruction
- 5% with aortic coarctation

Hornung et al.

Natural History
No intervention

- 90% die in infancy
- Those WITHOUT other defects do worse
  - Only 30% survive beyond 1 month
- Those with LVOT (Pulm Artery) obstruction and VSD tend to do better

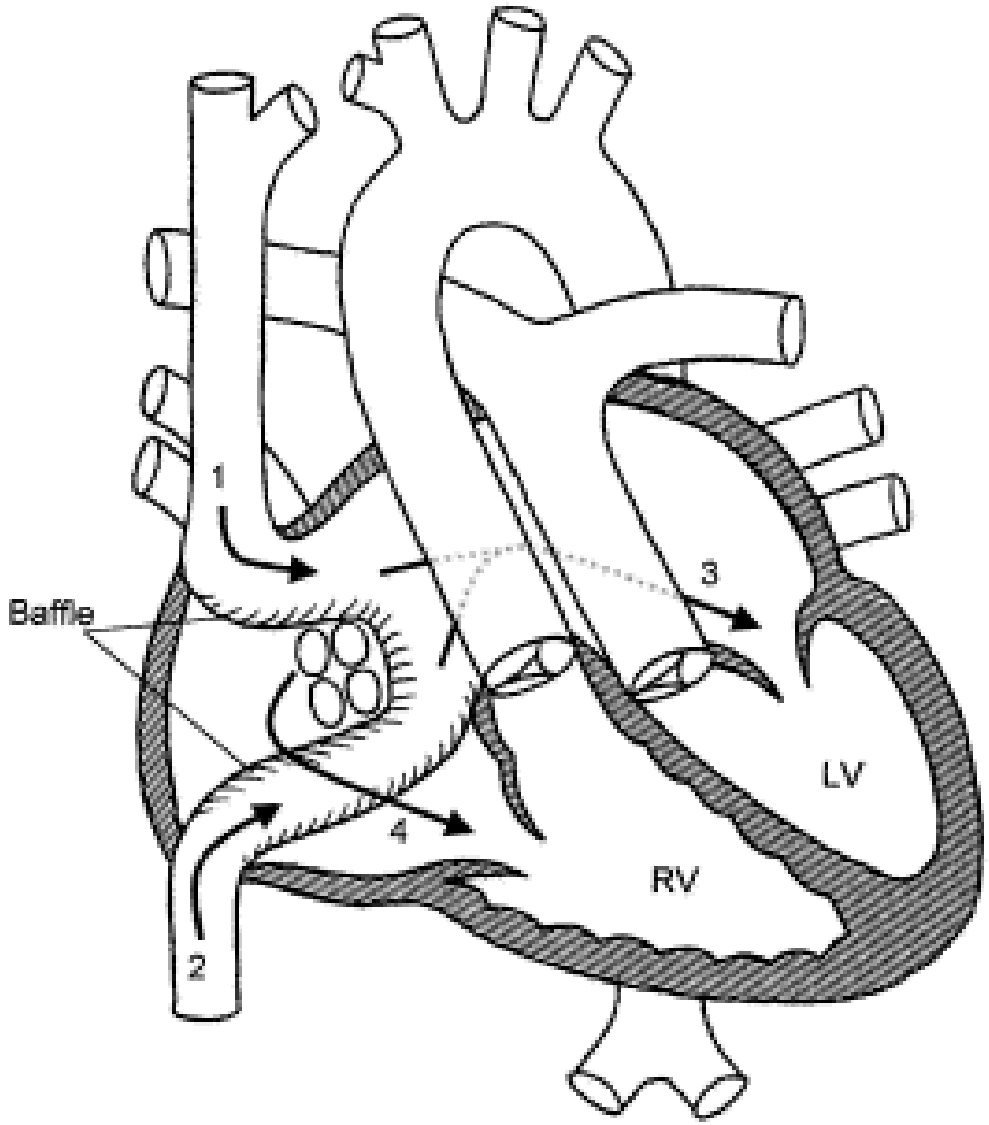
Hornung et al.

Surgical Therapy

- Mustard and Senning procedures started in 1960's
- Result in normal Pulmonary->systemic blood flow
- RV and TV still function systemically
- Operative mortality 5% in 1970's
  - Worse for patients WITH other defects

Hornung et al.

Mustard operation



- Atrial baffle diverts blood from SVC and IVC toward the MV and LV
- Pulmonary blood returned to TV and RV

Hornung et al.

Following surgery

- 85-90% 10 year survival
- 80% 20 year survival
- Late causes of death
  - Presumed arrythmia
  - RV failure
- Good functional capacity (60-80% NYHA class I)
- Many patients have chronotropic incompentence

Hornung et al.

Right Ventricle

- Morphologic RV destined to provide systemic circulation
- RVEF of 50% considered ‘normal’
- Concern for degeneration of function in 3rd-4th decade
- Geometry yields echo insufficient to evaluate function
- MRI considered gold standard*
- RV typically hypertrophies

Hornung et al.

Tricuspid Valve

- Mild TR (1-2+/4) common
- Significant TR may be due to RV dilitation
- In patients with VSD
  - Tricuspid may be damaged during repair of VSD; may necessitate further repair

Coronary anatomy

- Various origins
- Left cor supplies ANATOMIC LV
- Right cor supplies ANATOMIC RV

Arrythmia

- Loss of sinus rhythm
- Atrial tachycardia
- May require permanent pacemaker
- Atrial flutter is marker of increased risk of sudden death

Late surgical therapies

- Baffle leaks
  - May close with percutaneous occluder
- Pulmonary or systemic venous stenosis
  - Amenable to balloon dilitation
- Late arterial switch procedure
  - Attempted for RV dysfunction, refractory TR
  - Not very successful, requires ‘retraining’ LV

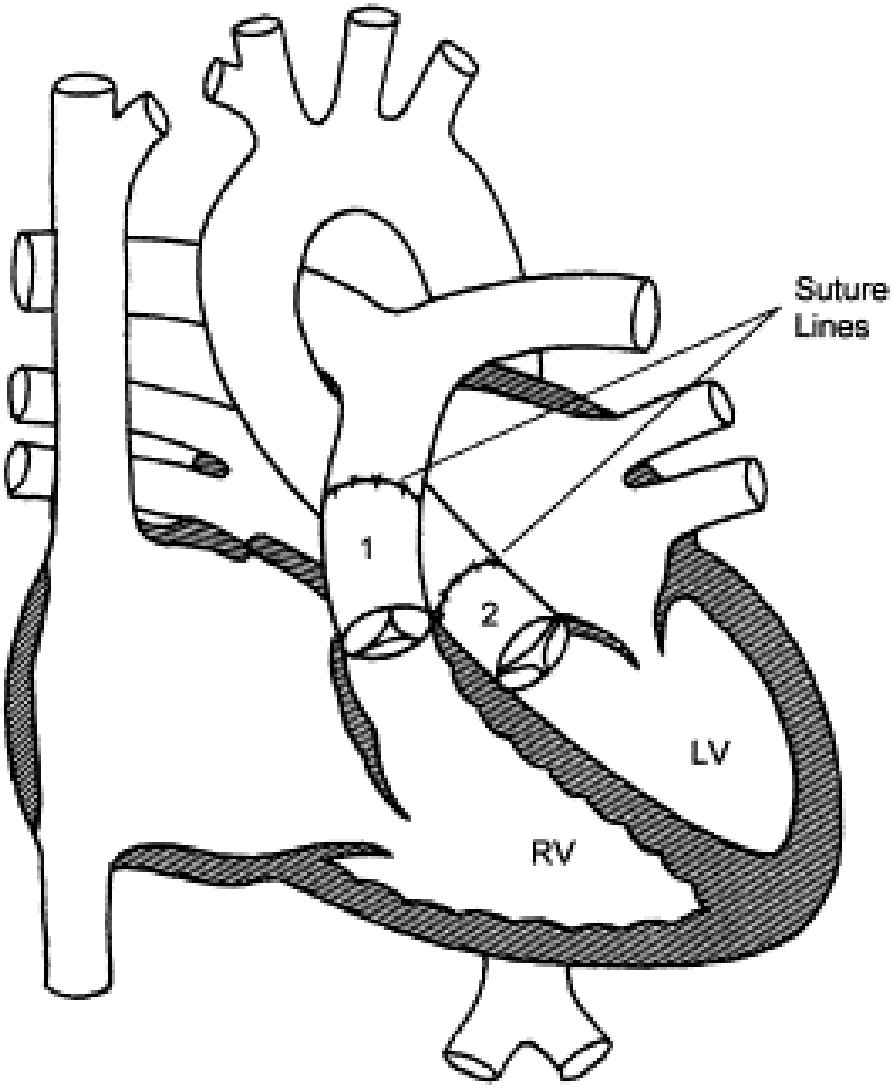
Medical therapies

- ACE inhibitor
  - No data, ongoing clinical trial
- Beta blockers
  - Presumed effect from ‘normal’ CHF patients
  - No data

Arterial Switch Procedure

- Initially high mortality, but theoretically better
- Should be performed age 3-4 weeks
- Operative mortality 5%
  - Increased risk with other defects, prior surgery
- 5 year survival good
- Patients in 20's now

Arterial Switch Procedure



- Great arteries transected above the valves and switched
- Cors are removed from aorta with cuff of wall and re-attached to the aorta after the switch

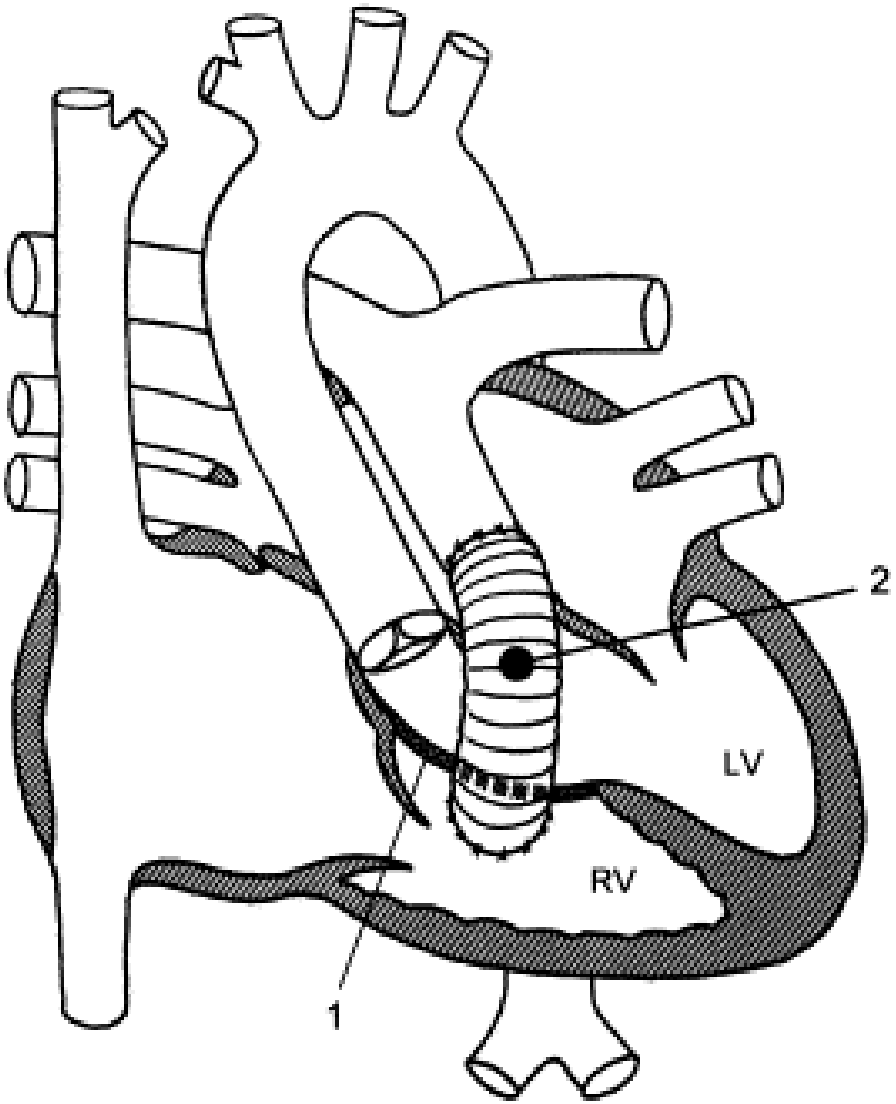
Late complications

- RVOT obstruction
  - LeCompte maneuver to flatten main PA
  - 18% reintervention in 10 years
- Aortic regurgitation
  - 9% incidence at 15 years
- Coronary occlusion/obstruction
  - 18% incidence

Transposition with VSD and Pulmonary Stenosis

- Rastelli procedure
- Operative mortality 5%
- 80% survival at 10 years
- 50% survival at 20 years
- High incidence of late arrythmia

Rastelli procedure



- VSD closed to become part of LVOT
- RV-PA conduit bypasses stenosis
- Conduit stenosis is inevitable!

Congenitally corrected transposition
L-Loop

- Rare, atrioventricular discordance and ventriculoarterial discordance
- 0.5% of congenital heart disease
- Blood flow pattern is normal
- RV supports systemic circulation
- 80-90% incidence of other defect
  - 60-80% with VSD
  - 30-50% with Pulmonary outflow obstruction
  - 90% with Ebstein's anomaly (clinical picture variable)
  - Complete heart block 2%/year

Natural history of congenital corrected transposition

- Variable
  - If no other defect, may only be discovered at autopsy
- Mean age of death about 38% in 1 series
- Various associated lesions make stats difficult
- Subnormal exercise capacity

Right ventricle

- Progressive dysfunction
- 67% with CHF by age 45
- No data to support medical therapy, though ACE inhibitor used

Tricuspid valve

- Regurgitation common
- Repair is feasible but replacement may be more beneficial

Double switch operation

- Small number of patients have
  - Great vessel arterial switch
  - Atrial baffle to redirect blood flow
- Anatomic LV->Aorta
- Anatomic RV-> Pulm Artery

Echo features

Feature	RV	LV
AV valve	Trileafet	Bicuspid (unless cleft)
Annulus	More apical	More basal
Trabeculation	Prominent	Less prominent
Moderator band	Present	Absent
Infundibulum	Present	Absent

Otto, 398

Cardiac Catheterization

- CX – OM1, OM2
- LAD small
- RCA – large RV branches, PLV, PDA branches

Cardiac Catheterization Hemodynamics

- RA 10
- LV 118/6 EDP 18
- PA 139/79, mean103
- RV 92/2 EDP 2
- AO 91/52 mean 70
- Oxygen saturation on RA: PA 23%, Ao 80%
- Following Oxygen administration: PA 35%, Ao 88%
- No change in Right sided pressures with Oxygen
- CO 2.34, CI 1.15
- Severe RV dysfunction EF 25%, 4+ regurgitation

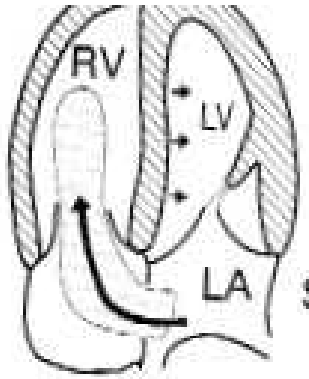
Followup of Pt

- Referred initially to Mayo, declined reintervention
- Accepted at Cleveland clinic (site of prior surgeries) for possible TV repair/replacement
- Ultimately received Heart-lung transplant

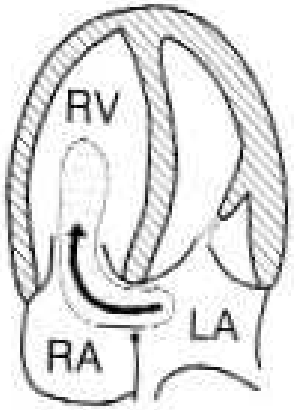
Patient #2

- 24 yo WF with ‘heart murmur’
- Mild DOE
- Previously negative TEE for ASD

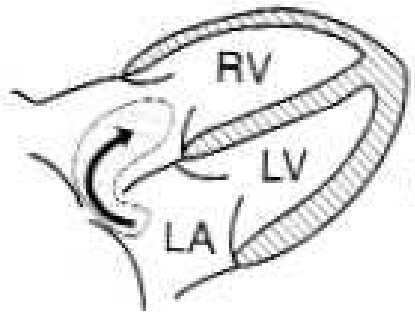
Types of ASD



Secundum ASD



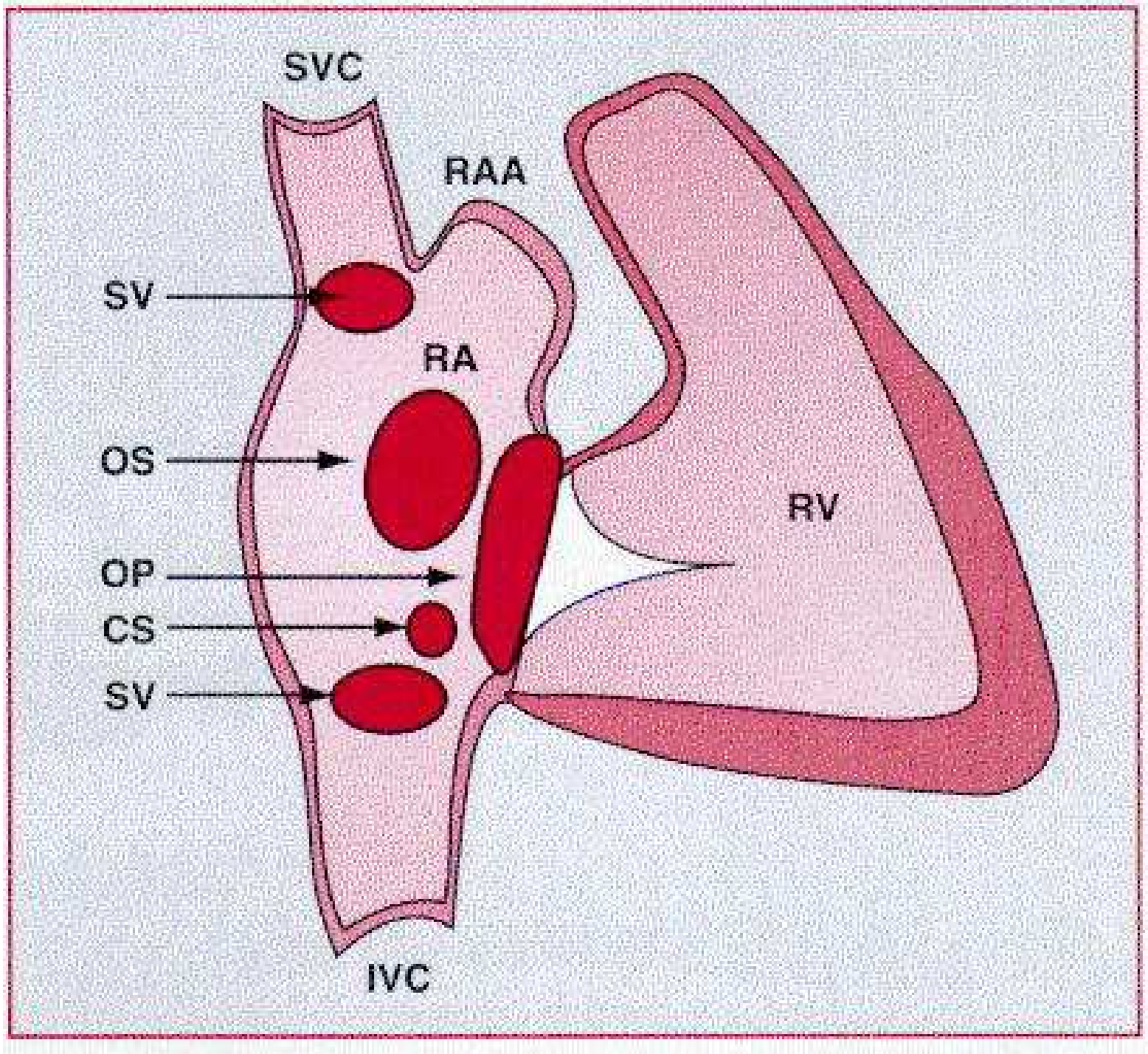
Primum ASD



Sinus venosus ASD

- Secundum – most common (7% of all congenital heart disease)
  - Failure of secundum atrial septum to cover foramen secundum
- Primum
  - Abnormal septum primum with failure of closure
  - Associated with **cleft mitral valve**
- **Sinus venosus**
  - Least common (2-3% of ASD)
  - Abnormal fusion of sinus venosus and atrium
  - Near junction of IVC/SVC and atrium

Otto, 408



Braunwald, 412

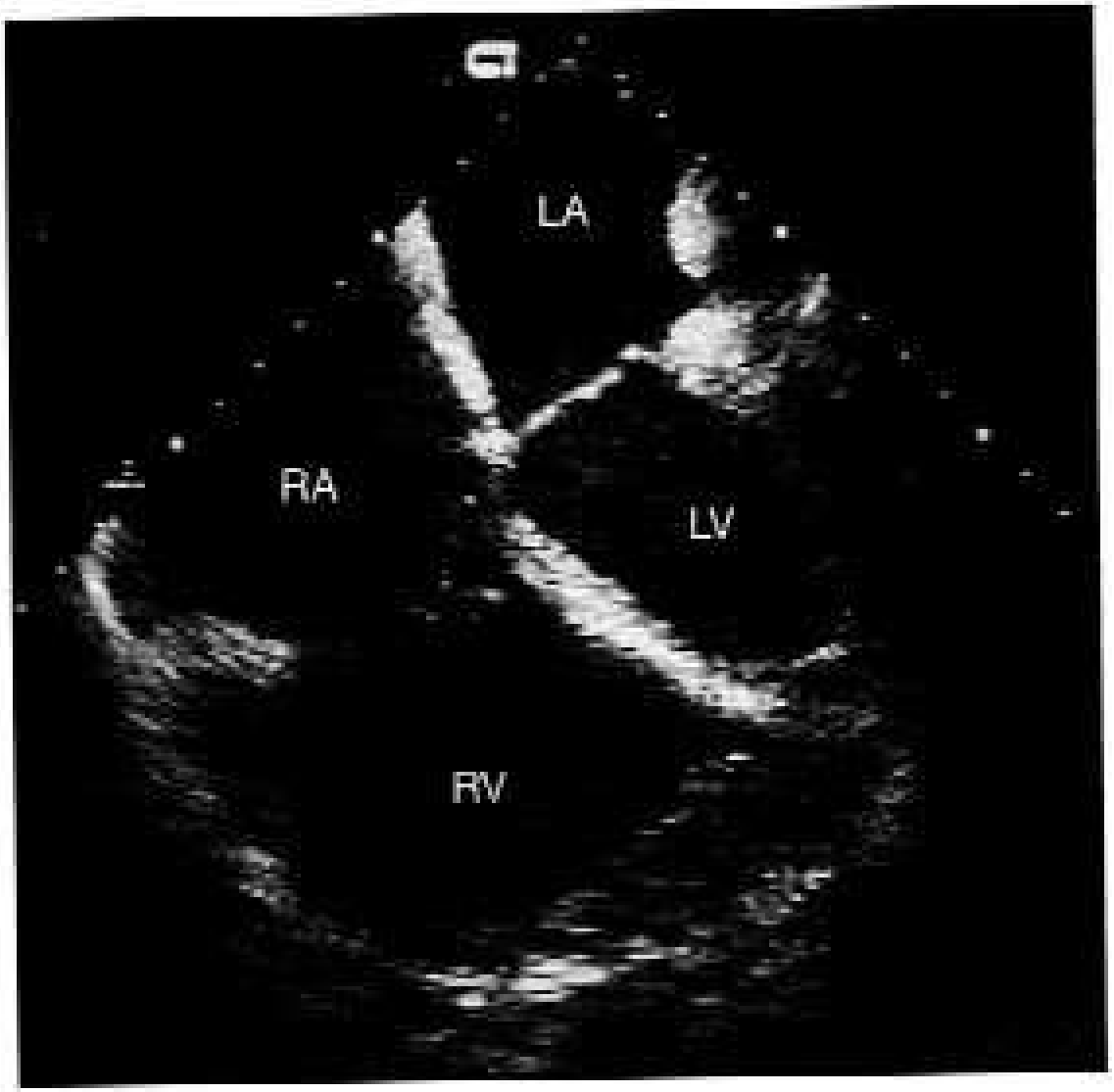


FIGURE 16–15. Sinus venosus atrial septal defect in a 74-year-old man. Although the transesophageal four-chamber view shows marked volume overload of the right side of the heart, the atrial septum appears intact in this view.

Otto, 412

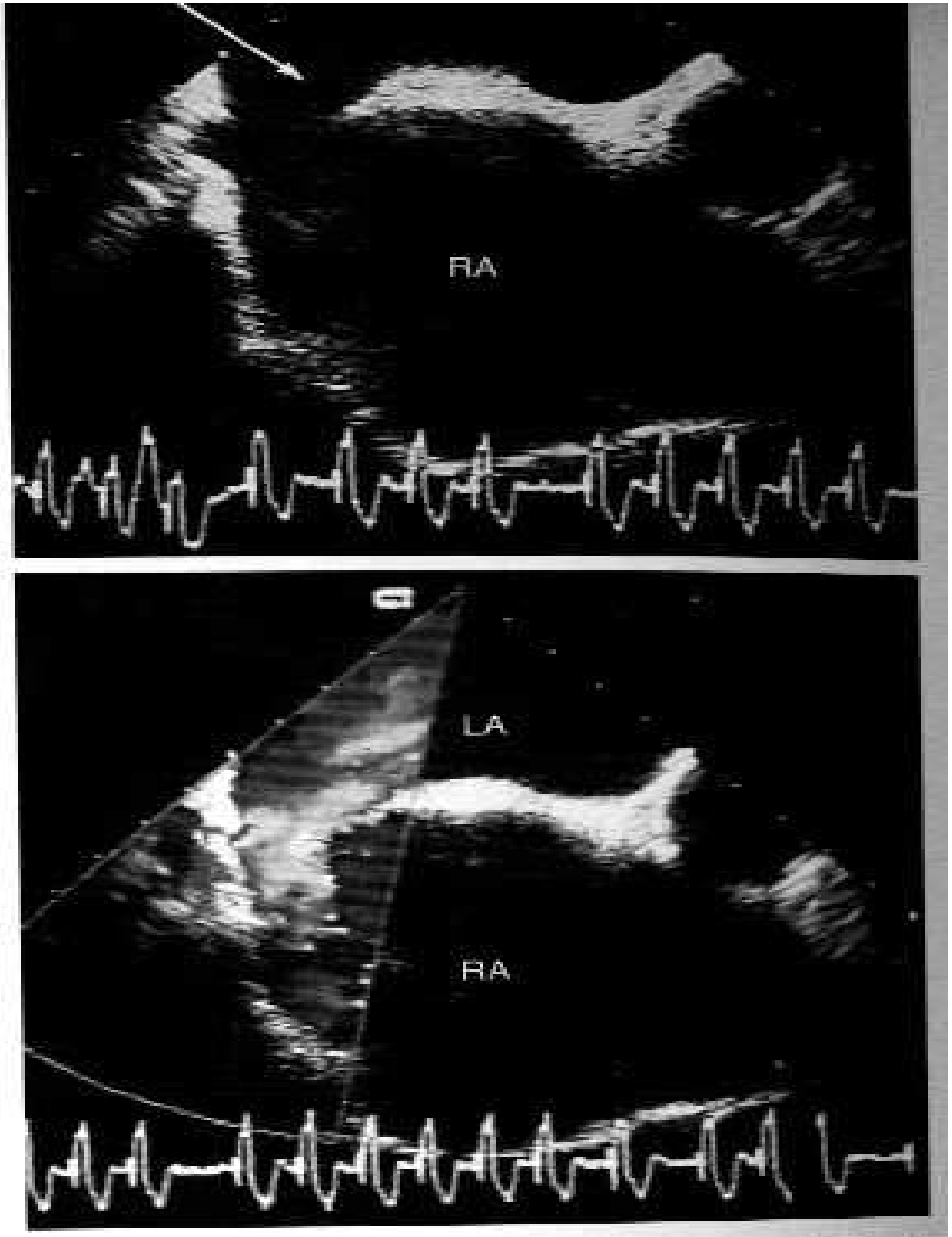


FIGURE 16–16. In the same patient as in Figure 16–15, the region of the atrial septum just inferior to the superior vena cava is absent *(above)*, and color flow demonstrates left-to-right flow across this defect *(below)*. (See Color Plate 19, p. 386.)

Otto, 412

Pearls for ASD

- Secundum
  - Most common type
  - Commonly presents in adulthood
- Primum
  - Associated with cleft MV
- Sinus venosus
  - Associated anamolous pulmonary vein return
  - Difficult to visualize
  - Catheter may ‘slip’ easily into pulmonary vein

References

- Hornung TS, Derrick GP, Deanfield JE, Redington AN. Transposition complexes in the adult: a changing perspective. Cardiol Clin. 2002 Aug;20(3):405-20.