Pulmonary venous anomalies

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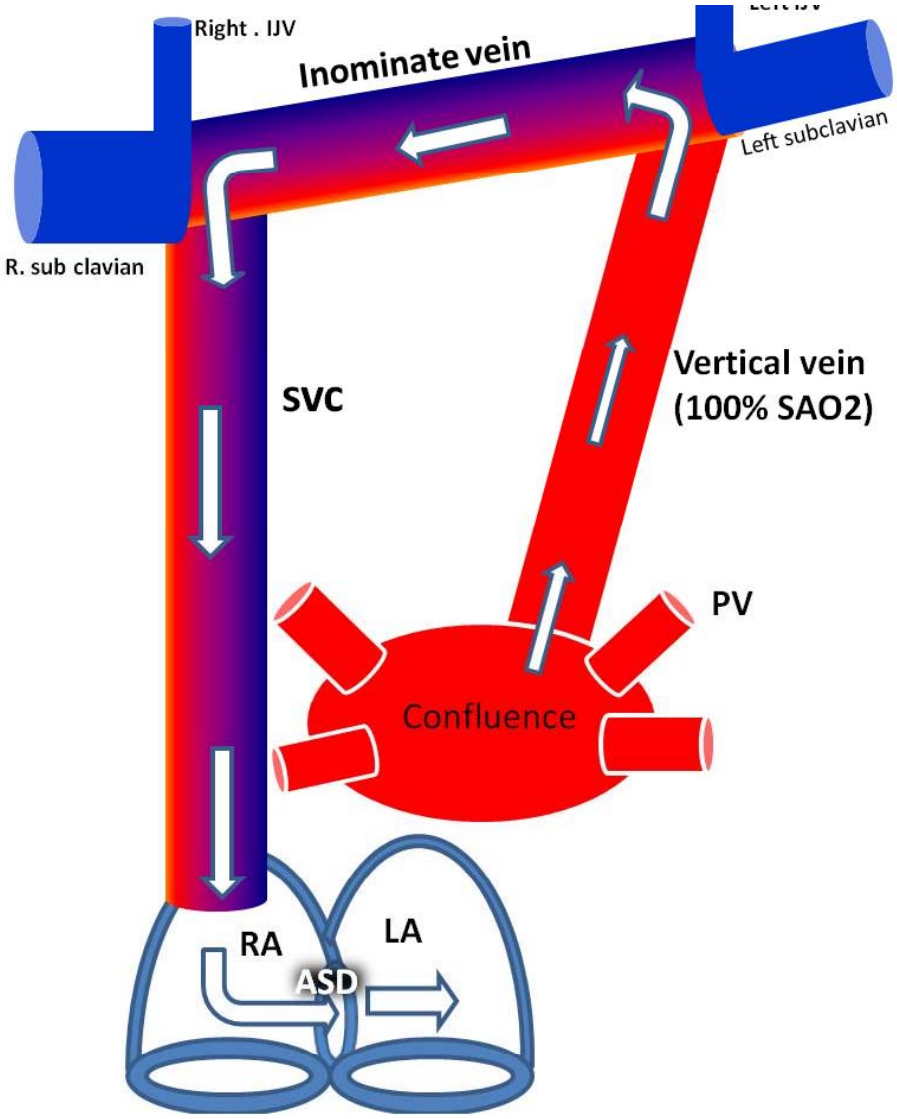
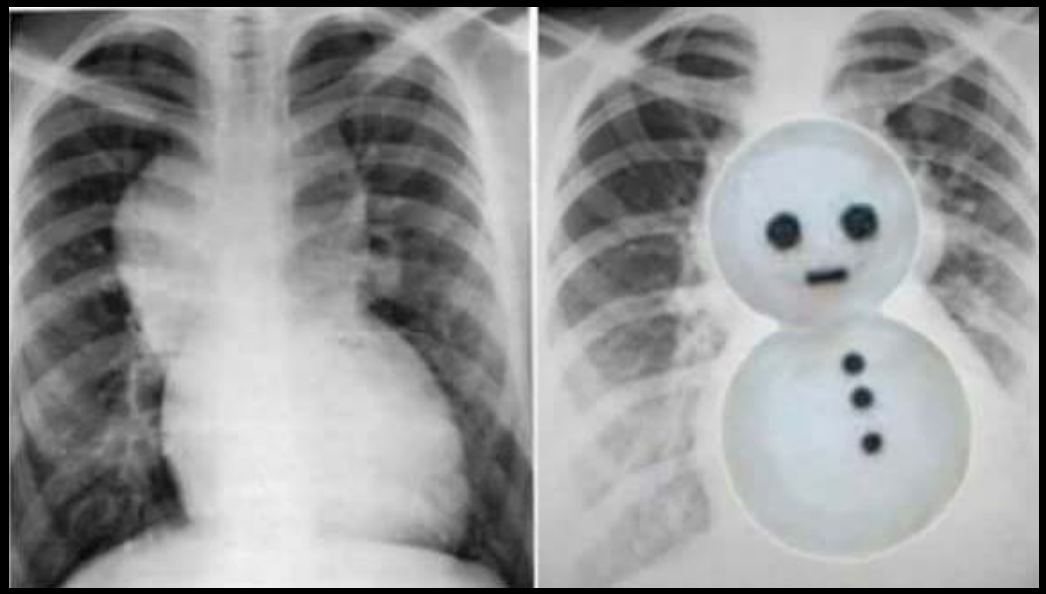
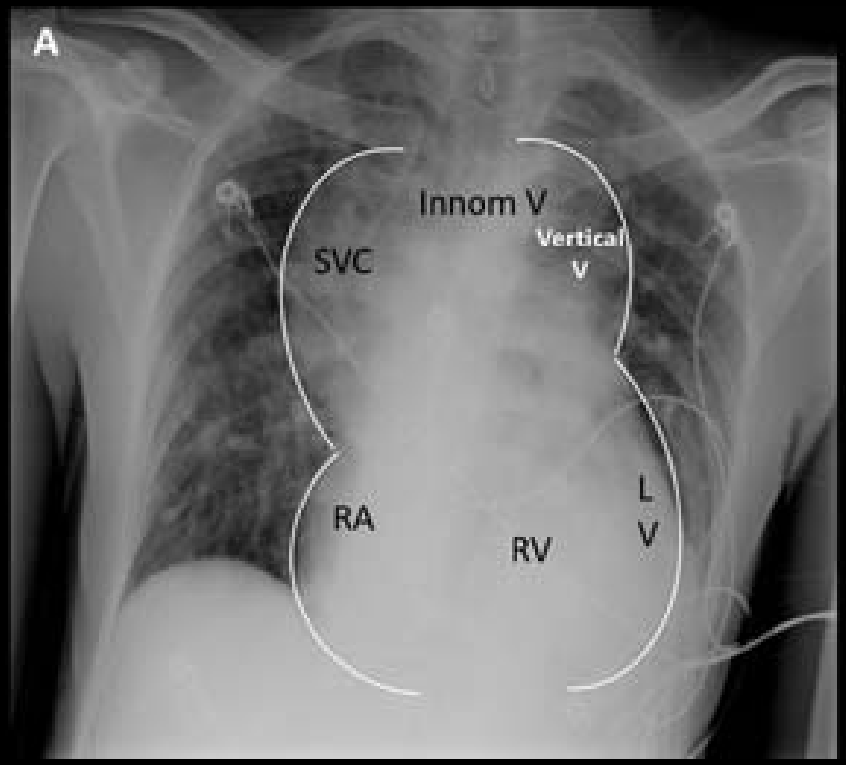
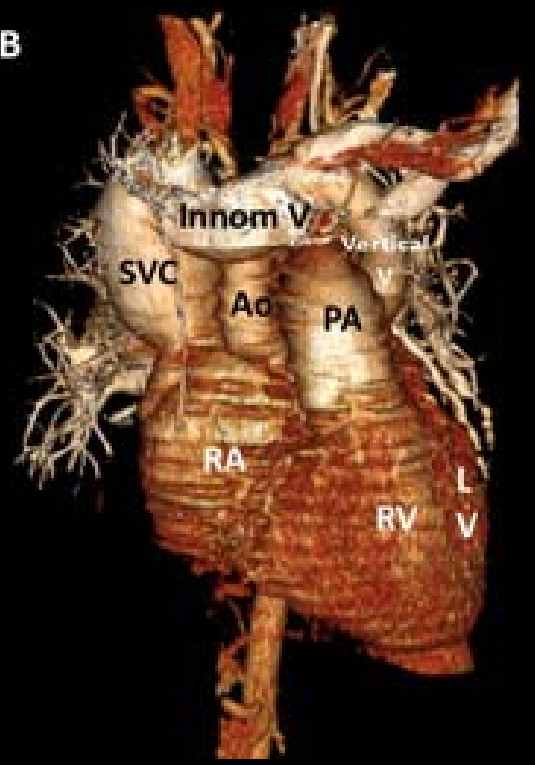
Total anomalous pulmonary venous returns (TAPVR)

- All pulmonary venous blood flow returns anomalously to the systemic veins or directly to the right atrium
- Prevalence estimated at 1 in 10,000
- **Acutely cyanotic infant in shock**
- One of the true surgical **emergencies** across the entire spectrum of congenital heart surgery.









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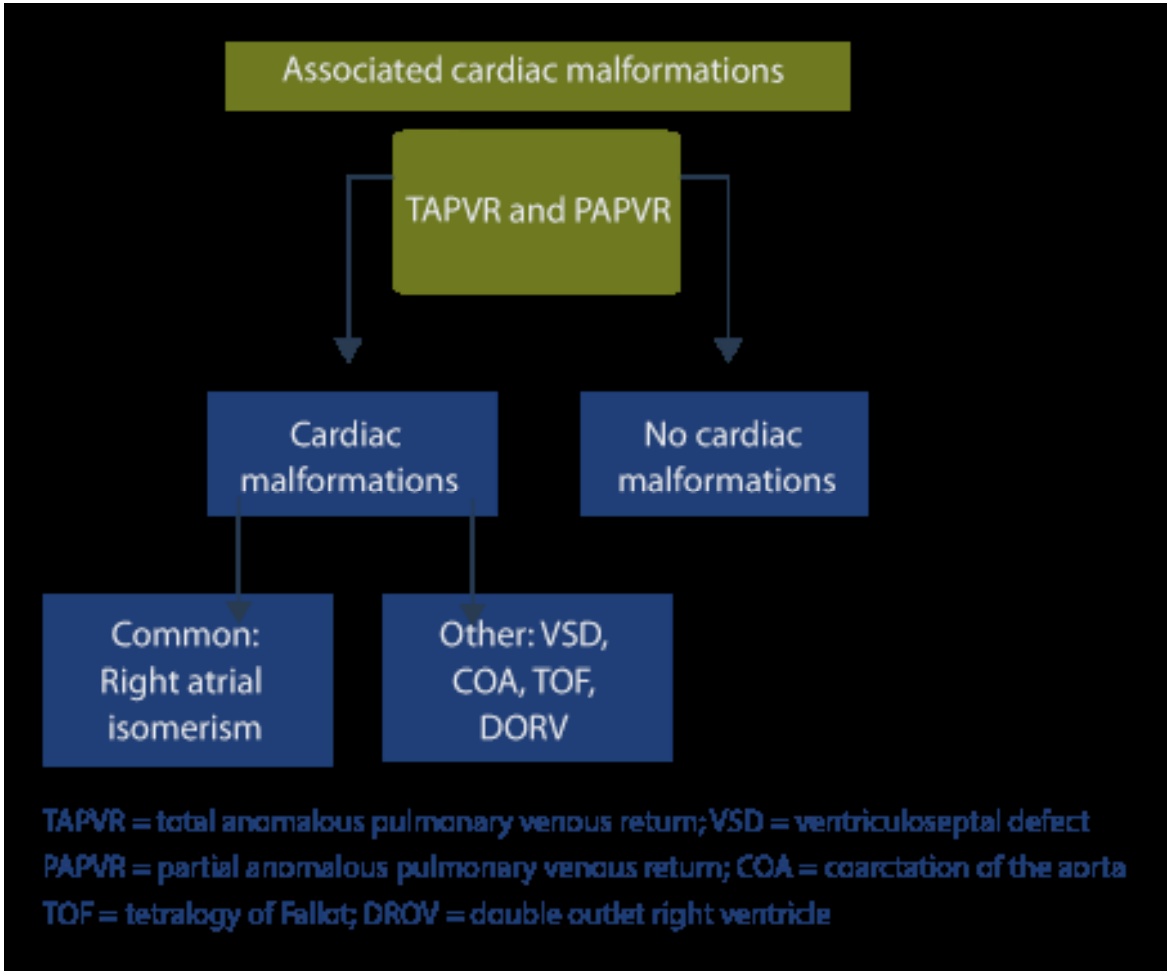
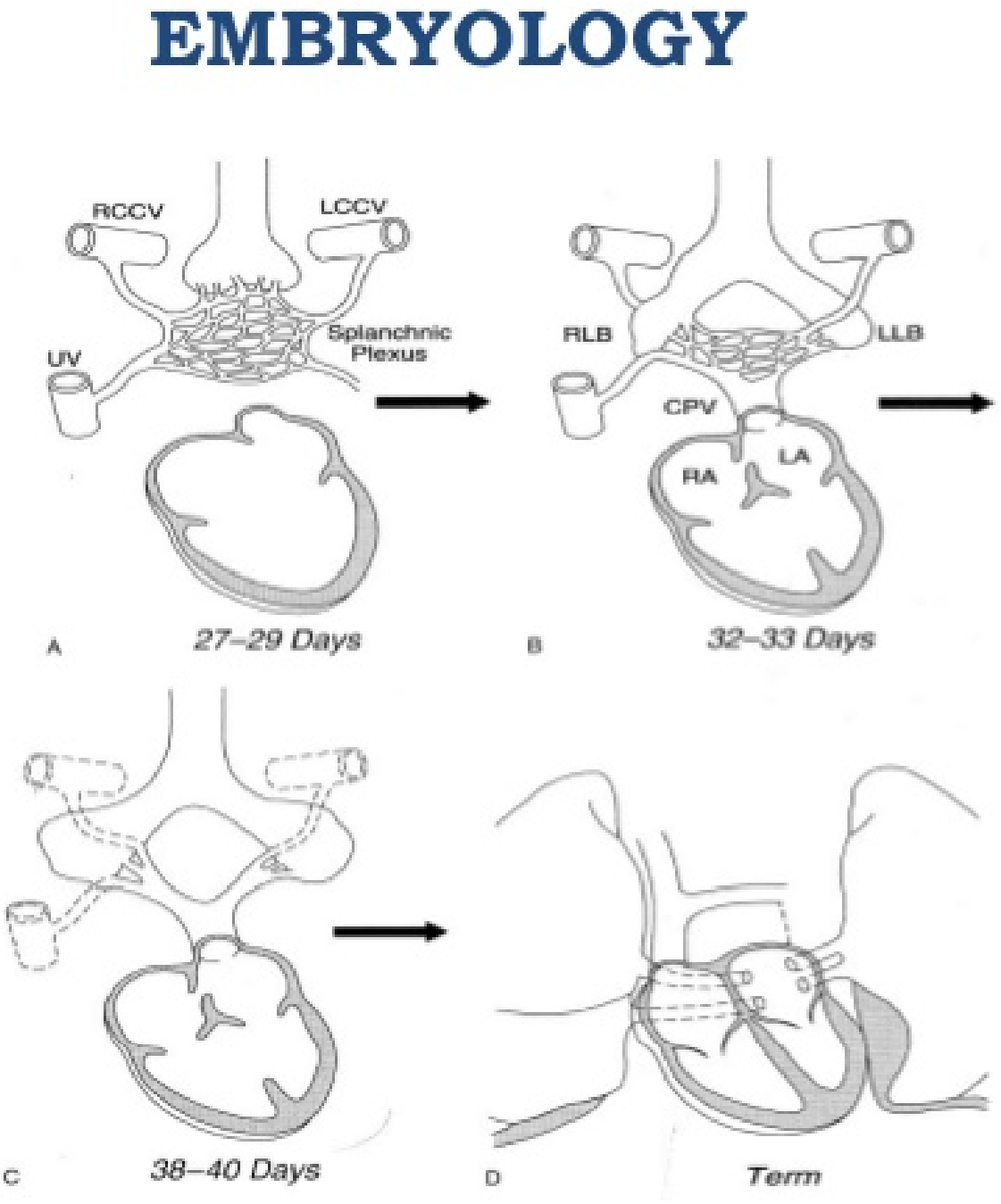
TAPVR

- Biventriclular heart

- Single ventricle
- Heterotaxy syndrome



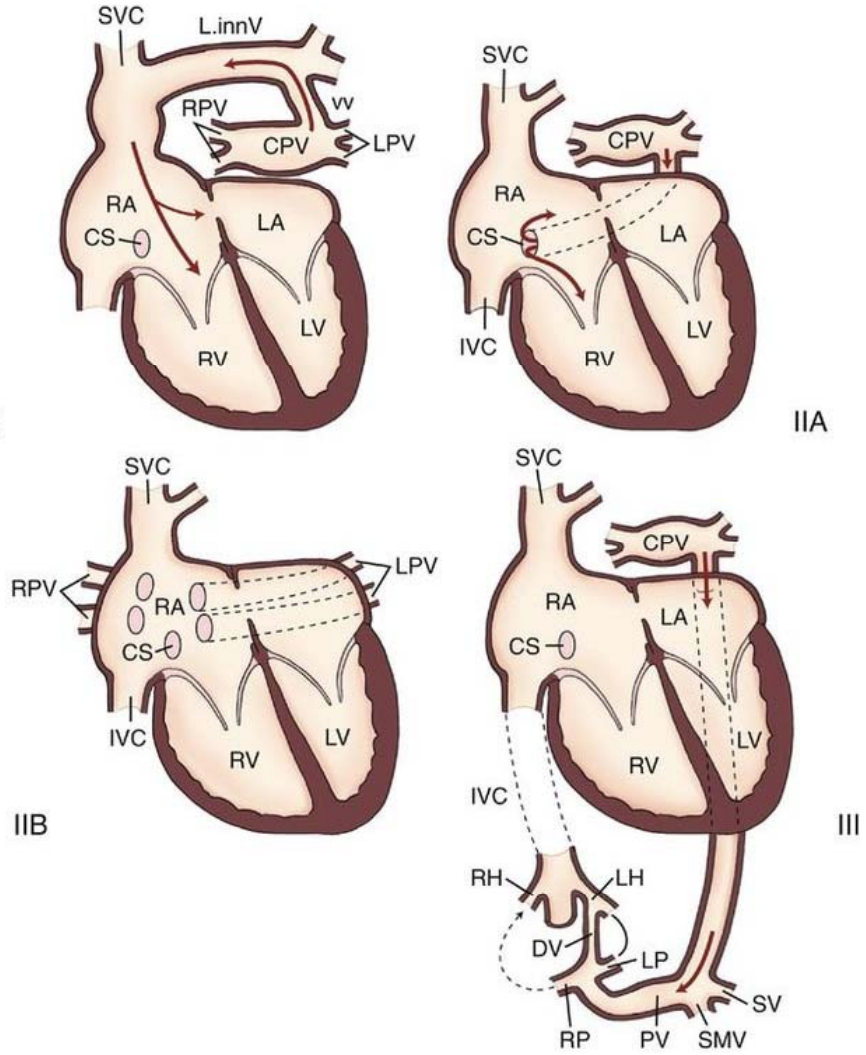
Embryology


EMBRYOLOGY
RCCV
LCCV
Splanchnic Plexus
UV
A
27–29 Days
RLB
LLB
CPV
RA
LA
B
32–33 Days
C
38–40 Days
D
Term

Anatomic subtype

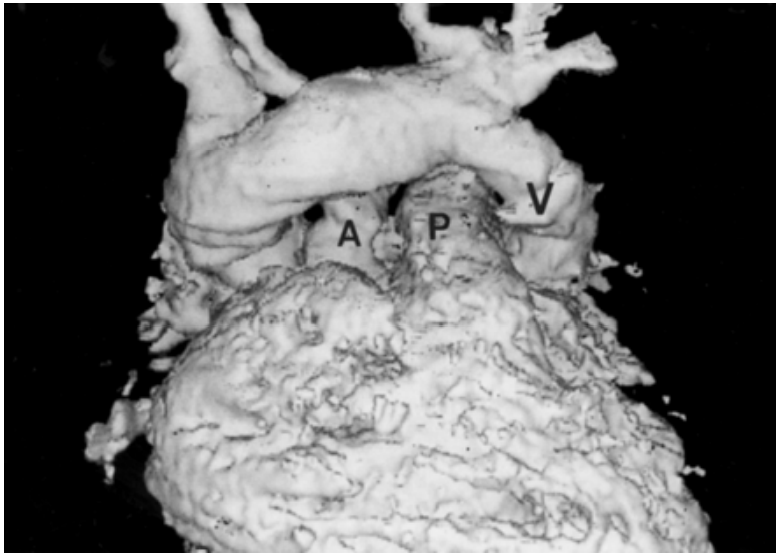
- Type 1 : supracardiac (43-50%)
- Type 2 : Cardiac (18-20%)
- Type 3 : Infracardiac type (20-27%)
- Type 4 : Mixed (10-12%)

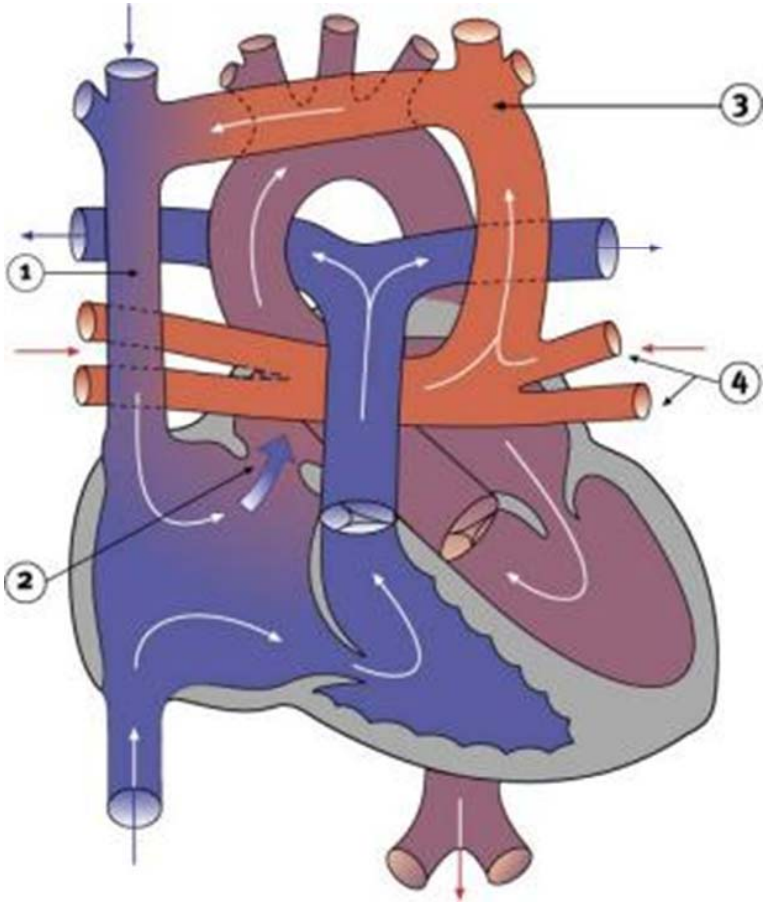
- Non-opstructed vs. Obstructed

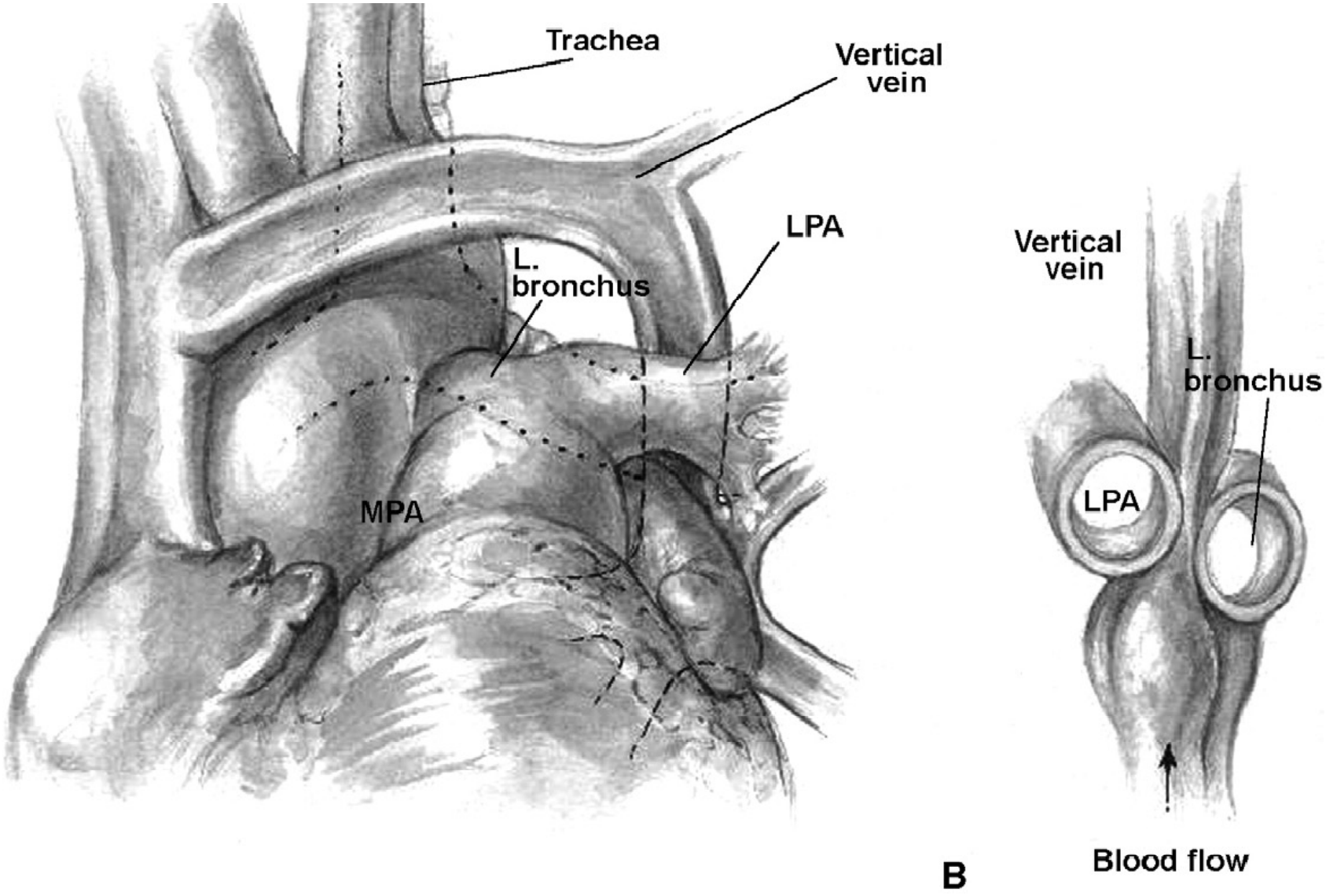


Type 1 : Supracardiac type

- Vertical vein most often drains to LIV
- Course between LPA and left main bronchis
- May present obstructed (around 50%)

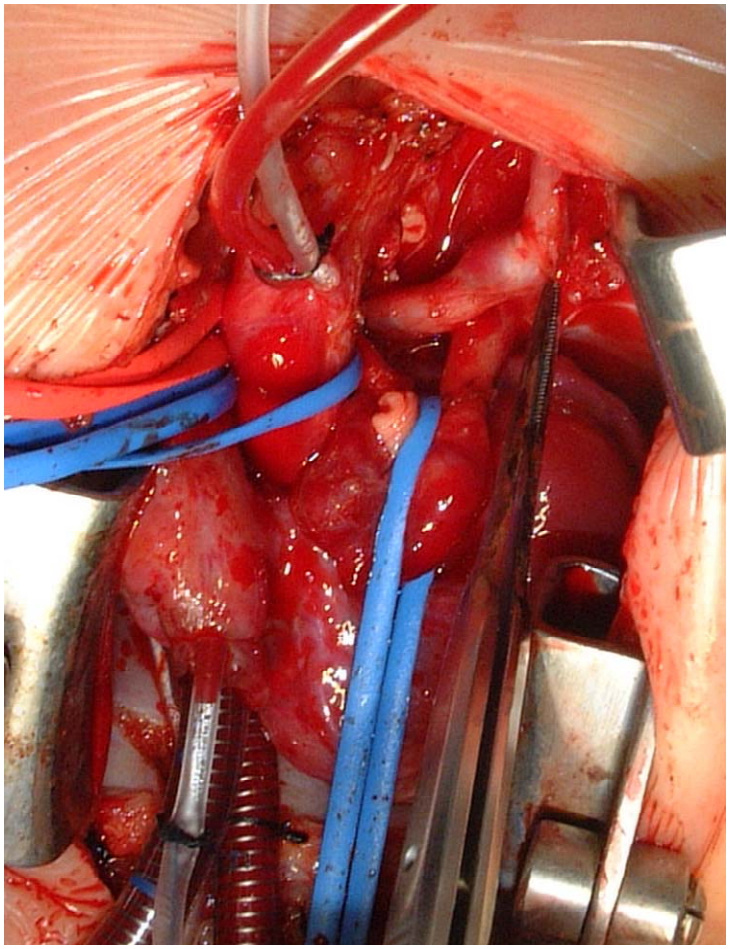


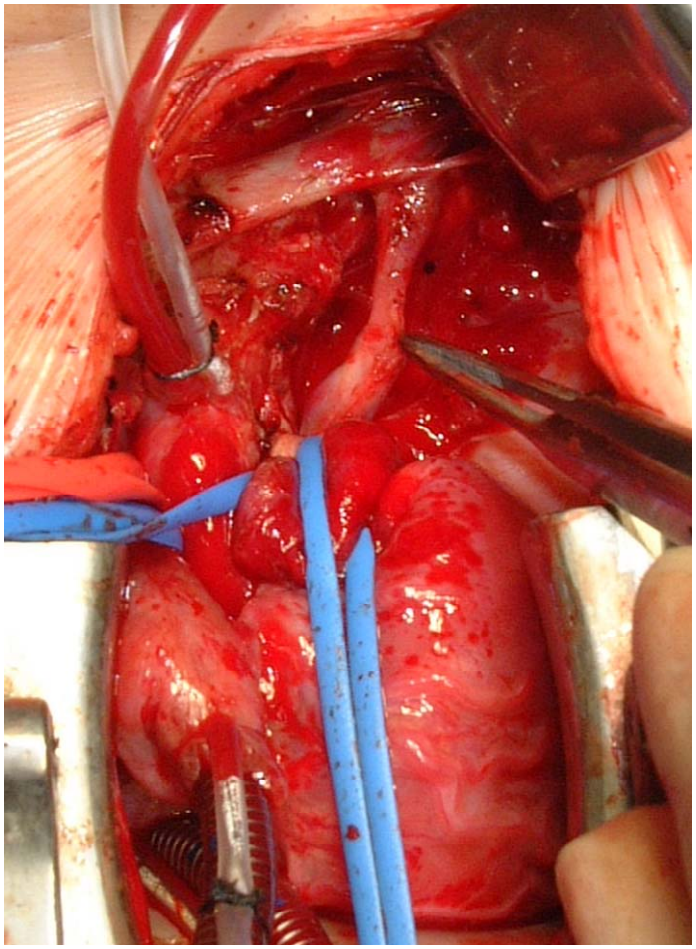



Trachea
Vertical vein
LPA
L. bronchus
MPA
Vertical vein
L. bronchus
LPA
Blood flow

B

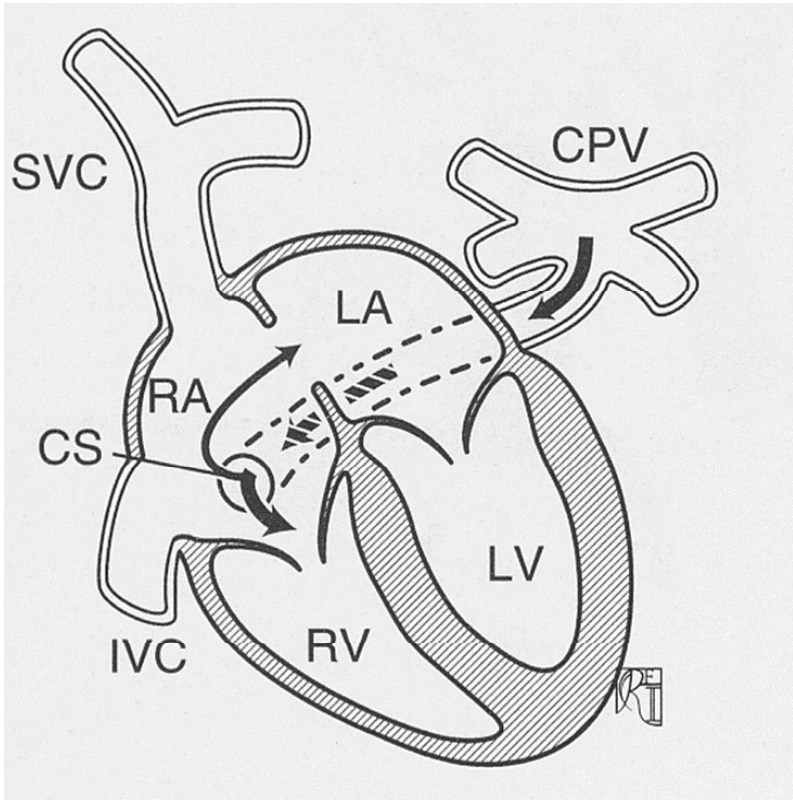
TAPVR(to innominate, obstructive type)





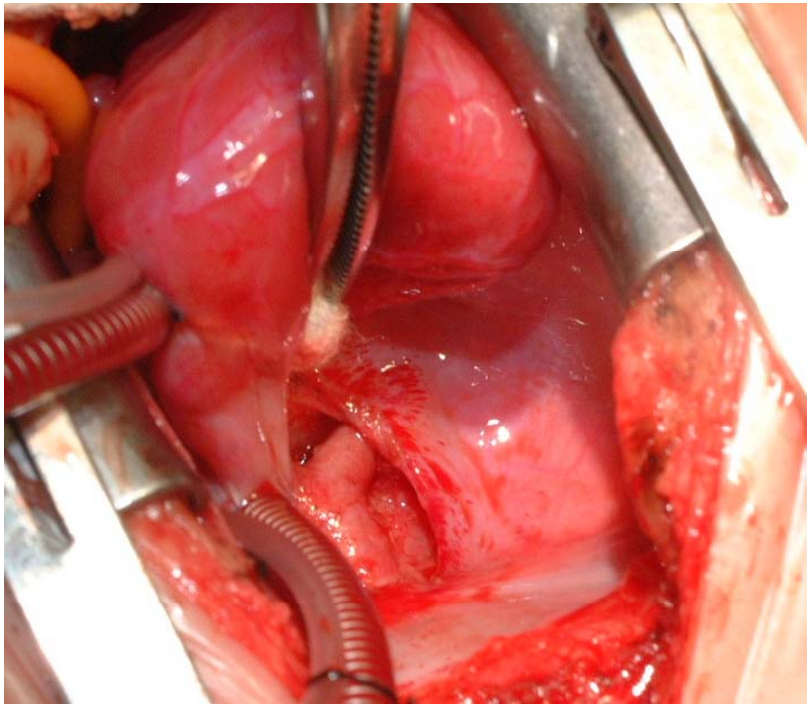
Type 2 : Cardiac type

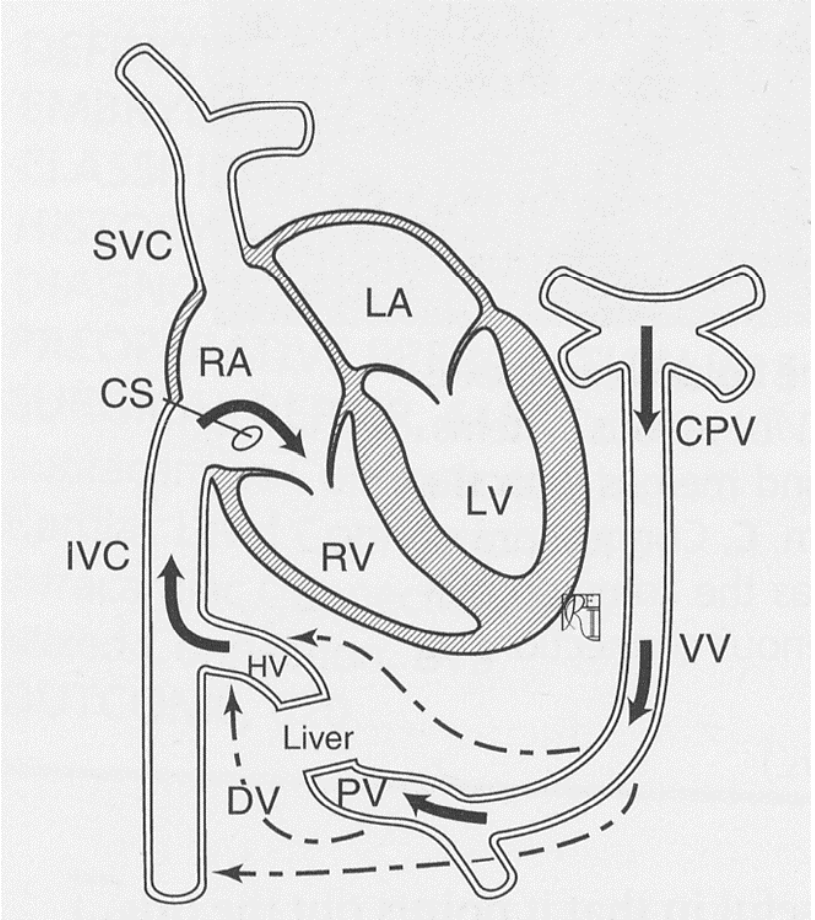
- Typically to the coronary sinus
- Less likely to be obstructed
- Can present later



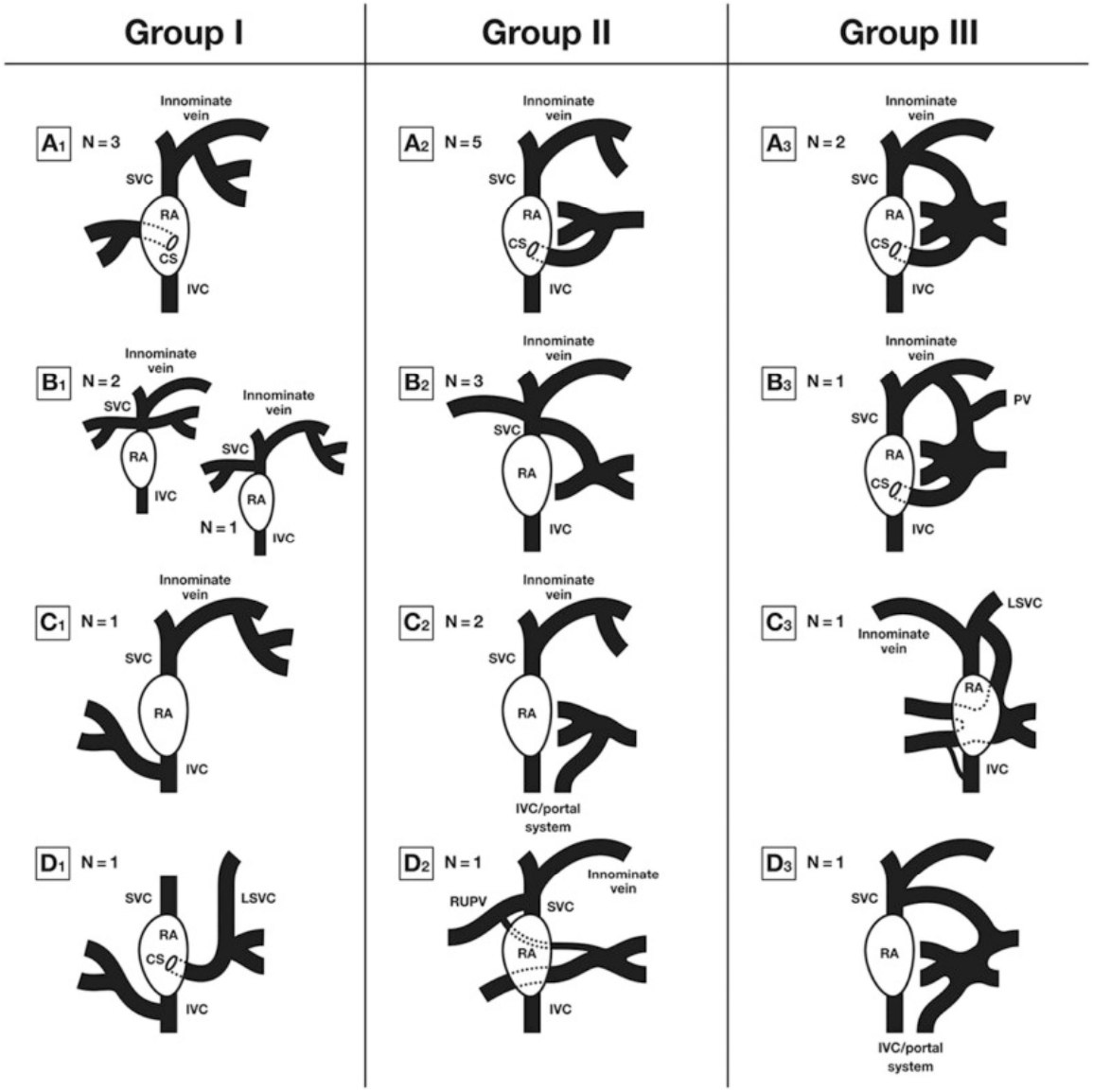
Type 3: Infracardiac type

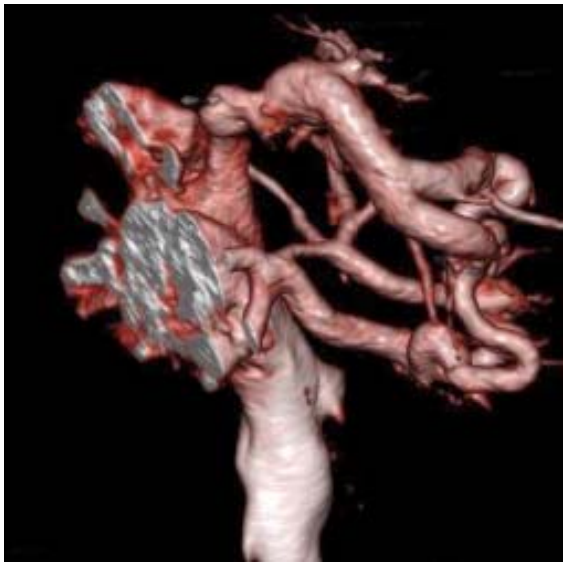
- Descending vein to portal vein, IVC, hepatic vein, or ductus venosus
- Nearly all obstructed → present at birth

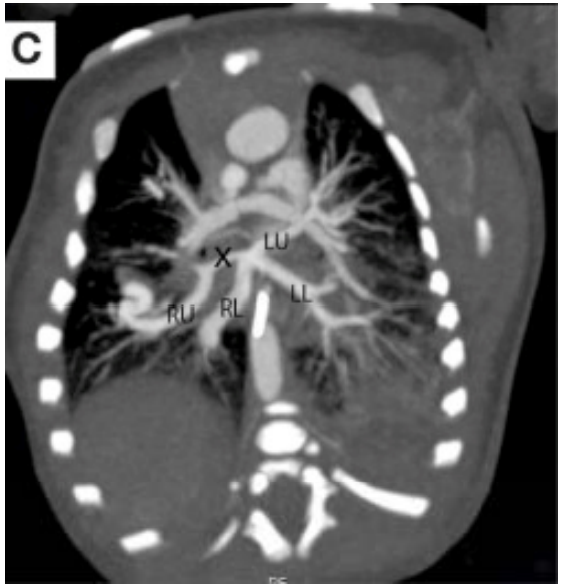




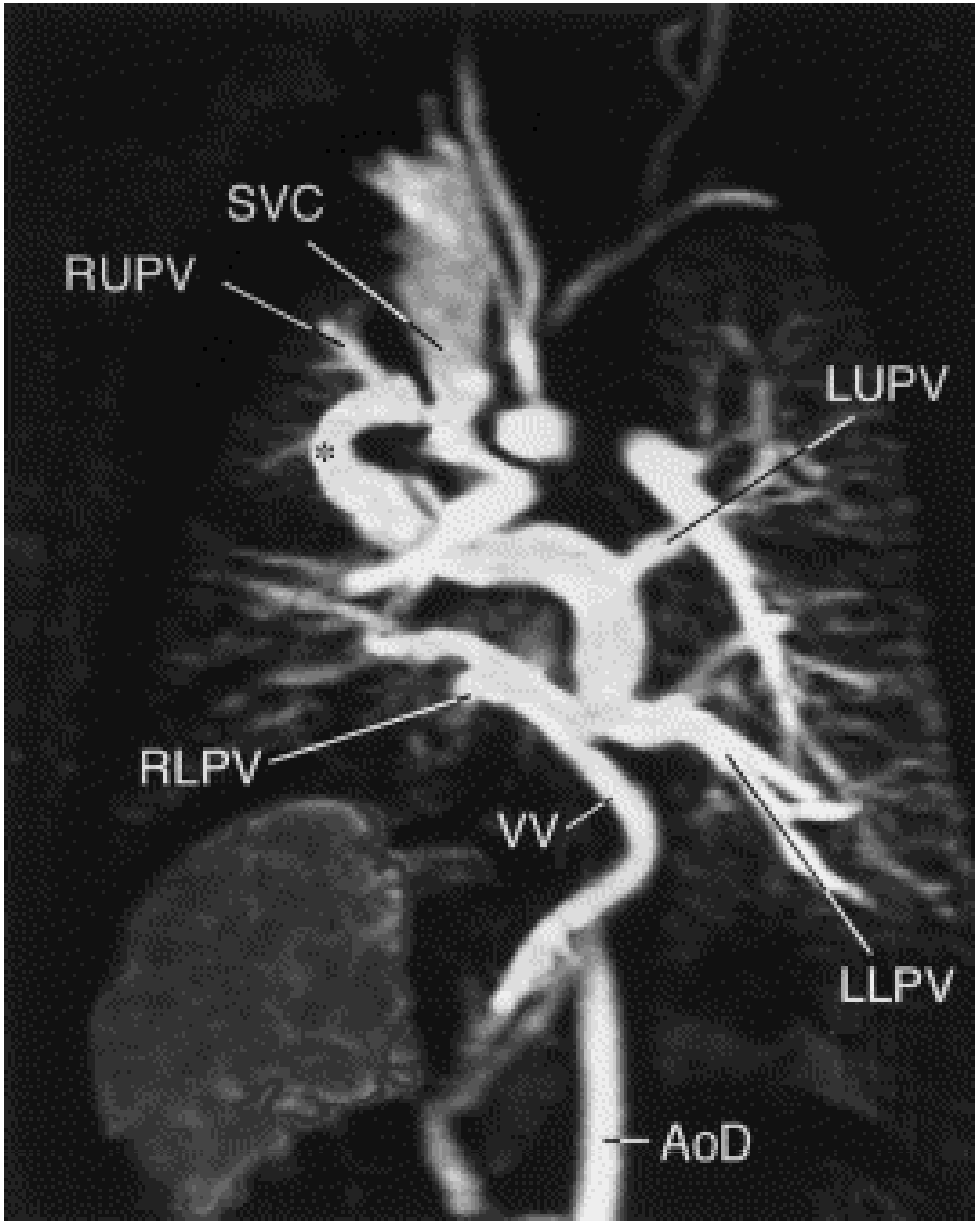
Type 4 : mixed type



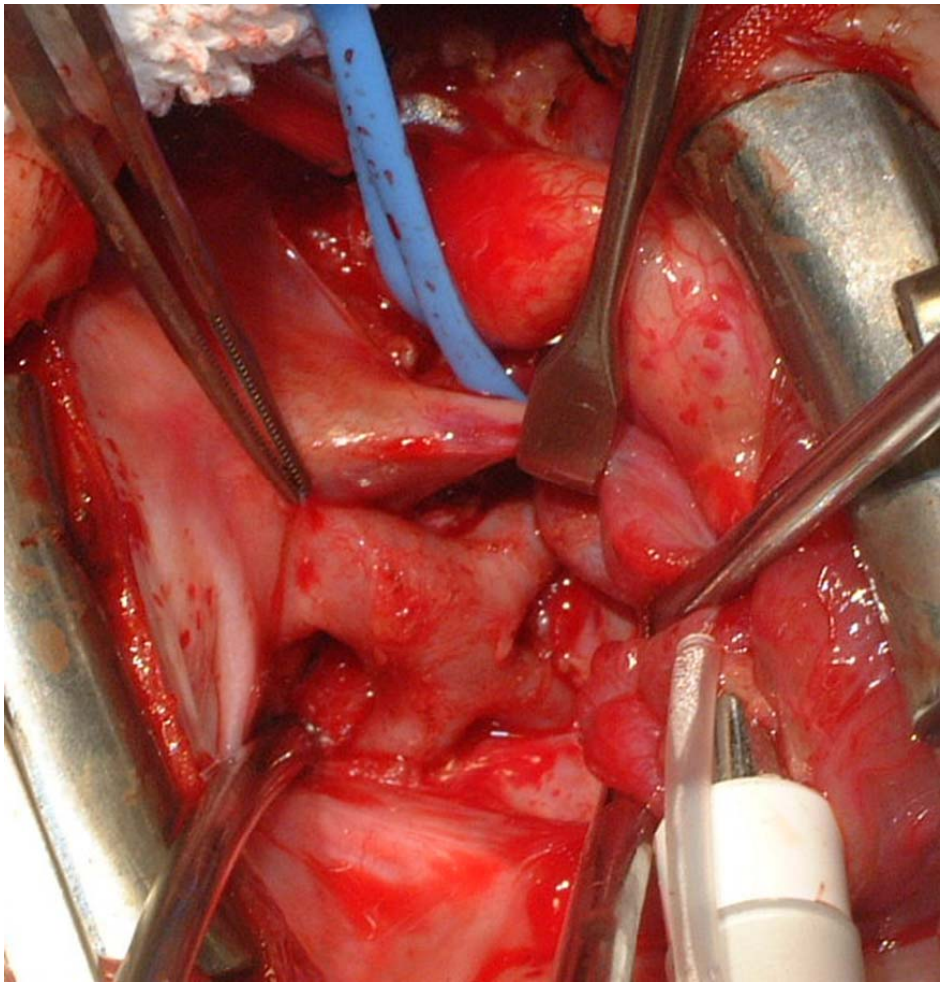




TAPVR (mixed type)



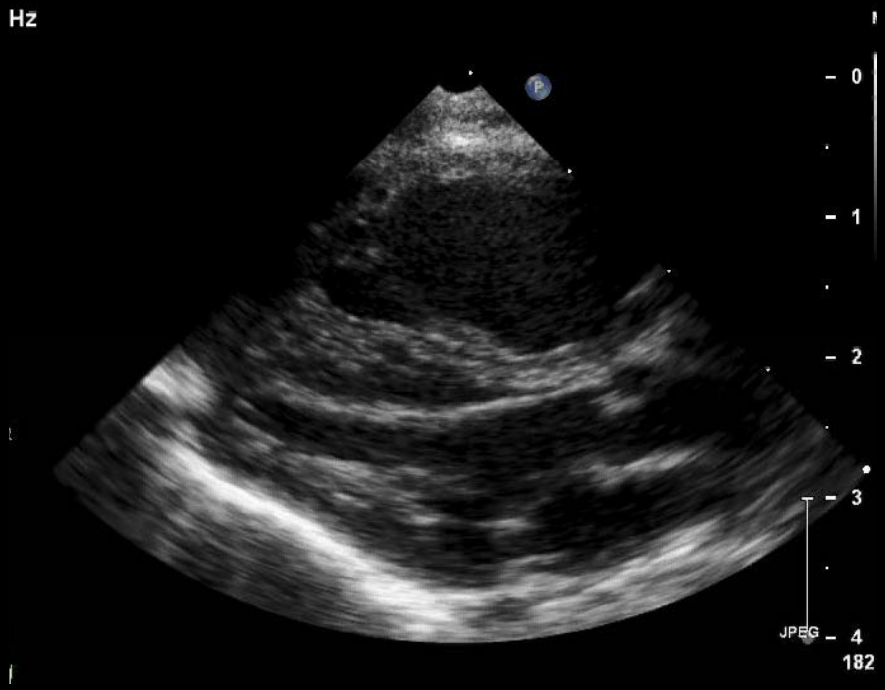
TAPVR (Rt. Isomerism)



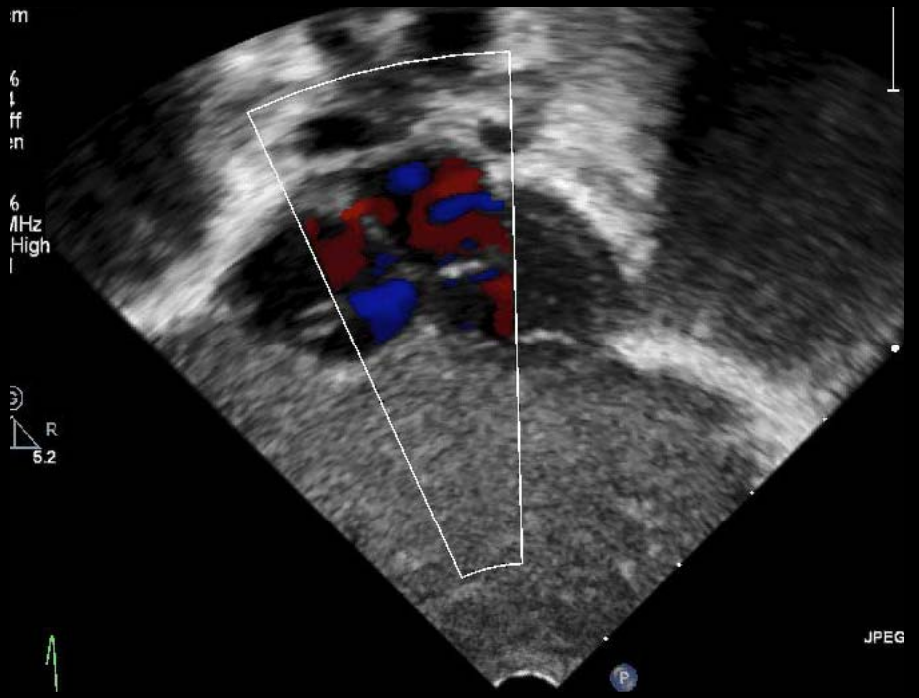
Diagnosis

- Echocardiography
- Cardiac angiography
- CT
- Cardiac MRI

Key feature of echo



Small appearing Left heart

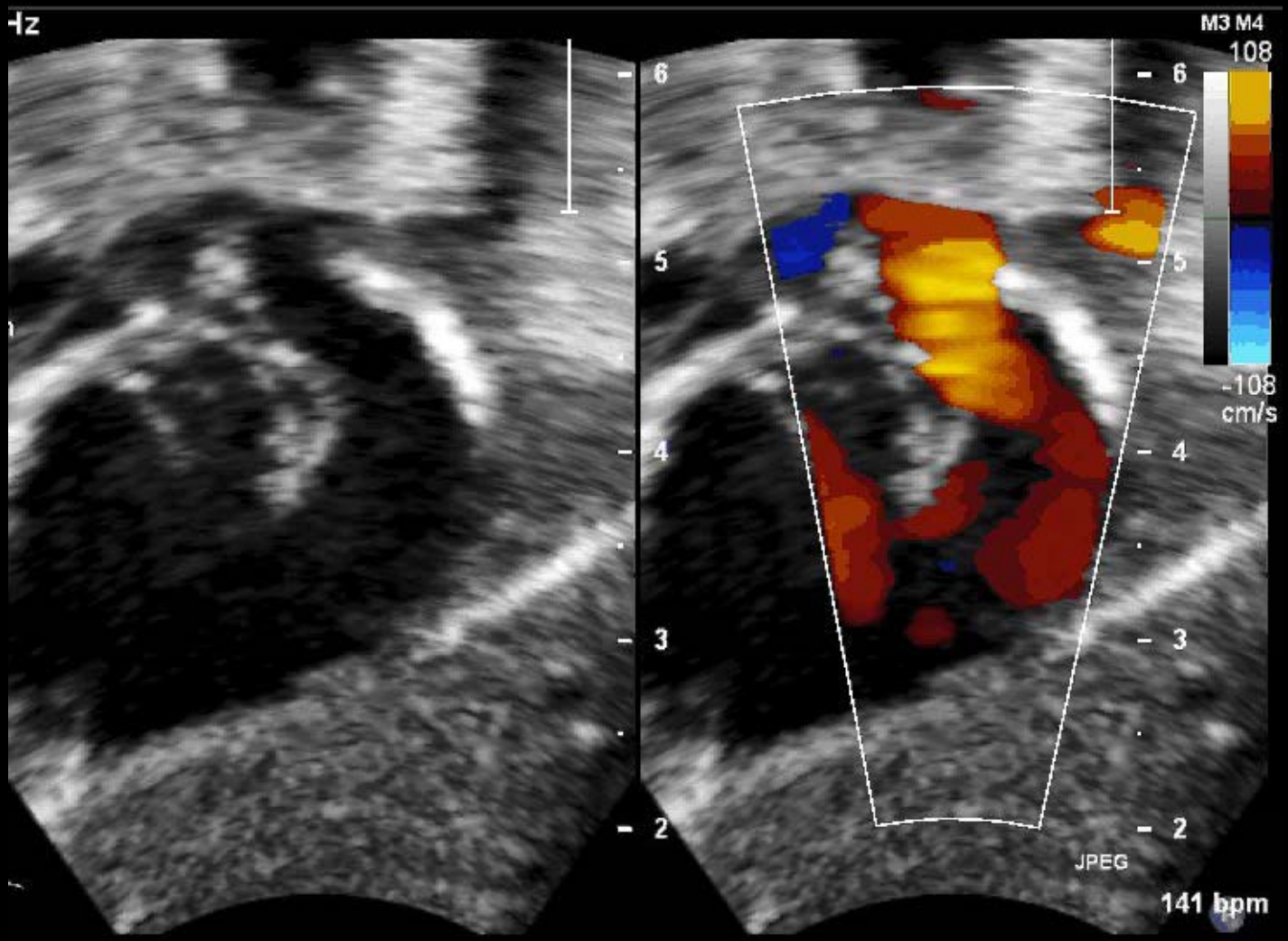


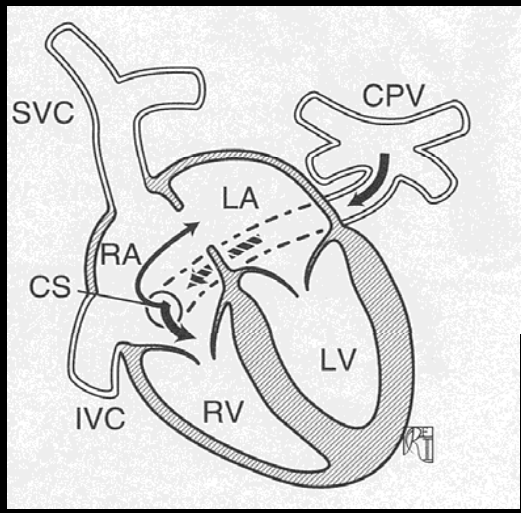
Pure R → L shunt at PFO

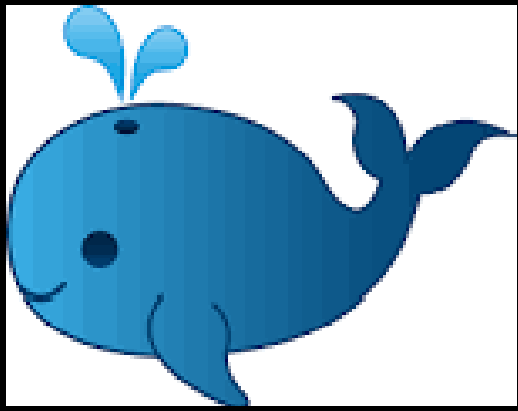
Key feature of ech.o

PV veins to confluence

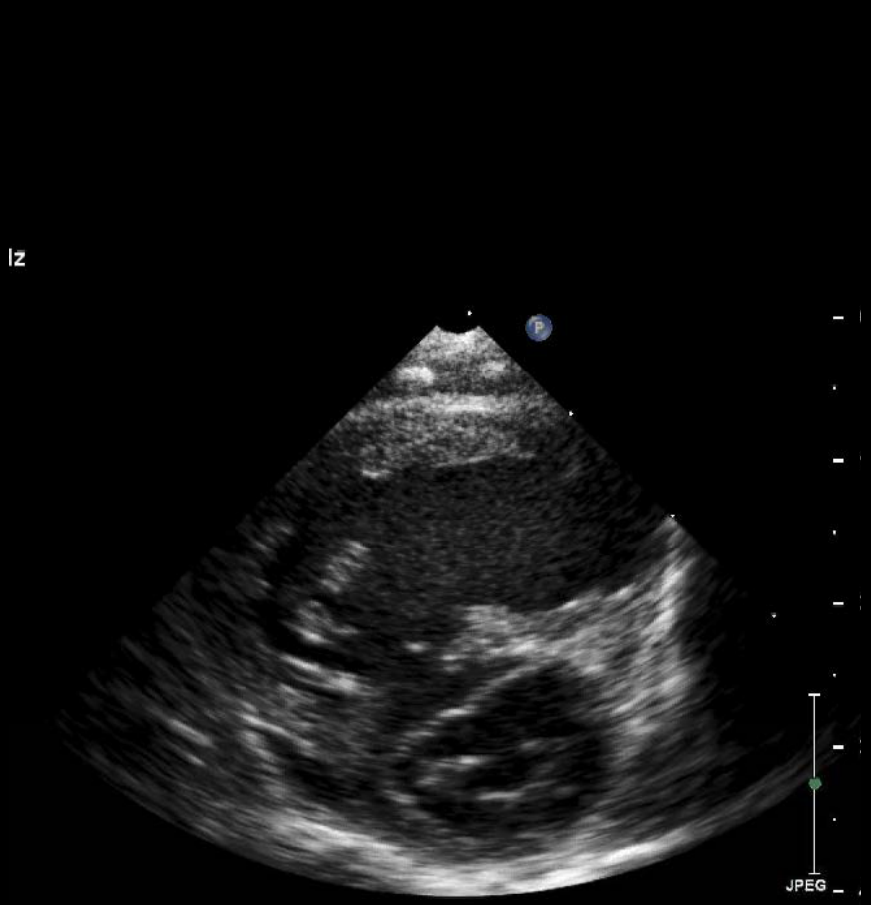
Type 2 – to the CS

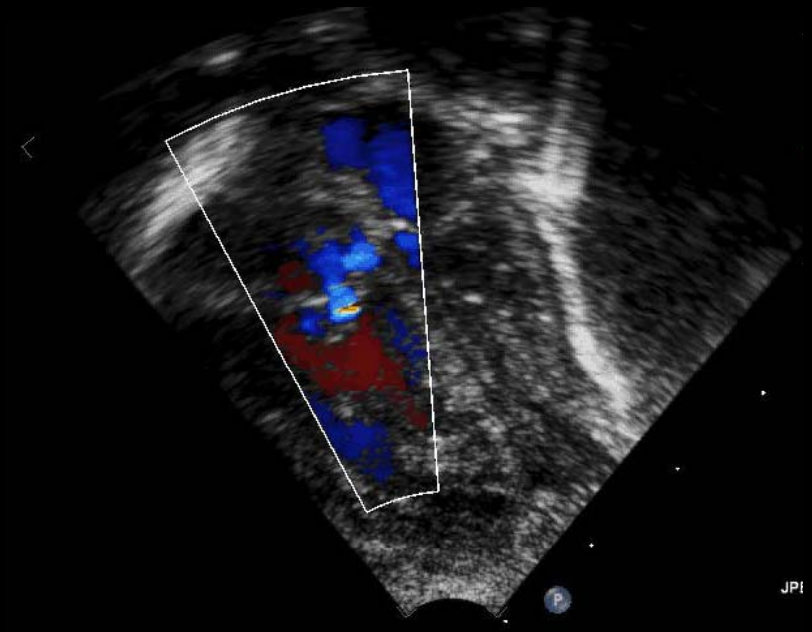


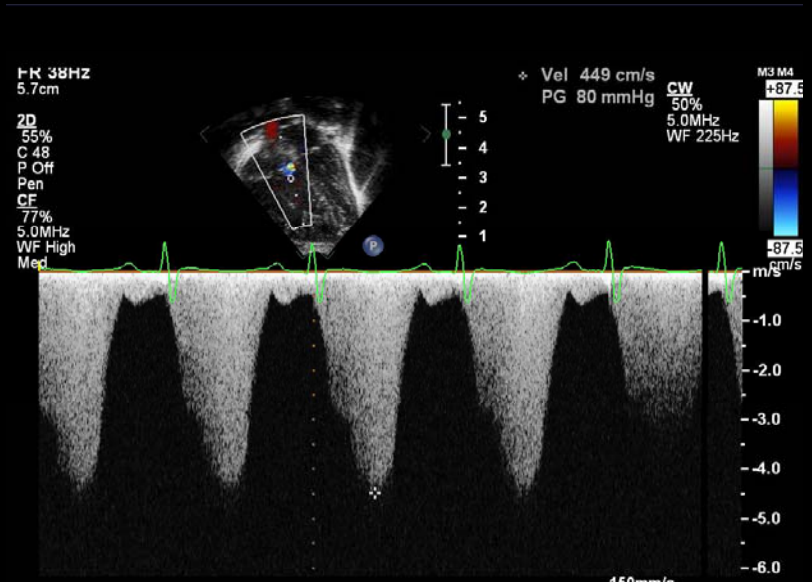




“Pulmonary hypertension”



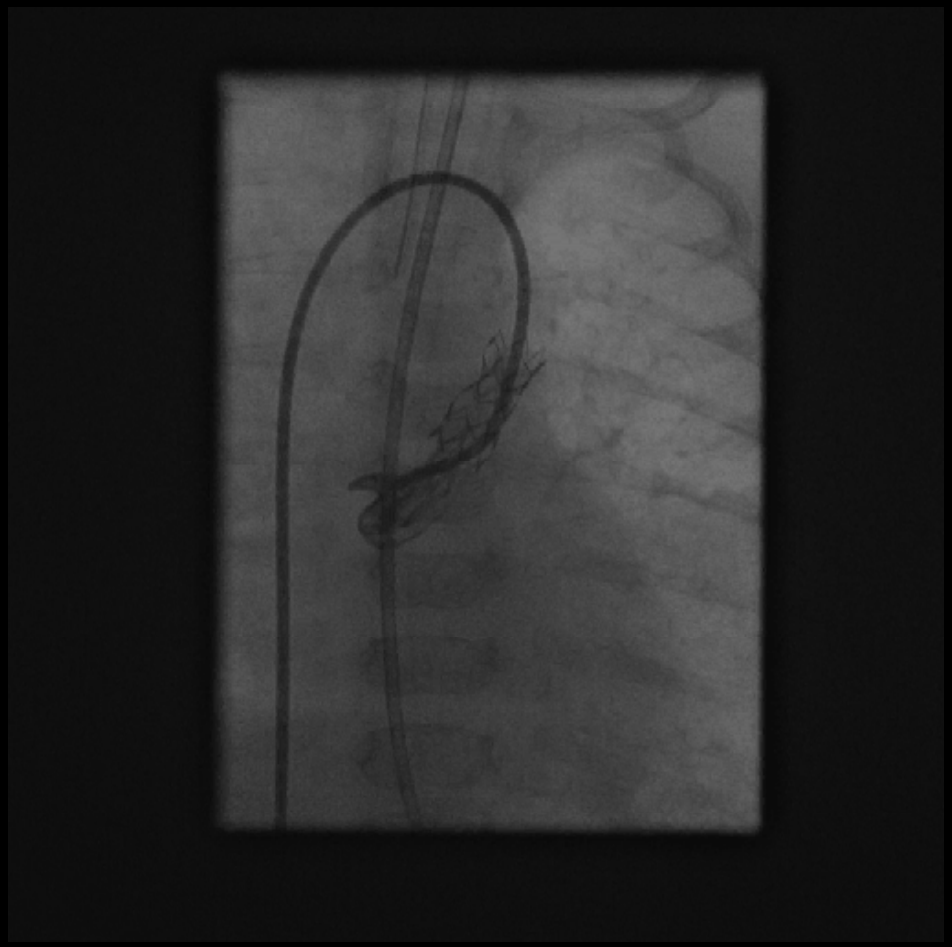




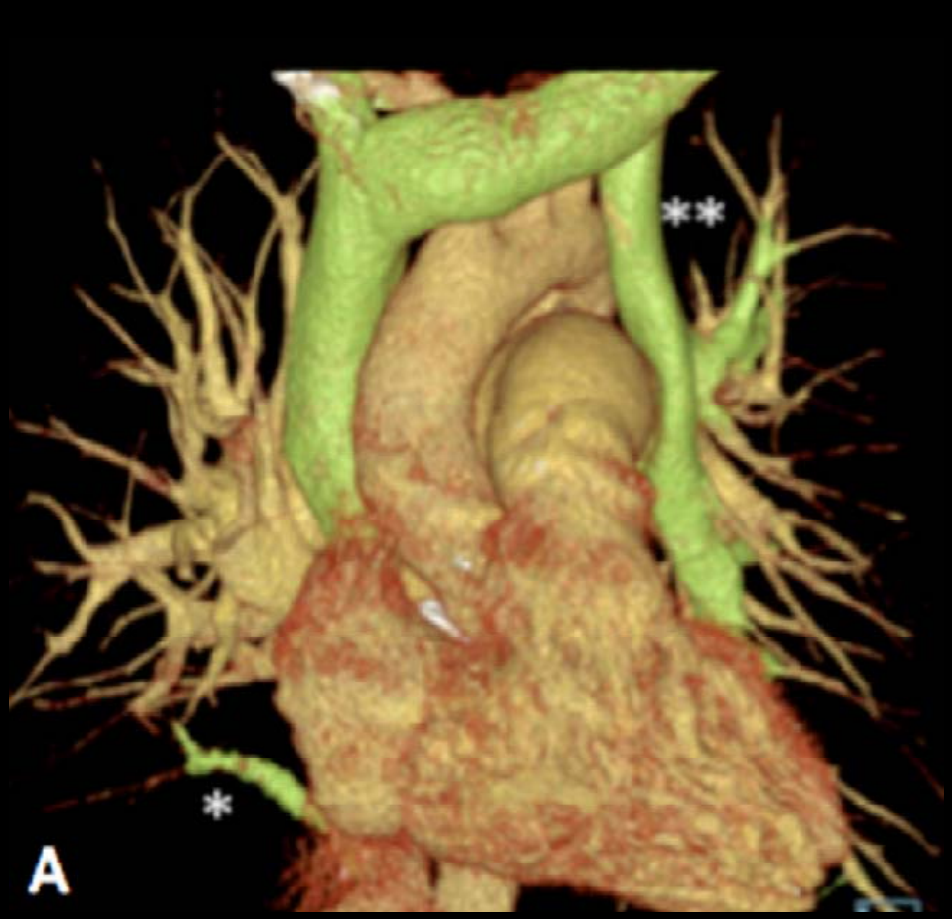
The importance of the Atrial communication

All preload to the LV is supplied by right to left shunting across the atrial communication

Cardiac catheterization – stenting vertical vein



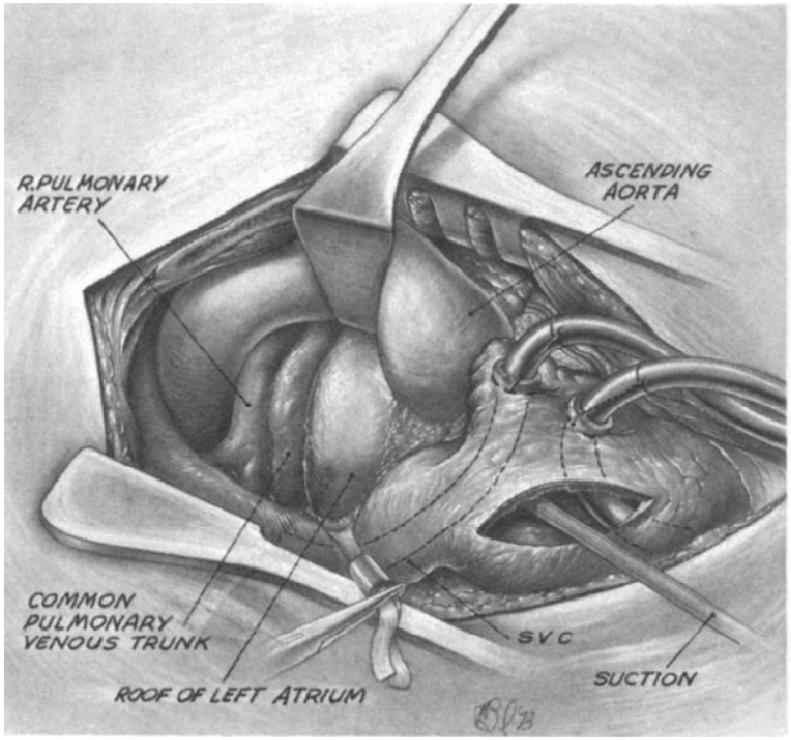
CT angiogram

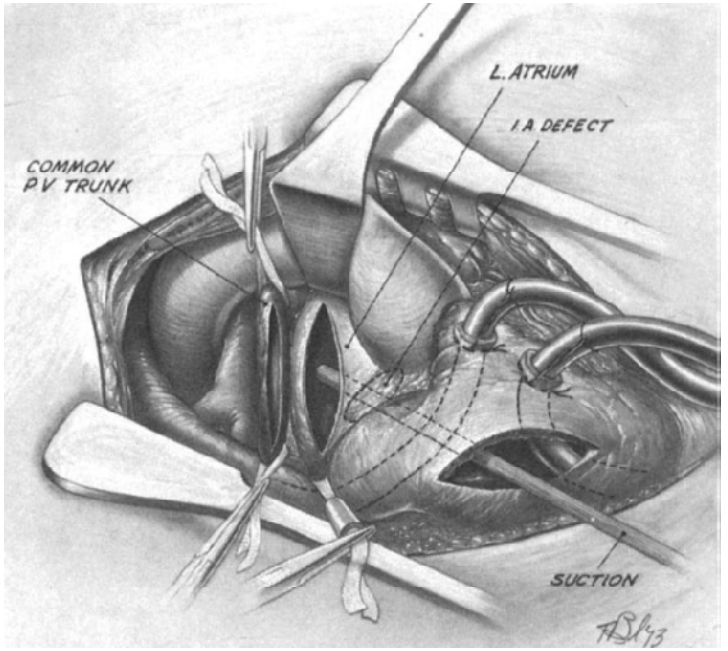


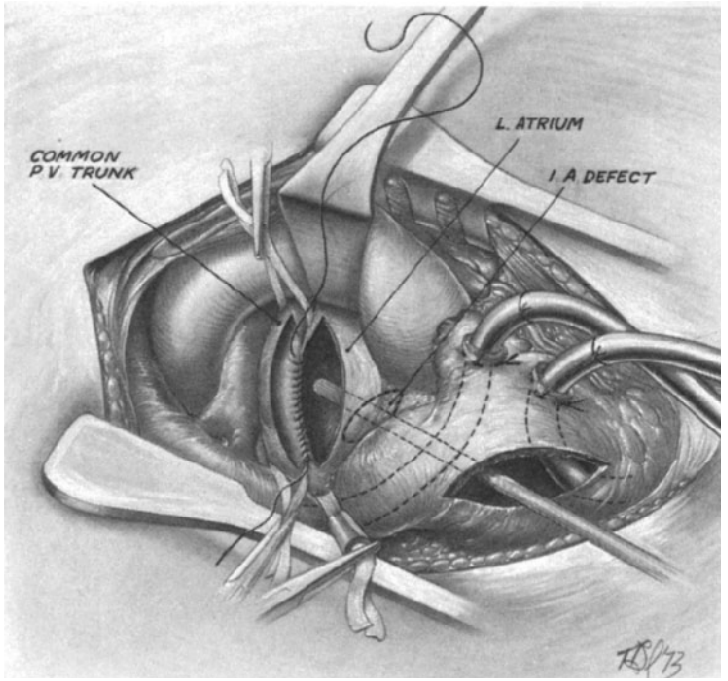
Surgery

- The superior approach
- Lateral approach
- “In situ” technique
- Sutureless repair
  - Surgery for pulmonary venous obstruction after repair of TAPVC
- Primary sutureless Repair

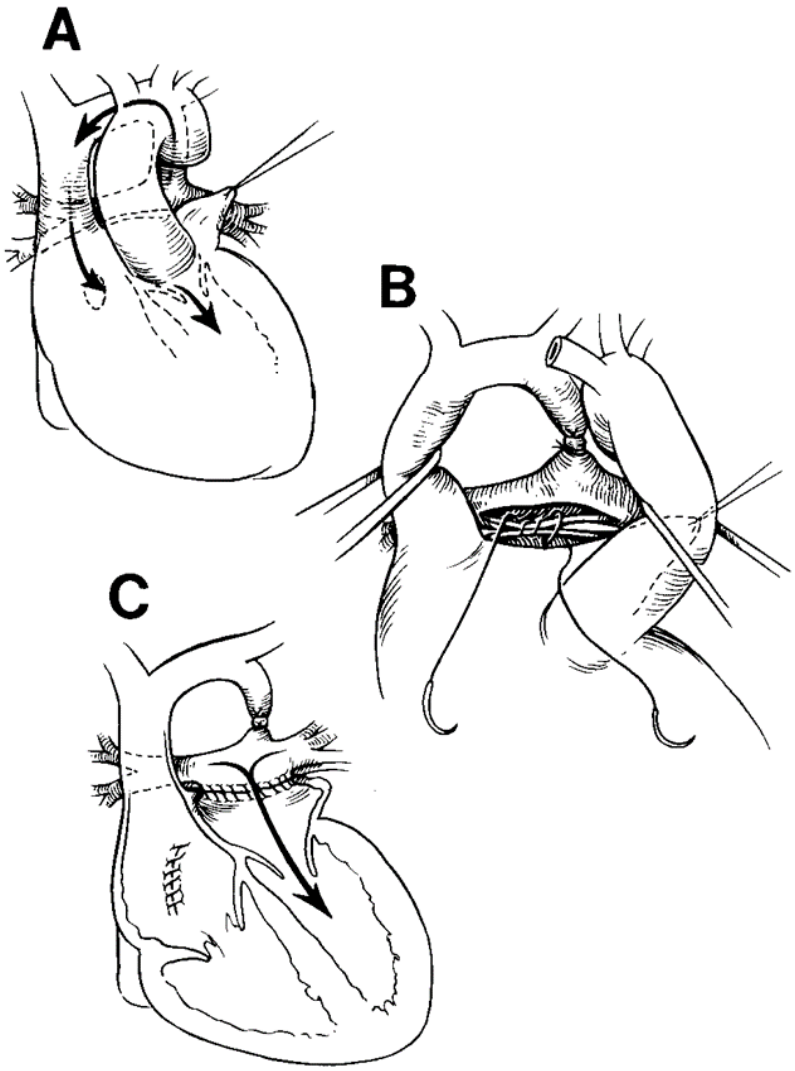
Superior approach

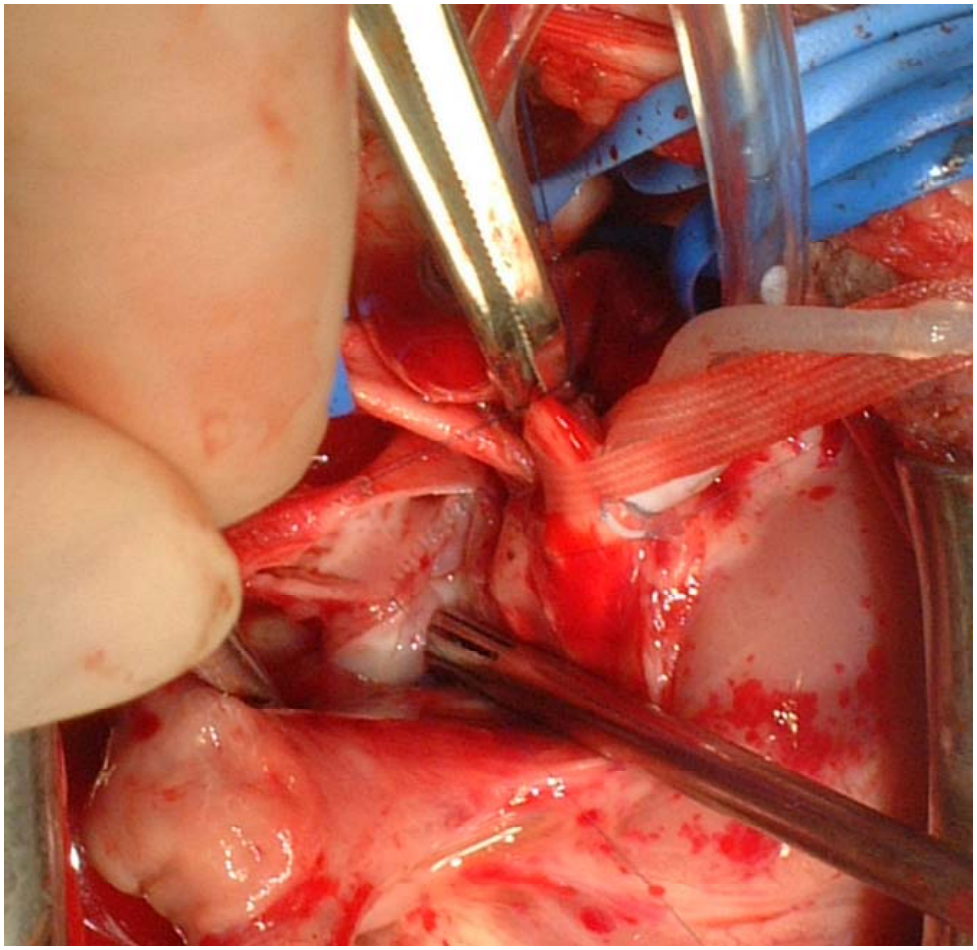




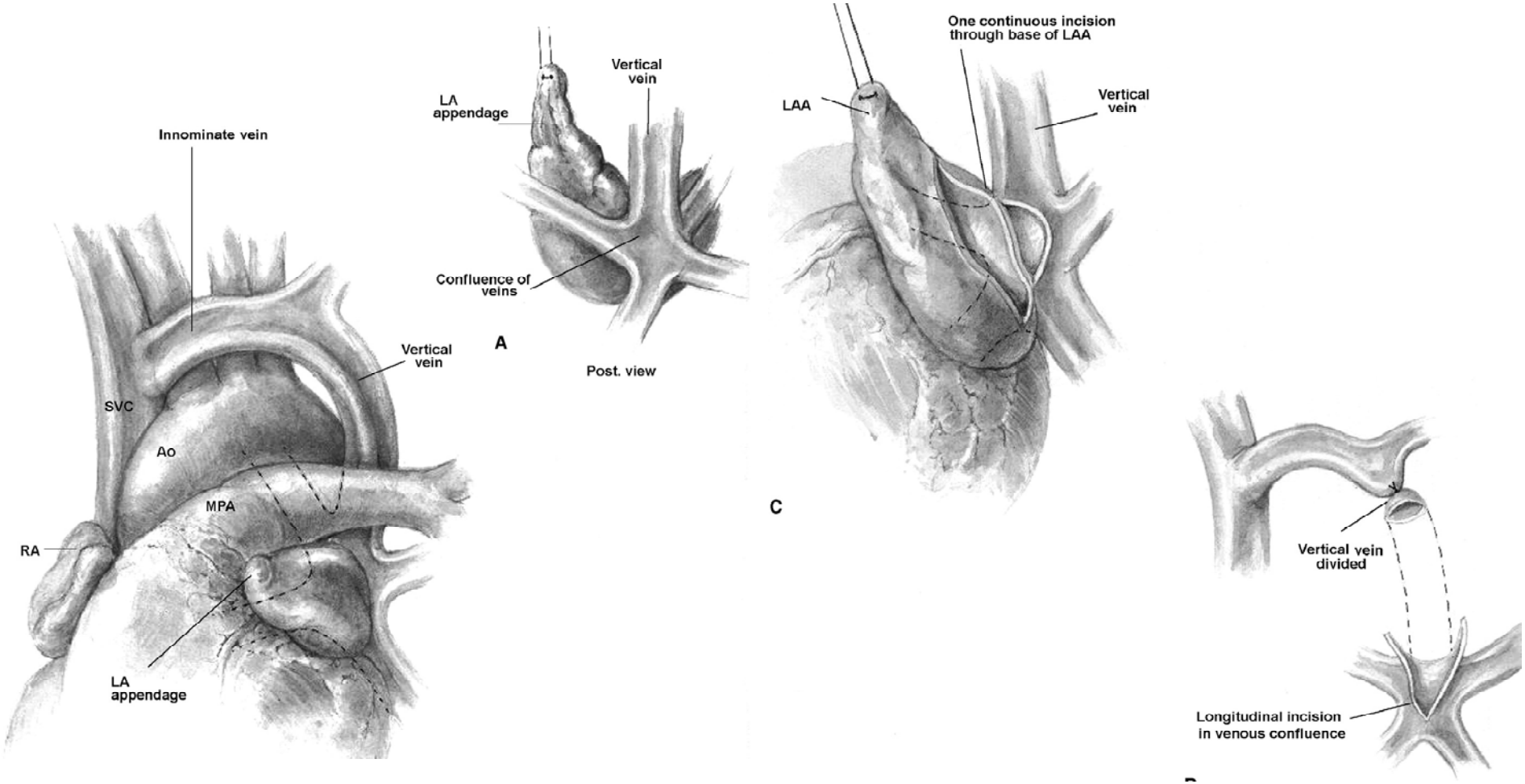


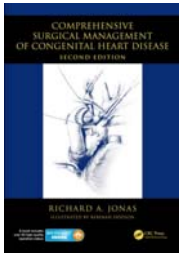
Transverse sinus approach



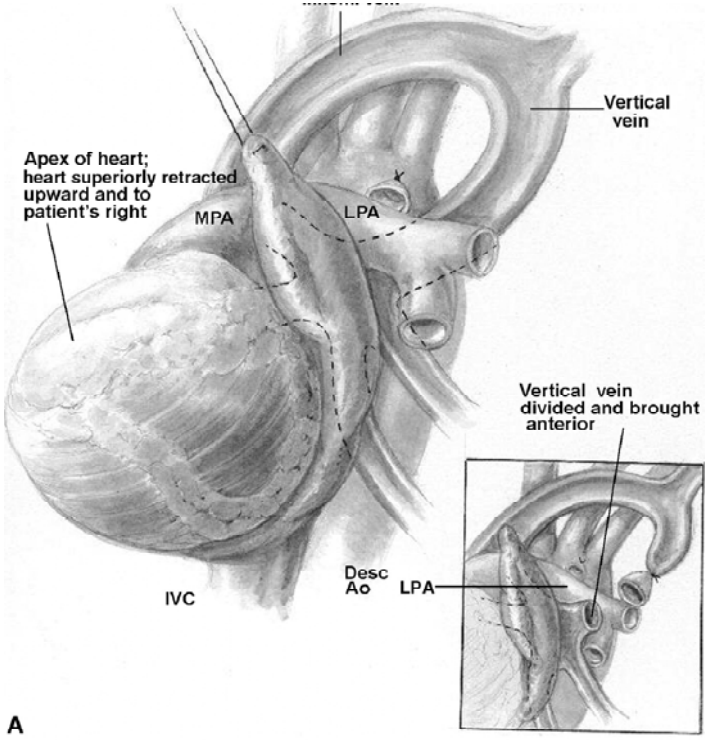


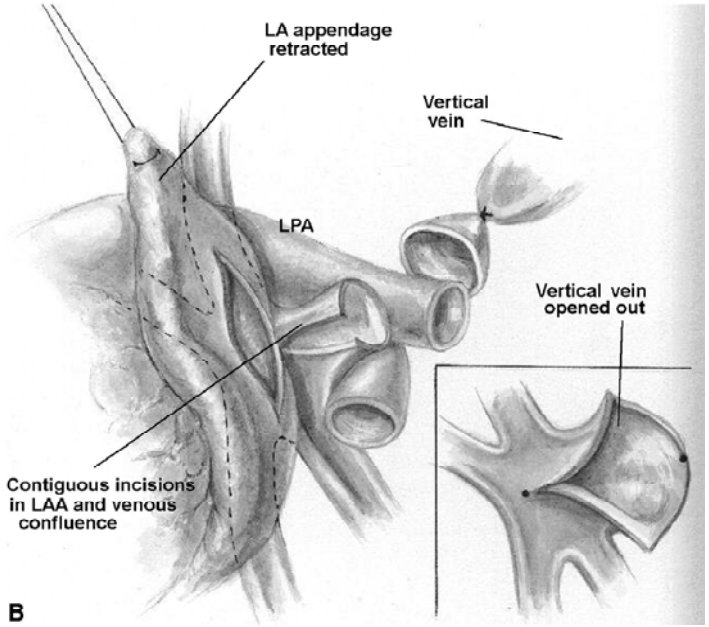
Lateral approach

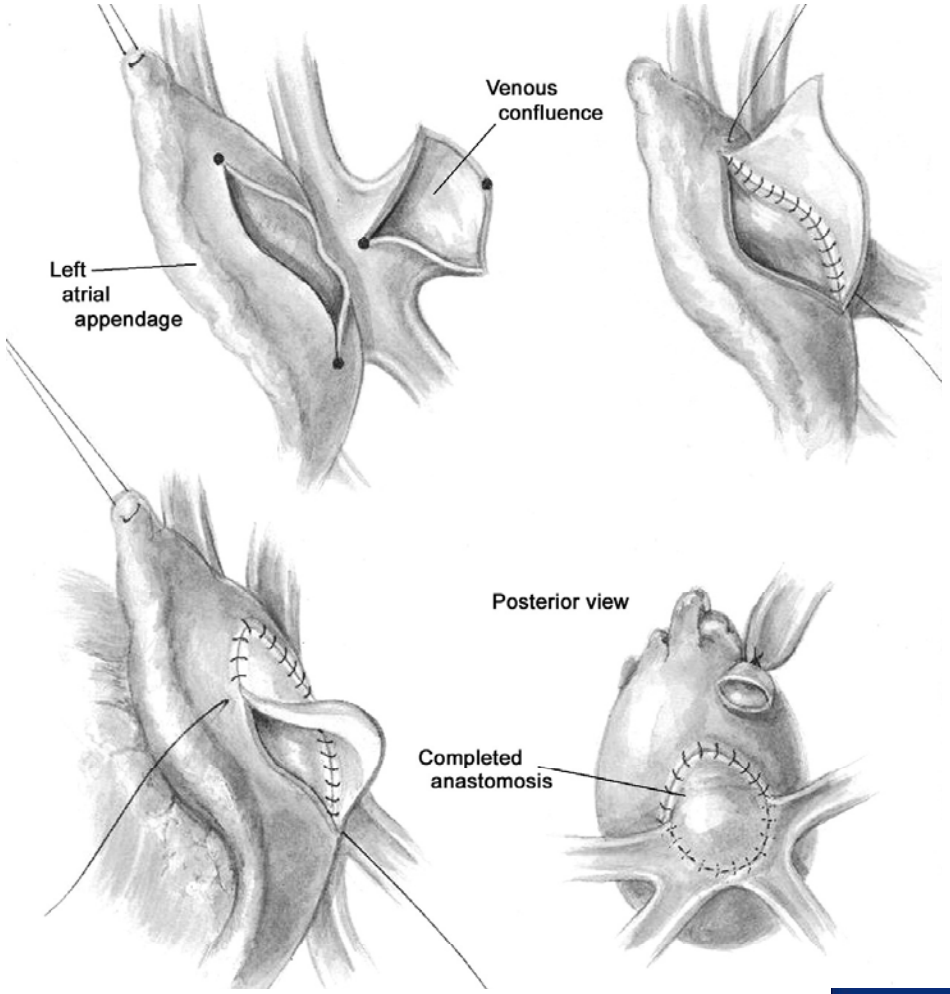


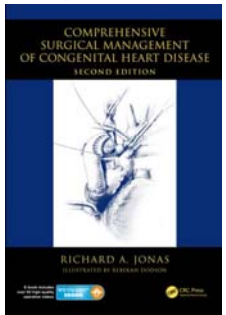


Lateral approach

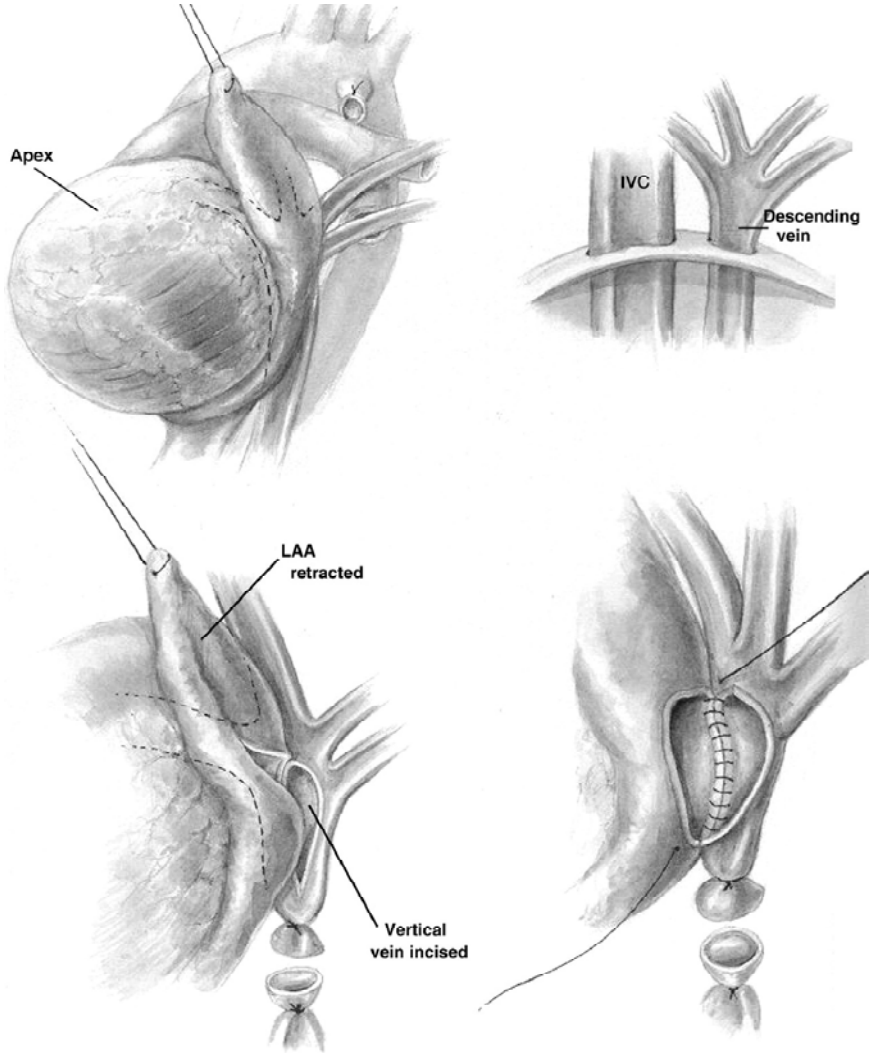


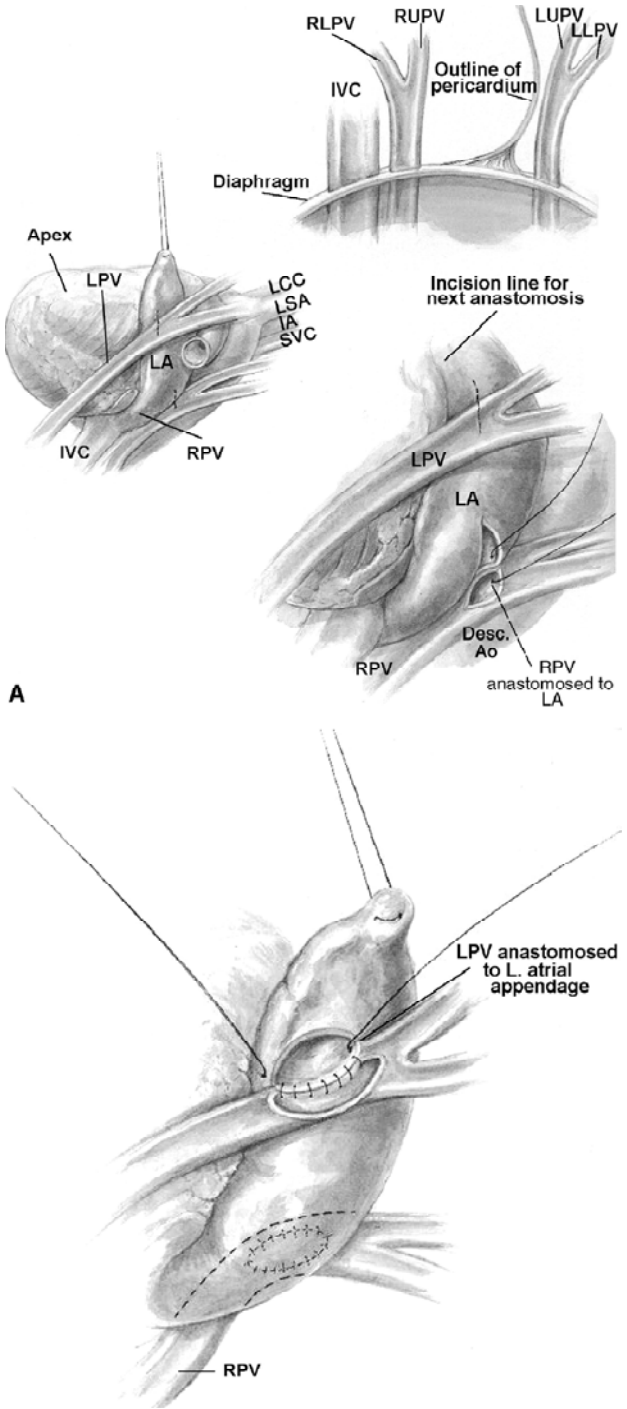


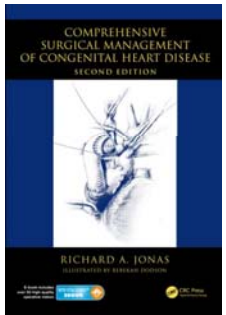




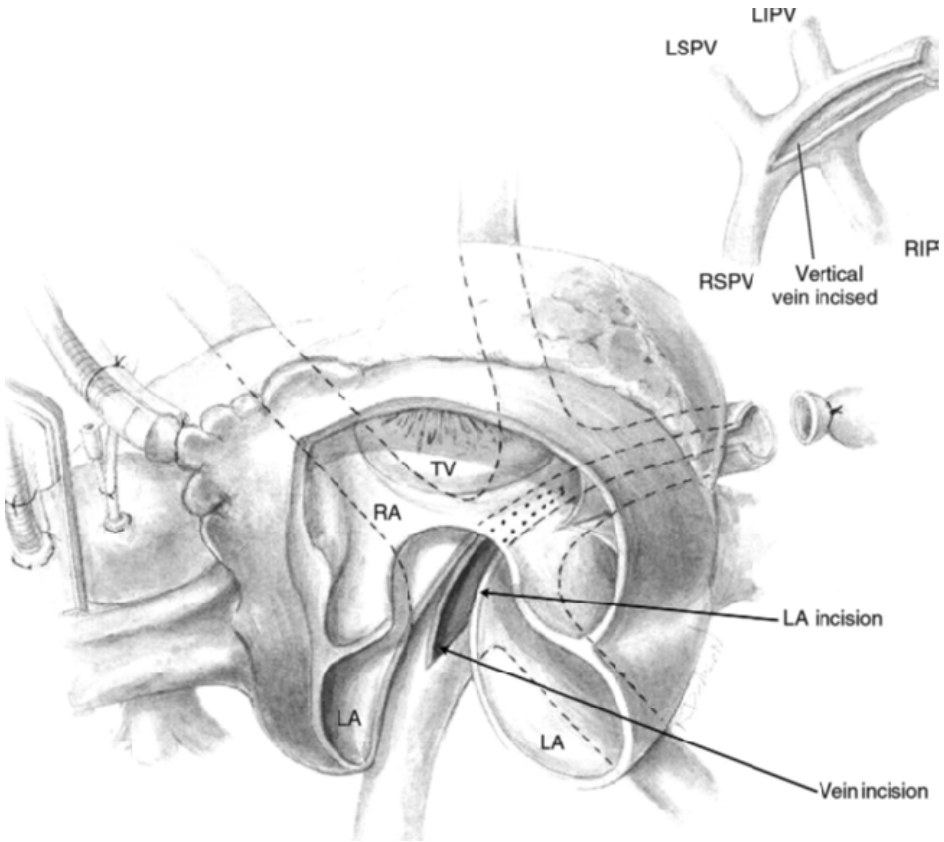
Lateral approach

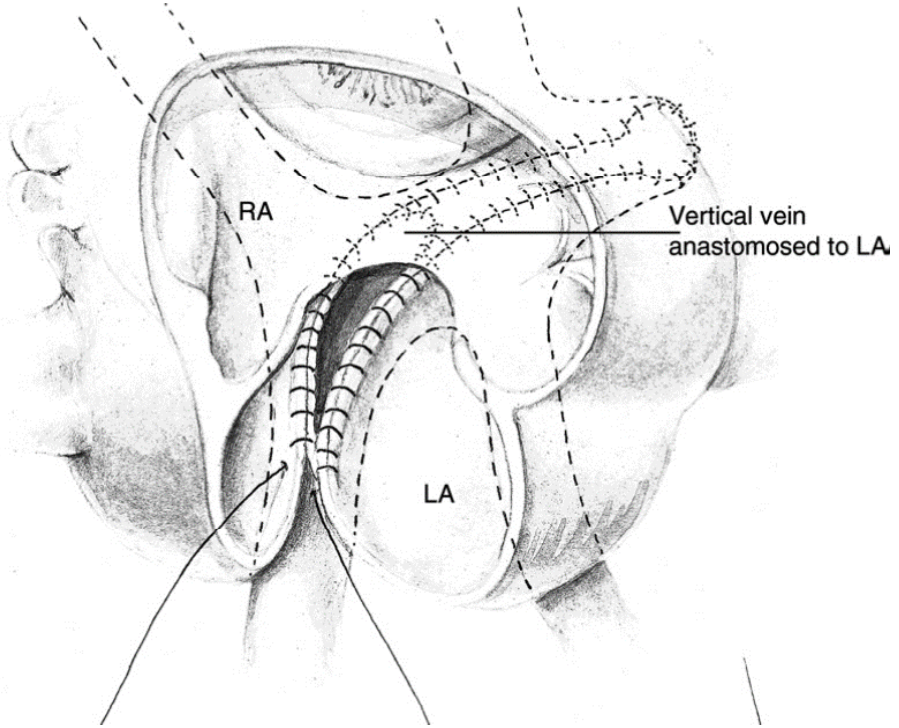


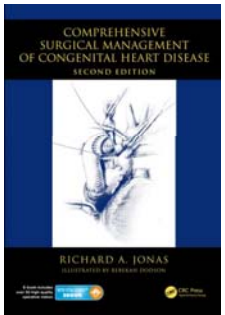




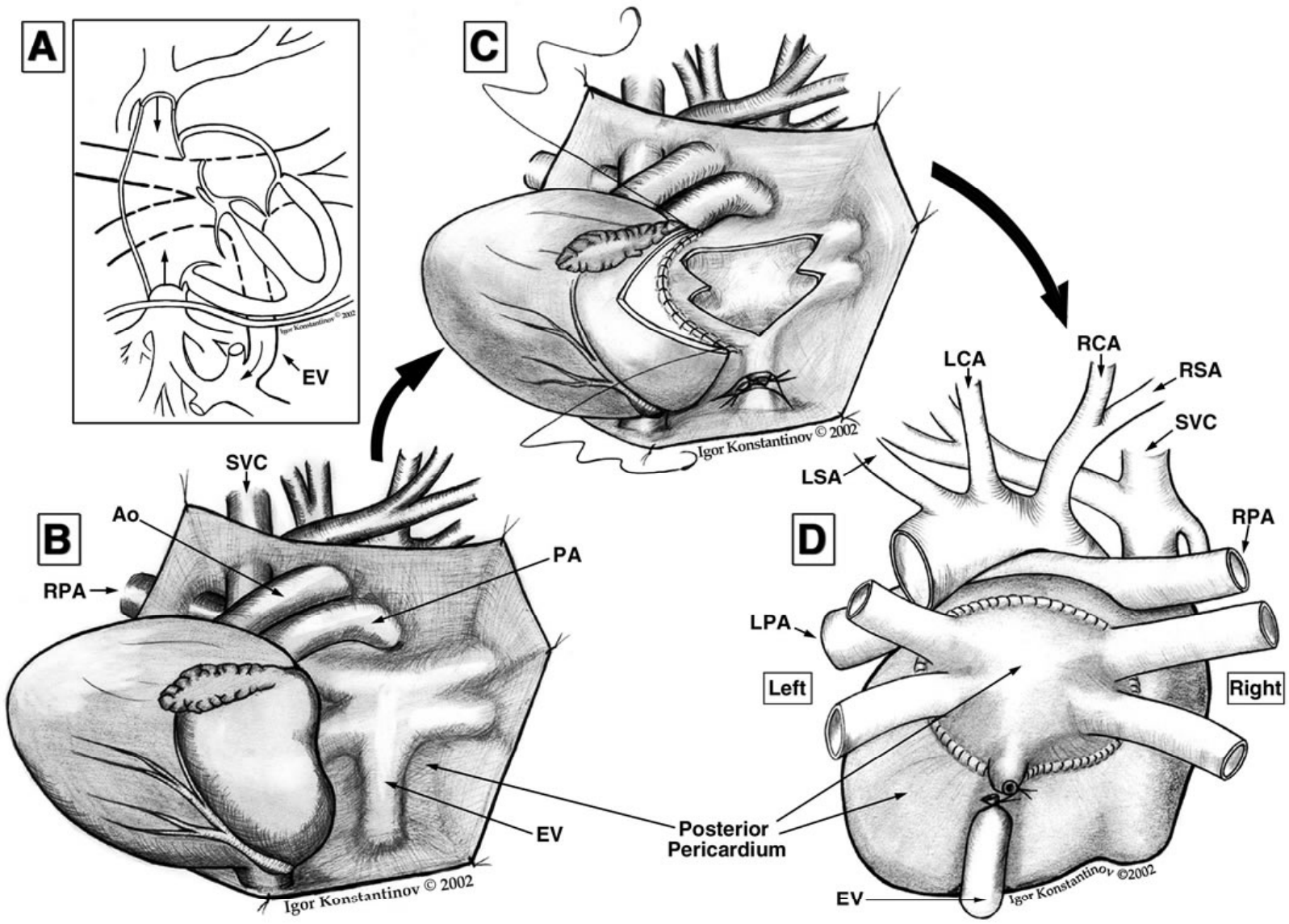
“In situ” technique



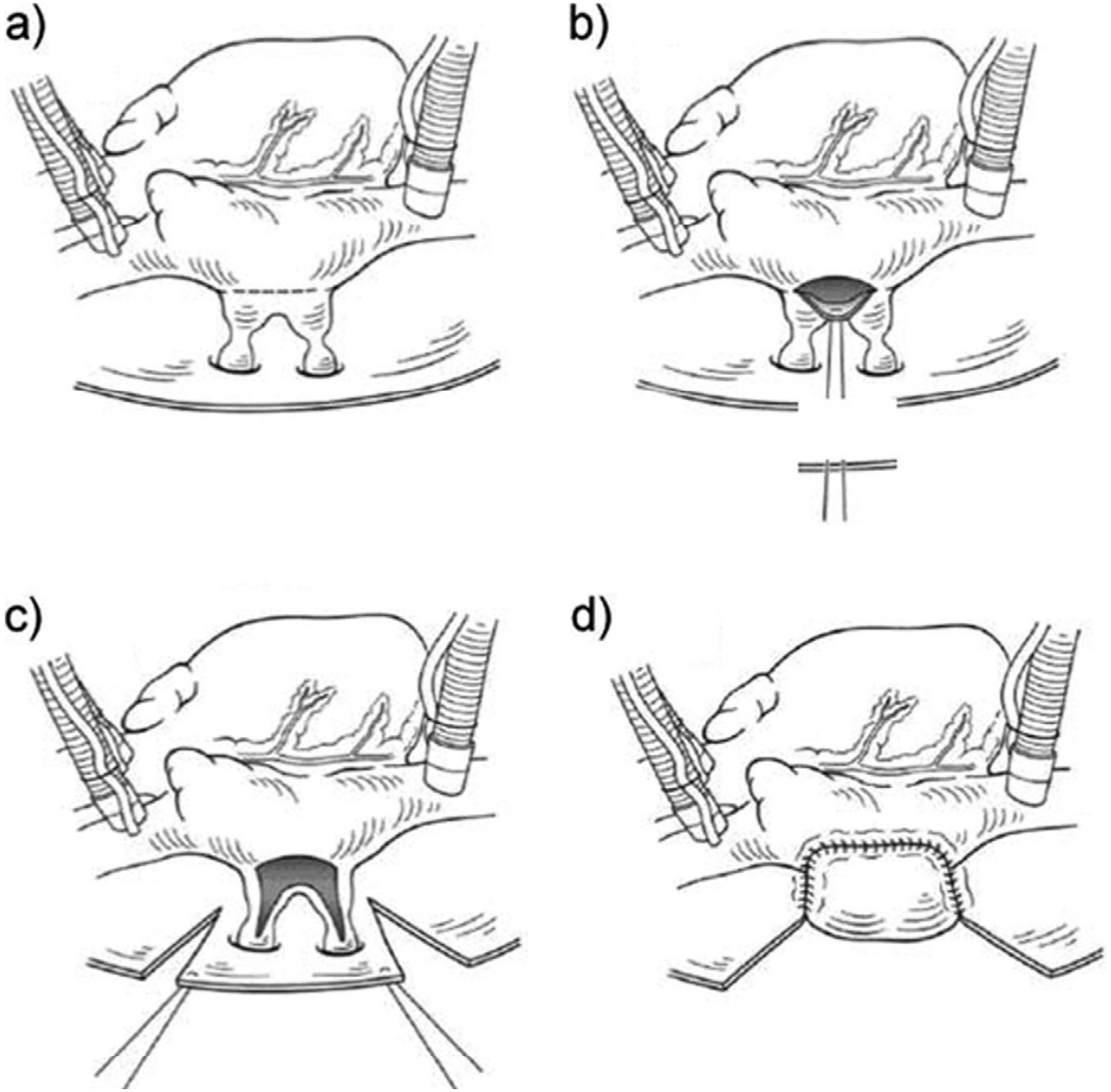




Sutureless repair



Surgery for post-repair pulmonary vein stenosis



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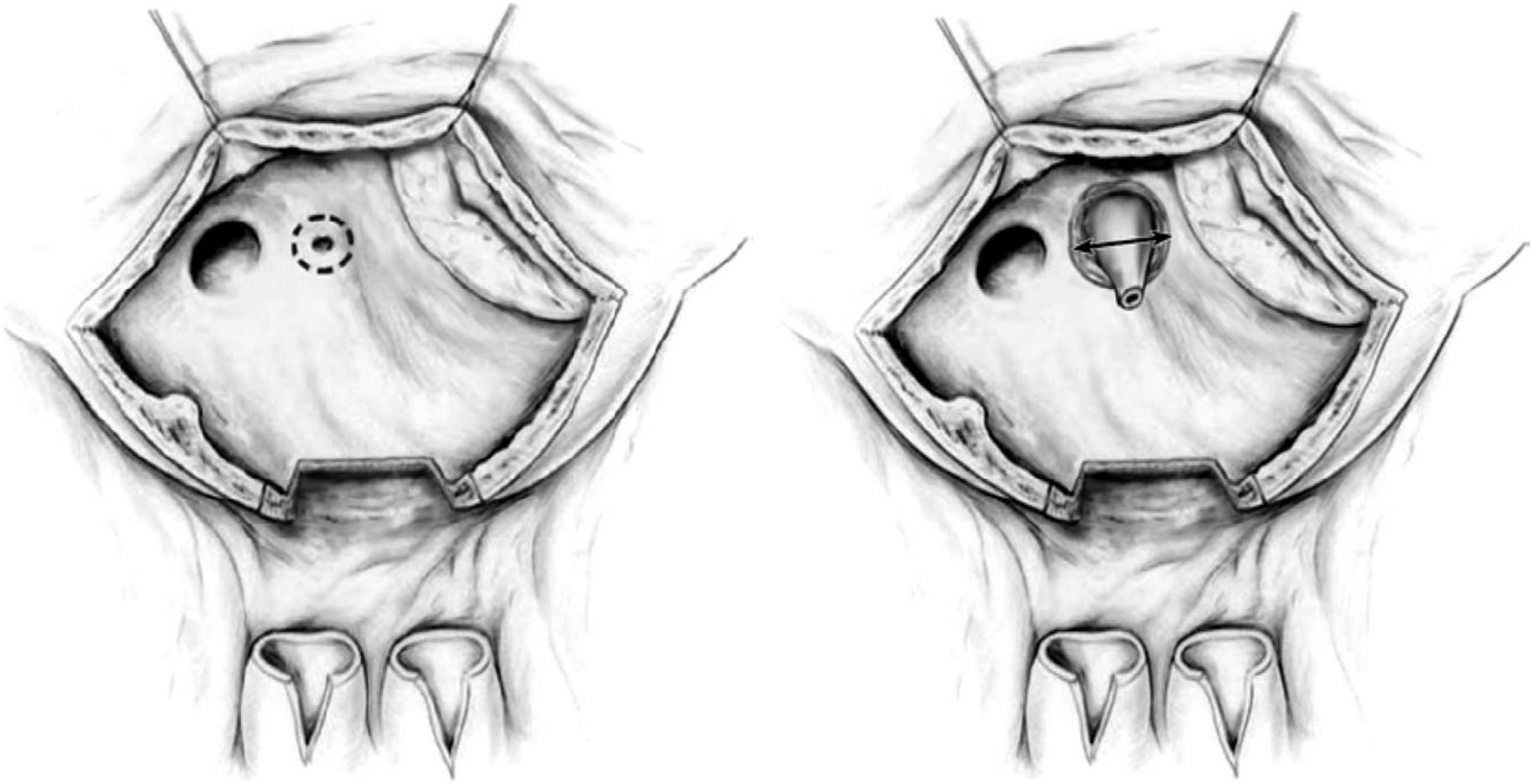
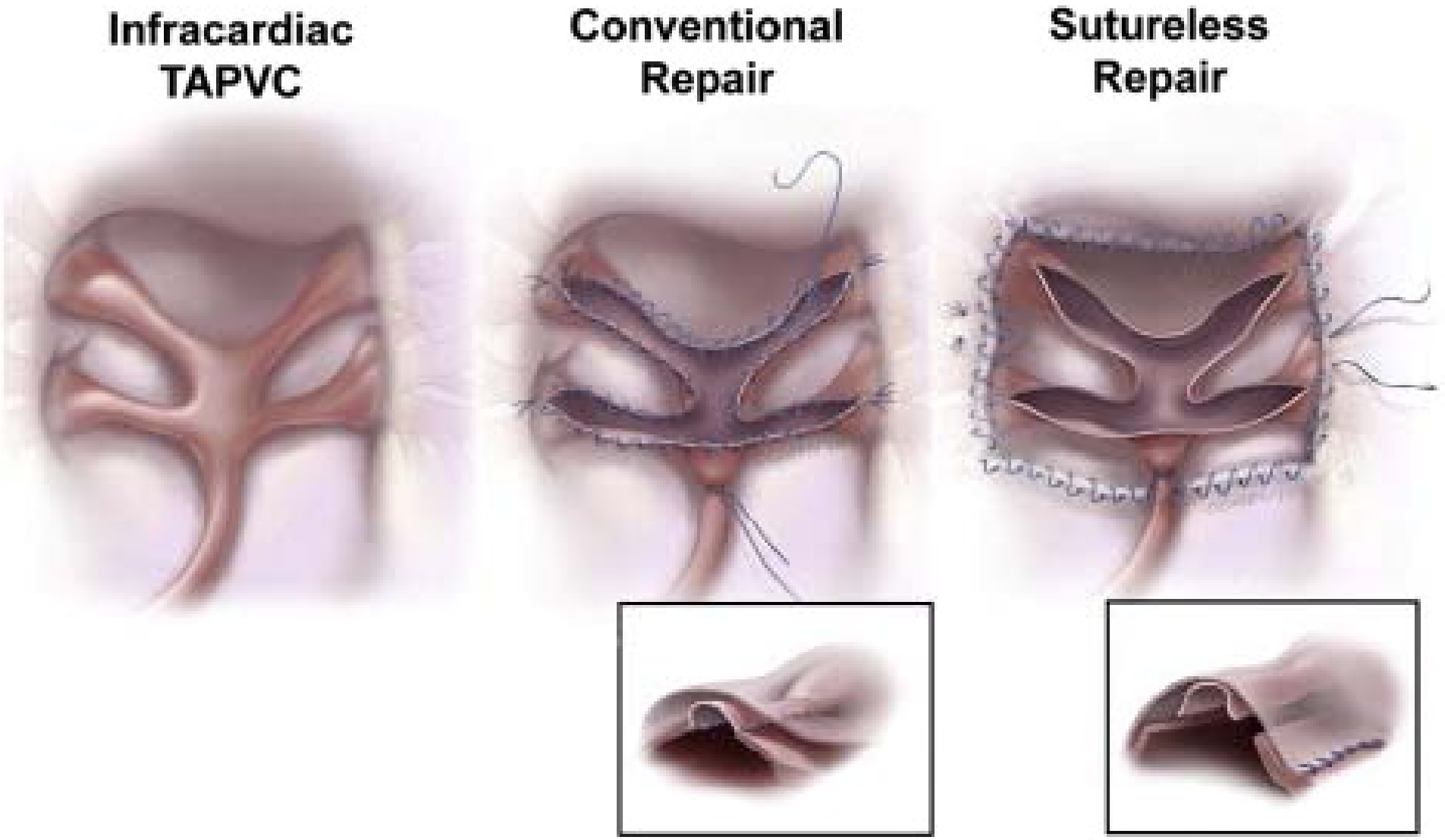
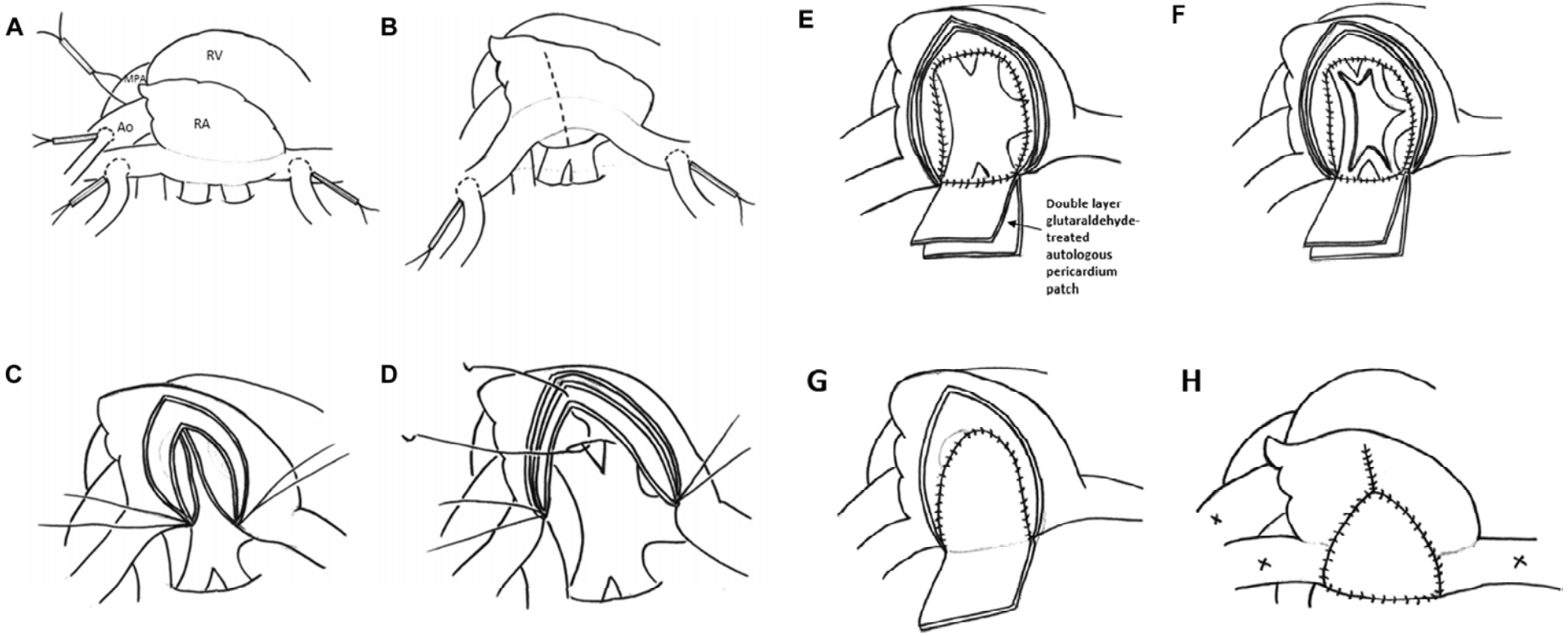


Primary sutureless repair



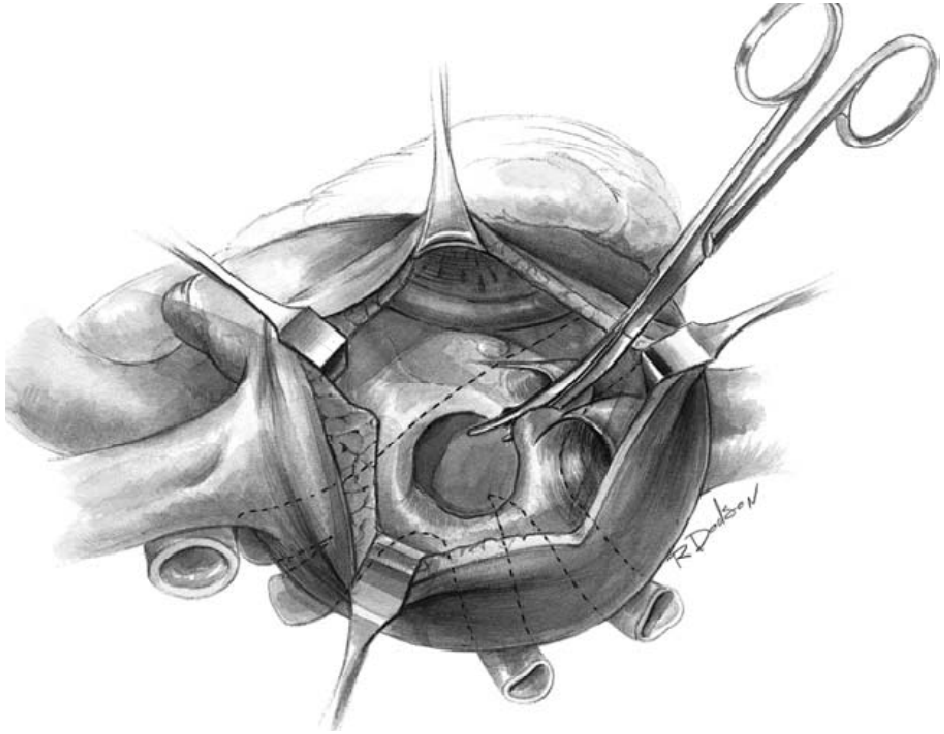
Primary sutureless repair

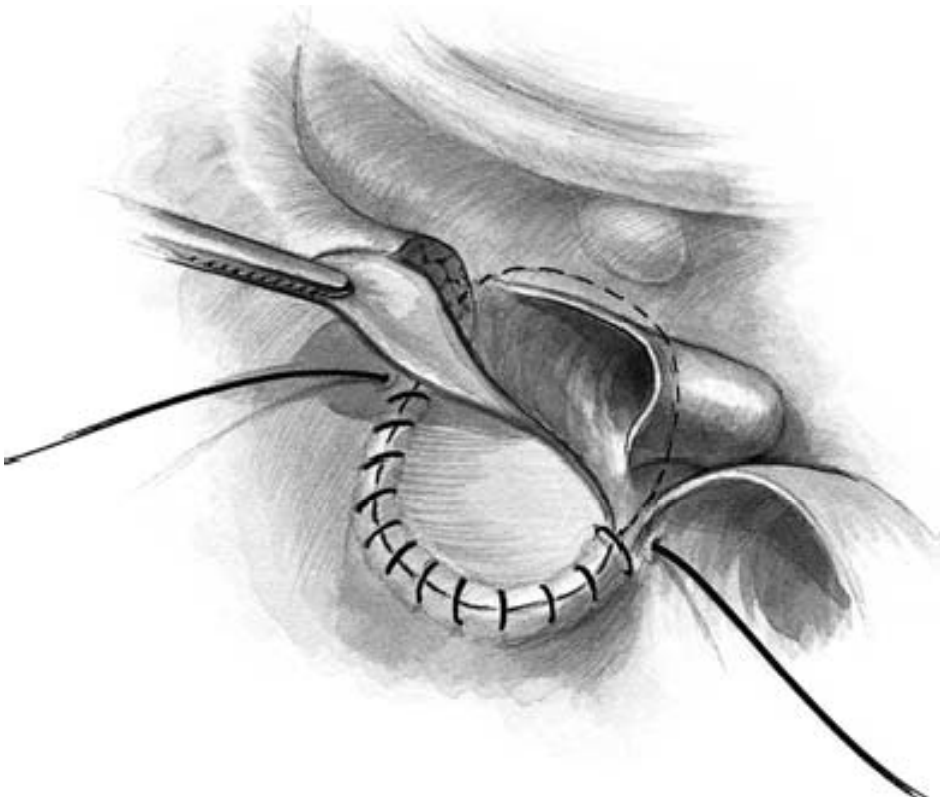
Suture and open technique

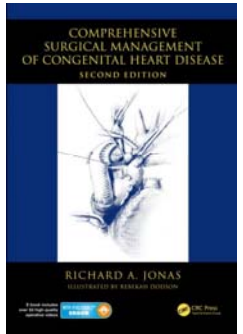


Ann Thorac Surg 2018;106:e273–6

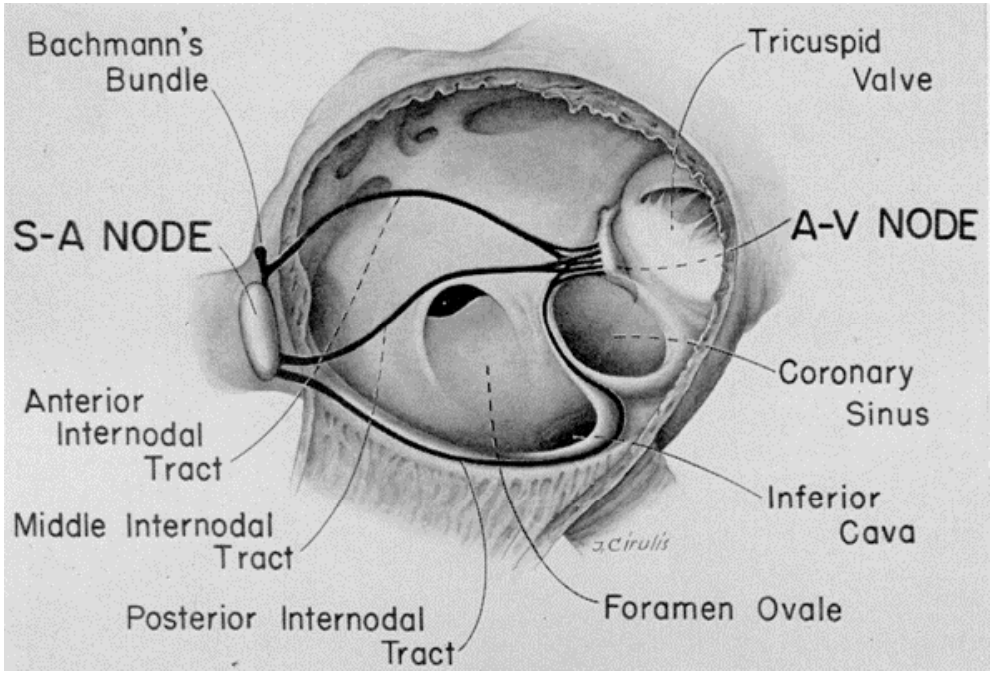
Cardiac type repair

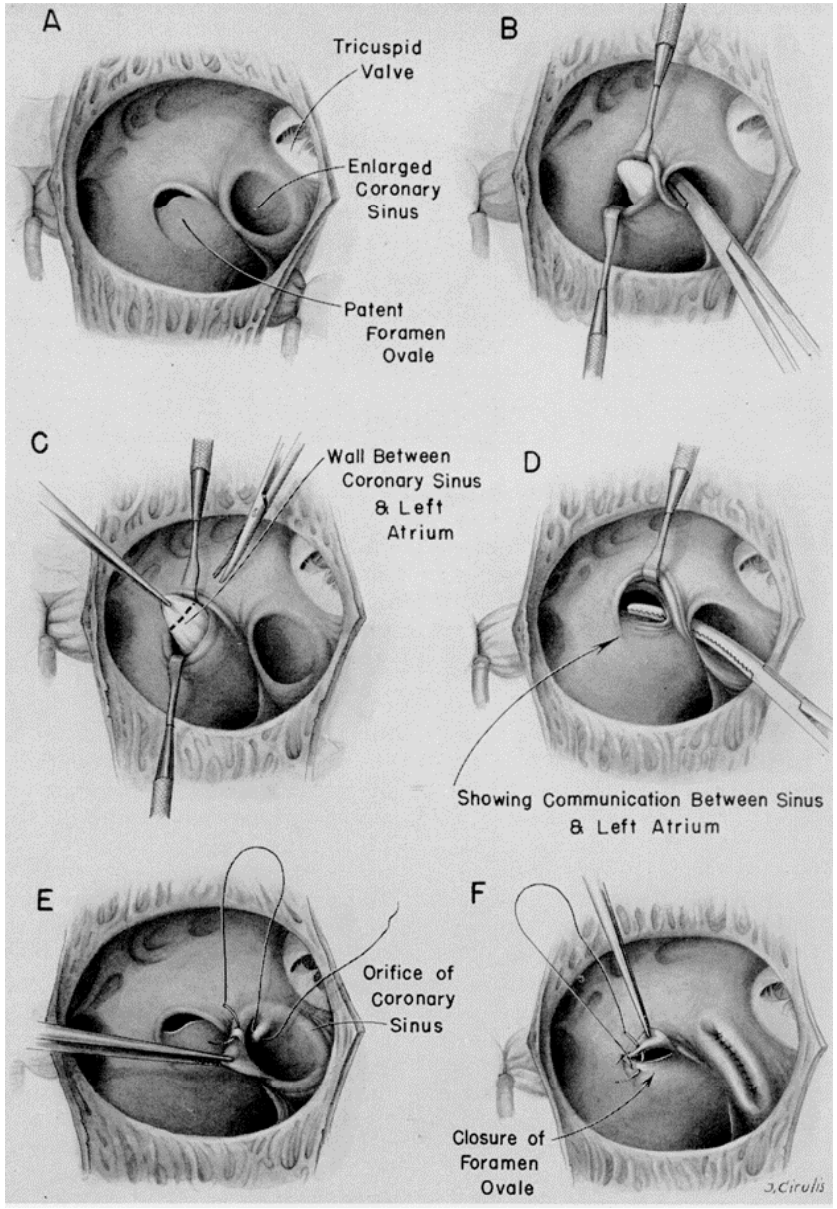






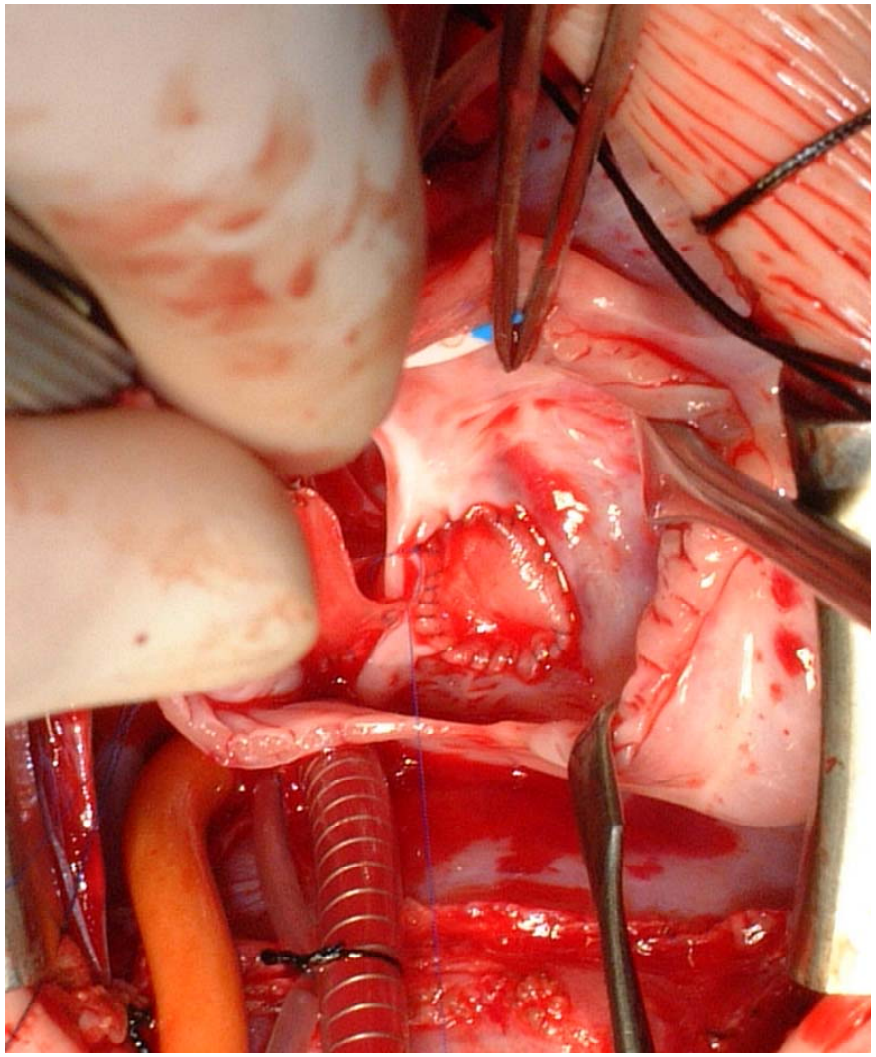
Cardiac type repair





Cardiac type repair

- TAPVR repair
- Separate patch closure



Postop. management

- Consideration of muscularized pulmonary arteries (obstructive TAPVC)
- Minimization of pulmonary resistance
  - Appropriate ventilator care (PCO_2 level)
  - Oxygen, NO gas
  - Low dose isoproterenol (pulmonary vasodilatory effect)
  - Sedation
- Careful observation for pulmonary hypertensive crisis

Prognosis

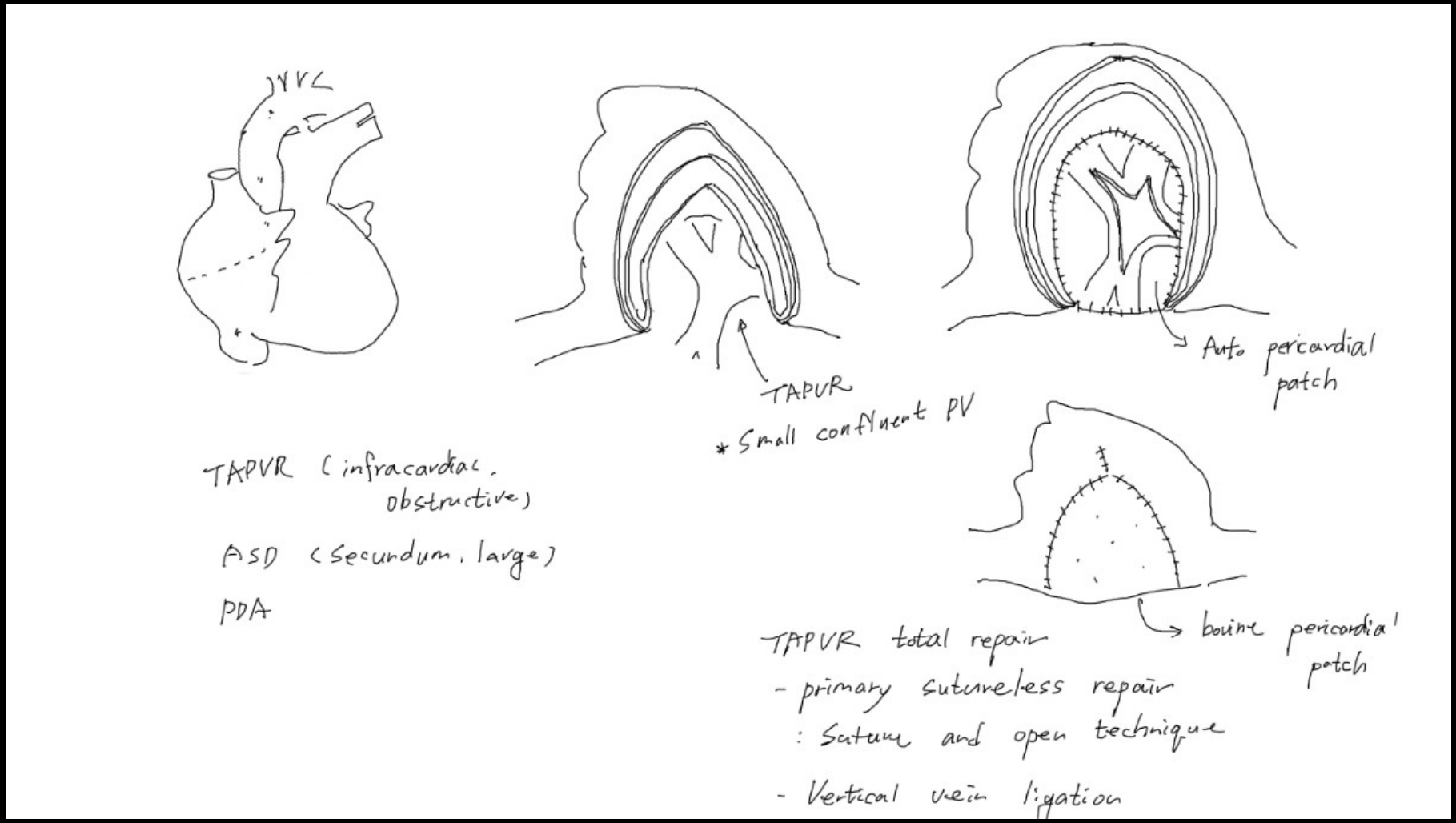
- Early mortality : 8 - 20% ?
- Current operative mortality rates < 5%
- Reoperation rates : 9 - 13 %

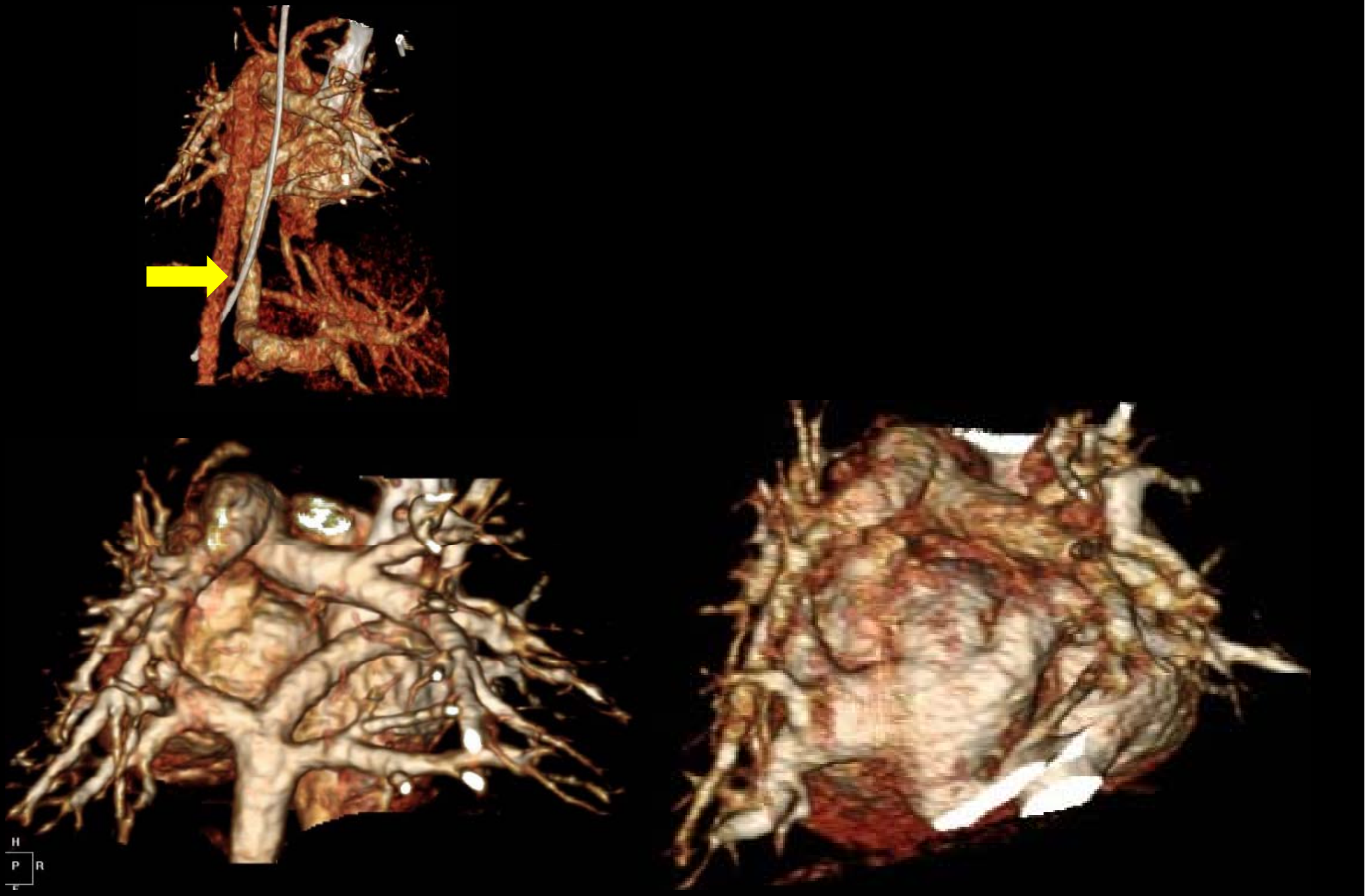
- Risk factors for early mortality
  - Preoperative pulmonary venous obstruction
  - Single ventricle anatomy
  - Chromosomal anomaly
  - Small pulmonary confluence
  - Diffuse pulmonary vein

- Pulmonary vein stenosis : 10-20%
  - Presence of preoperative obstruction
  - Endocardial sclerosis (recurrent obstruction)

1day/2.7kg/F

TAPVR (infracardiac type)





Preop.

Postop.

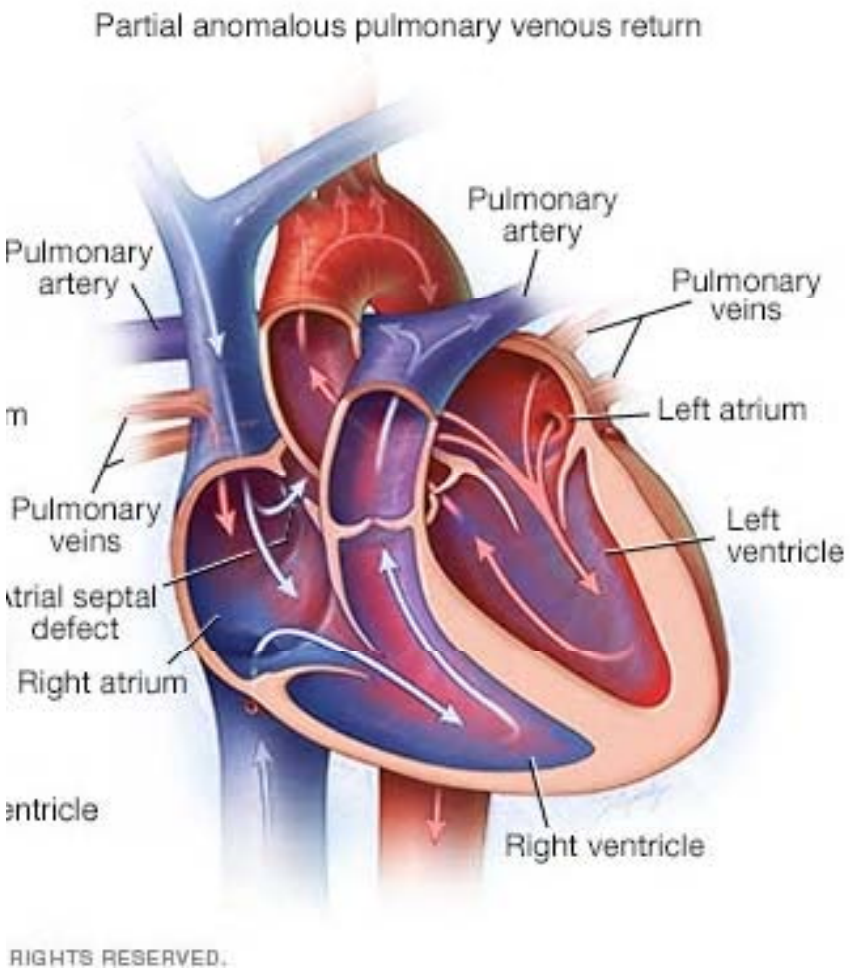
Preop.

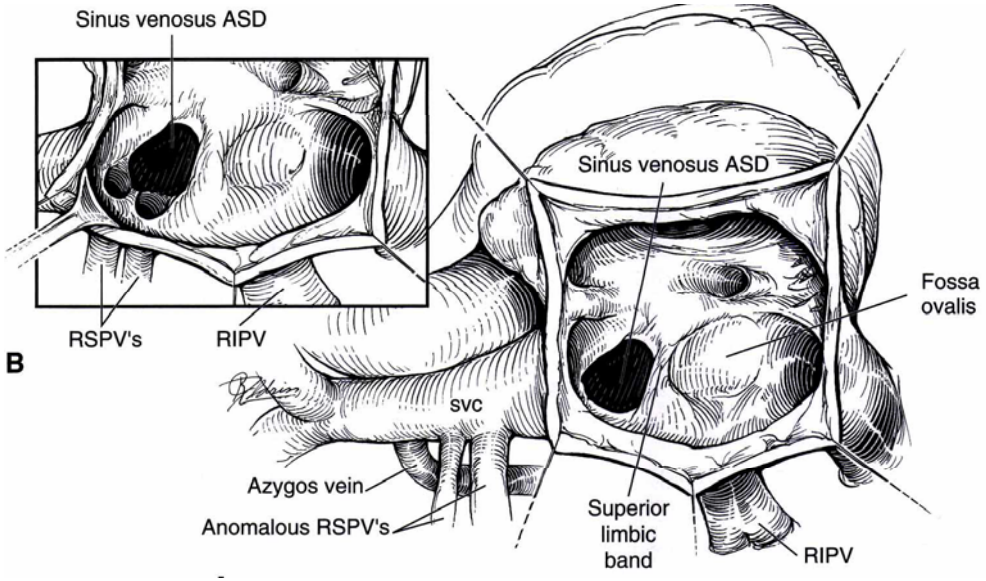
Postop.

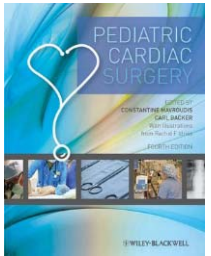
Partial anomalous pulmonary venous return

- M/C associated with sinus venosus ASD
- Usually, RUL draining to SVC
- Rare anomaly in right pulmonary veins
  - Single vertical trunk descends in a curve to enter the IVC (Scimitar syndrome)
- No symptoms and signs when Qp/Qs is less than 1.5

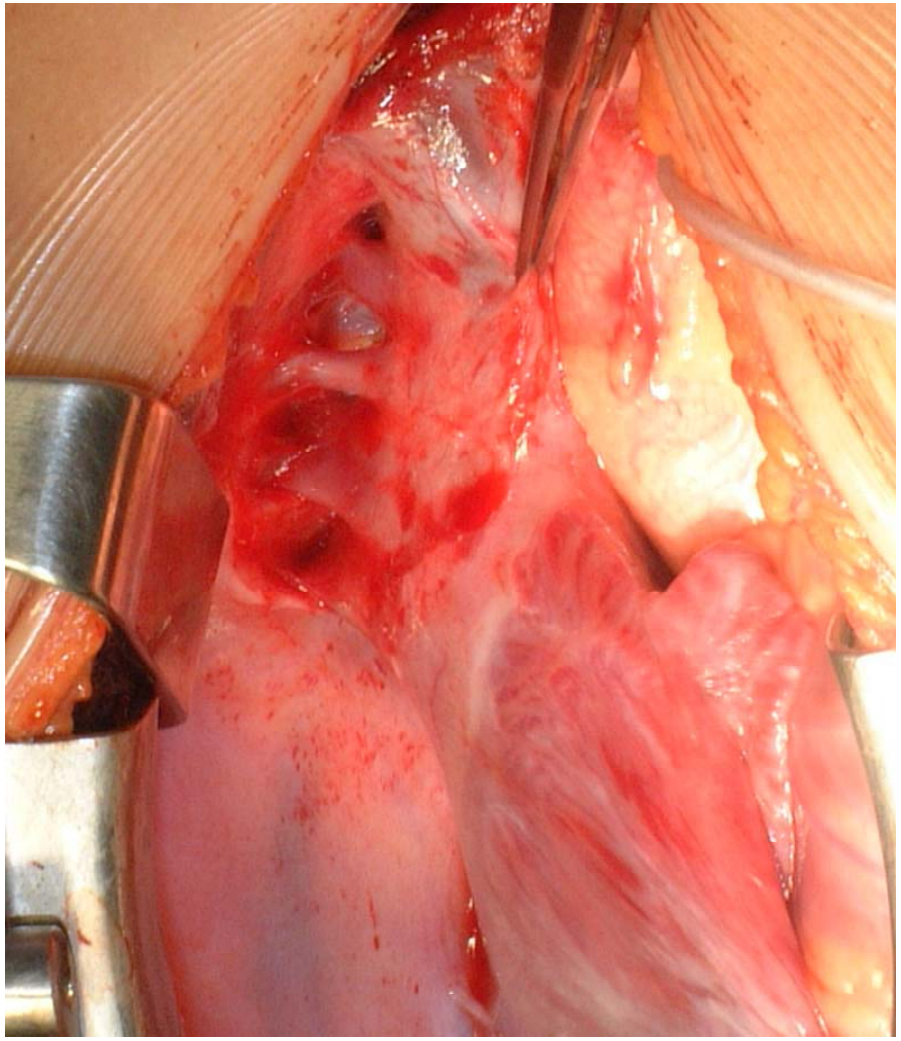
PAPVR

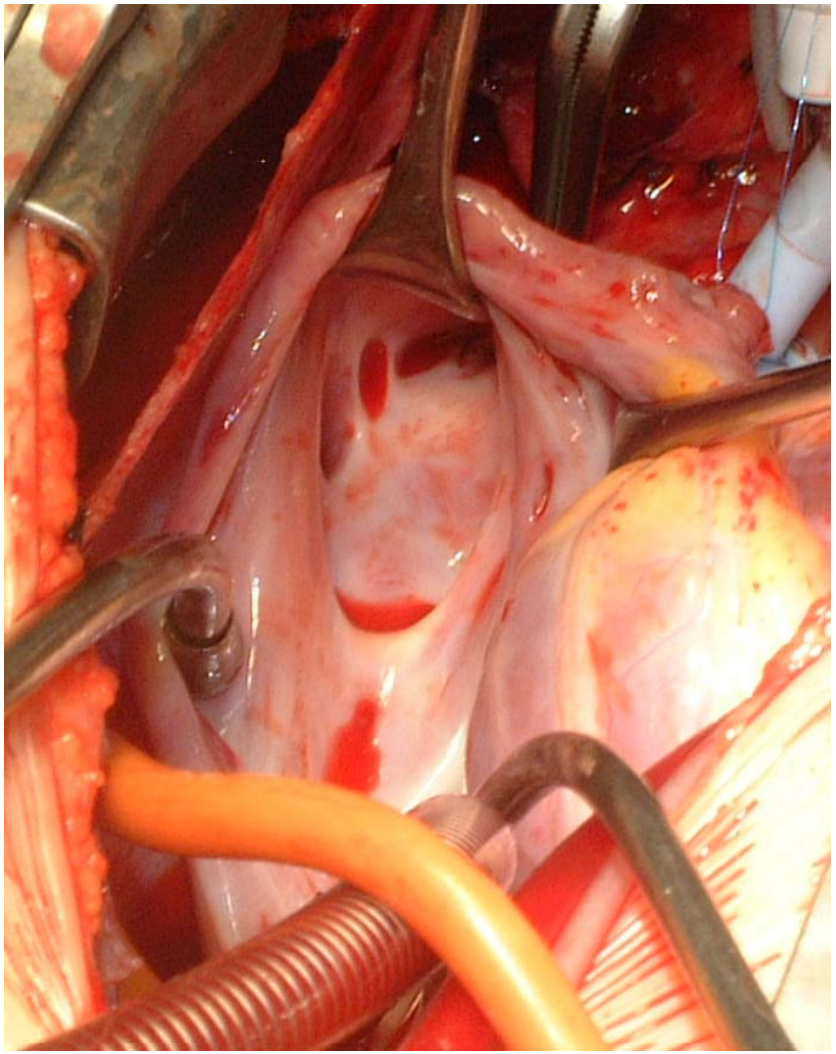

Partial anomalous pulmonary venous return
Pulmonary artery
Pulmonary artery
Pulmonary veins
Left atrium
Pulmonary veins
Left ventricle
trial septal defect
Right atrium
ntricle
Right ventricle
RIGHTS RESERVED.


Sinus venosus ASD
RSPV's
RIPV
B
Sinus venosus ASD
Fossa ovalis
SVC
Azygos vein
Anomalous RSPV's
Superior limbic band
RIPV

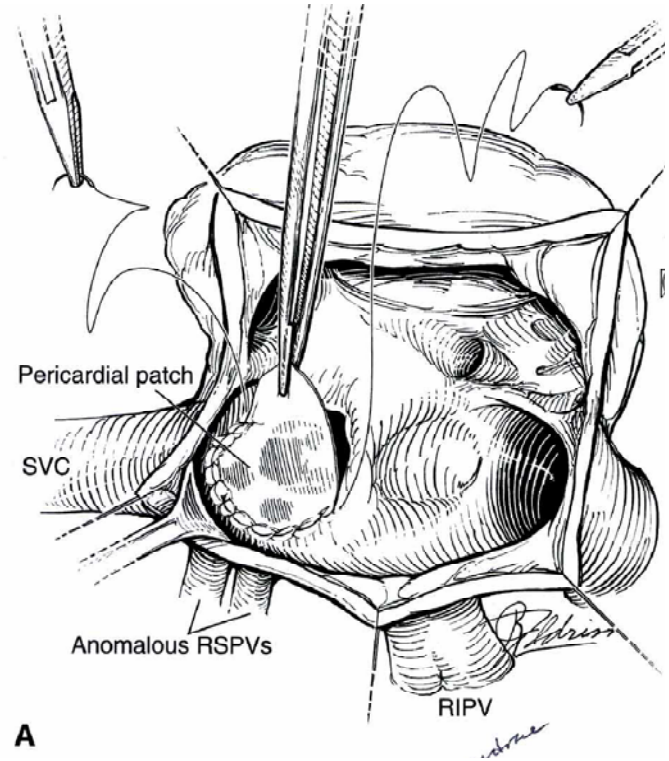

PEDIATRIC CARDIAC SURGERY

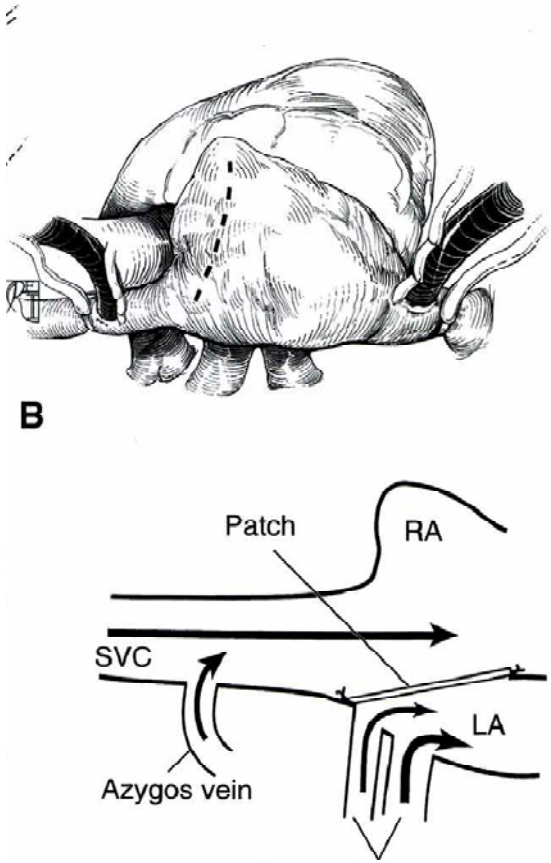
PAPVR

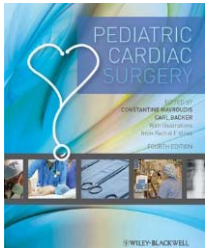




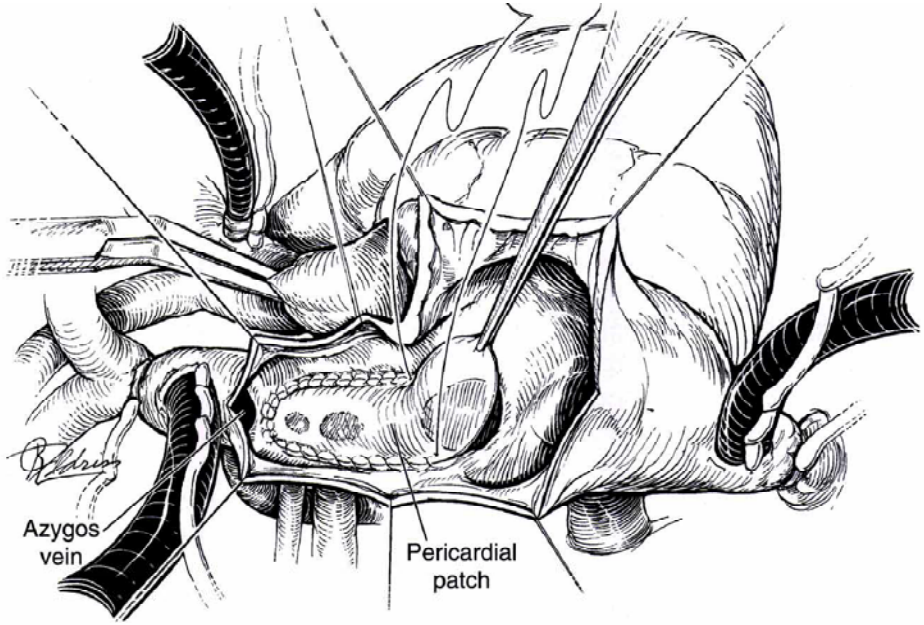
PAPVR repair

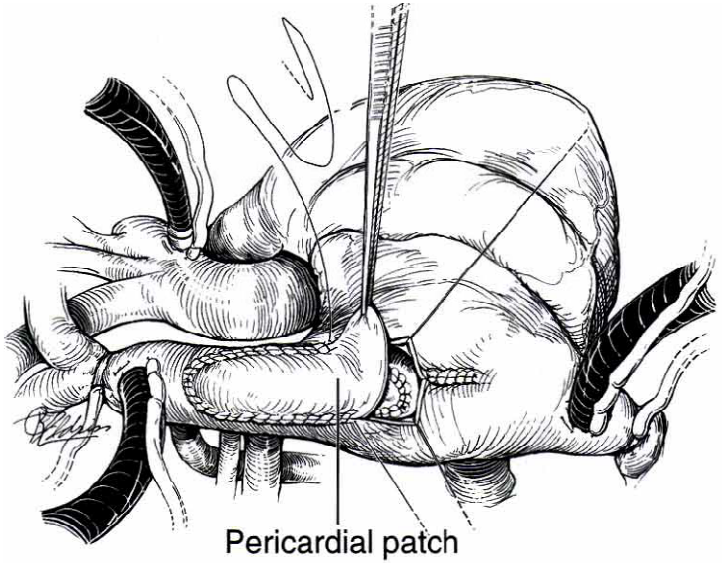


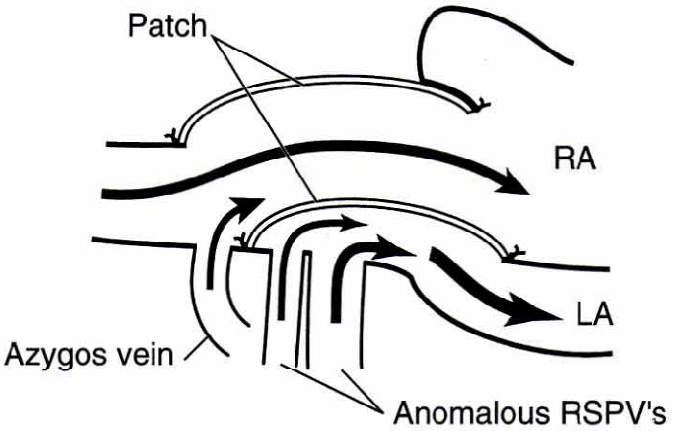


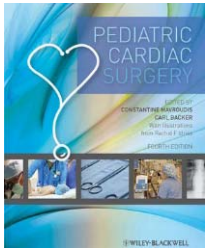


PAPVR repair

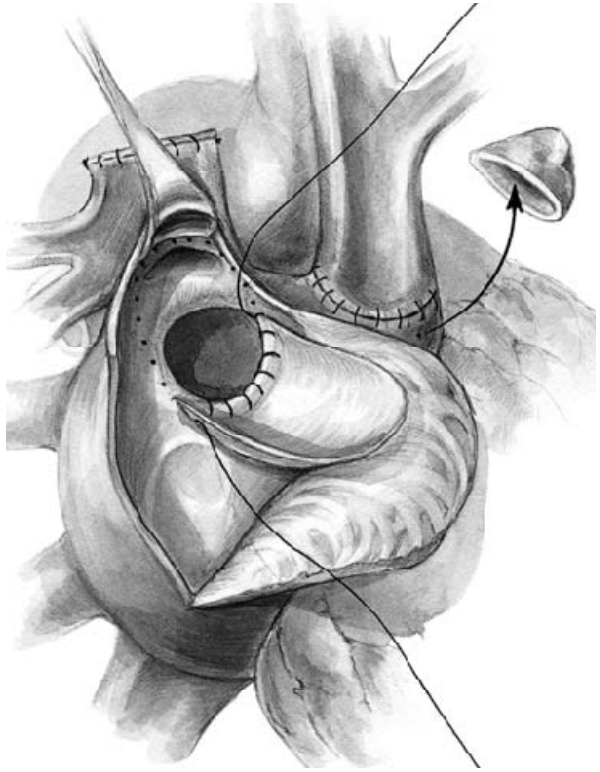


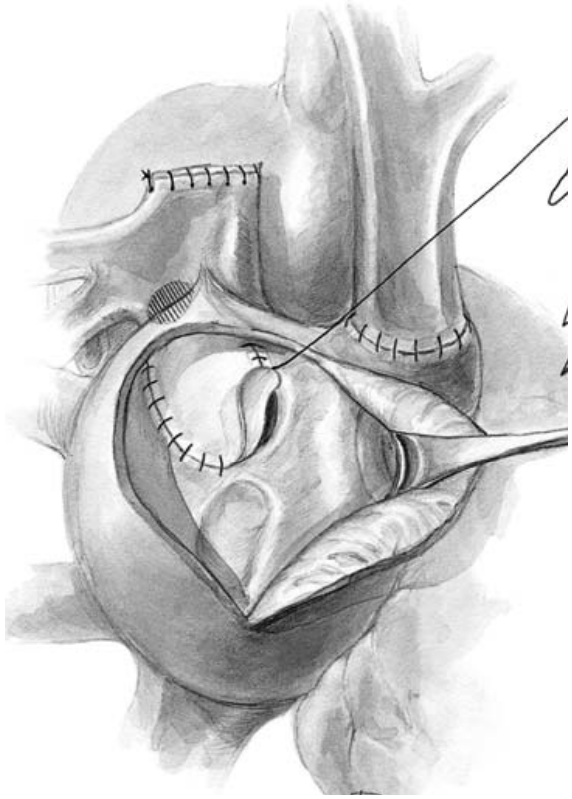


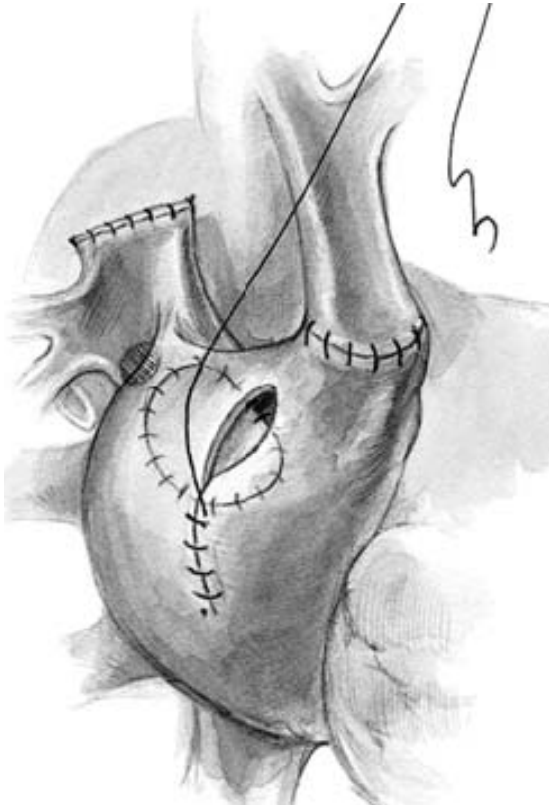


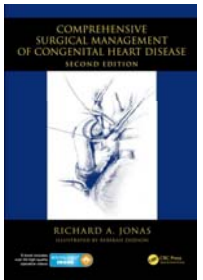


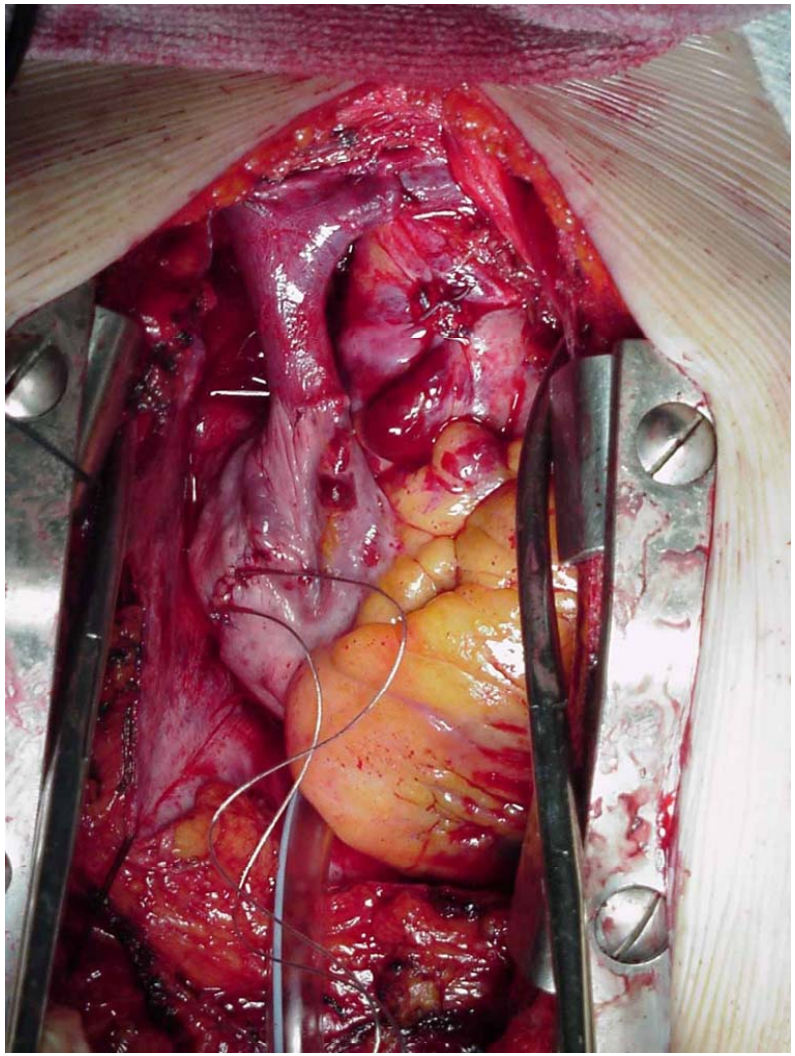
Warden procedure

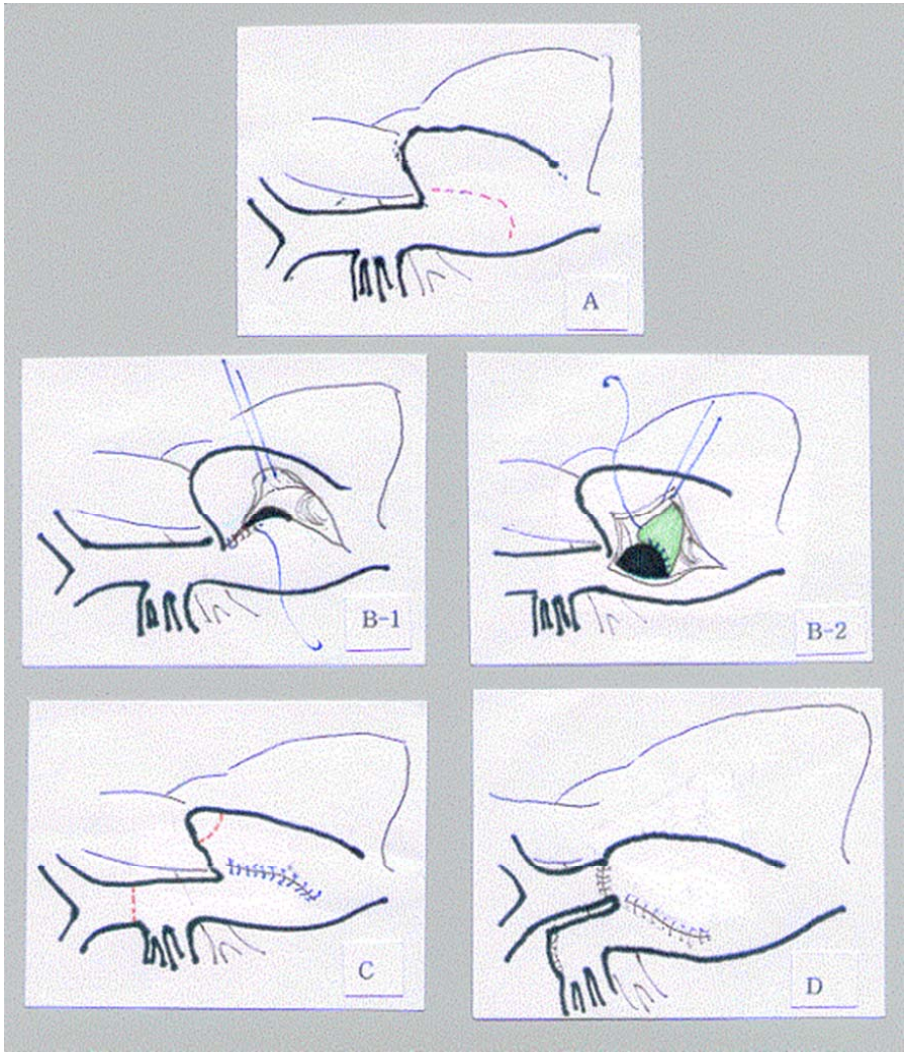








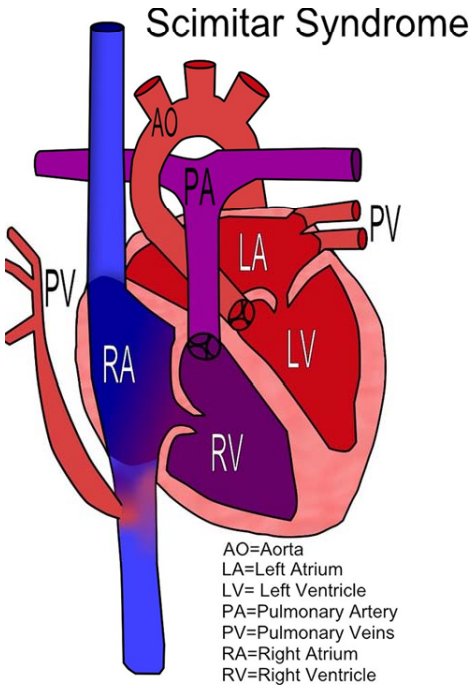


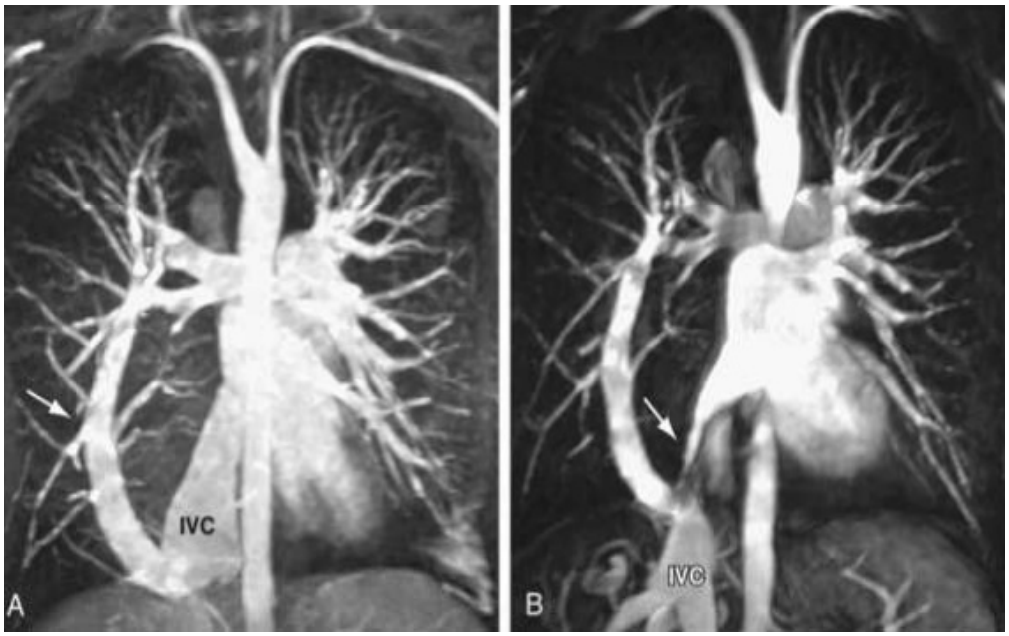

A
B-1
B-2
C
D

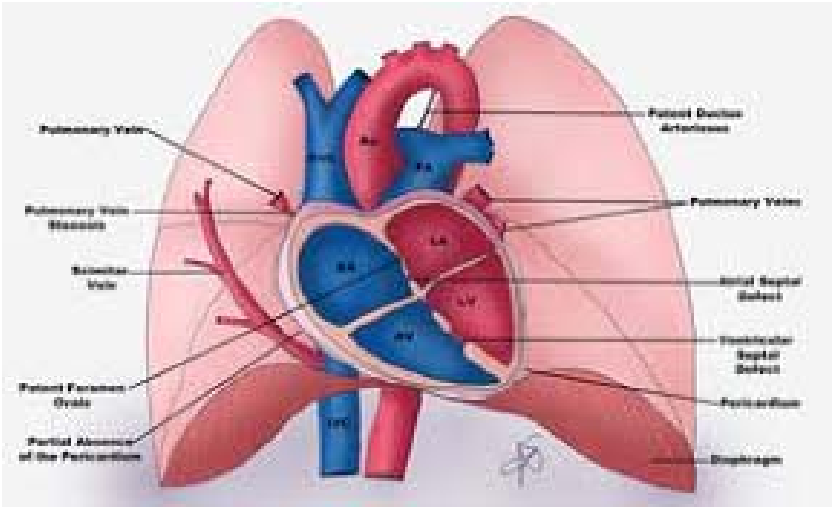
Possible complications

- SVC obstruction
- Pulmonary venous stenosis
- Sinus node dysfunction
- Sick sinus syndrome

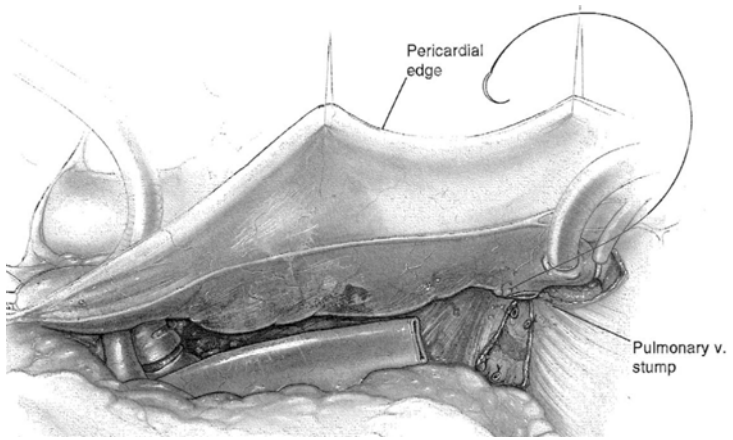
Scimitar syndrome

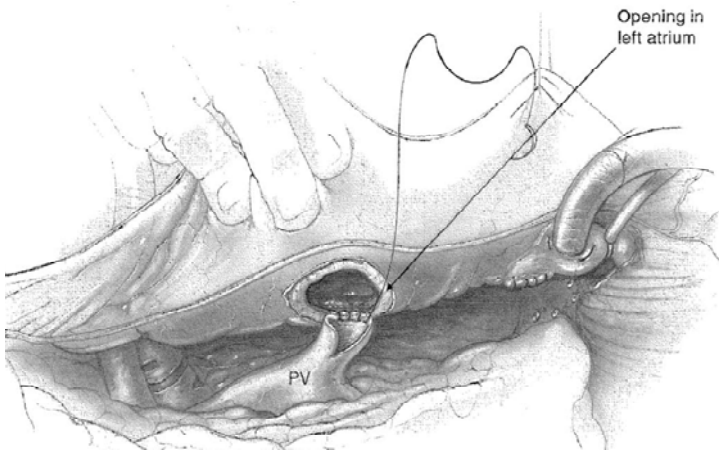


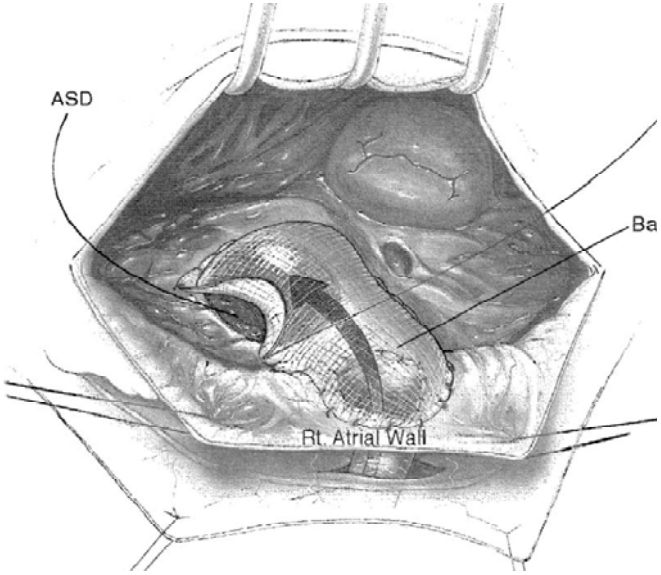


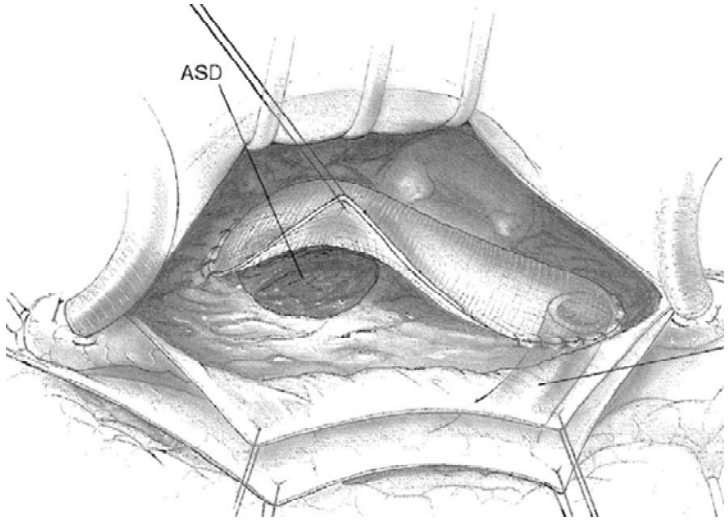


Repair of scimitar syndrome



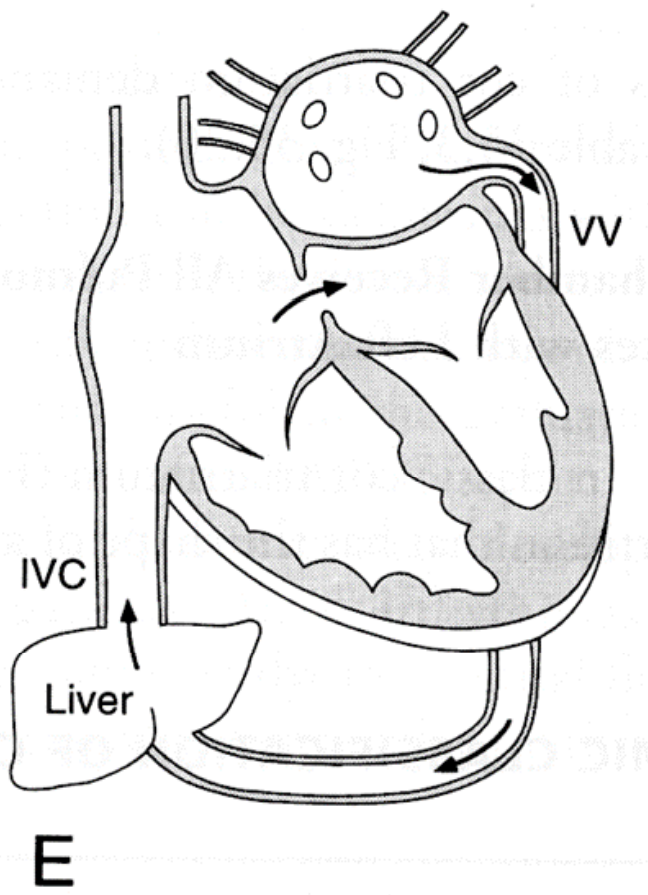






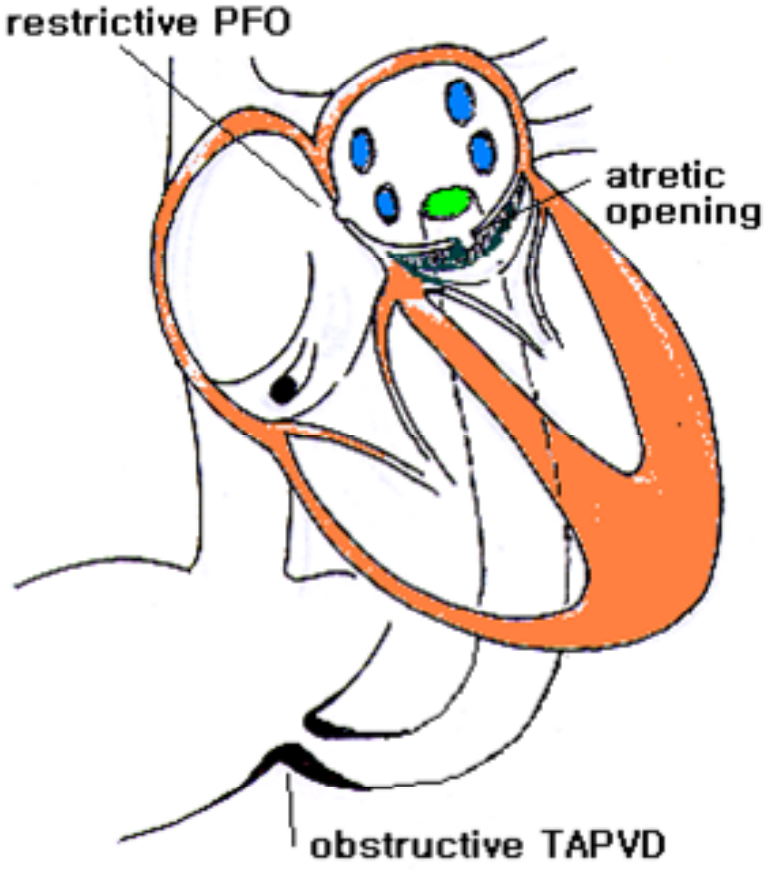
Cor Triatriatum

1. Communication with LA
   - No other connection
   - Other connection
     1. To RA
     2. TAPVC
2. No communication with LA
   - Connection to RA
   - TAPVC



E

Cor Triatriatum



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