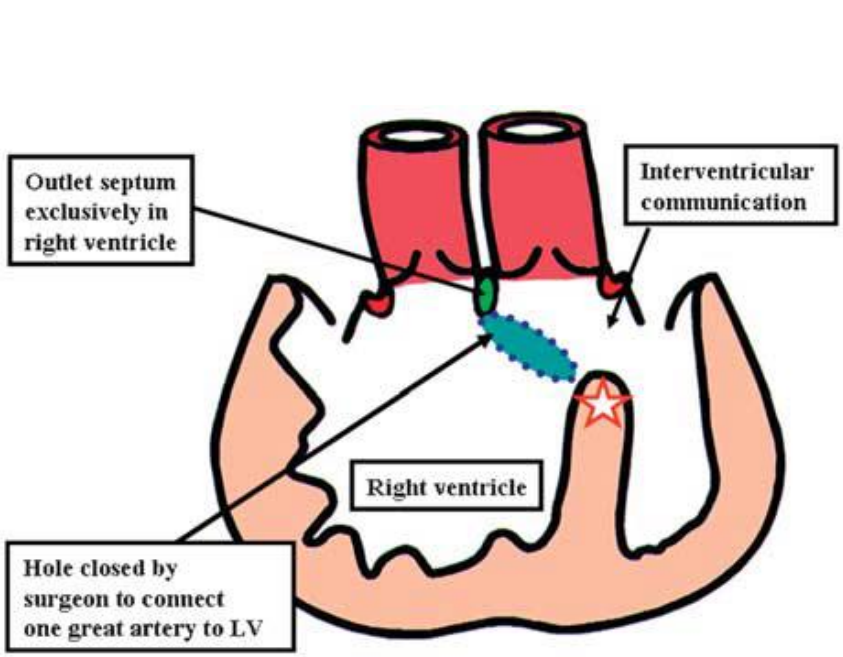
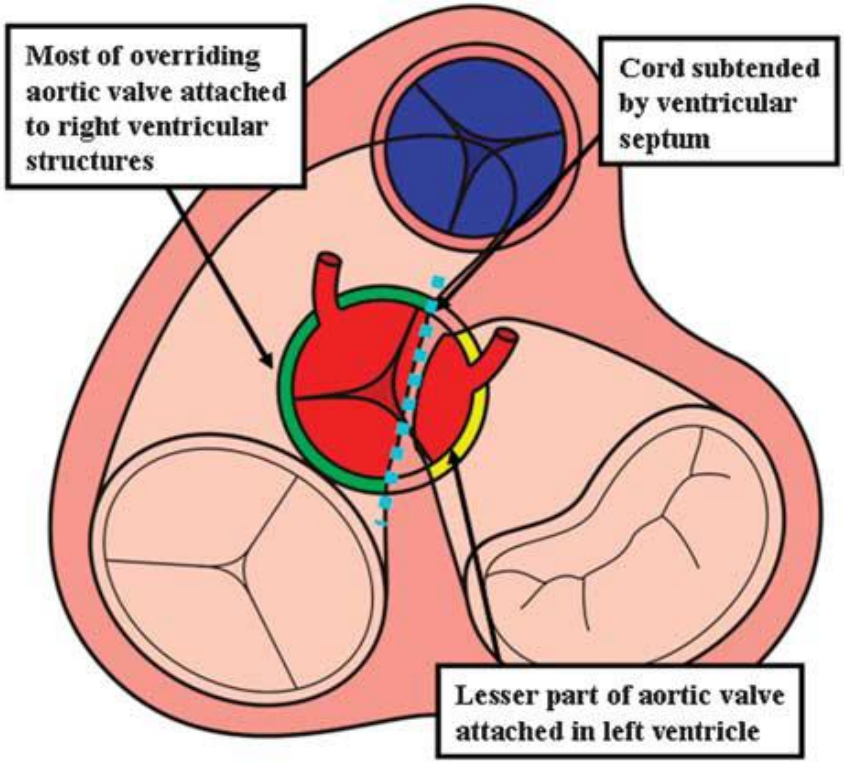
Double Outlet Right Ventricle (DORV)

조성규

Double Outlet Right Ventricle

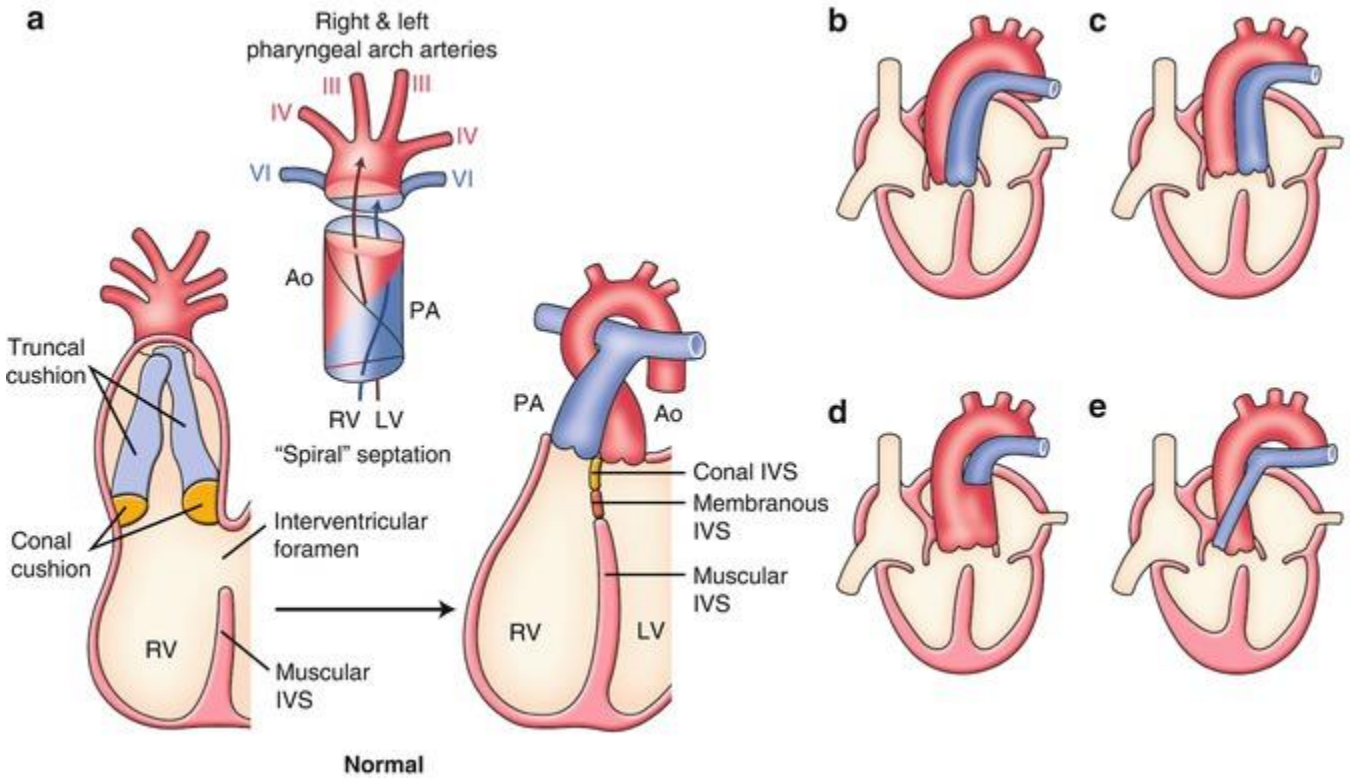
- Definition
  - Type of ventriculoarterial connection in which both great vessels arise either entirely or predominantly from the right ventricle
  - 50% rule(At least 50% each GA's from RV)
  - 200% rule(Both the GA's completely from RV)
- Controversy
  - Aorto-mital continuity
  - Double aortic coni





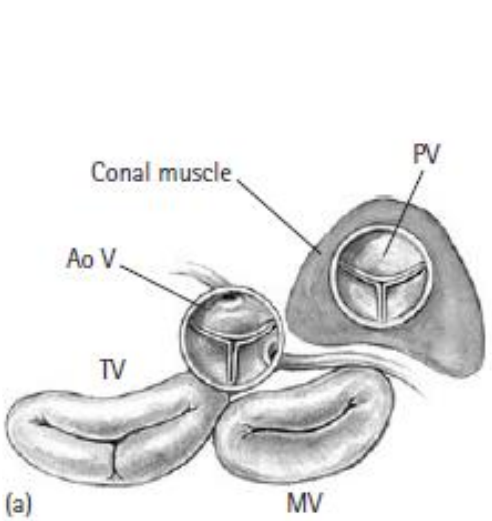
Mahle WT et al, Cardiol Young 2008;18:39-51

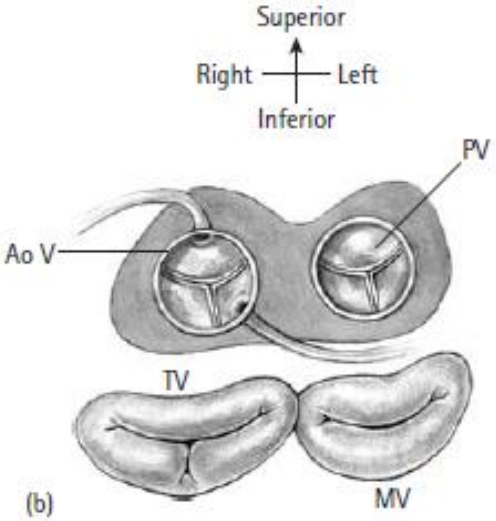
Classic Theory of Conotrucal Malseptation

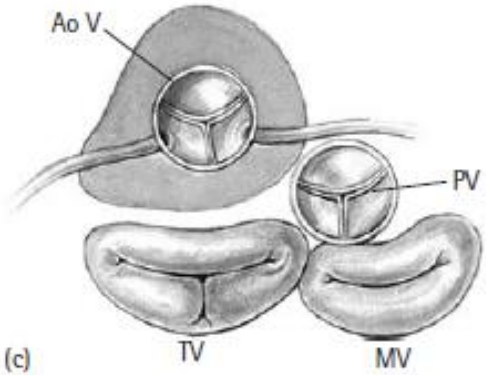


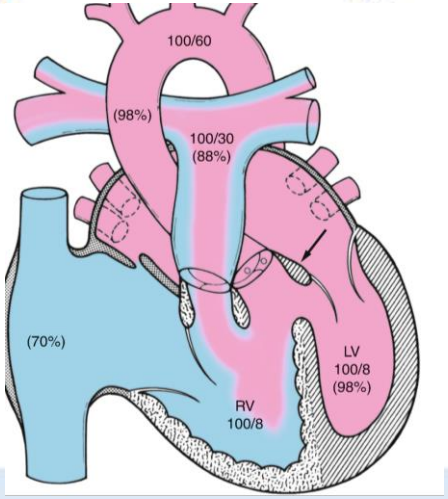
H Yamagish, Cardiac CT and MR for Adult Congenital Heart Disease p13

Van Praagh's Theory of Conal Underdevelopment









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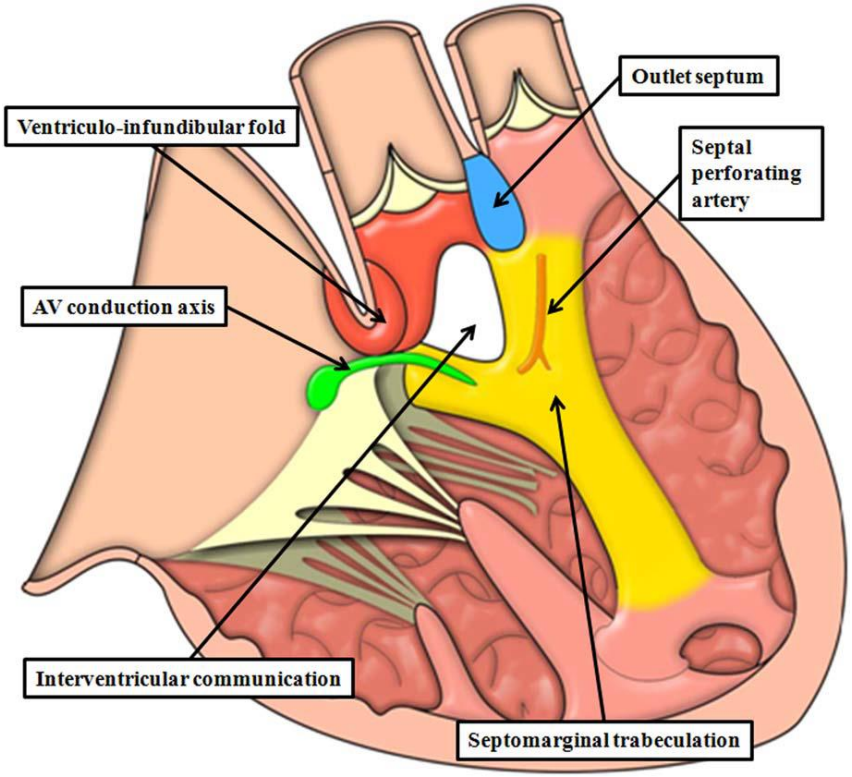
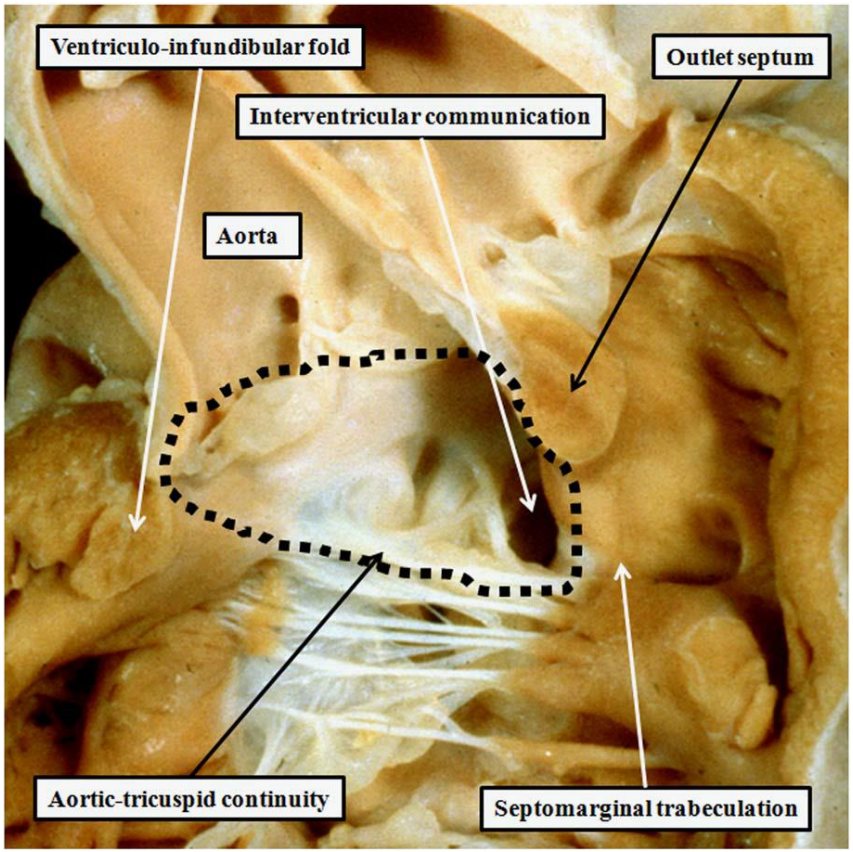
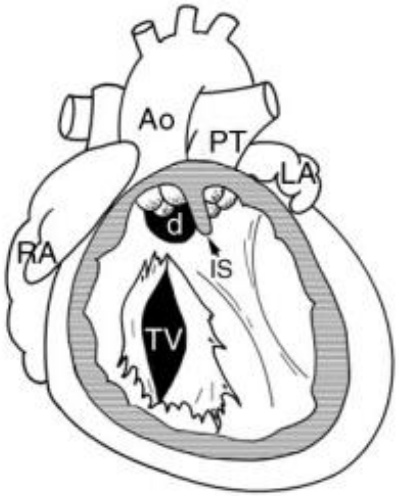




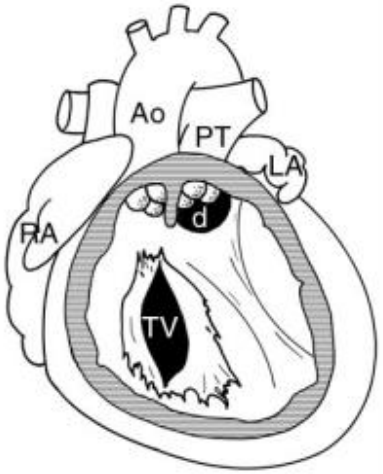
Bharucha T et al, Cardiol Young 2017;27:1-15

Classification



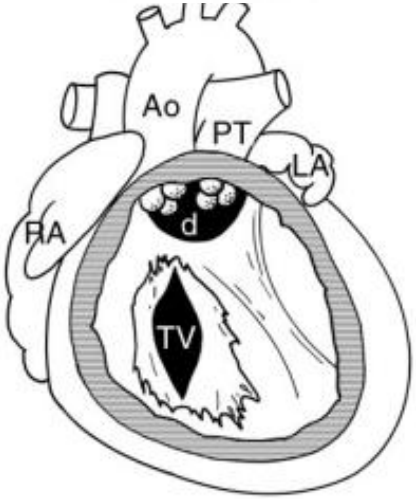
Subaortic VSD

VSD physiology

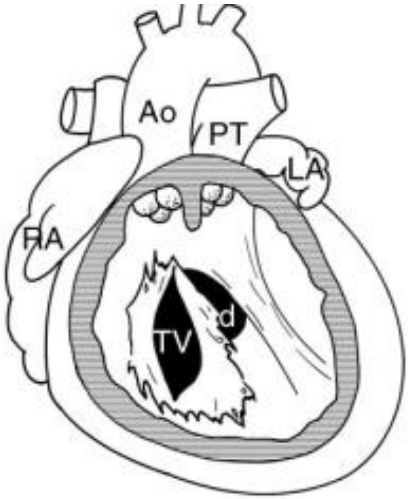


Subpulmonic VSD

C- TGA physiology



Doubly commited juxtaarterial VSD



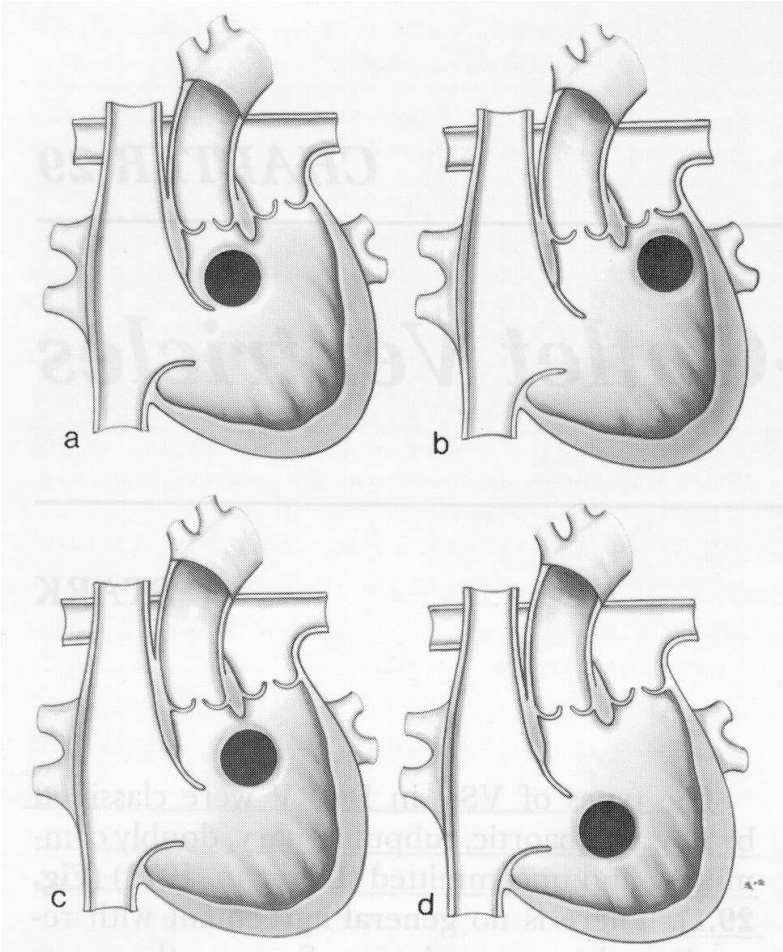
Remote VSD

Lev M et al, J Thorac Cariovasc Surg 64:271-281, 1972

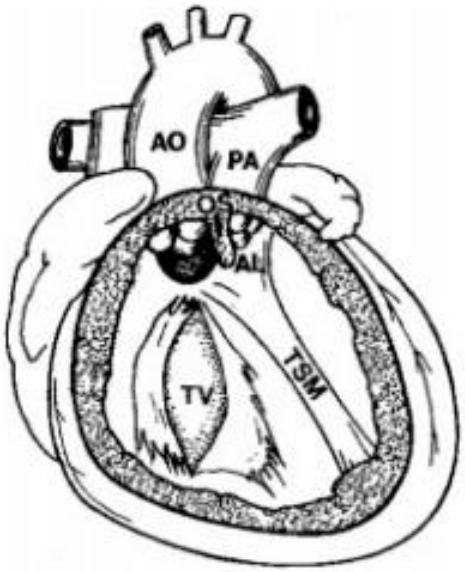
Freedom RM, Yoo SJ Ped Card Sug Ann Semi Thorac Cardiovsc Surg 3.3-19, 2000

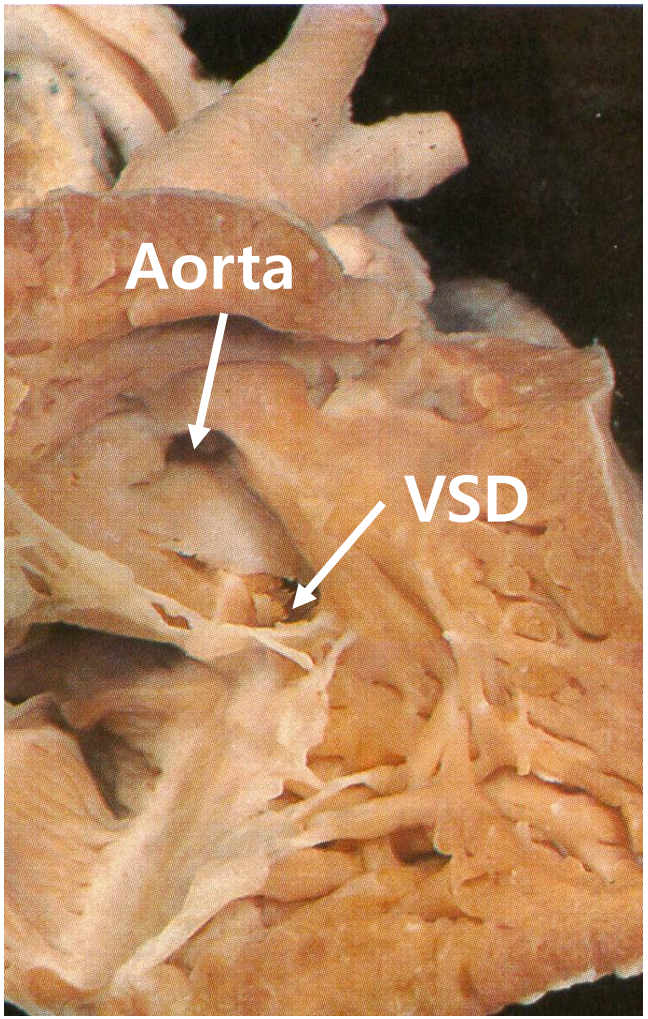
Classification
Locations of VSD in DORV

- a ; Subaortic
- b ; Subpulmonic
- c ; Doubly committed
- d ; Noncommitted

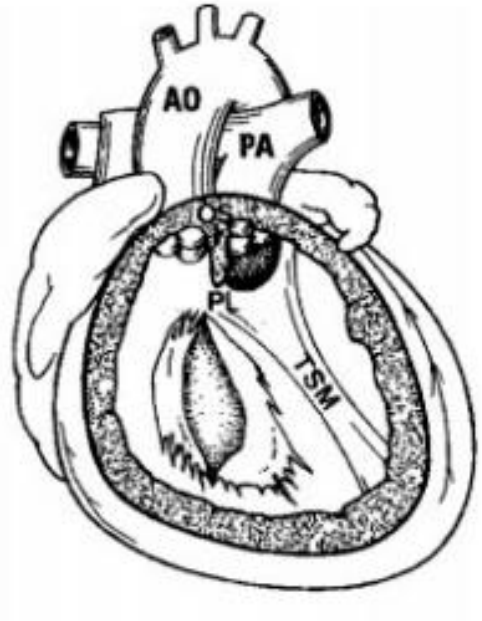


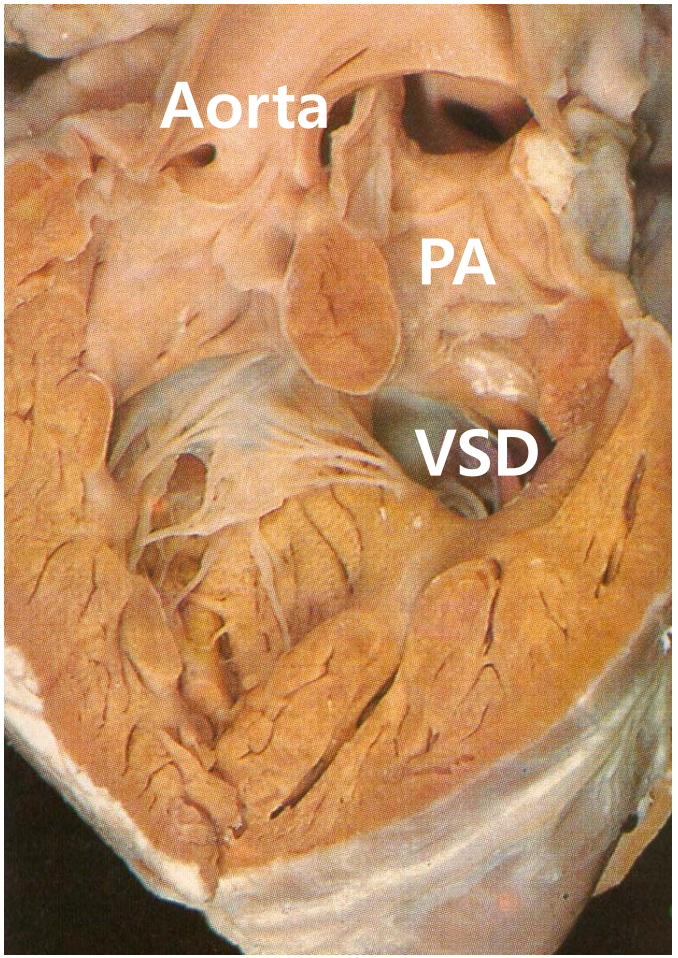
- **DORV with Subaortic VSD**





- **DORV with Subpulmonic VSD**





STS-EACTS-APEC classification

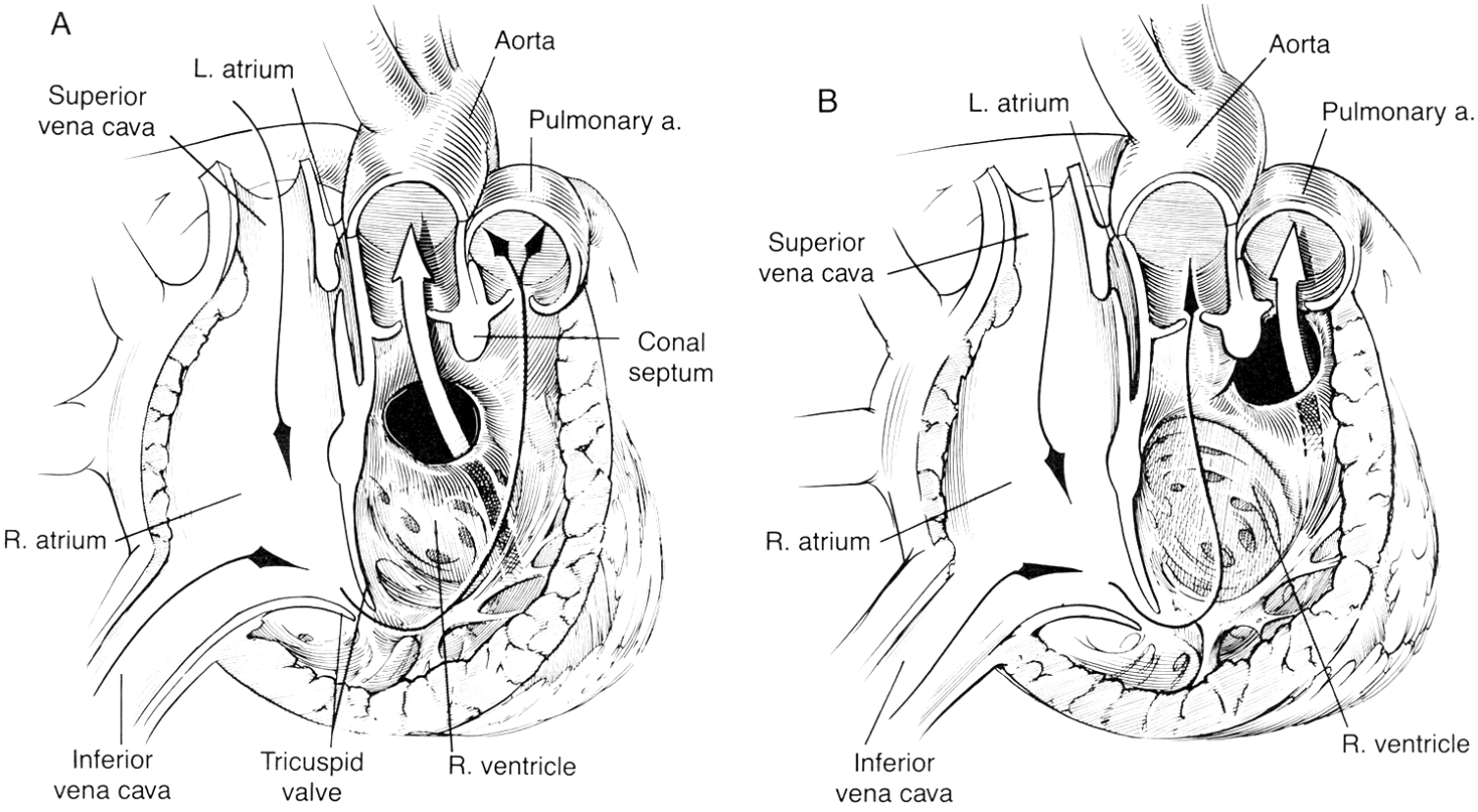
- DORV based on clinical presentation
  - VSD-type : DORV with subaortic or doubly committed VSD
  - Follot-type : DORV with subaortic or doubly committed VSD and RVOTO
  - TGA type (Taussig-Bing) : DORV woth subpulmonary VSD
  - Non-committed VSD-type : DORV with a remote VSD, possible RVOTO

Ann Thorac Surg 2000;69:S249

Anatomical Variables

- VSD location
- Orientation of outlet septum
- Great arterial relationship
- Extent of muscular infundibulum
- Straddling / overriding of AV valves
- Ventricular outflow tract stenosis
  - Subaortic stenosis / aortic valve stenosis
  - Subpulmonic stenosis / PV stenosis
- Aortic arch obstructive lesion

Hemodynamics of DORV



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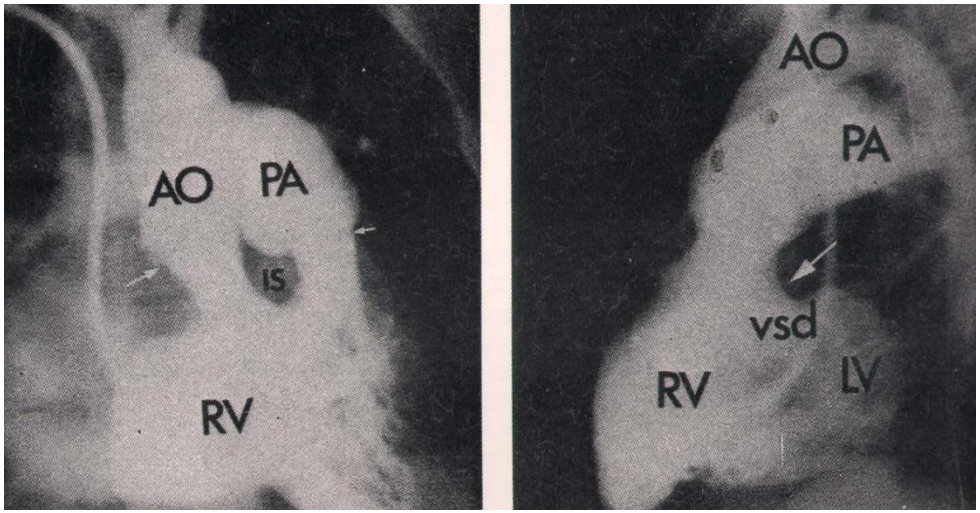
Diagnostic Evaluation

- Echocardiography
  - Single most useful tool
- Cardiac catheterization
  - Degree of pulmonary hypertension
  - Pulmonary resistance
- Computerized tomography

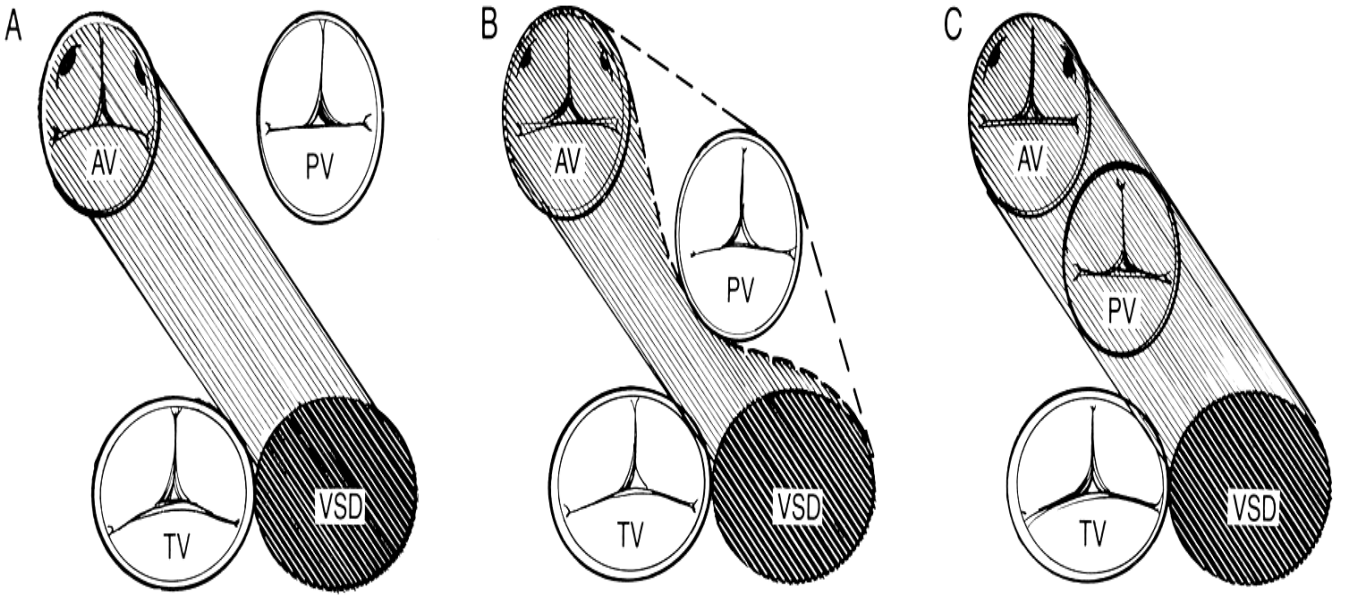


Decision Making for Surgical Treatment

- Determined location and size of the VSD
- Presence and site of pulmonary obstruction
- The degree of aorto-mitral valve separation
- Position of the aortic valve with respect to the pulmonary valve
- Distance between pulmonary and tricuspid valves
- Coronary artery distribution
- Chordal attachments to conal septum

Separation between the TV and PV

- Determining anatomic suitability for an intraventricular baffle repair



Adequate separation

Risk of subaortic stenosis

Rastelli procedure

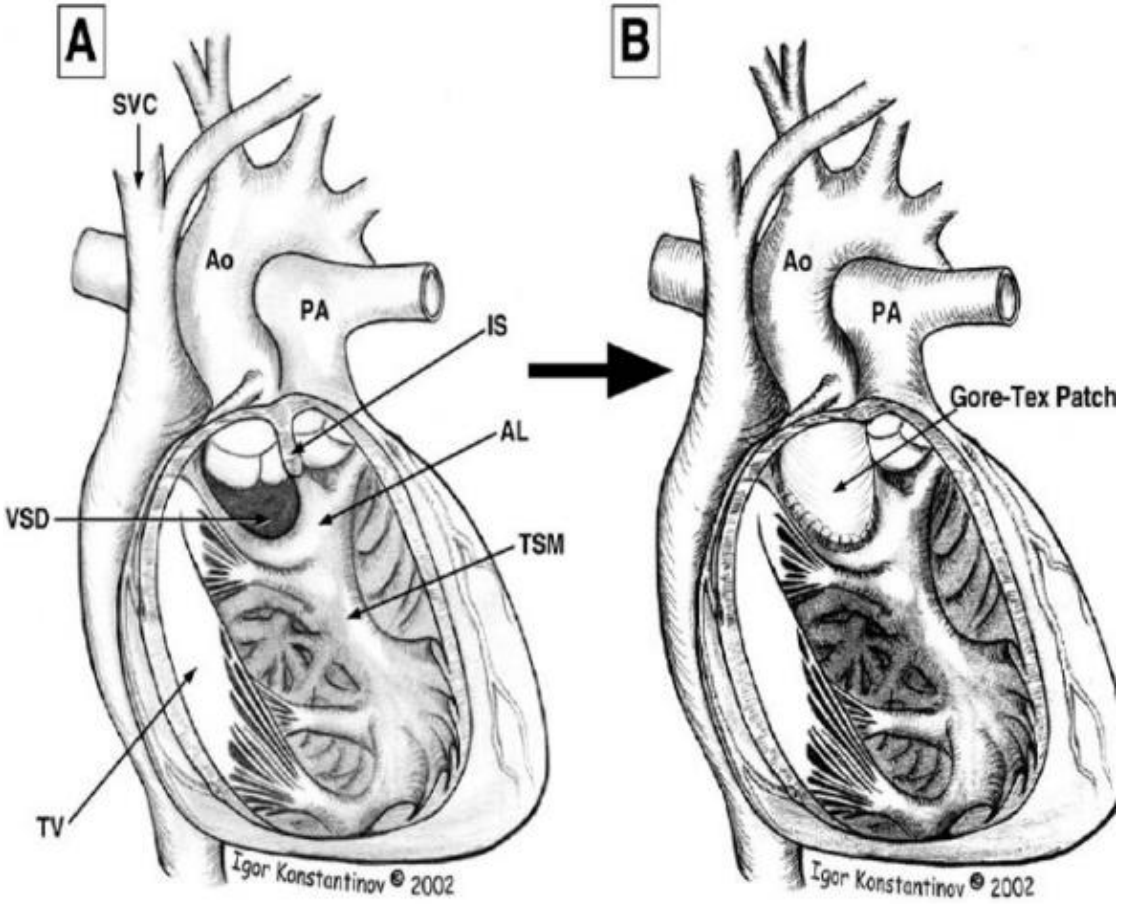
Surgial Techniques

- Intraventricular tunnel/ baffle repair
- Rastelli procedure
- REV procedure
- Aortic translocation (Nikaidoh procedure)
- Double root procedure
- Arterial switch operation

DORV – VSD type

- Subaortic or Doubly-Committed VSD without Pulmonary Stenosis
- Clinical sign of overcirculation
- Pulmonary vascular obstructive disease
- Complete repair in early infancy

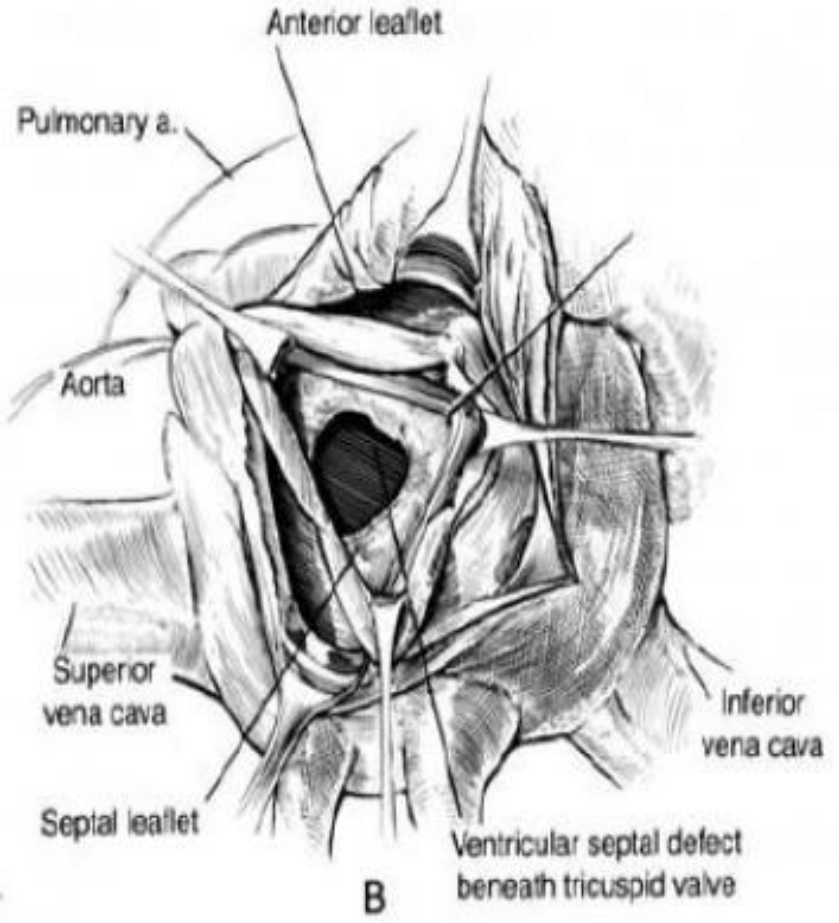
Intraventricular tunnel repair (VSD to aorta)



VSD Baffle to aortic valve

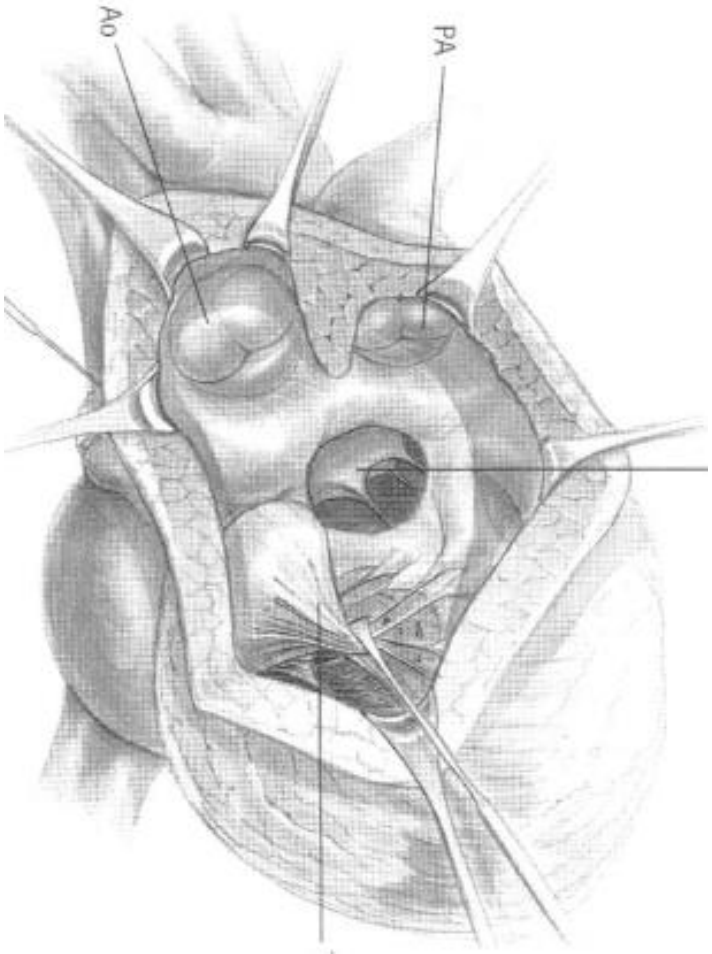
RA approach

- Avoids ventriculotomy
- Looking around a corner
- Difficulty visualizing conal septum / aortic valve



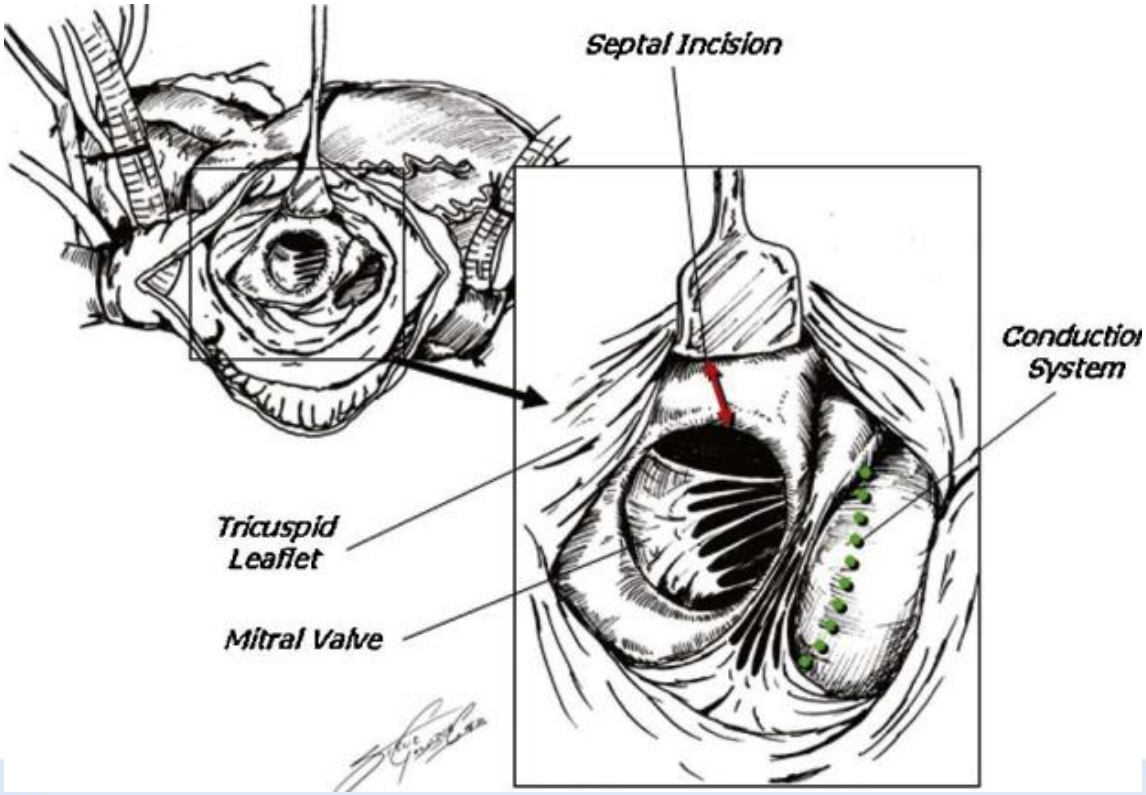
RV approach (RV infundibulum)

- Direct view of LVOT
- Ventriculotomy location critical
  - Conal branch
  - LAD



VSD enlargement

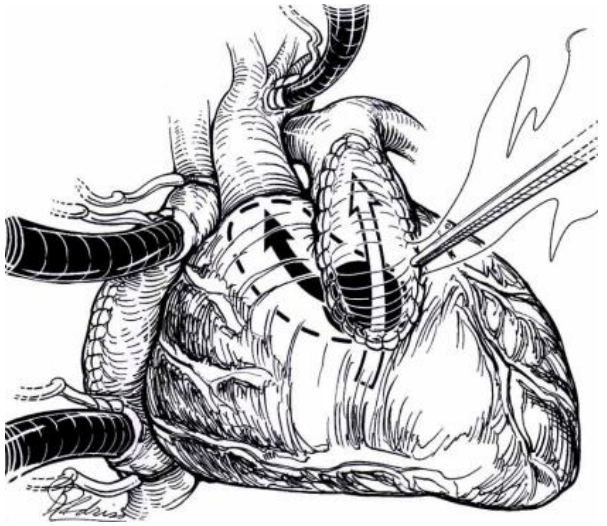
- Restrictive VSD
  - Incision anterior superioly
  - Resecting a wedge of the interventricular septum
- Risk of injury
  - Mitral valve & tensor apparatus
  - Ant. Vent wall & LAD, septal perforator

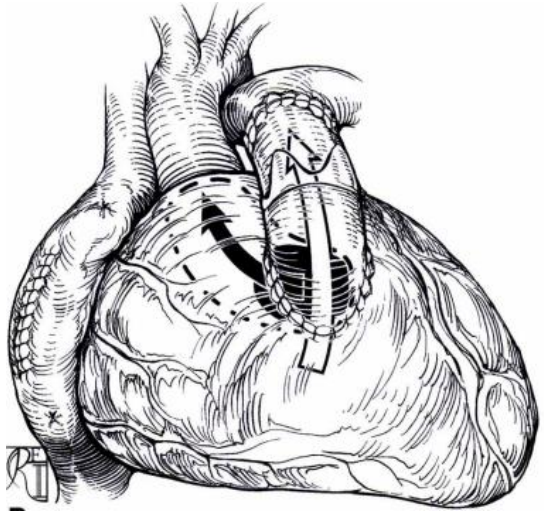


DORV-Fallot type

- Subaortic of Doubly-Committed VSD with Pulmonary Stenosis
- Similar to Tetralogy of Fallot
- RVOTO : Pulmonary stenosis or atresia
- VSD baffle patch closure
- RVOT reconstruction
- If preop. condition is poor : B-T shunt could be an option.

- RVOT reconstruction
  - Non transannular
  - Transannular
  - Division of septal and parietal extension of conal septum
  - Aggressive resection of hypertrophied obstructive muscle bundle
- RV-PA conduit
  - PTFE valved conduit
  - Homograft





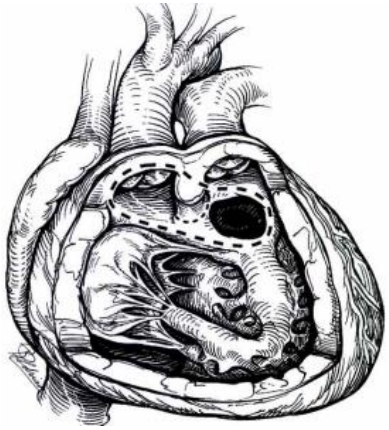
DORV with Subpulmonary VSD (DORV-TGA type, Taussig-Bing)

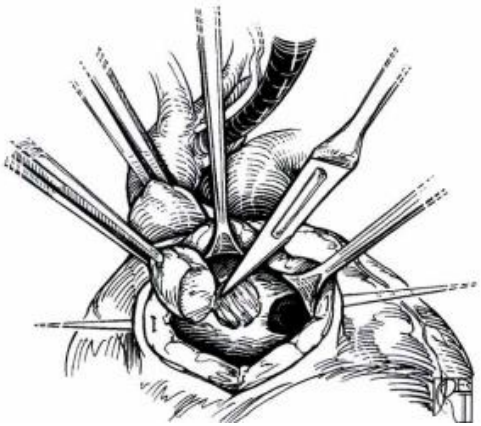
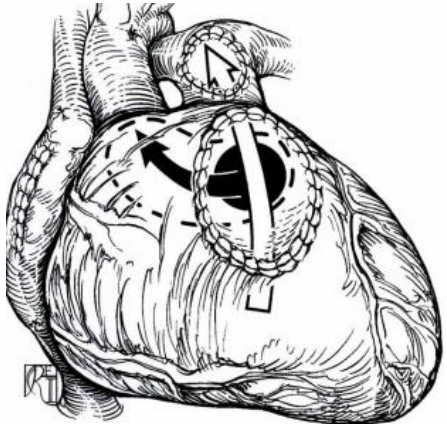
- Neonatal period with cyanosis typical of transposition physiology
- Side-by-Side semilunar valve, Bilateral conus

- Kawashima operation
- Arterial switch operation
- Rastelli / REV operation, intracardiac baffling
- Aortic Translocation (Nikaidoh procedure)

Kawashima operation

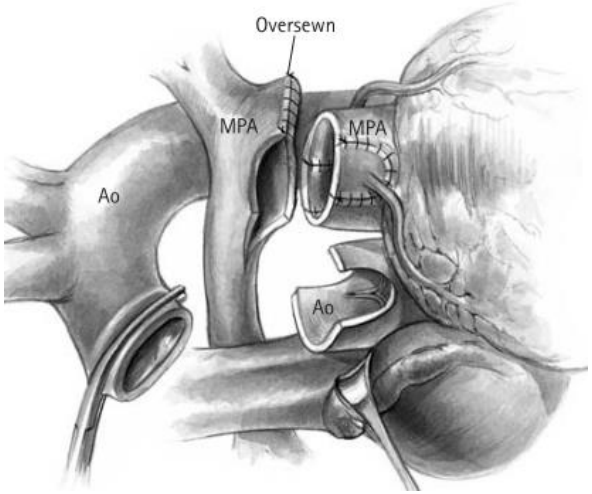
- Side-by-side relationship of great arteries
- Tunneling of Left ventricle directly to the aorta
- Resection of infundibular septum
- VSD enlargement

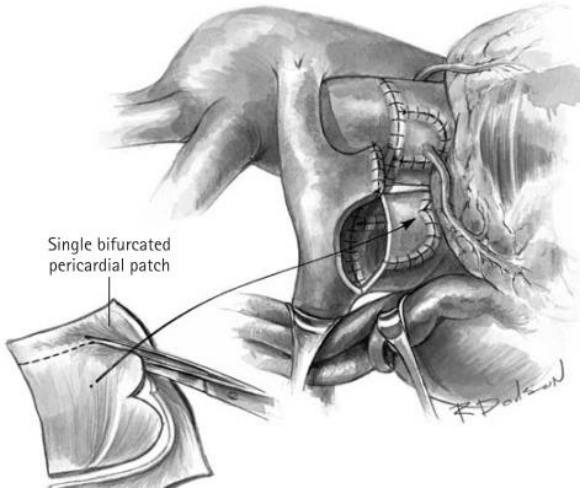


Arterial switch operation with tunnel closure of the VSD

- Frequently great vessel lie side-by-side
- Difficult exposure of VSD
- With or without Lecompte maneuber

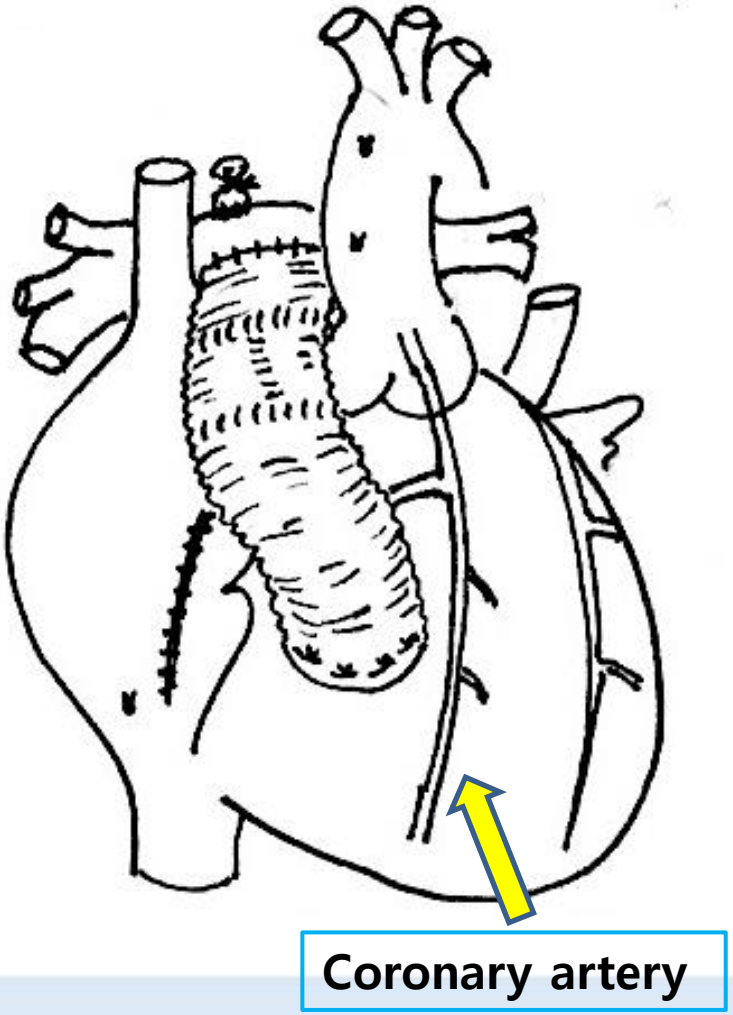




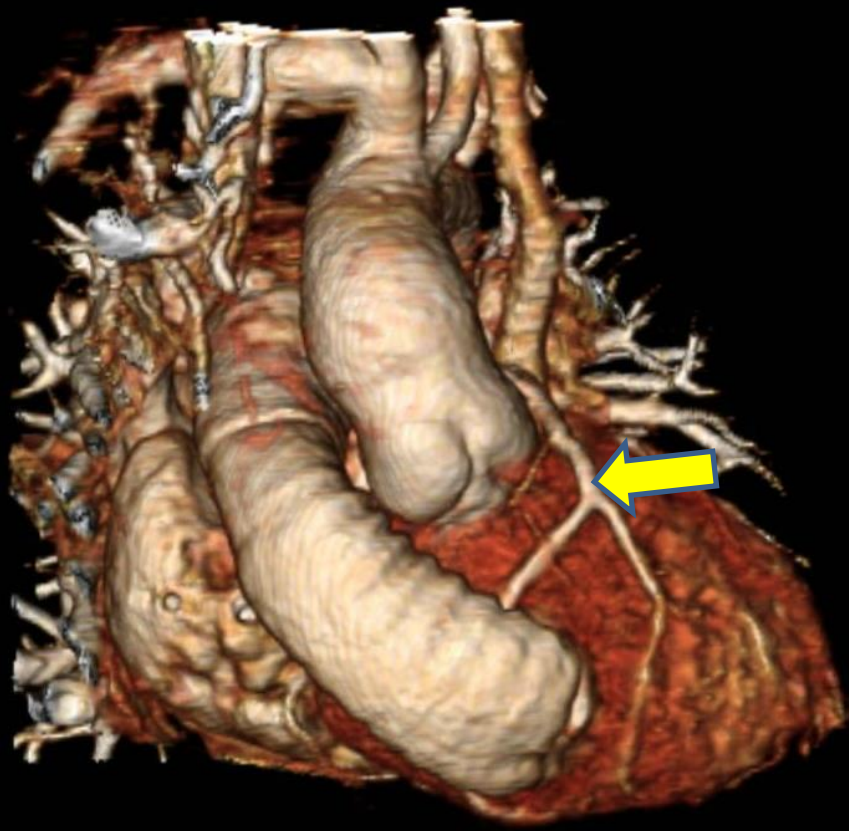
Jonas RA, Comprehensive Surgical Management of Congenital Heart Disease 2nd edition. p350

Rastelli / REV repair

- Midspectrum to Transposition-like DORV With subpulmonary stenosis or Inadequate pulmonary to tricuspid valve separation
- Risk of subaortic stenosis

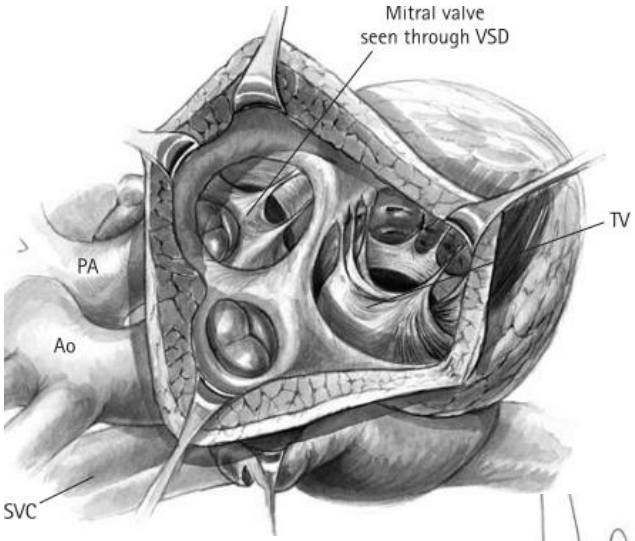


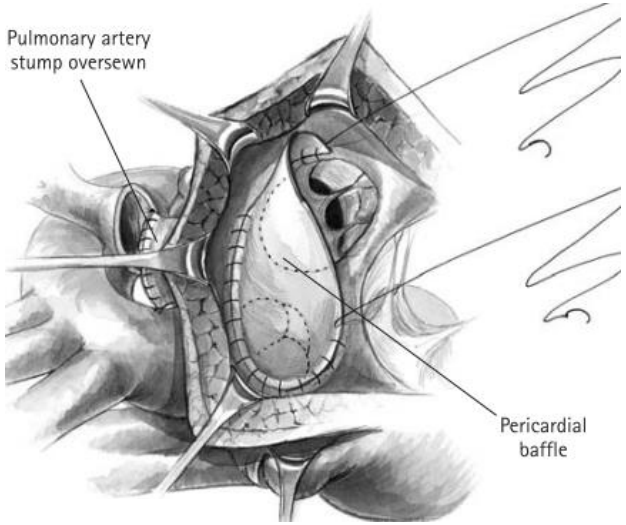
Pre Op.

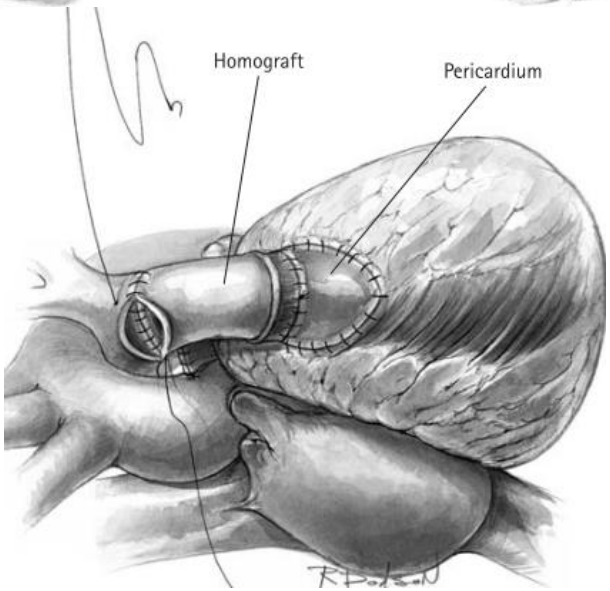


Post Op.

Rastelli procedure







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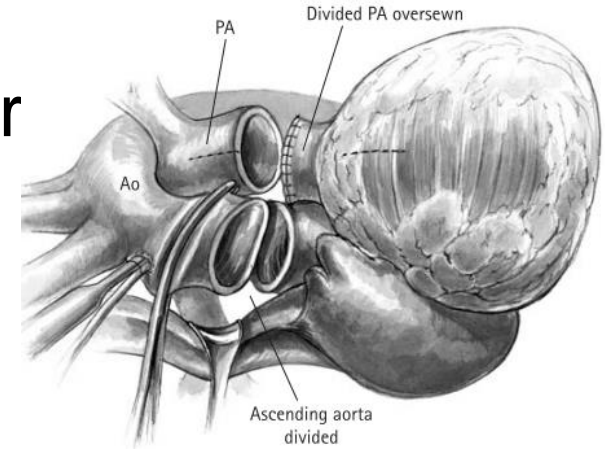
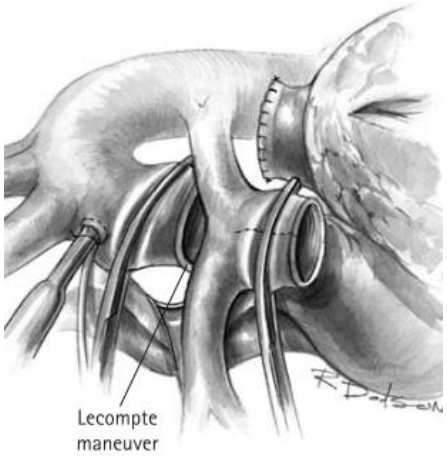
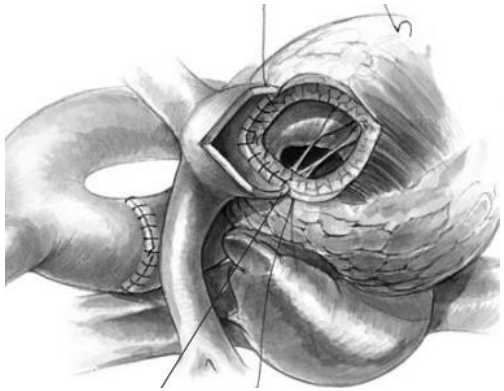
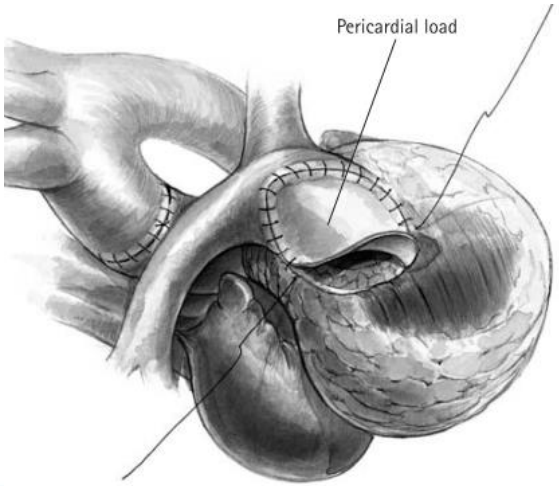
REV (reparation a l'etage ventriculaire) procedure

- Lecompte maneuver
- Dividing of aorta
- Wide mobilization of the pulmoanry arteries









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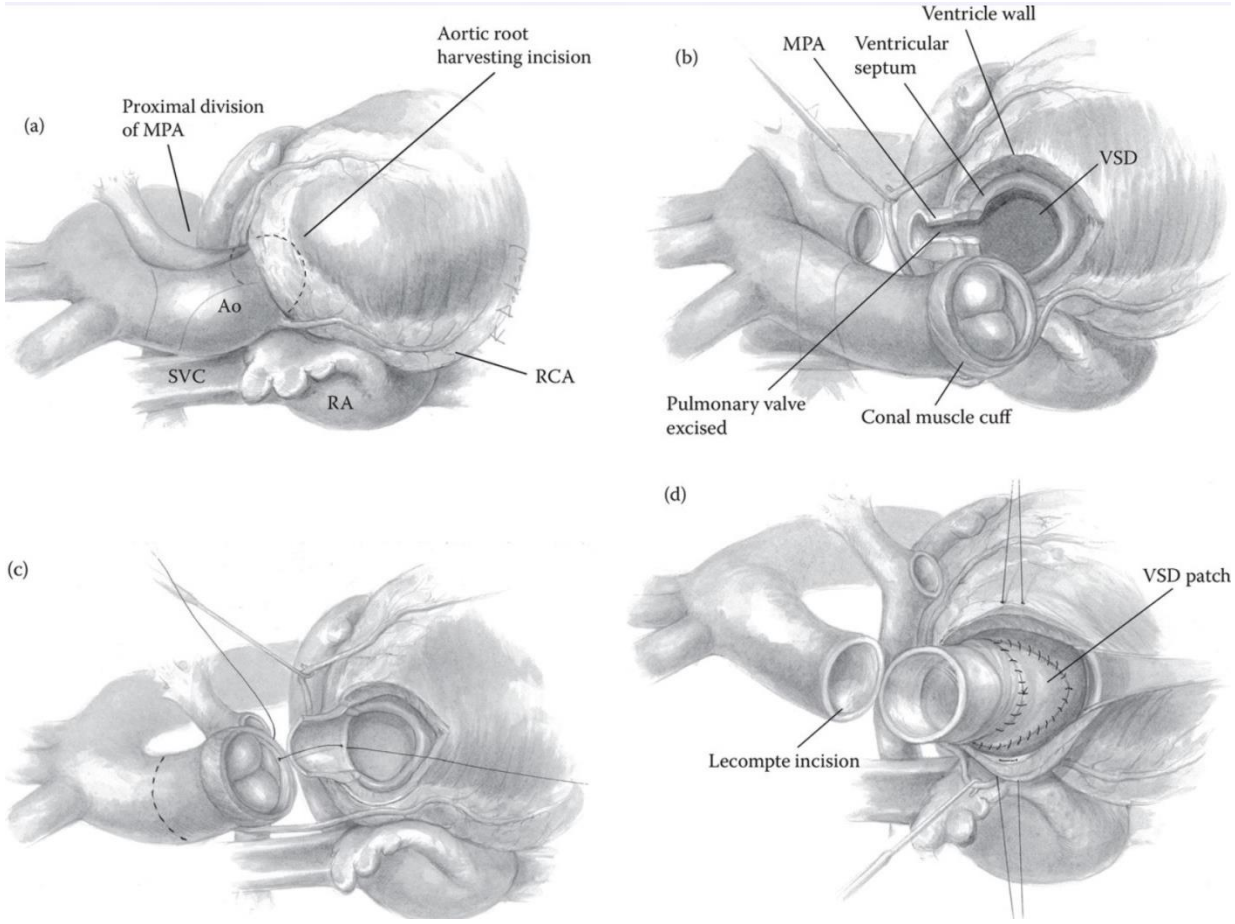
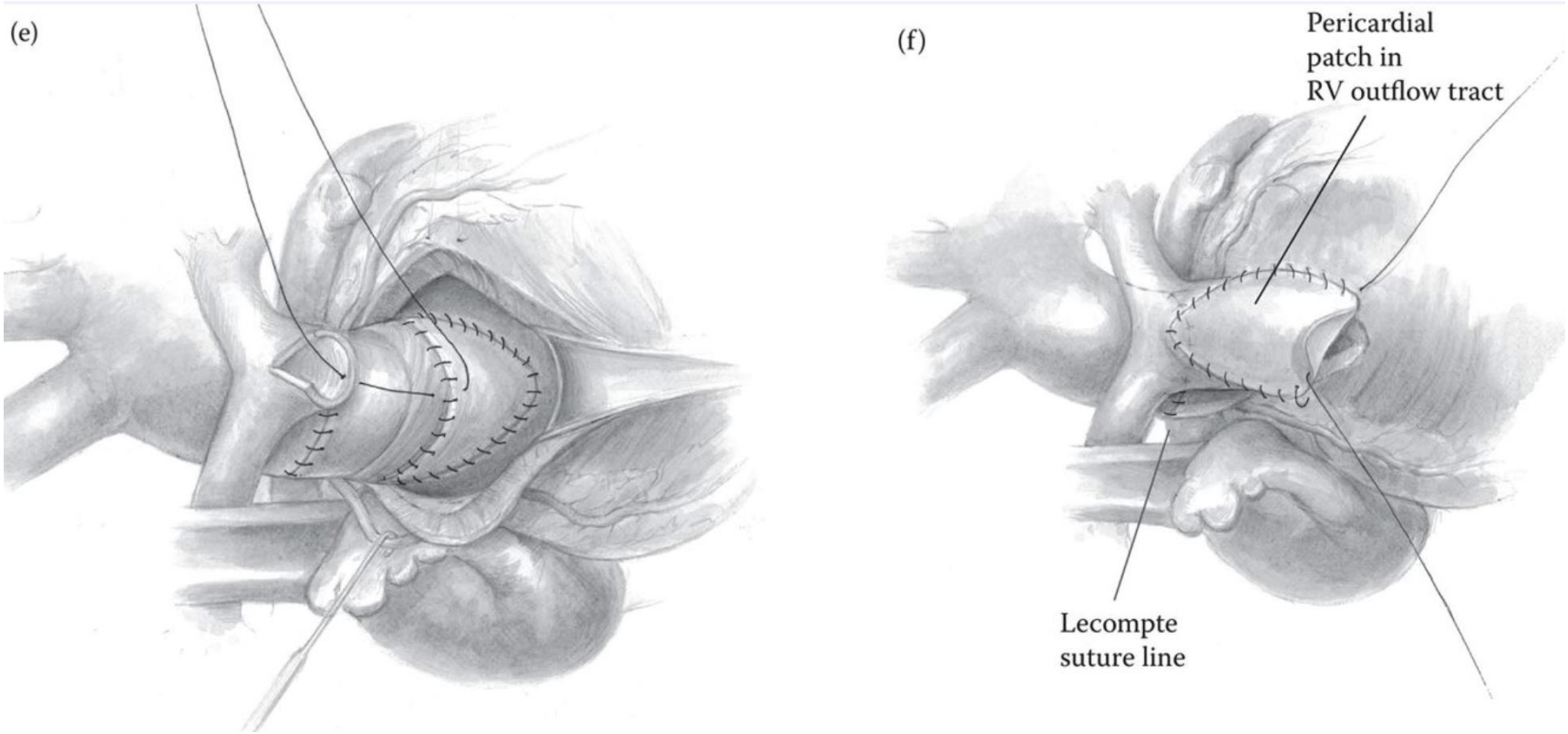
Aortic Translocation (Nikaidoh Procedure)

- Should be careful in patients selection
- DORV with subpulmonary VSD
- Anterior-posterior relationship of the great arteries
- Pulmonary stenosis

Nikaidoh Procedure



Nikaidoh Procedure



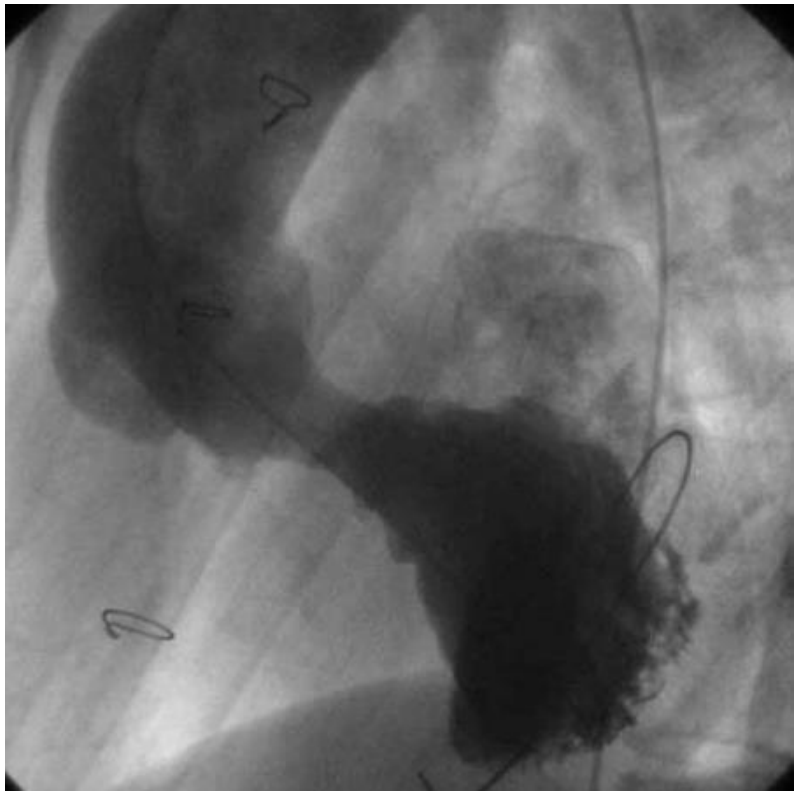
Pre Op.

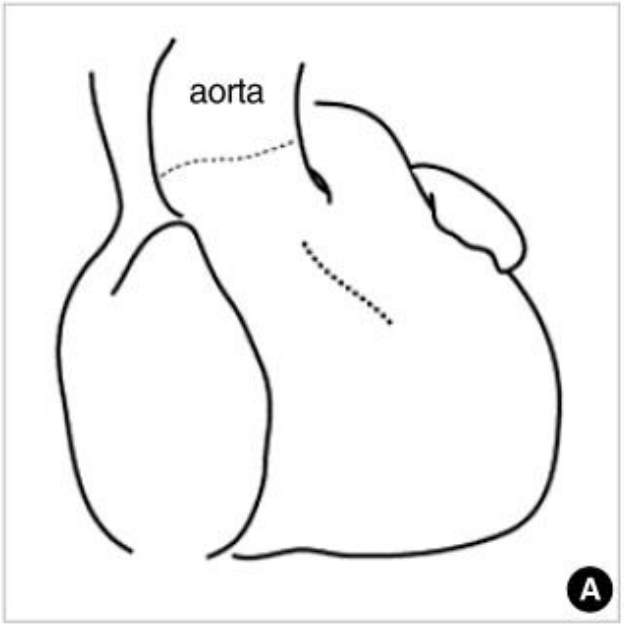
Post Op.

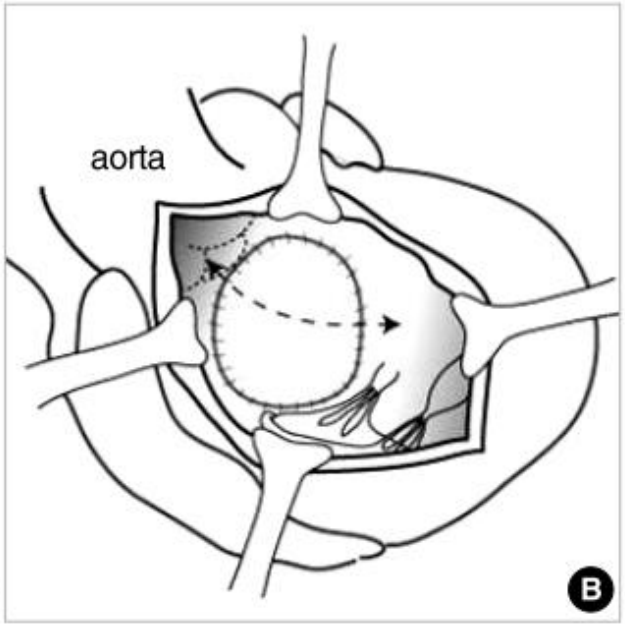
LVOTO after DORV repair

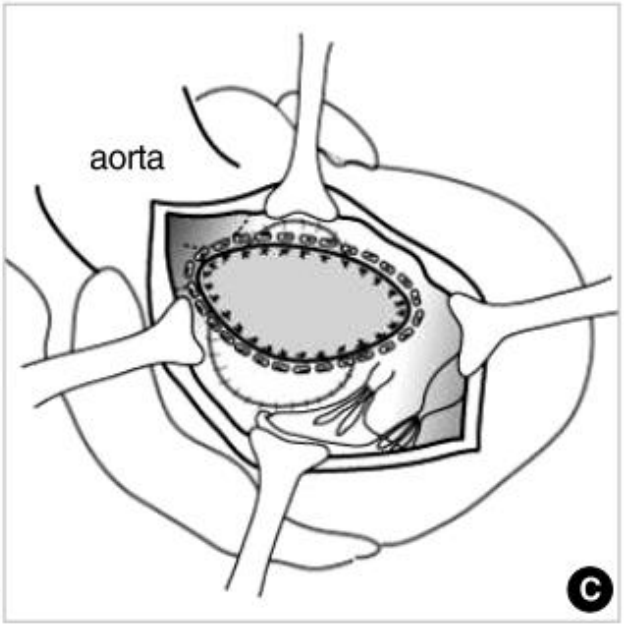
- Subaortic stenosis after biventricular repair of
  a DORV in 3.5-5.5 %

- Before bi-ventricular repair
  - Restrictive VSD
  - Subaortic conal or septal hypertrophy
  - Non committed VSD
  - Even in the presence of a sufficient LVOT and non-restrictive VSD









- Extended septoplasty
- (A) Right ventriculotomy

- (B) Longitudinal septal incision at previous patch
  - Extended toward the apex, in to the interventricular septum,
  - and toward the aortic valve, into the conal septum

- (c) The new patch along the enxtended septal incision

Surgical management of left ventricular outflow tract obstruction after biventricular repair of DORV J Korean Med Sci 2010;25:374-9

Thank you !