

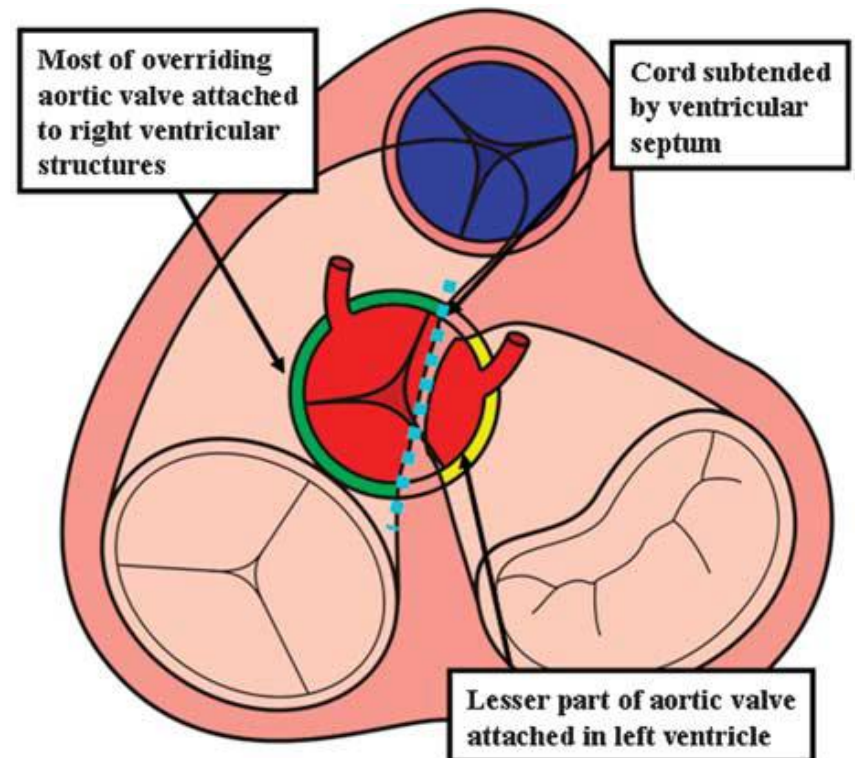
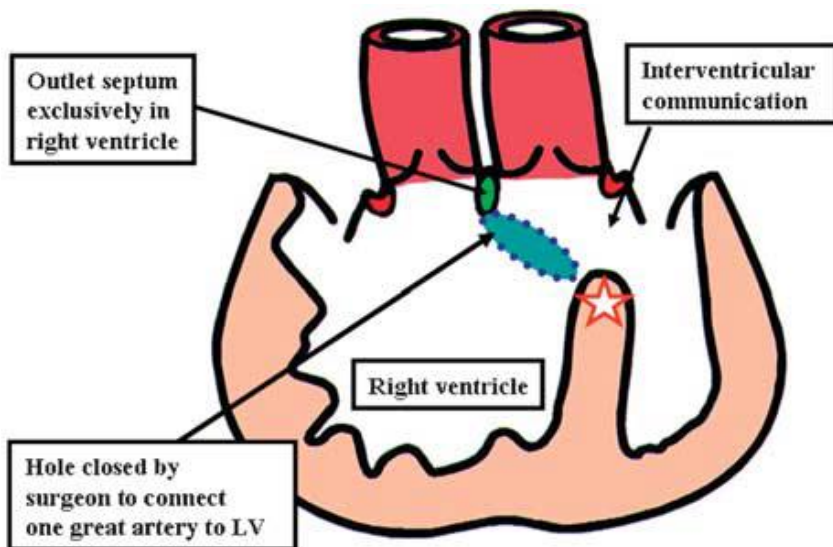
Double Outlet Right Ventricle (DORV)

조성규



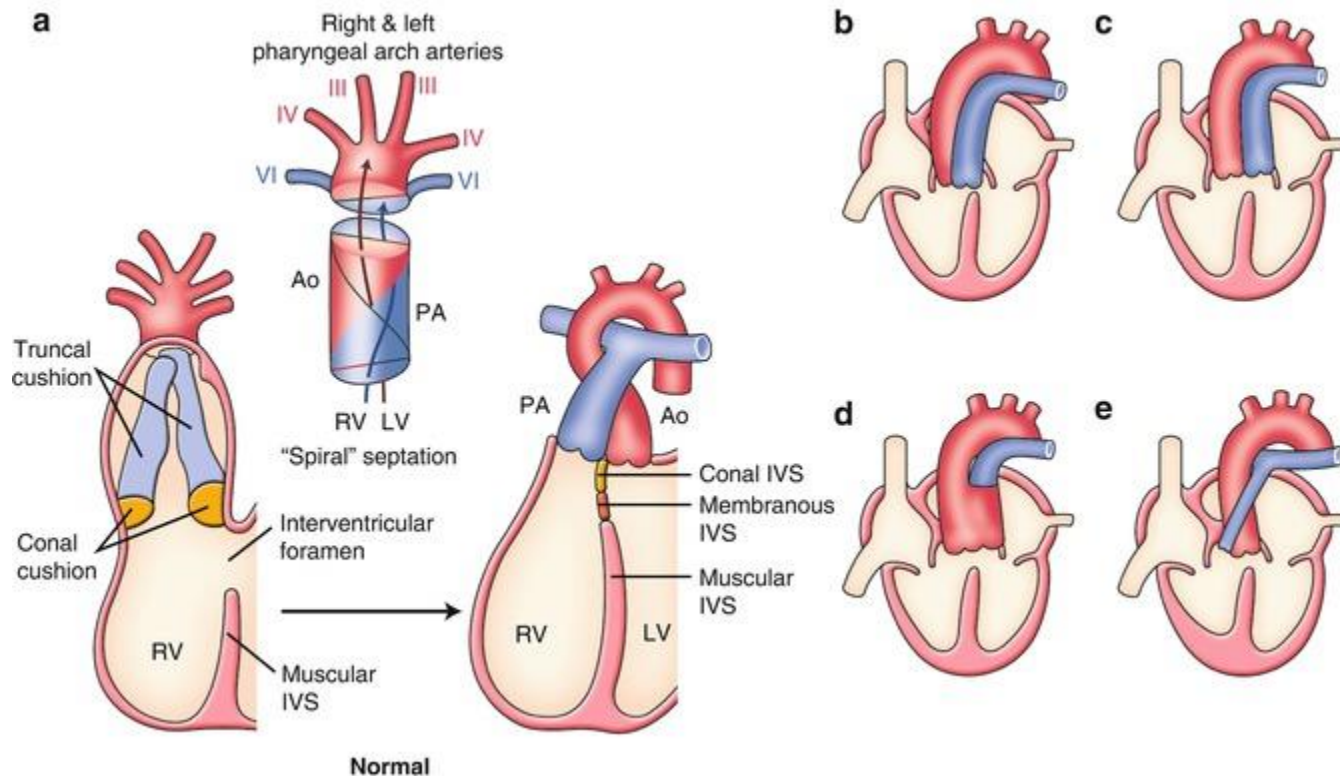
Double Outlet Right Ventricle

- 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 con



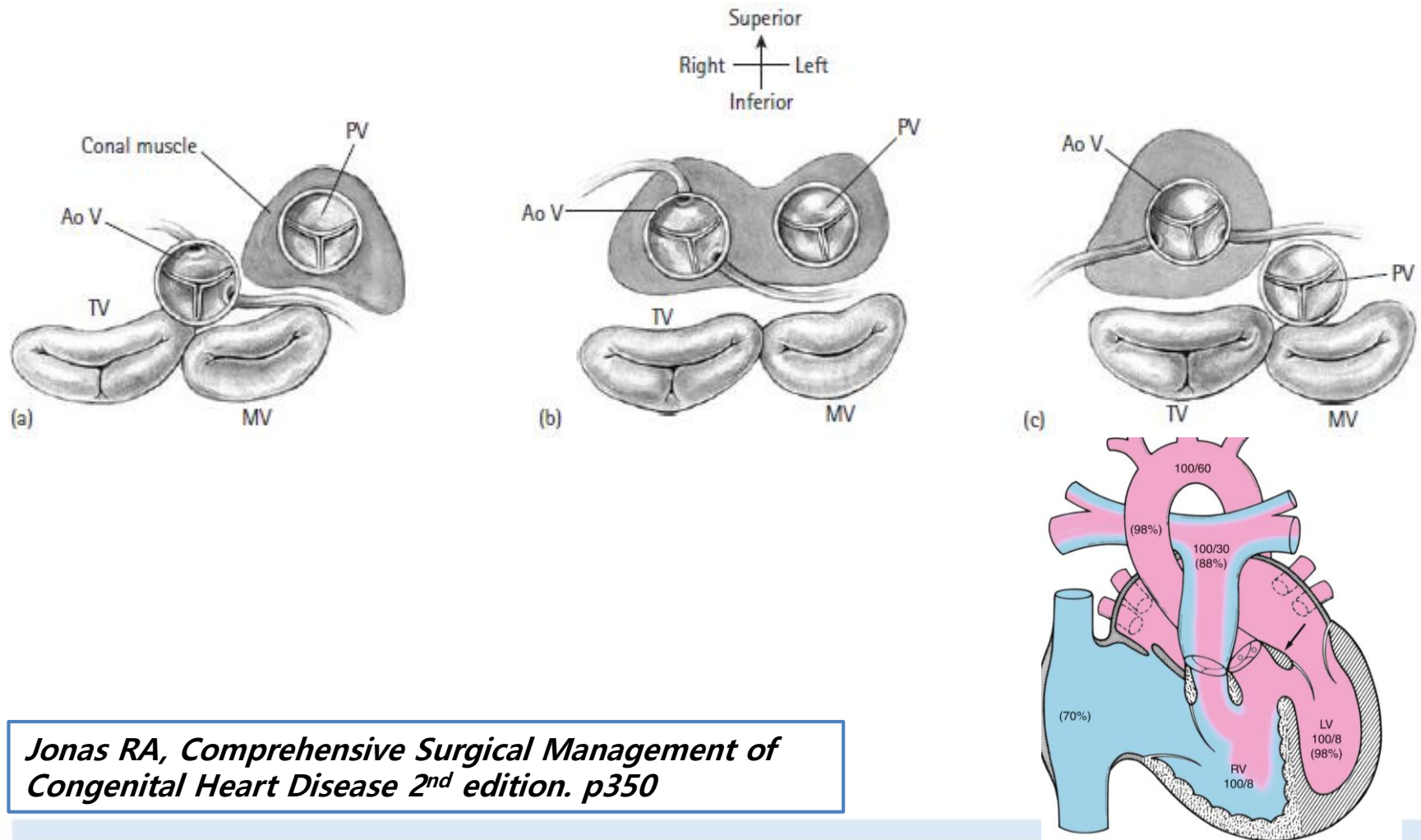
Mahle WT et al, Cardiol Young 2008;18:39-51

Classic Theory of Conotruncal Malseptation

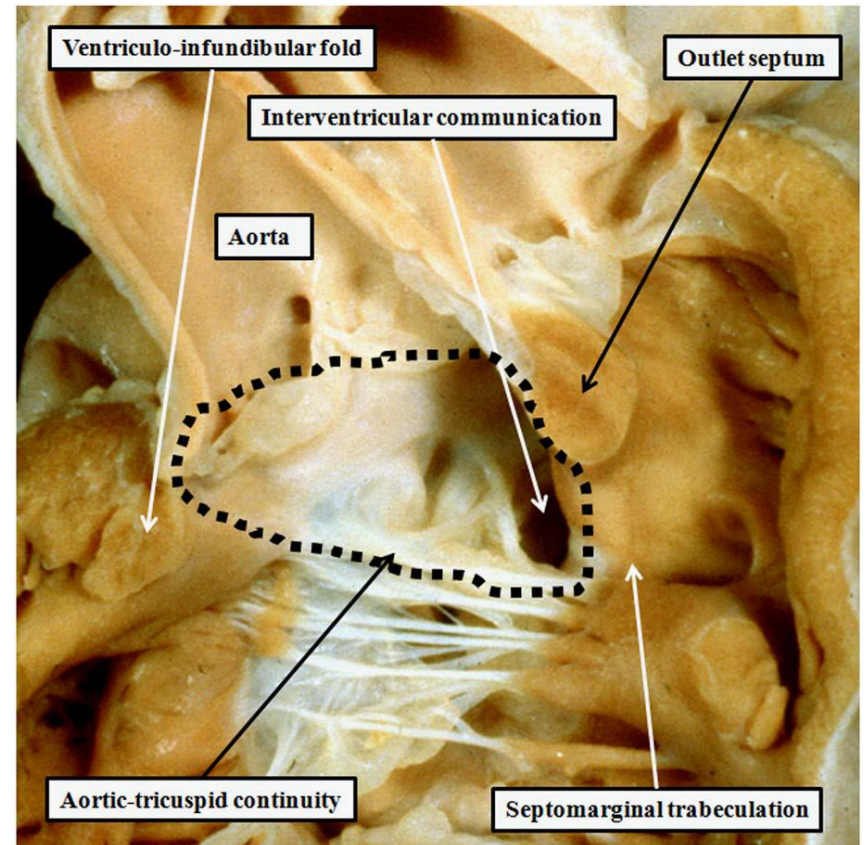
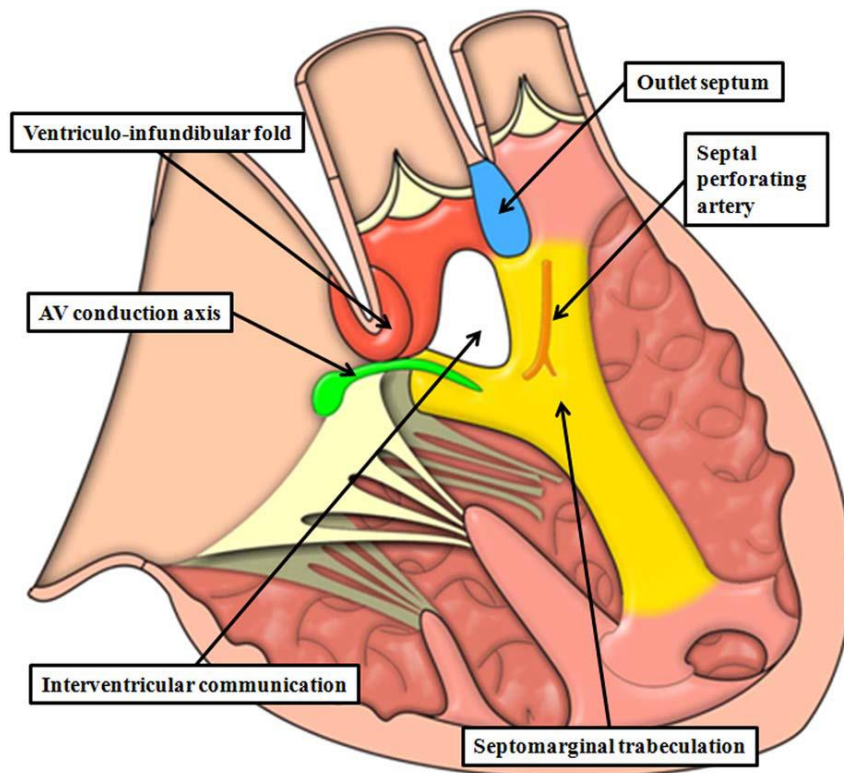


H Yamagish, Cardiac CT and MR for Adult Congenital Heart Disease p13

Van Praagh's Theory of Conal Underdevelopment

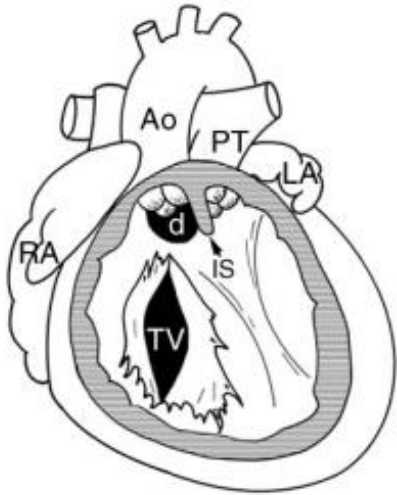


Jonas RA, Comprehensive Surgical Management of Congenital Heart Disease 2nd edition. p350



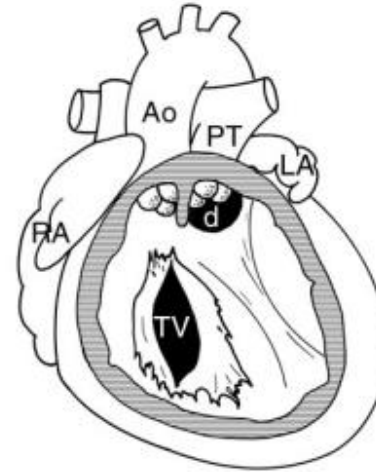
Bharucha T et al, Cardiol Young 2017;27:1-15

Classification



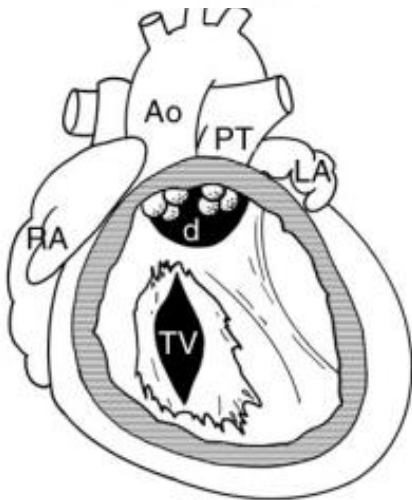
Subaortic VSD

VSD
physiology

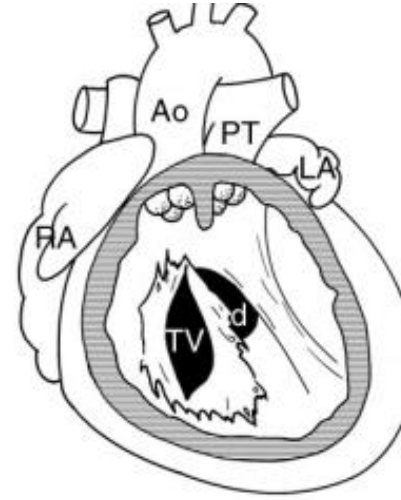


Subpulmonic VSD

C- TGA physiology



Doubly committed
juxtaarterial VSD



Remote VSD

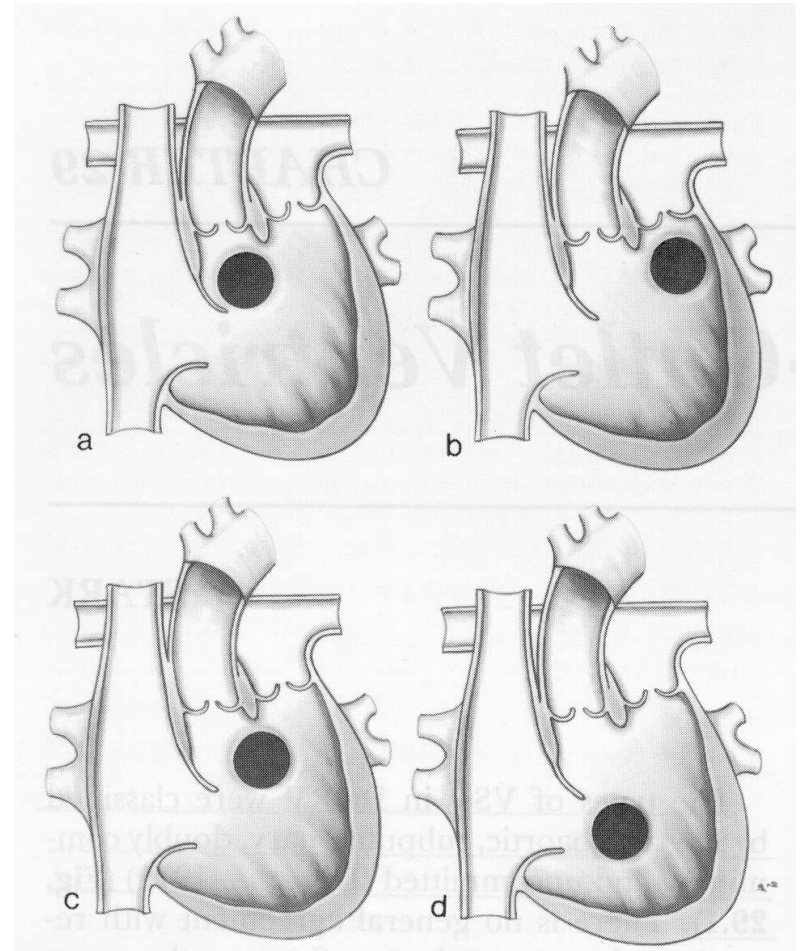
Lev M et al, J Thorac Cardiovasc Surg 64:271-281, 1972

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

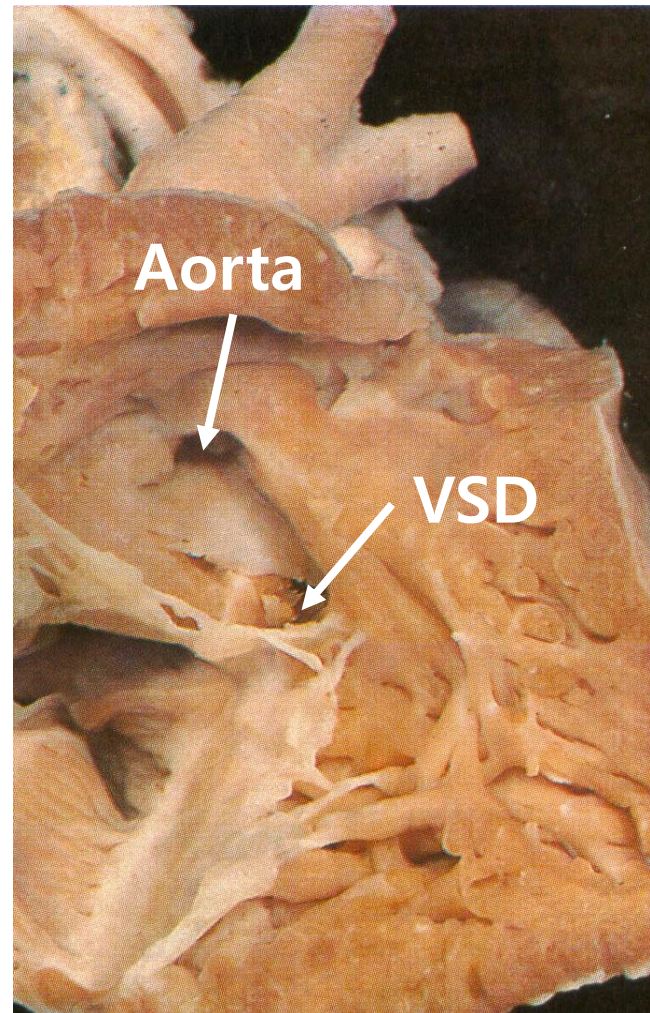
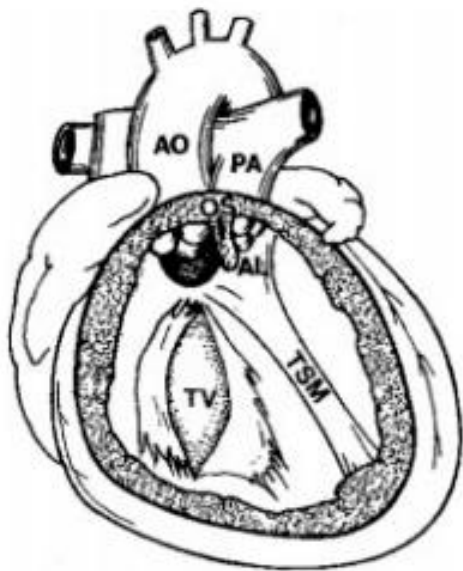
Classification

Locations of VSD in DORV

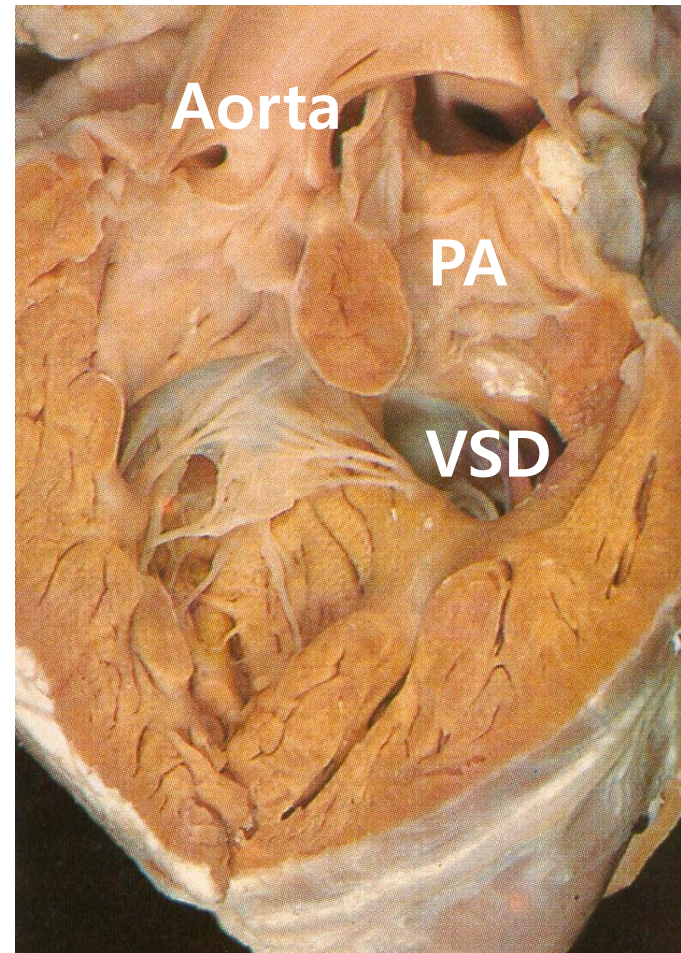
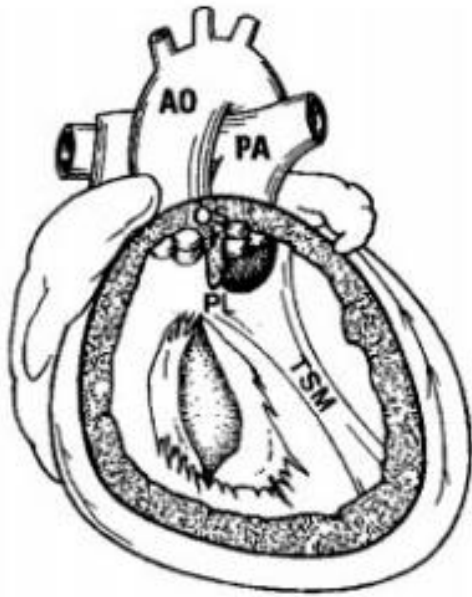
- a ; Subaortic
- b ; Subpulmonic
- c ; Doubly committed
- d ; Noncommitted



- **DORV with Subaortic VSD**



- **DORV with Subpulmonic VSD**



STS-EACTS-APEC classification

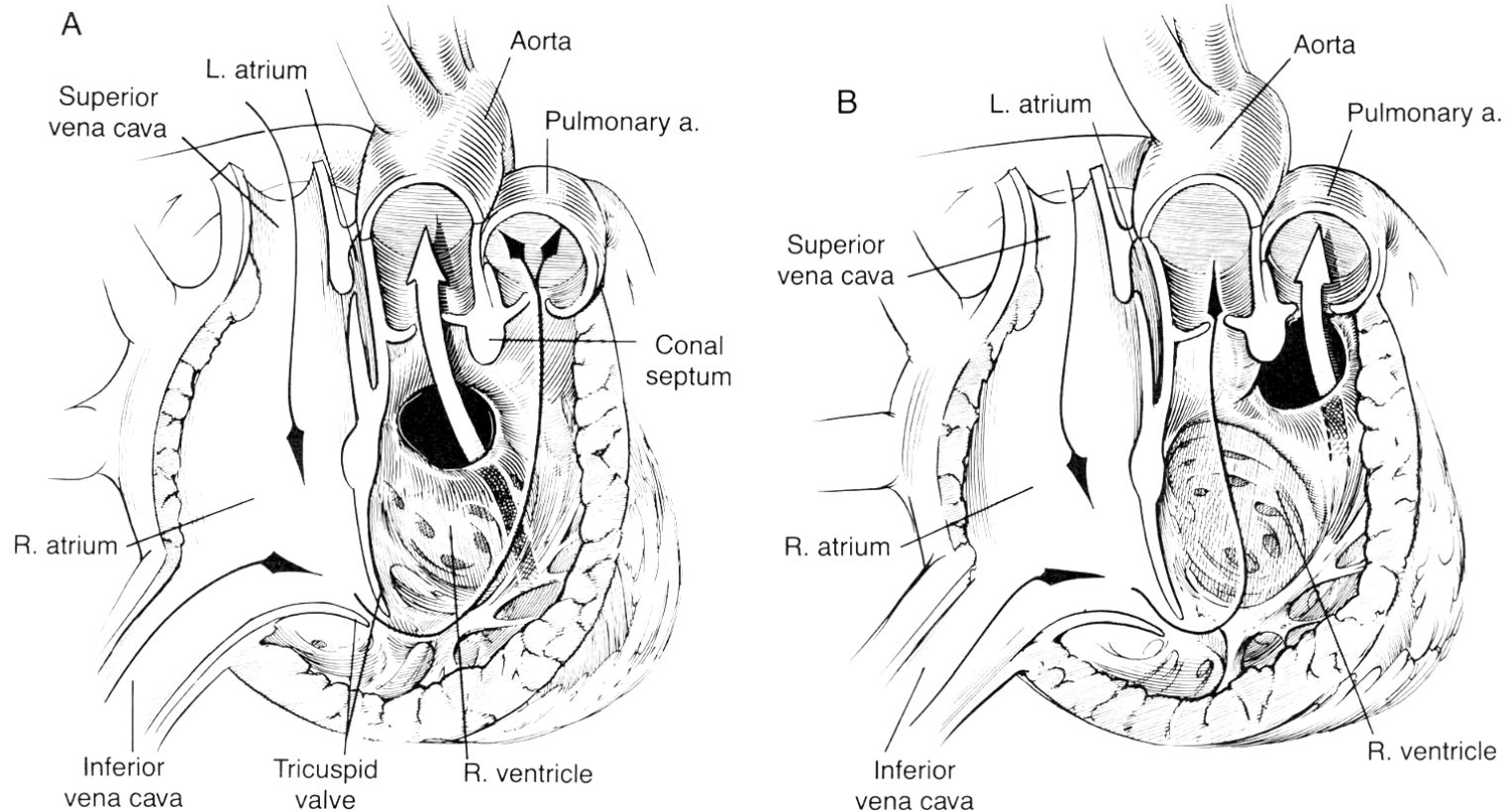
- DORV based on **clinical presentation**
 - VSD-type : DORV with subaortic or doubly committed VSD
 - Follot-type : DORV with subaortic or doubly committed VSD and RVOTO
 - TGA type (Taussig-Bing) : DORV with subpulmonary VSD
 - Non-committed VSD-type : DORV with a remote VSD, possible RVOTO

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

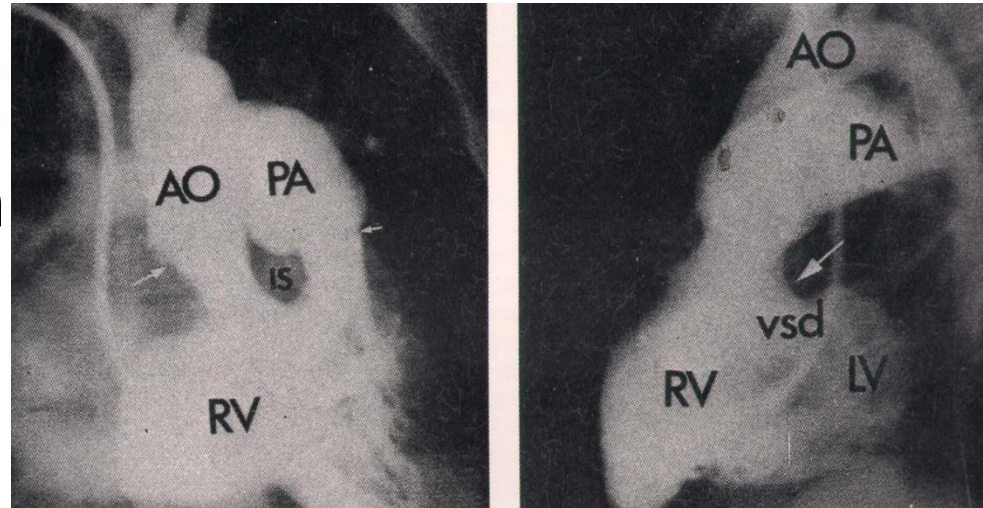
Hemodynamics of DORV



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

Diagnostic Evaluation

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



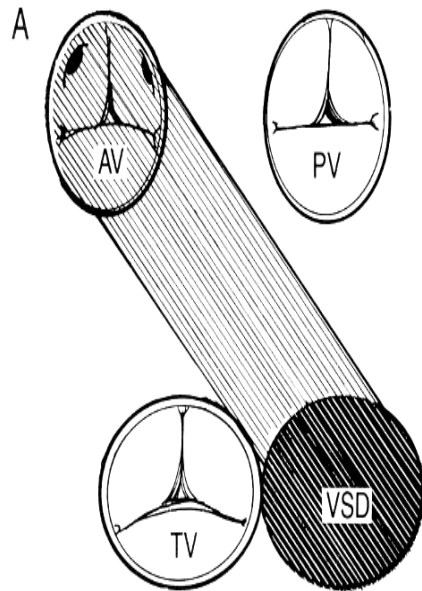
- Computerized tomography

Decision Making for Surgical Treatment

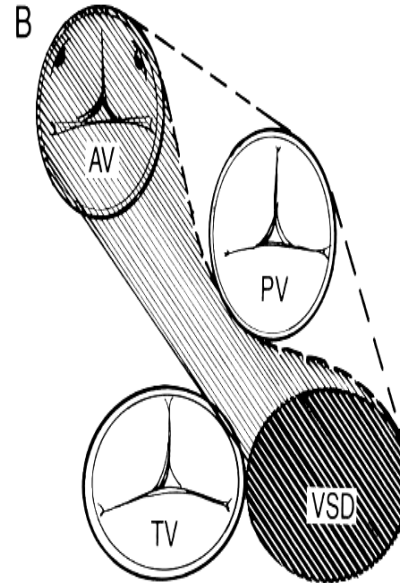
- Determined location and size of the VSD
- Presence and site of pulmonary obstruction
- The degree of aorto-mitral valve separation
- Position of the aortic valve with respect to the pulmonary valve
- Distance between pulmonary and tricuspid valves
- Coronary artery distribution
- Chordal attachments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

Separation between the TV and PV

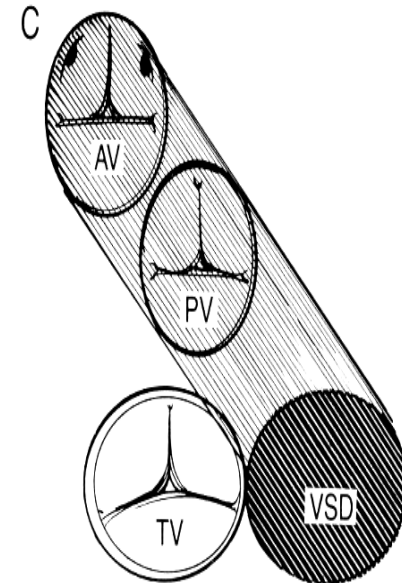
- Determining anatomic suitability for an intraventricular baffle repair



**Adequate
separation**



**Risk of subaortic
stenosis**



Rastelli procedure

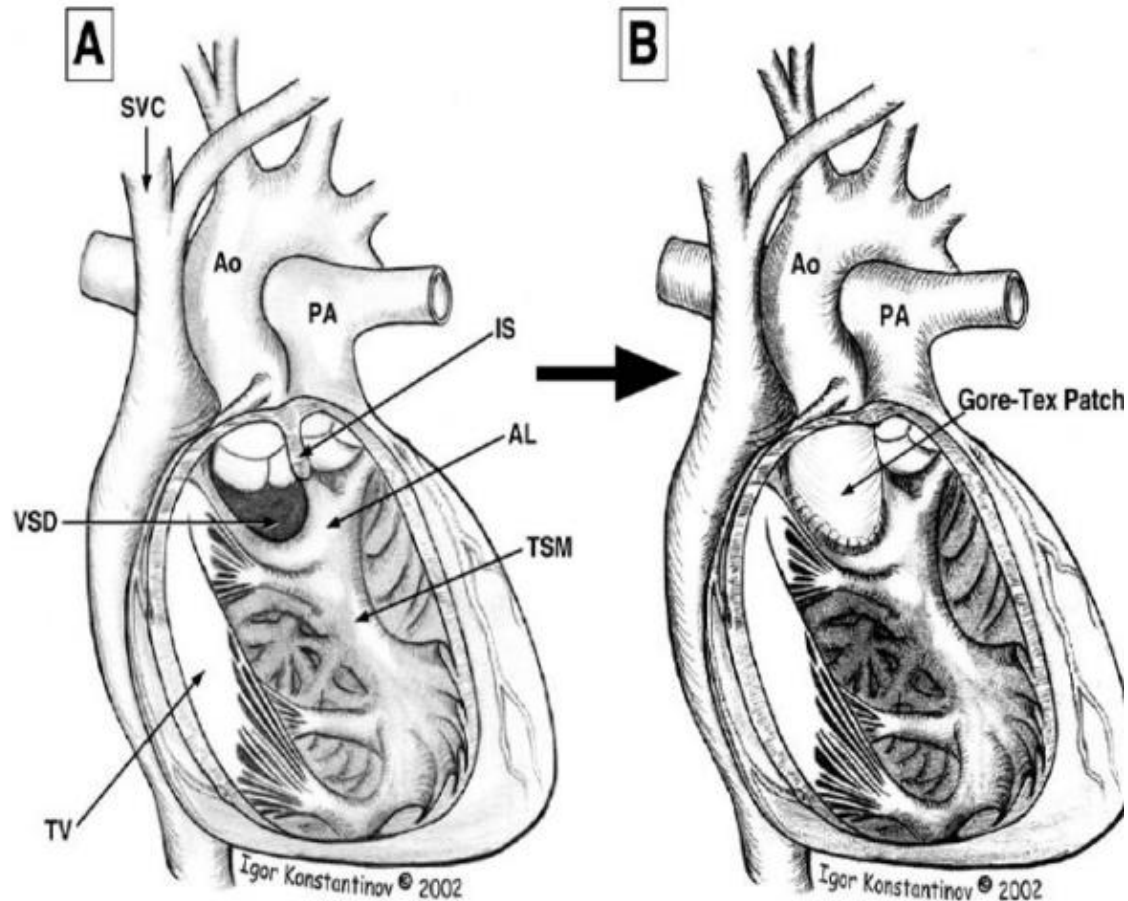
Surgical Techniques

- 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

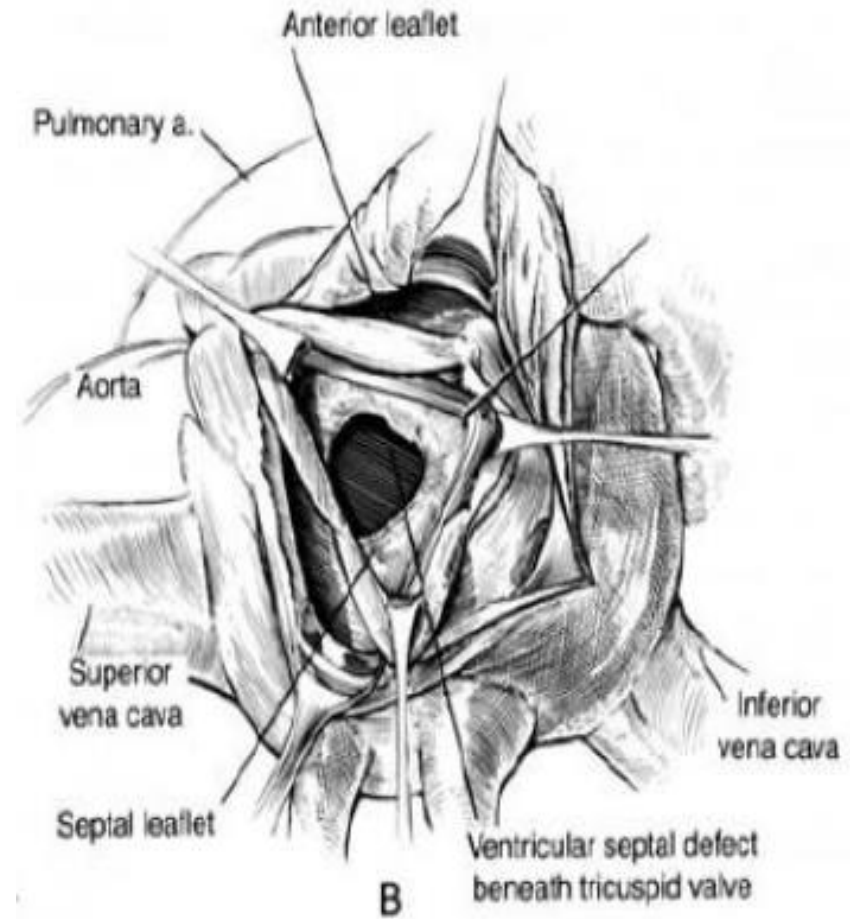
Intraventricular tunnel repair (VSD to aorta)



VSD Baffle to aortic valve

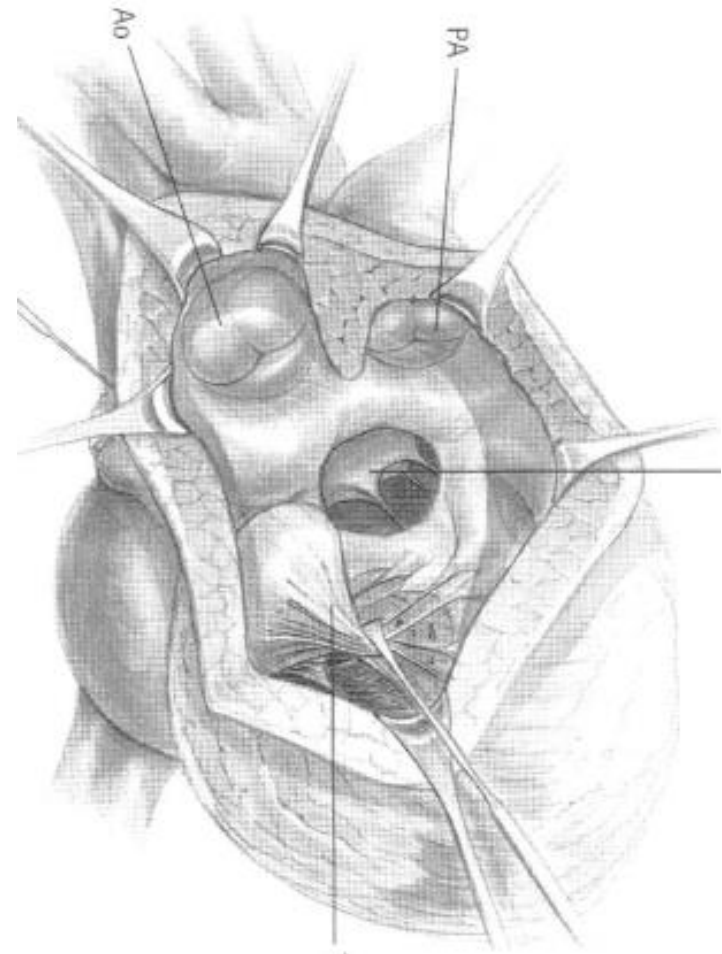
RA approach

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



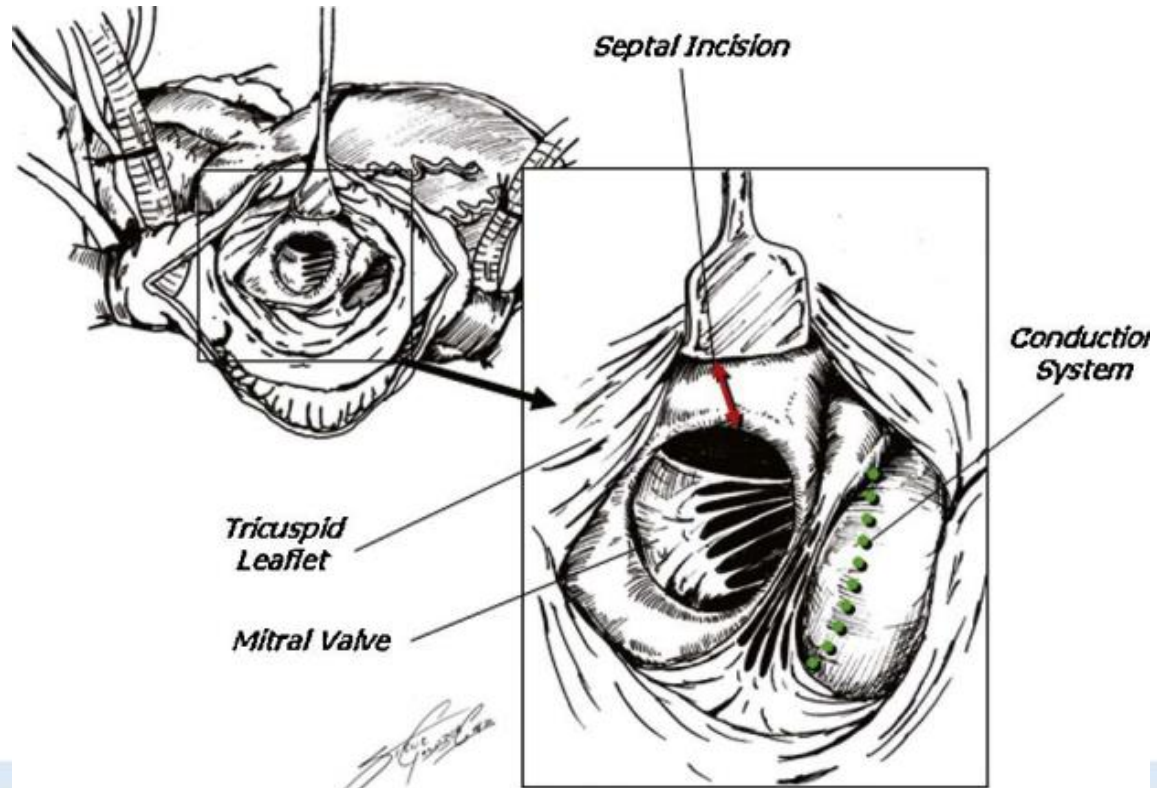
RV approach (RV infundibulum)

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



VSD enlargement

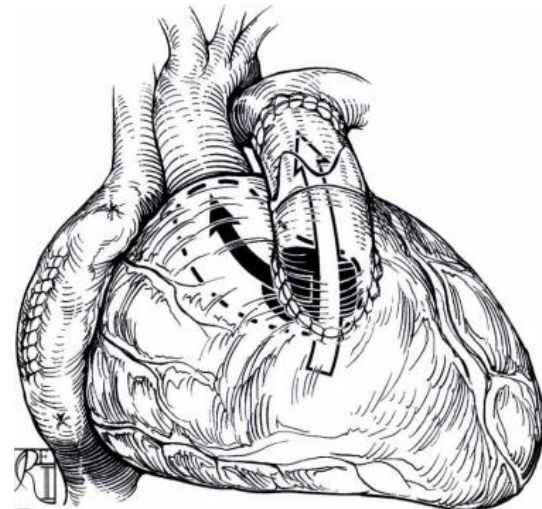
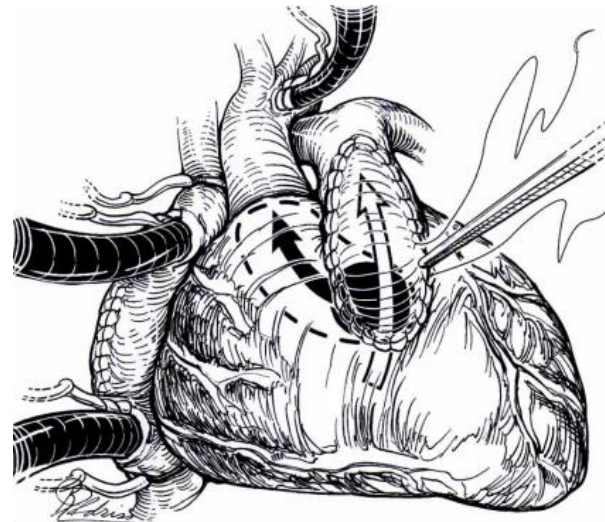
- Restrictive VSD
 - Incision anterior superiorly
 - Resecting a wedge of the interventricular septum
- Risk of injury
 - Mitral valve & tensor apparatus
 - Ant. Vent wall & LAD, septal perforator



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

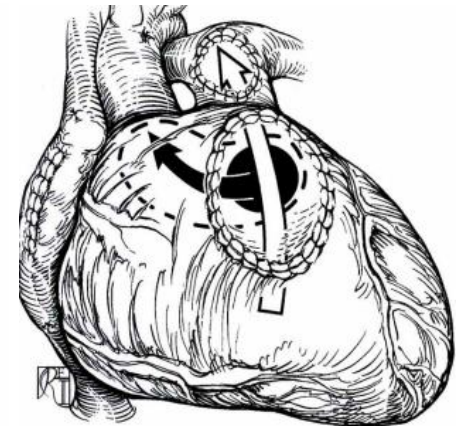
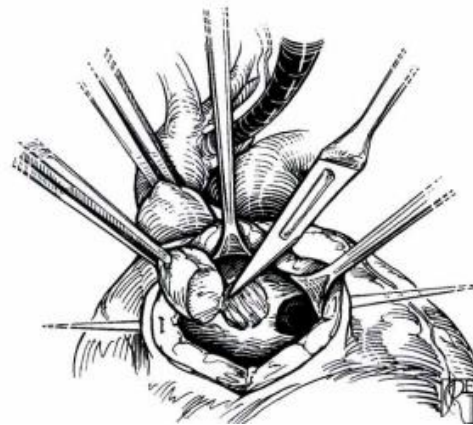
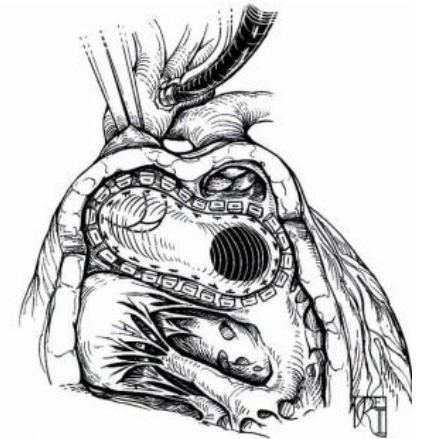
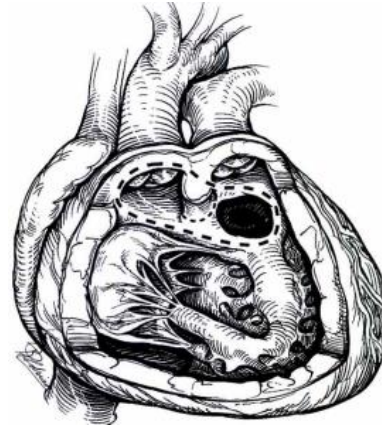


DORV with Subpulmonary VSD (DORV-TGA type, Taussig-Bing)

- 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)

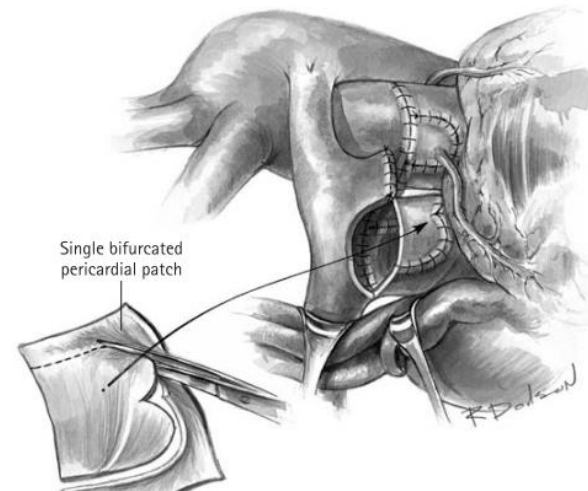
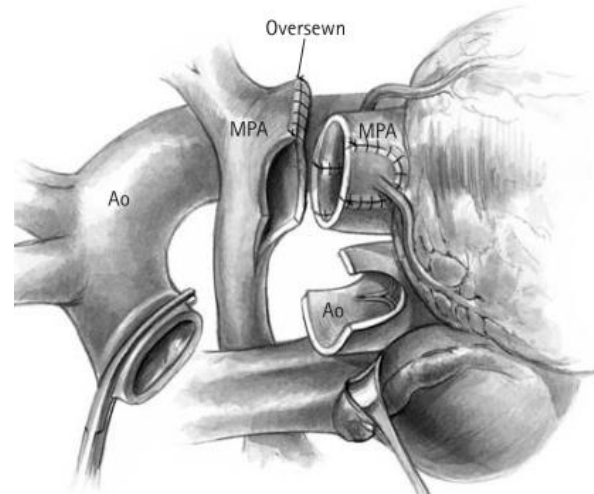
Kawashima operation

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



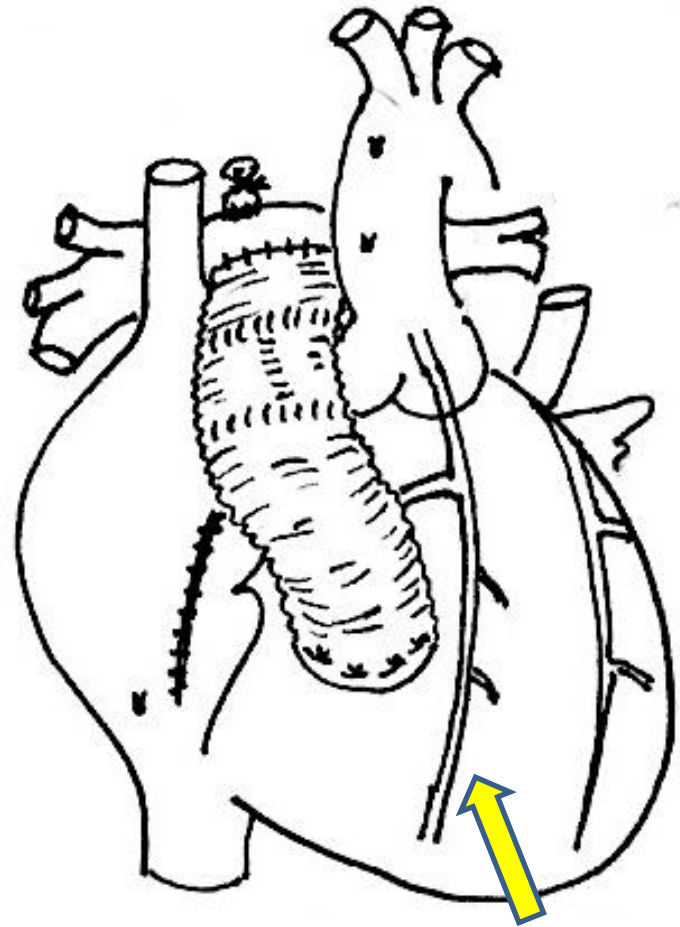
Arterial switch operation with tunnel closure of the VSD

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

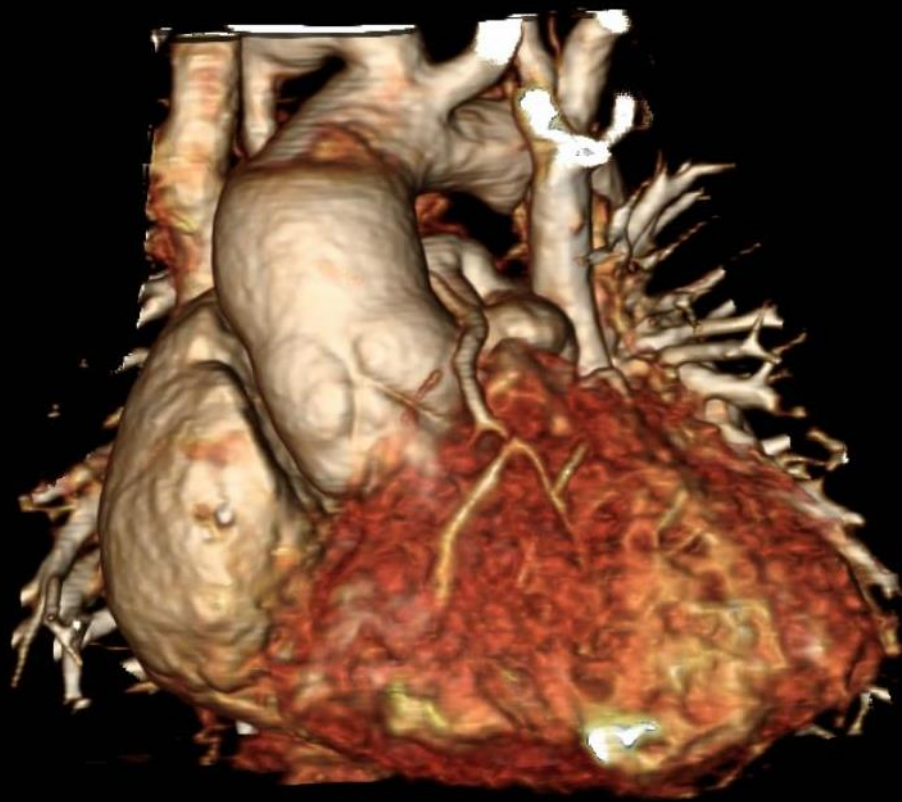


Rastelli / REV repair

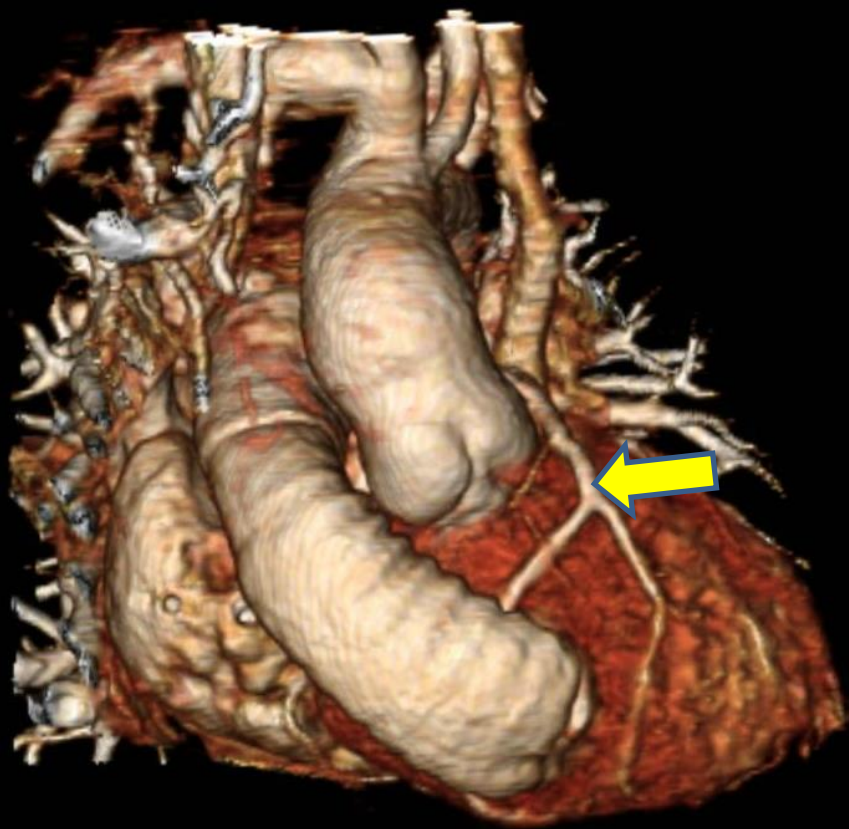
- Midspectrum to Transposition-like DORV With subpulmonary stenosis or Inadequate pulmonary to tricuspid valve separation
- Risk of subaortic stenosis



Coronary artery

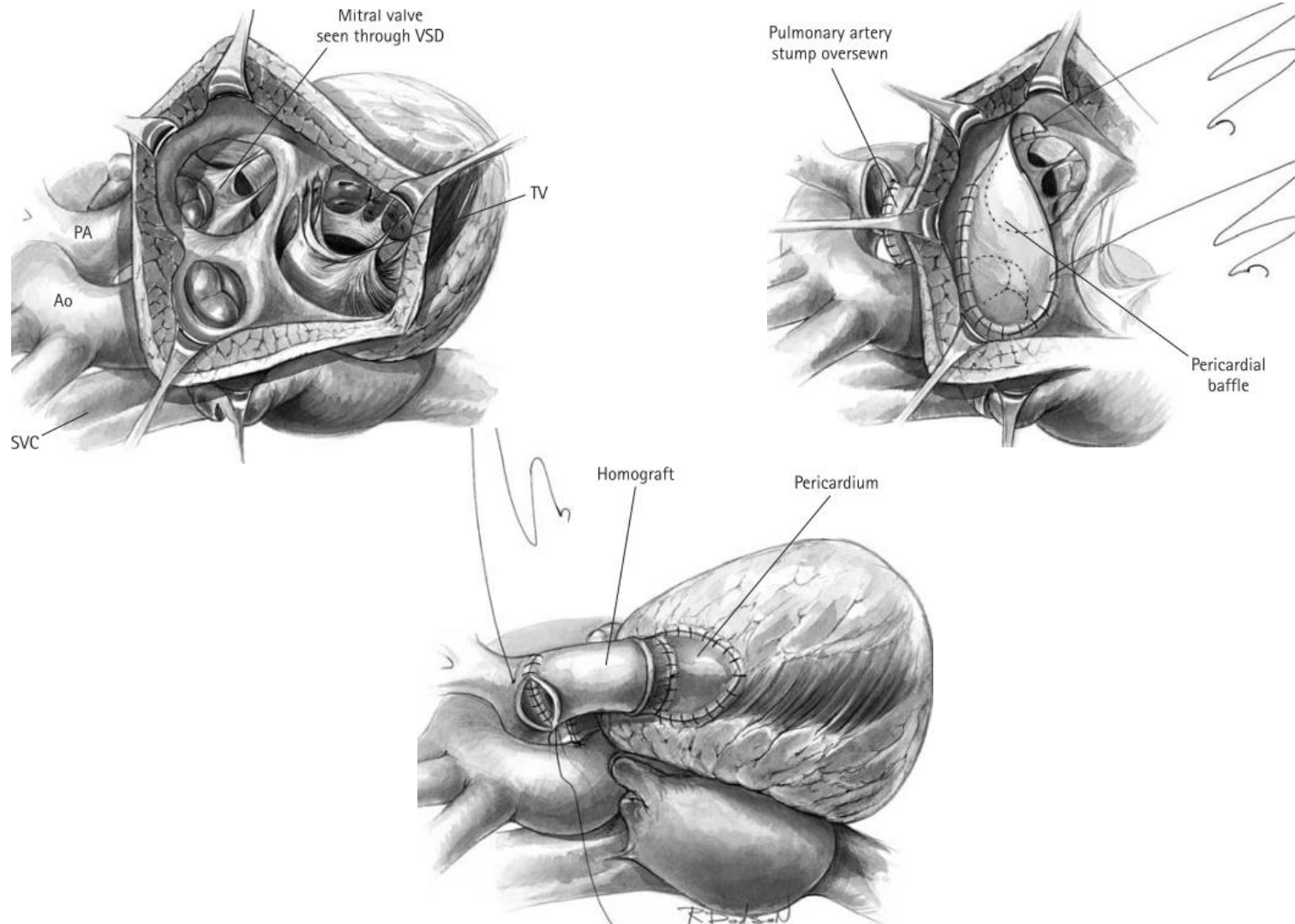


Pre Op.



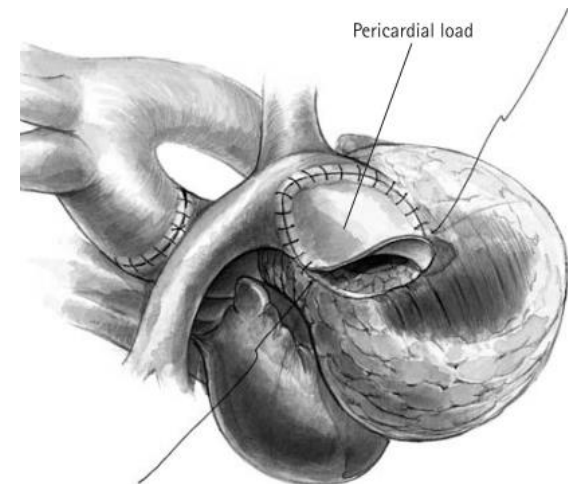
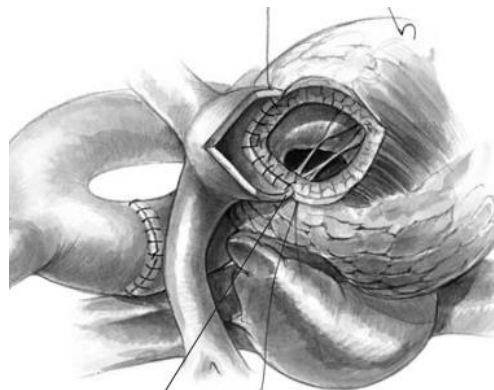
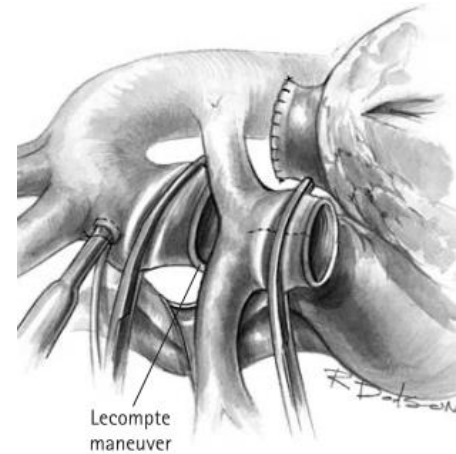
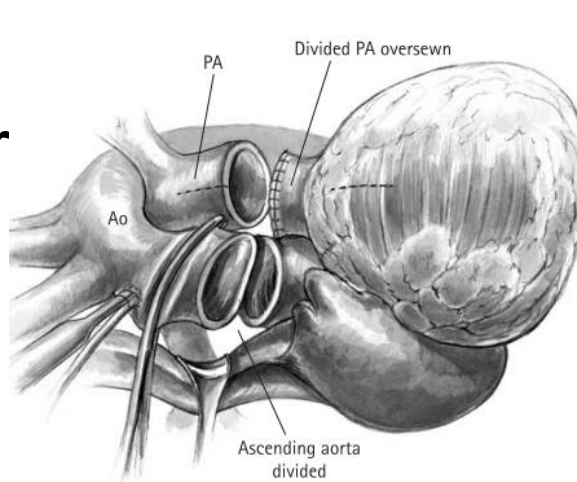
Post Op.

Rastelli procedure



REV (reparation a l'etage ventriculaire) procedure

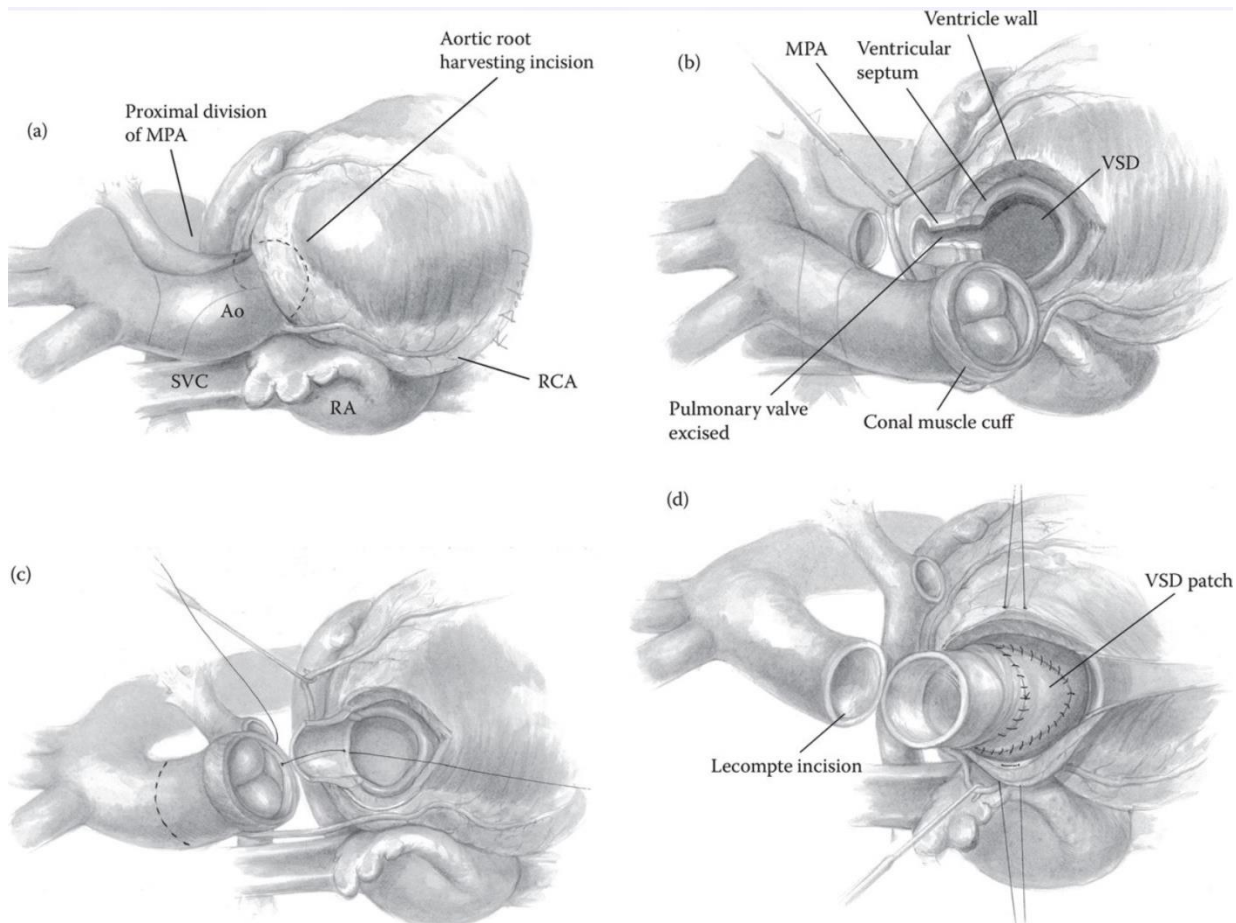
- Lecompte maneuver
- Dividing of aorta
- Wide mobilization of the pulmonary arteries



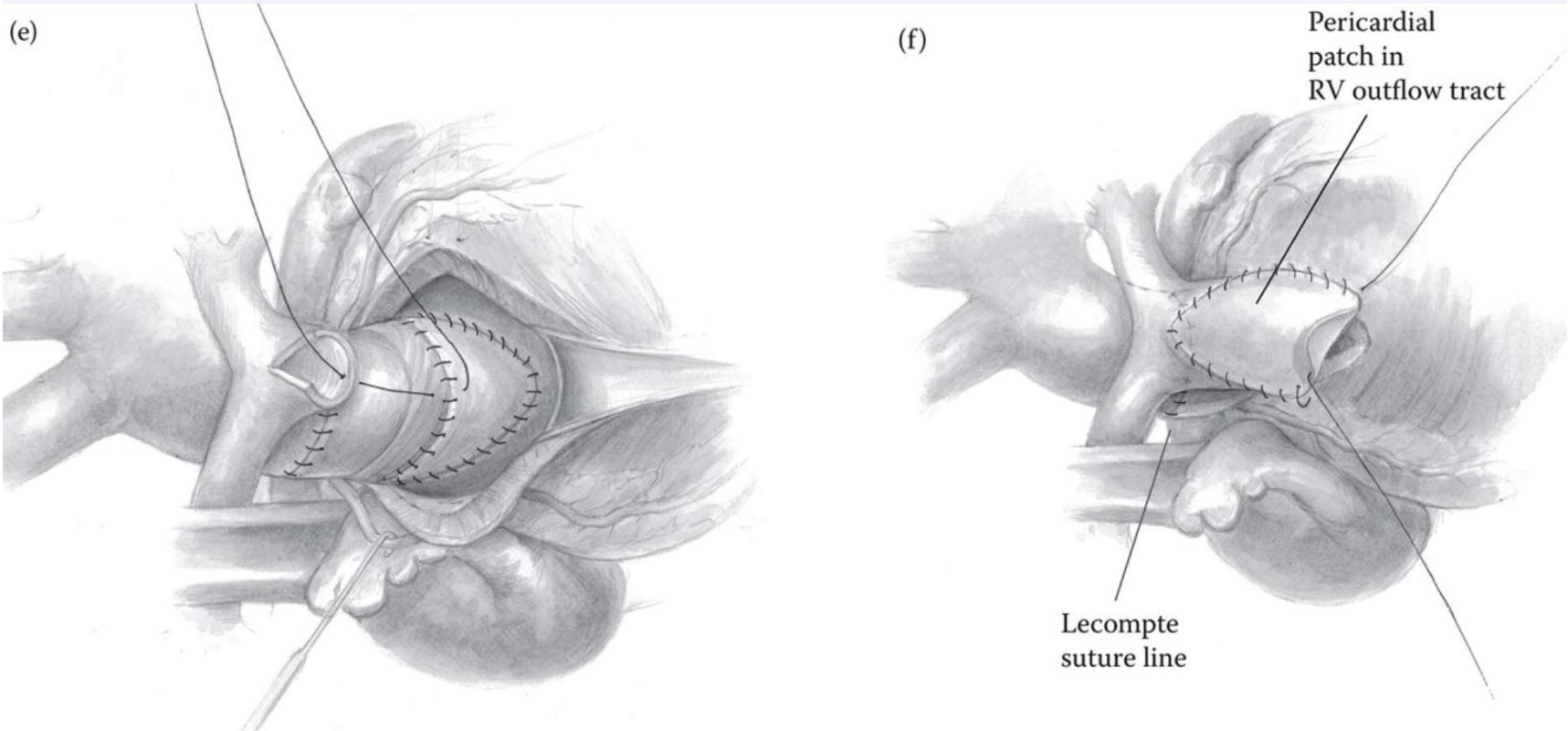
Aortic Translocation (Nikaidoh Procedure)

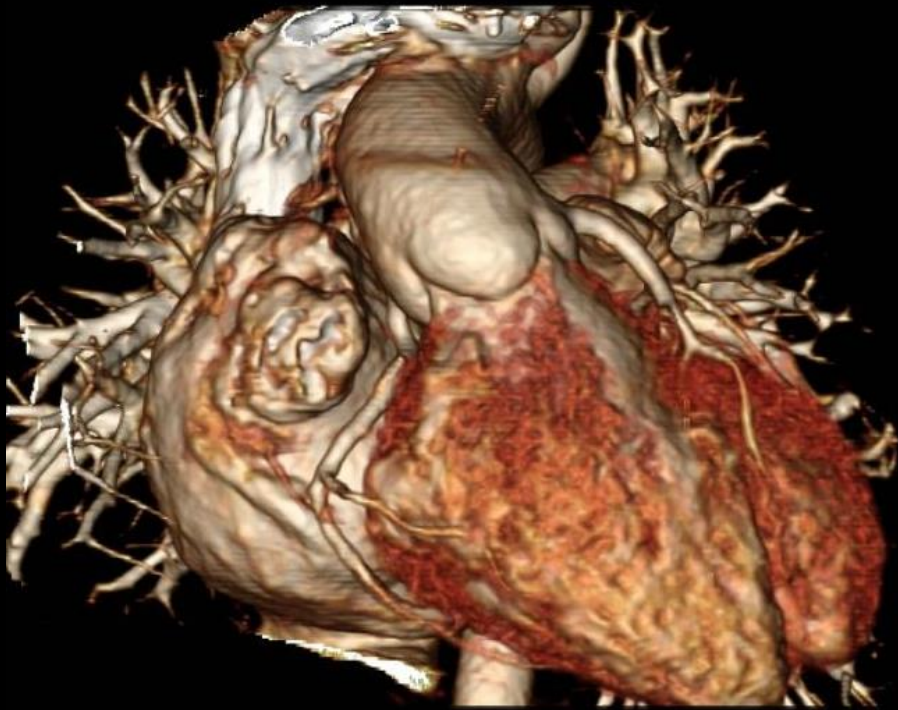
- Should be careful in patients selection
- DORV with subpulmonary VSD
- Anterior-posterior relationship of the great arteries
- Pulmonary stenosis

Nikaidoh Procedure



Nikaidoh Procedure





Pre Op.

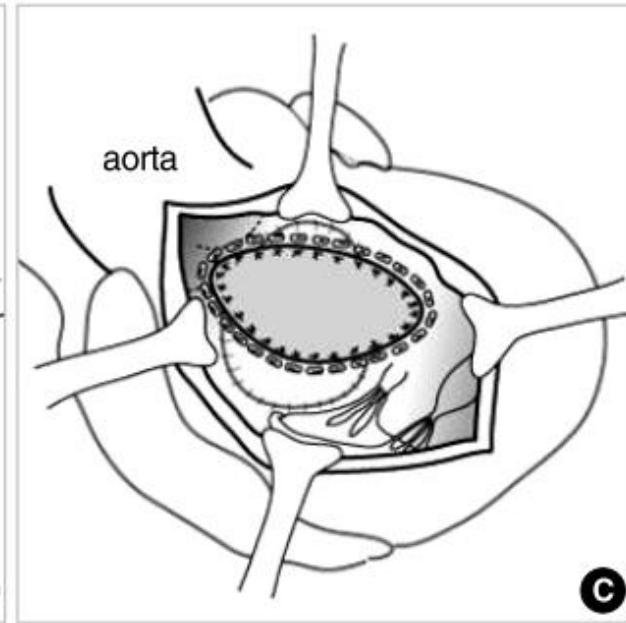
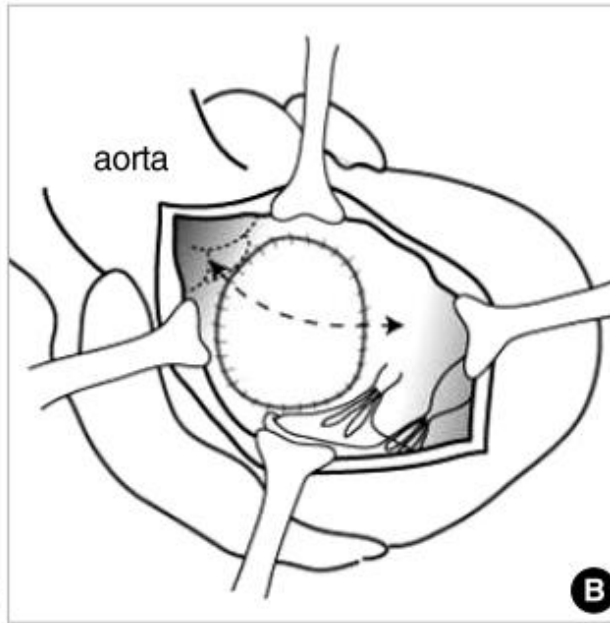
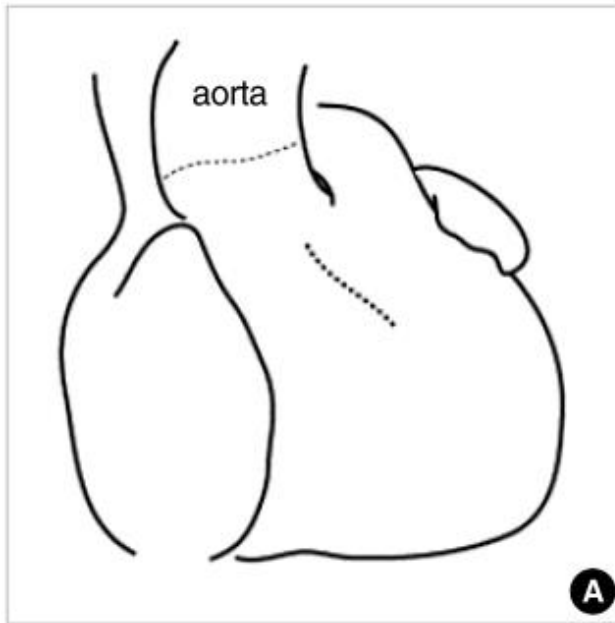


Post Op.

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 extended septal incision

Thank you !