Surgical Options in DORV

Bastien PROVOST

Marie Lannelongue Hospital

SickKids Hospital

Plan

- Rappels historiques / anatomiques
- Classification(s)
- Indication chirurgicale et Timing
- Techniques chirurgicales
- Univentriculaire ?

VDDI

=Ventricule Droit a Double Issue

= Malposition vasculaire

= DORV Double outlet right ventricle

Anomalies conotroncales

Transposition des gros vaisseaux +/- CIV (D-TGA)

Tétralogie de Fallot

Truncus

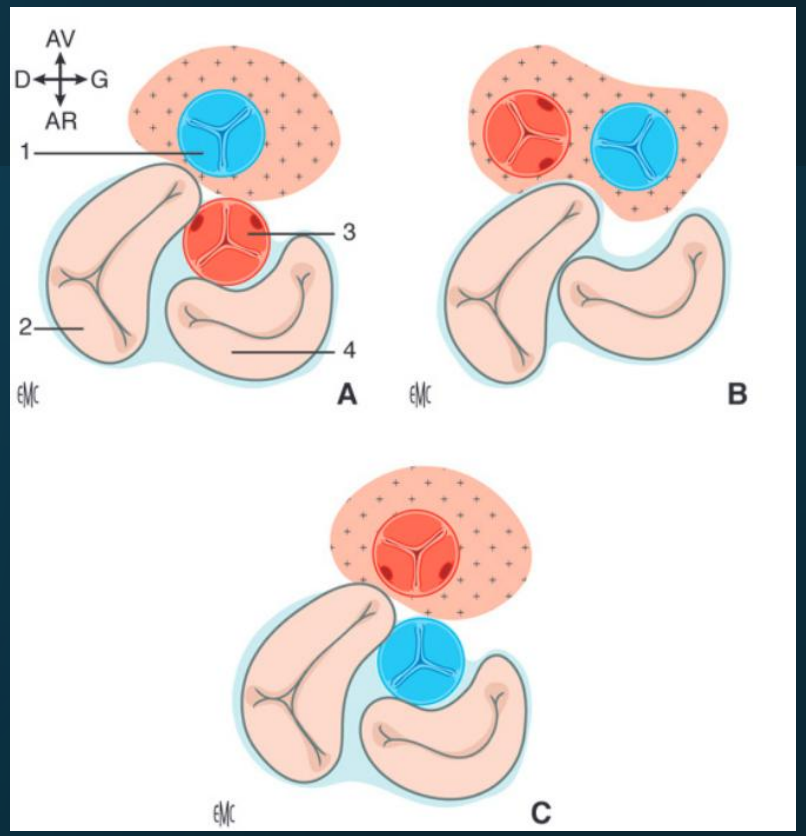
Double discordance (L-TGA) ccTGA

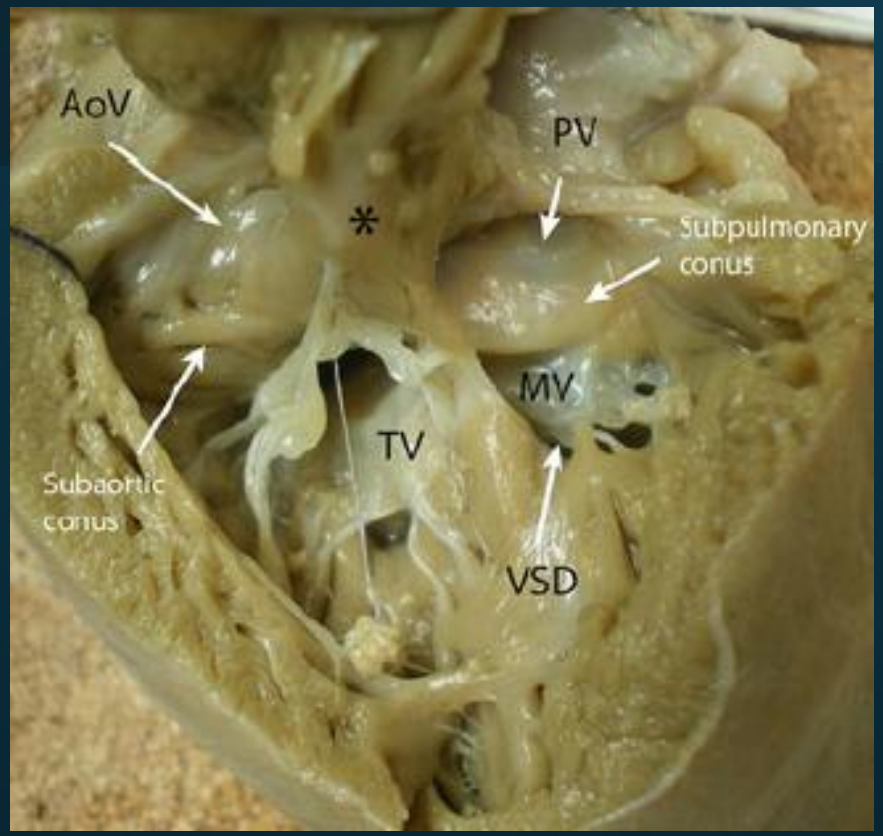
VDDI

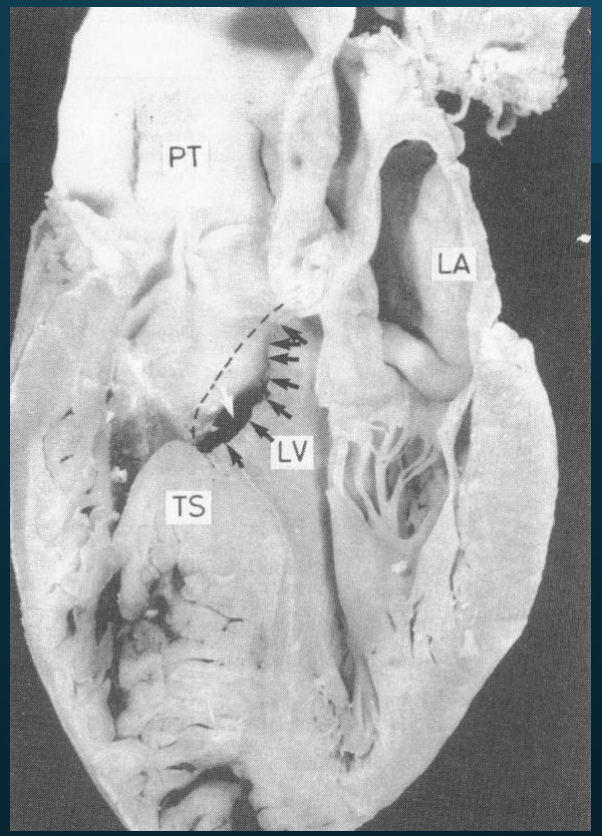
VGDI

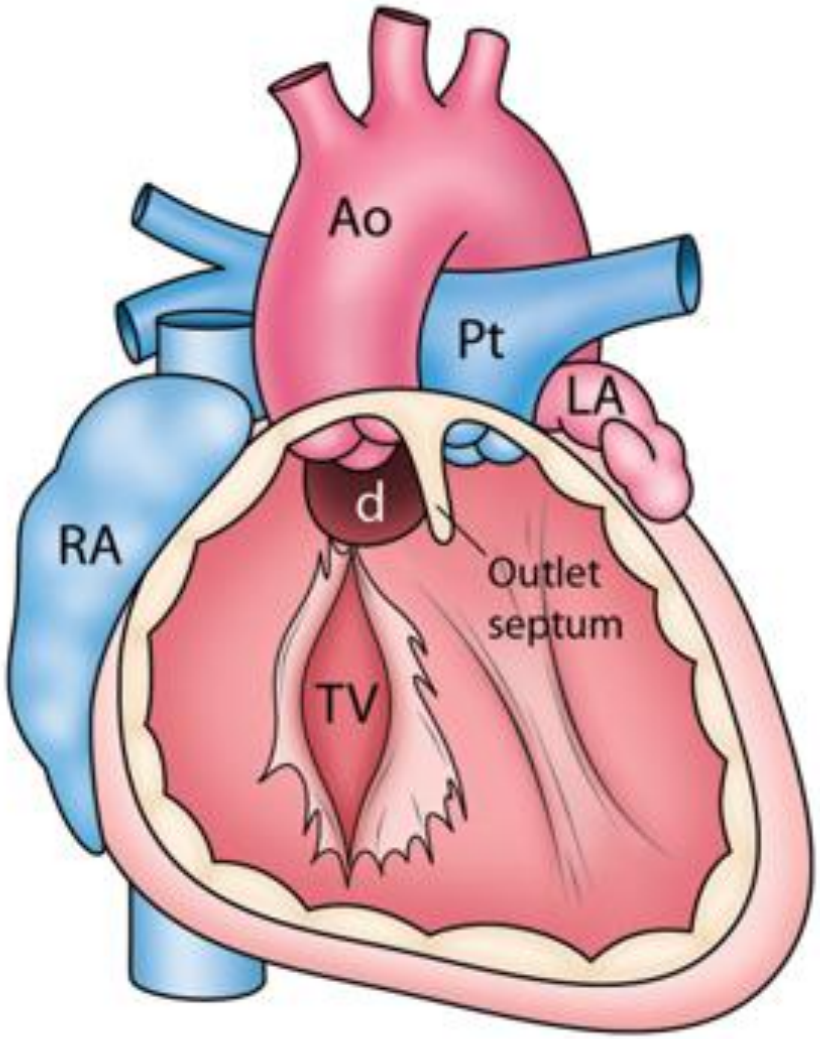
+/- Interruption de l’arche aortique

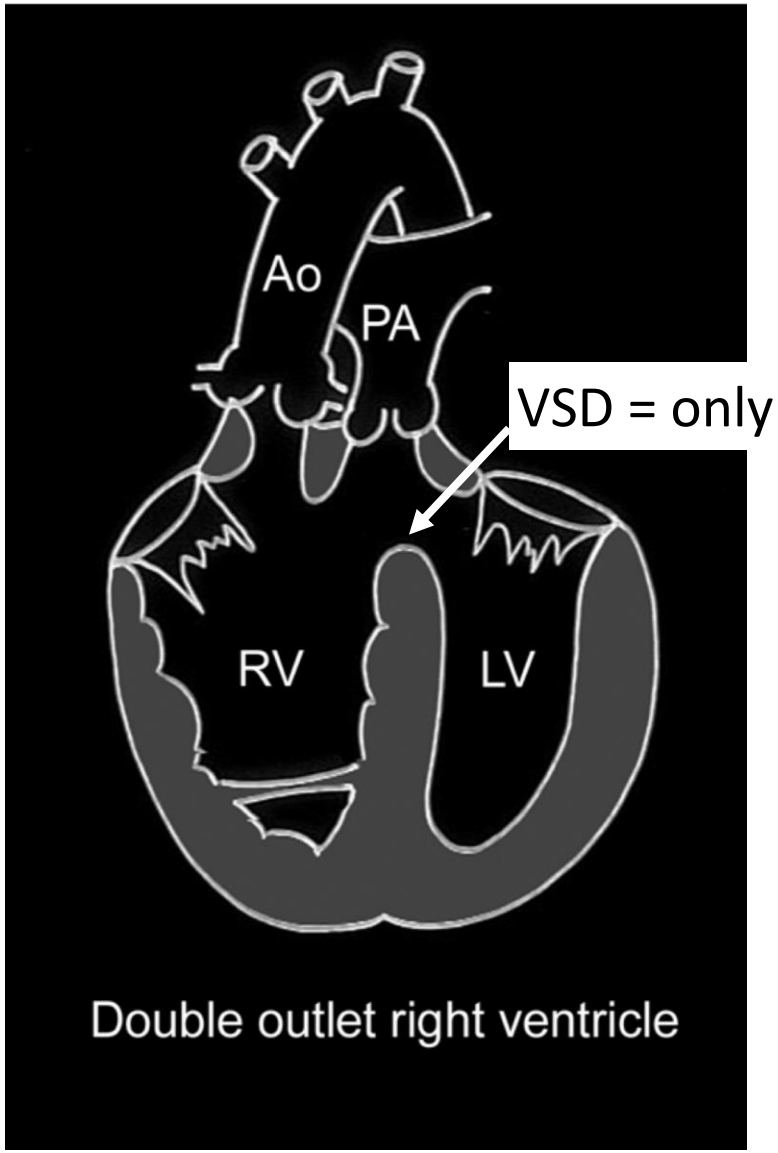
Discontinuite mitro-aortique/mitro-pulmonaire ??








Ao
Pt
LA
RA
d
Outlet
septum
TV


Ao
PA
VSD = only outlet of LV
RV
LV
Double outlet right ventricle

History & Classifications

- 1949: Taussig-Bing description
- 1957: Term DORV / Witham
- 1961: 14 cases series / Neufeld
- 1968: DORV first classification
- 1972: **Lev Classification**

Great Vessel Relation	Location of Ventricular Septal Defect: Subpulmonary	Subaortic	Subaortic and Subpulmonary	Remote	Total
Normal	0 (0†)	2 (0)	0 (0)	0 (0)	2 (0)
Side by side	4 (0)	28 (14)	2 (0)	5 (2)	39 (16)
Dextro-malposition	6 (3)	9 (7)	0 (0)	0 (0)	15 (10)
Levo-malposition	3 (2)	1 (1)	0 (0)	0 (0)	4 (3)
Total	13 (5)	40 (22)	2 (0)	5 (2)	60 (29)

Double Outlet Right Ventricle: Hemodynamic and Anatomic Correlations

July 1976 The American Journal of CARDIOLOGY Volume 38

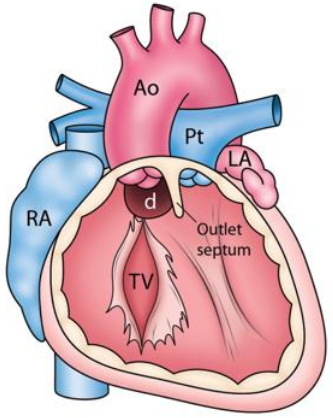
SOMKID SRIDAROMONT, MD*
ROBERT H. FELDT, MD, FACC
DONALD G. RITTER, MD, FACC
GEORGE D. DAVIS, MD
JESSE E. EDWARDS, MD, FACC

Rochester, Minneapolis and St. Paul, Minnesota

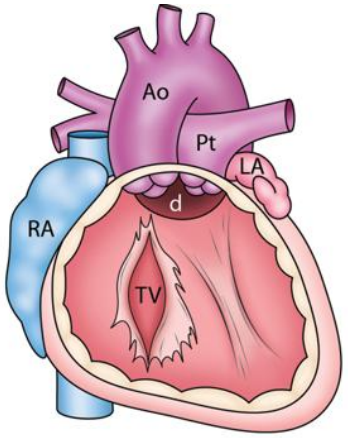
There ar[illegible] 16 possible variations [illegible]f double outlet right ventricle with regard to interrelations of the great arteries and to location of the ventricular septal defect. In a series of 62 cases, approximately two thirds of patients had the great arteries in a side by side relation, and most (28 of 41) had the ventricular septal defect in a subaortic position. In double outlet right ventricle with malposition of the great arteries, the ventricular septal defect was either subpulmonary or subaortic. Four of the 13 patients with subpulmonary ventricular septal defect had a supracristal defect with side by side relation of the great arteries (Taussig-Bing anomaly), and 9 patients had malposition of the great arteries with an infracristal ventricular septal defect. In all patients with subpulmonary ventricular septal defect,

Classifications

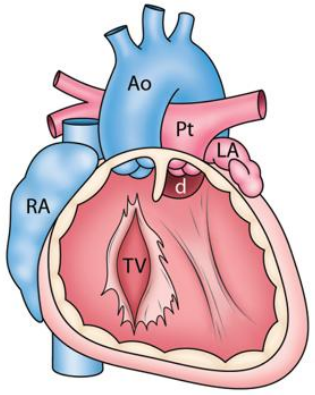
Lev Classification



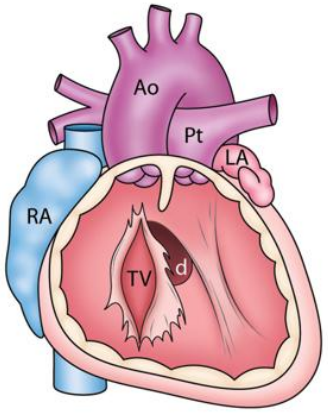
Subaortic defect



Doubly committed defect

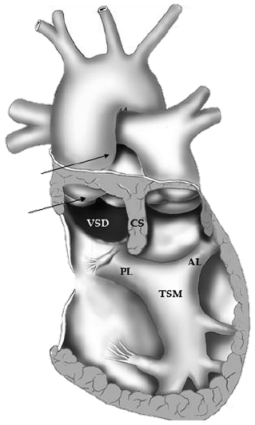


Subpulmonary defect

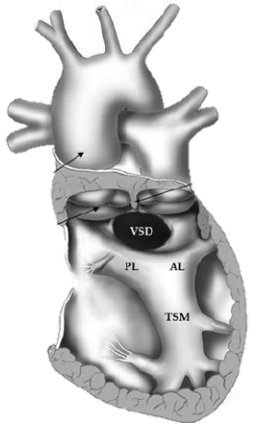


Remote defect

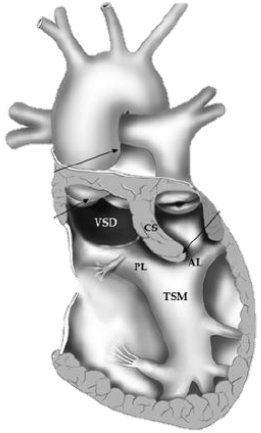
Lacour-Gayet



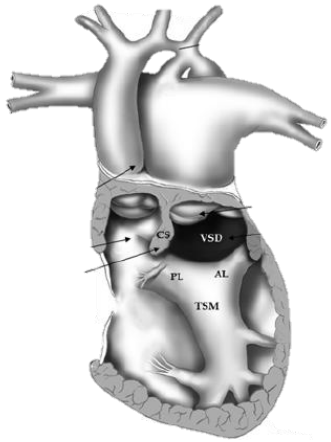
Subaortic



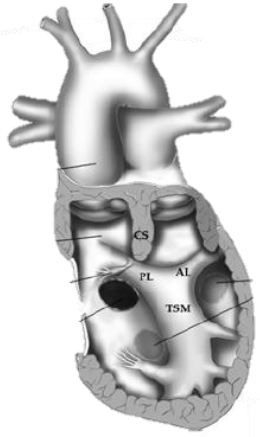
Doubly-committed



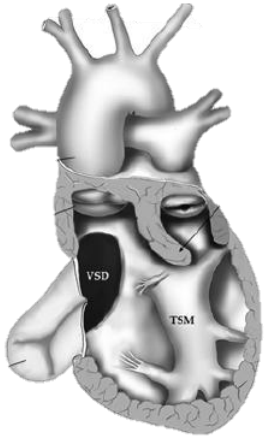
Fallot-type



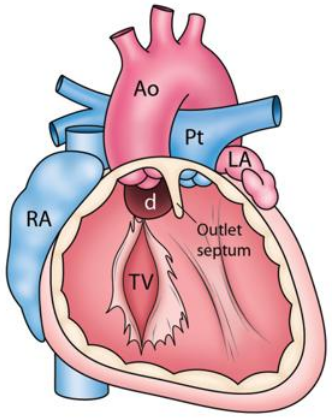
Subpulmonary/TGA



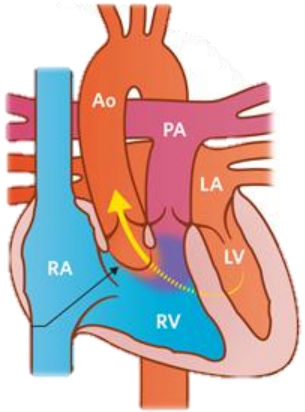
Non-committed

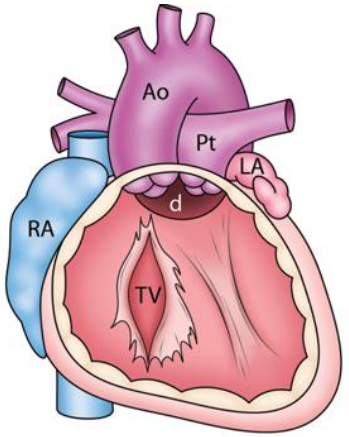


AVSD-type

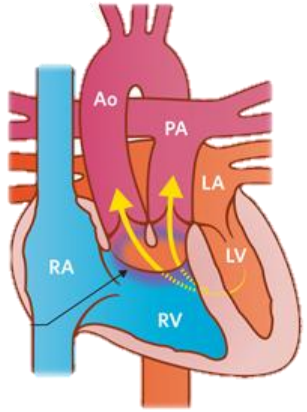


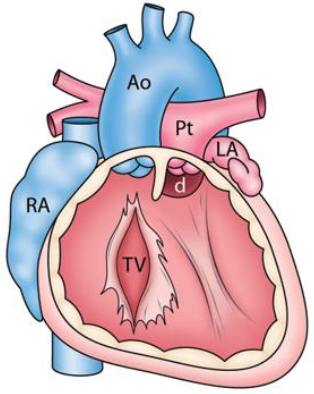
Subaortic defect



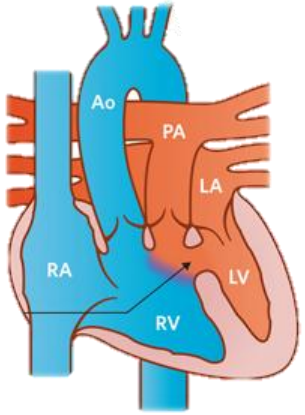


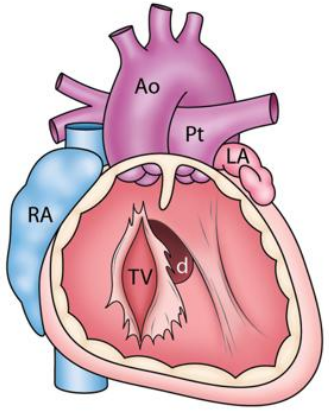
Doubly committed defect



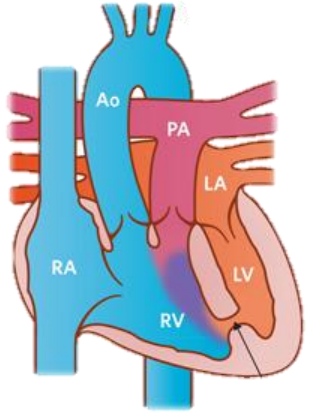


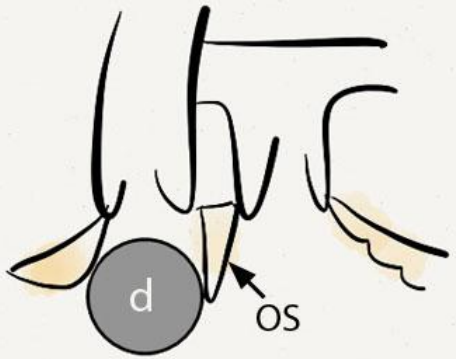
Subpulmonary defect



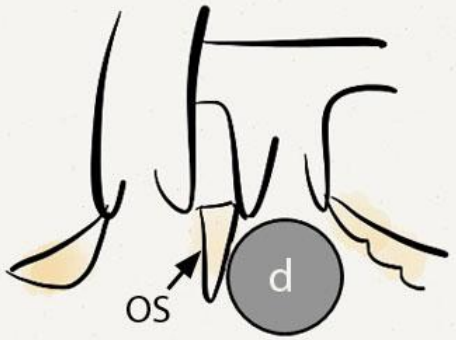


Remote defect



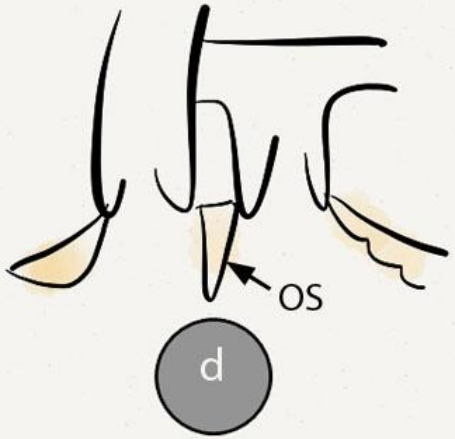


A. Outlet septum fused to the left margin of the VSD

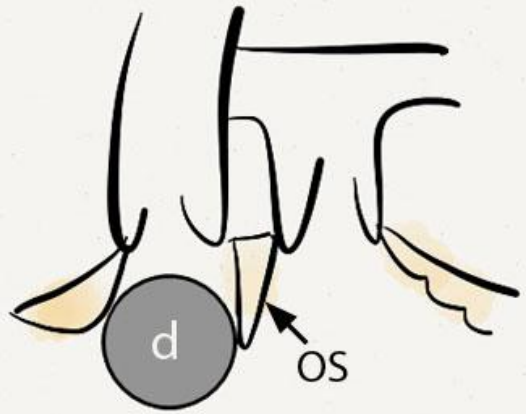


B. Outlet septum fused to the right margin of the VSD

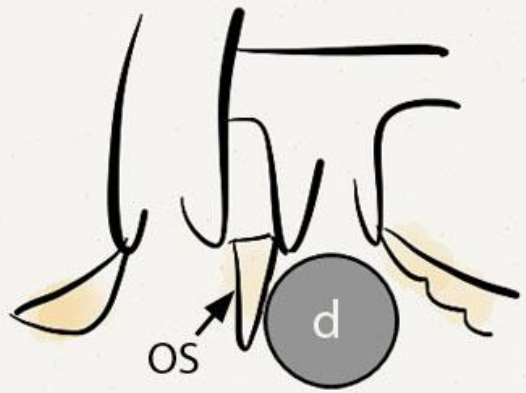
C. Outlet septum parallelwith the plane of the VSD



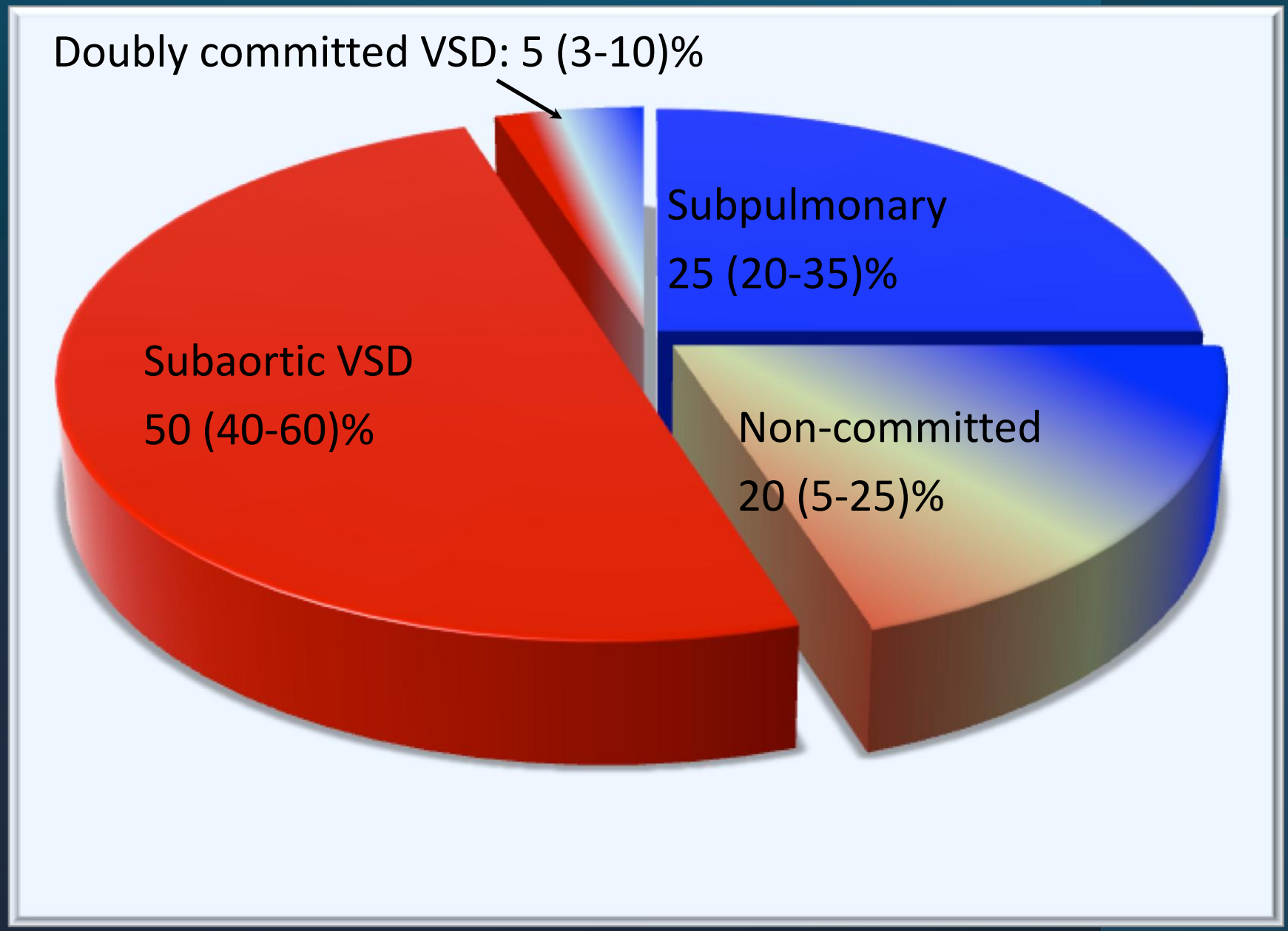
D. Outlet septum fused to neither side of the VSD



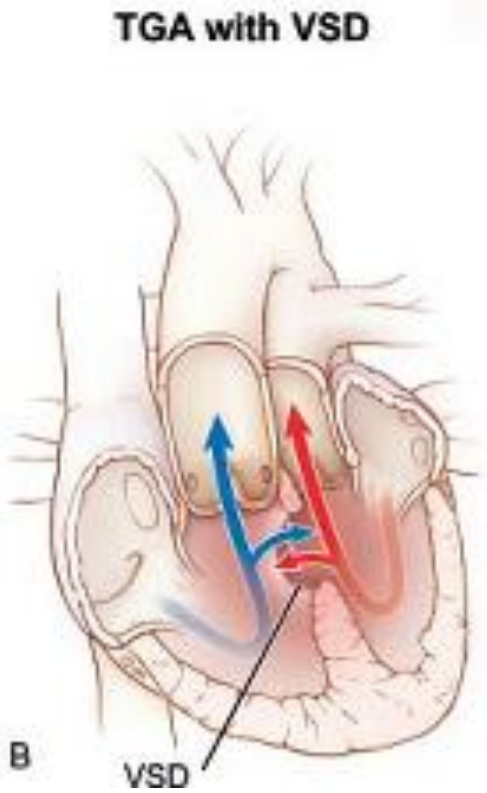
A. Outlet septum fused to the left margin of the VSD



B. Outlet septum fused to the right margin of the VSD

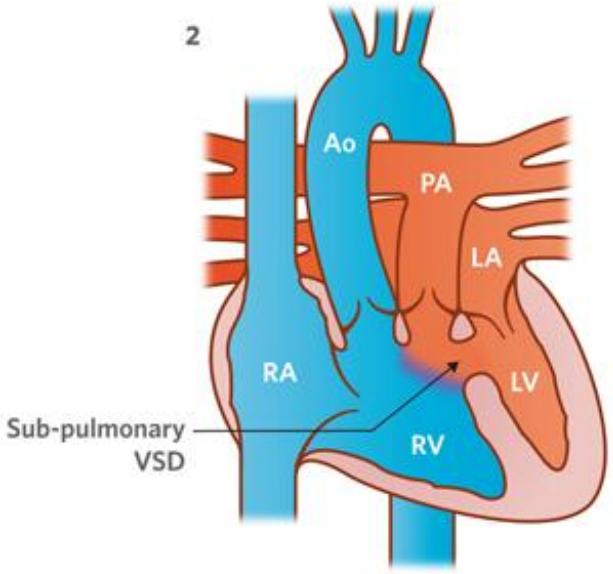

Doubly committed VSD: 5 (3-10)%
Subpulmonary
25 (20-35)%
Non-committed
20 (5-25)%
Subaortic VSD
50 (40-60)%

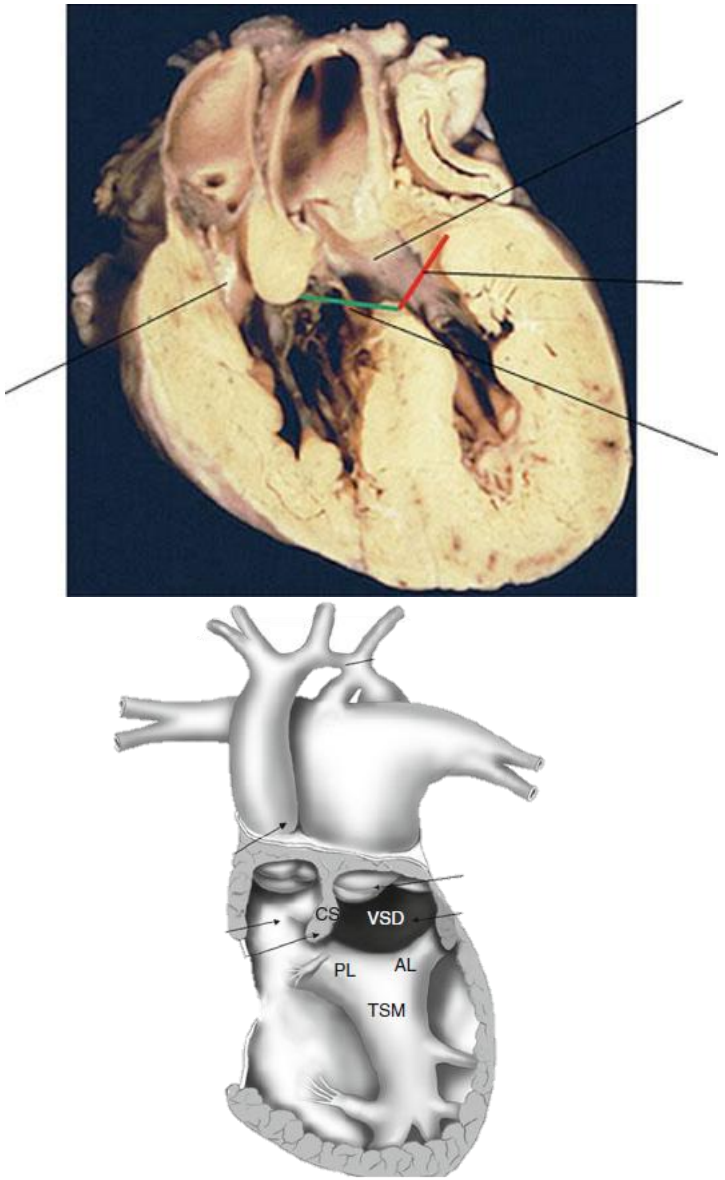
TGV+CIV ou VDDI ??



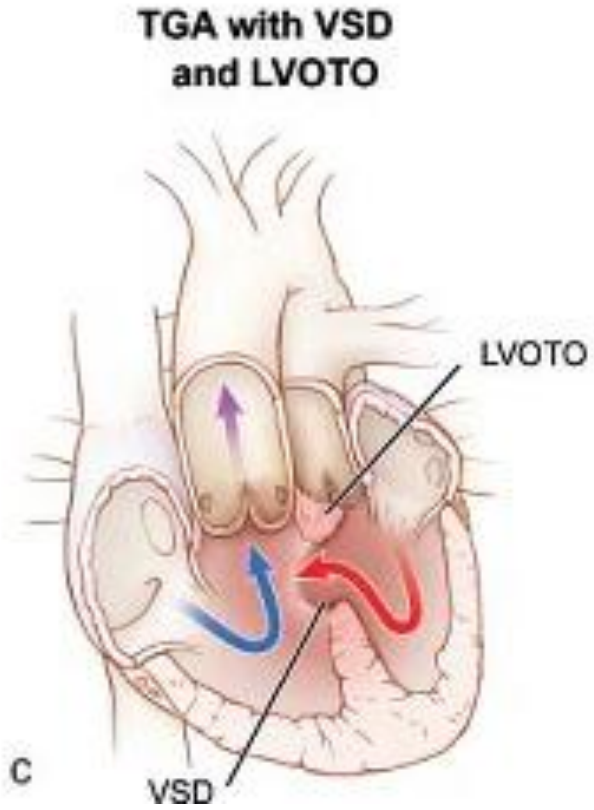
VS

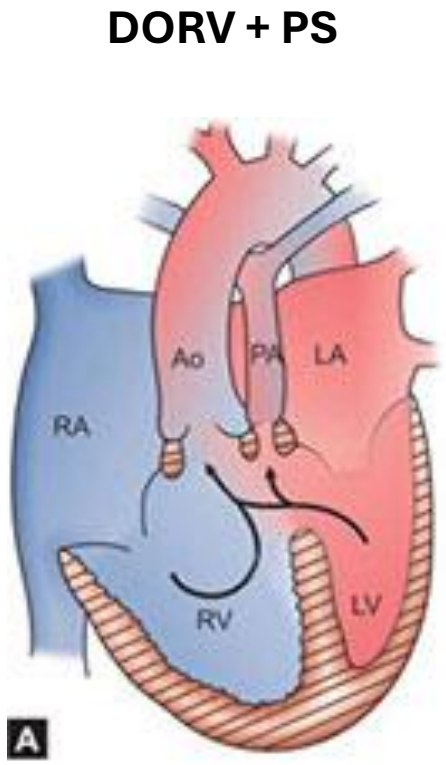
DORV-TGA type





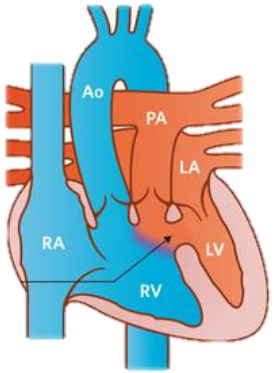
TGV-CIV-SP et VDDI-SP si différents ?





Ventricule Droit à Double Issue VDDI

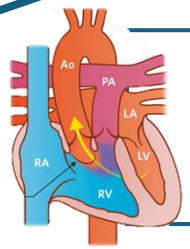
TGA-type ou Circulation systémique ducto-dépendante



Surgery during infancy

Primary repair ++

Staged repair



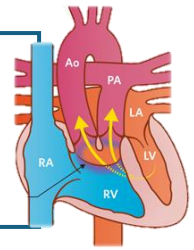
PAS de protection pulmonaire

Qp/Qs >2

Anatomie favorable (VSD-type, normally related GA)

Primary repair +/- neonatal

Protection pulmonaire (sténose, Fallot-type)



Qp/Qs **< 0.8**

Anatomie défavorable (CAV, tunnelisation difficile, abnormal CA, PA hypoplasia, VSDs)

Initial Palliation / staged repair

Banding

Shunt (Blalock) Stent canal/RVOT

Qp/Qs **≈1-1.5**

Equilibré / asymptomatique

Repair at 3-12 months

Techniques chirurgicales

Types d’intervention

- Palliation

Chirurgie d’attente

Croissance

Shunt

Banding AP

Stent canal

Stent RVOT

Glenn

- Réparation

Biventriculaire

IVR

Rastelli

REV

DREAM

Baffle + Switch

Root translocation Nikaidoh

Double Root Translocation

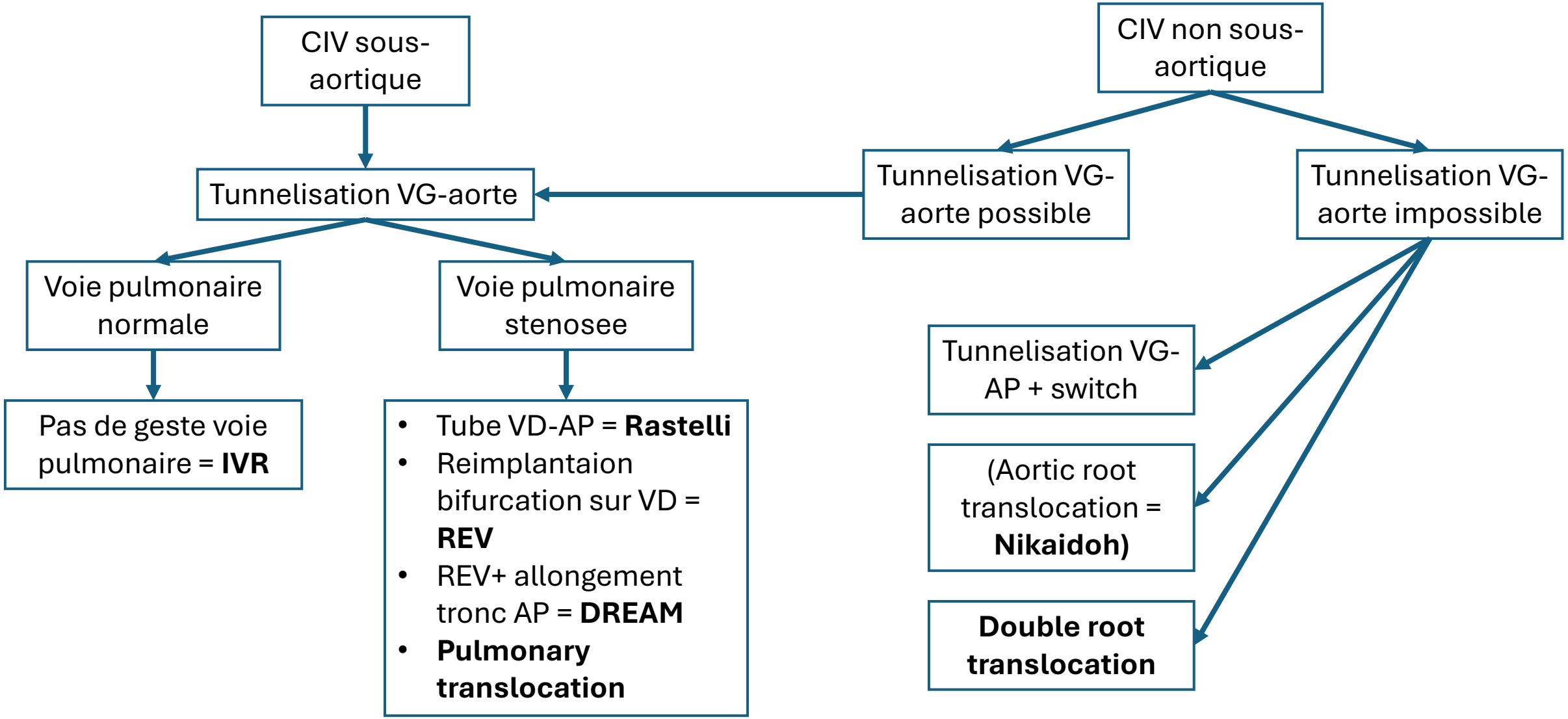
- Fontan

2 ventricules septables

2 VAV

1 Outlet

Reparations BiVentriculaires



Prerequis pour la strategie

1. The spatial relationship of the VSD to the septal leaflet of the tricuspid valve, and AVV abnormalities

2. The size of VSD

3. The presence and extent of the muscular infundibulum

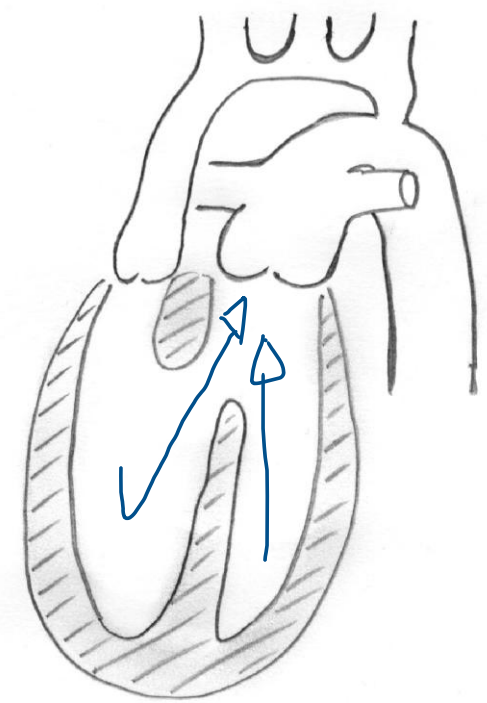
4. The orientation of the conal septum in relation to the VSD margin

5. The volume/size of ventricles

6. Any associated abnormalities (such as coronary artery anomalies, arch abnormalities)

Focus on Taussig-Bing = Subpulmonary type

→ If no RVOTO present, then TGA physiology



Preferential Flow in Main PA
Malaligned outlet septum
Risk of Sub-Aortic stenosis

Arch Hypoplasia and CoA 50%

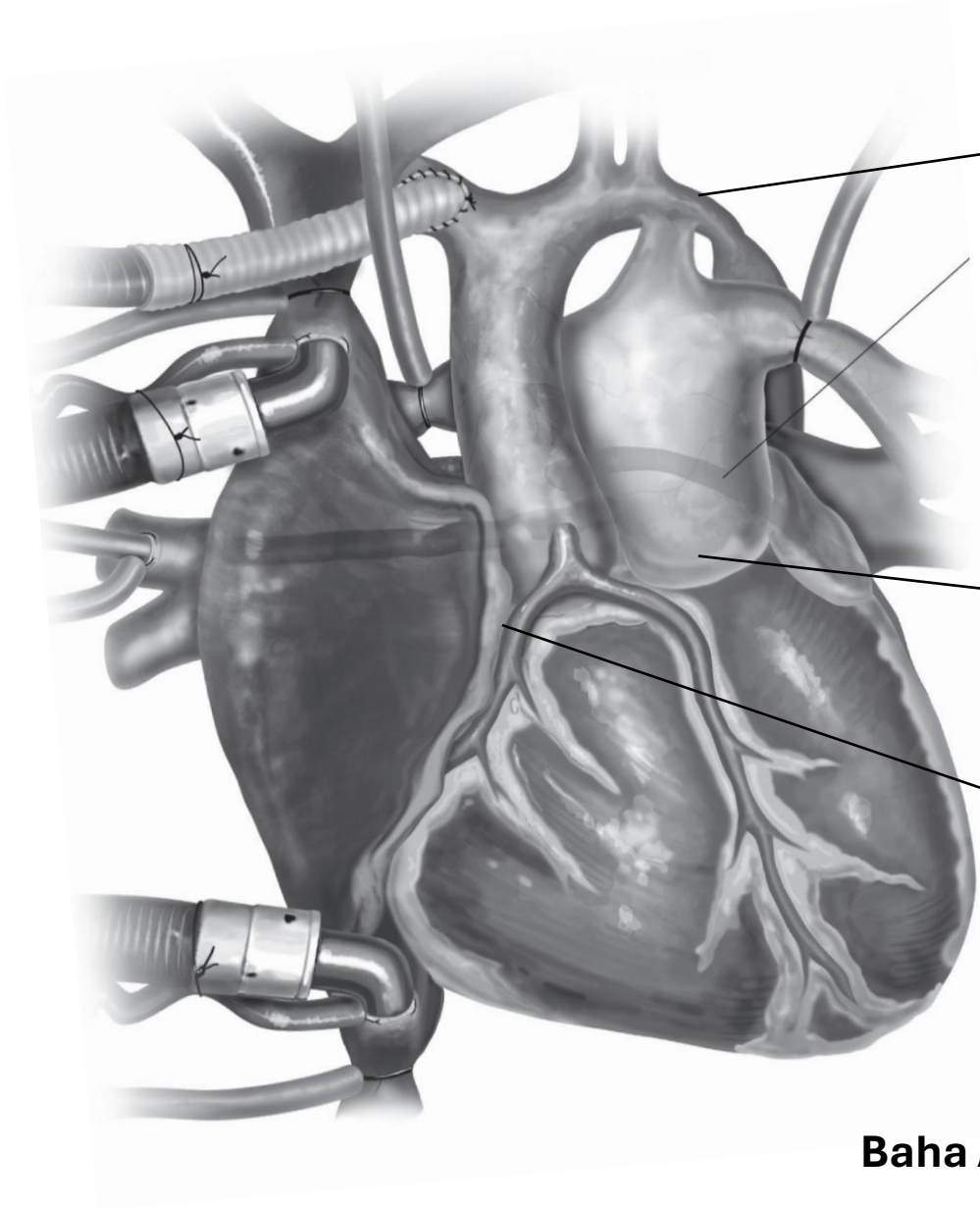
Abnormal Coronary Patterns in 50-60%

Need for Early Repair

Staged repair is an option but

- **-cyanosis**
- **-subaortic stenosis**
- **-risk of biventricular outflow obstruction**

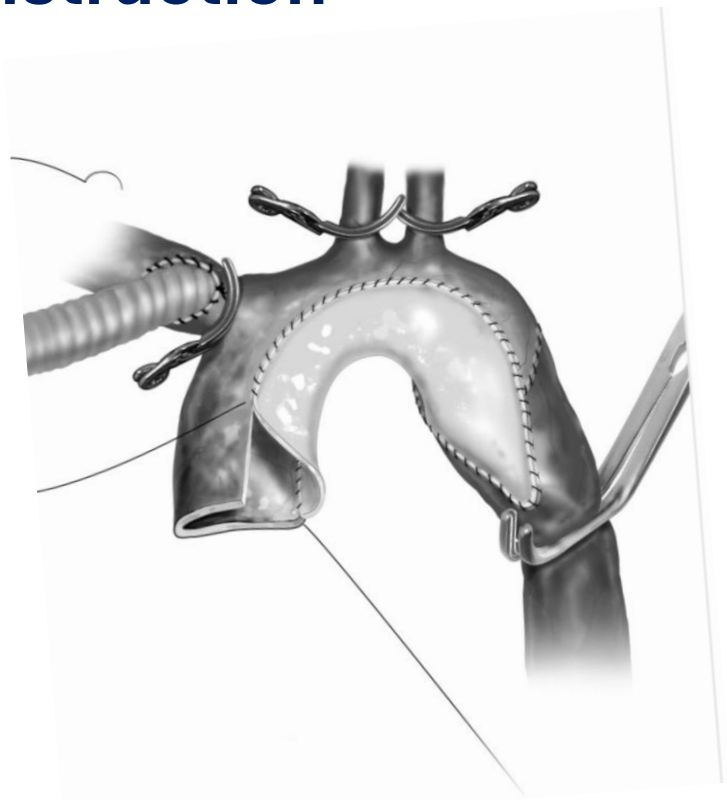
The Arterial Switch in Taussig- Bing

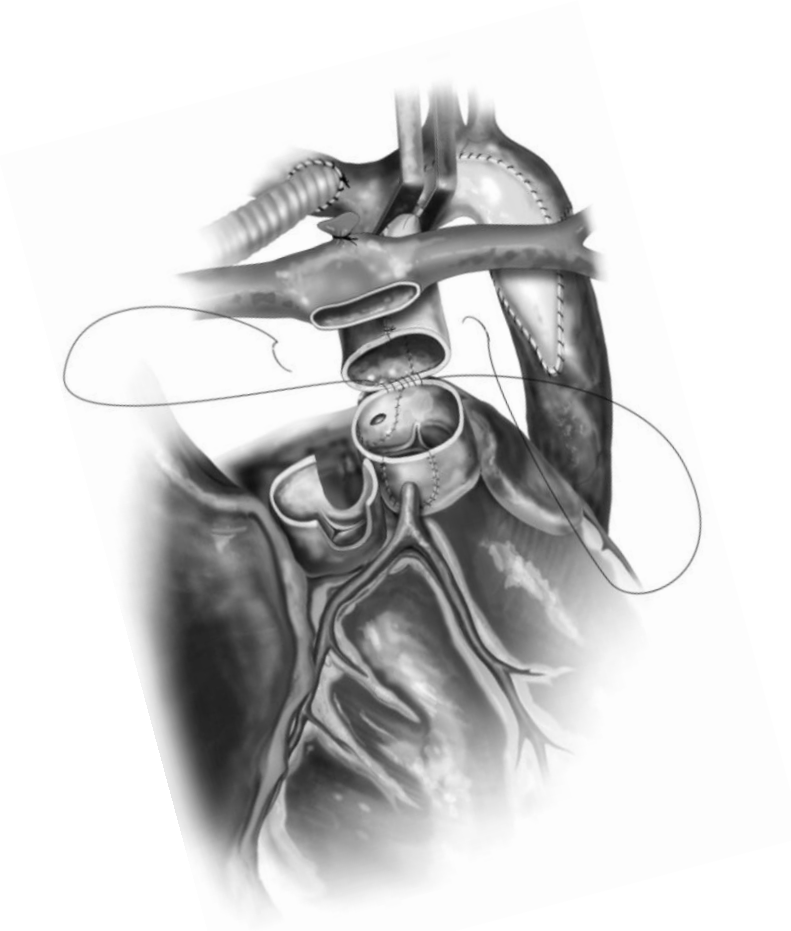


VSD Closure:
Via RA 60%
Via PA 30%
Via ventriculotomy 10%

Baha Alsoufi Op Tech CVS 4: 464 2015

DORV-TGA Type: Arch Reconstruction



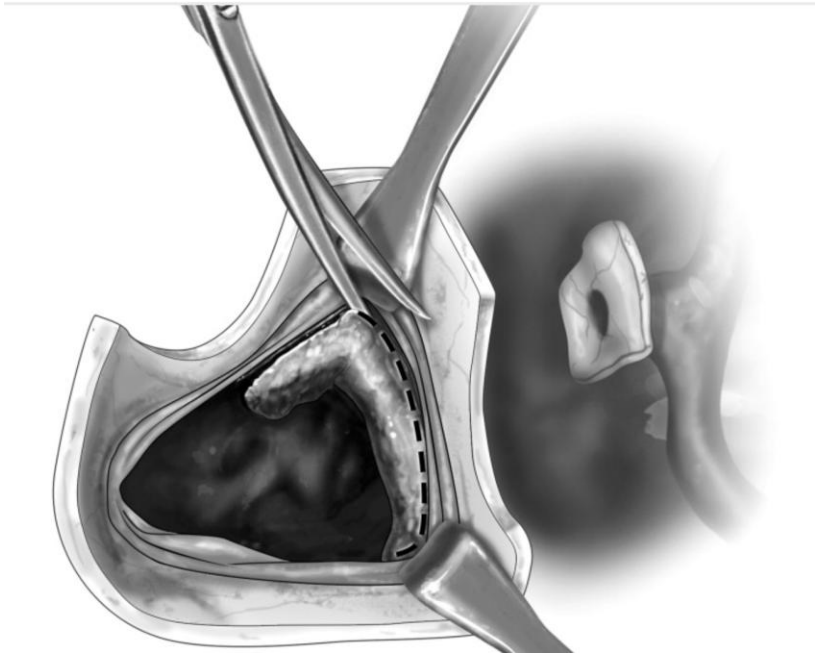


Op Tech CVS 4: 464 2015

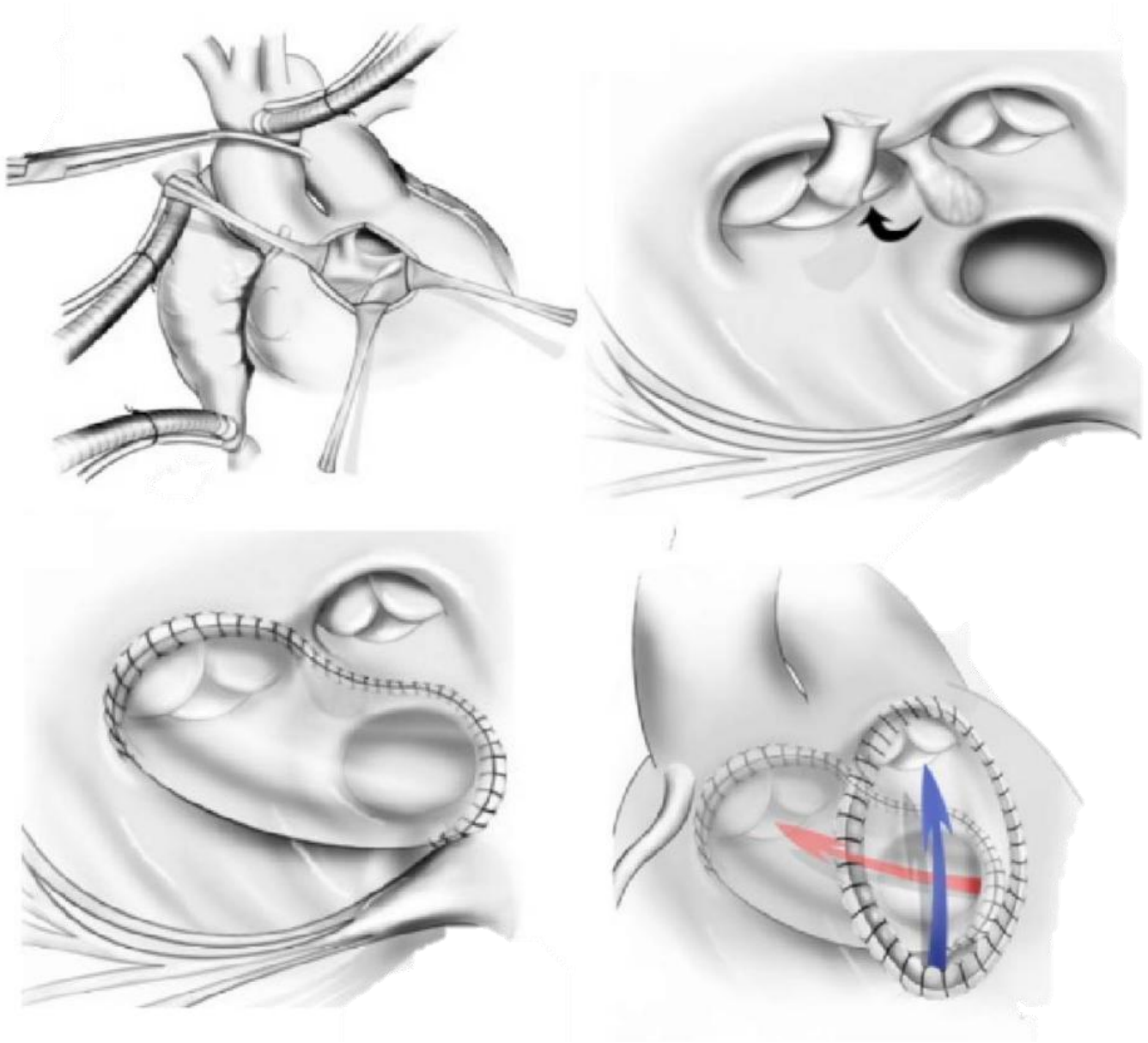
DORV-TGA Type: RVOT

Importance of assessing RVOT

Resect any muscle bars or deviated outlet septum

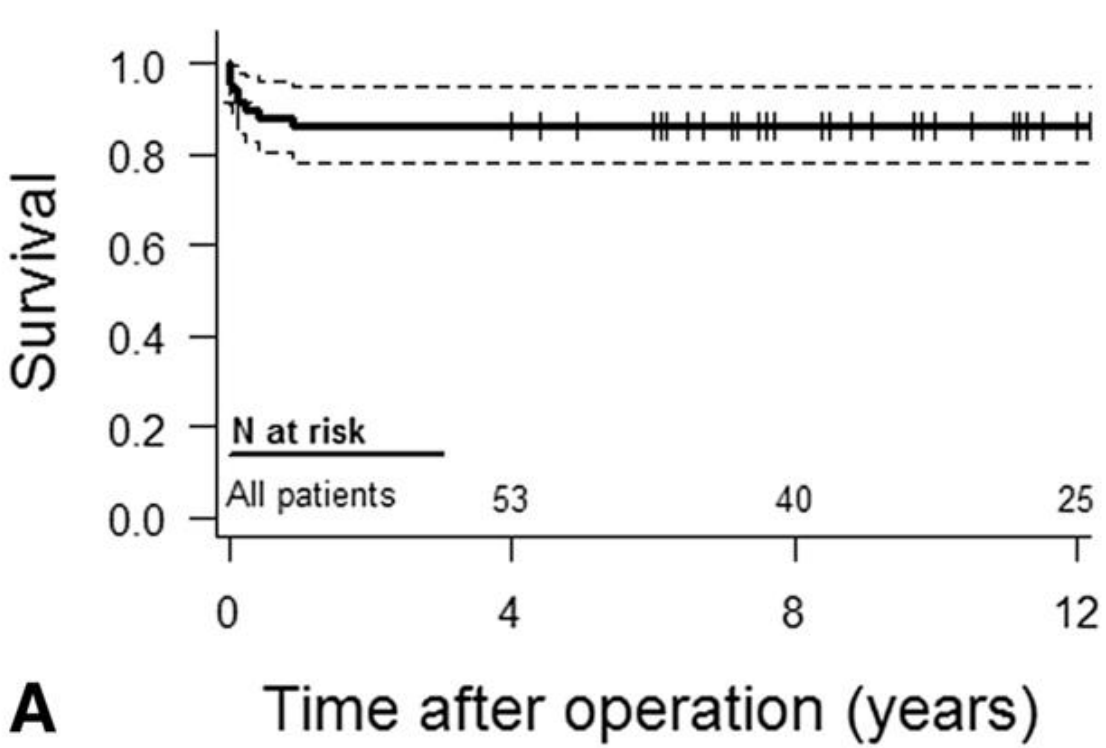


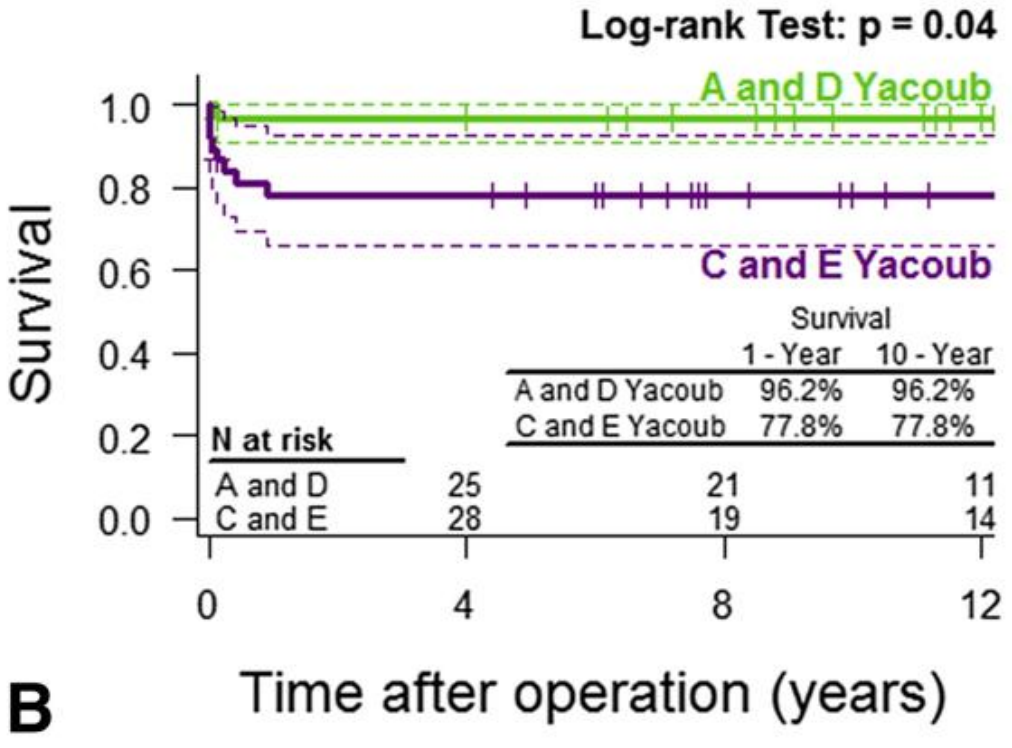
DORV-TGA Type: Kawashima repair



DORV TGA-type: Outcomes after Arterial Switch

Marie-Lannelongue, 1997-2010





Most Studies report Early Mortality 6-11%

Almost all associated with intramural or Type E coronary patterns
Almost all risk is within first year

JTCVS 149: 1124, 2015

Taussig Bing: Staged vs Complete Repair

Staged ??

Taussig Bing: Staged vs Complete Repair

Initial Arch Repair and PA Band

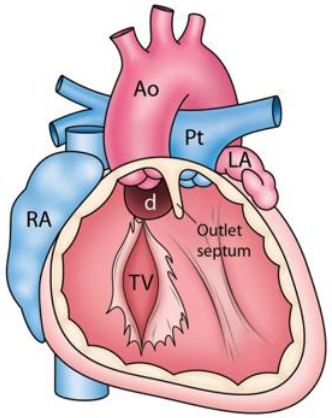
Remains an option: Risks are: Persistent cyanosis
May not address the sub-aortic stenosis/hypoplasia
May lead to biventricular outflow tract obstruction

Most studies have shown better outcomes with complete single-stage repair

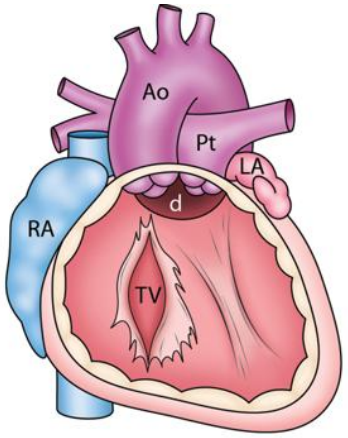
Still consider if: Multiple VSDs
Straddling MV
Concern over size of RV

Classifications

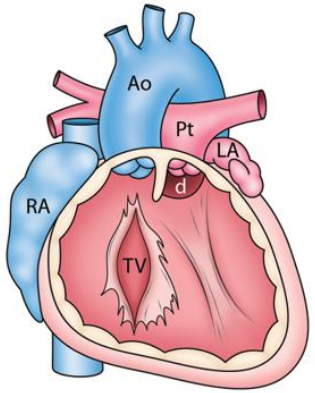
Lev Classification



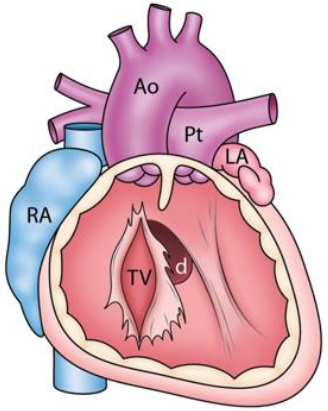
Subaortic defect



Doubly committed defect

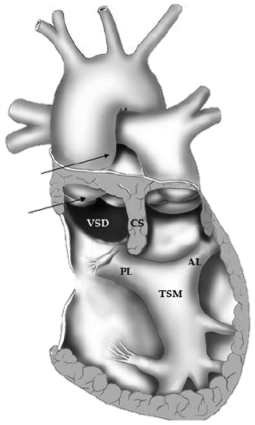


Subpulmonary defect

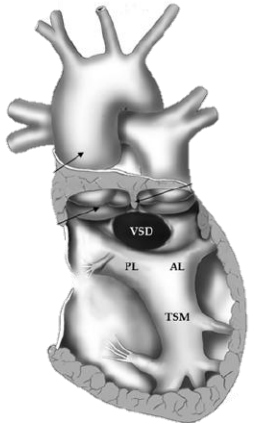


Remote defect

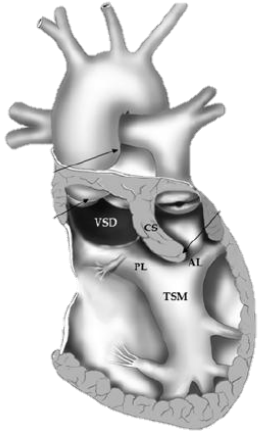
Lacour-Gayet



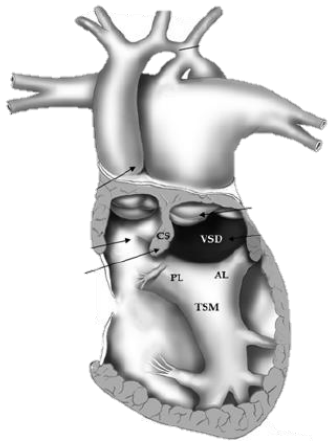
Subaortic



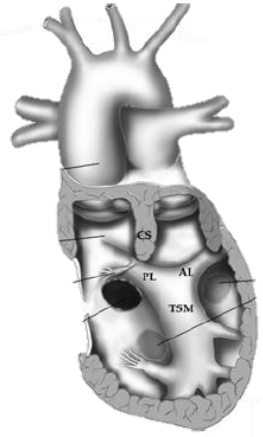
Doubly-committed



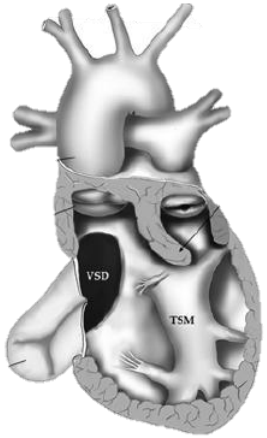
Fallot-type



Subpulmonary/TGA

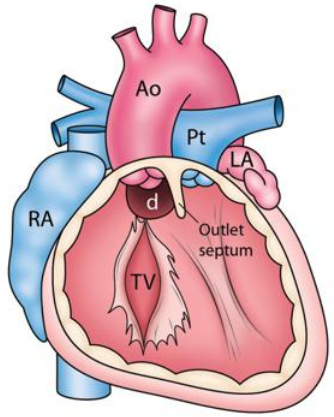


Non-committed

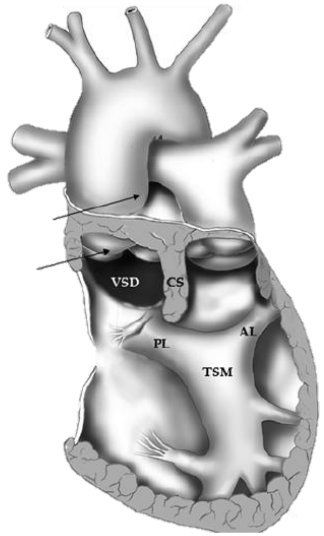


AVSD-type

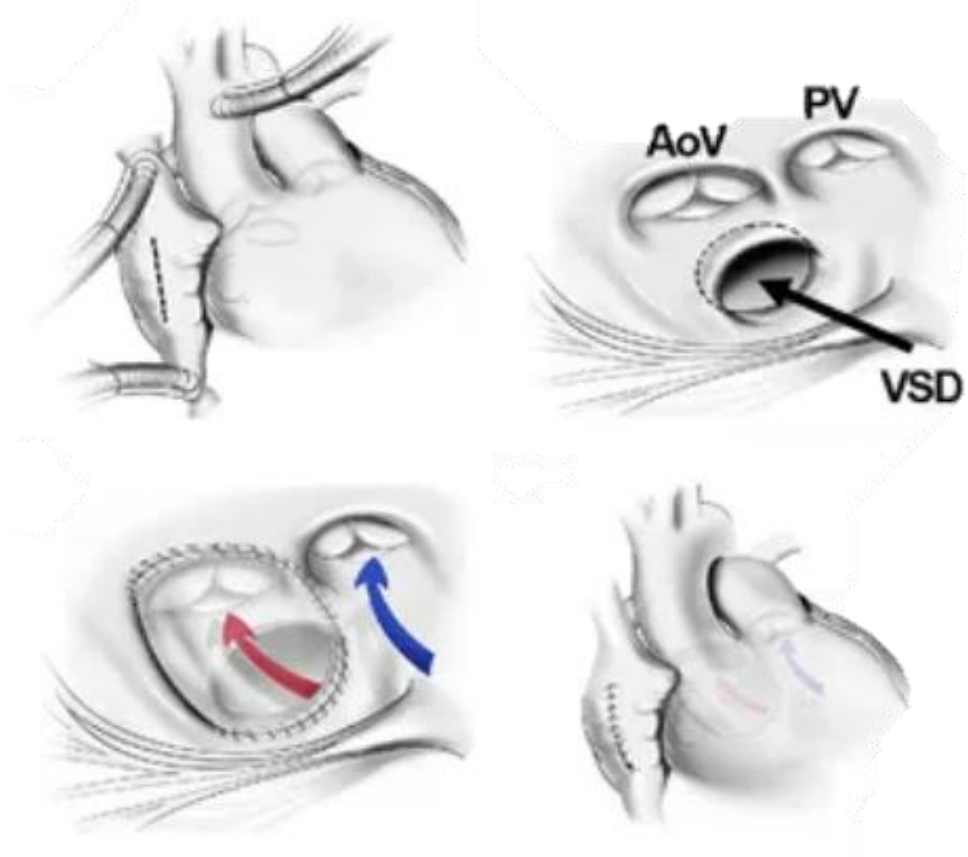
VSD-type



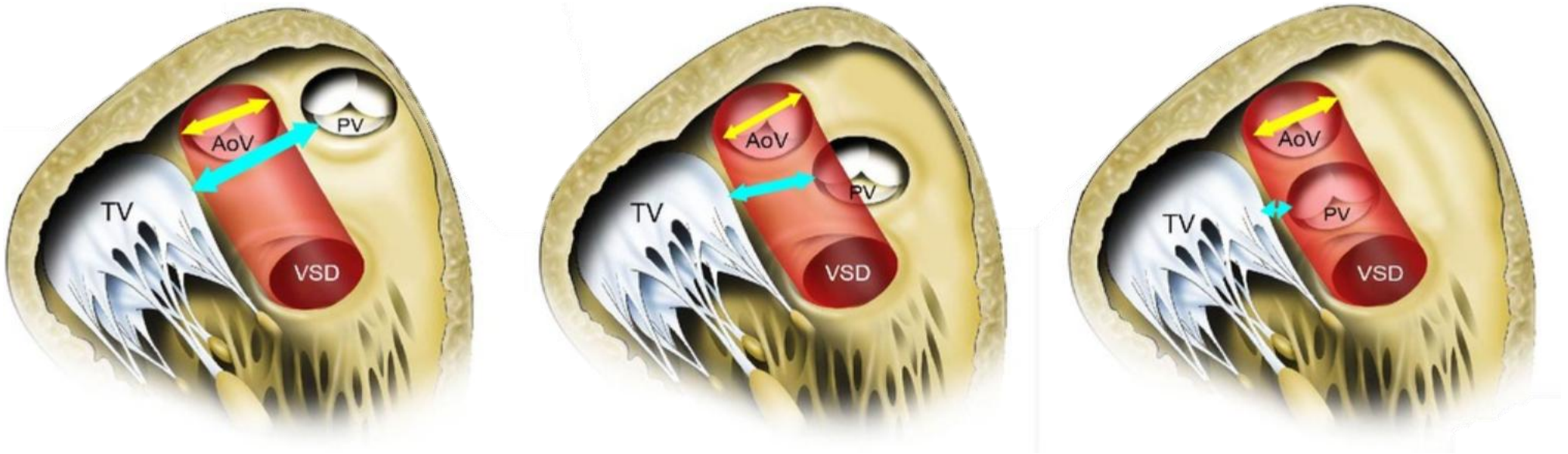
Subaortic defect



Intra Ventricular Repair

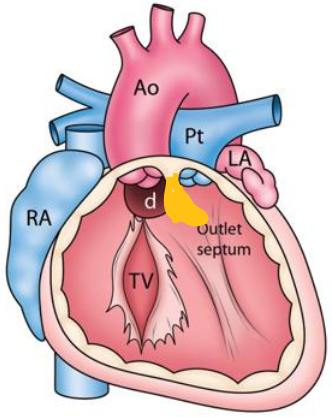


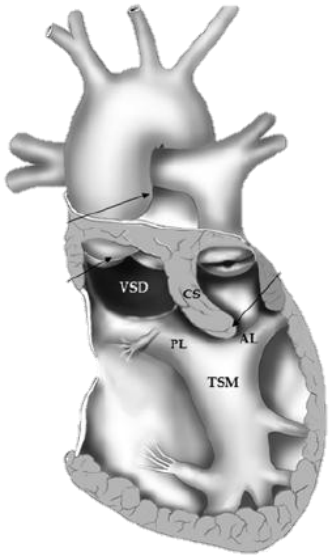
Distance tricuspido-pulmonaire



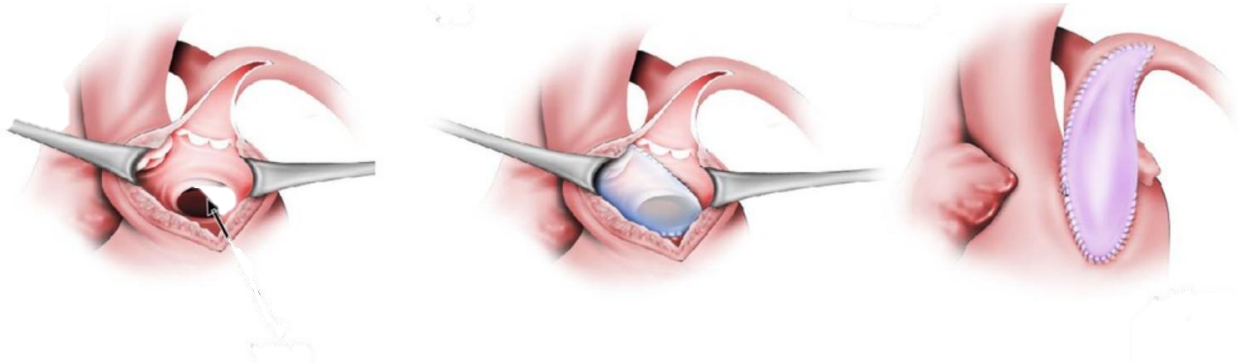
→ **Risk of RVOT Obstruction**

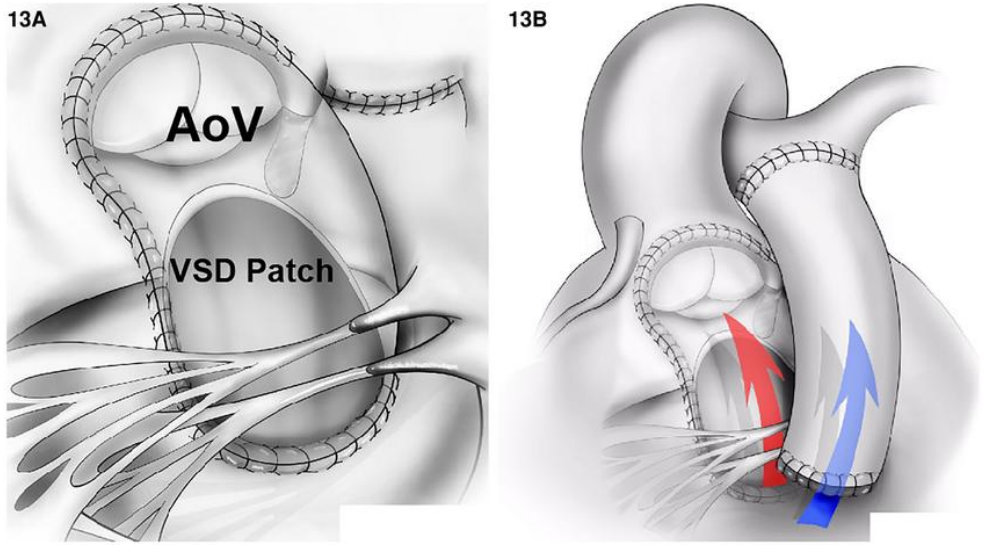
VSD Fallot-type





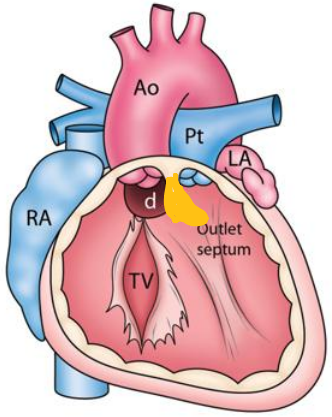
LV-Aorta baffle +
Transannular patch

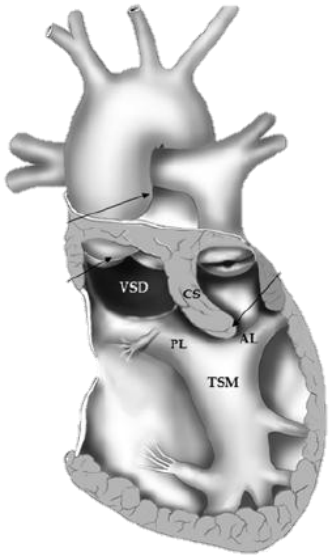


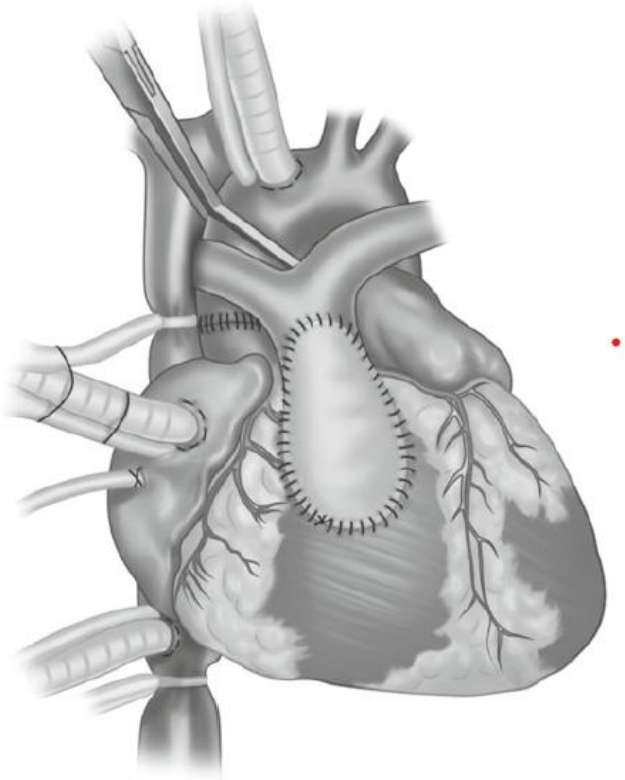


Rastelli

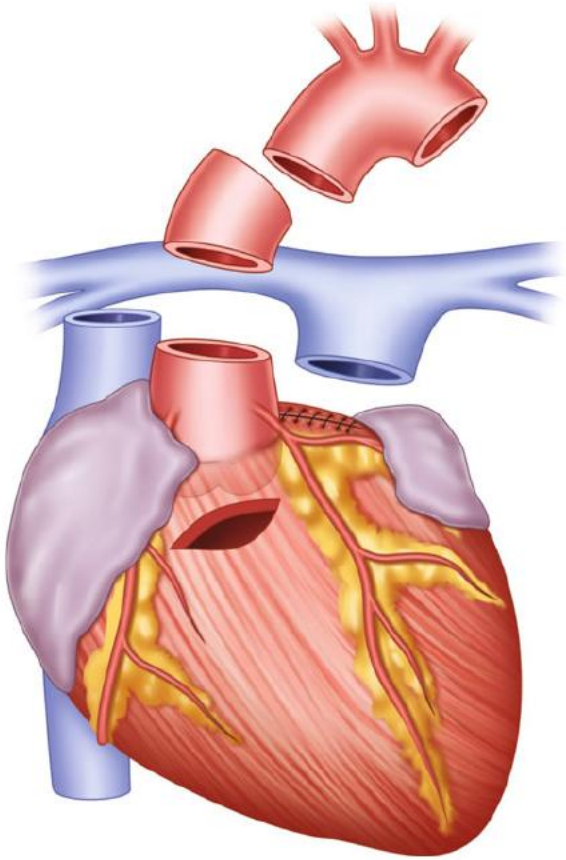
VSD Fallot-type







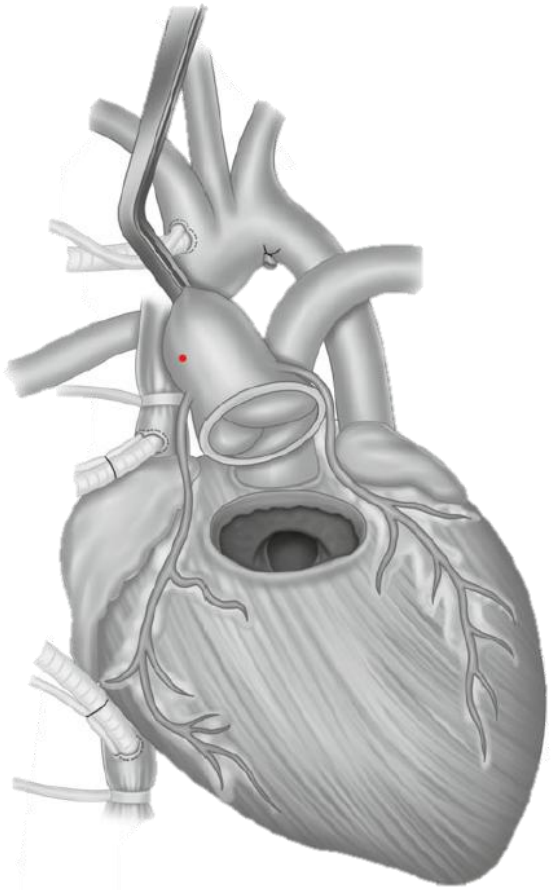
REV procedure

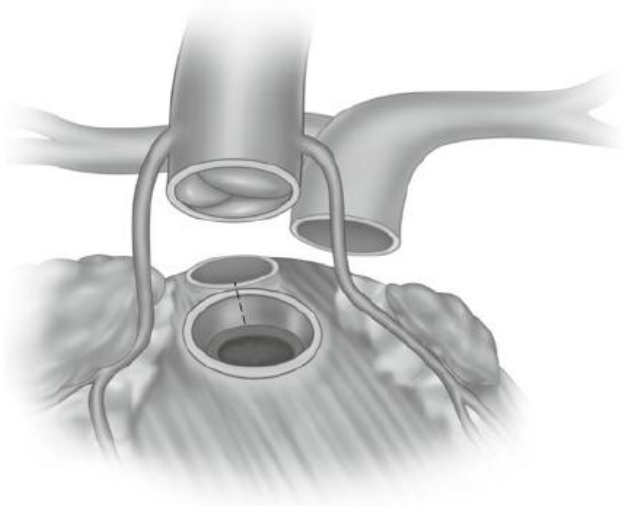


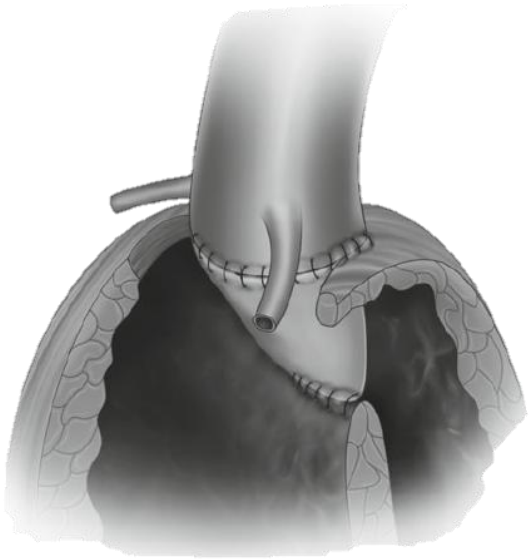
DREAM procedure

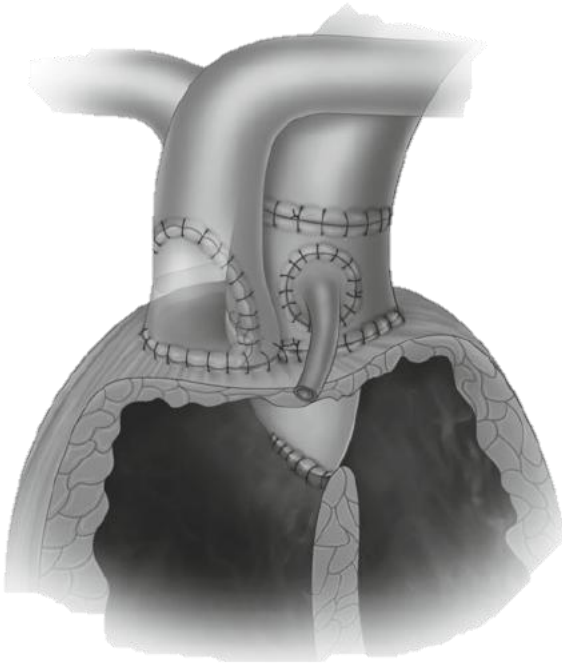
+ Pulmonary translocation

DORV TGA-type + PS
Nikaidoh procedure = Aortic translocation



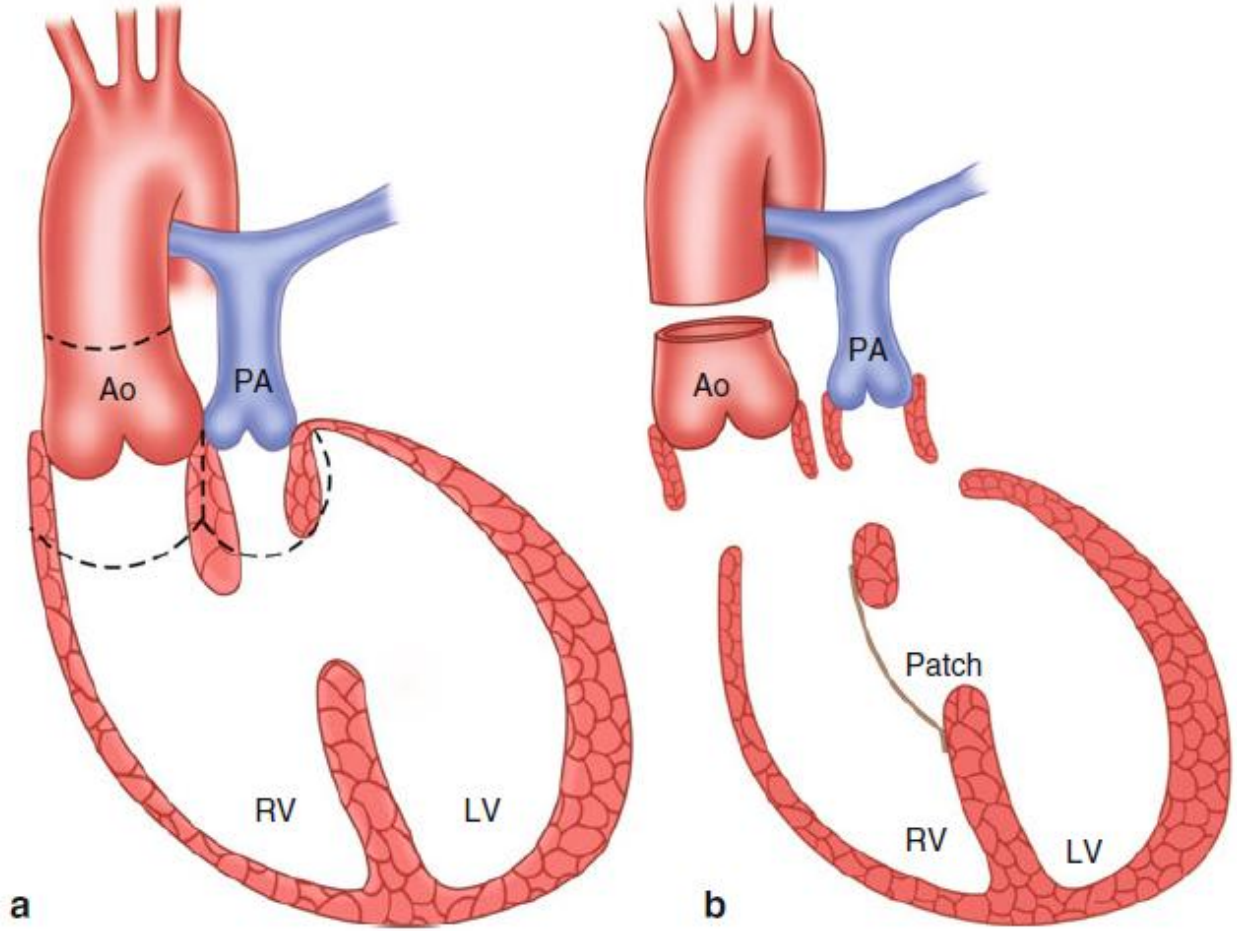


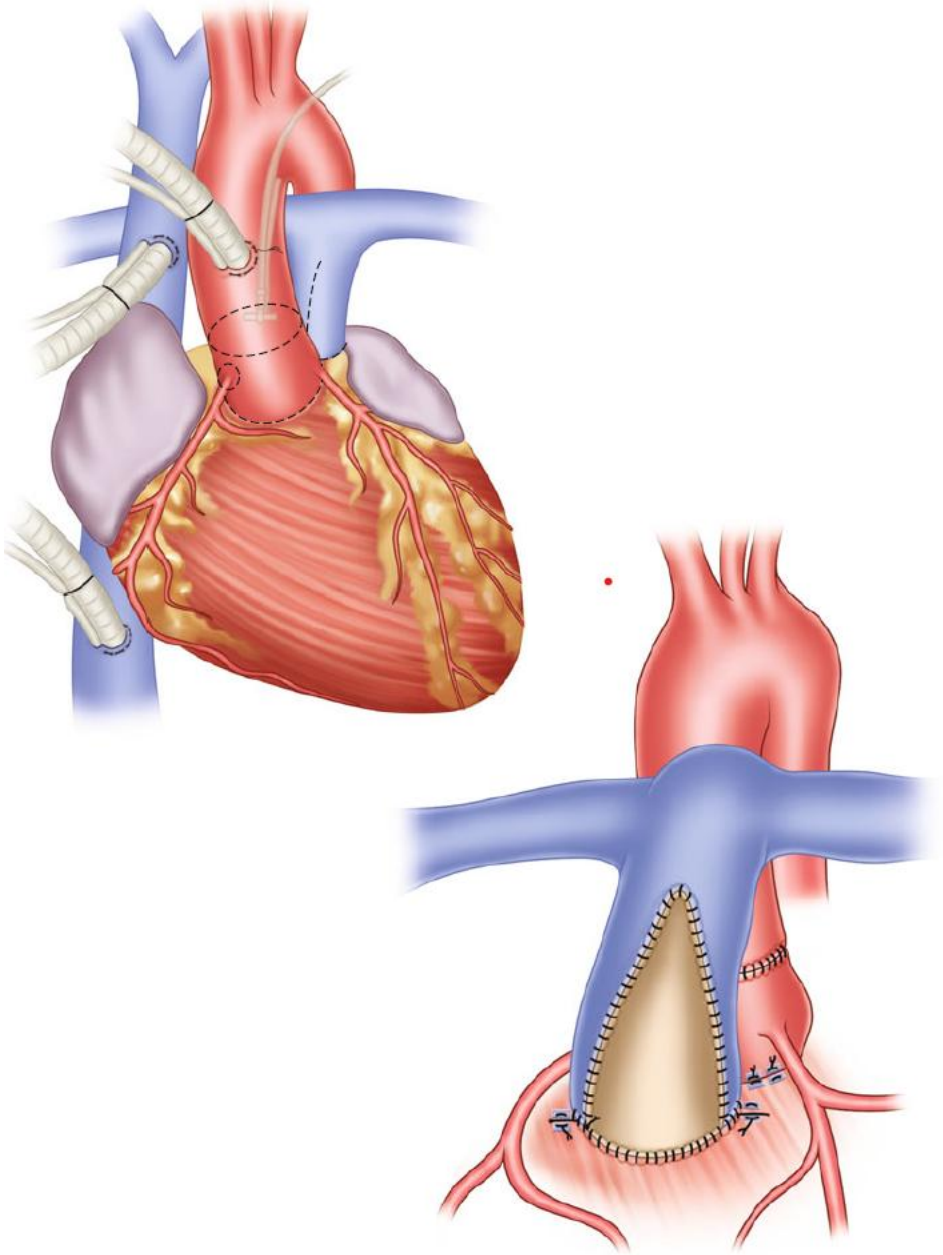




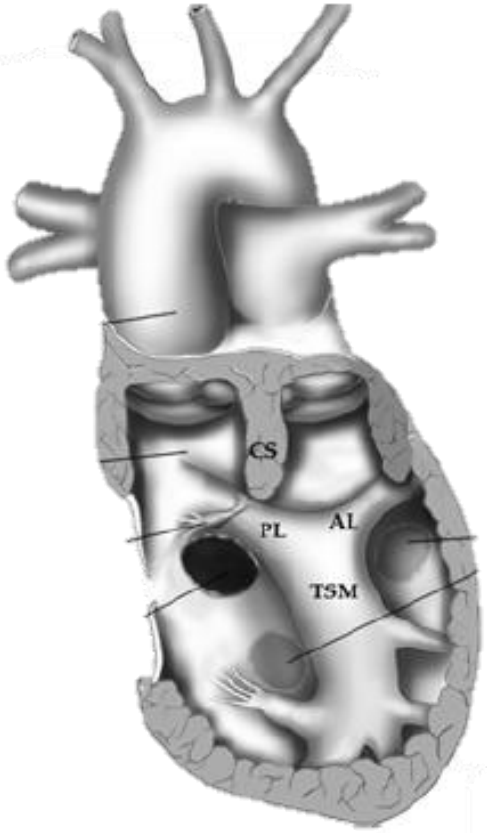
RVOT = Conduit or direct implantation

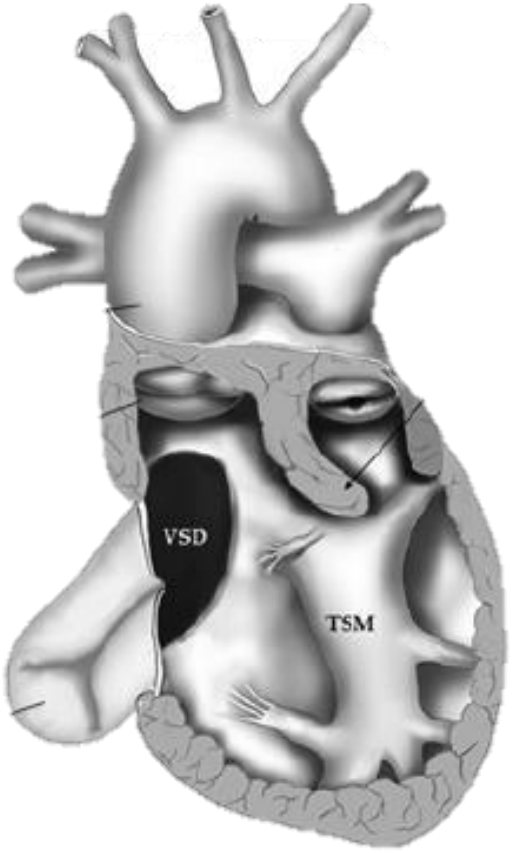
DORV TGA-type + PS
Double root translocation





Cas + rares



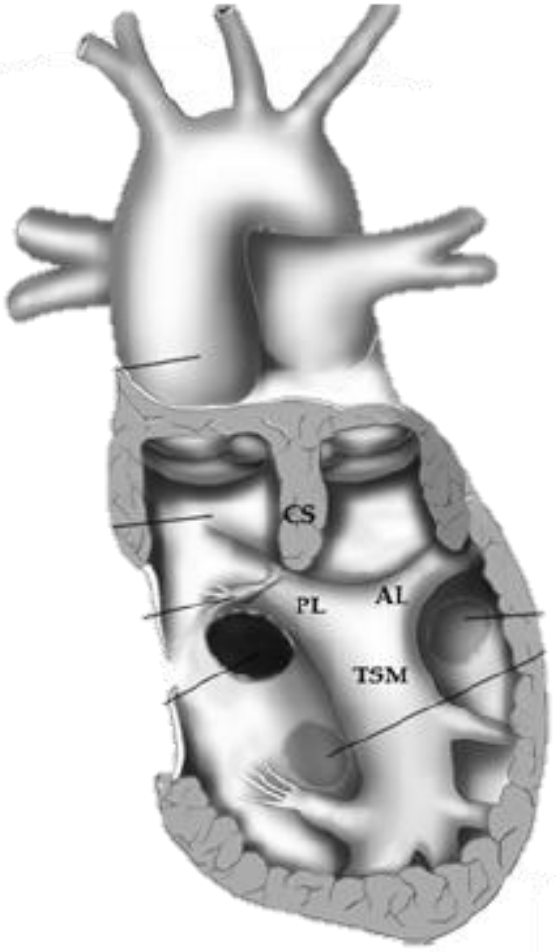


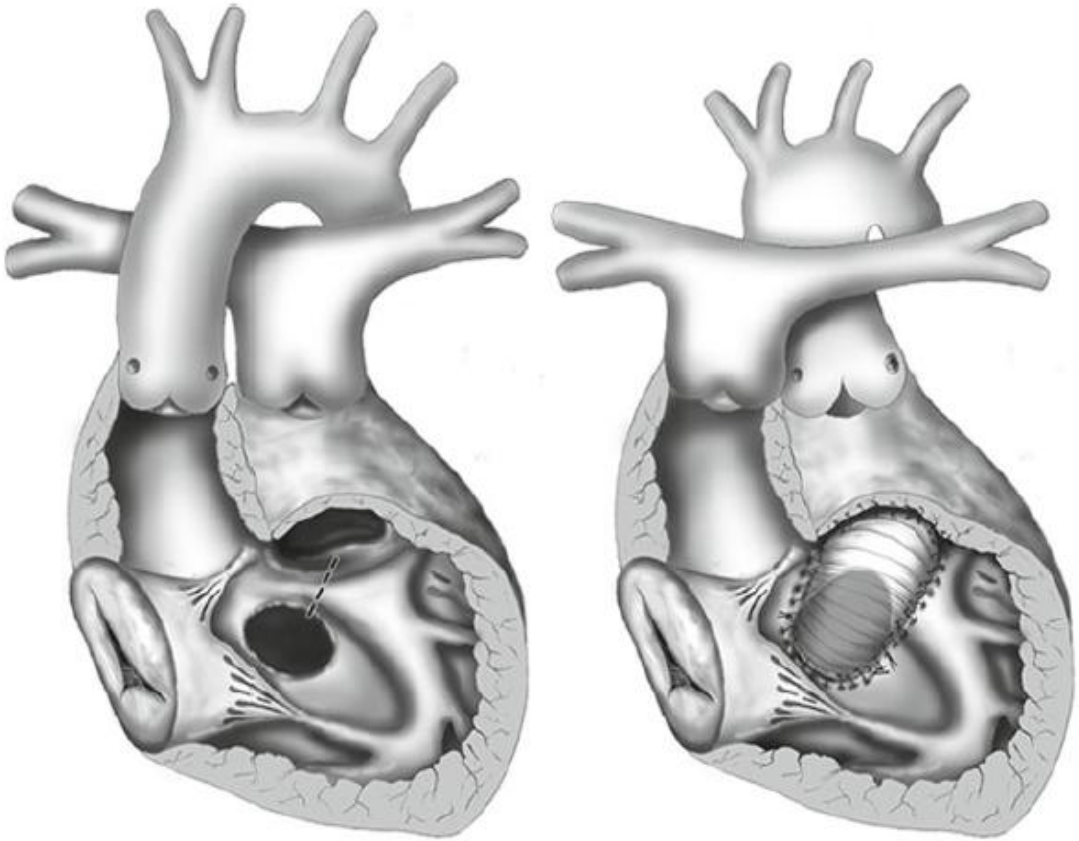
VDDI non-committed

Straddling VAV

VDDI-CAV Hétérotaxie Sténose pulmonaire RVPAt Retour azygos

Non-committed / remote VSD





Tunnelization + arterial switch

Univentricular pathway = Fontan

- Pour une réparation **Biventriculaire** il faut:
  - 2 Ventricules de bonne taille septables (tunnelisation possible)
  - 2 VAV
- Sinon : Fontan / Dans l'absolu VDDI avec 2 ventricules bons candidats car … 2 ventricules
- Attention risque obstacle sous aortique → élargissement CIV / DKS
- Alternative
  - 1.5 Ventricle repair

Timing de la réparation ?

- Physiologie de TGV et/ou symptomatique : chirurgie néonatale
- Anatomie favorable : a partir de 3 mois (ideal 6-10kgs)
- Anatomie défavorable/ complexe :
  - Besoin de + de temps
  - Palliation d'attente si besoin
    - Nouveau Shunt
    - Glenn/DCPP
  - Biventriculaire impossible → Fontan

Resume

Classification des DORV

- VSD-type
- TGA-type
- Fallot-type
- Doubly-committed
- Non-committed

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Fréquence : 70-80% = VSD or TGA=type
<20% formes très complexes (non-committed)

Timing de la chirurgie

Types de réparation

IVR / Fallot-like / REV / DREAM / Rastelli / Baffle+switch / aortic root translocation

Fontan si pas de réparation possible