Double outlet right ventricle

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Malformations Cardiaques Congénitales Complexes-M3C

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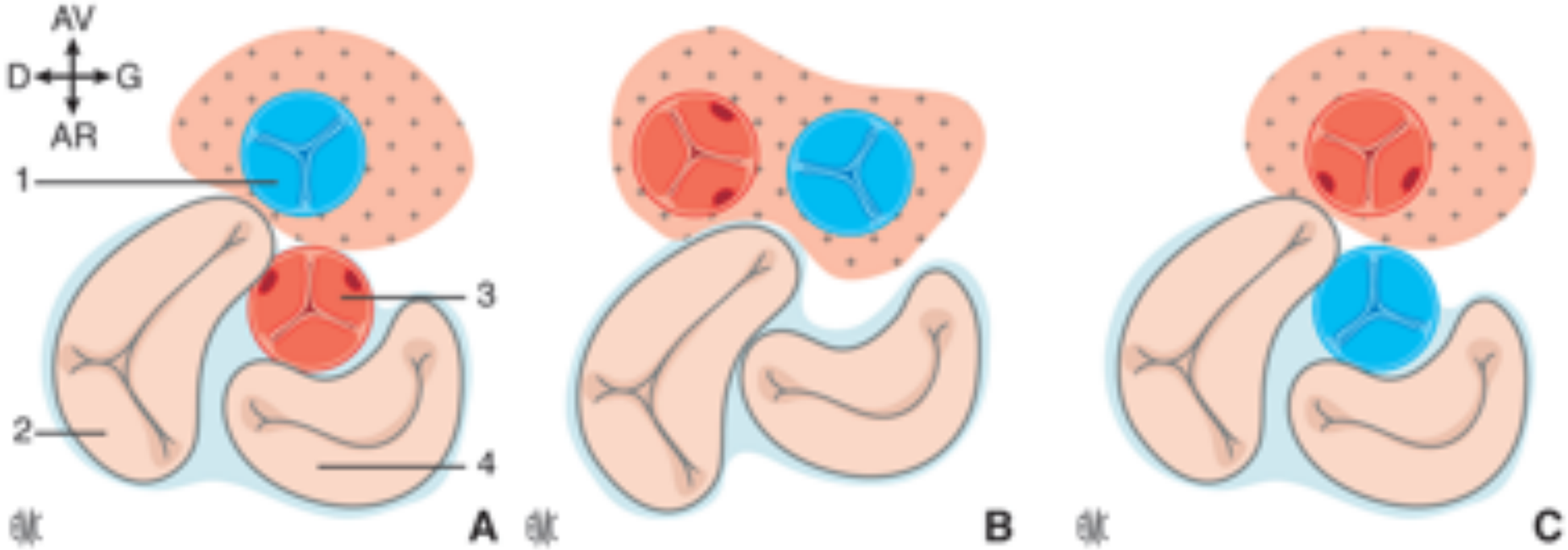
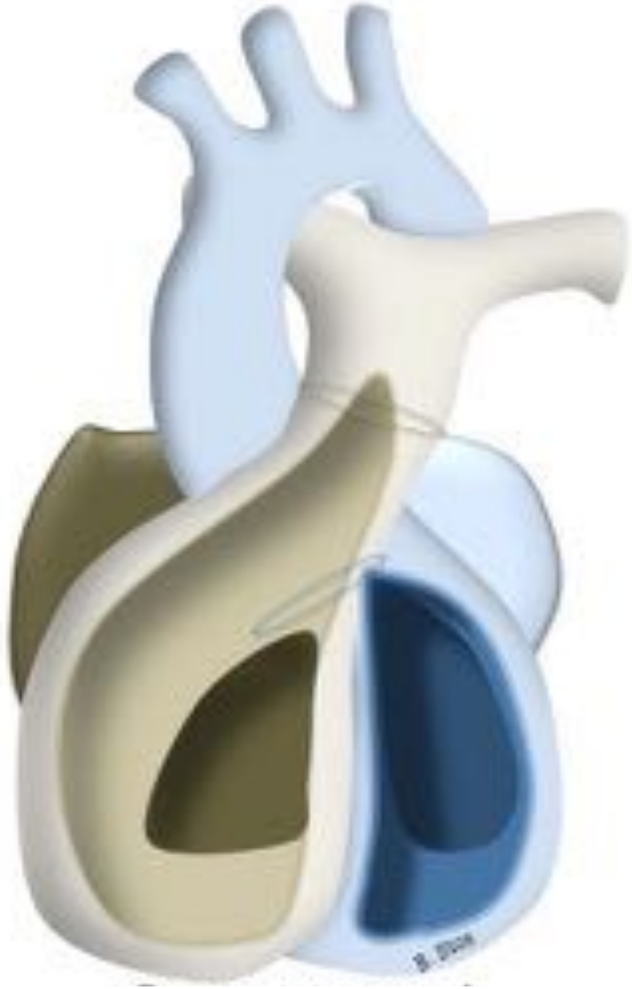
Definition, classification, embryology of DORV



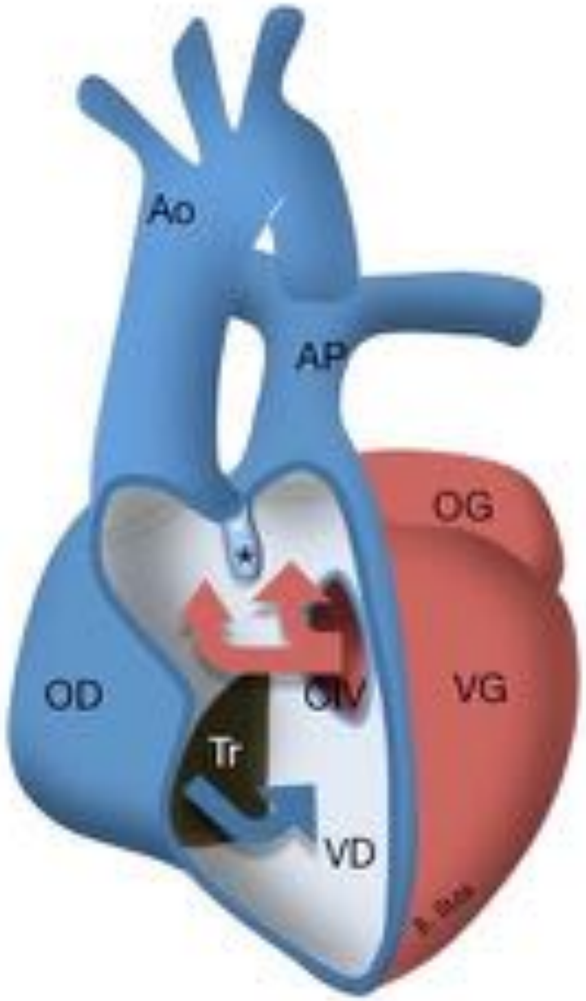
Normal

Malpositions
DORV

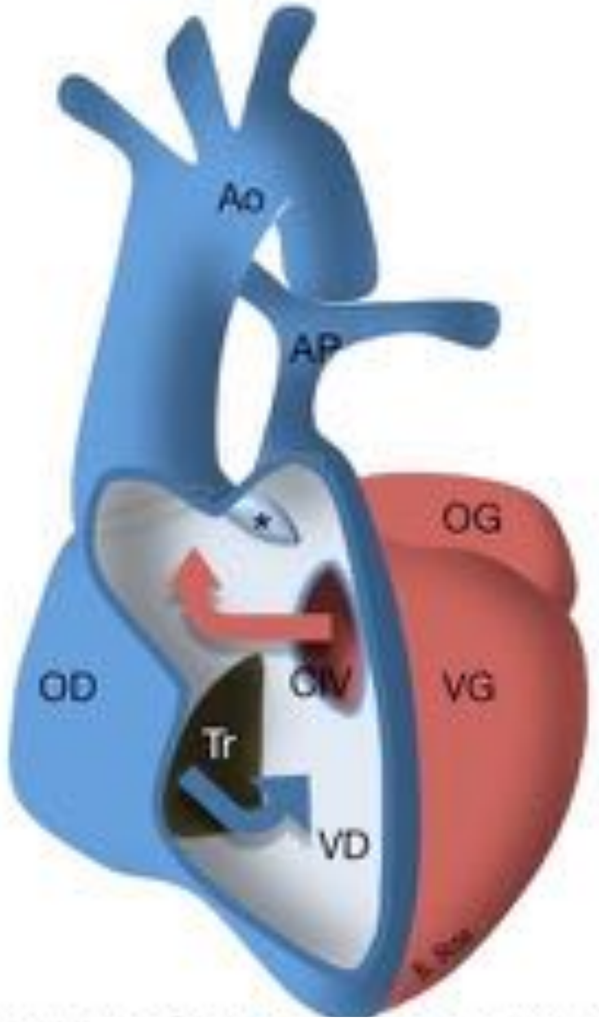
Transposition



Normal heart



DORV



DORV

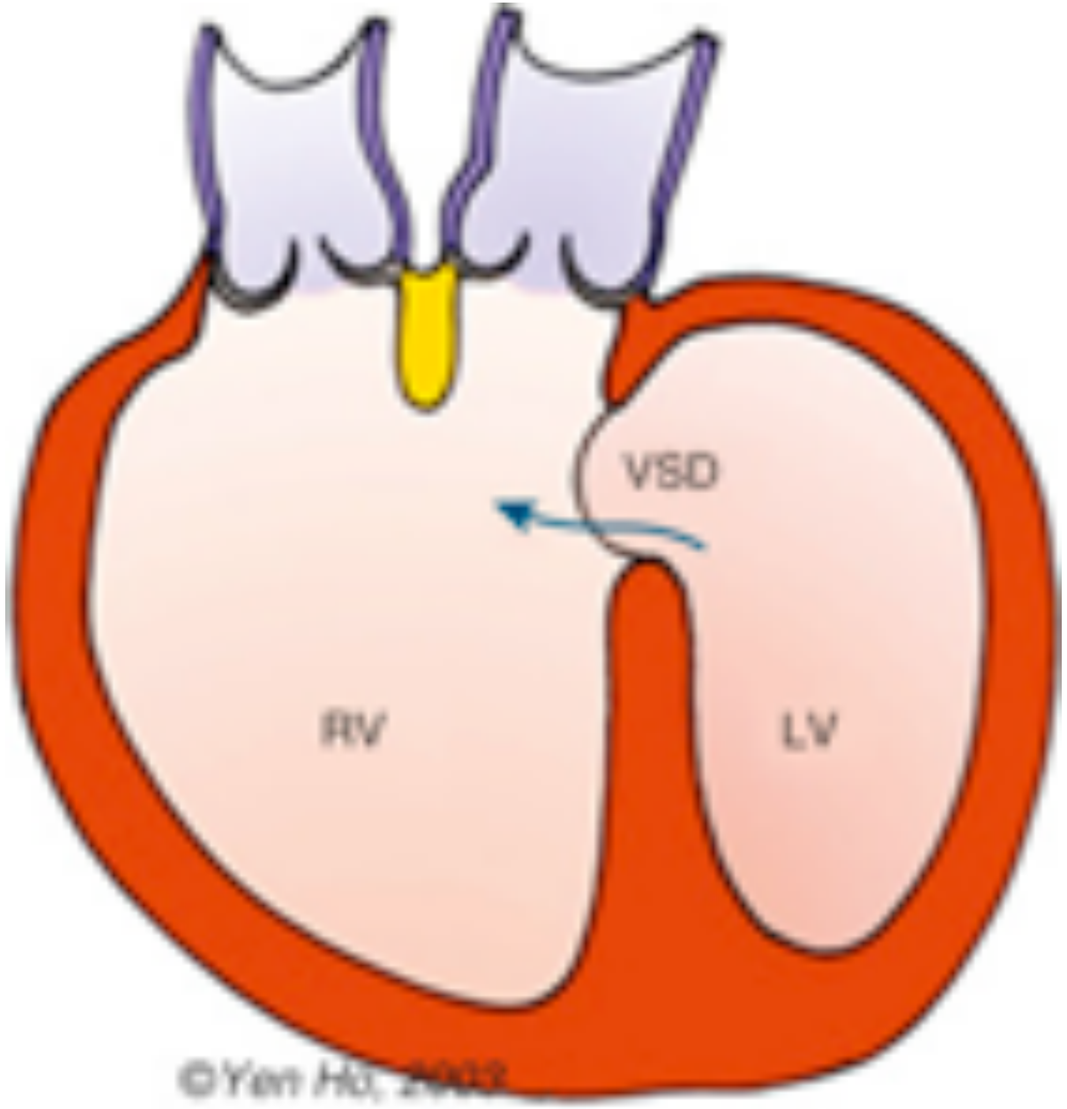
Courtesy Bertrand Stos

DORV - Definition

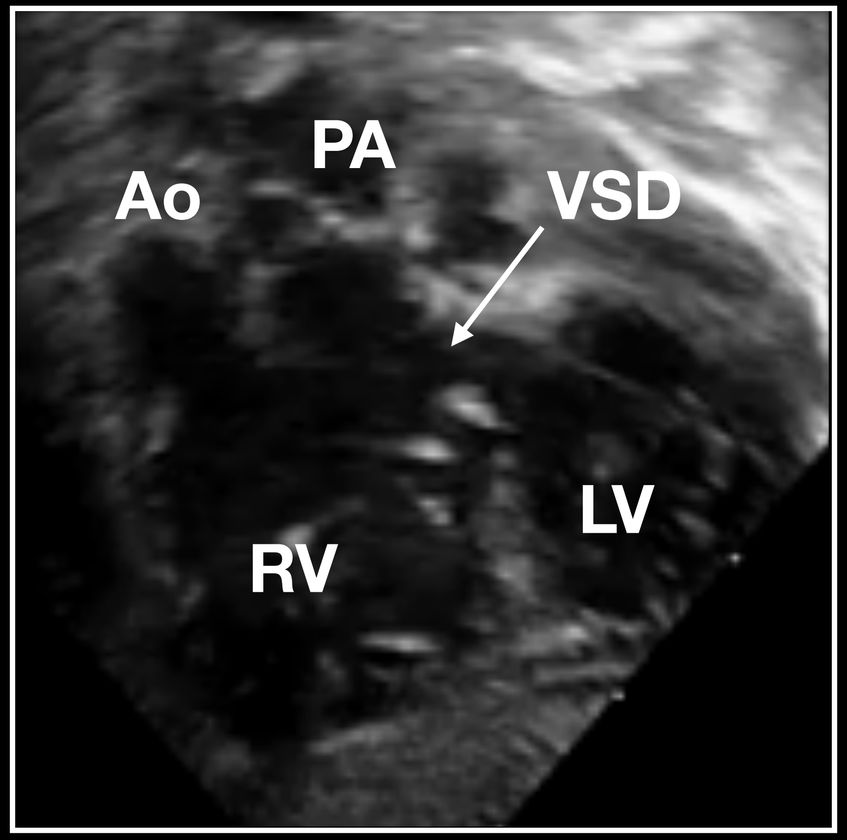
- Different definitions
  - Rule of 50%
  - Conal septum above the right ventricle
  - Sub-aortic + sub-pulmonary conus

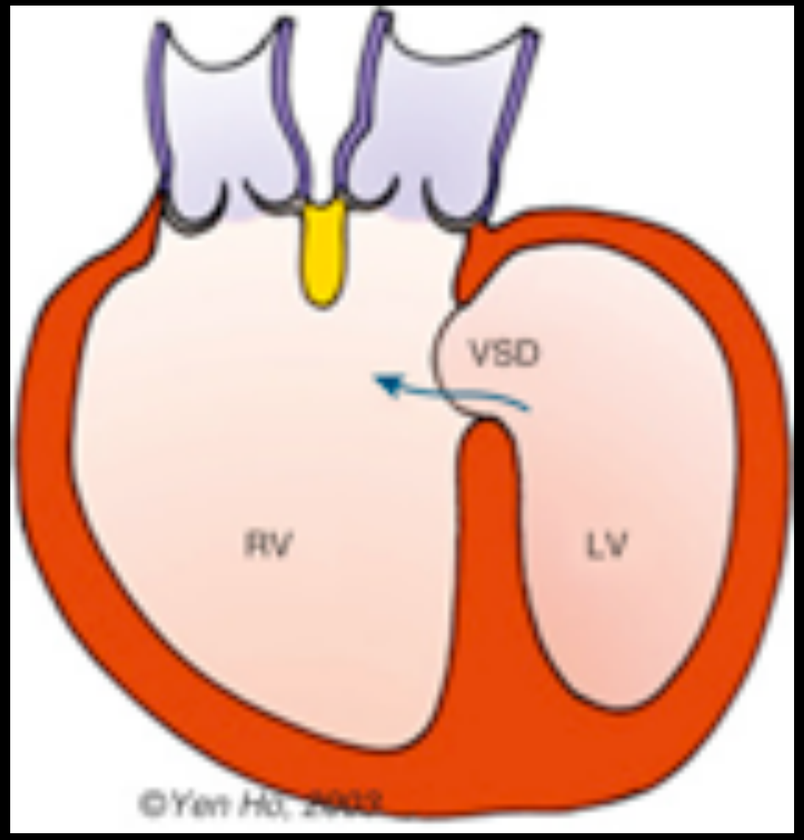
Only one clear rule

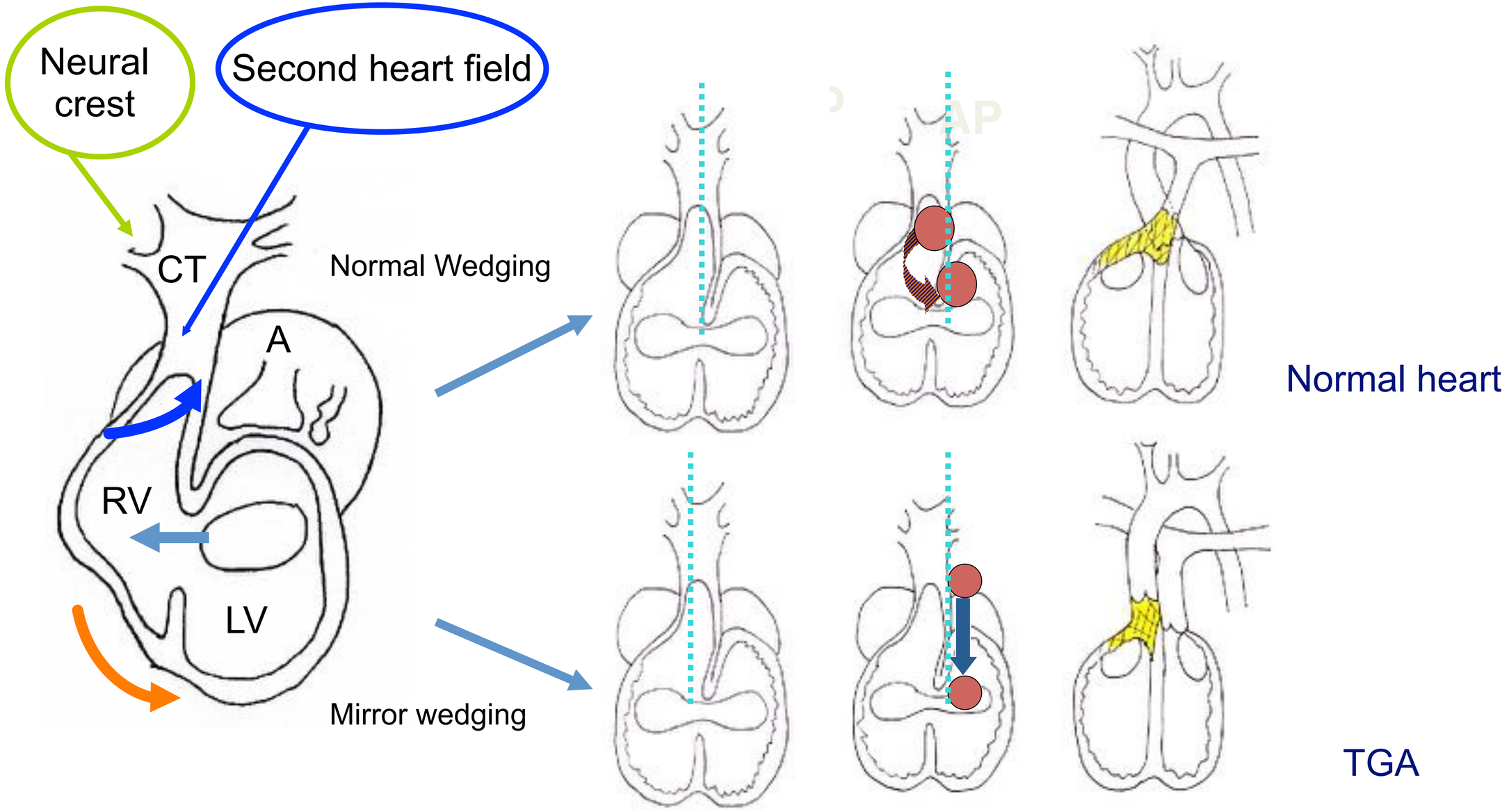
The two great vessels are above the right ventricle
or predominantly above the RV
& the only left ventricular outlet is the VSD



Double outlet right ventricle






Neural crest
Second heart field
CT
A
RV
LV
Normal Wedging
Mirror wedging
Normal heart
TGA
Early looping
Convergence
Wedging

DORV - Embryology

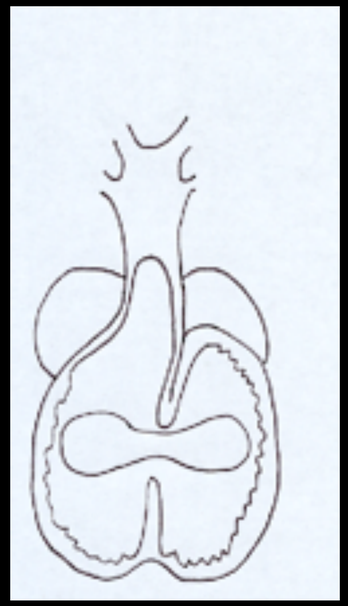
- DORV is a stop in cardiac development
  - at the « early looping » stage or even earlier
    - DORV is obligatory
    - left ventricle is under developed
  - at a later stage there is a combination of convergence and wedging anomalies leading to a malalignment of the conal septum and the lower part of the septum
    - VSD are located in the outlet
    - fibrous discontinuity between mitral and aortic valves (normal wedging) or between mitral and pulmonary valve in case of « mirror wedging »

3 groups of DORV (van Praagh)

- Groupe 1 : DORV with isolated anomalies of the outflow tracts
  « Late » DORV due to insufficient wedging
- Groupe 2 : DORV with outflow tracts anomalies + ventricles + AV valves
  « Early » DORV during « early looping »
- Groupe 3 : Looping anomalies
  DORV associated with heterotaxy

DORV

« Early » DORV



Vessel 1

Vessel 2

RA

LA

RV

LV

6

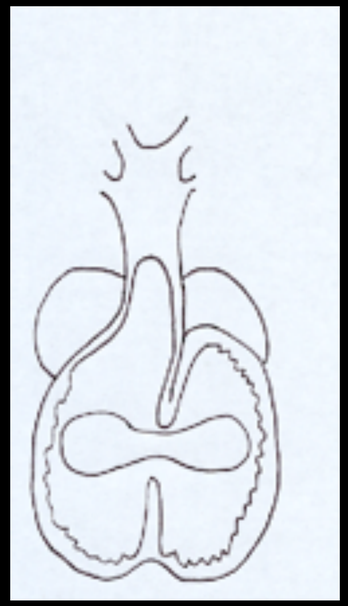
4

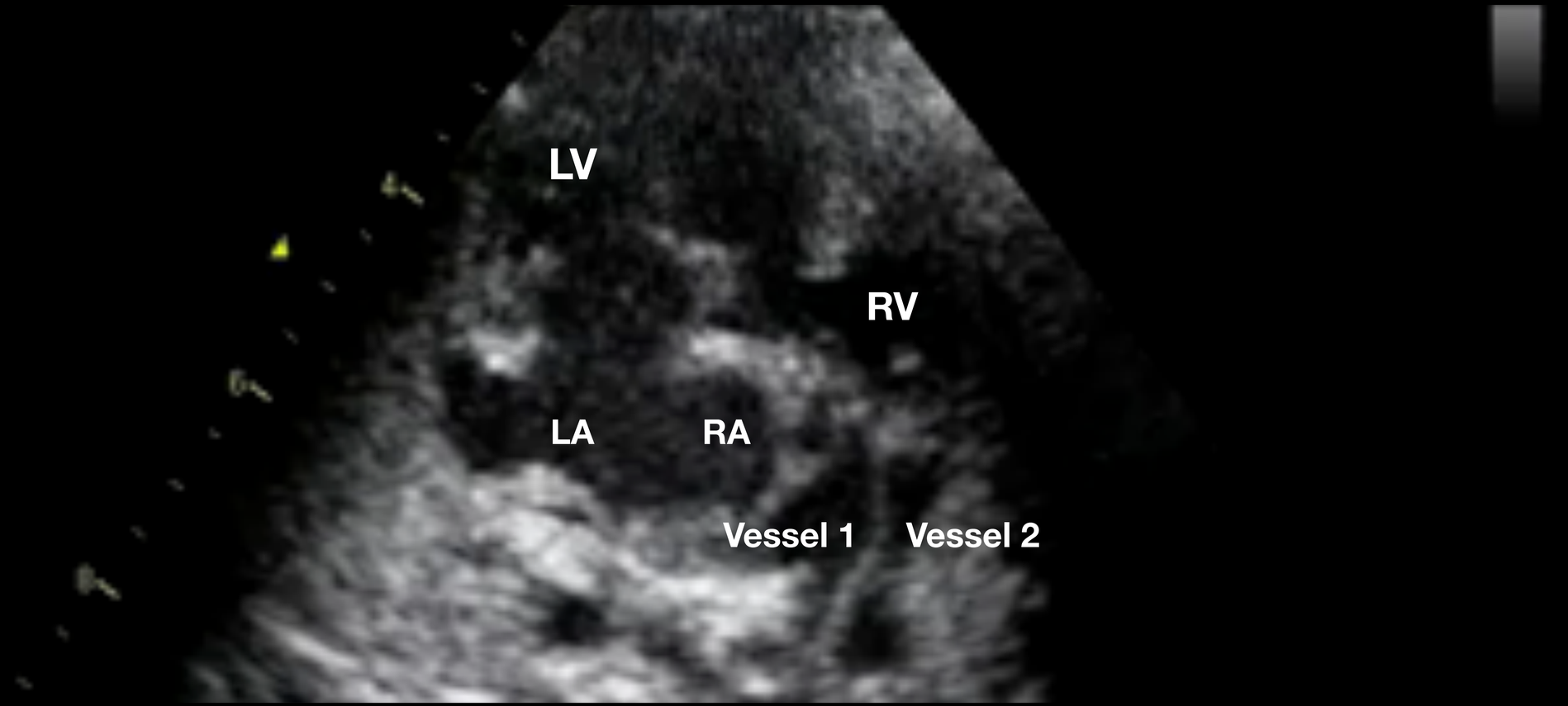
2

Group 3 DORV

DORV

« Early » DORV

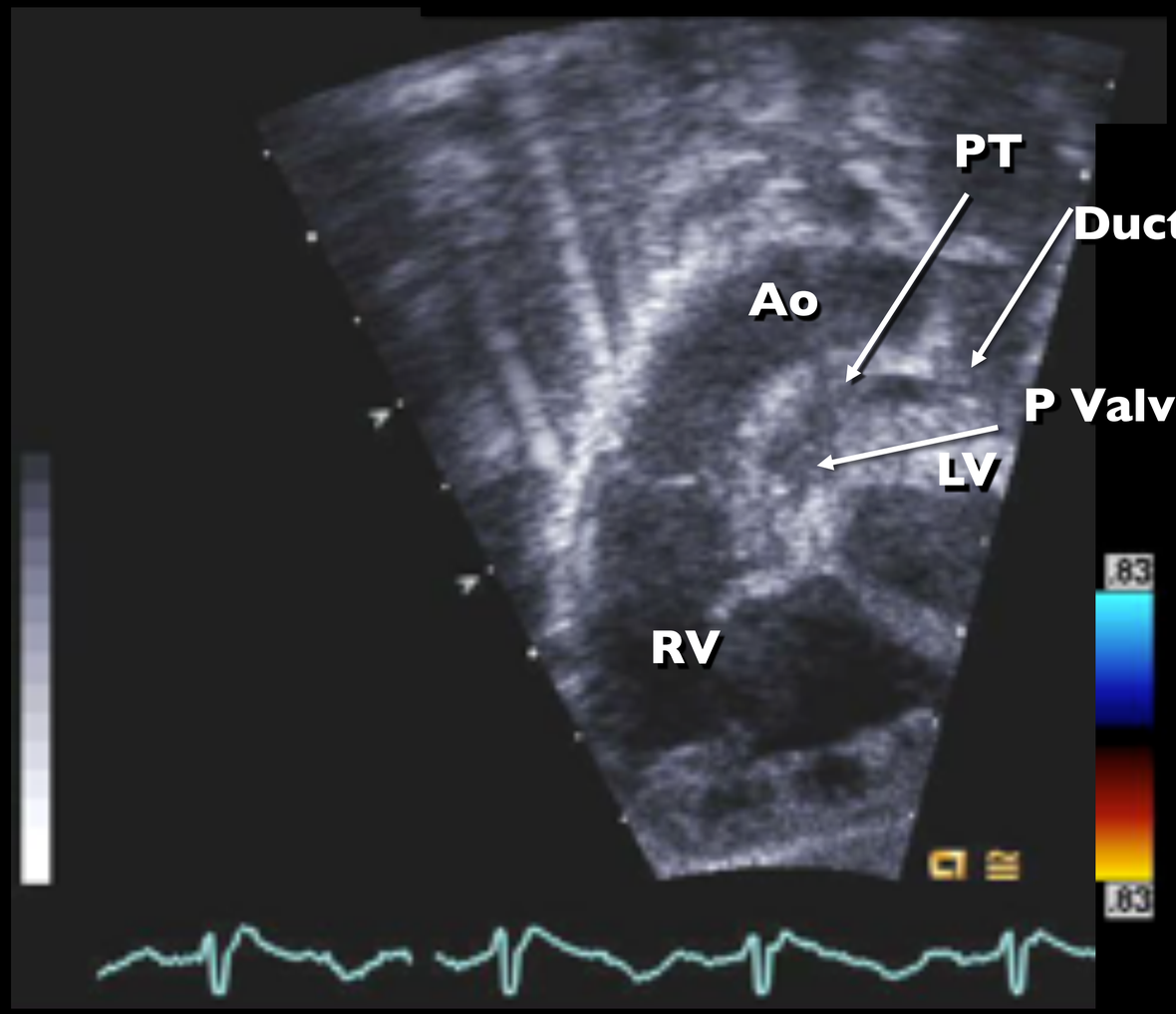


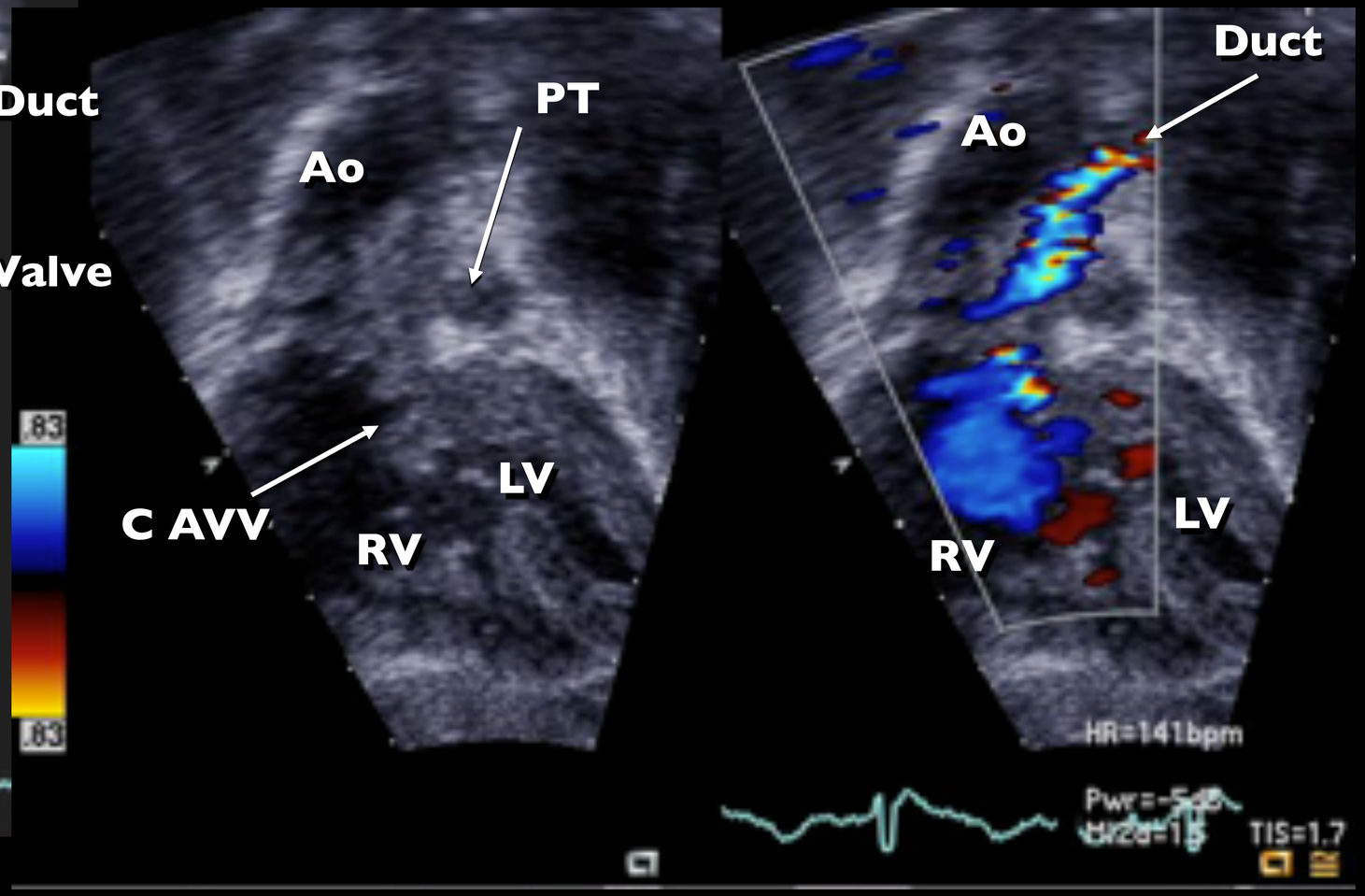


Group 3 DORV

Group 3 DORV

DORV in Right Isomerism
Pulmonary Stenosis/Atresia with DORV





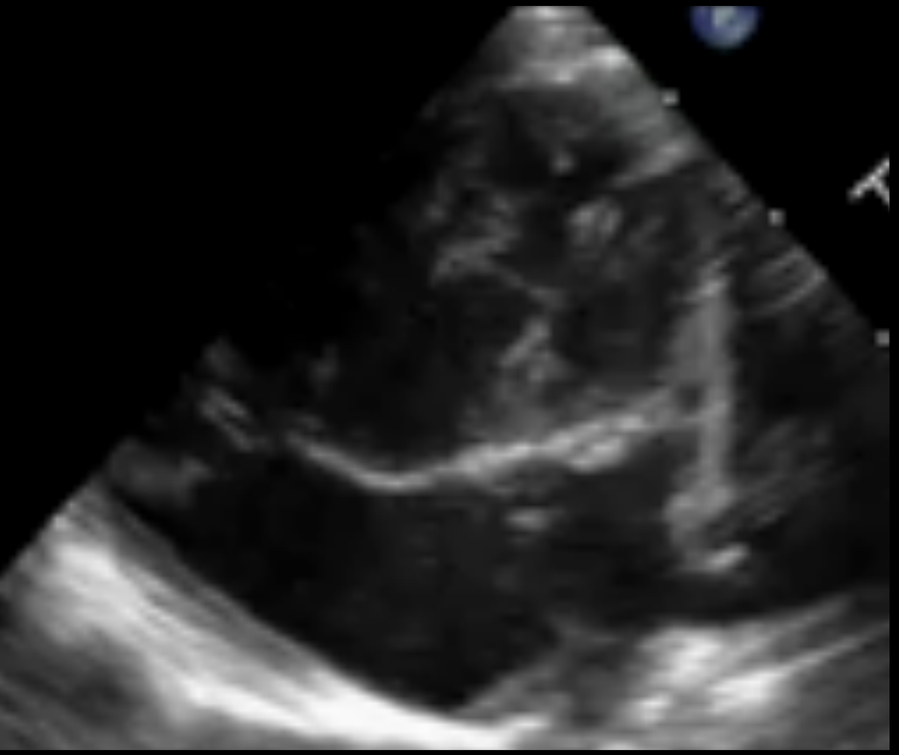
Subcostal Sagittal Cut

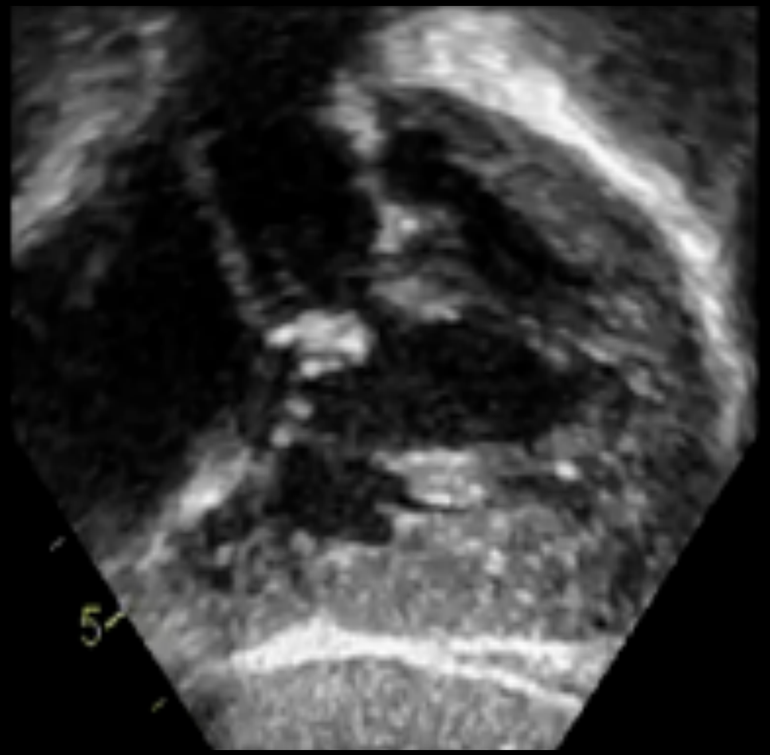
Subcostal Coronal Cuts

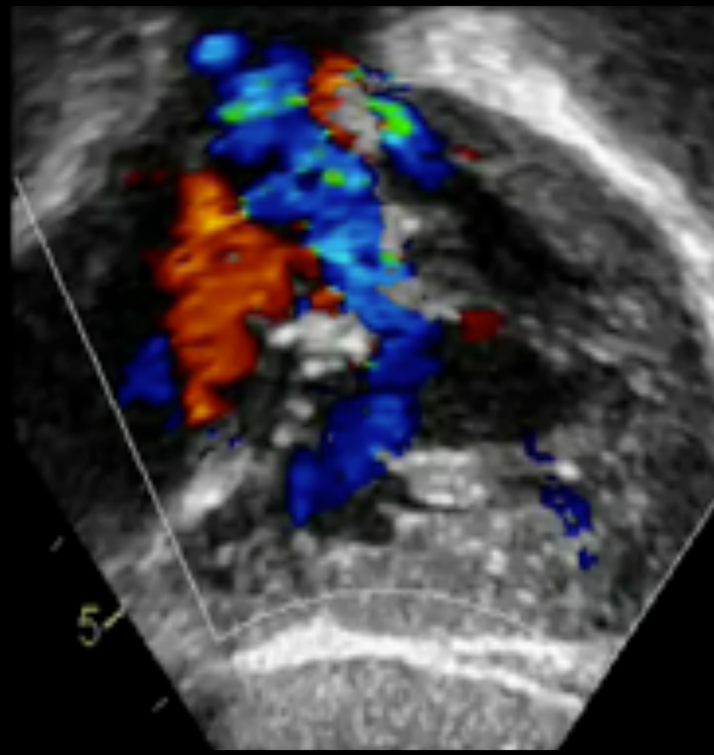
DORV

Mitral atresia and DORV

Group 2 DORV

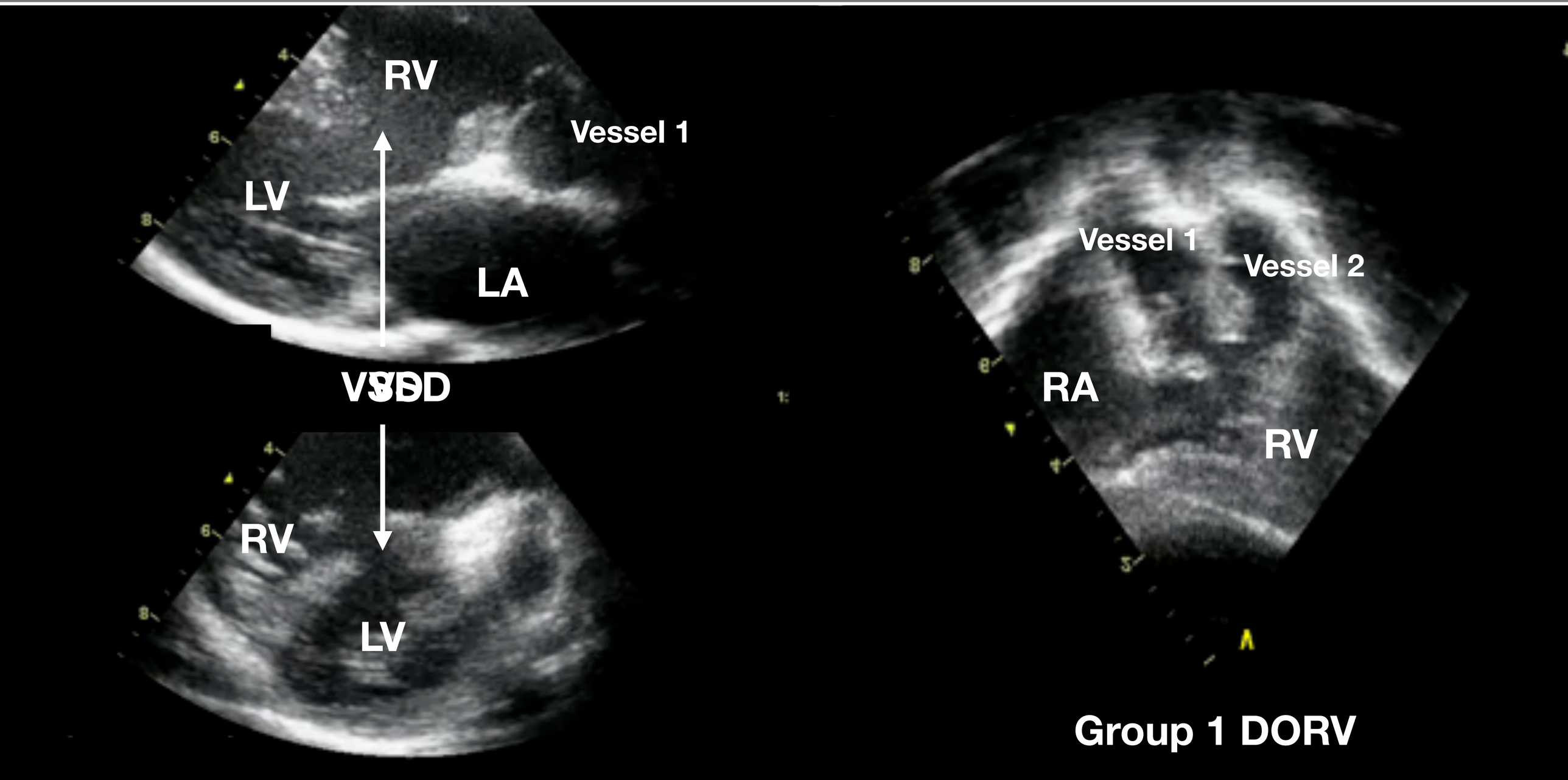






DORV

« Late » DORV



Group 1 DORV

DORV - classifications

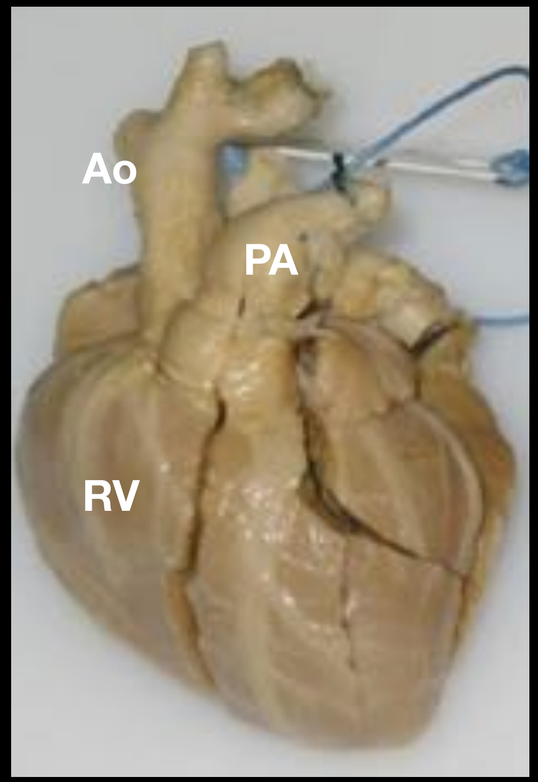
1-Relationship between VSD and great vessels (Lev 1972) : 4 types

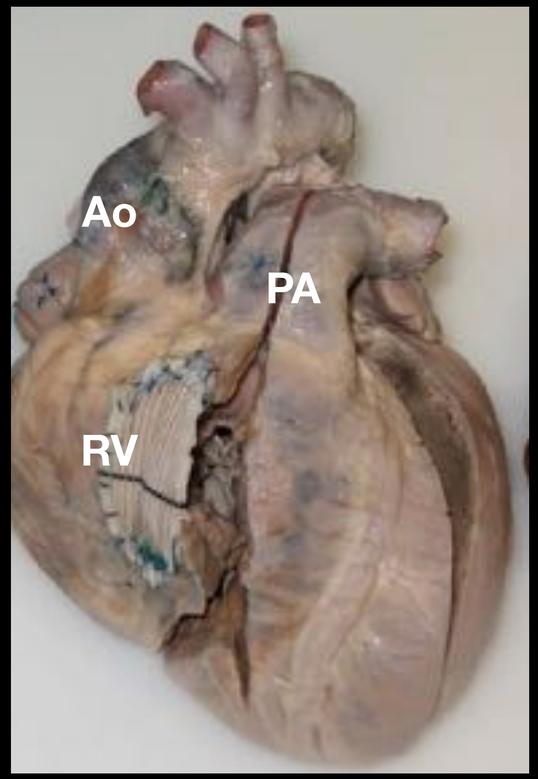
- sub-aortic
- Sub-pulmonary
- Double committed
- Non committed

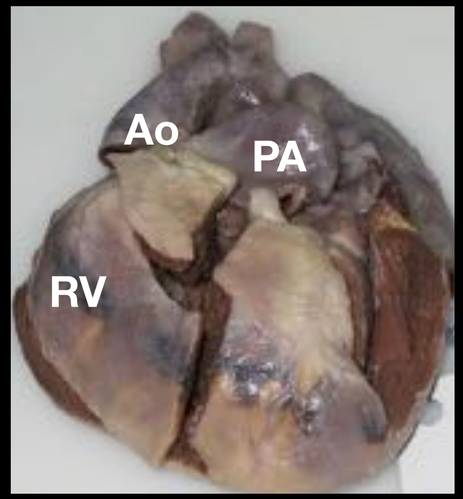
Physiological classification

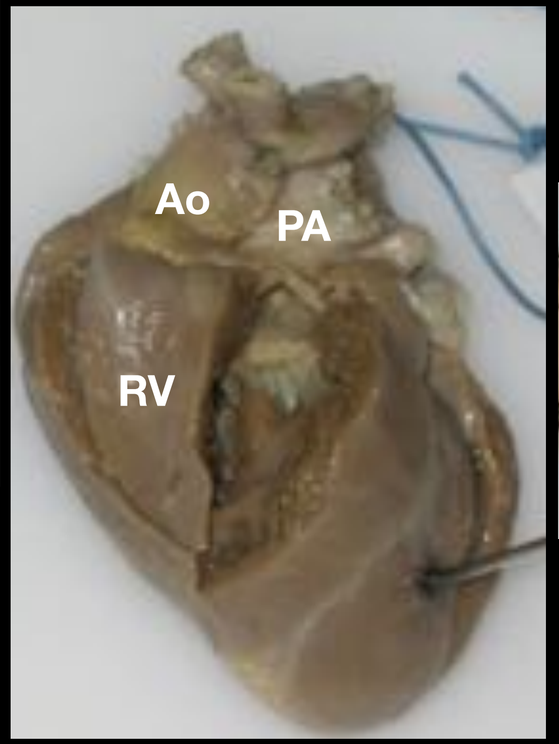
2-Relationship between the two great vessels (De La Cruz 1992)

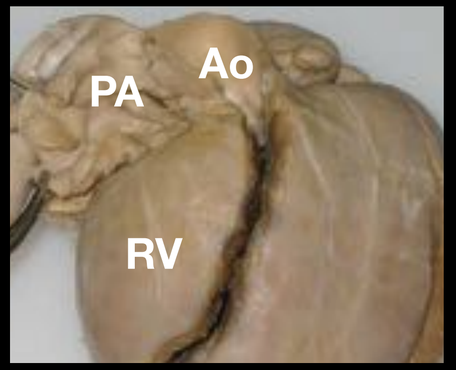
Relative position of the great vessels in DORV

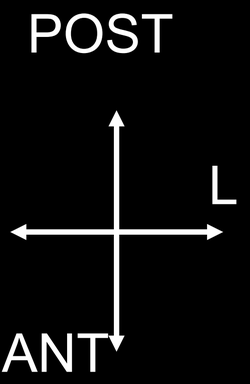




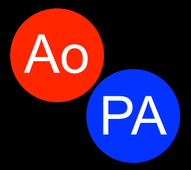


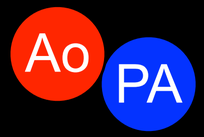


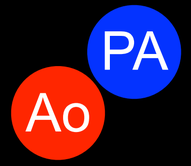


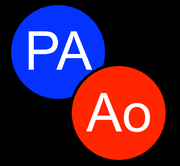


S,D,L

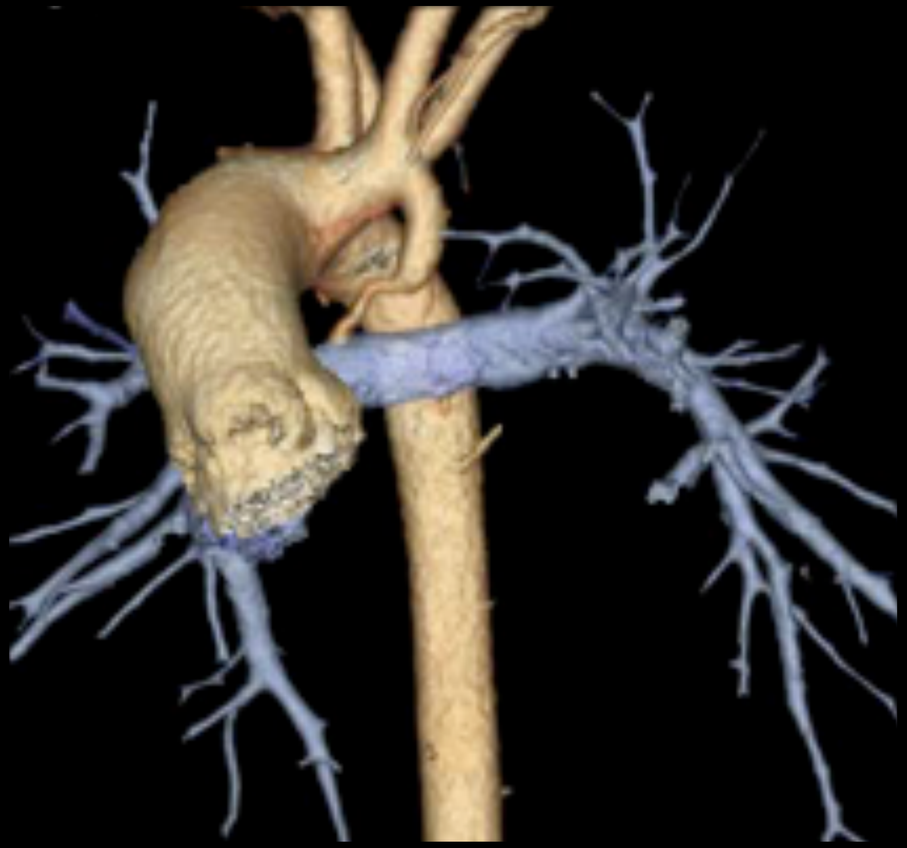






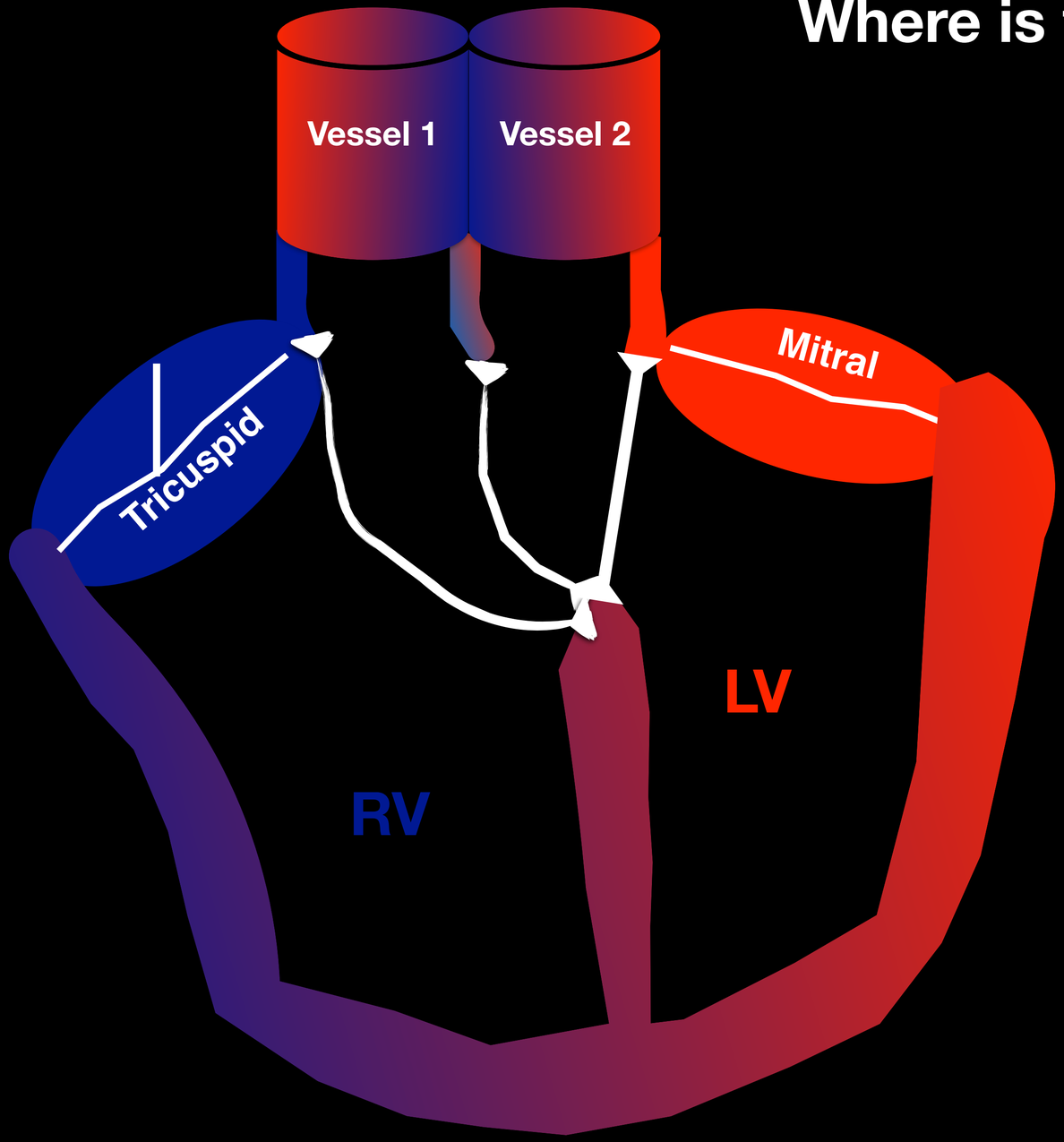


Relative position of the great vessels in DORV

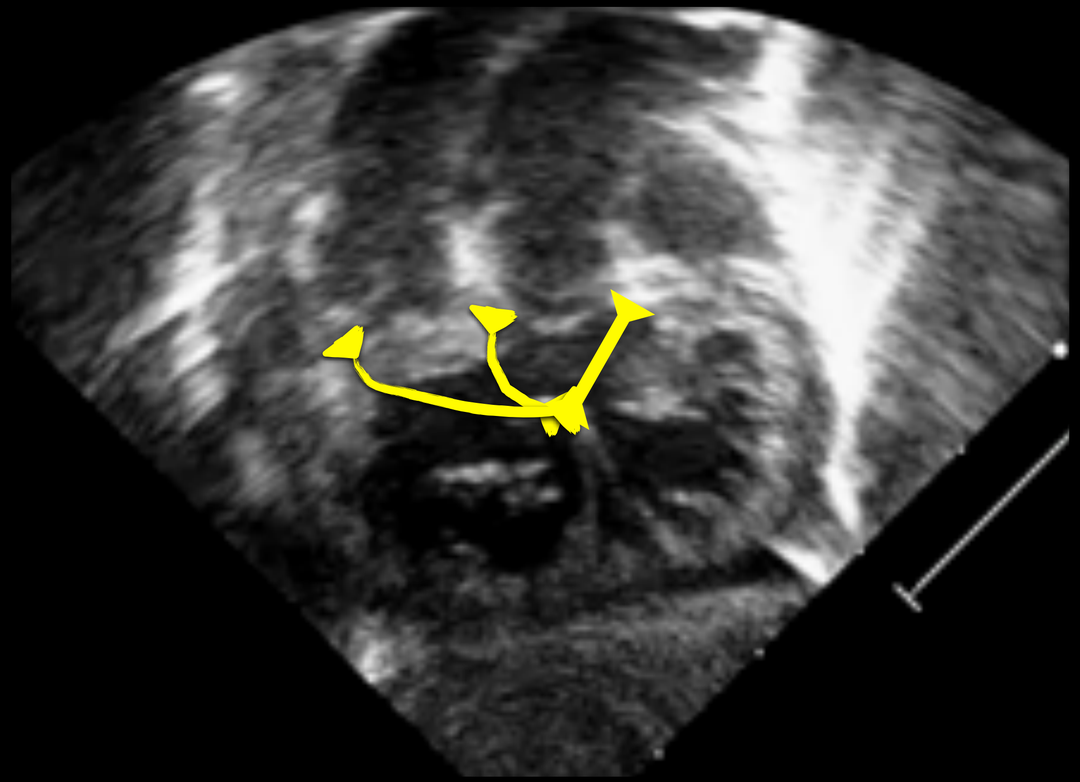


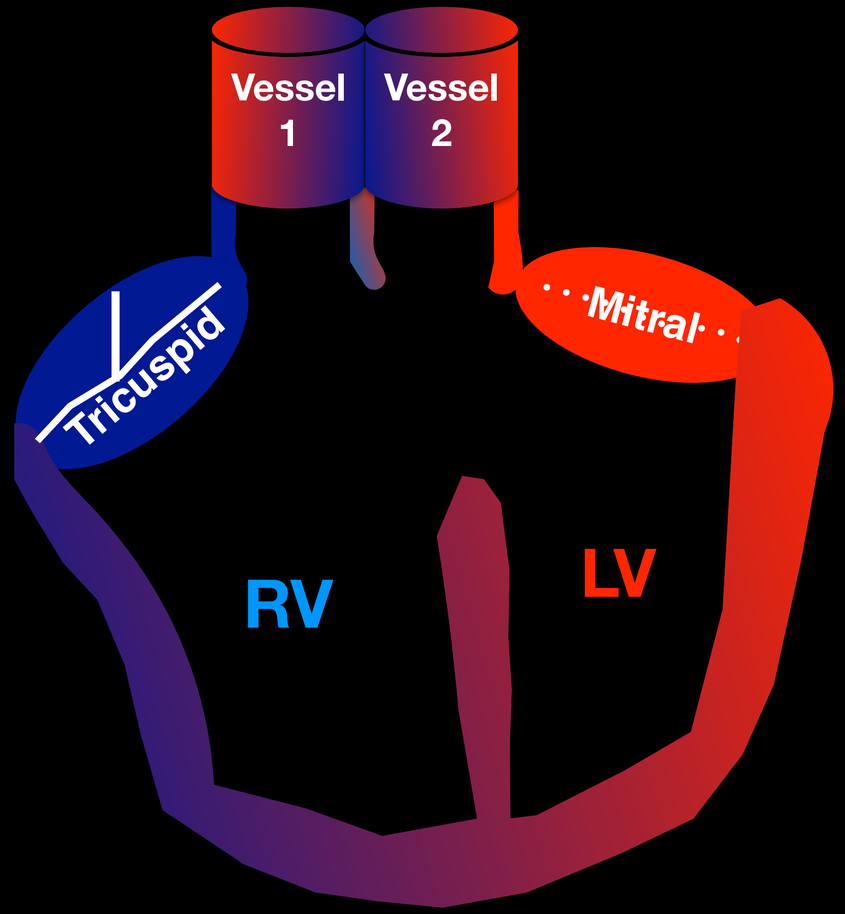
The position of the great vessels does not predict where is the VSD

Where is the « VSD » in DORV ?

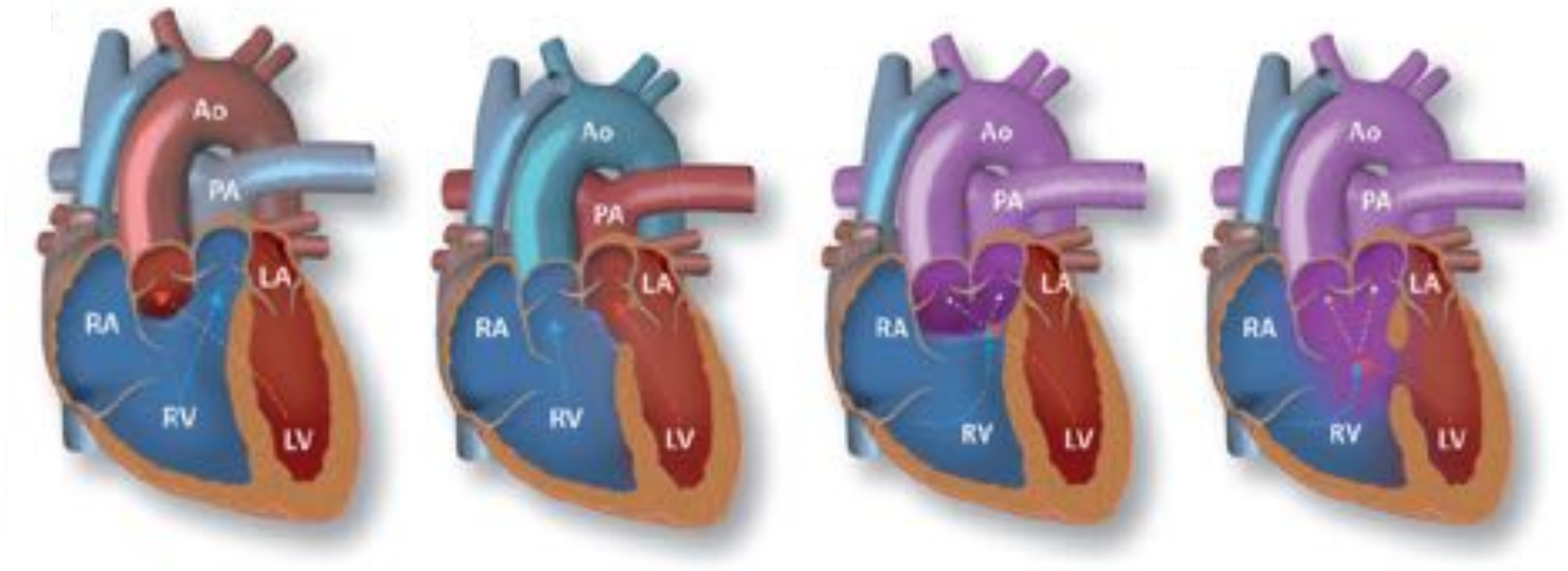


Where is the « VSD » in DORV ?

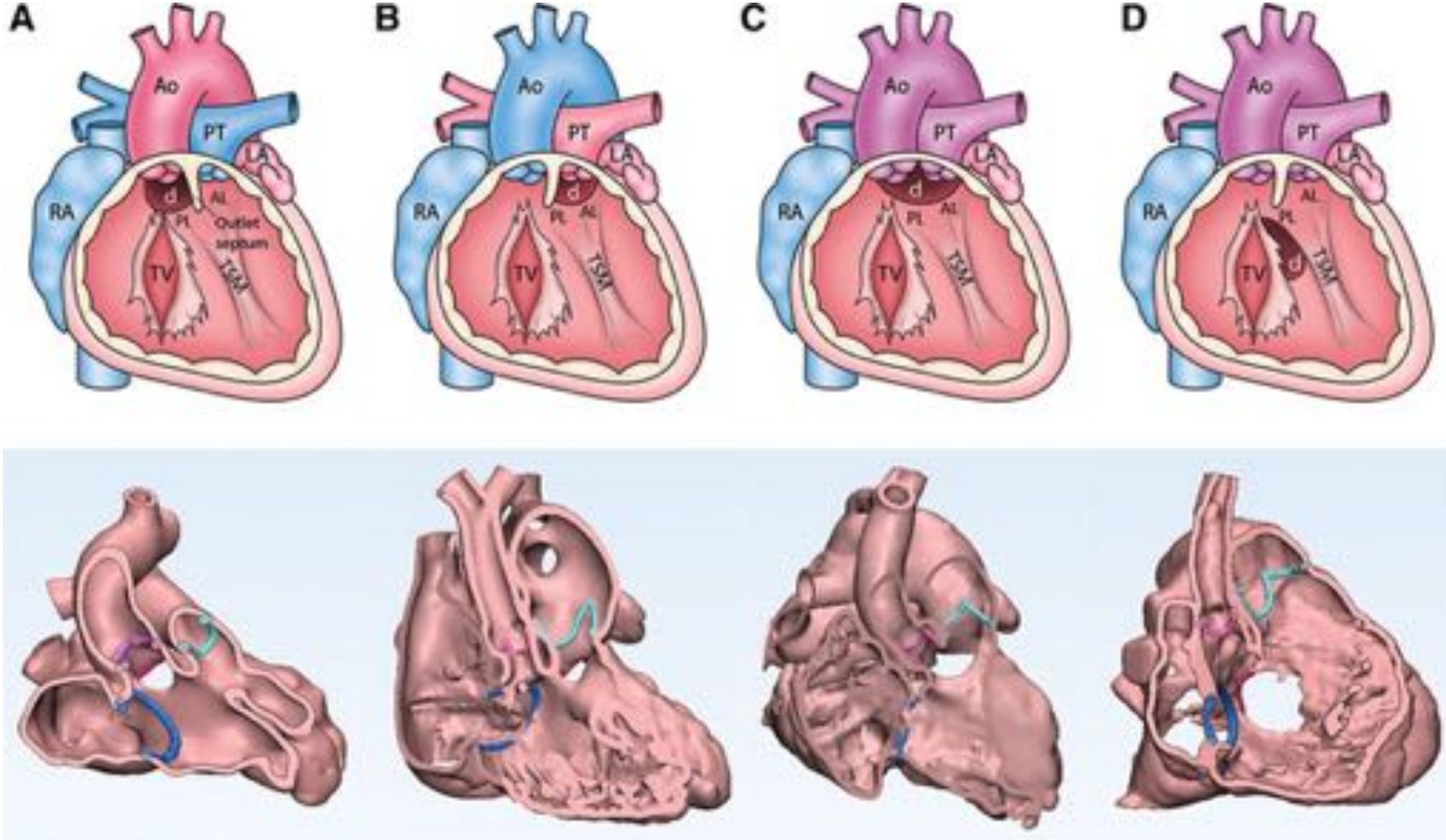




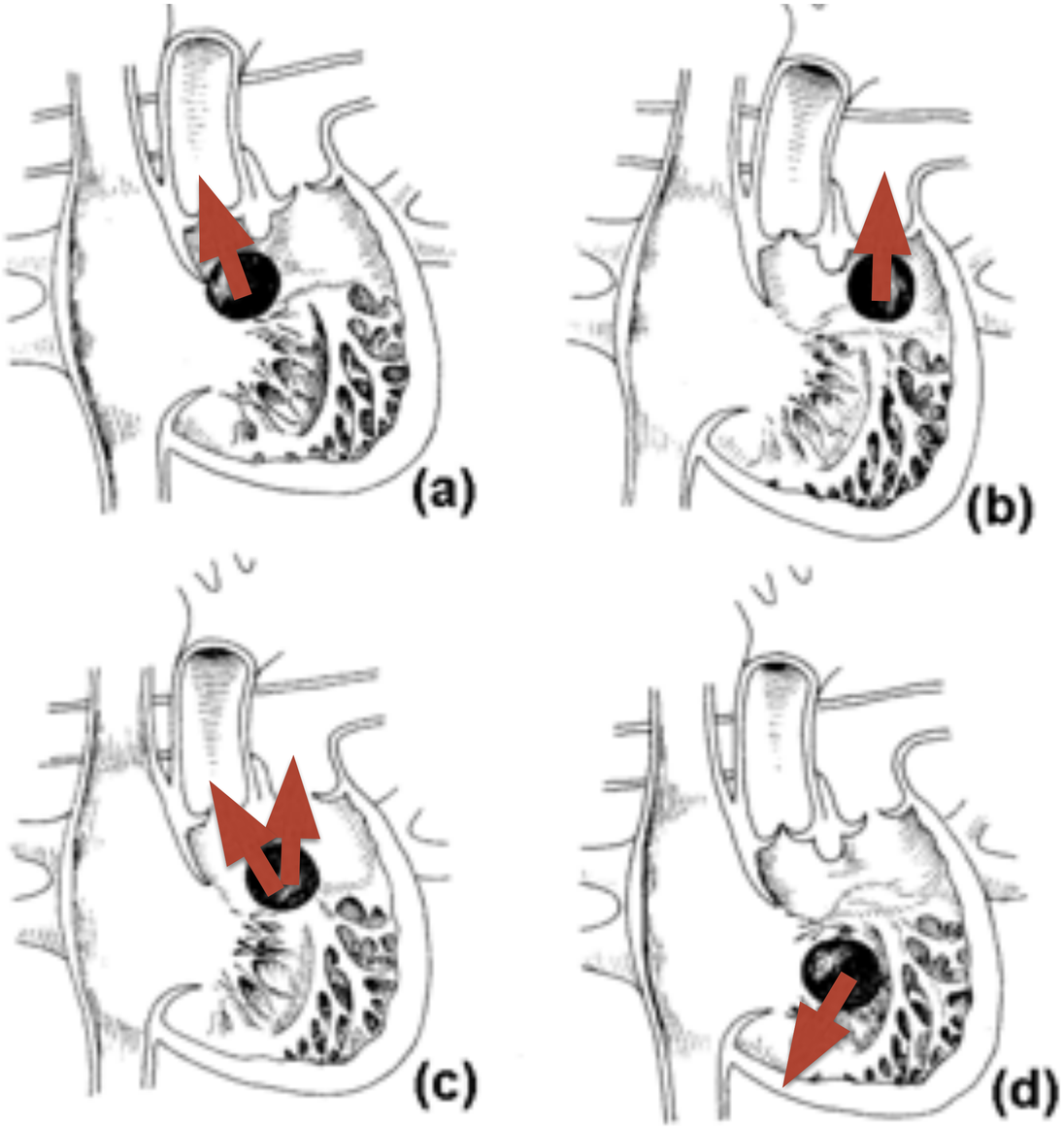
DORV - Relationship of VSD with great vessels



Sub-aortic **Sub-pulmonary** **Double committed** **Non committed**


A
B
C
D
Ao
PT
LA
RA
d
AI
PI
Outlet septum
TV
TSM
Subaortic VSD
Subpulmonary VSD
Doubly committed VSD
Non-committed (remote) VSD

DORV - Relationship of VSD with great vessels



Sidi D, Lecompte Y. Transposition and malposition of the great arteries with VSDs. In Moller JH, Hoffman JIE (eds). Pediatric cardiovascular medicine. 2000 : pp 363-73.

DORV-Surgical Repair

How to repair DORV ?

1. is biventricular repair possible ?

   if « YES »

2. is "anatomic" repair feasible ?

   if « NO »

3. which extra-anatomic repair is indicated ?

1. Is biventricular repair possible ?

- Problems related to ventricles and AV valves
  - size and function of ventricles
  - anatomy of A-V valves
    - abnormal insertions on conal septum
    - straddling
    - malformation (stenosis/regurgitation)

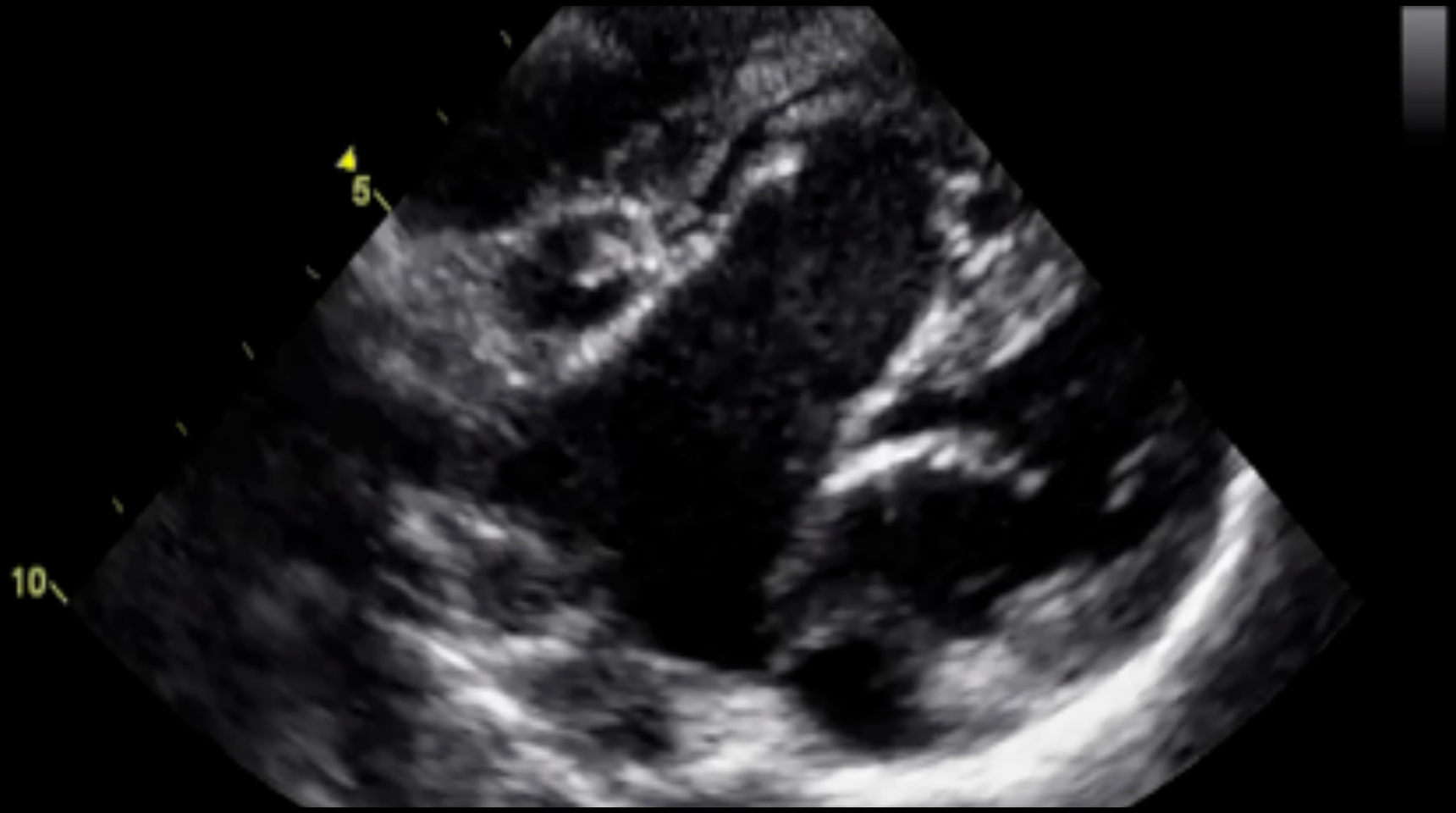
- Problems related to VSD
  - too large or multiple VSDs (swiss-cheese)
  - too small and impossible to enlarge by resecting conal septum
  - **non outlet VSD (muscular)**

1. is biventricular repair possible ?

- biventricular repair is impossible
- biventricular repair is possible but hazardous
- univentricular pathway (Fontan) is indicated
  (< 20% of cases)

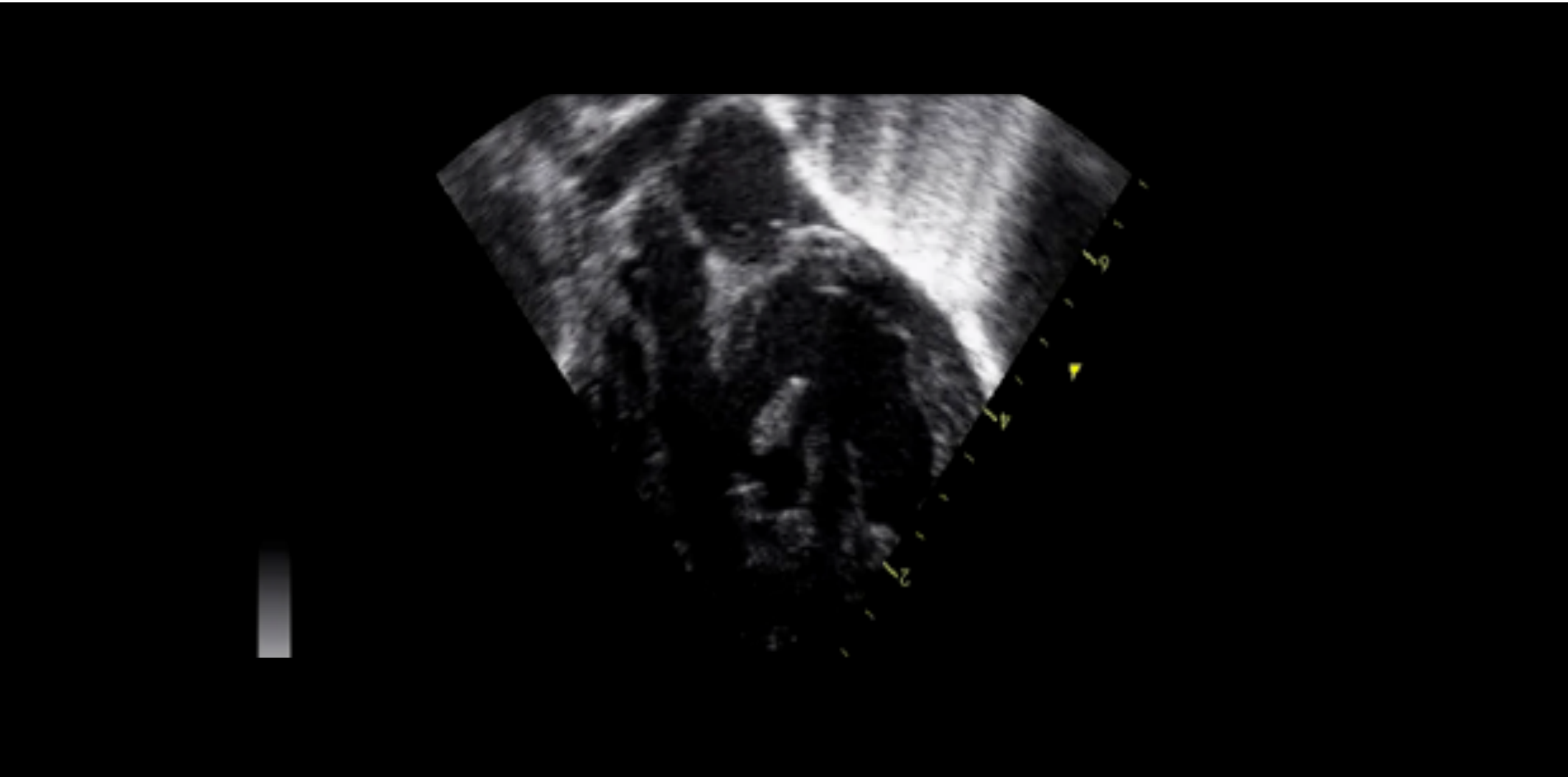
DORV

DORV non committed VSD



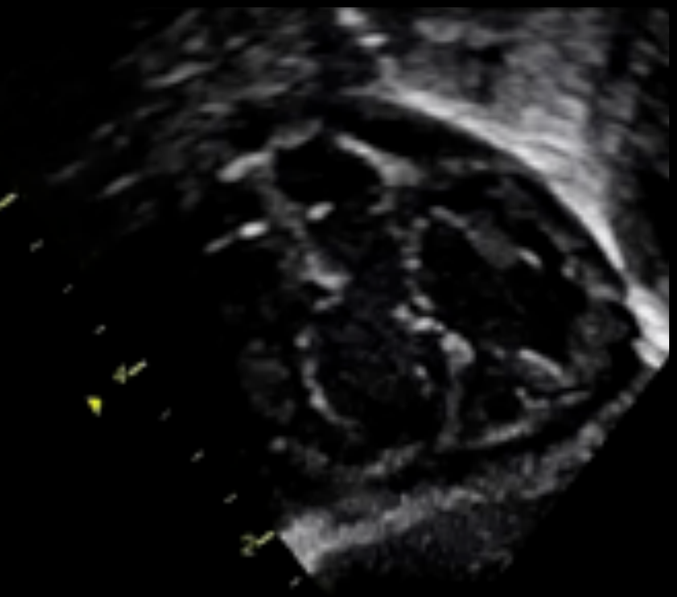
DORV

DORV Abnormal tricuspid valve-Hazardous repair



DORV

DORV Mitral straddling-Hazardous repair



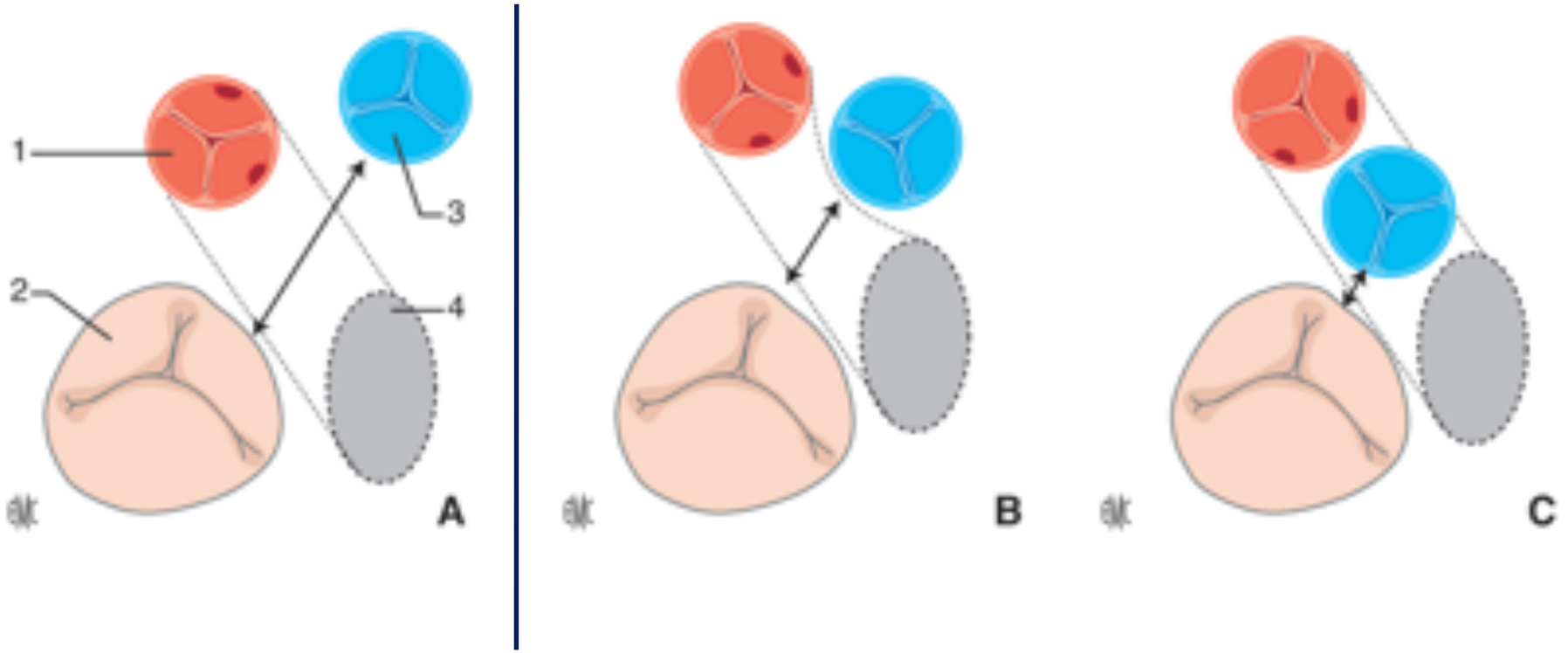
2. Is "anatomic" repair feasible ?

. LV connected to Aorta
. RV connected to PA
. Arterial valves in native position
. No extracardiac conduit

IntraVentricular Repair (IVR)

- VSD-type (no pulmonary stenosis)
- Fallot-type (pulmonary stenosis)

Determinant: tricuspid-to-pulmonary distance (lenght of subpulmonary conus)



IVR possible

IVR impossible

DORV

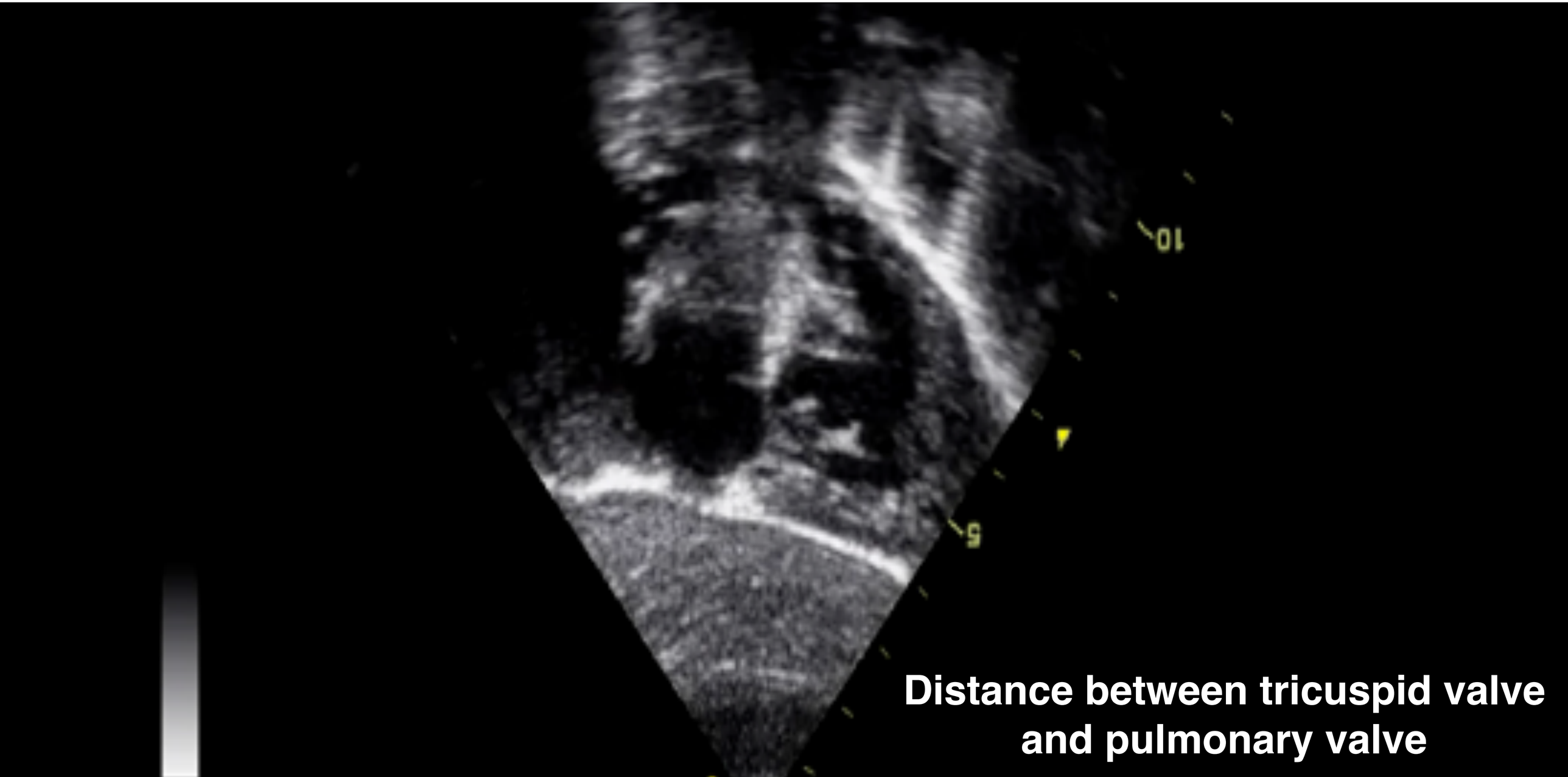
« Late » DORV sub aortic VSD-Evaluation for IVR

DORV sub-aortic VSD

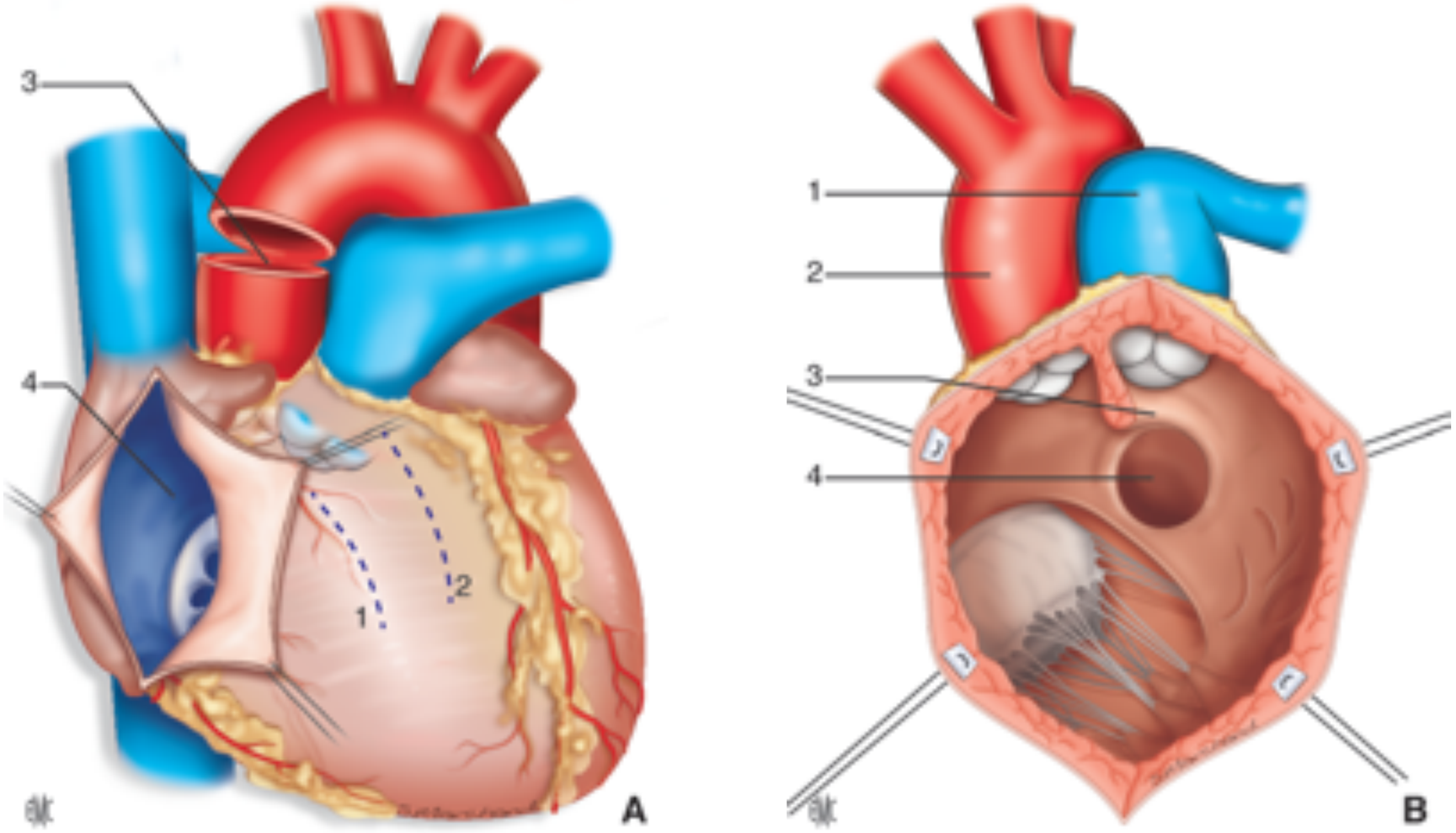
Distance between tricuspid valve and pulmonary valve

DORV

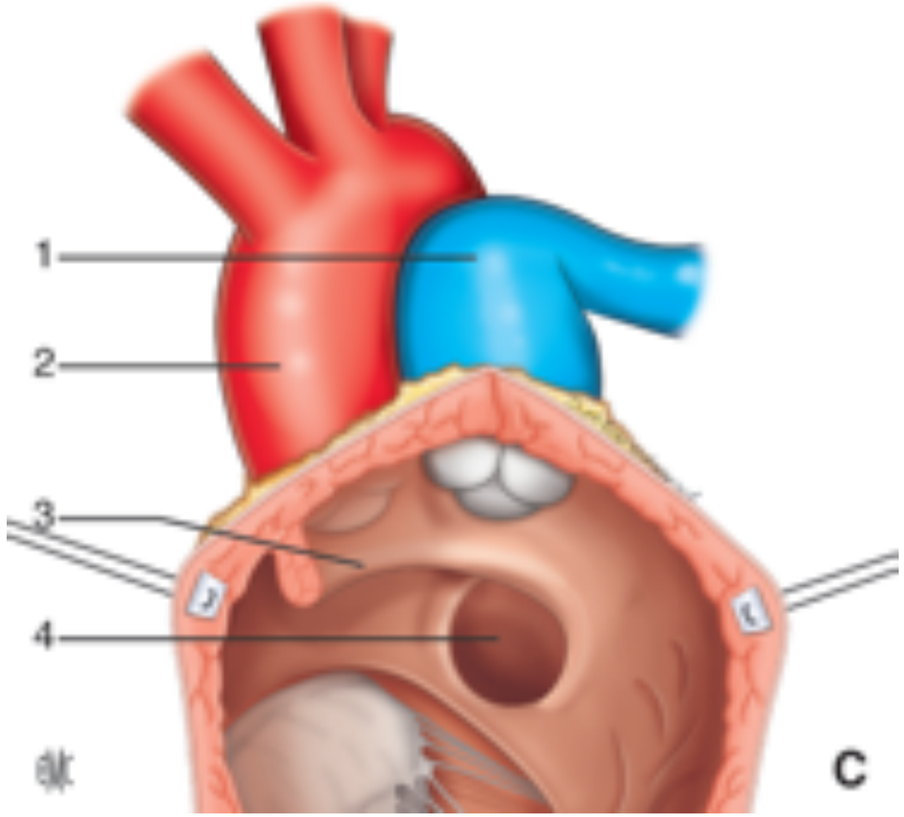
« Late » DORV sub aortic VSD-Evaluation for IVR

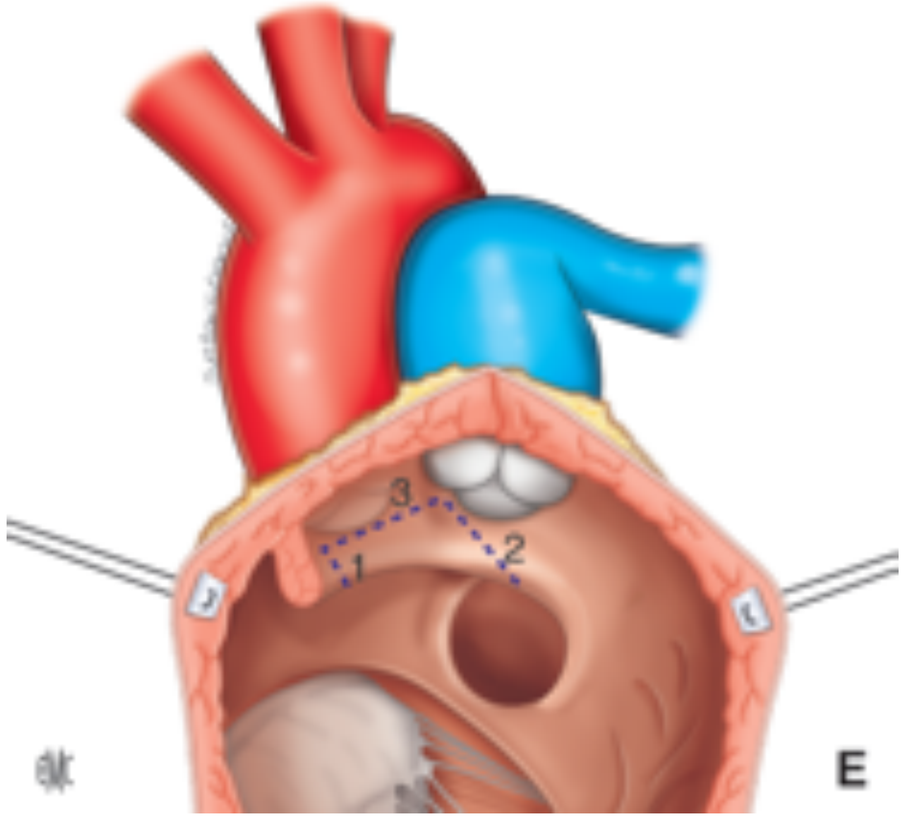


Intraventricular repair

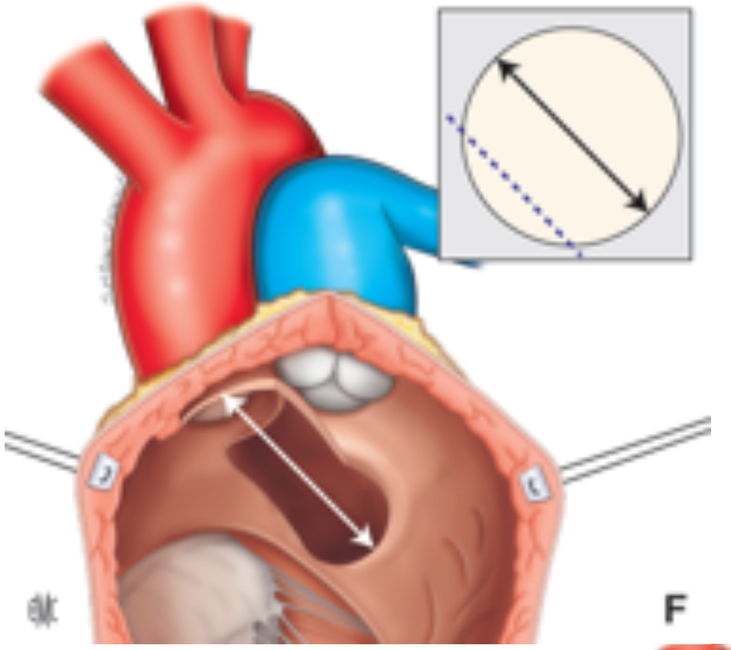


Intraventricular repair





Intraventricular repair



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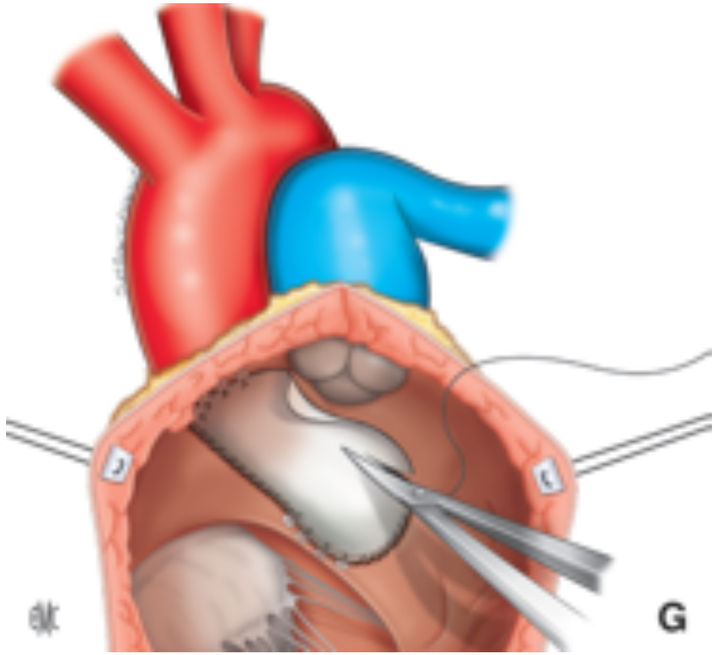
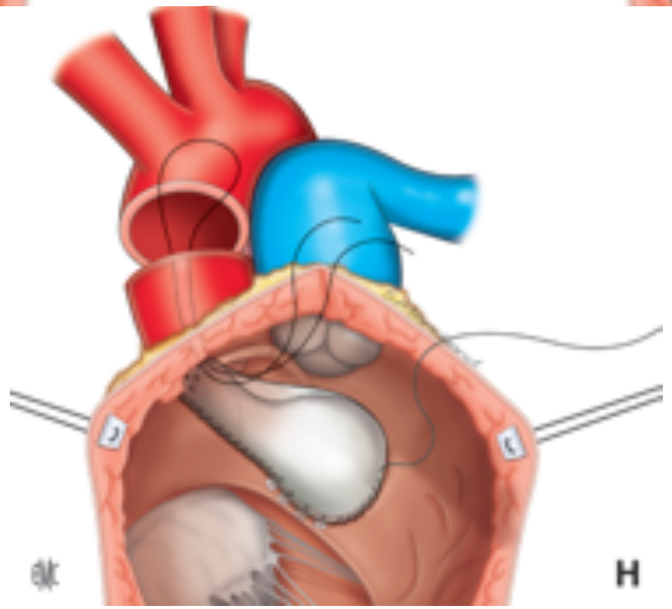


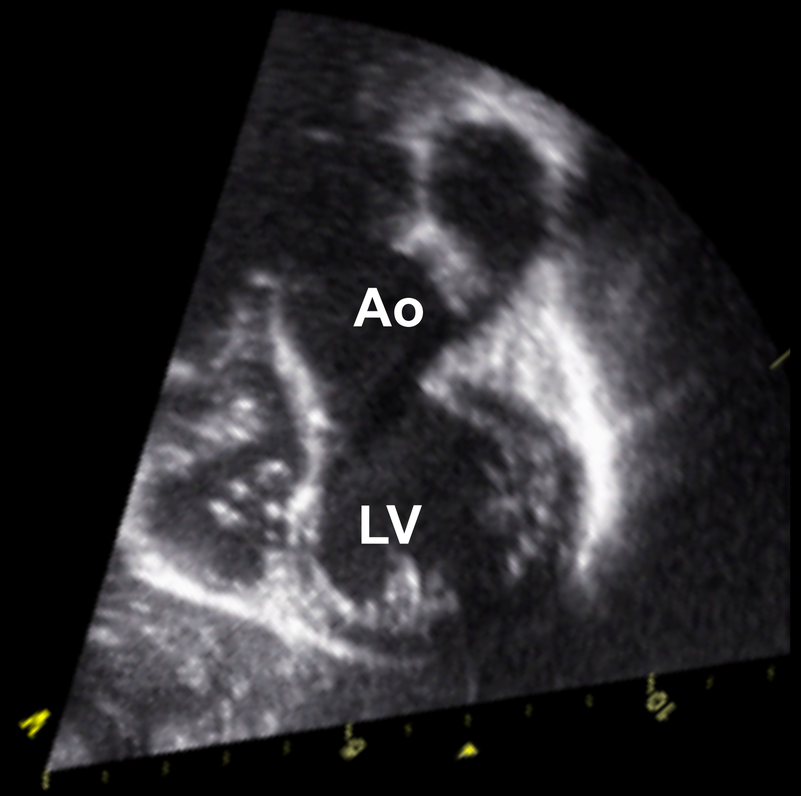
G

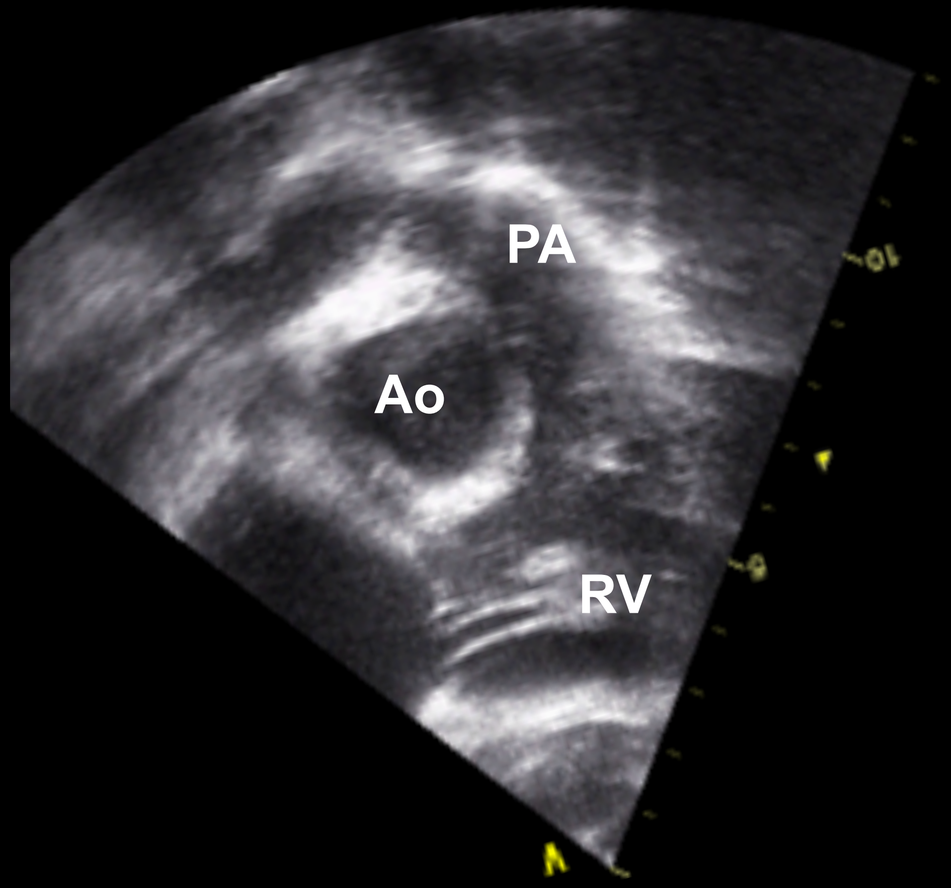


H

DORV

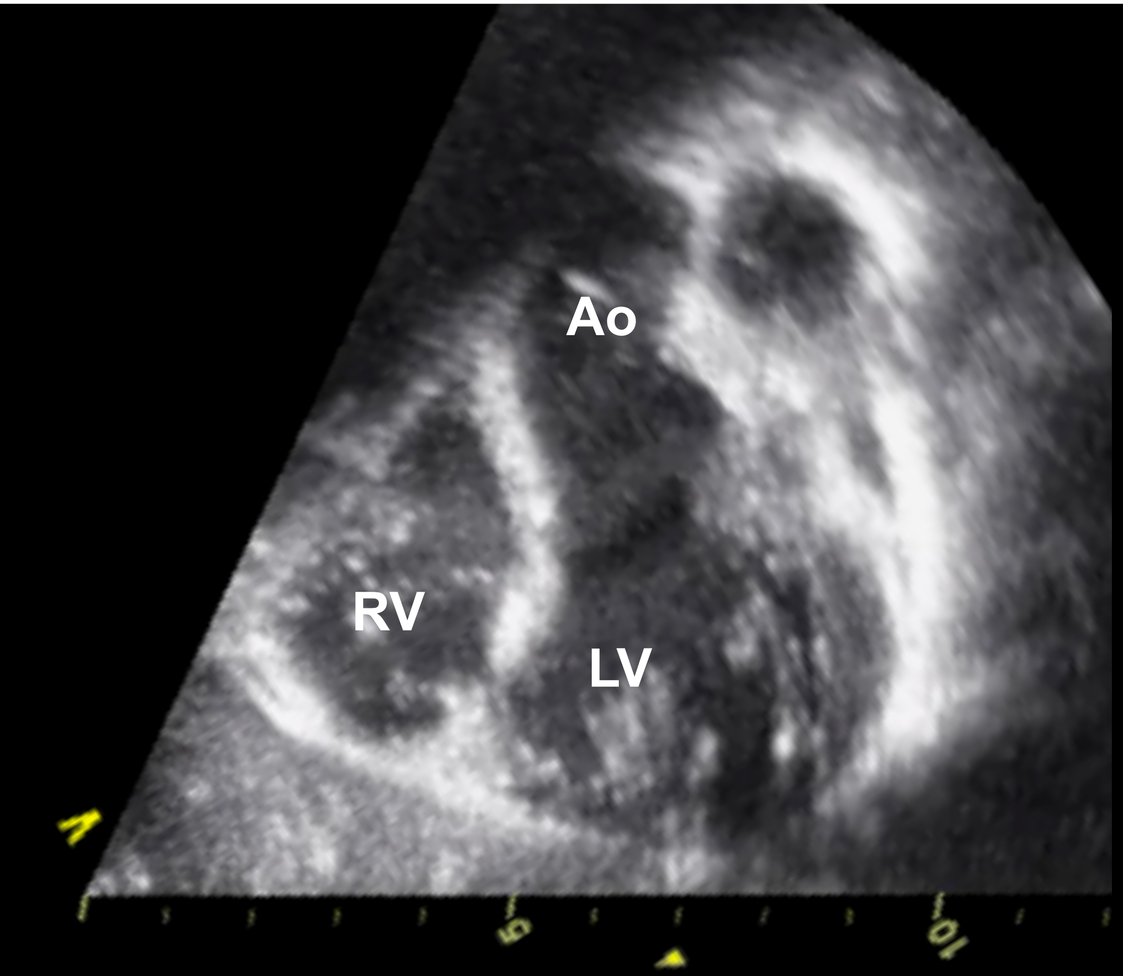
« Late » DORV sub aortic VSD-After IVR

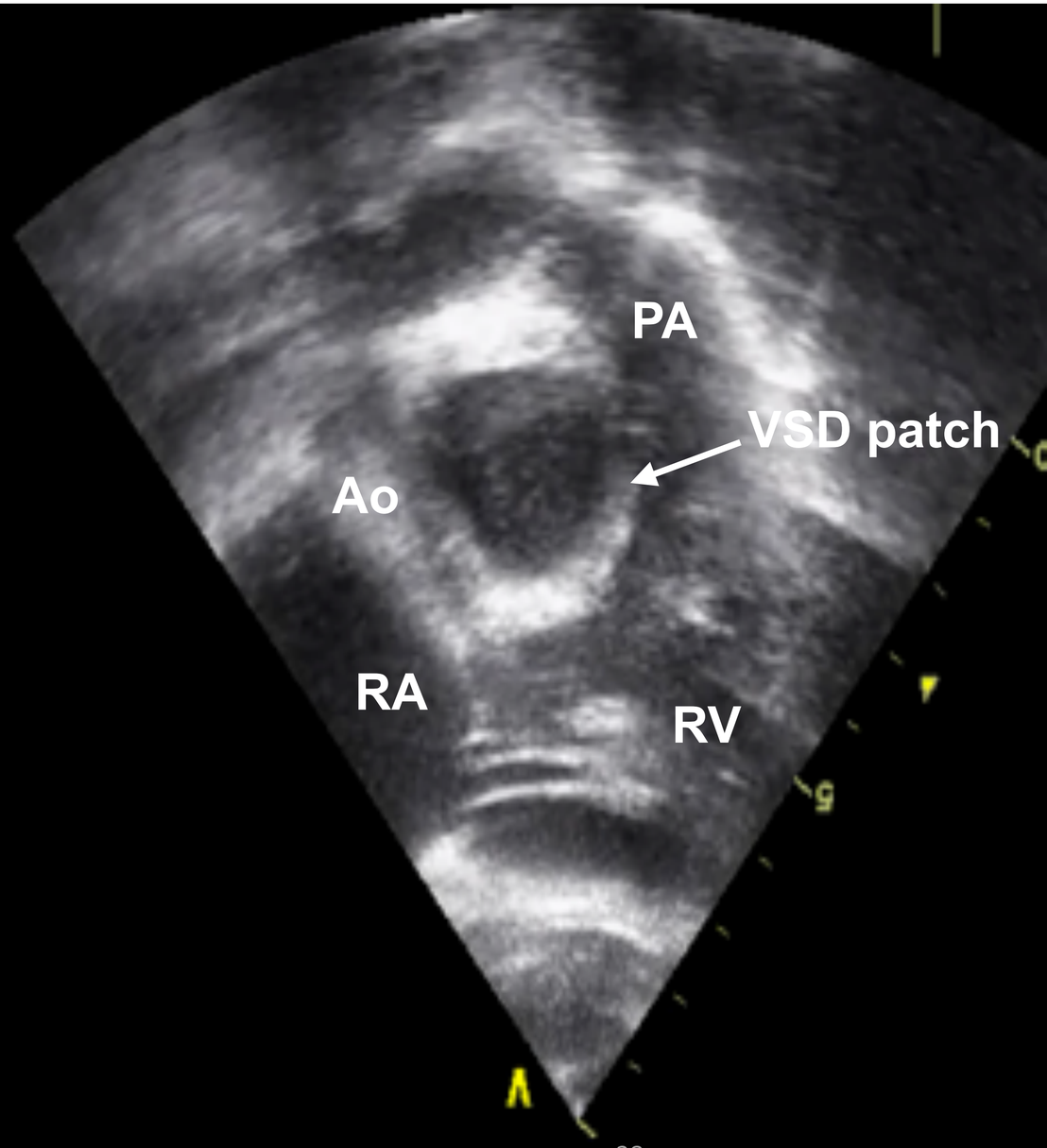




DORV

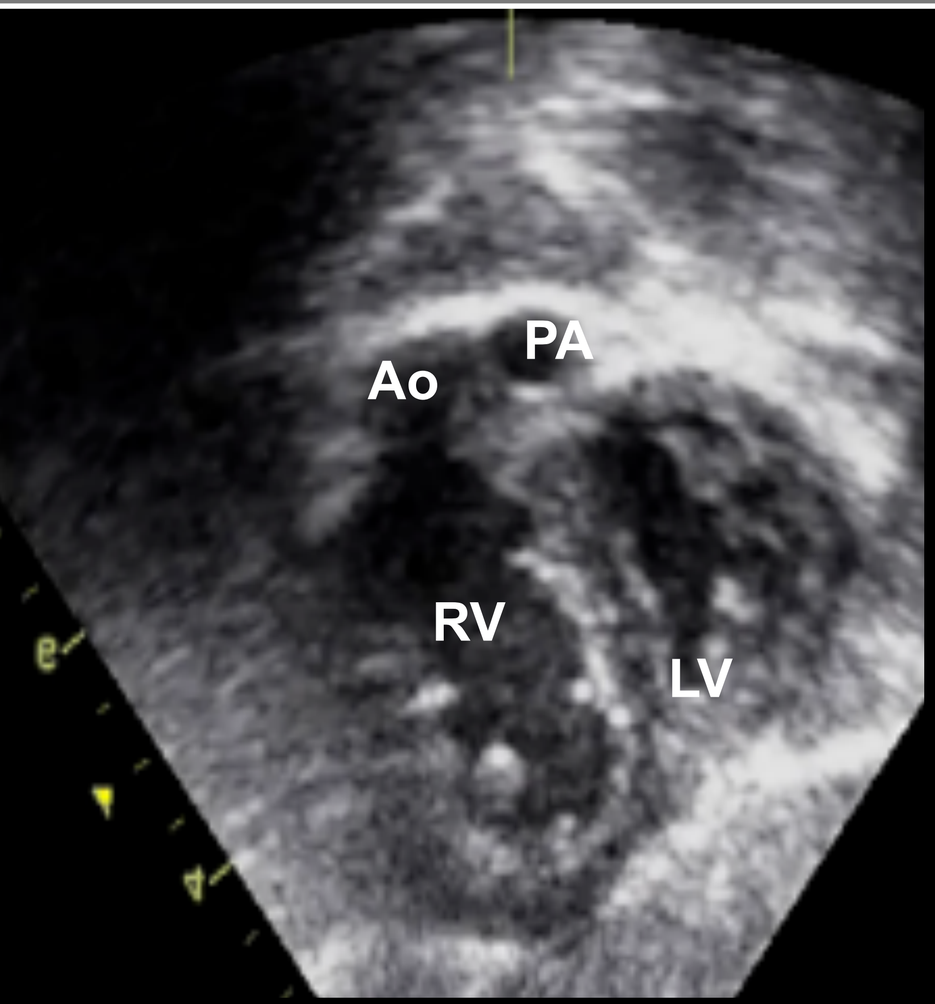
« Late » DORV sub aortic VSD-After IVR

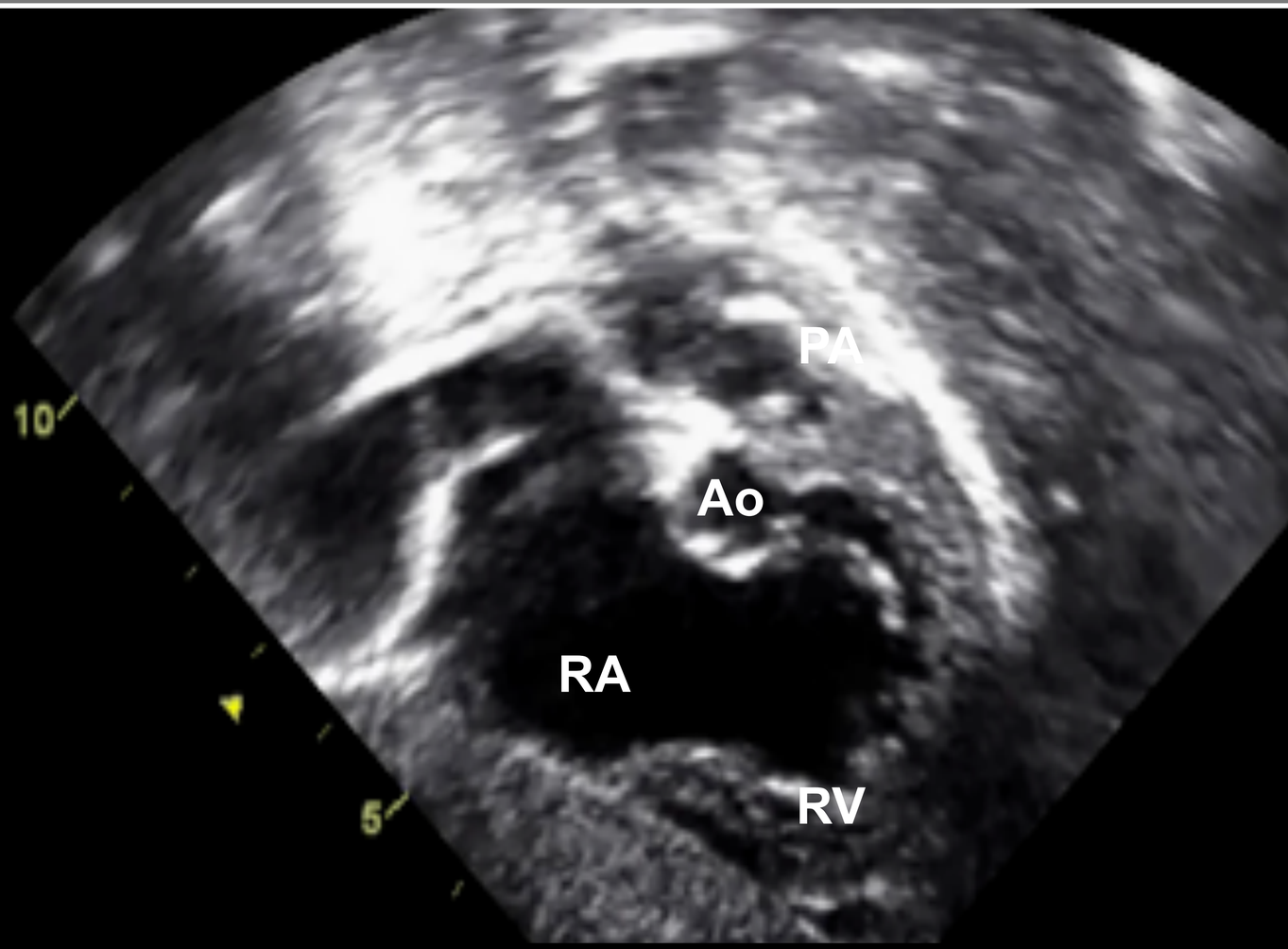




DORV

« Late » DORV sub aortic VSD + Pulmonary stenosis Fallot type





2. Is "anatomic" repair feasible ?

IVR may be possible but difficult :

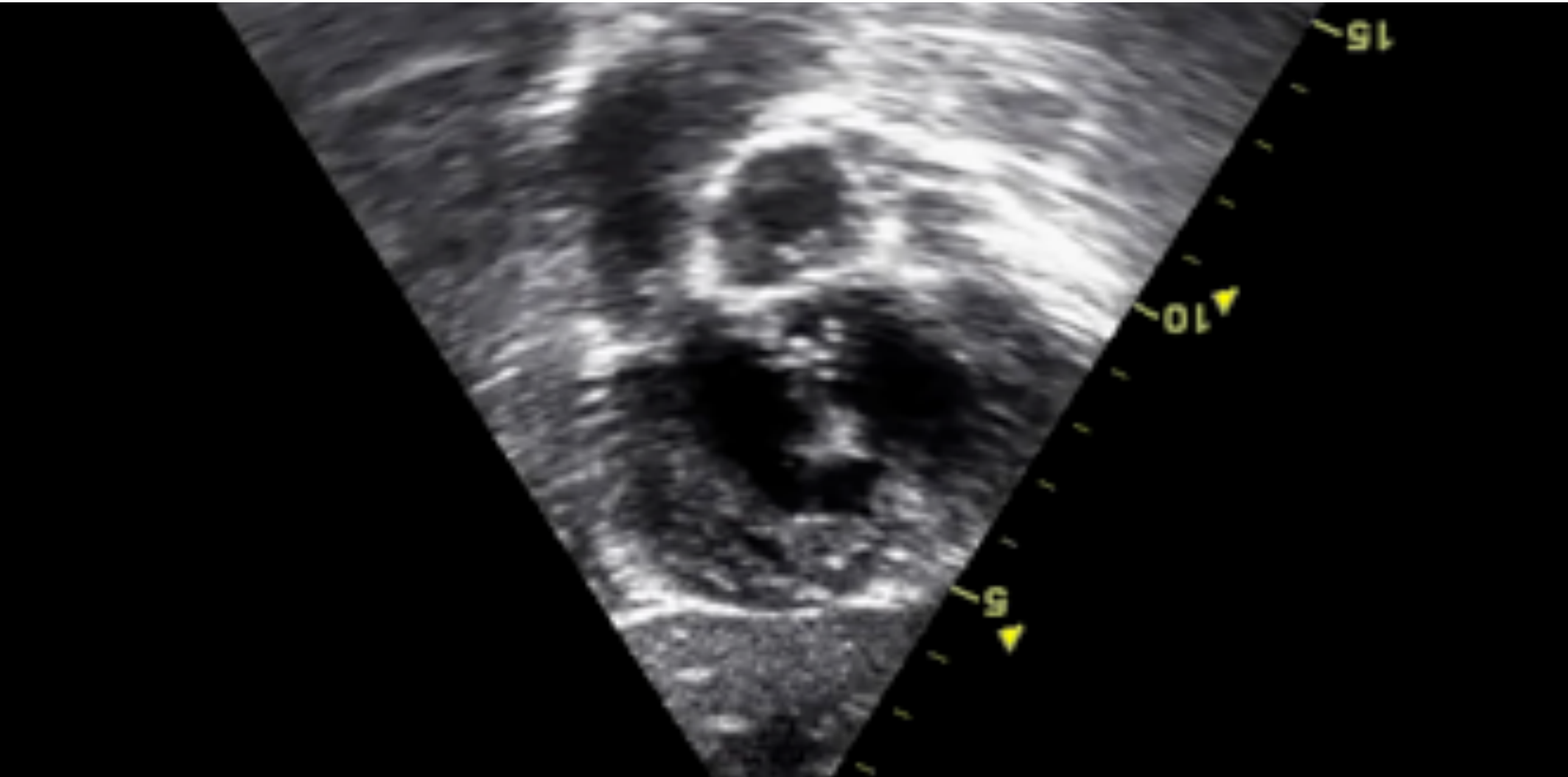
. extensive abnormal insertions of tricuspid and/or mitral valve

. asymmetric subpulmonary conus

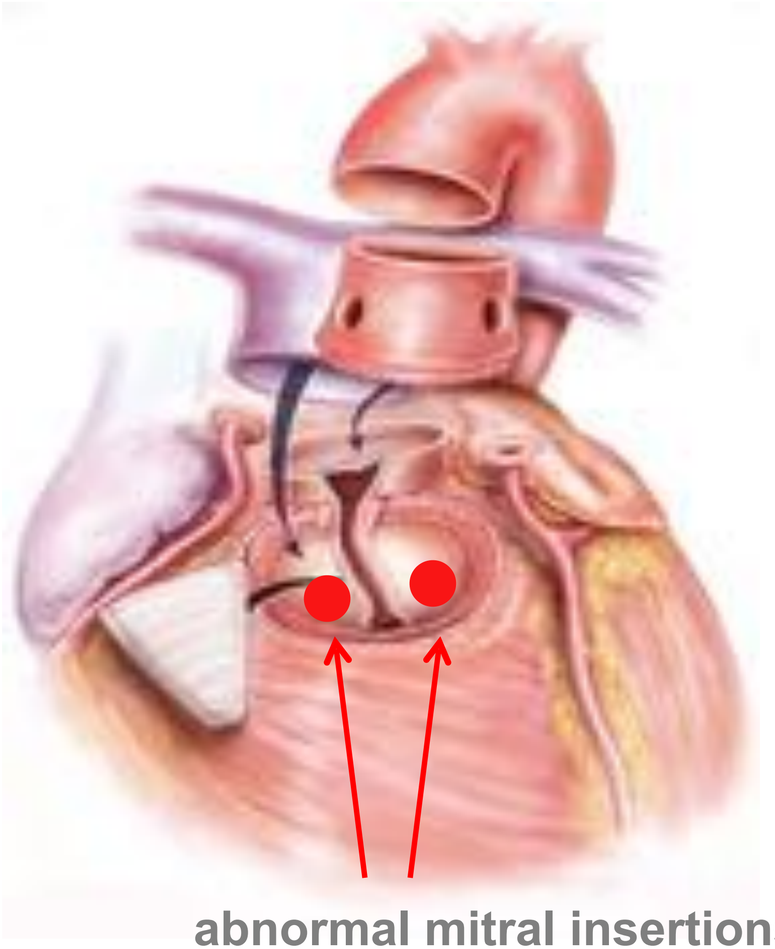
extra-anatomic repair may be preferable

DORV

« Late » DORV sub aortic VSD-Anormal tricuspid valve insertions



Abnormal insertions of mitral valve on conal septum



abnormal mitral insertions

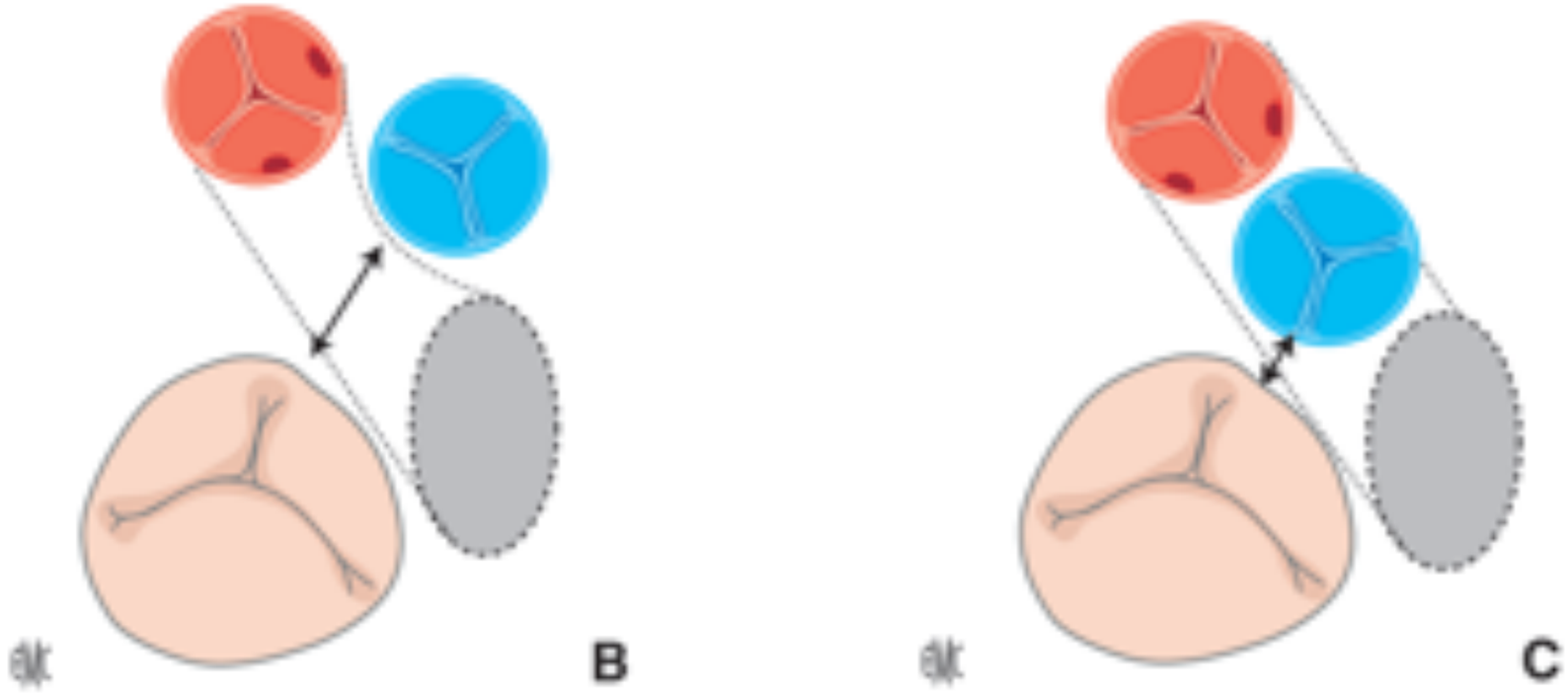
. rare

. difficult to manage

. resection of conal septum impossible

. Bex-Nikaidoh or cono-truncal rotation may be options in selected cases

Tricuspid-to-pulmonary distance < Ao diameter



IVR impossible

3. which extra-anatomic repair is indicated ?

- when "anatomic" repair (IVR) is impossible

- determinant: pulmonary outflow tract
  (particularly pulmonary valve)
  - normal
  - very abnormal (stenotic)
  - mildly abnormal (good enough for pulmonary)

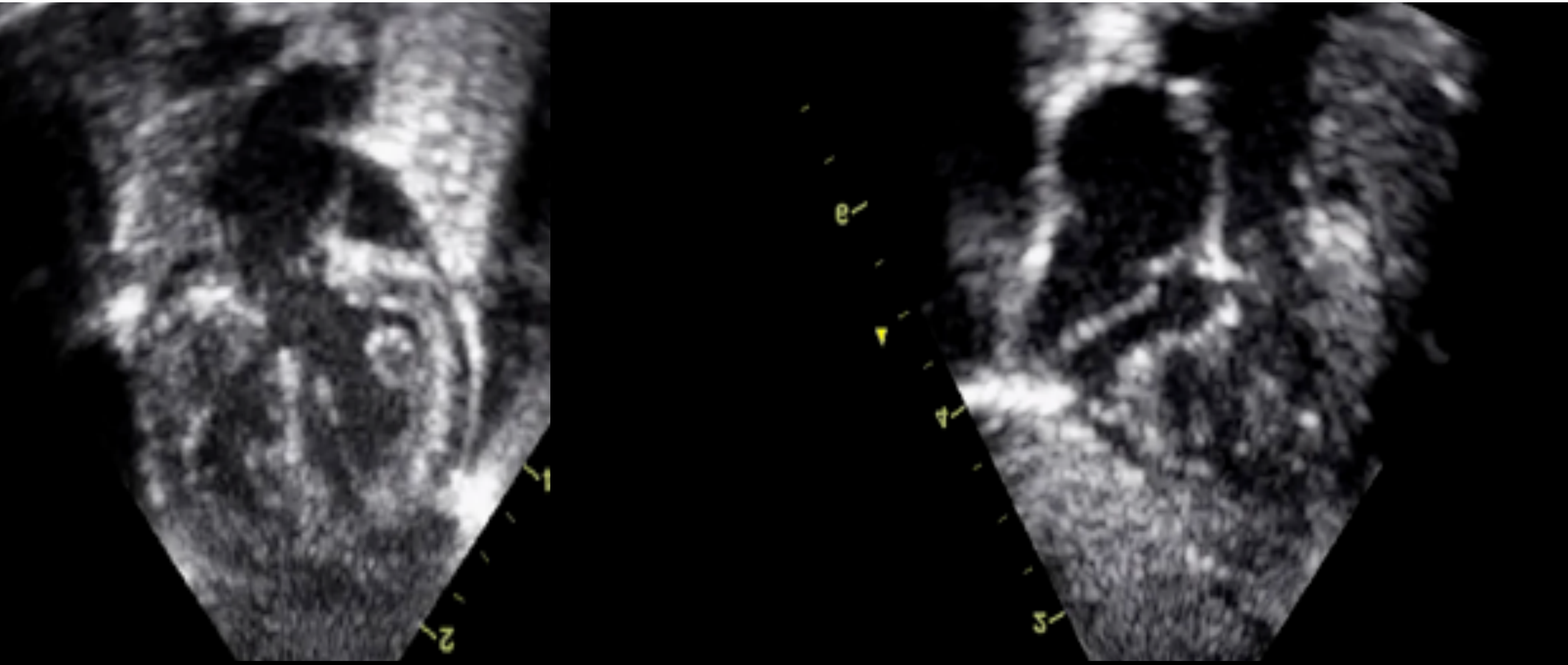
3. which extra-anatomic repair is indicated ?

- when LVOT can be used as neoaortic
  - normal pulmonary valve
  - subvalvar area
    - . normal
    - . stenosis which can be relieved

- **LV to PA connection + arterial switch**

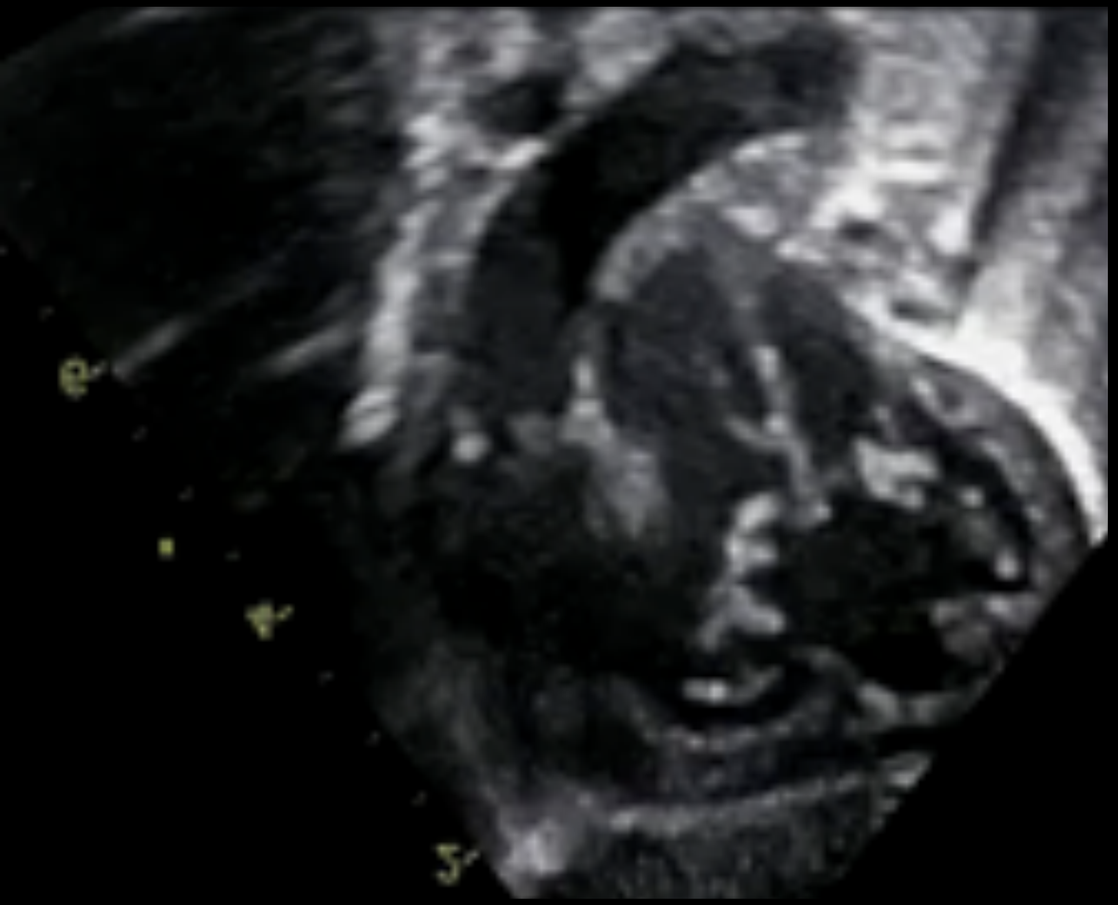
DORV

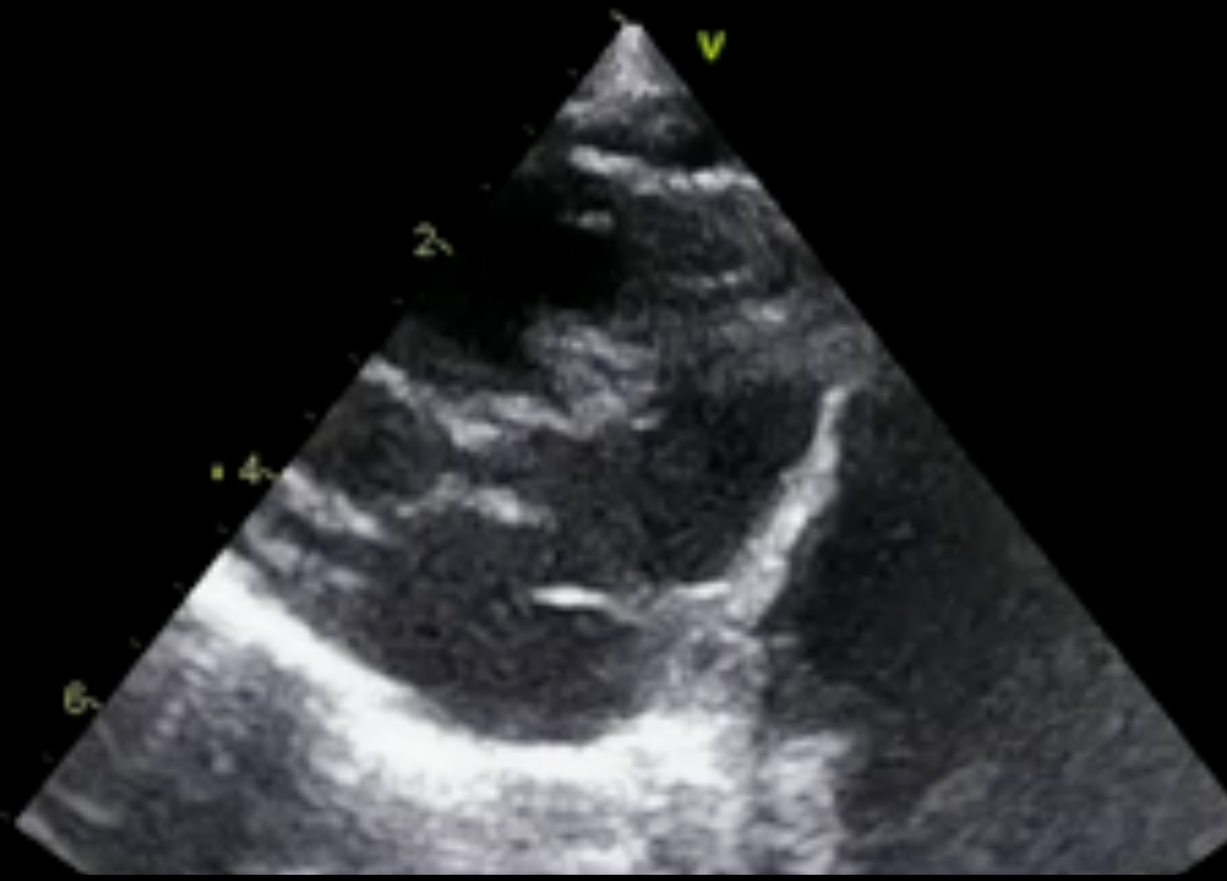
« Late » DORV -Short Tricuspid-Pulmonary valve distance



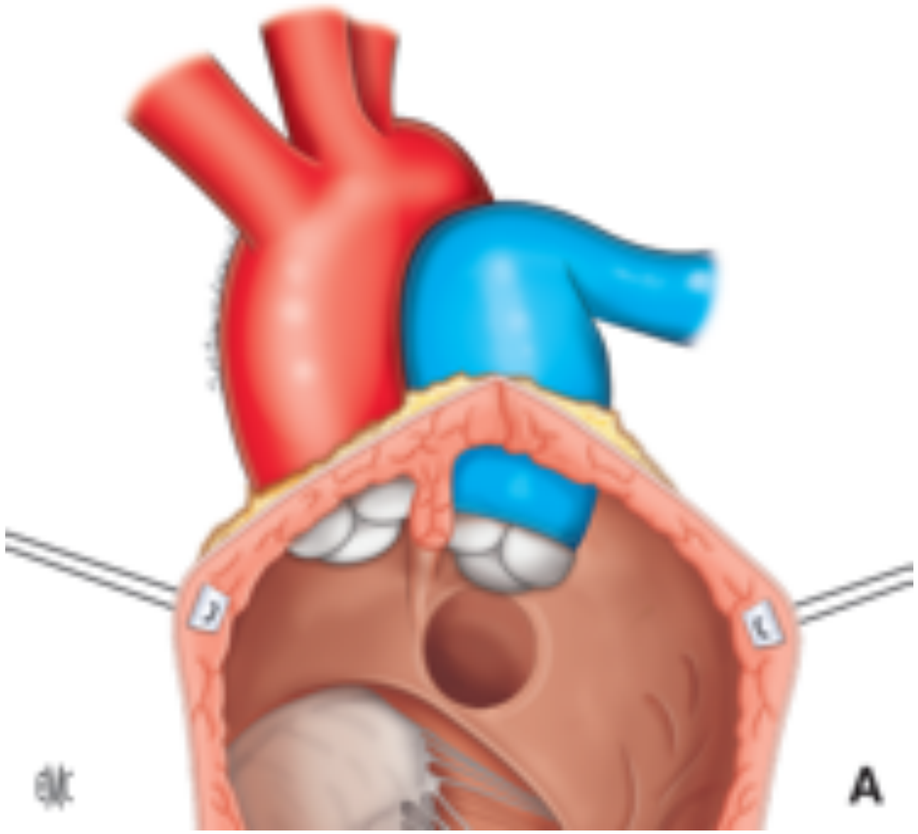
DORV

« Late » DORV -Sub-pulmonary VSD

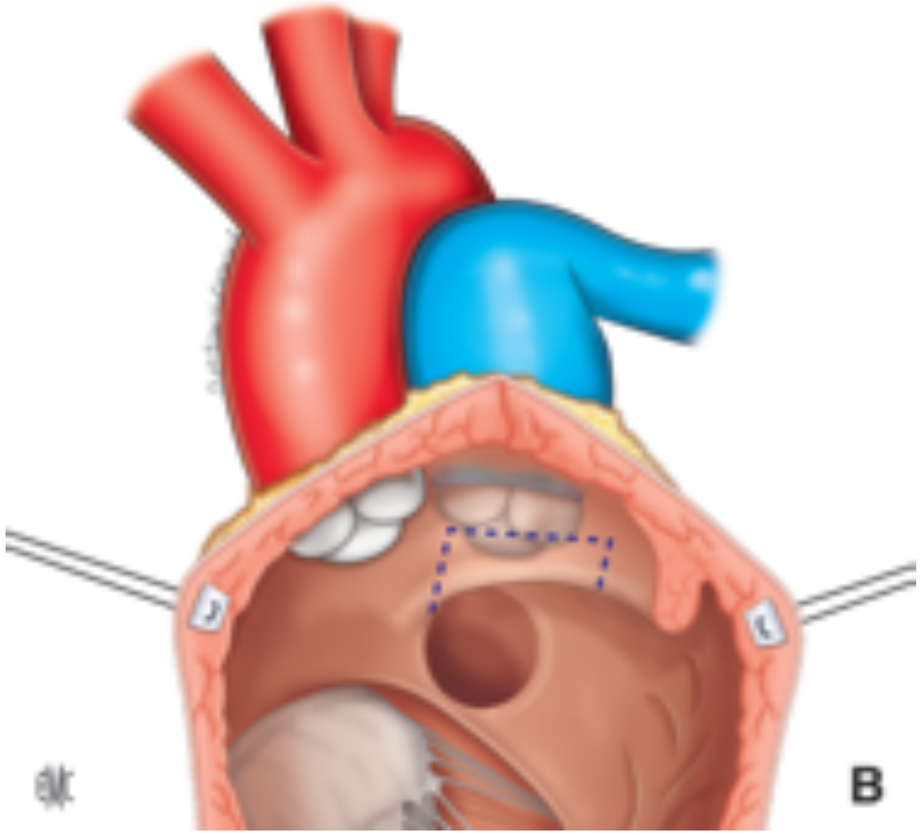




LV-PA connection + arterial switch



A



B

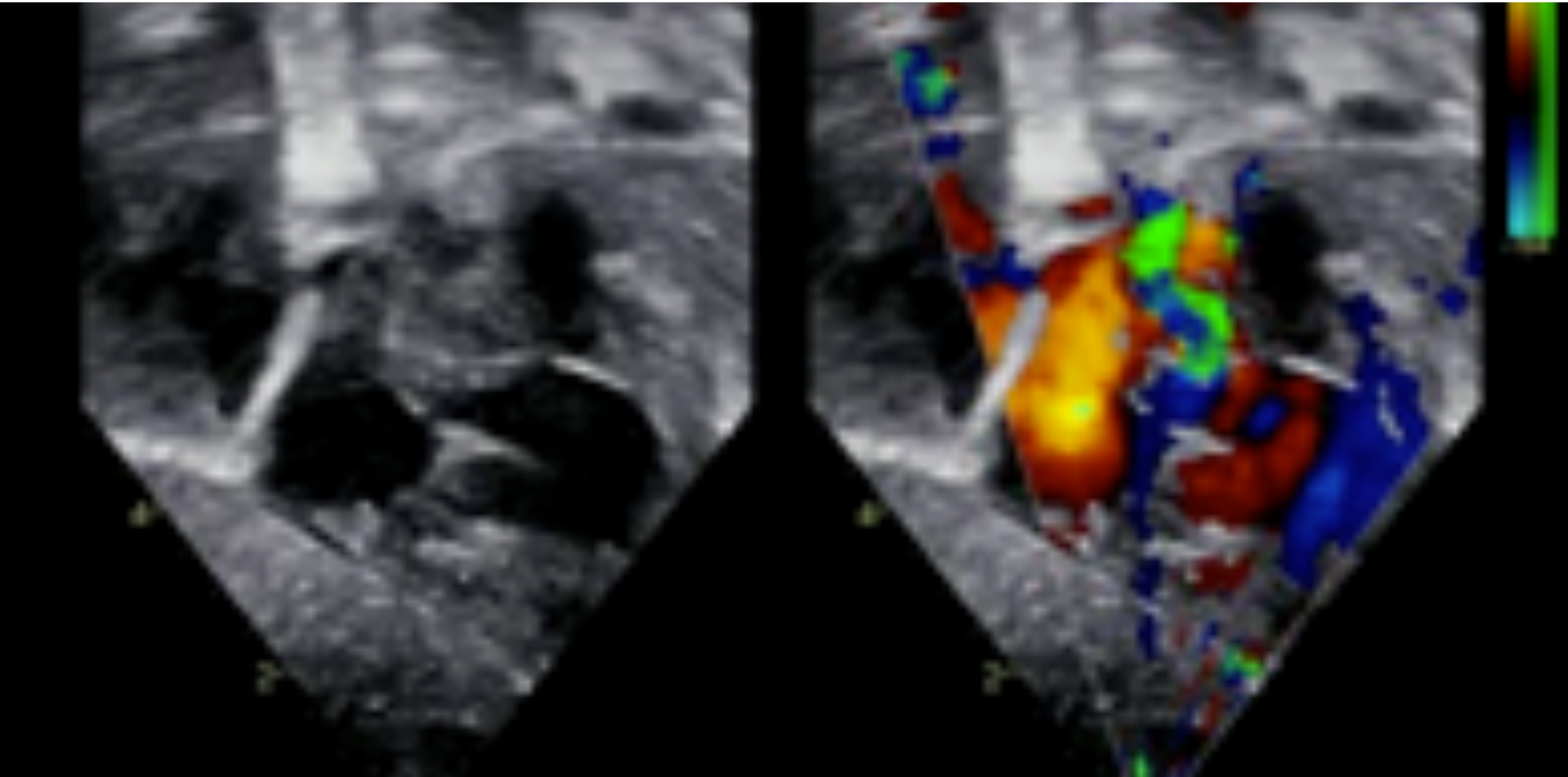
3. which extra-anatomic repair is indicated ?

- when LVOT cannot be used as neoaortic
  - severe valvar stenosis
  - non-resectable subvalvar obstruction

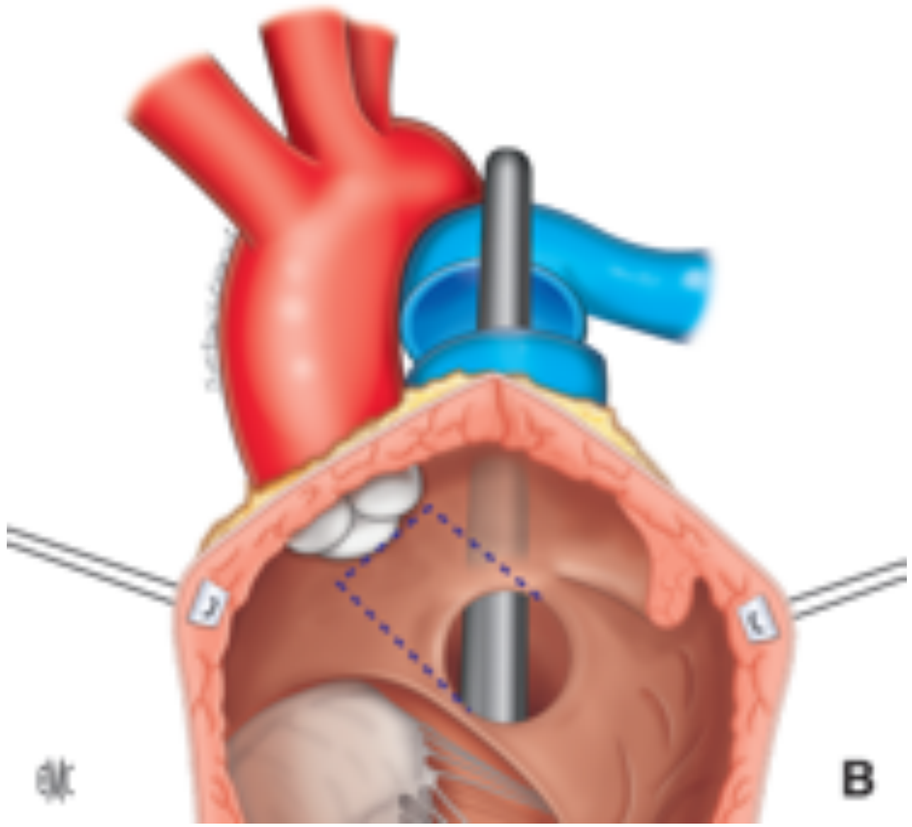
- **REV operation**
- **Bex-Nikaidoh operation**
- **Rastelli operation**

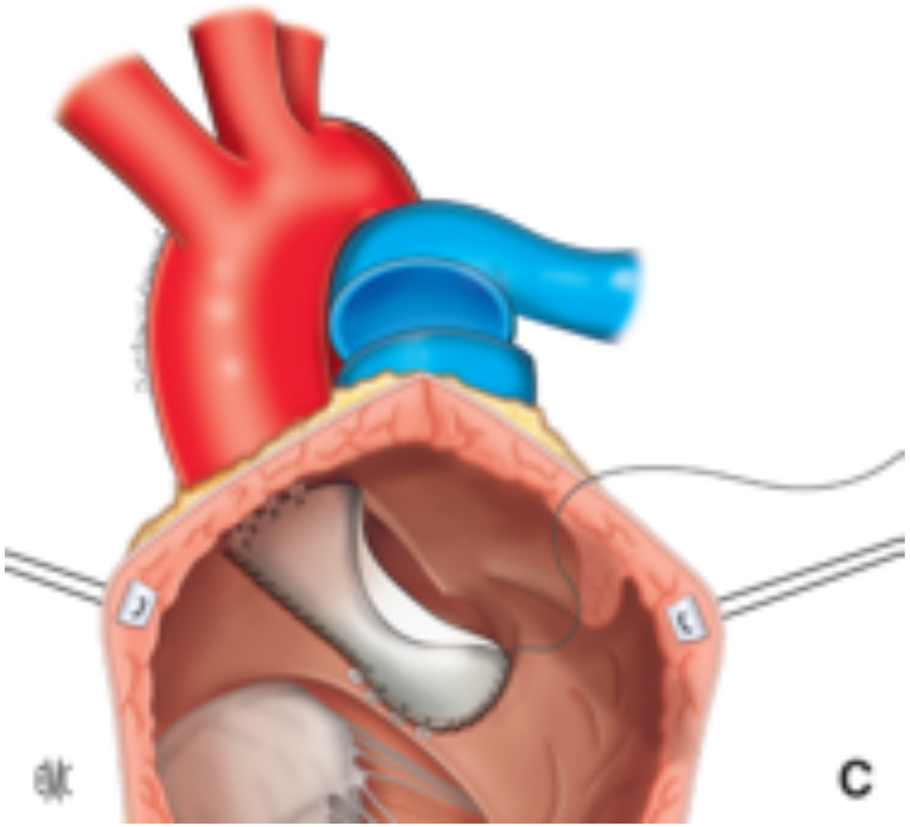
DORV

« Late » DORV -Short Tricuspid-Pulmonary valve distance-Severe subpulmonary stenosis

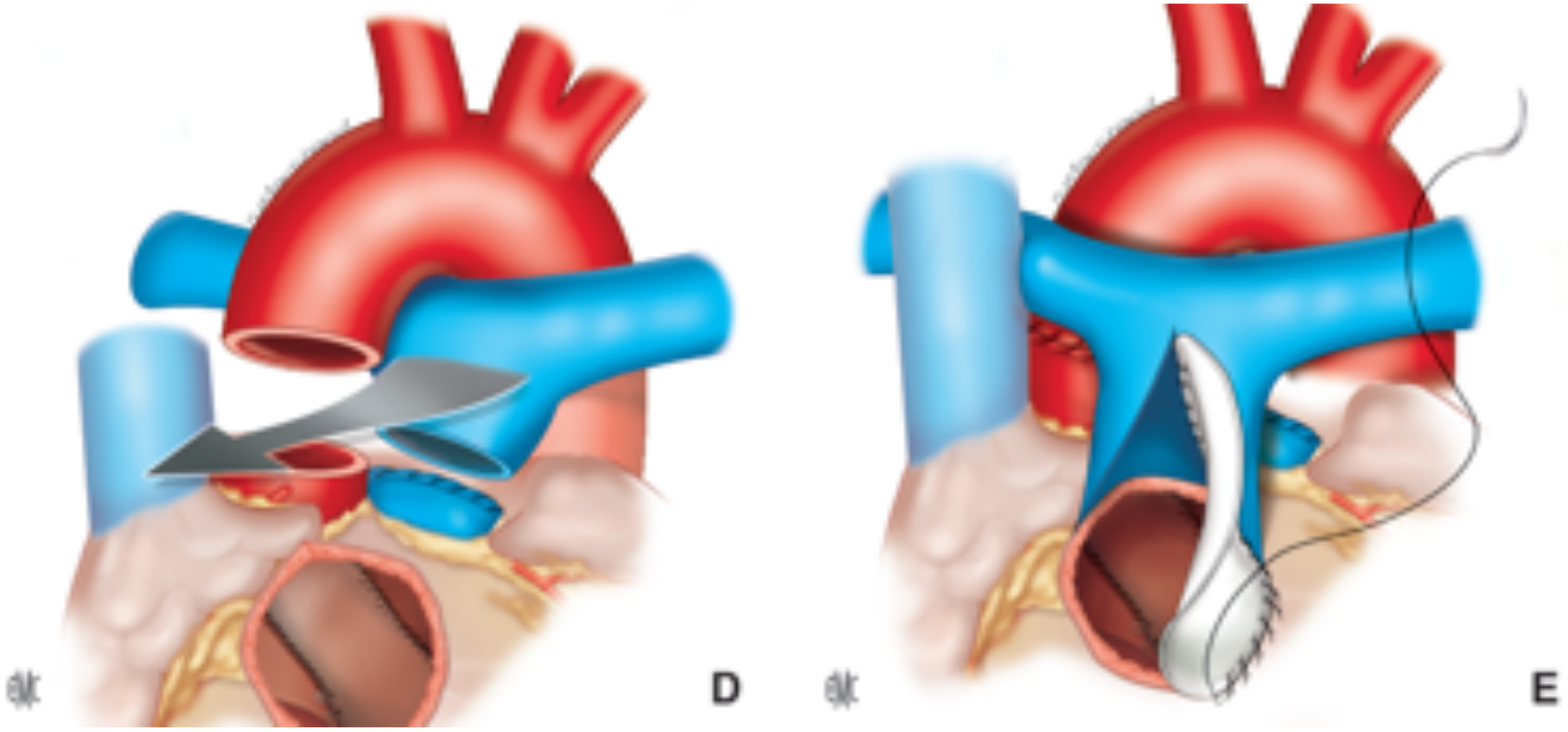


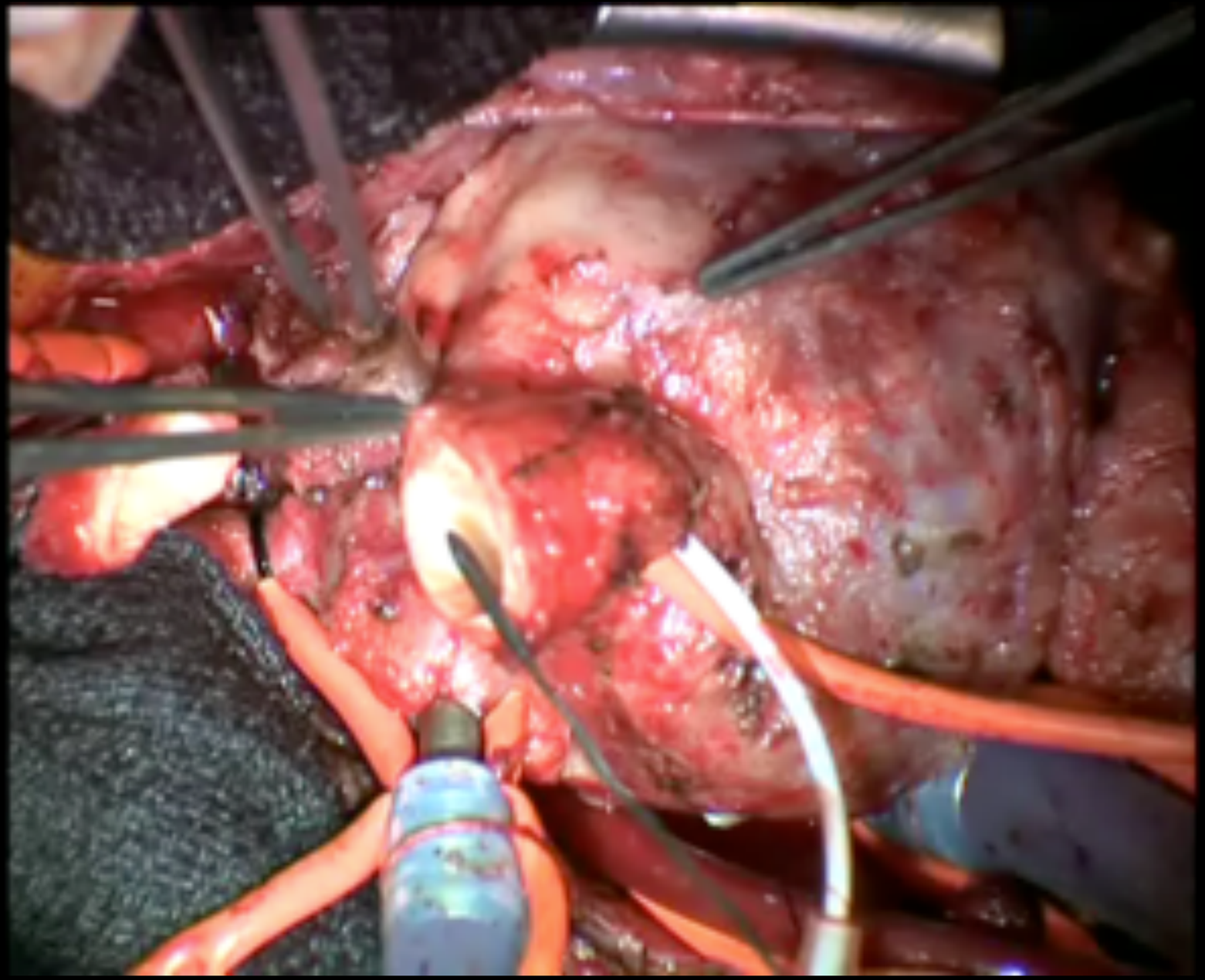
Extra-anatomic repair : REV



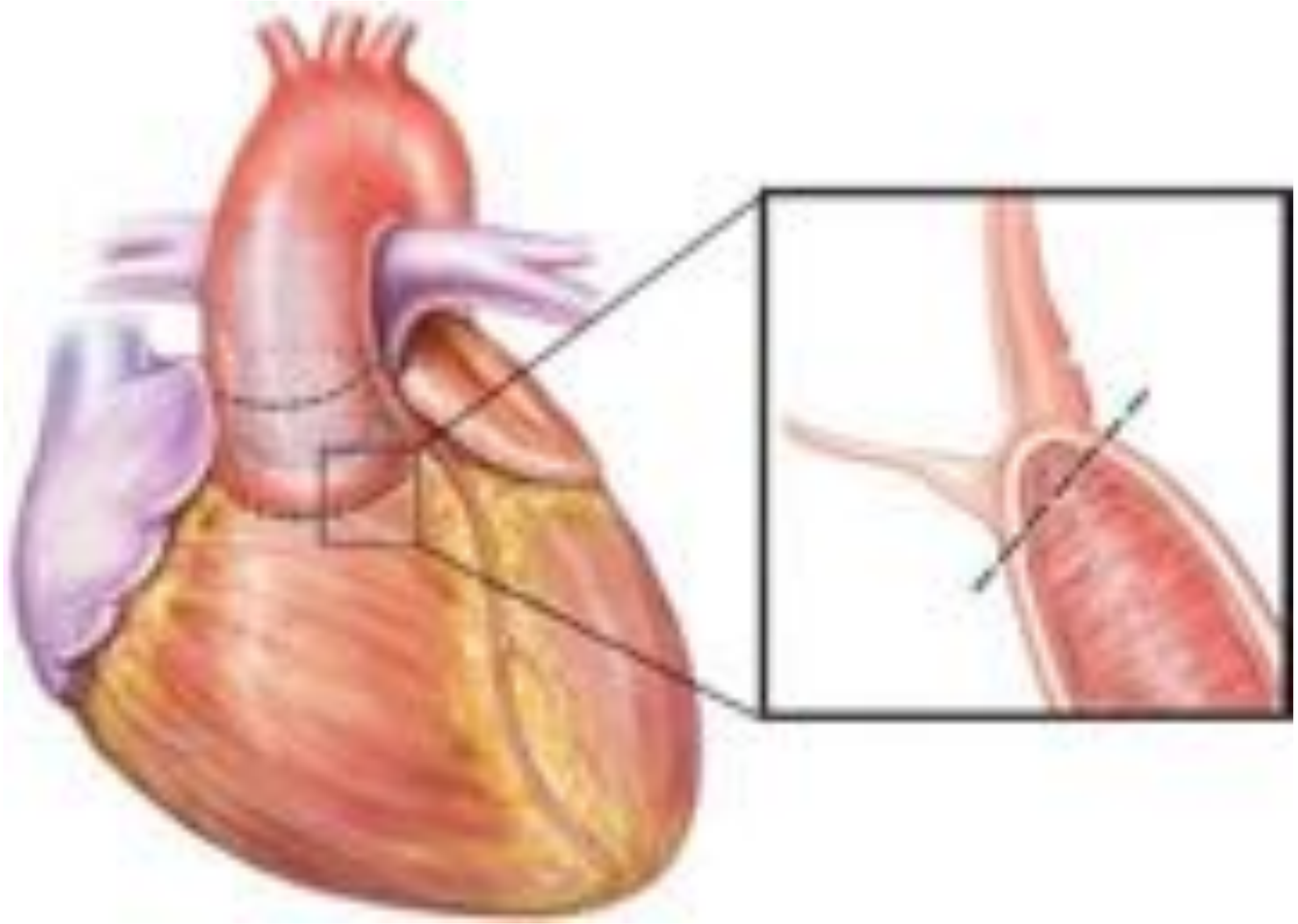


Extra-anatomic repair : REV

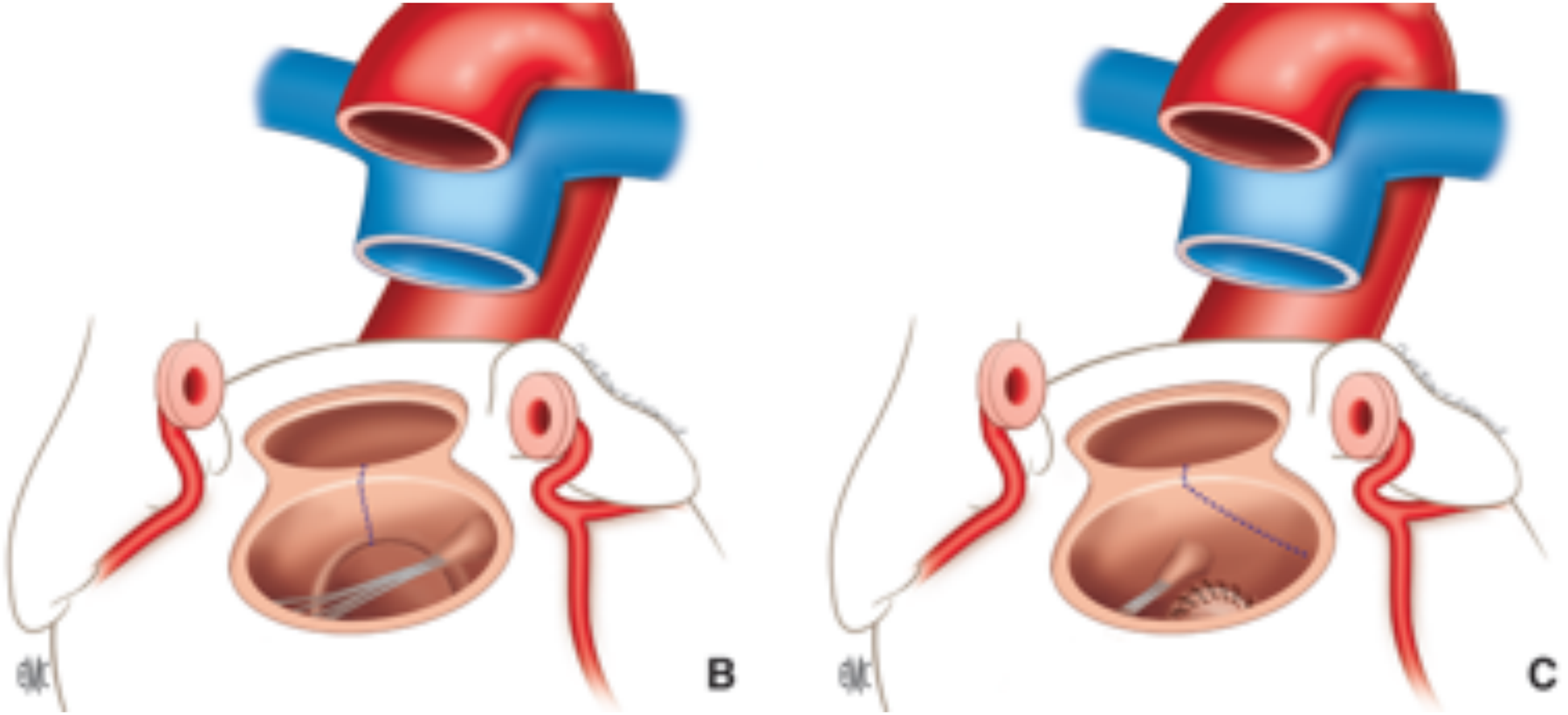




Extra-anatomic repair
Bex-Nikaidoh operation

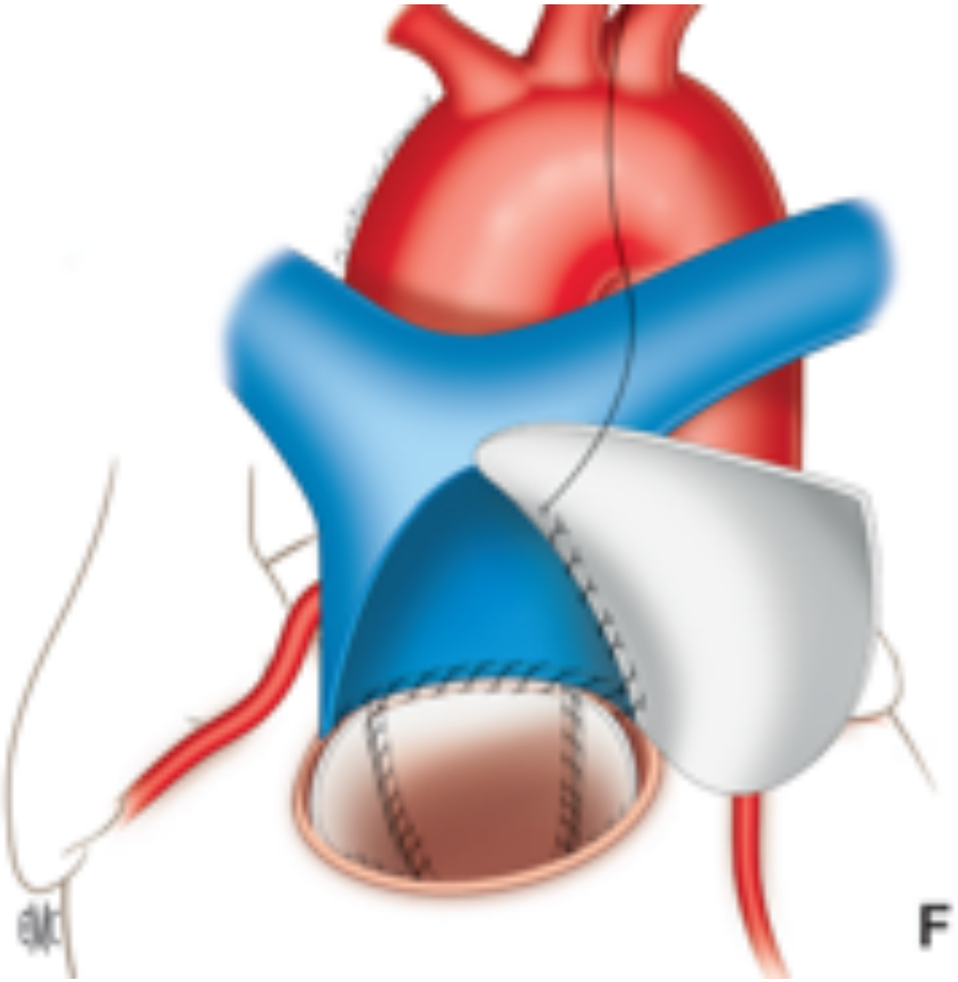


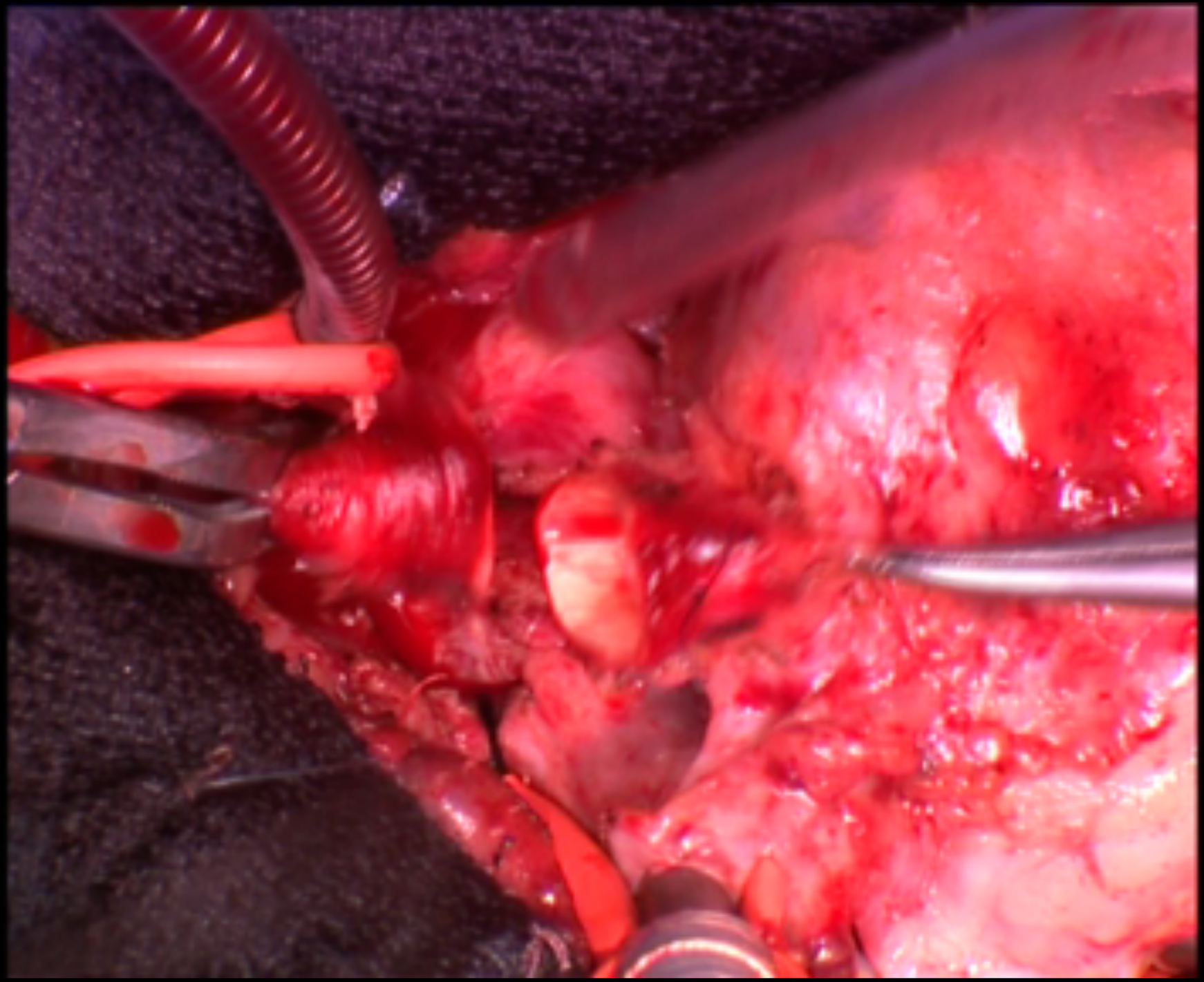
Extra-anatomic repair
Bex-Nikaidoh operation



Extra-anatomic repair
Bex-Nikaidoh operation

Extra-anatomic repair
Bex-Nikaidoh operation





Indications for Bex-Nikaidoh operation

when REV is difficult / impossible

- abnormal insertions of mitral valve on conal septum
- extensive abnormal insertions of tricuspid valve
- remote VSD (inlet, muscular)
- absence of VSD

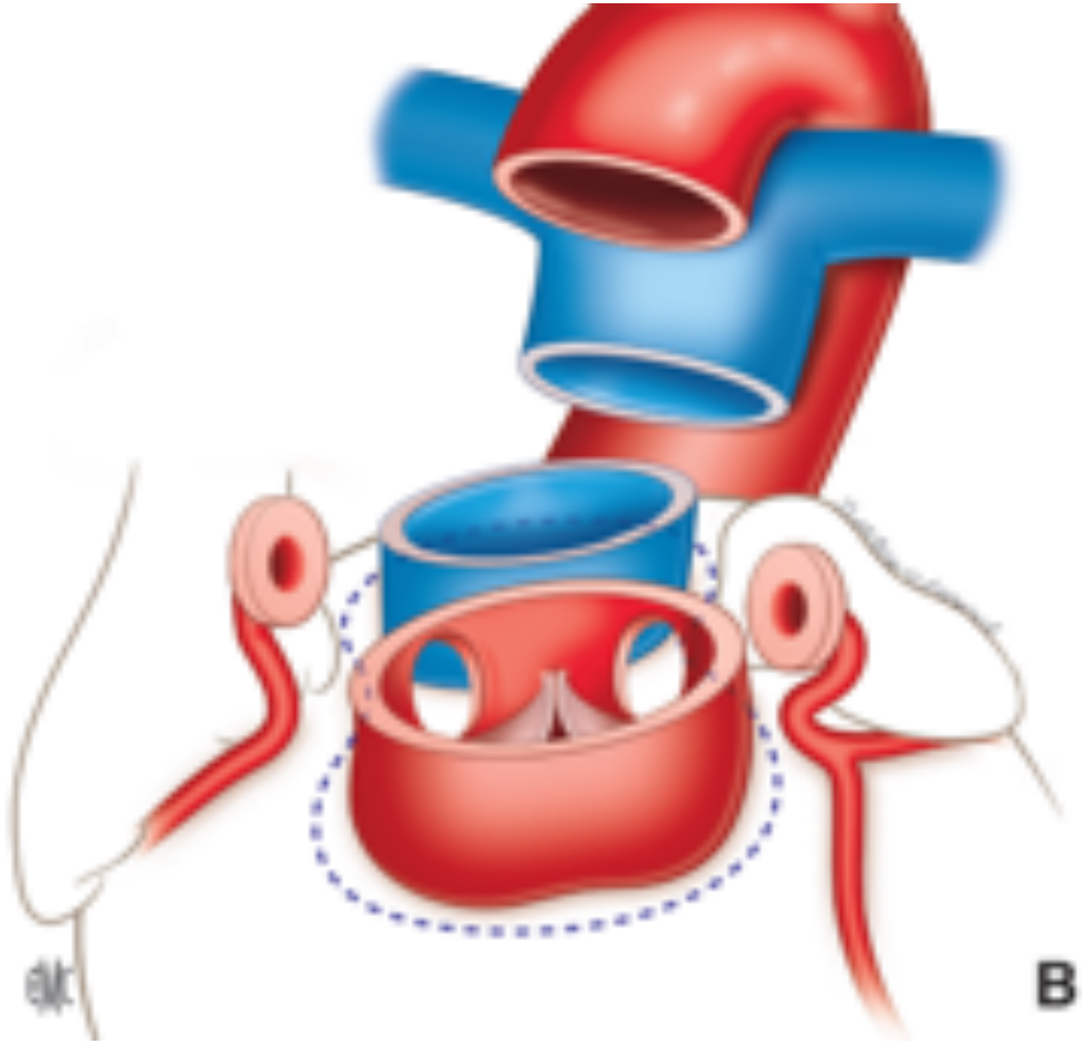
- coronary anatomy must allow Bex-Nikaidoh
  (no anterior loop)

3. which extra-anatomic repair is indicated ?

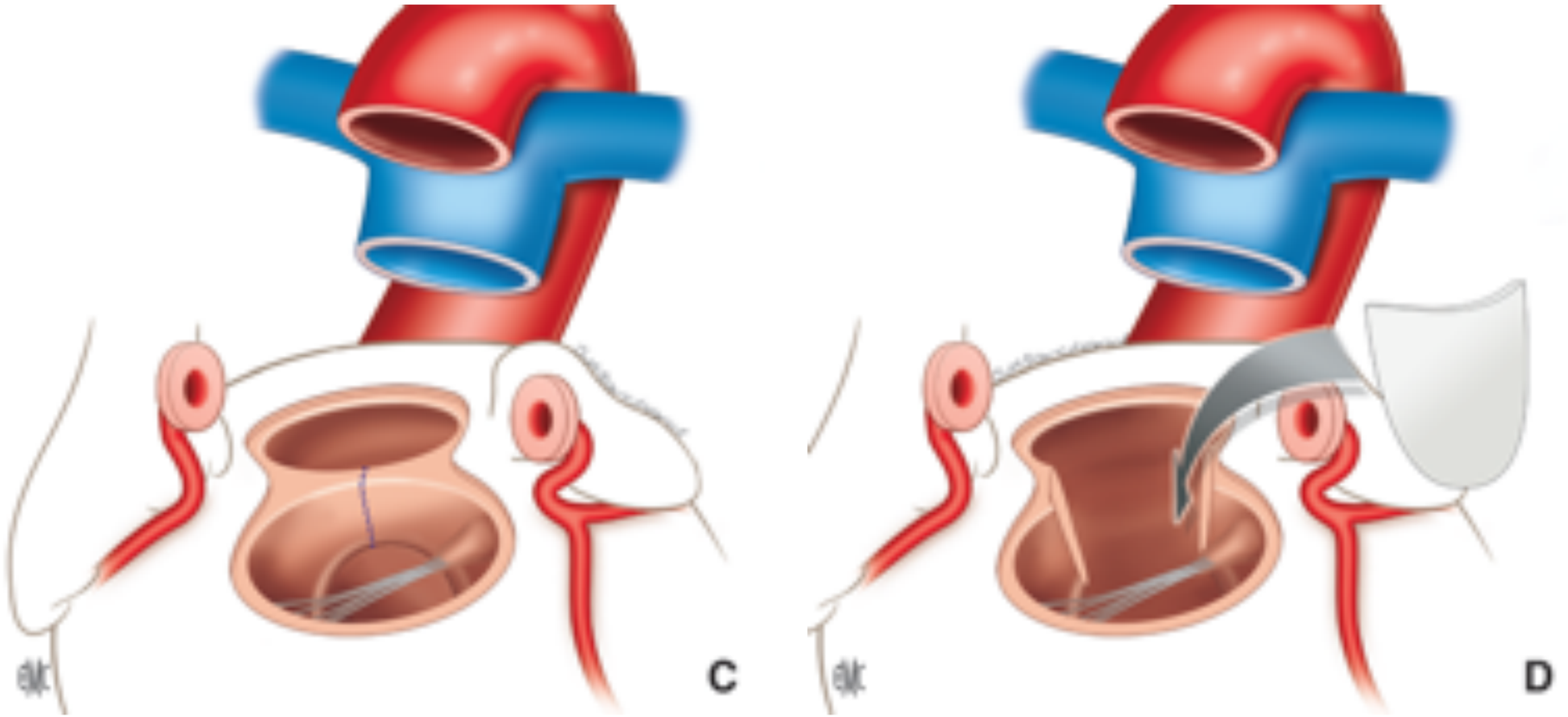
- when LVOT cannot be used as neoaortic
  but can be used as pulmonary
  - bicuspid pulmonary valve
  - mildly-dysplastic pulmonary valve

- **conotruncal rotation procedure**
  (if allowed by coronary anatomy)

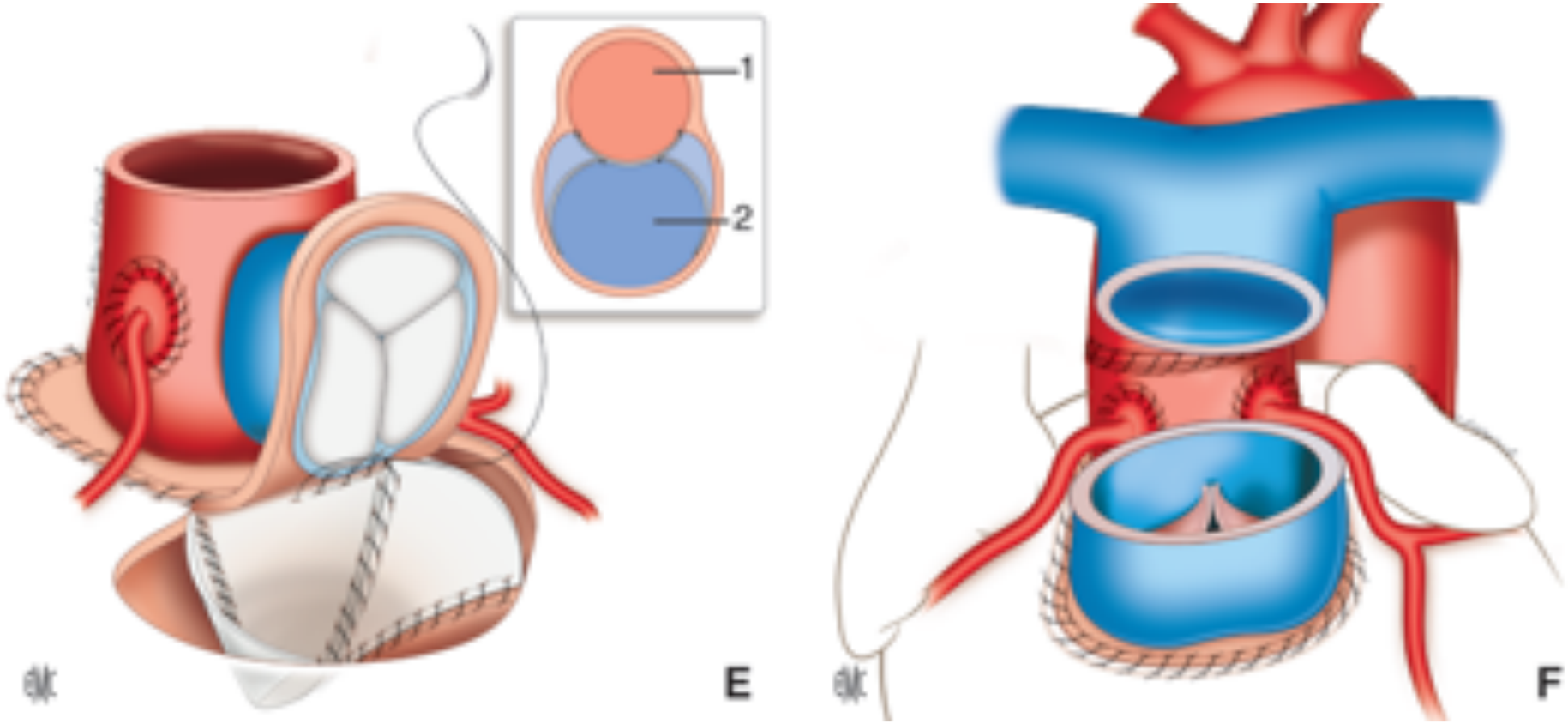
Conotruncal rotation procedure

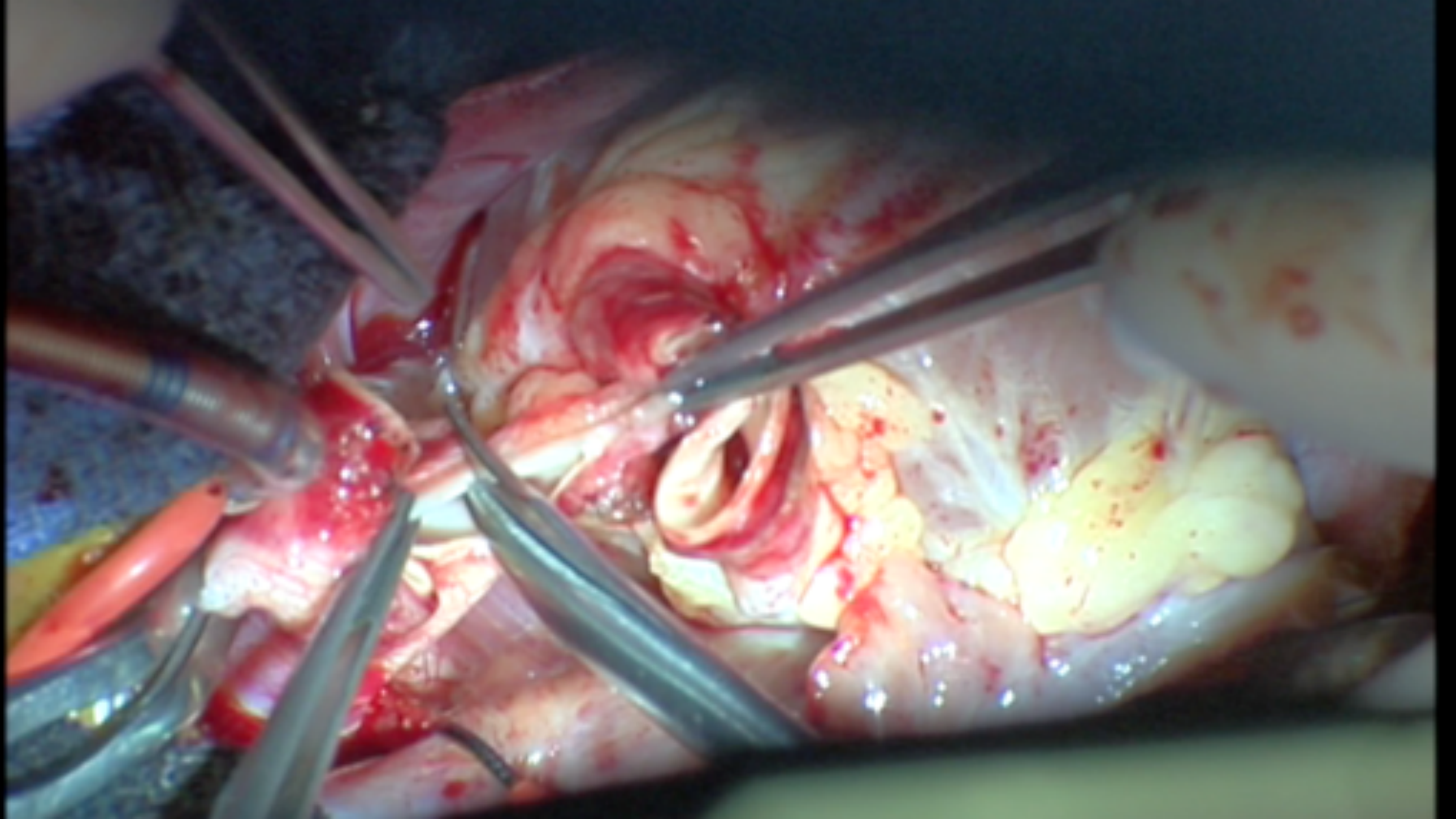


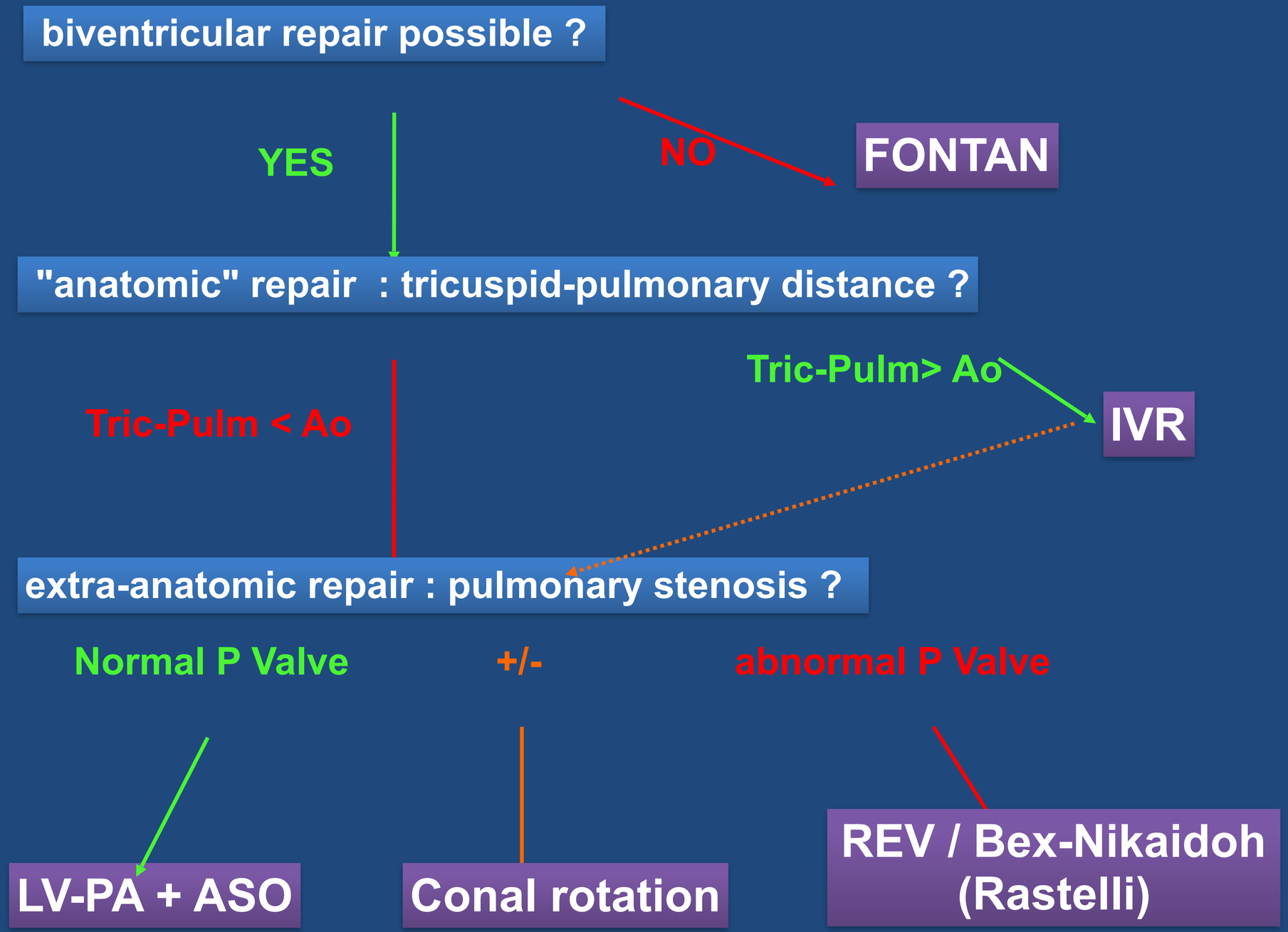
Conotruncal rotation procedure



Conotruncal rotation procedure






biventricular repair possible ?
YES
NO
FONTAN
"anatomic" repair : tricuspid-pulmonary distance ?
Tric-Pulm < Ao
Tric-Pulm> Ao
IVR
extra-anatomic repair : pulmonary stenosis ?
Normal P Valve
+/-
abnormal P Valve
LV-PA + ASO
Conal rotation
REV / Bex-Nikaidoh (Rastelli)

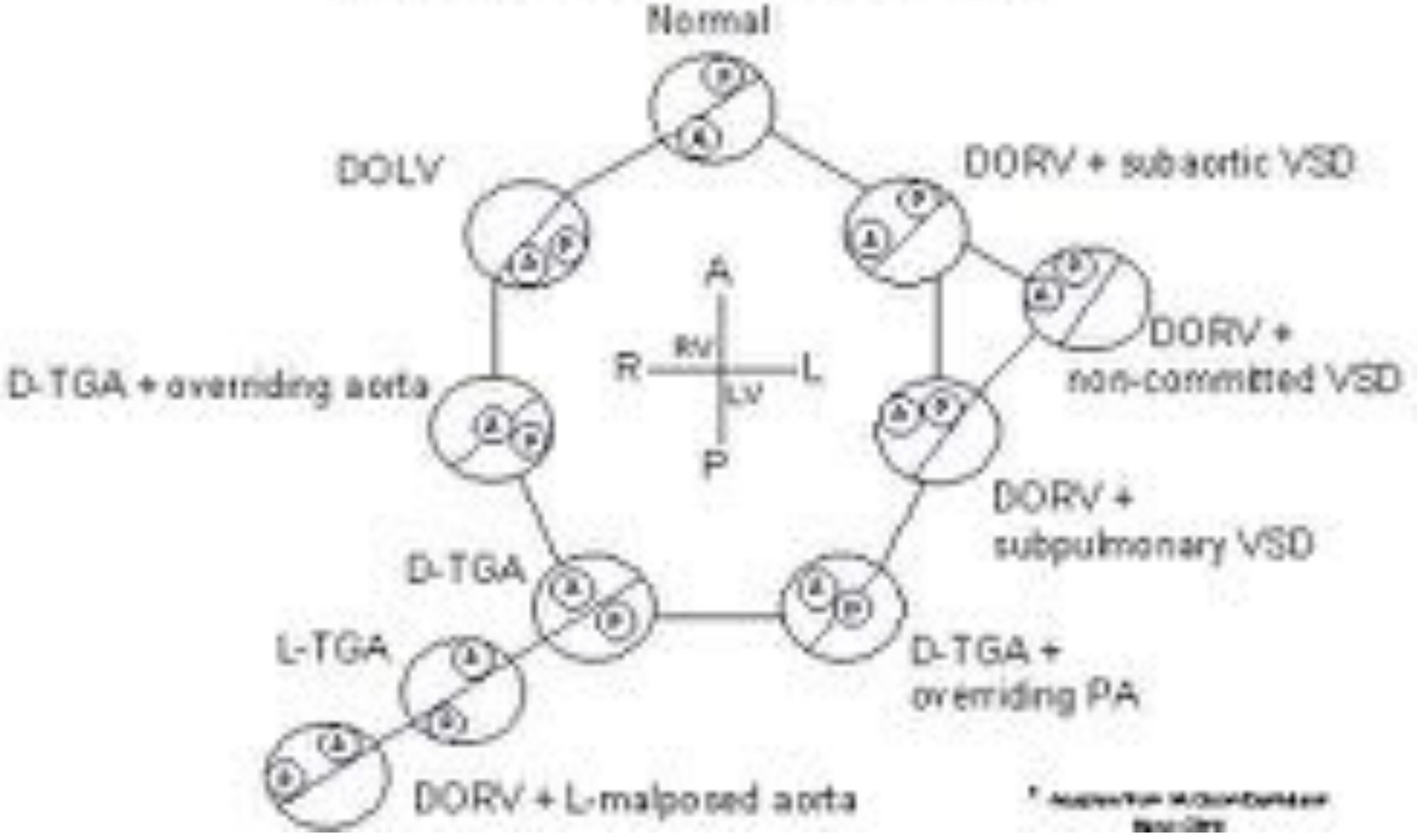

M3C
Thank you

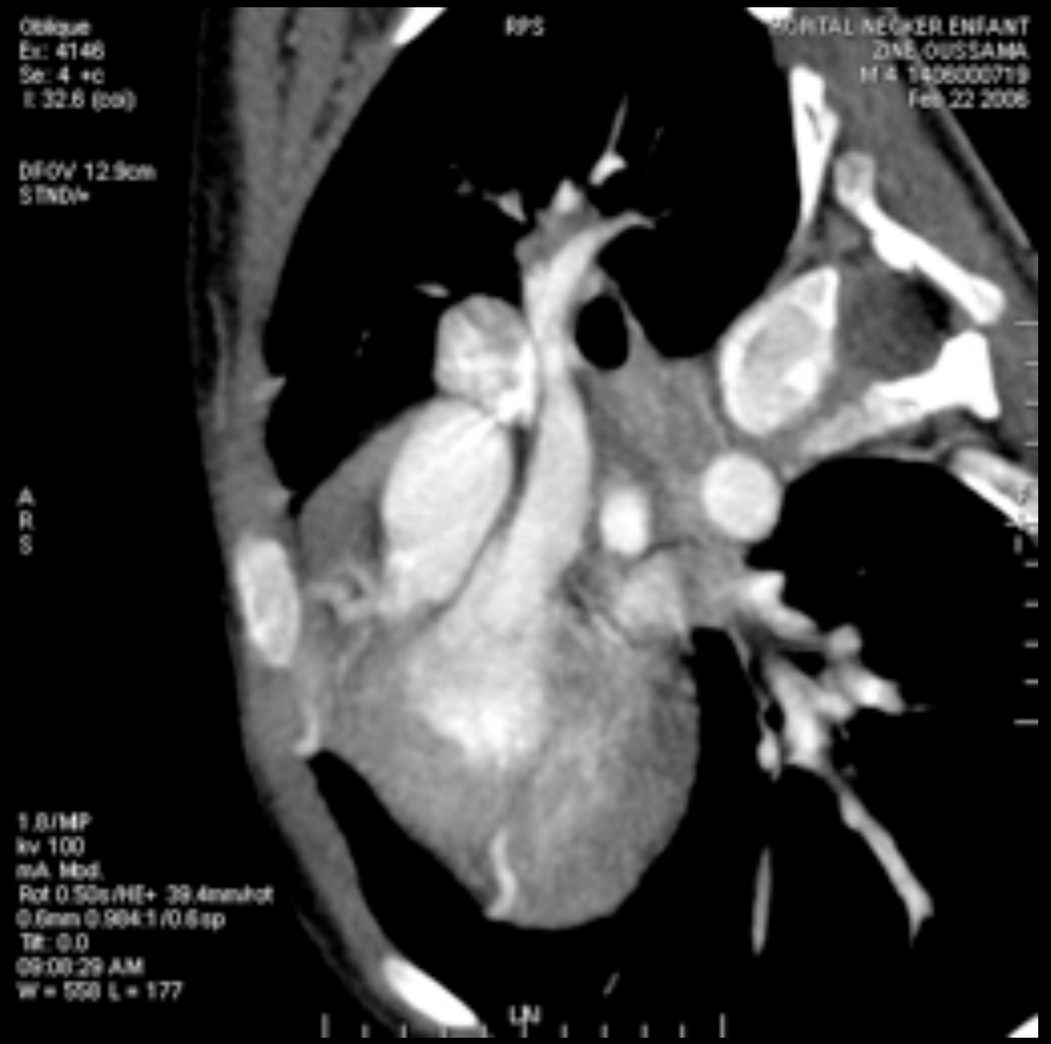
Operative risk (2007-2014)

	NEM	EACTS
IVR	*54 (0%)*	*530 (7%)*
Rastelli	*4 (0%)*	*105 (5.7%)*
REV	*41 (0%)*	*63 (4.8%)*
Bex-Nikaïdoh	*12 (8.3%)*	*37 (5.4%)*
conal rotation	*6 (0%)*	*---*

Positional Anomalies of Conotruncus

Atrioventricular Concordance*




Oblique
Ex: 4146
Se: 4 +c
I: 32.6 (cal)
DFOV 12.9cm
STND+
A
R
S
1.8/1MP
kv 100
mA Mod.
Rot 0.50s/HE+ 39.4mm/rot
0.6mm 0.984:1/0.6sp
Tilt: 0.0
09:08:29 AM
W = 558 L = 177
RPS
HOPITAL NECKER ENFANT
ZINE OUSSAMA
M 4 1806000719
Feb 22 2006


Oblique
Ex: 4146
Se: 4 +c
L: 1.0 (bol)
DFOV 12.9cm
STND+
SPR
HOPITAL NECKER ENFANT
ZINE OUSSAMA
M 4 1406000719
Fev 22 2006
R
A
S
1.8/MP
kv 100
mA Mod.
Rot 0.50s/HE+ 39.4mm/rot
0.6mm 0.984:1/0.6sp
Tilt: 0.0
09:08:29 AM
W = 558 L = 177