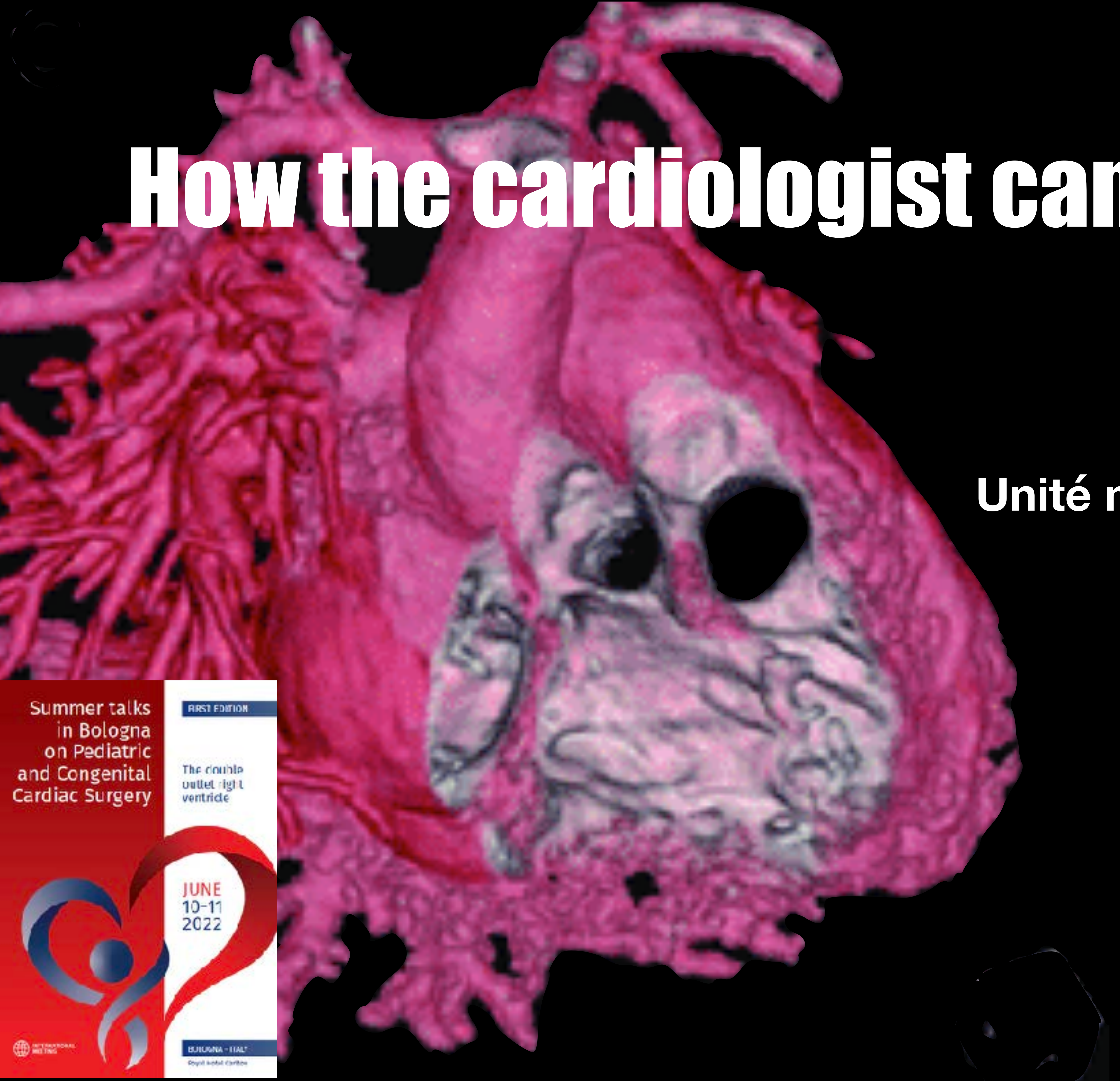

Double outlet right ventricle

How the cardiologist can influence the surgical strategy ?

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Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN

M3C
ARCFA
Association pour la Recherche en Cardiologie du Fœtus à l'Adulte
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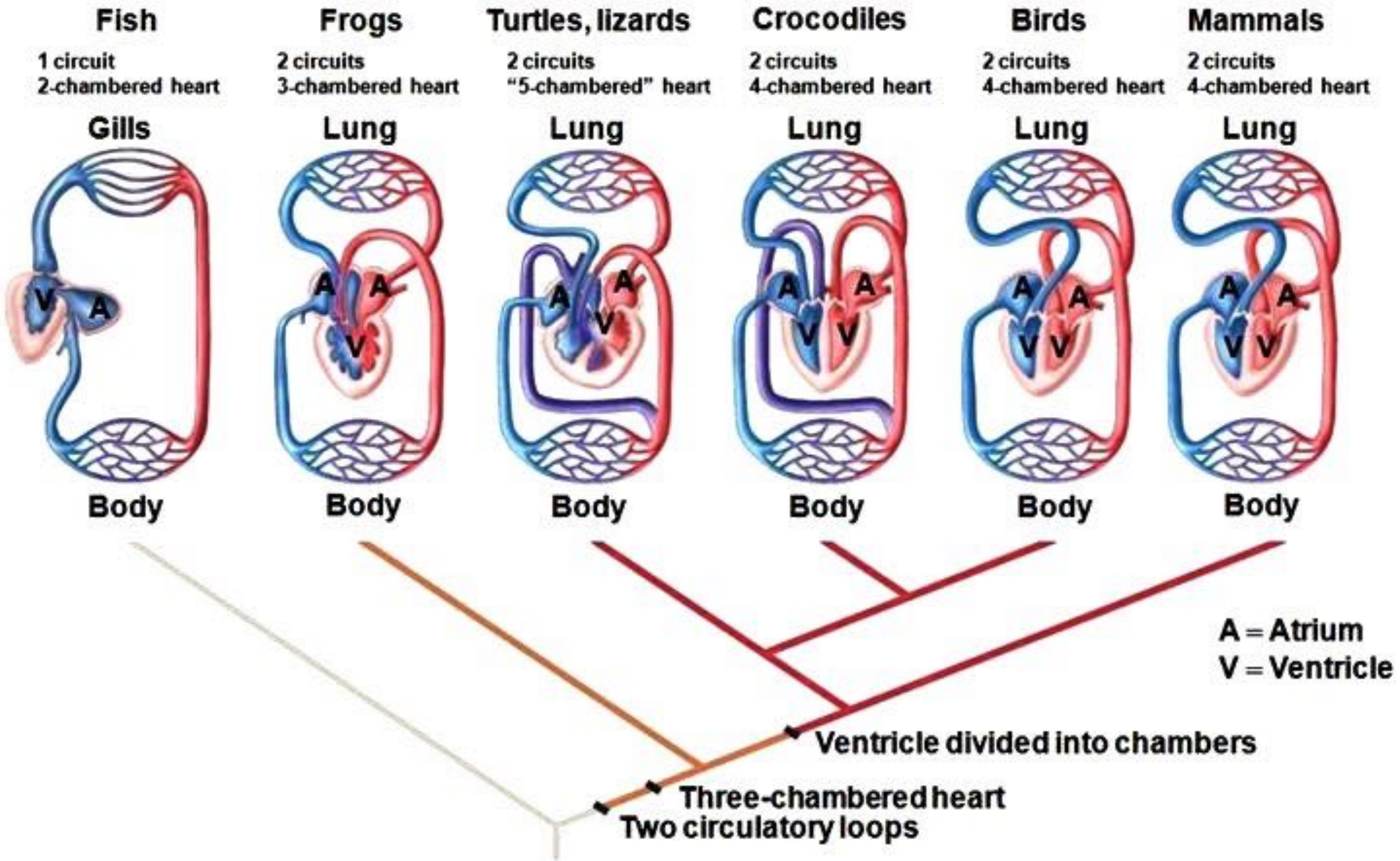
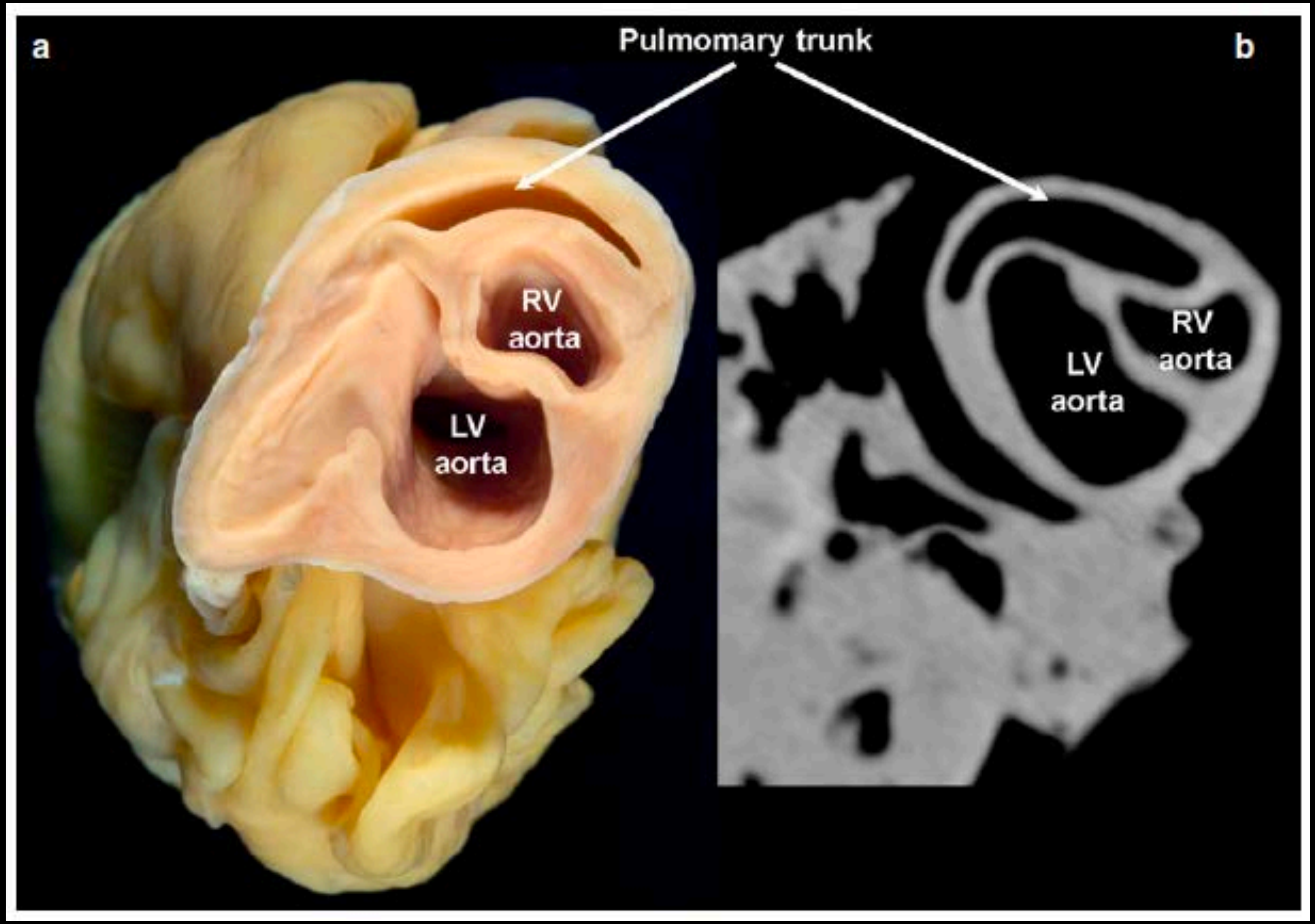
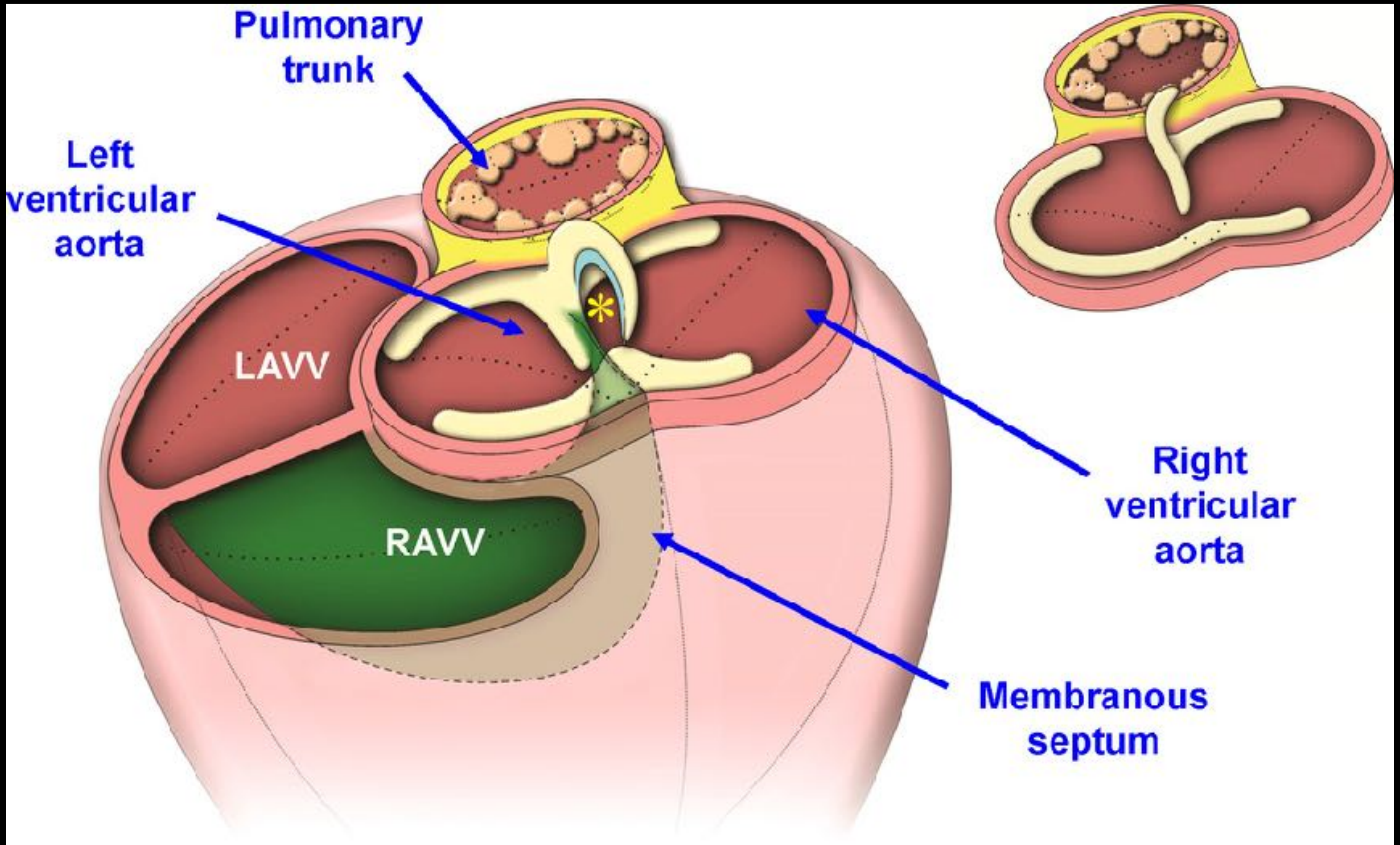
Necker ENFANTS MALADES HÔPITAL UNIVERSITAIRE
FONDATION imagine INSTITUT DES MALADIES GÉNÉTIQUES
UNIVERSITÉ PARIS DESCARTES
Université de Paris
Inserm
Institut national de la santé et de la recherche médicale
cardiogen
Filière nationale de santé maladies cardiaques héréditaires
Centre de Référence des Maladies Cardiaques Héréditaires
European Reference Network
for rare or low prevalence complex diseases
Network Respiratory Diseases (ERN-LUNG)
European Reference Network
for rare or low prevalence complex diseases
Network Heart Diseases (ERN GUARD-HEART)

Comparative Anatomy of Vertebrate Hearts

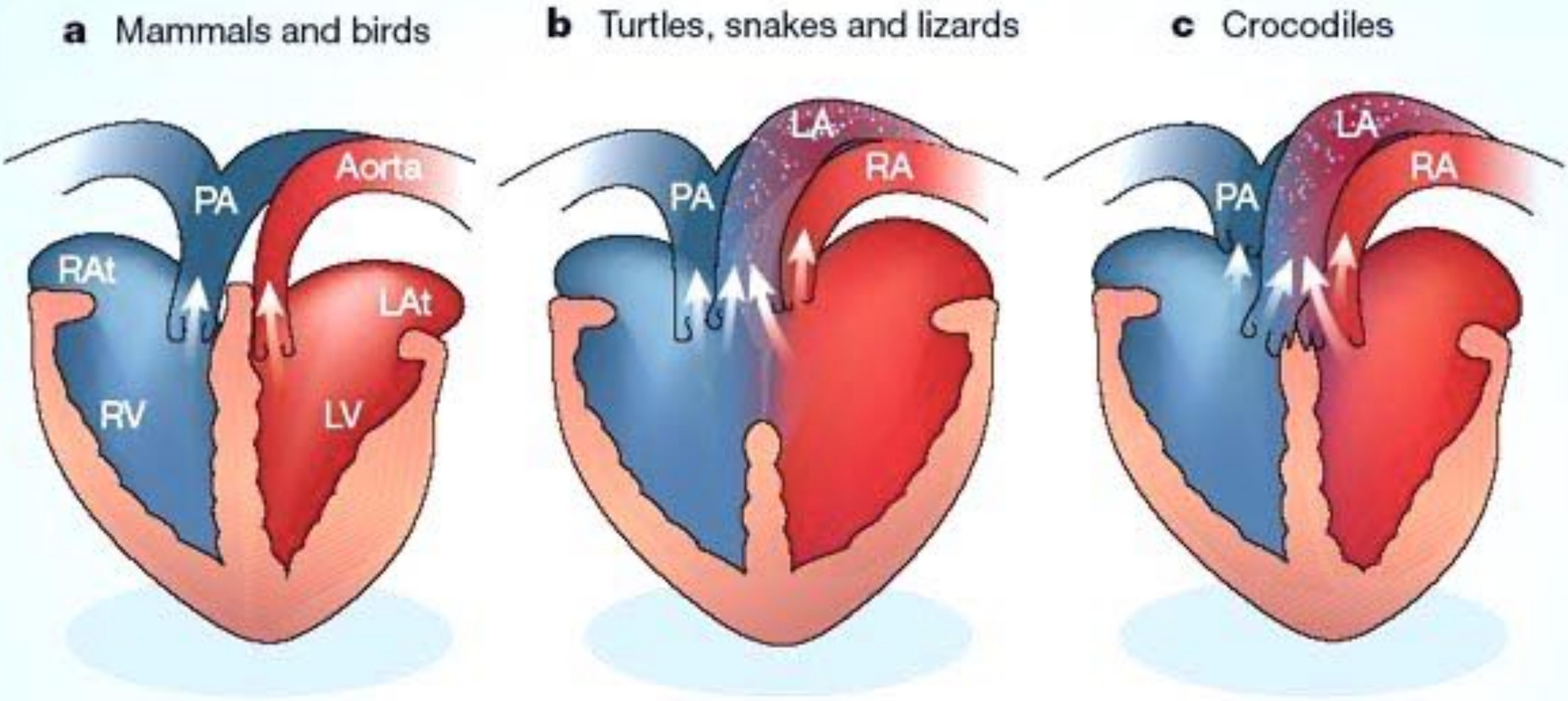




a
Pulmonary trunk
b
RV
aorta
LV
aorta
LV
aorta
RV
aorta

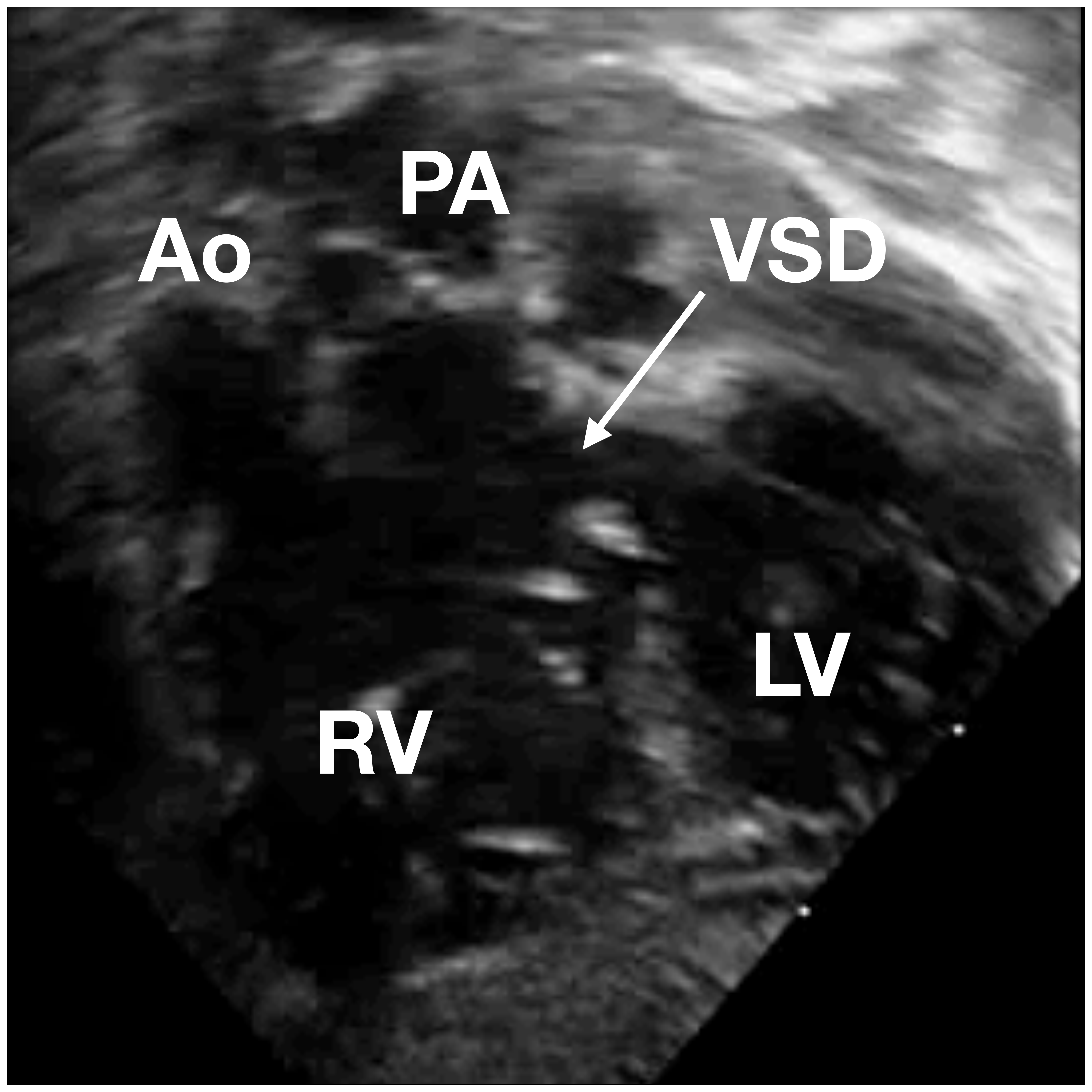


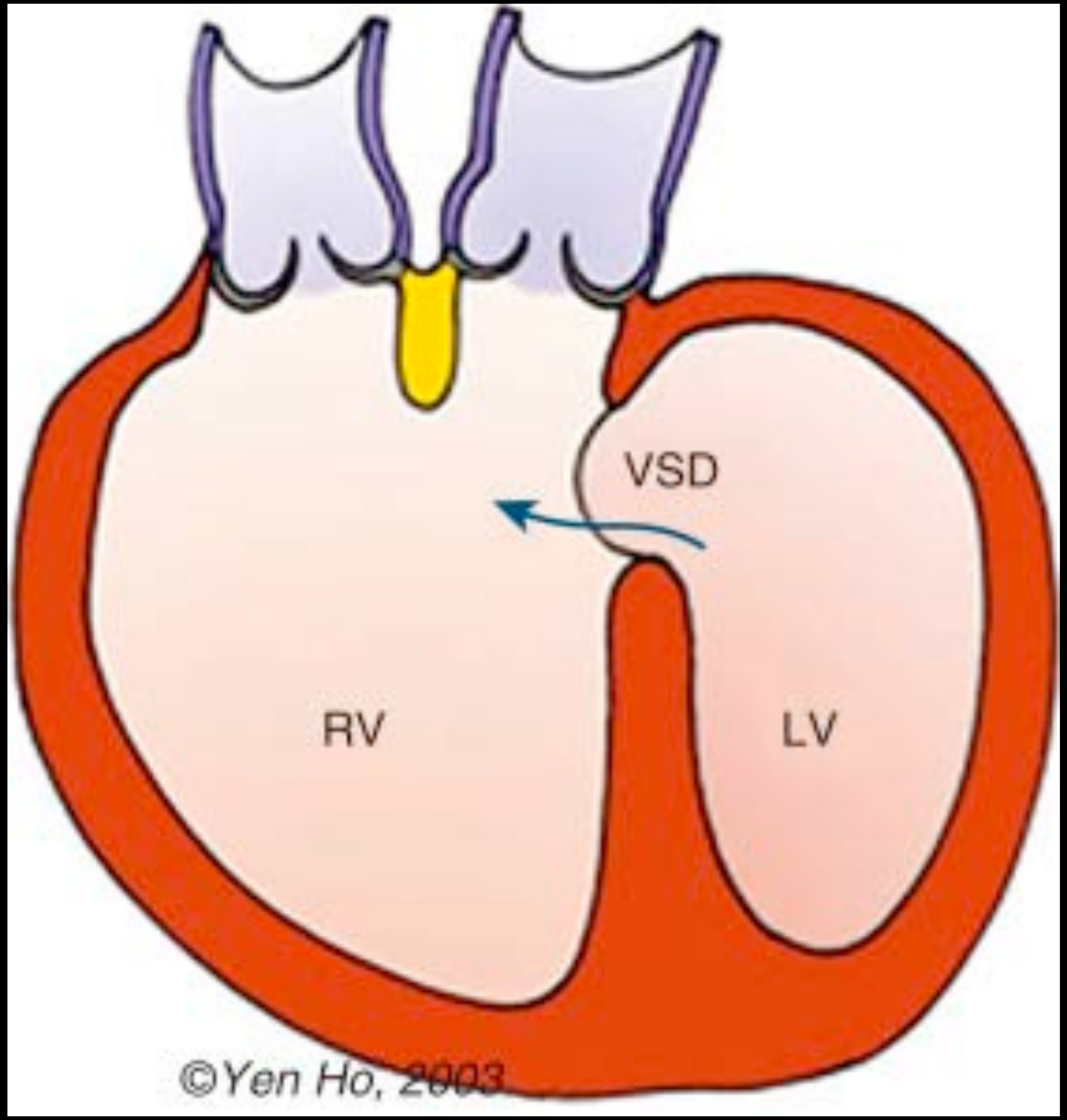
Pulmonary
trunk
Left
ventricular
aorta
LAVV
RAVV
*
Right
ventricular
aorta
Membranous
septum



a Mammals and birds
b Turtles, snakes and lizards
c Crocodiles
PA
Aorta
RAt
LAt
RV
LV
LA
RA
PA
LA
RA
PA

Double outlet right ventricle





Neonatal strategy in DORV

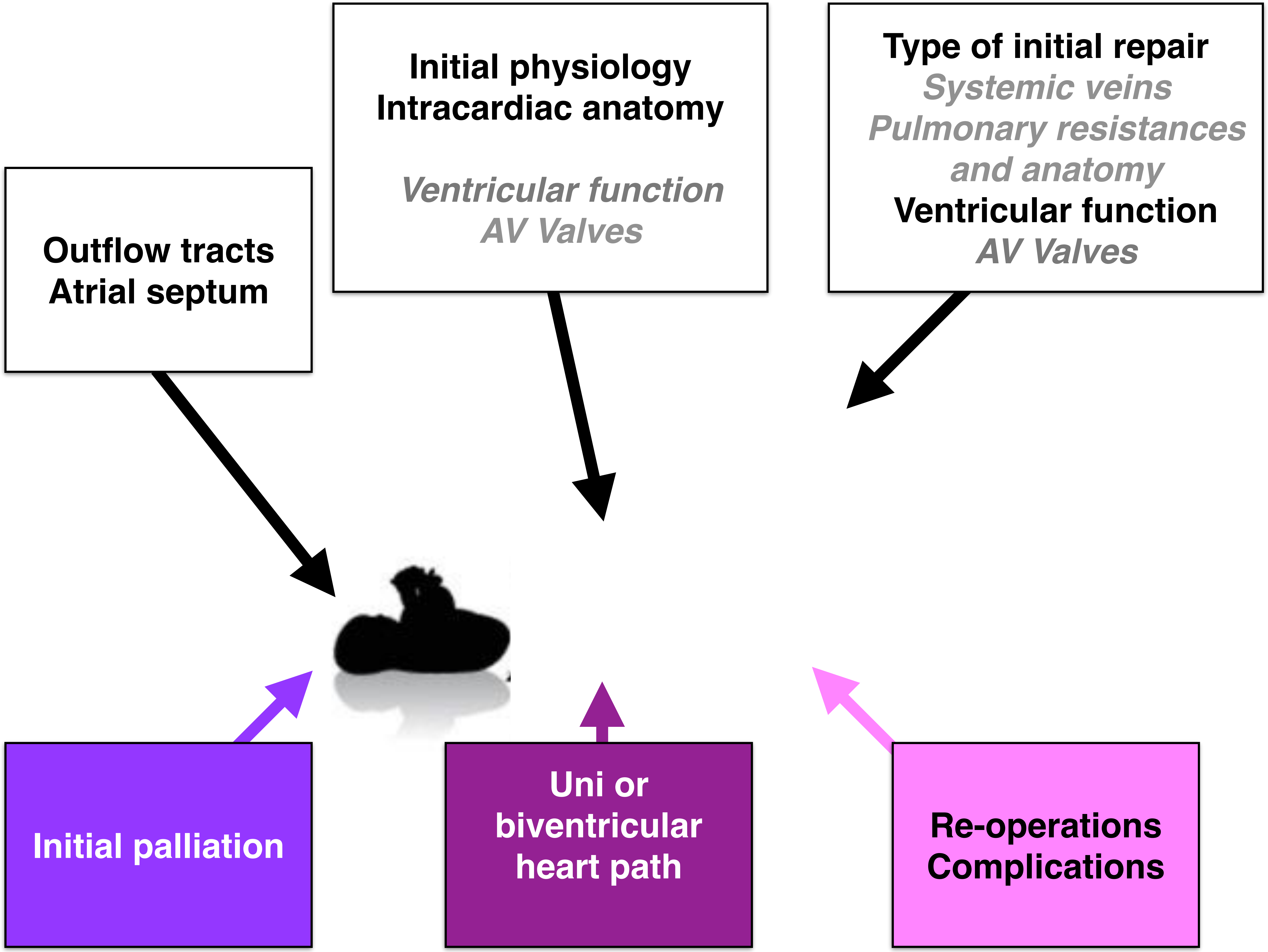
What is the plan during infancy ?

DORV is a **progressive disease** with a potential increase in severity with time.

In situations where biventricular repair is uncertain or hazardous, **setting the ground for optimal univentricular heart palliation** is mandatory.

When biventricular repair is possible, **early choice of the optimal type of repair** is key.

Limiting the risk of re-operations from the start will lead to a better long-term outcome.


Outflow tracts
Atrial septum
Initial physiology
Intracardiac anatomy
Ventricular function
AV Valves
Type of initial repair
Systemic veins
Pulmonary resistances
and anatomy
Ventricular function
AV Valves
Initial palliation
Uni or
biventricular
heart path
Re-operations
Complications

SHF 0 to 1

NCC 0 to 1

Wedging 0 to 1

Convergence 0 to 1

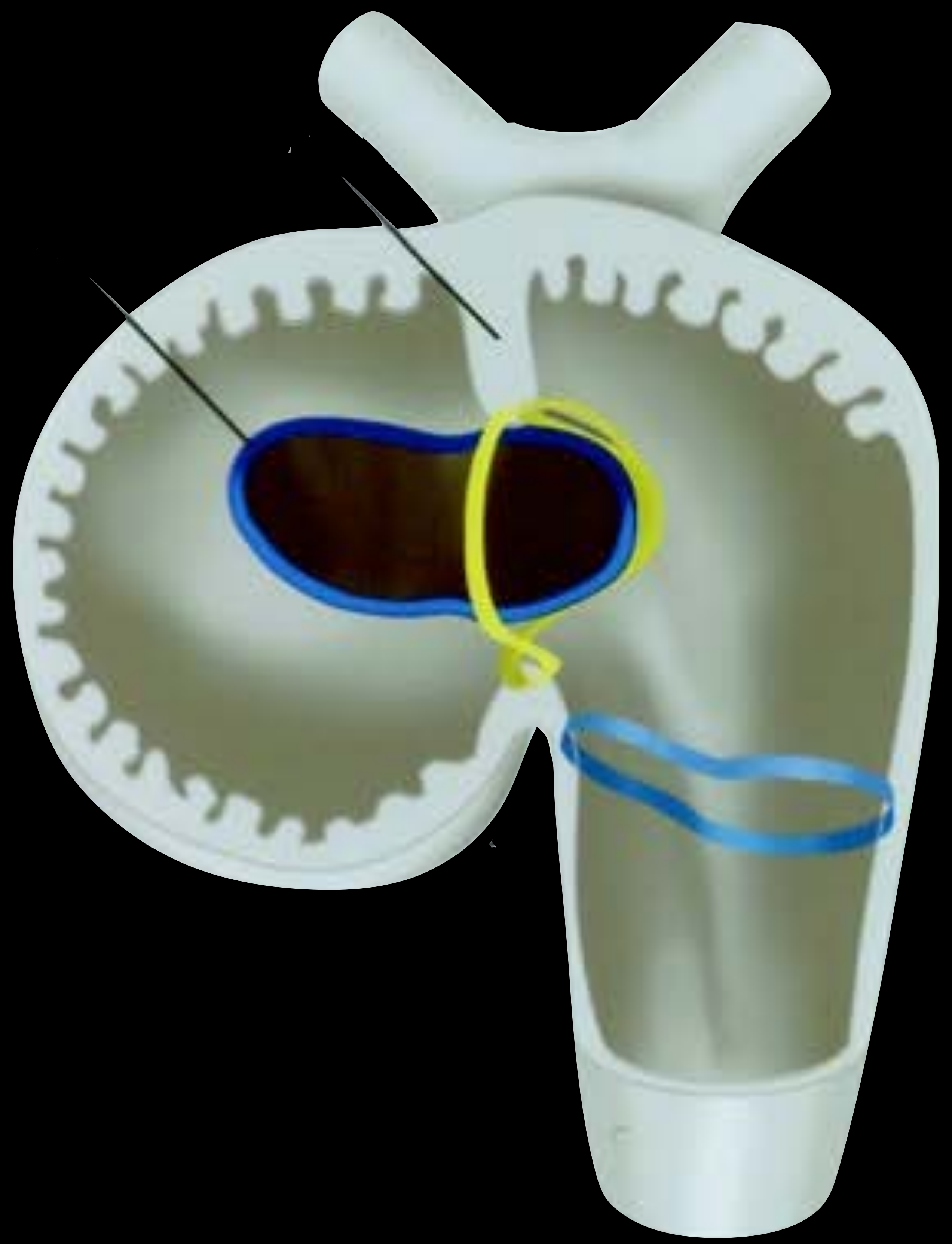
Gittenberger-de Groot A. et al. Basics of Cardiac Development for the Understanding of Congenital Heart Malformations. Pediatr Res 2005;57:169–176

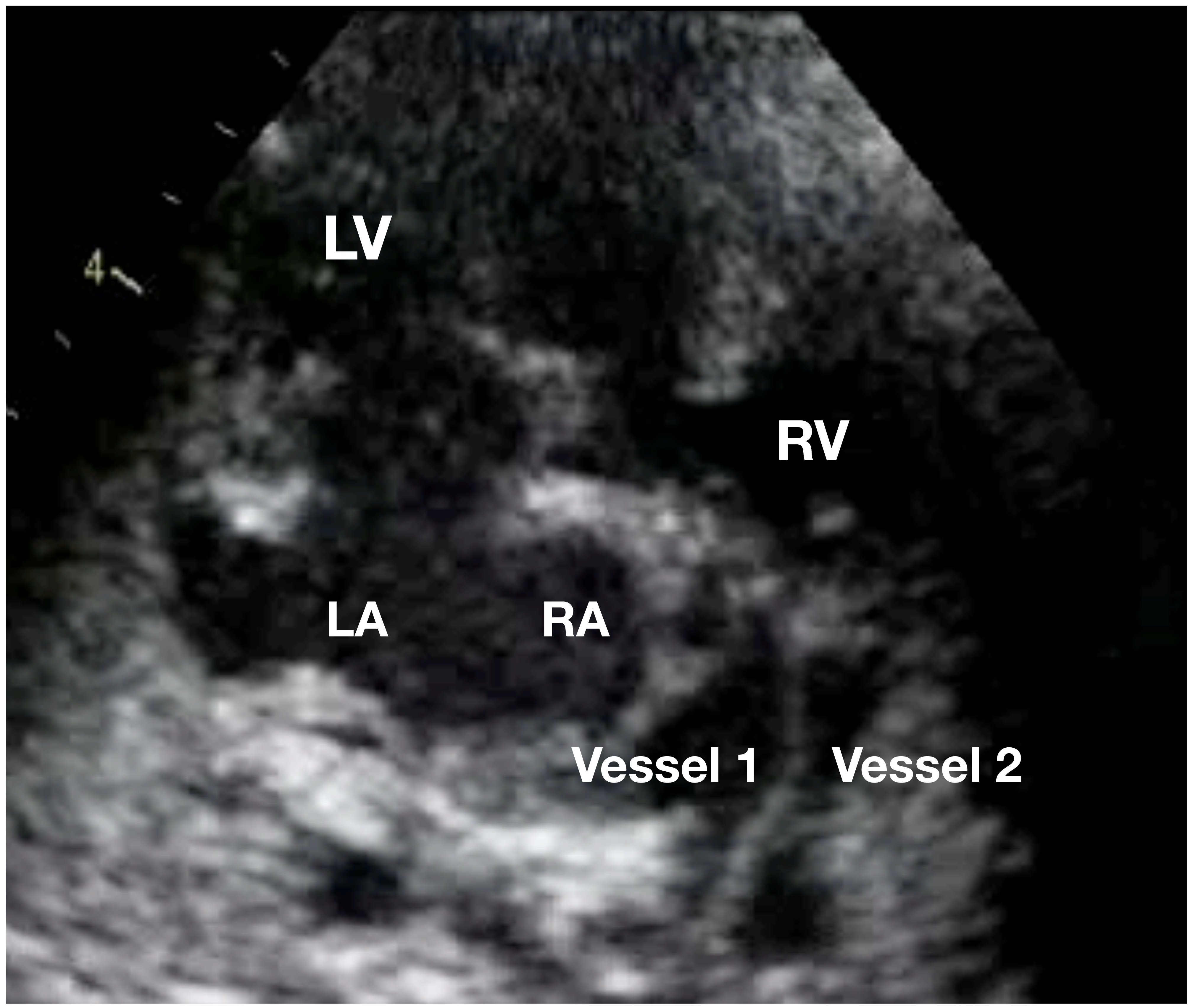
3 groups of DORV (van Praagh)

- 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

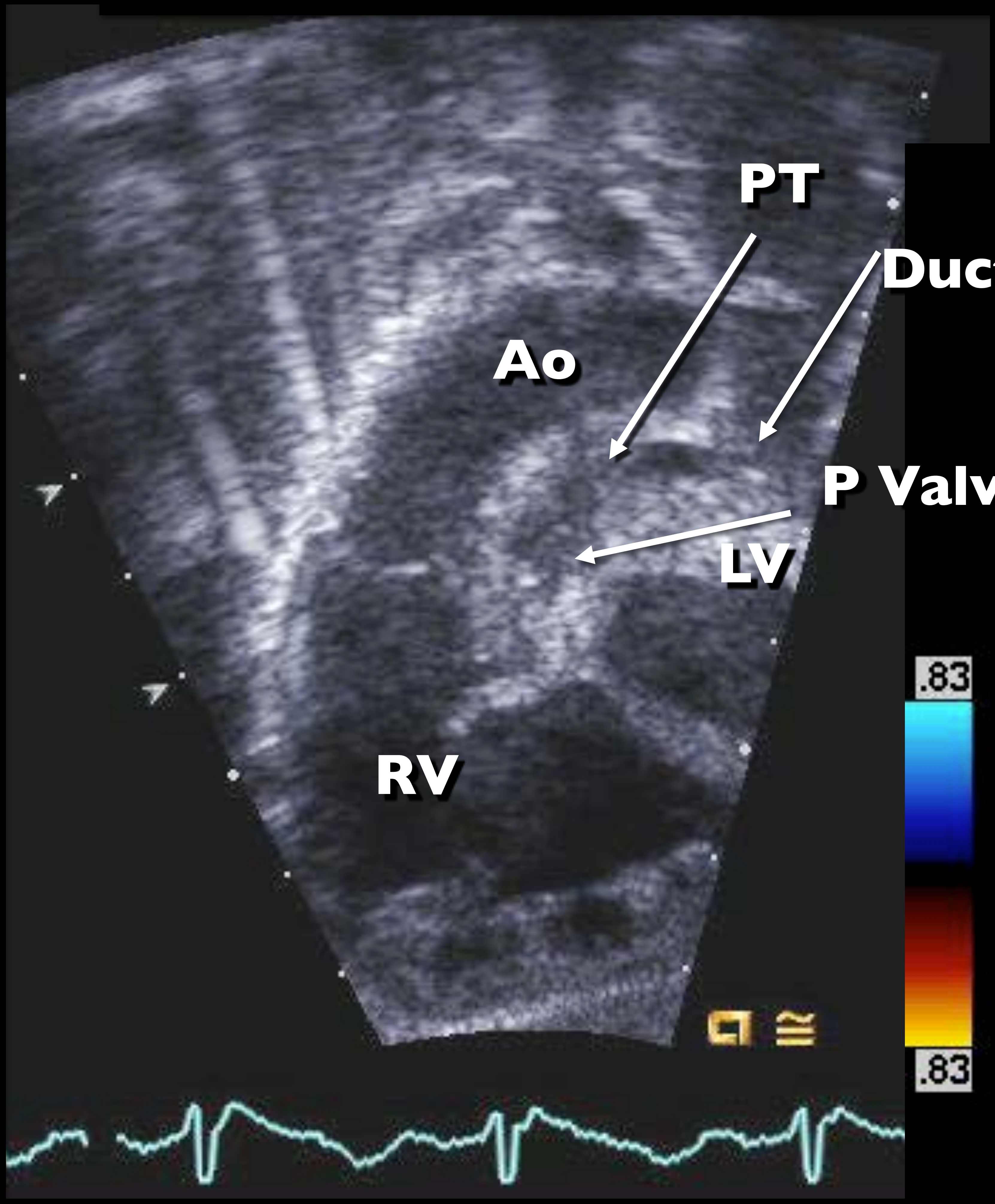


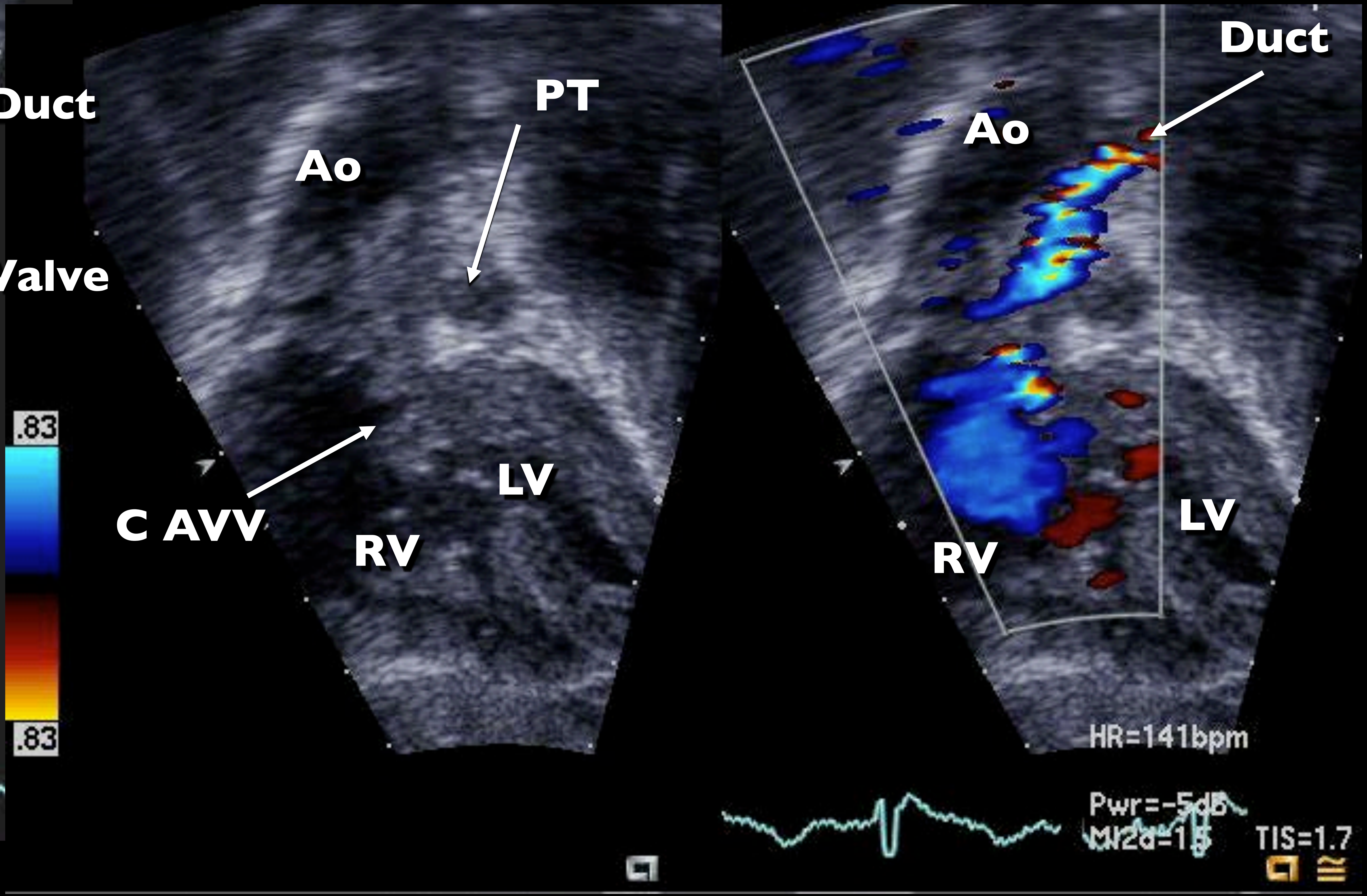


Group 3 DORV

Group 3 DORV

DORV in heterotaxy syndrome
Pulmonary Stenosis/Atresia with DORV





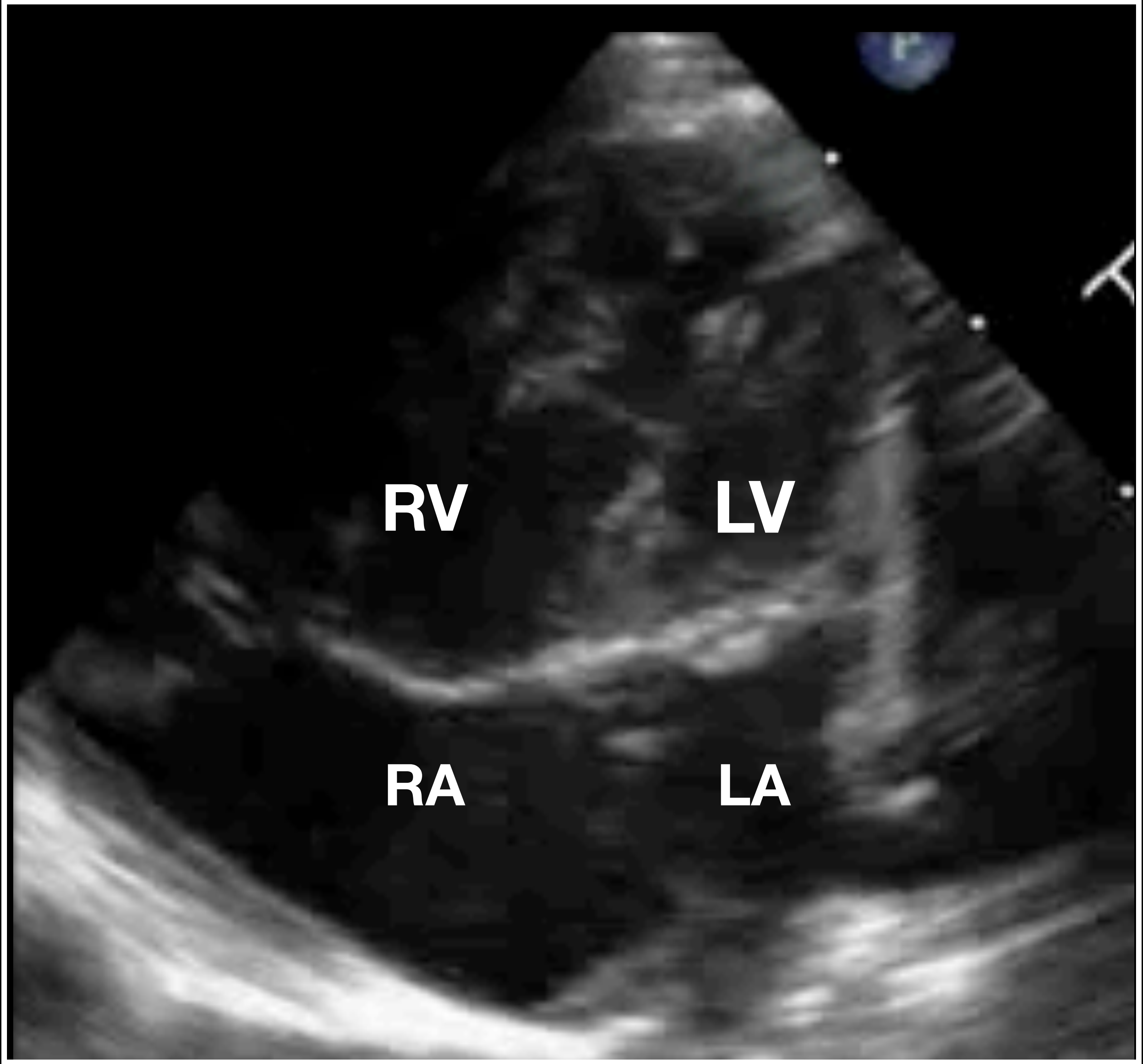
Subcostal Sagittal Cut

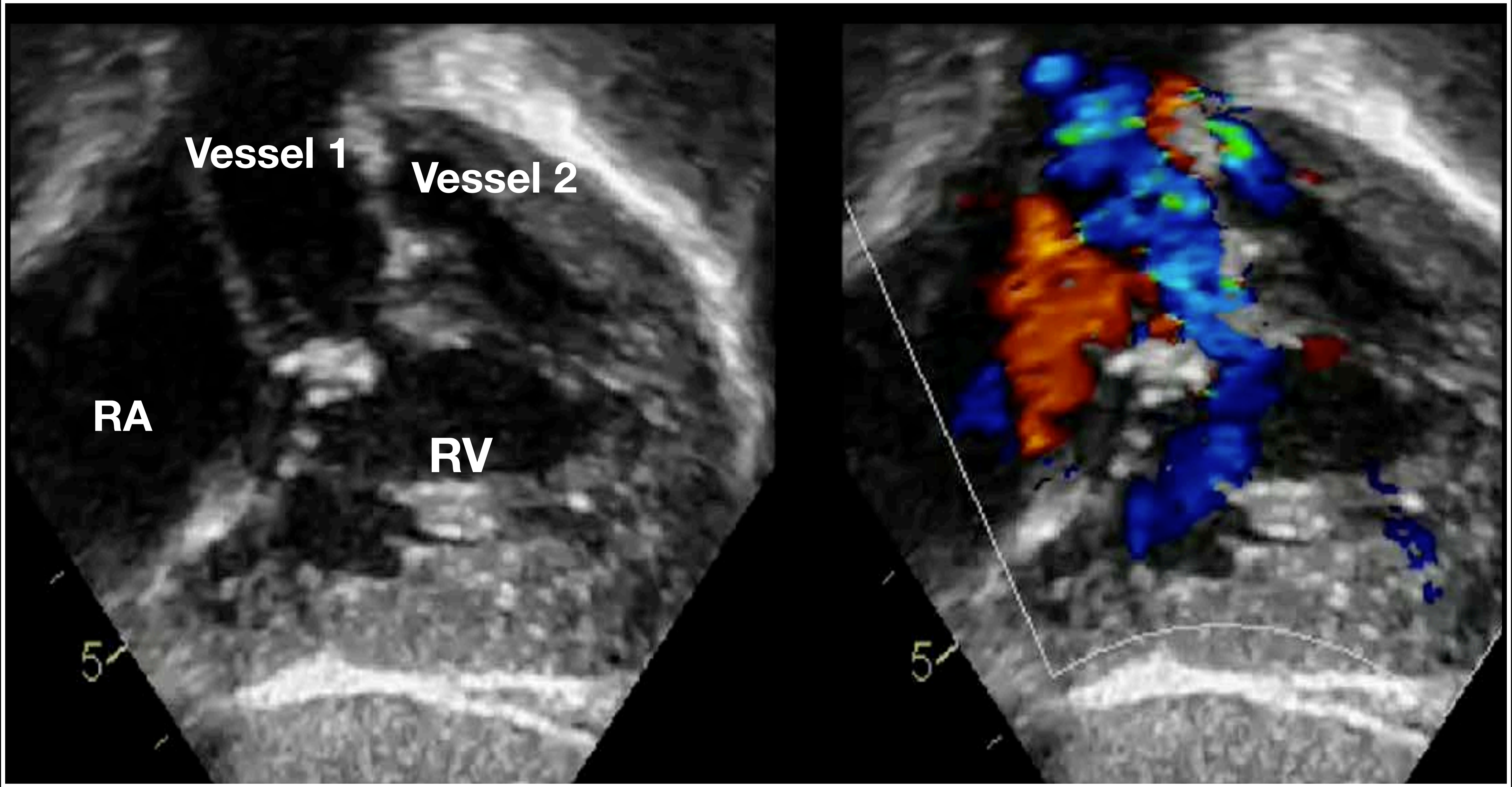
Subcostal Coronal Cuts

DORV

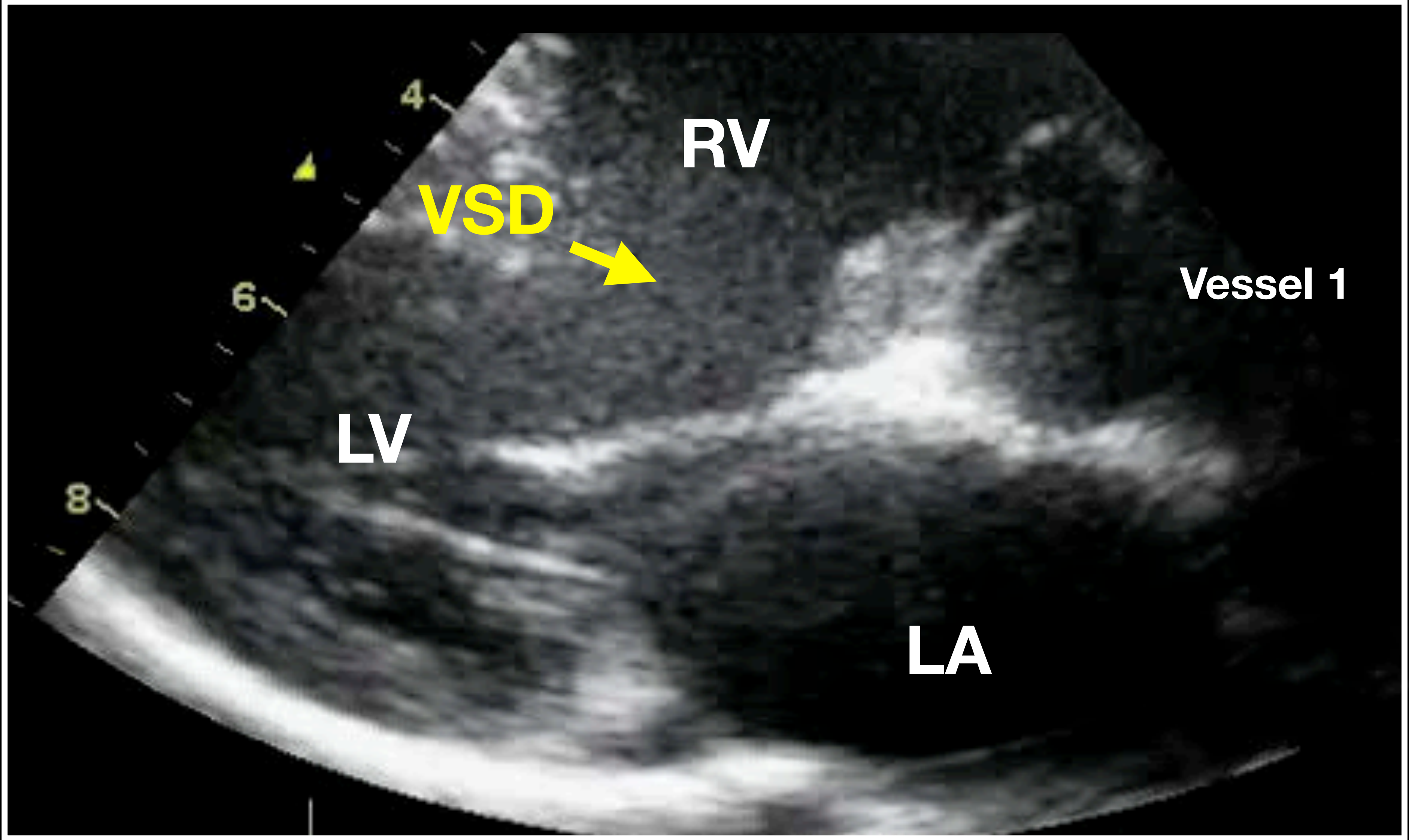
Mitral atresia and DORV

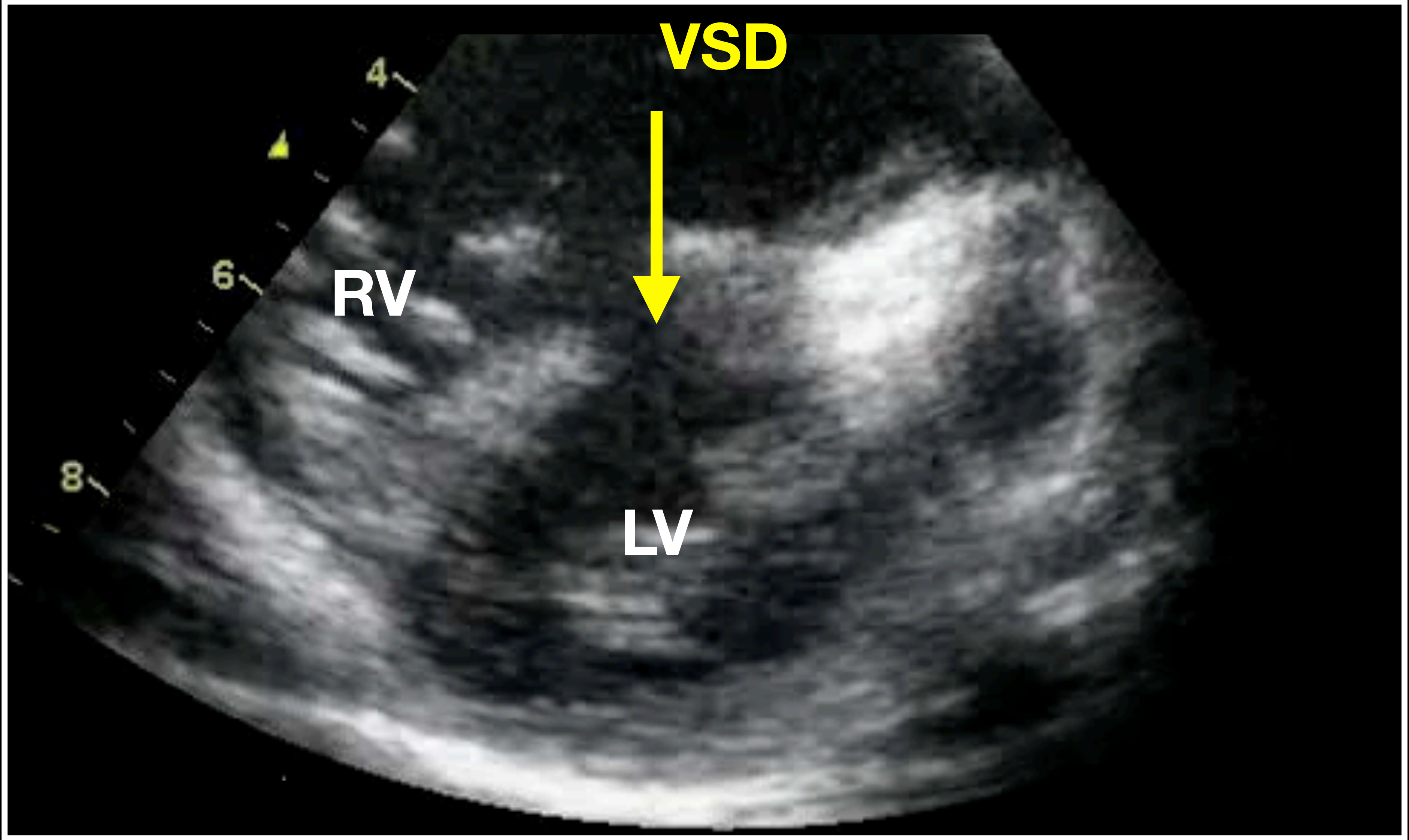
Group 2 DORV

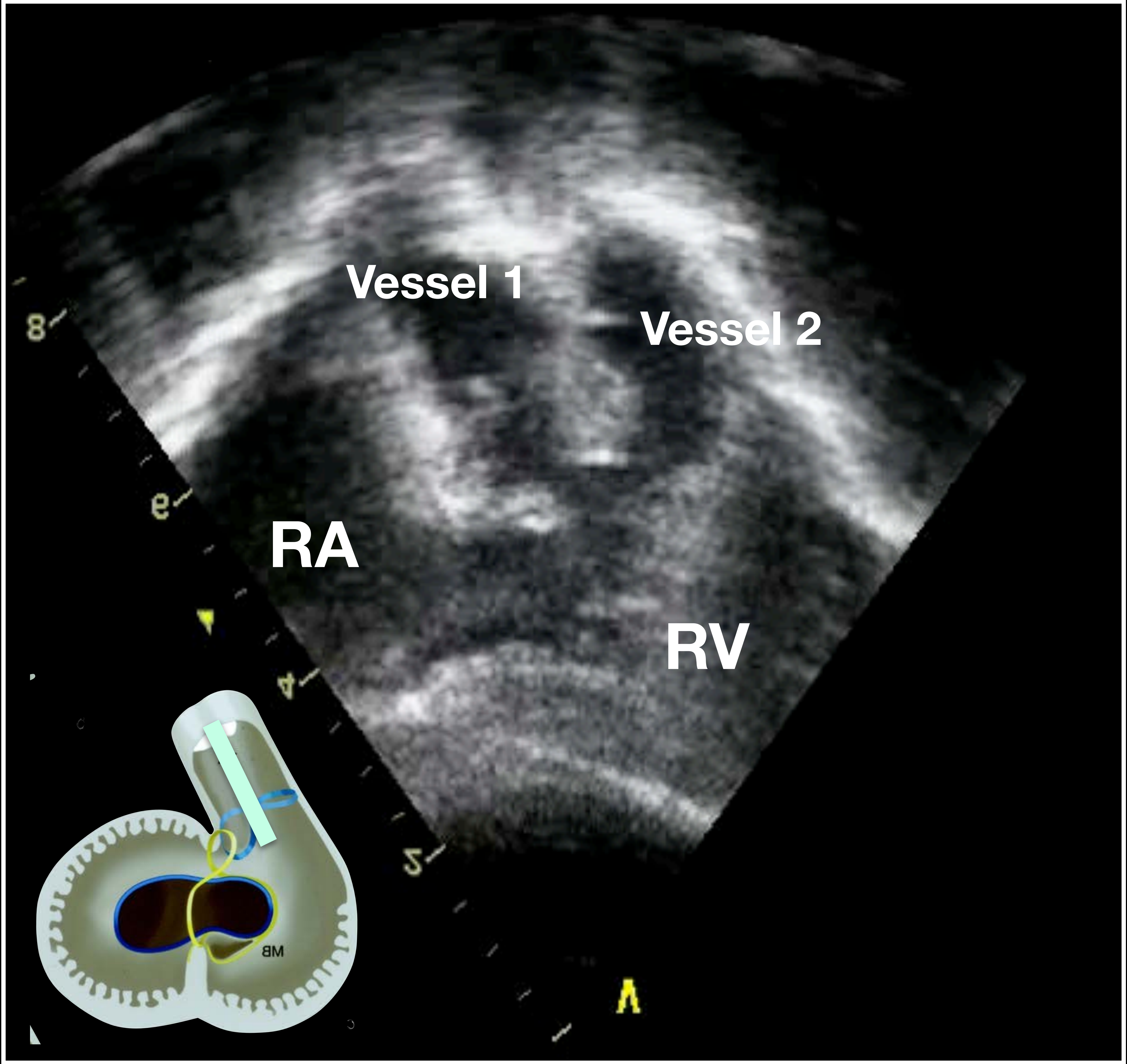




DORV
« Late » DORV

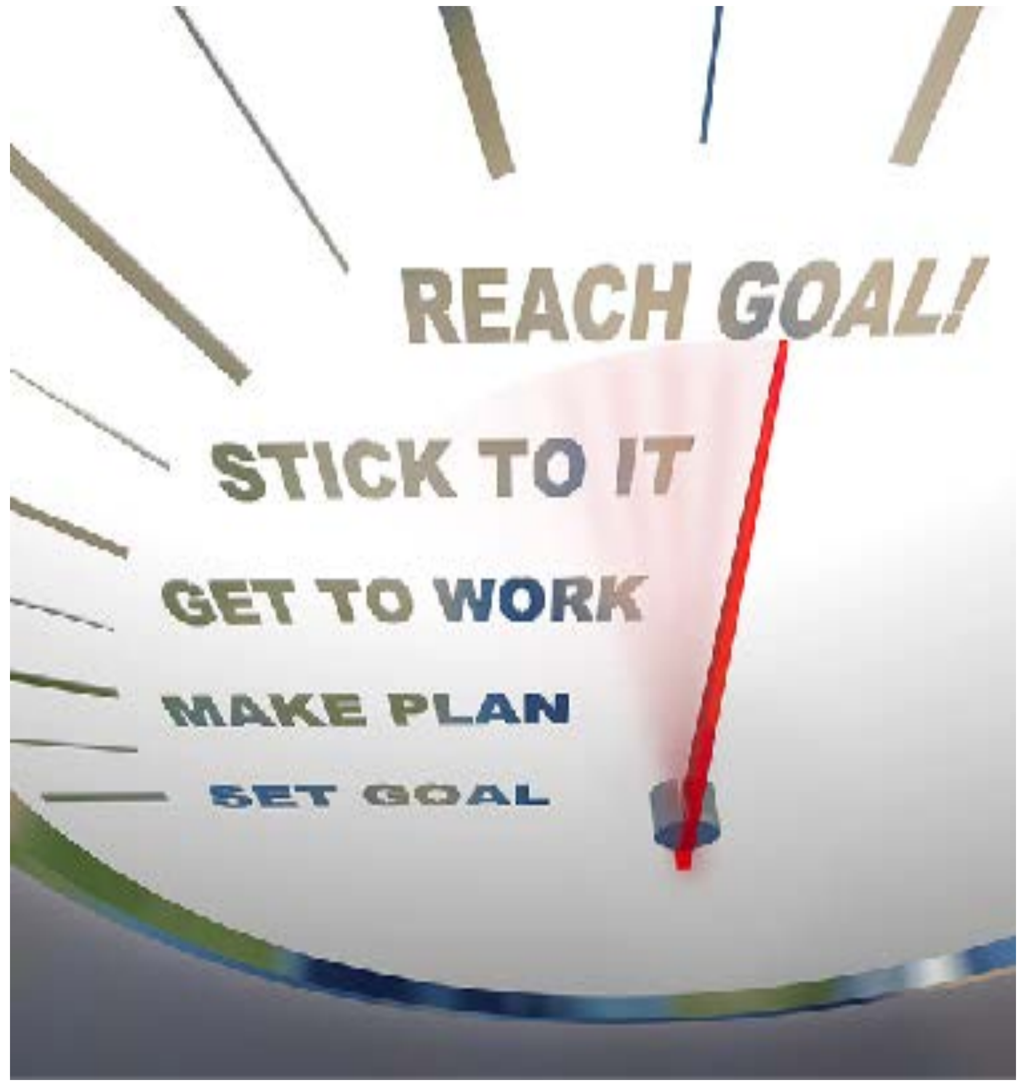






Group 1 DORV

Strategies in DORV type 1



Set goal: normal LV&RV function, no LVOTO/RVOTO, no/mild aortic regurgitation, preserved AV valve function, normal growth of pulmonary artery branches, limited risk of reoperation on RVOT/LVOT

Make plan : elective repair or patient's dependent repair (staged or one step)

Get to work: when ? and how ? and **stick to it**

Reach goal: initial strategy and long-term outcomes

Patients characteristics
Different categories

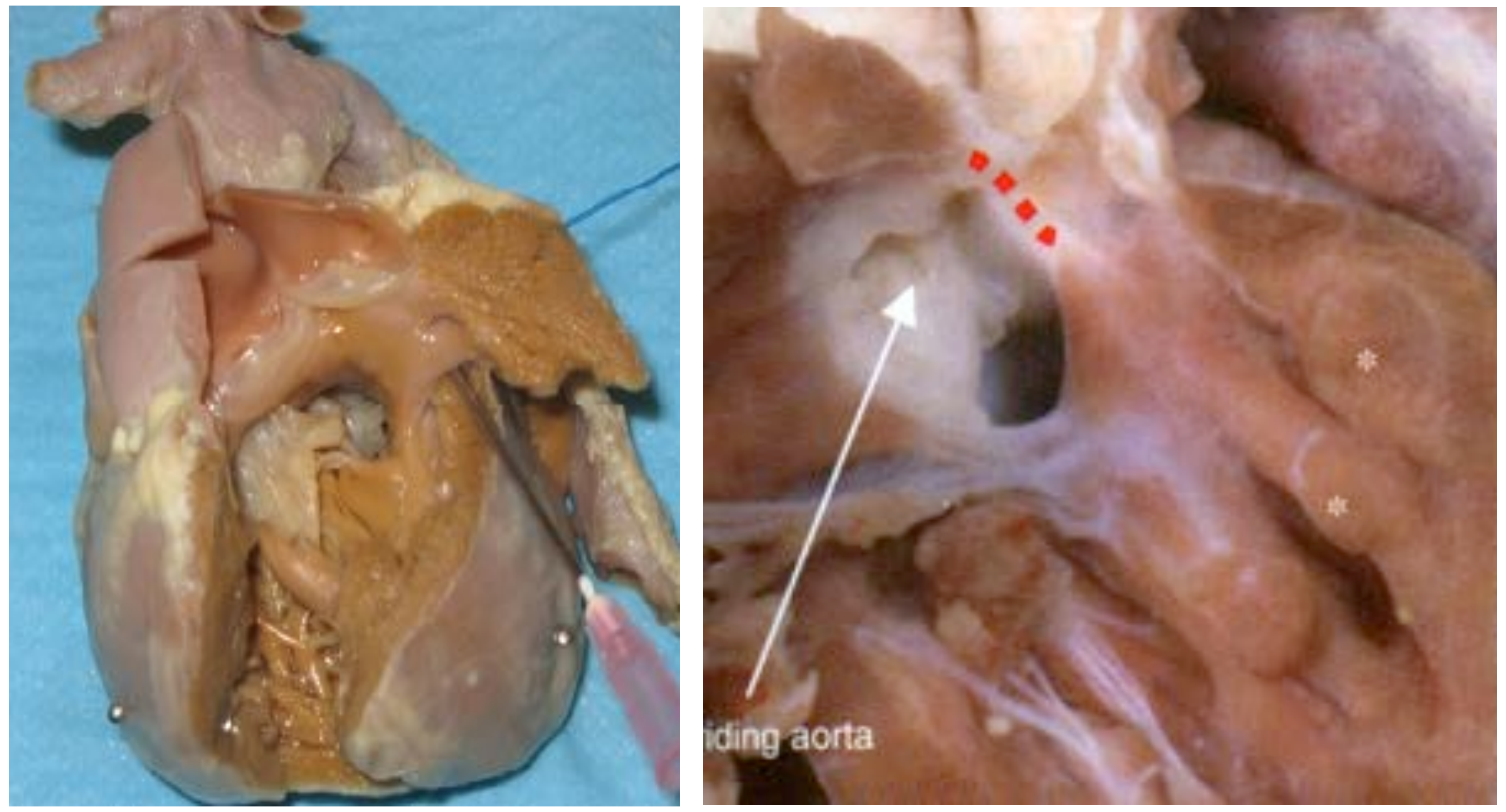
Non modifiable

-underlying genetic conditions

Time-dependent

-age and weight

-symptoms



Anatomical characteristics

Non modifiable

-location of the VSD/physiology of the DORV

-coronary artery anatomy

Time-dependent/modifiable

-anatomy of atrioventricular valves

-pulmonary tree anatomy (valve, annulus, branches)

DORV - classifications

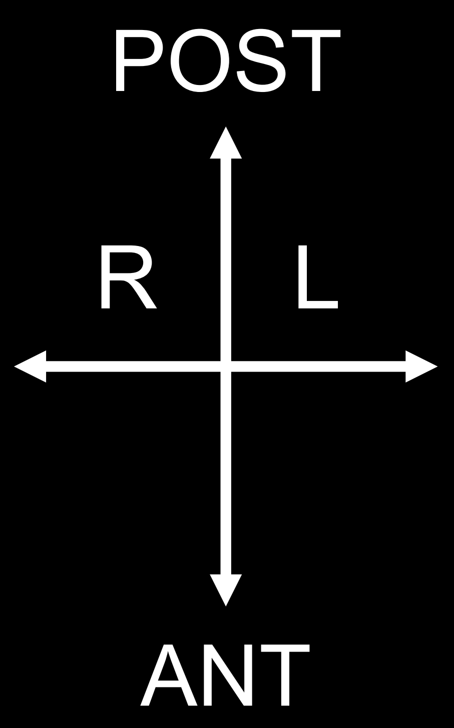
1-Relationship between VSD and great vessels (Lev 1972) : 4 types

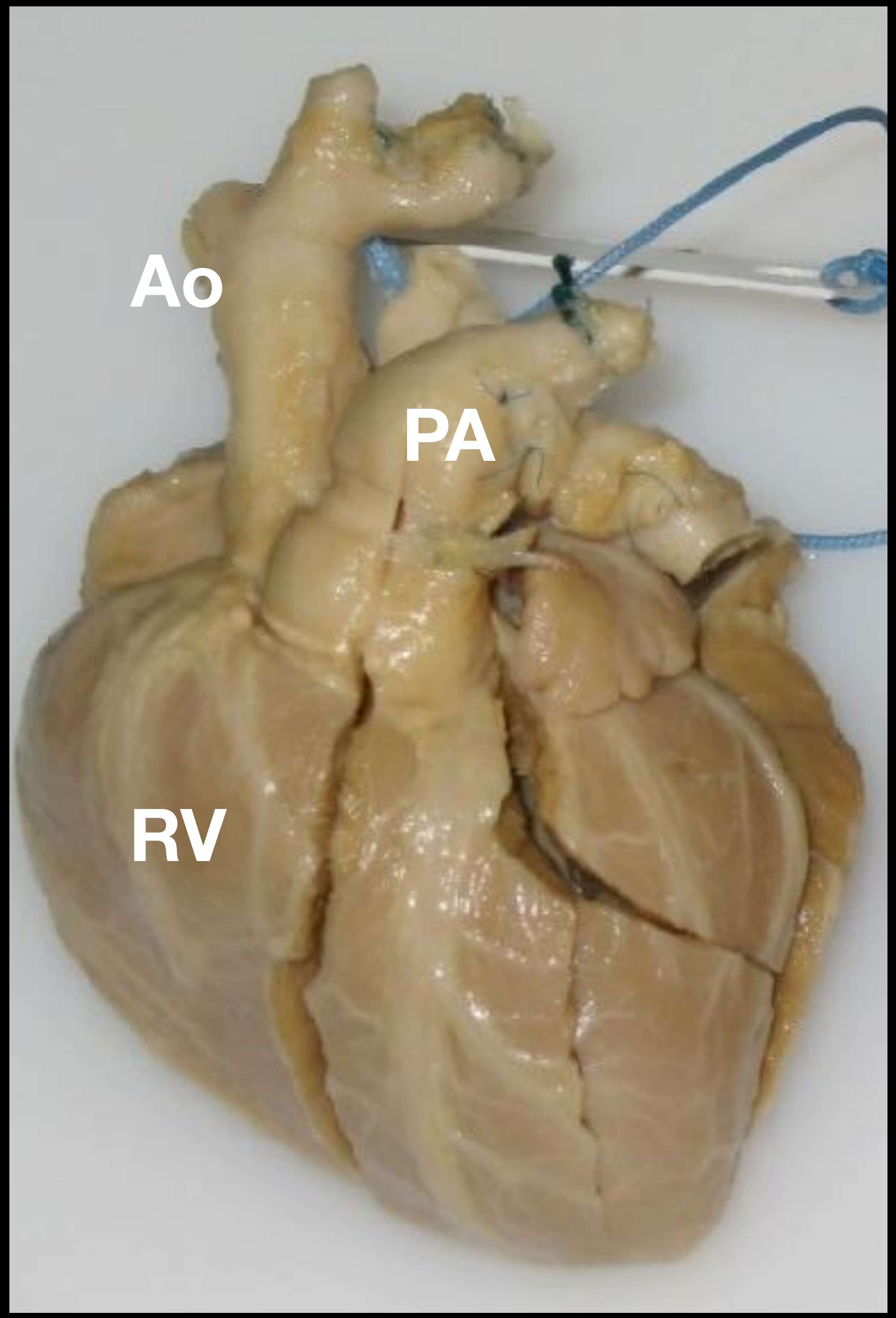
- Sub-aortic
- Sub-pulmonary
- Double committed
- Non committed

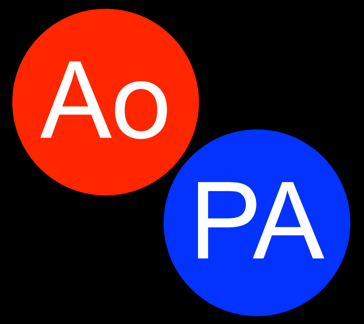
« Physiological classification »

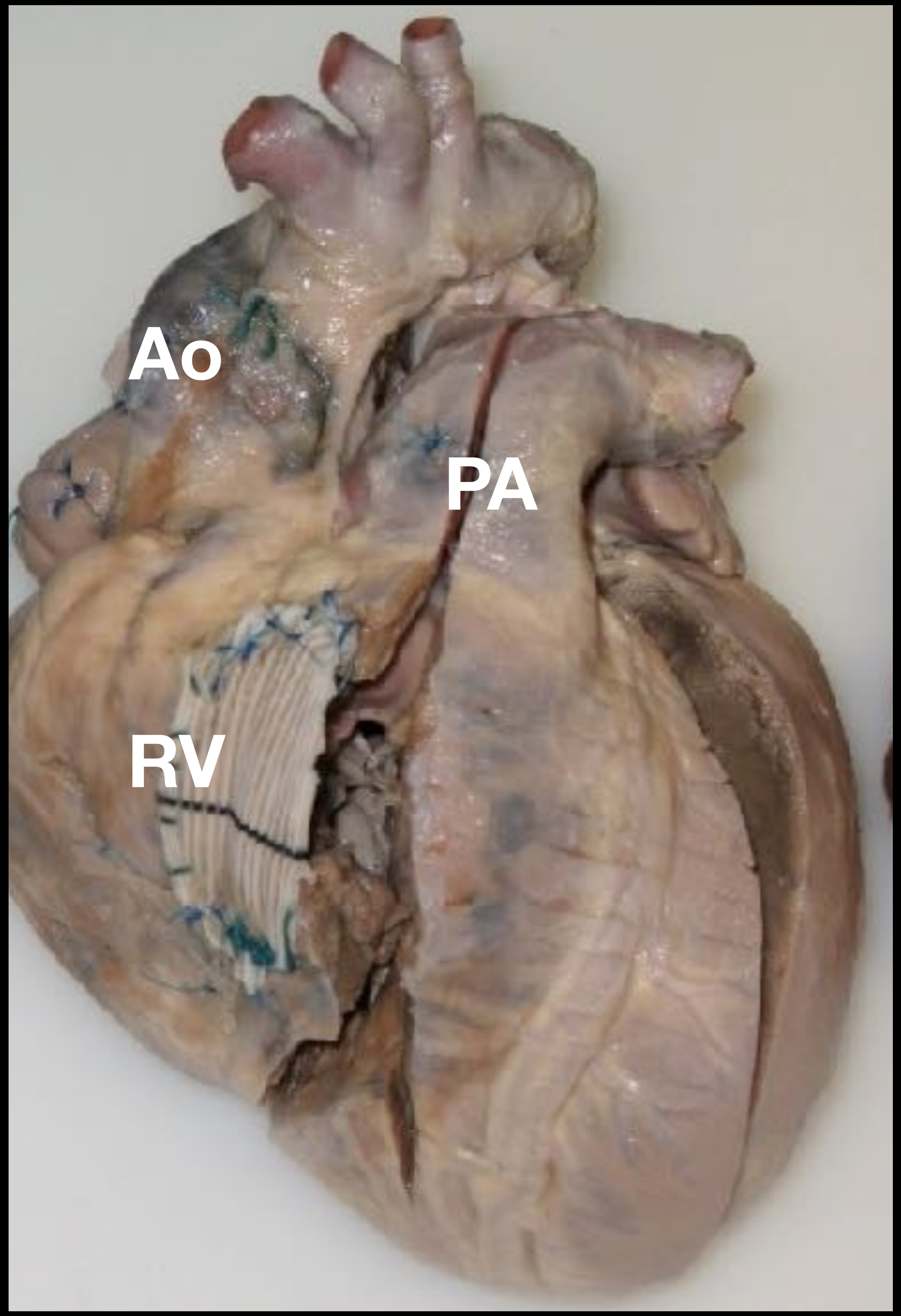
2-Relationship between the two great vessels (De La Cruz 1992)

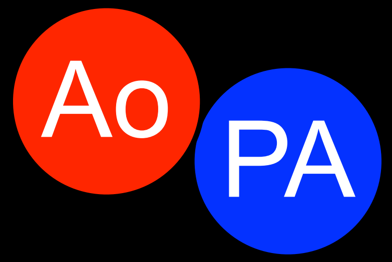
Relative position of the great vessels in DORV

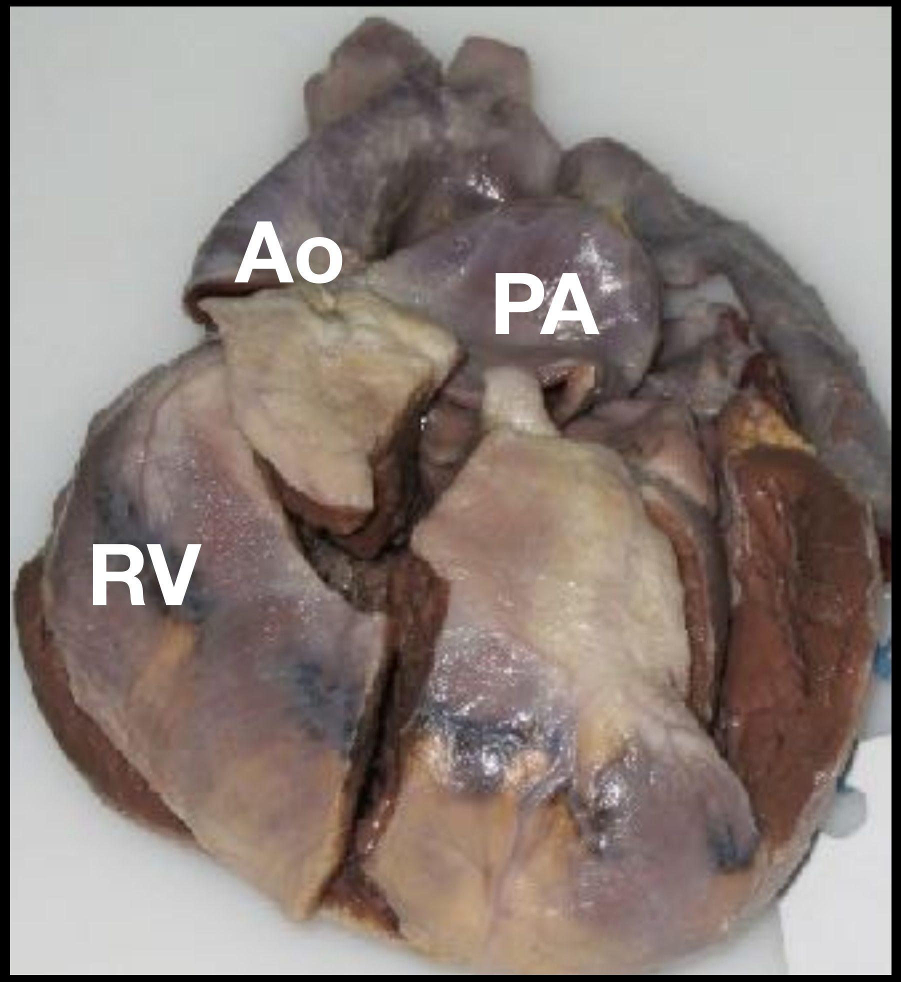


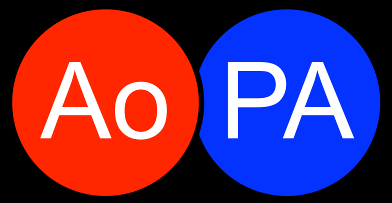


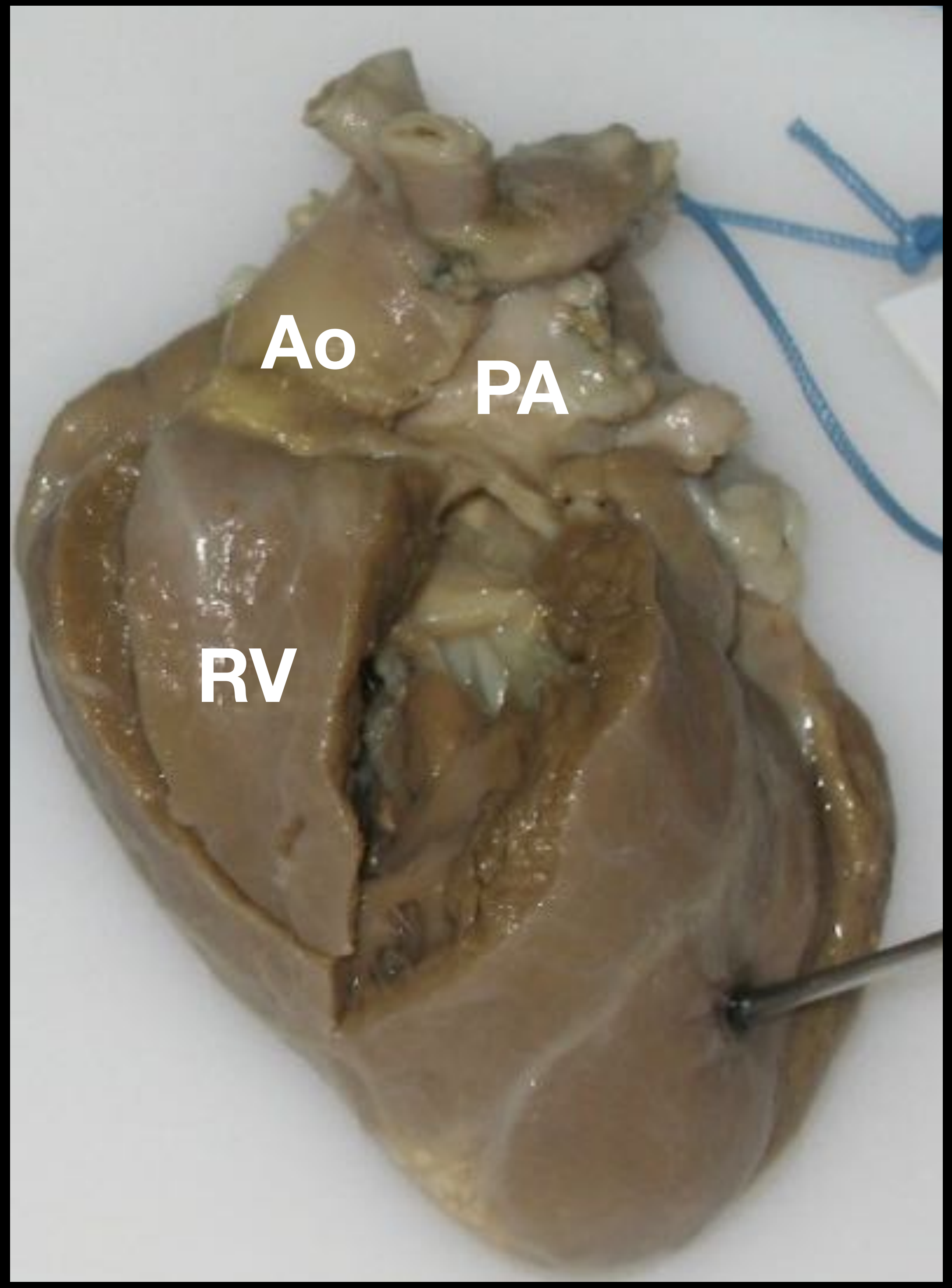


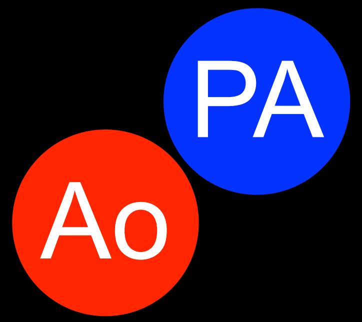


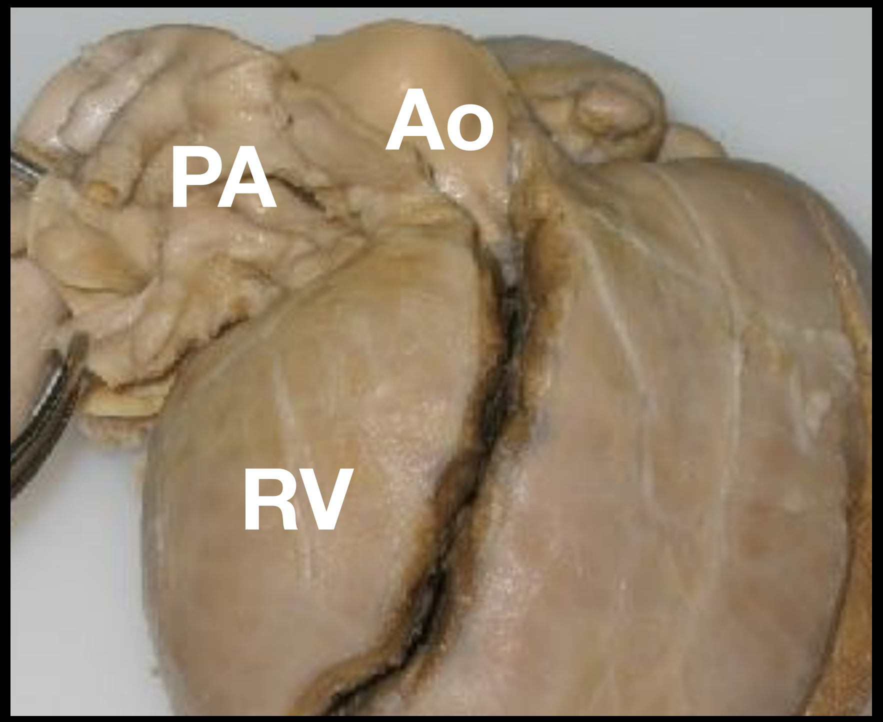




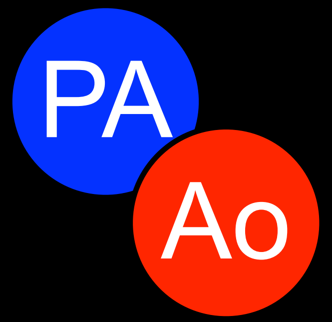




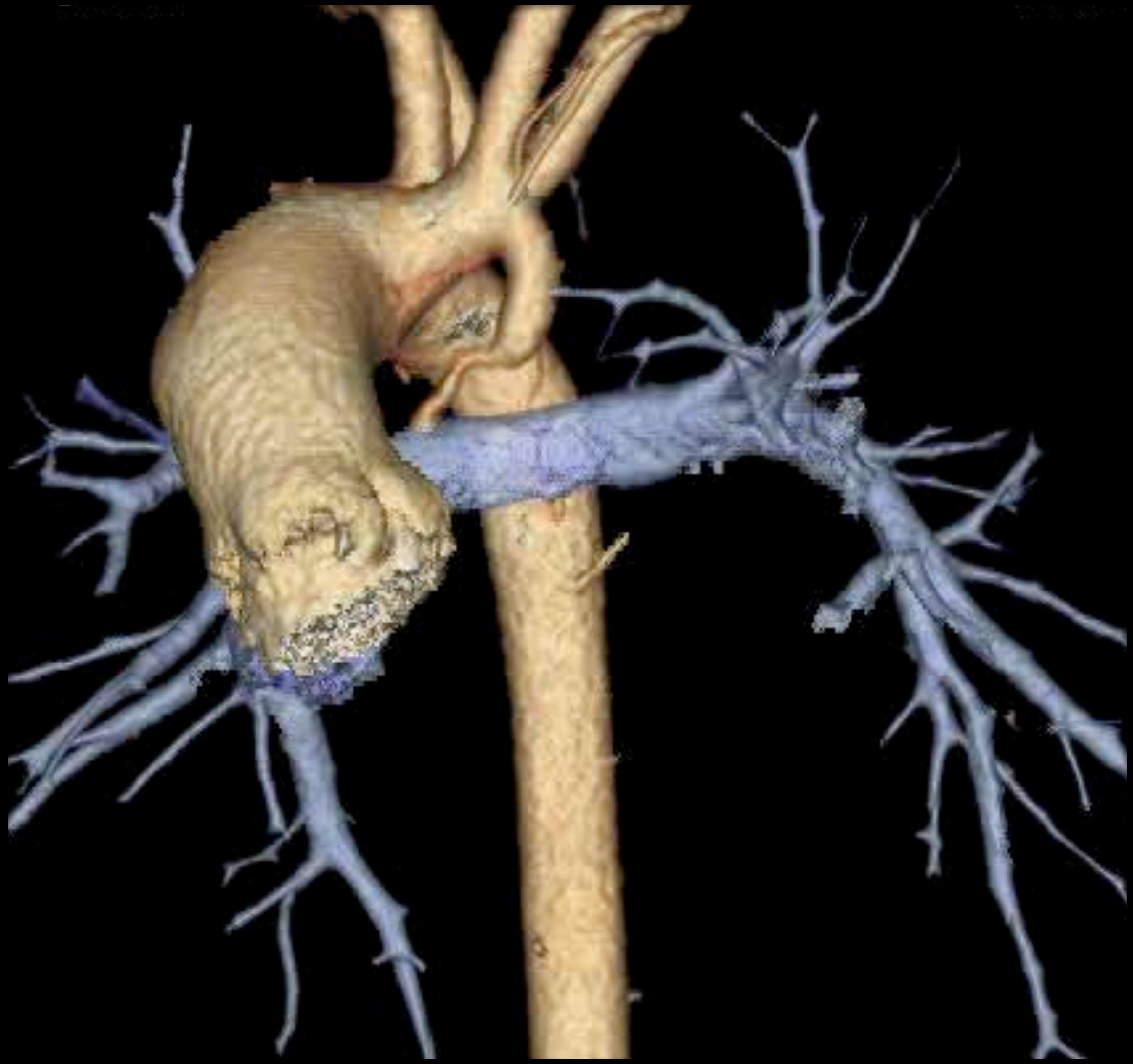




S,D,L



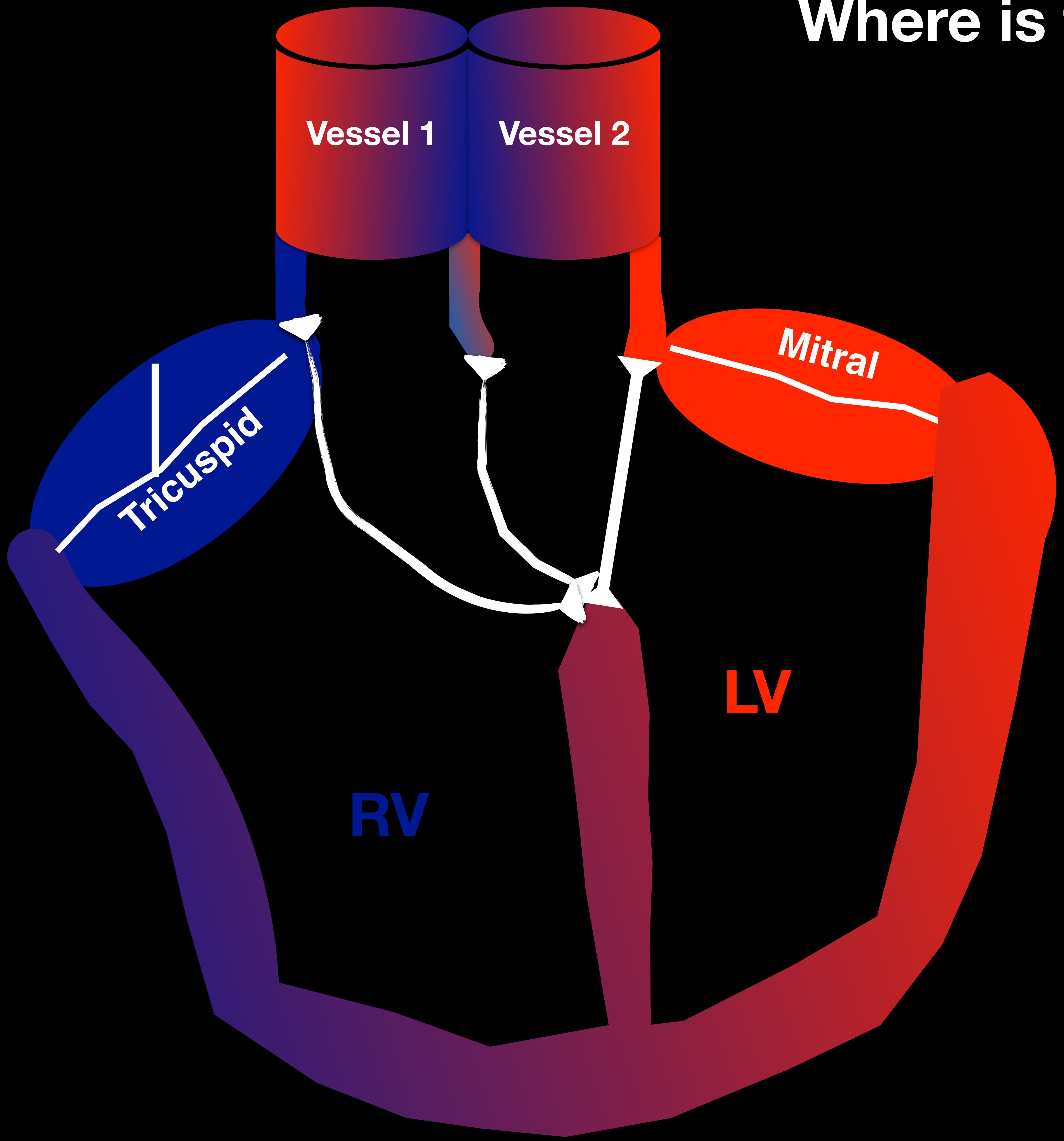
Relative position of the great vessels in DORV



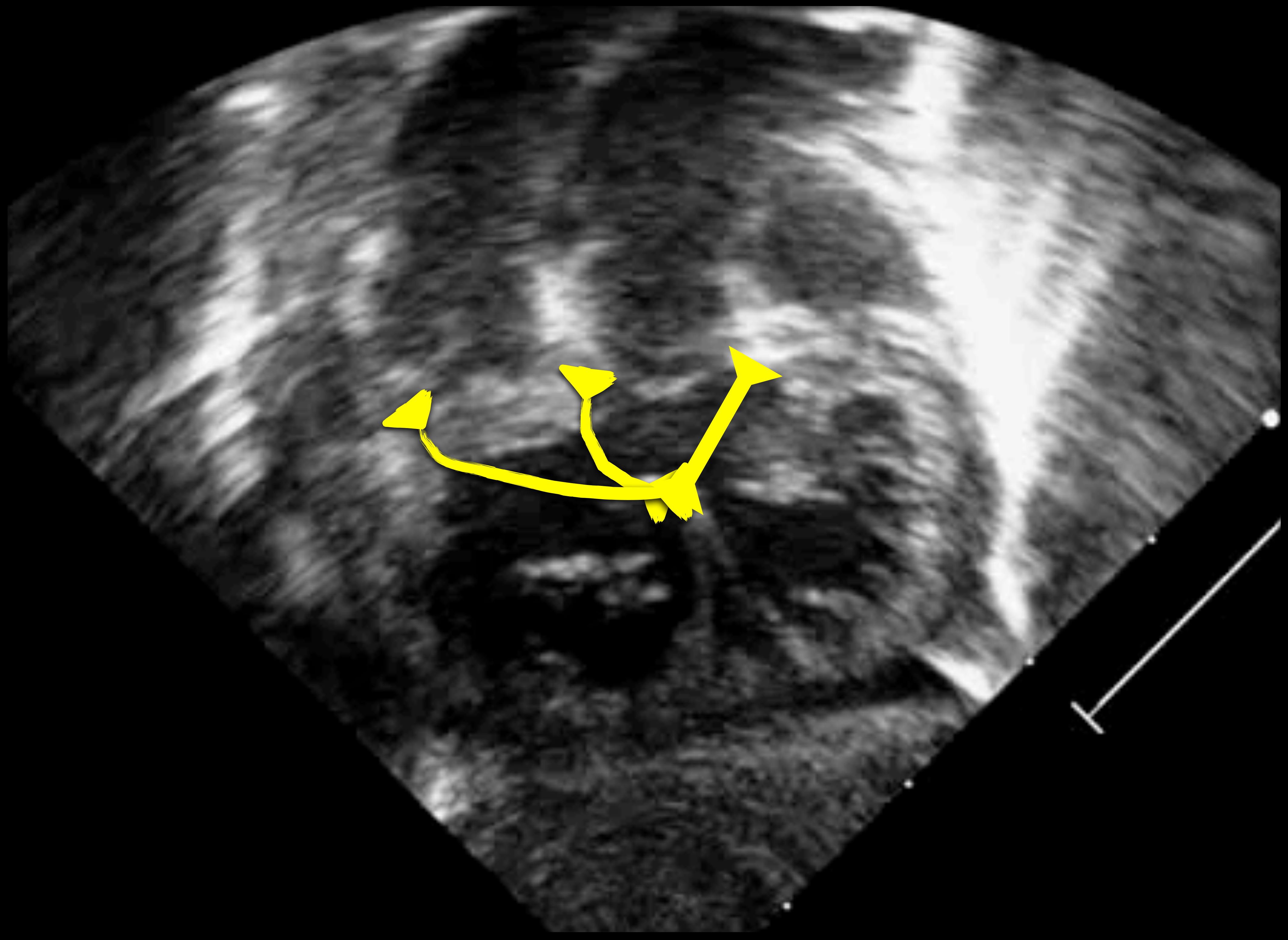
The position of the great vessels does not predict where is the VSD

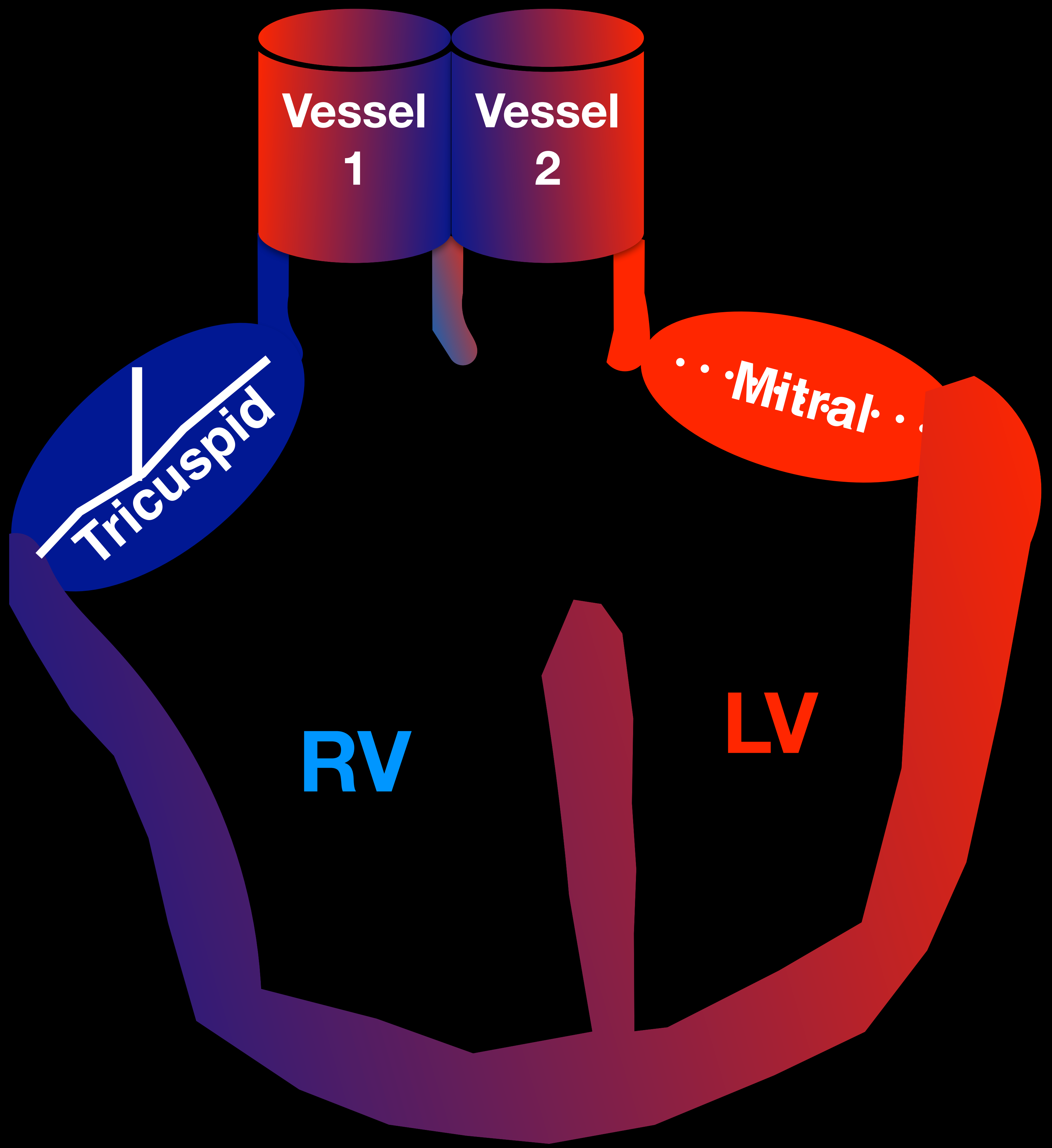
A-The position of the great vessels does not predict where is the VSD but
1-is of importance for surgical planning (ASO, Bex-Nikaido, Conotruncal rotation)
2-May complicate coronary transfer when ASO is indicated (side-by-side)

Where is the « VSD » in DORV ?

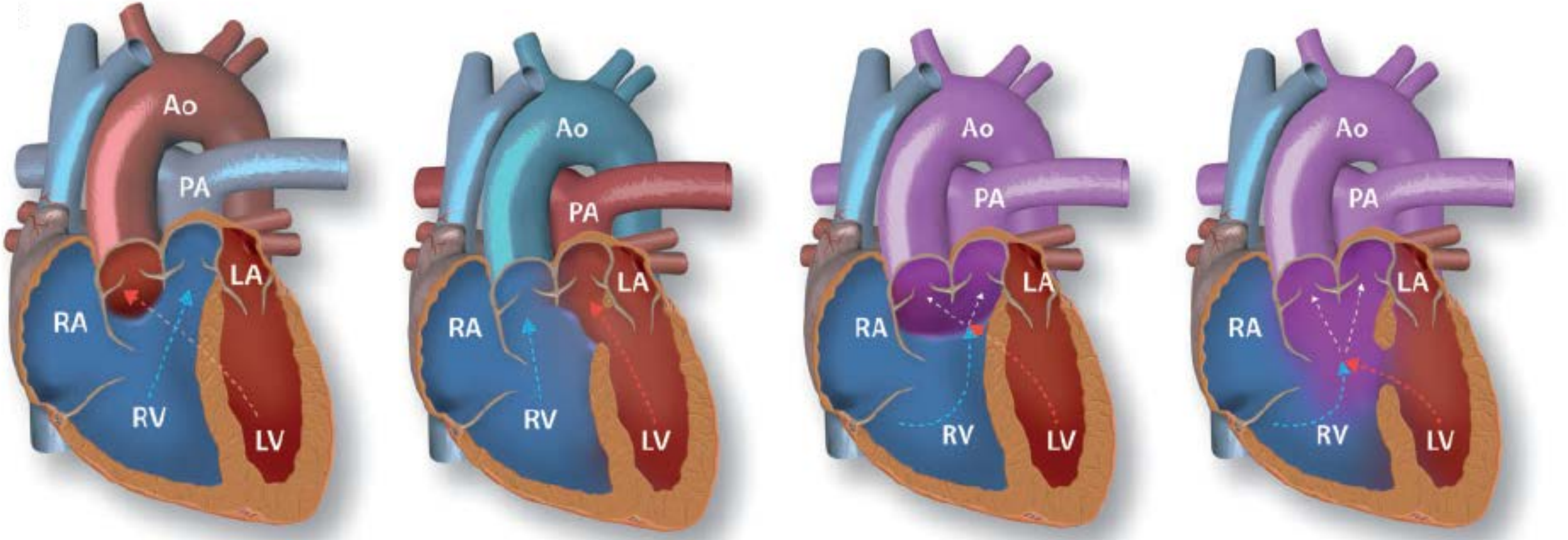


Where is the « VSD » in DORV ?



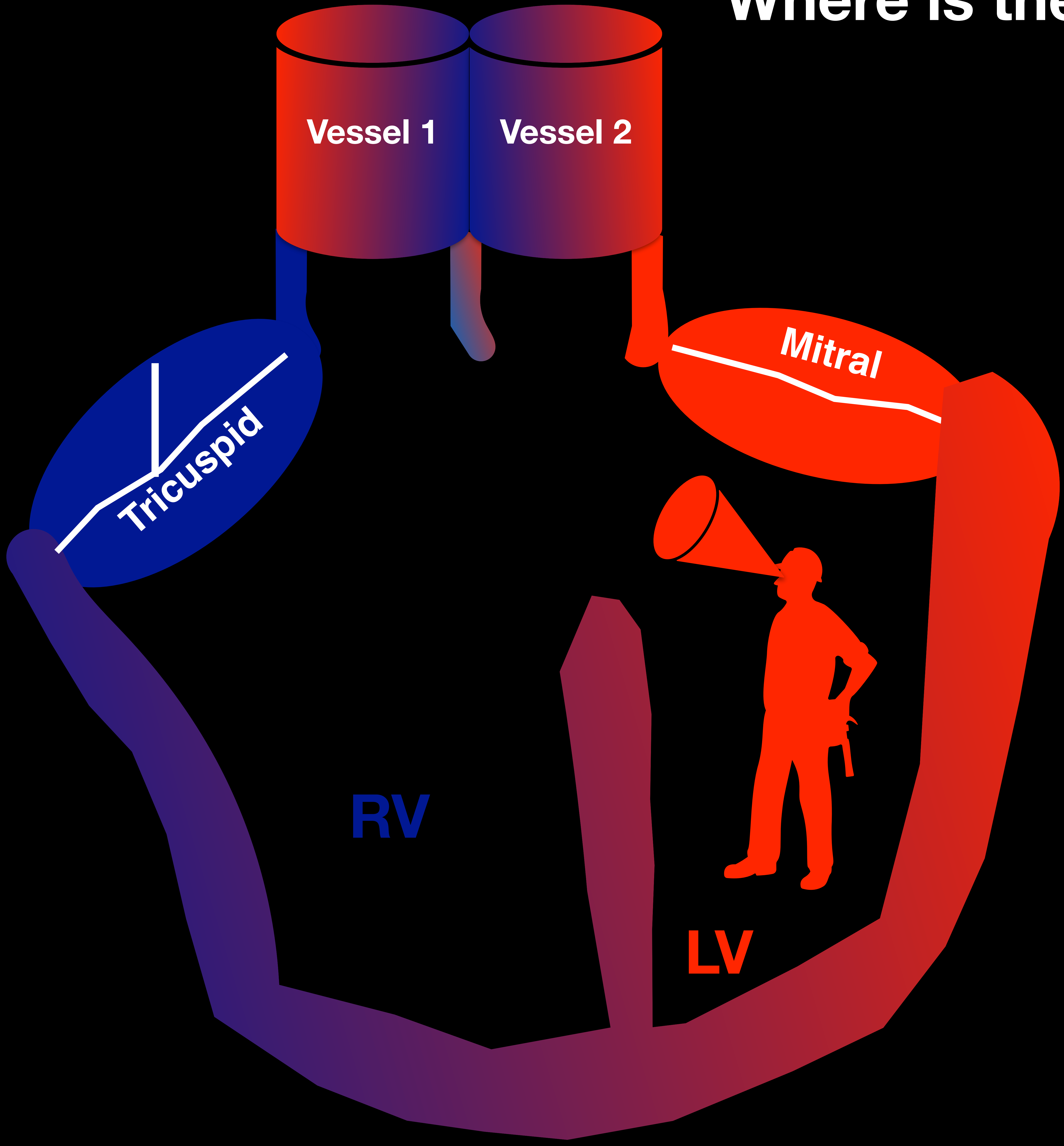


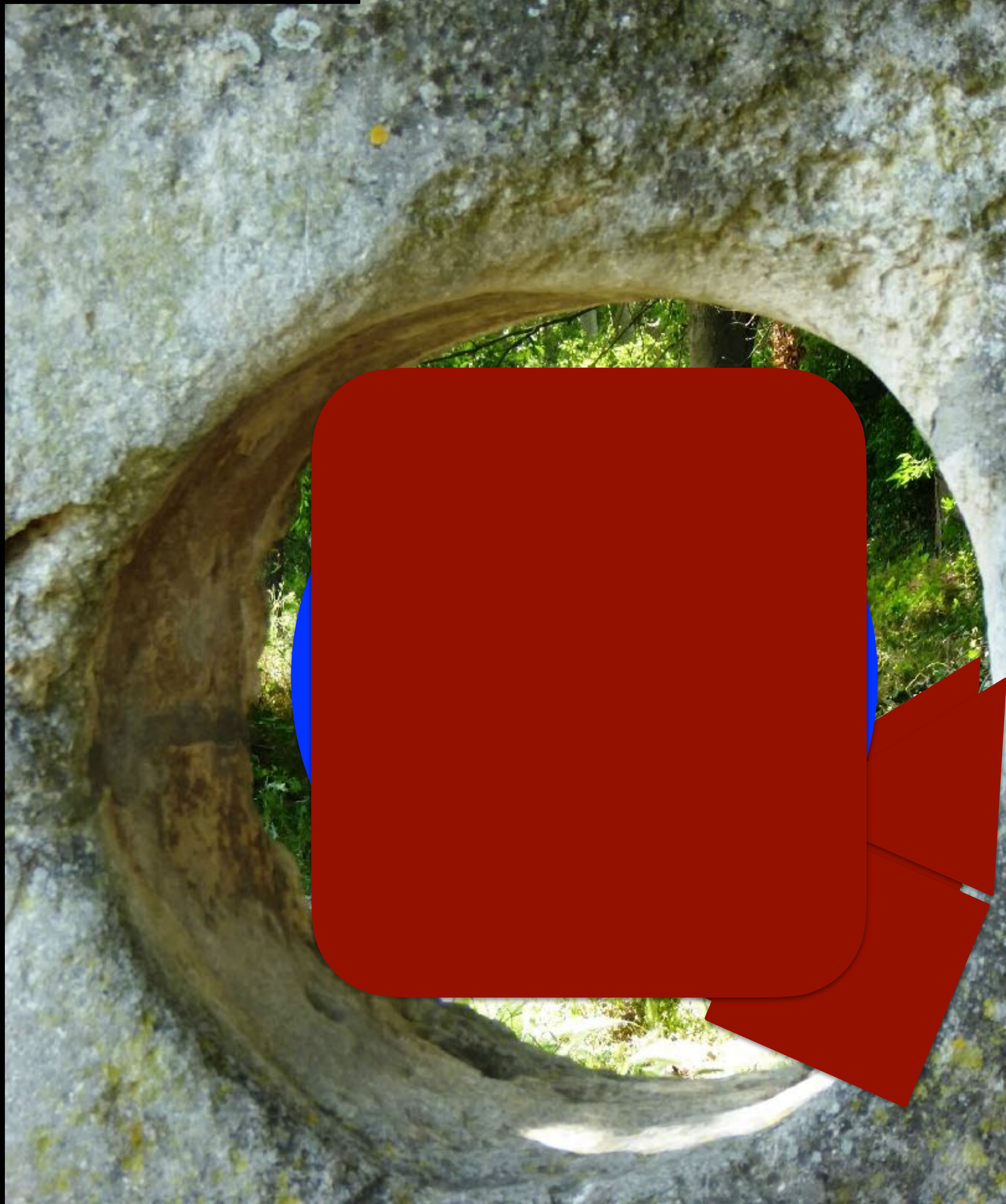
DORV - Relationship of VSD with great vessels



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

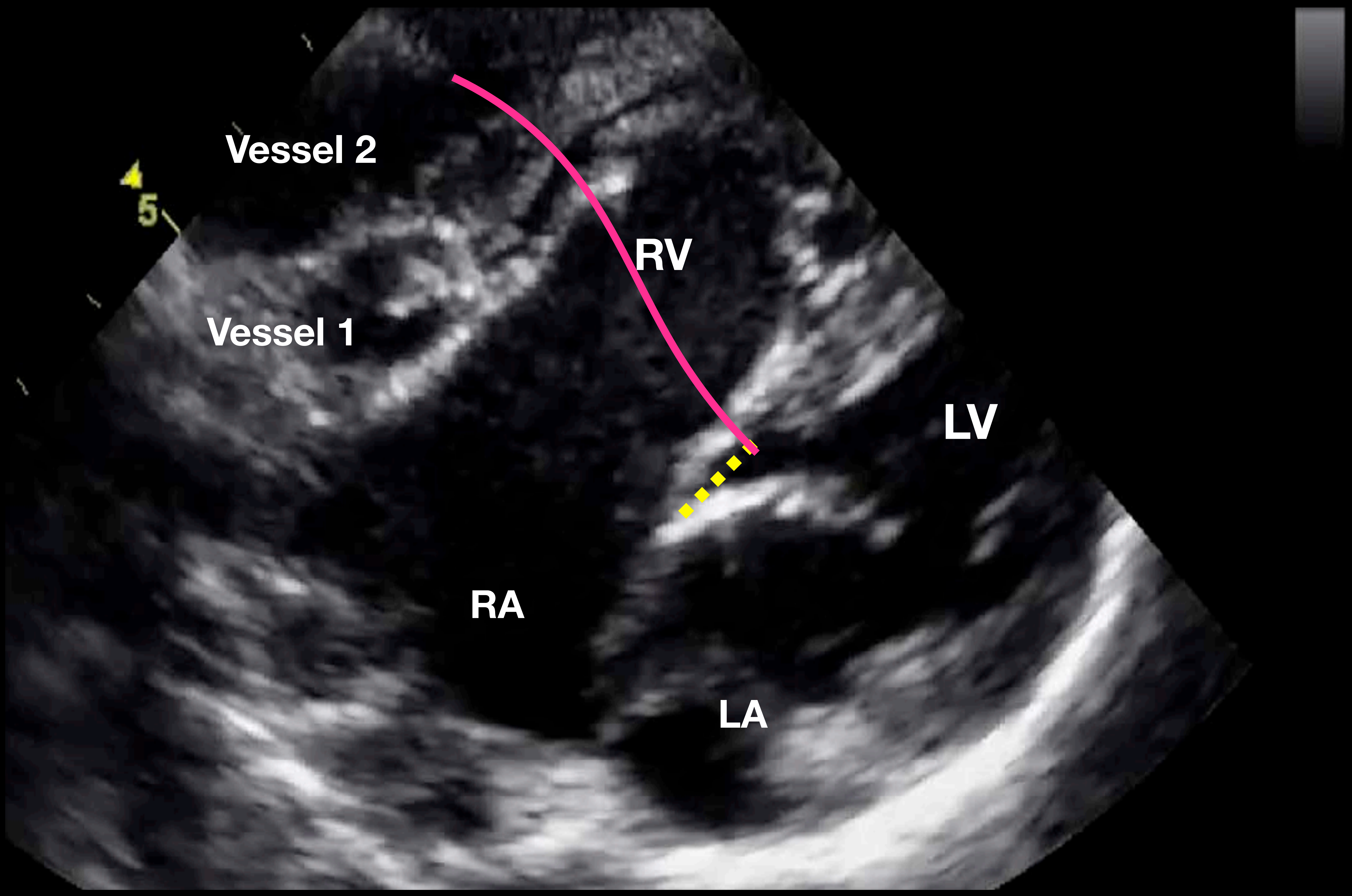
Where is the « VSD » in DORV ?





DORV

DORV non committed VSD



A-The position of the great vessels does not predict where is the VSD but
1-is of importance for surgical planning (ASO, Bex-Nikaido, Conotruncal rotation)
2-May complicate coronary transfer when ASO is indicated (side-by-side)

B-The position of the VSD with regards to flow will define the future LV outlet *but the strategy may also depend on*

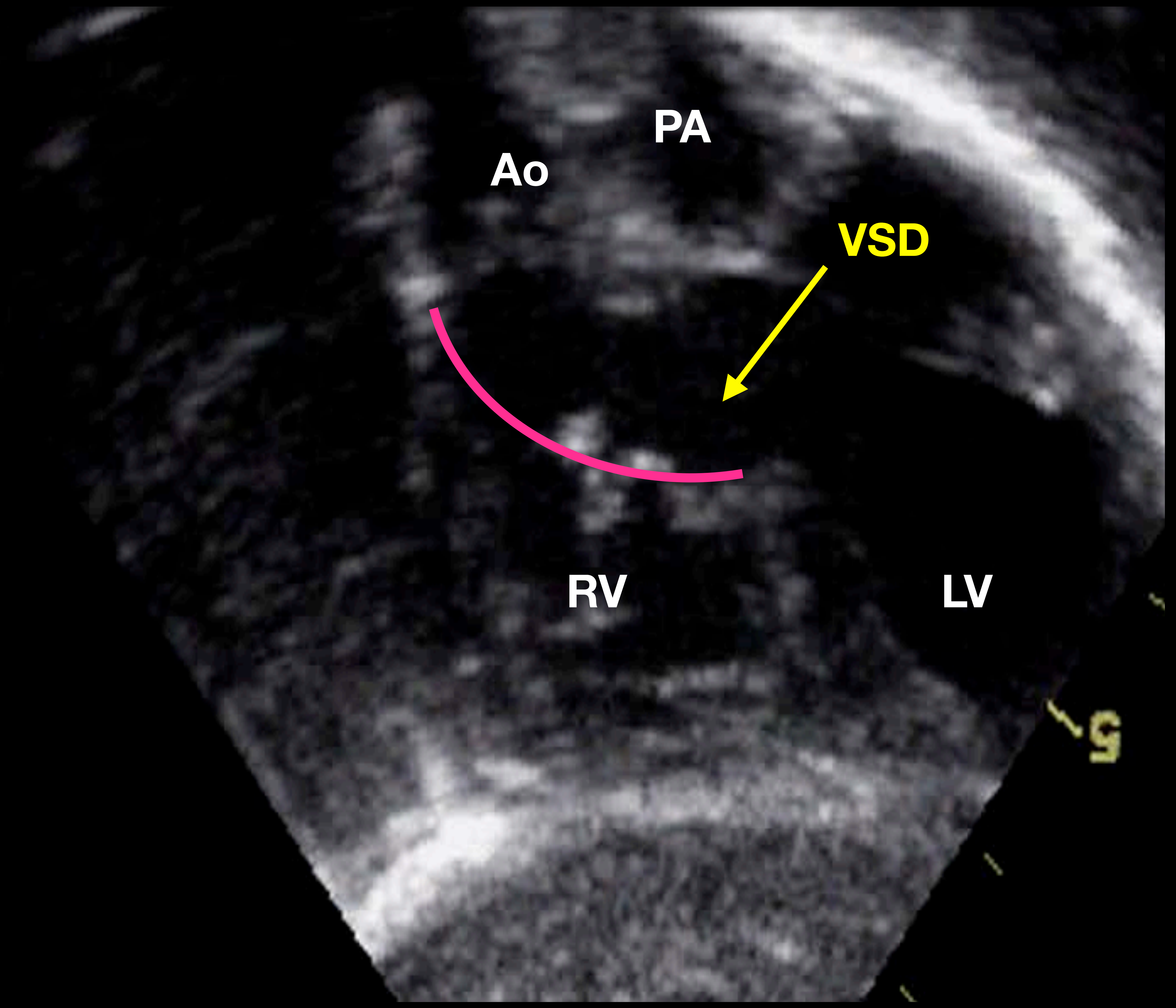
1-Time modifiable factors

- weight

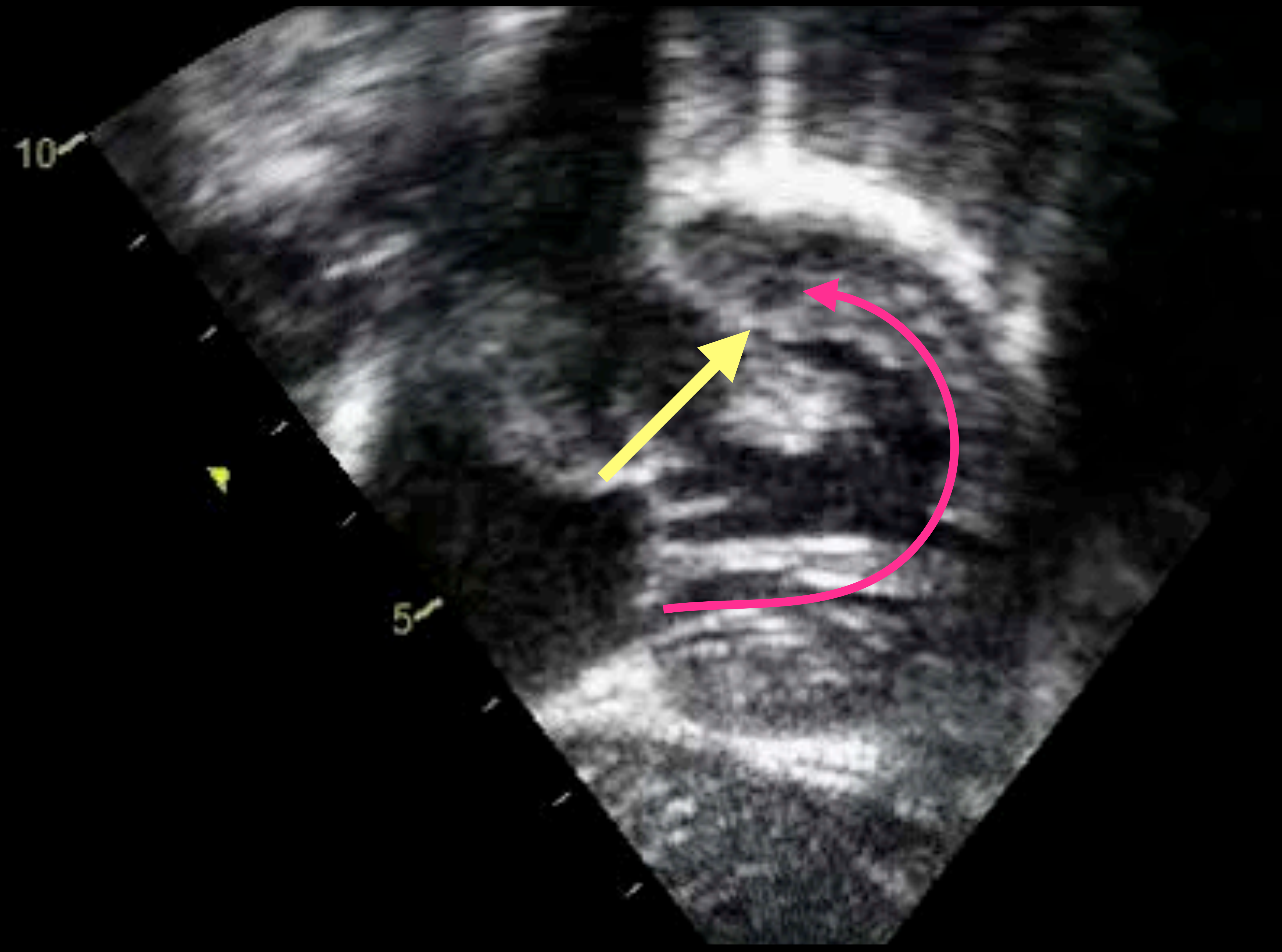
2-Contra-indication for theoretically optimal repair

DORV

« Late » DORV sub aortic VSD-Evaluation for IVR



DORV sub-aortic VSD



Distance between tricuspid valve and pulmonary valve

DORV

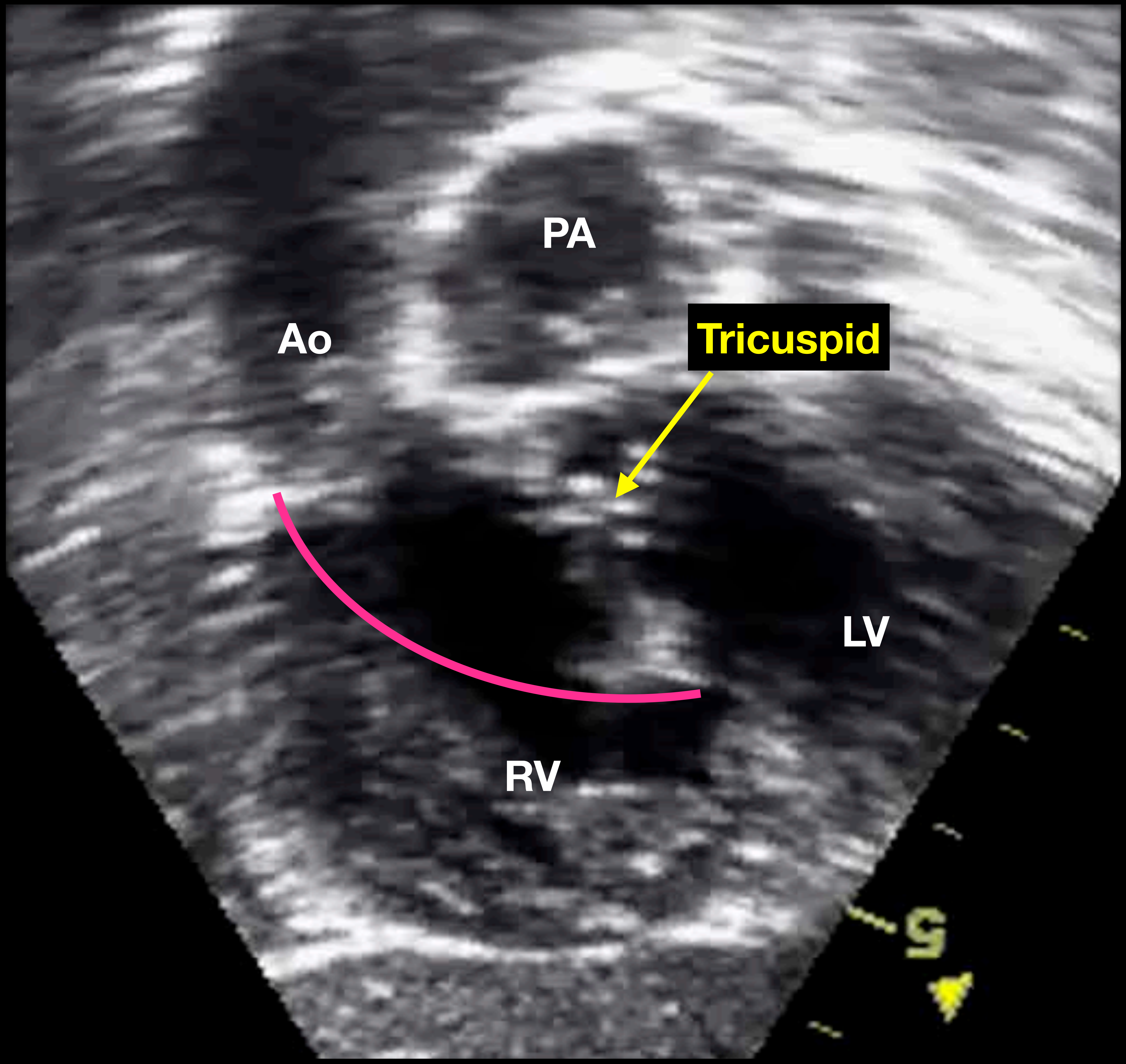
« Late » DORV sub pulmonary VSD-Evaluation for anatomical repair

DORV sub-pulmonary VSD

Short distance between tricuspid valve and pulmonary valve

DORV

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



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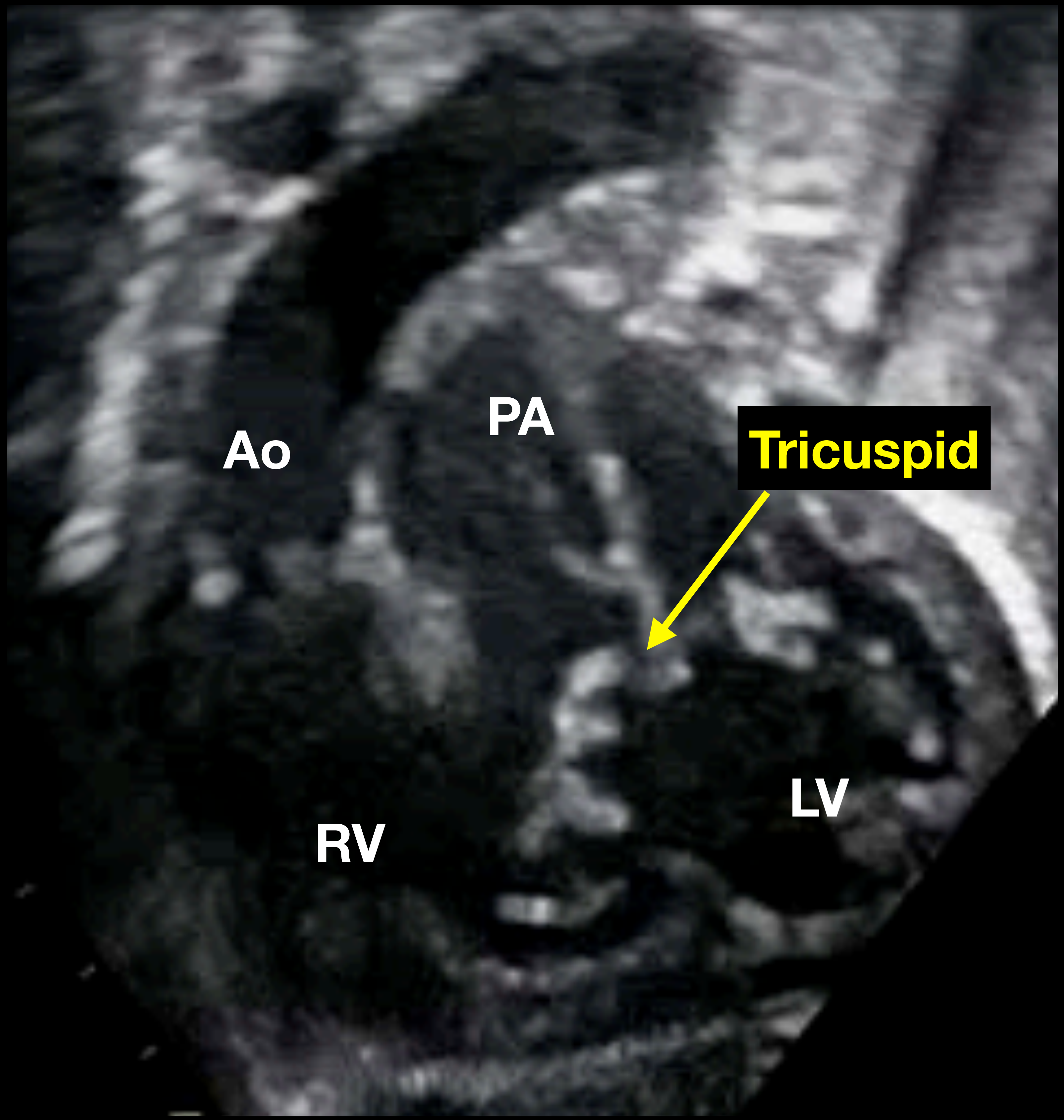
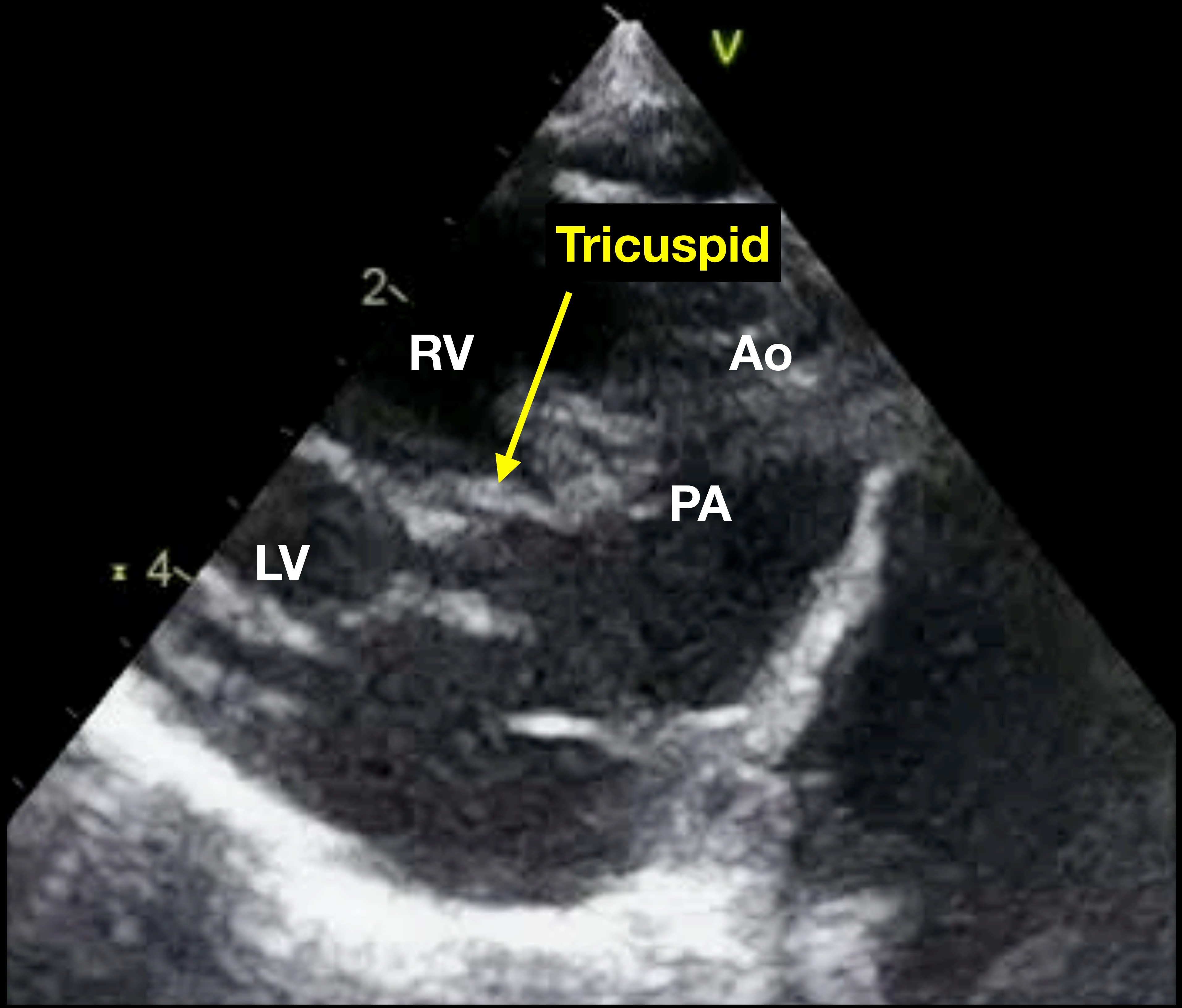
1-Time modifiable factors
- weight

2-Contra-indication for theoretically optimal repair
3-The associated risk of the future repair will lead to staged management adding other risks

DORV

« Late » DORV -Sub-pulmonary VSD





Rashkind, banding and Arterial switch operation + tricuspid valve repositioning

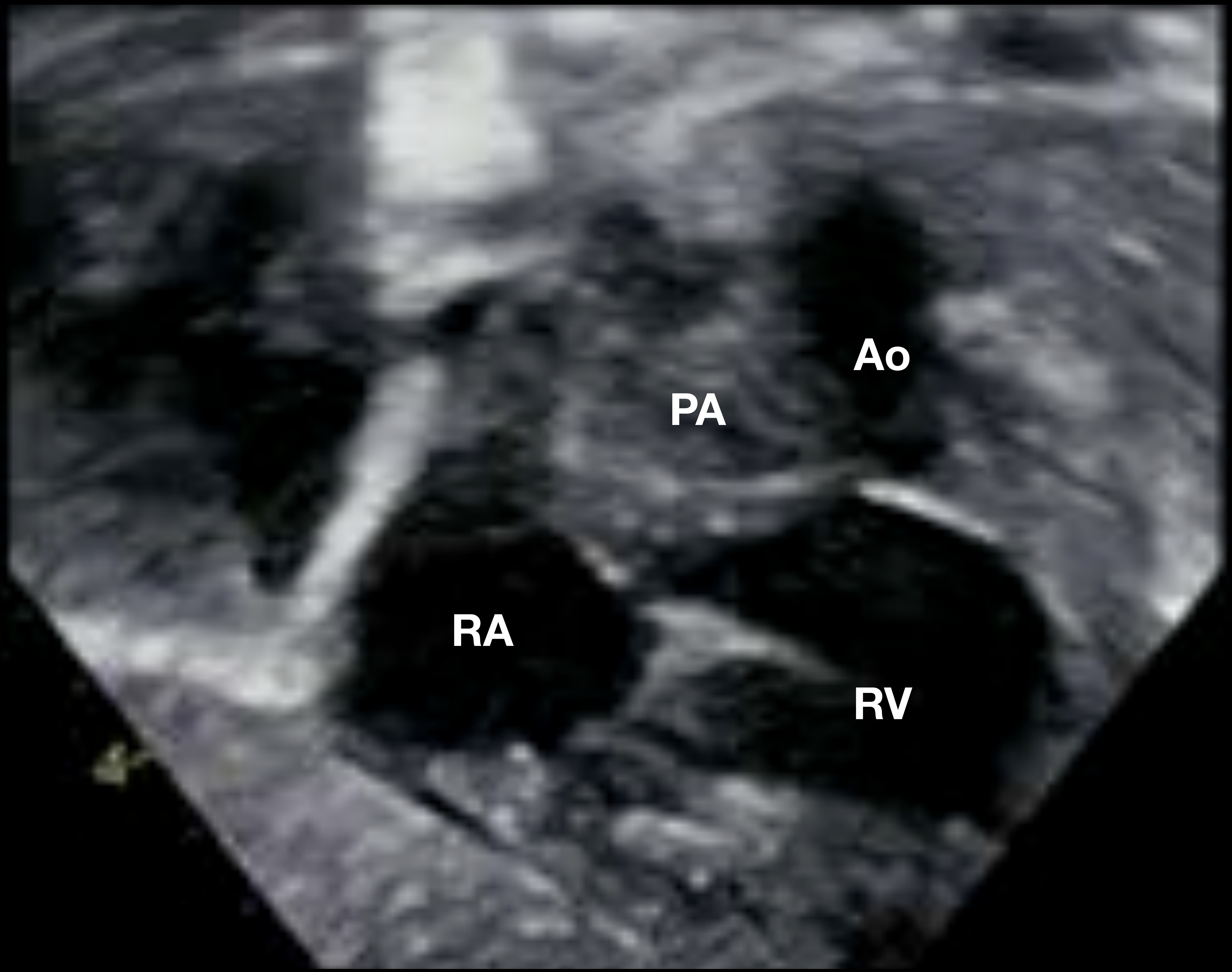
A-The position of the great vessels does not predict where is the VSD
1-is of importance for surgical planning (ASO, Bex-Nikaido, Conotruncal rotation)
2-May complicate coronary transfer when ASO is indicated (side-by-side)

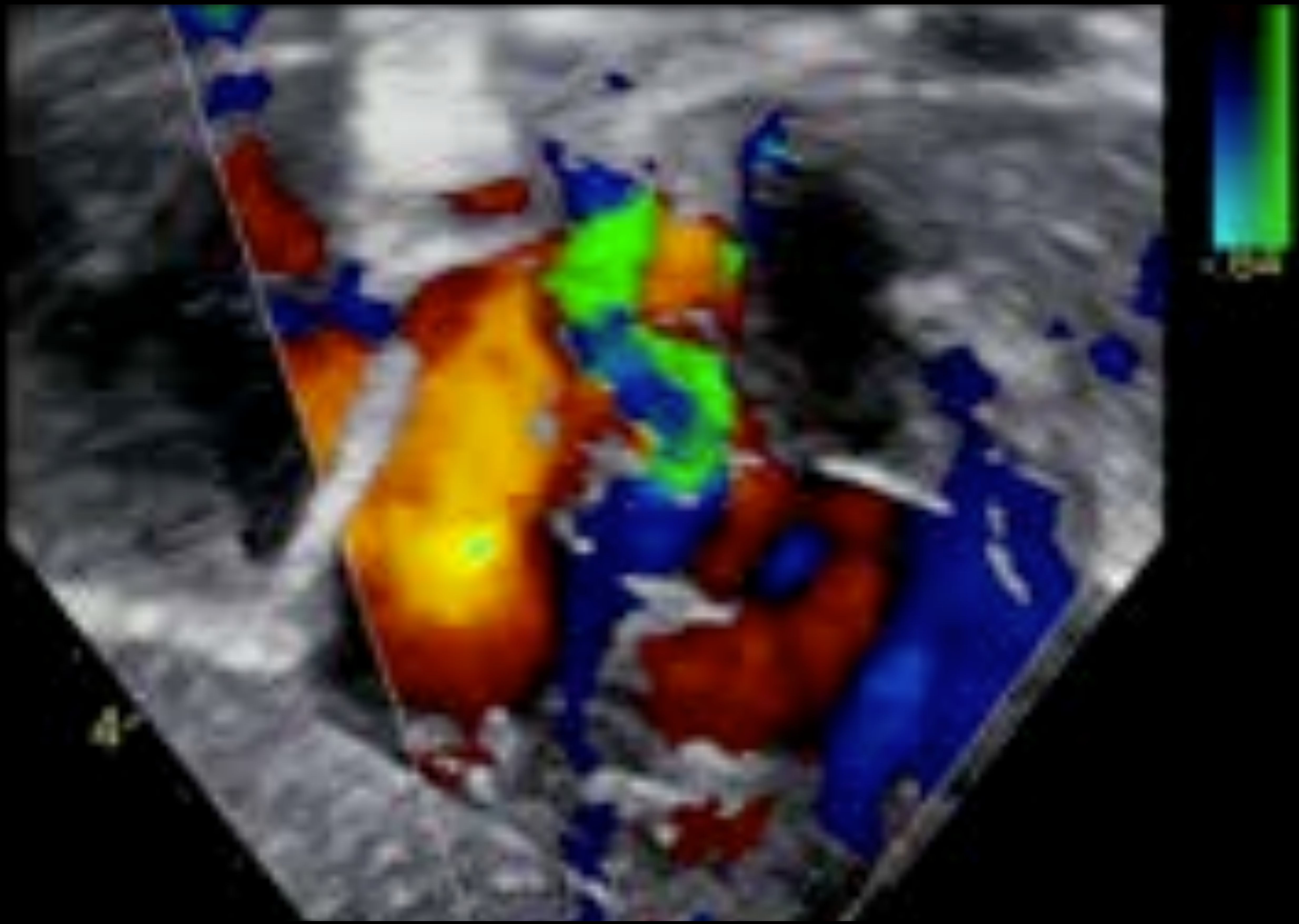
B-The position/physiology of the VSD with regards to flow will define the future LV outlet *<u>but the strategy may also depend on</u>*
1-Time modifiable factors
2-Contra-indication for theoretically optimal repair
3-The associated risk of the optimal repair will lead to staged repair adding other risks

C-The anatomy of the « future » right ventricle outflow tract will define the optimal type of repair, in association with the VSD physiology/position
1-If anatomical repair is feasible (IVR) or IVR-Fallot's strategy
2-If anatomical repair is not feasible
a-when pulmonary and aortic valves are normal (ASO)
b-when pulmonary valve is abnormal (not usable as aortic valve), numerous factors will influence final decision: size of pulmonary outflow tract, function of abnormal pulmonary valve, anatomy of coronary arteries (loops)…

DORV

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





Rashkind, BT shunt and REV operation

A-The position of the great vessels does not predict where is the VSD
1-is of importance for surgical planning (ASO, Bex-Nikaido, Conotruncal rotation)
2-May complicate coronary transfer when ASO is indicated (side-by-side)

B-The position/physiology of the VSD with regards to flow will define the future LV outlet *<u>but the strategy may also depend on</u>*
1-Time modifiable factors
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b-when pulmonary valve is abnormal (not usable as aortic valve), numerous factors will influence final decision: size of pulmonary outflow tract, function of abnormal pulmonary valve, anatomy of coronary arteries (loops)…

D-When a type of repair has been chosen, additional details may complicate or modify the final choice

1-Difficult REV operation (abnormal mitral valve insertions on the conal septum)
2-Anterior loop of coronary artery for Bex-Nikaido
3-Pulmonary artery branches anatomy and decision to insert a conduit with or without valve…


Initial palliation
PA Banding
Coarctation repair
Rashkind
BTT shunt
Uni- or biventricular management
Physiology/position of the VSD
Definition of future LV outlet
Definition of future RV outlet
IVR
IVR/Fallot
TCPC
ASO
REV
Bex-Nikaidoh
Rastelli
Conotruncal rotation

How the cardiologist can influence the surgical strategy ?

DORV is a pleomorphic defect and probably the paradigm CHD for personalized surgical strategy.

Detailed anatomical evaluation using appropriate modalities will define the strategy that will include initial palliation when required and optimal repair technique

Sequential in depth anatomical and physiological evaluations are required at all stages to confirm initial strategy or to reorient it in a more appropriate direction.

Cardiologist will not influence surgical strategy that depends on the patients and DORV characteristics but the « heart team » has to produce comprehensive informations to facilitate surgery for both planing and performing the optimal operation.

Details make perfection and perfection is not a detail.

Leonardo Da Vinci

Collective ignorance is our motivation
Curiosity is our strength
Research is our path

Individual experience is the brake
Indifference is the weakness
Argument from authority is the threat