



# Tetralogy of Fallot

Damien Bonnet

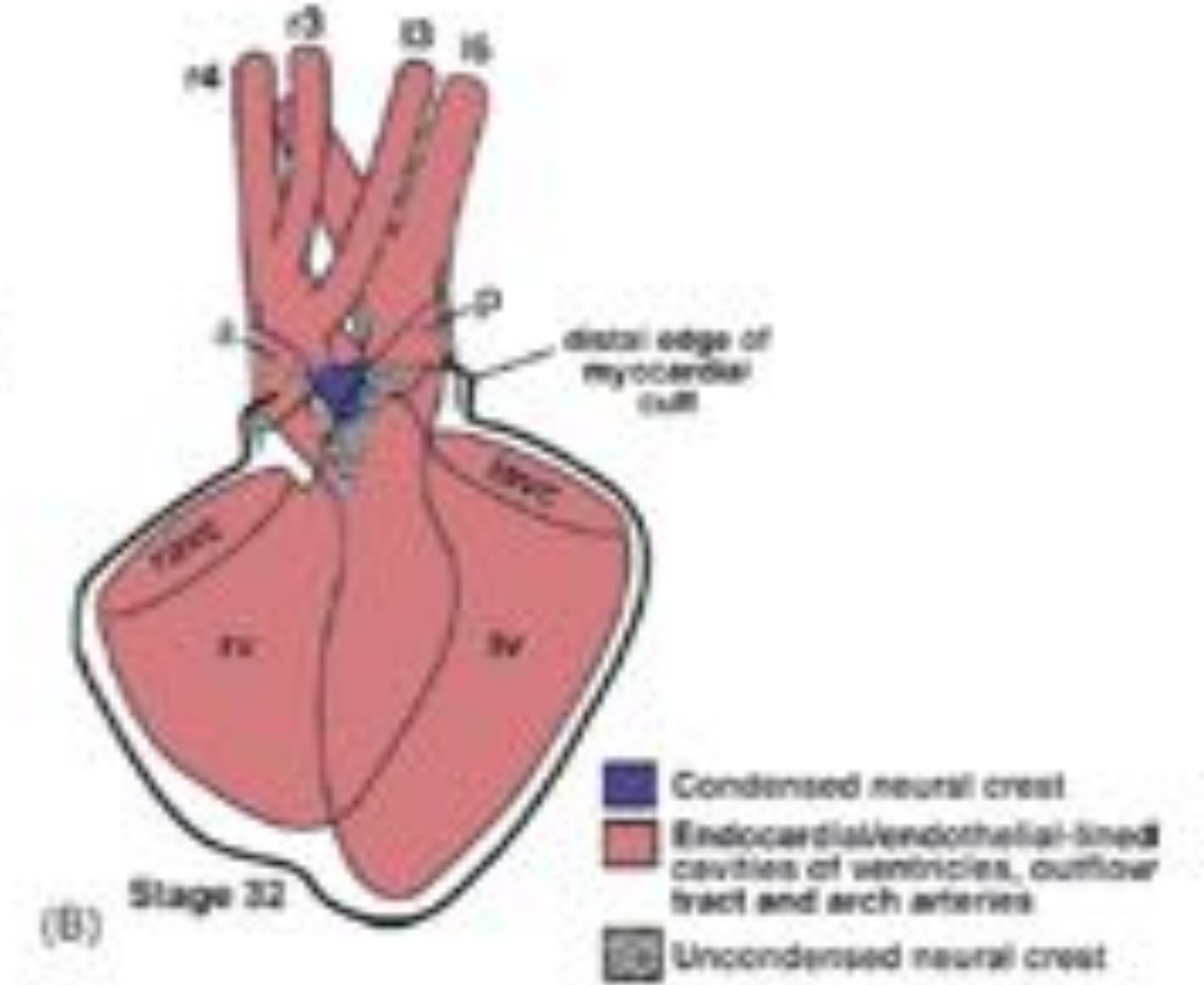
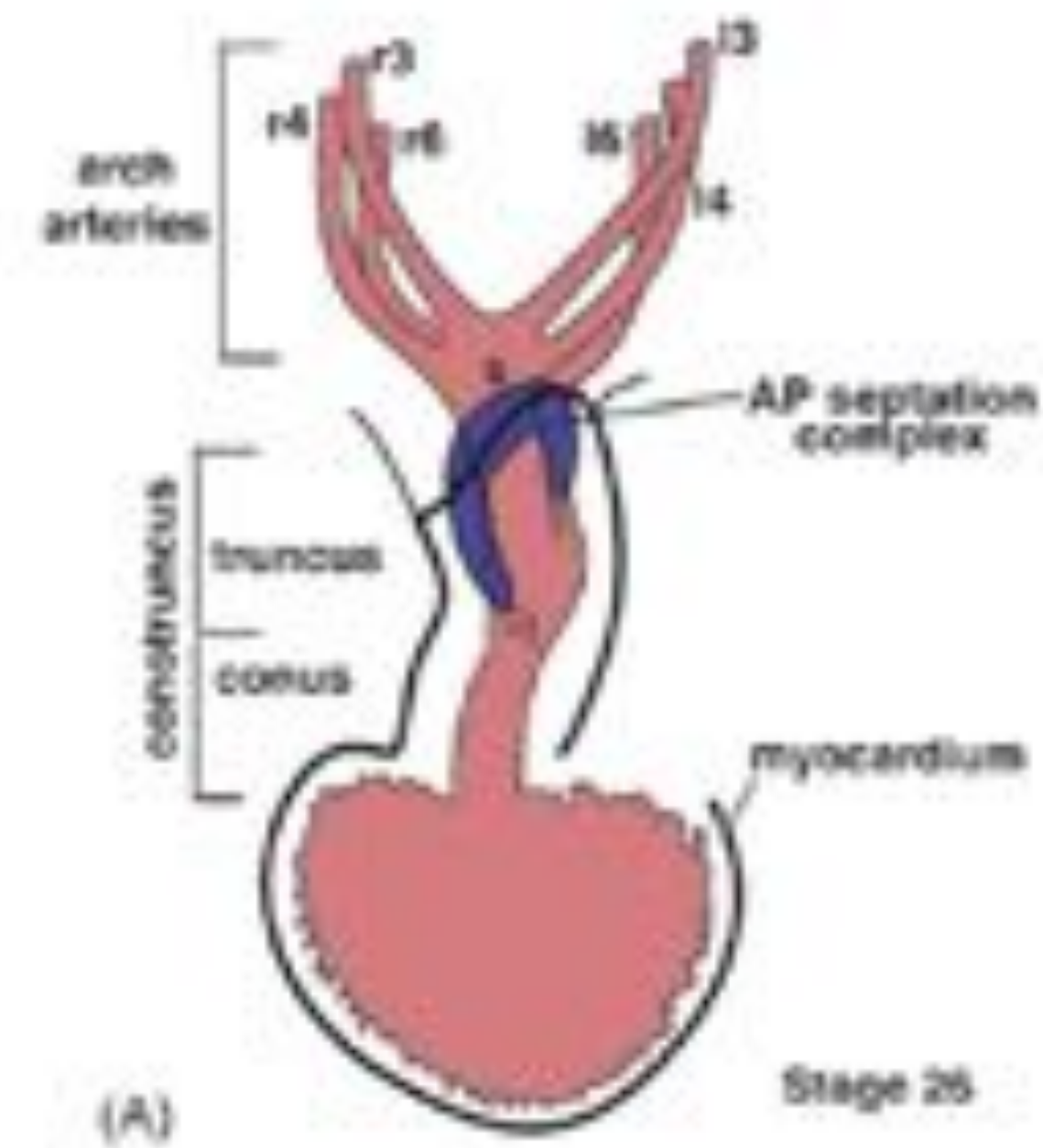
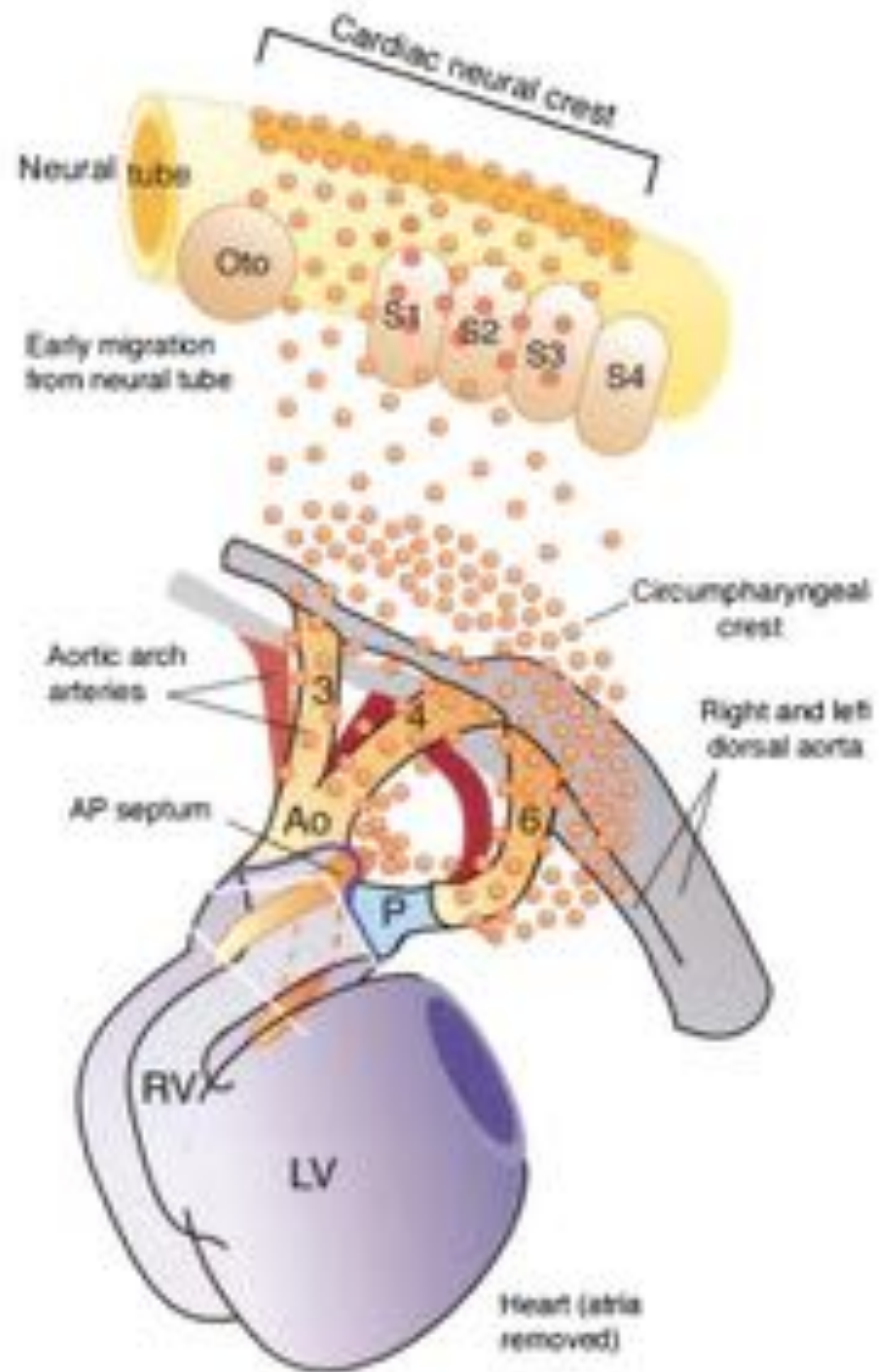
Unité médico-chirurgicale de Cardiologie Congénitale et Pédiatrique  
Hôpital Universitaire Necker Enfants malades – APHP, Université Paris Descartes, Sorbonne Paris Cité  
IcarP Cardiology, Institut Hospitalo-Universitaire IMAGINE

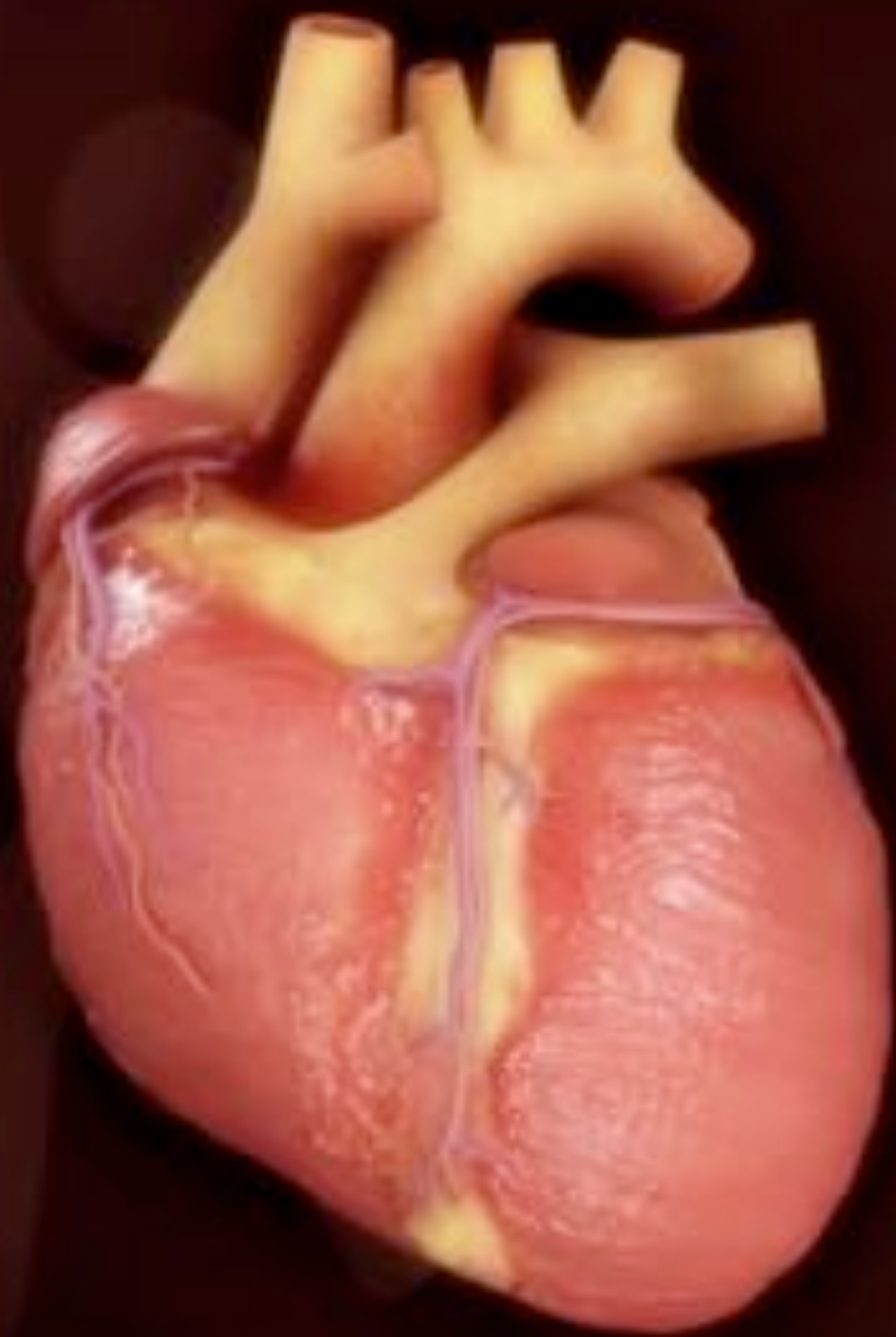
Centre de Référence Maladies Rares  
Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares  
Maladies Cardiaques Héréditaires- CARDIOGEN



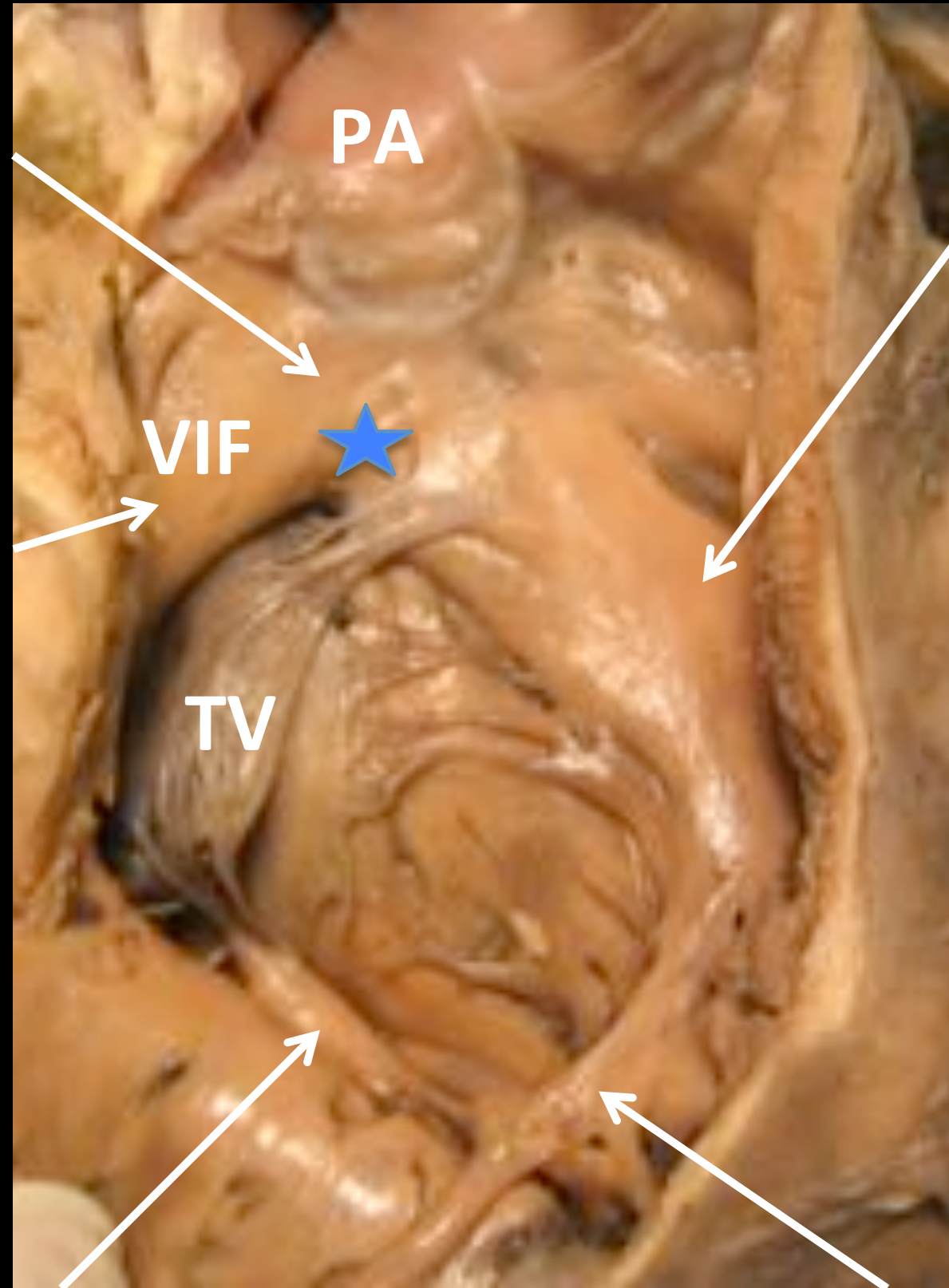
# Migration of neural crest cells into the outflow tract





Subpulmonary conus

Ventriculo-infundibular fold



PA

VIF

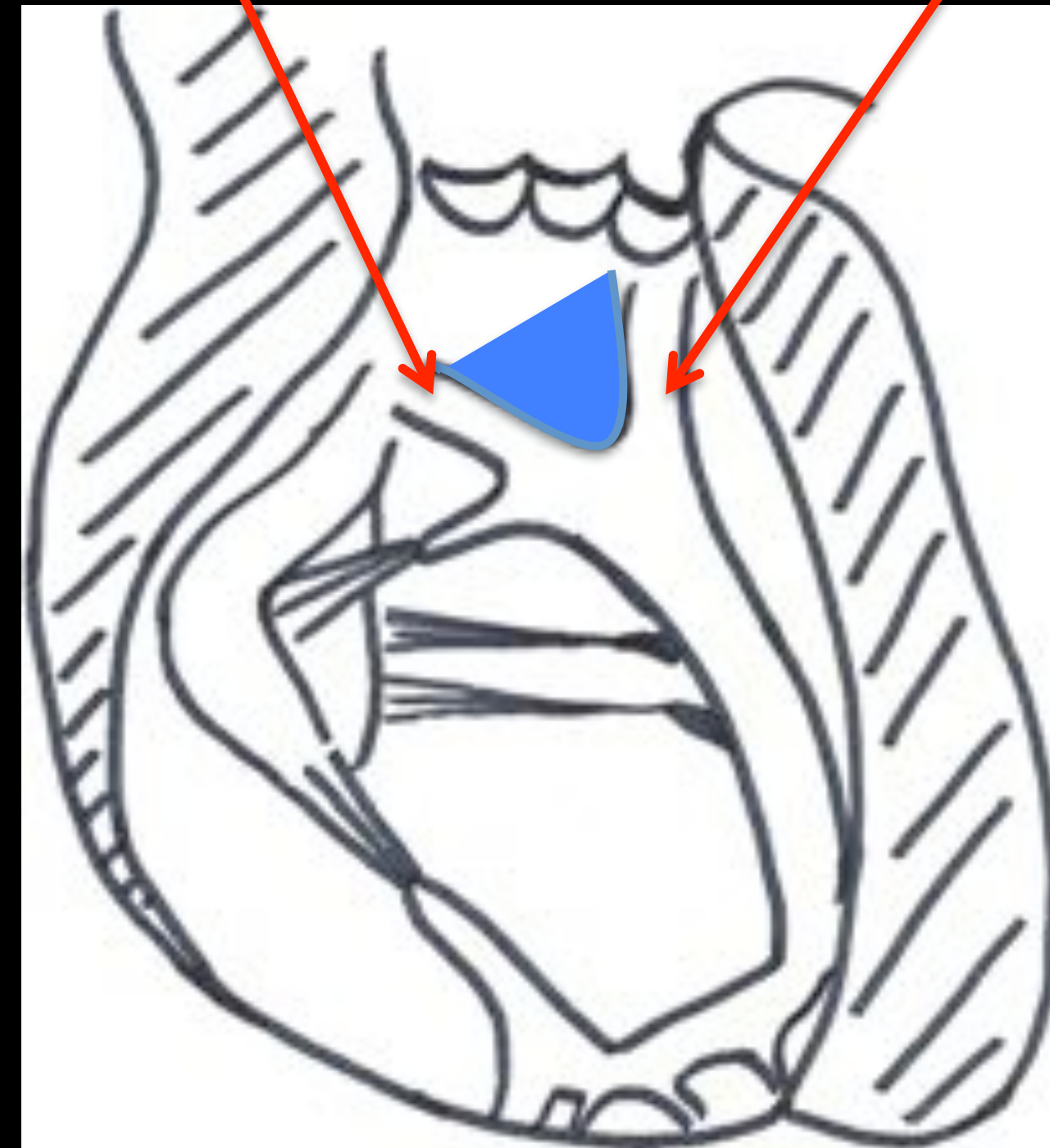
TV

Septal band

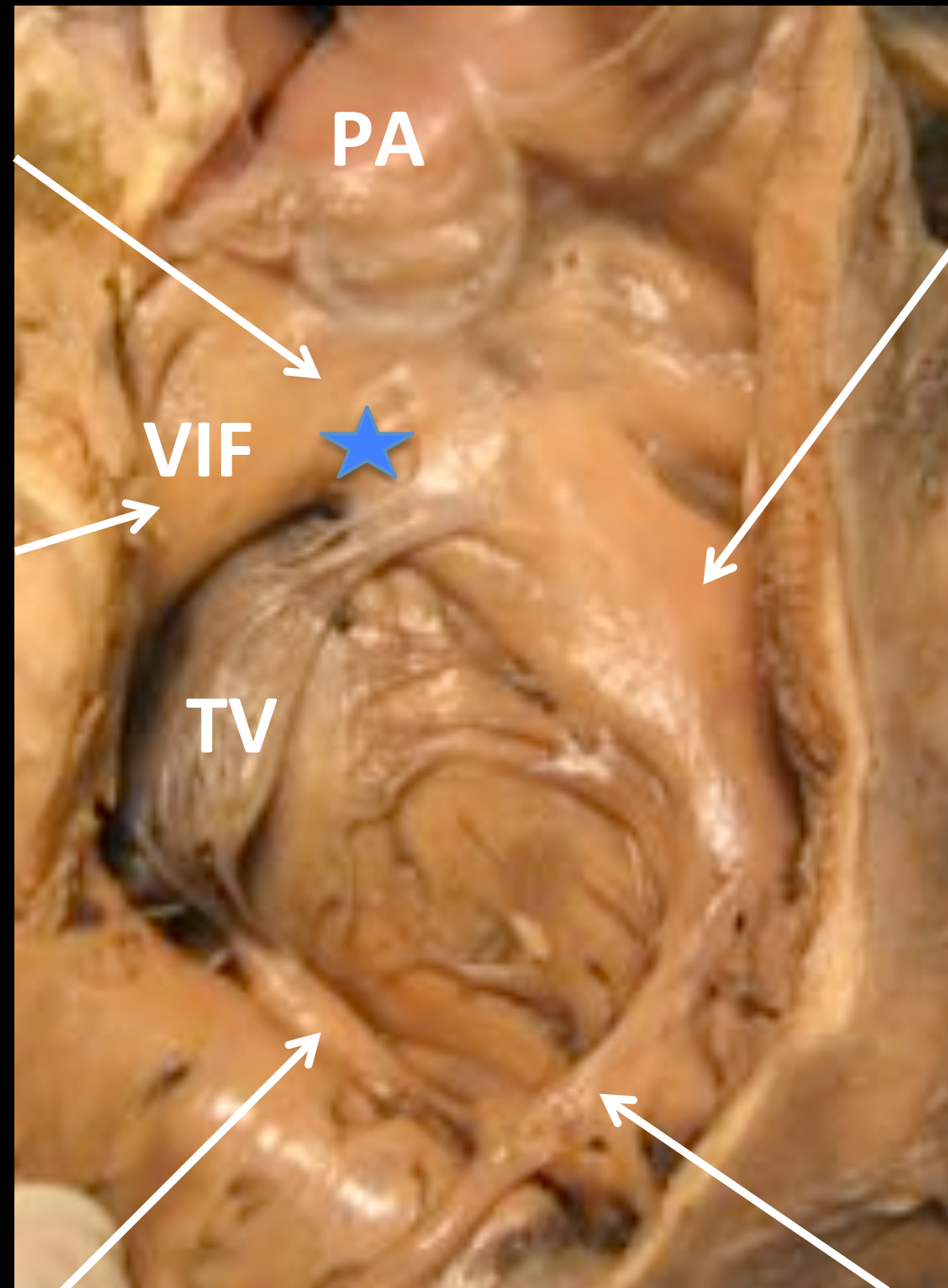
Anterior papillary muscle of the tricuspid valve Moderator band

Postero-inferior limb of the septal band

Anterior limb of the septal band



Subpulmonary conus

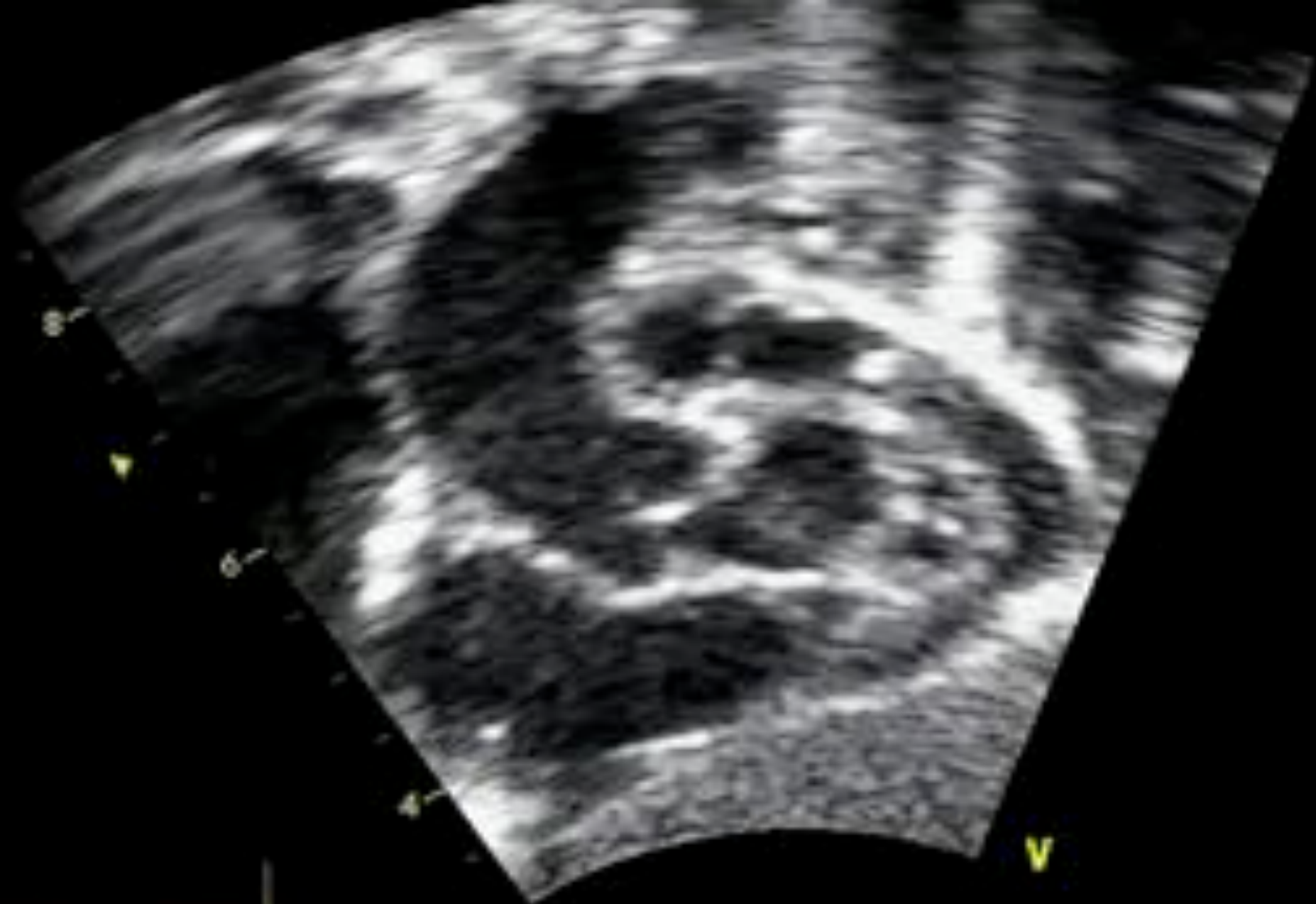


Ventriculo-infundibular fold

Septal band

Anterior papillary muscle of the tricuspid valve Moderator band





71  
HR



1:160

# **Main topics in tetralogy of Fallot**

- Prenatal diagnosis
- Perinatal management
- Strategy for repair
- Late outcomes

# **Prenatal diagnosis of tetralogy of Fallot**

## **What are the main issues ?**

1. Associated cytogenetic and extra-cardiac anomalies



Trisomie 21



Délétion 22q11



Alagille



CHARGE

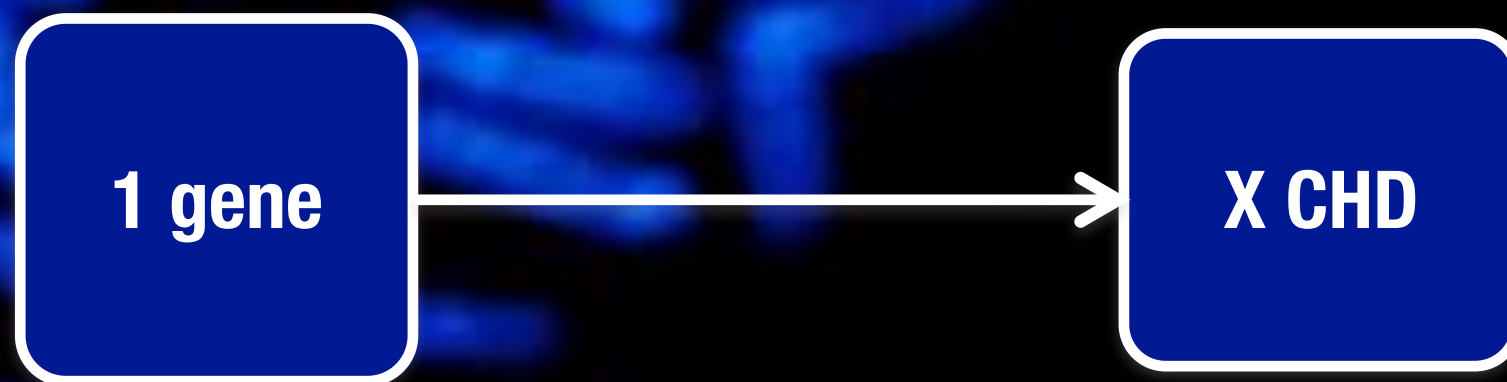
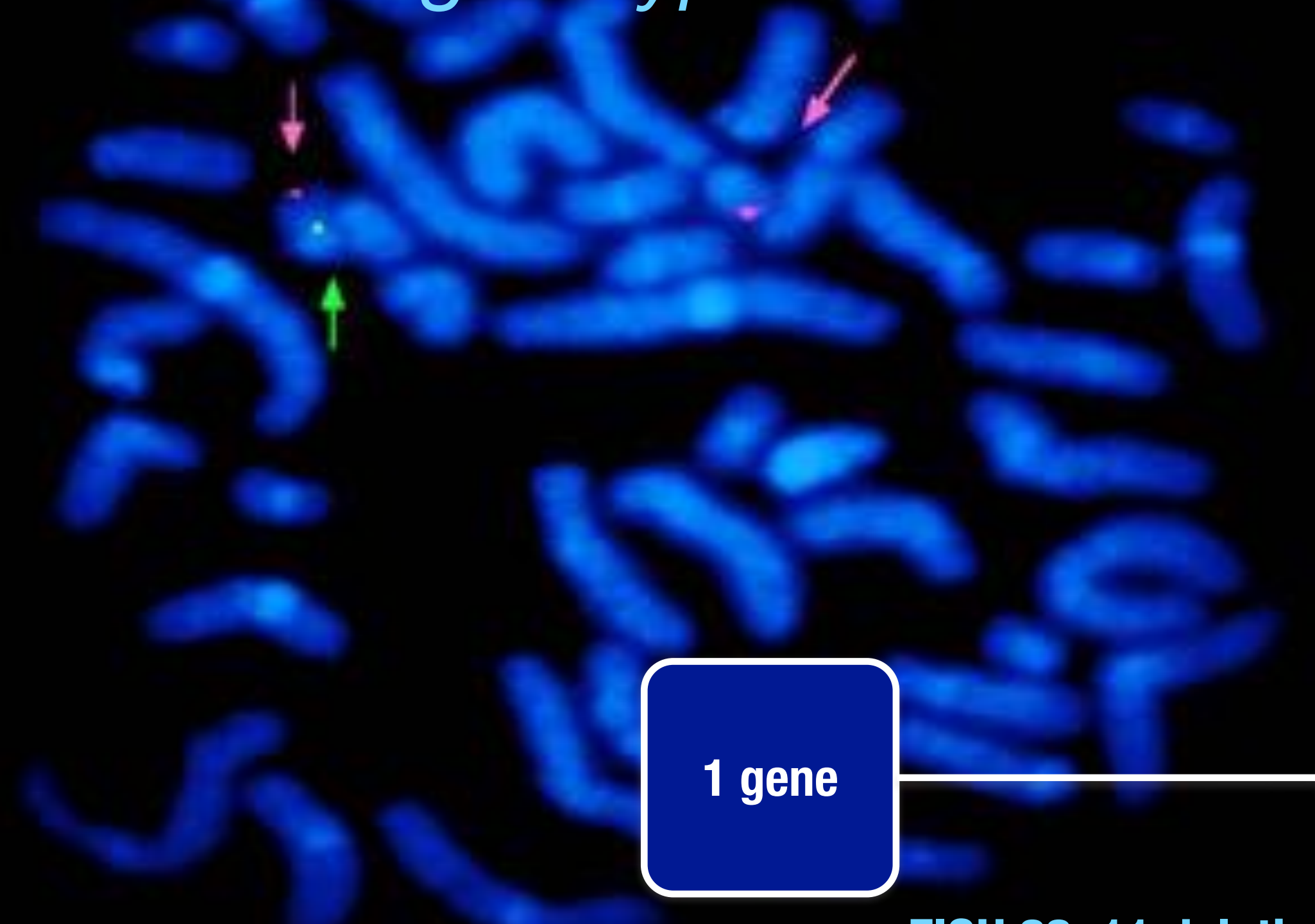


Waardenburg

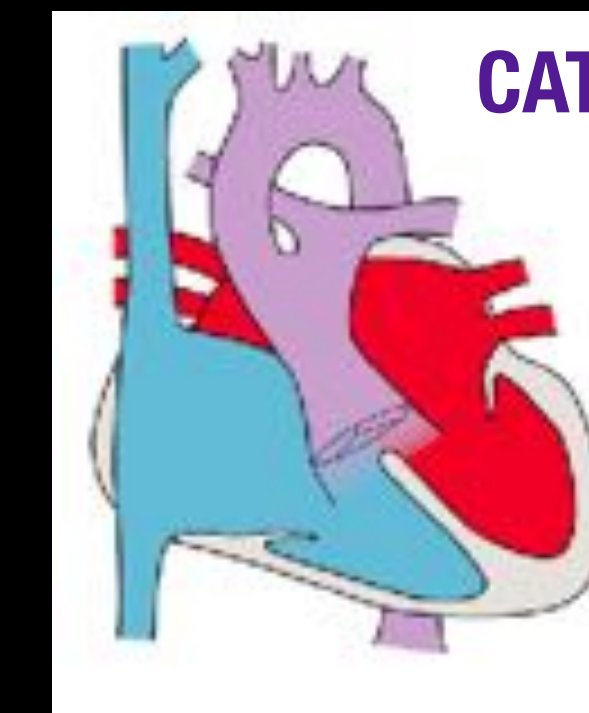
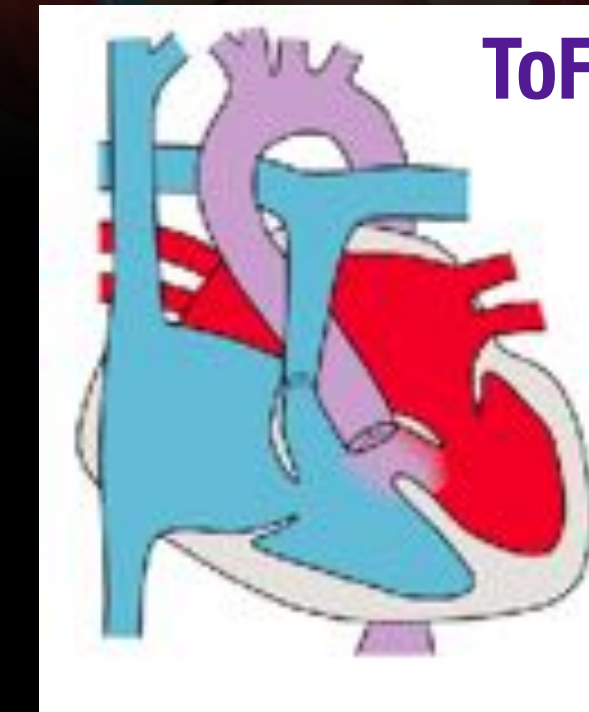
10 % karyotypic anomalies on standard analysis  
18% 22q11 deletion : 15% in ToF-PS, 26% in ToF-PA

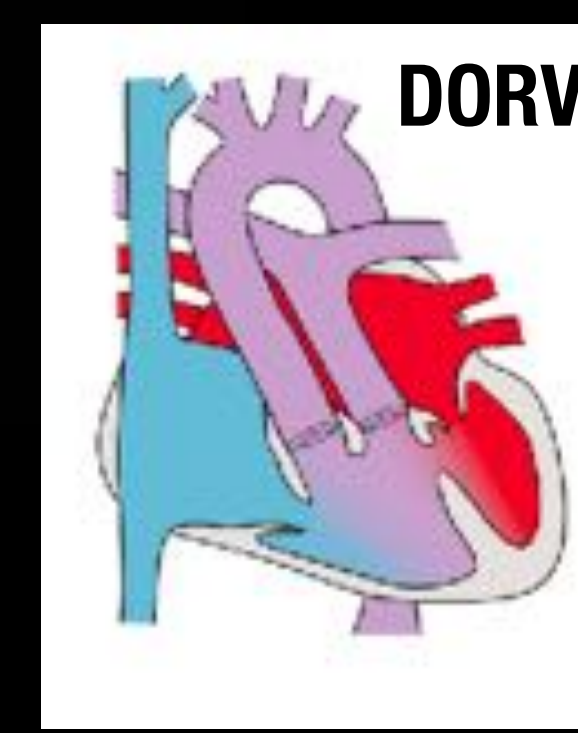
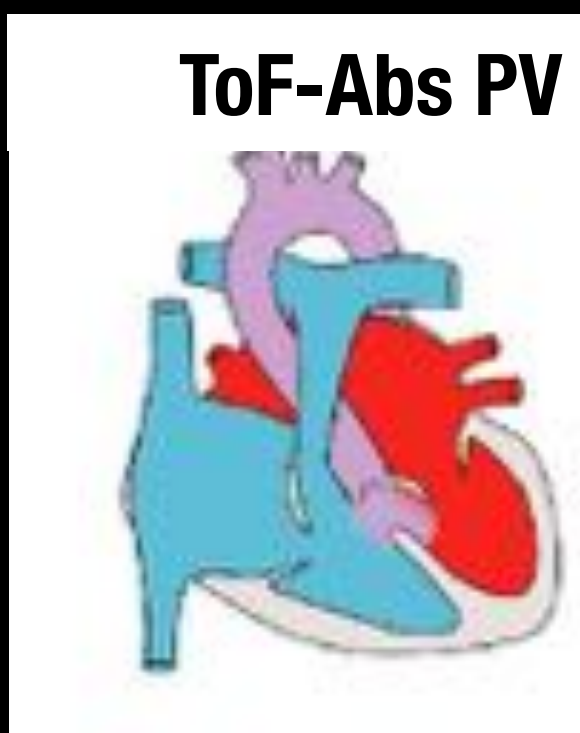
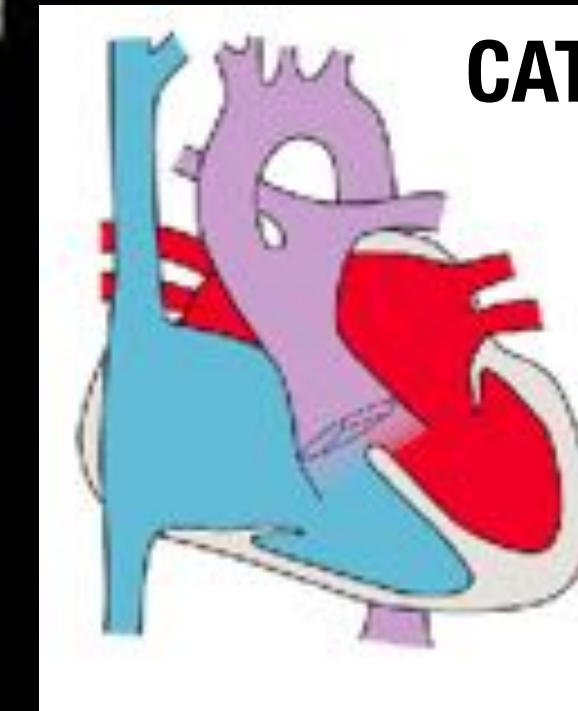
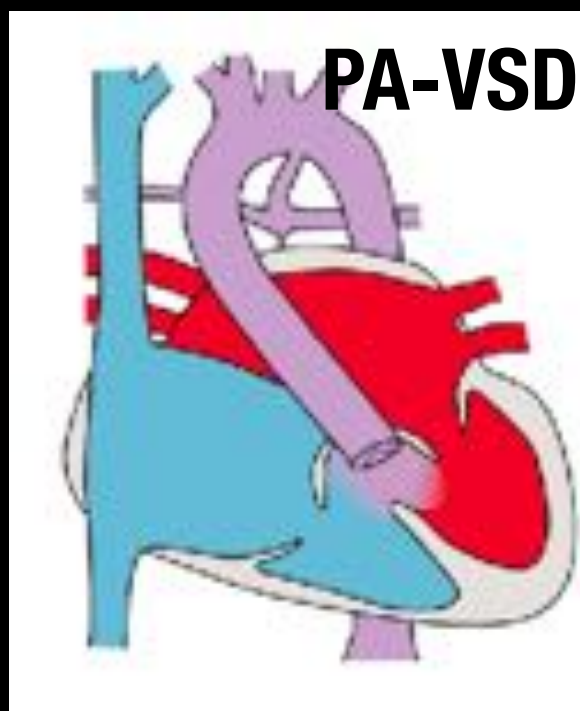
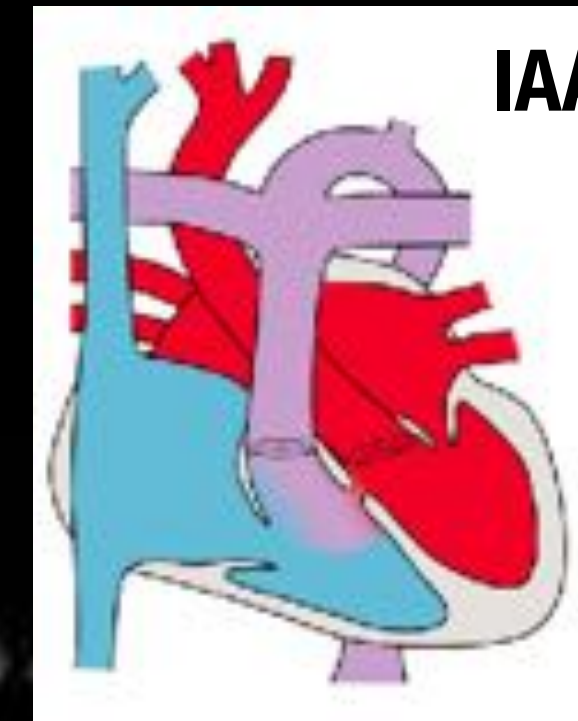
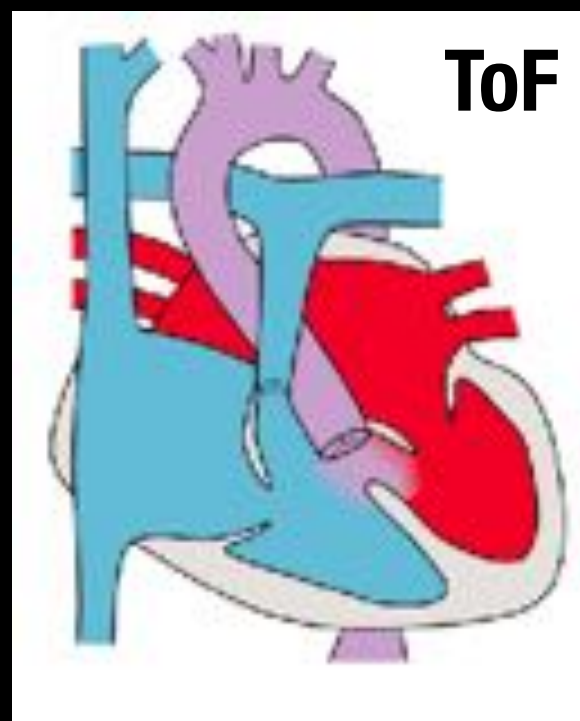


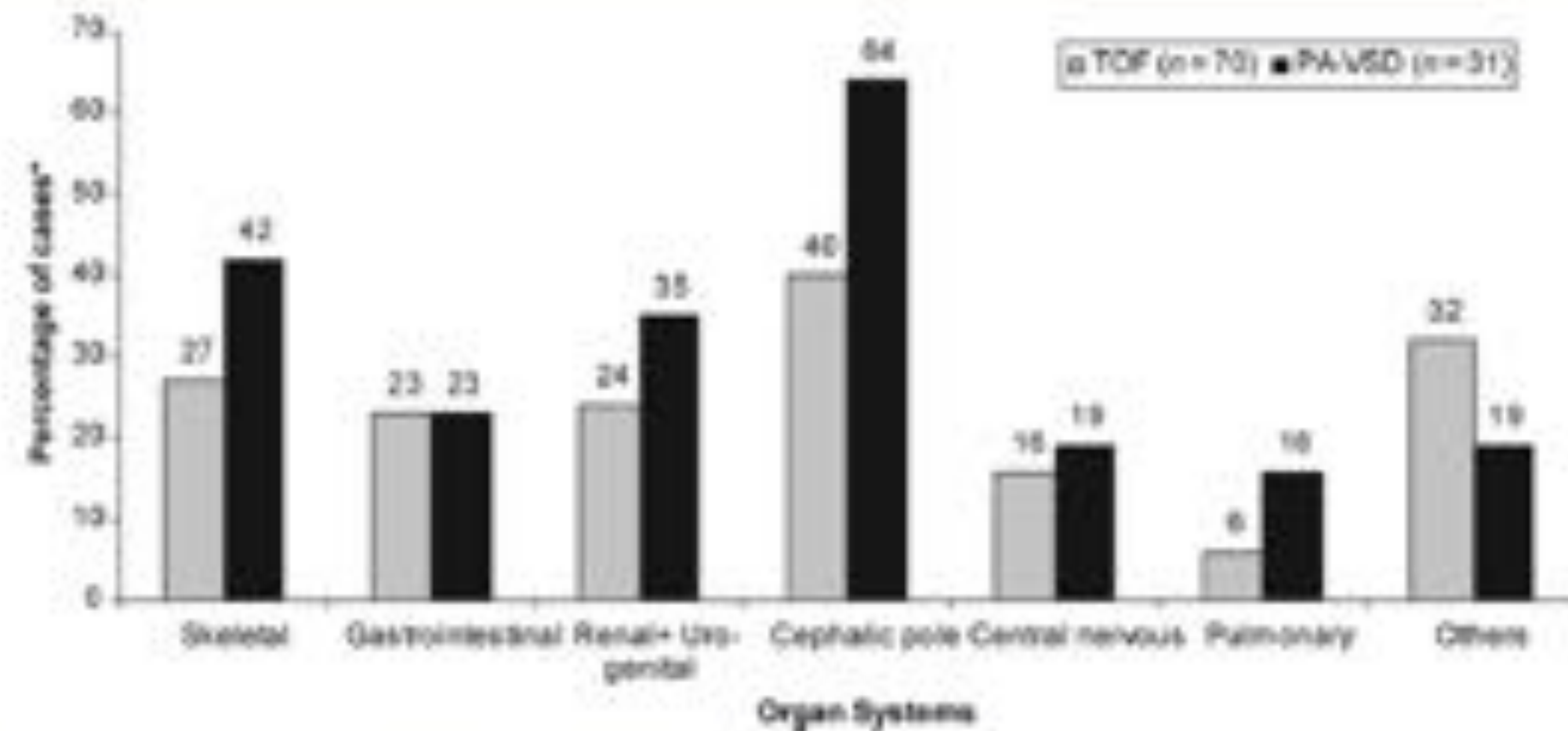
Phenotypic heterogeneity  
*One genotype-Different cardiac phenotypes*



FISH 22q11 deletion







**Figure 1** Details on extracardiac anomaly in foetal TOF and PA-VSD. Others, growth and endocrinological abnormalities, mental retardation, ophthalmological anomalies, unic umbilical artery, cystic hygroma; TOF, tetralogy of Fallot; PA-VSD, pulmonary atresia with ventricular septal defect; the asterisk indicates the number of abnormalities by system/number of fetuses with associated extracardiac malformations per type of CHD (TOF or PA-VSD).

One third of fetuses with ToF had extra cardiac anomalies  
 15% had intra-uterine growth retardation

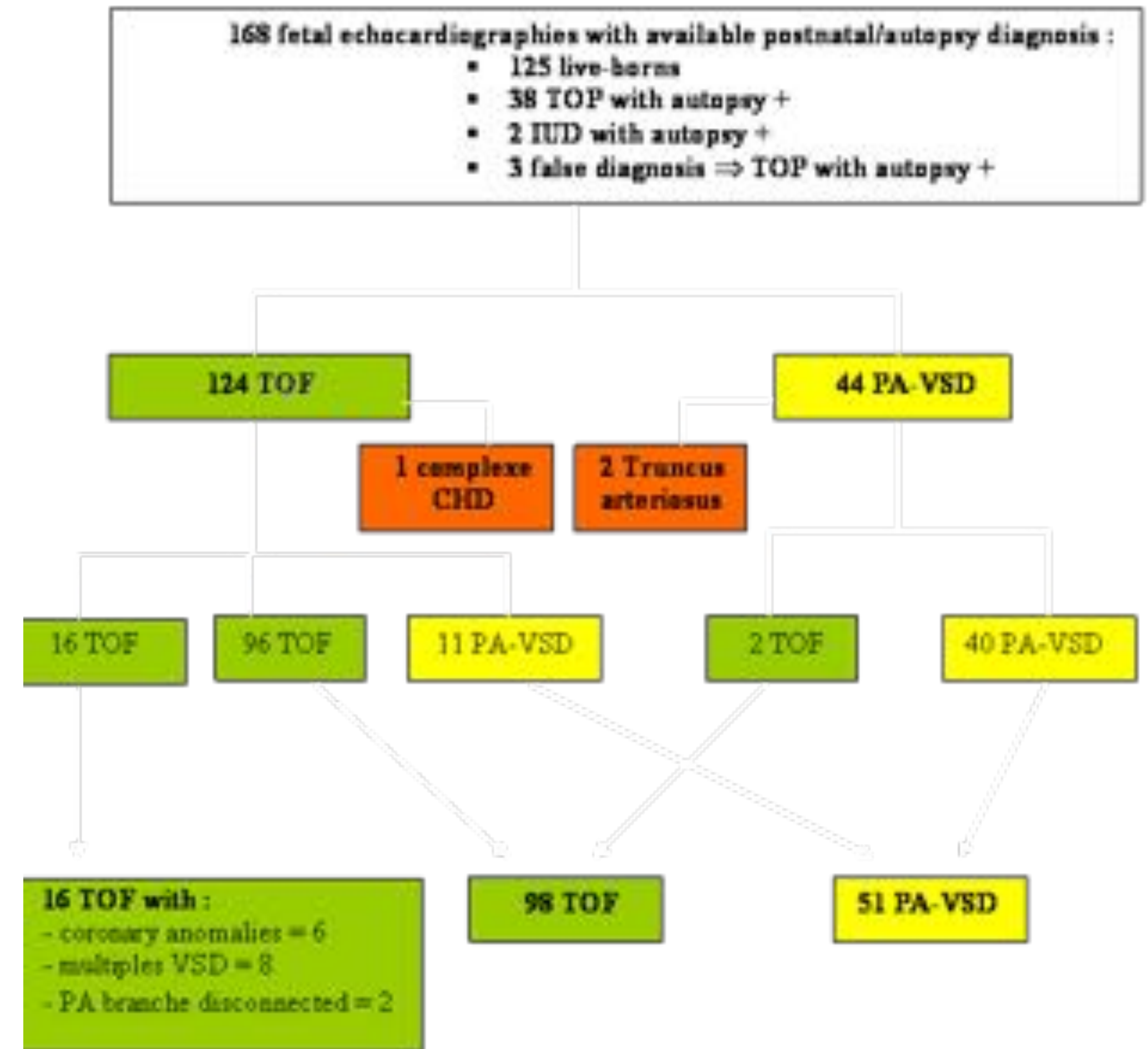
# **Prenatal diagnosis of tetralogy of Fallot**

## **What are the main issues ?**

1. Associated cytogenetic and extra-cardiac anomalies
2. Accuracy of diagnosis and decision for in utero transfer

# Accuracy of diagnosis

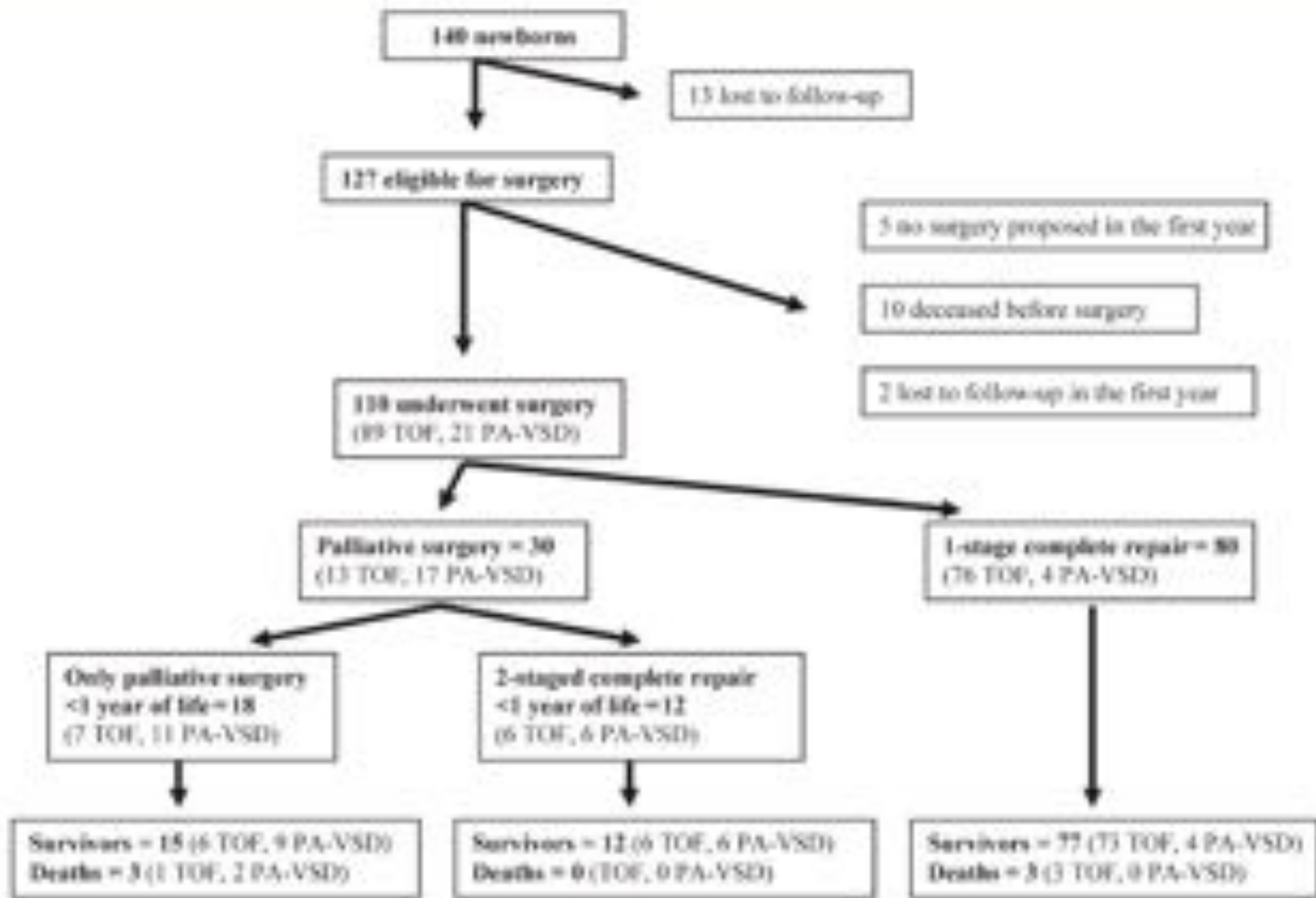
- ToF-PS can be ToF-PA
- ToF-PA can be Common arterial trunk
- Left pulmonary artery branch can arise from the arterial duct



# **Prenatal diagnosis of tetralogy of Fallot**

## **What are the main issues ?**

1. Associated cytogenetic and extra-cardiac anomalies
2. Accuracy of diagnosis and decision for in utero transfer
3. Prediction of repair



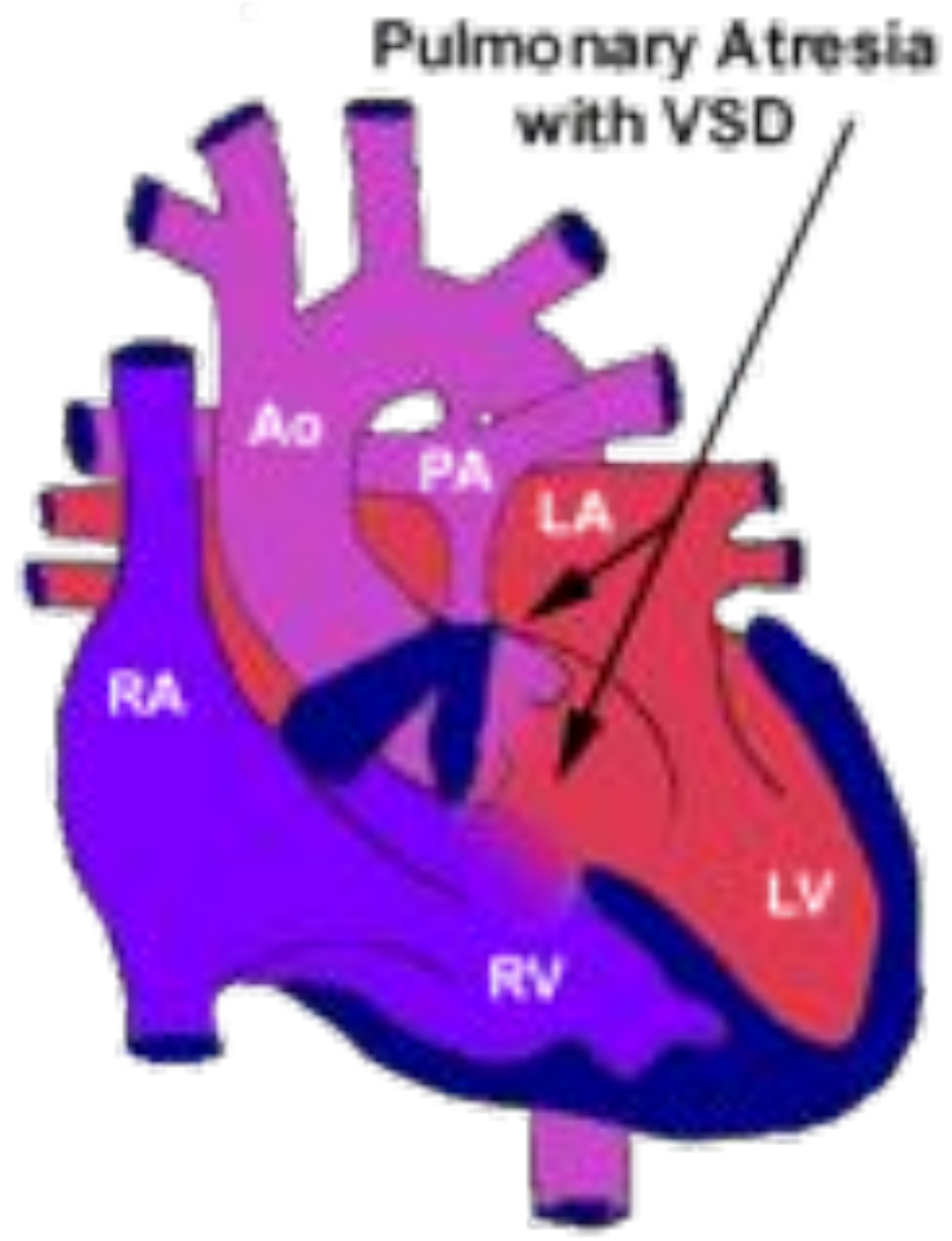
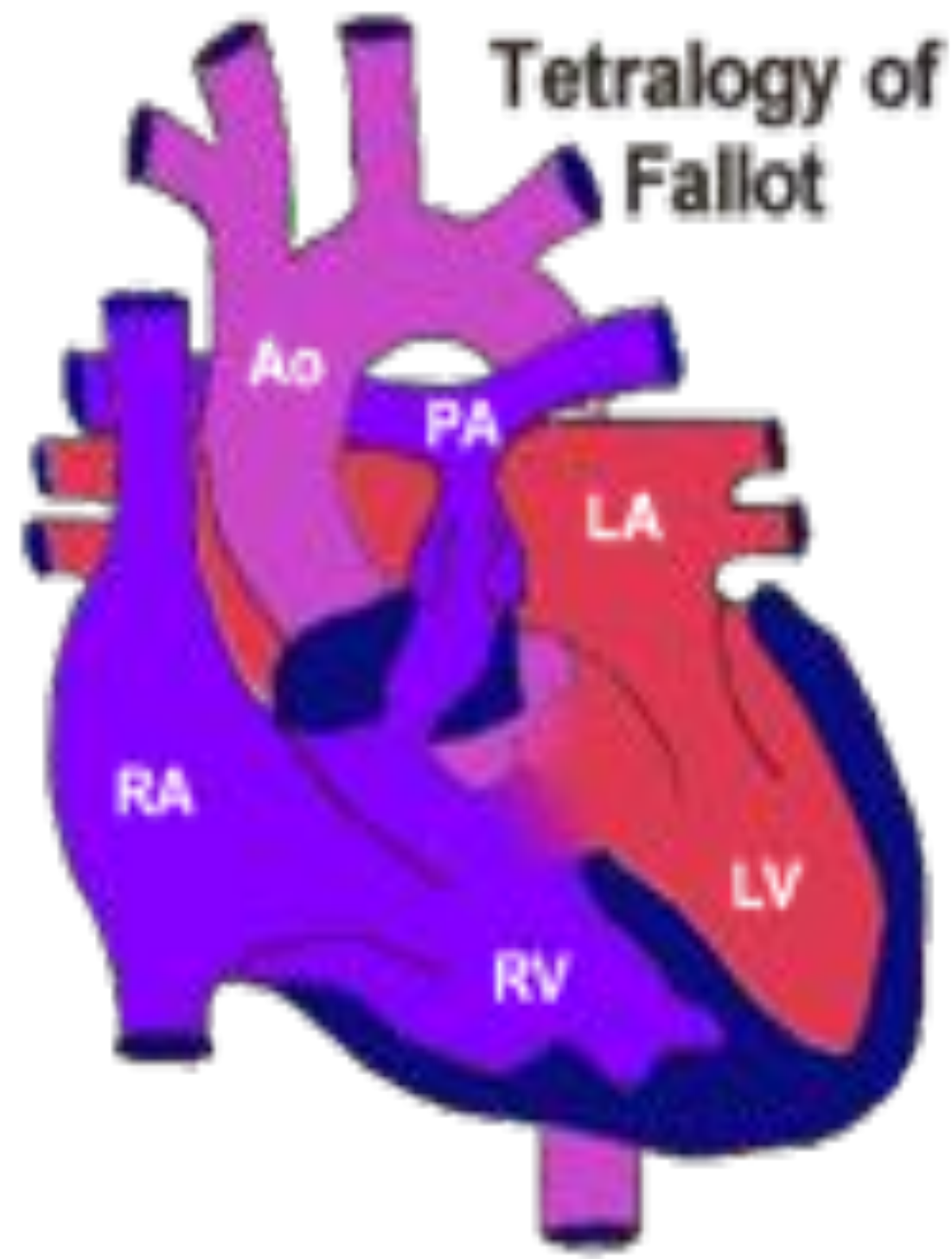
**Figure 3** Outcome of newborns after foetal diagnosis of tetralogy of Fallot in the first year of life.

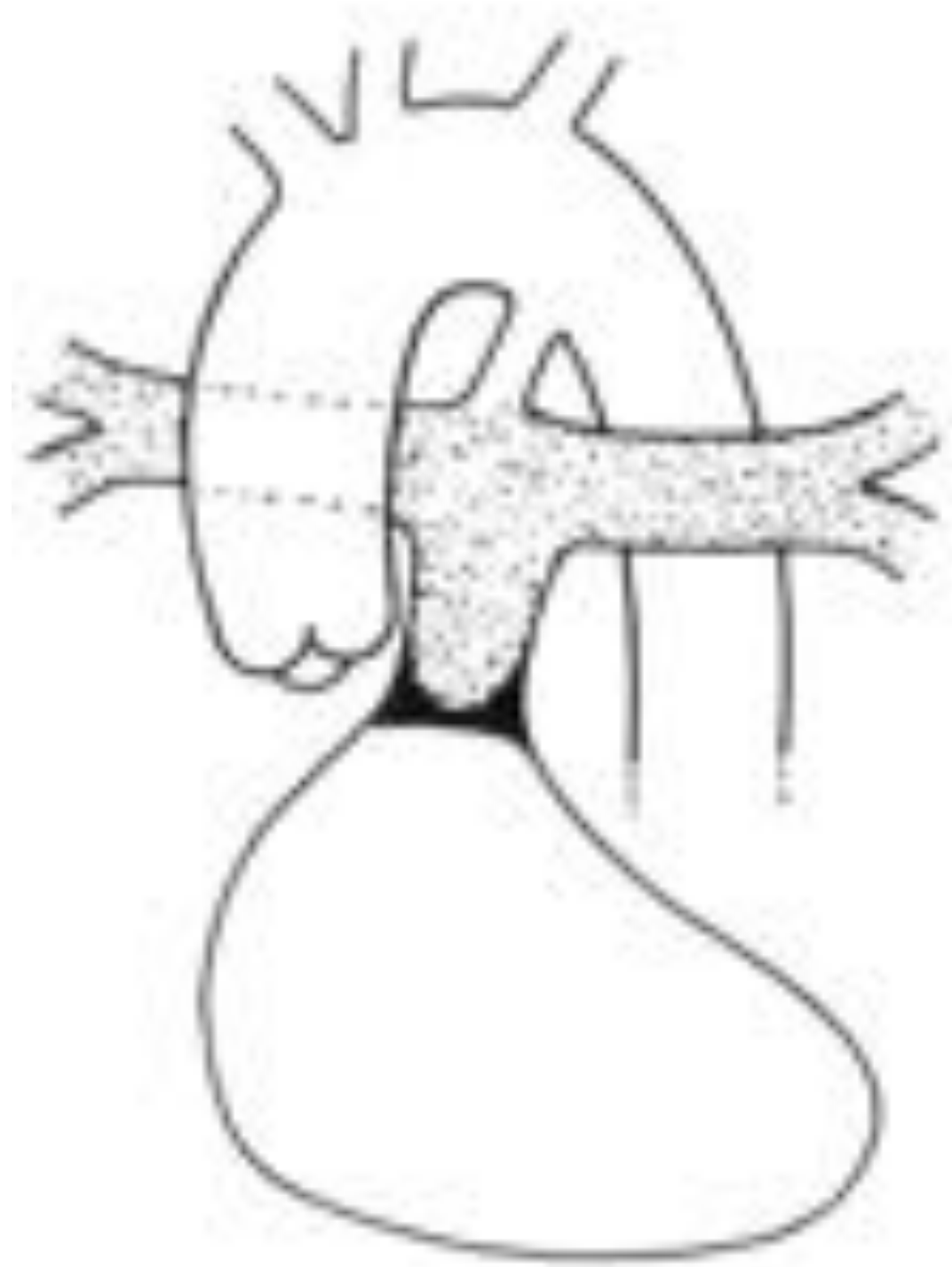


	PA branches small/absent	PA branches normal	p	Missing Data
Complete repair <1 an : number (%)	20/36 (55%)	→ 66/77 (86%)	<0,001	10
	Main PA absent	Main PA present	p	Missing Data
Complete repair <1 an : number (%)	1/6 (16%)	⇒ 90/114 (79%)	0,003	3
	PA branches and/or Main PA absent	PA branches and Main PA present	p	Missing Data
Complete repair <1 an : number (%)	1/6 (17%)	→ 84/104 (81%)	0,0022	13
	MAPCA (+)	MAPCA (-)	p	Missing Data
Complete repair <1 an : number (%)	3/6 (50%)	89/117 (76%)	0,17	0

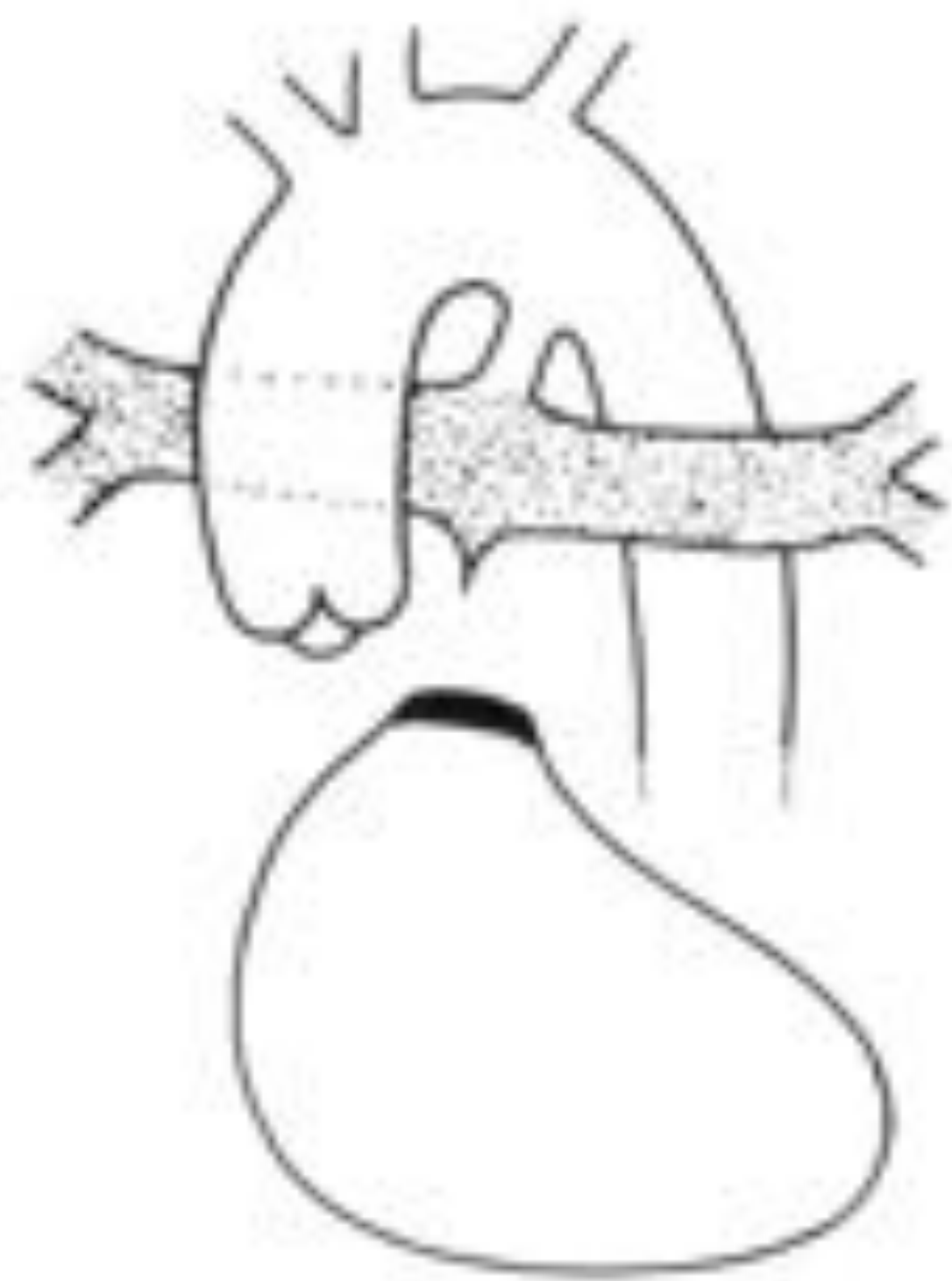
# Perinatal management

1. Ducto-dependent defect ?

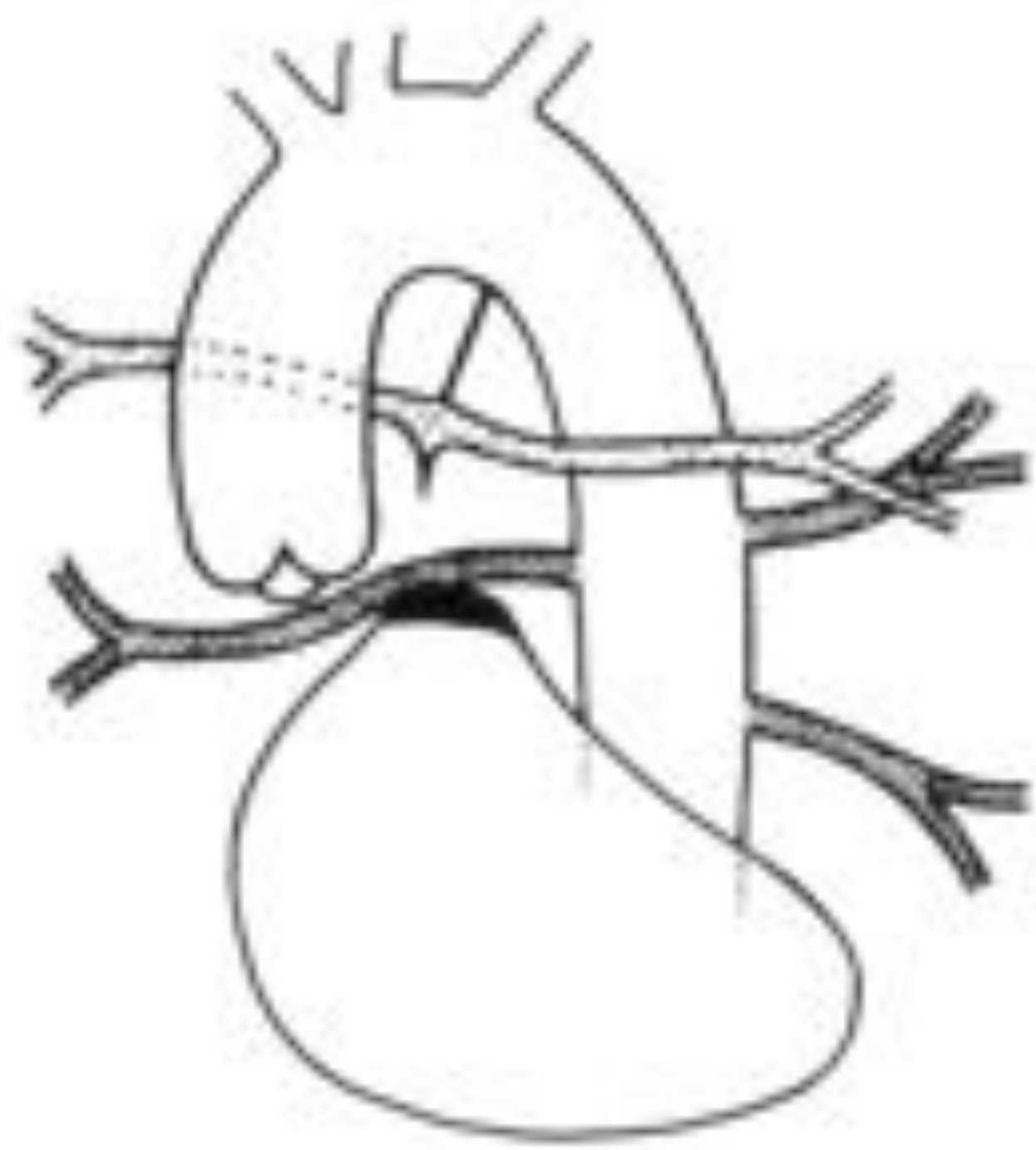




I



II



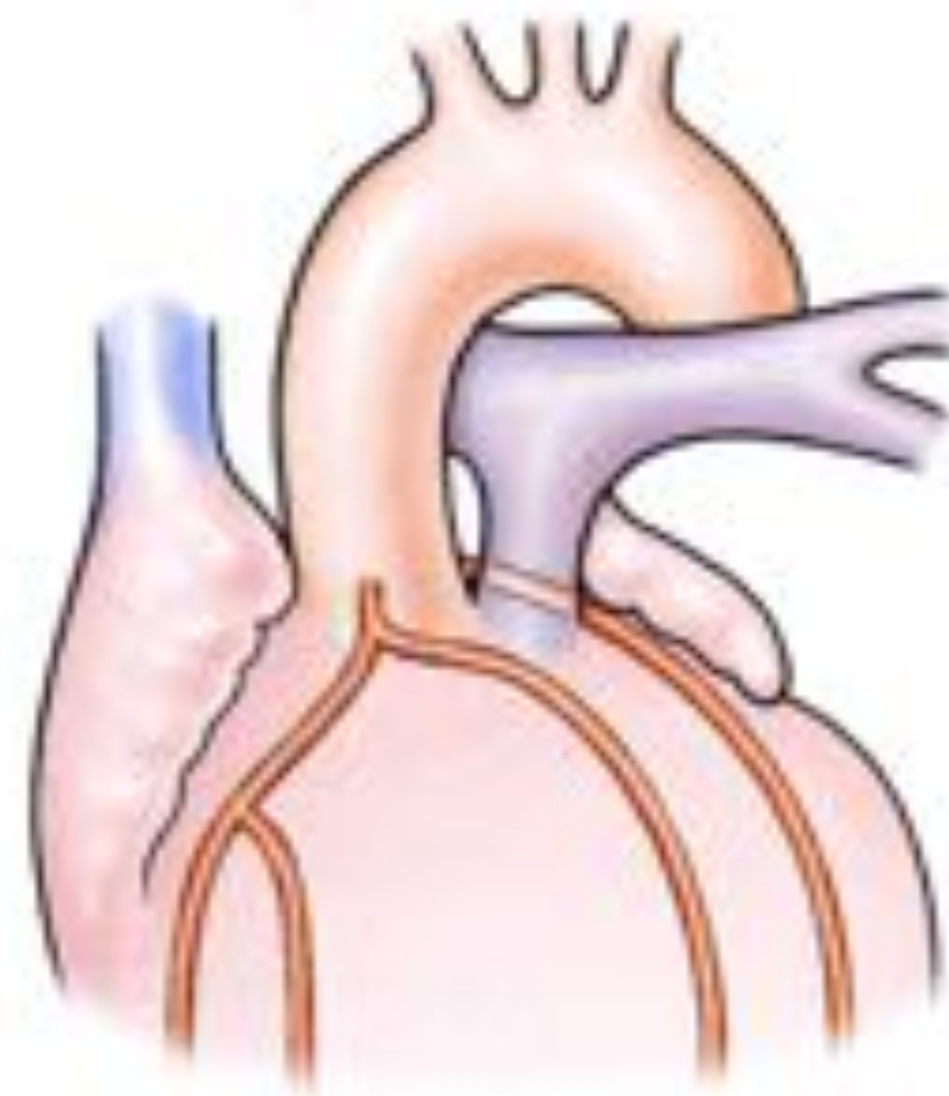
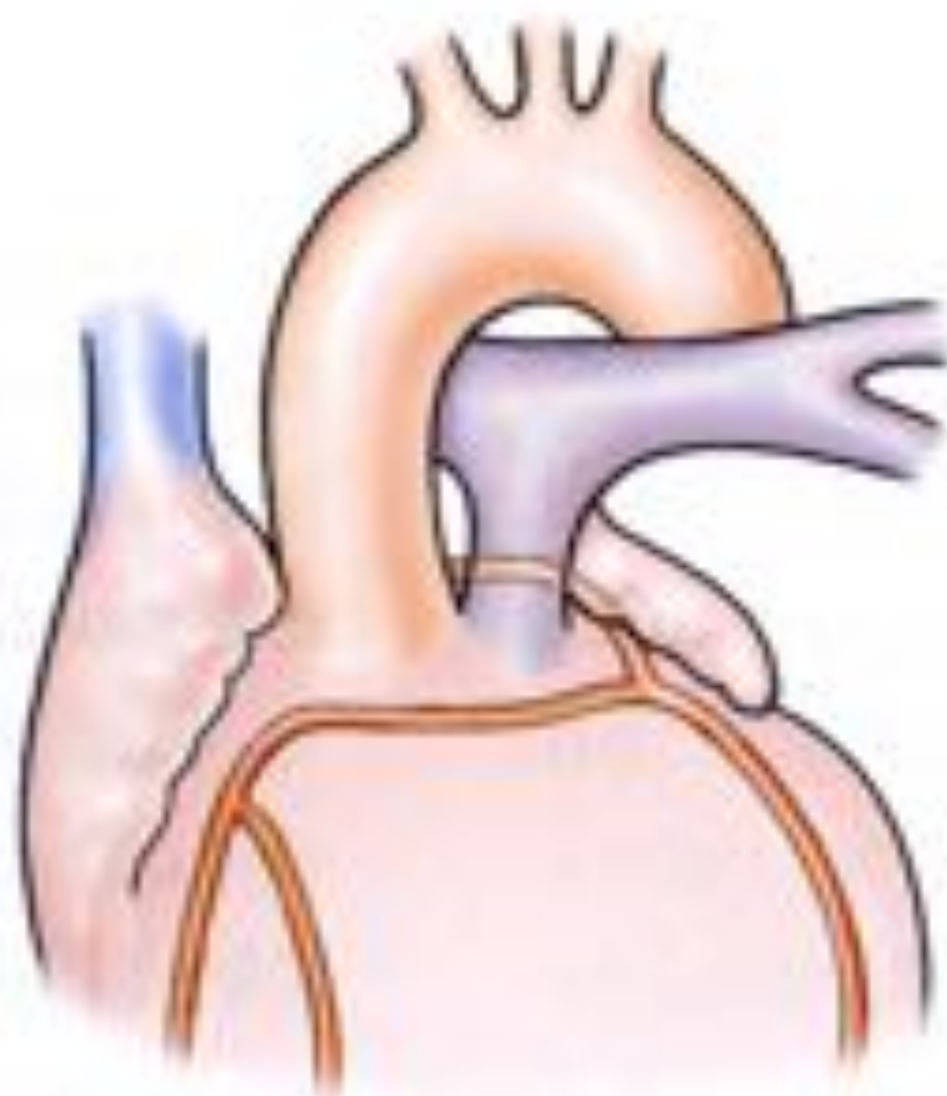
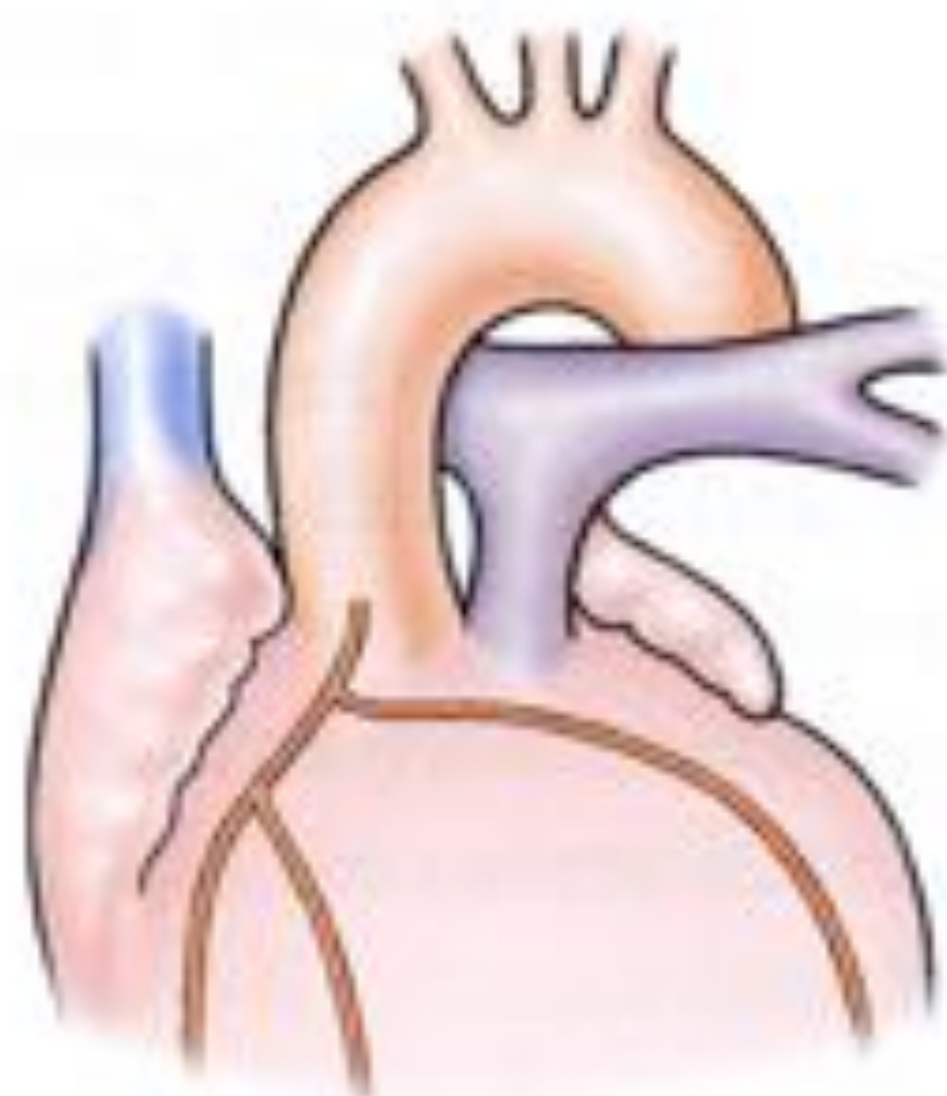
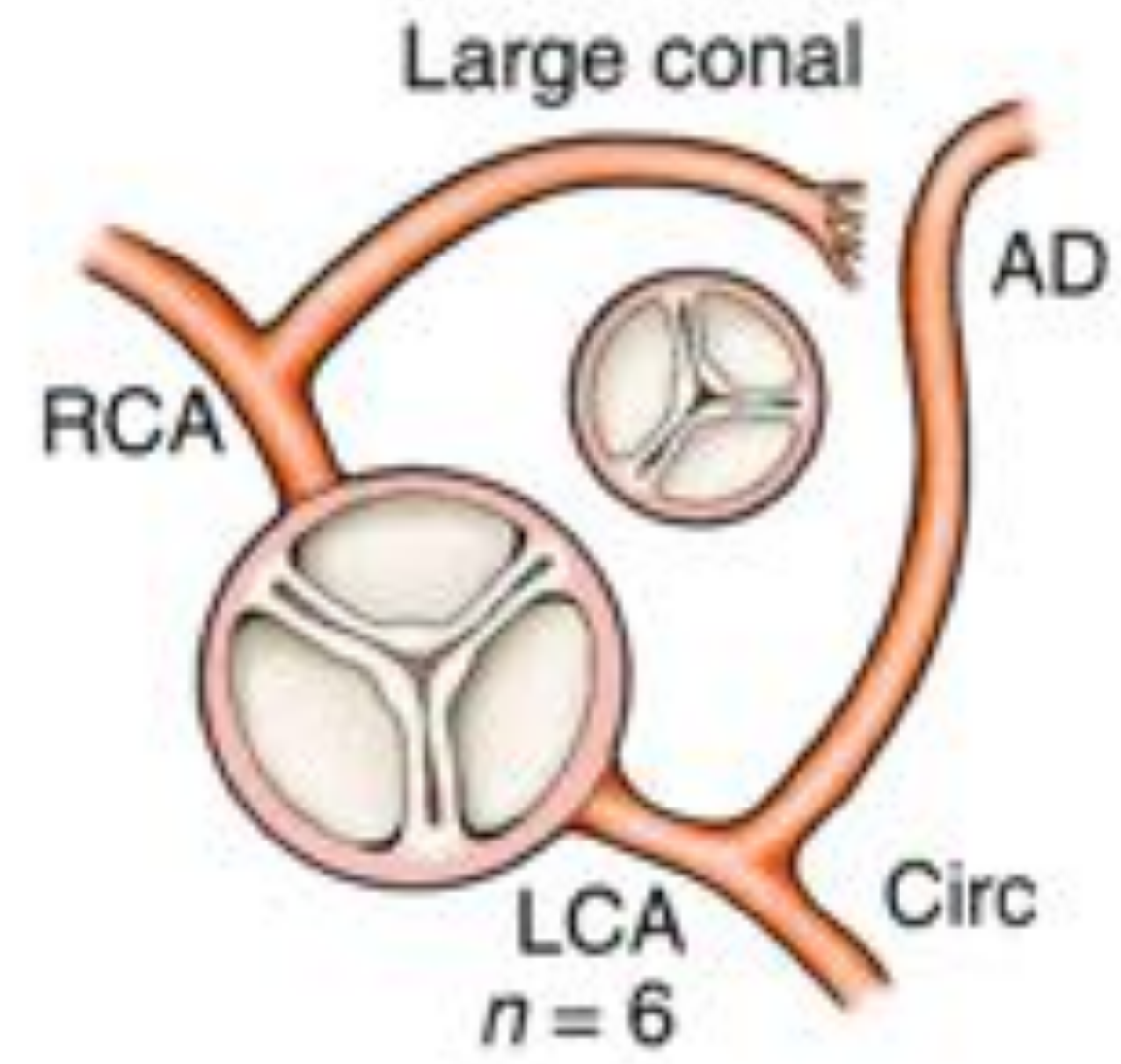
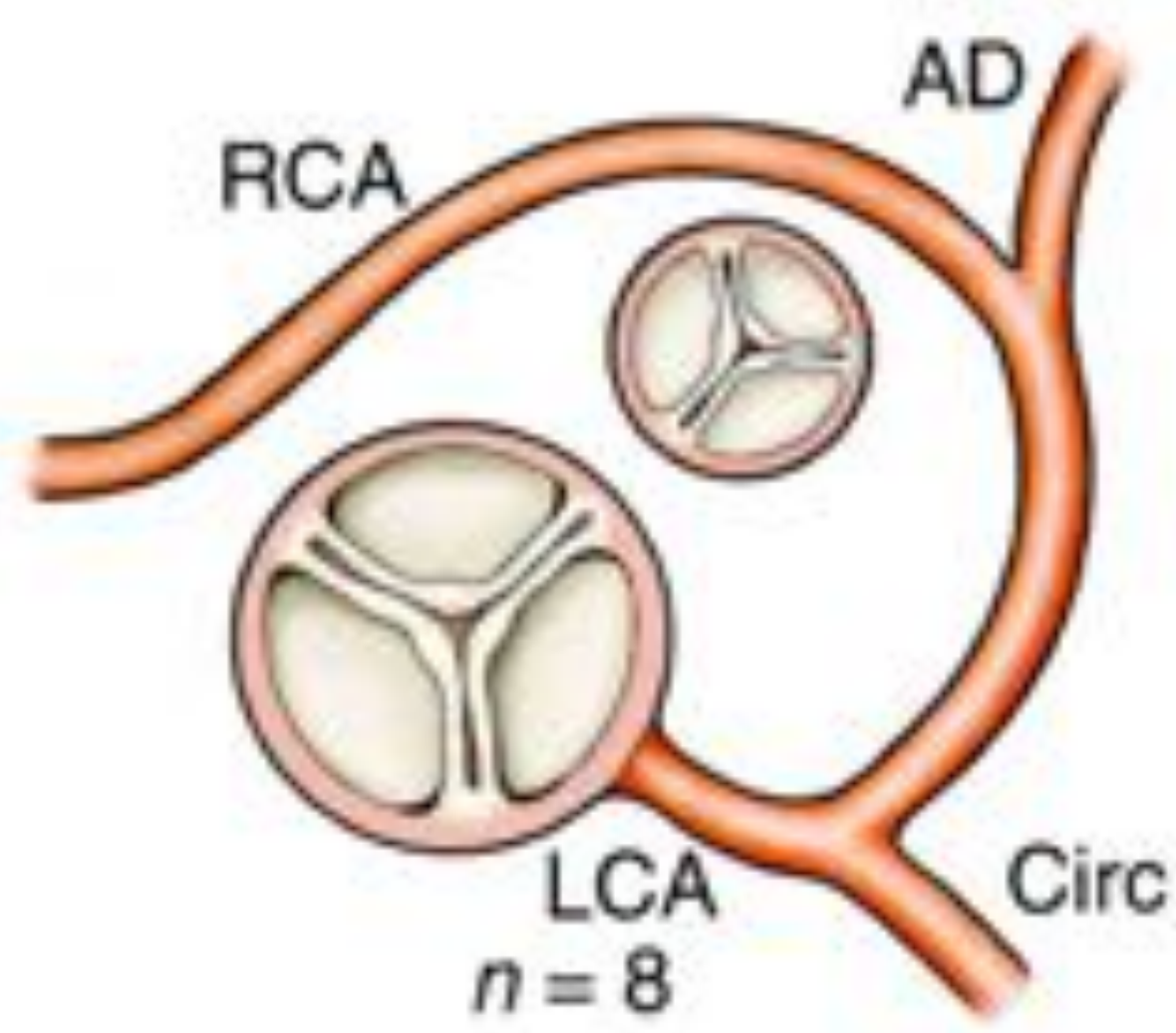
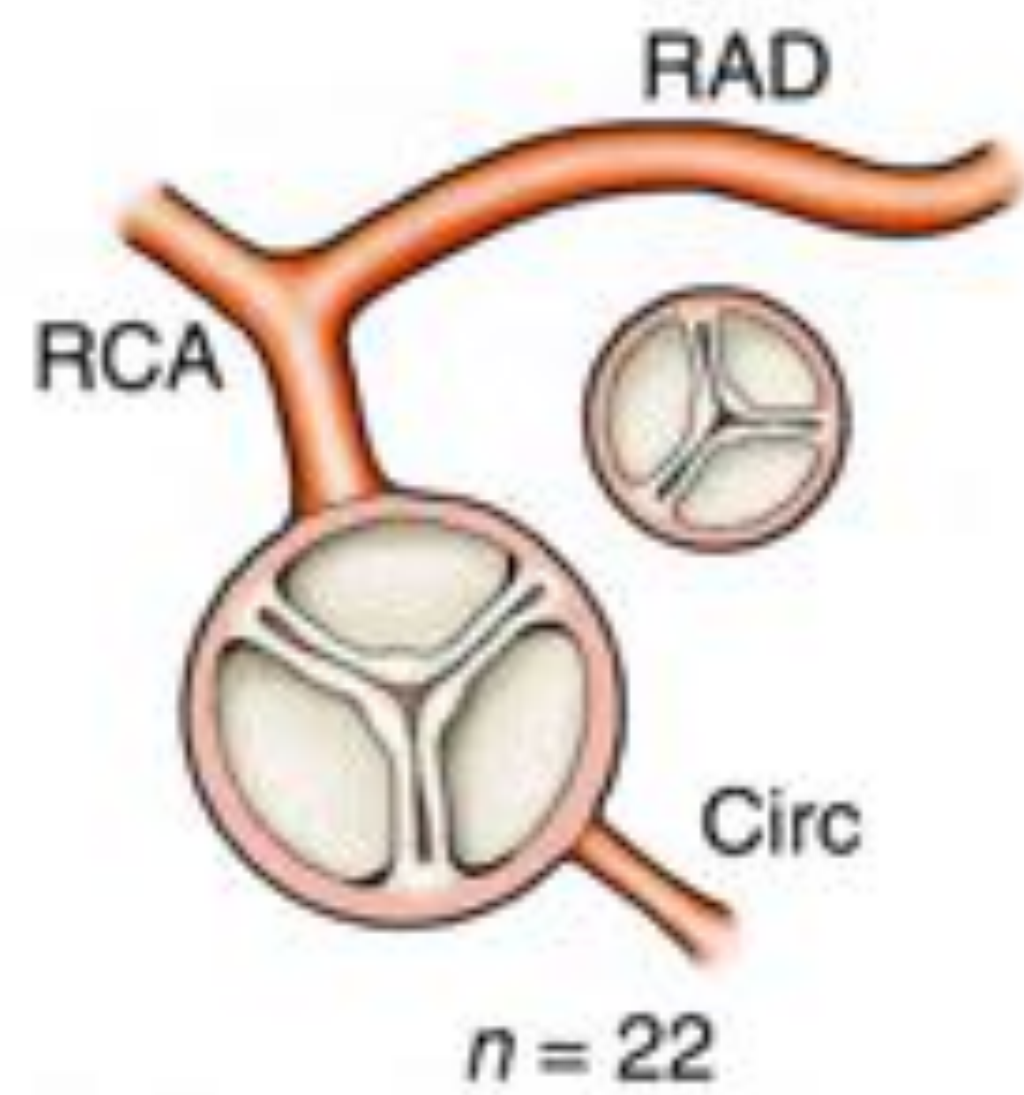
III



IV

# Perinatal management

1. Ducto-dependent defect ?
2. Associated cardiac anomalies





# Multiple VSDs

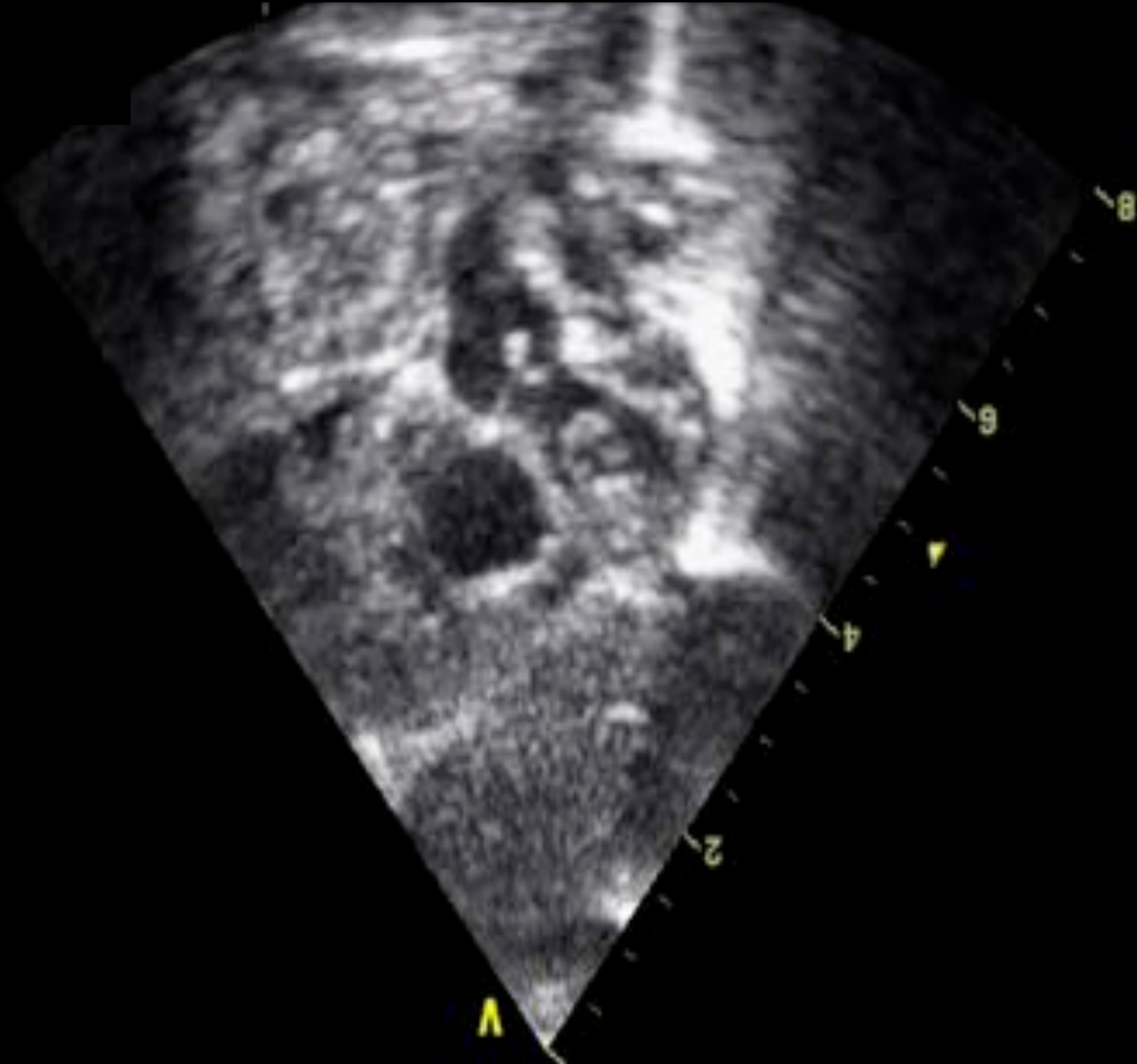
# Pulmonary branches stenosis

DFOV 17.4cm  
STNDV+

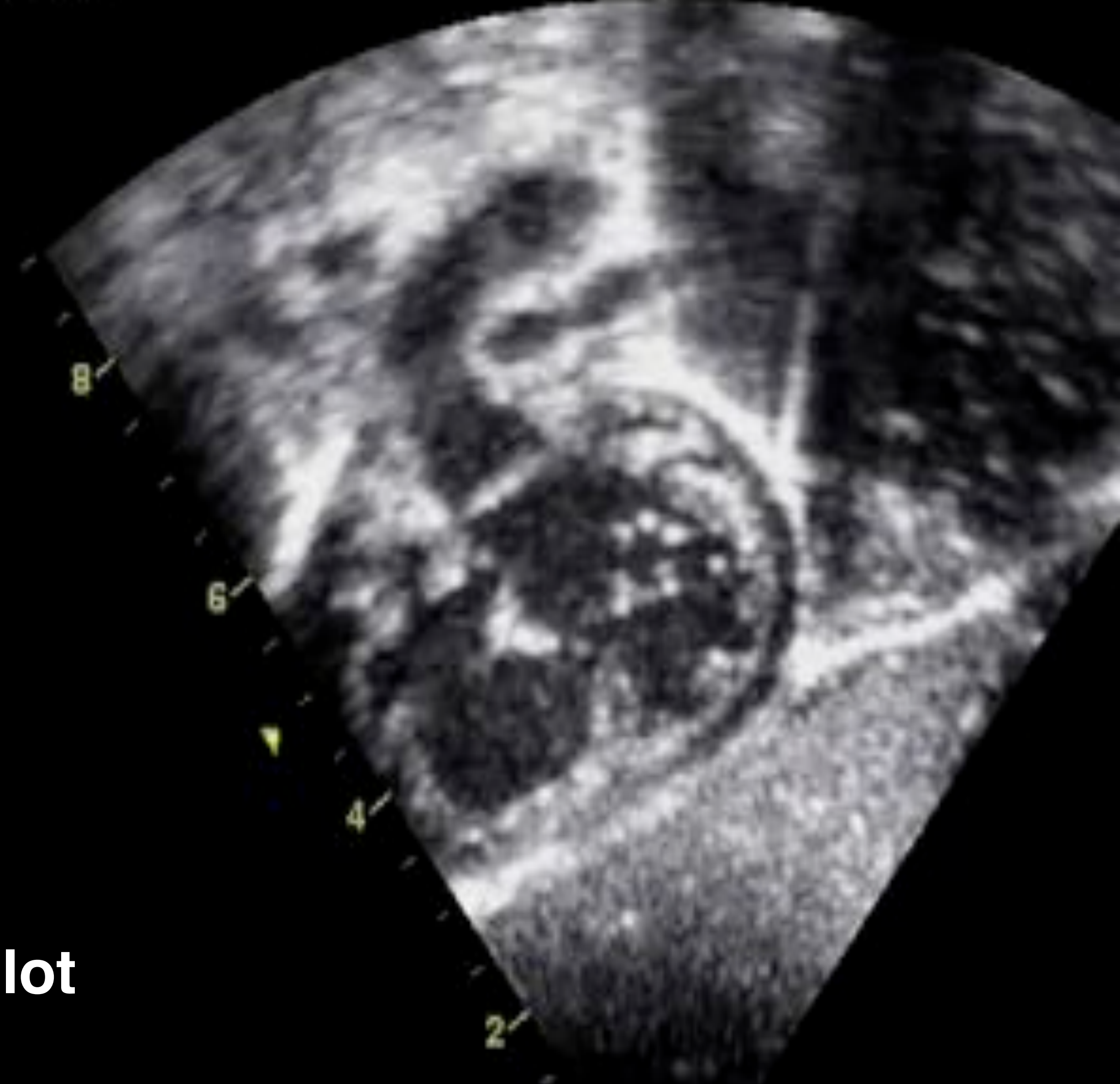
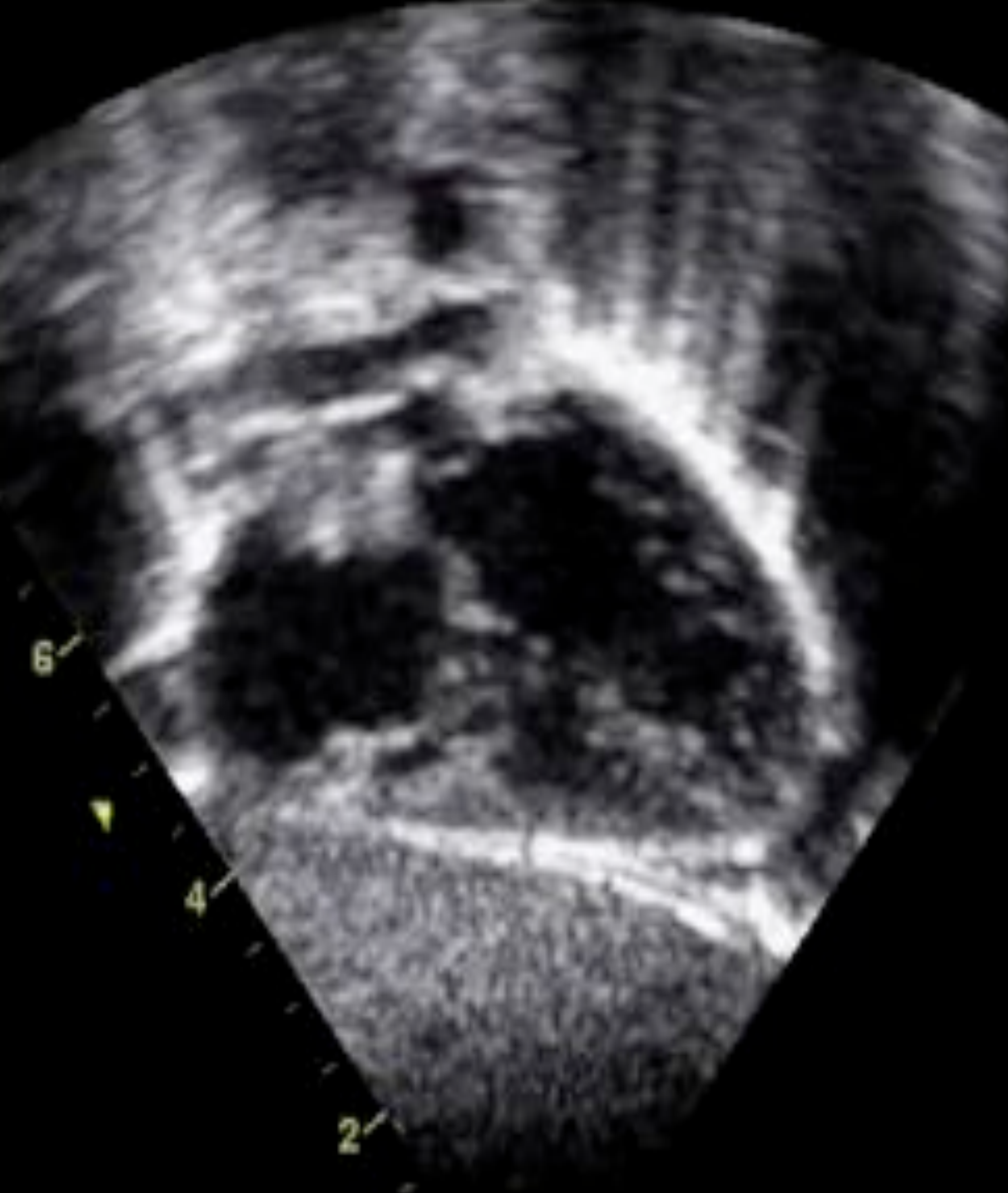
RAS



No VCI  
kv 120  
mA 149  
Rot 0.50s/HE+ 39.4mm/hot  
1.2mm @ 904.1 / 0.9sp  
TR: 0.0  
11:20:45 AM  
W = 4095 L = 2048

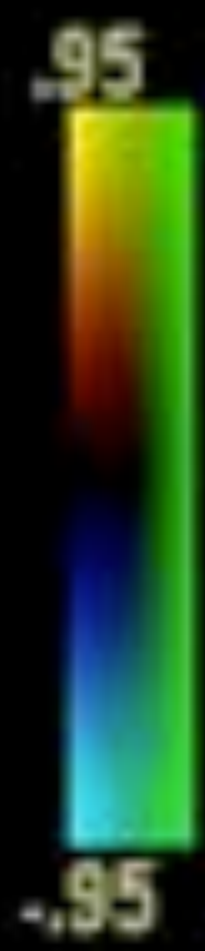
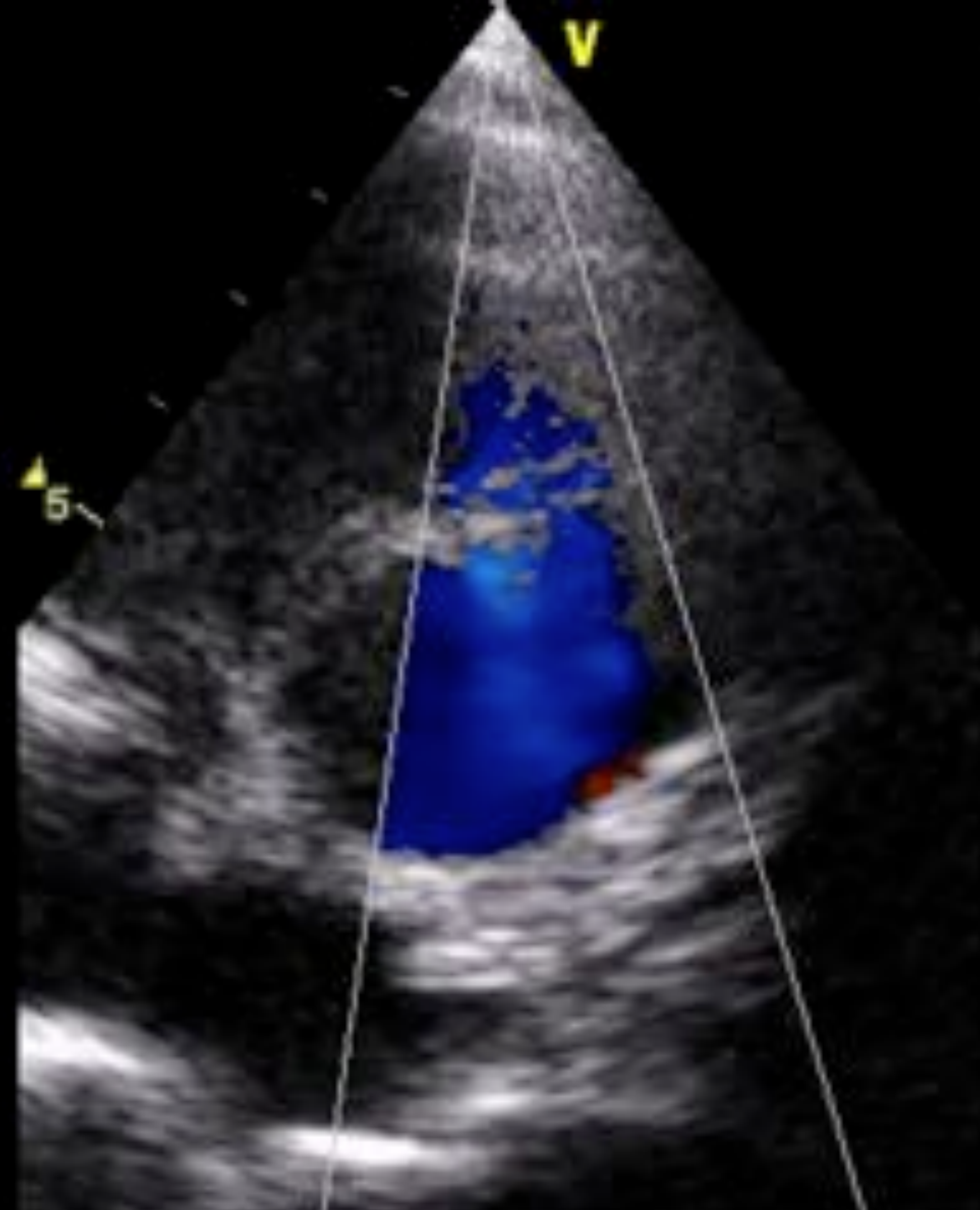






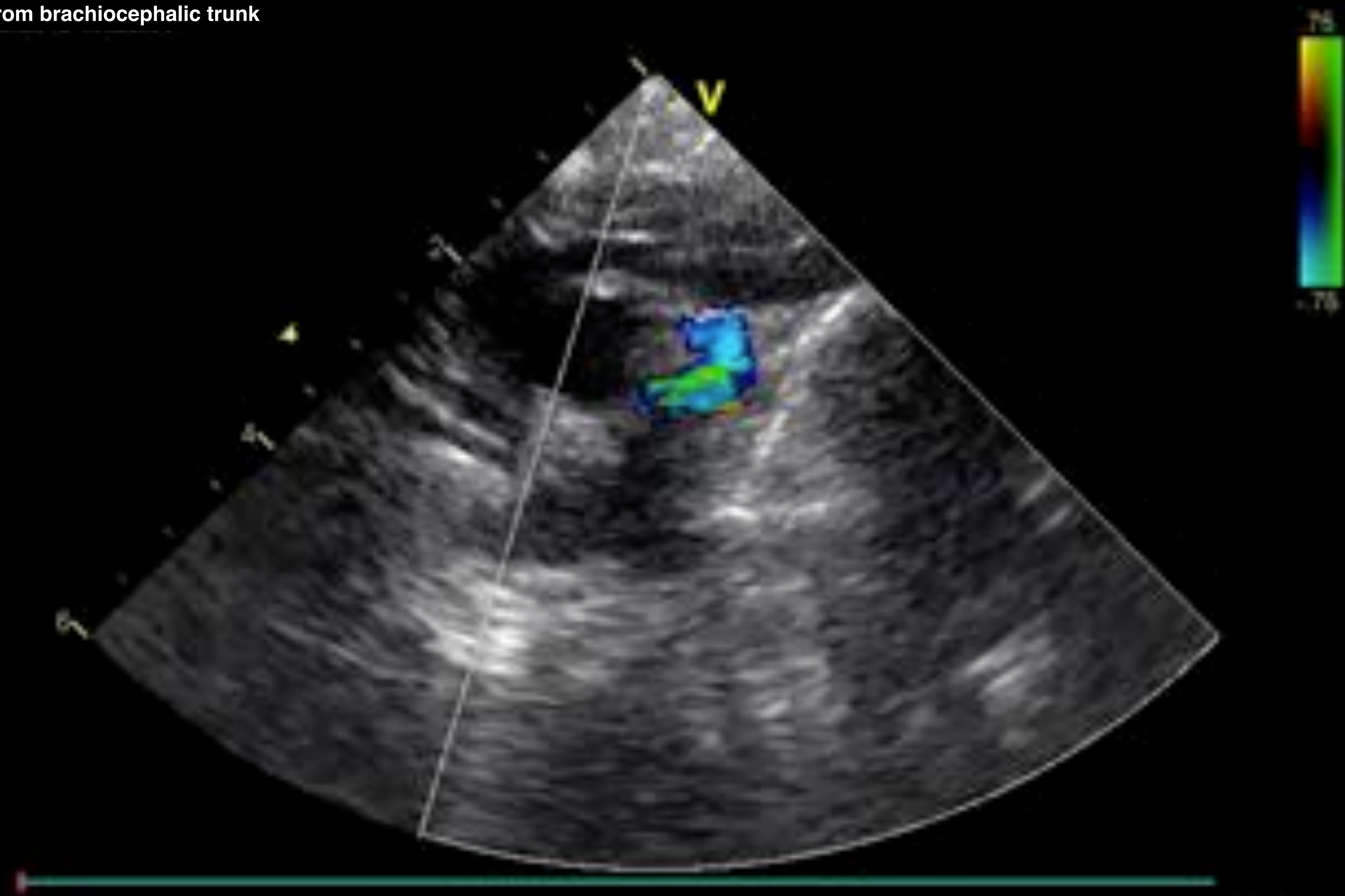
**AVSD Fallot**

18/11/2008 11:41:23



**Outflow VSD without conal septum**

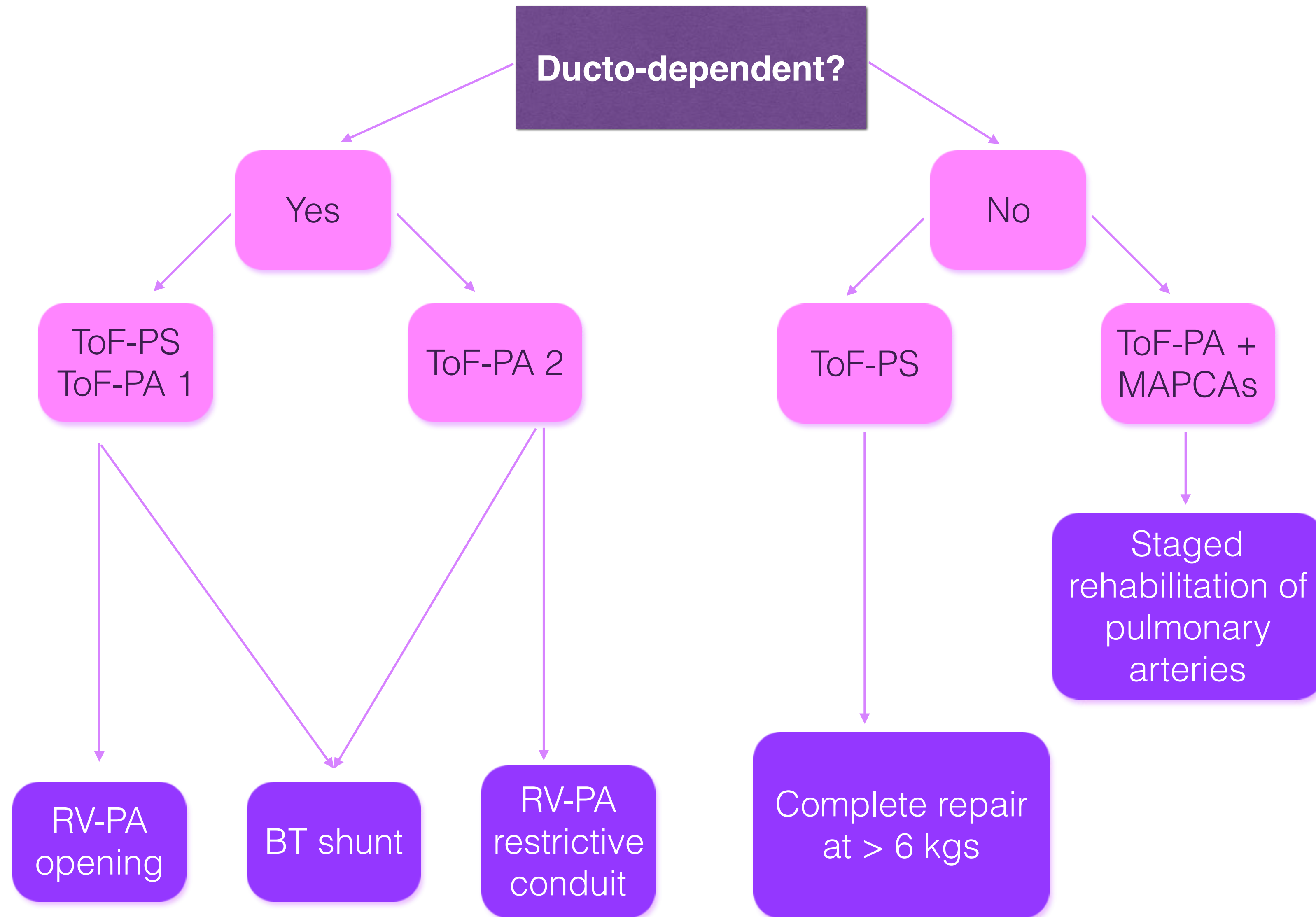
Patent arterial duct from brachiocephalic trunk

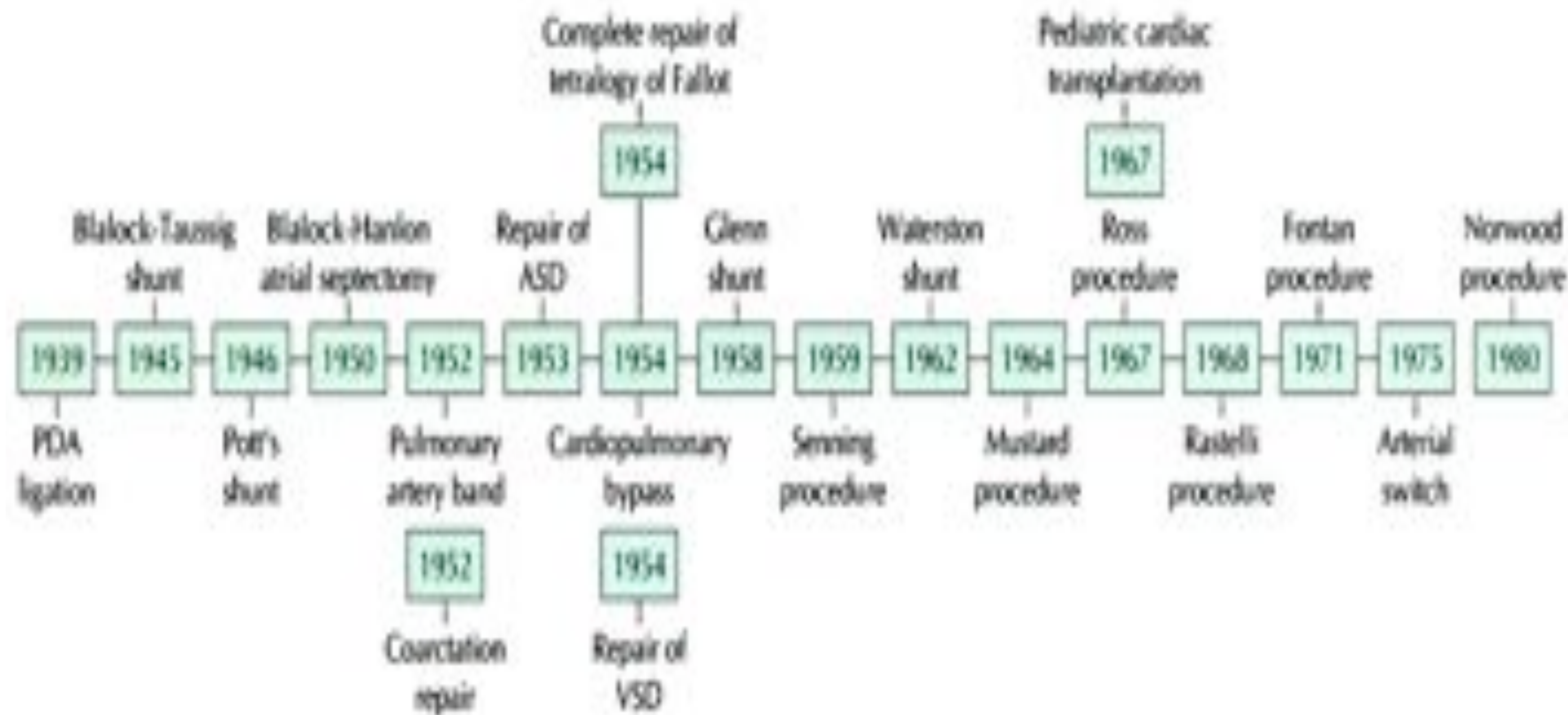


# Perinatal management

1. Ducto-dependent defect ?
2. Associated cardiac anomalies
3. Palliative surgery

# Palliative surgery







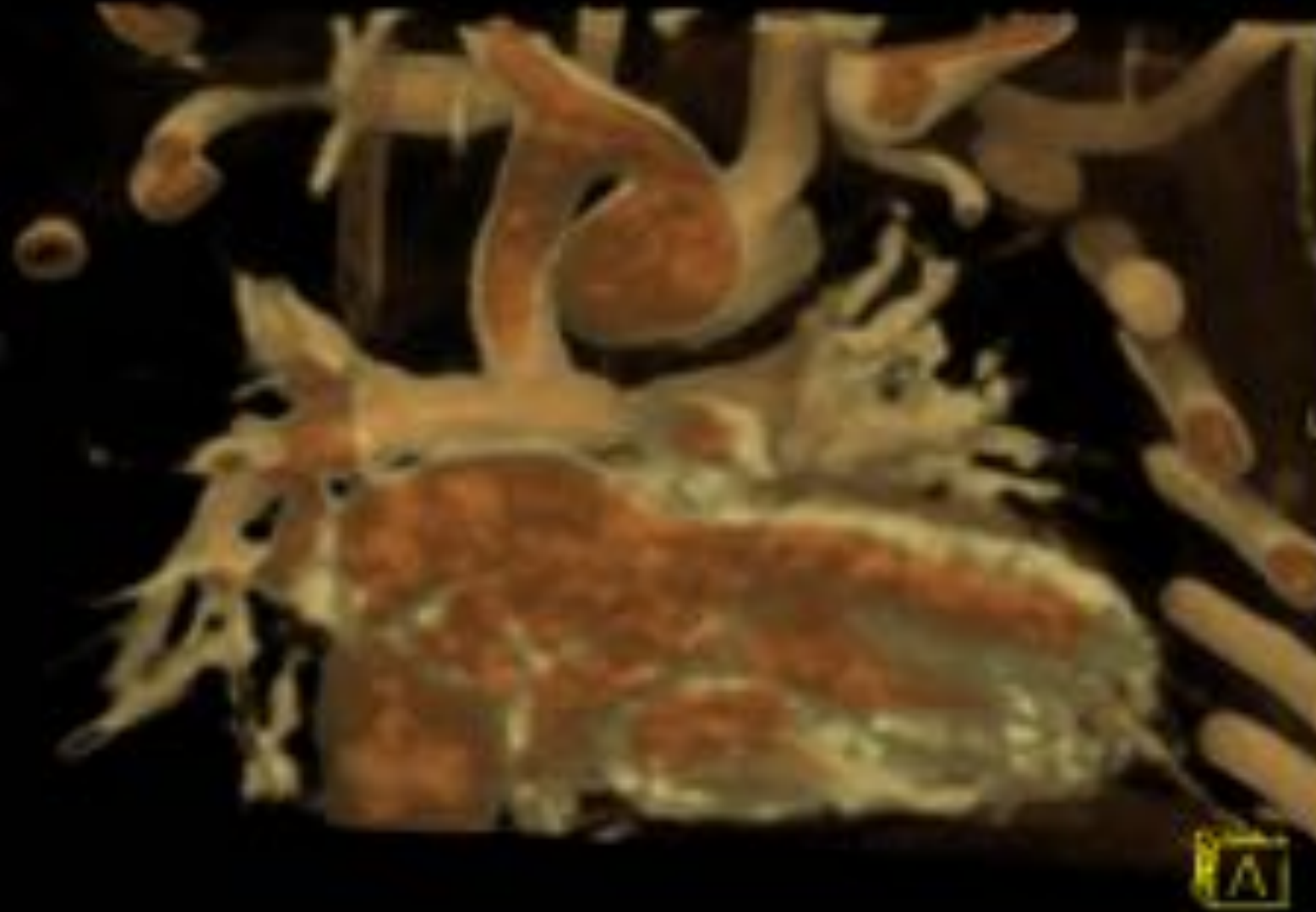
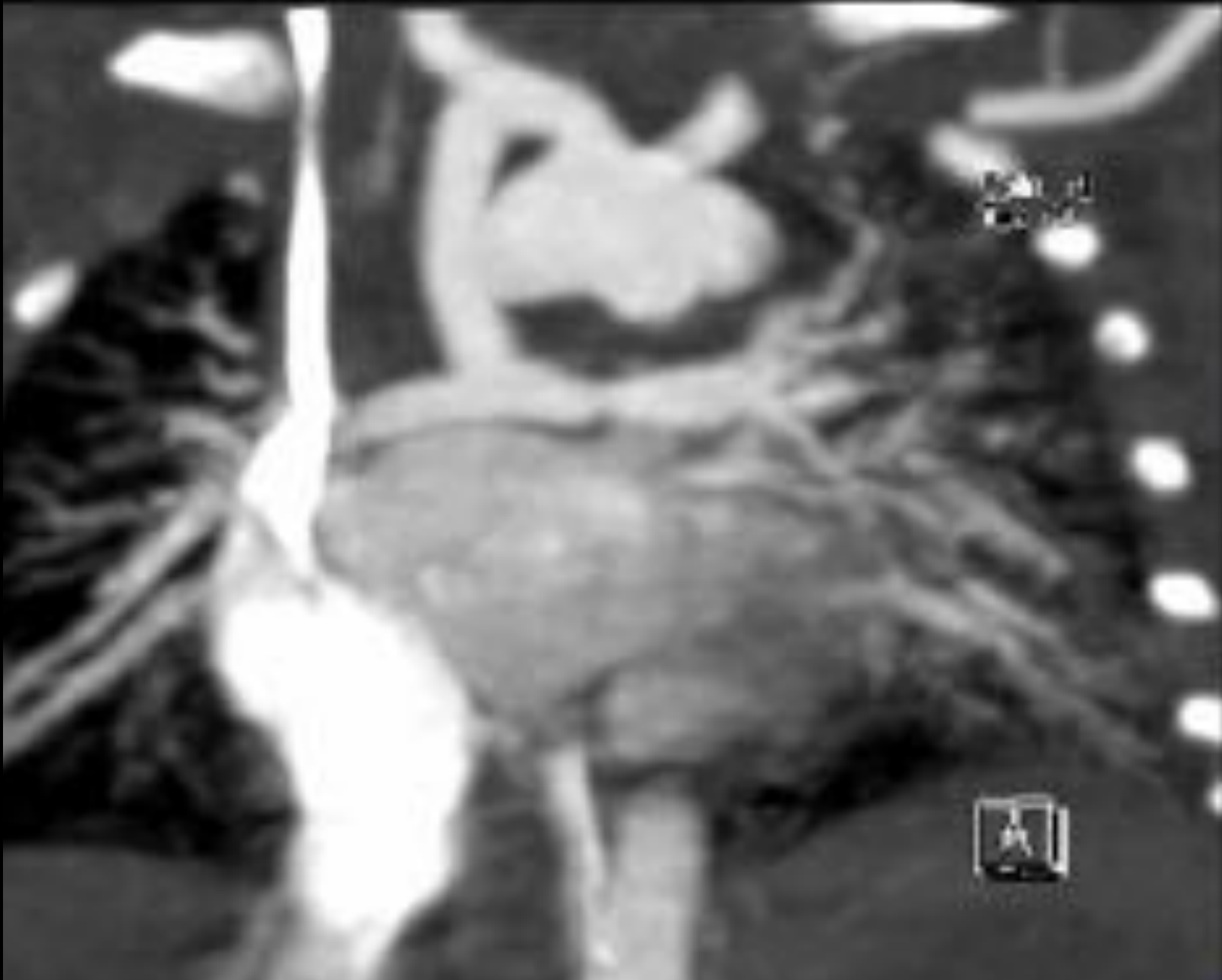
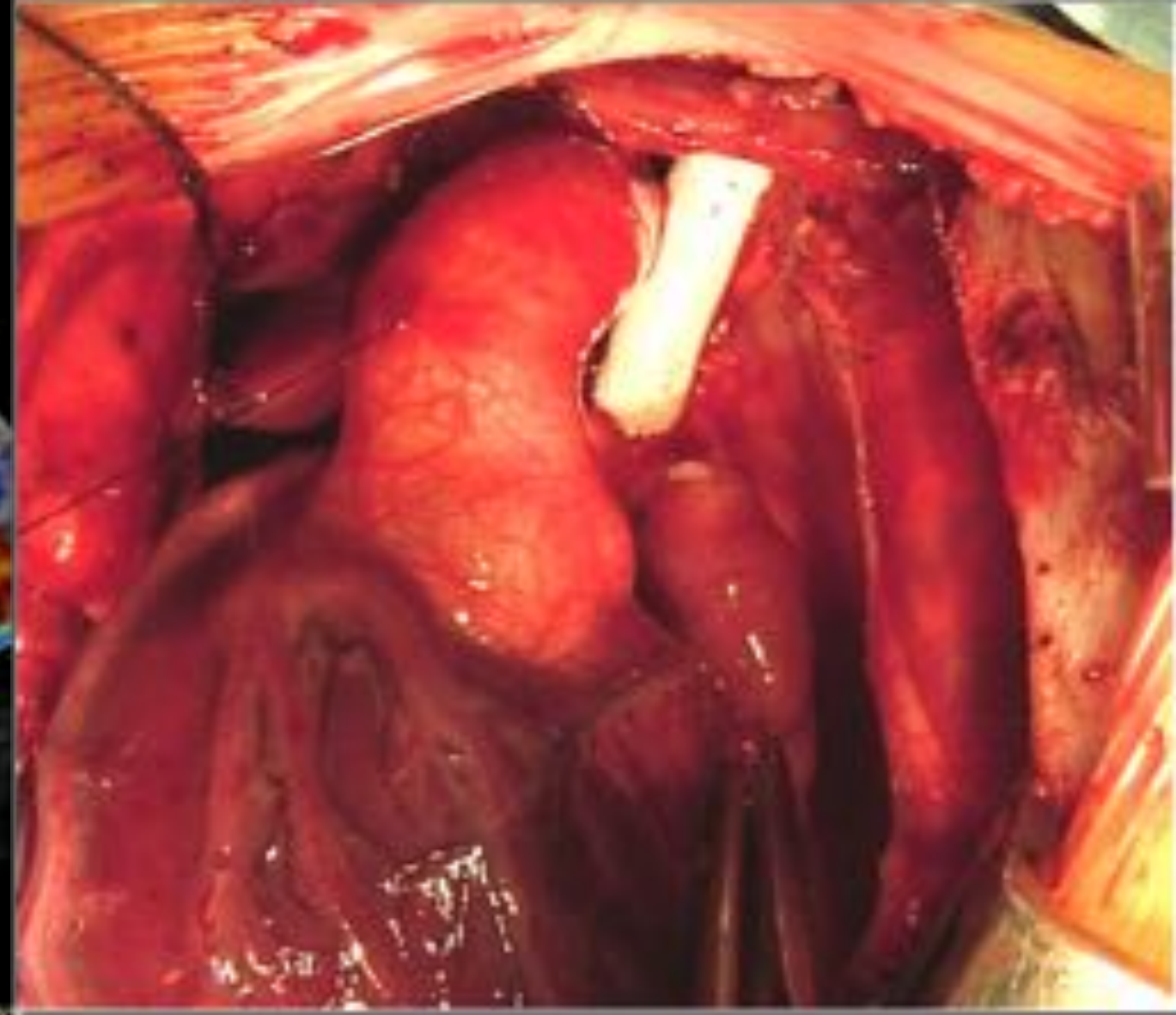
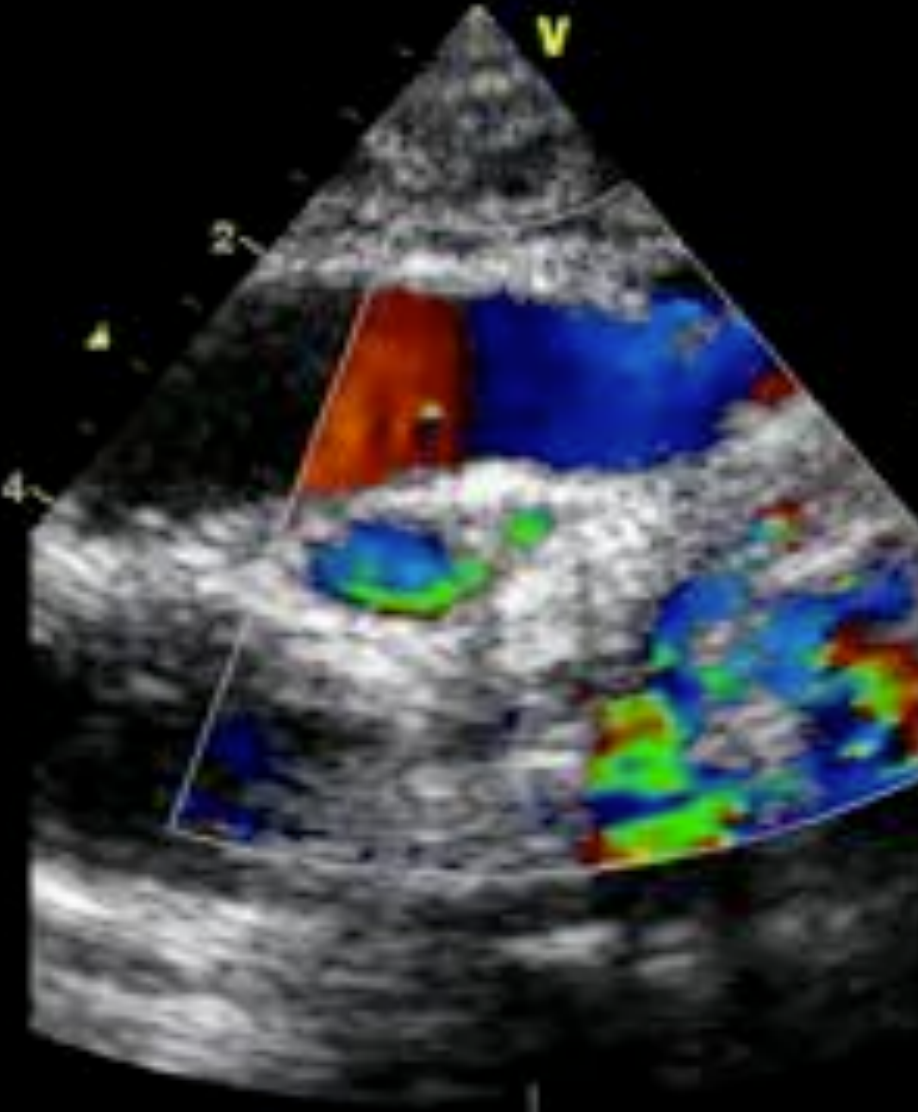
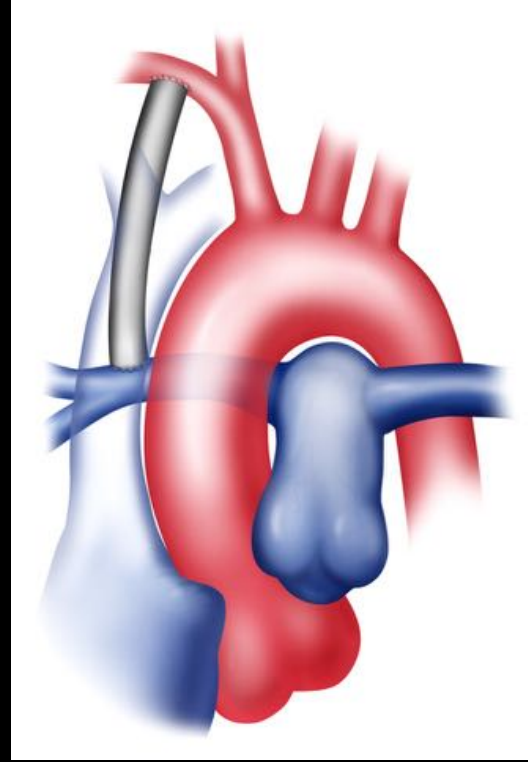
**Helen Taussig**



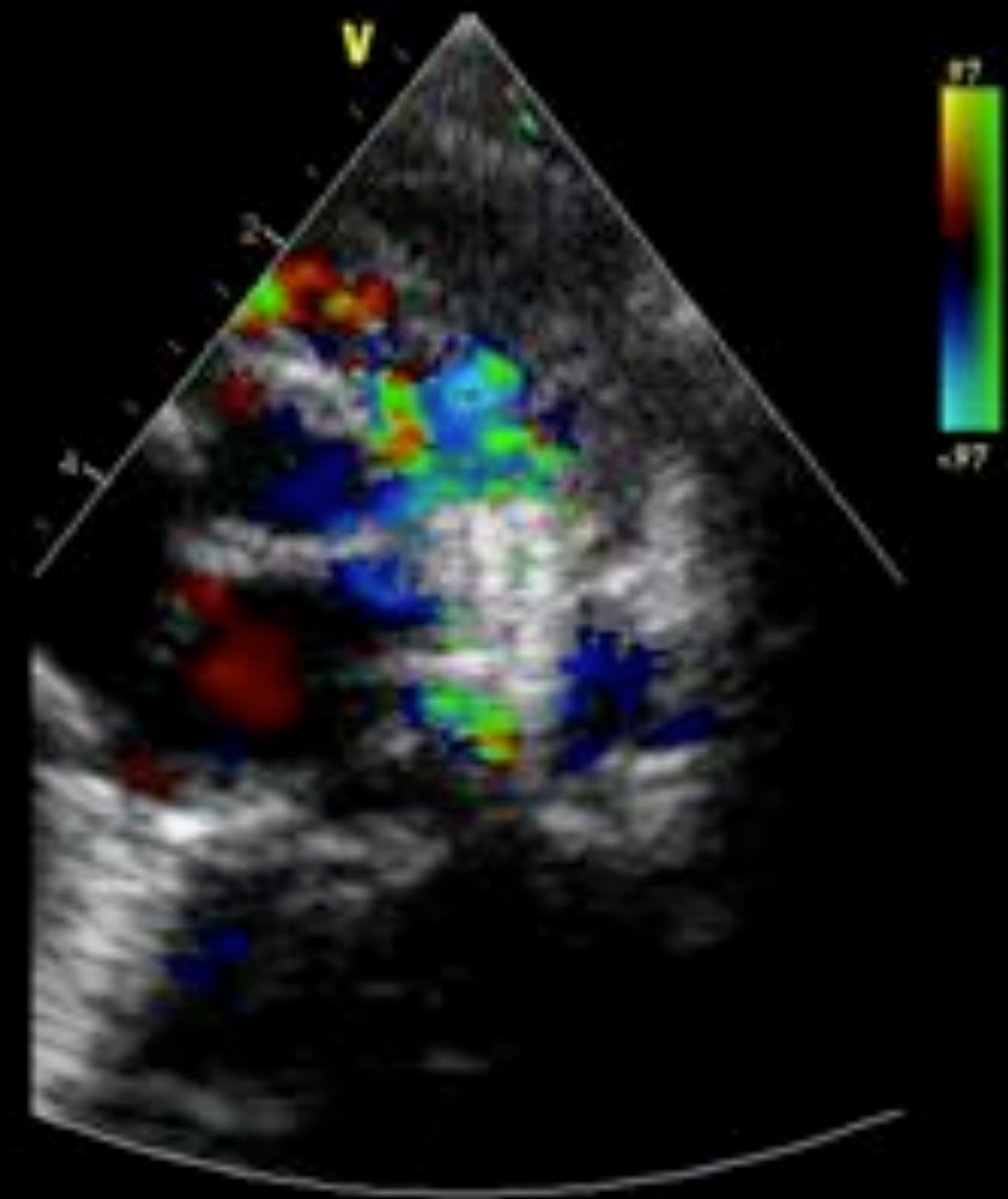
**Alfred Blalock and Eileen Saxon**

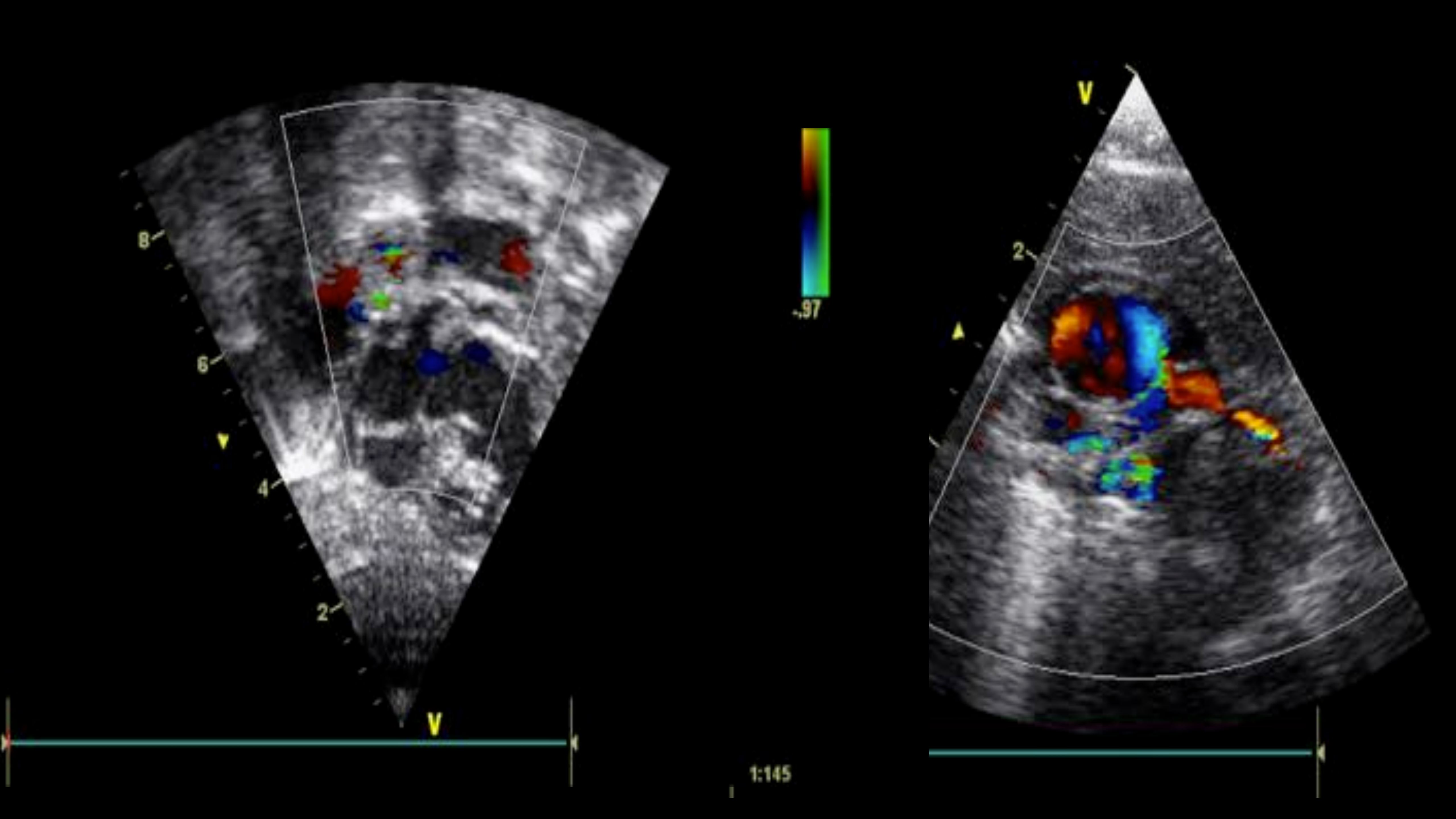


**Vivien Thomas**

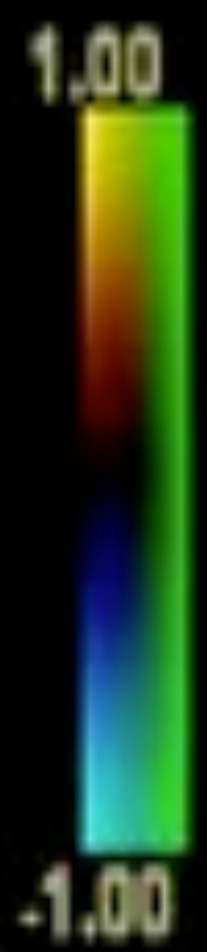




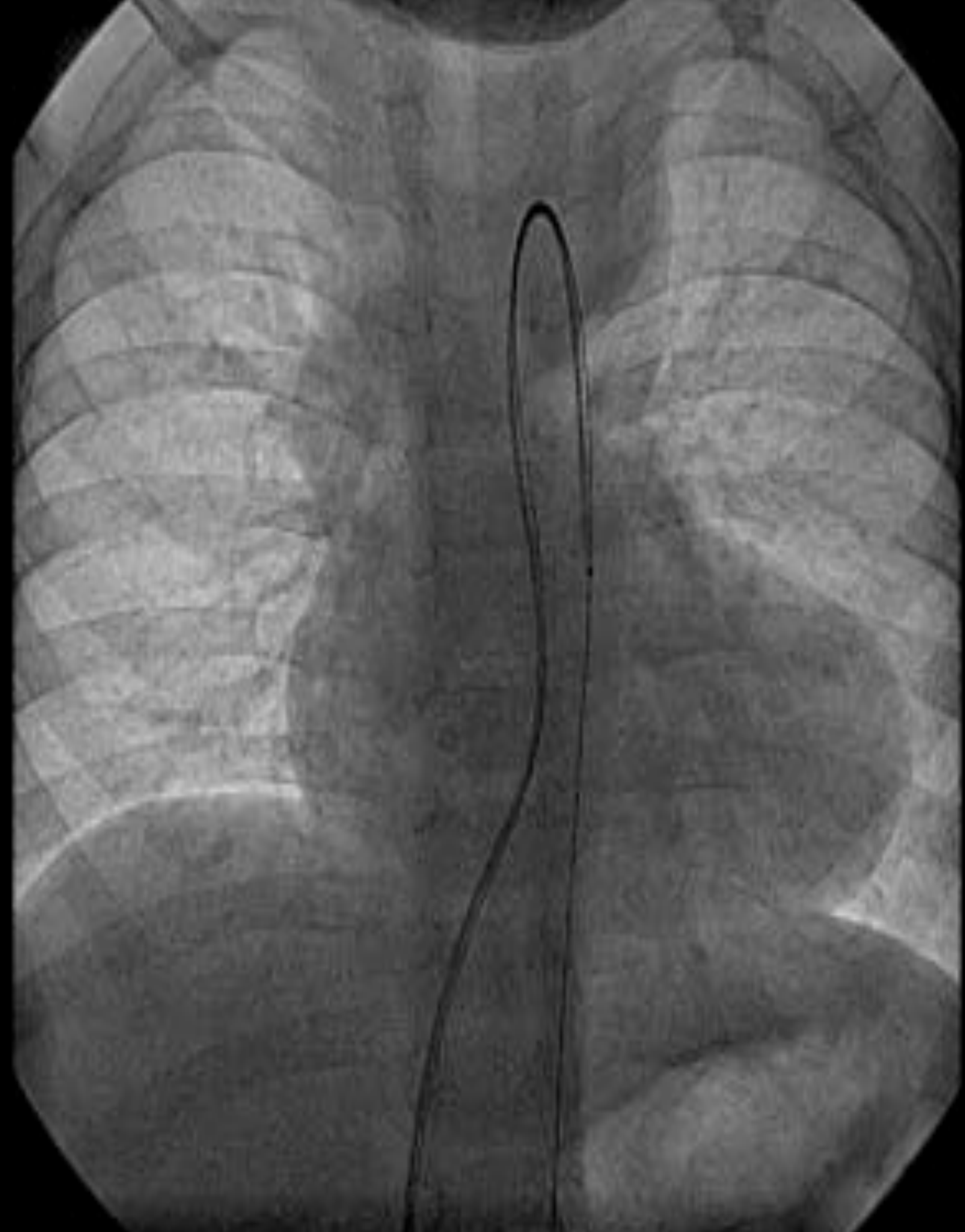




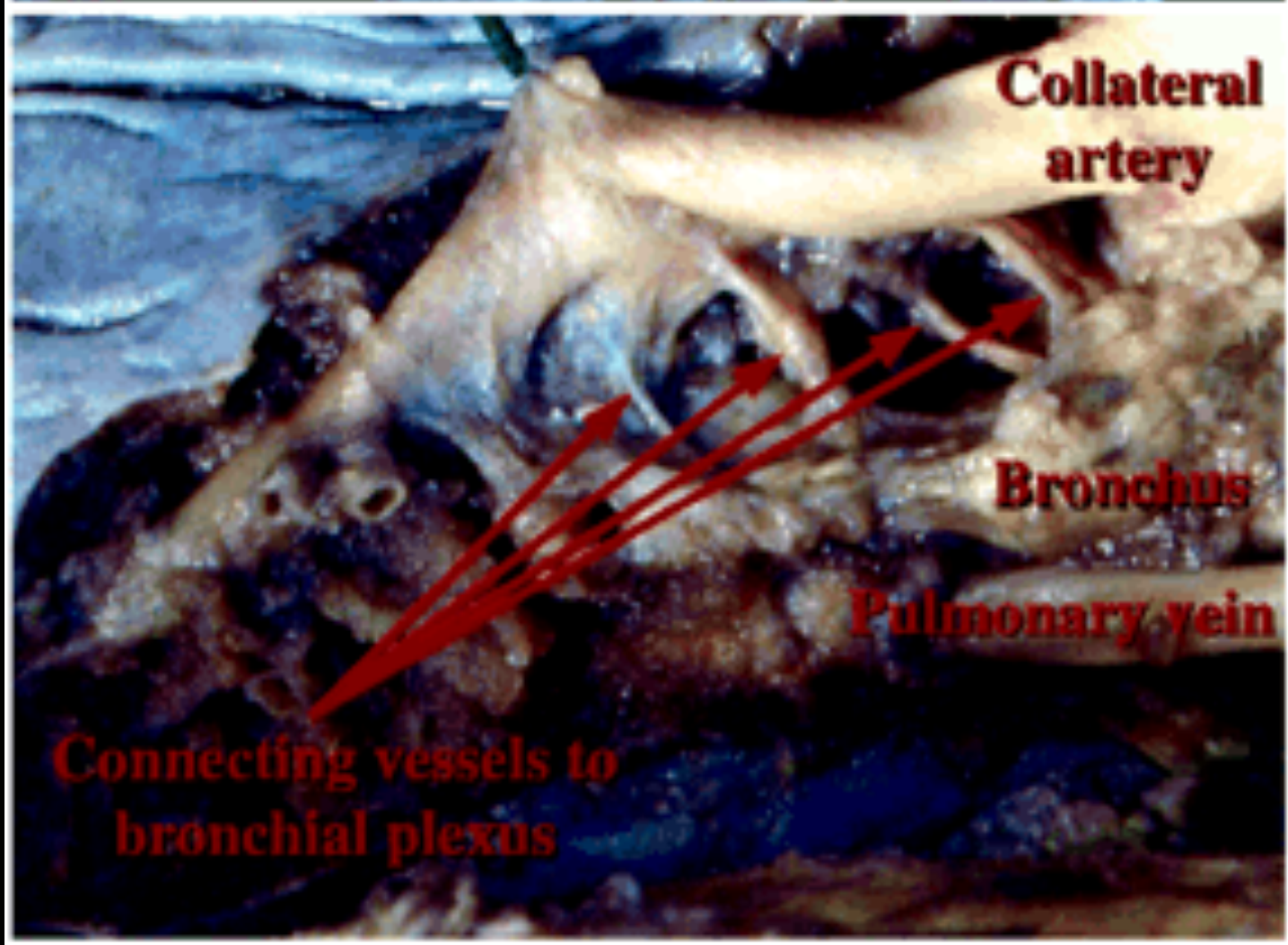
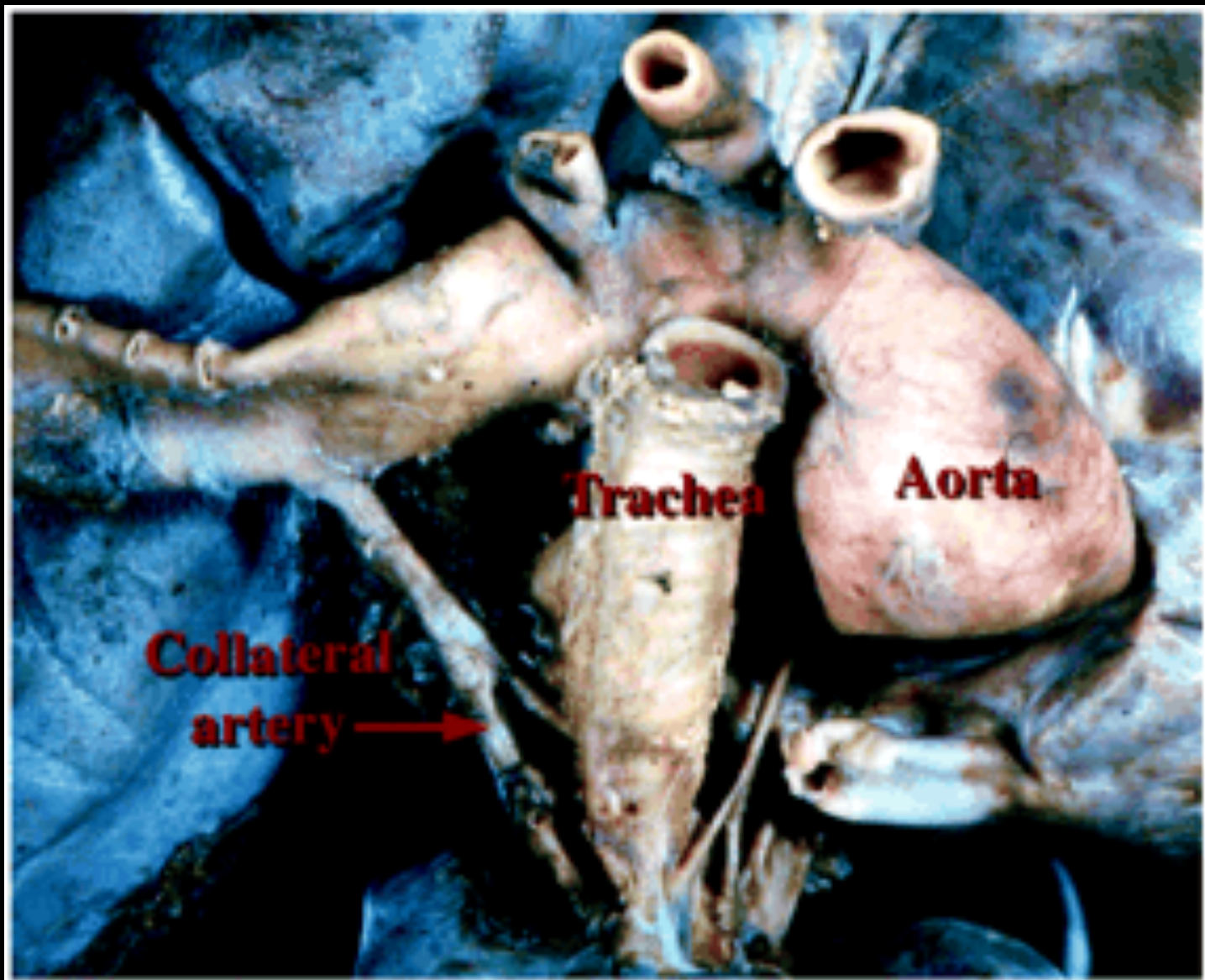
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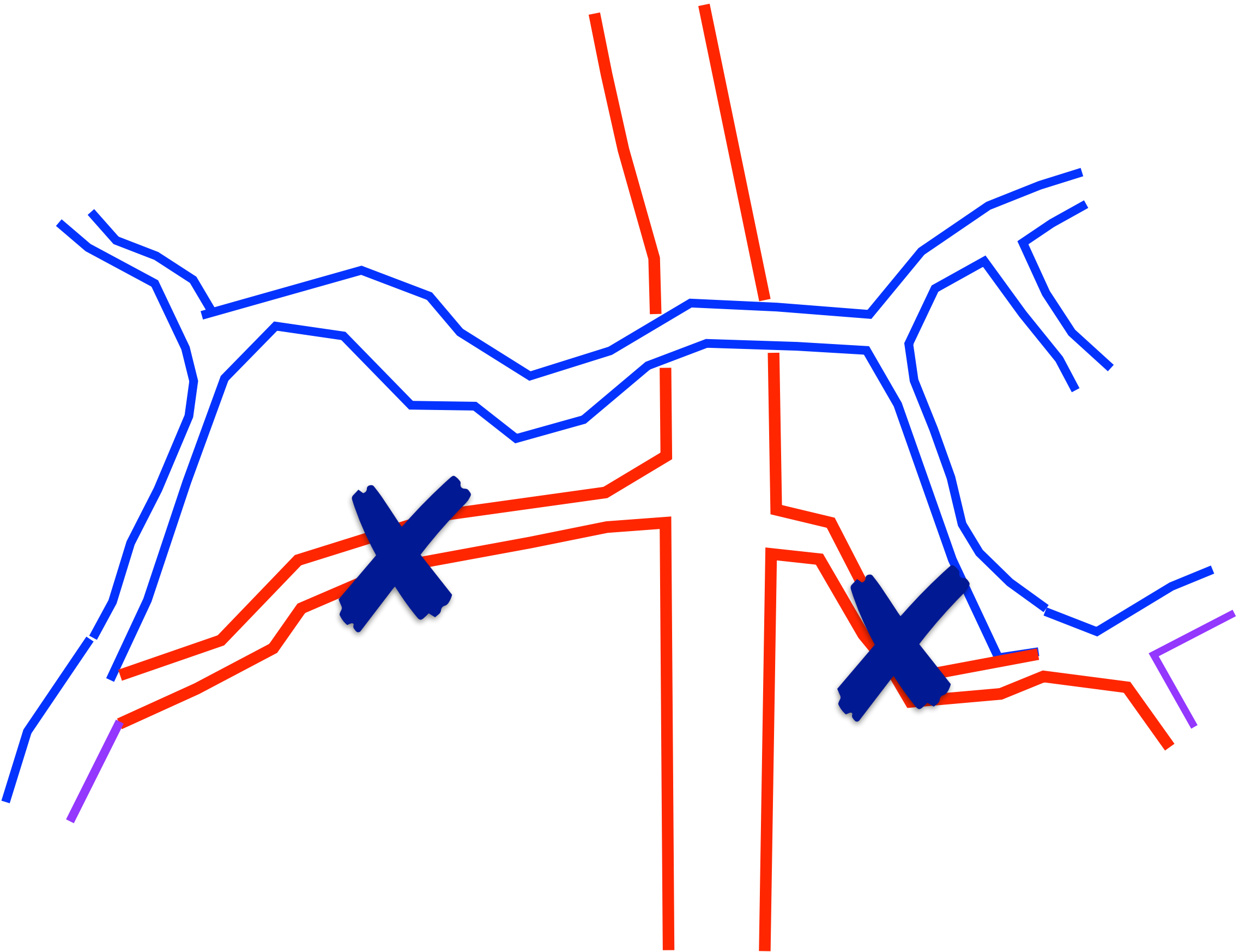
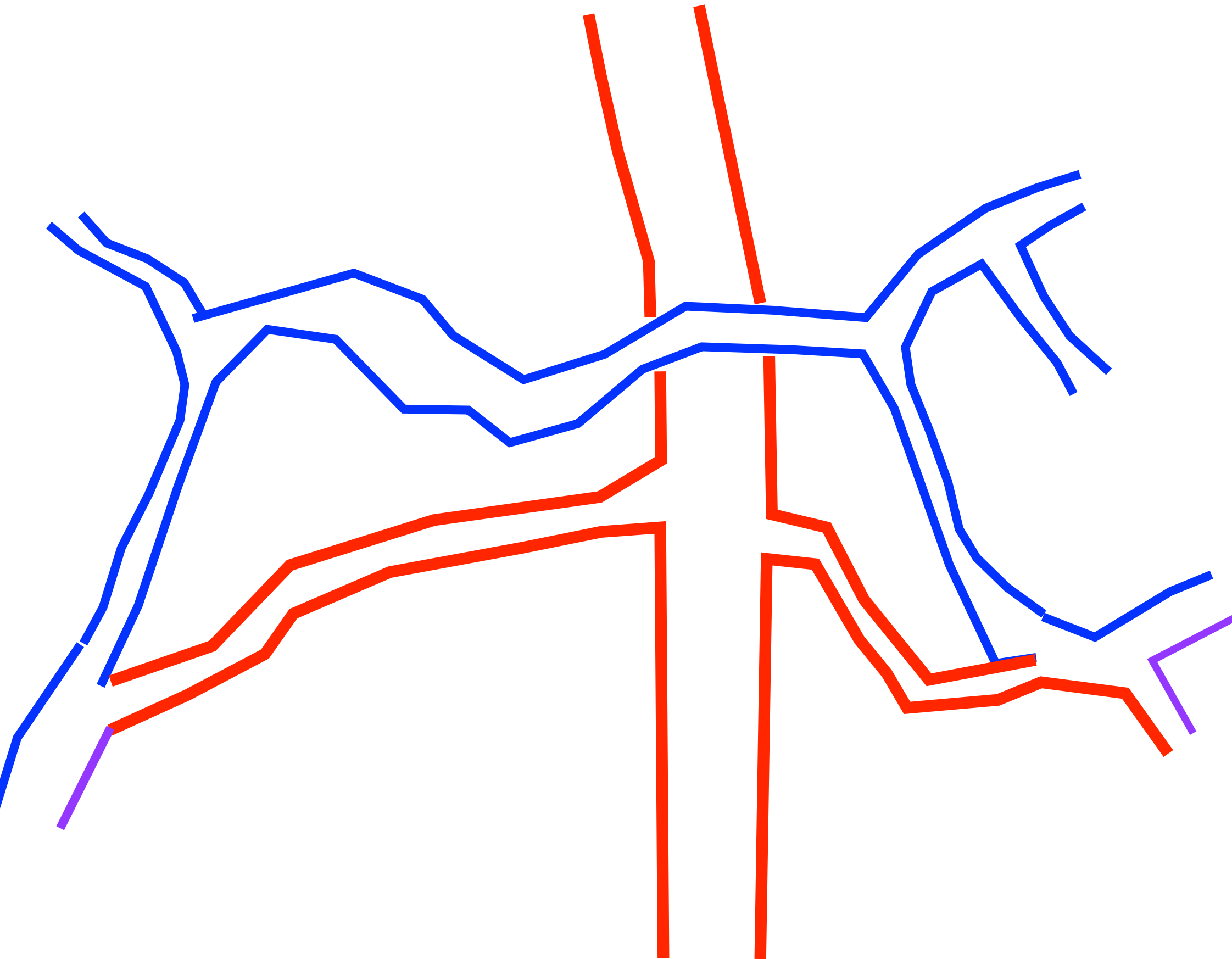
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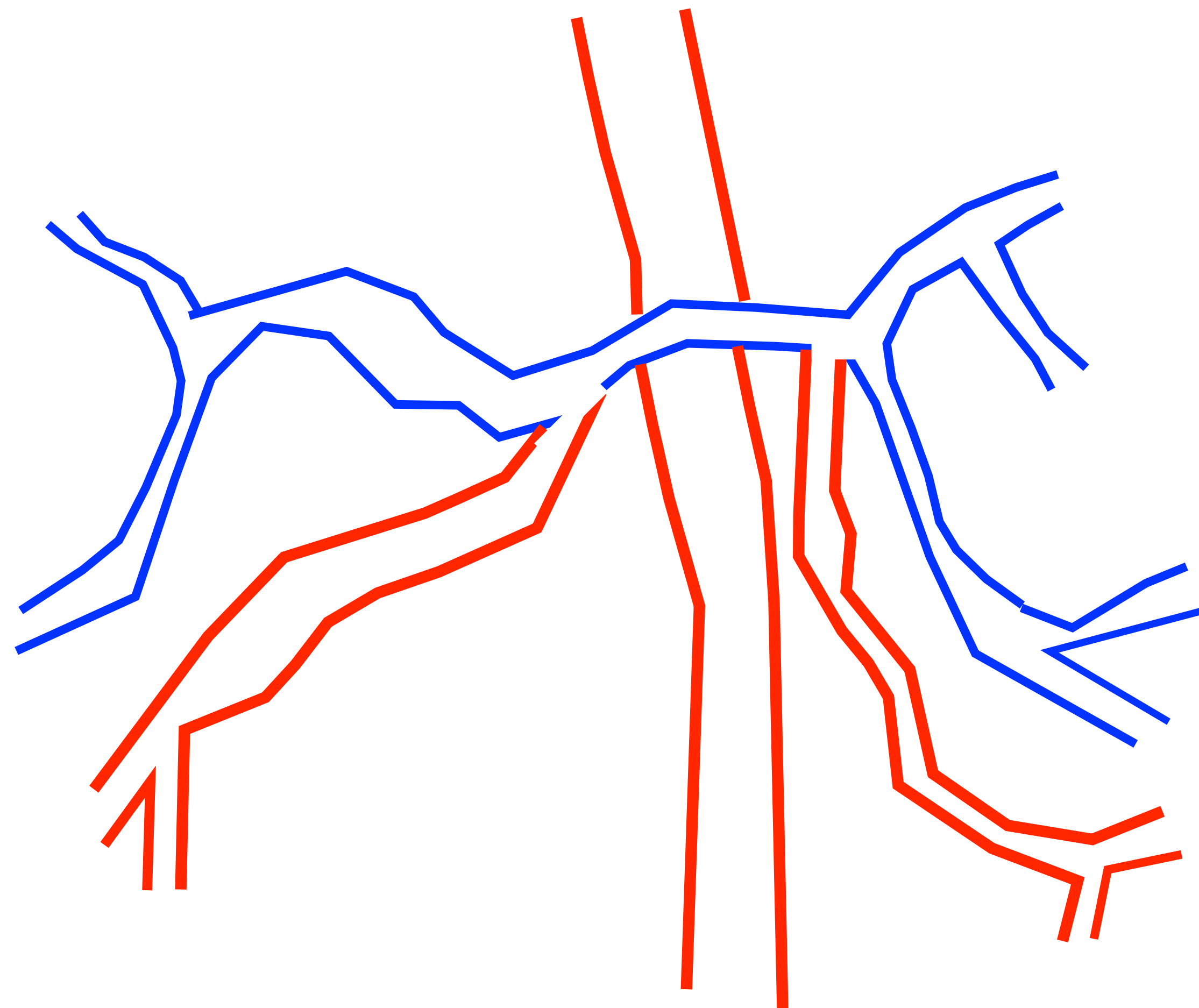
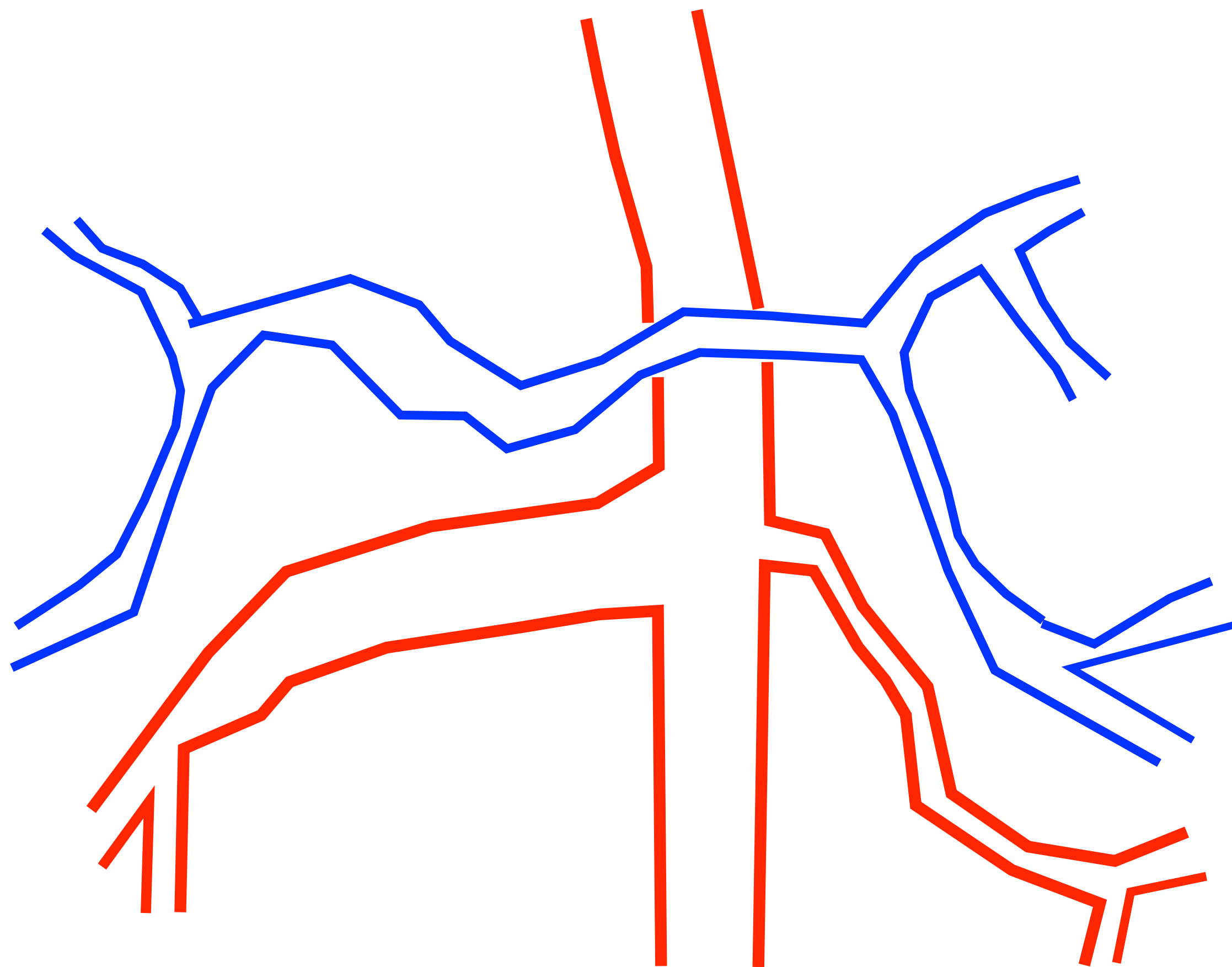




# MAPCAs with or without communication with the pulmonary arteries

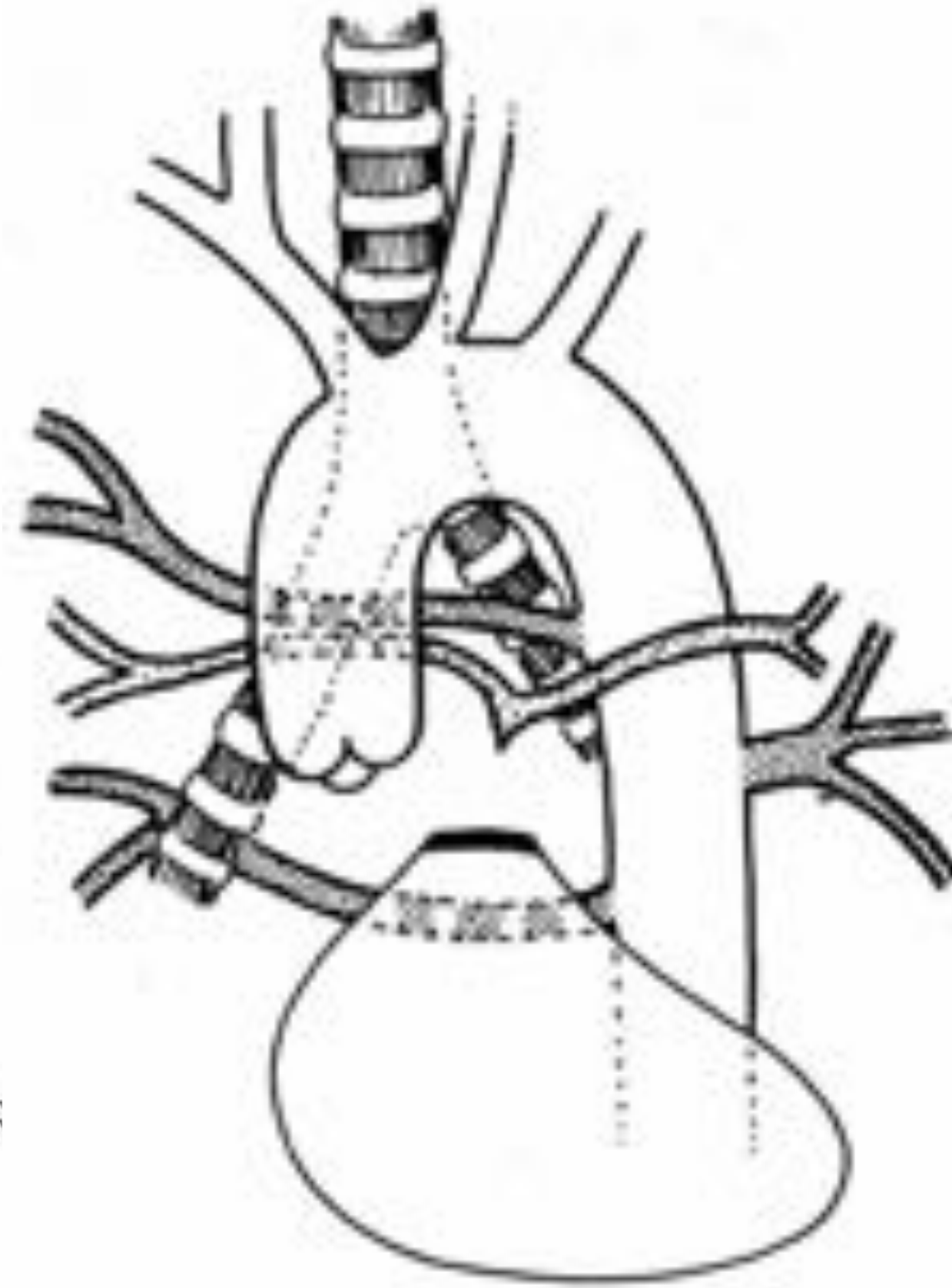
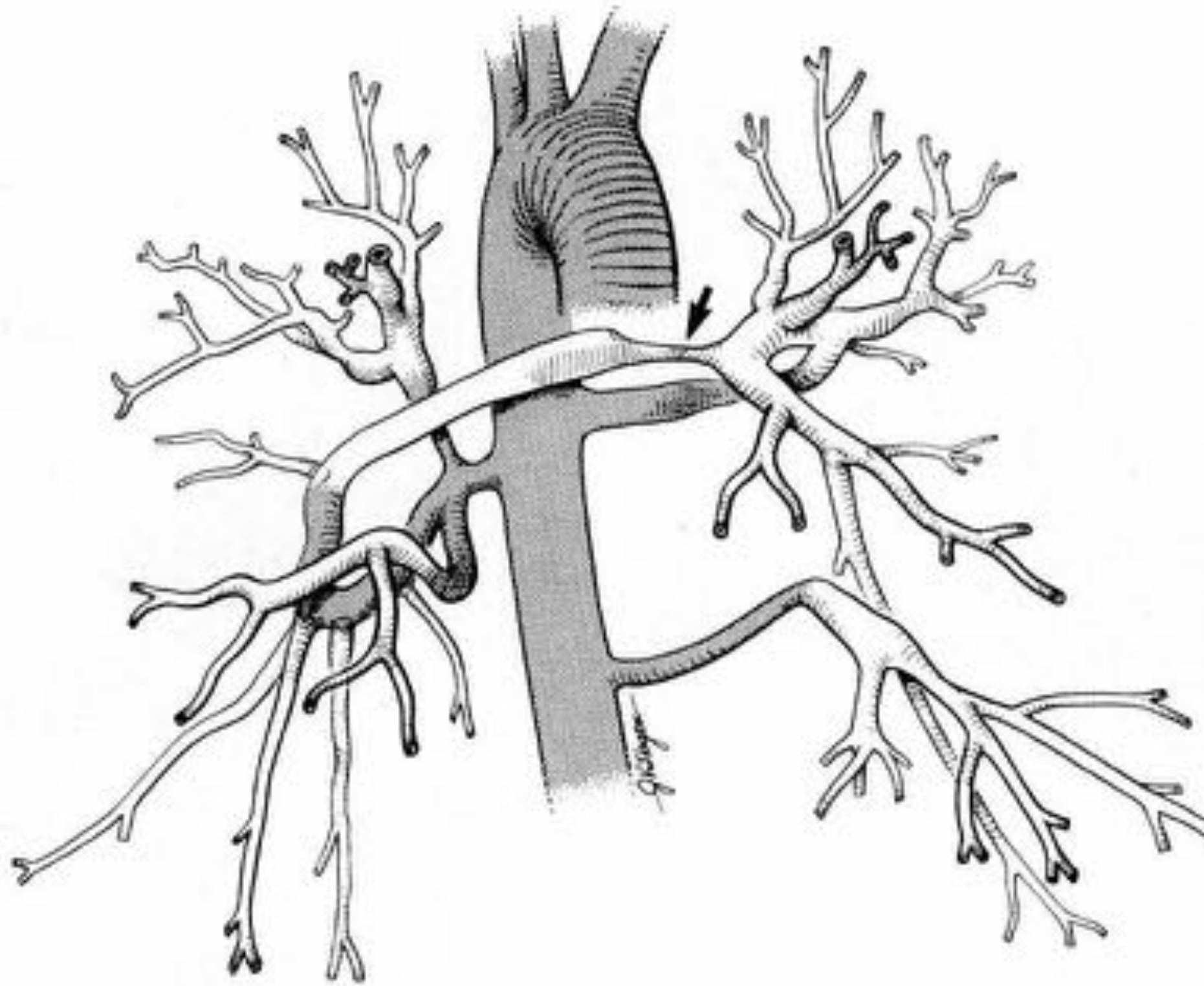


# MAPCAs with or without communication with the pulmonary arteries

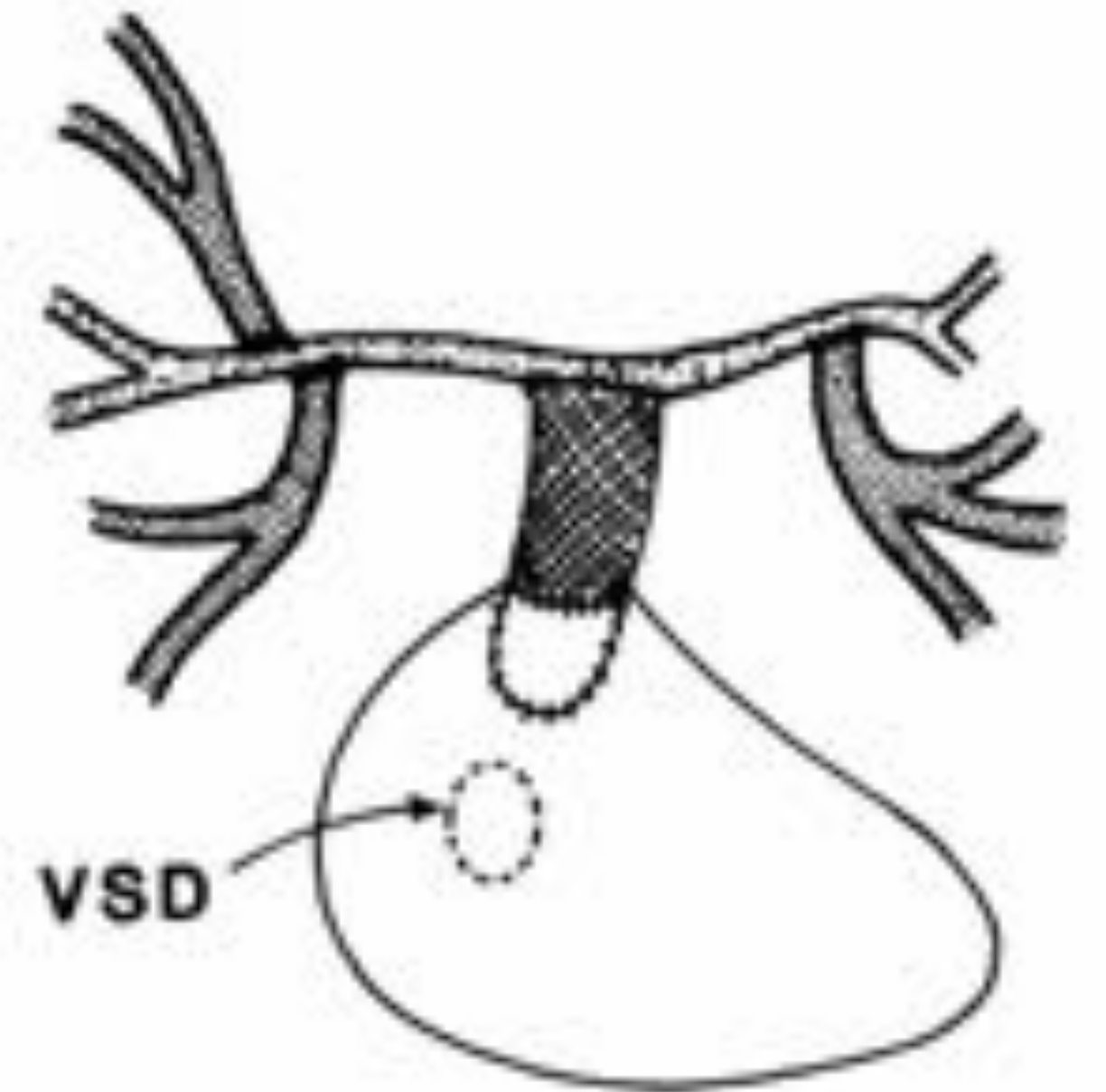




# Unifocalisation



**TOF/PA, MAPC**  
**(PAi 50 mm<sup>2</sup>/M<sup>2</sup>)**

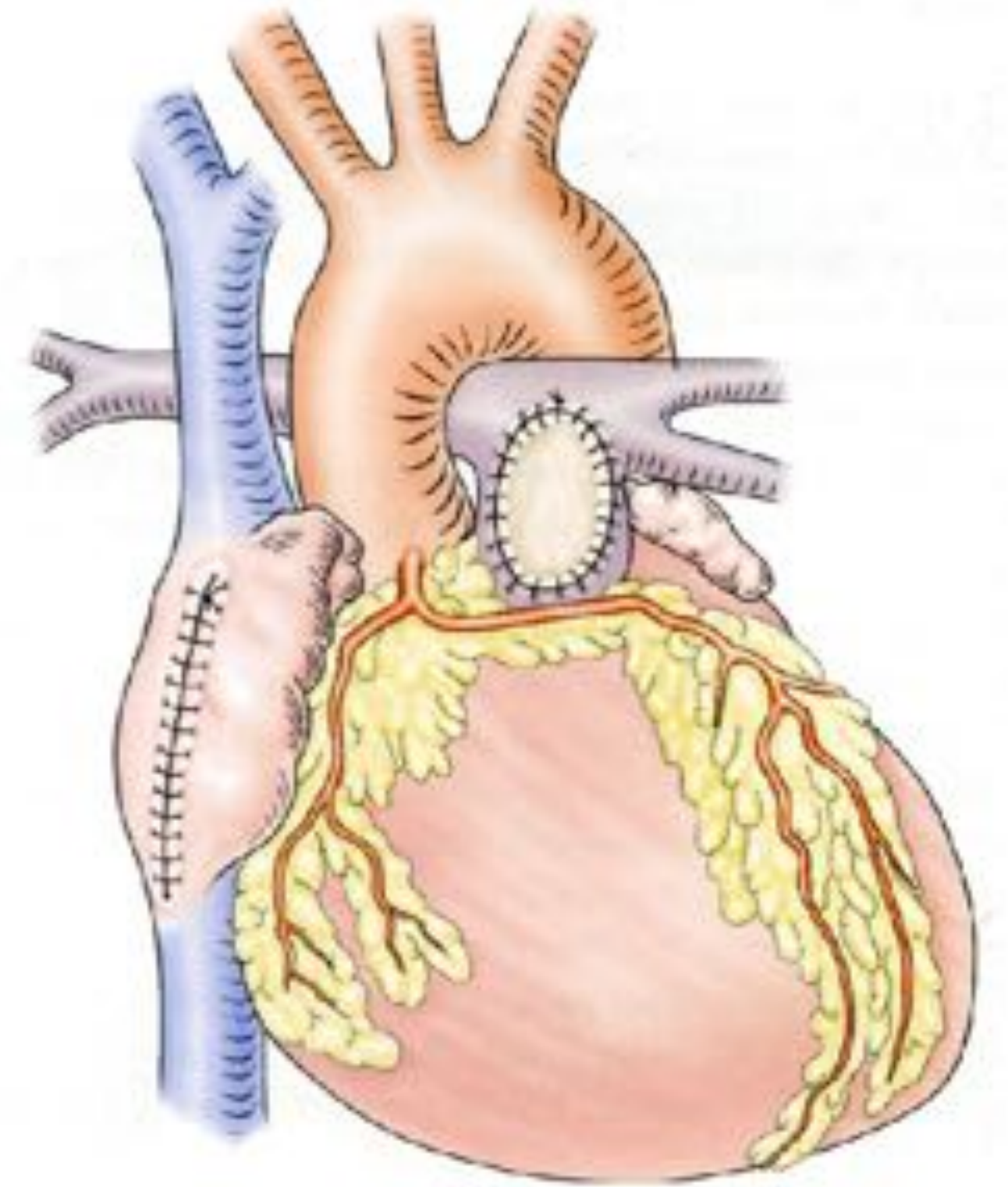
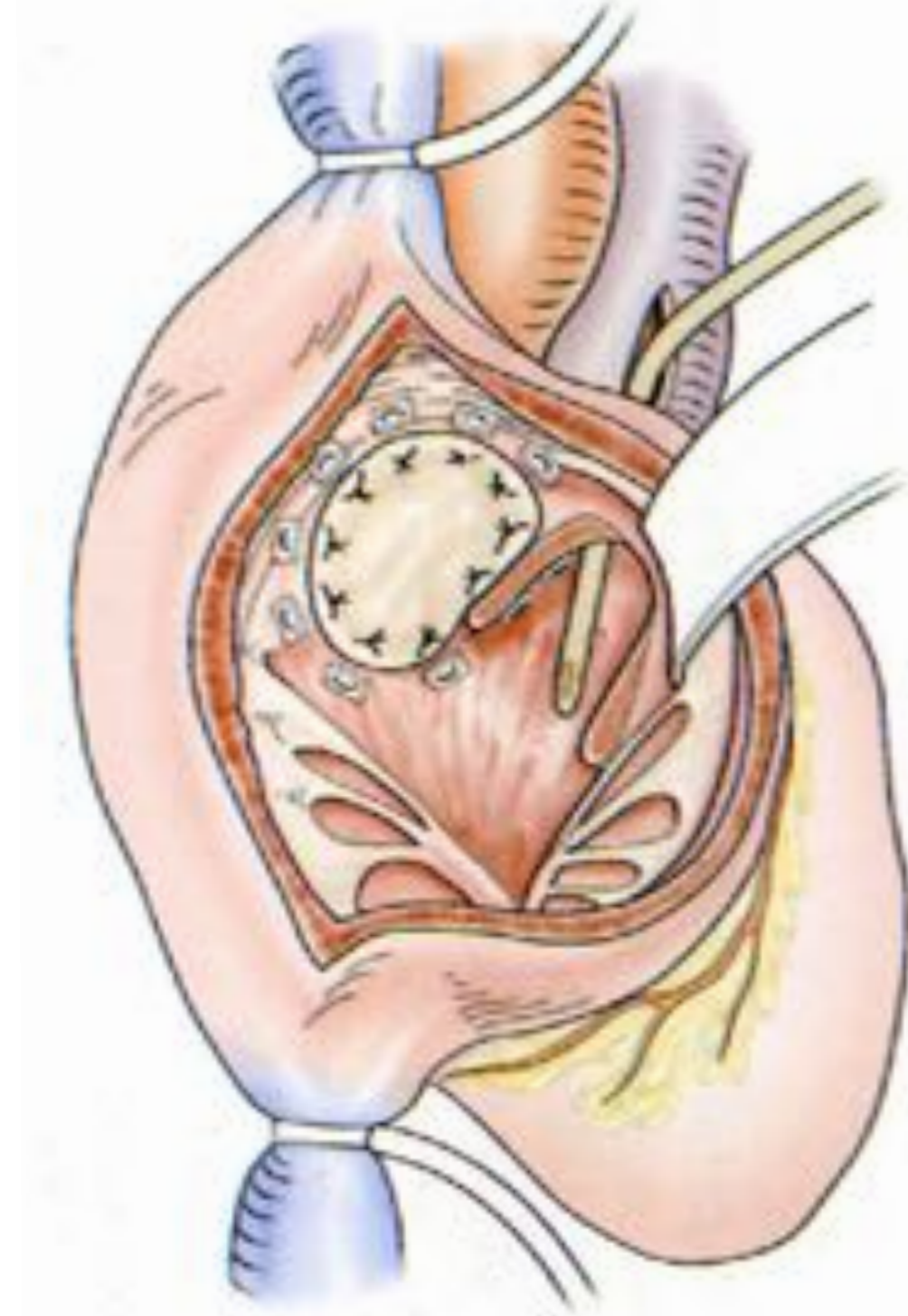
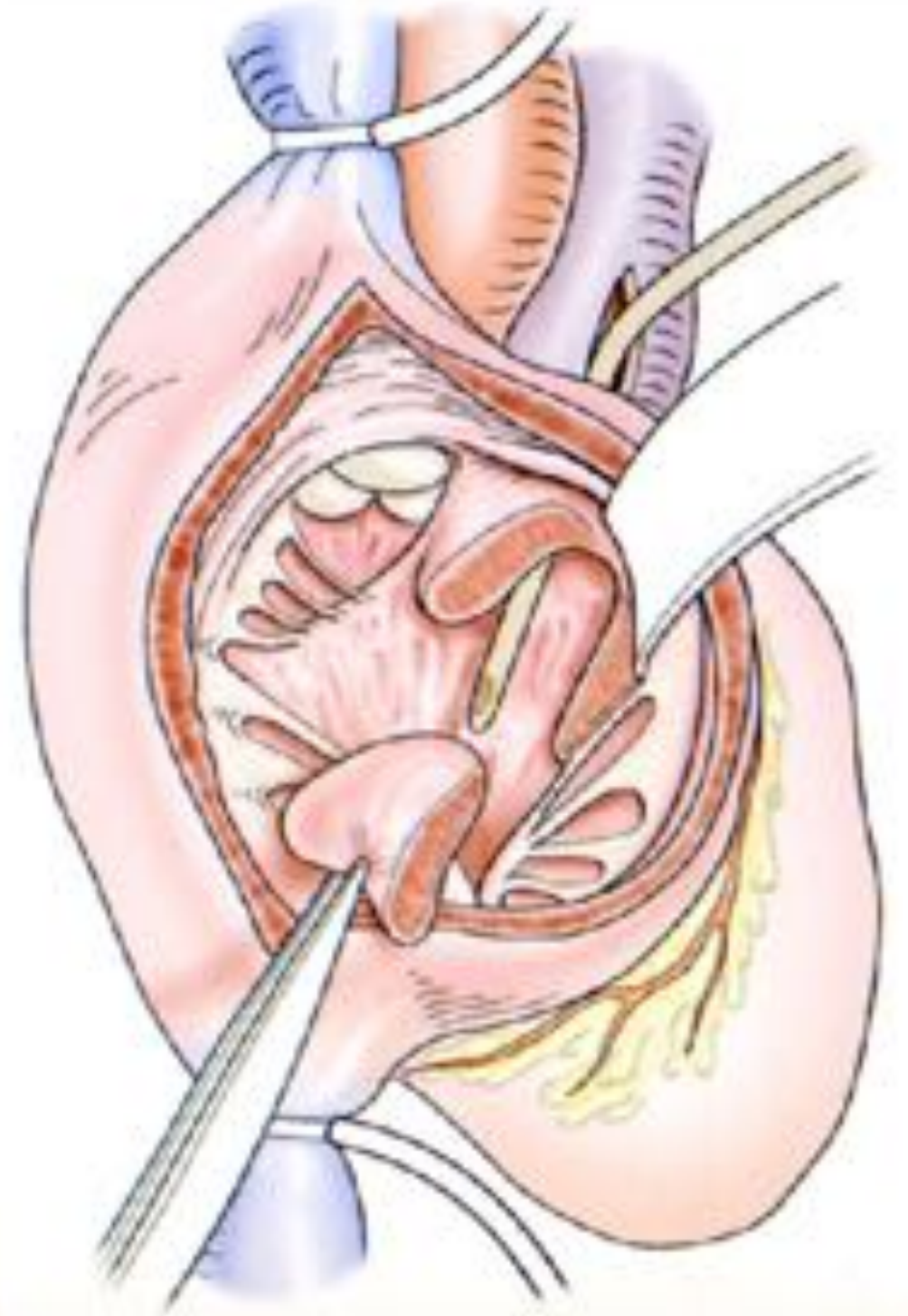
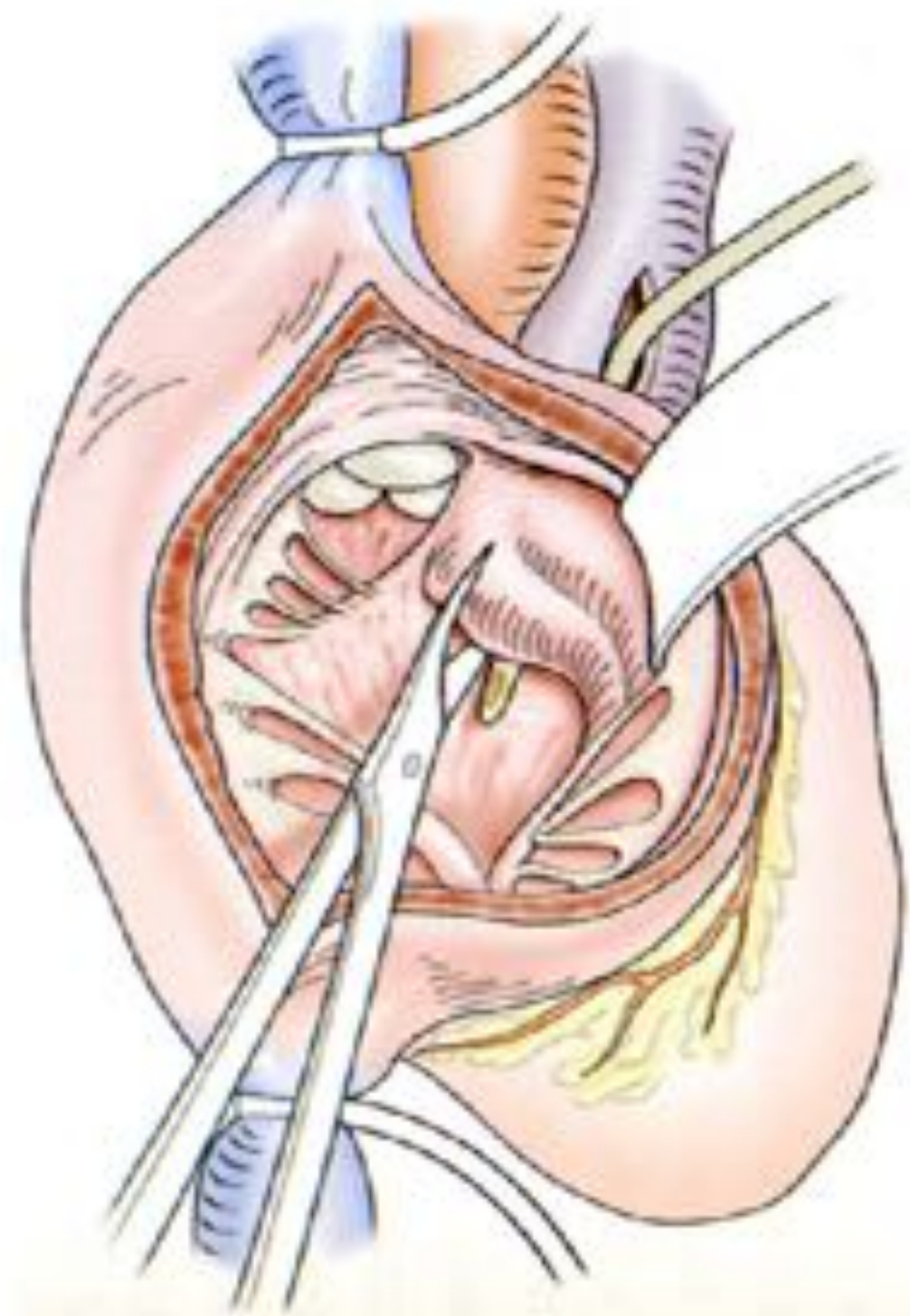


**RV-PA homograft**  
**and bilateral**  
**unifocalization**

# Strategy for repair

## What are the main issues ?

- Neonatal repair could be performed
  - closure of VSD is difficult
  - pulmonary arteries size and PVR do not always allow repair without conduit
- Repair with closure of the VSD in children > 6 kgs
  - If closure of VSD is possible : multiple VSD
  - Without conduit : coronary artery anatomy
  - Without valve : size/stenosis of pulmonary artery branches

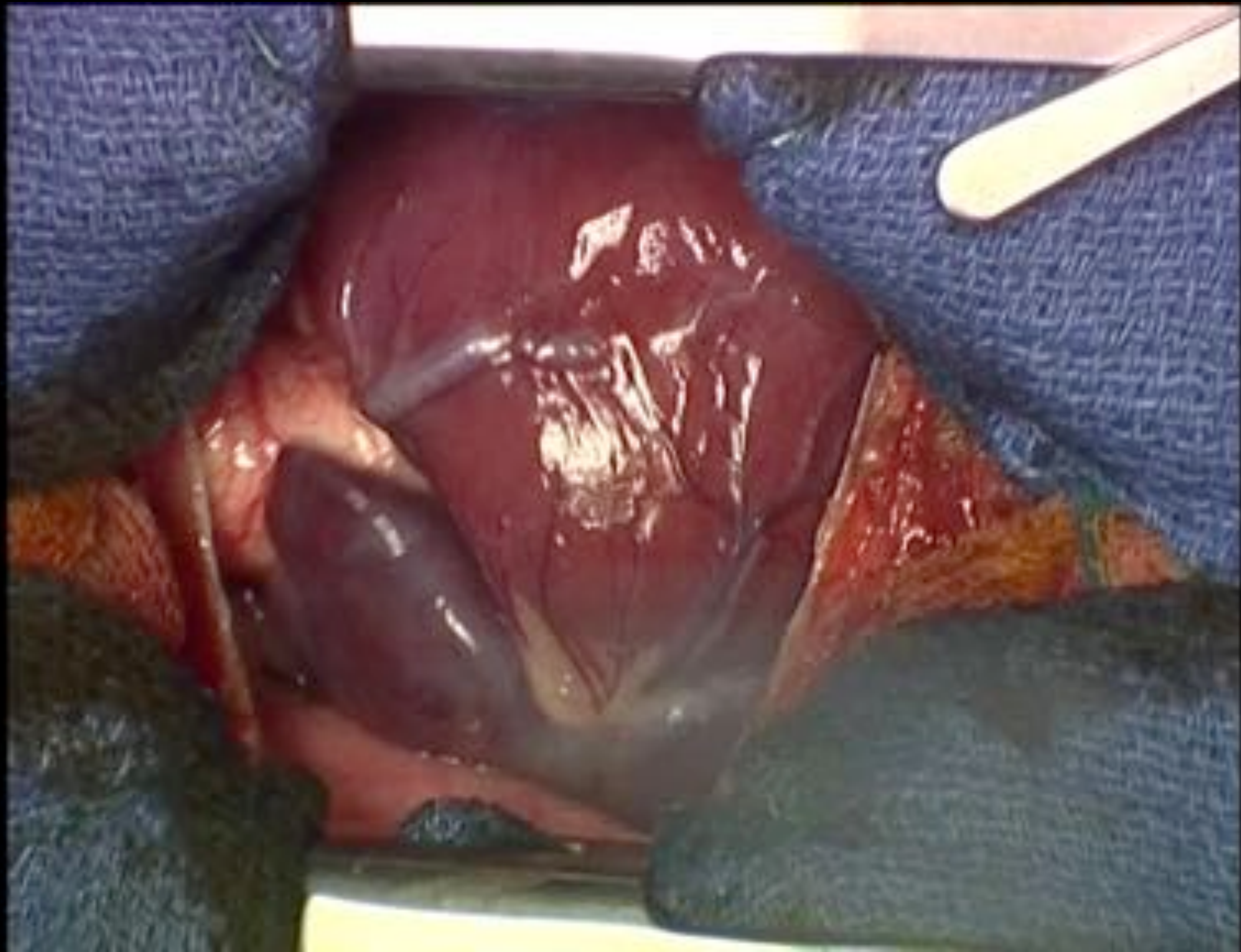




Tetralogy of Fallot Repair



Cincinnati  
Children's



# Late outcome

## What are the issues ?

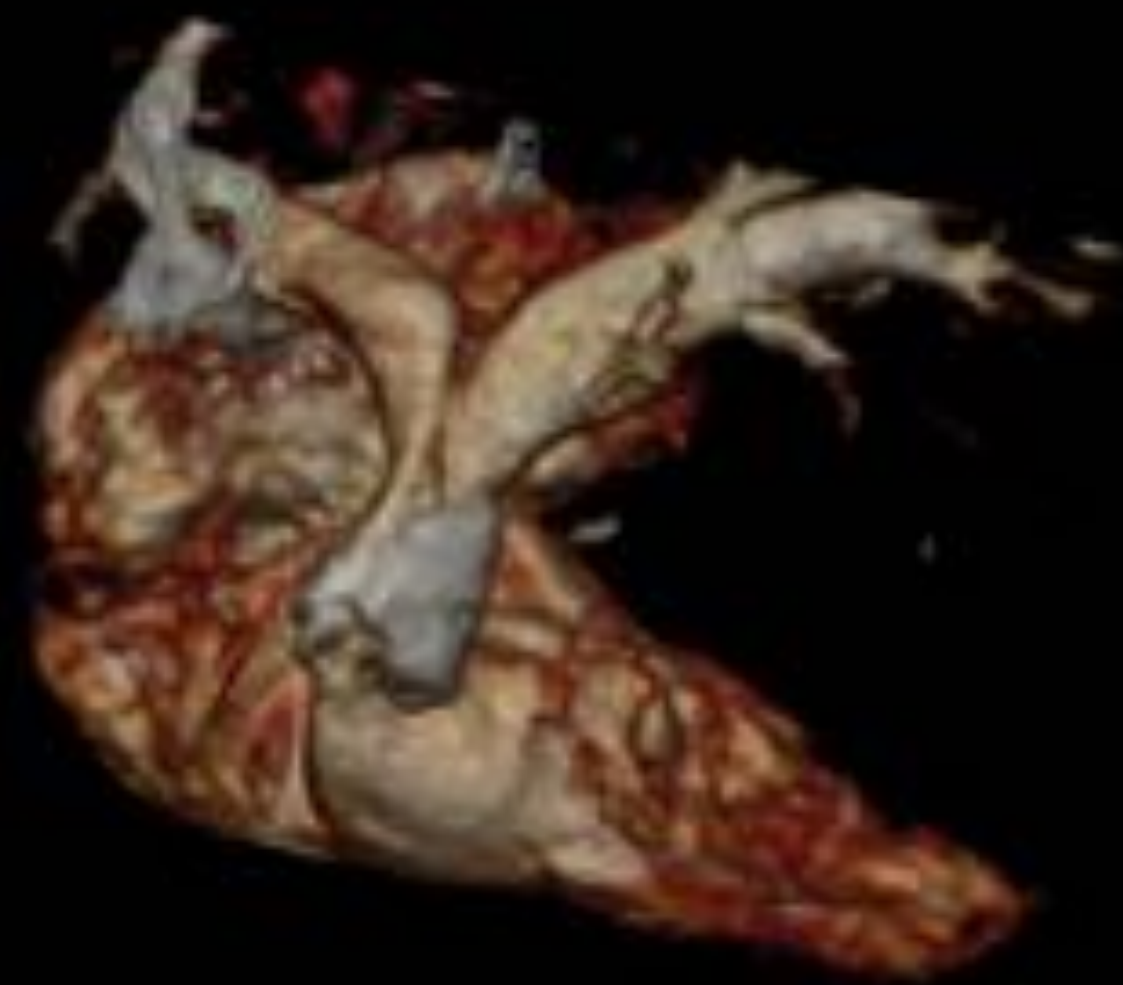
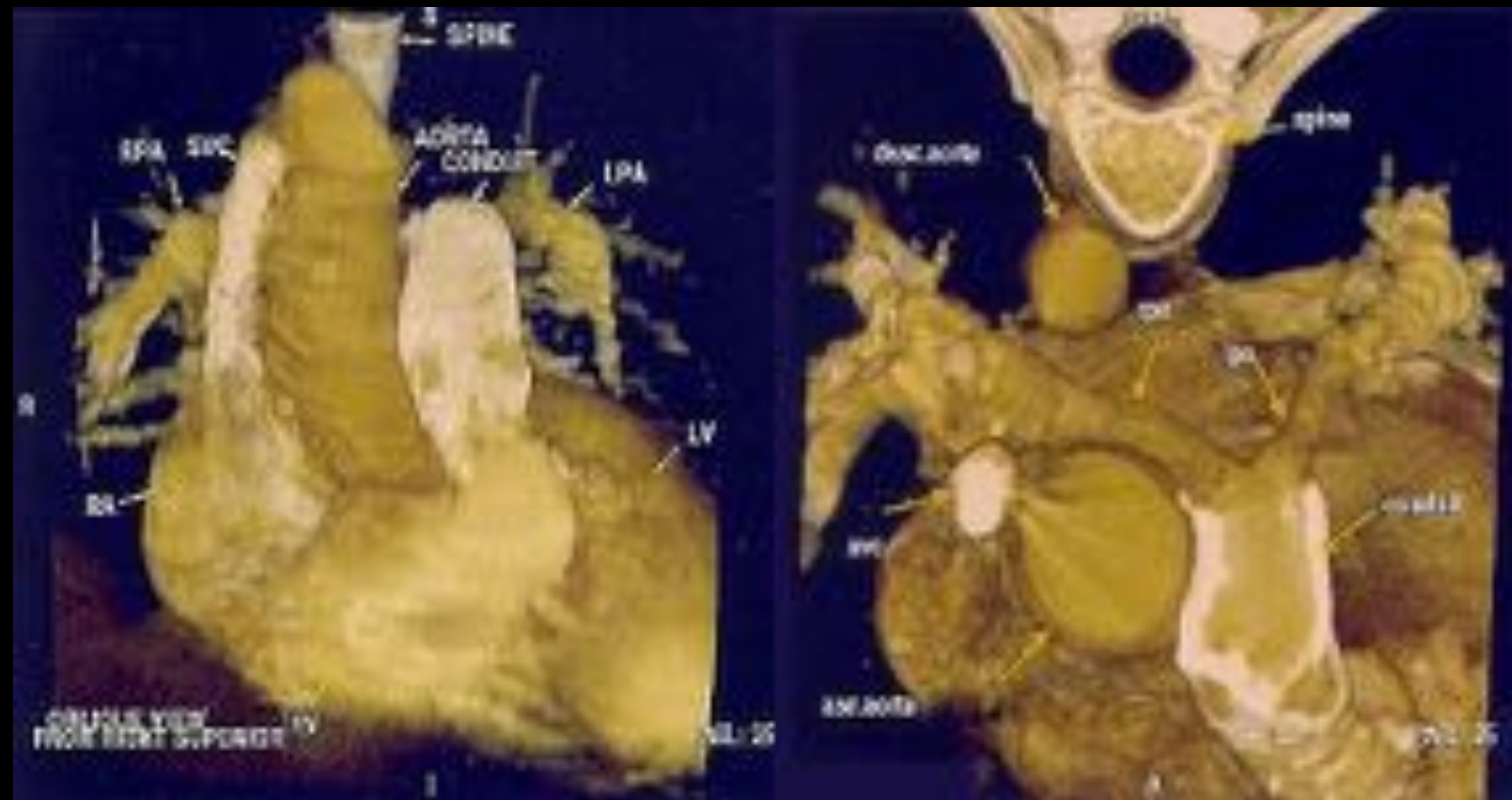
- Right outflow tract obstructions
- Pulmonary branches stenoses
- Chronic pulmonary regurgitation
- Ventricular tachycardia, other arrhythmias and sudden death

040V 25.3cm  
STNO Pt.75%

RAS



No VOI  
kv 100  
mA 300  
Rot 0.35s/CH 8 Dens/af  
0.6mm 0.21/0.6cp  
TR: 0.0  
11:50:43 AM  
W = 40



50  
Ex: 5099  
Se: 2  
Volume Rendering: No cut

SP

HORTAL NECKER INFANT

BFOW 22.8cm  
STNDv

R  
9  
5



No V0r  
kv 120  
mA 398  
Rot 0.40s.HZ= 39.4mm/hot  
0.6mm 0.904.1/0.5ap  
TM: 0.0  
11 08 37 AM  
W = 4095 L = 2048

LA

L  
1  
3  
3

11/2/2008 15:46:24





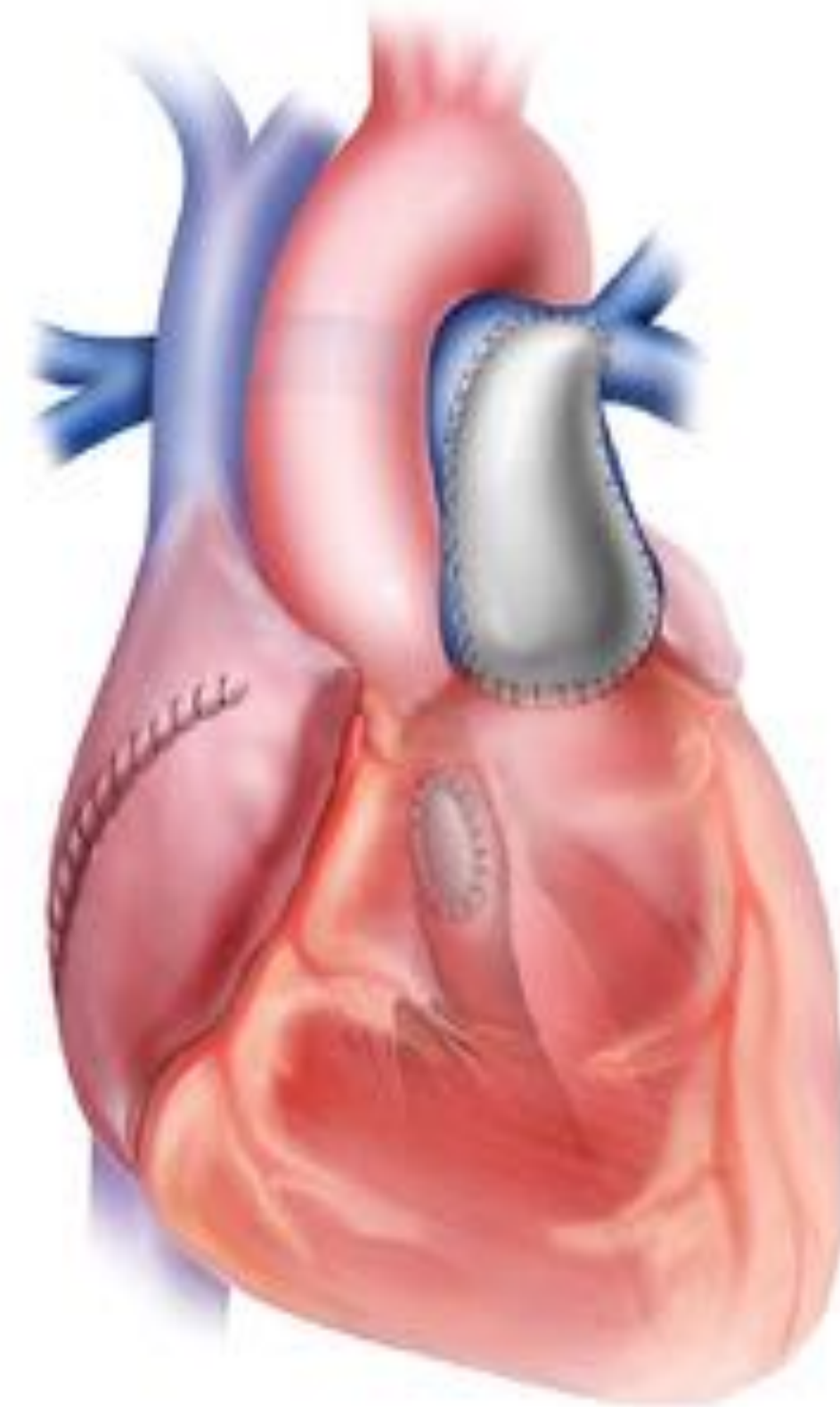
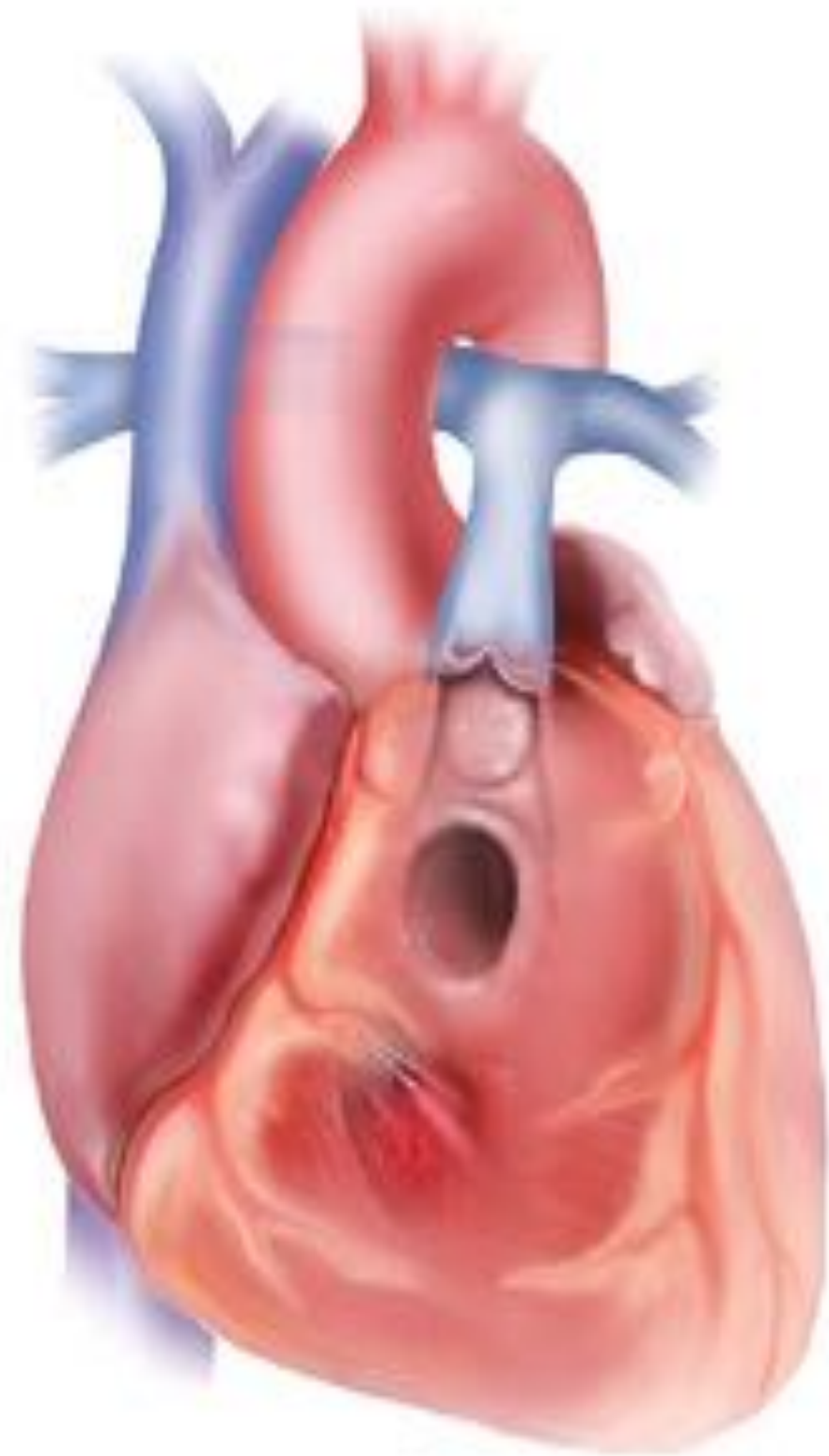


# Conduits VD-AP

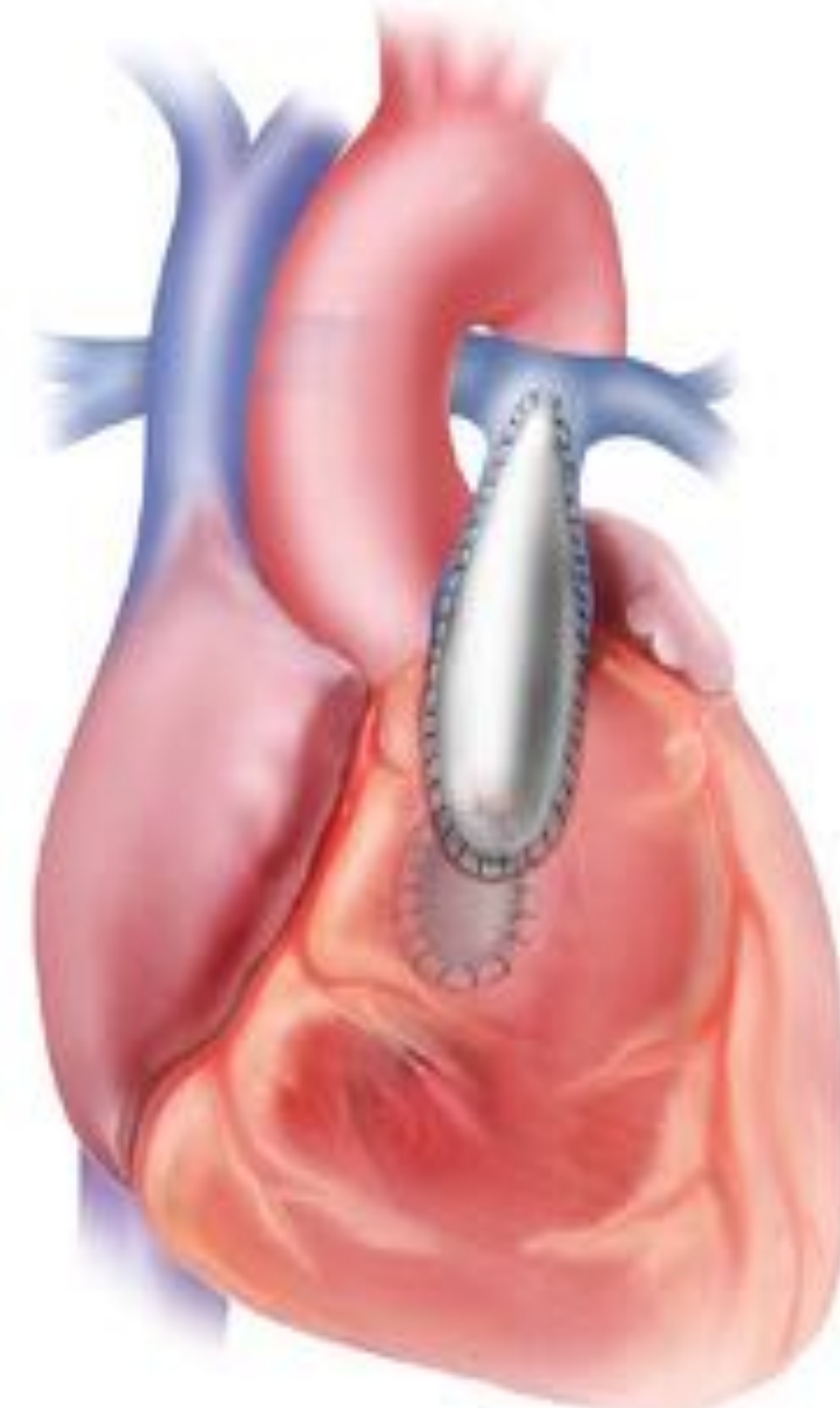


# Traitement chirurgical de la tétralogie de Fallot

## Réparation

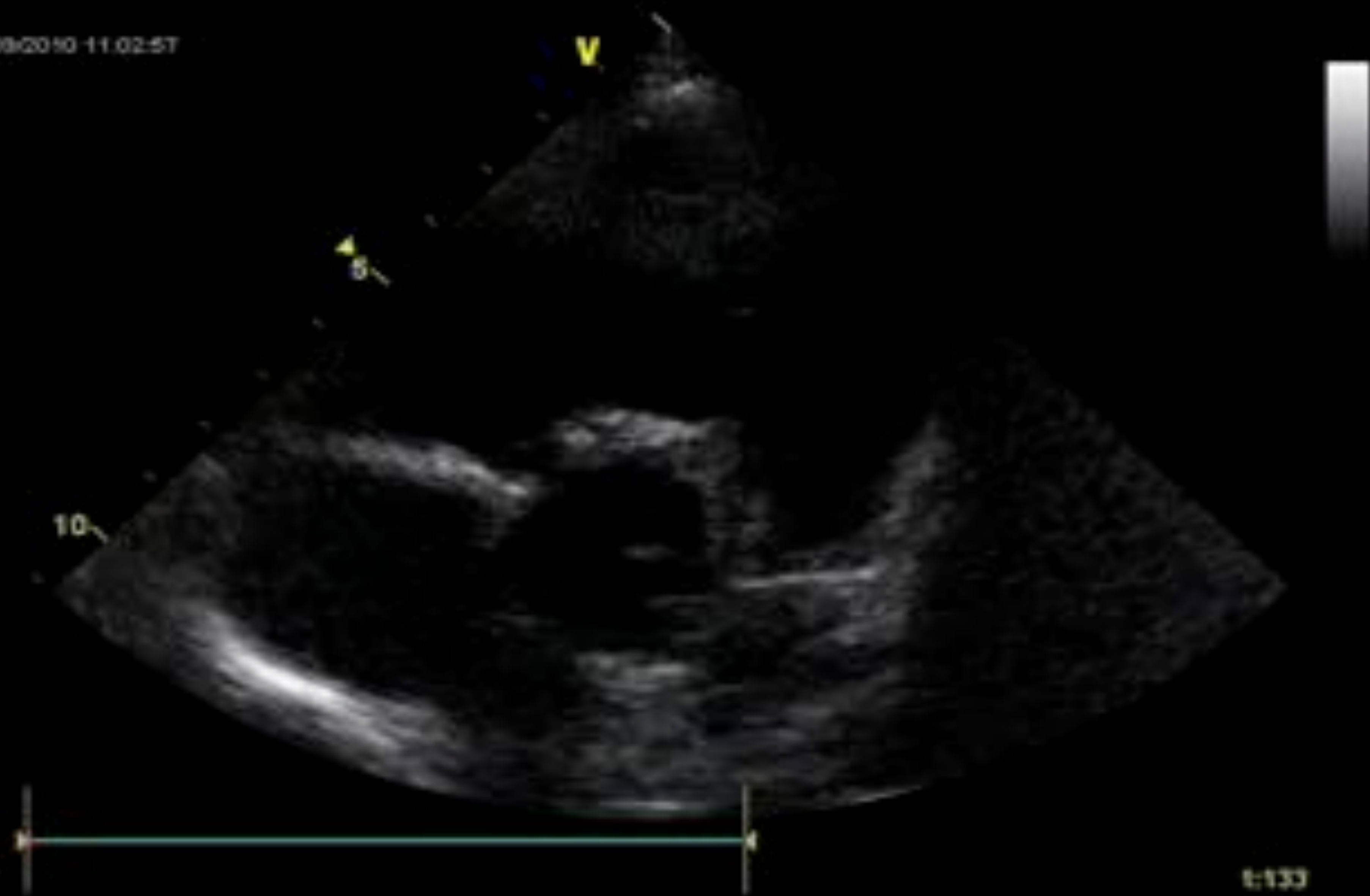


Voix atriale  
Patch pulmonaire



Voix infundibulaire  
Patch infundibulo-pulmonaire

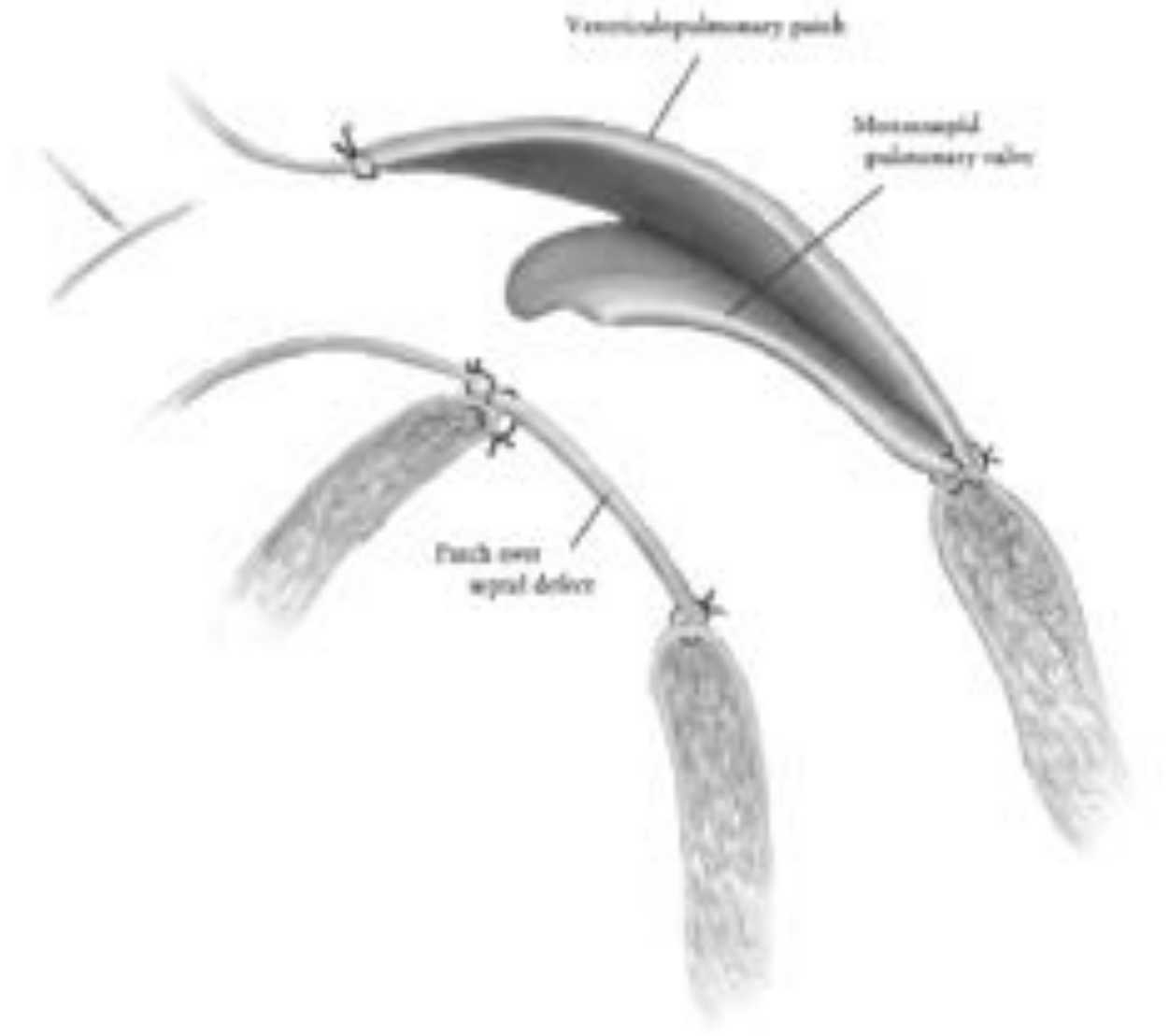
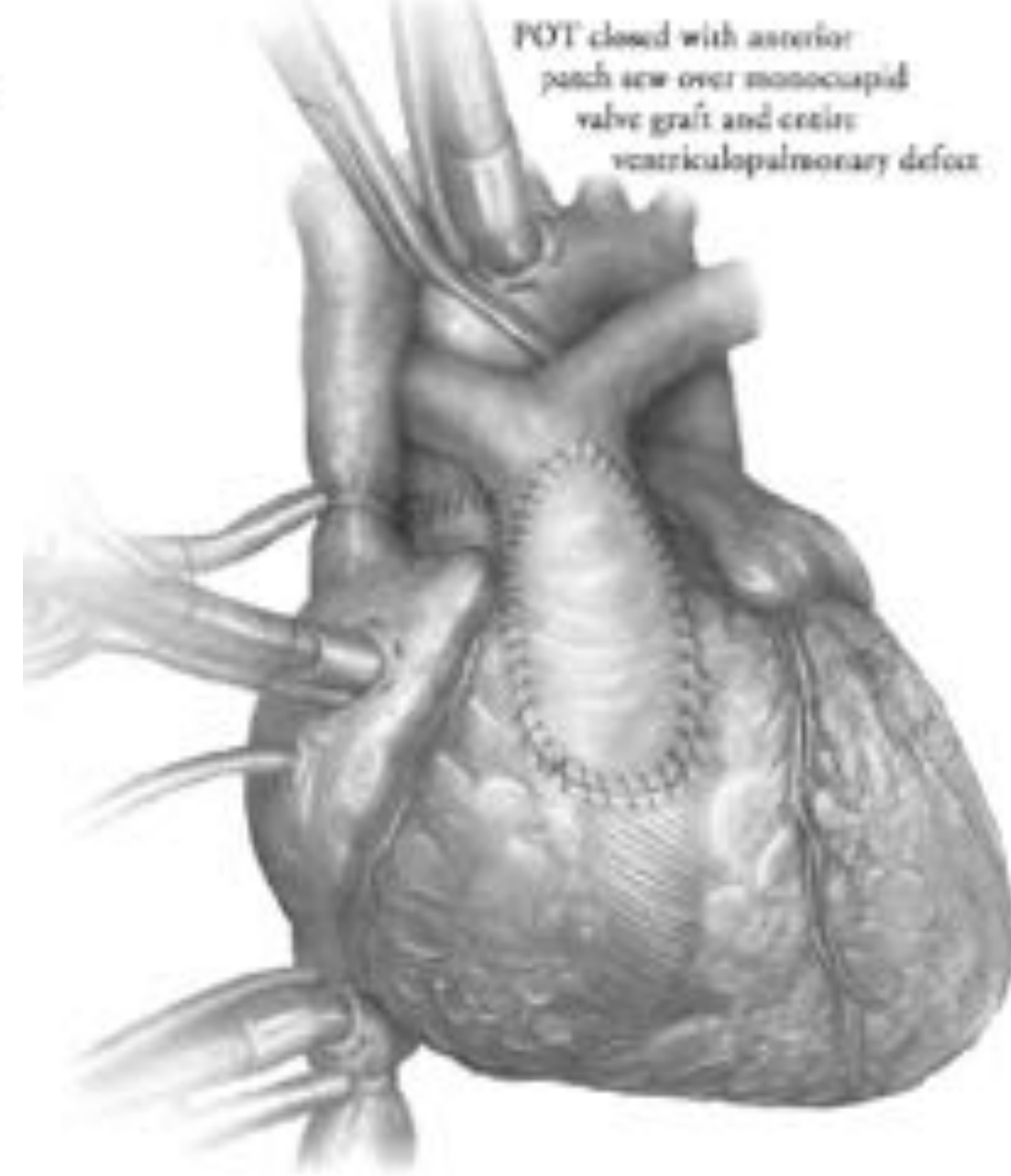
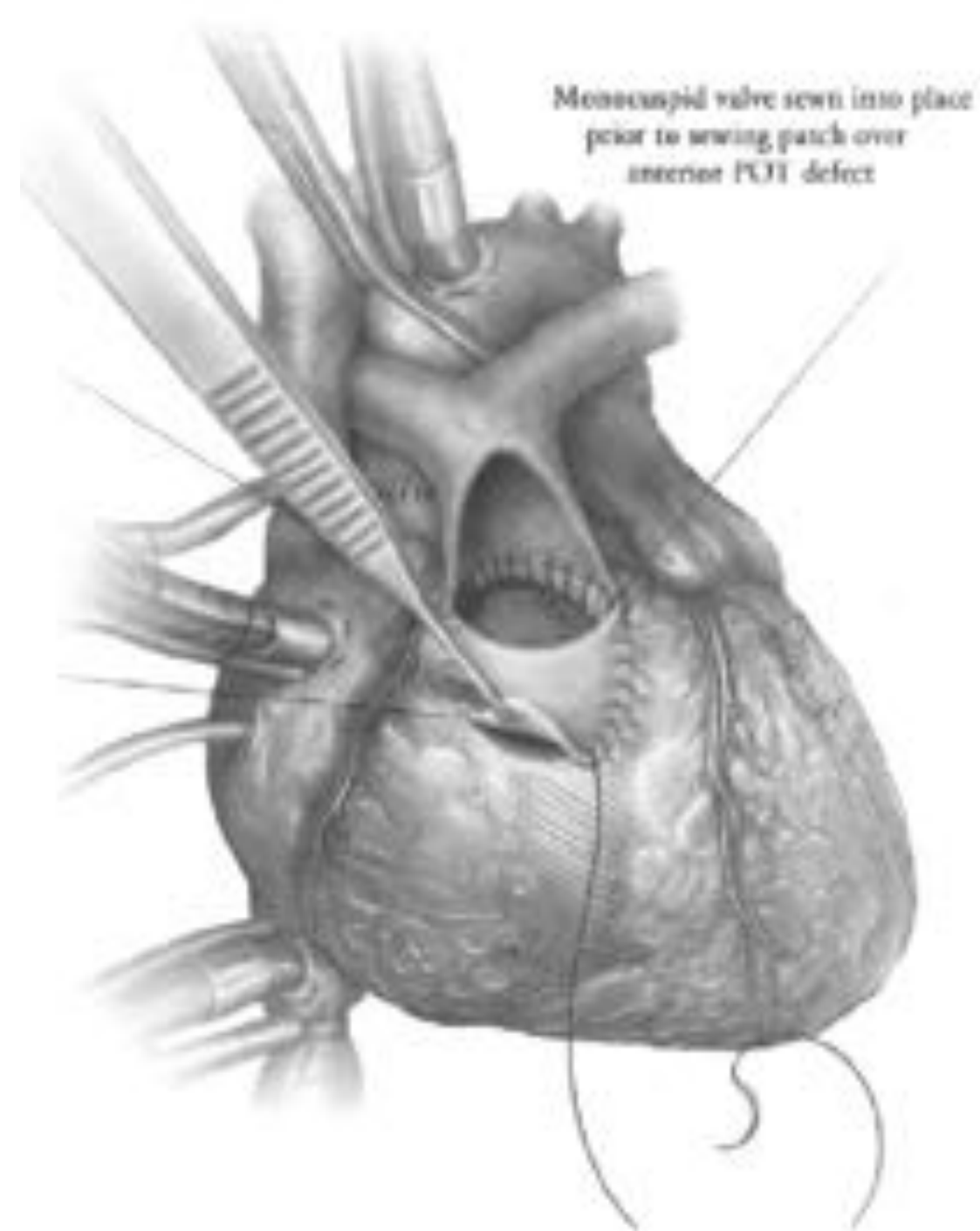
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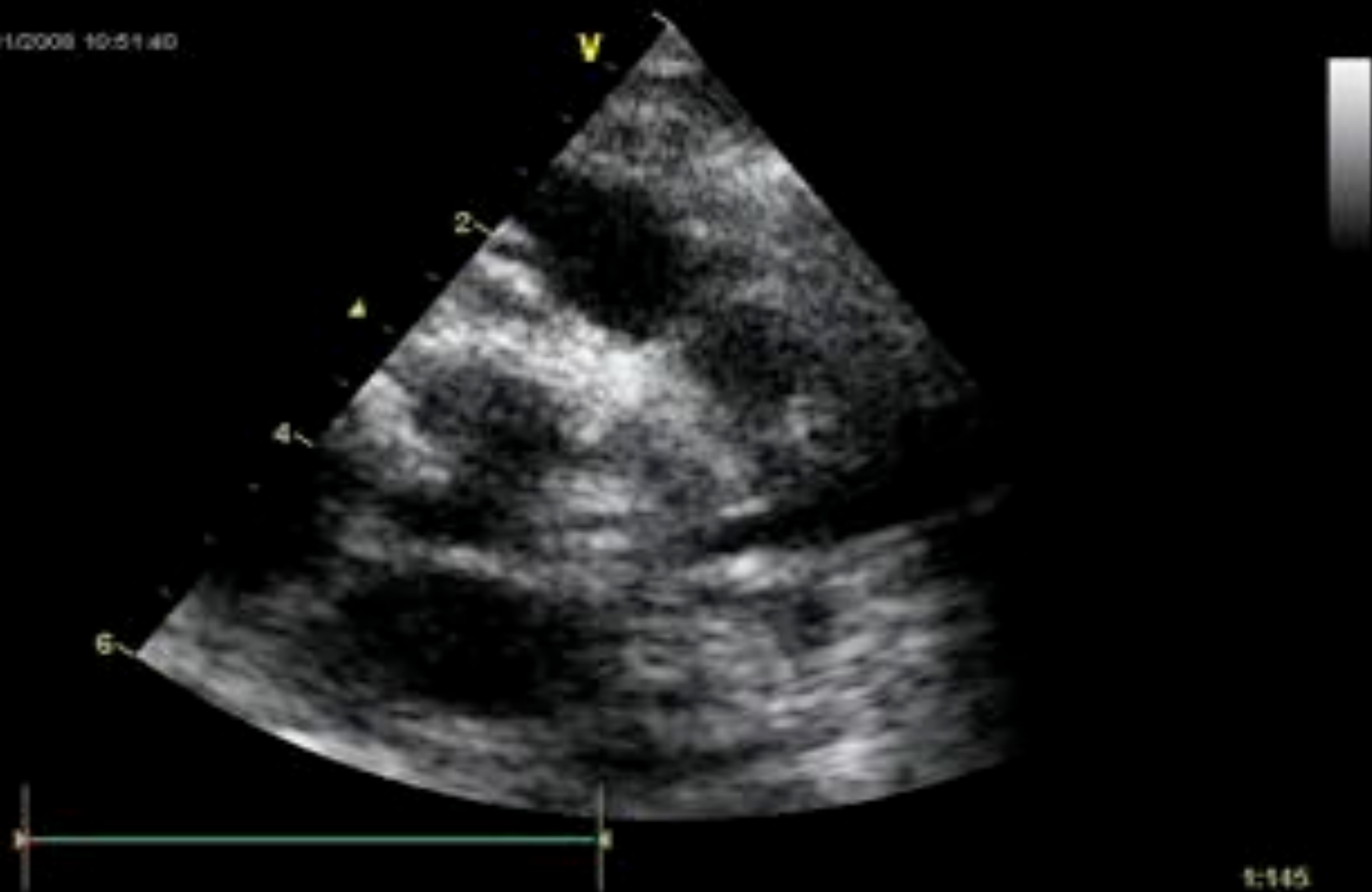
E-133

# Traitement chirurgical des tétralologies de Fallot

Fallot + Agénésie des valves ou petites branches : « Monocusp »



10/5/2008 10:51:40

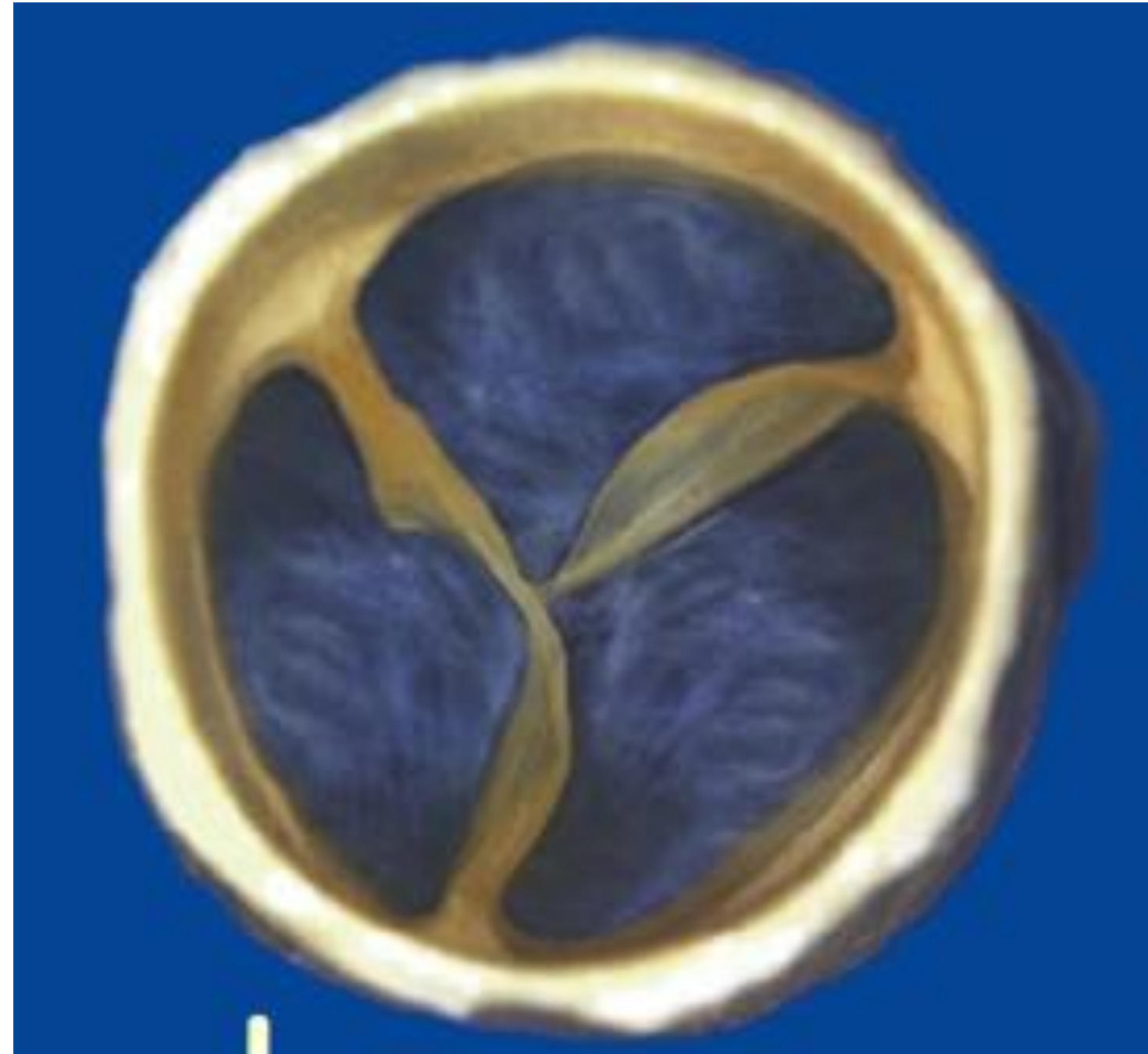


1:145

# Homogreffe pulmonaire



# Tube Contegra - Venpro





27/09/2006 04:48:16

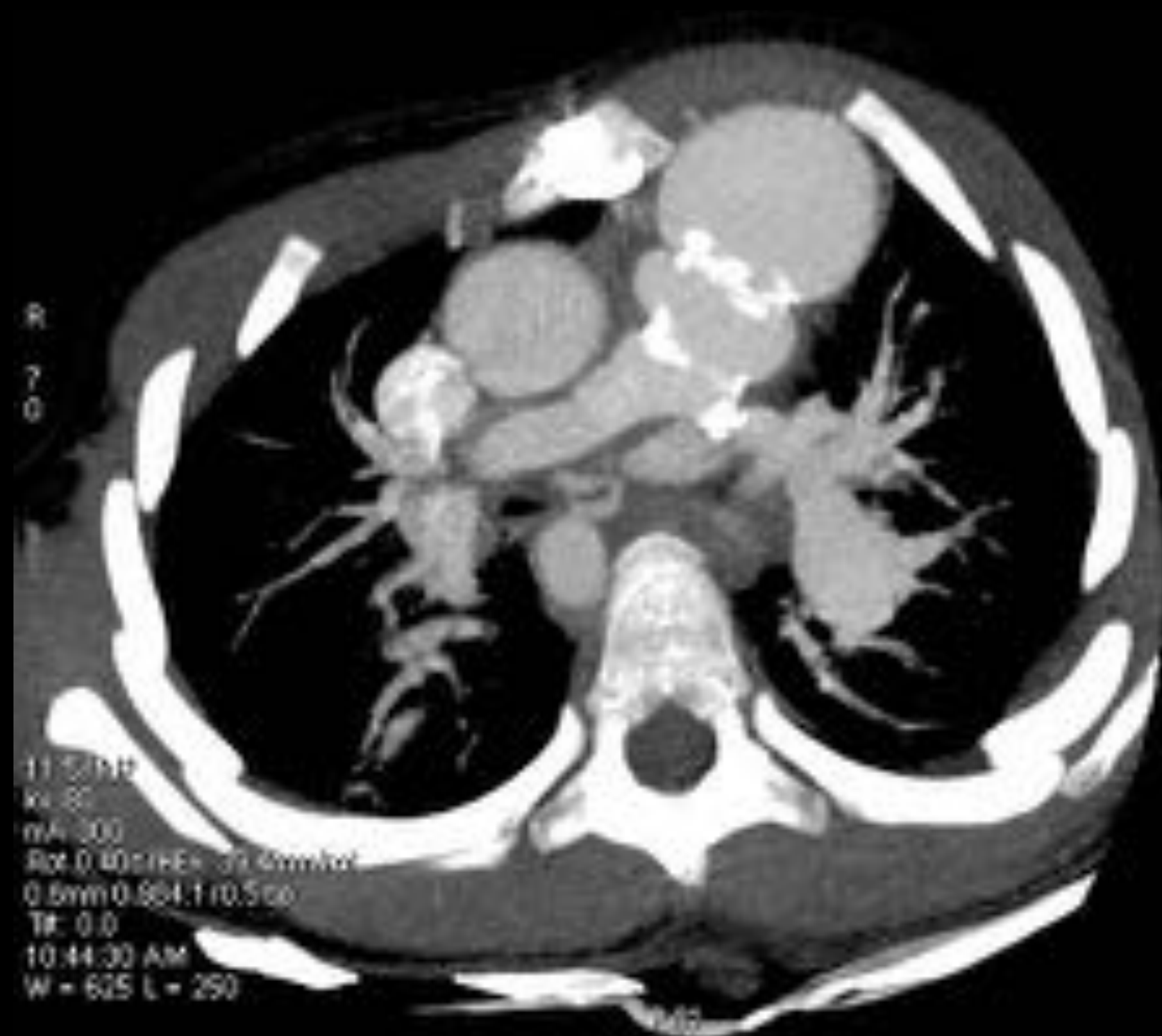


Labcor



VenPro





30  
Ex: 7300  
Se: 2  
Volume Rendering No cut

SPR

ANTAL NECKER INFANT

DF: 0V 25.3cm  
S: TMO Fr: 75%



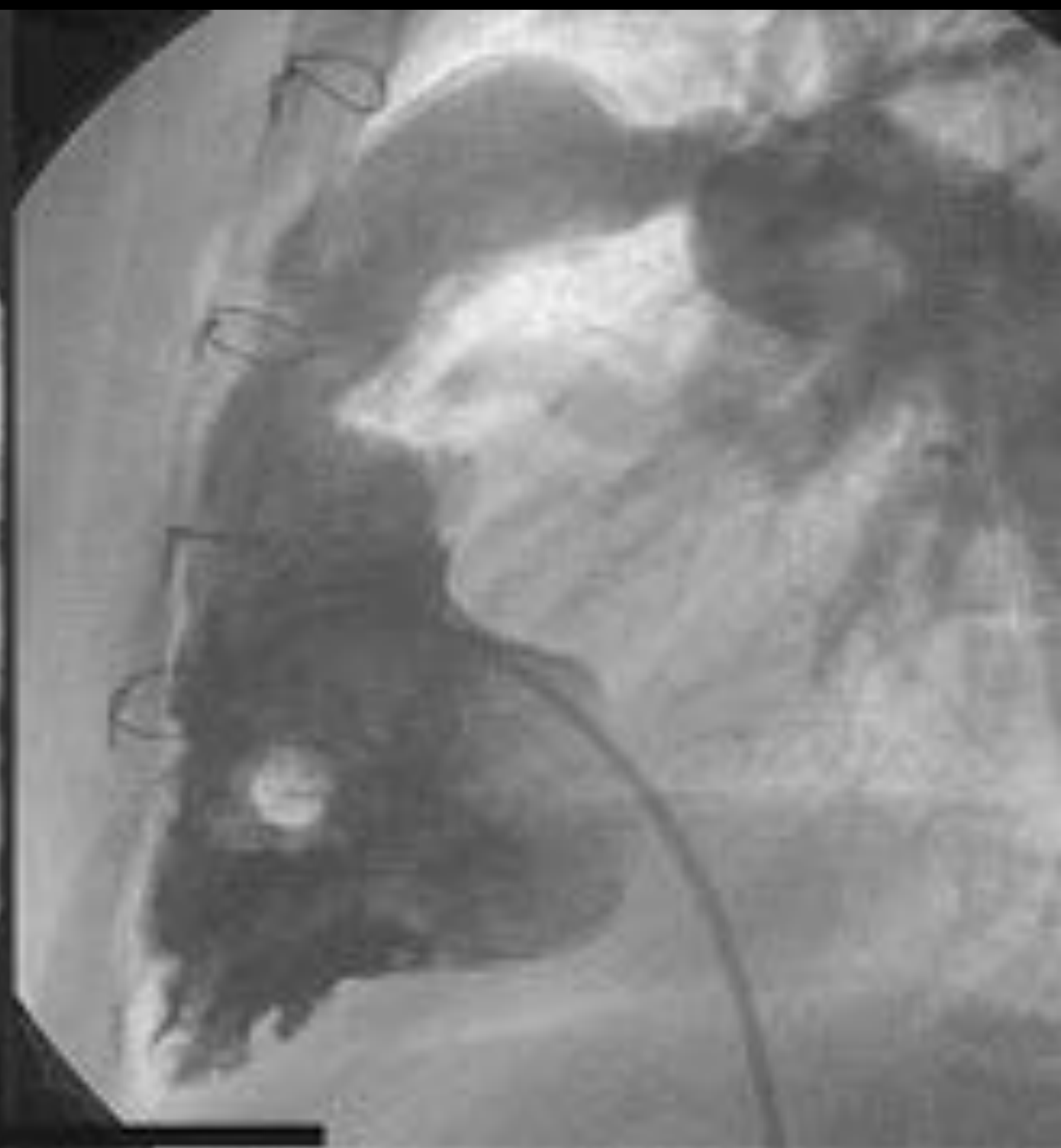
R  
A  
L

R  
A  
L

No VOF  
kv: 100  
mA: 300  
Rot: 0.35p/CH: 8.0mm/rot  
0.6mm 0.2:1 / 0.6tp  
TM: 0.0  
11:58:43 AM  
W = 4095 L = 2048

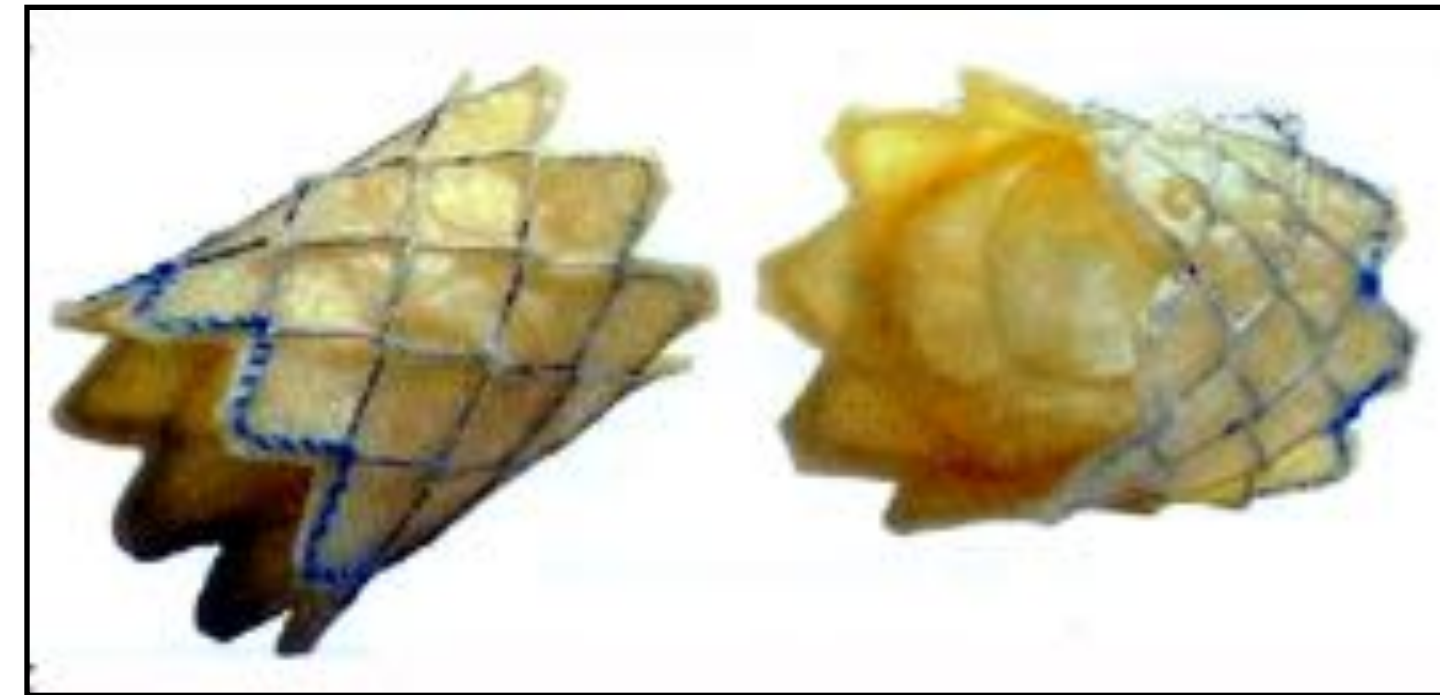
IAL





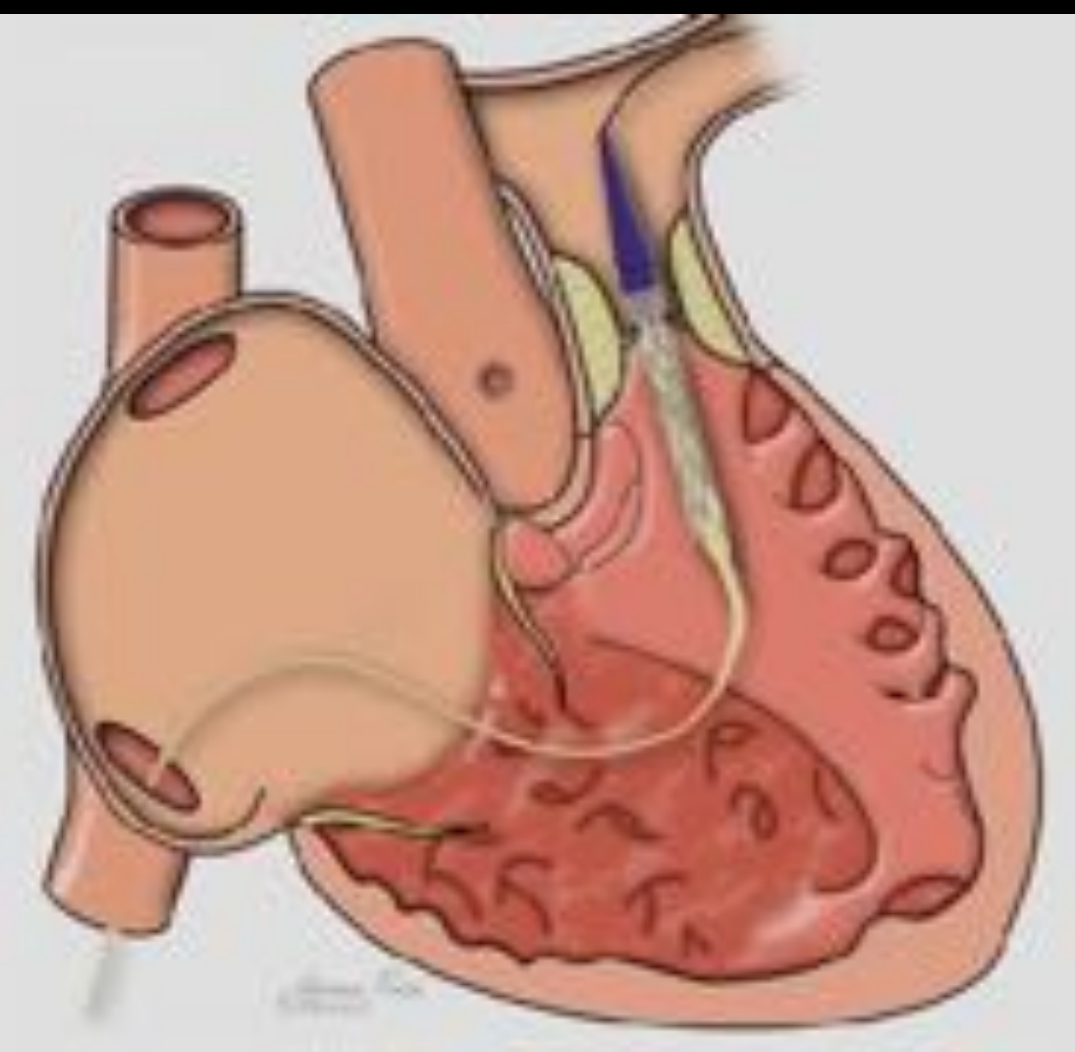
# Valves pulmonaires

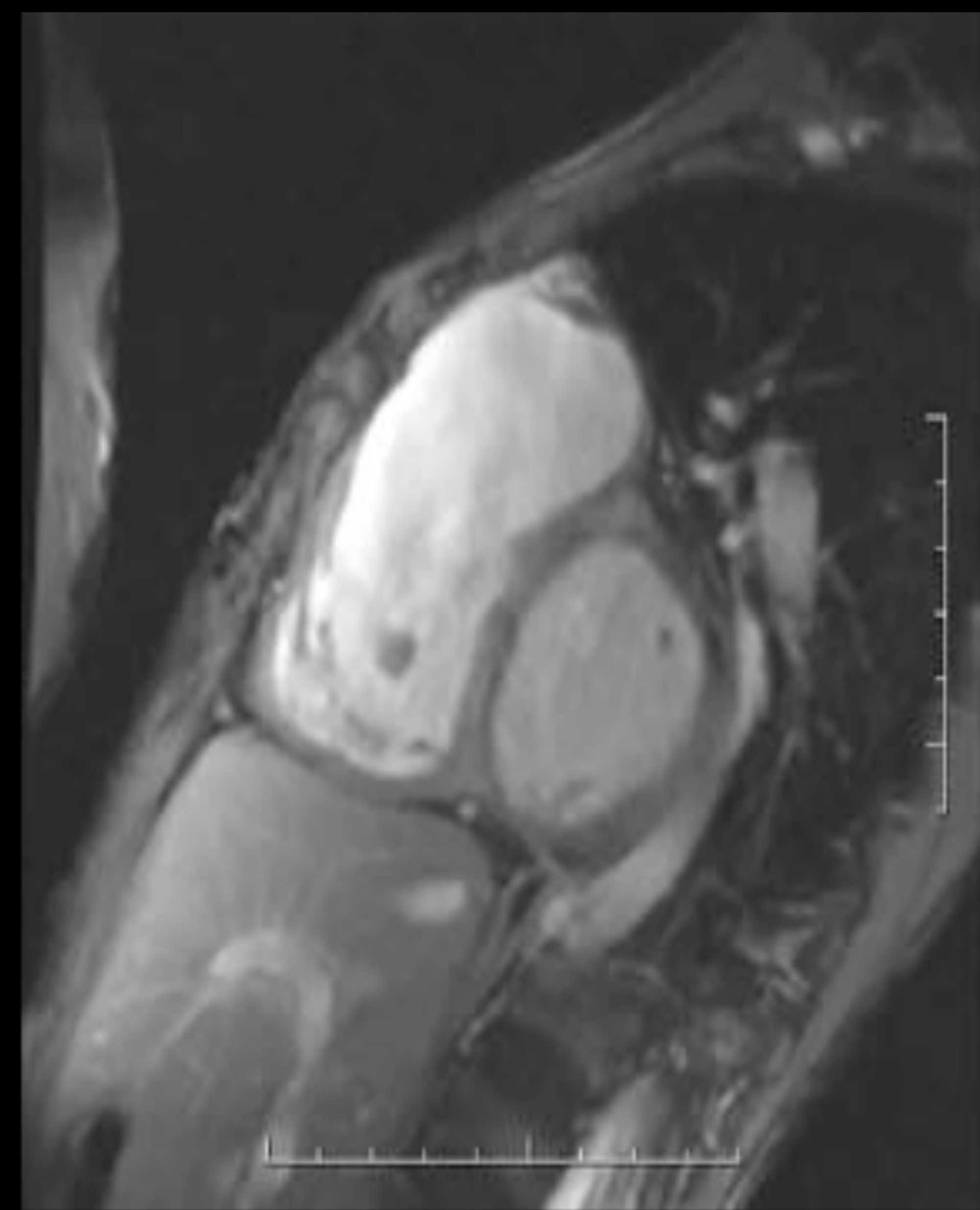
Valve pulmonaire  
«Melody»



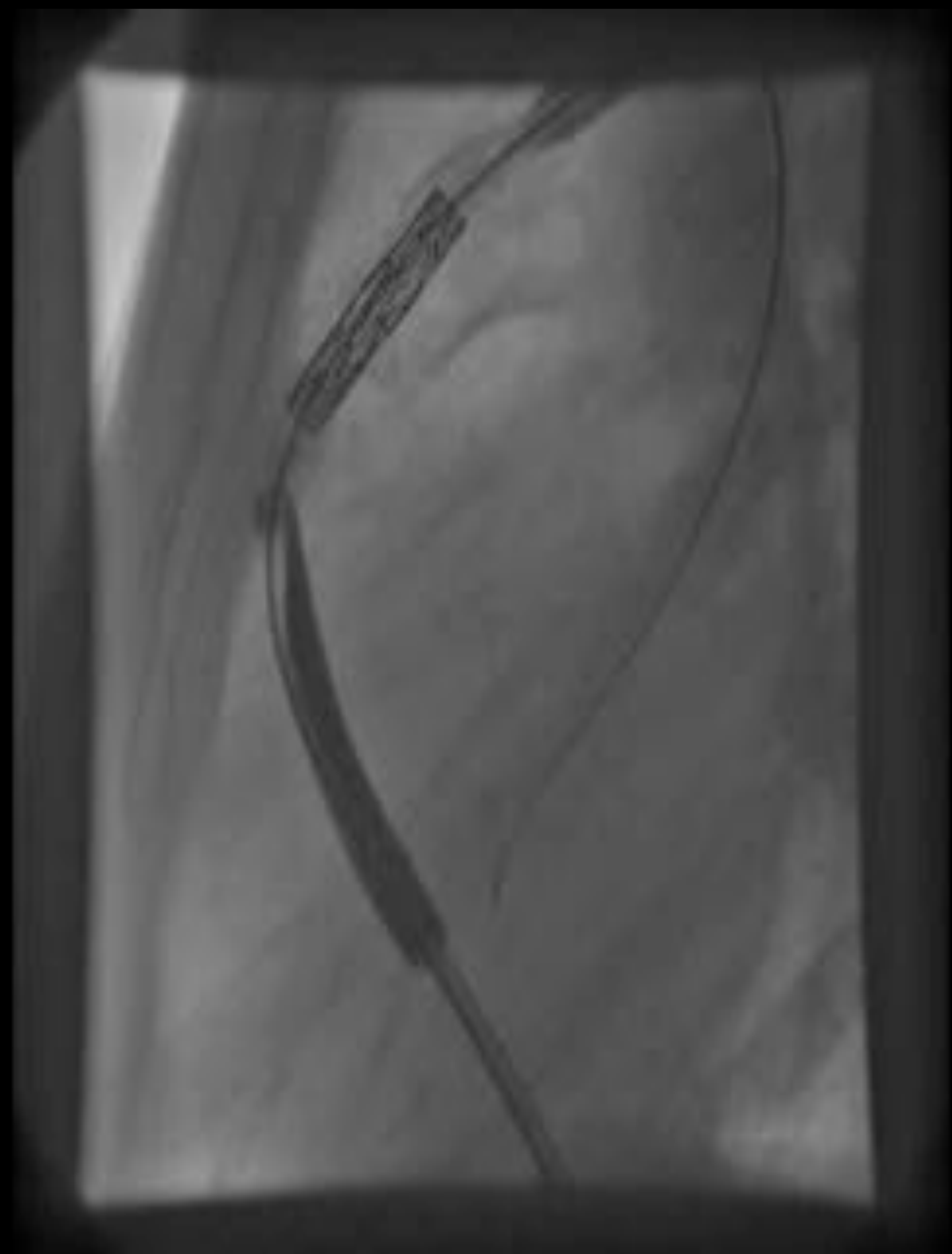
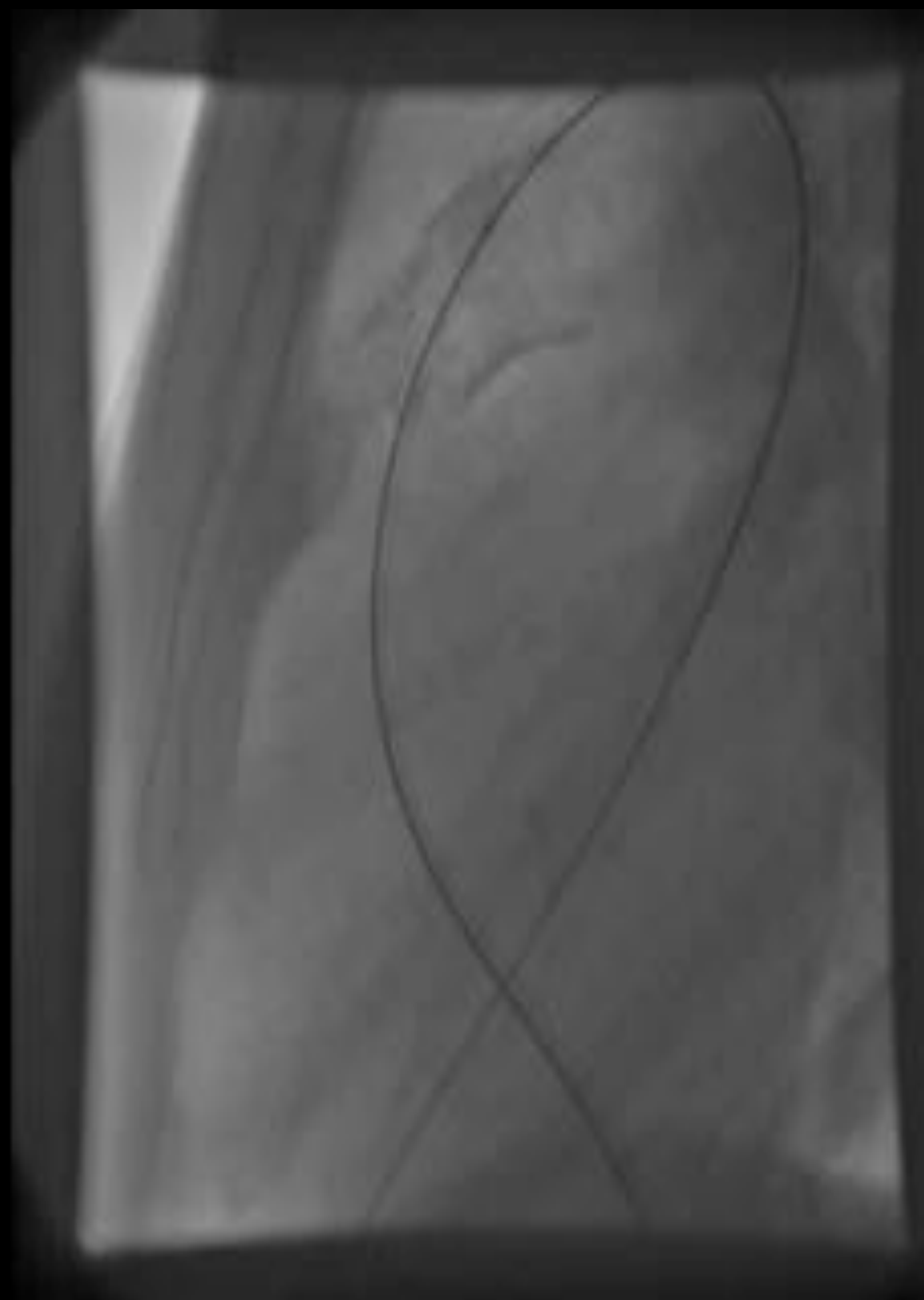
Valve injectable  
«Shelhigh»

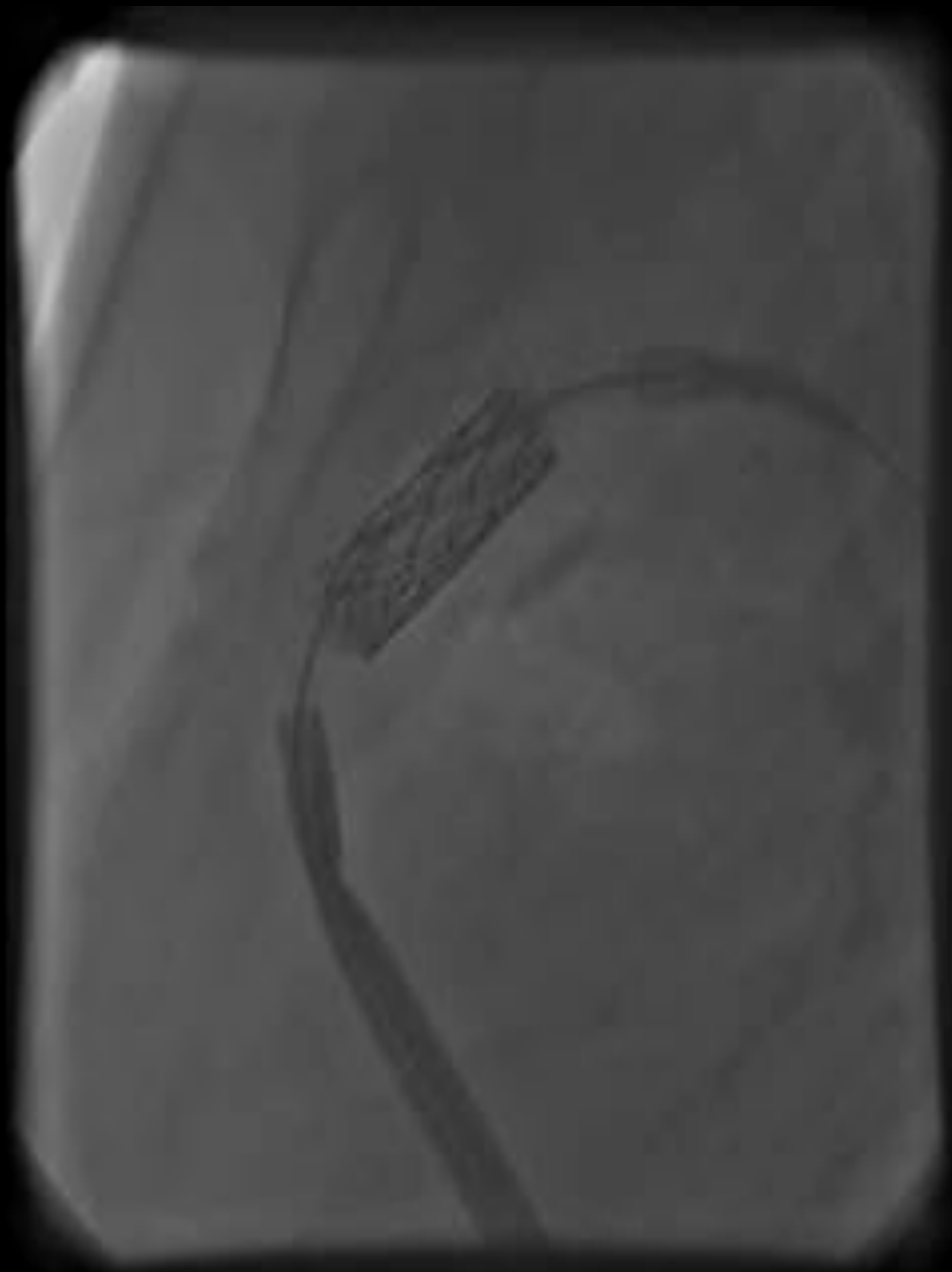




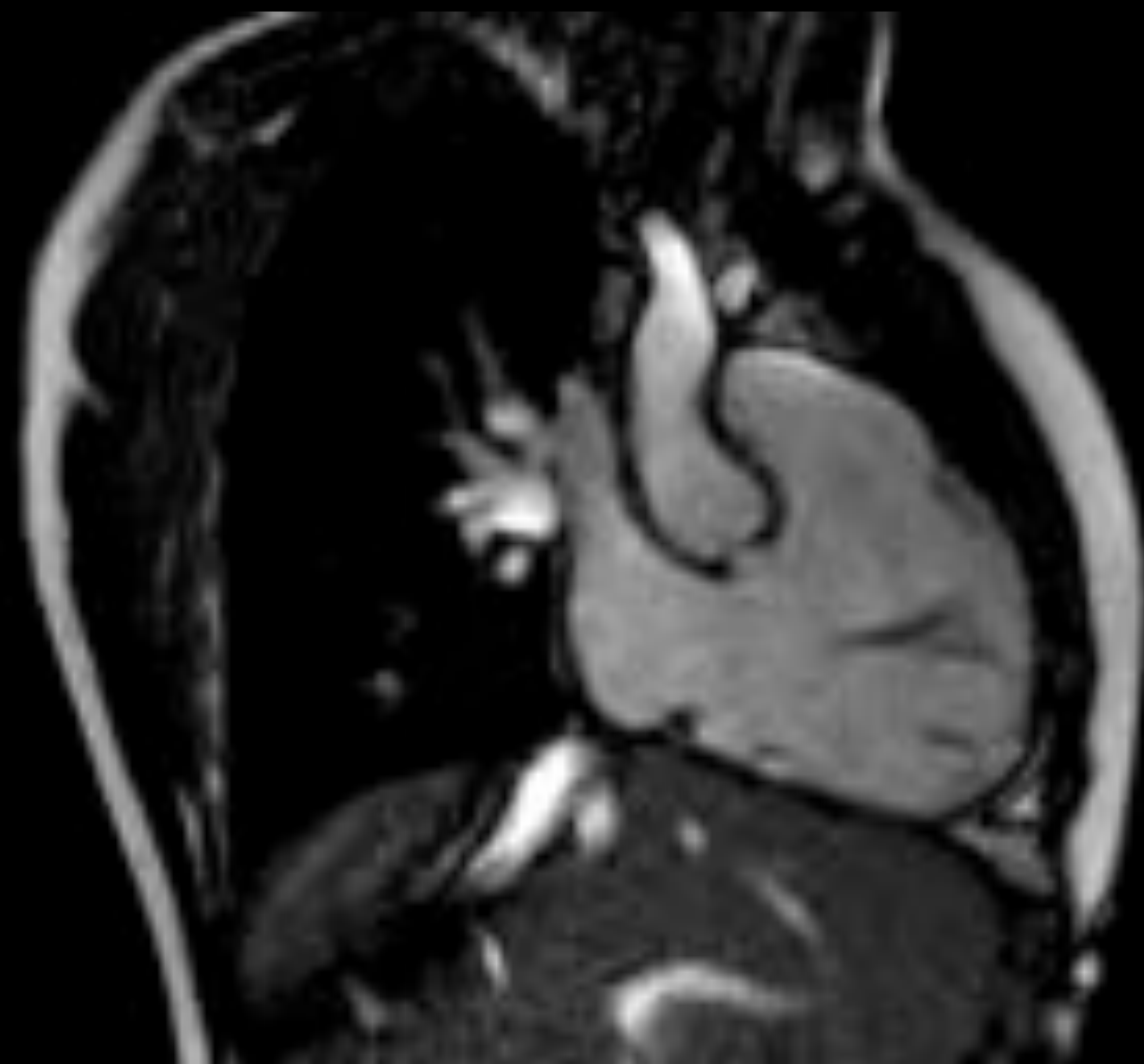
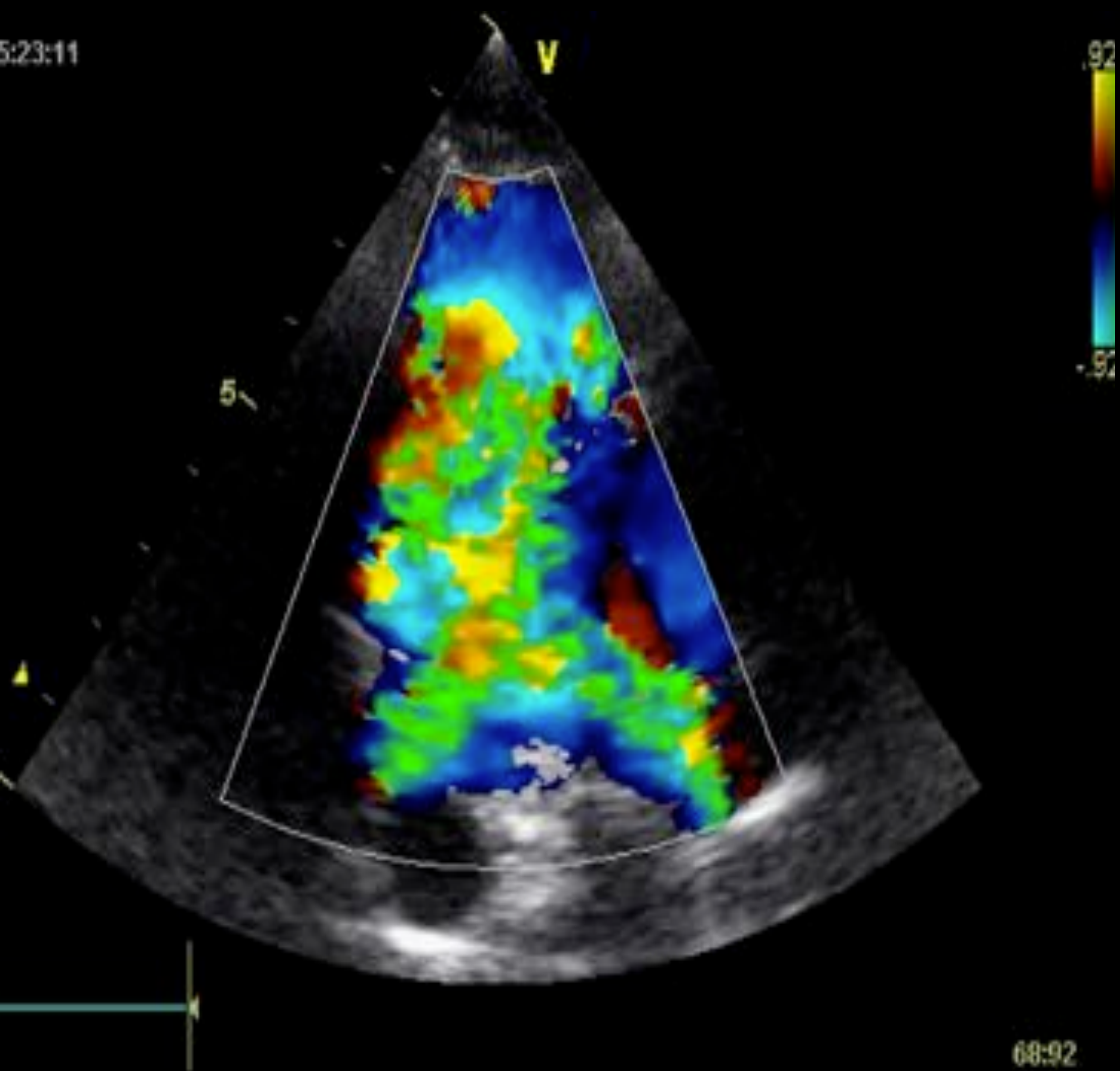




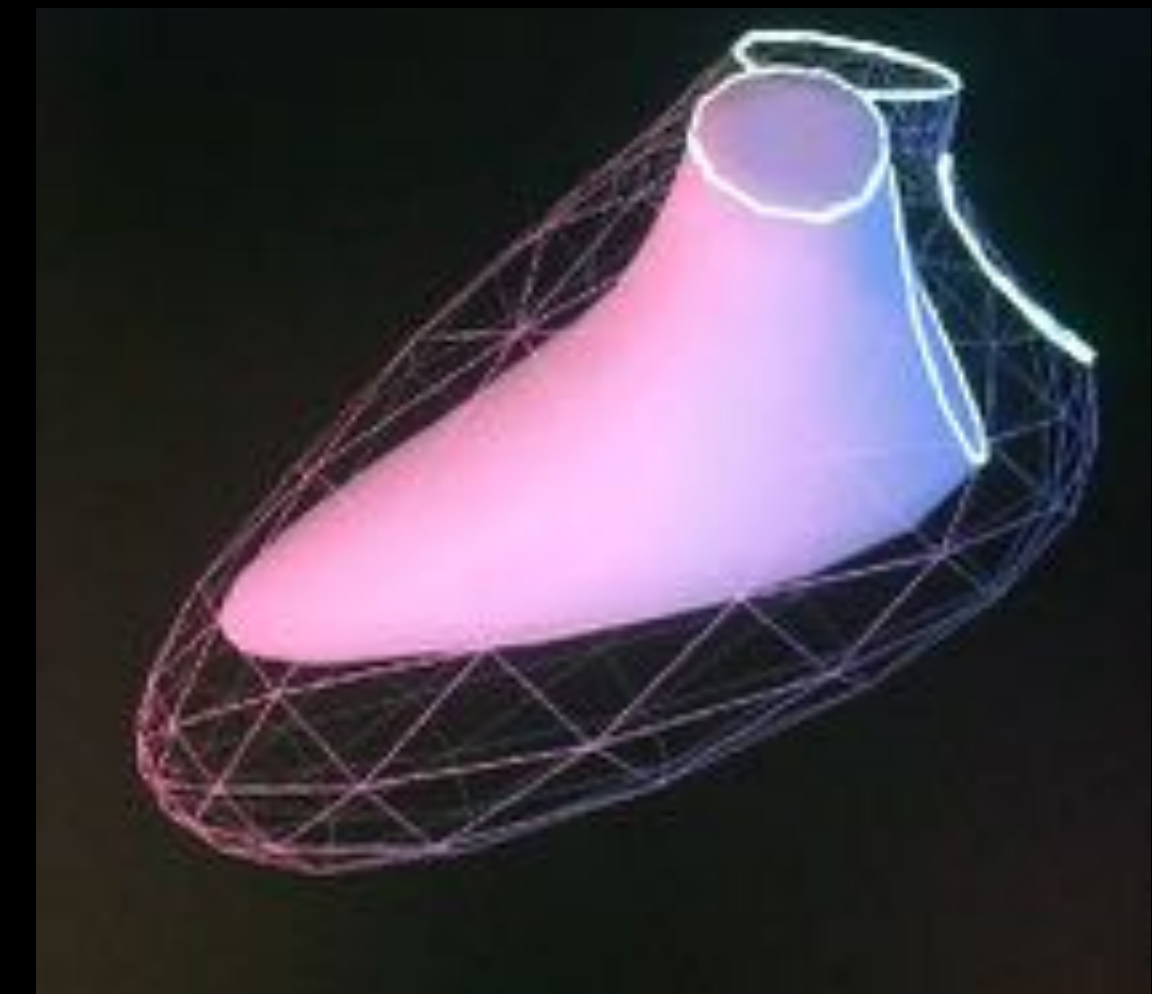
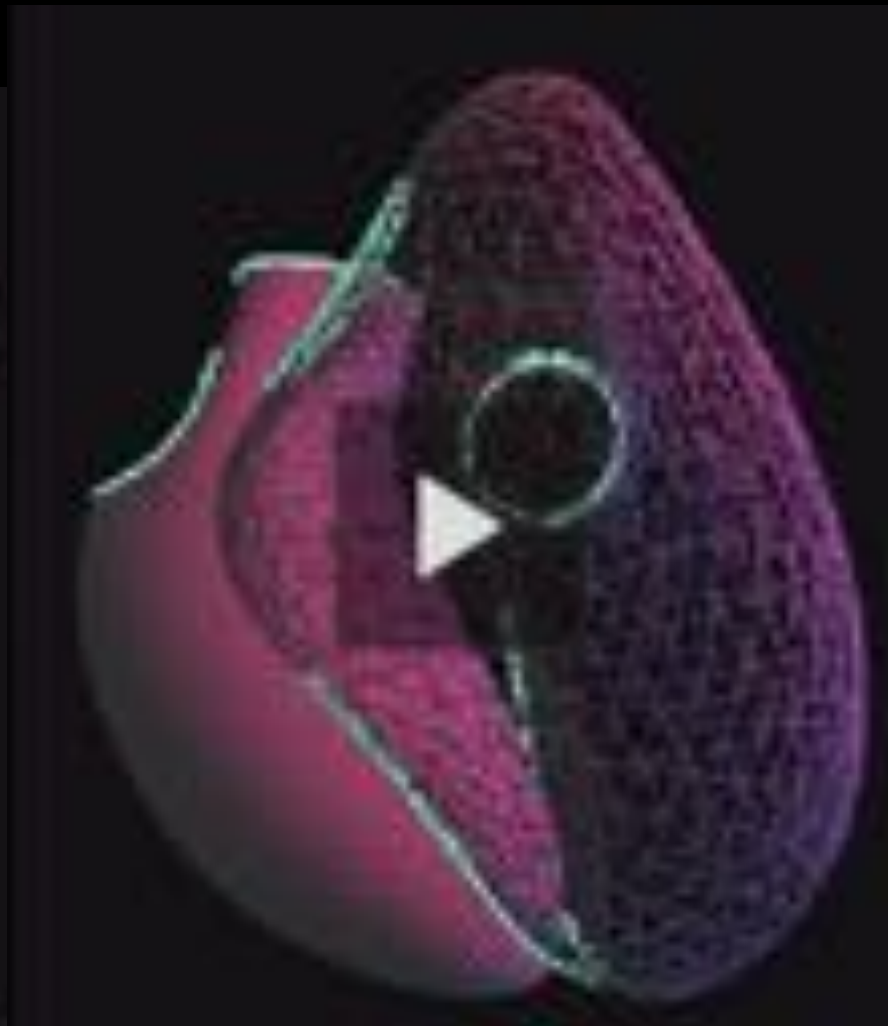
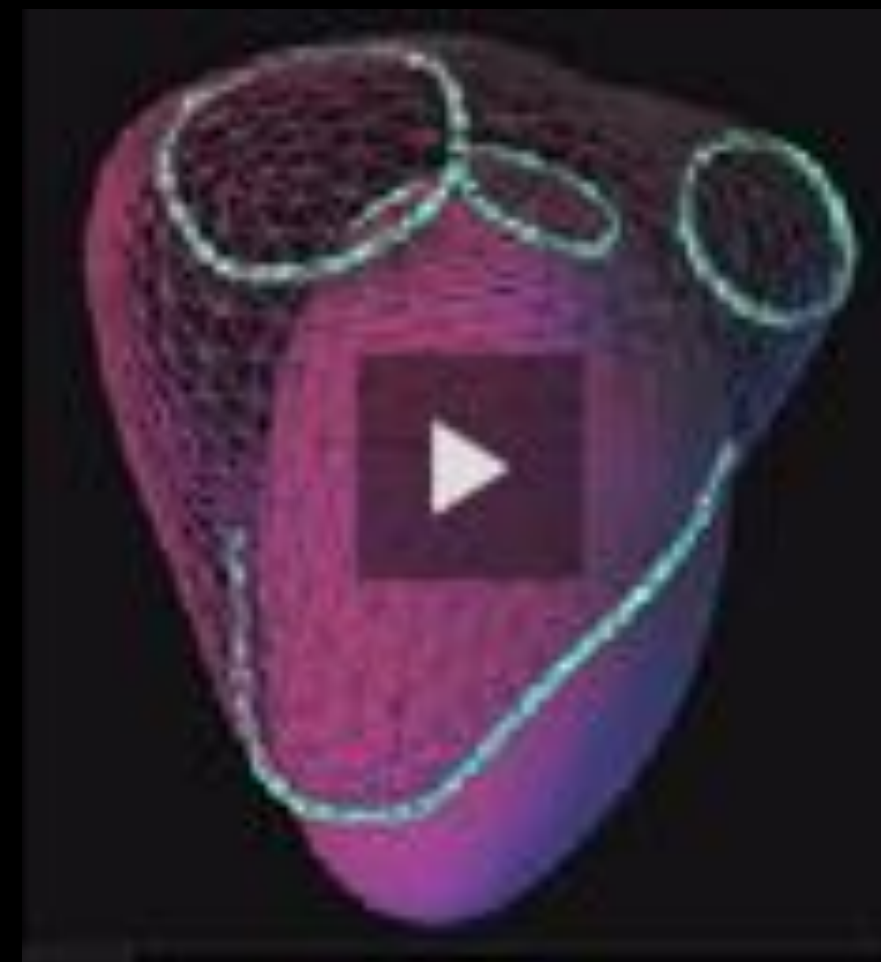
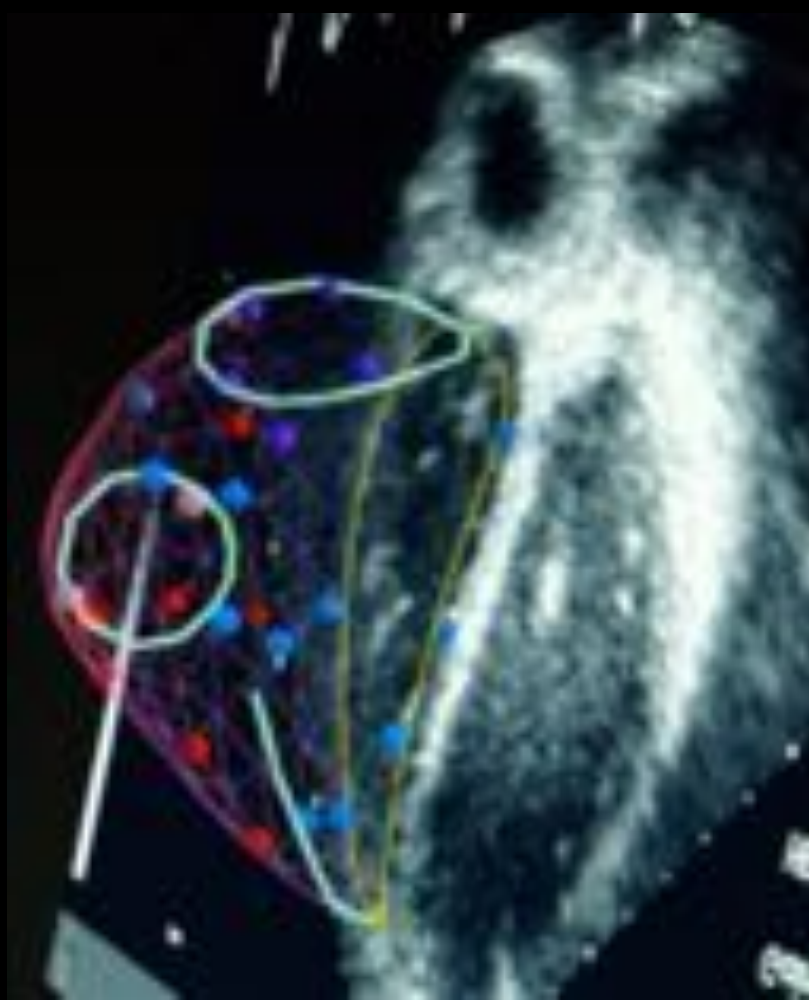
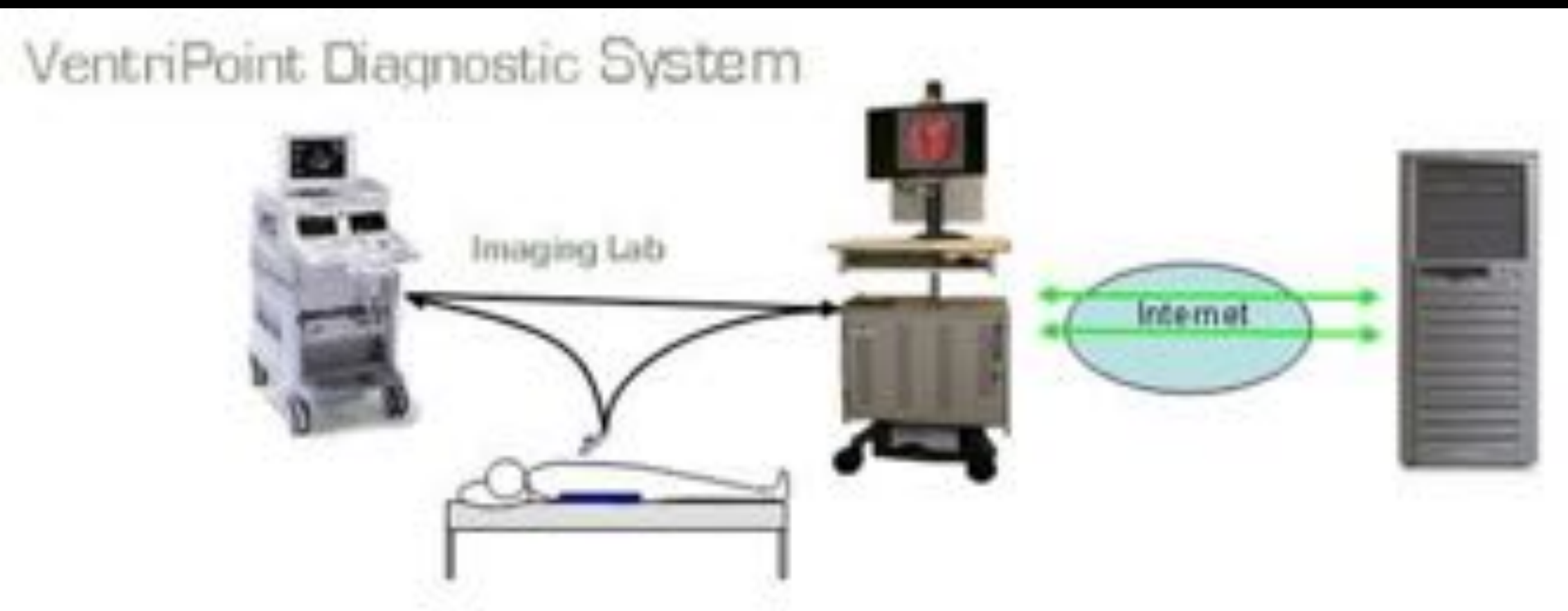




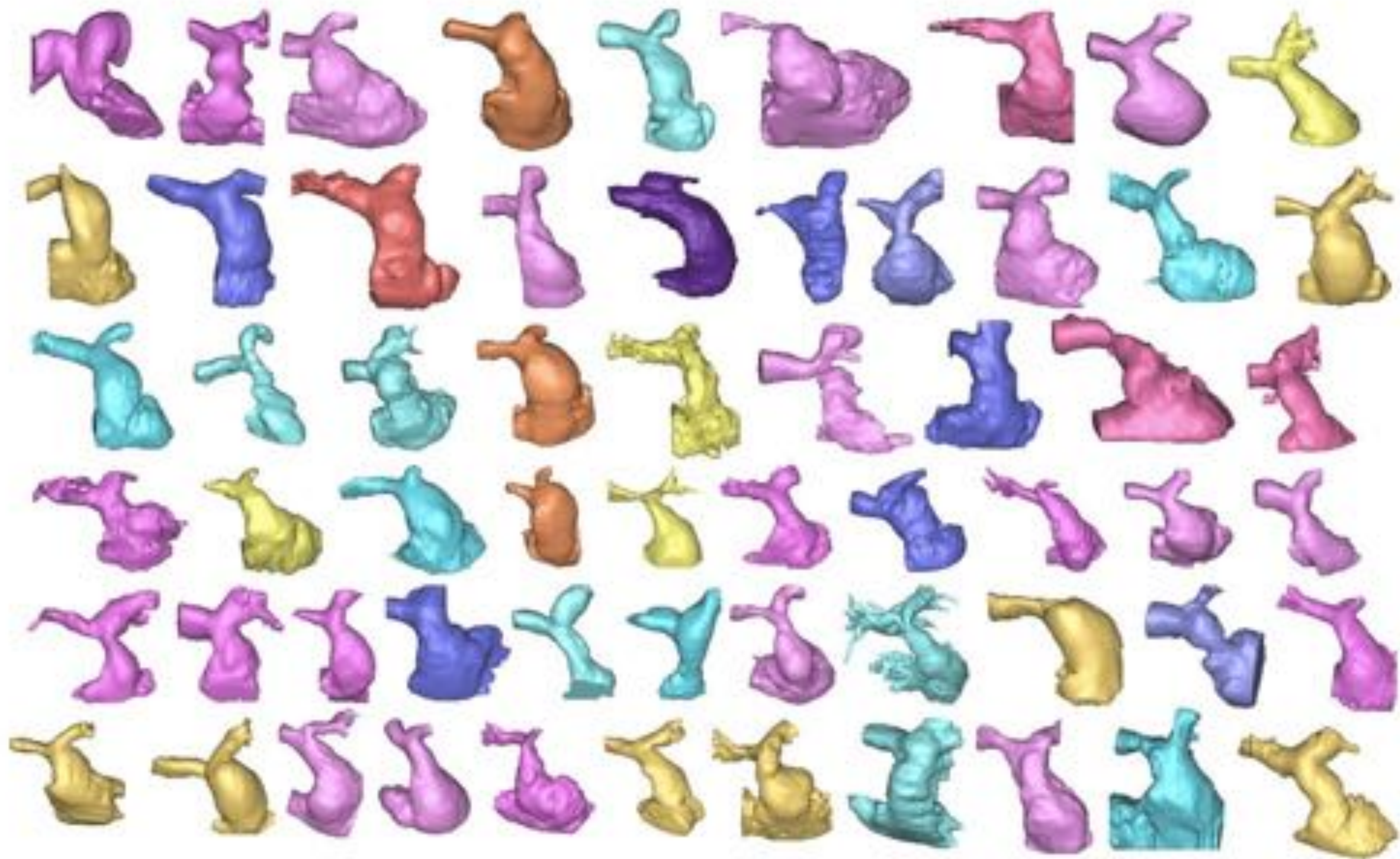
5:23:11

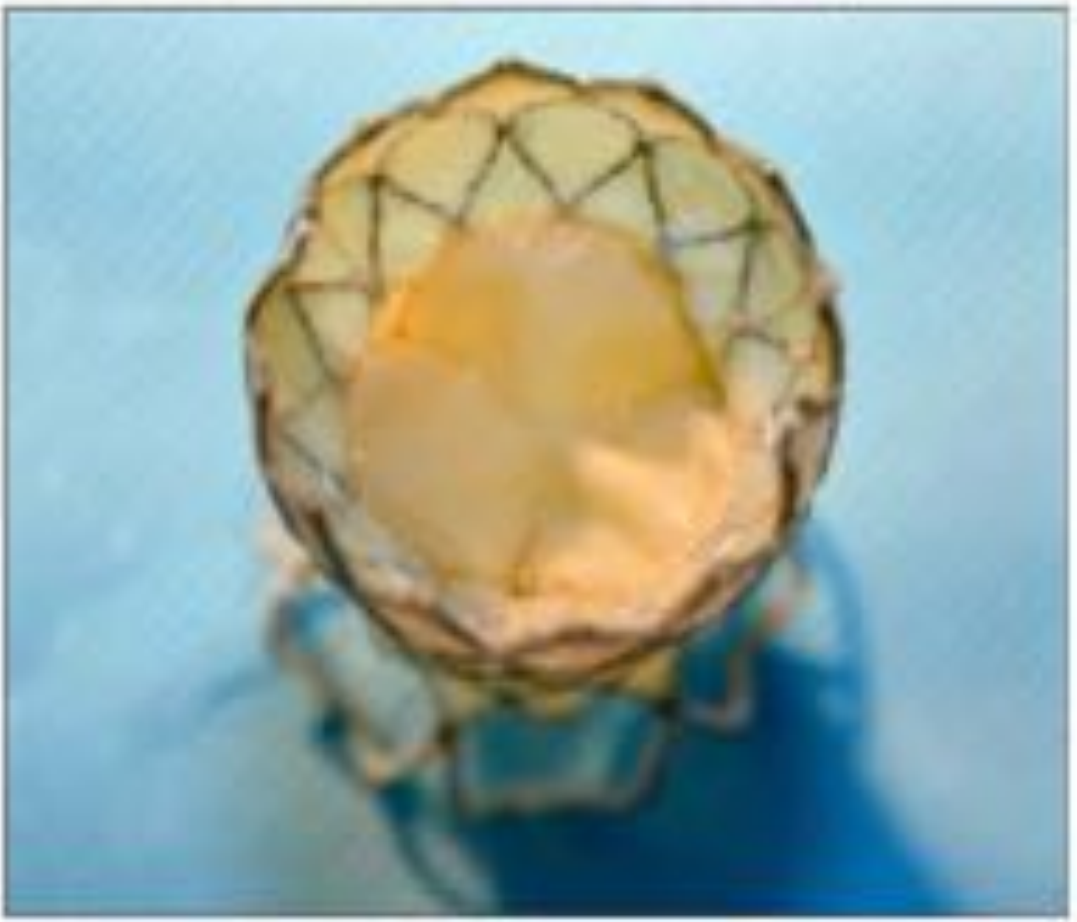
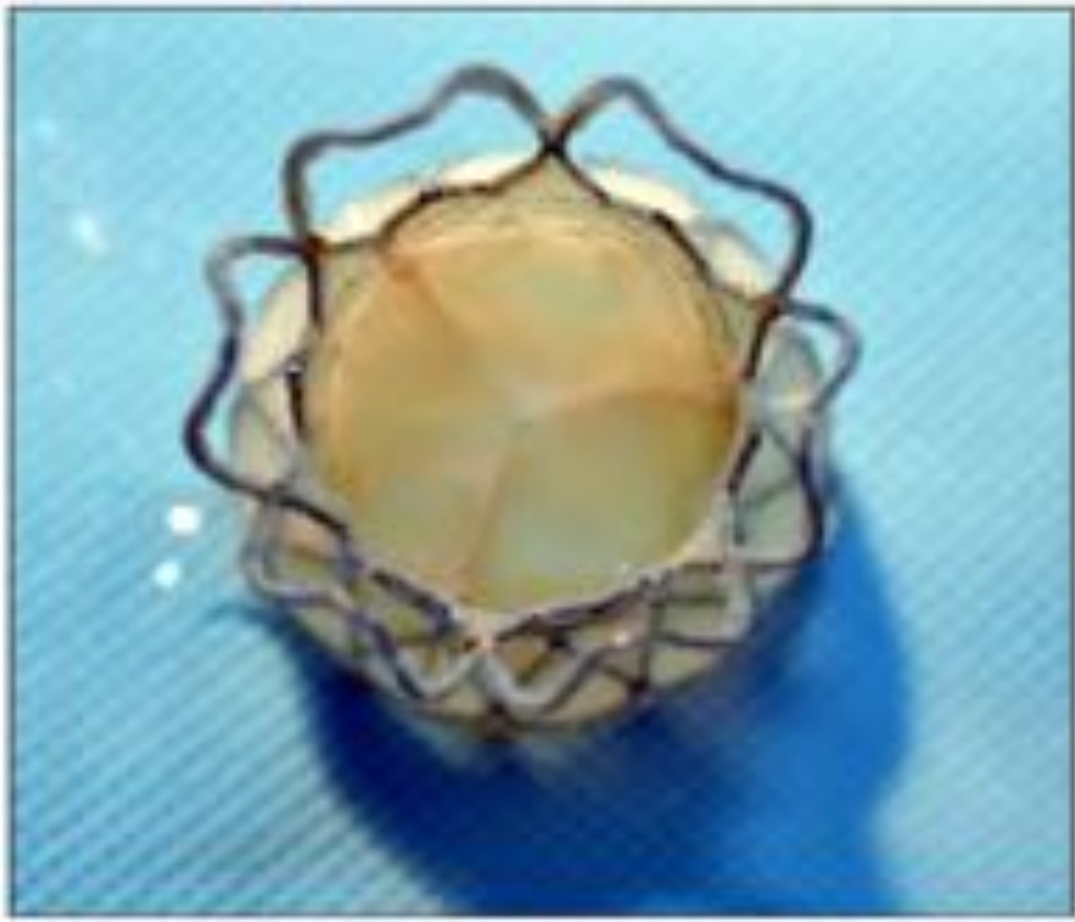
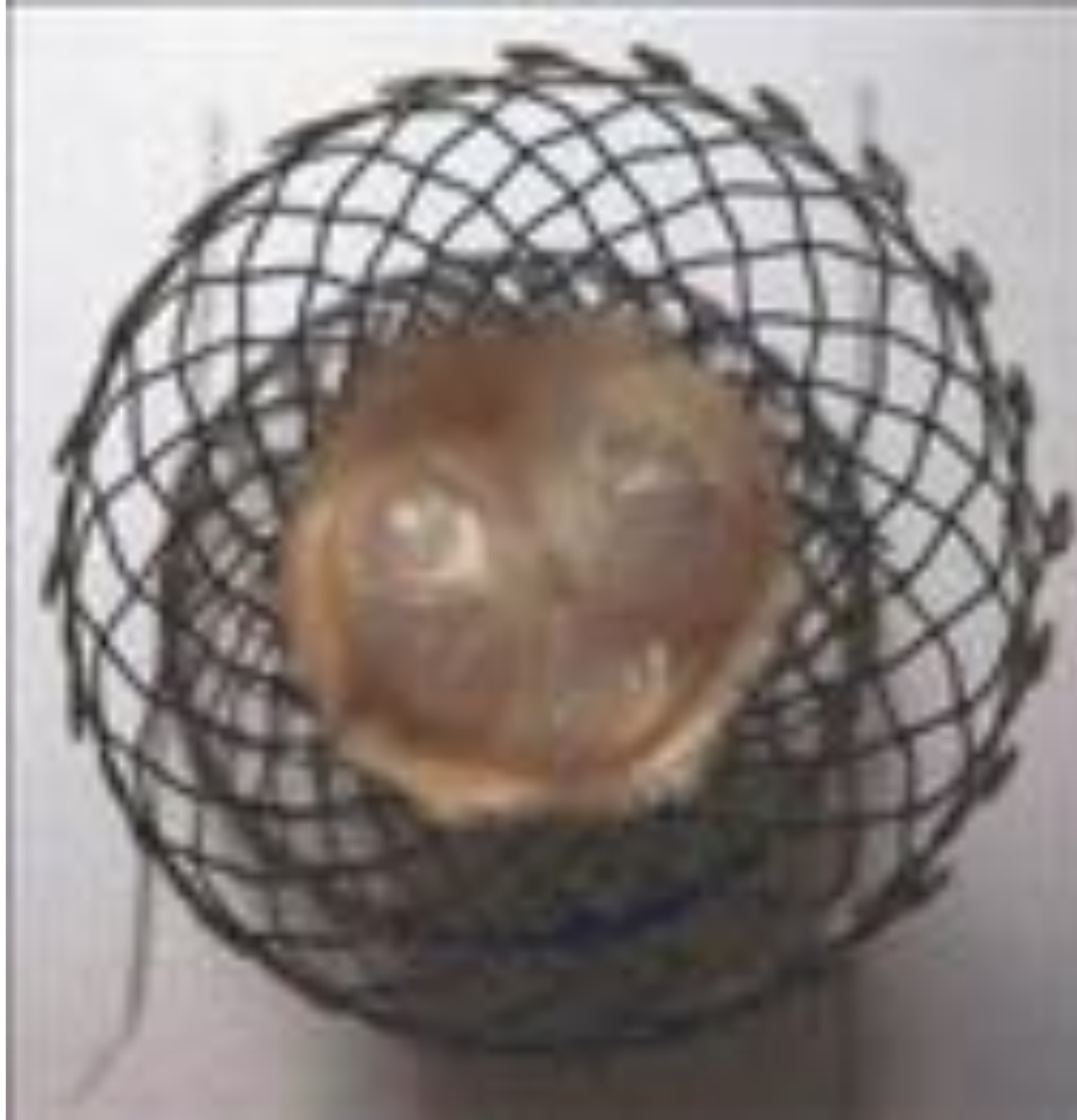
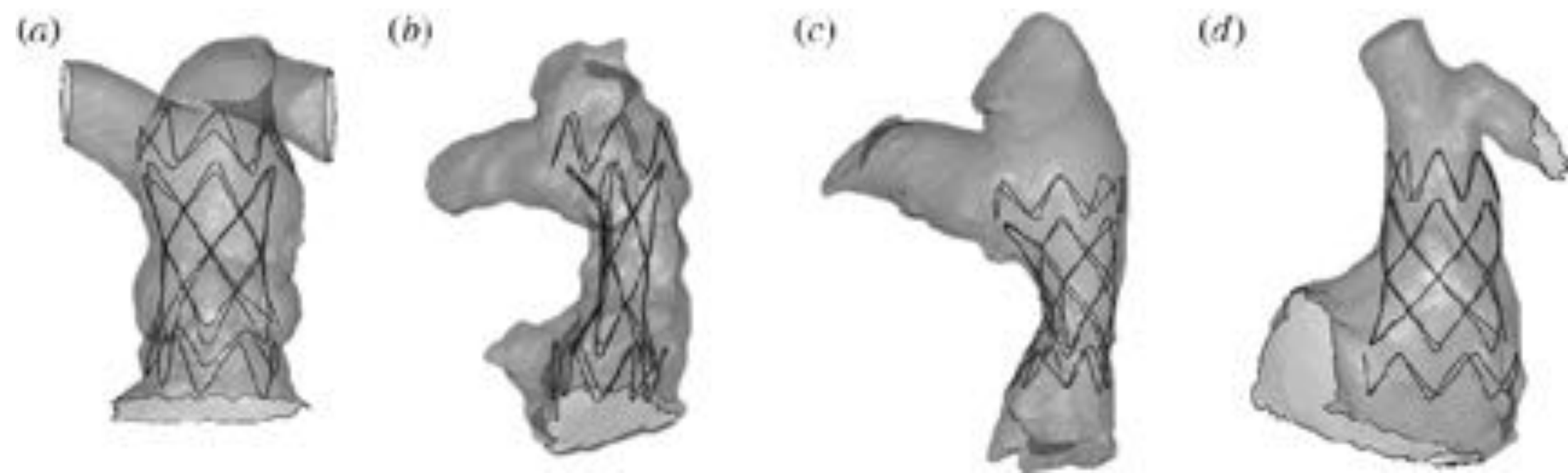


68:92



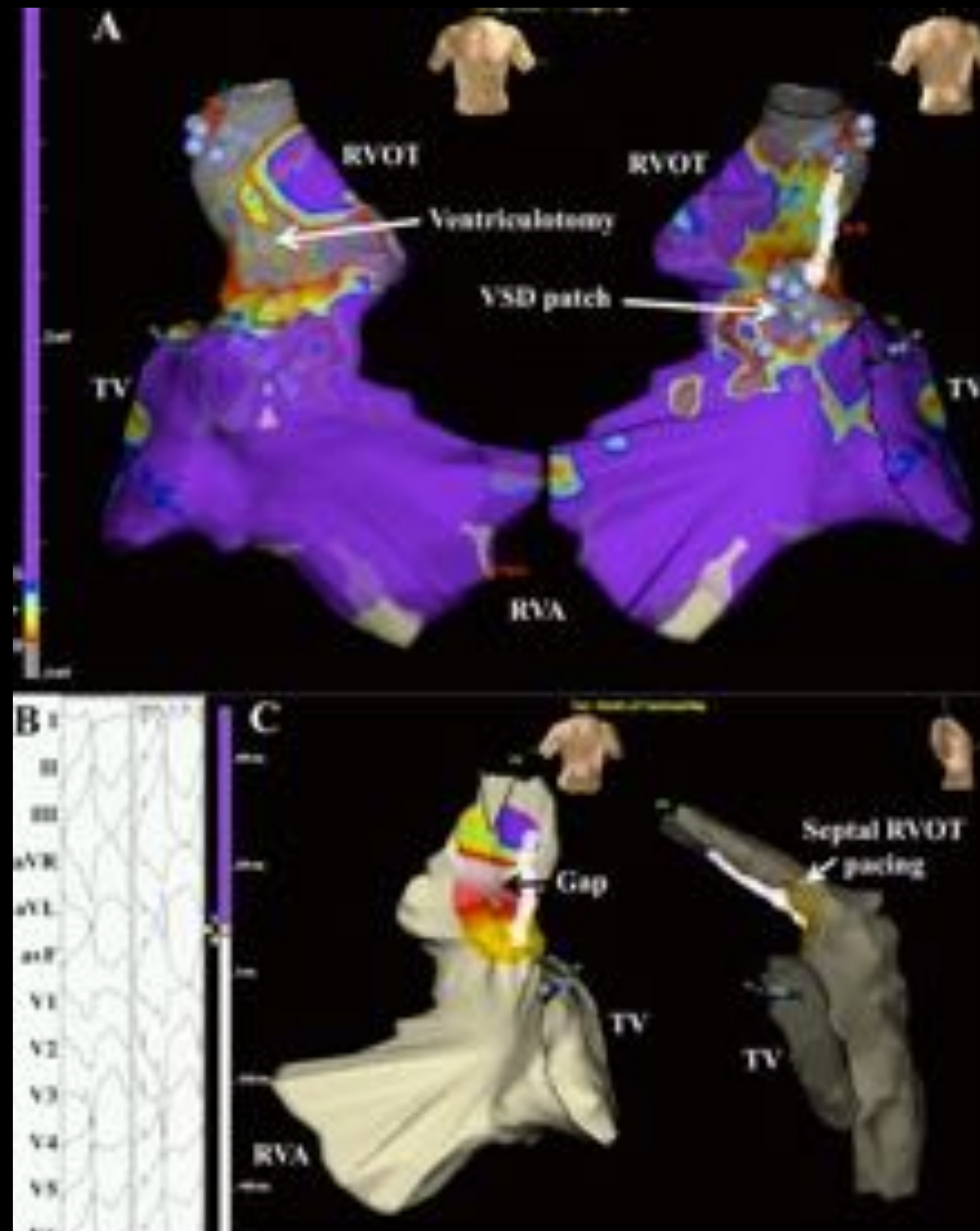
3D Knowledge system



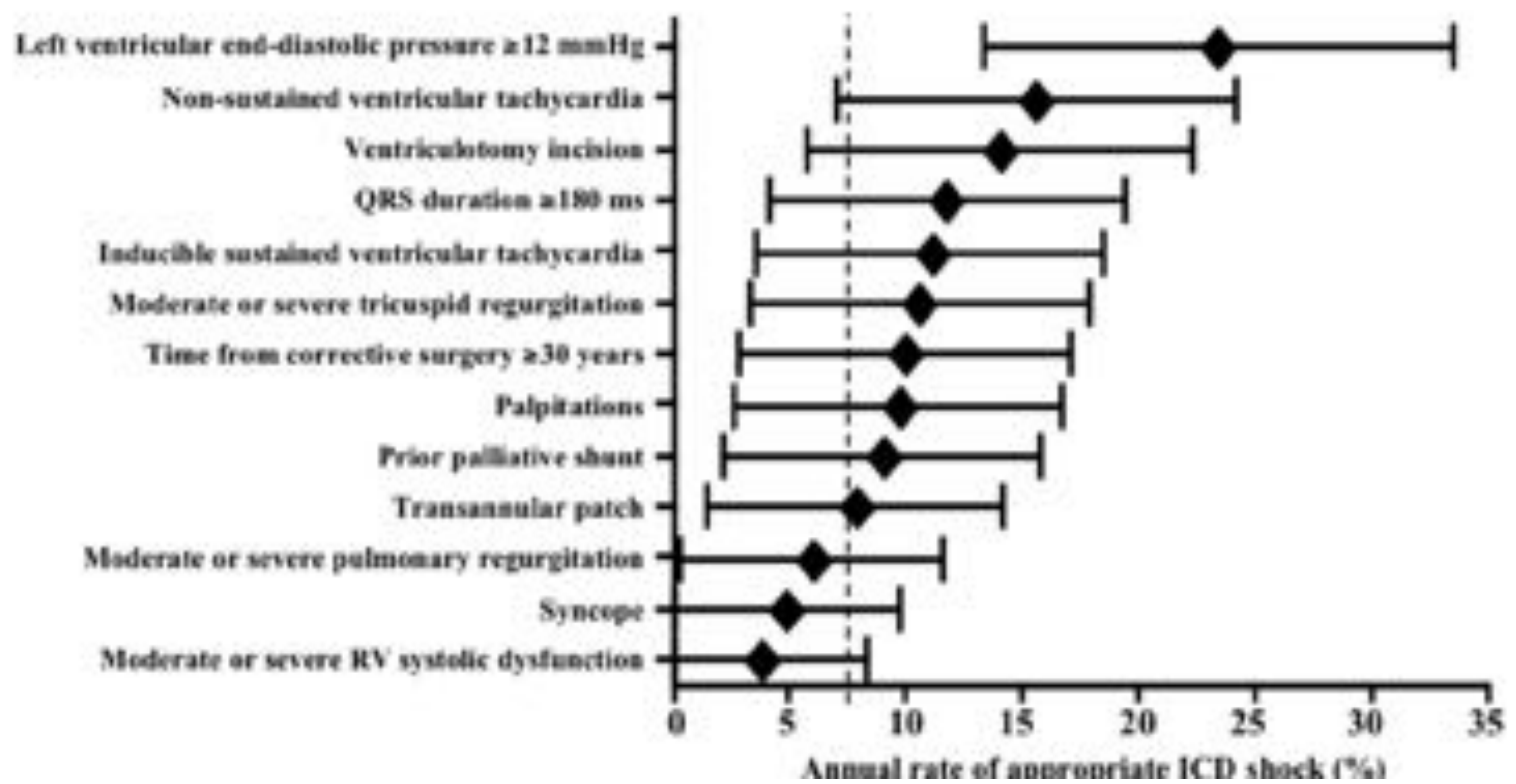


Indications	Class <sup>a</sup>	Level <sup>b</sup>
Aortic valve replacement should be performed in patients with severe AR with symptoms or signs of LV dysfunction	I	C
PVRep should be performed in symptomatic patients with severe PR and/or stenosis (RV systolic pressure >60 mmHg, TR velocity >3.5 m/s)	I	C
<p>PVRep should be considered in asymptomatic patients with severe PR and/or PS when at least one of the following criteria is present:</p> <ul style="list-style-type: none"> <li>• Decrease in objective exercise capacity</li> <li>• Progressive RV dilation</li> <li>• Progressive RV systolic dysfunction</li> <li>• Progressive TR (at least moderate)</li> <li>• RVOTD with RV systolic pressure &gt;80 mmHg (TR velocity &gt;4.3 m/s)</li> <li>• Sustained atrial/ventricular arrhythmias</li> </ul>	IIa	C
VSD closure should be considered in patients with residual VSD and significant LV volume overload or if the patient is undergoing pulmonary valve surgery	IIa	C

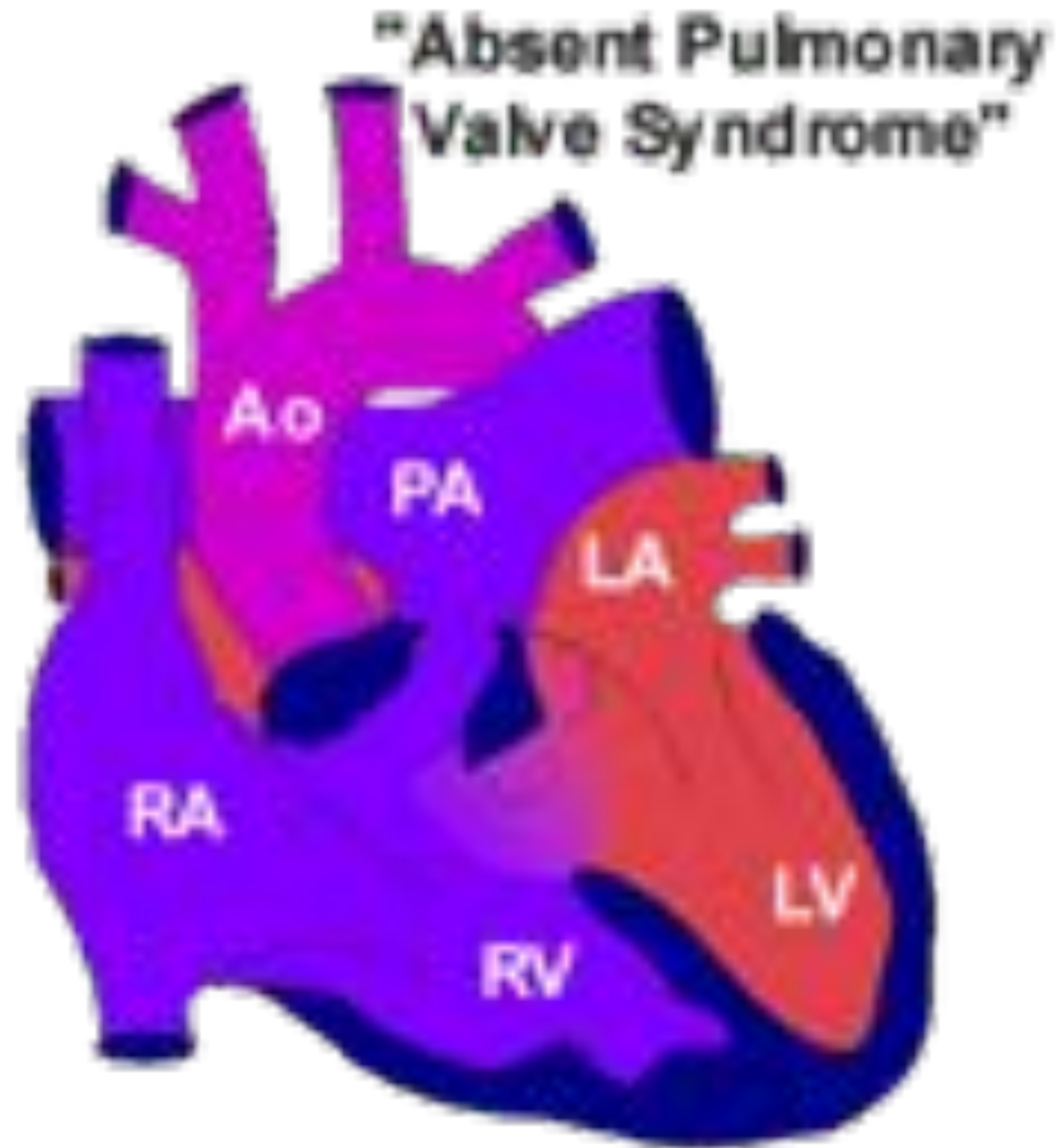
# Voltage and pace mapping of ventricular tachycardia in tetralogy of Fallot

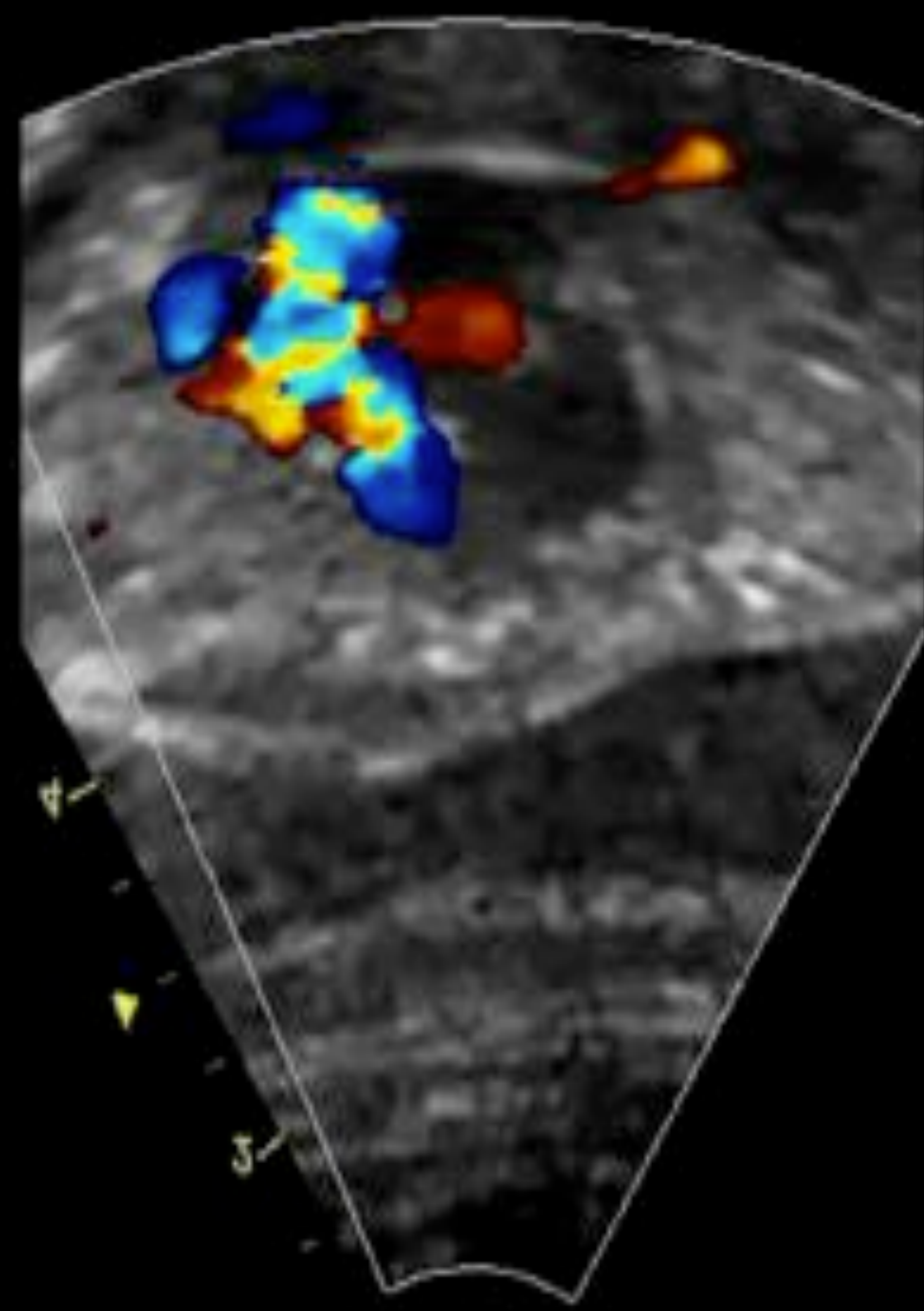
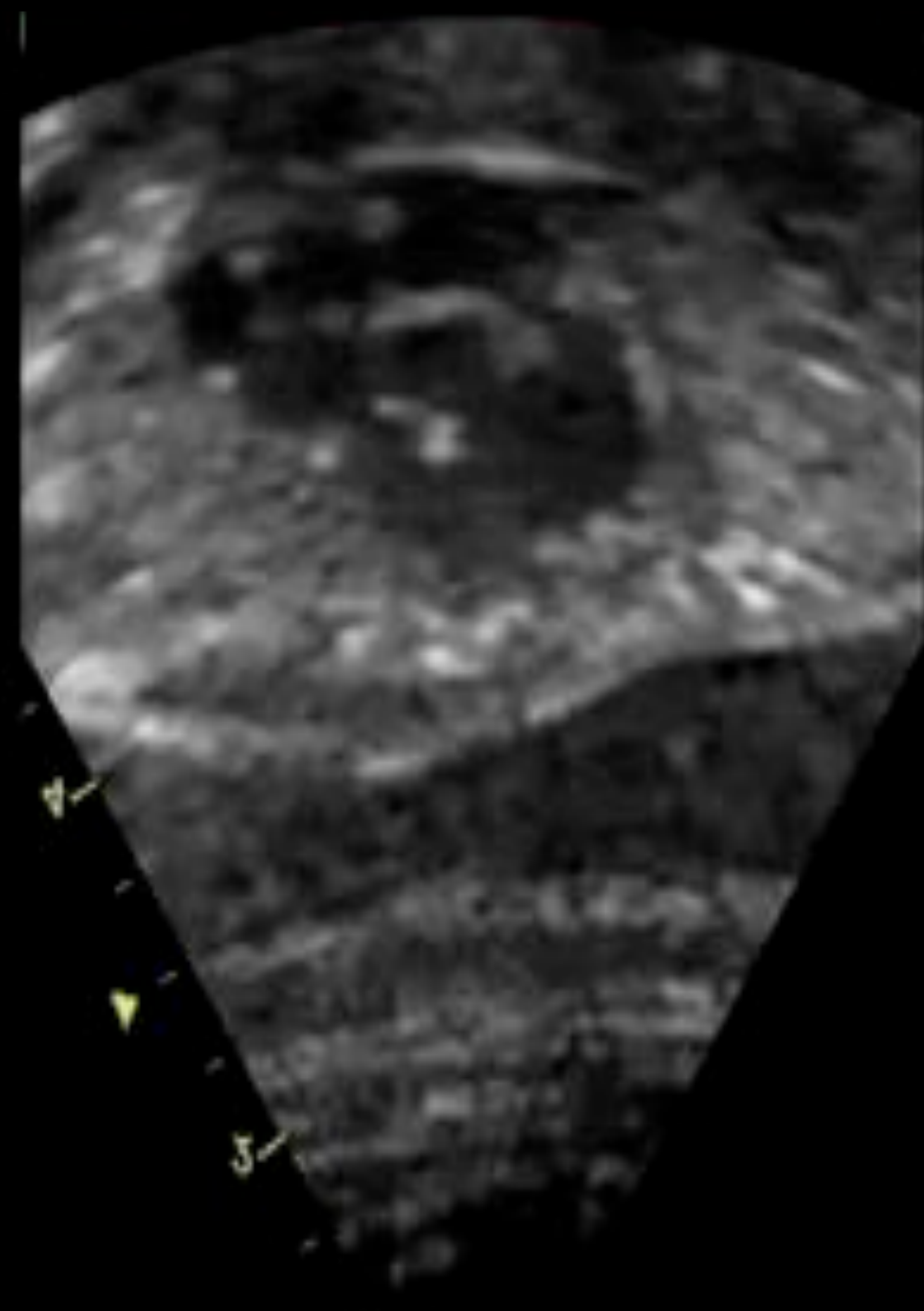


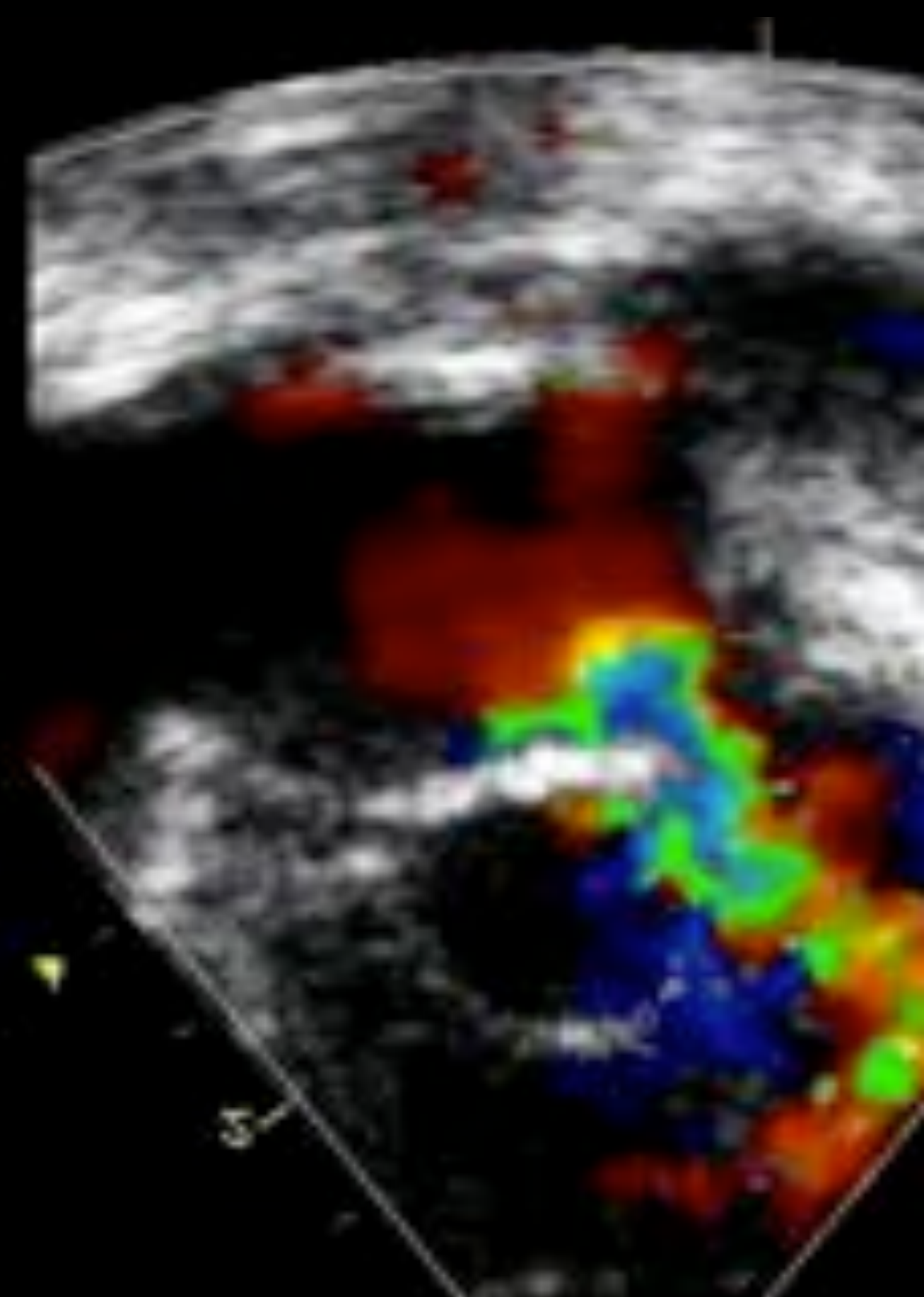
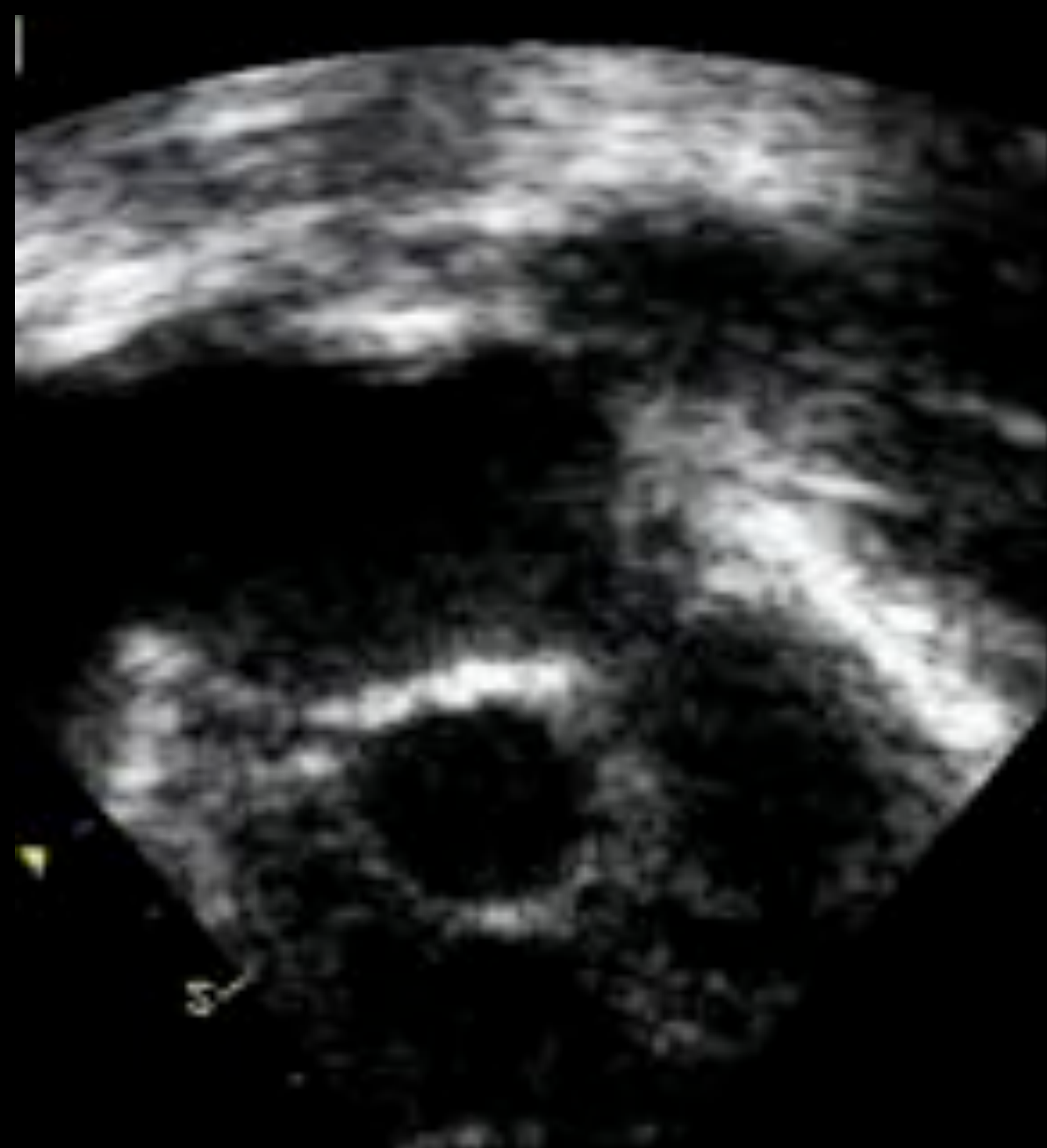


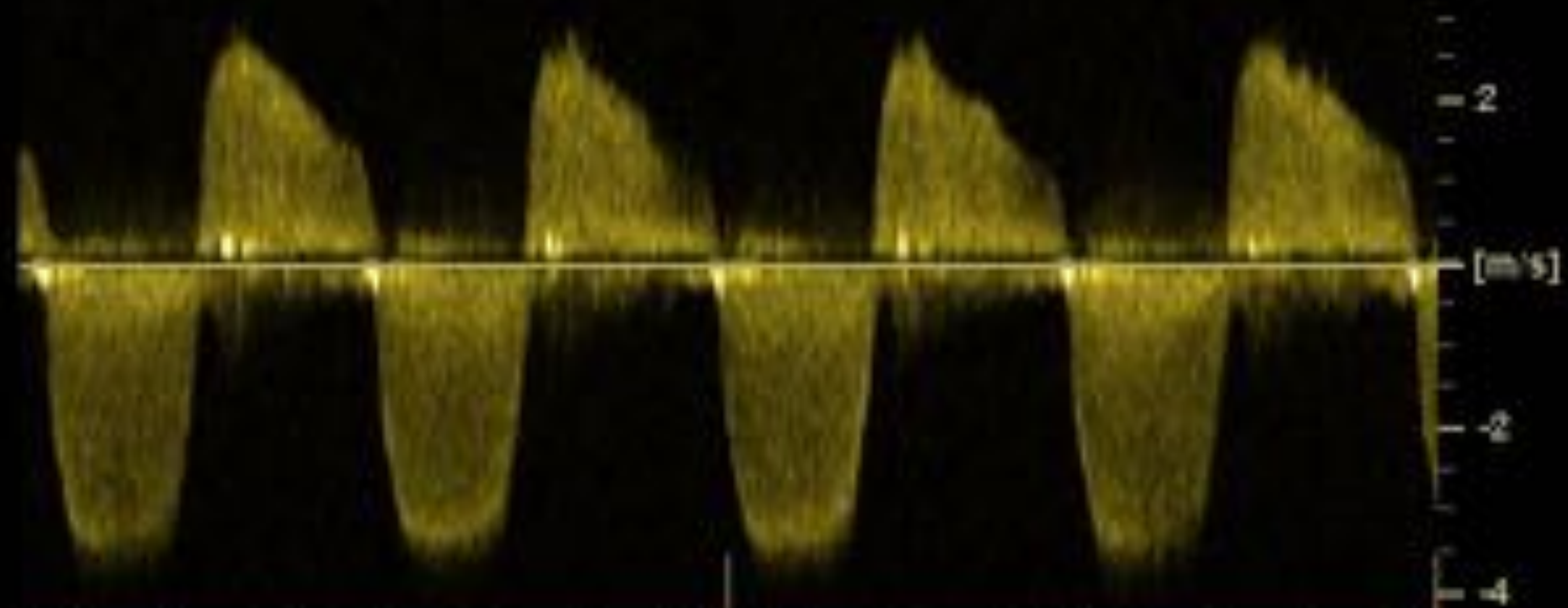
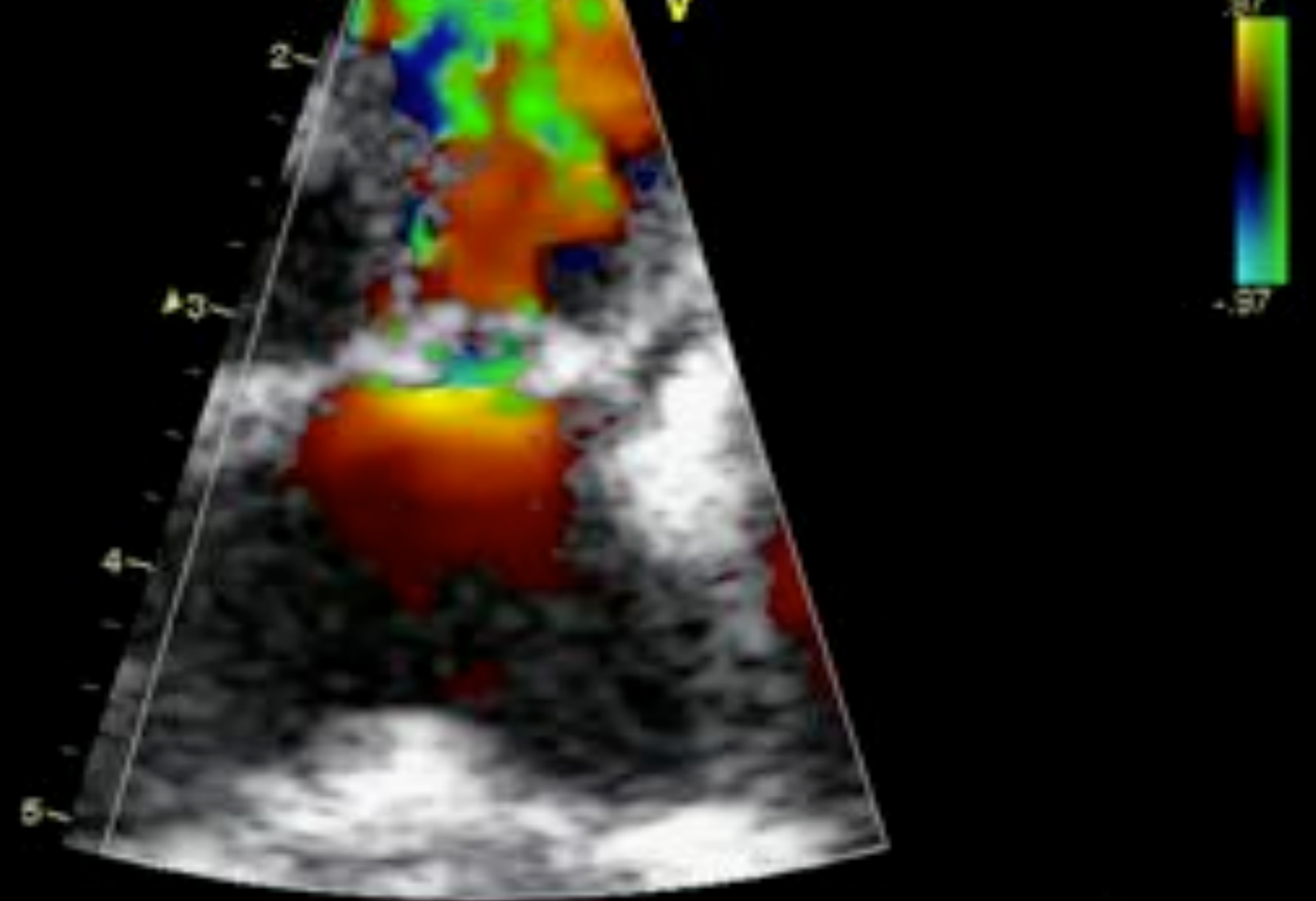


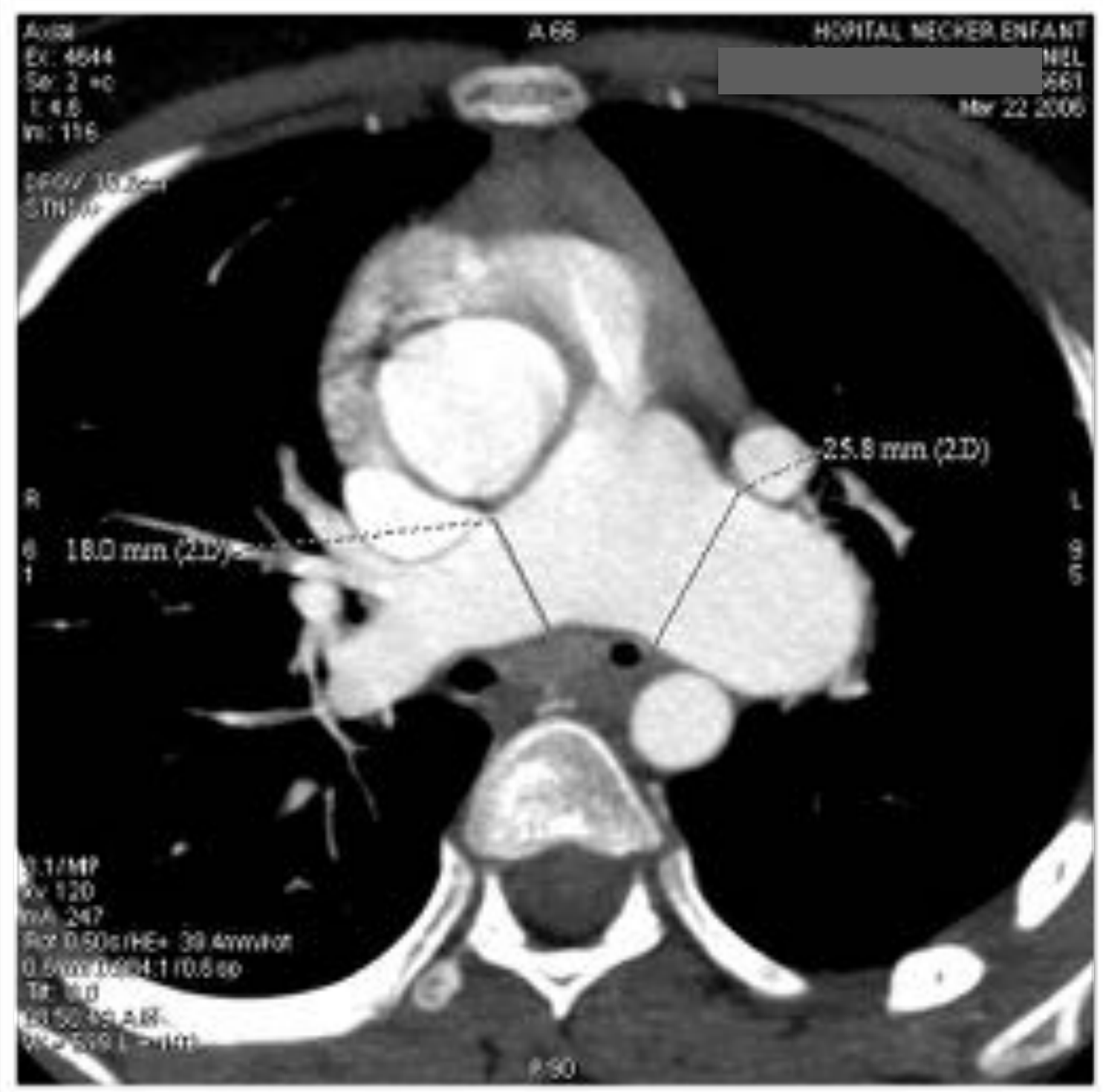
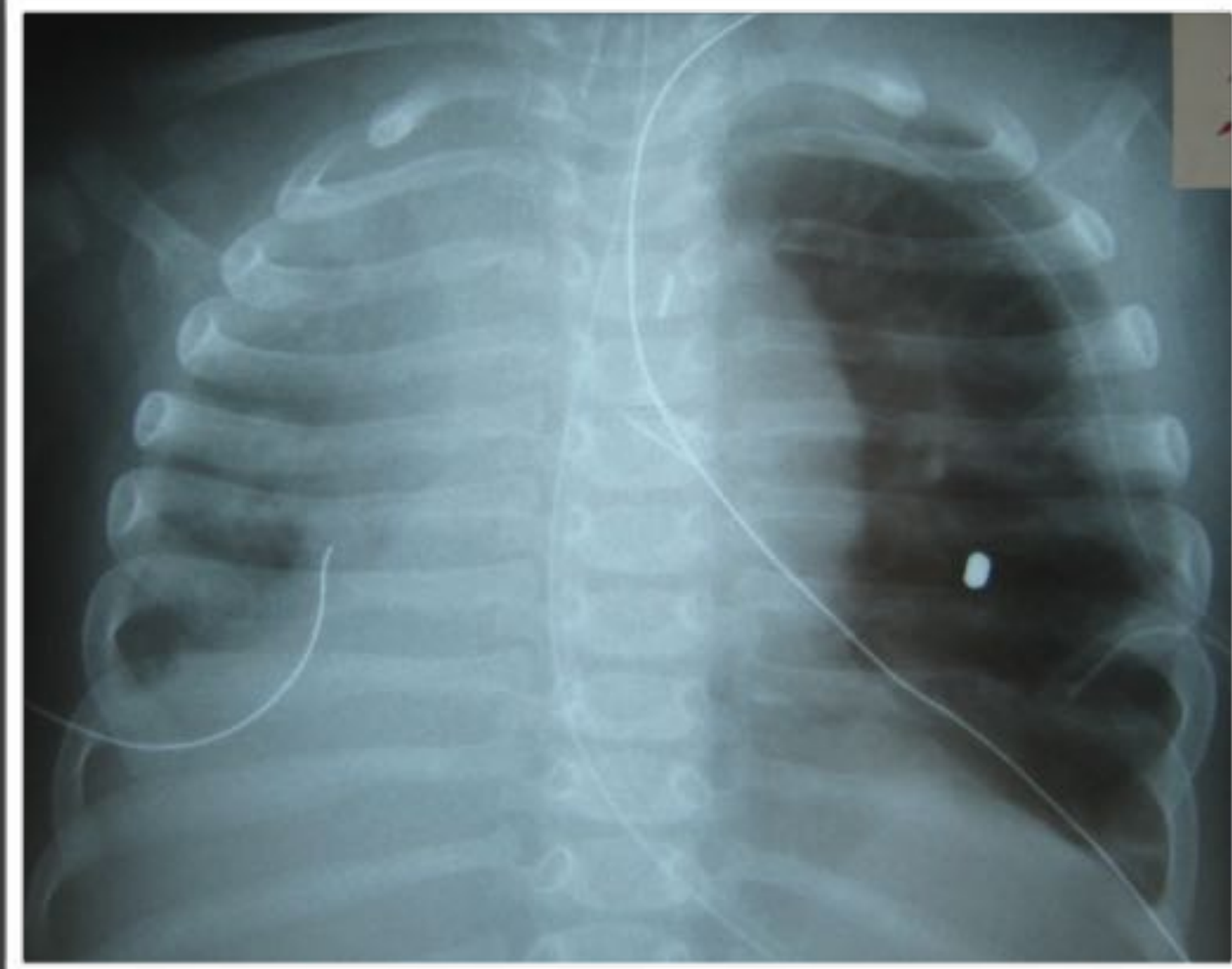
# Tetralogy of Fallot with absent pulmonary valve

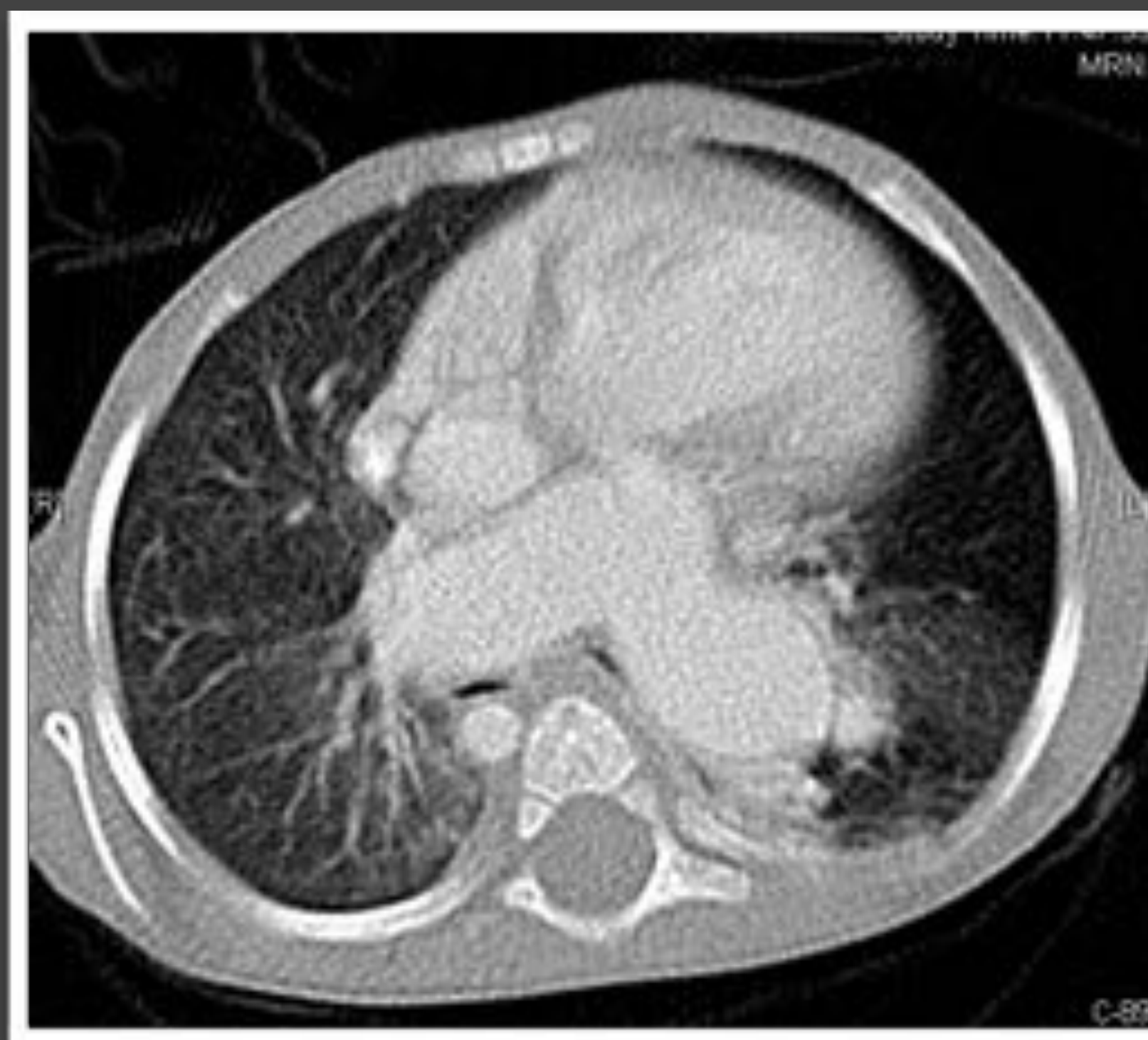
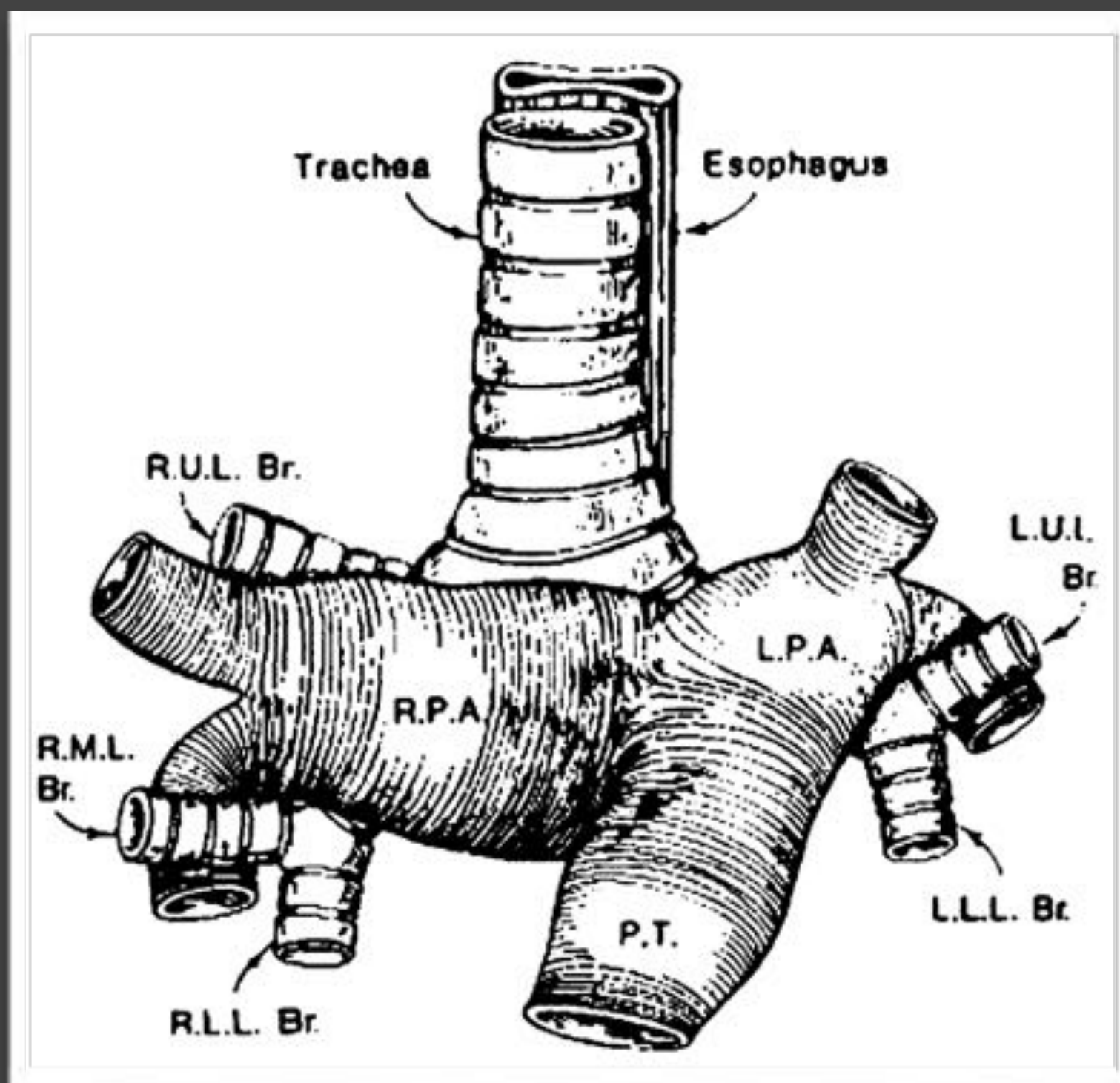




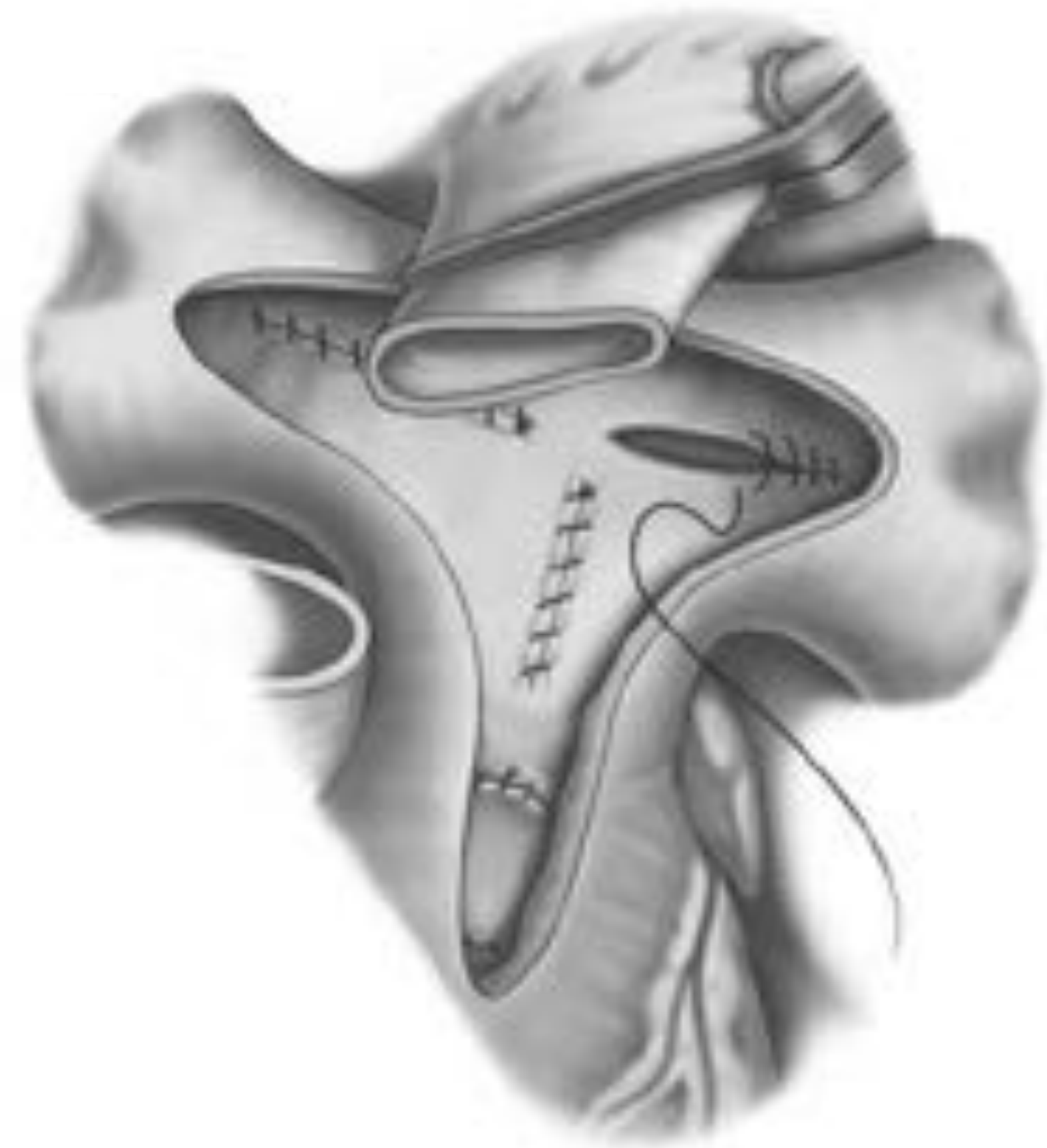
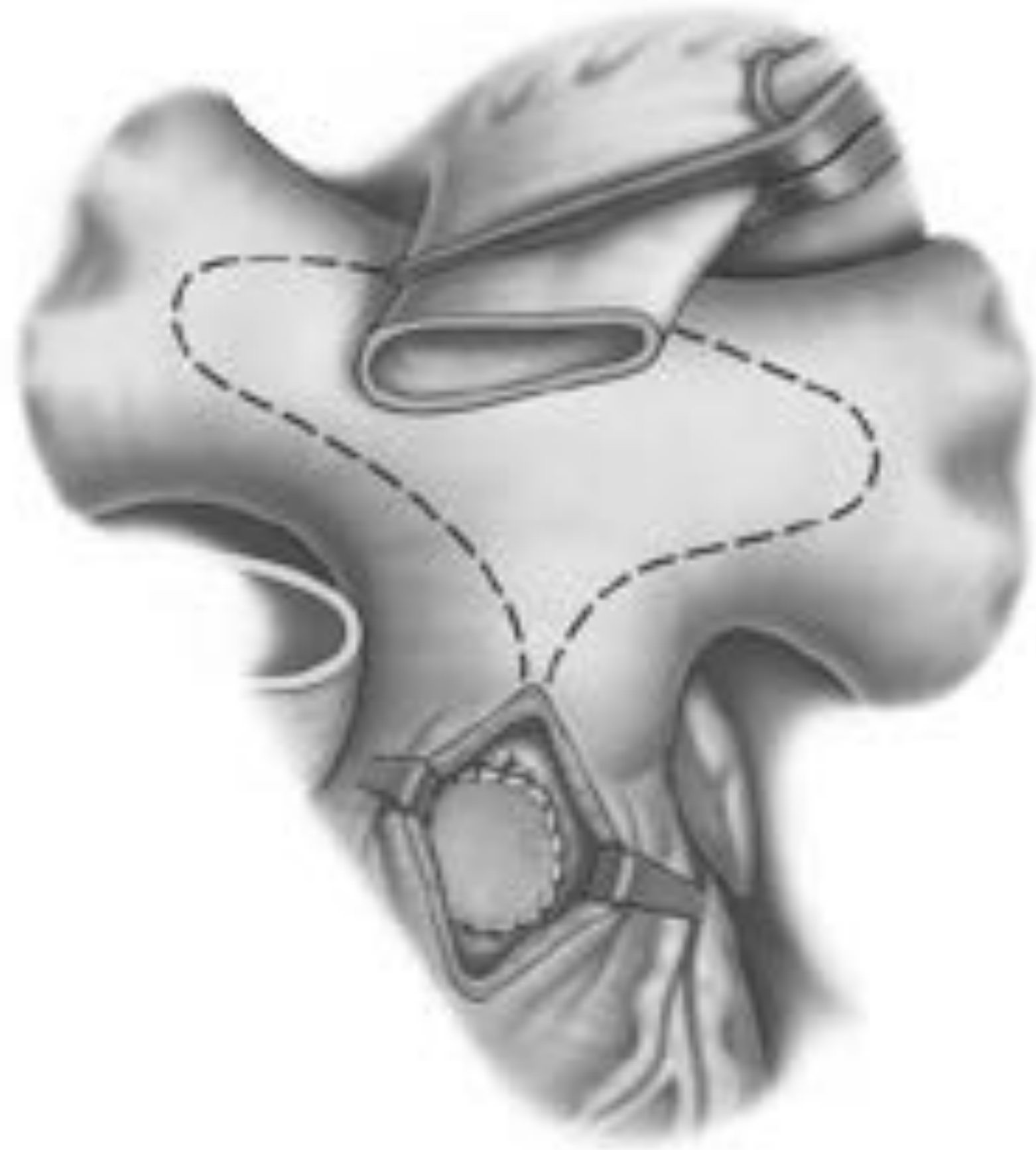








# Tetralogy of Fallot-Absent pulmonary valve: repair





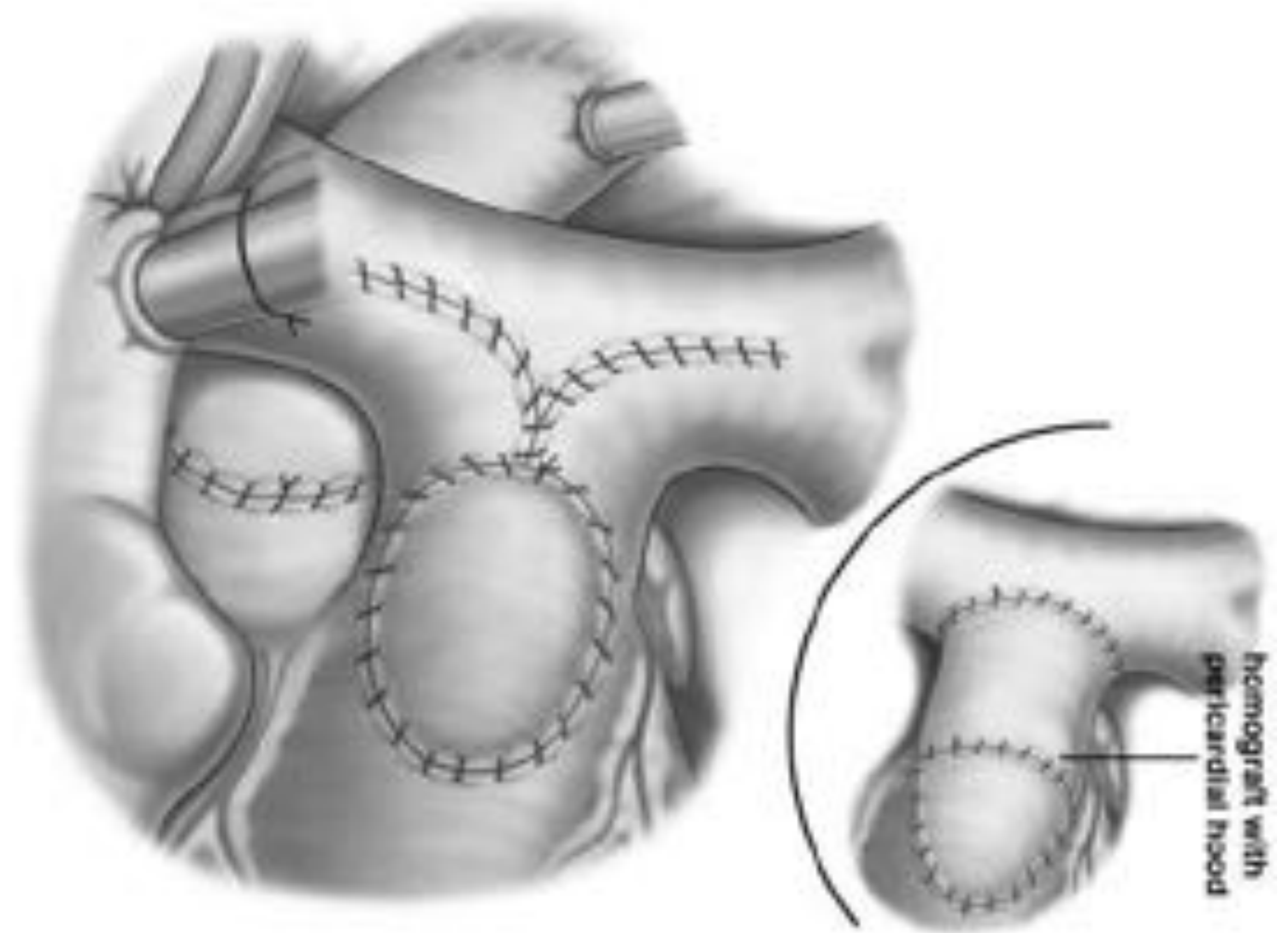
**B** preoperative



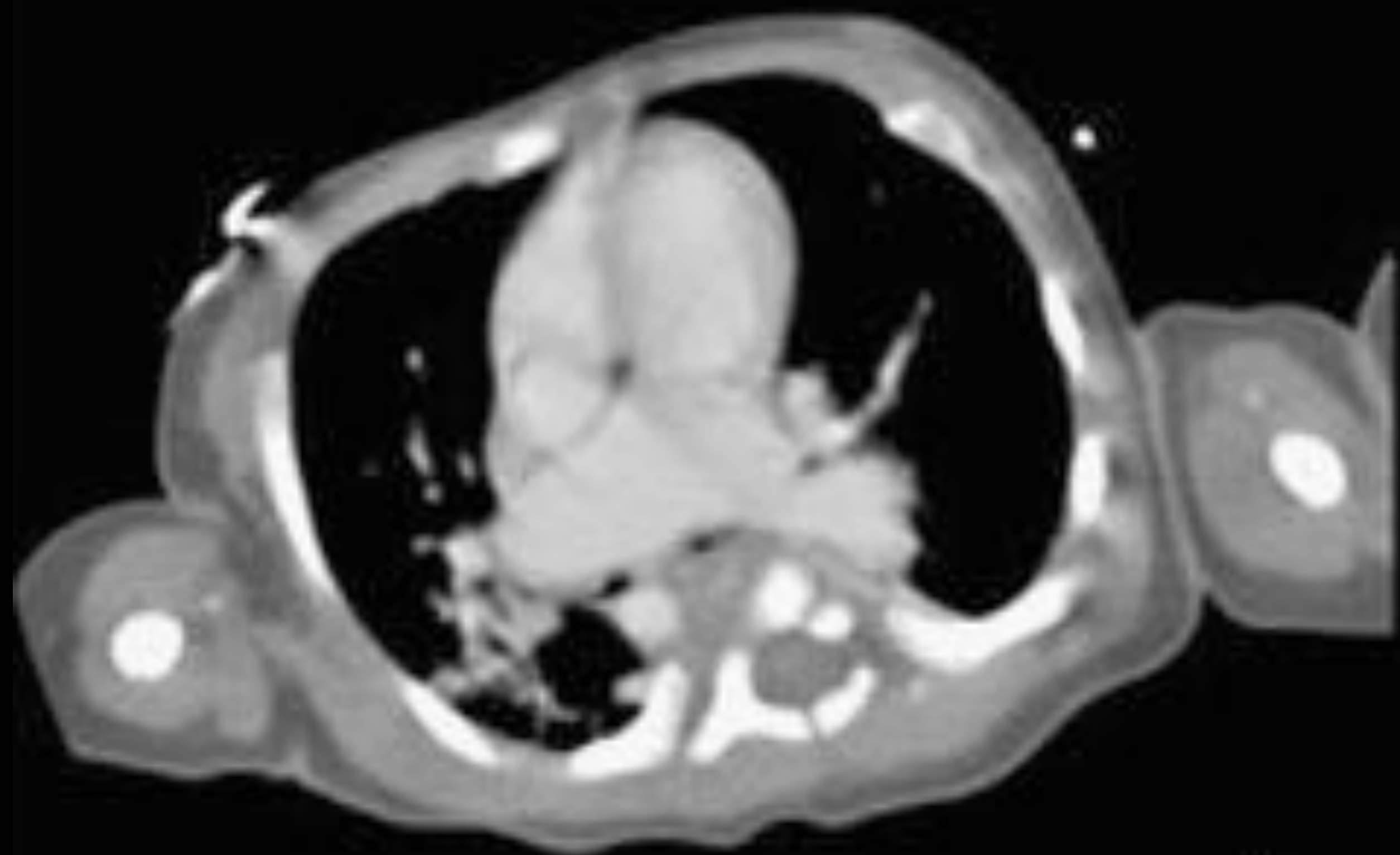
postoperative



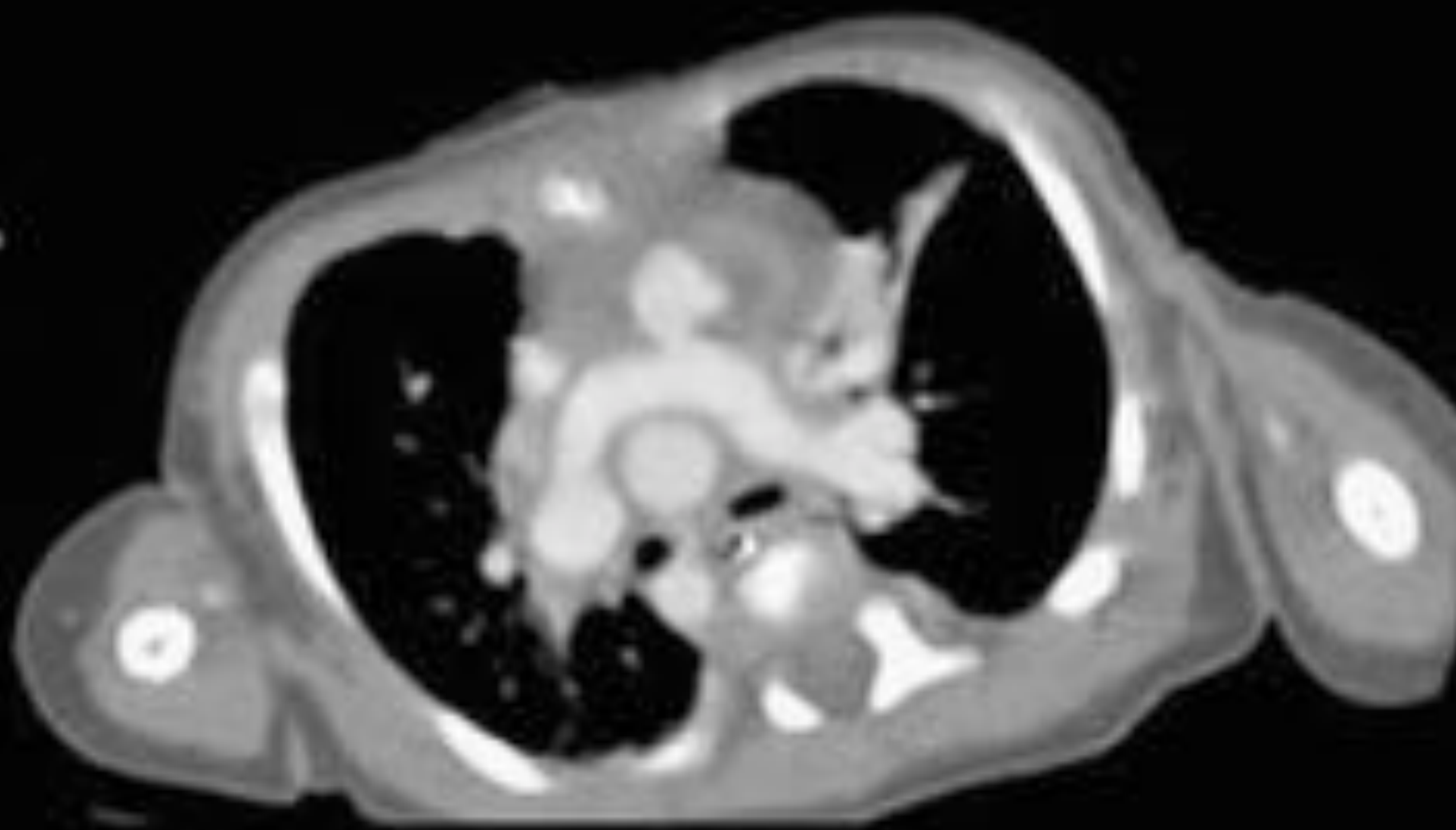
# Tetralogy of Fallot-Absent pulmonary valve: repair *With Lecompte manoeuvre*



A preoperative



postoperative





## « Tetralogy of Fallot »

Thank you