DIU RCC

Circulation de Fontan

Dr Xavier Iriart

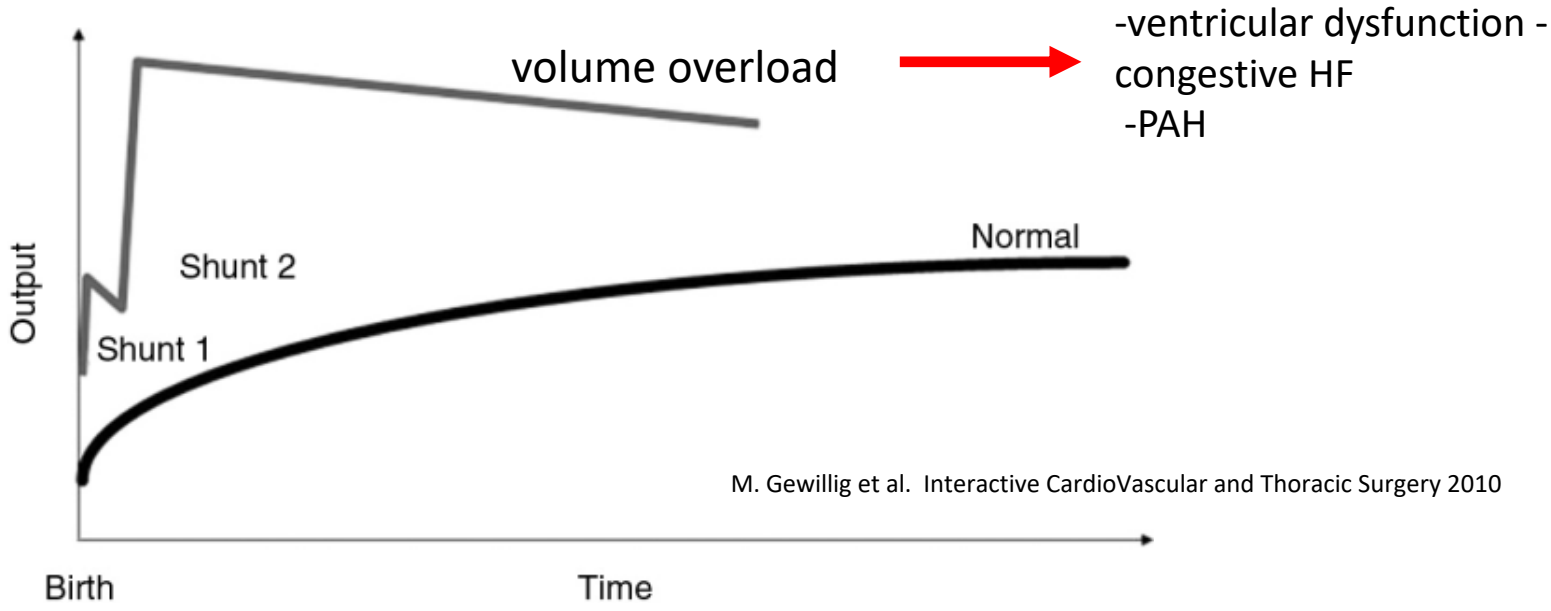
Fontan circulation Areas to be covered

- Key concepts for “single ventricle” physiology
- Stage 1: Shunt, banding and Norwood procedure
- Stage 2: PCPC
- Stage 3: TCPC

Single ventricle physiology

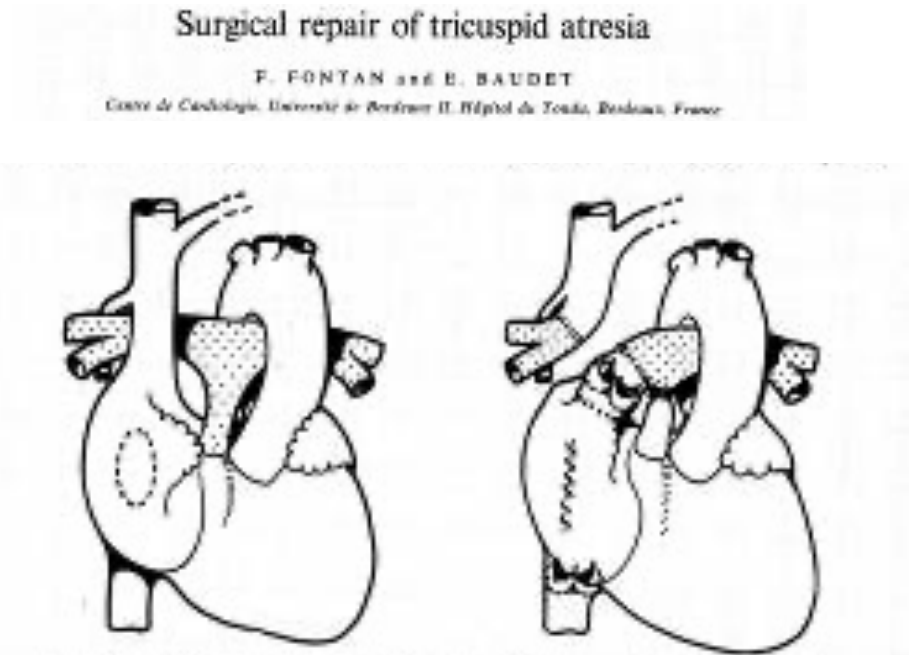
Palliation for UVH in the 1950–1960s

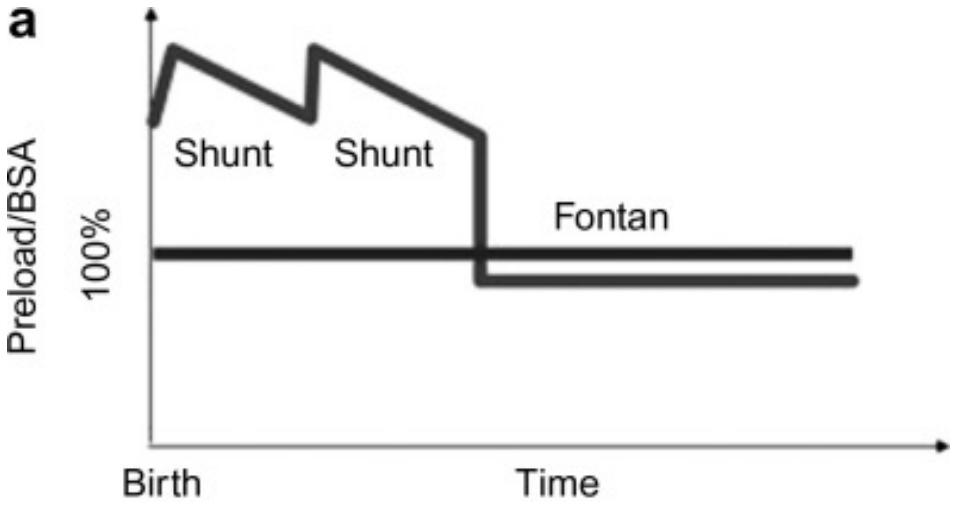
- large systemic to pulmonary artery (PA) shunts for adequate long-term relief of cyanosis
- dictum: “as pink as possible for as long as possible”
- few survivors beyond the 4th decade



M. Gewillig et al. Interactive CardioVascular and Thoracic Surgery 2010

The Fontan operation



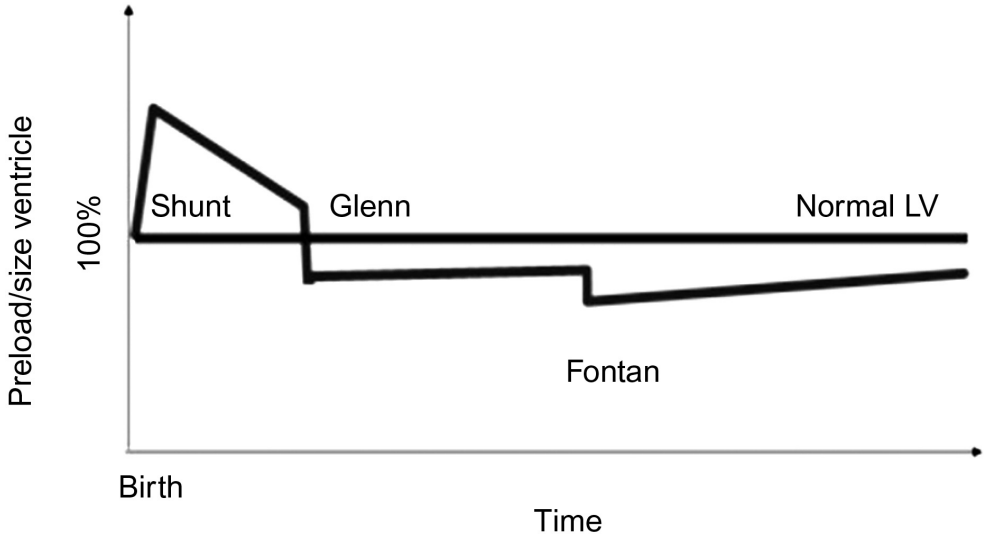


Emphasis shifted towards reducing the volume load of the ventricle

M. Gewillig et al. Interactive CardioVascular and Thoracic Surgery 2010
Fontan et al. Ann Chir Thorac Cardiovasc 1971

UVH physiology: current strategy

- From 1990's: **The staged palliation**
  - early placement of a PCPC (Glenn)
- Technical modifications
  - Smaller neonatal shunt lasting few months
- Clinical outcomes improvements
- The dictum: "as blue as possible" in order to keep the ventricle maximally unloaded.

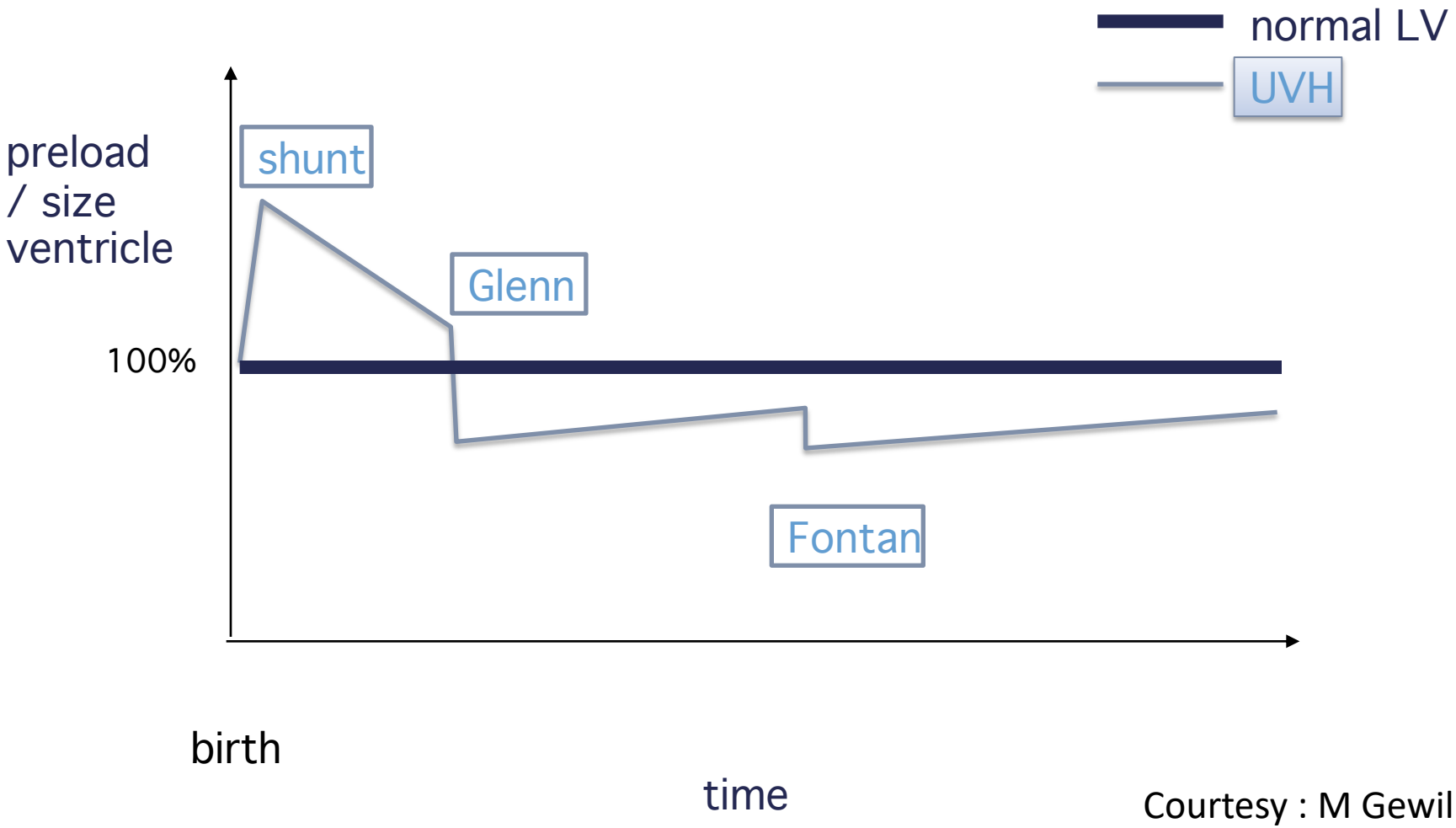


Emphasis further shifted to limitation of volume load as early as possible.

M. Gewillig et al. Interactive CardioVascular and Thoracic Surgery 2010

Preload : normal LV vs UVH

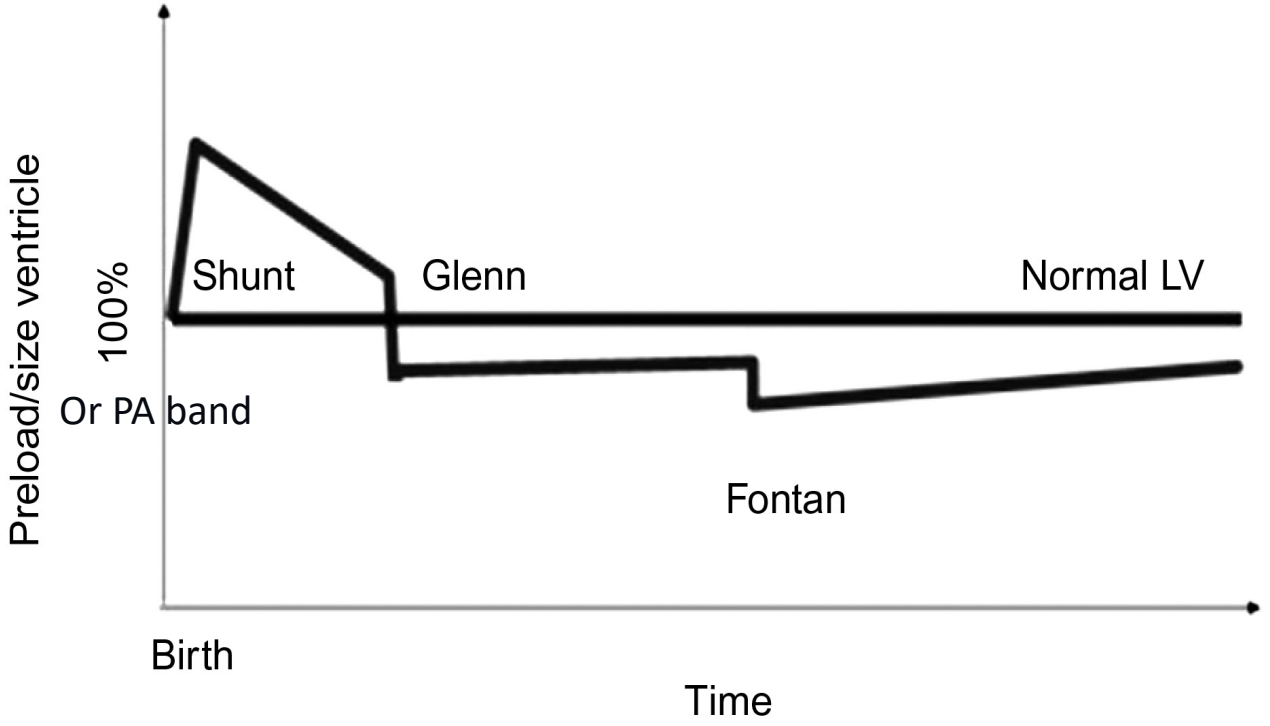
Staged Fontan palliation



Courtesy : M Gewillig

Goals of stage 1 palliation

- Balance aortic / pulmonary blood flow
- Optimize PA growth +++
- Protect from pulmonary vascular disease
- Surgical timing: limit the period of ventricular overload

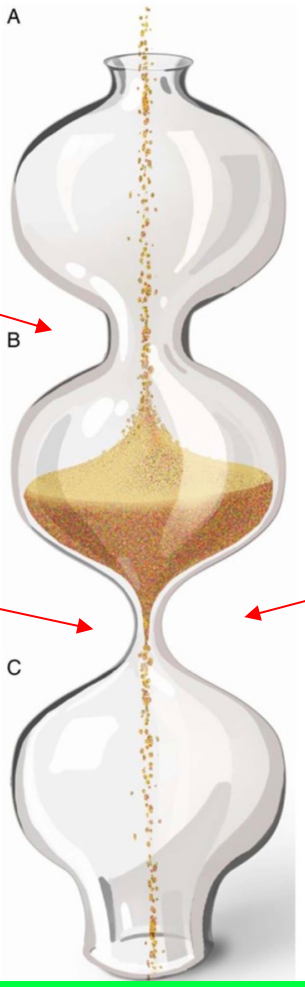


Multiple bootleneck concept

Absence of the RV to PA coupling: influence on the systemic venous return and the pulmonary circulation

No pump to propel blood into the PAs

Pulmonary impedance hampers venous return through the pulmonary vascular bed



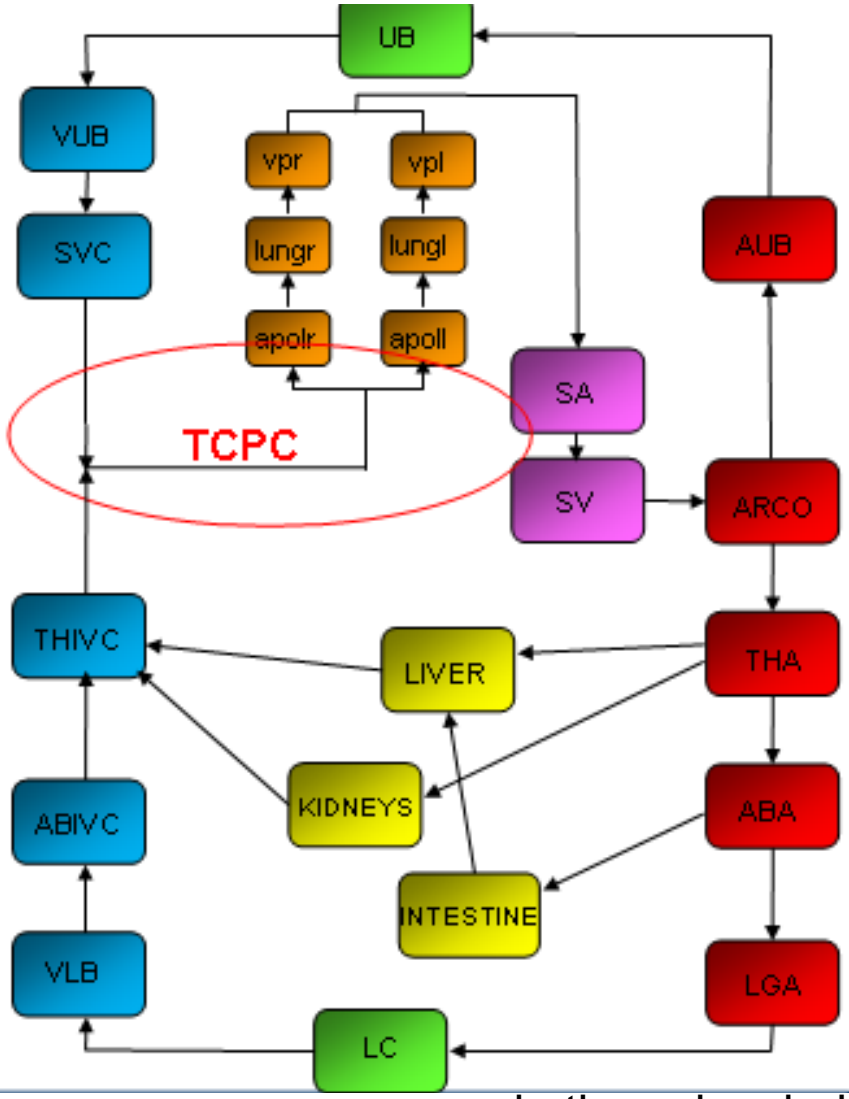
upstream venous congestion

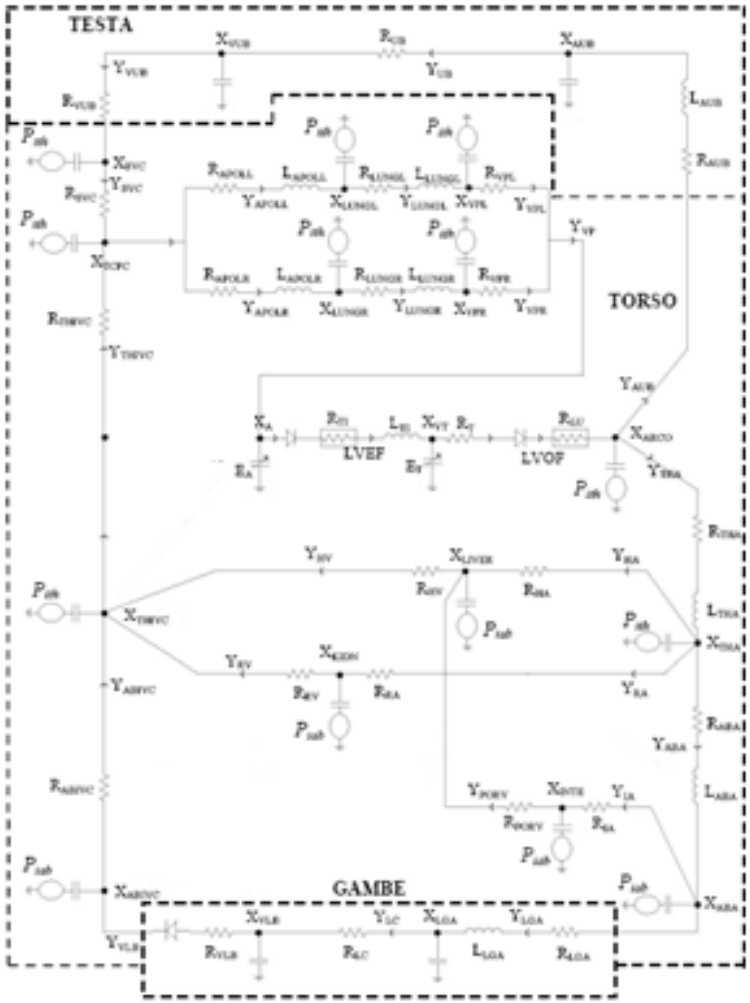
Risk of poor function of the Glenn and future Fontan if PAs are too small and impedance too high ++++++++

downstream decreased output

good ventricular function (systolic AND diastolic)

Gewilig and Brown. Heart 2015

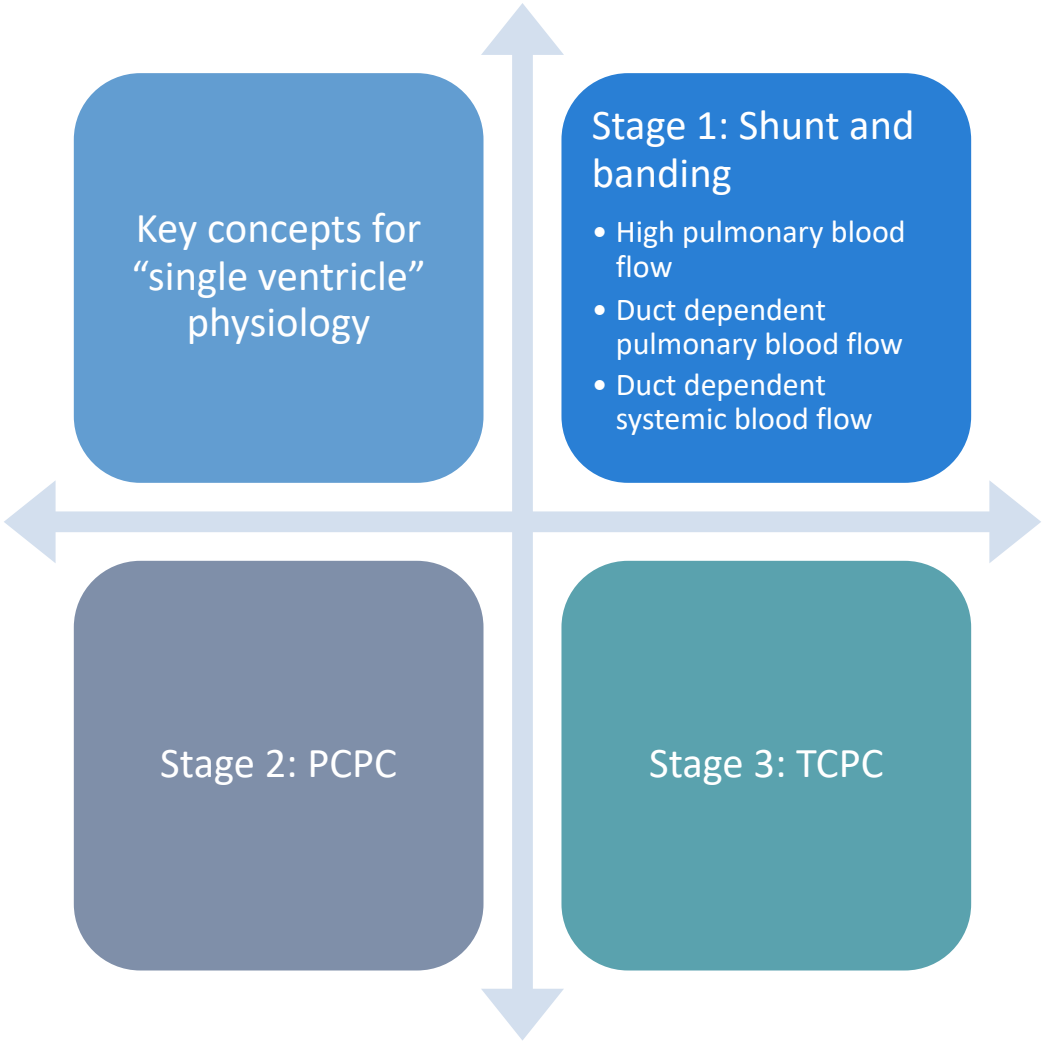




venous pressure in the splanchnic system is 2-3x higher than that in the systemic inferior caval system.

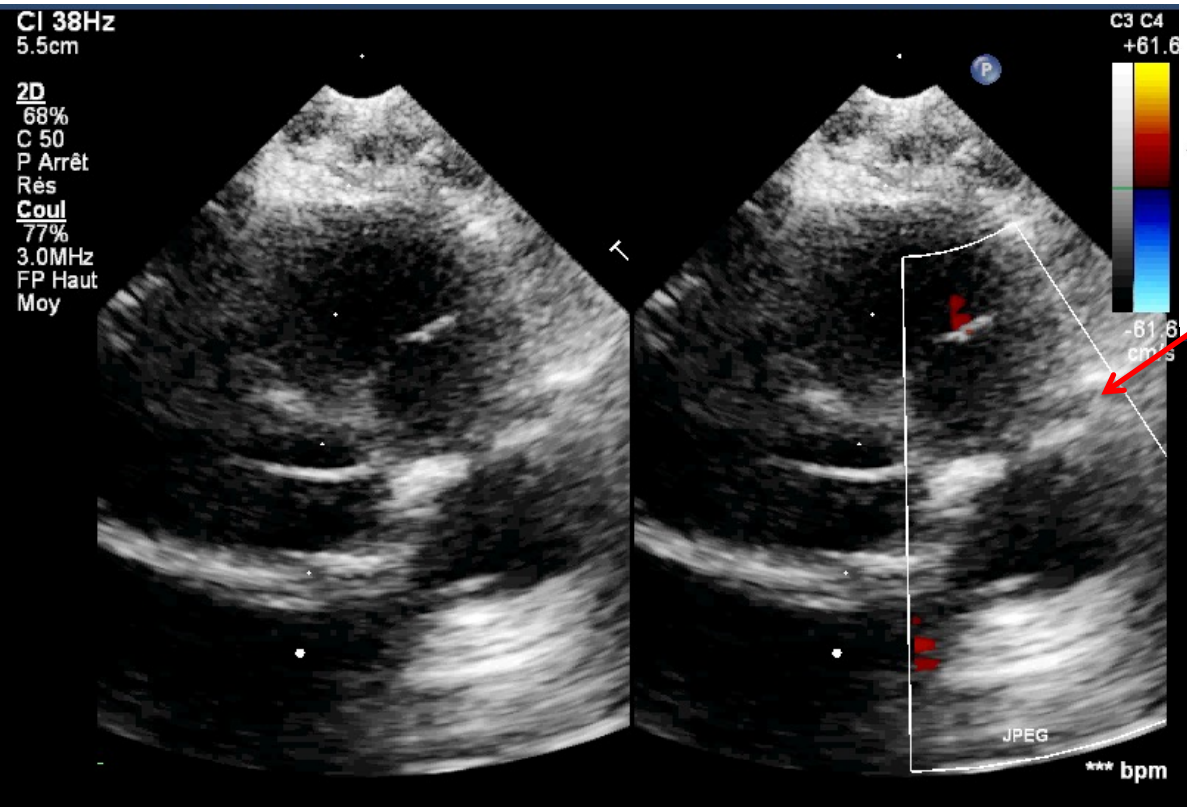
Courtesy of Daria Cosentino, Great Ormond street hospital and UCL London

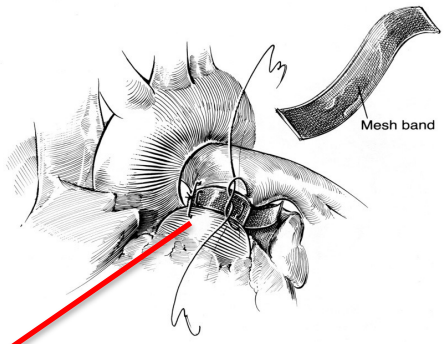
Areas to be covered

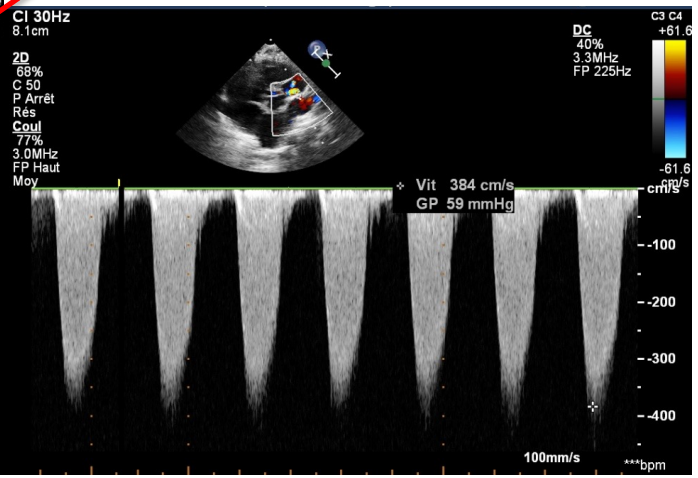


High pulmonary blood flow
PA banding

- SV-PA gradient: cw doppler (SAX/A4C). Longitudinal FU
- Pulmonary valve regurgitation: 2D/ color doppler
- Migration of PA banding: pulmonary branch distortion (RPA)

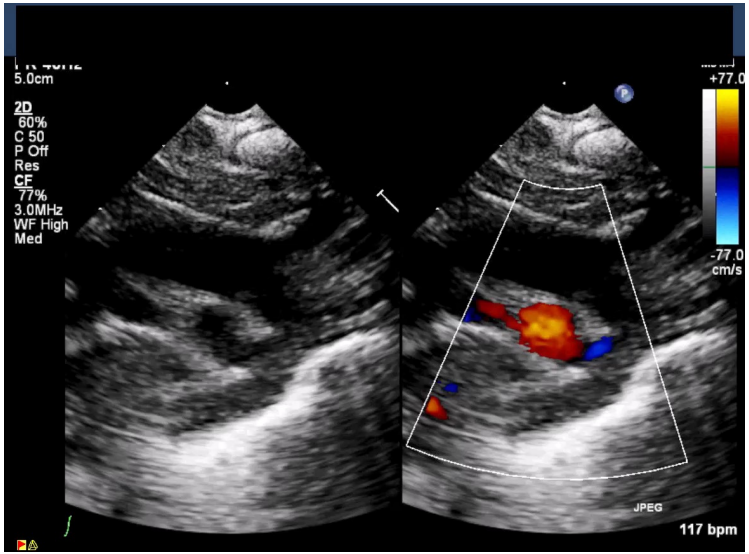


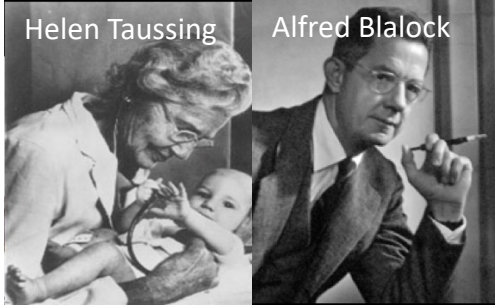




Duct dependent Pulmonary Blood Flow

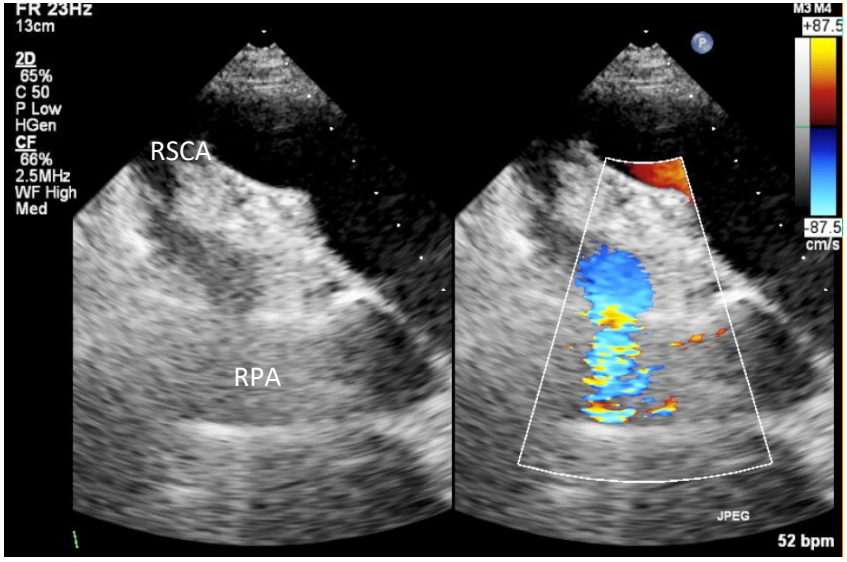
- Anatomy: Atresia/Hypoplasia of Right Heart structures (Tric/ pulm Atresia)
- **Immediate** action to establish **adequate** Pulmonary Blood Flow
  - Prostaglandin to maintain ductal patency
    - Flow: Aorta to Pulmonary artery
- Subsequent need for **Reliable** Pulmonary Blood Flow
  - Modified BT shunt
  - PDA stent

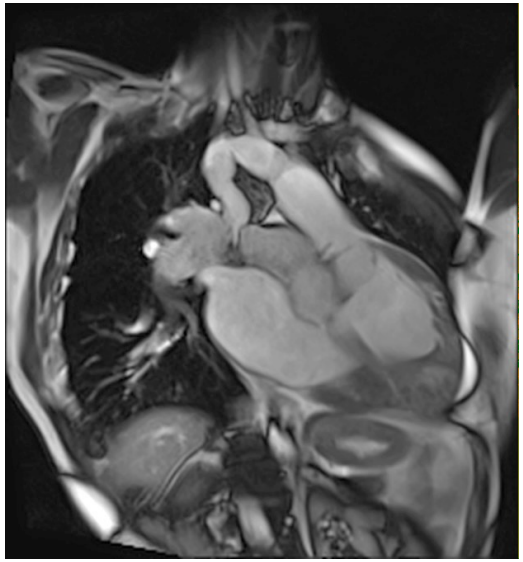


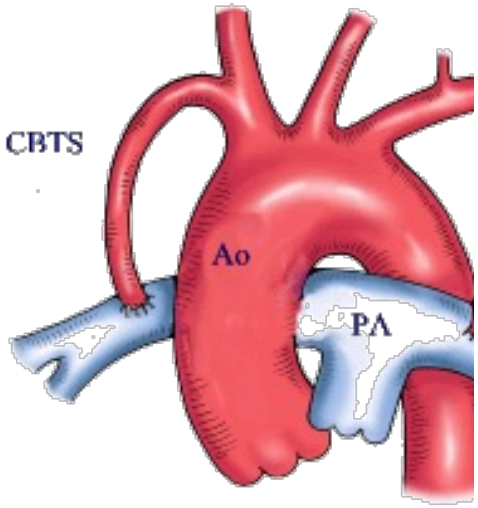


Duct dependent Pulmonary Blood Flow

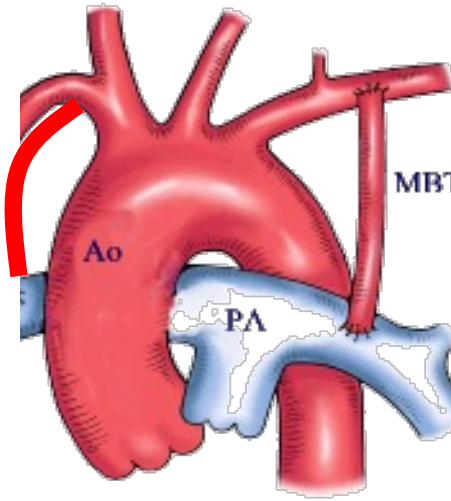
- First SPS: direct connection between SCA and ipsilateral PA
- Frequent complications
  - Unpredictibility of shunt flow
  - PA branch distortion



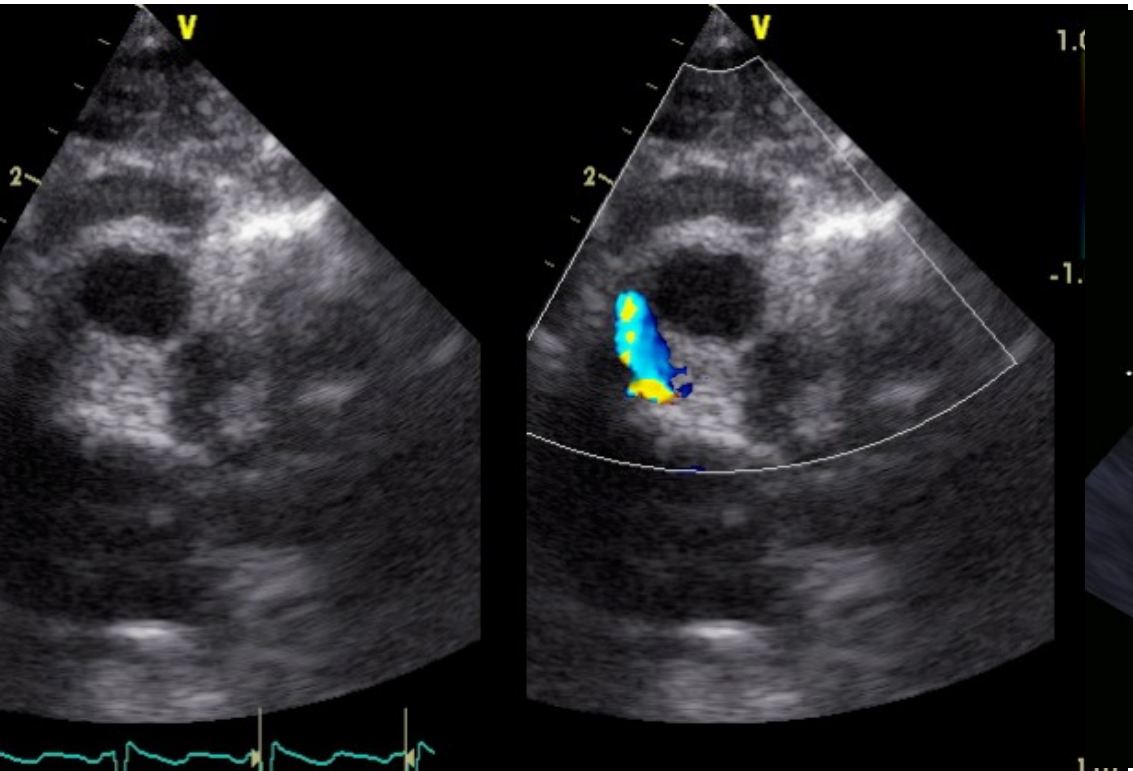


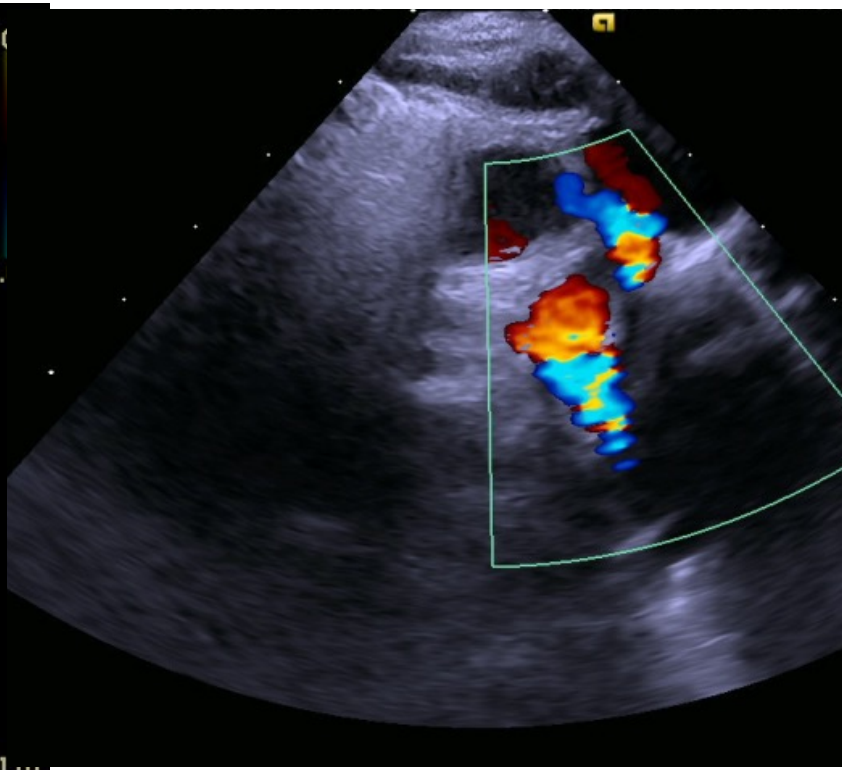


Systemic to pulmonary shunt

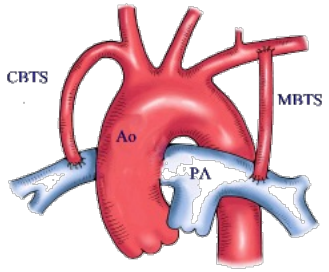


- modified BT shunt: prosthetic PTFE graft (Ø3-4mm)
- Innominate artery or SCA connected to ipsilateral PA branch
- Echo: suprasternal frontal view/ color doppler/ CW doppler



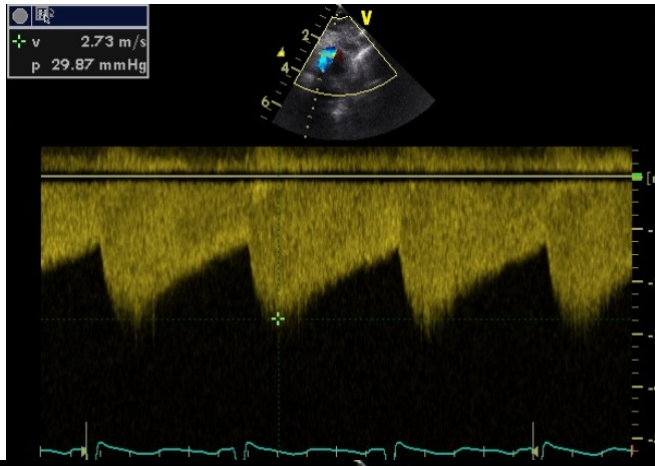


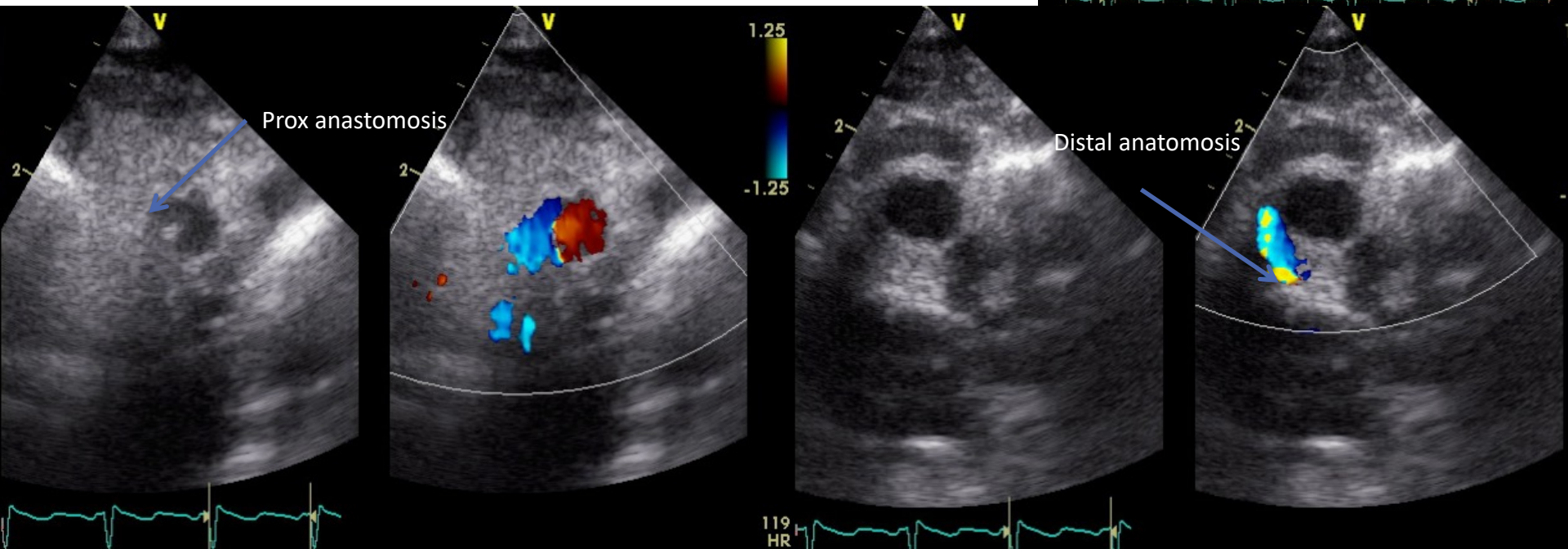
Systemic to pulmonary shunt



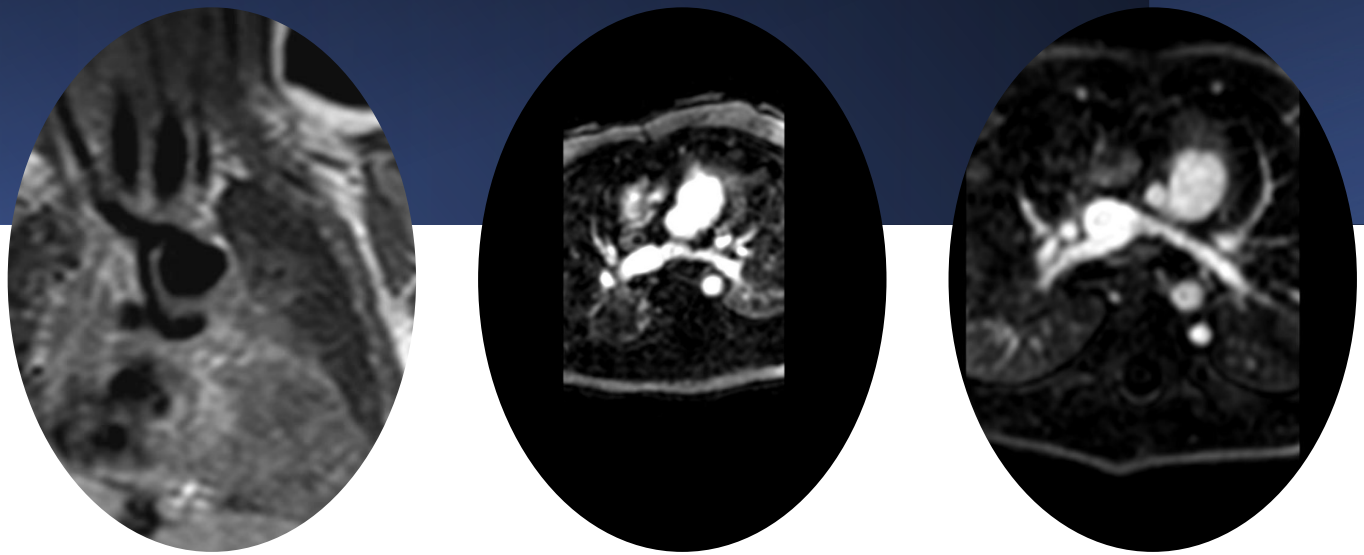
- Imaging: CW doppler: characteristic sawtooth doppler pattern

- Potential anomalies: distortion of inn Artery or PA branch, narrowing of prox or distal anastomosis (challenging)





Visualisation of shunt and PAs



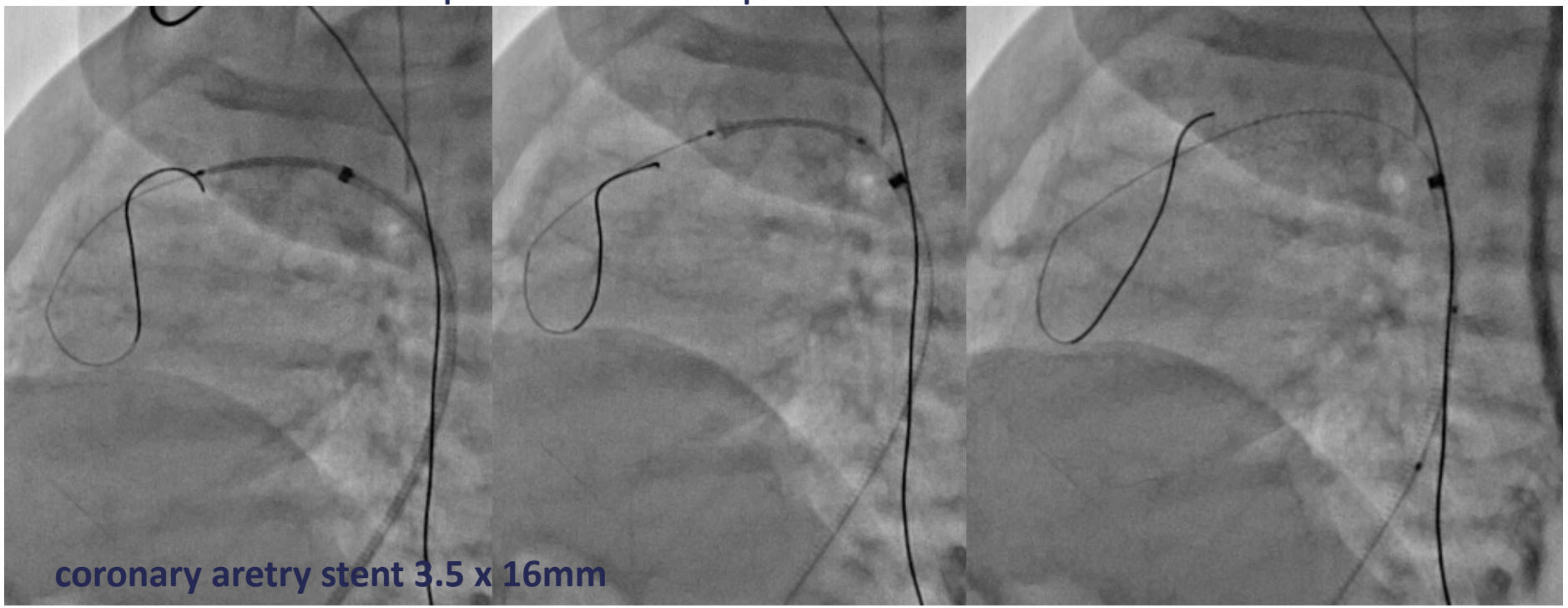
- If there is doubt or for planning next stage of surgery
- CT / MRI / Catheter
- To delineate shunt and branch PA anatomy

DUCT STENTING

Pulmonary atresia with IVS. Severe hypoplastic RV. Duct dependent pulmonary flow

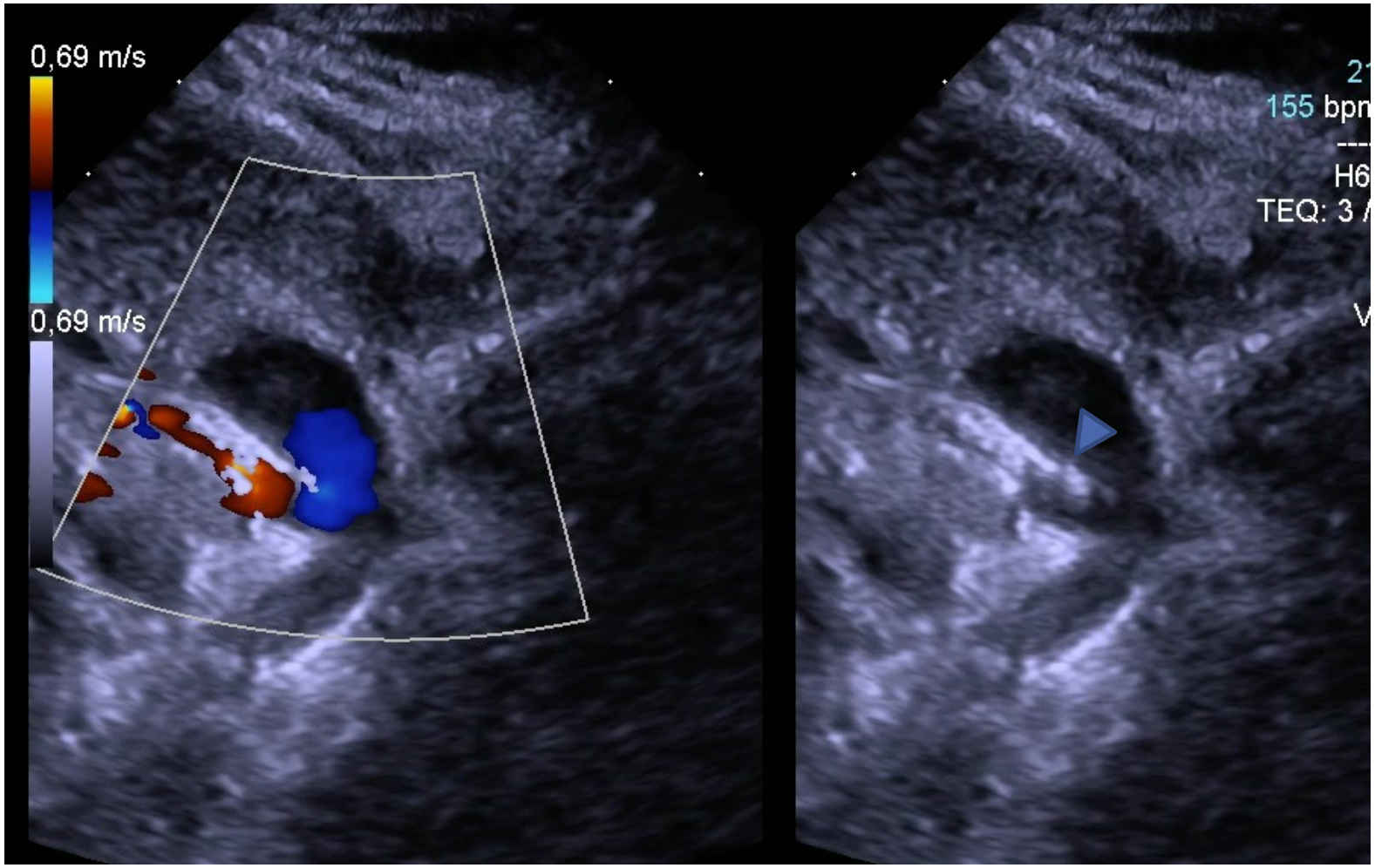
Weight 2kg. O2 Saturation 88% under PGE1

Stop PGE1 6h before the procedure



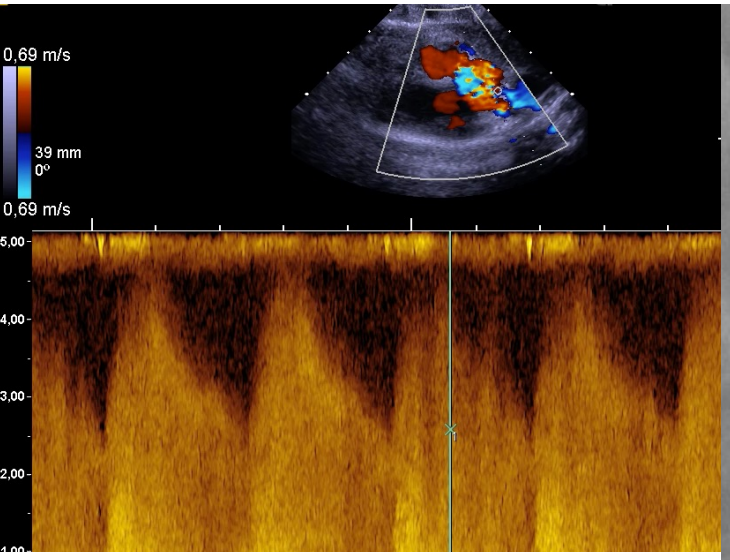
Discharge Day 5 O2 Sat85-90%

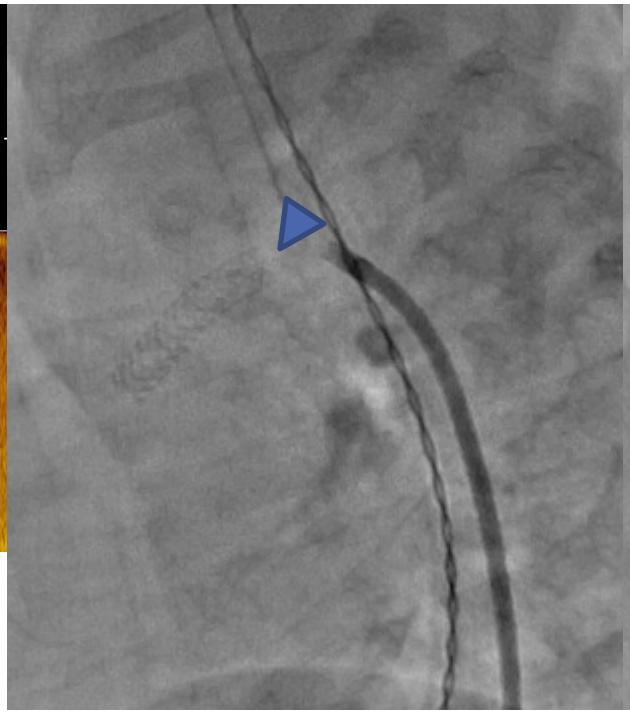
DUCT STENTING

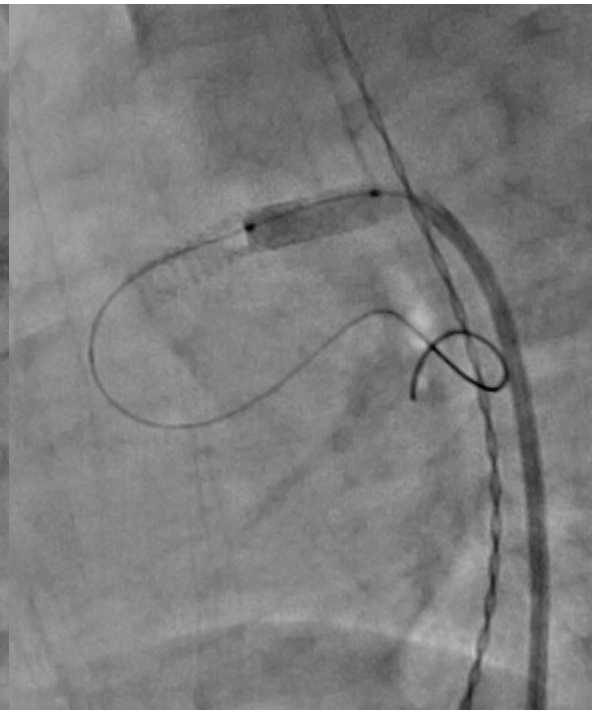


DUCT STENTING

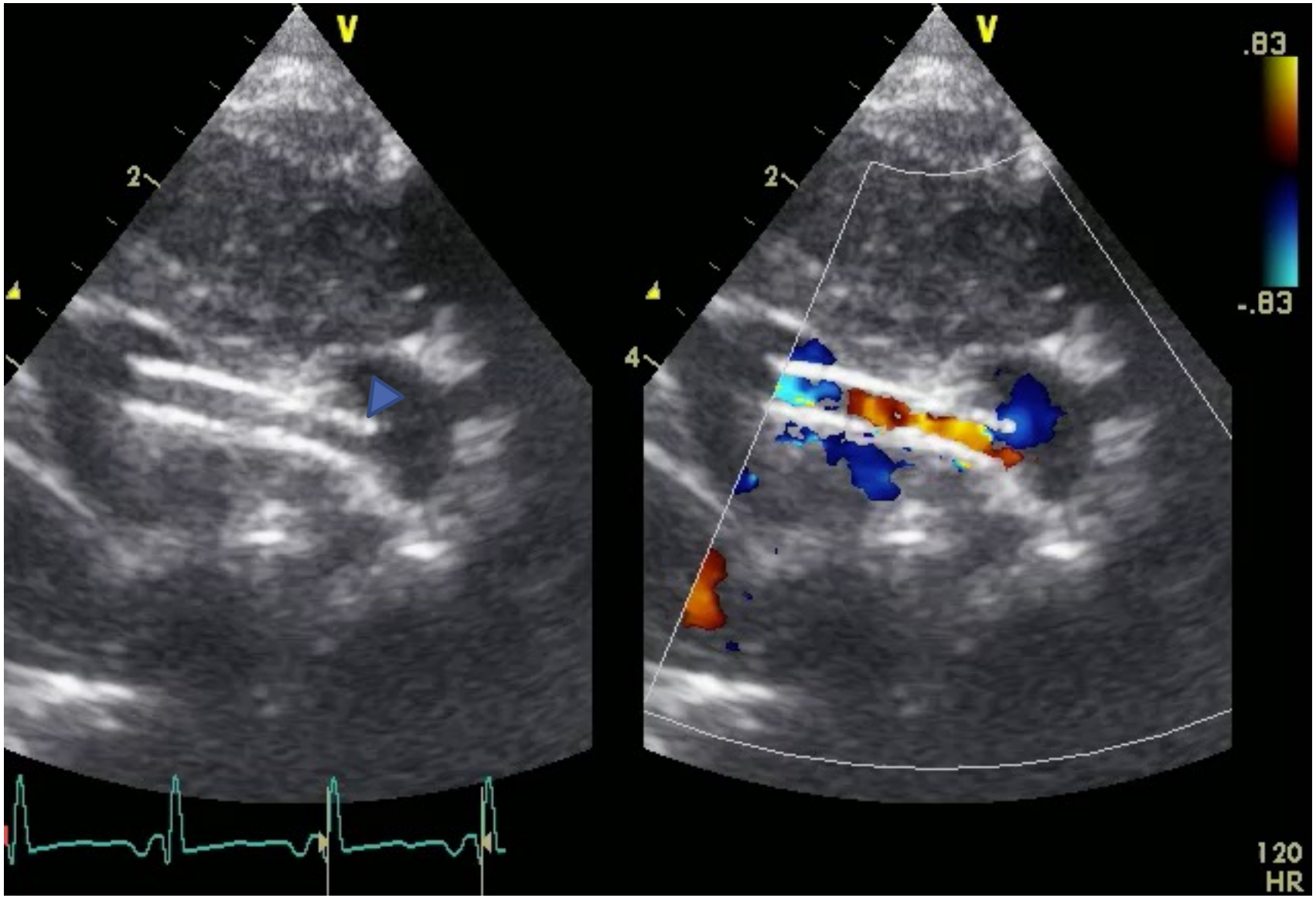
- Acute desaturation at 3 months 60 vs 85%
- Increased peak velocity from 3.5 to 5m/s



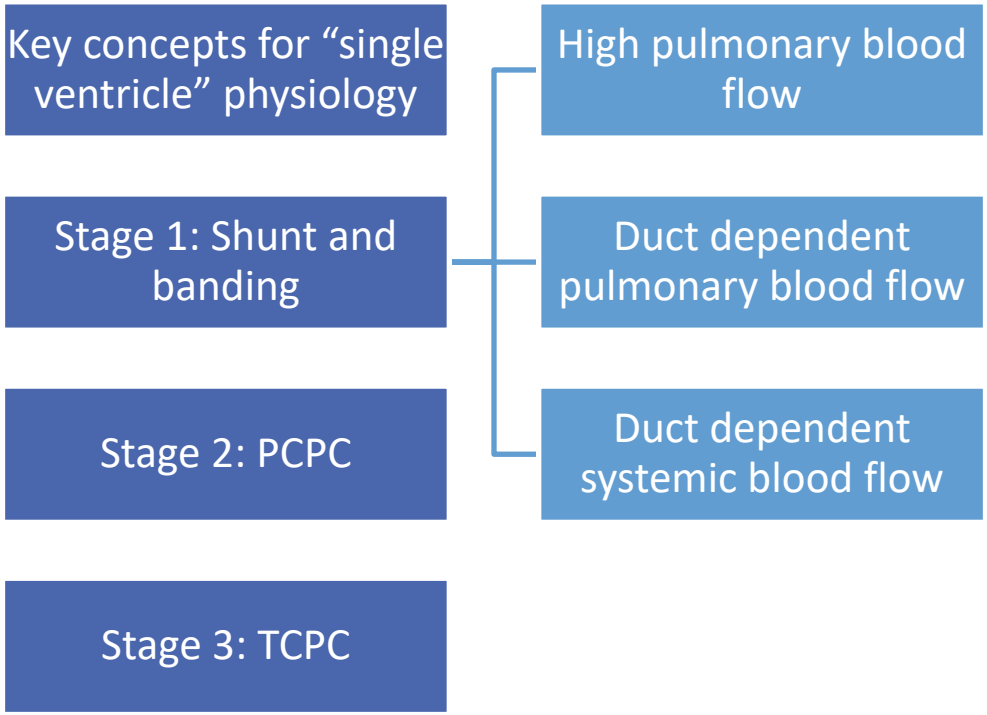




Additional stent on the aortic side

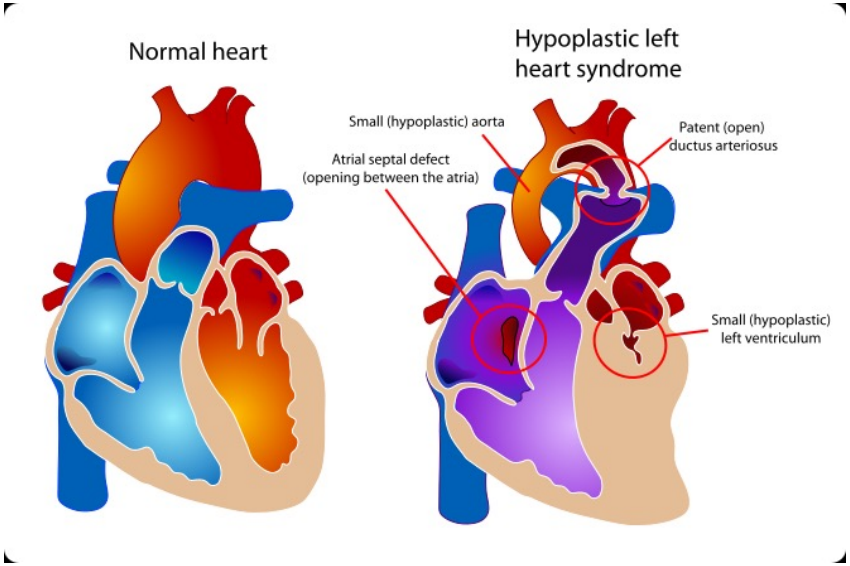


Areas to be covered



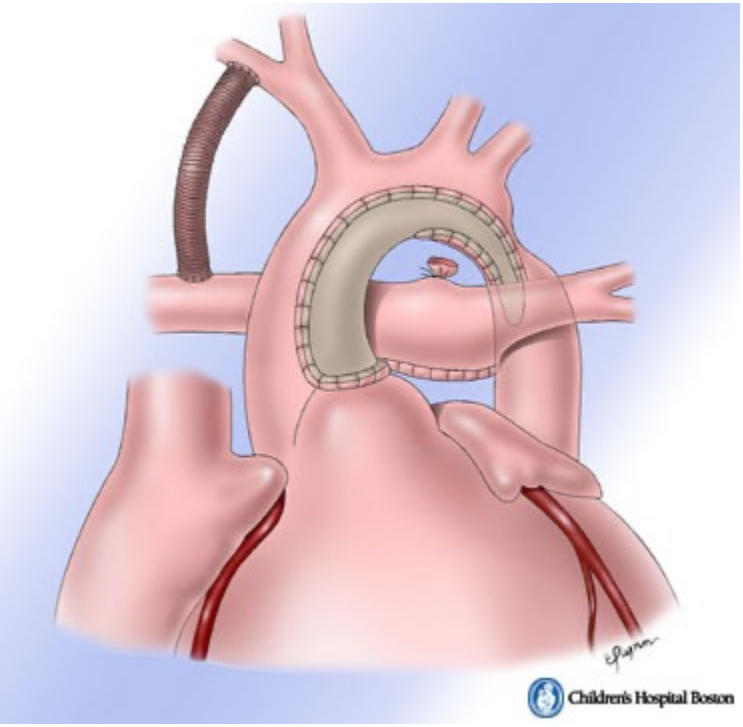
Duct dependent Systemic Blood Flow

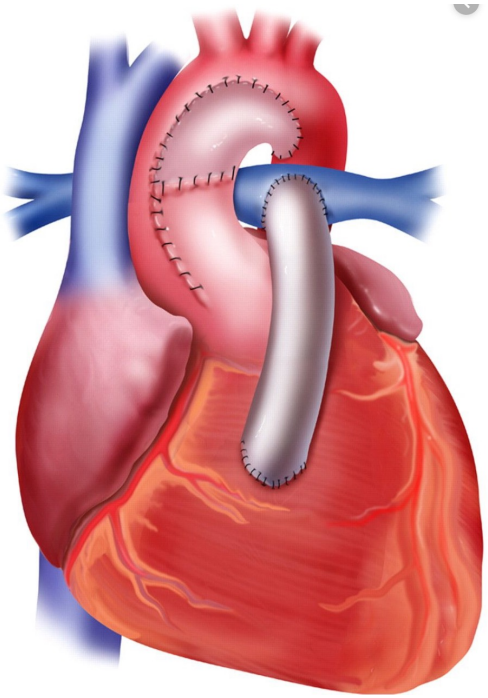
- Anatomy: large spectrum of HLHS
- **Immediate** action to establish **adequate** Systemic Blood Flow
  - Prostaglandin to maintain ductal patency (R to L flow)

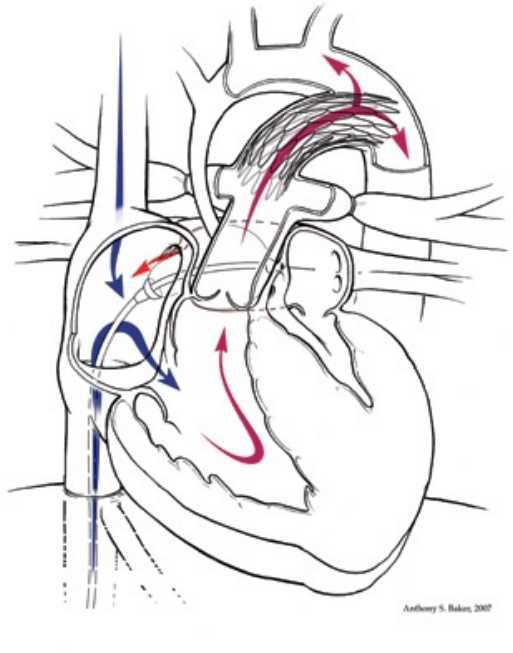


- Subsequent need for **Reliable** systemic circulation
  - Norwood/Hybrid for HLHS
- Balloon or Surgical Aortic Valvotomy in Critical AS

Palliative approach in HLHS

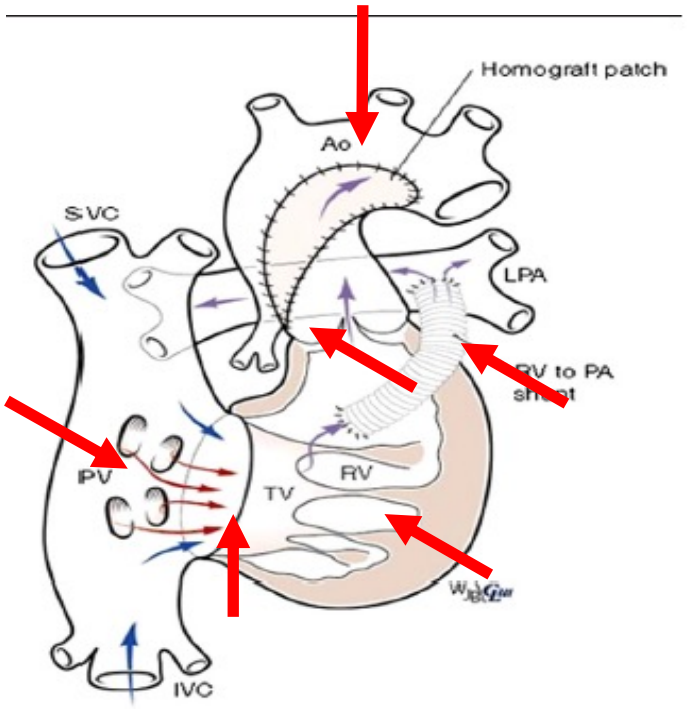


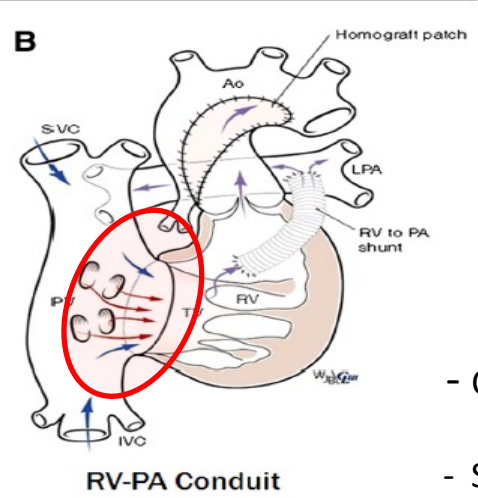




Post Norwood stage I echocardiography

- <u>Sequential segmental analysis</u>
  - Atrial septectomy
  - Tricuspid valve function
  - RV function
  - DKS anastomosis
  - Neoaortic arch
  - Systemic to pulmonary artery shunt

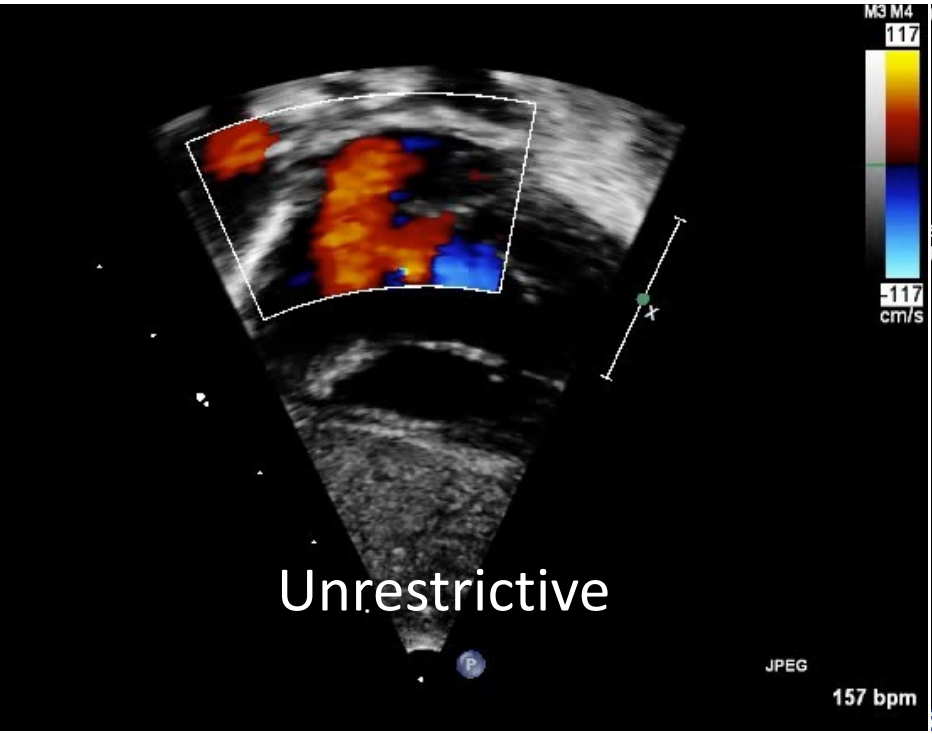




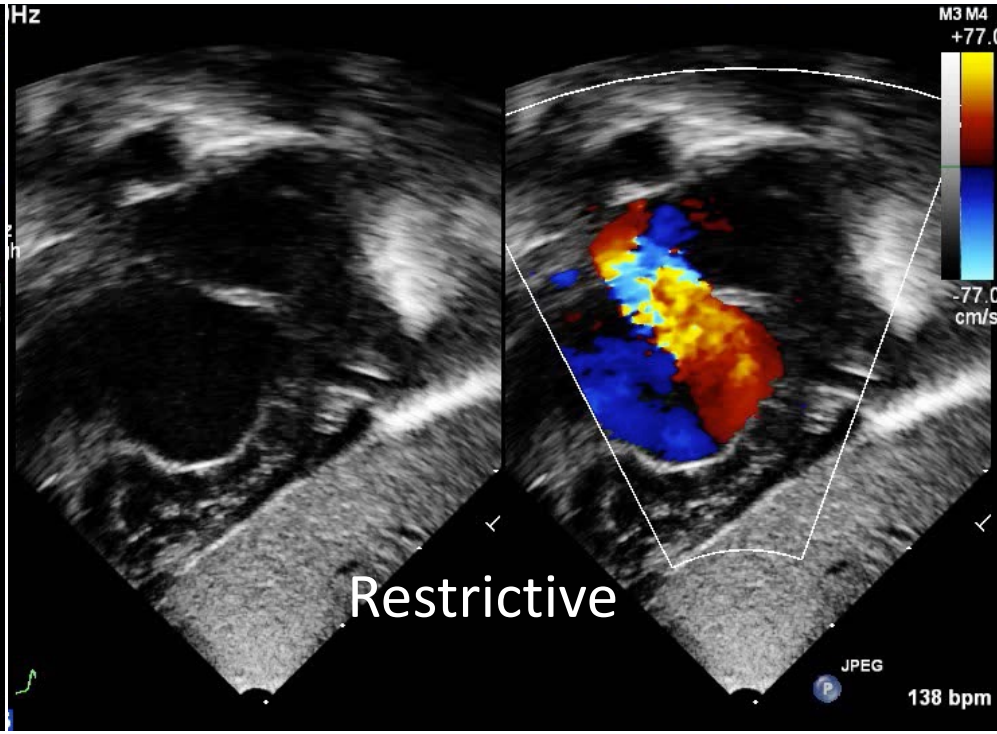
RV-PA Conduit

Assessment of atrial septum

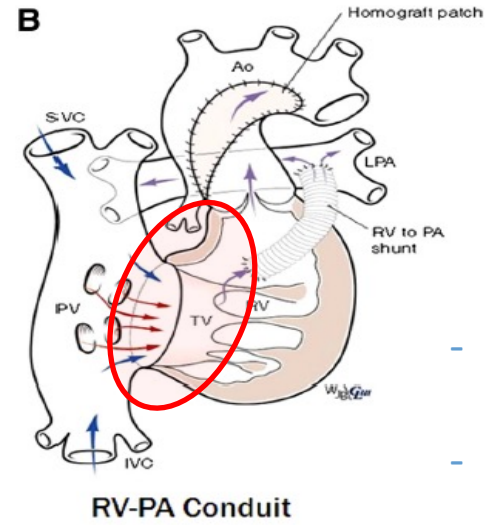
- Crucial for decision-making in the immediate post-natal period
- Subcostal view: color doppler



Unrestrictive



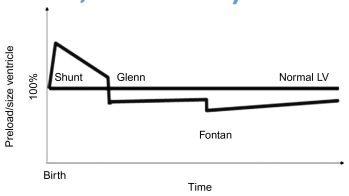
Restrictive

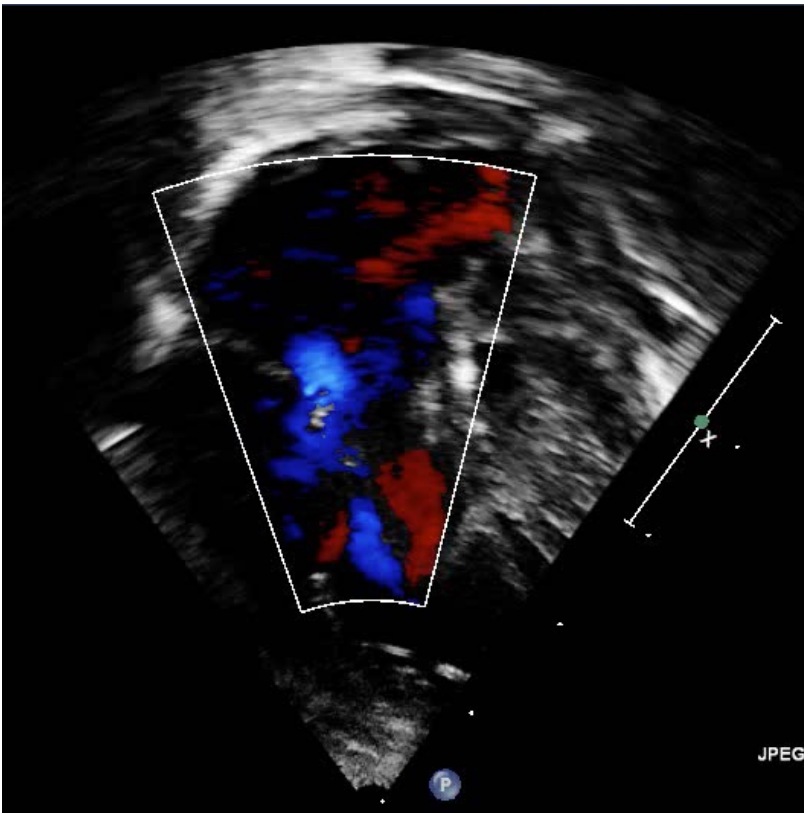


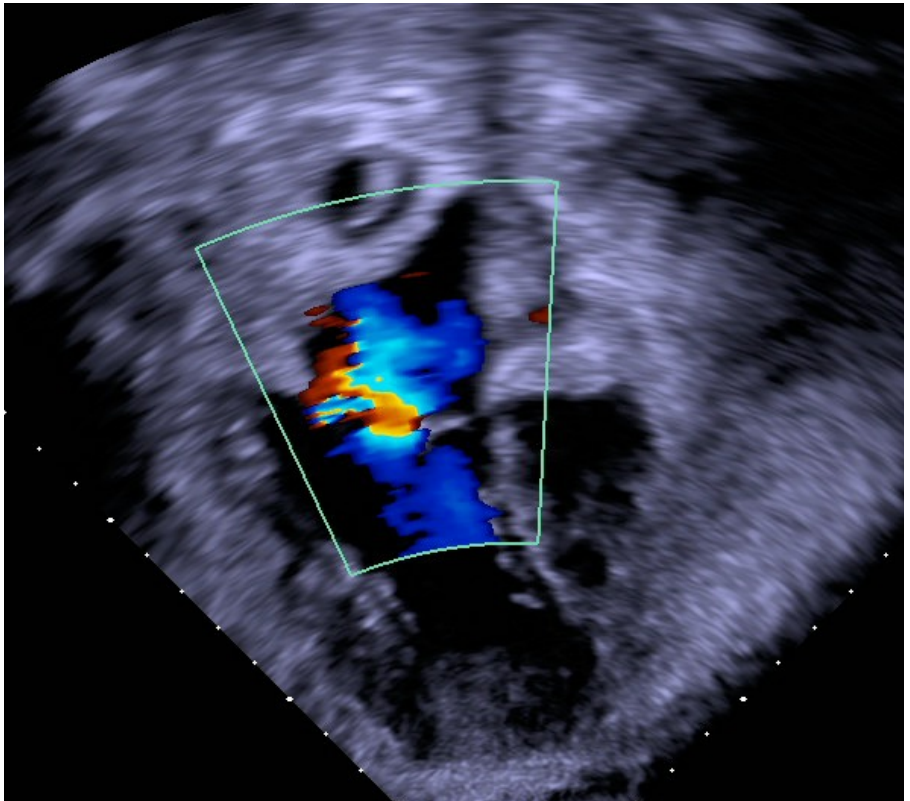
RV-PA Conduit

Assessment of TR

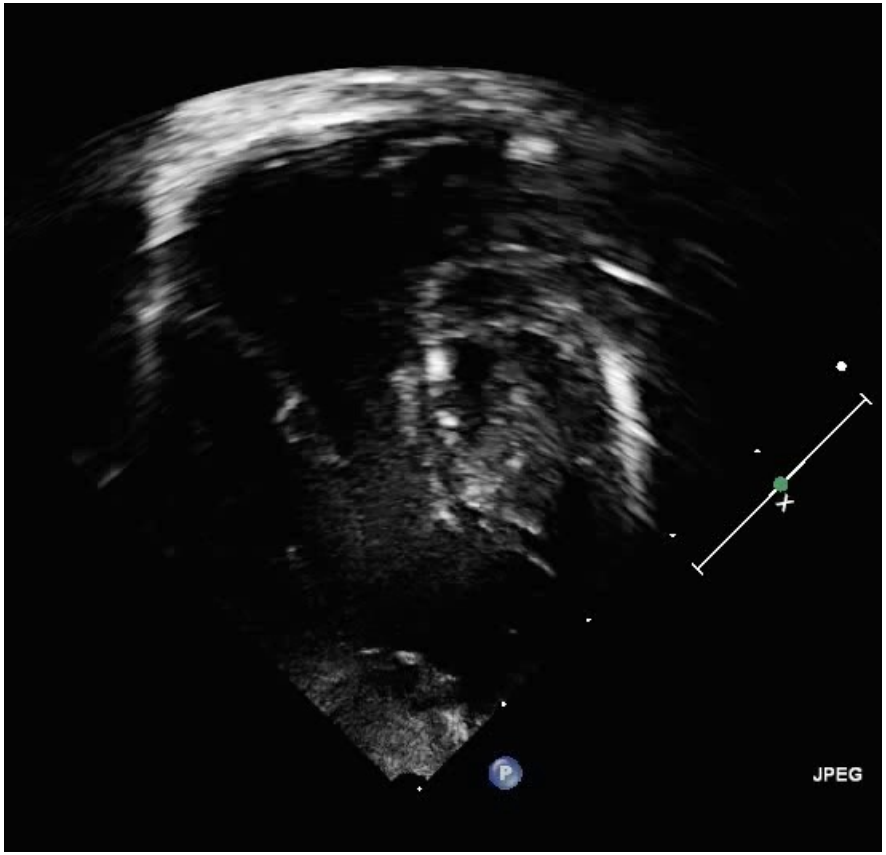
- Semi quantitative assessment (mild, moderate, severe)
- Consider change in loading conditions

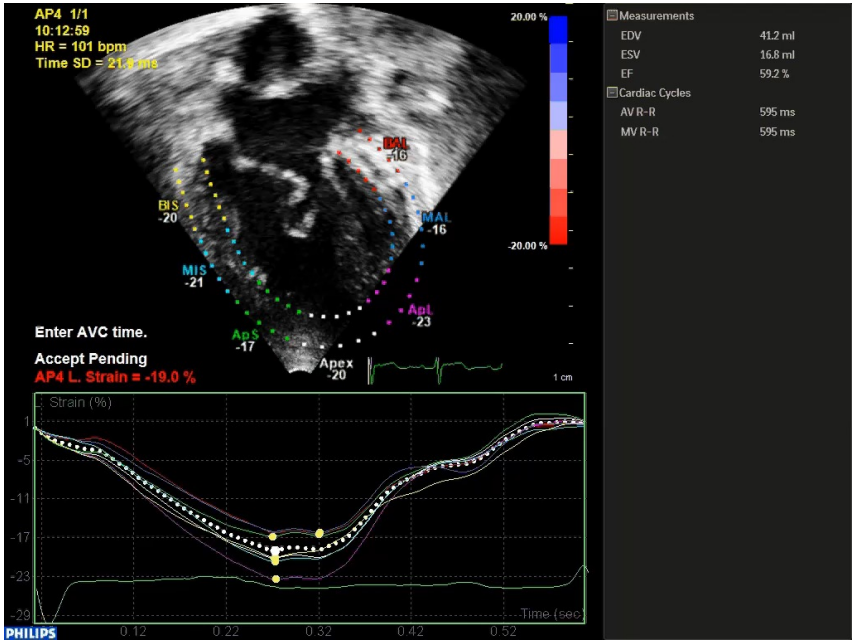






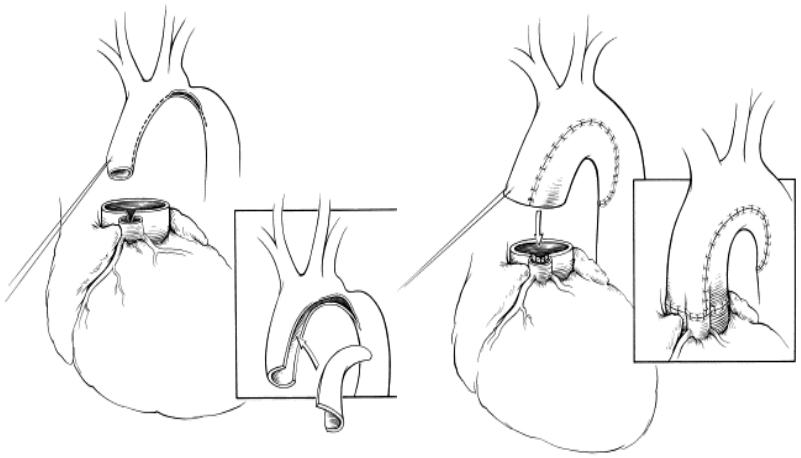
Eyeballing of RV contraction

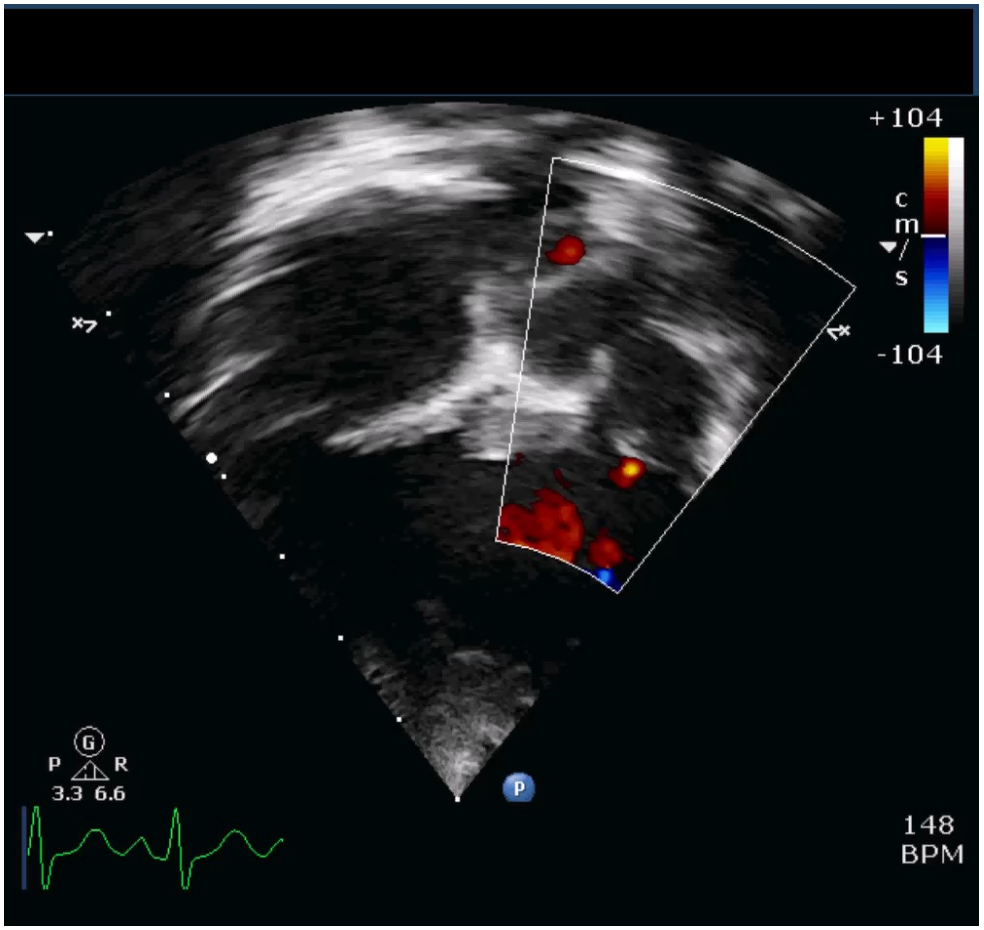




- Strain analysis

Asessement of DKS anastomosis

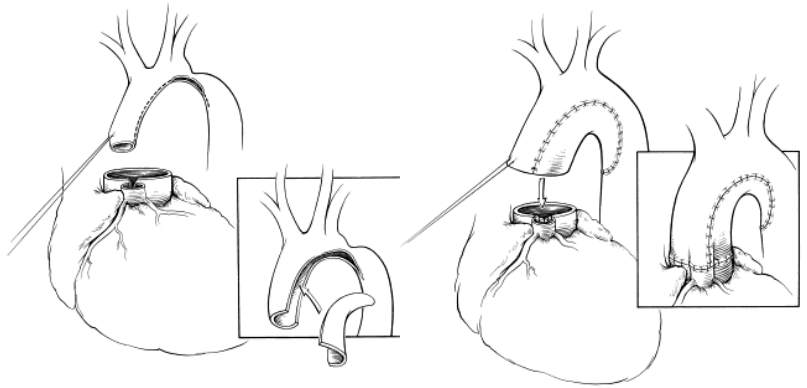


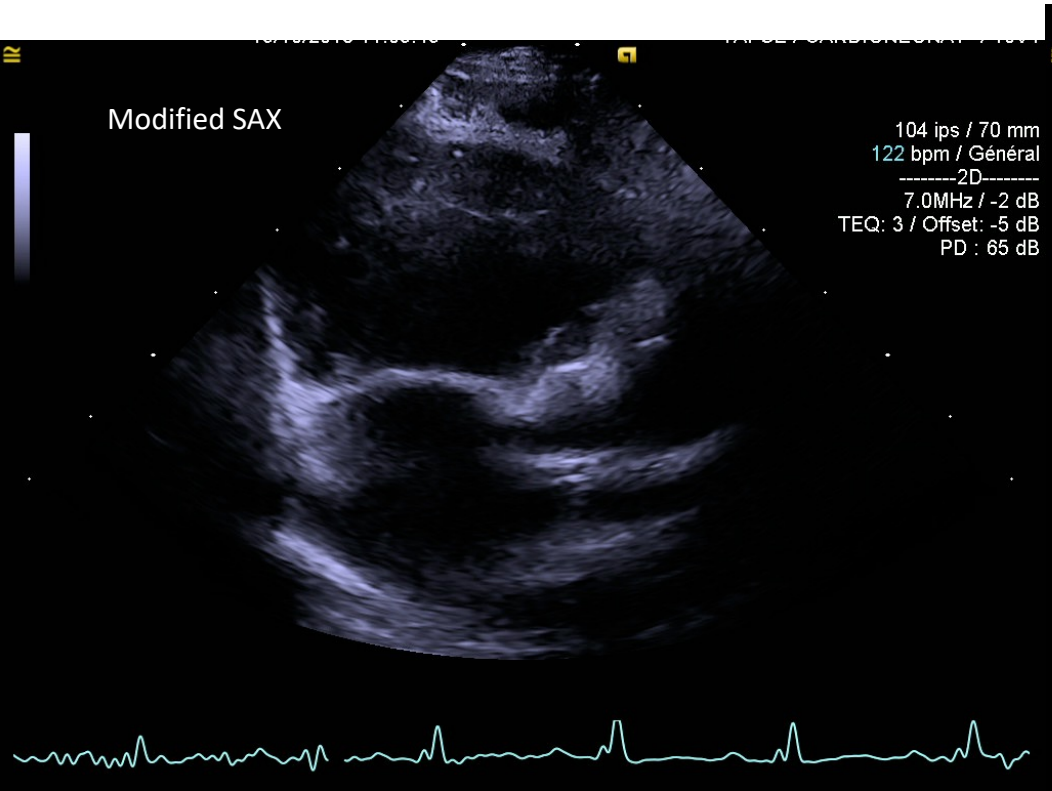


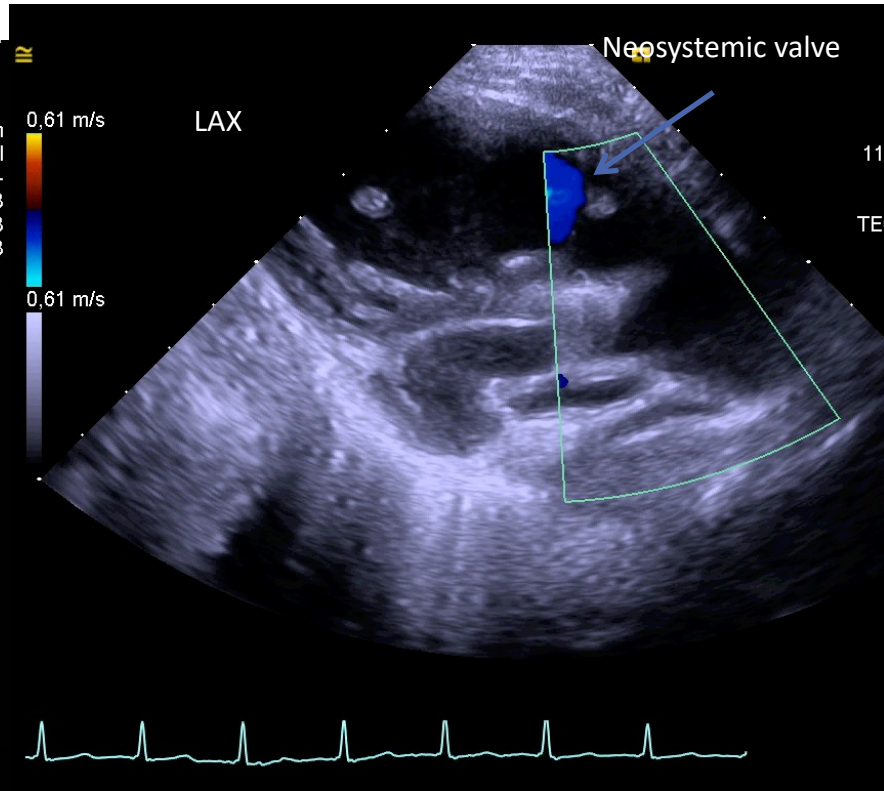
- Coronary blood flow is retrograde

Asessement of DKS anastomosis

- DKS anastomosis
- Neoaortic valve regurgitation

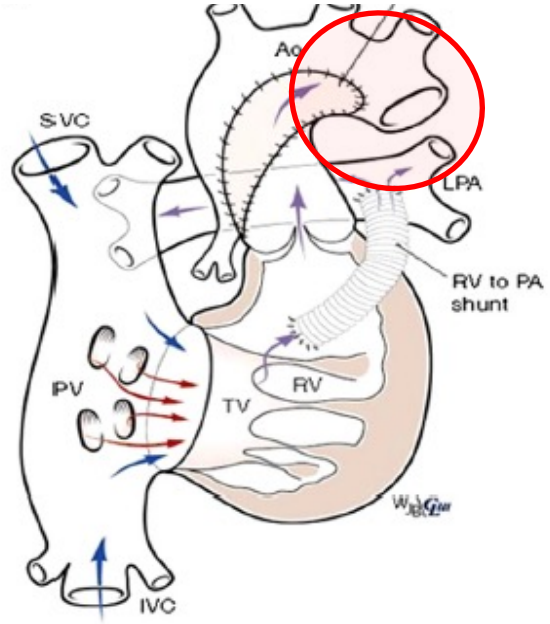


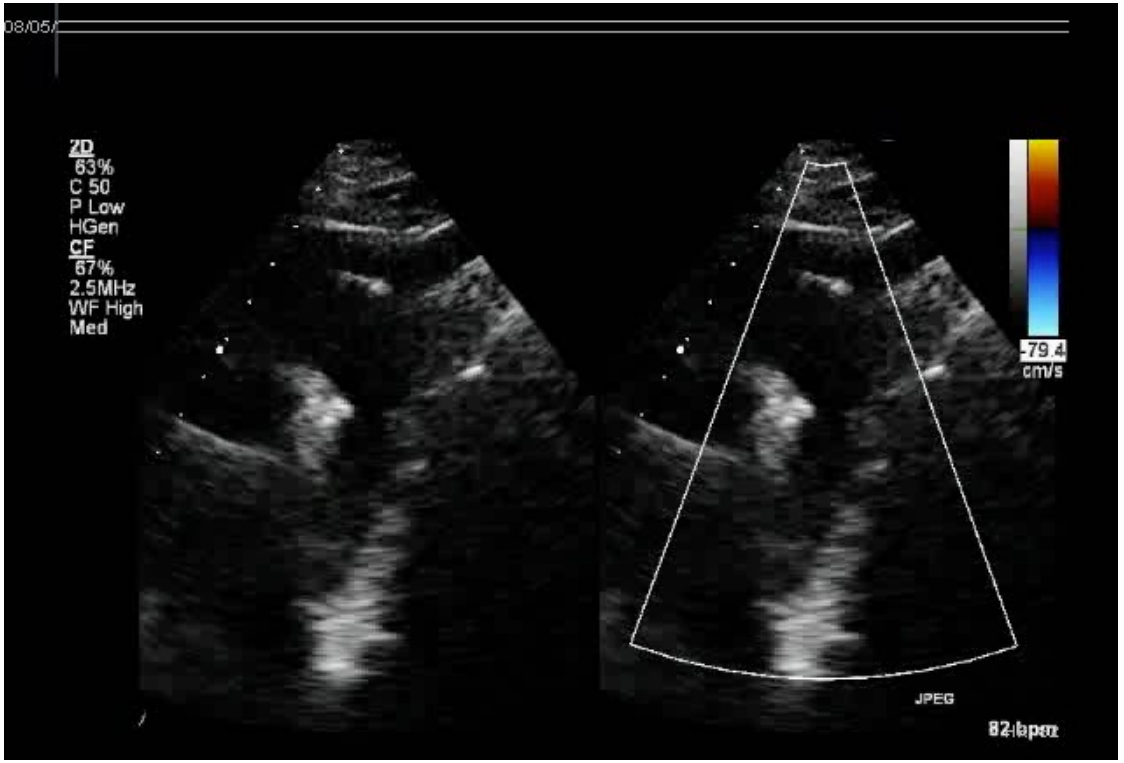




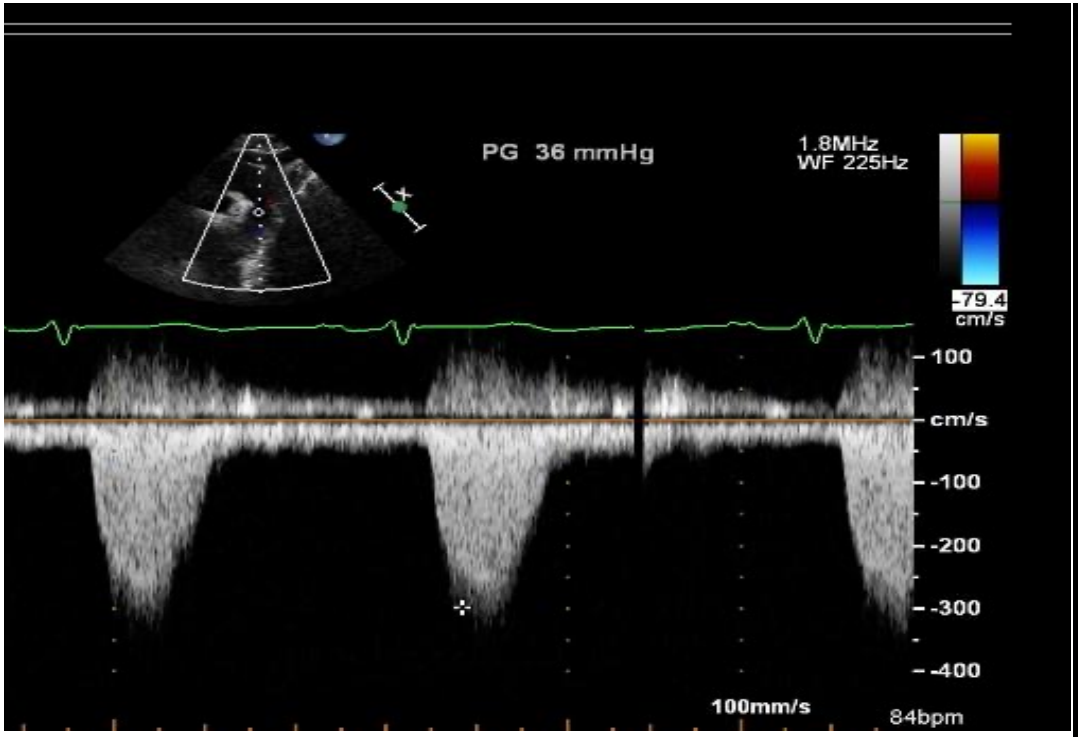
Neoaortic arch obstruction

- suprasternal sagittal view
- Increased flow velocity potentially related with change of caliber between large reconstructed neotransverse arch and native descending aorta

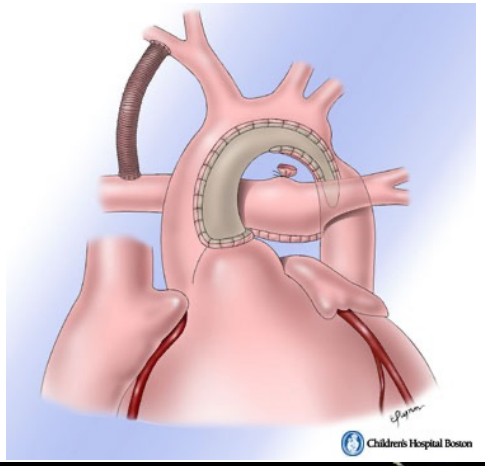


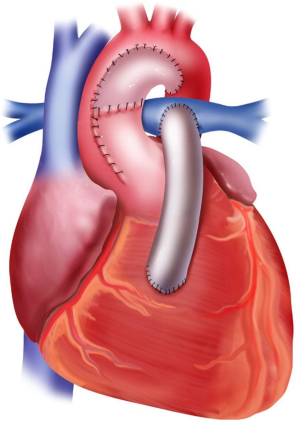


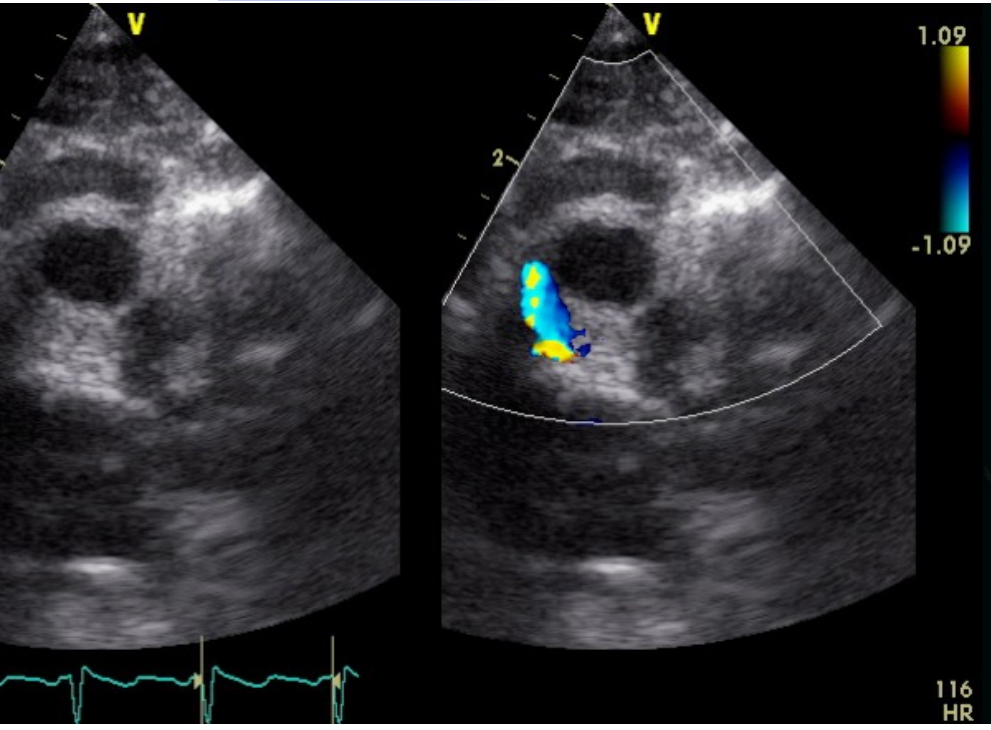
Neo aortic arch obstruction

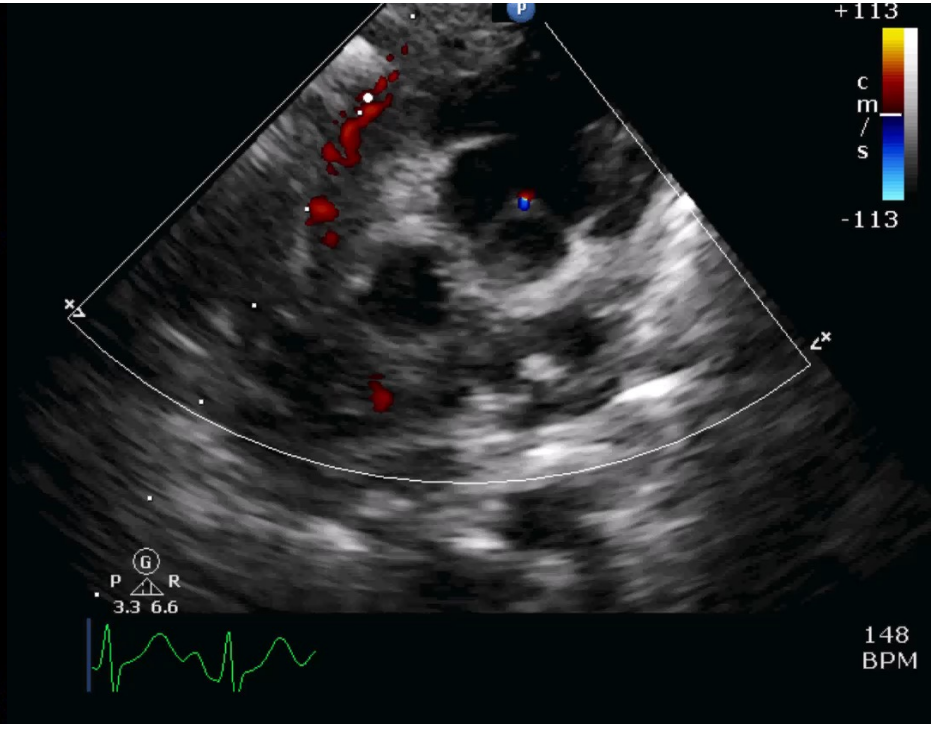


Different shunt types

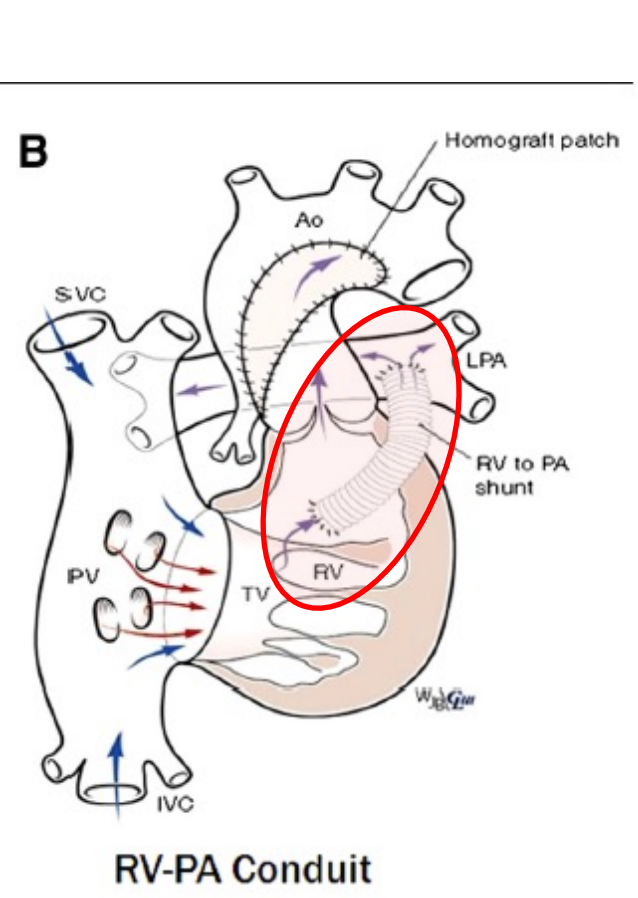


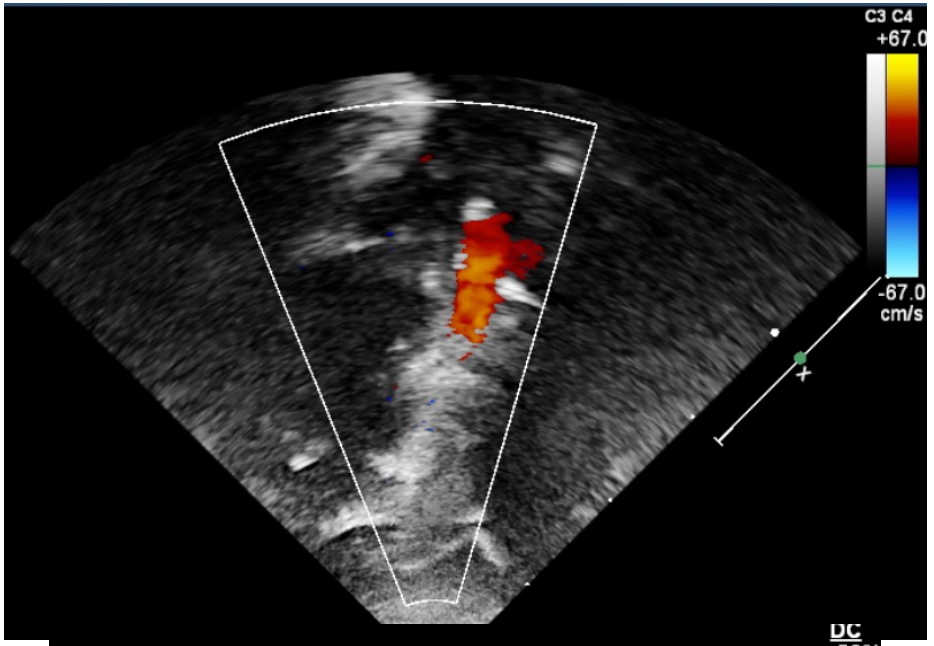


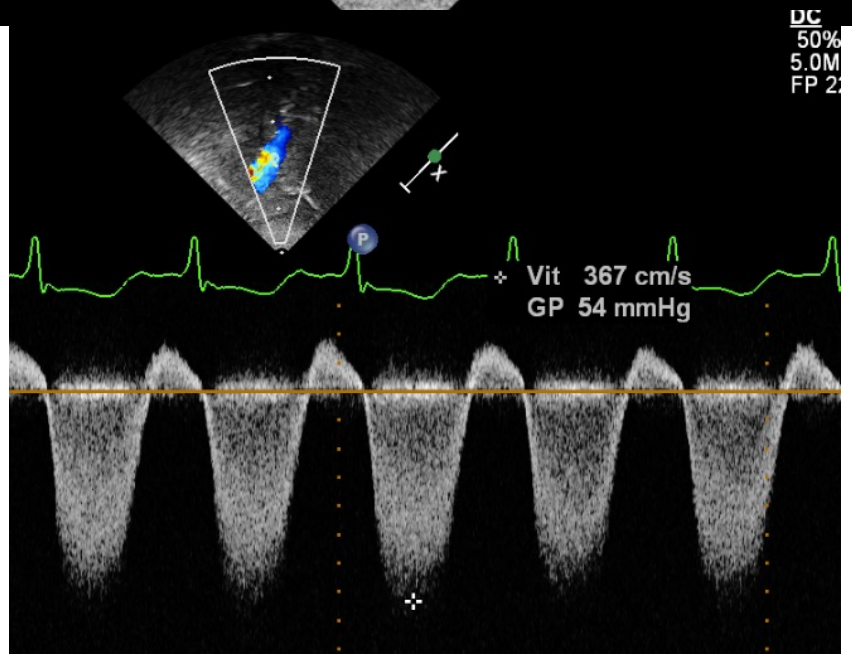


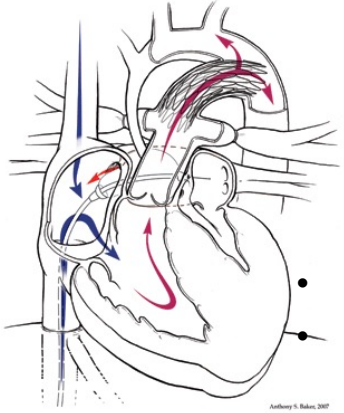


RV to PA conduit



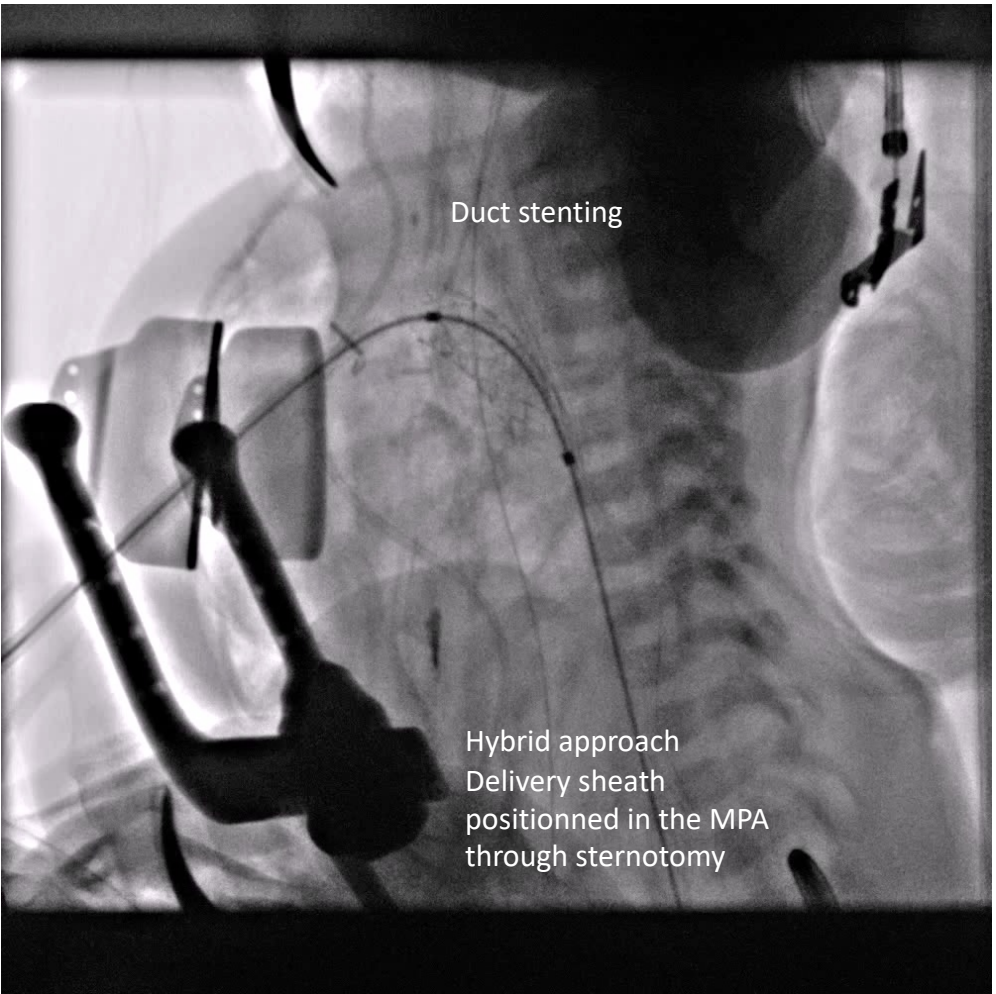


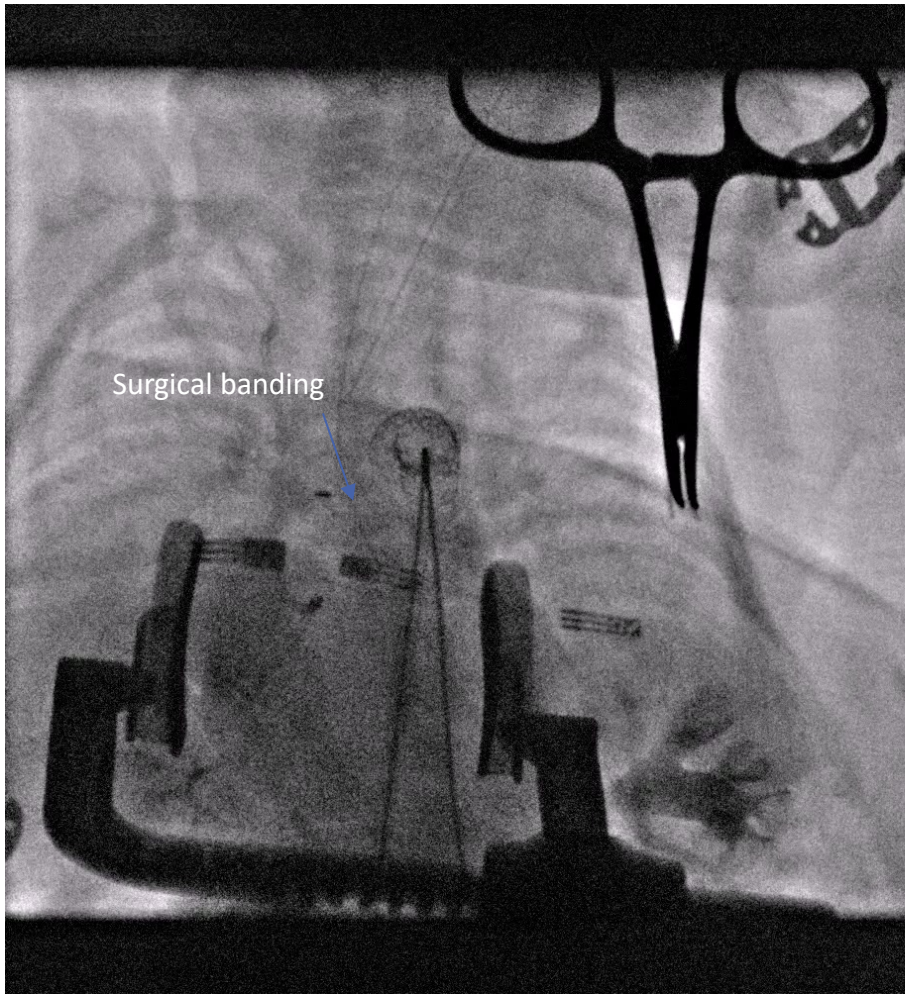




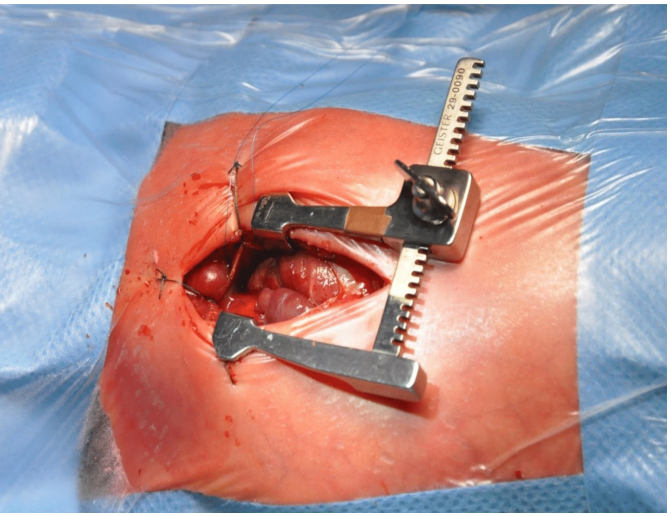
Hybrid palliation

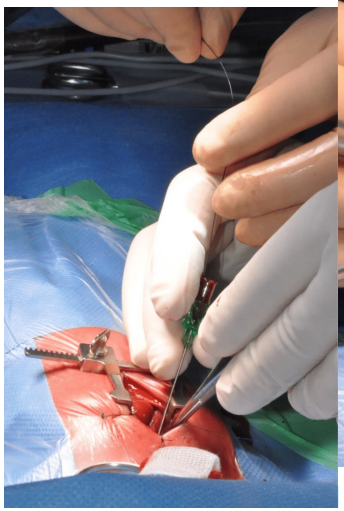
- create a balanced physiology where the single RV supplies the systemic and the pulmonary circulati
- complex aortic arch reconstruction (including CPB) is postponed to the stage 2 (Glenn)

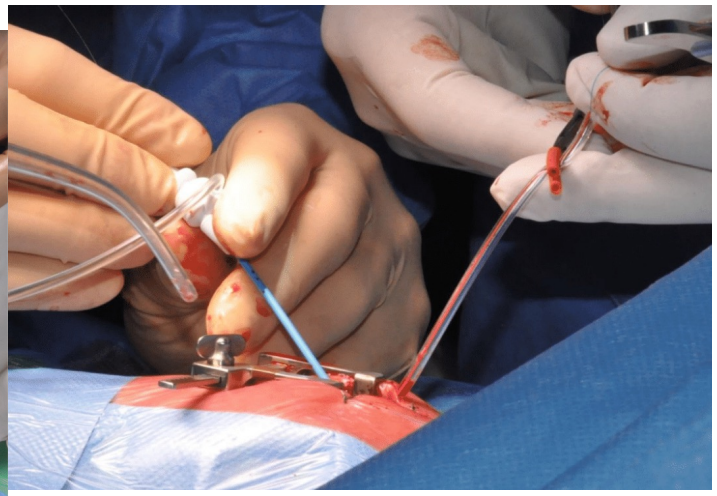


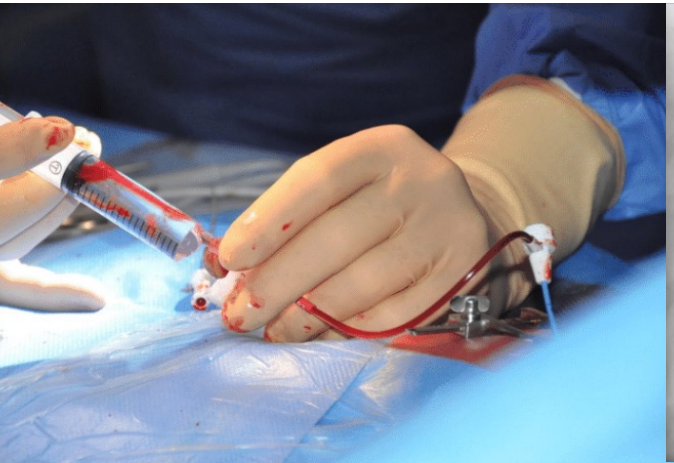


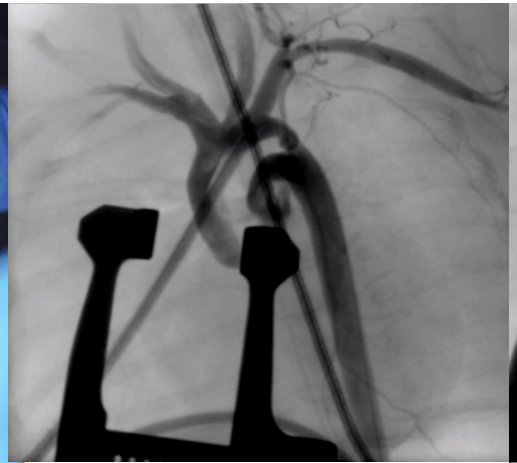
Hybrid procedure

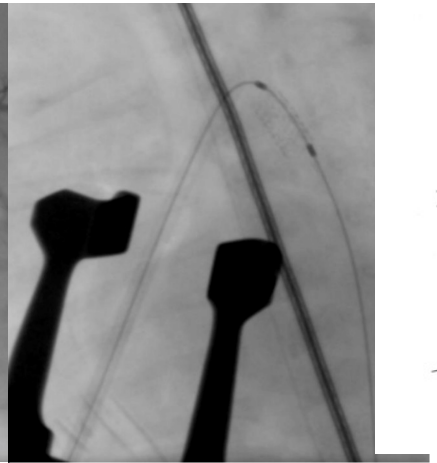


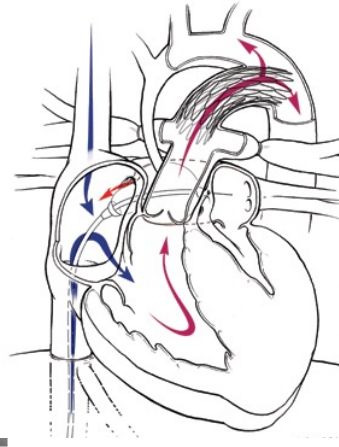




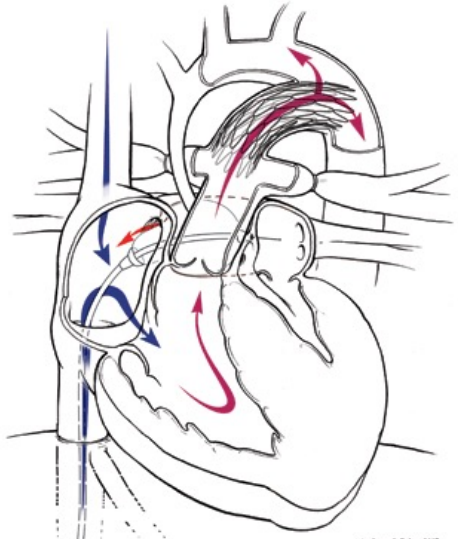


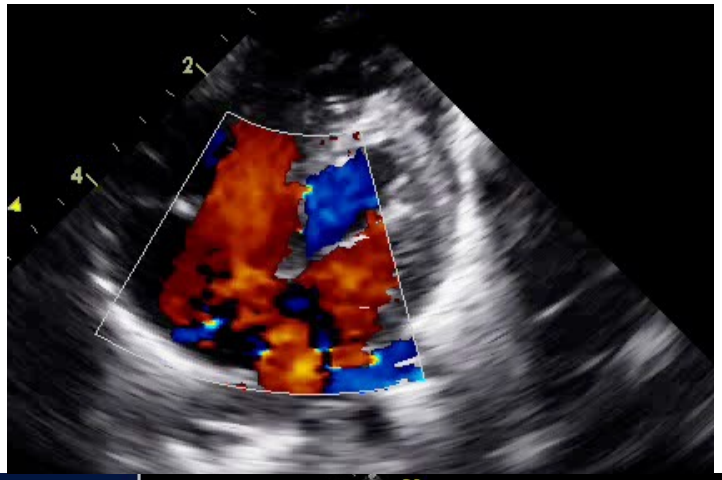


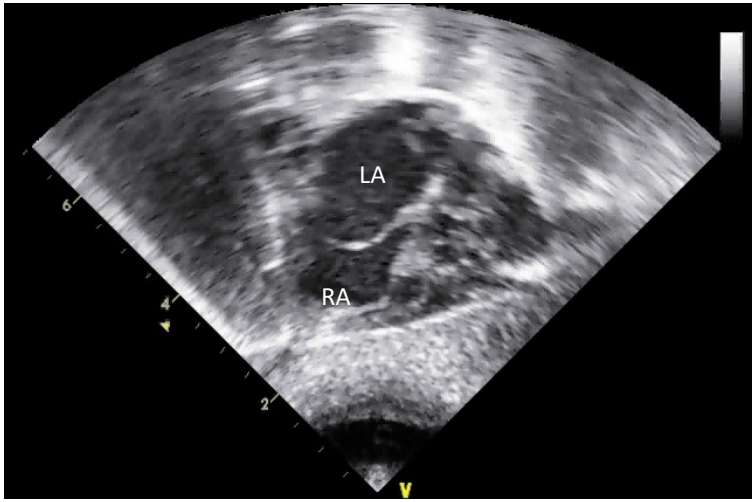


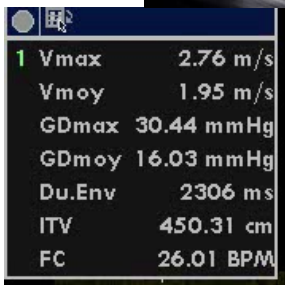


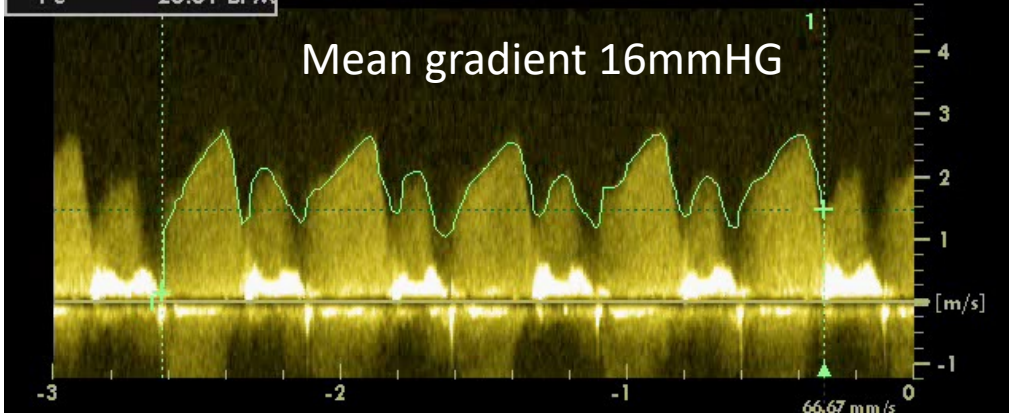
Restrictive PFO in HLHS



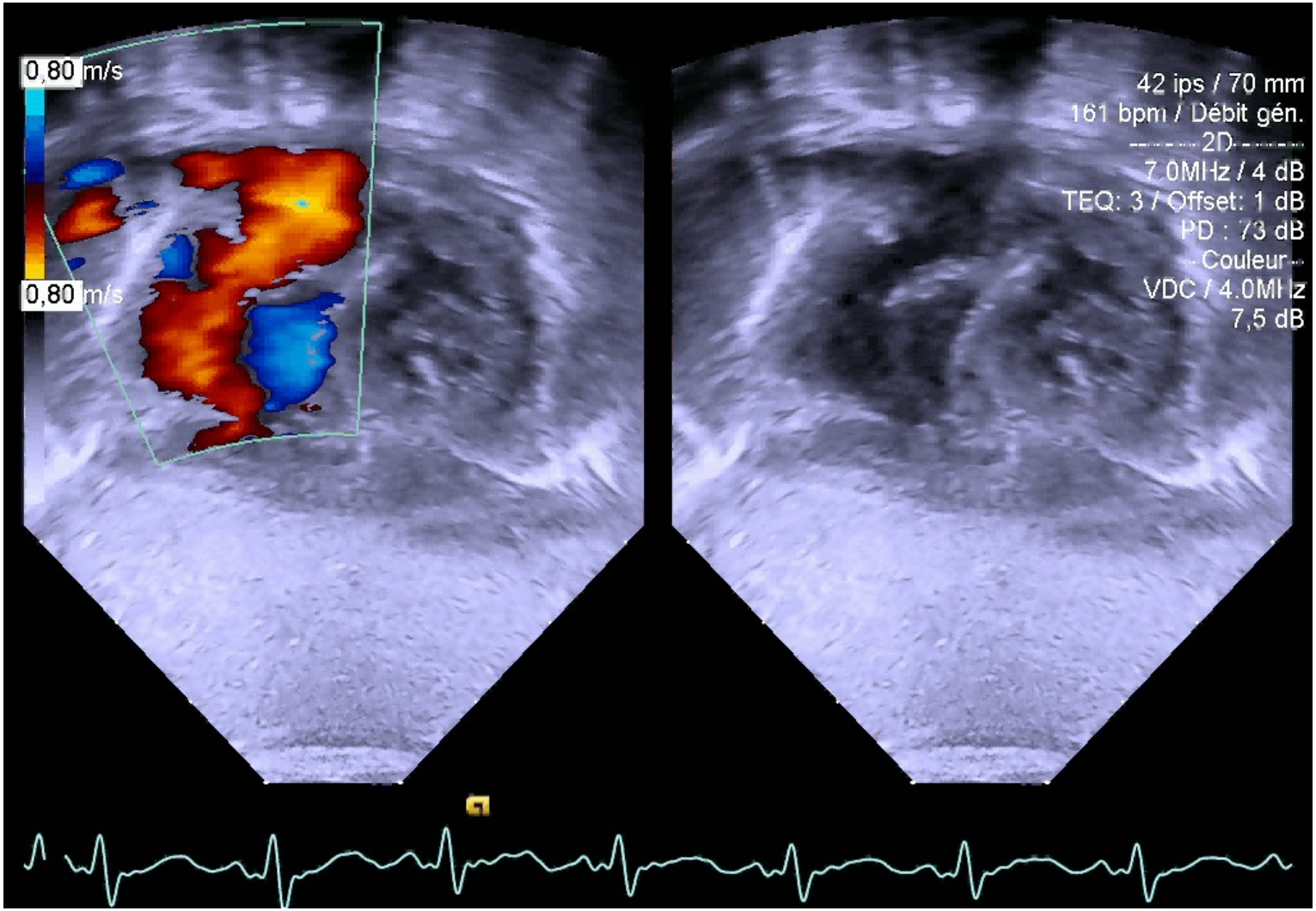


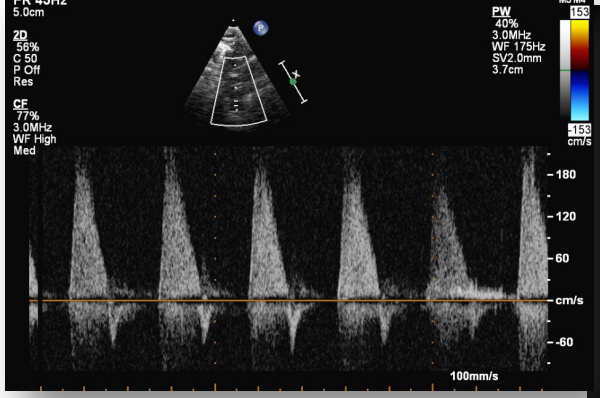


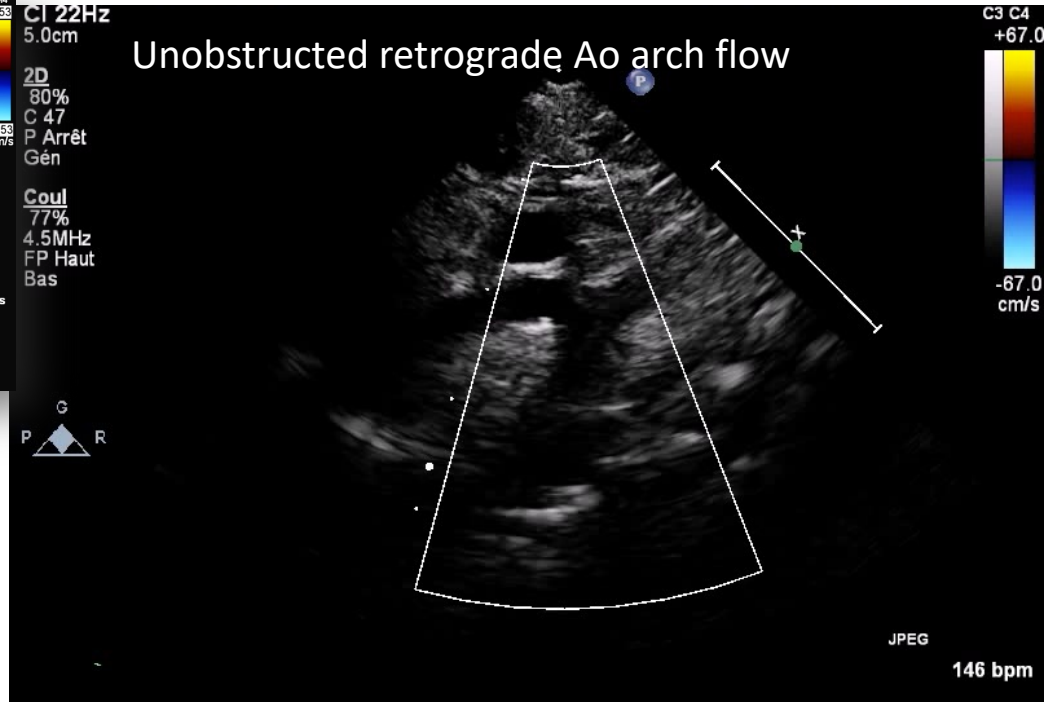




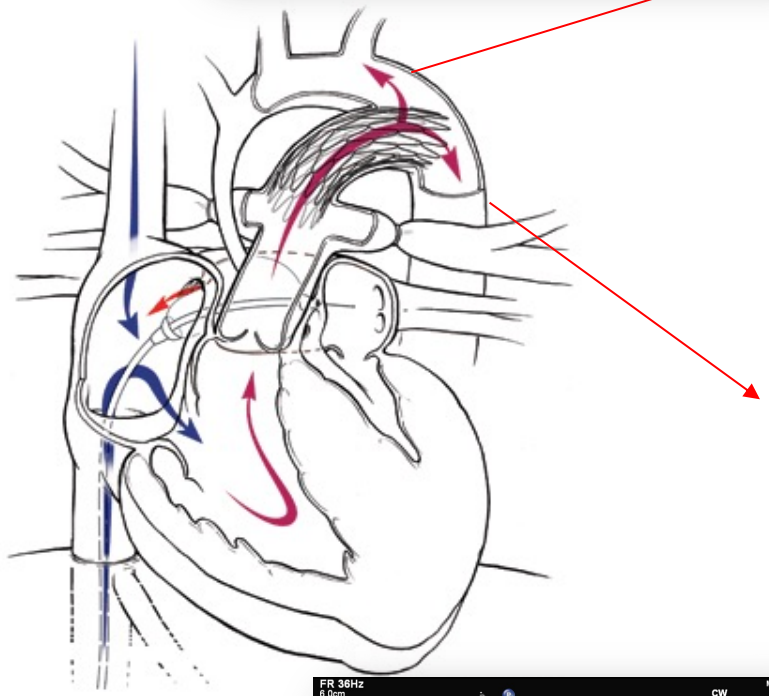
Same patient post balloon atrial septostomy

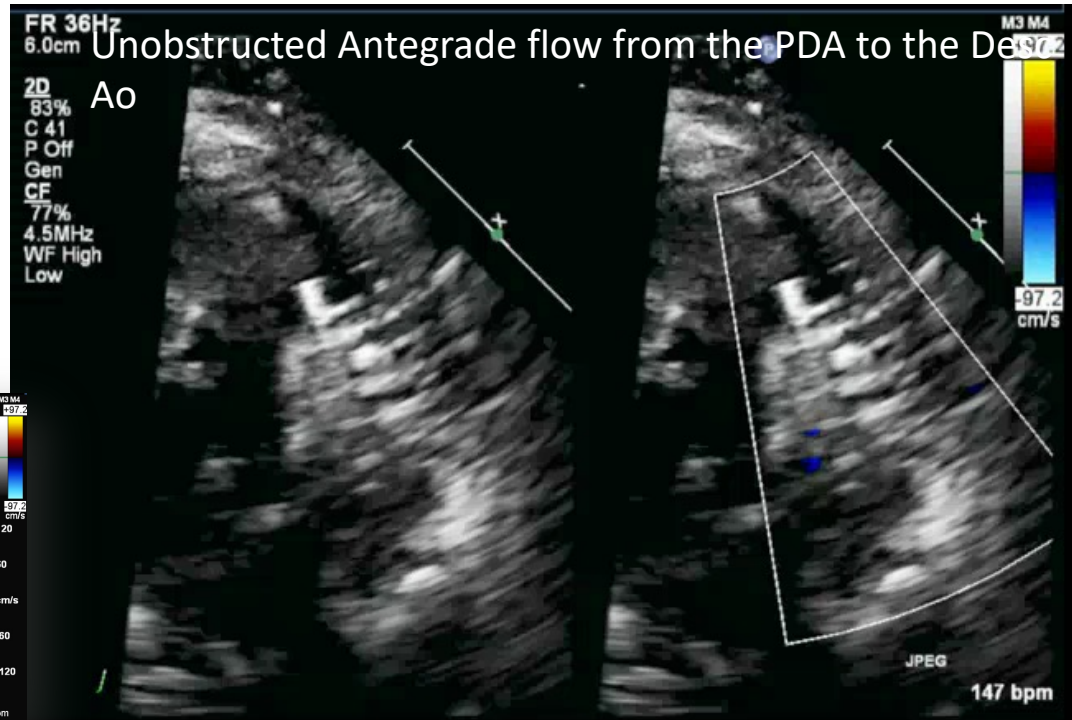




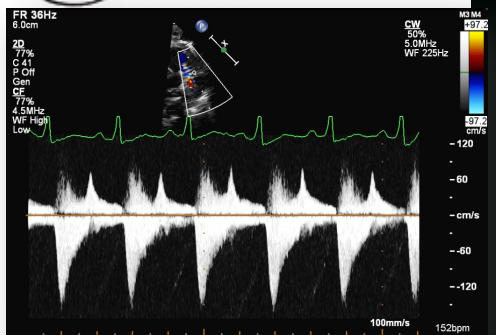


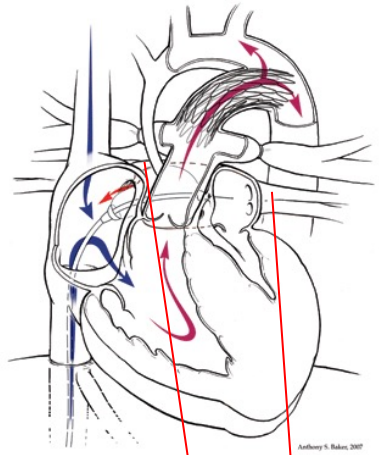
Unobstructed retrograde Ao arch flow

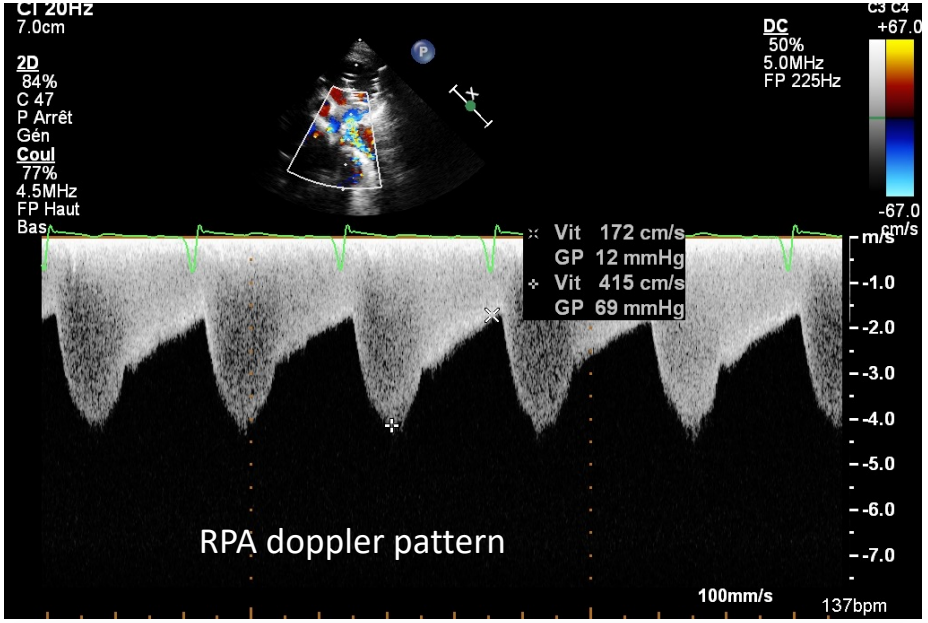


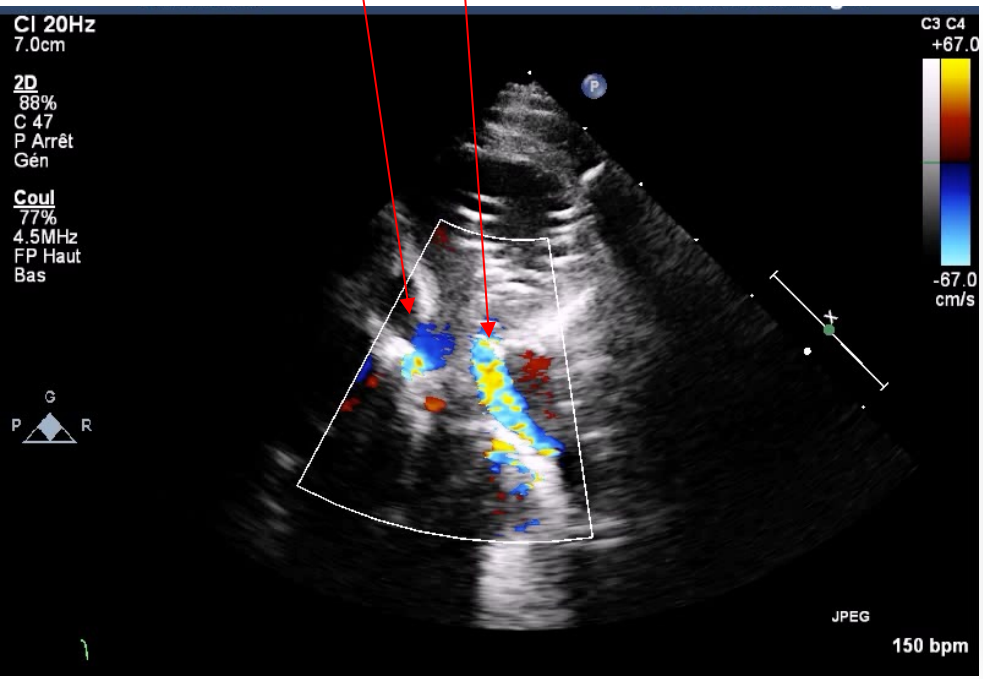


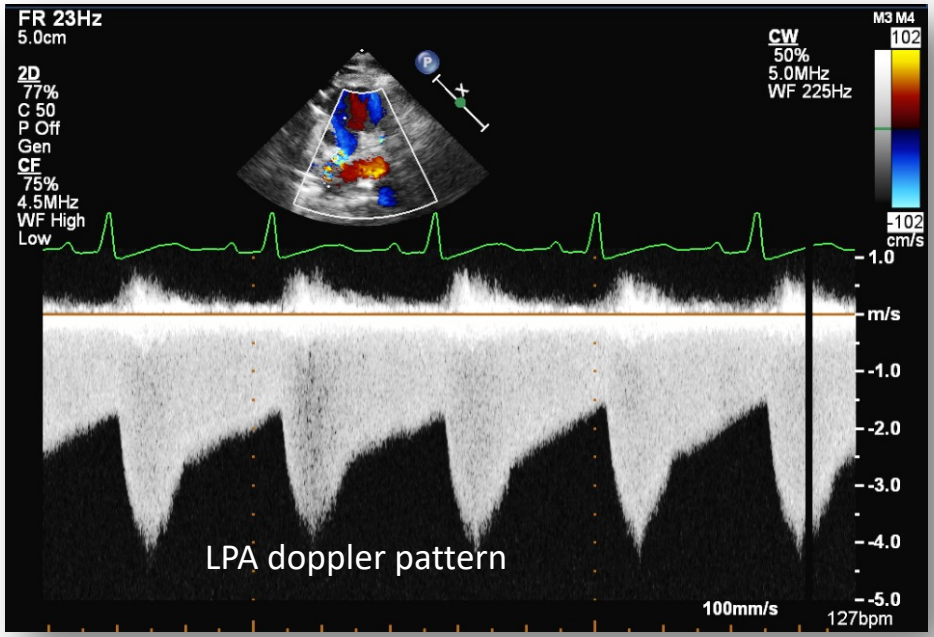
Unobstructed Antegrade flow from the PDA to the Desc Ao






CI 20Hz
7.0cm
2D
84%
C 47
P Arrêt
Gén
Coul
77%
4.5MHz
FP Haut
Bas
DC
50%
5.0MHz
FP 225Hz
C3 C4
+67.0
-67.0
cm/s
Vit 172 cm/s
GP 12 mmHg
Vit 415 cm/s
GP 69 mmHg
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
-6.0
-7.0
RPA doppler pattern
100mm/s
137bpm


CI 20Hz
7.0cm
2D
88%
C 47
P Arrêt
Gén
Coul
77%
4.5MHz
FP Haut
Bas
C3 C4
+67.0
-67.0
cm/s
G
P
R
JPEG
150 bpm


FR 23Hz
5.0cm
2D
77%
C 50
P Off
Gen
CF
75%
4.5MHz
WF High
Low
CW
50%
5.0MHz
WF 225Hz
M3 M4
102
-102
cm/s
1.0
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
LPA doppler pattern
100mm/s
127bpm

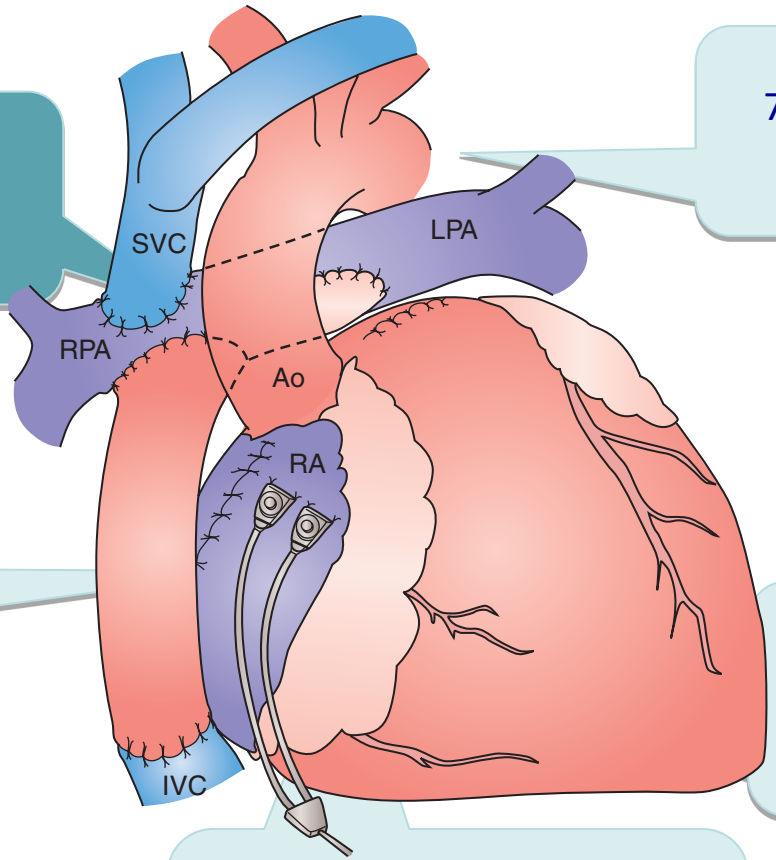
Echographic assessment after Glenn and TCPC completion: sequential segmental approach

1. Cavopulmonary connection
2. Branch PAs

PRELOAD

3. Pulmonary veins

4. Atrial septum



7. Systemic outflow tract
8.Aortic arch

AFTERLOAD

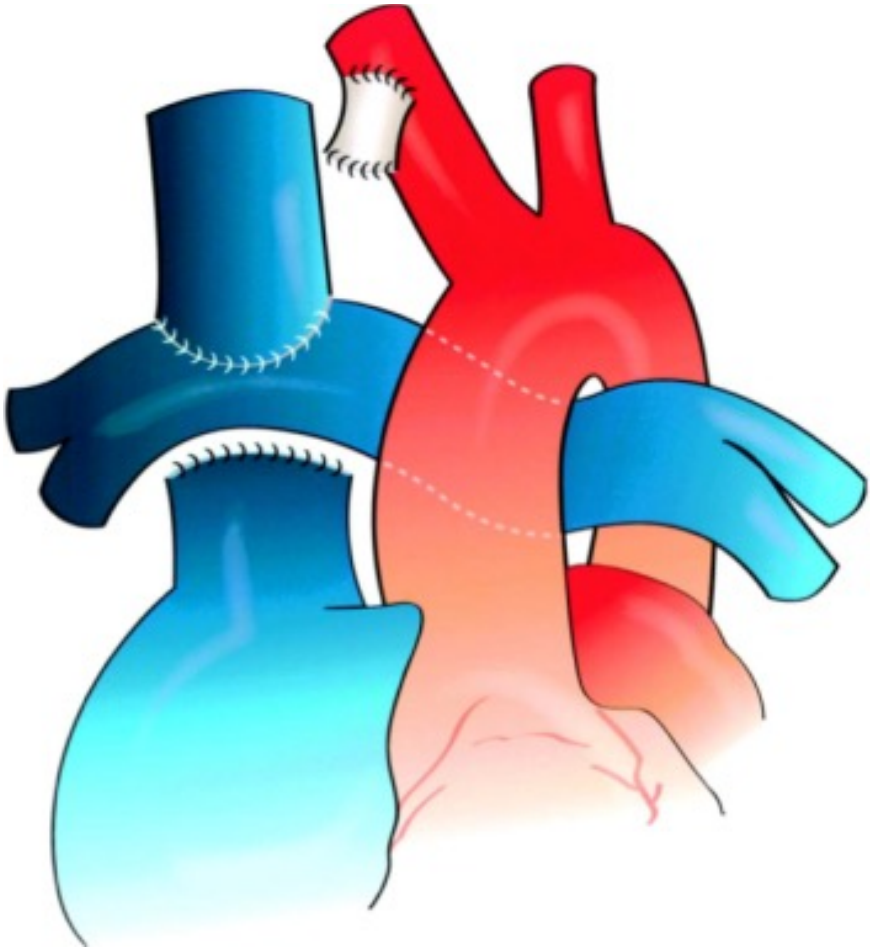
6.Systemic ventricular function

FUNCTION

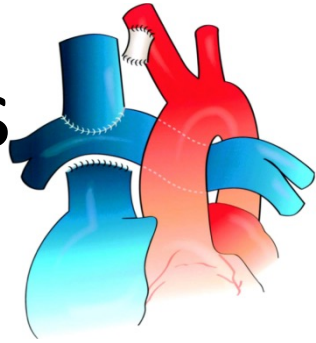
5. Atrioventricular valves

Glenn anastomosis

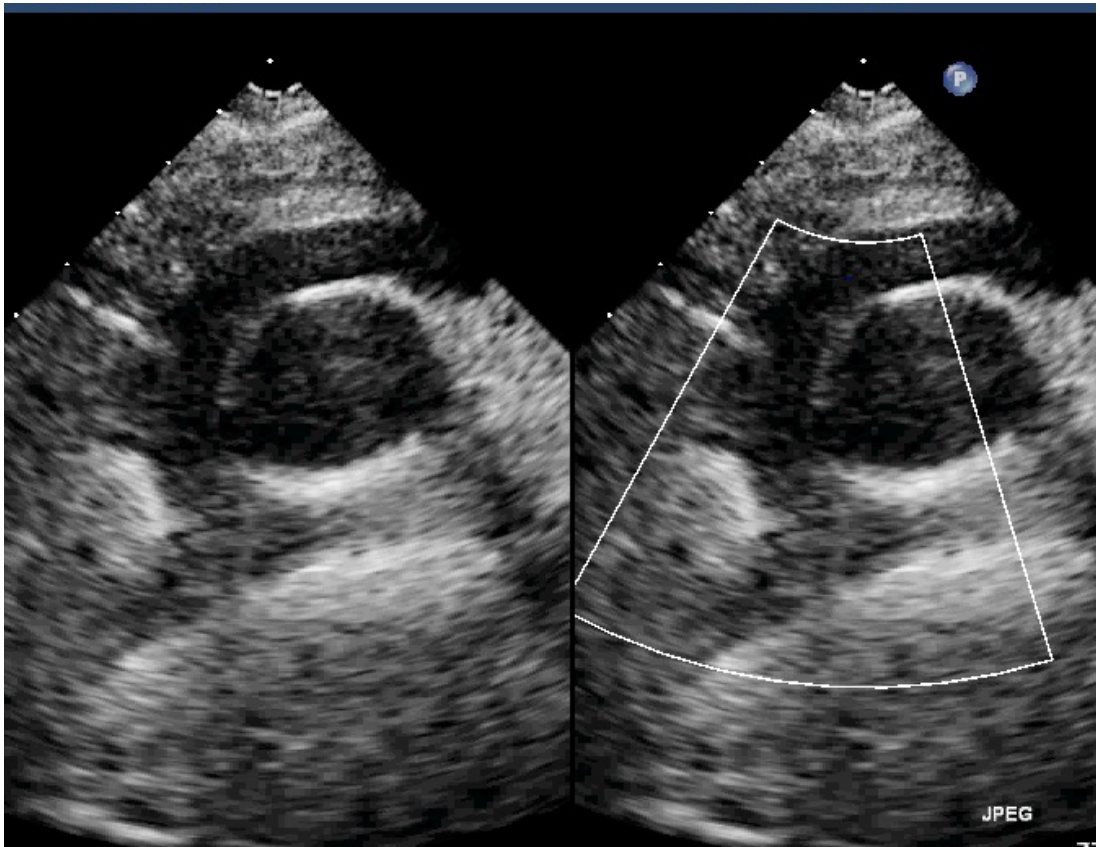
1. Cavopulmonary connection
2. Branch PA

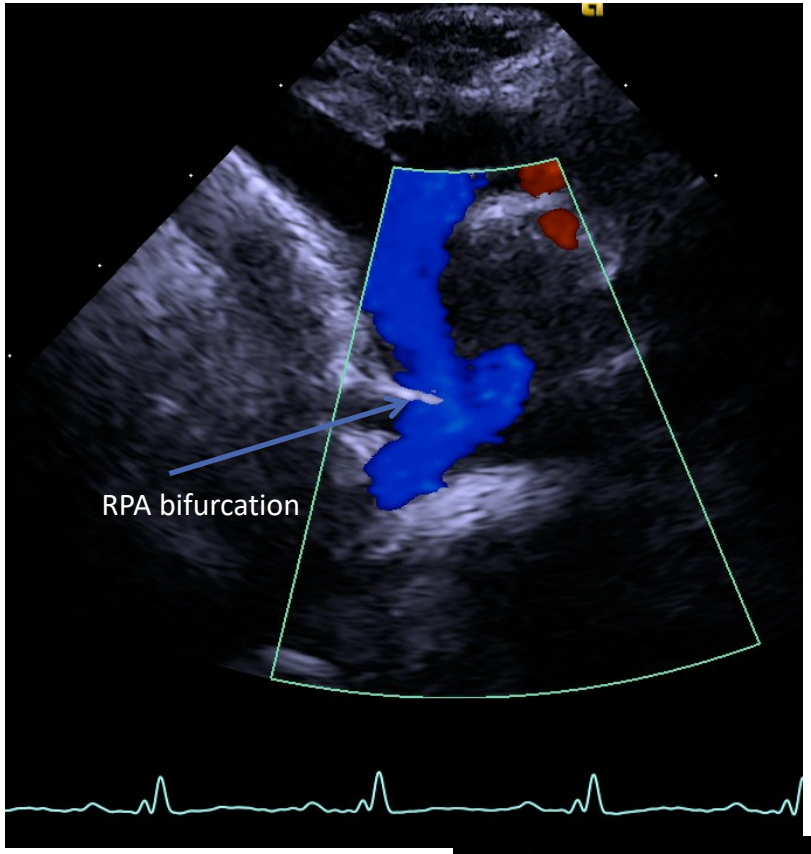


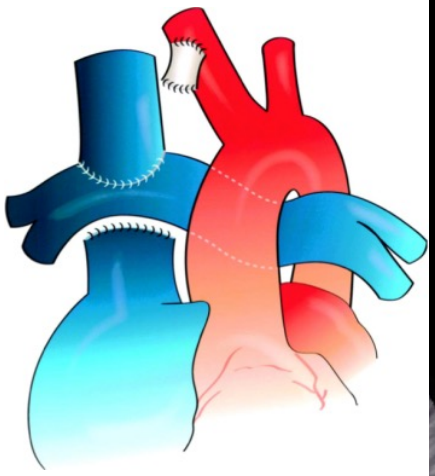
Assessment of the Glenn anastomos

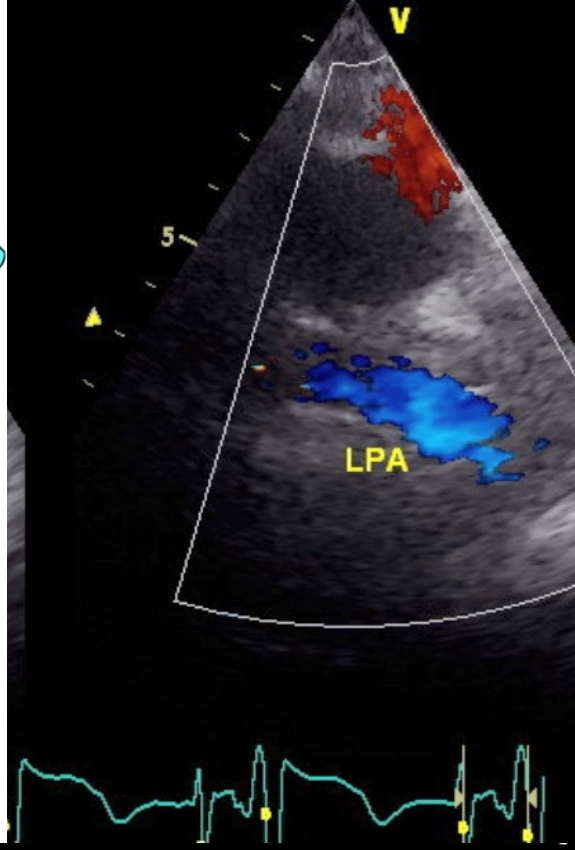


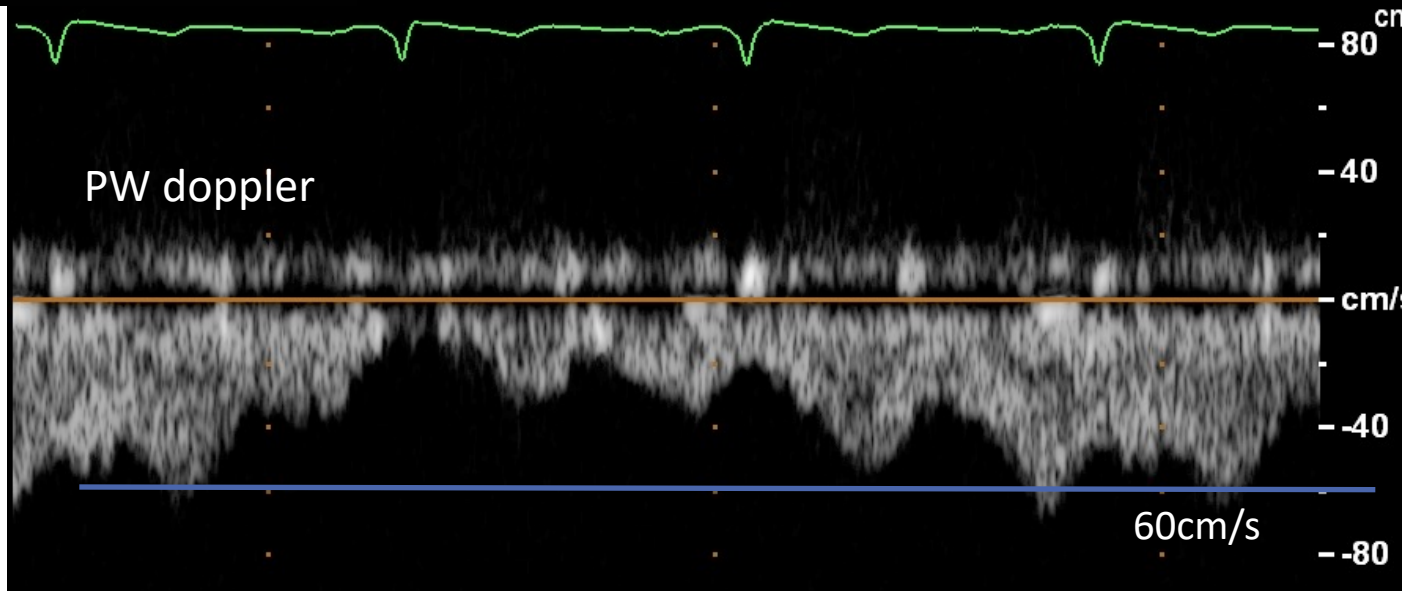
- Suprasternal / high parasternal view
- Laminar flow of low velocity with respiratory variation (adapt the velocity range+++)
- Rule out stenosis at the anastomosis site

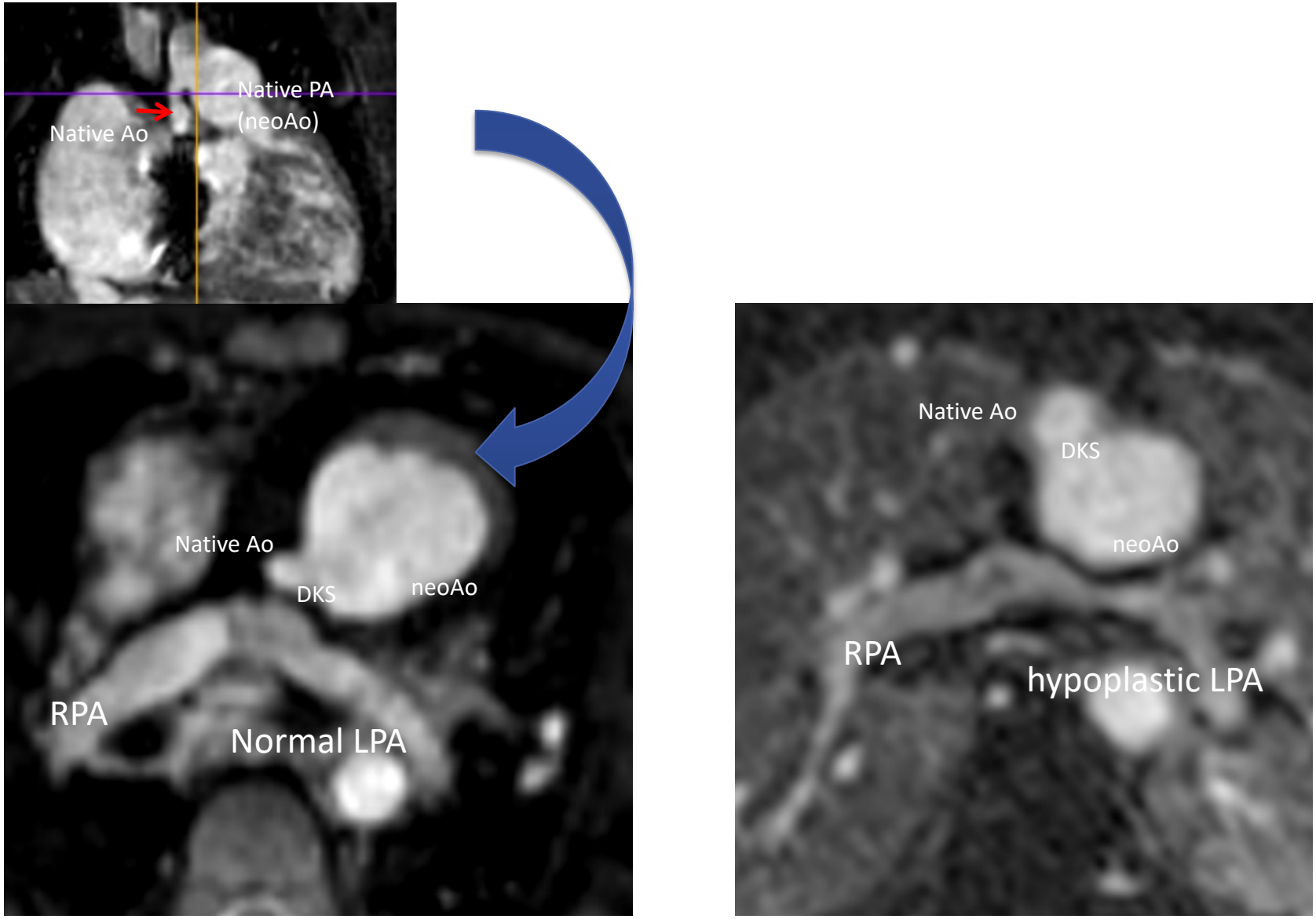



RPA bifurcation

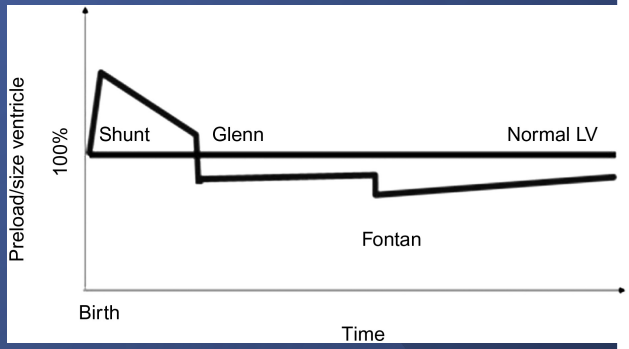



V
5
LPA


80
40
cm/
-40
-80
PW doppler
60cm/s

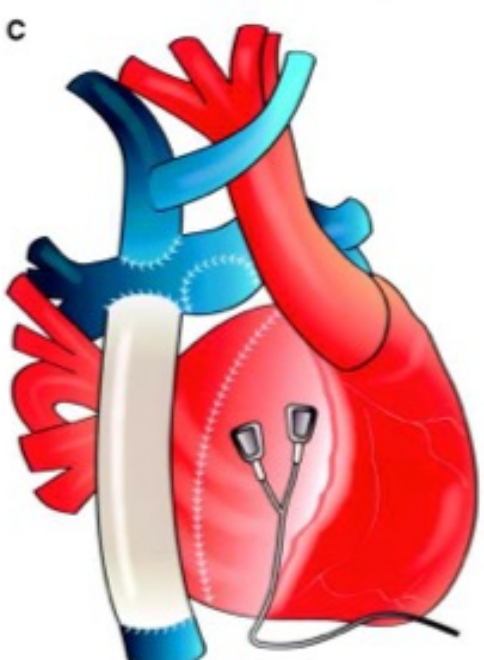


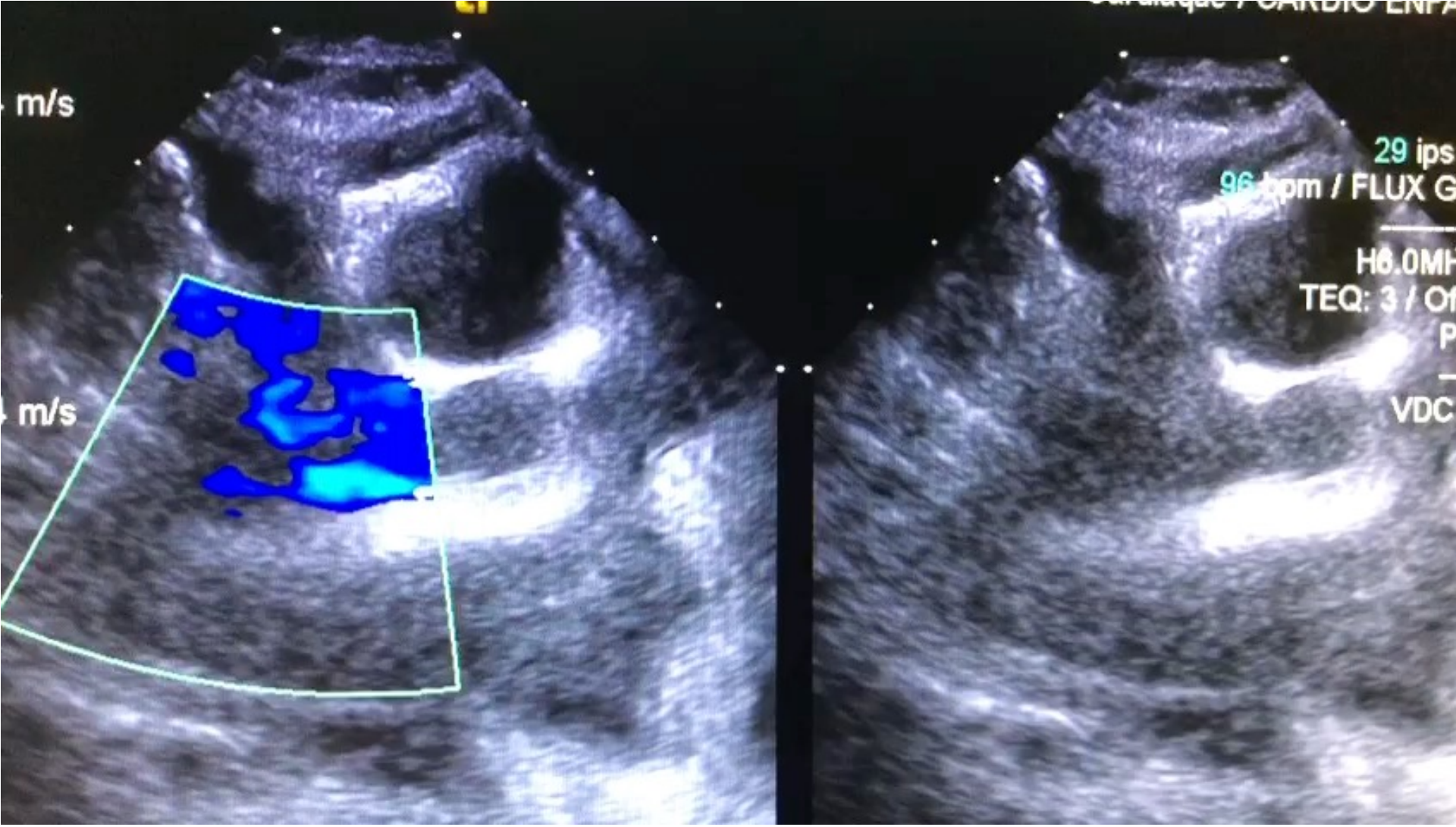
Look at the portion of the PA behind the reconstructed neo aorta

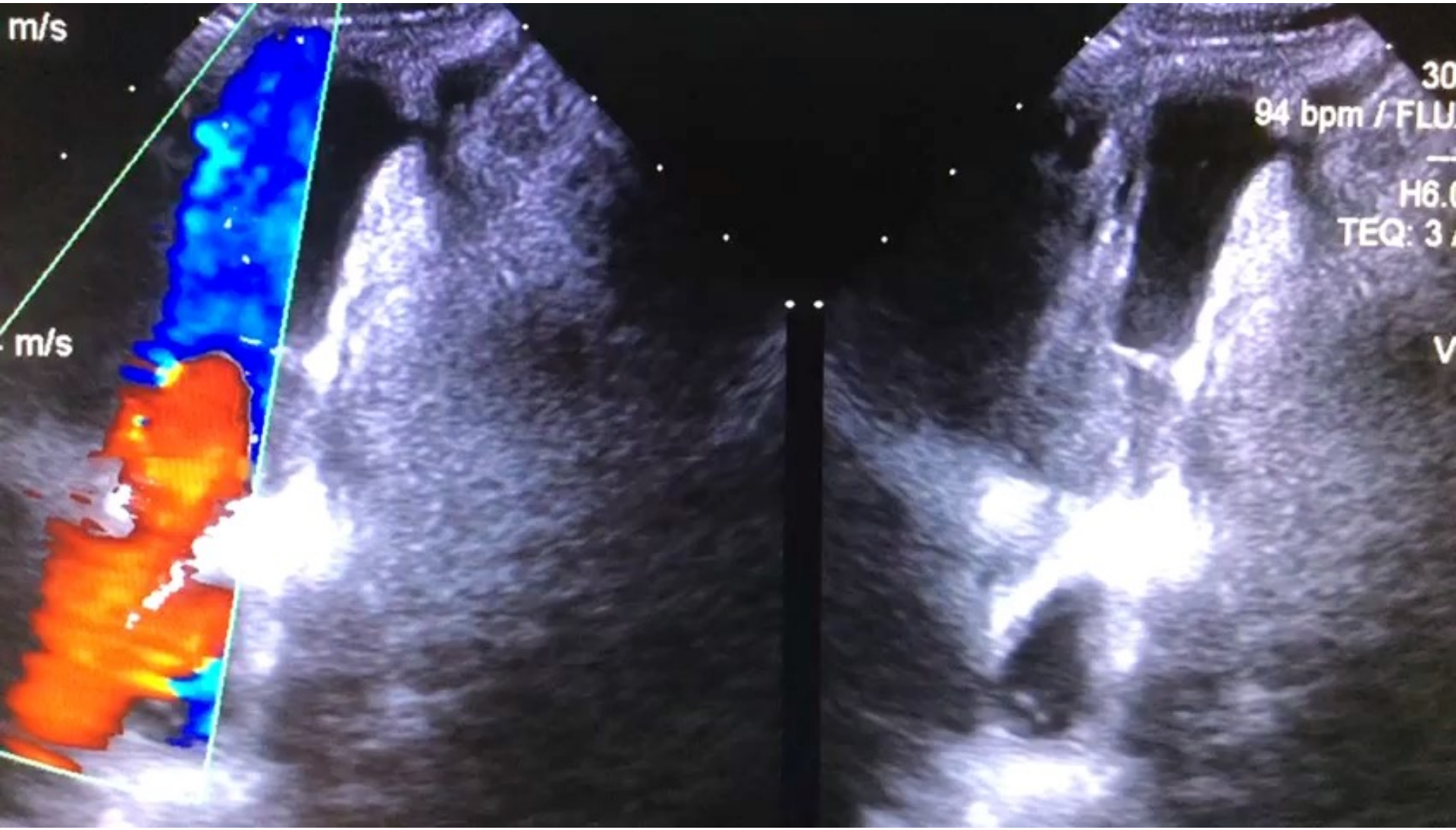


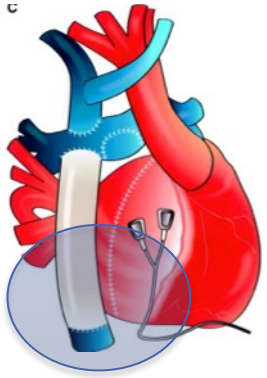
stage 3: TCPC

- SVC and Glenn anastomosis
- IVC to PA conduit assessment
- Conduit fenestration
- IVC and HV flow
- Thrombus in the Fontan pathway



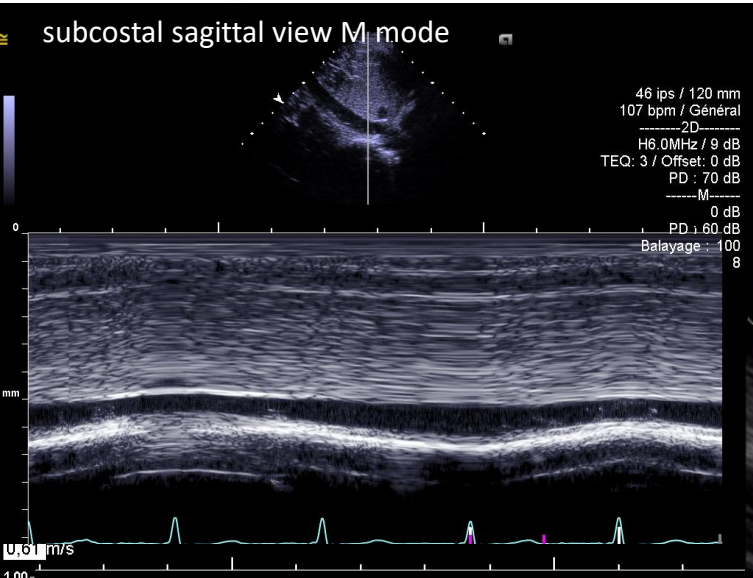

m/s
m/s
29 ips
FLUX G
TEQ: 3 / Of
VDC

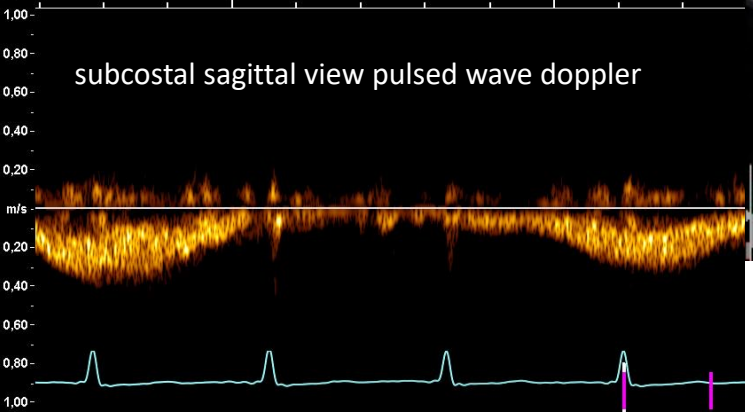

m/s
m/s
94 bpm / FLU
TEQ: 3

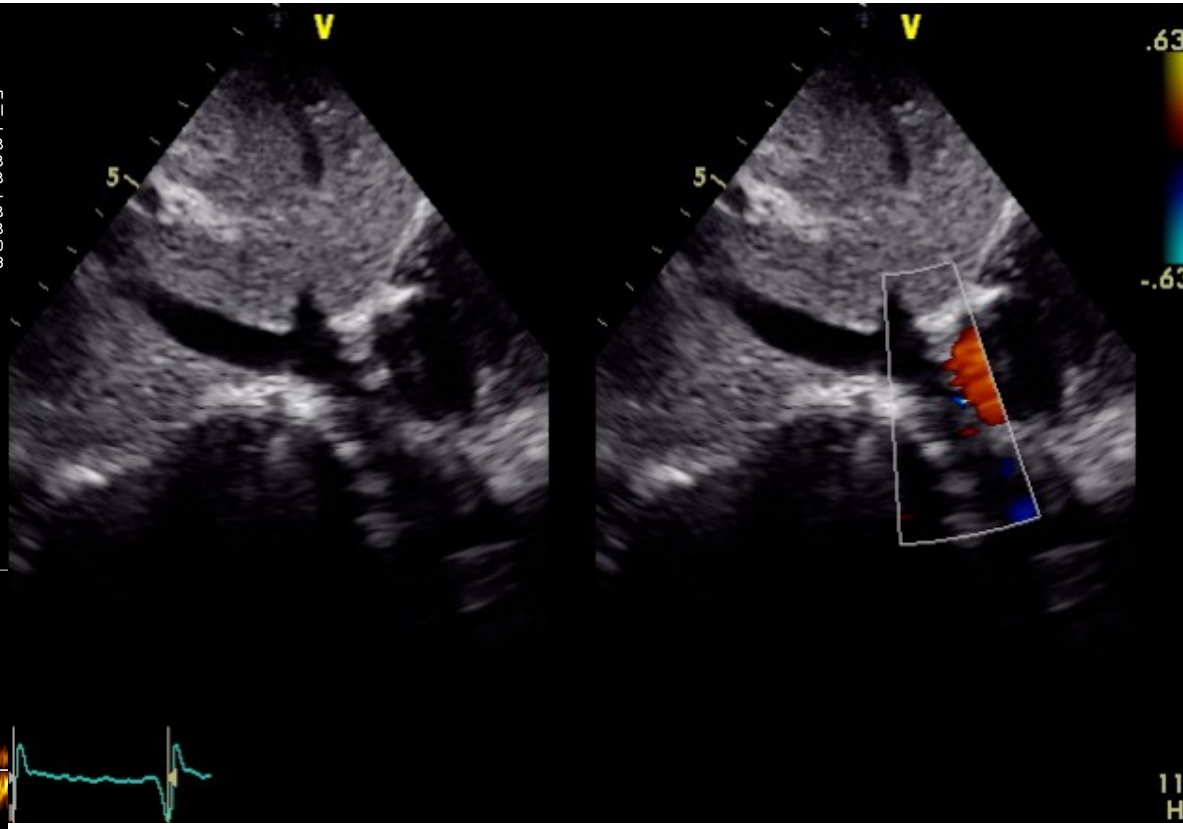


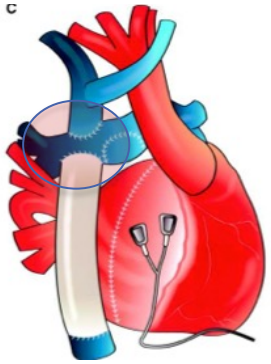
TCPC

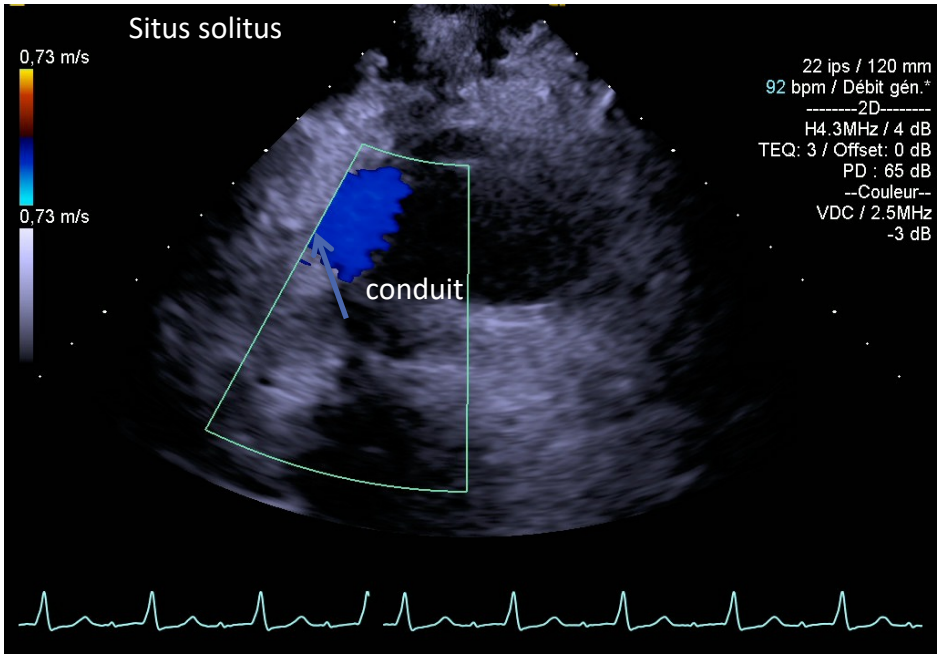
Proximal connection subcostal view

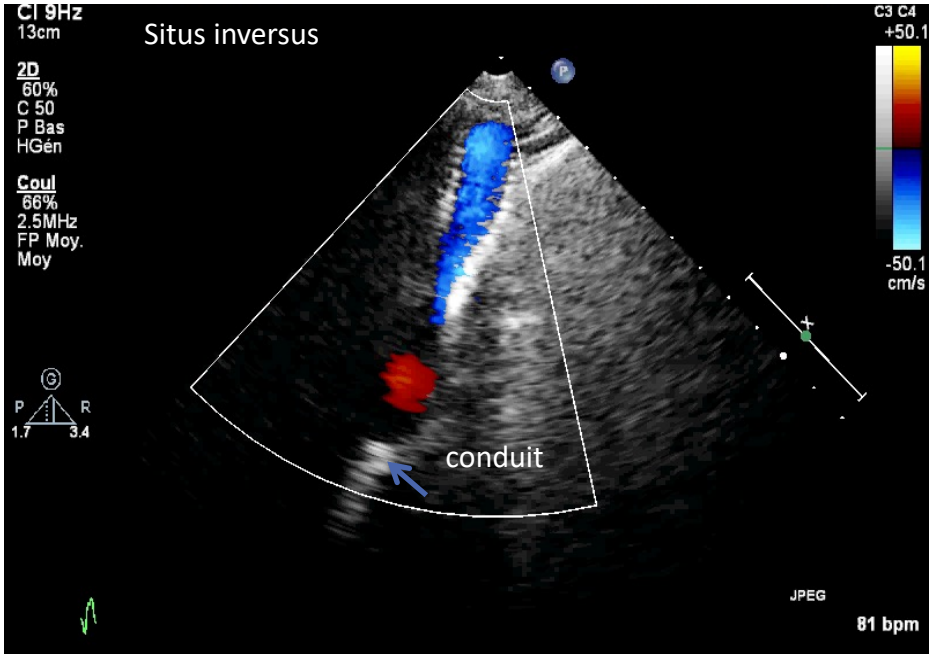




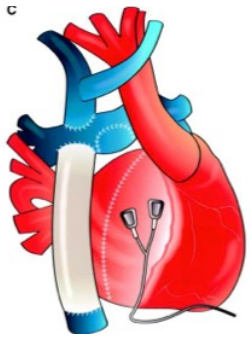


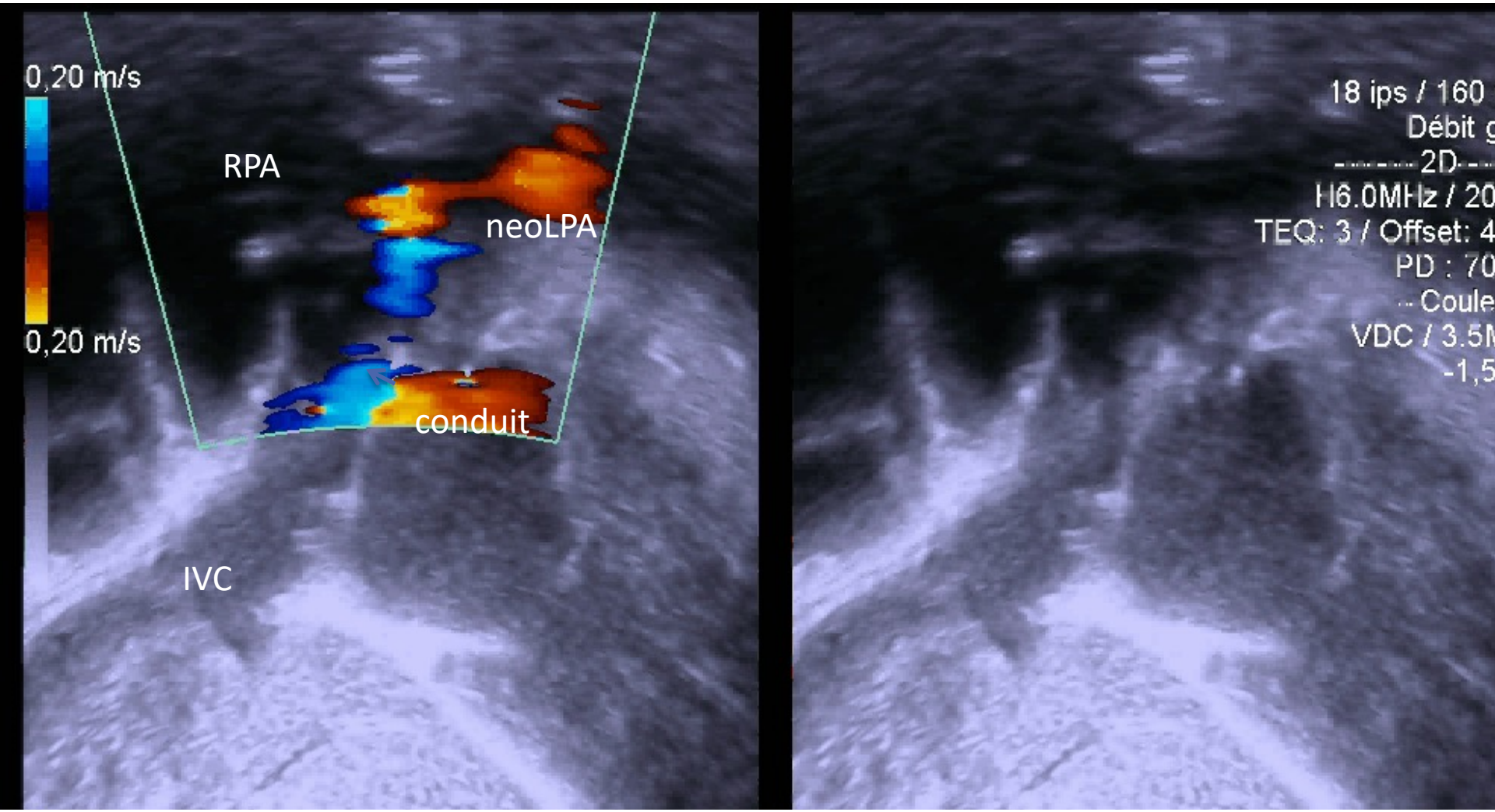


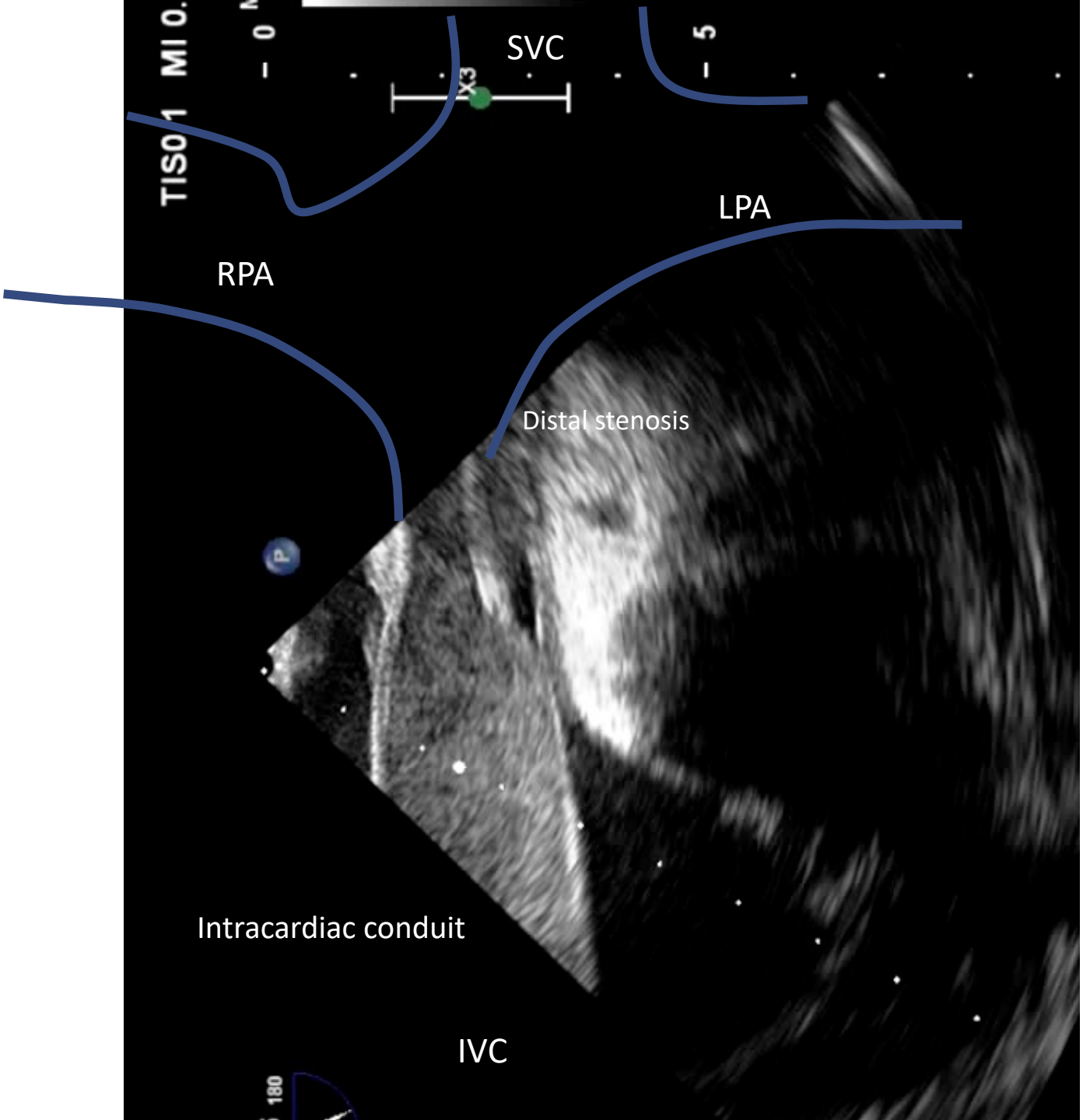




Conduit to PA distal connexion: high parasternal view

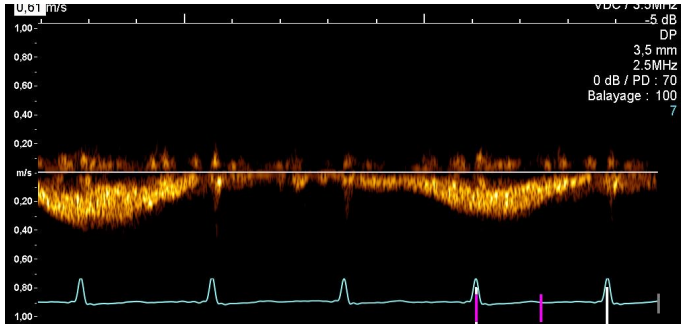



0,20 m/s
RPA
neoLPA
0,20 m/s
conduit
IVC
18 ips / 160
Débit
2D
H6.0MHz / 20
TEQ: 3 / Offset: 4
PD : 70
Coule
VDC / 3.5
-1,5

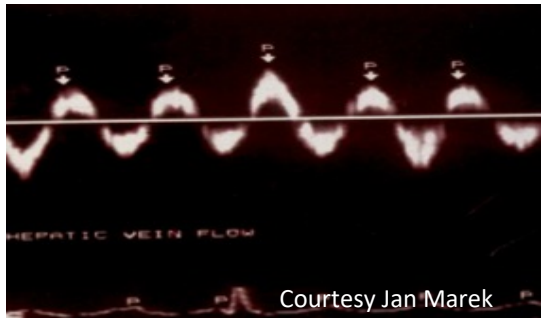

SVC
LPA
RPA
Distal stenosis
Intracardiac conduit
IVC

IVC flow after TCPC

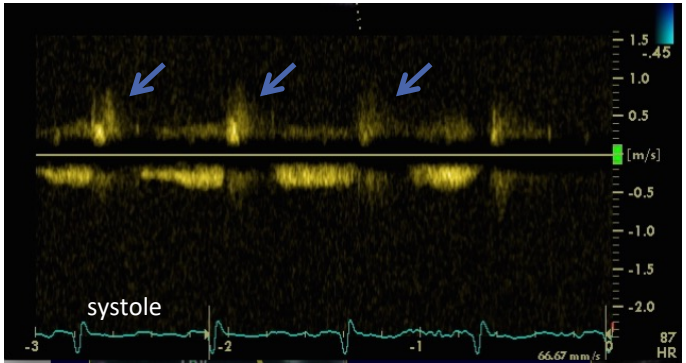
- Normal: continous anterograde flow of low velocity, respiratory variation



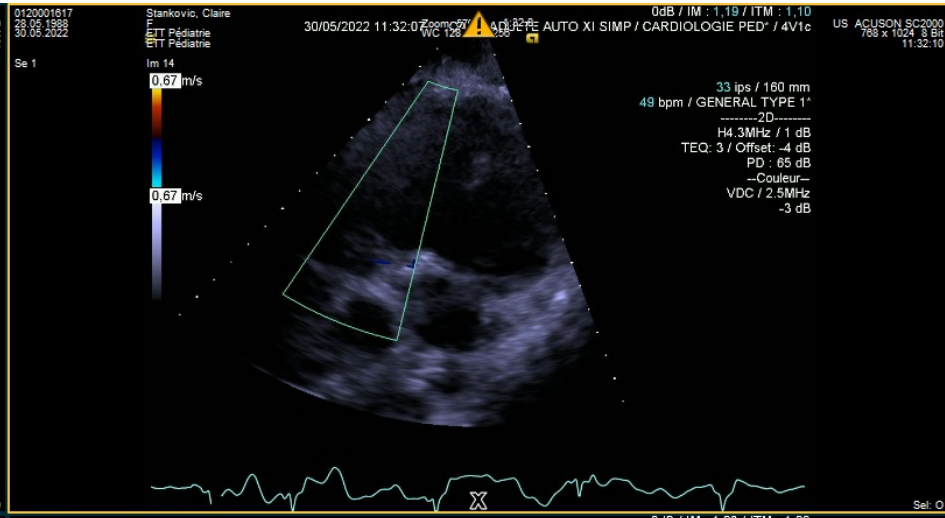
- Retrograde A wave:
  - failing fontan (↗CVP-arrythmia)

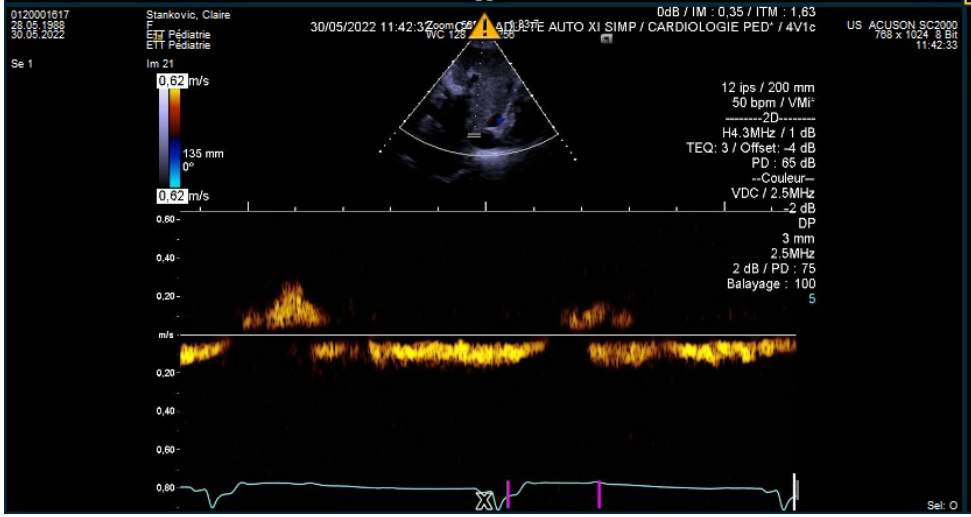


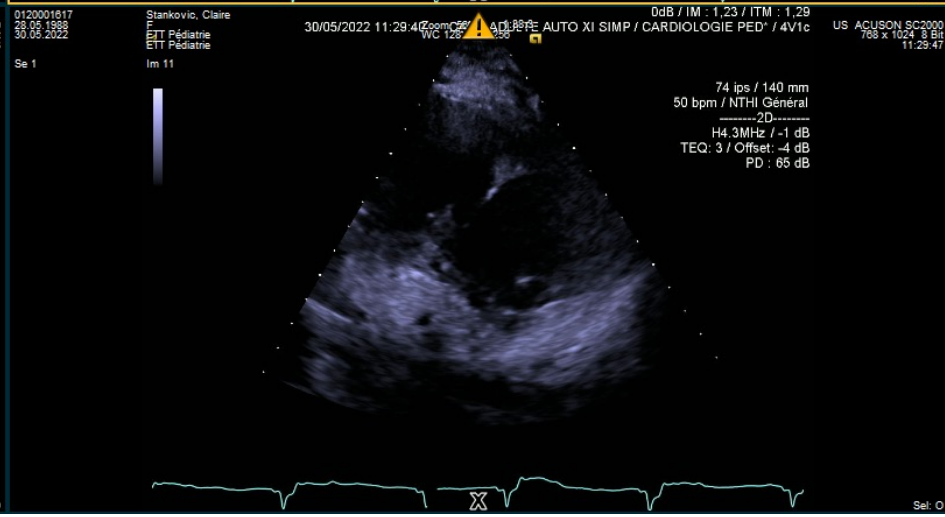
- Retrograde S wave:
  - AV regurgitation: IVC
  - Antegrade flow (pulm stenosis): SVC



Failing fontan IVC doppler pattern

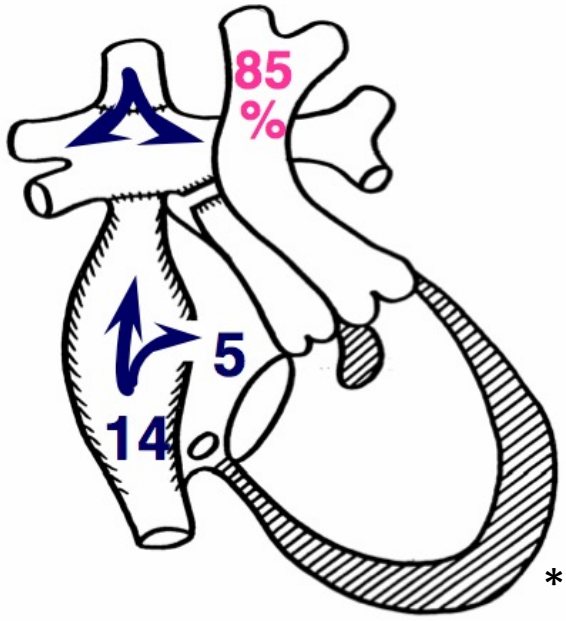


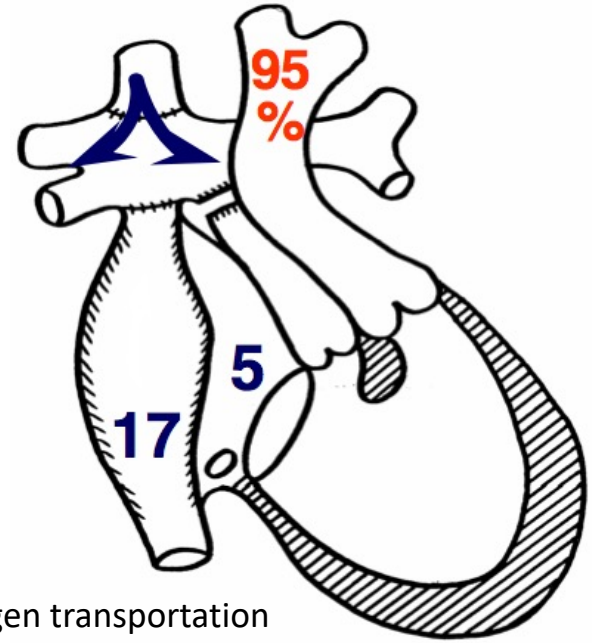




Fenestration assessement

- Right to left shunt
  - Decompress the systemic venous pathway
  - Maintain cardiac output



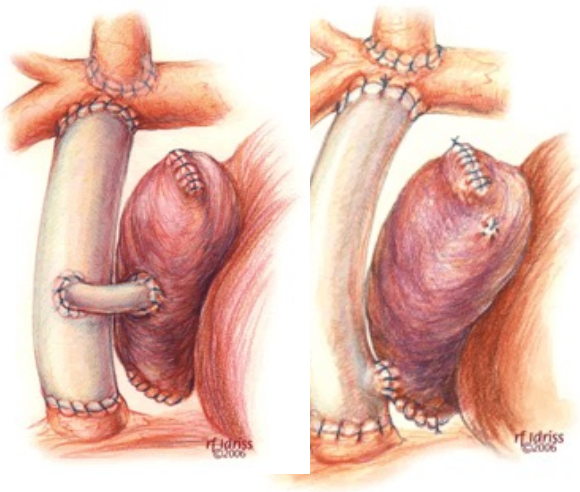


* Systemic oxygen transportation

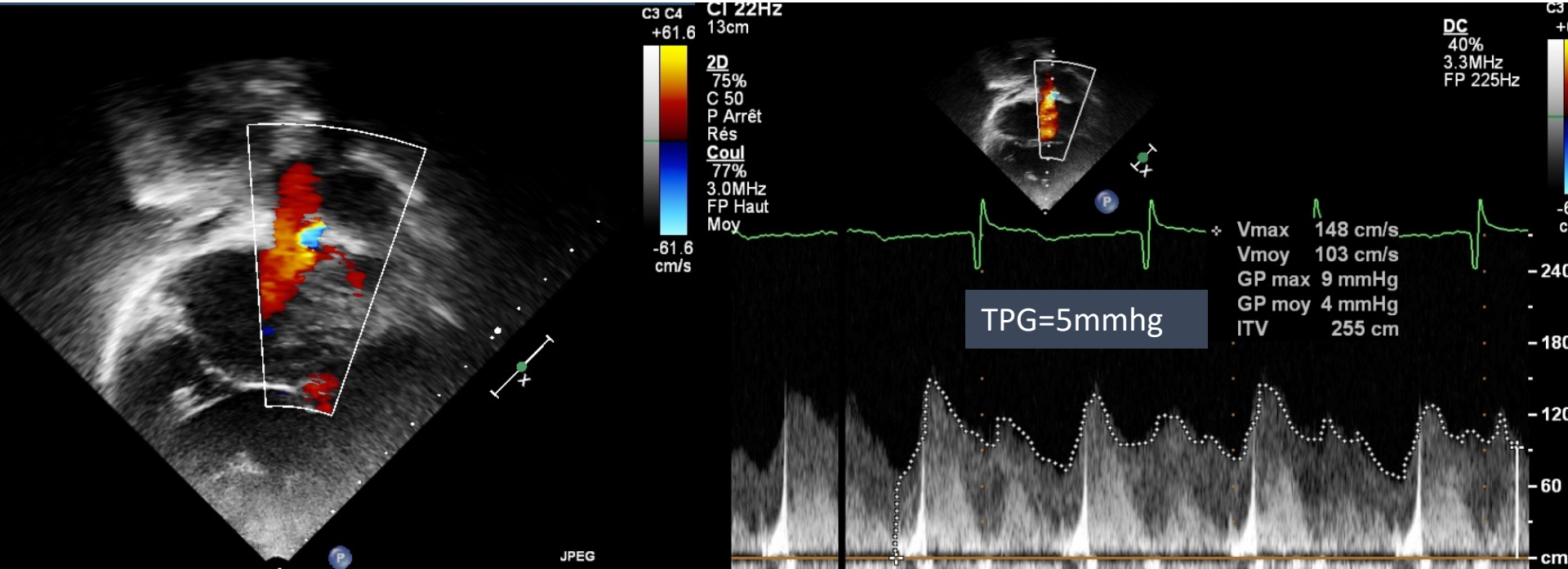
AO SaO2 84±6%		AO SaO2 95±3%
Qs:2.4±0.7 l1*min'*m-2	>	Qs: to1.8+0.4l1*min'*m-2
SOT*425±154ml		SOT: 366±112ml

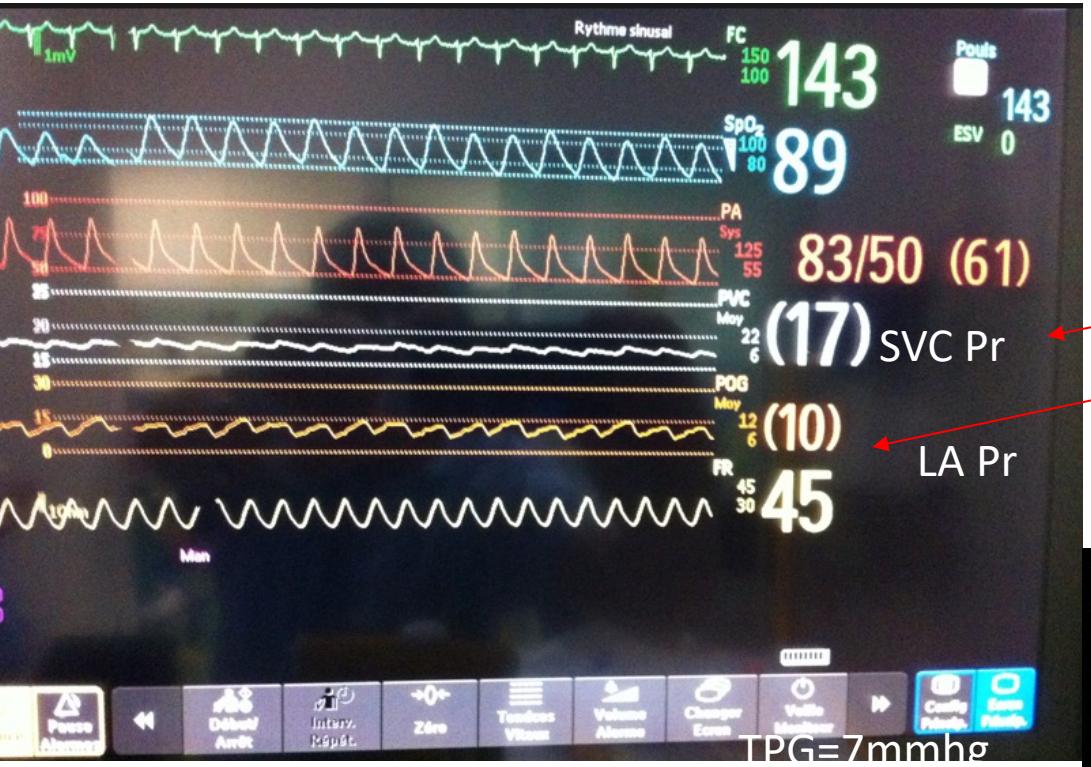
Hijazi et al. Circulation 1992

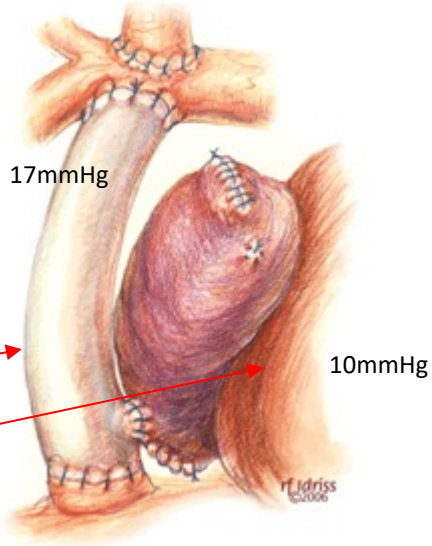
Fenestration assessement

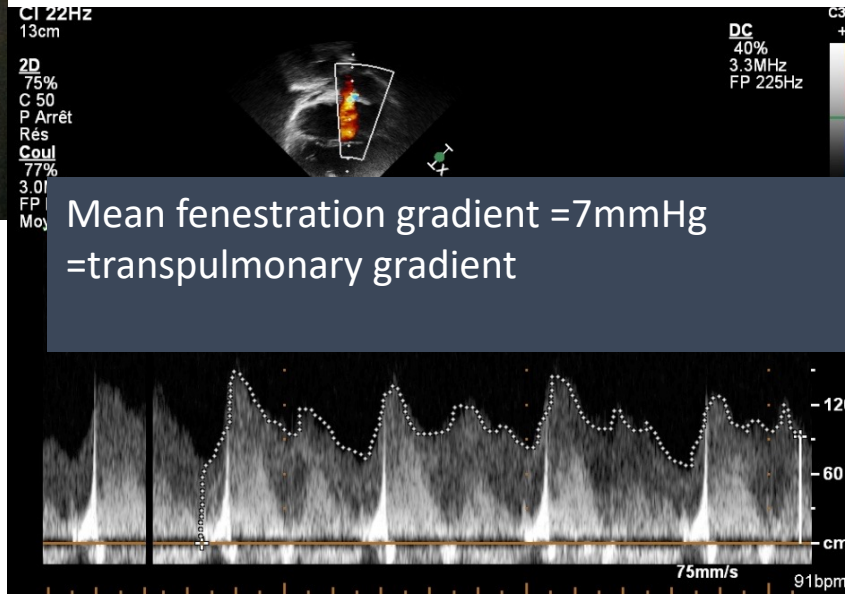


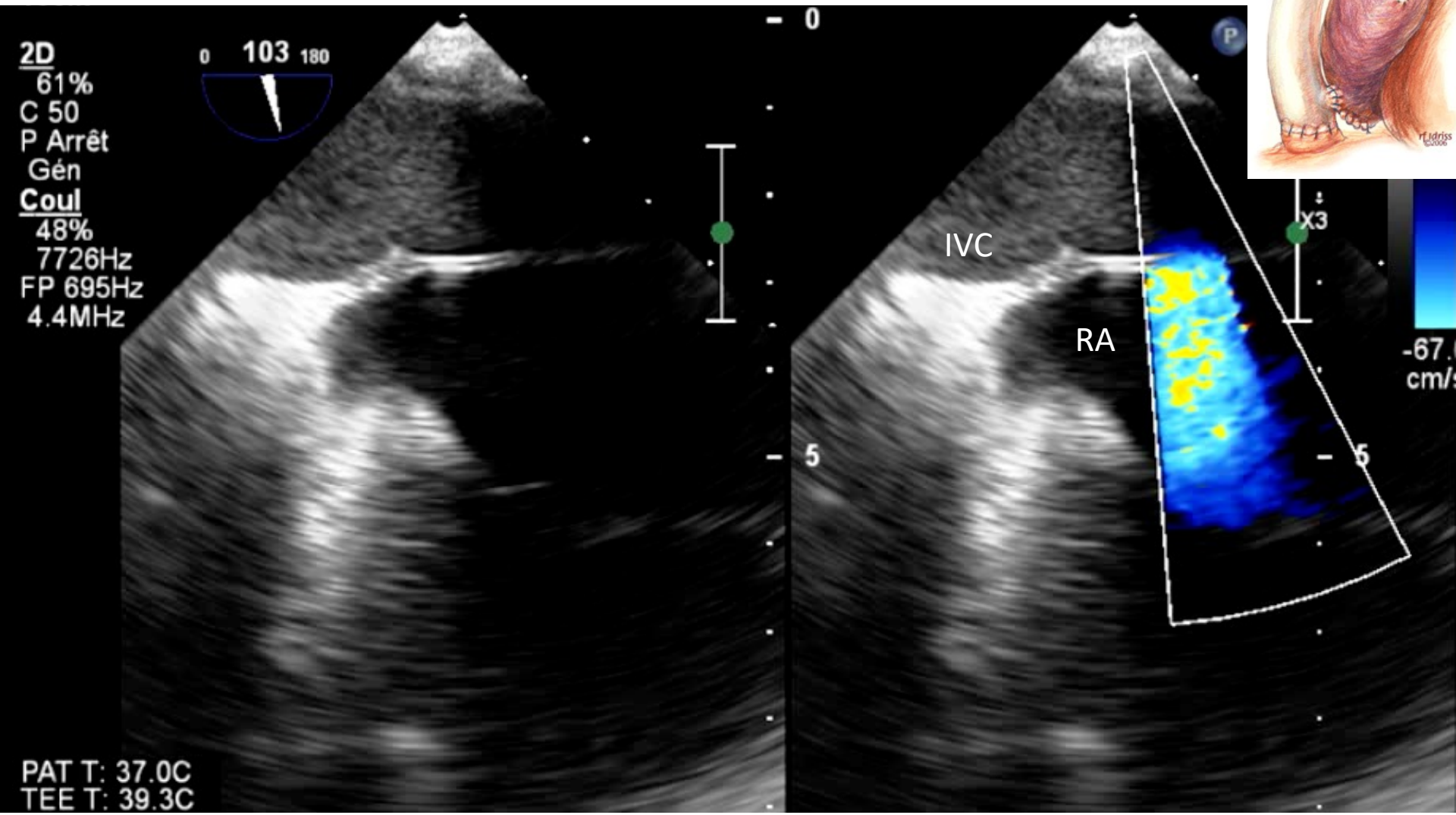
- Mean fenestration gradient = Transpulmonary gradient
  - Mean gradient over several cardiac cycles



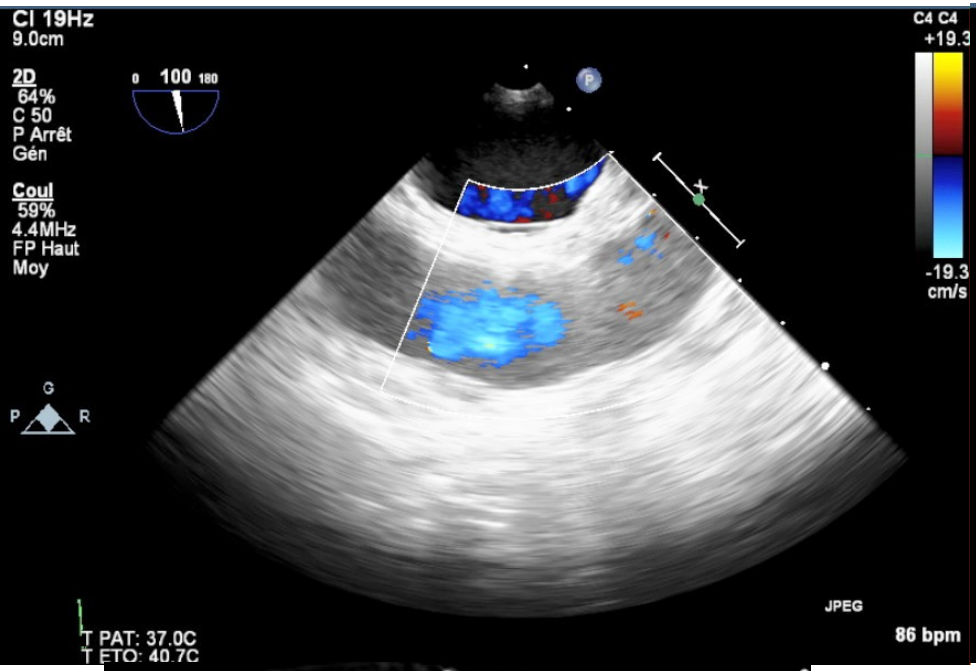

Rythme sinusal
FC
143
Pouls
143
ESV 0
89
PA
83/50 (61)
PVC
(17)
SVC Pr
POG
(10)
LA Pr
FR
45
TPG=7mmhg

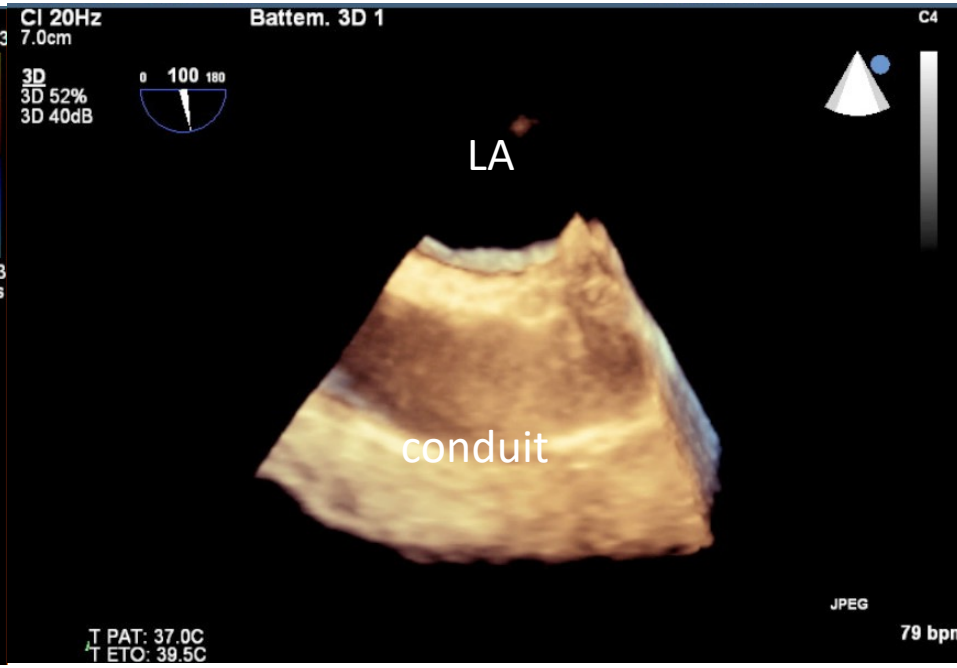

17mmHg
10mmHg

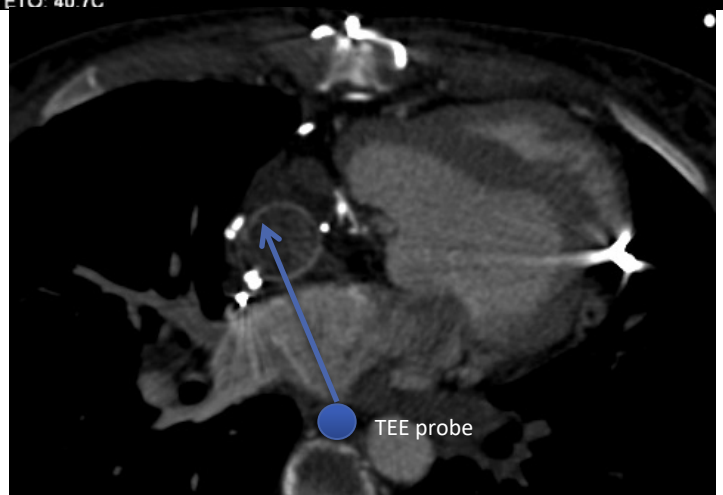

CF 22Hz
13cm
2D
75%
C 50
P Arrêt
Rés
Coul
77%
DC
40%
3.3MHz
FP 225Hz
Mean fenestration gradient =7mmHg
=transpulmonary gradient
75mm/s
91bpm


2D
61%
C 50
P Arrêt
Gén
Coul
48%
7726Hz
FP 695Hz
4.4MHz
0
103
180
0
5
IVC
RA
X3
-67.
cm/s
0
5
PAT T: 37.0C
TEE T: 39.3C

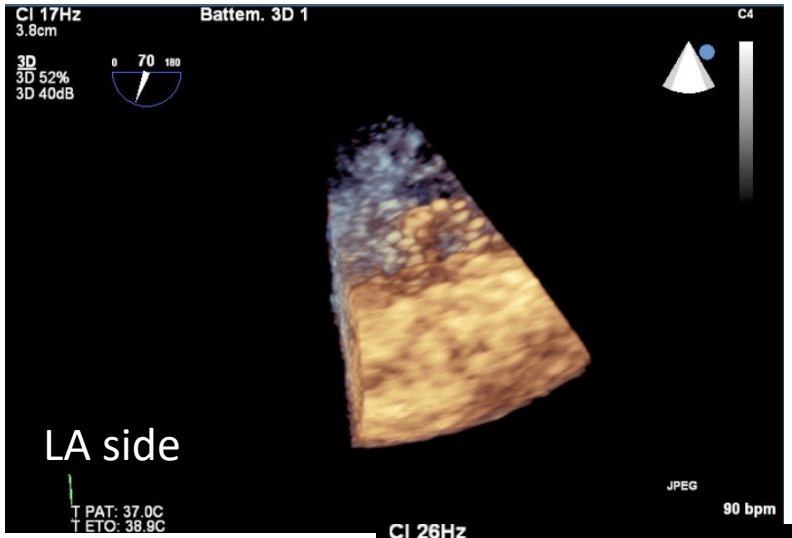
TOE guided-Percutaneous fenestration

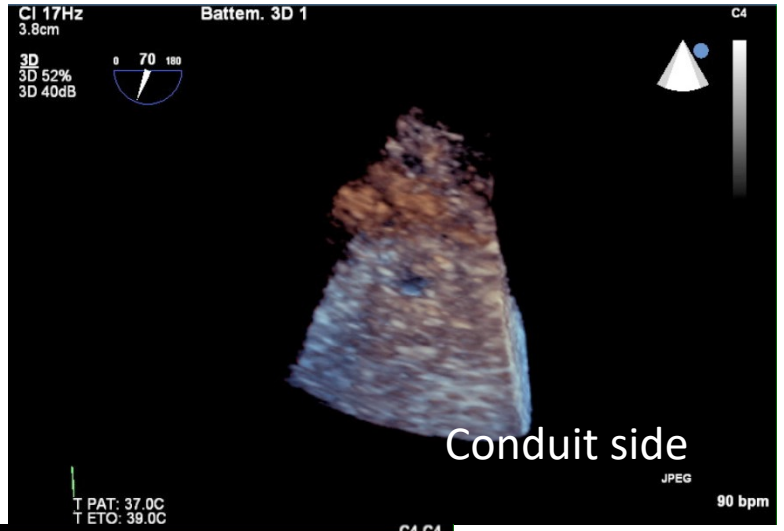


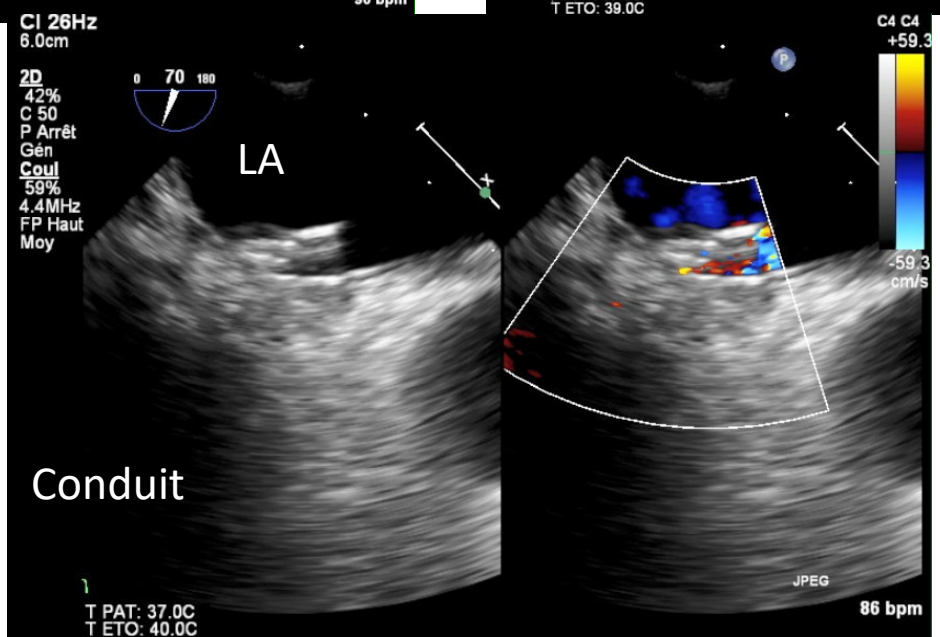




Percutaneous fenestration guiding







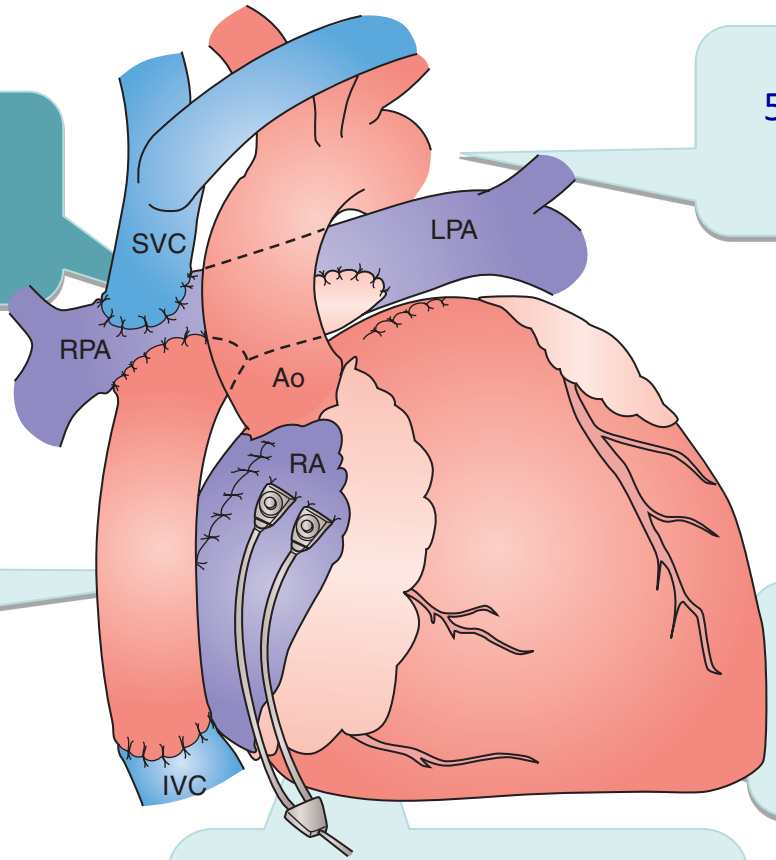
Echographic assessment after Glenn and TCPC completion: sequential segmental approach

1. Cavopulmonary connection
2. Branch PAs

PRELOAD

3. Pulmonary veins

4. Atrial septum



5. Systemic outflow tract
8.Aortic arch

AFTERLOAD

6.Systemic ventricular function

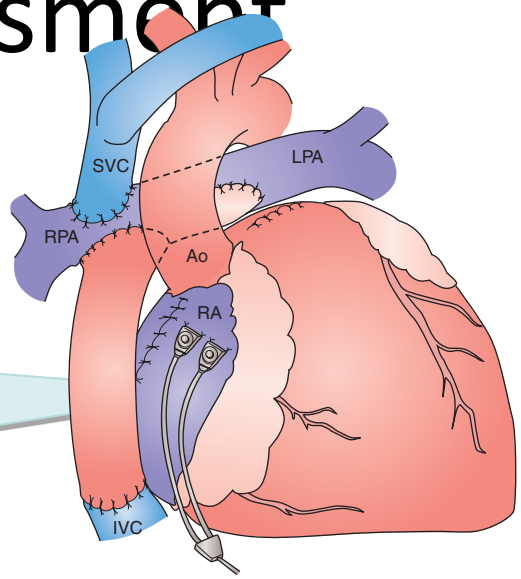
FUNCTION

7. Atrioventricular valves

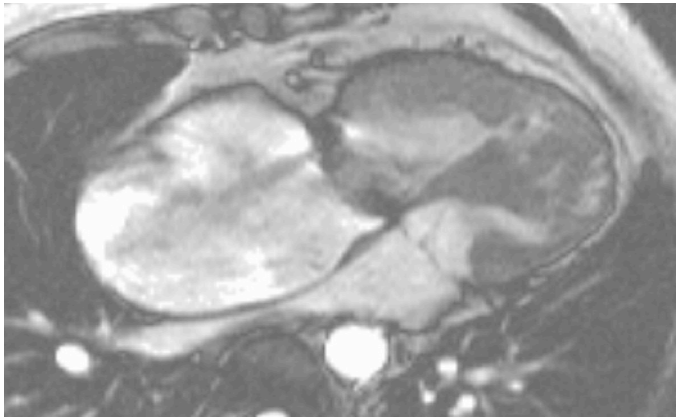
Echographic assessment

3. Pulmonary veins

4. Atrial septum



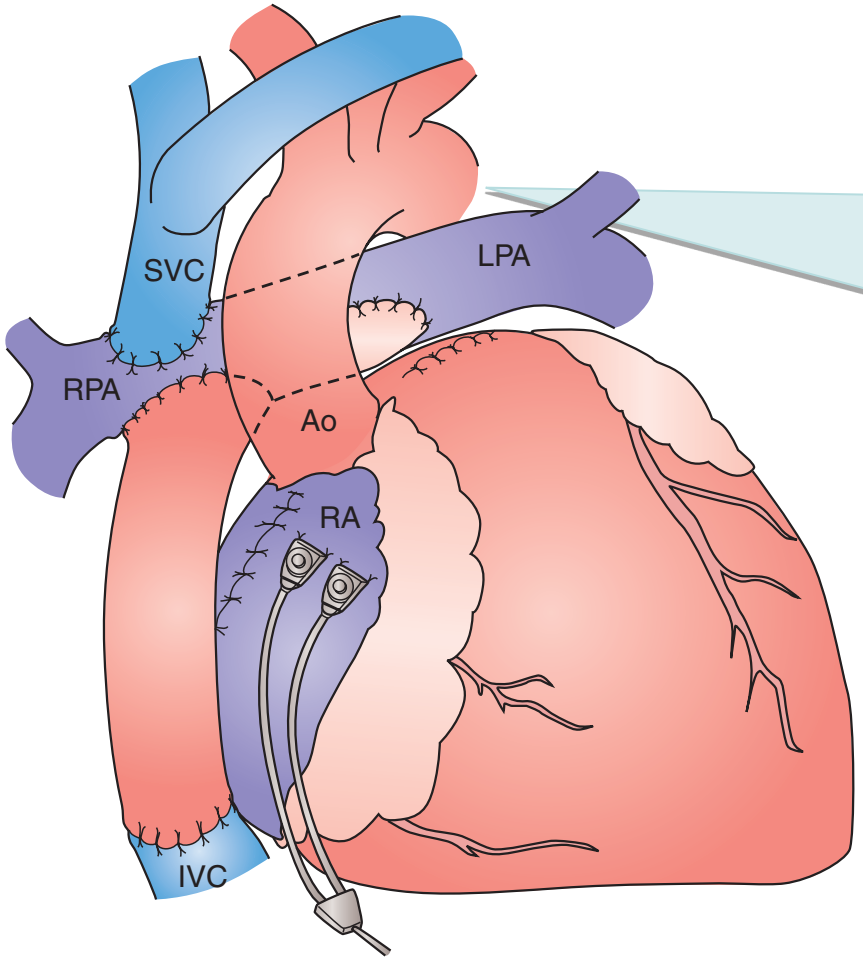
- PV compression
  - Atriopulmonary connection
  - Intracardiac tunnel
  - Heterotaxy syndrome



- Restriction of interatrial shunt in HLHS

- Potential cause of elevated PA pressure

Echographic assessment



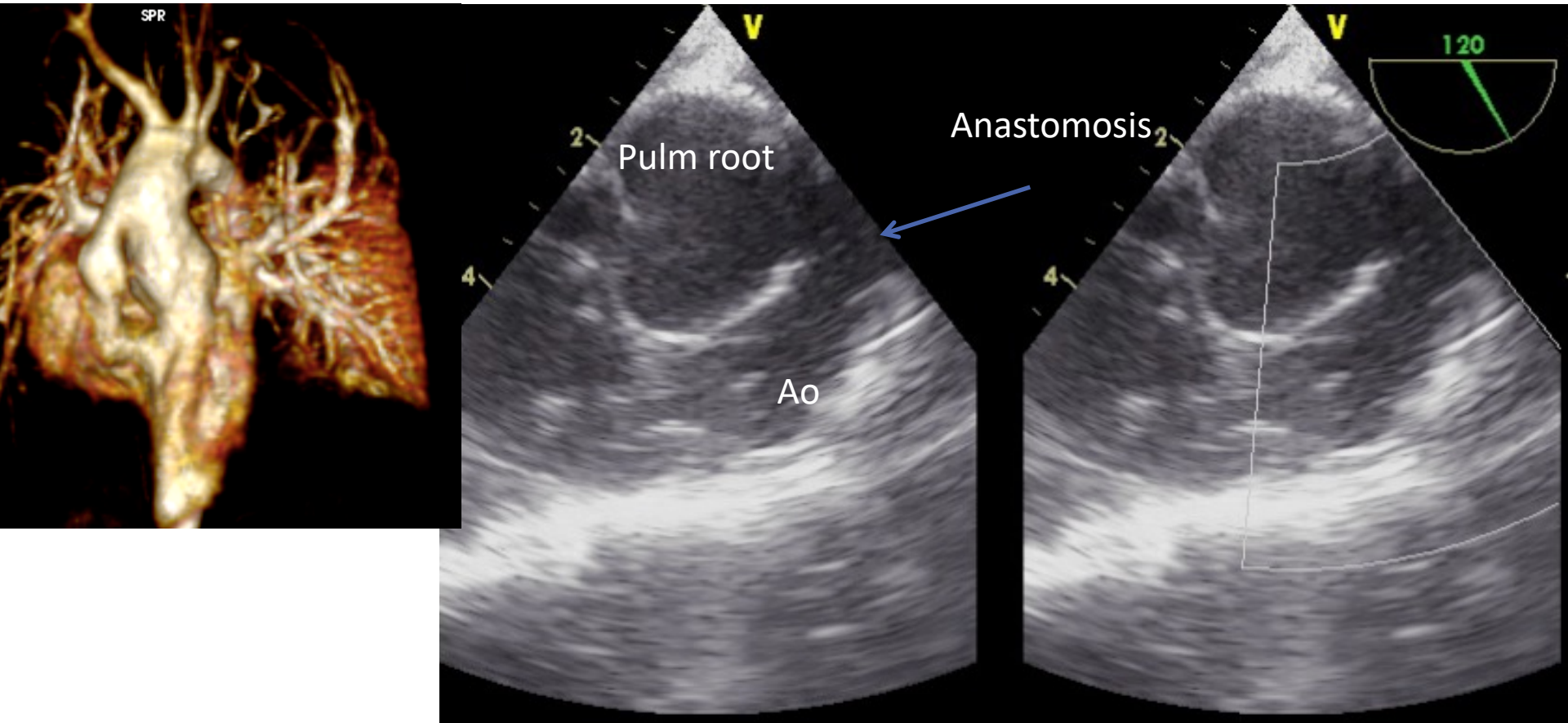
SV AFTERLOAD

6. Systemic outflow tract

7.Aorta

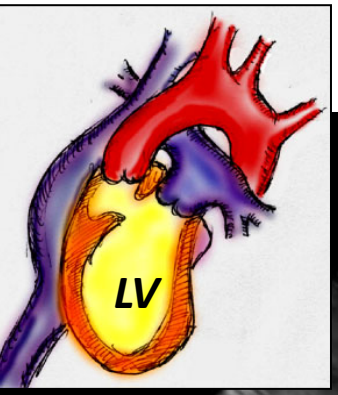
Assessment of DKS anastomosis

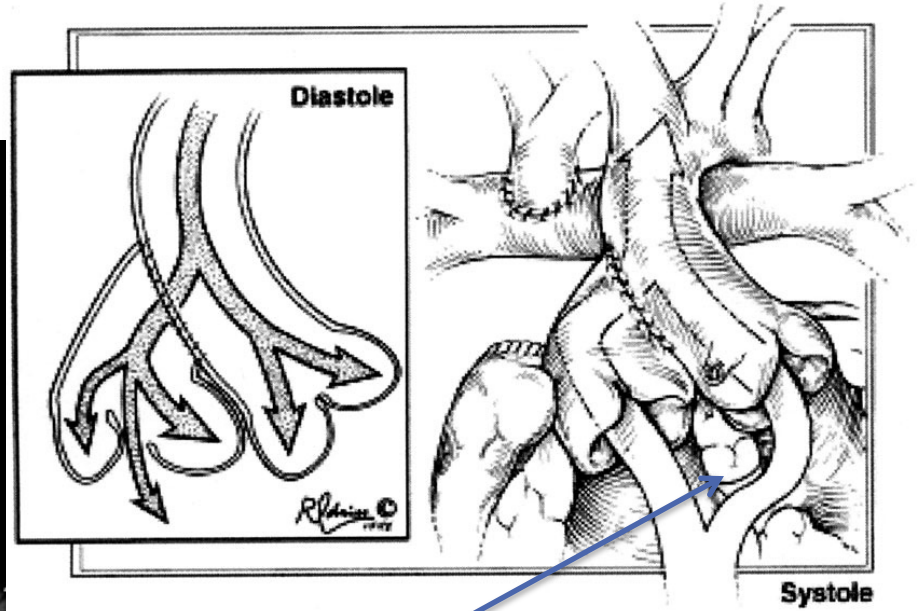
Exemple of a DKS anastomis in DILV with restrictive bulboventricumar foramen



Restrictive systemic outflow tract in DILV

- Proximal anastomosis
  - Restritive bulboventricular foramen: DKS



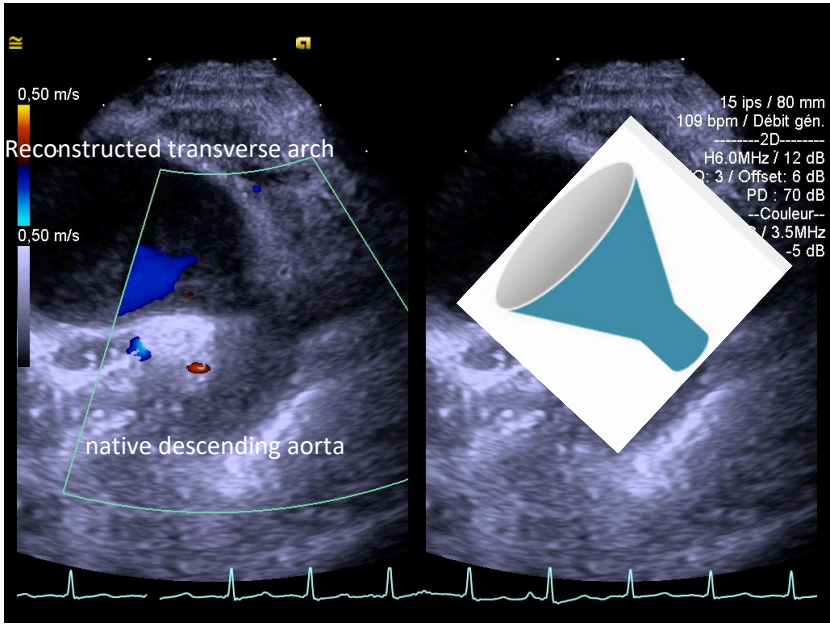


Restritive bulboventricular

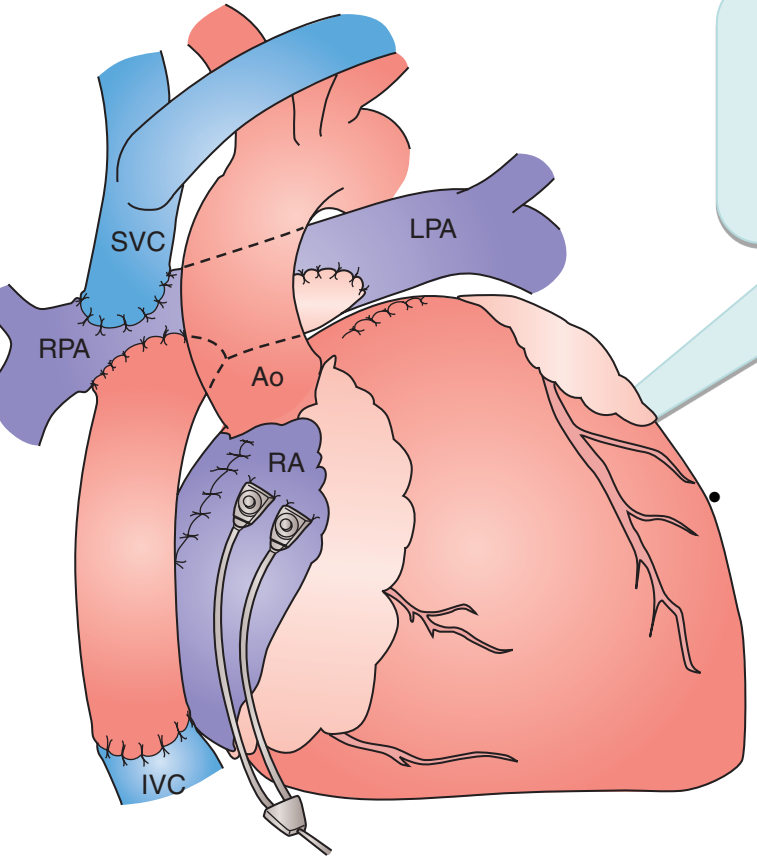
Aortic arch assessment

- Distal anastomosis
  - Supra sternal sagital view
  - Potential increased velocity (HLHS)

  - Pitfall: potential absence of diastolic runoff
    - change in Ao arch geometry/ patch/mBTshunt

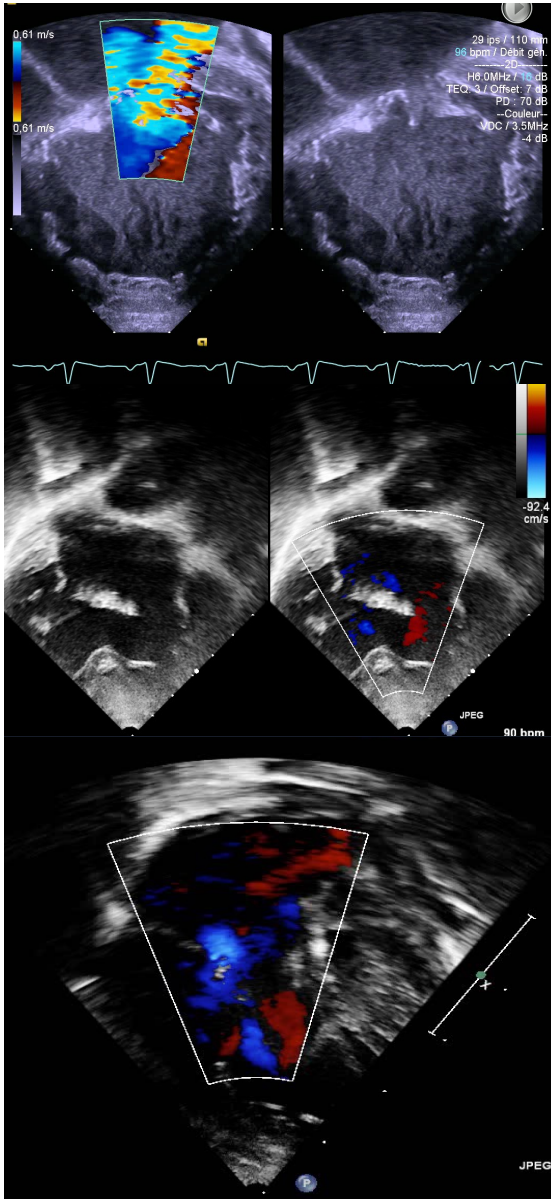


AVV assessment

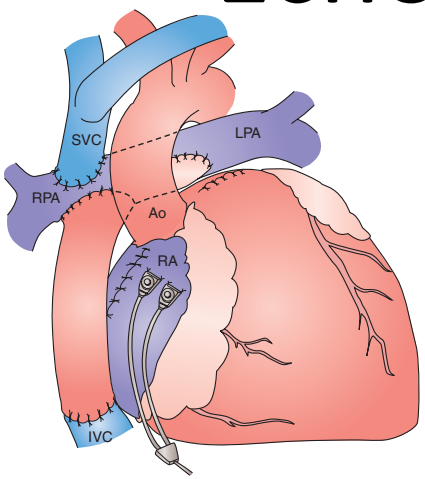


5. Atrio-ventricular valves

- Different anatomies
- Different loading conditions



Echographic assessment



8.Systemic ventricular function

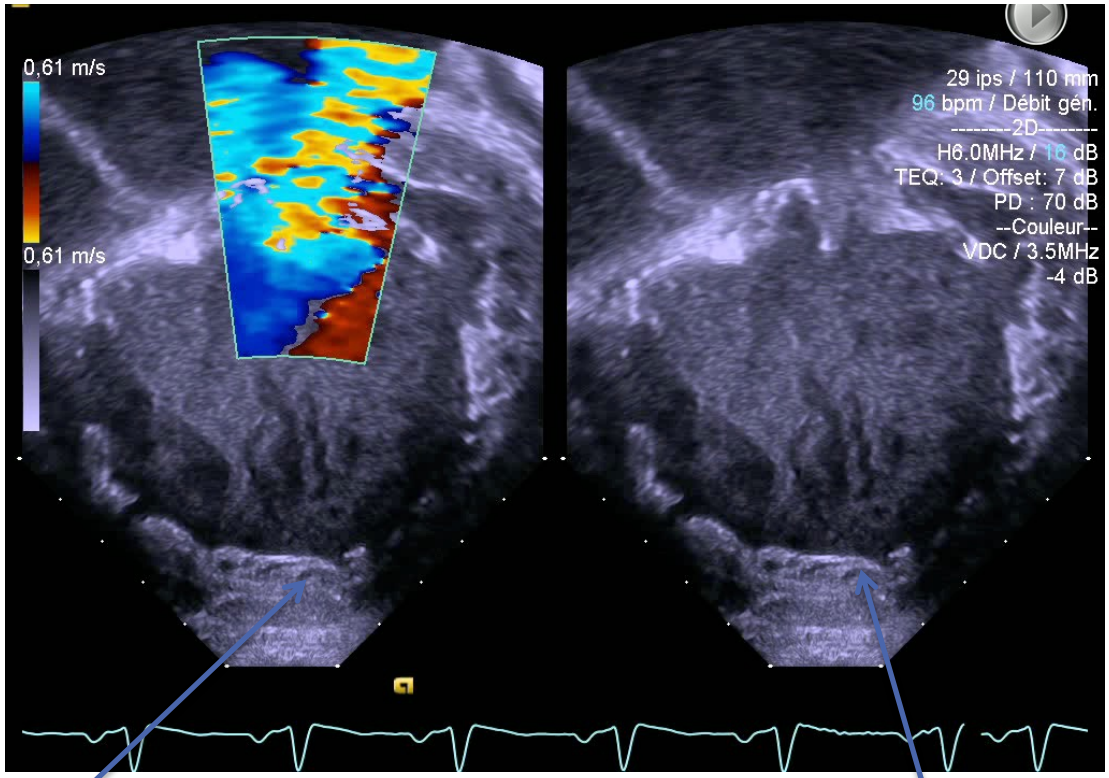
- Systolic and diastolic(+++) function
  - No single morphology
  - Important confunding variables
    - Preload (surgical stage/ AVVR/ PVR/PA compliance)
    - Afterload (restrictive outflow trat, CoA)

- Eye balling/ EF (TM, 2D) / myocardial deformation

Diagnosis and managment of post operative complication

- Pleural/abdominal effusion: multifactorial+++
- SV dysfunction
- Thrombosis
- Abnormal cyanosis

SV dysfunction

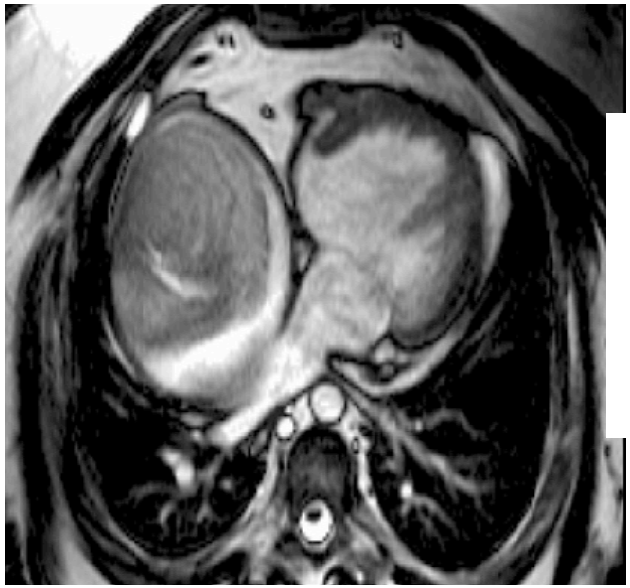


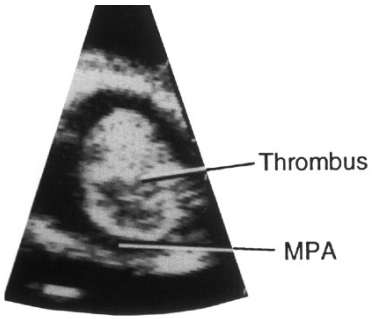
Restrictive bulboventricular foramen
Increased AFTERLOAD

PA banding/MR
Increased PRELOAD

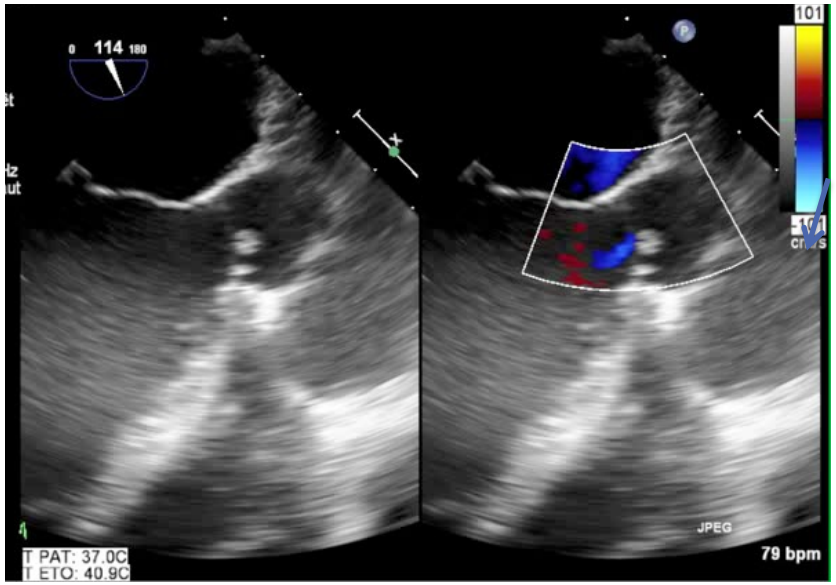
LV spherical remodeling : secondary MR worsening

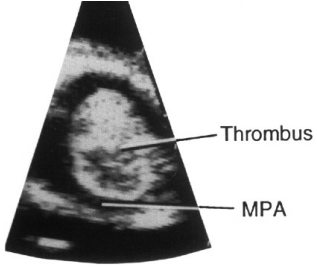
Thrombosis in TCPC

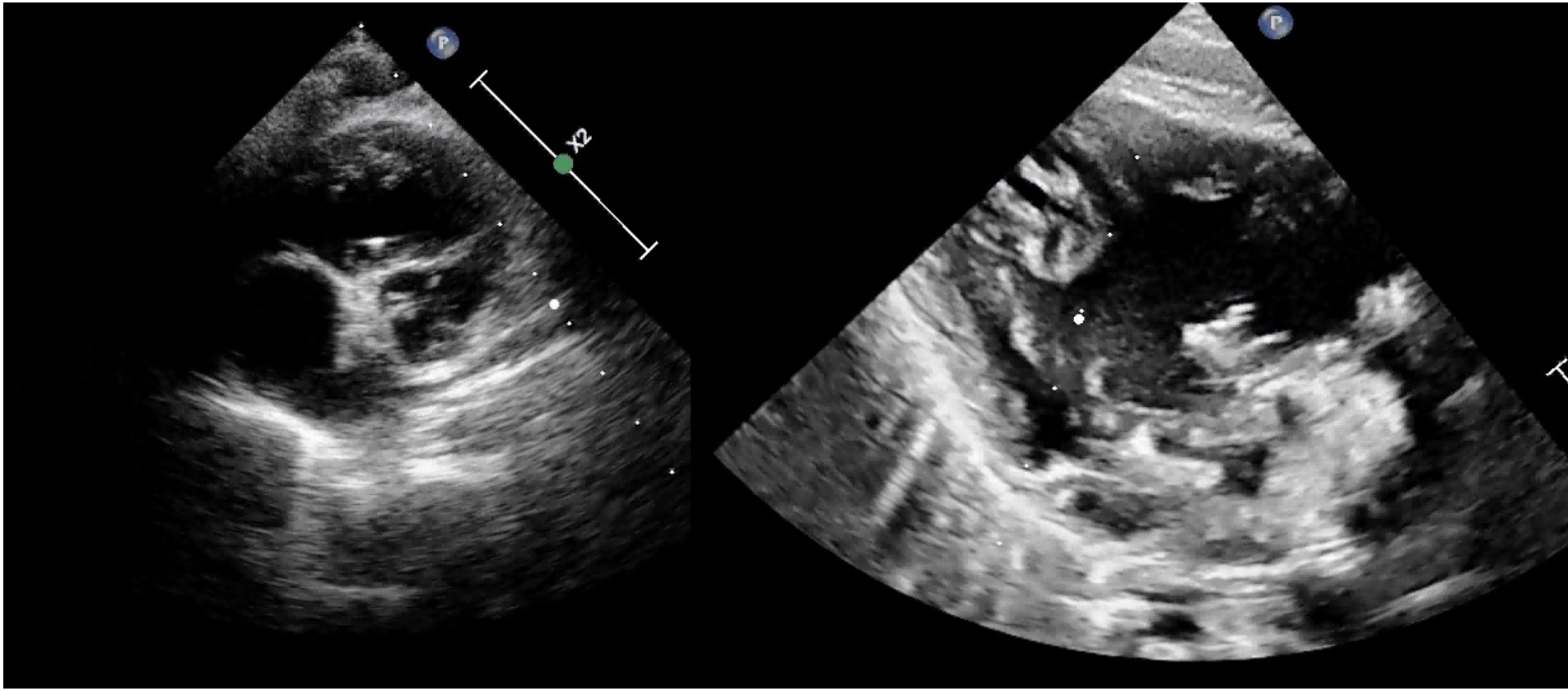




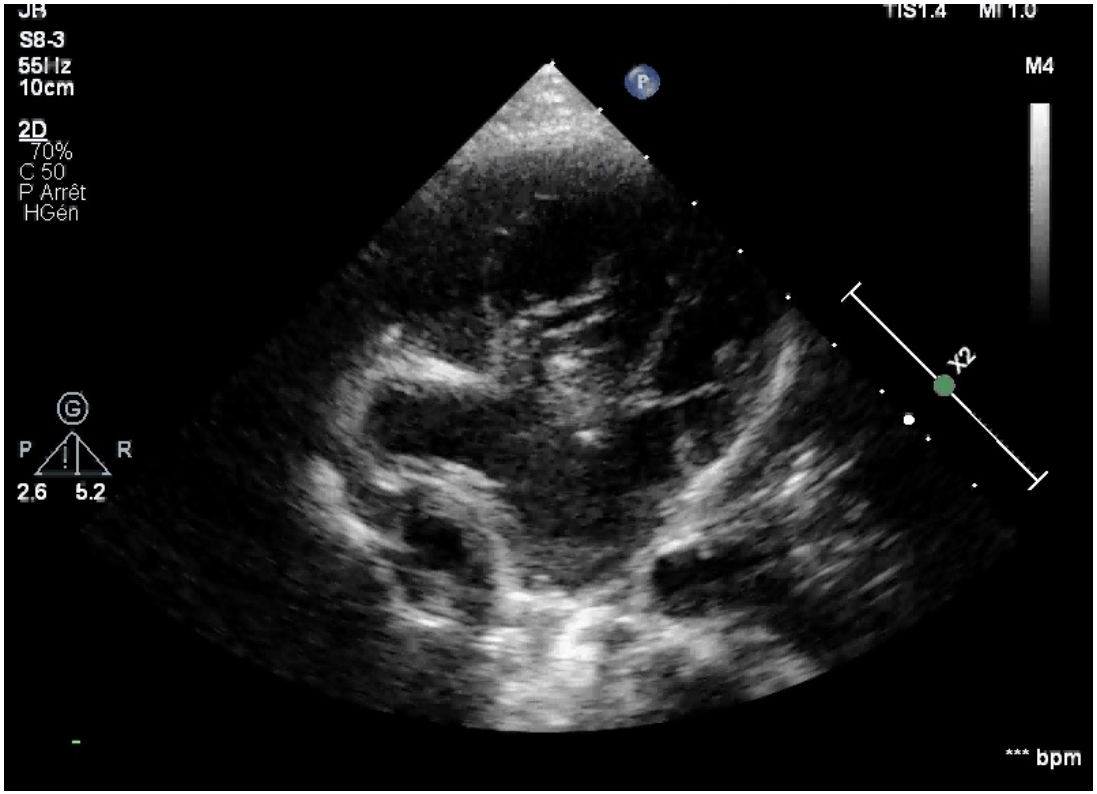
- Systemic venous pathway (TCPC)
  - IVC, RA (atriopulmonary connection), lateral tunnel/ conduit, fenestration,PAs, SVC
- Intra cardiac chambers
  - Intracardiac: LA, LAA, SV (poor systolic function)
- Native PA trunk
  - PAs divided and pulm valve non sutured

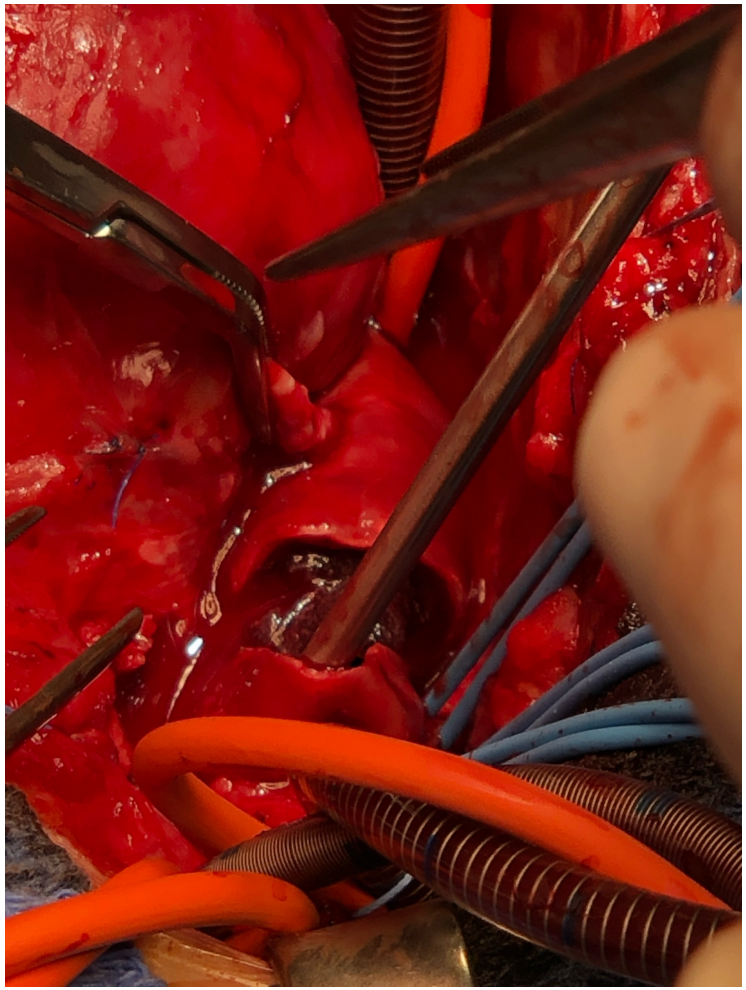



Thrombus
MPA


X2

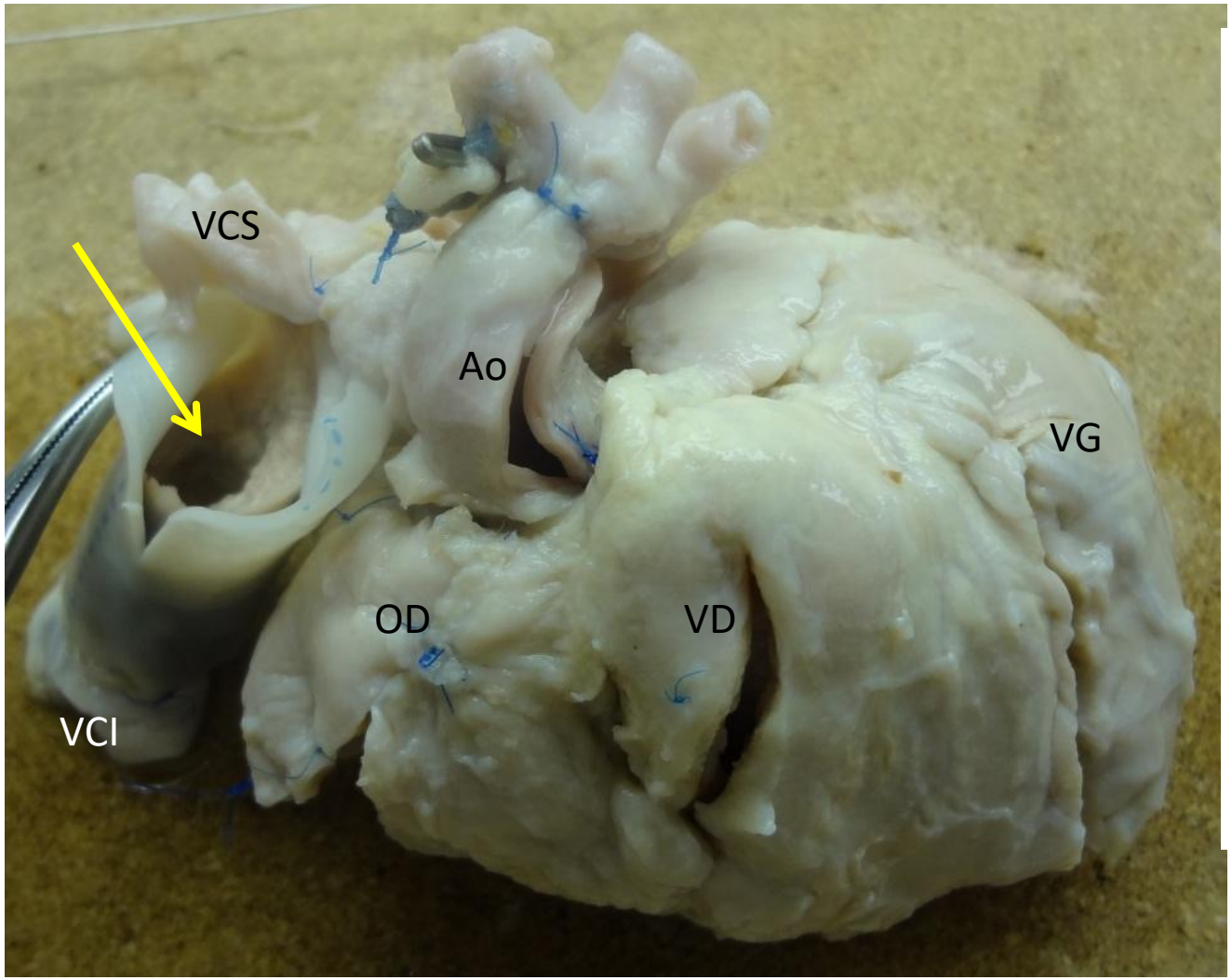
Thrombotic complications

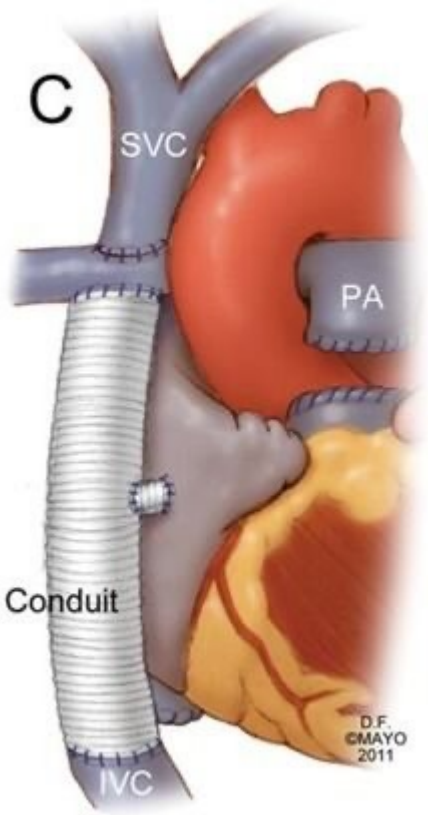




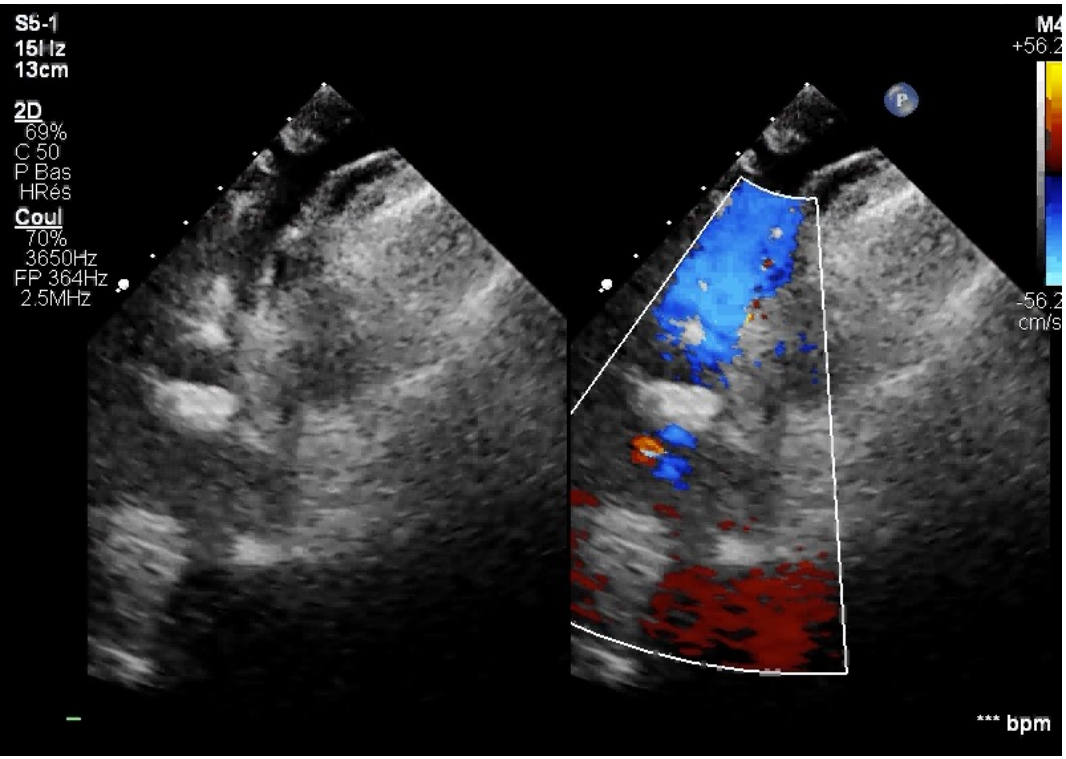
DCPT : thrombose du tube extra-cardiaque

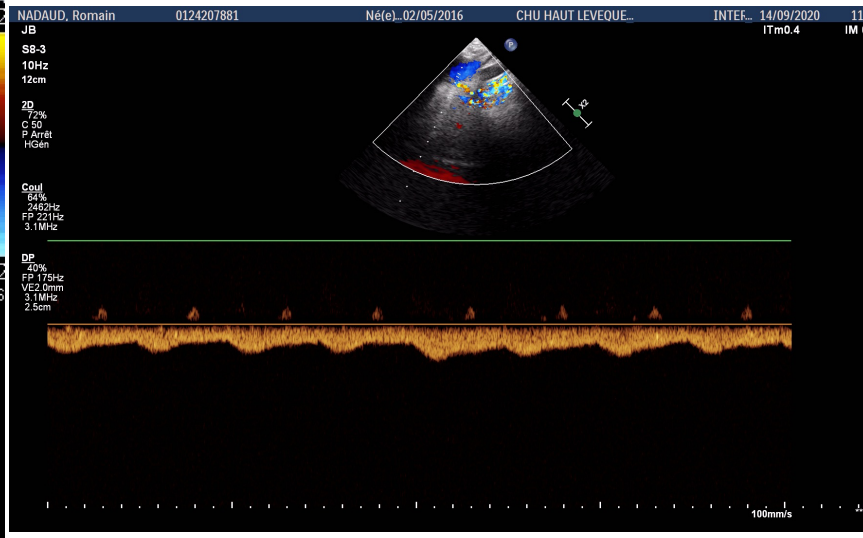
Atrésie tricuspide {S,D,S}

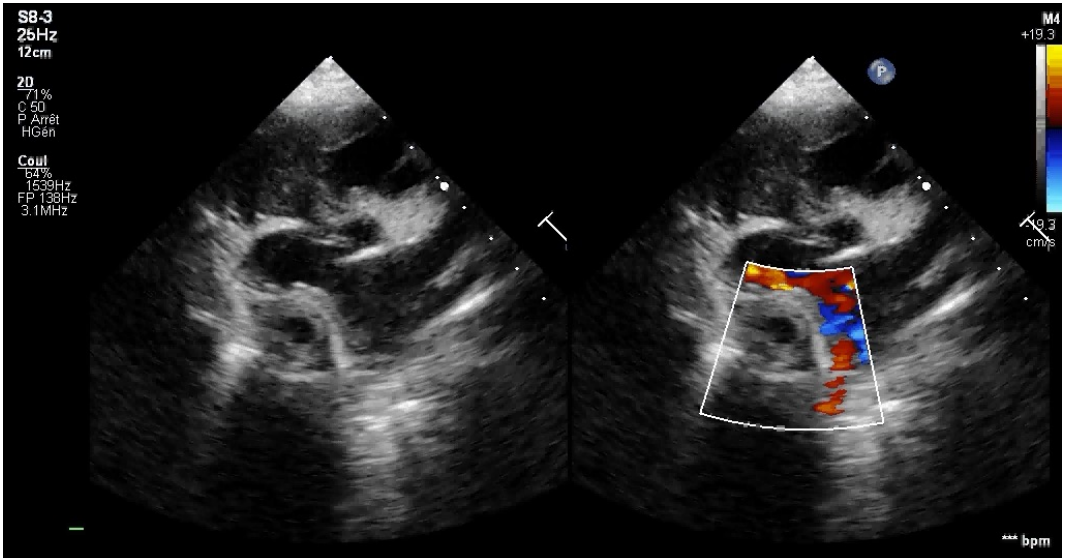


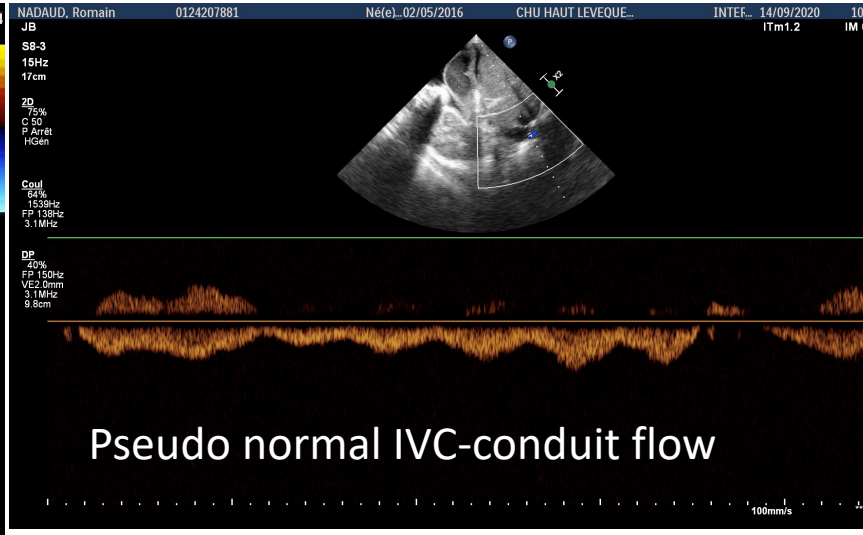


M3C


S5-1
15Hz
13cm
2D
69%
C 50
P Bas
HRés
Coul
70%
3650Hz
FP 364Hz
2.5MHz
M4
+56.2
-56.2
cm/s
*** bpm

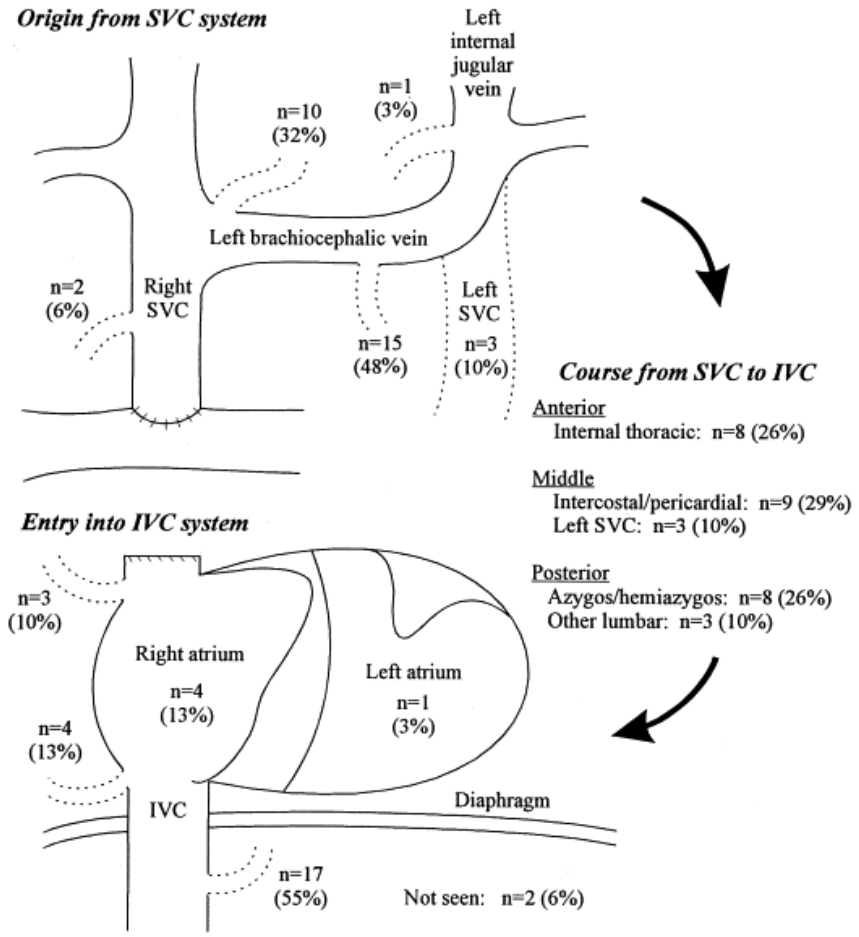

NADAUD, Romain
0124207881
Né(e)...02/05/2016
CHU HAUT LEVEQUE...
INTEF... 14/09/2020
JB
S8-3
10Hz
12cm
2D
72%
C 50
P Arrêt
HGén
Coul
64%
2462Hz
FP 221Hz
3.1MHz
DP
40%
FP 175Hz
VE2.0mm
3.1MHz
2.5cm
100mm/s


S8-3
25Hz
12cm
2D
71%
C 50
P Arrêt
HGén
Coul
64%
1539Hz
FP 138Hz
3.1MHz
M4
+19.3
-19.3
cm/s
*** bpm


NADAUD, Romain
0124207881
Né(e)...02/05/2016
CHU HAUT LEVEQUE...
INTEF... 14/09/2020
JB
S8-3
15Hz
17cm
2D
75%
C 50
P Arrêt
HGén
Coul
64%
1539Hz
FP 138Hz
3.1MHz
DP
40%
FP 150Hz
VE2.0mm
3.1MHz
9.8cm
Pseudo normal IVC-conduit flow
100mm/s

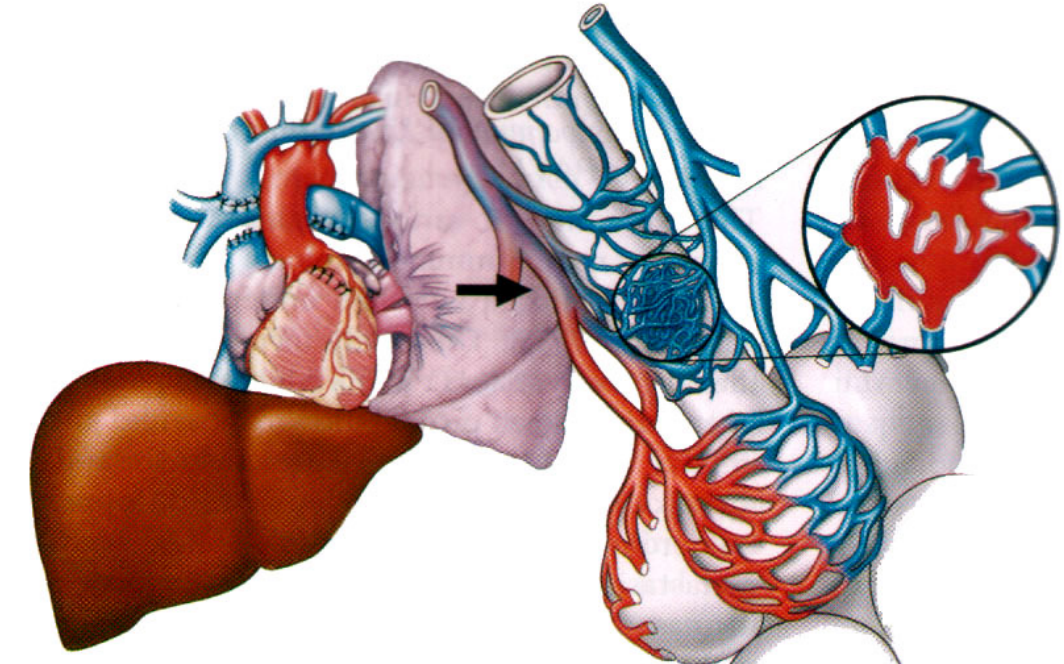
Abnormal cyanosis after BCPC

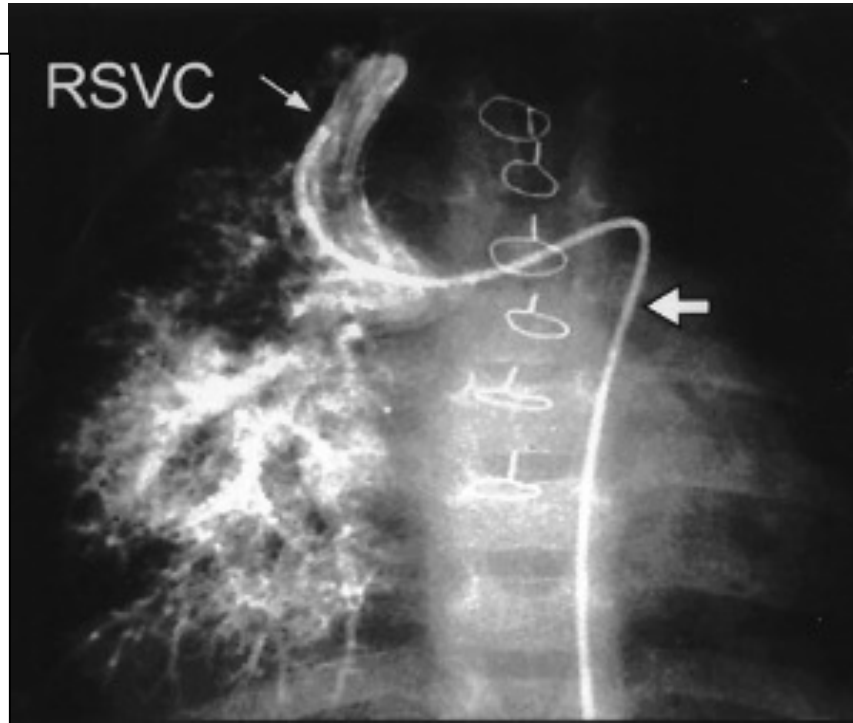
- Reopening of decompressing veins from SVC
  - In the IVC: azygos vein
  - In the atria: left SVC
  - Diagnosis:
    - Suprasternal frontal view
    - color doppler/ saline contrast

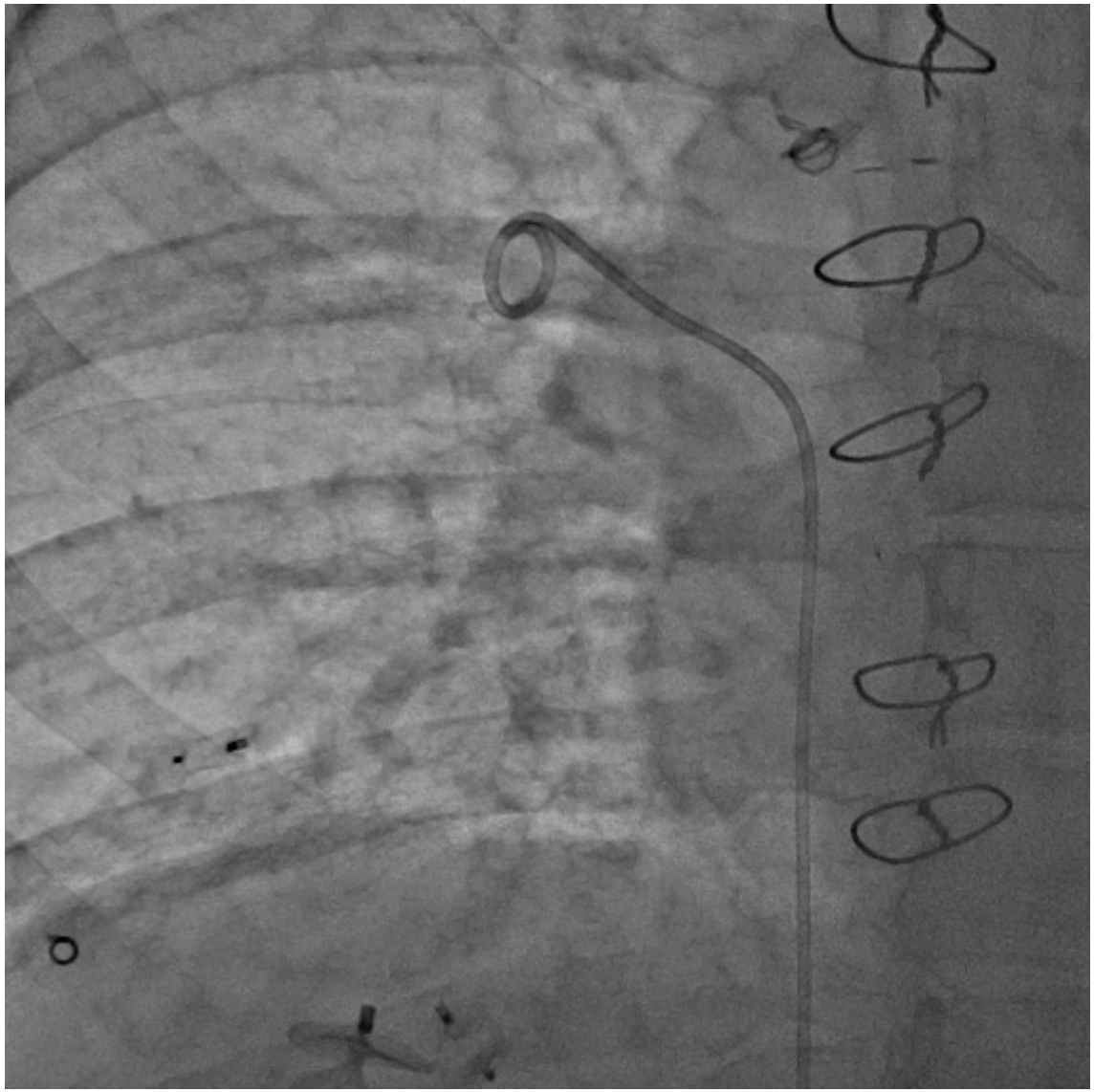


Abnormal cyanosis after BCPC

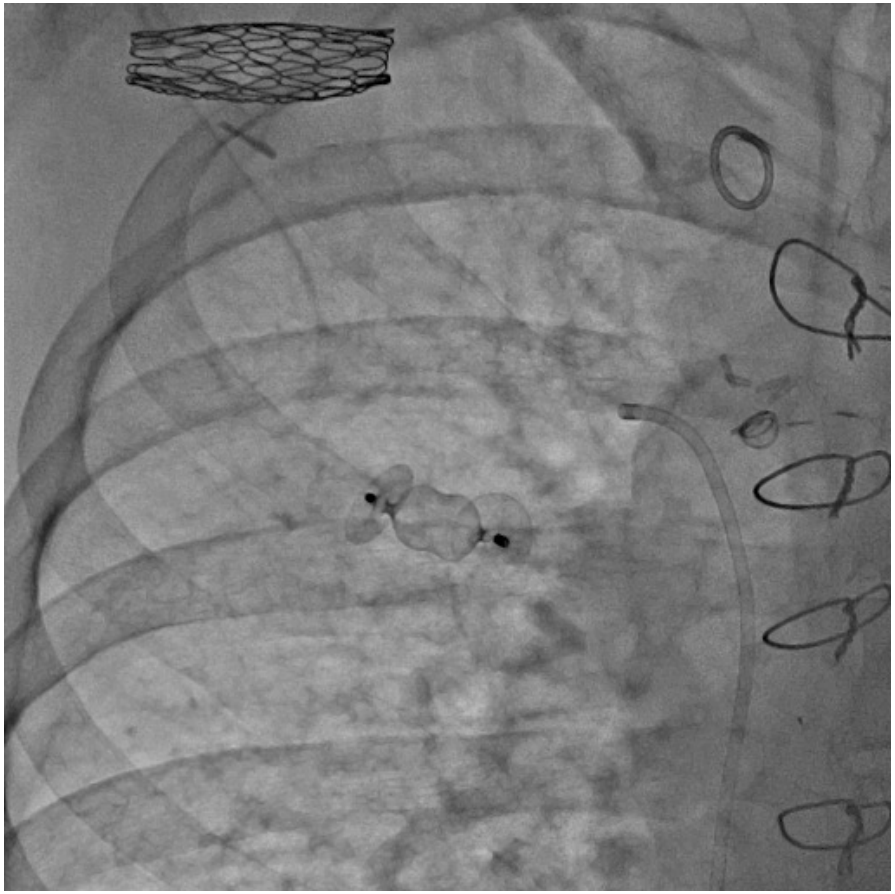
- Pulmonary AV malformations
  - heterotaxy syndrome
  - Lack of hepatic factor
  - Diagnosis: saline contrast

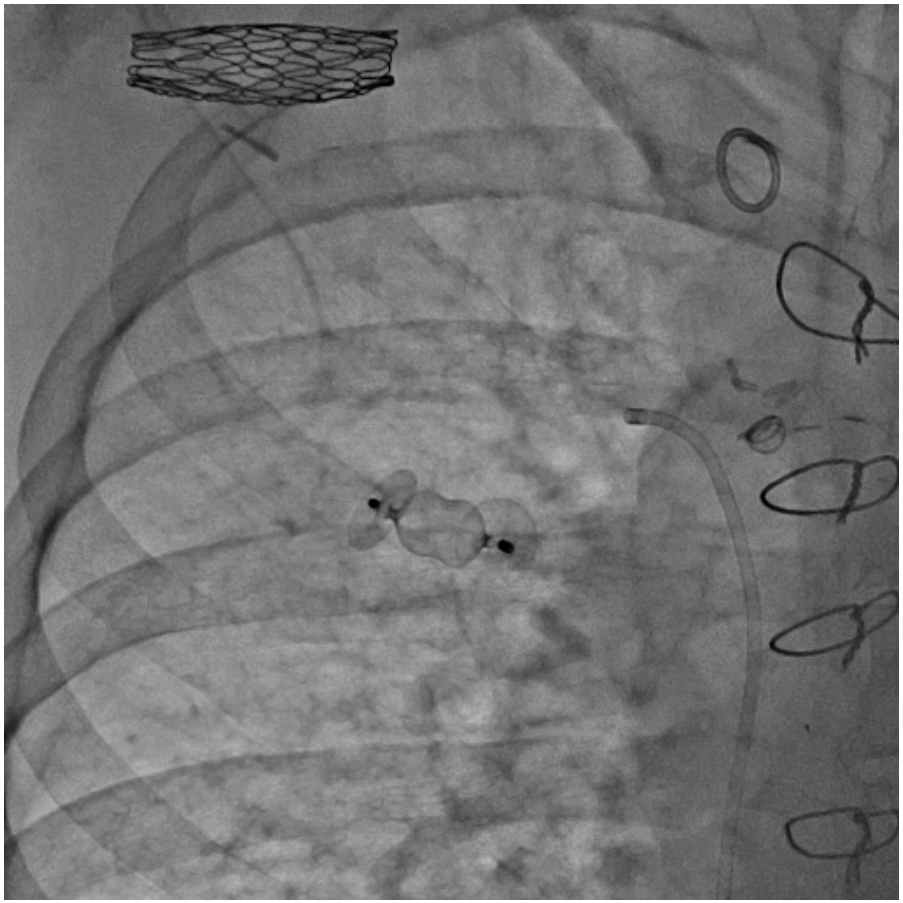







A
A: 7.8mm
CF: 0.1231mm/p (x1.4)



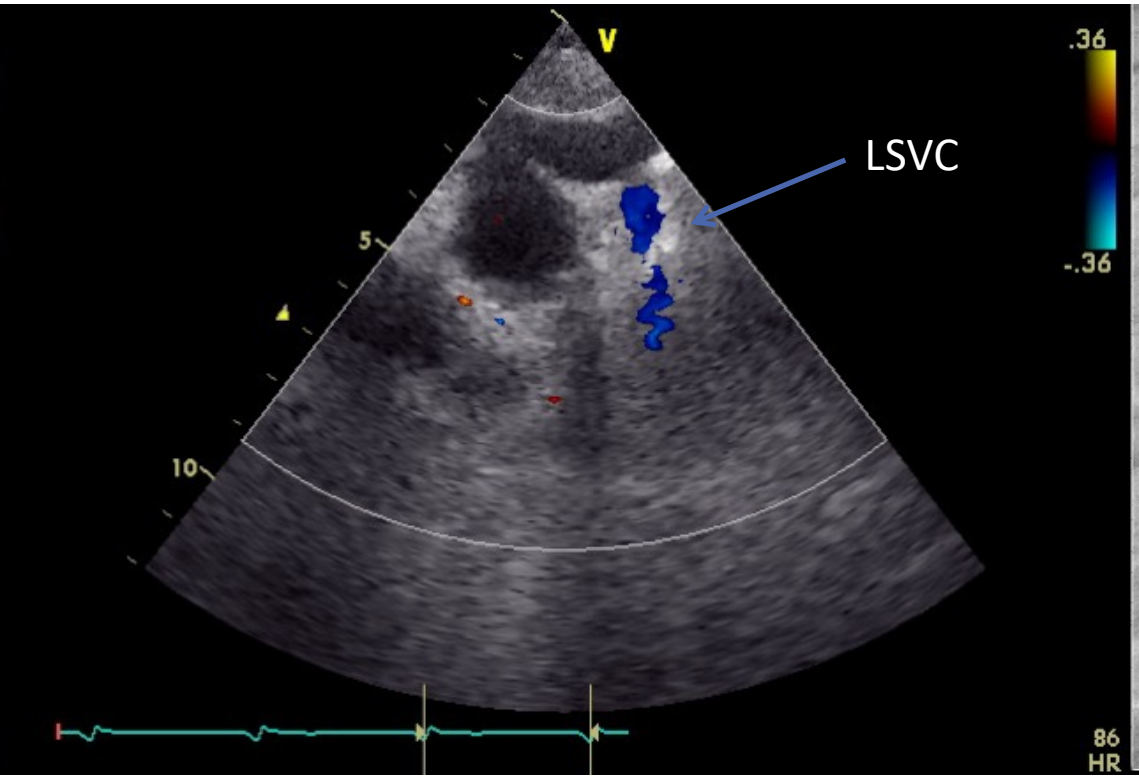


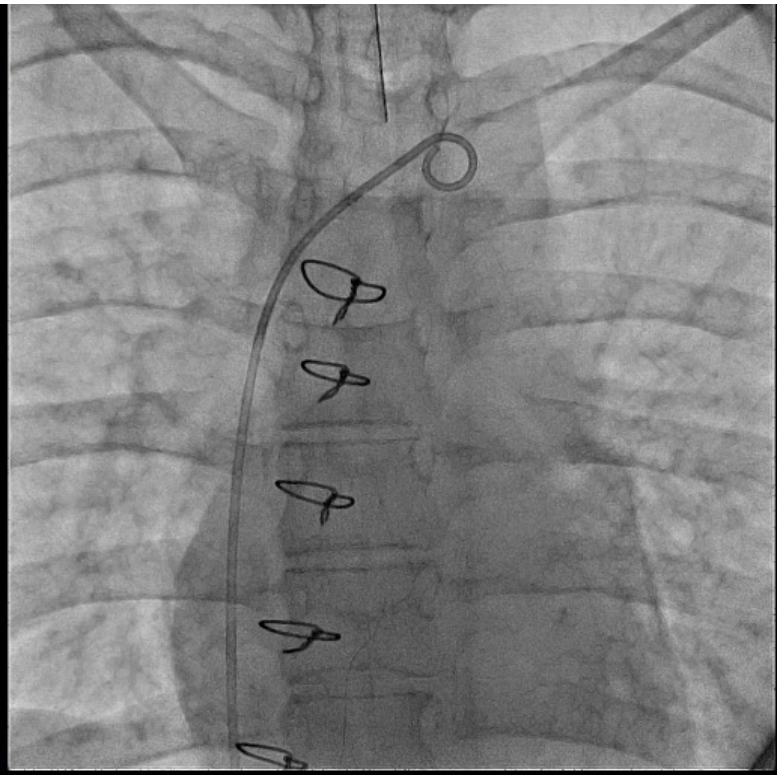
Persistent cyanosis after TCPC

- Conduit fenestration
  - Balance between PVR and early diastolic function
- Systemic venous pressure> pulmonary vein pressure
  - VV collaterals to pulmonary veins or systemic atrium
    - origin: LSVC, RSVC, inn Vein, hepatic veins
- Baffle leaks (intra cardiac type of connection)

Superior veno venous collateral

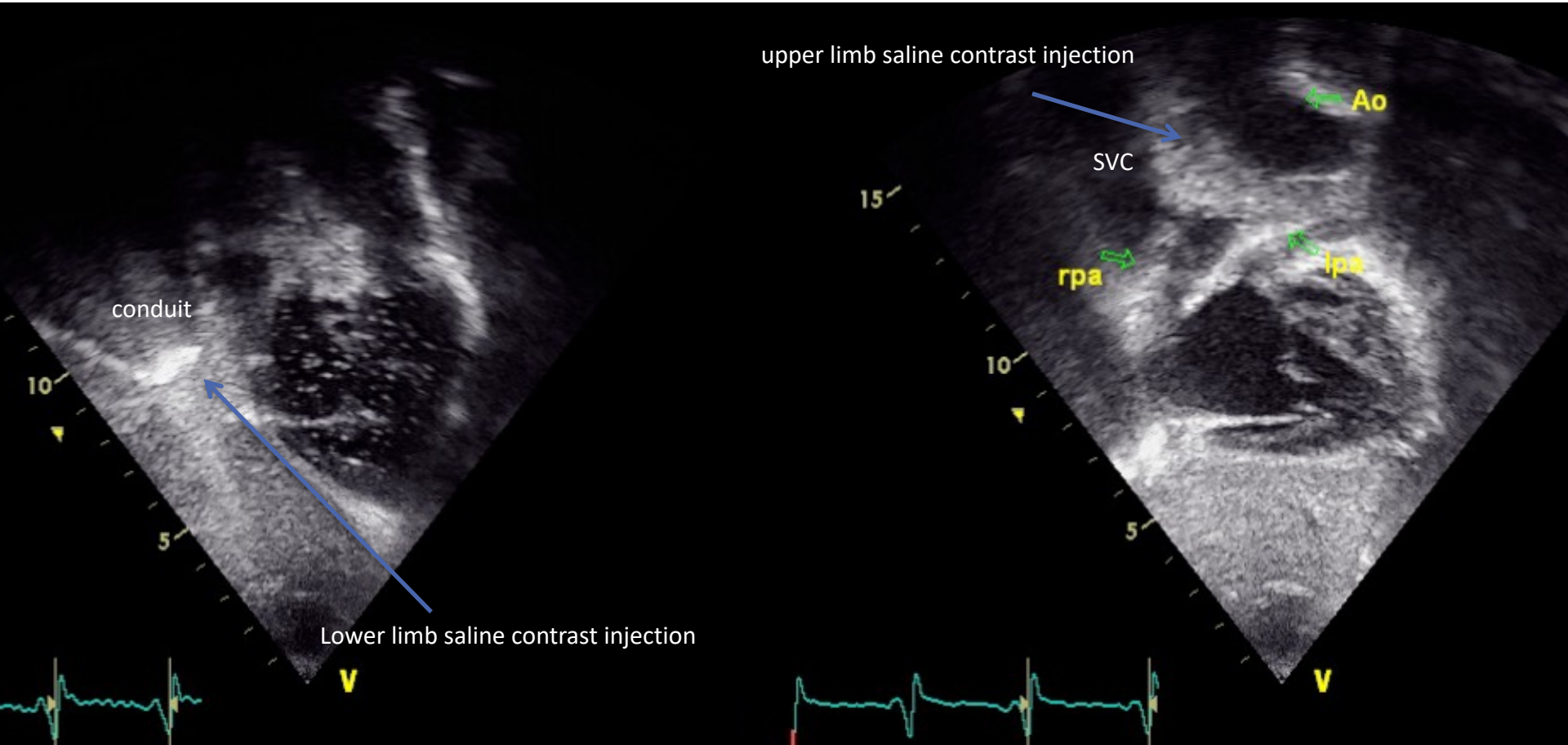
- Left « SVC » to the RA

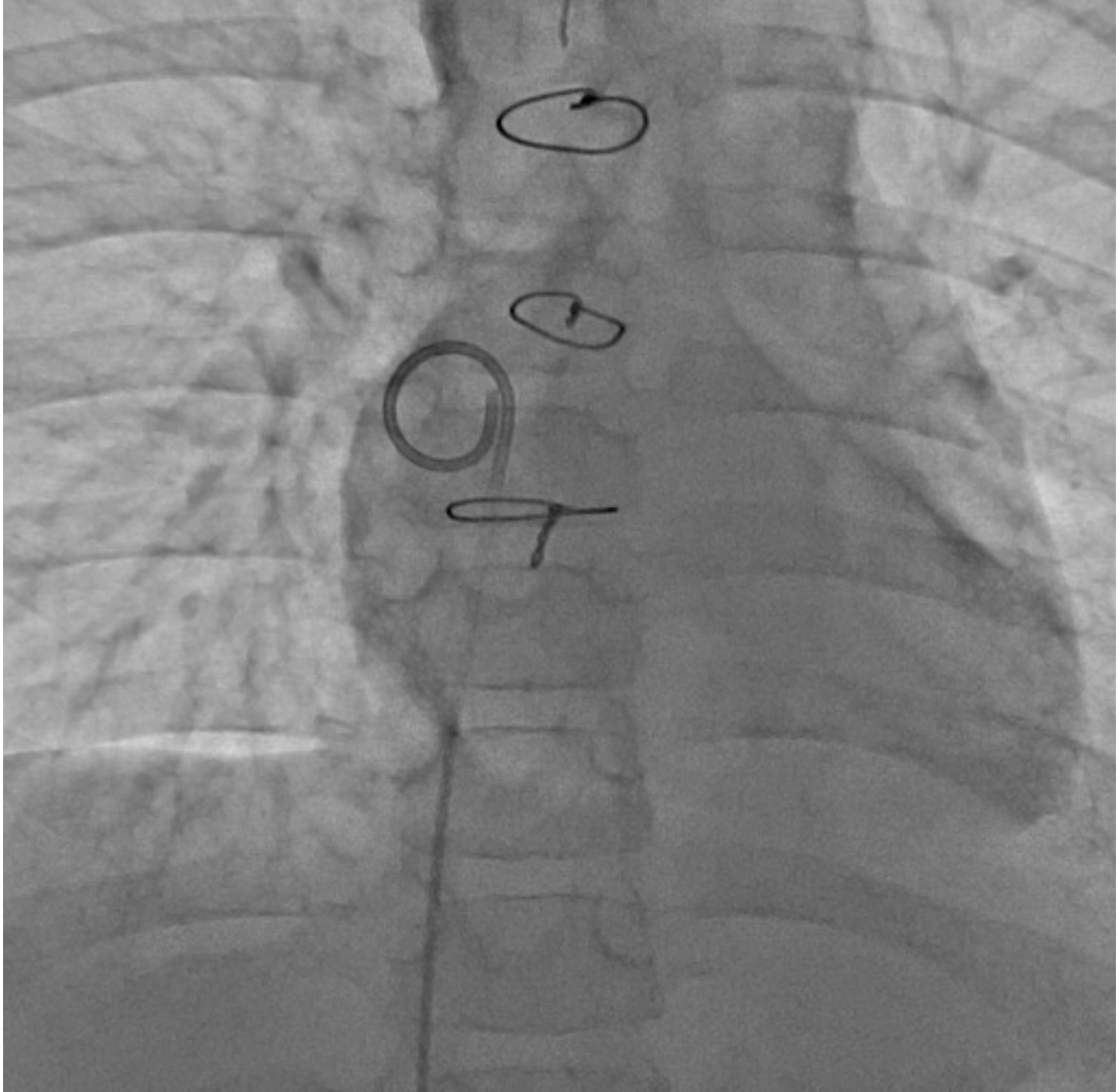




Inferior veno venous collateral

- HLHS S/P TCPC. Persistant mild hypoxemia
  - Shunt between HV and cardiac veins





Conclusion

- Understand Fontan physiology is key.
- Echo:Easily accessible and cost-effective tool: Sequential segmental analysis to not miss a treatable lesion
- Good assessment of the Fontan pathway in children but may be limited in adults. CT and MRI +++
- Global clinical assessment is mandatory: extracardiac complications drive the prognosis
- UVH systolic and diastolic function assessment is challenging. Spot extra cardiac targets and AVV dysfunctions

Merci