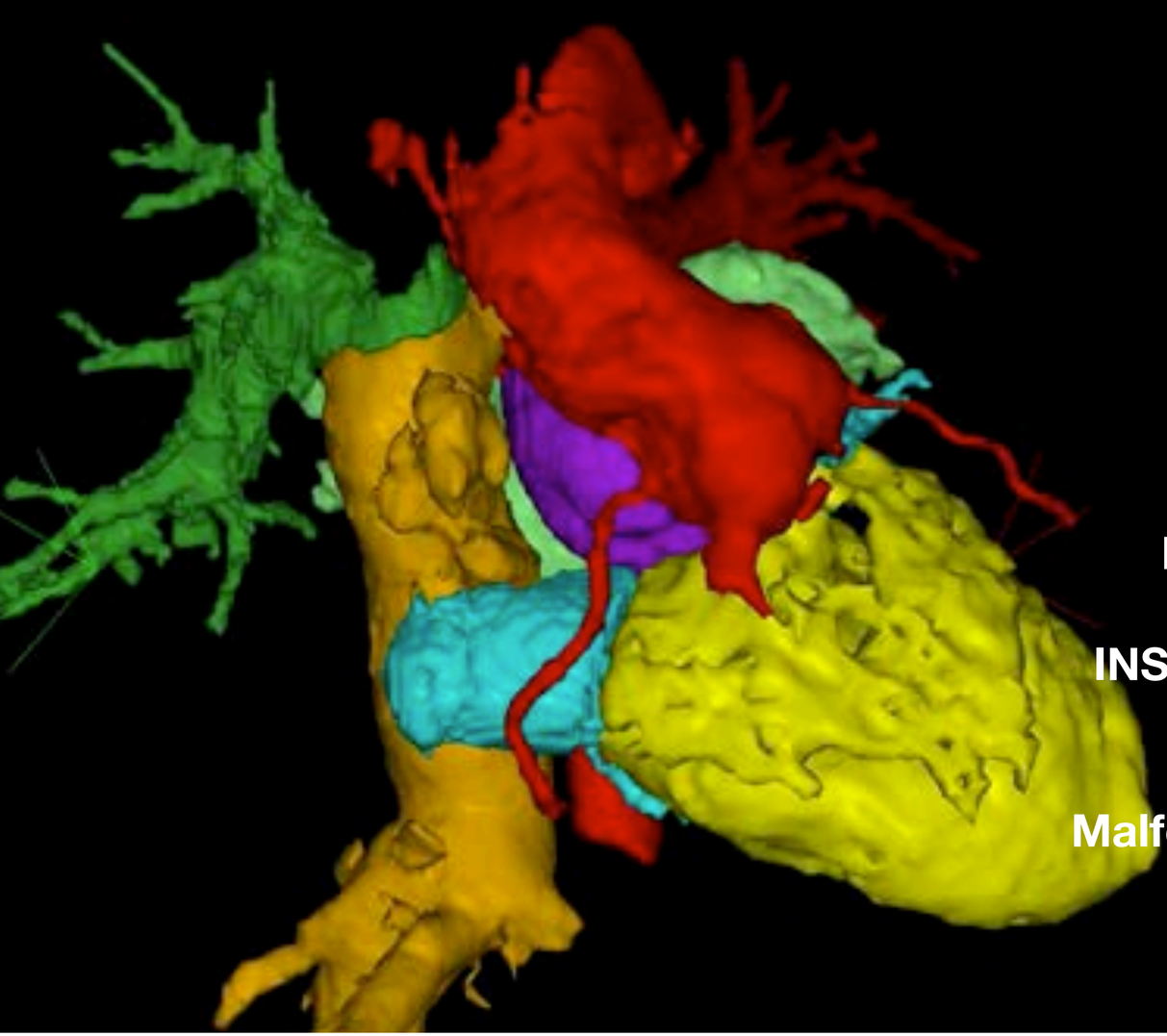

Univentricular hearts

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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

ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS
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Université de Paris

Inserm
Institut national de la santé et de la recherche médicale

cardiogen
Filière nationale de santé maladies cardiaques héréditaires

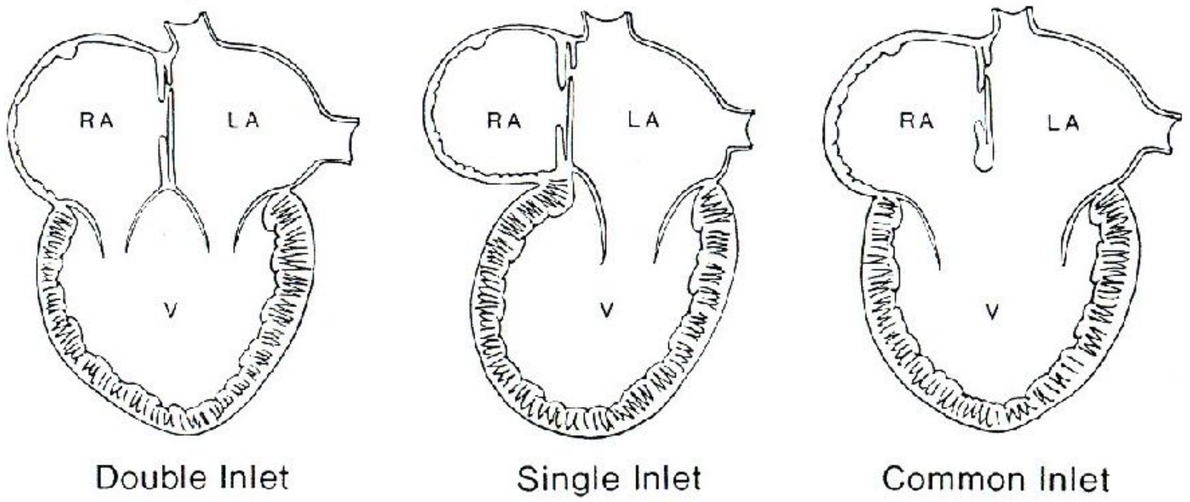
Centre de Référence des Maladies Cardiaques Héréditaires

European Reference Network
for rare or low prevalence complex diseases
Network Respiratory Diseases (ERN-LUNG)

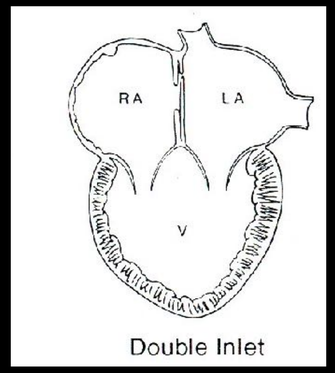
European Reference Network
for rare or low prevalence complex diseases
Network Heart Diseases (ERN GUARD-HEART)

Anatomic definition of univentricular hearts

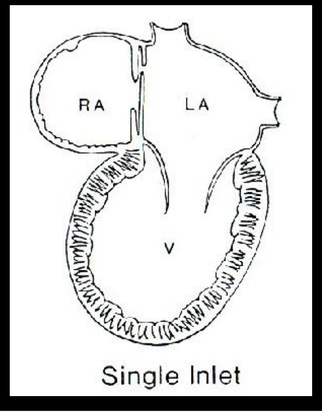
- Unique ventricle definition :
  - Double inlet ventricle
  - The two AV junctions are connected to a single ventricle
  - or the AV connection is unique
- AV valves
  - Both patent
  - One atretic
  - Common AV valve
- Accessory ventricle
  - does not have an inlet
  - Has an outlet
  - Hypoplastic apex



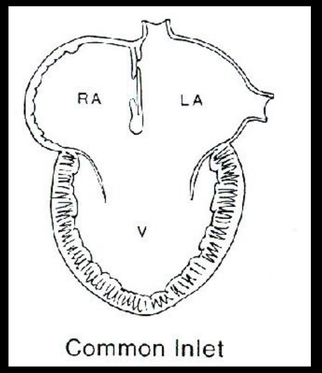
Double Inlet Single Inlet Common Inlet



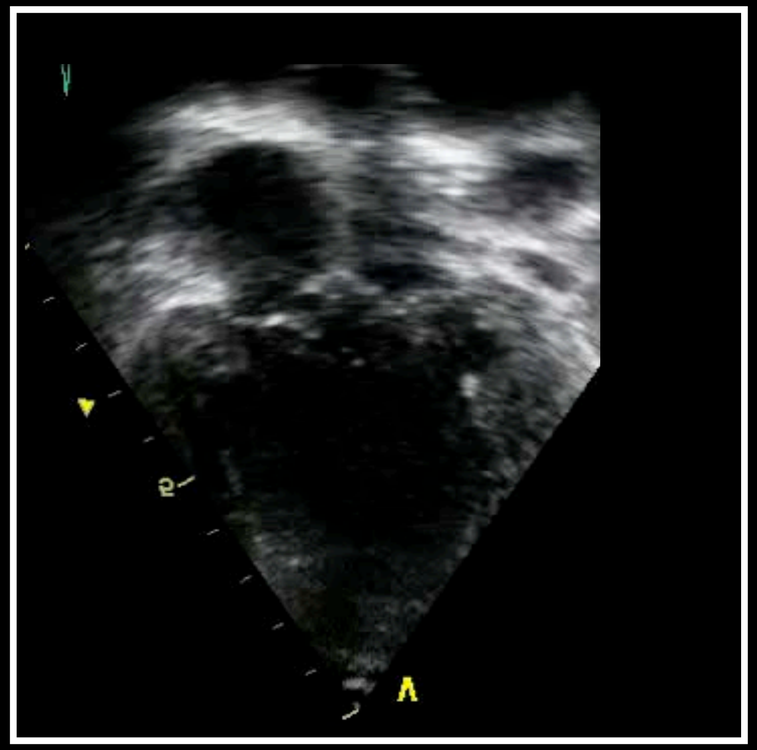
Double Inlet

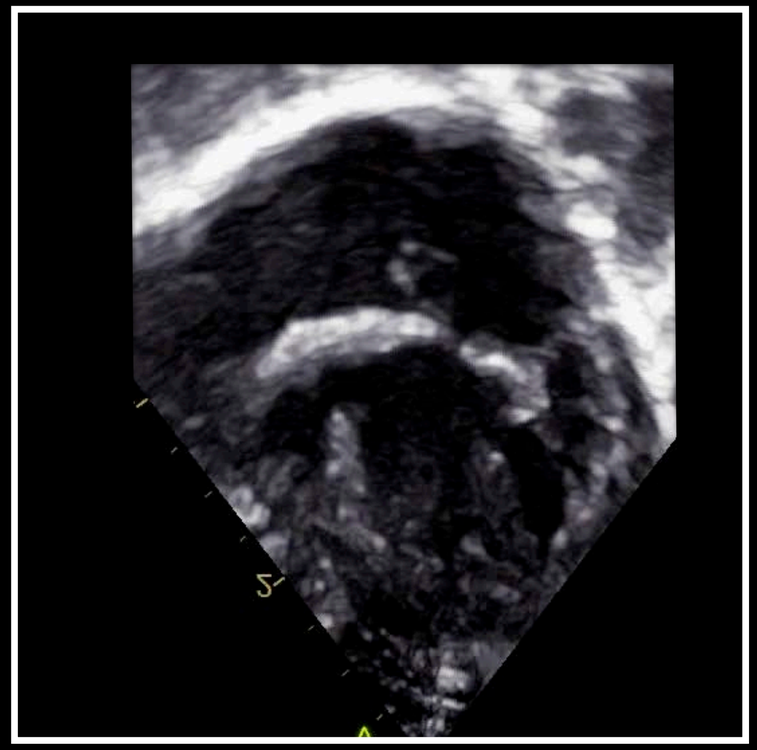


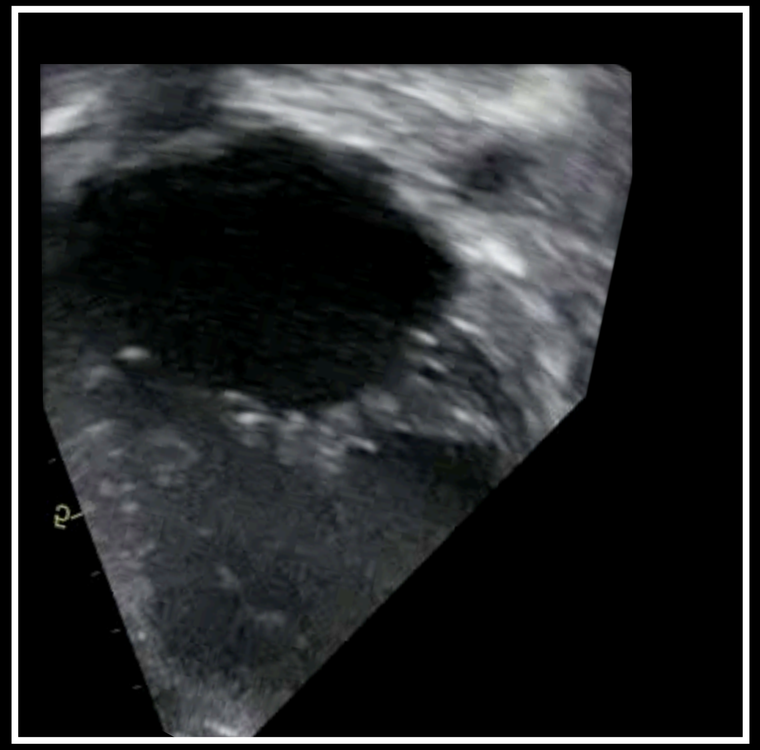
Single Inlet

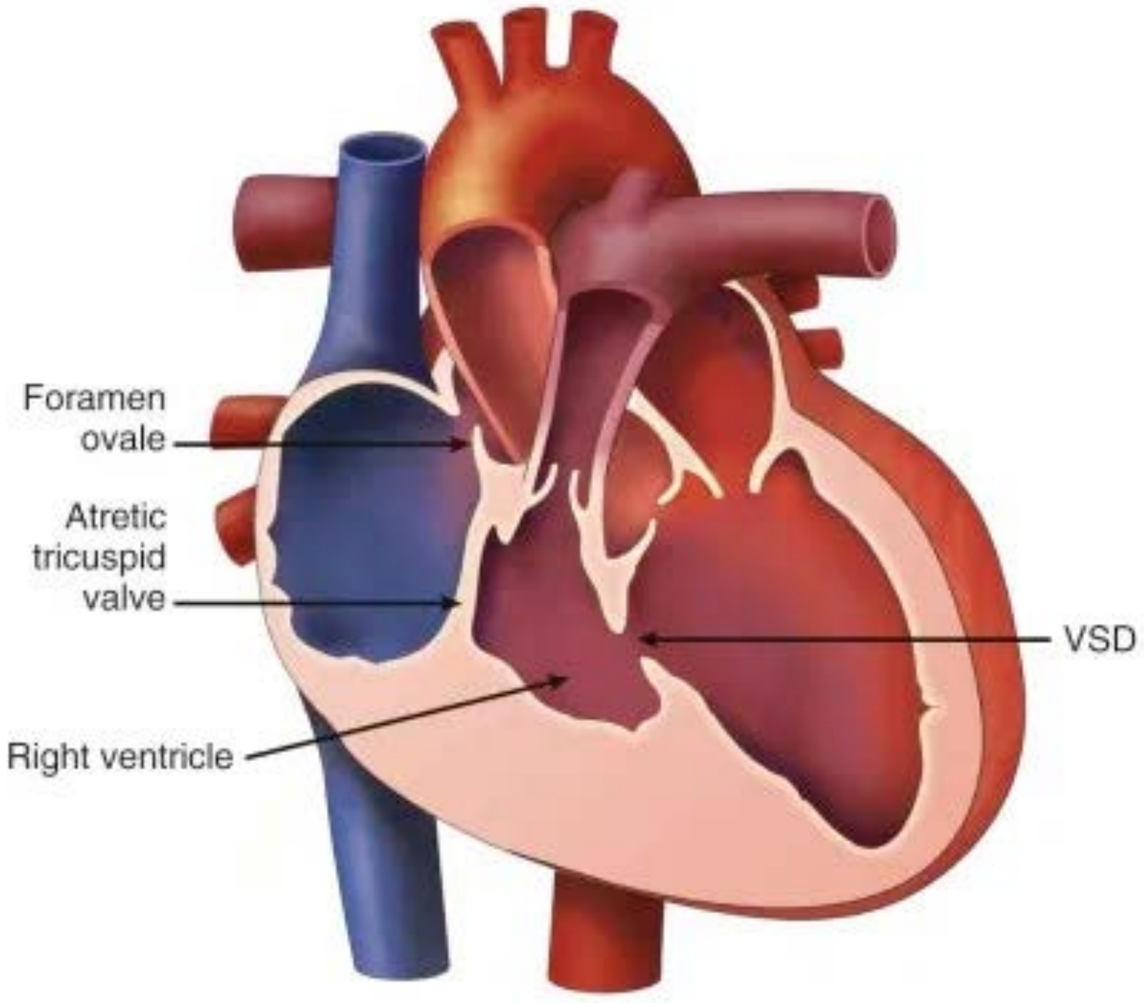


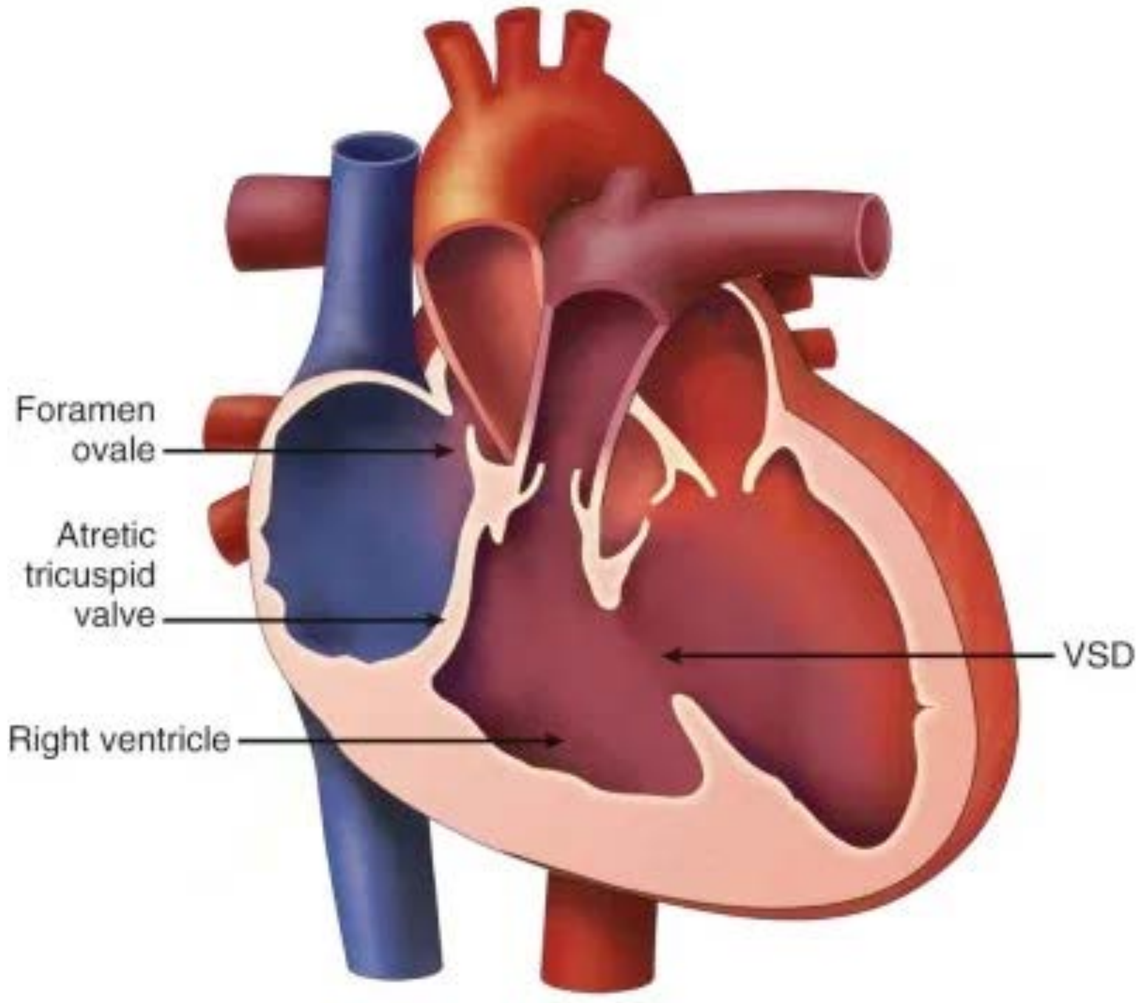
Common Inlet





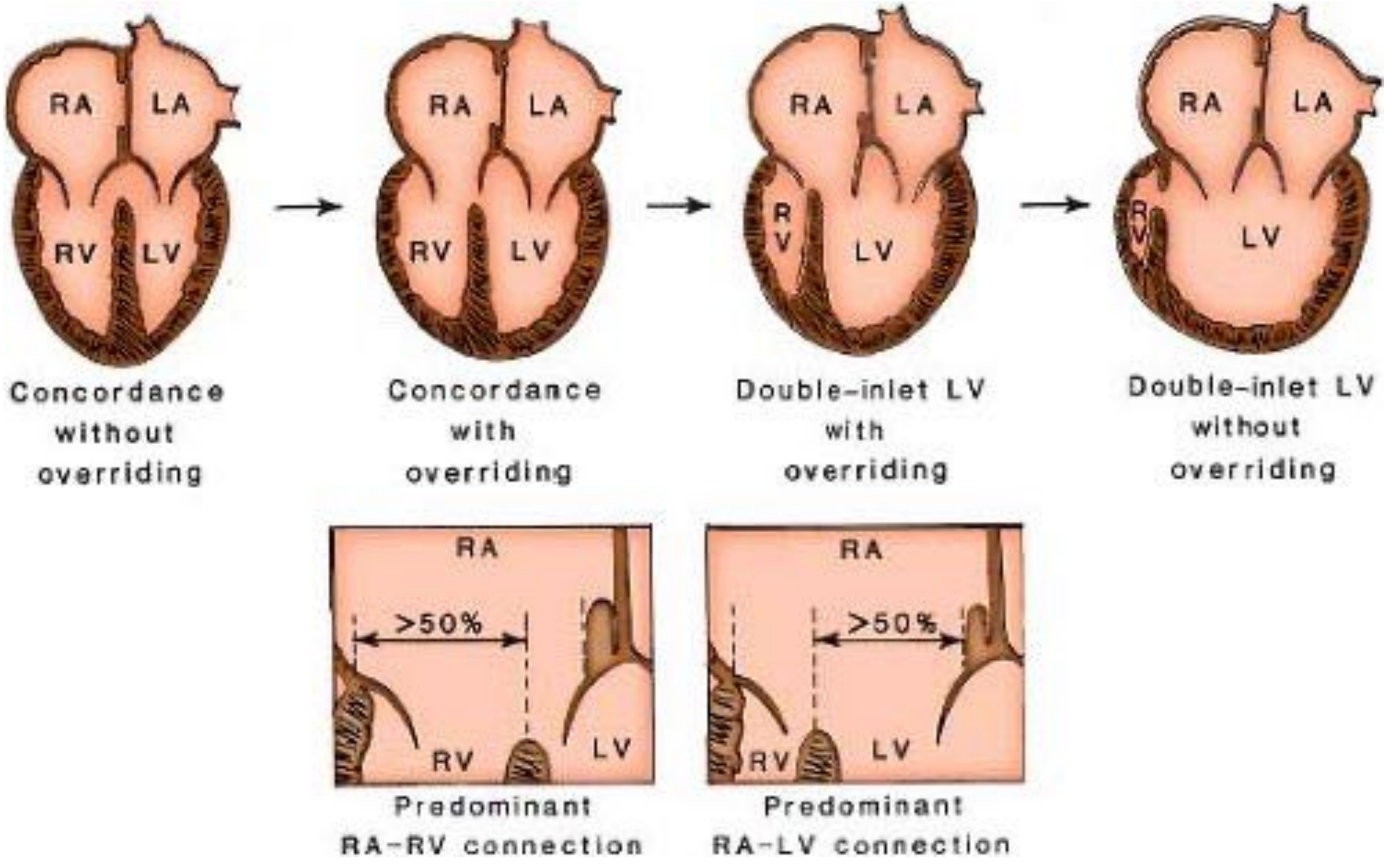



Foramen ovale
Atretic tricuspid valve
VSD
Right ventricle

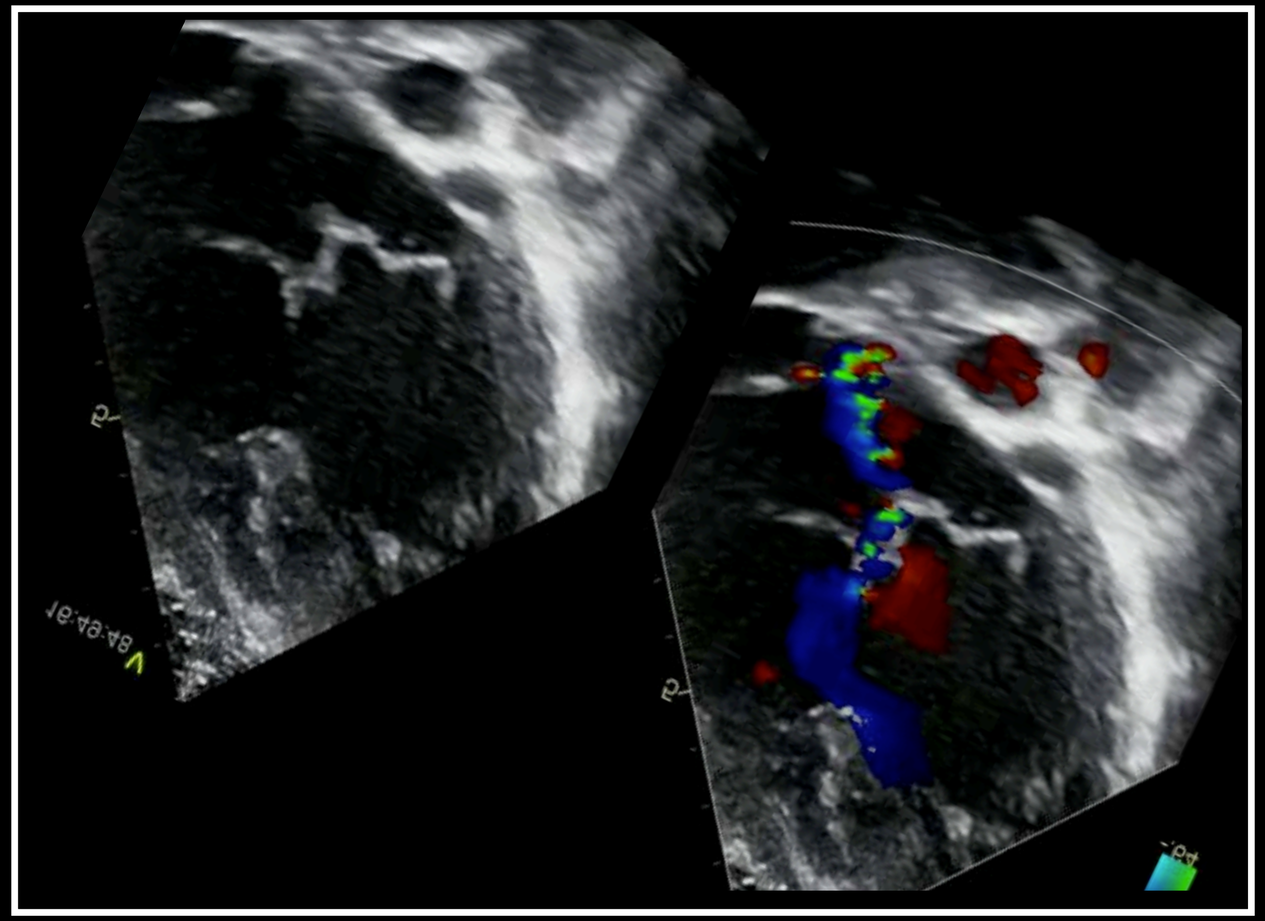

Foramen ovale
Atretic tricuspid valve
VSD
Right ventricle

Physiological definition of univentricular hearts

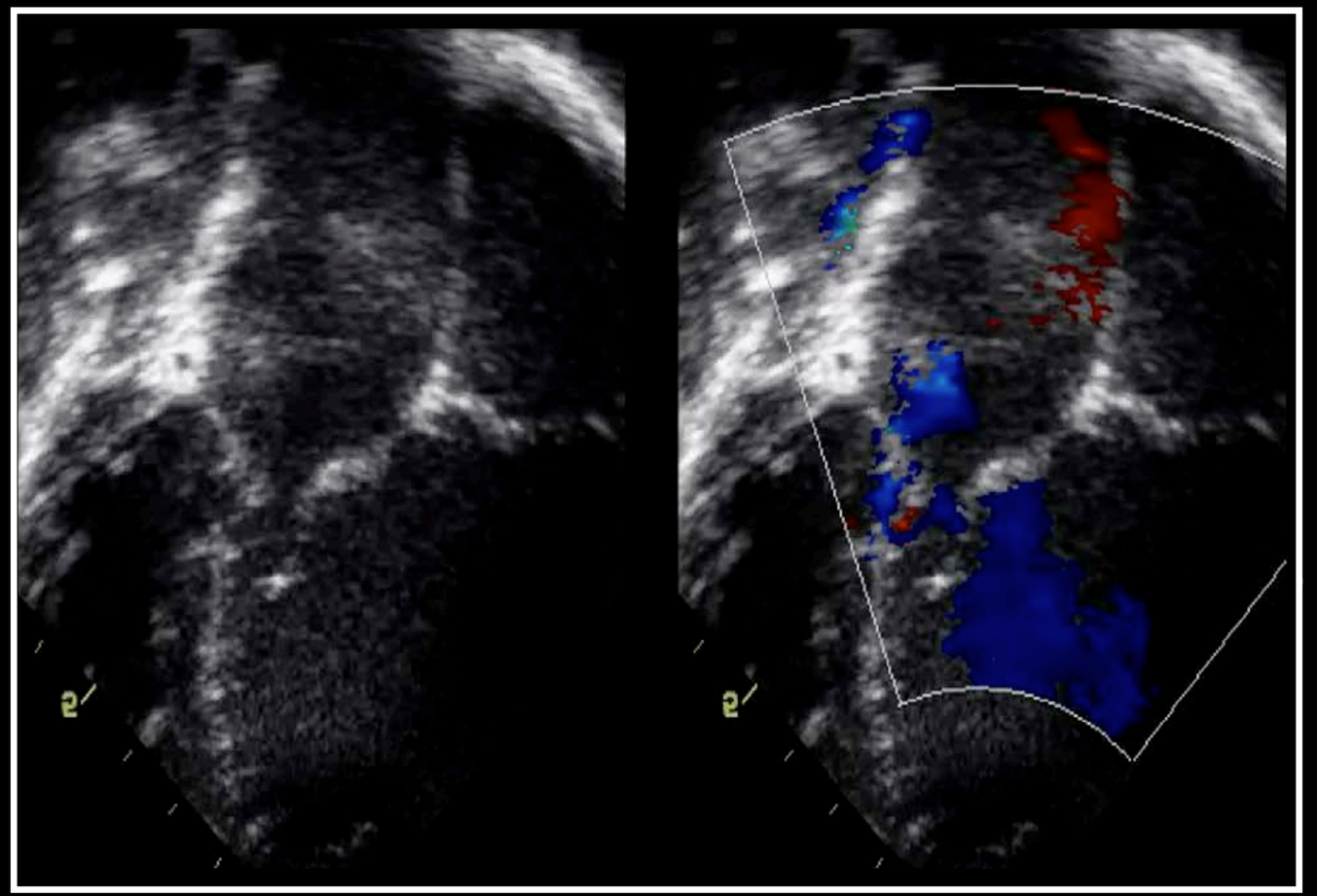
- All hearts for which biventricular repair is not possible
- 2 big categories
  - AV connection of the univentricular type
    - Double inlet ventricle
      - left, right, not defined
    - Absence of one AV connection
      - Tricuspid atresia, mitral atresia, imperforate valve
  - AV connection of the biventricular type
    - Very large or mutliple VSDs
    - Hypoplastic left hearts
      - HLHS, small left ventricles with multiple obstructions, DORV with non committed VSD and small left ventricle
    - Hypoplastic right hearts
      - PA-IVS, hypoplastic right ventricle
    - Unbalanced AVSD
    - Straddling of one AV valve with hypoplasia of one ventricle

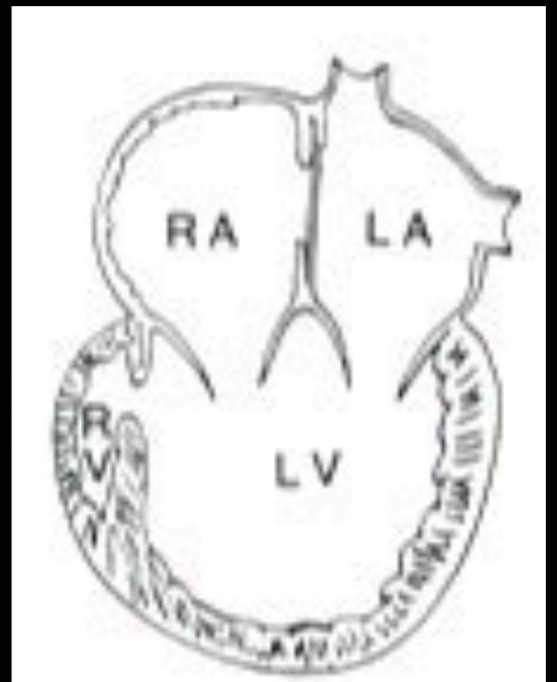

RA
LA
RV
LV
Concordance without overriding
Concordance with overriding
Double-inlet LV with overriding
Double-inlet LV without overriding
>50%
Predominant RA-RV connection
Predominant RA-LV connection


RA
LA
RV
LV
Concordance
with
overriding

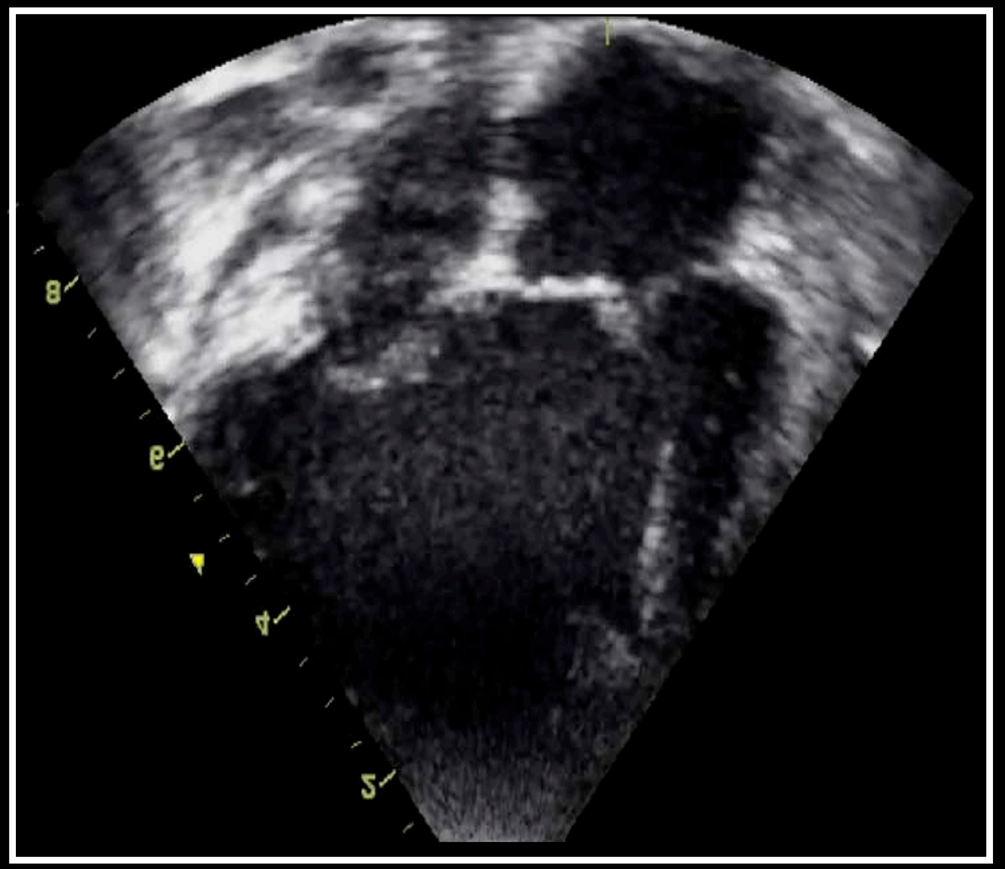



RA
LA
R
V
LV
Double Inlet LV
with
Overriding





Double Inlet LV without Overriding

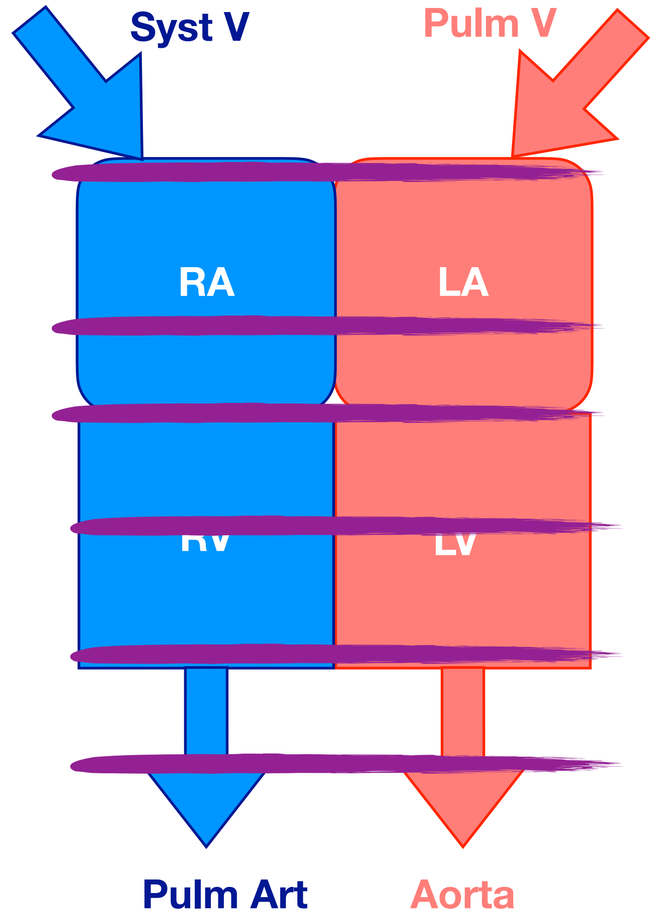


The EPICARD study

EPIdémiologie des enfants ou fœtus ayant une **CARD**iopathie congénitale

Anatomic and Clinical Classification of Congenital Heart Diseases ACC-CHD

ACC-CHD categories	Examples
Heterotaxy	Heterotaxy syndromes
Anomalies of venous connections	Total anomalous pulmonary venous return
Anomalies of atria	Atrial septal defect
Anomalies of AV junction and AV valves	Atrioventricular septal defect
Complex anomalies of AV junction	Double discordance
Functionally univentricular heart	Hypoplastic left heart syndrome
Ventricular septal defects	Perimembranous VSD
Anomalies of ventriculo-arterial connections	Transposition of the great arteries, DORV
Anomalies of extra pericardial trunks	Coarctation of the aorta
Congenital anomalies of coronary arteries	ALCAPA



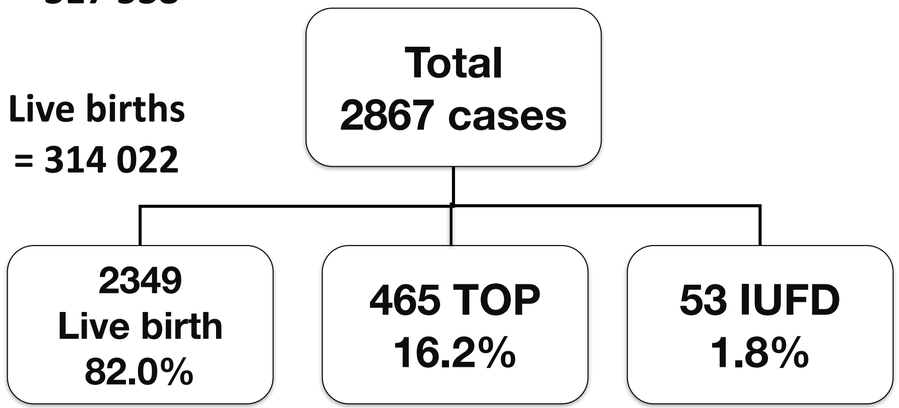
Houyel L, Bonnet D. Orphanet J 2011

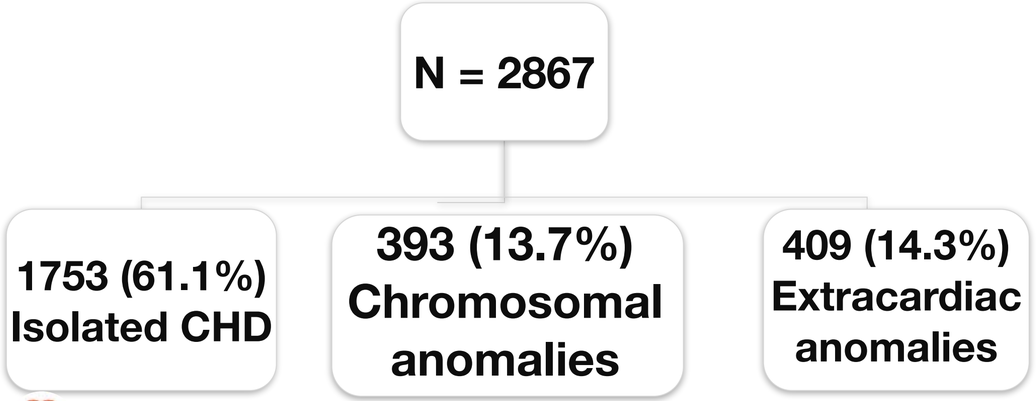
Prevalence, pre- and post-natal diagnosis, and infant mortality of newborns with congenital heart defects: A population-based study using the International Paediatric and Congenital Cardiac Code (IPCCC)

The EPICARD Study Group

Total number of birth = 317 538

Live births = 314 022





Distribution of categories of CHD and associated anomalies

ACC-CHD categories	Total		Live births	
	% of chromosomal anomalies	% of extra cardiac anomalies	% of chromosomal anomalies	% of extra cardiac anomalies
Heterotaxy	0	24.3	0	25.0
Anomalies of venous connections	19.4	16.1	7.7	15.4
Anomalies of atria	9.9	19.8	7.5	19.0
Anomalies of AV junction and AV valves	57.3	12.7	43.1	13.8
Complex anomalies of AV junction	0	7.7	0	0
Functionally univentricular heart	15.8	19.6	8.3	20.8
Ventricular septal defects	9.3	11.1	3.9	11.0
Anomalies of ventriculo-arterial connections	10.7	18.8	4.5	14.1
Anomalies of extra pericardial trunks	15.9	31.2	3.2	26.4
Congenital anomalies of coronary arteries	0	0	0	0

Khoshnood B et al. Heart 2012;98:1667-73

Prevalence, pre- and post-natal diagnosis, and infant mortality of newborns with congenital heart defects

A population-based study using the International Paediatric and Congenital Cardiac Code (IPCCC)

The EPICARD Study Group

Proportion of prenatal diagnosis

All CHDs

ACC-CHD categories	% of prenatal diagnosis
All cases excluding chromosomal anomalies	25.6
All cases excluding chromosomal and other extra cardiac anomalies	23
All cases excluding chromosomal, other anomalies and simple VSD	40.2

In categories of CHDs

ACC-CHD categories	% of prenatal diagnosis (n)
Heterotaxy	89.2 (37)
Anomalies of venous connections	16.0 (25)
Anomalies of atria	4.3 (164)
Anomalies of AV junction and AV valves	67.0 (91)
Complex anomalies of AV junction	100.0 (13)
Functionally univentricular heart	92.5 (133)
Ventricular septal defects	9.6 (1353)
Anomalies of ventriculo-arterial connections	39.2 (503)
Anomalies of extra pericardial trunks	44.7 (143)
Congenital anomalies of coronary arteries	0 (9)

Specific CHDs

Type of CHD	% of prenatal diagnosis
Congenitally corrected transposition of the great	100
Functionally univentricular heart	92.5
TGA	74
DORV	98

Khoshnood B et al. Heart 2012;98:1667-73

Prevalence, pre- and post-natal diagnosis, and infant mortality of newborns with congenital heart defects:
A population-based study using the International Paediatric and Congenital Cardiac Code (IPCCC)

The EPICARD Study Group

Live birth - Termination of pregnancy

All CHDs

ACC-CHD categories	% TOP
All cases excluding chromosomal anomalies	9.8
All cases excluding chromosomal and other extra cardiac anomalies	6.4
All cases excluding chromosomal, other anomalies and simple VSD	14.0

In categories of CHDs

ACC-CHD categories	% TOP
Heterotaxy	75.7
Anomalies of venous connections	16.1
Anomalies of atria	4.4
Anomalies of AV junction and AV valves	42.7
Complex anomalies of AV junction	46.2
Functionally univentricular heart	**62.7**
Ventricular septal defects	5.7
Anomalies of ventriculo-arterial connections	18.5
Anomalies of extra pericardial trunks	23.5
Congenital anomalies of coronary arteries	0

Khosnood B et al. Heart 2012;98:1667-73

Pre- and postnatal diagnosis in CHD without chromosomal anomalies
The EPICARD Study Group

ACC-CHD categories	Prenatal diagnosis	Postnatal diagnosis			
	%	<7days	8-28 days	29 days-3 months	3 months -1 year
Heterotaxy	89.2	8.1	0.0	2.7	0.0
Anomalies of venous connections	16.0	32.0	16.0	28.0	4.0
Anomalies of atria	4.3	29.3	26.8	26.8	11.6
Anomalies of AV junction and AV valves	67.0	19.8	3.3	2.2	2.2
Complex anomalies of AV junction	100.0	0.0	0.0	0.0	0.0
Functionally univentricular heart	92.5	6.0	0.7	0.0	0.0
Ventricular septal defects	9.6	67.4	9.0	9.8	3.8
Anomalies of ventriculo-arterial connections	39.2	29.6	7.1	14.5	5.4
Anomalies of extra pericardial trunks	44.7	28.7	9.8	10.5	2.1
Congenital anomalies of coronary arteries	0	0	0	44.4	55.6
All except chromosomal anomalies and /or anomalies of other systems	**40.2**	**28.7**	**10.4**	**14.4**	**5.6**

Infant mortality in newborns with congenital heart defects
The EPICARD Study Group

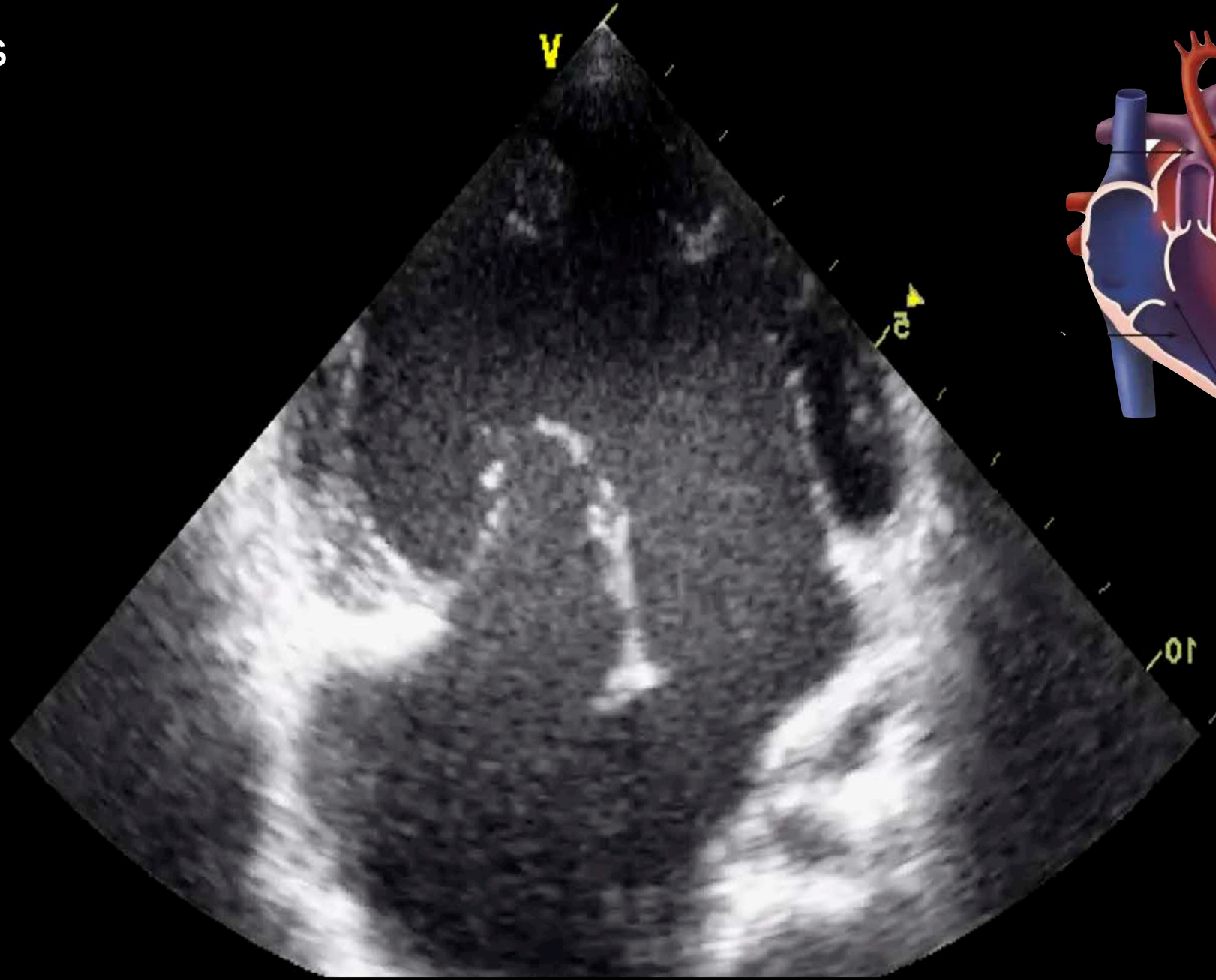
ACC-CHD categories	Prenatal diagnosis	Postnatal diagnosis			Infant mortality	
	N	<7days %	8-28 days %	29 days-1 year %	%	95%CI
Heterotaxy	8	25.0	0.0	12.5	**37.5**	8.5-75.5
Anomalies of venous	26	3.9	11.5	11.5	**26.9**	11.6-47.8
Anomalies of atria	174	0.6	0.6	2.3	**3.5**	1.3-7.3
Anomalies of AV junction and AV valves	109	8.3	7.3	12.8	**28.4**	20.2-37.0
Complex anomalies of AV	7	0.0	0.0	14.3	**14.3**	0.4-57.9
Functionally univentricular	48	41.7	12.5	4.1	**58.3**	43.2-72.4
Ventricular septal defects	1396	0.2	0.5	0.9	**1.6**	1.0-2.4
Anomalies of ventriculo-arterial connections	447	2.3	2.0	4.0	**8.3**	5.9-11.2
Anomalies of extra pericardial trunks	124	3.2	6.5	2.4	**12.1**	6.9-19.2
Congenital anomalies of coronary arteries	9	0	0	11.1	**11.1**	0.3-48.2
All	2348	2.1	1.8	2.5	**6.4**	5.5-7.5
All except chromosomal anomalies and /or anomalies of other systems and IVSD	**784**	**2.9**	**2.2**	**3.6**	**8.7**	**6.8-10.9**

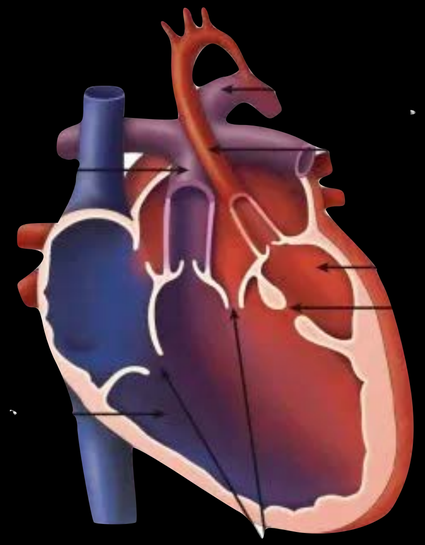
Résumé épidémiologie des cardiopathies univentriculaires

- Les cardiopathies univentriculaires représentent 5 à 7% de toutes les cardiopathies congénitales.
- *Le diagnostic est fait avant la naissance dans plus de 90% des cas depuis 20 ans.*
- Elle sont associées à des anomalies chromosomiques une fois sur 6 et à des anomalies extracardiaques une fois sur 5.
- *Elles représentent environ 20% des cardiopathies motivant un transfert in utero vers un centre tertiaire.*
- Dans 2/3 des cas ce transfert est justifié par la nécessité d'une intervention précoce.
- *Elle représentent avec le diagnostic précoce (<22 SA) et les anomalies extracardiaques, le principal facteur de décision d'interruption de grossesse.*
- Elle sont avec les atteintes extracardiaques majeures la principale cause de mortalité de la première année.
- *La survie à un an des nouveau-nés vivants est de 50% (4% pour les hypoplasies du coeur gauche).*

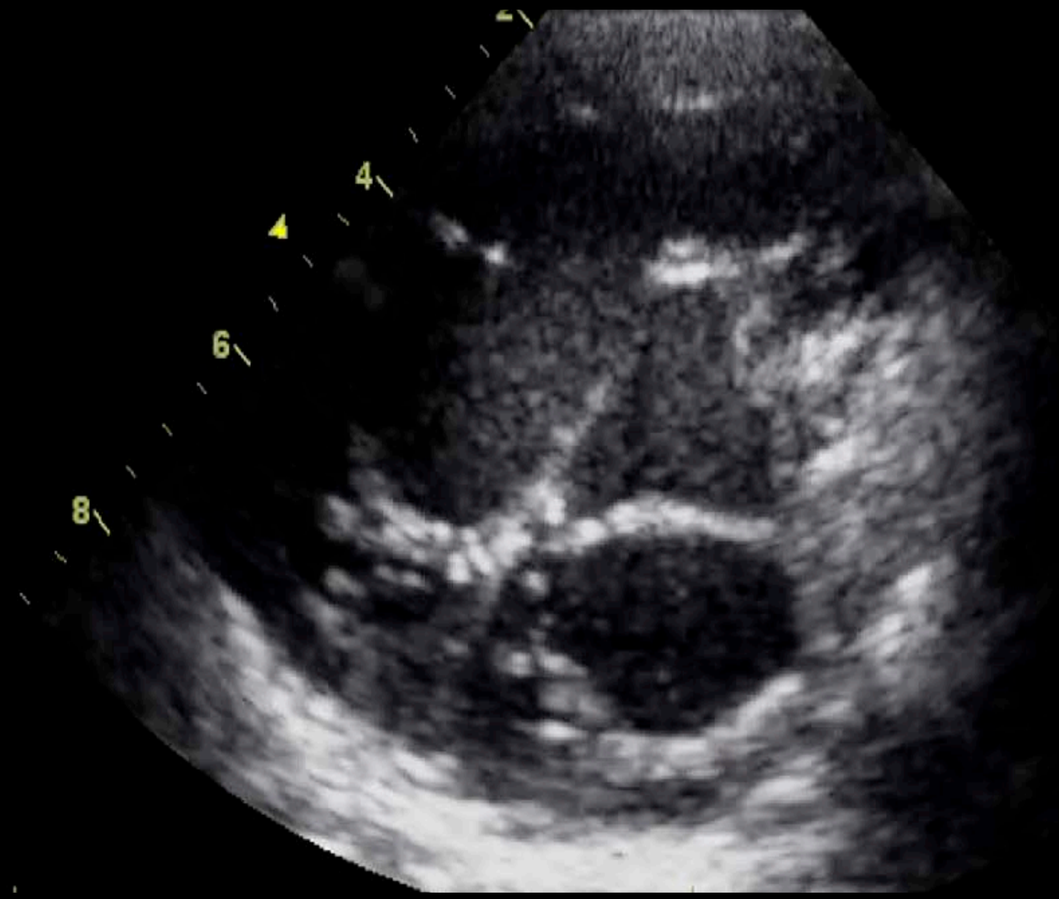
Echocardiography of univentricular hearts

UVH 2 AV Valves
AV Discordance
L-Loop

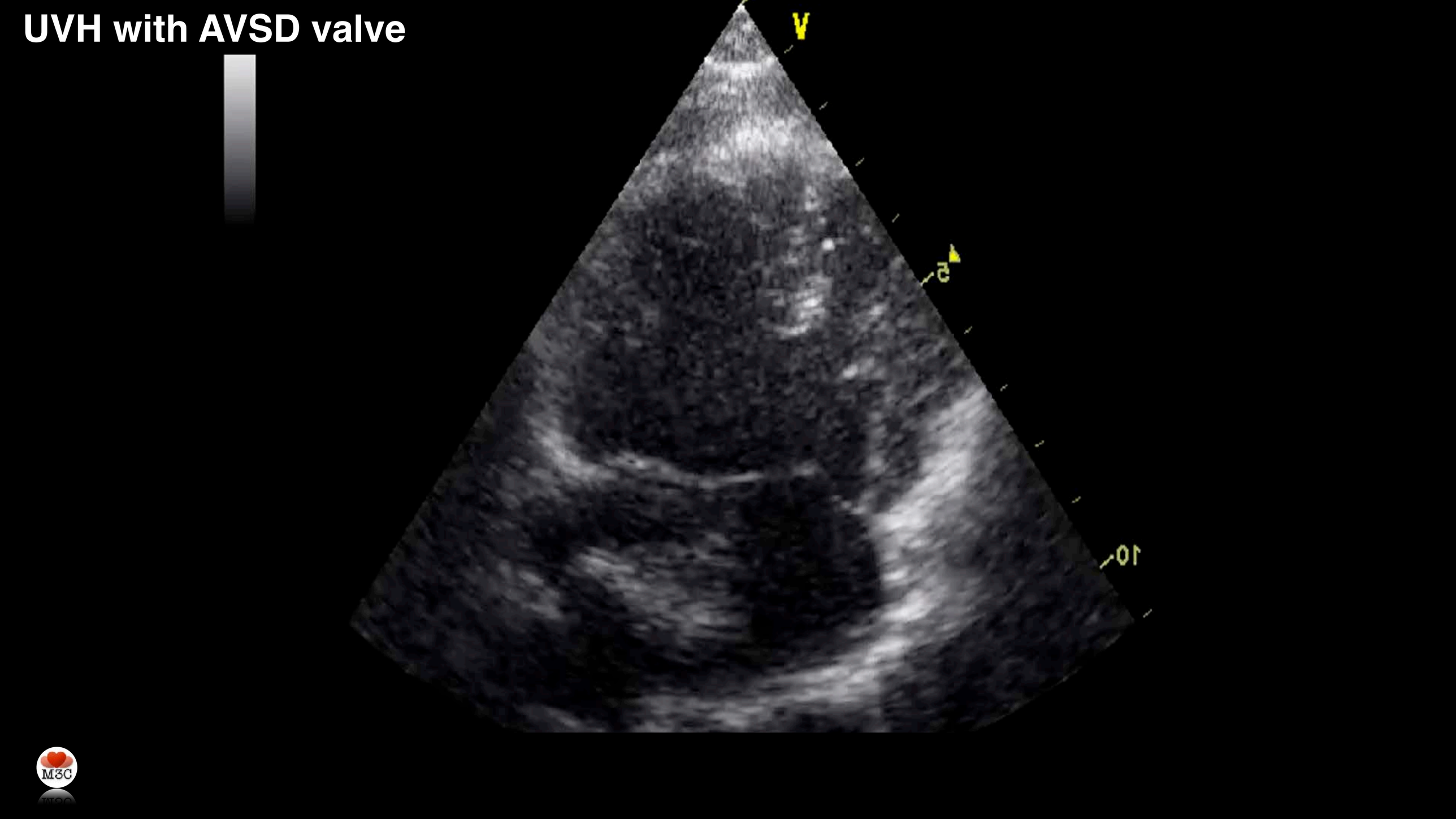




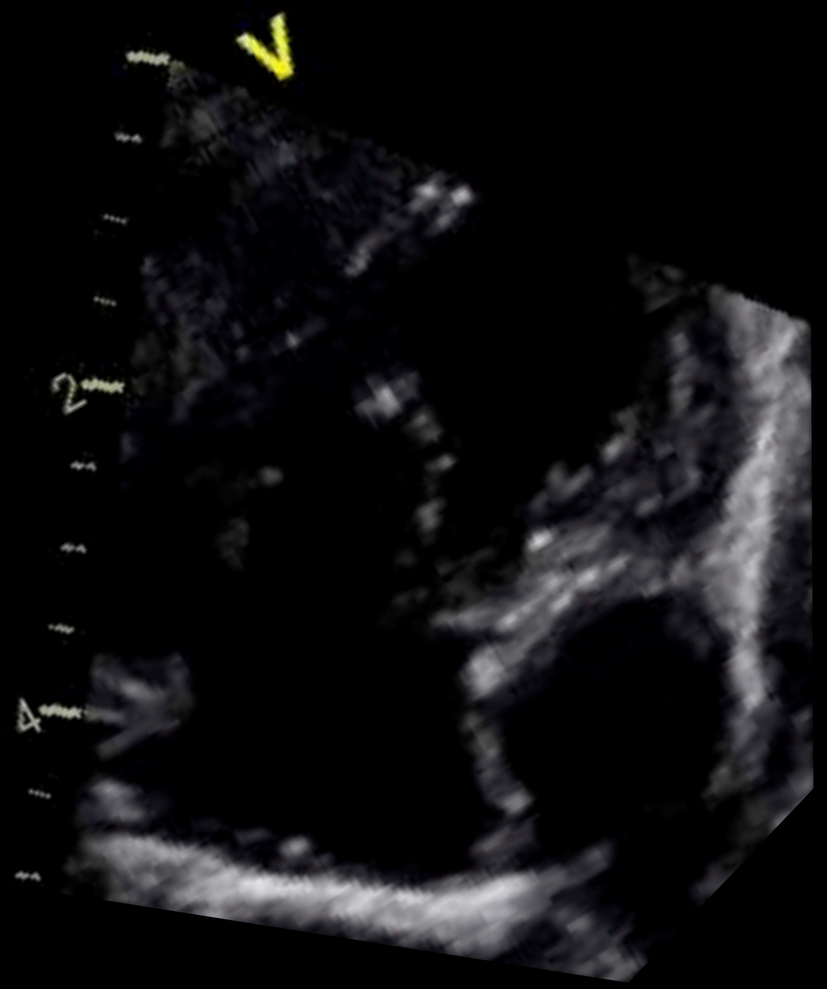
UVH 2 AV Valves



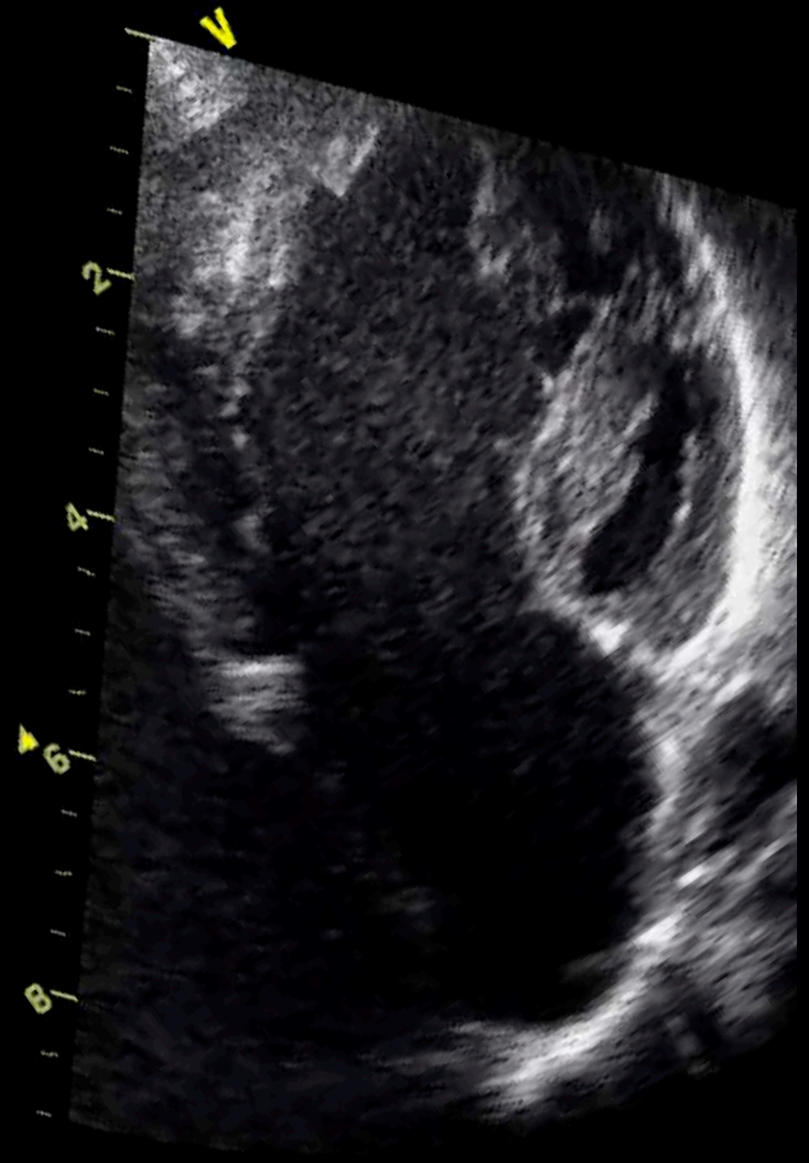
UVH with AVSD valve

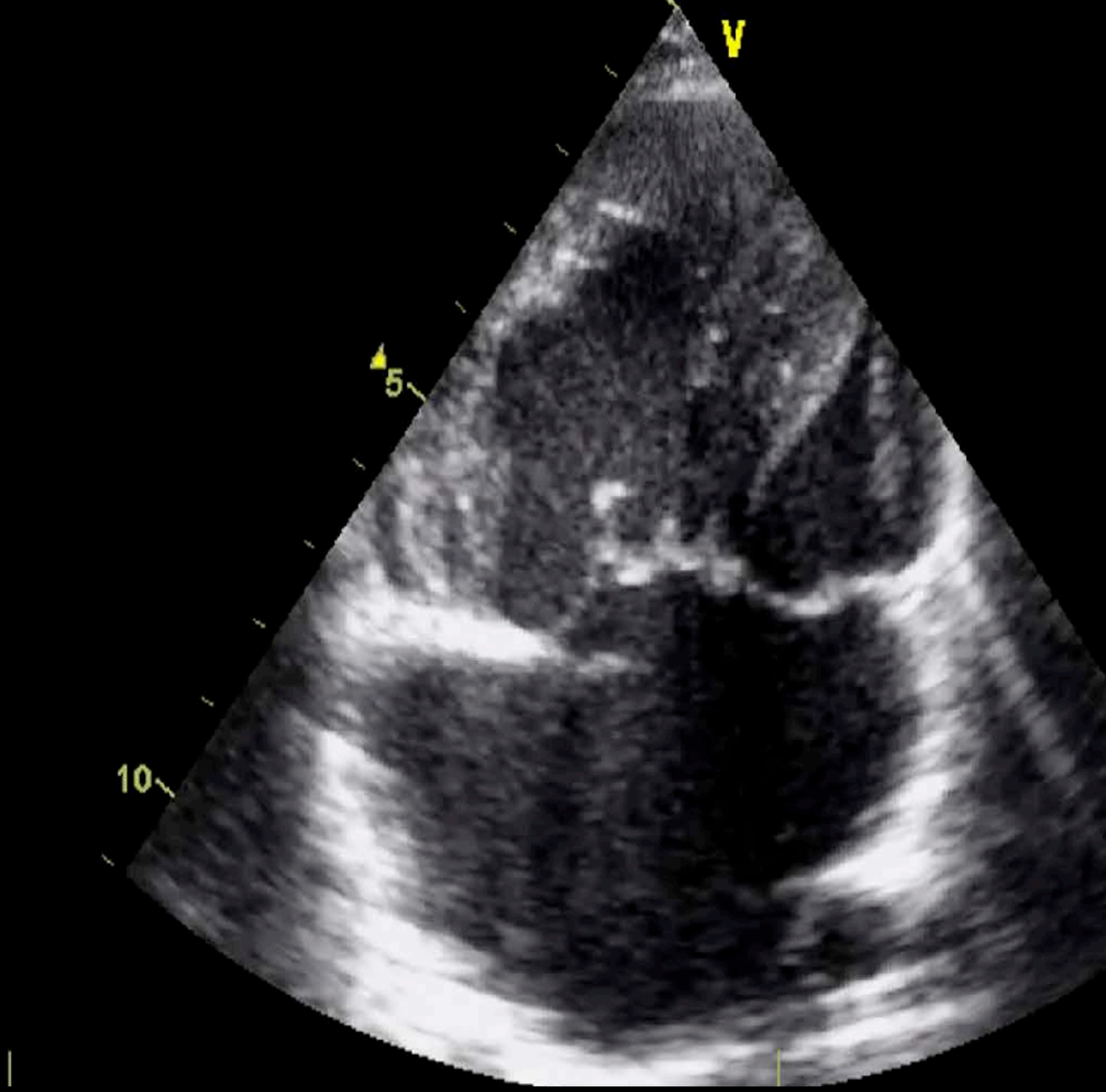


Mitral atresia

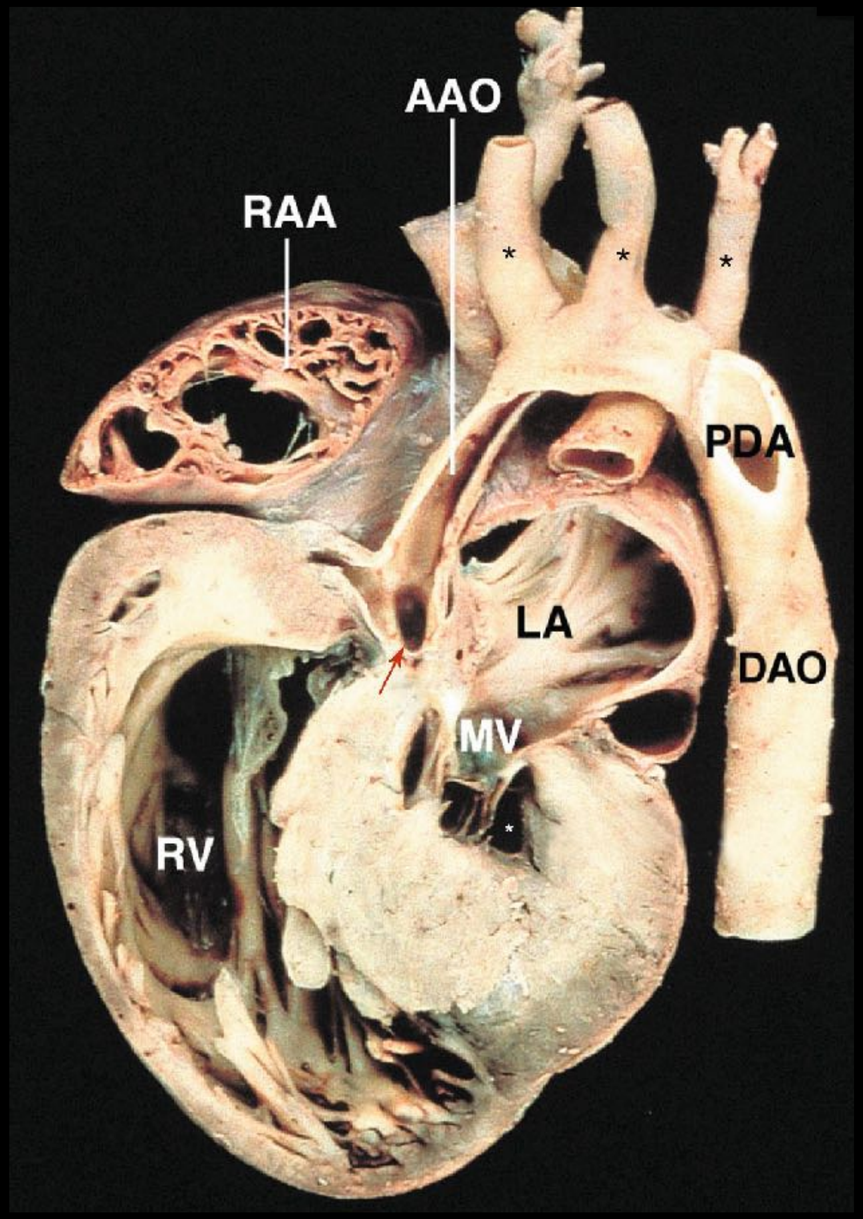


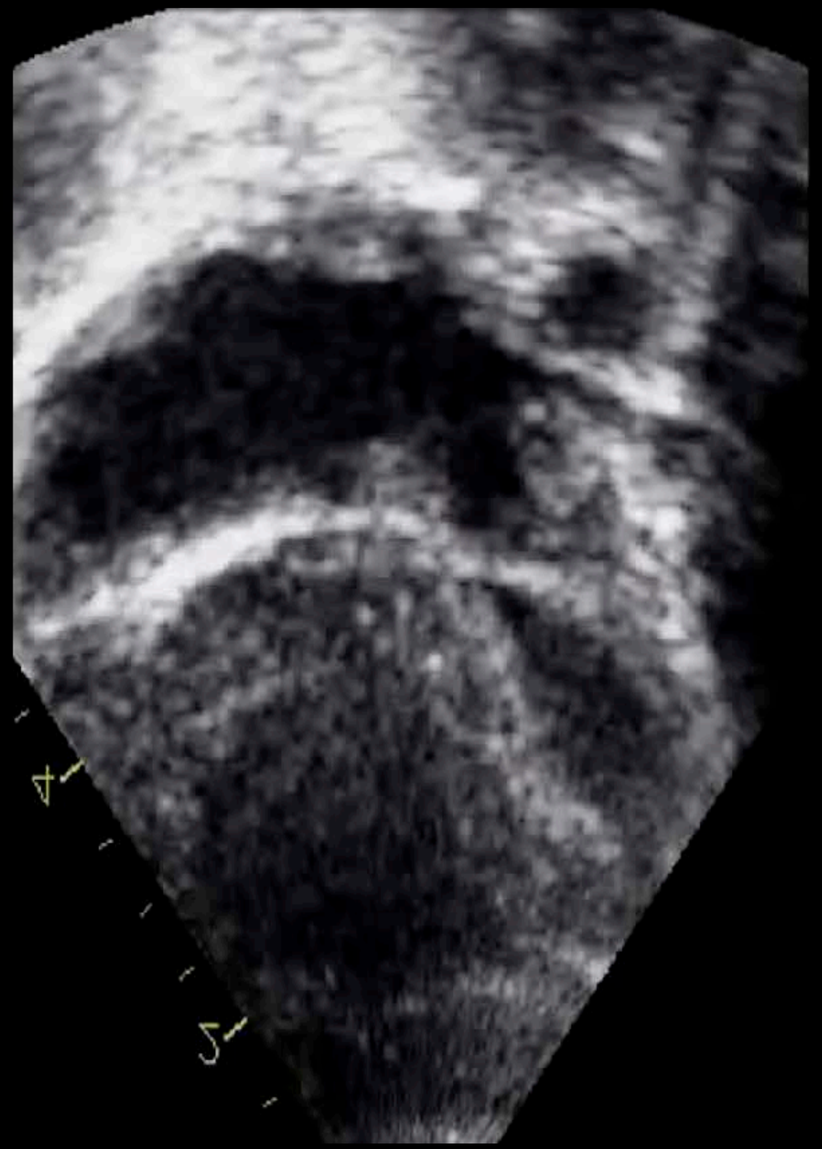
Mitral valve hypoplasia



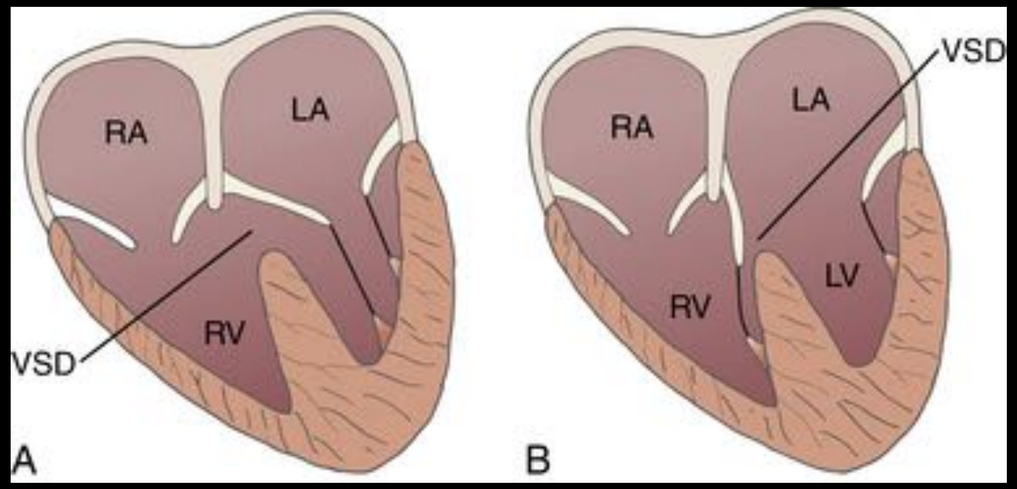


Hypoplastic left heart syndrome

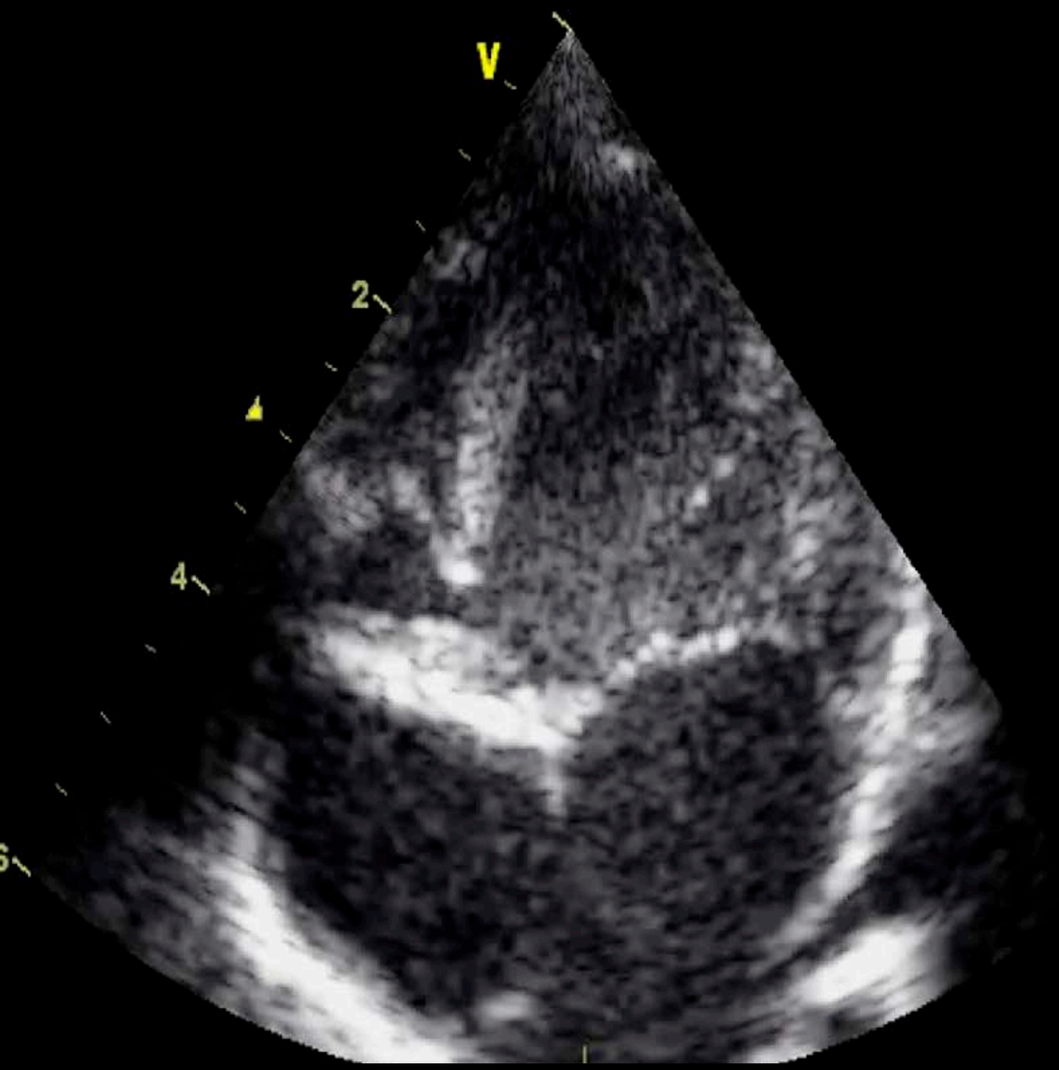


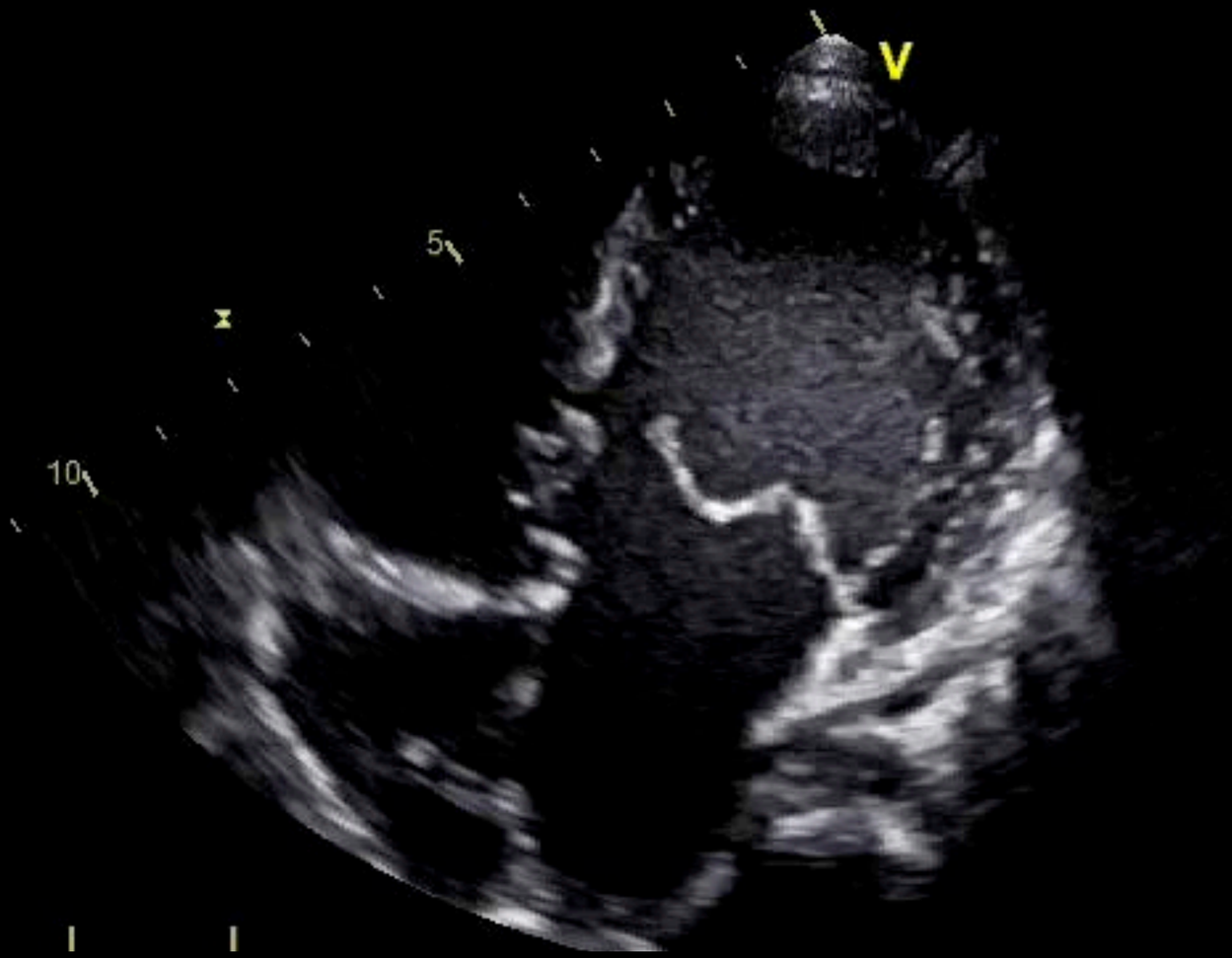


Straddling of the mitral valve

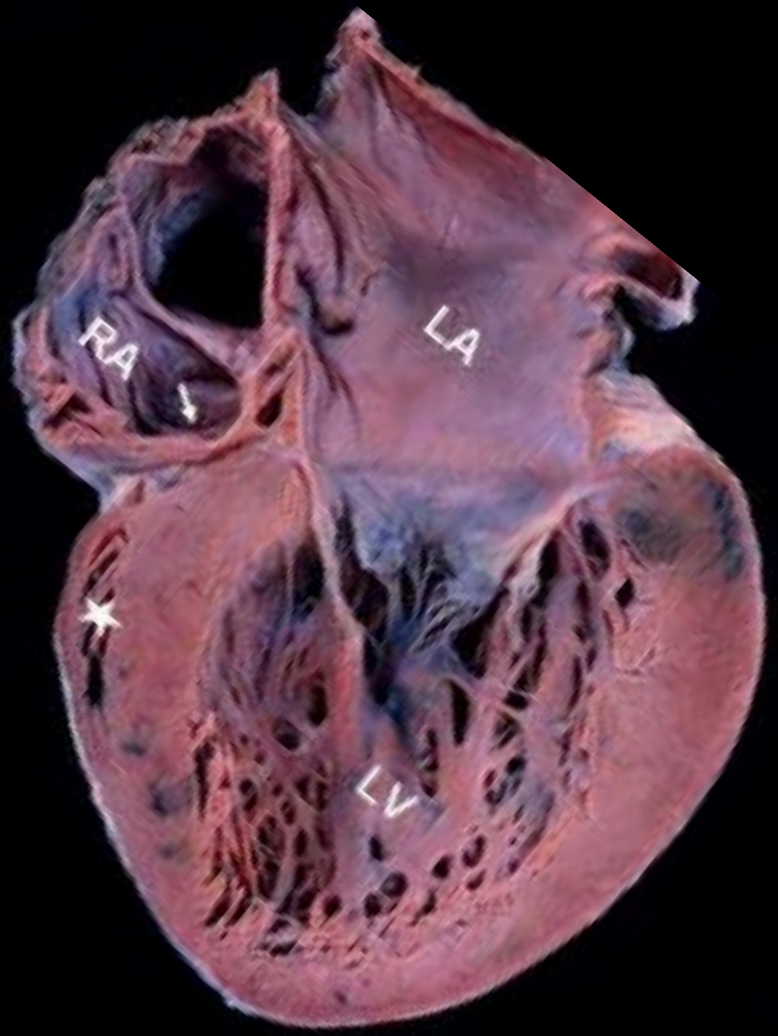


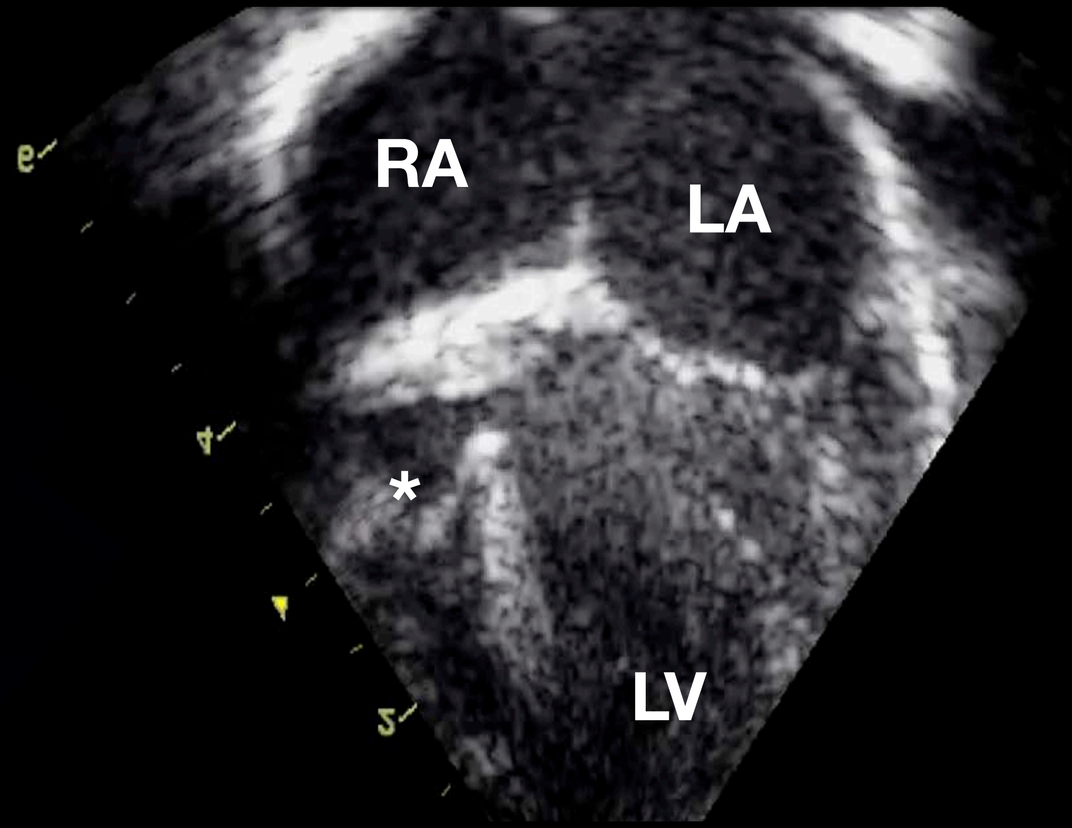
Tricuspid atresia



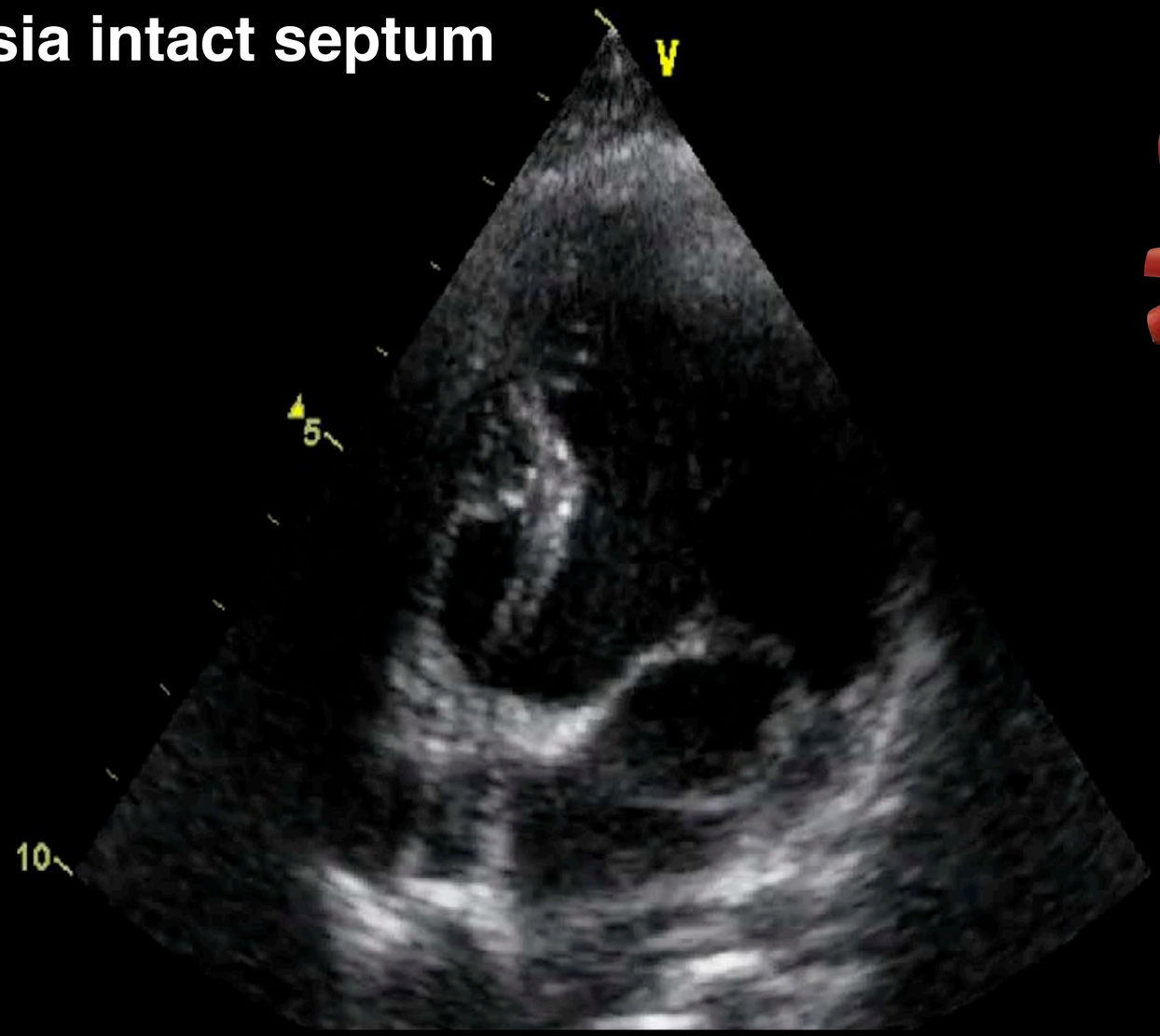


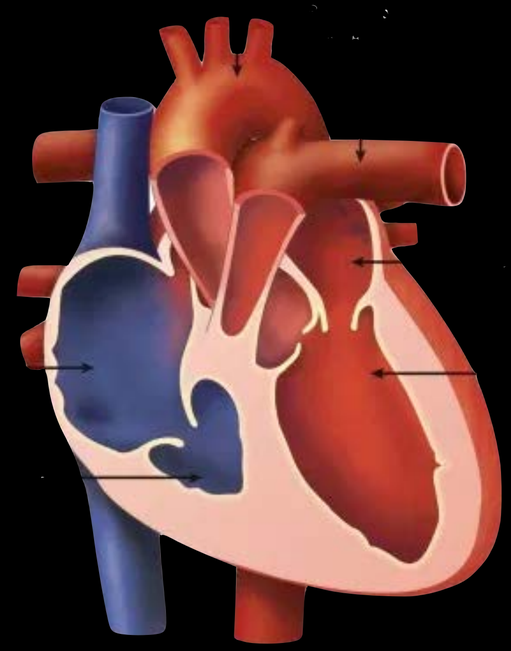
Tricuspid atresia



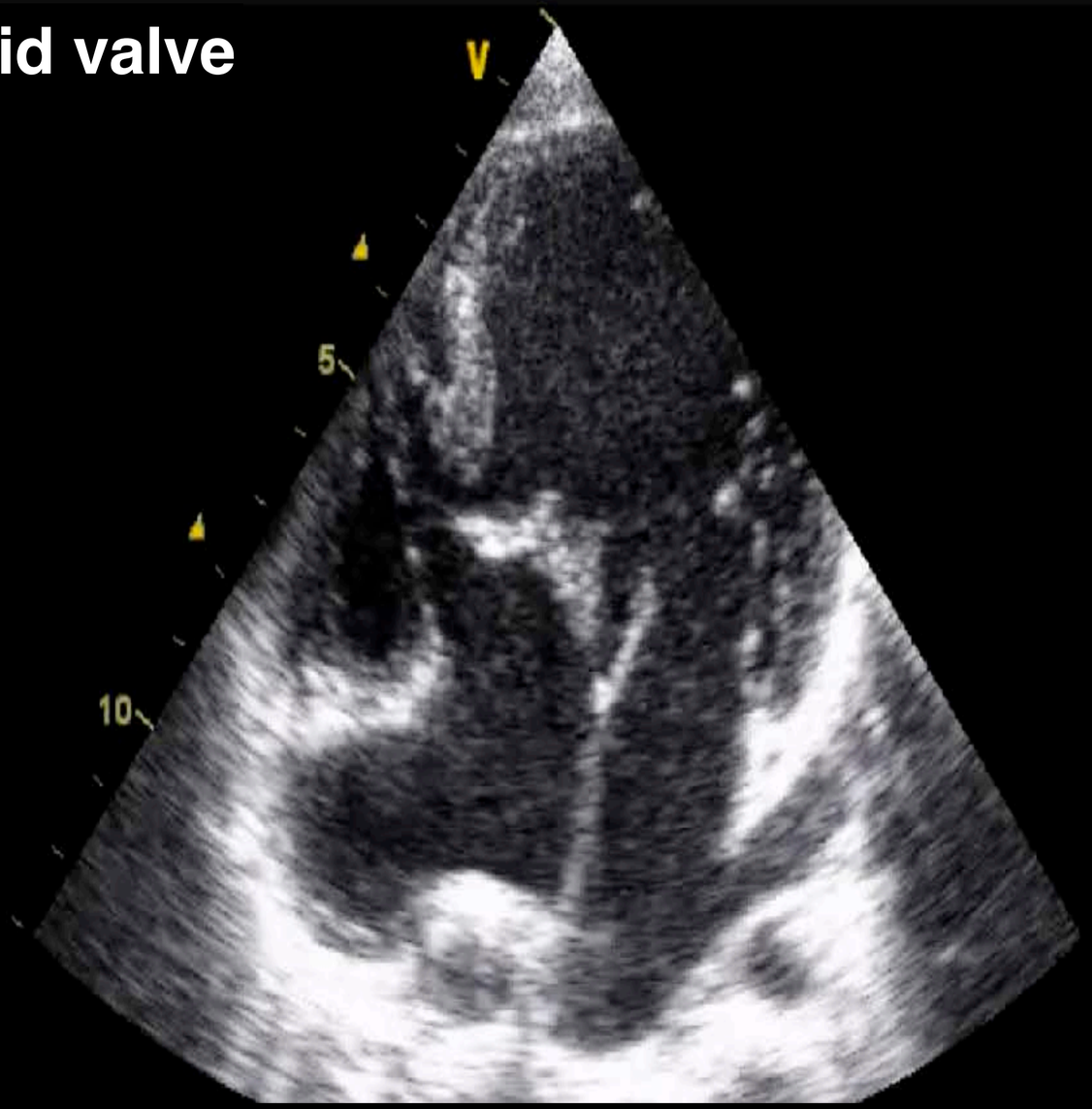


Pulmonary atresia intact septum

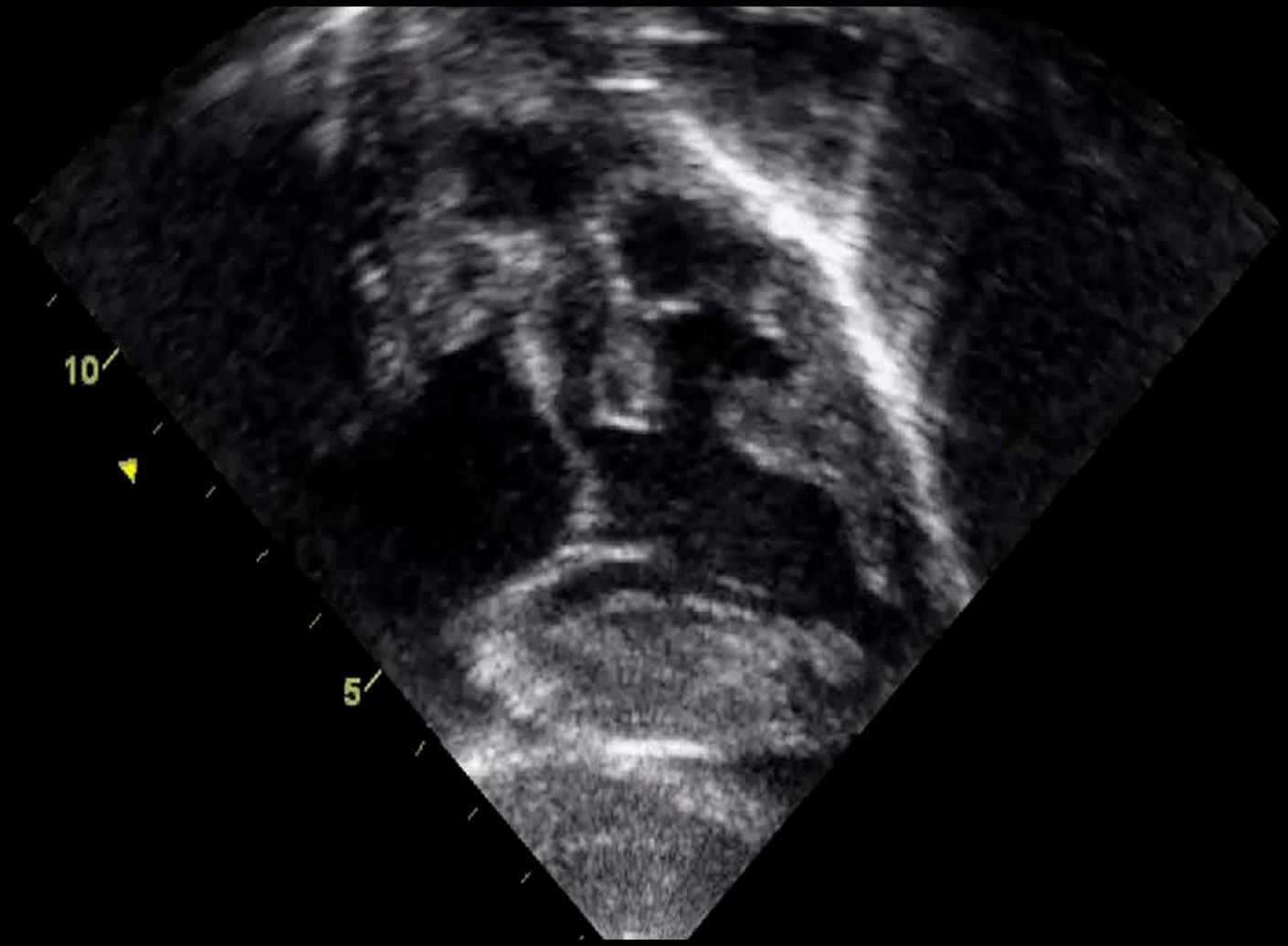




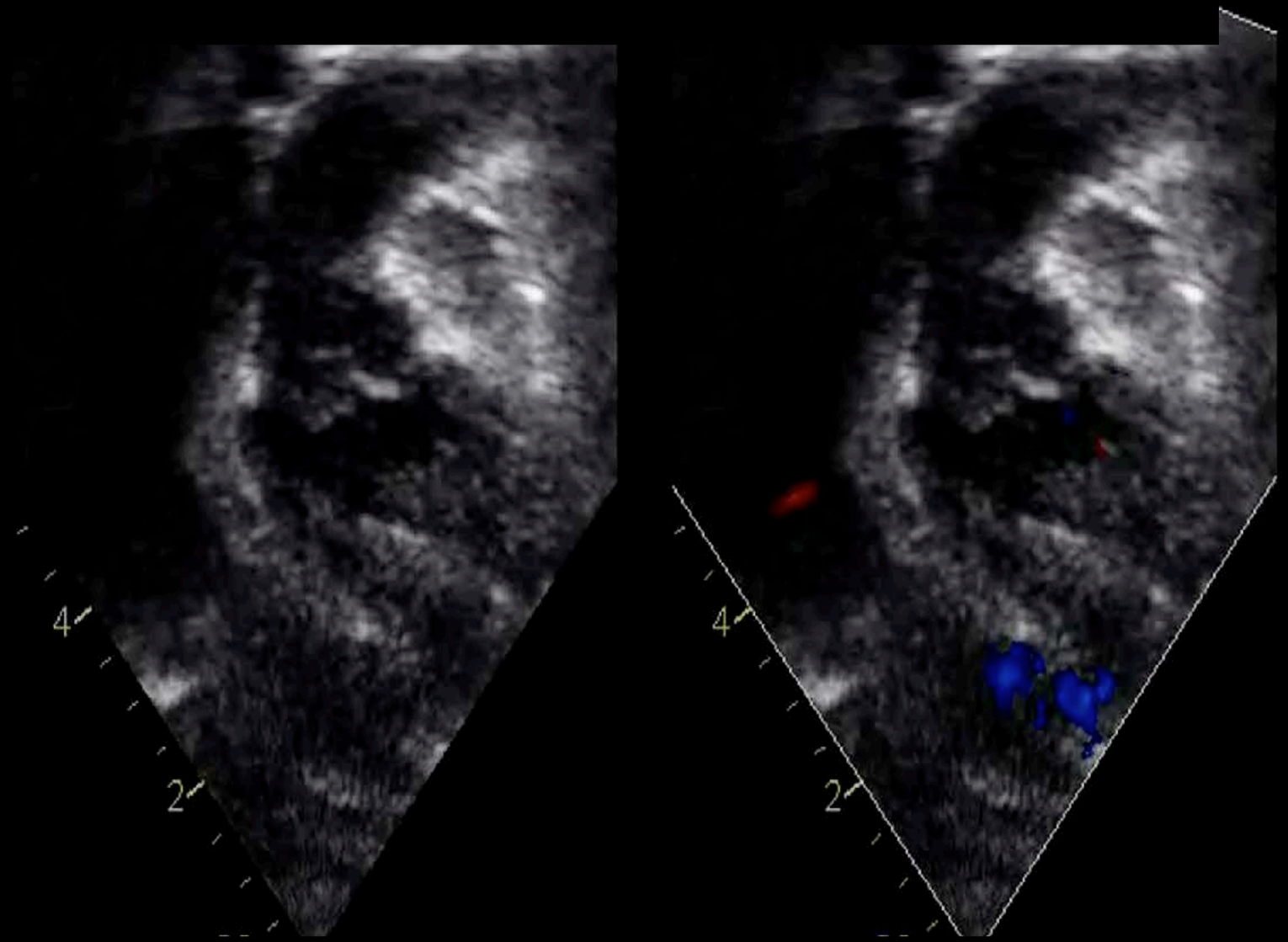
Straddling of tricuspid valve



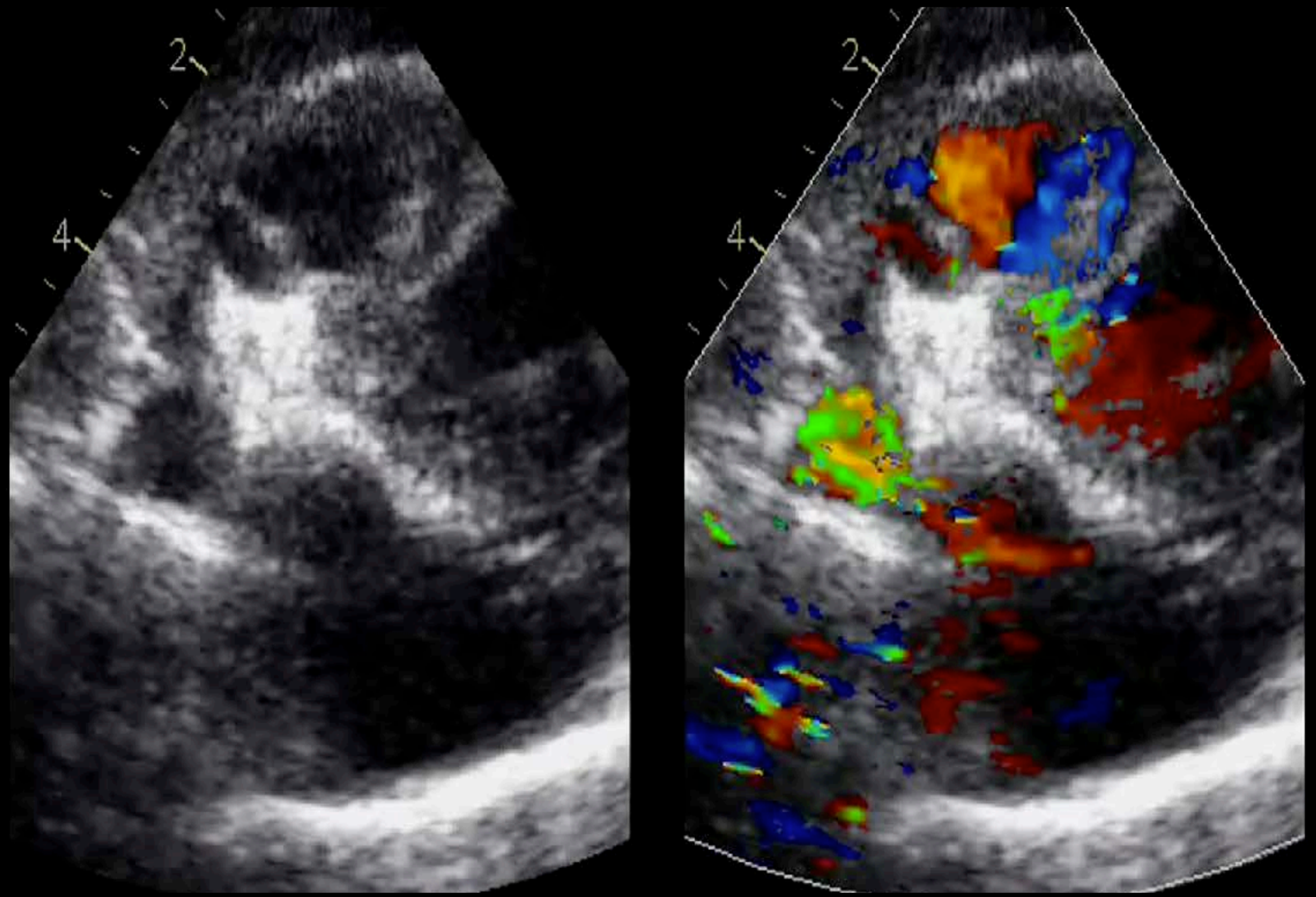
Outflow tracts in univentricular heart: accessory ventricle



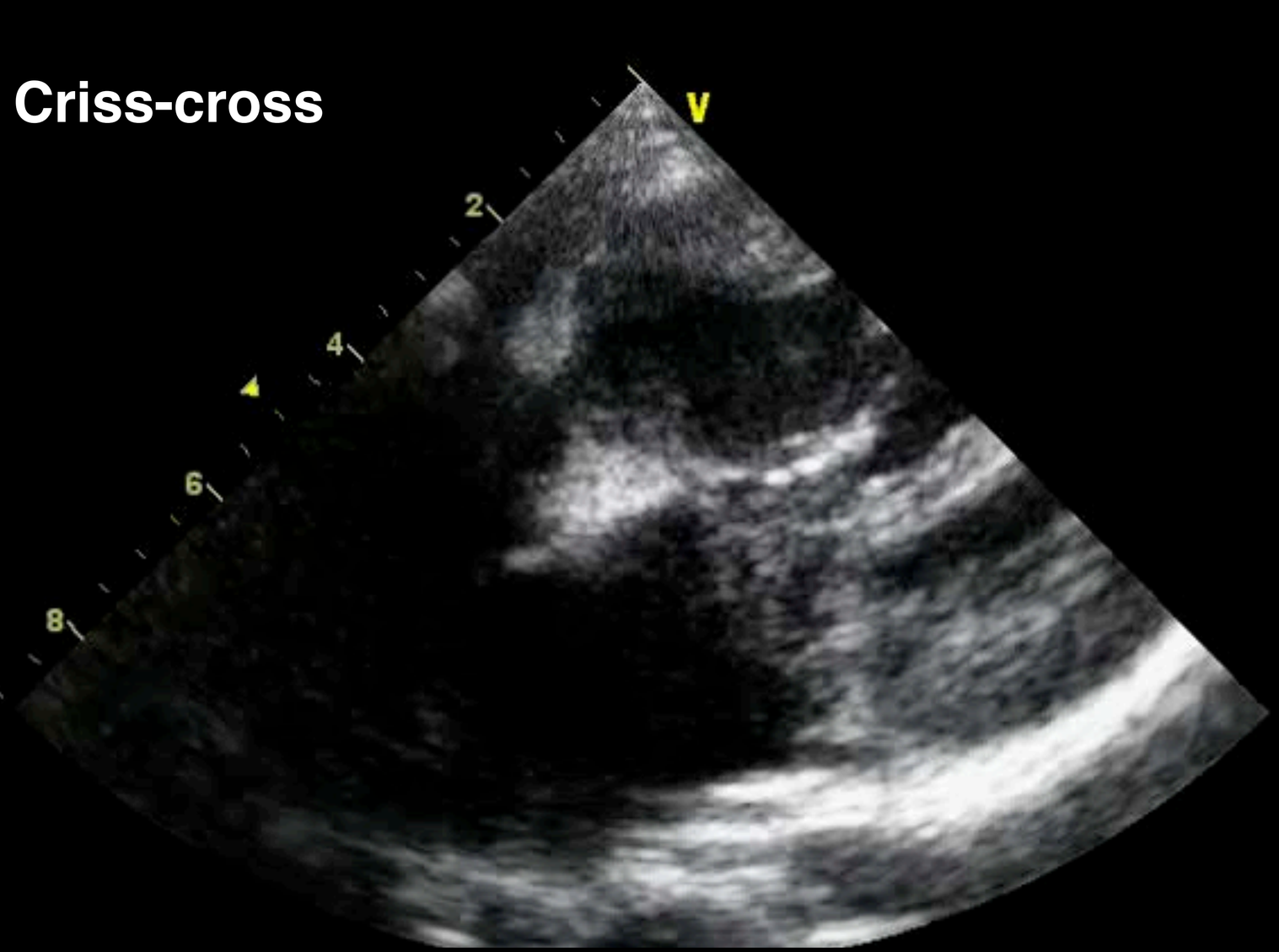
Outflow tracts in univentricular heart: accessory ventricle

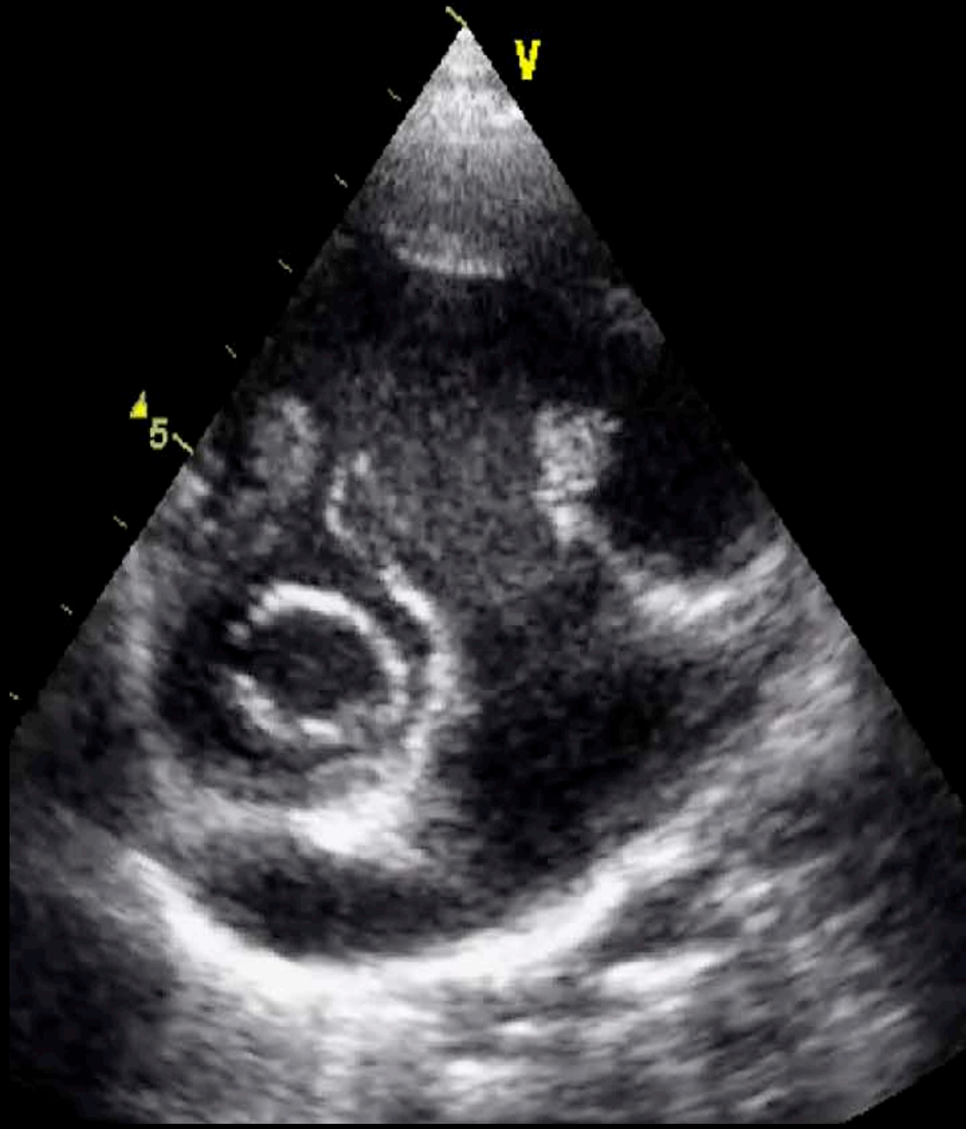


Outflow tracts in univentricular heart: accessory ventricle

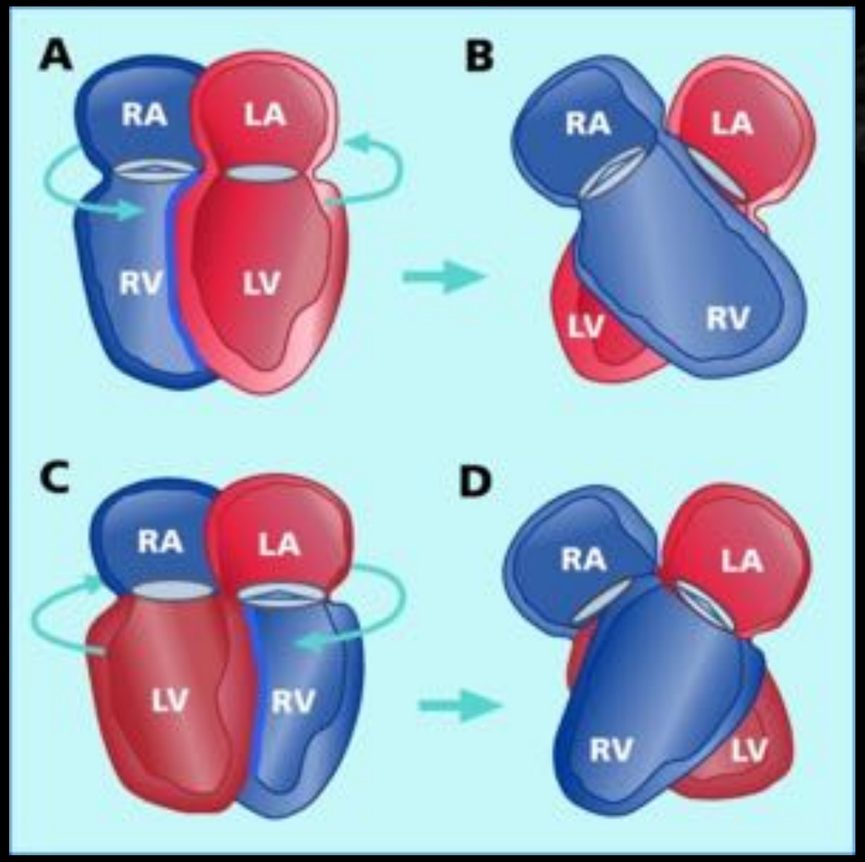


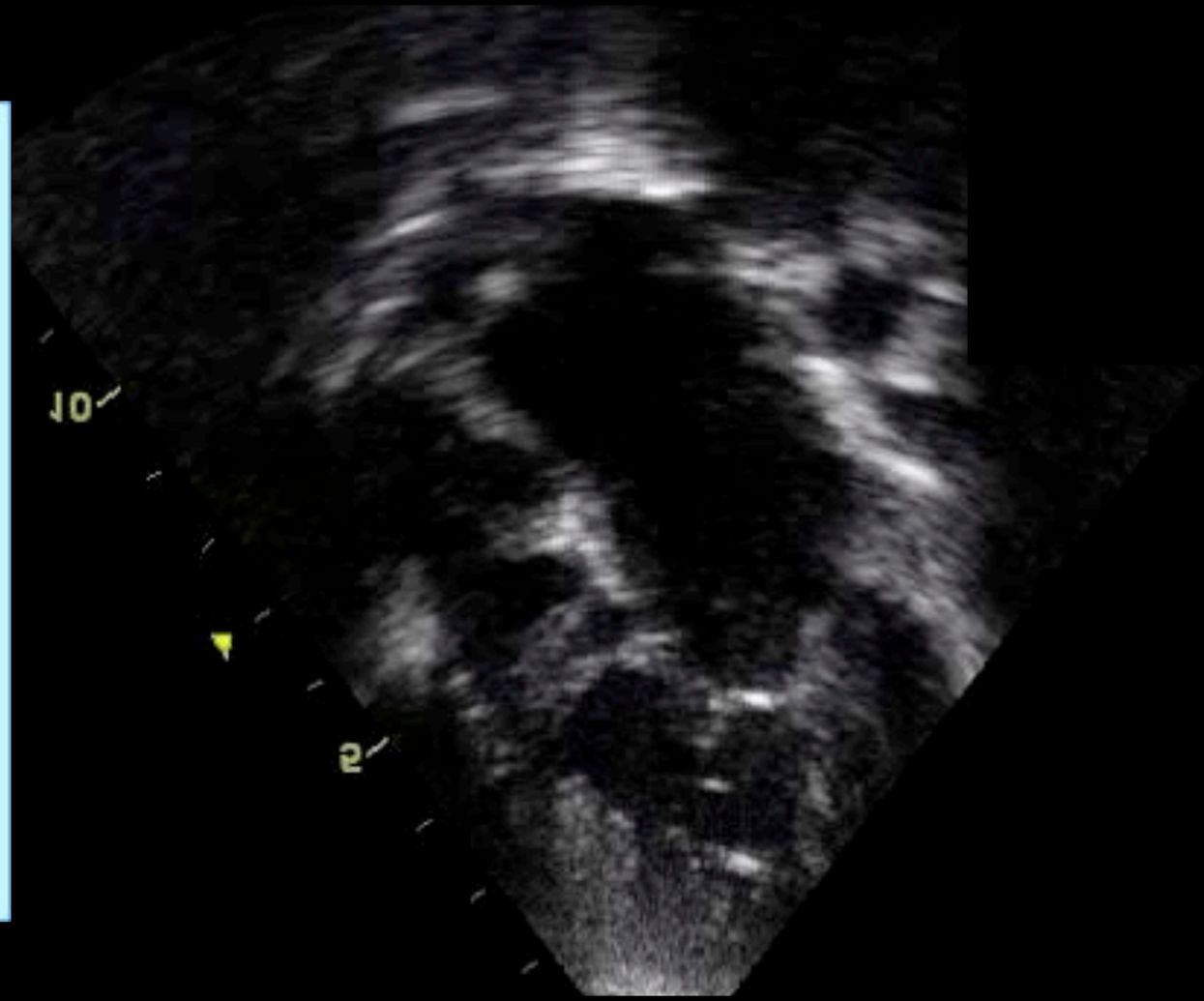
Criss-cross



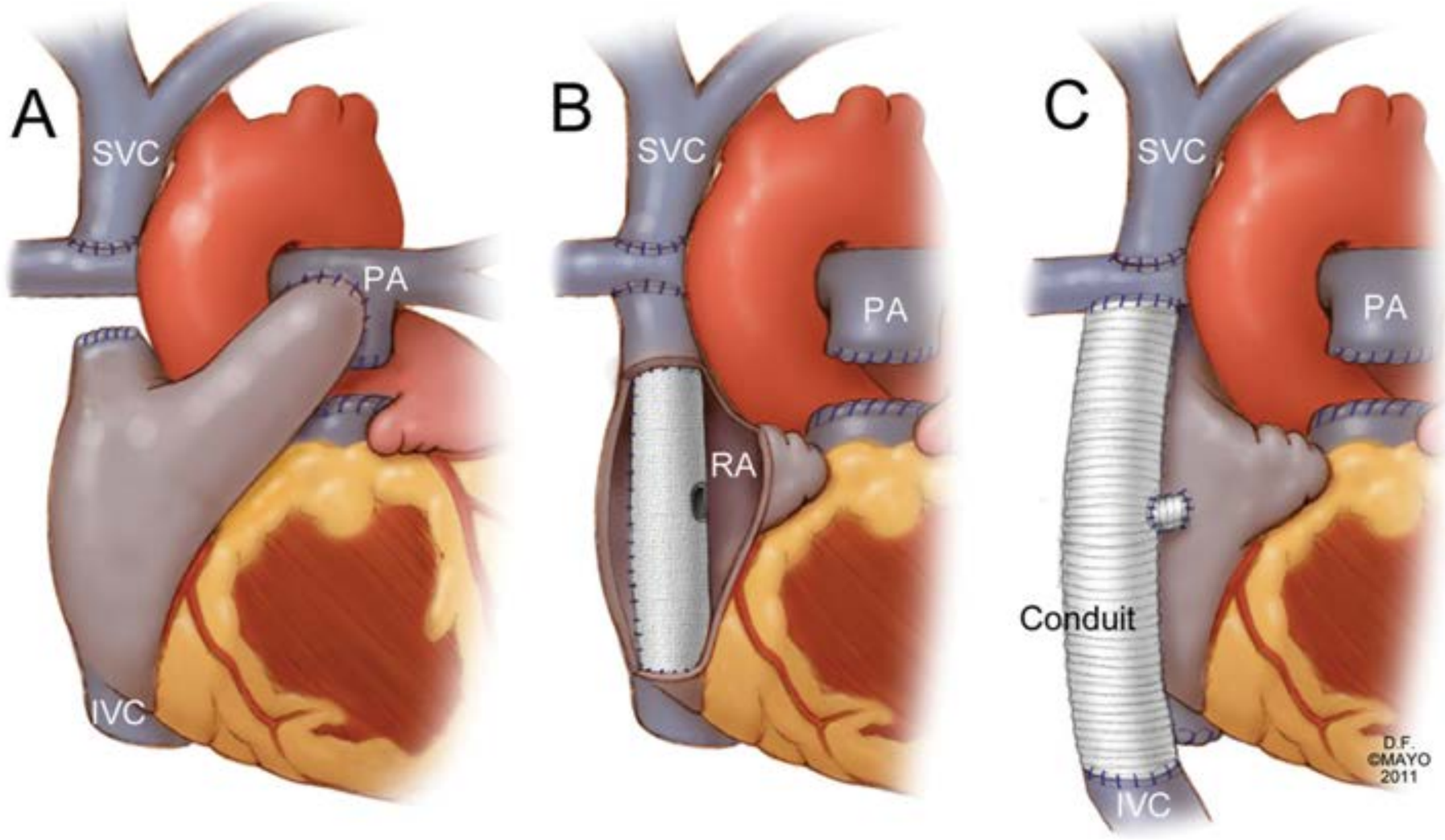


Criss-cross



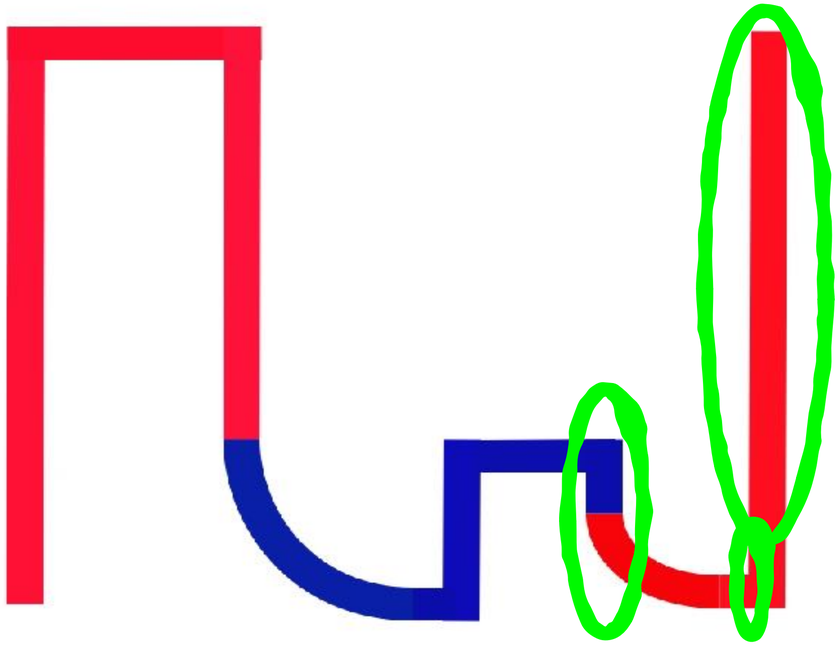


TCPC program



Critical bottlenecks in various circulations

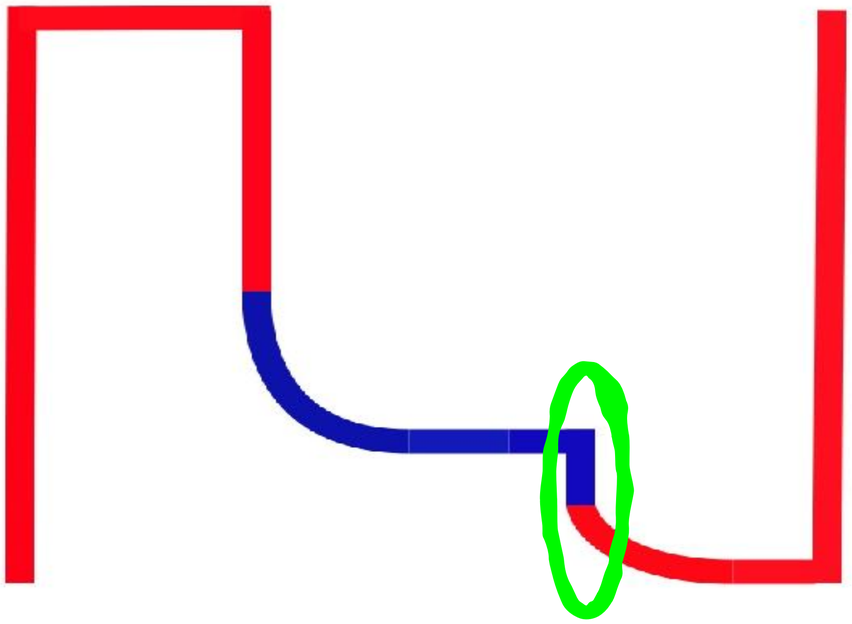
The Fontan circulation - a new portal system



Cardiomyopathies

Pulmonary hypertension

Mitral stenosis



Fontan TCPC

Courtesy Marc Gewillig

Quels sont les objectifs du traitement des VU ?

- Protéger la circulation pulmonaire
  - Cerclage
  - Lever les obstacles au retour veineux: CIA – RVPA bloqués
- Calibrer le débit pulmonaire
  - Blalock, conduit VU-AP
- Préserver la fonction du ventricule et des VAV
  - Réduire la post-charge
    - Coarctation, sténose sous aortique
  - Limiter la surcharge diastolique
    - Calibrer le débit pulmonaire
  - Essayer de limiter la durée de clampage aortique pendant la chirurgie
- Préserver les artéres pulmonaires et les veines systémiques
- Limiter les cicatrices atriales et ventriculaires

Le décalogue : 10 commandements (Choussat-Fontan)

1. situs solitus
2. connection veineuse, systémique et pulmonaire, normale
3. hypertrophie oreillette droite
4. fonction ventriculaire normale
5. fonction normale des valves auriculo-ventriculaires
6. rythme sinusal
7. artères pulmonaires de taille normale
8. pression pulmonaire basse (< 15 mmHg)
9. résistances vasculaires pulmonaires basses
10. âge > 4 ans

Critères actuels

- "bon" lit vasculaire pulmonaire
    . absence d'hypoplasie diffuse
    . absence de sténose localisée (native ou iatrogène)
    . résistances vasculaires pulmonaires basses
    . gradient transpulmonaire bas

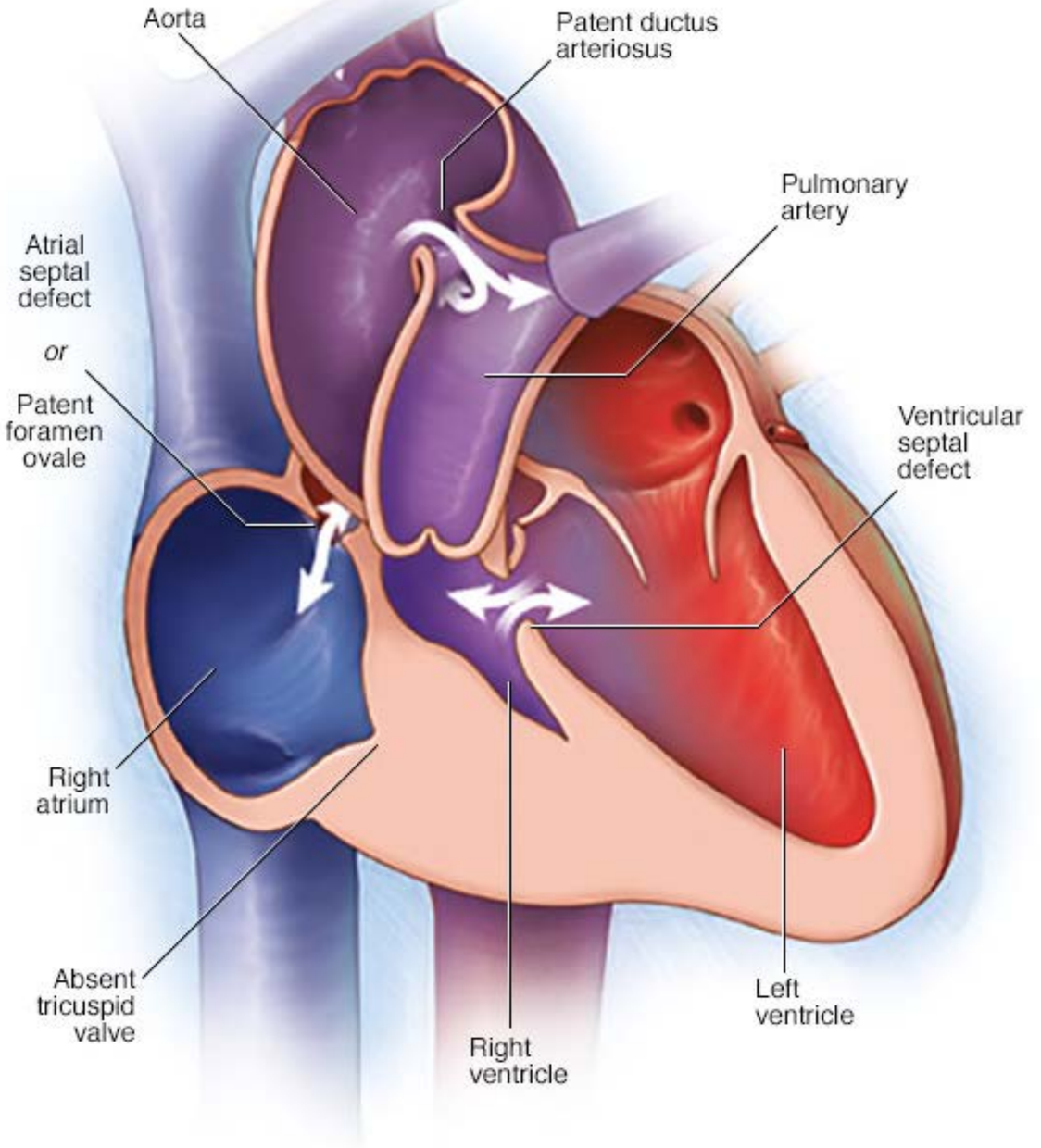
- "bon" ventricule
    . fonction normale (systolique and diastolique)
    . valve AV normale (au moins une)
        . voie de sortie sans obstacle
        . rythme sinusal stable
    . pression auriculaire gauche basse

Good and evil

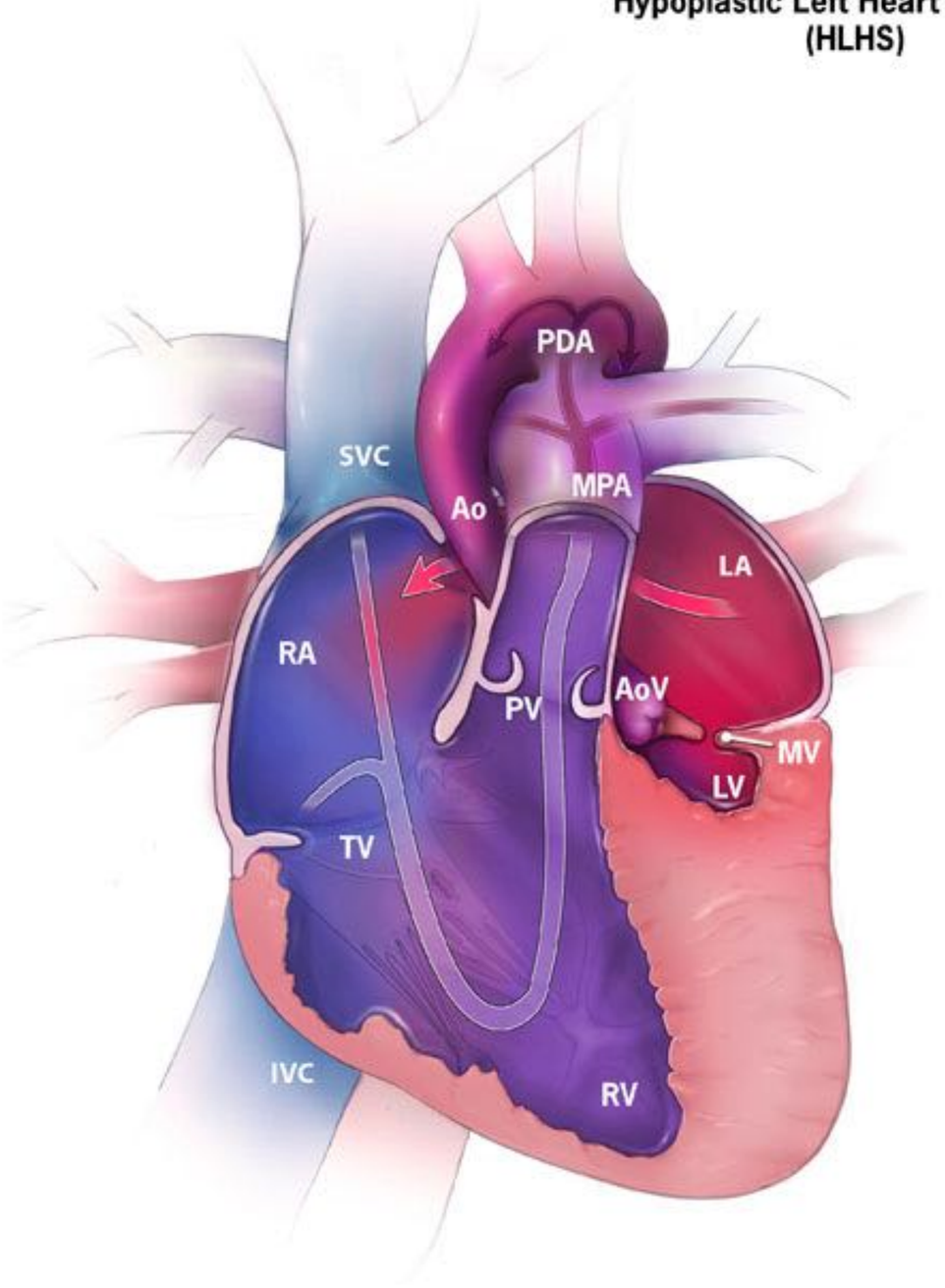
- VG versus VD versus Vx
- Mitrale vs tricuspide vs CAV
- Obstacle aortique
- Absence de protection pulmonaire
- Anatomie artérielle pulmonaire
- RVPAT ou PFO restrictif (atrésie mitrale)

Situations néonatales

- Ducto-dépendance pour la circulation pulmonaire
  - PGE1 puis Blalock
- Ducto-dépendance pour la circulation systémique
  - Norwood
- Hypertension pulmonaire
  - Cerclage


Aorta
Patent ductus arteriosus
Pulmonary artery
Atrial septal defect
or
Patent foramen ovale
Ventricular septal defect
Right atrium
Absent tricuspid valve
Right ventricle
Left ventricle

Hypoplastic Left Heart Syndrome (HLHS)

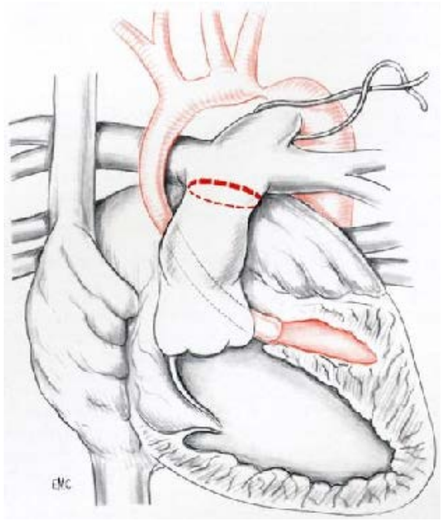


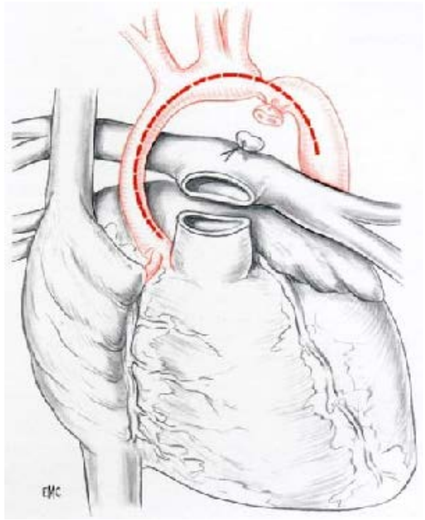
RA. Right Atrium
RV. Right Ventricle
LA. Left Atrium
LV. Left Ventricle

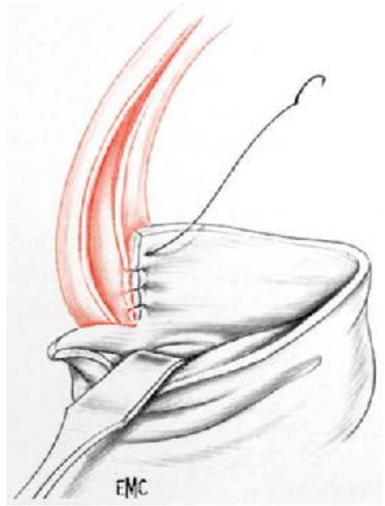
SVC. Superior Vena Cava
IVC. Inferior Vena Cava
MPA. Main Pulmonary Artery
Ao. Aorta
PDA. Patent Ductus Arteriosis

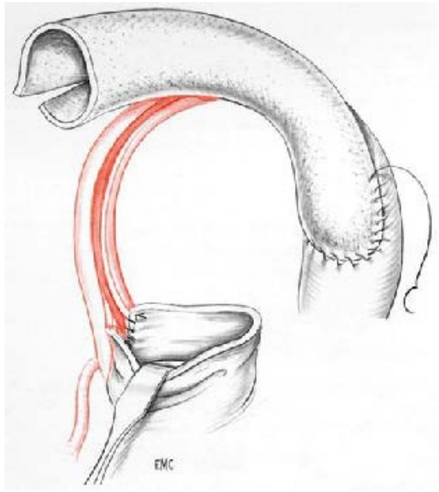
TV. Tricuspid Valve
MV. Mitral Valve
PV. Pulmonary Valve
AoV. Aortic Valve

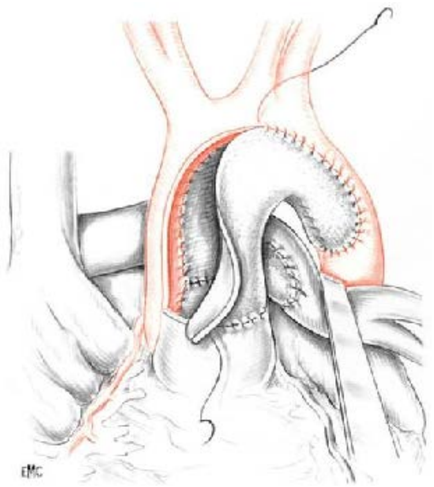
Norwood procedure

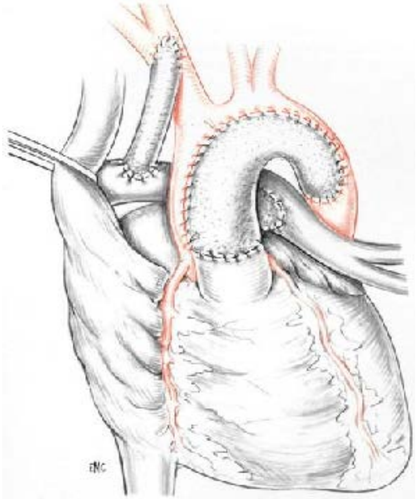




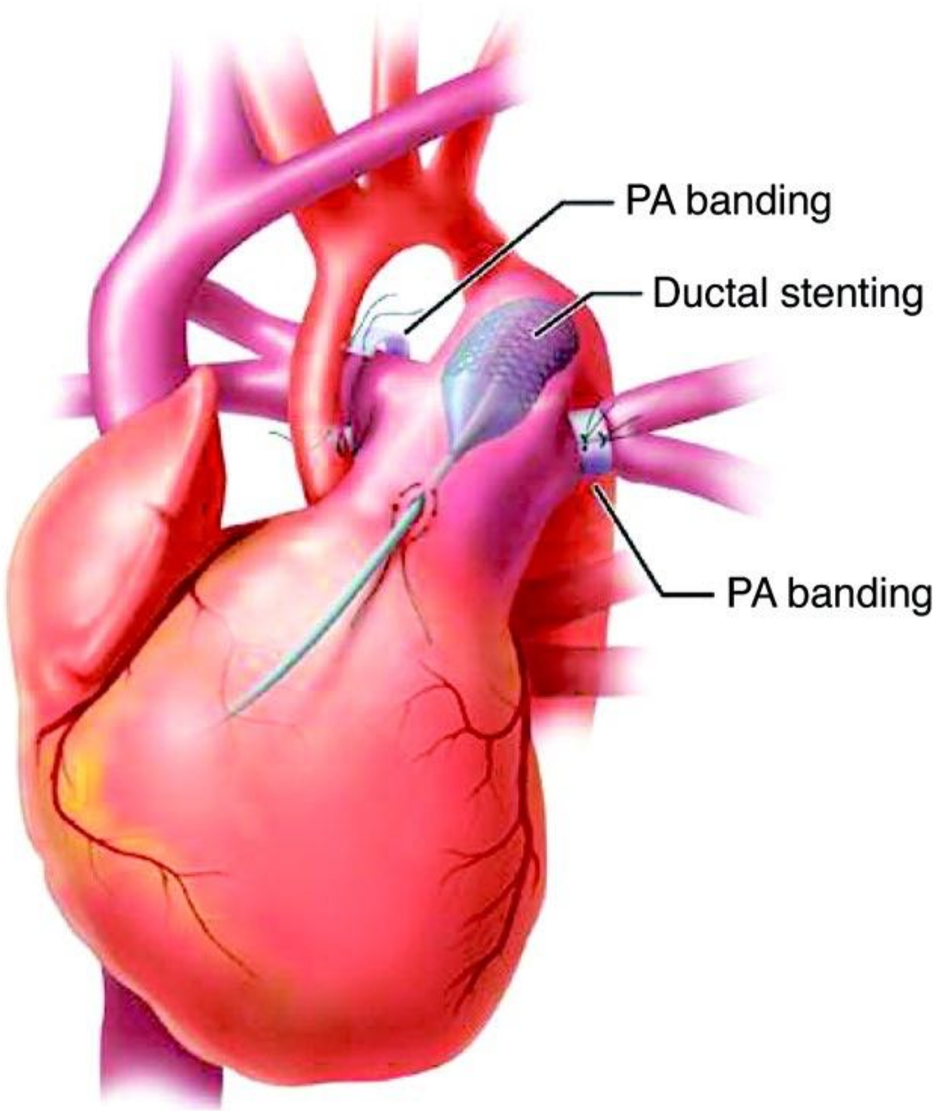


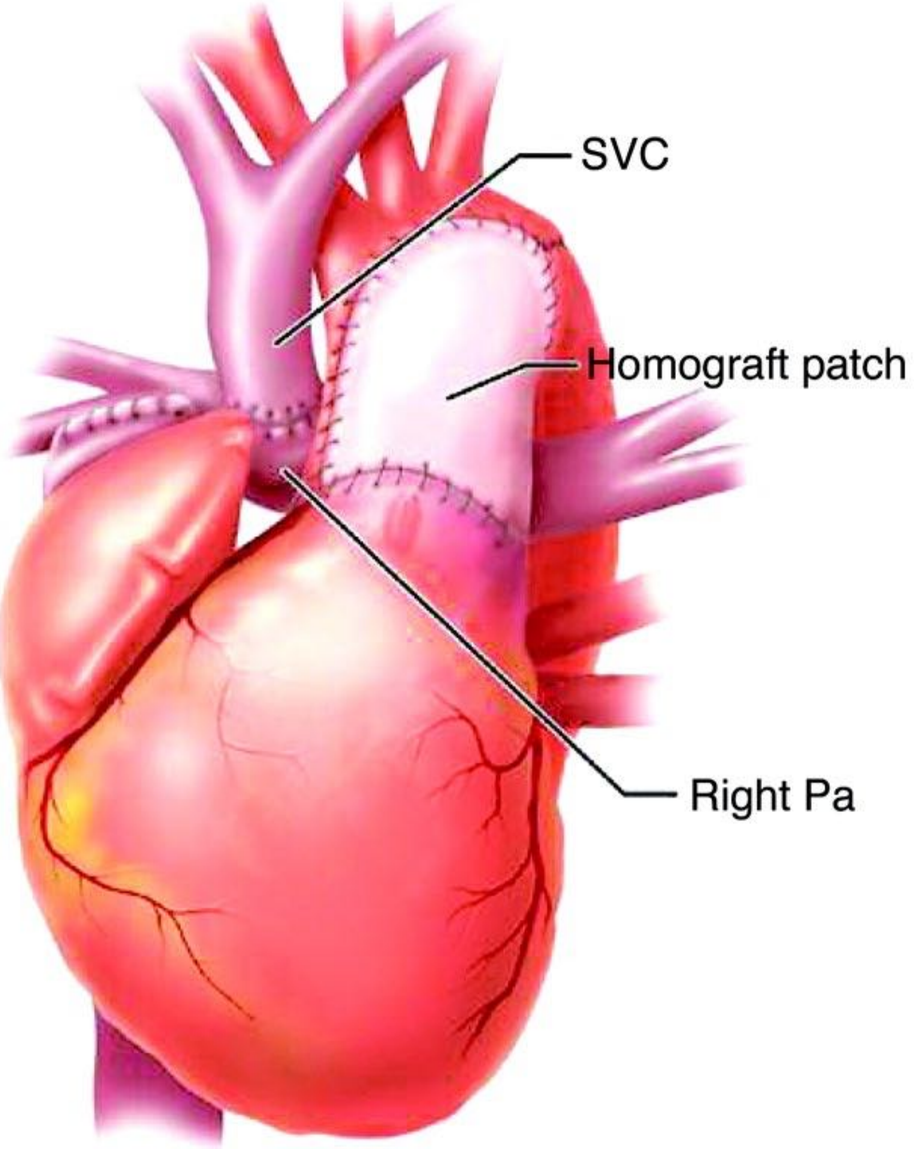






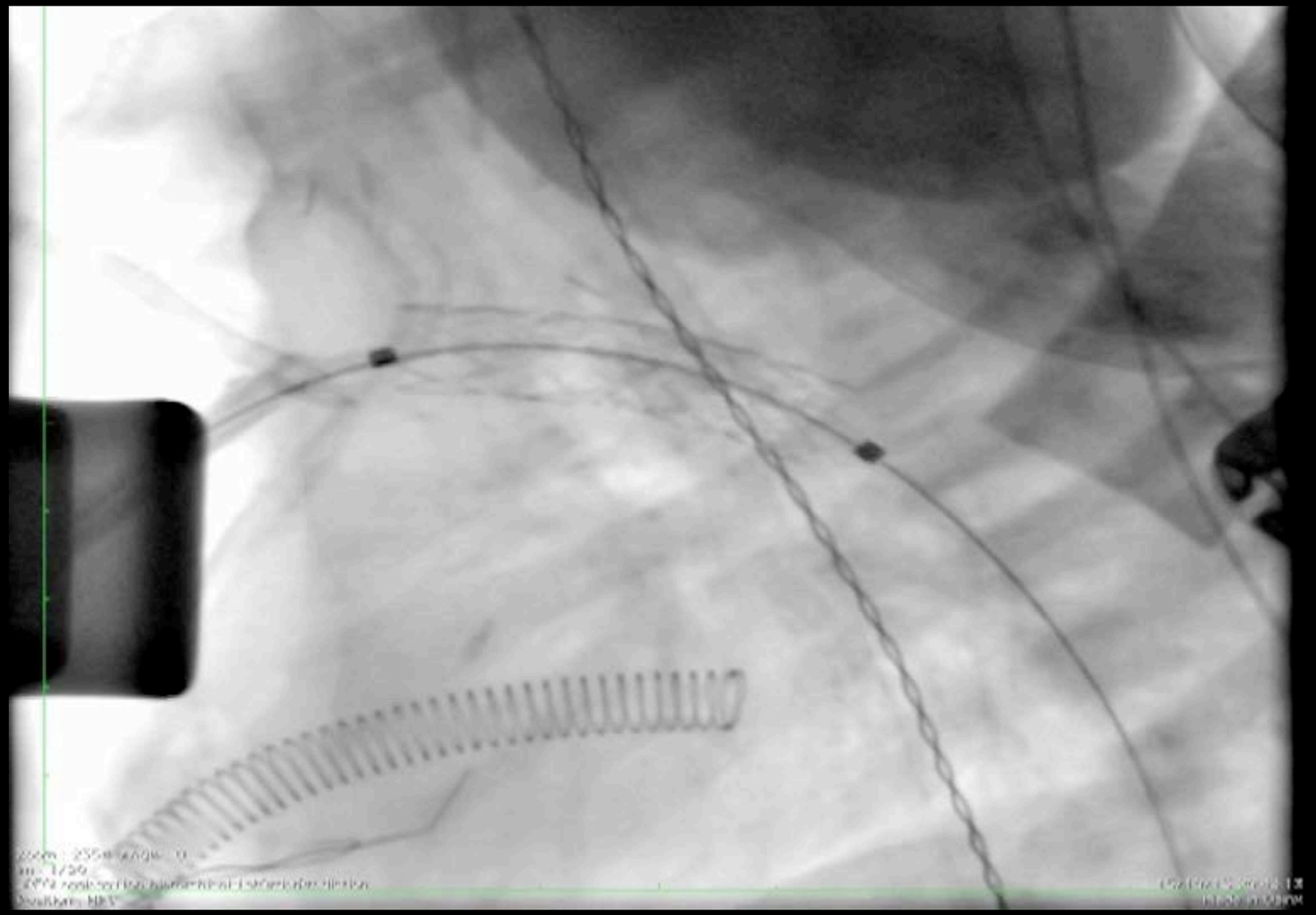
Percutaneous procedures in UVH





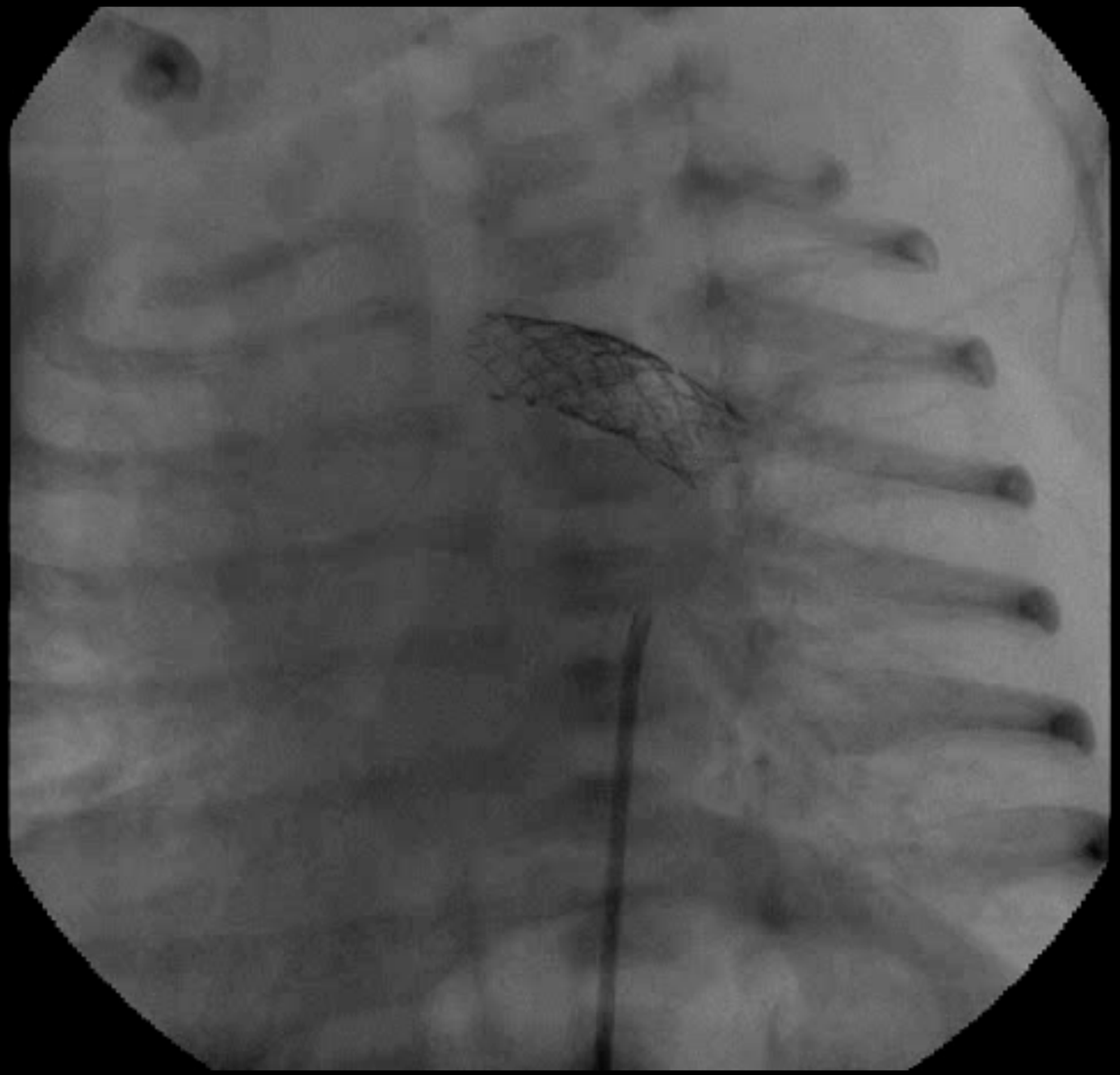
Hypoplasie du cœur gauche

1er temps de l’opération de Norwood-Hybride

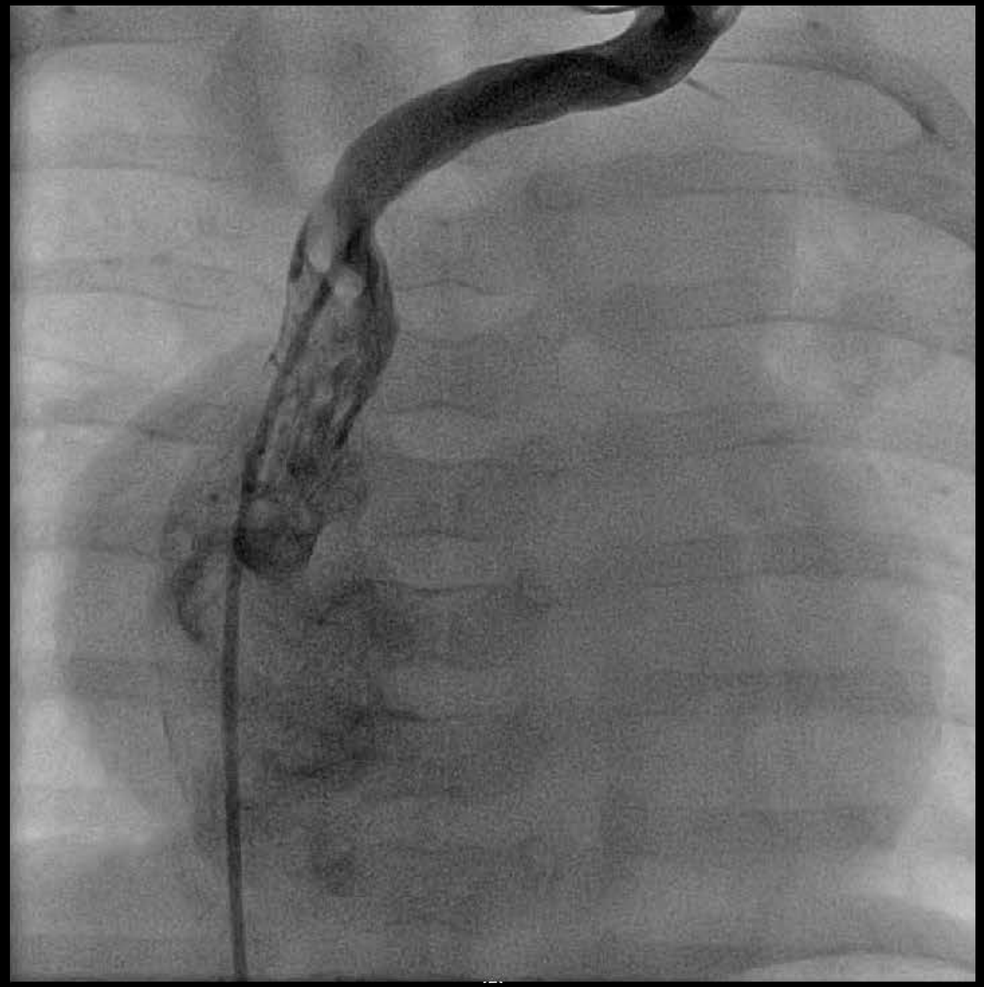


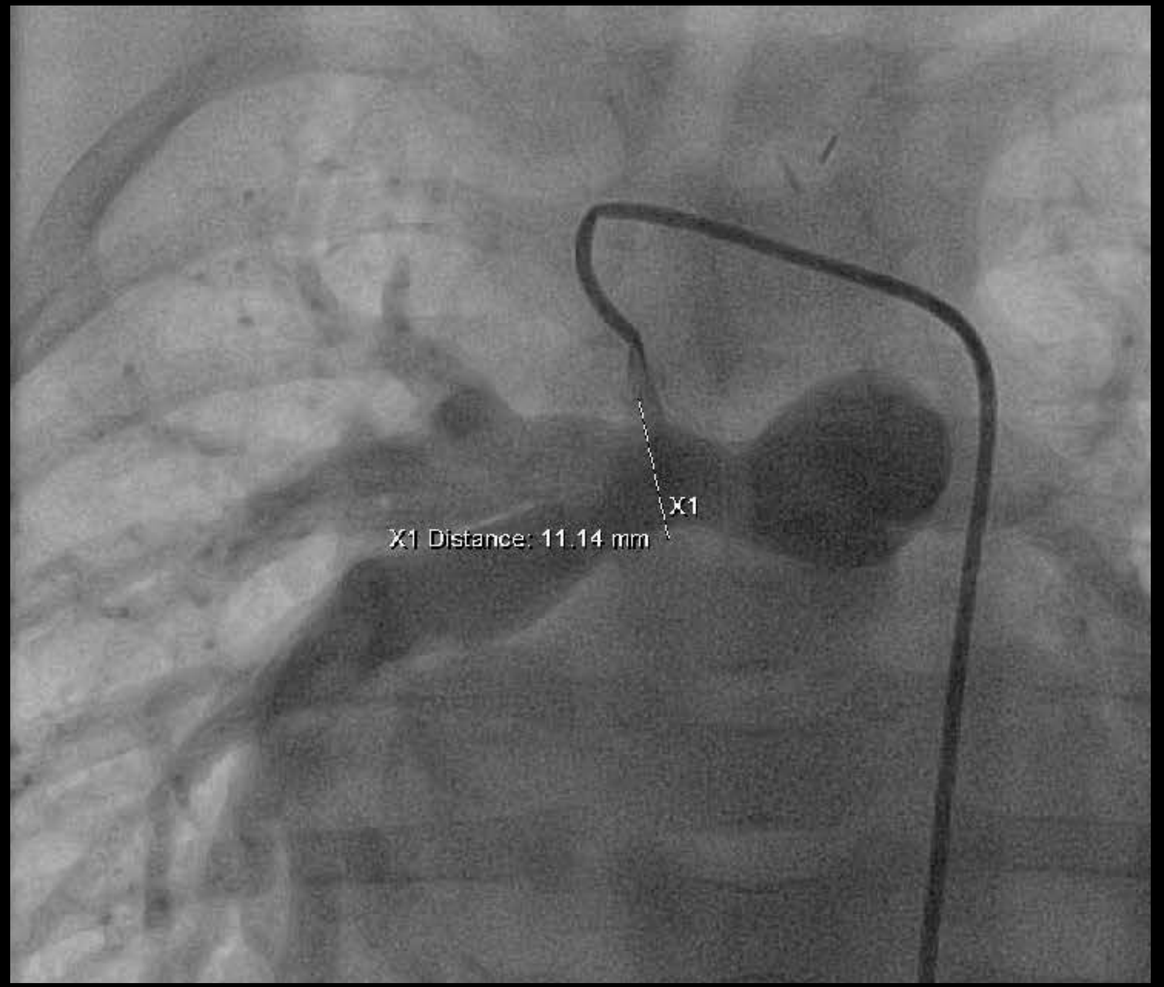
Hypoplasie du cœur gauche

1er temps de l’opération de Norwood-Hybride

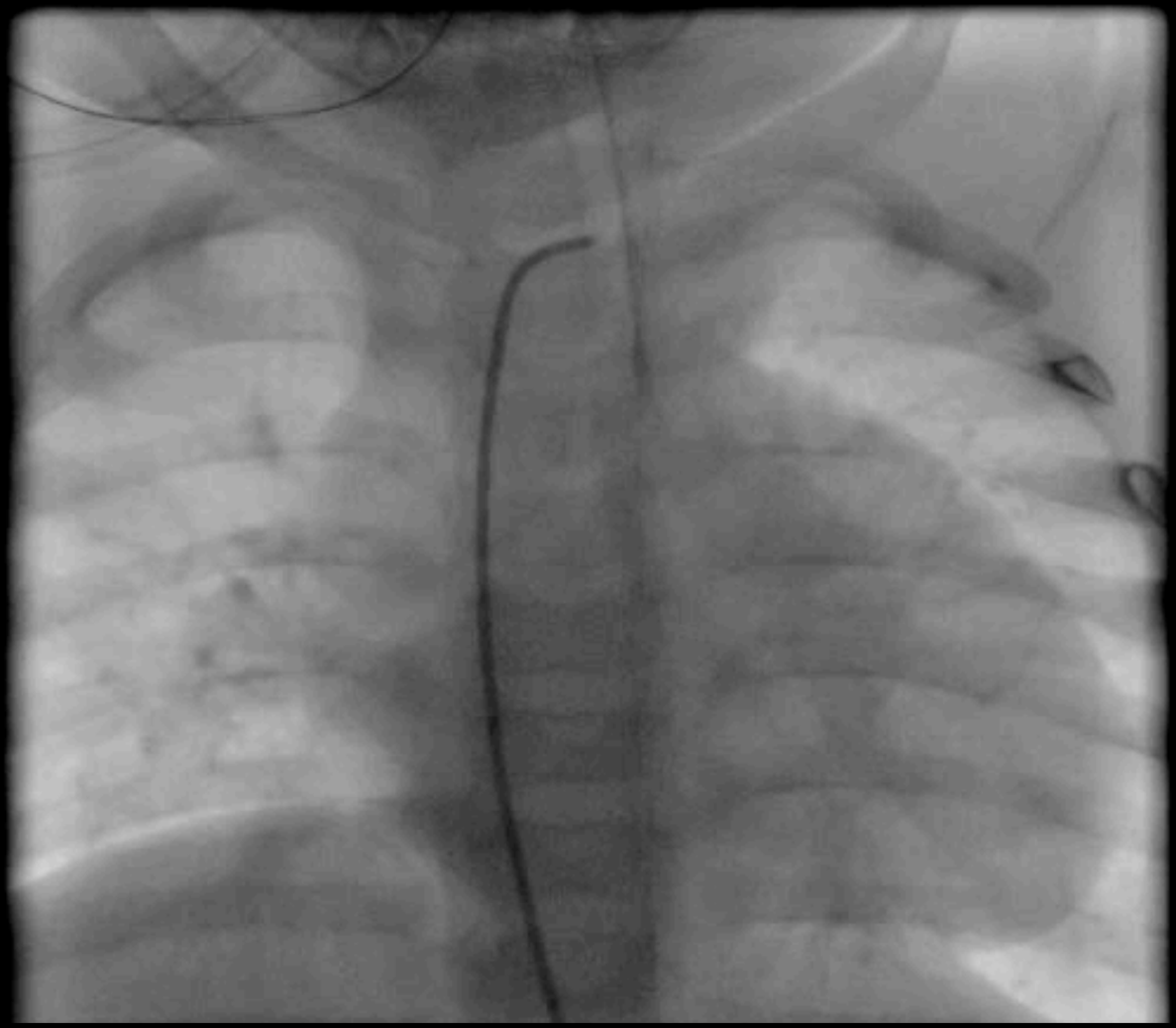


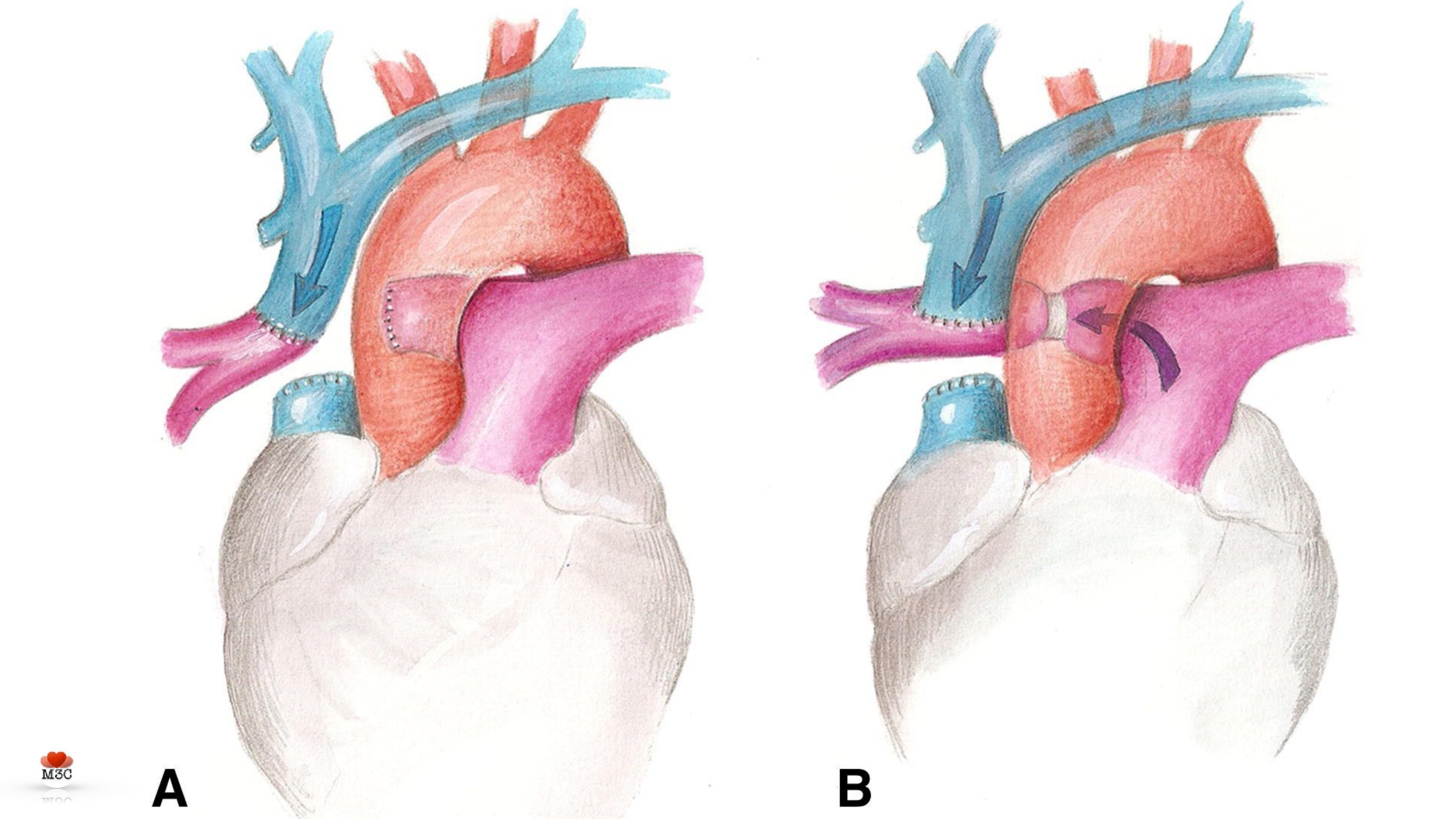
Evaluation pré DCPP





Evaluation pré DCPP

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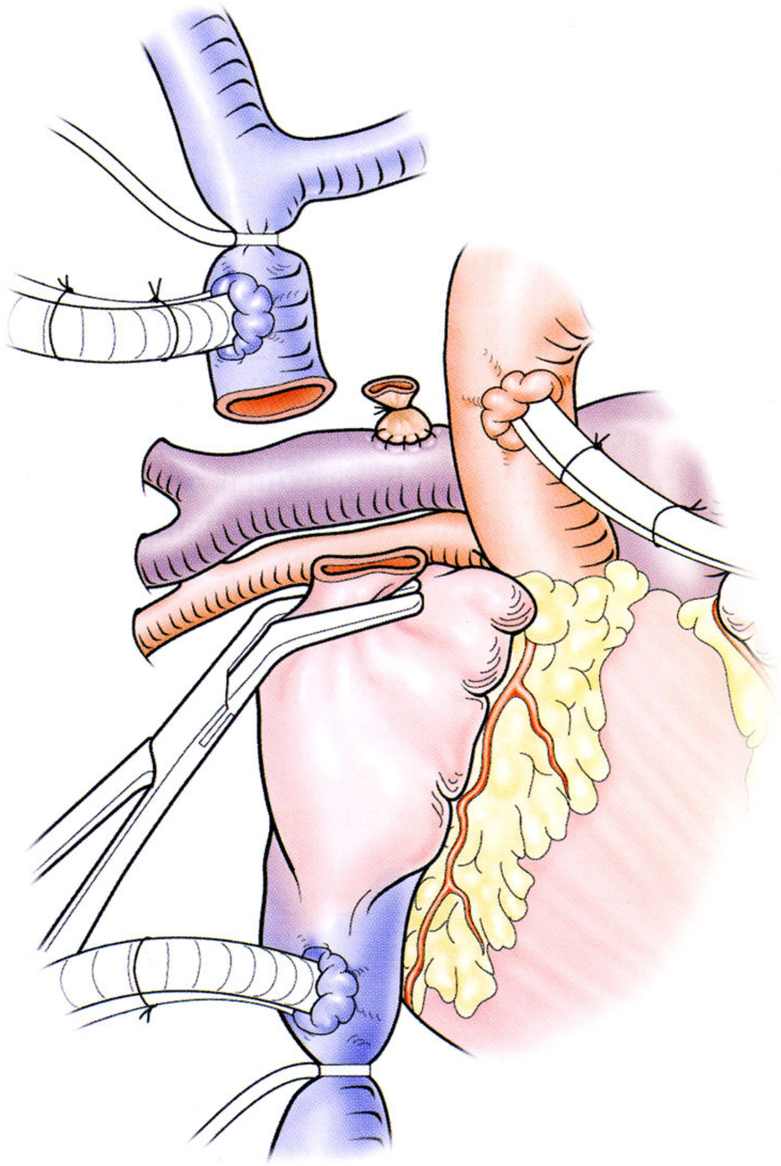
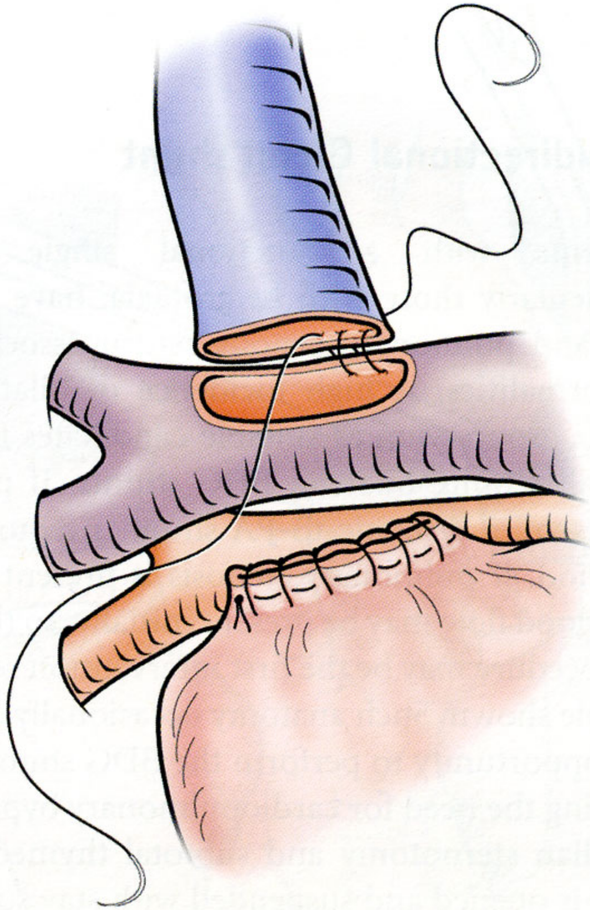
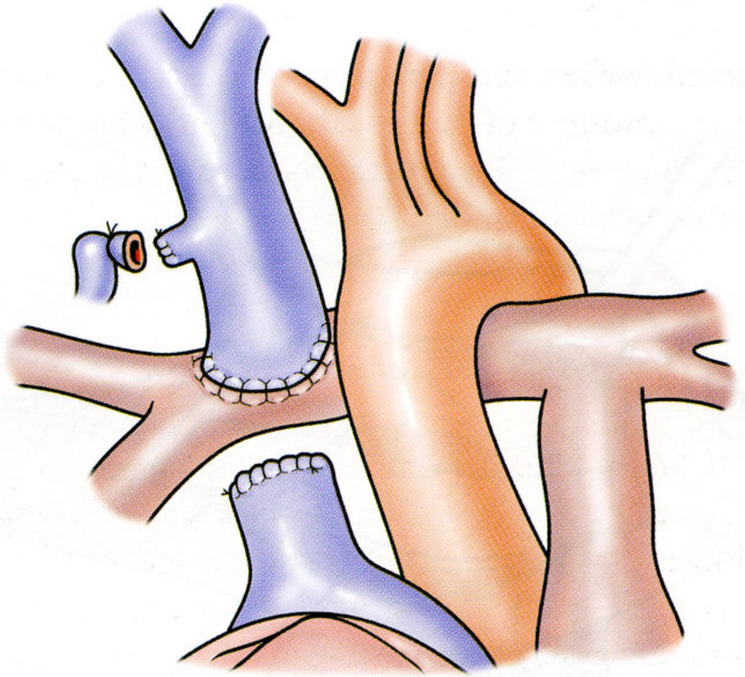
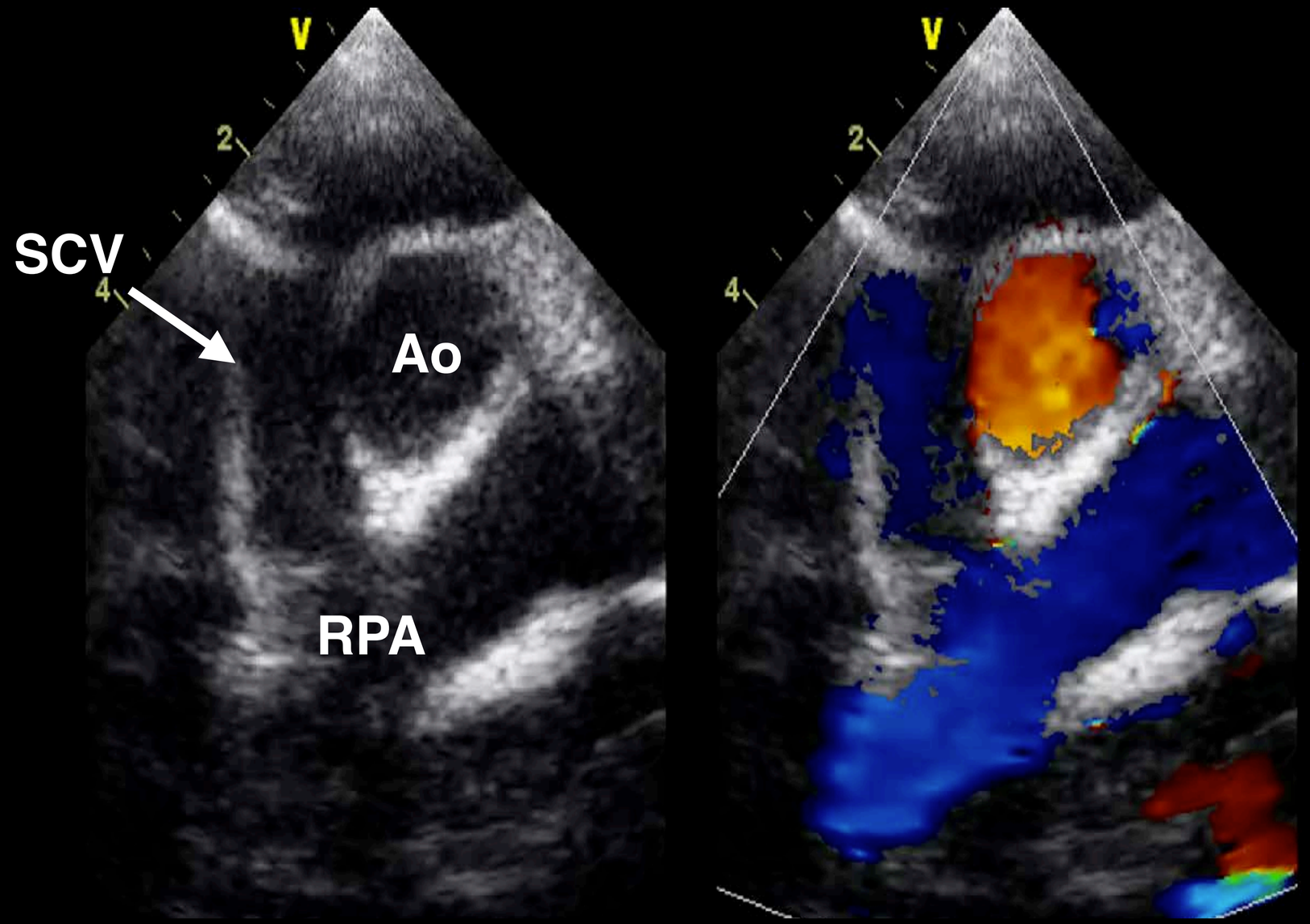
B





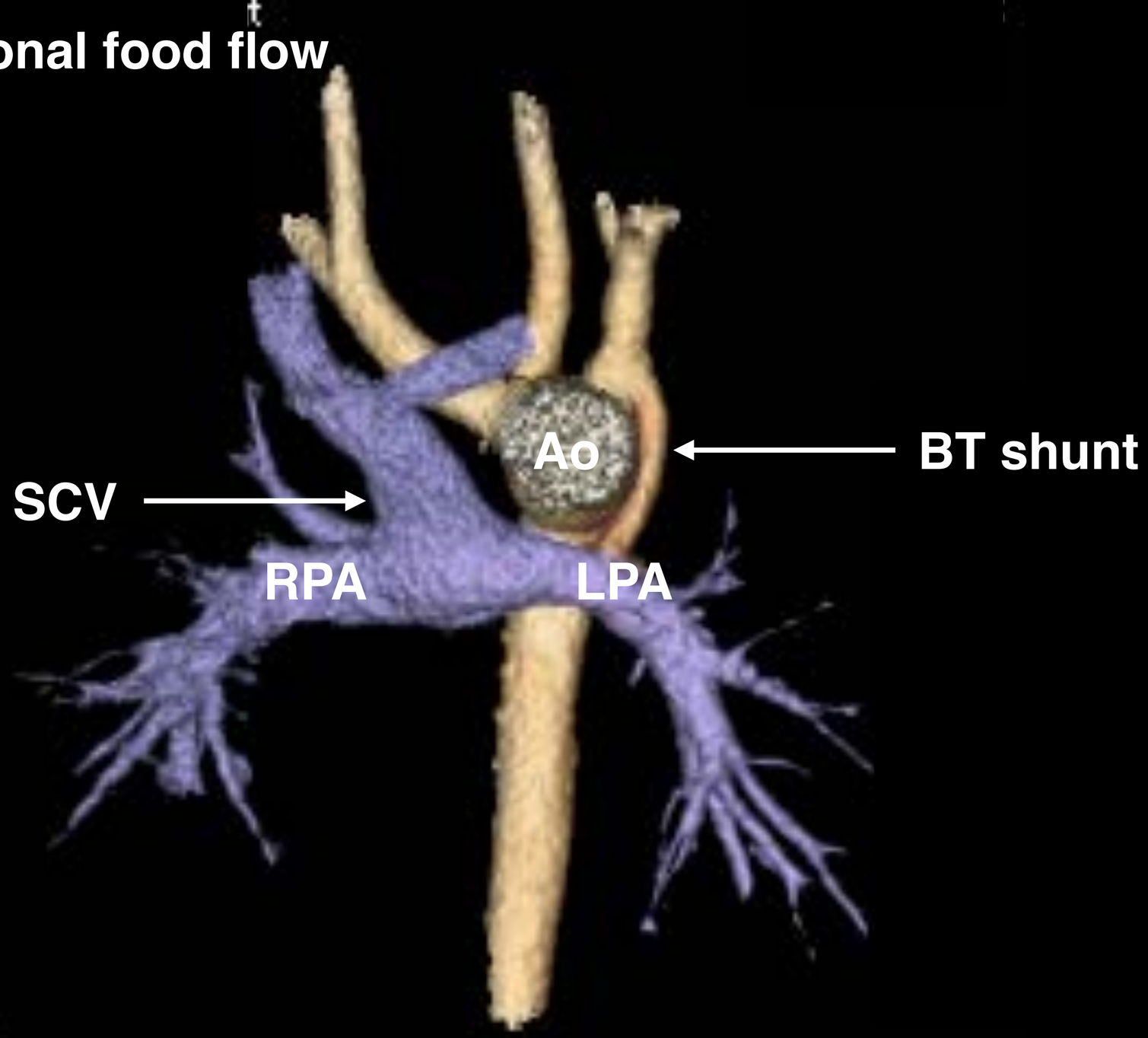


Partial cavopulmonary connection



Partial cavopulmonary connection

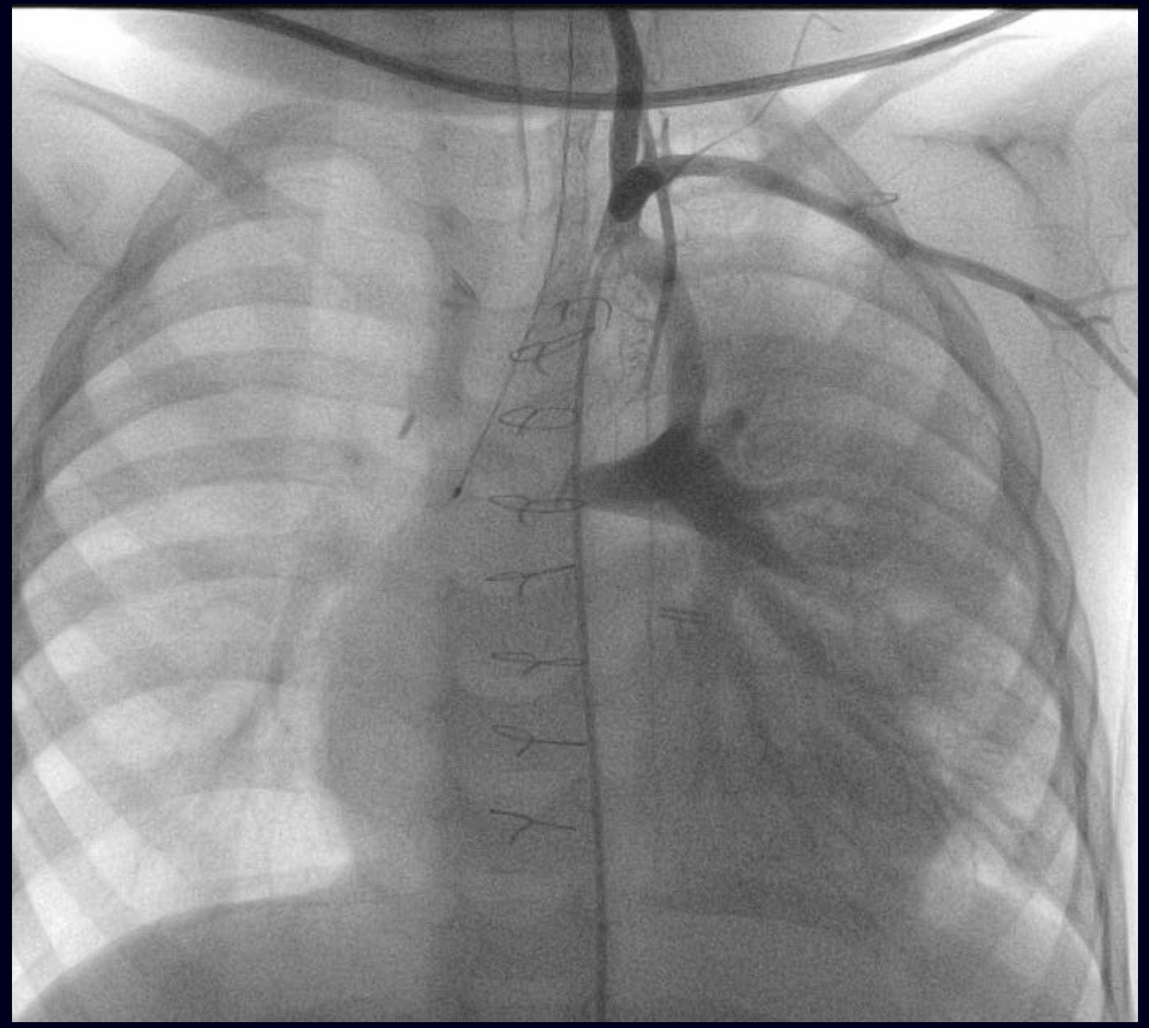
Partial CPC with additional food flow



HLHS, 3 years pre-Fontan

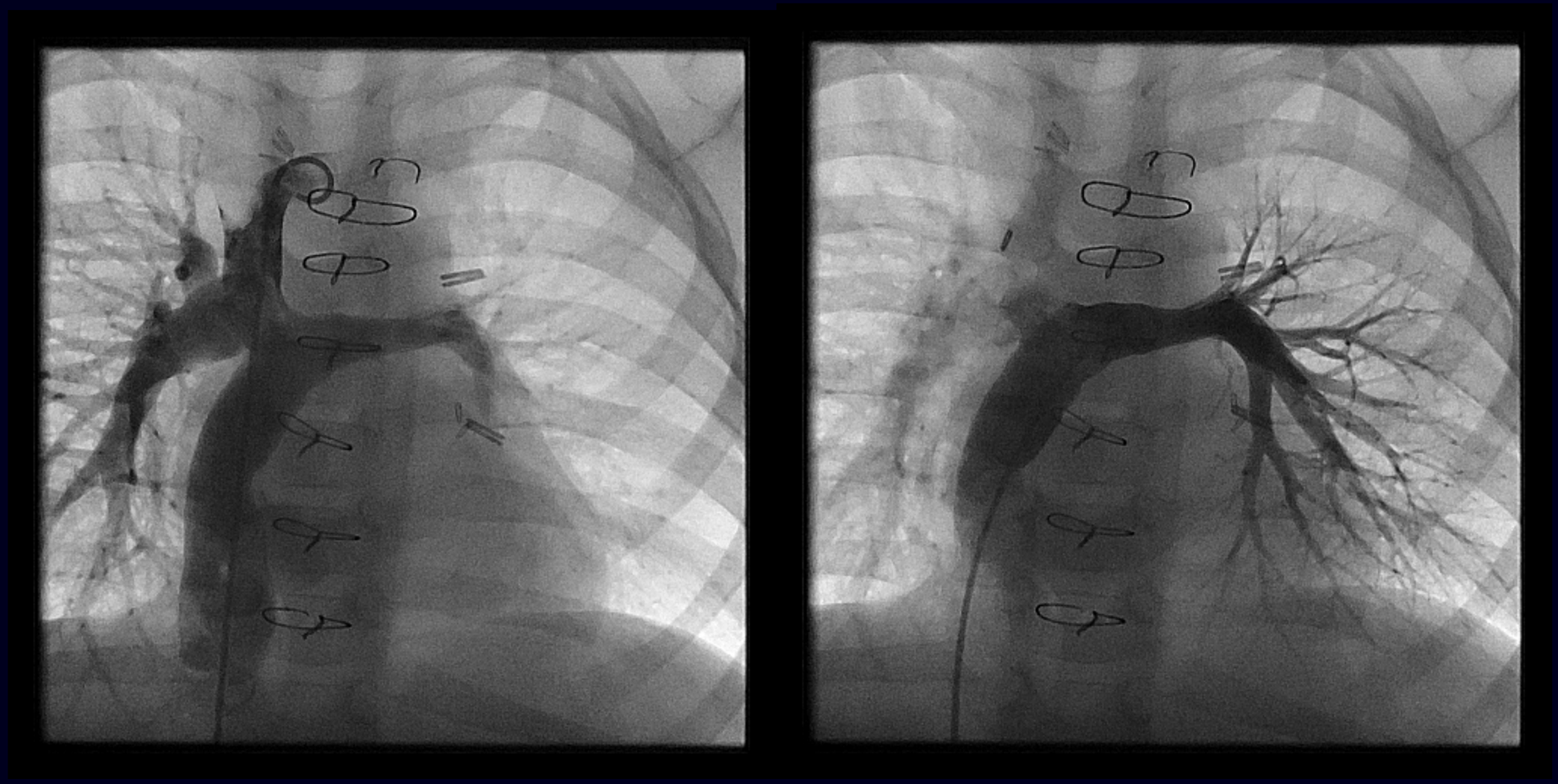
Status PO Norwood 3.0 central shunt; PO Glenn

HLHS, 3 years pre-Fontan



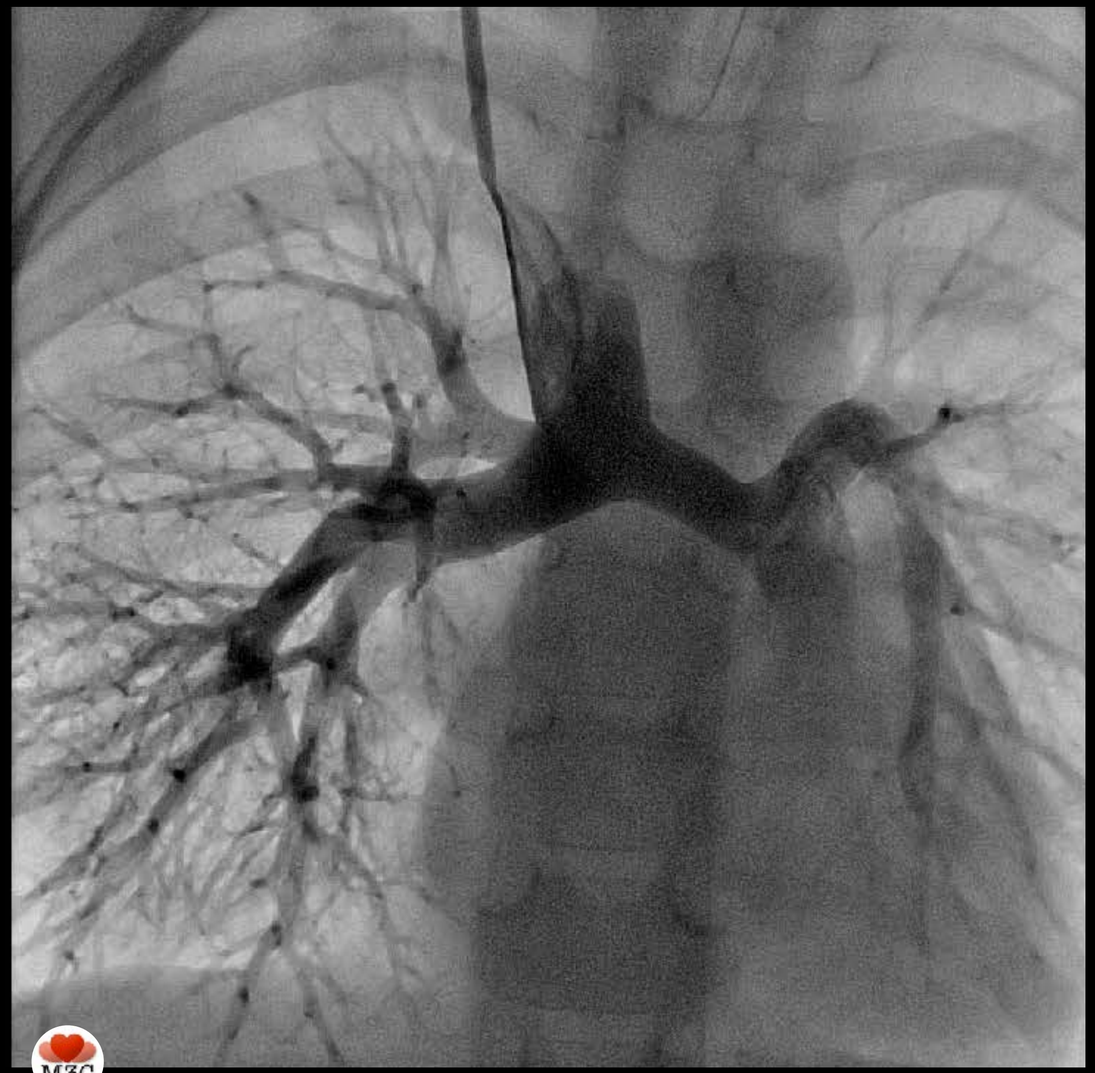
6 mm GT mBT shunt , clip on central PA

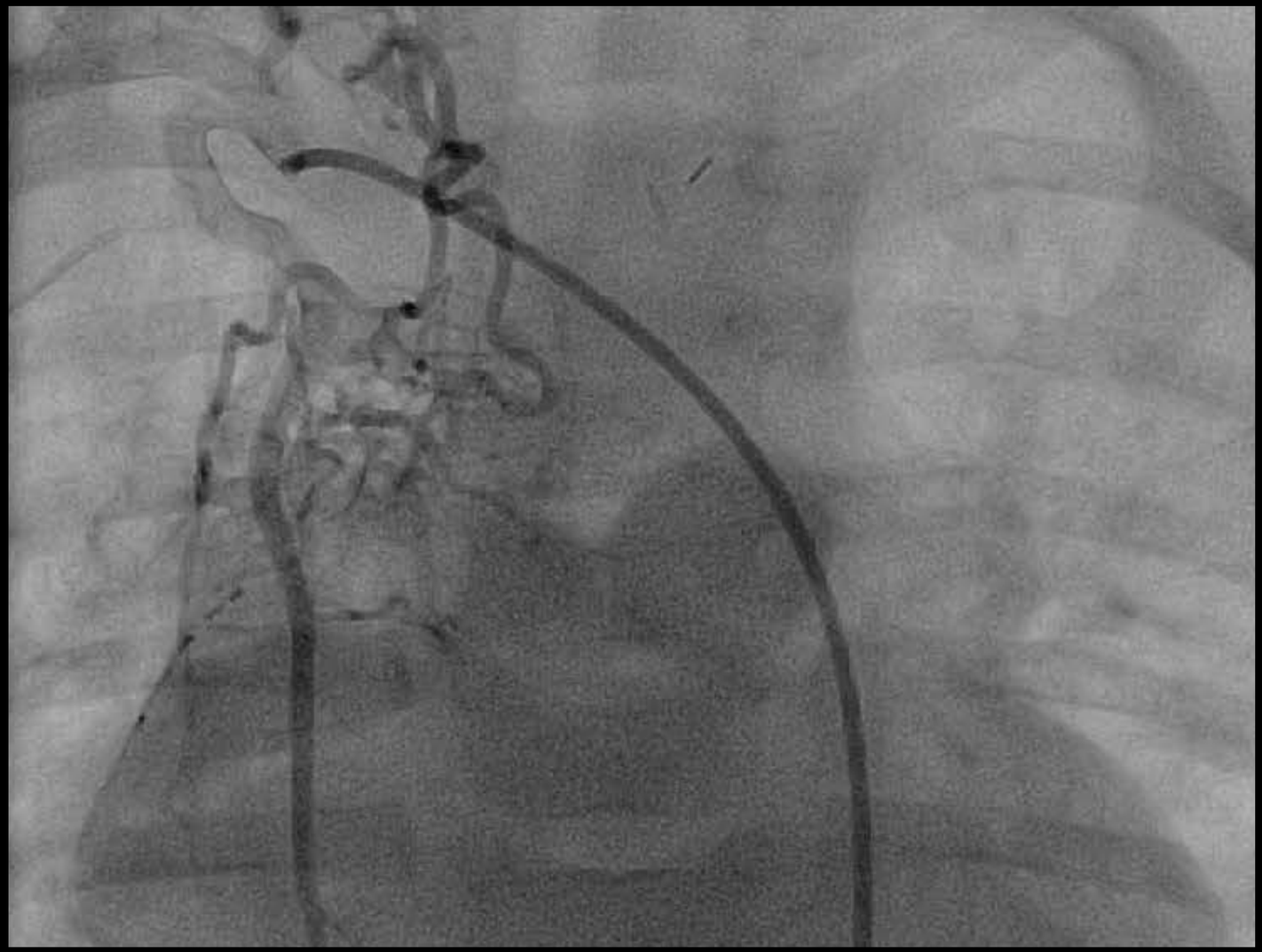
HLHS, 1 year post-Fontan



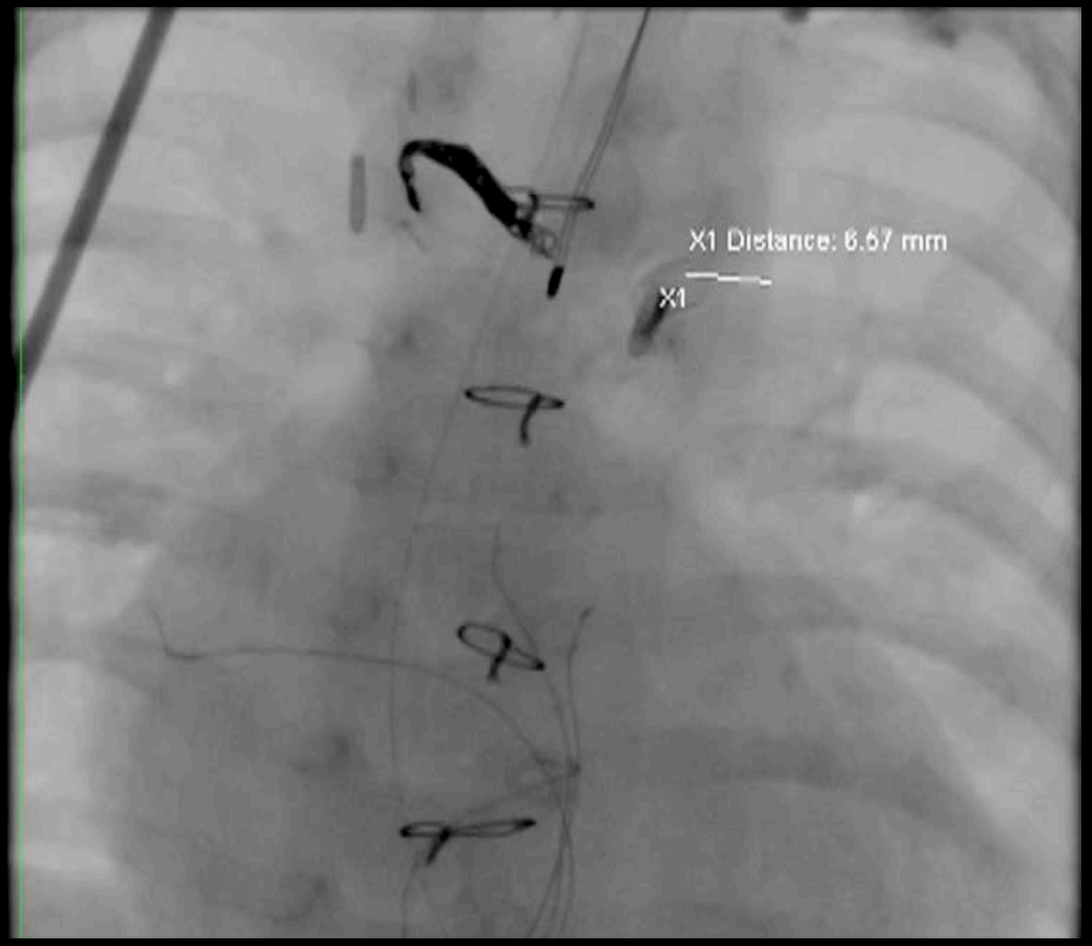
Le PA reconstructed with 12 GT

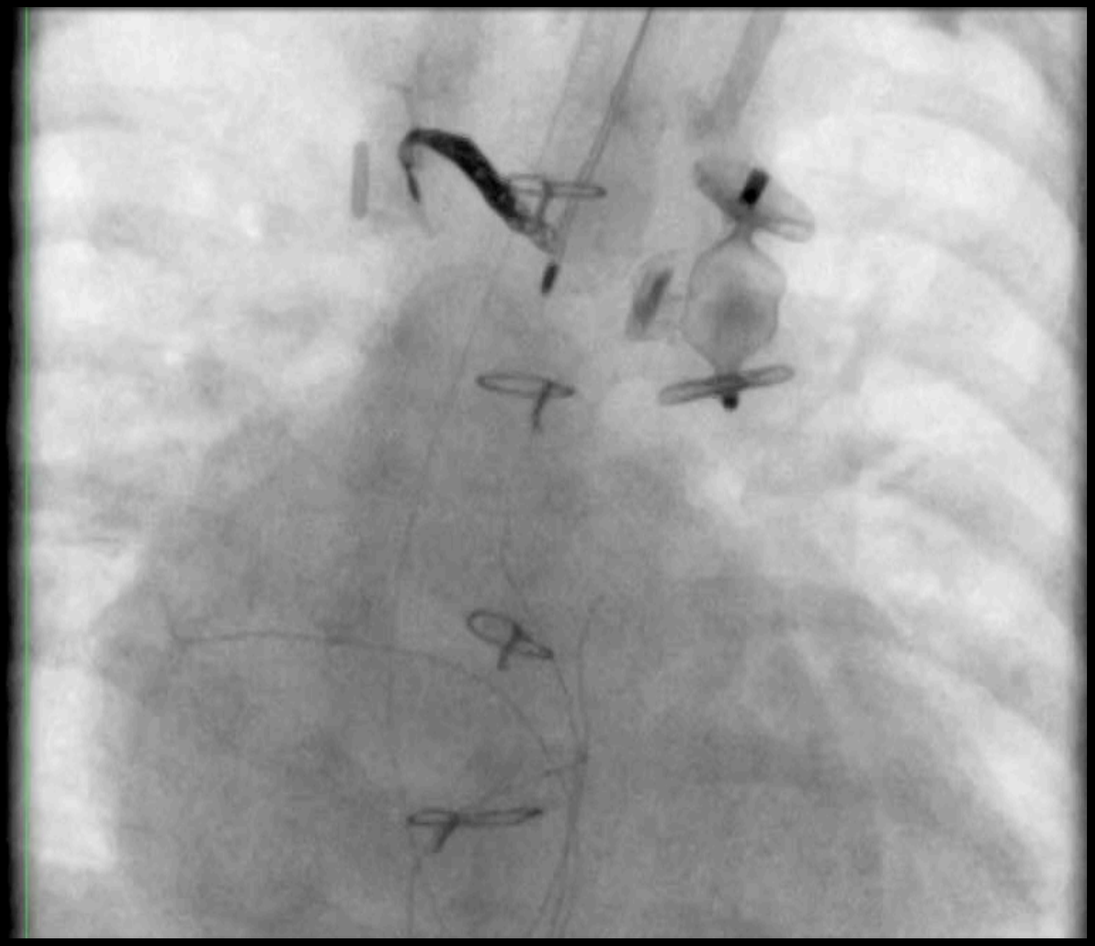
Evaluation pre-TCPC



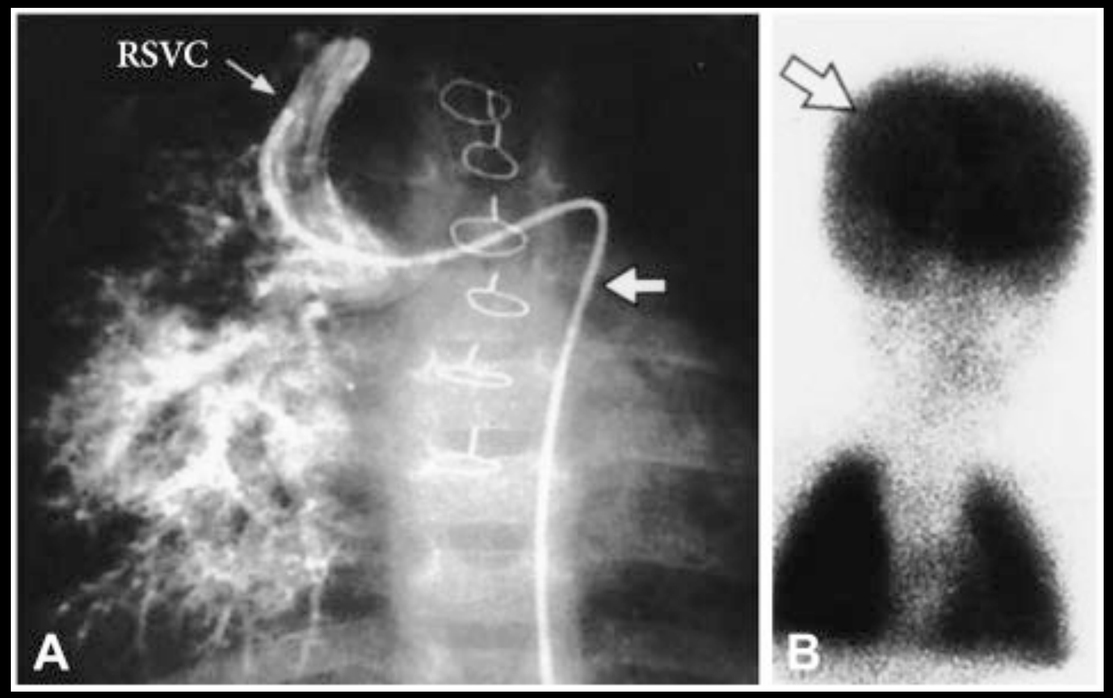


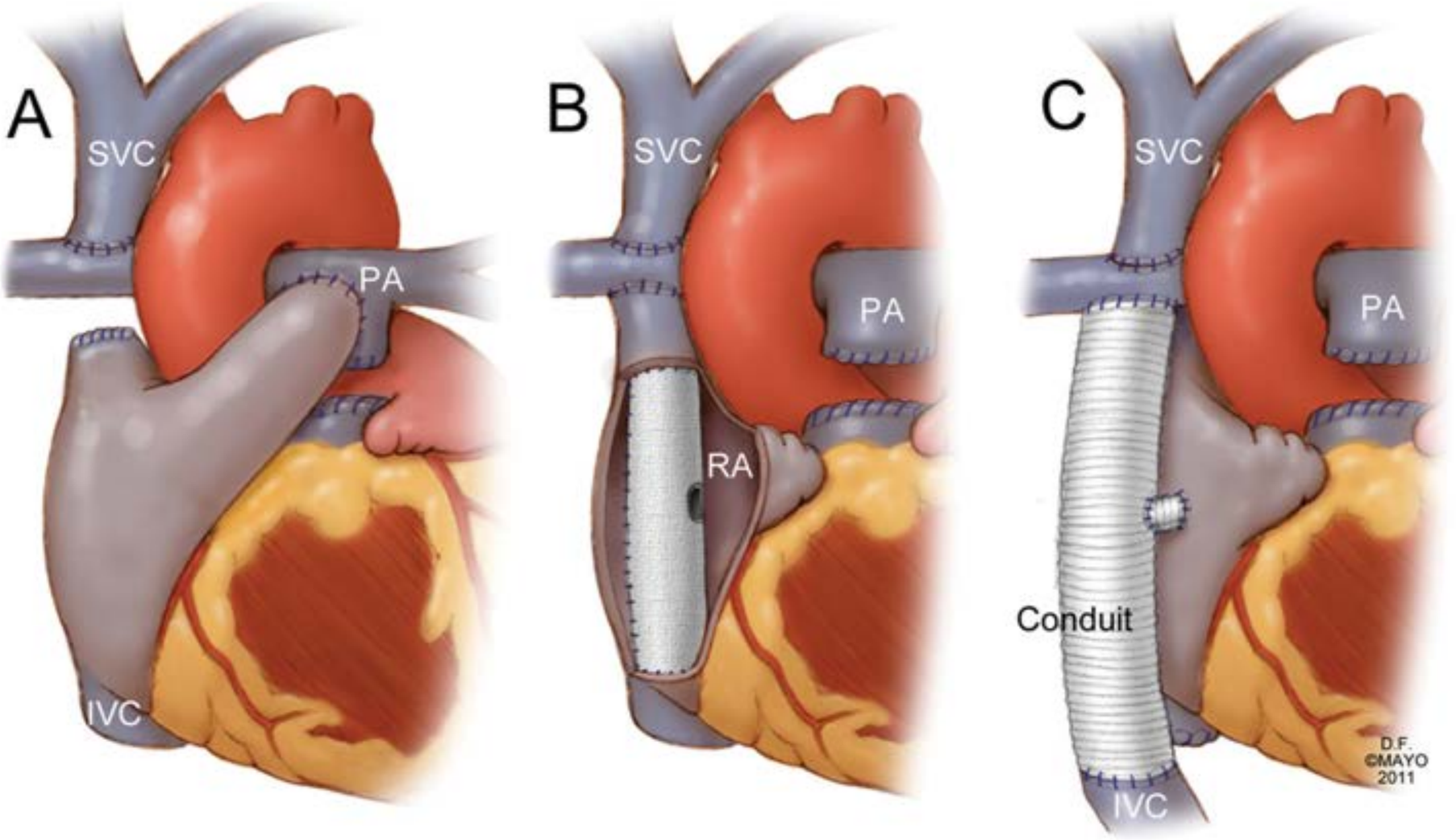
Veno-venous fistulae

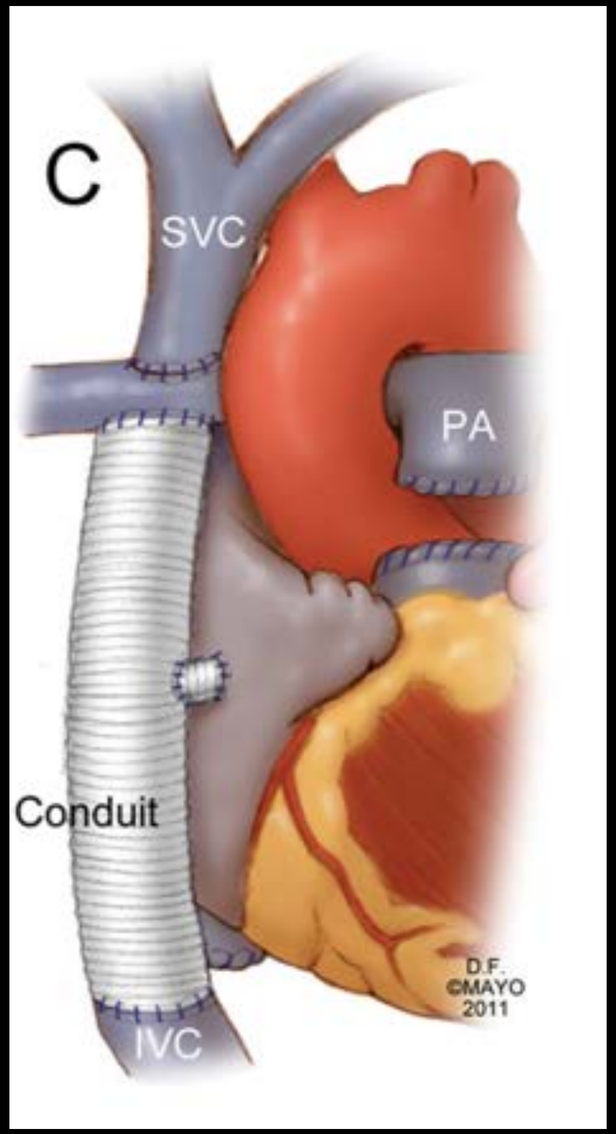




Arterio-venous lung fistulae




A
SVC
PA
IVC
B
SVC
PA
RA
C
SVC
PA
Conduit
IVC
D.F.
©MAYO
2011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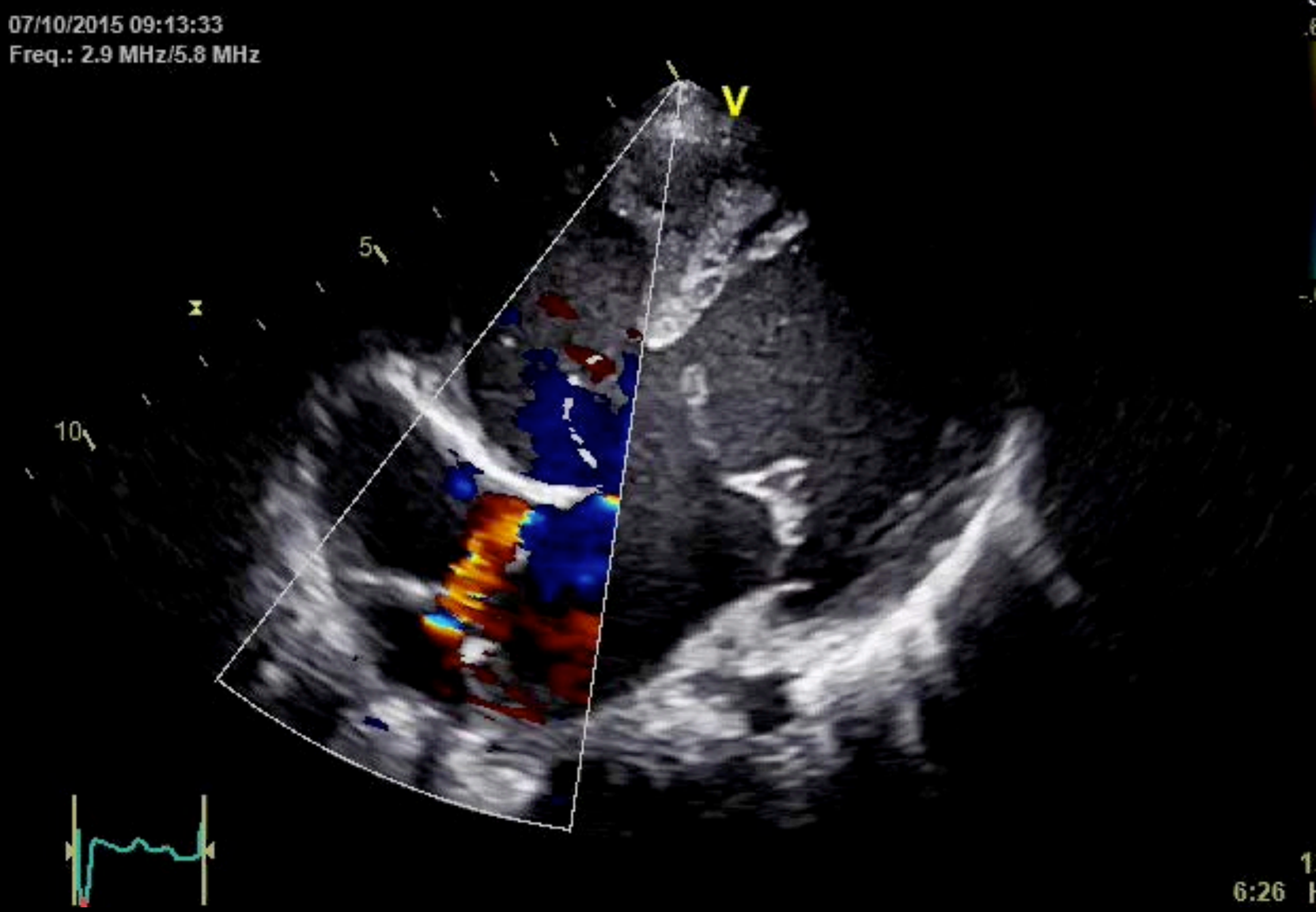
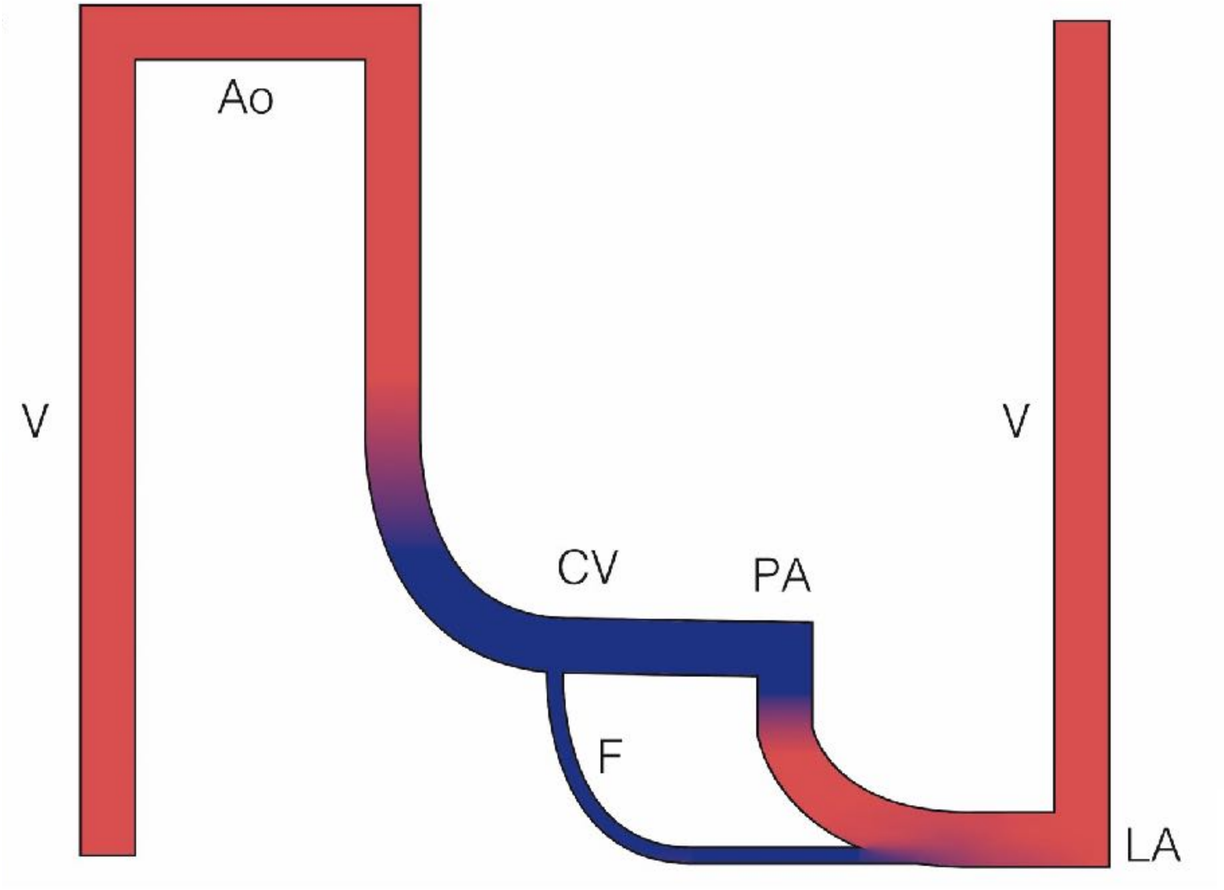
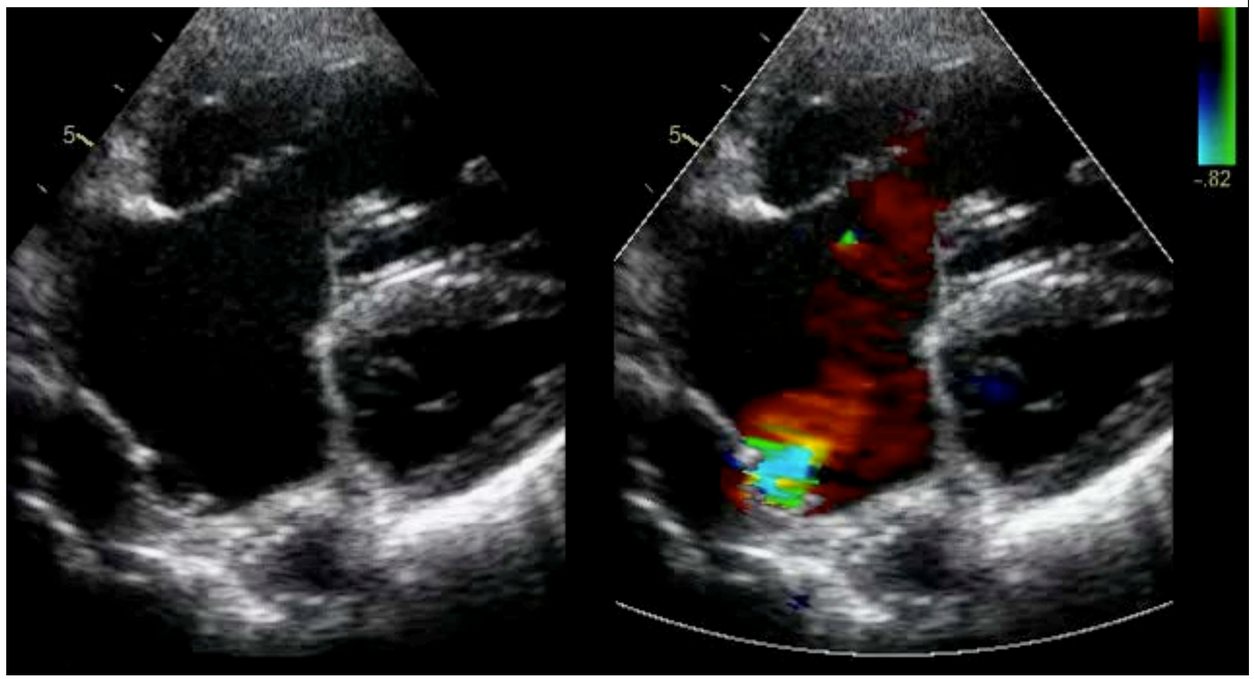

07/10/2015 09:13:33
Freq.: 2.9 MHz/5.8 MHz
V
5
10
6:26

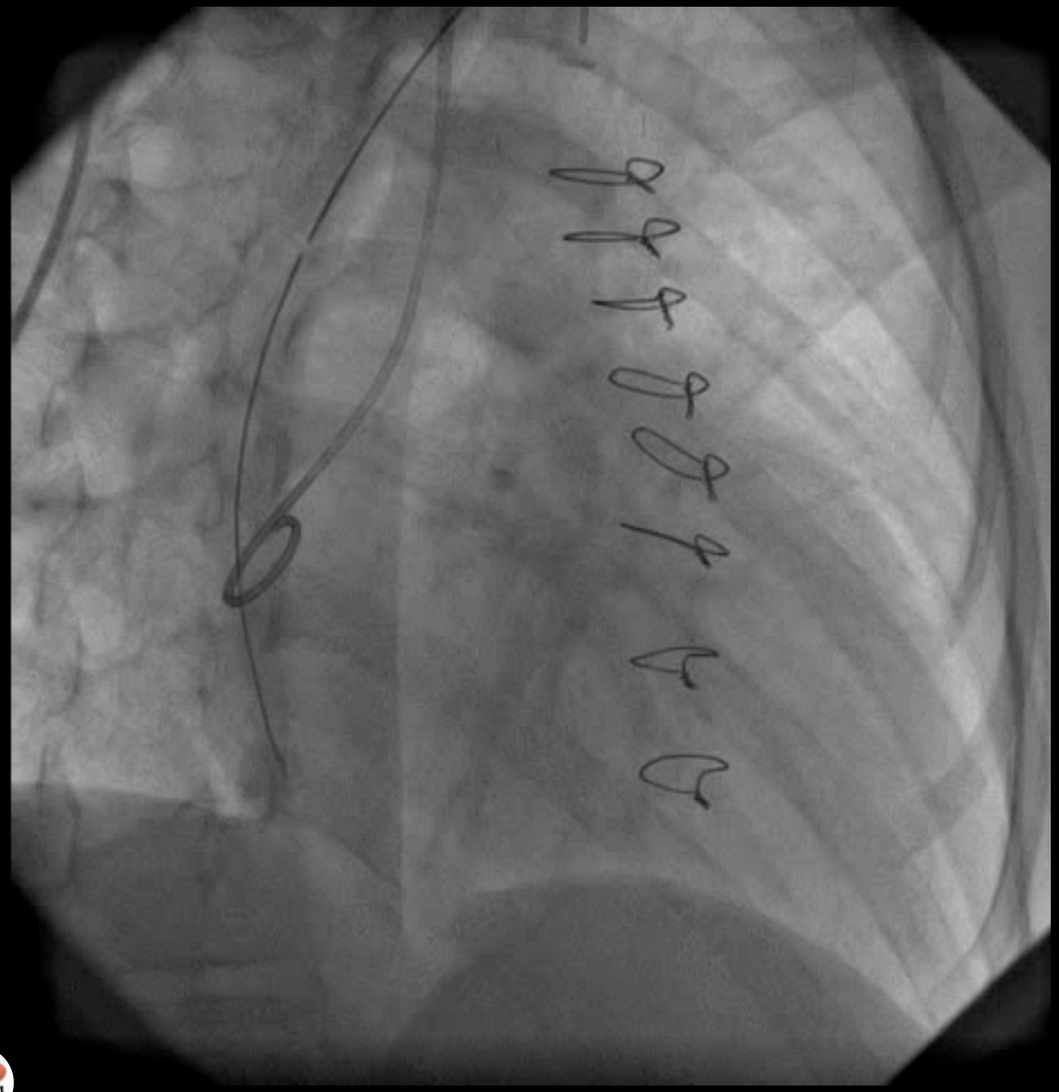
Should we close fenestrations to prevent stroke ?
or leave them open to improve cardiac output ?

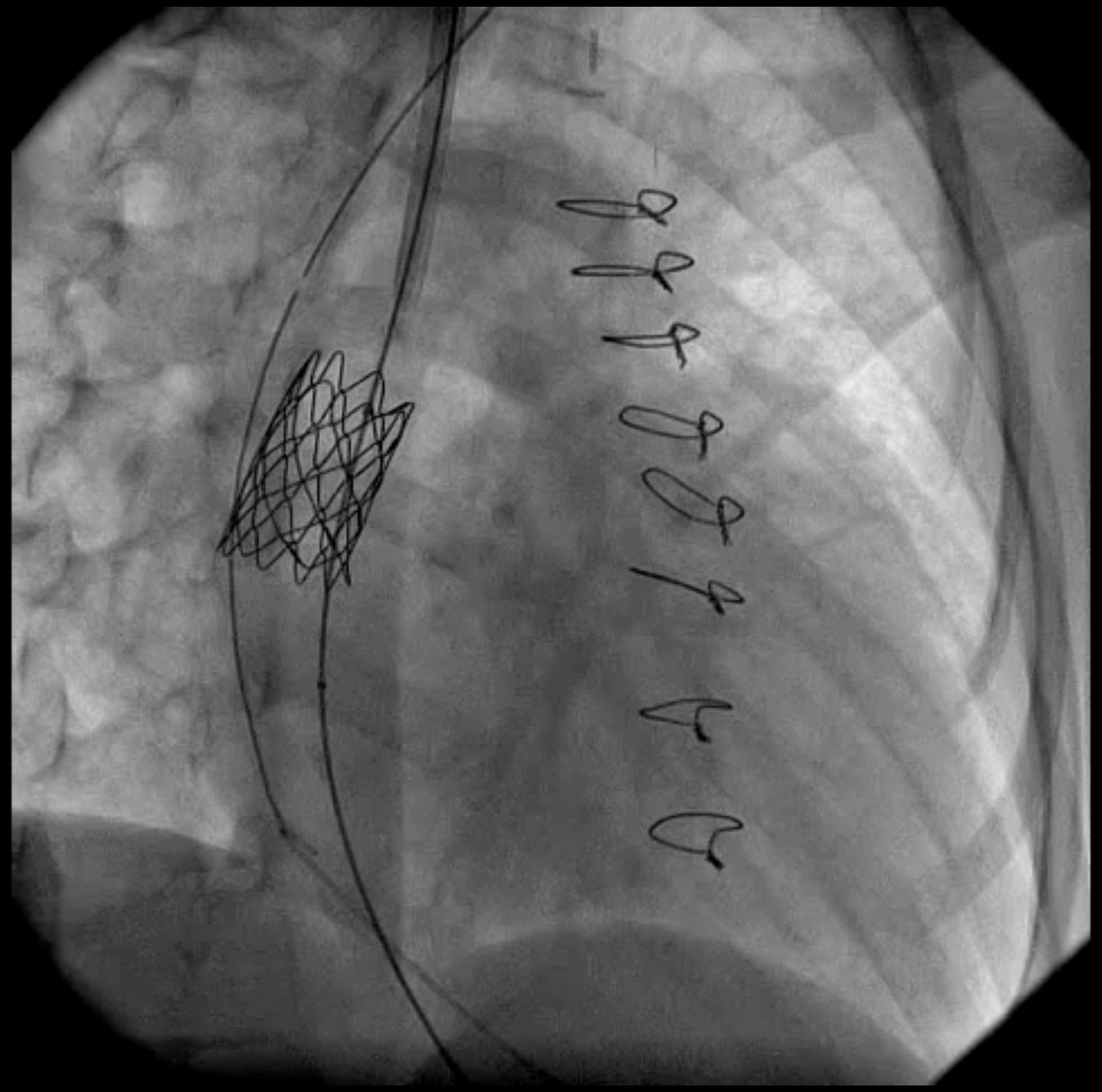




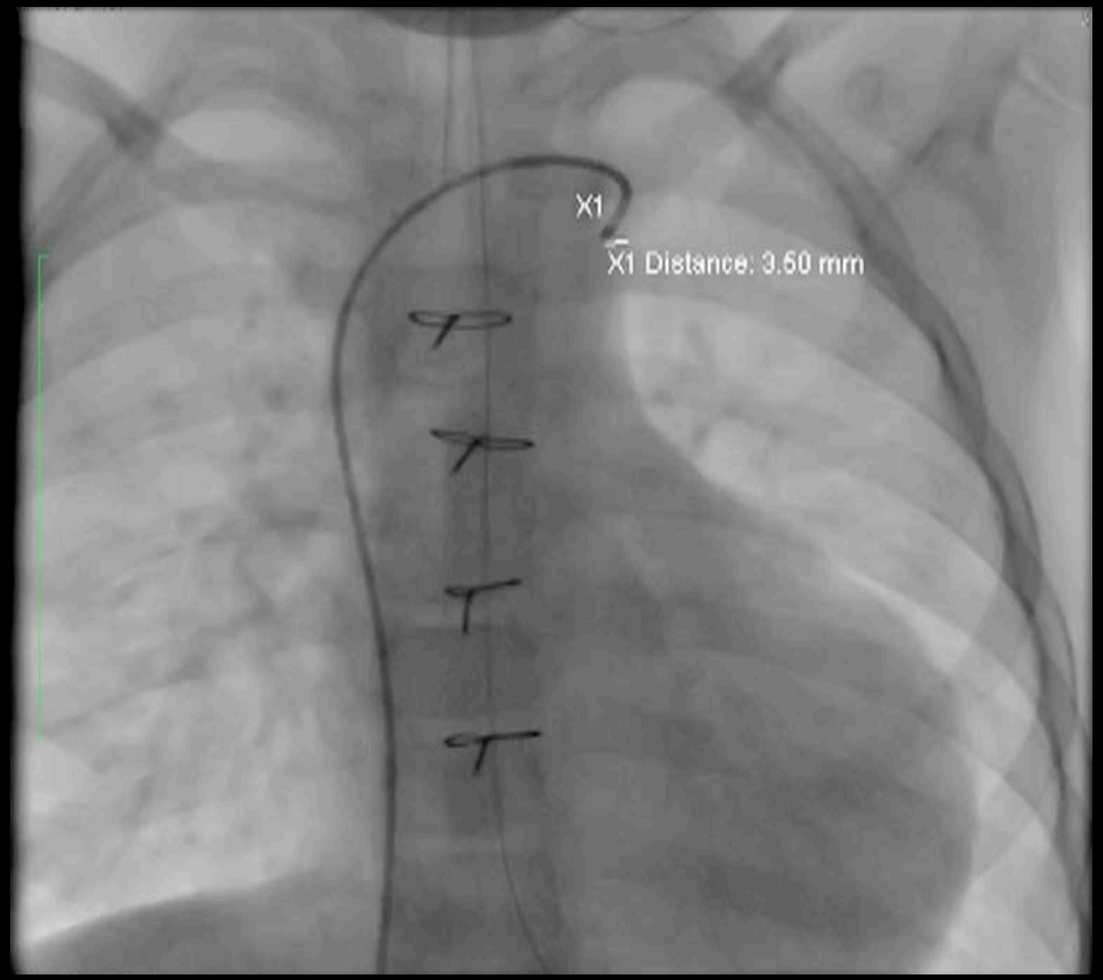
Fenestration device closure

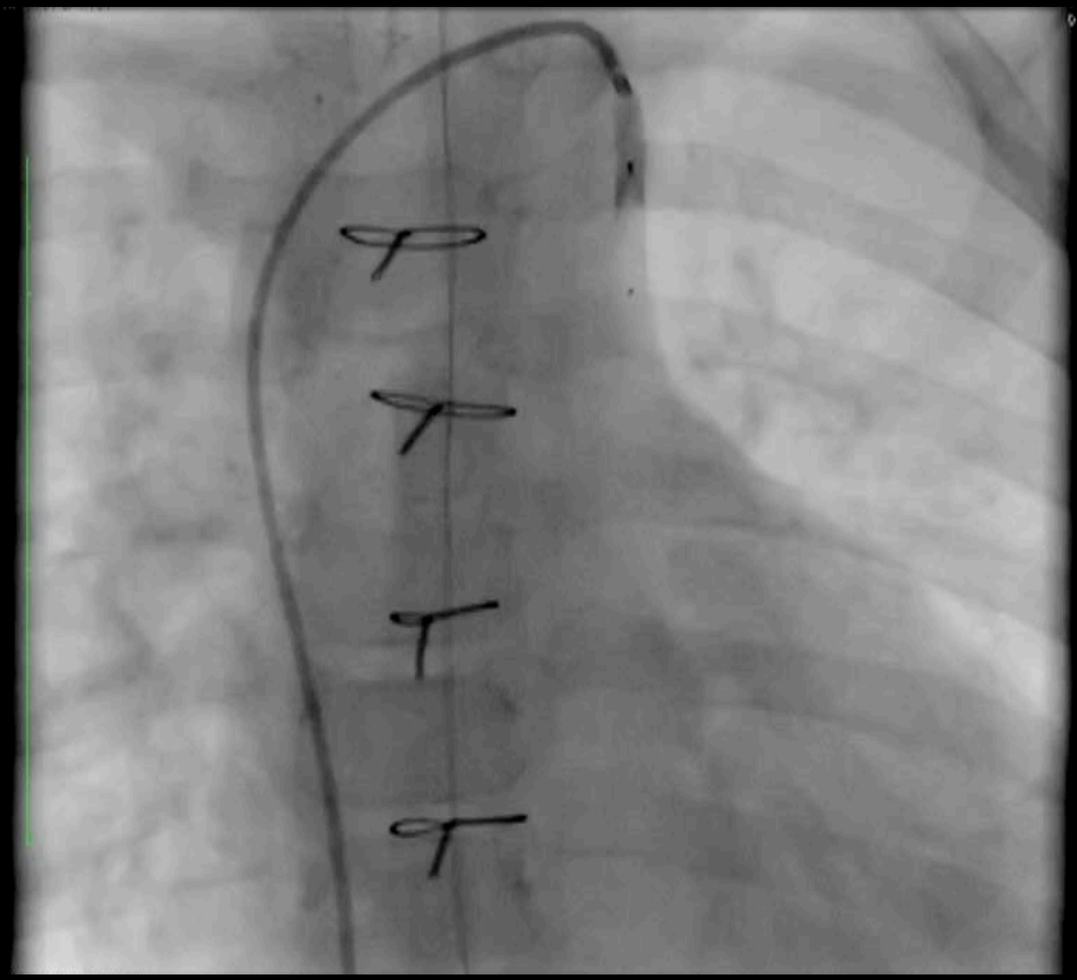
Fenestration covered stent closure



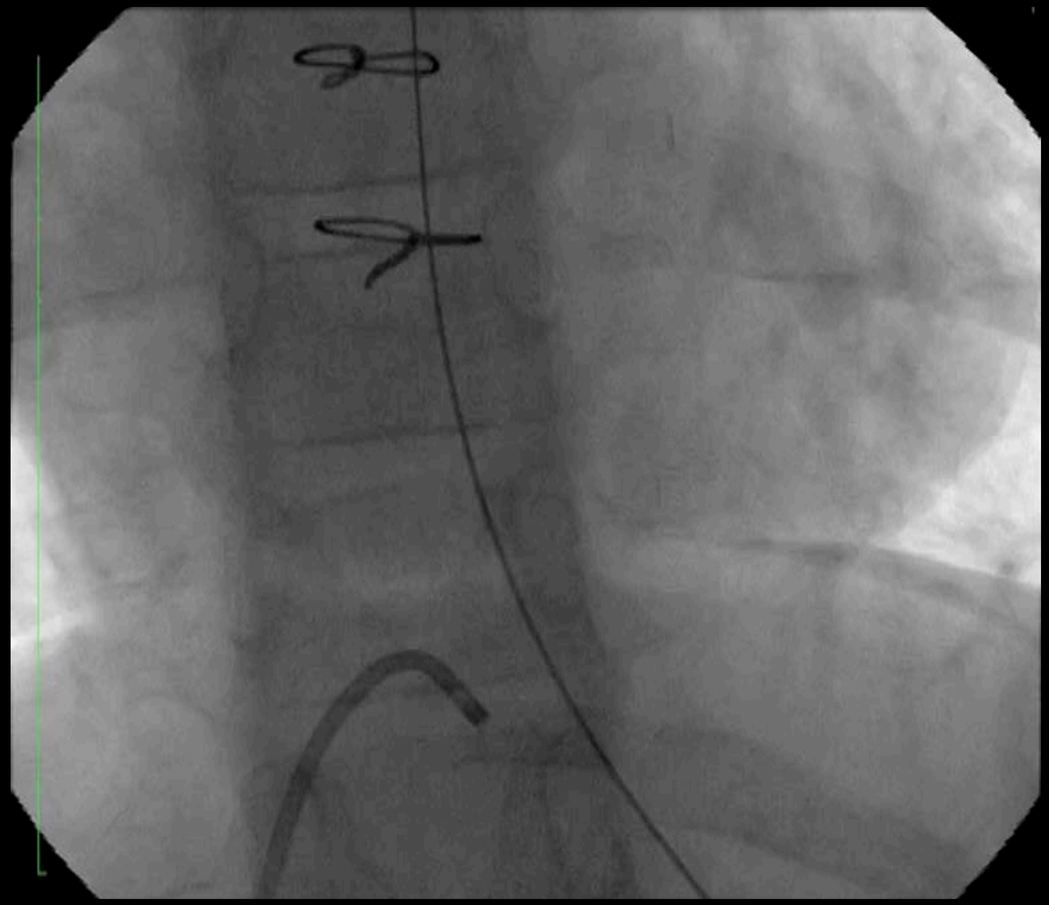


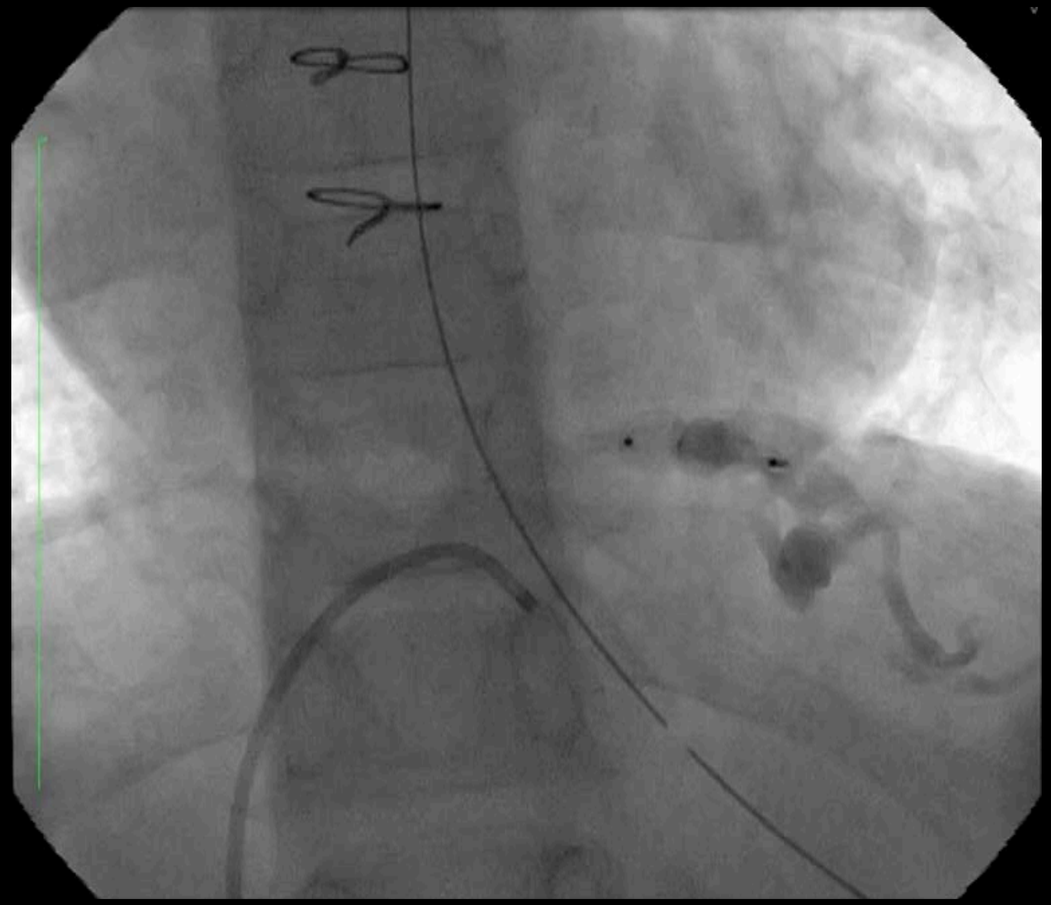
Venous fistulae in coronary sinus in TCPC

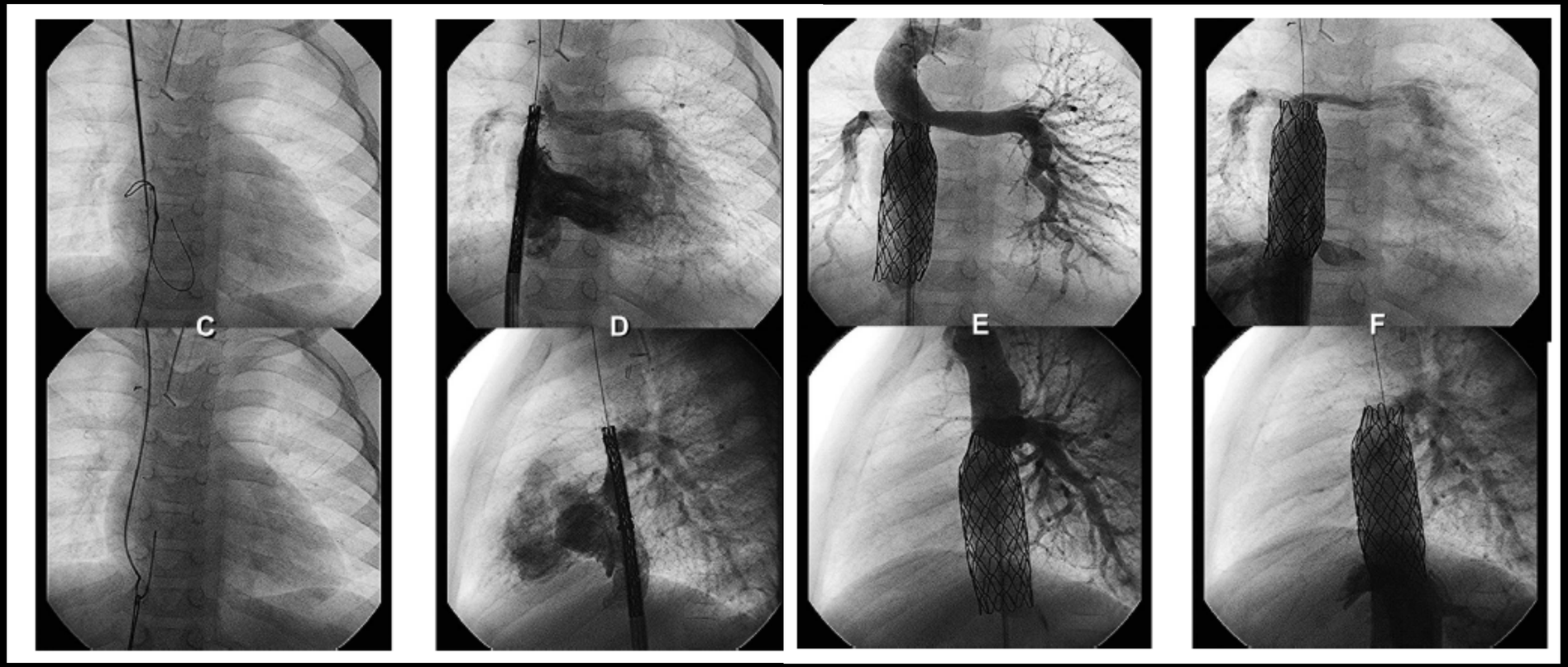




Venous fistulae in TCPC

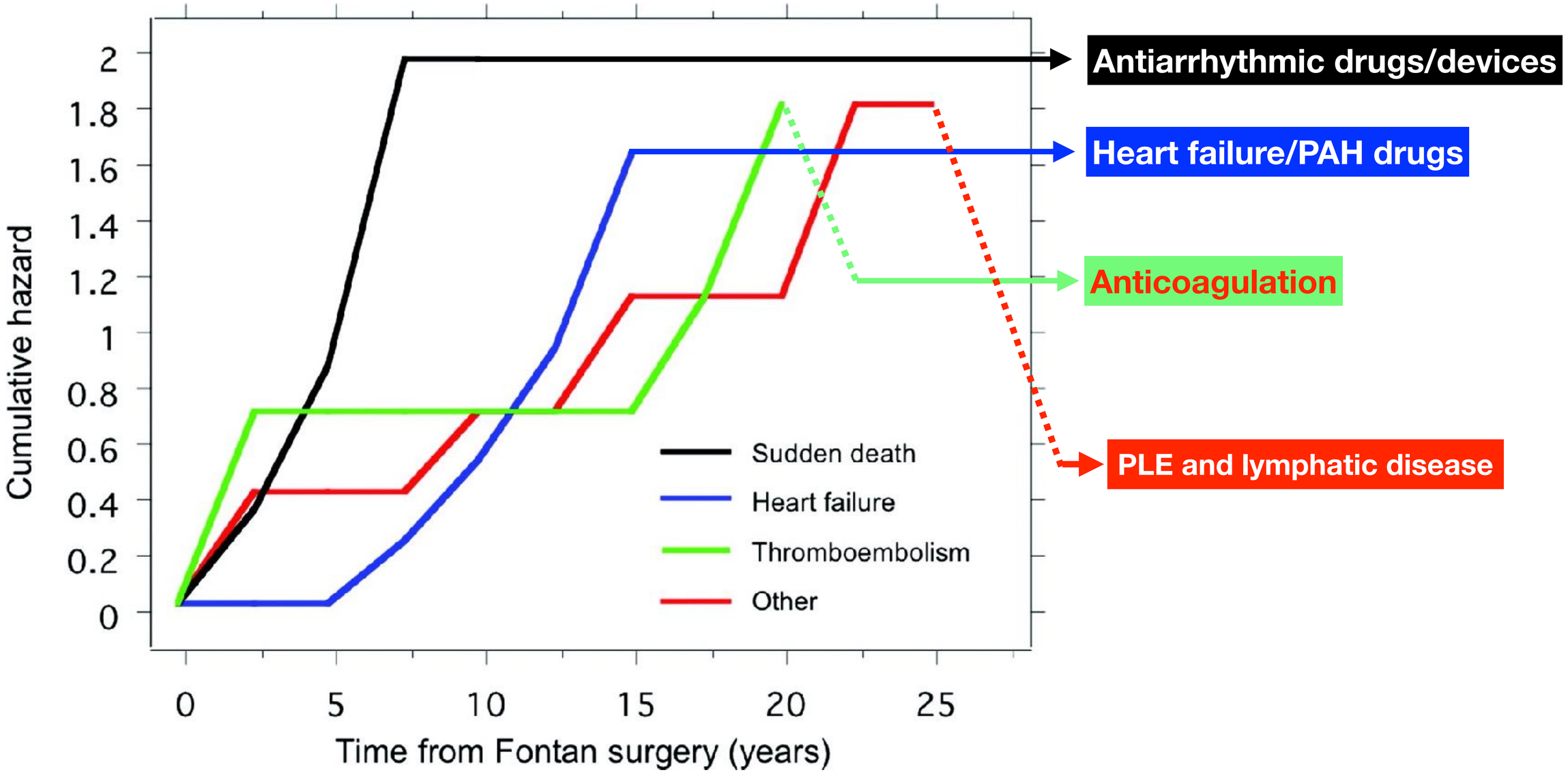





C
D
E
F

Follow-up of patients with TCPC

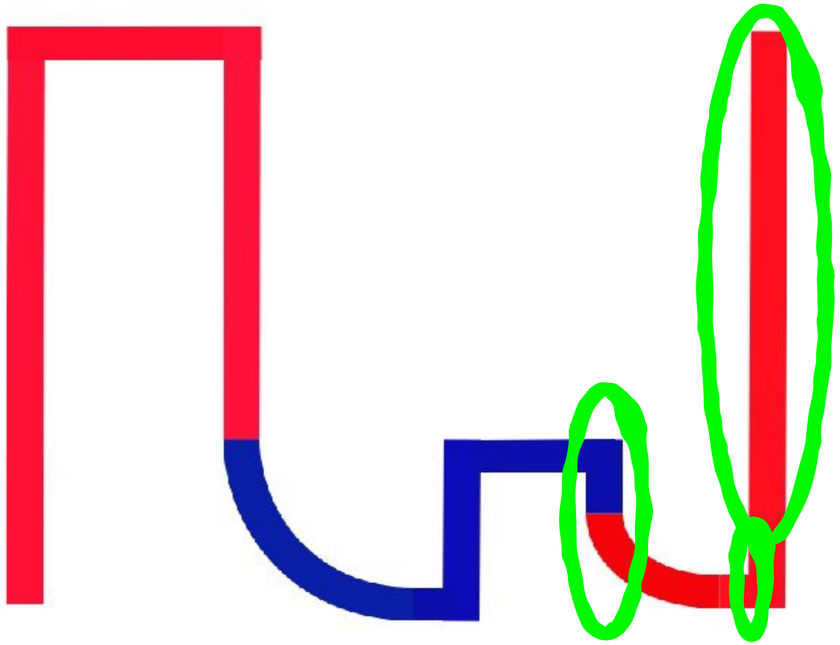
Cumulative hazard by mode of death



Khairy, P. et al. Circulation 2008;117:85-92

Critical bottlenecks in various circulations

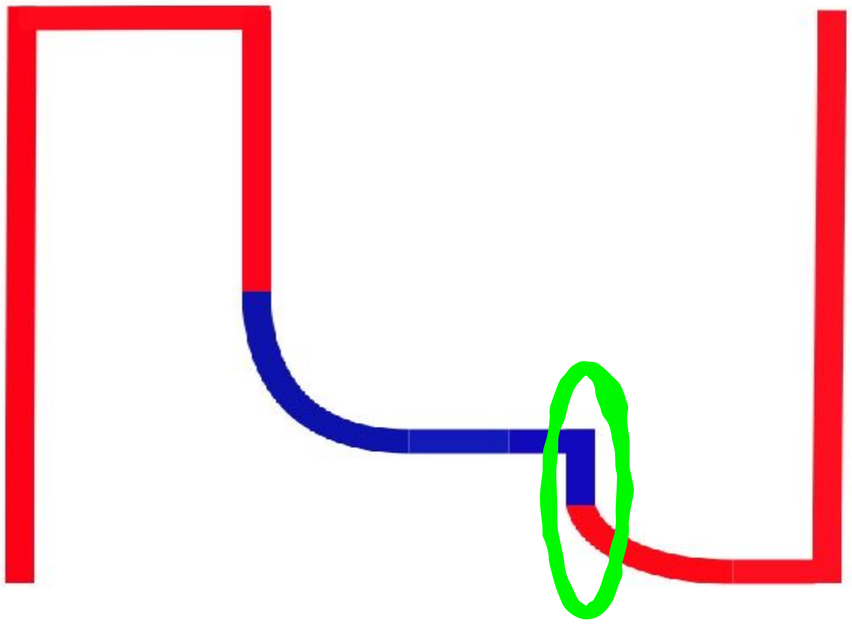
The Fontan circulation - a new portal system



Cardiomyopathies

Pulmonary hypertension

Mitral stenosis

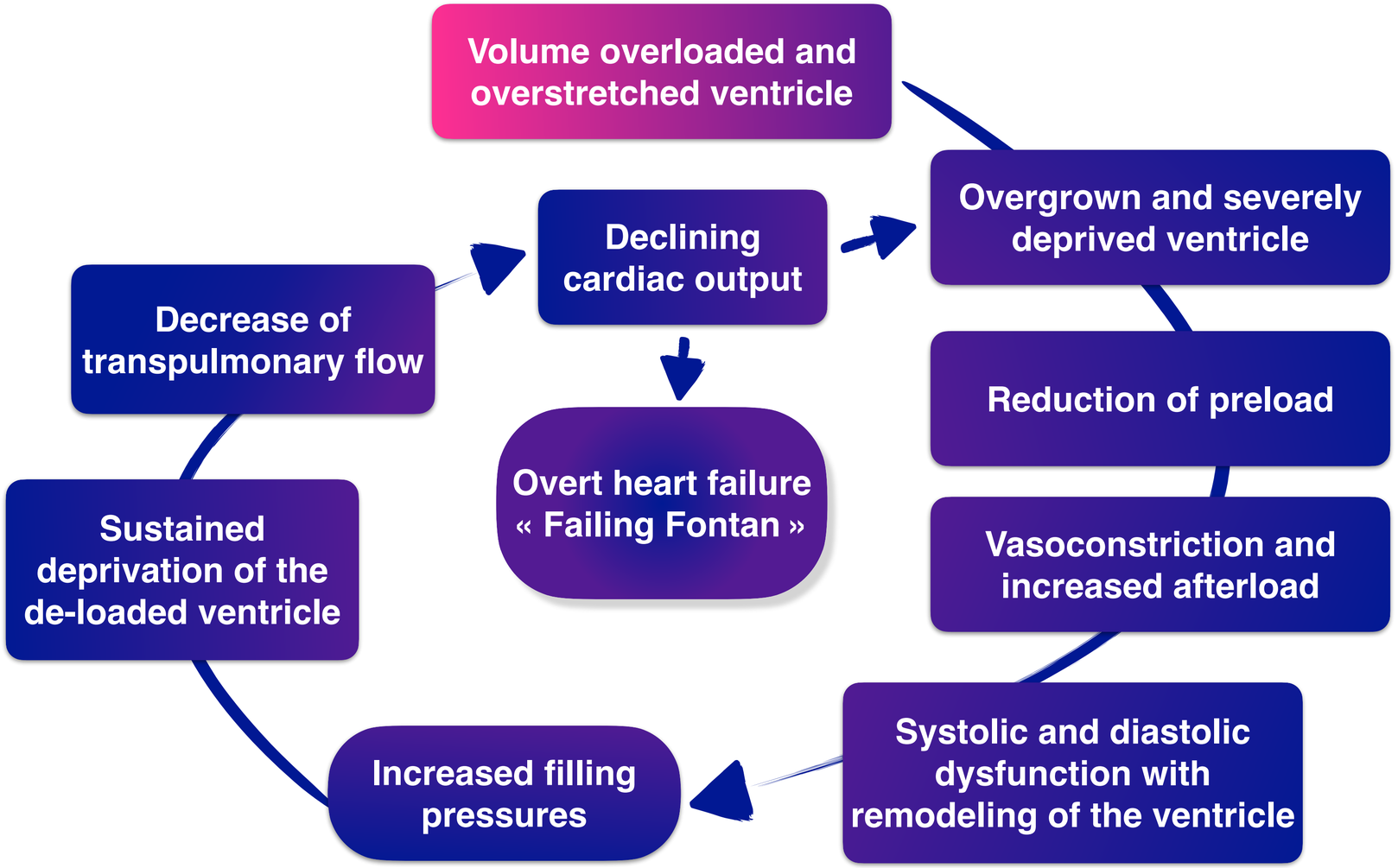


Fontan TCPC

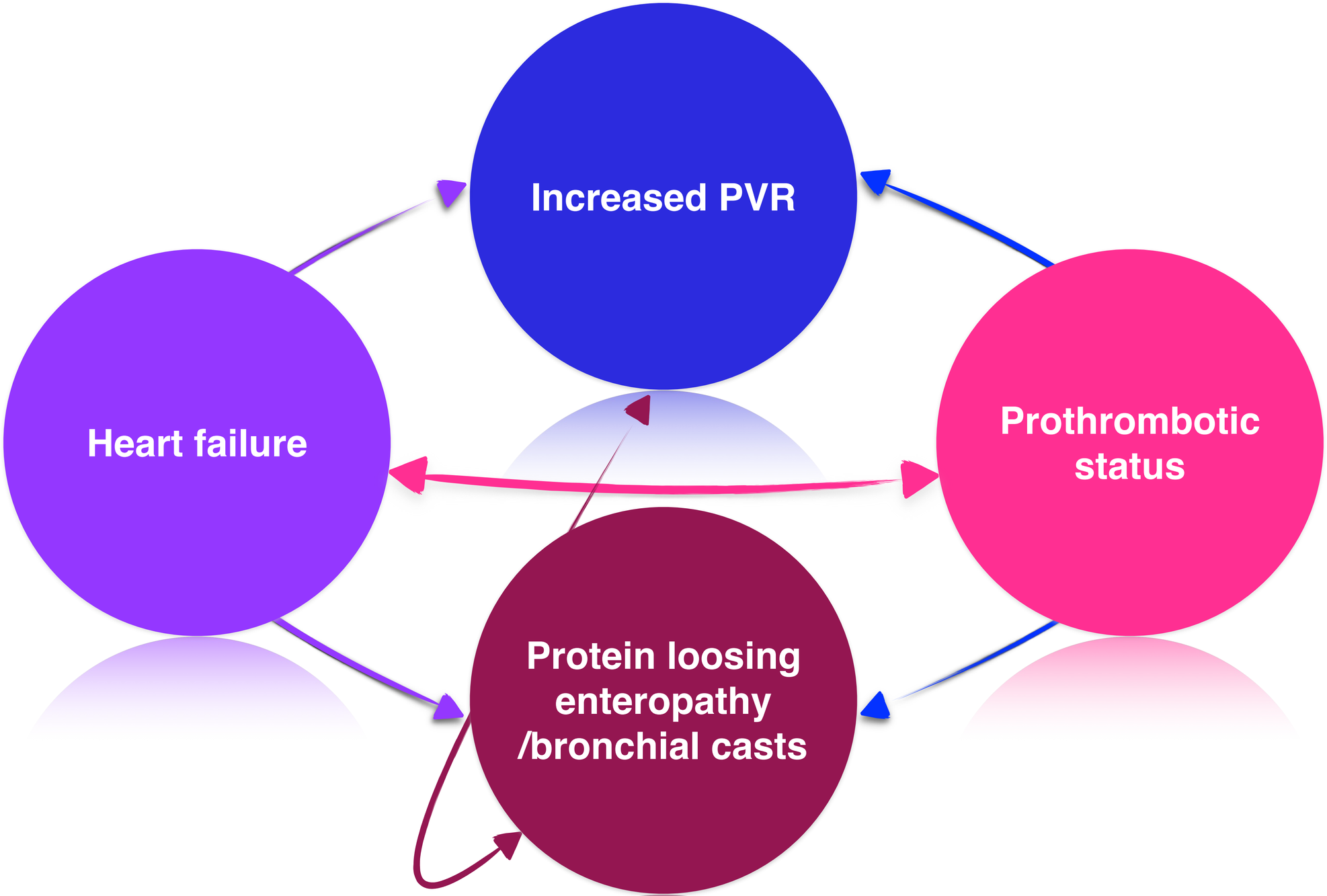
Courtesy Marc Gewillig

The Fontan circulation - a new portal system

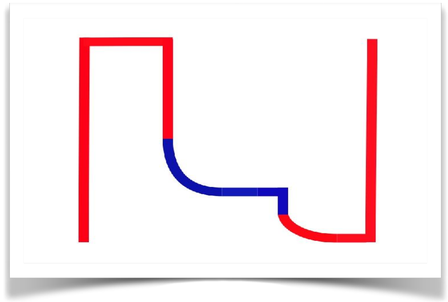
The vicious circle to failing Fontan



Adapted from Marc Gewillig-Heart 2016


Increased PVR
Heart failure
Prothrombotic status
Protein loosing enteropathy /bronchial casts

Optimization of the Neo-portal Fontan system



- **Connection**
  - No gradient
  - Minimal turbulence & flow collision; hepatic X-factor to both lungs
- **Lungs**
  - ***Total PVR as low as possible***
    - ✓ capacitance
    - ✓ recruitment
  - ***Reasons increased PVR***
    - ✓ Hypoplasia, stenosis, kink, loss of segments
    - ✓ Collateral flow
    - ✓ **Pulmonary vascular disease**
    - ✓ **(micro)Thrombi**
- **Ventricular succion**

Anatomical solutions

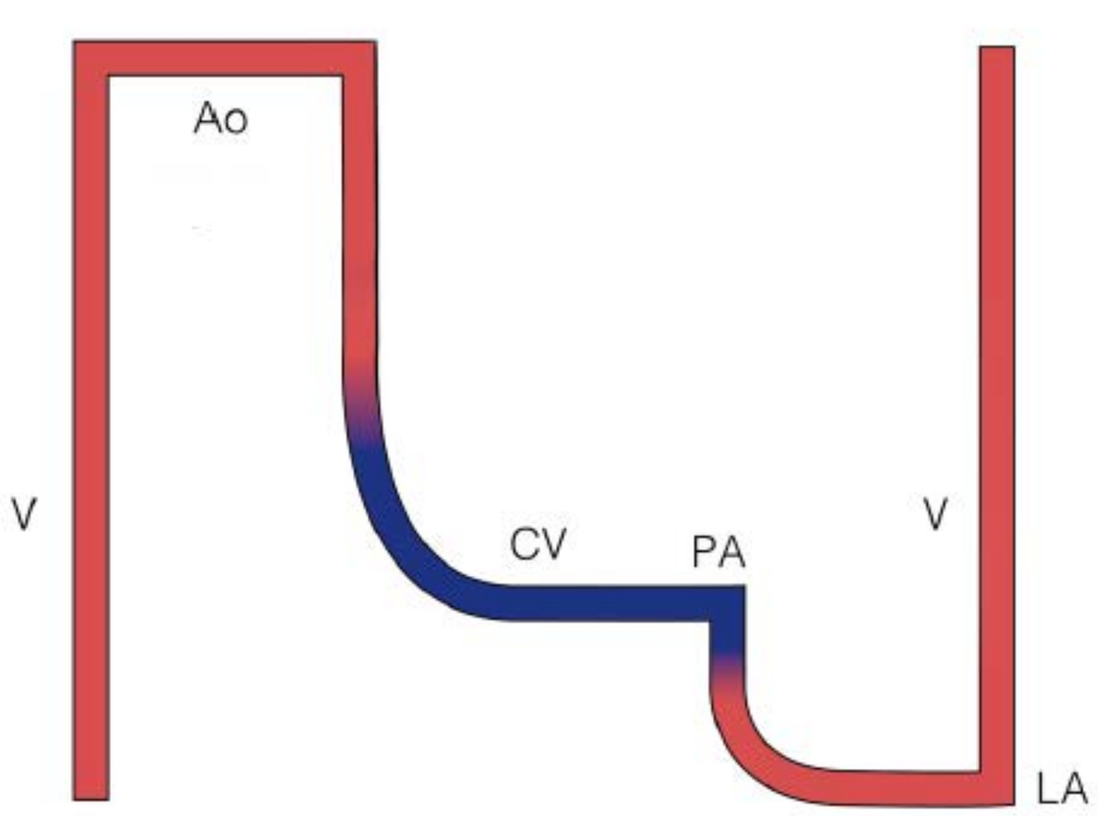
PAH drugs

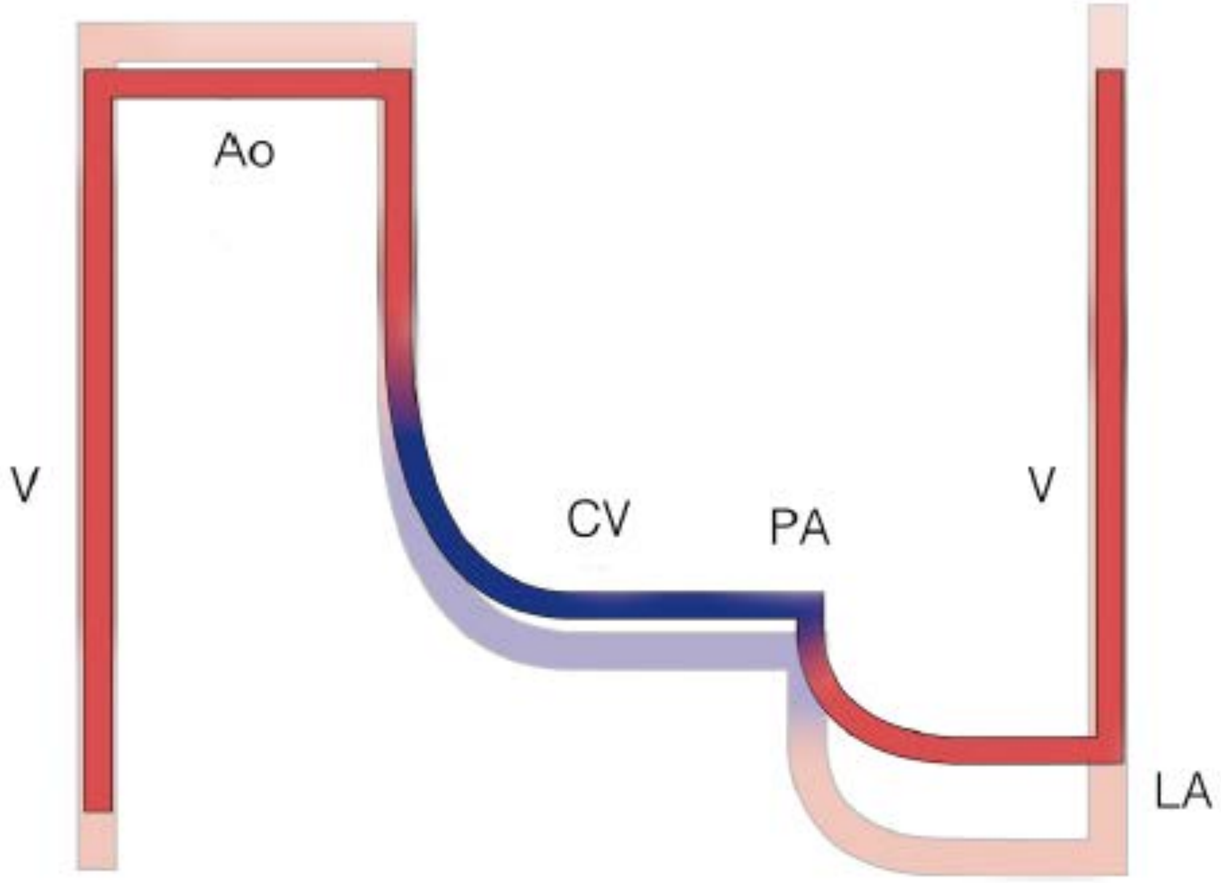
Anticoagulants

Lusitropic drugs

The Fontan circulation - a new portal system

Changes with aging - Preventing strategies

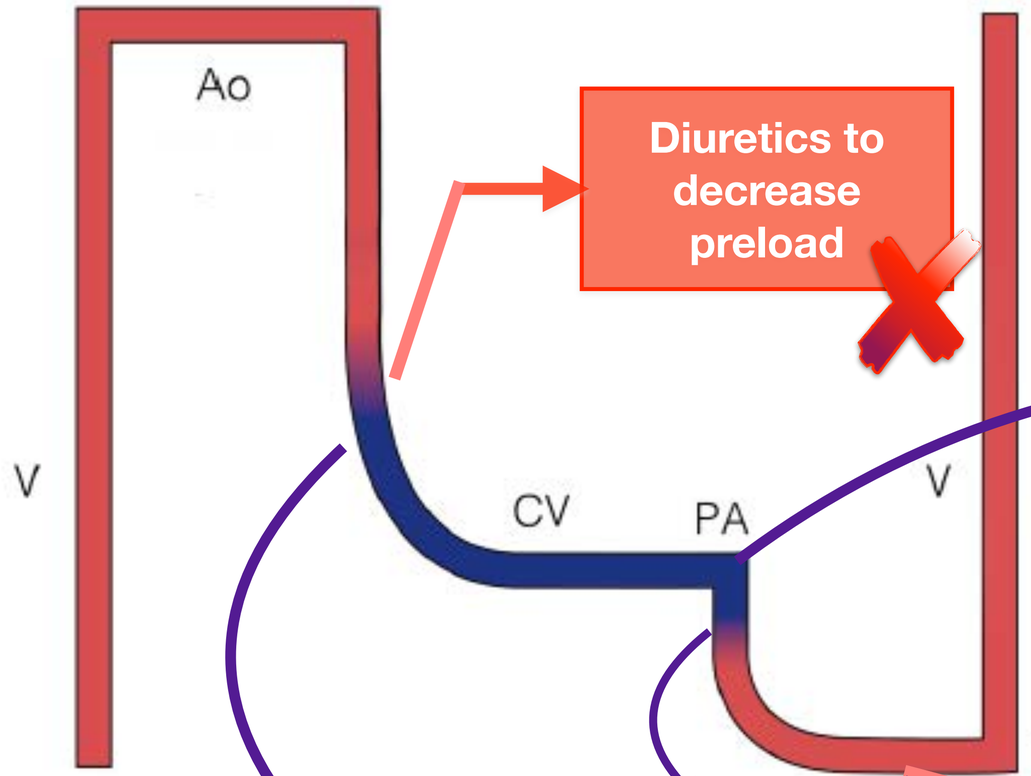




The lack of a robust definition of Fontan failure has contributed to the limited understanding of the prevalence of HF in Fontan-palliated SVs

Heart failure drugs in Fontan circulation

Potentially a wrong reasoning and a predictable minimal effect



Diuretics to decrease preload

ACE Inhibitors to decrease after load

Pulmonary vasodilatation to increase preload

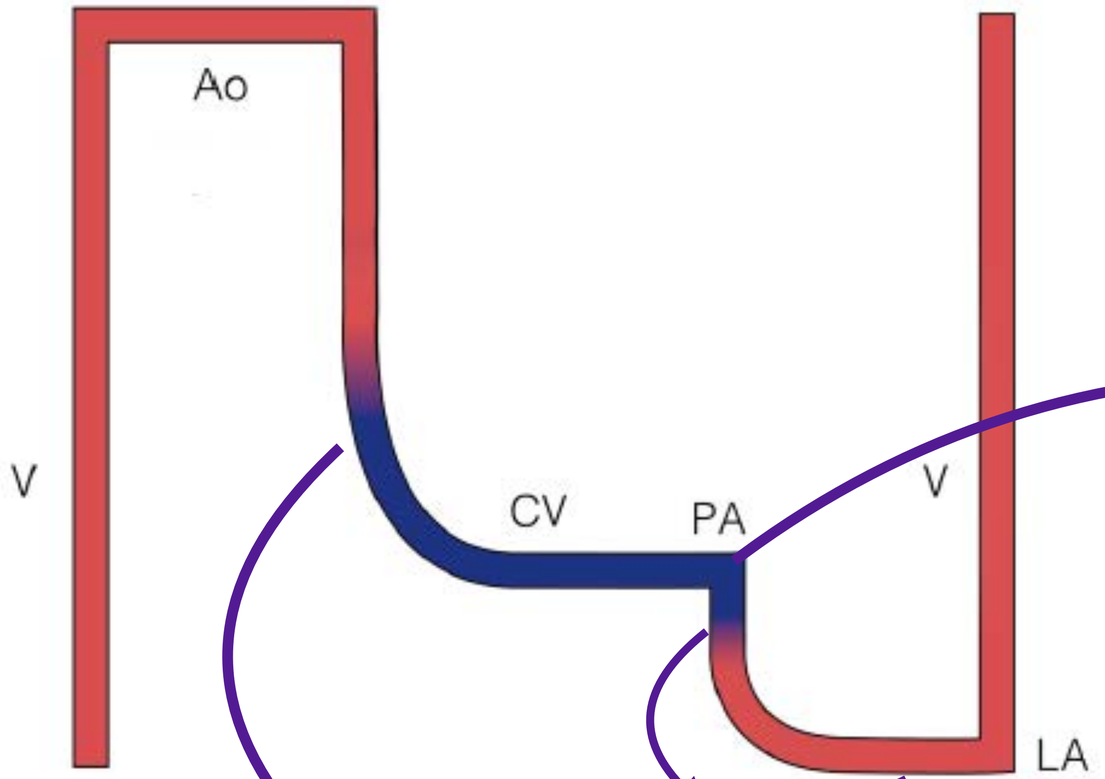
Beta-blockers to lengthen diastole and ventricular filling

Exercise to increase preload and decrease PVR

Lusitropic drugs sGC stimulators

Heart failure drugs in Fontan circulation

Potentially efficient therapies



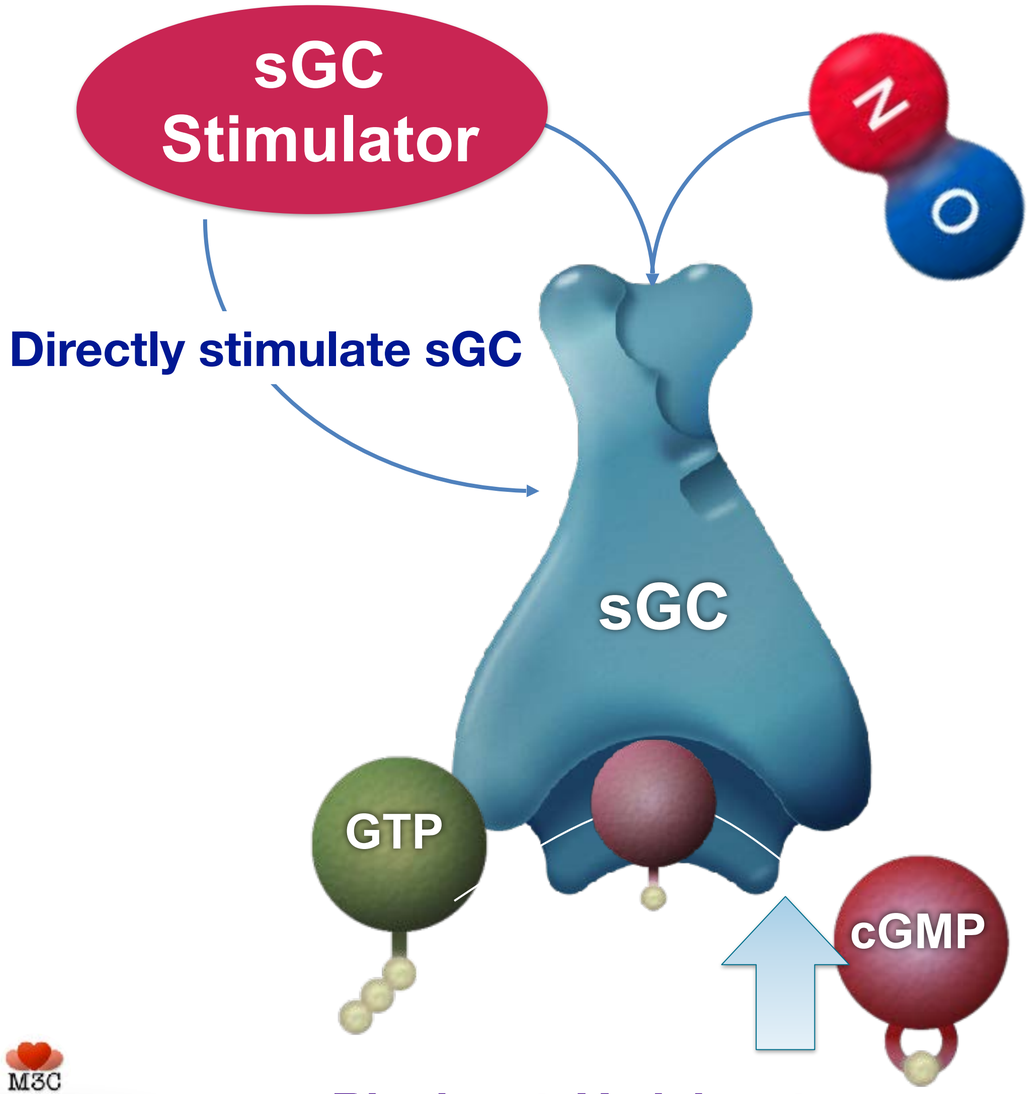
Pulmonary vasodilatation to increase preload

Exercise to increase preload and decrease PVR

Lusitropic drugs sGC stimulators

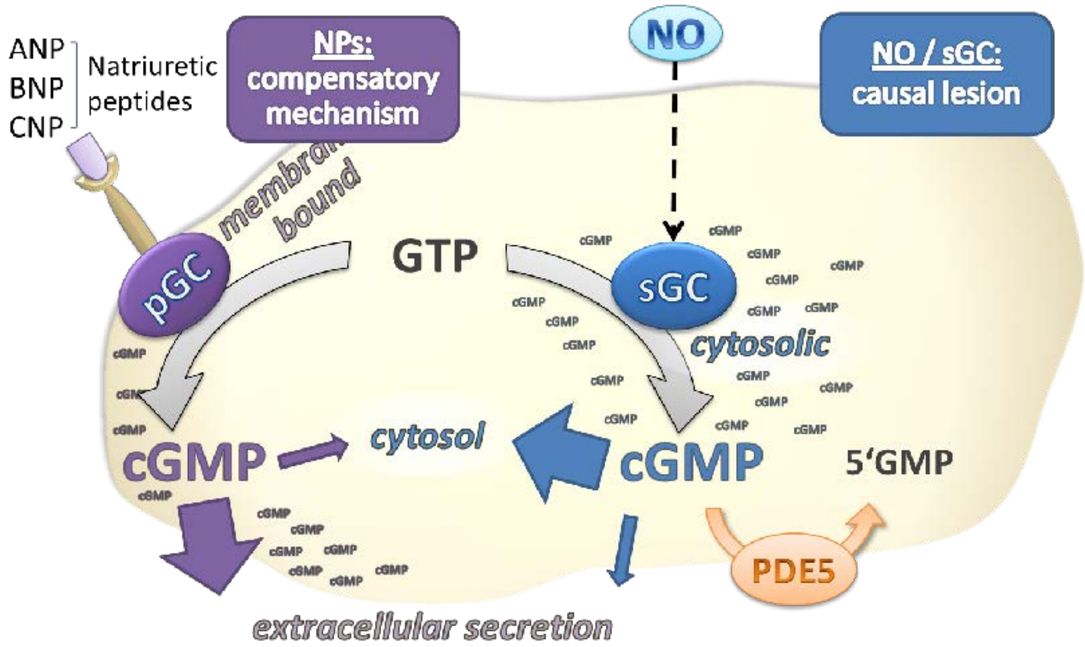
Lusitropic drugs in univentricular hearts

Enhances sensitivity of sGC to NO



Riociguat, Vericiguat

Different cGMP-augmenting pathways targeted in clinical trials

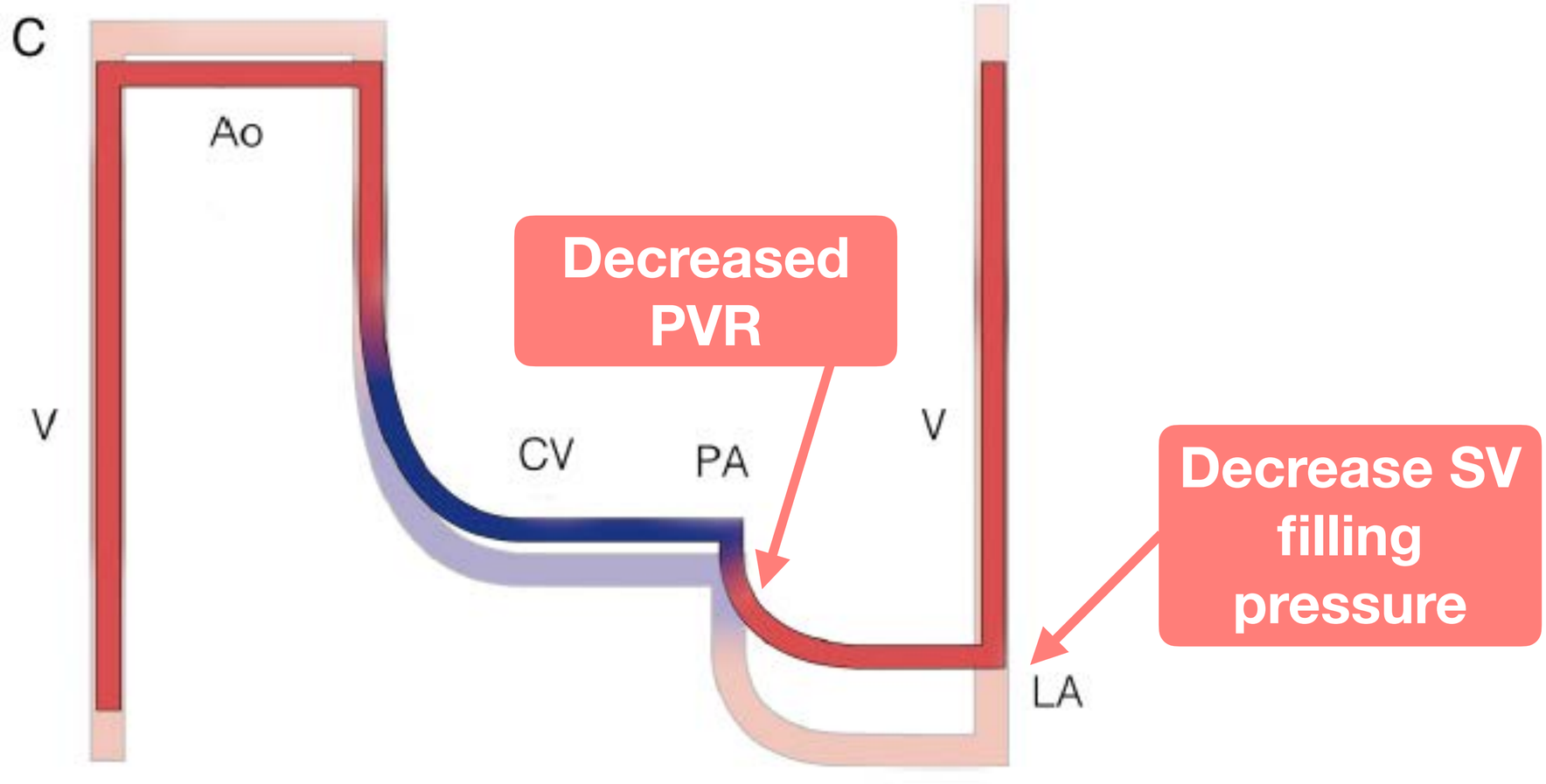


sGC stimulators, PDE-5 inhibitors, Neprilysin inhibitors (LCZ)

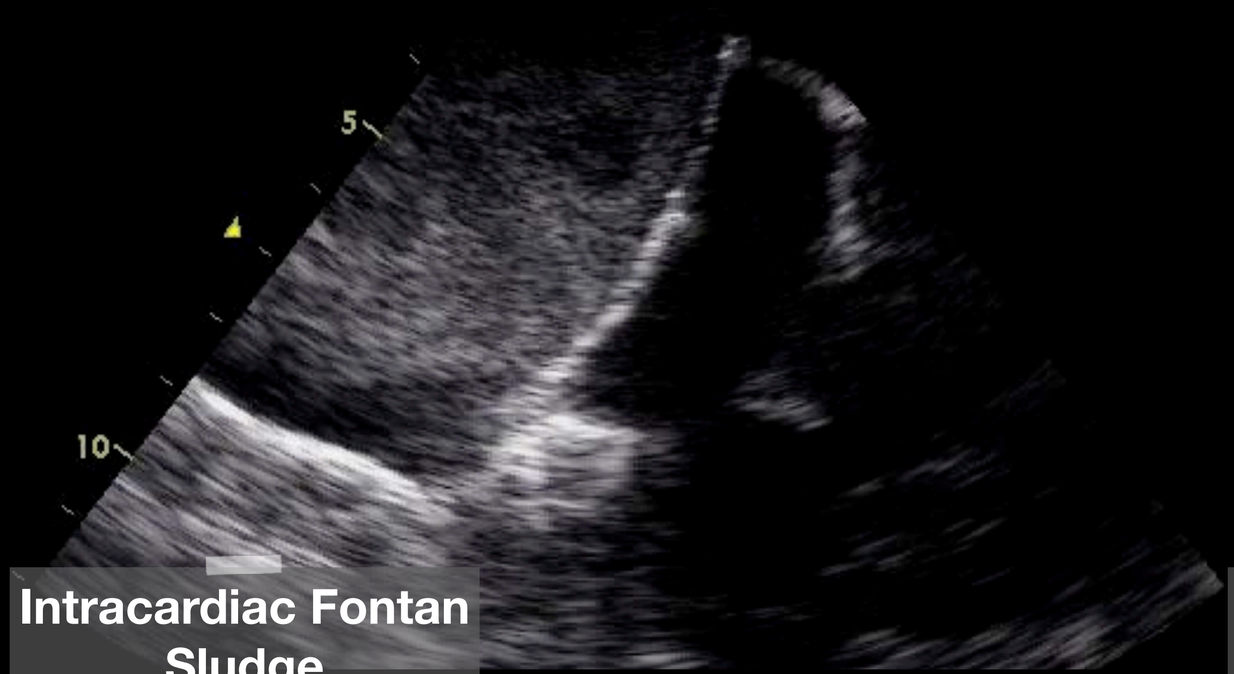
ANP, atrial natriuretic peptide; BNP, brain natriuretic peptide; CNP, c-type natriuretic peptide; NO, nitric oxide; PDE5, phosphodiesterase-5; pGC, particulate guanylyl cyclase; sGC, soluble guanylyl cyclase. modified after Senni et al., Eur Heart J. 2014 Oct 21;35(40):2797-815

Stasch JP et al. Nature 2001;410:212–15;
Greene SJ et al. J Am Heart Assoc 2013;2:e000536;

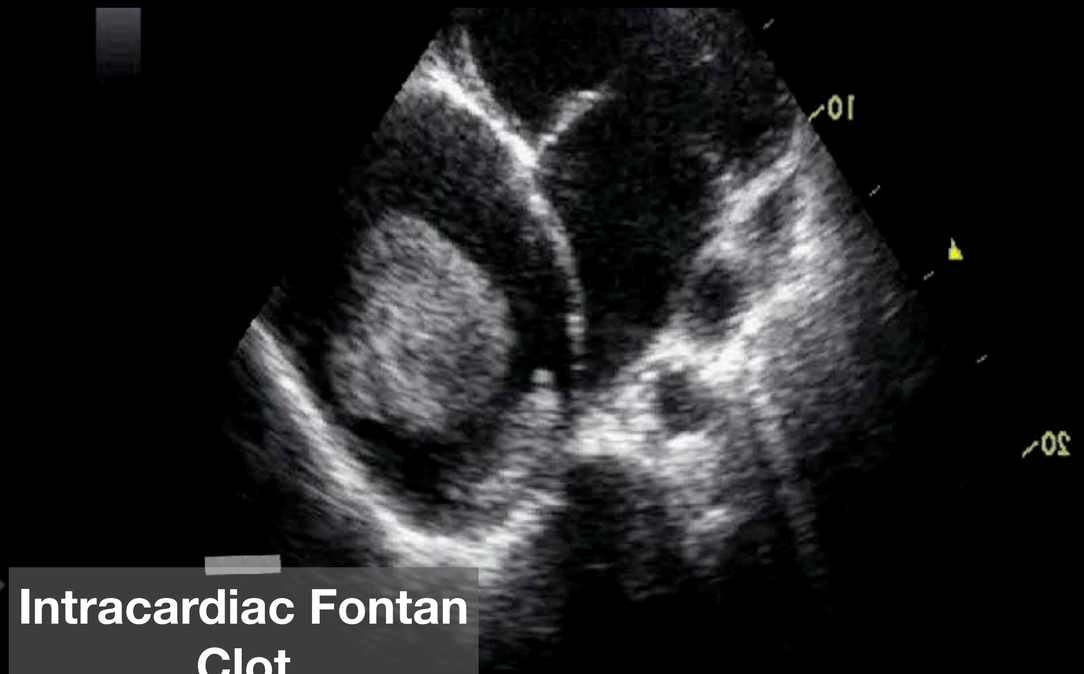
The RioFontan study : riociguat in patients with Fontan circulation



The antithrombotic treatment in Fontan circulation

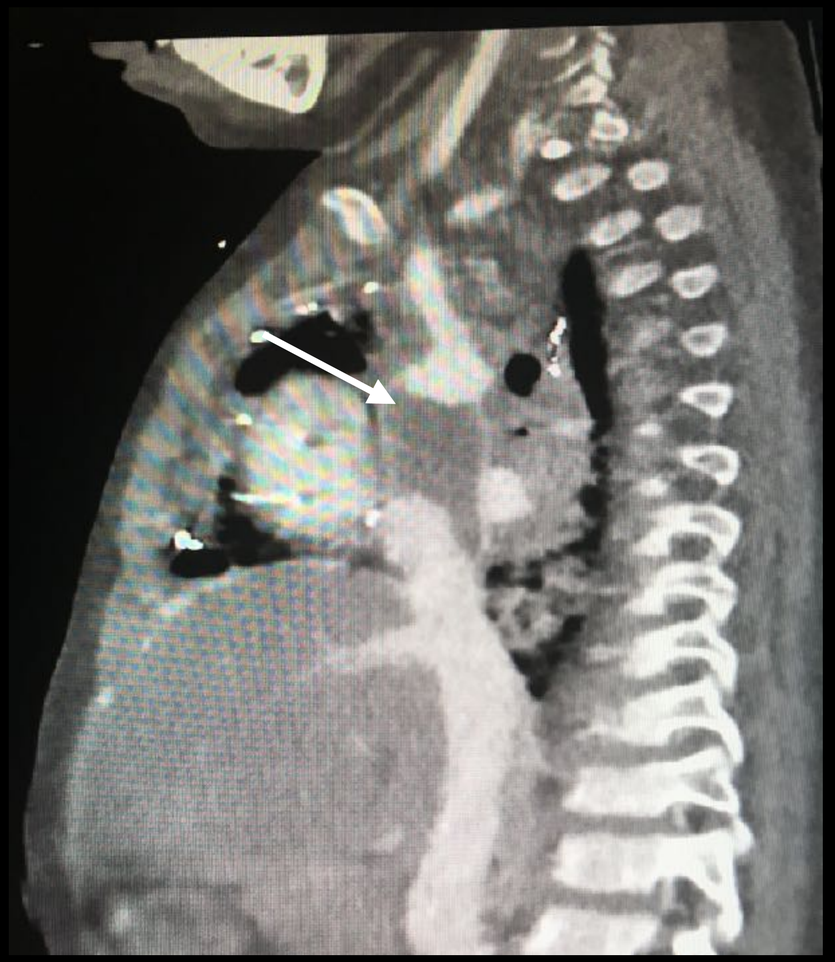


Intracardiac Fontan Sludge



Intracardiac Fontan Clot

The antithrombotic treatment in Fontan circulation



Extracardiac tube
Fontan Clot

Predictors of thrombo-embolic deaths in survivors

Characteristic	Hazard ratio	*95%CI*	*P*
Univariate			
Atrial fibrillation	5.4	*1.0-29.4*	0.0529
Lack of aspirin or warfarin therapy	5.7	*1.0-32.3*	0.0515
RA pressure on follow-up, mmHg	1.26	*1.03-1.53*	0.0247
Thrombus within Fontan	4.9	*2.1-11.6*	0.0002
Multivariate			
Thrombus within Fontan	22.7	*4.3-120.0*	0.0002
Lack of aspirin or warfarin therapy	91.6	*4.2-2004.8*	0.0041

All but nothing

Effect of aspirin and warfarin therapy on TE events in patients with Fontan palliation

Meta-analysis

10 studies with 1200 patients, average F/U 7.1 yrs

Incidence of TE

OR 0.43 (0.19-0.93) for some vs. none
OR 0.36 (0.18-0.74) for ASA vs. none
OR 0.33 (0.17-0.63) for warfarin vs. none
OR 0.94 (0.61-1.44) for warfarin vs. ASA

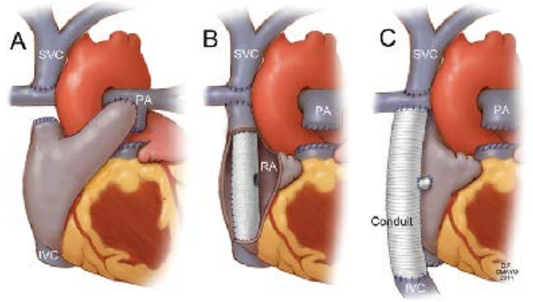
No difference for early or late TE

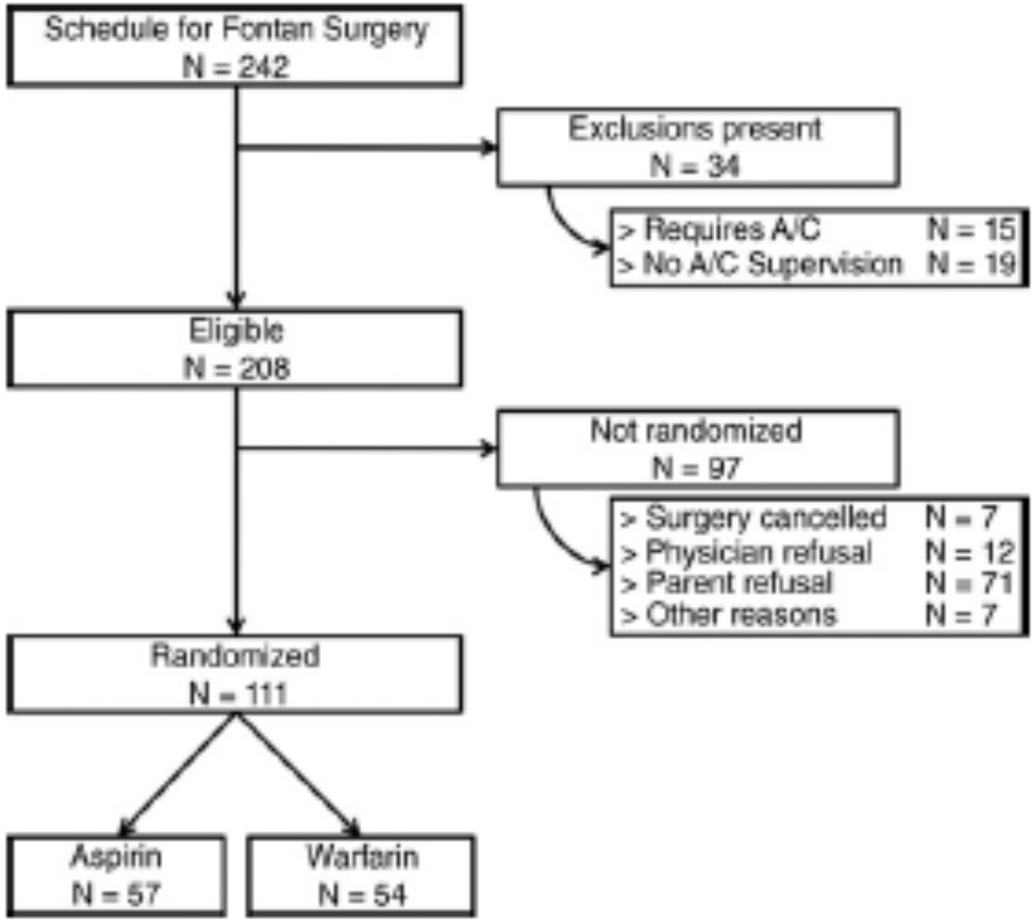
No difference for Fontan connection type

M3C

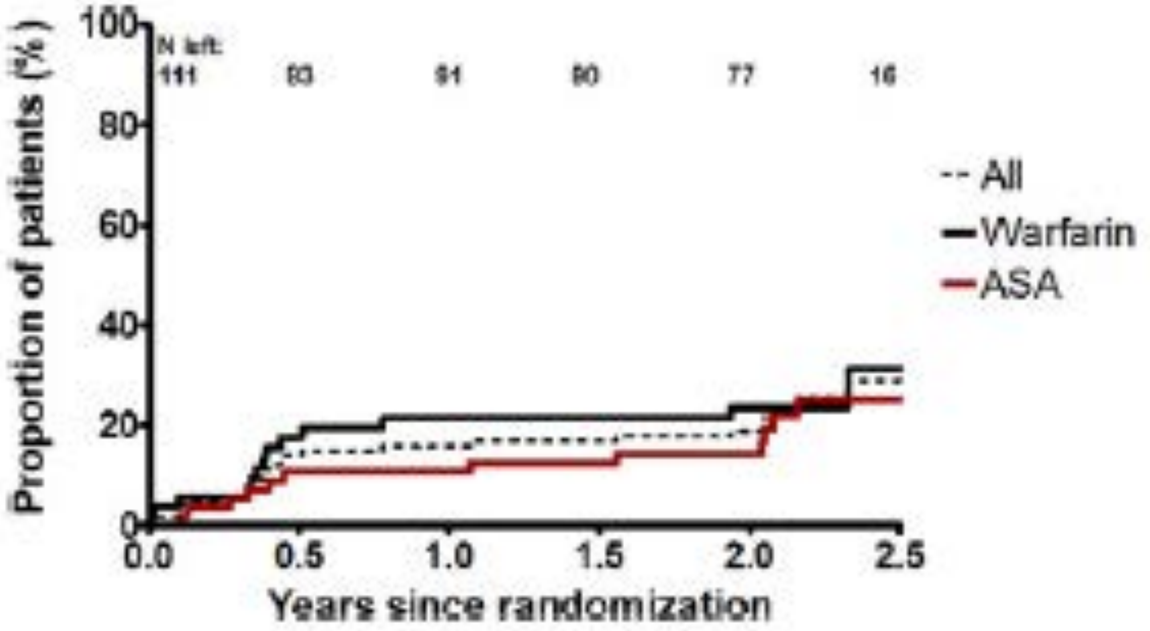
Alsaled et al. Heart 2015

A Multicenter, Randomized Trial Comparing Heparin/Warfarin and Acetylsalicylic Acid as Primary Thromboprophylaxis for 2 Years After the Fontan Procedure in Children





Thrombosis after randomization

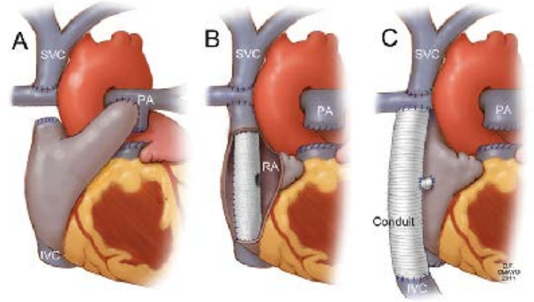


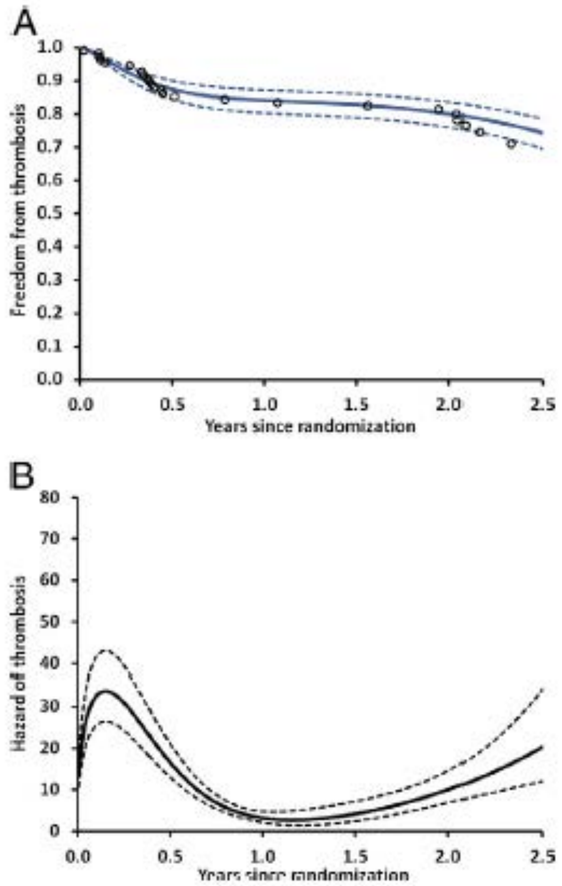
Kaplan-Meier estimate for the proportion of patients with thrombosis 2 years after randomization was 19% (24% in patients randomized to warfarin vs. 14% in patients randomized to aspirin). Hazard ratio for thrombosis for patients randomized to warfarin vs. acetylsalicylic acid (ASA) was 1.35 (95% confidence interval: 0.62 to 3.00), p 0.45.

Monagle P et al. J Am Coll Cardiol 2011;58:645-51

Factors Associated With Thrombotic Complications After the Fontan Procedure

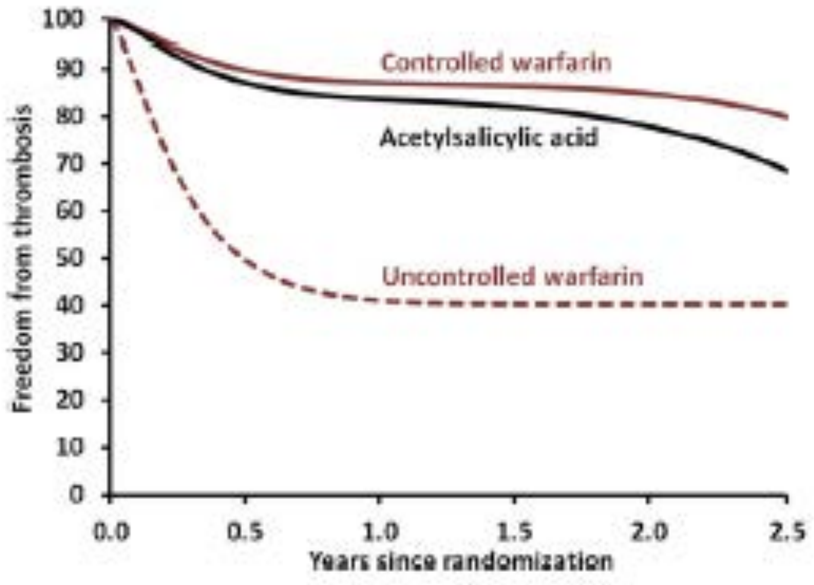
A Secondary Analysis of a Multicenter, Randomized Trial of Primary Thromboprophylaxis for 2 Years After the Fontan Procedure





Freedom From and Hazard of Thrombosis Over Time in Patients After Fontan Surgery

Time-related freedom from thrombosis was 69% (all venous, no arterial events), with 28% of thrombosis presenting with clinical signs or events



Freedom From Thrombosis Over Time Stratified by Thromboprophylaxis Choice and Effectiveness

McCrindle BW et al. J Am Coll Cardiol 2013;61:346-53

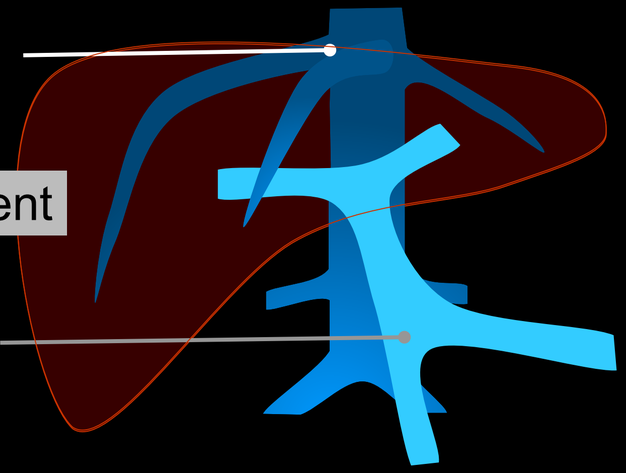
Liver and Fontan

6	6	17
4	15	4
10	21	21
NI		TCPC

Hepatic V

Gradient

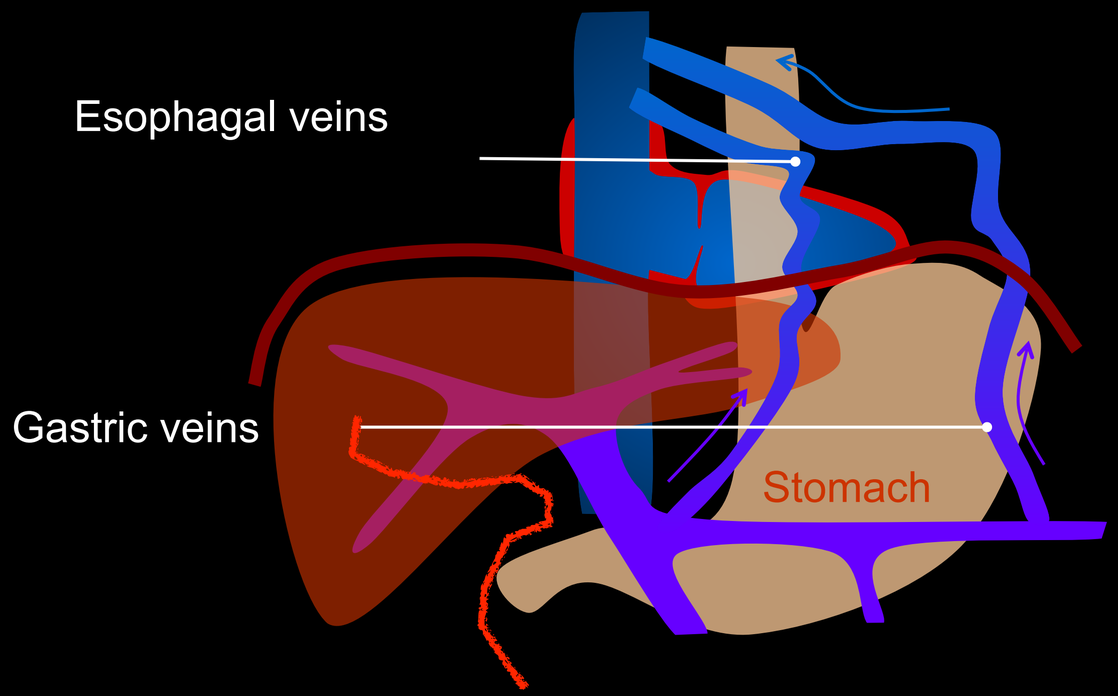
Portal V.

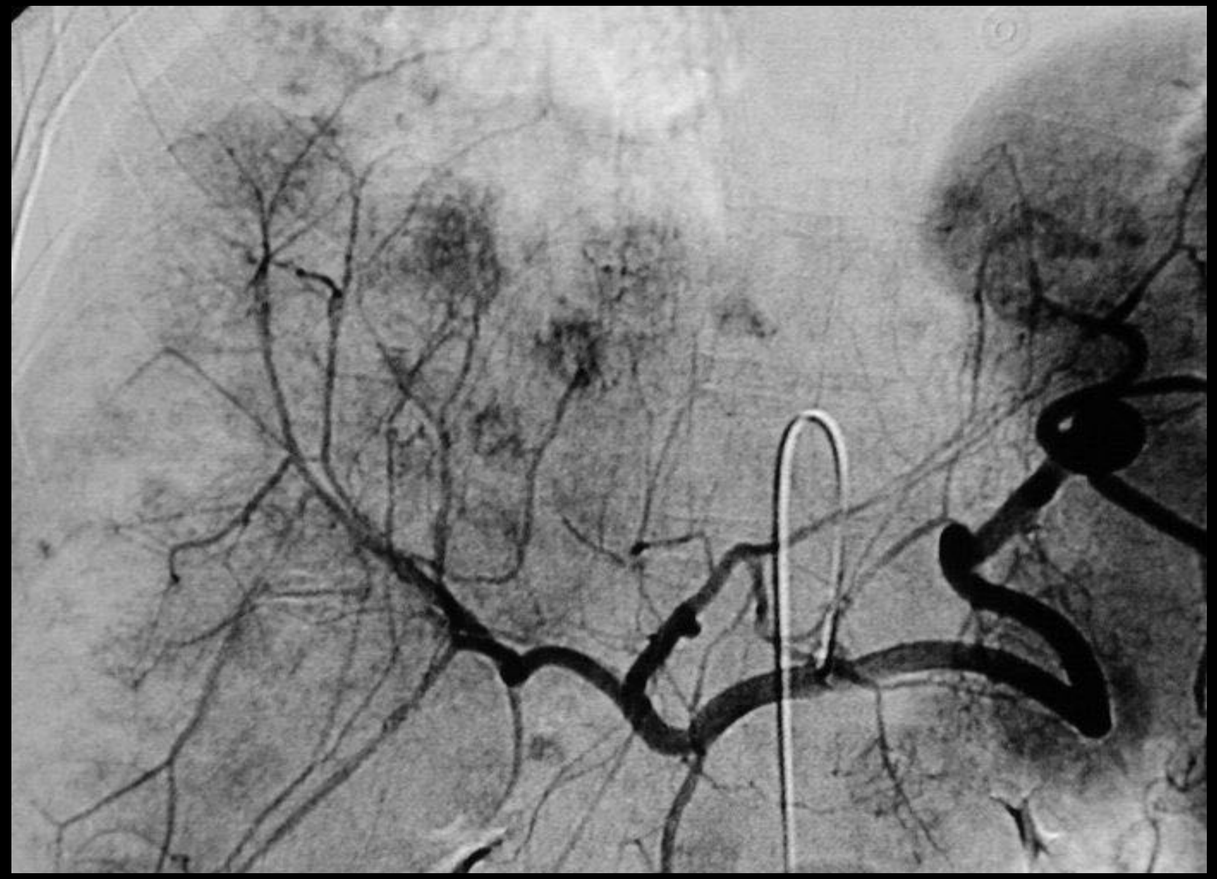


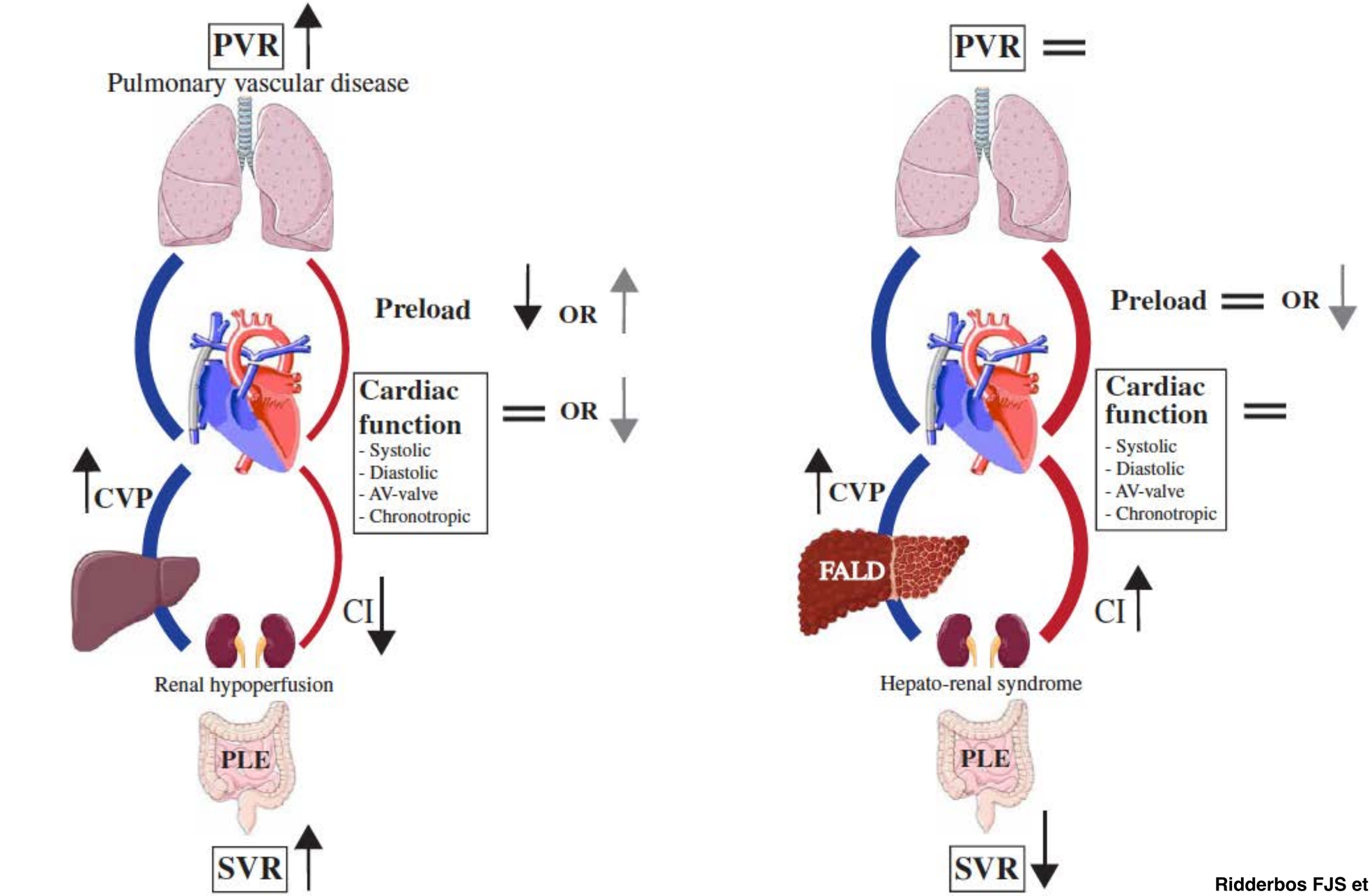
Portal Hypertension

Kiesewetter Heart 2007

Liver and Fontan



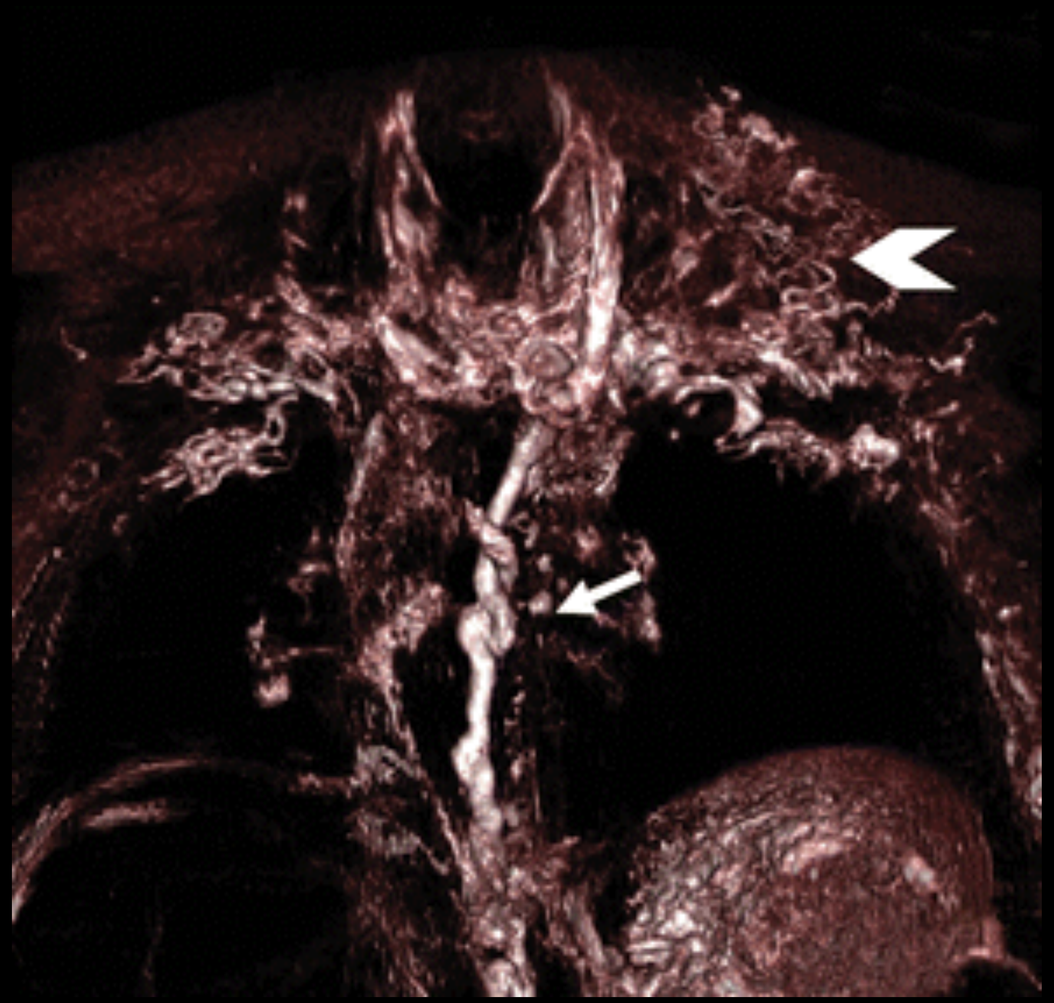




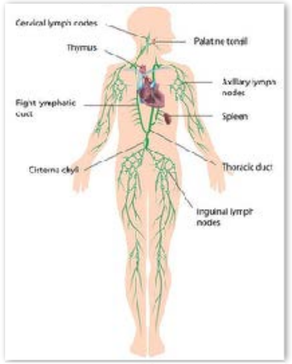
Ridderbos FJS et al. Eur Heart J 2019

The lymphatics in Fontan circulation

Bronchial casts



Lymphatics



T2 imaging of lymphatic circulation

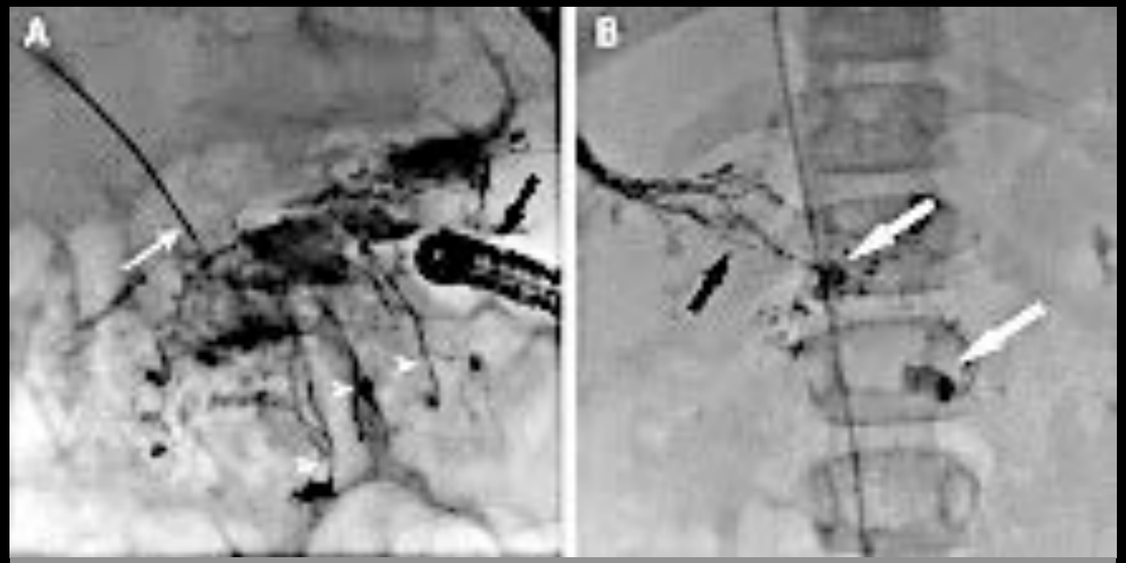
The new horizon for the treatment of PLE/bronchial casts

Lymphangectasia collaterals

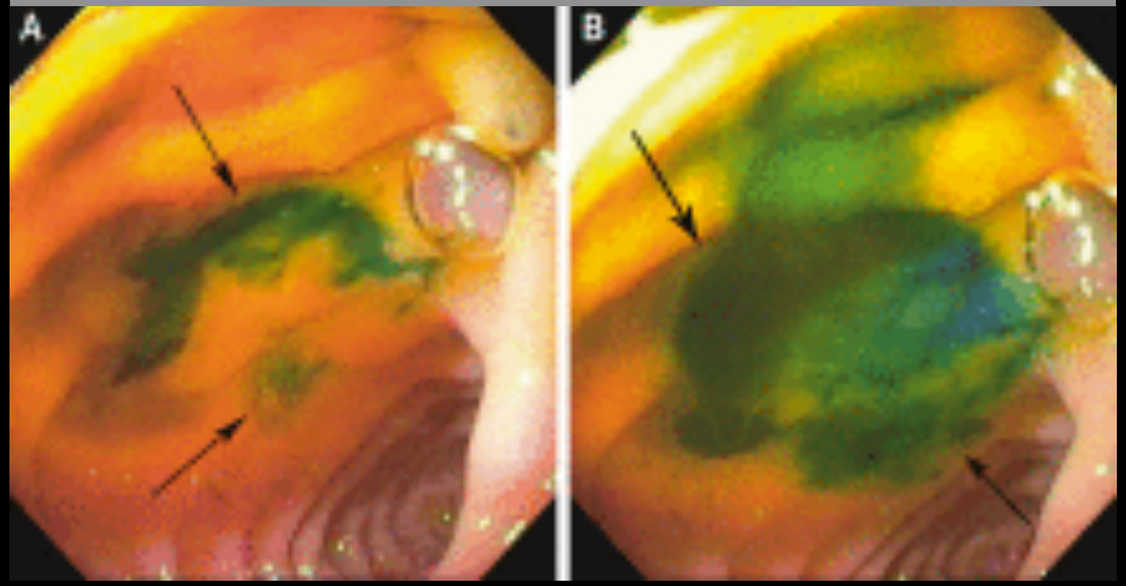
Thoracic duct dilation & tortuosity

Tissue edema

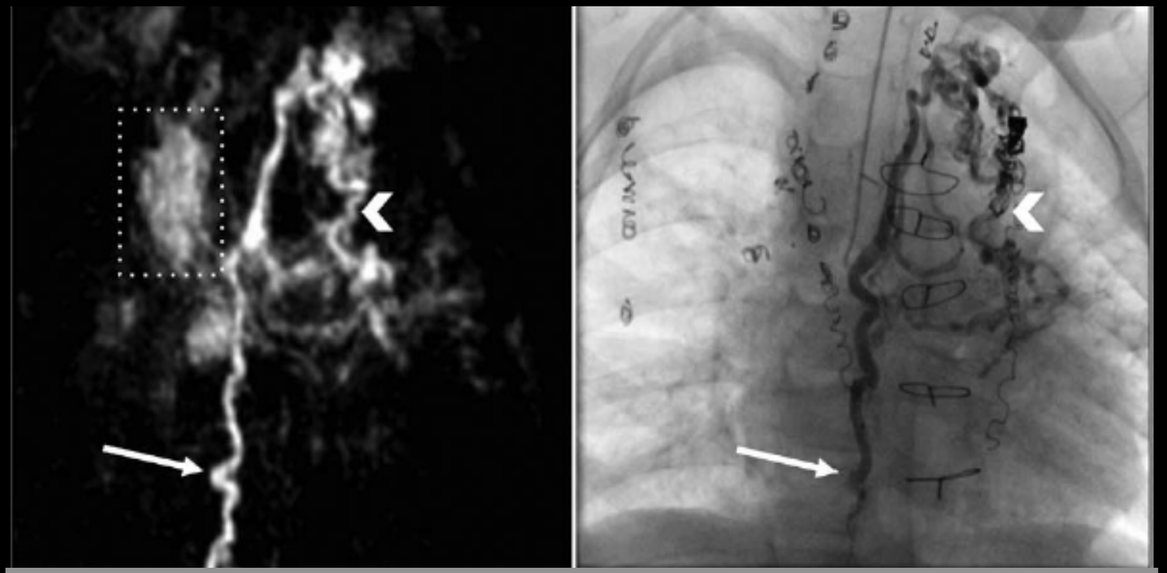
Dori Y et al. AJR 2014;203:426



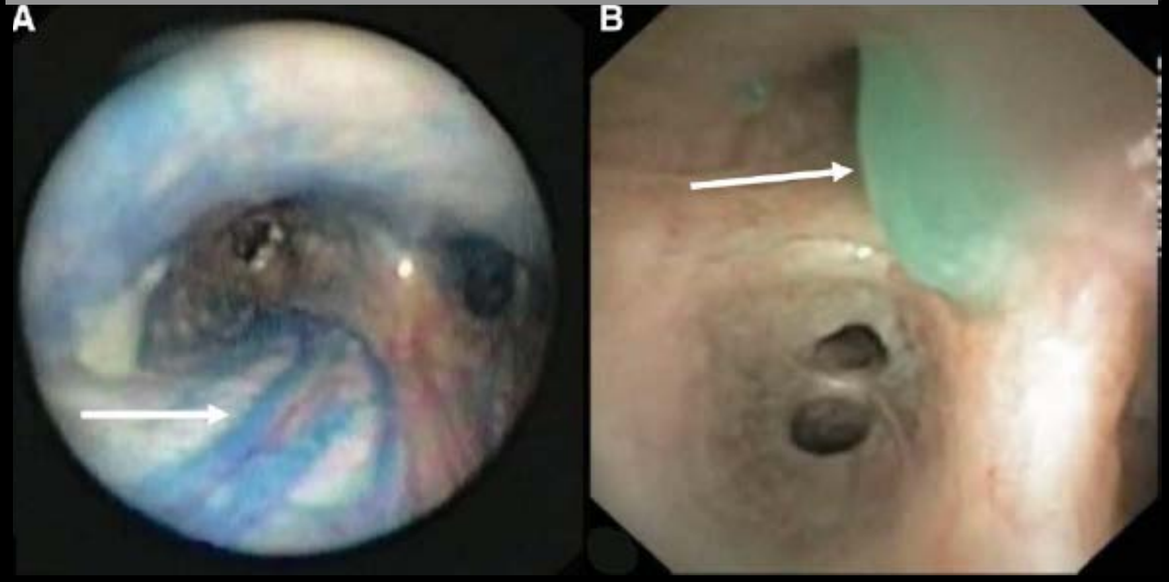
PLE Hepatic lymphogram & endoscopy



Itkin M et al. JACC 2017



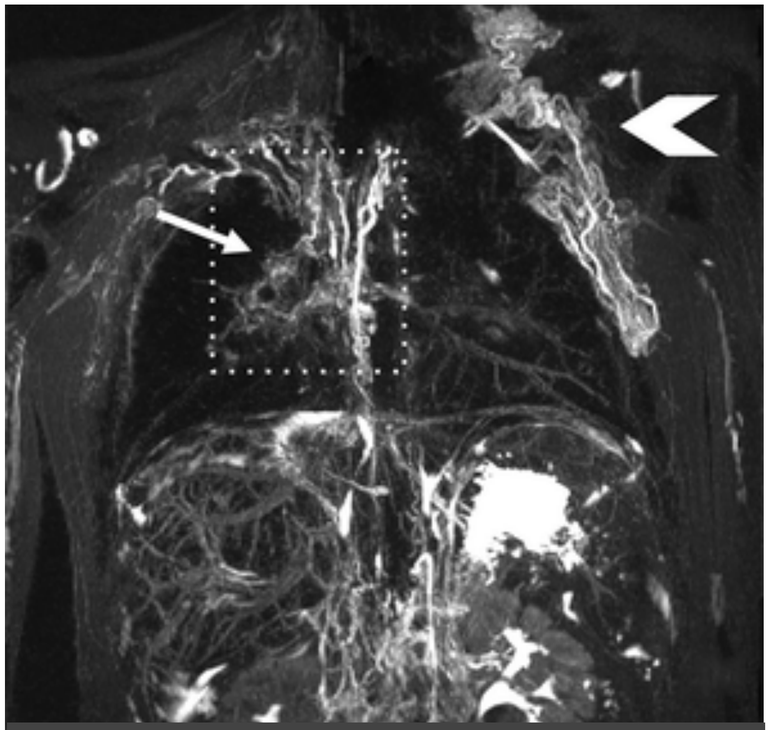
Plastic bronchitis lymphogram & bronchoscopy



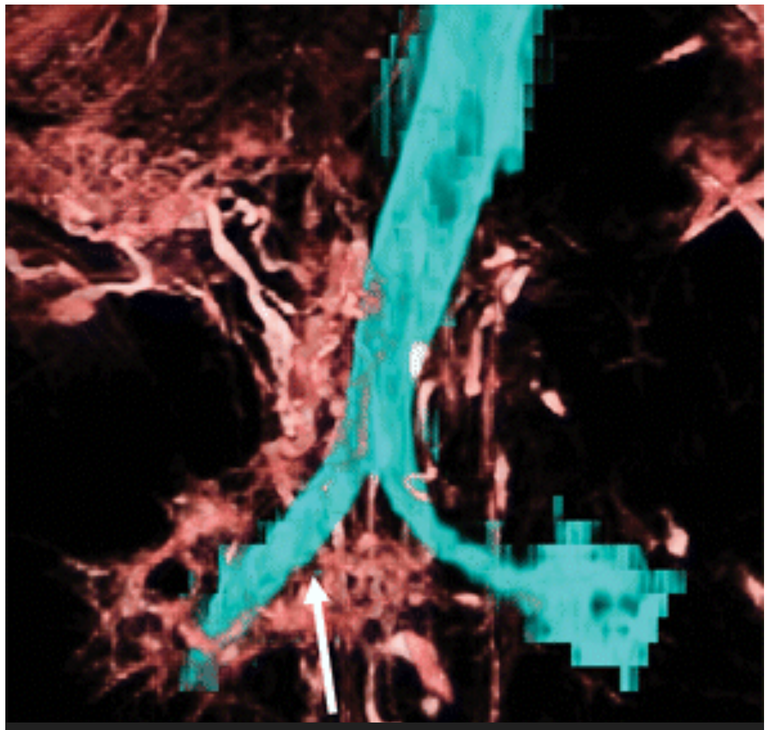
Dori Y et al. Circulation 2016

Lymphatic interventional catheterization

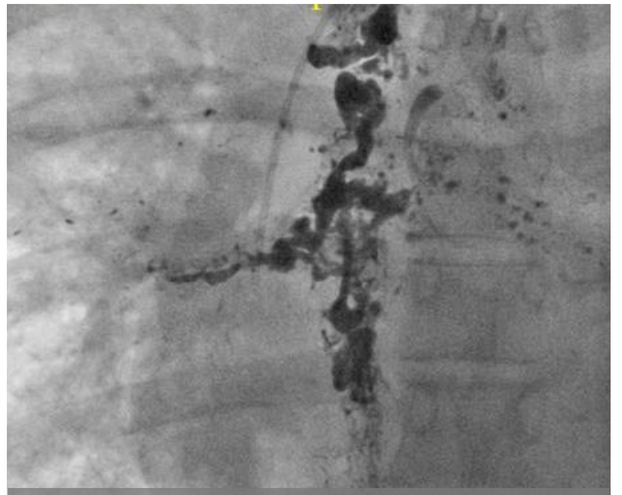
The new horizon for the treatment of PLE/bronchial casts



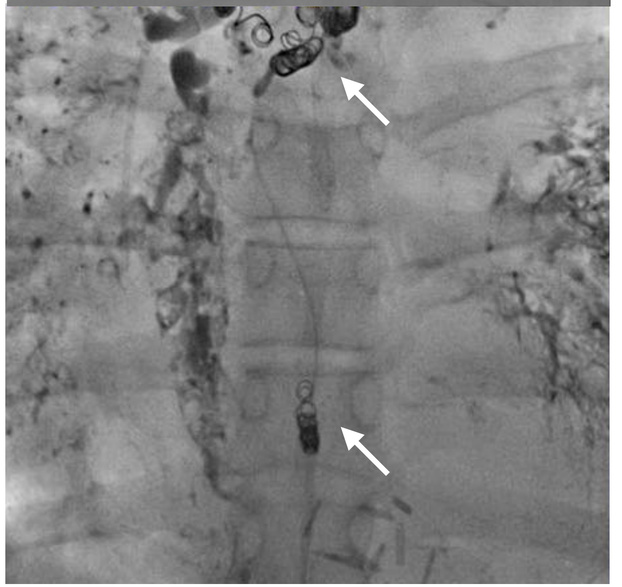
Leak in right lung



Leak into right bronchus



Embolisation coils & glue

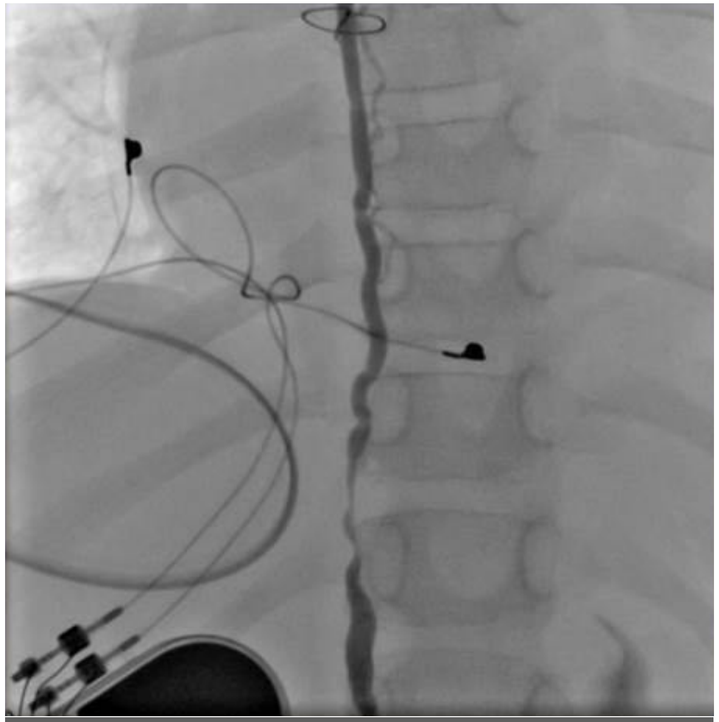


15 years TCPC, plastic bronchitis

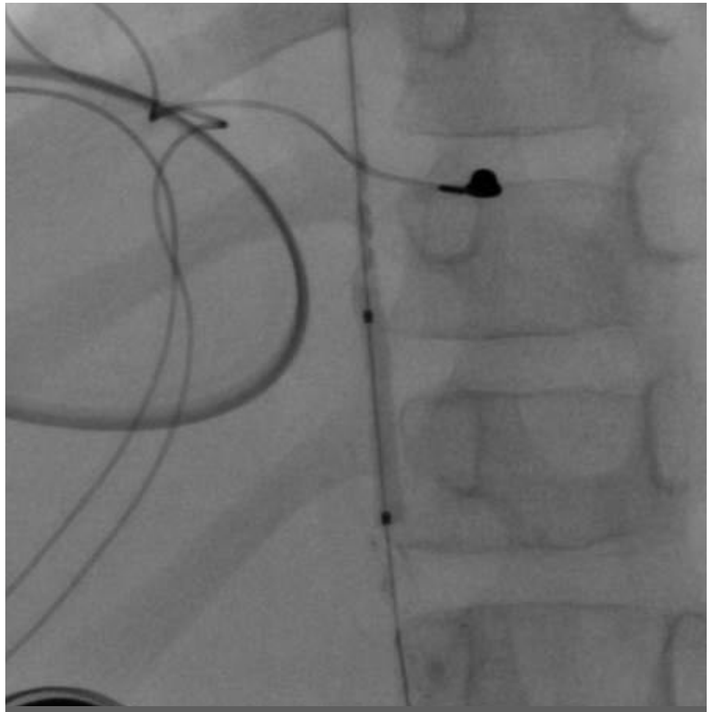
Dori Y et al. AJR 2014;203:426

Lymphatic interventional catheterization

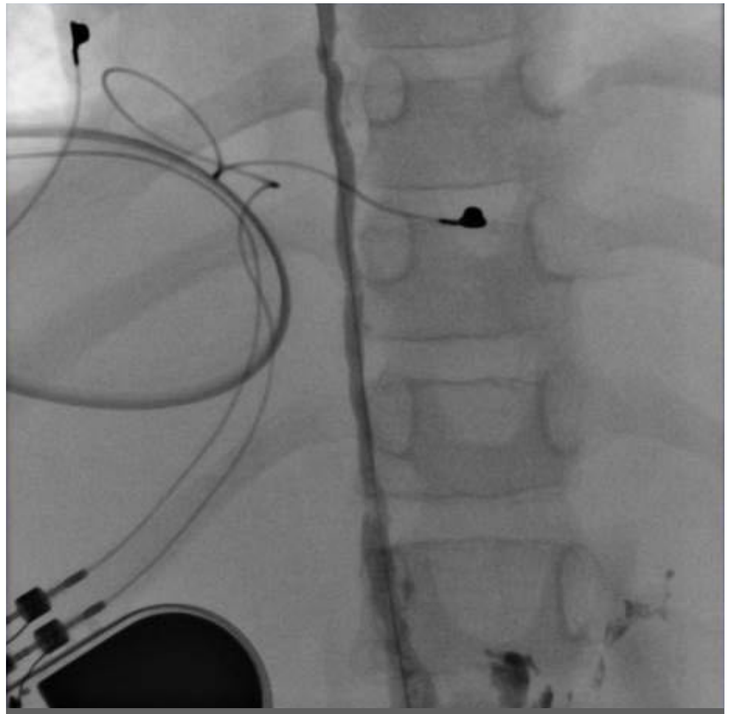
The new horizon for the treatment of PLE/bronchial casts



Stenosis of the thoracic duct



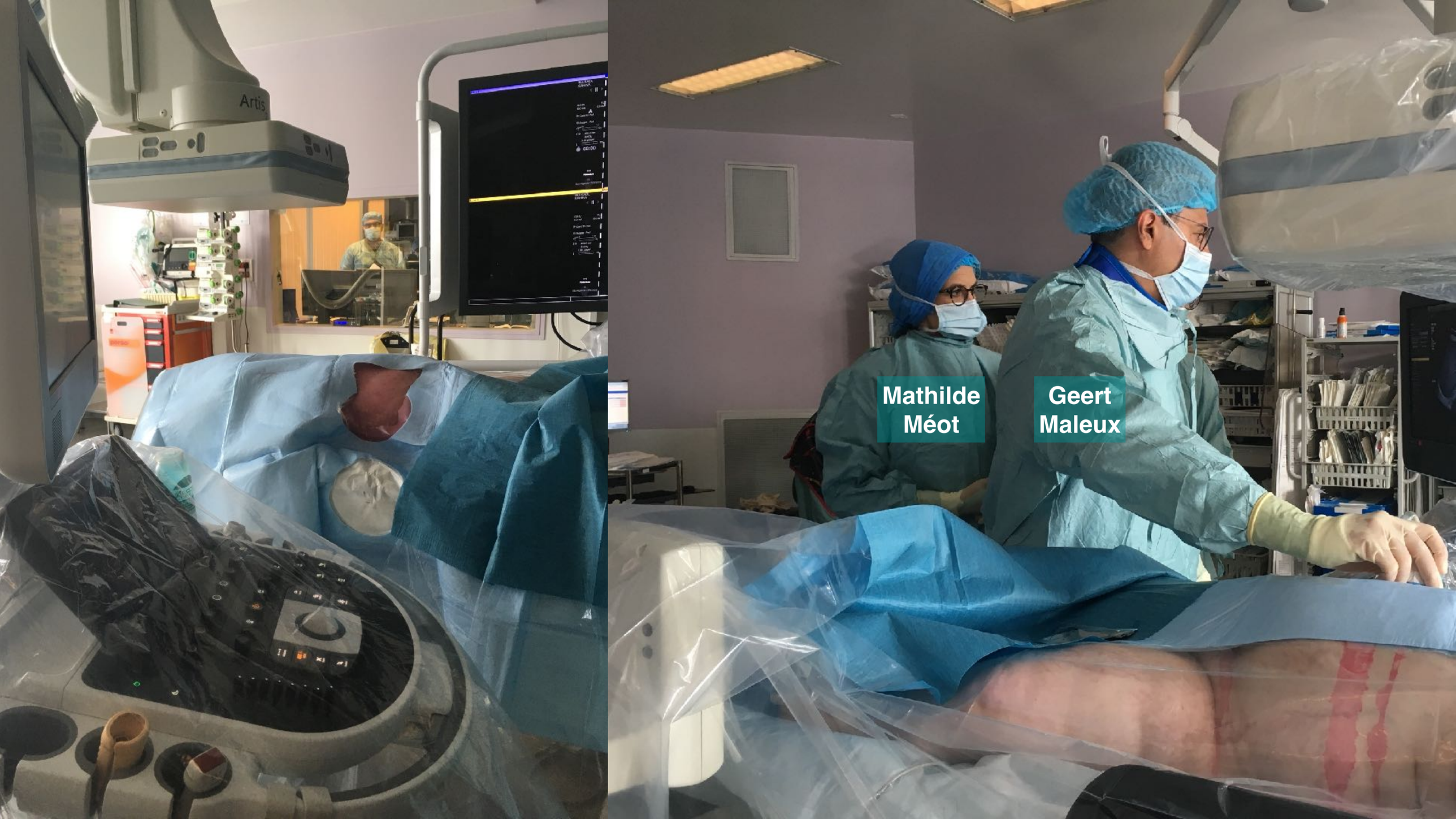
Balloon dilatation
of the thoracic duct



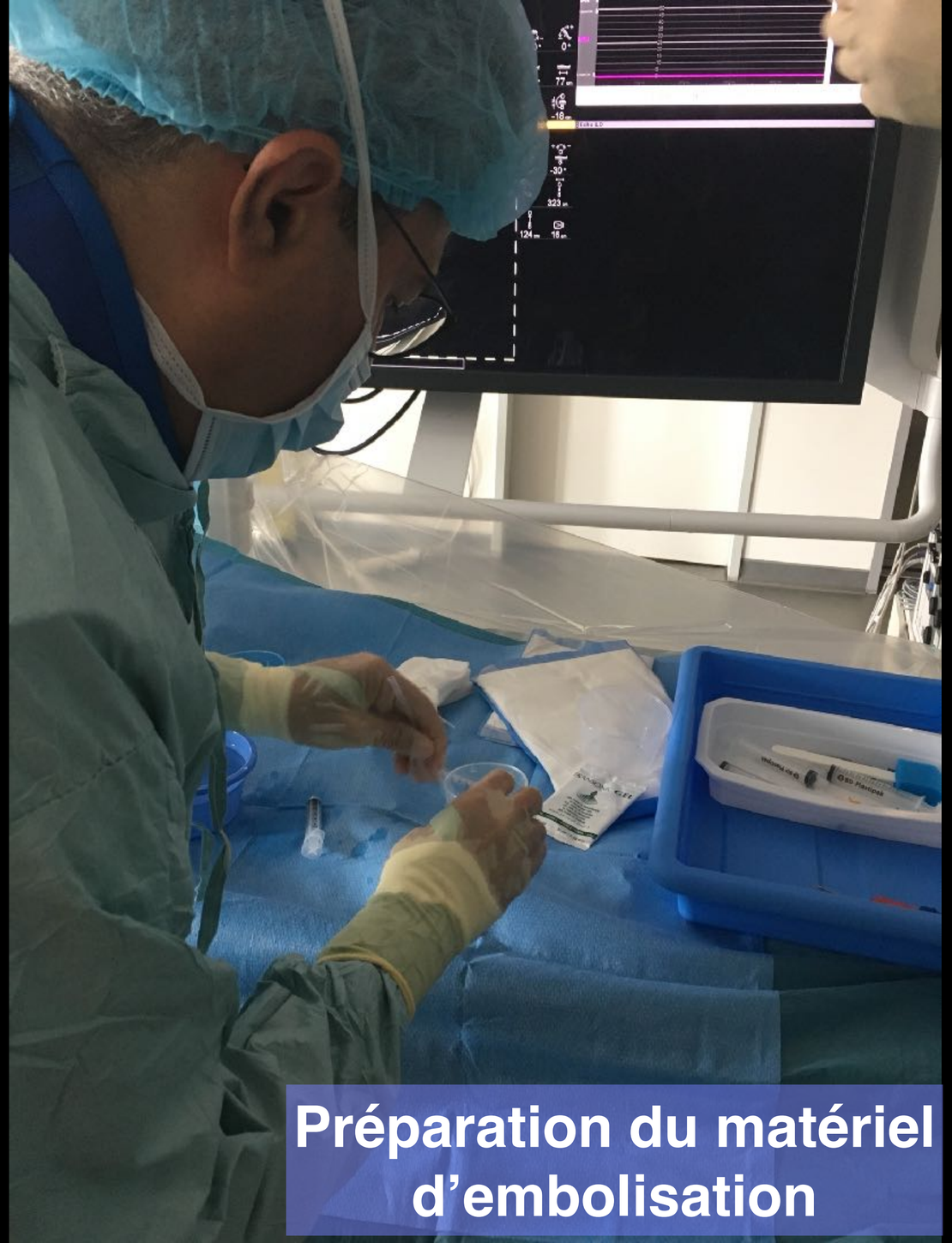
Control after dilatation
of the thoracic duct

23 years TCPC, PLE

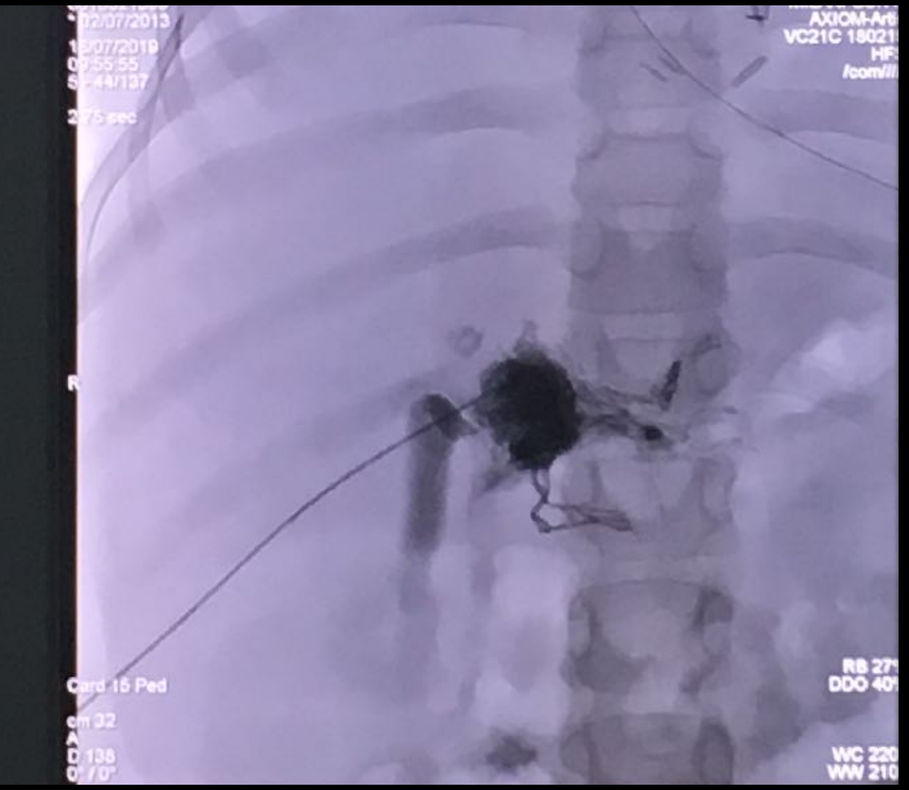
Dori Y et al. AJR 2014;203:426

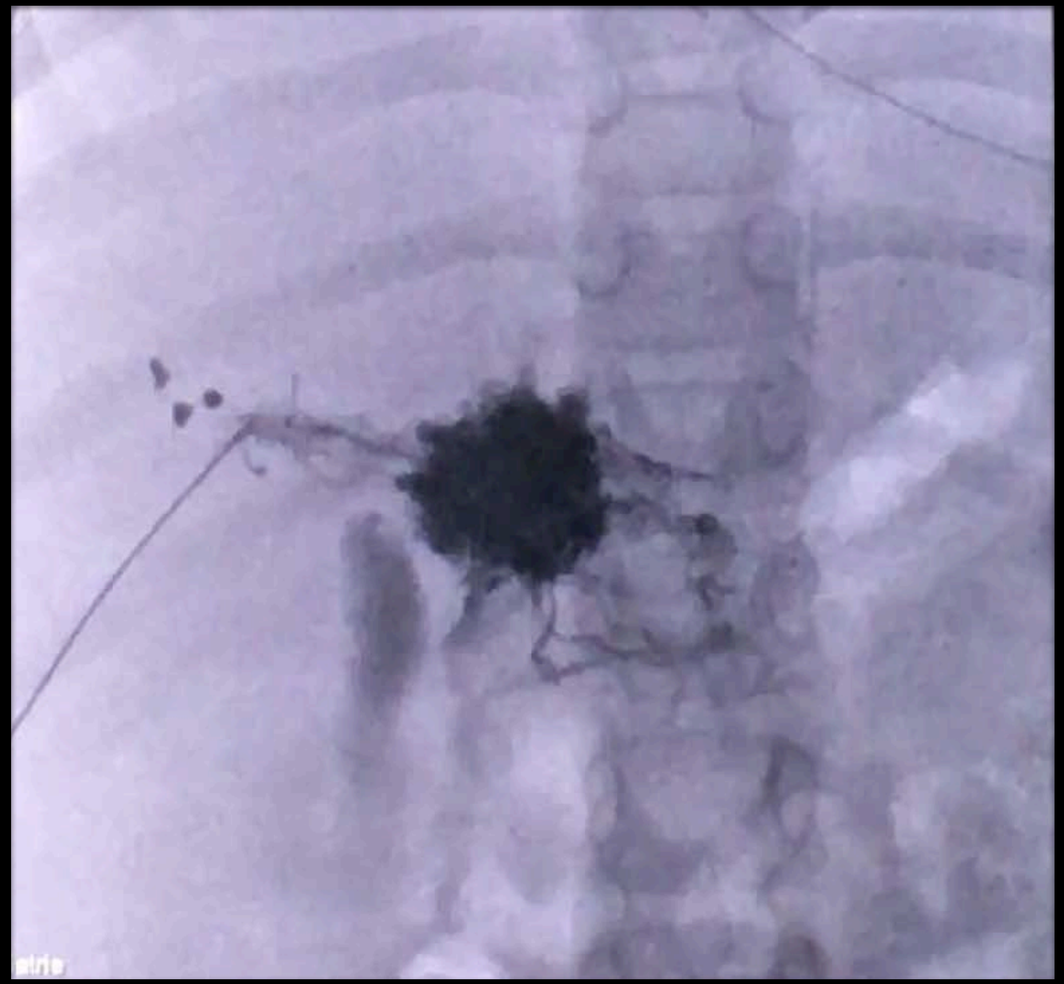

Artis
Mathilde Méot
Geert Maleux

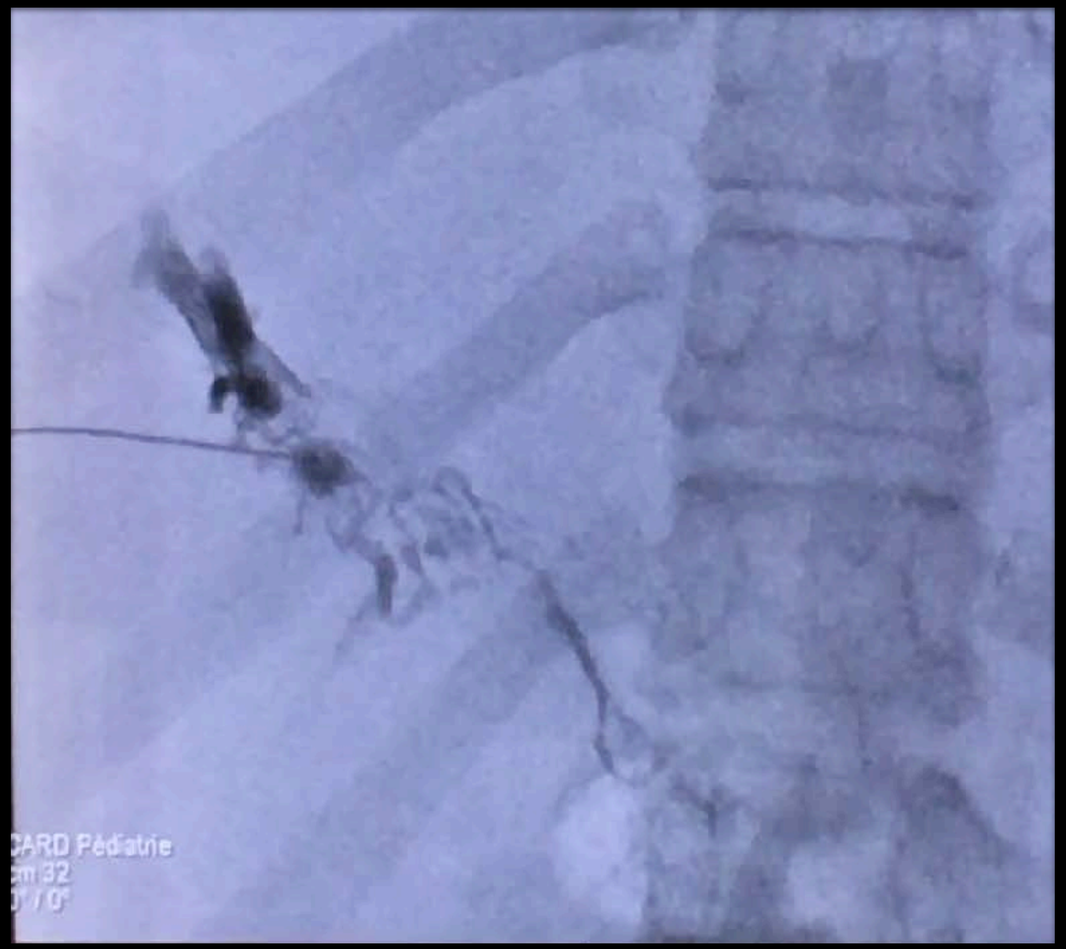
Ponction échoguidée
de l’espace périportal

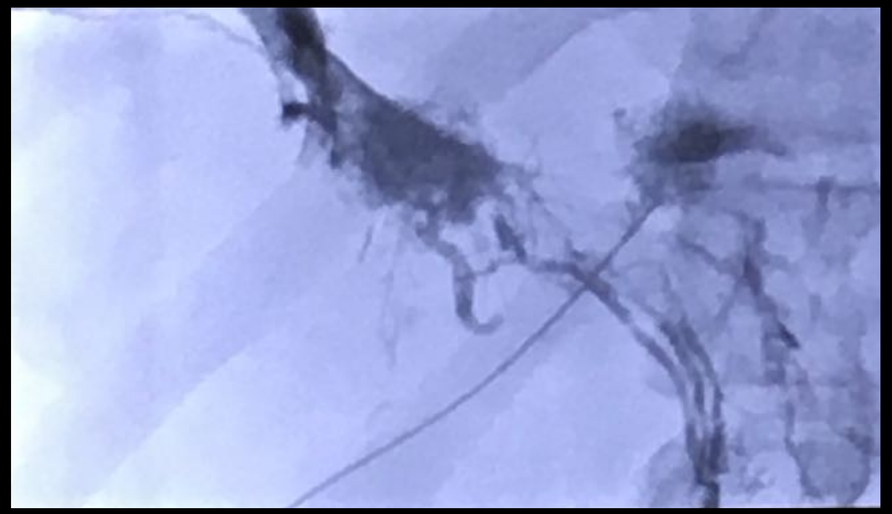


Préparation du matériel
d’embolisation


AXIOM-Arti
Card 15 Ped




CARD Pédiatrie



1-Sequentially phenotyping patients is the key to define the baseline and follow-up predictors of outcome

2-Phenotyping patients and building large registries are the keys to identify real surrogates (therapeutic targets)

3-Treatment of heart failure in Fontan circulation uses different paradigms than for HF with reduced EF

4-Thromboprophylaxis should be given to ALL patients and if given appropriately monitored

5-The « lympho-cardiology imaging/intervention » is the new horizon to improve PLE/bronchial casts

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

Individual experience is the brake
Indifference is the weakness
Authority argument is the threat

Thank you

Collective ignorance is the motivation
Curiosity is the strength
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Individual experience is the brake
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