Left outflow tract obstructions

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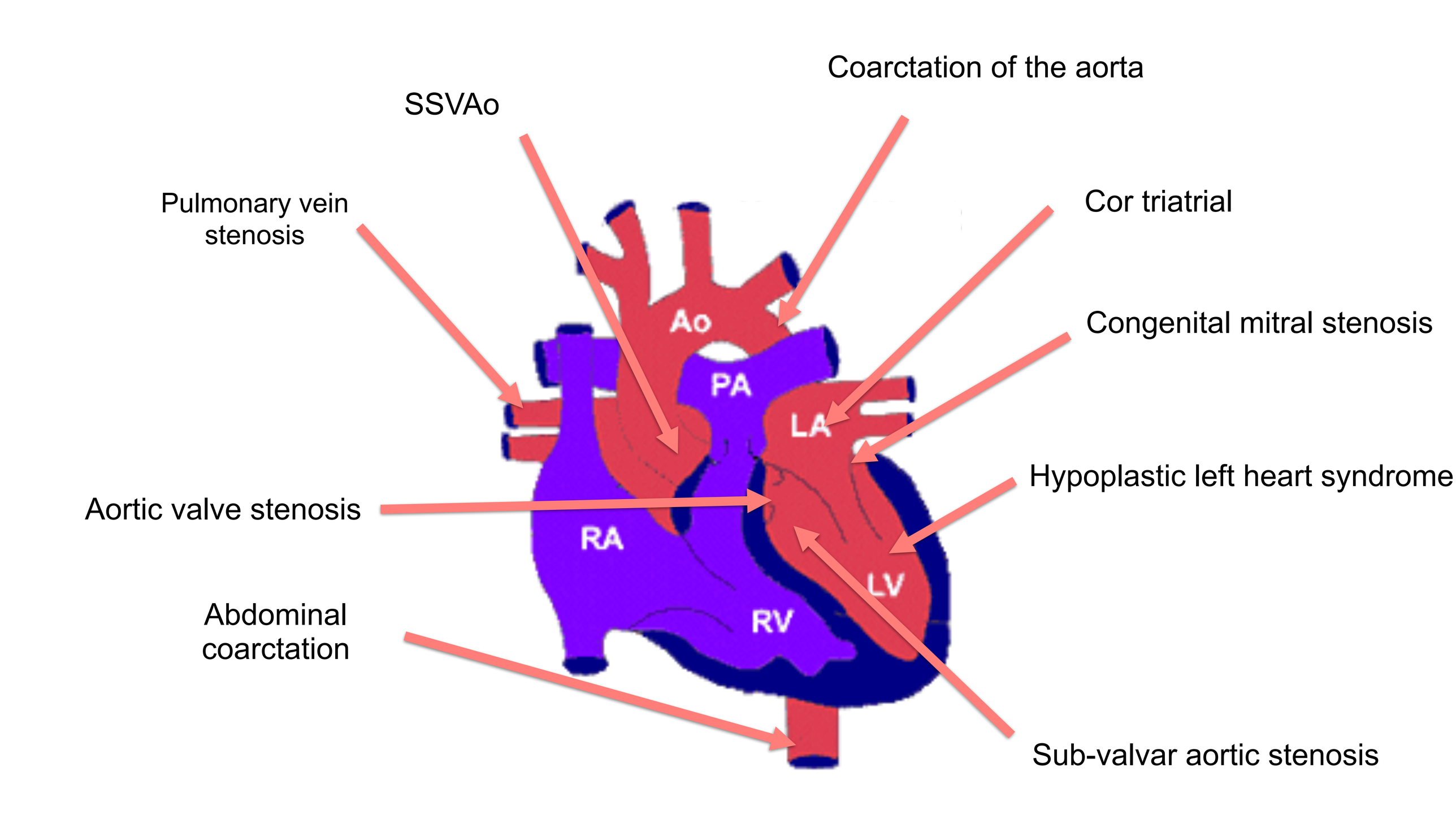
IcarP Cardiology, Institut Hospitalo-Universitaire IMAGINE

Centre de Référence Maladies Rares
Malformations Cardiaques Congénitales Complexes-M3C

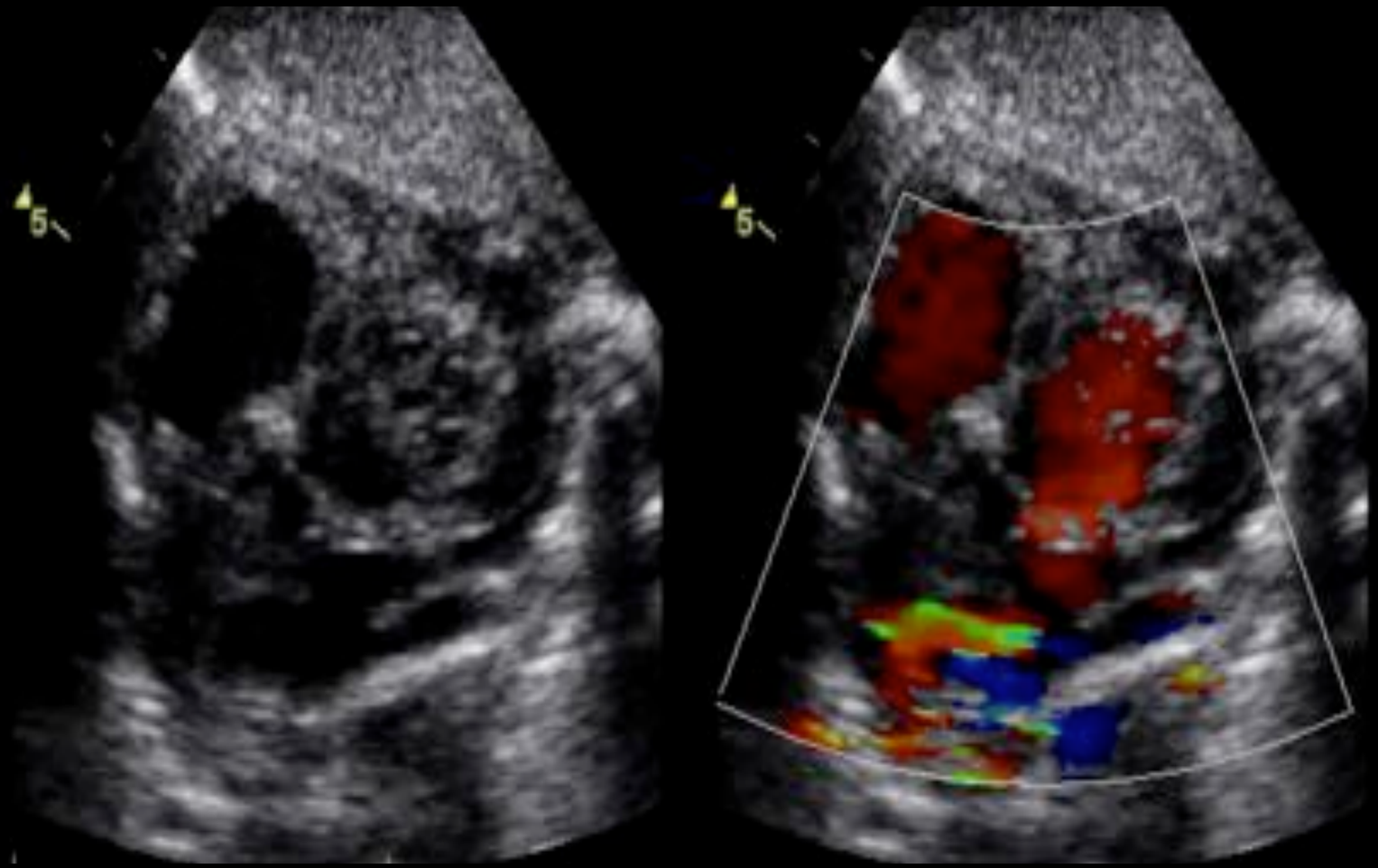
Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN


Coarctation of the aorta
SSVAo
Pulmonary vein stenosis
Cor triatrial
Congenital mitral stenosis
Hypoplastic left heart syndrome
Aortic valve stenosis
Abdominal coarctation
Sub-valvar aortic stenosis
Ao
PA
LA
RA
LV
RV

Pulmonary veins stenosis

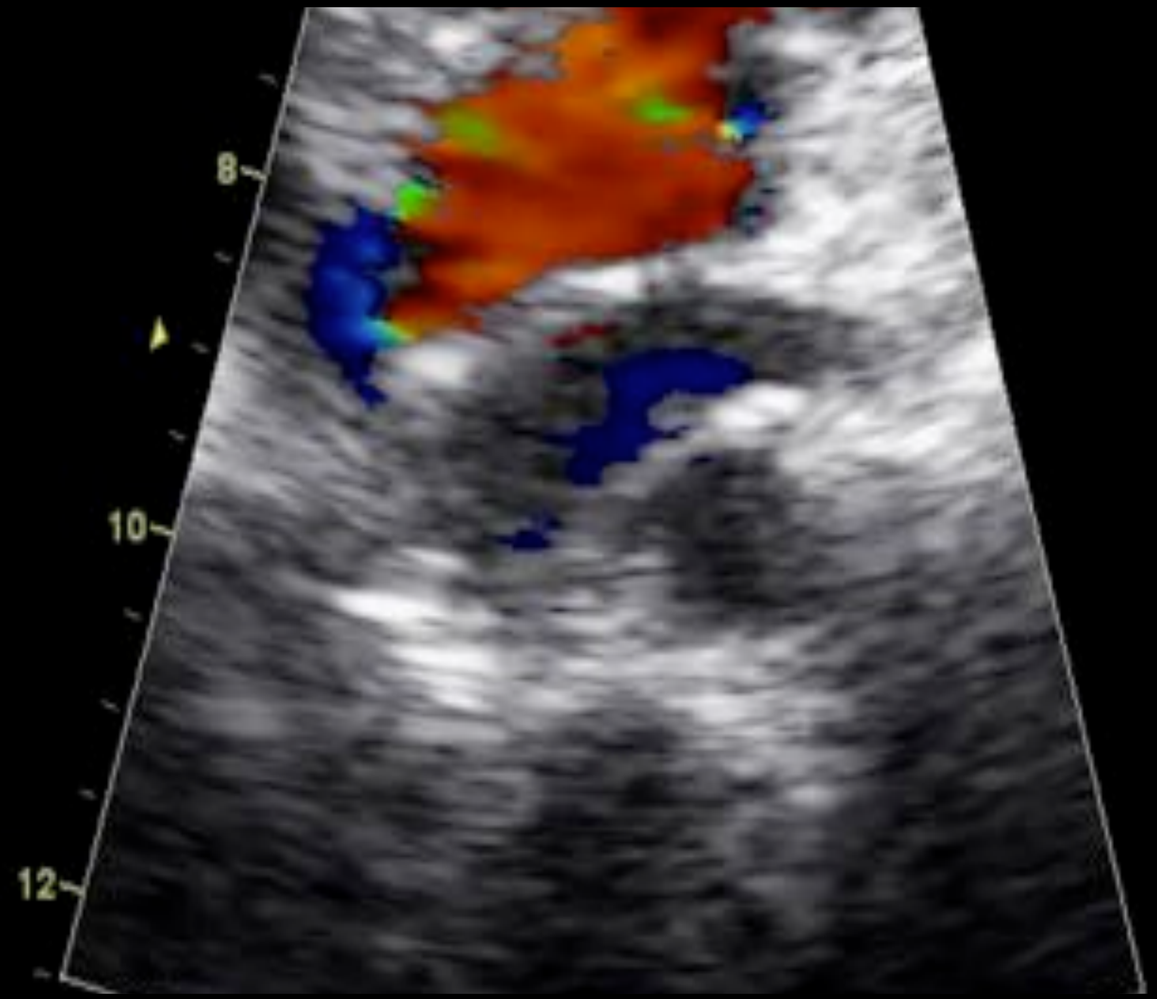
Sténose des veines pulmonaires

Collecteur RVPAT anastomose large postopératoire



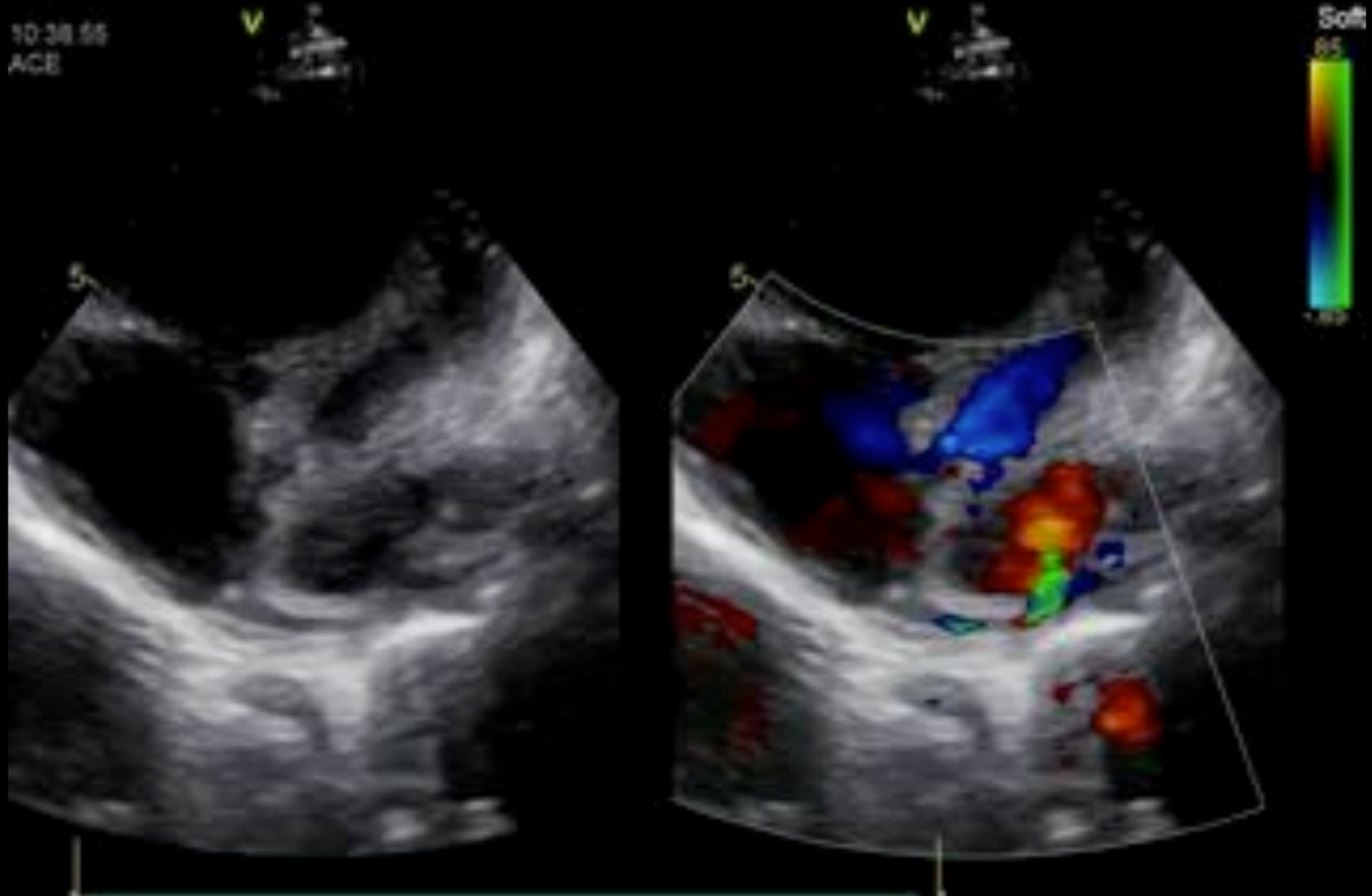
Sténose des veines pulmonaires

Collecteur RVPAT anastomose sténosée en postopératoire



Sténose des veines pulmonaires

Sténose isolée d’une veine pulmonaire



Sténose des veines pulmonaires

Sténose après ablation de fibrillation auriculaire

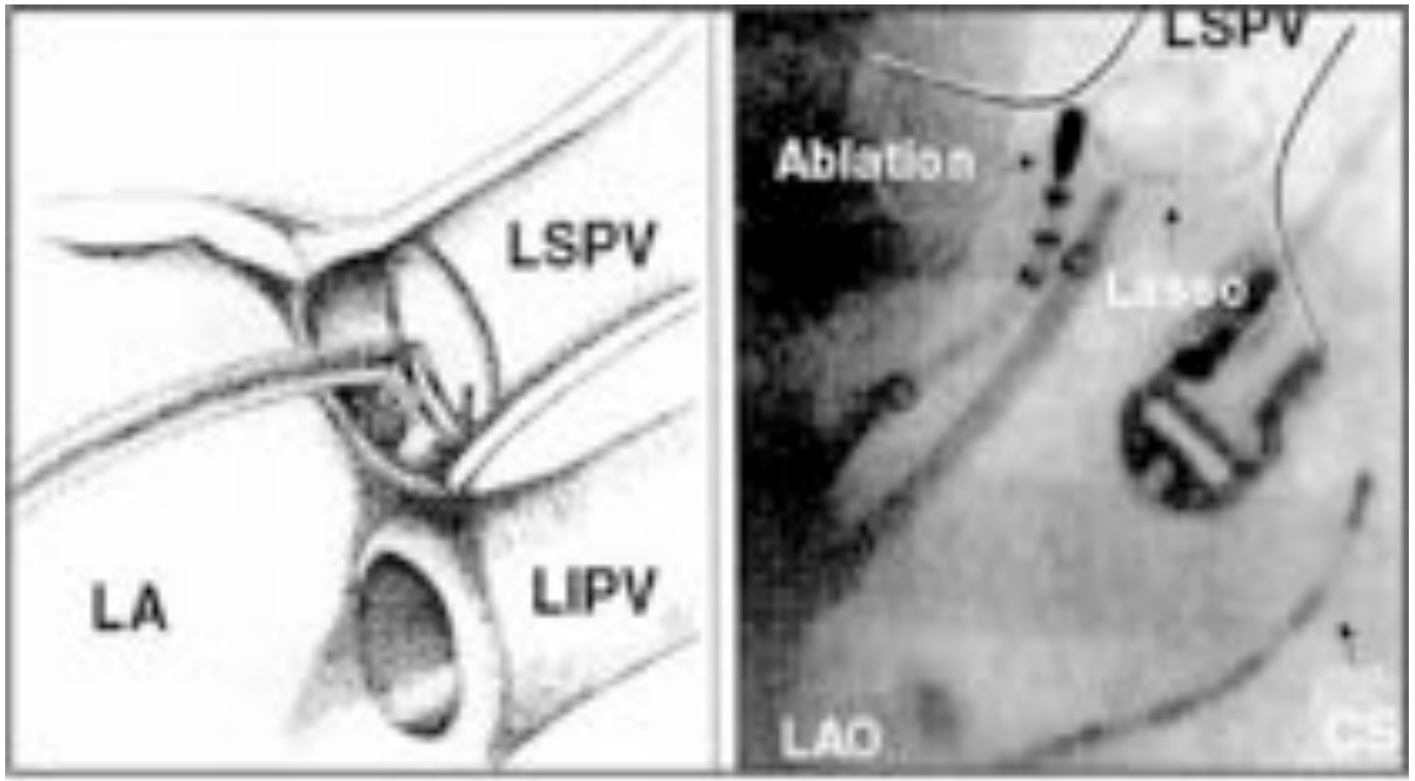
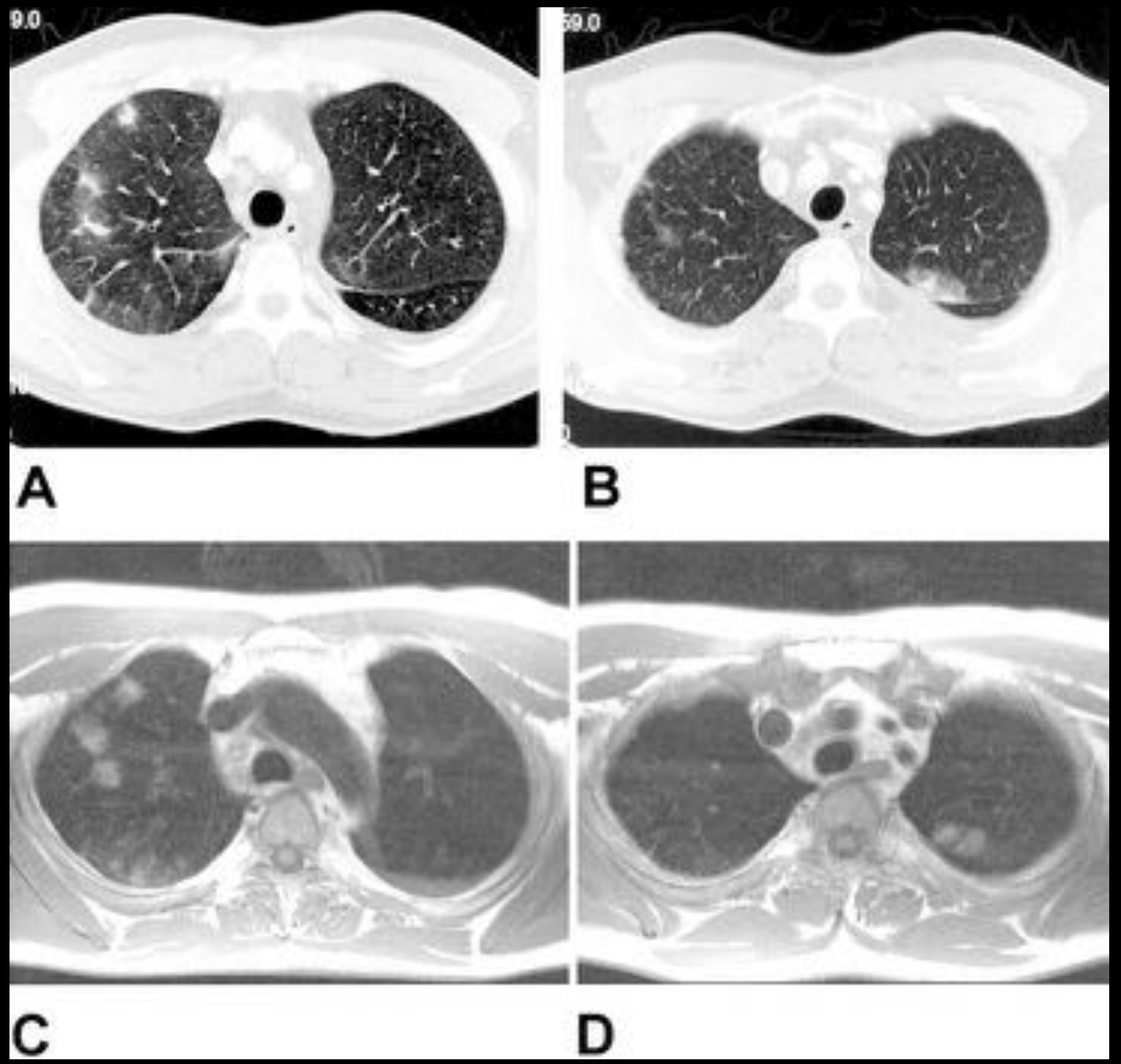


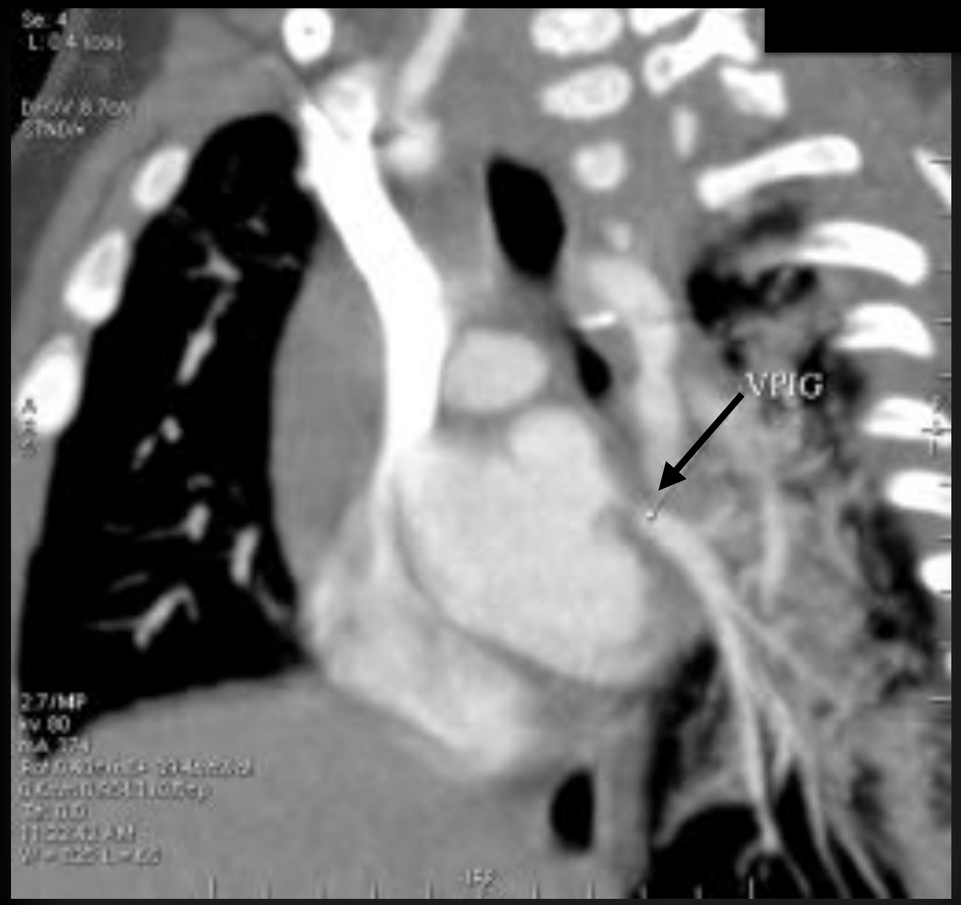
Figure 1 - Pulmonary vein isolation technique to treat paroxysmal atrial fibrillation. A) Representation of catheter position in the left superior pulmonary vein ostium (LSPV). B) Left anterior oblique (LAO) fluoroscopic projection during LSPV isolation. Note radiofrequency ablation catheter at the LSPV ostium. LA - left atrium; LIPV - left inferior pulmonary vein; CS - coronary sinus catheter.

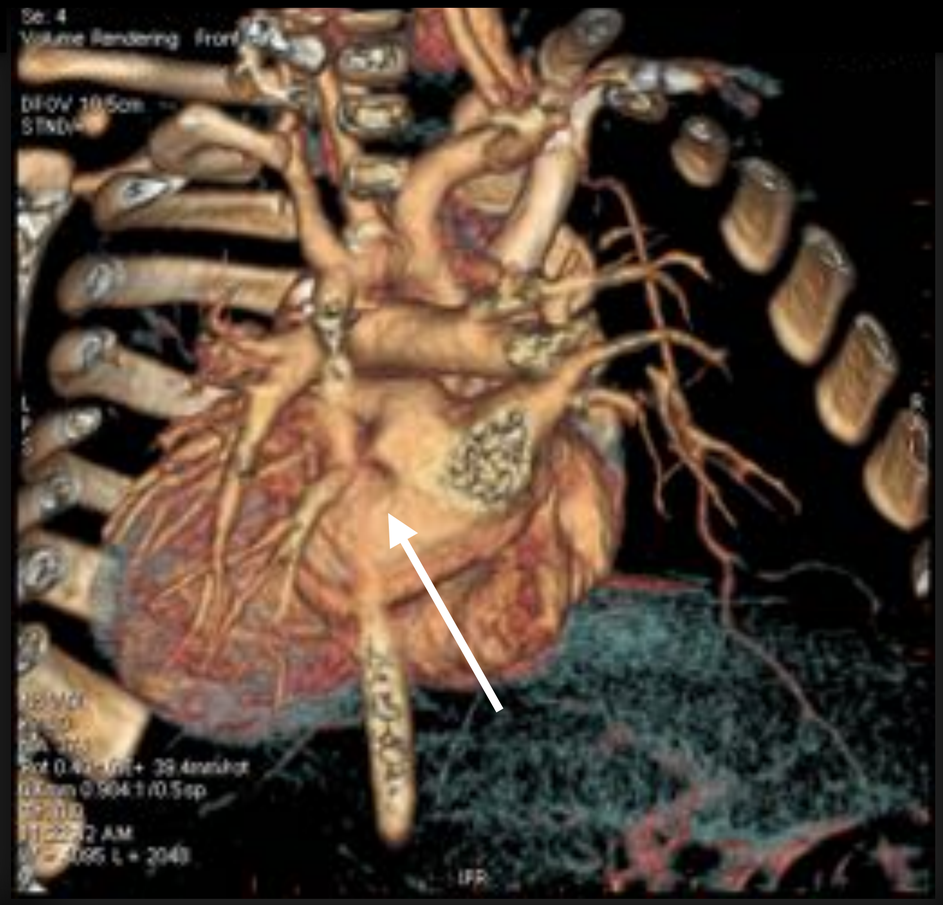
Sténose des veines pulmonaires

Aspect du parenchyme pulmonaire



Sténose des veines pulmonaires

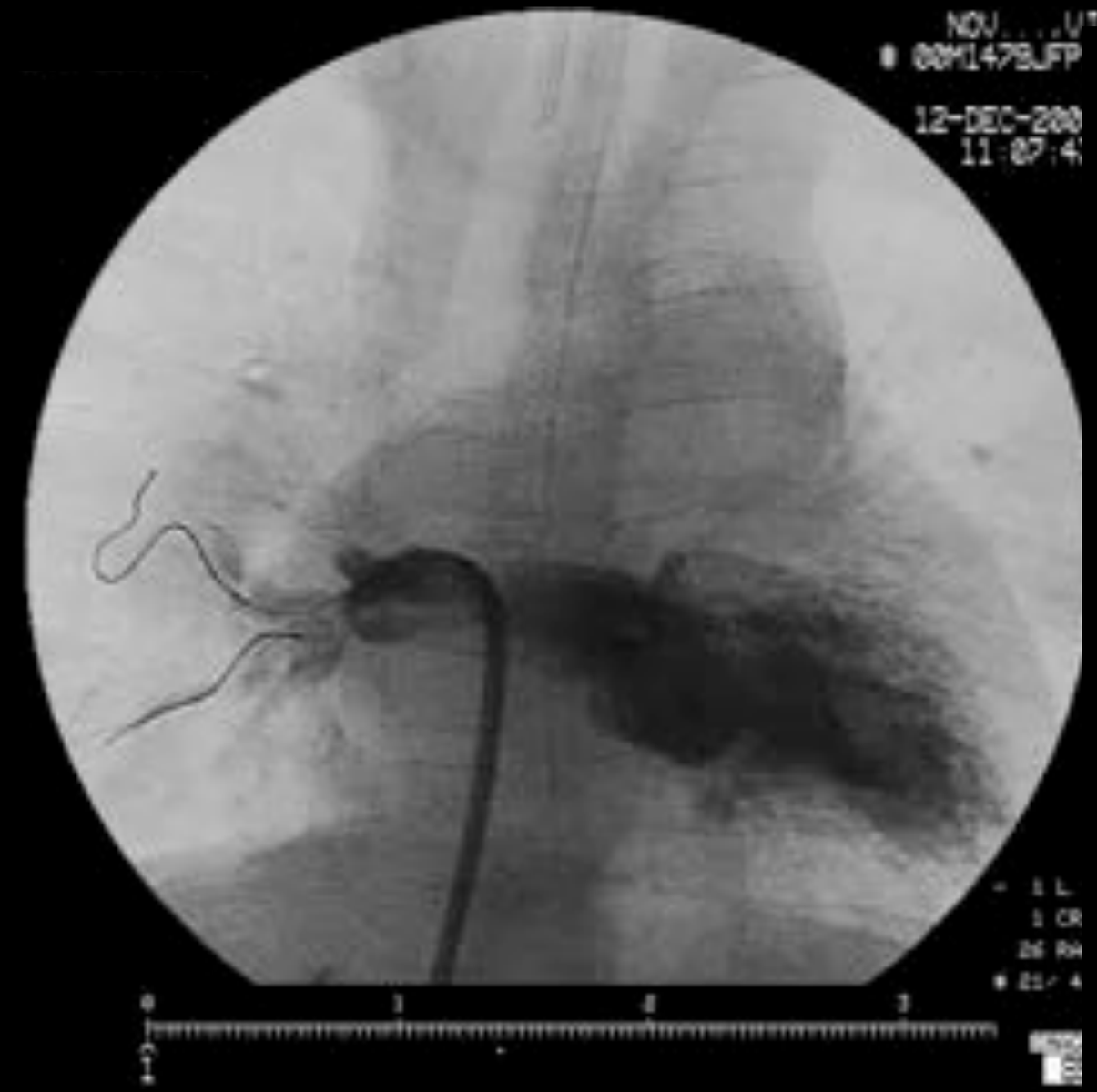




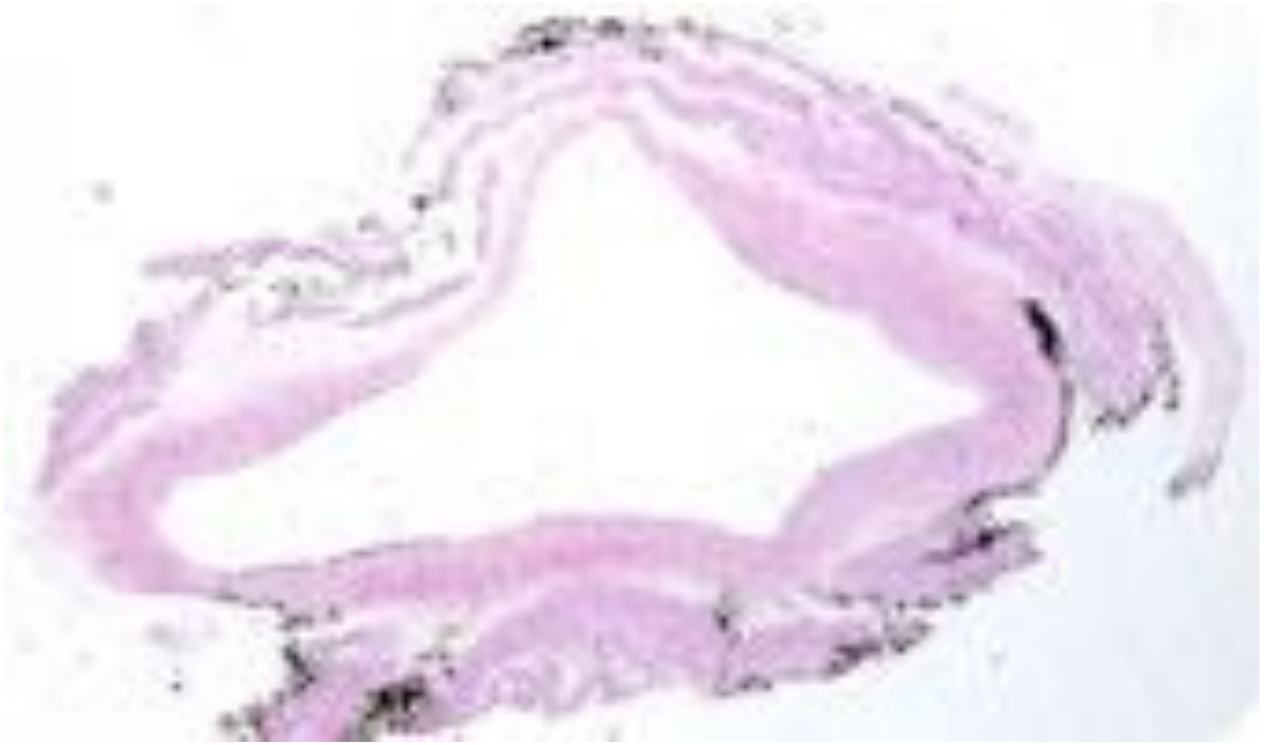
Sténose des veines pulmonaires

Sténose des veines pulmonaires

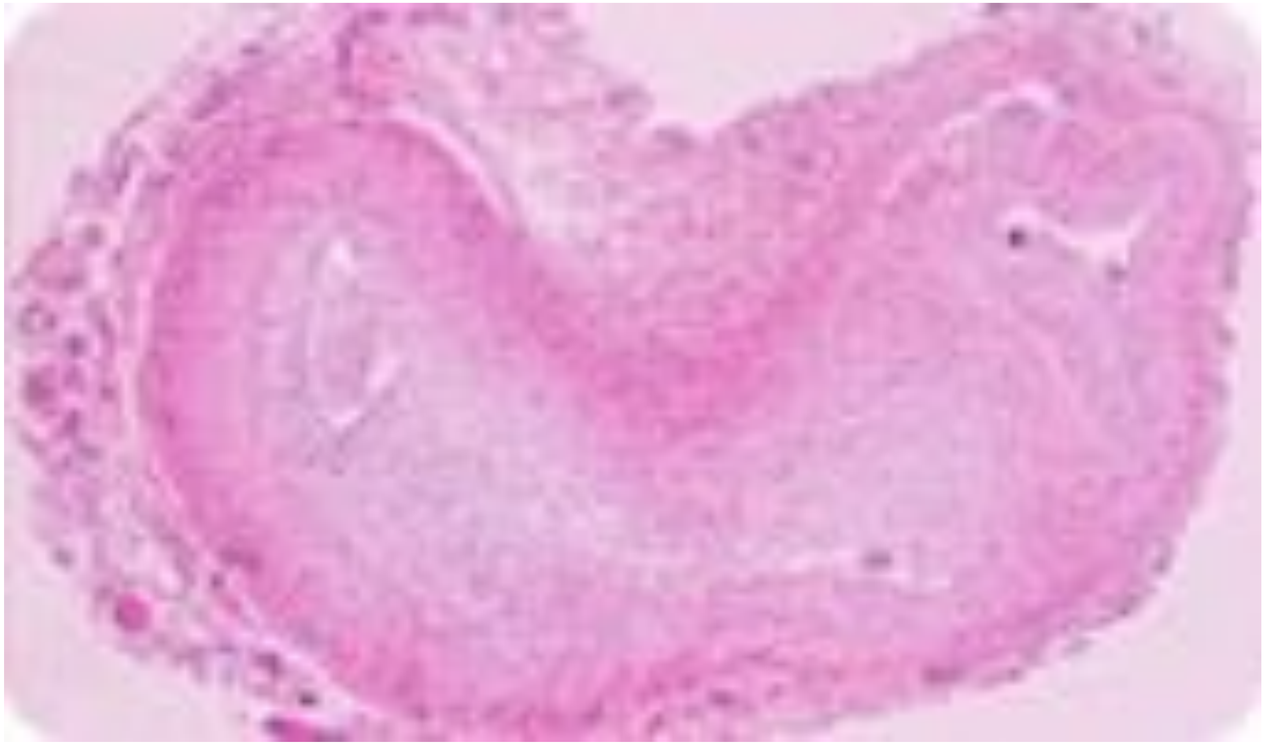
Sténose des veines pulmonaires



Sténose des veines pulmonaires
Histologie



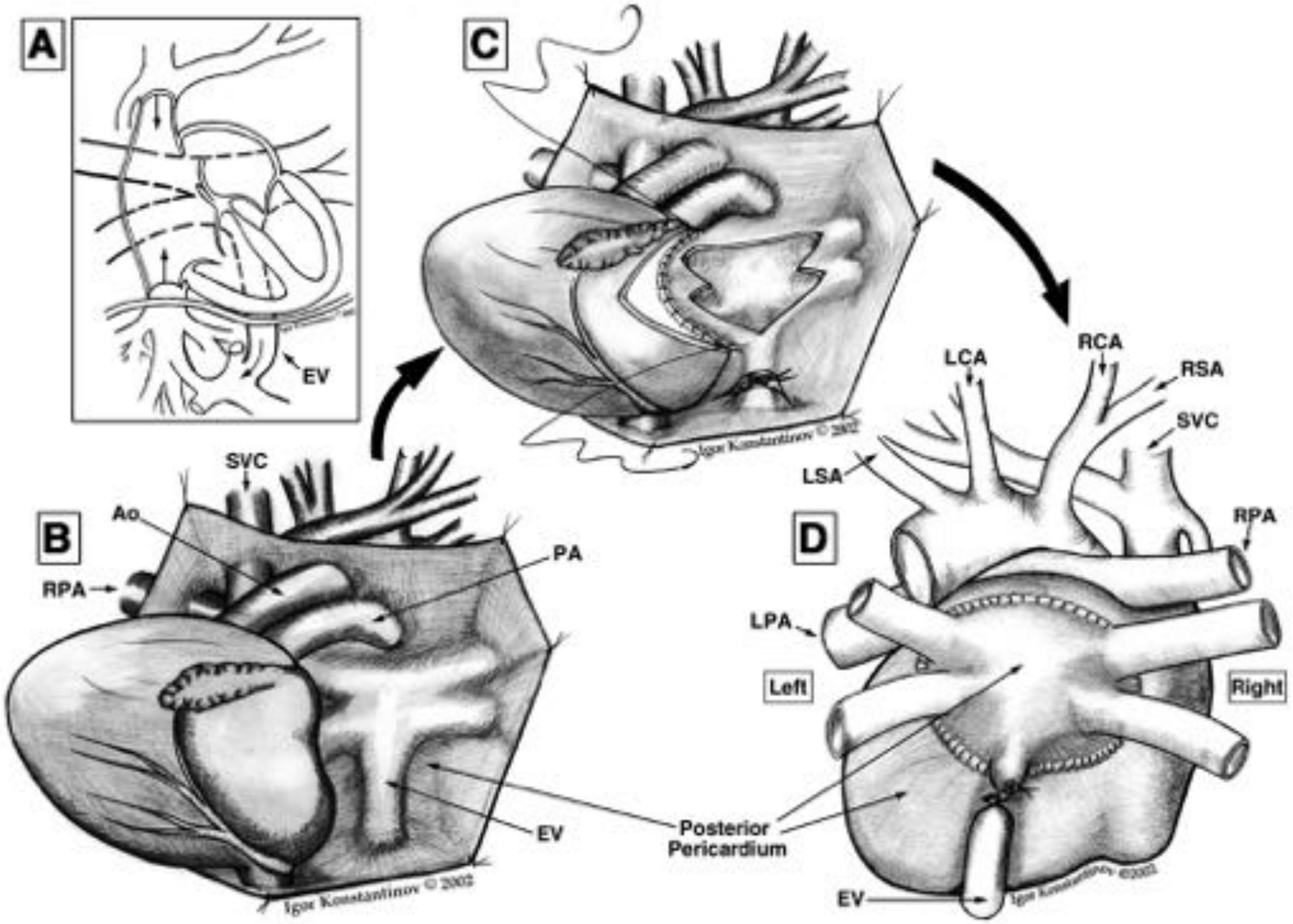
Veine pulmonaire normale



Veine pulmonaire sténosée

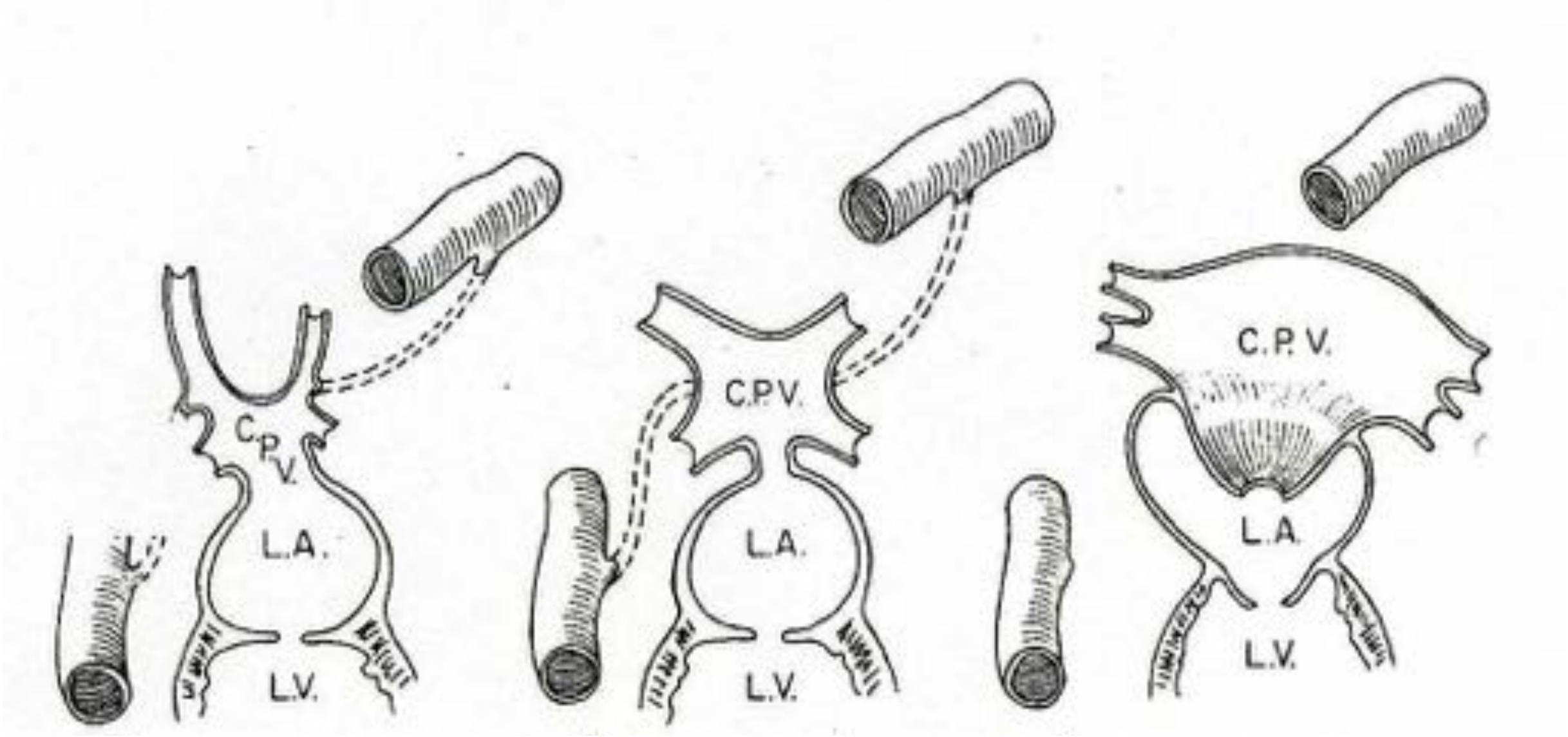
Sténose des veines pulmonaires

Technique dite « sutureless »

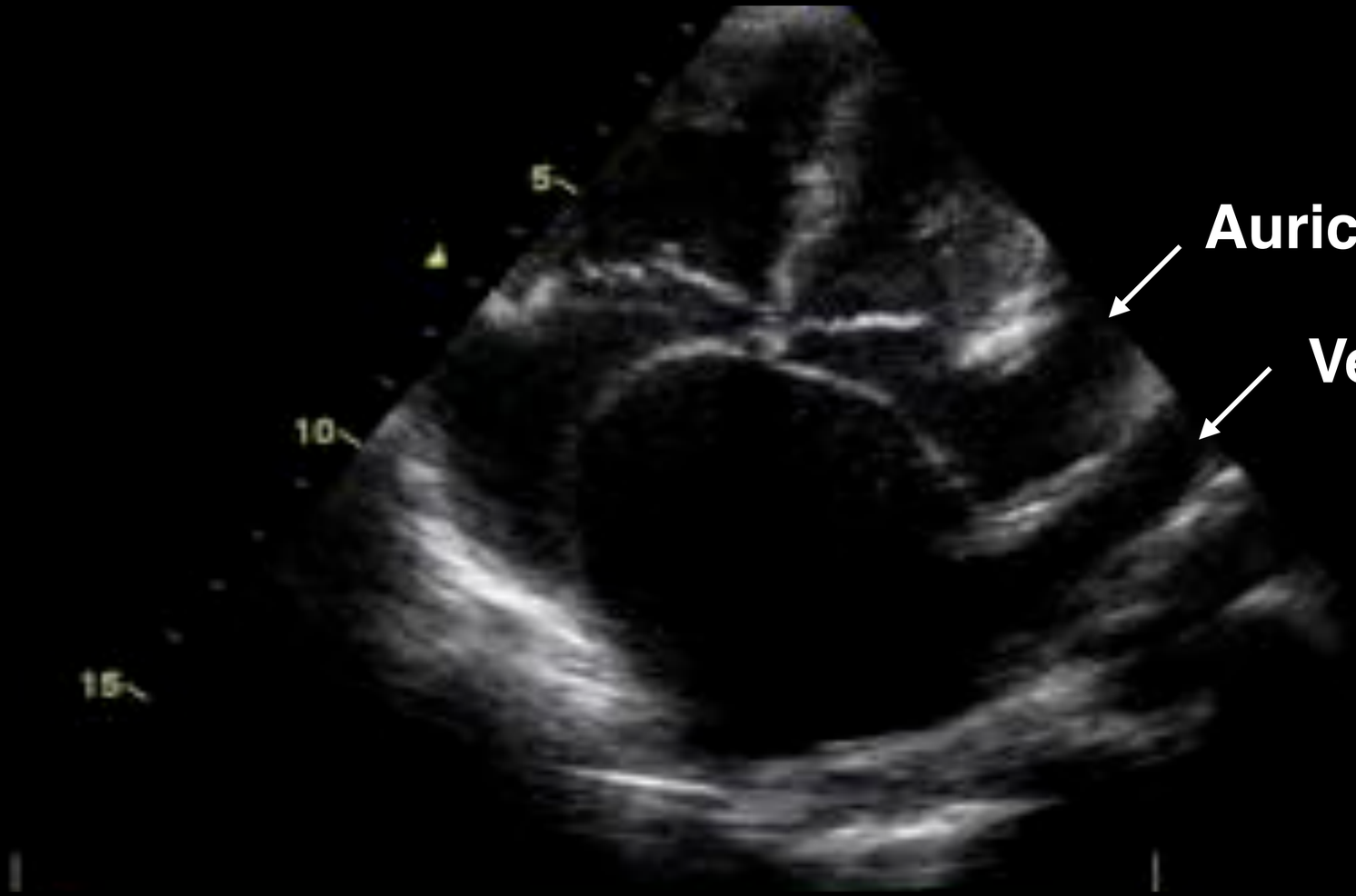


Cœur triatrial

Sténose de la veine pulmonaire commune
Cœur triatrial



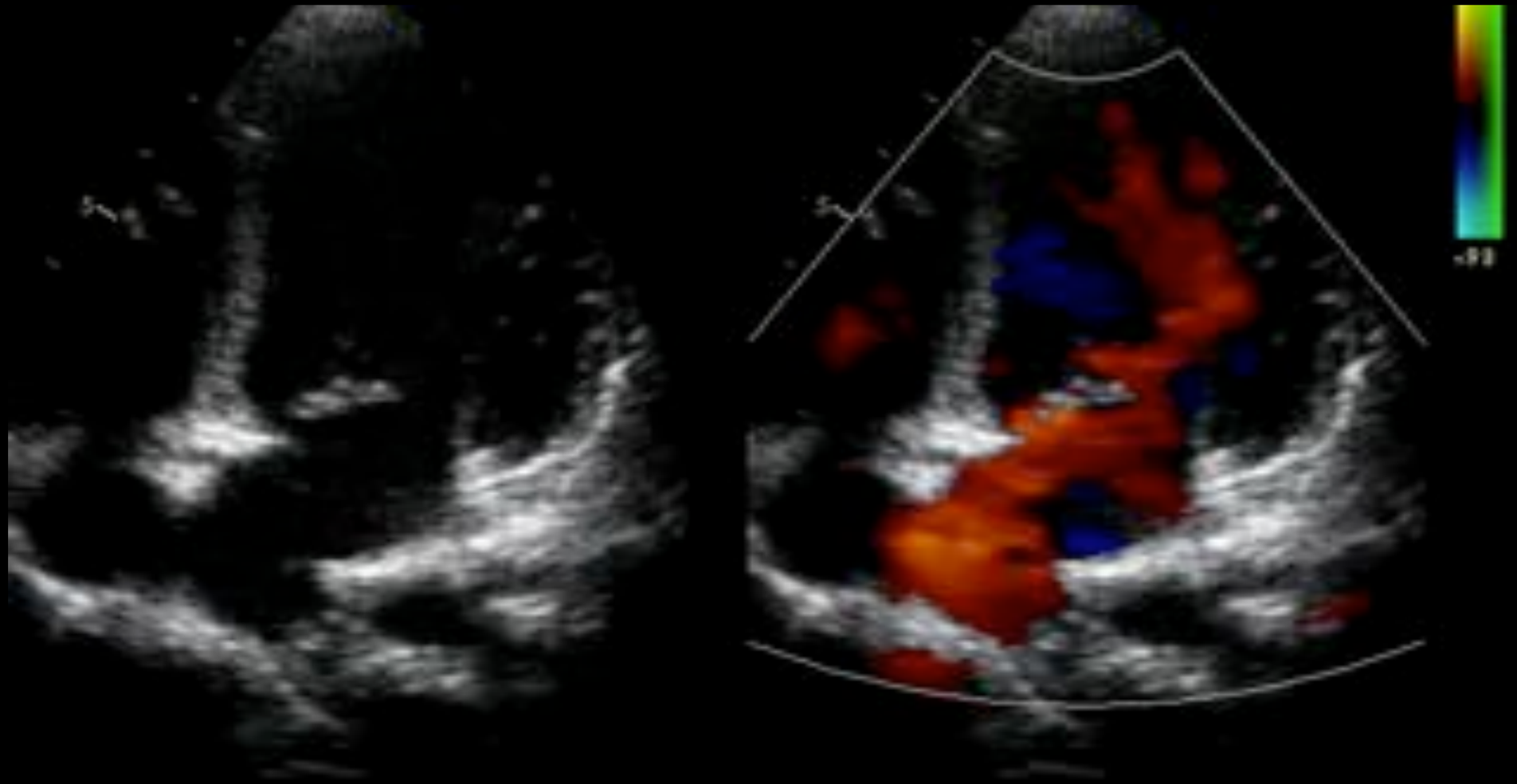
Cœur triatrial



Auricule gauche

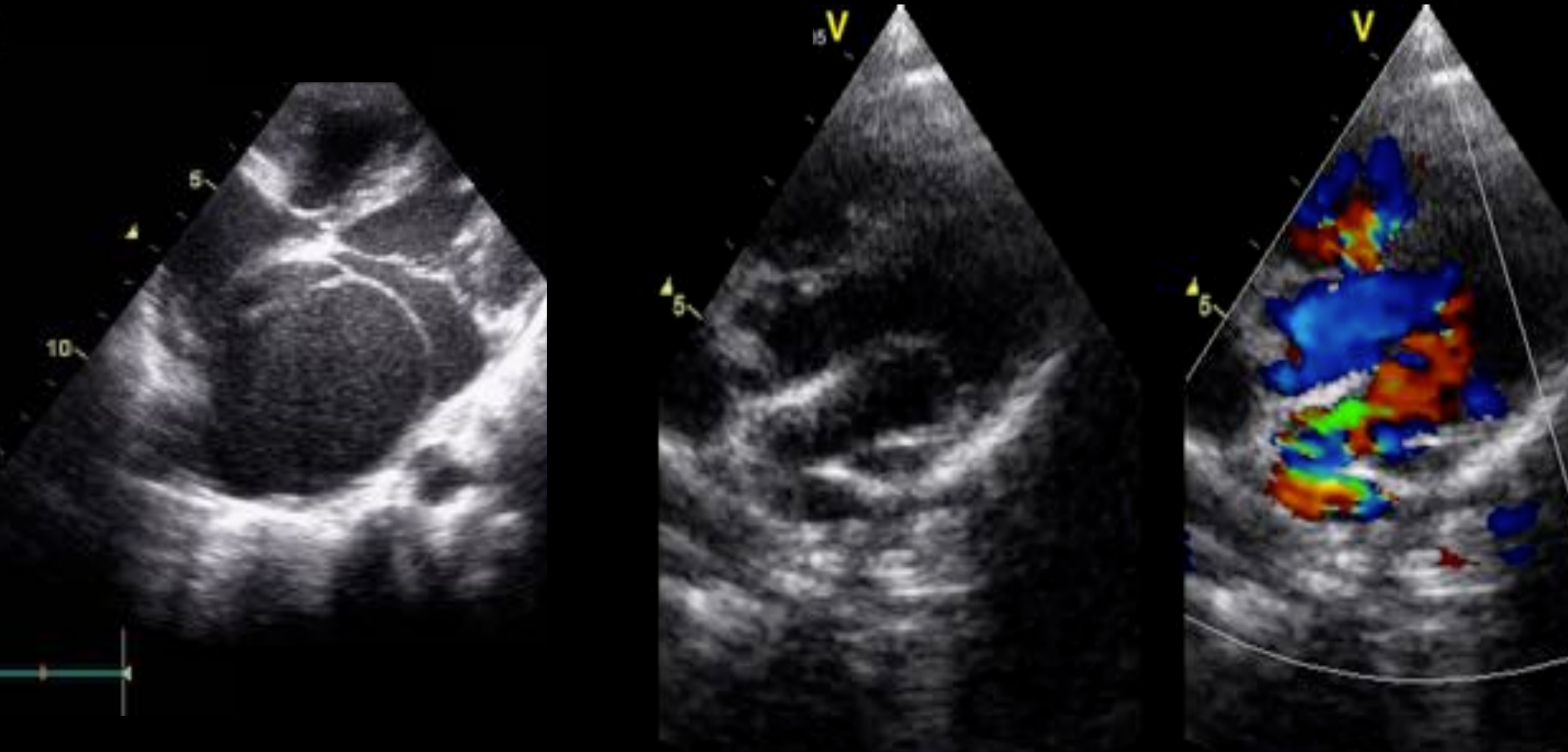
Veine pulmonaire

Cœur triatrial

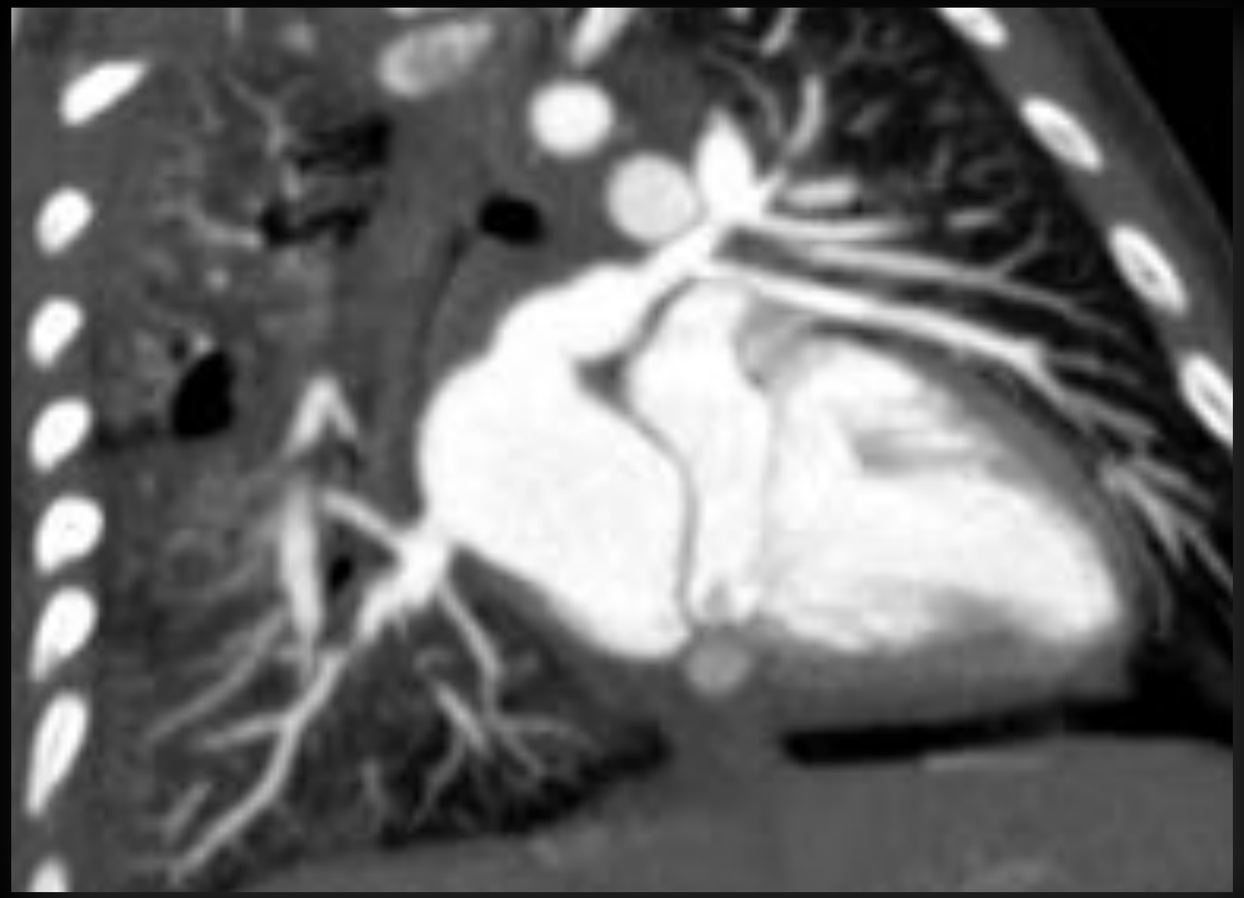


Cœur triatrial

Cœur triatrial



Cœur triatrial



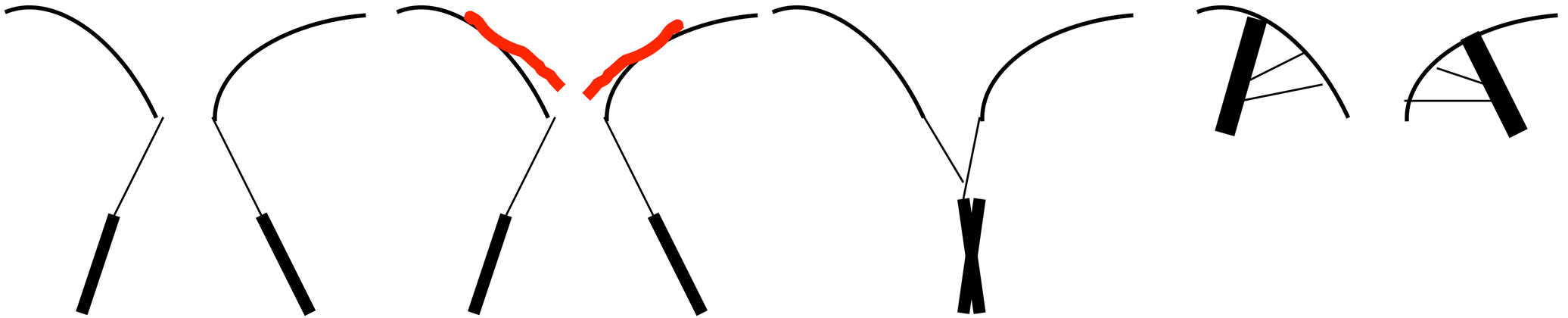
Les anomalies des feuillets mitraux

- Prolapsus
- Cleft isolée
- Double orifice
- Anneau supramitral
- Ebstein mitral

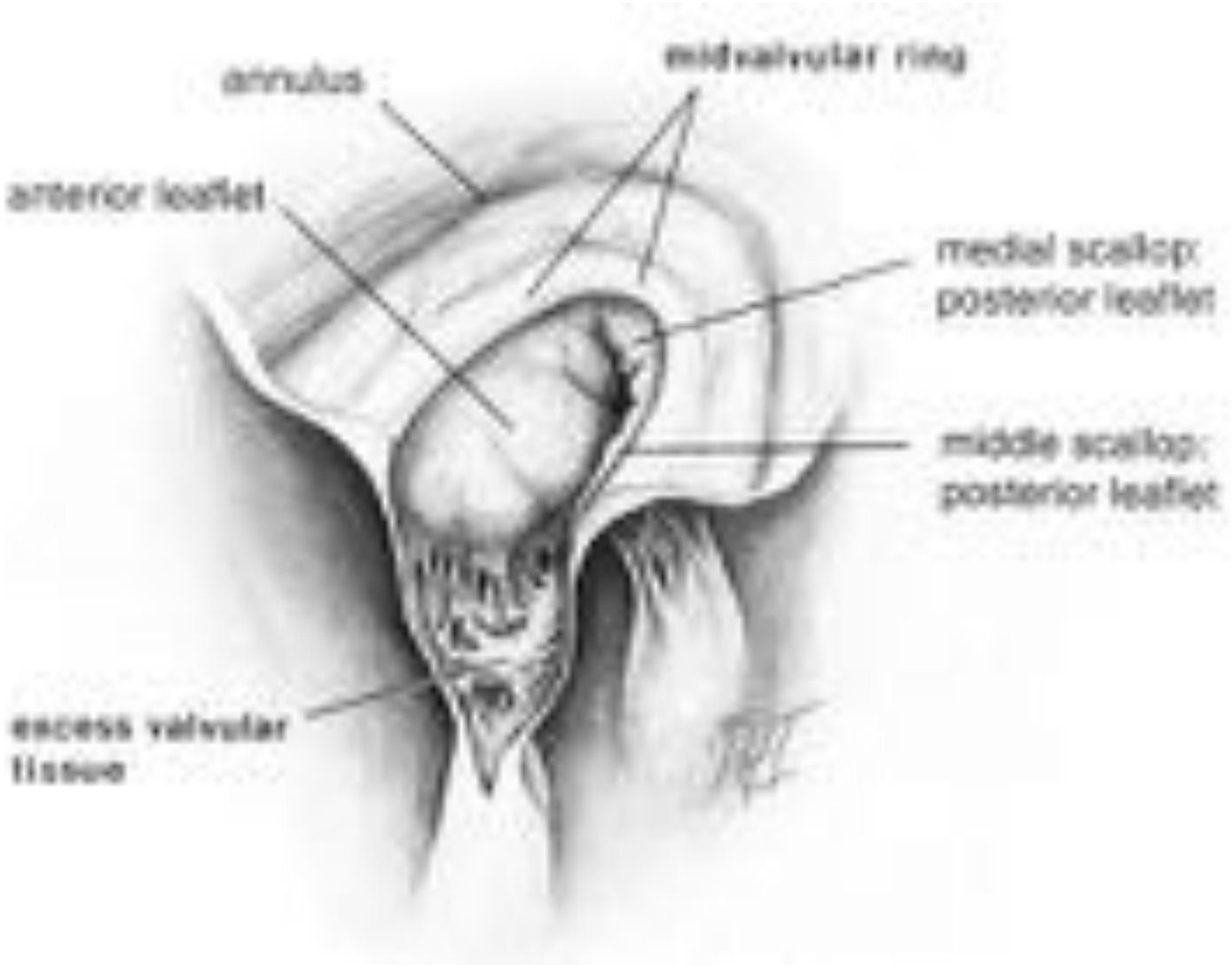
Rétrécissement mitral congénital

Anatomie

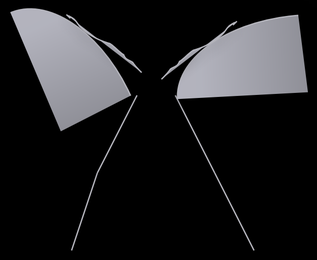
- Membrane supramitrale
- Parachute
- Hamac



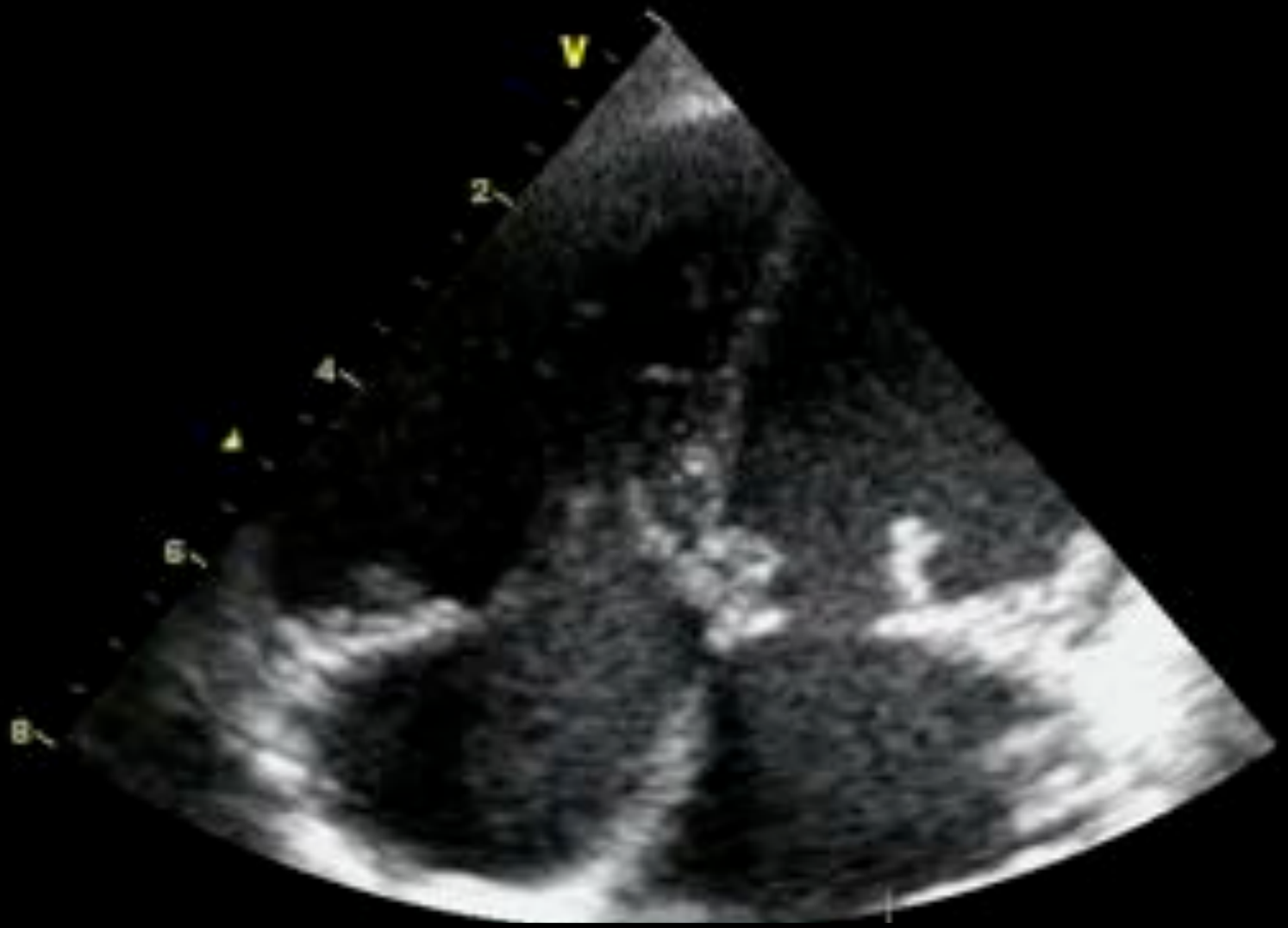
Membrane supramitrale



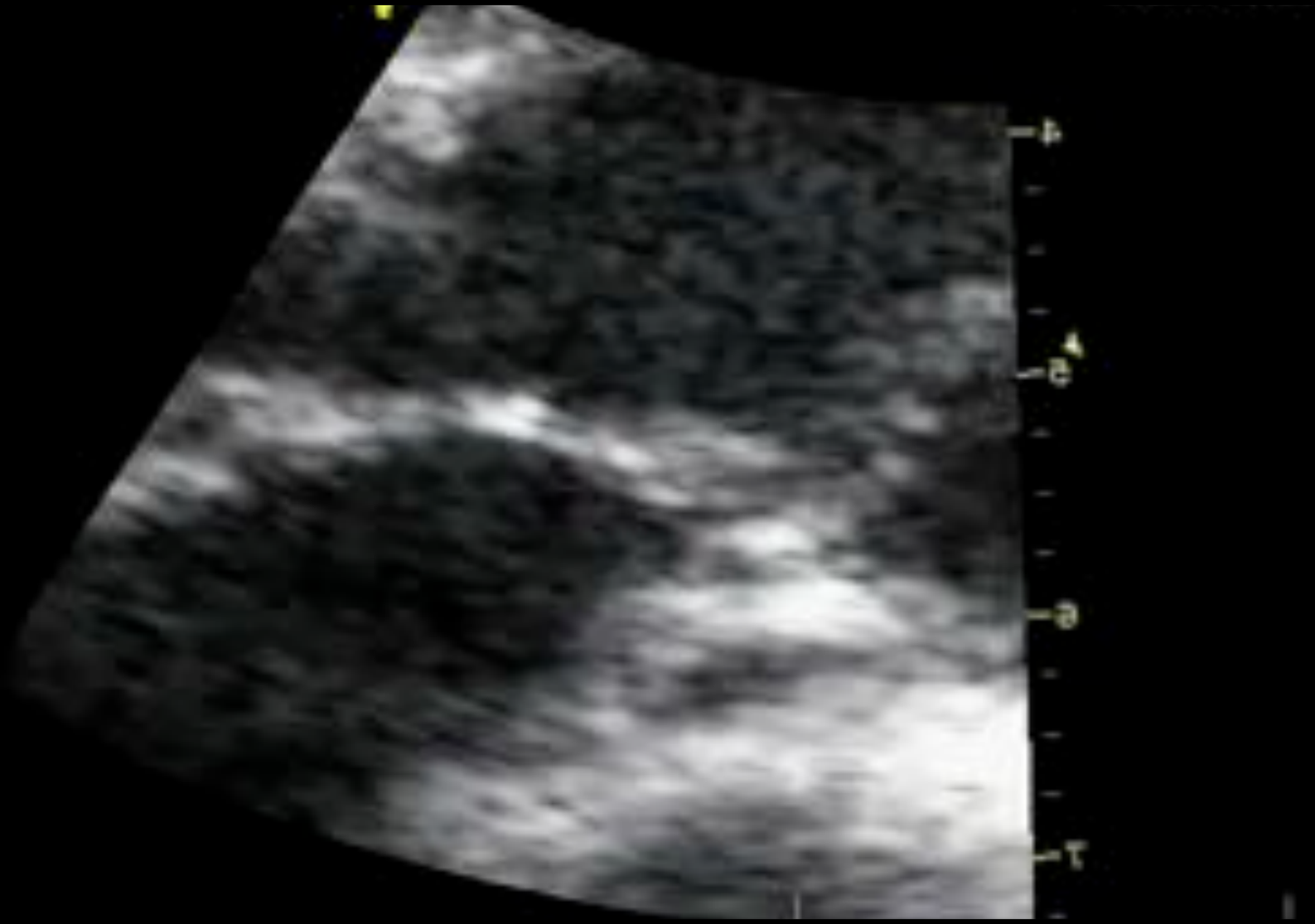
Membrane supramitrale



Membrane supramitrale



Membrane supramitrale



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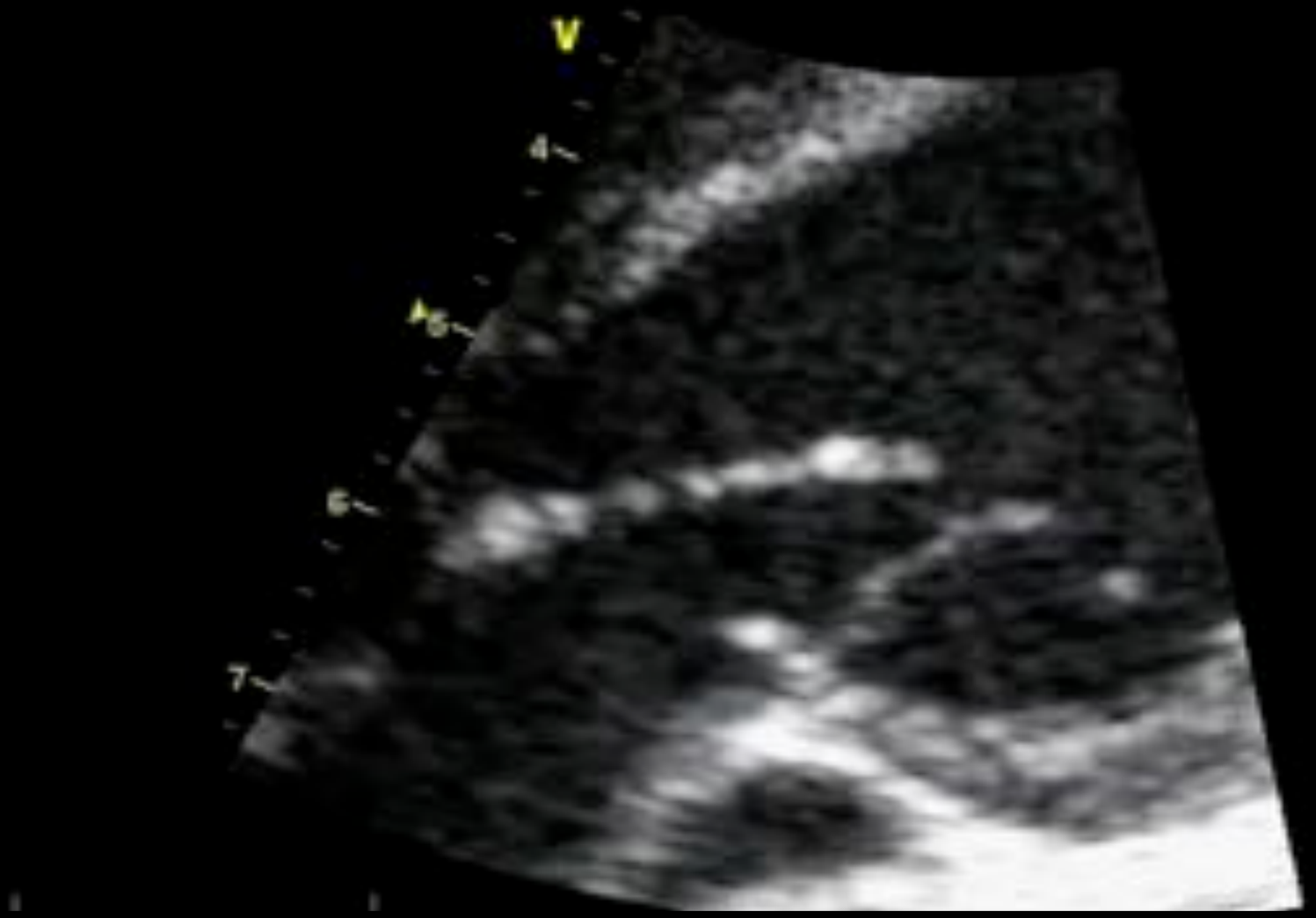

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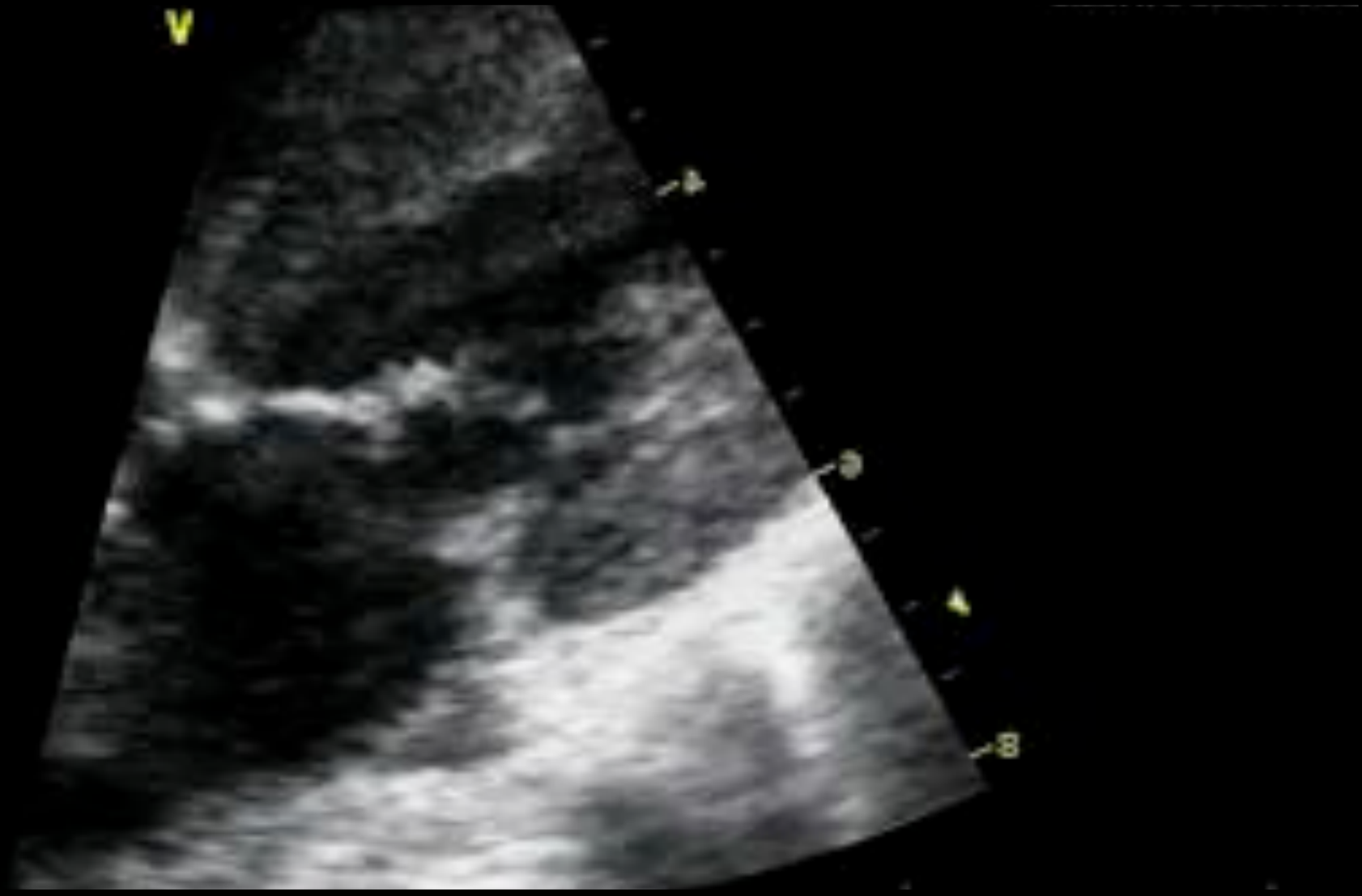
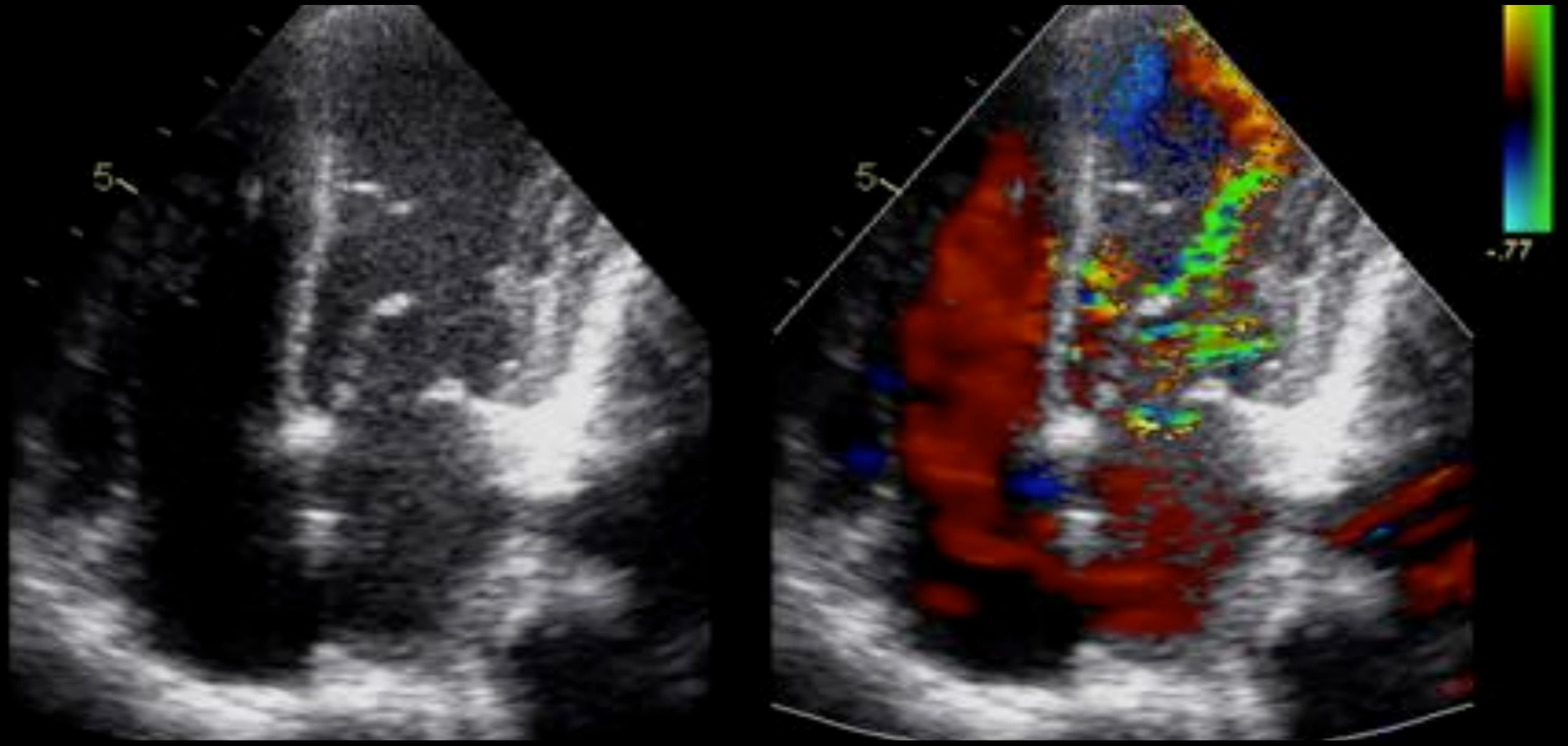
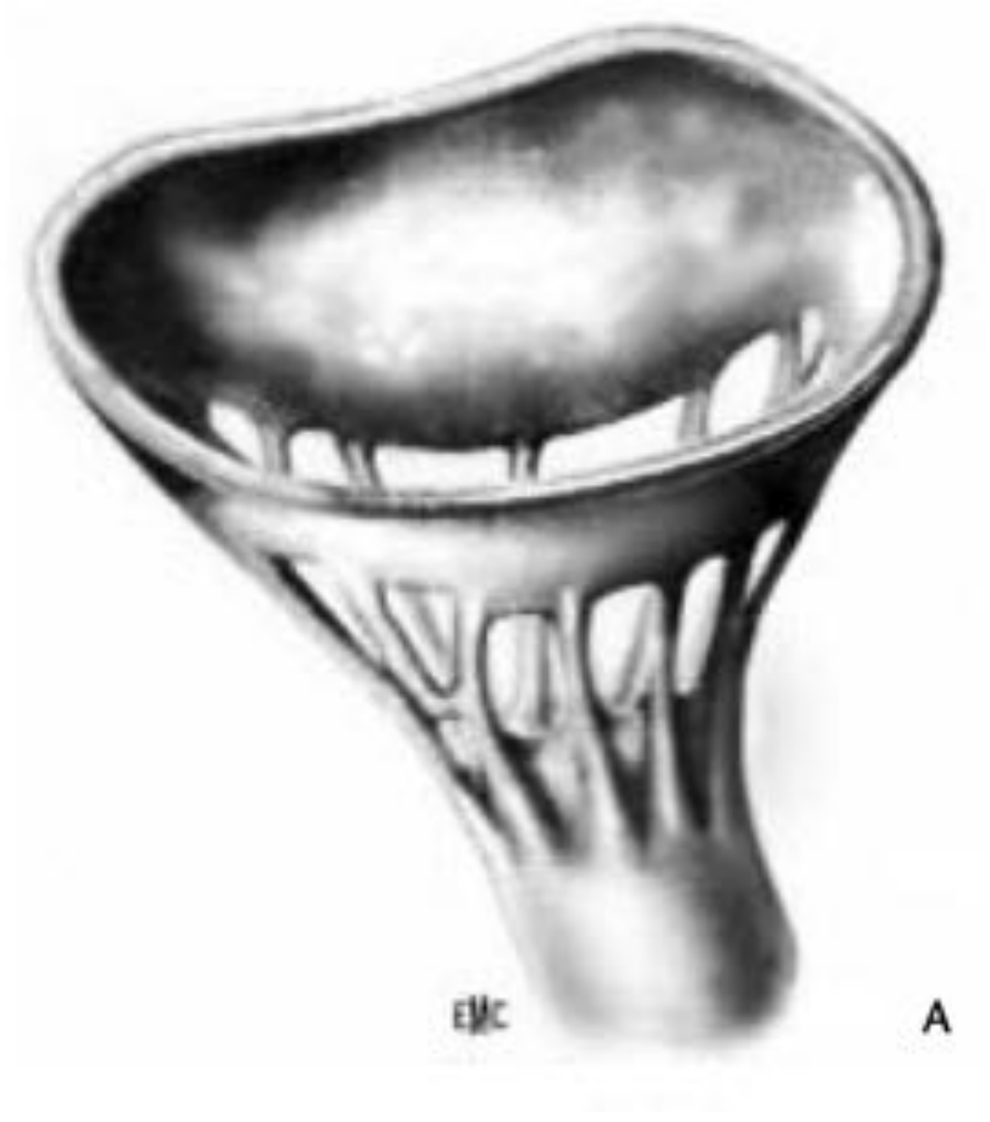


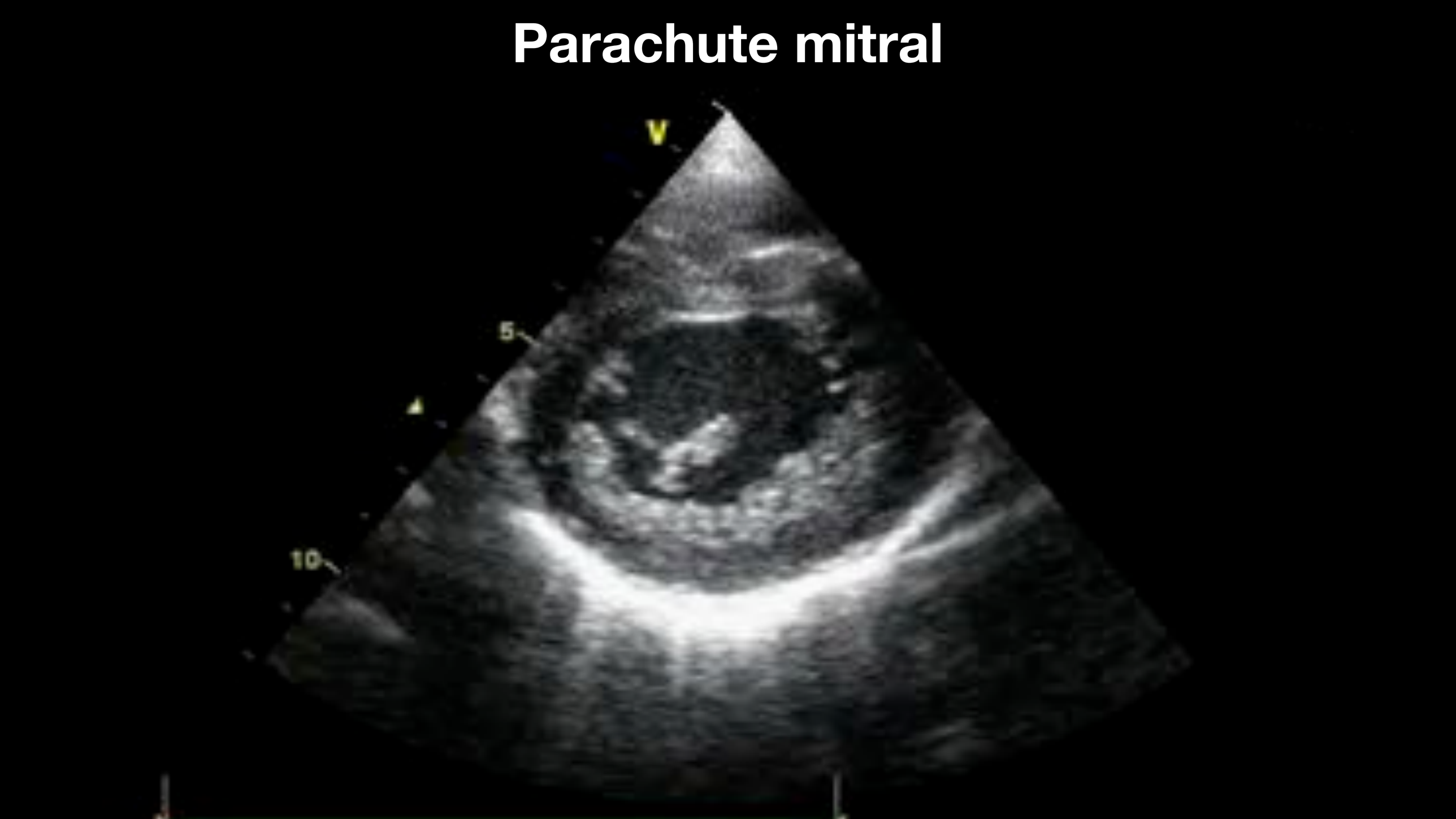
Membrane supramitrale



Parachute mitral



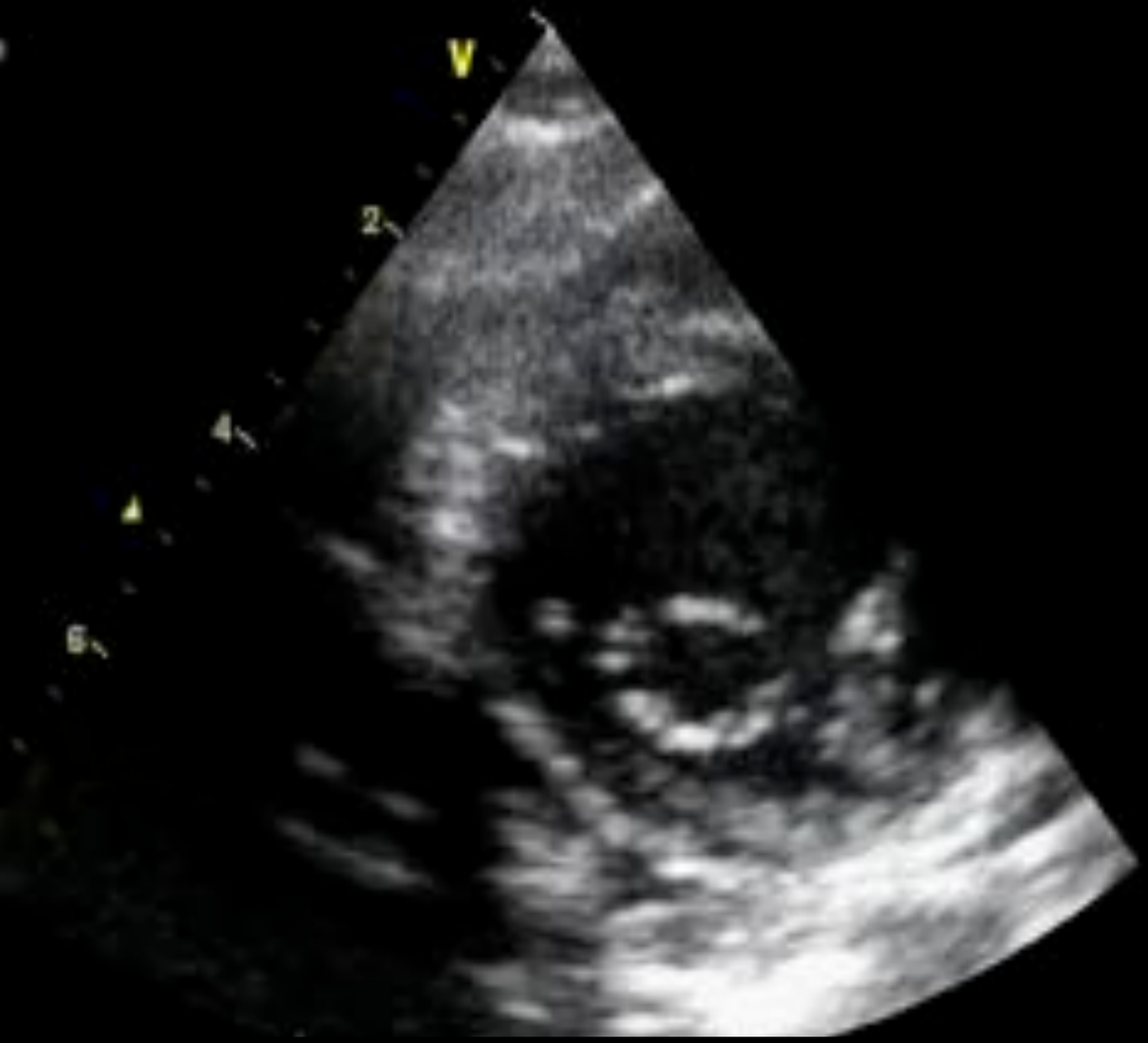
Parachute mitral

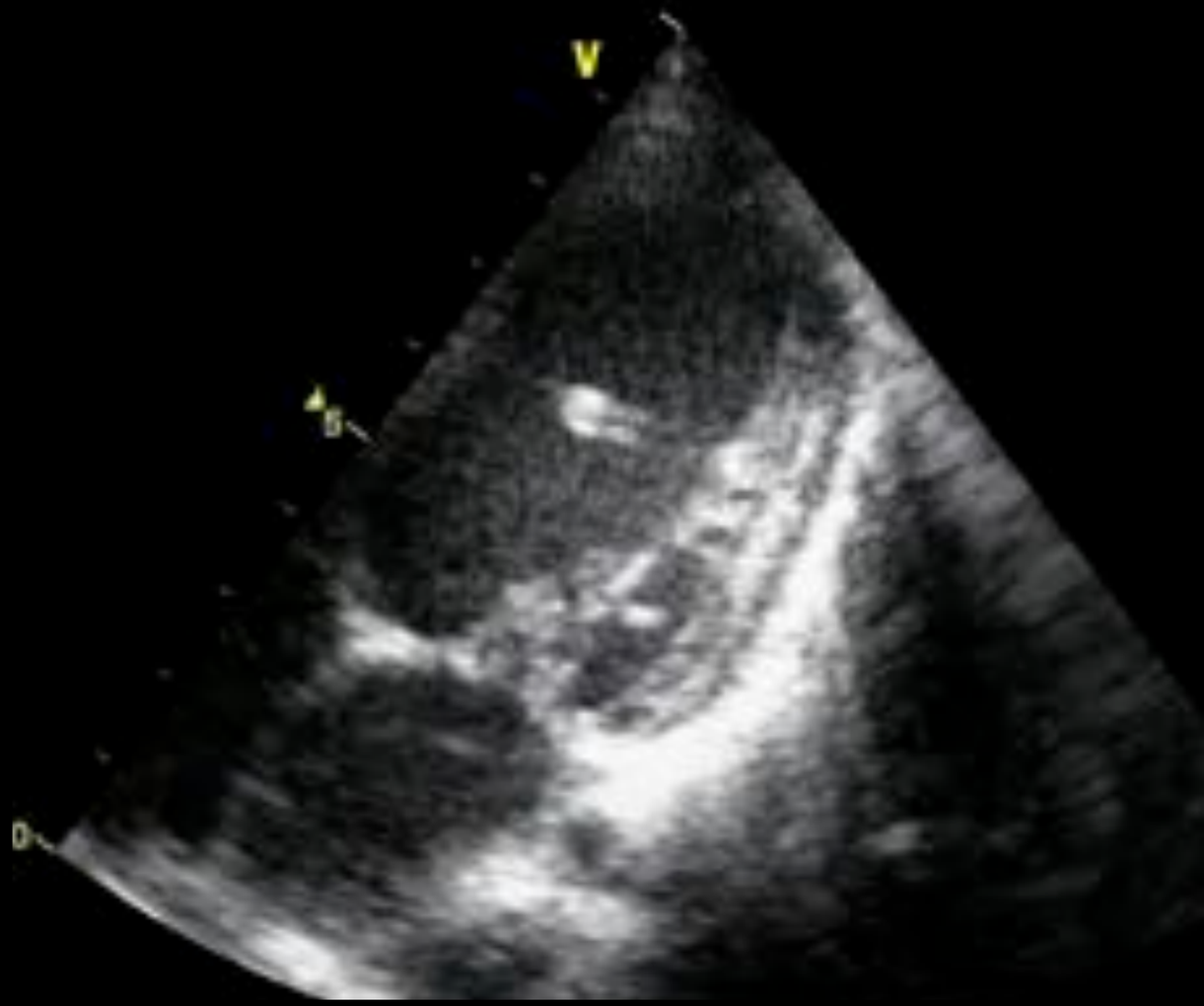


Parachute mitral

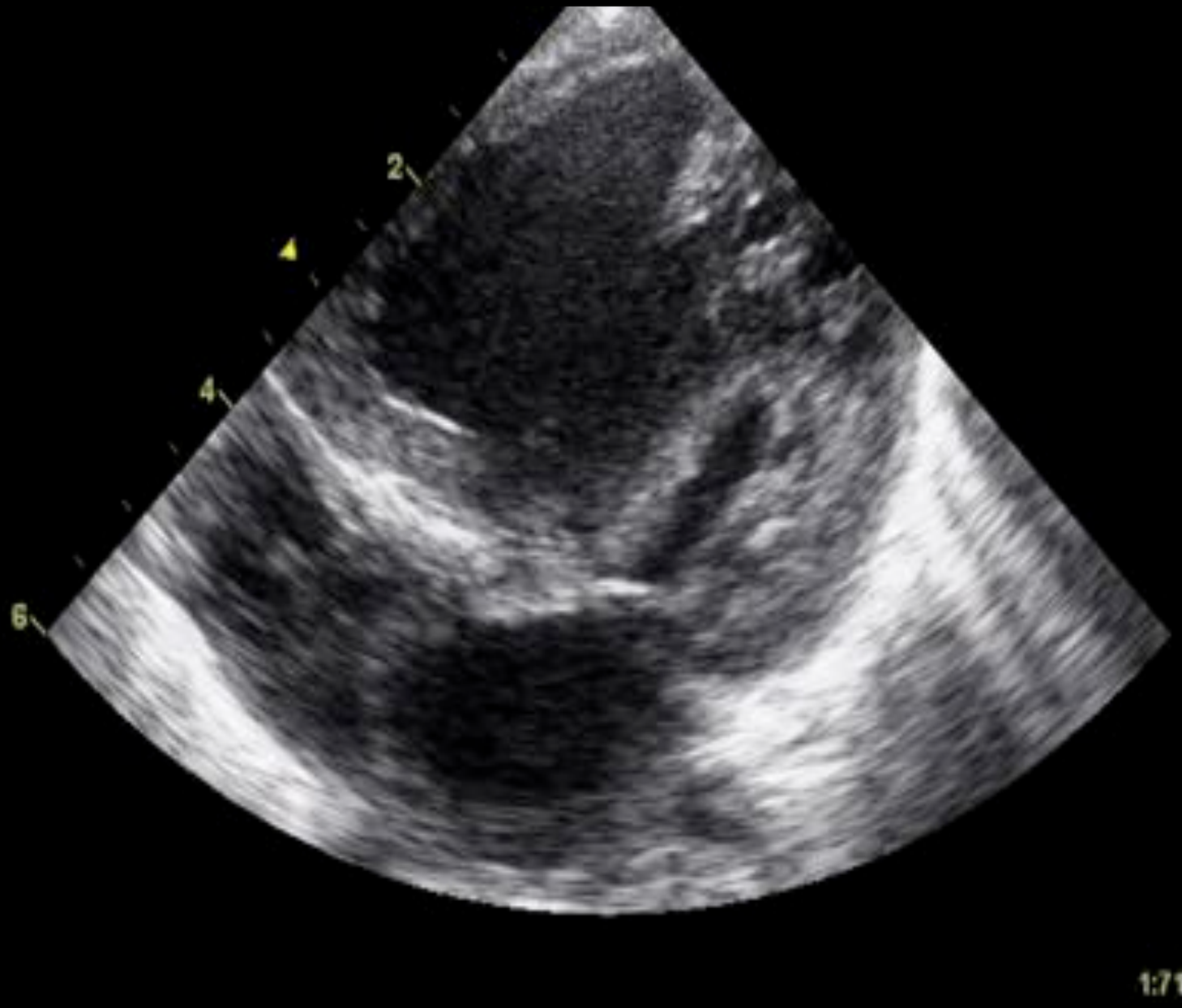
Parachute mitral

Parachute mitral

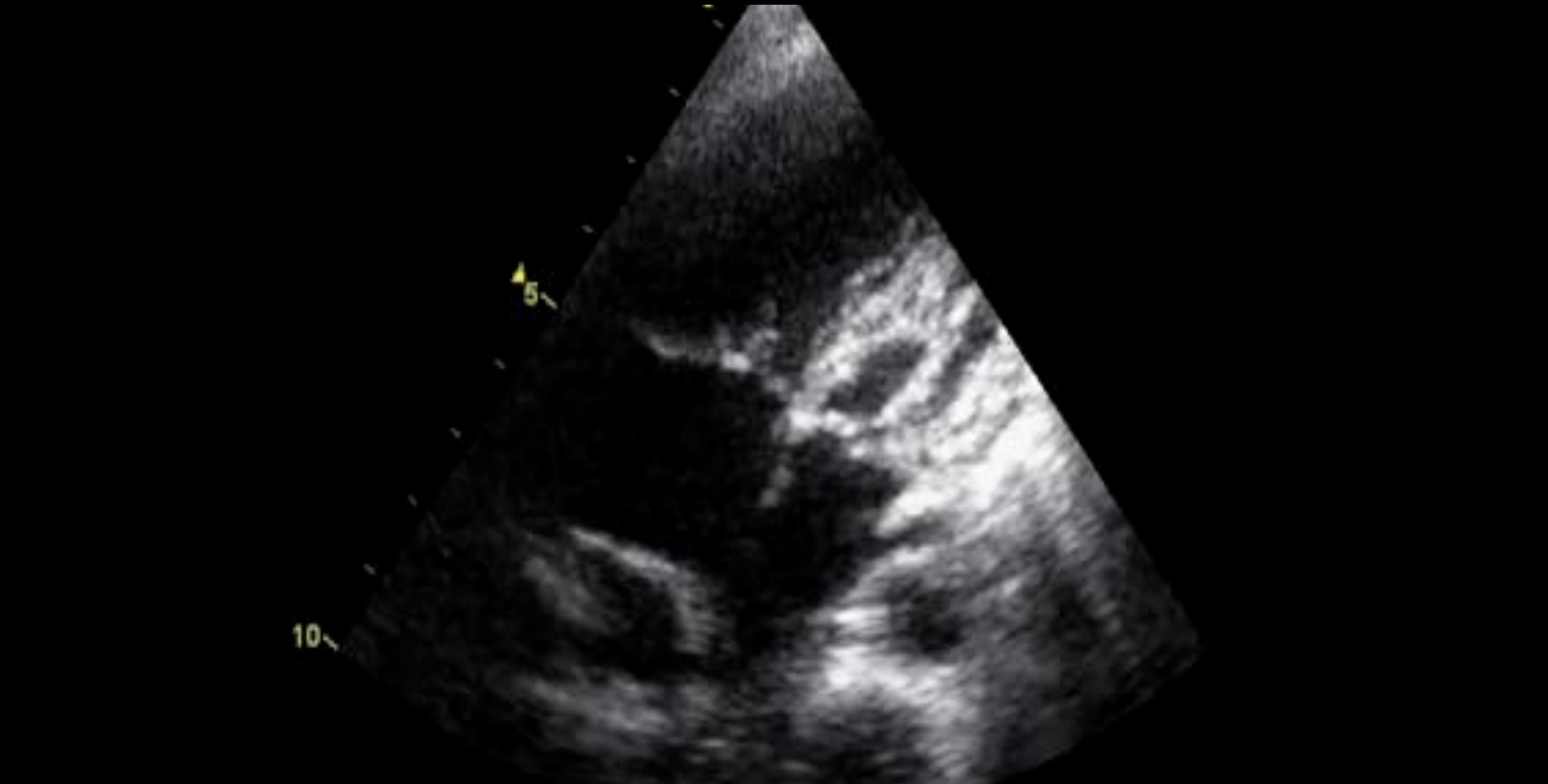




Hypoplasie mitrale



Hypoplasie mitrale

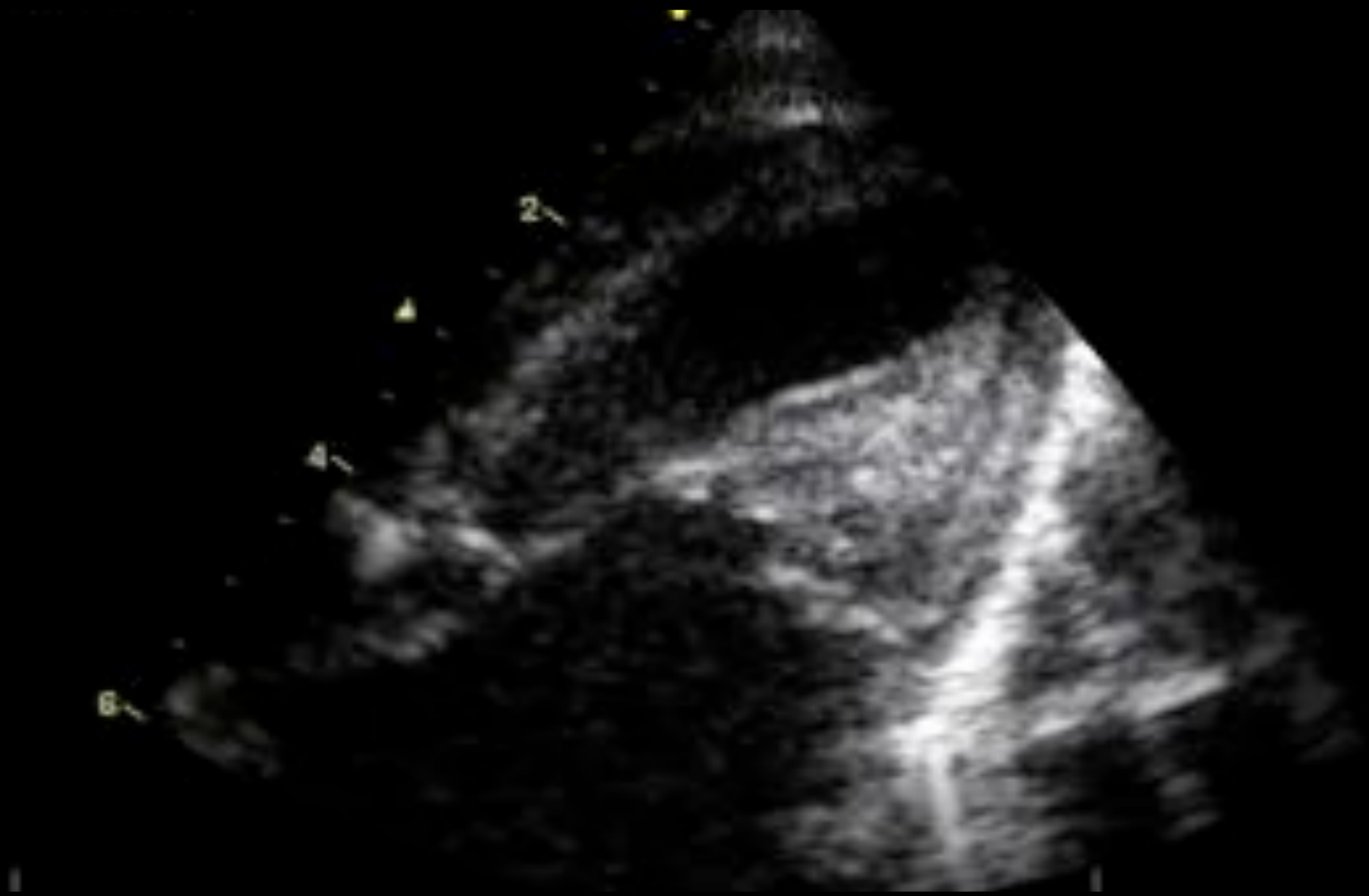


Hamac mitral

A

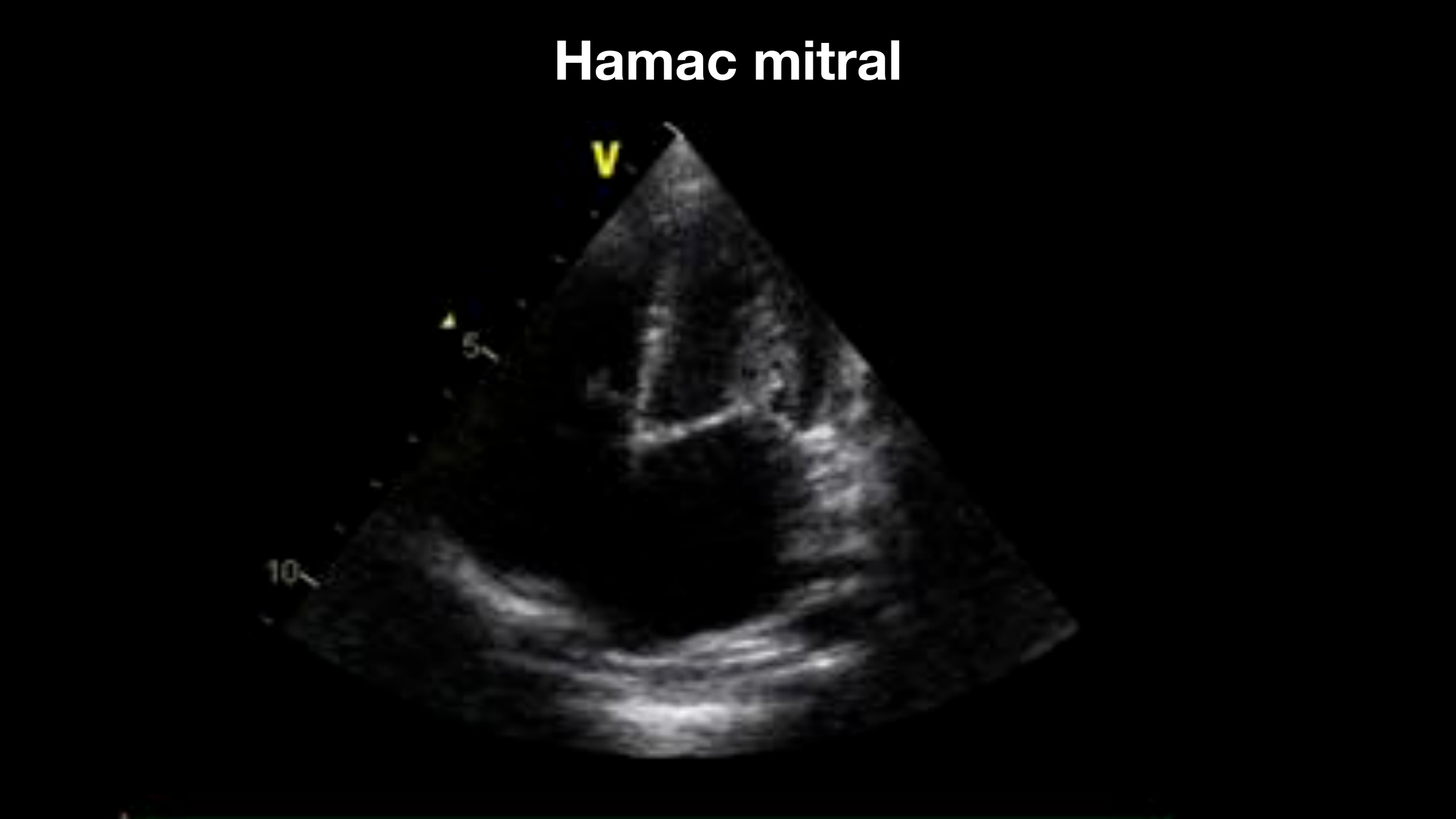
B

Hamac mitral



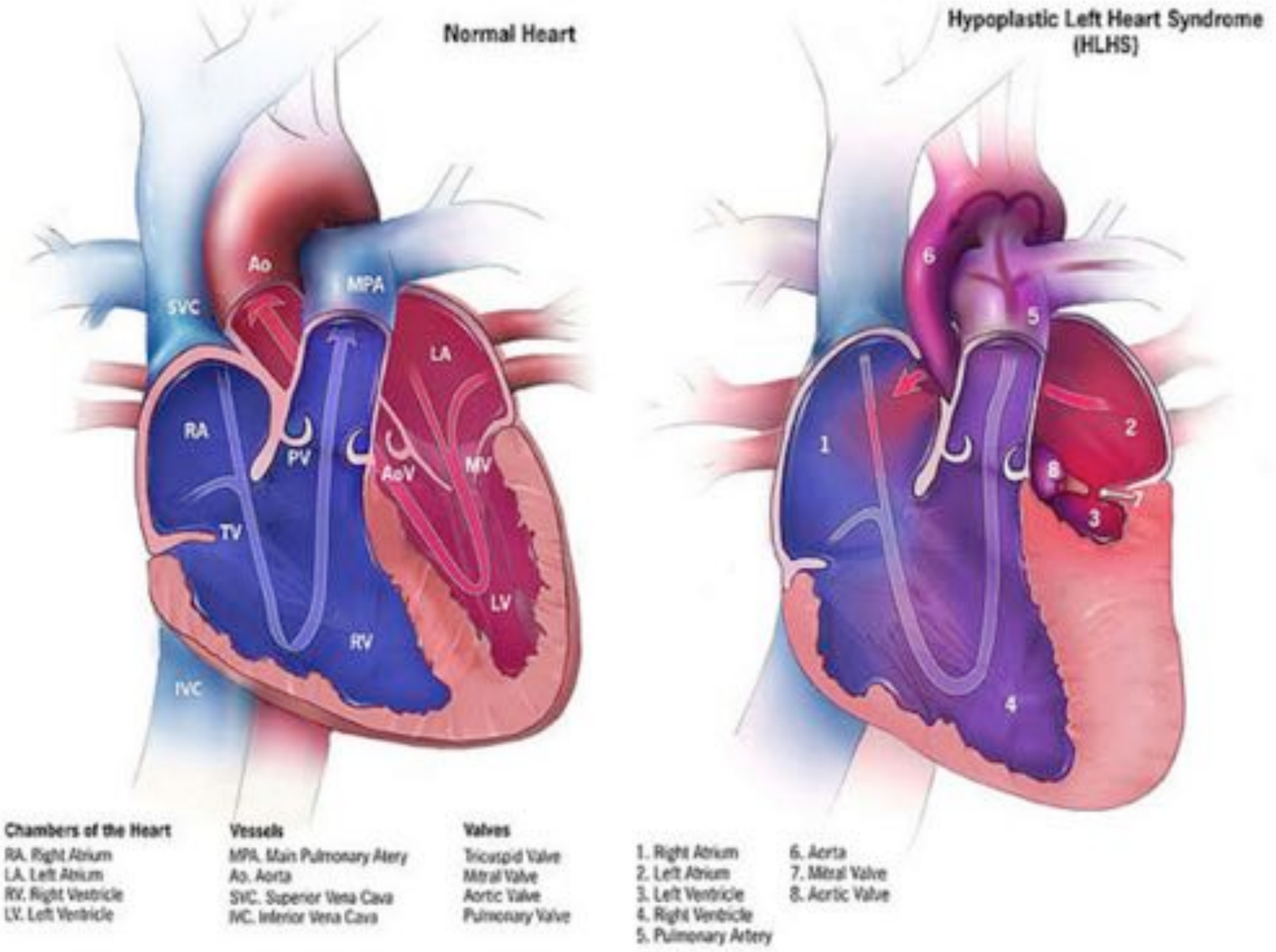
Hamac mitral

Hamac mitral



Hamac mitral

Hypoplasie du cœur gauche

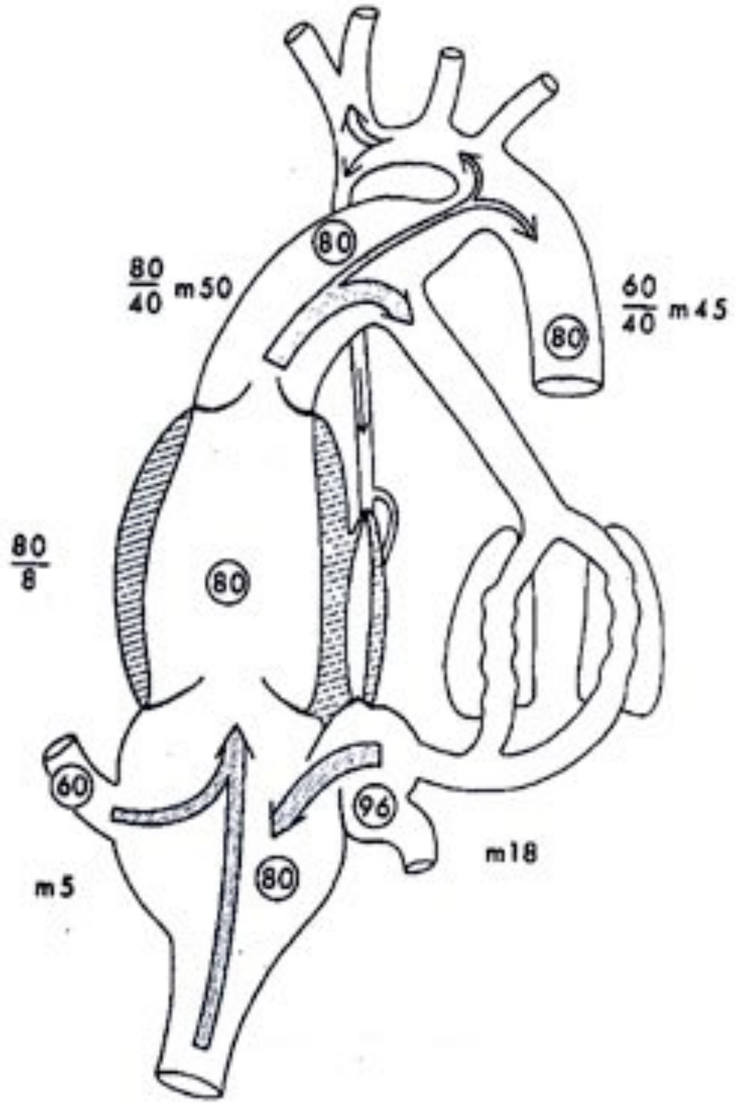


Hypoplasie du cœur gauche


Cincinnati
Children's

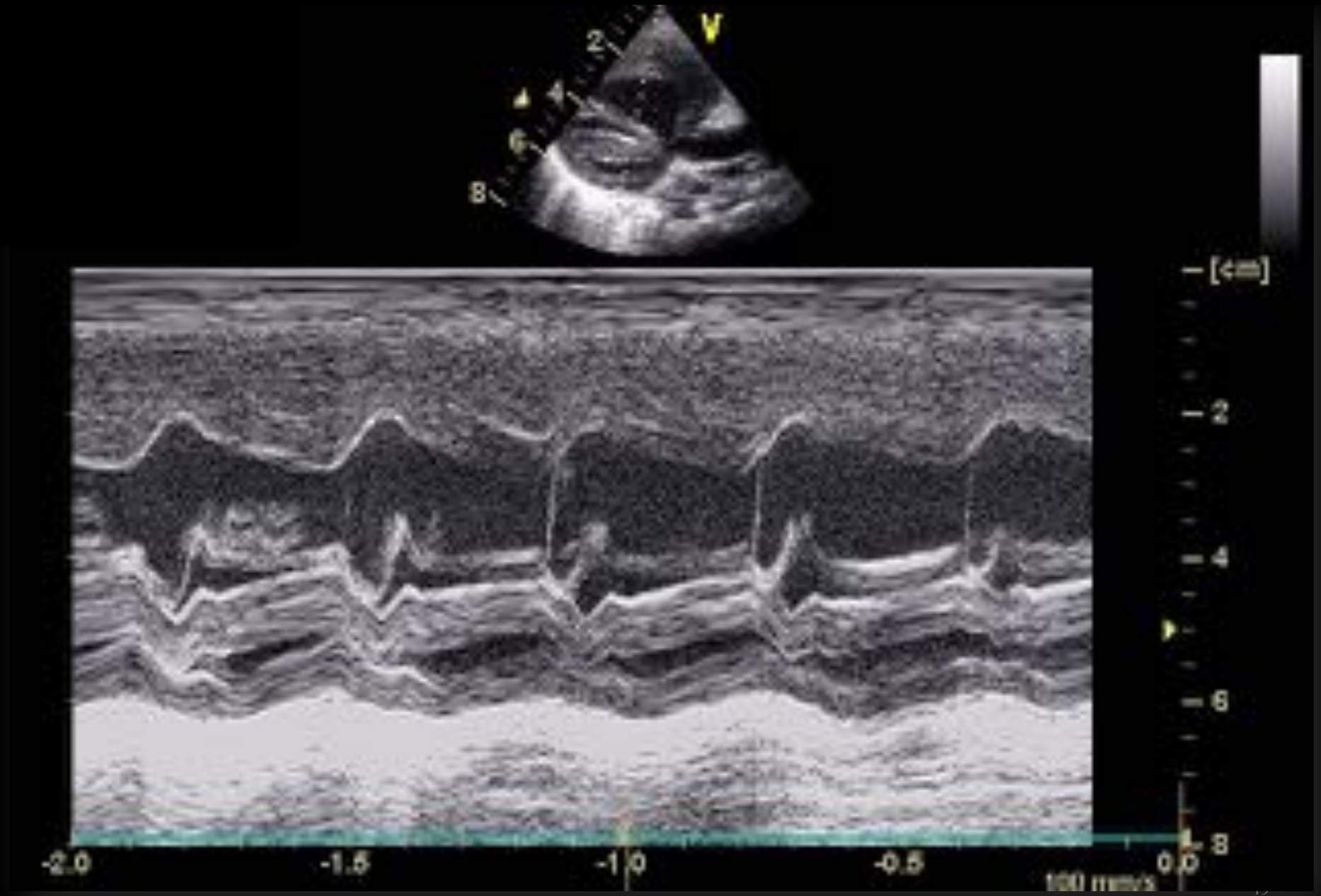
Hypoplasie du cœur gauche

Physiologie foetale



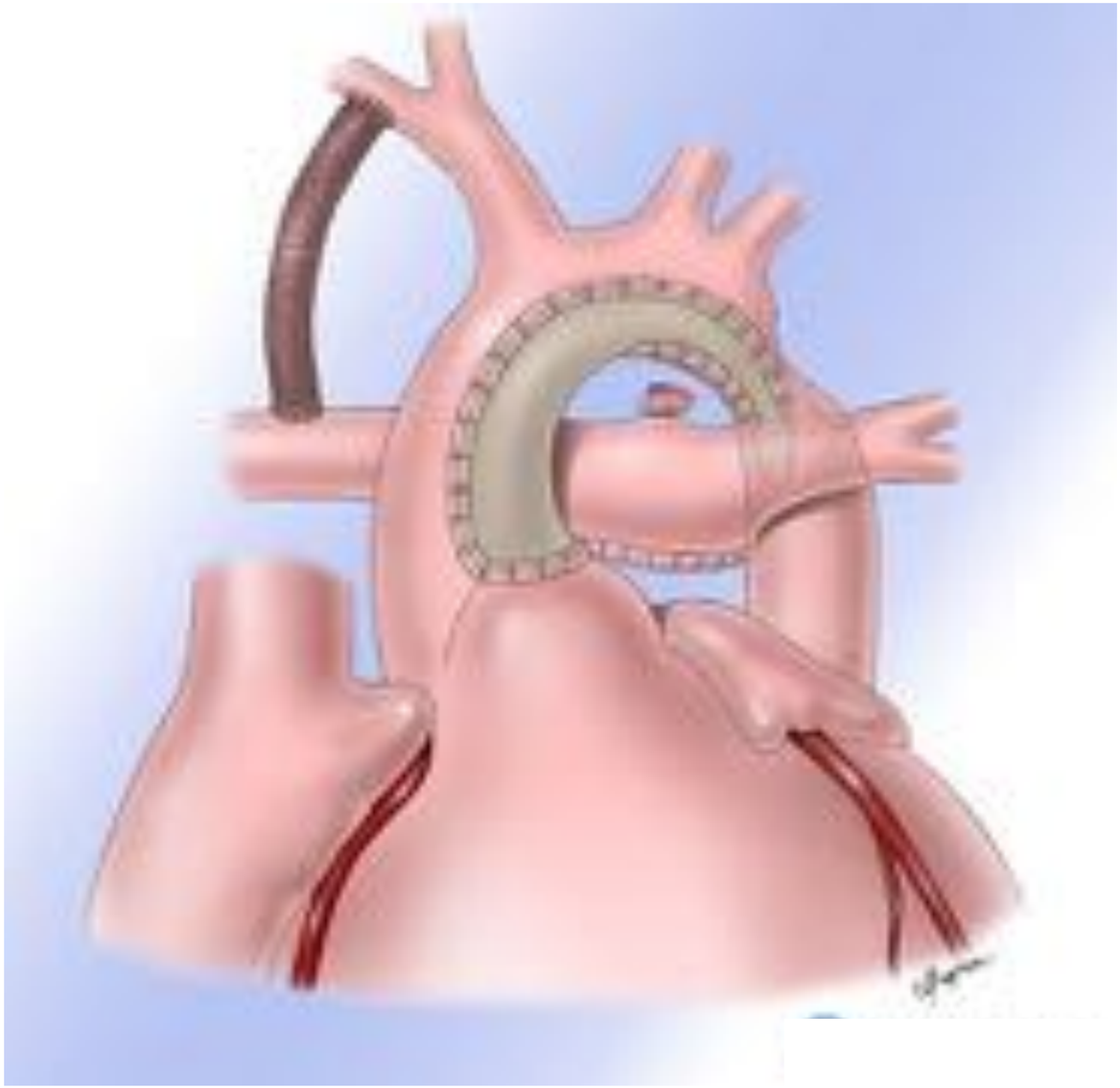
Hypoplasie du cœur gauche

Hypoplasie du cœur gauche



Hypoplasie du cœur gauche

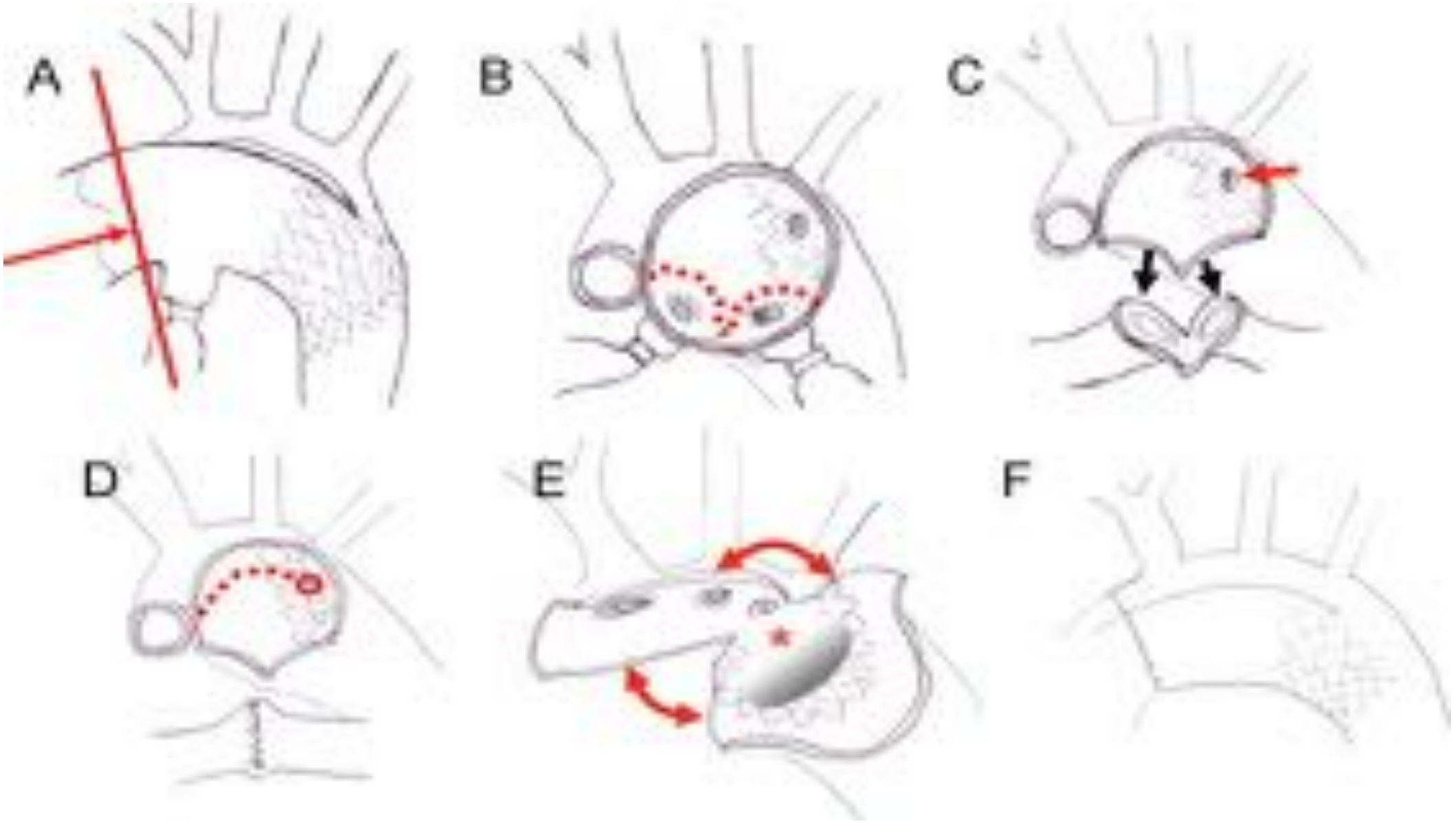
1er temps de l’opération de Norwood



Programme de Norwood

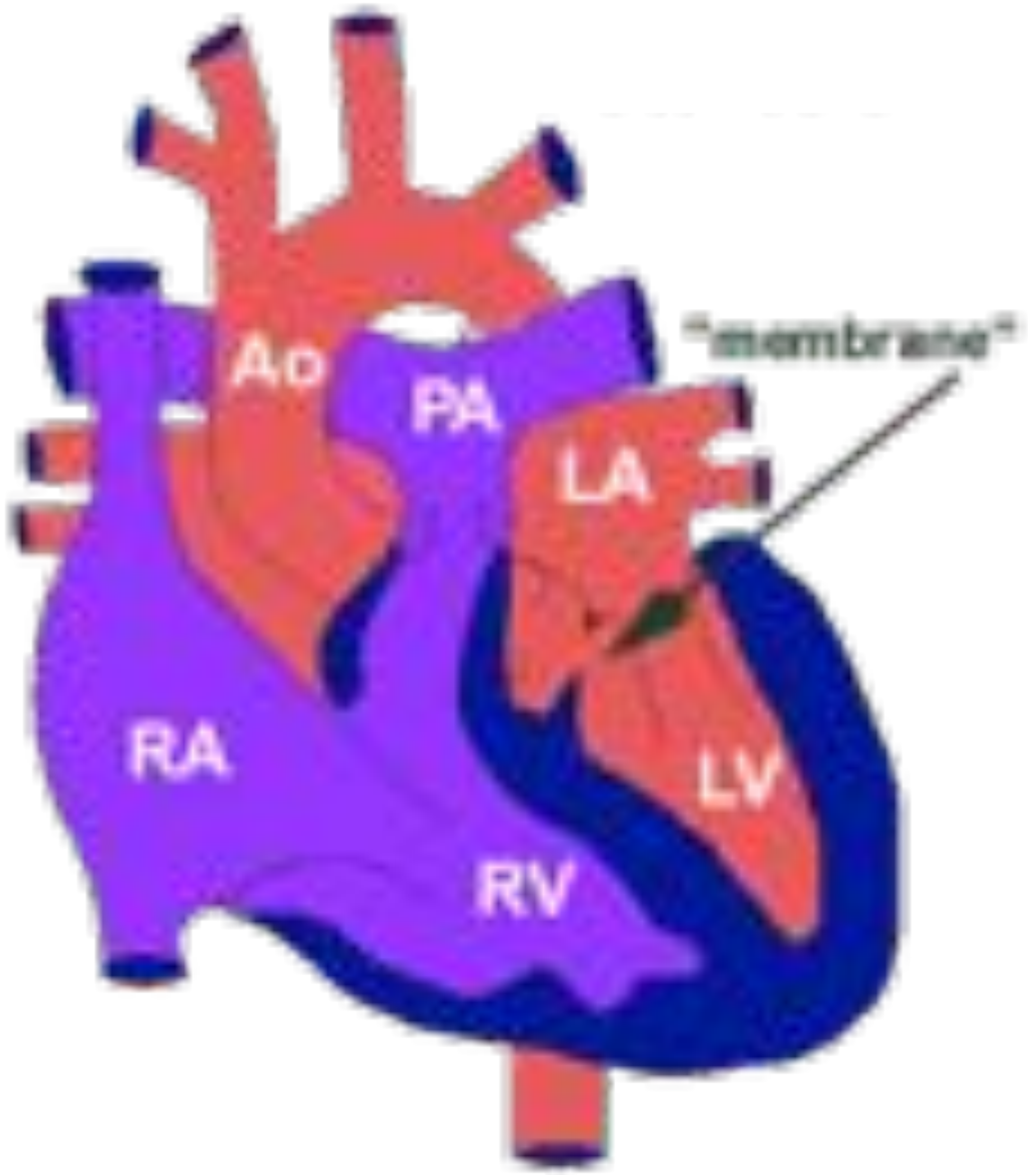
- Stage 1: Blalock ou tube VD-AP (Sano)
- Complications post-opératoires
  - Coarctation
  - Dysfonction VD ou IT
- Stage 2 : DCPP
- Stage 3 : DCPT

Hybride dans l’hypoplasie du coeur gauche



Sténose sous-aortique

Sténose sous-aortique



Sténose sous-aortique

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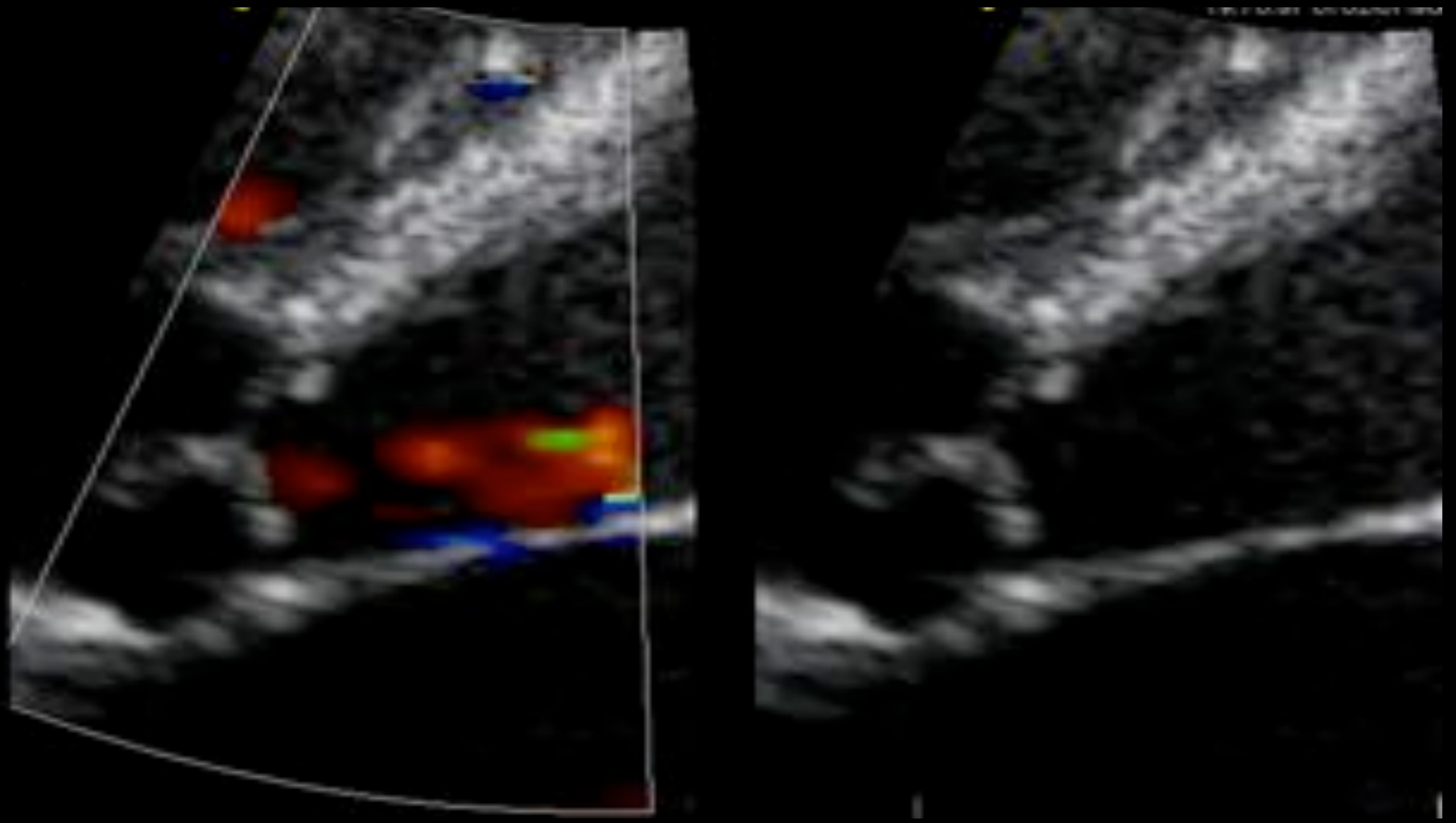
Sténose sous-aortique



Sténose sous-aortique

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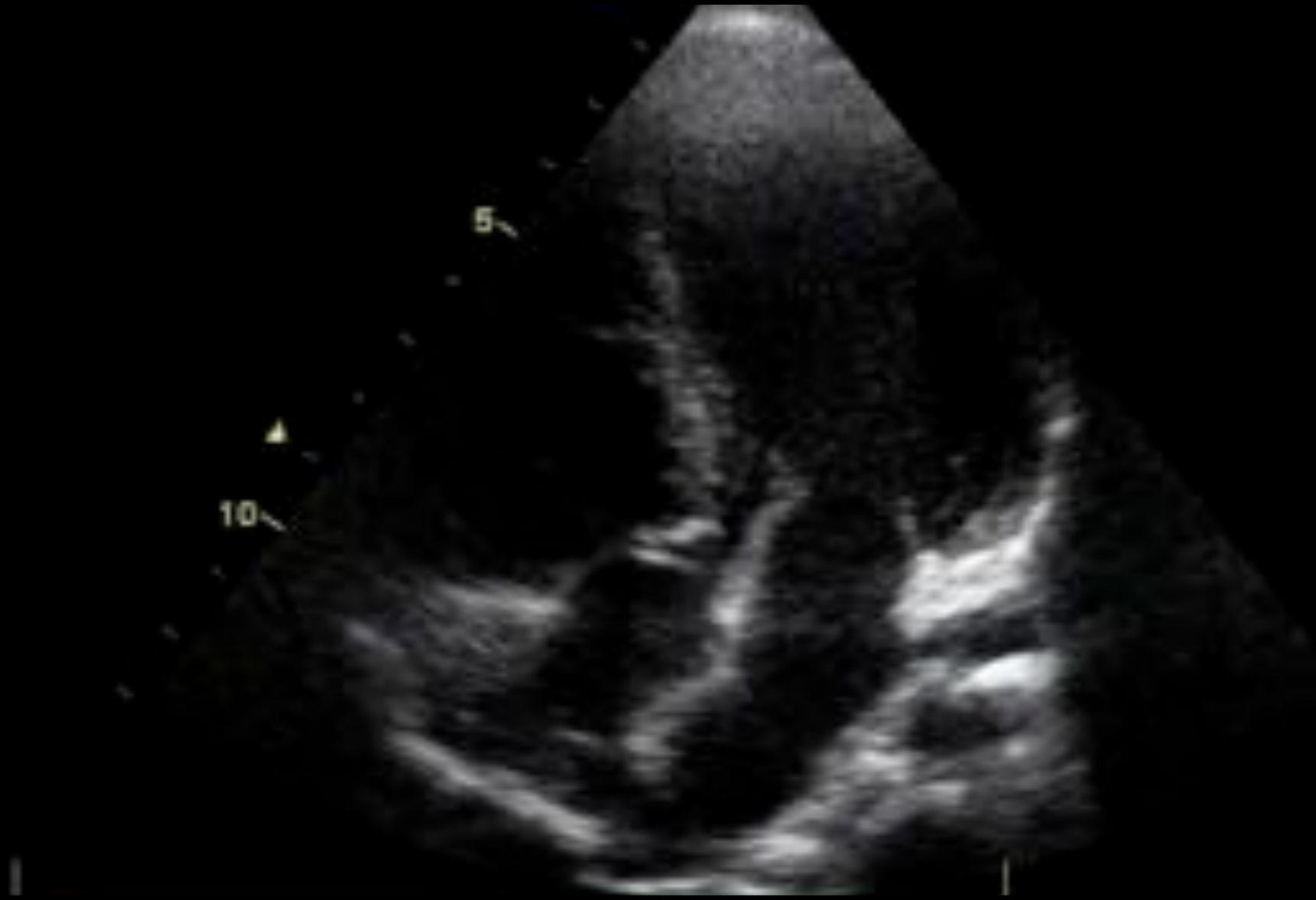

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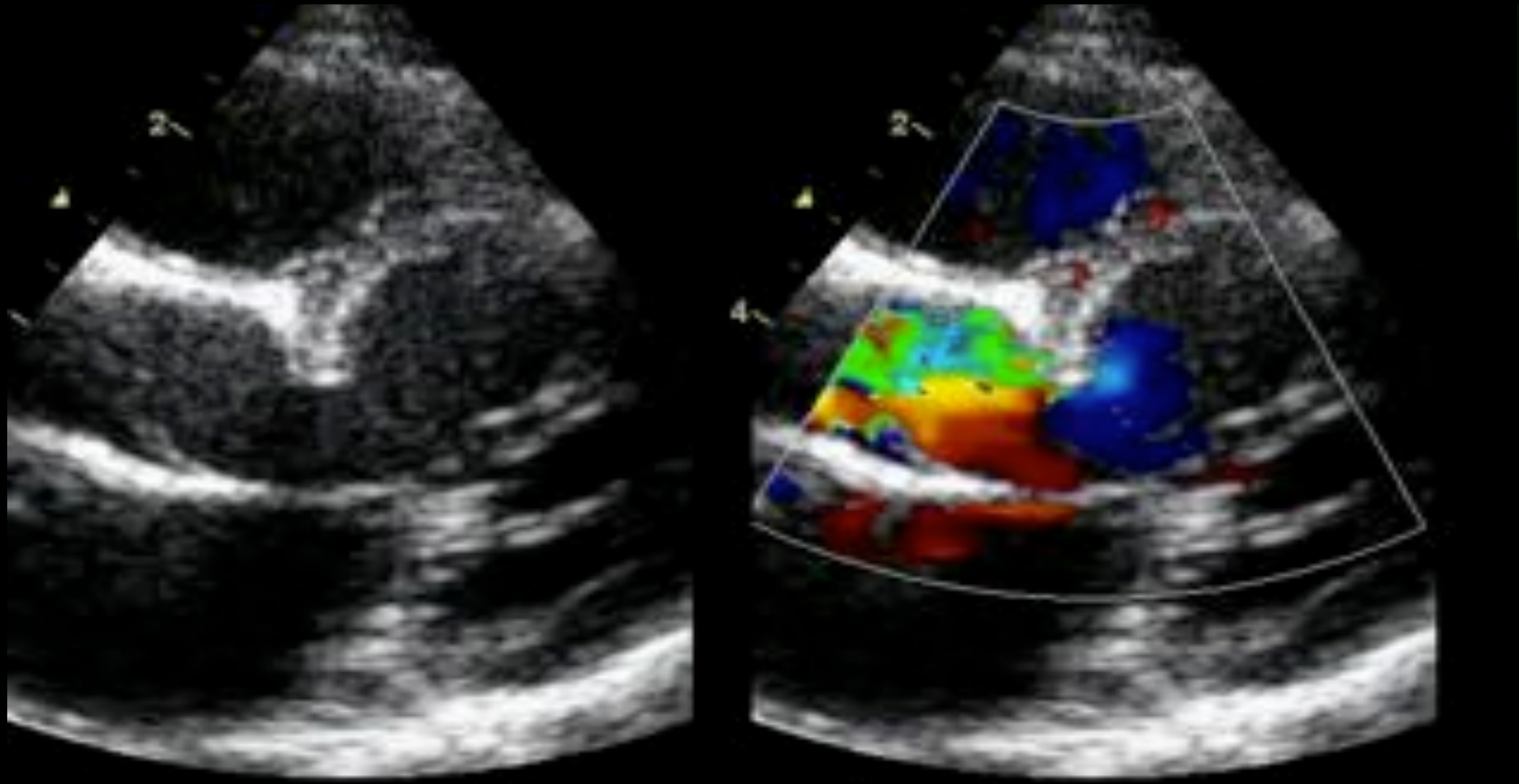

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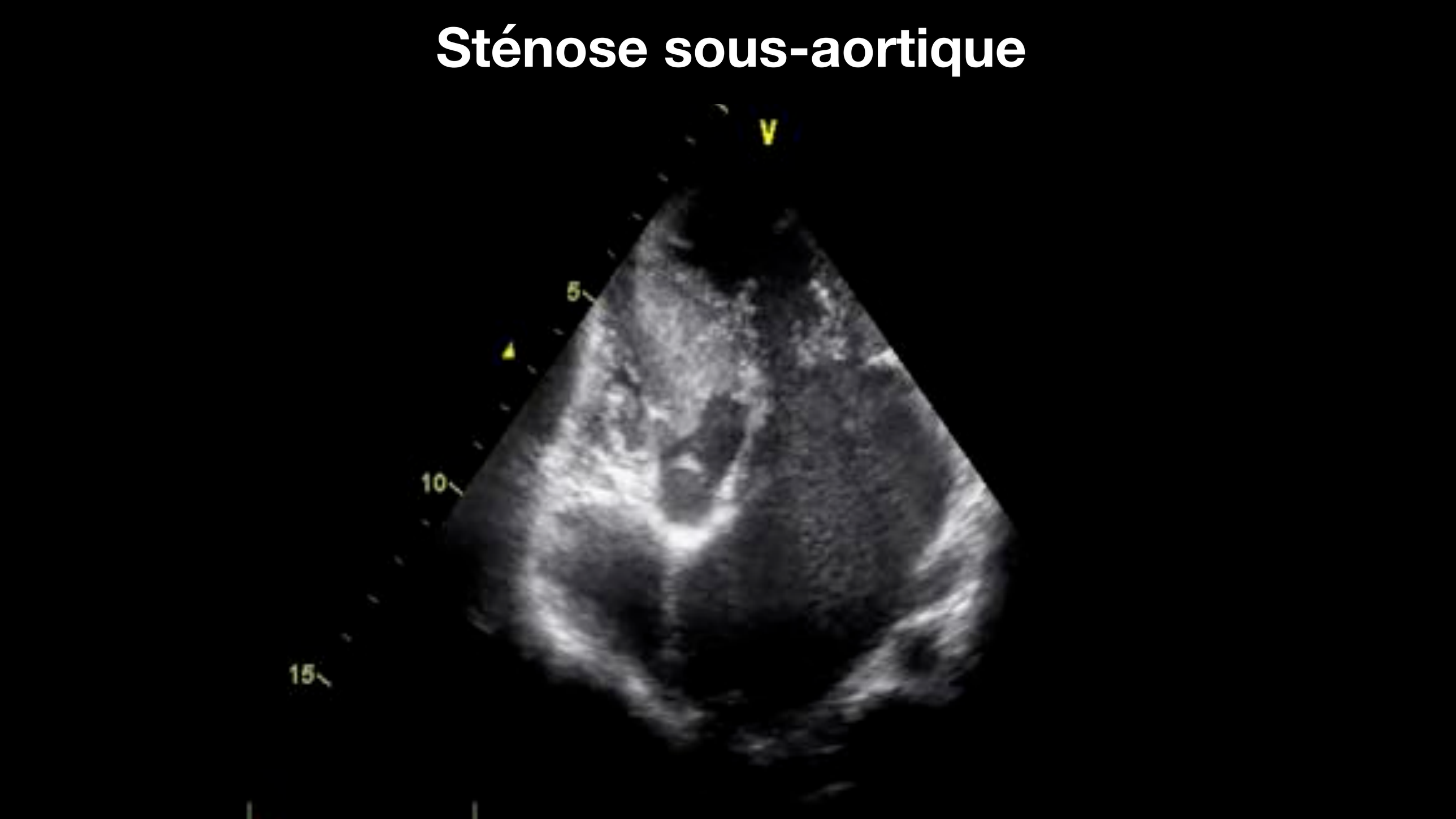
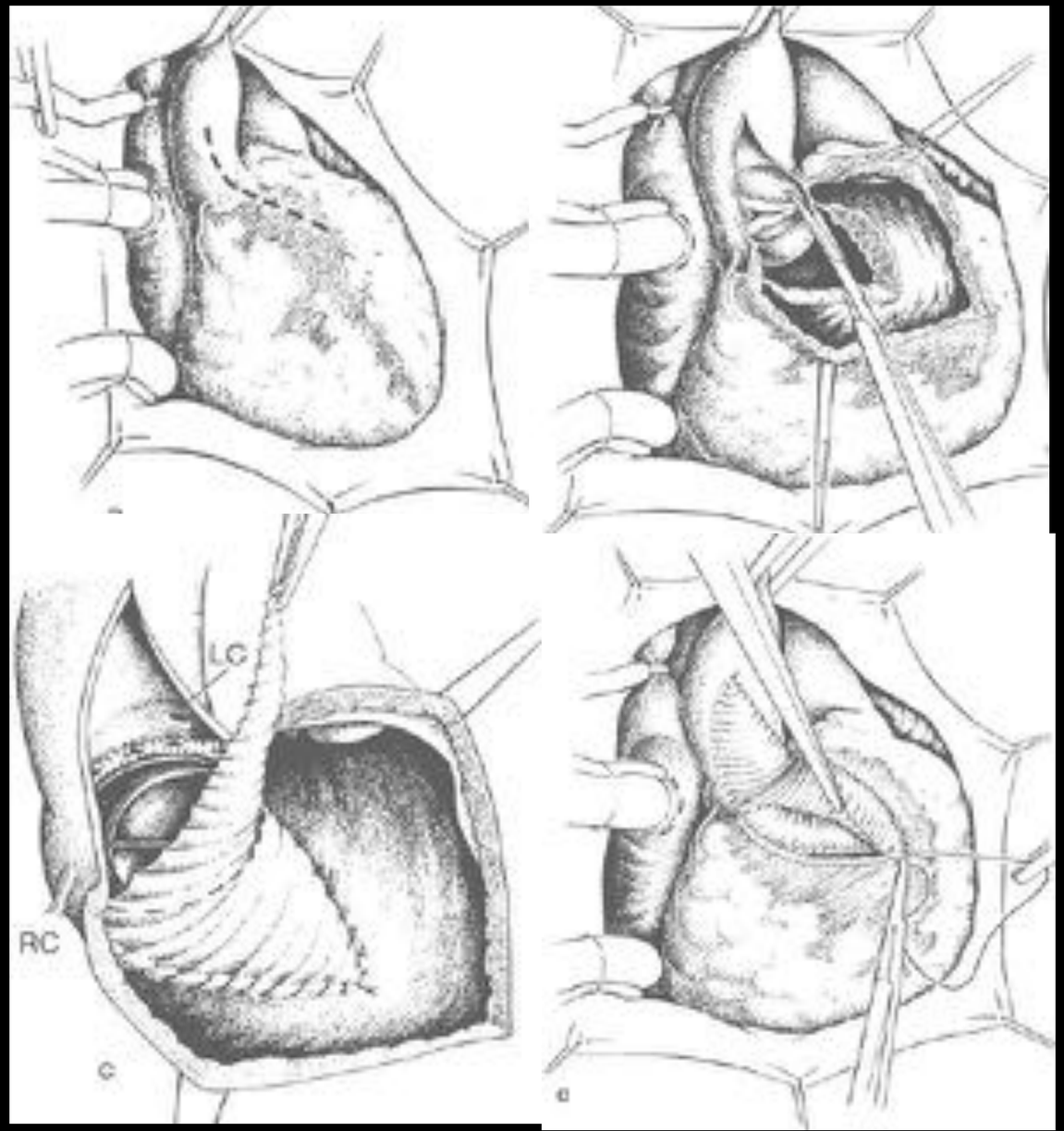
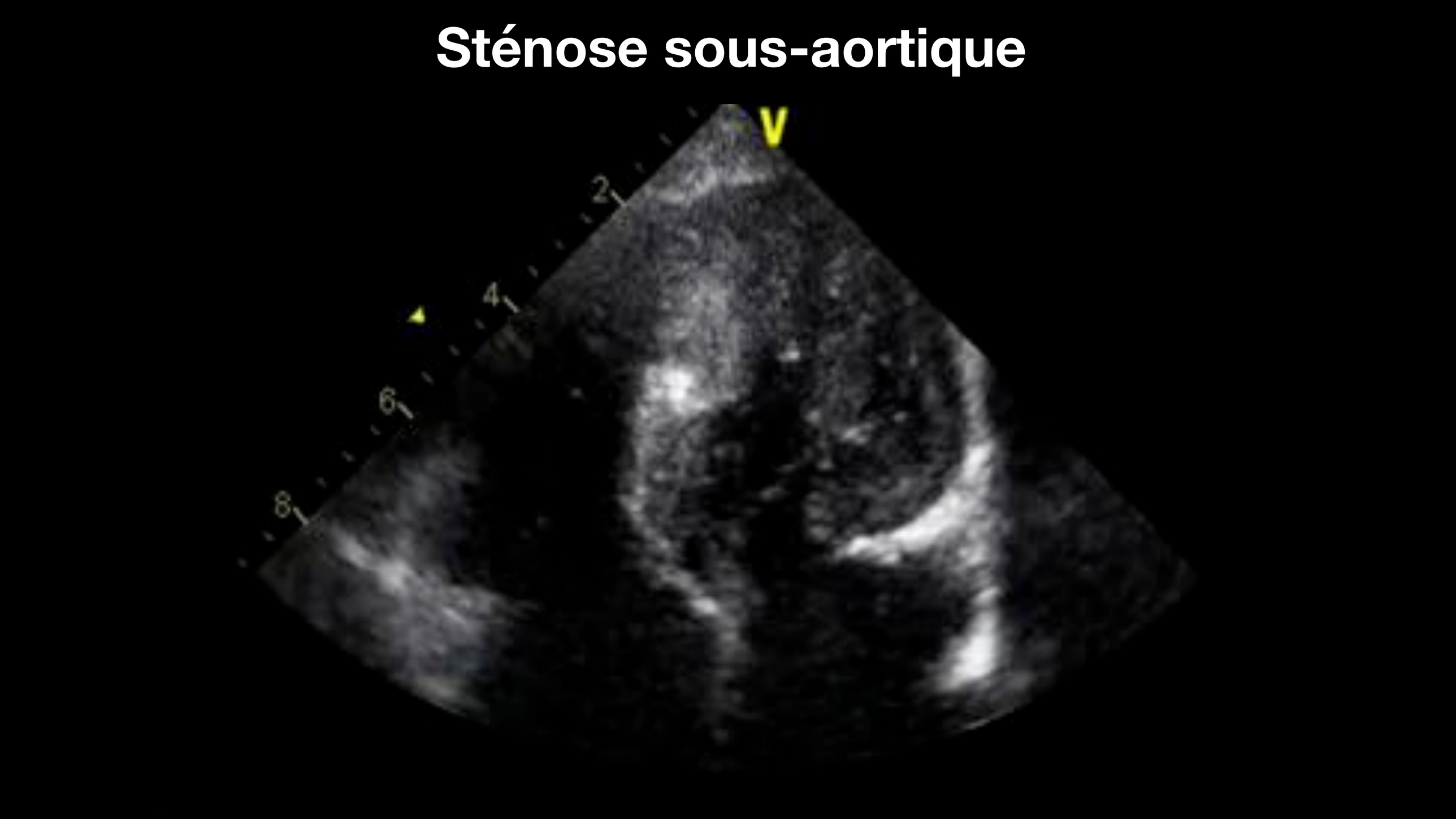


Intervention de Konno modifiée

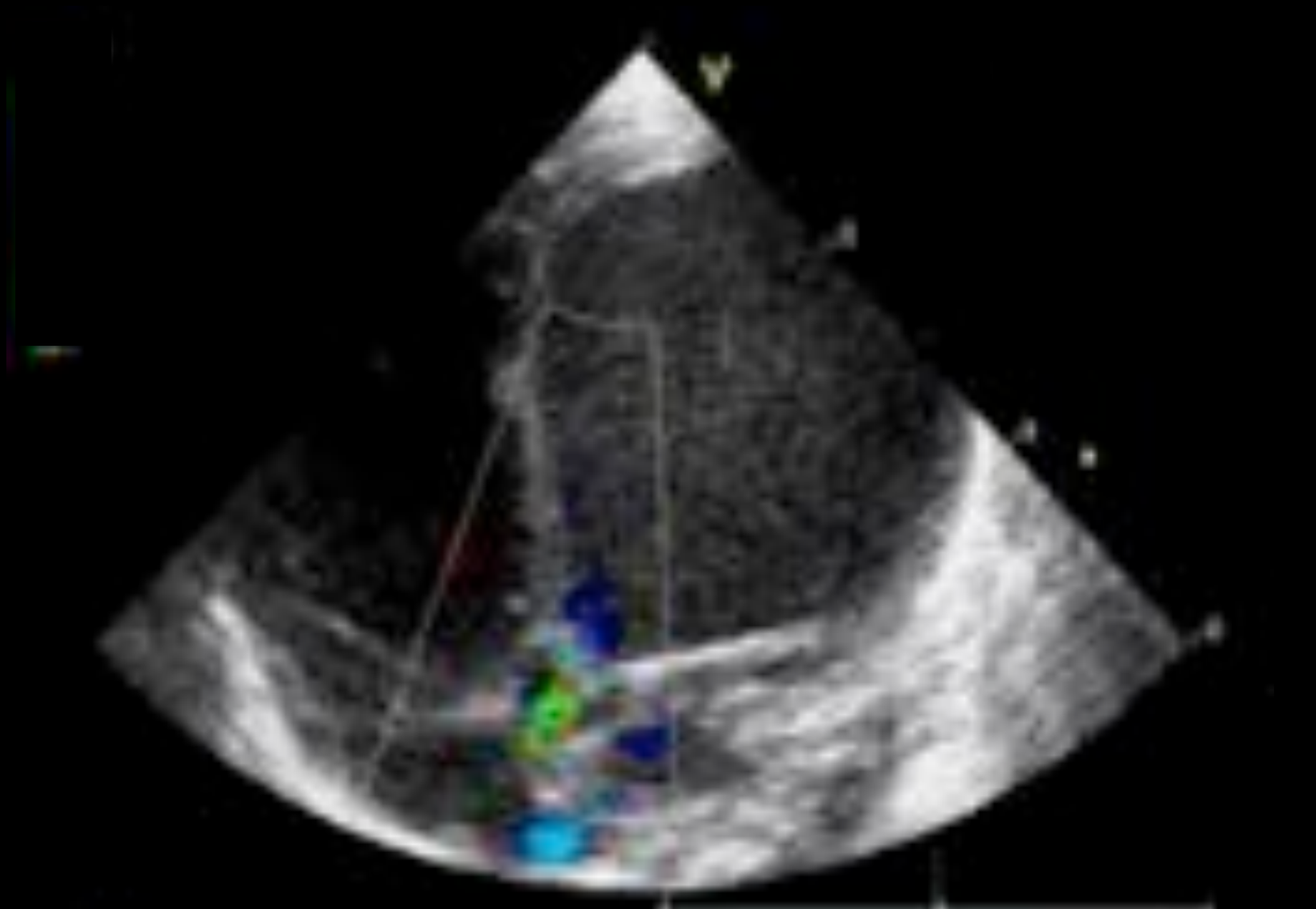


Sténose sous-aortique



Rétrécissement valvulaire aortique

Rétrécissement valvulaire aortique

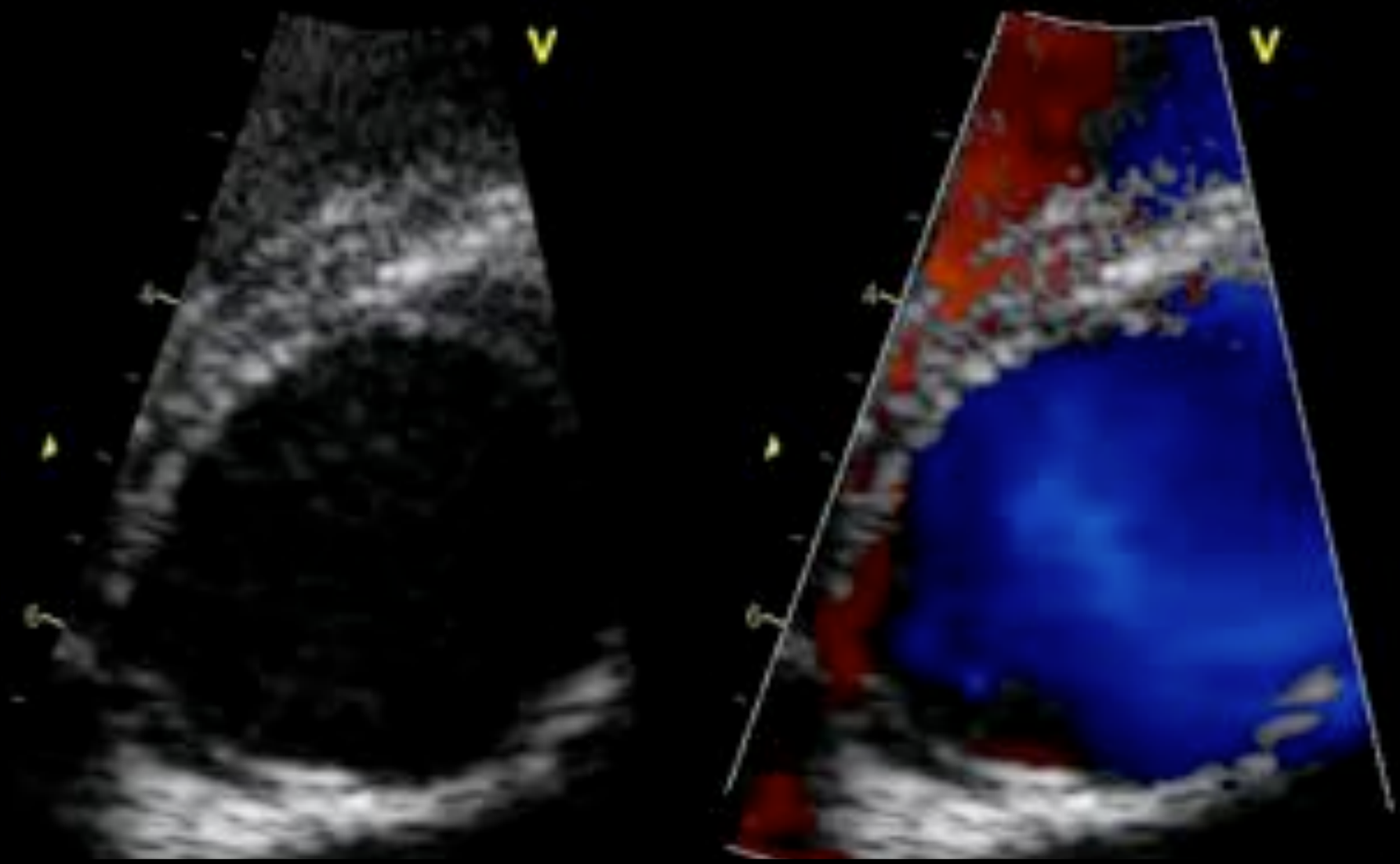


Rétrécissement valvulaire aortique

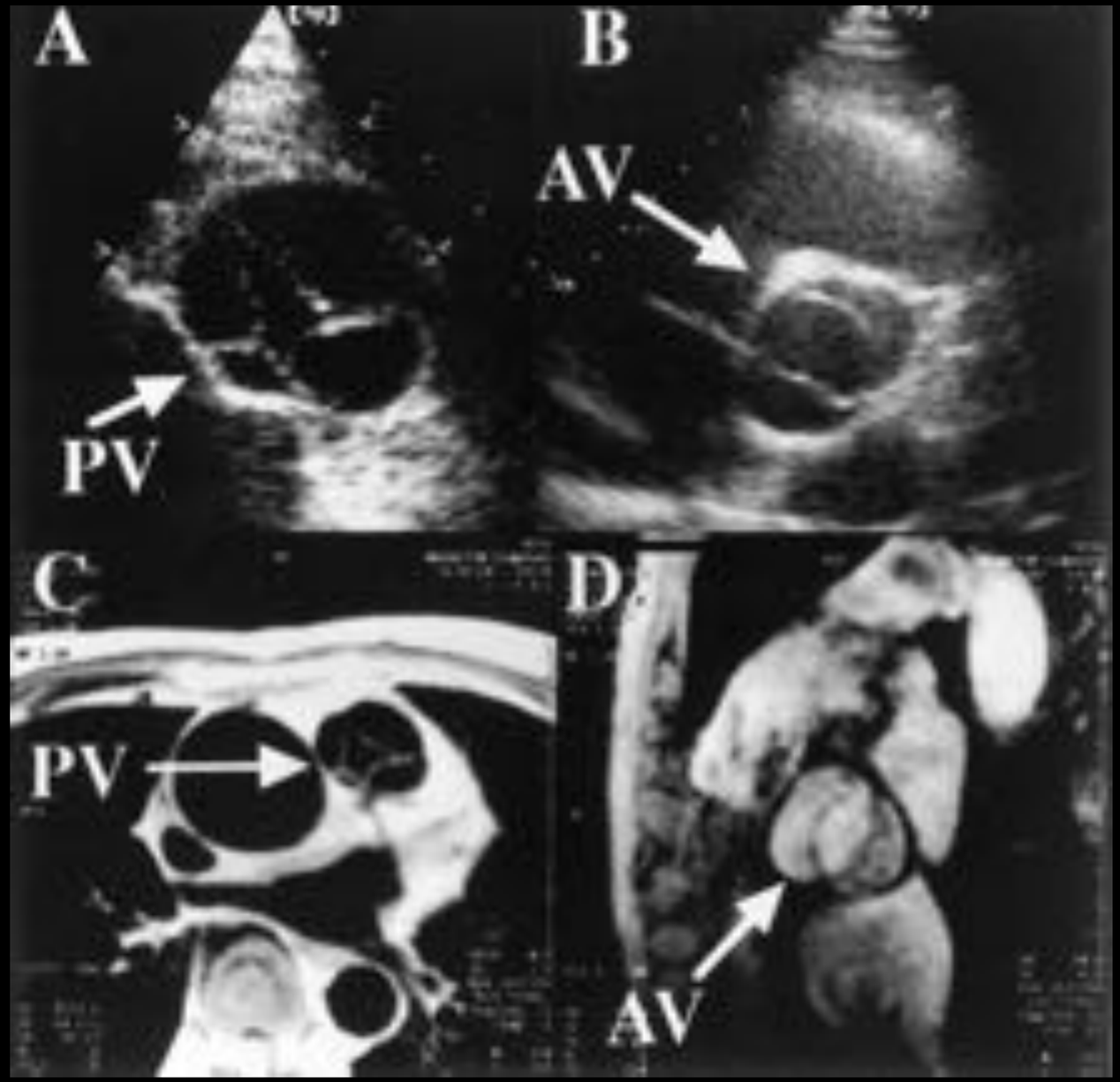
Rétrécissement valvulaire aortique
Bicuspidie type 0 de Sievers

Rétrécissement valvulaire aortique
Bicuspidie type 0 de Sievers

Rétrécissement valvulaire aortique
Bicuspidie type 2 de Sievers



Rétrécissement valvulaire aortique



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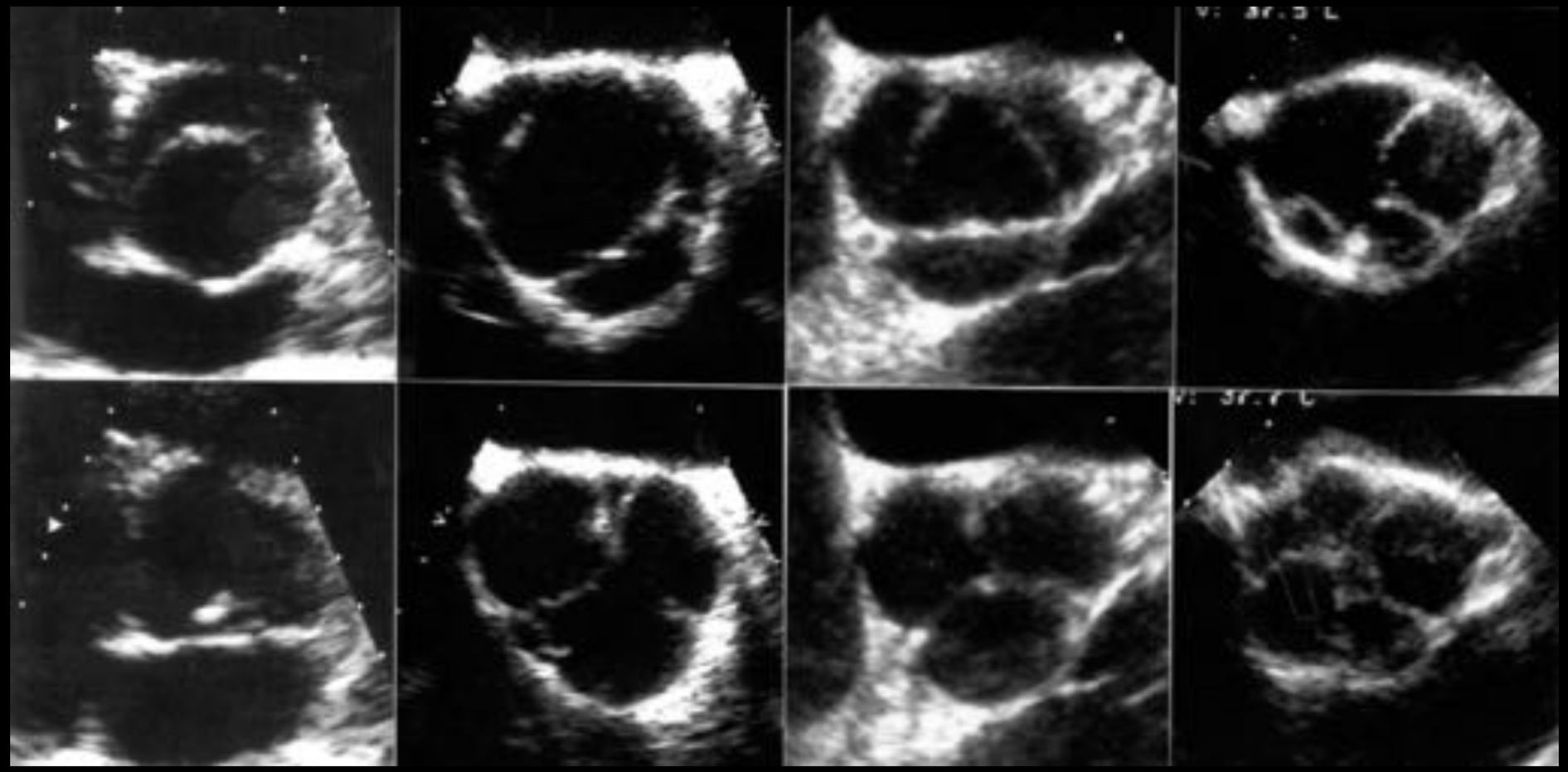
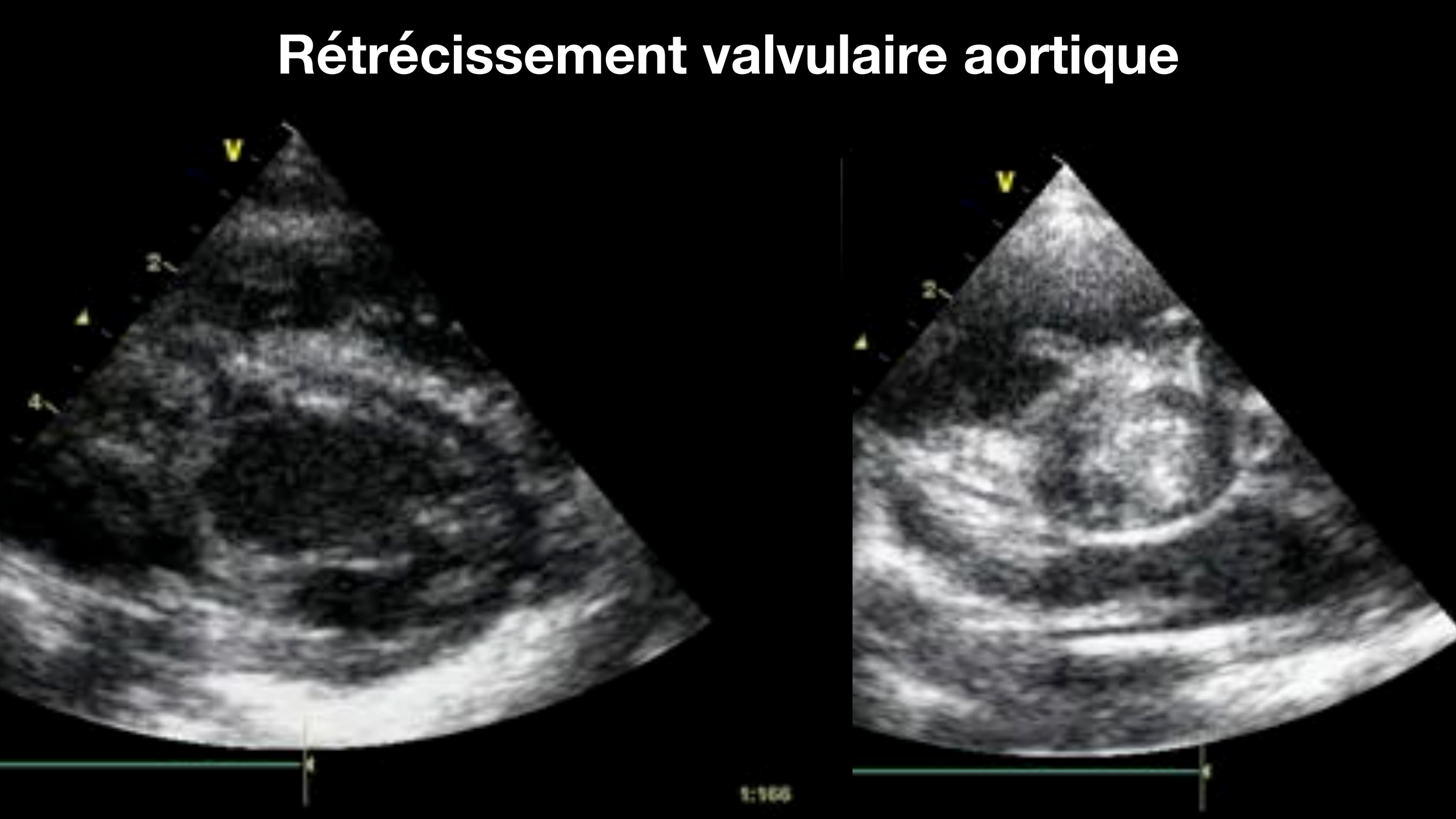
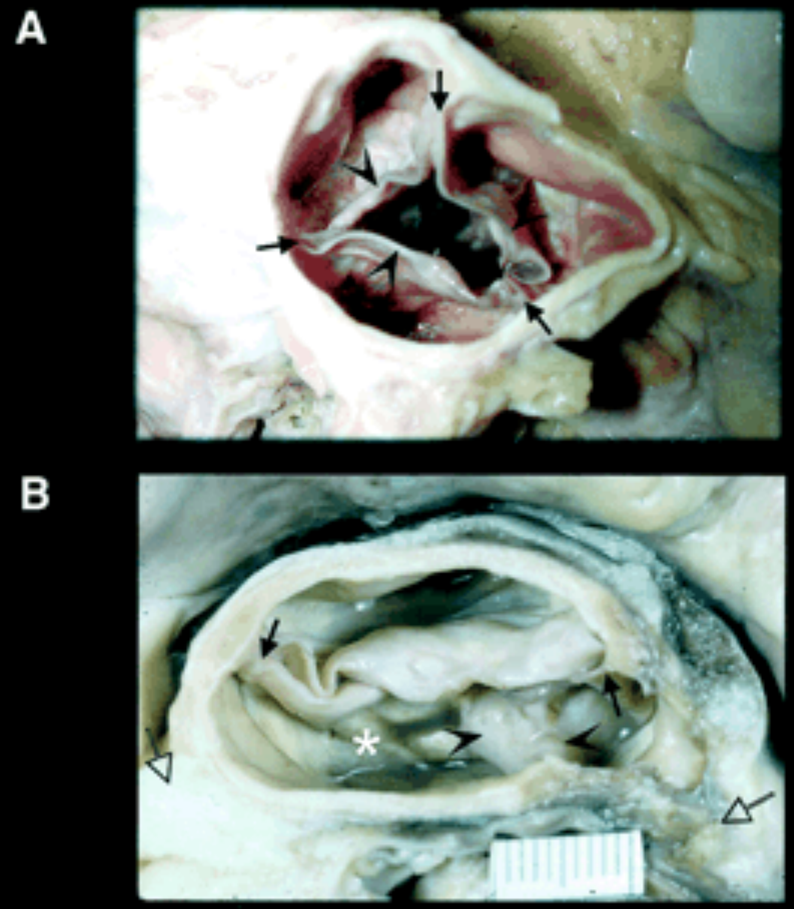
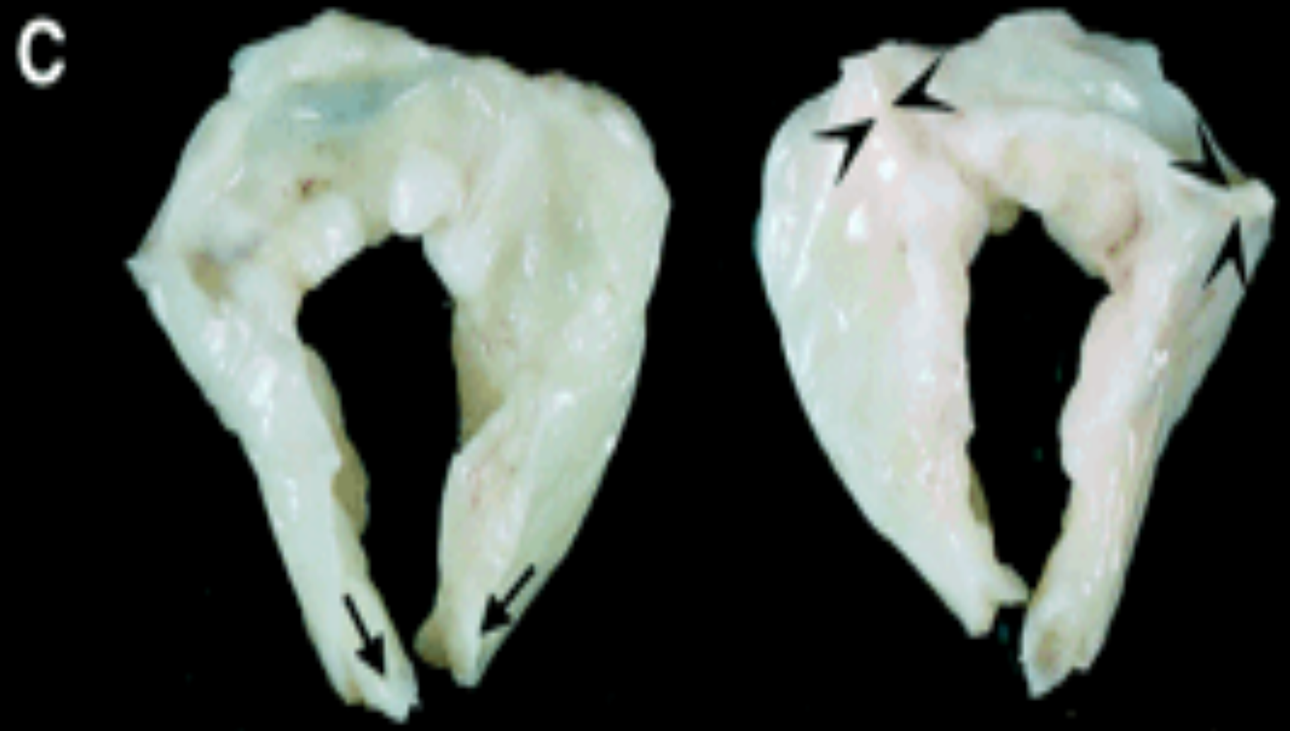


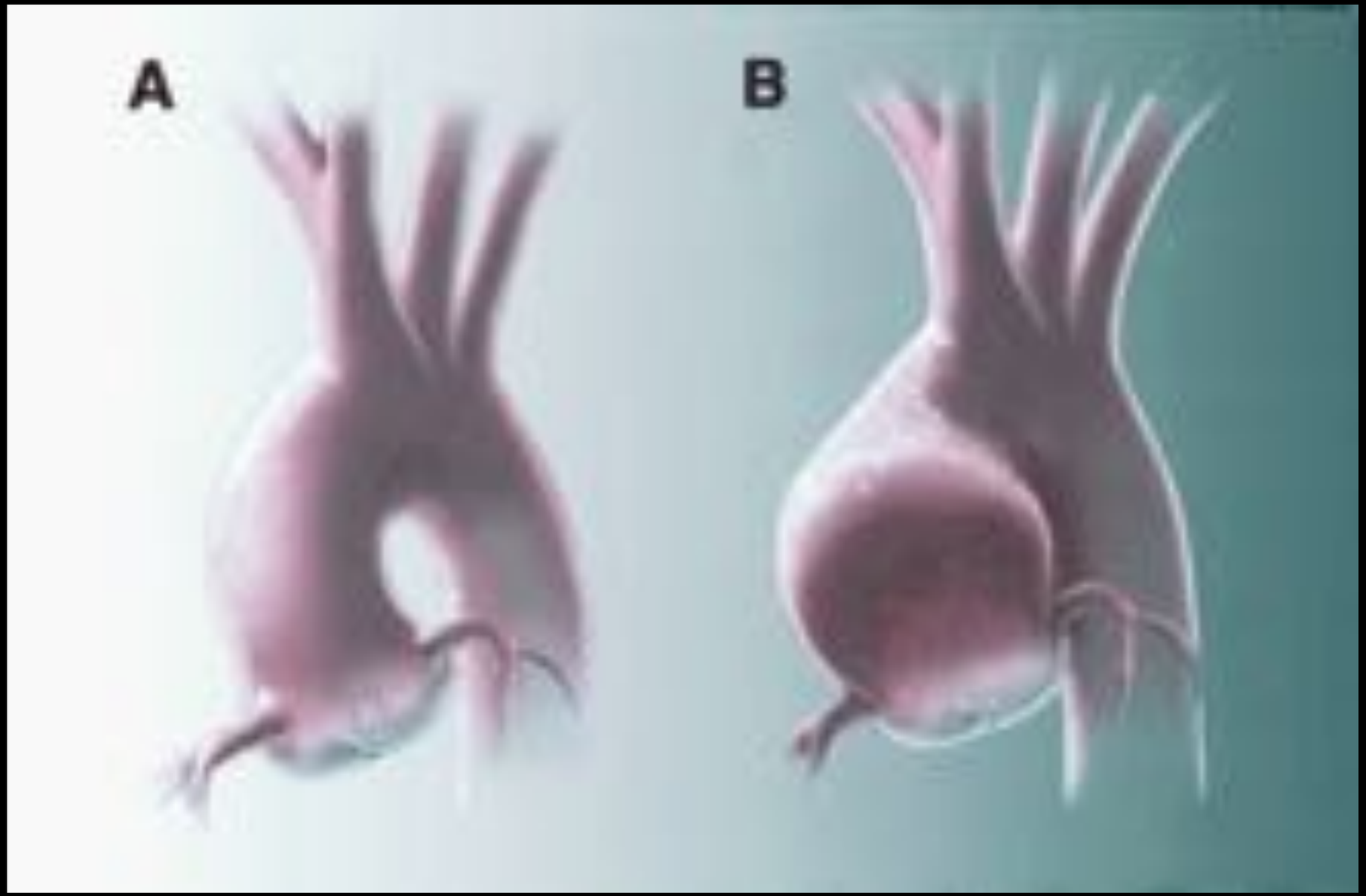
Rétrécissement valvulaire aortique



Rétrécissement valvulaire aortique







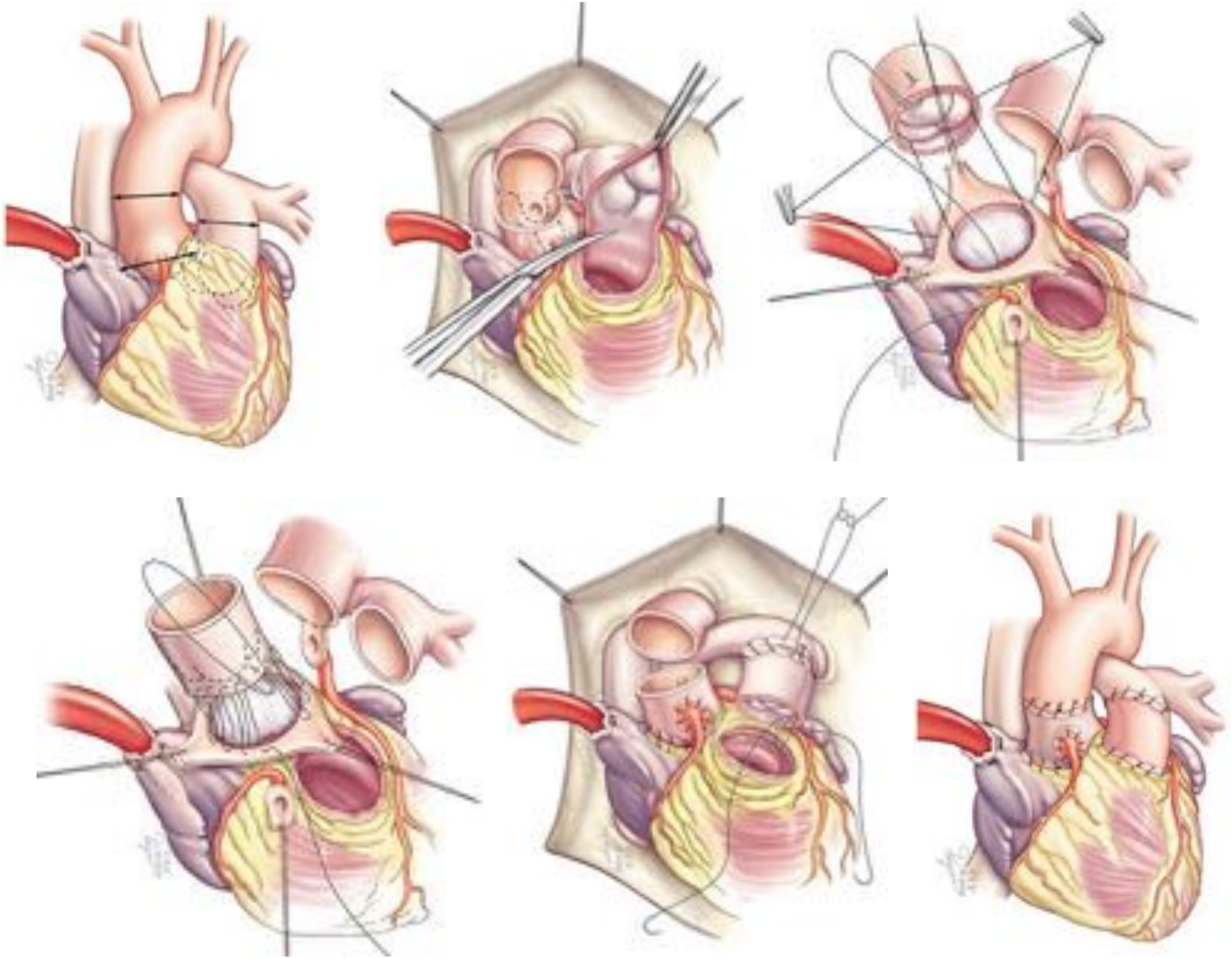
Dilatation du culot aortique et bicuspidie

Traitement des sténoses aortiques

- Valvuloplastie percutanée
- Commissurotomie chirurgicale
- Opération de Ross
- Remplacement valvulaire aortique

Rétrécissement valvulaire aortique

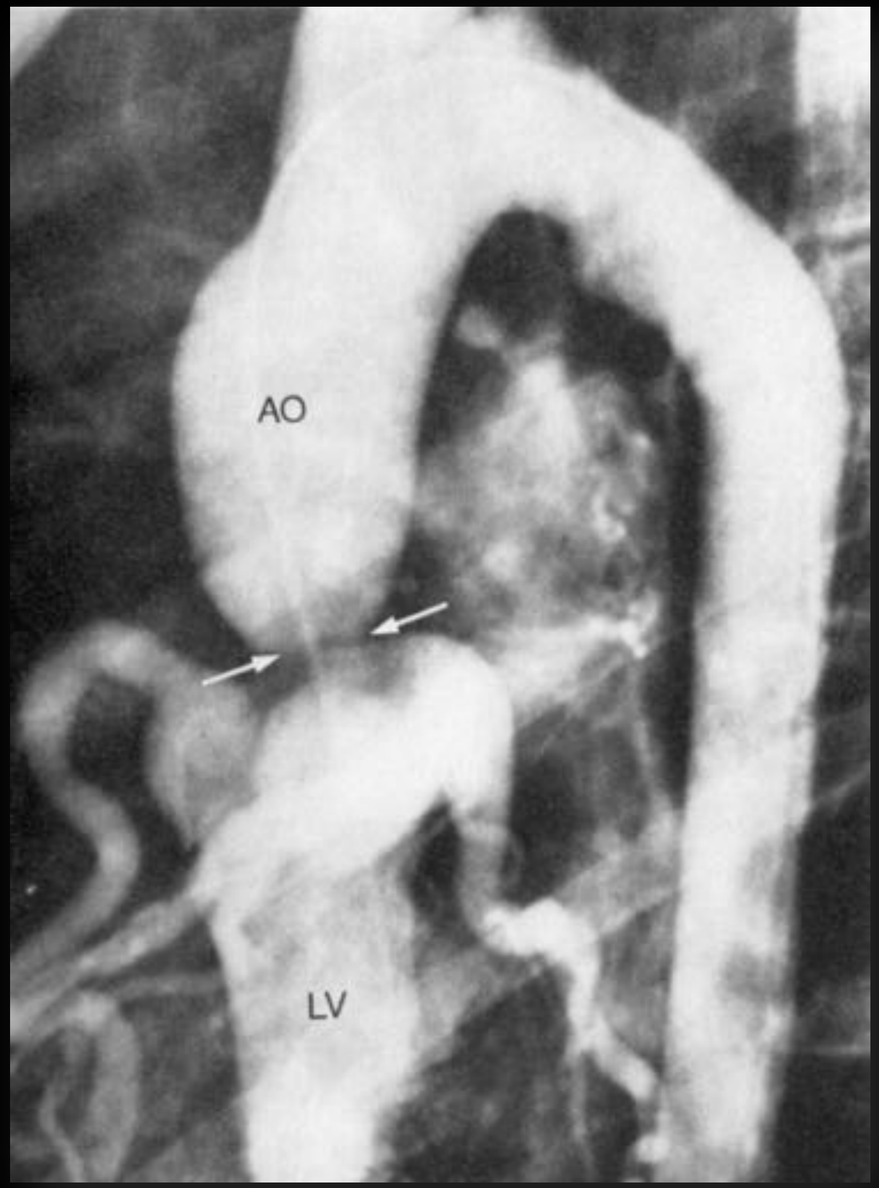
Opération de Ross

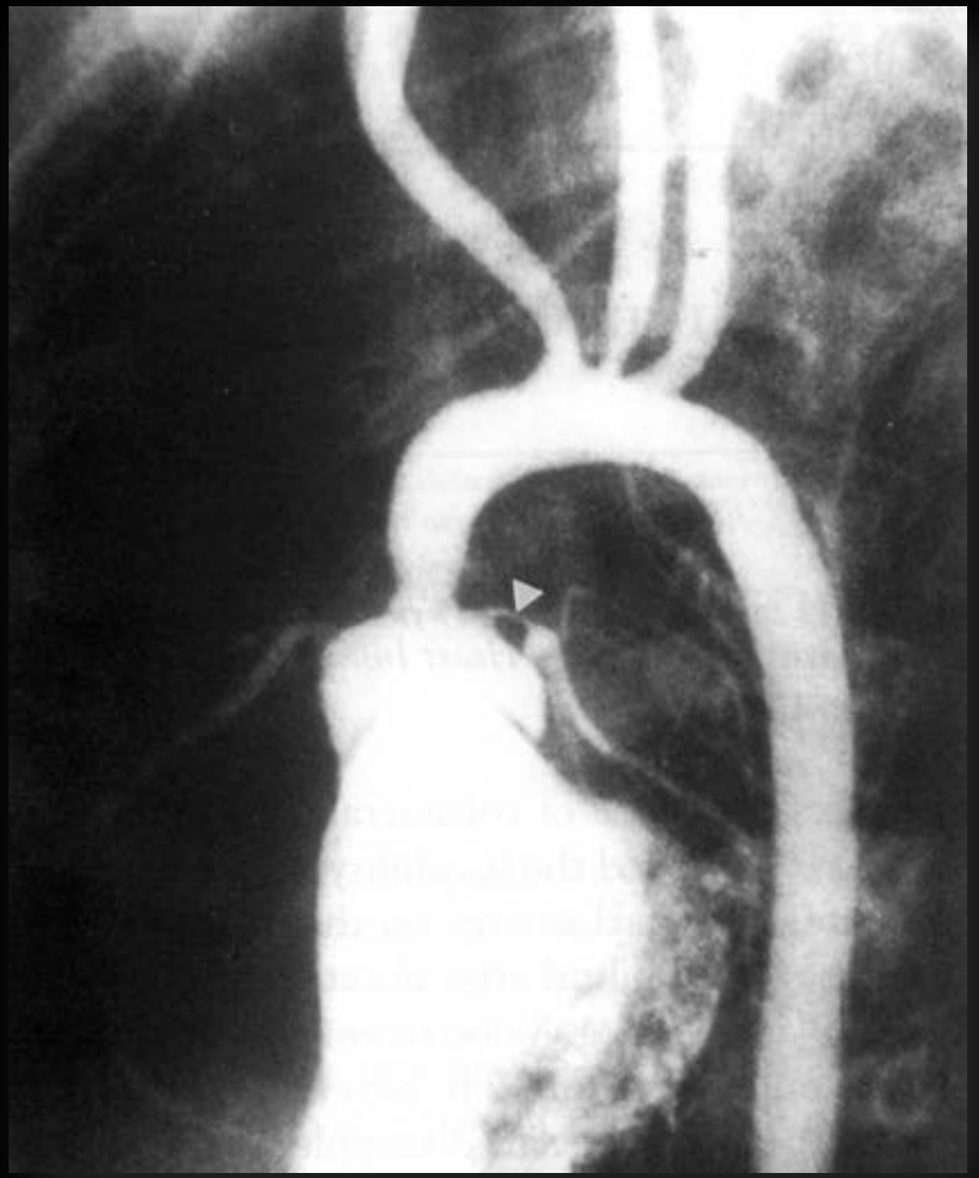


Sténose supravalvulaire aortique

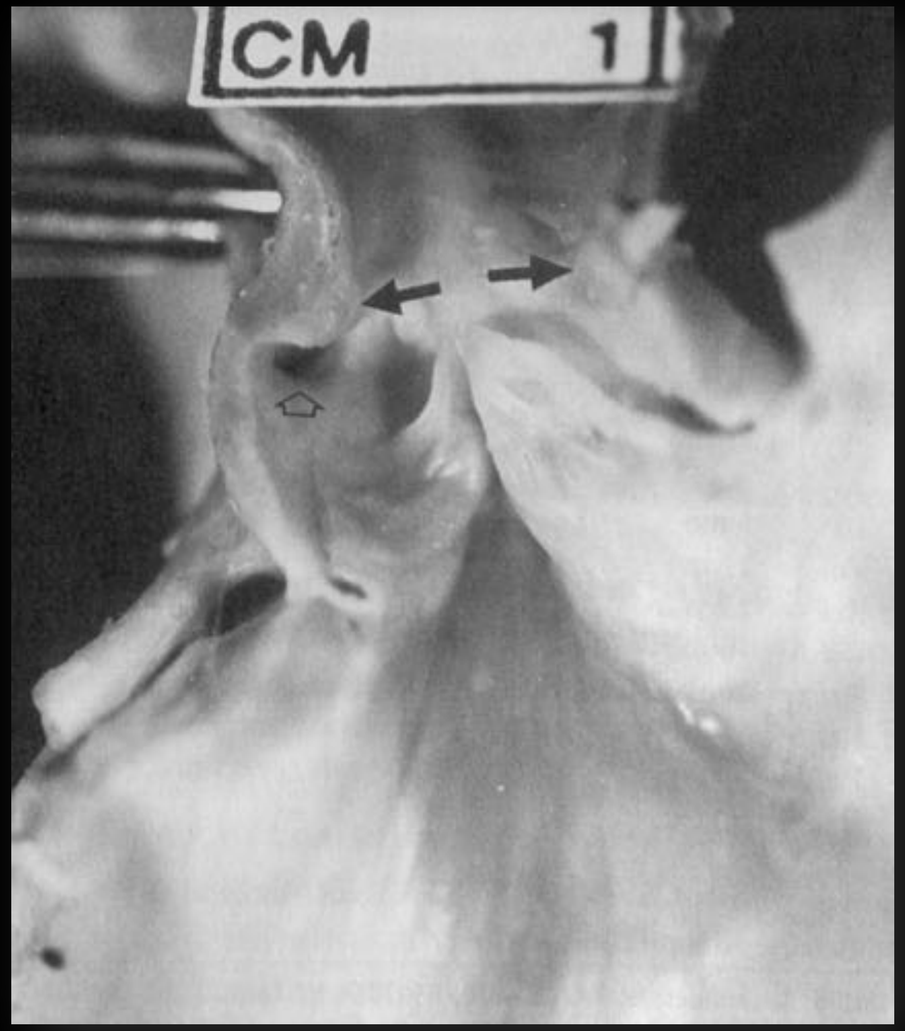

SUNKEN NASAL BRIDGE
PUFFINESS AROUND THE EYES
CAN STILL SEE THE EPICANTHAL FOLD (EYE LID)
BLUE EYES WITH A STARRY PATTERN
LONG UPPER LIP LENGTH (PHILTRUM)
SMALL AND WIDELY SPACED TEETH
WIDE MOUTH (EAR TO EAR SMILE)
PROMINENT LOWER LIP
SMALL CHIN

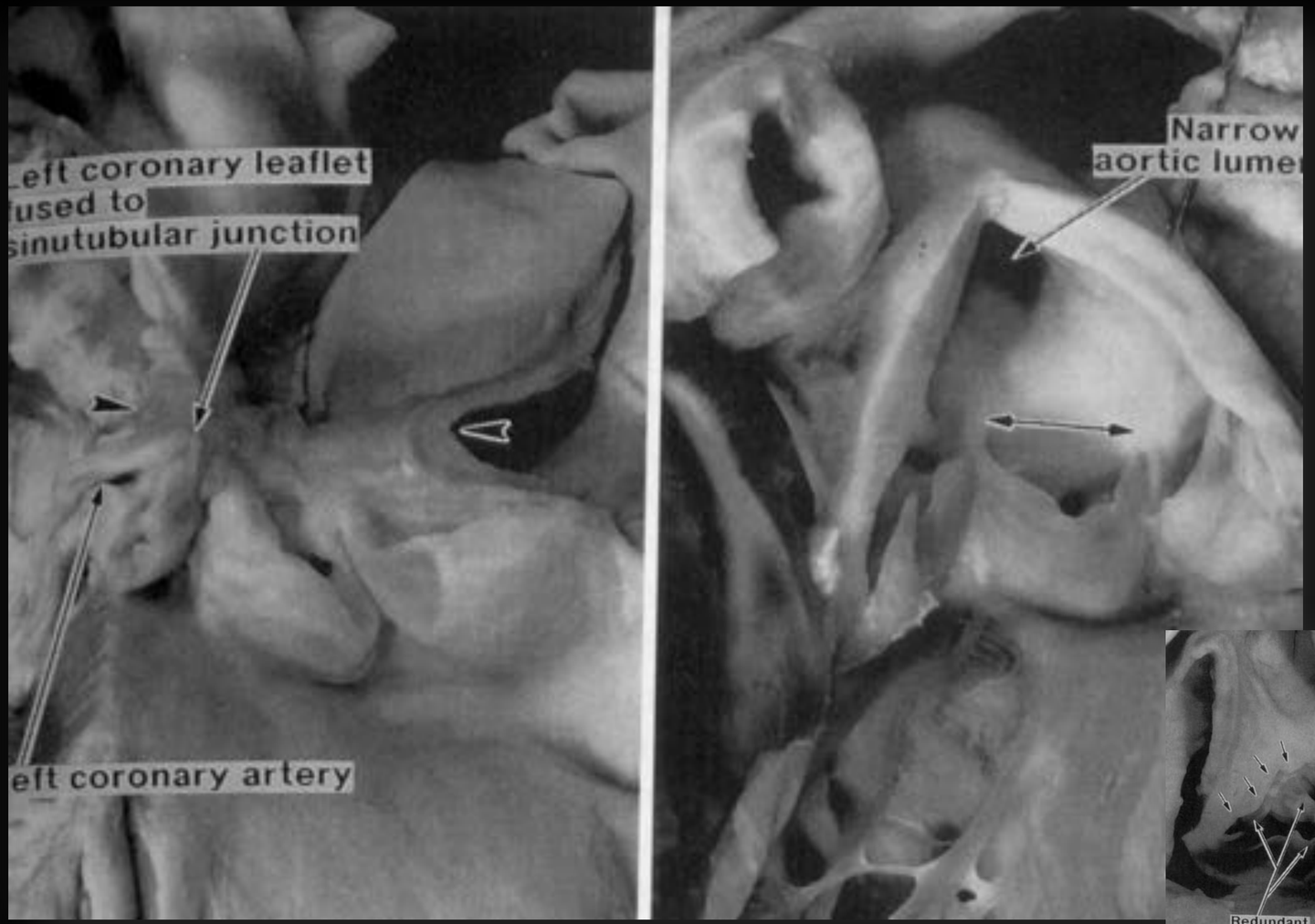
Sténose supravalvulaire aortique





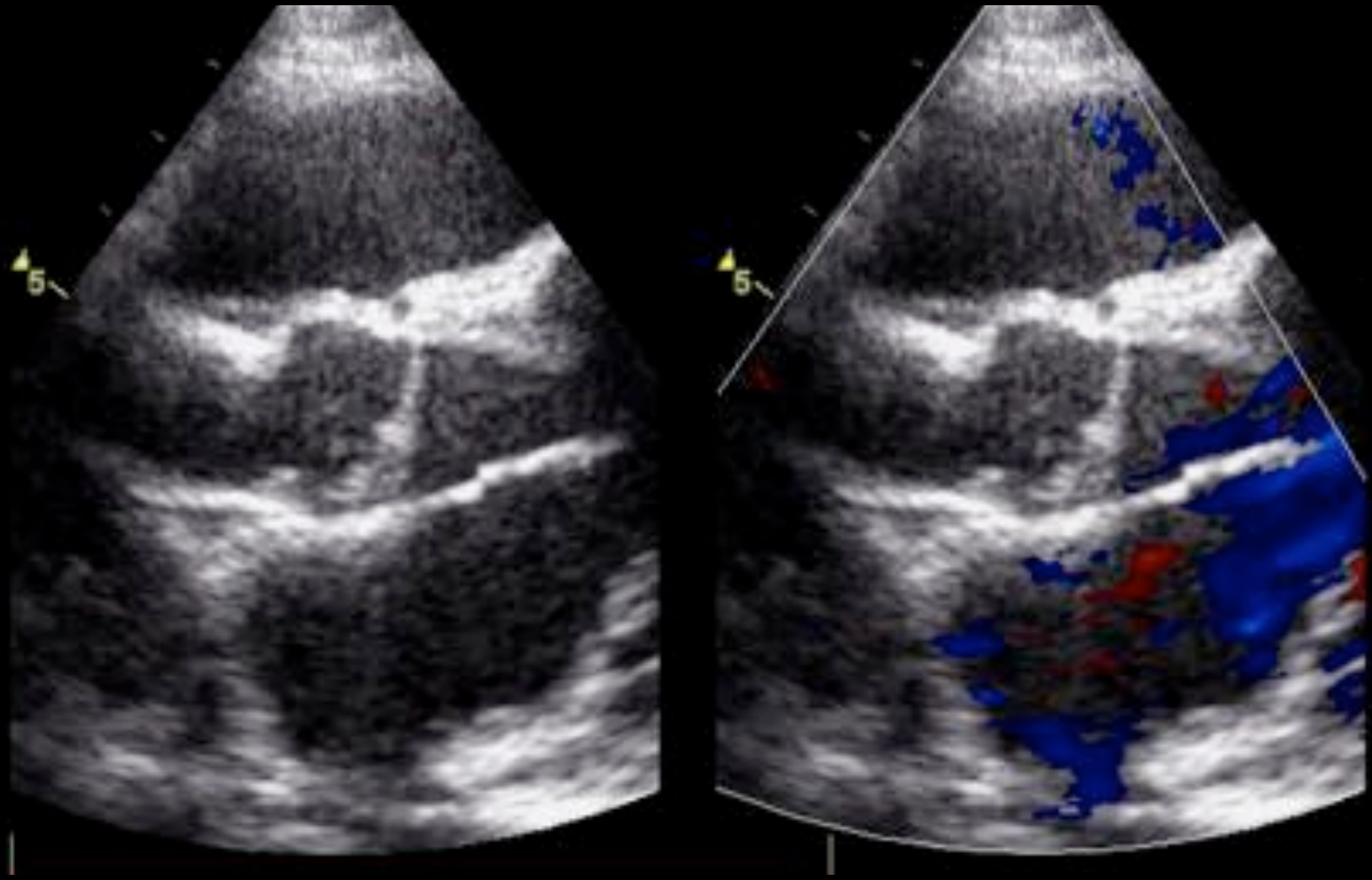
Sténose supravalvulaire aortique





Sténose supravalvulaire aortique

Sténose supravalvulaire aortique

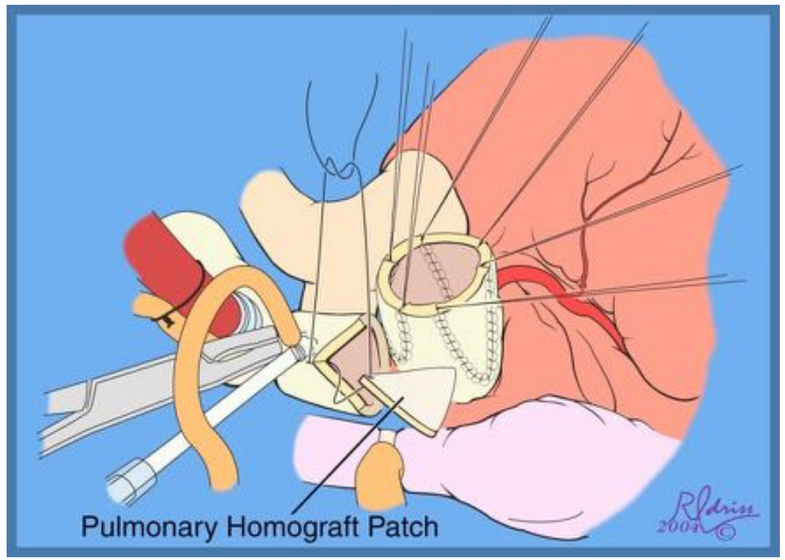


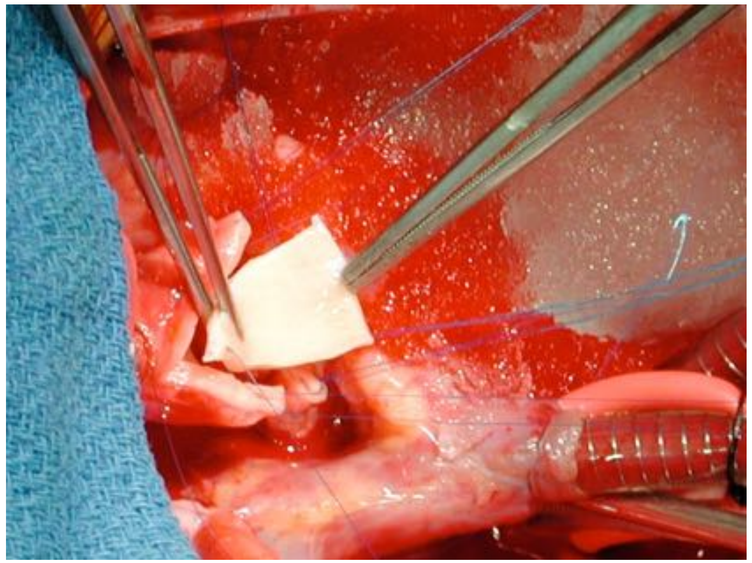
Sténose supravalvulaire aortique

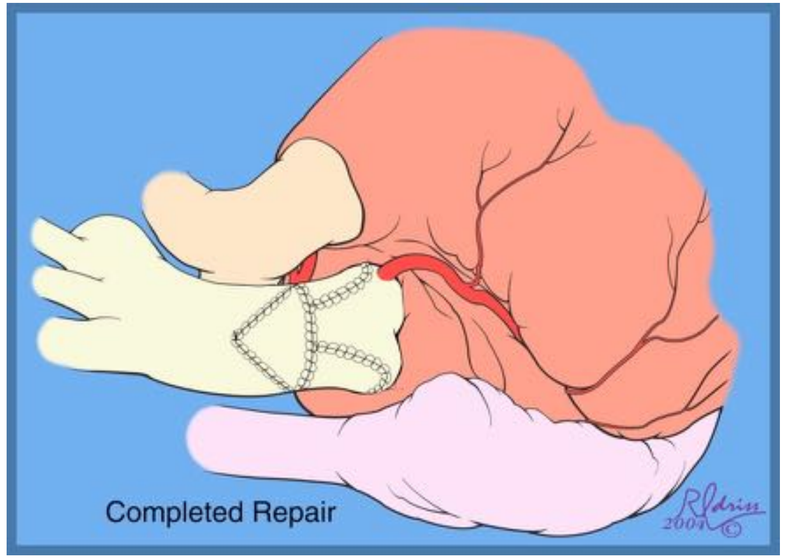
Sténose supravalvulaire aortique

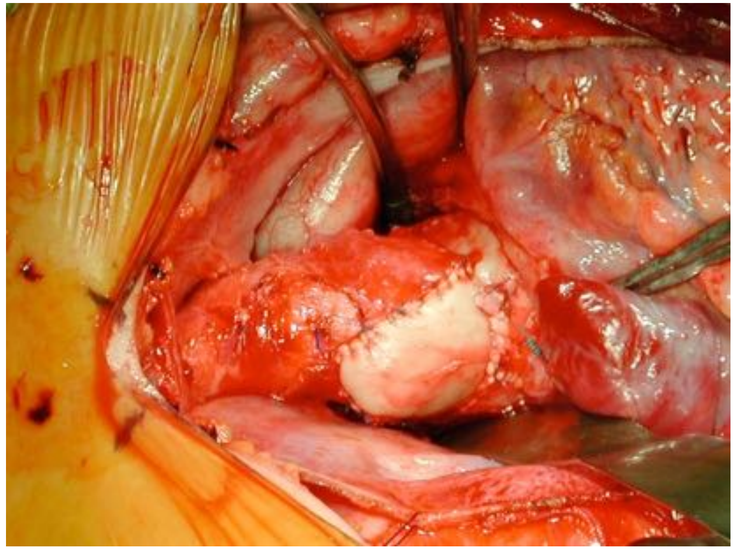
Sténose supravalvulaire aortique

Opération de Brom

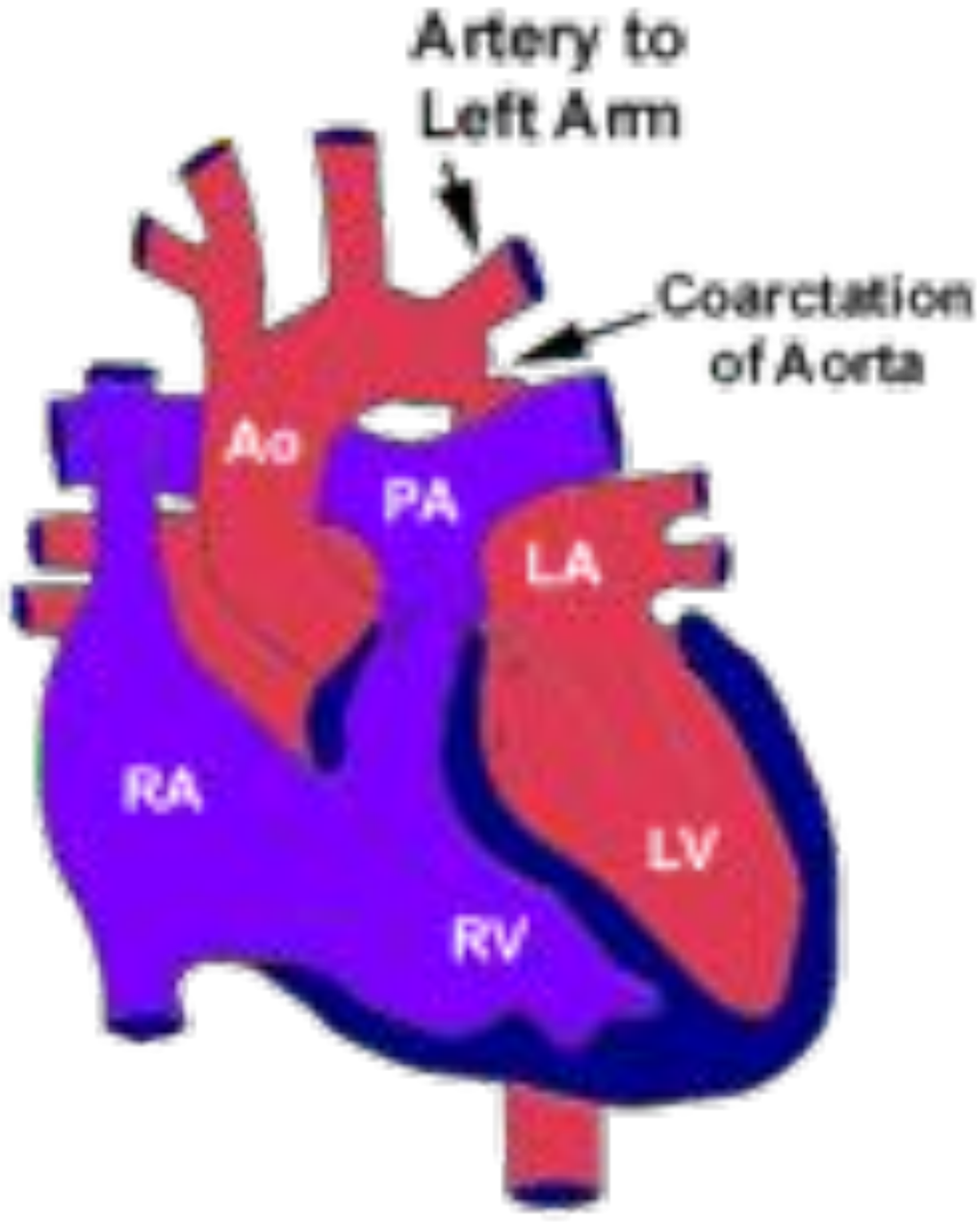




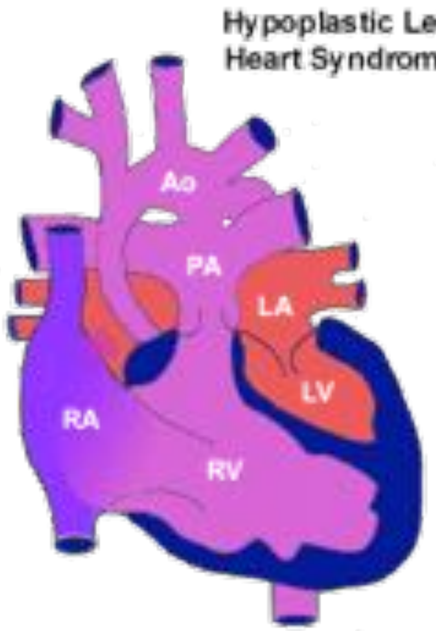


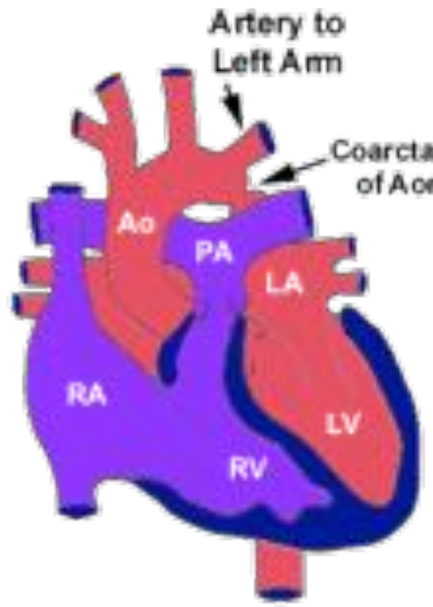


Coarctation isthmique


Artery to
Left Arm
Coarctation
of Aorta
Ao
PA
LA
RA
LV
RV

Notion de continuum phénotypique



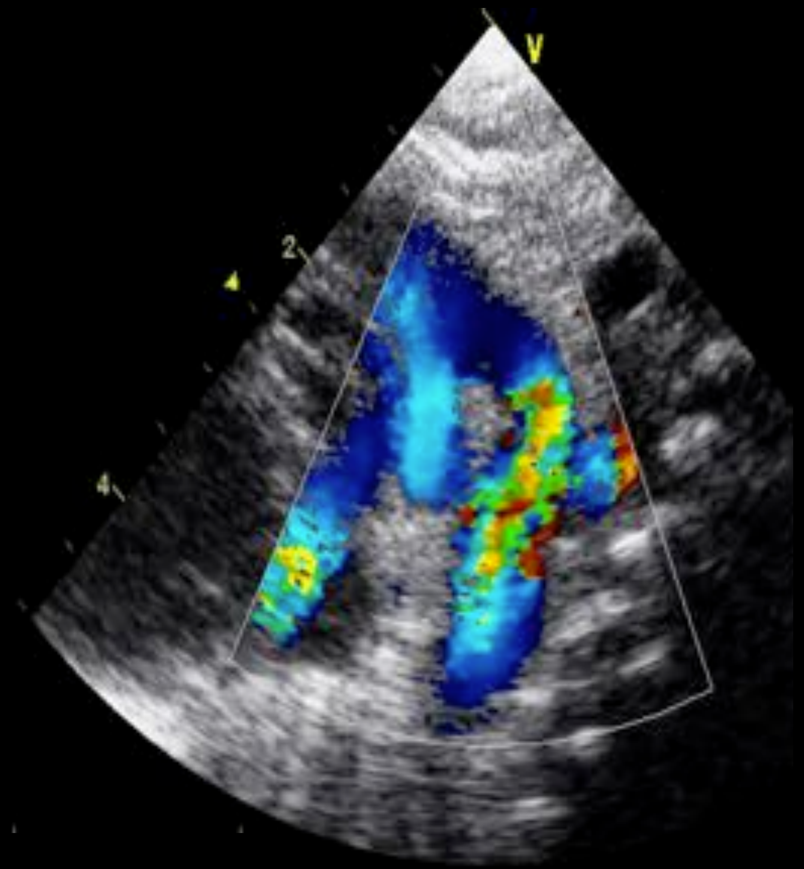


Bicuspidie aortique

Stratégie chirurgicale

- Ducto-dépendance ou défaillance cardiaque
  - Chirurgie néonatale
- Hypertension artérielle
  - Chirurgie
- Tout va bien
  - Chirurgie entre 1 et 3 mois

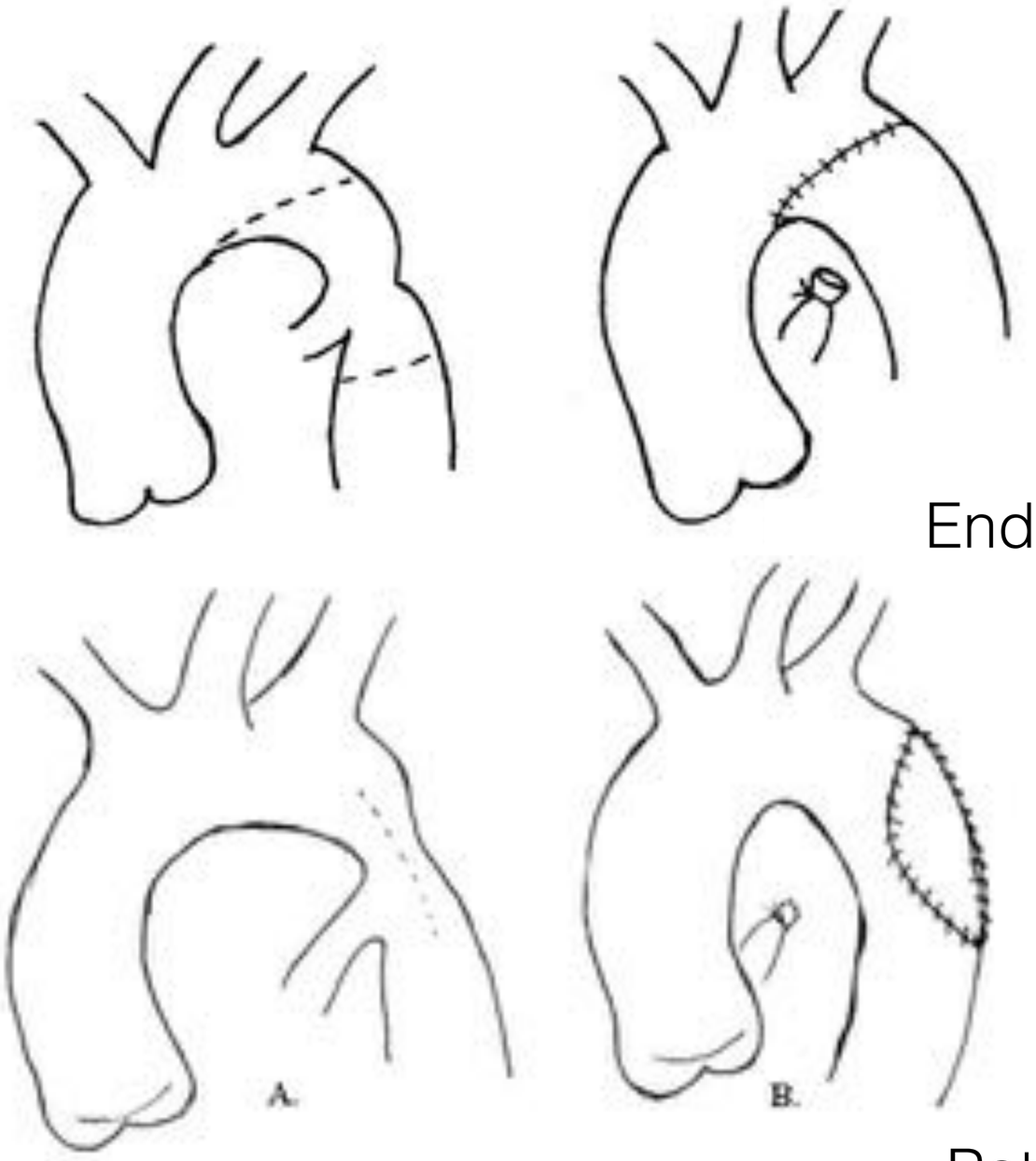
Perfusion systémique ducto-dépendante ?




V
2
4
6

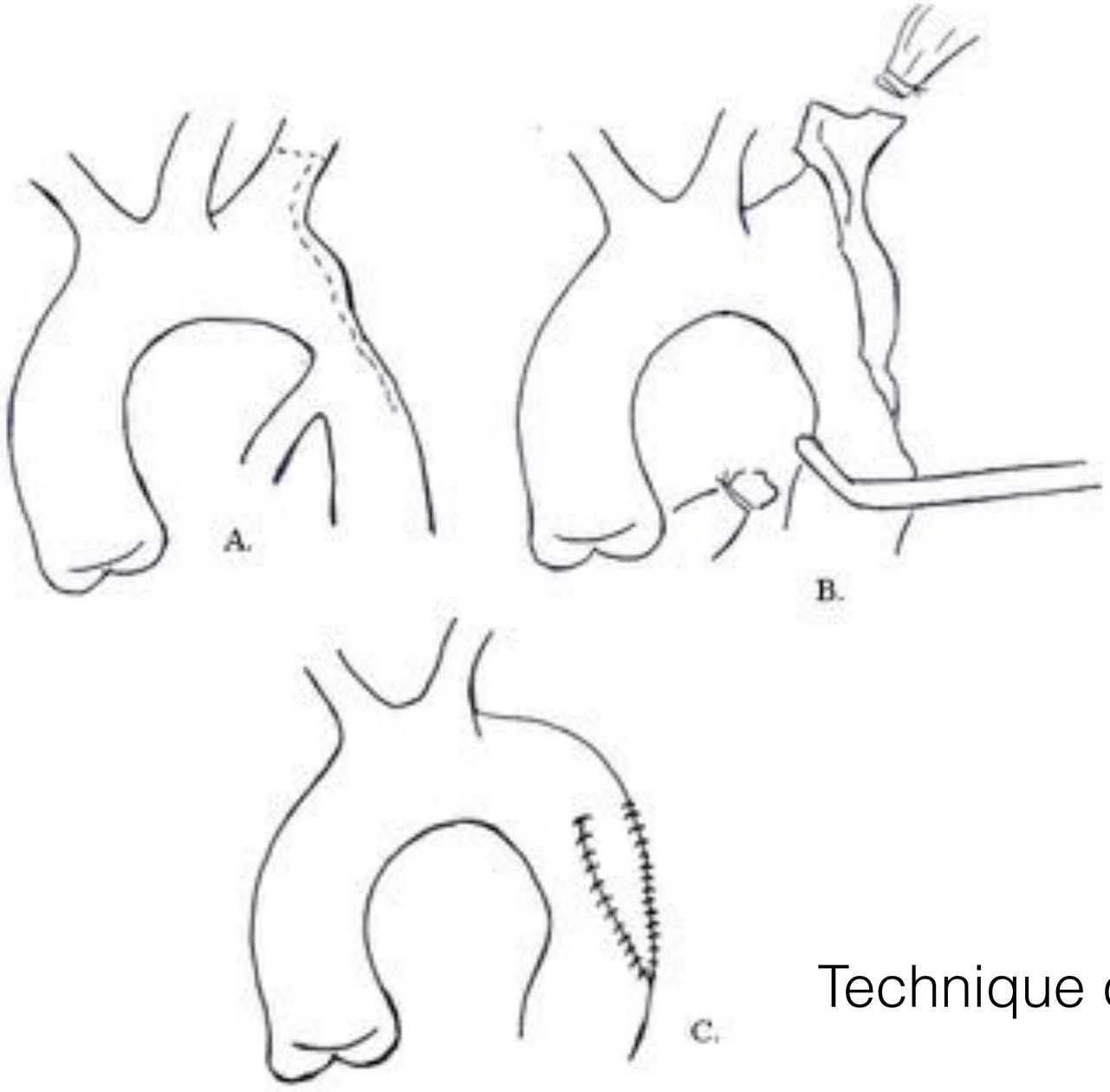

SLA
HOPITAL NECKER ENFANT
3 W 1406012256
Mar 07 2006
P
L
S
IRP


5.5 mm (2D)
4.5 mm (2D)

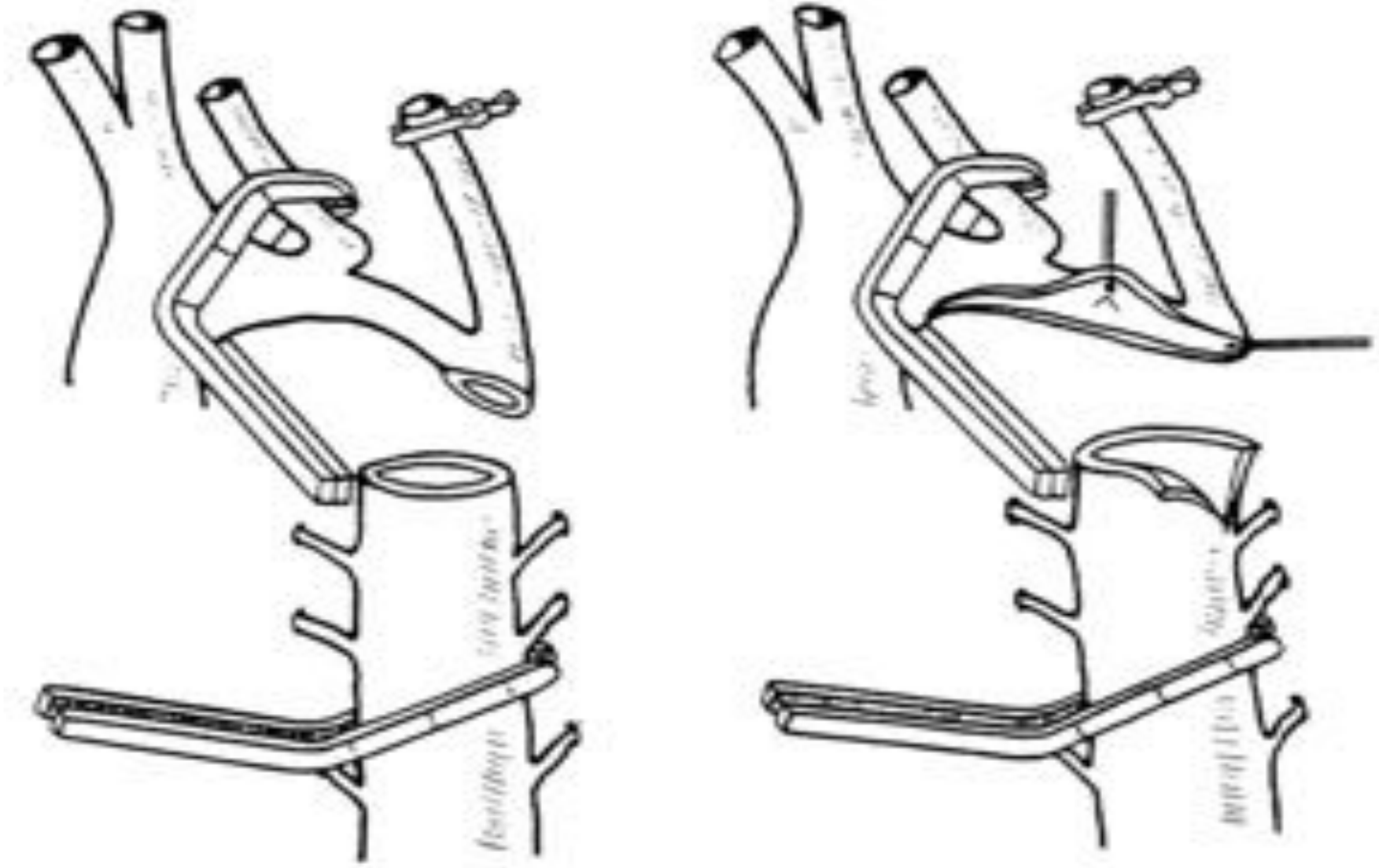


End-to-end anastomosis

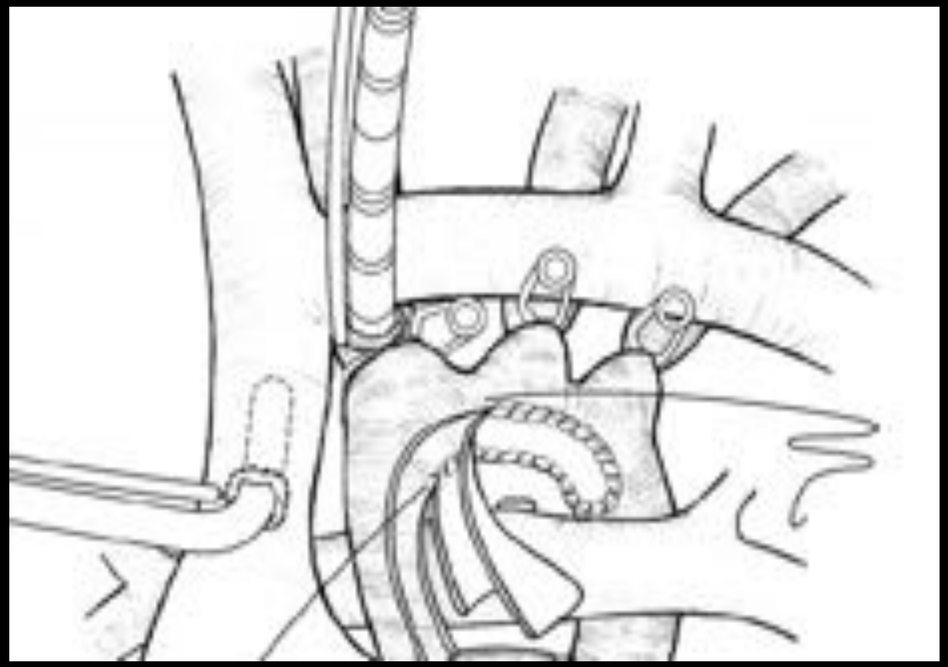
Patch



Technique de Waldahausen



Extended end-to-end anastomosis



Repair with selective brain perfusion

Arterial tortuosity syndrome

Descending aorta coarctation

Osler of coarctation

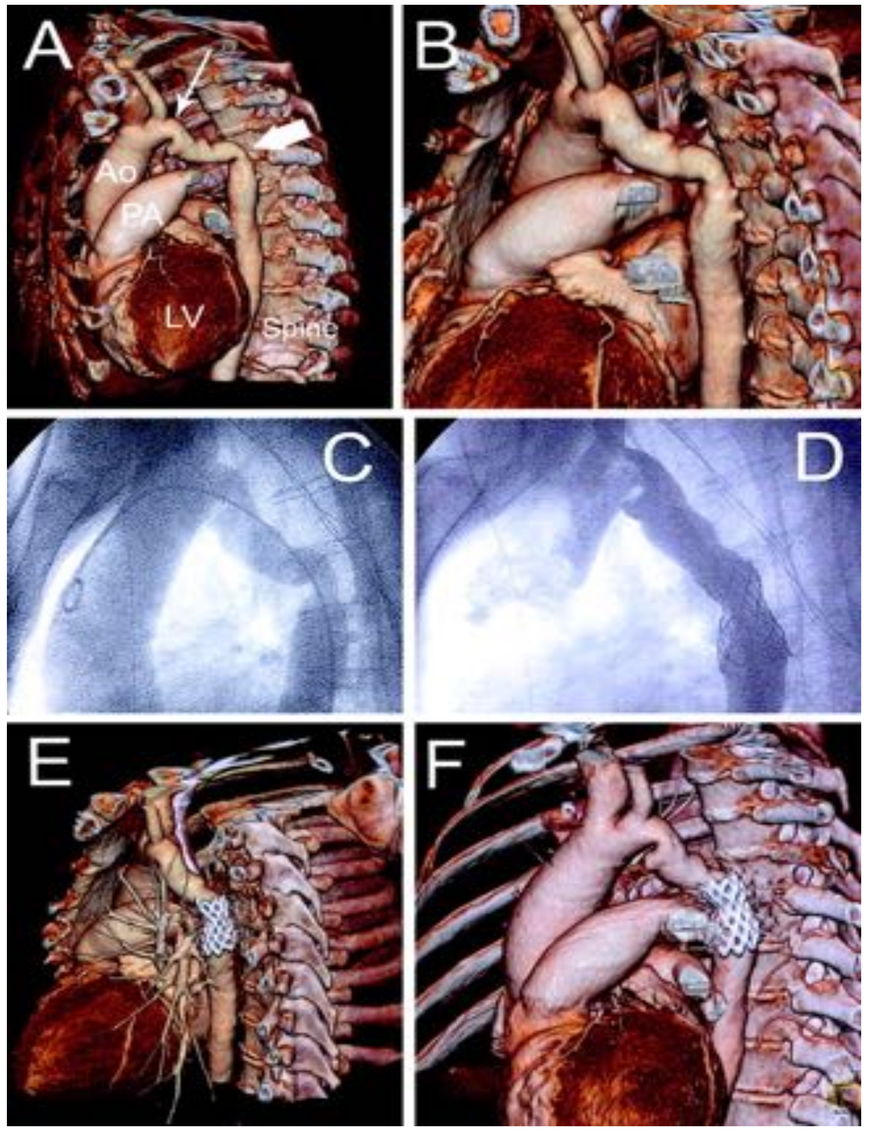
Other complex anatomical forms of coarctation of the aorta

Les anomalies associées

- La valve aortique
  - Hypoplasie annulaire, sténose, bicuspidie
- La voie sous aortique
  - Désaxation et membrane sous-aortique
  - Anomalies d'insertion mitrale
- La valve mitrale
  - RM ou IM congénitaux

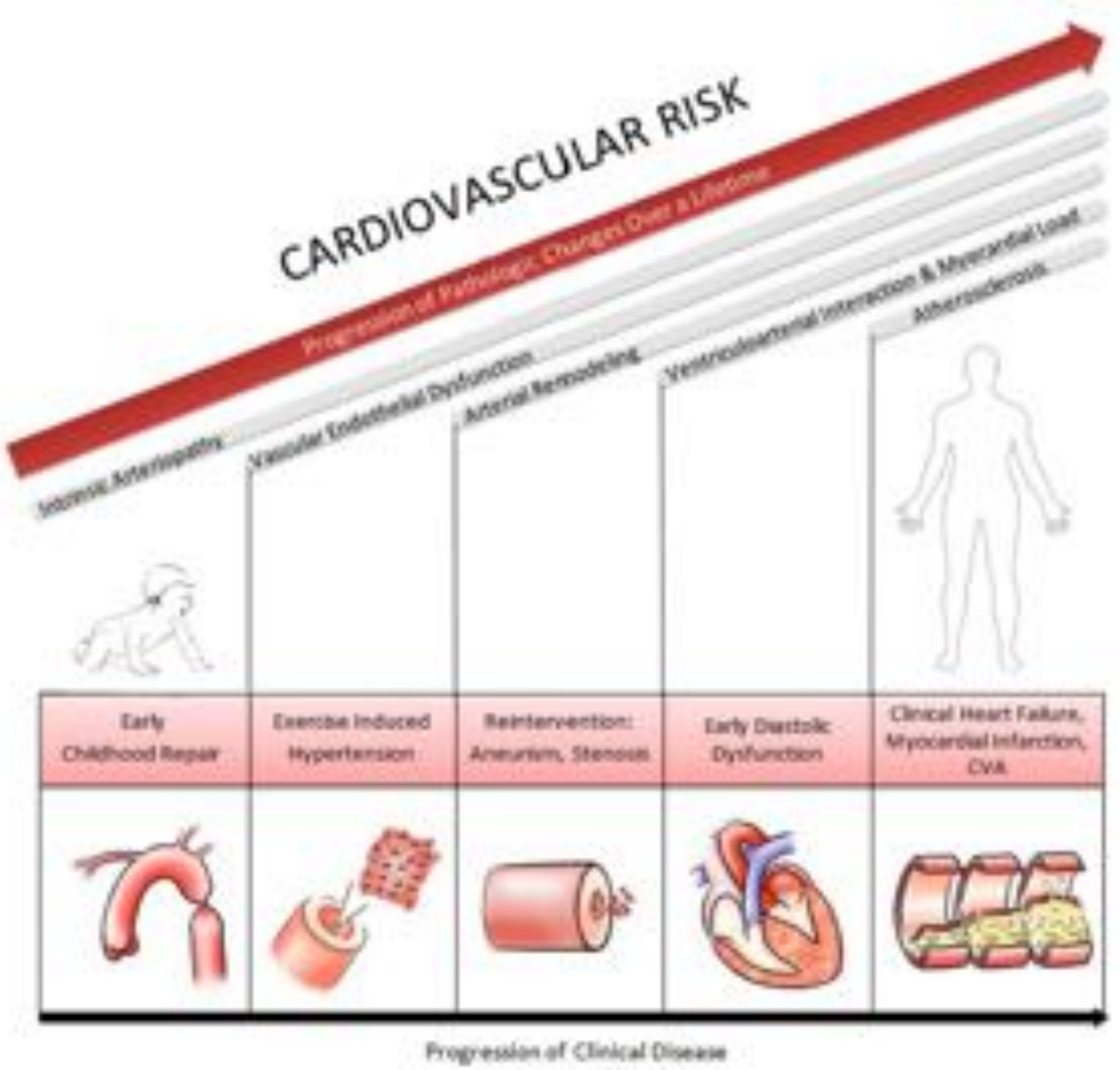
Au-delà de la période néonatale

- Indications opératoires idem
- Méthodes
  - Chirurgie chez l'enfant ?
  - Stenting direct chez l'adulte ?



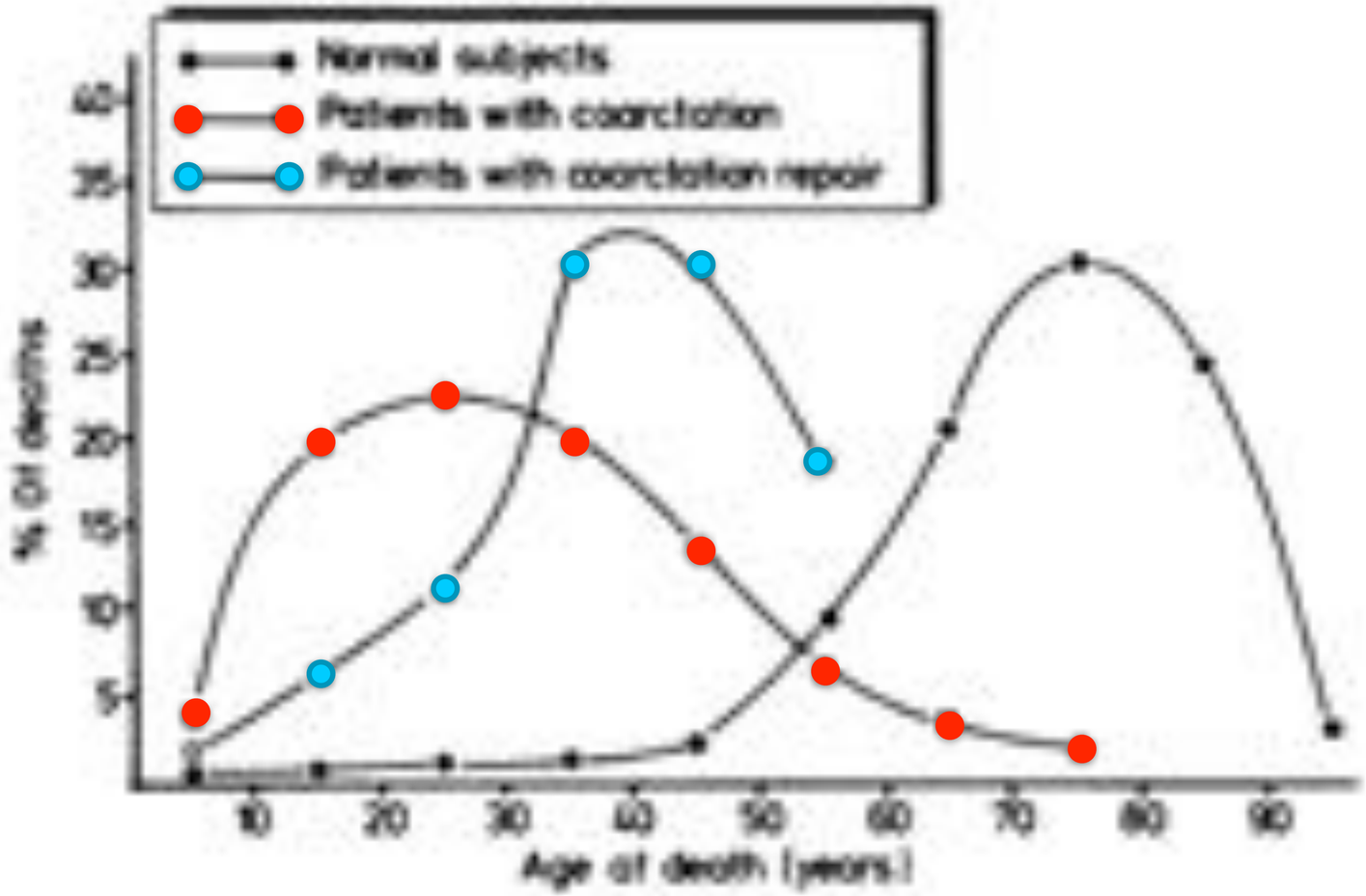
Complications immédiates

- Poussée hypertensives post-opératoires
- Infection de l’anastomose



Bhatt AB et al. Heart 2015;101:1173-5

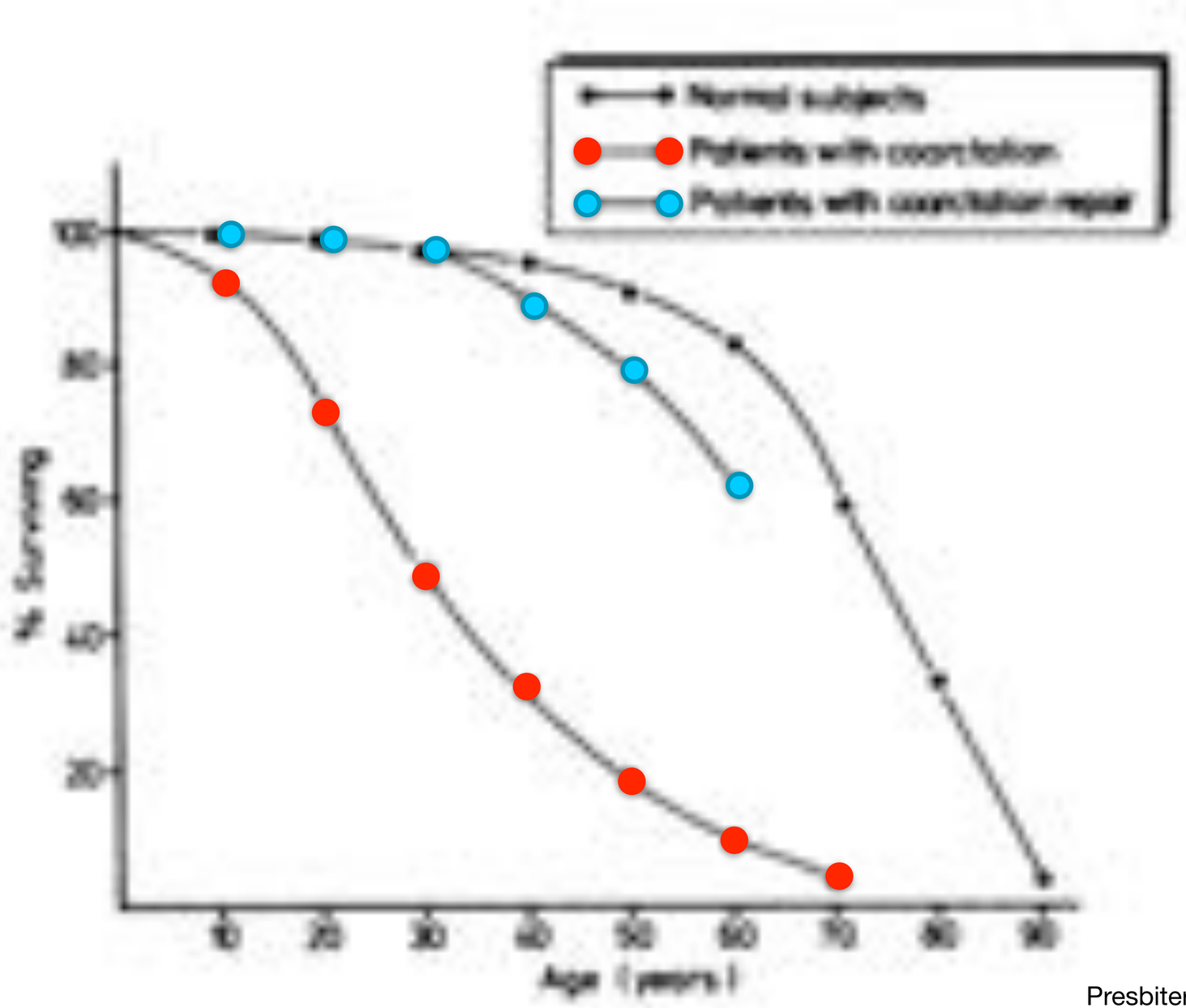
Distribution of death by age of subjects with **coarctation**, of subjects with **coarctation repair**, and of the **general population**



The curves on the left and right are taken from Campbell.
The middle curve shows the distribution of death in our patients with coarctation repair. These curves have been adjusted for age.

Presbitero P et al. Br Heart 1987;57:462-7

Percentage of subjects still alive at the end of each decade



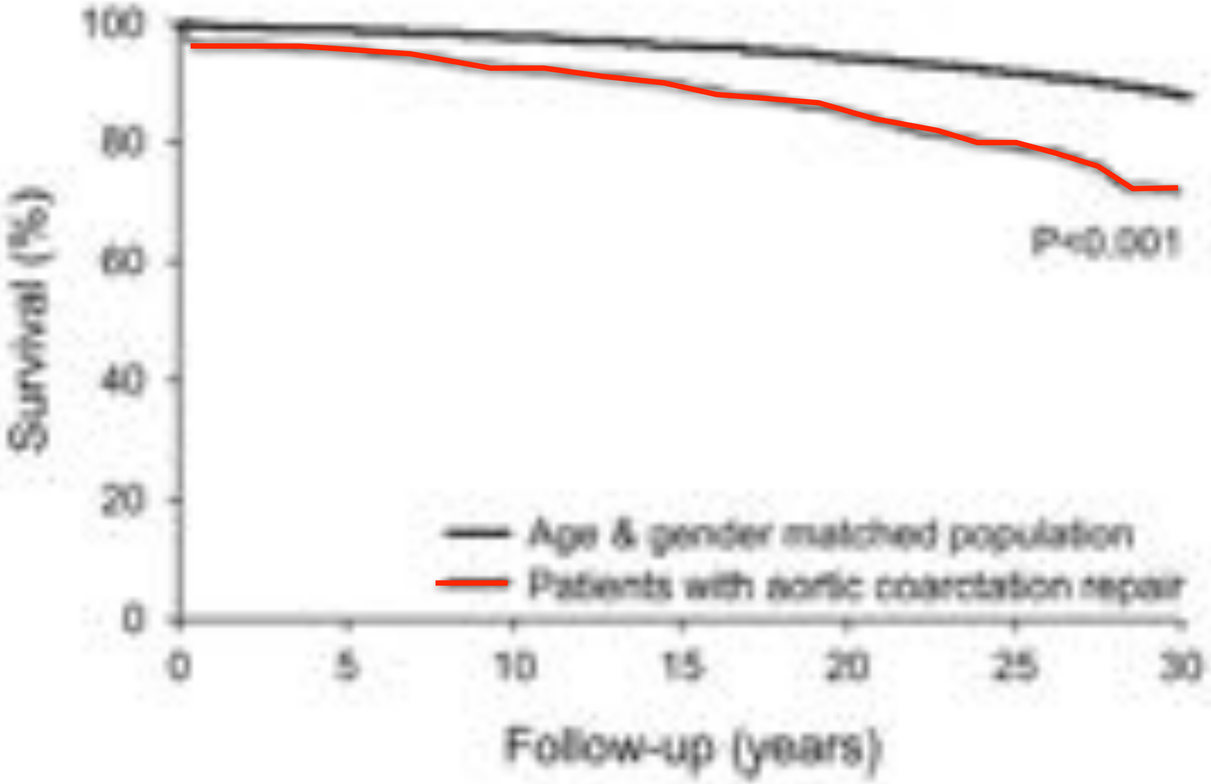
Presbitero P et al. Br Heart 1987;57:462-7

Overall survival after coarctation repair

Mean age at surgery 17.2 years

Table 1 Pre-Operative Characteristics (N = 853)

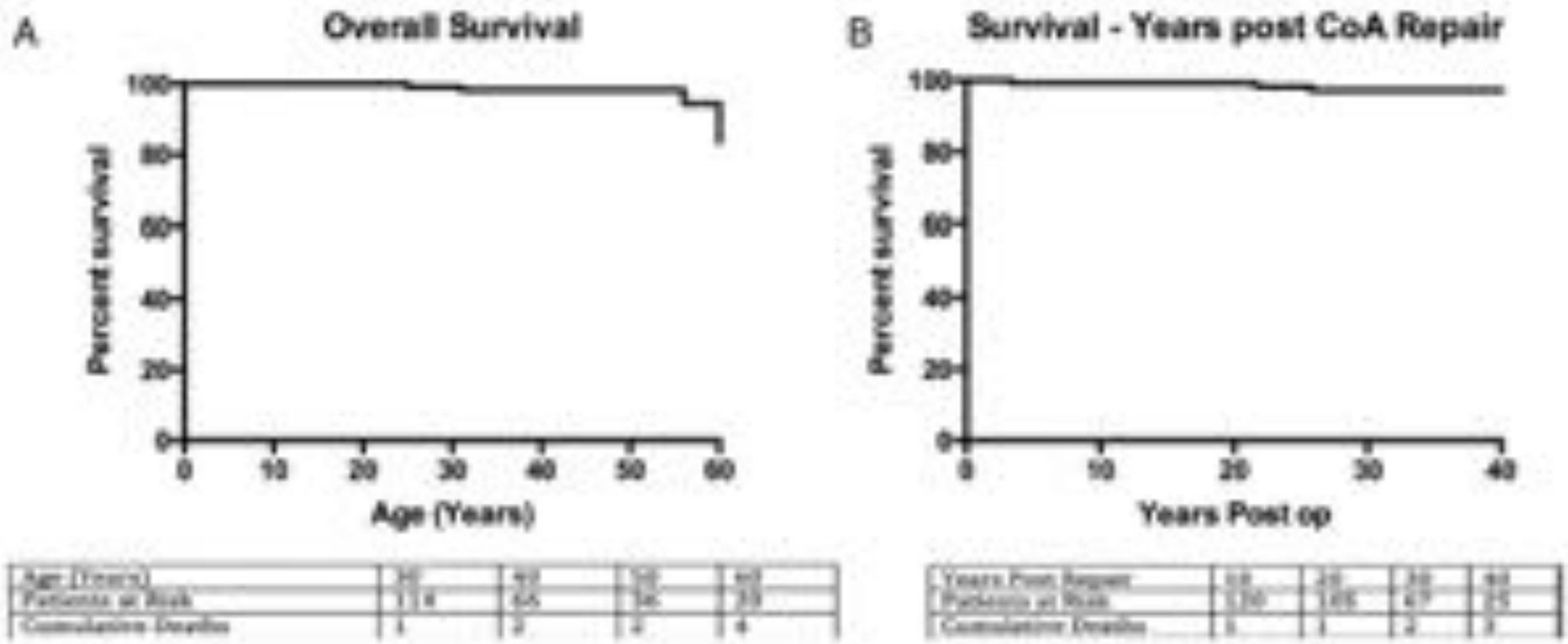
Characteristic	Value
Age at operation	
Mean ± SD, yrs	17.2 ± 13.6
Range	1 day to 72.2 yrs
Age group	
≤1 yr	136
>1–≤5 yrs	76
>5–≤10 yrs	123
>10–≤20 yrs	235
>20 yrs	269
Female	343 (30)
Pre-operative hypertension	663 (83)
NYHA class III or IV	32 (6)



Brown ML et al. JACC 2013;62:1020-5

Kaplan–Meier curve showing long-term survival in patients with coarctation of the aorta (CoA) repair

Median age at surgery 60 months



Choudhary P et al. Heart 2015;101:1190-95

Residua, sequelae, and complications are listed below
ESC Guidelines 2010

- Arterial hypertension *at rest or during exercise* is common, even after successful treatment, and it is an important risk factor for premature CAD, ventricular dysfunction, and rupture of aortic or cerebral aneurysms. ***The geometry of the arch*** may play a role in the development of hypertension. ***The significance of isolated, exercise-induced hypertension*** is a matter of debate.

- Recurring or residual CoA may induce or aggravate systemic arterial hypertension and its consequences

- Aneurysms of the ascending aorta or at the intervention site present a risk of rupture and death. Patch repair are at particular risk of repair site aneurysms and should be imaged on a regular basis.

- Attention is required for BAV, mitral valve disease, premature CAD, and berry aneurysm of the circle of Willis.

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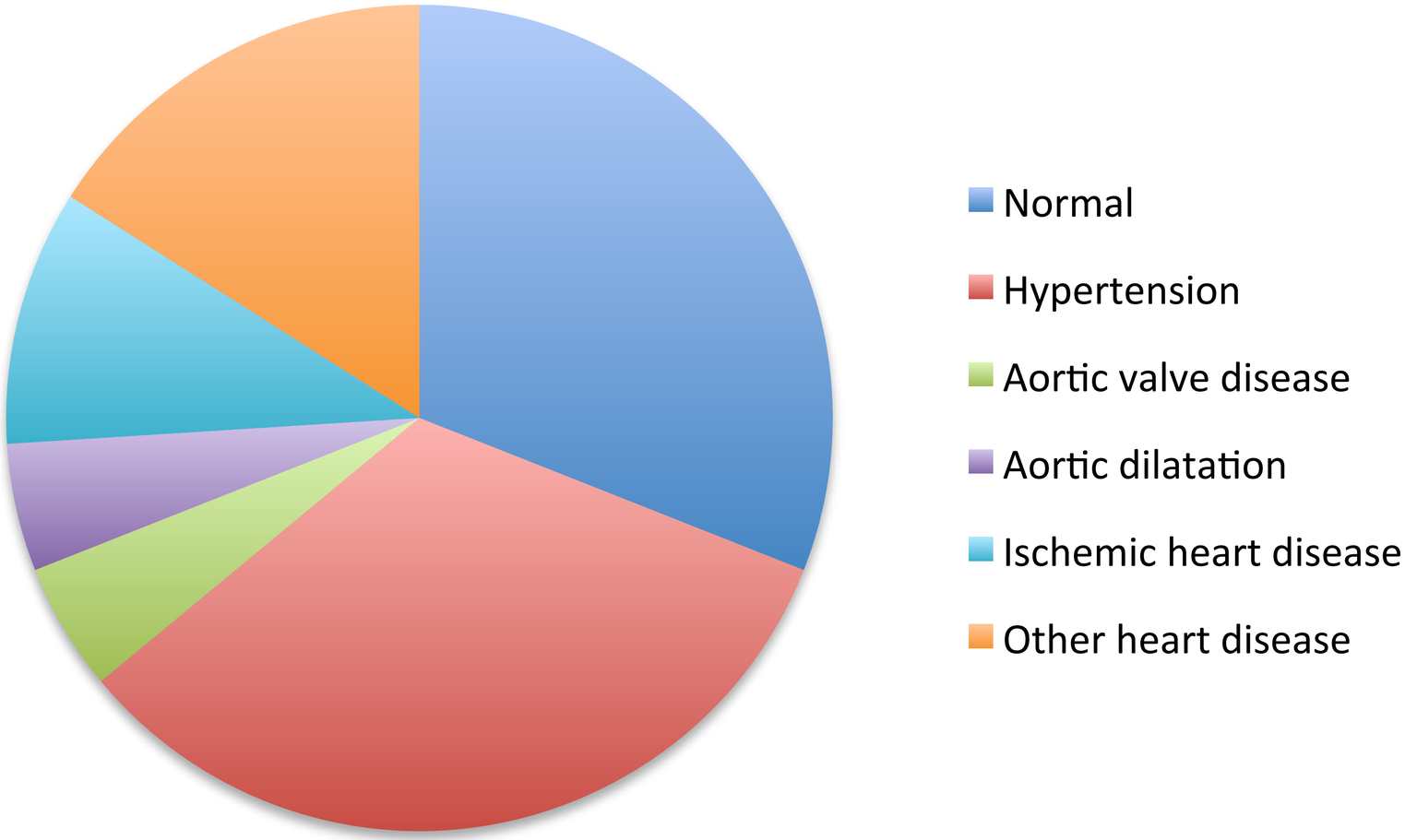
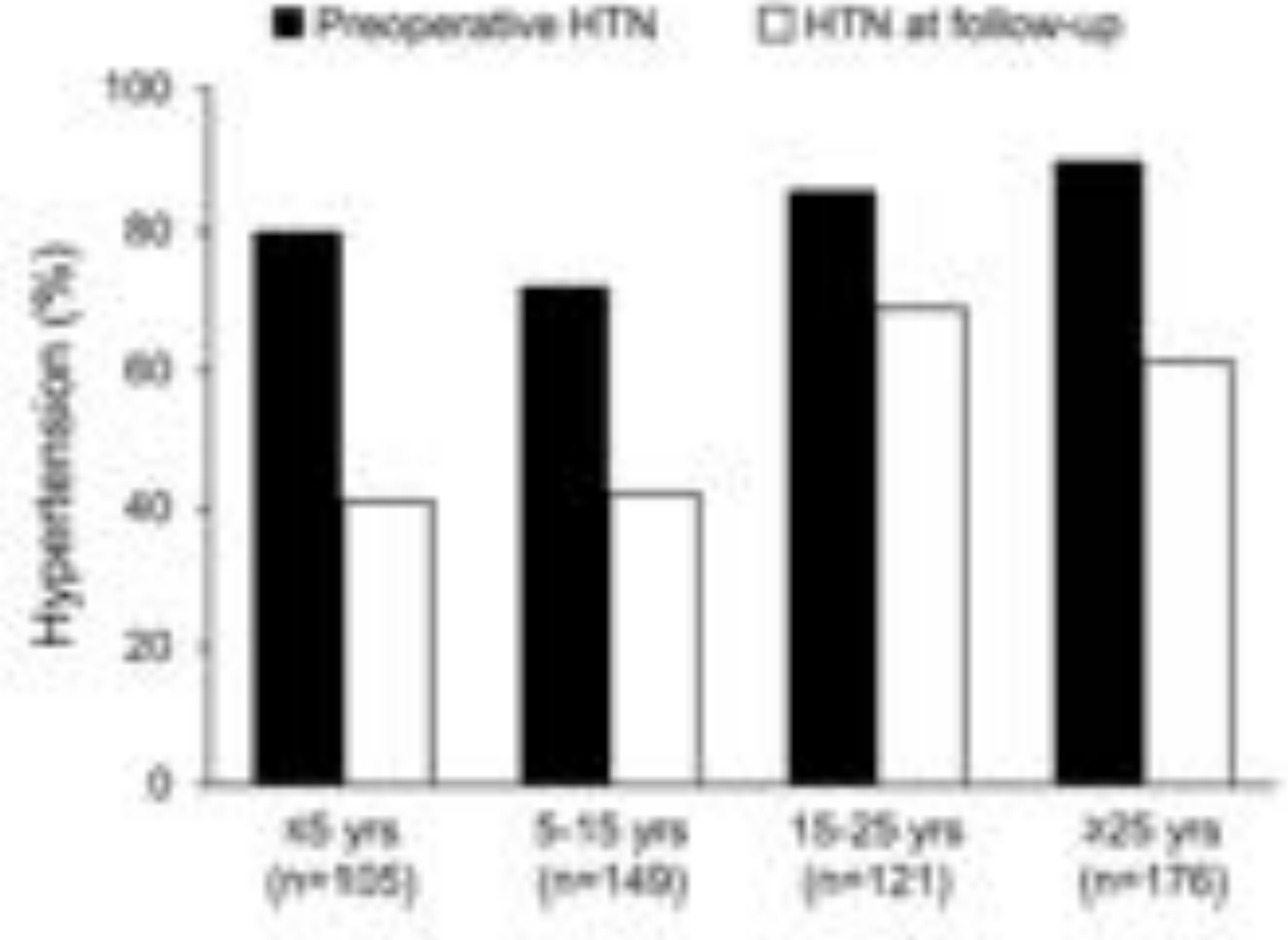
Cardiovascular morbidity in adults after coarctation repair



Stewart AB et al. Br Heart 1993;69:65-70

Comparison of hypertension pre- and postoperatively at various time intervals



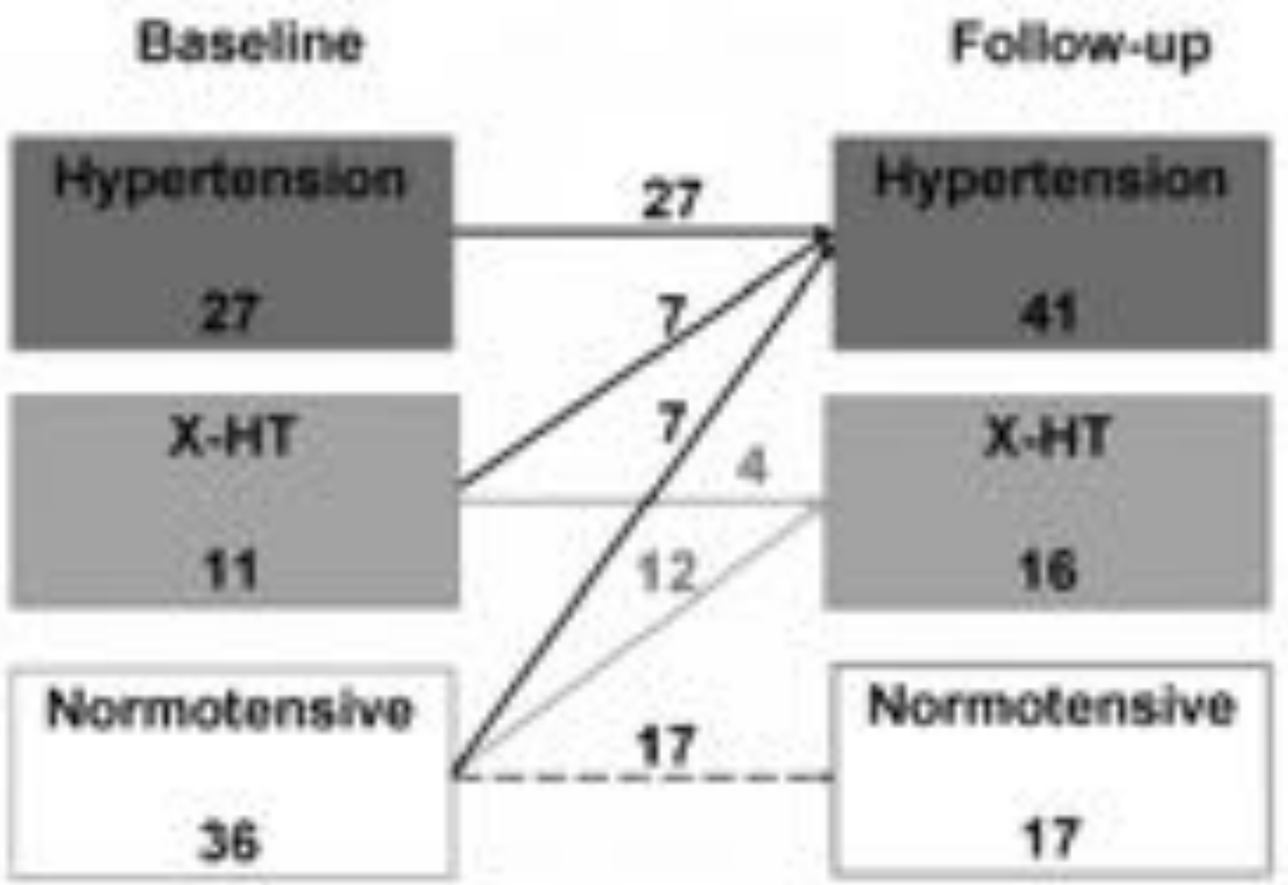
Brown ML et al. JACC 2013;62:1020-5

What type of HT ?

- Exercise HT
- 24h Ambulatory Blood Pressure Monitoring
  - 60% abnormal pressure profile
  - 24% night HT in daytime normotensive patients
- Systolic HT with increased pulse pressure
- Resting HT

Lee MGY et al. J Thorac Cardiovasc Surg 2012;144:1110-8

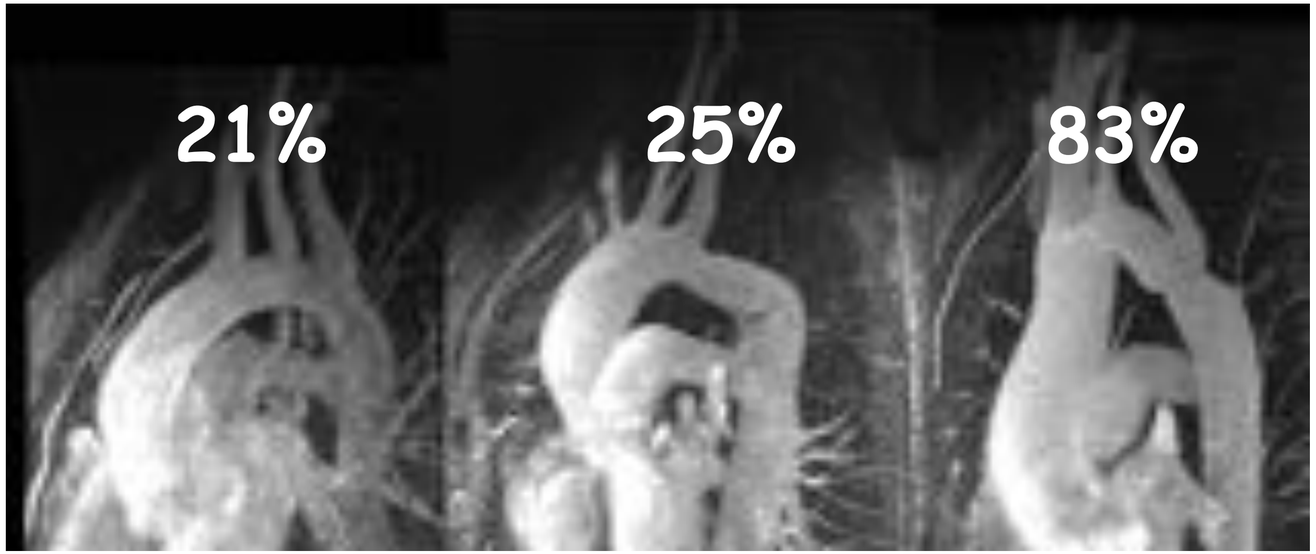
Exercise hypertension predicts late resting hypertension after coarctation repair



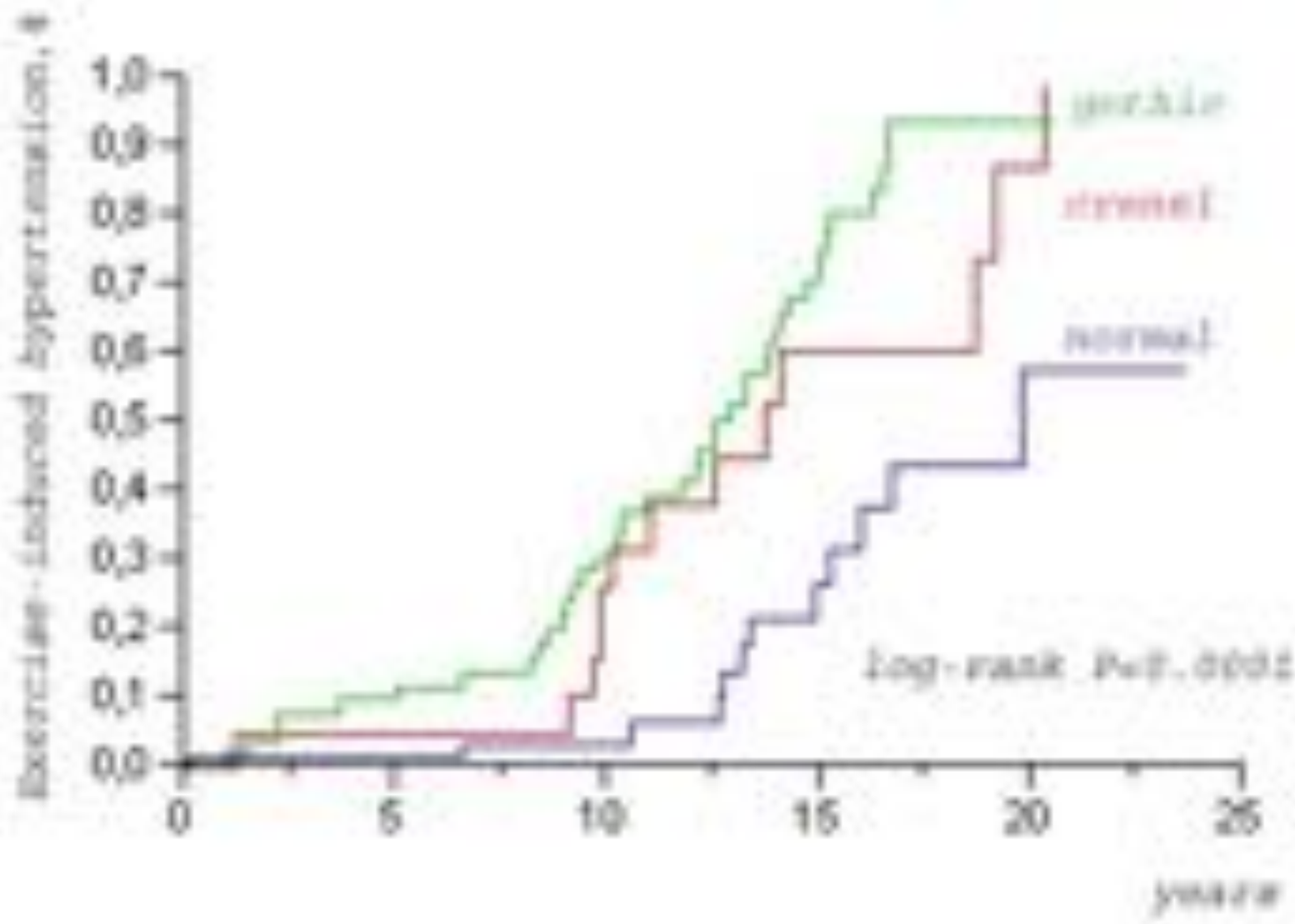
Baseline maximum exercise systolic BP was independently associated with the mean systolic BP at follow-up (p <0.005)

Luijendijk P et al. Am J Cardiol 2011;108:435-9

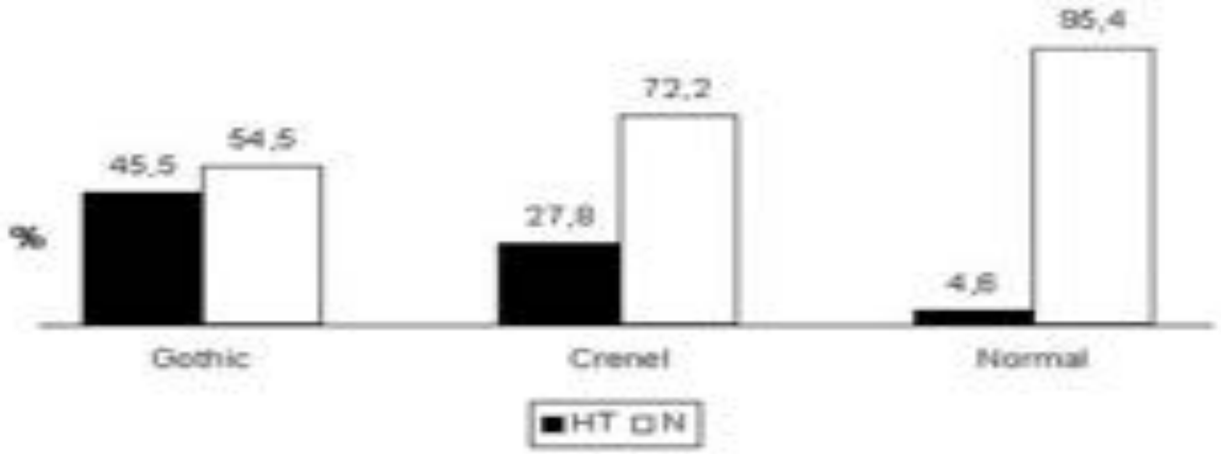
Correlation between aortic arch geometry and BP at exercise



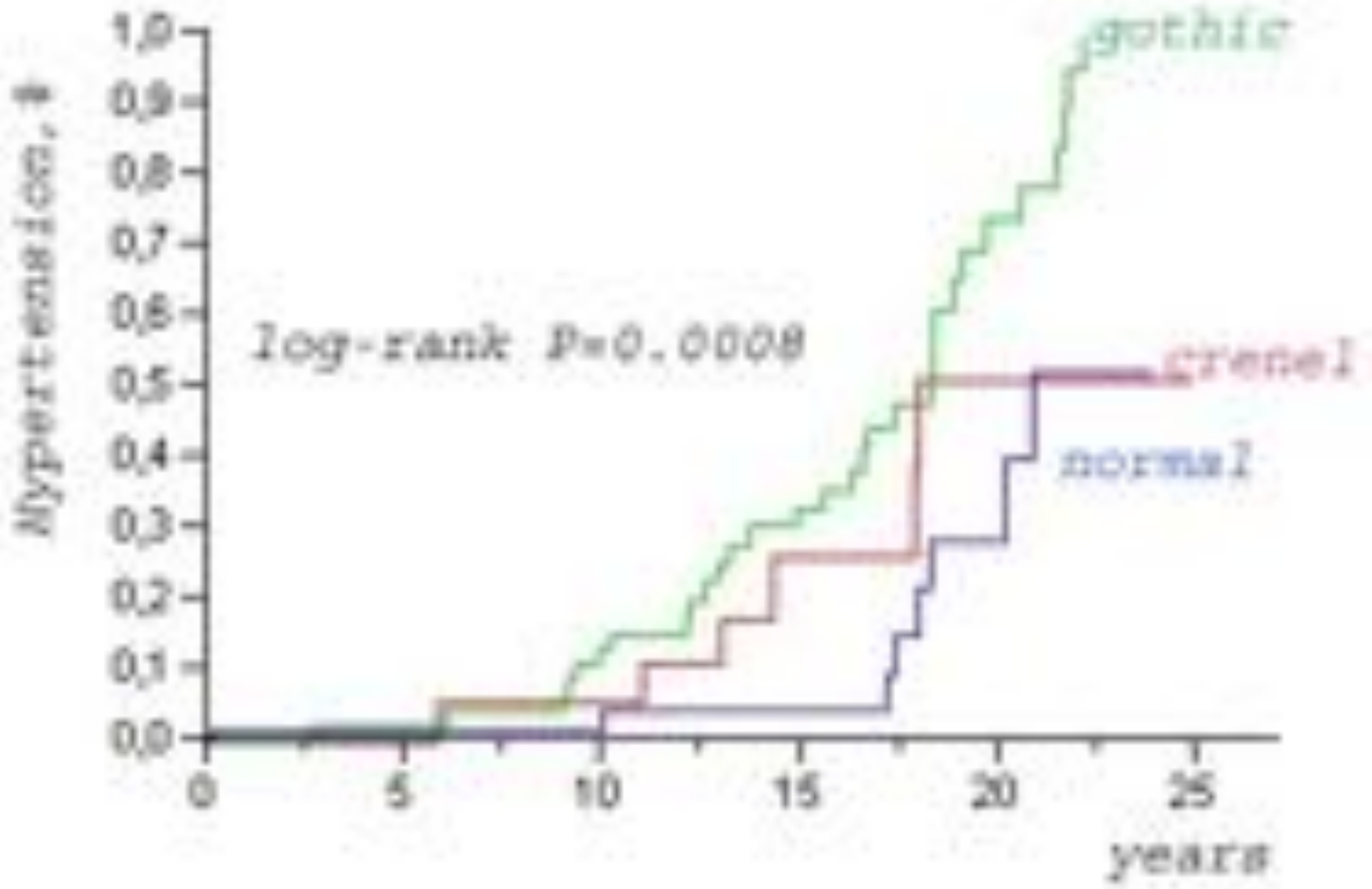
Exercise induced HT was correlated to the gothic arch form and to A/T


Exercise-induced hypertension, %
1,0
0,9
0,8
0,7
0,6
0,5
0,4
0,3
0,2
0,1
0,0
0
5
10
15
20
25
years
severe
normal
Log-rank P<0.0001

Aortic arch geometry and resting HT after CoA repair



Ou P. et al, Eur. Heart J. 2004


Hypertension, %
1,0
0,9
0,8
0,7
0,6
0,5
0,4
0,3
0,2
0,1
0,0
log-rank P=0,0008
gothic
trend
normal
0
5
10
15
20
25
years

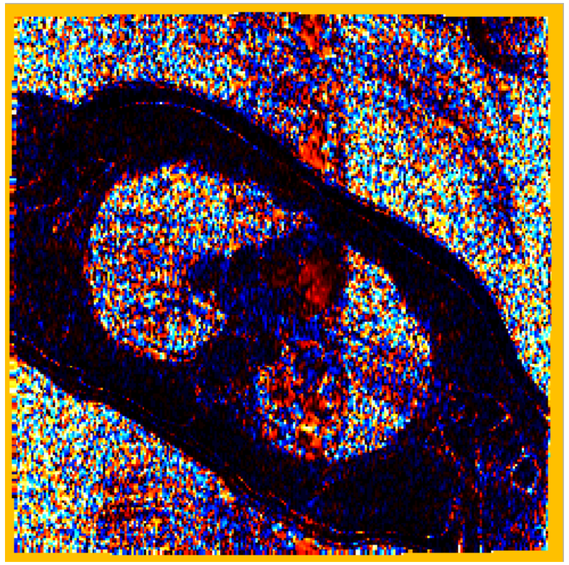
Aortic arch geometry and flow dynamics after CoA repair

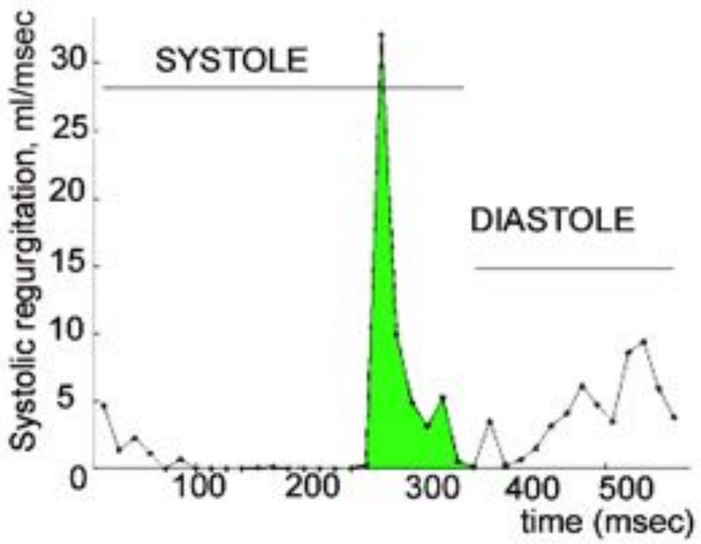
	GOTHIC	CRENEL	NORMAL	*P value*
PWV, (m/sec)	8.6 ±3.4	4.5 ±1.6	3.7 ±0.5	*<0.0001*
Systolic regurgitation (ml/msec)	14.4 ±5.3	7.9 ±1.3	5.3 ±1.9	*<0.001*

Aortic arch geometry and flow dynamics after CoA repair

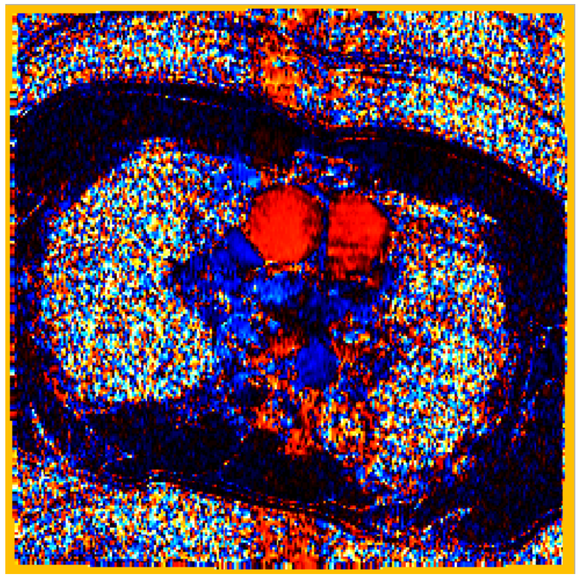
Flow dynamics in the ascending aorta

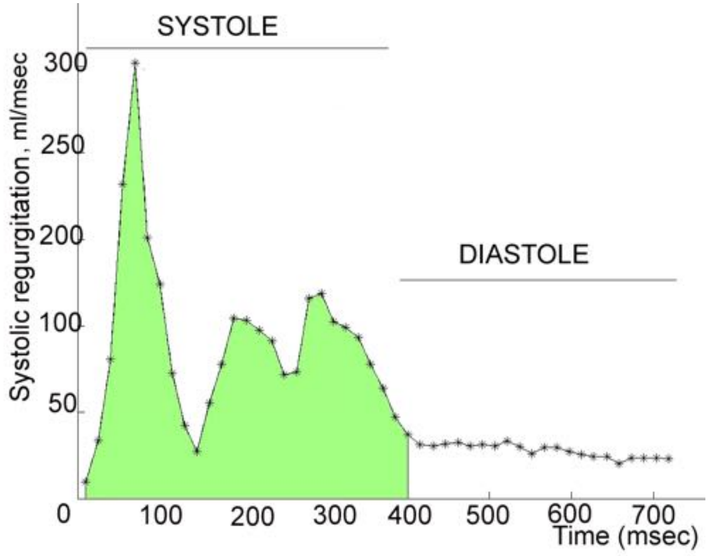
NORMAL

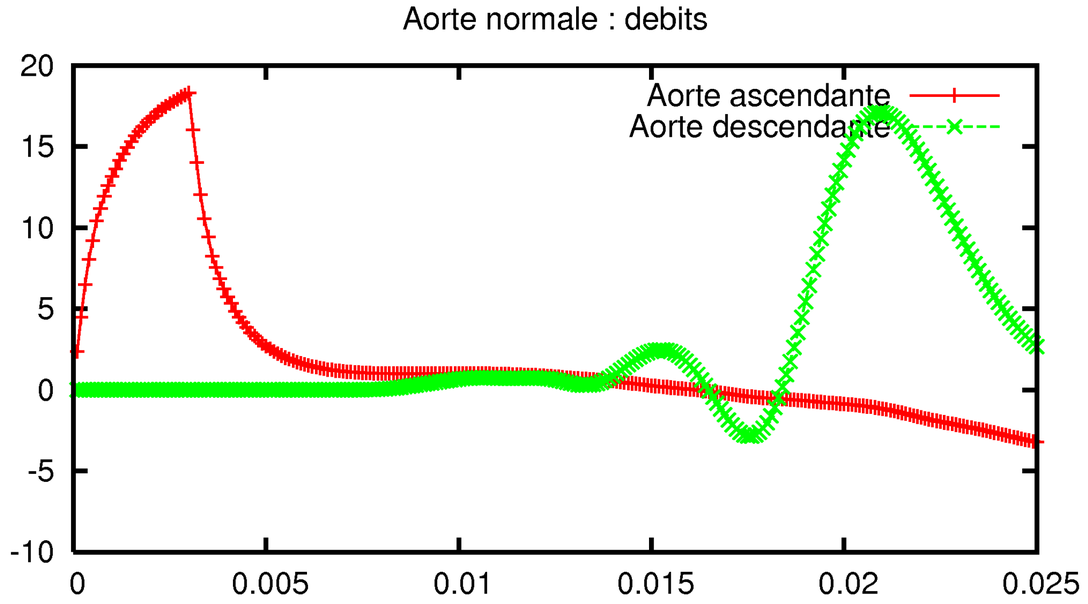


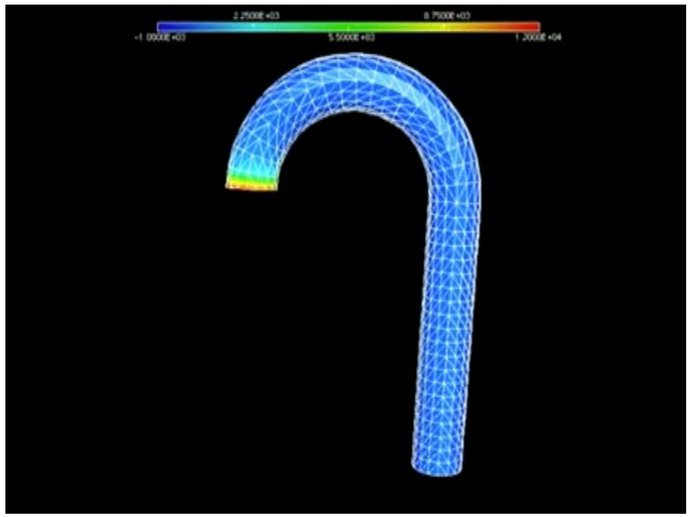


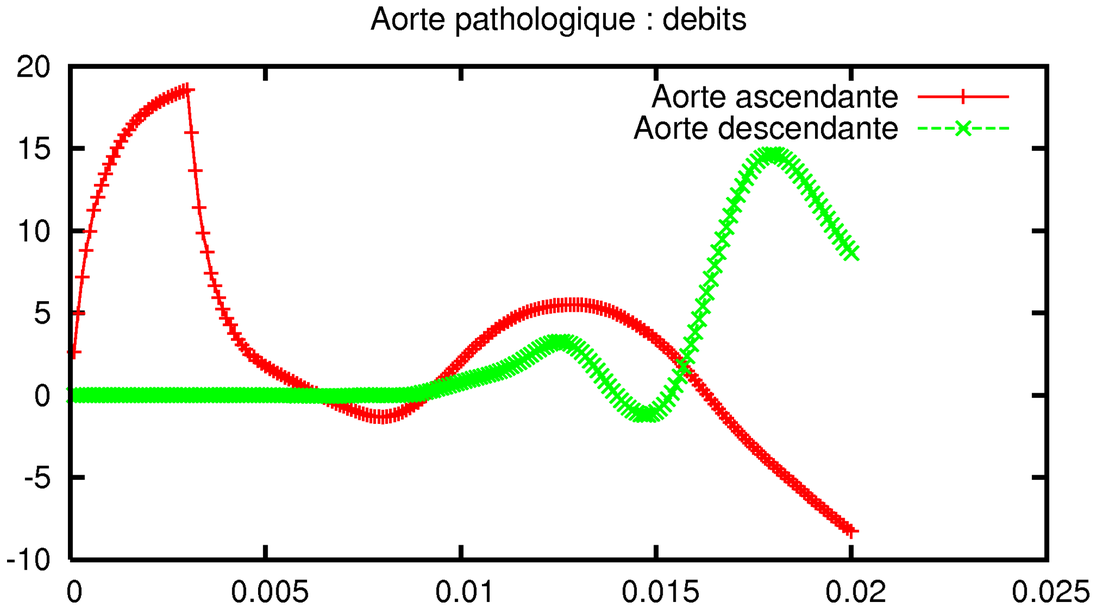
GOTHIC

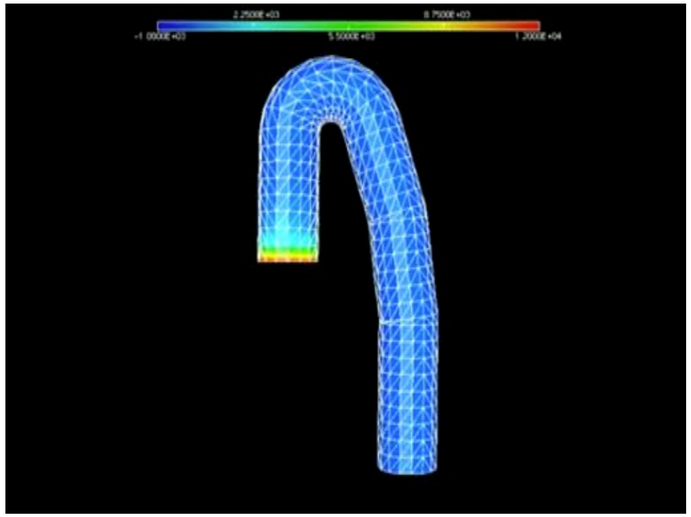




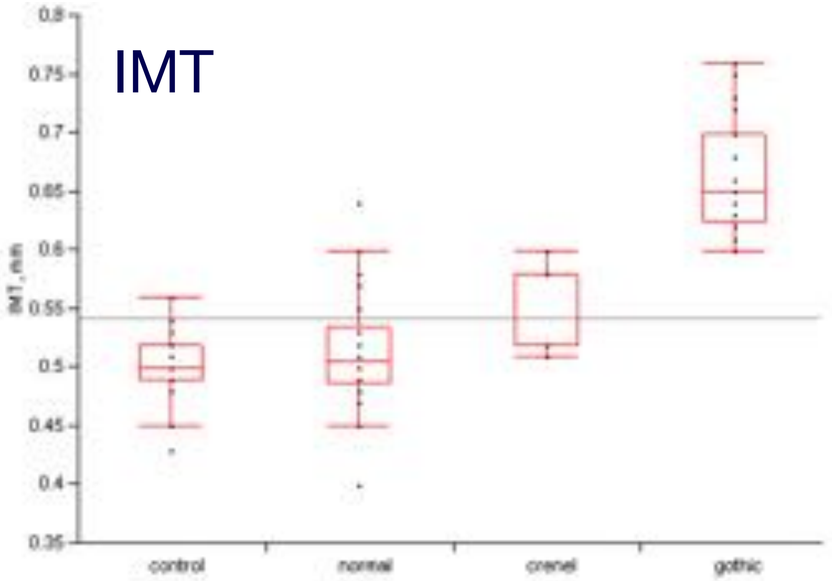


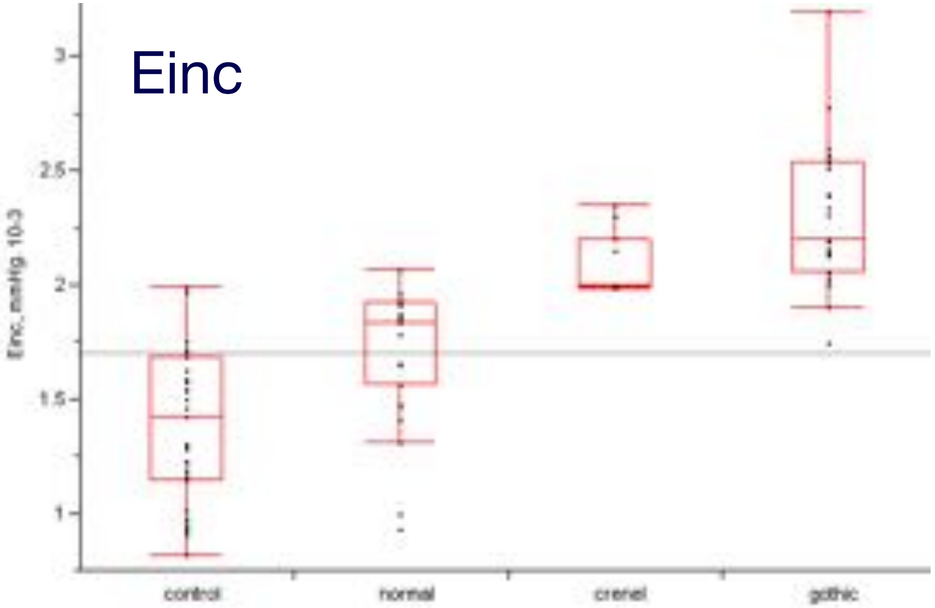


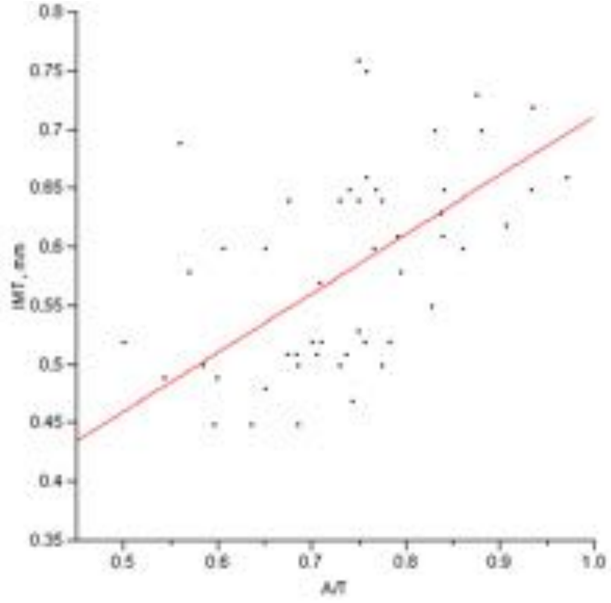


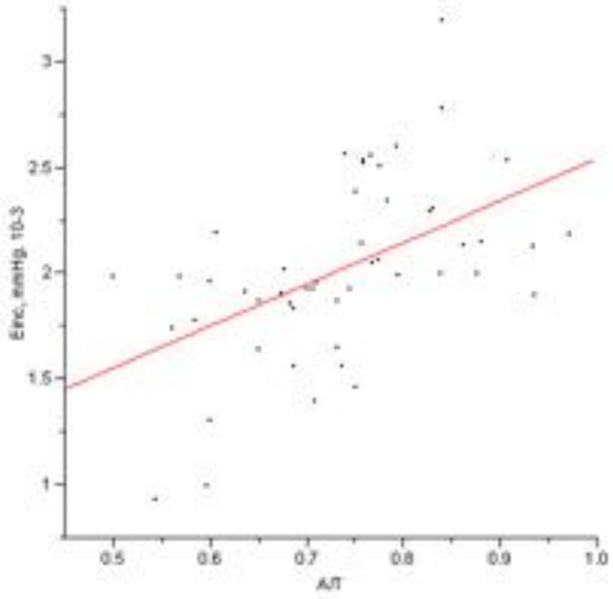


Szopos M et al. J Thorac Cardiovasc Surg. 2014;148:1572-82.









Ou P et al. J Am Coll Cardiol. 2007;49:883-90.
Ou P et al. J Thorac Cardiovasc Surg. 2008;135:62-8.

The significance of isolated, exercise-induced hypertension is a matter of debate.

- Exercise HT does not help to detect recoarctation.
- Exercise HT predicts resting HT.
- Exercise HT is associated with vascular remodeling and increased LV mass.
- Should exercise HT lead to early antihypertensive treatment ?

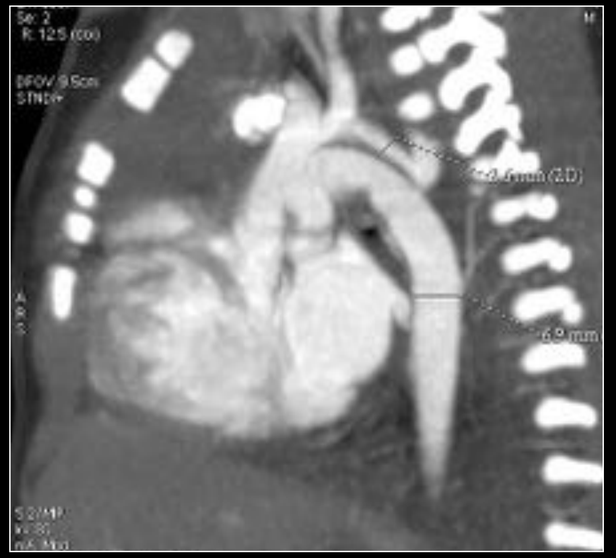
Residua, sequelae, and complications are listed below
ESC Guidelines 2010

- Arterial hypertension *at rest or during exercise* is common, even after successful treatment, and it is an important risk factor for premature CAD, ventricular dysfunction, and rupture of aortic or cerebral aneurysms*. The geometry of the arch* may play a roel in the development of hypertension. *The significance of isolated, exercise-induced hypertension* is a matter of debate.

- Recurring or residual CoA may induce or aggravate systemic arterial hypertension and its consequences

- Aneurysms of the ascending aorta or at the intervention site present a risk of rupture and death. Patch repair are at particular risk of repair site aneurysms and should be imaged on a regular basis.

- Attention is required for BAV, mitral valve disease, premature CAD, and berry aneurysm of the circle of Willis.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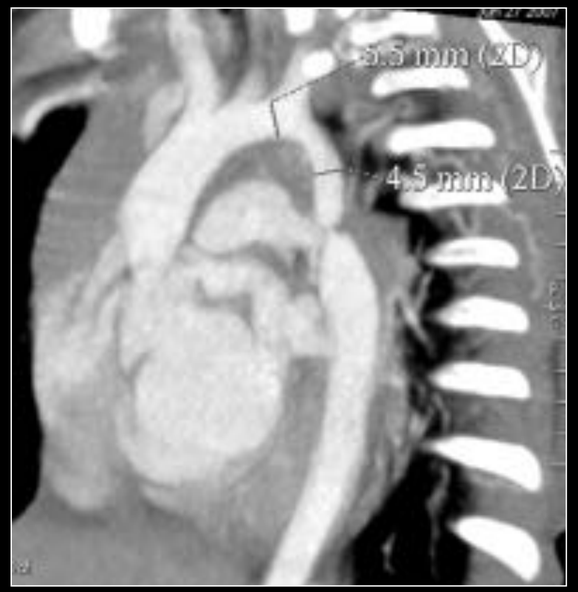
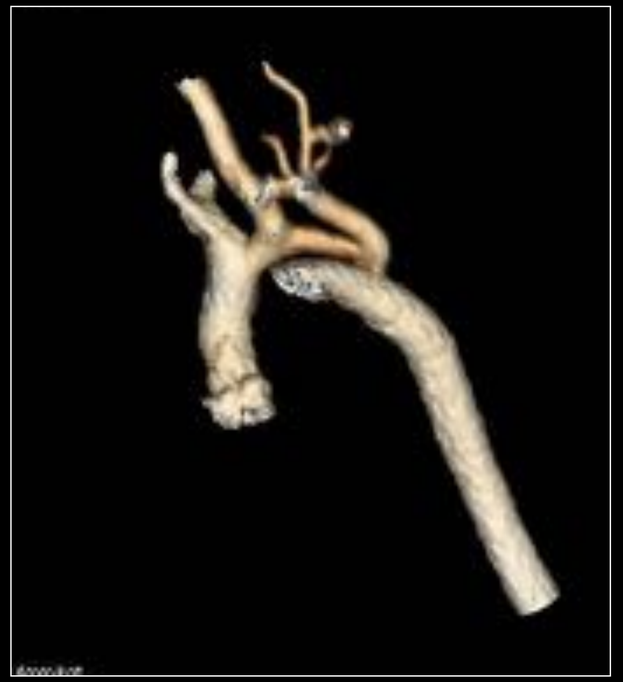
6.9 mm


4.0 mm (2D)


5.5 mm (2D)
4.5 mm (2D)




IPR
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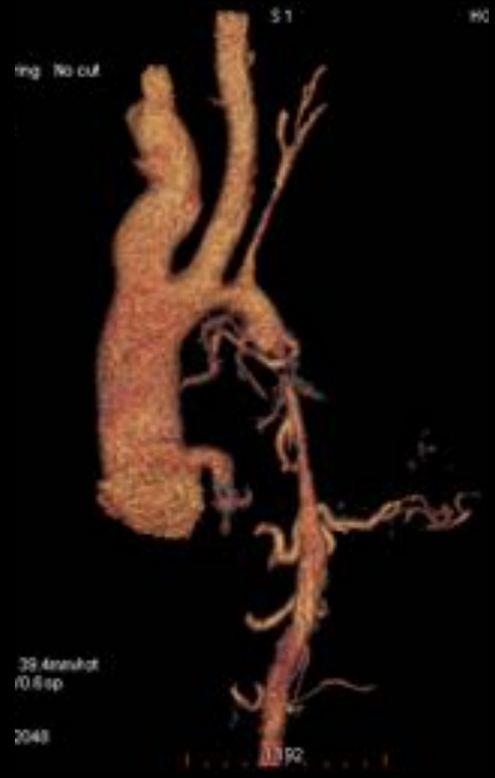


3D
Ex: 15217
Se: 2
Volume Rendering No cut


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Table 11 Indications for intervention in coarctation of the aorta

Indications	Class[a]	Level[b]
All patients with a non-invasive pressure difference >20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mmHg in adults), pathological blood pressure response during exercise, or significant LVH should have intervention	I	C
Independent of the pressure gradient, hypertensive patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) should be considered for intervention	IIa	C
Independent of the pressure gradient and presence of hypertension, patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) may be considered for intervention	IIb	C

[a]Class of recommendation.
[b]Level of evidence.
CMR = cardiac magnetic resonance; CoA = coarctation of the aorta; CT = computed tomography; LVH = left ventricular hypertrophy.

When to treat ?
How to treat residual or re-coarctation ?

Residua, sequelae, and complications are listed below
ESC Guidelines 2010

- Arterial hypertension *at rest or during exercise* is common, even after successful treatment, and it is an important risk factor for premature CAD, ventricular dysfunction, and rupture of aortic or cerebral aneurysms*. The geometry of the arch* may play a roel in the development of hypertension. *The significance of isolated, exercise-induced hypertension* is a matter of debate.

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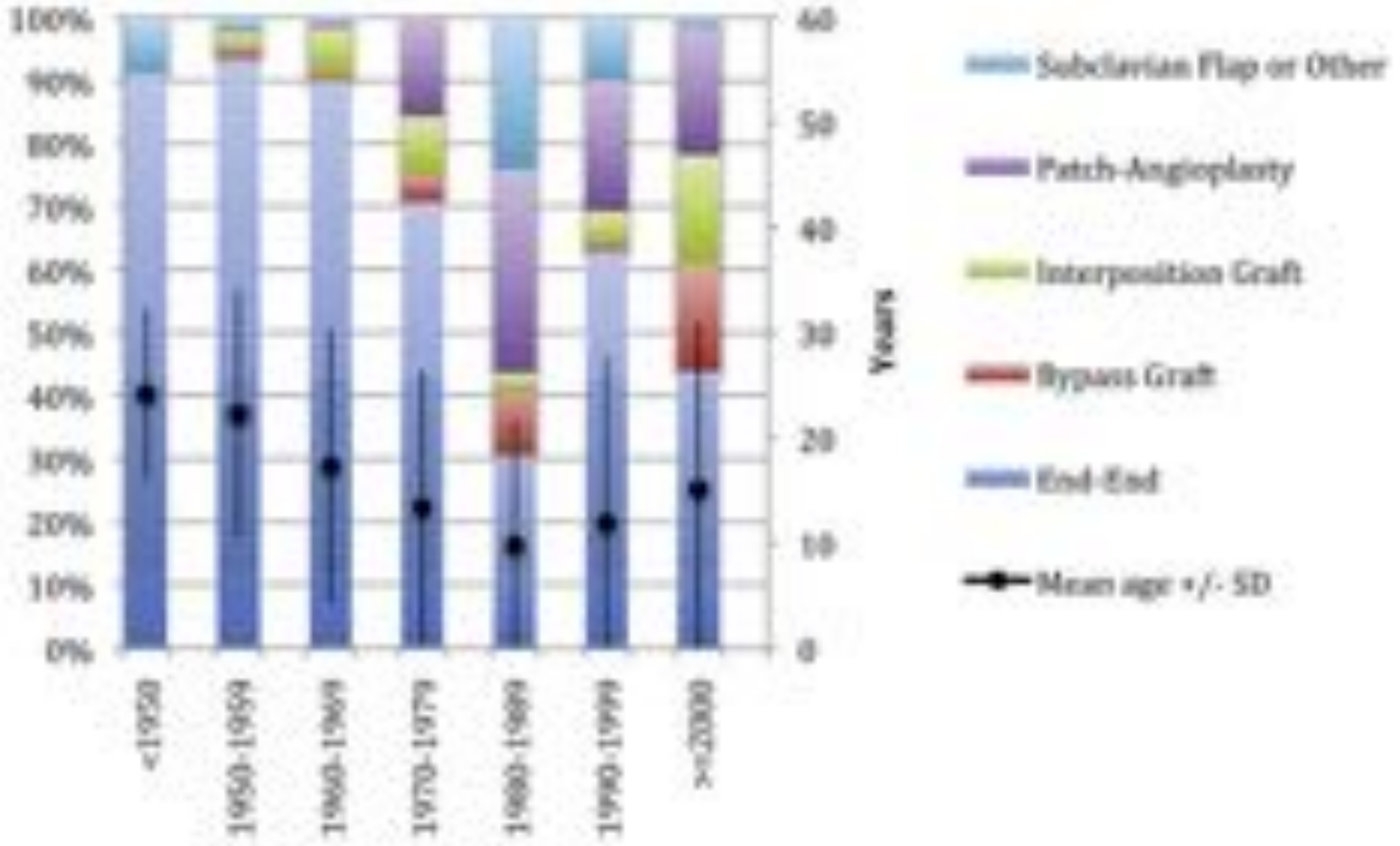
- Aneurysms of the ascending aorta or at the intervention site present a risk of rupture and death. Patch repair are at particular risk of repair site aneurysms and should be imaged on a regular basis.

- Attention is required for BAV, mitral valve disease, premature CAD, and berry aneurysm of the circle of Willis.

Type of repair by decade



Brown ML et al. JACC 2013;62:1020-5

Aortic coarctation endocarditis

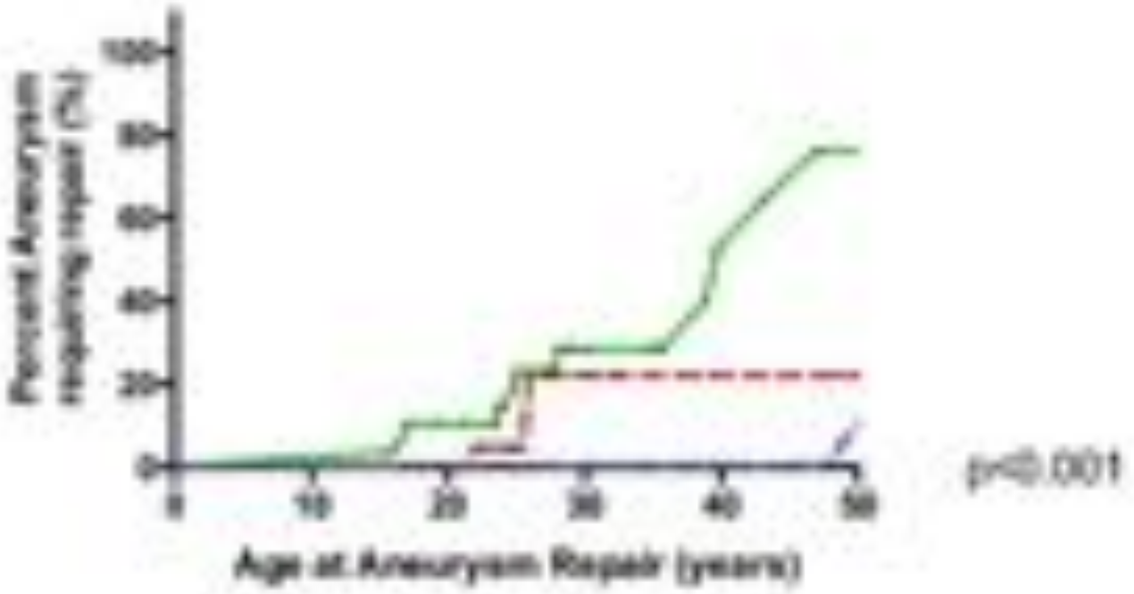
Aneurysm at the repair site

Location	Number (%)
Site of prior repair or DTA	42 (76.4%)
Ascending aorta	18 (32.7%)
Left subclavian artery	8 (14.5%)
Distal aortic arch	8 (14.5%)
Aortic root	6 (10.9%)
Abdominal aorta	1 (1.8%)
Iliac artery	1 (1.8%)
Innominate artery	1 (1.8%)

DTA = descending thoracic aorta.

Preventza O et al. Ann Thorac Surg 2013;95:1960-7

Incidence of descending aortic aneurysms requiring repair



Patients at risk	Total	Legend	20 years	30 years	40 years	50 years
Mid-to-end repair	43		39	23	13	7
Subclavian flap augmentation	28		23	7	3	2
Patch Aortoplasty	22		23	9	3	3

Choudhary P et al. Heart 2015;101:1190-95

Aneurysm of the ascending aorta

32% of patients with aortic root dilatation
21% with aneurysm of ascending aorta
74% had Bicuspid aortic valve

Padang R et al. Heart, Lung and Circulation 2014;23: 332–338

Residua, sequelae, and complications are listed below
ESC Guidelines 2010

- Arterial hypertension *at rest or during exercise* is common, even after successful treatment, and it is an important risk factor for premature CAD, ventricular dysfunction, and rupture of aortic or cerebral aneurysms*. The geometry of the arch* may play a roel in the development of hypertension. *The significance of isolated, exercise-induced hypertension* is a matter of debate.

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Common cardiovascular anomalies associated with coarctation

Anomaly	No. of patients	% (n = 500 unless otherwise specified)
Bicuspid aortic valve (including functional bicuspid valve)	268	59.6 (n = 449)
Arch hypoplasia	71	14.2
Ventricular septal defect	64	12.8
Mitral valve abnormalities	41	8.2
Mitral regurgitation	20	
Mitral valve prolapsed	6	
Mitral stenosis	4	
Double-orifice mitral valve	3	
Parachute mitral valve	2	
Imperforate mitral valve (associated with Shone complex)	2	
Patent ductus arteriosus	36	7.2
Subaortic stenosis	28	5.6
Other arch anomalies	21	4.2
Aberrant right subclavian artery	8	
Aberrant left subclavian artery	1	
Bovine pattern	7	
Other arch anomalies	5	
Left superior vena cava	21	4.2
Atrial septal defect	10	2

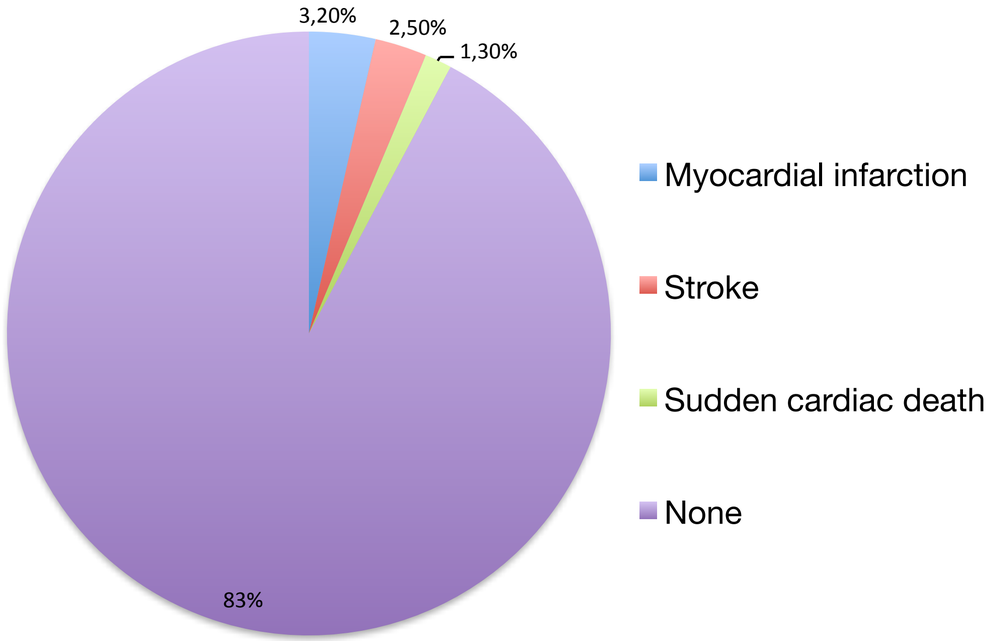
High proportion of Sievers type 0 BAV

Teo LLS et al. Pediatr Cardiol 2011;32:1120-7

Risk of premature CAD after coarctation repair

CoA did not independently predict for the development of CAD (OR, 1.04; 95% CI, 0.68 –1.57) or premature CAD (OR for CoA versus ventricular septal defect, 1.44; 95% CI, 0.79 –2.64) after adjustment for other cardiovascular risk factors.

Incidence of CV events in 160 patients after coarctation repair over a period of 10 years



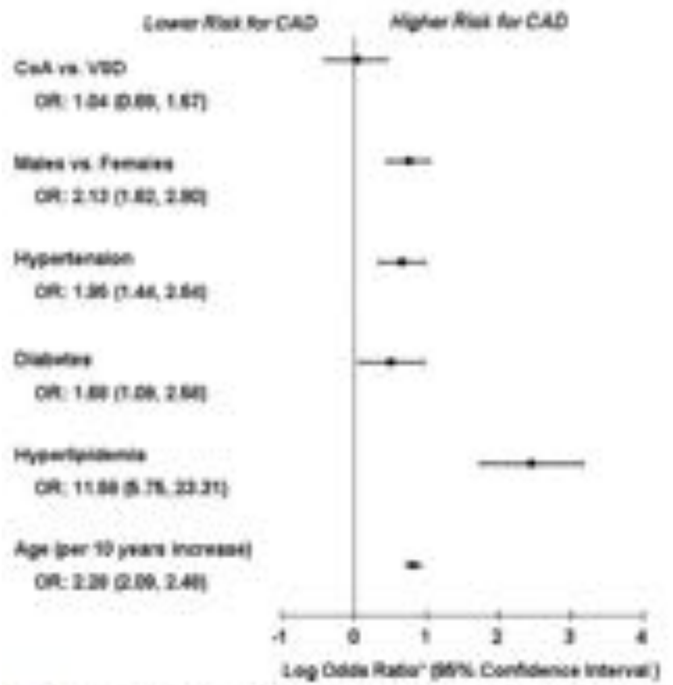


Figure 2. Multivariable analysis of the effect of aortic coarctation (CoA) vs ventricular septal defect (VSD) on coronary artery disease (CAD; nested case-control sample). OR indicates odds ratio.

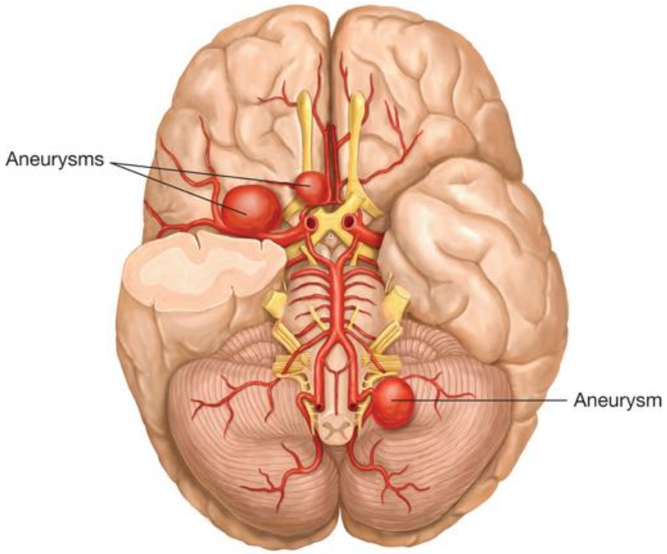
Luijendijk P et al. Int J Cardiol 2014;176:776-81
Roifman I et al. Circulation 2012;126:16-21

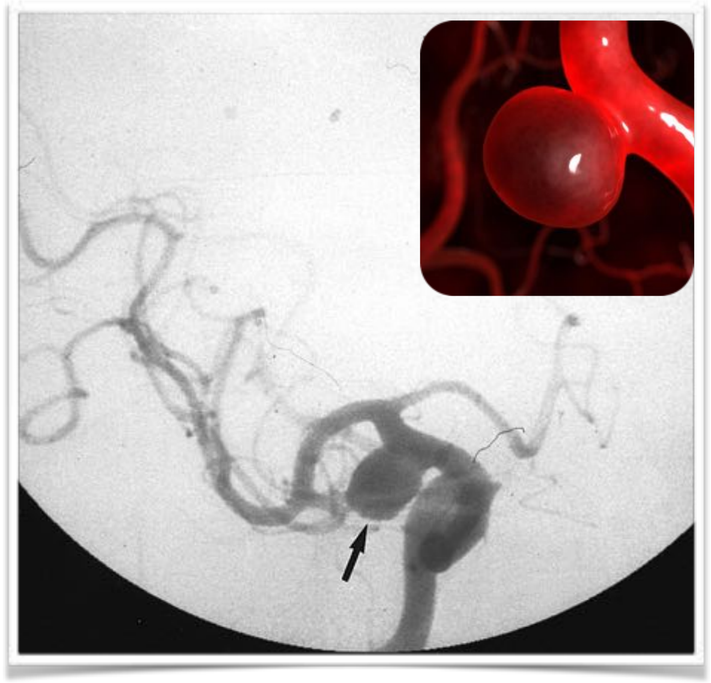
Berry aneurysms of the circle of Willis and coarctation

Controversial prevalence in patients with Coarctation

5-18% in a MRI study in adults (mean age 42 years)[1]

0% in a MRI study in adolescents (mean age 16 years) [2]





1-Connolly HM, et al. Mayo Clin Proc 2003;78:1491-9.
2-Donti A et al. Am J Cardiol 2015;116:630-3

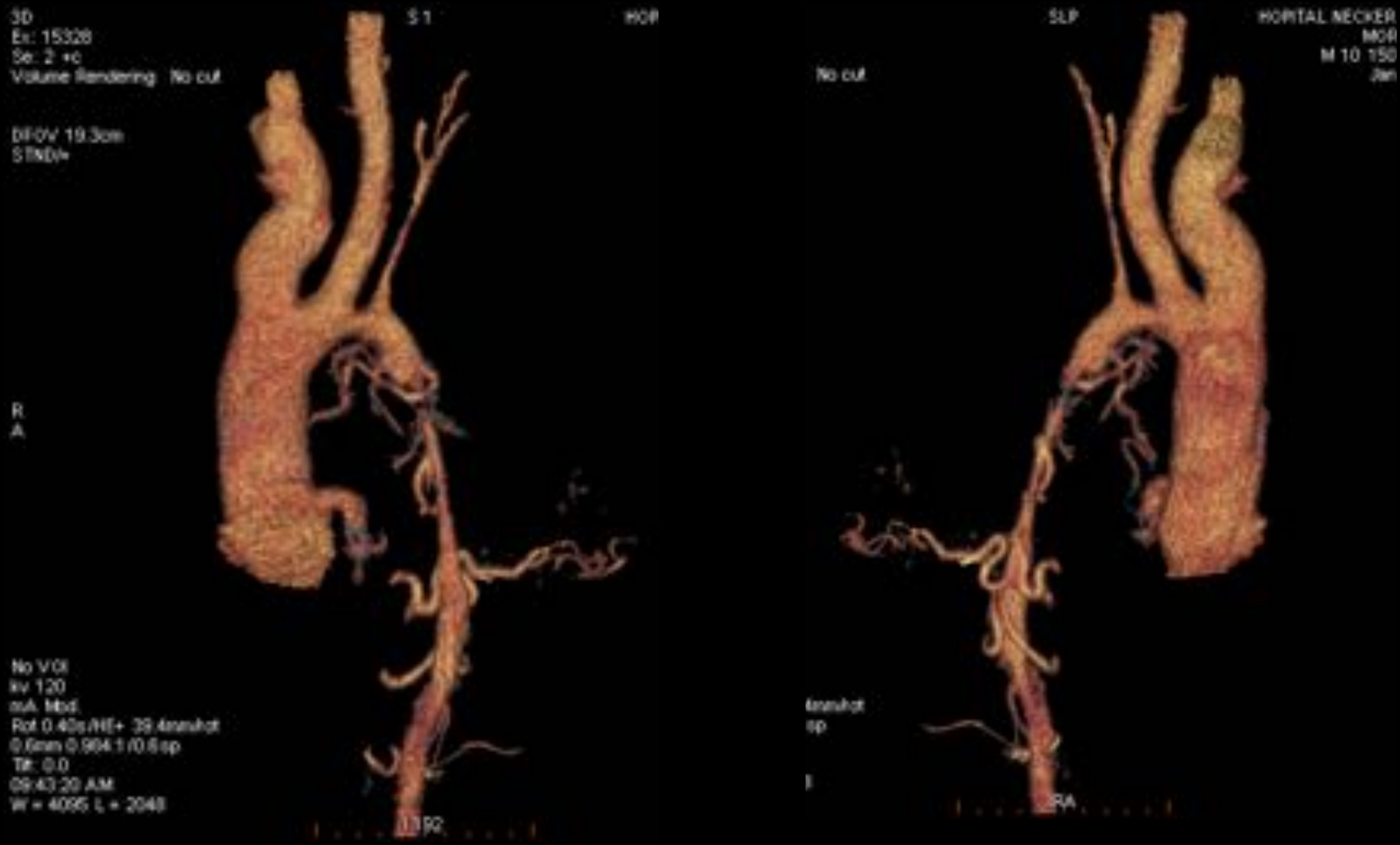
Tailored treatment and follow-up

- Optimize aortic geometry during repair
  - to prevent/delay systemic HT
- Detect systemic hypertension
- MRI best available tool for aortic anatomy-geometry, aortic function, LV mass and function
- Intra-cardiac surveillance : echocardiography
- Prevent CV events by minimizing usual CV risk factors
- Pending questions:
  - Reinforce monitoring of patients with exercise HT ?
  - Reinforce follow-up in patients with gothic geometry, vascular dysfunction, associated BAV…
  - Screening for aortic aneurysms in patients with patch aortoplasty
  - Screening for berry aneurysms in patients operated at an older age and in hypertensive patients

Coarctation thoracique basse et abdominale

Coarctation thoracique basse et abdominale

Coarctation thoracique basse et abdominale



Coarctation thoracique basse et abdominale
Création d'une aorte extra-anatomique

Coarctation thoracique basse et abdominale
Création d'une aorte extra-anatomique