

Sténoses de la voie d'éjection gauche

Coarctations de l'aorte

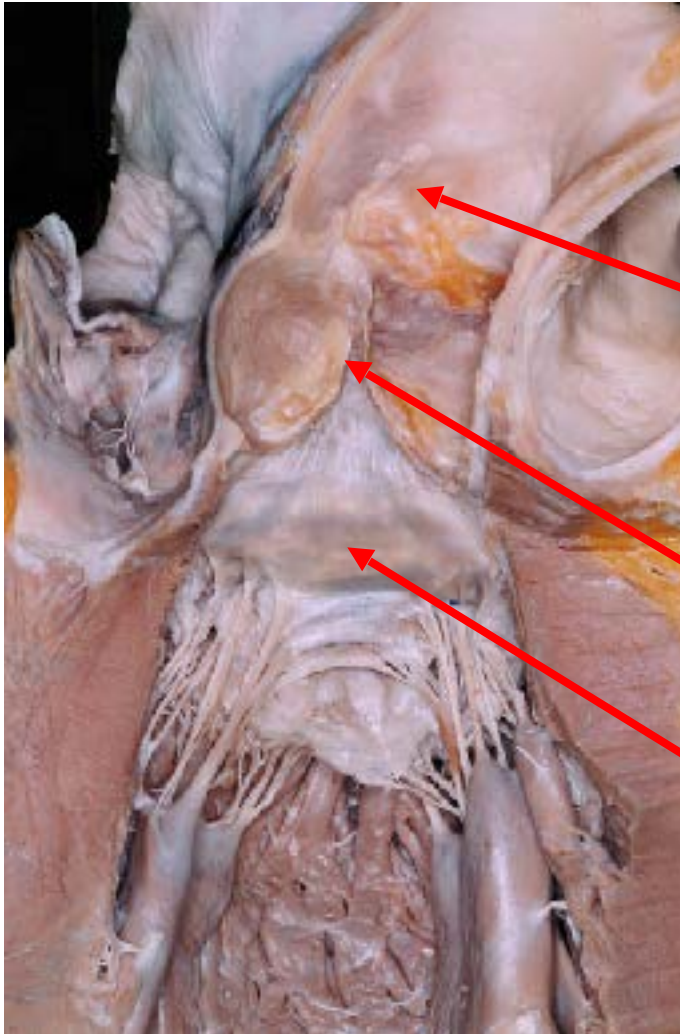
Pauline Parisot

Hôpital Armand Trousseau - Paris
Institut de l'enfant - Boulogne-Billancourt

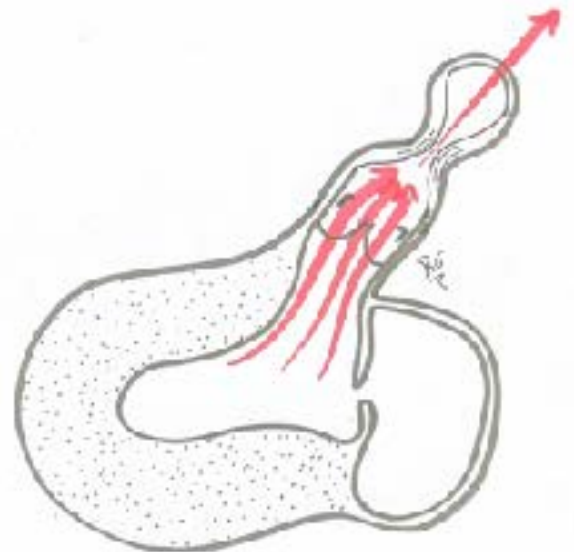
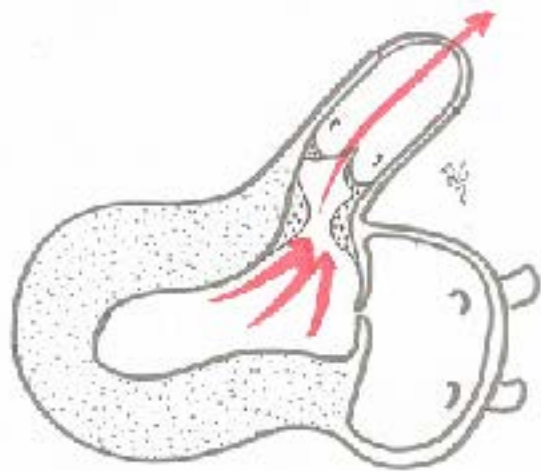
DU cardiologie congénitale et pédiatrique
22.01.2024

- sténoses sous valvulaires aortiques
- sténoses valvulaires aortiques
- sténoses supravalvulaires aortiques
- coarctation de l'aorte
 - isthmique
 - abdominale

Anatomie de la voie d'éjection gauche



- Coarctation de l'aorte
- Sténose aortique supra valvulaire
- Sténose valvulaire aortique
- Sténose sous valvulaire aortique



Sténoses sous valvulaires aortiques

Sténoses sous valvulaires aortiques

- obstacle associé à un CIV (malalignement)
- obstacle musculaire (CMH)
- membrane sous aortique = membrane fibreuse
- obstacle complexe fibro musculaire
- obstacle localisé (membrane)
- obstacle étendu : tunnel
- plus rarement :
 - tissu accessoire (septum membraneux ou valvulaire mitral: bleb)
 - insertions anormales de cordage ou pilier mitral

Valvar stenosis

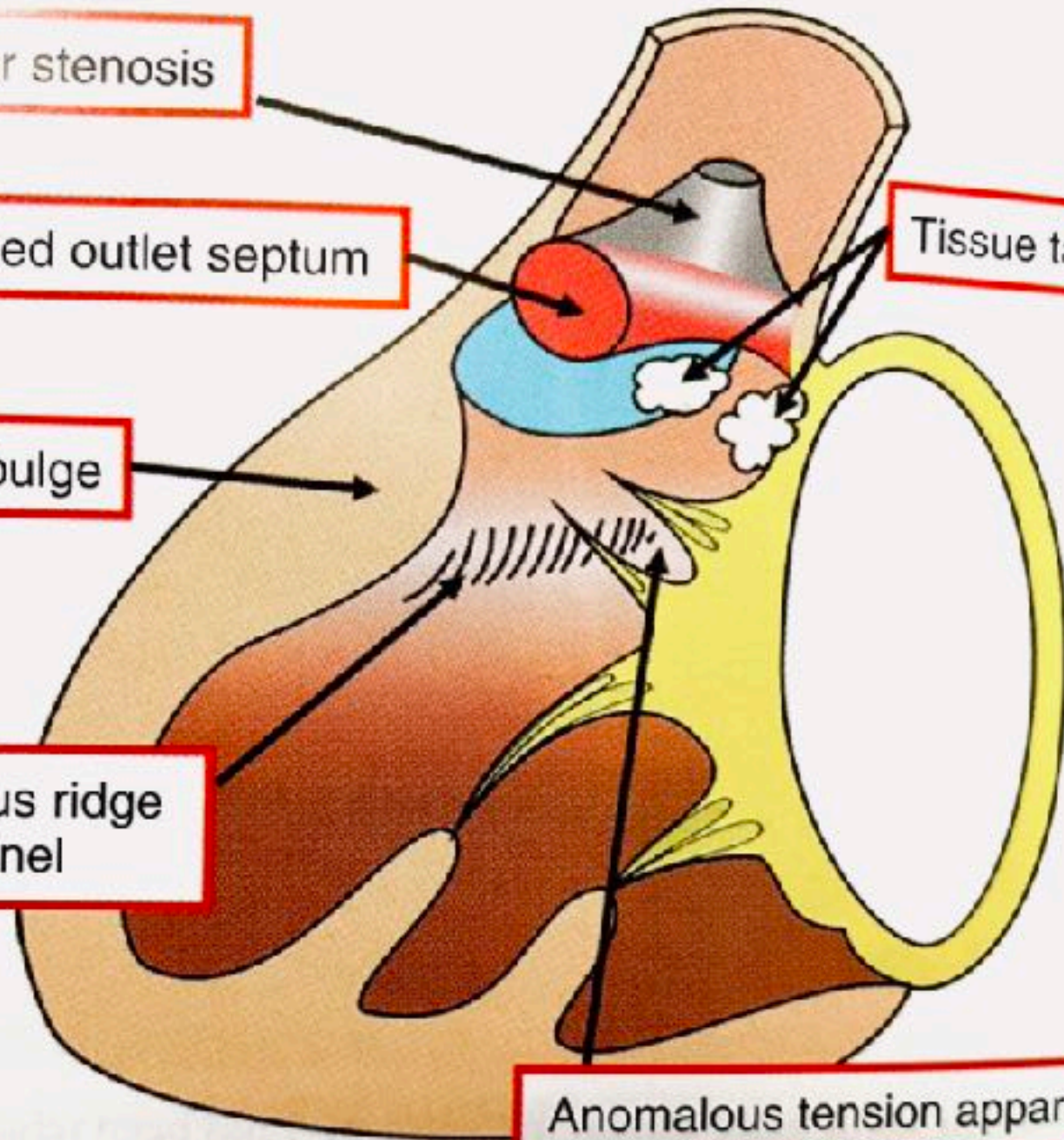
Deviated outlet septum

Tissue tags

Septal bulge

Fibrous ridge
or tunnel

Anomalous tension apparatus

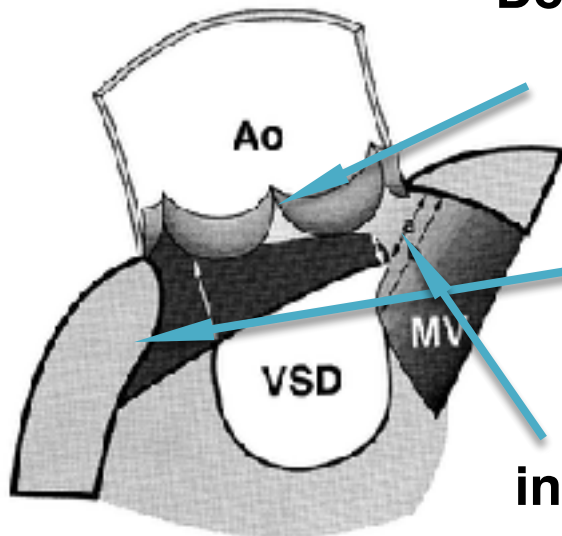


Anatomie de la voie d'éjection gauche

The Surgical Anatomy of the Left Ventricular
Outflow Tract in Hearts With Ventricular Septal
Defect and Aortic Arch Obstruction

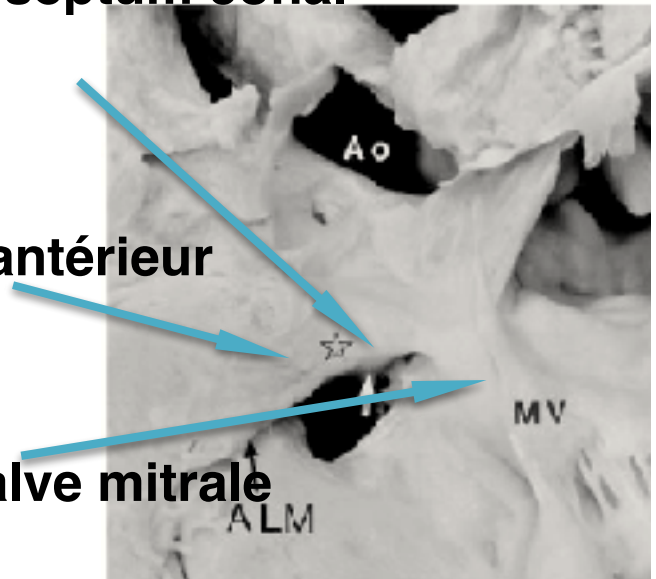
Yuzhi Shiohara, MD, and Anken E. Becker, MD

Déplacement postérieur du septum conal

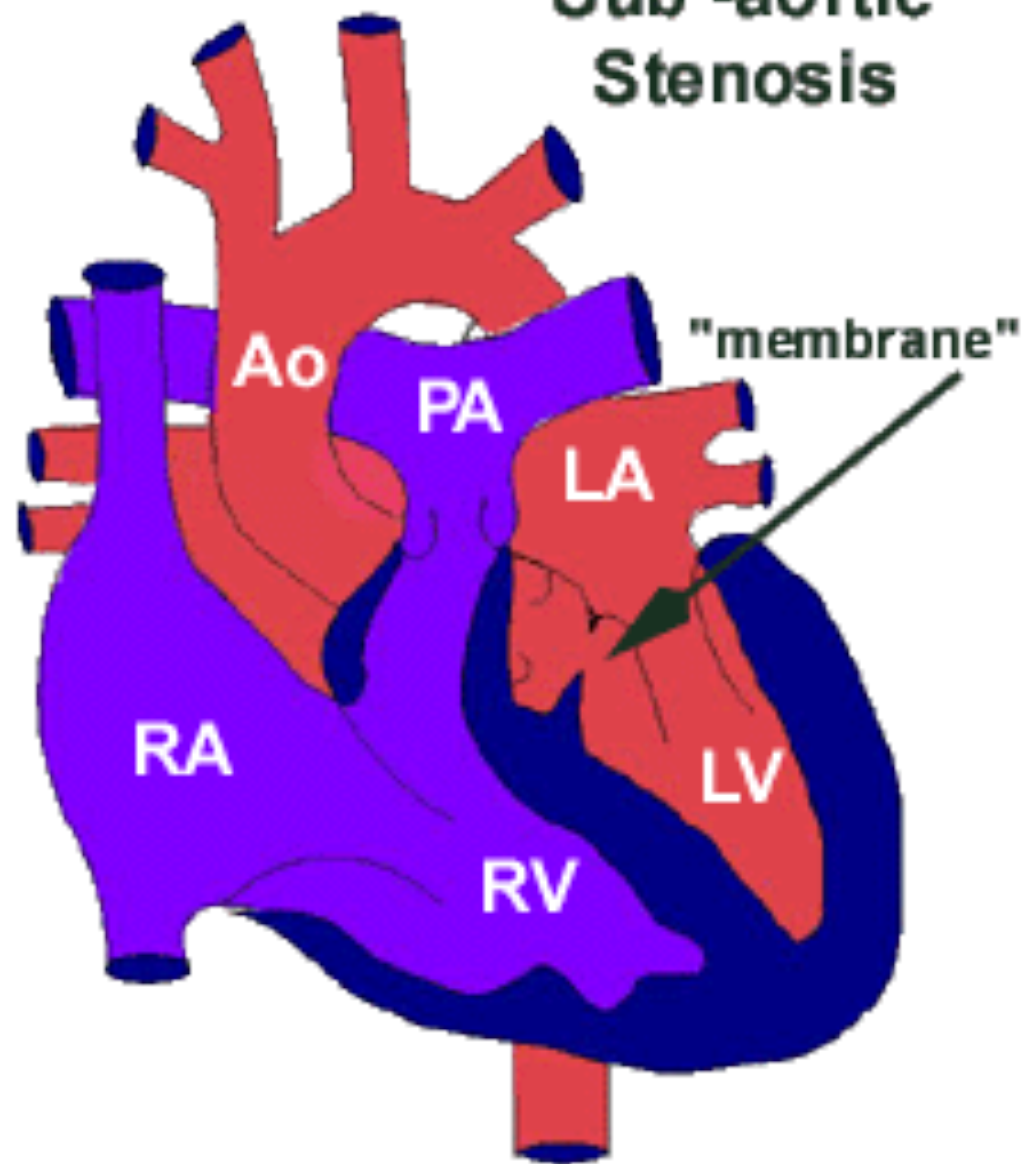


« bulge » musculaire antérieur

insertion anormale de la valve mitrale



Sub-aortic Stenosis



membrane sous aortique



membrane sous aortique

- lésions le plus souvent acquises
- lésions évolutives
- facteurs anatomiques prédisposants

conséquences hémodynamiques

- hypertrophie ventriculaire gauche
- insuffisance aortique
- risque d'endocardite

Clinique

- souffle mesosystolique bord sup D/G du sternum, irradiant vers la carotides
- +/- S protoD si lao associée
- forme sévère : ↘ pouls périphériques, syncope, angor, insuffisance cardiaque

Diagnostic échographique

- nature de l'obstacle
- siège (proximité valve aortique)
- sévérité : gradient P
- retentissement : HVG
- IAo
- lésions associées : bicuspidie aortique, CIV, Coa, anomalie mitrale (fente de la voie d'éjection)
- surveillance évolutivité

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V

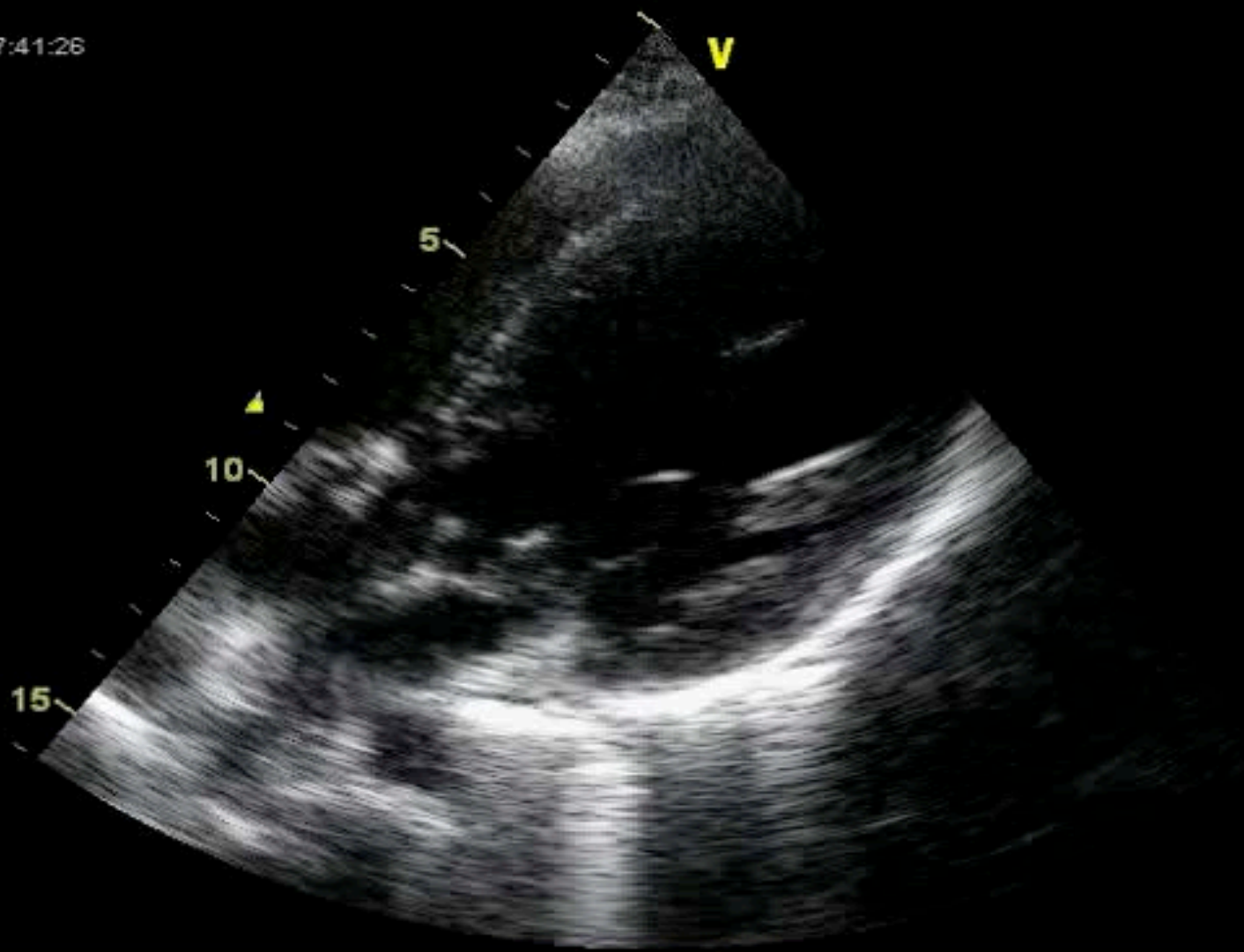
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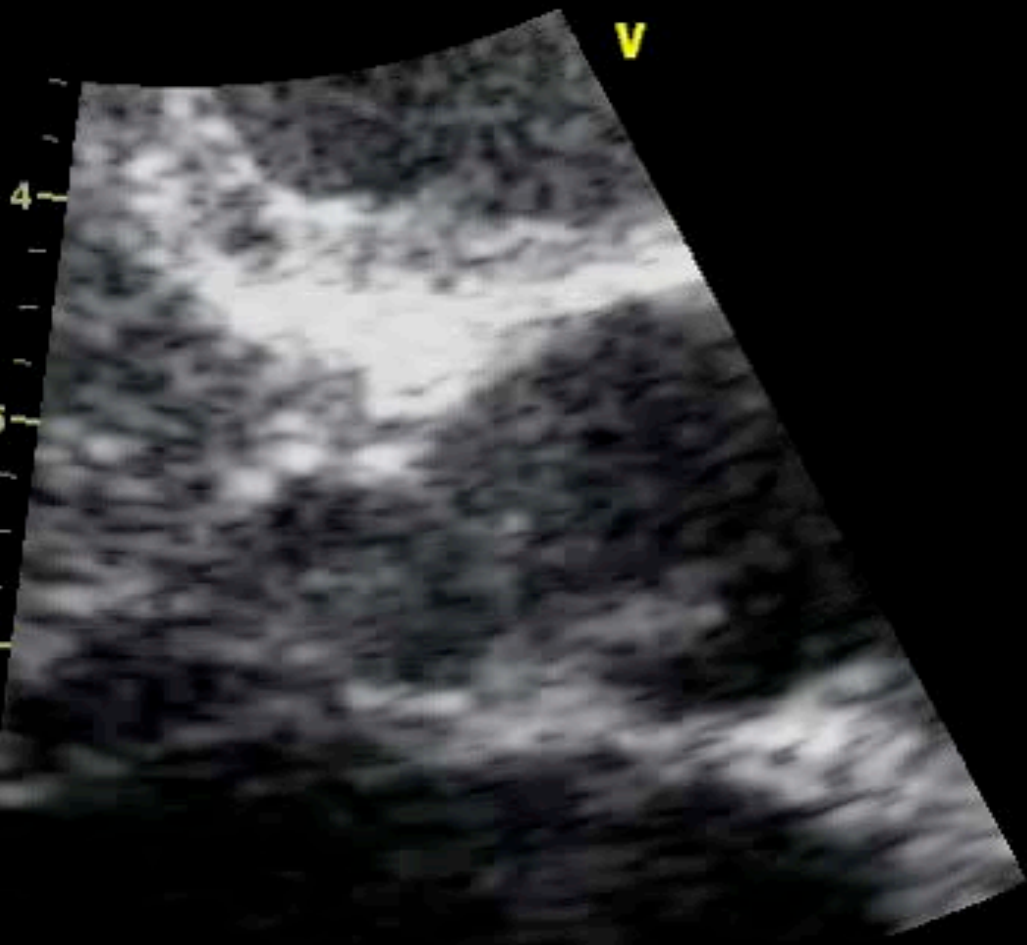
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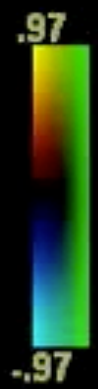
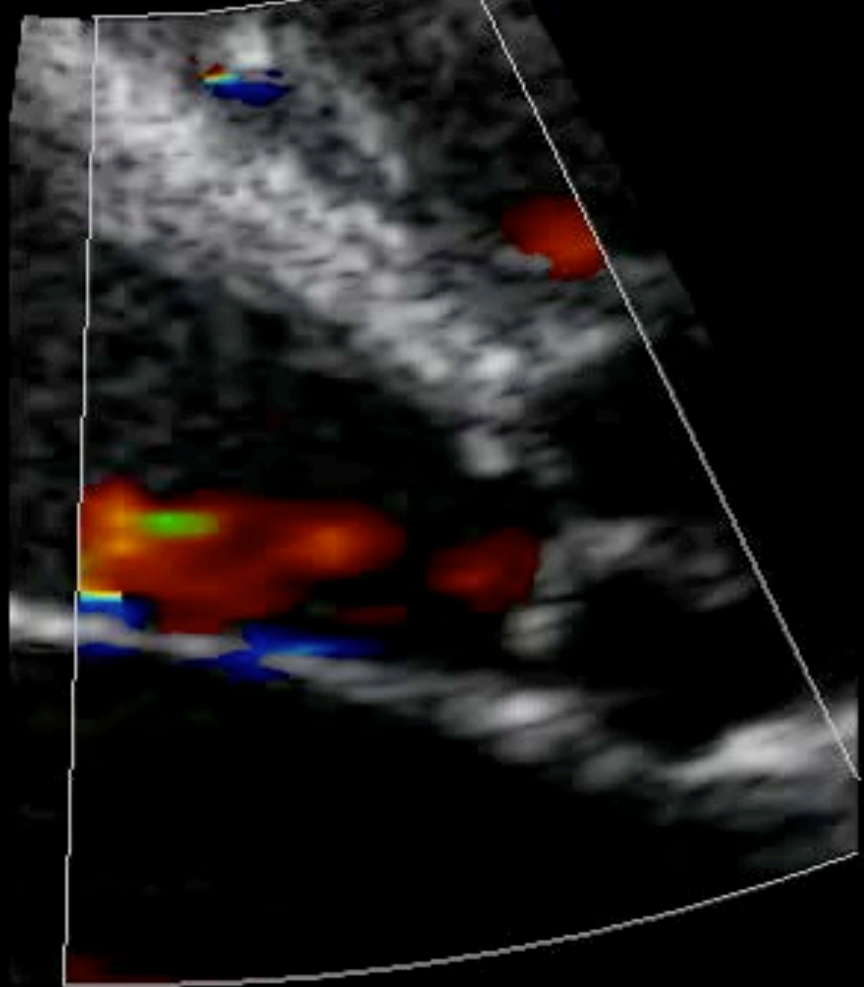
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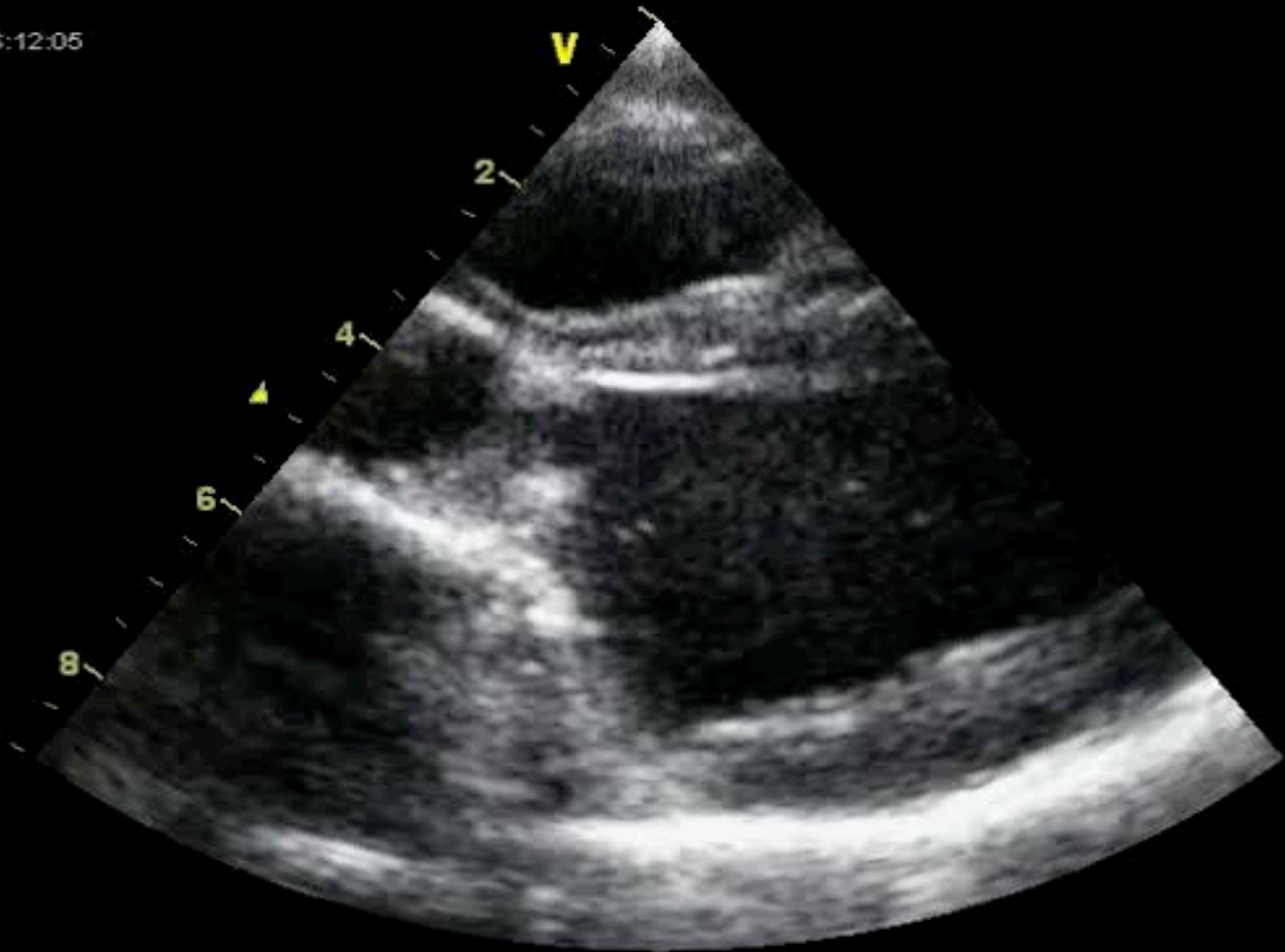


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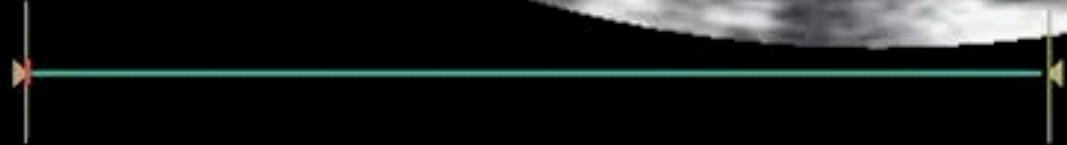
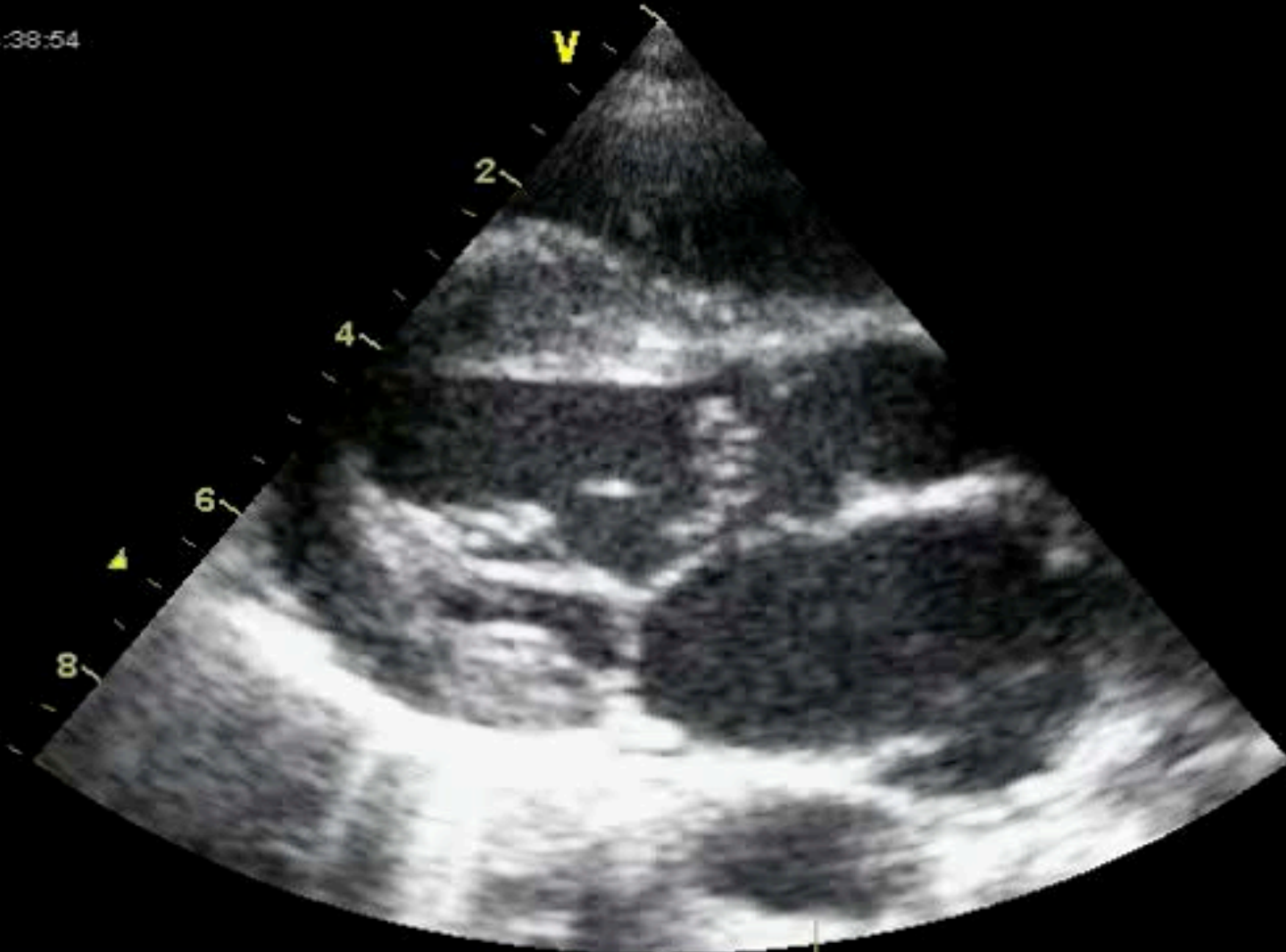
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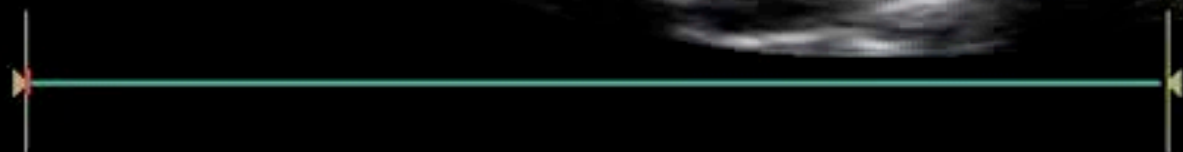


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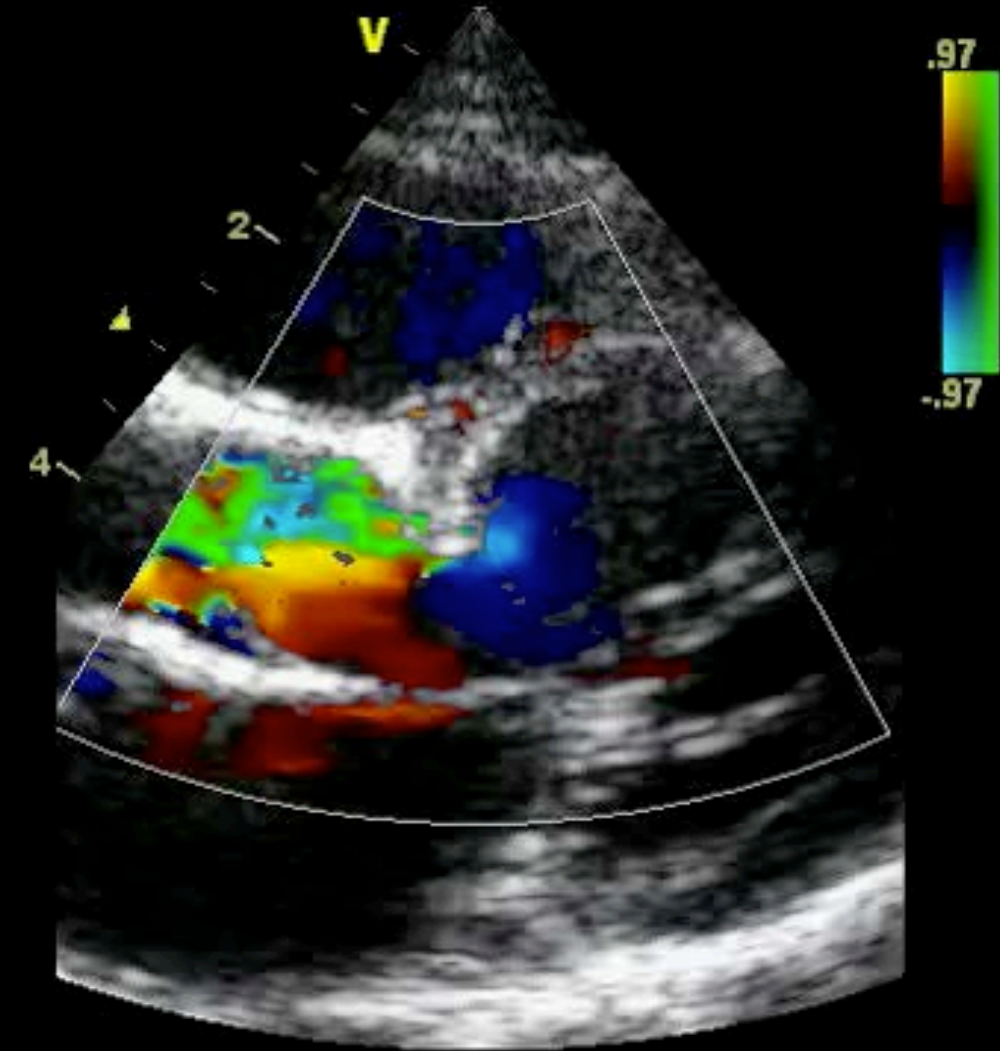
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Facteurs d'évolutivité

- sévérité obstruction VG
- distance obstacle - valve
- dysplasie valvulaire
- âge au diagnostic

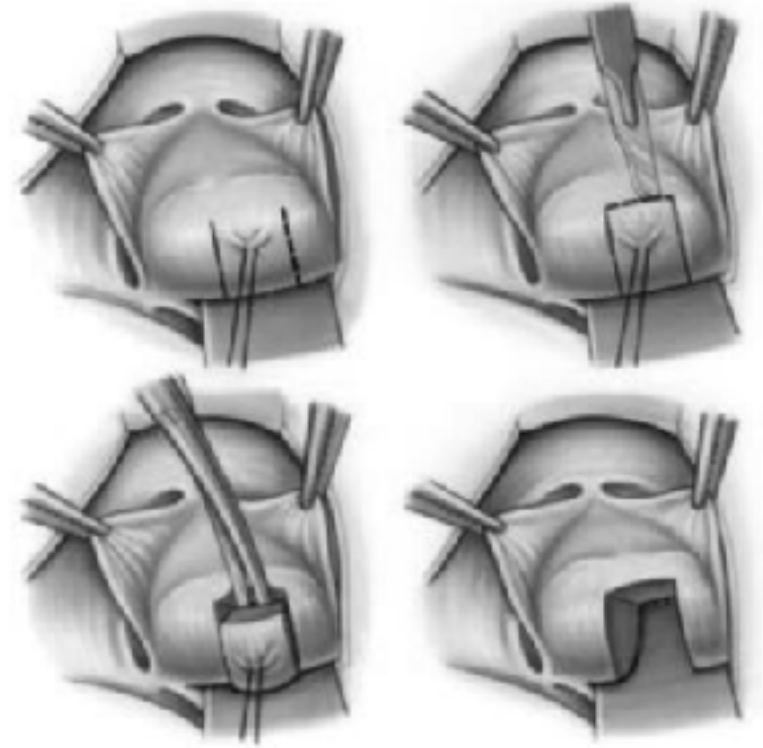
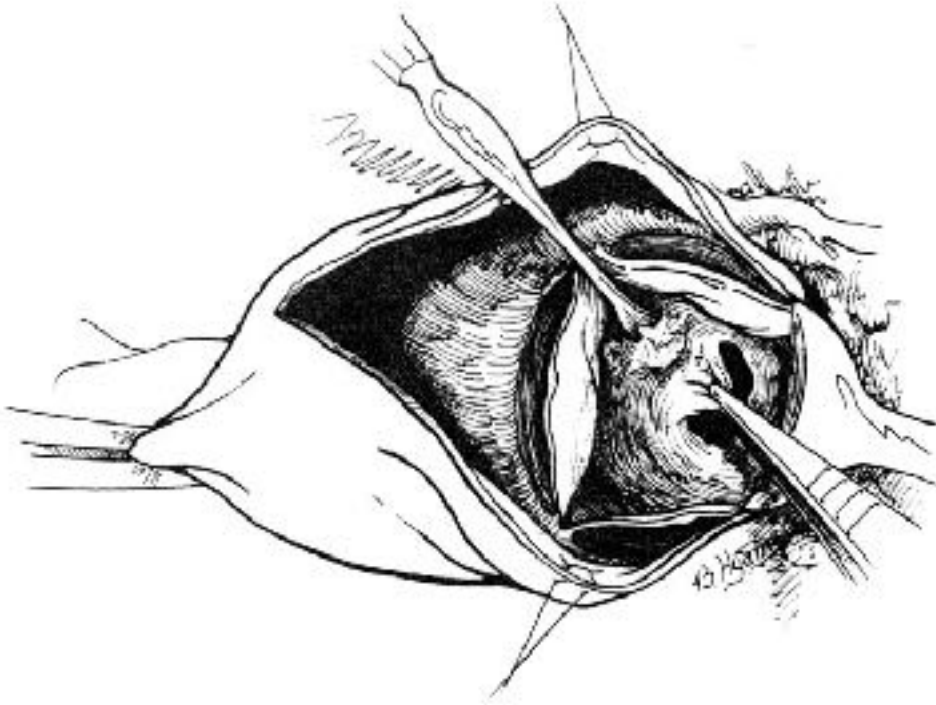
Membrane sous aortique

Traitement : chirurgical

- exérèse par énucléation
- réséquer les extensions sur valve aortique / mitrale
- myectomie systématique (prévention de la récurrence)



Enucléation + myectomie septale



abord transvalvulaire aortique

Traitement : chirurgical

- mortalité $\simeq 0\%$
- BAV post-opératoire : 0 - 6%
- CIV iatrogène : 2 - 4%
- lésion valve aortique : 3 - 6%
- lésion valve mitrale : 0 - 4%

Traitement chirurgical : indications

- indication “curative” : obstacle sévère

UE : gradient moyen 40mmHg vs US grad max 50mmHg

- indications “préventives” ?
 - obstacle peu sévère avec IAo minime ou pas d'IAo?

indications préventives

- sténose peu sévère (gdt moy < 30)
- sans insuffisance aortique

- . chirurgie préventive non justifiée
- . surveillance régulière

- sténose modérée (30 < gdt moy < 50)
- avec ou sans IAo

- . chirurgie « préventive » sans doute justifiée
- . risque évolutif accru

- sténose peu sévère (gdt moy < 30)
- avec lao minime

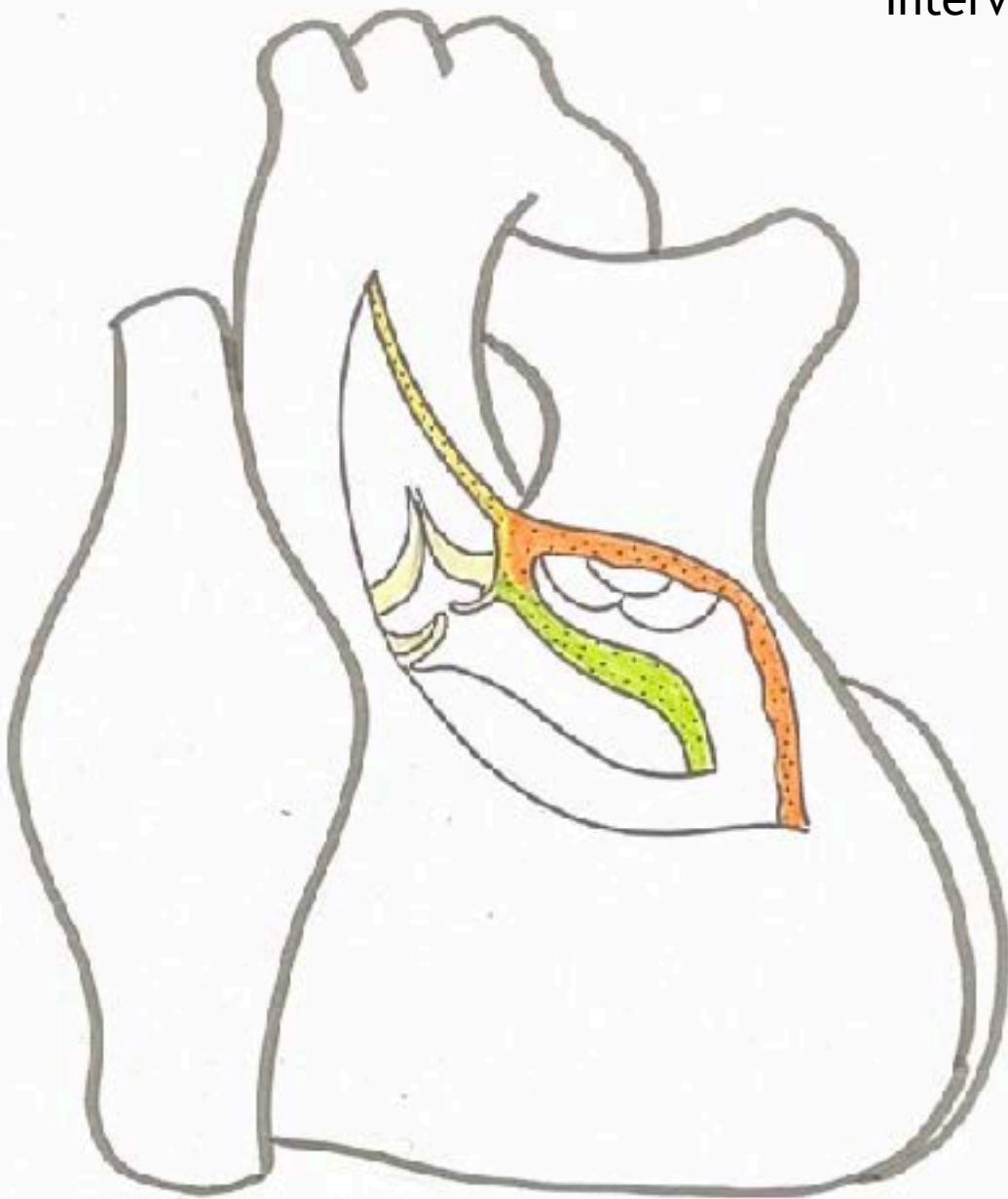
- . chirurgie préventive non justifiée
- . surveillance plus rapprochée surtout si
 - proximité obstacle-valve
 - dysplasie valvulaire
- jeune enfant ou adulte

Intervention de Konno

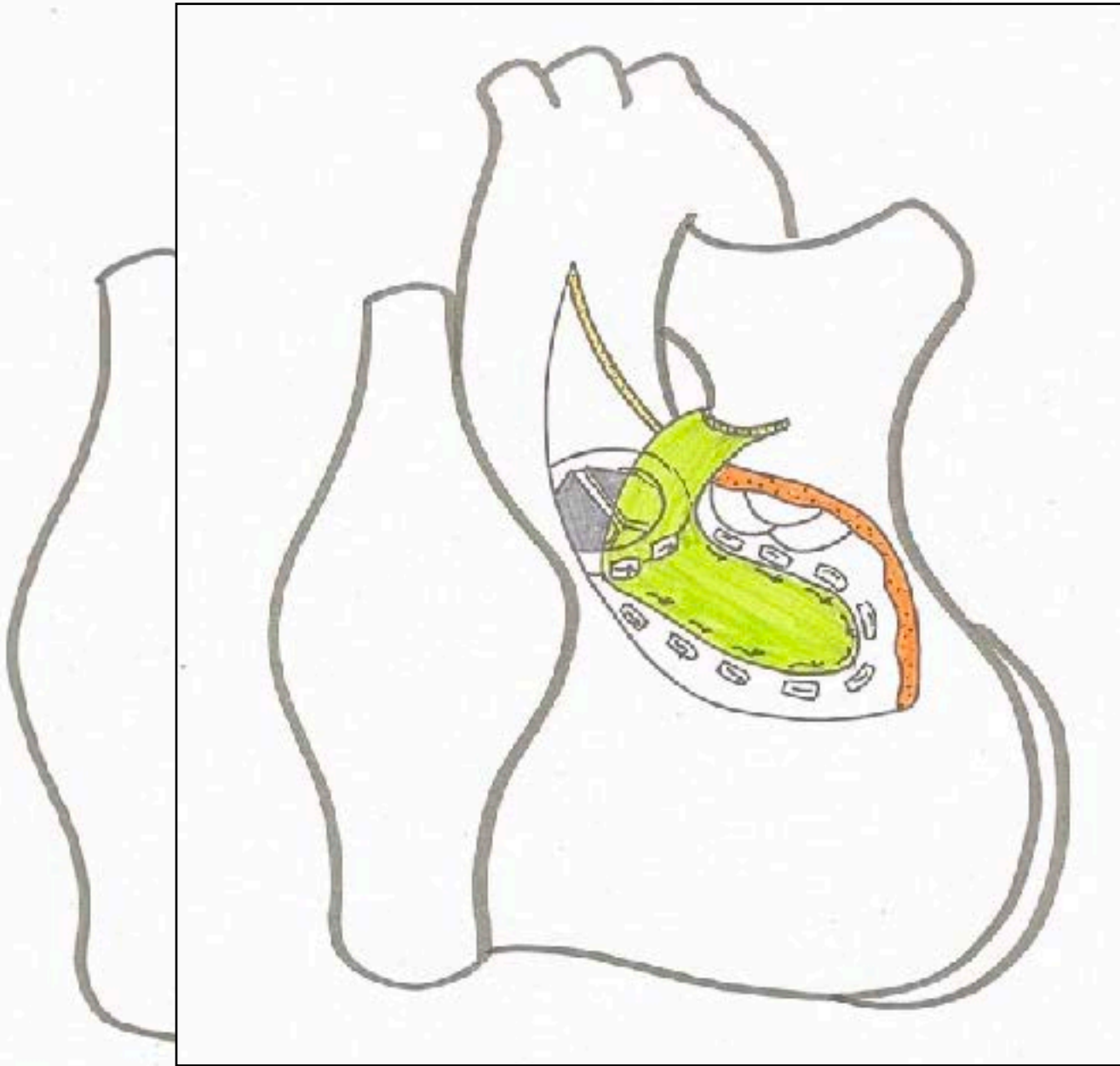
Obstacle sous valvulaire et valvulaire aortique : Konno ou Ross Konno

Obstacle sous valvulaire ao complexe et étendu : Konno modifié

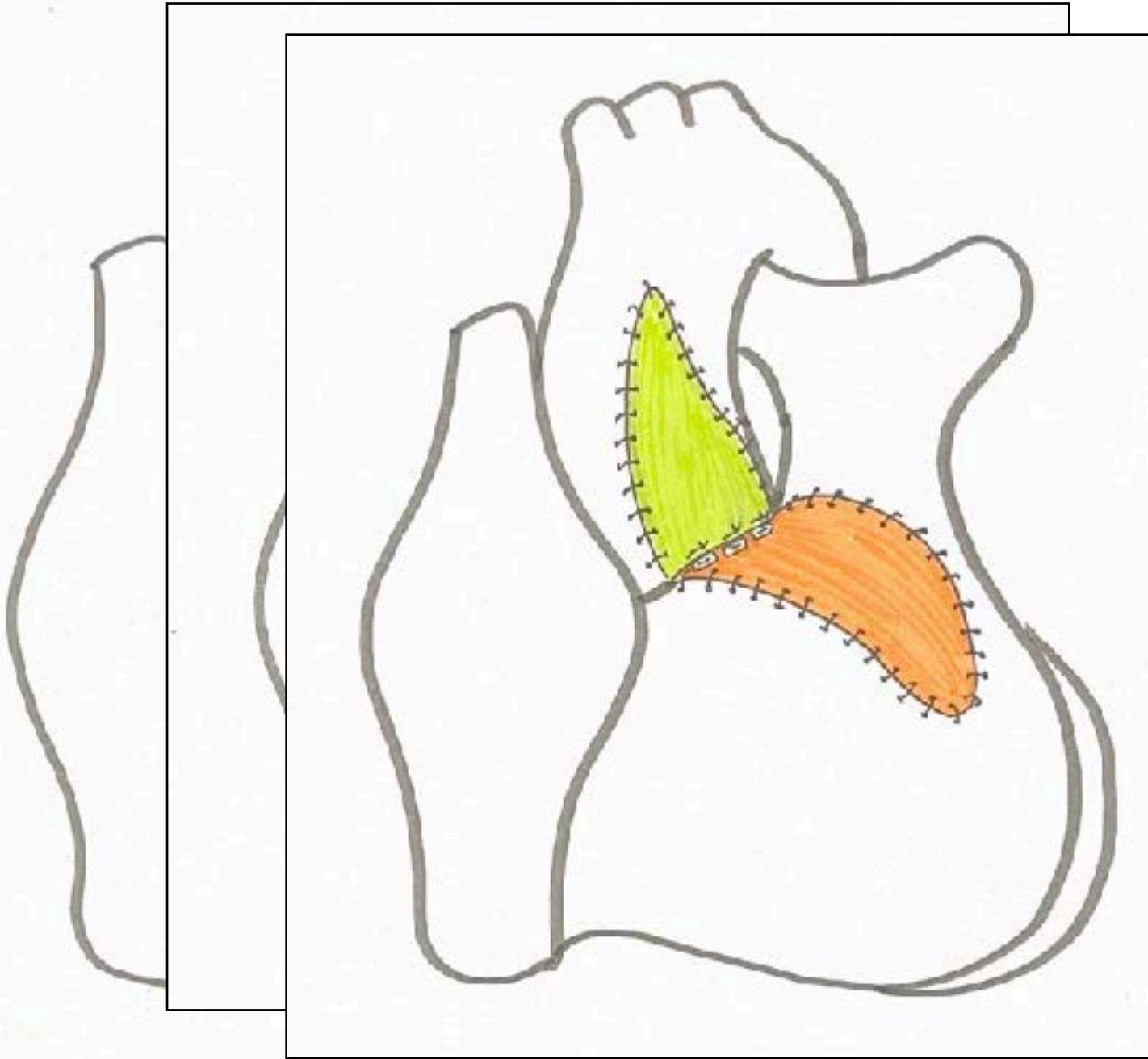
Intervention de Konno



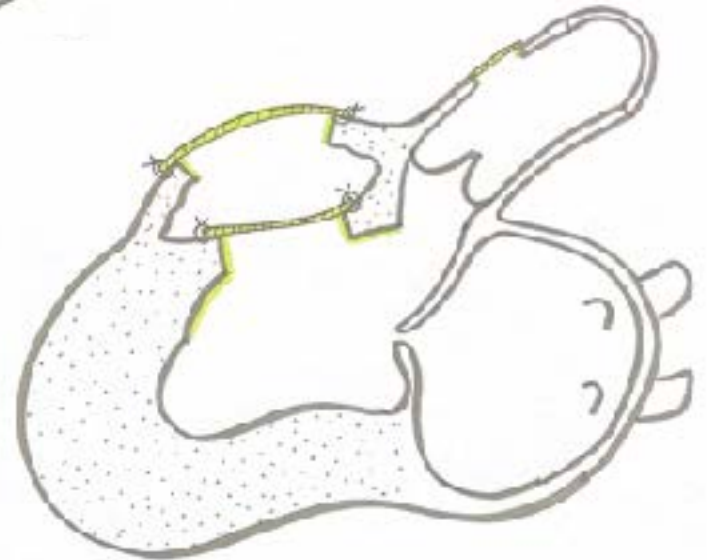
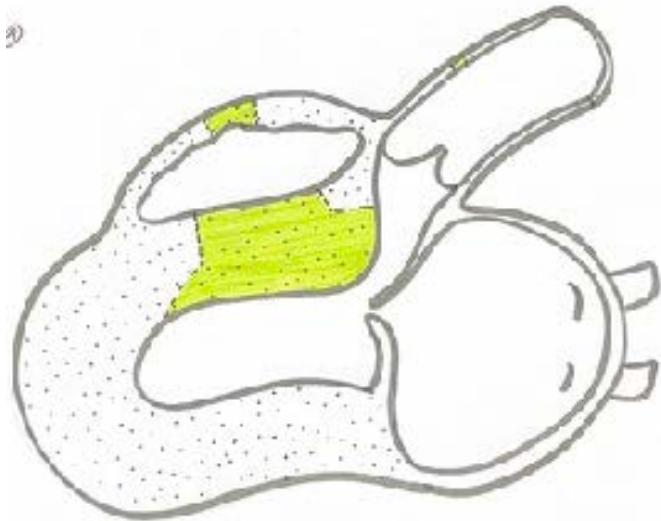
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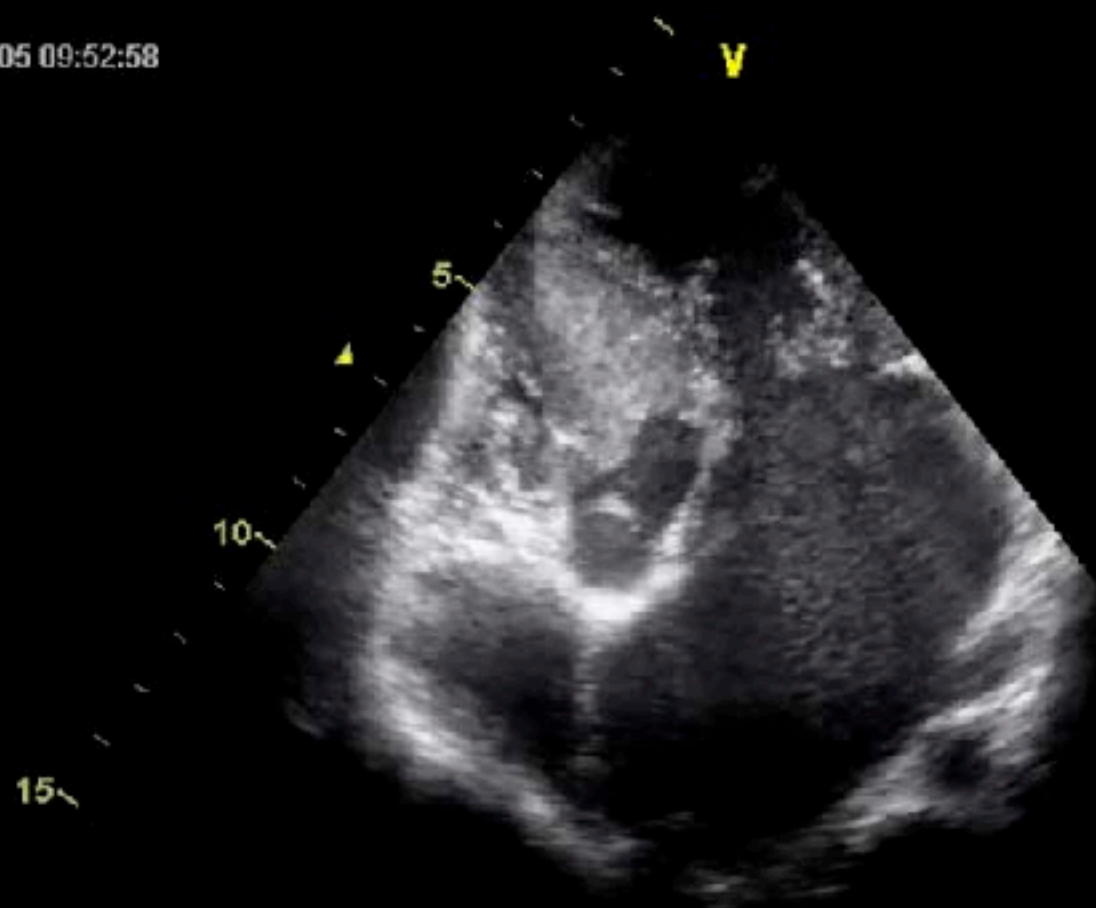
Intervention de Konno



Intervention de Konno modifiée

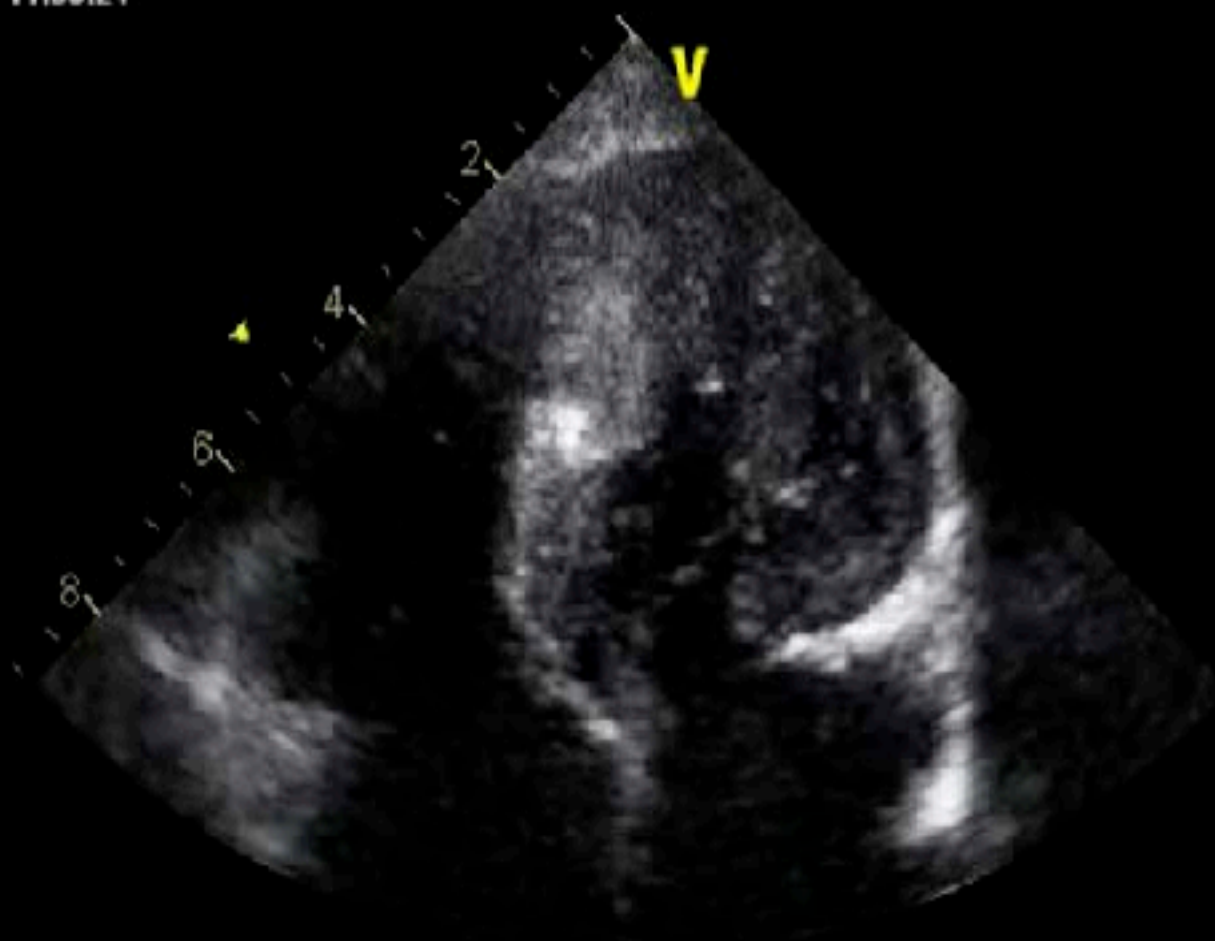


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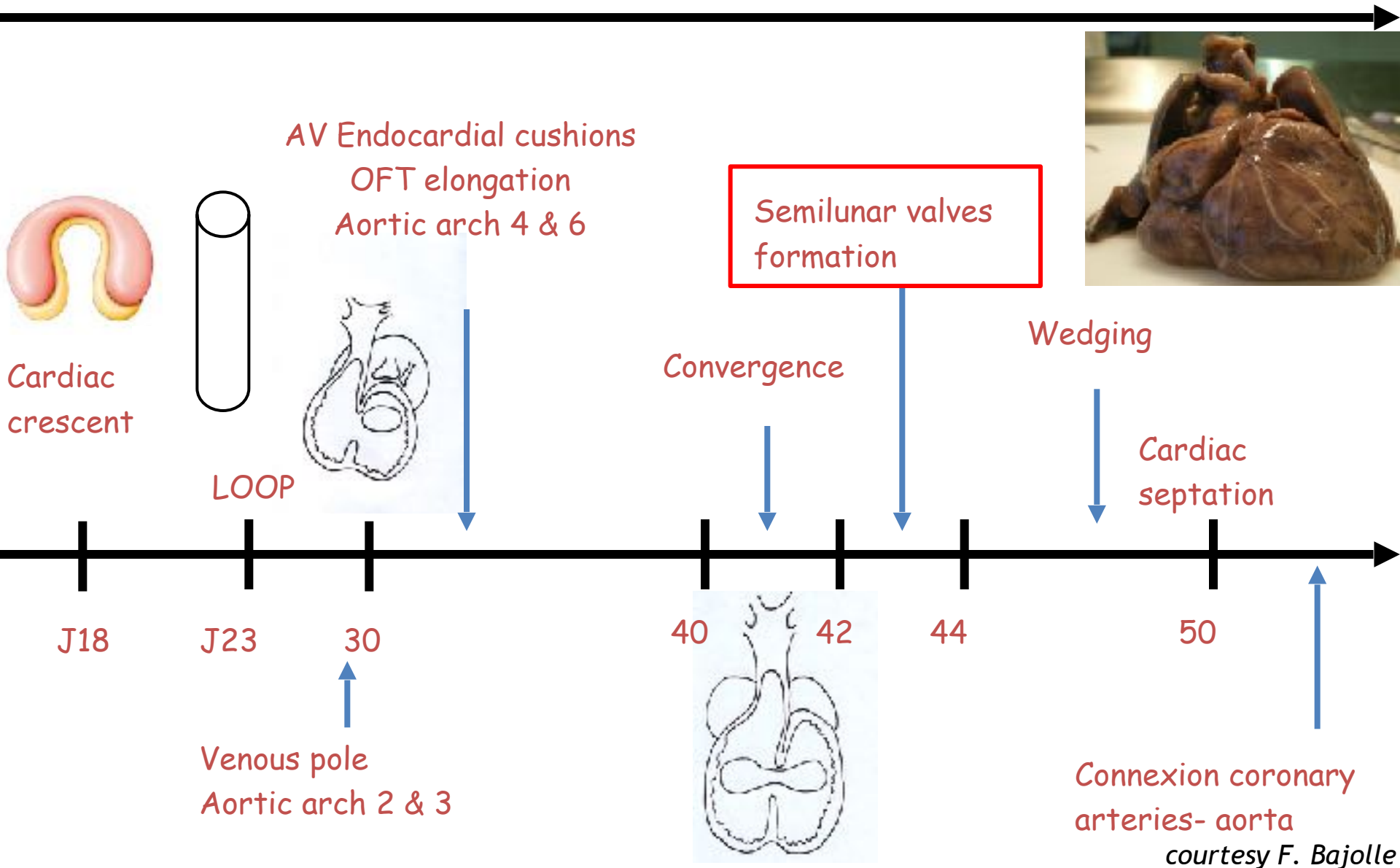
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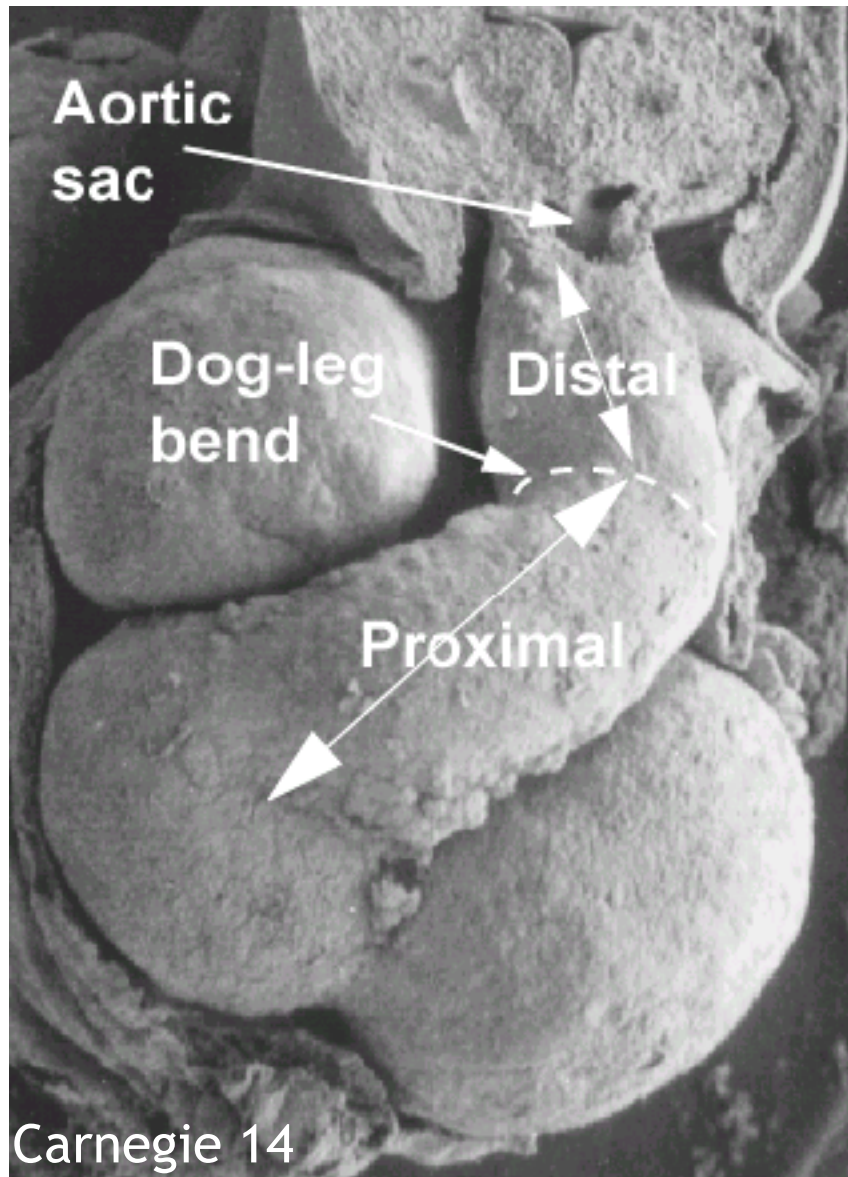
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sténose valvulaire aortique

Développement cardiaque

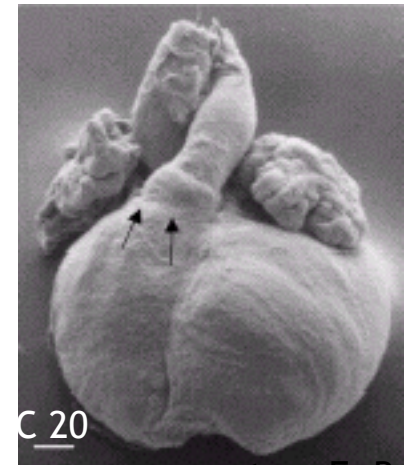


Morphogénèse de la voie efferente



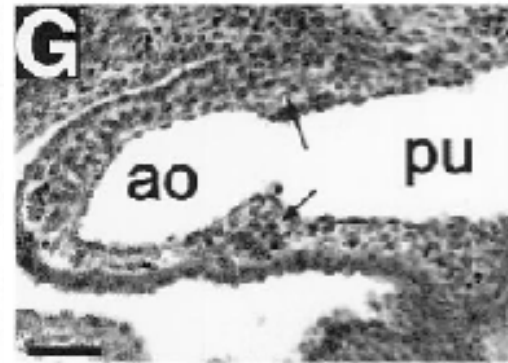
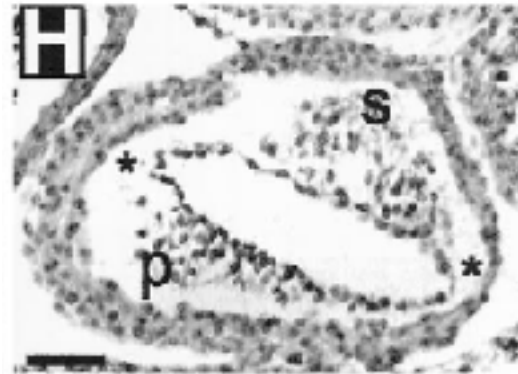
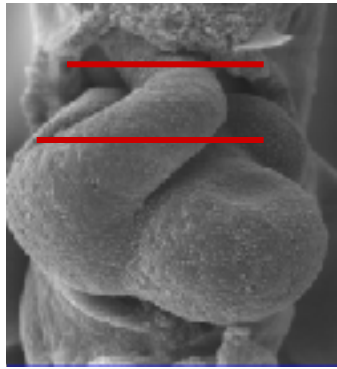
Voie efferente proximale

- feuillets valvulaires et parois sinus
- septum de la voie d'éjection



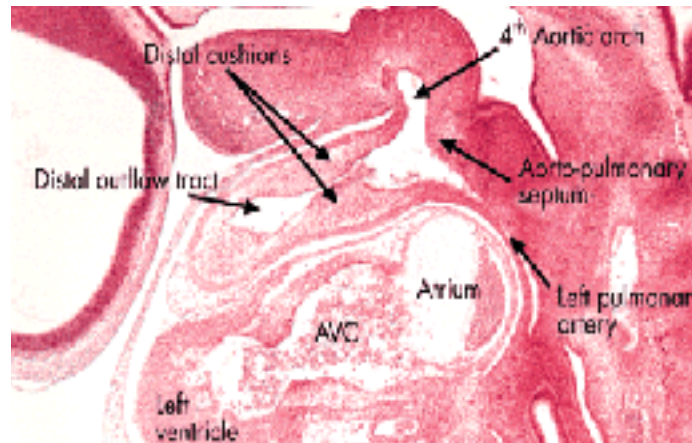
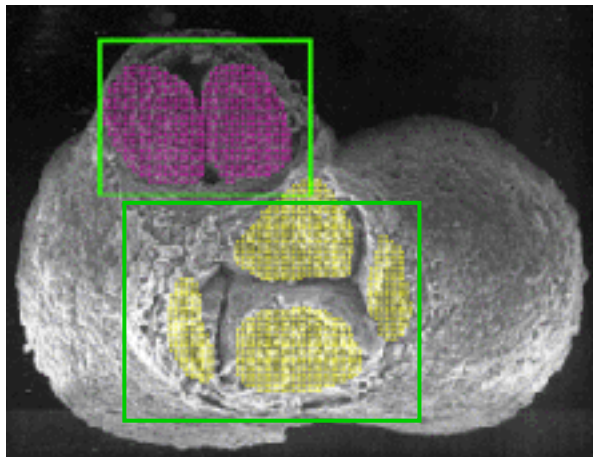
Coussins endocardiques

Les valves semilunaires ont pour origine une production accrue de MEC qui initie la formation des bourgeons endocardiques dans la voie efferente



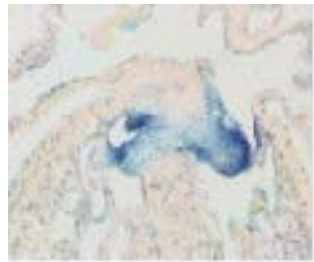
Costell et al., 2002

La formation des coussins endocardiques est initiée par des signaux émanant du myocarde de la VE qui va diffuser dans la MEC et subir une transition épithéliomesenchymateuse

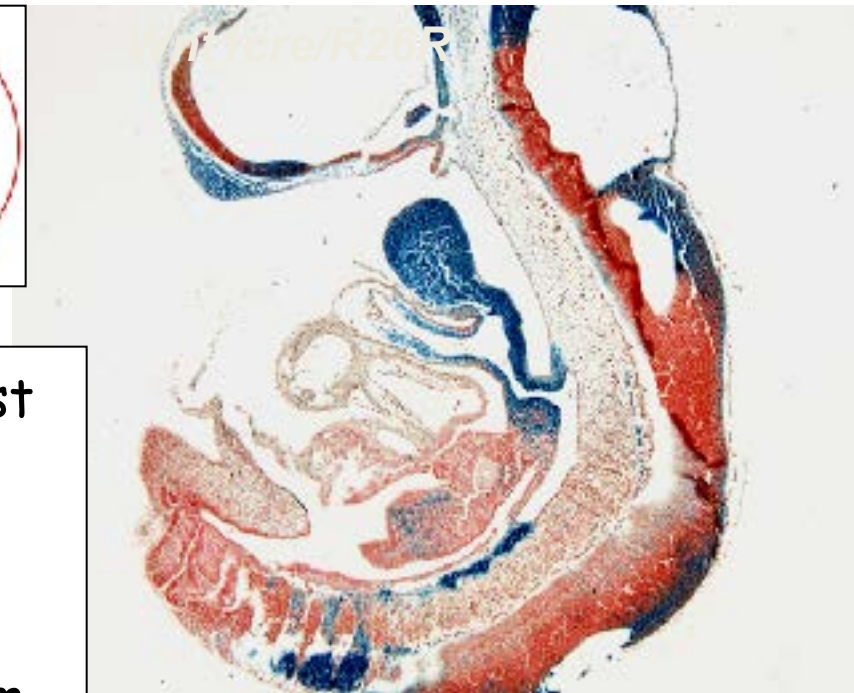
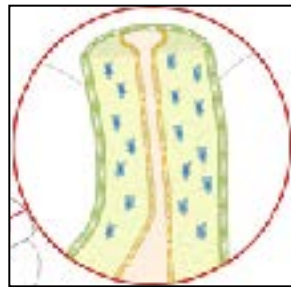


Anderson et al., 2003

Endocardial cushions contribute to the OFT remodeling



Tie2cre/R26R



Neural crest



Cushions

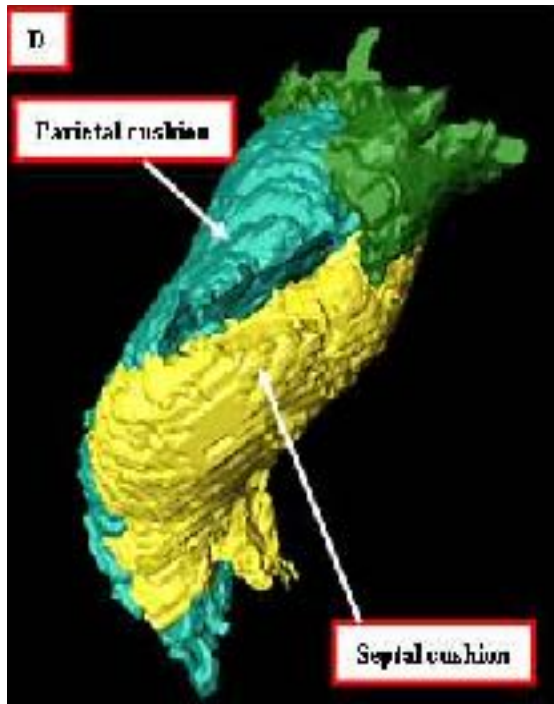


Myocardium

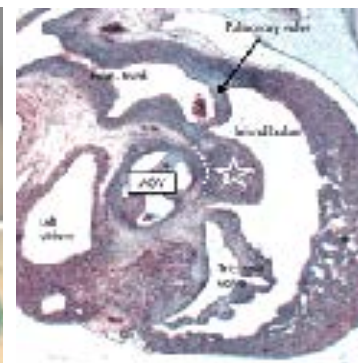
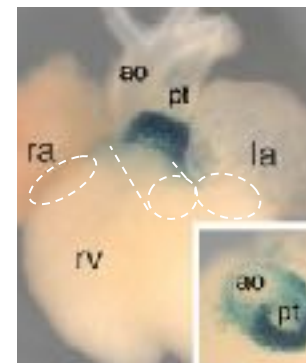
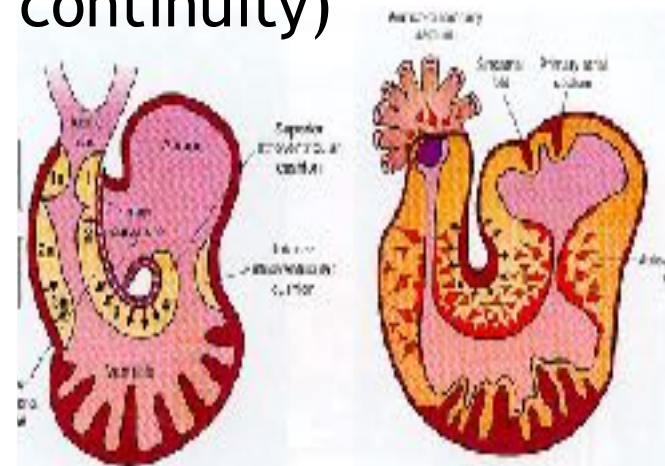
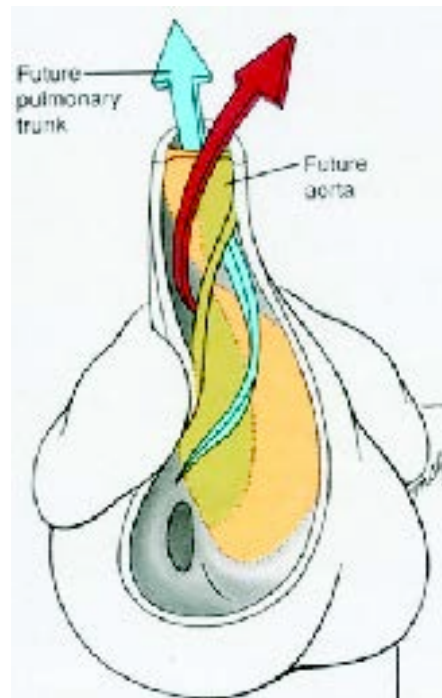
- Endocardial cushions of the proximal outflow tract and the AVC-derived from the endocardium, due to signals from myocardium
- Endocardial cushions of the distal outflow tract - contribution from cardiac neural crest

Proximal outflow tract remodeling

- Shape of cushions : spiraling cushions
 - proximal OFT: parietal and septal cushions
 - distal OFT: caudal and cranial cushions
- Myocardization: cushions are replaced by myocardial cells in the inner curvature (mitro-aortic continuity)

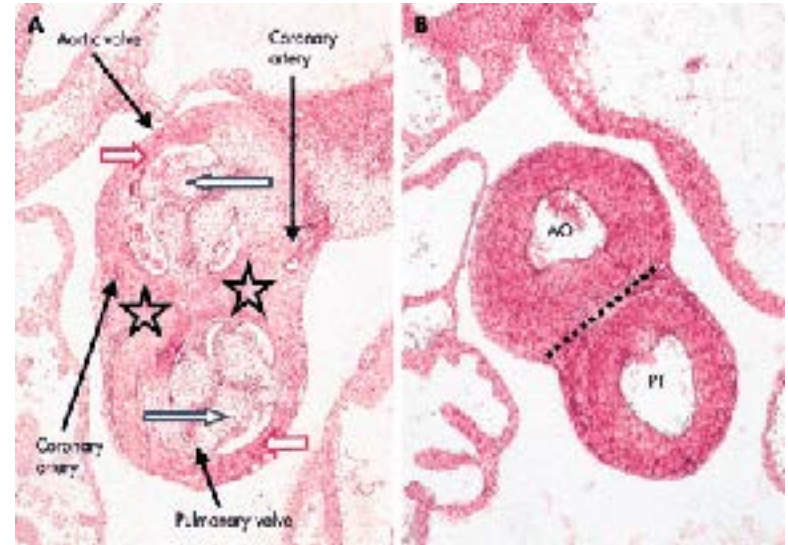
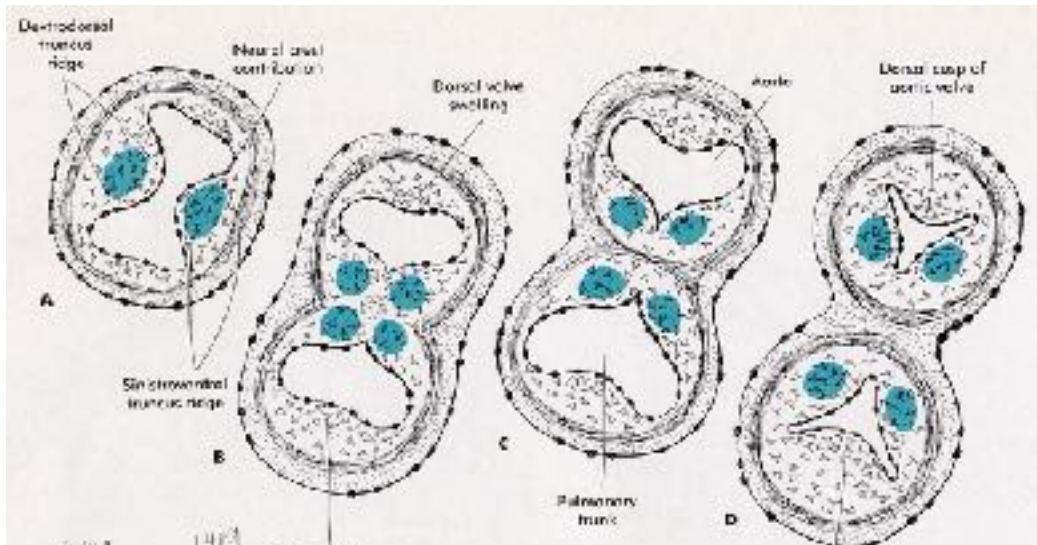


Courtesy B. Anderson



Semilunar valve formation

- Cushions undergo cell proliferation and grow towards each other until a process of fusion between the two cushions.

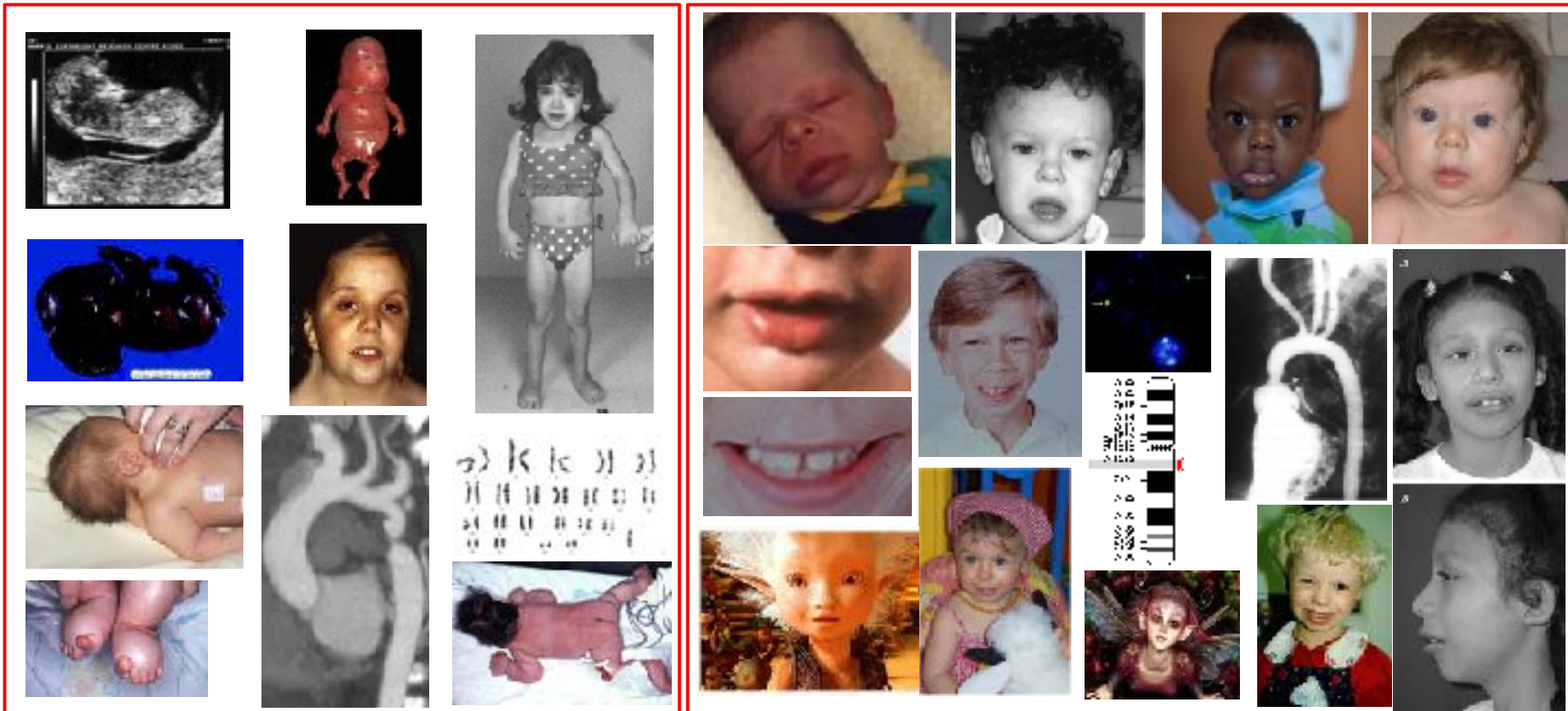


- Differentiation, apoptosis and ECM remodeling: formation of thin protruding leaflet valves composed of endocardial cells and ECM
- Cardiac neural crest cells reach the OFT cushions and contribute to the formation of the aortic and pulmonary valves.
- The wall of the distal OFT change from myocardium phenotype to an arterial one

Bicuspid Aortic valve

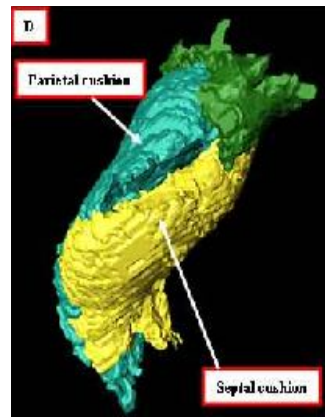
- Embryologic hypothesis for BAV development
 - Perturbations in endocardial cushions development
 - Improper development of the conotruncal ridges
 - Conotruncal malseptation (quadricuspid pulmonary valve?)
 - Valve cushions agenesis
 - Abnormal extracellular matrix
 - Fusion between cushions
- Associated congenital cardiovascular malformations (20-50%)
 - Coarctation (50% BAV)
 - PDA
 - VSD
 - Shone's Syndrome (Left heart obstruction): supraaortic mitral ring, parachute mitral valve, subaortic stenosis, aortic coarctation , BAV (2/3)

- Anomalies génétiques associées
 - Syndrome de Turner : 30% of CHD, 2/3 LHO, 30% BAV
 - Syndrome de Williams et sténose supra-avalvulaire aortique: 15% BAV



Development of BAV and Aortopathy

Transcription factors and signaling pathways implicated in EMT and cushions morphogenesis: TGF- β superfamily, VEGF, ErbB, NFATc1, Notch, WntB-catenin, Twist, Sox9, Tbx20, GATA4....



- *Associated genetic anomalies*
 - Gene TGFB receptor
 - Filamin A...

Conclusion

- Some BAV result of
 - genetic disease
 - of the fusion of 2 normal cusps
 - associated with head/neck anomaly may be due to a disorder of NCC
 - extracellular matrix anomaly....
- Altered fetal blood flow through the aortic valve may be an important factor producing the bicuspid condition

BAVs are likely the result of a complex developmental process involving genetic, cell behavior anomaly, cell interaction disorders and haemodynamics

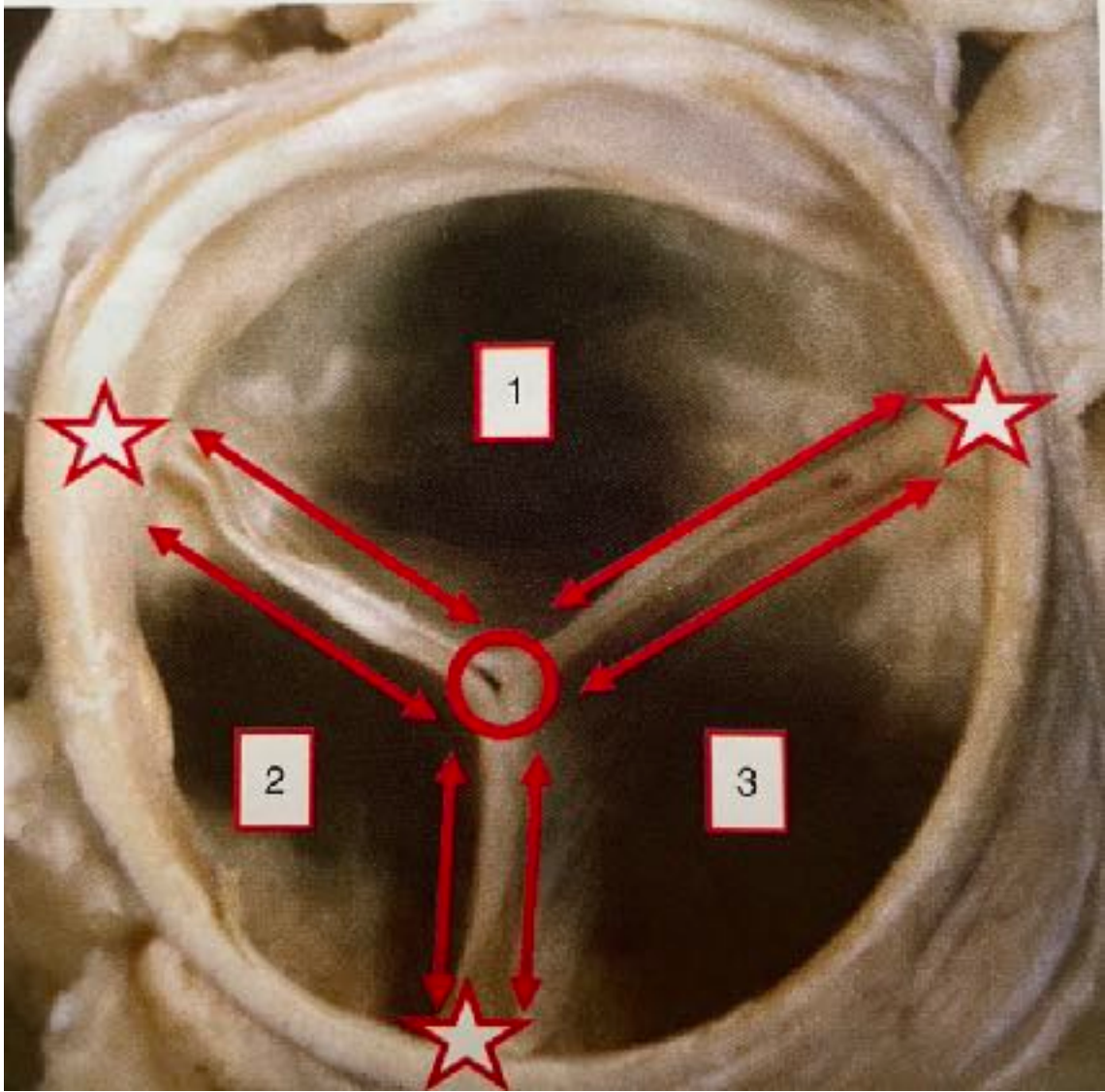
Haemodynamic approach: mechanisms of Left Heart Obstruction

Familial screening in adults

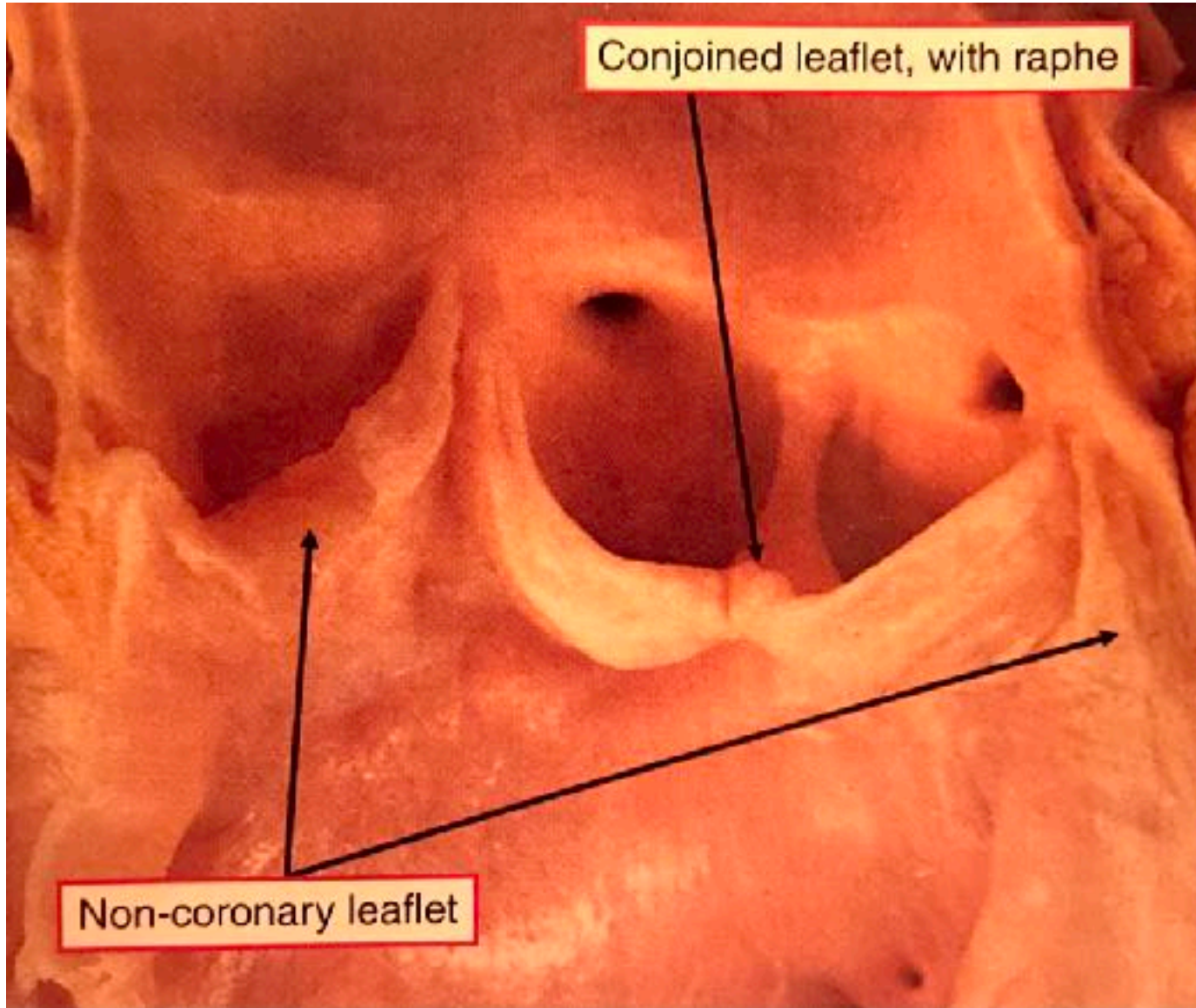
- Echocardiographic familial screening is recommended for first degree relatives of patients with BAV
- Familial recurrence of BAV occurs in approximately 9%
- Autosomal-dominant with reduced penetrance

These LHO anomalies belonged to a same embryological

- The development of heart chambers is related to the pattern of the combined foetal blood flow that passes through them (Abraham Rudolph)
- recurrences of different severity within the same family
- prenatal progression of obstructive left heart diseases
- identification of the same mutation in *NOTCH1* in patients of the same family with a different cardiac phenotype



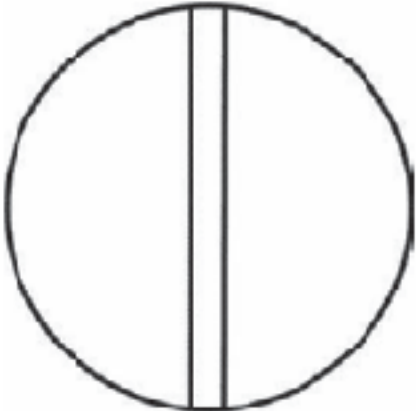



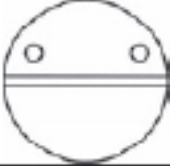




Sténose aortique : valve bicuspide

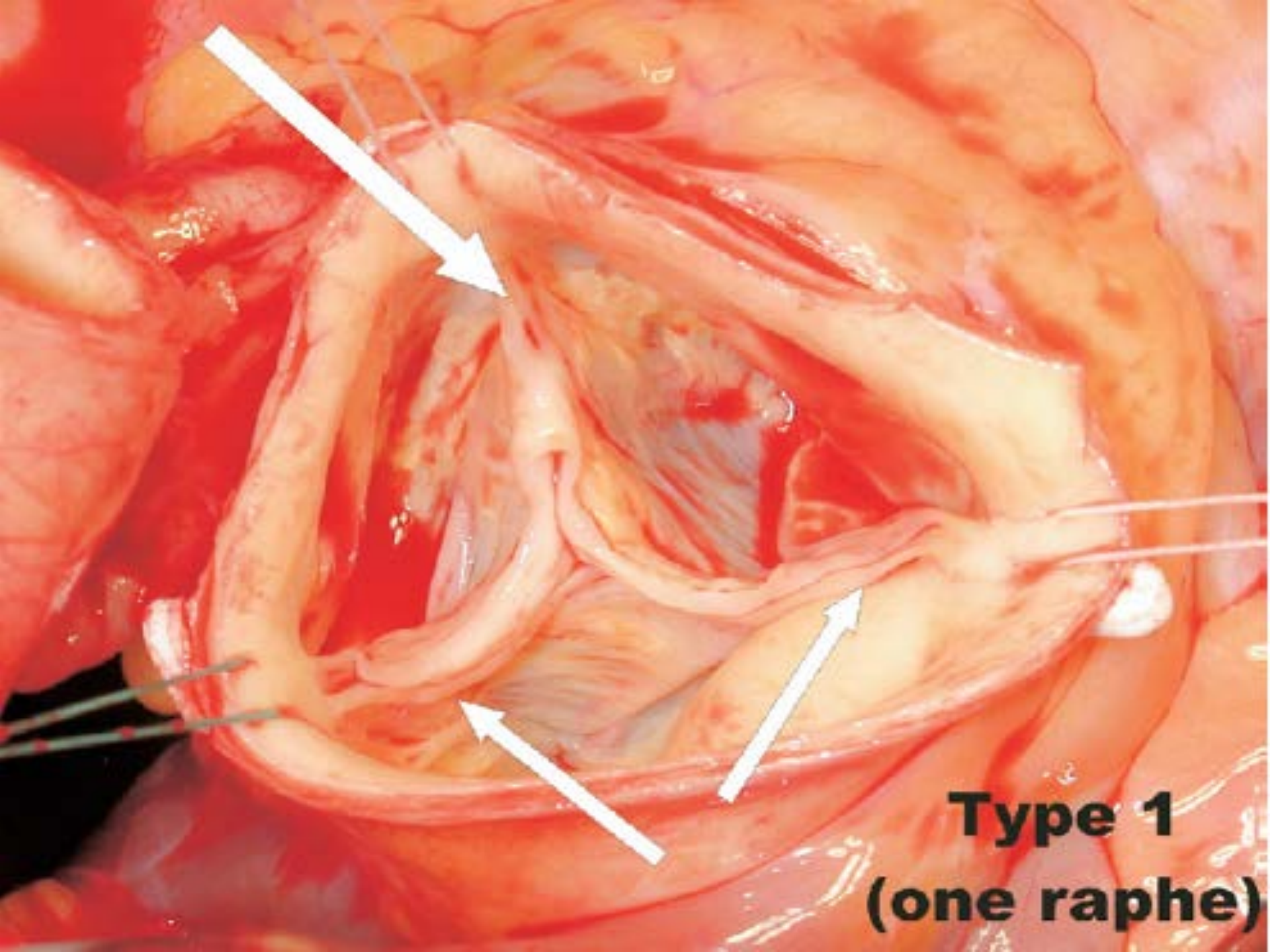


Bicuspidie aortique

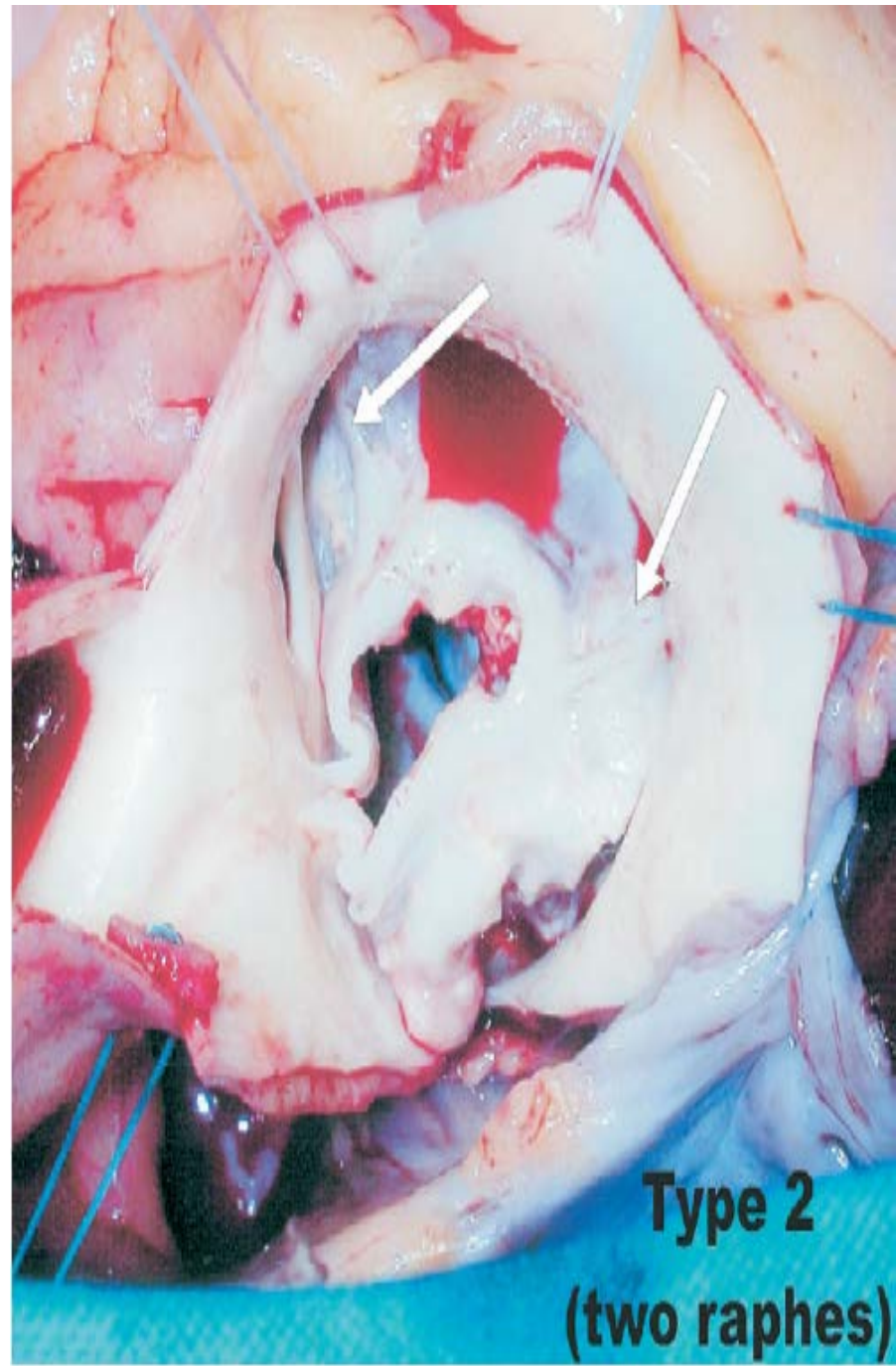
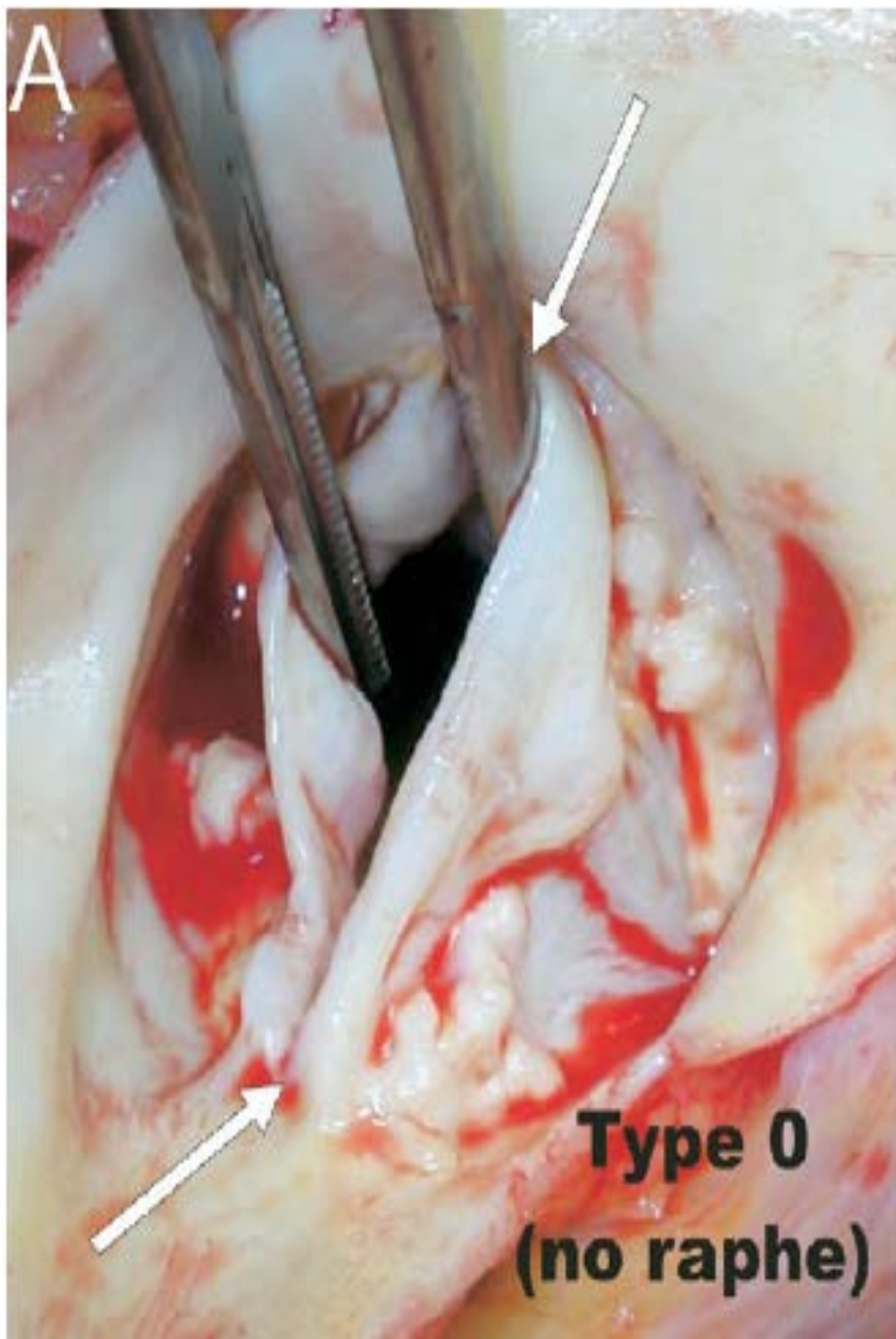
- incidence estimée 1%
- prévalence chez les NN : 0.46%
- forme familiale
- hérédité autosomique dominante pénétrance variable (prévalence intrafamiliale 37%)
- hétérogénéité génétique; NOTCH1
- risque accru d'obstacle G dans les apparentés de 1^{er} degré
- Dépistage echo familial

CLASSIFICATION MORPHOLOGIQUE

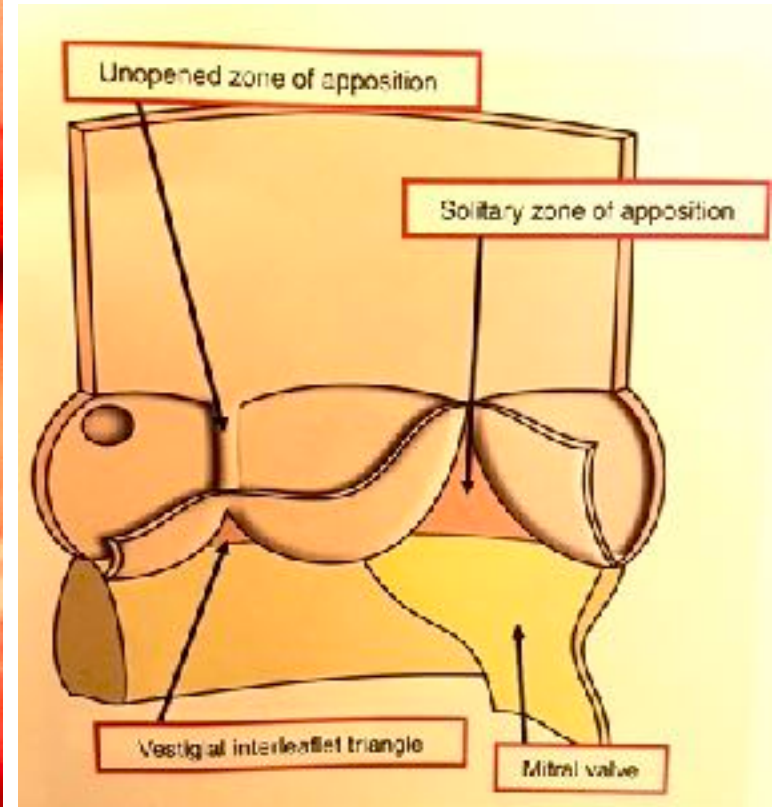
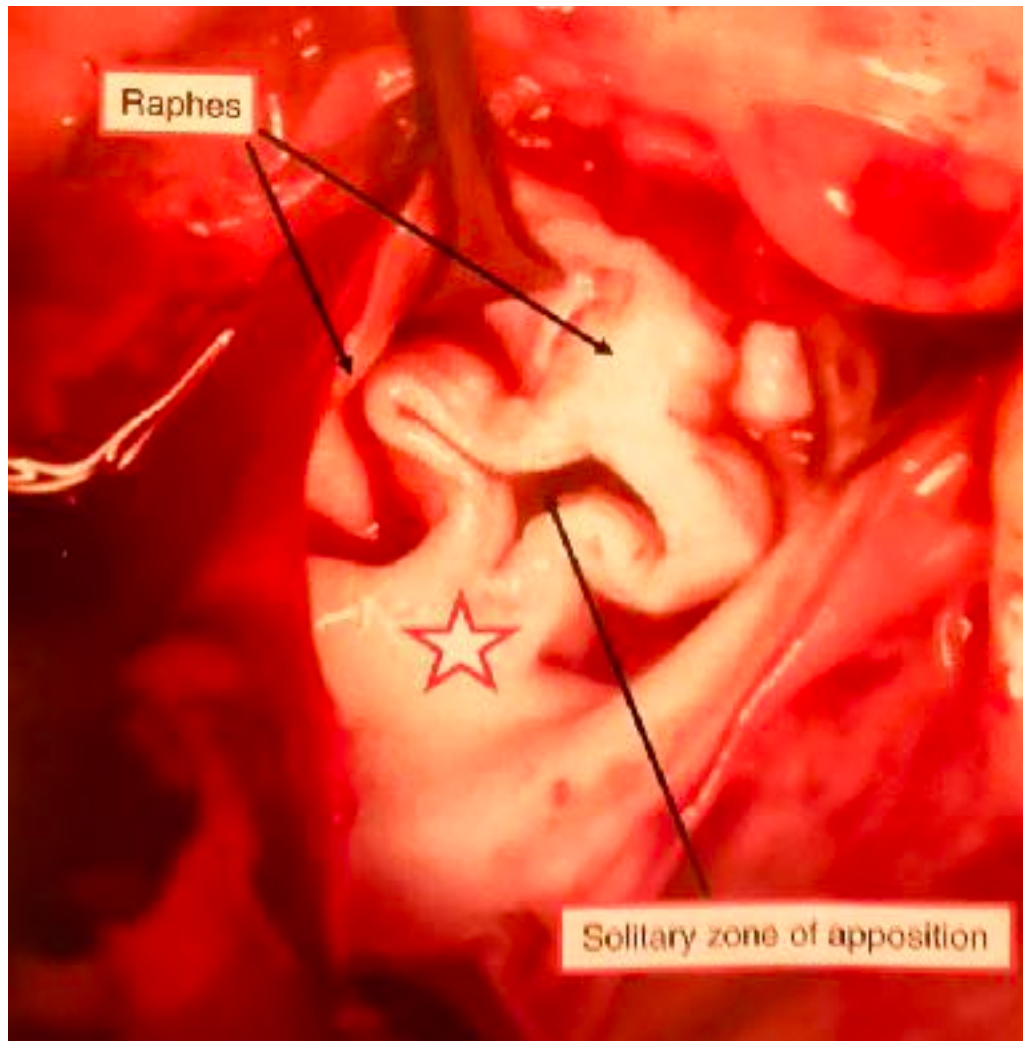
<p><u>main category:</u> number of raphes</p>	0 raphe - Type 0		1 raphe - Type 1			2 raphes - Type 2																			
	 <p>21 (7)</p>		 <p>269 (88)</p>			 <p>14 (5)</p>																			
<p><u>1. subcategory:</u> spatial position of cusps in Type 0 and raphes in Types 1 and 2</p>	<p>lat 13 (4)</p> 	<p>ap 7 (2)</p> 	<p>L - R 216 (71)</p> 	<p>R - N 45 (15)</p> 	<p>N - L 8 (3)</p> 	<p>L - R / R - N 14 (5)</p> 																			
<p><u>2. subcategory:</u></p> <table border="1" style="display: inline-table; vertical-align: middle;"> <tr> <td>V</td> <td>F</td> <td rowspan="5" style="vertical-align: middle;">B (I + S)</td> </tr> <tr> <td>A</td> <td>U</td> </tr> <tr> <td>L</td> <td>N</td> </tr> <tr> <td>V</td> <td>C</td> </tr> <tr> <td>U</td> <td>T</td> </tr> <tr> <td>L</td> <td>I</td> <td rowspan="2" style="vertical-align: middle;">No</td> </tr> <tr> <td>A</td> <td>O</td> </tr> <tr> <td>R</td> <td>N</td> <td></td> </tr> </table>	V	F	B (I + S)	A	U	L	N	V	C	U	T	L	I	No	A	O	R	N							
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A	U																								
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	I	6 (2)	1 (0.3)	79 (26)	22 (7)	3 (1)	6 (2)																		
	S	7 (2)	5 (2)	119 (39)	15 (5)	3 (1)	6 (2)																		
	No		1 (0.3)	15 (5)	7 (2)	2 (1)	2 (1)																		
	No			3 (1)	1 (0.3)																				



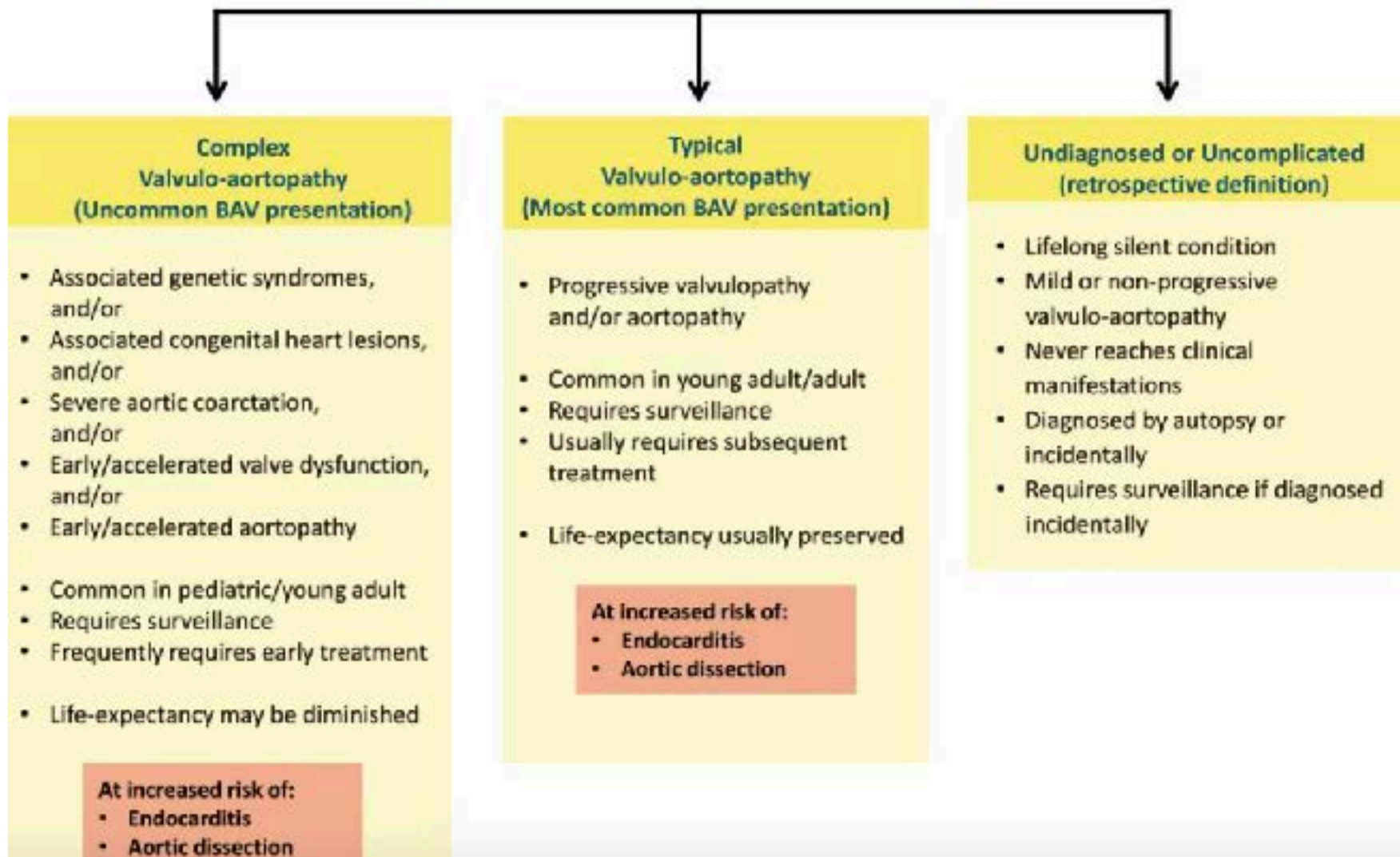
Type 1
(one raphe)



Sténose aortique : valve unicuspide



Bicuspid Aortic Valve Nosology



Michelena HI et al; International Consensus Statement on Nomenclature and Classification of the Congenital Bicuspid Aortic Valve and Its Aortopathy, for Clinical, Surgical, Interventional and Research Purposes. Radiol Cardiothorac Imaging. 2021 Jul 22;3(4):e200496.

Michelena HI et al; International Consensus Statement on Nomenclature and Classification of the Congenital Bicuspid Aortic Valve and Its Aortopathy, for Clinical, Surgical, Interventional and Research Purposes. Radiol Cardiothorac Imaging. 2021 Jul 22;3(4):e200496.

Bicuspid Aortic Valve Types and Phenotypes

Fused BAV (90-95%)

- 3 aortic sinuses
- 2 cusps: Usually different size/shape with asymmetric or symmetric non-fused commissural angle
- 2 commissures
- Raphe: Common, visible or not

Specific phenotypes of fused BAV

- Right-left cusp fusion (70-80%)
- Right-non cusp fusion (20-30%)
- Left-non cusp fusion (3-6%)
- Indeterminate cusp fusion

2-Sinus BAV (5-7%) (=Type 0)

- 2 aortic sinuses
- 2 cusps: Roughly same size/shape with symmetric non-fused commissural angle
- 2 commissures
- Raphe: No

Specific phenotypes of 2-sinus BAV

- Latero-lateral (most common)
- Anterior-posterior

Partial-fusion BAV (%?) (forme fruste)

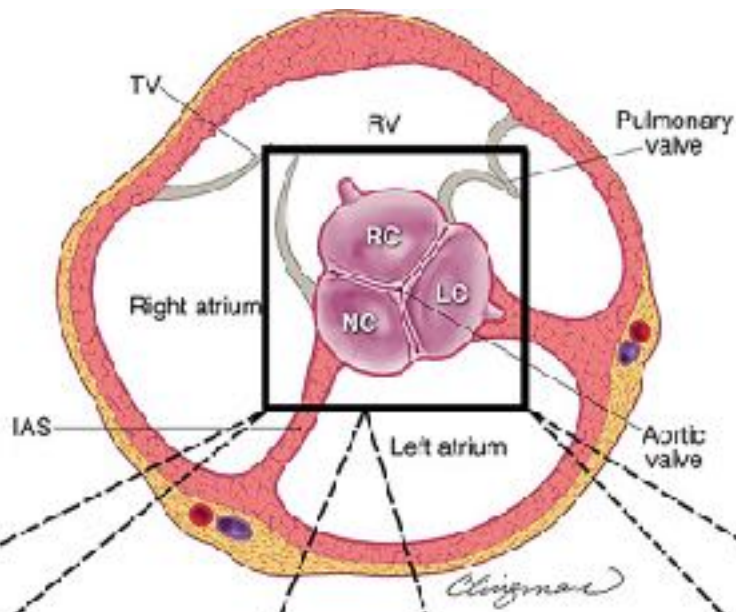
- 3 aortic sinuses
- 3 cusps: Usually symmetric
- 3 "apparent" commissures where 2 are normal and the third is fused <50%
- Raphe: Small, mini-raphe

Specific phenotype of partial-fusion BAV

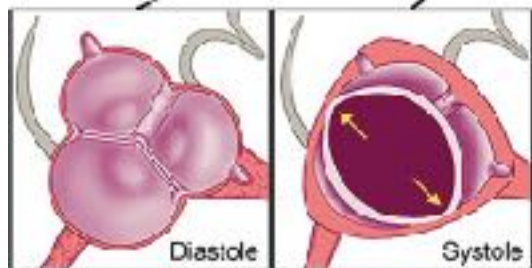
- Partial (<50%) fusion of one commissure

= Type 1

Fused BAV
(90-95% of BAV)
3 Phenotypes

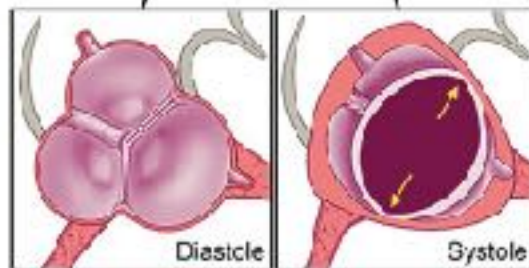


1.



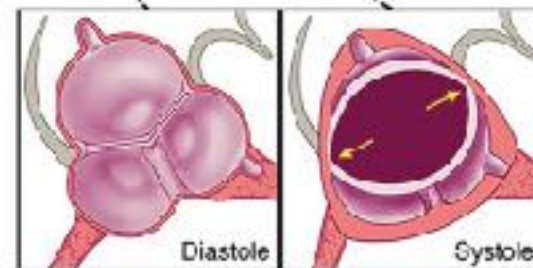
Right – Left Cusp Fusion
(70-80%)

2.



Right – Non Cusp Fusion
(20-30%)

3.



Left – Non Cusp Fusion
(3-6%)



ACE
FPS: 100/
T: 1.7 MHz/3 MHz

Sharp



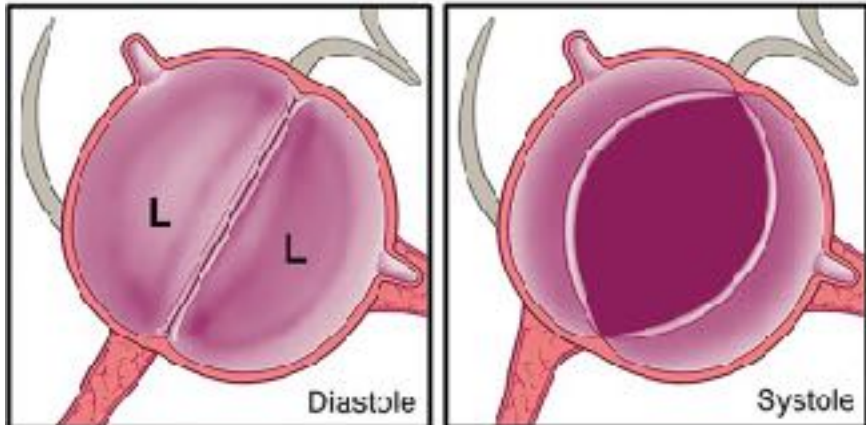
70
HR

2-Sinus BAV

(5-7% of BAV)

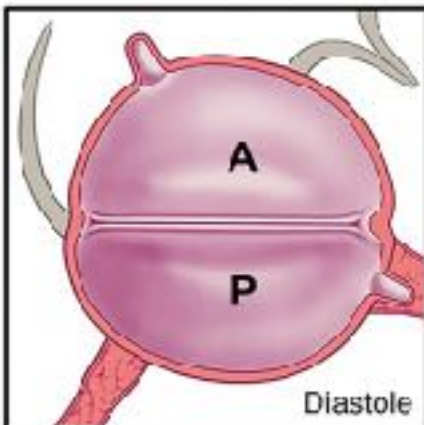
2 Phenotypes

1.



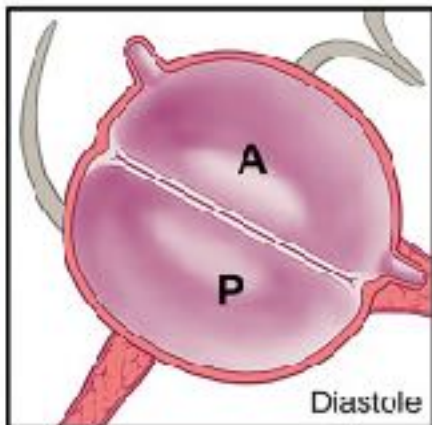
Latero-lateral
(most common)

2.A



Anteroposterior
(least common)

2.B

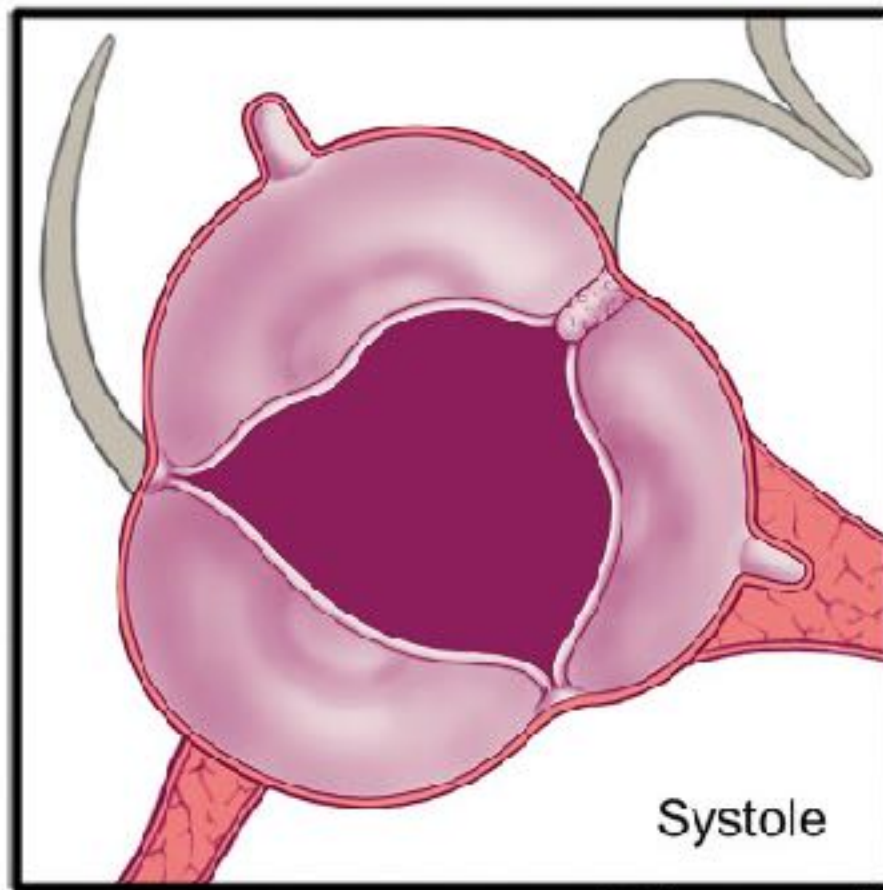
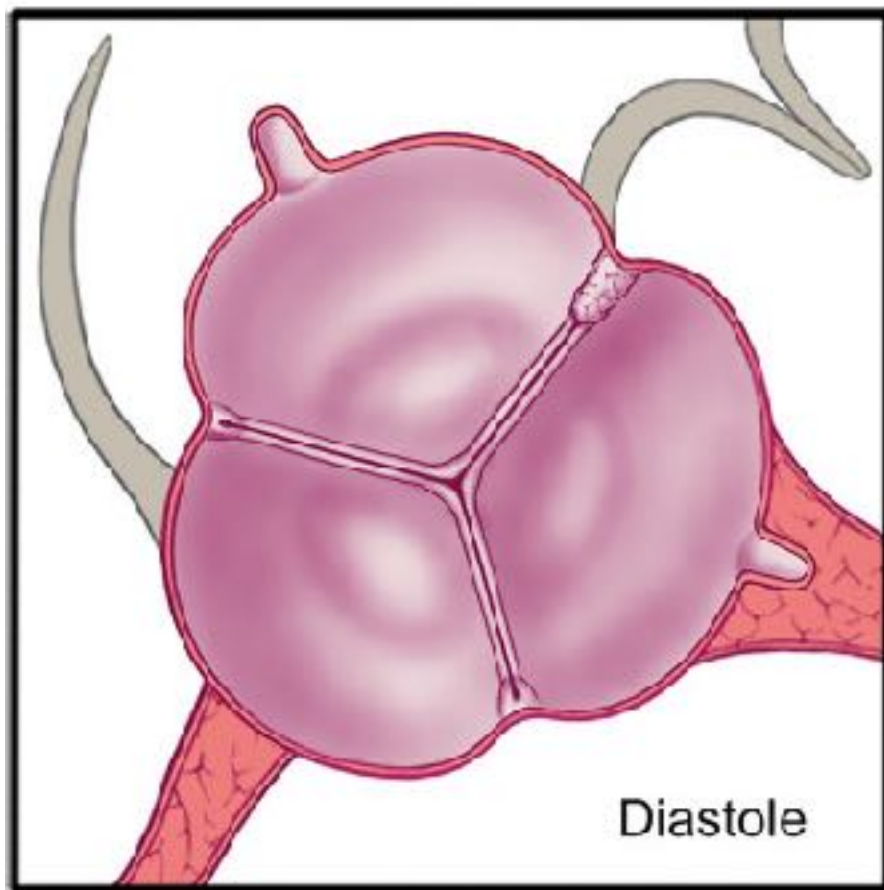


Chigman

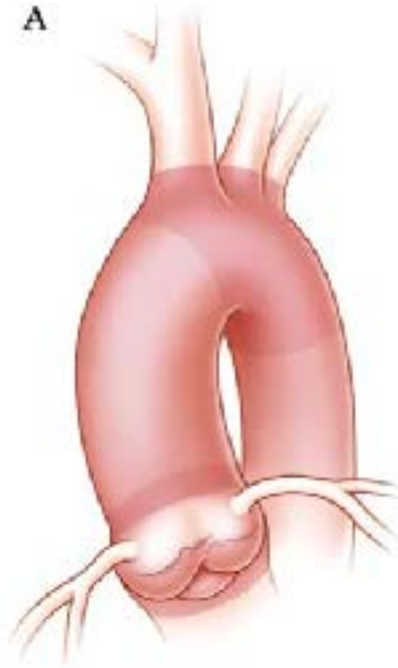
Partial-Fusion BAV

(Forme Fruste)

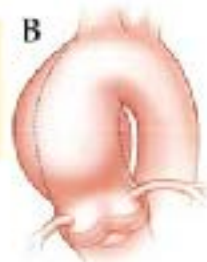
Short fusion of 1 commissure



Chigman



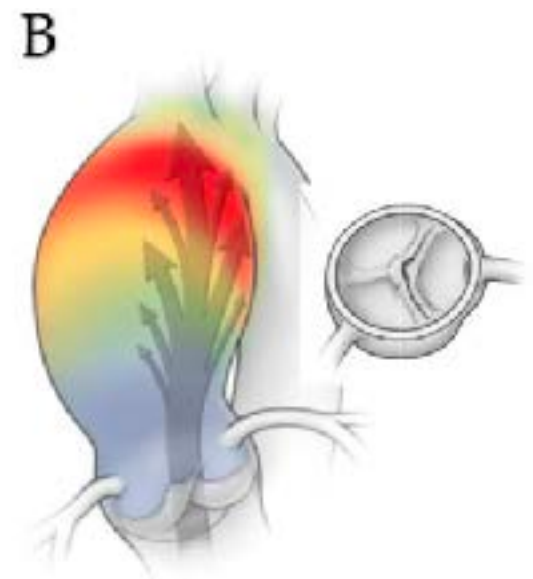
Type 1
Dilatation of tubular ascending aorta primarily along coronary of aorta, with mild-to-moderate



Type 2
Aortic dilatation with involvement of tubular ascending aorta, with relative sparing of root



Type 3
Isolated aortic root involvement with normal tubular ascending aorta and arch dilatation



Sténose valvulaire aortique

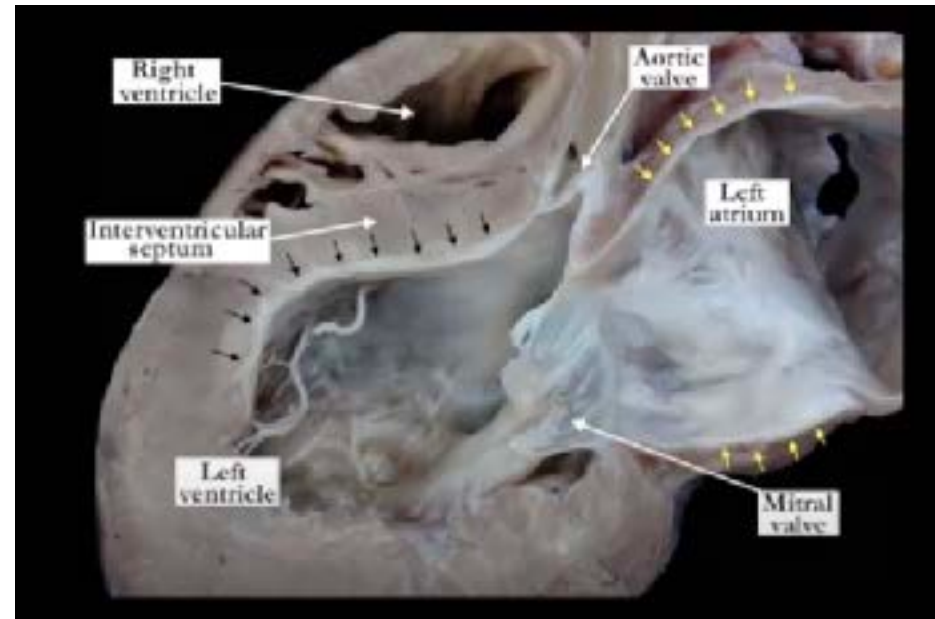
Formes cliniques pédiatriques

- sténose aortique néonatale critique
 - dg foetal

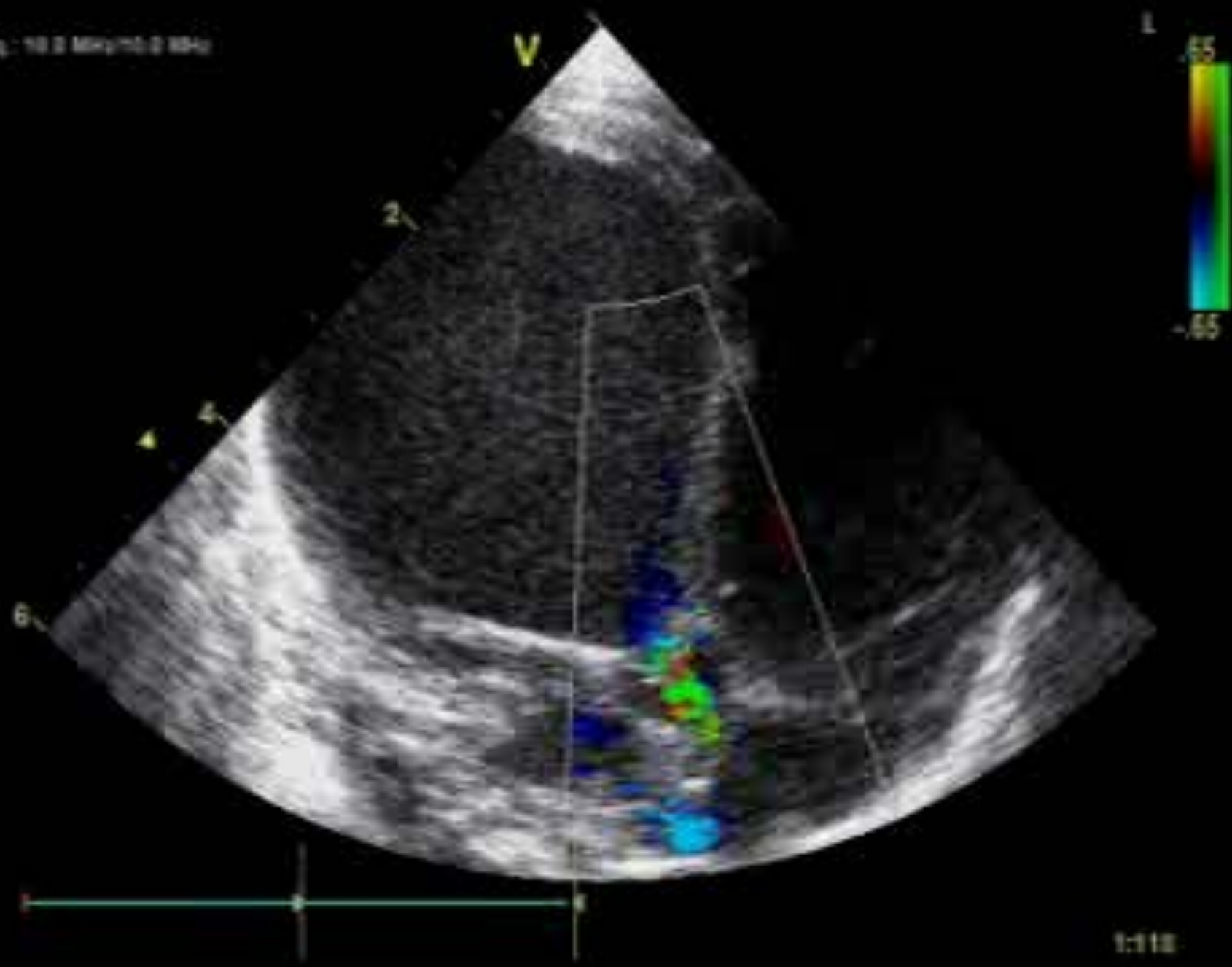
- sténose aortique non critique

sténose valvulaire aortique critique néonatale

- insuffisance cardiaque néonatale + cyanose MI (CA systémique)
- souvent valve monocuspide
- VG dilaté fibroelastotique
- fonction VG, pronostic?
- ductodépendance > prostine
- stratégie uni ou biventriculaire

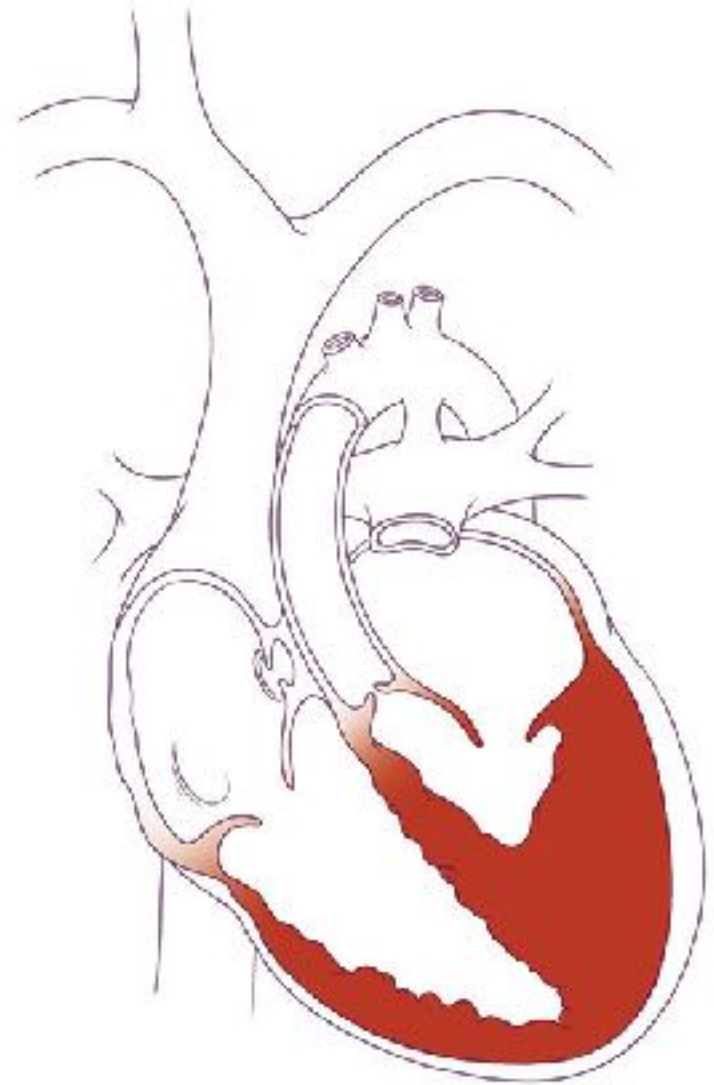
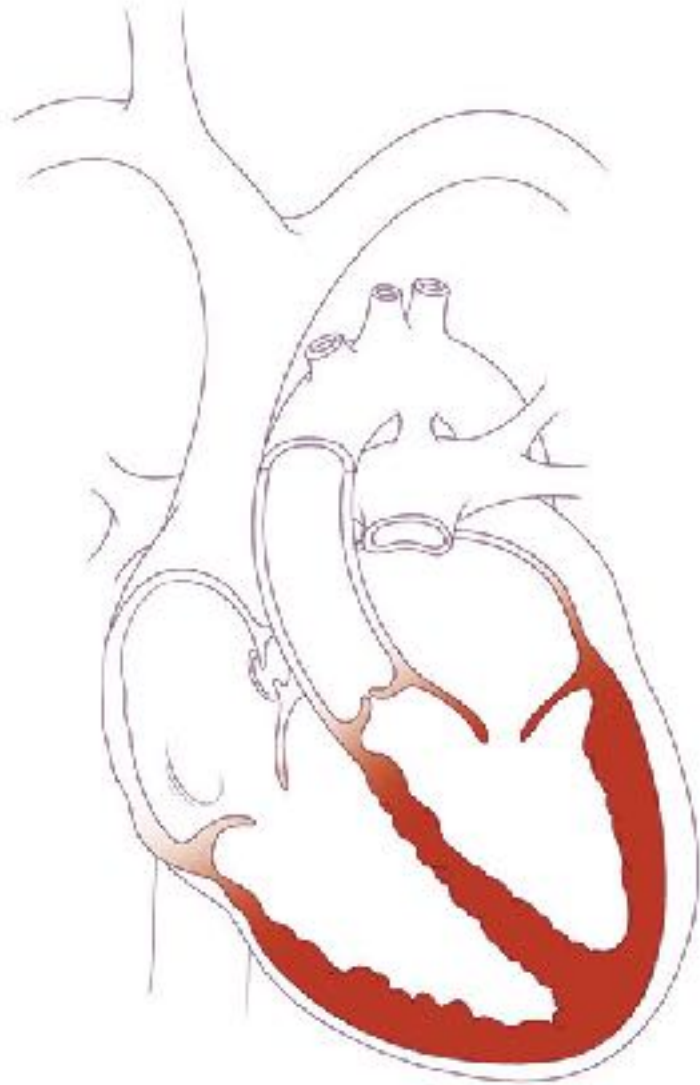


Freq.: 10.0 MHz/10.0 MHz



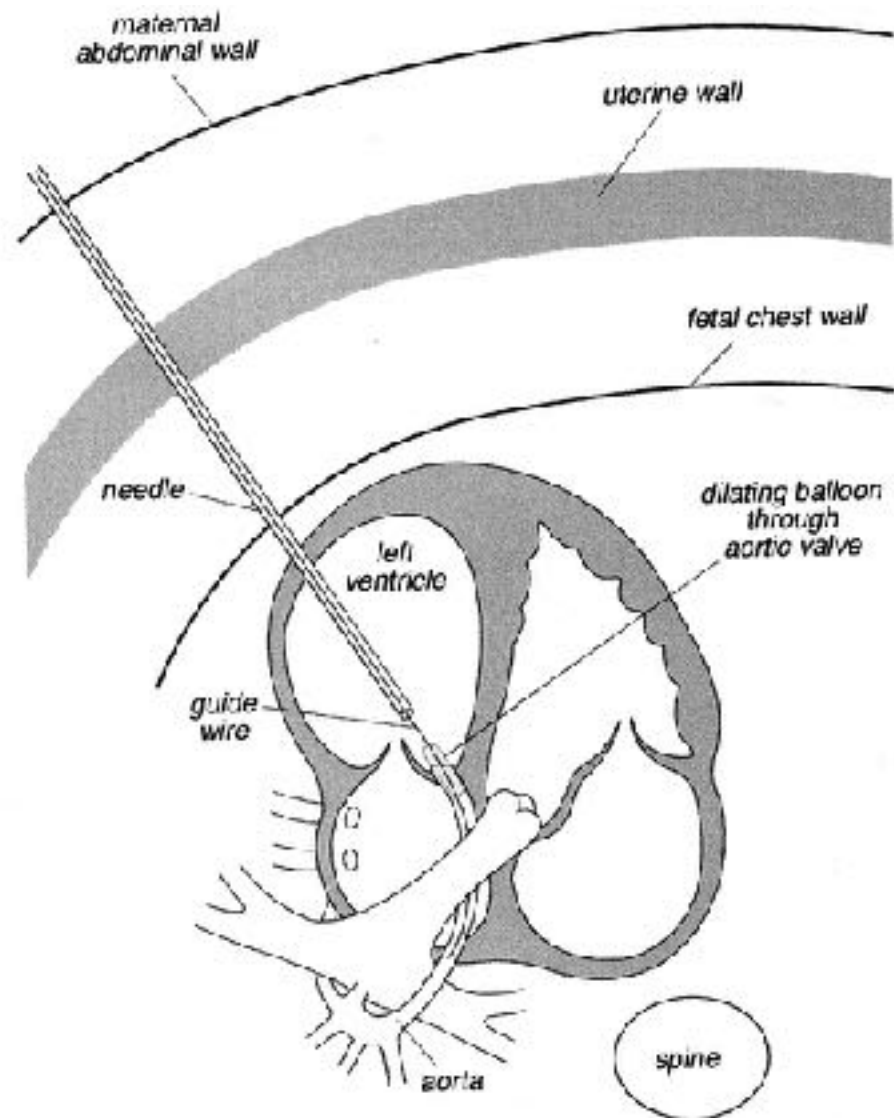
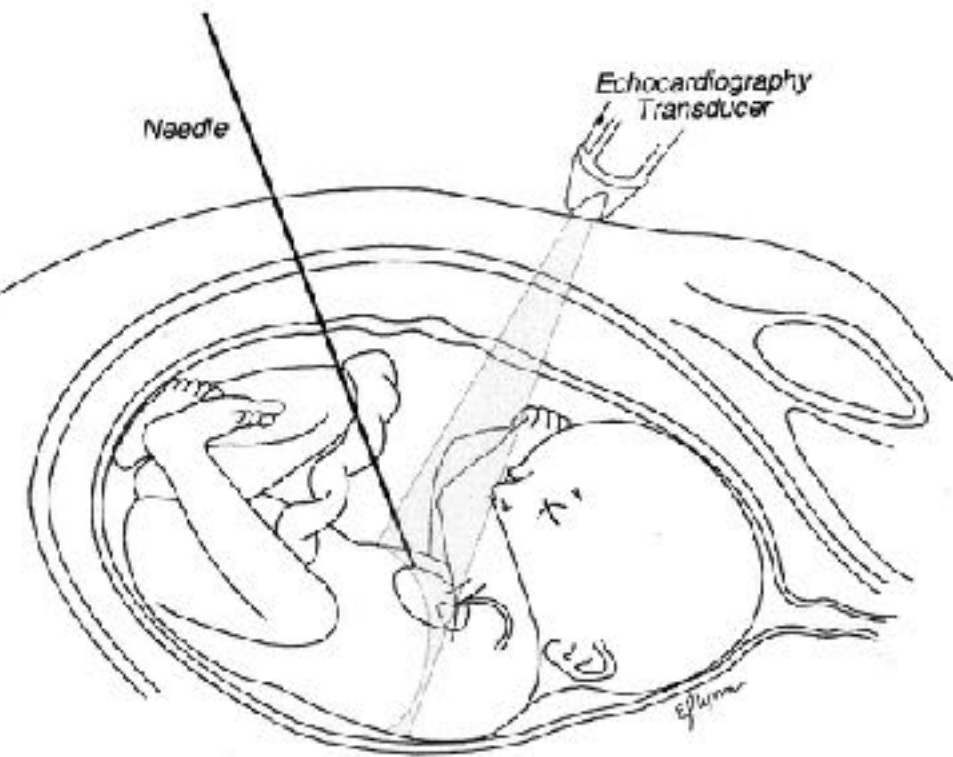
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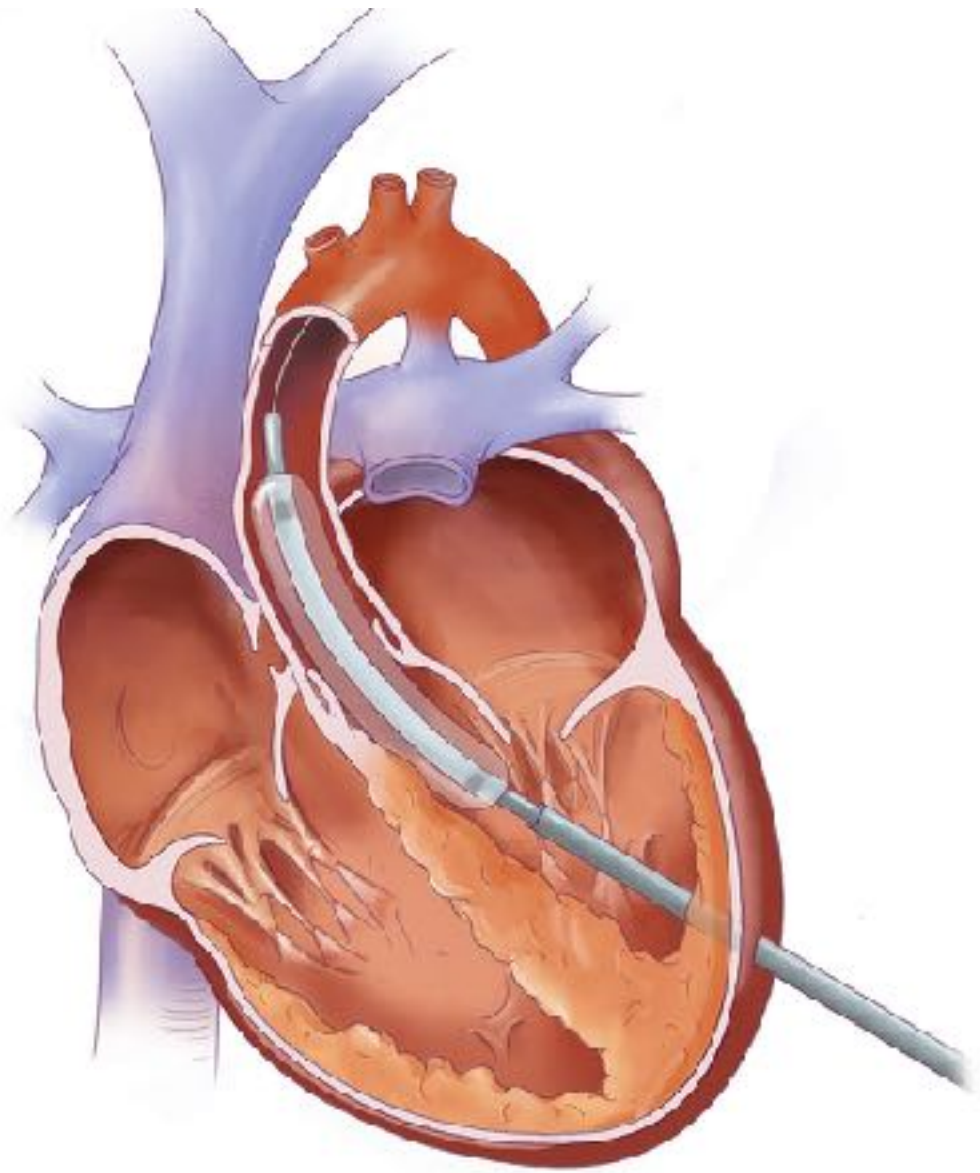
Forme foetale



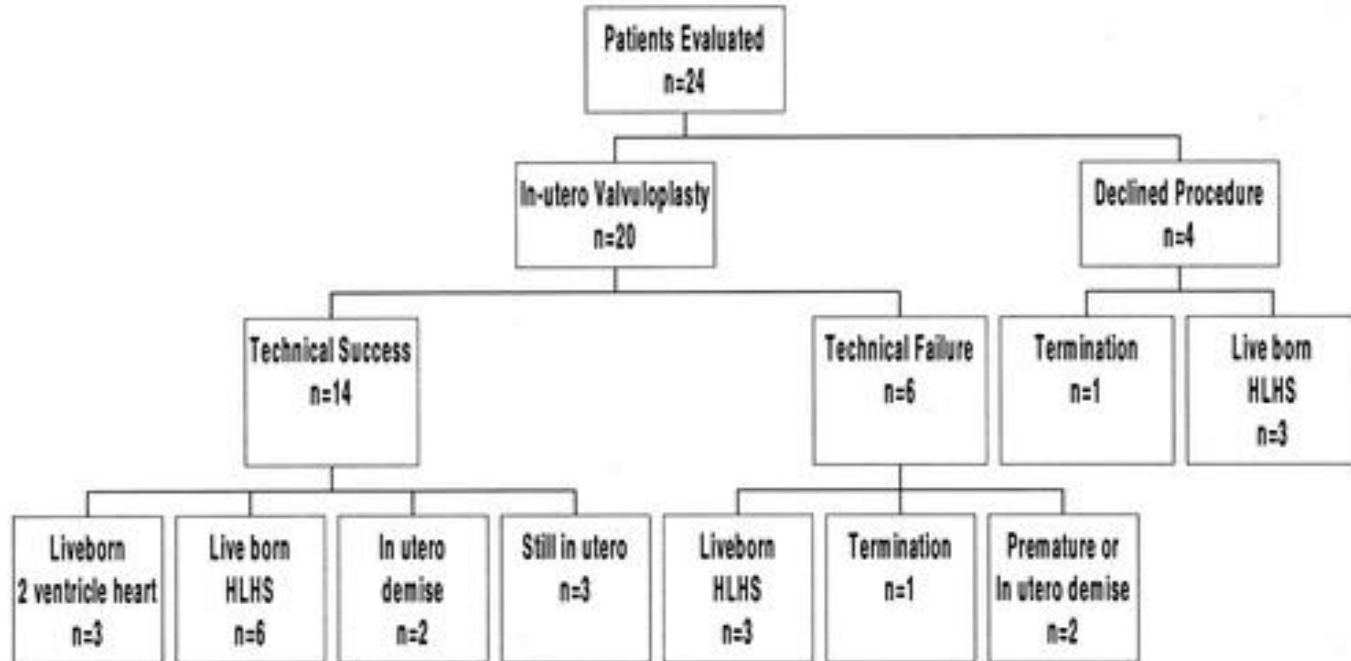
Dilatation valvulaire aortique foetale

- Arrêt de la progression de l'hypoplasie du VG
- Restaurer la fonction VG
- Améliorer la croissance des structures du coeur gauche
- Objectif : pouvoir envisager une stratégie biventriculaire à la naissance
- Succès si dilatation valvulaire possible et amélioration du flux antérograde





Flow Chart of All Patients Evaluated for Fetal Aortic Valvuloplasty



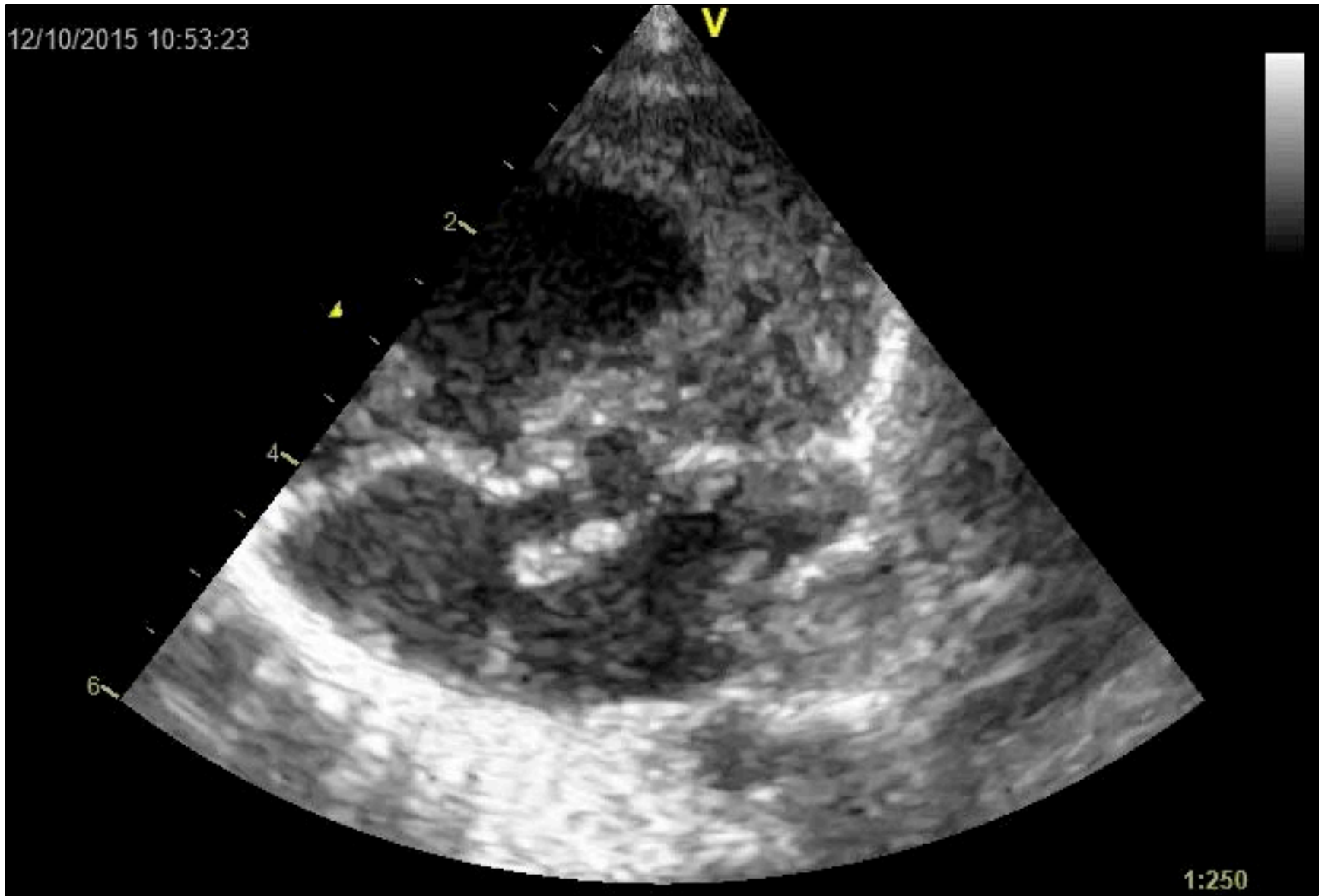
Wayne Tworetzky. Circulation 2004. Balloon Dilation of Severe Aortic Stenosis in the Fetus, Volume: 110, Issue: 15, Pages: 2125-2131. (Boston)

sténose aortique néonatale non critique

Lésions valvulaire

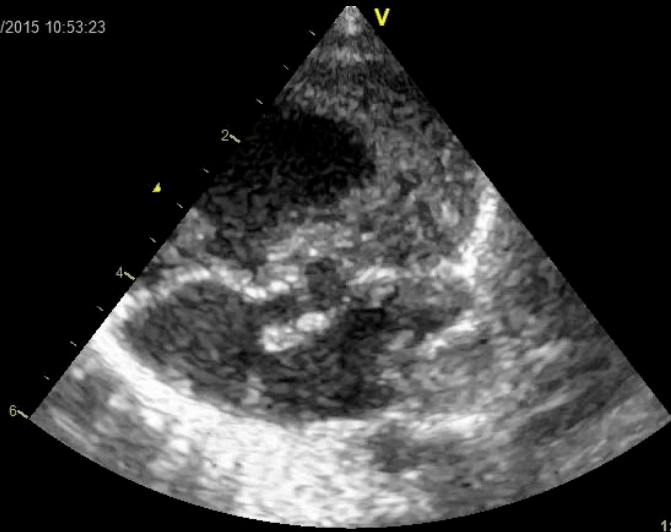
- valve bicuspidé ou monocuspide
 - rarement tricuspide
 - une seule commissure normale
 - feuillets dysplasiques et épais
 - tissu nodulaire fibreux
 - taille de l'anneau : +/- hypoplasique
-
- Lésions associées : obtacle ss ao, supravalulaire ao, dilatation de l'aorte ascendante anomalie mitrale, Coa

12/10/2015 10:53:23

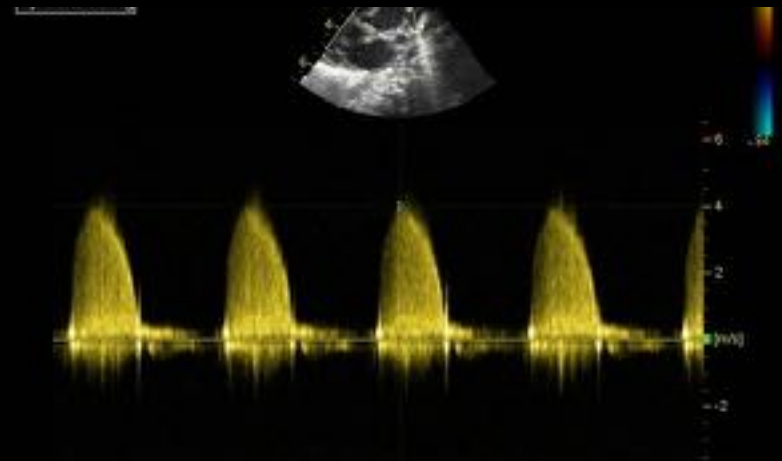
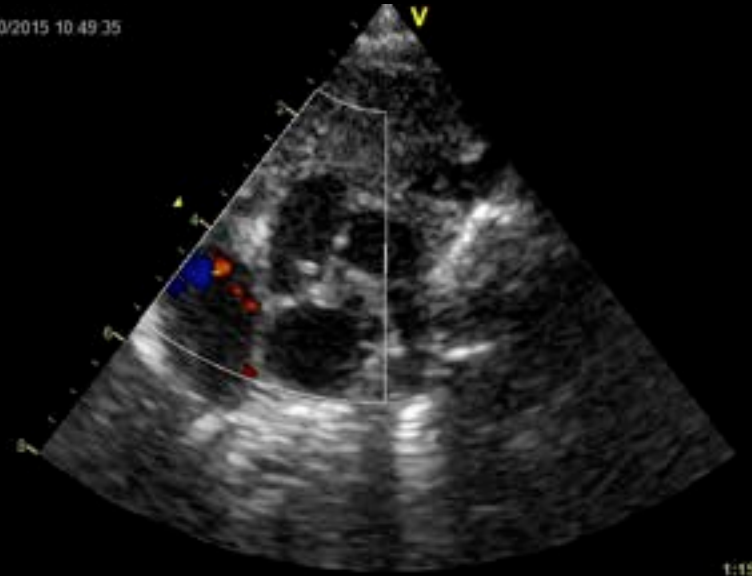


1:250

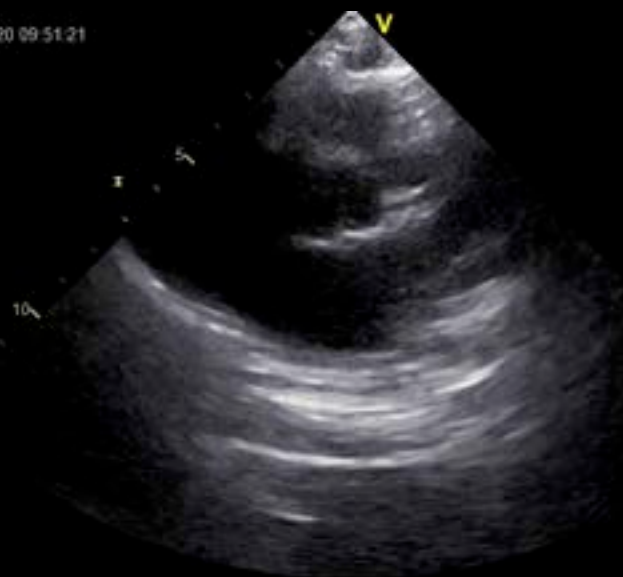
12/10/2015 10:53:23



12/10/2015 10:49:35



16/09/2020 09:51:21



CTO

16/09/2020 09:46:37



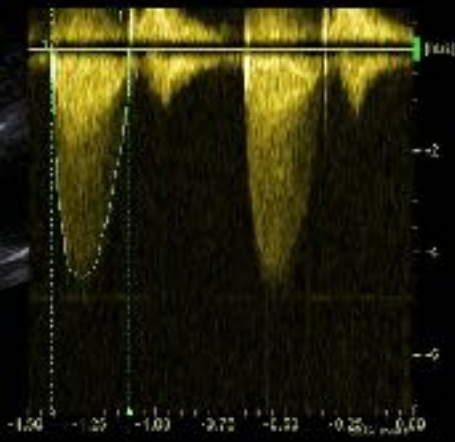
CTO

1:148

1:265

66.5	
VA Vmax	1.97 m/s
VA Vmoy	3.35 m/s
VA QDmax	80.83 mmHg
VA QDmoy	50.18 mmHg
VA Ery.Ts	391 ms
VA ITV	103.78 cm
FC	109.57 BPM

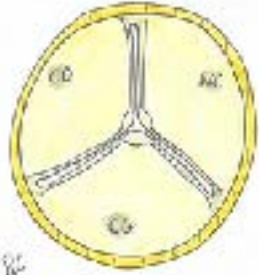
CTO



Traitement chirurgical

- commissurotomie extensive (bicuspidie)
- « peler » les feuillets valvulaires
- excision des nodules fibreux
- reconstruction commissurale
- élargissement du culot

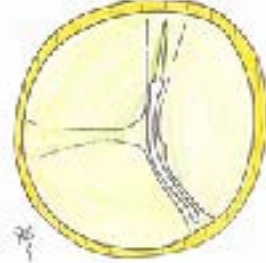
- ligature du canal artériel
- fermeture totale ou partielle CIA
- plastie aortique si coa



卷



卷



卷



卷

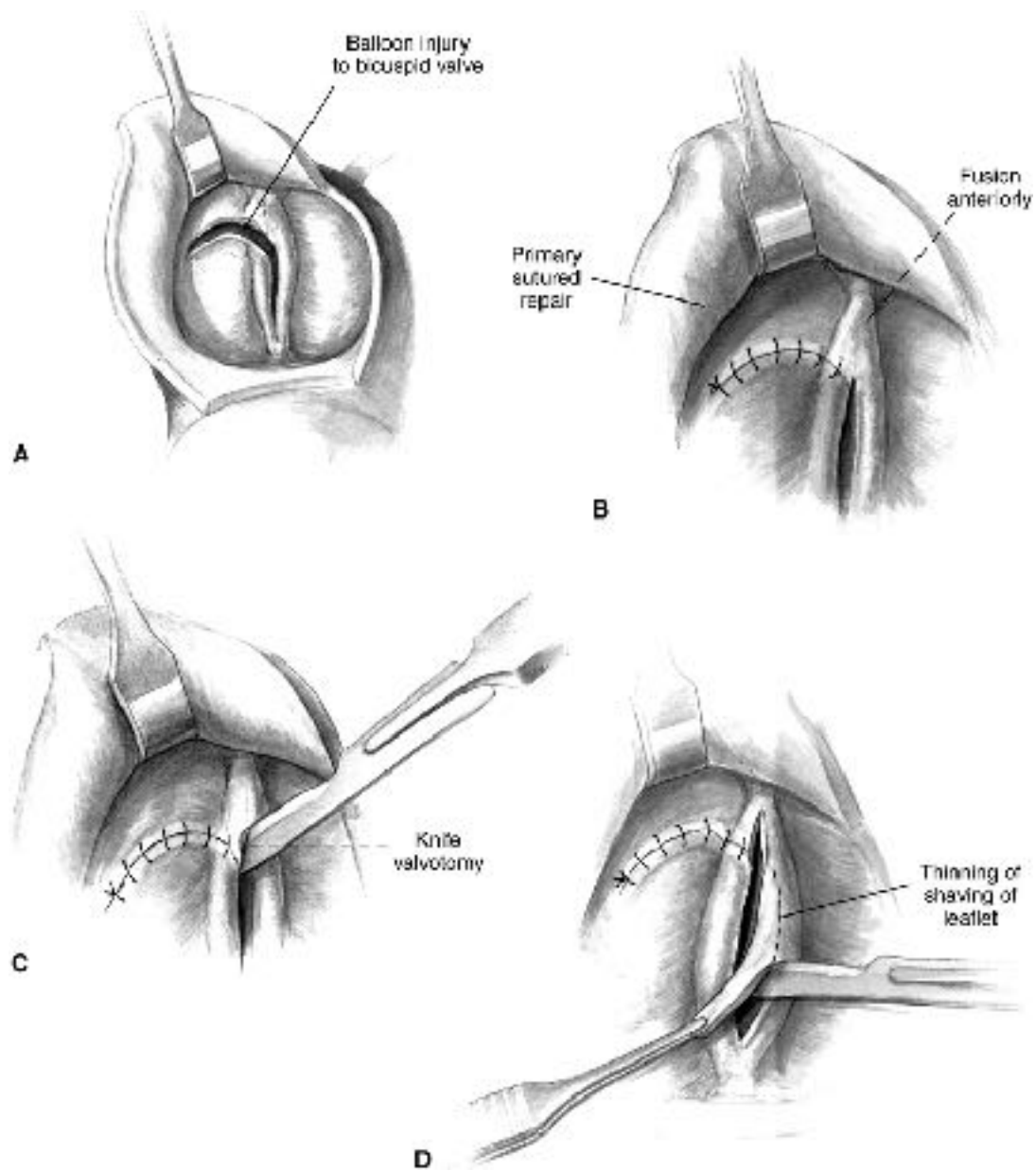


卷



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Indications

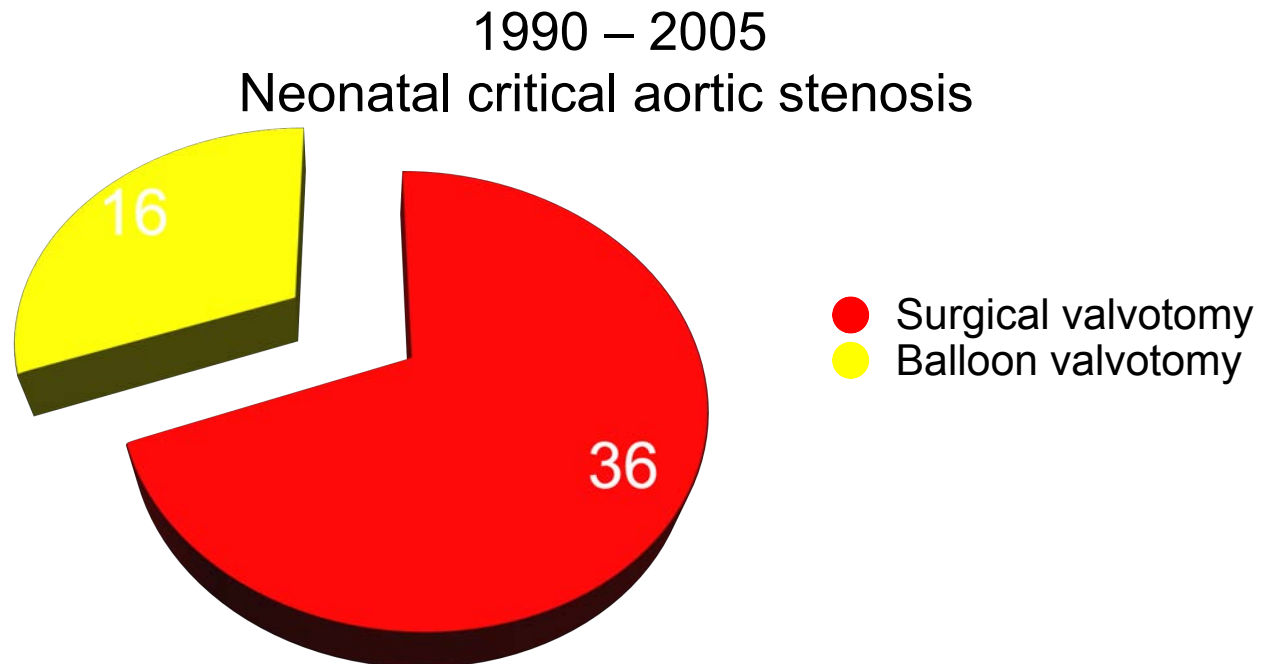
- VG normal (taille et fonction)
 - valvuloplastie chirurgicale : excellents résultats
diminution grad moy, amelioration fonction VG, peu de IAO
 - dilatation percutanée: non
- dysfonction VG
 - dilatation valvulaire percutanée
 - puis valvuloplastie ou Ross Konno

Neonatal Surgical Aortic Commissurotomy: Predictors of Outcome and Long-Term Results

Gabriella Agnoletti, MD, PhD, Olivier Raisky, MD, Younes Boudjemline, MD, PhD,
Phalla Ou, MD, Damien Bonnet, MD, PhD, Daniel Sidi, MD, PhD, and
Pascal Vouhé, MD, EBTCs

Pediatric Cardiology and Cardiac Surgery, Necker-Enfants Malades, Paris, France

Ann Thorac Surg 2006;82:1585-1593



Balloon Valvotomy

- High mortality 9/16 patients (56%) =>
 - left ventricular failure
 - persistent pulmonary hypertension
 - mitral dysfunction
- Moderate to severe aortic regurgitation: 7 patients (44%)
- 5 Early re-operation: surg. valvoplasty, 2 rescue Ross, 1 coarctation => 4 deaths
- late redo-operation: 3 patients out the 7 surviving
 - (ross konno, valvotomy, mitral replacement) =>no death

Indications

Chirurgie

- valve dysplasique
- feuillets épais
- nodules fibreux
- valve monocuspide
- obstacles LVOT associées

Valvuloplastie percutanée

- dysfonction VG
- VG fibroelastotique
- resténose post chirurgicale tardive

*Alexiou et al, Eur J Cardiothorac Surg, 2001.
Bhabra MS et al, Ann Thorac Surg, 2003.*

Traitement des sténoses aortiques

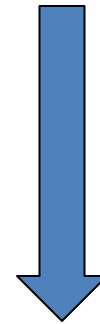
- Commissurotomie chirurgicale
- Valvuloplastie percutanée
- Remplacement valvulaire aortique
- Opération de Ross

Remplacement valvulaire aortique chez l'enfant



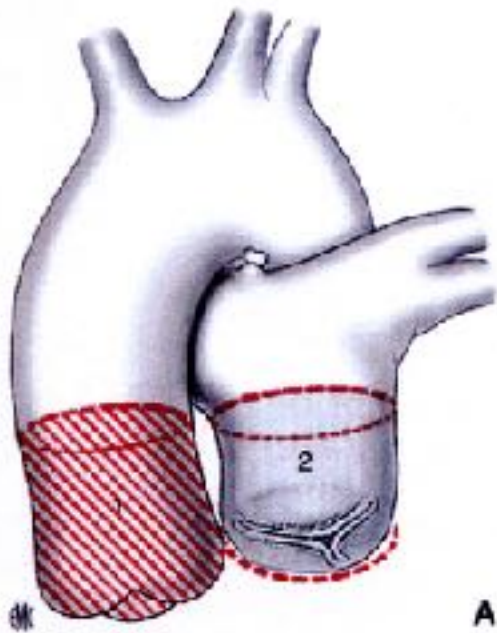
Valve mécanique (taille définitive)

2 Stratégies finales

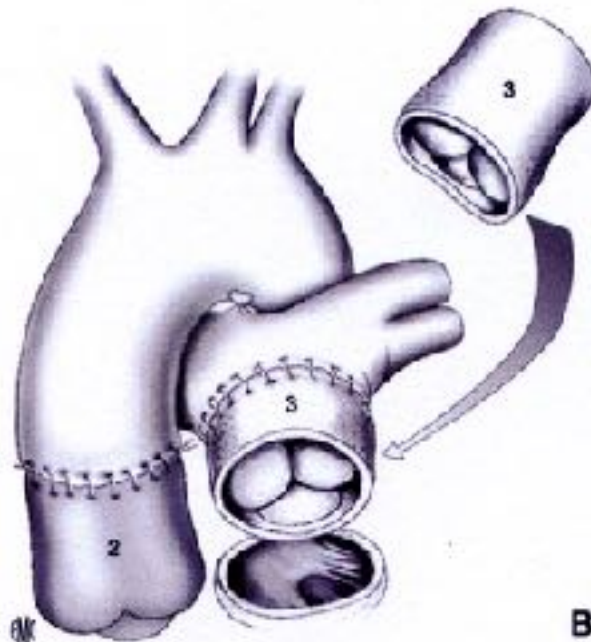


- Autogreffe pulmonaire Ross et Ross-Konno

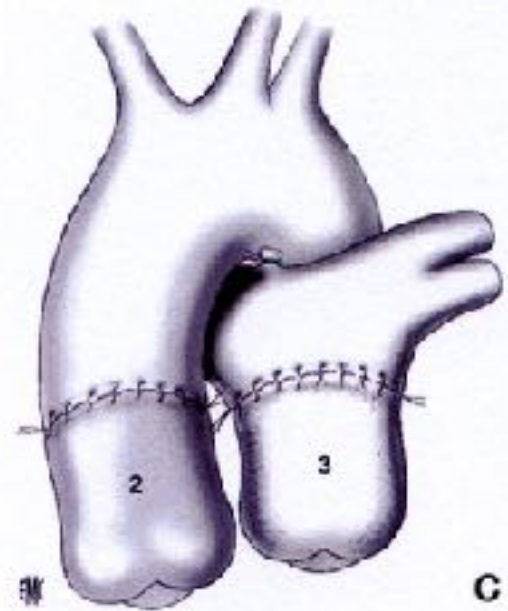
Ross / Ross-Konno chez l'enfant



A



B



C

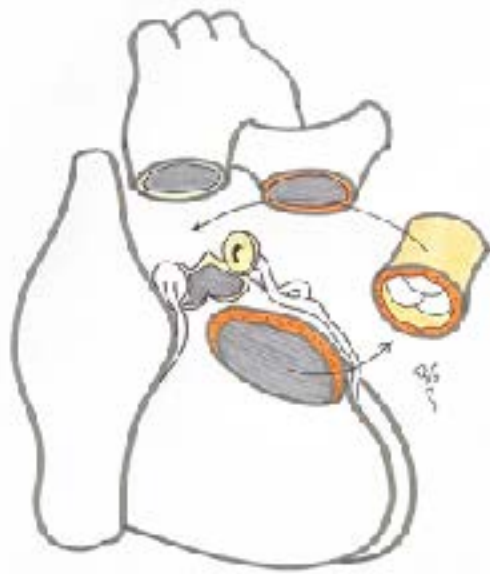
Ross / Ross-Konno chez l'enfant

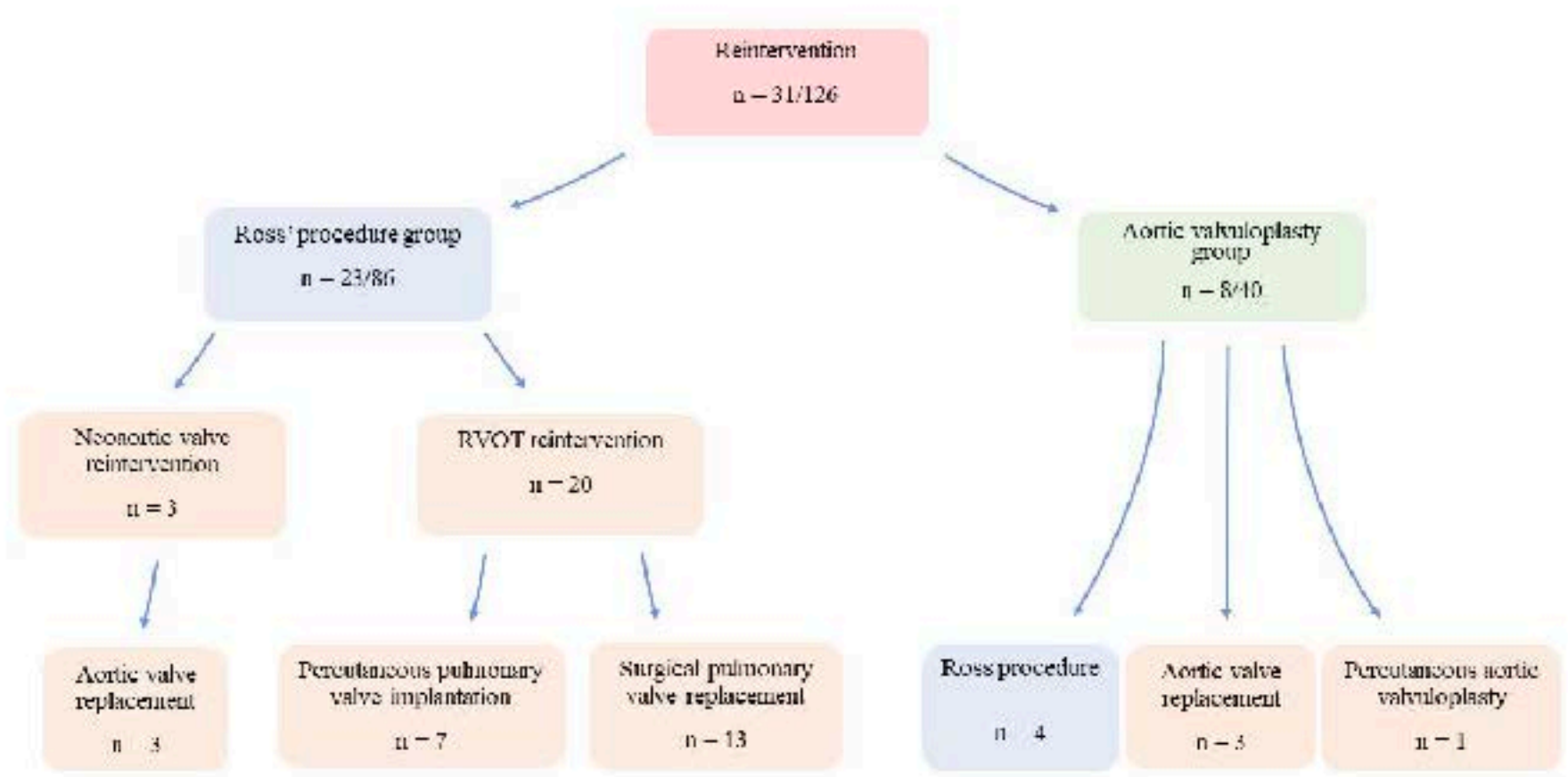
Indications

- Obstacles complexes voie gauche
(Shone, malformation mitrale, bourrelet septal, ...)
- sténose aortique congénitale
(post - dilatation)
- insuffisance aortique congénitale

Reconstruction voie droite

- reconstruction orifice ventriculaire droit
- options
 - . Homogreffe pulmonaire
 - . Hétérogreffe
 - . Monocusp
 - . Double autogreffe



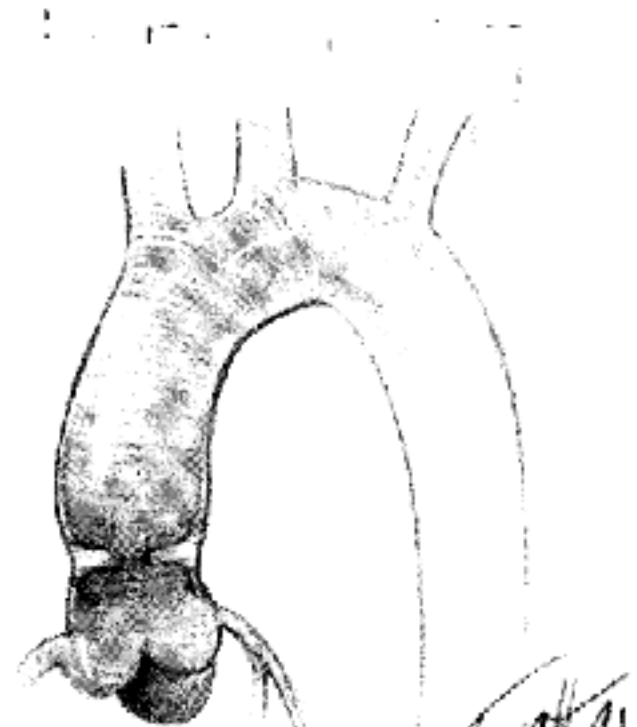
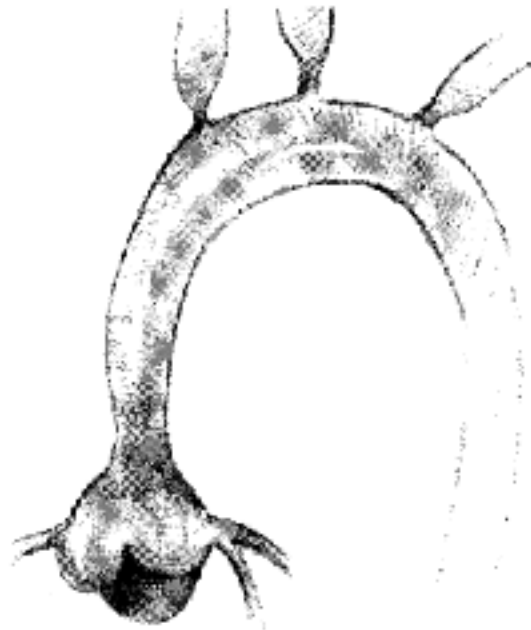


Ross procedure or complex aortic valve repair using the pericardium in children: a real dilemma
 Danial *et al*; JTCS 2021

Sténose supravalvulaire aortique

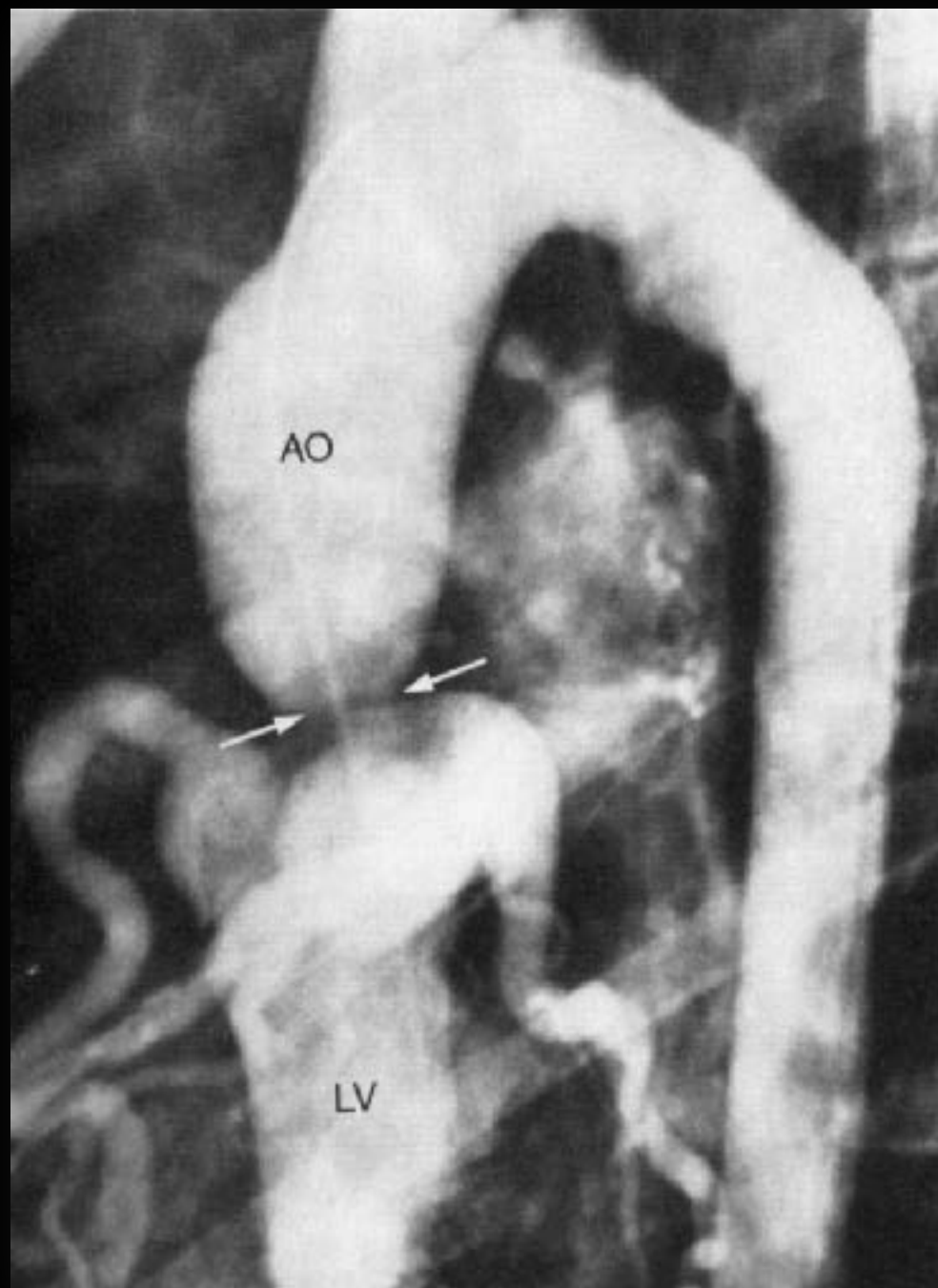
Sténose supravalvulaire aortique

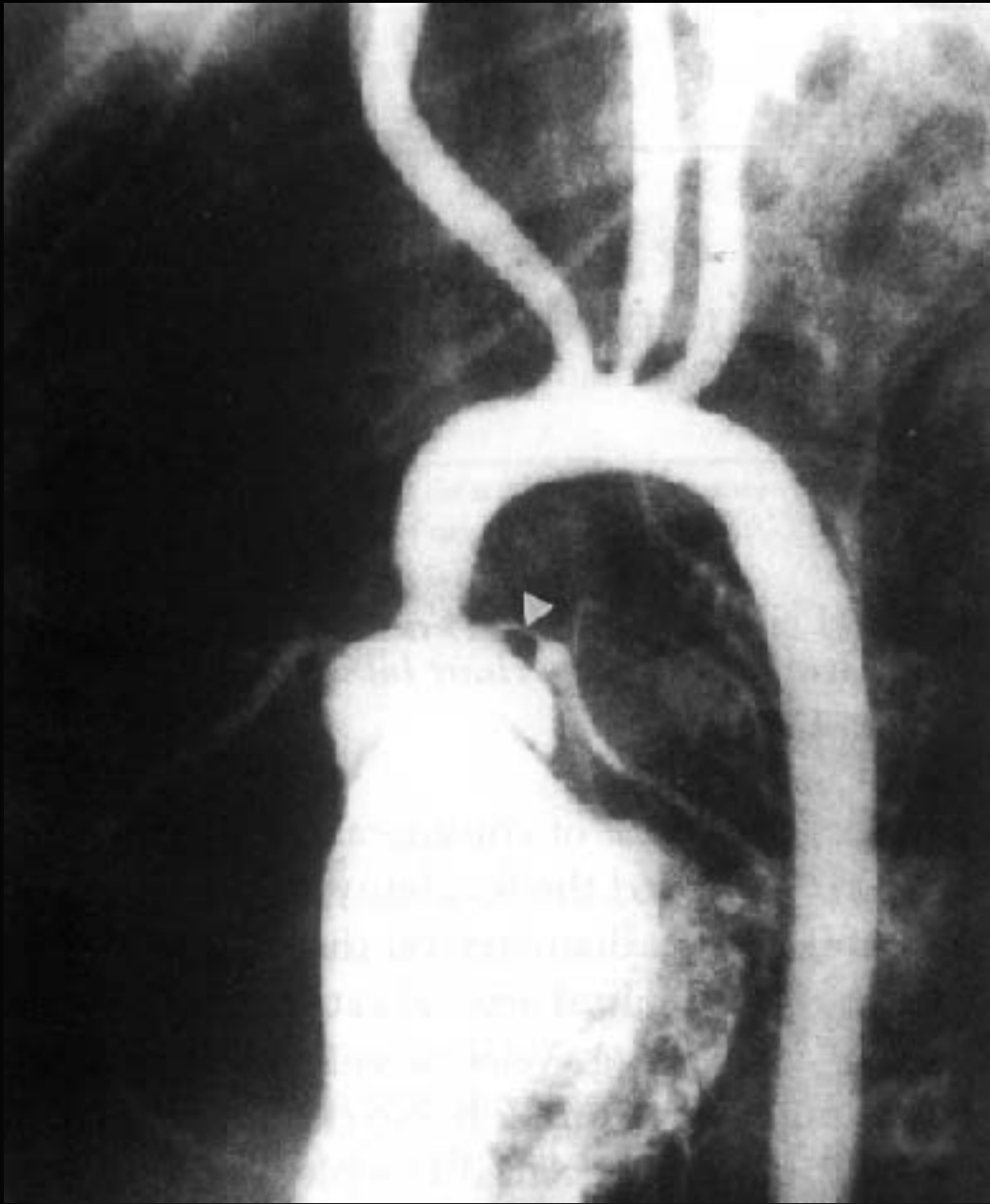
- plus rare
- lésions focales (JST) ou diffuses



- sd de Williams Beuren
- del 7q11.23
- mutation gène élastine
- réduction et désorganisation des fibres élastiques dans la media aortique: perte d'élasticité paroi, hypertrophie du muscle lisse et dépôt collagène (aspect feuilleté)

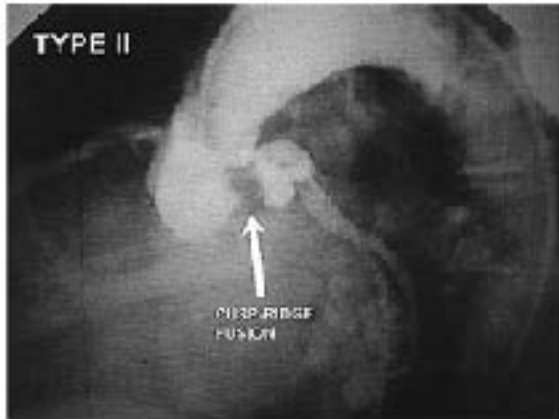
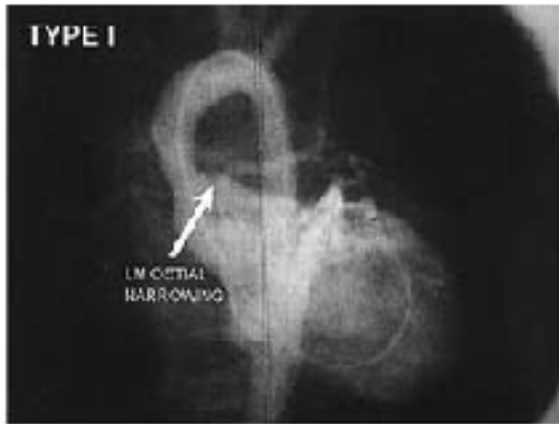






- lésions associées :
 - bicuspidie aortique
 - sténose ostiale coronaire
 - sténose distale des artères pulmonaires
 - sténose des troncs supra aortiques, artères rénales, artères mésentériques

Sténose ostiale coronaire



Type I



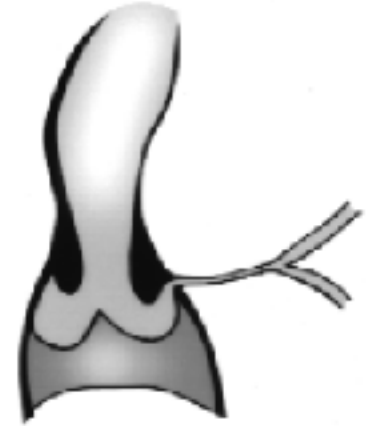
Ostial
Narrowing

Type II



Cusp-Ridge
Fusion

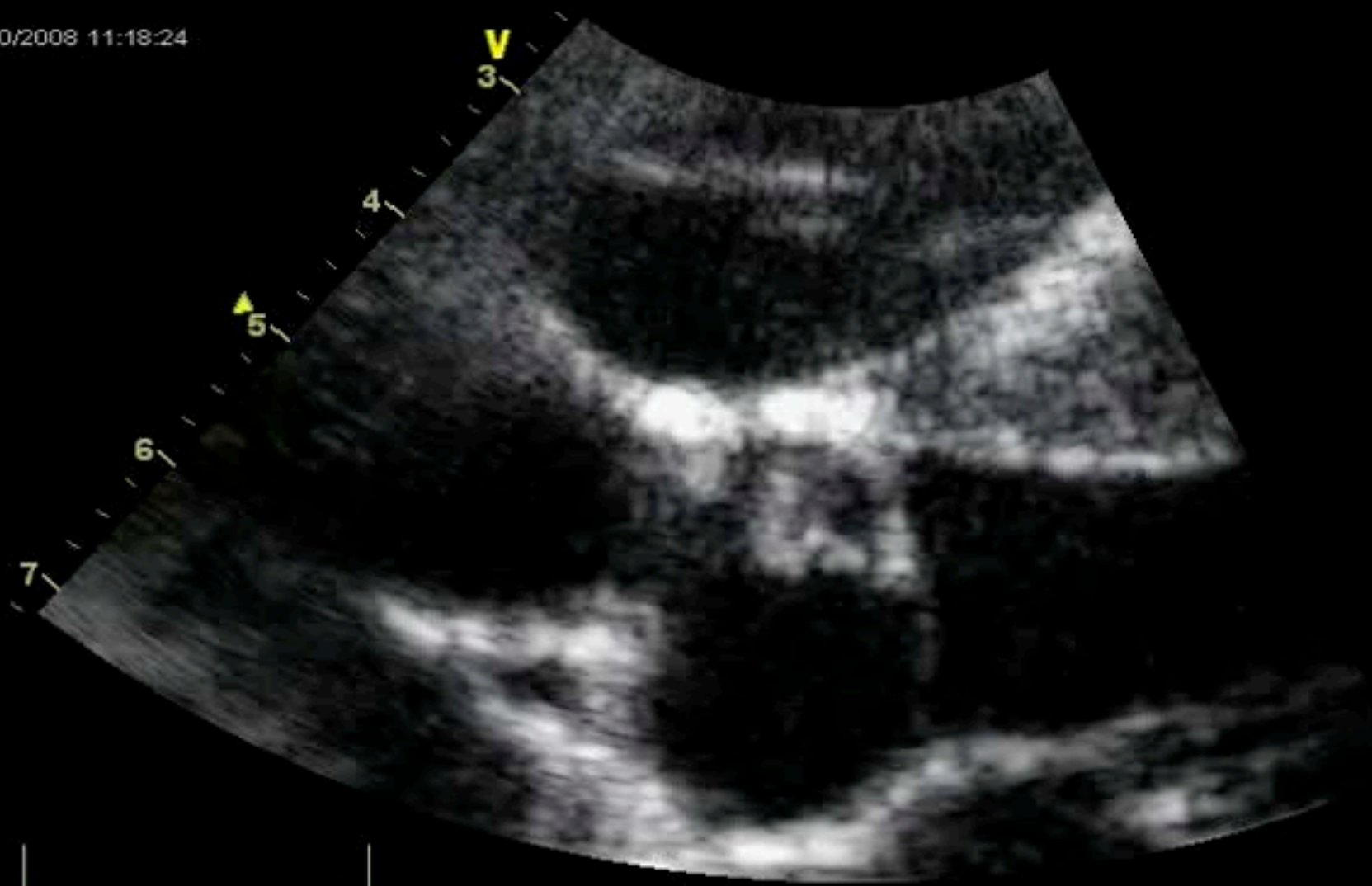
Type III



Fusiform
Narrowing



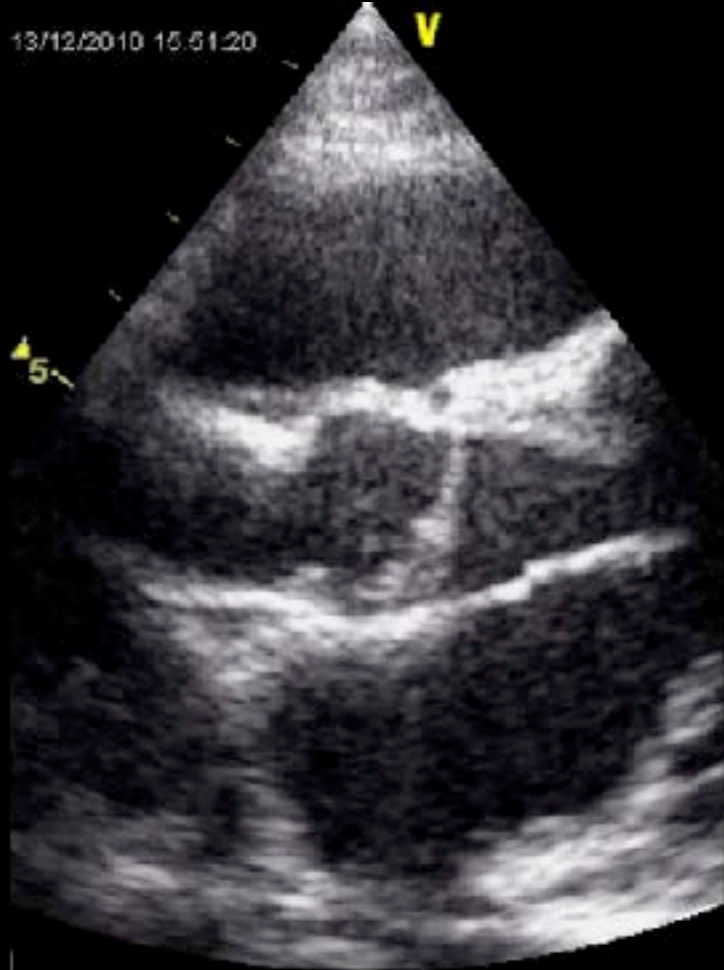
14/10/2008 11:18:24



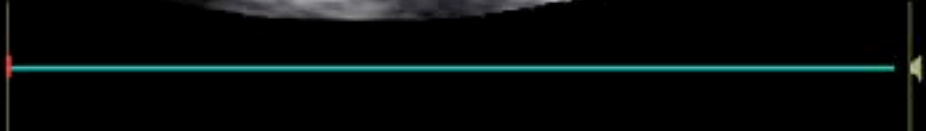
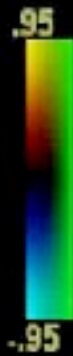
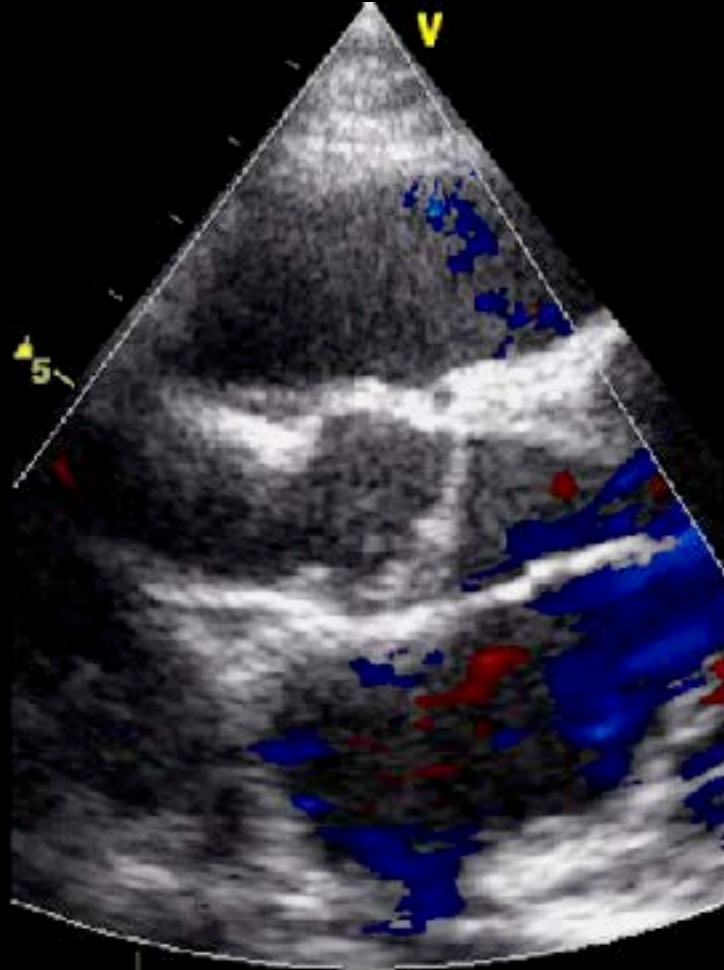
1:47

13/12/2010 15:51:20

V



V



1:59

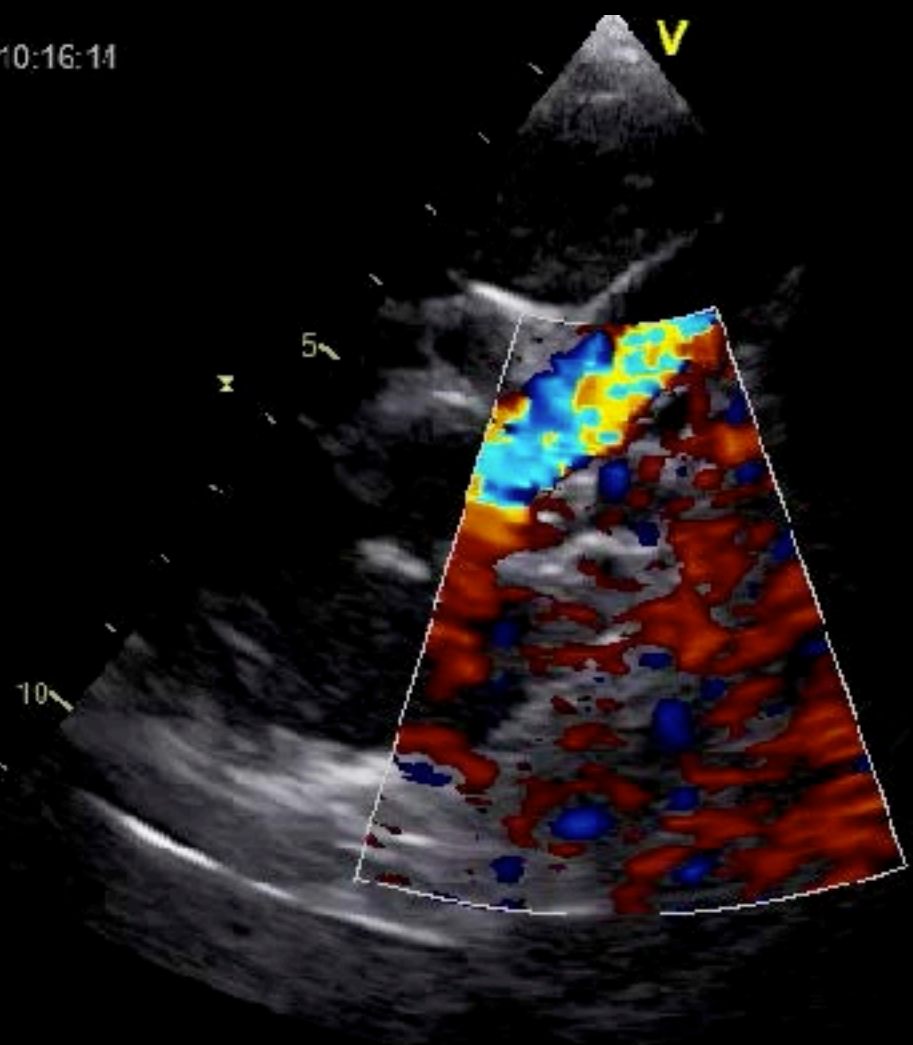
15/06/2018 10:16:14

V

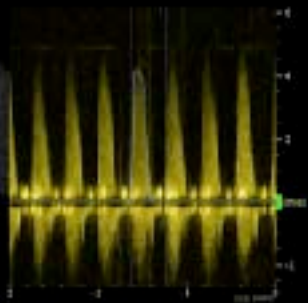
CTO

.80

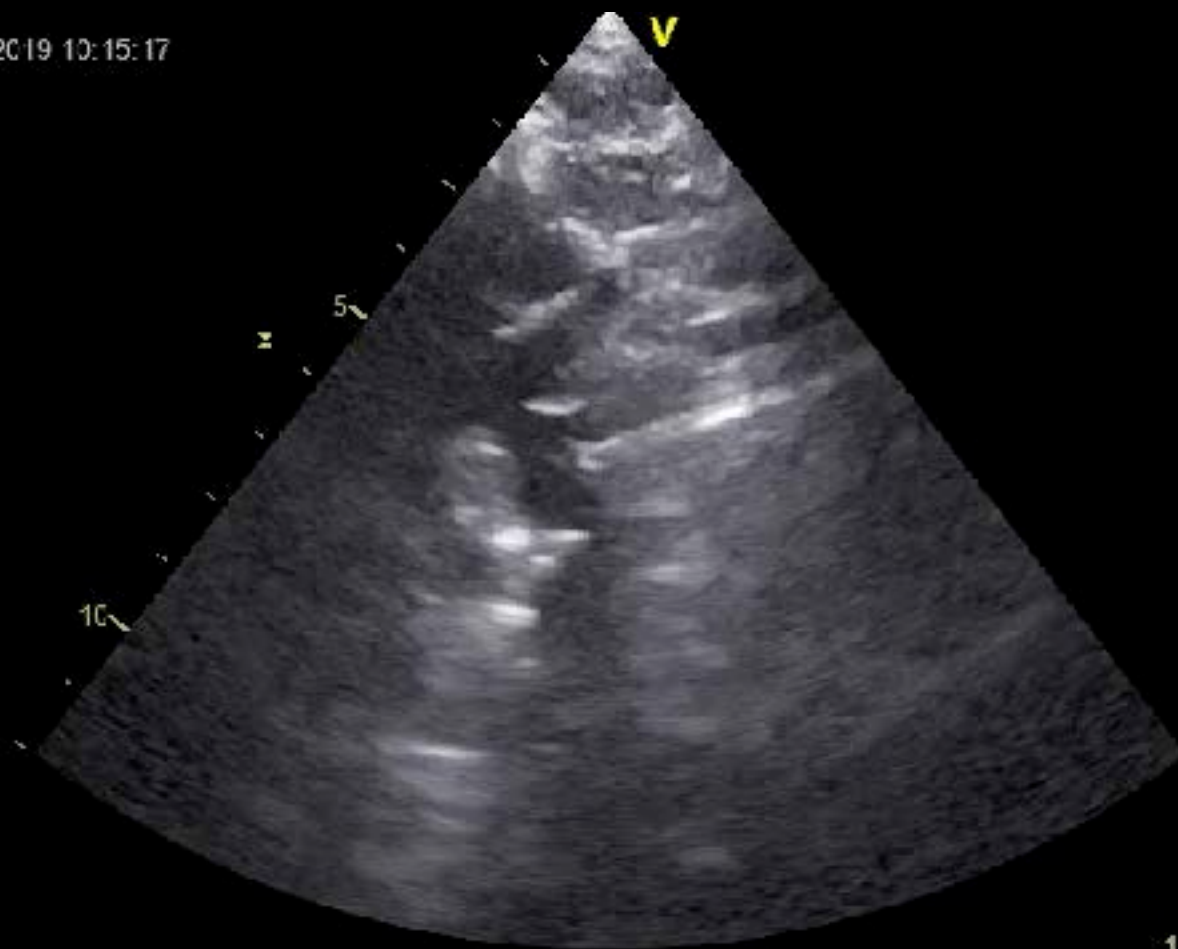
-.80



VT	
VA Area	4.30 cm ²
VA Area	2.70 cm ²
VA CFlow	60.66 mm ³ /s
VA CFlow	30.17 mm ³ /s
VA Flow %	27%
ABTY	61.82 mm
PL	152.06 BPM



07/01/2019 10:15:17



CTO



1:172

ED
Ex: 6255
Se: 2
Volume Rendering No cut

RSP

HOPITAL NECKER ENFANT

DFOV 11.7cm
STND+

A
P
I



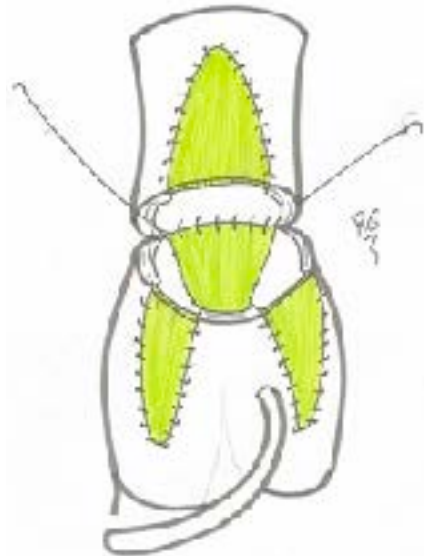
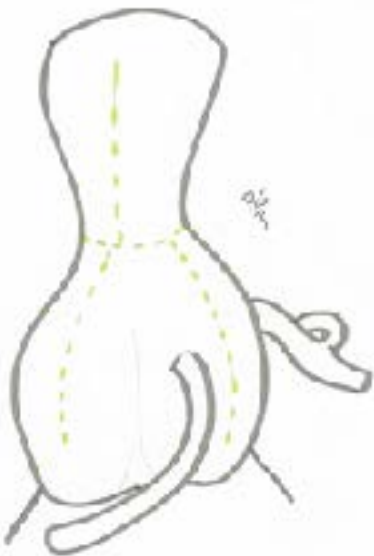
S
5

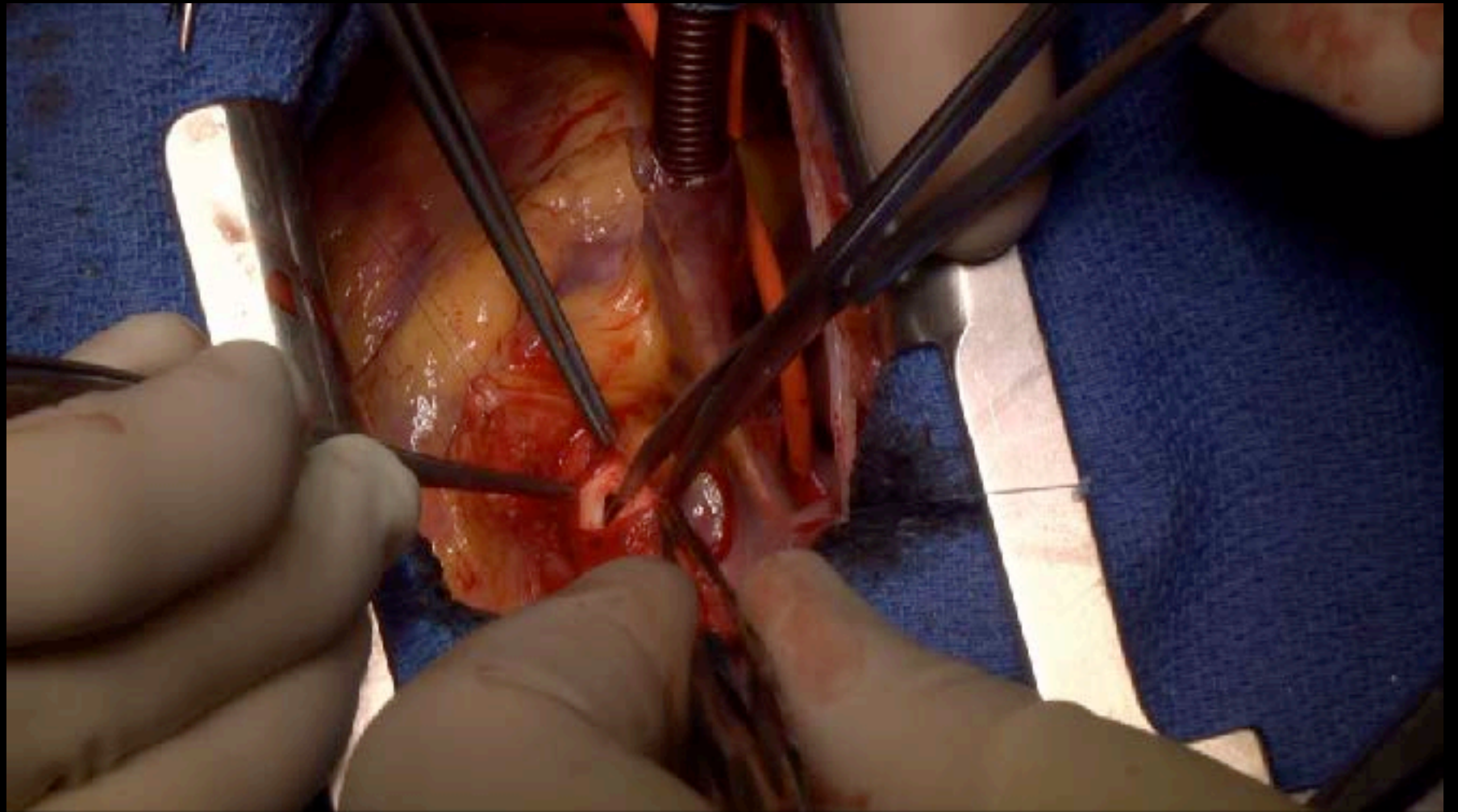
No vOl
lv 80
mA Mod.
Rot 0.43s/HE: 30.4min/rot
C: 6mm 3.084:1/0 Esp
Tilt: 0.0
11:40 53 AM
W - 4095 L - 2043

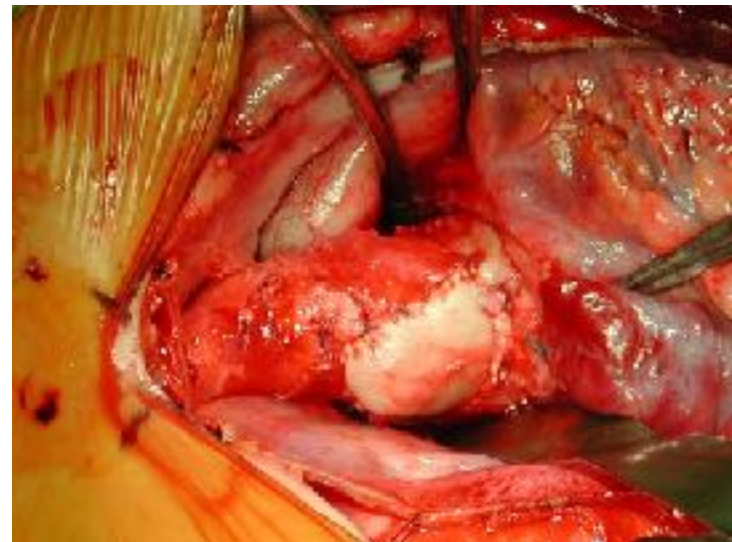
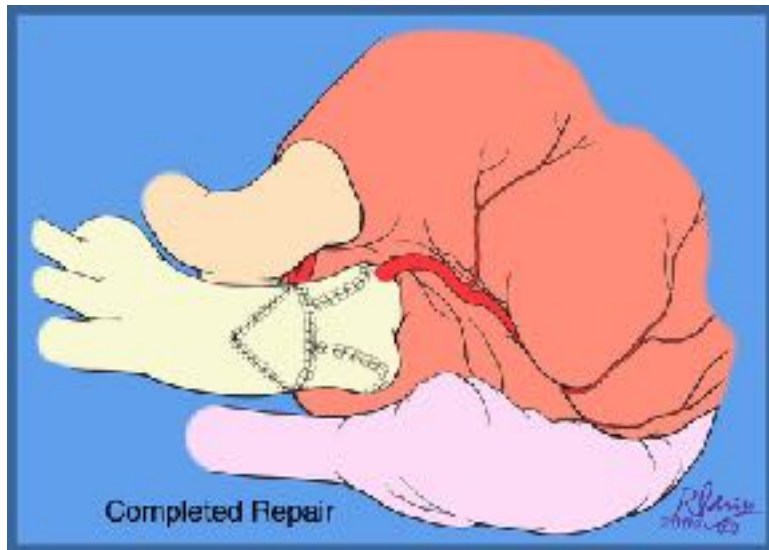
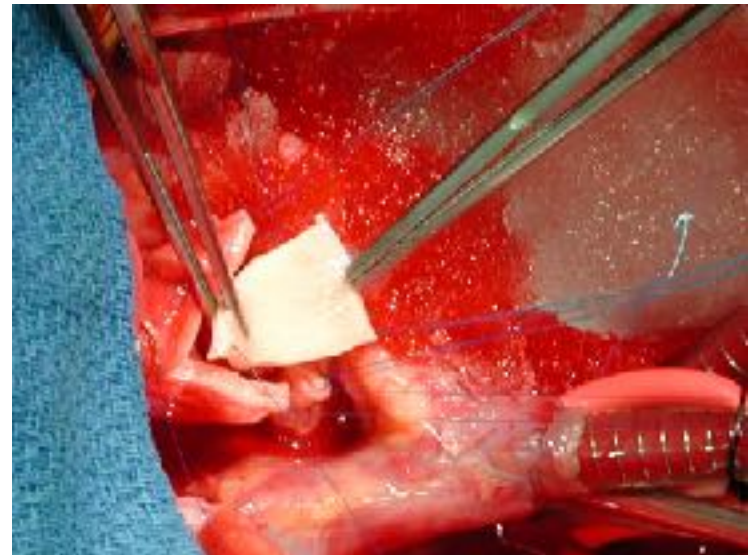
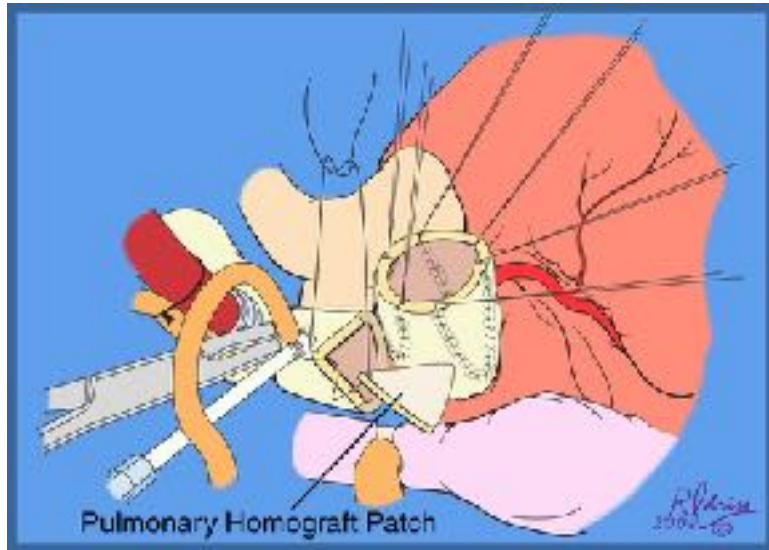
LVA

SVAS CT 3D 1

Sténose supravalvulaire aortique : Technique de Brom

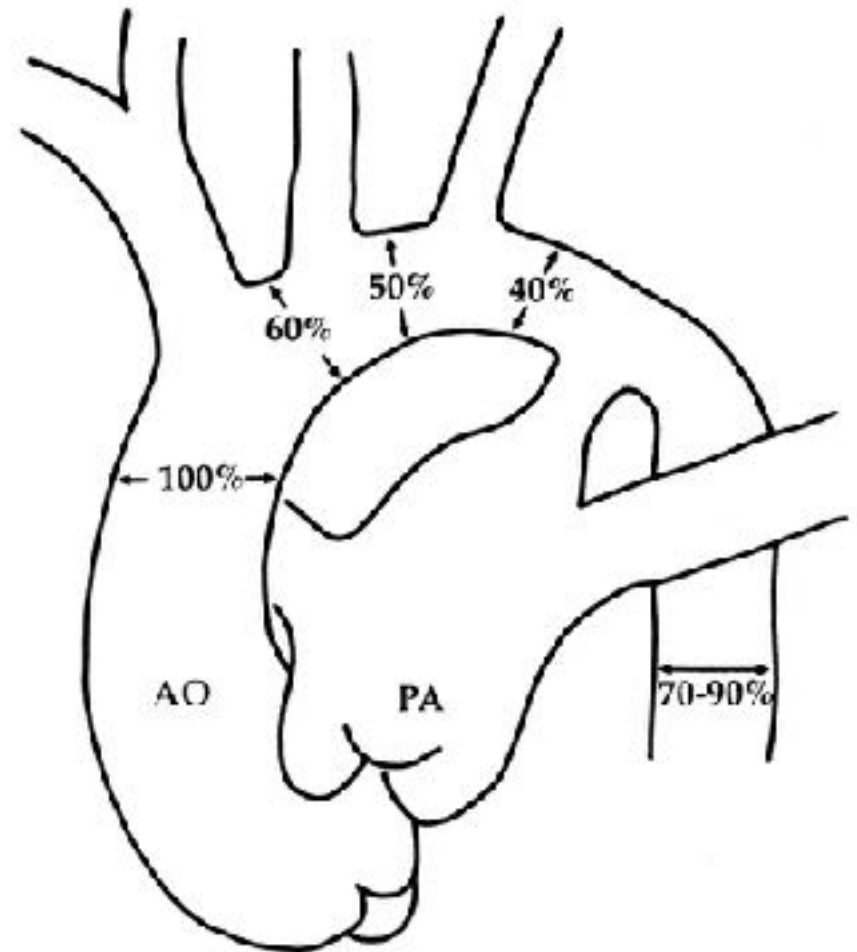
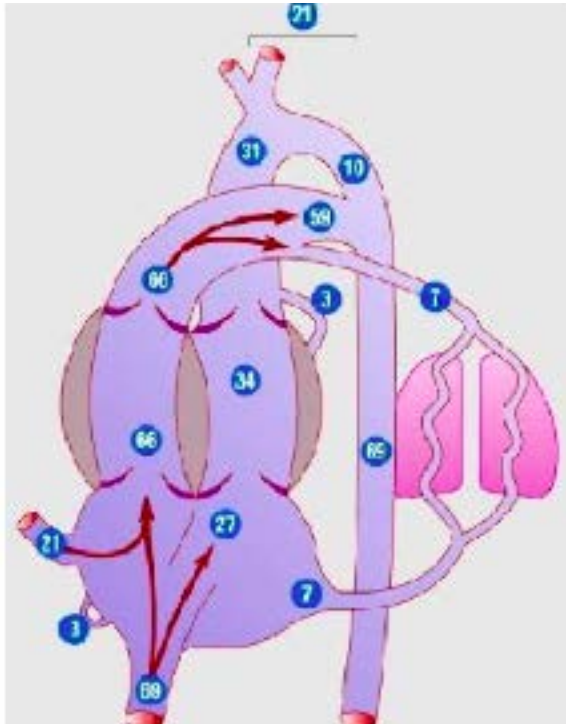




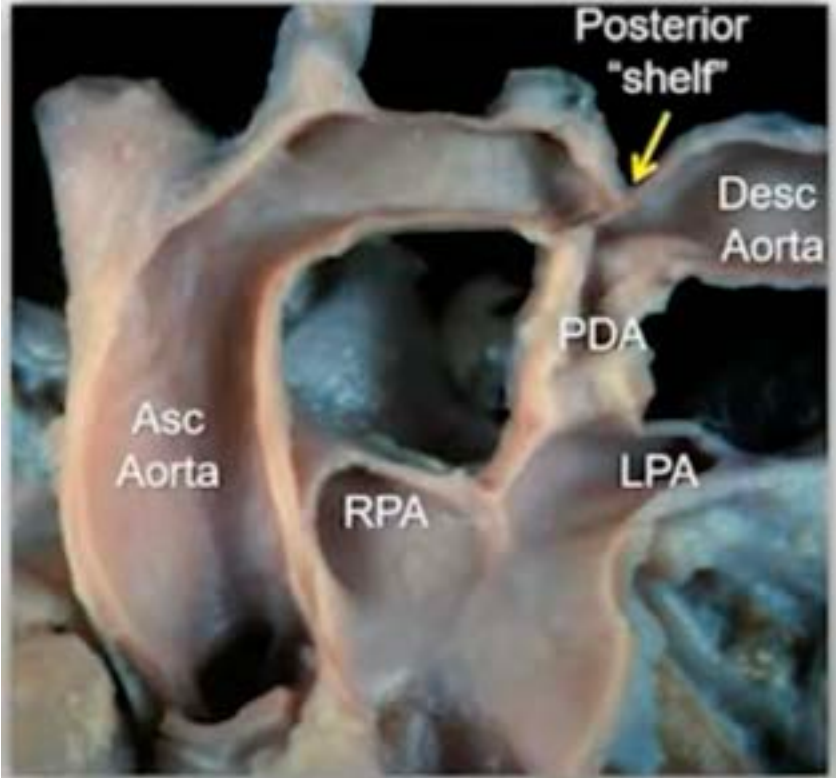
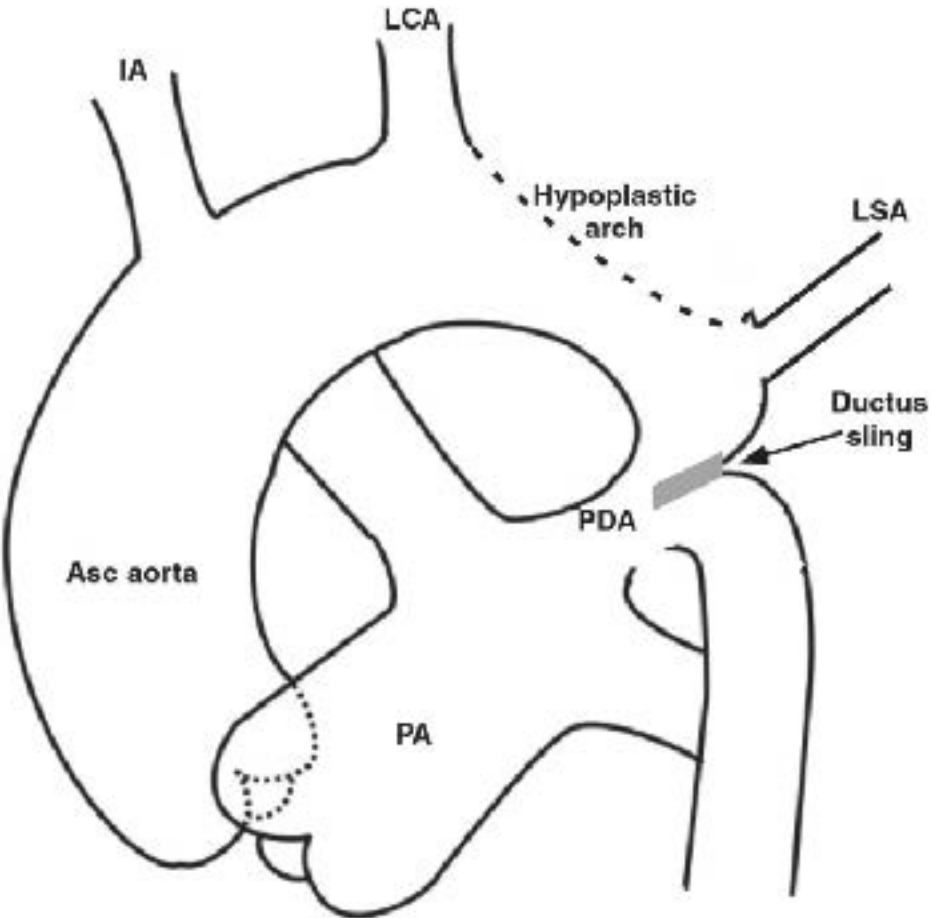


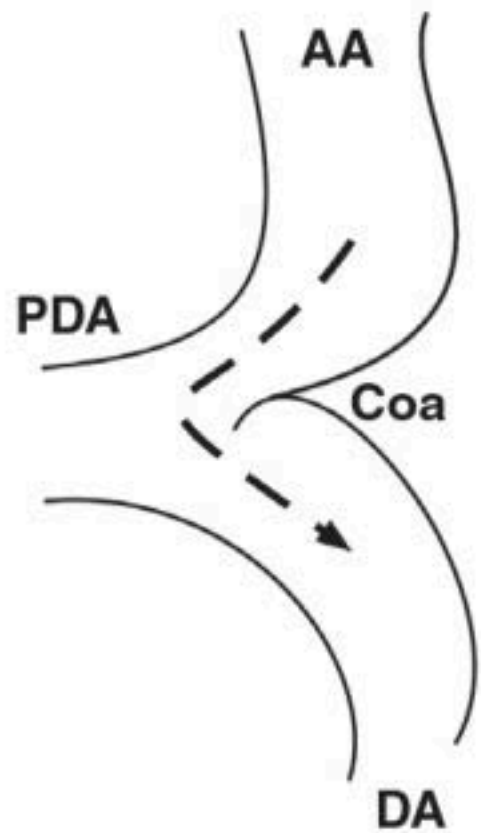
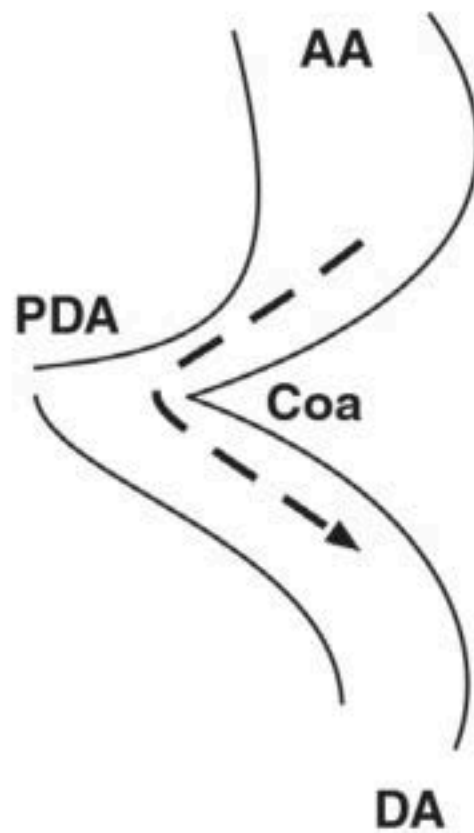
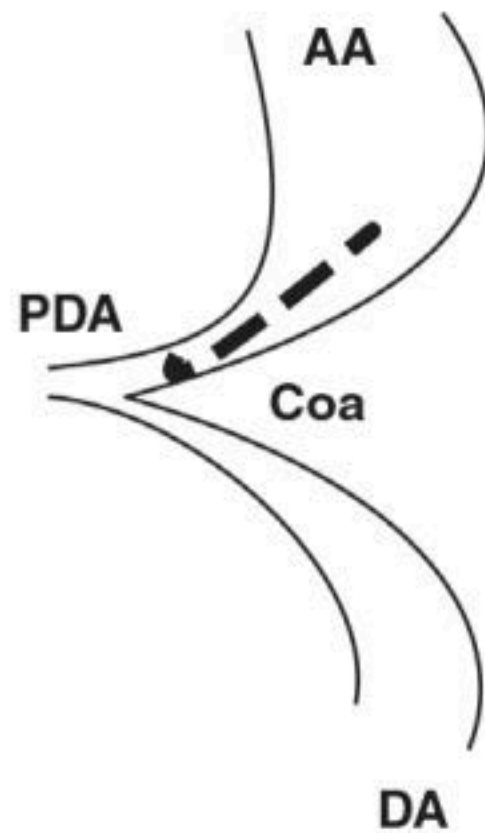
Coarctations de l'aorte

L'arche aortique normale n'est pas un tube régulier

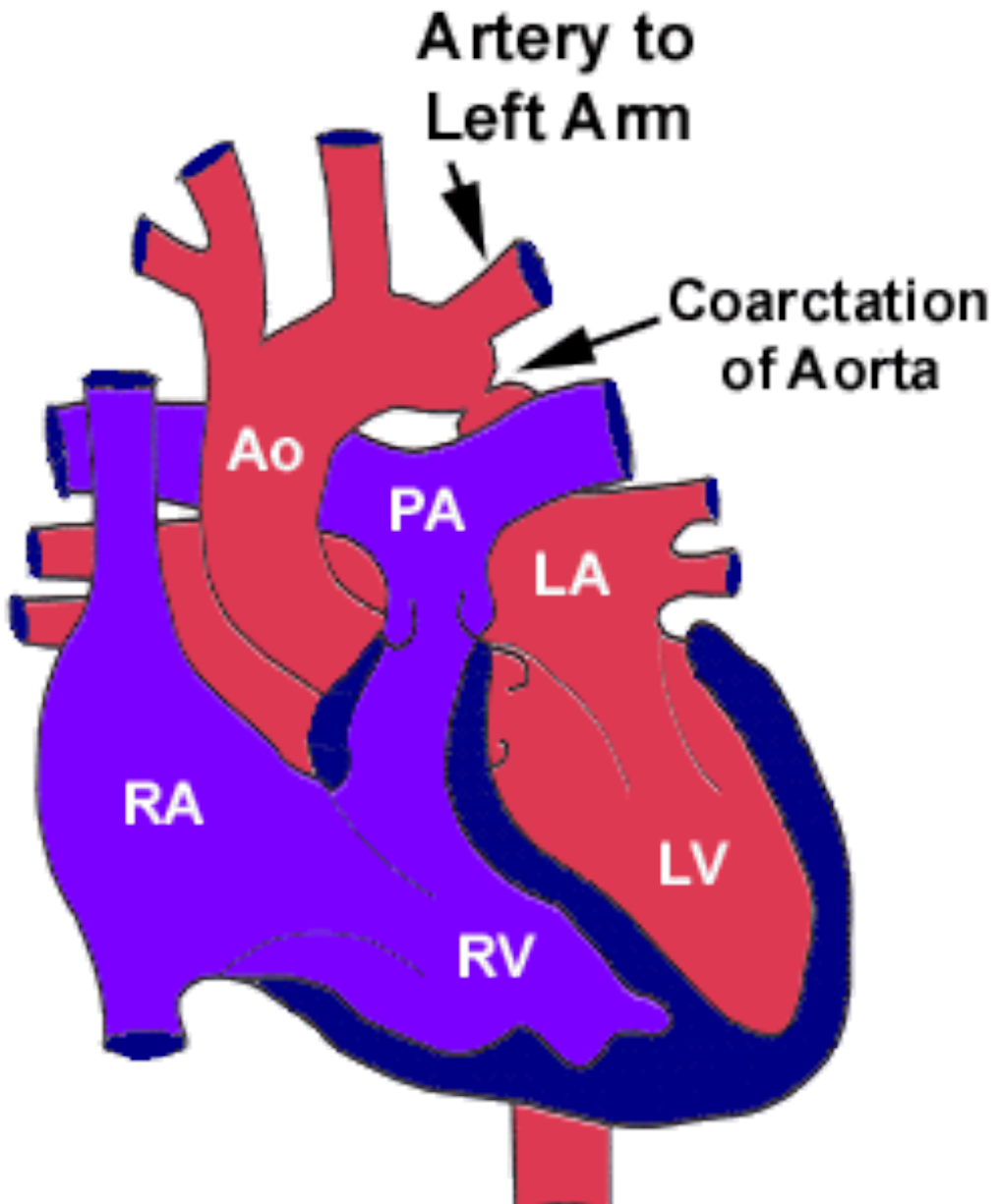


Coarctation de l'isthme aortique



A**B****C**

Coarctation de l'isthme aortique



Epidemiologie

- 40-50/100000 naissances
- 6% des cardiopathies
- Sex ratio H/F 2/1
- Association avec BAV 50-60%
- Sd de Turner (45X0) : 20%

Lésions associées

- valve mitrale
 - RM ou IM congénitaux
- voie sous aortique
 - Désaxation et membrane sous-aortique
 - Anomalies d'insertion mitrale
- **valve aortique**
 - Hypoplasie annulaire, sténose, bicuspidie
- **Hypoplasie de l'arche aortique**
- Syndrome de Shone : mb supraV mitrale, VM en parachute, sténose sous aortique, CoA
- **CIV** : perimembraneuse, malalignement
- Cardiopathies complexes : CAVc , TGA, VU..

diagnostic : présentation clinique

- Nouveau né /enfant/adulte
- pouls fémoraux faibles ou abolis
- asymétrie MS (pouls huméral ou carotidien trop forts) /MI
- HTA membre sup (! Si ASCDRO ou coa comprenant l'ASCG)
- gradient MSup -Mb inférieurs
- Souffle systolique doux dorsal

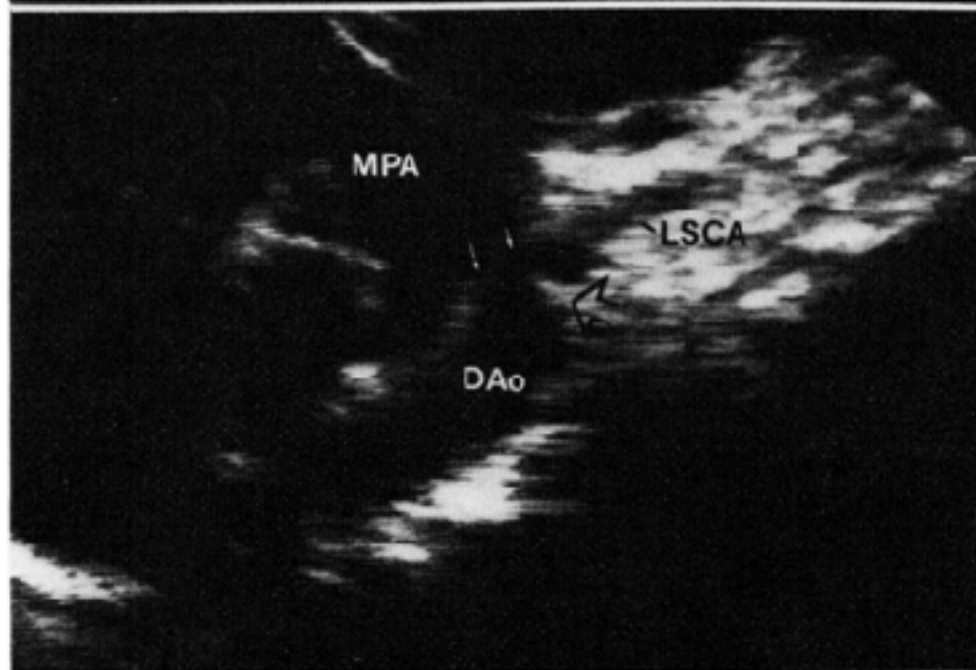
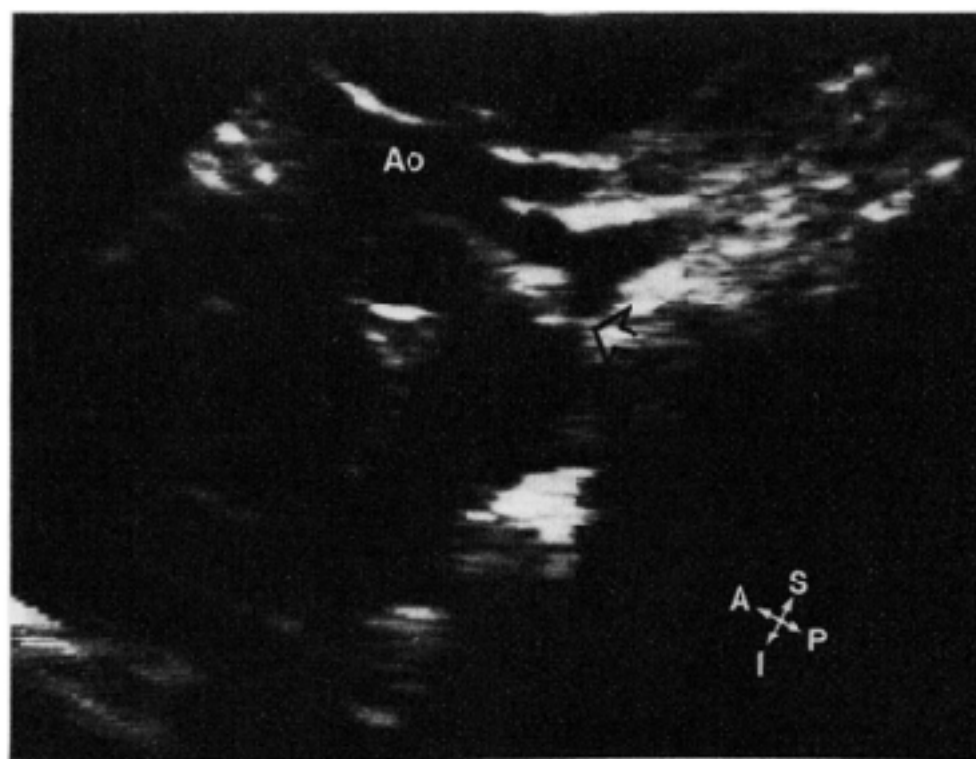
- insuffisance cardiaque si dysfonction VG (tachycardie, polypnée, difficultés alimentaires, signes congestifs et bas débit)

Diagnostic : écho

- échographique
 - gradient sur l'isthme ao $>$ ou $=$ 20mmHg et prolongement diastolique du flux
 - étude de la crosse et isthme aortique
 - VG
 - lésions associées : bicuspidie, obstacles étagés, CIV

Écho

- 2D anatomie : zone étroite
- Doppler Couleur : supra sternale : crosse aliasing isthmique, flux systole diastolique ao descendante
- Doppler continu : pic vitesse systolique, prolongement diastolique
- CW ou PW dans l'aorte abdominale
- Canal artériel : fermé ? Perméable? systémique?
- Mode TM: VG : hypertrophie; fonction



PHILIPS

FR 25Hz
14cm

2D
61%
C 53
P Low
HGen

CF
72%
2.5MHz
WF High
Med



M4 M4

+80.2

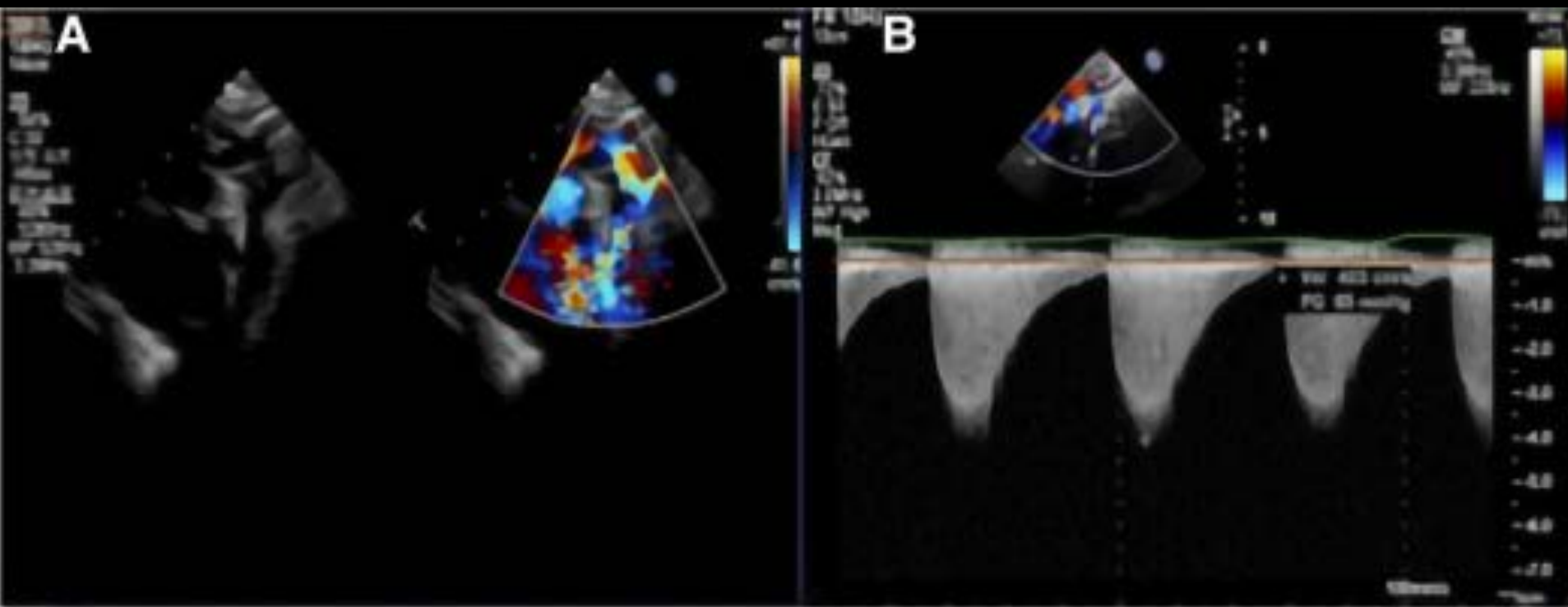


80.2

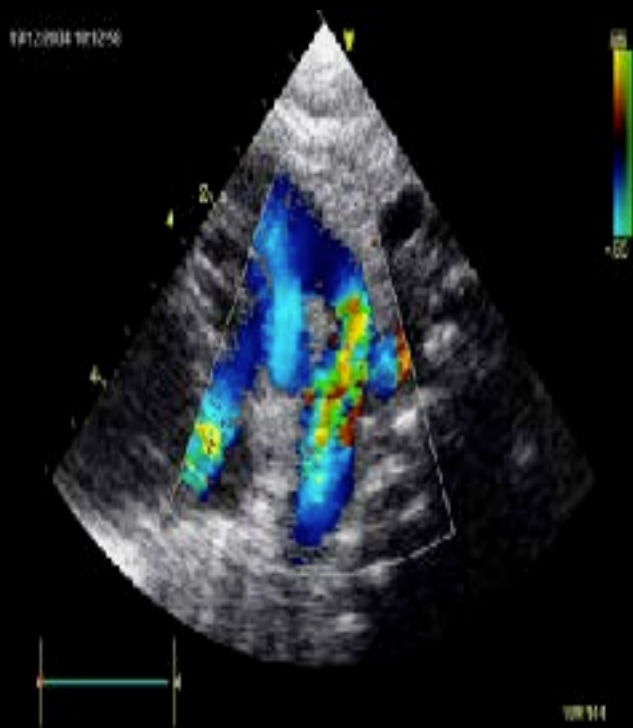
cm/s

JPEG

77 bpm



Perfusion systémique ducto-dépendante ?



3D
BC 488
Sc 2
Volume Rendering: BC ref

176

FORTAL-BROEHRWENT

F 20 1480/040
Mm 17 308

DATA 18.1cm
1.78cm

F
A



No V3
M 02
M4 204
NOT 0400.004 30 000000
0.0cm 0004 0.3 10
M 30
03 00 15 00
W -4000 L - 104

1 85





dering No cut

:m

E+ 39.4mm/rot
k170.5 sp

= 2048



A1



Diagnostic chez l'adulte

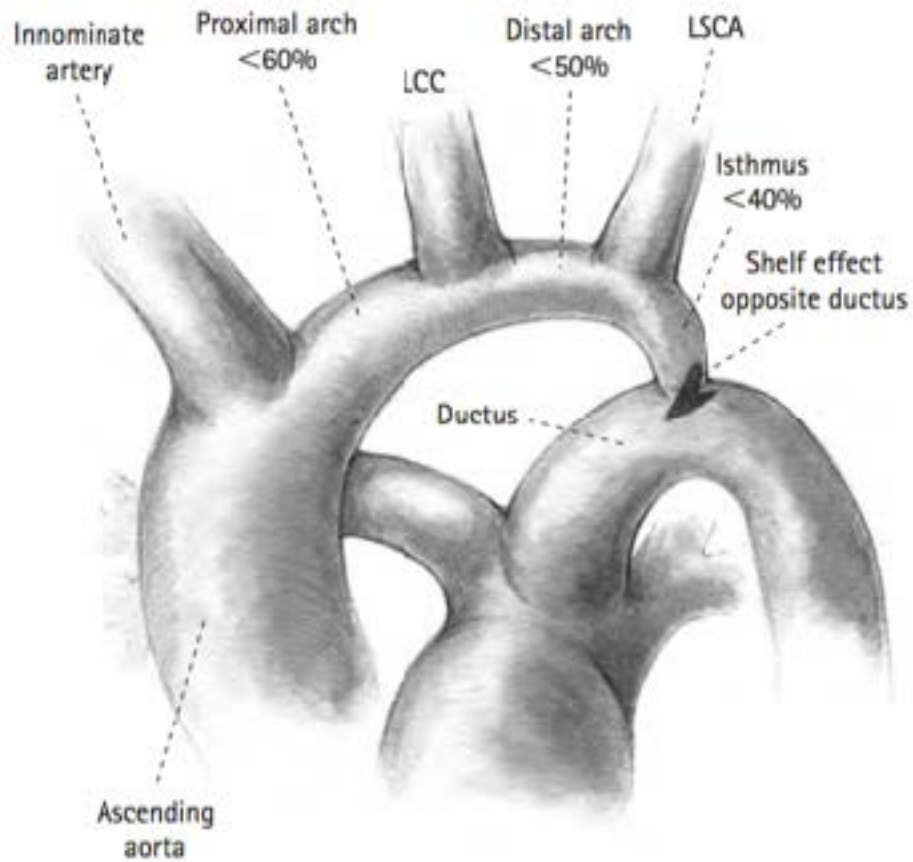
- Présentation clinique : HTA
- Peu de lésion associée sauf BAV
- Dg : écho difficile
- Imagerie multimodale





Stratégie thérapeutique

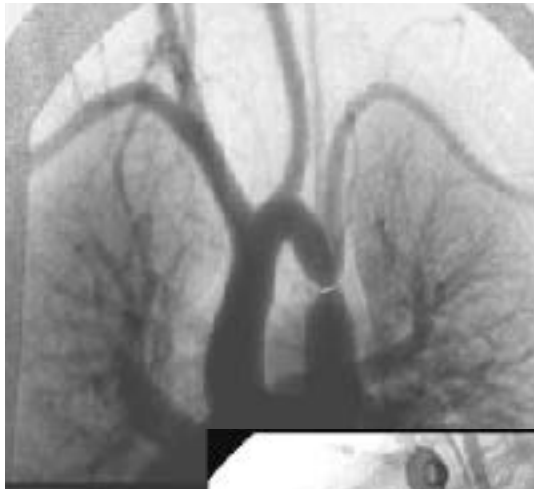
- Ducto-dépendance ou défaillance cardiaque
 - Chirurgie néonatale
- Hypertension artérielle
 - Chirurgie
- Tout va bien : VG adapté , pas d'HTA
 - Chirurgie après 1 mois



Le diamètre des 3 segments

- Arche proximale
- Arche distale
- Isthme

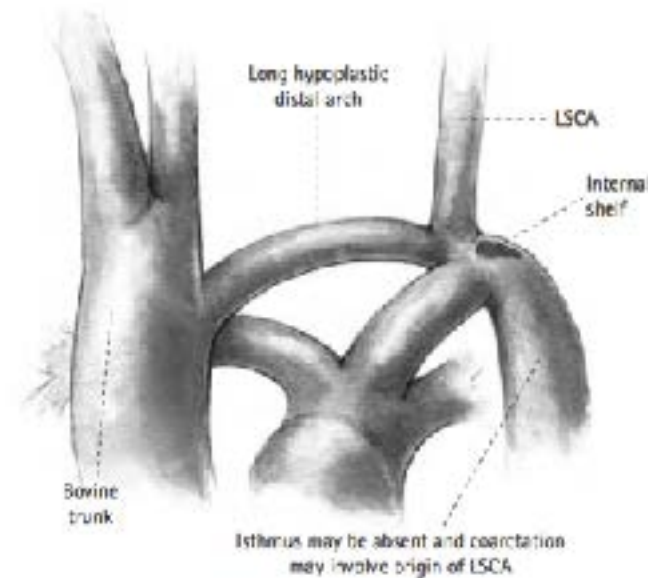
Comment définir l'hypoplasie de l'arche?



- Diamètre (mm) < poids (kg) +1
- Z score inférieur à -2
- Diamètre inférieur à 50% de l'Ao Asc

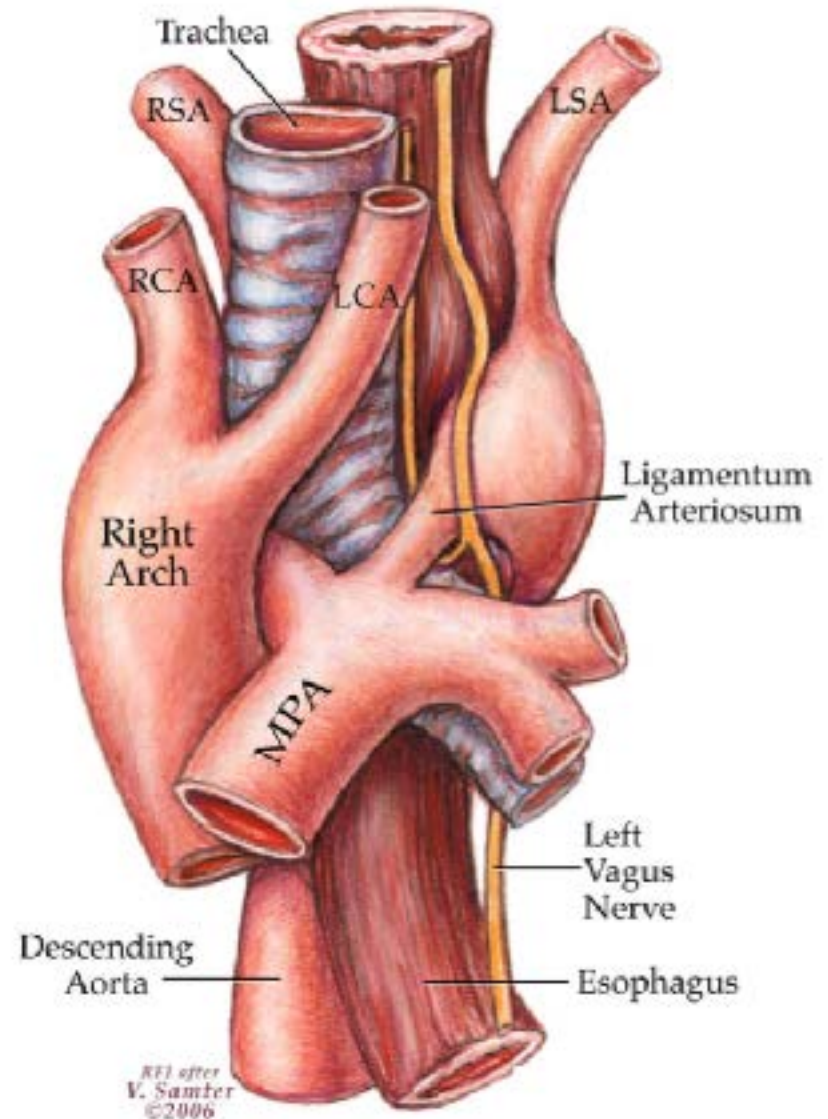
préciser aussi:

La distribution des troncs et la longueur des segments -> peut conditionner la réparation

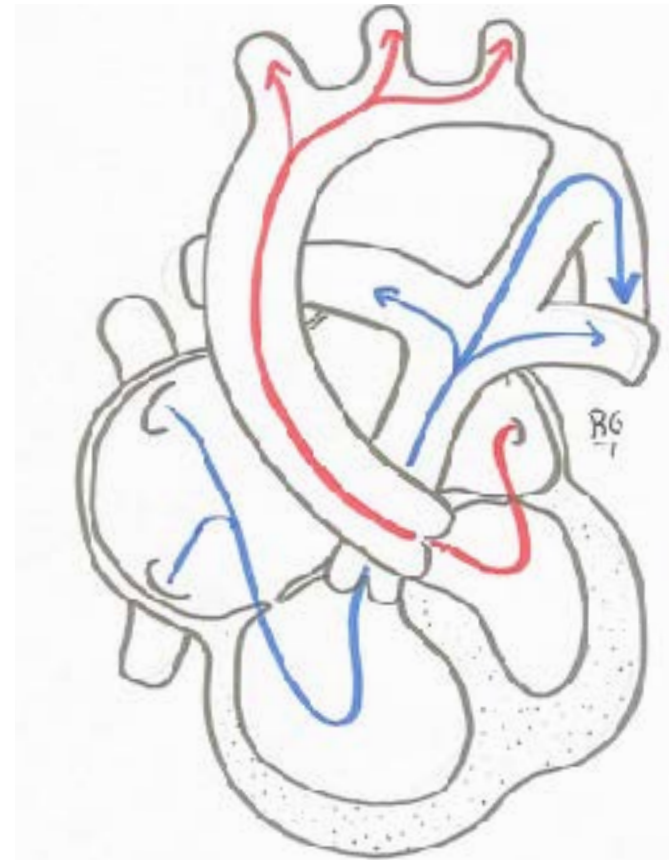
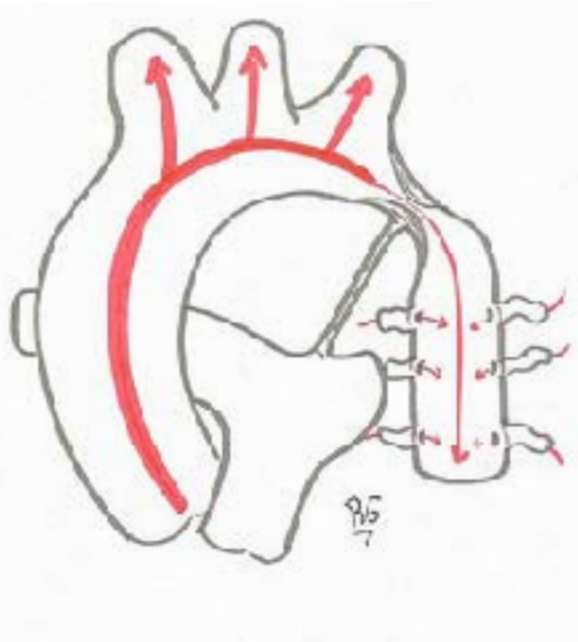


Préciser
Le coté de la crosse aortique

**Chirurgie impossible par
thoracotomie gauche
en cas d'arche droite**



traitement chirurgical



traitement chirurgical : abord

Par thoracotomie



Par sternotomie



stratégie établie selon :

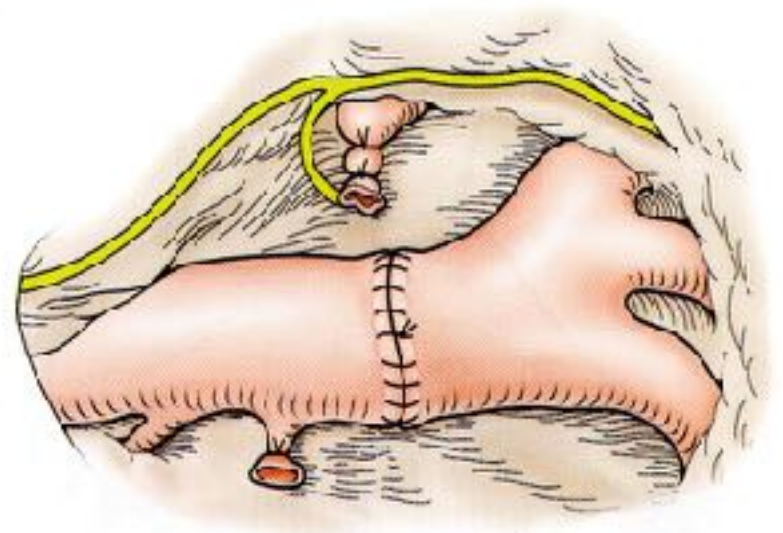
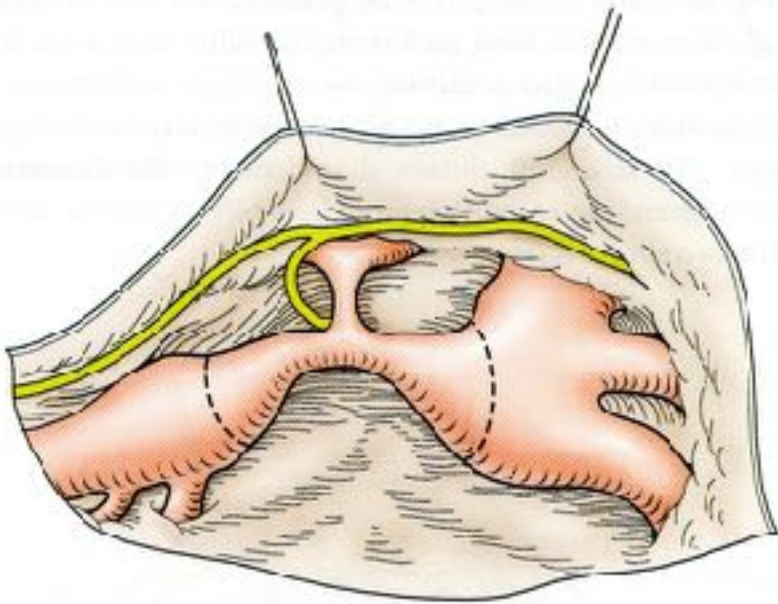
- 1° Le degré d'hypoplasie du segment proximal de l'arche
- 2° La cardiopathie associée

techniques chirurgicales

Hypoplasie	Lésions associées	Techniques de réparation
isthme	0	anastomose termino-terminale thoracotomie
+ arche distale	+ (CIV)	anastomose termino-terminale élargie thoracotomie sternotomie – CEC (si CIV)
+ arche proximale	++ (cardiopathie complexe)	termino-terminale élargie + patch d'élargissement sternotomie – CEC

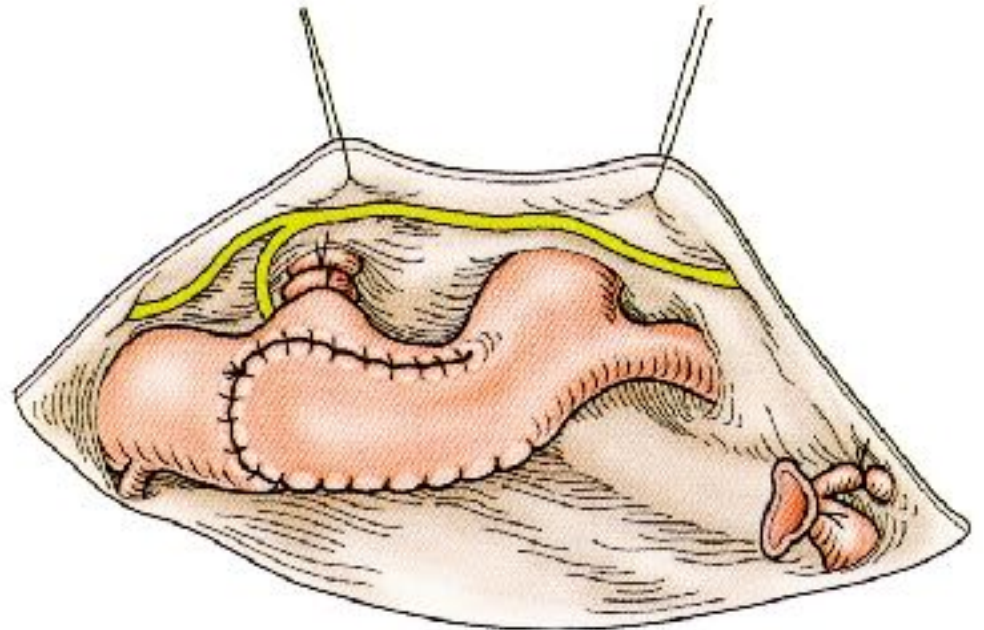
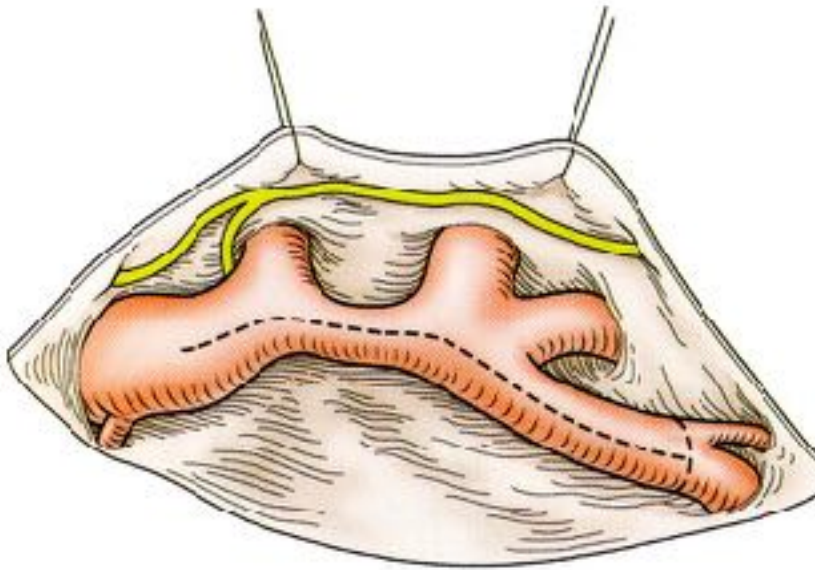
Réparation par Thoracotomie

Résection anastomose: Crafoord



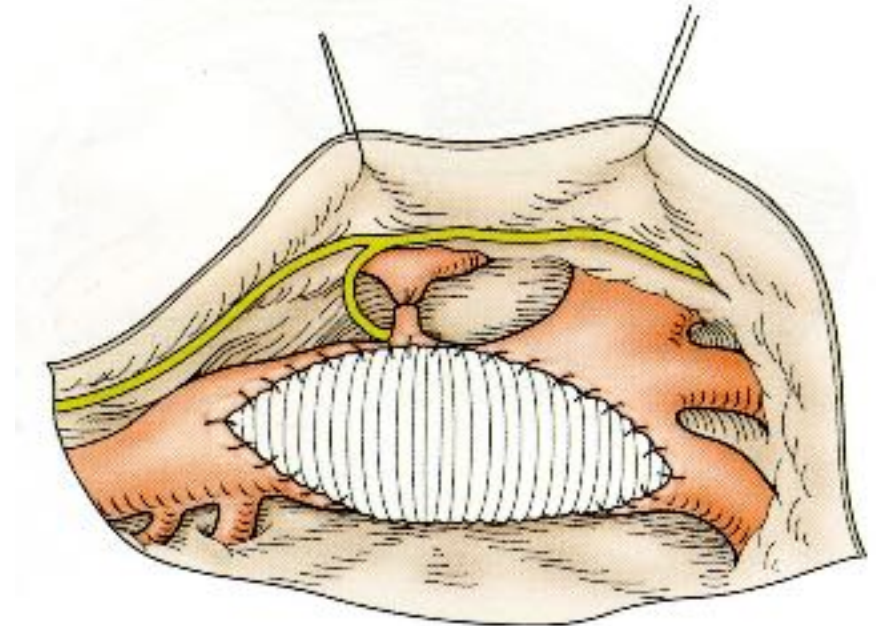
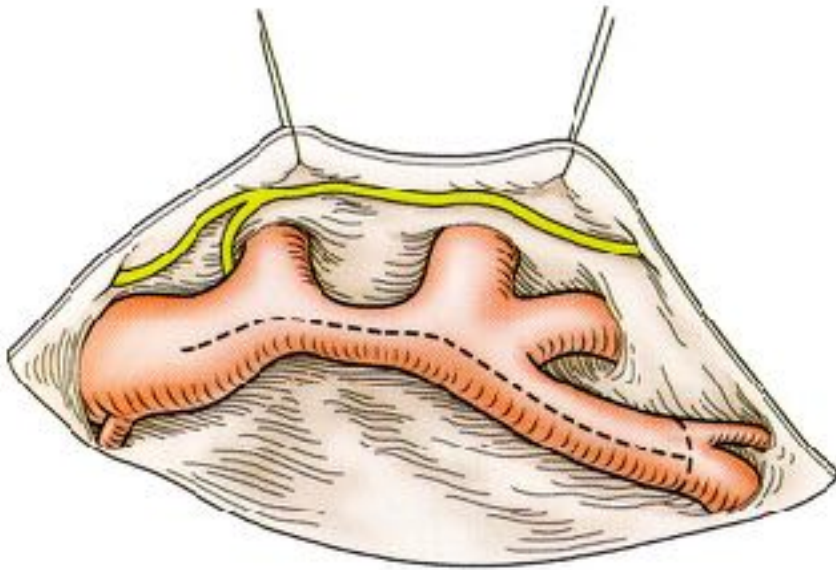
Réparation par Thoracotomie

Réparation avec un flap subclavier: Waldhausen



Réparation par Thoracotomie

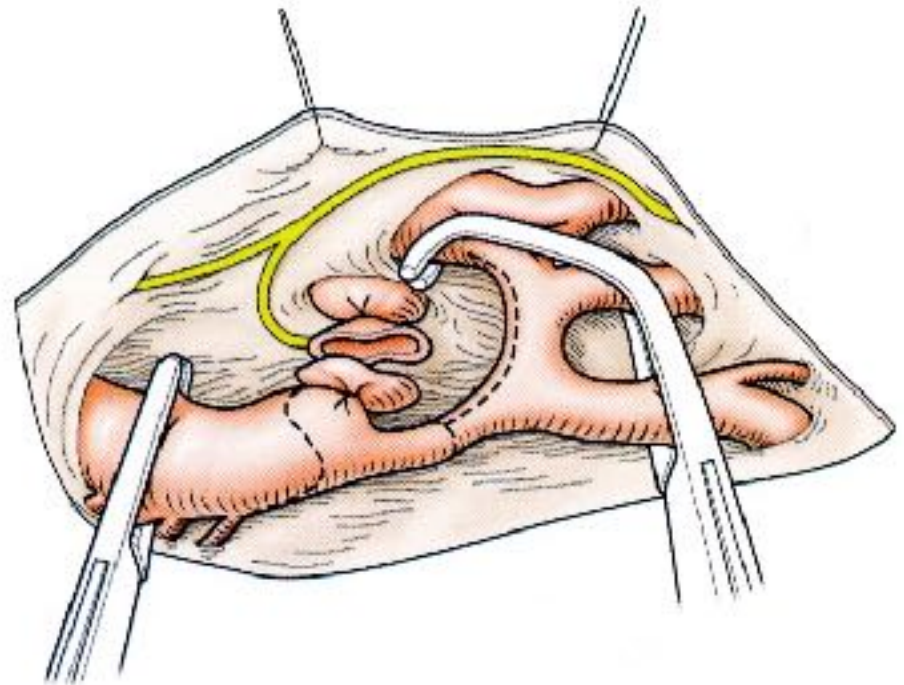
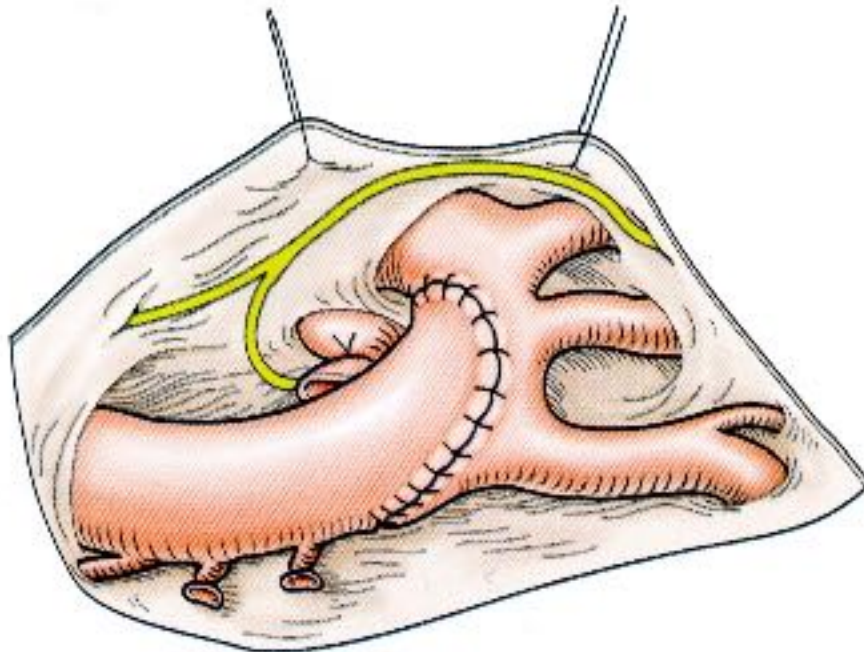
Plastie d'élargissement avec patch



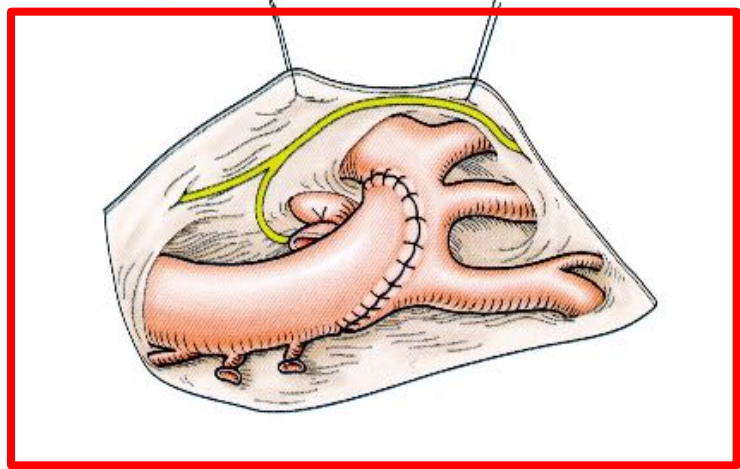
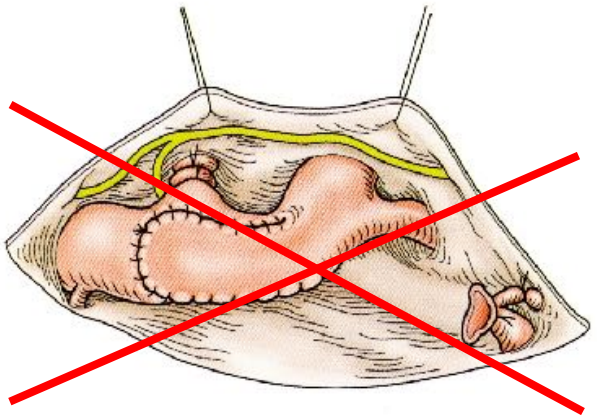
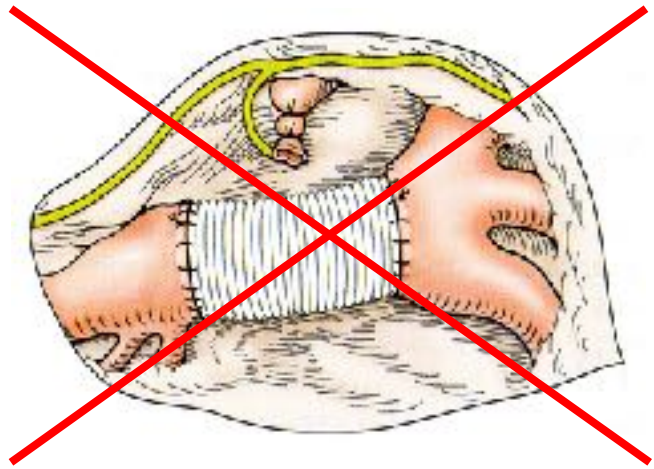
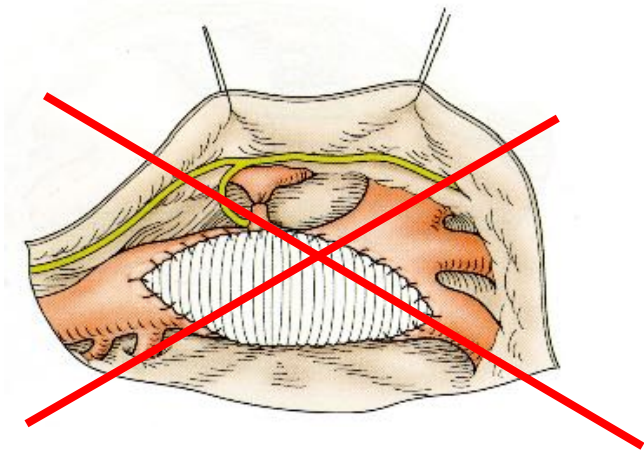
Quelle technique chirurgicale?

Réparation par Thoracotomie

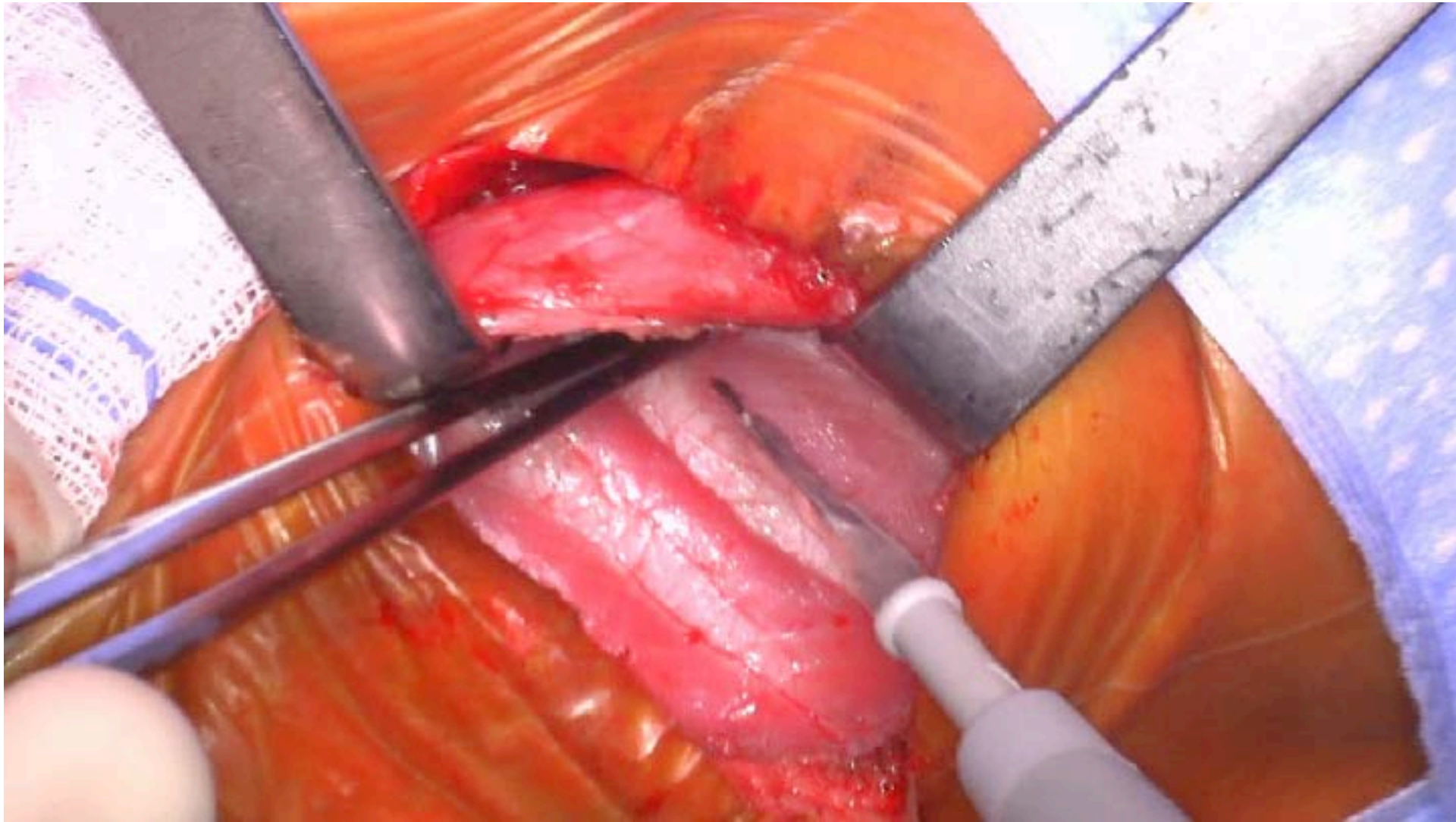
Résection anastomose étendue: Crafoord « modifié »



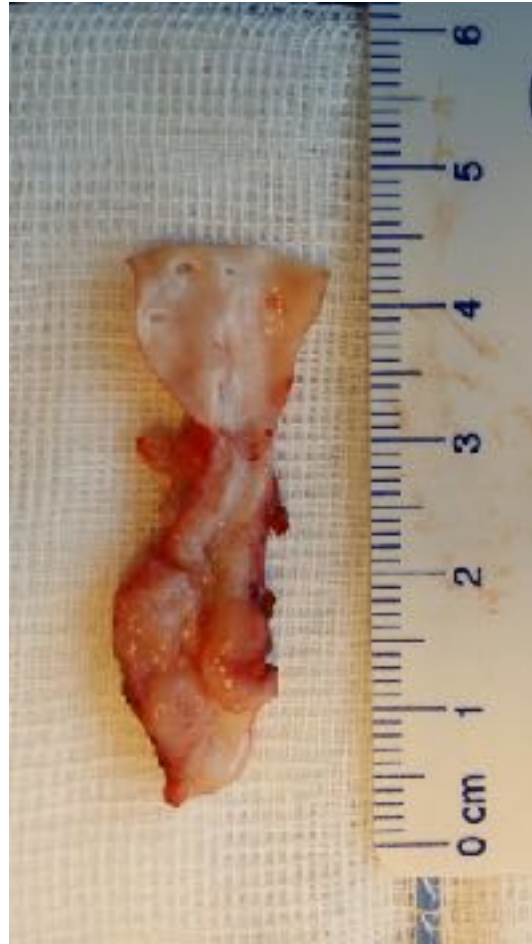
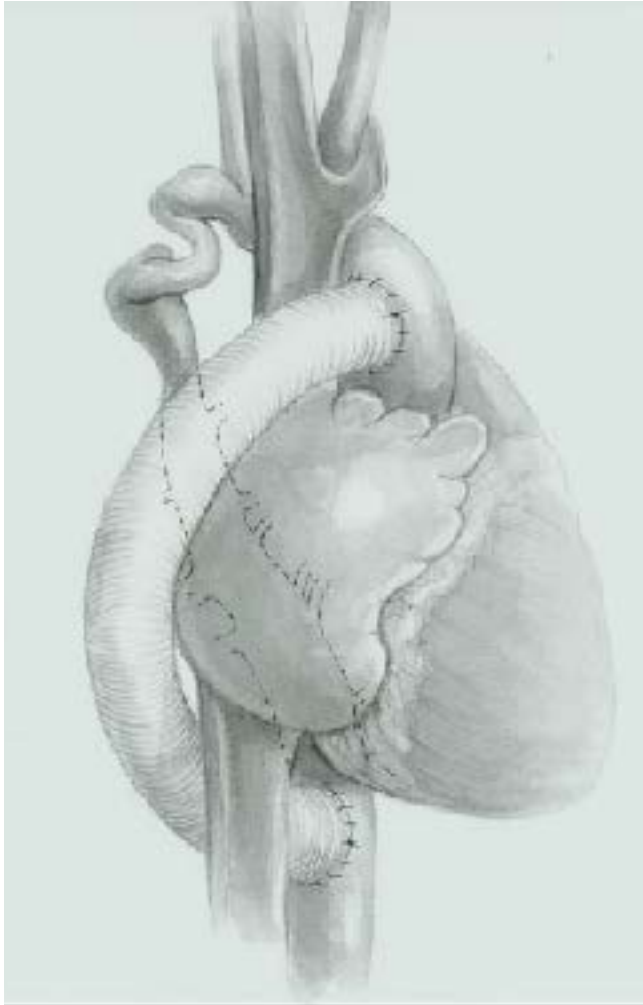
Réparation par Thoracotomie



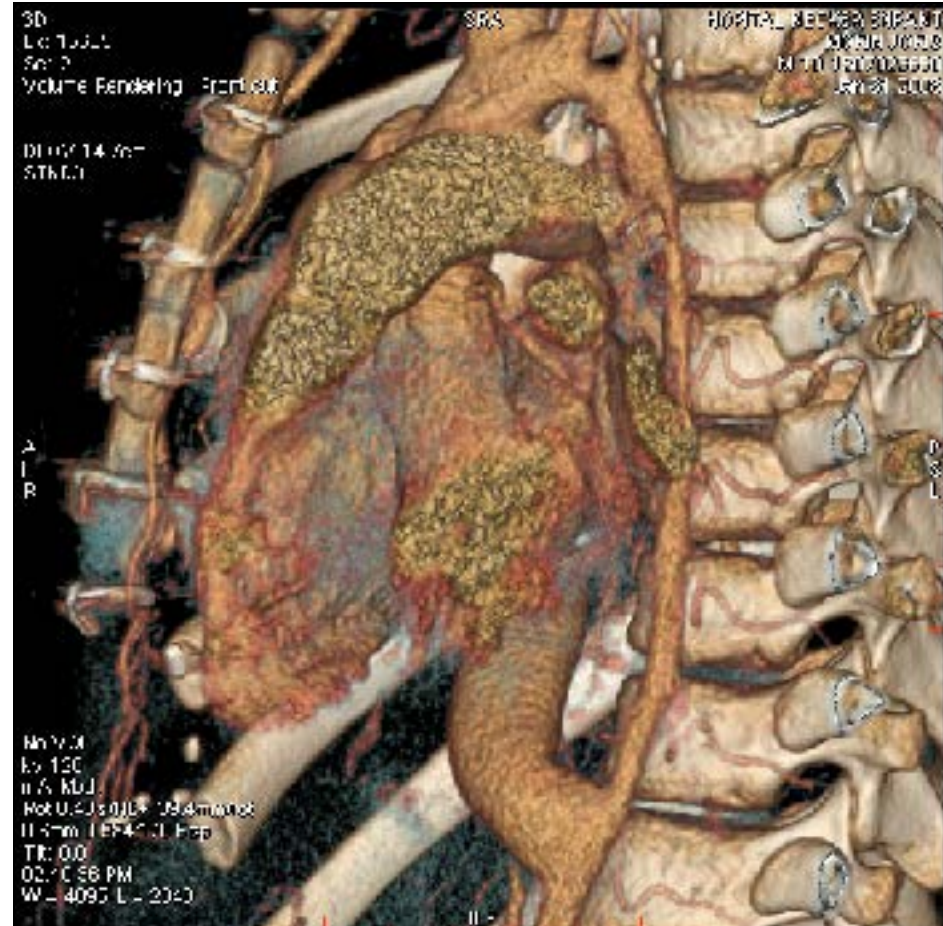
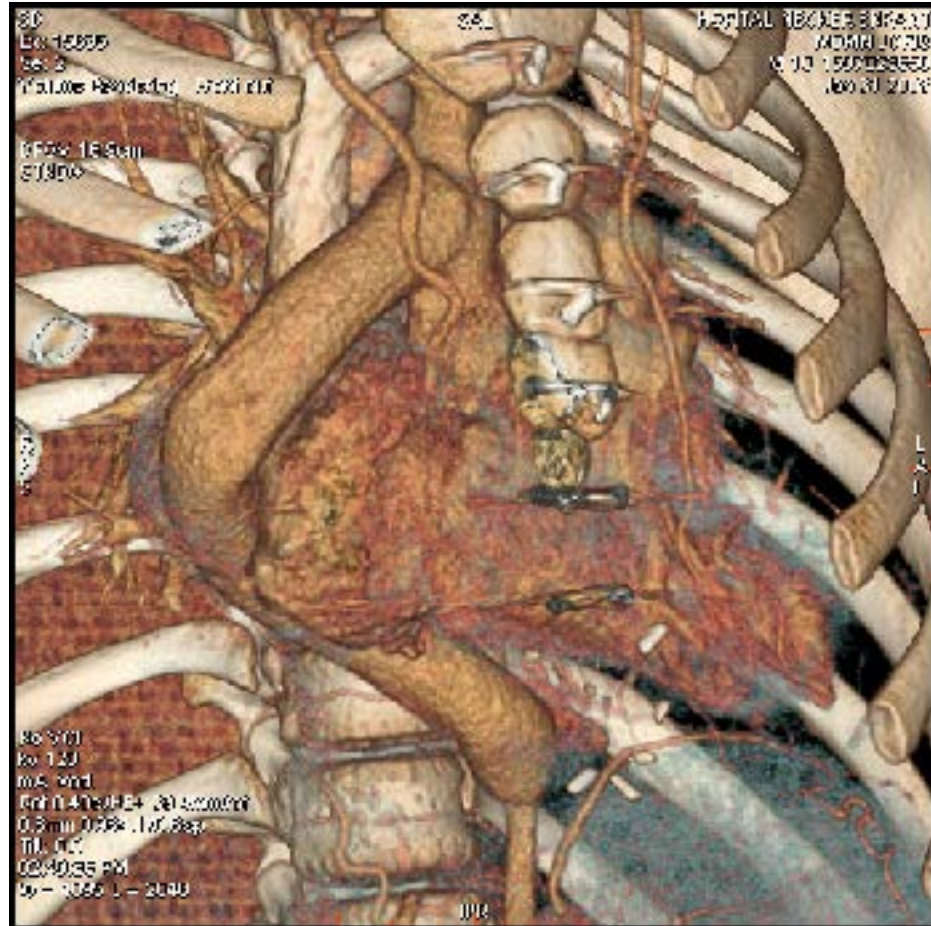
cure de coarctation par
thoracotomie



Formes complexes d'hypoplasie PHACE syndrome



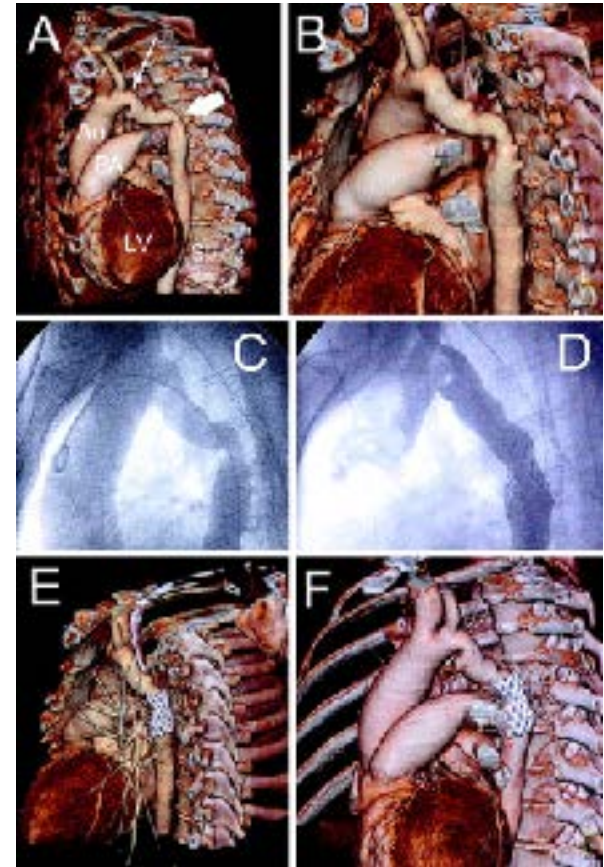
P: anomalie fosse Posterieure
Hemangiomes
Anomalies Arterielles
Coarctation de l'aorte
Eye : anomalies oculaires
S: anomalies Sternales





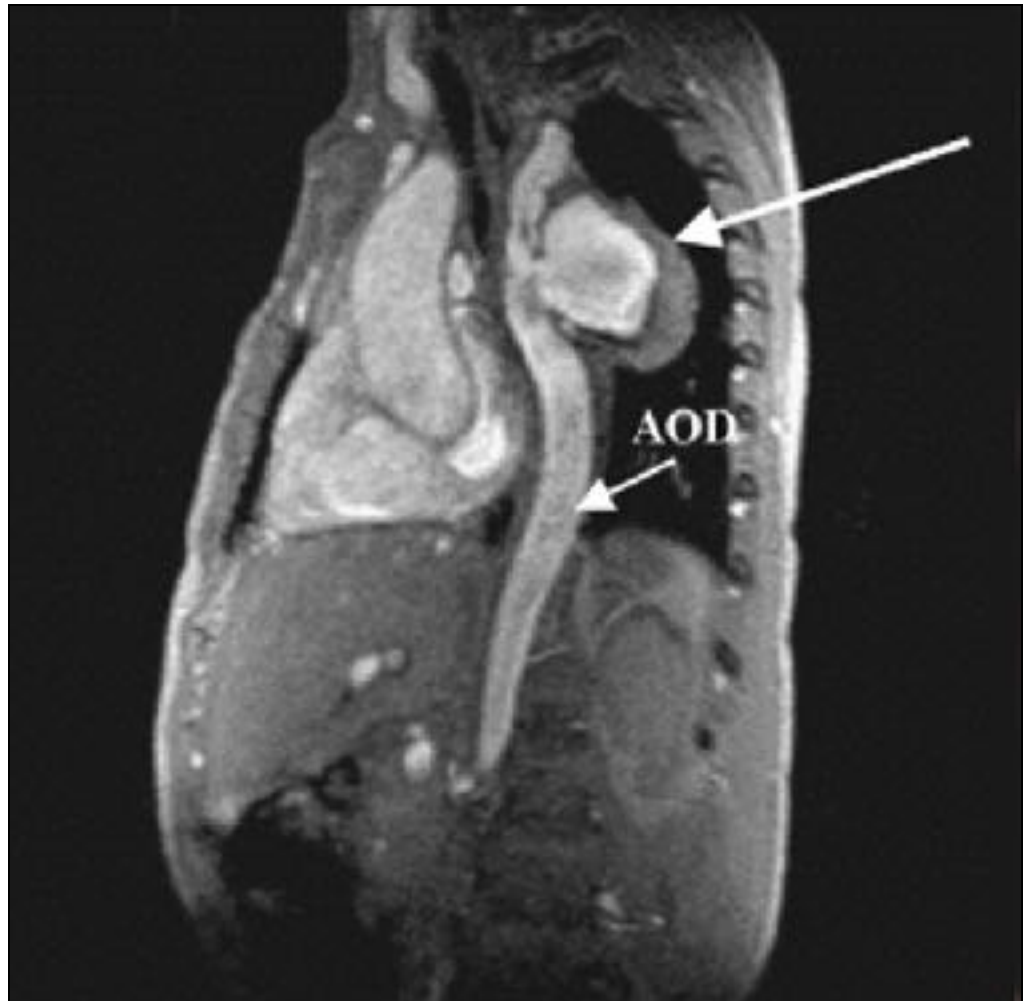
Au-delà de la période néonatale

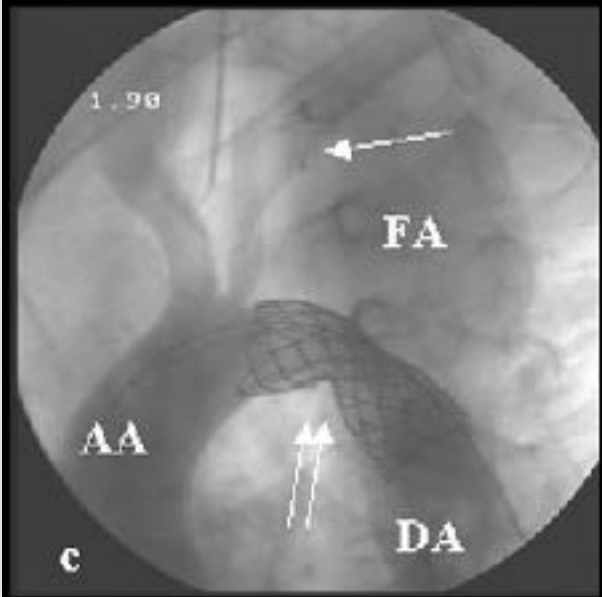
- Indications opératoires
- Méthodes
 - Chirurgie chez l'enfant ?
 - Stenting direct chez l'adulte ?



Complications immédiates

- Poussée hypertensives post-opératoires
- Infection de l'anastomose
- thoracotomie :
 - Chylothorax
 - Paralysie récurrentielle
 - Syndrome occlusif





Complications précoces

- Re-coarctation
 - Surtout la première année
 - Dilatation au ballonnet
 - Coarctation résiduelle
 - Hypoplasie aorte horizontale non traitée

Prévalence de l'HTA

- 49% d 'HTA à 17 ans *Kappetein 1994*
- 68% d 'HTA à 30 ans *Presbitero 1987*
- *Influencée par l'anatomie de l'arche aortique (gothique vs romane)*

Hypertension artérielle tardive

Surmortalité après cure de coarctation de l'aorte

- 182 pts avec 40 ans de suivi
 - 22 décès (18 cardiovasculaires) : 12%

Bobby 1991

- 0.8% décès par pt/année

Koller M 1987

- 87 décès/646 pts

– âge moyen du décès 38 ans

Cohen M 1989

Mortalité cardiovasculaire

- Causes de décès
 - coronaropathie
 - mort subite
 - insuffisance cardiaque
 - accident vasculaires cérébraux
 - rupture d 'anévrisme aortique
 - valvulopathie aortique

Coarctation abdominale

- Étiologies
 - Aortites inflammatoires Takayashu
 - NF1
- Lésions étendues : mésentérique sup -> iliaques, artères rénales
- HTA
- ETT: HVG
- Ttt chirurgical