

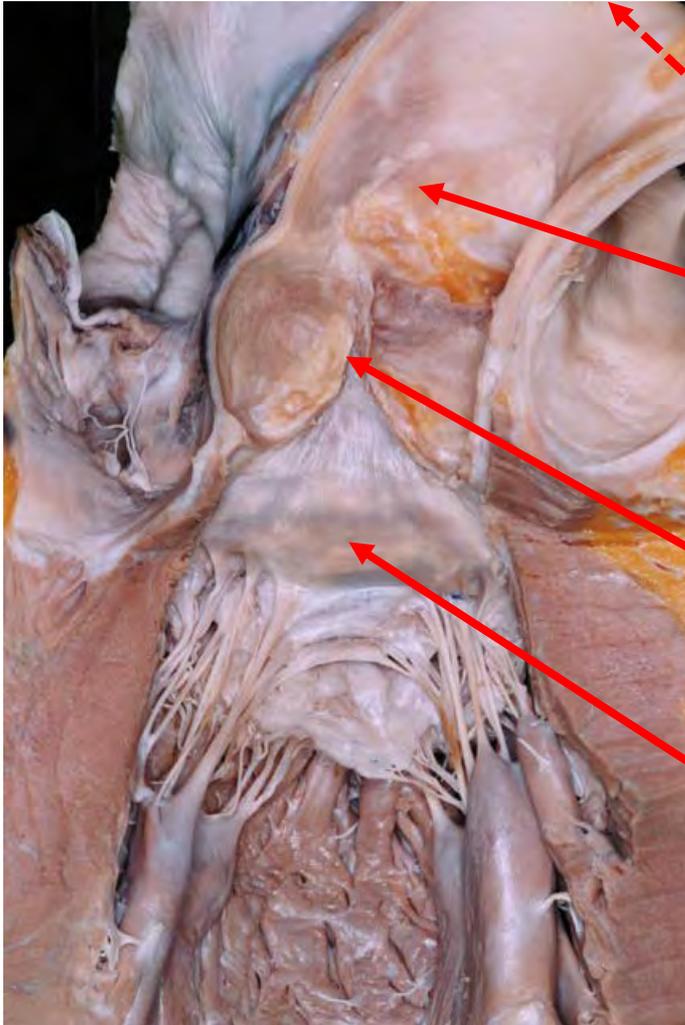
Sténoses de la voie d'éjection gauche
Coarctations de l'aorte

Pauline Parisot

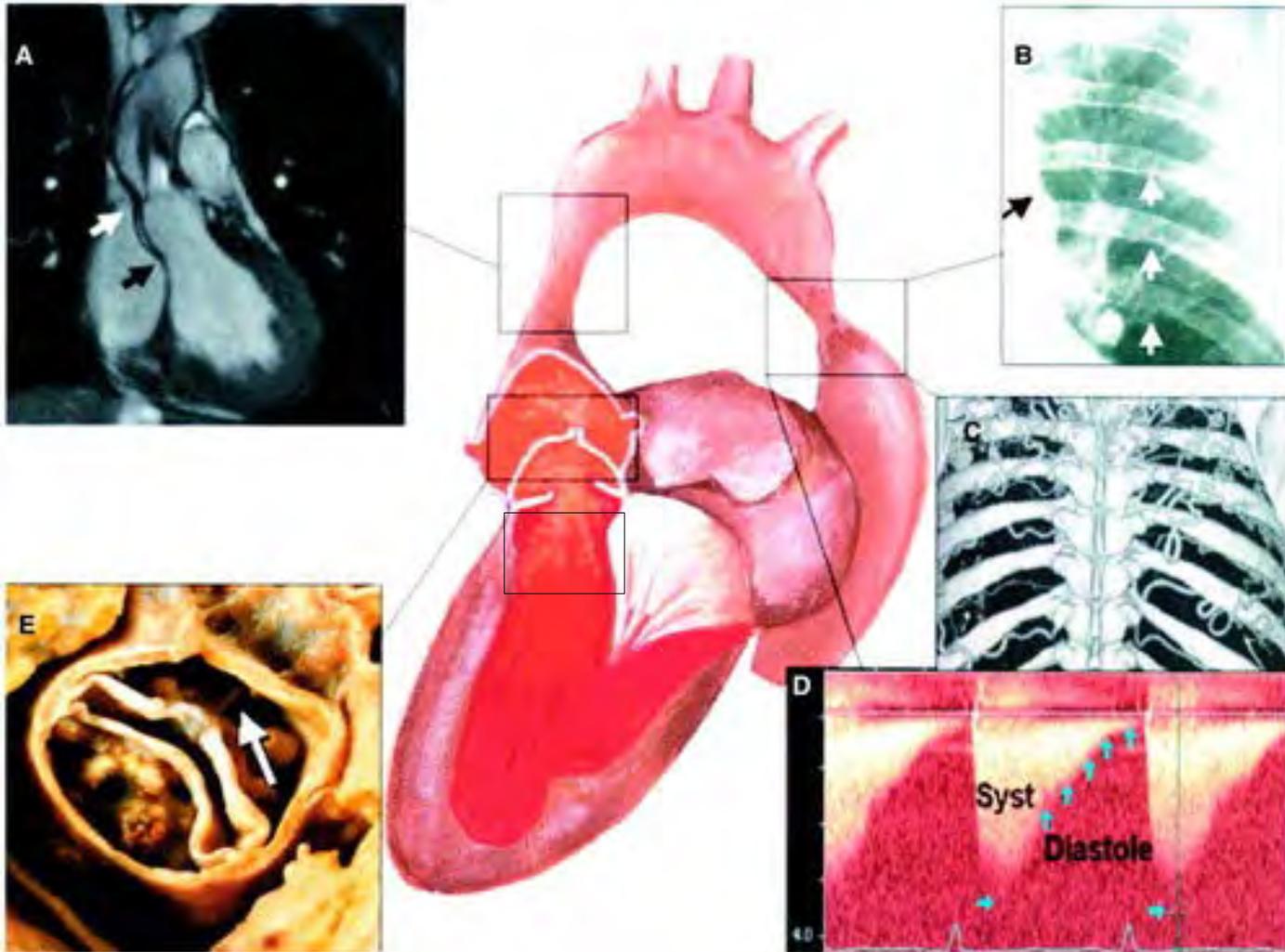
Hôpital Armand Trousseau – Paris

Institut de l'enfant - CCBB

Anatomie de la voie d'éjection gauche



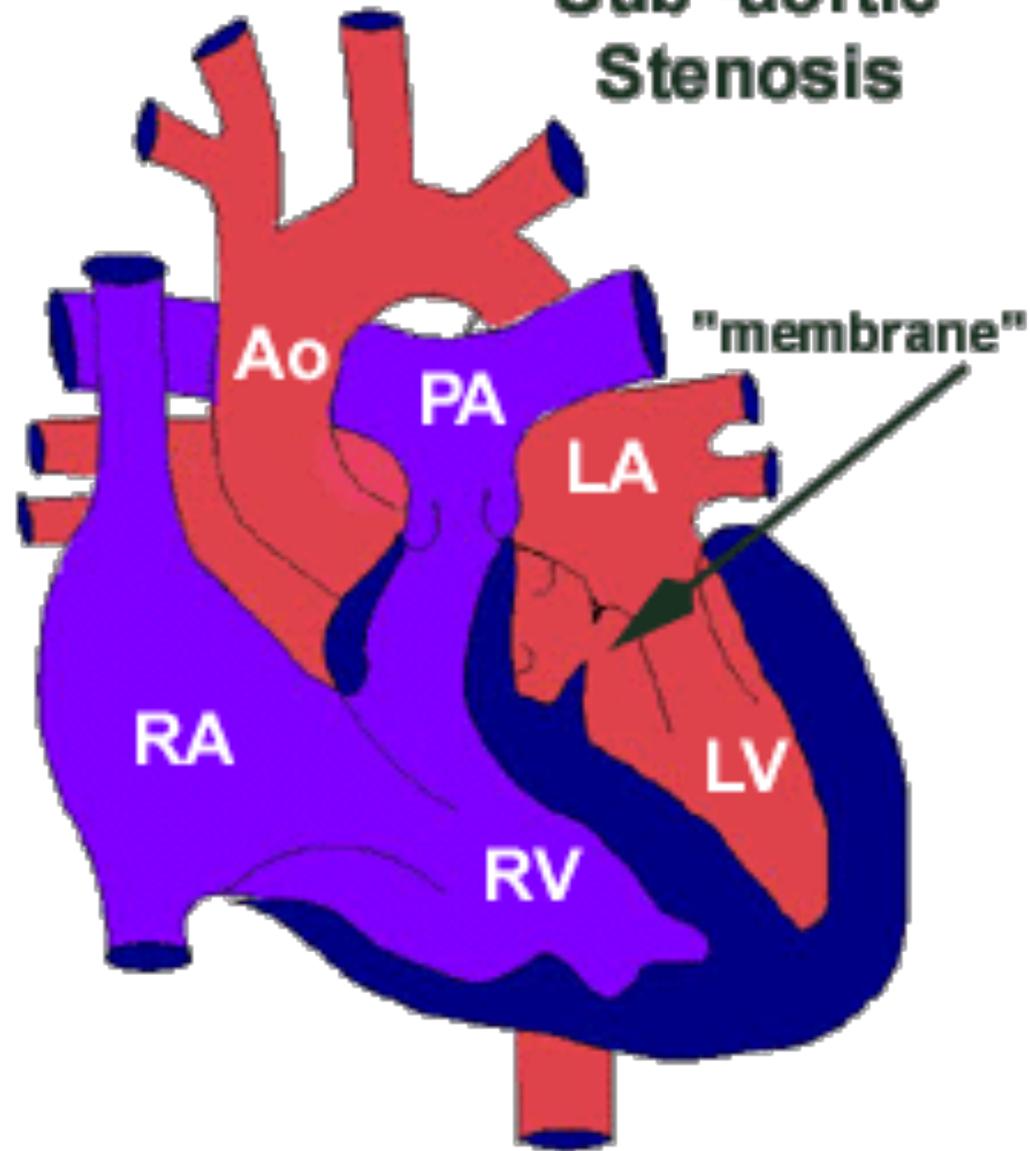
- Coarctation de l'aorte
- Sténose aortique supra valvulaire
- Sténose valvulaire aortique
- Sténose sous valvulaire aortique



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

Sténoses sous valvulaires aortiques

Sub-aortic Stenosis



Sténoses sous valvulaires aortiques

- membrane sous aortique = membrane fibreuse
- obstacle musculaire
- obstacle complexe fibro musculaire
- obstacle localisé (membrane)
- obstacle étendu : tunnel
- plus rarement :
 - bleb mitral
 - insertion anormale de cordage ou pilier mitral

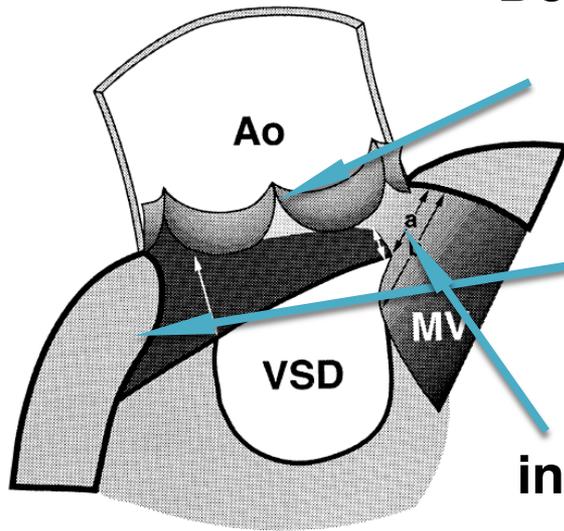
- lésions congénitales ou acquises
- lésions évolutives
- facteurs génétiques
- facteurs anatomiques prédisposants

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

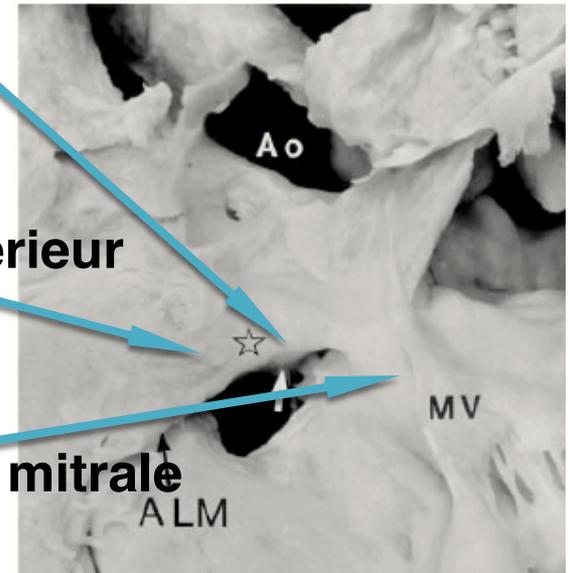
Yuichi Shiokawa, MD, and Anton E. Becker, MD

Déplacement postérieur du septum conal



« bulge » musculaire antérieure

insertion anormale de la valve mitrale



conséquences hémodynamiques

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

Clinique

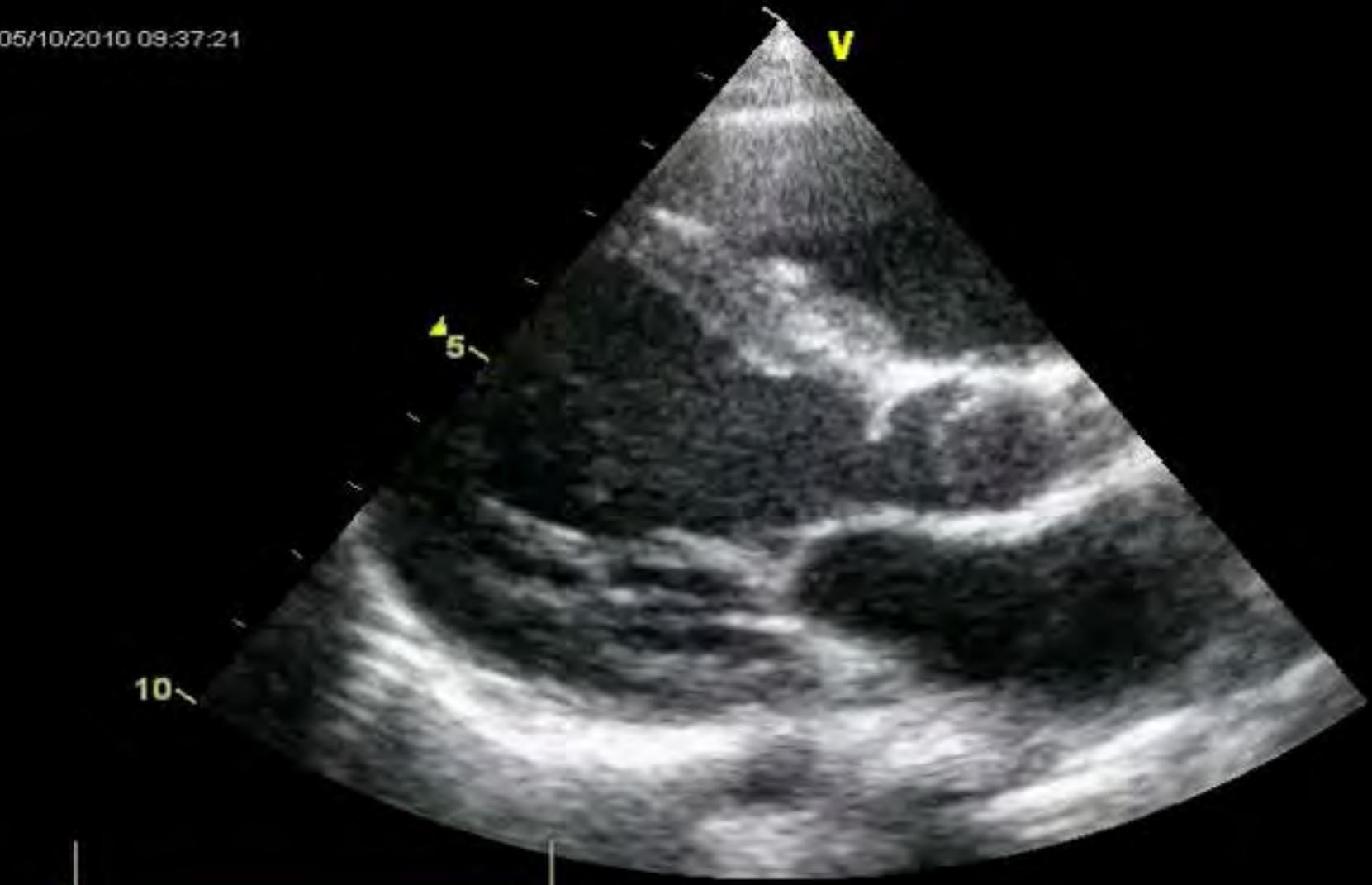
- souffle mesosystolique bord sup D/G du sternum, irradiant vers la carotides
- +/- S protoD si lao associée
- forme sévère : \sphericalangle pouls périphériques, syncope, angor, insuffisance cardiaque

Diagnostic échographique

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

- surveillance évolutivité

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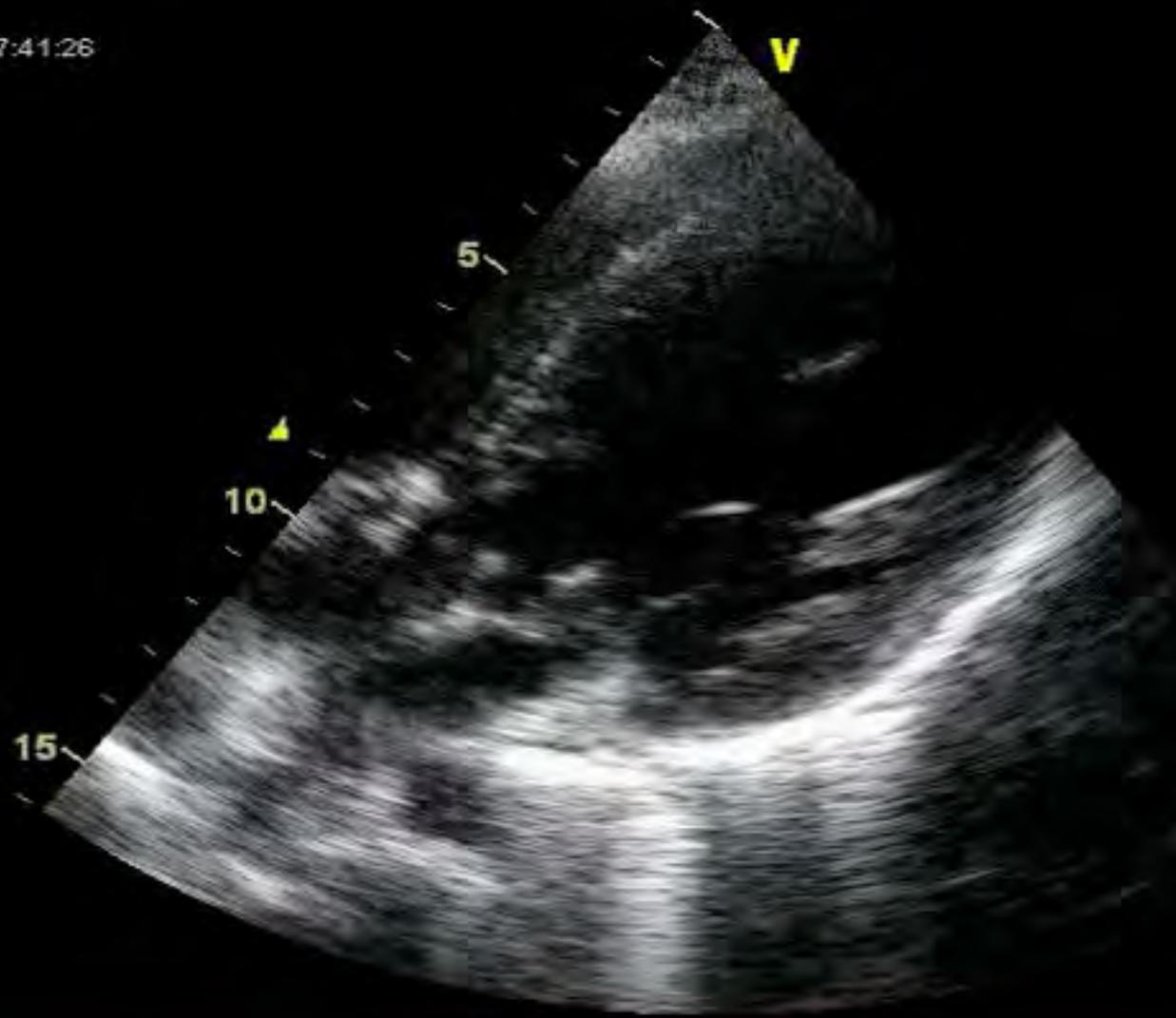


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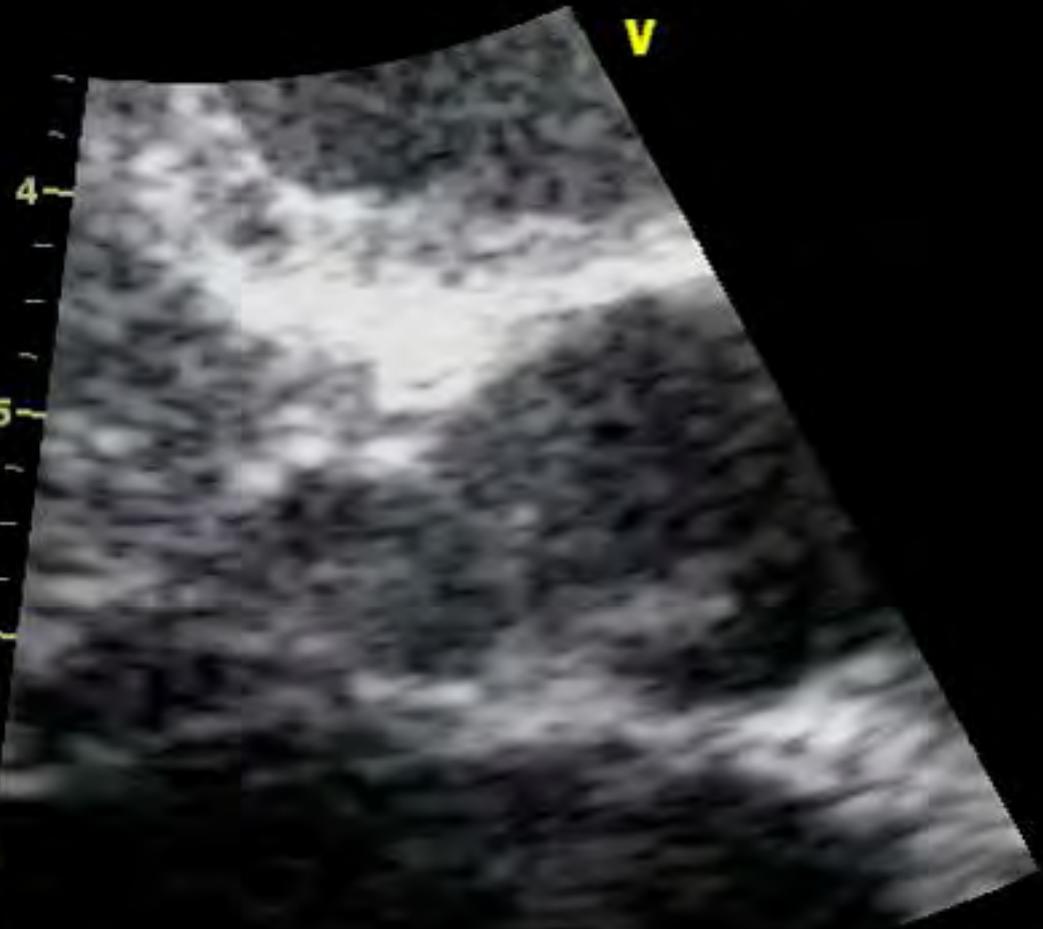
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04/10/2010 16:01:19

V



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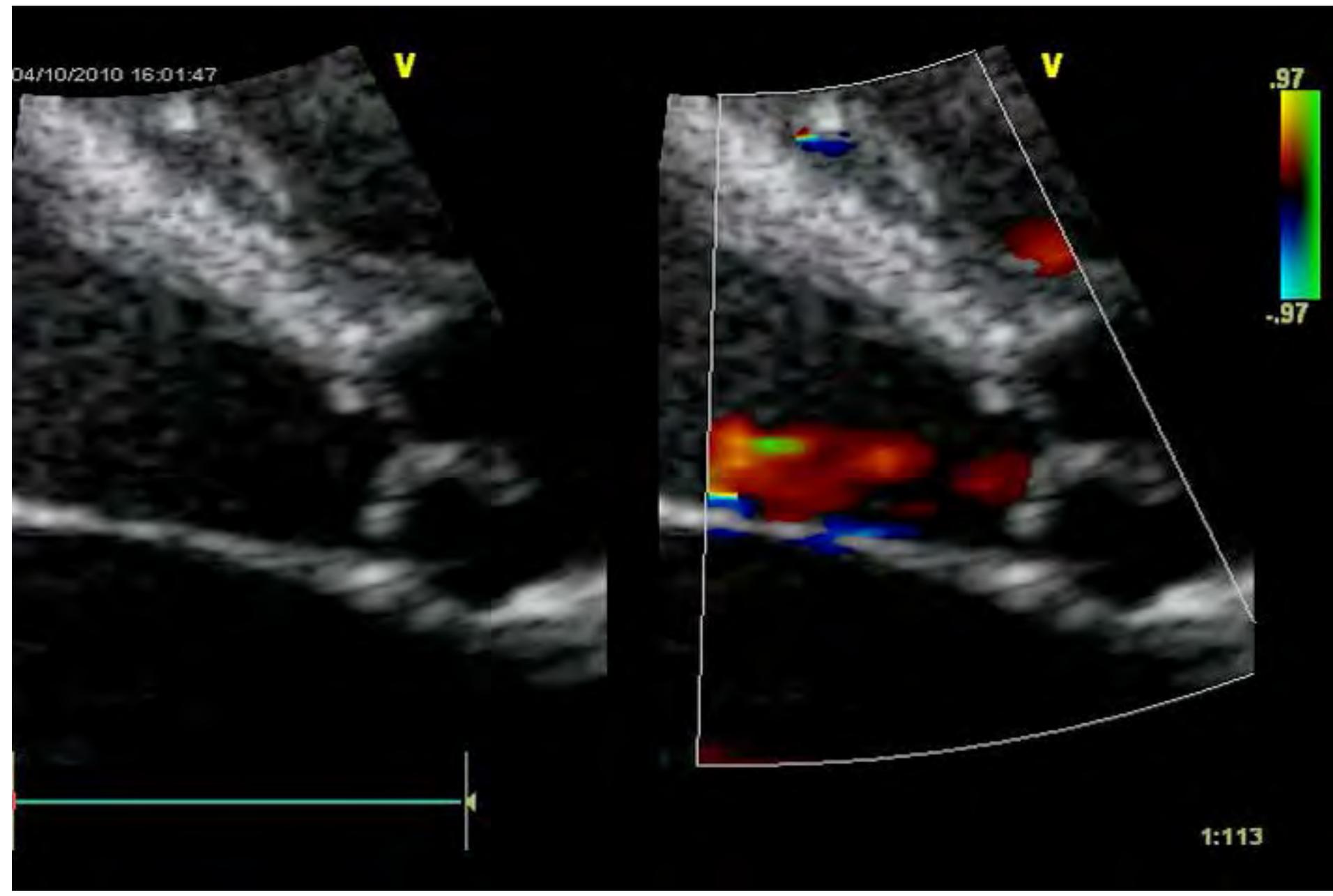
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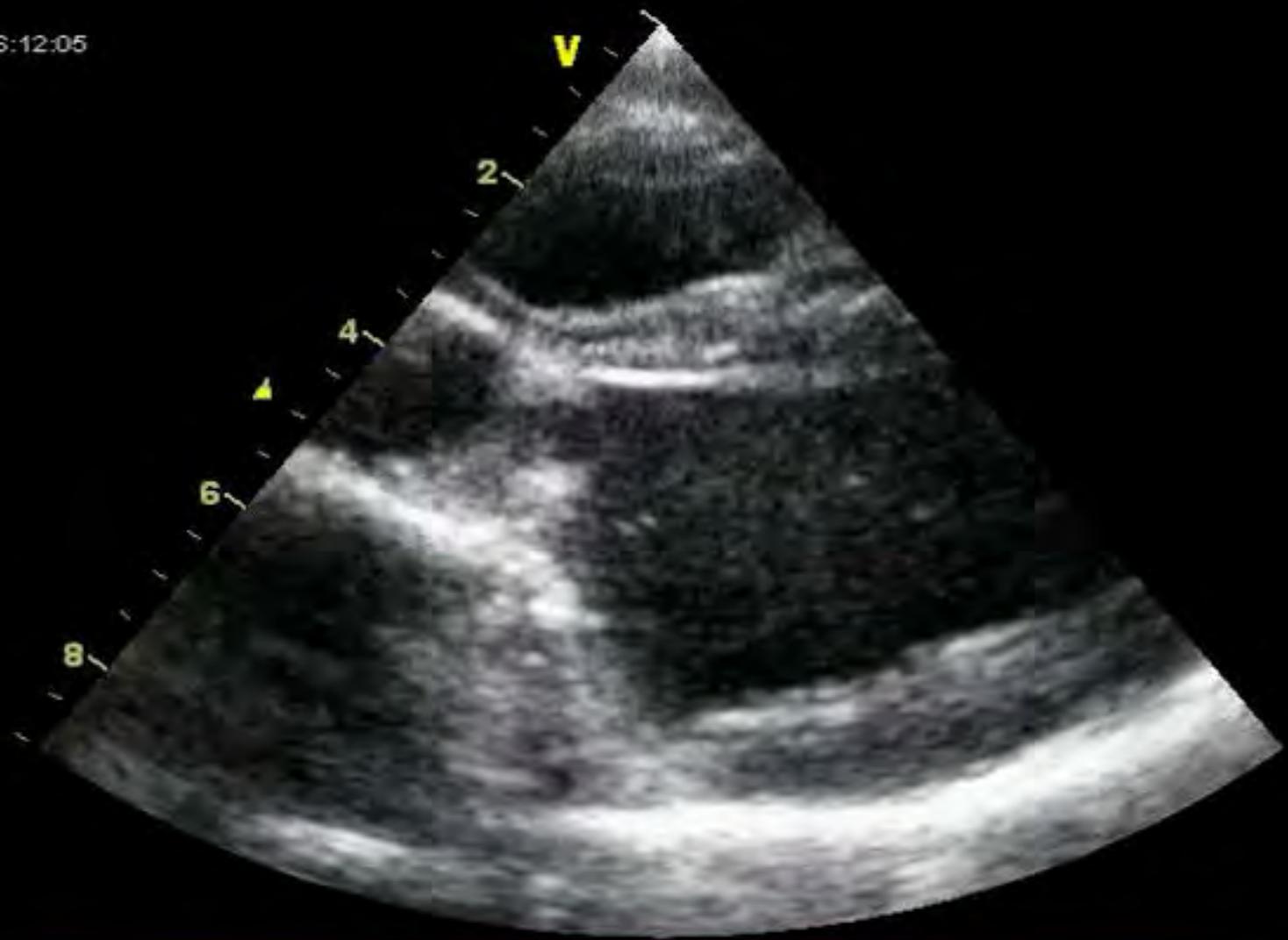
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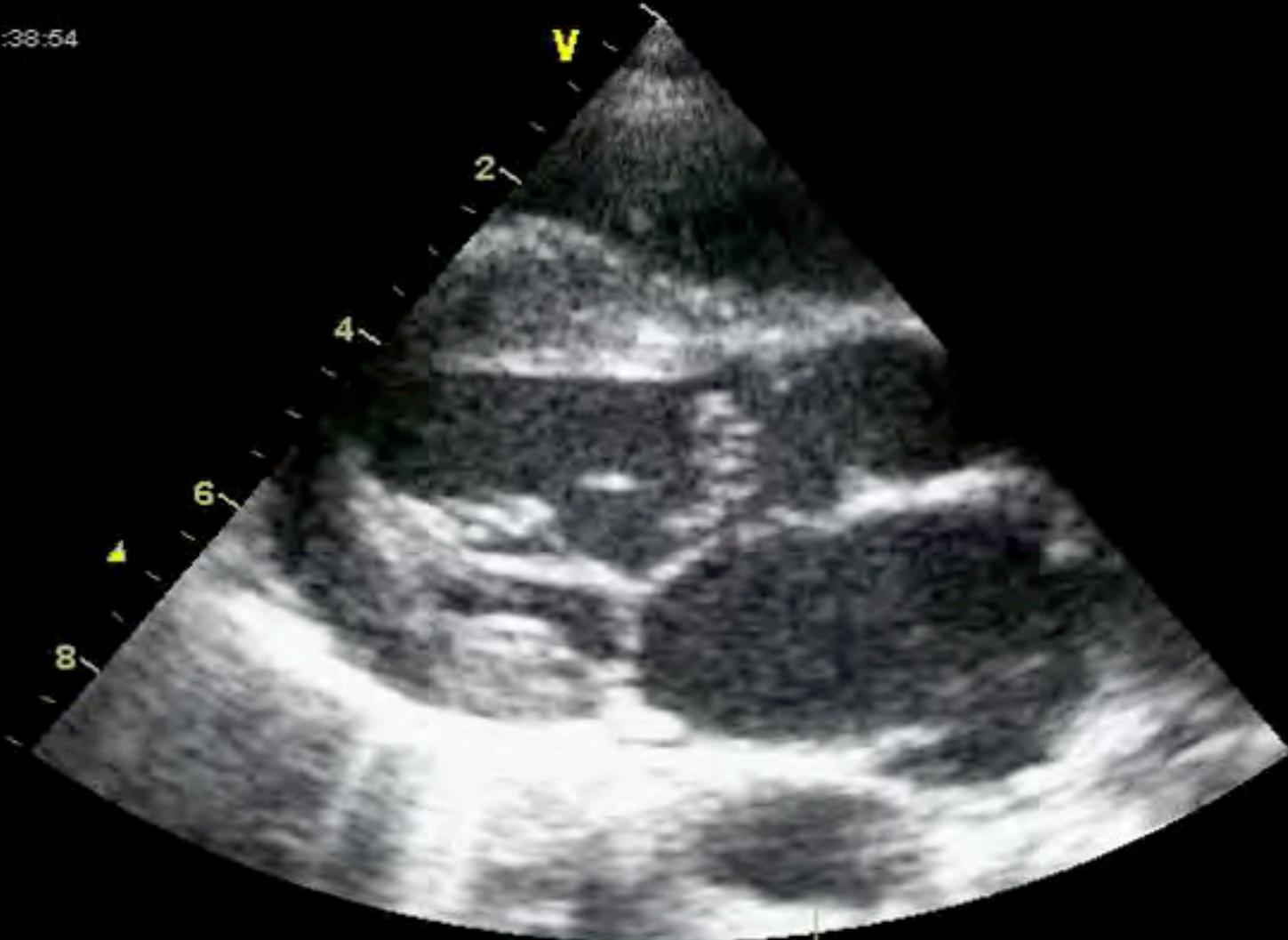


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17/03/2008 14:38:54



26/10/2010 12:11:09

V

5

10



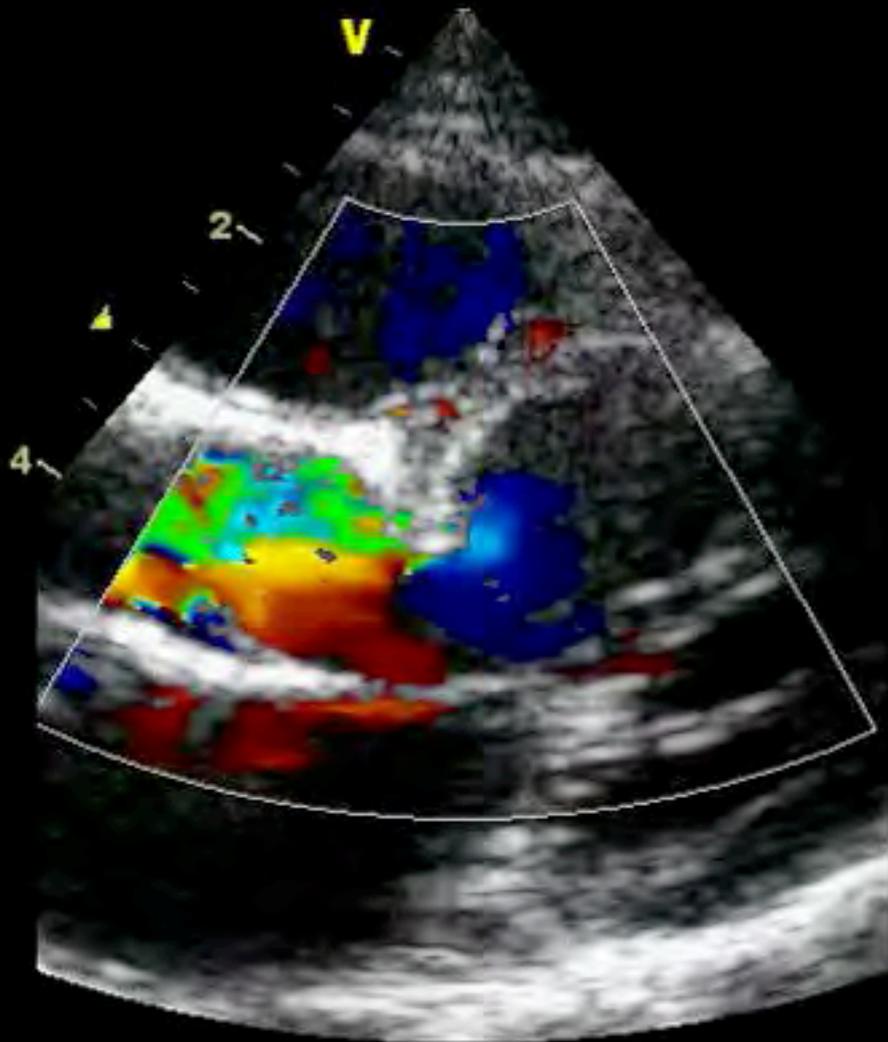
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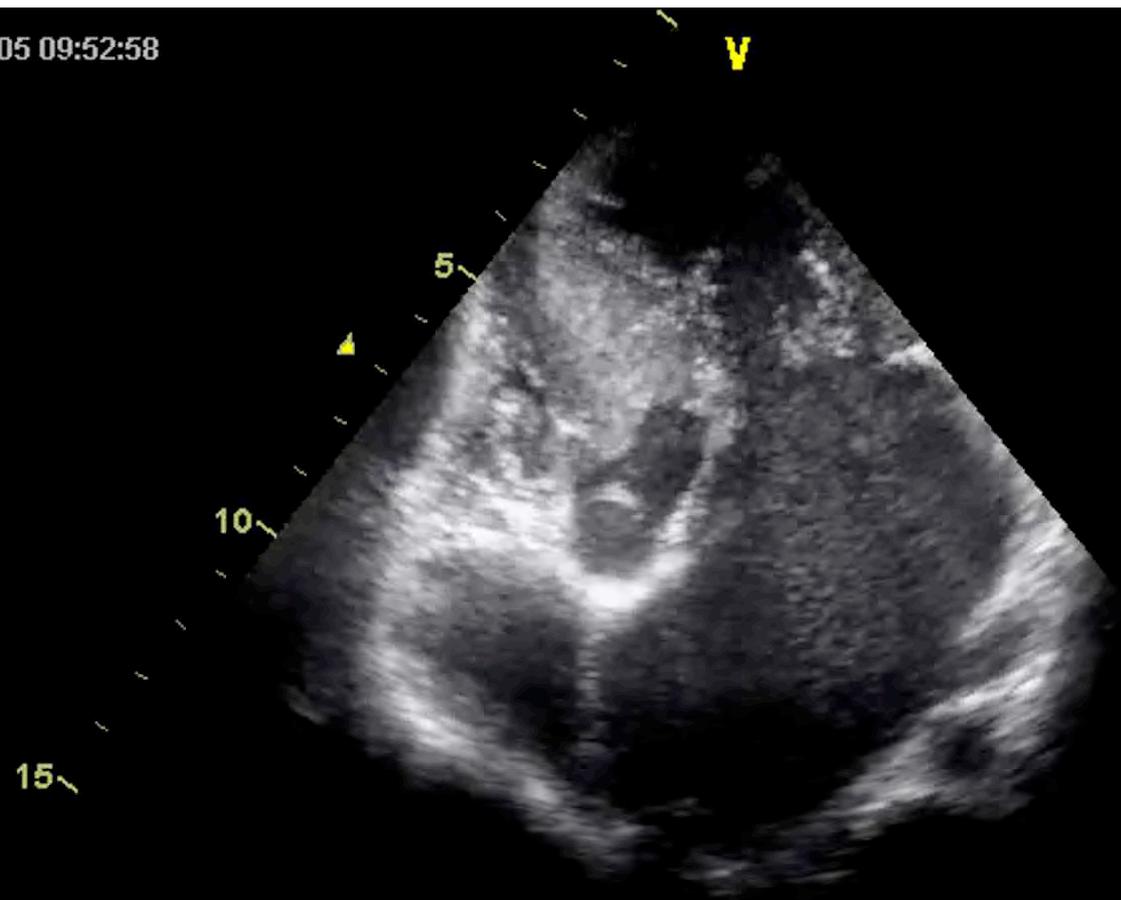
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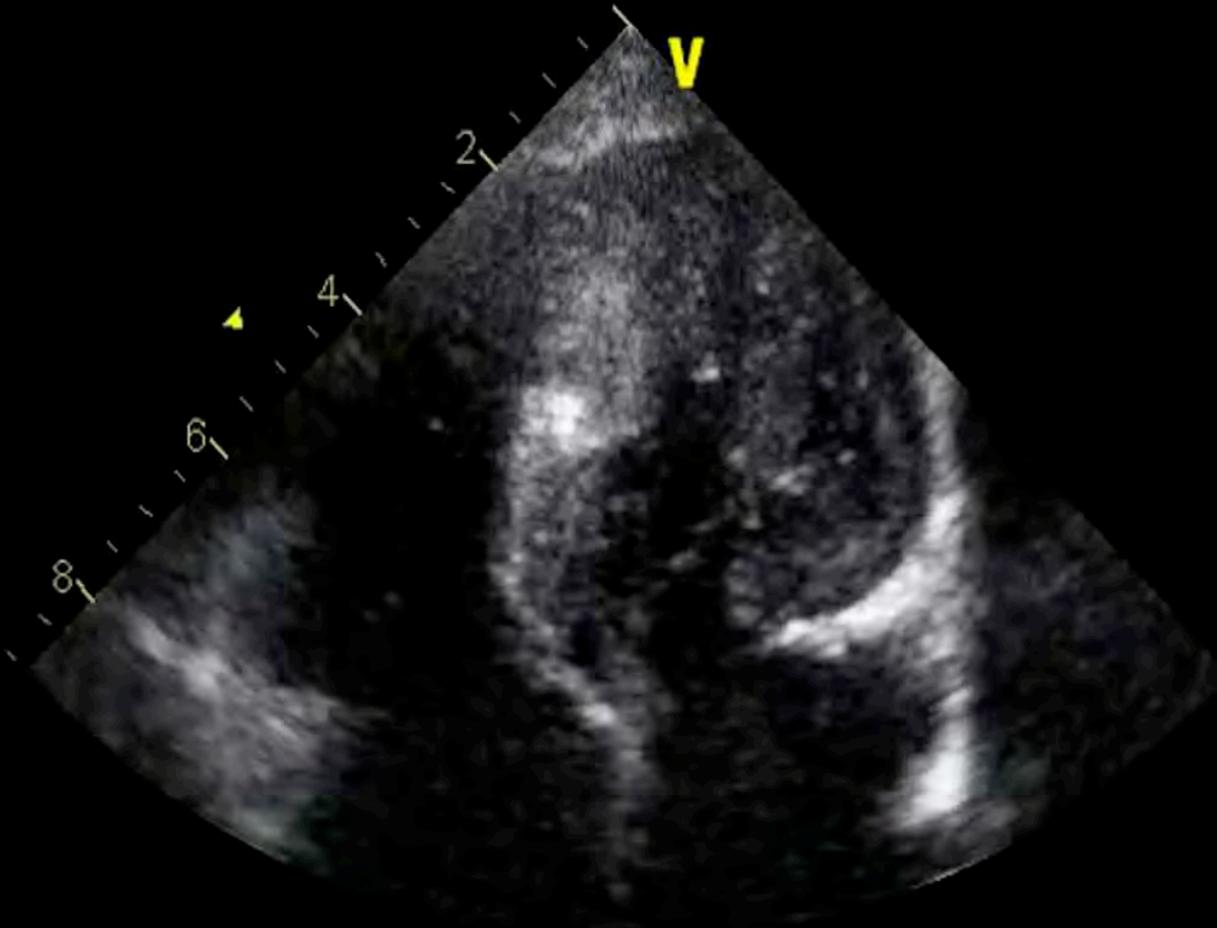


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02/05/2012 11:30:24



Facteurs d' évolutivité

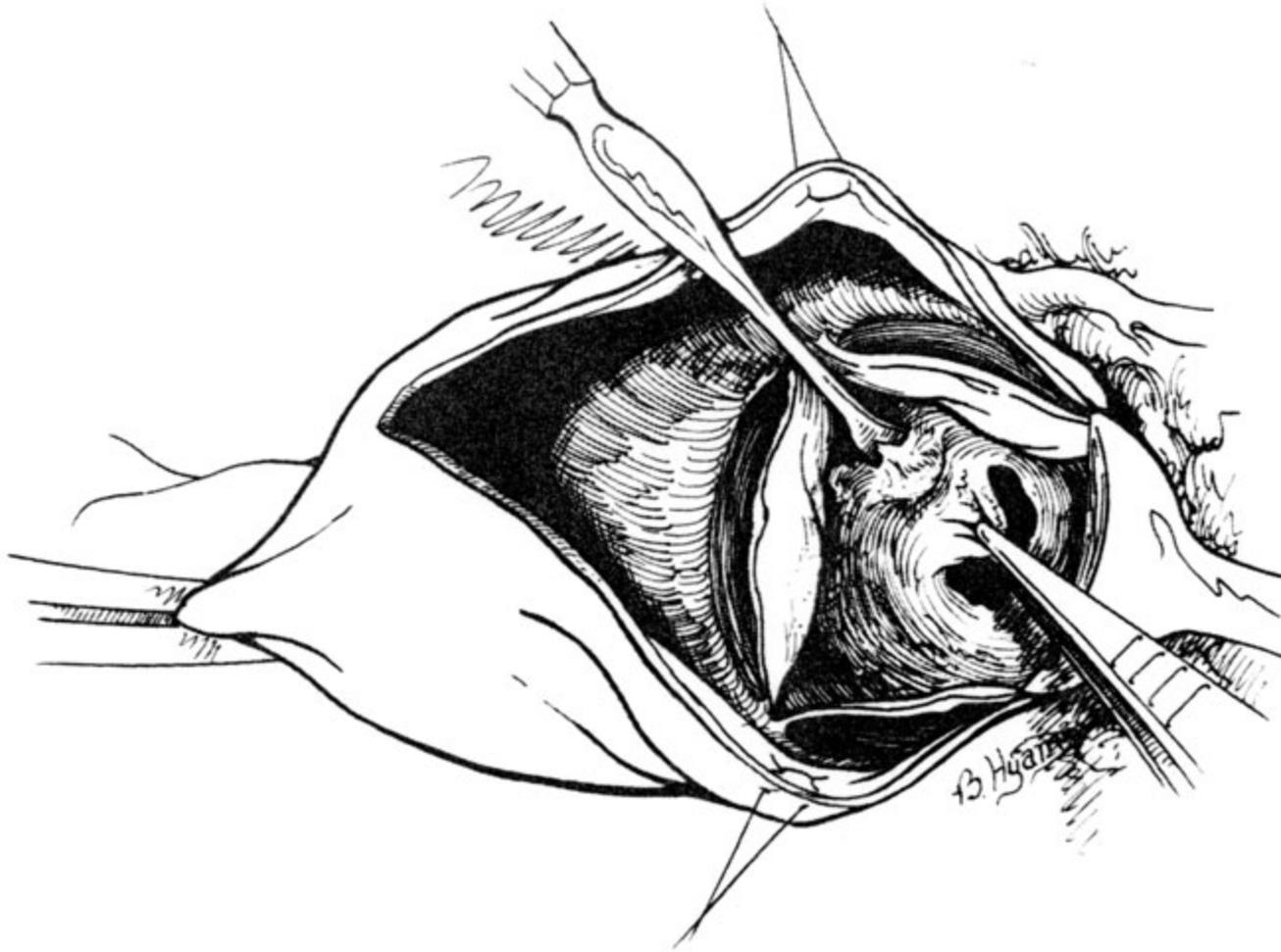
- sévérité obstruction VG
- distance obstacle – valve
- dysplasie valvulaire
- âge au diagnostic

Membrane sous aortique

Traitement : chirurgical

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

Enucléation + myectomie



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 dIAo?

indications préventives

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

. chirurgie préventive non justifiée

. surveillance régulière

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

. chirurgie préventive non justifiée

. surveillance plus rapprochée surtout si

- proximité obstacle-valve
- dysplasie valvulaire

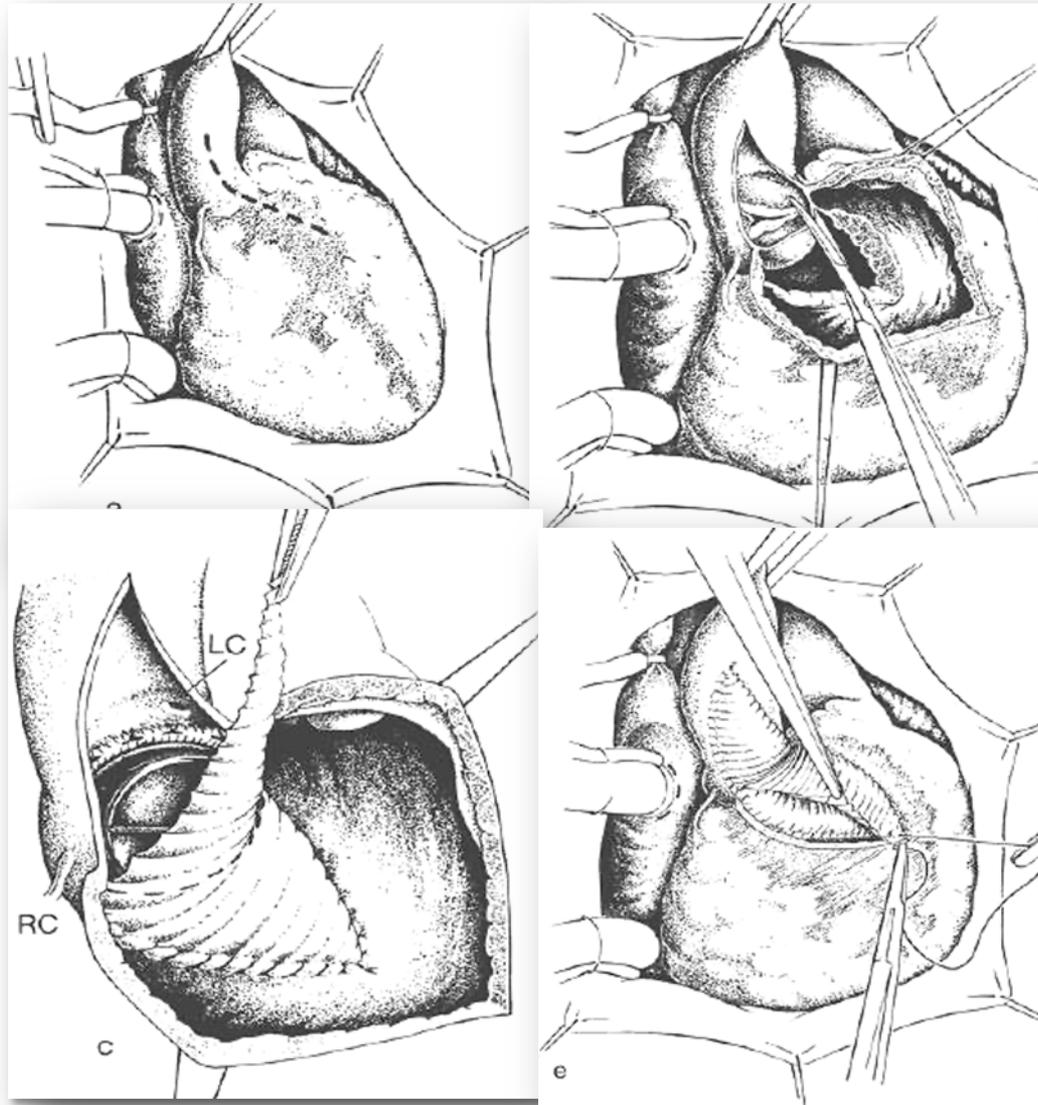
- jeune enfant ou adulte

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

. chirurgie « préventive » sans doute justifiée

. risque évolutif accru

Intervention de Konno

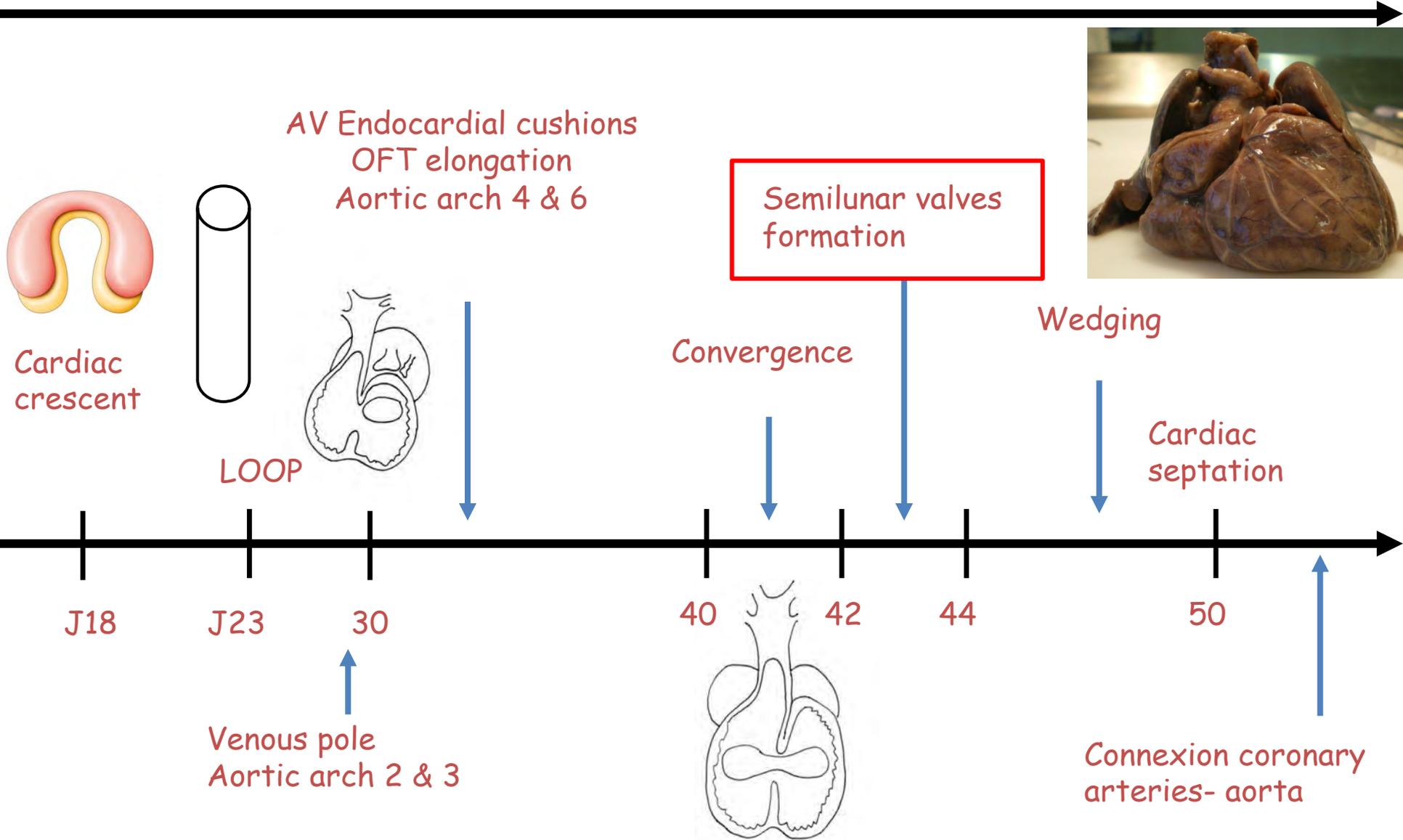


Obstacle sous valvulaire ao complexe et étendu : Konno modifié

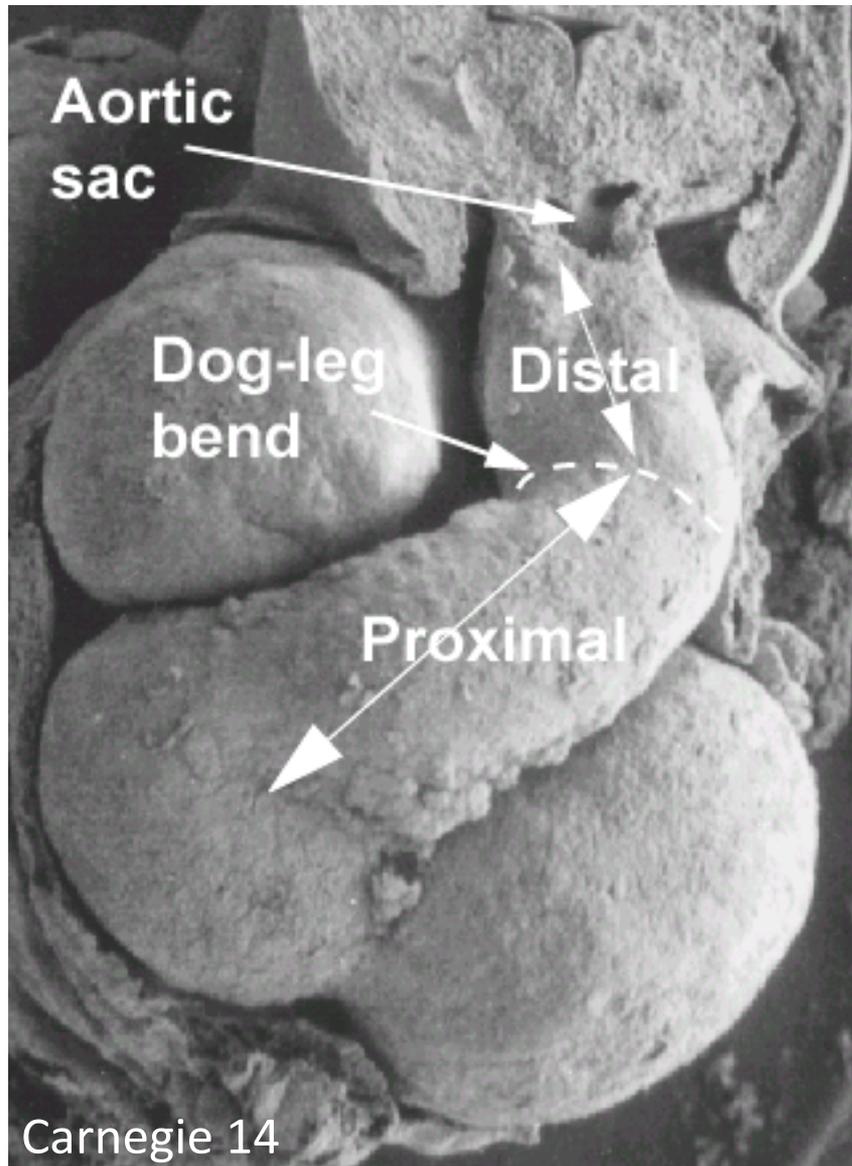
Obstacle sous valvulaire et valvulaire aortique : Konno ou Ross Konno

sténose valvulaire aortique

Développement cardiaque

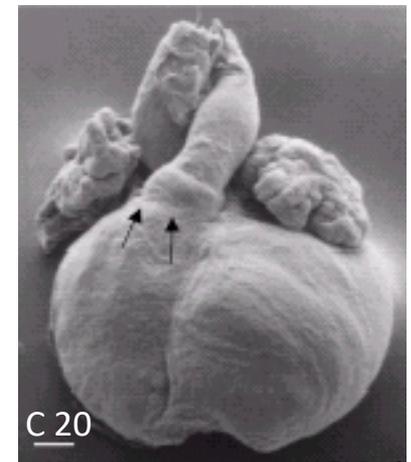
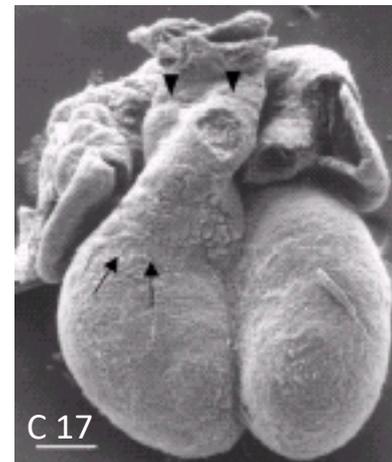


Morphogénèse de la voie efferente



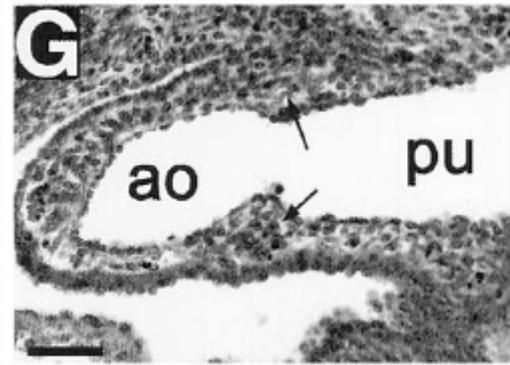
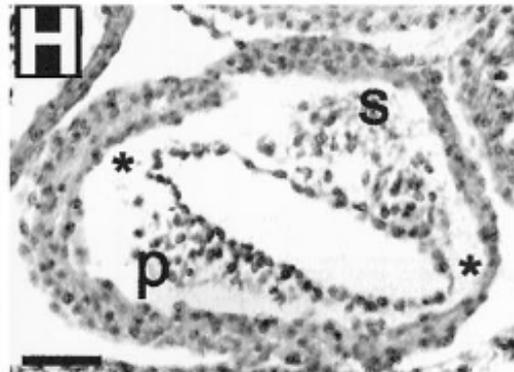
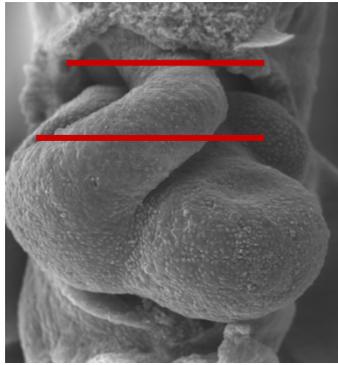
Voie efferente proximale

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



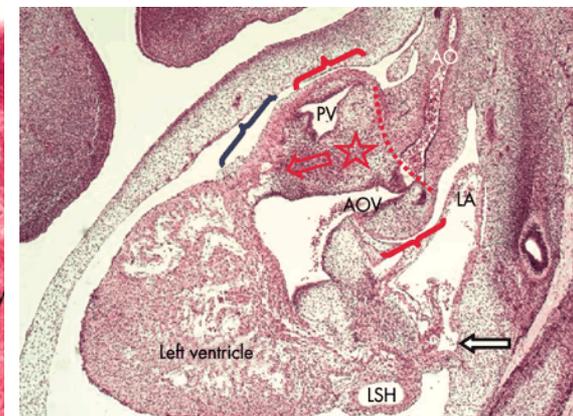
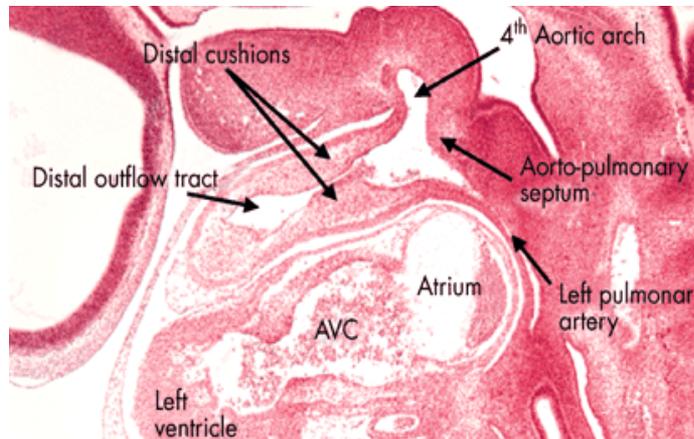
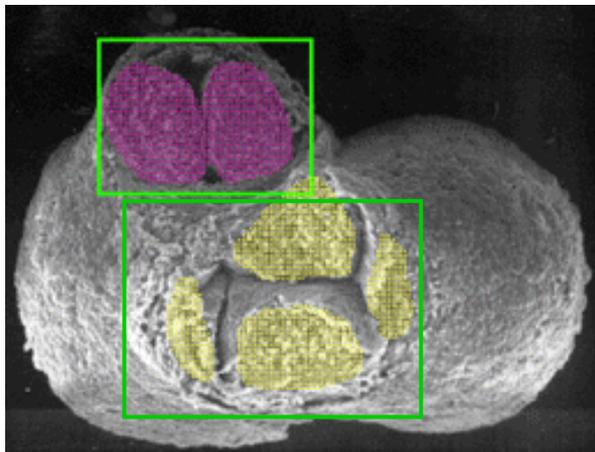
Coussins endocardiques

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



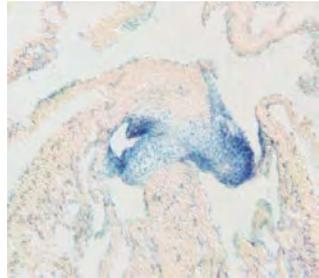
Costell et al., 2002

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 epiteliomesenchymateuse

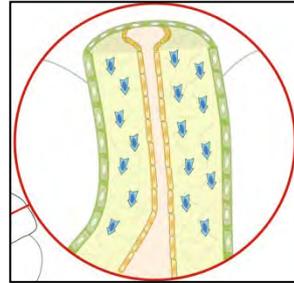


Anderson et al., 2003

Endocardial cushions contribute to the OFT remodeling



Tie2cre/R26R



Neural crest



Cushions

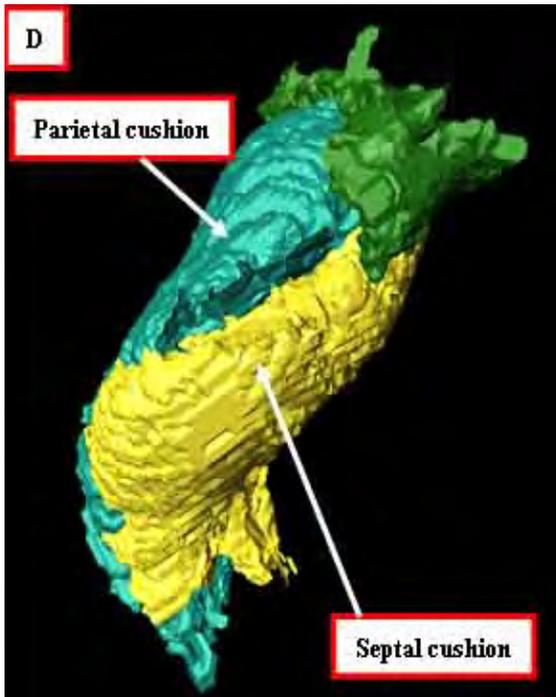


Myocardium

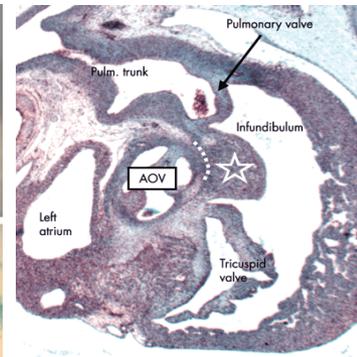
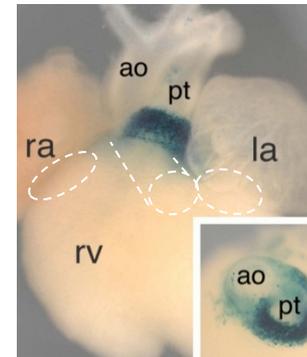
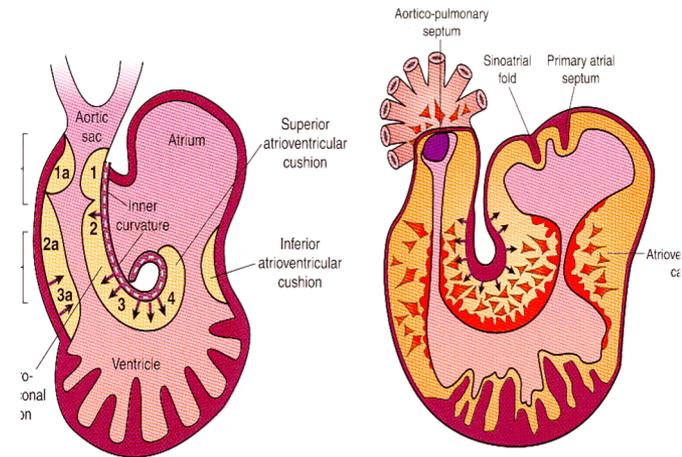
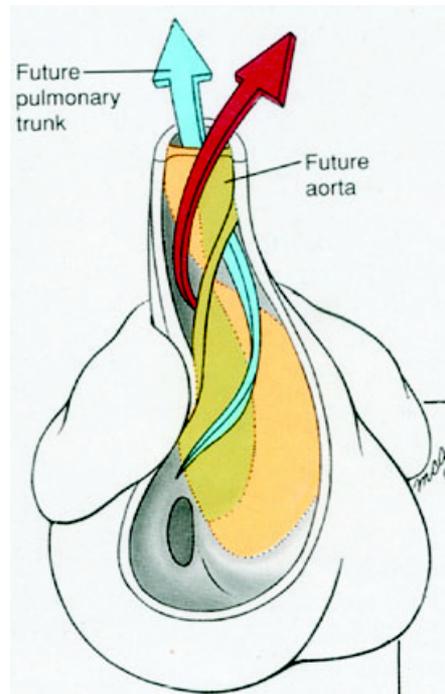
- Endocardial cushions of the proximal outflow tract and the AVC-derived from the endocardium, due to signals from myocardium
- Endocardial cushions of the distal outflow tract - contribution from cardiac neural crest

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)

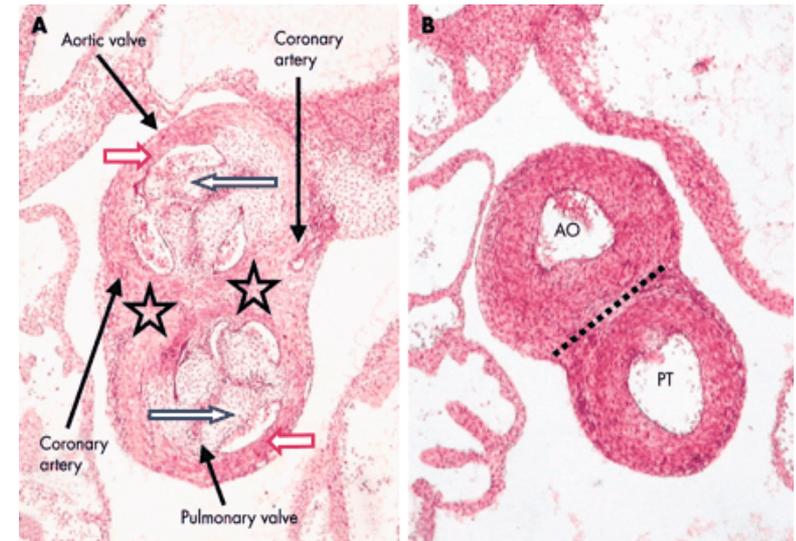
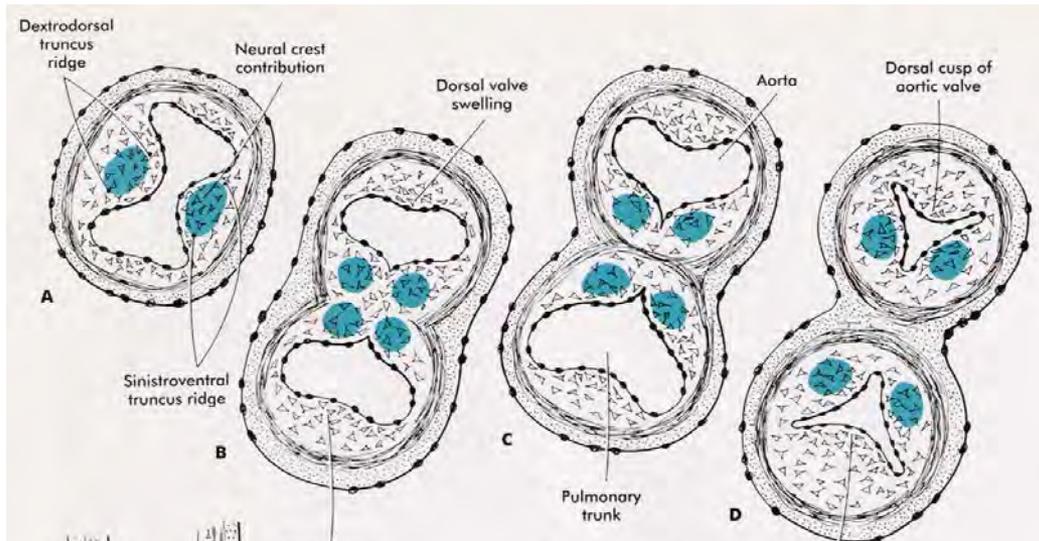


Courtesy B. Anderson



Semilunar valve formation

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



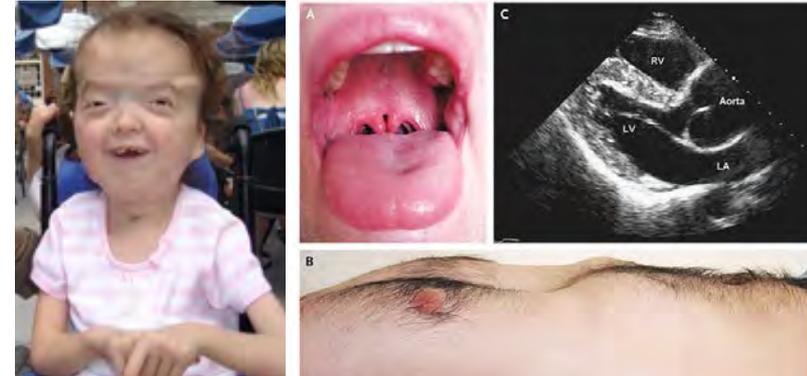
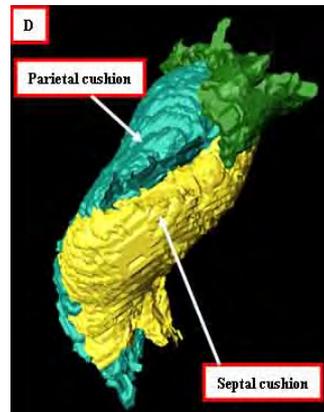
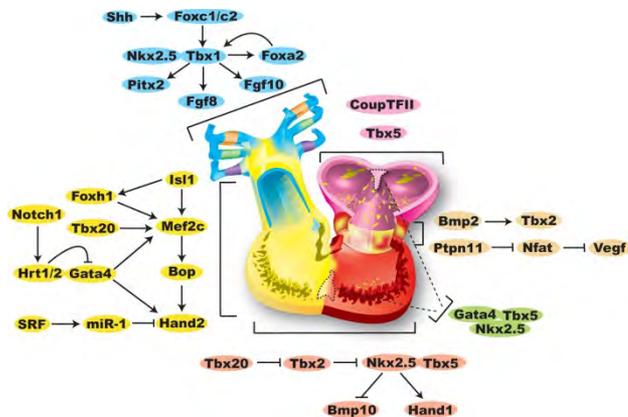
- Differentiation, apoptosis and ECM remodeling: formation of thin protruding leaflet valves composed of endocardial cells and ECM
- Cardiac neural crest cells reach the OFT cushions and contribute to the formation of the aortic and pulmonary valves.
- The wall of the distal OFT change from myocardium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

Bicuspid Aortic valve and Aortopathy

- **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)

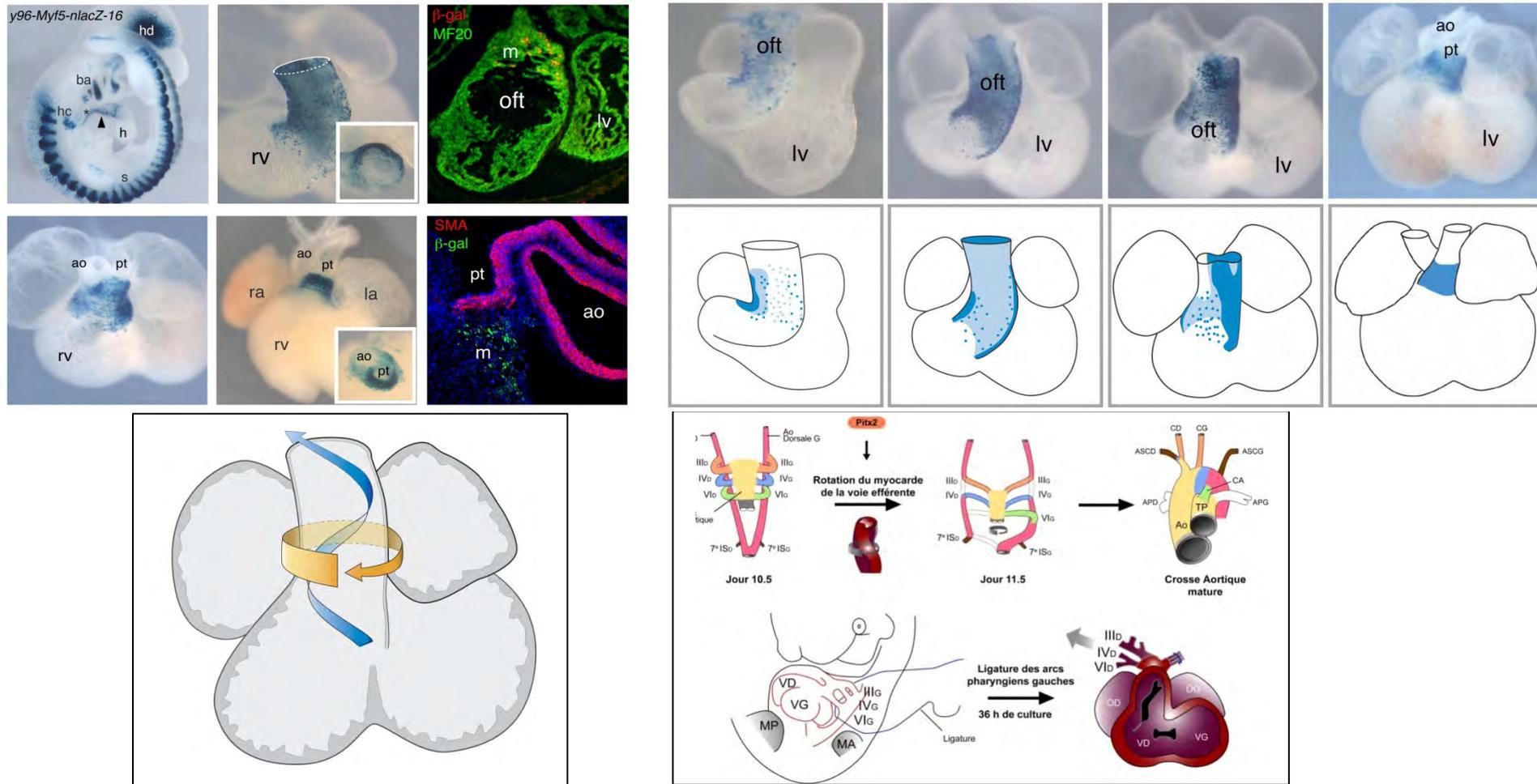
Development of BAV and Aortopathy

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



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

Does a link between fetal heart blood flow and gene expression exist?



Haemodynamics determined by a genetic programme govern asymmetric development of the aortic arch

Bajolle et al., 2006; Yashiro et al., Nature 2007

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

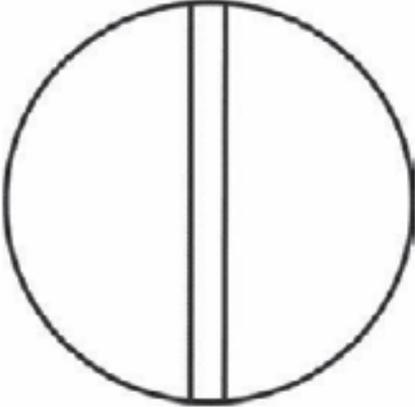
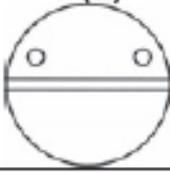
These LHO anomalies belonged to a same embryological

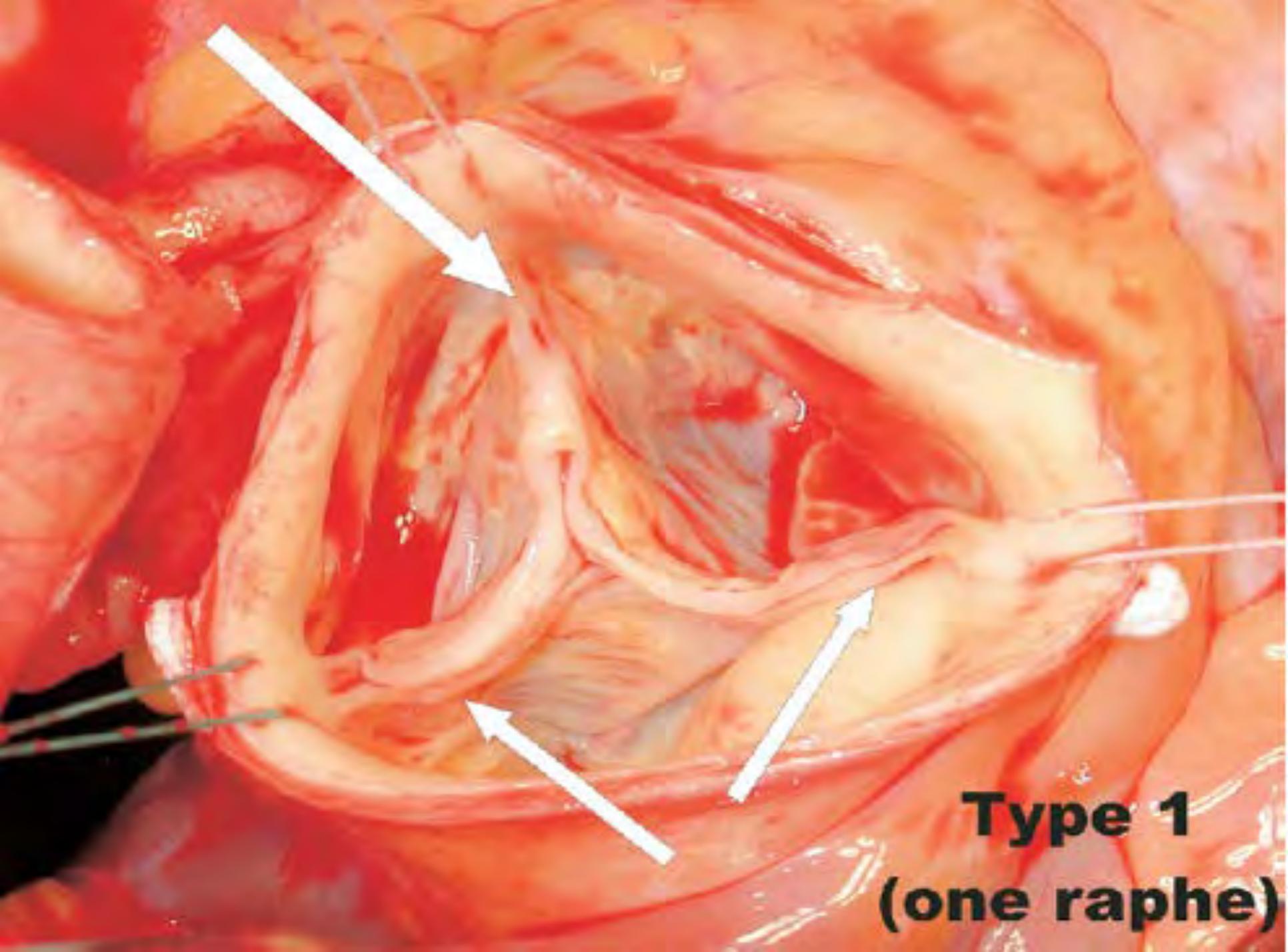
- The development of heart chambers is related to the pattern of the combined foetal blood flow that passes through them (Abraham Rudolph)
- recurrences of different severity within the same family
- prenatal progression of obstructive left heart diseases
- identification of the same mutation in *NOTCH1*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

Bicuspidie aortique

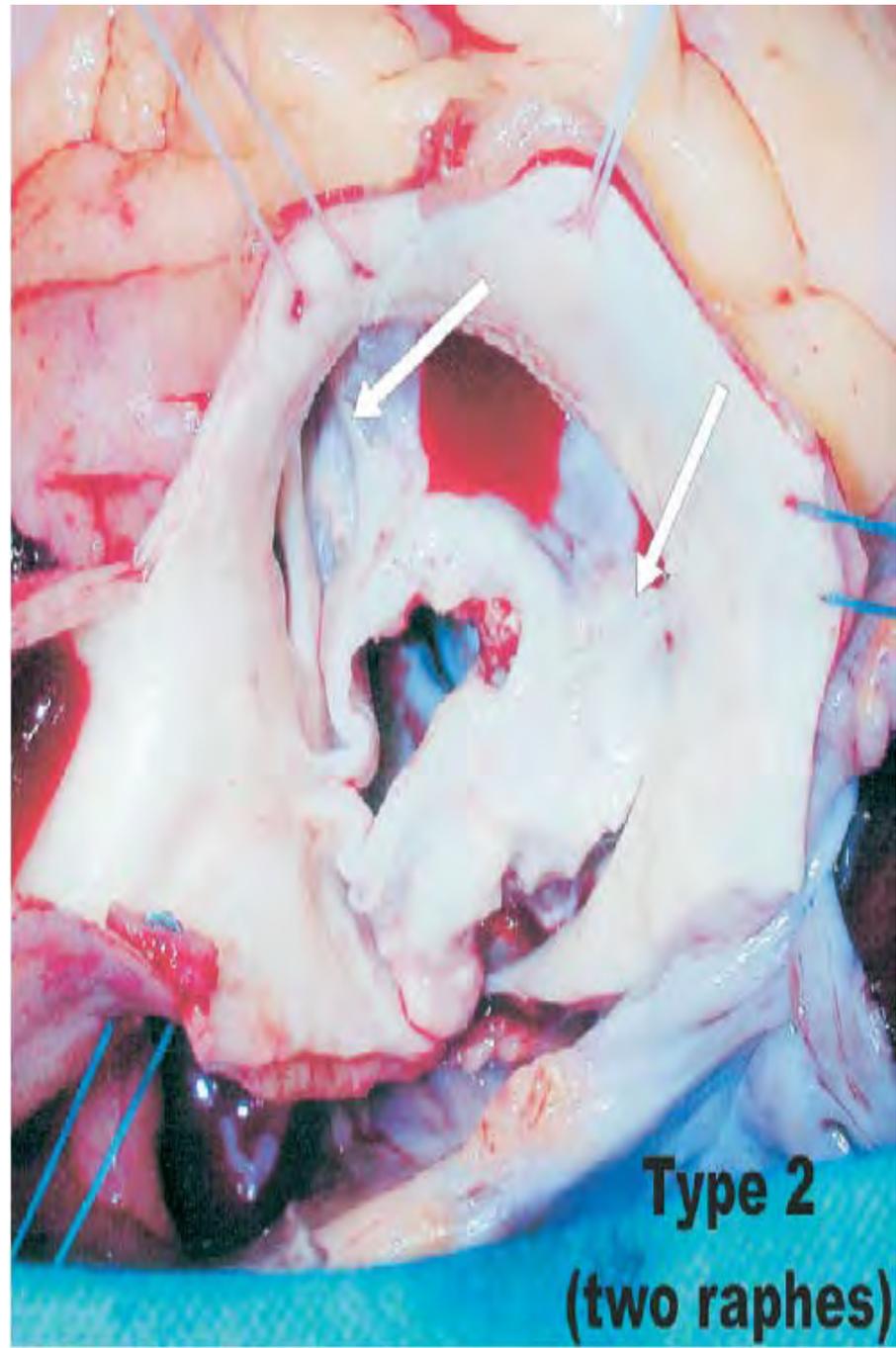
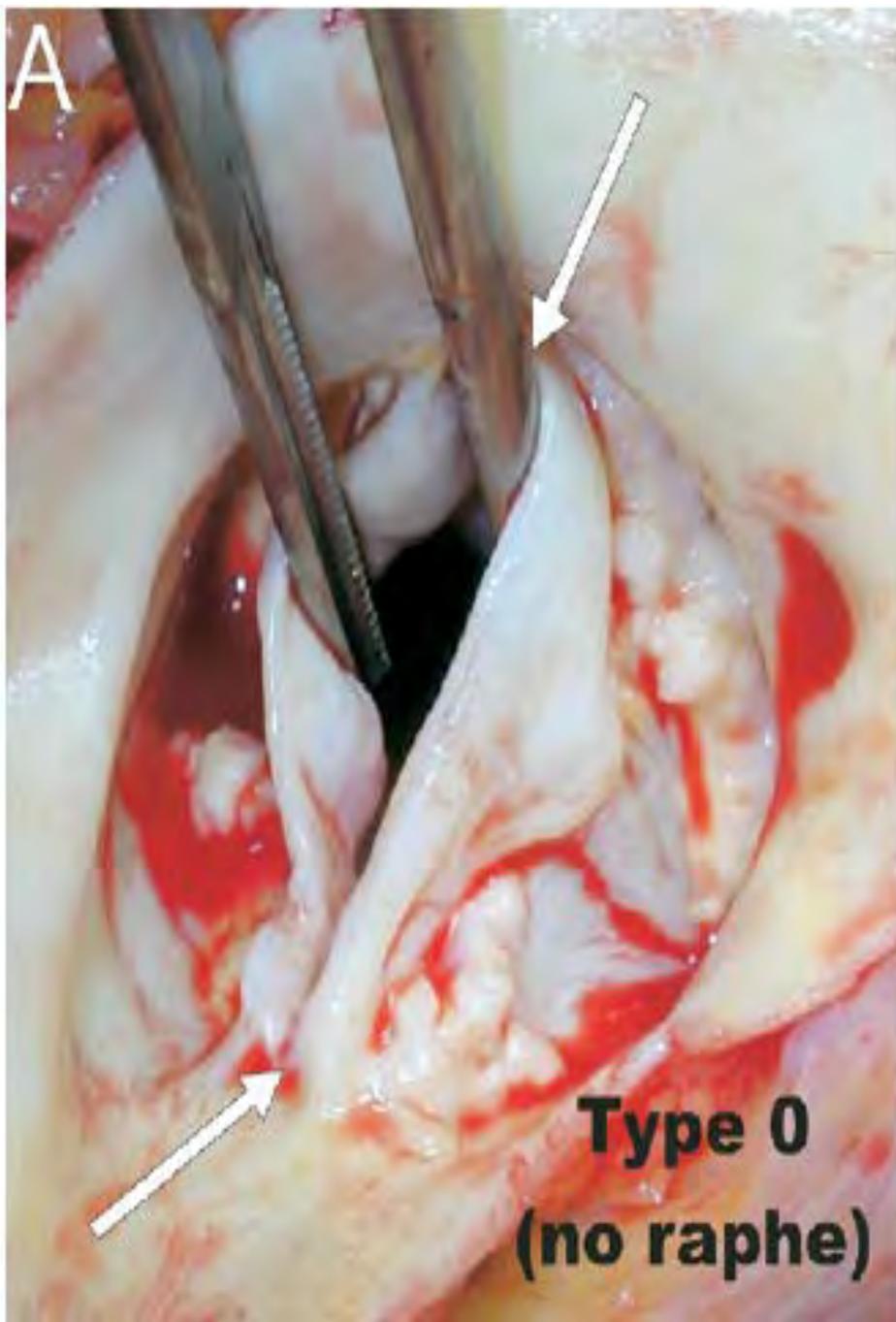
- malformation la plus fréquente incidence estimée 1%
- prévalence chez les NN : 0.46%
- forme familiale
- hérédité autosomique dominante pénétrance variable (prévalence intrafamiliale 37%)
- hétérogénéité génétique; NOTCH1
- risque accru d'obstacle G dans les apparentés de 1^{er} degré
- screening echo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

CLASSIFICATION MORPHOLOGIQUE

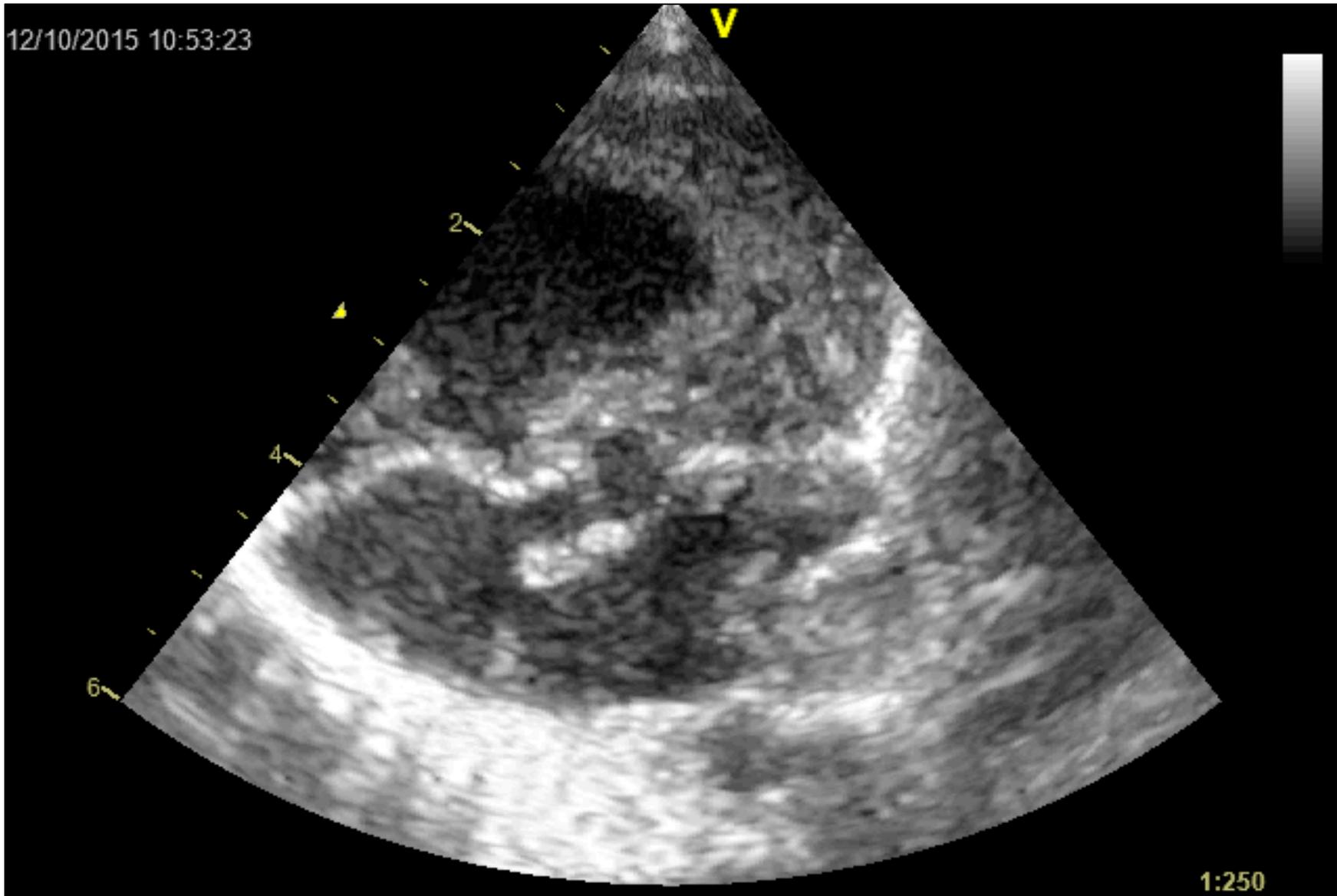
<u>main category:</u> number of raphes	0 raphe - Type 0		1 raphe - Type 1			2 raphes - Type 2
	 21 (7)		 269 (88)			 14 (5)
<u>1. subcategory:</u> spatial position of cusps in Type 0 and raphes in Types 1 and 2	lat 13 (4) 	ap 7 (2) 	L - R 216 (71) 	R - N 45 (15) 	N - L 8 (3) 	L - R / R - N 14 (5) 
<u>2. subcategory:</u> V A L V U L A R F U N C T I O N						
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)
B (I + S)		1 (0.3)	15 (5)	7 (2)	2 (1)	2 (1)
No			3 (1)	1 (0.3)		



Type 1
(one raphe)

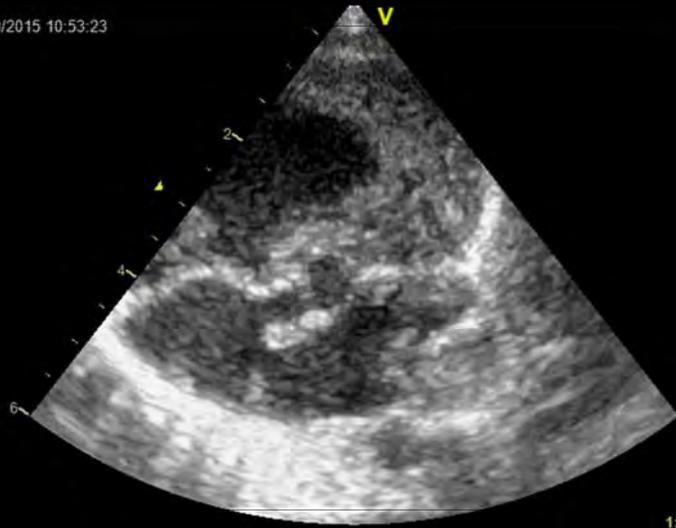


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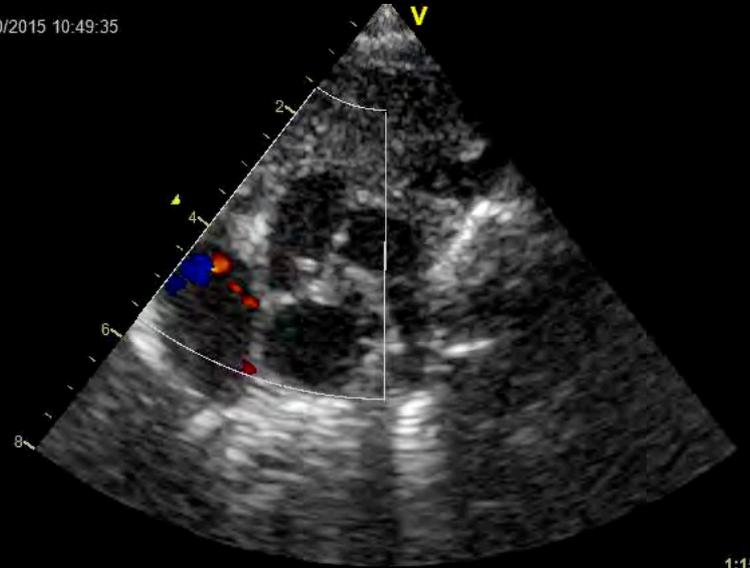
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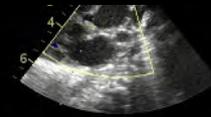


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STRATEGIE CHIRURGICALE

Annular dilatation

Annuloplasty

Commissural plication (7 patients)

Simple annuloplasty (6 patients)

Reinforced annuloplasty (2 patients)

Redundant leaflet tissue

Leaflet plication (7 patients)

Midleaflet excision (11 patients)

Restricted leaflet motion \pm deficient leaflet tissue

Commissurotomy (13 patients)

Leaflet shaving (4 patients)

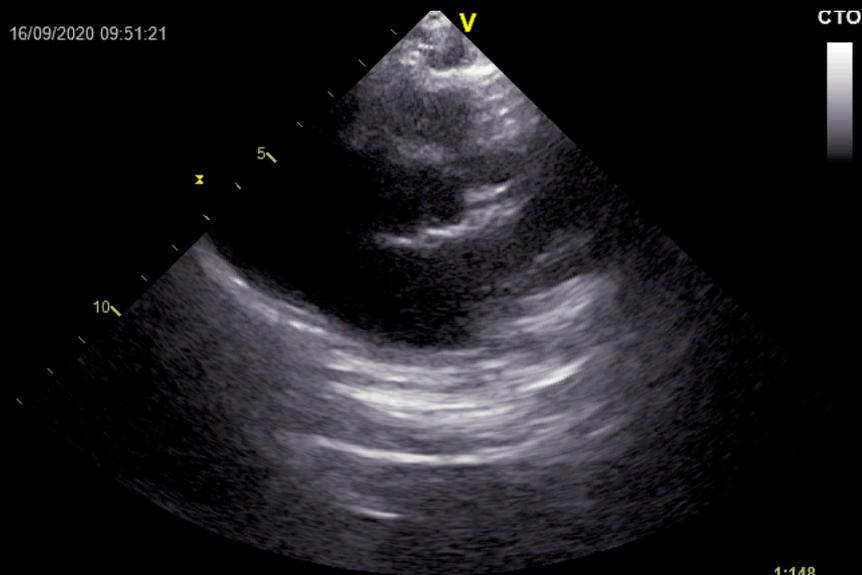
Repair of leaflet perforation (2 patients)

Augmentation commissurorrhaphy (2 patients)

Leaflet extension (19 patients)

Leaflet replacement (1 patient)

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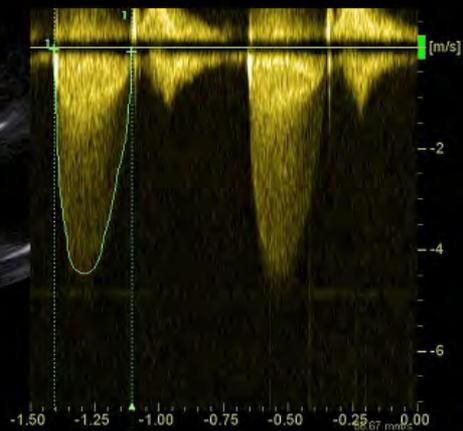


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1 VA Vmax	4.47 m/s
VA Vmoy	3.35 m/s
VA GDmax	80.08 mmHg
VA GDmoy	50.16 mmHg
VA Env.Ti	301 ms
VA ITV	100.76 cm
FC	199.57 BPM

CTO



sténose valvulaire aortique critique néonatale

- souvent valve monocuspide
- VG dilaté fibroelastotique
- fonction VG, pronostic?
- ductodépendance
- stratégie uni ou biventriculaire

Freq: 10.0 MHz @ 0.00 Hz

V

L



2

4

6



100%



sténose aortique néonatale

Lésions valvulaire

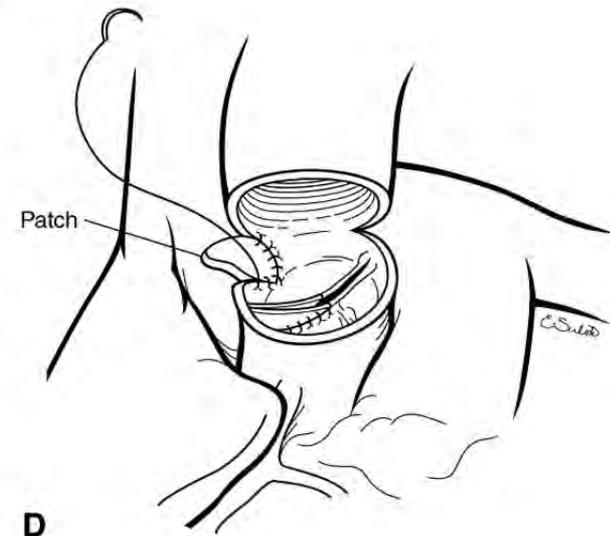
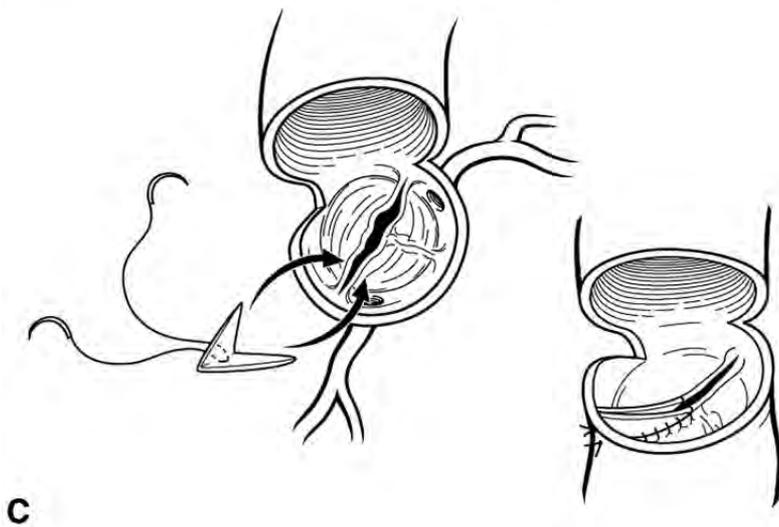
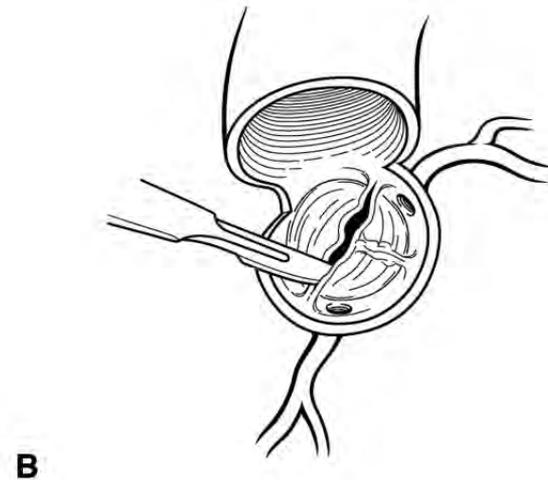
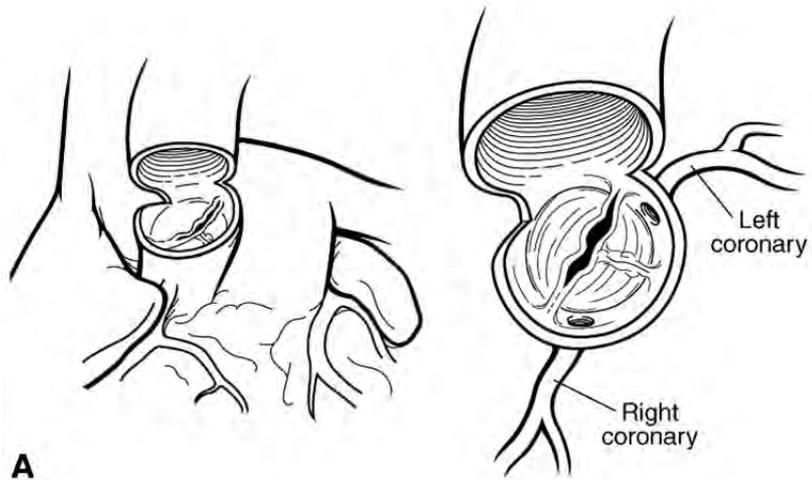
- valve bicuspide ou monocuspide
- rarement tricuspide
- une seule commissure normale
- feuillets dysplasiques et épais
- tissu nodulaire fibreux
- taille de l'anneau : +/- hypoplasique

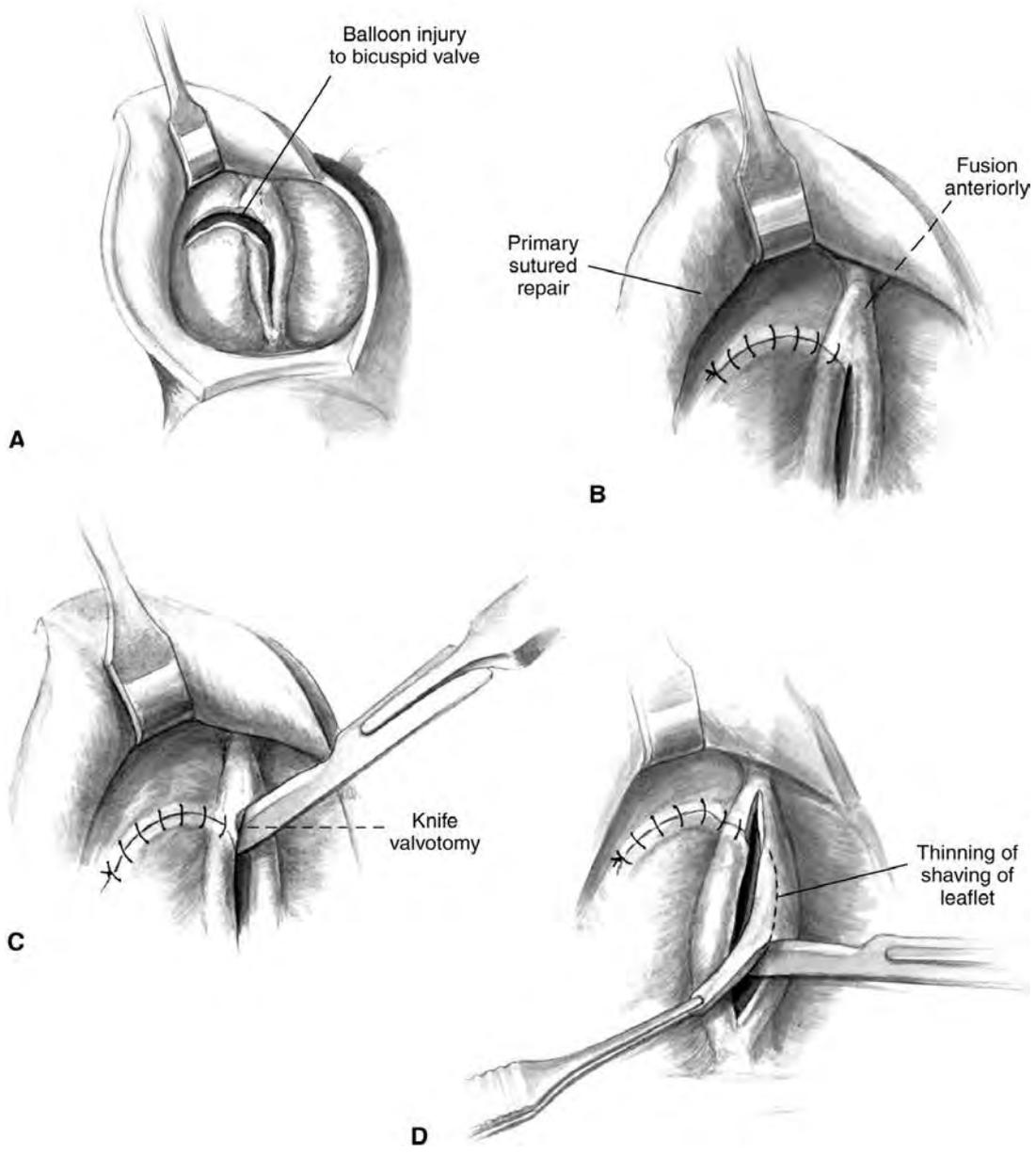
- Lésions associées : obtacle ss ao, supraalulaire ao, anomalie mitrale, Coa

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





Indications

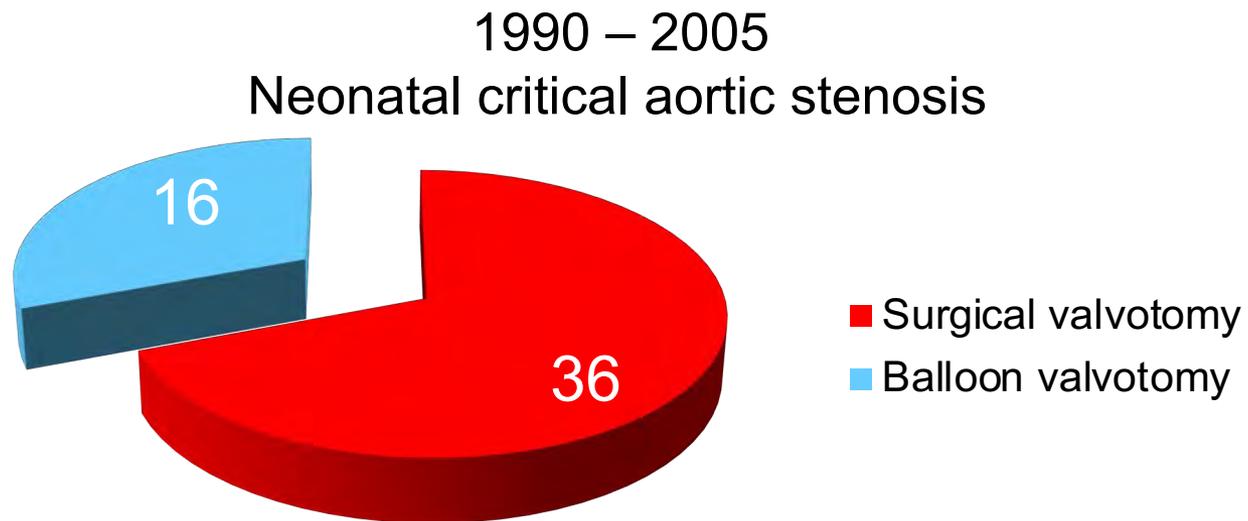
- VG normal (taille et fonction)
 - valvuloplastie chirurgicale : excellents résultats
diminution grad moy, amelioration fonction VG, peu de IAo
 - dilatation percutanée: non
- dysfonction VG
 - dilatation valvulaire percutanée
 - puis valvuloplastie ou Ross Konno

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

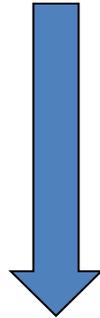
*Alexiou et al, Eur J Cardiothorac Surg, 2001.
Bhabra MS et al, Ann Thorac Surg, 2003.*

Traitement des sténoses aortiques

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

Remplacement valvulaire aortique chez l'enfant

2 Stratégies finales



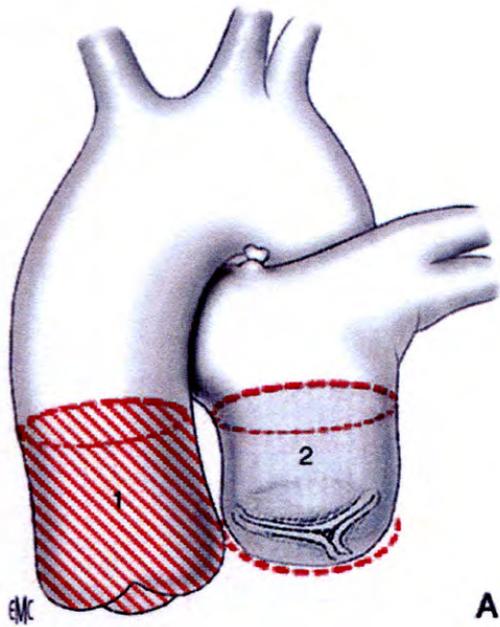
Valve mécanique (taille définitive)



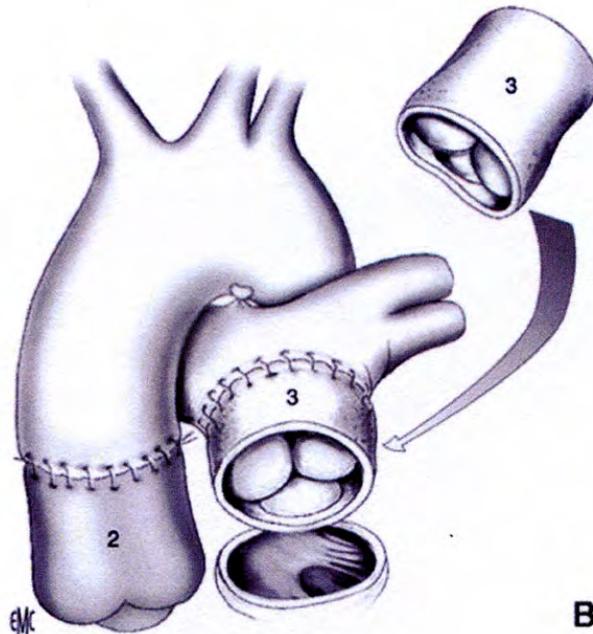
- Autogreffe pulmonaire
Ross et Ross-Konno

Ross / Ross-Konno chez l'enfant

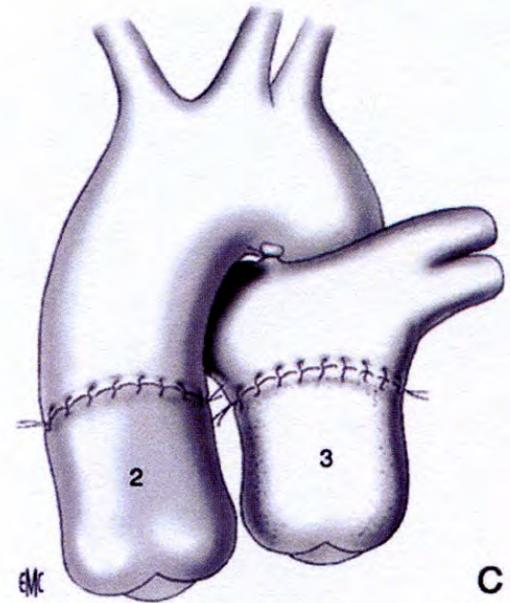
Principes



A



B



C

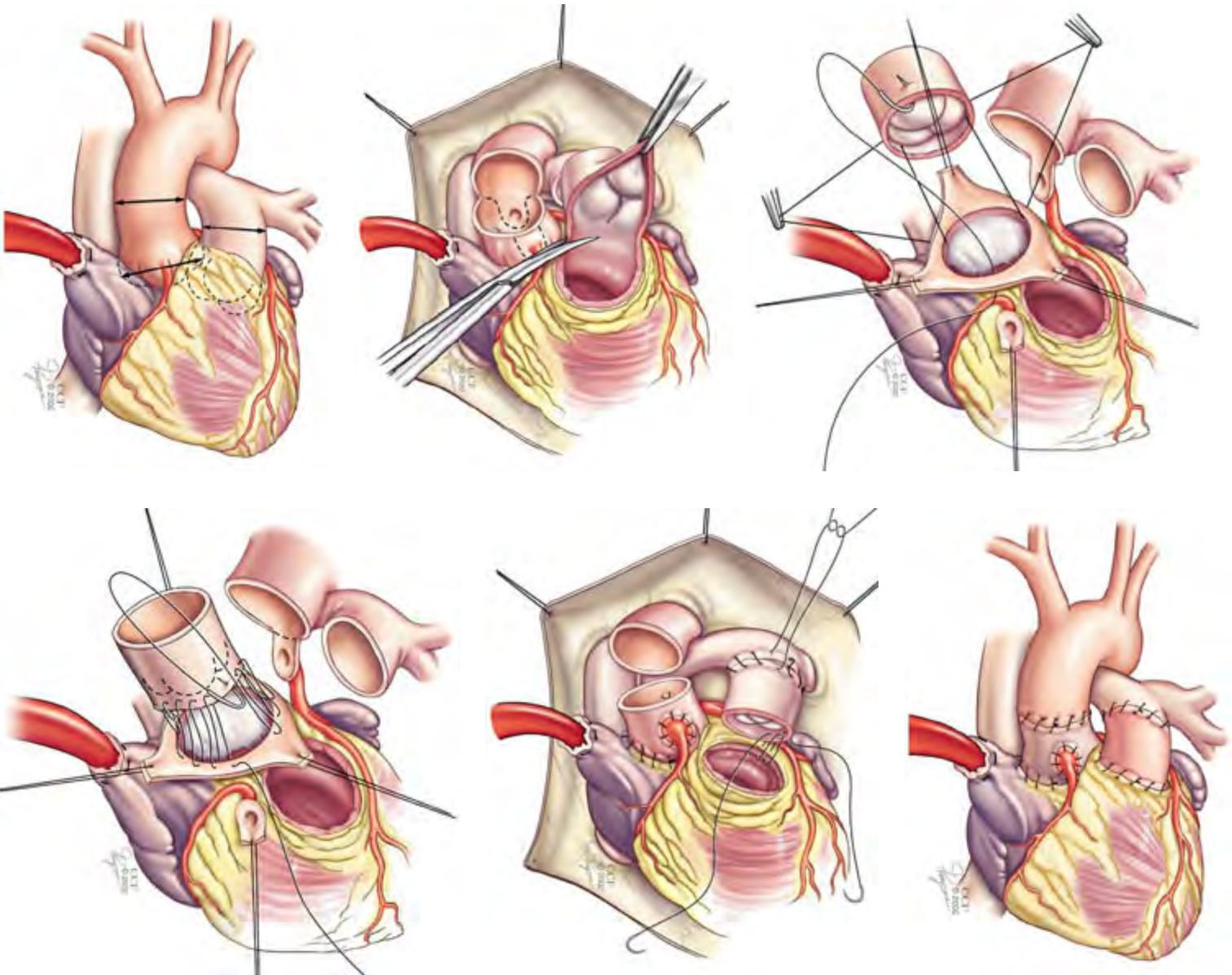
Ross / Ross-Konno chez l'enfant

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

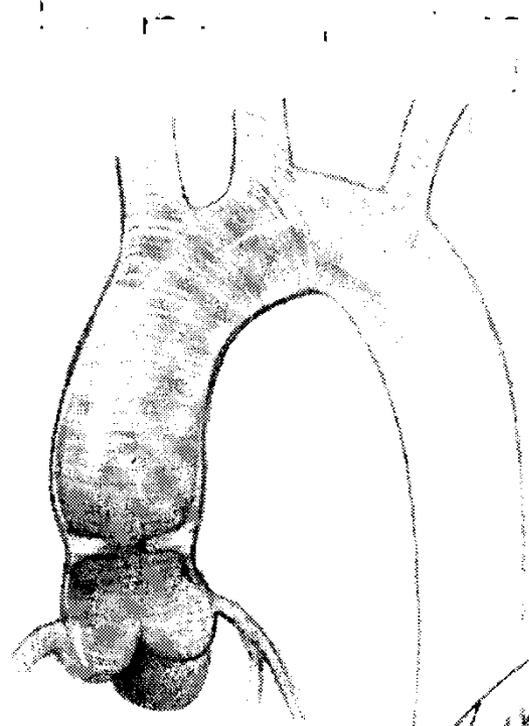
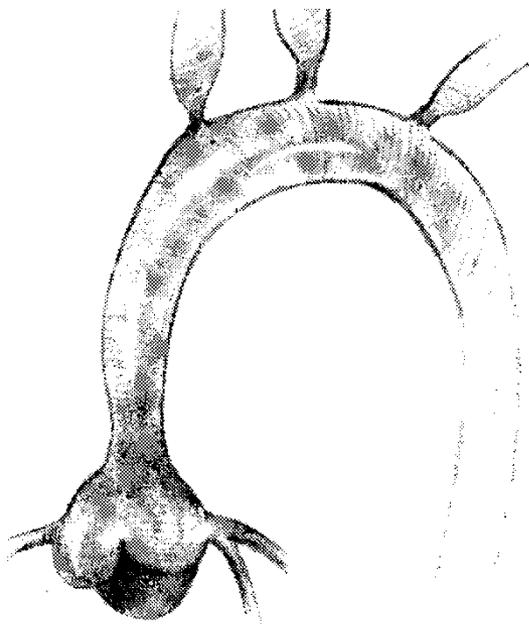
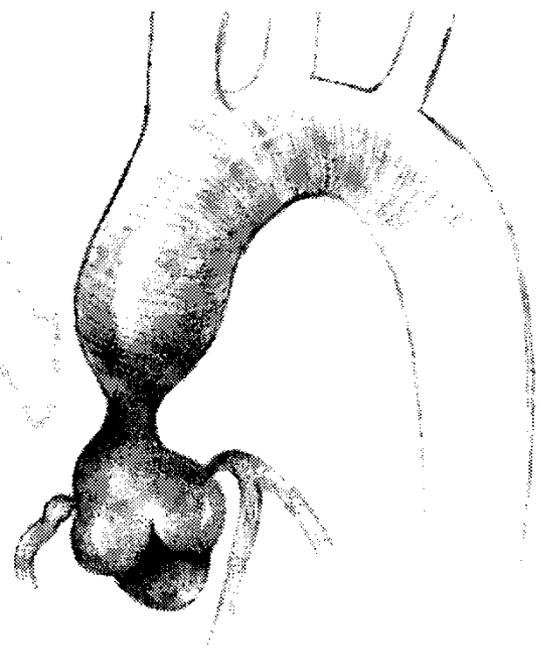


Sténose supravalvulaire aortique

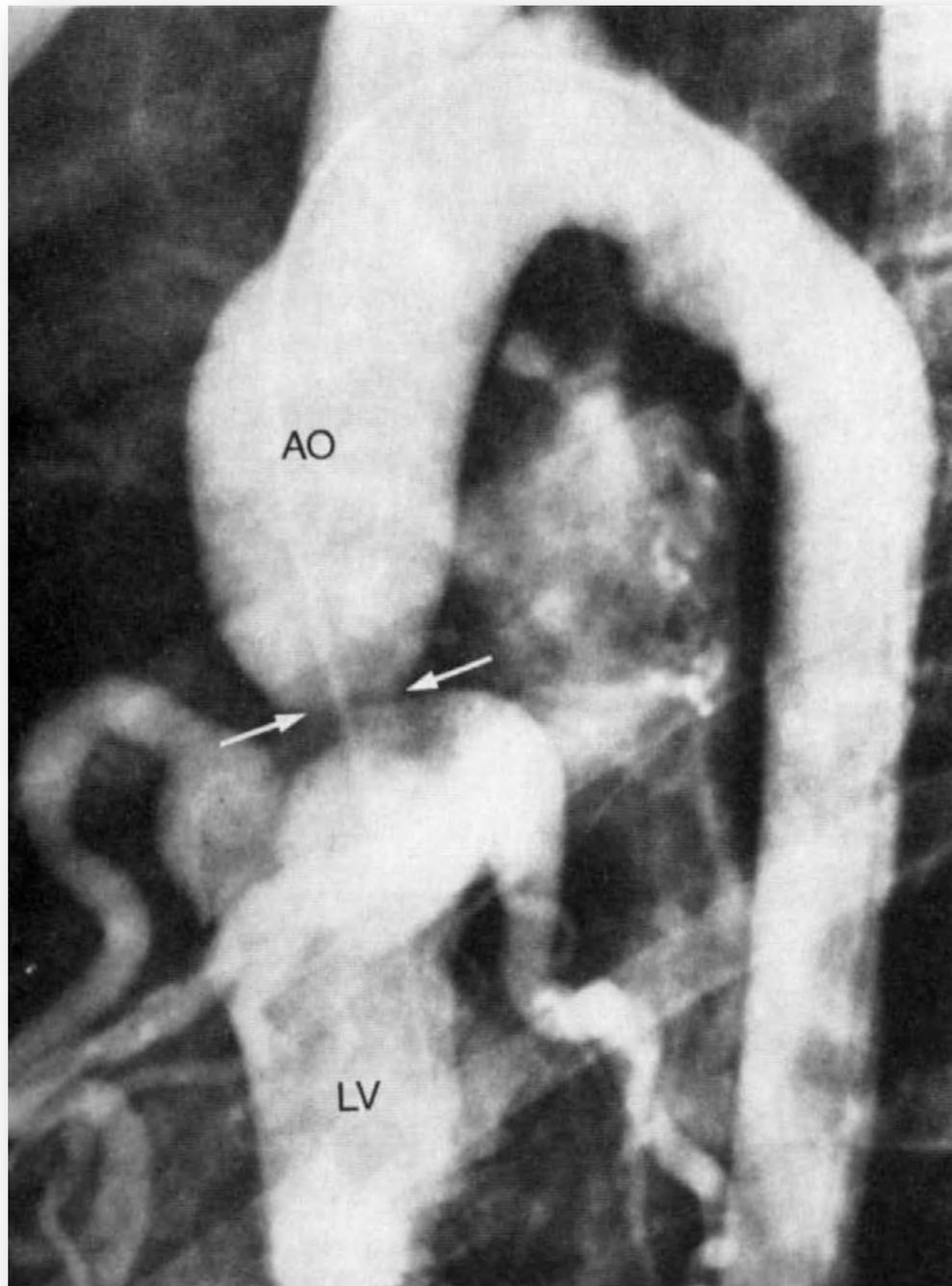


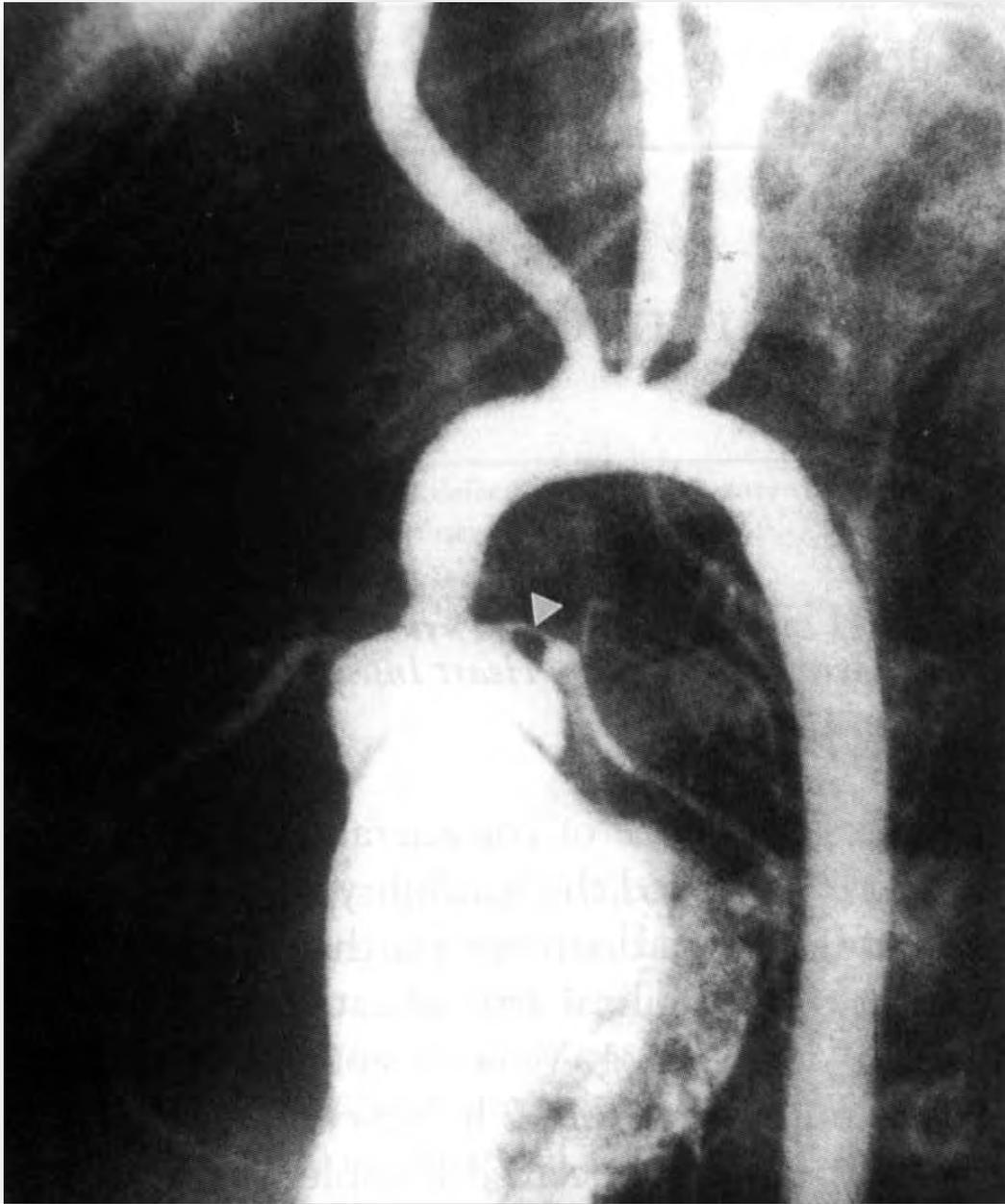
- sd de Williams Beuren
- del 7q11.23
- mutation gene élastine
- réduction et desorganisation des fibres élastiques dans la media aortique: perte d'élasticité paroi, hypertrophie du muscle lisse et depot collagene (aspect feuilleté)

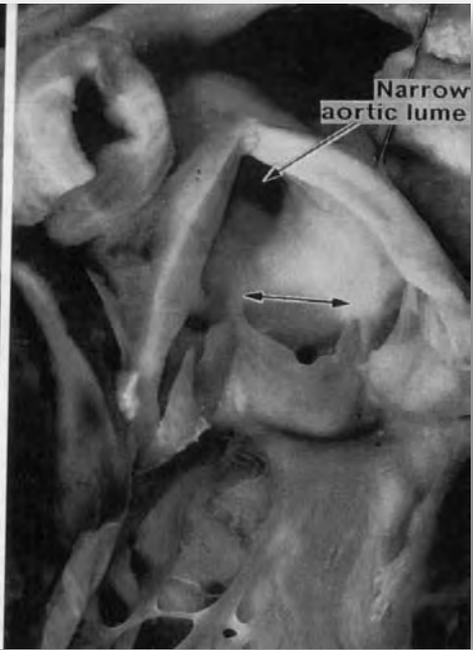
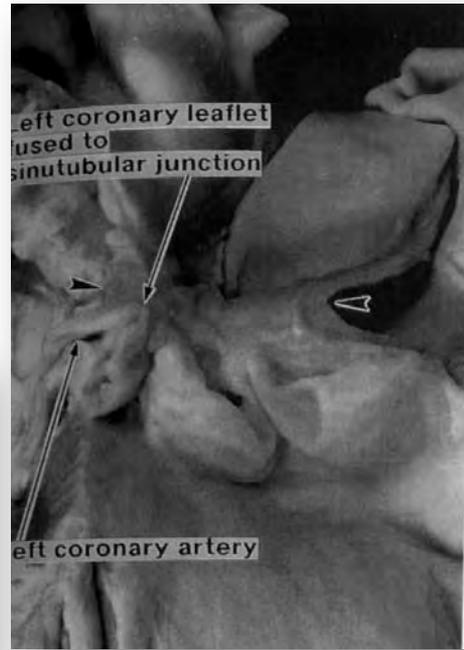




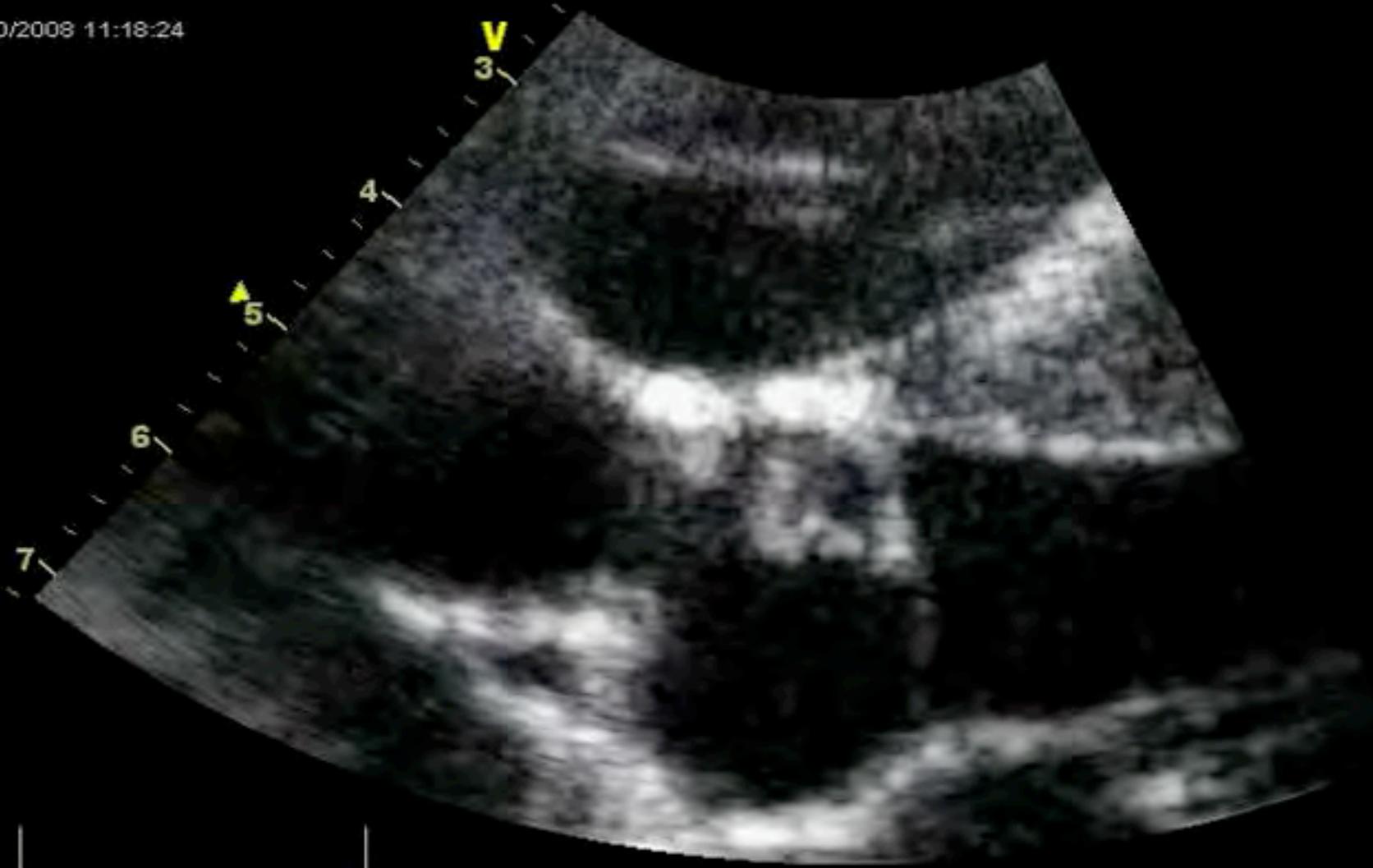
F. H. 91







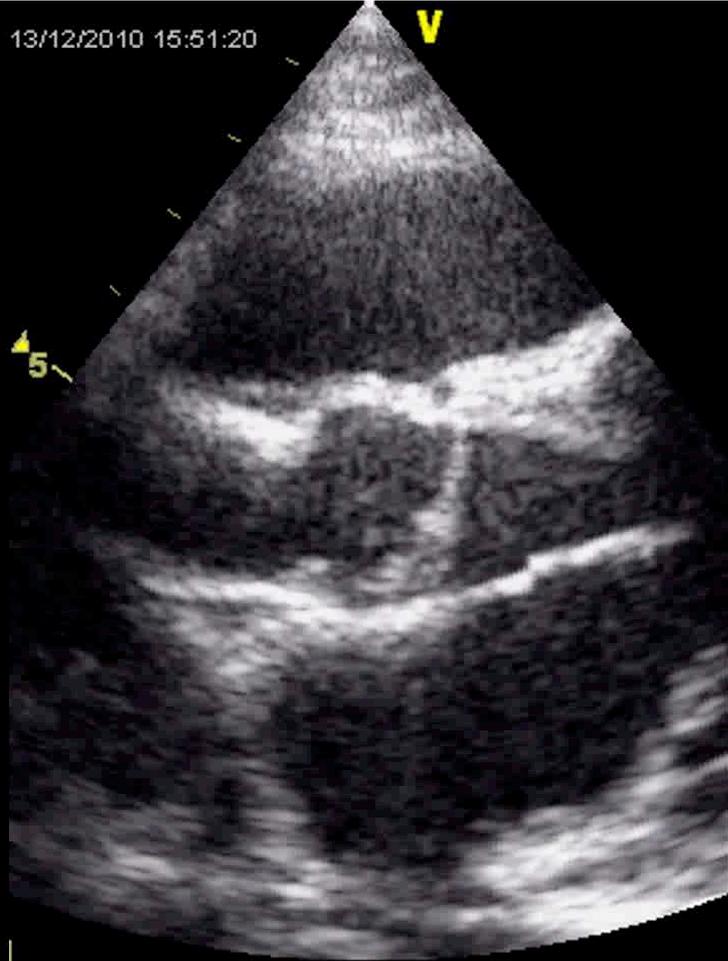
14/10/2008 11:18:24



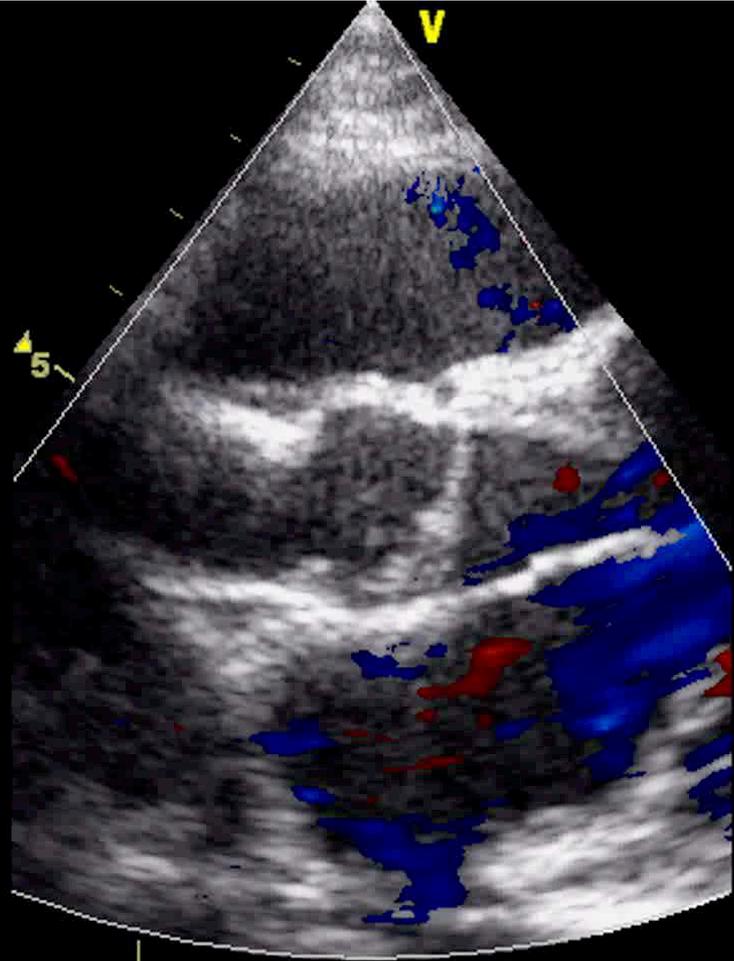
1:47

13/12/2010 15:51:20

V



V

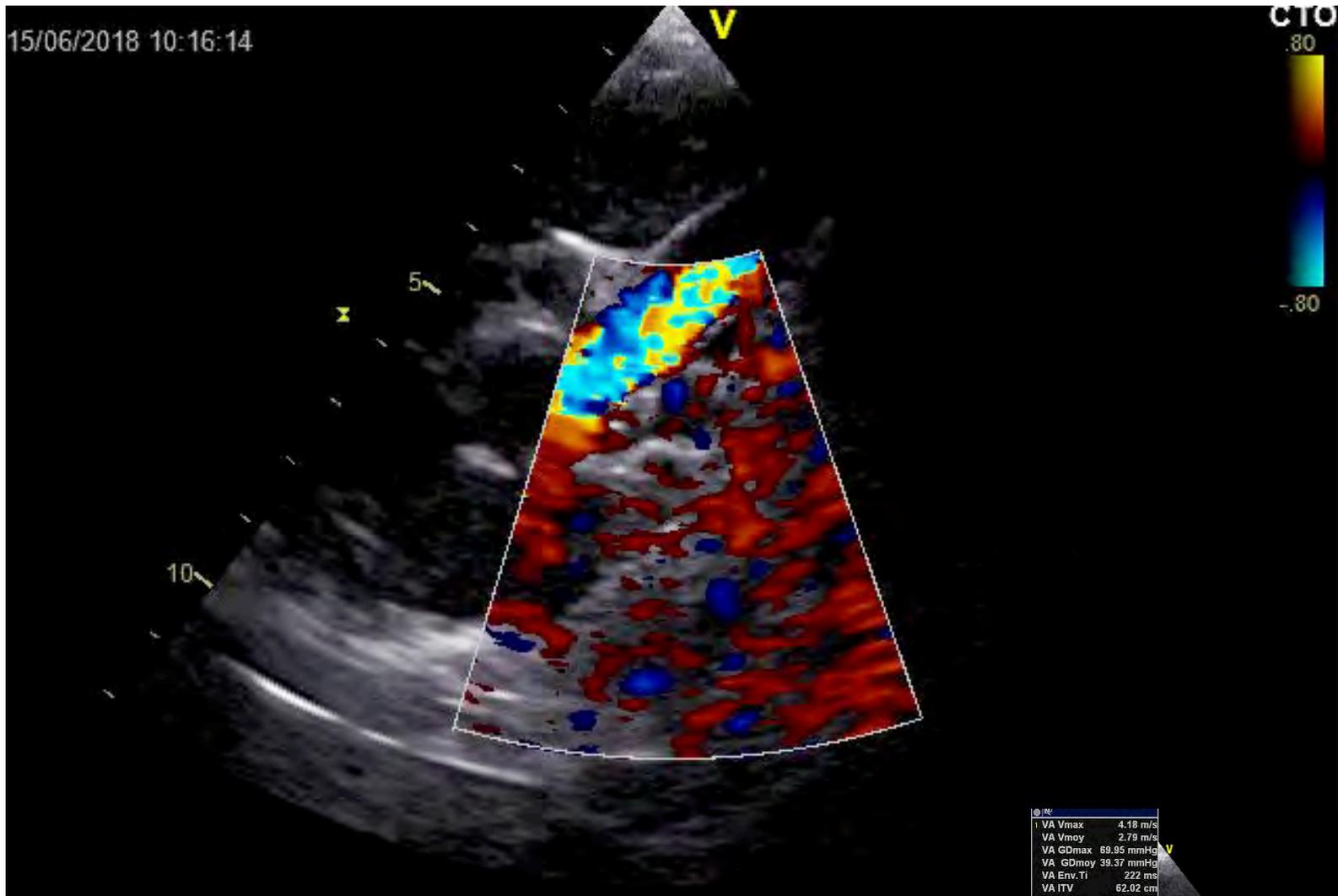


.95

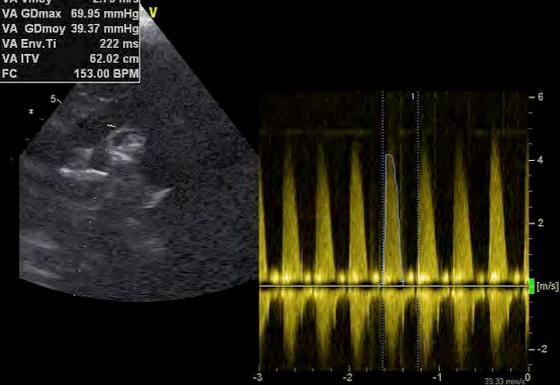
-.95

1:69

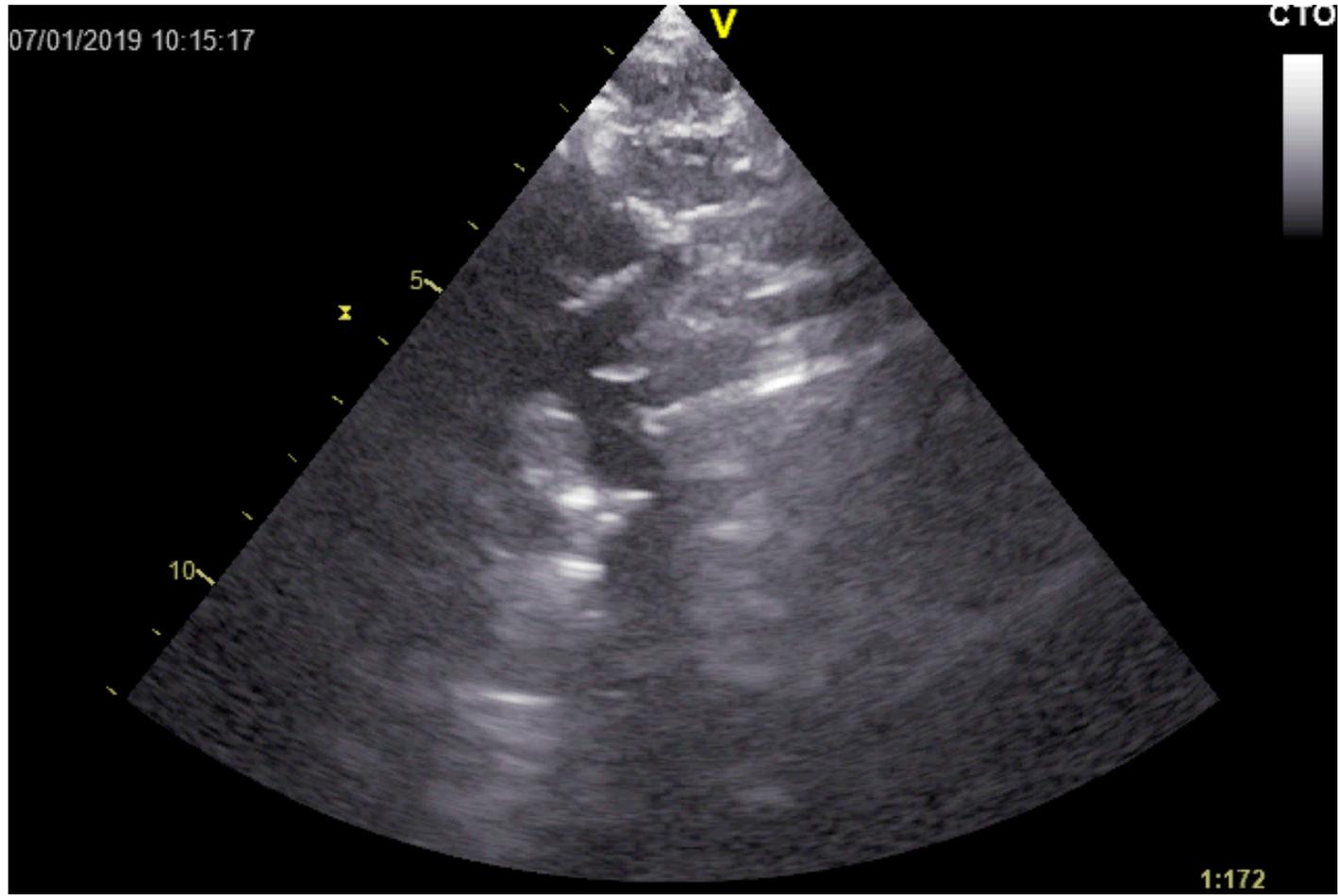
15/06/2018 10:16:14



1 VA Vmax	4.18 m/s
VA Vmoy	2.79 m/s
VA G _D max	68.95 mmHg
VA CD _{moy}	39.37 mmHg
VA Env.TI	222 ms
VA ITV	62.02 cm
FC	153.00 BPM



07/01/2019 10:15:17



CTO

1:172

3D
Ex: 6255
Se: 2
Volume Rendering No cut

RSP

HOPITAL NECKER ENFANT

DFOV 11.7cm
STND+

A
R
I

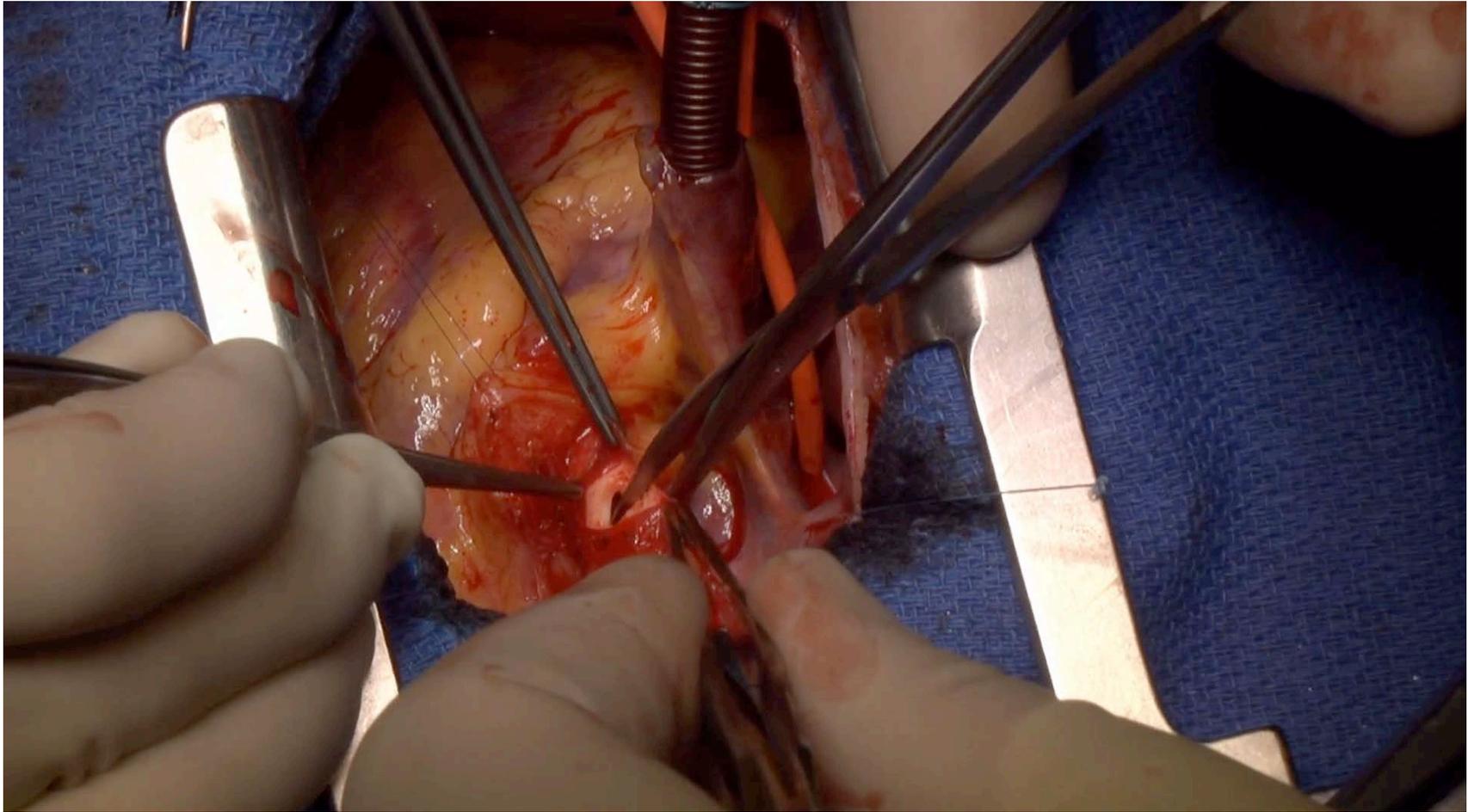


P
L
S

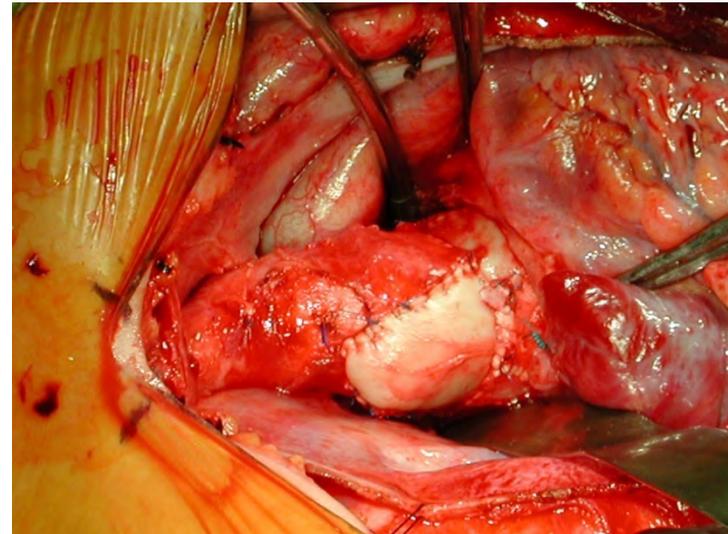
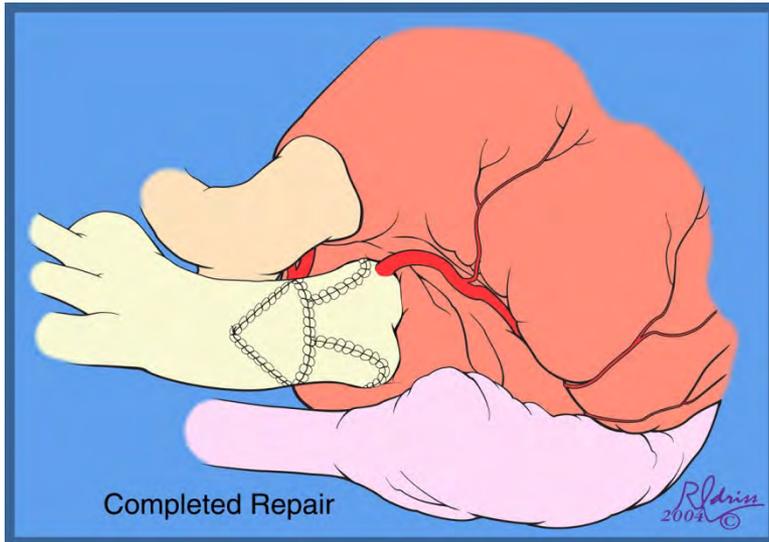
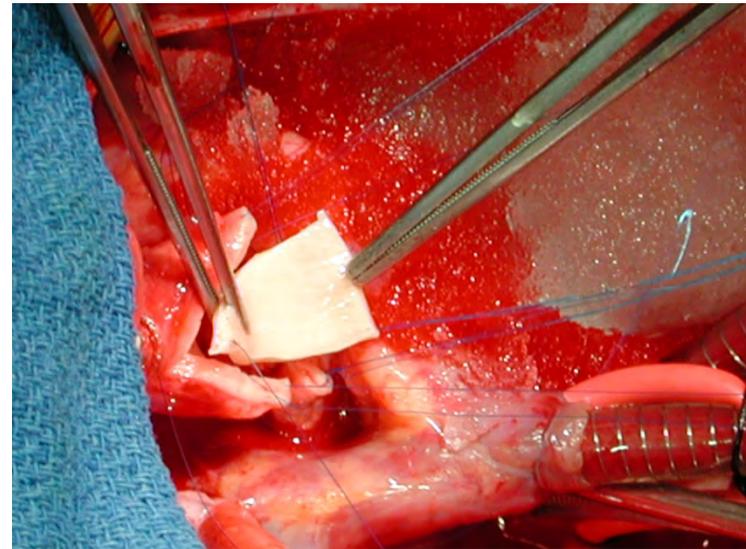
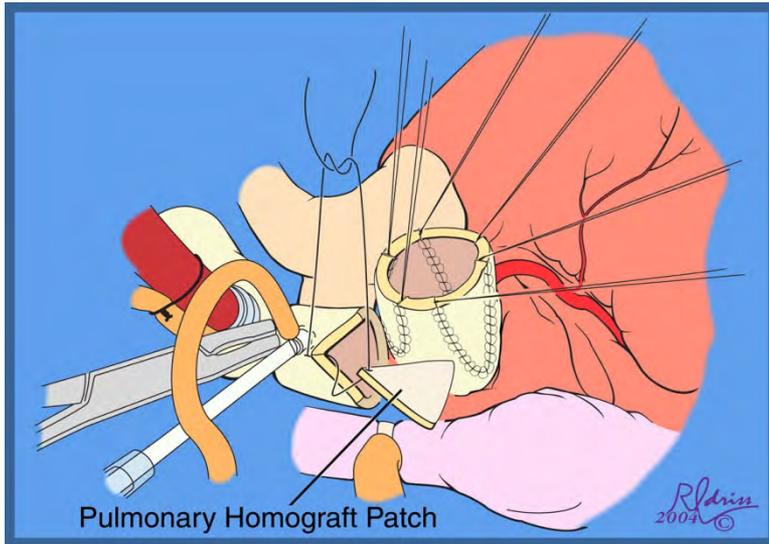
No VOl
kv 80
mA Mod.
Rot 0.40s/HE+ 39.4mm/rot
0.6mm 0.964:1 /0.6sp
Tilt: 0.0
11:49:53 AM
W = 4095 L = 2048

LIA

SVAS CT 3D 1

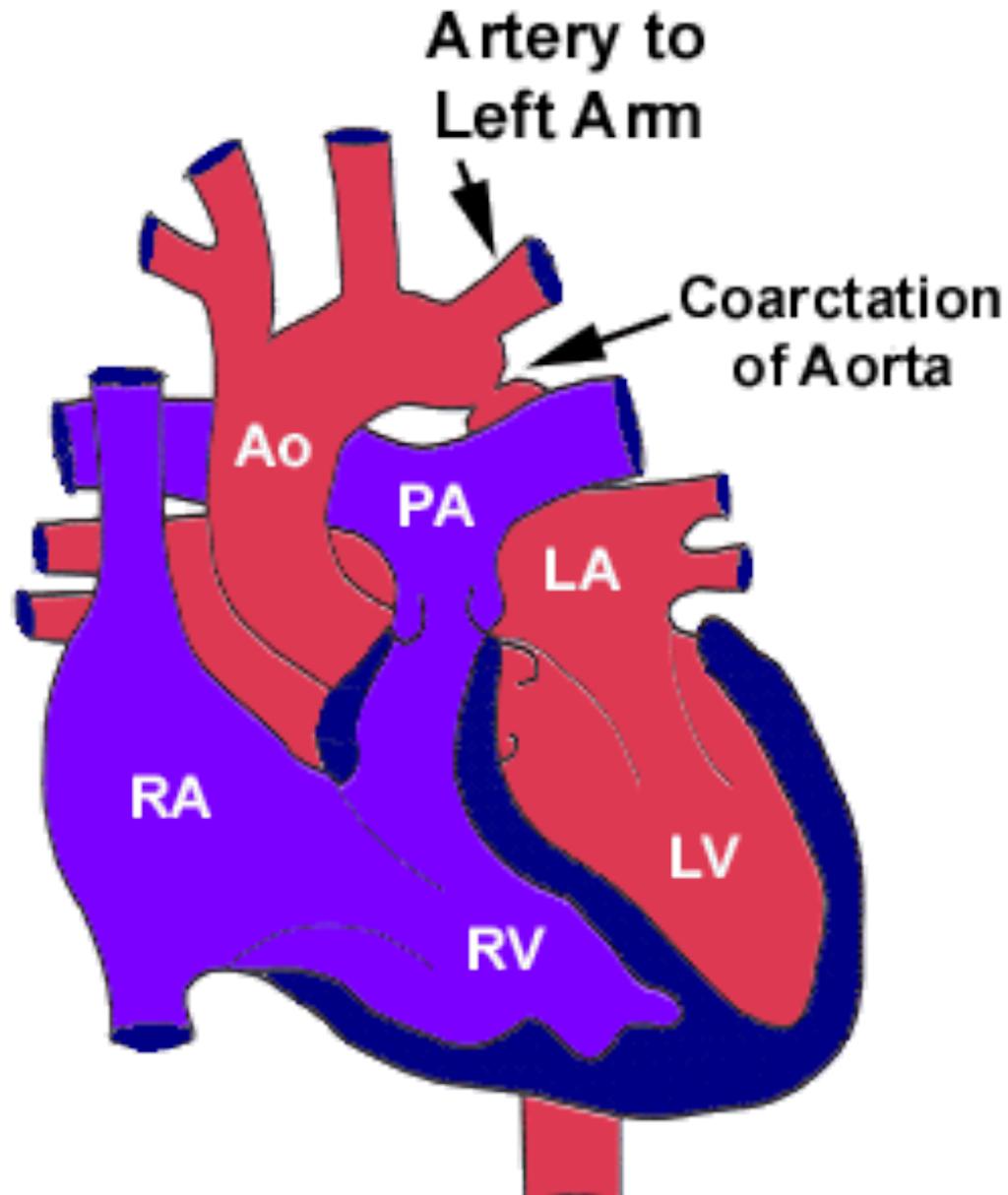


Sténose supravalvulaire aortique : Technique de Brom



Coarctations de l'aorte

Coarctation isthmique du nouveau-né

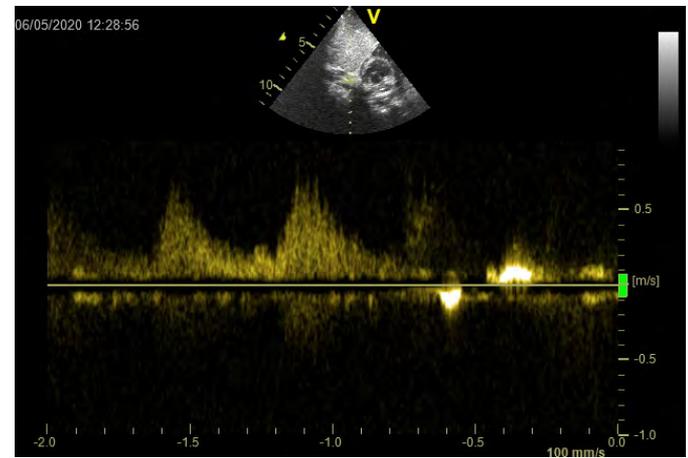
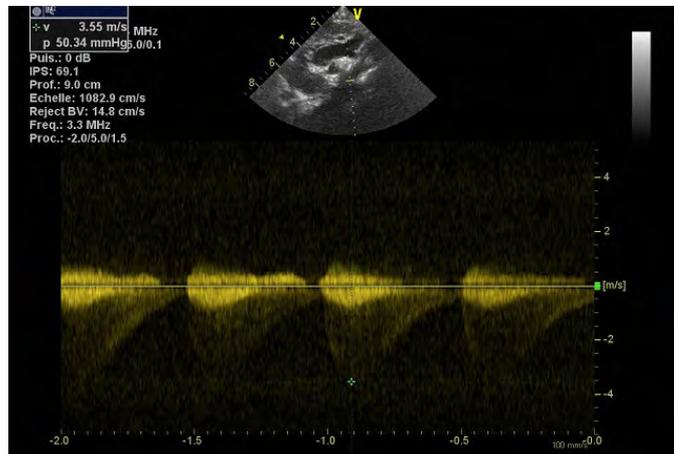
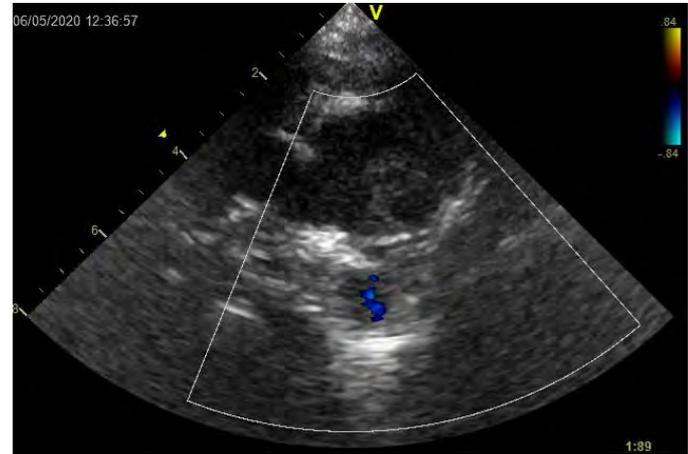
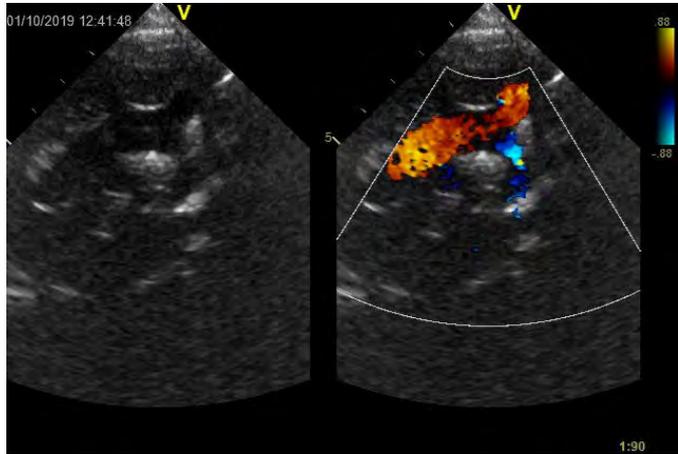


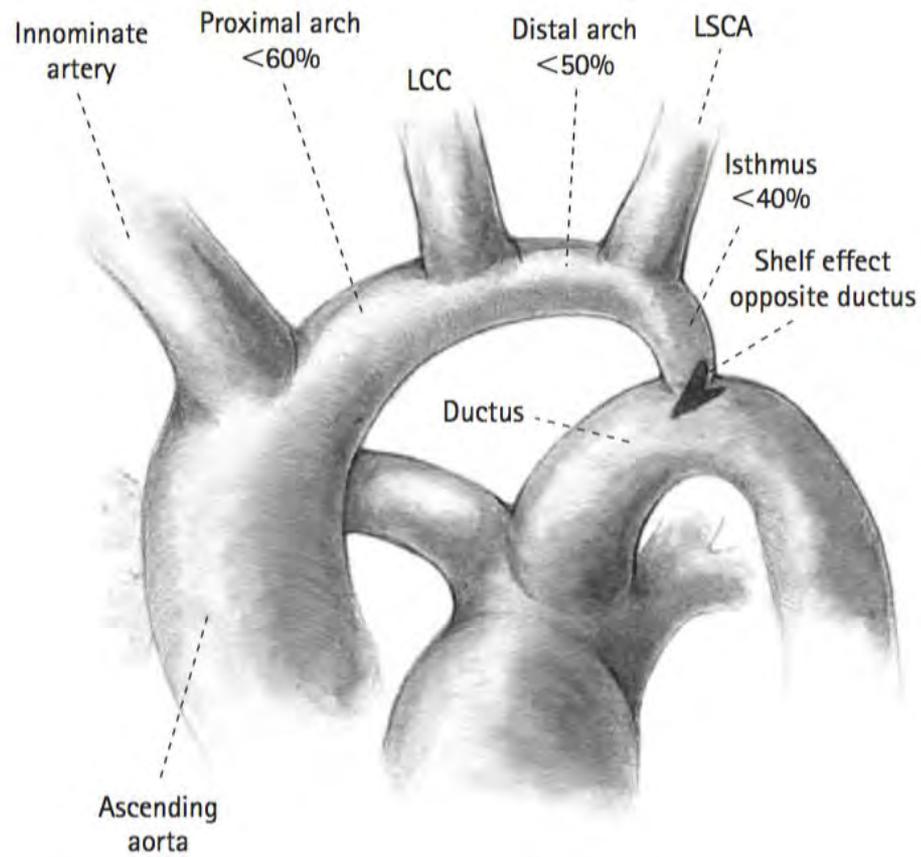
Les anomalies associées

- 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

diagnostic

- clinique :
 - pouls fémoraux faibles ou abolis/ asymetrie MS MI
 - HTA membre sup dt/ gradient MSup –Mb inferieurs
 - insuffisance cardiaque si dysfonction VG
- échographique
 - gradient sur l'isthme ao $>$ ou $=20$ mmHg et prolongement diastolique du flux
 - étude de la crosse et isthme aortique
 - VG
 - lésions associées :bicuspidie, obstacles étagés,CIV

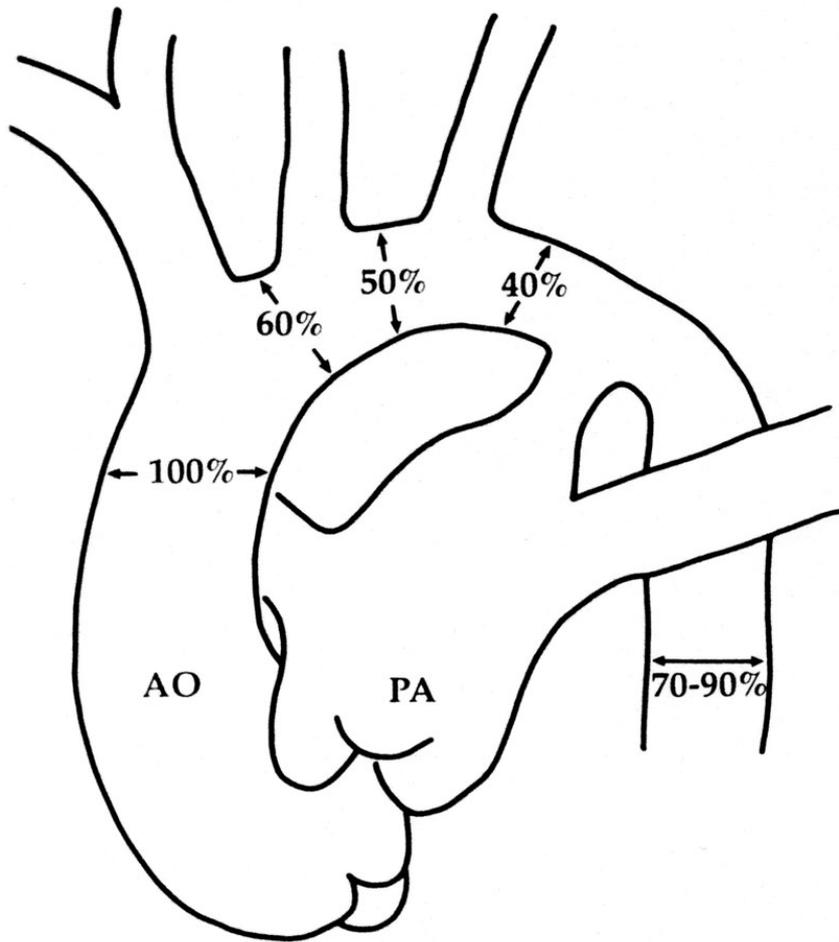




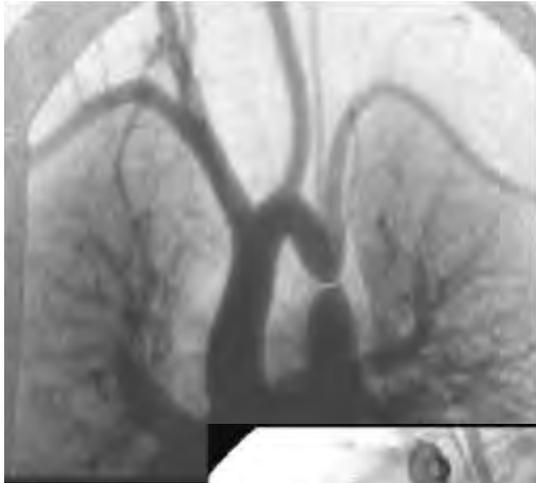
Le diamètre des 3 segments

- Arche proximale
- Arche distale
- Isthme

L'arche normale n'est pas un tube régulier



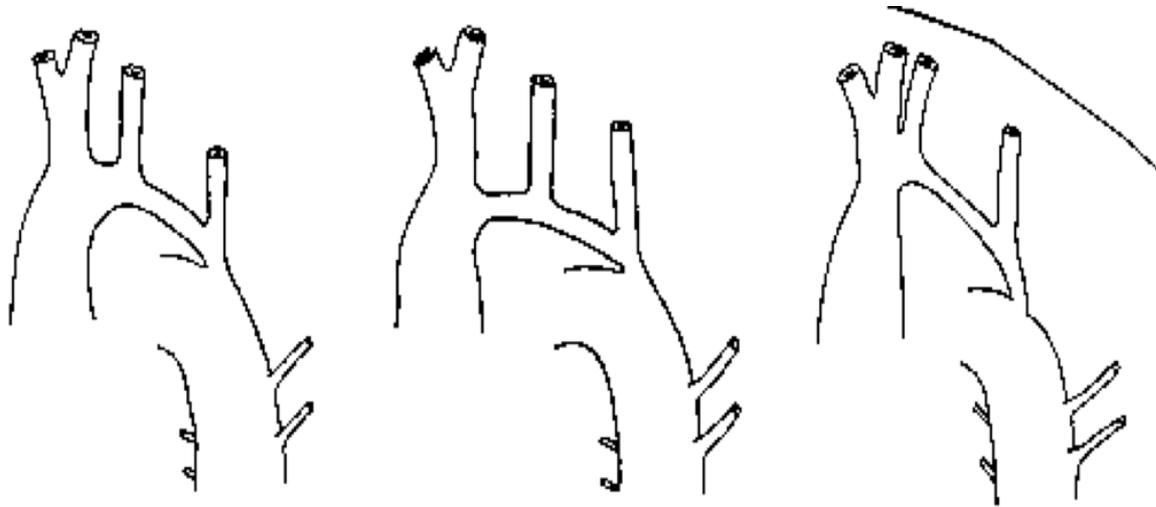
Comment définir l'hypoplasie de l'arche?



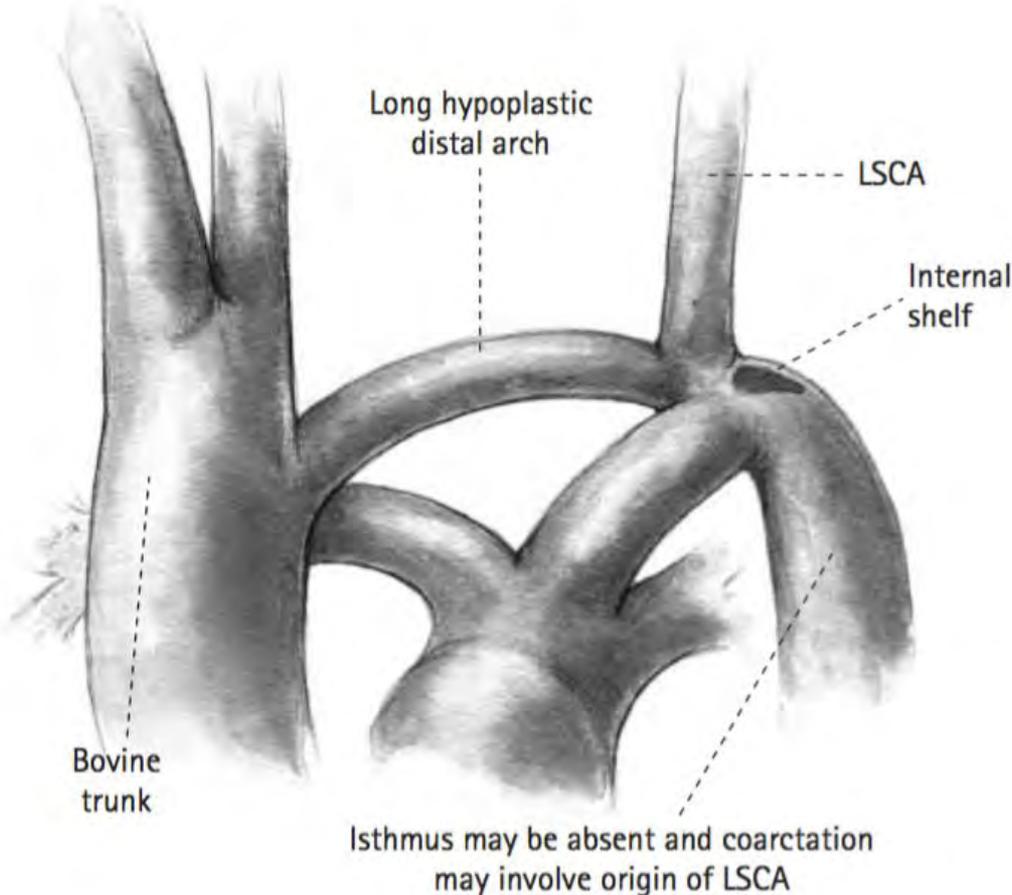
- Diamètre (mm) inférieur au poids (kg) +1
- Z score inférieur à -2
- Diamètre inférieur à 50% de l'Ao Asc

préciser aussi:

La distribution des troncs et la longueur des segments

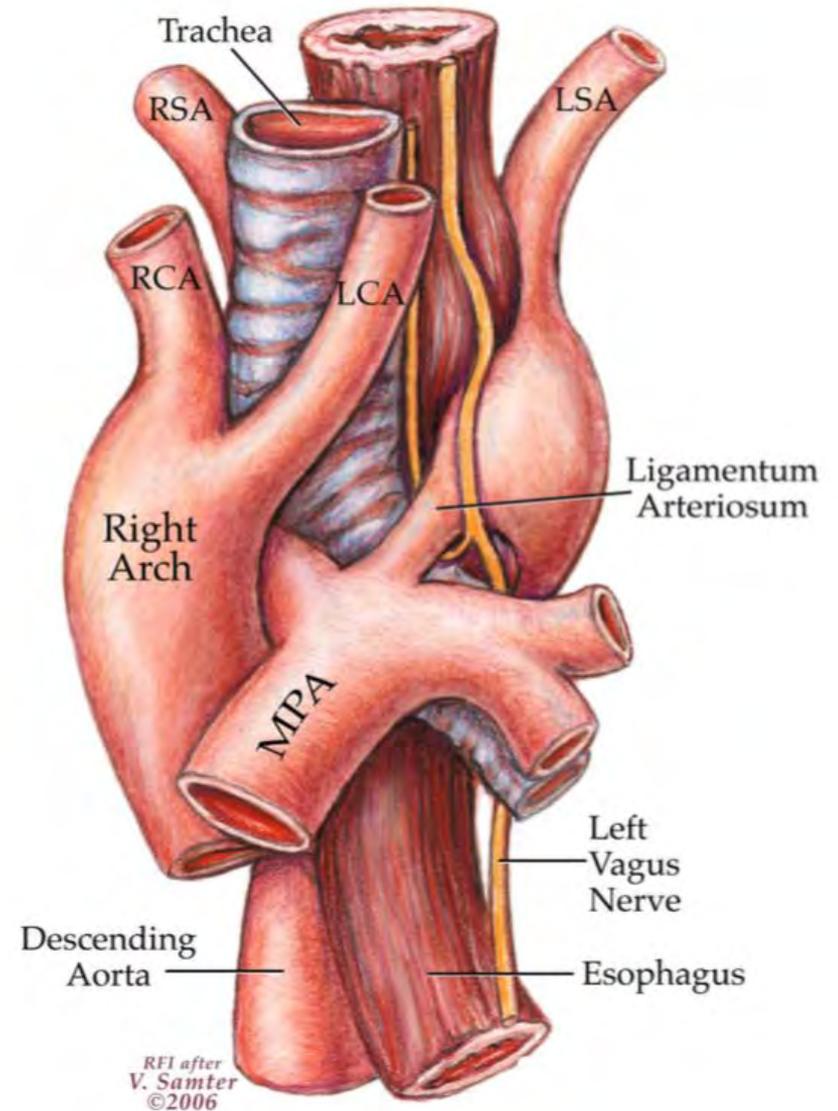


La distributions des troncs supra aortiques peut conditionner la réparation



Préciser
Le coté de la crosse aortique

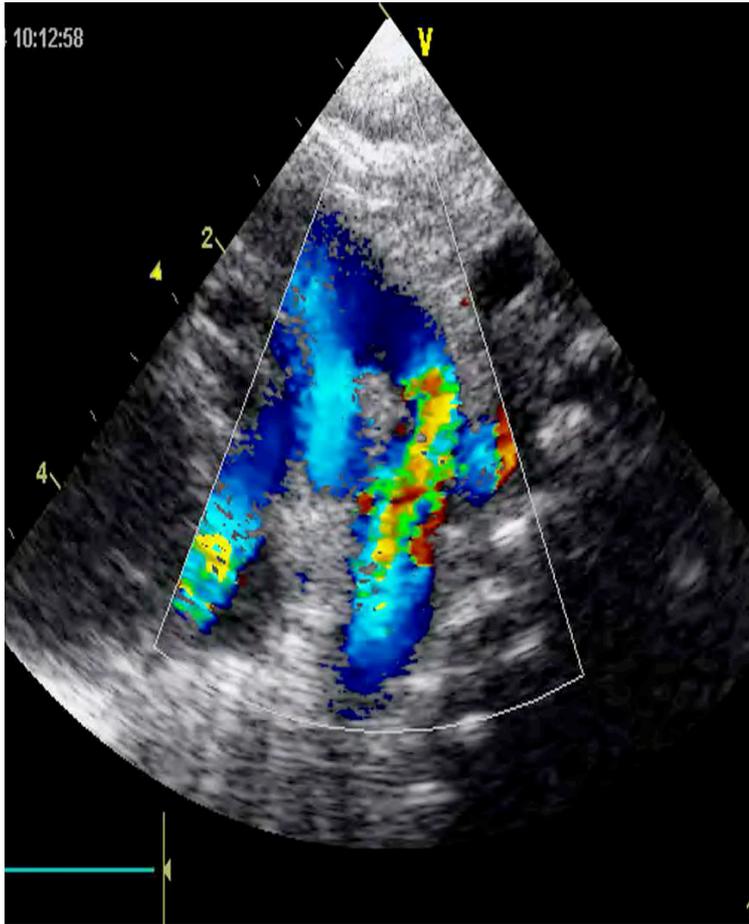
**Chirurgie impossible par
thoracotomie gauche
en cas d'arche droite**

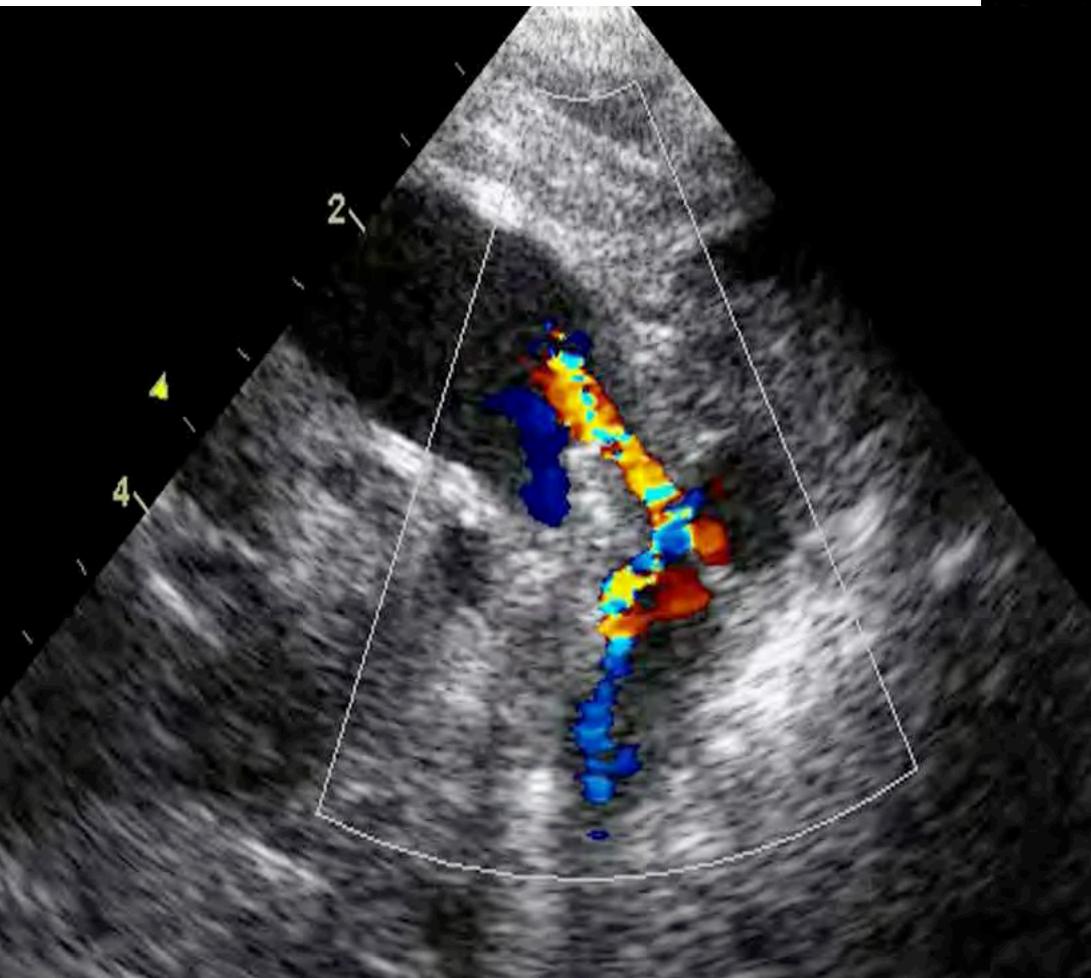


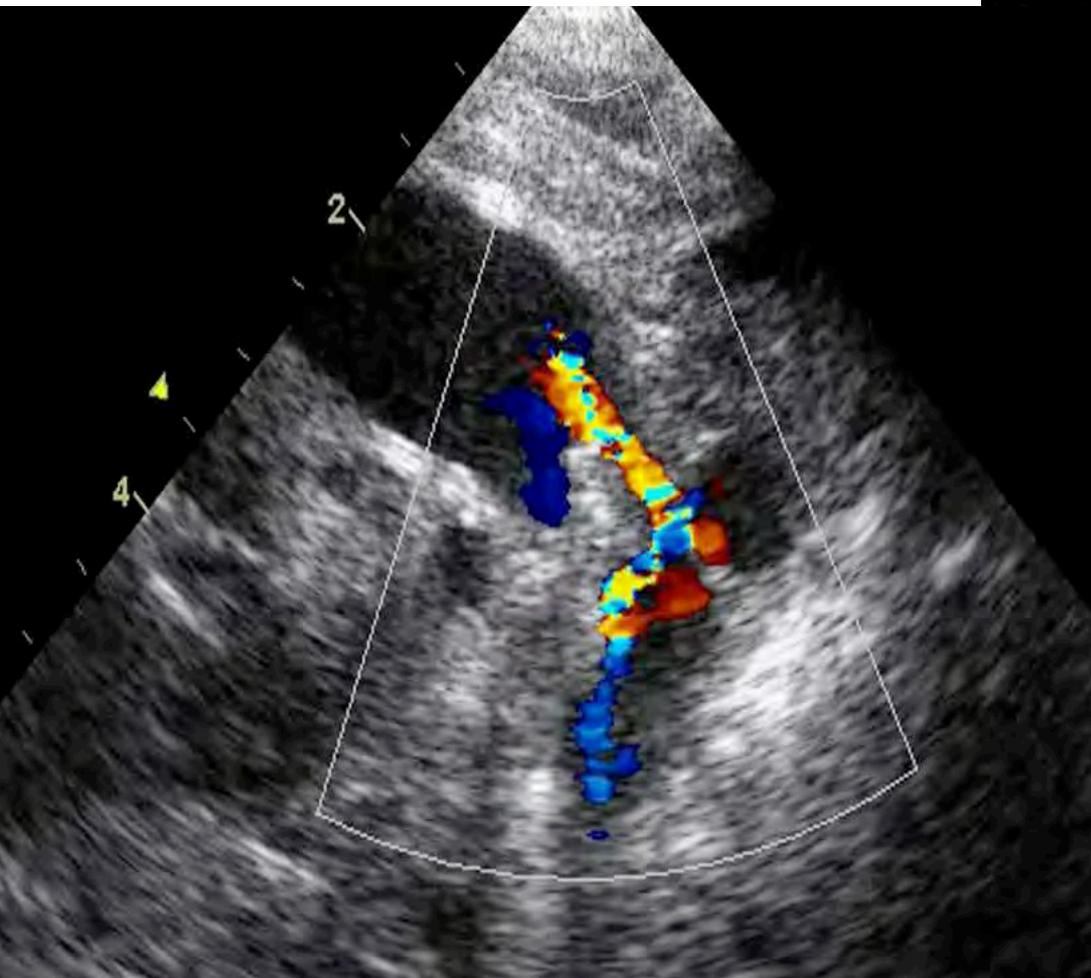
Stratégie chirurgicale

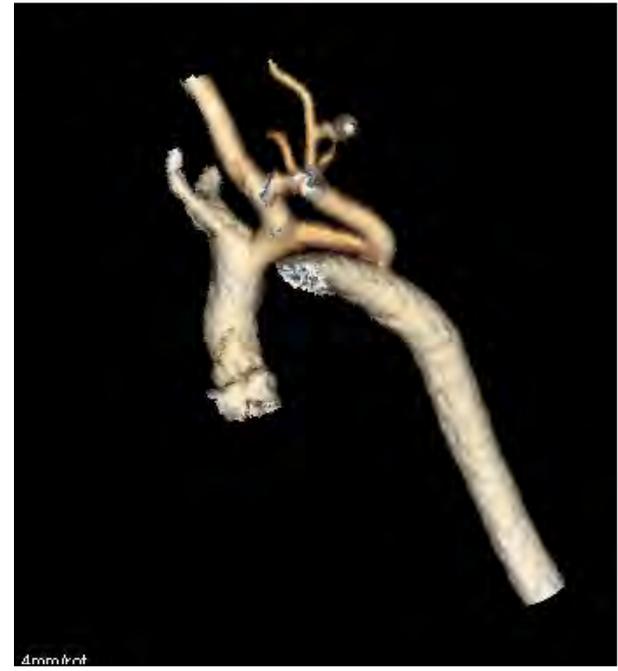
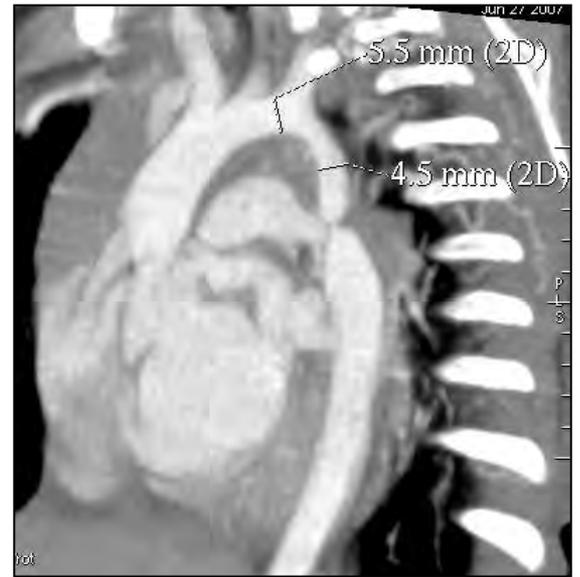
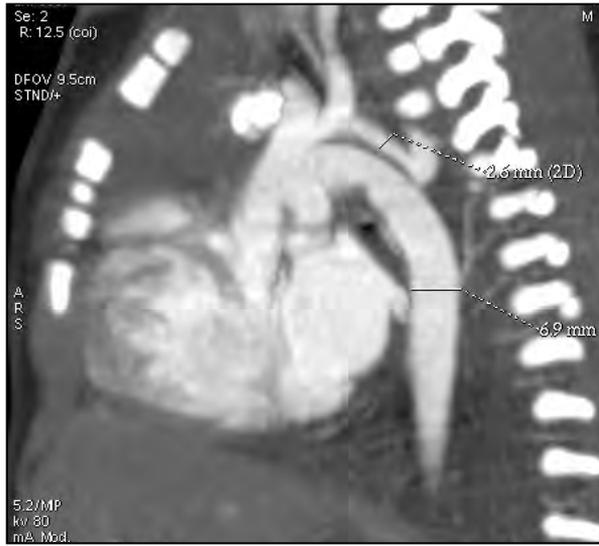
- Ducto-dépendance ou défaillance cardiaque
 - Chirurgie néonatale
- Hypertension artérielle
 - Chirurgie
- Tout va bien
 - Chirurgie apres 1 mois

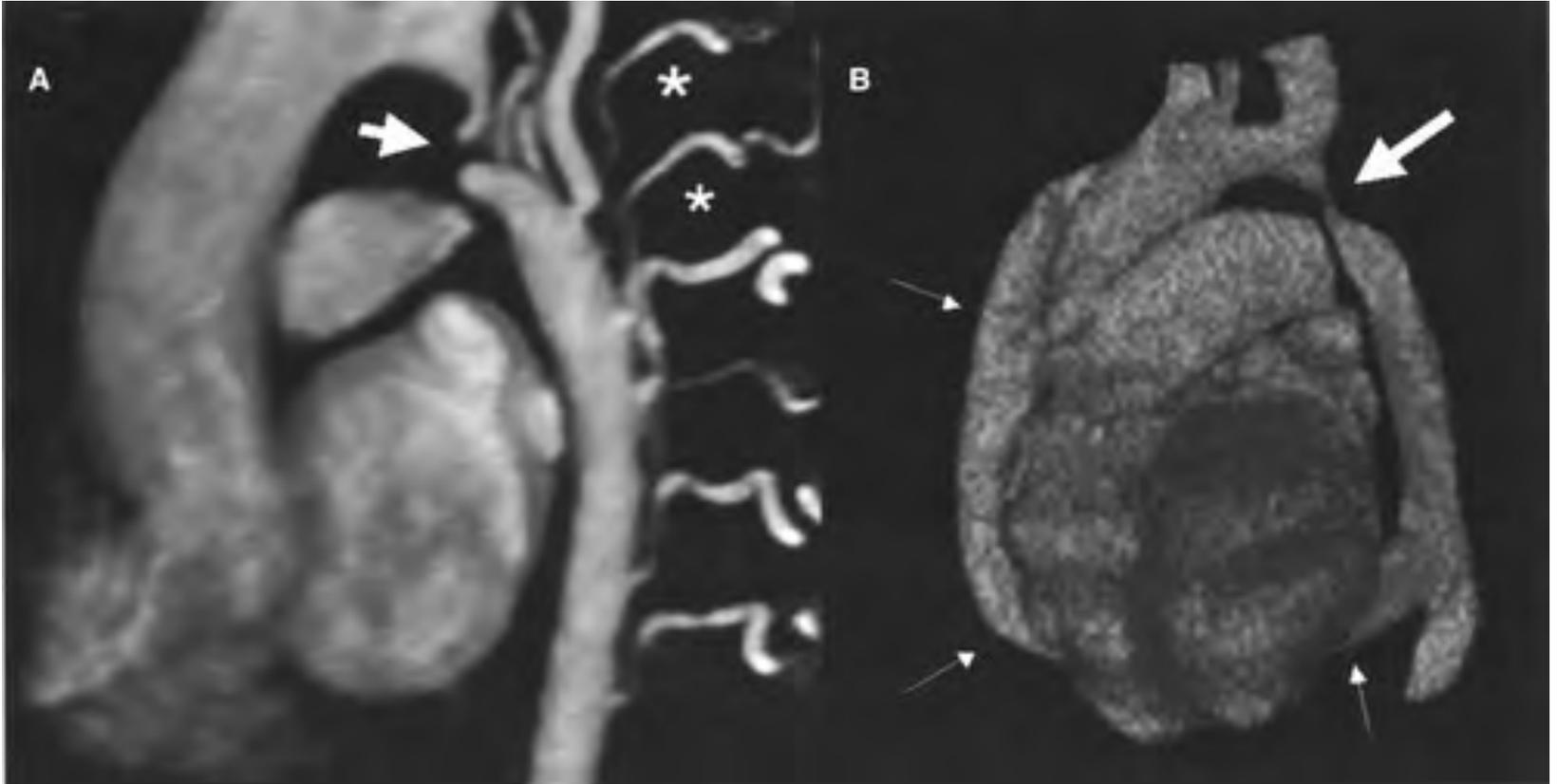
Perfusion systémique ducto-dépendante ?









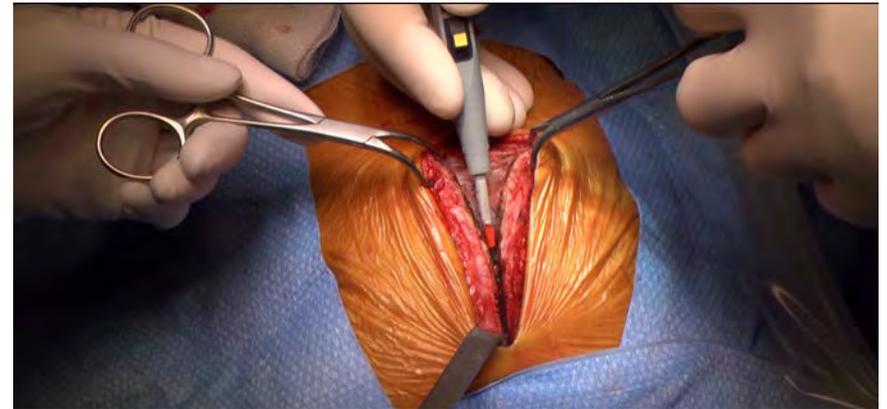


traitement chirurgical : abord

Par thoracotomie



Par sternotomie



stratégie établie selon :

1° Le degré d'hypoplasie du segment proximal de l'arche

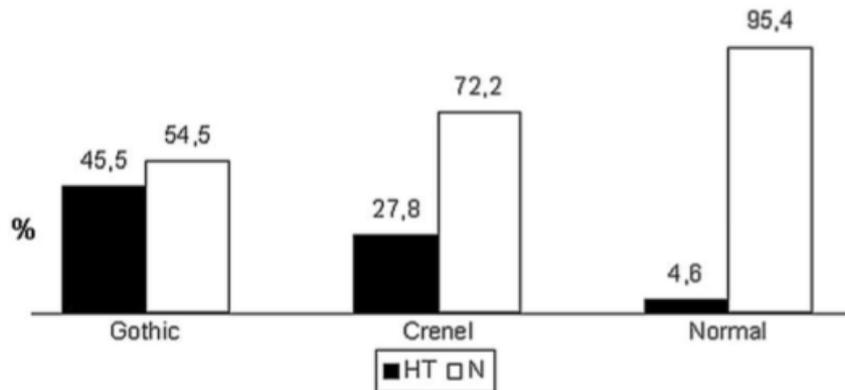
2° La cardiopathie associée

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

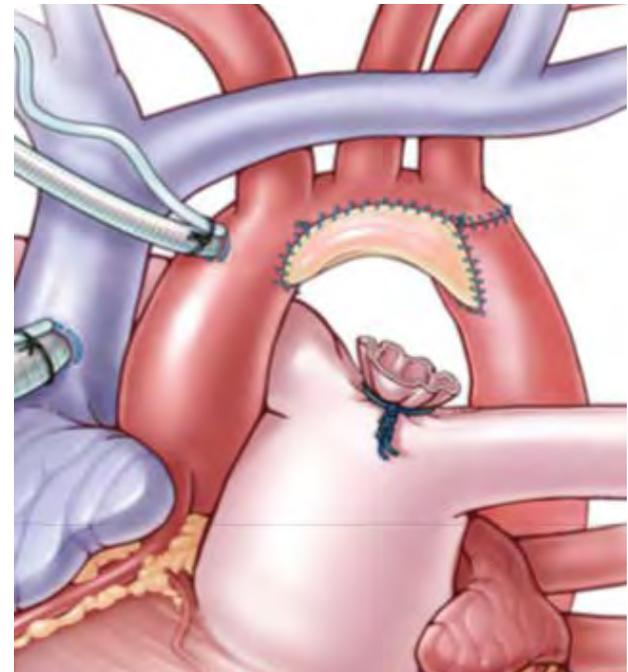
Réparation par sternotomie

Géométrie et hypertension artérielle



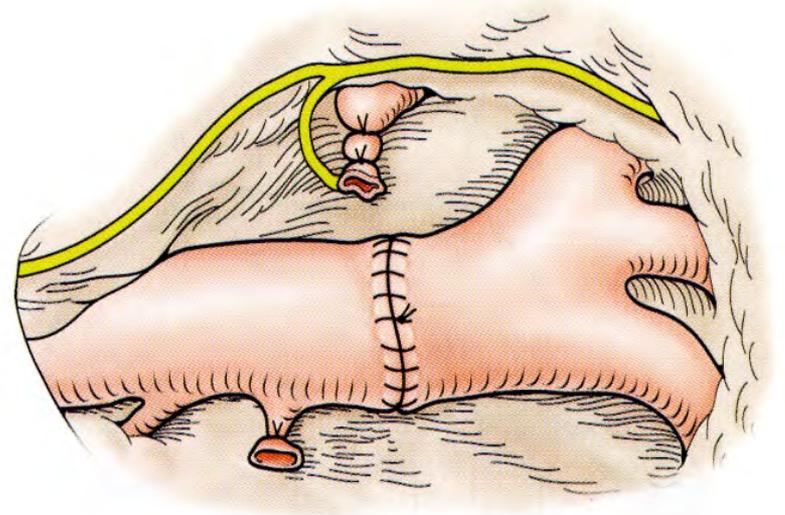
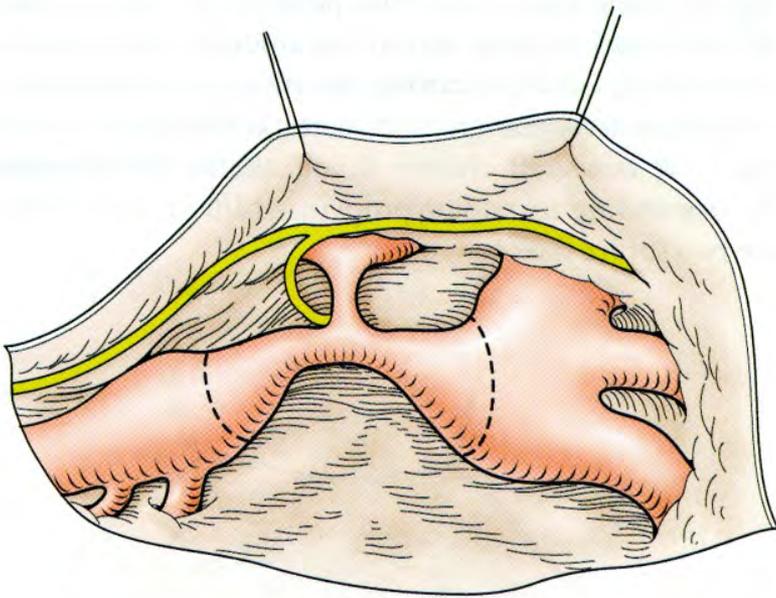
Late systemic hypertension and aortic arch geometry after successful repair of coarctation of the aorta

Phalla Ou^{a,c}, Damien Bonnet^{a,b,*}, Louis Auriacombe^a, Elisa Pedroni^a, Fanny Balleux^c, Daniel Sidi^a, Elie Mousseaux^{c,d}



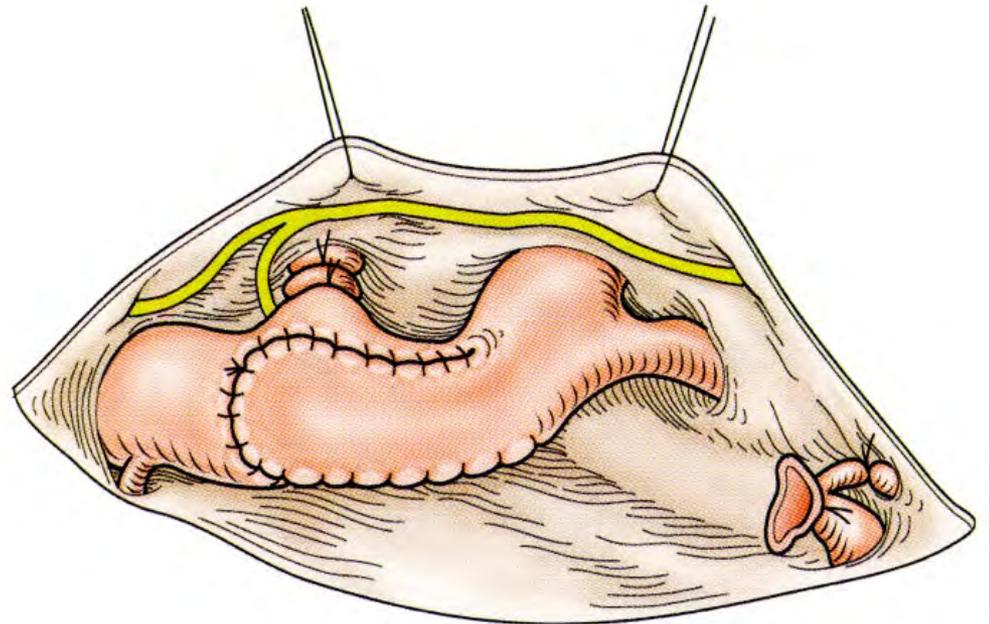
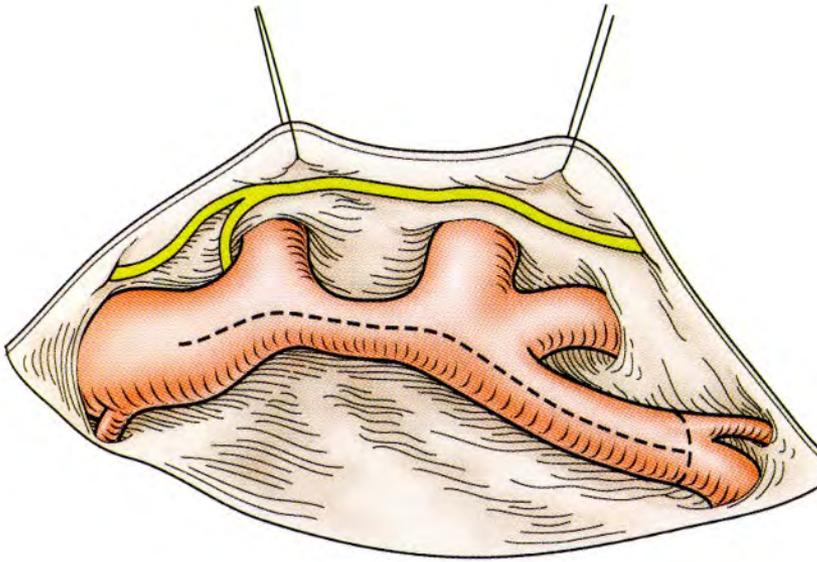
Réparation par Thoracotomie

Résection anastomose: Crafoord



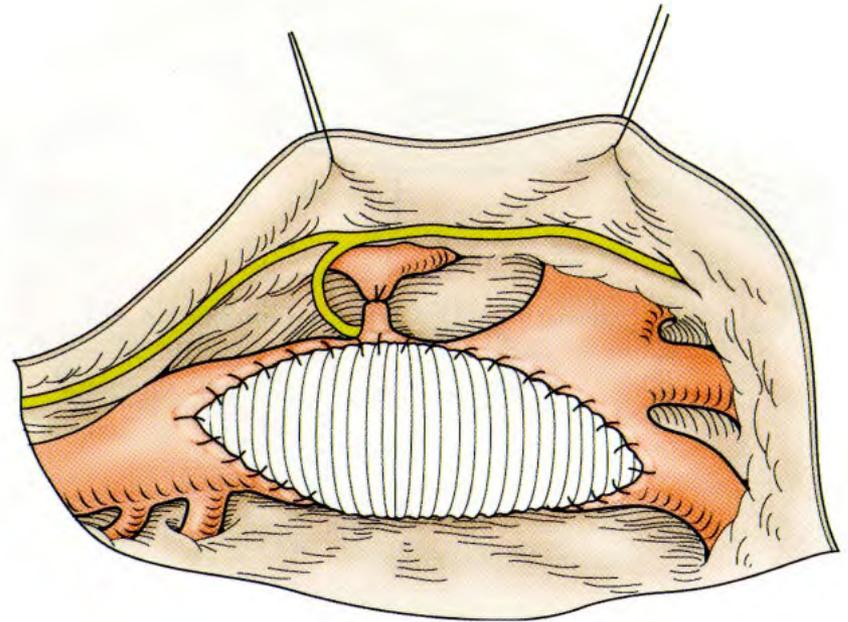
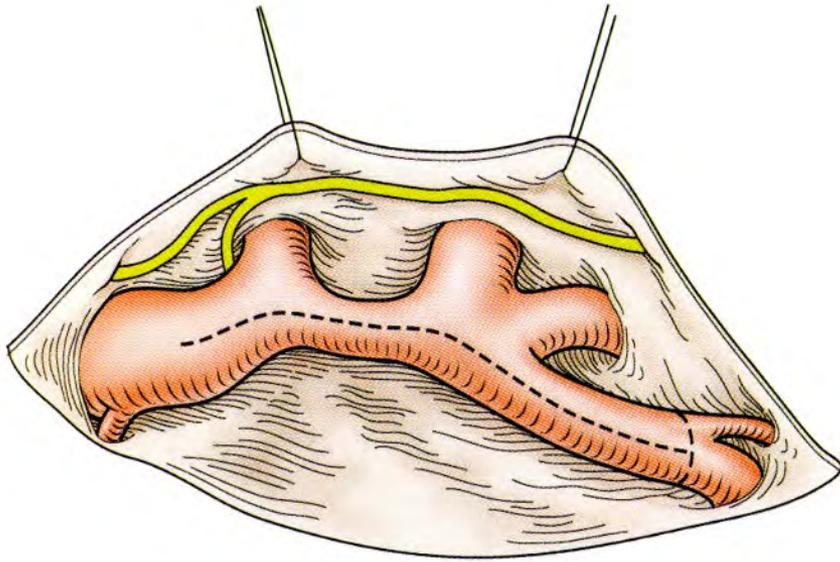
Réparation par Thoracotomie

Réparation avec un flap subclavier: Waldhausen



Réparation par Thoracotomie

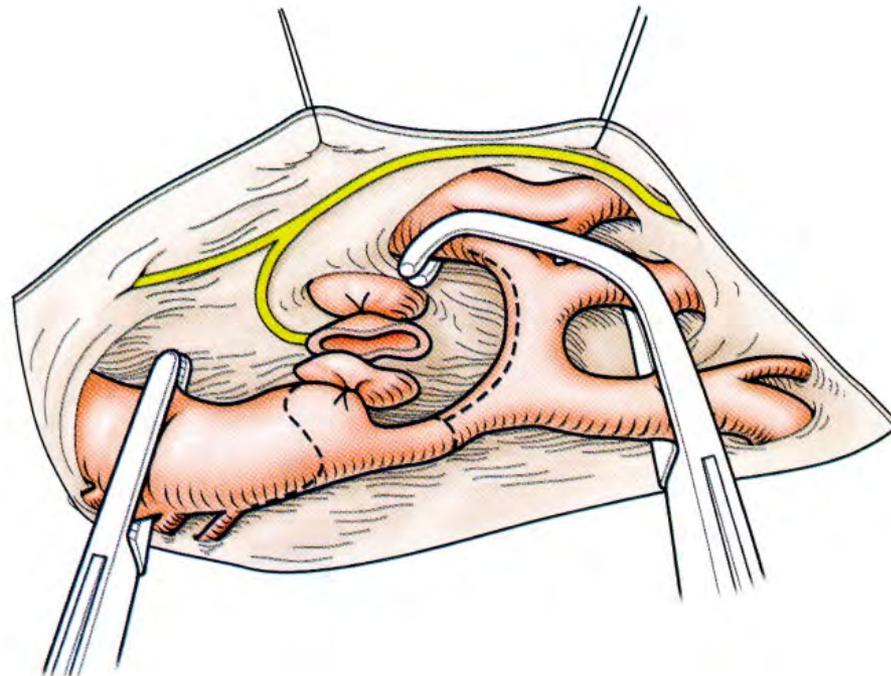
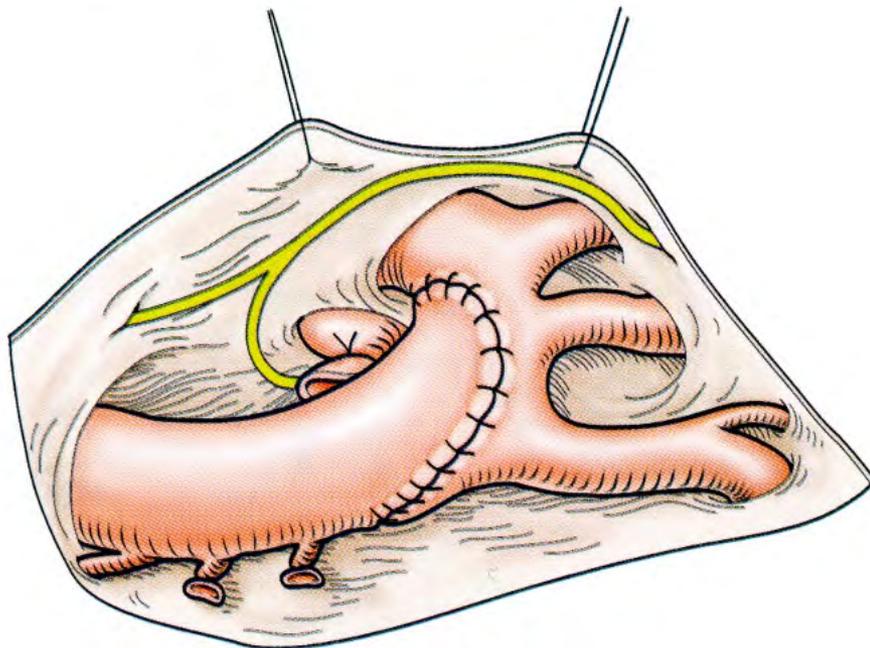
Plastie d'élargissement avec patch



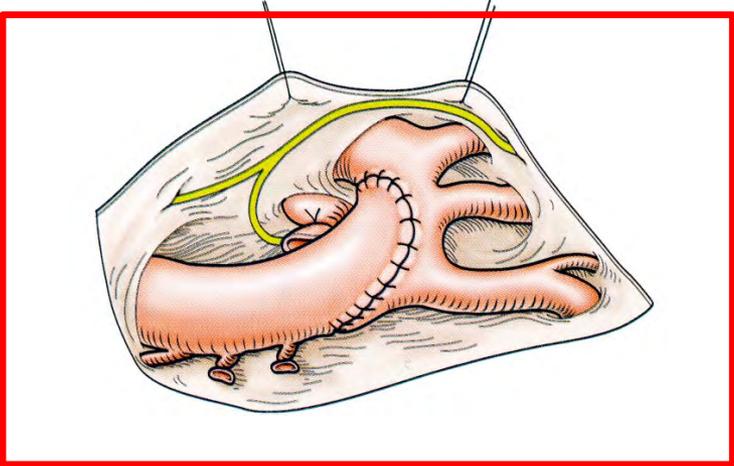
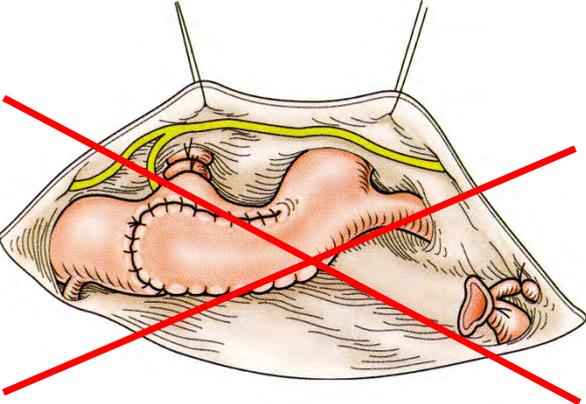
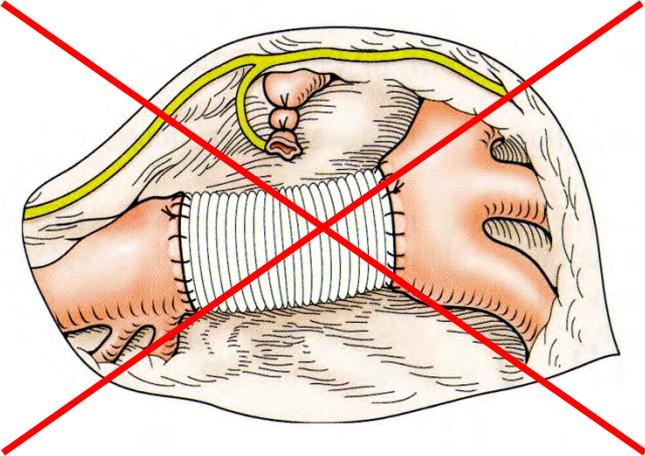
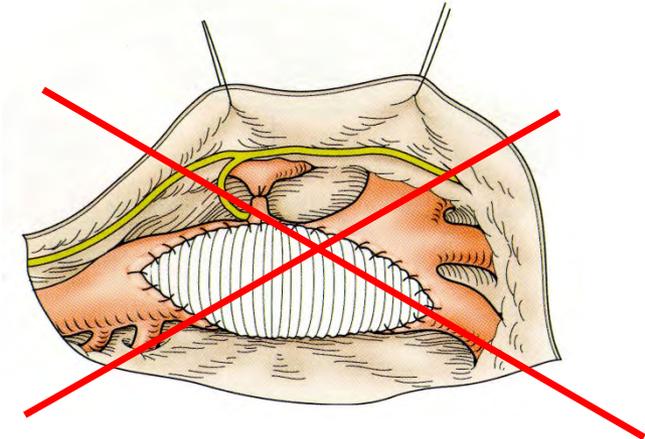
Quelle technique chirurgicale?

Réparation par Thoracotomie

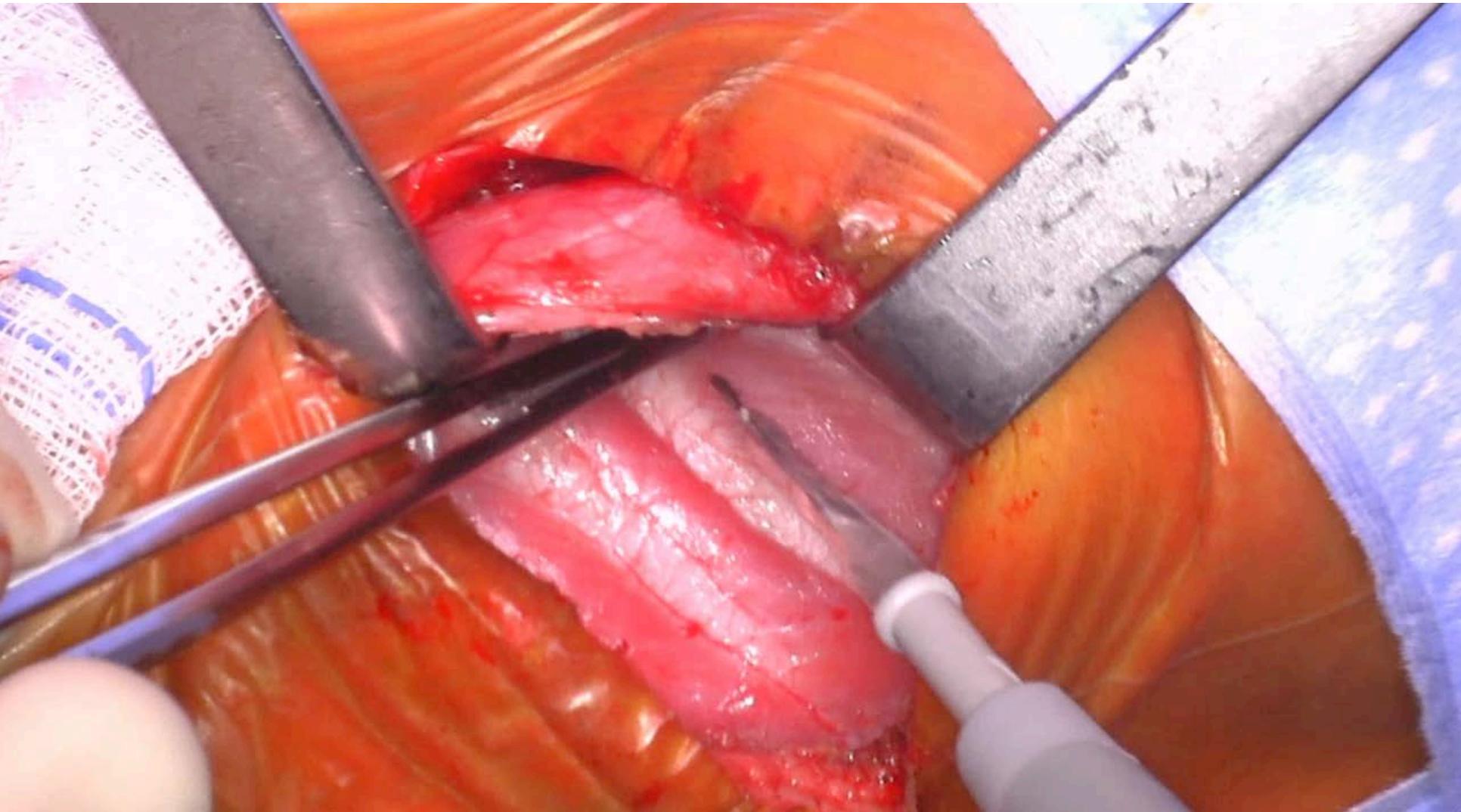
Résection anastomose étendue: Crafoord « modifié »

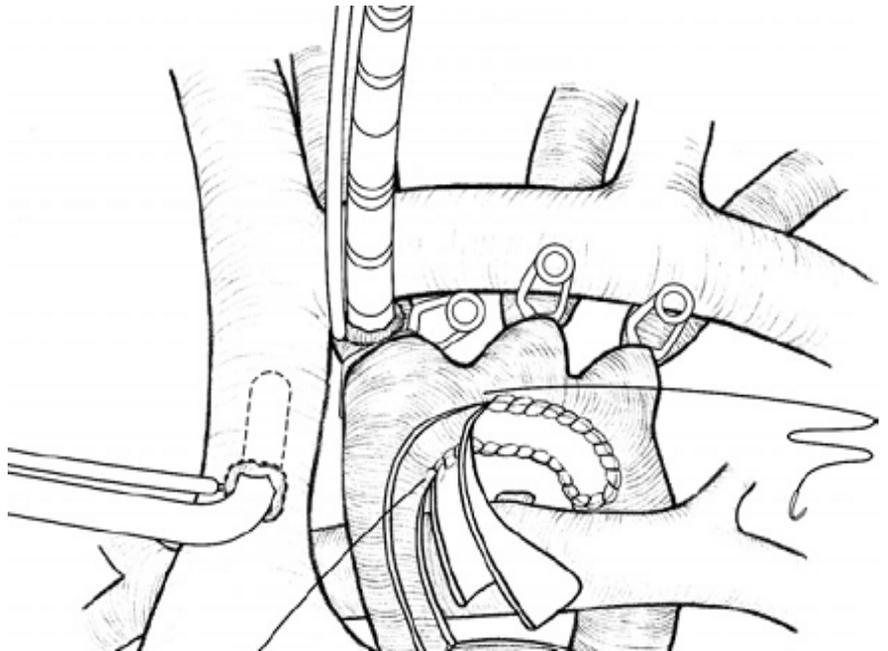


Réparation par Thoracotomie



cure de coarctation par thoracotomie

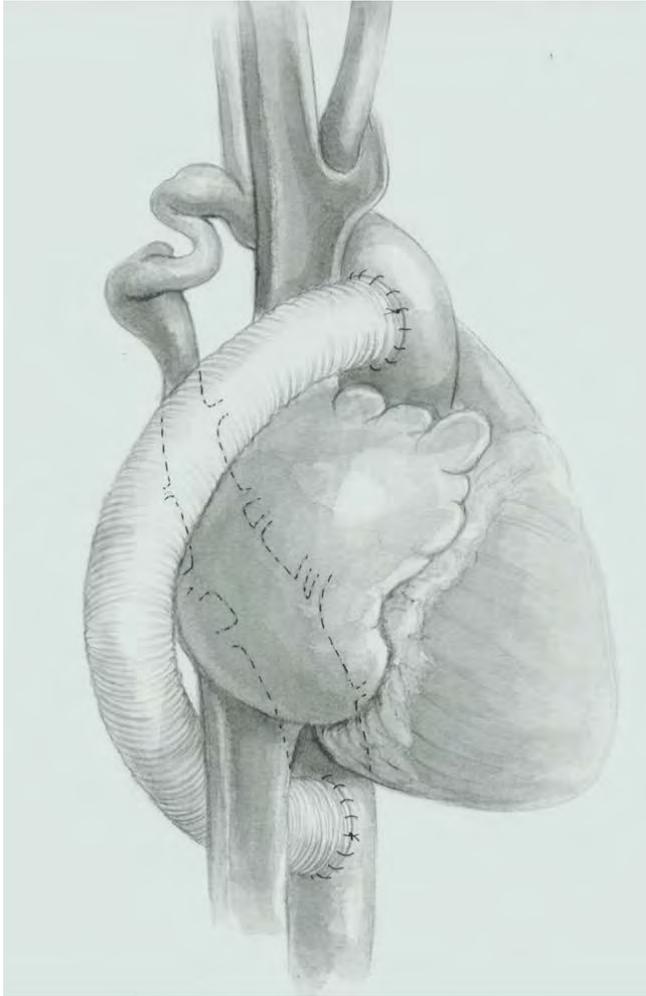




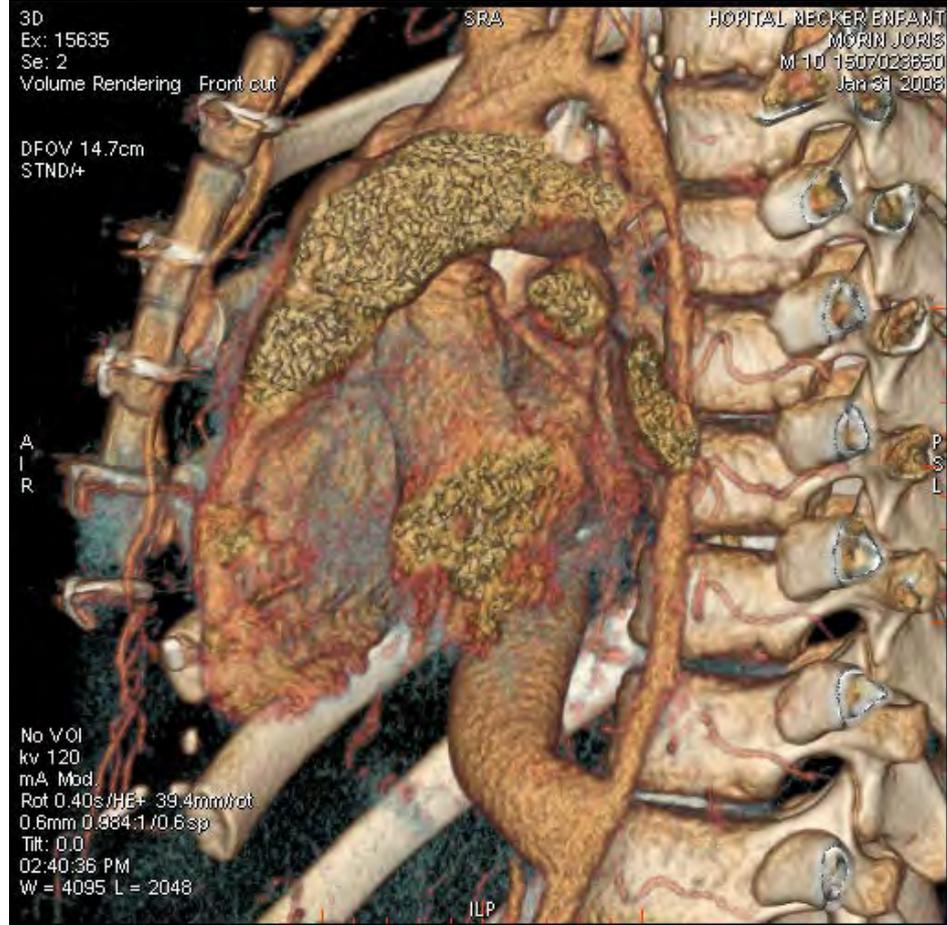
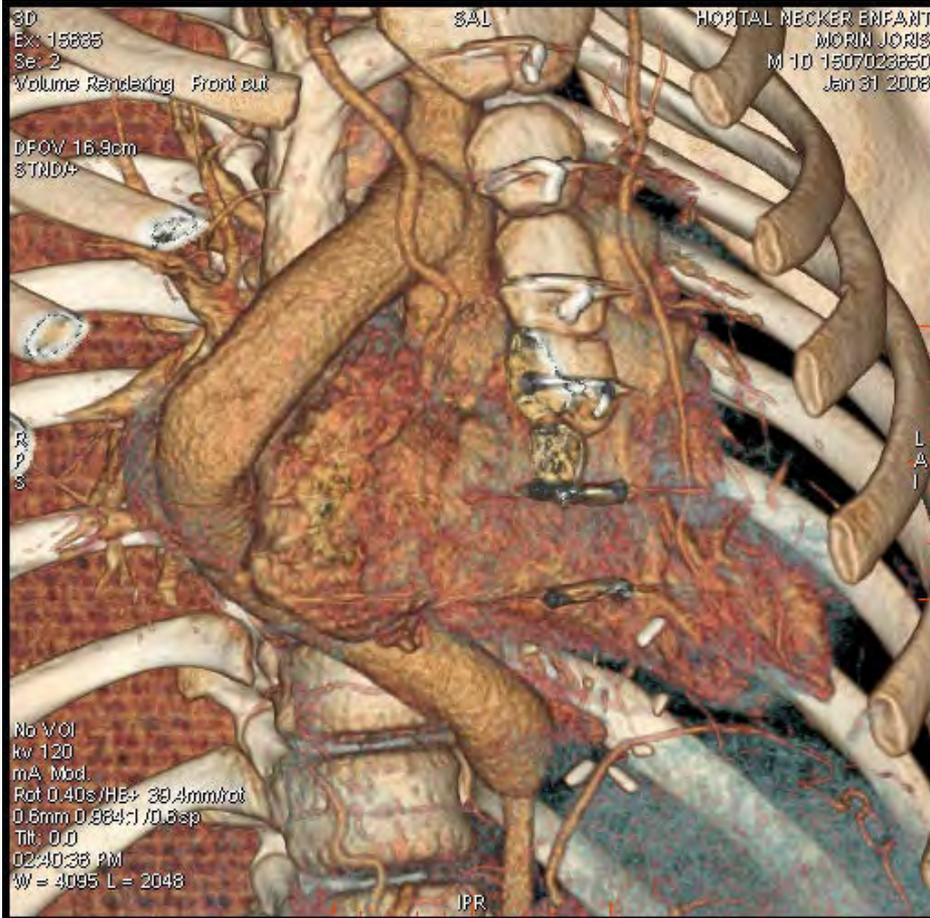


thorax
dissection rétro-sternale
anévrisme oculuse

Formes complexes d'hypoplasie PHACE syndrome



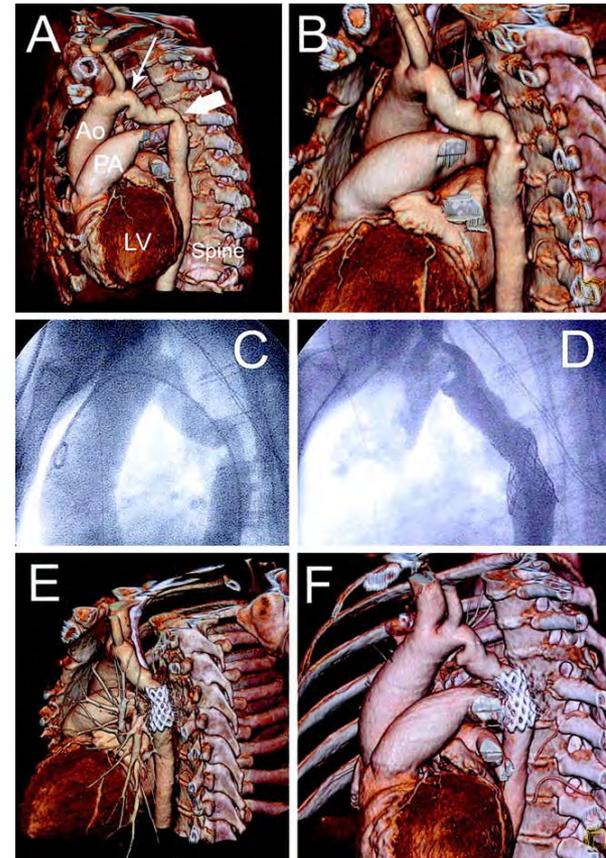
P: anomalie fosse Posterieure
Hemangiomes
Anomalies Arteriellles
Coarctation de l'aorte
Eye : anomalies oculaires
S: anomalies Sternales





Au-delà de la période néonatale

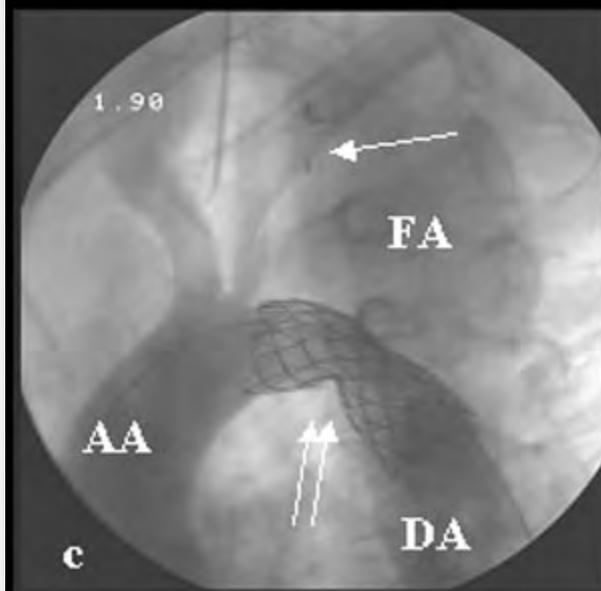
- Indications opératoires idem
- Méthodes
 - Chirurgie chez l'enfant ?
 - Stenting direct chez l'adulte ?



Complications immédiates

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





Complications précoces

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

Complications tardives



Hypertension artérielle tardive

Position du problème

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

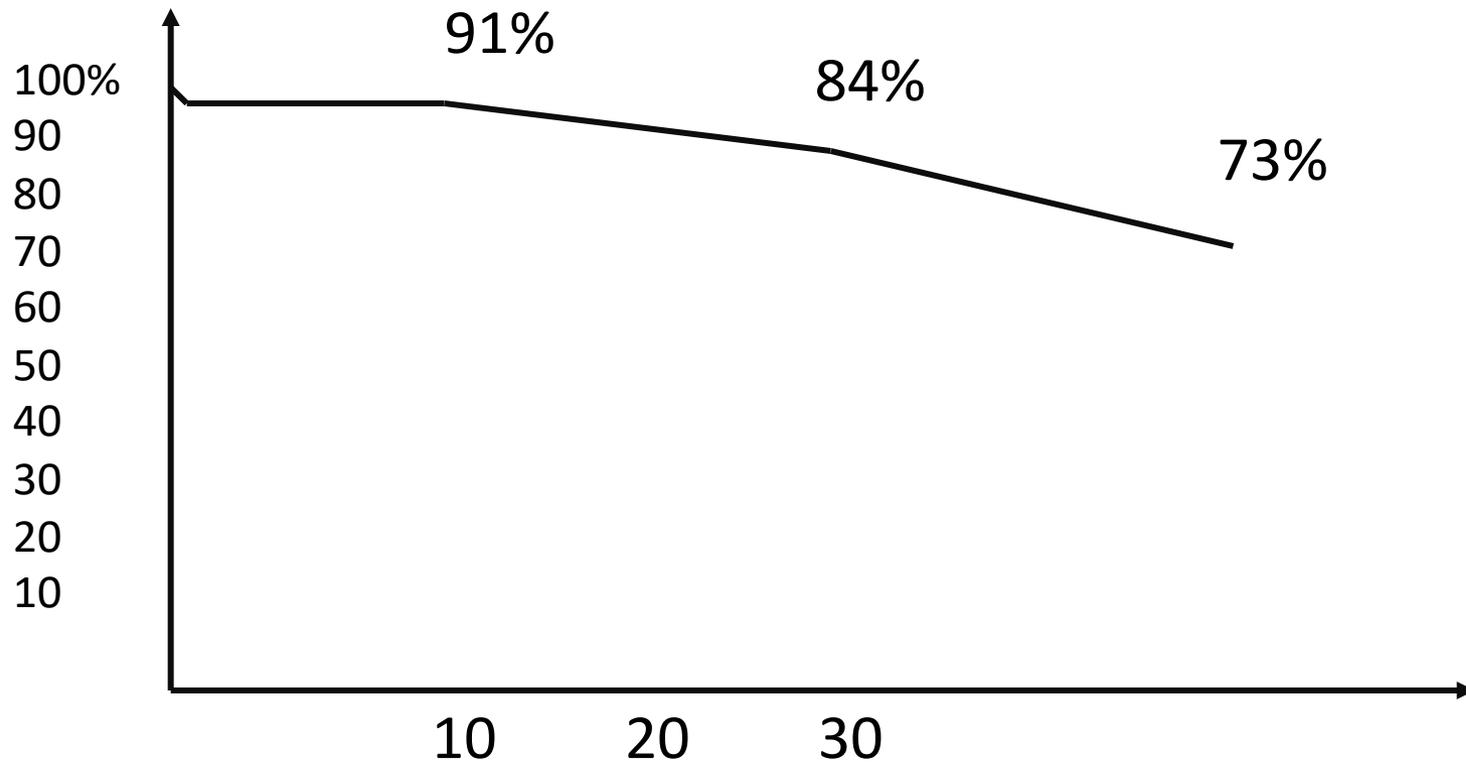
- 87 décès/646 pts

- âge moyen du décès 38 ans

Cohen M 1989

Mortalité et mortalité cardiovasculaire

Survie actuarielle



Mortalité cardiovasculaire

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

Prévalence de l' HTA

- 49% d' HTA à 17 ans *Kappetein 1994*
- 68% d' HTA à 30 ans *Presbitero 1987*

Coarctation abdominale

