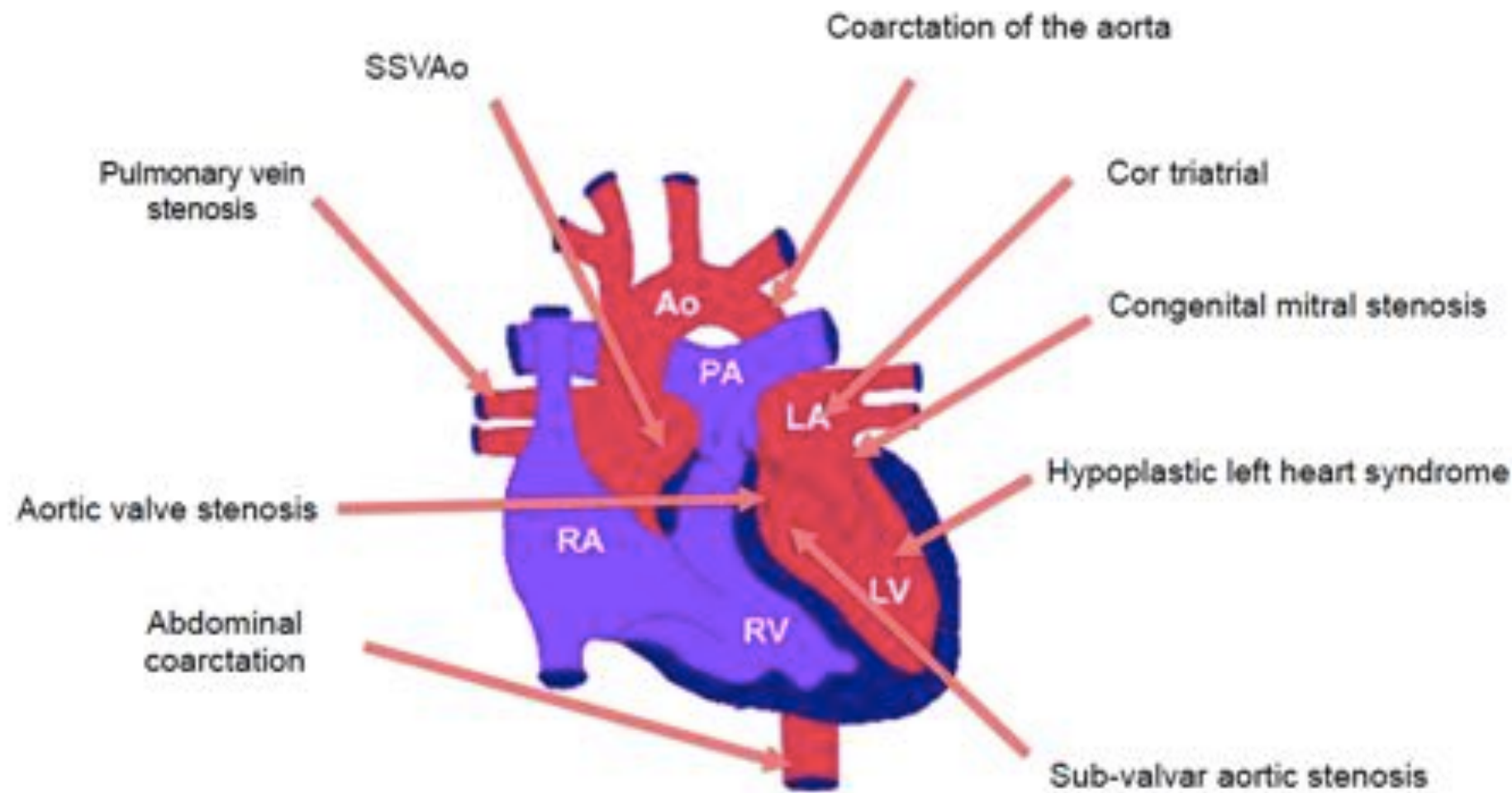


OBSTACLES GAUCHES

C Karsenty

CHU Toulouse

07/06/2019



OBSTACLES

EN AMONT DE LA VALVE MITRALE

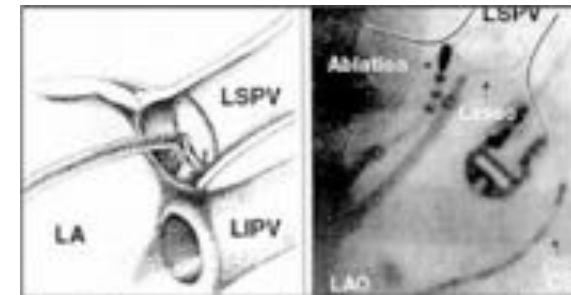
Sténose des veines pulmonaires

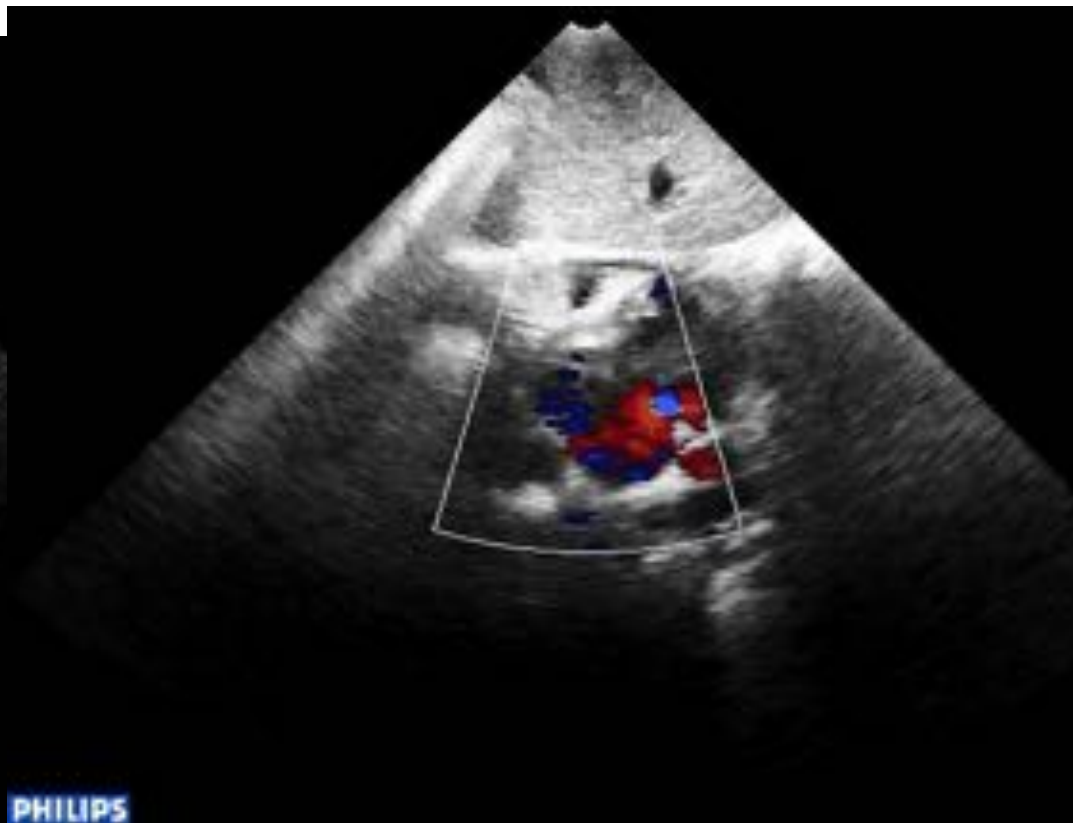
Cœur triatrial

Membrane supramitrale

Sténose veines pulmonaires

- Sténose isolée congénitale/primaire) d'une/des veine pulmonaire (0.4%, 50% associée)
- Secondaire :
 - Collecteur anastomose RVPAtotal sténosé
 - Sténose postablation FA
 - Acquis: compression extrinseque
- → Révélation **respi** : pnp, hémoptysie
- HTAP postcapillaire

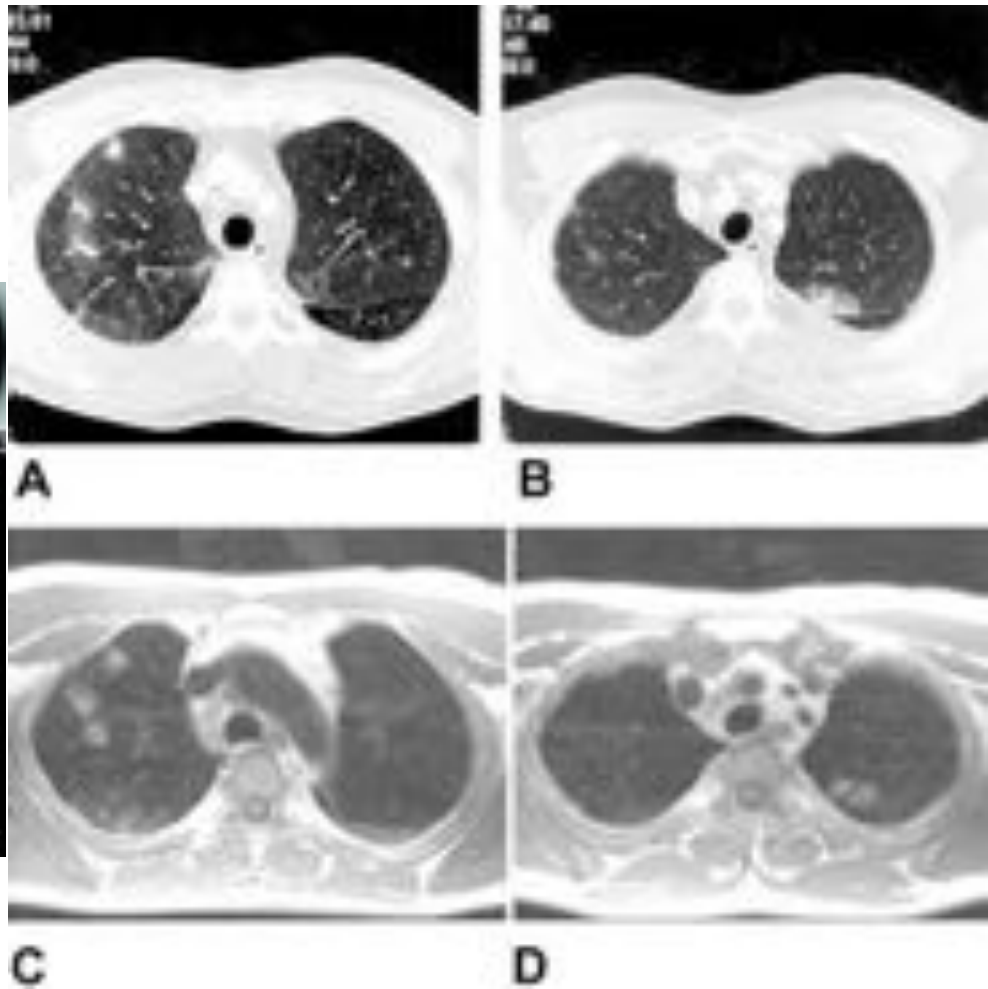
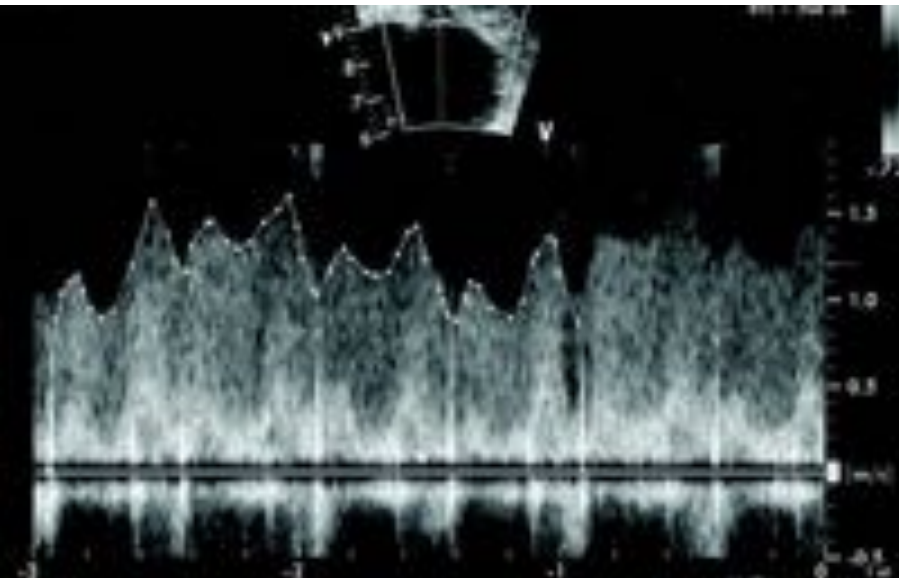




Sténose veines pulmonaires

- Aspect parenchyme pulmonaire

Echo - doppler



Sténose veines pulmonaires

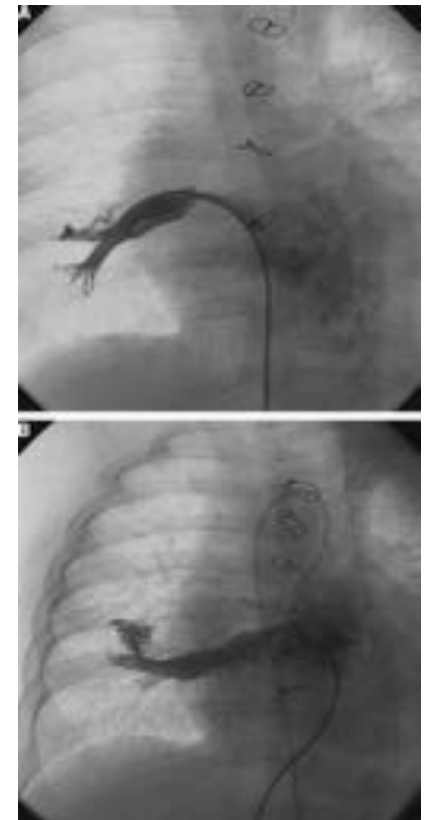
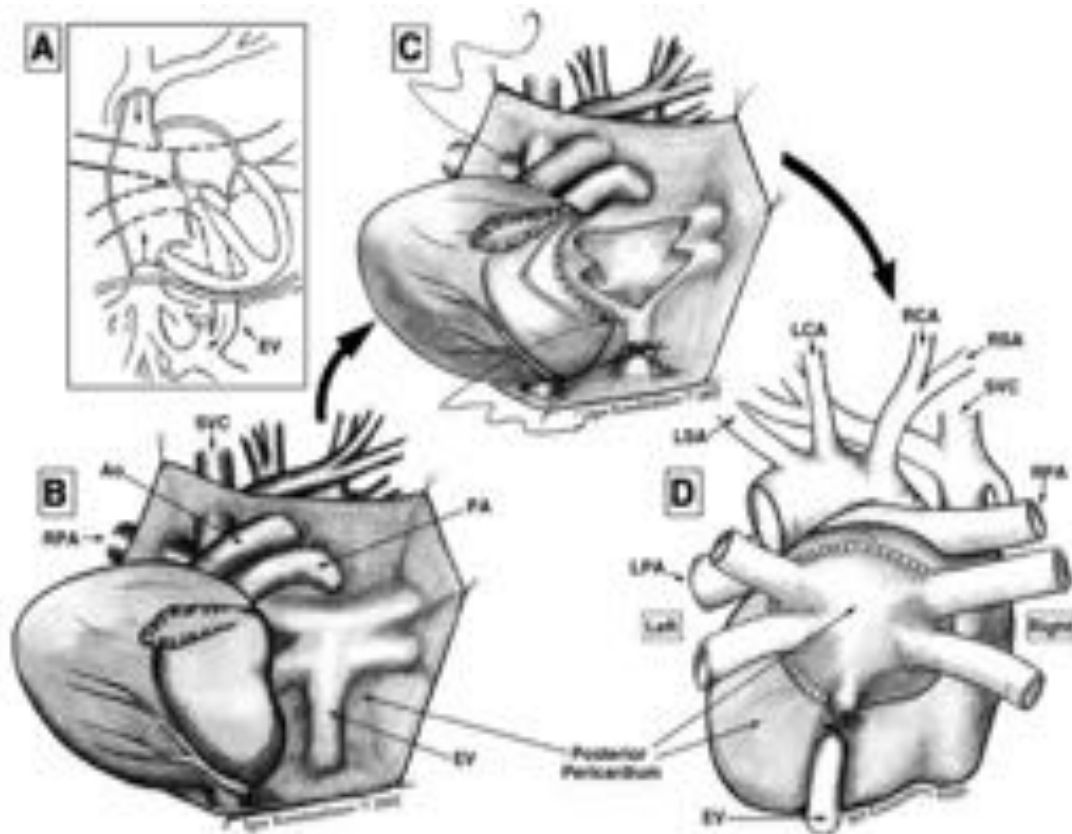
- Scanner injecté ++



Sténose veines pulmonaires : Traitement

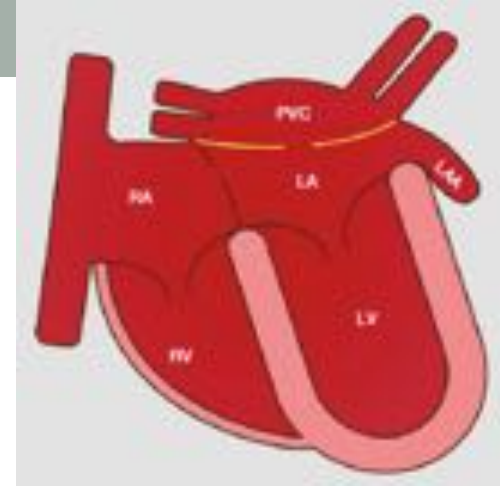
- Chir (suturless)

- Kt (adulte / répétés)
- Ballon haute pression
- Vs Stent (>10mm)

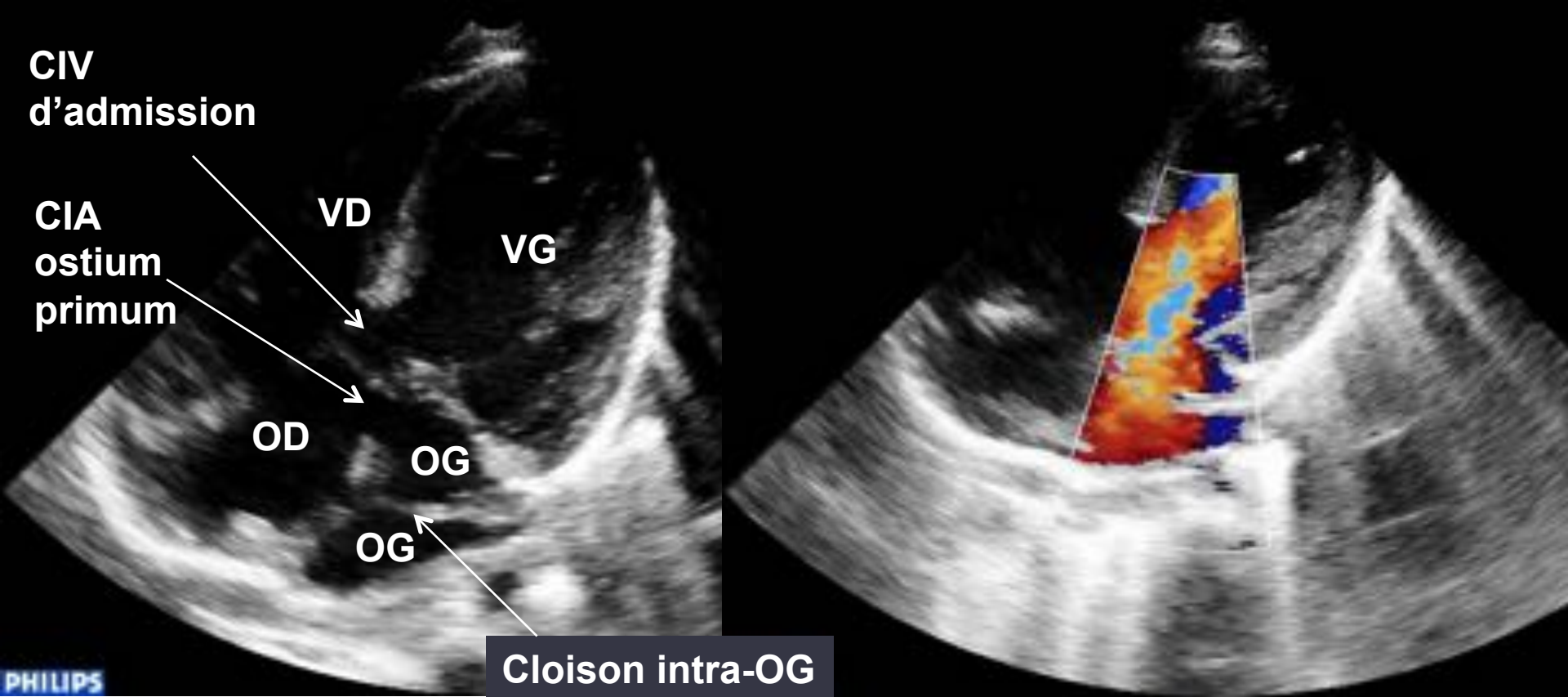


Obstacle dans l'OG:
Cœur triatrial
Membrane supramitrale

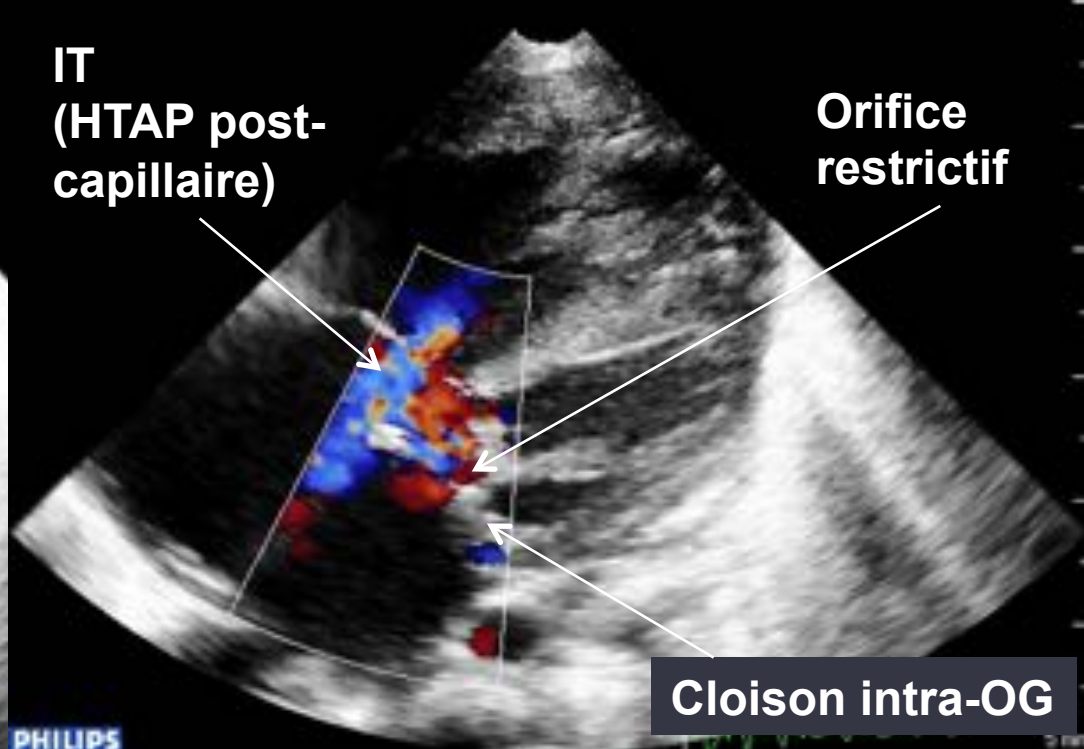
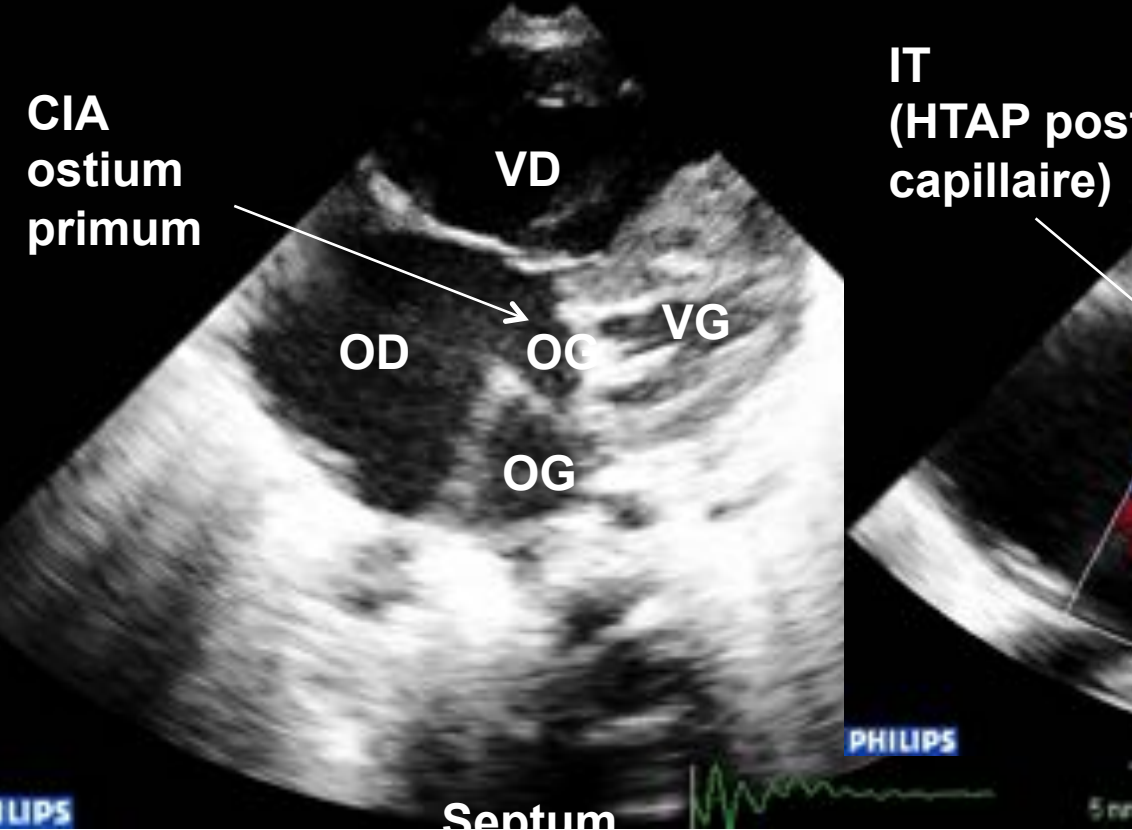
Cœur triatrial



- <0,1% des cardiopathies congénitales
- Cloison fibro-musculaire séparant l'OG en 2 chambres :
 - une chambre **distale** : recevant les veines pulmonaires
 - une chambre **proximale** : en connexion avec l'auricule gauche et la valve mitrale (+/- FO)
- Sévérité : taille de l'orifice de la cloison (sténose mitrale)
 - Si restrictif : HTAP post-capillaire et possible bas débit cardiaque
- Possible lésions associées : RVPA, CIA, CAV, VCSG...



Cœur triatrial + CAV complet
4 cavités



Cœur triatrial avec orifice restrictif + CAV partiel
 4 cavités &
 3D vue atriale

Coeur triatrial

Echo +++

Imagerie si besoin



Coeur triatrial : traitement

Depend symptomes ++
Obstruction ou non??

Symptomes

- Poor growth (children)
- Dyspnea
- Pulmonary HTN
- Atrial fibrillation
- Hemoptysis

Age and severity of presentation depends on severity of LV inflow obstruction

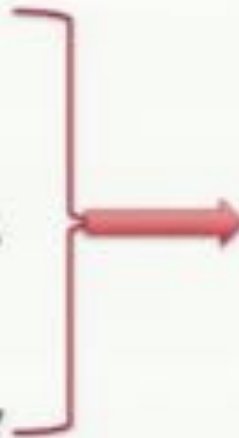
Cor Triatriatum – Management

- Asymptomatic & non-obstructive CTS
 - No pathologic change in cardiac chambers



Conservative management

- Development of Symptoms:
 - Atrial fibrillation
- Cardiac changes
 - Elevation in inflow gradients or PA pressures
 - LA dilation or thrombus
- Need for additional cardiac surgery



Consider Surgical Resection

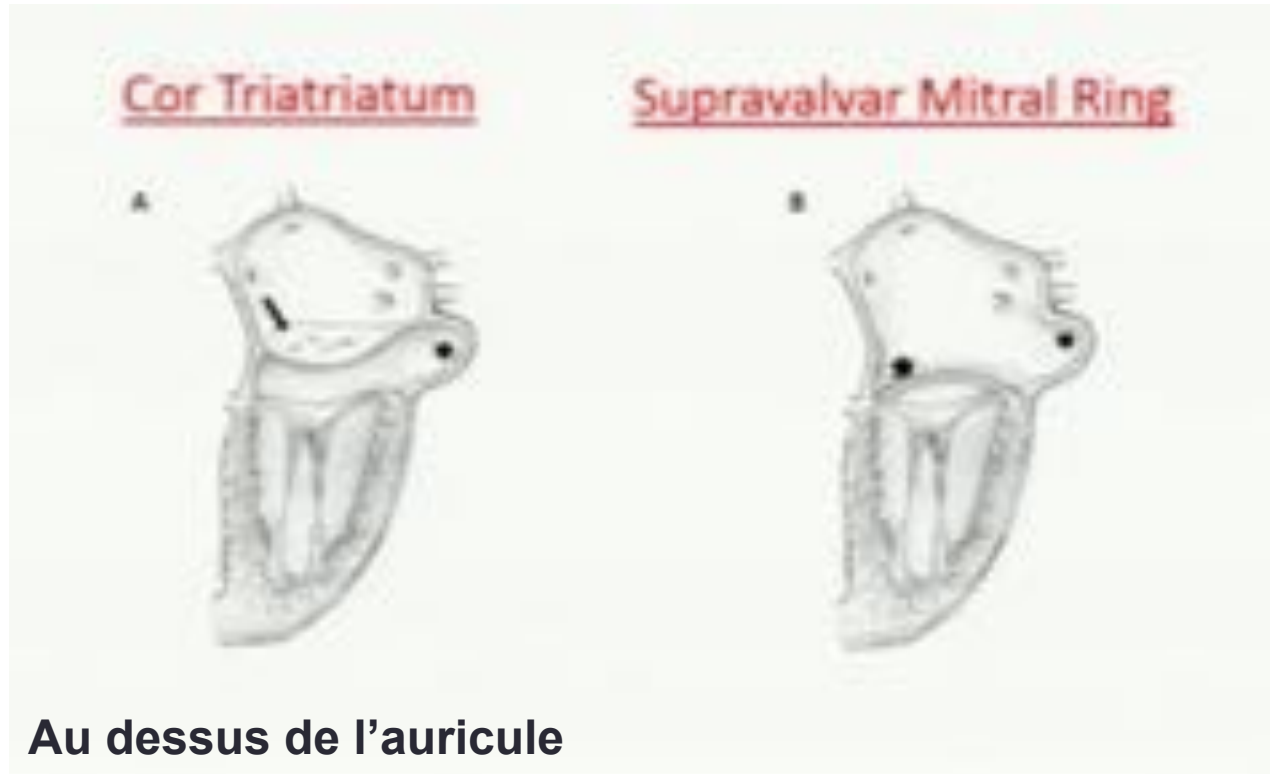
Excellents résultats

Coeur triatrial : adulte

I	B-NR	1. Adults presenting with cor triatriatum sinister should be evaluated for other congenital abnormalities, particularly ASD, VSD, and anomalous pulmonary venous connection. ^{14.2.1-1}
IIa	B-NR	2. In adults with prior repair of cor triatriatum sinister and recurrent symptoms, it is reasonable to evaluate for pulmonary vein stenosis. ^{14.2.1-2}
Therapeutic		
I	B-NR	3. Surgical repair is indicated for adults with cor triatriatum sinister for symptoms attributable to the obstruction or a substantial gradient across the membrane. ^{14.2.1-3}

> 8 mmHg

Coeur triatrial vs supravulvaire



Au dessus de l'auricule

Vers mitrale en diastole

S'éloigne mitrale en diastole
Mvt feuillets anormaux

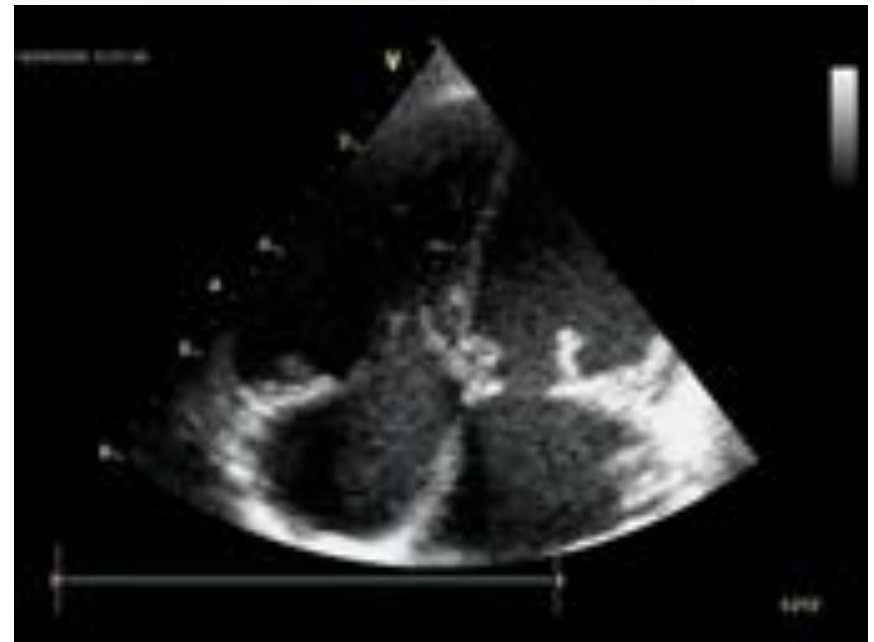
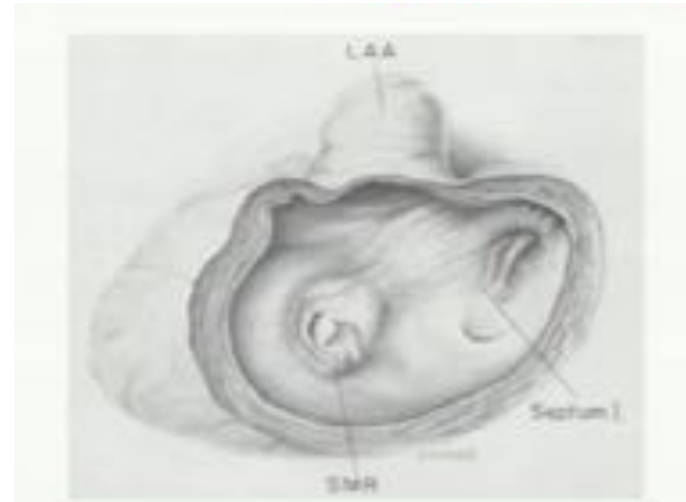
Stenose supravulvaire mitrale

Rare : 0.5% des CC

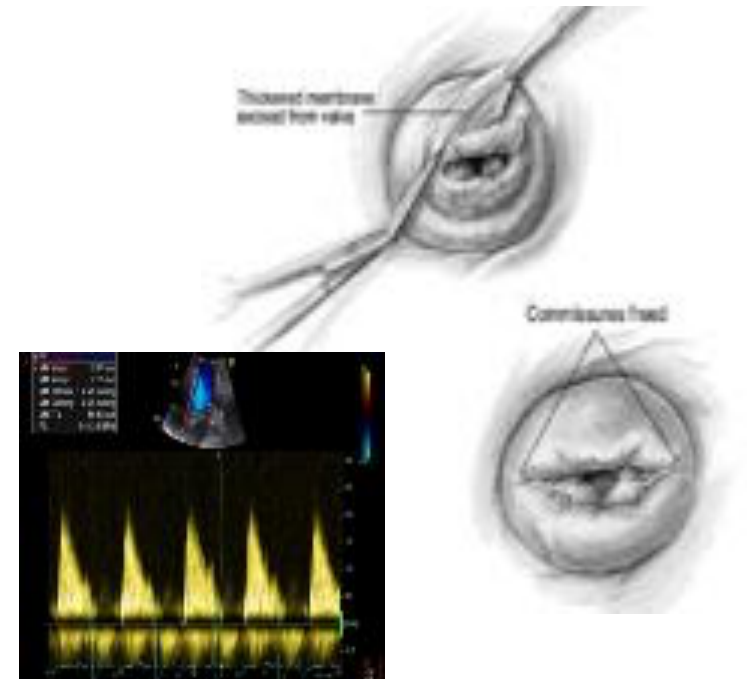
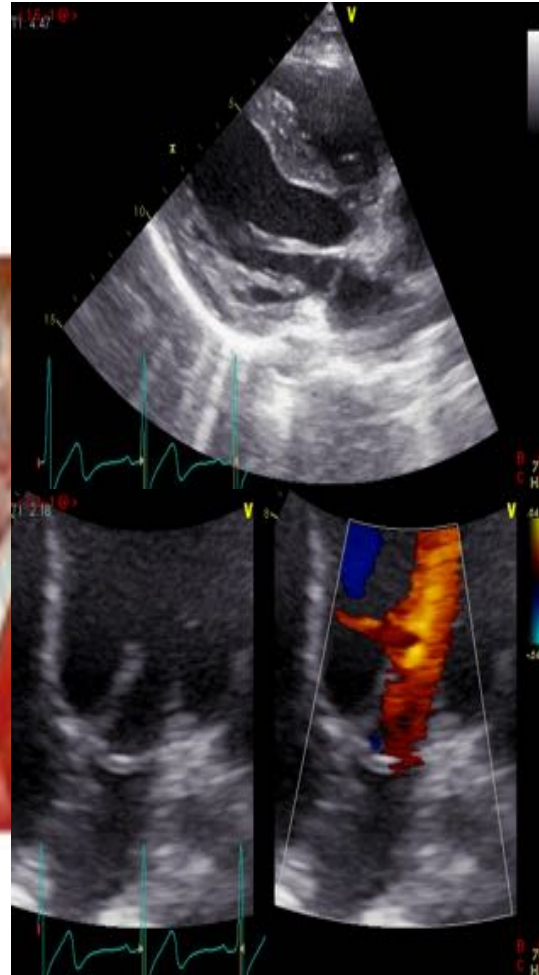
Physio : stenose mitrale +/- fuite

Souvent associée (Shone, CIA...)

Diagnostic : ETT++

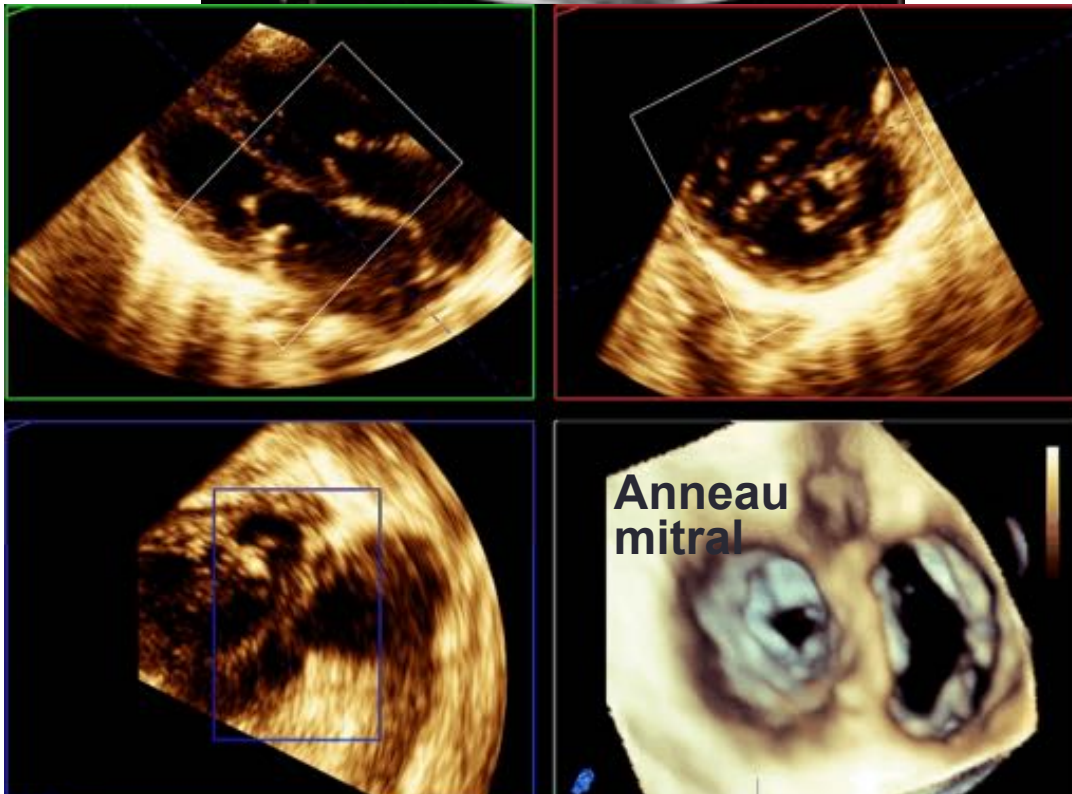


Membrane supra-valvulaire



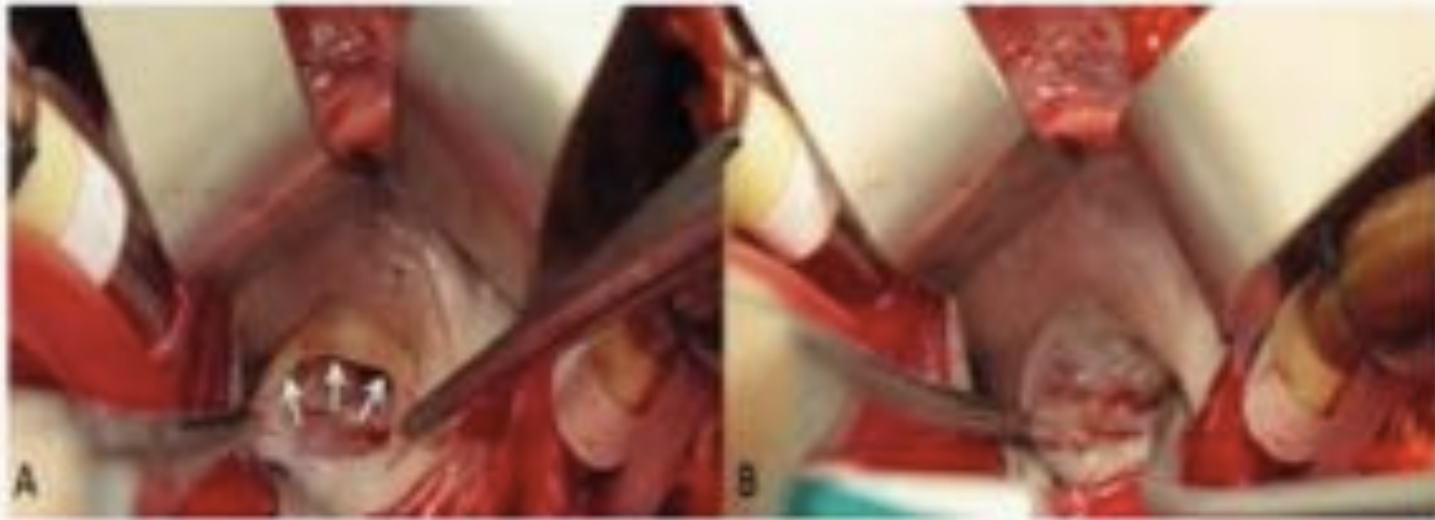


PSPA



Stenose supravulvaire mitrale

Traitement : si isolée = excellent résultats



Operative view of supravulvar mitral ring (white arrows) (A) and after its resection (B).

J Card Surg. 2015 Jul;30(7):591-4

Si associée : parfois RVM

PATHOLOGIES MITRALES

Anomalies des feuillets

Anomalies des cordages

Anomalies des piliers

Anomalies mitrales

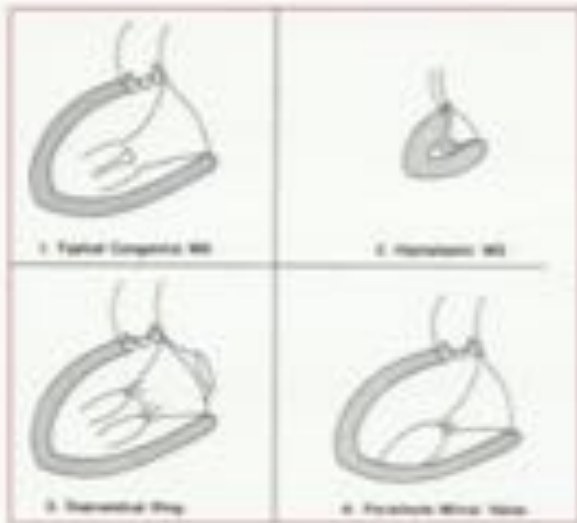
- Rare:
 - 0.36 à 0.5% des cardiopathies congénitales:
 - la fente, le parachute, le hamac, la fusion piliers-commissures et le double orifice mitral.
 - Formes complexes :
 - Le complexe de Shone
 - Autres malformations cardiaques : CAV, malformations cono-troncales
 - Formes acquises :
 - Infx: RAA
 - Ischémique : kawa...
 - Maladies du tissu conjonctif : Marfan, Ehler-Danlos
 - Postop: CAV
- **Fuite ou sténose ou mixte**
- sténose mitrale = gradient télédiastolique OG-VG → HTAP post-capillaire (S. IC)

	Cases encountered	% Frequency
Frequency of lesions		
Cleft mitral valve	58*	0.001
Supramitral ring 1 PMs	35*	0.000583
Supramitral ring 2 PM	22*	0.000367
Double orifice mitral valve	19*	0.00032
Mitral stenosis 1 PM	51*	0.00085
Mitral stenosis 2 PM	72*	0.0012
Mitral prolapse	44†	0.0012
Mitral straddle	5†	0.00014

Banerjee A *et al.*, Am J Cardiol, 1995
Silverman *et al.*, CIY, 2014

Stenose mitrale congenitale : classifications

Ruckman & Van Praagh



Classification and Frequency of 48 Cases of Congenital Mitral Stenosis

Frequency	Type of Congenital Mitral Stenosis	Cases (No.)	Percent of Series
1	Typical	24	48
1	Horseshoe	20	41
1	Triangular	1	2
1	Pericardial mitral valve	1	2
4	Total	46	95

AJG 1978; 42: 592.

Congenital Mitral Stenosis Classification



Catherine M Ofri Heart 2003;89:100-108

Carpentier

Table 4. Carpentier's classification of congenital mitral stenosis (20)

Stenosis pattern	No. patients (%)
Type I: Prolapsed mitral valve	7 (35)
Annoy chordae	1
Papillary chordae	1
Bicuspid mitral	1
Type II: Prolapsed mitral valve	10 (50)
Prolapsed mitral	10
Bicuspid mitral	1
Type III: Prolapsed mitral valve	2 (10)
Prolapsed mitral	2

1. Prolapsed mitral valve: Prolapsed mitral valve, not all of these could be classified as the category "bicuspid mitral valve" because the posterior leaflet was the papillary muscle valve.

Congenital Heart Surgery Nomenclature and Database Project

CONGENITAL MITRAL VALVE DISEASE (CMVD): An anatomic abnormality of the mitral valve of a functional left ventricle that results in structural and/or functional alterations.

CMVD, TYPE 1—SUPRAVALVAR: An abnormality of the mitral valve above the level of the leaflets or annulus.

CMVD, TYPE 2—VALVAR: An abnormality of the mitral valve at the level of the leaflets or annulus.

CMVD, TYPE 3—SUBVALVAR: An abnormality of the mitral valve at the level of the chordae tendineae or the papillary muscles.

CMVD, TYPE 4—MIXED: An abnormality of the mitral valve that is best described as a mixture or combination of two or more of the above three types.

Lamberti et al. Ann Thorac Surg 2000;69:5132

Classification fonctionnelle



Type I (normaux): IM

- Anneau : dilatation

déformation

- Feuillelet : cleft
agénésie partielle

Type III (restrictif) a :
muscle papillaire normal : SM(± IM)

- Fusion pilier-commissure
- Raccourcissement des cordages
- Feuillelet redondant
- Anneau valvulaire
- Hypoplasie de l'anneau



Type II (prolapsus): IM

- Élongation des cordages
- Élongation des piliers
- Agénésie de cordages

Type III b : muscle papillaire anormal : SM(± IM)

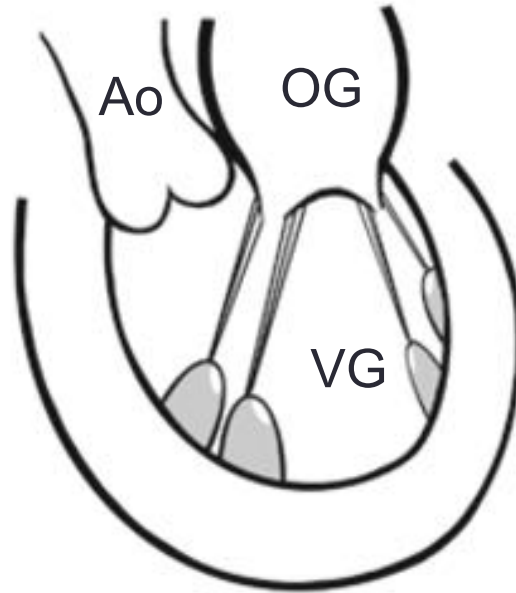
- Mitrale parachute
- Hamac
- Hypoplasie muscle papillaire

Carpentier *et al*, Reconstructive valve, 2010
Mitruka *et al* Ann Thorac Surg 2000

Anomalies des feuillets

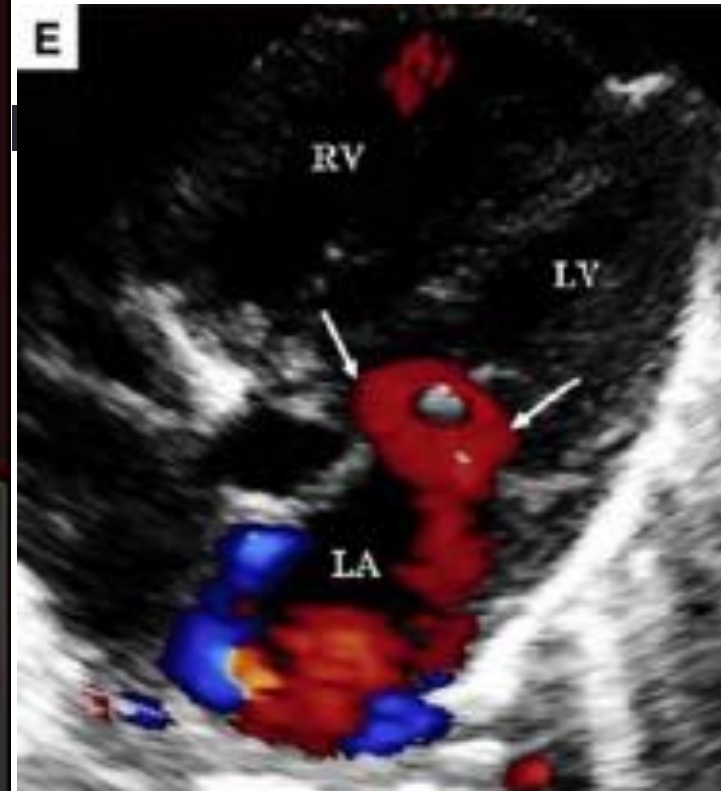
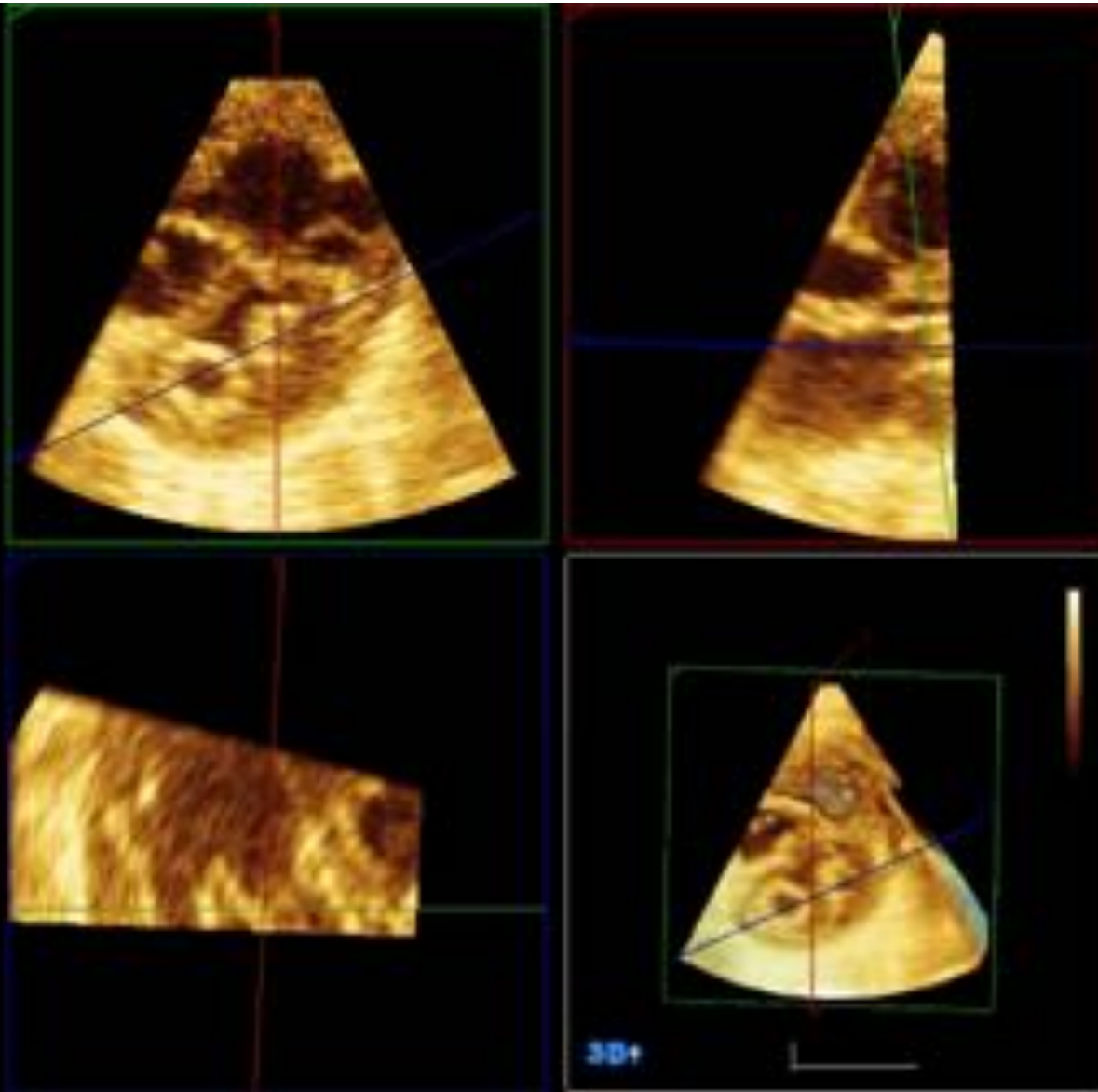
Double orifice mitral

- **IM** et/ou **sténose mitrale**
- Orifice médial et orifice latéral
- Double flux antérograde par la mitrale



Anomalies des feuillets

Double orifice mitral

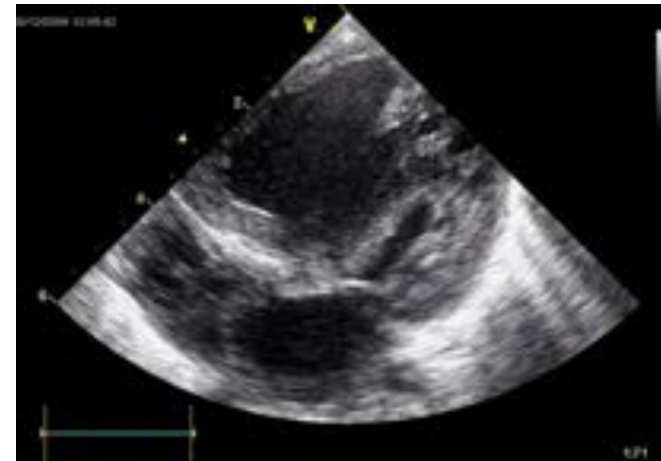


4 cavités
& 3D PSPA

Anomalies des feuillets

Autres

- Dysplasie valvulaire mitrale : feuillets épaissis
- Fente (Fuite)
- Hypoplasie de l'anneau : Z-score < -2

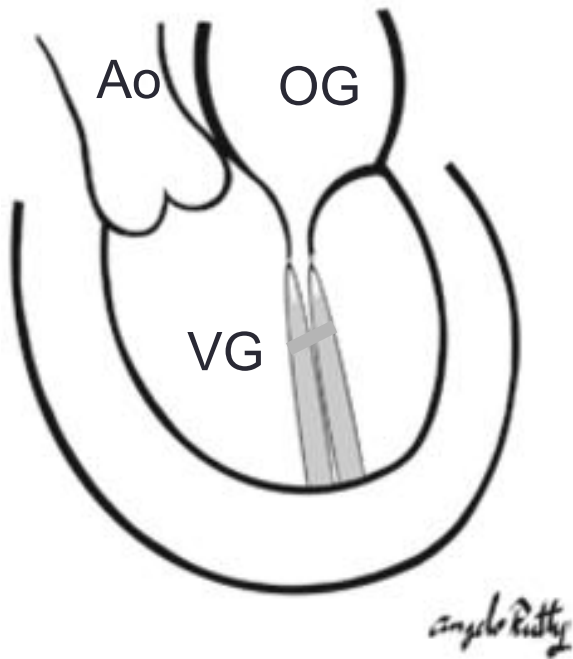


M. D Pettersen et al., "Regression Equations for Calculation of z Scores of Cardiac Structures in a Large Cohort of Healthy Infants, Children, and Adolescents: An Echocardiographic Study," *Journal of the American Society of Echocardiography* 21, no. 8 (2008): 922-934.

Anomalies des cordages

Valve mitrale en arcade/hammac

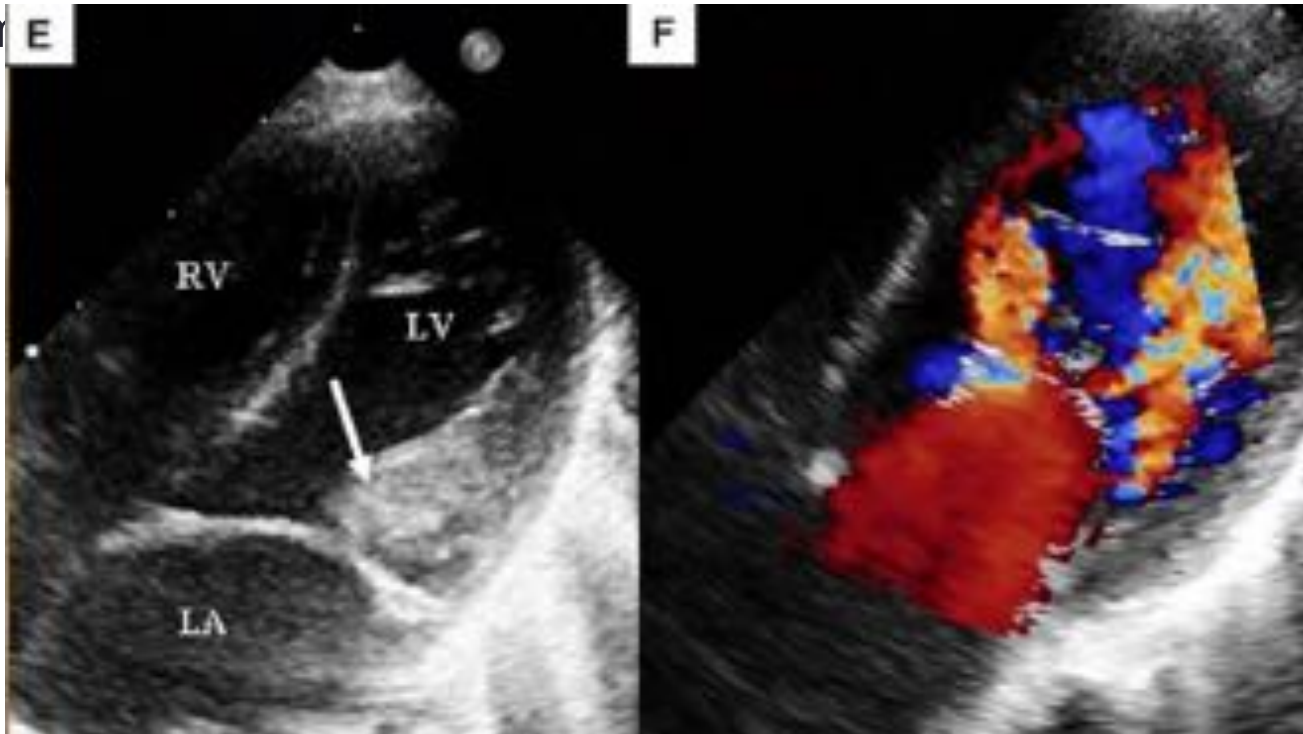
- **IM** et/ou **sténose mitrale**
- Cordages épais et courts →
 - espaces inter-cordages réduits avec multiples jets en doppler couleur
 - mouvement restrictif des feuillets



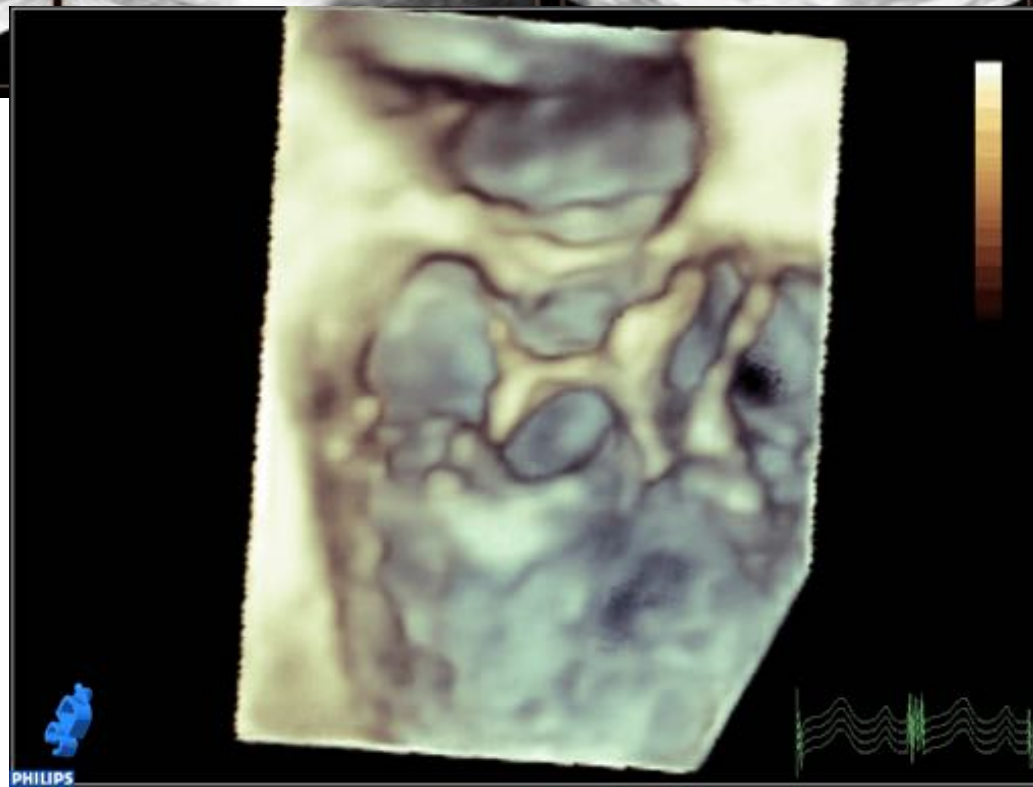
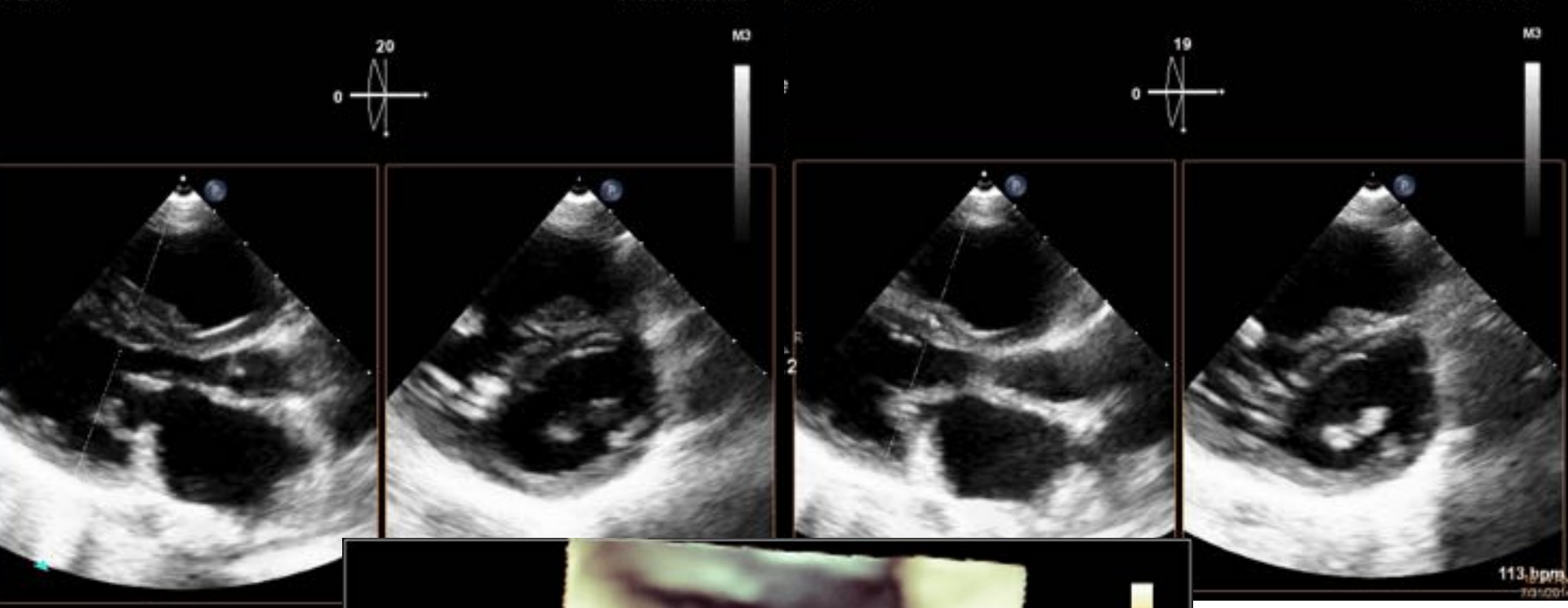
Anomalies des cordages

Valve mitrale en arcade/hammac

- **IM** et/ou **sténose mitrale**
- Cordages épais et courts →
 - espaces inter-cordages réduits avec multiples jets en doppler couleur



4 cavités

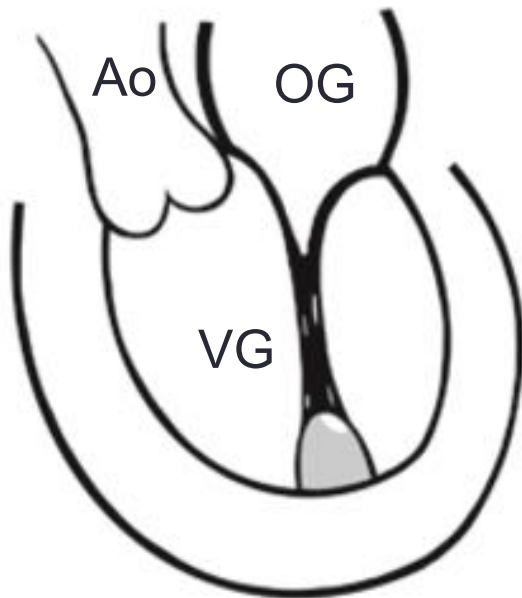


**PSGA, PSPA
& 3D vue VG**

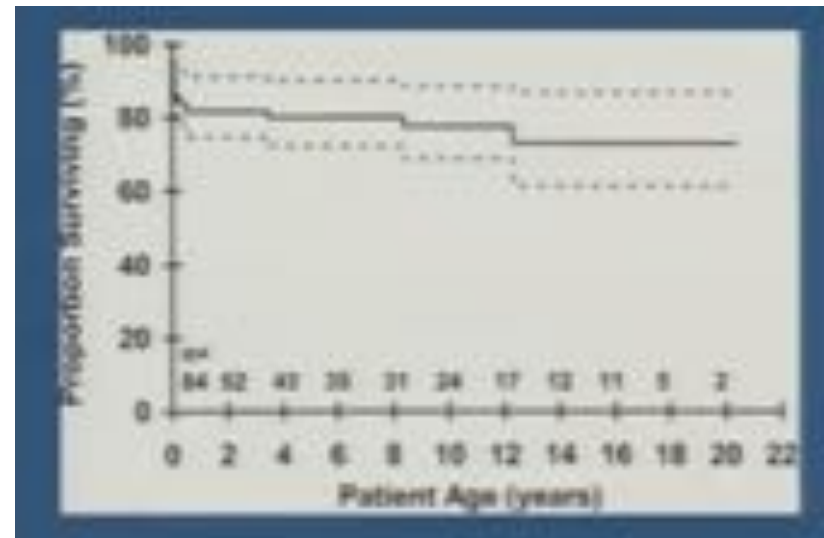


Valve mitrale parachute

- **IM** et/ou **sténose mitrale**
- Un seule muscle papillaire
(ou Parachute-like : 2 piliers mais un pilier prédominant)
- Cordages peu mobiles → mouvement restrictif des feuillets



Histoire naturelle : pediatrique





**4 cavités
& PSPA**

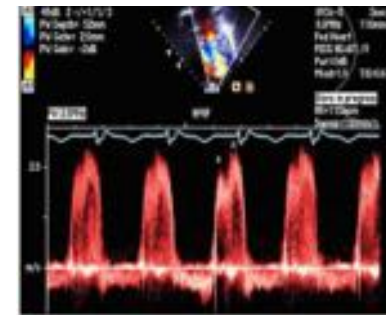
Sténose mitrale congénitale : ETT

- Mécanisme :
 - Supra-valvulaire mitral
 - Valves mitrales de face
 - Anneau mitral
 - Appareils sous valvulaire mitral
- + Chambre de chasse VG
 - + Taille du VG
- Sévérité : Gradient moyen transmitral
 - Modérée si gradient moy = 10-15 mmHg
 - Sévère si gradient moy >15 mmHg
- Retentissement :
 - PTDVG élevée
 - HTAP post-capillaire
- Recherche de lésions associées

Grosse-Wortmann et al, JTCS, 2008

Indications pédiatriques : différence avec l'adulte

- Mais limites
 - Pic de l'onde E et PHT et l'estimation des surfaces d'ouverture mal définies en Doppler
 - CIA
 - Diastole raccourcie chez l'enfant
 - Fusion E/A, $A > E$
 - Planimétrie & PISA non applicables
 - Car souvent étagé (pas que feuillets)



Banerjee A *et al.*, Am J
Cardiol, 1995

La stratégie

- Pondération : symptômes, HTAP, complexité, âge, taille de l'anneau, lésions associées
- Plastie mitrale (souvent palliatif = « gagner des années »)
 - Croissance de l'anneau
 - Capacité de récupération de la fonction ventriculaire
- RVM pédiatrique : mortalité et réintervention
 - Mismatch anneau natif/taille de la prothèse
 - Anticoagulation au long cours



Evaluation post-opératoire : ETO au bloc

- Dysfonction valvulaire résiduelle = Risque de réopération
 - IM ou RM
 - Moyenne à sévère
 - A la sortie d'hôpital

Résultats : plastie

Stellin G et al, J Thorac Cardiovasc Surg, 2010

- 93 patients (4.5 ans), 52% IM



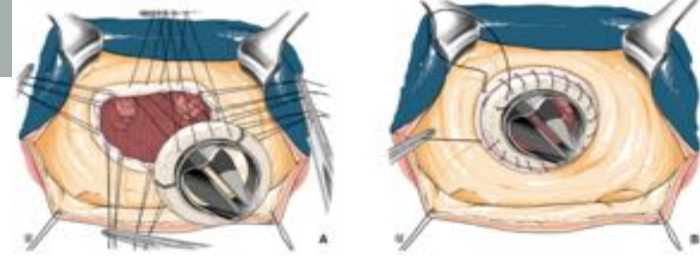
TABLE 5. Significant risk factors for several outcomes at logistic regression analysis

Outcomes	Variables				
	Complex MV (OR)	MV stenosis (OR)	Parachute MV (OR)	Before 1984 (OR)	Age, y (OR)
Any adverse outcome	3.173	2.520	–	–	–
Early death	–	–	7.122	18.40	0.636
Late death	–	6.973	–	–	–
Preoperative medical therapy	–	–	4.889	–	–
Reoperation on MV	–	–	6.817	–	–
Operation on MV before 1 y of age	–	7.219	–	–	–
Heart failure at follow-up	–	–	6.100	–	–
Arrhythmias	–	–	6.377	4.744	–

Variables significant at a level of $0.05 < P < .1$ are shown in *italics*. MV, Mitral valve; OR, odds ratio.

Résultats : RVM

- 94 children underwent 108 SAVR (4.2 years)(Necker)



Mortalité 30 jours : 9% (<1 an, Shone)
Survie 10 ans 79%.

Meilleur si position annulaire, taille
prothèse (23 vs 17mm)

C° : pacemaker (13%) saignement (8%),
thrombose (3%), AVC (3%) endocardite
(4%).

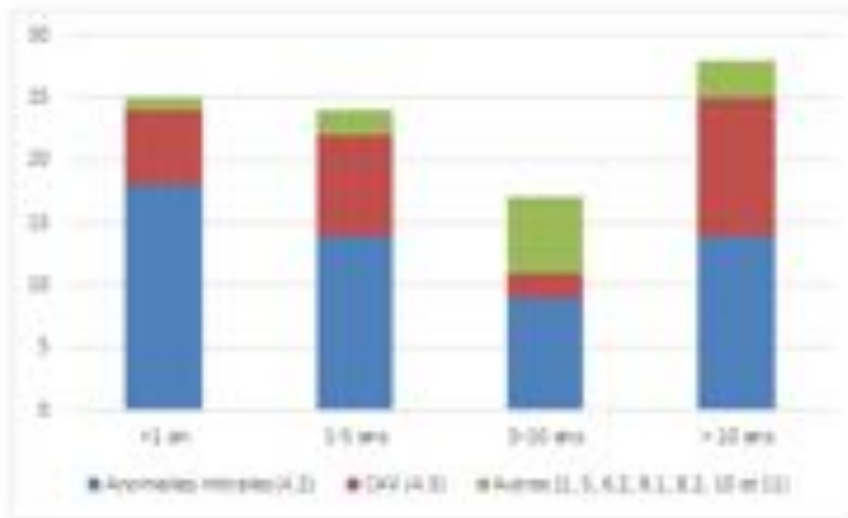
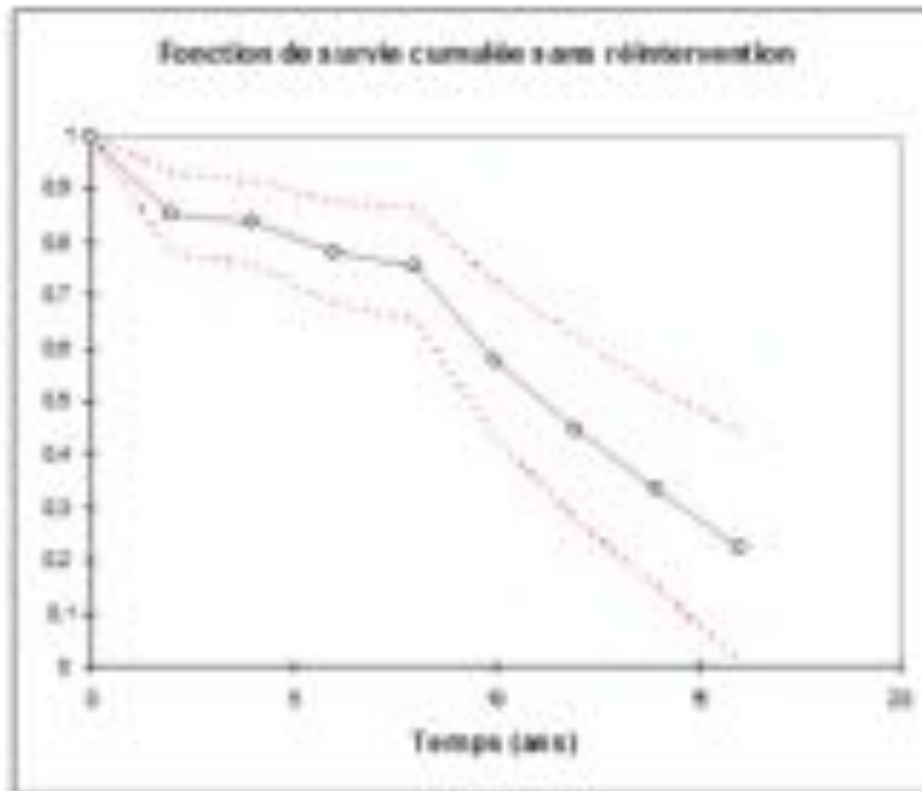


Figure 7 : Répartition de la cohorte RVM par sous-groupes ACC-CDH et par âge
Les pathologies complexes (bicuspidie, double discordance, VU, TGV et VDDC) de faible effectif ont
été regroupées dans une seule catégorie avec l'ALCAPA et la CMD.

Survie sans réintervention

La survie sans réintervention (RVM uniquement) à 1 an, 5 ans et 10 ans est respectivement de 85%, 78% et 58%. (Figure 12).



Valve Mitrale

- Les indications tiennent compte de :
 - L'âge
 - La complexité
 - Symptomes
- Bilan pré-opératoire : penser aux lésions associées (complexe de Shone)
- Peropératoire : avant tout c'est l'exposition chirurgicale qui compte
- ETO en fin de procédure est capitale : ne pas laisser sortir un malade du bloc avec une IM ou un RM moyen à sévère
- Surveillance car risque de réintervention
 - sur la mitrale chez l'enfant
 - Sur la "region" aortique chez l'adulte

OBSTACLES EN AVAL DE LA VALVE MITRALE

Sténose sous-valvulaire aortique

Sténose valvulaire aortique

Sténose supra-valvulaire aortique

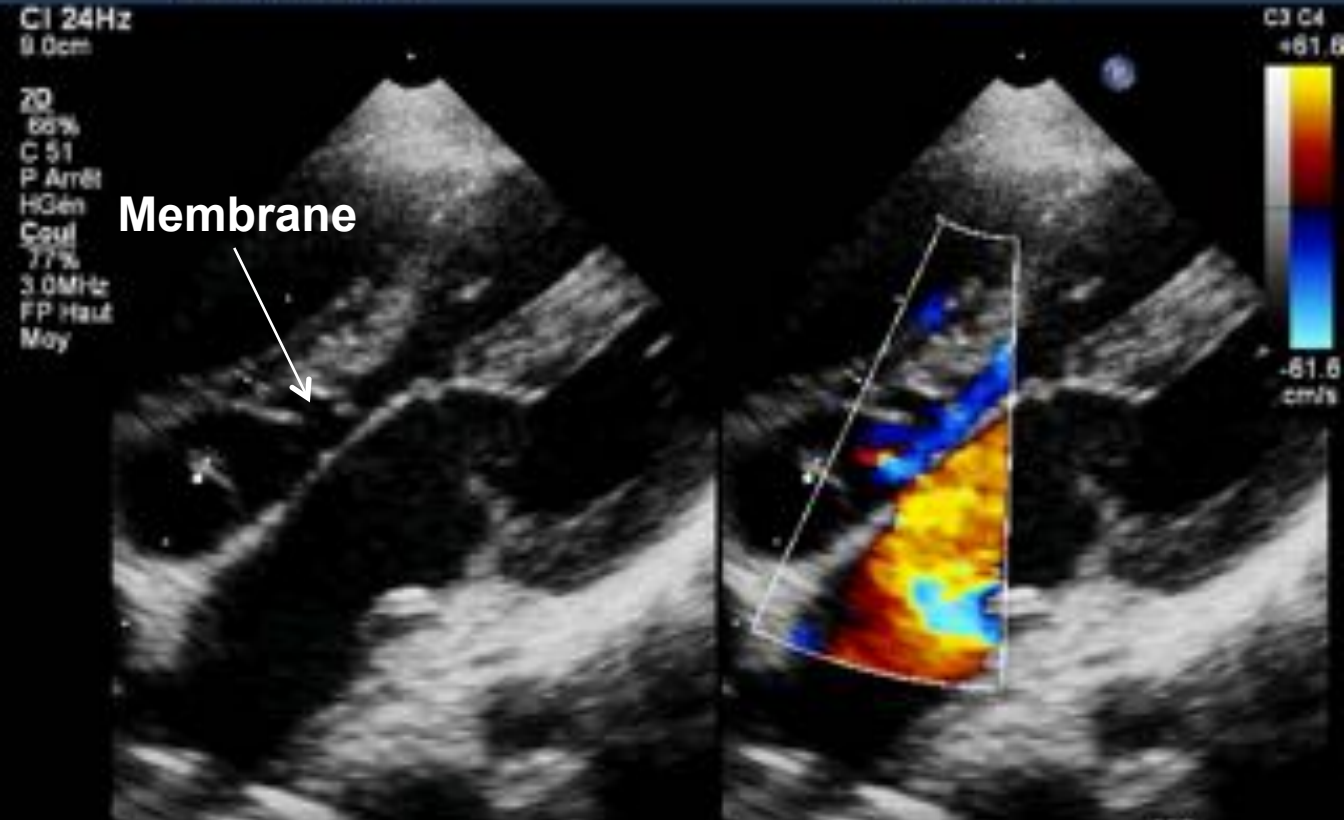
Coarctation de l'aorte

Sténose sous-valvulaire aortique

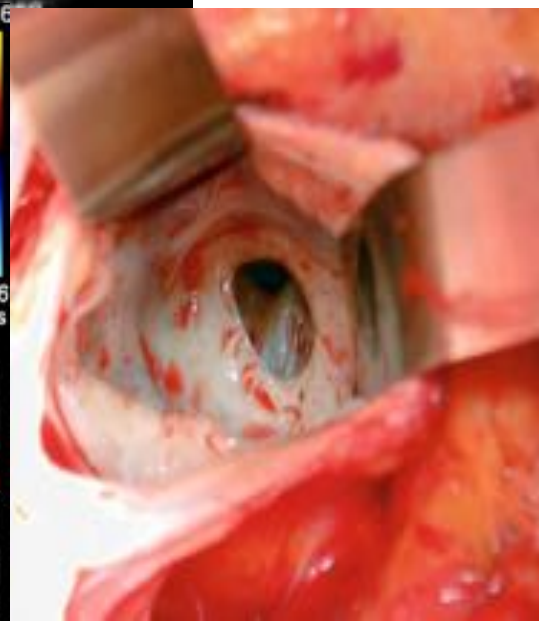
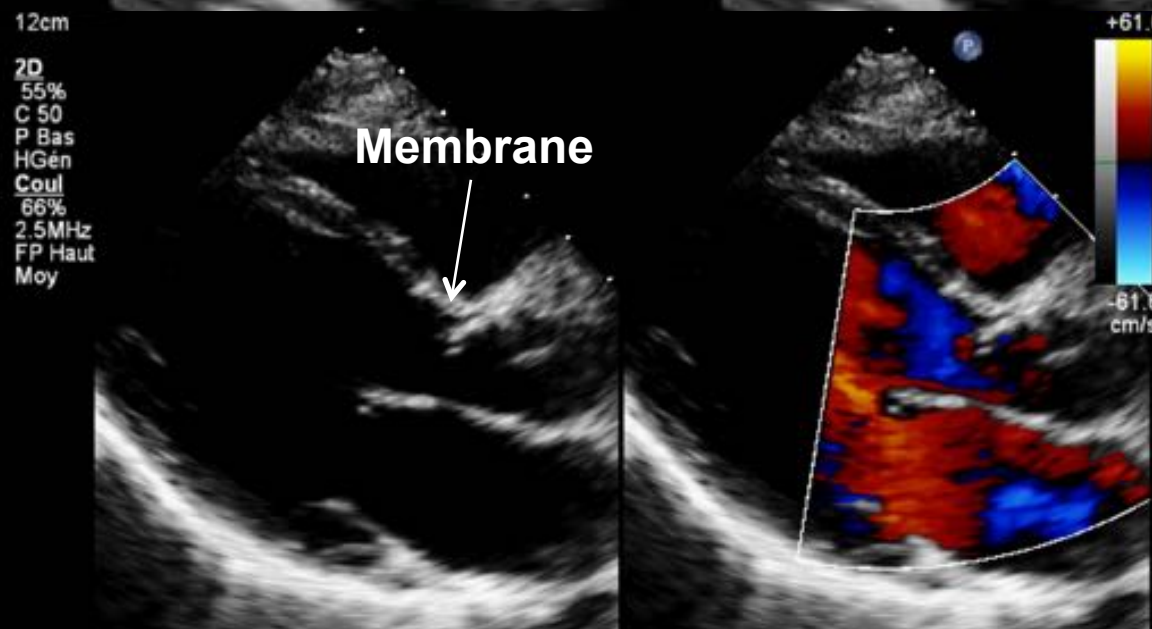
- 3 types de lésions :
 - **Membrane**
 - **Bourrelet fibreux**
 - **Tunnel fibro-musculaire** : obstacle étendu
- } obstacle localisé

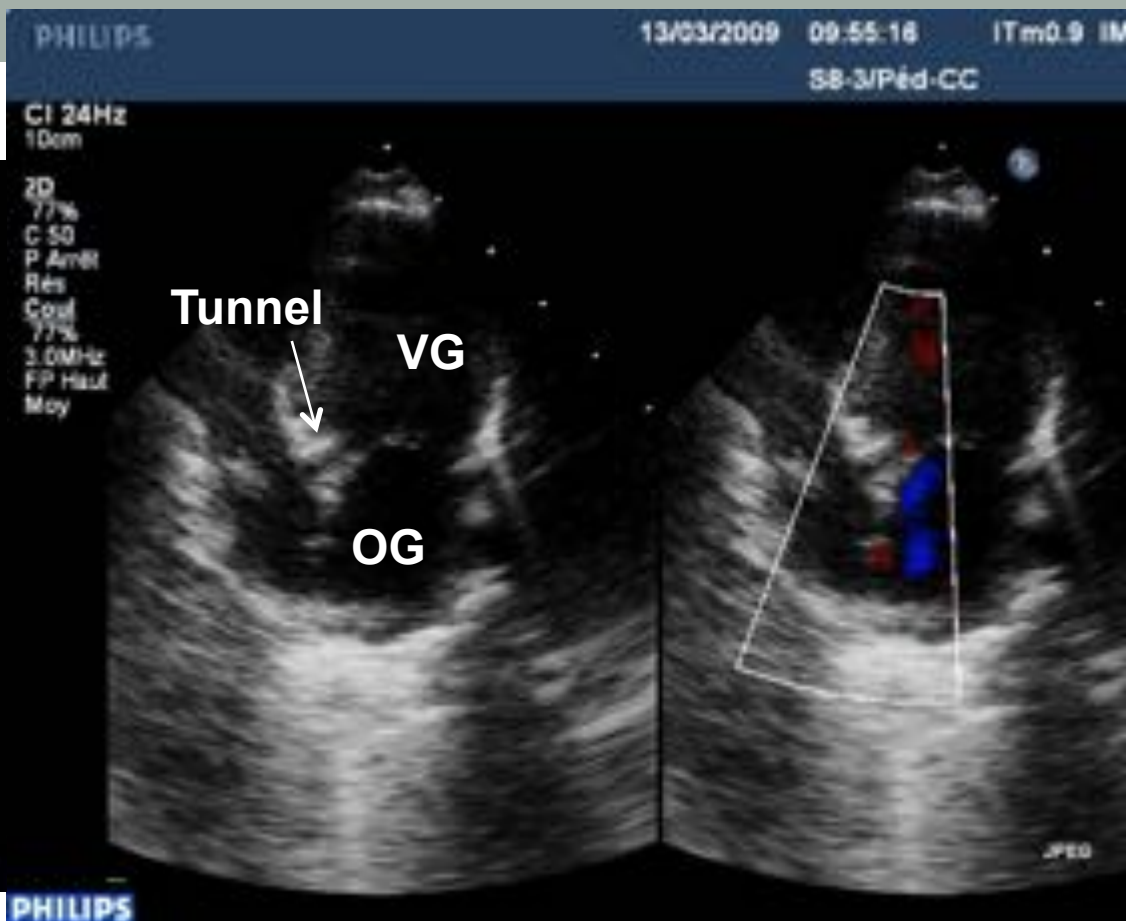
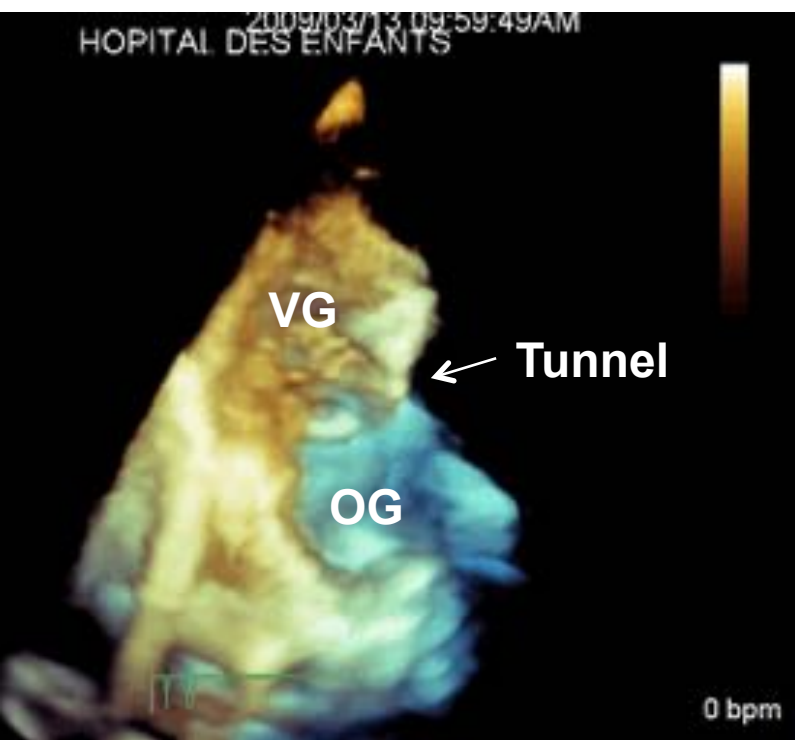
Sténose sous-valvulaire aortique : ETT

- Mécanisme
- Siège (distance avec la valve aortique) et étendue
- Sévérité : Gradient moyen
 - Sévère si gradient moy >30 ou 50 mmHg
- Retentissement : HVG, fuite aortique (**lésions de jet**, défaut de coaptation par restriction d'une des sigmoïdes)
- Recherche de lésions associées (anomalie mitrale, coarctation aortique; CIV, CAV...)



**Membrane
sous-valvulaire
aortique
5 cavités &
PSGA**





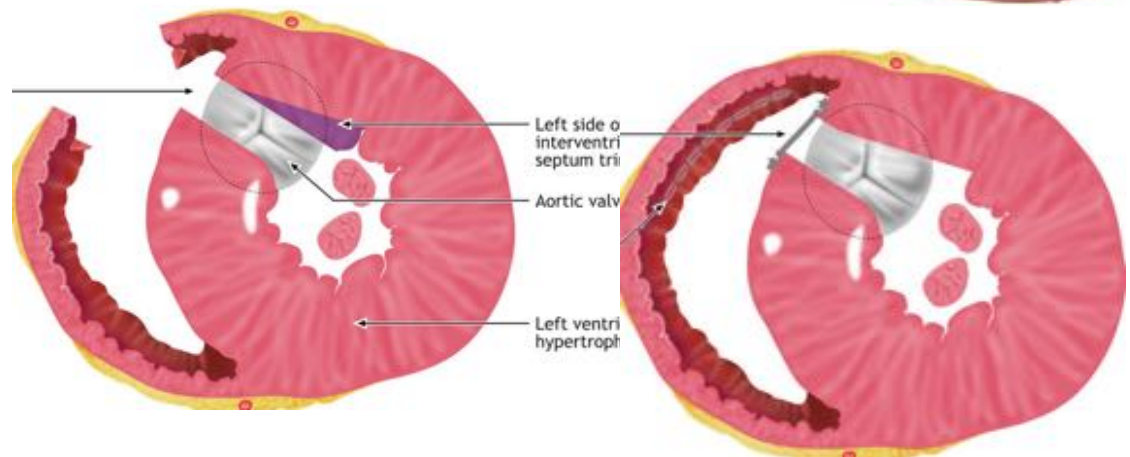
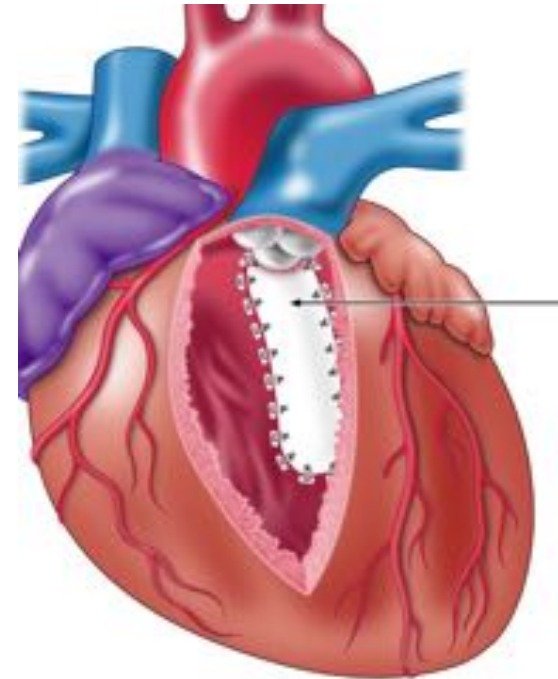
**Tunnel fibro-
musculaire**
5 cavités et 3D

Obstacle sous aortique : resultats pediatriques

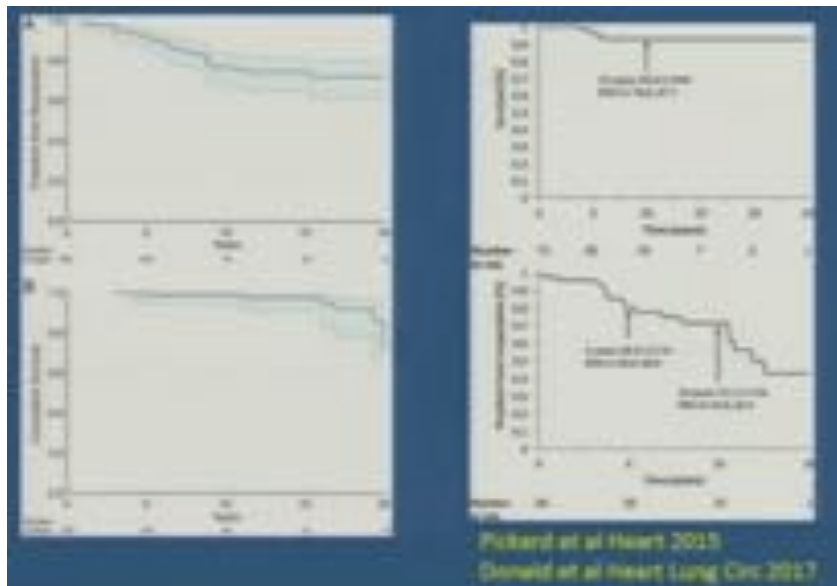
- Resection mb sous aortique +/- myectomie
- Myectomie
- Konno modifié

Complications post opératoires:

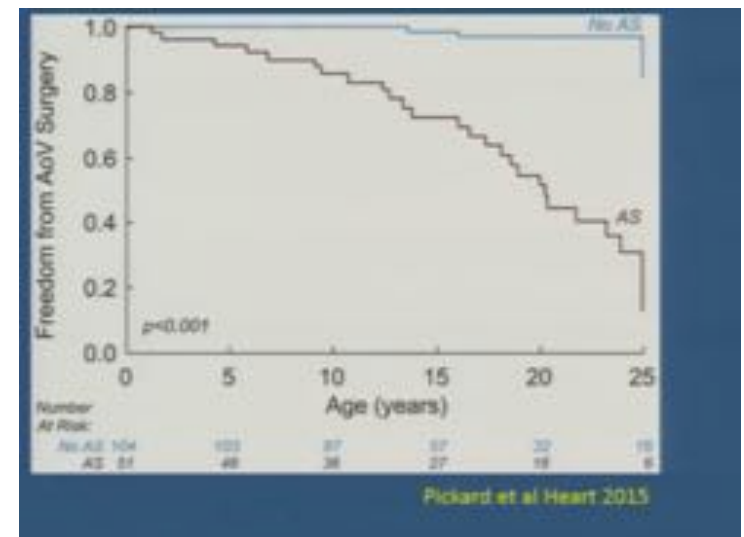
- BAV post-opératoire (10%)
- lésion coronaire
- lésion valve aortique
- CIV iatrogène



Obstacle sous aortique : resultats pediatriques



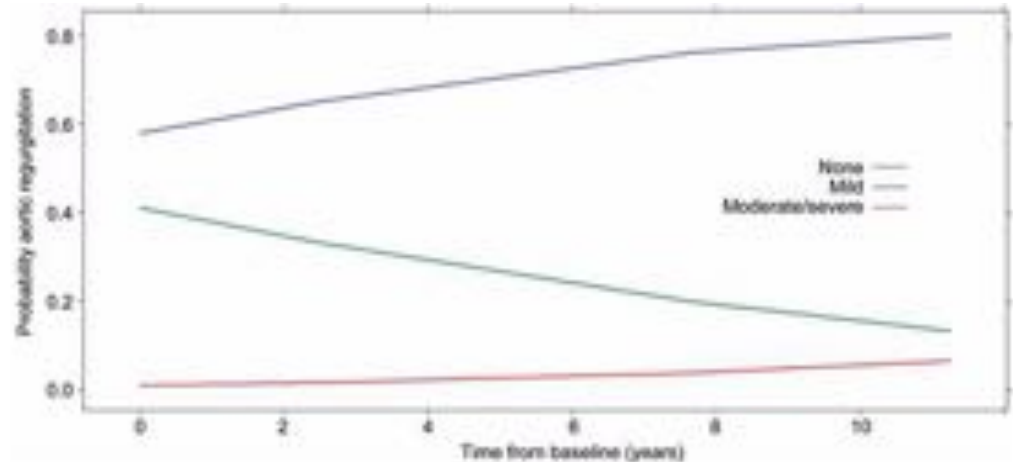
Besoin de reintervention !
Quand : Gradient > 50mmHg...



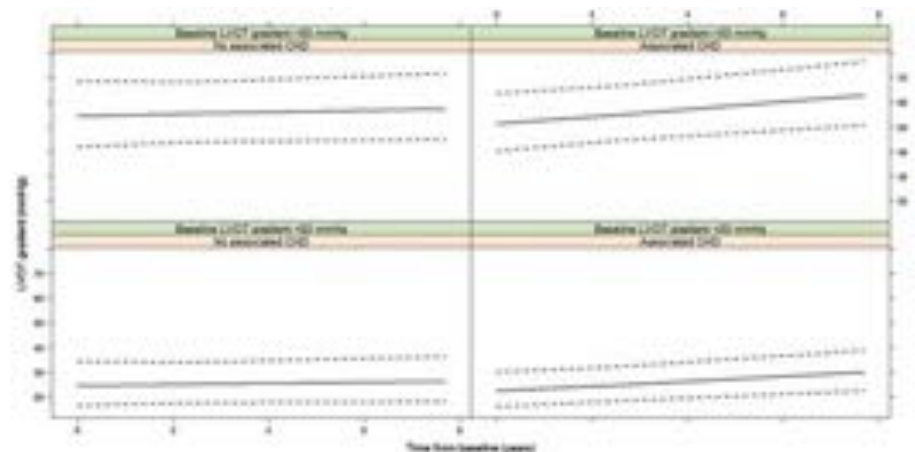
Reintervention si stenose aortique...

Obstacle sous aortique : adultes

Fuite aortique très fréquente, minime et progresse pas



Gradient évolue peu (**sauf si associé**; ex CIV)



Obstacle sous aortique : adultes

I	C-EO	2. Surgical intervention is recommended for adults with subAS, a maximum gradient 50 mm Hg or more and symptoms attributable to the subAS.
I	C-LD	3. Surgical intervention is recommended for adults with subAS and less than 50 mm Hg maximum gradient and HF or ischemic symptoms, and/or LV systolic dysfunction attributable to subAS. ^{14.2.3.3}
IIb	C-LD	4. To prevent the progression of AR, surgical intervention may be considered for asymptomatic adults with subAS and at least mild AR and a maximum gradient of 50 mm Hg or more. ^{14.2.3.4-14.2.3.6}

Attention si tunnel + complexe (konno)

Suivi : obstruction, AR, and LV function-size.
 post-operative : restenose, progressive AR,
 complications such as arrhythmias, heart block,
 and iatrogenic VS

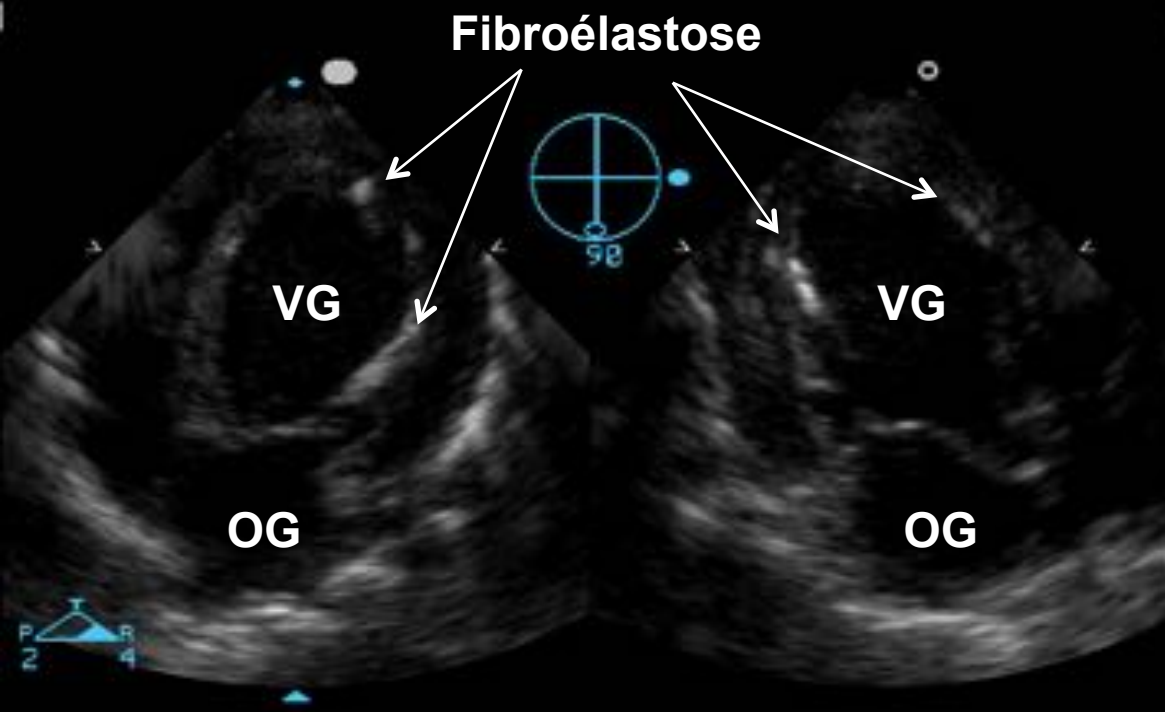
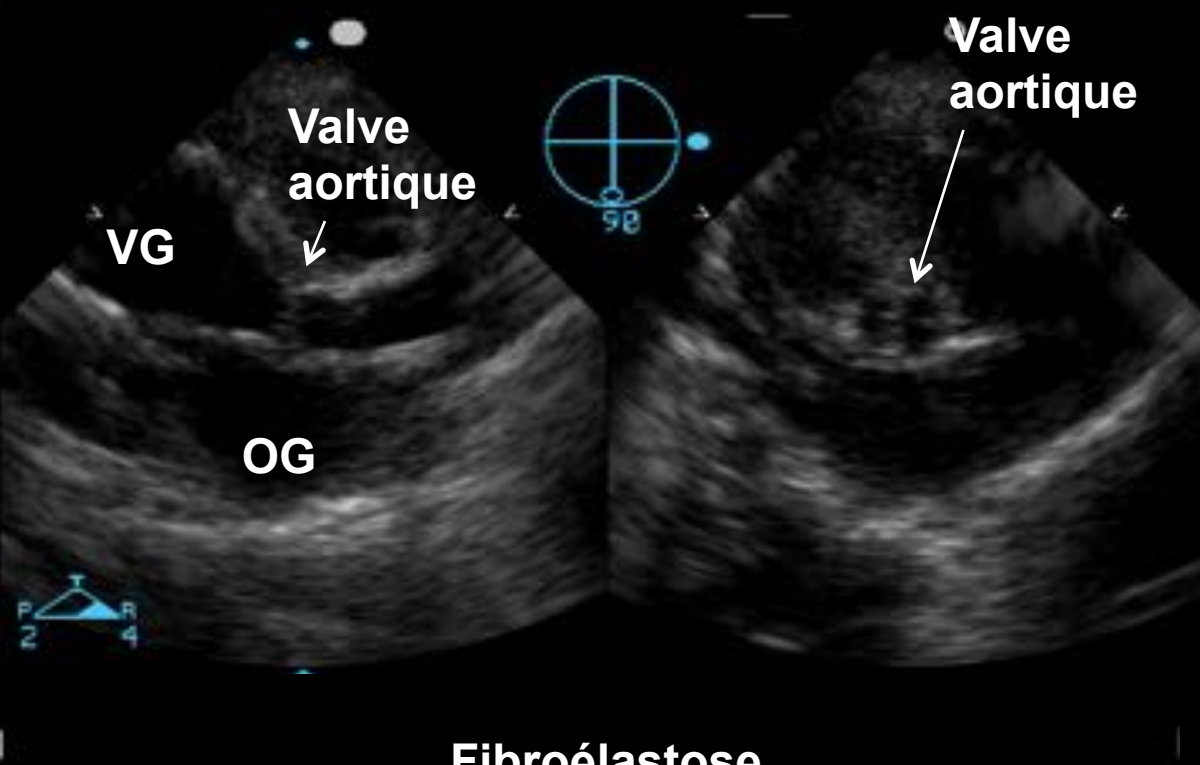
Indications	Class ^a	Level ^b
Symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient ≥ 50 mmHg ^c or severe AR should undergo surgery	I	C
Asymptomatic patients should be considered for surgery when:		
• LVEF is $< 50\%$ (gradient may be < 50 mmHg due to low flow)	IIa	C
• AR is severe and LVESD > 50 mm (or 25 mm/m ² BSA) and/or EF $< 50\%$ ^d	IIa	C
• mean Doppler gradient is ≥ 50 mmHg ^c and LVH marked	IIa	C
• mean Doppler gradient is ≥ 50 mmHg ^c and blood pressure response is abnormal on exercise testing	IIa	C
Asymptomatic patients may be considered for surgery when:		
• mean Doppler gradient is ≥ 50 mmHg ^c , LV normal, exercise testing normal, and surgical risk low	IIb	C
• progression of AR is documented and AR becomes more than mild (to prevent further progression)	IIb	C

Sténoses aortiques

- 5% des cardiopathies congénitales

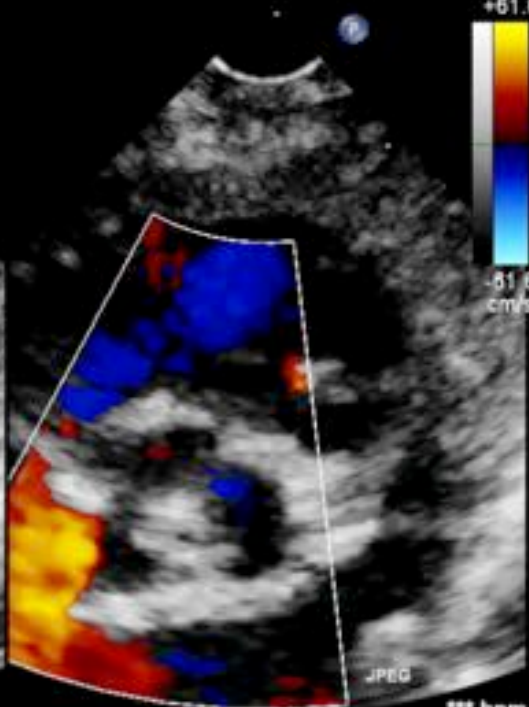
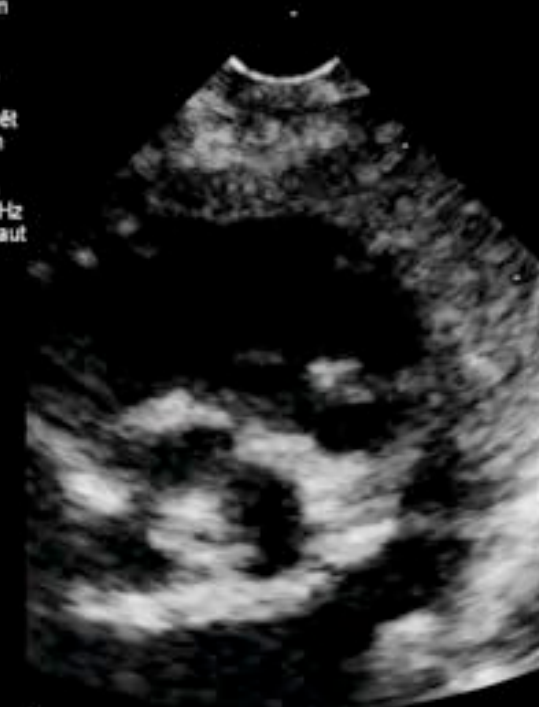
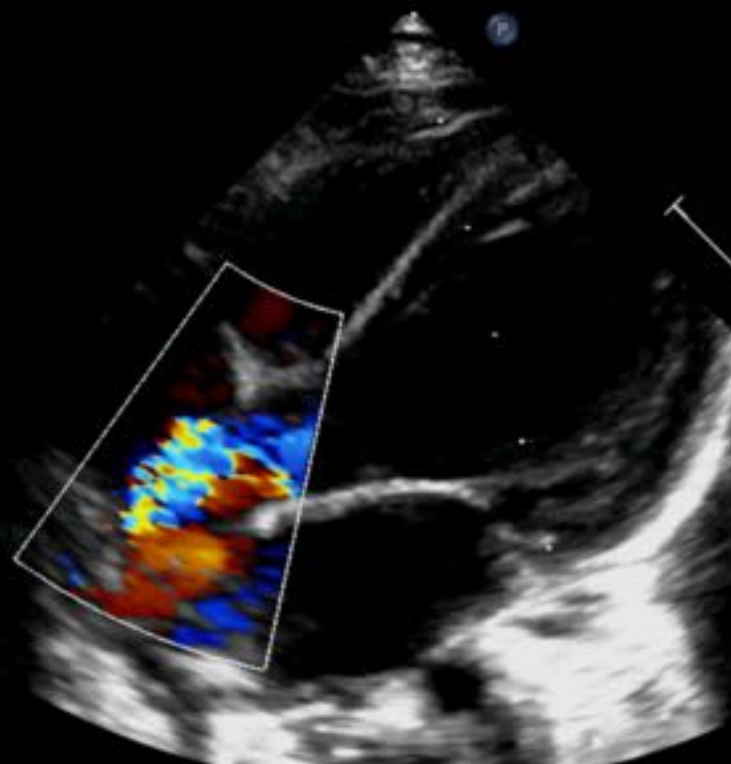
Sténose valvulaire aortique

- 2 tableaux cliniques :
 - Sténose aortique critique : Nouveau-né
 - Dysfonction VG / insuffisance cardiaque / collapsus
 - Souffle souvent modéré
 - Ducto-dépendance pour la perfusion systémique
 - Sténose aortique non critique :
 - Asymptomatique ; parfois dyspnée d'effort; (rarement syncope, mort subite, angor, douleur abdo)
 - Souffle systolique



**Sténose valvulaire
aortique critique**

**Mode biplan (PSGA,
PSPA, 4 cavités & 2
cavités)**



CI 31Hz
7.0cm

2D
67%
C 50
P Arrêt
HGen
Coul
77%
3.0MHz
FP Haut
Moy

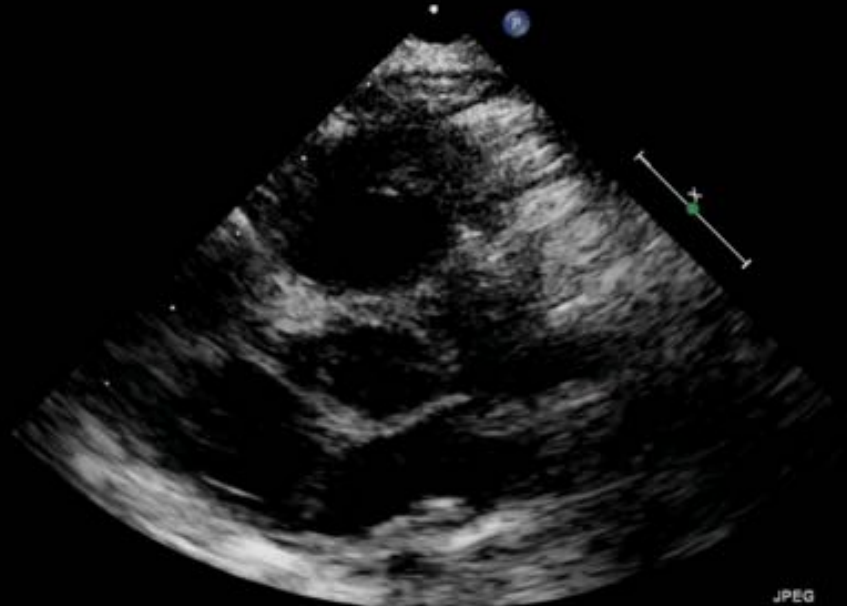
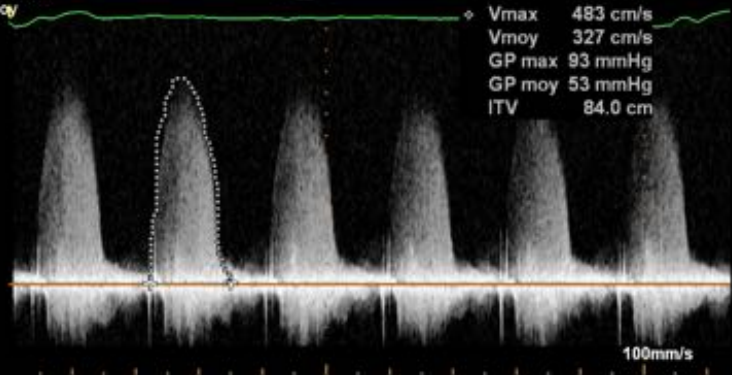


DC
45%
3.3MHz
FP 225Hz

CI 37Hz
4.0cm

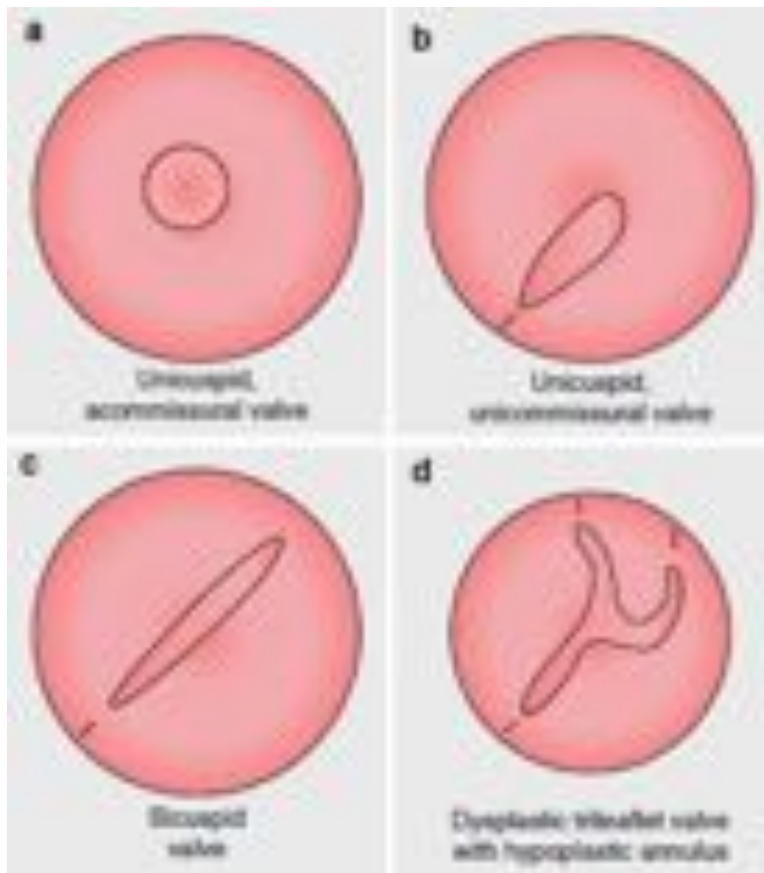
2D
62%
C 47
P Arrêt
Rés

61.6 cm/s



Sténose valvulaire aortique

- Mécanisme :



Bicuspidie aortique : Sievers

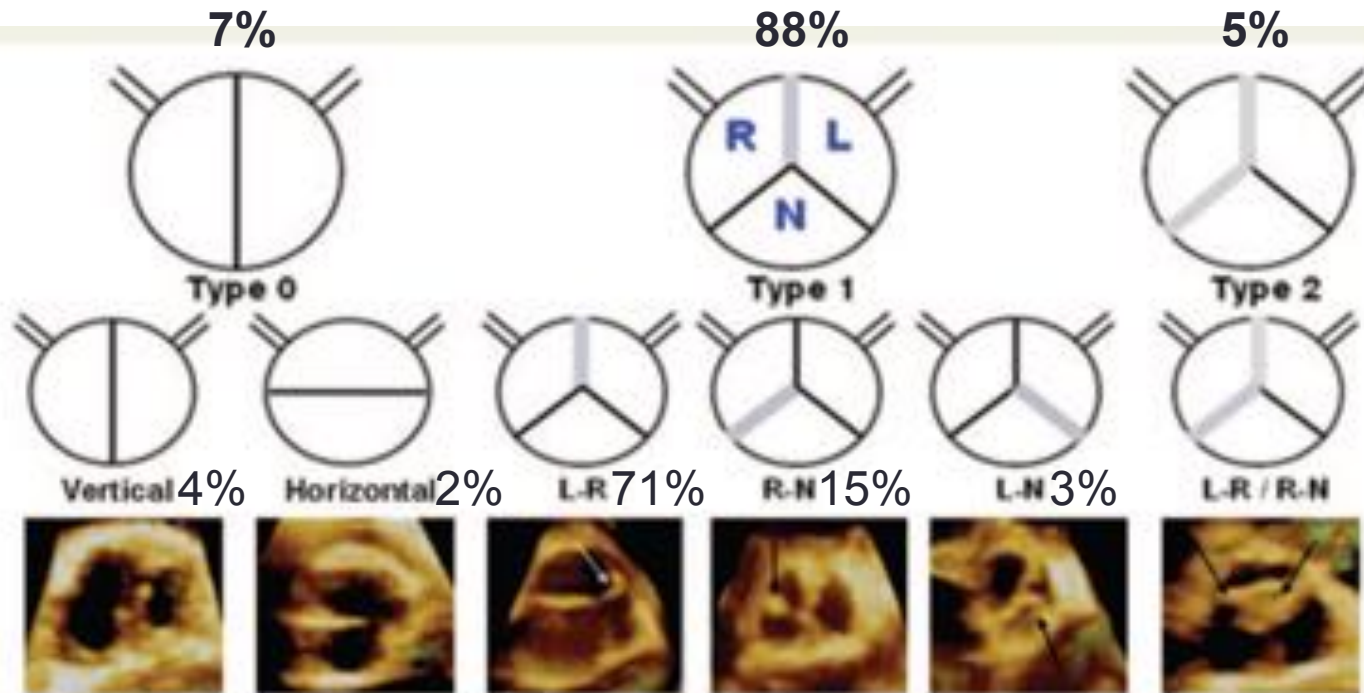
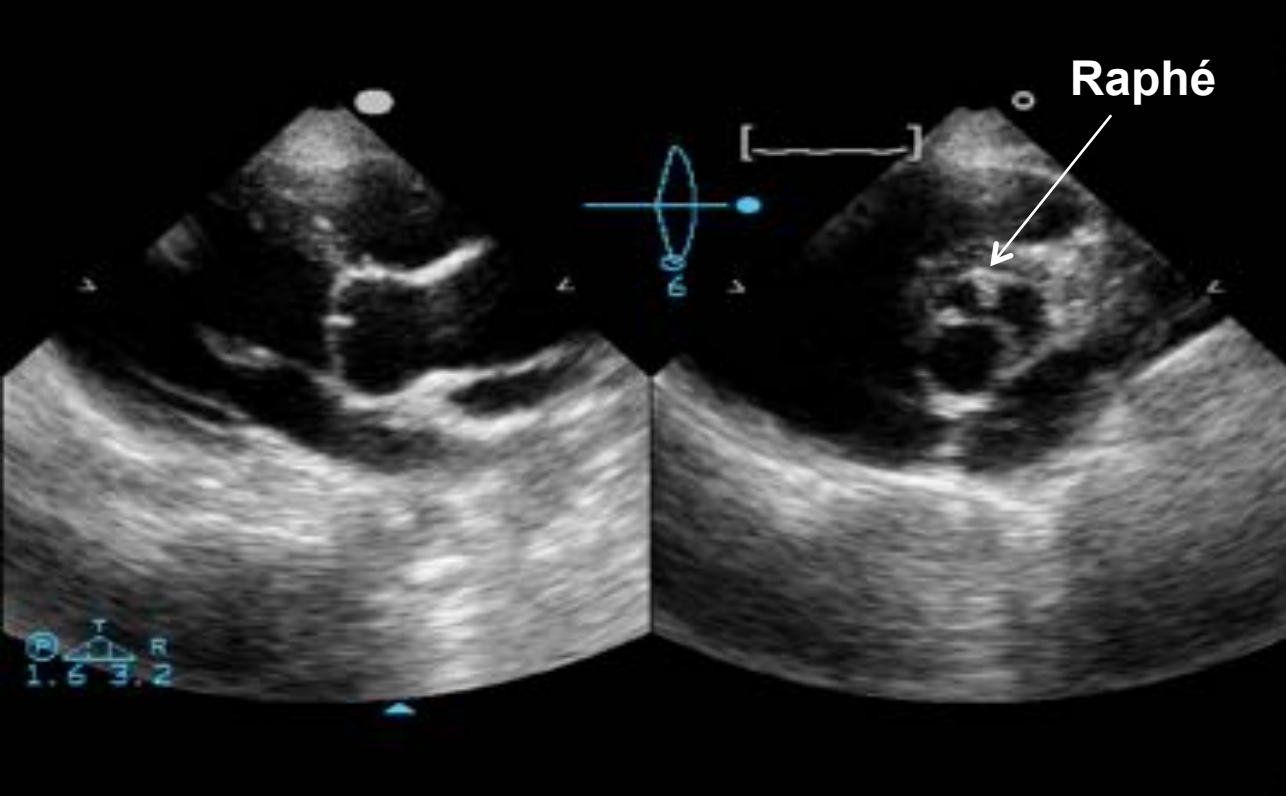


Figure 2 Correspondence between the classification of Sievers and three-dimensional echocardiographic views of BAVs. Aortic valves are seen in the short-axis section and from a left ventricular view. Grey bands represent the raphe (commissural fusion) and arrows show them. Type 0 denotes 'true' BAVs (it means with only two cusps and no raphe) and type 1 and type 2 denote 'false' BAVs (it means with three cusps and one or two raphe). Adapted from : Sievers HH, Schmidtke C. J Thorac Cardiovasc Surg. 2007 May; 133:1226-33.

Interet? → PRONOSTIQUE : Type 0 ou Type 2 = valvulopathie + précoce



VR 26Hz
7cm
Live 3D
3D 30%
3D 50dB



3D↑
PHILIPS

**Bicuspidie aortique
type 1 LR**

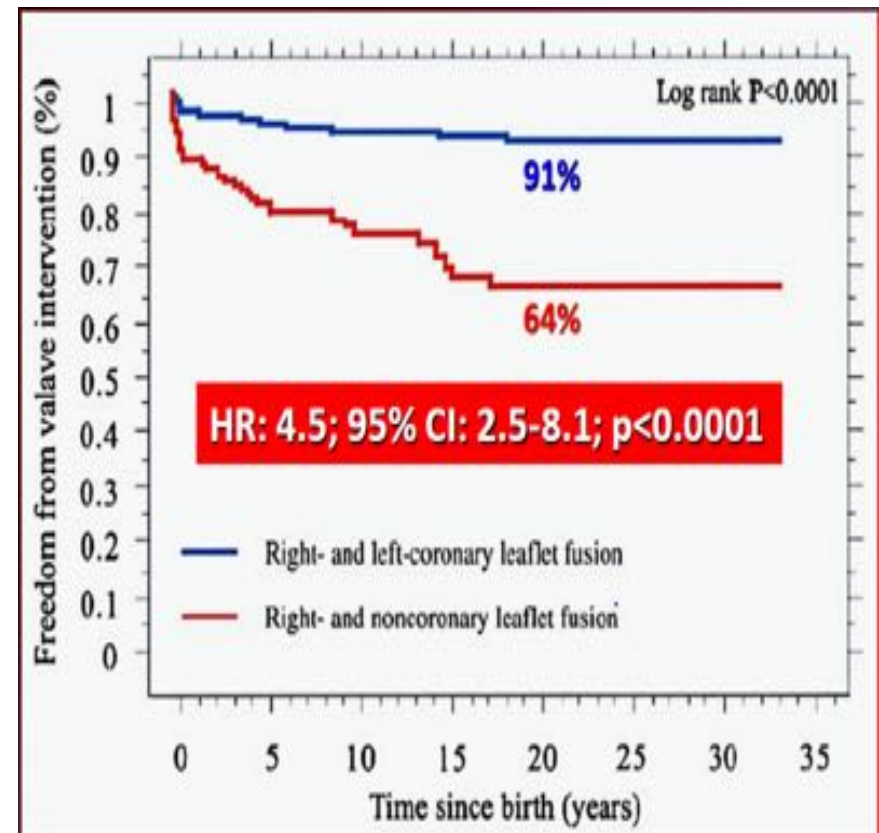
**Mode biplan (PSGA
et PSPA) & vue 3D
depuis l'aorte**

Bicuspidie aortique : classification Sievers

- Forme la plus fréquente >> 1 L – R.

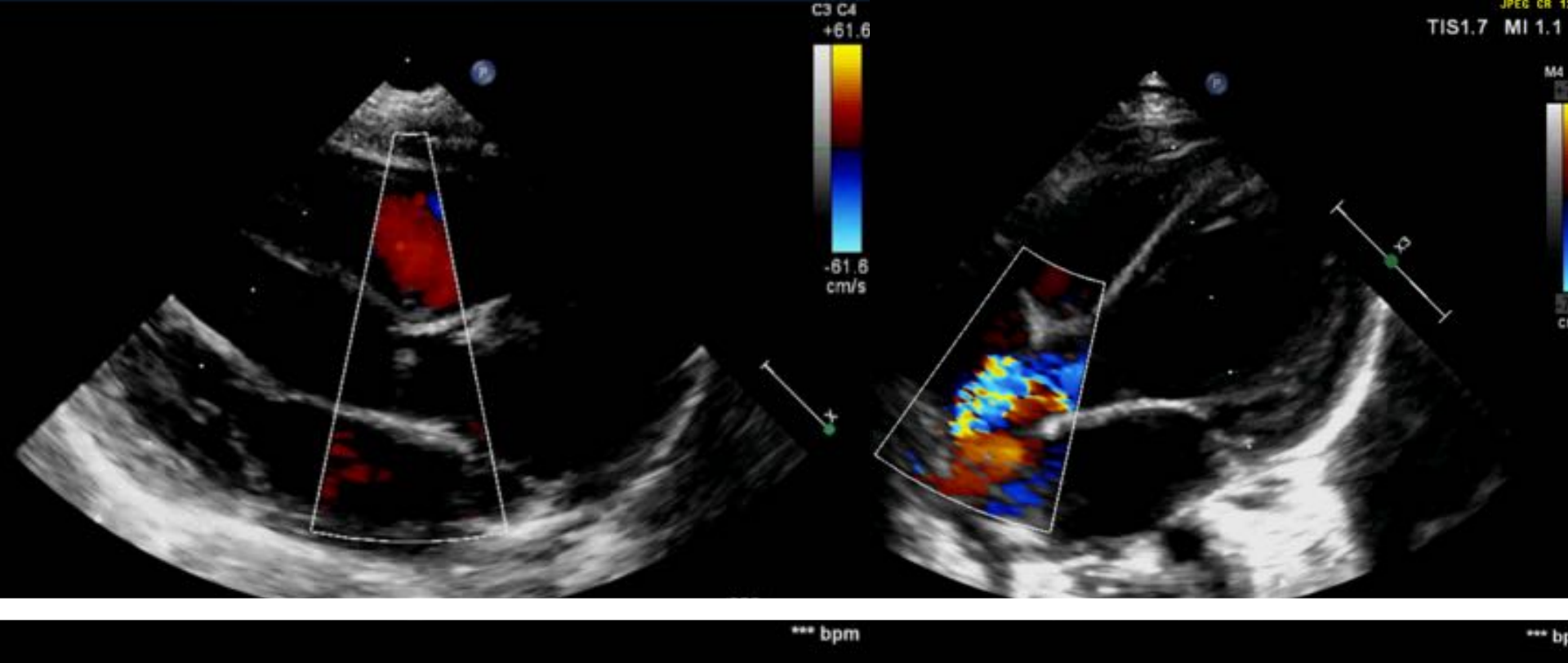
Association fréquente avec une coarctation, mais évolution plus tardive vers une valvulopathie

- Evolution plus rapide vers une valvulopathie quand fusion avec la cusp non coronaire



Sténose valvulaire aortique : ETT

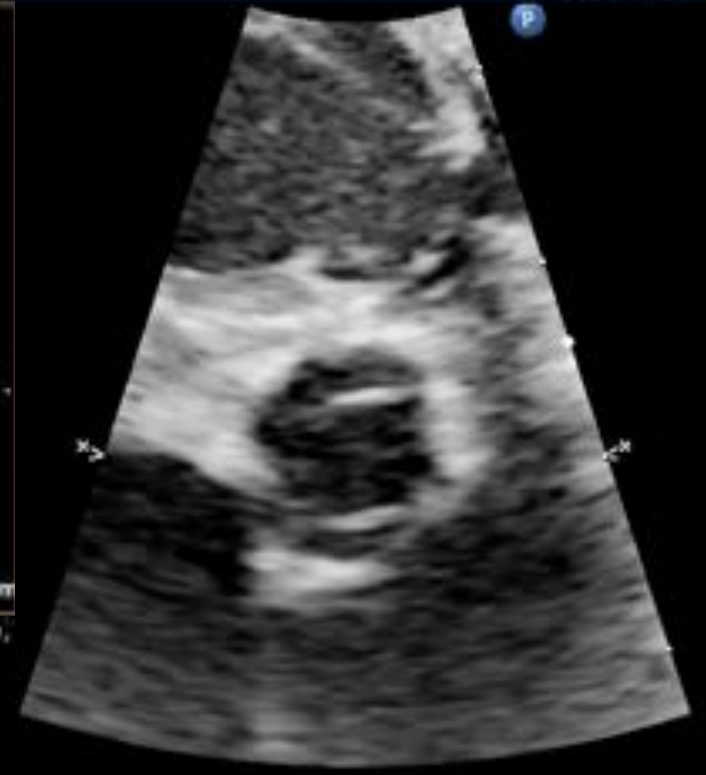
- Mécanisme : nombre sigmoïdes, dysplasie, taille de l'anneau aortique
- Sévérité : Gradient moyen (sous-estimé en cas de dysfonction VG)
 - Sévère si gradient moy >50 mmHg (**Suprasternal...**)
- Retentissement : **HVG**, ischémie, dysfonction VG
- Canal artériel
- Recherche de lésions associées (anomalie mitrale, coarctation aortique...)



Sténose valvulaire aortique
PSGA et 5 cavités

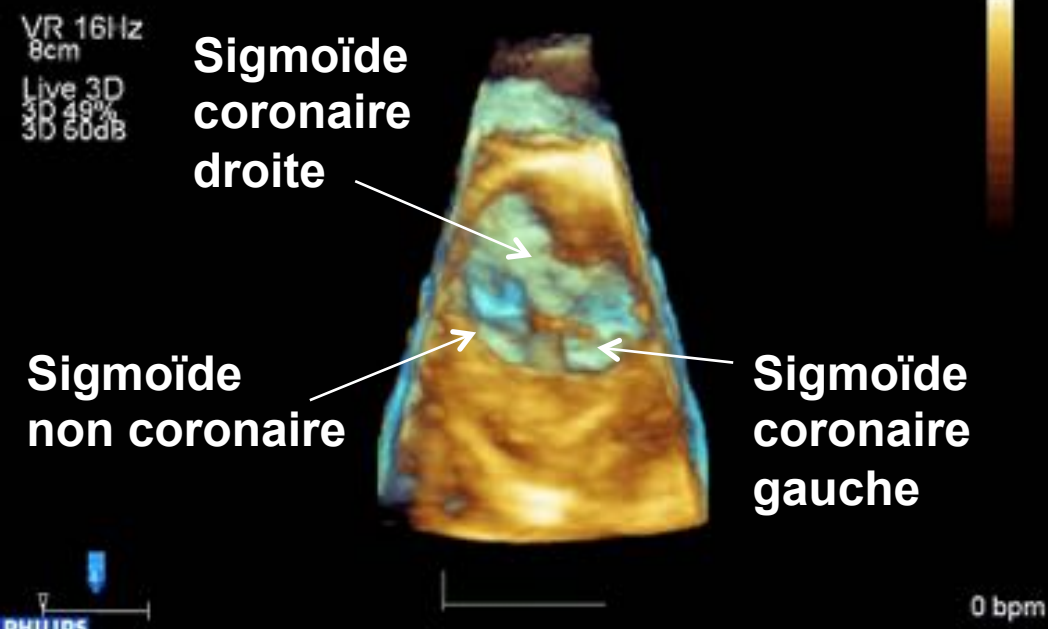
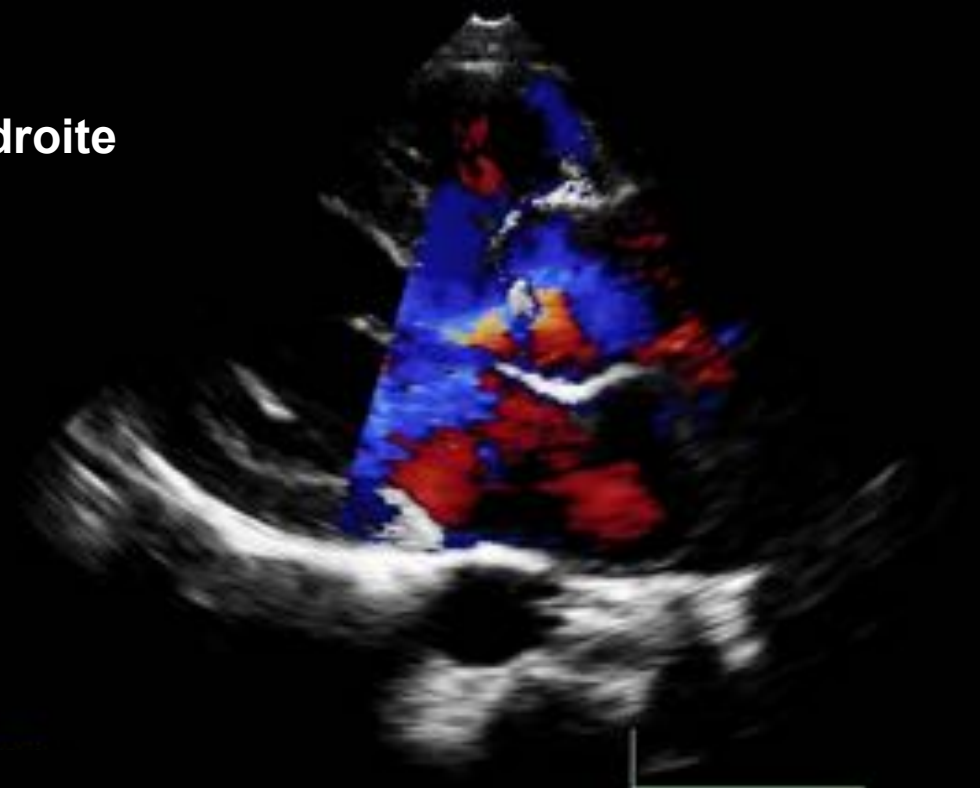
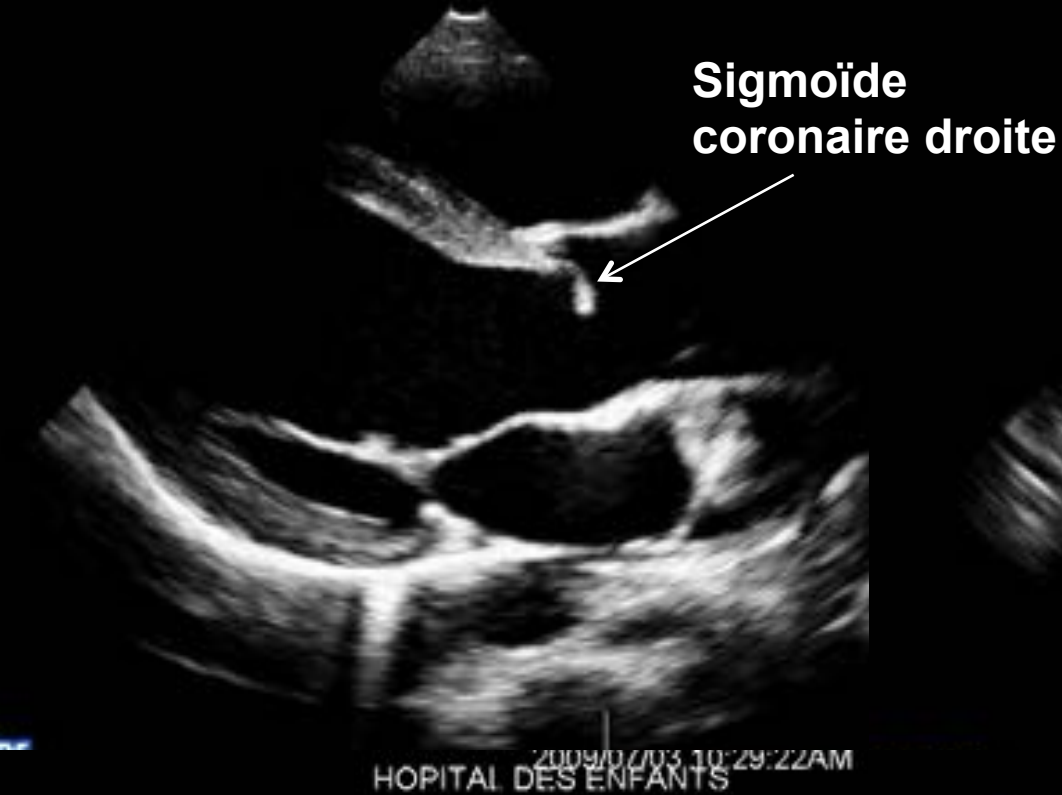


HOPITAL DES ENFANTS



**Bicuspidie aortique
type 0 horizontale**

Mode biplan (PSGA et
PSPA), PSPA et vue 3D
depuis l'aorte

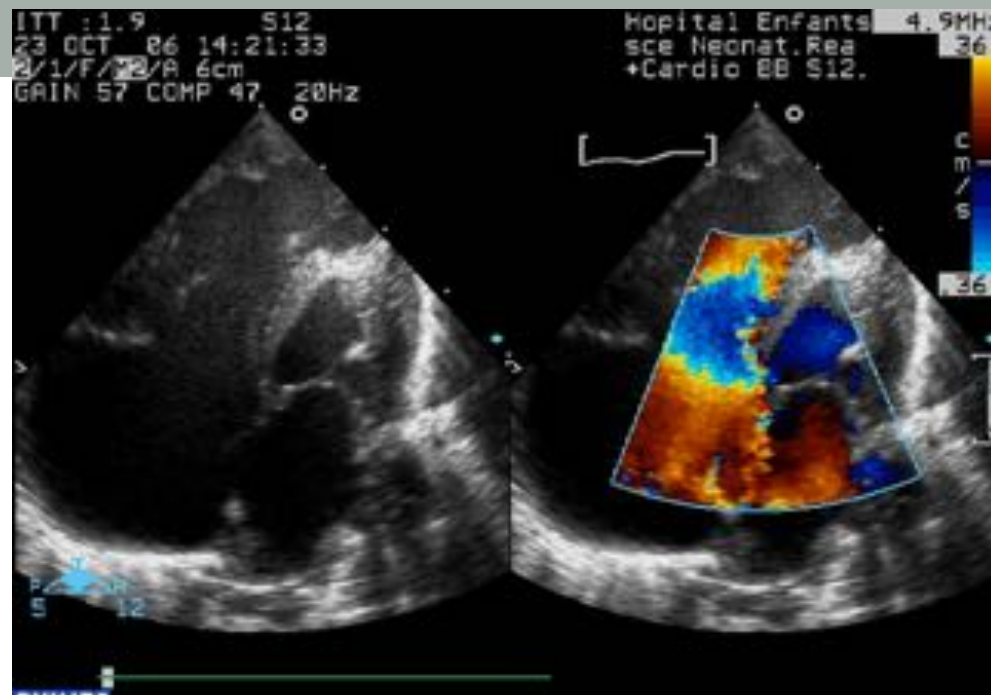


**Valve aortique dysplasique,
restriction de la sigmoïde
coronaire droite**

**PSGA & vue 3D depuis
l'aorte**

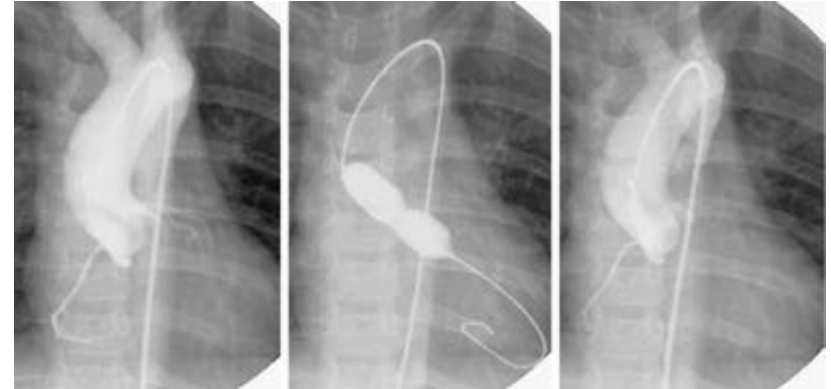
Traitement

- **Sténose valvulaire aortique critique** : traitement médical pour permettre récupération hémodynamique
 - Prostaglandine PGE1 (Prostine®) : maintient le canal artériel ouvert
 - Inotrope, ventilation mécanique
- Puis Valvuloplastie percutanée ou chirurgicale



Traitement néonatal

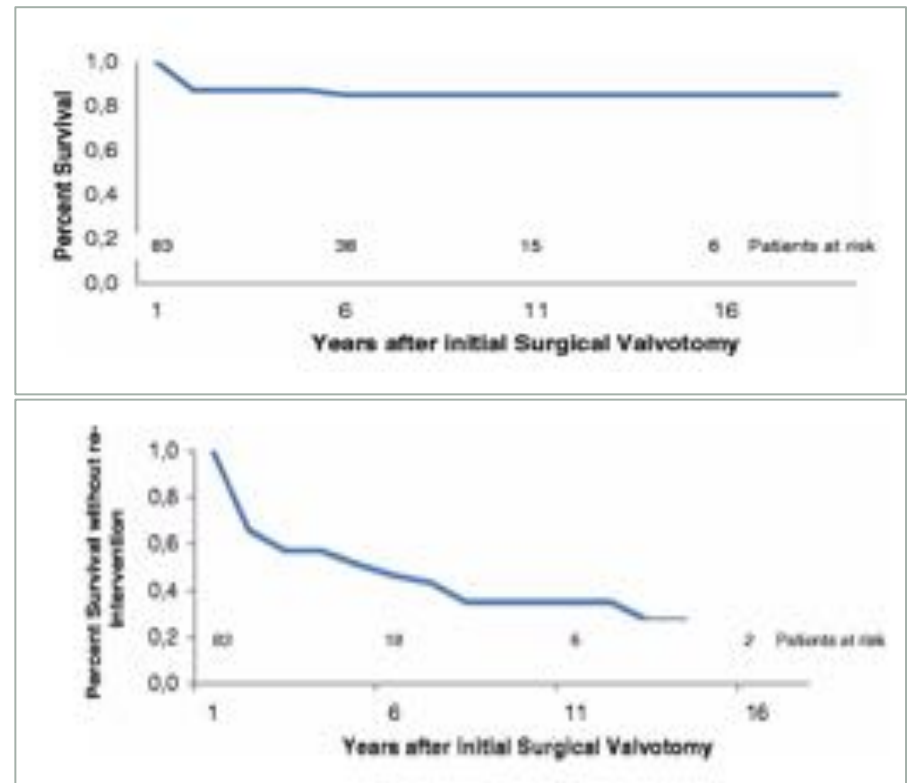
- KT vs Chir (valvulotomie)



Chir : Bons résultats en l'absence de
dysfonction VG sévère
Mais reintervention +++

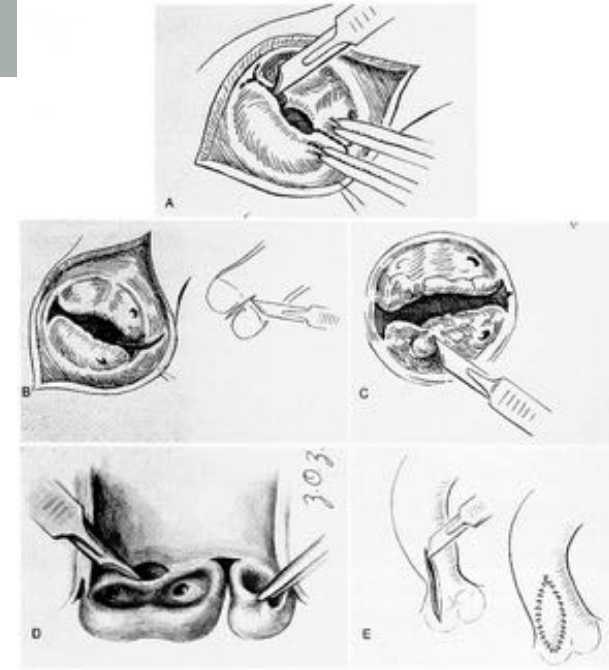
Kt si bas débit ++
Sinon à discuter (centre dpdt)

Agnoletti et al. ATS 2006
Belli JTCS 2016
Reich Heart 2004



Traitement

- **Sténose non critique :**
 - Valvuloplastie percutanée
 - Commissurotomie chirurgicale
 - Opération de Ross
 - Remplacement valvulaire aortique

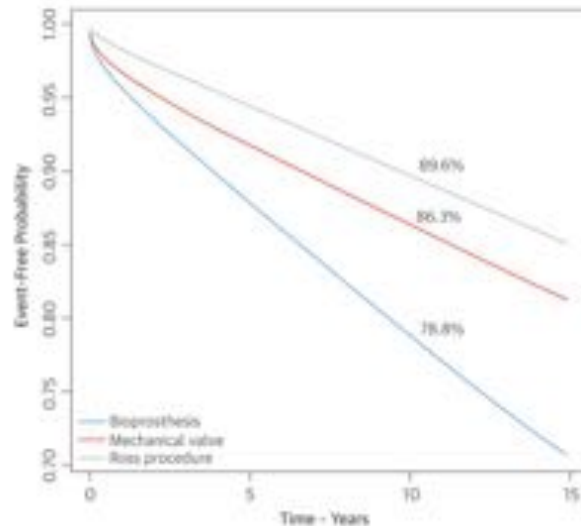


Ross

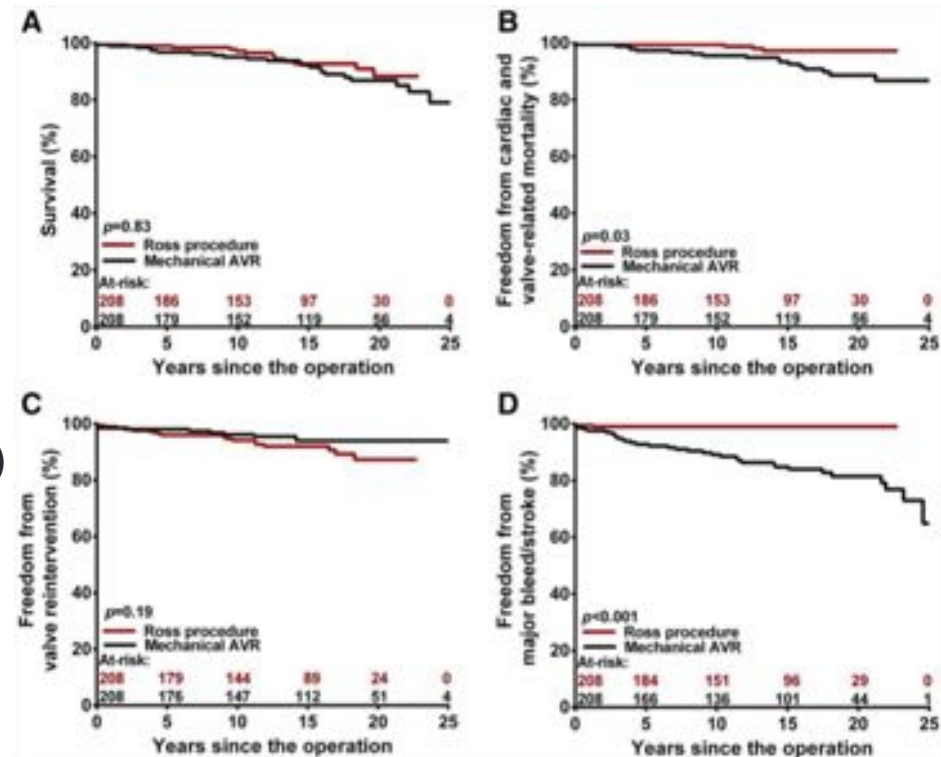
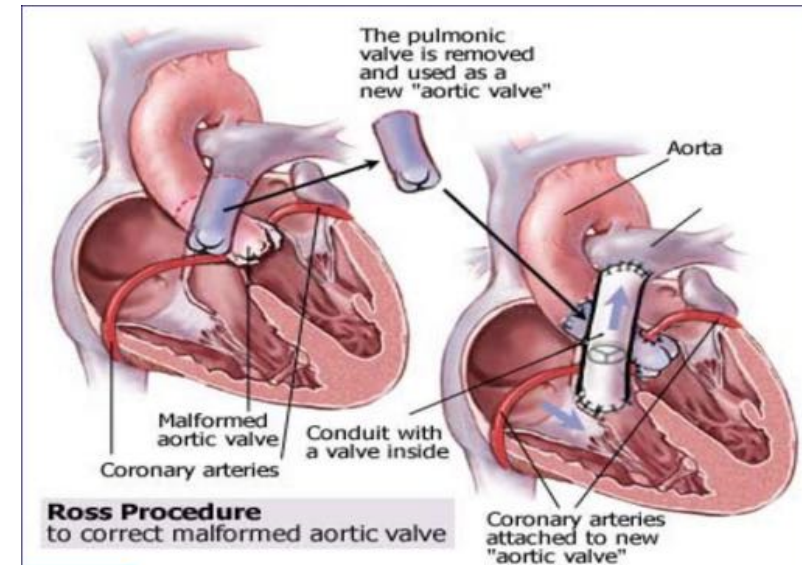
Evolue avec croissance

Pas d'anticoagulation

→ Courbe survie améliorées!



Mais : risque opératoire x3 (2.7% vs 1%)
(si centre inexperimenté)
+ Réintervention

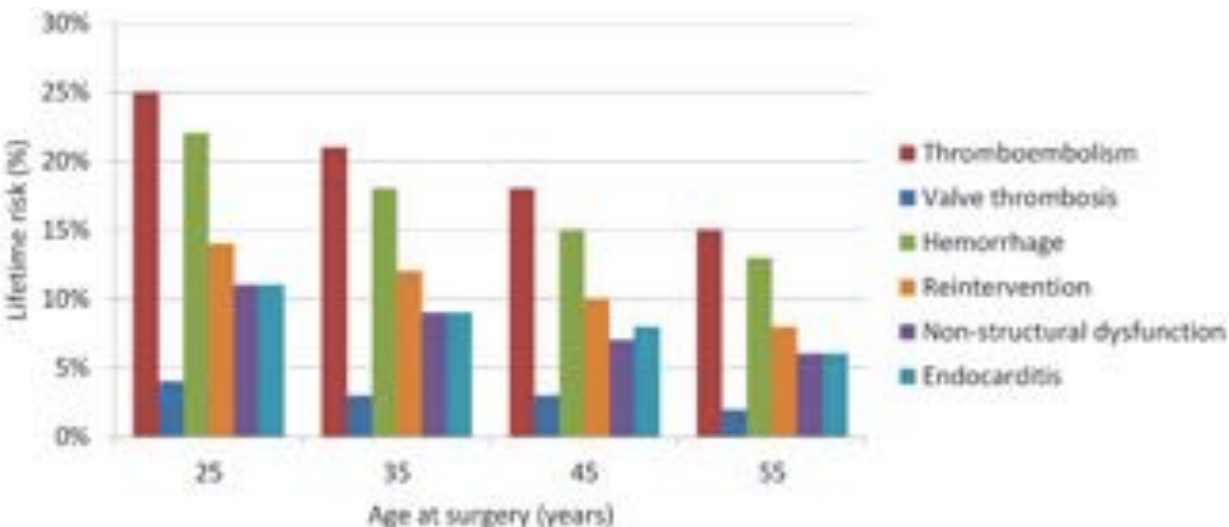


Valve mécanique adulte jeune

Meta analyse: 5728 patients (18-55 ans)

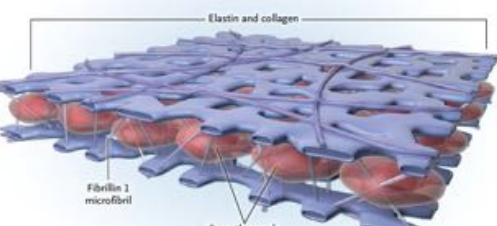
a 45ans = -15 ans d'esperance de vie

lifetime risks of thromboembolism, bleeding and reintervention of 18%, 15%, and 10%, respectively.

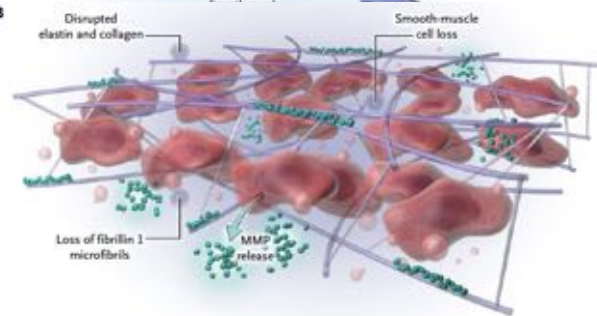


Bicuspidie : maladie du culot aortique

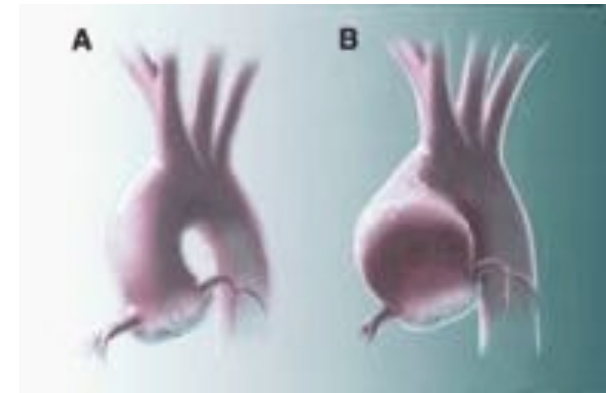
- Association quasi systématique : dilatation de l'aorte ascendante
- Physiopathologie : anomalies de la synthèse des protéines microfibrillaires de la média
- Risque >> dissection aortique



Structure normale



Bicuspidie
aortique



THE NEW ENGLAND JOURNAL of MEDICINE

REVIEW ARTICLE

Edward W. Campion, M.D., Editor

Aortic Dilatation in Patients
with Bicuspid Aortic Valve

Subodh Verma, M.D., Ph.D., and Samuel C. Siu, M.D.

A Patterns of bicuspid aortopathy

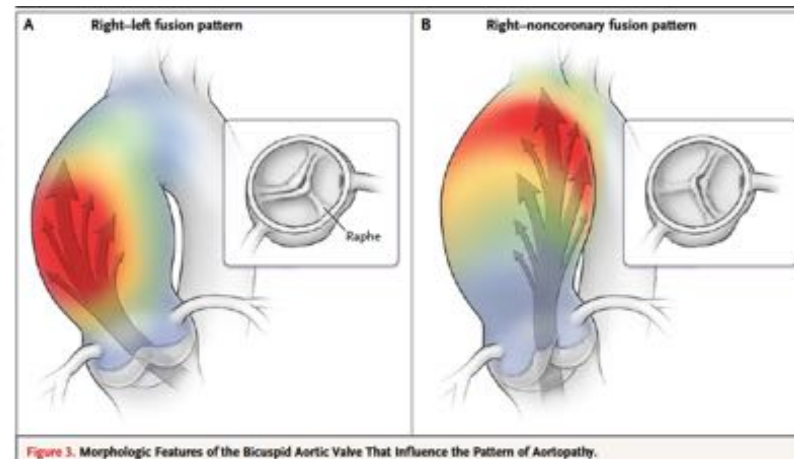
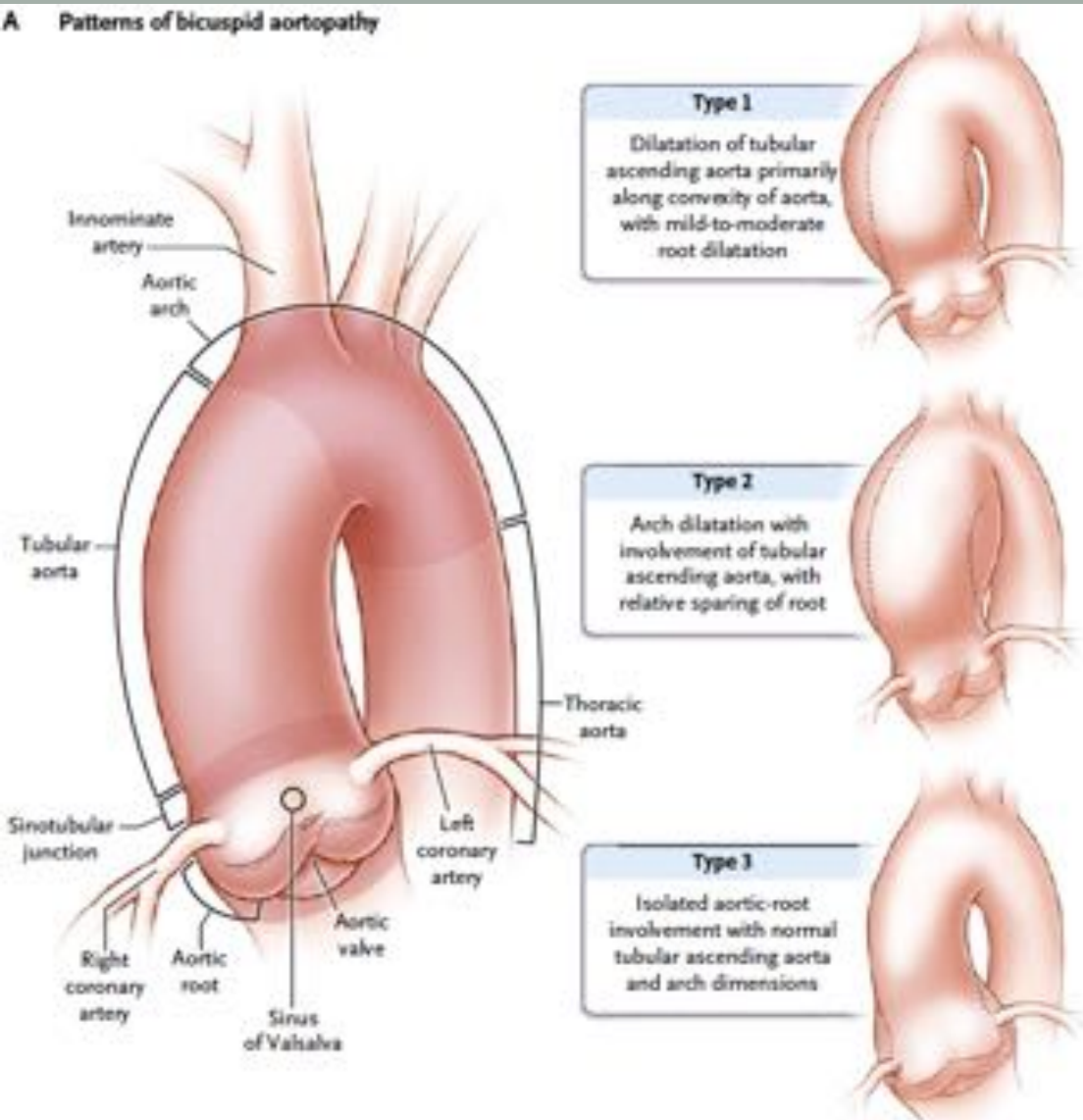


Figure 3. Morphologic Features of the Bicuspid Aortic Valve That Influence the Pattern of Aortopathy.

→ Imagerie de coupe si besoin +++

Bicuspidie : autre

Plus d'antibioprophylaxie

Suivi /an ou / 2 an

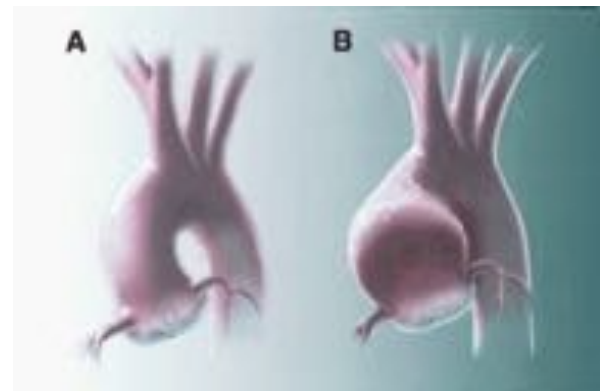
Screening Echo famille 1er degre (10% de risque vs 1% pop° general) (gene Notch1, Acta2)

IIa	B-NR	2. It is reasonable to screen first-degree relatives of patients with bicuspid aortic valve or unicuspid aortic valve with echocardiography for valve disease and aortopathy. ^{S4.2.4-2-S4.2.4-4}
-----	------	--

Grossesse: CI si >50mm

Traitement : aortopathie

Selon les recommandation ESC des valvulopathies



Baumgartner et al. EHJ 2017

Pas de donnée mais BB souvent prescrits

FdR =histoire familiale de dissection, l'ao sévères, désir de grossesse, HTA, >3mm/an

B. Aortic root or tubular ascending aortic aneurysm ^d (irrespective of the severity of aortic regurgitation)		
Aortic valve repair, using the reimplantation or remodeling with aortic annuloplasty technique, is recommended in young patients with aortic root dilation and tricuspid aortic valves, when performed by experienced surgeons.	I	C
Surgery is indicated in patients with Marfan syndrome who have aortic root disease with a maximal ascending aortic diameter ≥ 50 mm.	I	C
Surgery should be considered in patients who have aortic root disease with maximal ascending aortic diameter: ≥ 45 mm in the presence of Marfan syndrome and additional risk factors^e or patients with a TGFBR1 or TGFBR2 mutation (including Loeys-Dietz syndrome).^f ≥ 50 mm in the presence of a bicuspid valve with additional risk factors^e or coarctation. ≥ 55 mm for all other patients.	IIa	C
	IIa	C
	IIa	C
	IIa	C
When surgery is primarily indicated for the aortic valve, replacement of the aortic root or tubular ascending aorta should be considered when ≥ 45 mm, particularly in the presence of a bicuspid valve. ^g	IIa	C

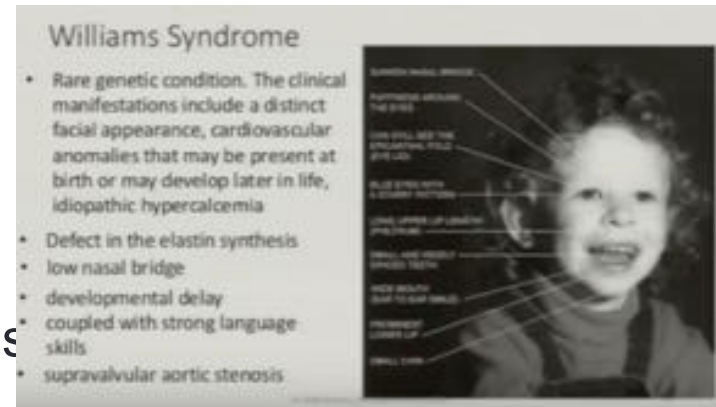
Sténose supra-valvulaire aortique

- 2 types de lésions :
 - **Sténose localisée à la jonction sino-tubulaire**
 - **Hypoplasie diffuse de l'aorte ascendante**
- 50% = Syndrome de Williams-Beuren (déficit synthèse élastine) : progresse++

Lésions associées fréquentes :

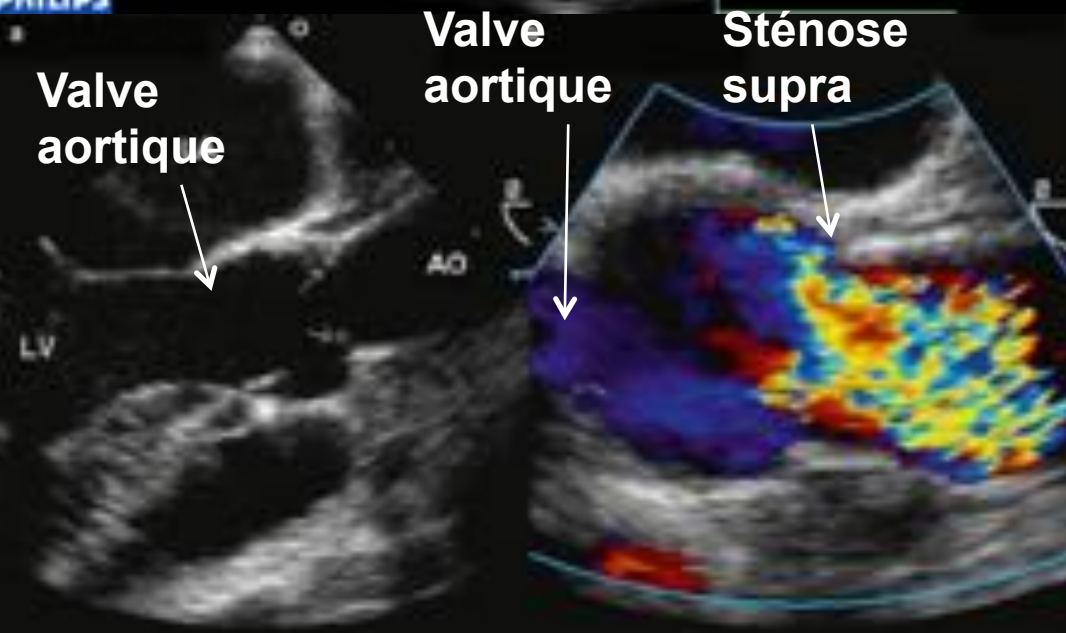
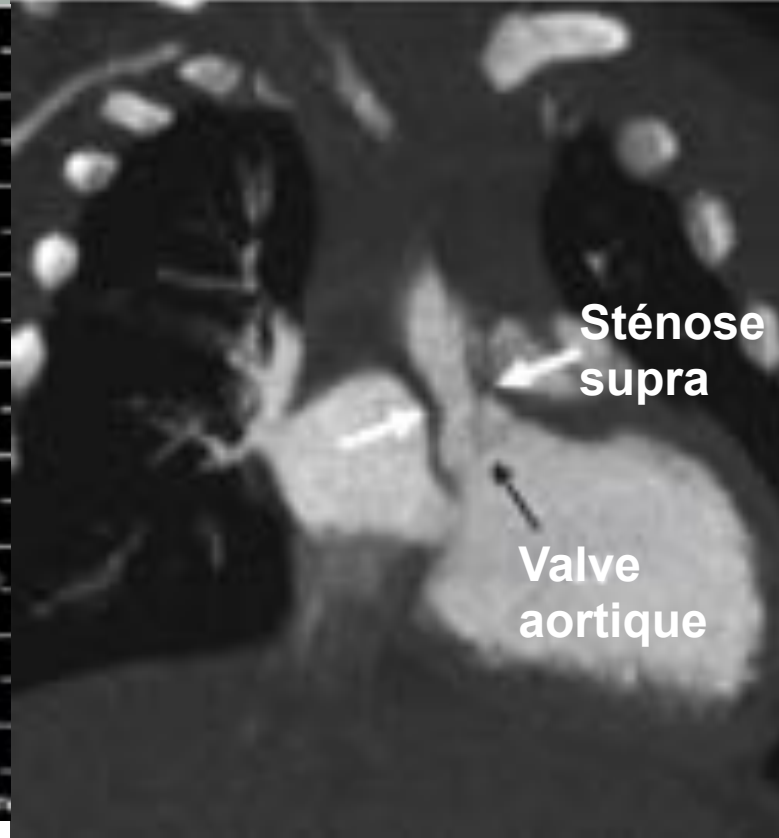
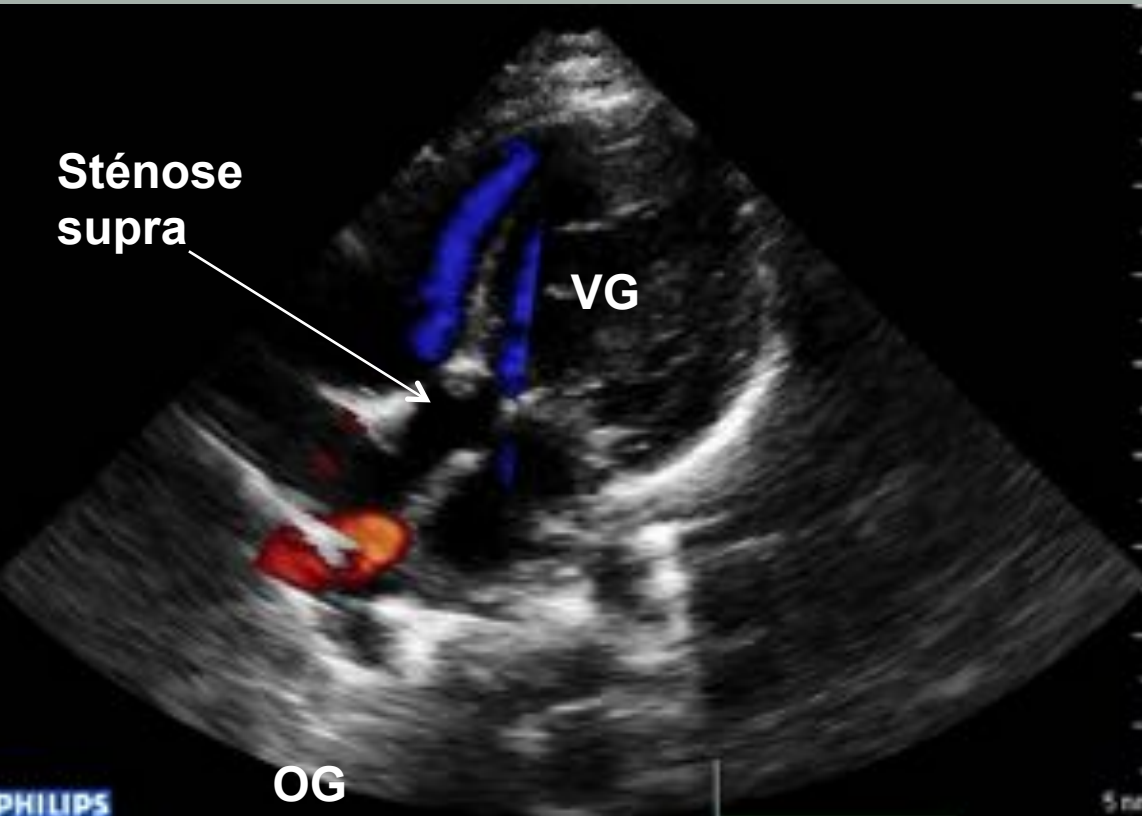
- Bicuspédie aortique
- Sténose des ostia coronaires
- Sténose des artères pulmonaires
- Sténose aorte abdominale, artères rénales

Evolutif +++



Sténose supra-valvulaire aortique : ETT

- Mécanisme
- Siège (distance avec les ostia coronaires) et étendue
- Sévérité : Gradient moyen
 - Sévère si gradient moy >50 mmHg
- Retentissement : HVG
- Recherche de lésions associées



**Sténose supra-
valvulaire aortique**
**5 cavités, PSGA &
coupe TDM**

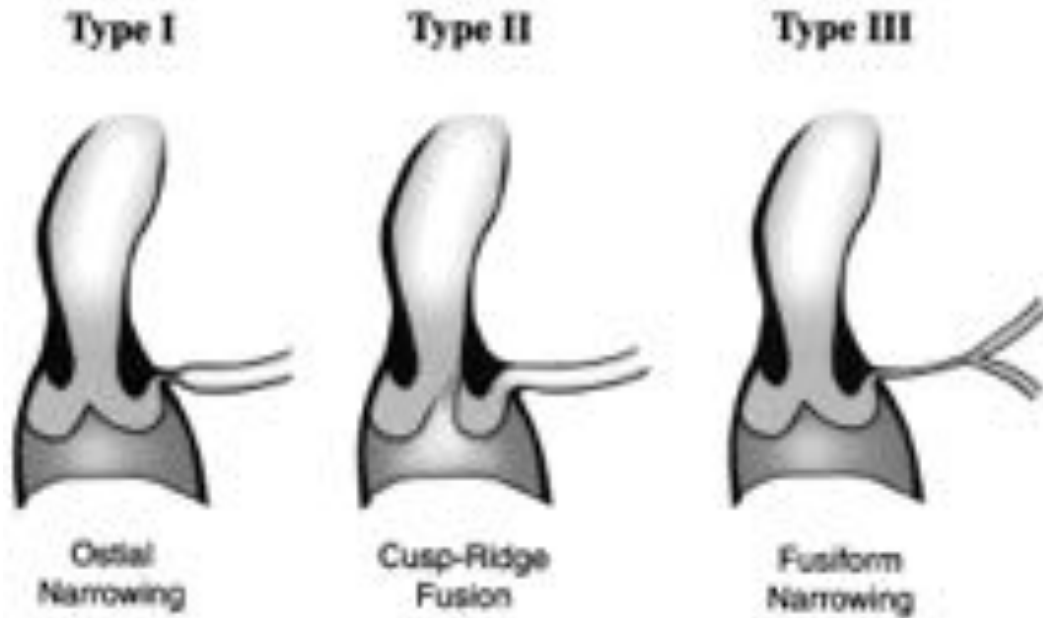
Sténose Supravalvulaire Aortique

Pathophysiologie

- Perfusion myocardique : (pas modifié dans les autres obstructions du cœur gauche)
(perfusion pendant la systole survient à une pression beaucoup plus élevée, obstruction à la jonction de sinotubulaire peut limiter la perfusion myocardique durant la diastole)

Attention en post op!

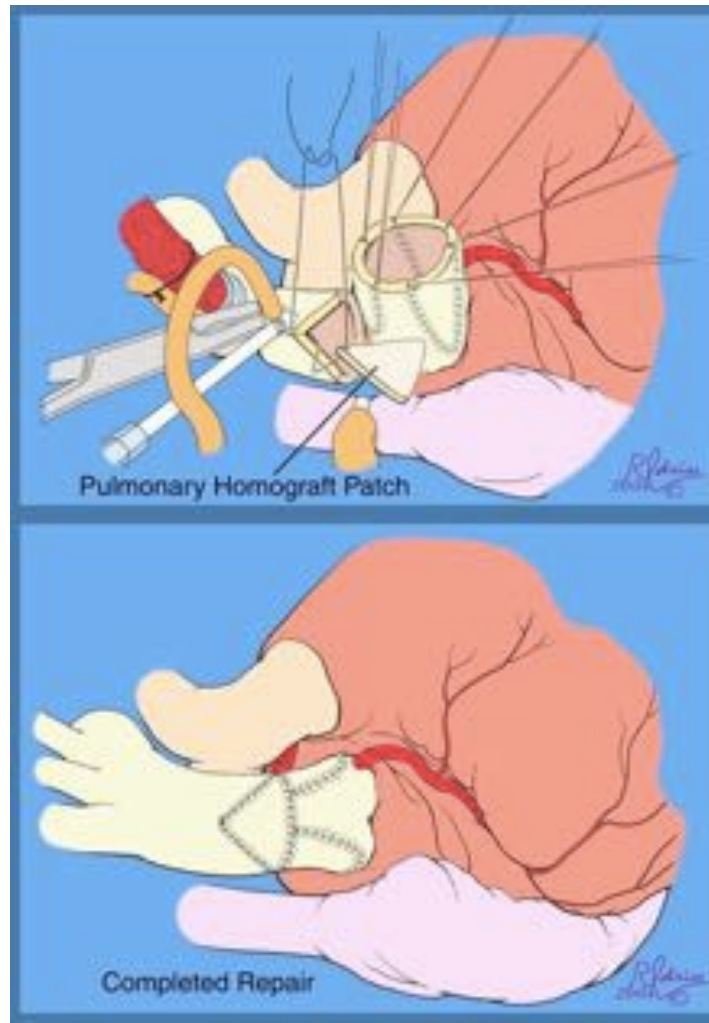
SSV aortique et sténoses coronaires



Thistlethwaite PA. JTCS 2000

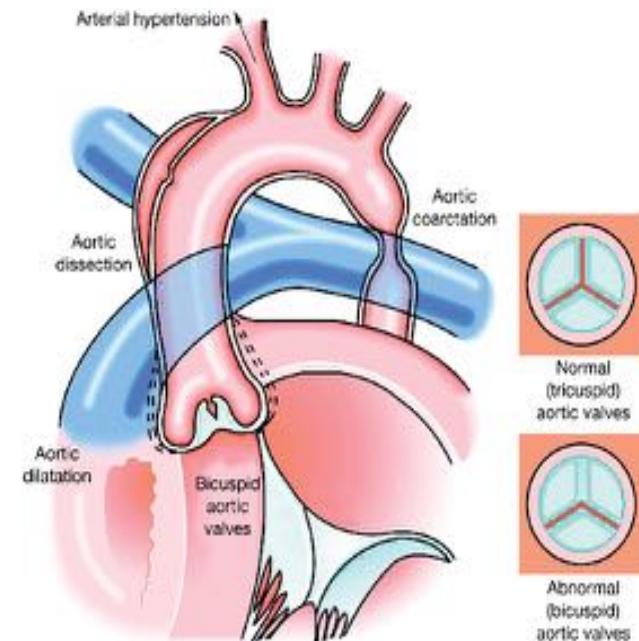
Stenose supravallulaire aortique

Intervention de Brom : reconstruction jonction sinotubulaire



COARCTATION AORTIQUE

- Sténose de l'isthme aortique
 - 5 % des cardiopathies congénitales
 - Garçons/ filles : 2-3/1
 - Parfois syndromique : Turner++,
Williams-Beuren
-
- Lésions associées : 2/3
 - hypoplasie de l'arche
 - **bicuspidie aortique (50%)**
 - anomalies mitrales, syndrome de Shone
 - CIV, CIA
 - VCSG



Arche aortique: anatomie

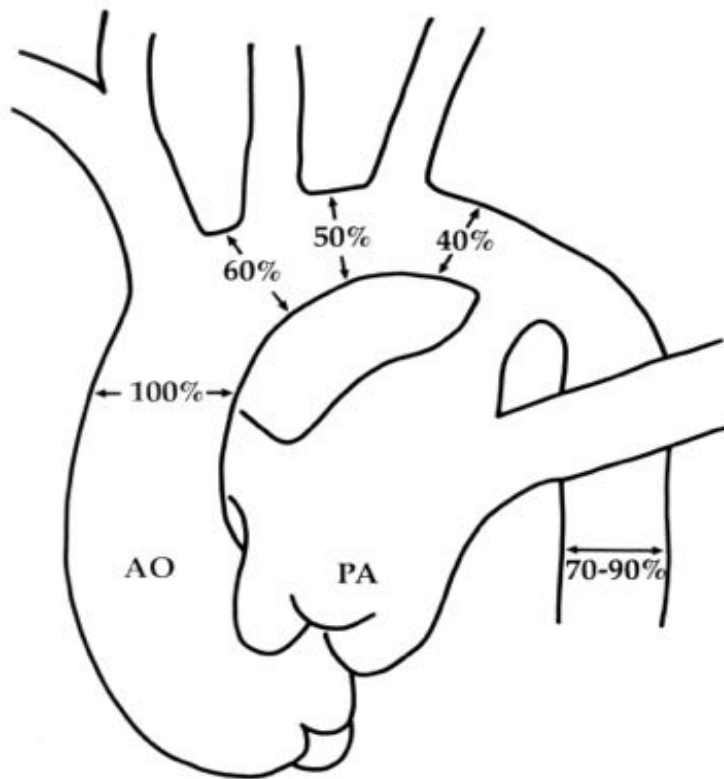


Fig 1. Minimal relative diameters of the normal thoracic aorta (AO) in infancy. (PA = pulmonary artery.) (Adapted from Moulaert AJ, Bruins CC, Oppenheimer-Dekker A. Anomalies of the aortic arch and ventricular septal defects. *Circulation* 1976;53:1101-5, by permission.)

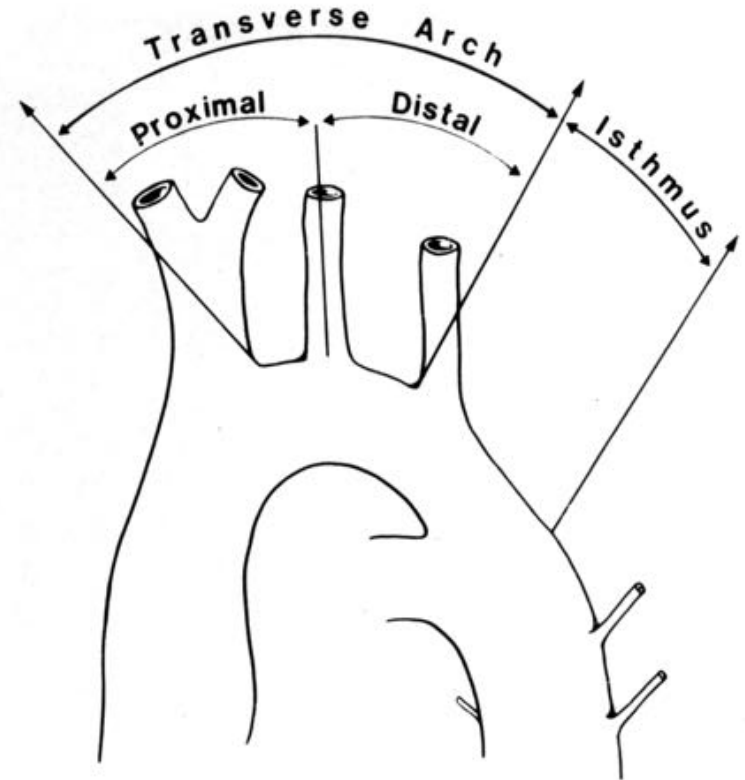
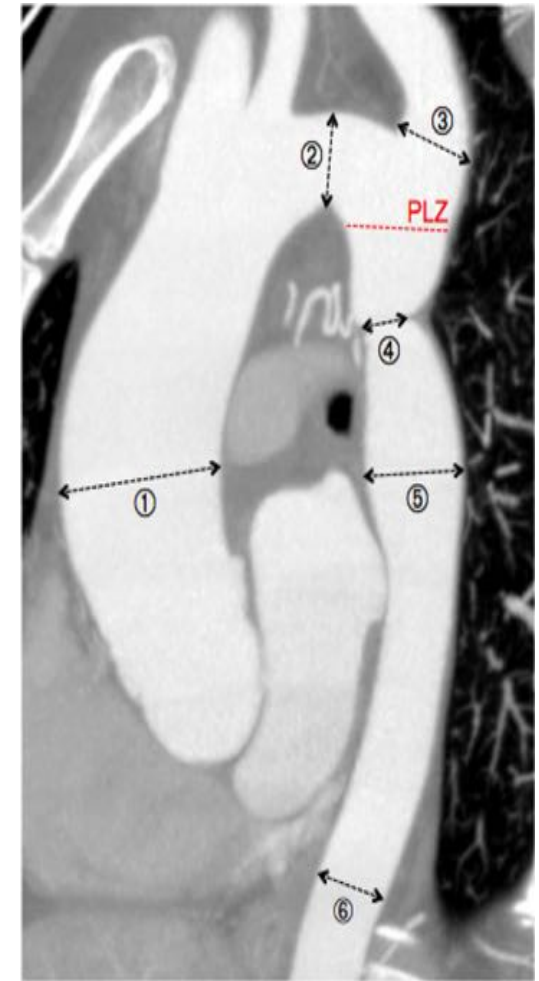
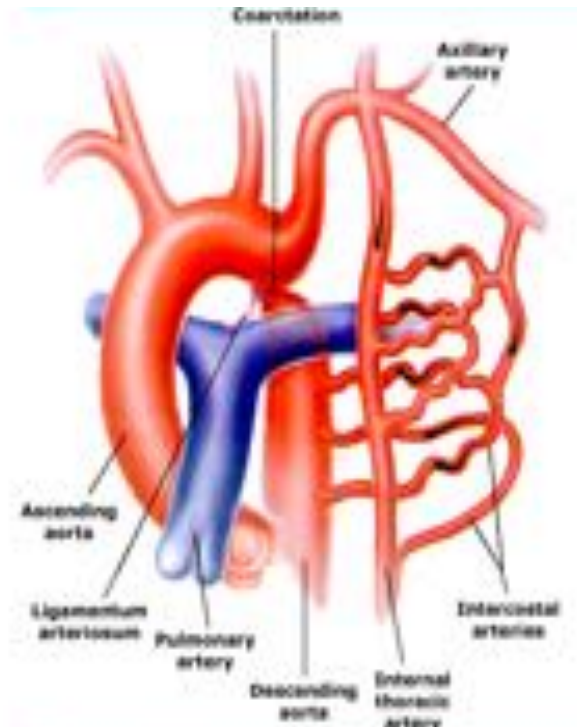


Fig. 1. Aortic arch anatomy in neonates.

Definition

- narrowing of the thoracic aorta $> 50\%$ compared to diaphragmatic aorta
 - Distal to left subclavian artery - ductus arteriosus
- Rarely long segment or tubular hypoplasia



Natural history of coarctation of the aorta

Maurice Campbell

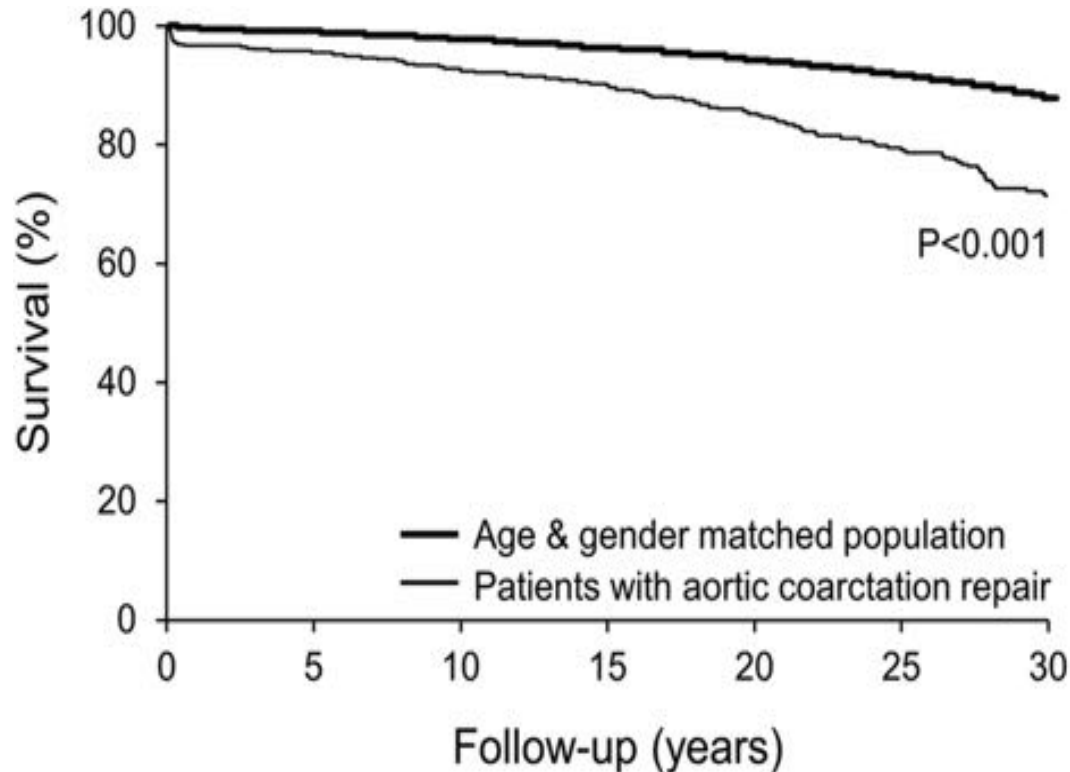
From the Cardiac Department, Guy's Hospital, London S.E.1, and the Institute of Cardiology, London W.1

TABLE 2 *Causes of death in coarctation of the aorta (304 necropsies)*

<i>Causes of death</i>	<i>Per cent</i>	<i>Mean age and usual decades</i>	
Congestive heart failure	25.5	39	3rd to 5th
Not directly connected	24	47	4th to 6th
Aortic rupture	21	25	2nd and 3rd
Bacterial endocarditis	18	29	First five
Intracranial haemorrhage	11.5	29	2nd and 3rd
All cases	100	34.4 years	

25% avant 20 ans
50% avant 32 ans
75% avant 46 ans
90% avant 58 ans

Survival lifetime reduced even treated

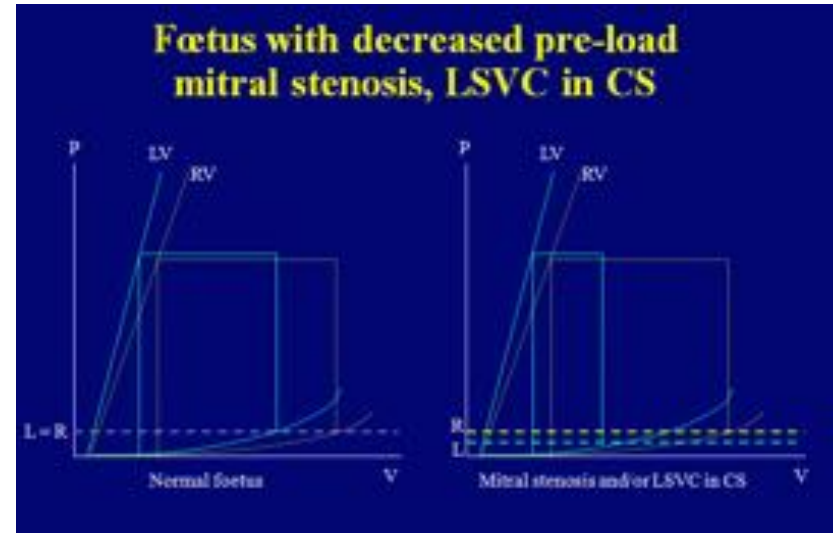


Survival of the entire cohort compared with an age- and sex-matched Minnesota population ($p < 0.001$)

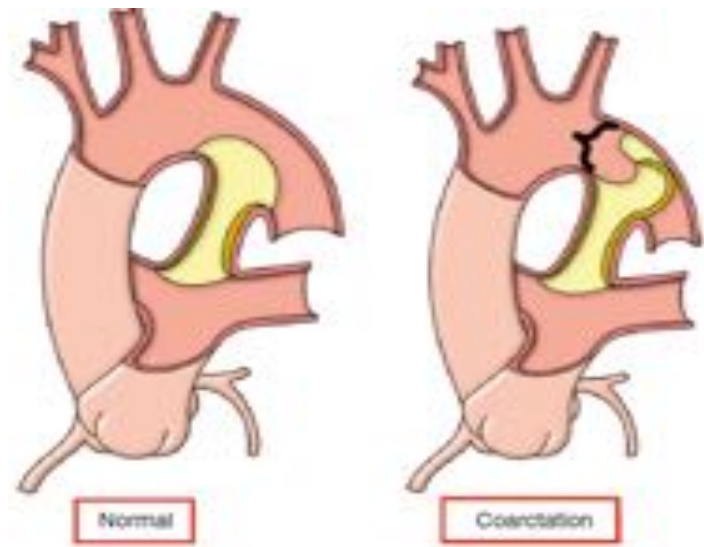
→ **Serious disease**

Morphogenèse et étiologie

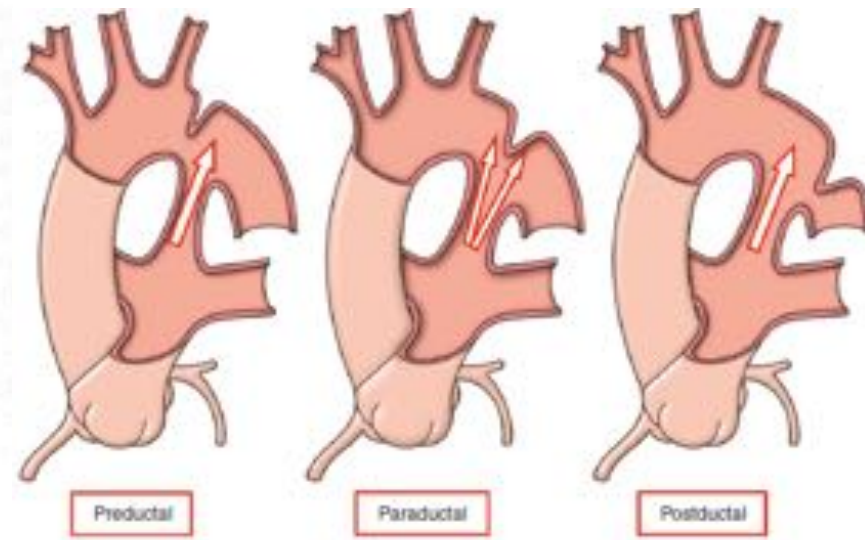
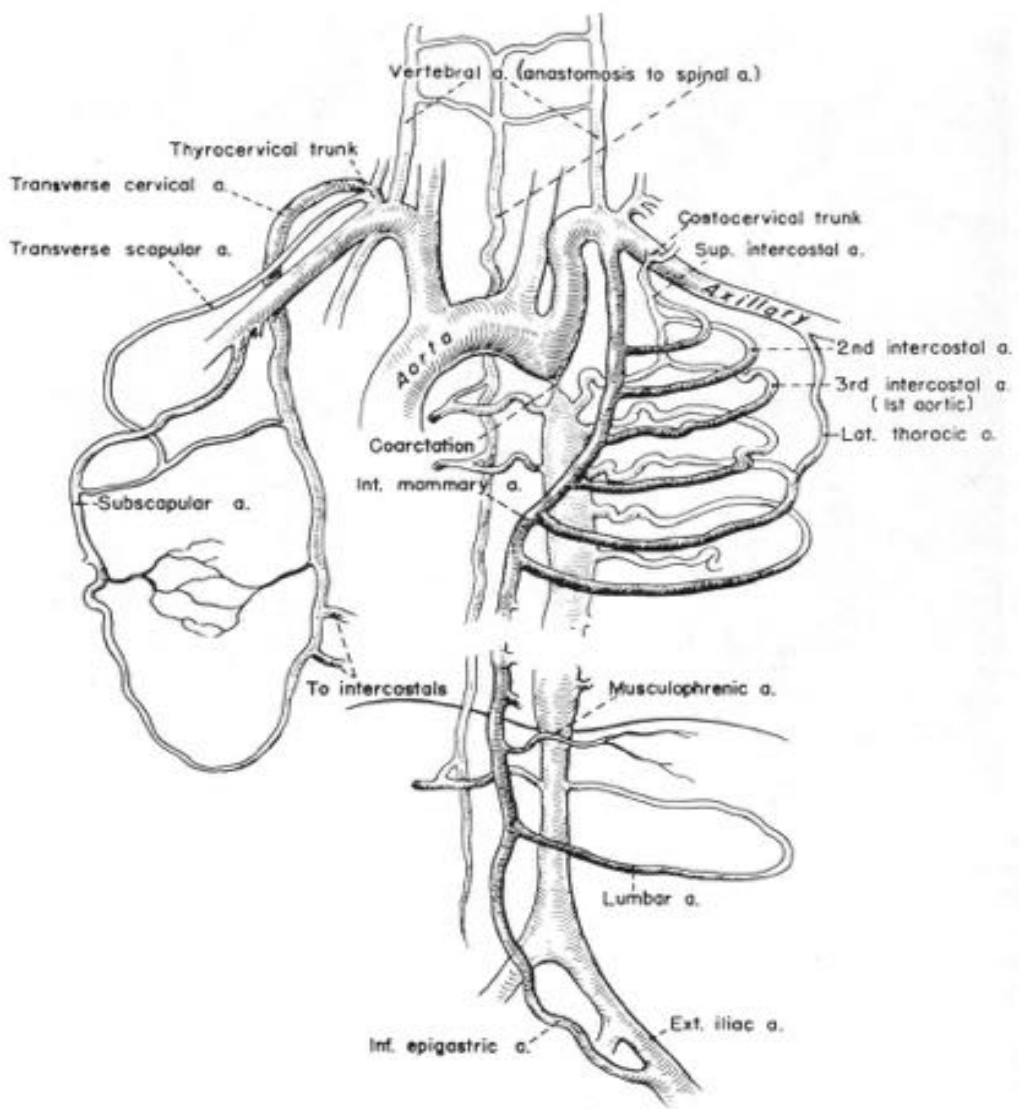
- **Théorie hémodynamique:**
Hypo-perfusion
anténatale (sténose
aortique, mitrale, FOP)>
Hypoplasie aortique



- **Hypothèse mécanique:** rôle du
tissu ductal
ectopique



Collatérales dans CoA

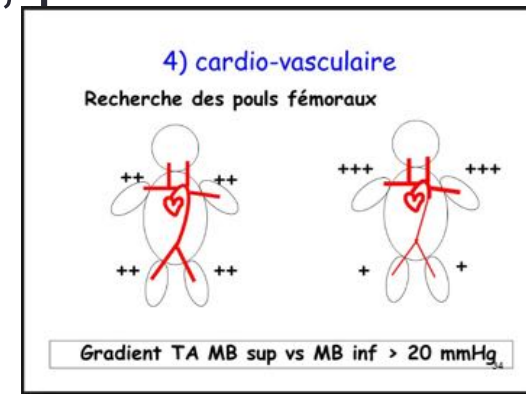


3 formes cliniques

- **Nouveau né :**

Coarctation serrée et de constitution brutale

- Latence 7-15 jours (fermeture du canal artériel)
- insuffisance cardiaque
voire choc cardiogénique/défaillance multiviscérale
- Abolition des pouls fémoraux
- Gradient tensionnel entre MS droit et MI, parfois faible du fait du choc.



3 formes cliniques

- **Nourrisson >1 mois :**

Coarctation de constitution plus progressive

- Asymptomatique le plus souvent (VG adapté)
- Souffle systolique sous-claviculaire gauche irradiant dans le dos
- Abolition des pouls fémoraux
- HTA MS droit
avec gradient tensionnel systolique >20 mmHg entre MS droit et MI.

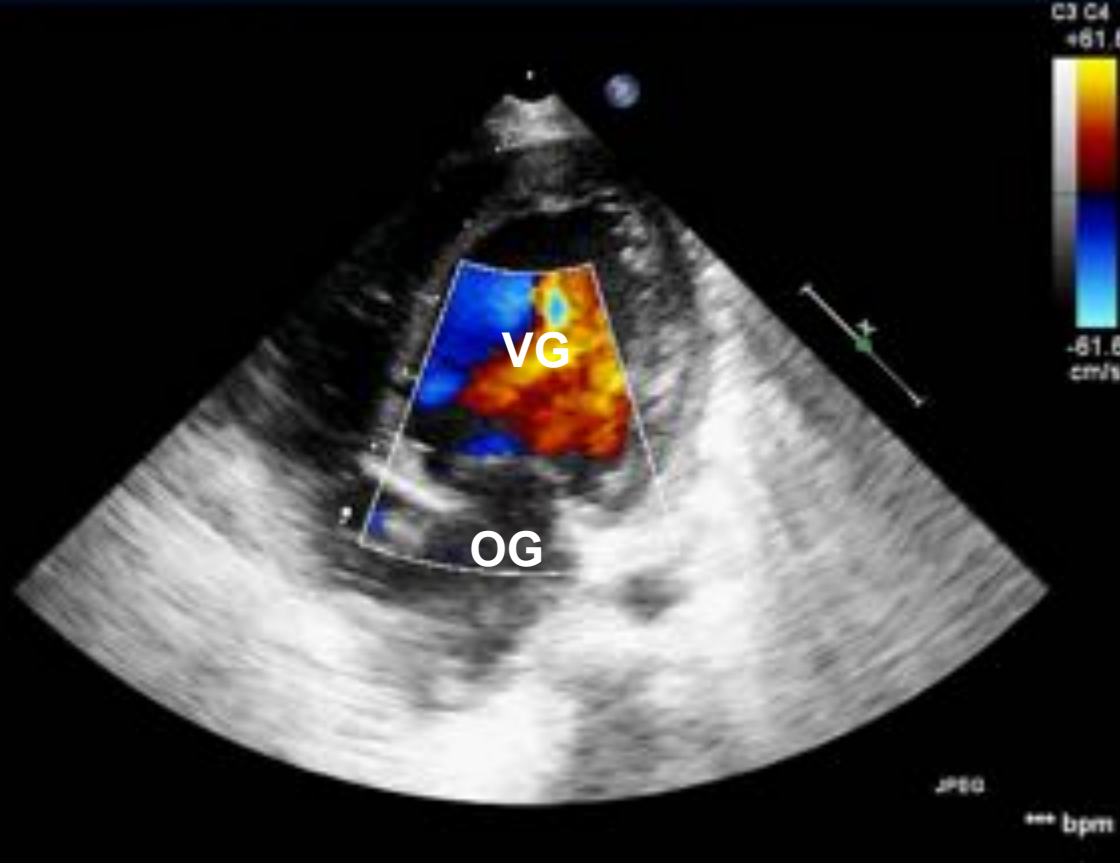
3 formes cliniques

–Grands enfants :

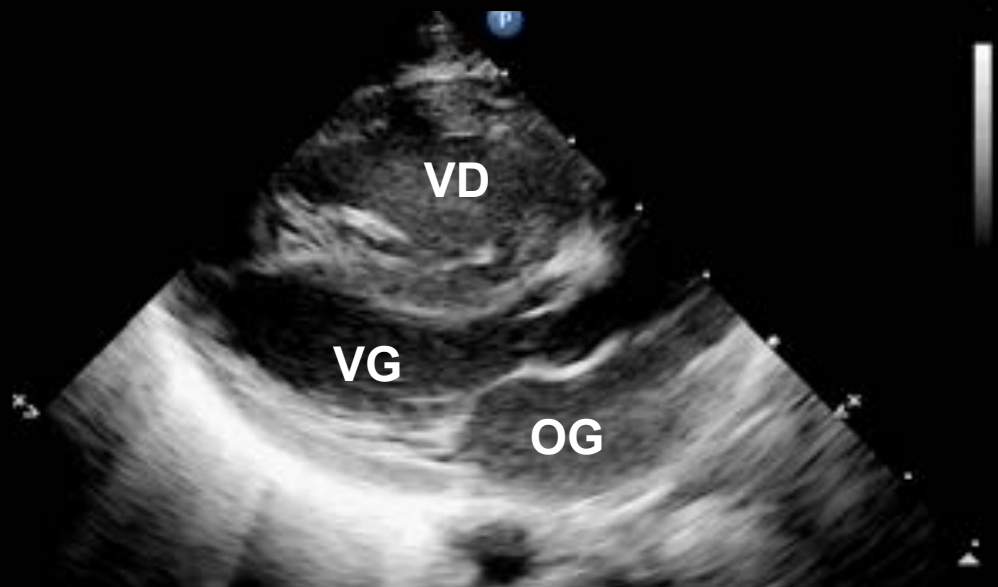
- HTA MS droit
- Insuffisance cardiaque très rare

Coarctation aortique : ETT

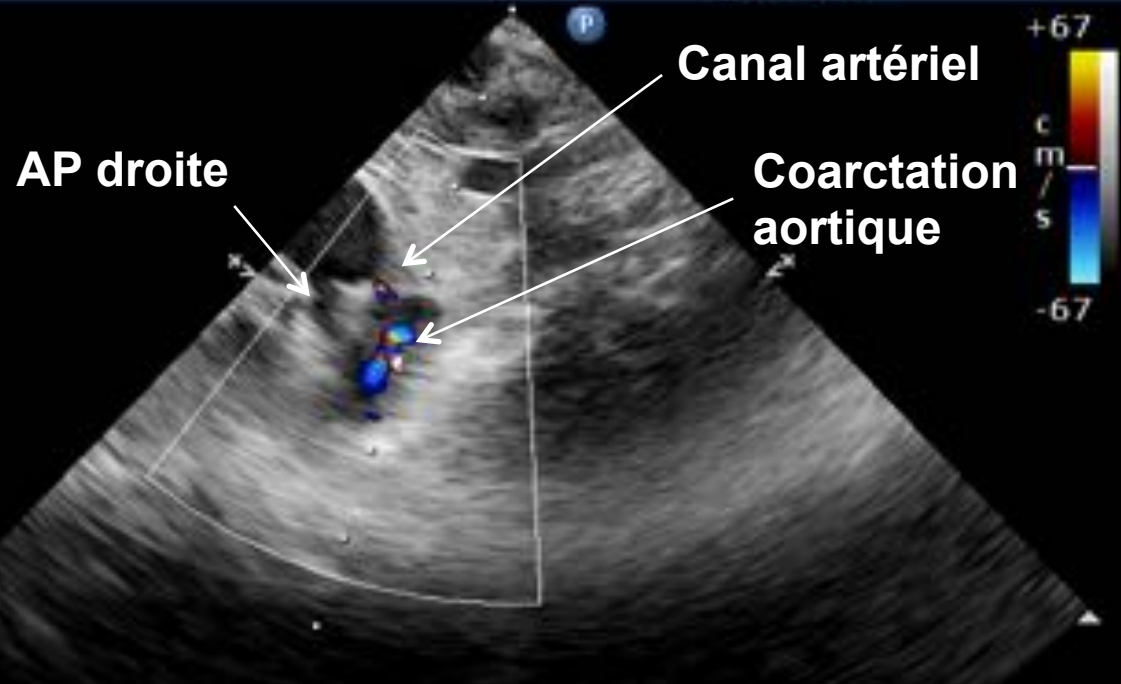
- Siège et étendue
- Doppler continu : accélération + prolongement diastolique
- Retentissement sur le VG :
 - Fonction VG
 - Hypertrophie VG
 - Estimation de la POG (vélocité du FOP)
 - HTAP post-capillaire
- Canal artériel
- Recherche de **lésions associées**



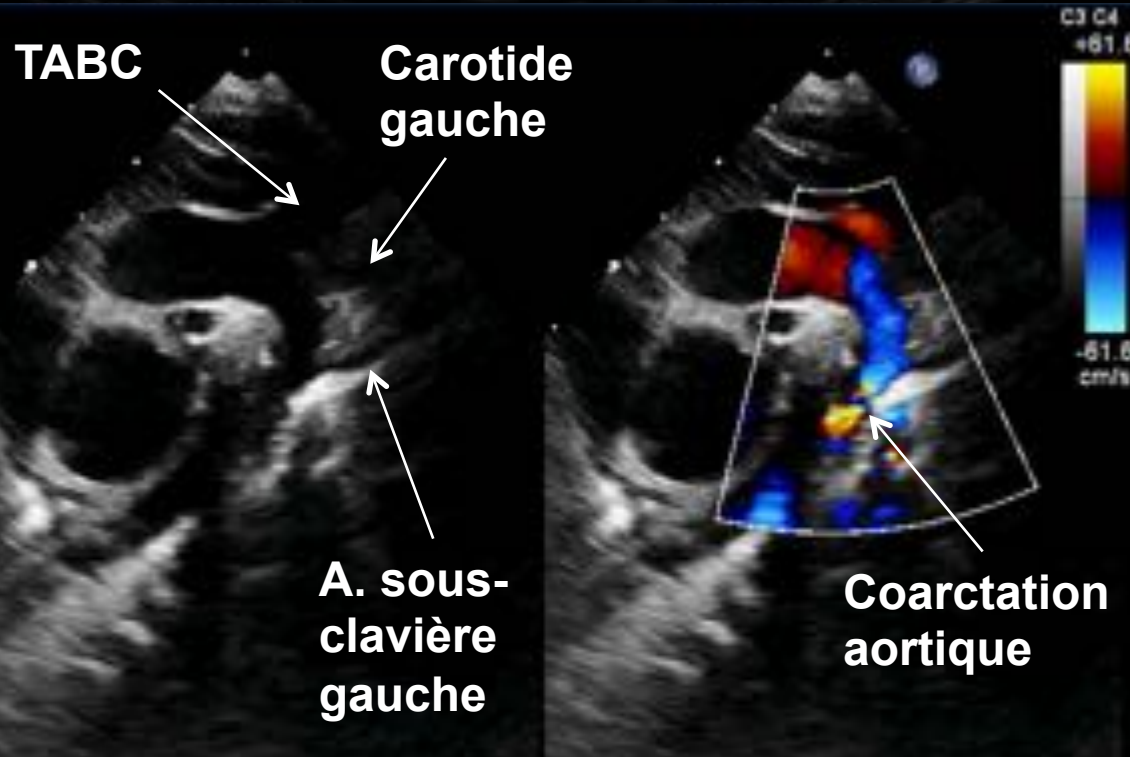
**VG dilaté
hypokinétique
4 cavités**



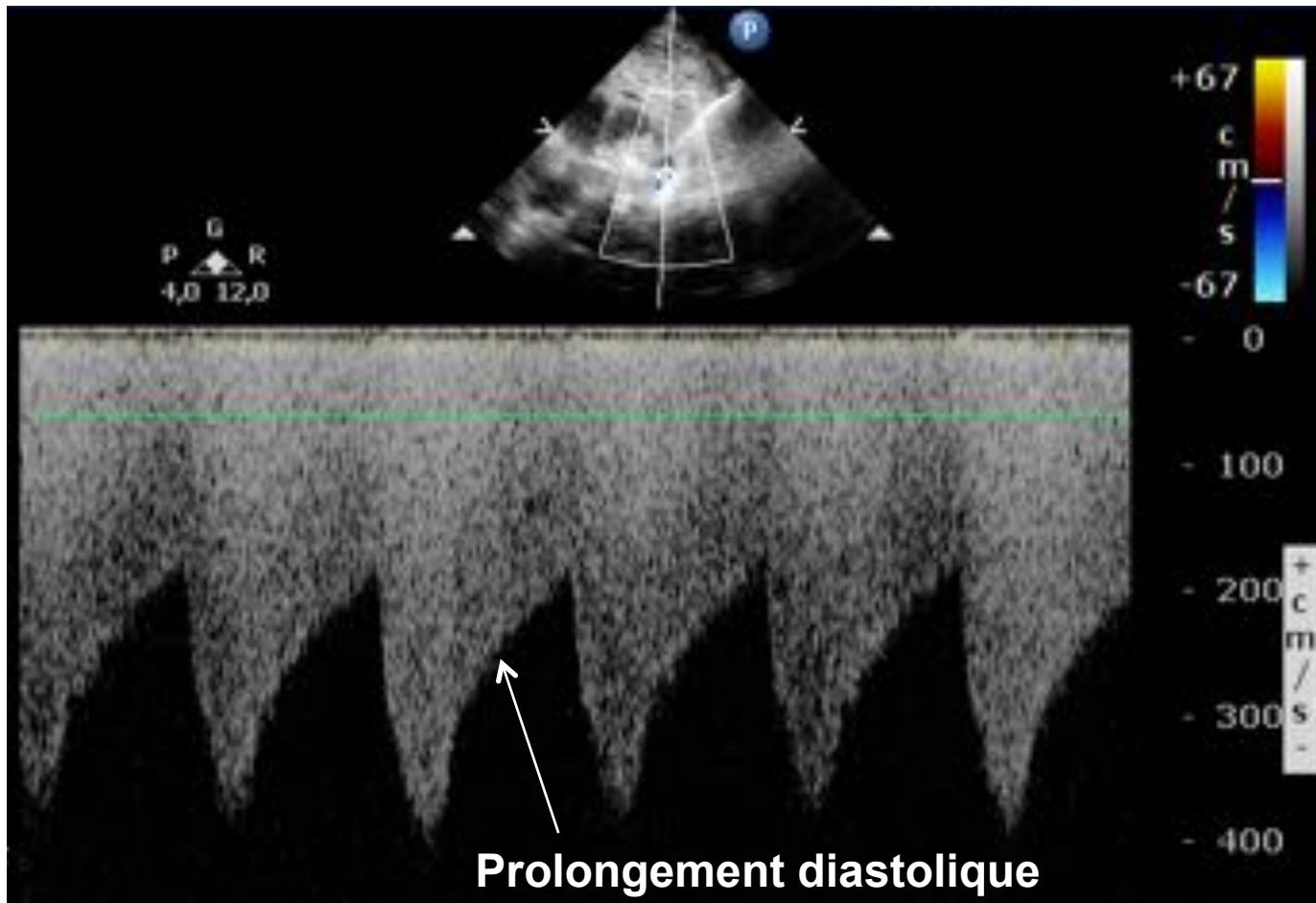
PSGA



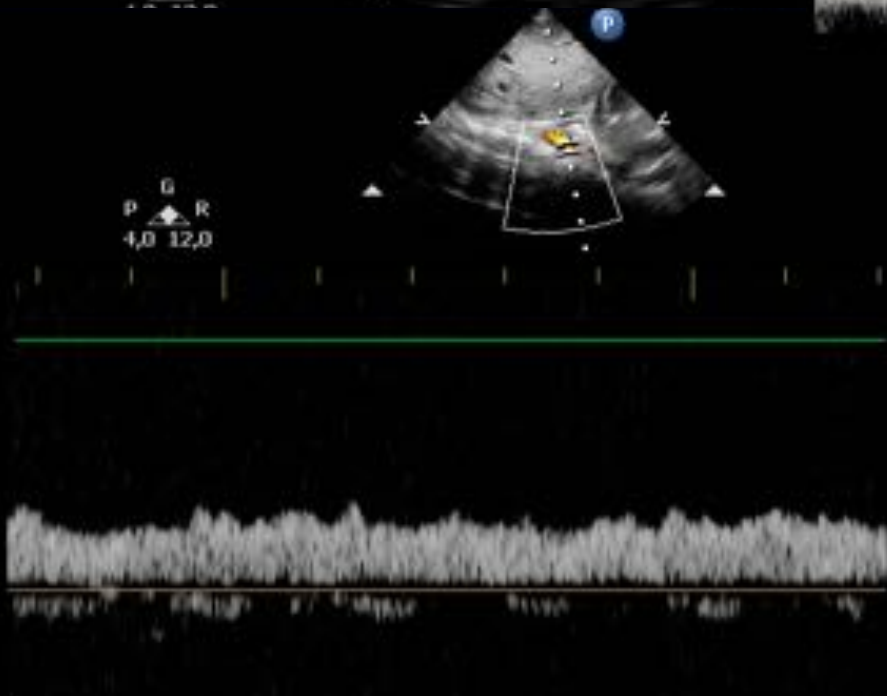
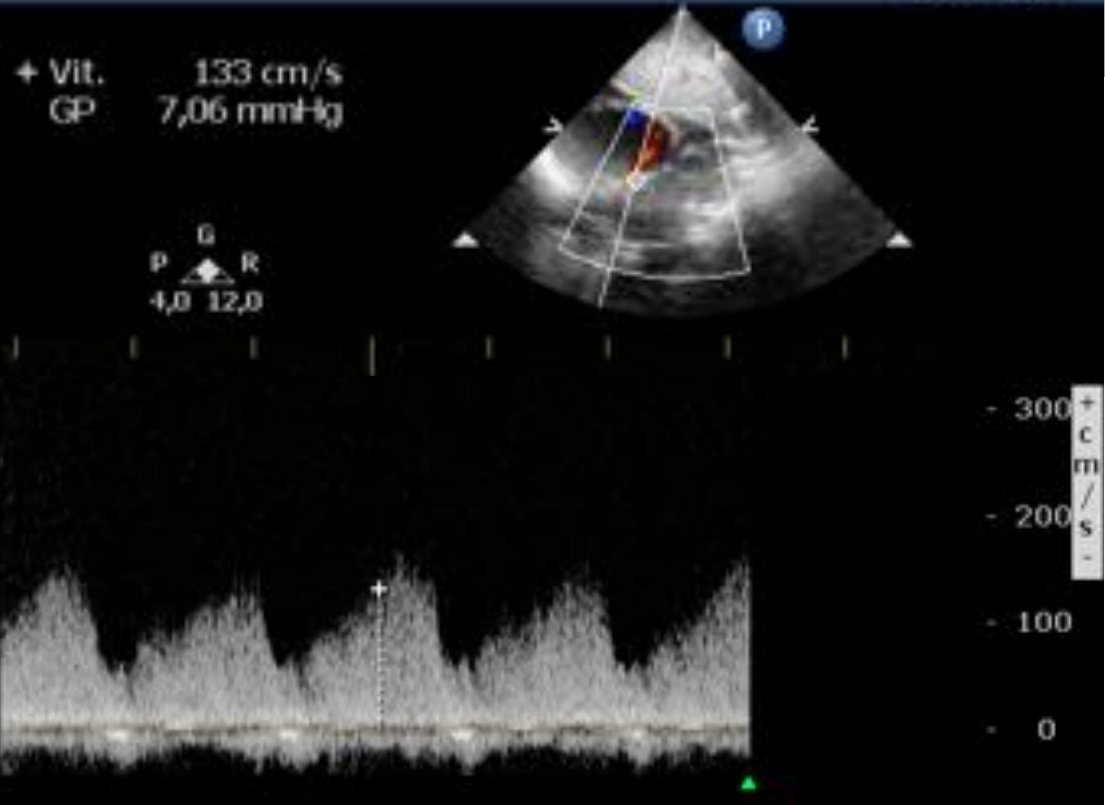
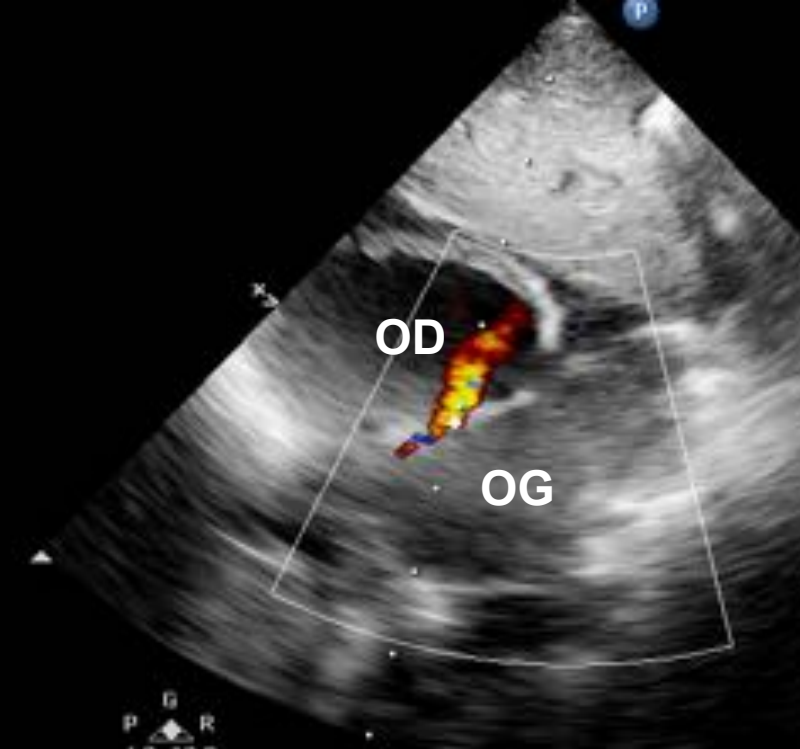
**Coarctation de
l'aorte
PSPA**



**Coarctation de
l'aorte
suprasternale**



**Coarctation de
l'aorte**
Doppler continu



**FOP véloce
sous-costale**

**Flux non pulsatile dans
l'aorte abdominale
sous-costale**

Imagerie de coupe

- **Forme néonatale** : pas systématique
- **Enfant ≥ 20 kg (6 ans)** : traitement percutanée (angioplastie + stent) ou traitement chirurgical

IMMOT-STACY
0815 F 003349713
100-Stat04 +c
Volume 35
Rendering Normal

HOPITAL DE PERPIGNAN
BERTRAND STACIUS
F 10 003349713-
008: Nov 12 2000
100 Sep 23 2011

DFOV 17.6 cm
STND Ph:80%

NOVL 100 LAD 39 CRA

0.5

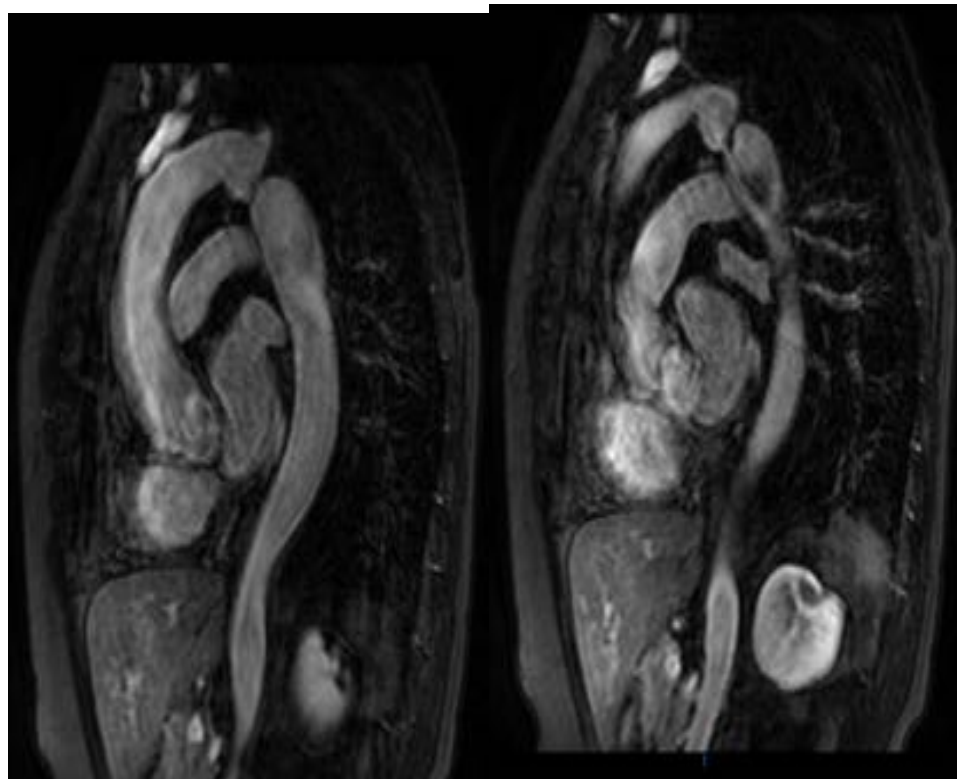
0.5

109/5

No VOI
kv 100
mA 550
Rot 0.35s/CI
0.6mm /0.6sp
Tilt: 0.0
20F000:08 AM
L1128 45344 -255

ILP



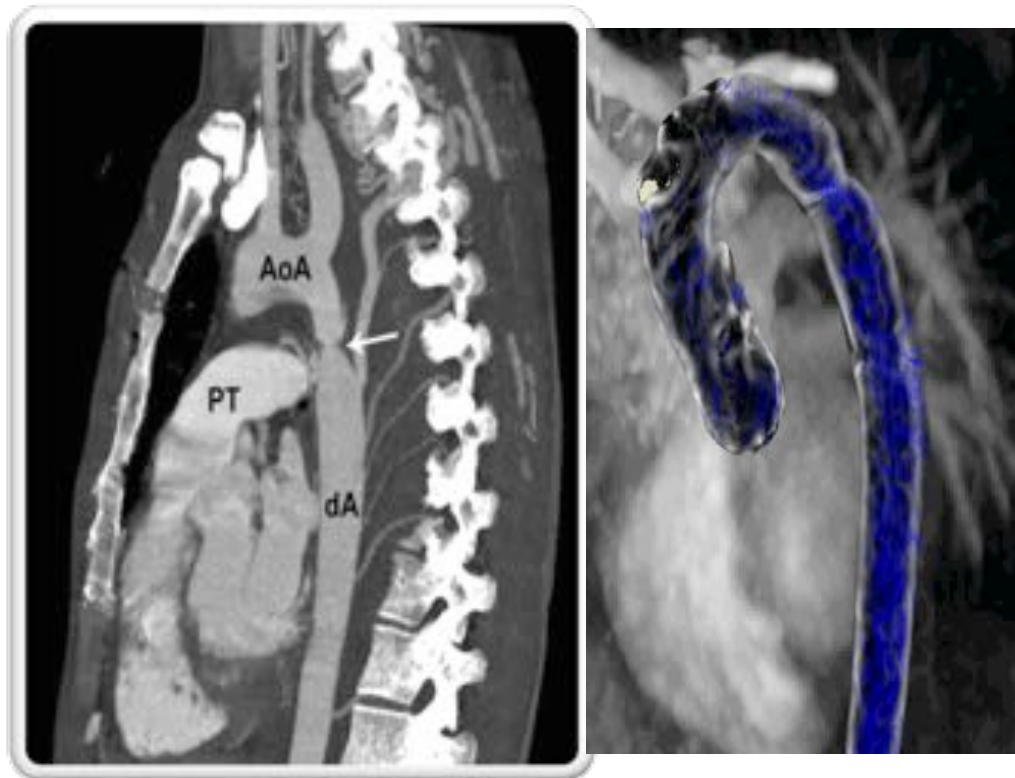


MRI

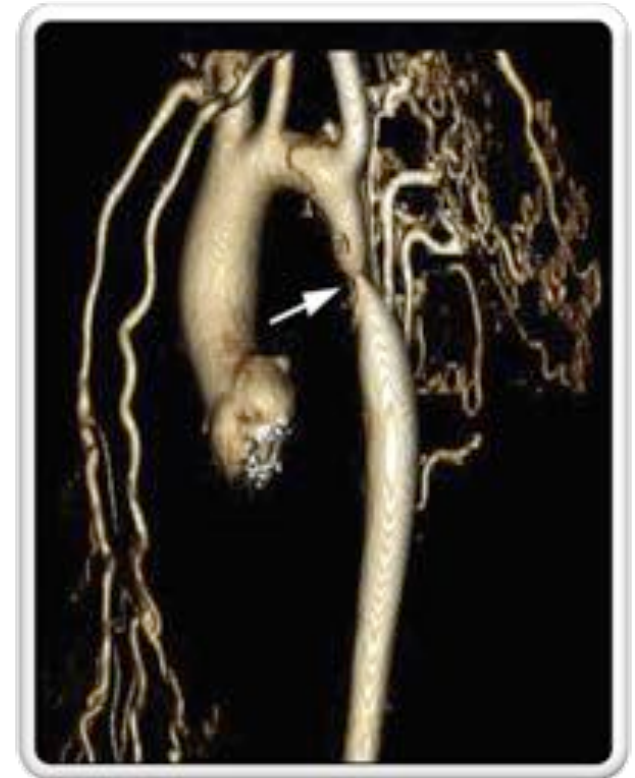


3D CT - scan

CT scan Vs IRM



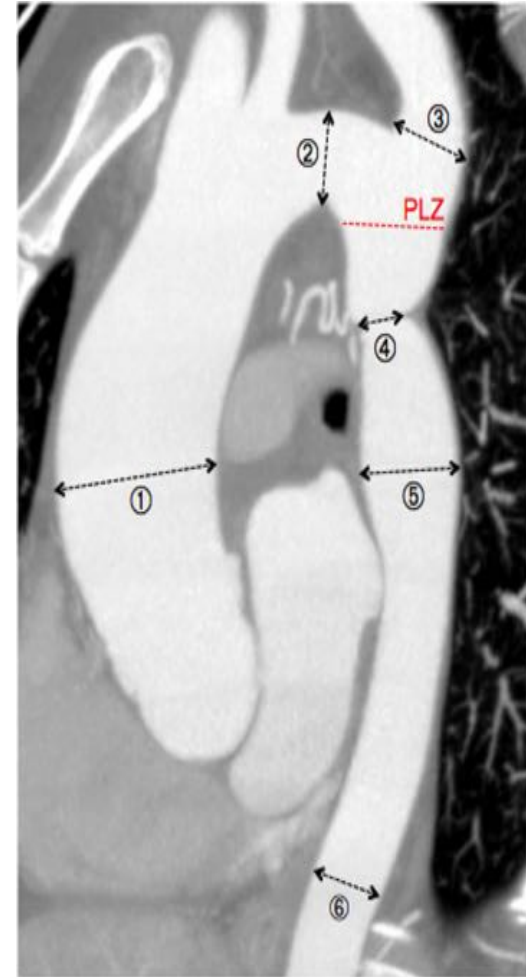
“Periodic imaging of the entire aortic arch, preferably with CMR, is recommended.”



Di Salvo et al. EHJ CVI 2018

Quel info?

- severity of coarctation
- anatomy of the aortic arch
- presence of collateral arteries
- suitability for stenting (cerebral vessels)
- pre-intervention LV mass, volumes and function



Traitement

- **Forme néonatale** : traitement médical pour permettre récupération fonction VG et hémodynamique avant chirurgie
 - Prostaglandine PGE1 (Prostine®) : lève l'obstacle isthmique + ouvre le canal artériel
 - Inotrope, diurétiques (Furosémide), ventilation mécanique
- Traitement étiologique :
 - **Pour nouveau-né, nourrisson et enfant <20 kg** : traitement chirurgical (clampage aortique sans CEC)
 - **Enfant ≥20 kg (6 ans)** : traitement percutanée (angioplastie + stent) ou traitement chirurgical

Quand traiter chez le grand?

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

Anatomic coarctation + gradient > 20mmHg
Or Gradient > 10mmHg with collaterals

Even without gradient with HTA

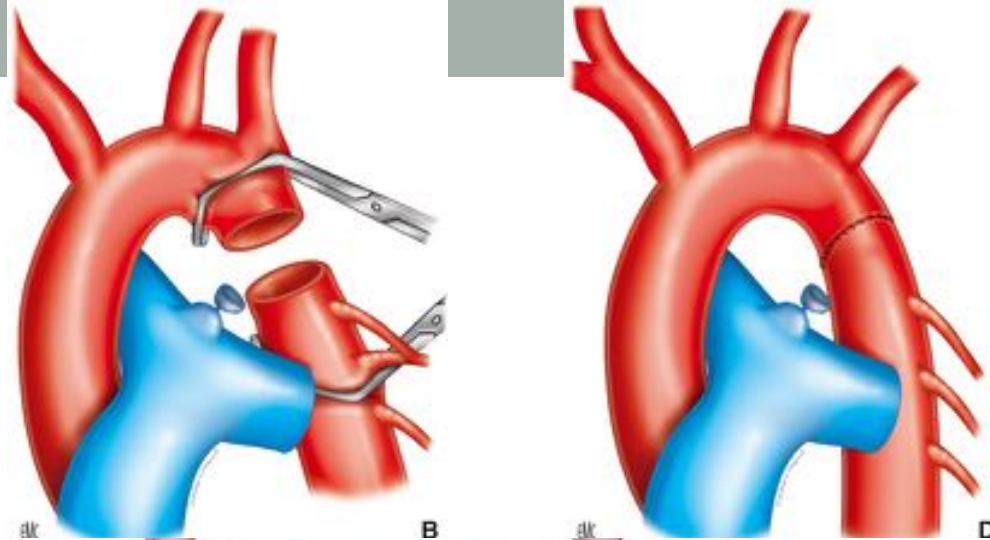
ESC GUIDELINES



European Heart Journal (2010) **31**, 2915–2957
doi:10.1093/eurheartj/ehq249

Chirurgie

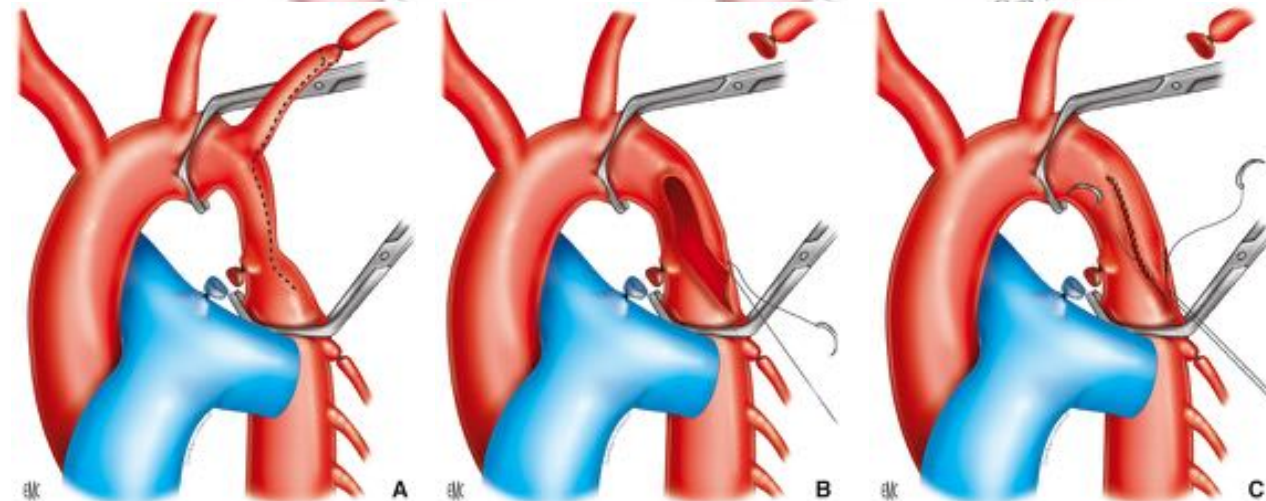
End to End (Crafoord
1945)



Extended end to end
(Crafoord modifié)



Flap sousclavière
(Waldhausen)



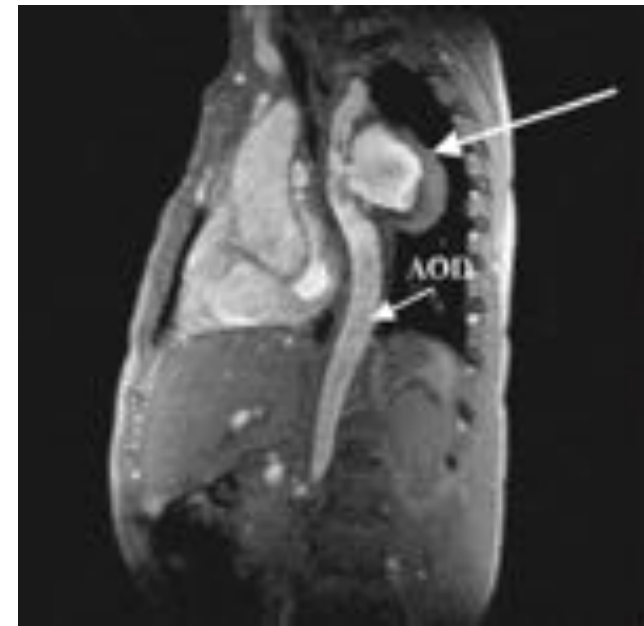
Résultats

- Mortalité :
 - 1 à 2 % si isolée
 - < 10 % si anomalies intracardiaques complexes associées
- Recoarctations :
 - 10 à 20% des Coarctations opérées en période néonatale
 - Facteurs de risques:
 - Age moins de 3 mois lors de la cure.
 - Presence d'hypoplasie de l'aorte transversale.



Complications post-opératoires

- Précoces:
 - HTA paradoxale
 - Troubles digestifs (syndrome post coarctectomie)
 - Pulmonaires : compression bronchique, chylothorax
 - Neurologique: paraplégie, paralysie récurrent
- Tardives:
 - HTA (effort)
 - Recoarctation
 - Anévrisme et faux anévrisme



Dilatation percutanée coarctation native ou recoarctation



European Journal of Cardio-thoracic Surgery 39 (2011) 898–904

SUPPLEMENTARY JOURNAL OF
CARDIO-THORACIC
SURGERY

www.elsevier.com/locate/ejcts

Comparison of surgical and interventional therapy of native and recurrent aortic coarctation regarding different age groups during childhood

Seraina Früh^{a,1}, Walter Knirsch^{b,1,*}, Ali Dodge-Khatami^a, Hitendu Dave^a,
René Prêtre^a, Oliver Kretschmar^b

^a Division of Pediatric and Congenital Cardiac Surgery, University Children's Hospital Zurich, Zurich, Switzerland

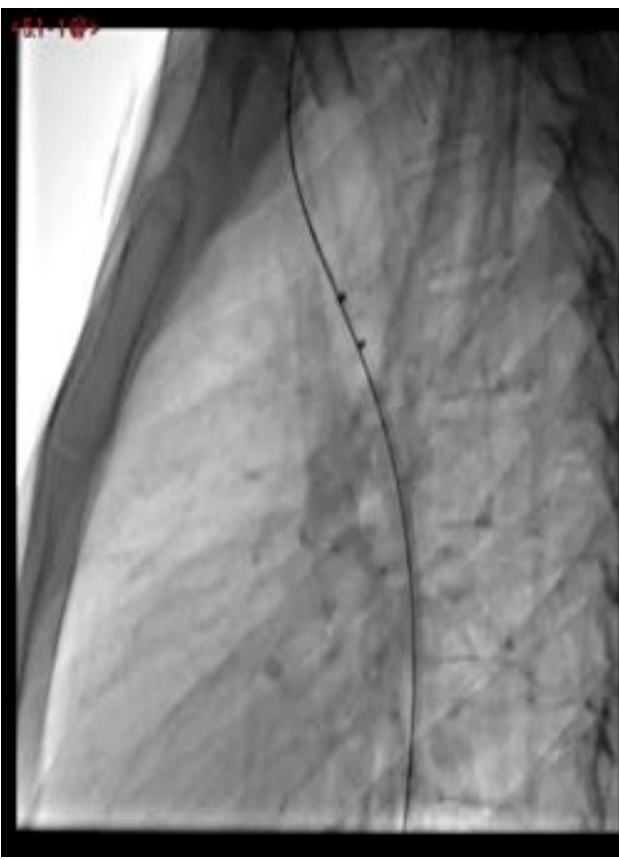
^b Division of Pediatric Cardiology, University Children's Hospital Zurich, Zurich, Switzerland

Received 2 March 2010; received in revised form 21 September 2010; accepted 22 September 2010; Available online 18 December 2010

In the very young patients <6 months of age (group A), the early and long-term results of surgical resection are excellent. These data confirm surgery as the treatment of choice in this specific age group.

In children between 6 months and 6 years of age (group B) with isolated CoA, the acute results of reduction of gradients between surgery and angioplasty are comparable, as shown in the literature for similar age groups. In our series, the rate of re-interventions in this age group was higher after angioplasty (37.5%). Stent implantation is not recommended in this age group due to the need for frequent redilatation in growing children, the lack of redilatable stents until adulthood, a high incidence of intimal proliferation in small stents and the potential development of post-stent aneurysm formation.

In children >6 years of age (group C), stent implantation is an excellent therapeutic alternative to surgery.



Stent vs Surgery (adulte)

Stent : 302 patients from 34 centers

Efficacy :

- Acute procedural success was 96%

Safe:

- Mortality <1% (0% in this serie)
- Risk = 5% (stent migration, aortic wall injury)
- unplanned reintervention of 4%

Less invasive !

1 study compare to surgery (217 vs 72) : Stent = shorter hospitalization (2.4 vs. 6.4 days; $p < 0.001$) and fewer complications (2.3 vs 8.1% $p < 0.001$)

No strict recommendation “suggests a need to perform a prospective randomized controlled clinical trial”

Hypertension

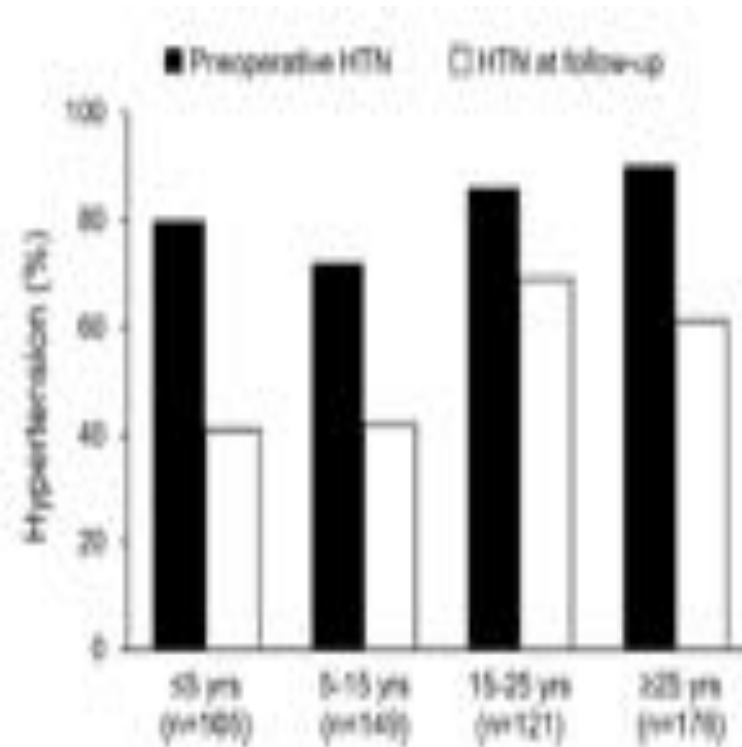


Figure 5 Hypertension Pre- and Post-Repair

Comparison of hypertension pre- and post-operatively at various time intervals of follow-up (All $p < 0.001$).

Traquer HTA

- Prise de TA Musp : Droit ++
 - sacrifice SCG
 - Attention aux anomalies des arcs
- Prise de TA Minf : gradient TA
 - Attention aux sténoses fémorales post KT
- MAPA TA (effet blouse blanche)
 - Msup droit ou TA la plus élevée

HTA et Coarctation de l'aorte opérée

Rôle de la géométrie de la crosse:

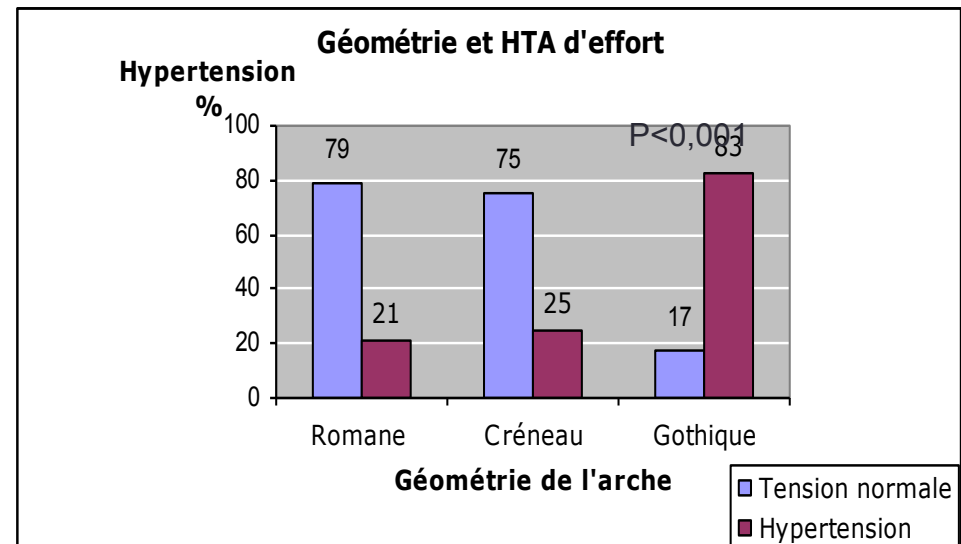
Phalla Ou MD; JAAC 2007



Normal

Gothique

Créneau



Suivi

- **Cause of early death**
- **Hypertension (be careful : right upper limb + look for gradient + Ambulatory BP if suspected)**
 - Pronostic factor = age at correction
- **Aortic disease**
- **Re-intervention 124/819 in surgical cohort**
- At least / 2ans in GUCH center

Table 2 All Reoperations and Re-interventions (n = 124) /819

Reoperation or Reintervention Type	No. of Interventions
Coronary reoperation	
Surgical	40
Catheter based	13
Aortic valve replacement	10
Aortic valve repair	17
Coronary artery bypass grafting	13
Mitral valve replacement	10
Ascending aorta repair	8
Mitral valve repair	8
Descending aorta anastomosis	8
Aortic dissection	2
Other (main, AED, PFO, pericardiotomy, subclavian anastomosis, valvular stenosis)	9
Total	124

AED, PFO = atrial septal defect, patent foramen ovale

Coarctation et anevrisme cerebral

10% des patients
Souvent petits

Study from american database : stroke at younger age
(18yo younger)!



IIb	B-NR	4. Screening for intracranial aneurysms by magnetic resonance angiography or CTA may be reasonable in adults with coarctation of the aorta (S4.2.6-5, S4.2.6-6).
-----	------	--

Curtis SL et al. AJNR 2012

Cook SC et al. CHD 2013

Pickard et al. JAHA 2018

Stout et al. JACC 2018

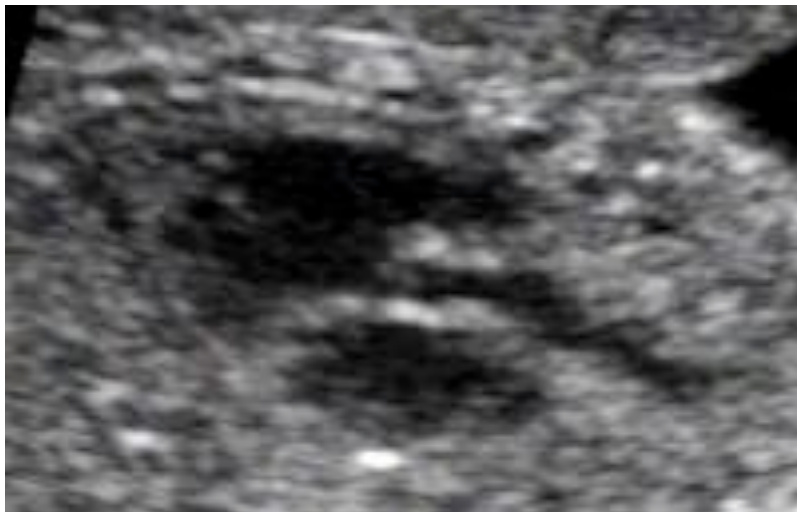
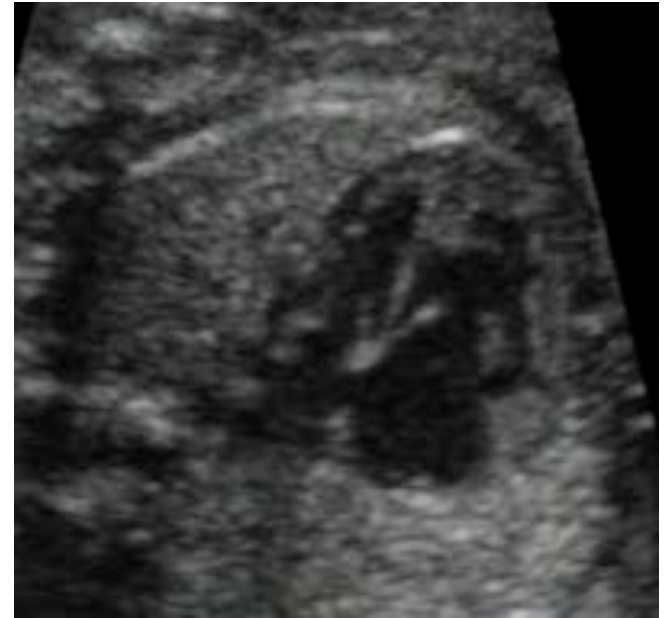
Grossesse et coarctation

- Bilan pré-conceptionnel
- Après réparation = WHO risk class II.
- Si non réparé :
 - + de pre-eclampsie et fausse couche + risque dissection
- → Surveiller PA +++ (stent à discuter)
- **Turner:**
 - Grossesse possible (après don d'ovules, exceptionnellement spontanément)
 - Contre indiquée si :
 - aorte $\geq 25\text{mm/m}^2$
 - histoire de la chirurgie aortique,
 - HTA non contrôlée

Dépistage anténatal

VG<VD

Suspectée devant une asymétrie
AO<AP



Risque de coarctation post natale

Durand et al. Pediatr Cardiol Mars 2015

- Asymétrie de taille dès le 2^{ème} trimestre de la grossesse
- Anneau *Ao* < 5 mm au 3^{ème} trimestre de la grossesse
- La crosse de l'aorte est *hypoplasique*
- Il existe des *anomalies associées*: CIV, anomalies de la valve mitrale, de la valve aortique



Recurrence of familial Left heart obstruction: Continuum

HypoVG



bicuspidie

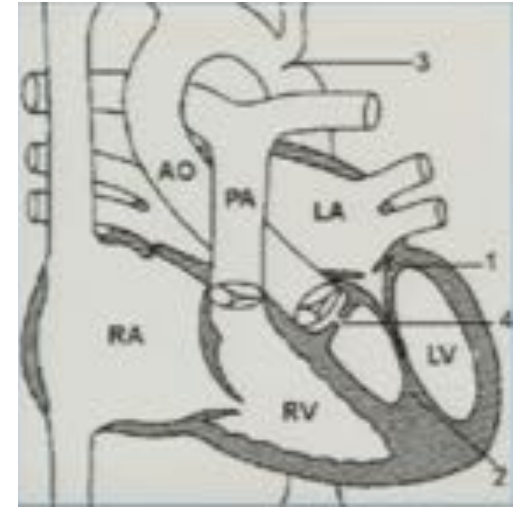


coarctation



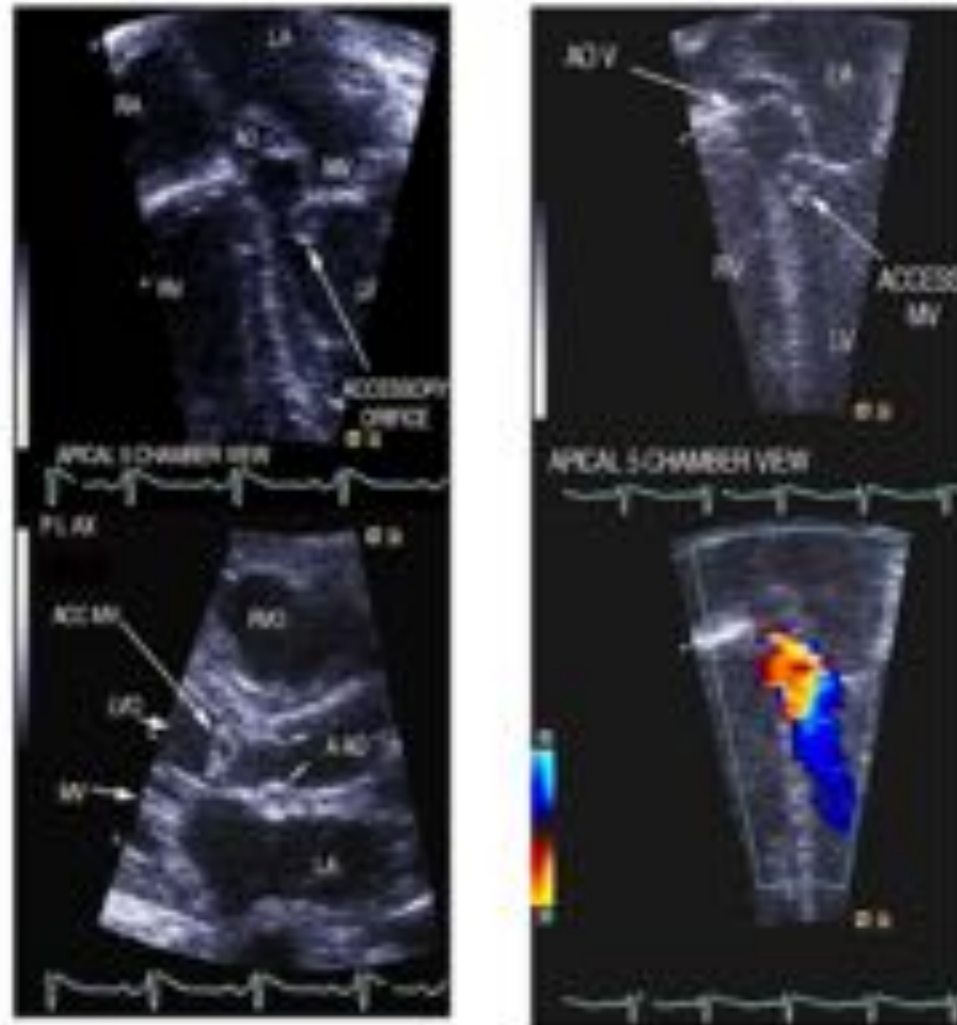
Le complexe de Shone

- Complet:
 - Valve mitrale parachute
 - Anneau mitral supra-valvulaire
 - Obstacle sous aortique
 - Coarctation aortique
- Partiel : 1 des obstacles OG-VG, + au moins 1 autres anomalies des structures gauches:
 - Obstacle sous aortique
 - Sténose aortique
 - Bicuspidie aortique
 - Hypoplasie de l'arche aortique
 - Coarctation aortique



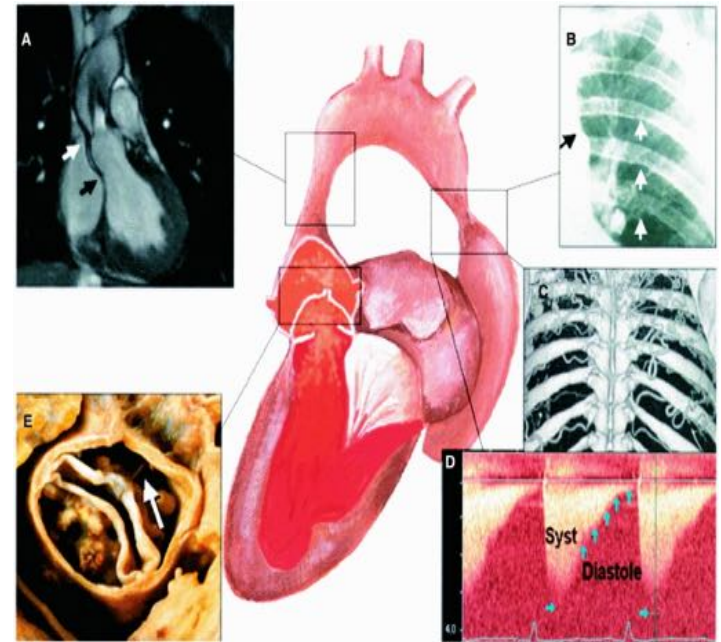
Shone *et al*, Am J cardiol 1963

Le complexe de Shone : la voie sous aortique



Conclusion

- Obstacles étagés du coeur gauche
 - Symptômes?
 - ETT: meca, sévérité, retentissement, lésions associées
-
- Merci : Khaled Hadeed, Romain Amadiou, Philippe Acar, Laurence Iserin



Coeur triatrium

Cor Triatriatum Sinister (CTS)

Anatomic Morphologies

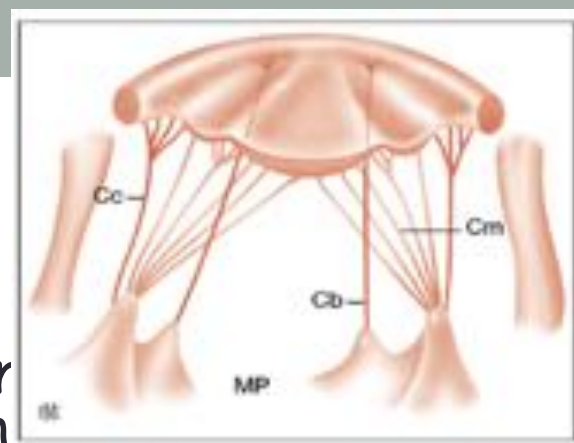


• Embryology: 3 hypothesis of development.

1. **Malseptation** – abnormal growth of septum primum
2. **Malincorporation** – Failure of pulmonary vein fusion to LA
3. **Entrapment** – Left horn of sinus venosus entraps common pulmonary vein – which remains in left atrial cavity

Historical Context	Year	Physician Scientists
First Recorded Description	1888	Church & Andral
Coined "Cor Triatriatum"	1905	Borst
Angiographic Description	1964	Miller et al
Echo Diagnosis	1984	Oldman-Smith
First Surgical Resection	1956	Vineberg & Giallaretto

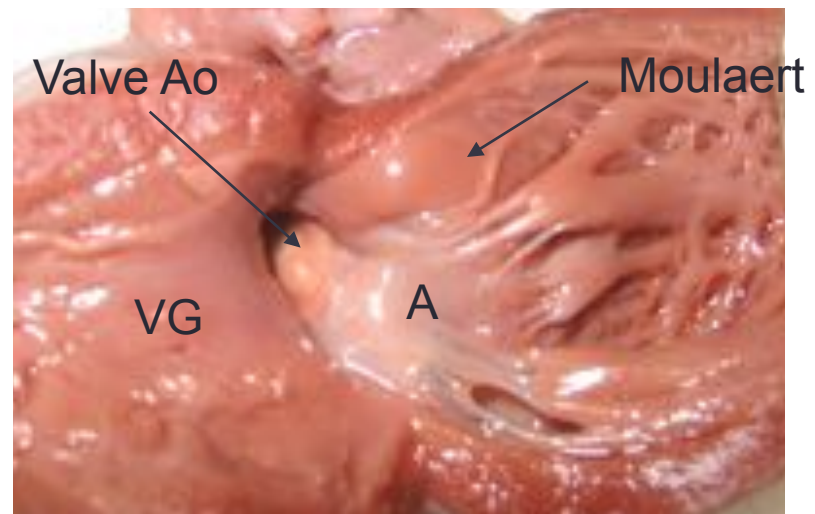
Valve mitrale



- La valve mitrale se forme à partir du mésobourrelets endocardiques du canal atrioventriculaire d'une partie de la cloison interventriculaire à partir de la cinquième semaine de vie embryonnaire.
- L'appareil mitral normal est constitué de quatre éléments, d'origine embryologique différente:
- le voile (feuillet mitral antérieur et feuillet mitral postérieur),
- l'anneau,
- les cordages tendineux (naissance à partir des portions apicales des muscles papillaires puis ramification et insertion sur le versant ventriculaire des valves), et
- les muscles papillaires ou piliers (pilier postéro-médial et pilier antéro-latéral).

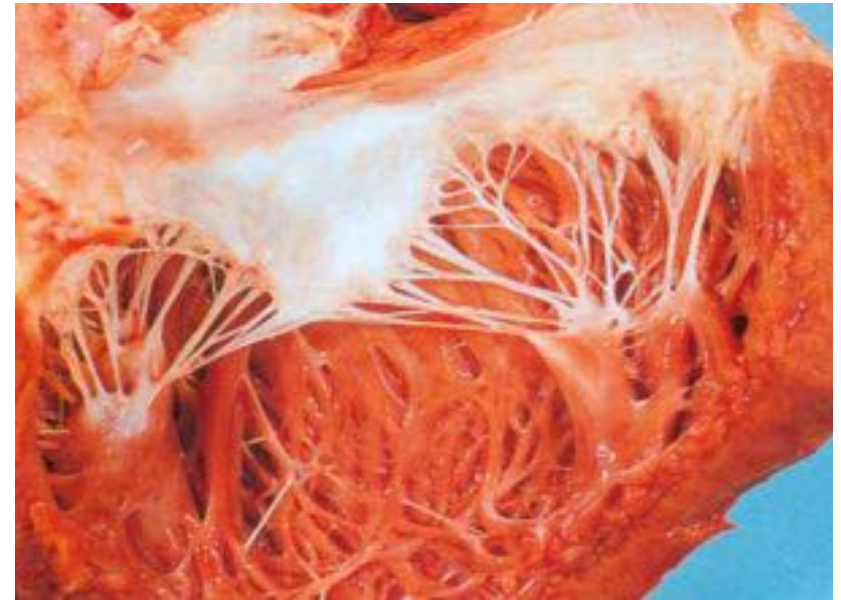
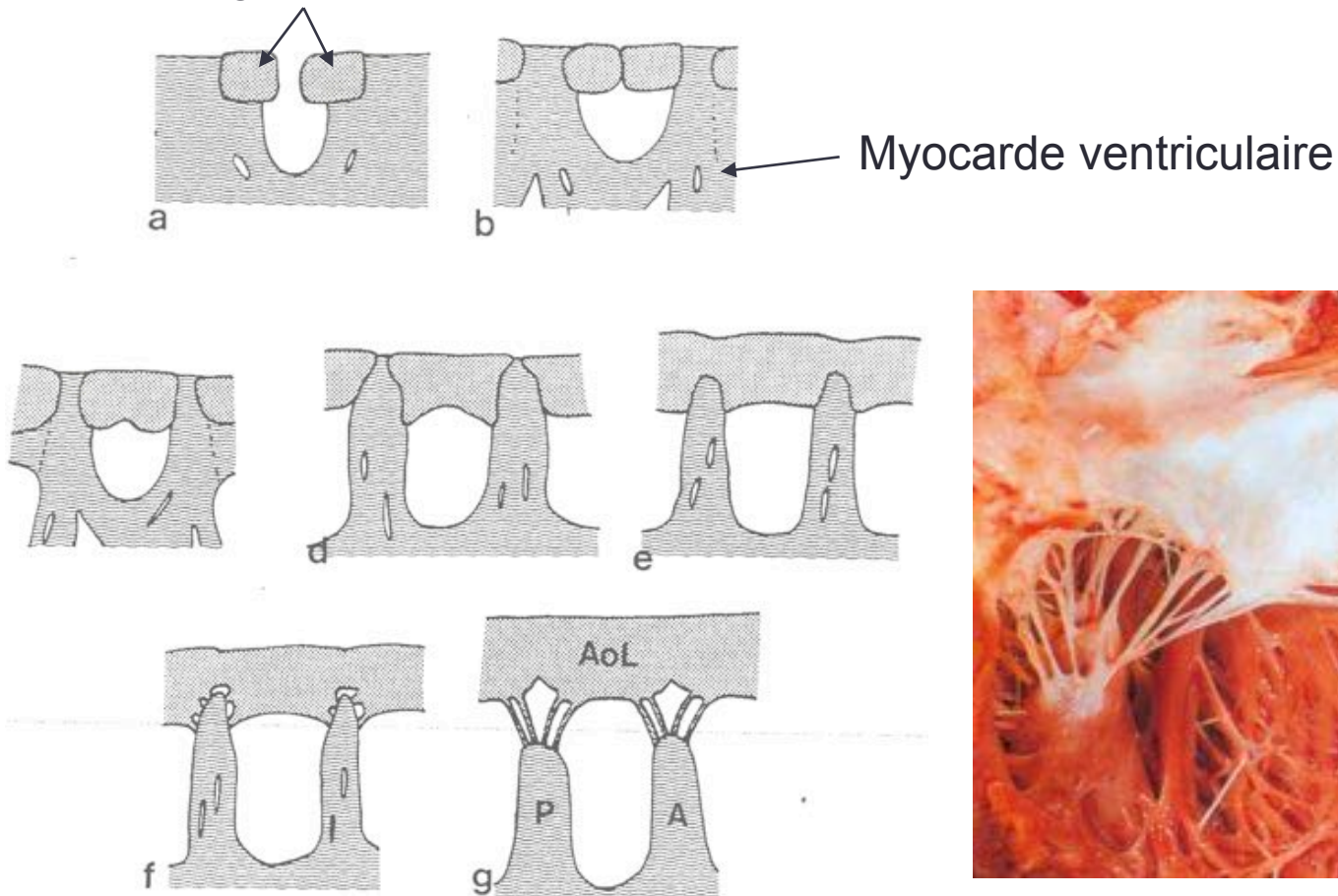
Formation de la valve mitrale

- Formation de la continuité mitro-aortique (wedging)
- Disparition des dérivés de la courbure interne (persistance = muscle de Moulaert)



Formation des piliers mitraux

Bourgeons endocardiques



Oosthoek et al. JTCS 1998 ; 116 : 36-46

Coarctation aortique in utéro

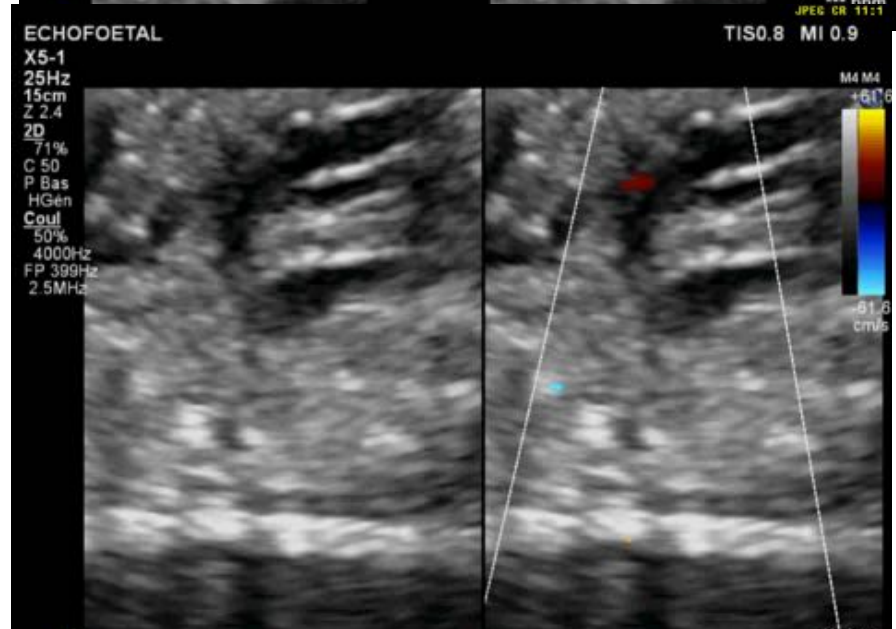
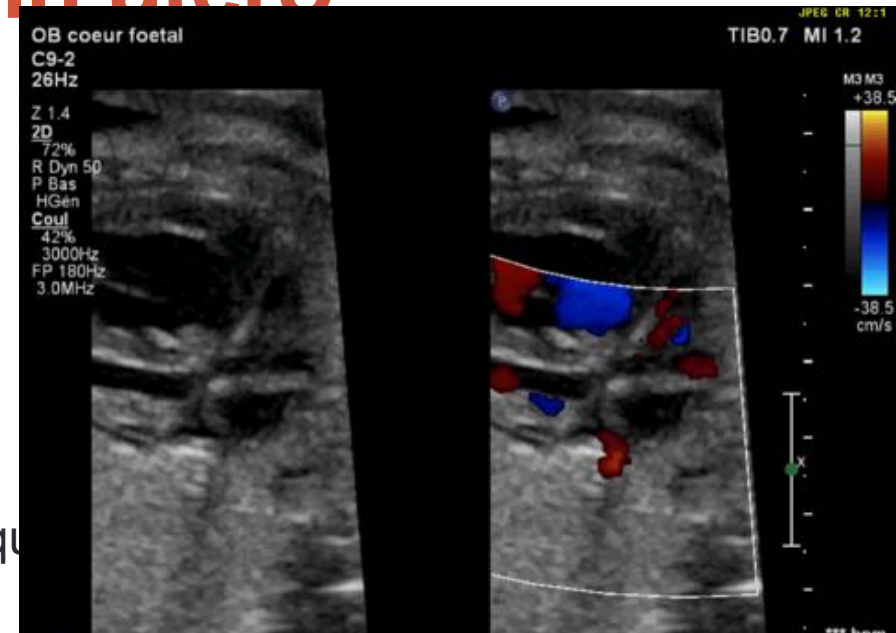
Asymétrie au profit VD:

$VD/VG \geq 1,5$

$AP/Ao \geq 1,5$

Hypoplasie Ao horizontale et isthmique

Shunt G-D auriculaire+++



• Le diagnostic prénatal réduit la morbidité (neurologique) et la mortalité néonatale.

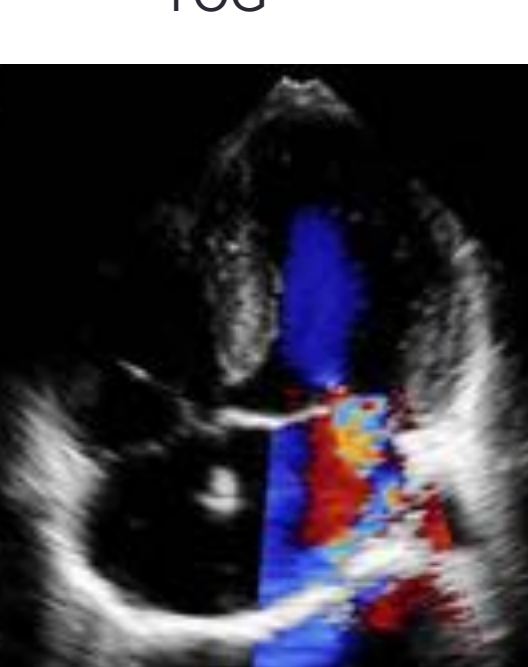
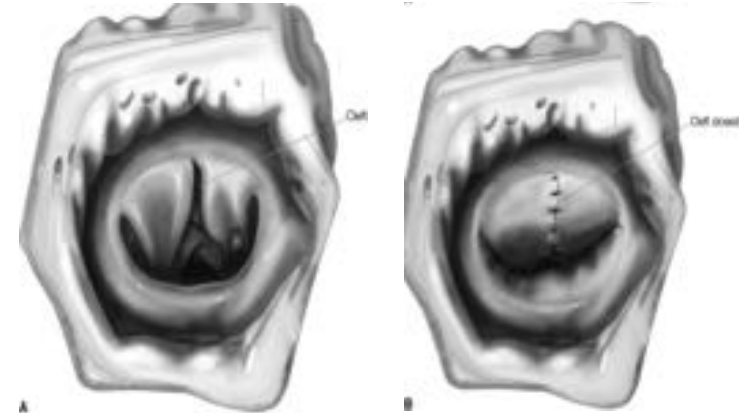
- Eapen RS et al. Am J Perinatol. 1988;14:237-41.
- Franklin O et al. Heart 2002;87:67-69.

DAN N	OUI 22	NON 10
----------	-----------	-----------

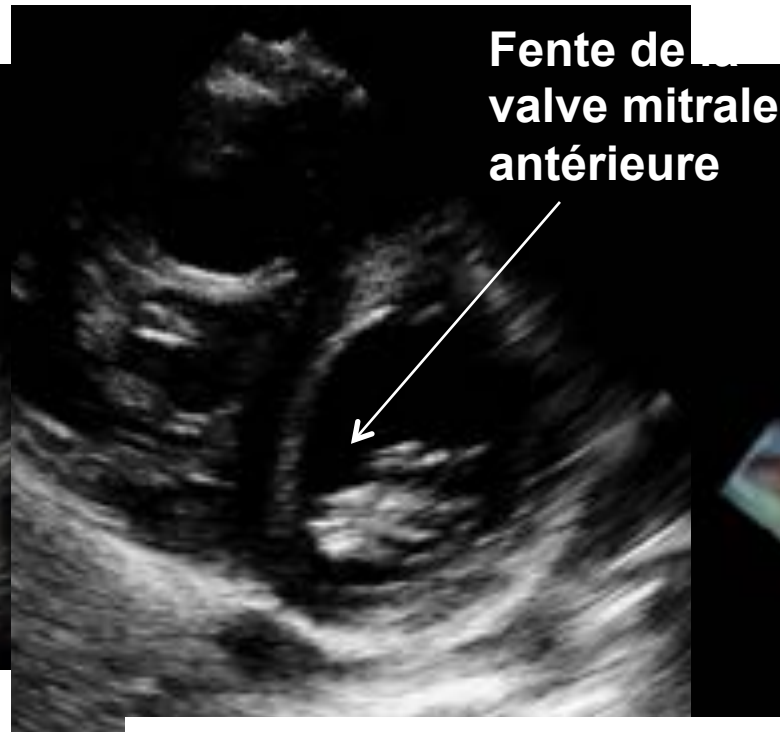
Age admission	H0	J7
Choc	0	15
Altération du VG	0	18
Canal large	8	6
Mortalité néonatale	0	3

Fente mitrale isolée

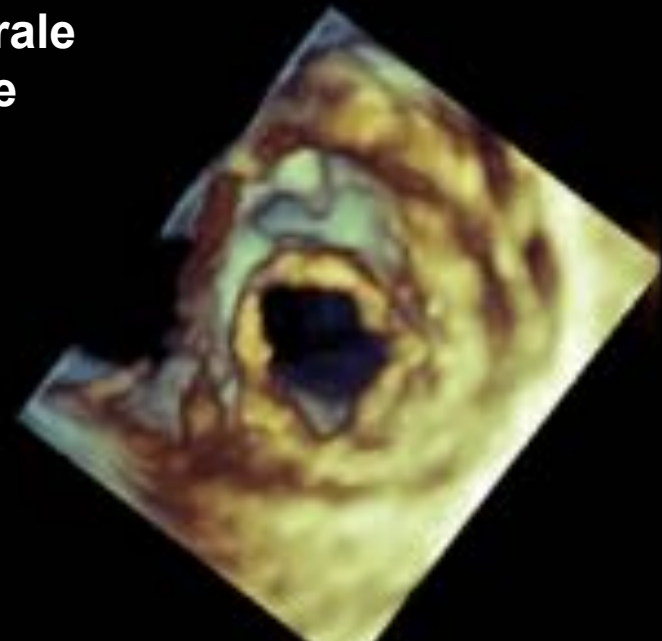
- Fente de la valve mitrale antérieure le plus souvent +++
- Muscles papillaires normo-posés
- Fente dirigée vers la chambre de chasse du VG
- **IM** : jet excentré, dirigé vers la paroi latérale de l'OG



4 cavités



PSPA



3D vue

HypoVG? Cas borderline

- Is left-sided structures are adequate to sustain the systemic circulation ?
- In very borderline situations, the hybrid approach might stabilize the circulation and help to select proper treatment plan later.
- In the majority of these cases, a single ventricular palliation may considered
 - Mitral valve annulus diameter is <7 mm in a neonate, or < -2 z-score
 - Hypoplasia of the aortic annulus (<5 mm),
 - Predominantly retrograde flow in the ascending aorta
 - Ratio of left and right ventricular lengths <0.8,
 - Cardiac apex not formed by the left ventricle,
 - Presence of endocardial fibroelastosis,

Coarctation

