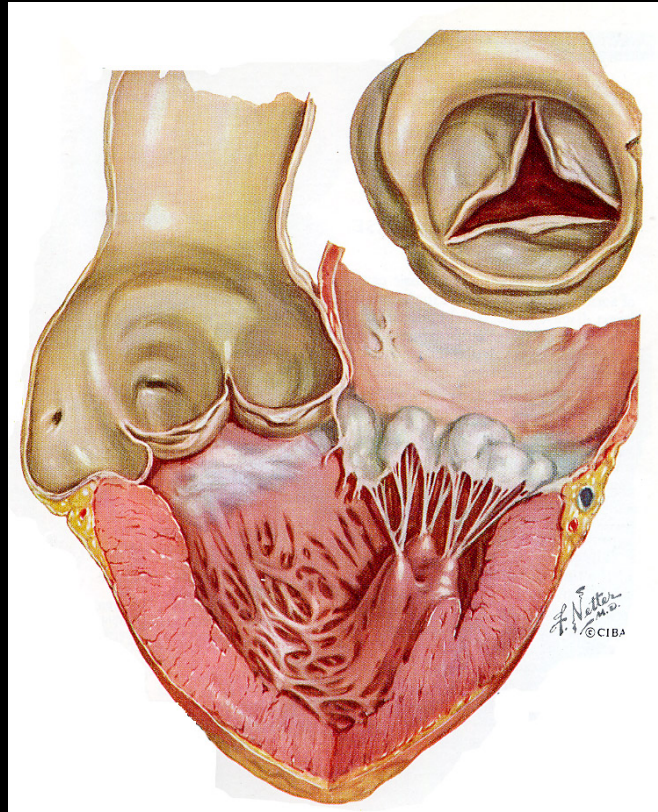


Maladies Aortiques



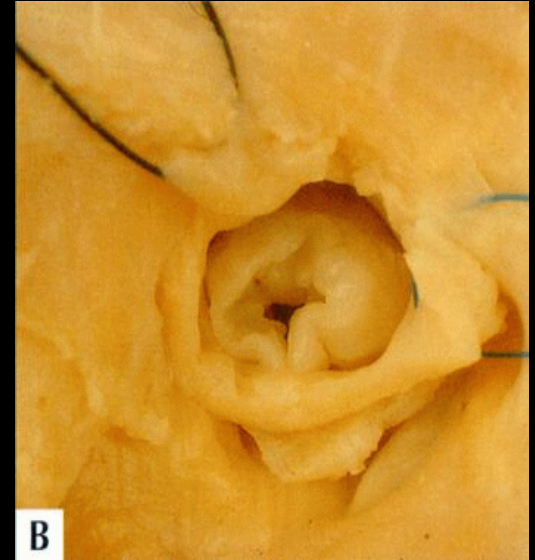
Ambroise BOUCHARD DE LA POTERIE / Xavier IRIART
Service des cardiopathies congénitales de l'enfant et de l'adulte
CHU de Bordeaux

Plan

- **RA néonatal**
- Sténose sous valvulaire aortique
- RA de l'adulte
- IA

RAo néonatal

- Valve pathologique : uni, bi ou tricuspide
- + souvent bicuspidie
- Bicuspidie la plus fréquente est la type 1 LR



main category: number of raphes	0 raphe - Type 0		1 raphe - Type 1			2 raphes - Type 2
	21 (7)		269 (88)			14 (5)
1. subcategory: 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)
2. subcategory:						
V F I	6 (2)	1 (0.3)	79 (26)	22 (7)	3 (1)	6 (2)
A U S	7 (2)	5 (2)	119 (39)	15 (5)	3 (1)	6 (2)
L N B (I + S)		1 (0.3)	15 (5)	7 (2)	2 (1)	2 (1)
V C No			3 (1)	1 (0.3)		
U L I						
A O N						
R N						

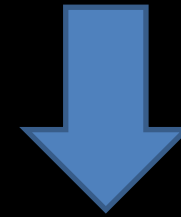
Sténose aortique sévère du fœtus

Evolution vers HLHS

Sténose aortique



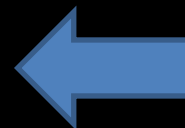
Ischémie sous endocardique
Dysfonction VG / HVG
Baisse compliance VG



Elevation PTDVG et POG



Shunt G>D par FOP



↓ Remplissage VG



Baisse croissance VG



HLHS :

hypoplastic left heart syndrome

Evaluation d'une voie aortique

- La valve mitrale
 - Anneau
 - Membrane supra mitrale*
 - Morphologie mitrale (parachute)*
 - Tissu accessoire
- Taille du VG, hypertrophie VG
- Voie sous aortique*
- Valve (uni, bi, tricuspide)
- Anneau aortique
- Crosse aortique*

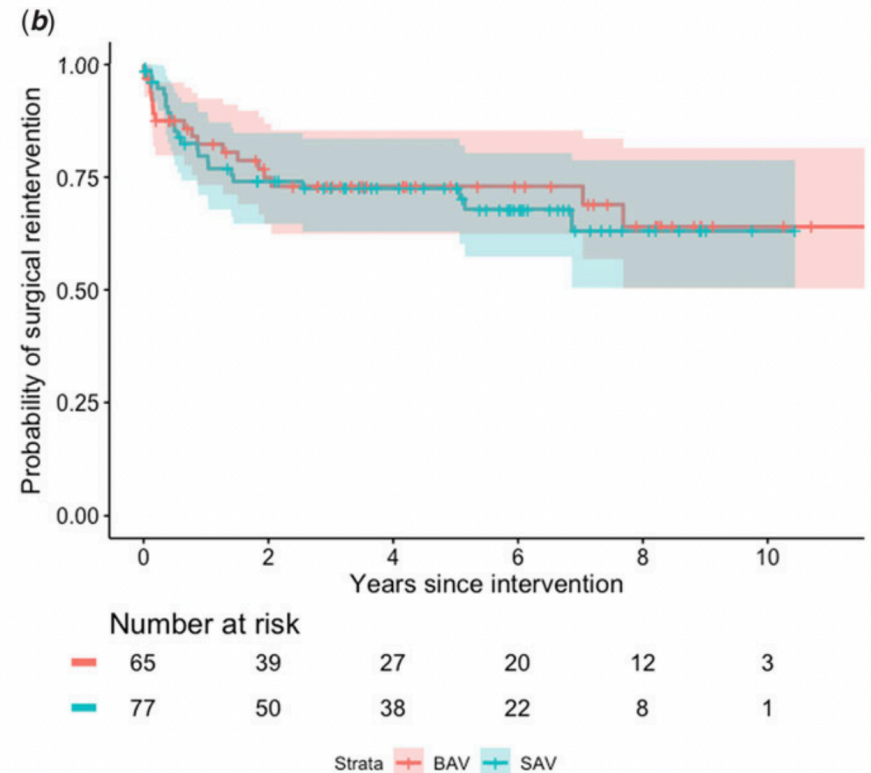
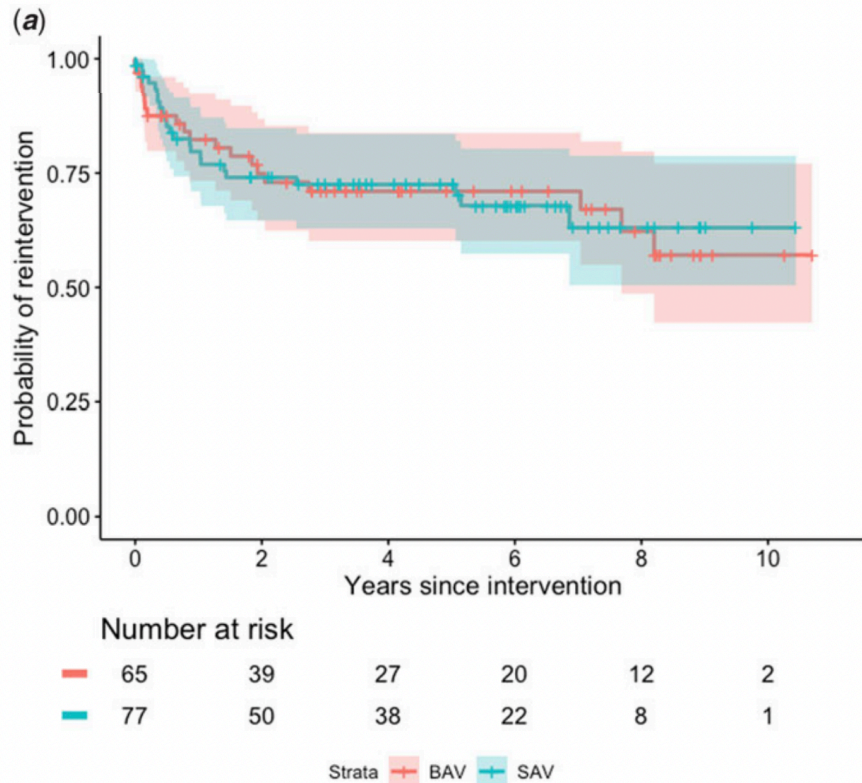


2*/4 = Syndrome de shone

Stratégie thérapeutique et timing

- Indications d'intervention :
 - Sténose valvulaire aortique critique : si ductodépendance
 - Dysfonction VG
 - Ischémie
 - Syncope
 - Retentissement staturo-pondéral
- Stratégie à 1 ou 2 ventricules ?
- Si stratégie à 2 ventricules : chirurgie ou angioplastie

Cathétérisme (BAV) VS Chirurgie (SAV)



Auld et al, Balloon dilatation versus surgical valvotomy for congenital aortic stenosis: a propensity score matched study, *Cardiol Youth*, 2021

Valvuloplastie au ballon

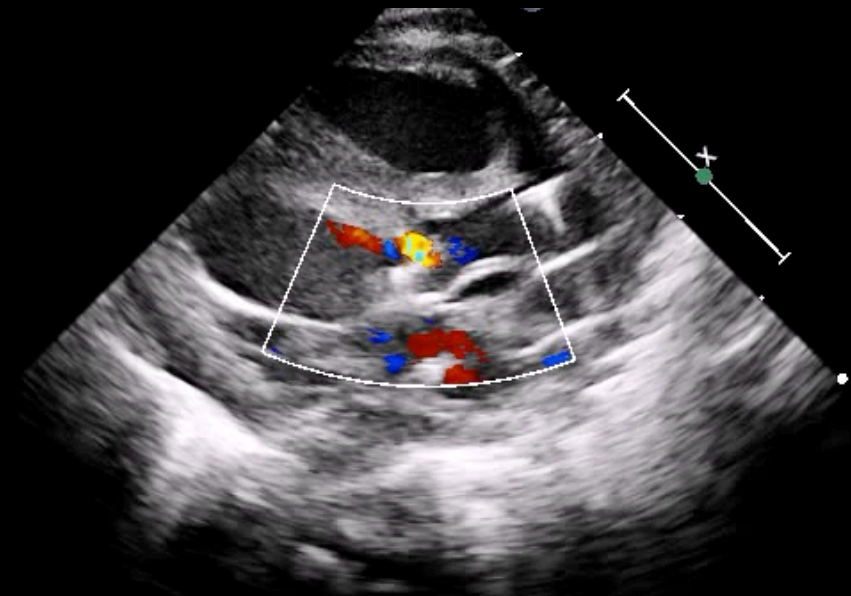
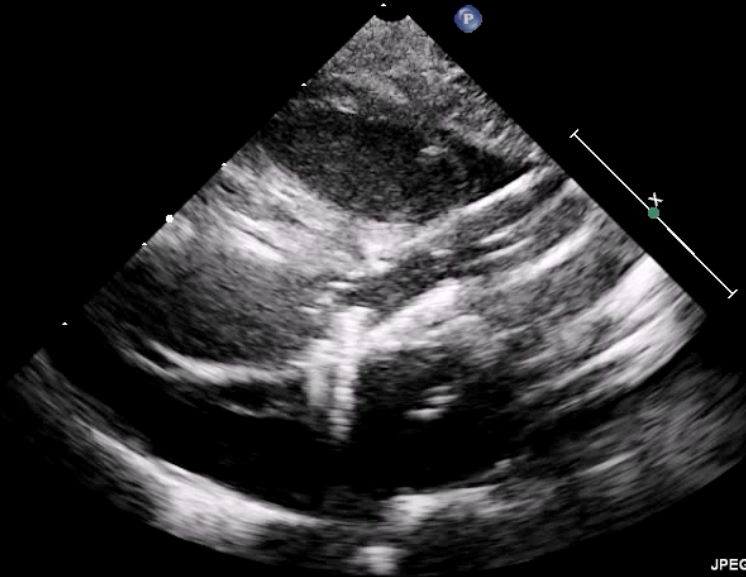
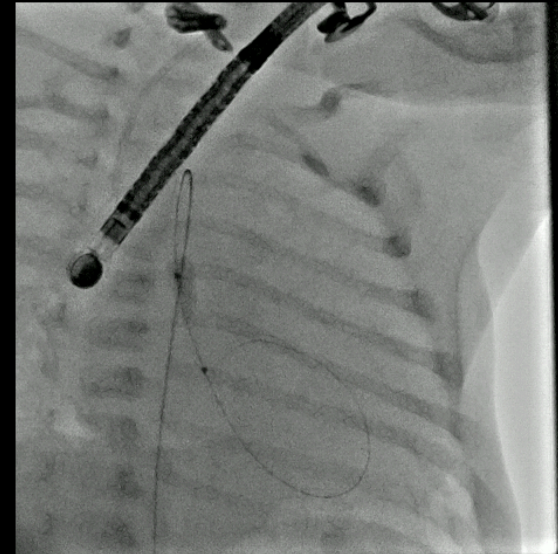
Recommendations for Aortic Valvuloplasty

Class I

1. Aortic valvuloplasty is indicated regardless of valve gradient in the newborn with isolated critical valvar AS who is ductal dependent or in children with isolated valvar AS who have depressed left ventricular systolic function (*Level of Evidence: B*).
2. Aortic valvuloplasty is indicated in children with isolated valvar AS who have a resting peak systolic valve gradient (by catheter) of ≥ 50 mm Hg[†] (*Level of Evidence: B*).
3. Aortic valvuloplasty is indicated in children with isolated valvar AS who have a resting peak systolic valve gradient (by catheter) of ≥ 40 mm Hg[†] if there are symptoms of angina or syncope or ischemic ST-T-wave changes on electrocardiography at rest or with exercise (*Level of Evidence: C*).

Valvuloplastie au ballon

- Monitoring ETT ou ETO
- Diamètre du ballon : $0.8 \times$ diamètre anneau aortique
- Arrêt si IA grade 2

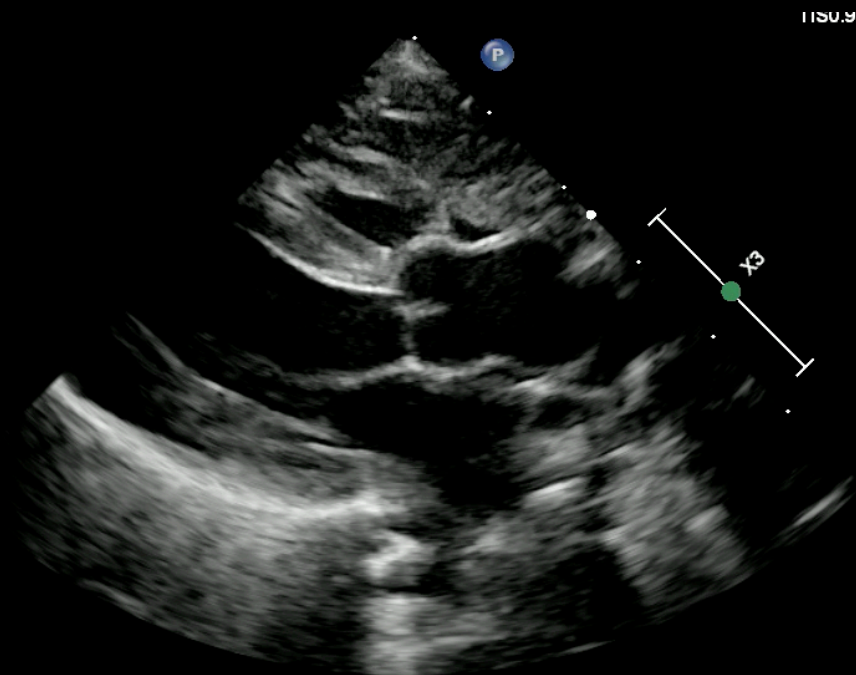
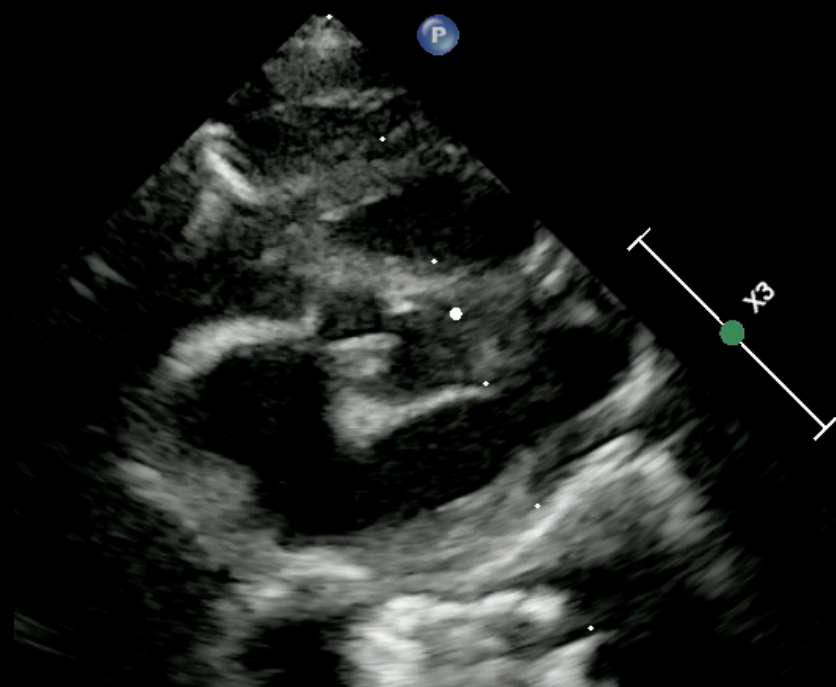


Cas n°1

- Evolution vers Rao serré à 3 mois.
- Poids lors du KT 2.8 kg

- ETT : Rétrécissement aortique serré (Gmoy 42mmHg). HVG modérée.

ETT / ETO pré-procédure



Angiographie (OAG/crânial et profil) : valve aortique épaissie, anneau 7.5 mm.

Guidance de la procédure par ETO : 7mm

Attitude :

Dilatation au ballon Tyshak 6x20mm jusqu'à disparition de l'encoche.

Contrôle :

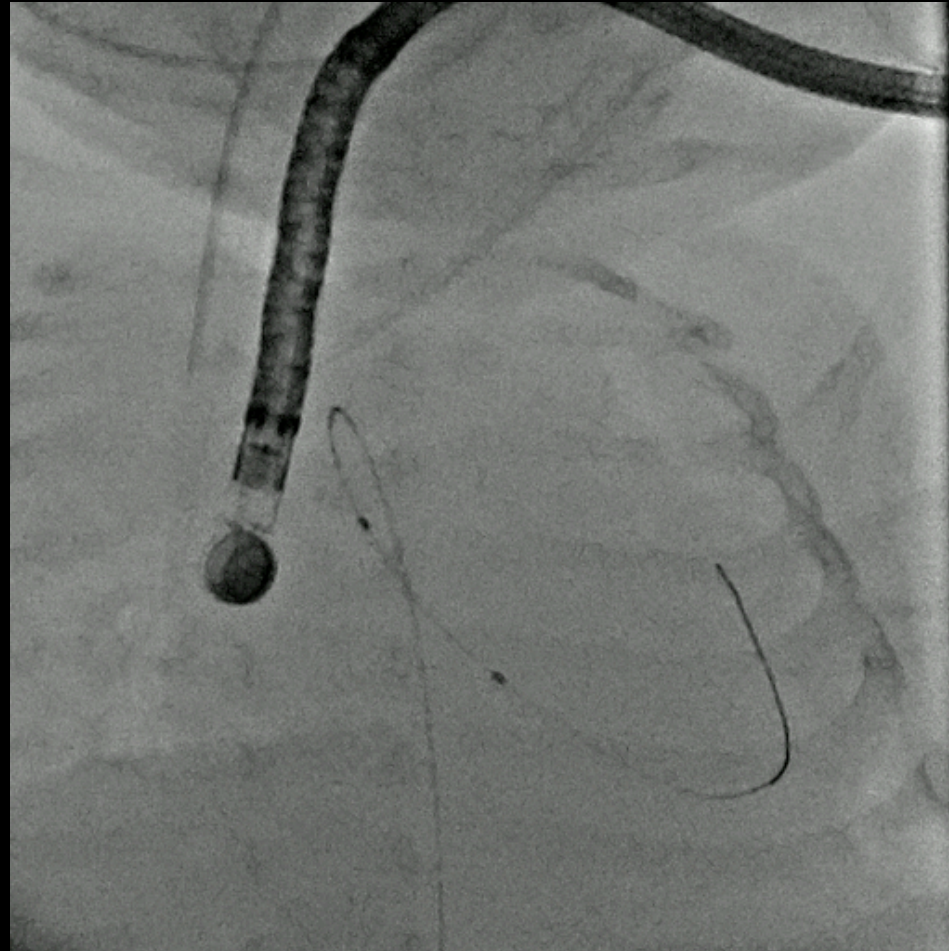
Pressions : VG=71/9, Ao=63/30/42

Evaluation échographique : Pas de fuite aortique

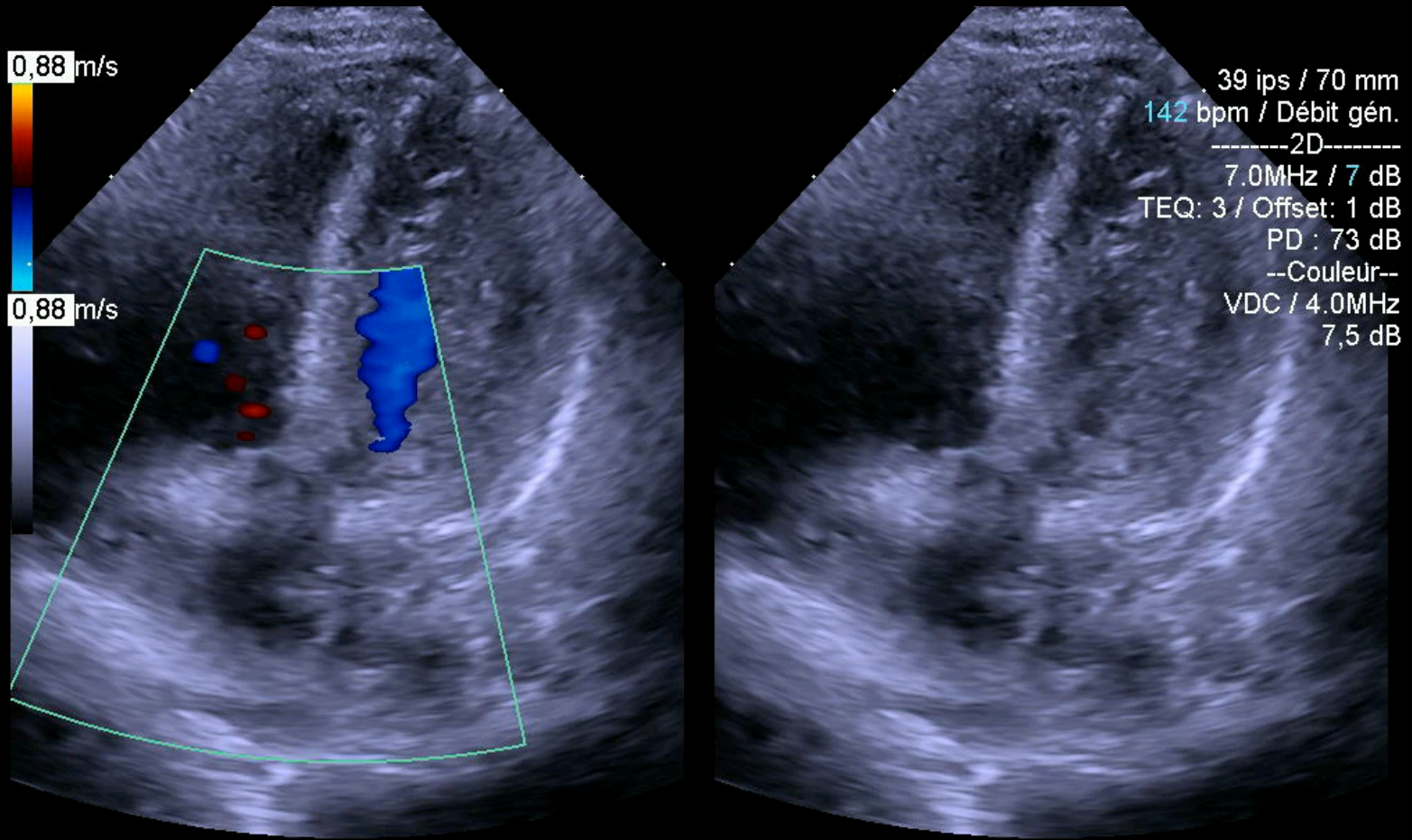
Conclusion :

Sténose valvulaire aortique serrée dilatée au ballon Tyshak 6x20 mm.

Bon résultat hémodynamique .

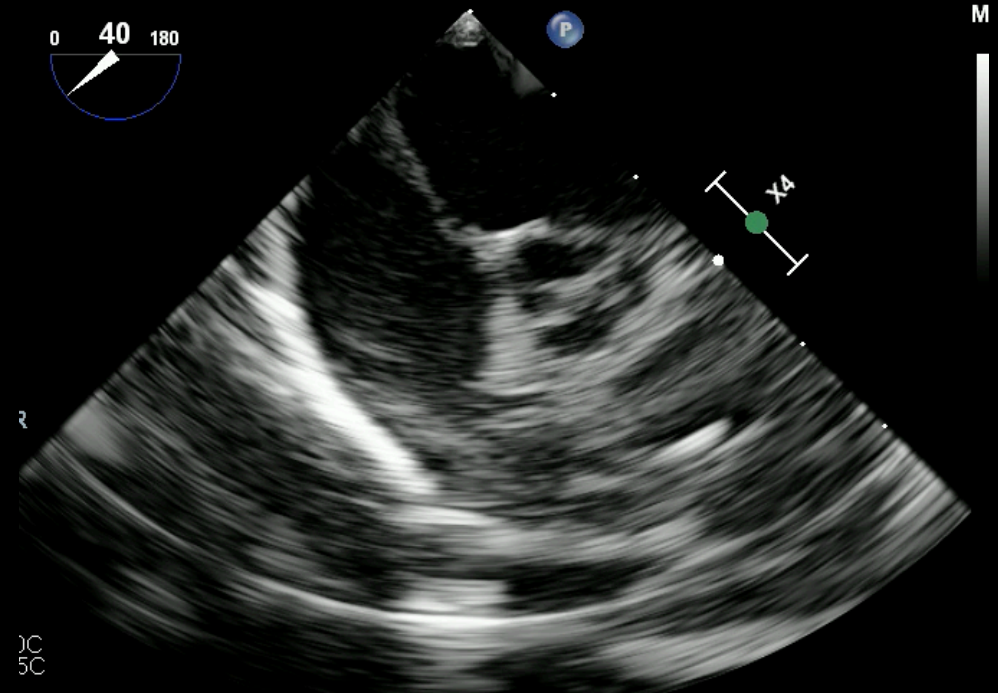
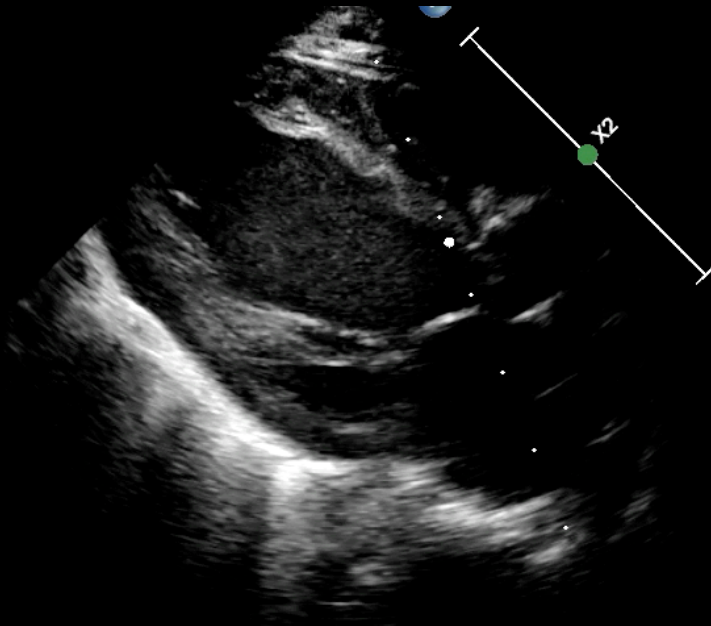
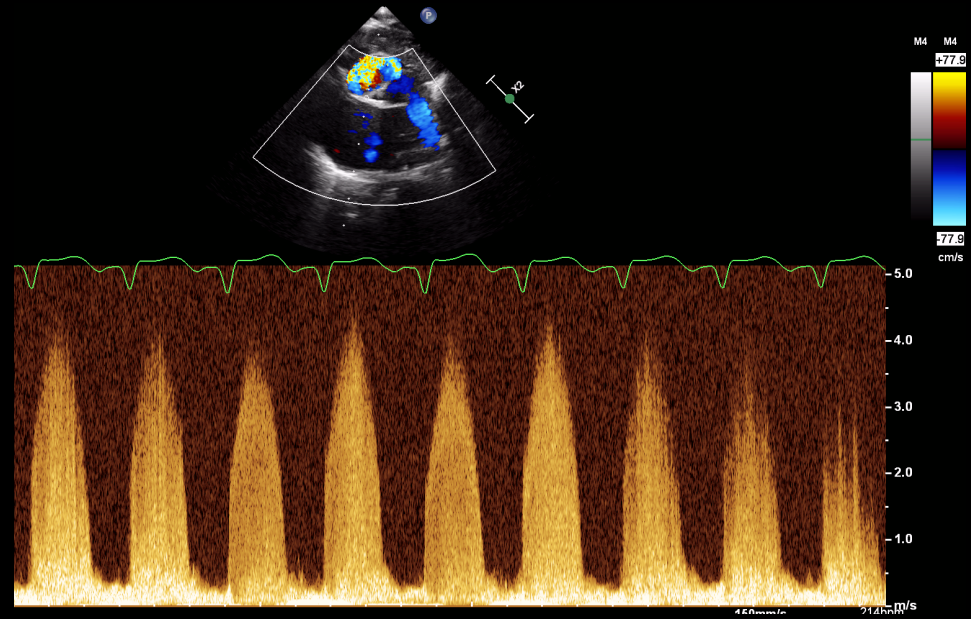


ETT / ETO post procédure



Cas n°2

- Diagnostique post natal :
stagnation pondérale + souffle + IC
- Poids 3.7 kg
- Bicuspédie type 2 (ETO)
- VG dilaté +4 Z-score avec FEVG 30%
- Vmax 5 m/s, GMoyen 70 mmHg



Cathétérisme

Angiographie (OAD/ profil) : valve aortique épaissie, anneau 8.5mm.

Attitude : Dilatation au ballon Tyshak 8x20 mm jusqu'à disparition de l'encoche.

Contrôle ETO: persistance d'une sténose serrée ($V_{max}=4m/s$) sans fuite

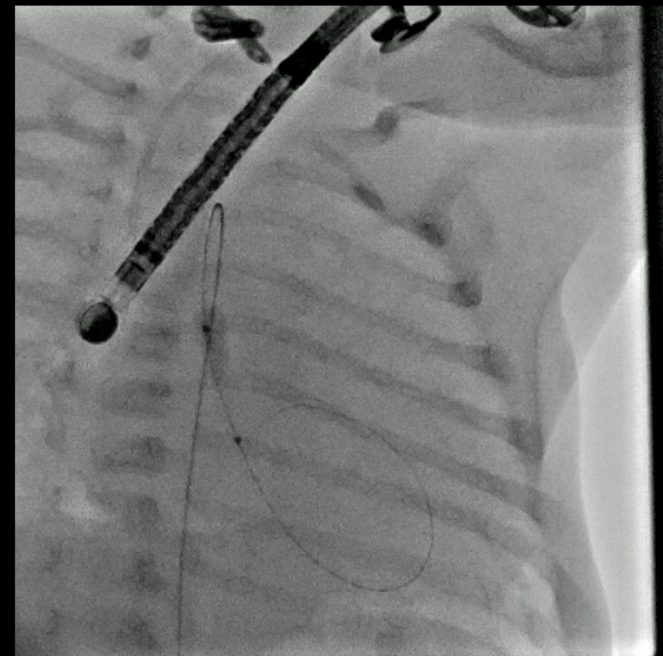
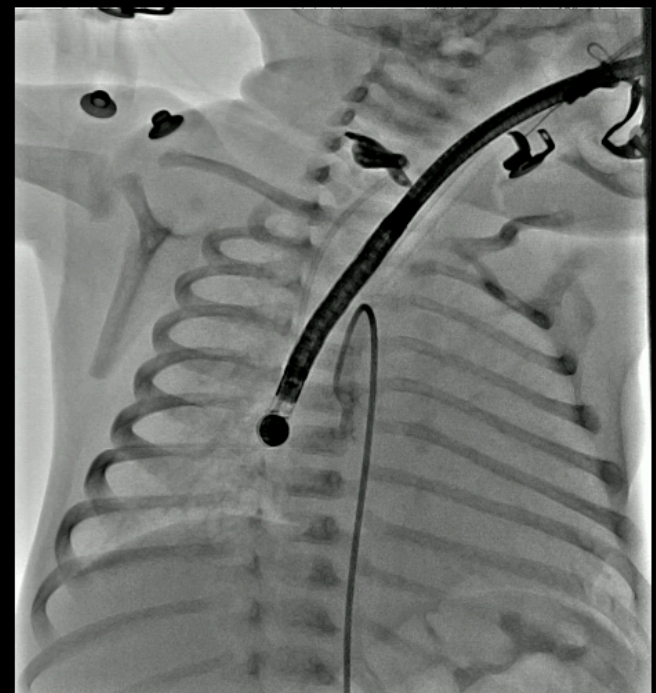
Dilatation au ballon Tyshak 9x20 mm jusqu'à disparition de l'encoche.

Contrôle ETO: bon résultat sur le gradient ($V_{max}=2.7m/s$), apparition d'une fuite sévère.

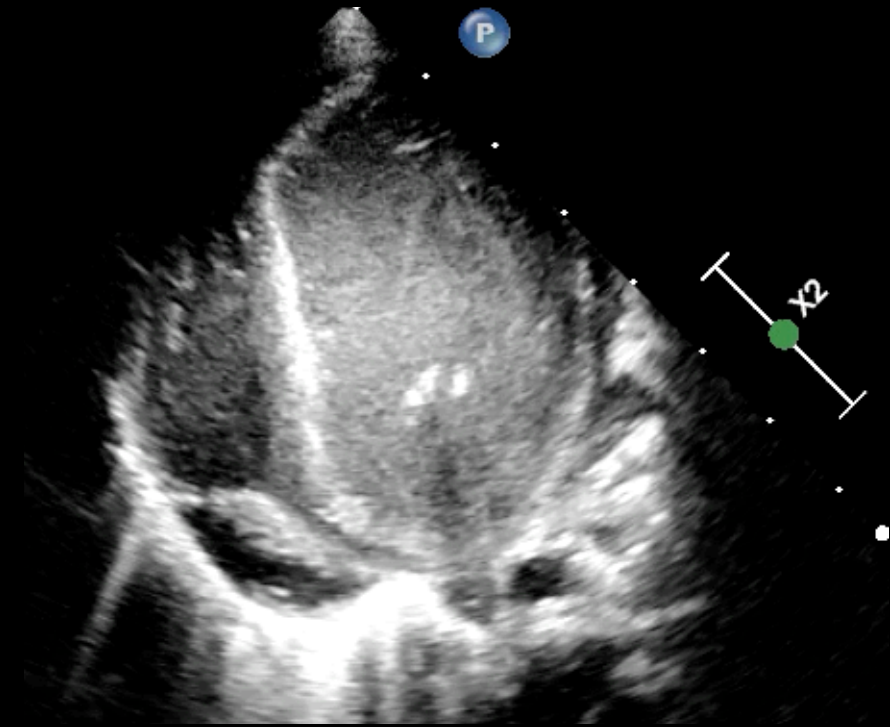
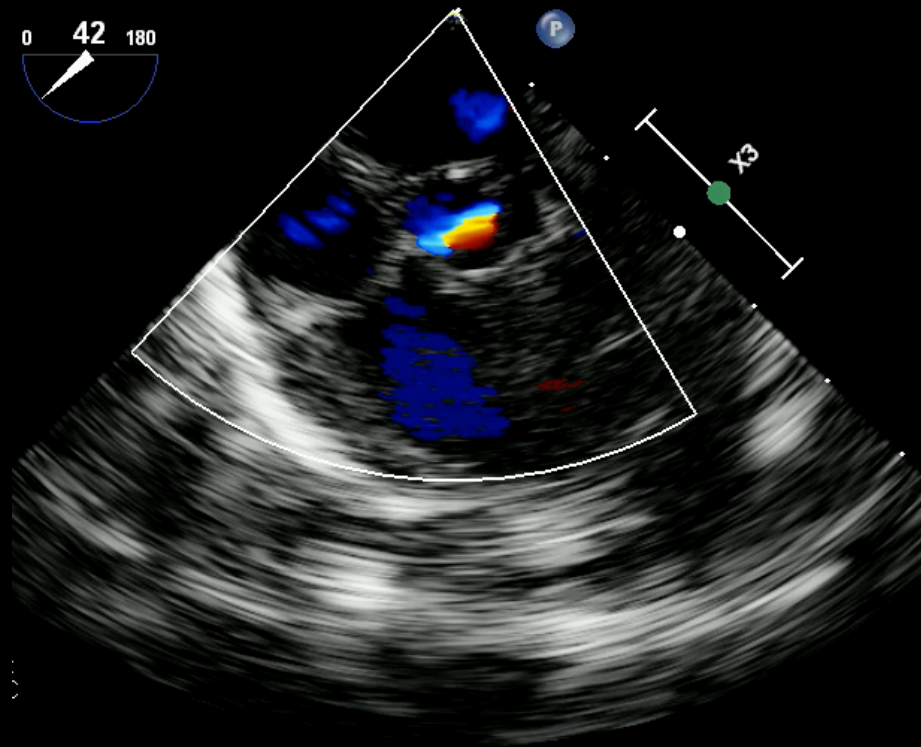
Conclusion :

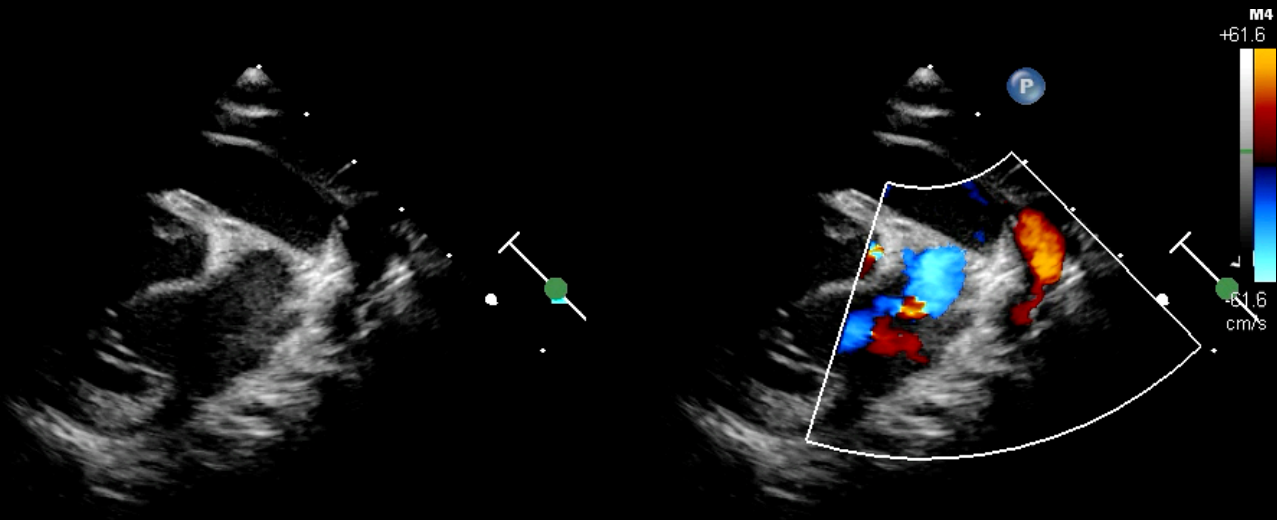
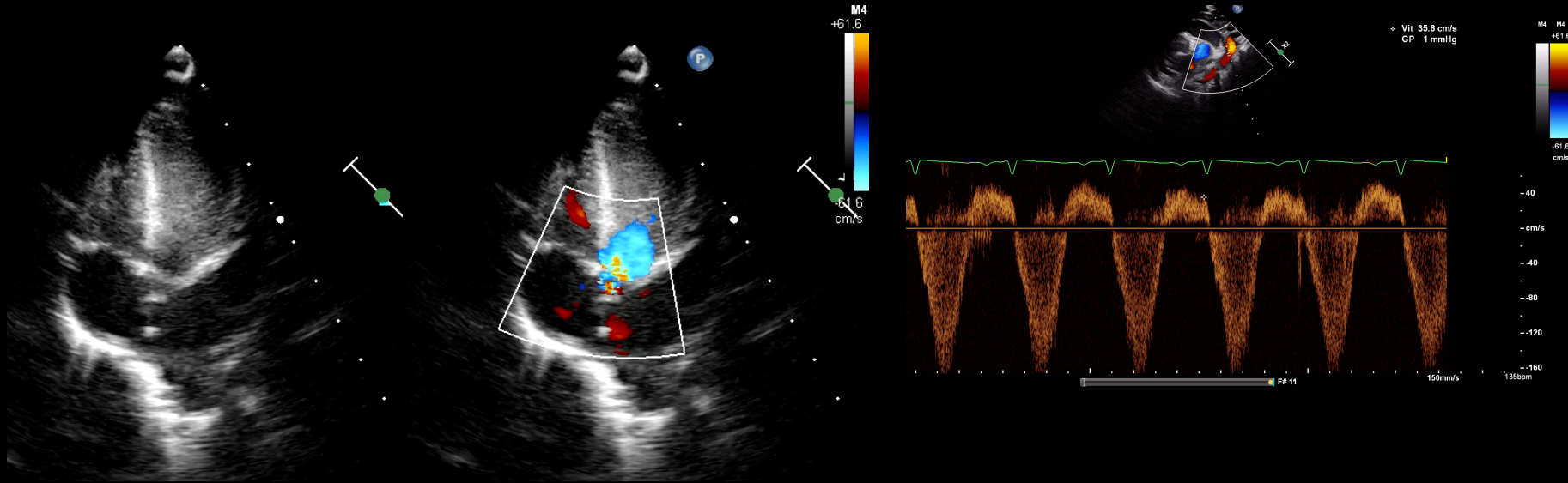
Sténose valvulaire aortique serrée dilatée au ballon Tyshak 8mm puis, devant une absence d'amélioration hémodynamique, utilisation d'un ballon de 9 mm

Bon résultat sur le gradient transvalvulaire, mais apparition d'une fuite aortique sévère.



ETT / ETO post procédure

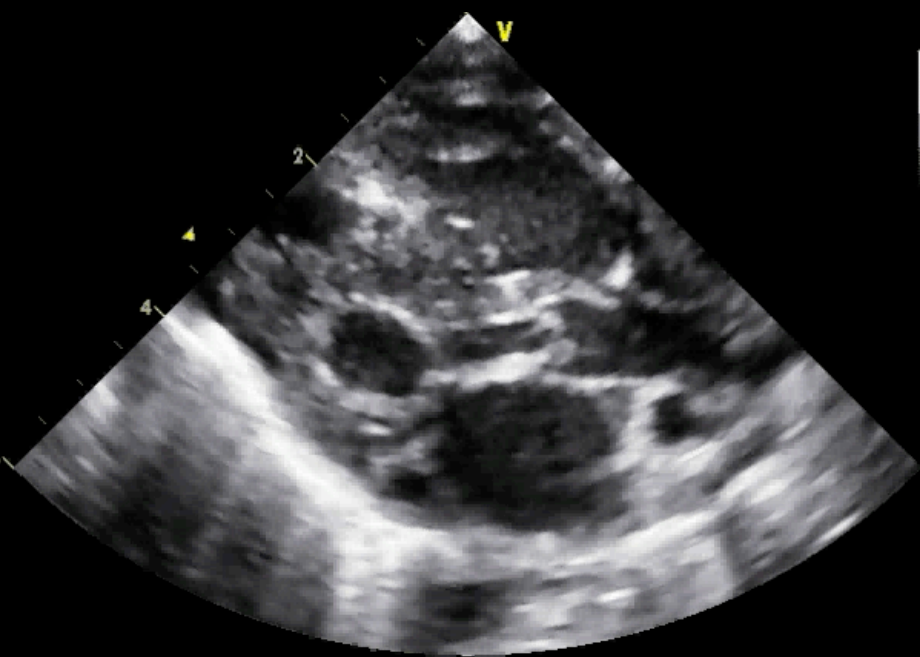




Après discussion en staff,
 décision de prise en charge par
 Ross néonatal.
 Chirurgie de Ross et
 reconstruction de la voie
 pulmonaire par un tube valvé
 Contegra n°12, par les Pr
 LEOBON et ROUBERTIE.

Stratégie à 1 ou 2 ventricules ?

Sténose aortique critique avec VG borderline



Hypoplasie du cœur gauche ou équivalent

- Sténose aortique critique avec désadaptation anténatale du VG = évolution possible mais non systématique vers l'hypoplasie du cœur gauche (HLHS)
- Formes frontières avec dysfonction moyenne du VG et taille limite : difficulté d'appréciation du potentiel évolutif (hypo VG vs circulation biventriculaire)
- Critères de Rhodes+++

Critical aortic stenosis

ECHO criteria for biventricular repair

LV / RV length	> 0.8
LV – AO junction	> 3.5 cm/m ²
MV area	> 4.75 cm ² /m ²
LV mass	> 35 g/m ²

Rhodes LA: Circulation 1991;84:2325-35

LV length	> 25mm
LV – AO junction	> 5mm
MV orifice	> 9mm
LV vol.	> 20 ml/m ²

Leung MP: J Thorac Cardiovasc Surg 1991;101:526-35

Rhodes discriminant score for survival with intended BiV circulation

- **Rhodes 1991** retrospective analysis of 65 pts (AS < 60d.)

95% correctly selected for biventricular circulation

Rhodes LA, Circulation 1991

- **Colan 2006** retrospective analysis of 126 pts

77% correctly selected for biventricular circulation

Colan SD, JACC 2006

Revised Rhodes formula

Changes: MiV excluded, EFE inserted, Z-scores applied

Score: $10.98 \times \text{BSA} + 0.56 \times \text{AoV annulus z-score} + 5.89 \times \text{Long axis ratio} - 0.79$
presence of grade 2 or 3 EFE $- 6.78$

Discriminate score:

< - 0.65 univentricular circulation

> - 0.65 biventricular circulation

• With old formula
77% correctly selected
for
biventricular circulation

• With new formula
90% correctly selected
for biventricular
circulation

CRITICAL AORTIC STENOSIS IN THE NEONATE: A MULTI-INSTITUTIONAL STUDY OF MANAGEMENT, OUTCOMES, AND RISK FACTORS

Initially intended BiV (116) - 5yr survival rate 70%

Initially UNiV (179) - 5yr survival rate 60%

Independent risk factors associated with greater survival benefit for UniV versus BiV:

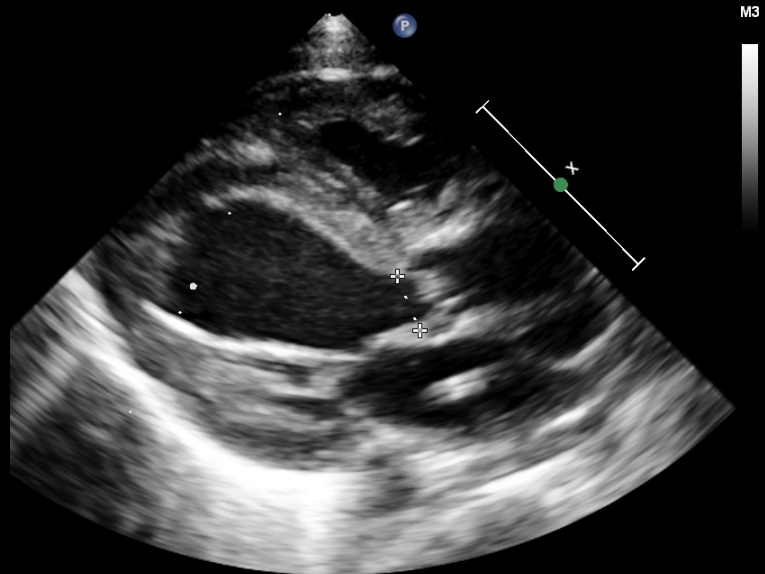
- Young age
- Low aortic valve Z-score
- Low LV length
- High Endocardial fibroelastosis
- No Tricuspid regurgitation

Multivariable hazard model

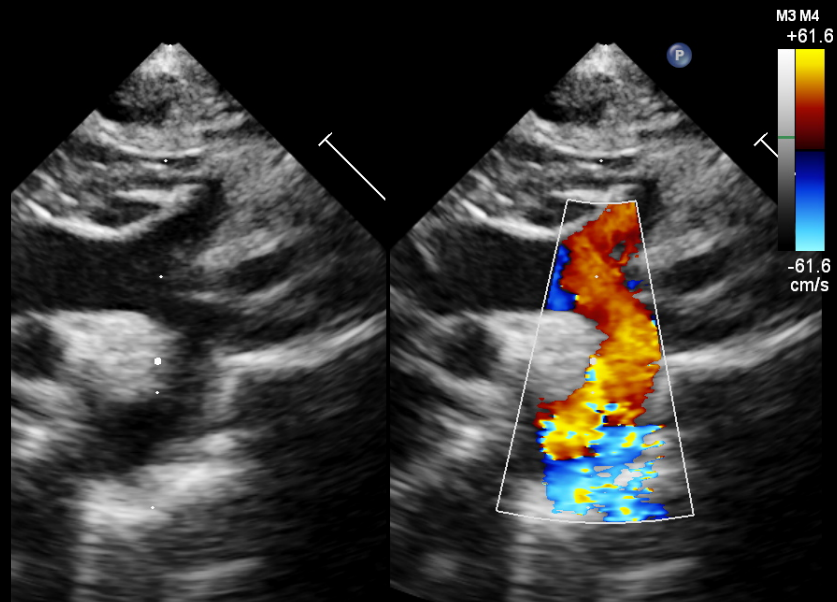
Small left ventricle: criteria used for routine clinical practise

- **Aortic valve annulus > 5.0 mm**
- **Mitral valve annulus > 9 mm**
(absence of severe morphological abnormalities)
- **Absence of EFE**
- **Left ventricle/heart long axis >0.8 in 4-CH**
- **Absence of retrograde flow in distal aortic arch**

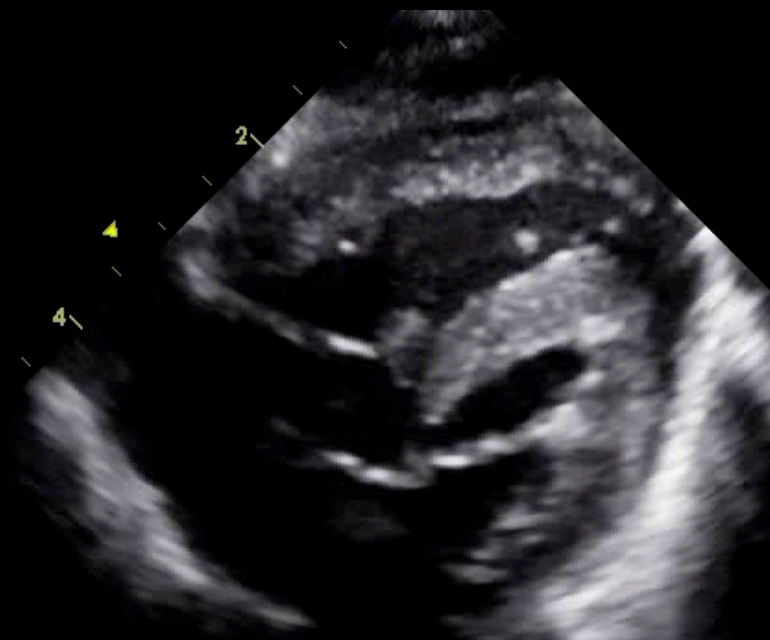
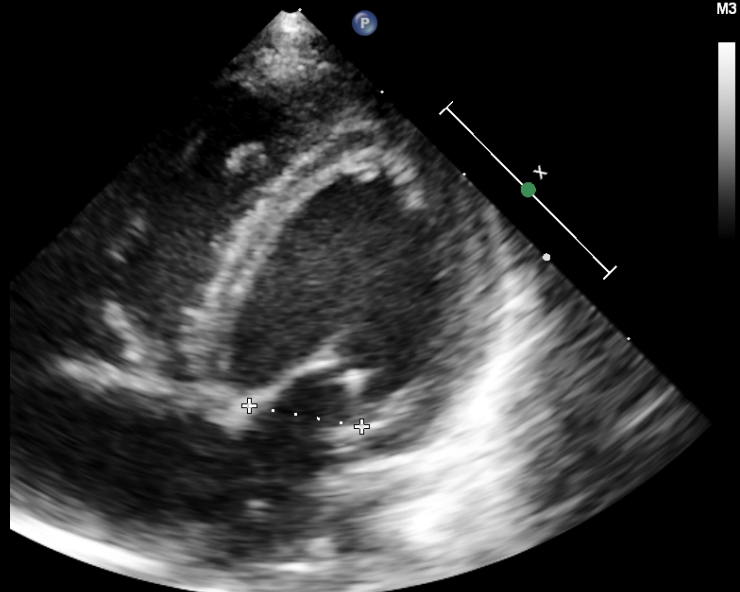
ITm1.2 IM 0.7



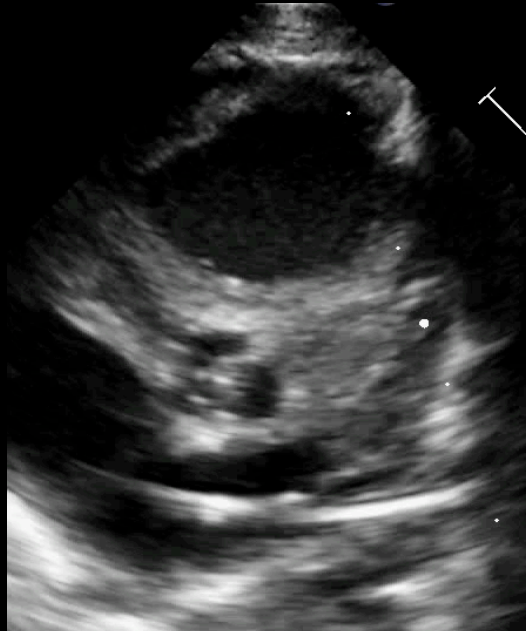
ITm0.8 IM 0.8



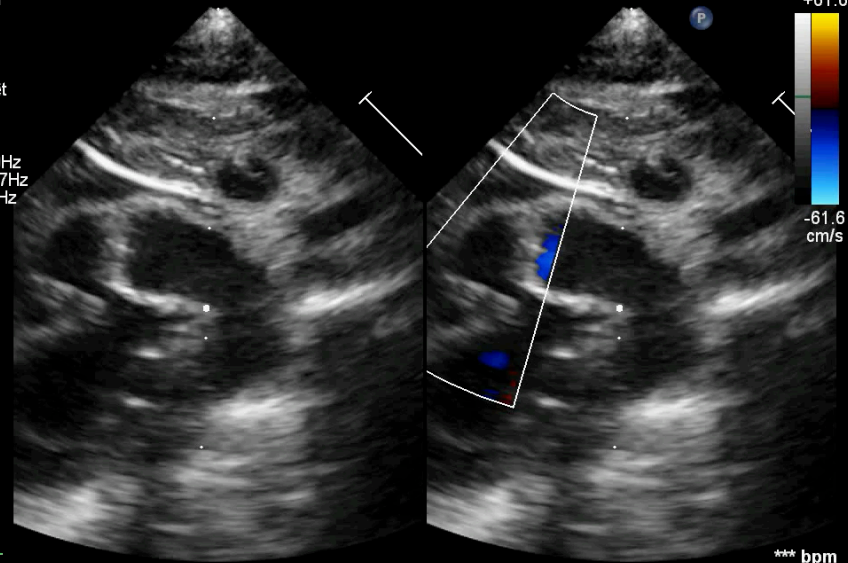
M3



Sténose aortique + Fibroélastose



S12-4
46Hz
5.0cm
2D
82%
C 52
P Arrêt
Gén
Coul
67%
7200Hz
FP 647Hz
4.5MHz

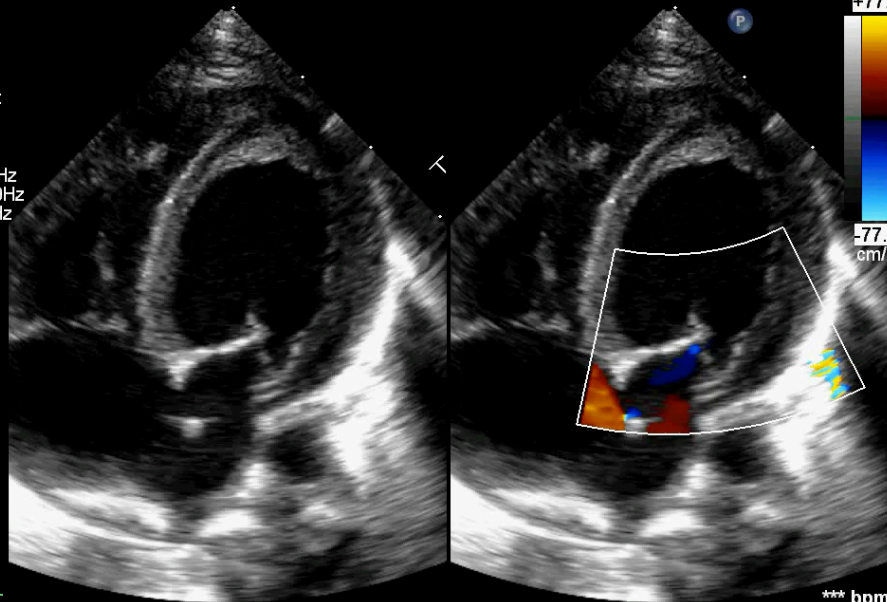


M3 M4
+61.6
-61.6
cm/s

*** bpm

26Hz
6.0cm

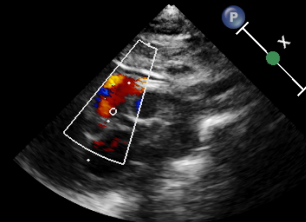
2D
49%
C 50
P Arrêt
Rés
Coul
40%
6600Hz
FP 659Hz
3.3MHz



M4 M4
+77.0
-77.0
cm/s

*** bpm

Pediatric
S12-4
46Hz
5.0cm
2D
81%
C 52
P Arrêt
Gén
Coul
67%
7200Hz
FP 647Hz
4.5MHz

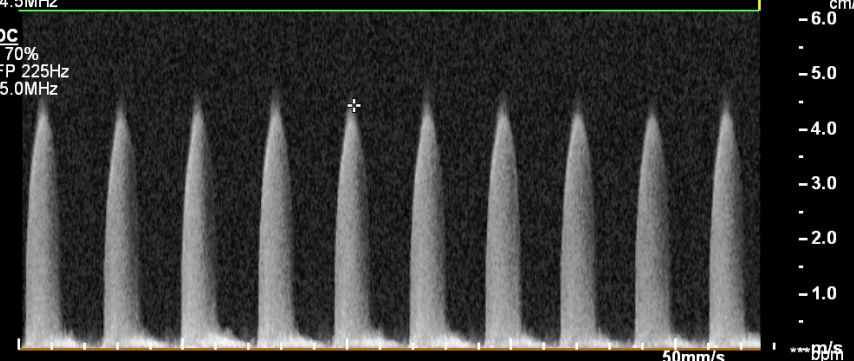


11 mu.3 1M 0.0

+ Vit 444 cm/s
GP 79 mmHg

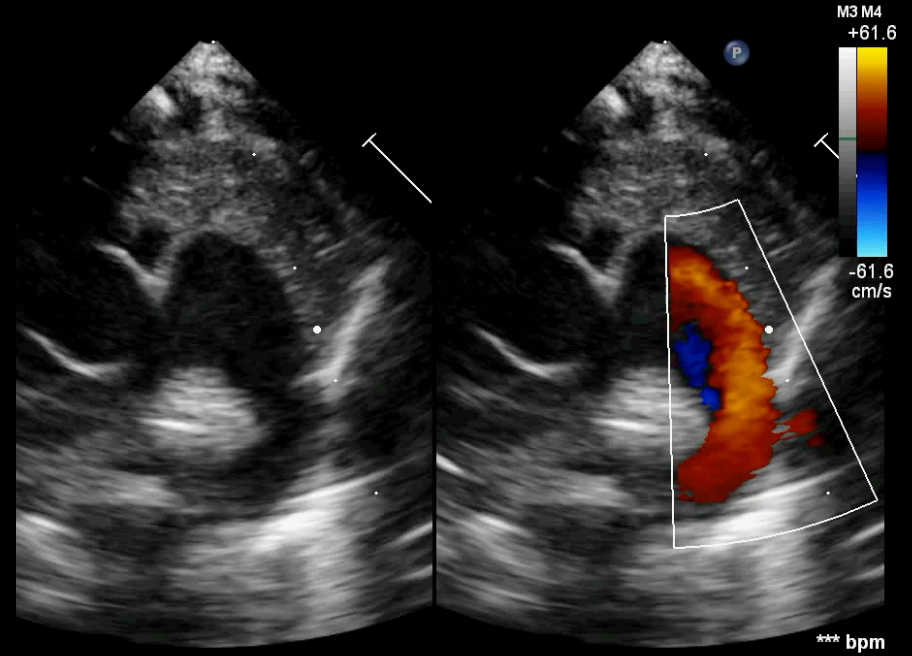
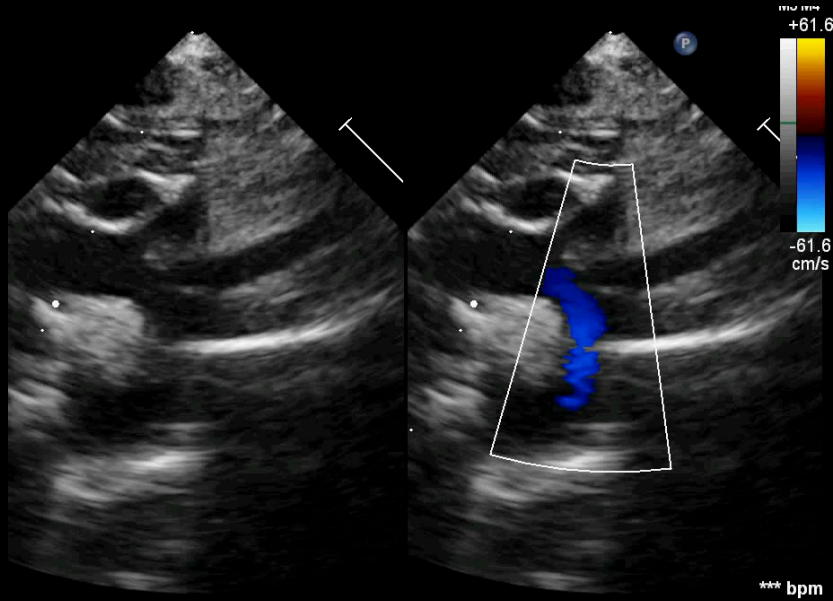
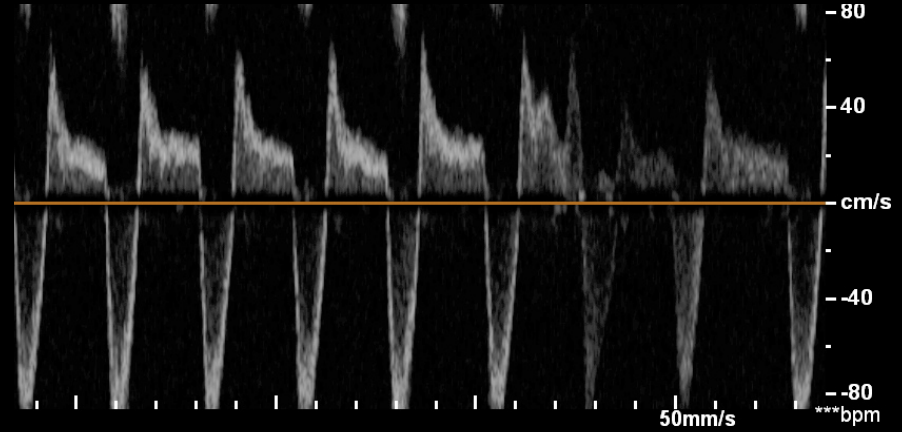
M3 M4
+61.6
-61.6
cm/s

DC
70%
FP 225Hz
5.0MHz



*** bpm

RA critique ductodépendant



Plan

- RA néonatal
- **Sténose sous valvulaire aortique**
- RA de l'adulte
- IA

Membrane sous aortique

- Membrane située sous anneau aortique
- Favorisée par angulation septo-aortique aigue avec shear stress

Evolution :

- Enfant : tendance à progresser
- Adulte : gradient évolue assez peu

Retentissement : HVG, IA (lésion de jet avec défaut de coaptation par restriction d'une sigmoïde)

Membrane sous aortique : Indications chirurgicales

Recommendations for intervention in subaortic stenosis

Recommendations	Class ^a	Level ^b
In symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient ≥ 40 mmHg ^c or severe AR, surgery is recommended.	I	C
Asymptomatic patients should be considered for surgery when one or more of the following findings are present: <ul style="list-style-type: none"> ● Mean gradient < 40 mmHg but LVEF $< 50\%$. ● AR is severe and LVESD > 50 mm (or 25 mm/m² BSA) and/or EF $< 50\%$^d. ● Mean Doppler gradient is ≥ 40 mmHg^c and marked LVH present. ● Mean Doppler gradient is ≥ 40 mmHg^c and there is a fall in blood pressure below baseline on exercise. 	IIa	C
Asymptomatic patients may be considered for surgery when one or more of the following findings are present: <ul style="list-style-type: none"> ● Mean Doppler gradient is ≥ 40 mmHg^c, LV is normal (EF $> 50\%$ and no LVH), exercise testing is normal, and surgical risk is low. ● Progression of AR is documented and AR becomes more than mild (to prevent further progression). 	IIb	C

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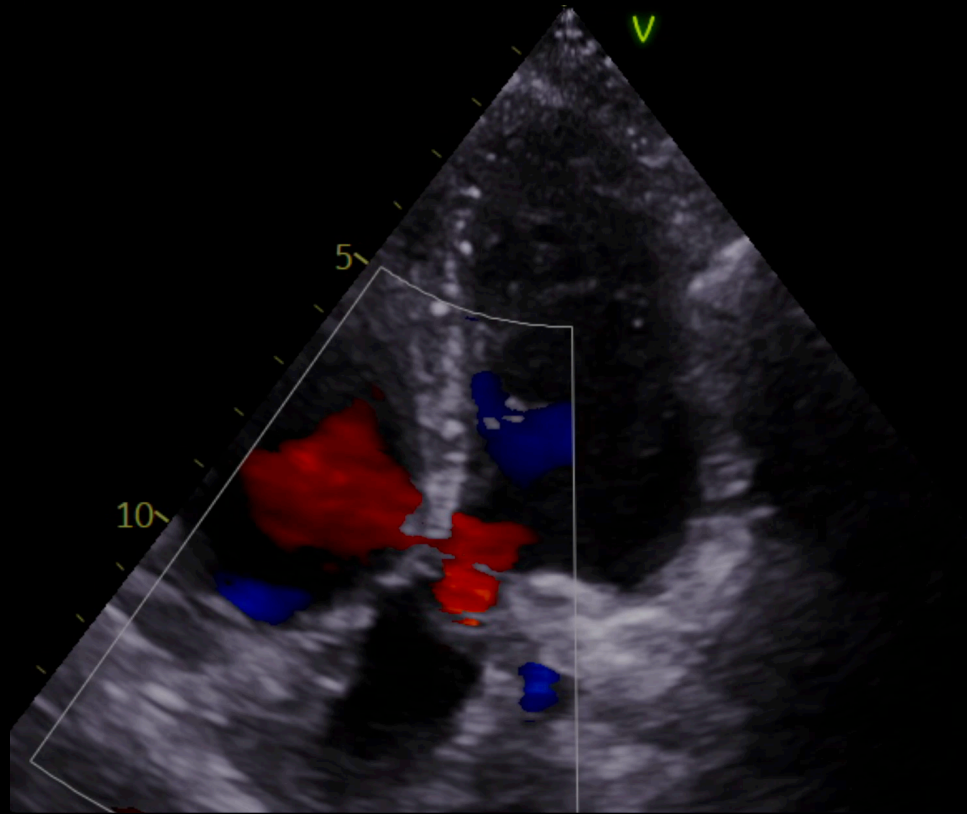
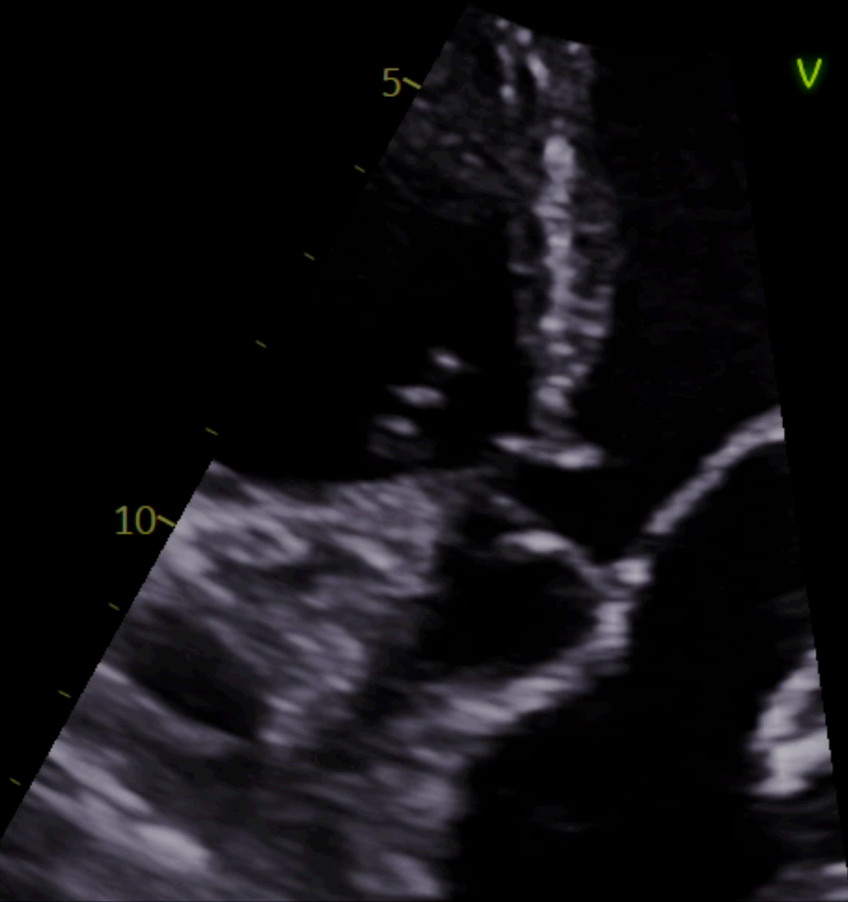
AR = aortic regurgitation; BSA = body surface area; EF = ejection fraction; ESC = European Society of Cardiology; LV = left ventricle/ventricular; LVEF = left ventricular ejection fraction; LVESD = left ventricular end systolic diameter; LVH = left ventricular hypertrophy.

^aClass of recommendation.

^bLevel of evidence.

^cDoppler-derived gradients may overestimate the obstruction and may need confirmation by cardiac catheterization.


^dSee 2017 ESC Guidelines on the management of valvular heart disease.²⁵



Plan

- RA néonatal
- Sténose sous valvulaire aortique
- **RA de l'adulte**
- IA

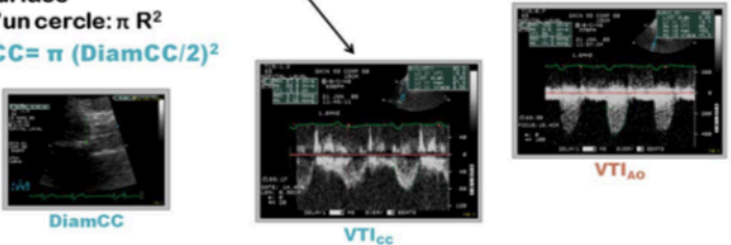
RA de l'adulte



$A_1 =$ Surface Chambre de Chasse: SCC
 $V_1 =$ Vitesse Chambre de Chasse: VTI_{CC}
 $A_2 =$ Aire Valvulaire Aortique: AVA
 $V_2 =$ Vitesse dans l'aorte: VTI_{Ao}

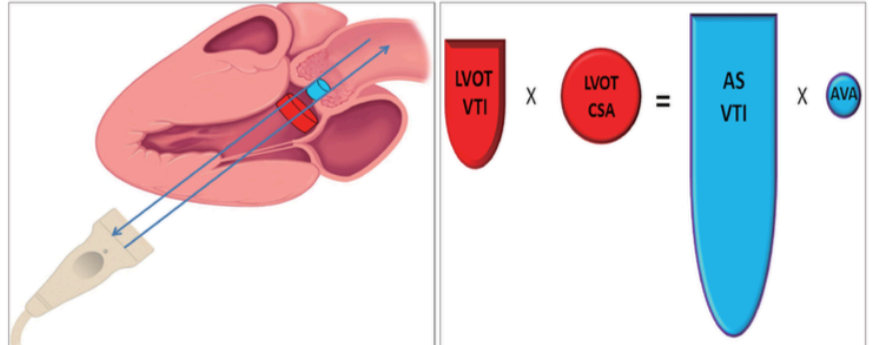
$$SCC \times VTI_{CC} = AVA \times VTI_{Ao}$$

Surface d'un cercle: πR^2
 $SCC = \pi (\text{DiamCC}/2)^2$



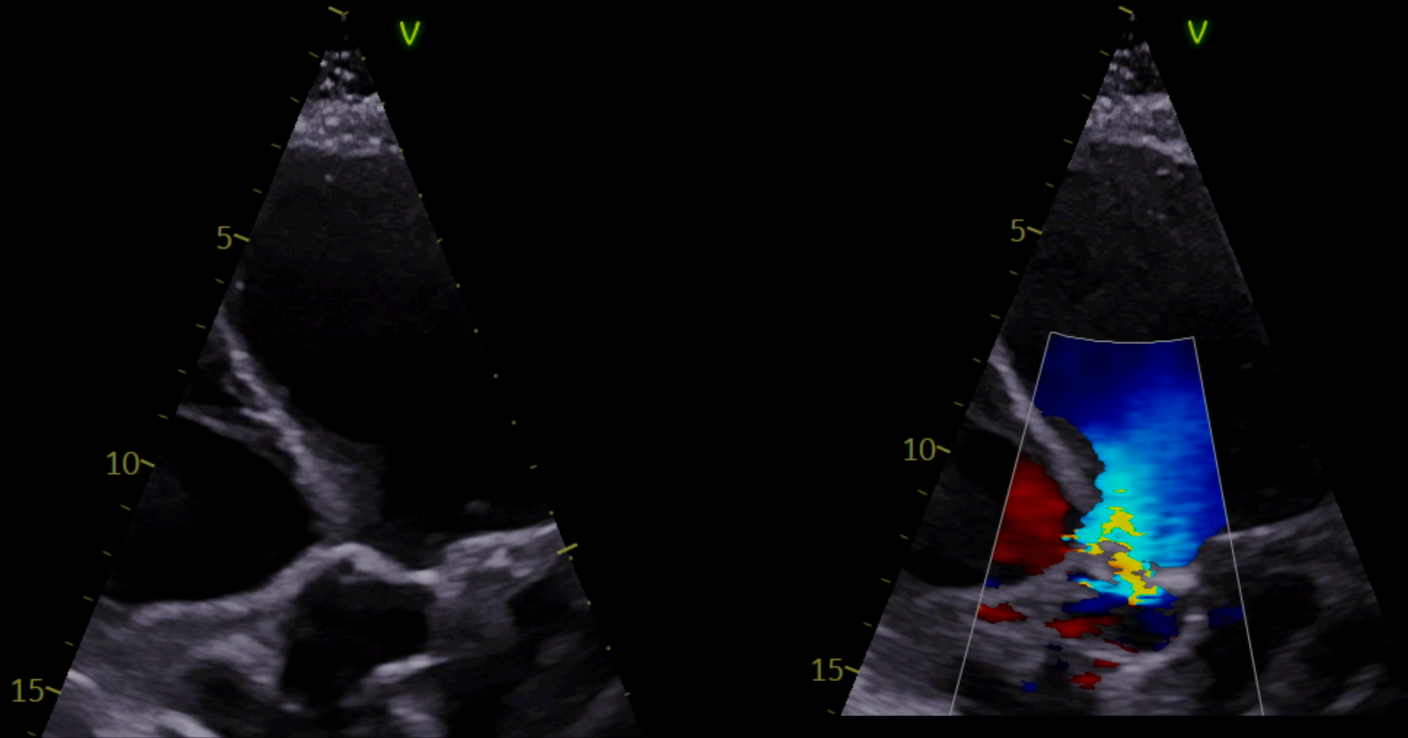
$AVA = \frac{\pi}{4} \times \text{DiamCC}^2 \times \frac{VTI_{CC}}{VTI_{Ao}}$

AVA < 1cm² = serré



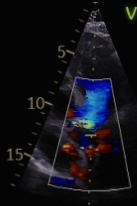
$LVOT \ VTI \times LVOT \ CSA = AS \ VTI \times AVA$

RA serré : évaluation échographique

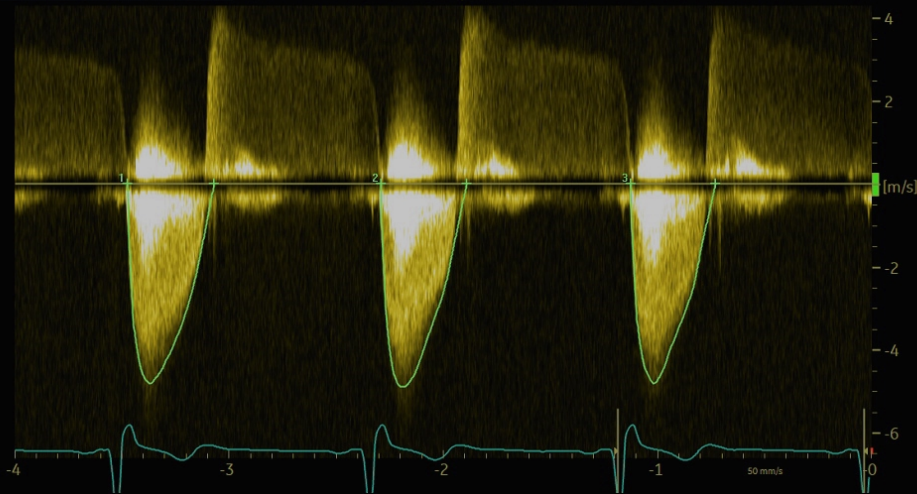


RA serré : évaluation échographique

Av	VA Vmax	4.83 m/s
	VA Vmoy	3.17 m/s
	VA GDmax	93.46 mmHg
	VA GDmoy	48.25 mmHg
	VA ITV	126.69 cm
	VA Durée	399.62 ms
	FC	51.55 BPM

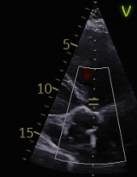


Spectrum Auto Recognized

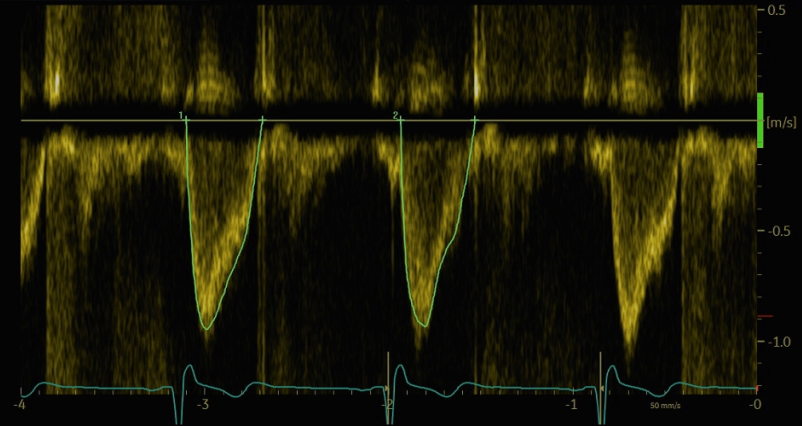


52 HR

+	ssAo Vmax	0.94 m/s
	ssAo Vmoy	0.62 m/s
	ssAo GDmax	3.55 mmHg
	ssAo GDmoy	1.81 mmHg
	ssAo ITV	25.40 cm
	ssAo Durée	409.13 ms
	FC	52.23 BPM



Spectrum Auto Recognized



52 HR

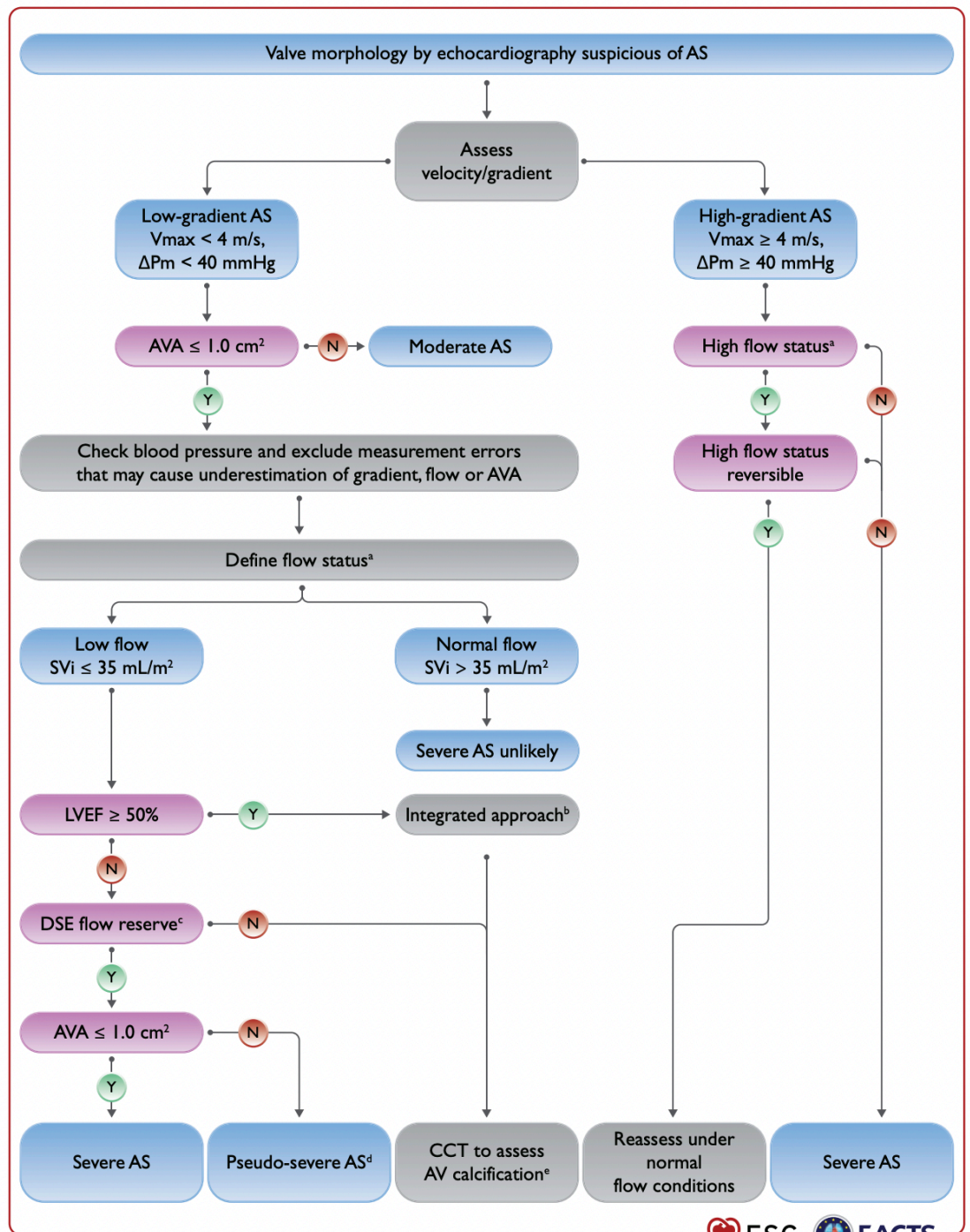
+

L 21 mm

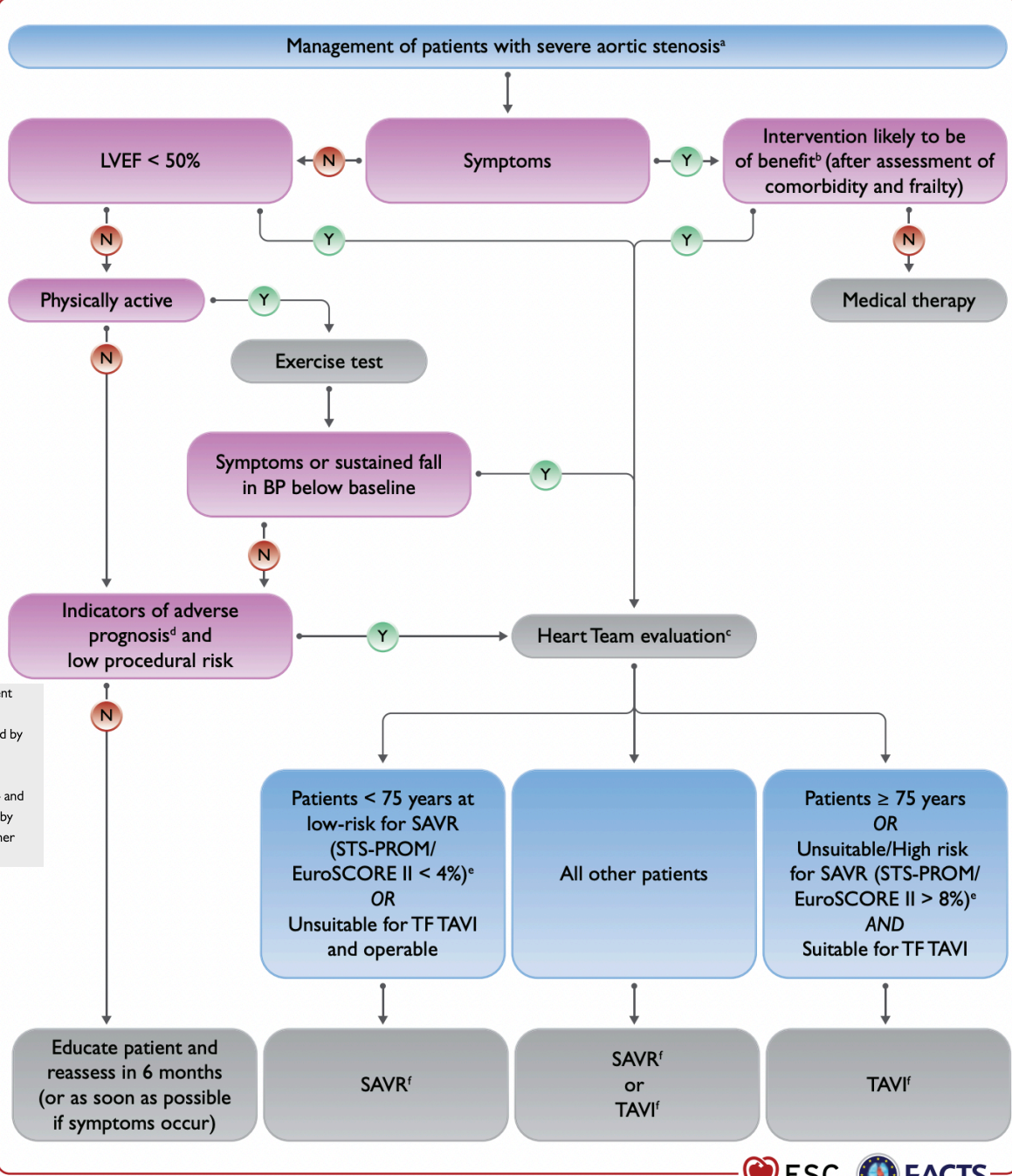


52 HR

RA



RA



- Very severe aortic stenosis (mean gradient ≥ 60 mmHg or $V_{max} > 5$ m/s).^{9,242}
- Severe valve calcification (ideally assessed by CCT) and V_{max} progression ≥ 0.3 m/s/year.^{164,189,243}
- Markedly elevated BNP levels ($> 3 \times$ age- and sex-corrected normal range) confirmed by repeated measurements and without other explanation.^{163,171}

Plan

- RA néonatal
- Sténose sous valvulaire aortique
- RA de l'adulte
- IA

IA : physiopathologie

Aigue :

- Volume regurgitant soudain sur VG de taille normale.
- Elevation rapide du VTDVG et de la POG
- Vol coronaire
- OAP, choc cardiogénique, ischémie

Chronique :

- Surcharge volumique chronique
- Remodelage progressif : Elevation VTDVG
- Altération FEVG tardive

IA de l'adulte

Table 5 Echocardiographic criteria for the definition of severe aortic valve regurgitation

Qualitative	
Valve morphology	Abnormal/flail/large coaptation defect
Colour flow regurgitant jet area ^a	Large in central jets, variable in eccentric jets
CW signal of regurgitant jet	Dense
Other	Holodiastolic flow reversal in descending aorta (EDV >20 cm/s)
Semiquantitative	
Vena contracta width (mm)	>6
Pressure half-time ^b (ms)	<200
Quantitative	
EROA (mm ²)	≥30
Regurgitant volume (mL/beat)	≥60
Enlargement of cardiac chambers	LV dilatation

Evaluation échographique

Diagnostic

- Etiologie
- Lésions associées

Réponse VG

- Dimensions VG
- Volumes VG
- FEVG

Sévérité

- Doppler continu aortique
 - R PISA
 - SOR, VR, PHT
- Vena contracta
- VTD isthme aortique
 - PAD (Clinique)

- Surveillance ?
- Intervention ?

Etiologies

Congenitale

- Bicuspidie
- Lésion de jet sur stenose sous valvulaire
- Laubry-Pezzi

Acquise

Anomalie des cusps

- Dégénérative
 - Iatrogène
- Rhumatismale
 - EI
- Marastique
- Traumatique

Anomalies aortiques

- Marfan
- Dissection aortique
- Aortite syphilitique
 - Spondylarthrite
 - Ehlers-Danlos
- Ostéogénèse imparfaite
 - Fallot

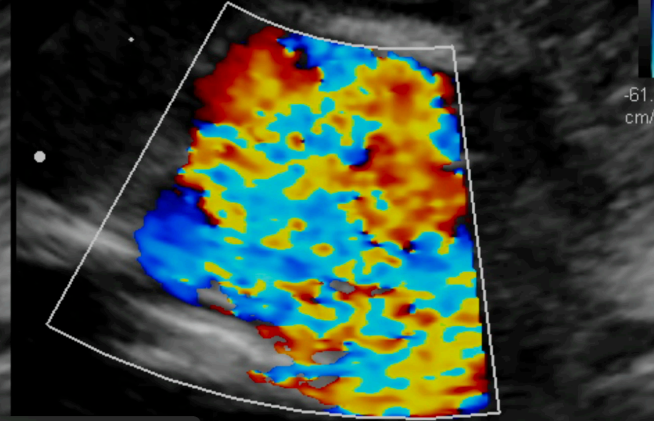
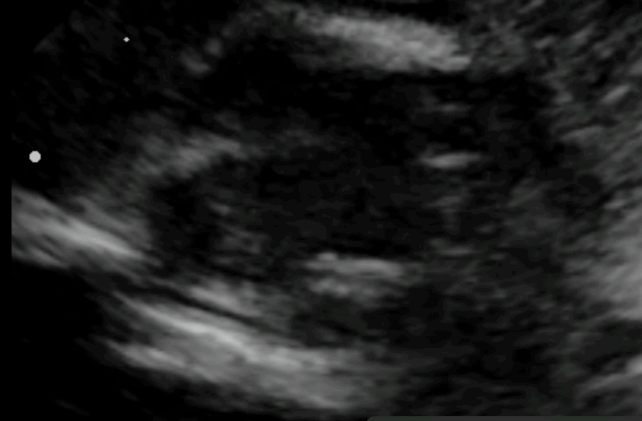
TAC

Fuite valve
troncale
sévère

Pédiatrique

S12-4
32Hz
6.0cm
Z 2.0
2D
77%
C 52
P Arrêt
Gén

Coul
65%
7110Hz
FP 639Hz
4.4MHz



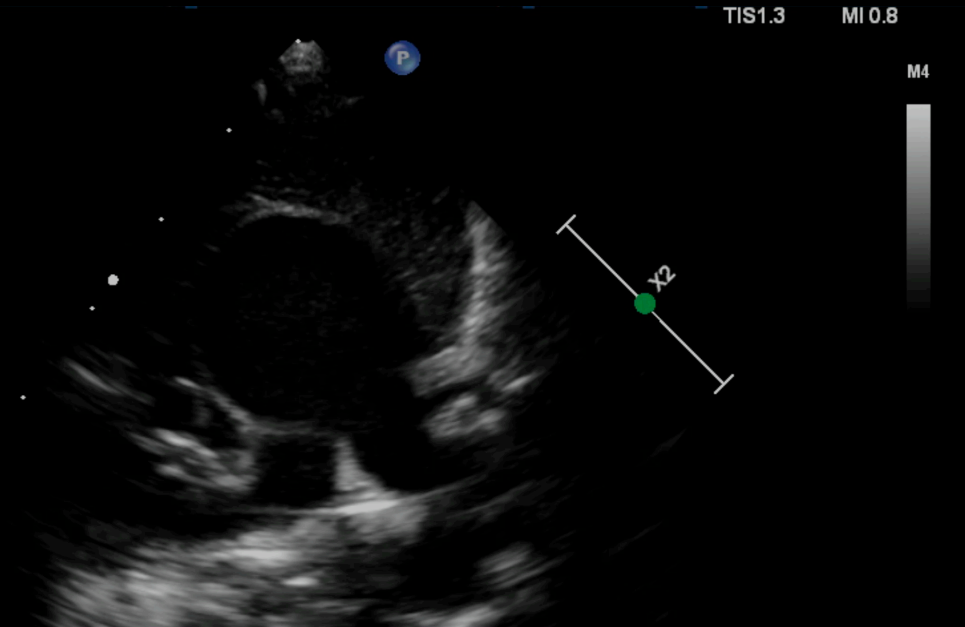
TIS1.0 MI 0.7

M4
+61.6
-61.6
cm/s

Pédiatrique

S12-4
48Hz
6.0cm

2D
75%
C 52
P Arrêt
Gén



TIS1.3 MI 0.8

M4

TAC

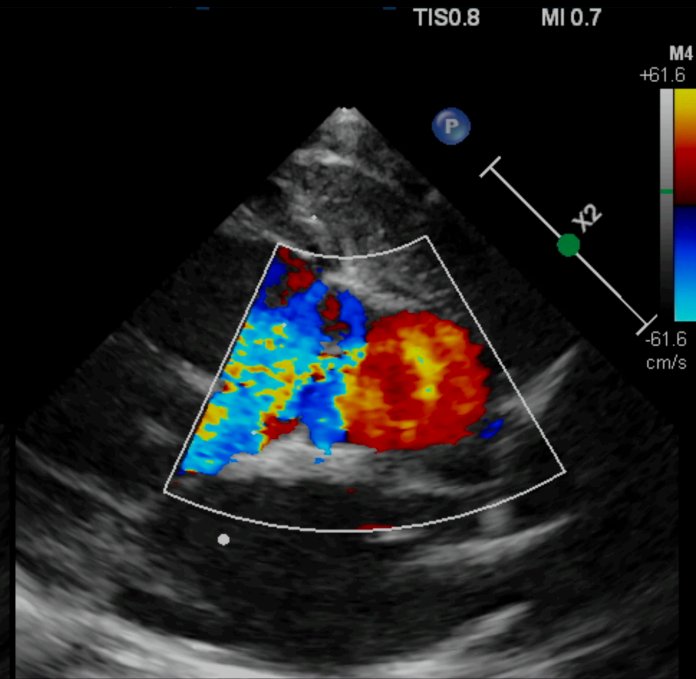
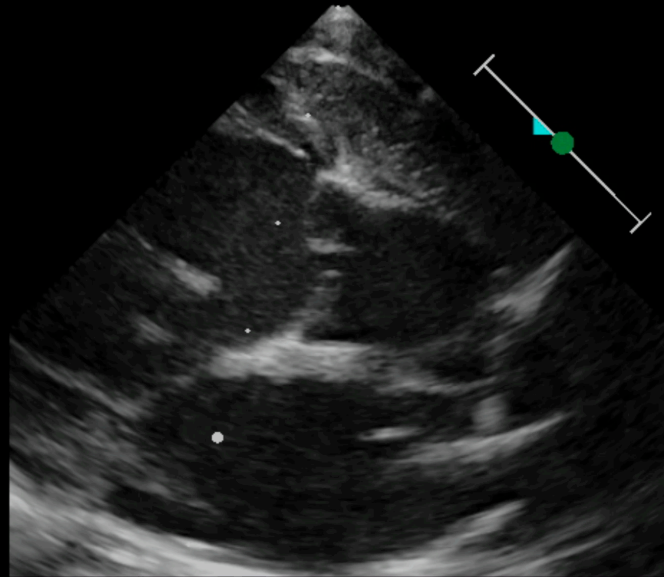
Fuite valve
troncale
sévère

Pédiatrique

S12-4
25Hz
6.0cm

2D
82%
C 52
P Arrêt
Gén

Coul
65%
7110Hz
FP 639Hz
4.4MHz

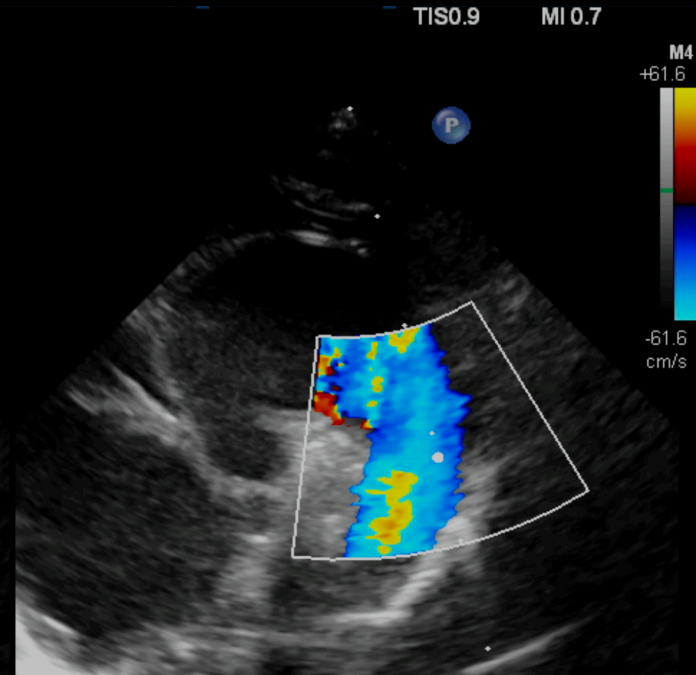
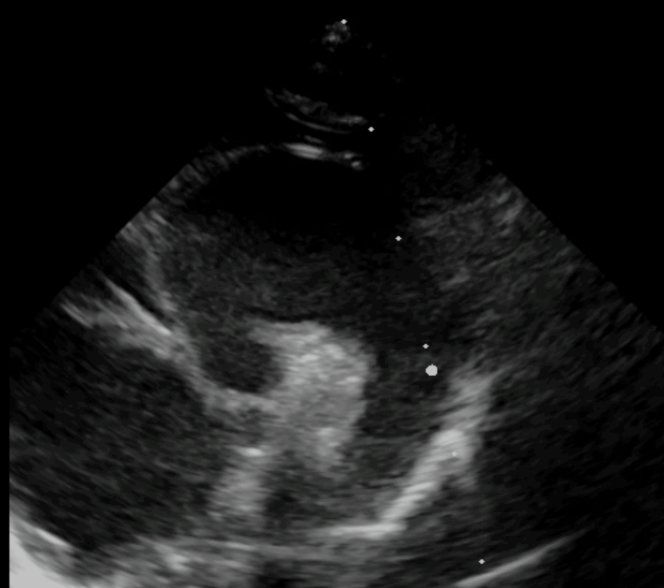


Pédiatrique

S12-4
18Hz
6.0cm

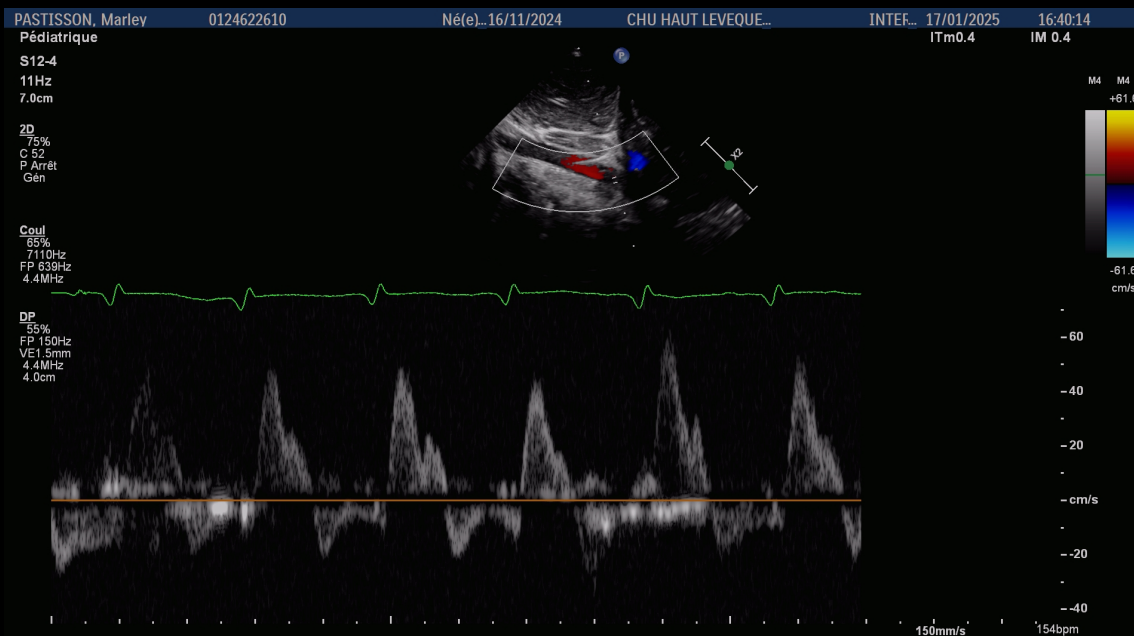
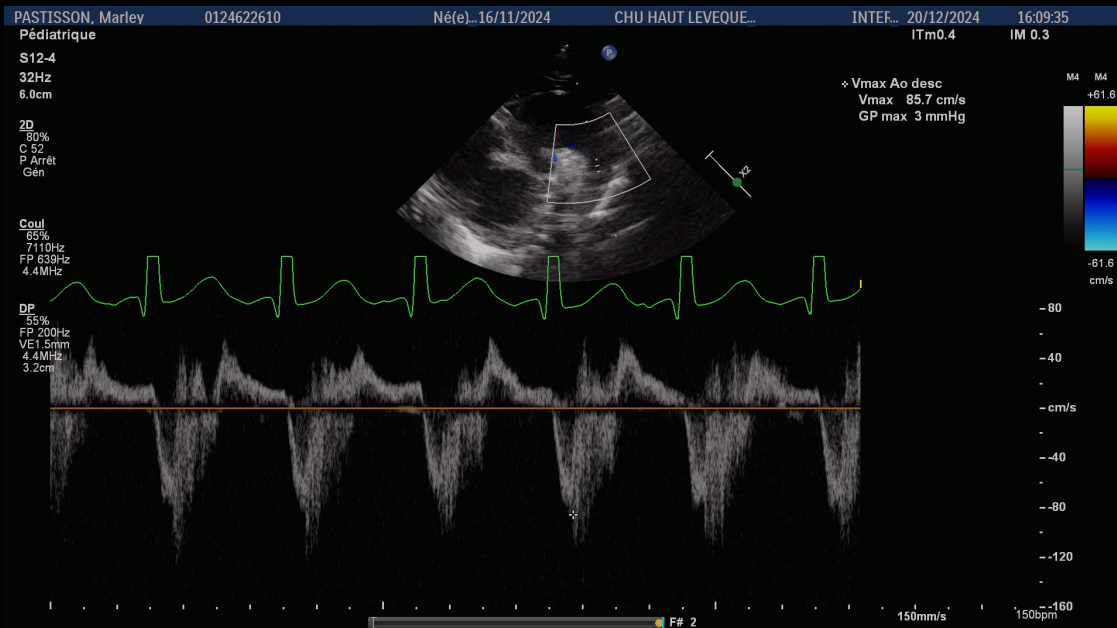
2D
77%
C 52
P Arrêt
Gén

Coul
65%
7110Hz
FP 639Hz
4.4MHz



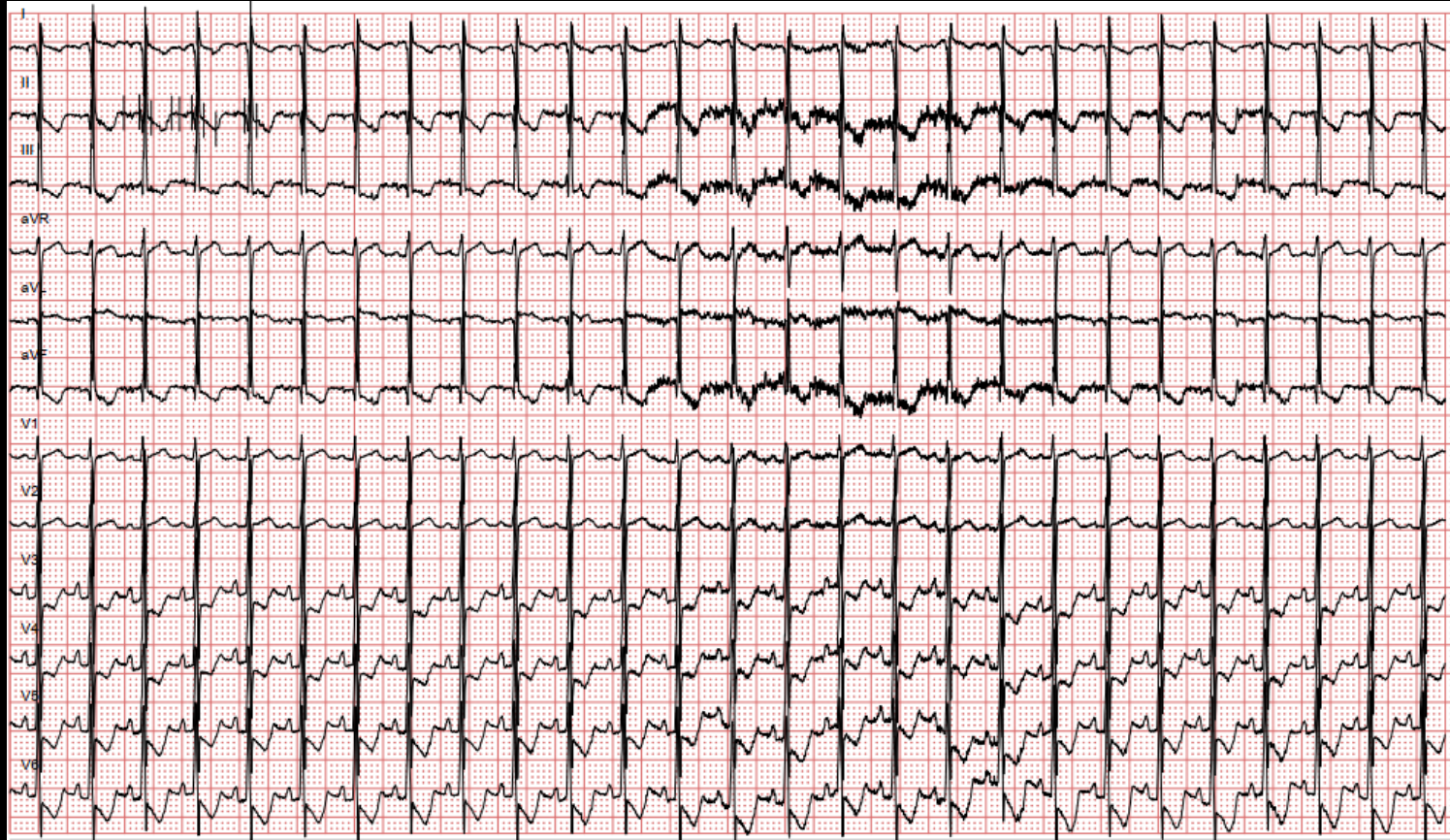
TAC

Fuite valve troncale sévère



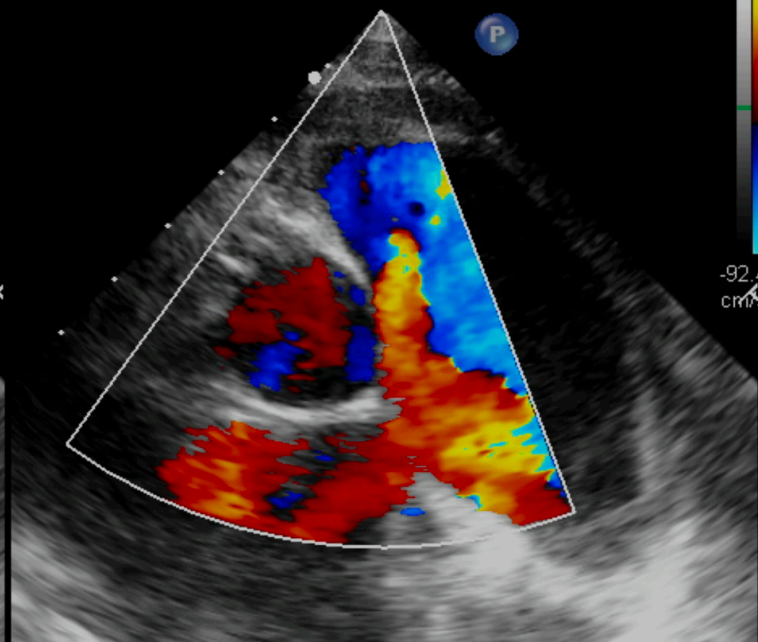
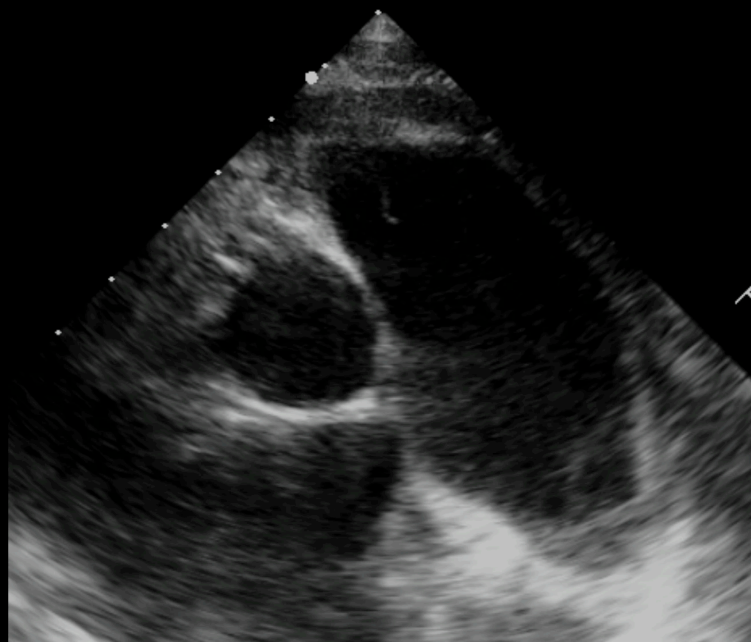
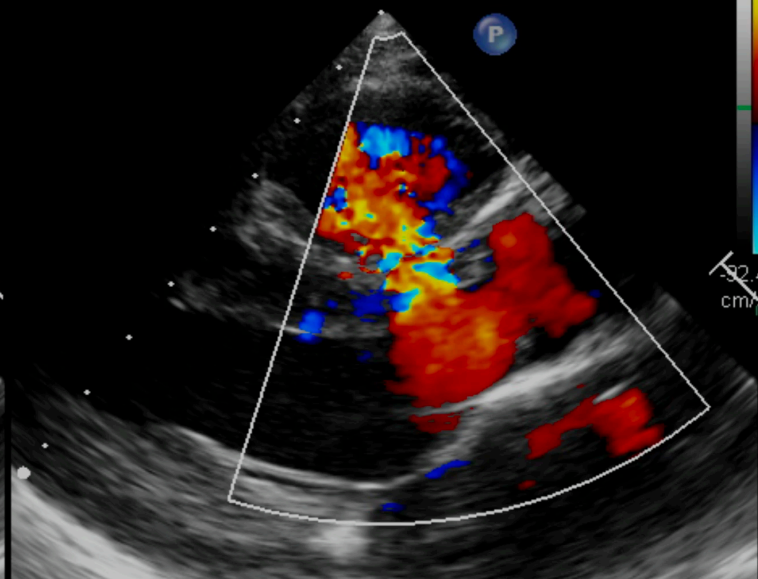
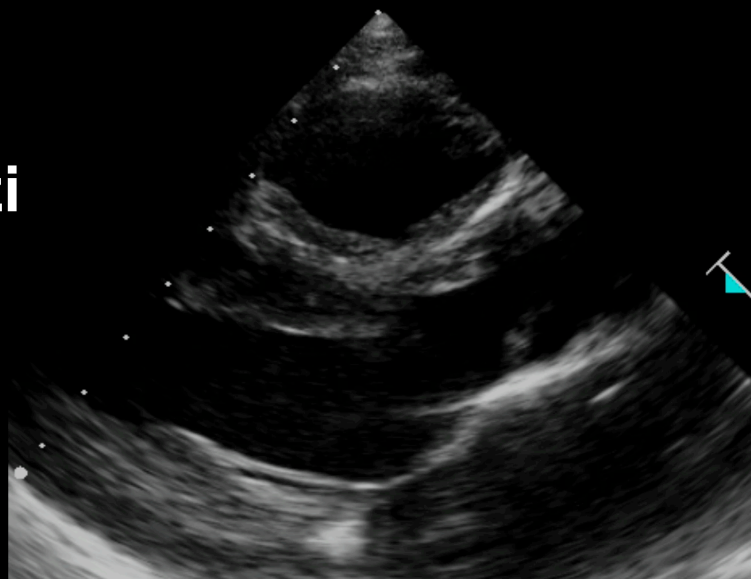
TAC

**Fuite valve
troncale
sévère**

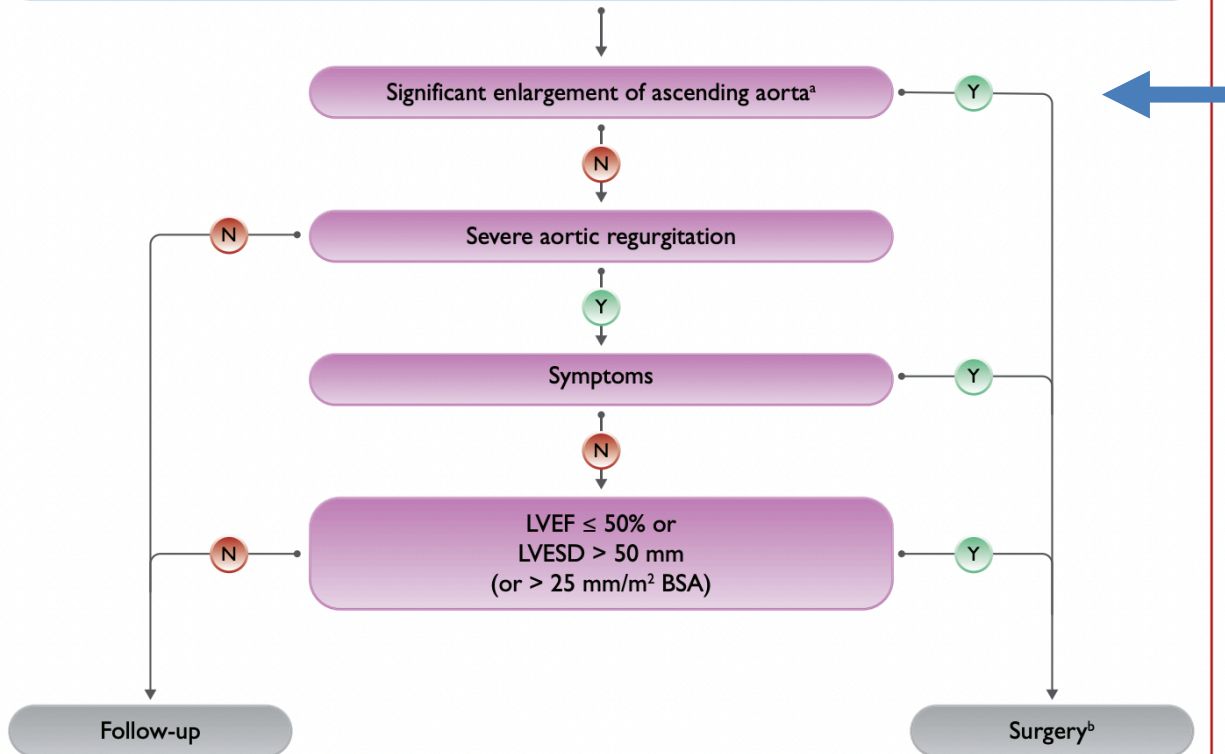


Laubry-Pezzi

CIV + IA



Management of patients with aortic regurgitation



Ascending aortic surgery is recommended in patients with Marfan syndrome who have aortic root disease with a maximal ascending aortic diameter ≥ 50 mm.

Ascending aortic surgery should be considered in patients who have aortic root disease with maximal ascending aortic diameter:

- ≥ 55 mm in all patients.
- ≥ 45 mm in the presence of Marfan syndrome and additional risk factors^d or patients with a *TGFBR1* or *TGFBR2* mutation (including Loeys–Dietz syndrome).^e
- ≥ 50 mm in the presence of a bicuspid valve with additional risk factors^d or coarctation.

^dFamily history of aortic dissection (or personal history of spontaneous vascular dissection), severe aortic or mitral regurgitation, desire for pregnancy, uncontrolled systemic arterial hypertension and/or aortic size increase >3 mm/year (using serial echocardiography or CMR measurements at the same level of the aorta confirmed by ECG-gated CCT).

I	C
IIa	C

Questions ?