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ACHD

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ESC European Society of Cardiology

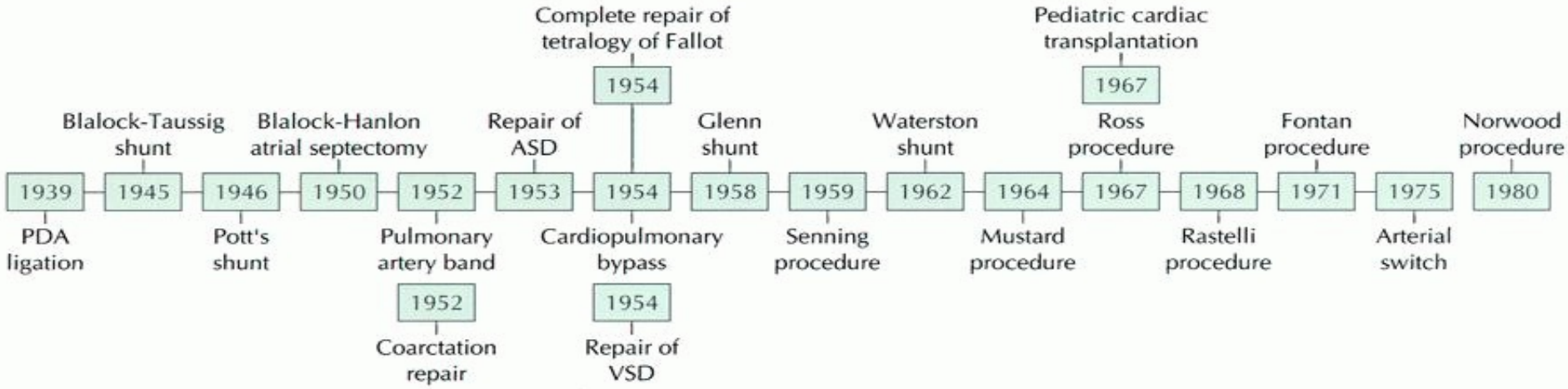
European Heart Journal (2021) **42**, 563–645
doi:10.1093/eurheartj/ehaa554

ESC GUIDELINES

2020 ESC Guidelines for the management of adult congenital heart disease

The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC)

Suivi des cardiopathies congénitales à l'âge adulte



800.000 patients aux USA
Augmentation de 25% de 2000 à 2010
Perspective 2010: la majorité des congénitaux opérés auront plus de 16 ans

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Heart 2004;91:207-212

JACC 2005;46:1-8

Plan

- Cardiopathies simples à l'âge adulte
  - Opérées
  - Non opérées

- Cardiopathies complexes opérées

Plan

- Cardiopathies simples à l'âge adulte
  - Opérées
  - Non opérées

Cardiopathies simples: CIV- CIA- CA

- réparation “à temps”
- Guérison complète ?
- Devenir à long terme

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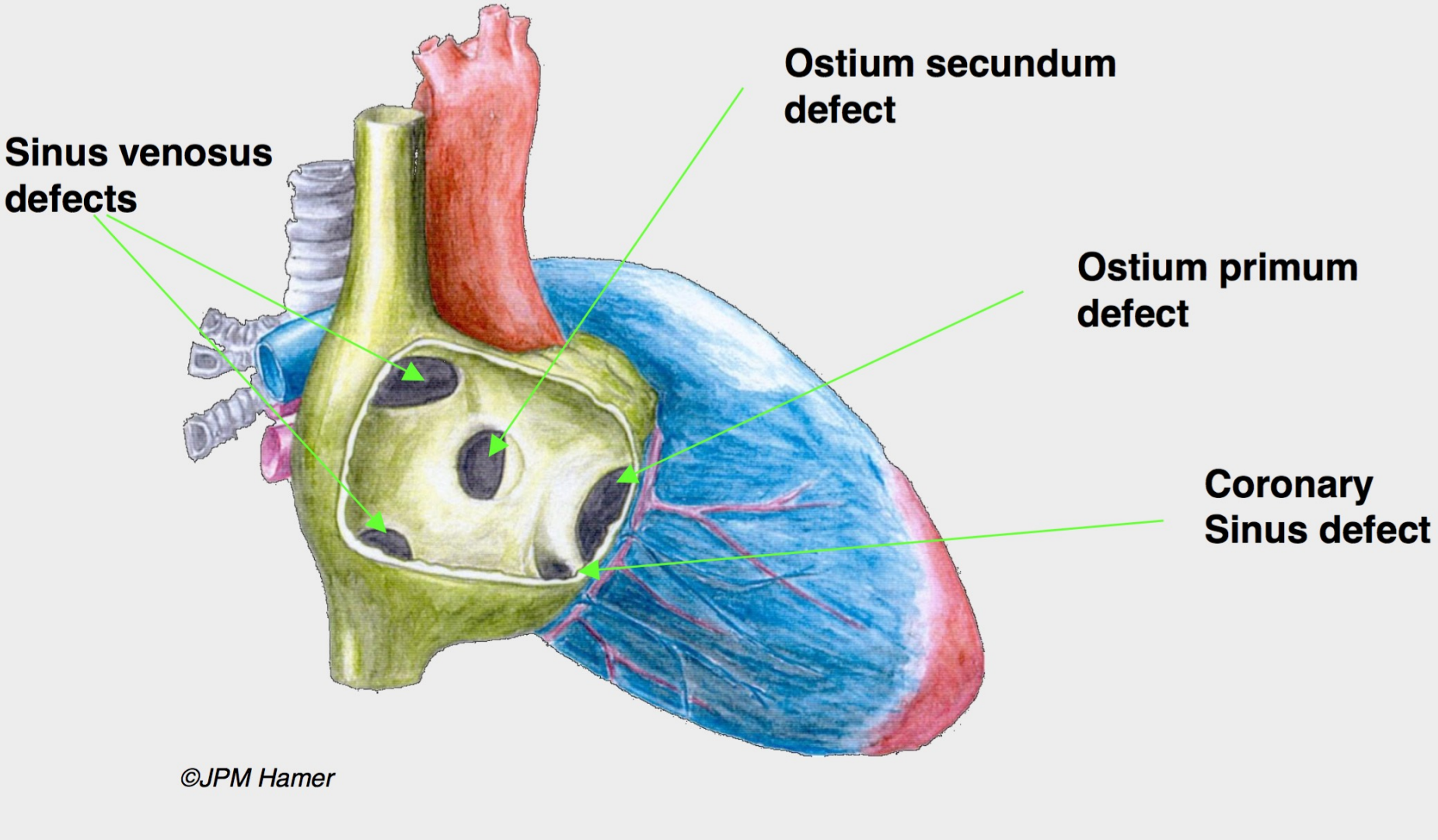
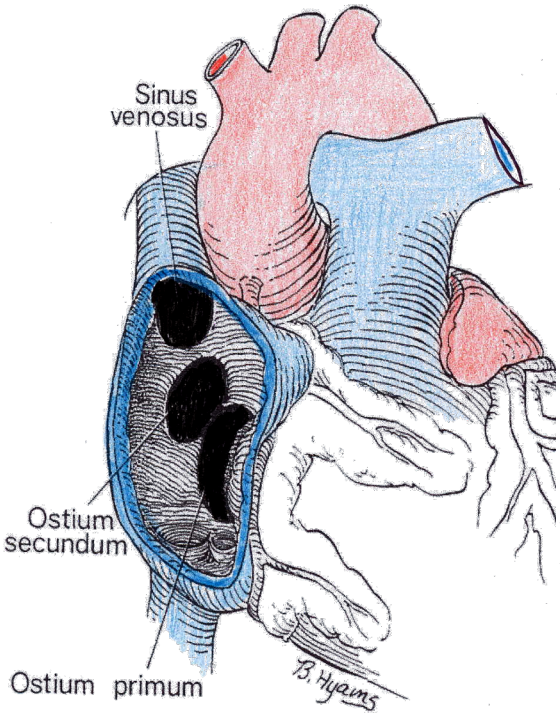
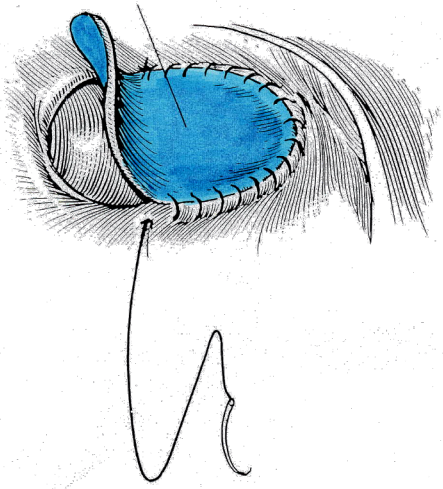
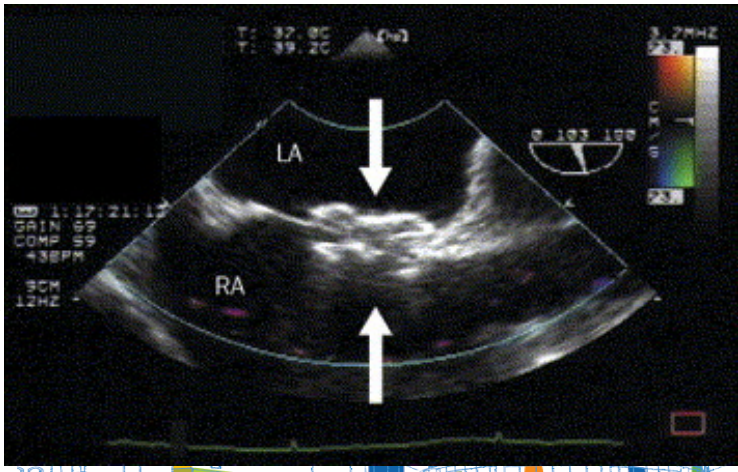


CIA

- Réparation “à temps”
- Guérison complète ?
- CIA : problèmes rythmiques et HTAP
- Connaitre les indications de fermeture percutanées

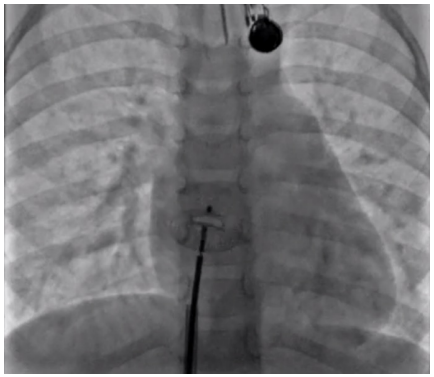




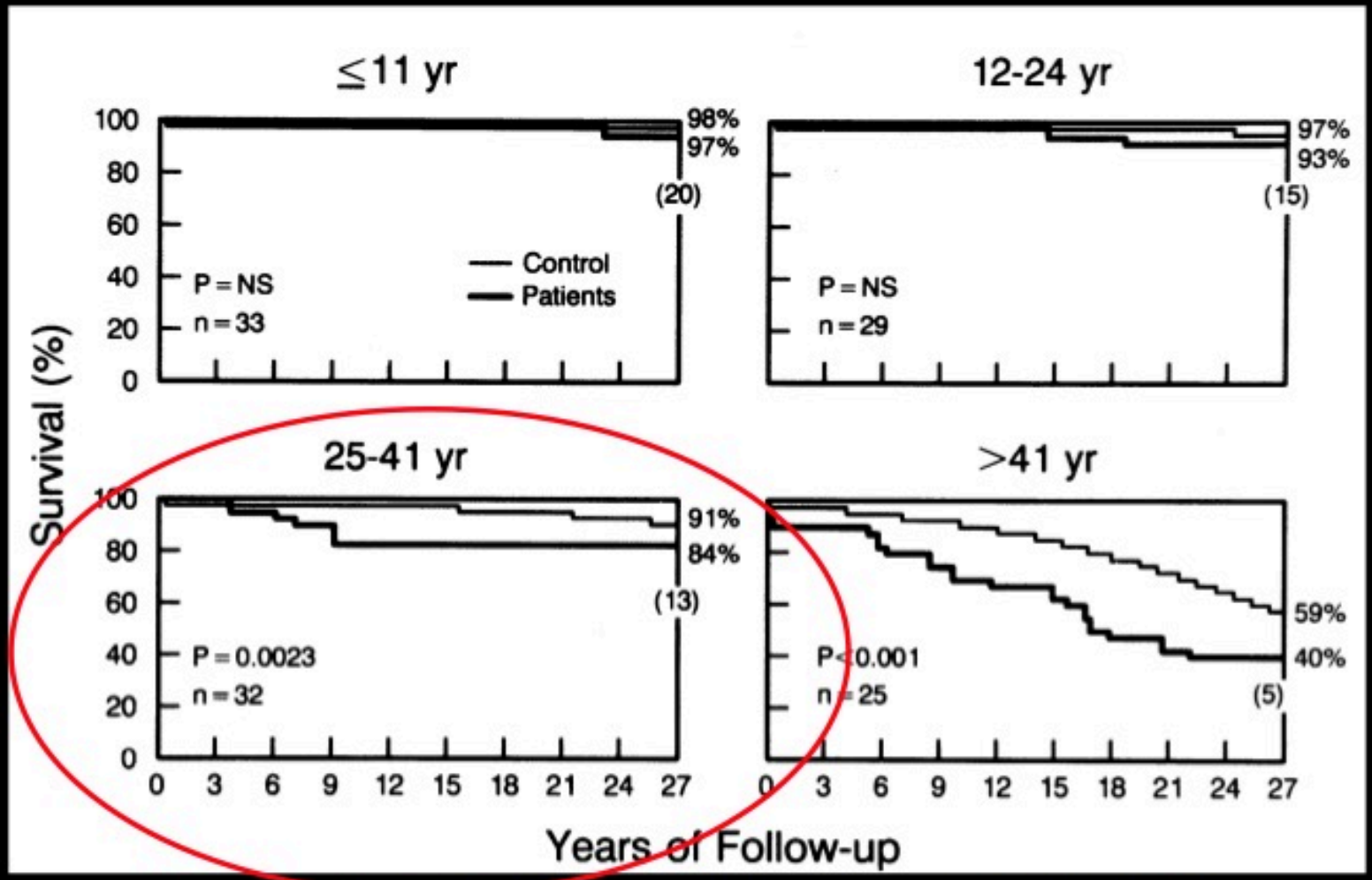


COMPLICATIONS TARDIVES

- Valvular failure
- Cardiac Erosion / perforation
- AV Conduction abnormalities
- Device thrombosis
- Nickel allergy
- Stroke
- Endocarditis
- Atrial arrhythmias

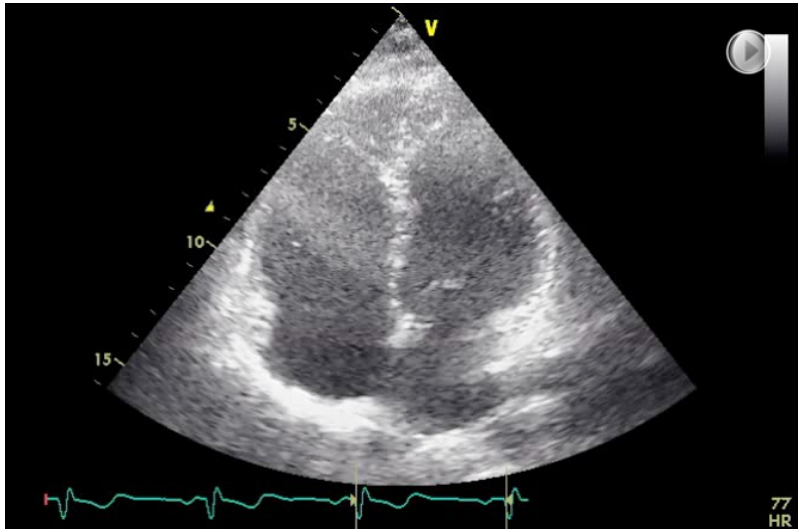


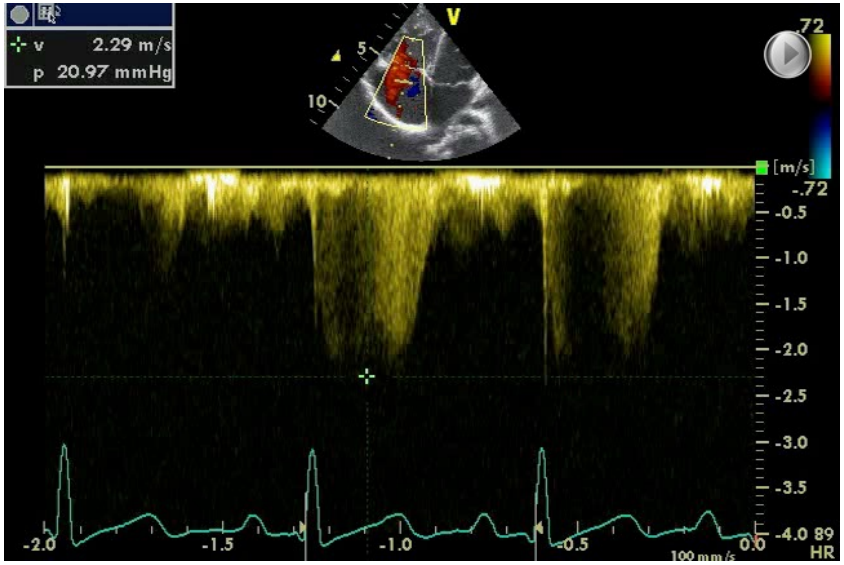
Long-Term Survival after ASD Closure by Age

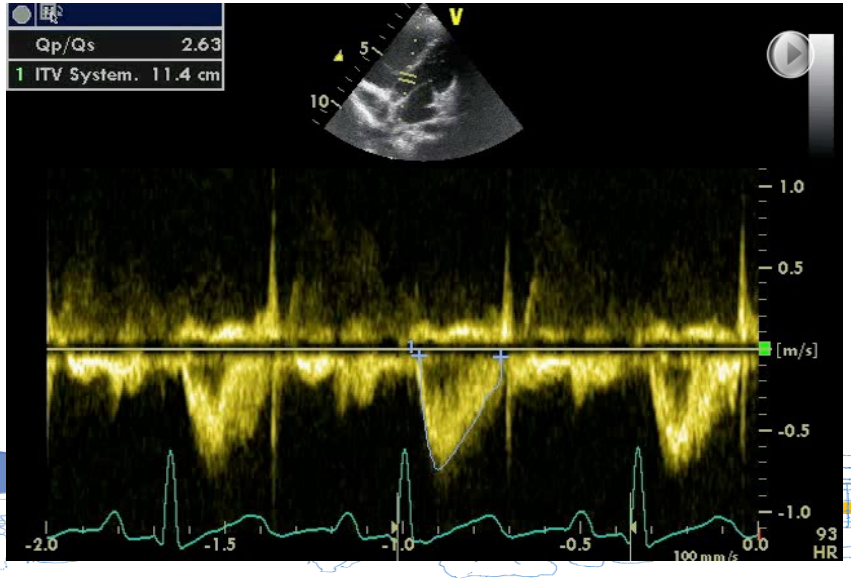


Murphy JG, et al. N Engl J Med 1990;323:1645-50

CIA Ostium secundum







Cas 1
Souffle et dyspnée minime
Chez un adolescent sportif

Dilatation des cavités droites
Pressions pulmonaires normales

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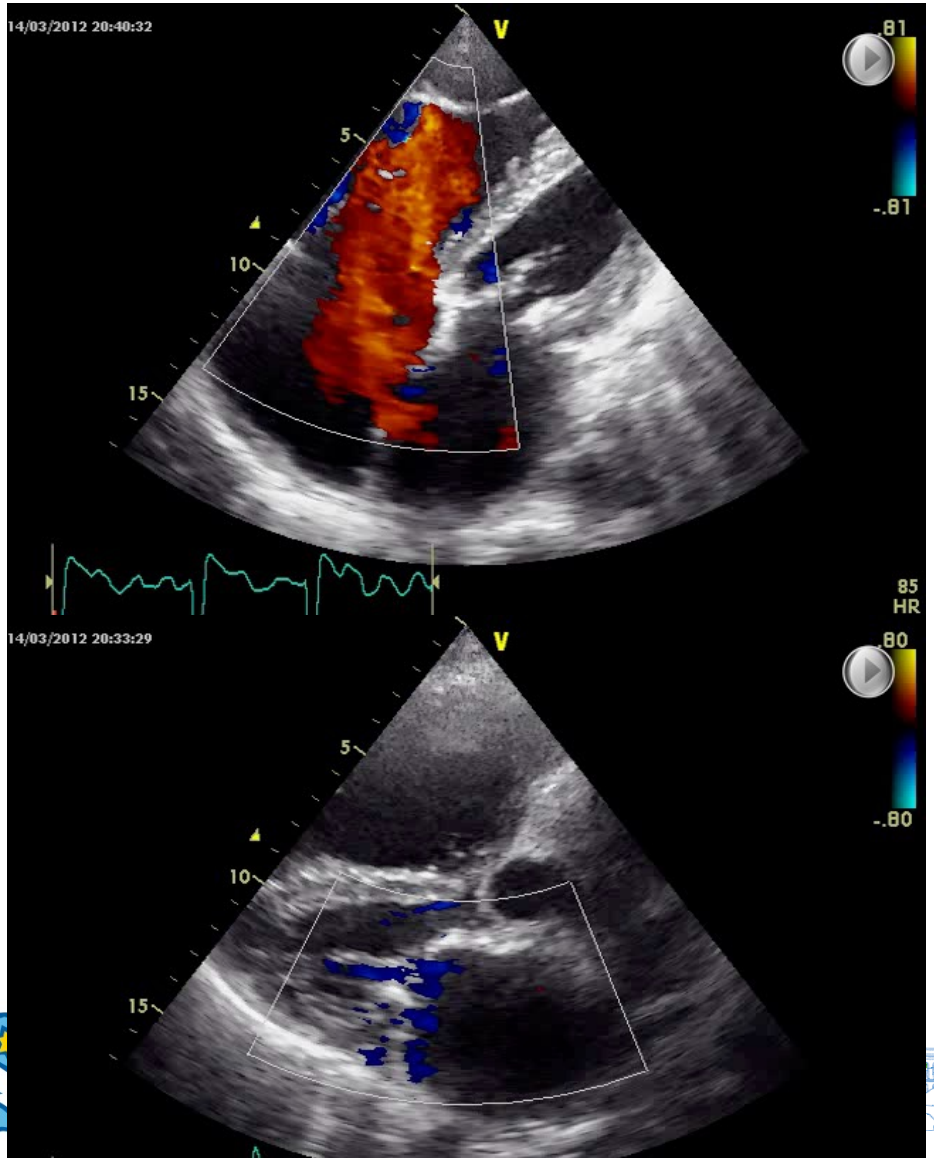
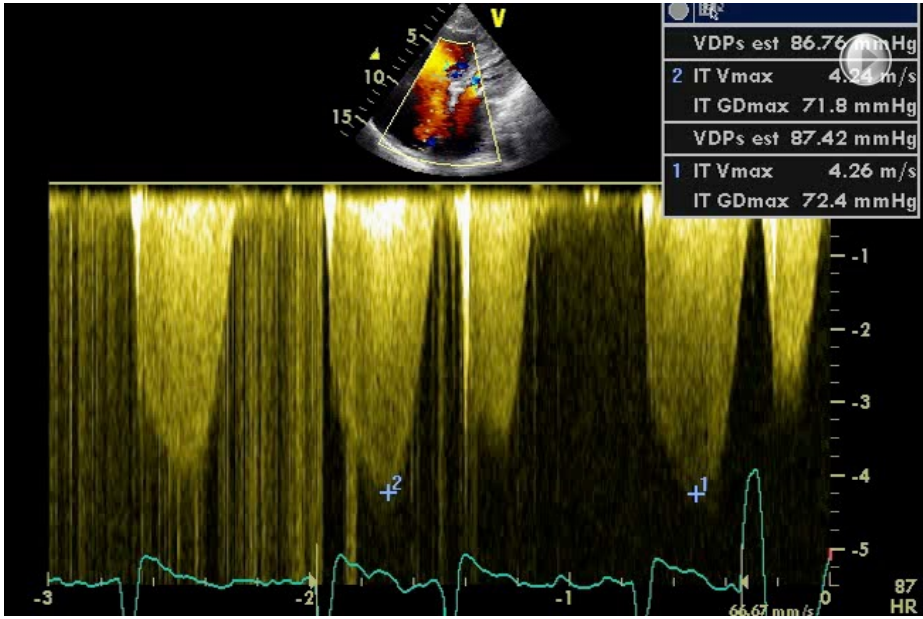
CIA de la personne âgée

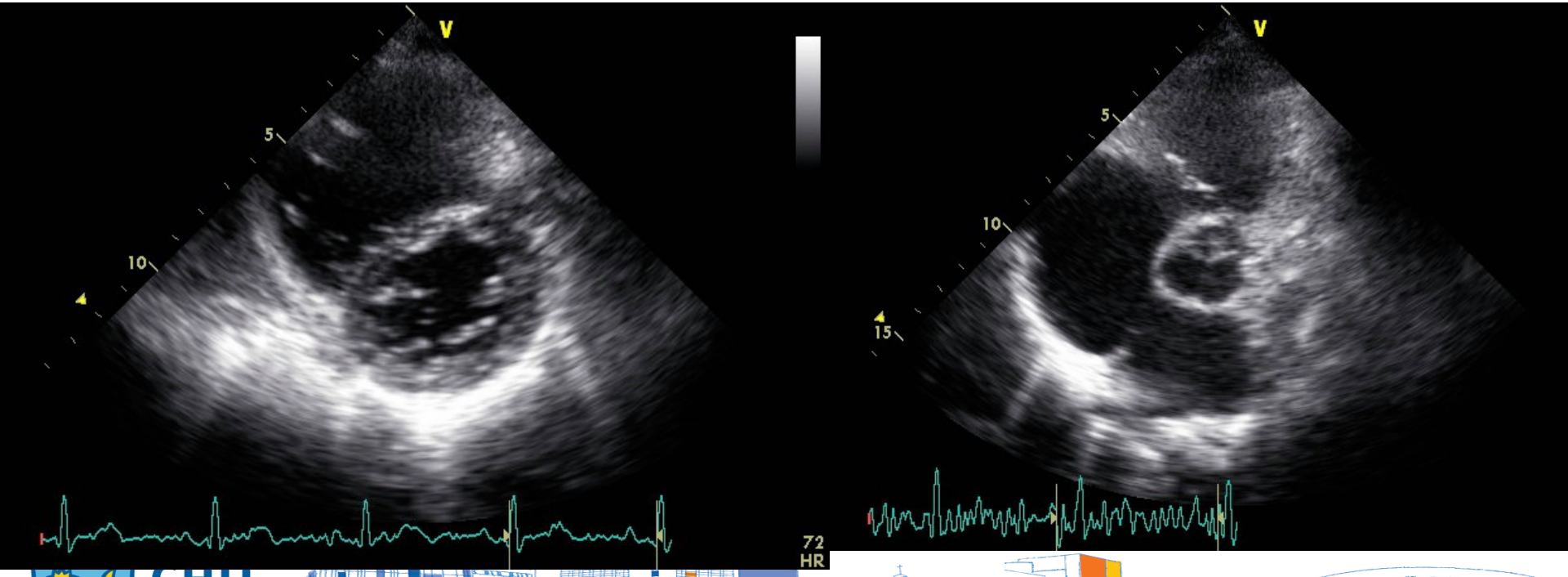




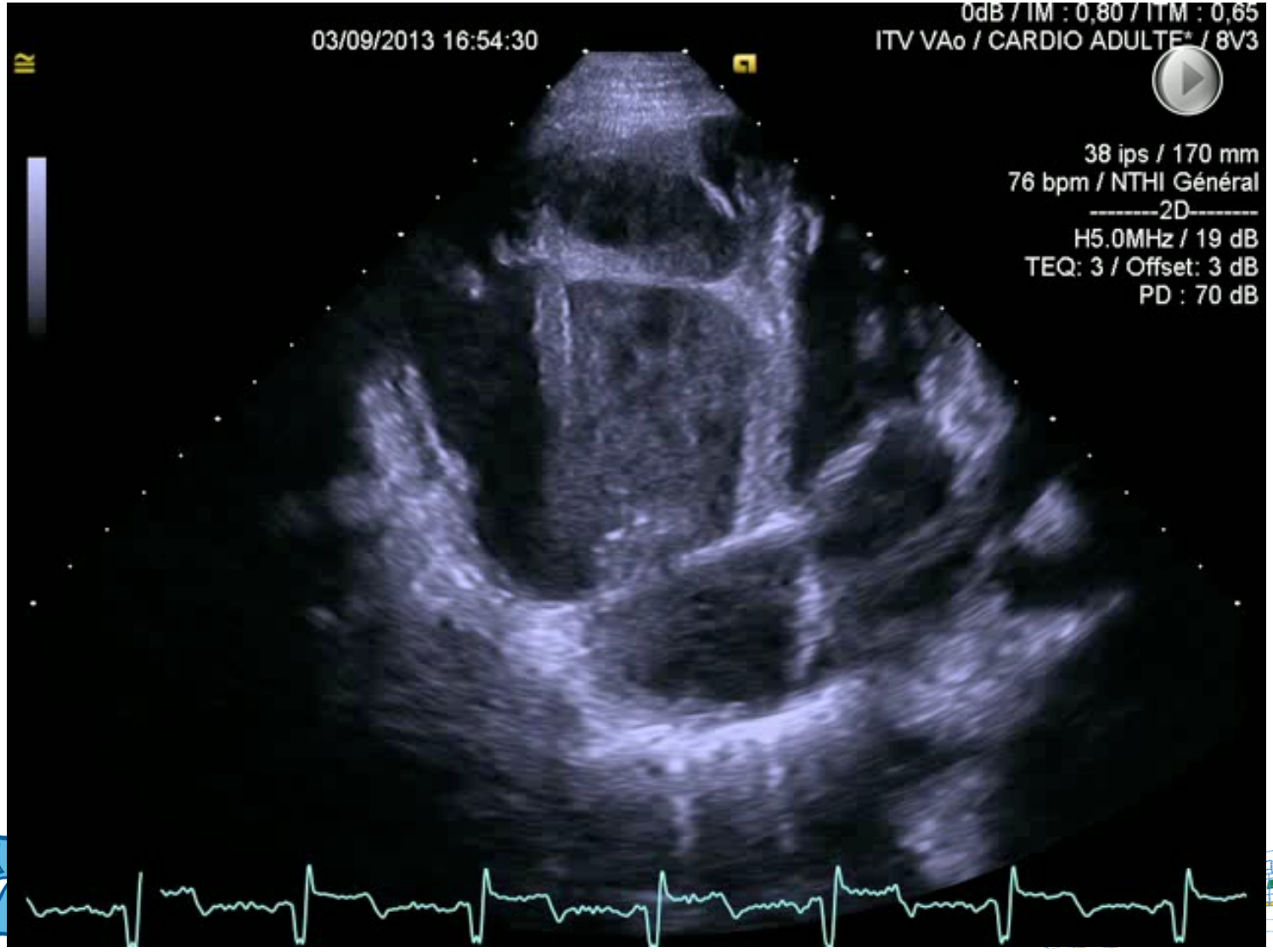
Cas 2
Femme 81 ans
AEG depuis fracture du fémur en 2009
Très peu mobile (lit-fauteuil)
1ère Décompensation cardiaque droite

FA permanente
HTAP 75+15mmHg
IT grade 3
IM grade 2fort Rao lâche

Cas3
Femme 24ans
Découverte fortuite d'une dilatation du TAP sur RP médecine du travail
Dyspnée stade 2 (sous estimée par la patiente jusqu'à présent)
ECG: HAD BBD
Echo: CIA 15mm, shunt bidirectionnel, dilatation des cavités droites
PAPS 55%
KT : PAPm 50mmH Pcap 3 RVP 10UW



CIA et HTAP


03/09/2013 16:54:30
0dB / IM : 0,80 / ITM : 0,65
ITV VAo / CARDIO ADULTE* / 8V3
38 ips / 170 mm
76 bpm / NTHI Général
--------2D--------
H5.0MHz / 19 dB
TEQ: 3 / Offset: 3 dB
PD : 70 dB

Prévalence de l'HTAP dans les CIA

- Prévalence de l'HTAP dans les CIA = 16 % du total
  - 4 % avant 20 ans
  - 18 % de 20 à 40 ans
  - 40 % après 40 ans
- HTAP deux fois plus fréquente dans les CIA de type *sinus venosus*
  - 16 % des adultes avec *sinus venosus vs* 4 % des adultes avec OS [1]
- Rôle du sexe : 4 femmes pour 1 homme dans les CIA avec HTAP *vs* 2 / 1 sans HTAP
- Age moyen du diagnostic : 36 ans

(1) Vogel M *et al.* Heart 1999.

Développement de la maladie vasculaire pulmonaire chez les patients avec shunts gauche-droite non opérés [18,19]

Shunt gauche-droite

↓

Augmentation du flux sanguin pulmonaire

↓

Augmentation des résistances vasculaires pulmonaires (RVP)

↓

En l'absence de correction chirurgicale,
la maladie vasculaire pulmonaire peut se développer
de type histologique similaire à celui de l'HTAP idiopathique (HTAPi) [18]

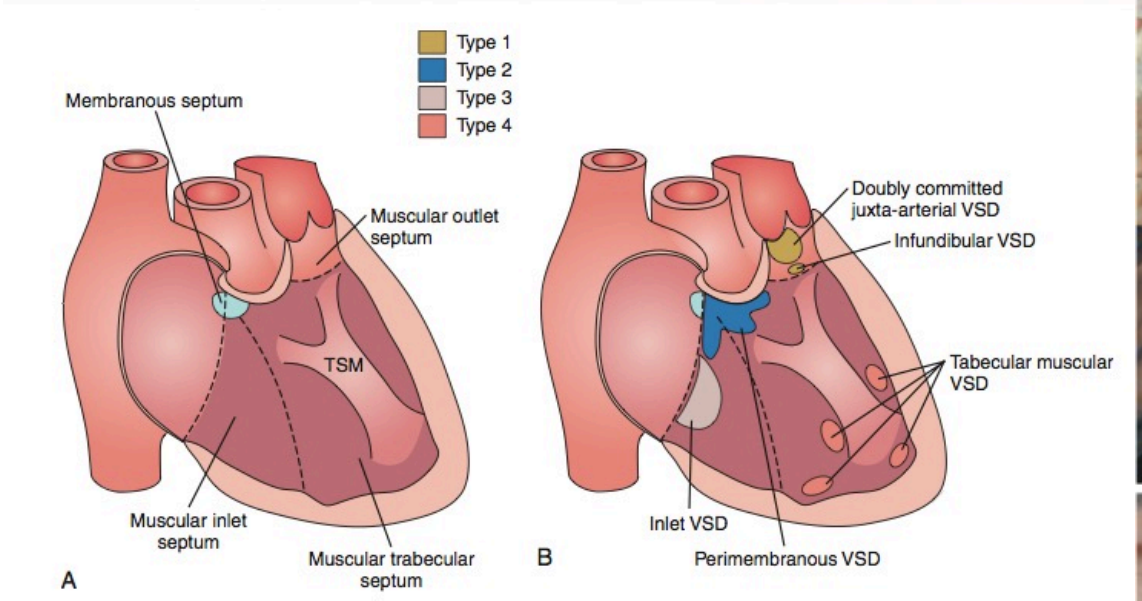
↓

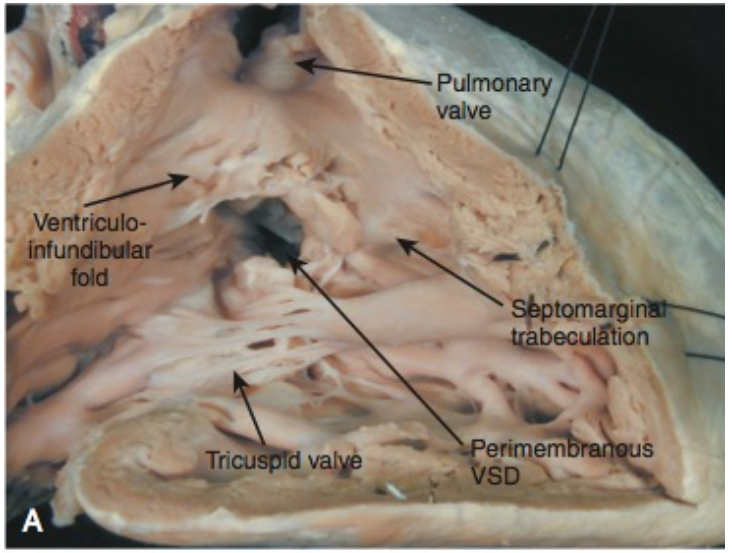
Dommages irréversibles du système vasculaire pulmonaire
et augmentation des RVP

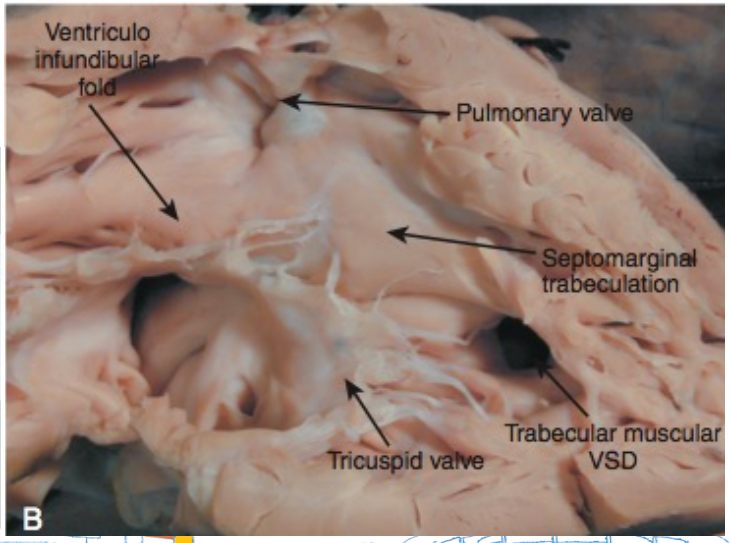
L'HTAP peut se développer après chirurgie correctrice ou palliative de la CC [18]

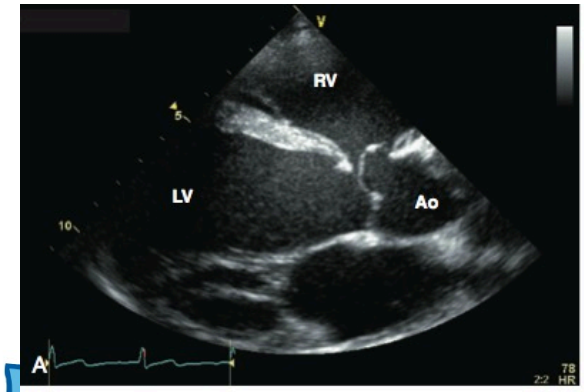
[18]Galié N et al. *Drugs 2008;68(8):1049-66* ; [19]Beghetti M et al. *J Am Coll Cardiol 2009;53(9):733-40*

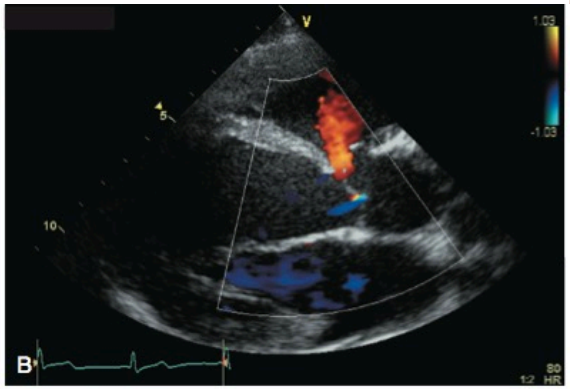
Communication interventriculaires





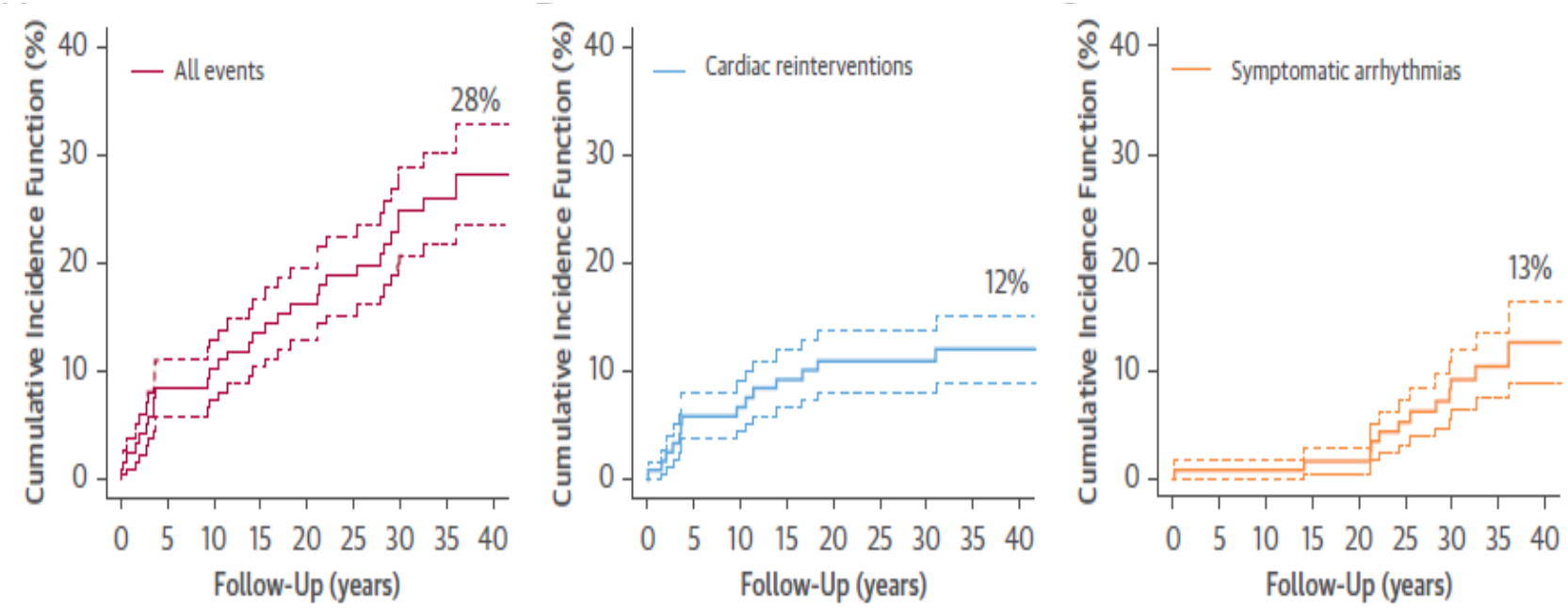






Fermeture chirurgicale et devenir à long-terme…

- 91 patients
- Fermeture chirurgicale avant 1980 et avant l'âge de 15 ans
- L'espérance de vie est réduite par rapport à la population générale



Am J Cardiol. 2010 Feb 1;105(3):404-7. doi: 10.1016/j.amjcard.2009.09.047. Epub 2009 Dec 22.

Behavior of unrepaired perimembranous ventricular septal defect in young adults.

Soufflet V[1], Van de Bruaene A, Troost E, Gewillig M, Moons P, Post MC, Budts W.

220 patients

8 endocardites

Aggravation pb aortiques

2 morts (1 MS, 1 ICC).

2 PM-DAI.

15 fermeture chir (7%).

8 fermetures spontanées

9% HTAP

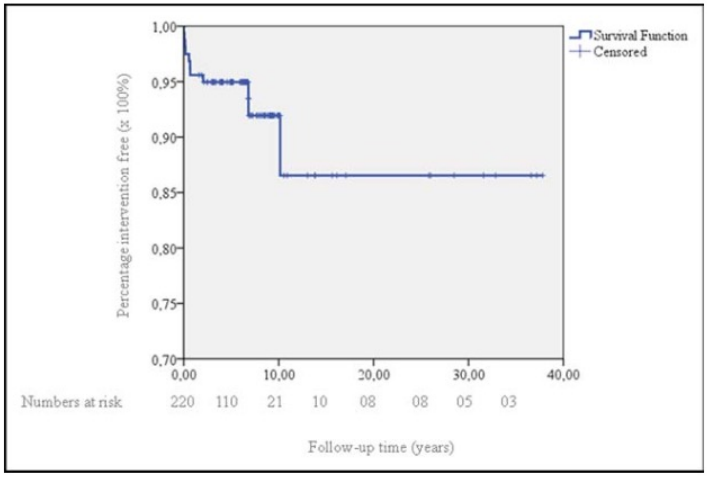


Figure 1. Kaplan-Meier curve of event-free survival, with event defined as surgical or interventional VSD closure.

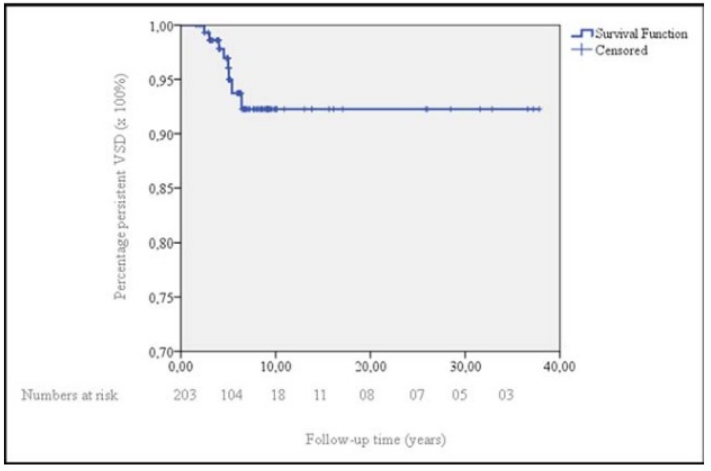


Figure 2. Kaplan-Meier curve of event-free survival, with event defined as spontaneous VSD closure.

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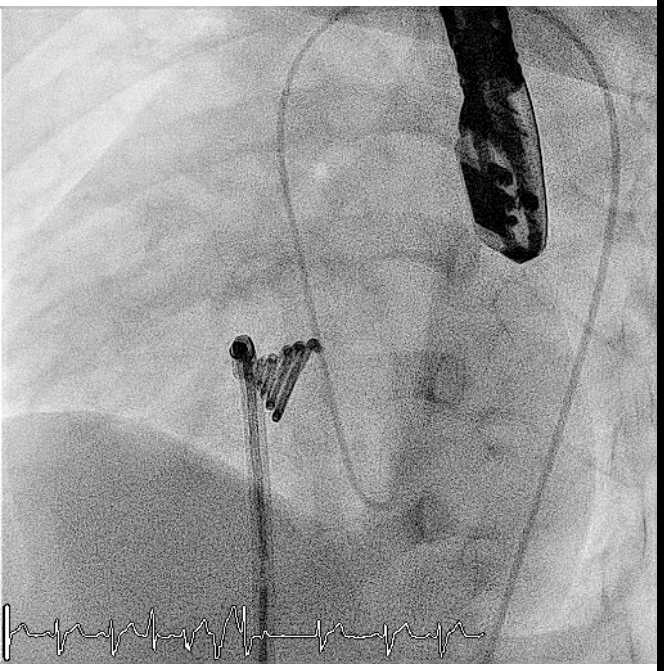
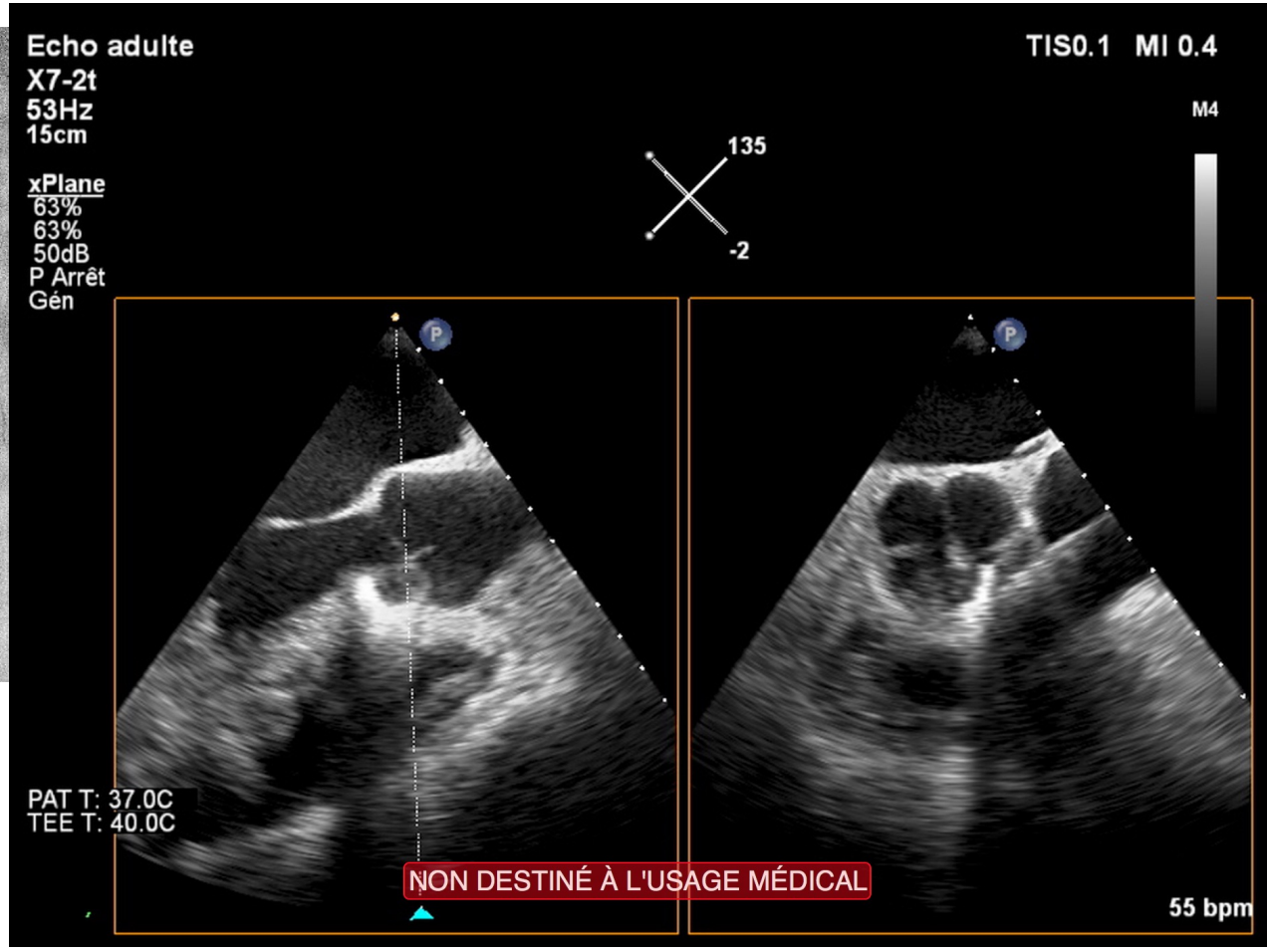
On estime qu'environ 20 à30% des CIV intermédiaires prise en charge de manière « conservatrice » pendant l'enfance auront besoin d'un traitement (chirurgie ou cathétérisme) à l'âge adulte.





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JACC: Cardiovascular Interventions

Volume 3, Issue 3, March 2010, Pages 290–297

Clinical Research

Indications and Outcomes of Surgical Closure of Ventricular Septal Defect in Adults

François-Pierre Mongeon, MD*, Harold M. Burkhart, MD†, Naser M. Ammash, MD*, Joseph A. Dearani, MD†, Zhuo Li, MS‡, Carole A. Warnes, MD*, Heidi M. Connolly, MD*

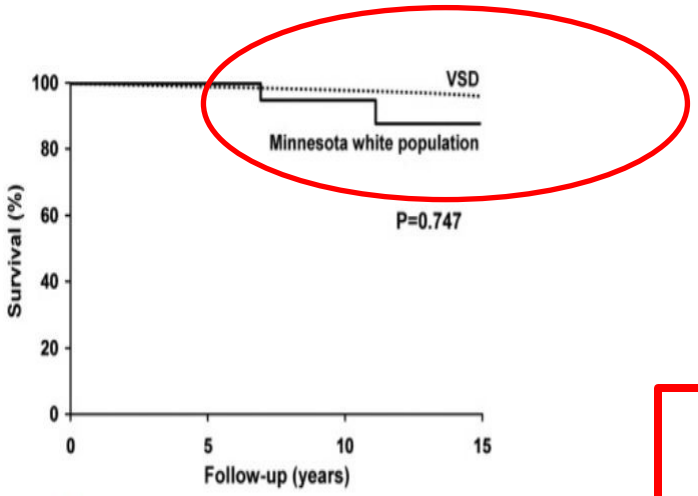

Figure 3.
Survival After Surgical VSD Closure Versus Reference Population
Survival after surgical ventricular septal defect (VSD) closure compared with a reference population. There was no difference in 15-year survival of operated patients compared with the Minnesota white population (p = 0.747).

Baseline Characteristics (n = 46)

Age at surgery, yrs	33.6±11.2
Female sex	24(52)
Society of Thoracic Surgeons type of VSD	
Type 1(subarterial)	12(26)
Type 2(membranous)	33(72)
Type 3(inlet)	1(2)
Type 4(muscular)	0
VSD diameter, mm	11.4±10.5
Associated lesions	
Coronary cusp prolapse	15(33)
With sinus of Valsalva aneurysm	5(11)
Patent foramen ovale	7(15)
Bicuspid aortic valve	6(13)
Mitral regurgitation	6(13)
Right ventricular outflow tract obstruction	6(13)
Atrial septal defect	5(11)
First degree atrioventricular block	3(7)
Patent ductus arteriosus	3(7)
Persistent left superior vena cava	3(7)
Coarctation of the aorta	2(4)
Warnes-Somerville ability index	
I	31(67)
II	15(33)
III	1(2)

Data are n (%) or mean ± SD.
VSD = ventricular septal defect.

Mortalité précoce: 0%
Mortalité tardive: 5%
Même survie à 15 ans

Table 2.
Pre-Operative Echocardiographic and Catheterization Data

Echocardiography		
Left ventricular end-diastolic diameter, mm	39	55±8
LVEF, %	42	61±7
Tricuspid regurgitation peak velocity, m/s*	26	2.81±0.46
Tricuspid regurgitation ≥ moderate	34	6(18)
Aortic regurgitation ≥ moderate	35	6(17)
Catheterization		
Qp/Qs	19	1.6±0.5
Left ventricular end-diastolic pressure, mm Hg	16	12±3
Right ventricular systolic pressure, mm Hg*	20	50±27
Mean pulmonary artery pressure, mm Hg	20	25±16
Pulmonary vascular resistance index, WU · m2	18	5.6±4.5

Data are n (number of patients for which the data were available), n (%), or mean ± SD.

LVEF = left ventricular ejection fraction.

* Patients with right ventricular outflow tract obstruction and double chamber right ventricle were excluded from the analysis of this parameter.

Indications for Surgery

Aortic regurgitation	13(28)
Left ventricular dilation	13(28)
Pulmonary hypertension	9(20)
Endocarditis	8(17)
Symptoms	6(13)
Left ventricular dysfunction	4(9)
Sinus of Valsalva aneurysm	5(11)
Ruptured	3(7)
Not ruptured	2(4)
Coronary cusp prolapse	3(7)
Double chamber right ventricle	3(7)
Mitral regurgitation	3(7)
Qp/Qs ≥2	3(7)
Subaortic stenosis	1(2)

Data are n (%). Total is >100% because many patients had multiple indications.

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Coarctation de l'aorte

Coarctation natives

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

Causes of death	*Per cent*	*Mean age and usual decades*	
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	

Natural history of coarctation of the aorta **635**

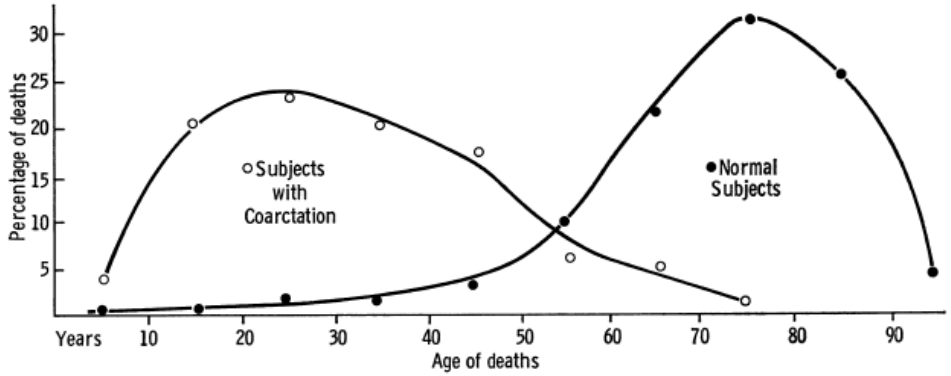

FIG. 1 *The distribution of deaths by age, excluding deaths in the first year of life. In coarctation on the left and in normal subjects on the right, there is relatively little overlapping. For coarctation, the more rounded curve from reported necropsies is shown but the flatter curve from my calculations from deaths of patients under observation is similar (see Fig. 2).*

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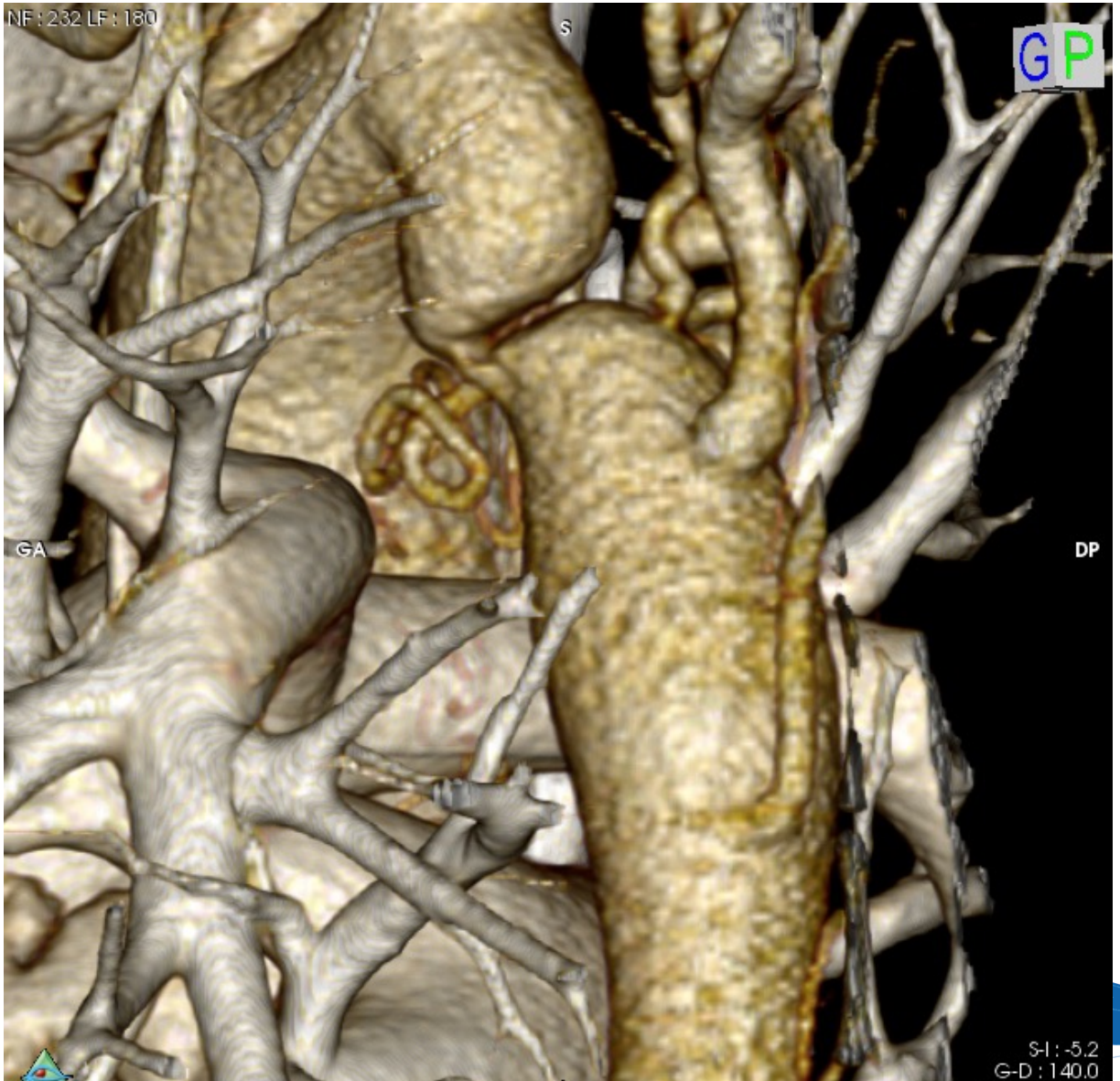
Ingénieur 42 ans, bilan d'HTA



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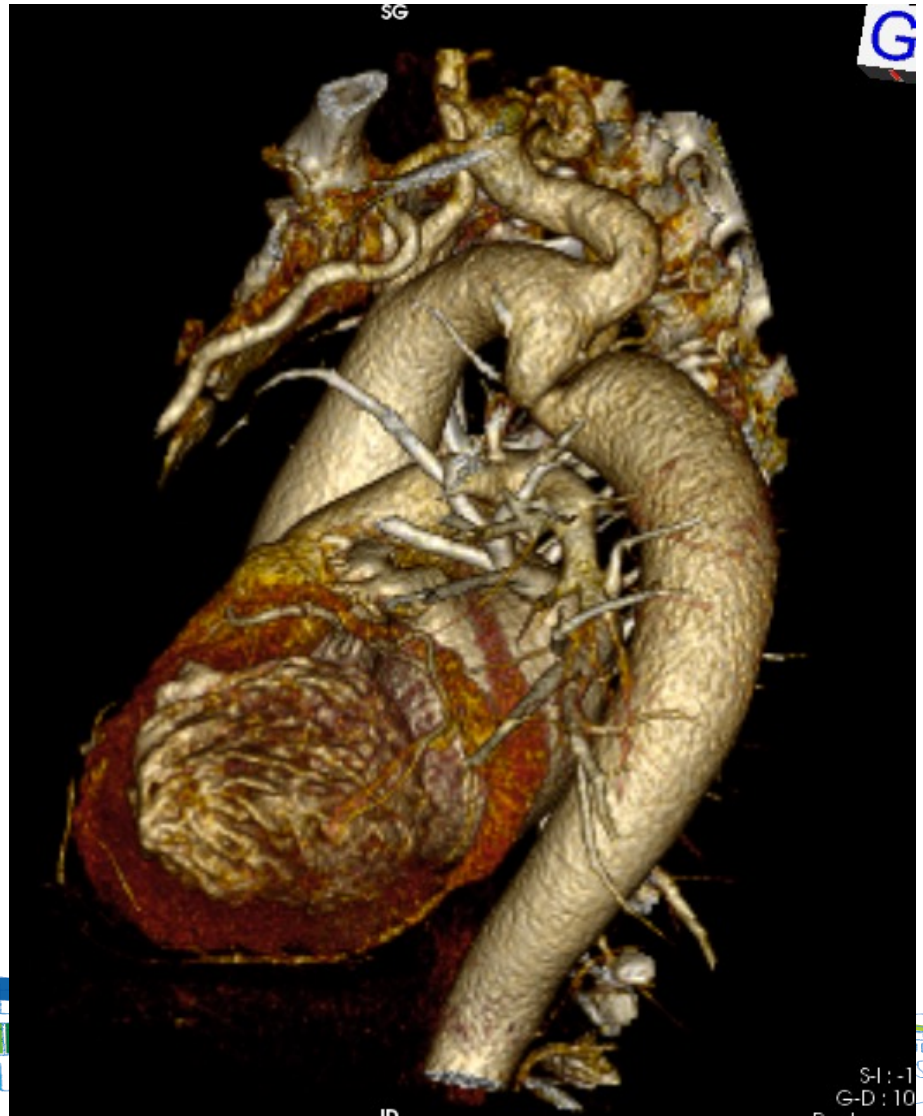
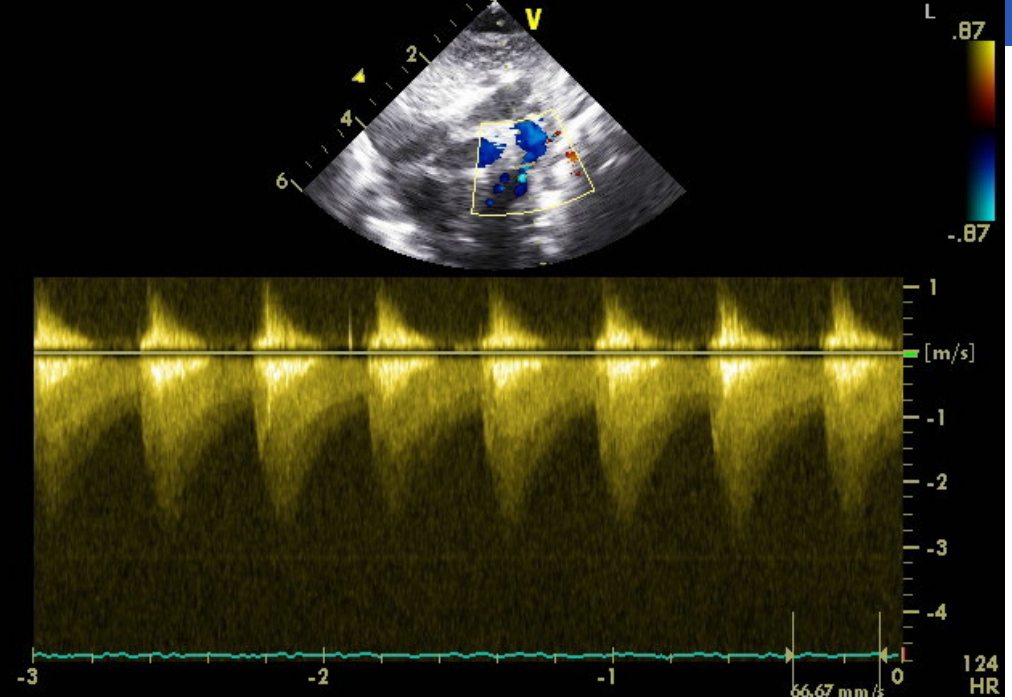
70 ans, bilan préopératoire de RA serré



Coarctations opérées

- Exercise-induced hypertension
- Late systemic hypertension
- Cerebrovascular accidents
- Premature coronary atherosclerosis
- Endocarditis
- Bicuspid aortic valve
- Aortic aneurysms or dissection
- Recoarctation

Vriend JW et al. Int J Cardiol. 2005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-.87
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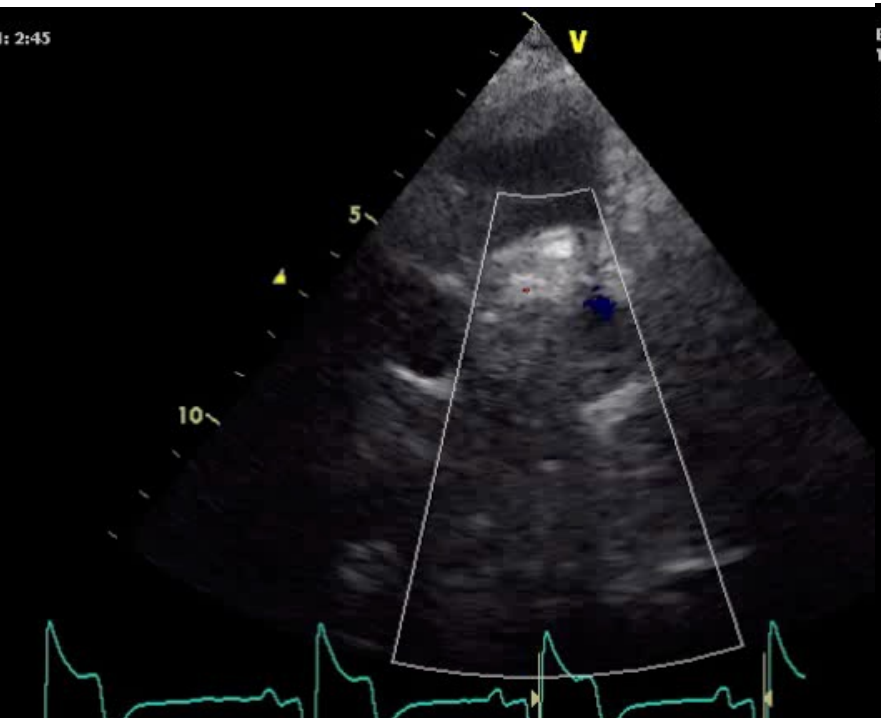
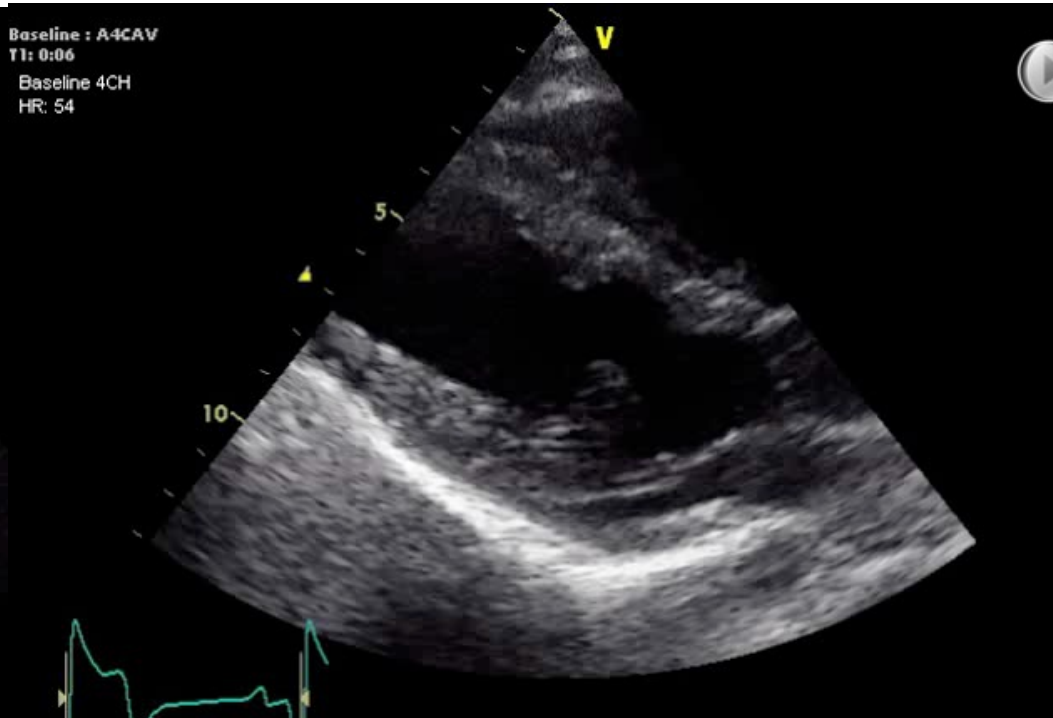
4
6
1
[m/s]
-1
-2
-3
-4
-3
-2
-1
0
L
124
HR


: 2:45
V
5
10


Baseline : A4CAV
T1: 0:06
Baseline 4CH
HR: 54
V
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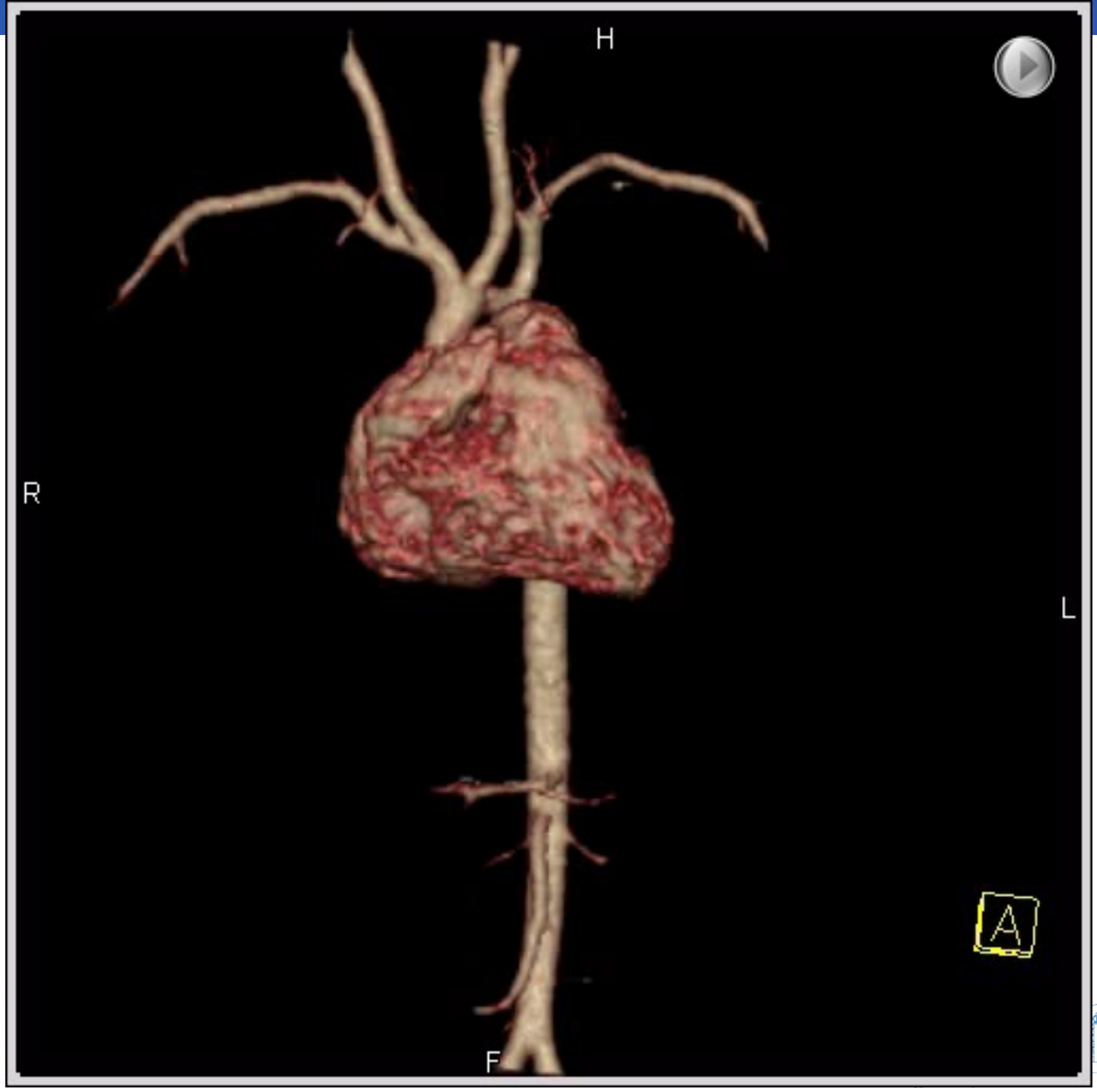
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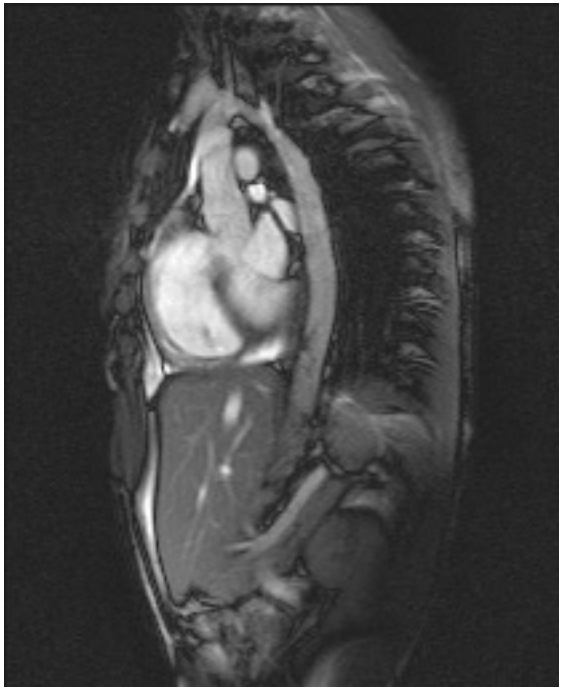
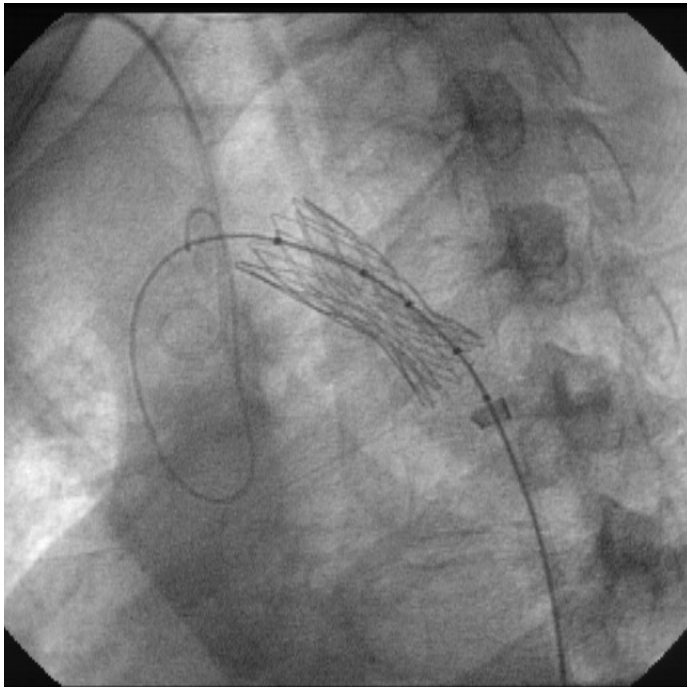
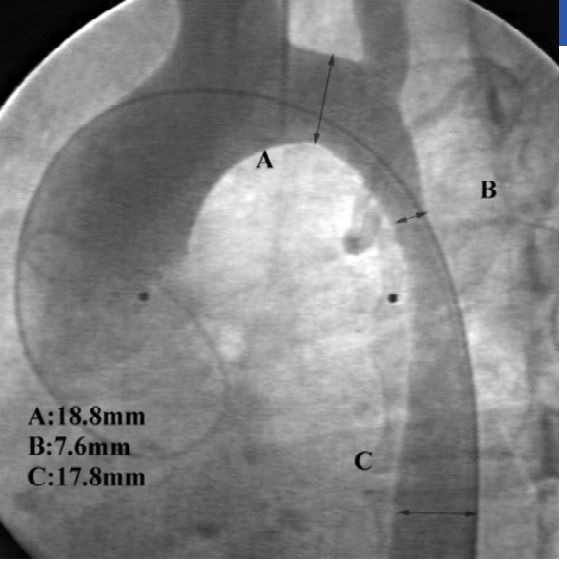
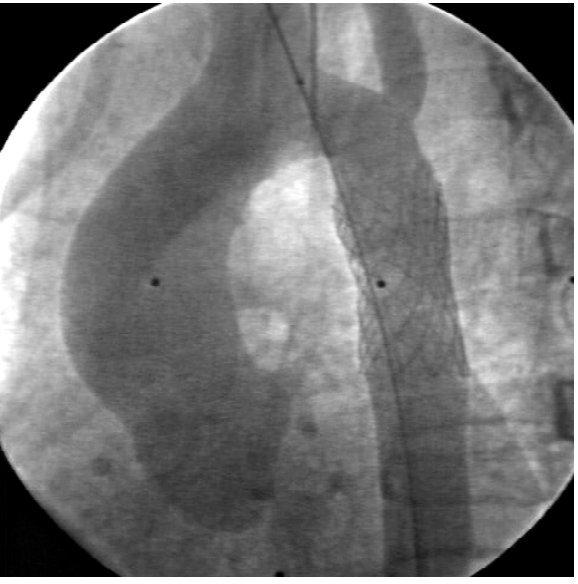
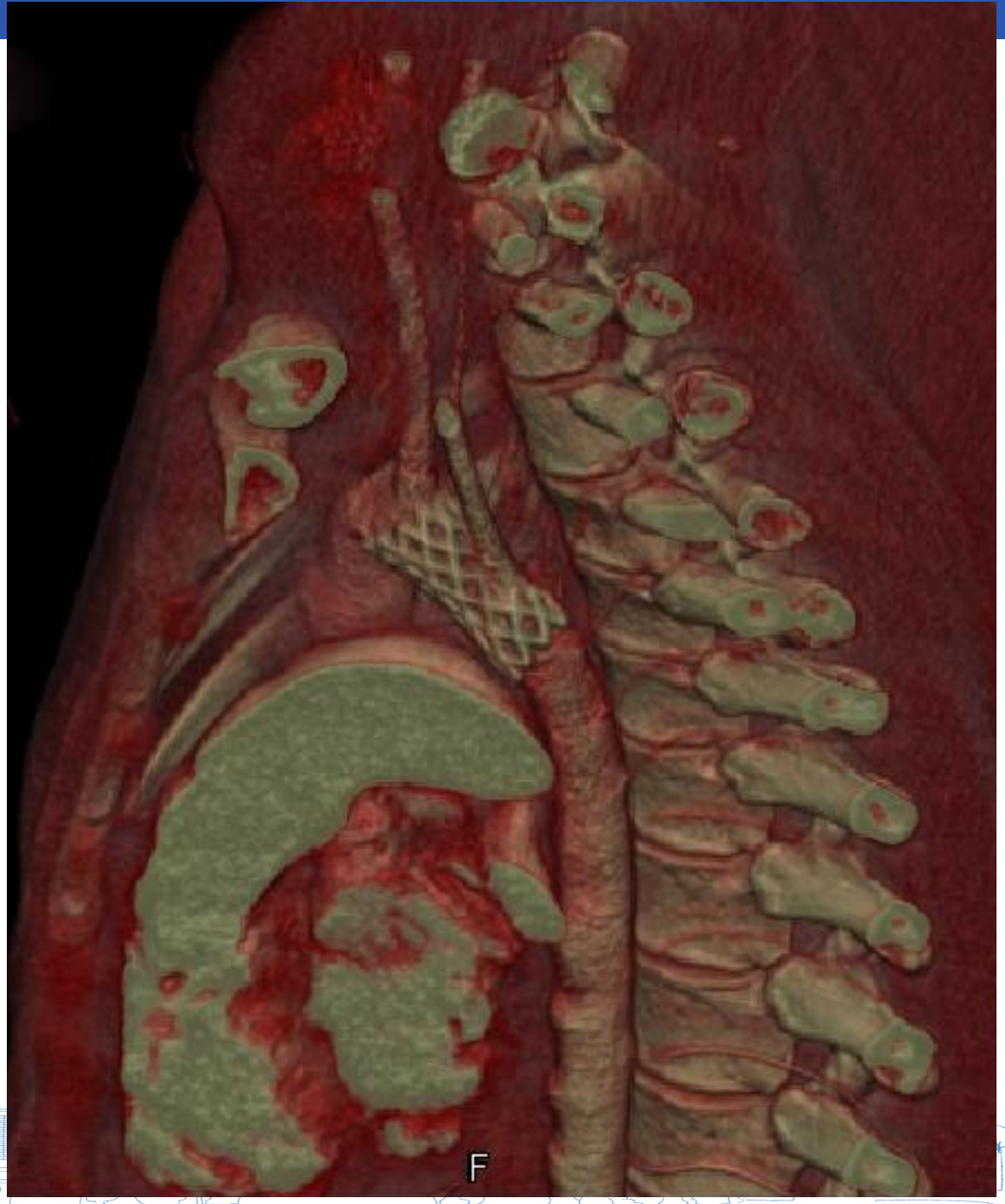
Coarctation de l'aorte native et récidive post opératoire











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Coarctation de l'aorte

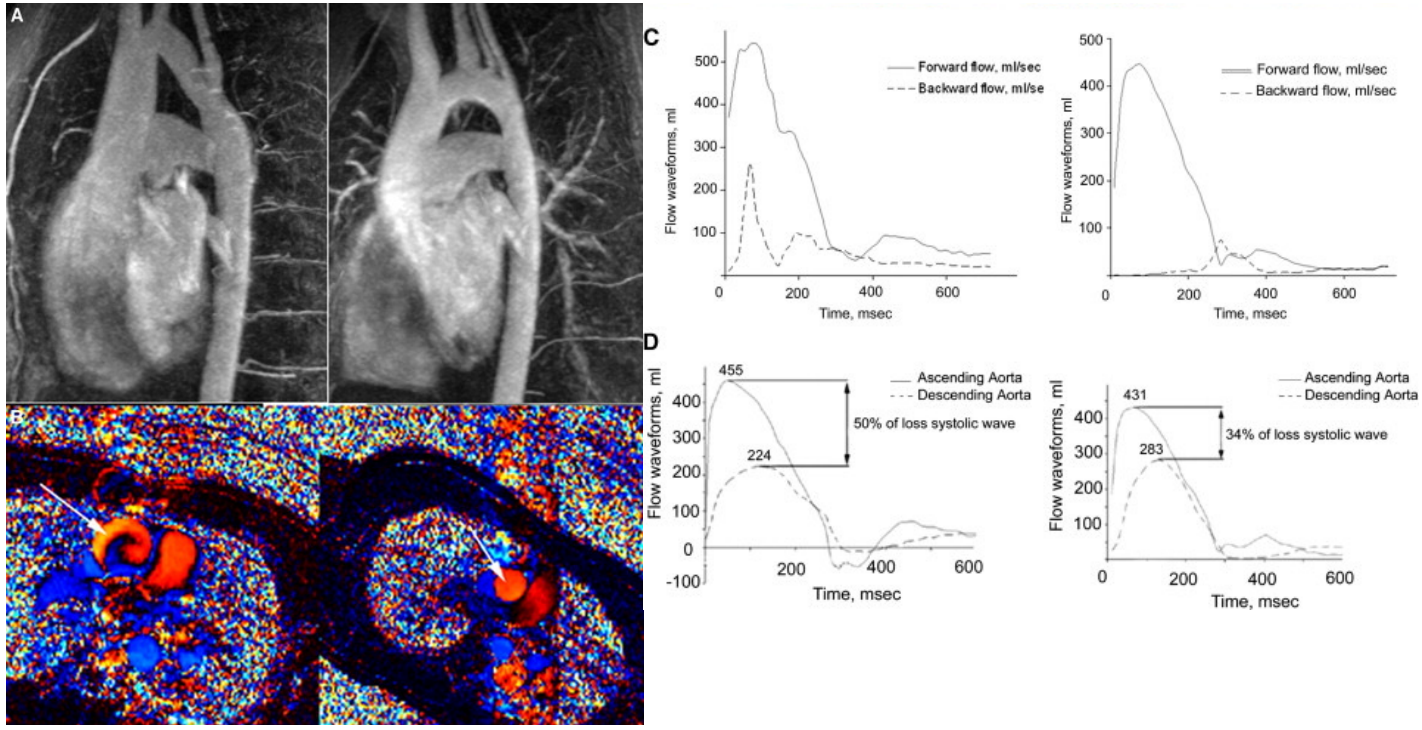
- SEQUELLES ET COMPLICATIONS :
  - Persistance HTA (opération tardive)
  - Recoarctation ou coarctation résiduelle
    - différence TA MS / MI > 20mmHg, HVG
    - aspect rétréci de la zone d'anastomose
    - flux aorte descendante > 3 m / sec,
    - prolongement diastolique ++
- Anomalies associées :
  - bicuspidie aortique (RAO, IAO) (30 - 80 % des cas)
  - autres anomalies du cœur G : mitrales, aortiques
  - Dilatation anévrysmale de l'aorte ascendante

Sport après cure de coarctation de l'aorte

- PTE normal :
  - PAS ≤ 200 mmHg (enfants)
  - PAS ≤ 230 mmHg (adolescents)
    - → Sports dynamiques recommandés

- PTE anormal :
  - PAS ≥200 mmHg (enfants)
  - PAS ≥ 230 mmHg (adolescents)
    - → Sports à faible contrainte dynamique et statique
    - → Réinvestigations ? Traitement ?

Relation between aortic arch geometry and fluid dynamics



A, Gothic *(left)* and Romanesque *(right)* arch. B, Color technique applied in a section of the ascending aorta, which separates the forward flow *(red)* and backward flow *(blue)* during a cardiac cycle.

Ou P. J Thorac Cardiovasc Surg. 2008

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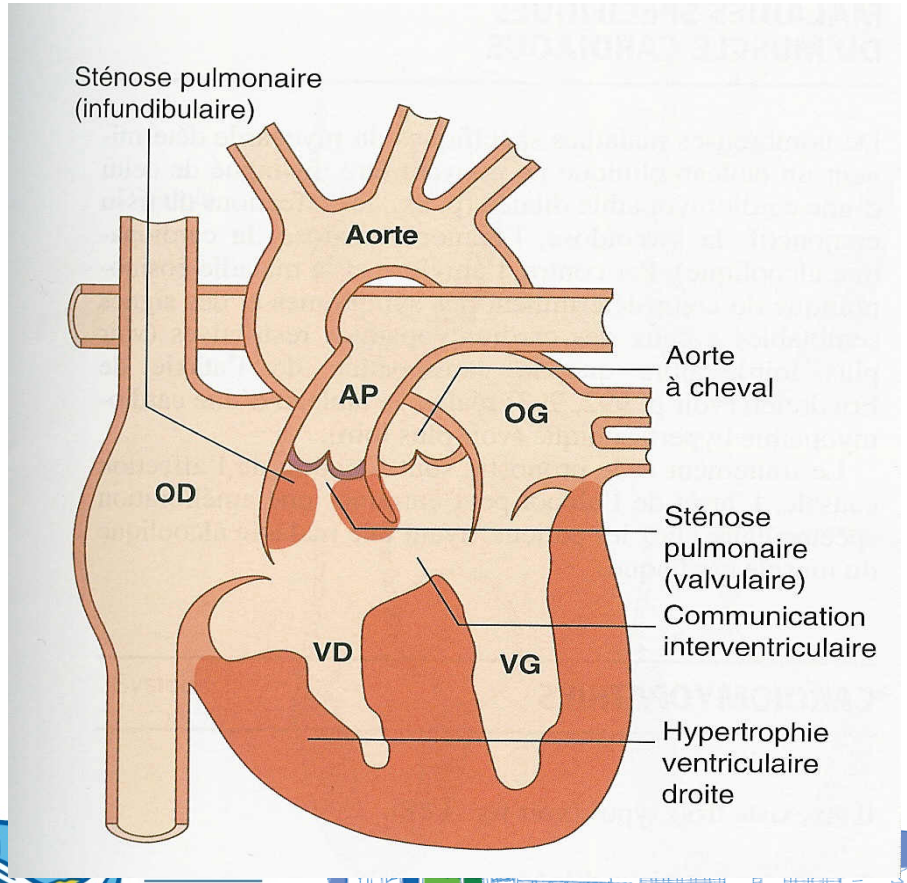
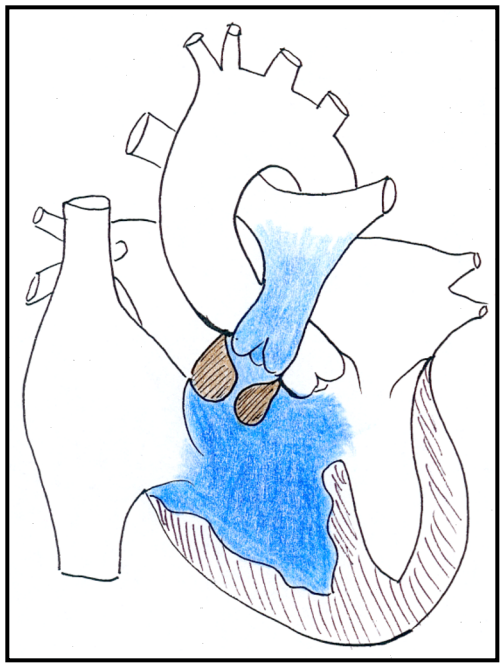
Plan

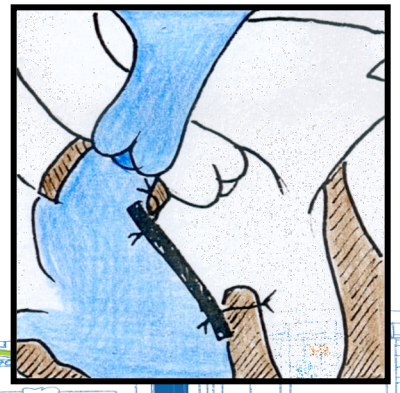
- Cardiopathies complexes opérées

Tétralogie de Fallot

7% des cardiopathies congénitales



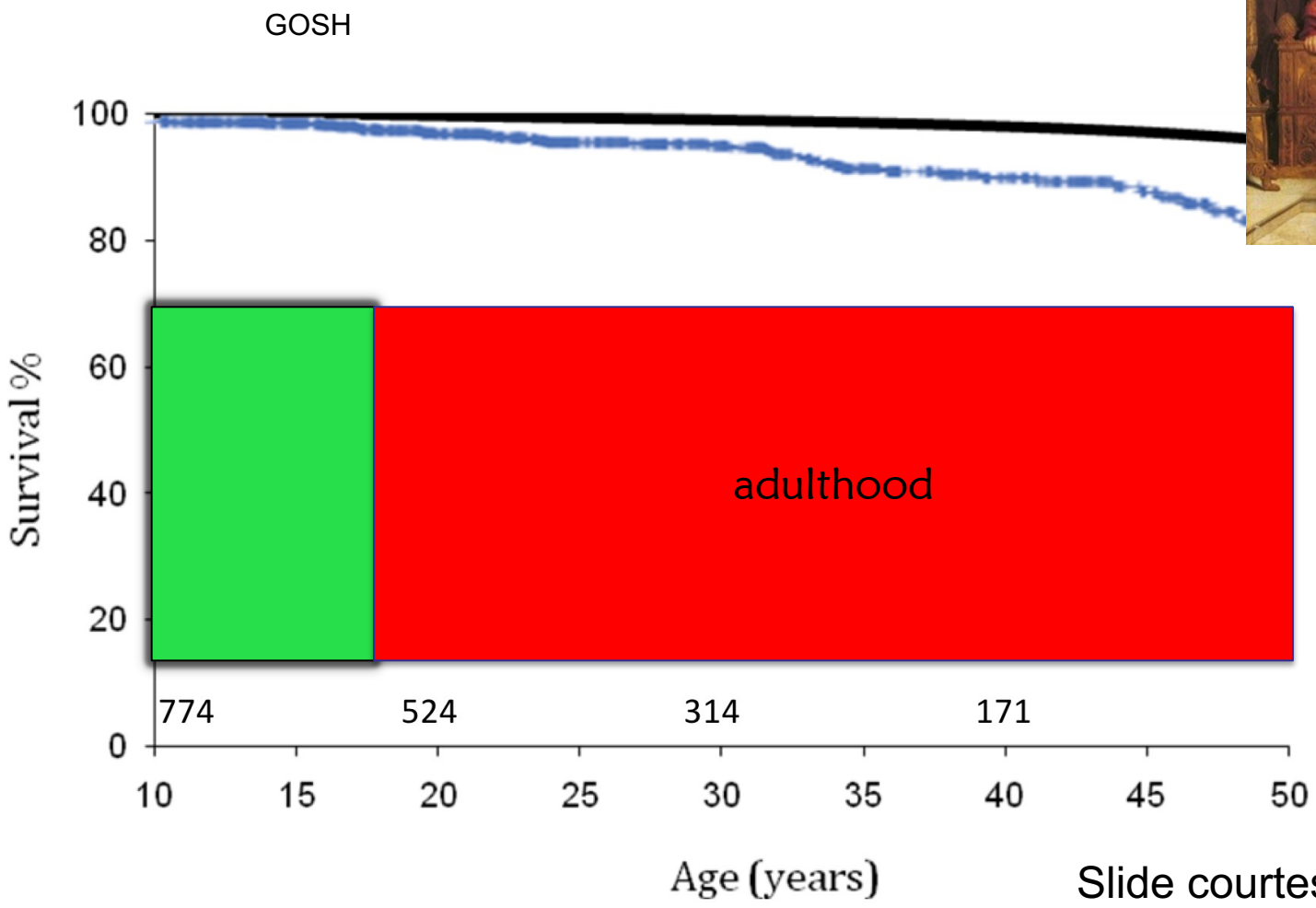




Suivi médico chirugical

- Evaluation de la fonction VD/VG et lien avec les surcharges de volume et de Pression et les lésions associées
- Traitement de la voie droite
- Traitement des lésions associées
- Traitement du risque rythmique
- Traitement électrique de l'insuffisance cardiaque

Why follow-up is important?

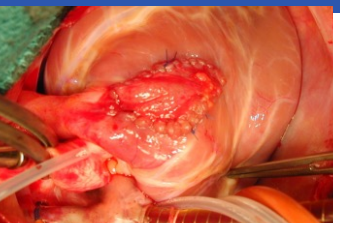


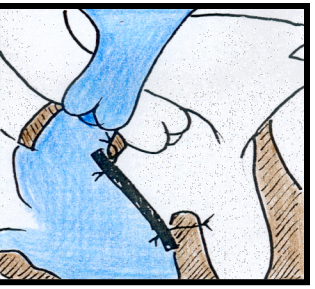
Slide courtesy of A Frigiola

Arythmies

- ⅓ to ½ of deaths in adult TOF are sudden
- 100% of TOF pts with sudden death has moderate or severe PR
- QRS > 180 ms &/or an increase in QRS duration
  > 3.5 ms/year predicts ventricular arrhythmia and sudden death

Repaired T4F pathophysiology



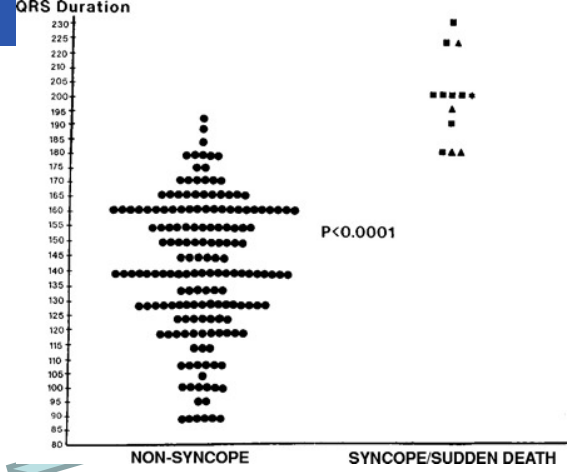


PA branch stenosis

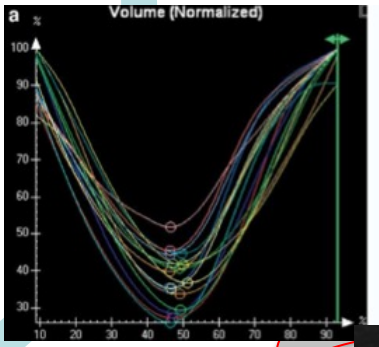
PR

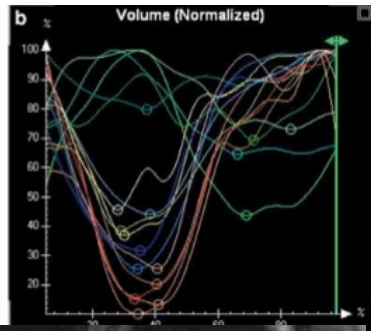
Chronic volume overload

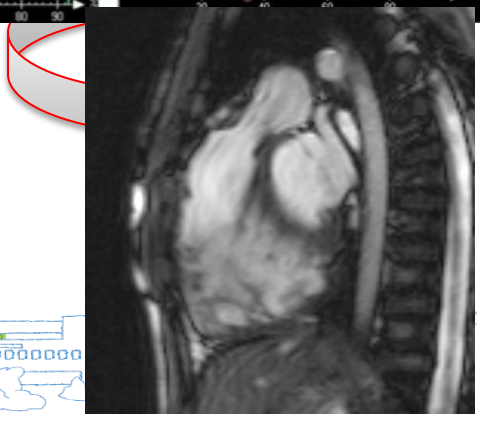
RV dilatation



RV/LV Interaction
-asynchronism
- P/V curve change



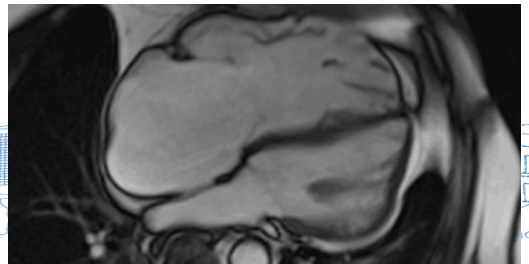


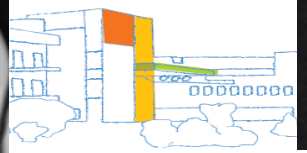


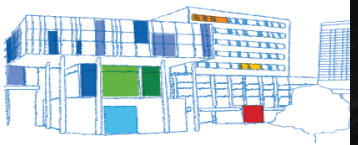
TR

RA Dilatation

atrial arrythmia







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Effects of chronic volume overload

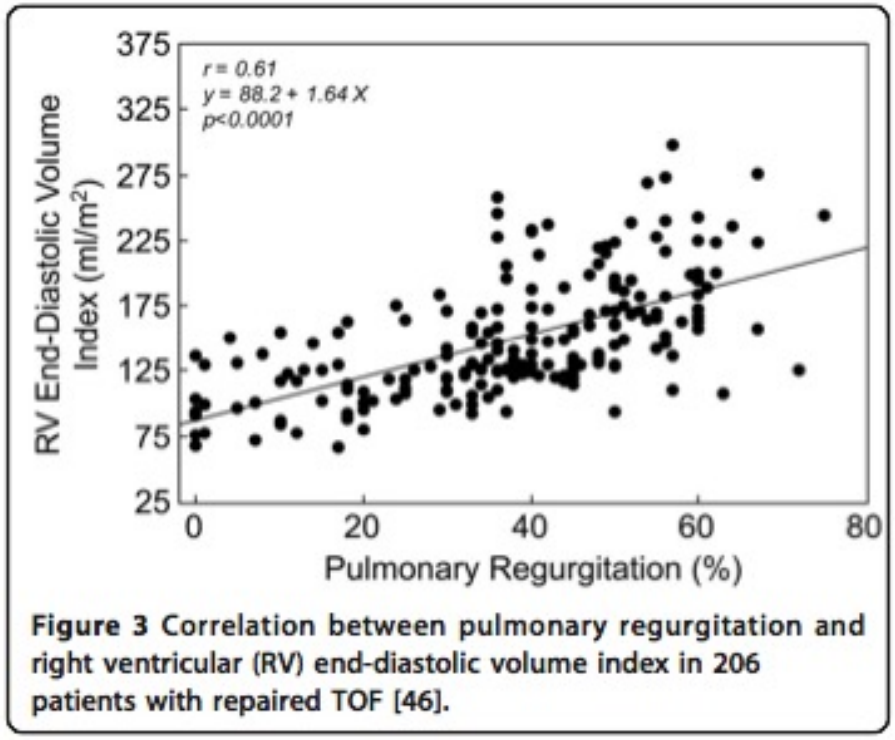


Figure 3 Correlation between pulmonary regurgitation and right ventricular (RV) end-diastolic volume index in 206 patients with repaired TOF [46].

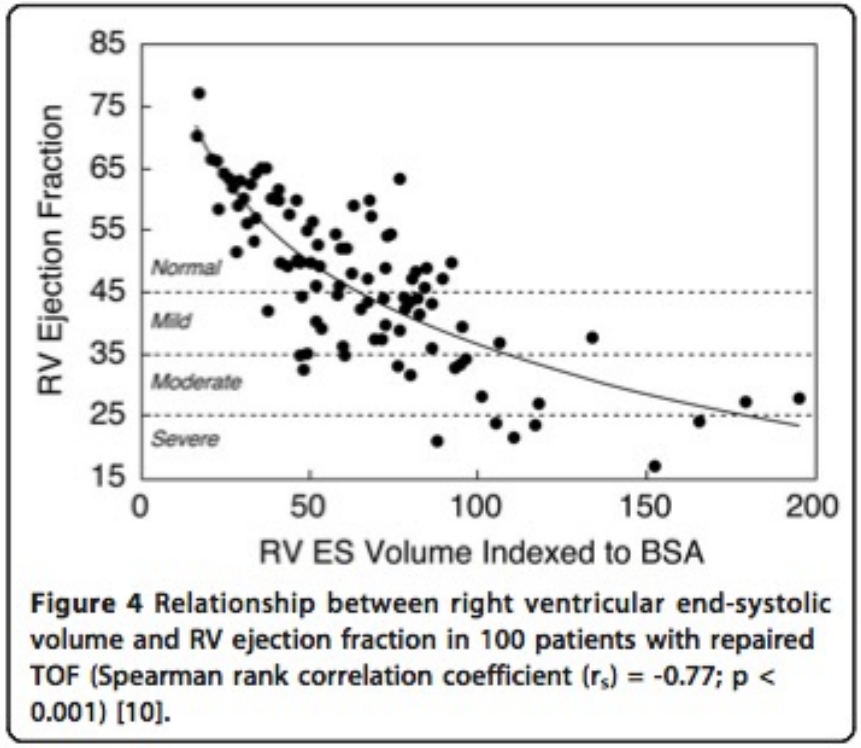


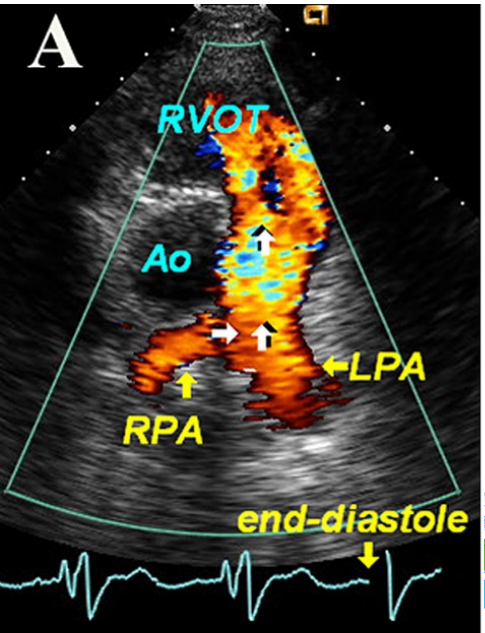
Figure 4 Relationship between right ventricular end-systolic volume and RV ejection fraction in 100 patients with repaired TOF (Spearman rank correlation coefficient (r_s) = -0.77; $p < 0.001$) [10].

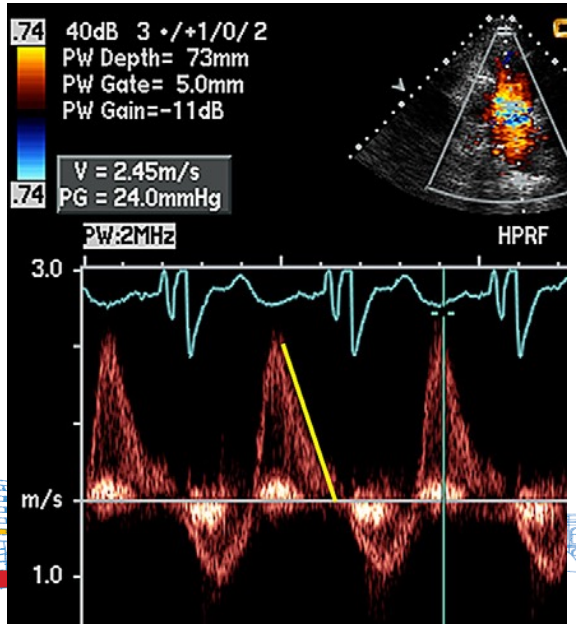
Samyn et al, J Magn Reson Imaging 2007

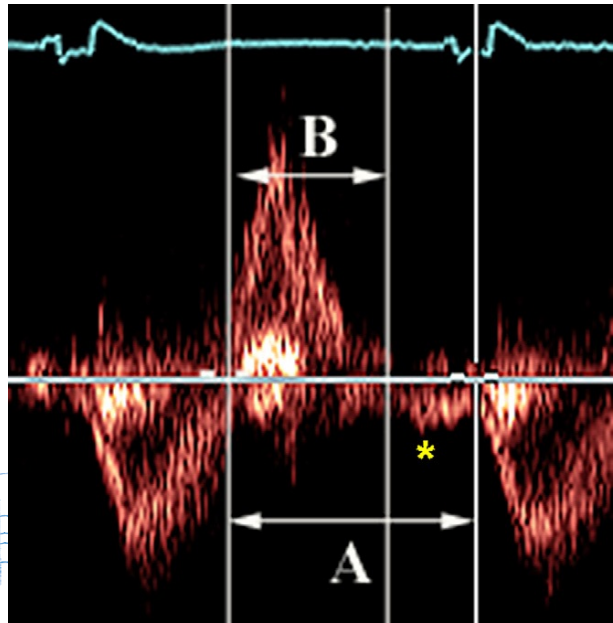
Geva et al, J Am Coll Cardiol 2004,

PR evaluation

	Mild PR	Moderate PR	Severe PR
diastolic reversal flow	Pulm annulus	Pulm trunck	Pulmonary Branches
Jet width/ RVOT Ø	<1/3	1/3-2/3	> 2/3
PHT			<100ms
Duration PR/ duration of diastole			>0.77

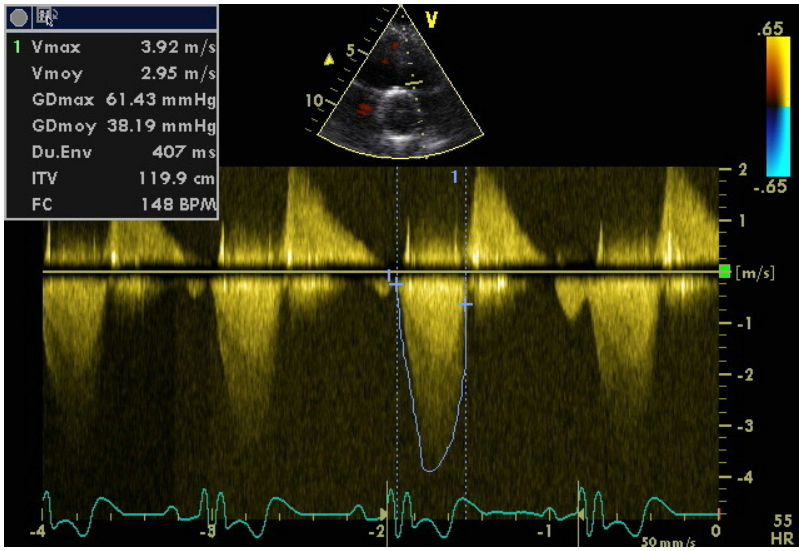






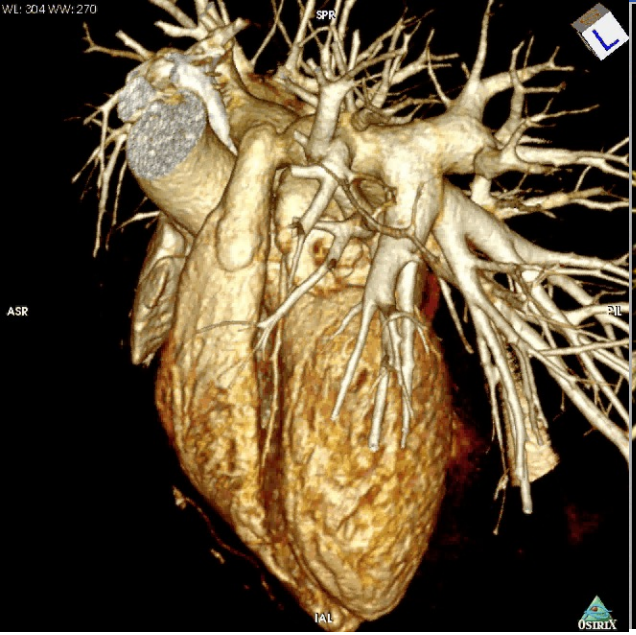
Pulmonary stenosis

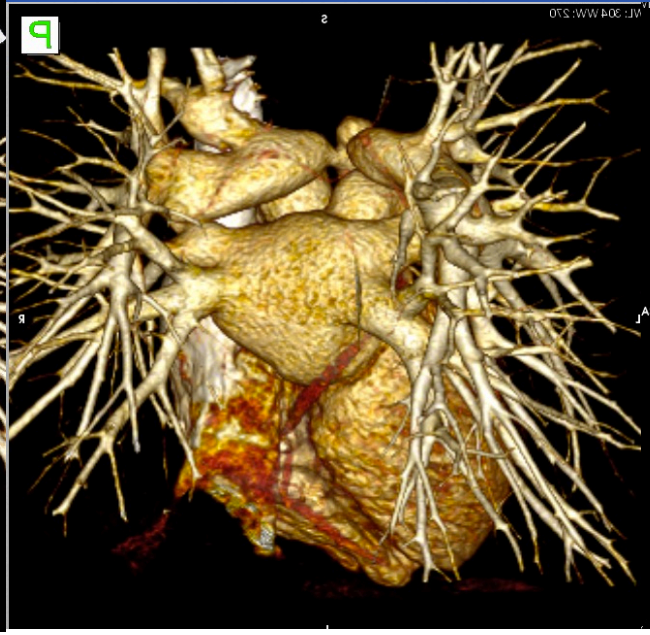
- Continuous-wave doppler estimates
  - RVOTO
  - Peripheral branch stenosis



  - May limited by
    - Suboptimal angle
    - Suboptimal acoustic window
    - Distal stenosis

- Complementary imaging modalities are usefull
  - MRI, CT
  - Angio


WL: 304 WW: 270
SPR
ASR
IAL
OSIRIX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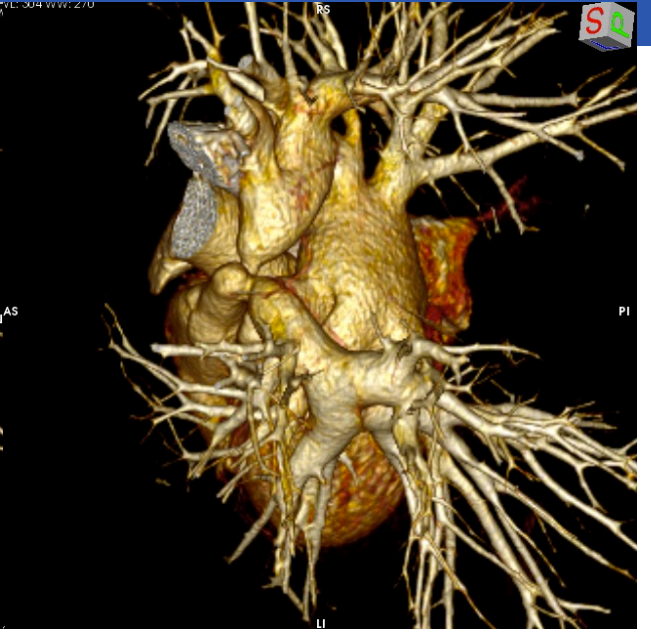
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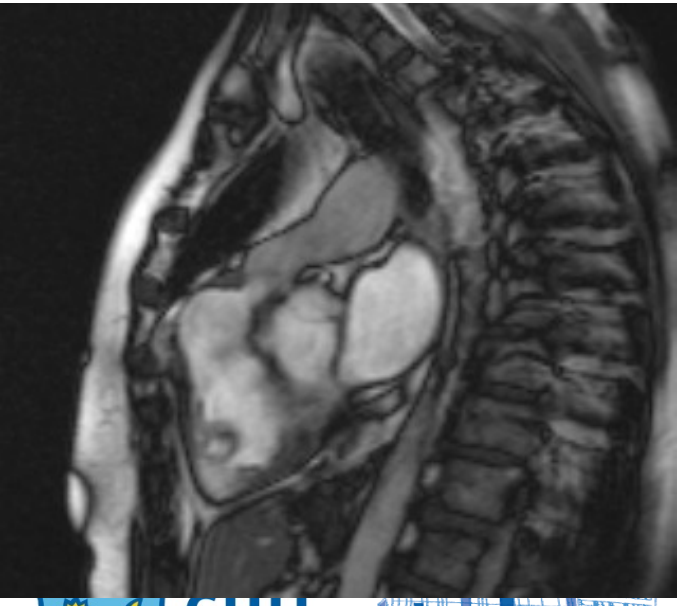
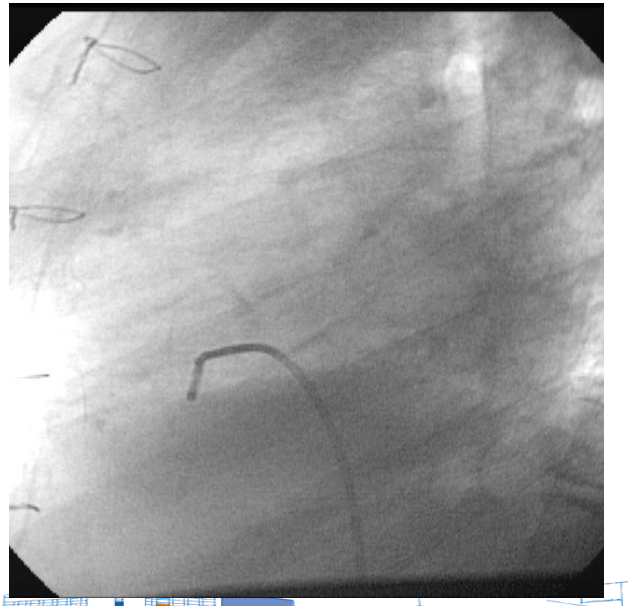




Apport de l'imagerie en coupe

-analyse des structures extracardiaques

-quantification des volumes et des fonctions VG/VD (gold standard)

-analyse des zones de fibrose

-quantification des valvulopathies et des shunts

Indication de valvulation pulmonaire des Fallot opérés:
-VTD>150ml/m²
-Anévrysme infundibulaire>40mm
-Altération de la fonction VD

Therrien J. Am J Cardiol 2005

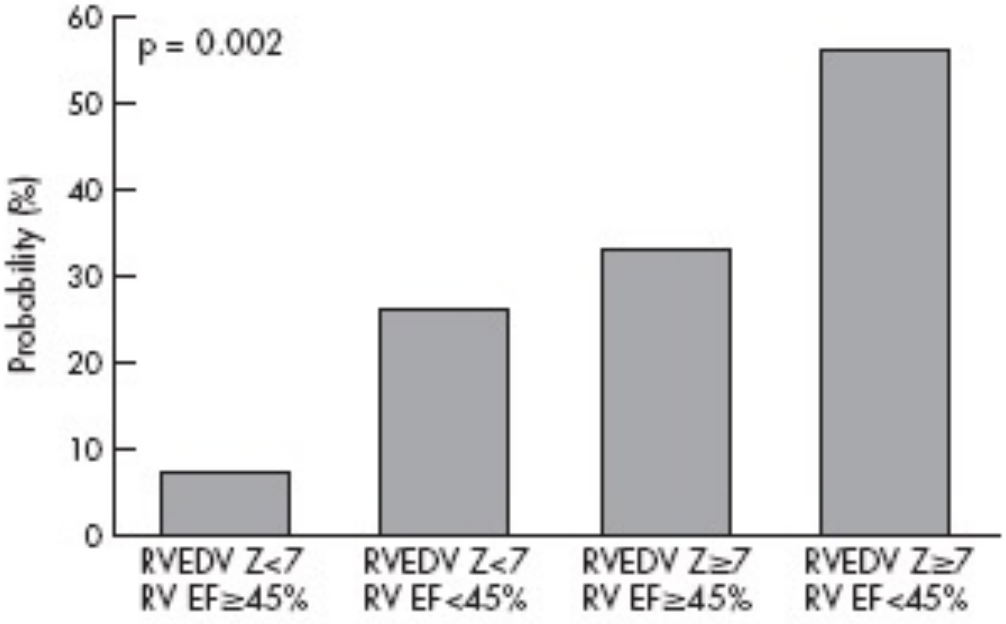
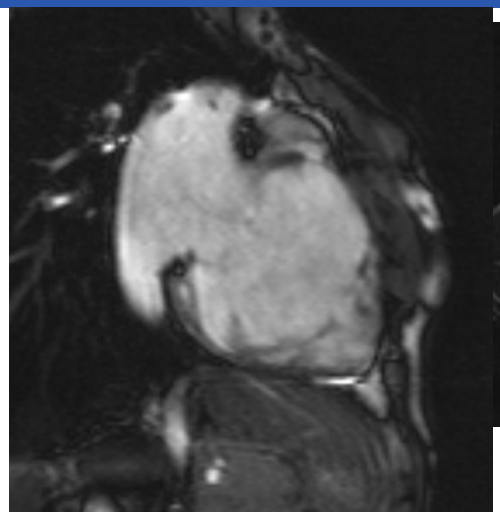


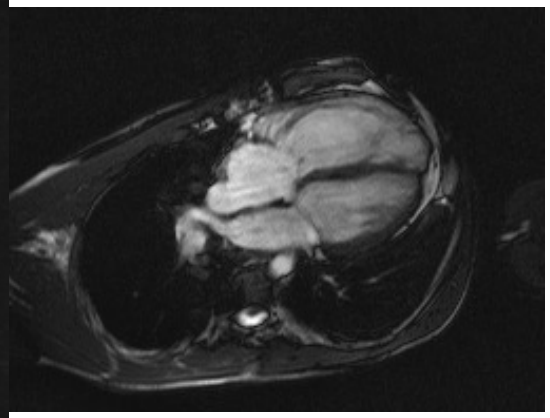
Figure 1 Probability of major adverse clinical outcomes based on multivariate logistic regression model containing right ventricular end-diastolic volume (RVEDV) Z-score and right ventricular ejection fraction (RV EF).

Ventricular size and function assessed by cardiac MRI predict major adverse clinical outcomes late after tetralogy of Fallot repair

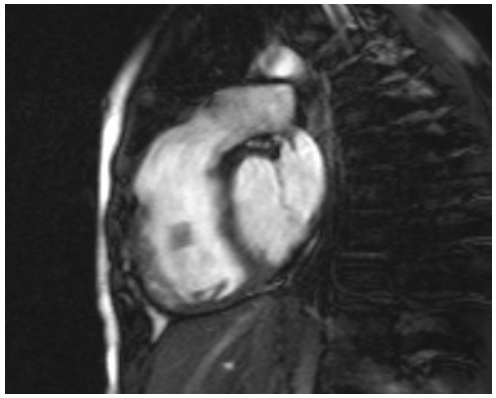
Knauth AL et al *Heart* 2008;94:211-216

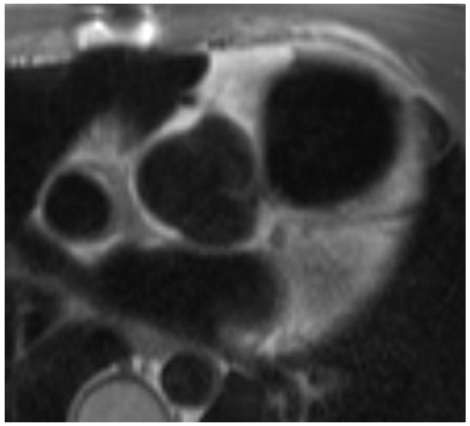
- Fonction et volumes



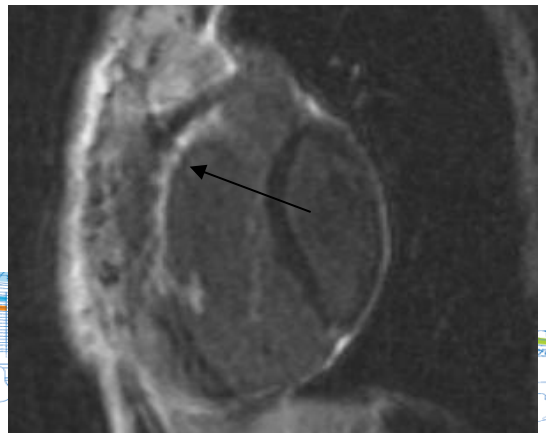


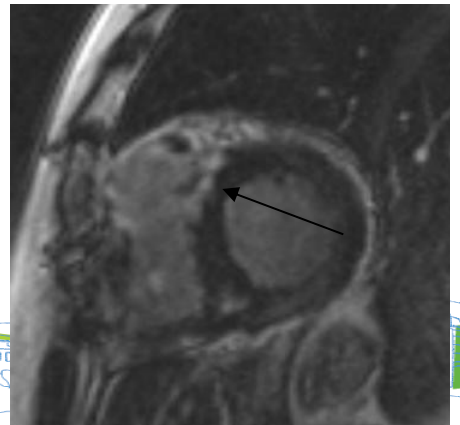
- Morphologie et mesure de l'anévrysme infundibulaire





- Fibrose analyse du réhaussement tardif





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Indications for PVR

I Asymptomatic pts with 2 or > of the following criteria

-RVEDV >150 ml/m2 or RV/LV EDV volume >2

-RV ESV >80 ml/m2

-RV EF <47%

-LV EF <55%

-QRS duration >140 ms, sustained tachyarrhythmia

II Hemodynamically significant lesions

-RVOTO with RV systolic pressure 2/3 systemic

-Severe branch PAs stenosis

-Moderate TR

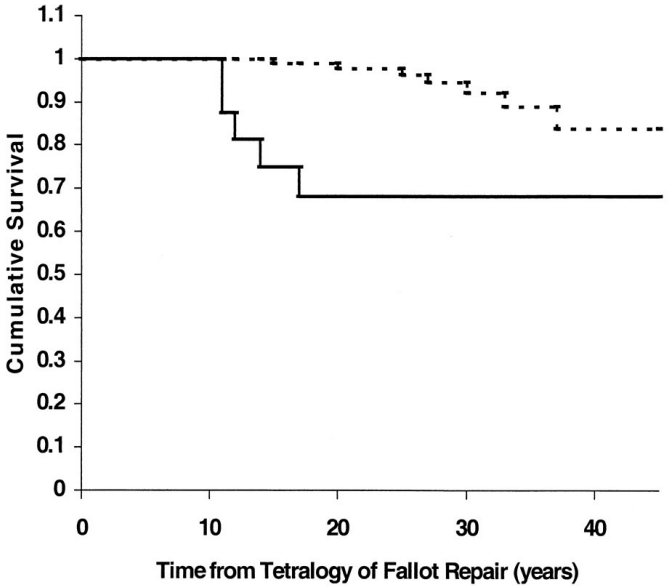
-L to R shunt with Qp:Qs ≥1.5

-Severe AR

-Severe aortic dilatation (diameter 5 cm)

Tétralogie de Fallot

Left ventricular dysfunction is a risk factor for sudden cardiac death in adults late after repair of tetralogy of fallot



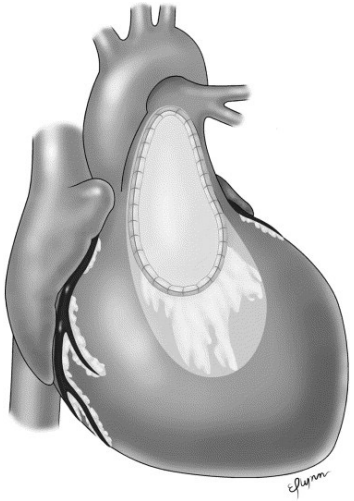
Normal or mild LV systolic dysfunction (n)	121	121	119	116	114
Moderate or severe LV systolic dysfunction (n)	16	16	11	11	11

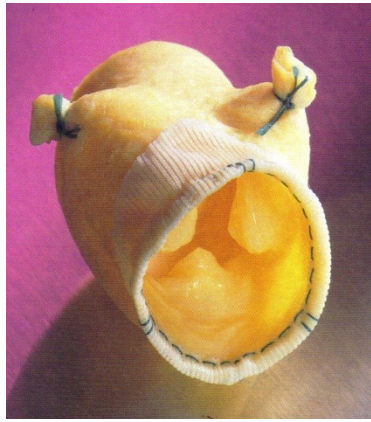
Kaplan-Meier survival curve beyond the age of 18

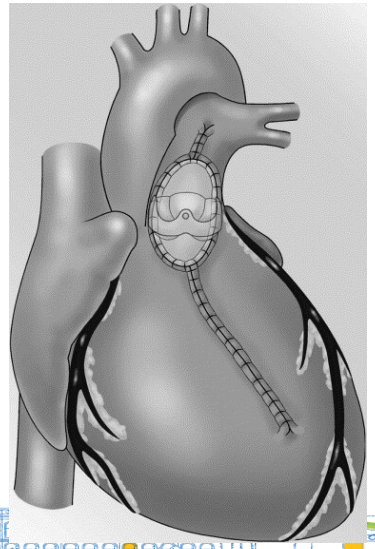
Ghai, A. et al. J Am Coll Cardiol 2002;40:1675-1680

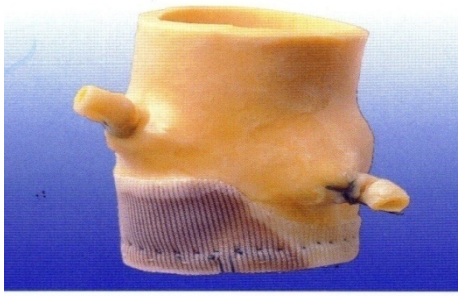
Place de la chirurgie

- Technique chirurgicale :
  - Remplacement valvulaire biologique
  - Gestes associés
    - TAP
    - IT
    - infundibulum
    - TDR
    - CIV r
    - Dilatation aorte



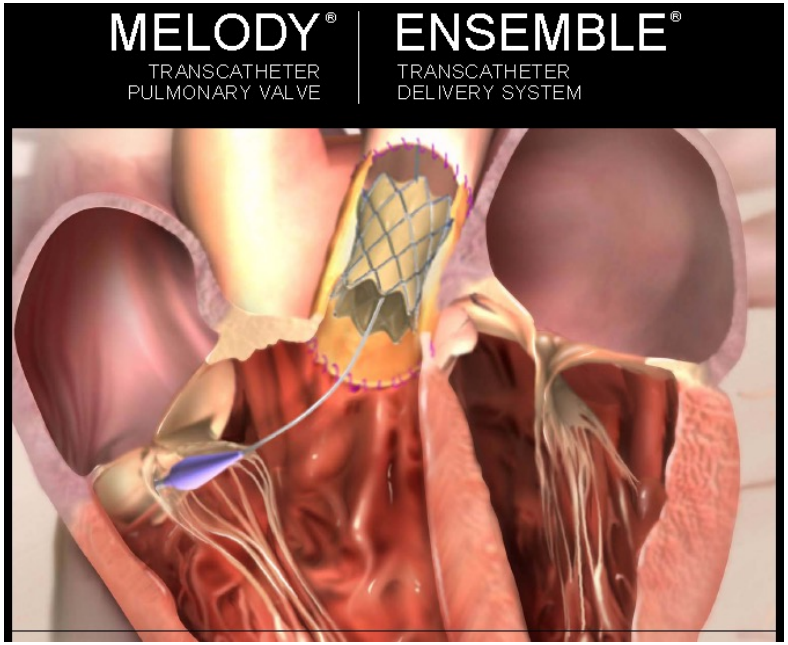


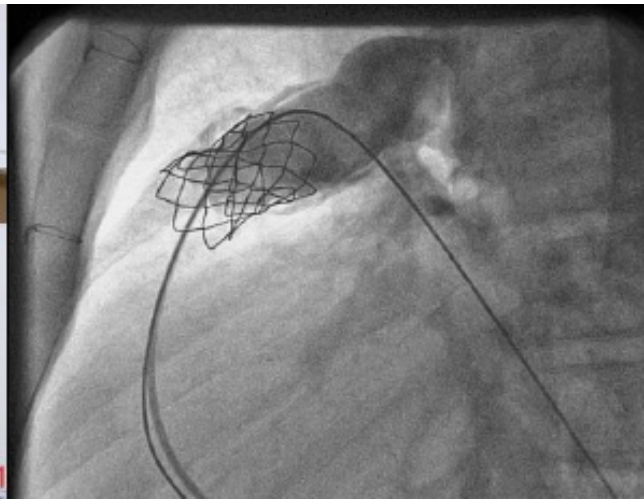


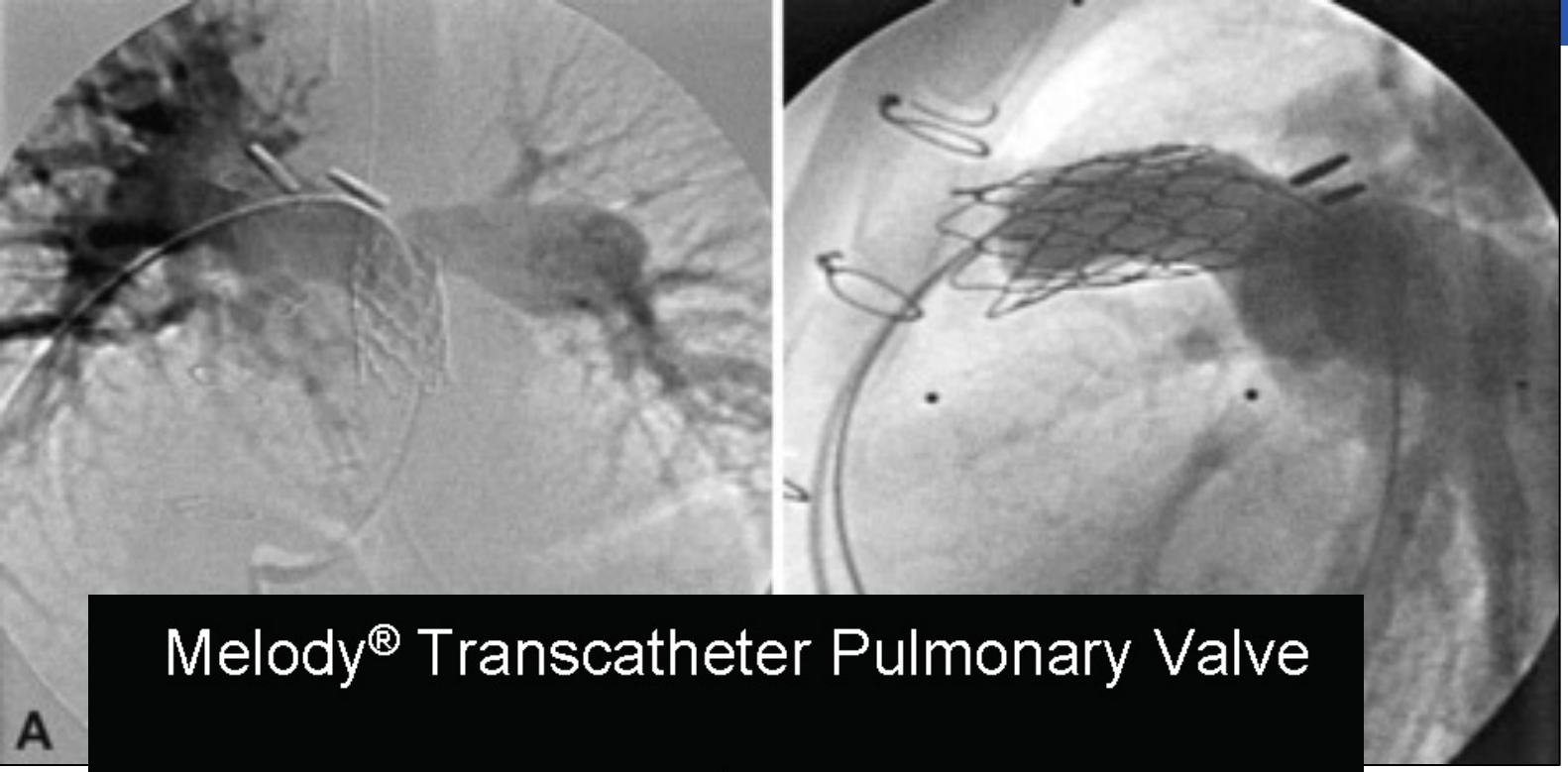


Vavulation percutanée: « melody »

- Indication:
  - Homogreffe ou tube VD-AP
- Limite
  - 18< Ø < 22 mm

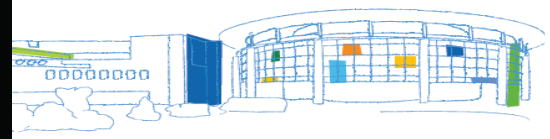






Melody® Transcatheter Pulmonary Valve

- 18mm Contegra modified-bovine jugular vein with valve segment
- Balloon Expandable system
- NuMed Platinum Iridium Stent
  - 28 mm length
  - Crimped down to 6mm, re-expanded 18mm up to 22mm



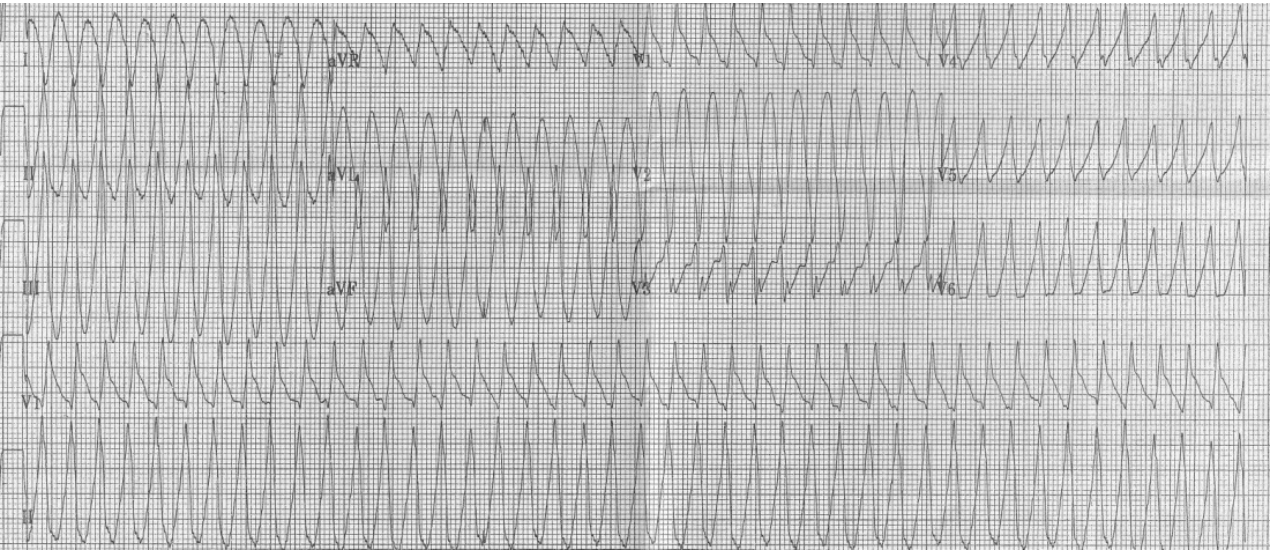
VT in rTOF

Tétralogie de Fallot

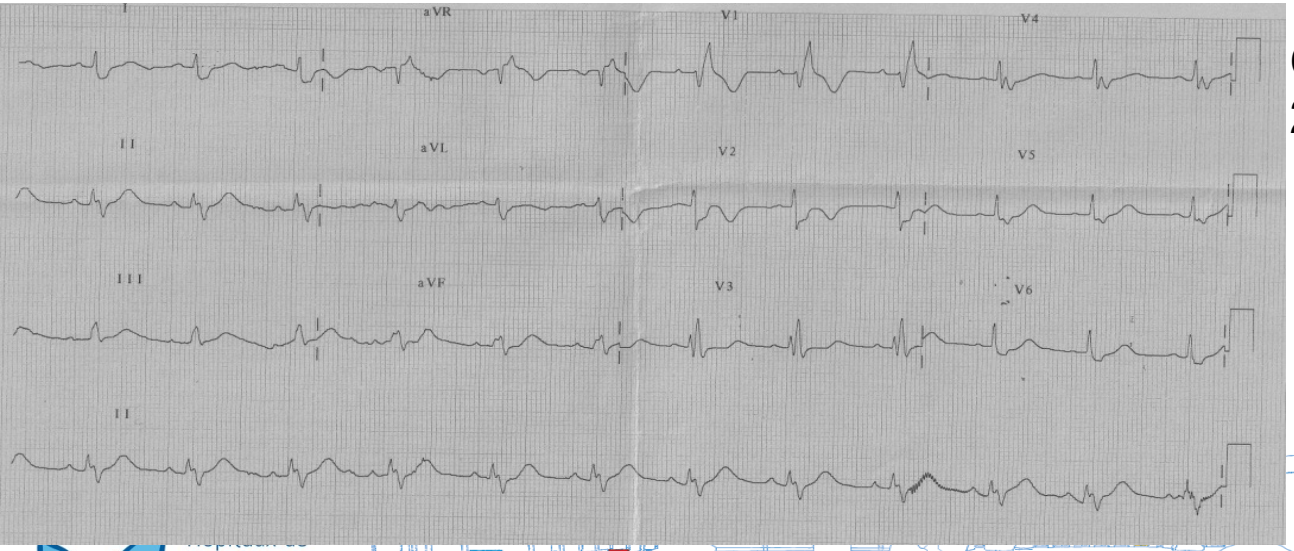
Cas clinique

-Jeune femme 21 ans
-Correction complète à 9 mois
-Suivi dans l'enfance
-QRS 170ms , ETT: VD dilaté IP III
-Absence de suivi depuis 10 ans
-1 Syncope en 2007 non explorée
- Hospitalisation en urgence pour malaise sans PCI

Tétralogie de Fallot



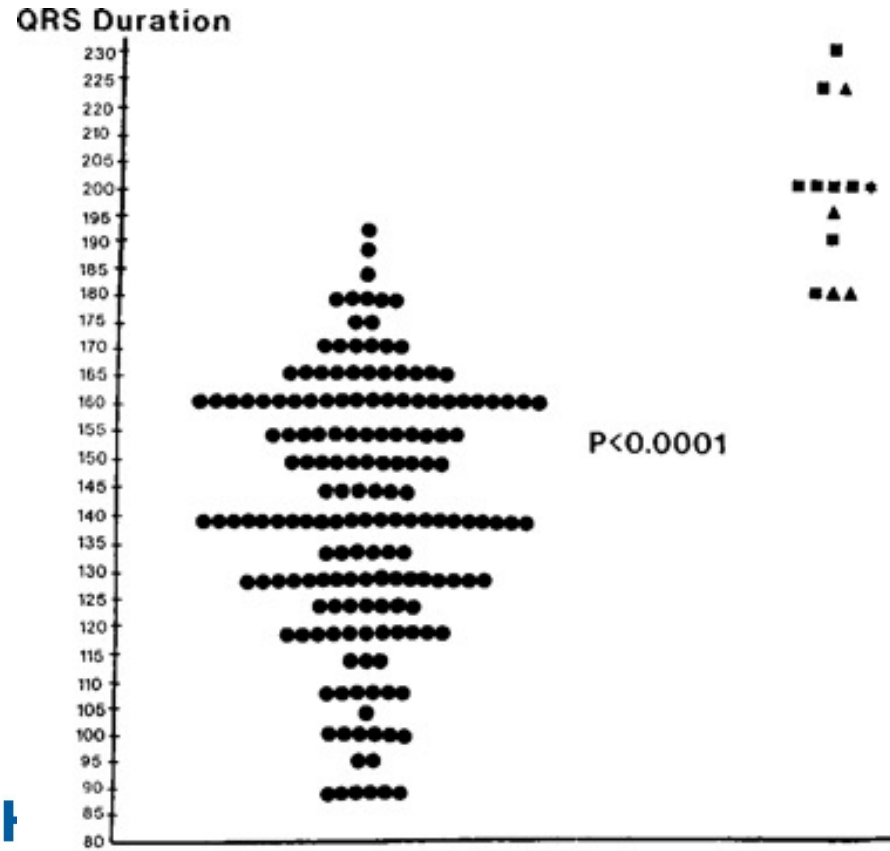
Hémodynamique instable
TA 80/40



Cardioversion sous AG
200J retour rythme sinusal

Tétralogie de Fallot

Corrélation largeur du QRS et mort subite



Gatzoulis Circ 1995

Tétralogie de Fallot

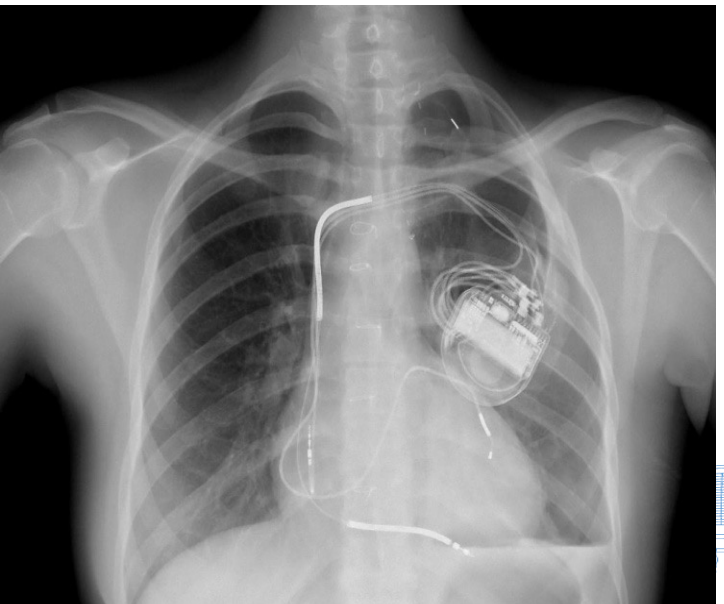
- Prise en charge rythmique : indication du DAI

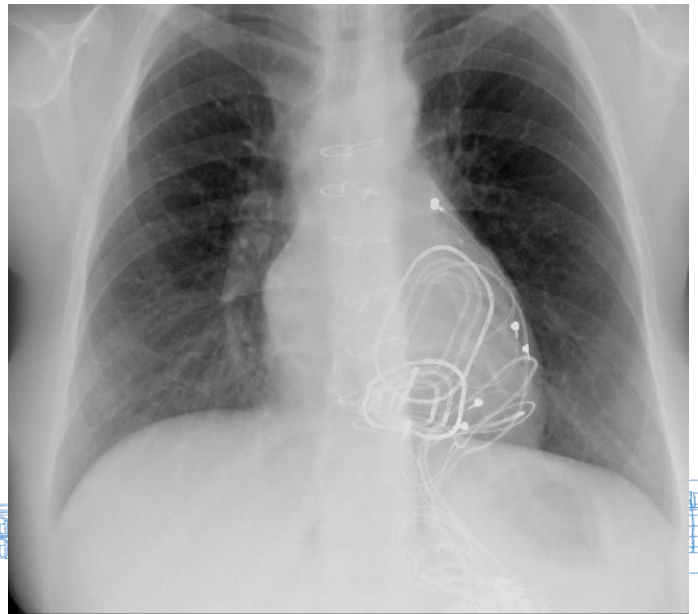
Congenital Heart Disease

Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;
Sangeetha Viswanathan, MRCPCH; Amanda Barlow, MD; Michael A. Gatzoulis, MD;
Susan M. Fernandes, MHP, PA-C; Luc Beauchesne, MD; Judith Therrien, MD; Philippe Chetaille, MD;
Elaine Gordon, MD; Isabelle Vonder Muhll, MD; Frank Cecchin, MD

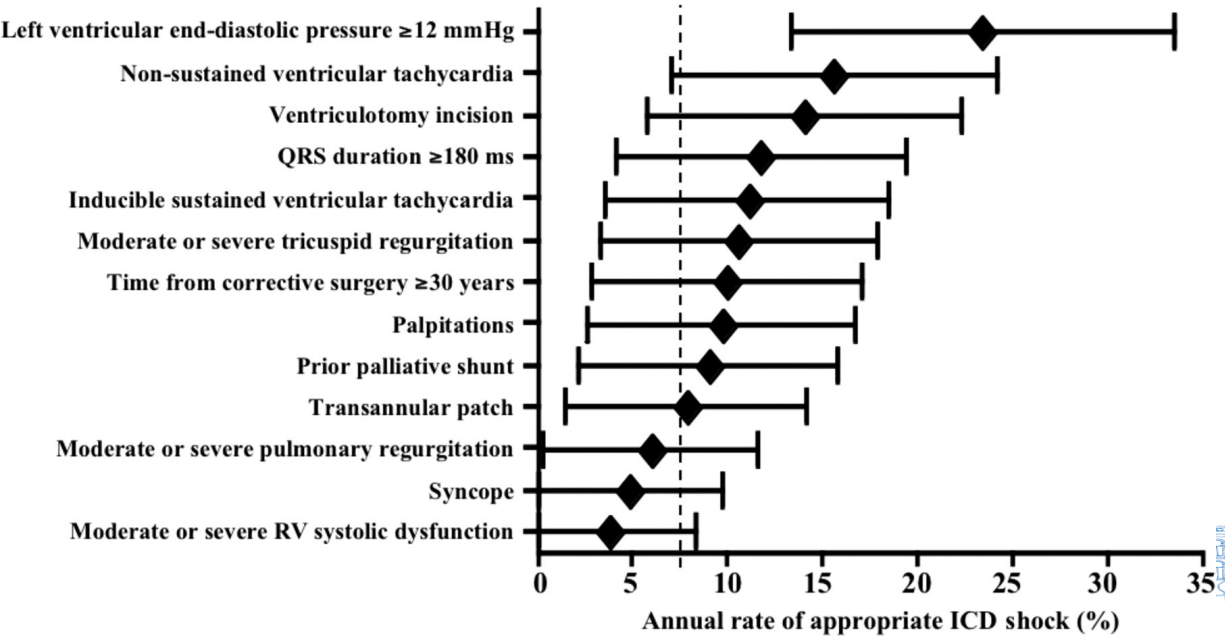
Khairy, P. et al. Circulation 2008;117:363-370



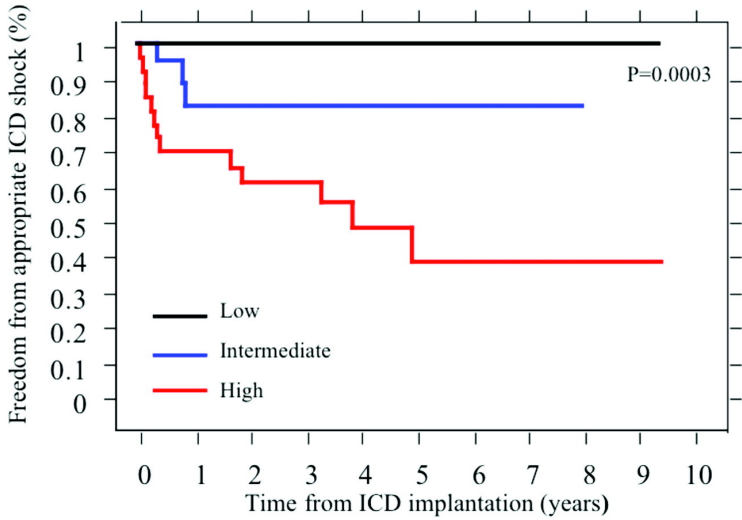


Risk Score for Appropriate ICD Shocks in Primary Prevention

Prior palliative shunt		2
Inducible sustained ventricular tachycardia		2
QRS duration	180 ms	1
Ventriculotomy incision		2
Nonsustained ventricular tachycardia		2
LVEDP	12 mm Hg	3
Total points		0–12

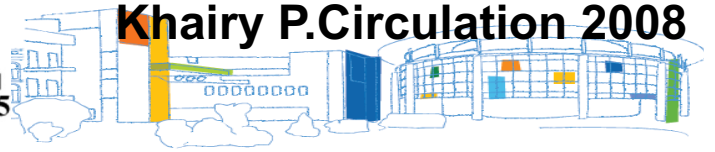


Freedom from appropriate ICD shocks in primary prevention according to risk category



Risk score	Risk category	N	Annualized rate of appropriate shocks
0-2	Low	18	0%
3-5	Intermediate	24	3.8%
6-12	High	26	17.5%

Khairy P.Circulation 2008

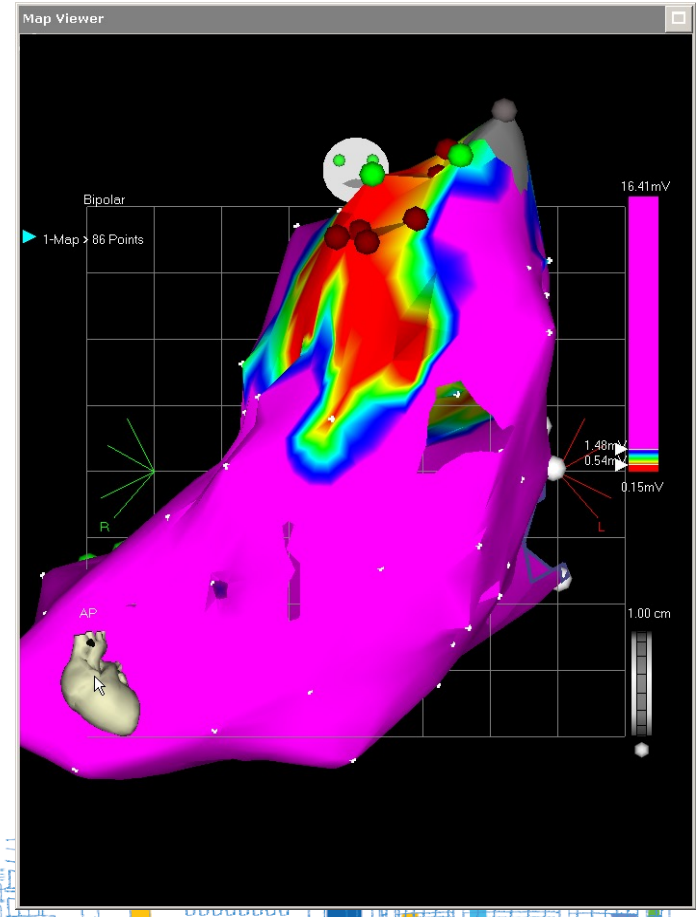


Tétralogie de Fallot

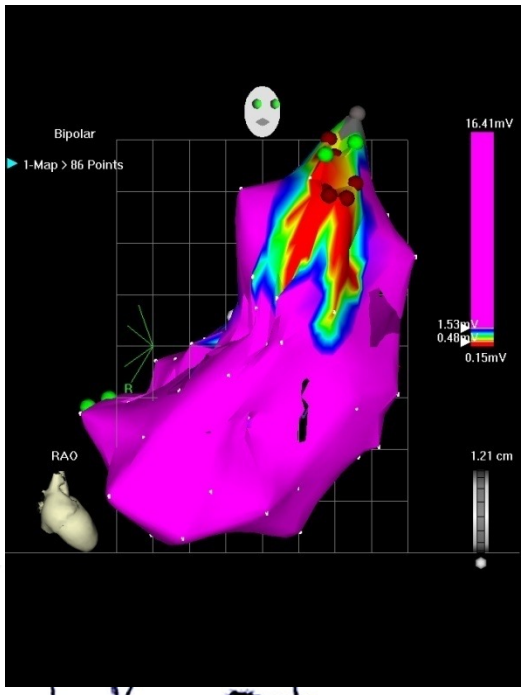
- Prise en charge rythmique: indication de l'ablation

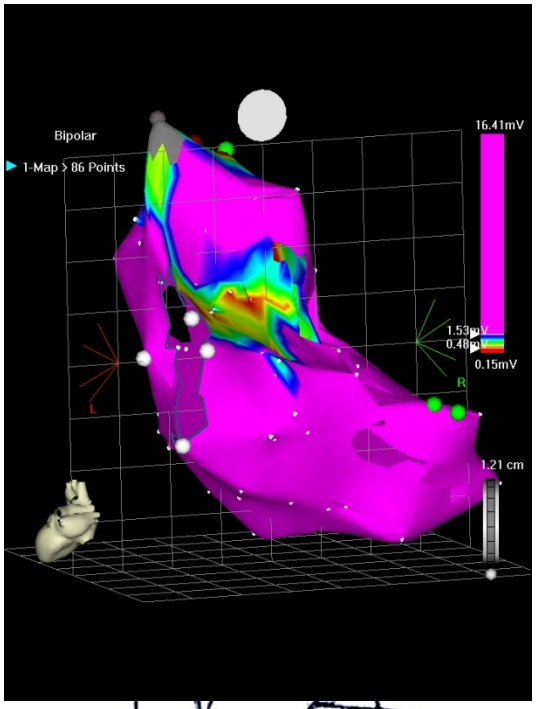
Mapping endocavitaire
et ablation par radiofréquence

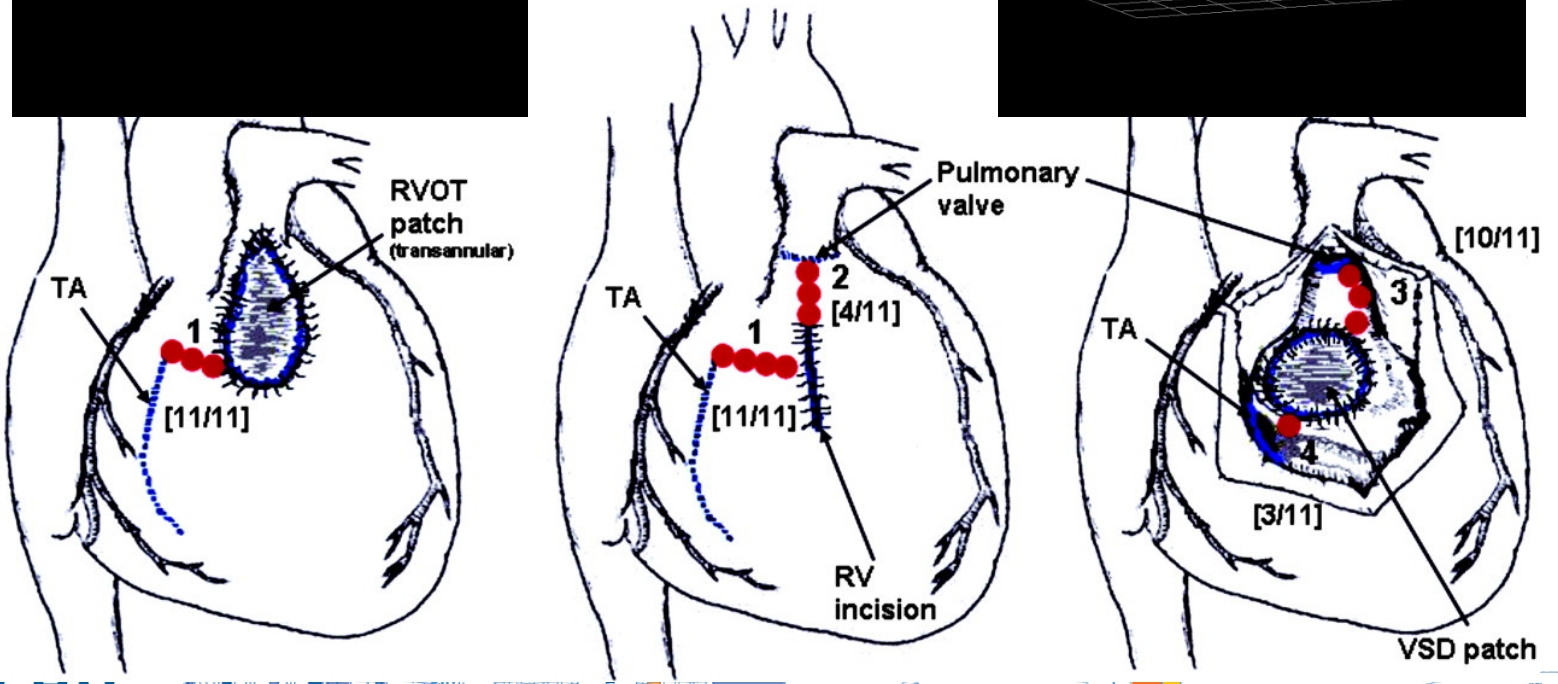
Indication:
-TV récidivante sous traitement médical
-traitement complémentaire du DAI
- Bilan systématique avant RVP (possibilité de TTT chir complémentaires)
- Traitement de première intention au CHU de Bordeaux +++++



CHU Hôpitaux de Bordeaux





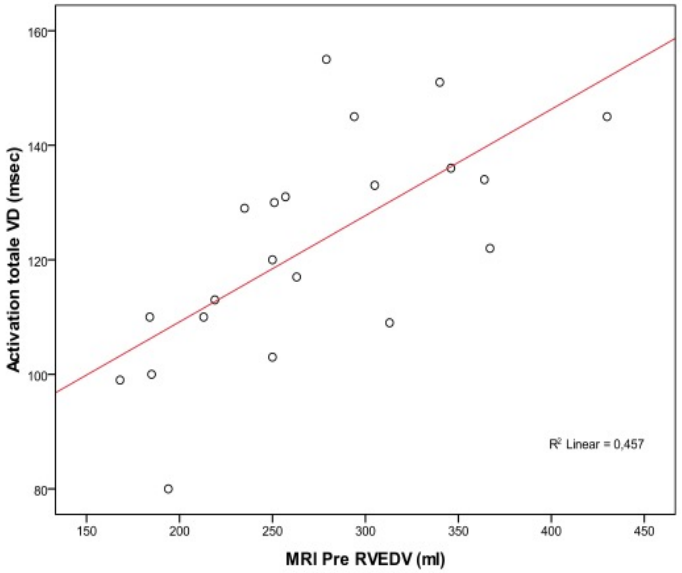


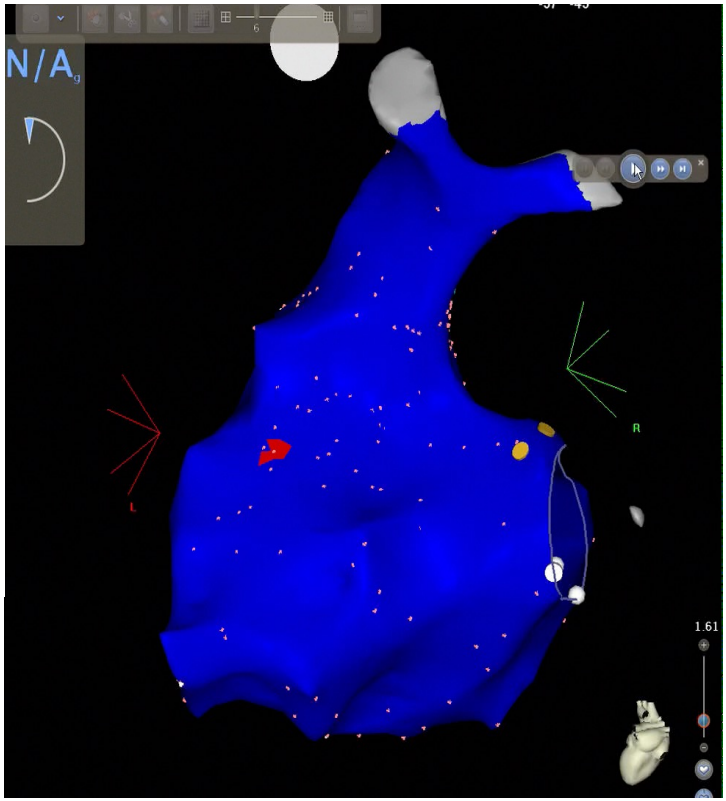
Zeppenfeld, K. et al. Circulation 2007;116:2241-2252

CRT

Endocardial RV activation mapping

- Avantage of end
  - Septal activa
- Activation patt
  - Starts from b
  - Ends on the
  - Late activatic



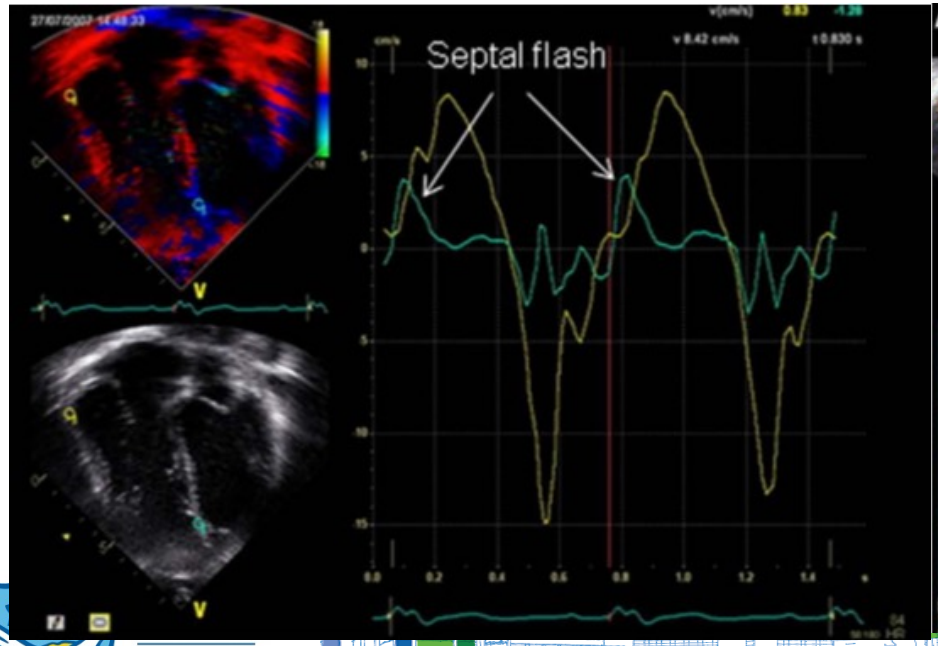


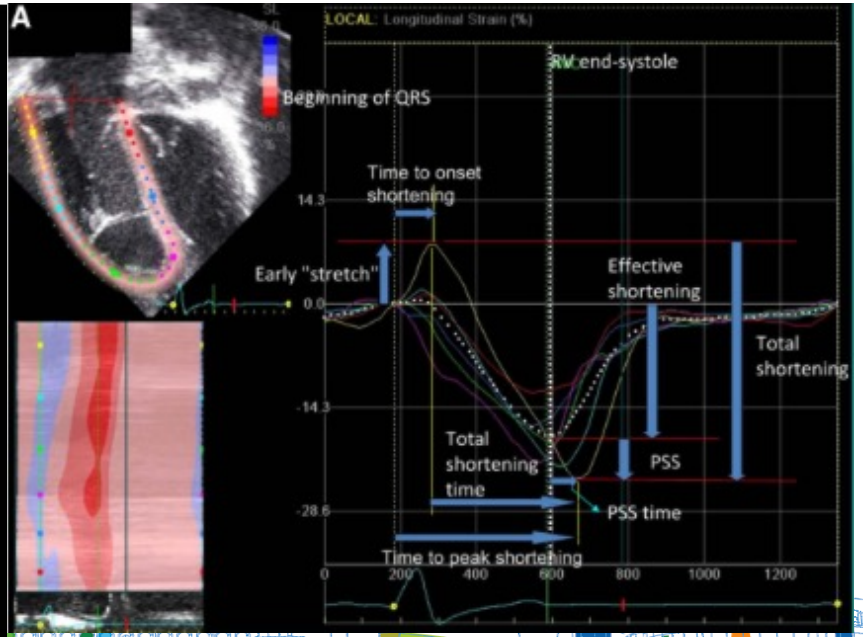
- Conduction delay correlated with RV volume (MRI)
- Hypothesis
  - diffuse RV disease not limited to RVOT
  - Hemodynamic/electrical interaction

Jalal et al. AEPC meeting 2013

RV mechanical activation pattern in rTOF

- Impact of dyssynchrony on RV function
- Abnormal mechanical activation pattern
  - Right sided septal flash/ early septal activation
  - Prestretch of laterobasal wall
  - Late contraction of laterobasal wall and PSS



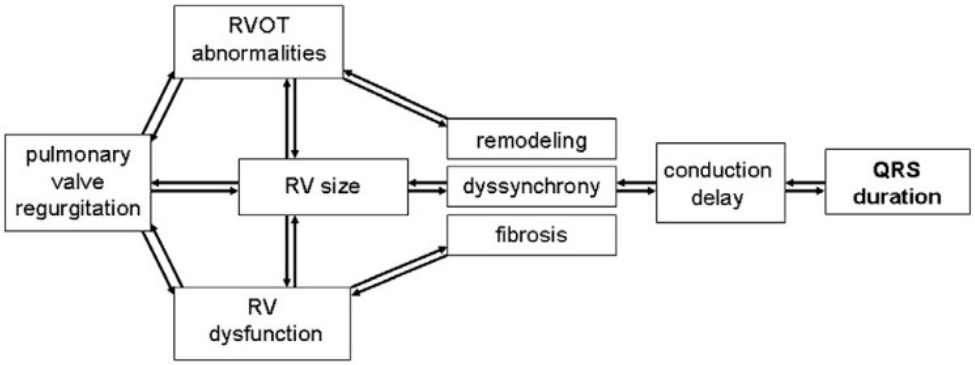


Hôpitaux de Bordeaux

Hui, Friedberg. Circ Cardiovasc Imaging. 2014

Conclusion

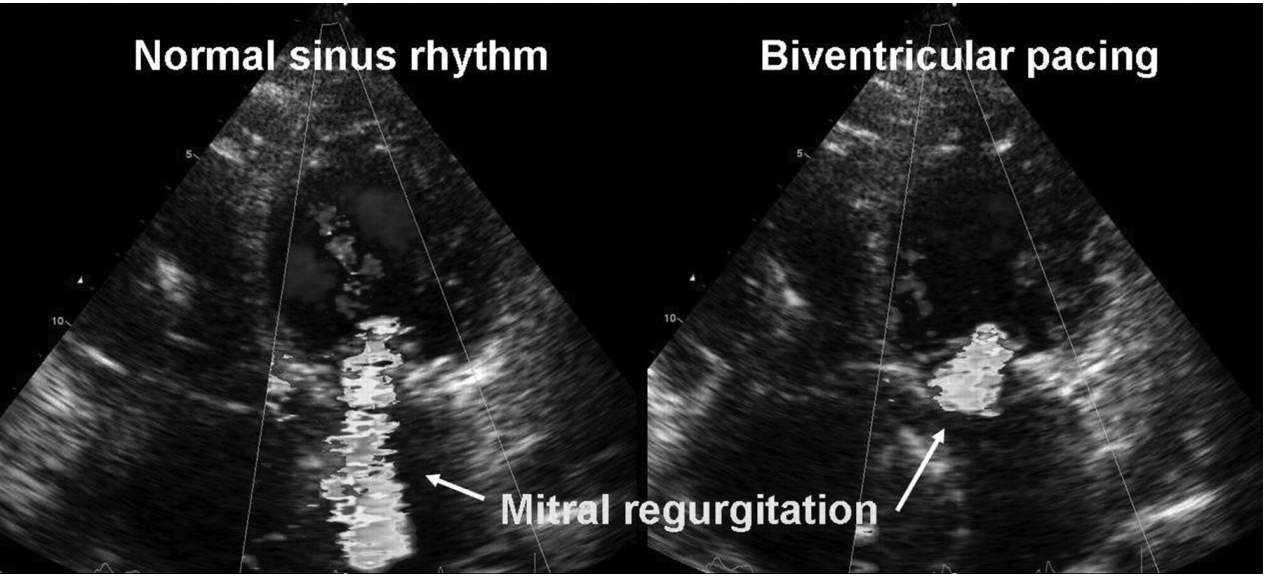
- CRT indication for RV failure: complementary to PVR



- Potential effects on RV failure
  - Reduce RV activation delays
  - Correct inefficient PSS

conclusion

-potential improvement of LV function
-increase FC
-LV remodeling with improvement of MR

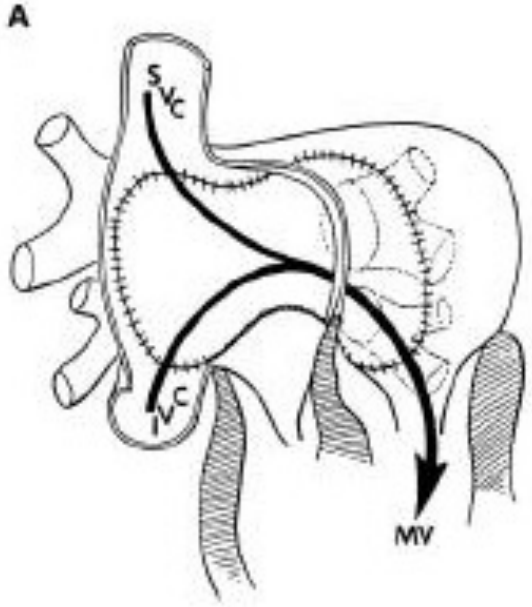


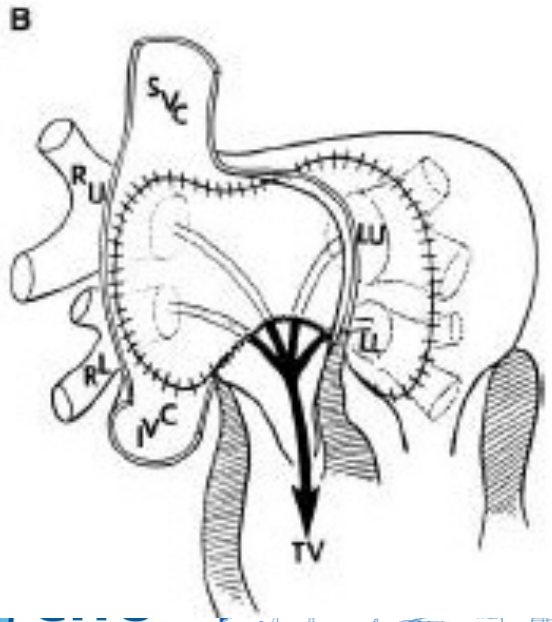
Thambo J et al. Europace. 2007 9:1184.
Diller GP et al. Europace 2006 8:267–72
Kirsh JA et al. Circulation. 2006 11:113:691-2

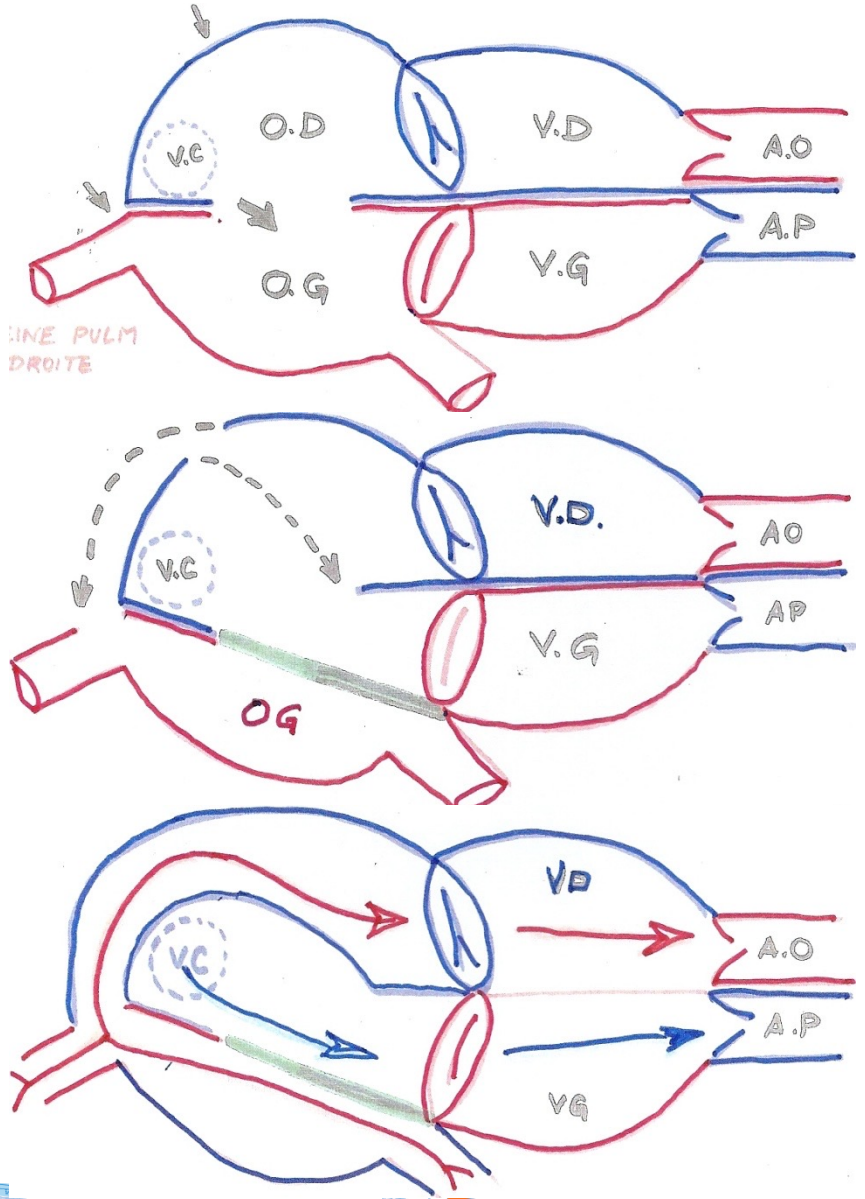
Systemic RV

Systemic right ventricle

- Atrial switch operation
- ccTGA


A
SVC
IVC
MV


B
SVC
RU
LU
RL
LL
IVC
TV

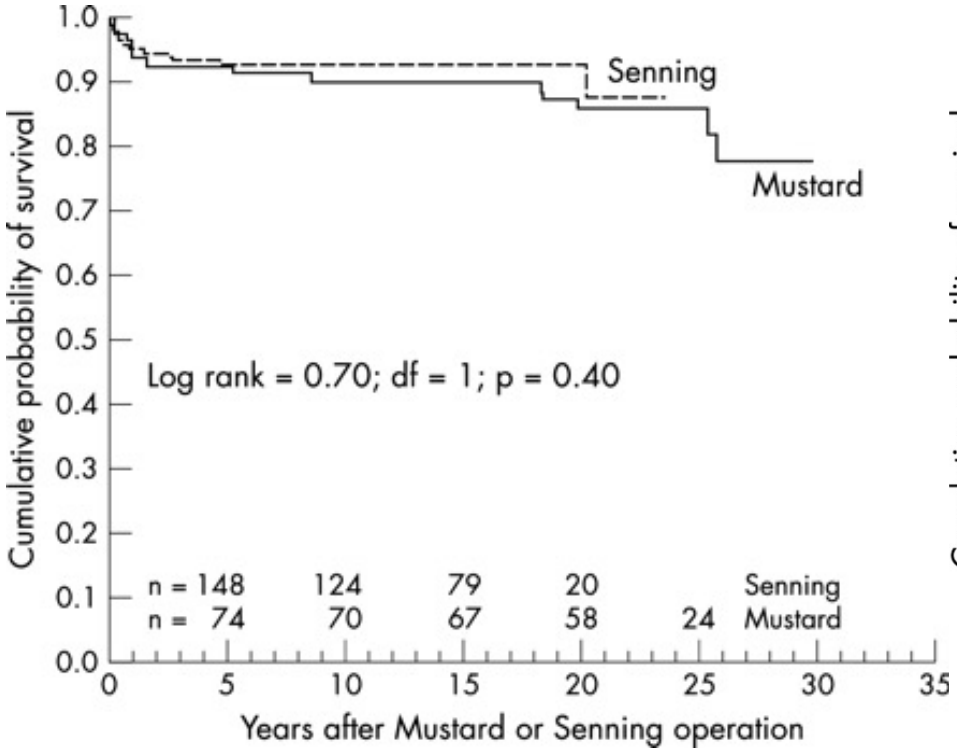

O.D
V.C
V.D
A.O
A.P
V.G
O.G
V.D.
AO
AP
V.G
OG
VD
A.O
A.P
VG

Senning/Mustard

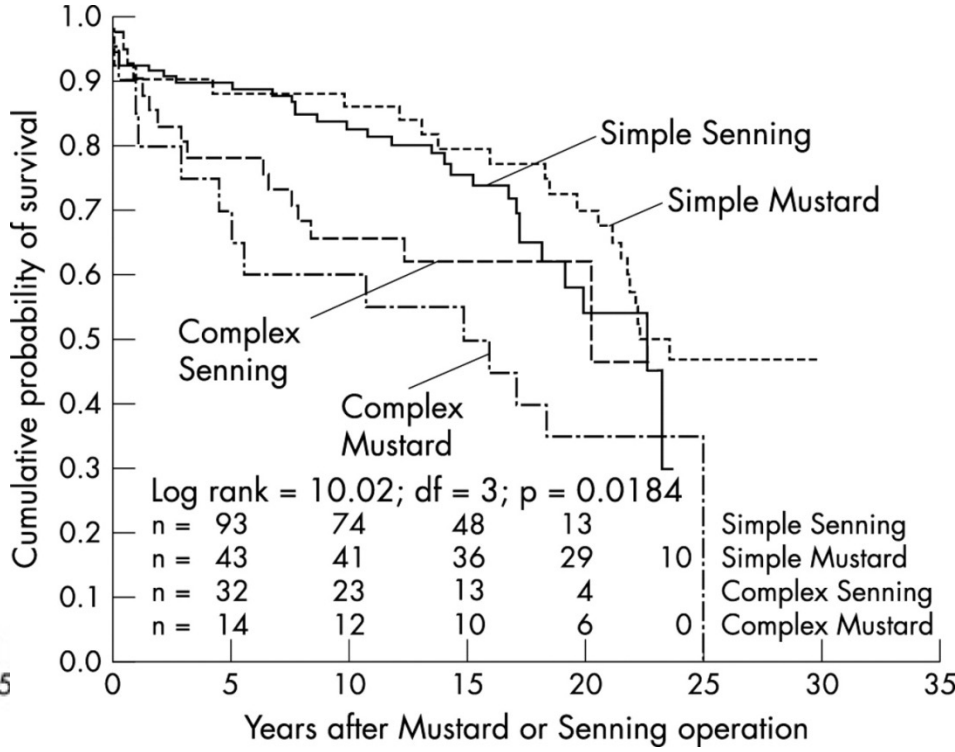
- Dysfonction VD 10 % à 10 ans
- Insuffisance tricuspide 35 % à 10 ans
- Sténoses des veines caves et des pulmonaires
- Perte de la fonction contractile de l'oreillette

- Troubles du rythme
  - Arythmie supra ventriculaire 20 %
  - Dysfonction sinusal 50 %

    - Limitation de la prescription des anti-arythmiques
    - Difficultés d'implantation des PM (VG sous pulm)
    - Difficultés de l'ablation (position SC, anatomies chenaux)



Deanfield. Circ 1999



Moons. Heart 2004

Facteurs de risque:
- dysfonction du ventricule systémique
- plicature ou thrombose des chenaux

Arythmie supra ventriculaire

Complications médicales

Mort subite et arythmie ventriculaire

Troubles du rythme auriculaires

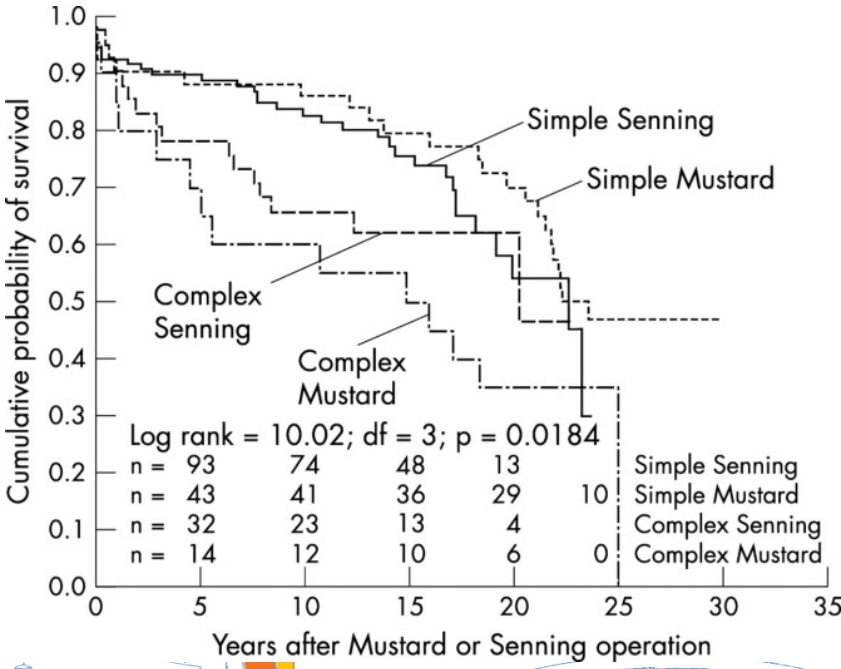
Dysfonction sinusale (difficultés thérapeutiques)

Obstacle sur les chenaux

Dysfonction du VD systémique

Strategie diagnostique et therapeutique?

Evaluation des patients
Traitement medical
Stimulation multisite
Cerclage préparateur puis switch
Transplantation



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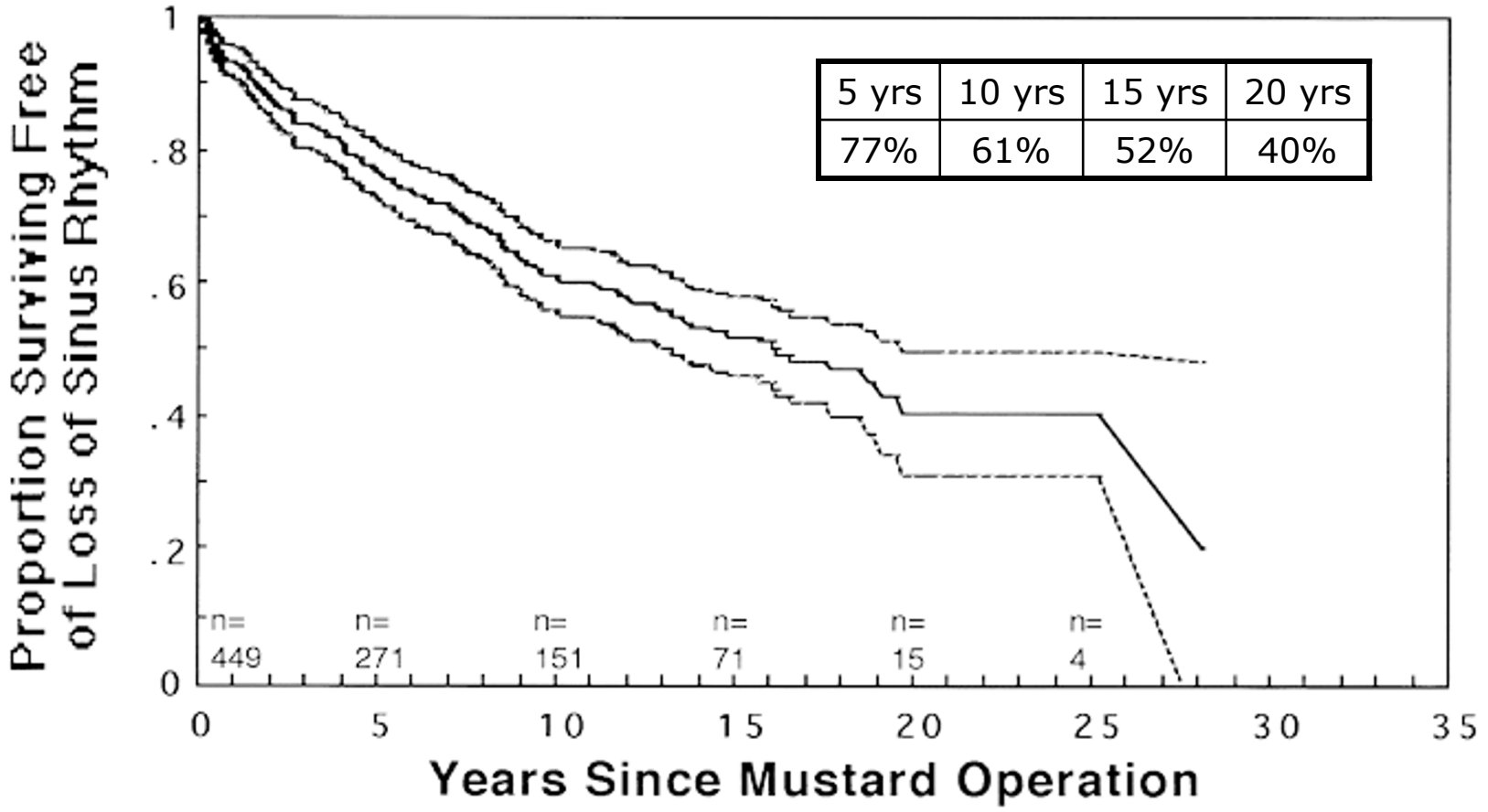
Moons et al. Heart 2004

Dysfonction sinusale

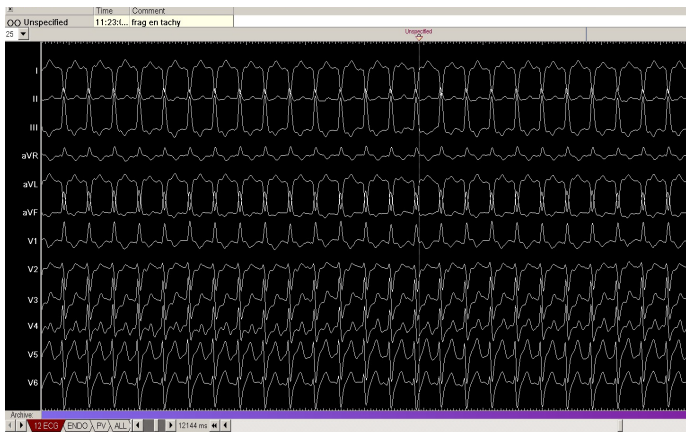


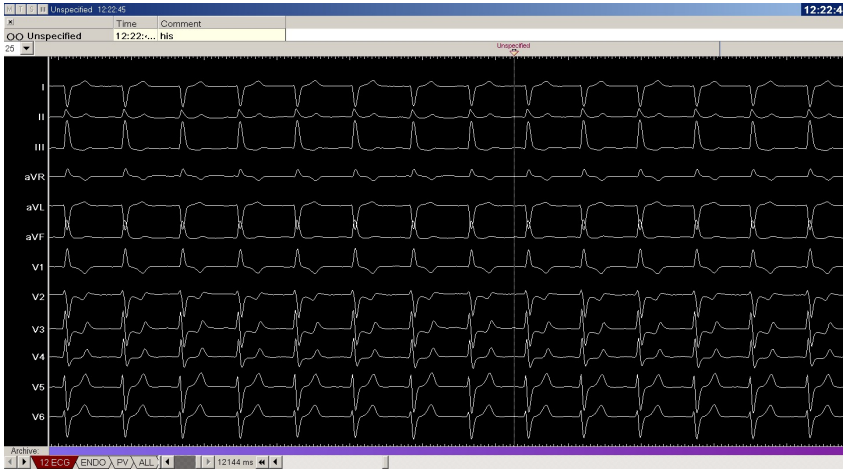
5 yrs	10 yrs	15 yrs	20 yrs
77%	61%	52%	40%

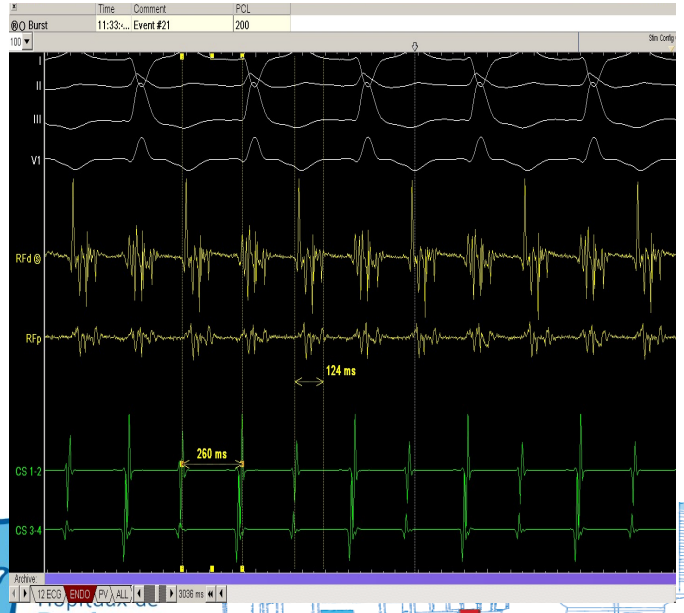
Gelatt M et al. JACC 1997;29:194-201

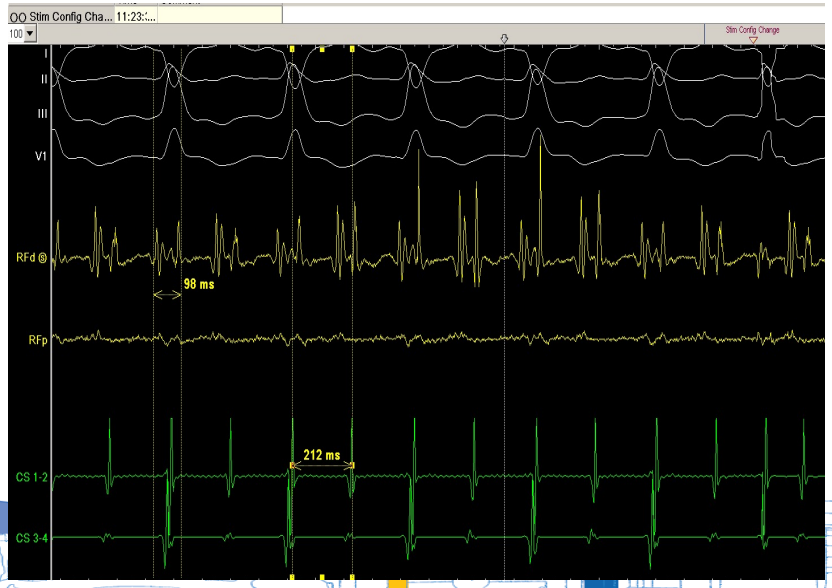
20% de Flutter, 50% de dysfonction sinusale

End of procedure



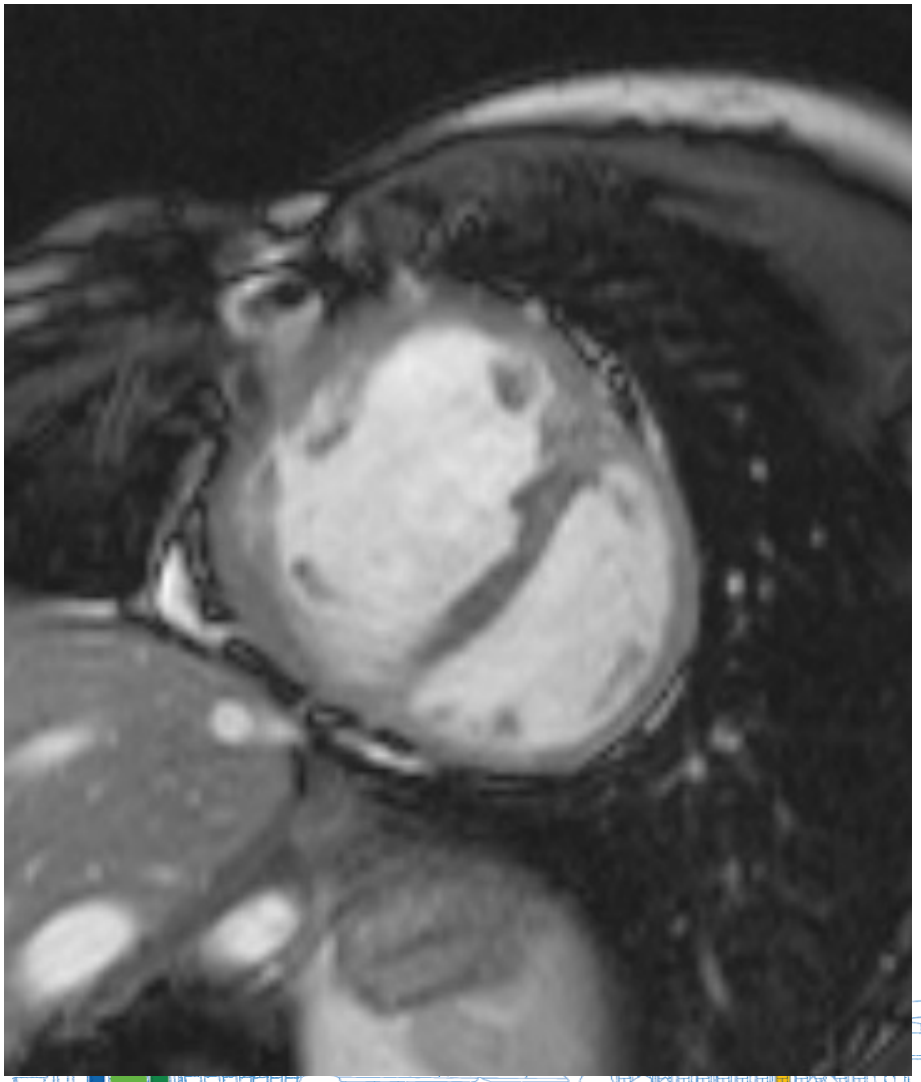




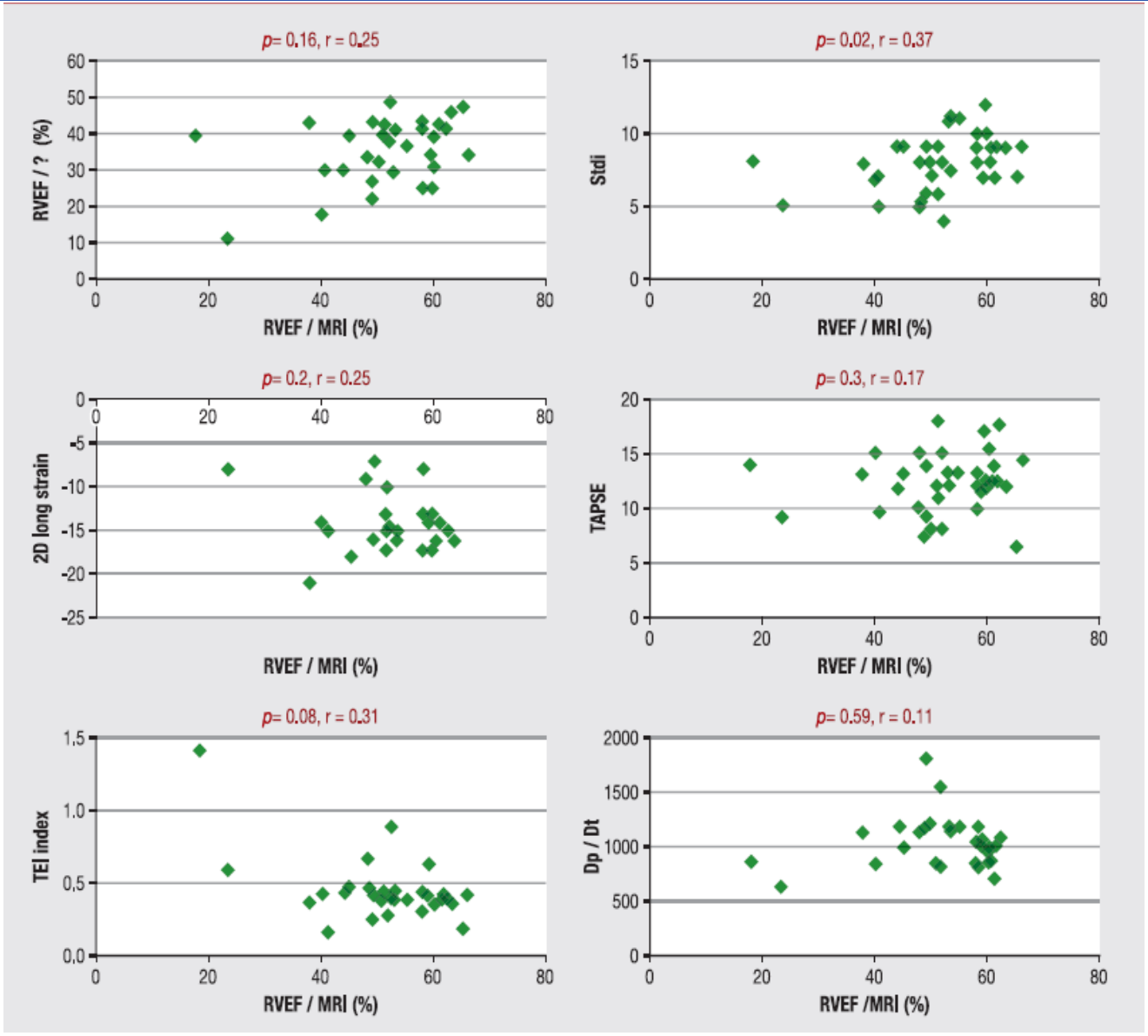


Bordeaux

MRI: gold standard

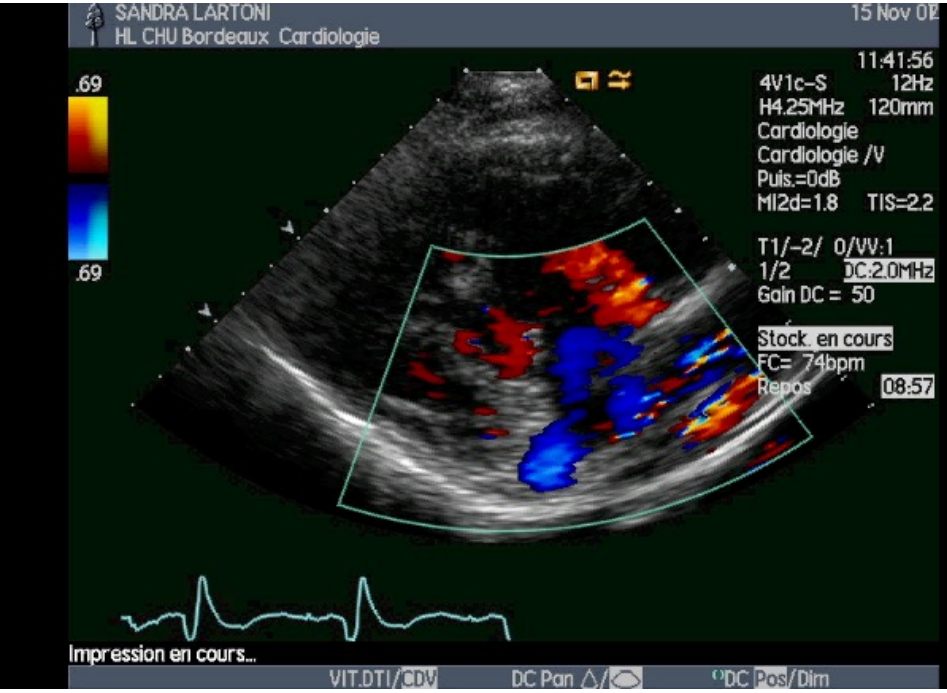


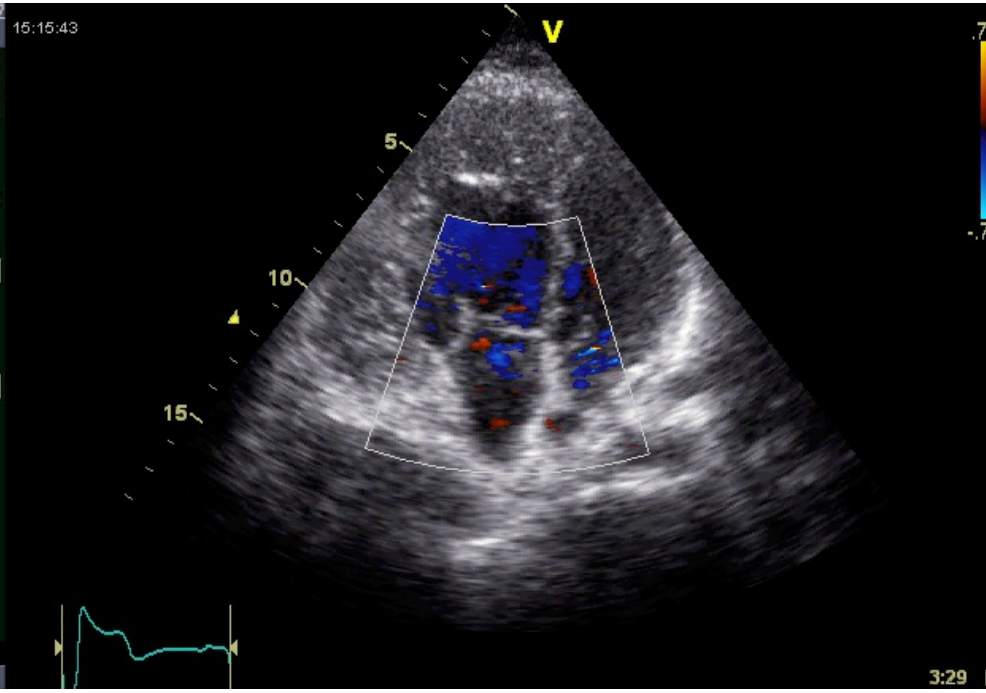
CHU Hôpitaux de Bordeaux



Iriart X et al. ACVD 2012

Systemic AVV assessment





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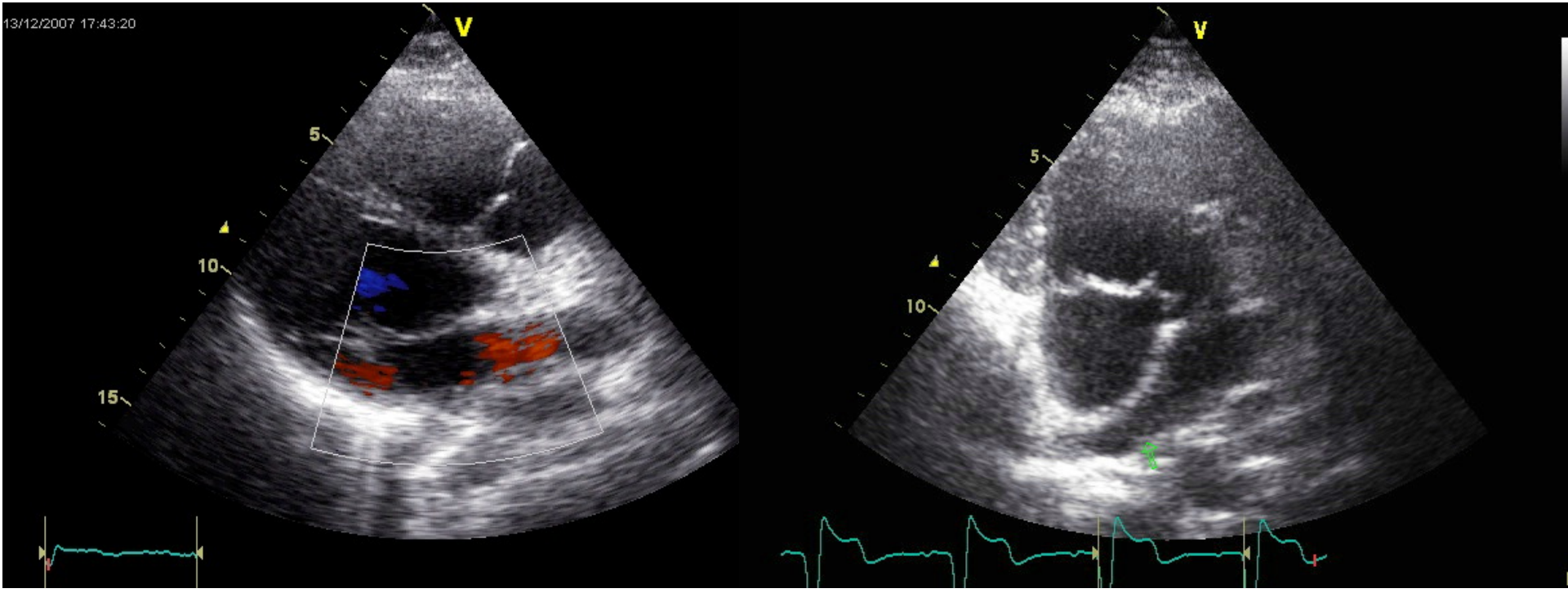
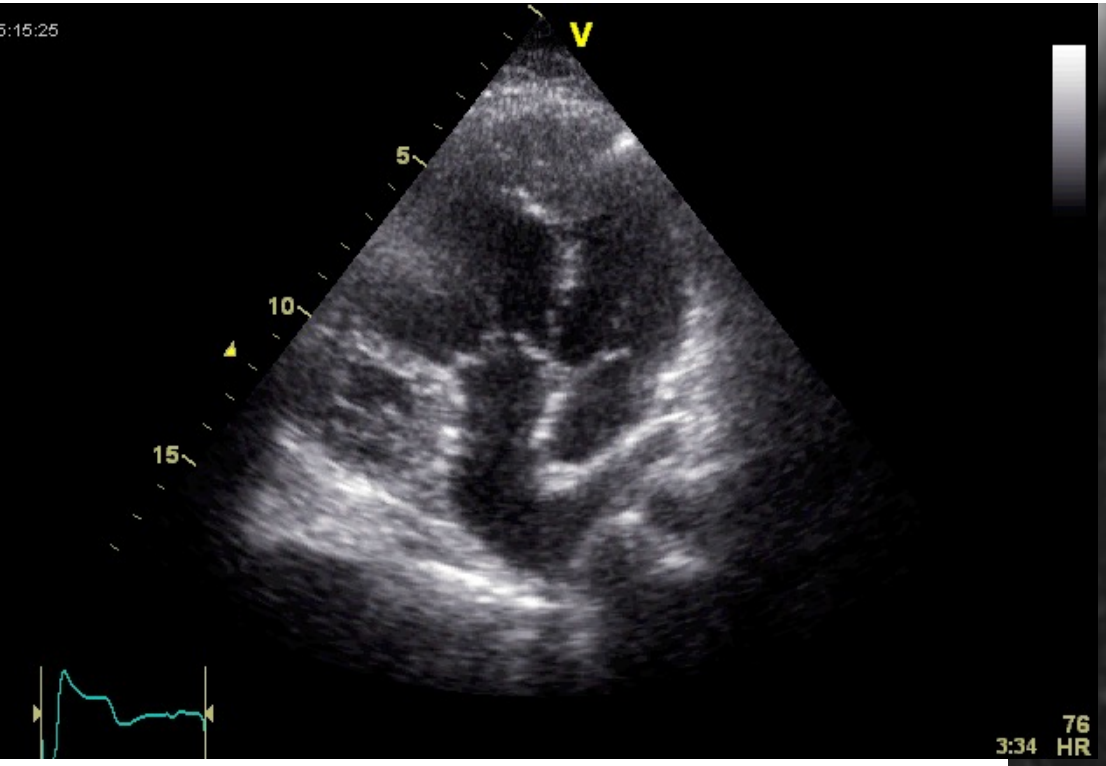
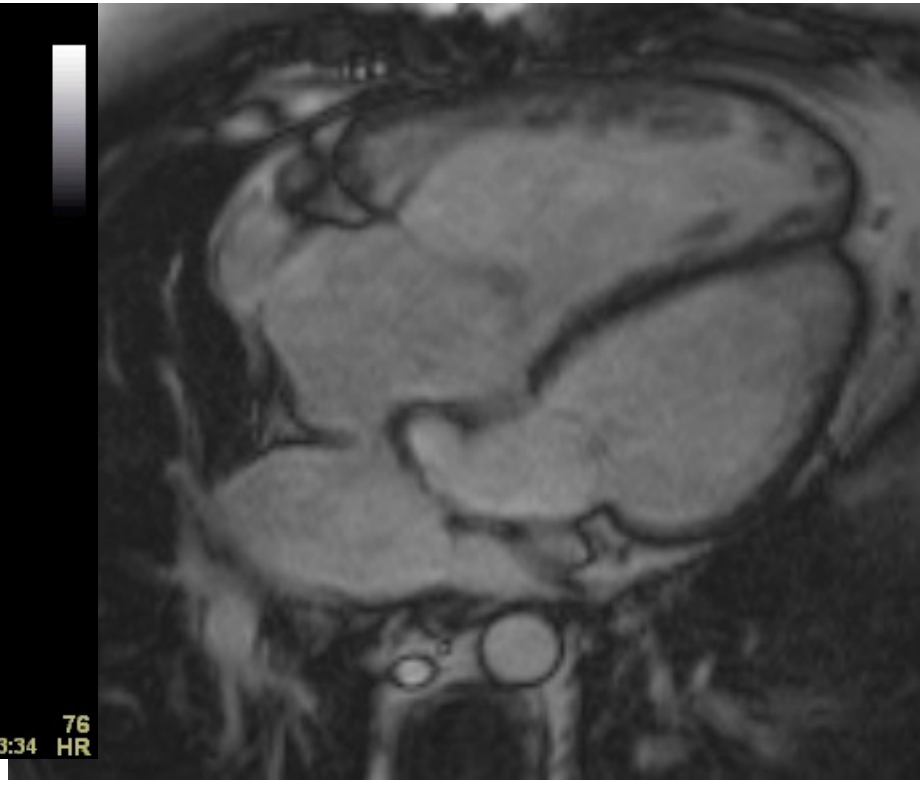
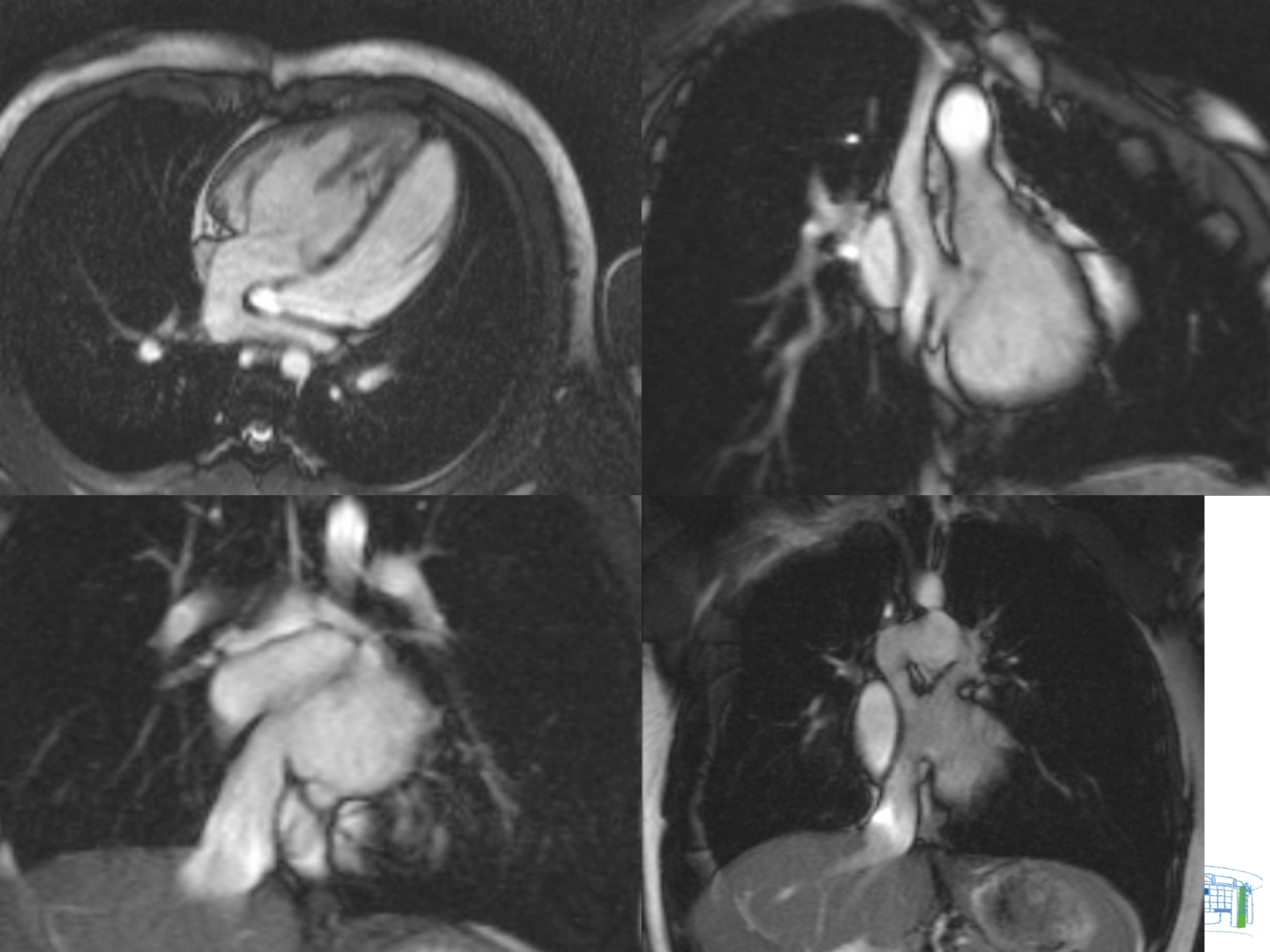
Baffles assessement

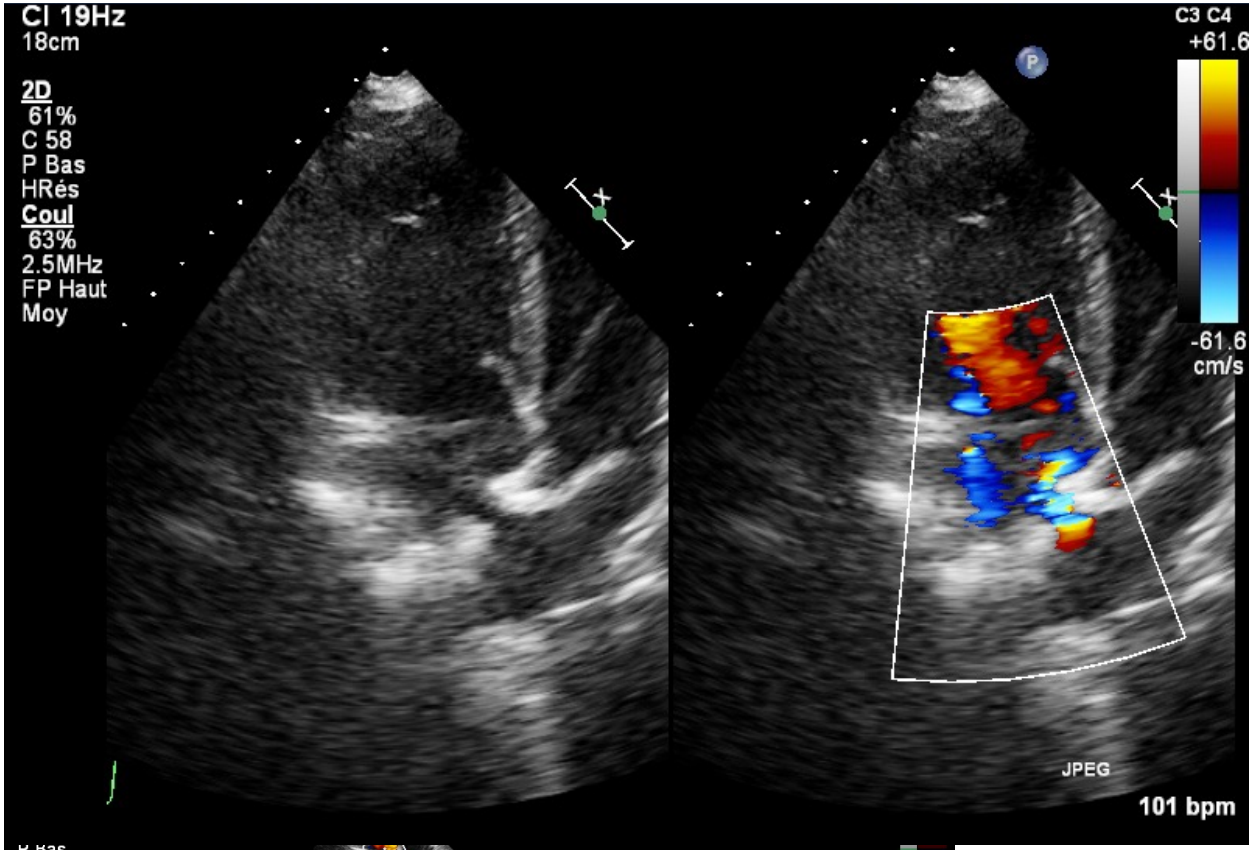


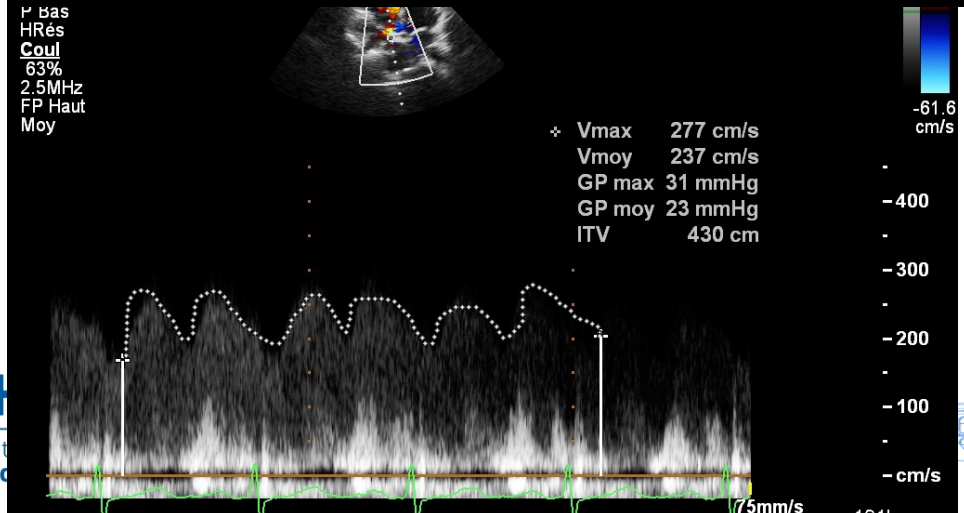


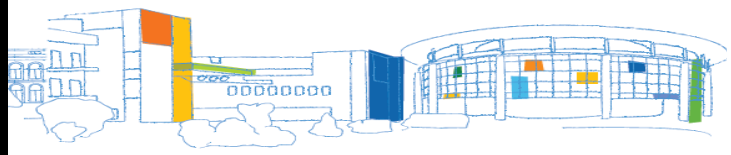


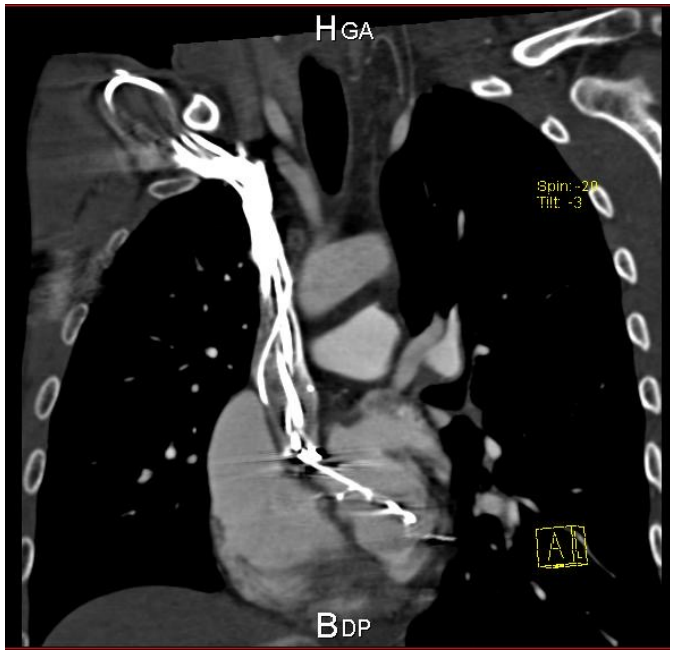
CHU Hôpitaux de Bordeaux

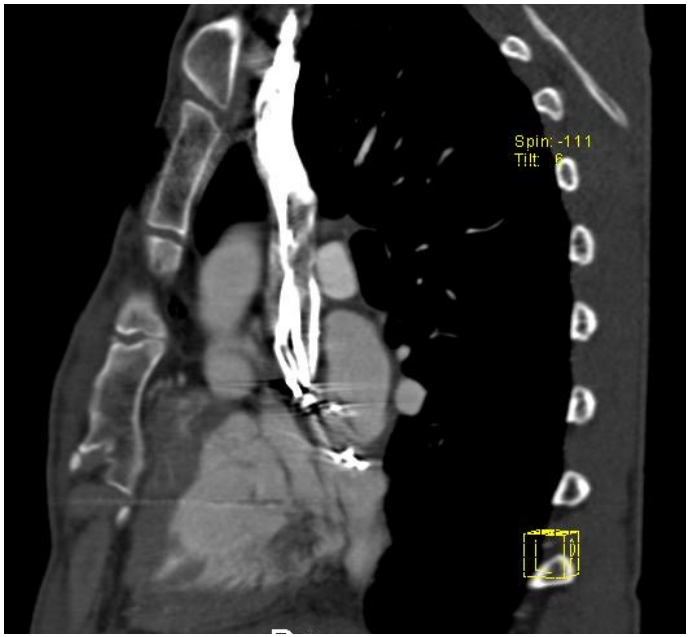


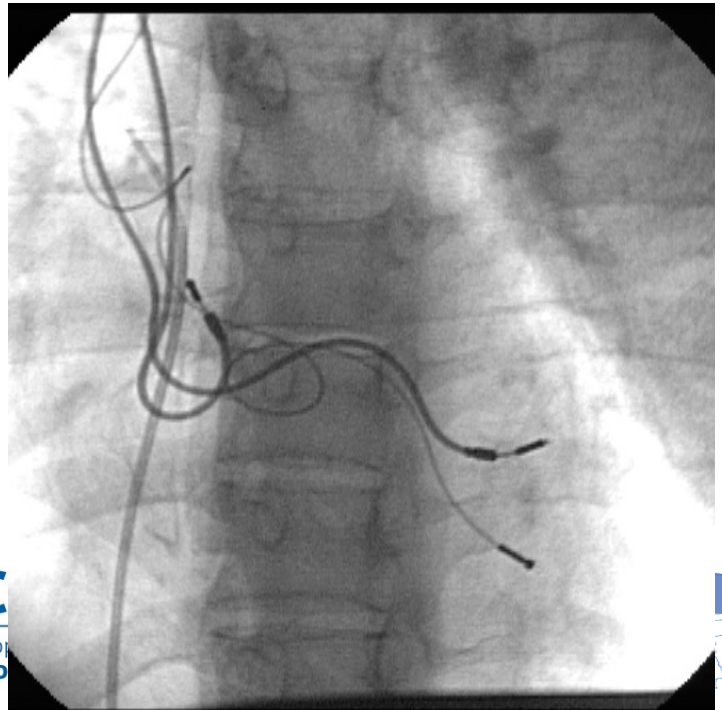

CI 19Hz
18cm
2D
61%
C 58
P Bas
HRés
Coul
63%
2.5MHz
FP Haut
Moy
C3 C4
+61.6
-61.6
cm/s
JPEG
101 bpm

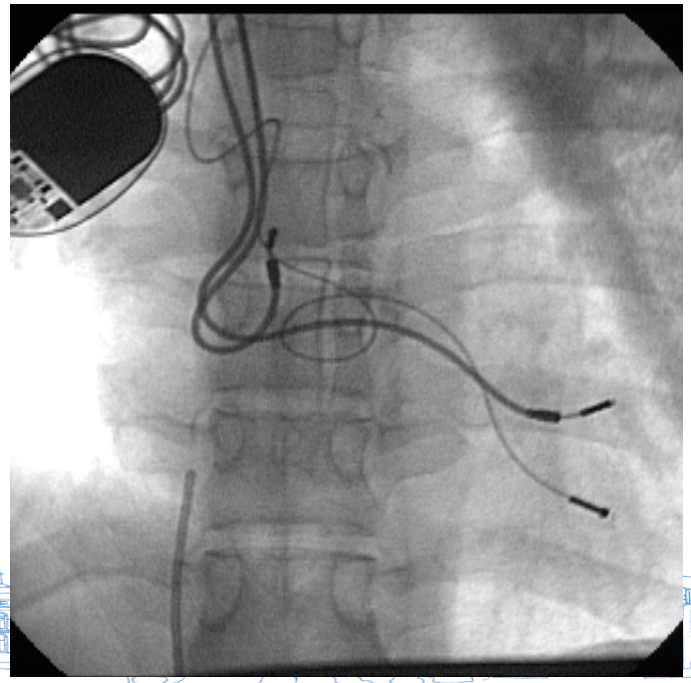

P Bas
HRés
Coul
63%
2.5MHz
FP Haut
Moy
-61.6
cm/s
Vmax 277 cm/s
Vmoy 237 cm/s
GP max 31 mmHg
GP moy 23 mmHg
ITV 430 cm
400
300
200
100
cm/s
75mm/s

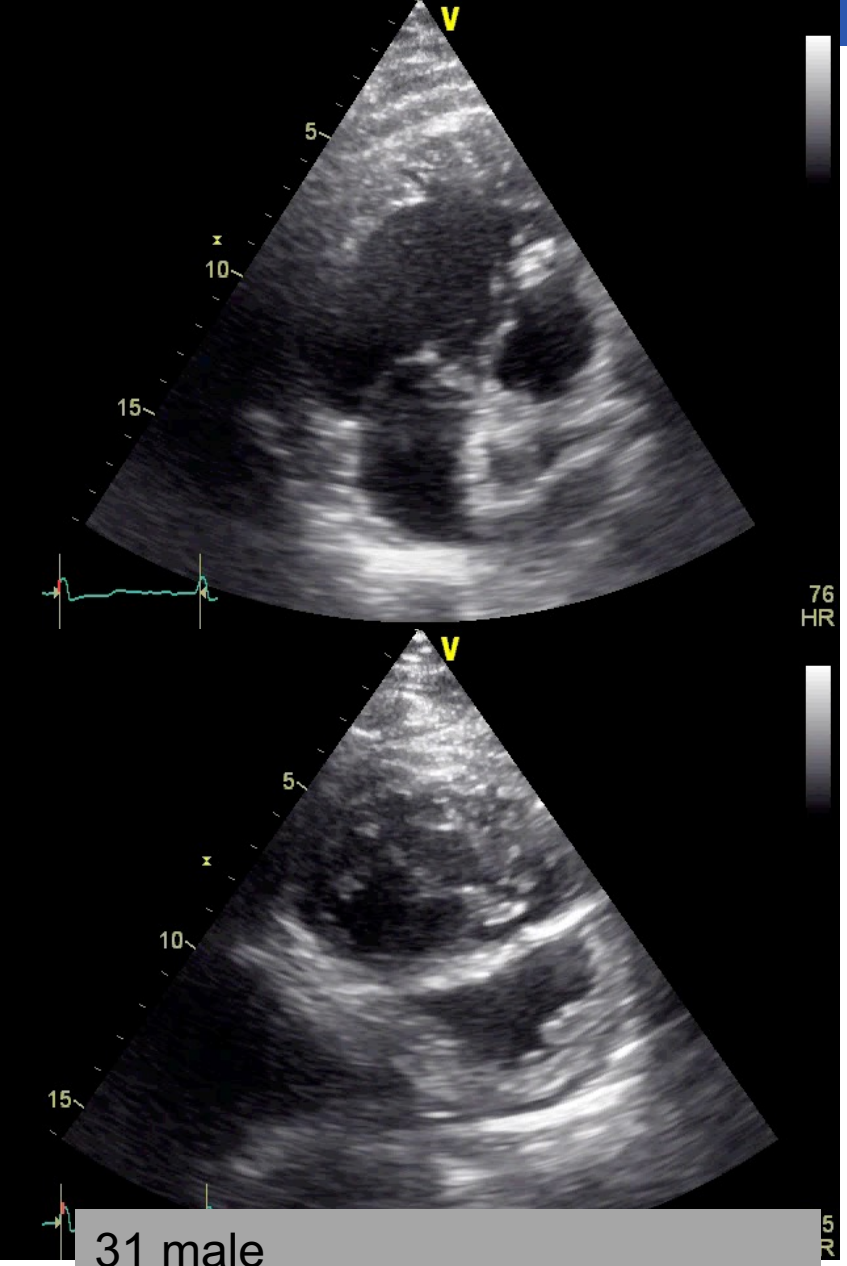



HGA
Spin: -20
Tilt: -3
BDP

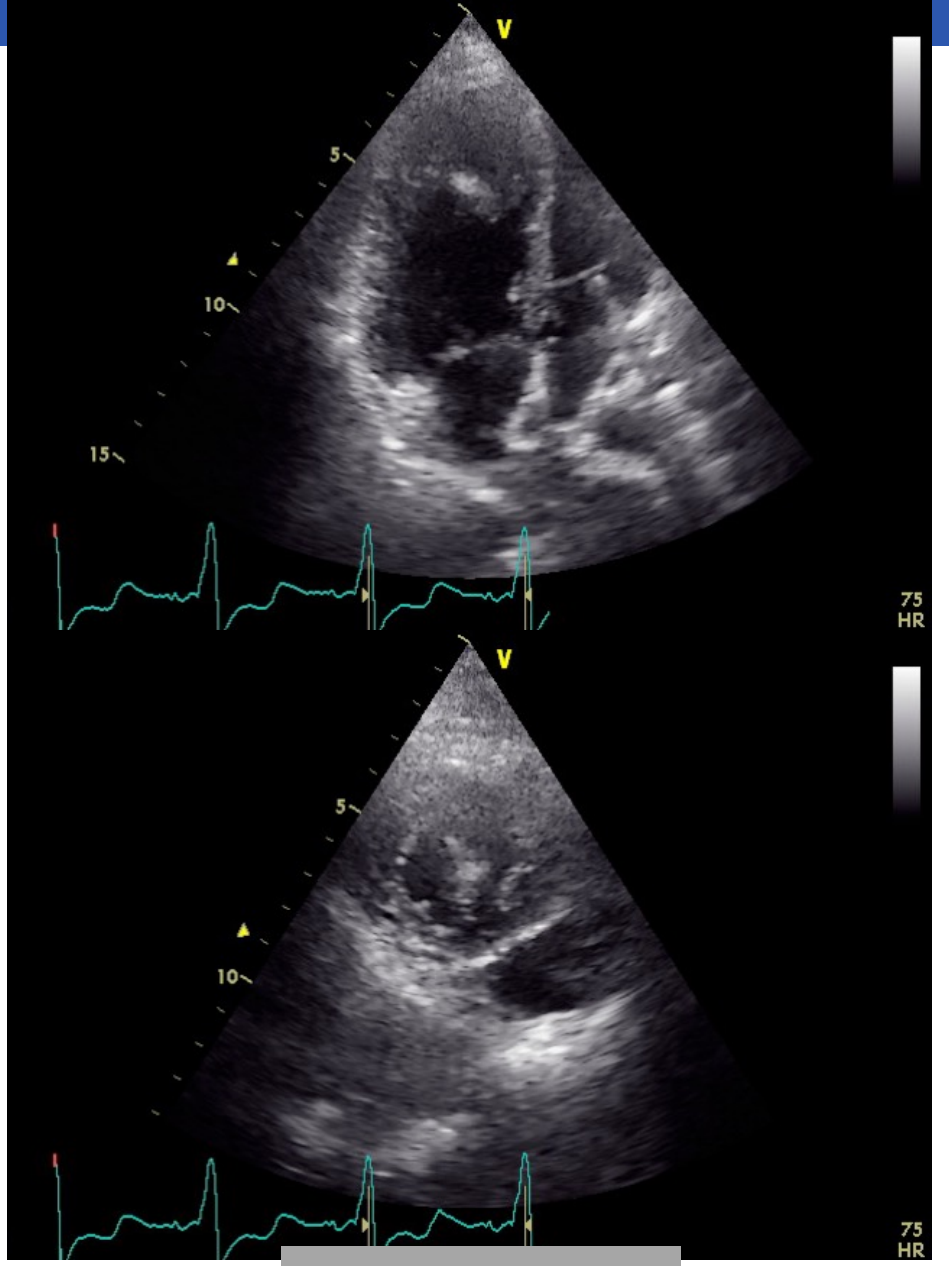

Spin: -111







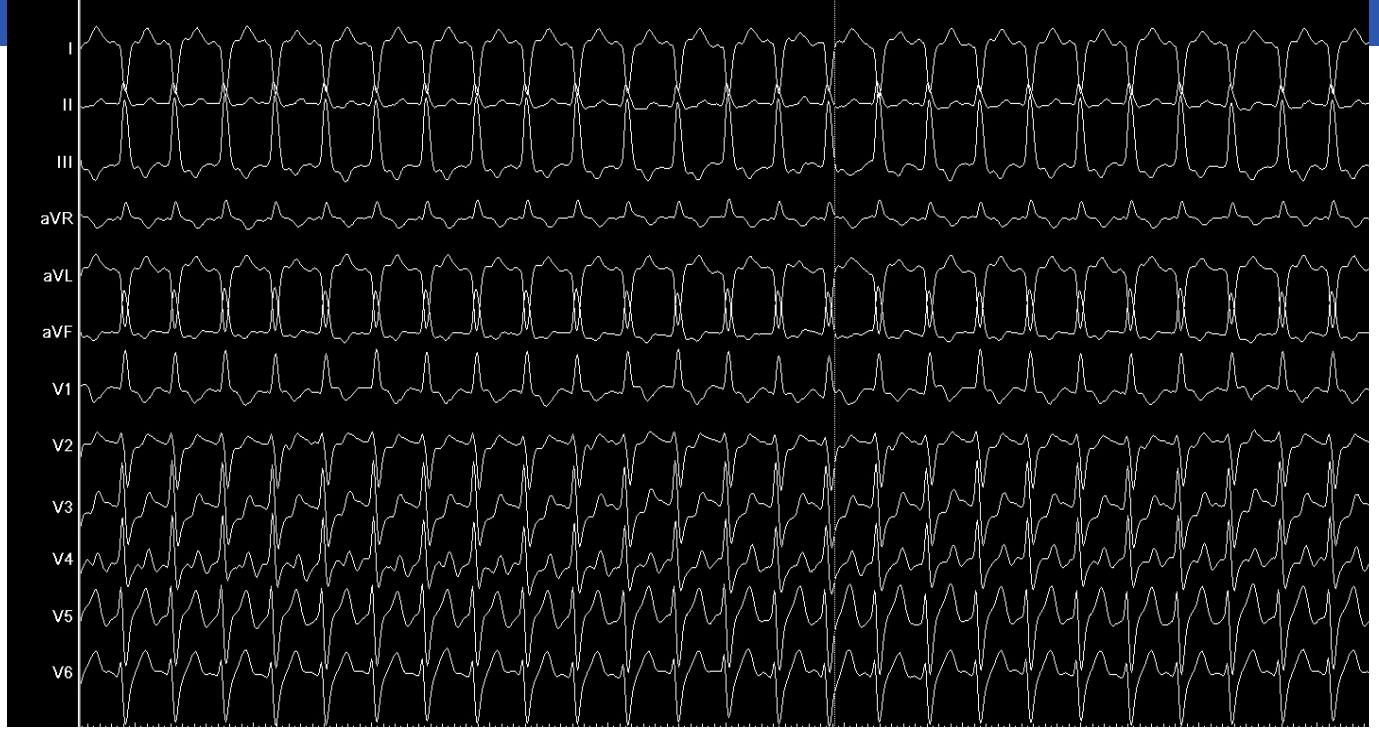
31 male
Asymptomatic, mild obesity
Leisure sport activity
CPET 270W

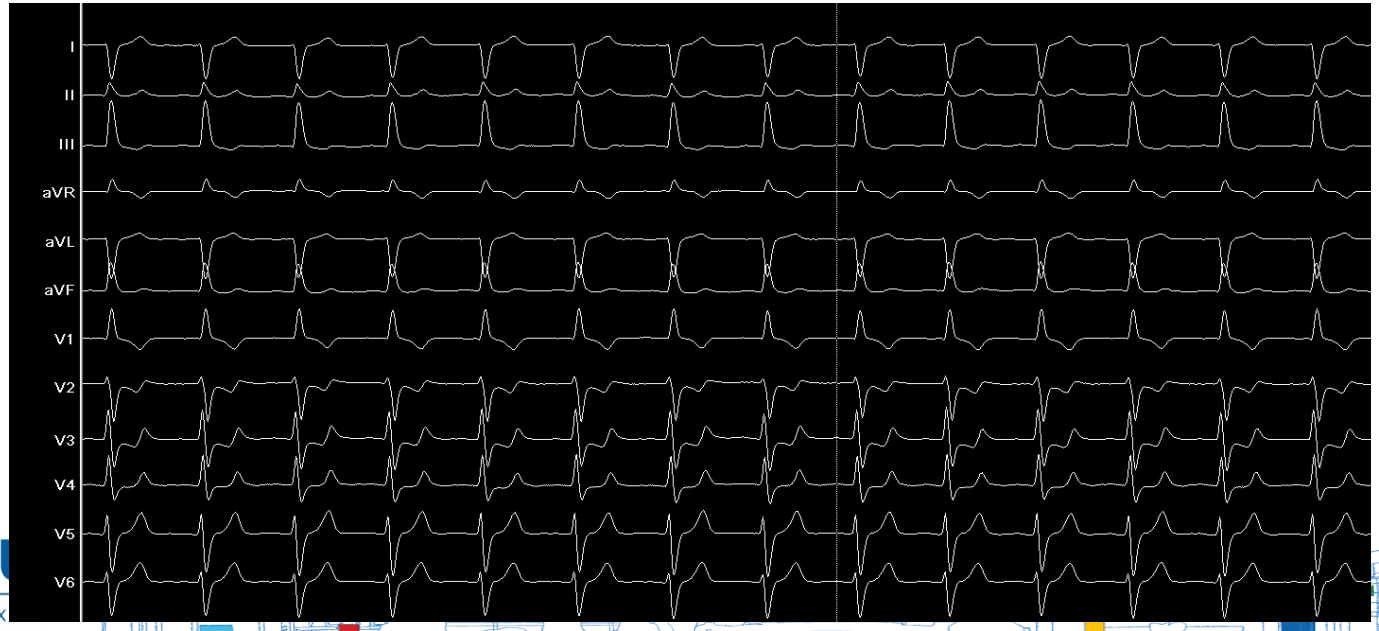


28 female
NYHA FC3
Pic VO2 14ml/kg

Cas clinique

- Jeune homme de 26 ans
- D-TGV traité par atrioseptostomie de Rashkind à la naissance puis correction par switch atrial à l'âge de 9 mois
- Excellente tolérance à l'effort (280W à EE), très sportif
- Dysfonction sinusale asymptomatique FC de repos 45-50, FC<40 la nuit
- Arythmie supraventriculaire mal tolérée en 2007 et 2008 réduit par cordarone IV, relai sotalol très mal toléré avec asthénie et majoration des bradycardies
- Aggravation de l'arythmie déclenchable aux efforts violents
- CAT?

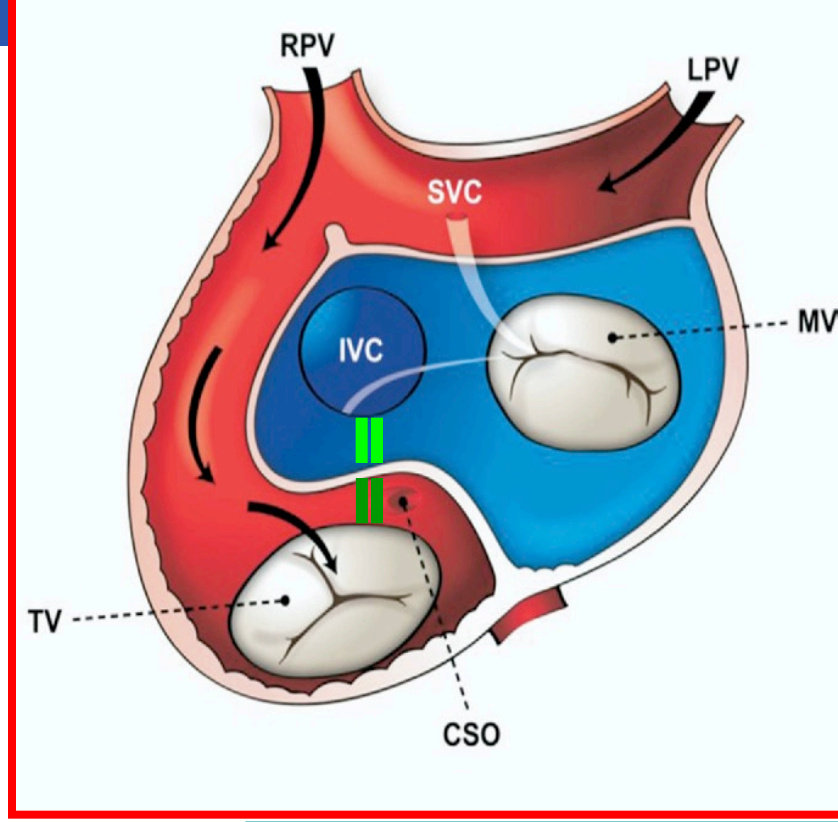

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

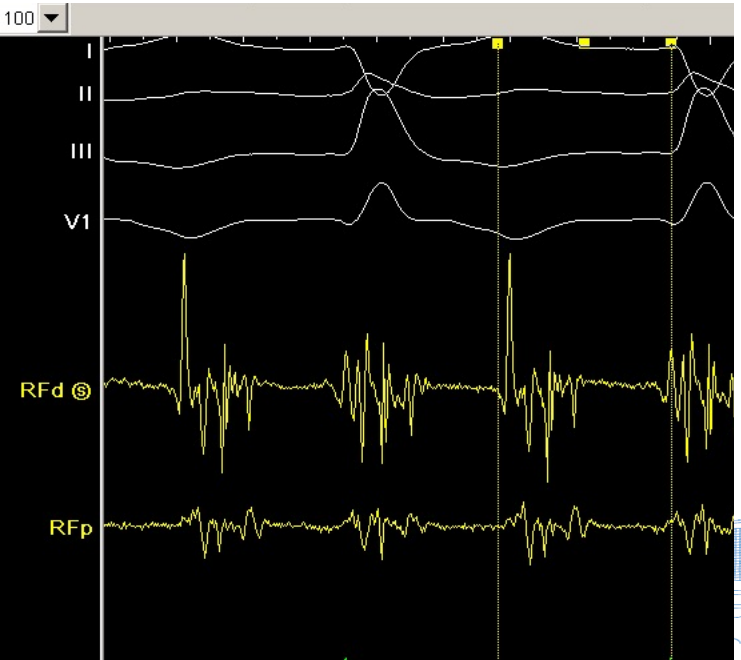

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

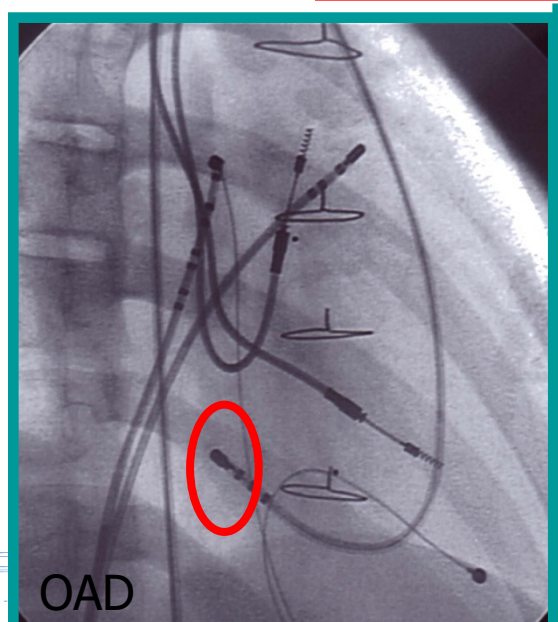
Problème d'accès aux cavités cardiaques

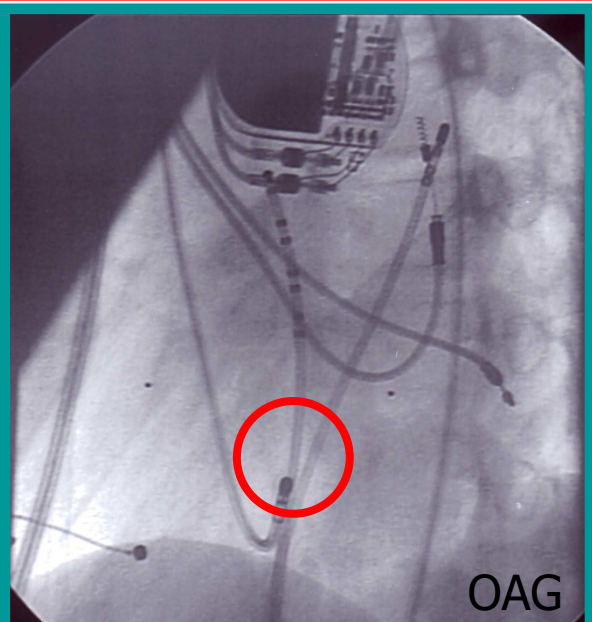
Flutter Atrial et switch atrial

- Isthme Cavo-Tricuspide est séparé par le “baffle”
- Localisation du sinus coronaire depend de la position du baffle









Transposition des gros vaisseaux opérée par switch artériel

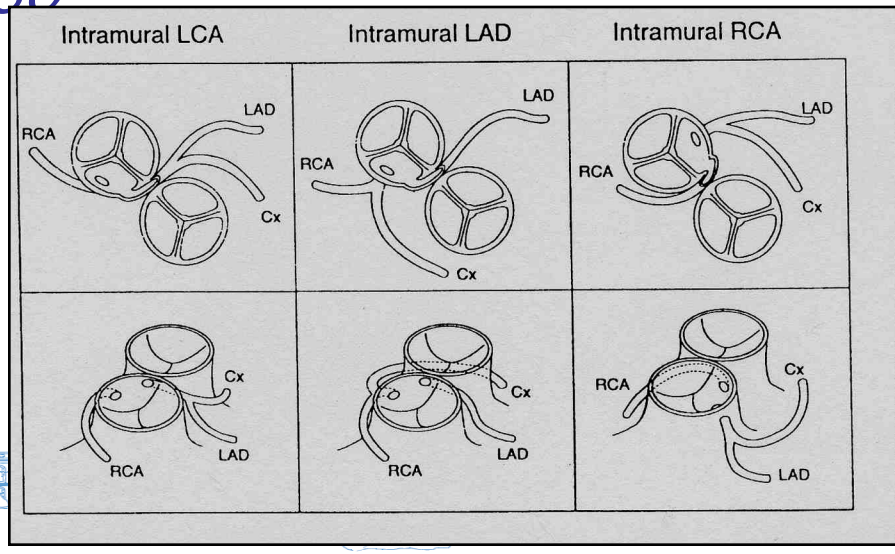
TGV - switch artériel

- Surveillance voie pulmonaire (gradient APG)
- surveillance néo-valve aortique, dilatation du culot
- problèmes coronaires à distance :
  - épreuve d'effort - thallium d'effort – scanner multibarettes- coronarographie

Coronaires et switch

- 1304 switch – 60 mois de suivi - *Circ 2003;108:186-90*
  - 7.2% évènement coronaire
  - Free
    - 1 an : 92.7%; 10 ans : 91%; 15 ans : 88.2%

- Méta-analyse : 1942 switch , mortalité par évènement coronaire - *Circ 2002;106:2575-80*
  - Ostium unique OR 2.9
  - Intramurale OR 6.5
  - Autre Ano coronaire OR 1.7



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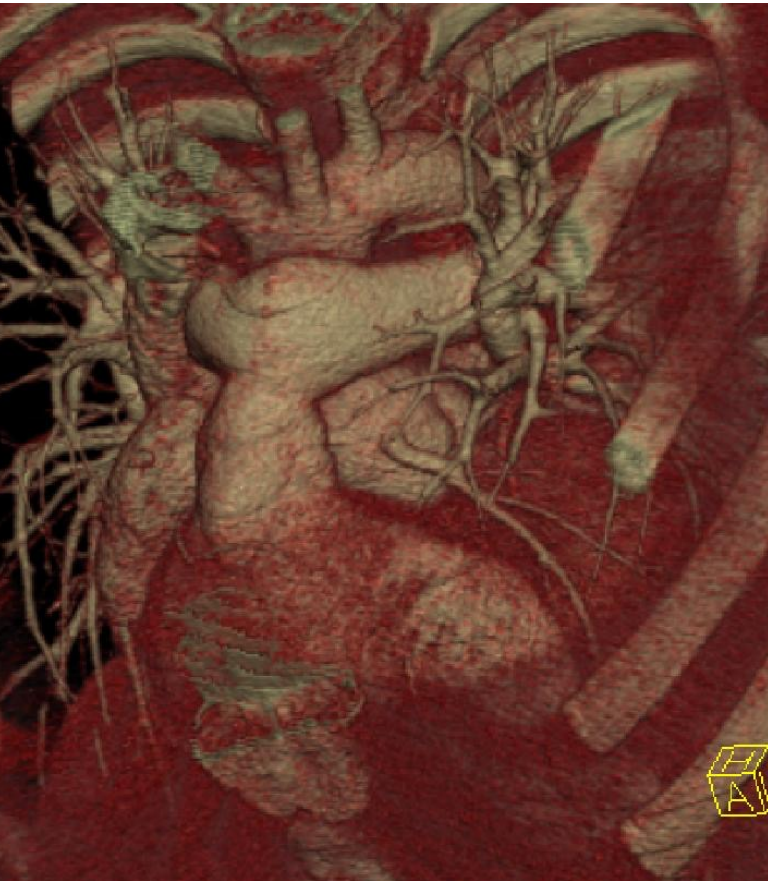
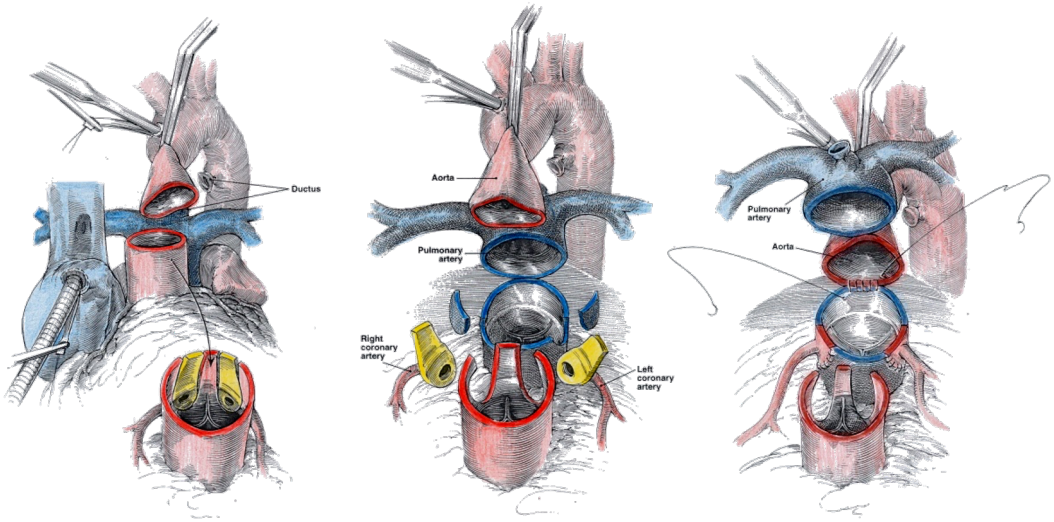
VG et switch

- écho –dobu 9.4 ± 2 ans après switch néonatal (15.5 ± 4.3 jours)
  - coronaires normales à l'angiographie
  - FR, FE abaissés au repos
  - stress : 74% anomalie cinétique segmentaire
    - facteurs favorisants:
      - âge tardif ($p < 0.01$)
      - durée de CEC ($p < 0.01$)
      - durée de CA ($p < 0.045$)

Heart 2005;91:68-72

Switch artériel





Facteurs pronostiques:
-Sténoses coronaires
-Sténoses des branches pulmonaires
-Dilatation du néo culot aortique

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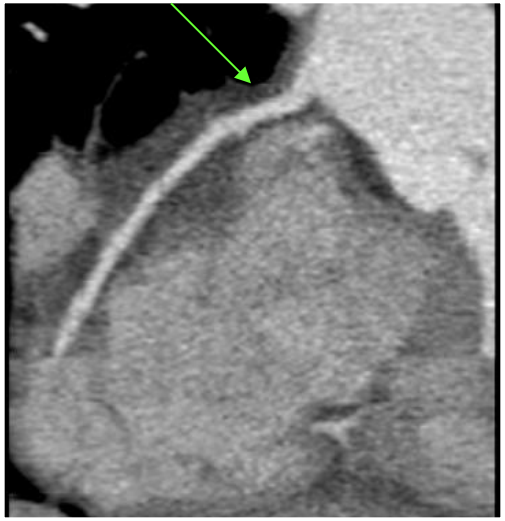
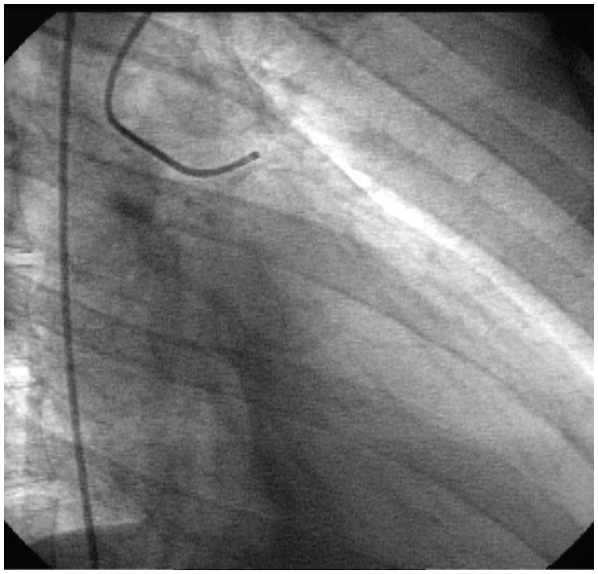
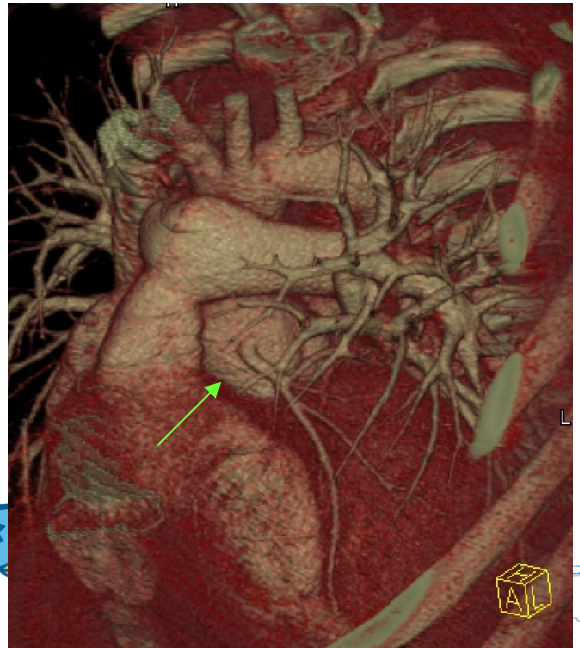
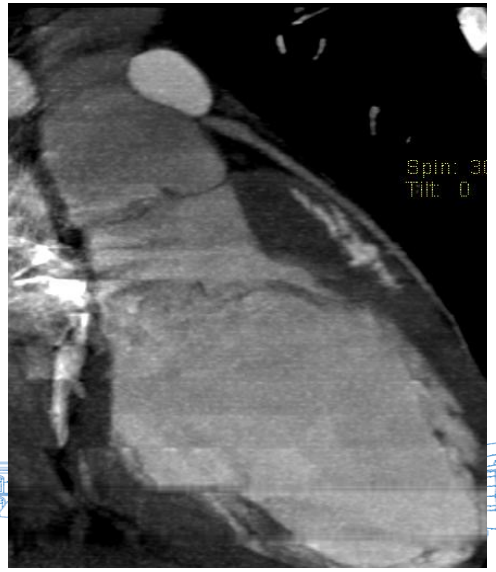
analyse des coronaires et de l'aorte post switch artériel

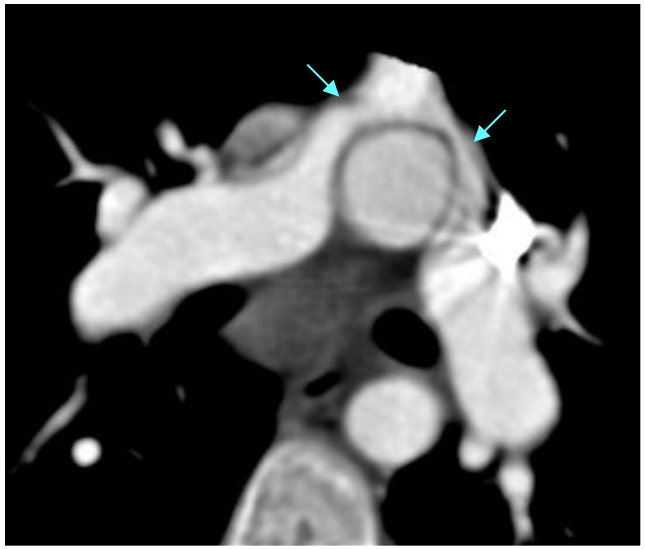


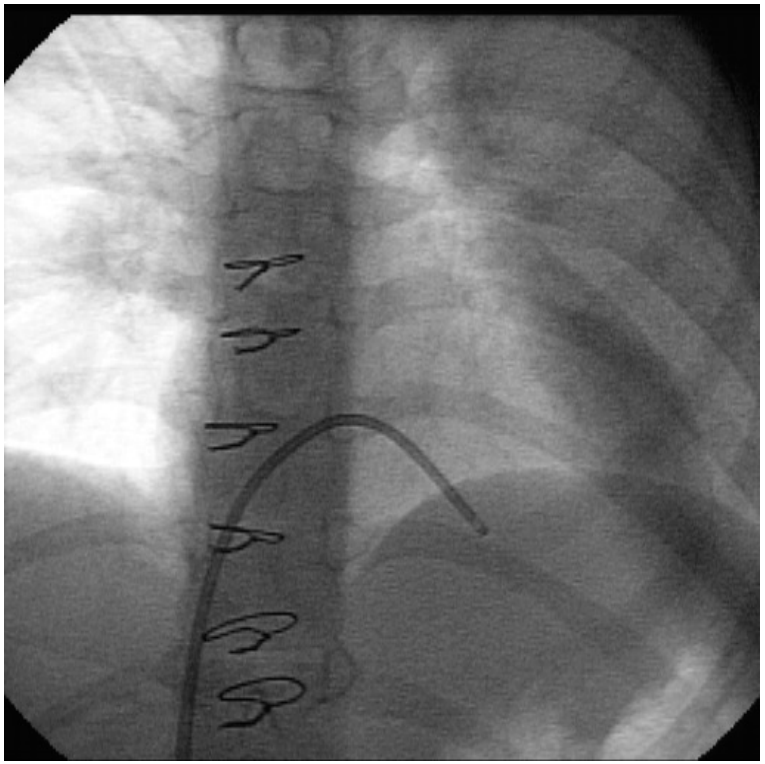


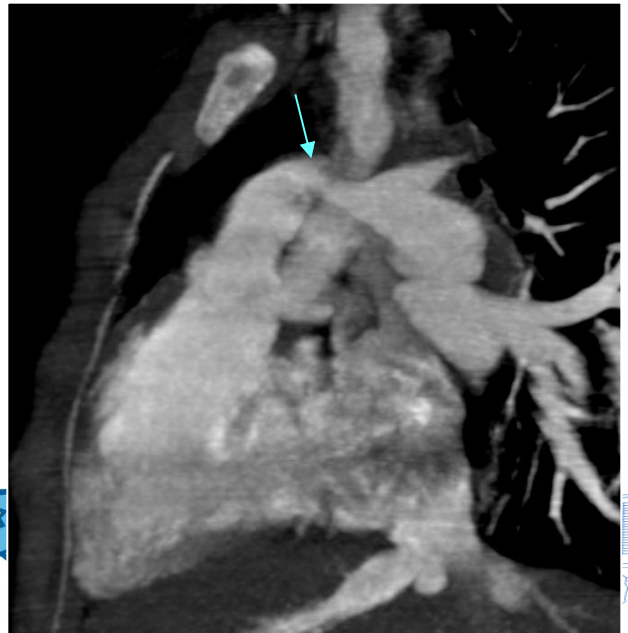




Sténose des AP post-switch

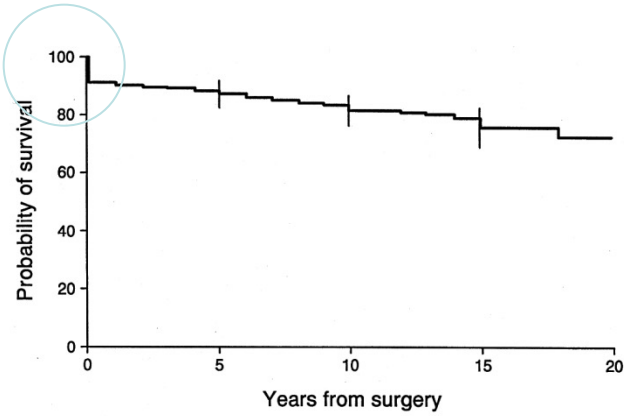




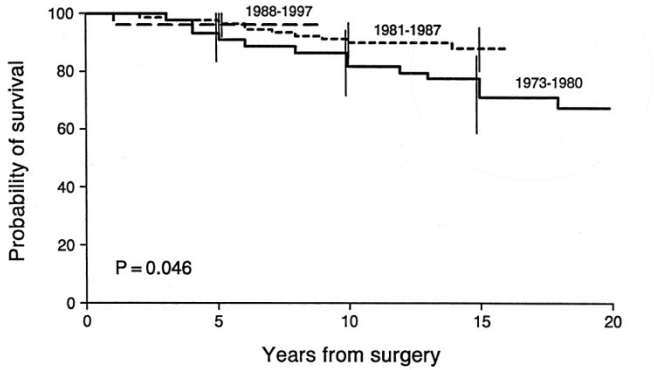


VU et circulation de FONTAN

Complications tardives du Fontan



n=216, 1973-1998 (TCPC < age 3)



Facteurs de risque :

- Age de la correction
- ATCD procédure palliative
- période opératoire
- Anatomie autre que AT
- Hétérotaxies
- PAP élévées
- Fuite valve AV
- NYHA >2

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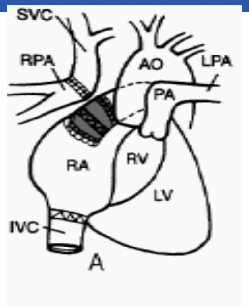
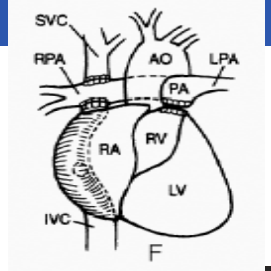
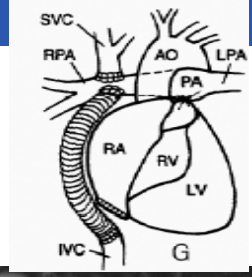
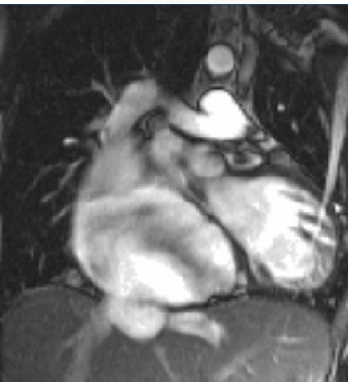
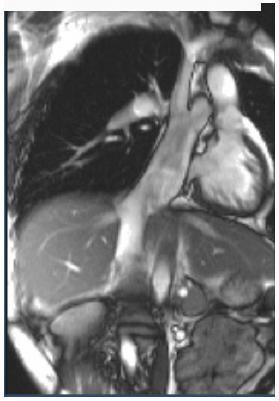
Mair et al. JACC 2001
Fontan et al. Circulation 1990

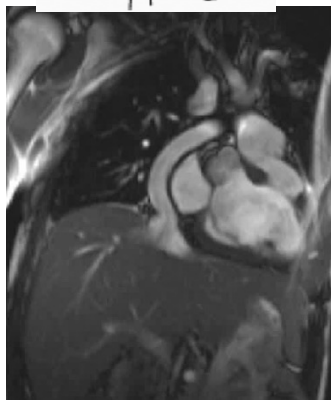


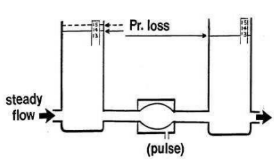


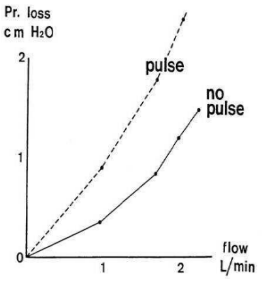


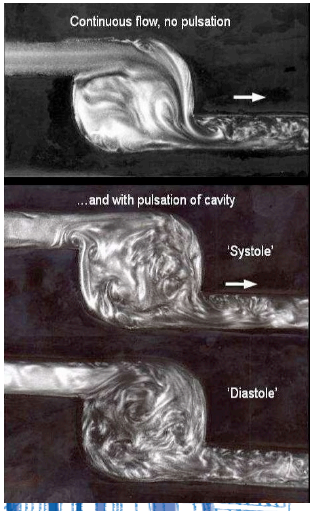


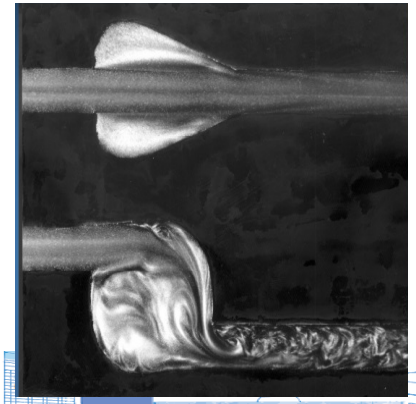


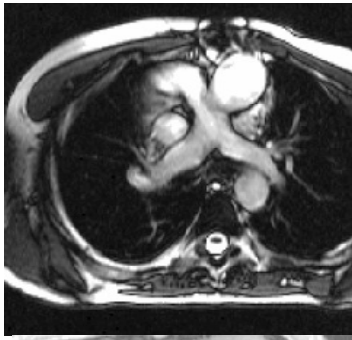


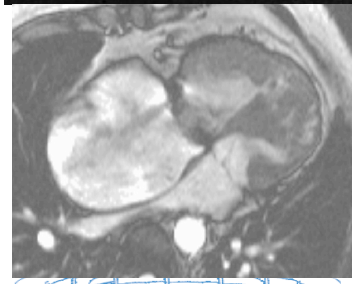










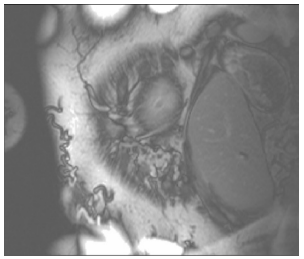
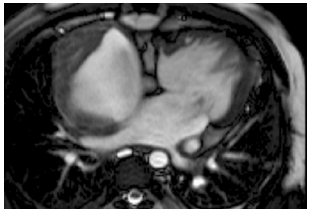
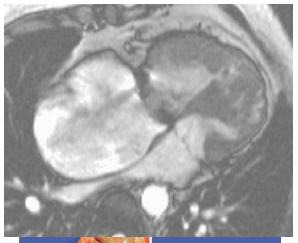
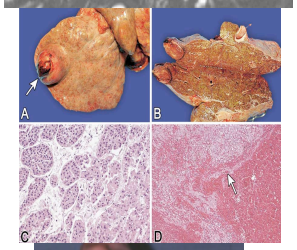


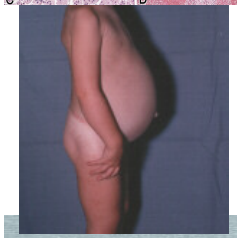
Hôpitaux de Bordeaux

De Leval M, Kilner P, J Thorac Cardiovasc Surg 1988

Problèmes Post Fontan

- 1. Obstruction des Connections
- 2. Dysfonction ventriculaire (progressive)
- 3. Dysfonction VAV systémique
- 4. Obstacle à l'éjection VU
- 5. Dilatation atriale
- 6. Compression VP
- 7. Thrombus
- 8. Cyanose due à
  - • Fenestration ou fuite patch
  - • Collatérales AV
  - • Malformations pulmonaires artérioveineuses
  - • CollateralesVéno-veineuses
- 9. Arythmie (50% à 10ans; PM 5%)
- 10. Intolérance à l'exercice
- 11. Dysfonction endothéliale
- 12. Protein Losing Enteropathy
- 14. Dysfonction hépatique
- 15. moules bronchiques

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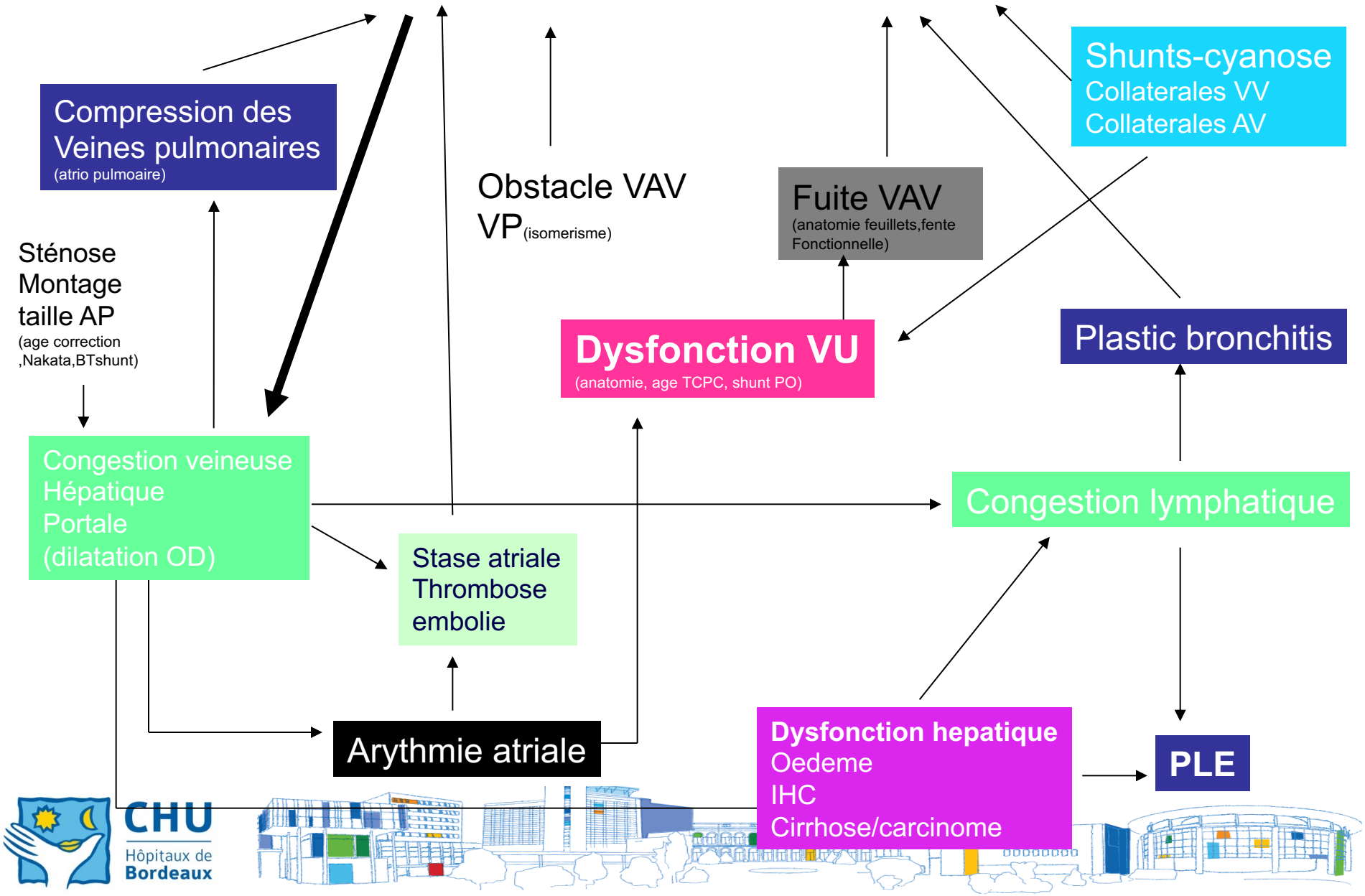
Elévation résistance cavo (atrio)pulmonaires



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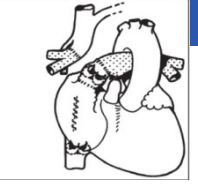
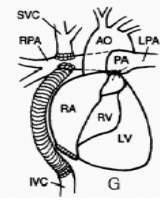
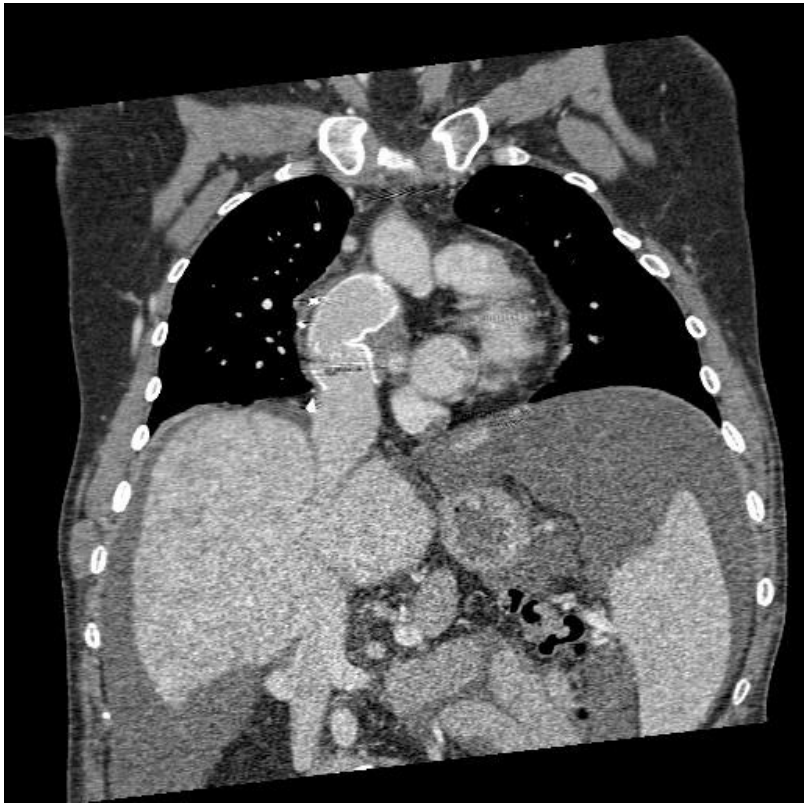
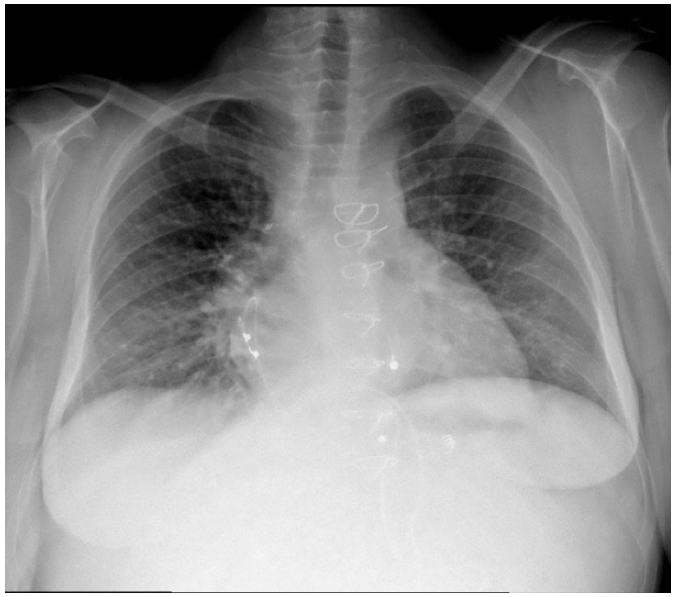


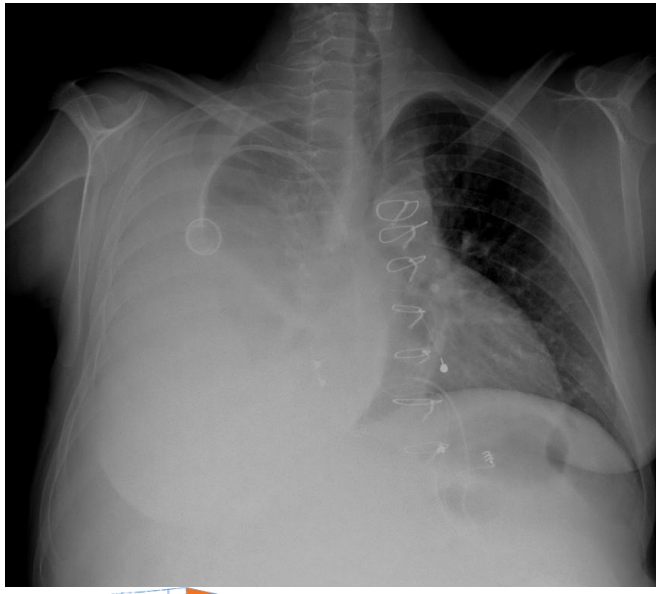


Stratégie diagnostique et thérapeutique du « failing Fontan »

- Evaluation anatomique et hémodynamique détaillée
- Traitement des lésions obstructives du circuit
- Traitement précoce des arythmies et maintien du rythme sinusal
- Traitement symptomatique de la PLE et bronchite plastique
- Gestion des RVP et RVS
- Conversion chirurgicales des anastomoses atriopulmonaires+ chirurgie ryhtmique
- transplantation







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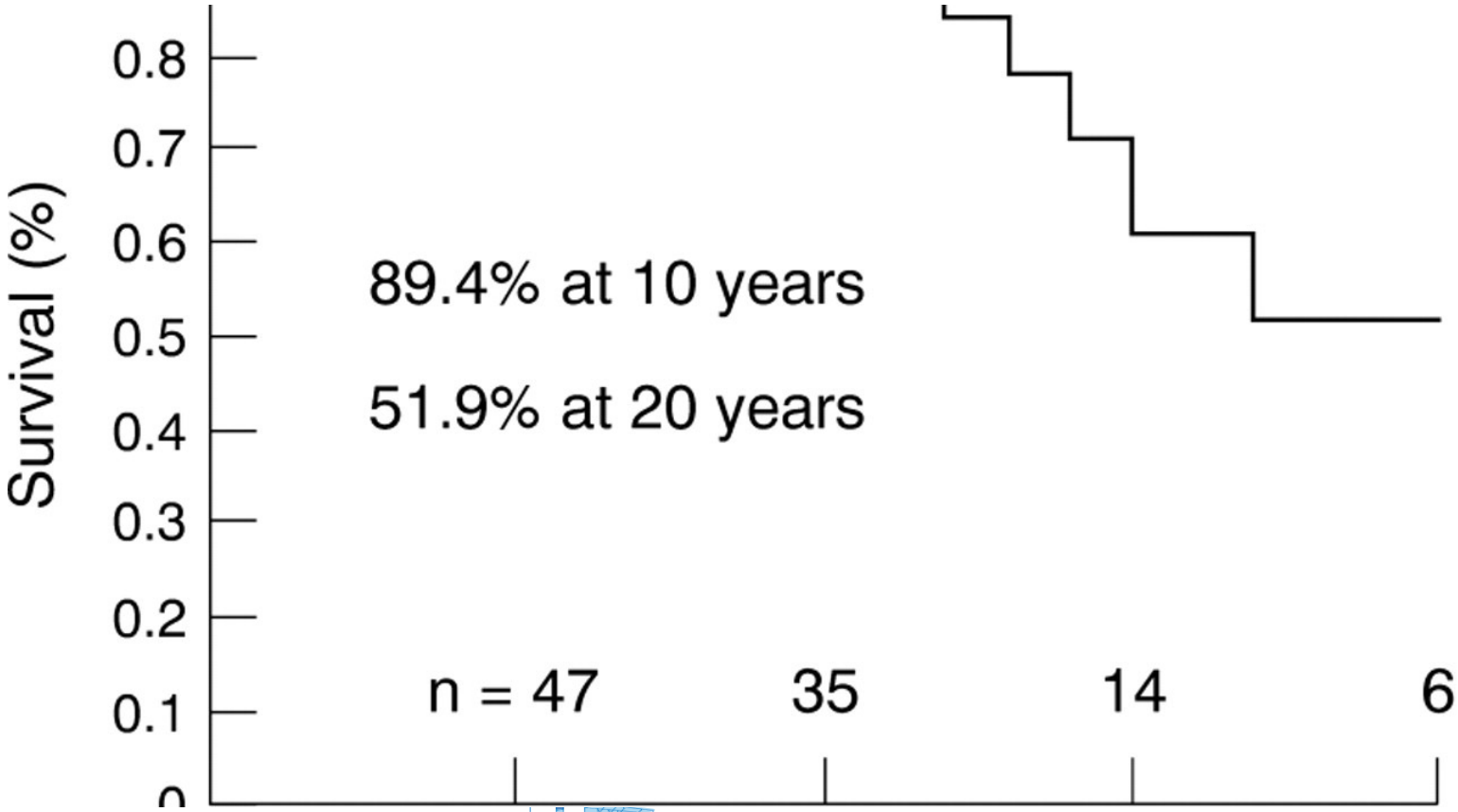
Bordeaux

Courbe de survie des VU admis à l'âge adulte en centres spécialisés GUCH



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Comment améliorer cette évolution?

- Préserver l'arbre pulmonaire+++
- Préférer la DCPP au Blalock
- Dérivation cavopulmonaire totale par tube extra-cardiaque fenestré pour améliorer le post-opératoire immédiat
- Fermeture de la fenêtre dans les situations favorables

Les inconnues

- Le meilleur âge pour la DCPT
- Les csq d'un flux veineux continu sur les fonctions hépatiques, surtout si les pressions sont élevées
- Savoir poser l'indication de transplantation à temps lorsque la situation est précaire

Stratégie thérapeutique: sidénafil?

Limites de la physiologie type Fontan

- Défaut de précharge: rôle de la circulation pulmonaire
- Paradoxe du niveau de PVC chronique
- Élévation des resistances pulmonaire: rôle de la pression pulsée

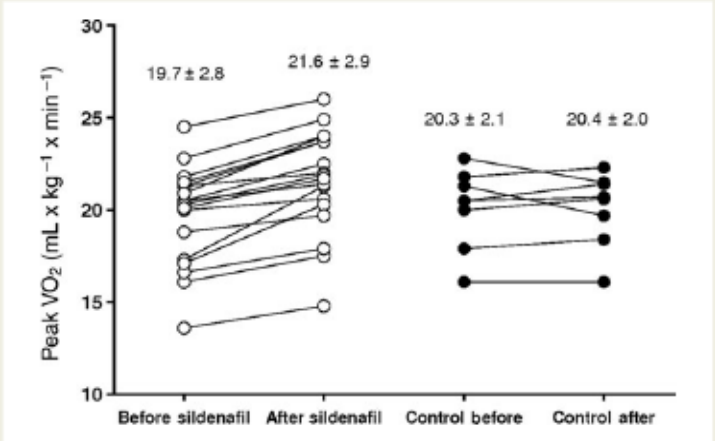

Figure 3 Individual improvement in peak VO_2 observed in the sildenafil treatment group (on the left side) and in the control group (on the right side). Data are presented as mean ± SD. VO_2 indicates oxygen uptake.

Giardini A. European Heart Journal 2008
Hager A. European Heart Journal 2008

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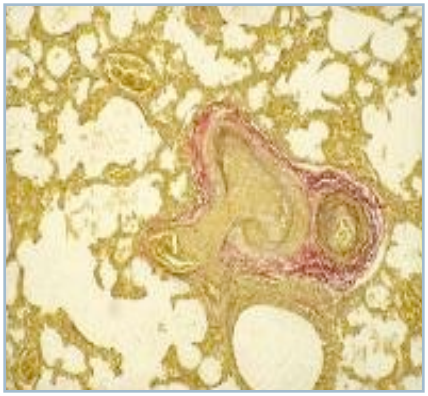
Dysfonction du VU

- dilatation pré-op peu réversible
- Recherche de MAPCA (fréquent si cyanose)
- Remodelage:dysfonction systolique et diastolique
  - Intérêt de la Milrinone en PO
  - Intérêt des IEC en chronique (élévétion des taux circulants d'ADH, rénine ,AII, aldostérone)?
    - Pas de bénéfice clinique prouvé

Eisenmenger



Prolifération des CML
↗ Matrice extra-cellulaire
Thrombose intravasculaire

Shunt gauche-droite

↓

↗ du débit sanguin pulmonaire
"shear stress"

↓

Dysfonction endothéliale

↓

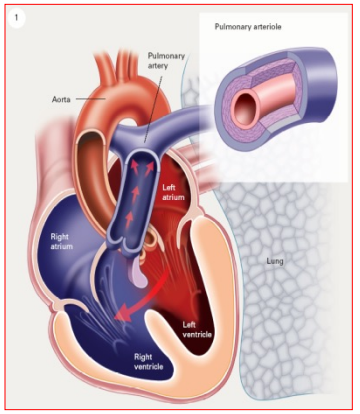
Augmentation des RVP

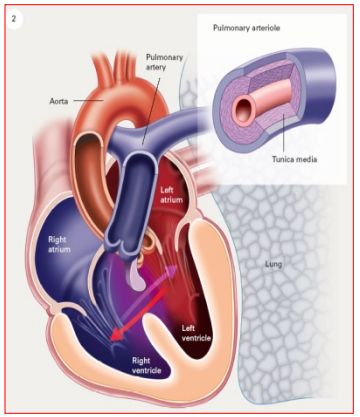
↓

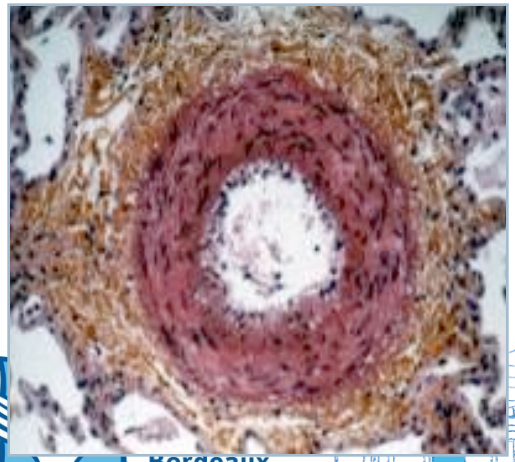
Inversion du shunt : droite-gauche

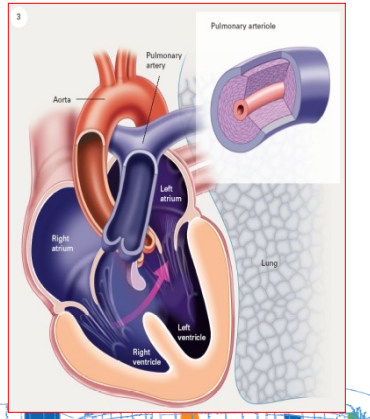
↓

Cyanose (Eisenmenger)









Bordeaux

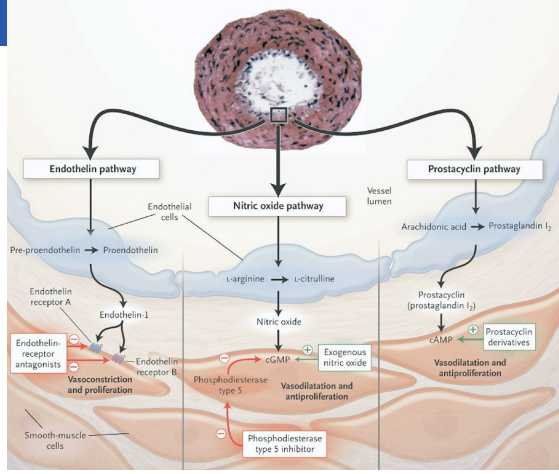
Beghetti M. Rev Mal Resp 2006

complications

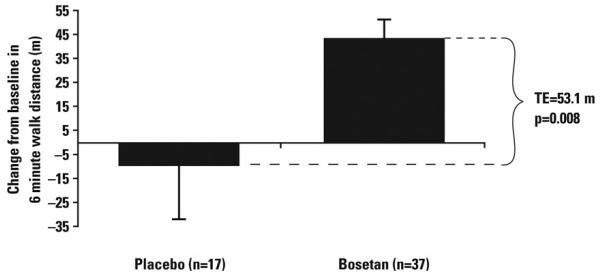
- Hyperviscosité
- AVC et autres thromboses(0-14%)
- Abcès cérébral (4%)
- Hémoptysie
- Dysfunction rénale
- Hyperuricémie, crise de goutte
- Lithiases biliaires
- Troubles de la coagulation
- Mort subite

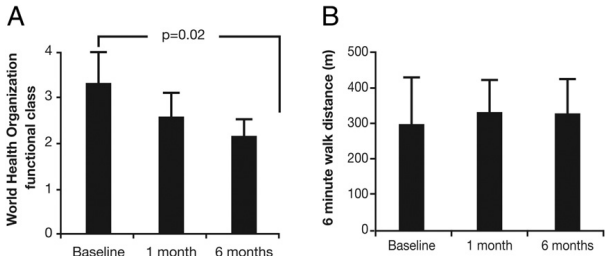
Beghetti JACC 2009
Galie N Eur Heart J 2009
Vongpatanasin W Ann int med 1998

Thérapeutique



- Traitement conventionnel
  - Diurétique?
  - Anticoagulants?
  - Inh Ca+?
  - O2
- Bosentan (Breathe 5)
- Sitaxentan
- Sildenafil
- Prostacycline





Humbert et al. NEJM 2004
Galie N et al. Circulation 2006
Chau et al. Int J cardiol 2007
Beghetti M et al. JACC 2009

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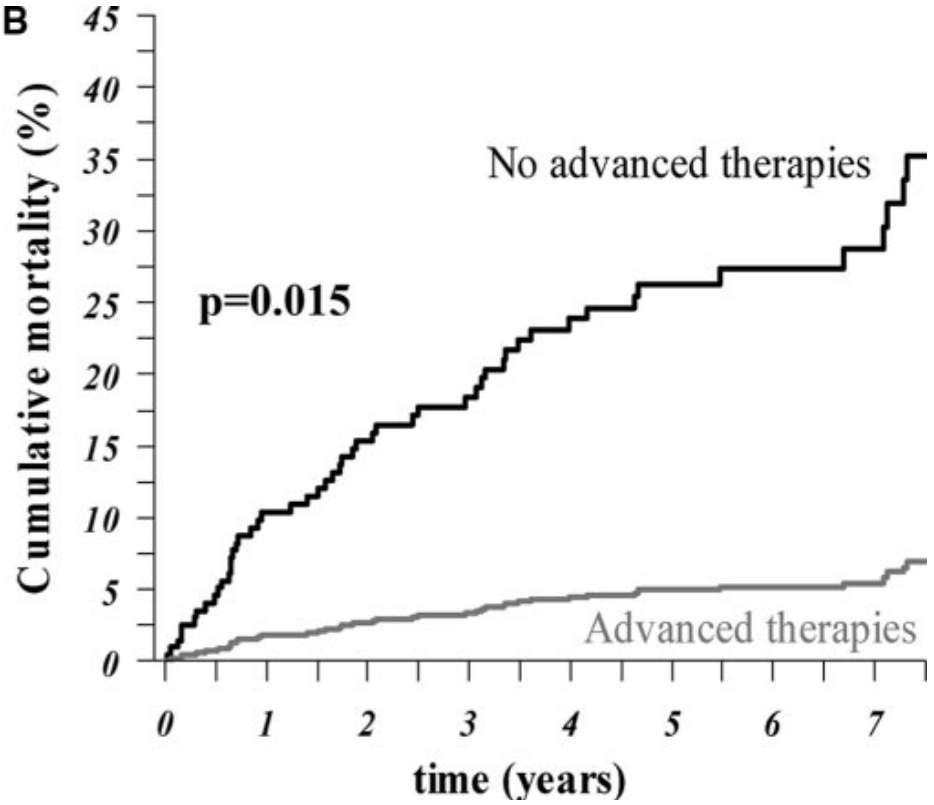
Ourien?

Eisenmenger

- Traitement spécifique
  - NYHA II ou III
  - Mono/bithérapie
  - Critères de suivi



Dimopoulos K et al. Circulation 2010

Gestion de la polyglobulie

Stratégie thérapeutque: Réduction de l'Ht à volémie constante

- Phlébotomie?
  - indication:symptomes cliniques
  - inconv: hyperviscosité paradoxale à Ht constant++++
- Supplémentation martiale (Foalte B12): monitoring+++
- Inhibiteur synthèse érythrocytaire : hydréa
  - Absence d'essai randomise/durée/ risque cutané???

Ammash N et al. JACC 1996

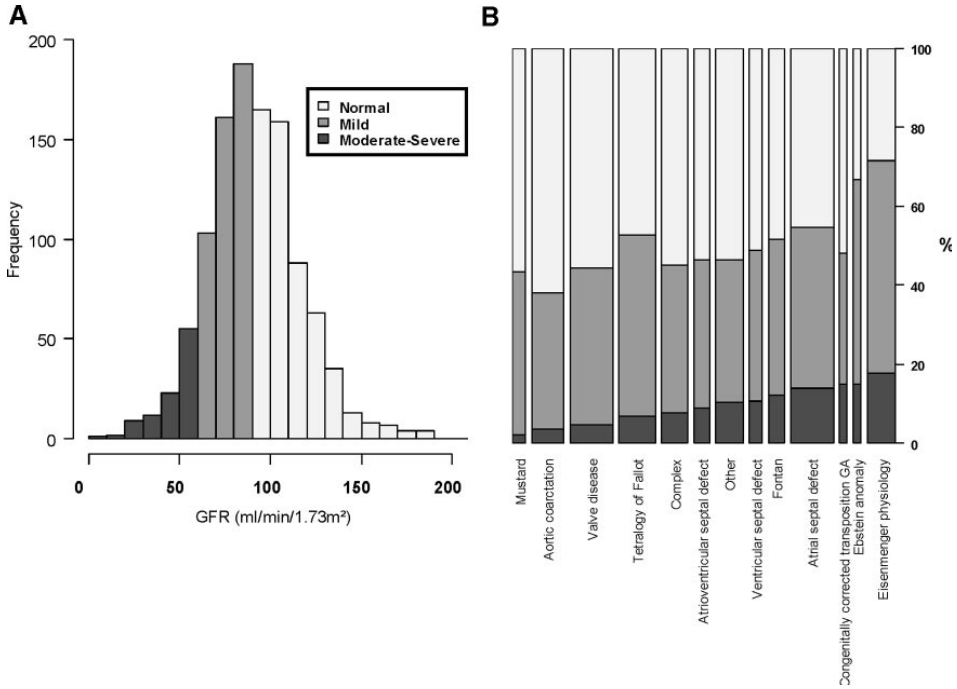
Endocardite infectieuse

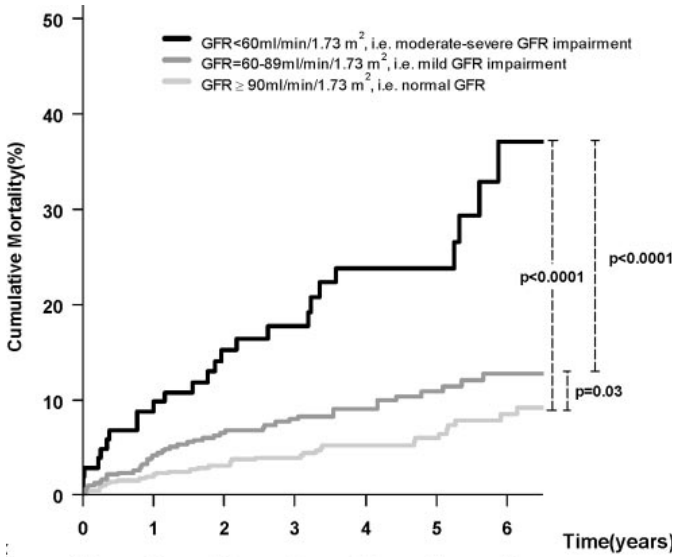
- Incidence. 15–140x supérieure à la population générale
- Prévalence: 2 à 18% (nette prédominance masculine)
- Germes habituels (streptococcus et staphylococcus)
- Mortalité: 4–10%. Meilleur pronostic que dans les cardiopathies acquises (grande proportion d'endocardite du coeur droit).
- antibioprophylaxie seulement dans les groupes à haut risque.

Recommendations: prophylaxis	Class[a]	Level[b]
Antibiotic prophylaxis should only be considered for patients at highest risk of IE 1. Patients with a prosthetic valve or a prosthetic material used for cardiac valve repair 2. Patients with previous IE 3. Patients with congenital heart disease a. cyanotic congenital heart disease, without surgical repair, or with residual defects, palliative shunts or conduits b. congenital heart disease with complete repair with prosthetic material whether placed by surgery or by percutaneous technique, up to 6 months after the procedure c. when a residual defect persists at the site of implantation of a prosthetic material or device by cardiac surgery or percutaneous technique	IIa	C
Antibiotic prophylaxis is no longer recommended in other forms of valvular or congenital heart disease	III	C

ESC guidelines 2009

Fonction rénale et GUCH





	HR	95% CI	*P*
Propensity score weight-adjusted models			
Moderate/severe impairment vs normal GFR	3.25	(1.54–6.86)	0.002
Moderate/severe impairment vs mild GFR impairment	1.72	(0.97–3.05)	0.06
Mild GFR impairment vs normal GFR	1.25	(0.69–2.27)	0.46
Multivariate models of death with diuretic as stratum			
Moderate/severe GFR impairment	2.31	(1.17–4.56)	0.015
Mild GFR impairment	1.22	(0.70–2.10)	0.48
NYHA class >2	2.50	(1.52–4.10)	0.0003
Any systemic ventricular dysfunction	2.66	(1.62–4.38)	0.0001
Previous reparative surgery/sternotomy	0.69	(0.41–1.14)	0.15
Previous palliative surgery	2.42	(1.45–4.03)	0.001
Age, y*	1.09	(0.92–1.29)	0.35
Cyanosis	1.16	(0.65–2.09)	0.61

- 1102 patients. Age moyen 36 ans
- 41% d'IRC mineure
- 9% d'IRC modérée à sévère
- FDR
  - Cyanose
  - IC

Hôpitaux de Bordeaux

Dimopoulos et al. circulation 2008

Conclusions « ère de la perspective et de l'innovation » et de l'organisation

- 80-85% des patients atteignent l'âge adulte: nouvelle spécialités cardiologique
- Désir de vie normale
- Organisation autour de cardiologues référents et d'unités spécifiques
  - Centre de compétence: imagerie, EP, chirurgie, anesth
  - Réseau cardio-obstétrique
  - Réseau cardio-anesthésie chirurgie

Fin