



How do I follow my patients after an atrial switch for TGA?

Laurence Iserin

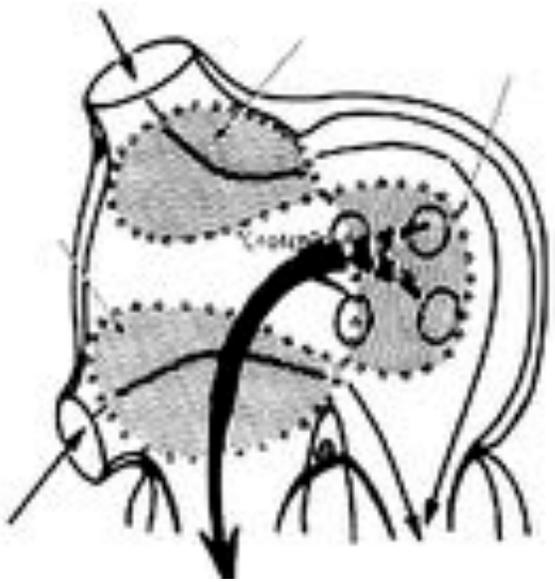
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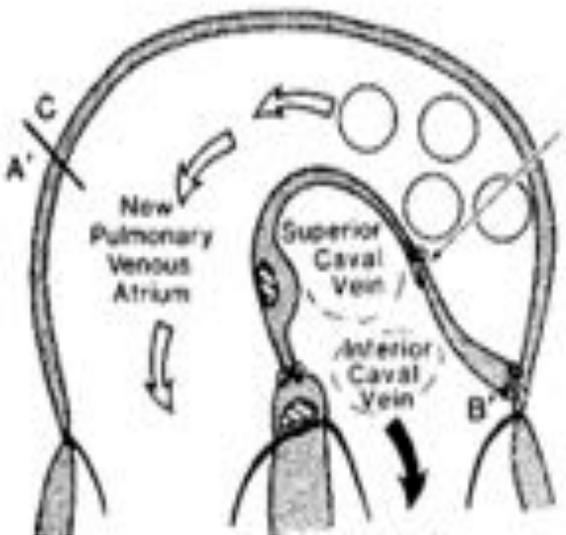
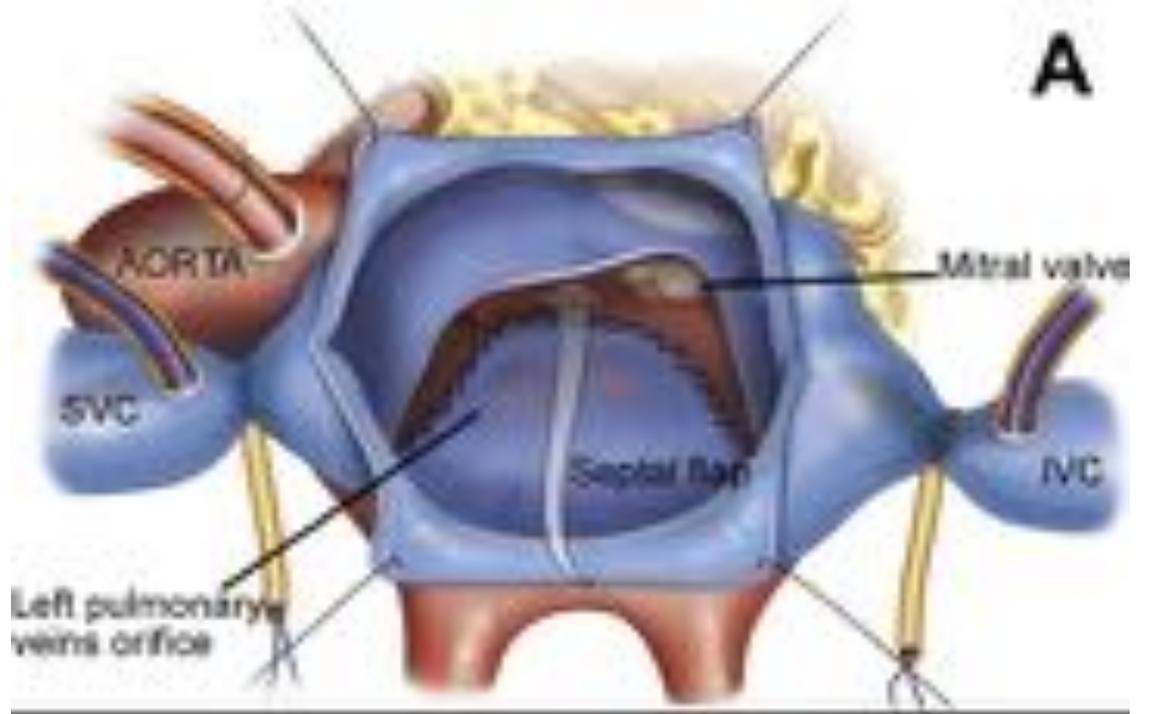
Centre de Référence Maladies Rares

Malformations **C**ardiaques **C**ongénitales **C**omplexes-M3C
CARDIOGEN

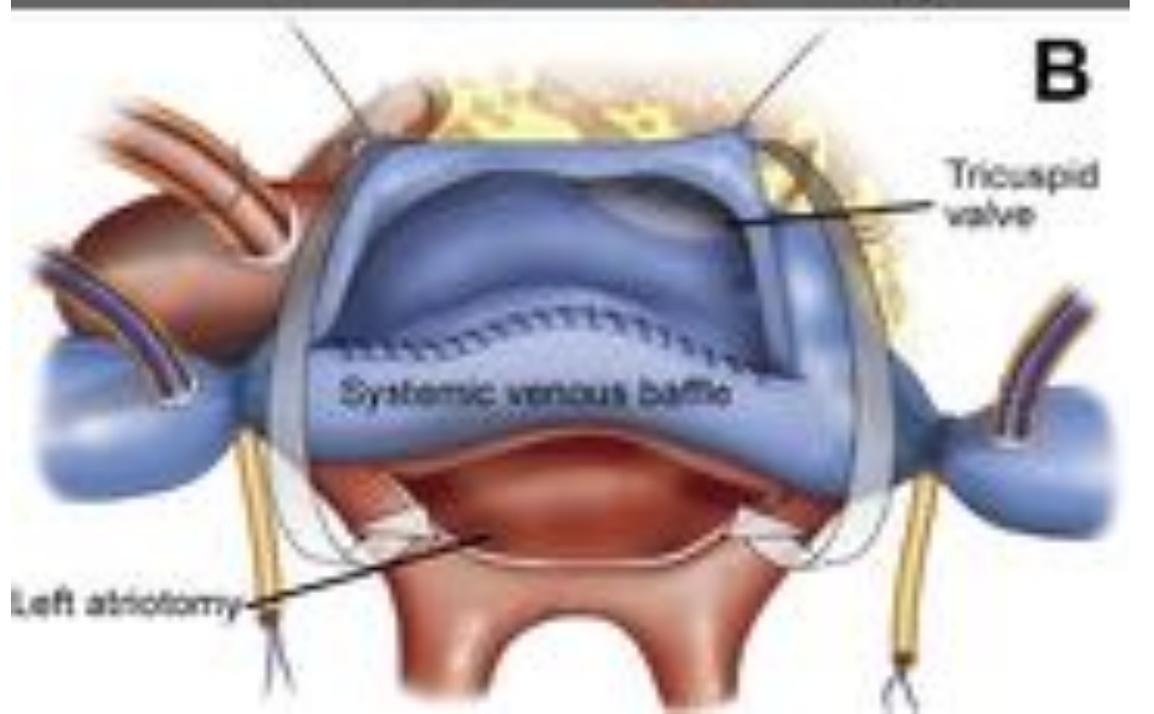




Mustard

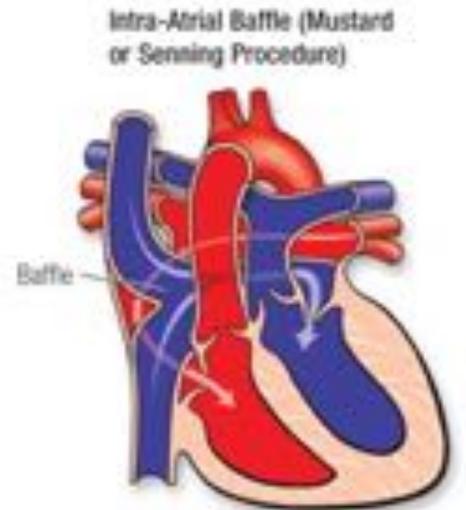
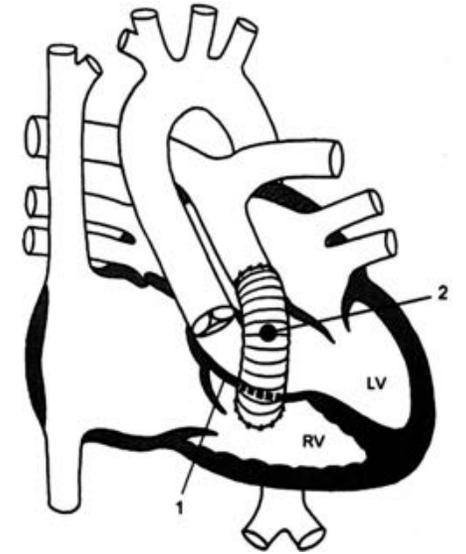
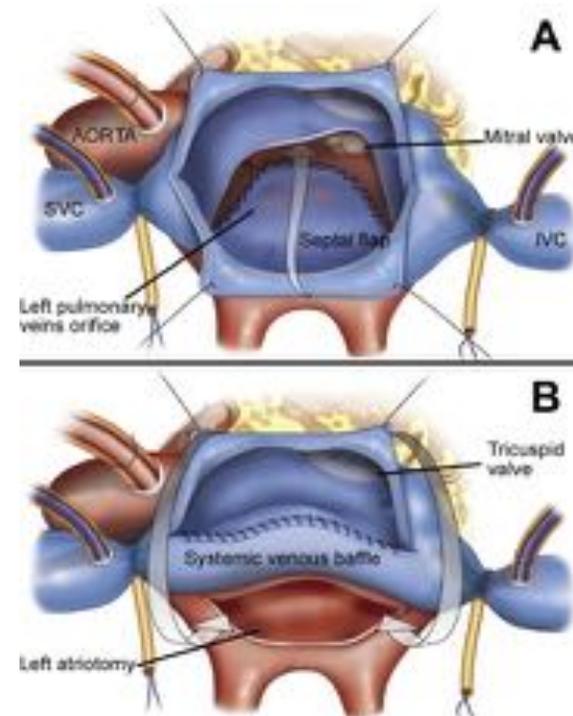


Senning

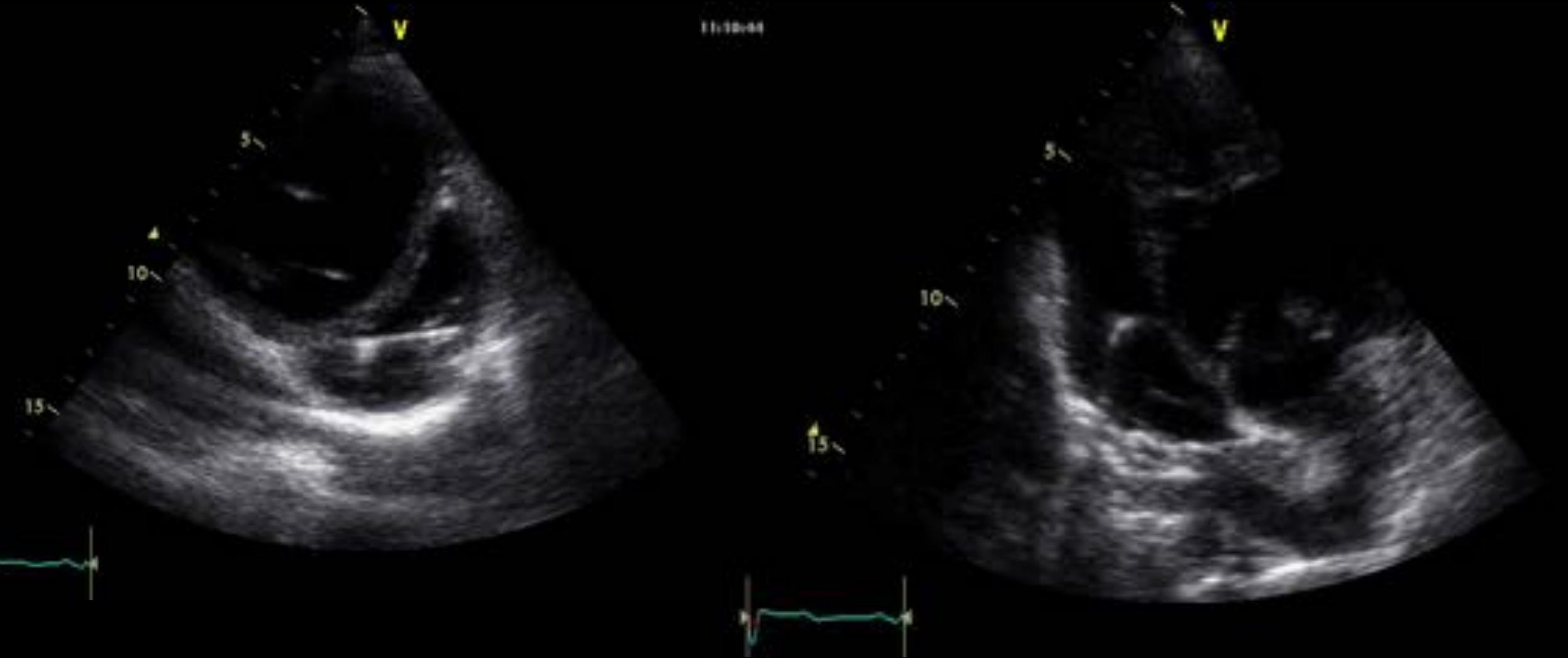


Corrections chirurgicales

- Rastelli TGV et CIV
 - En cas de CIV large: technique de choix en première intention
 - Complications:
 - Sténose et détérioration du tube VD-AP
- Mustard ou Senning avant 1980



ECHO after Mustard



Complications de la pathologie

- Décès

- 16% dus à
 - Défaillance cardiaque
 - Arythmies

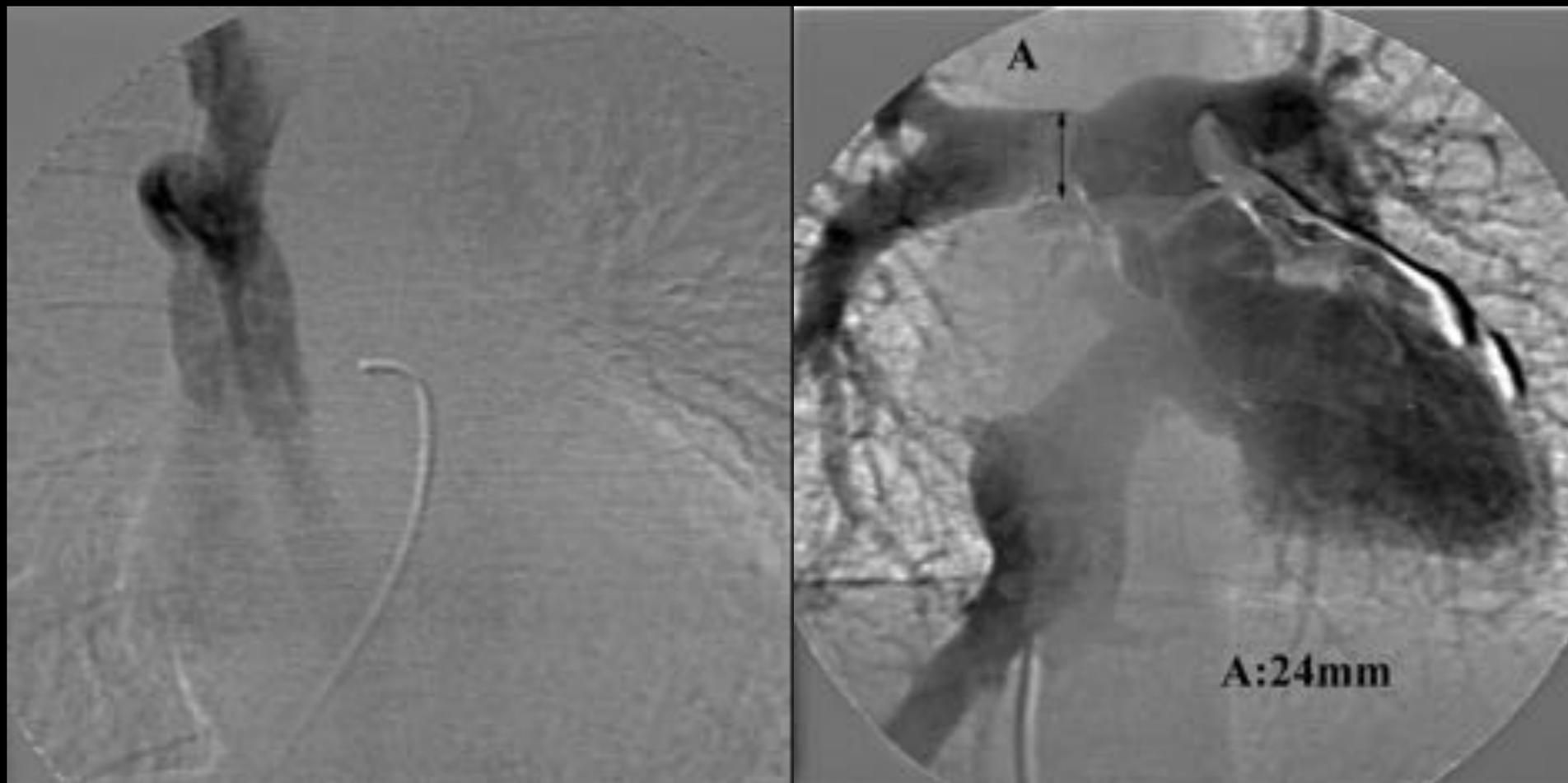
- Arythmies

- Supraventriculaires
 - BradyFA et tachyFA (60% à l'âge de 20ans)
 - Flutter (14%): de mauvais pronostic (FDR de mort subite)
- Dysfonction sinusale chez l'adulte (11% de PM)
 - » Restauration du rythme++++

Complications de la pathologie

- Dysfonction du VD morphologique:
 - Évaluée en echo coeur:
 - dP/dT tricuspide, appréciation qualitative strain
 - Traitement: peu de preuve:
 - » IEC
 - » Béta-bloquant: avec précaution car:
 - pas de preuve de leur bénéfice
 - risque rythmique
- Insuffisance tricuspide:
 - Secondaire la plupart du temps à la dilatation de l'anneau
 - Endocardite

Intervention de Mustard



Sudden death and atrial switch

variable	Atrial Arrhythmias (n = 20)	No Atrial Arrhythmias (n = 44)	P
Op age (m)	19 (11–40)	12 (5–17.5)	.000 ^a
Age last review (y)	36 (31.5–40)	30 (28–37)	.013 ^a
Time post op (y)	34 (27.5–36)	30 (28–34.5)	ns ^b
RV function (moderate dysfunction)	21	22	.024
RV size (cm)	5.5 (5.0–6.0)	5.1 (4.6–5.8)	ns ^b

RV size (cm) using the base of the RV in the apical four-chamber view. Statistical comparison is generally P value using parametric comparison (†) for continuous and its square (–) for categorical variables.

Table E. Comparing the Patients with SCD to the Rest of the Population

variable	SCD (n = 6)	No SCD (n = 71)	P value
Age at operation (m)	23 (21.5–23.0)	14 (9–23.0)	.27 ^a
Age at last review (y)	29 (28.5–30.5)	33 (29–36)	ns ^b
Tricuspid regurgitation (moderate) n (%)	4 (100%)	20 (27%)	.003
RV dysfunction (moderate)	3 (50%)	61 (86%)	ns
Atrial arrhythmia	5 (100%)	22 (30%)	.004
Cardiac failure admission (†)	3 (50%)	11 (15%)	.04
ACE inhibitor use (†)	3 (50%)	38 (53%)	ns
Beta blocker use (†)	1 (20%)	14 (19%)	ns

Median (IQR) for age at operation in months (m) and age at last review in years (y), number (n) of patients. The statistics used were nonparametric comparison (†) for continuous and its square (–) for categorical variables.

ACE, angiotensin-converting enzyme; IQR, interquartile range; RV, right ventricle; SCD, sudden cardiac death.

Complications de la pathologie

- Sténose

- Des veines caves:

- Supérieure

- Risque de thrombose/obstruction à la pose des PM

- Inférieure

- Des veines pulmonaires:

- HTAP 7% des adultes survivant

- Autres FDR pour HTAP:

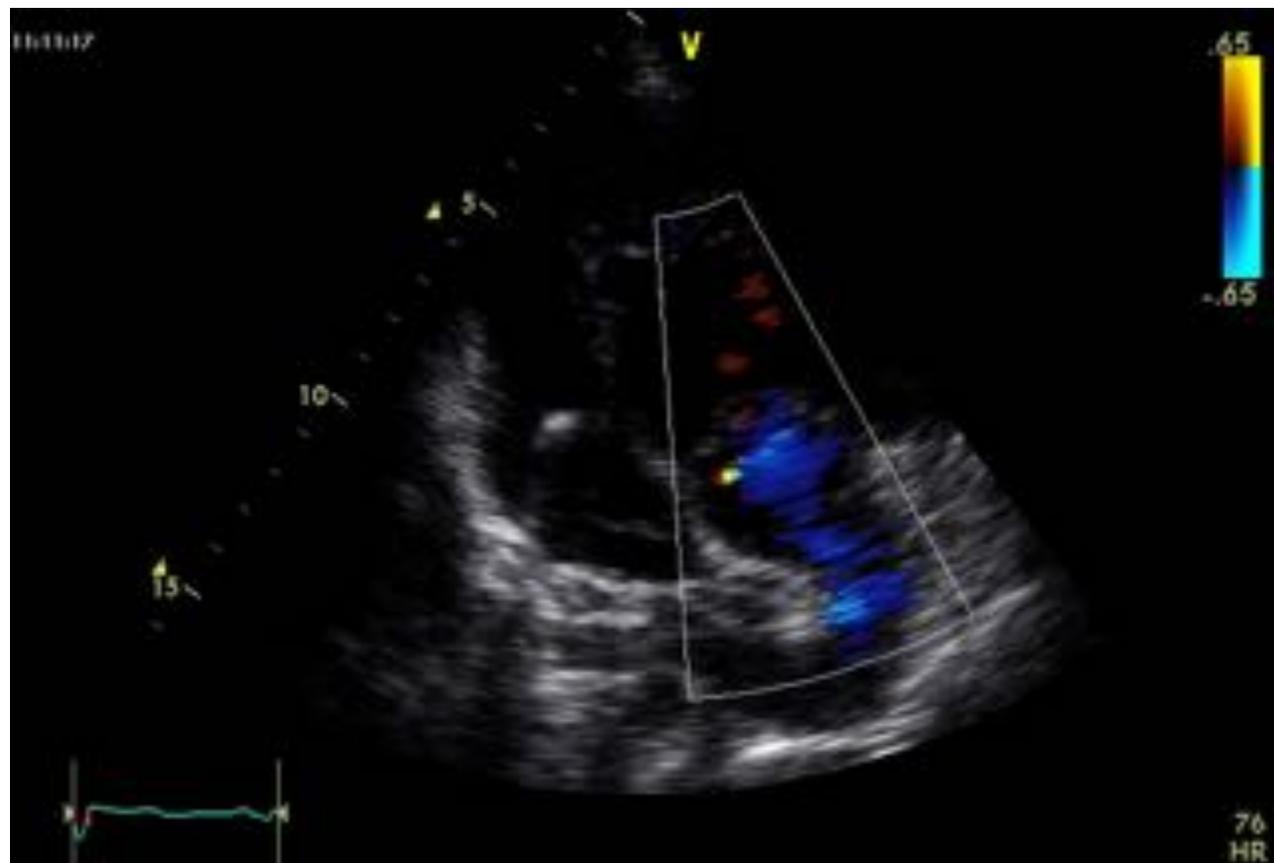
- Chirurgi>2ans

- Shunt ventriculaire ou des grosses vaisseaux

- Grossesse:

- A risque mais possible

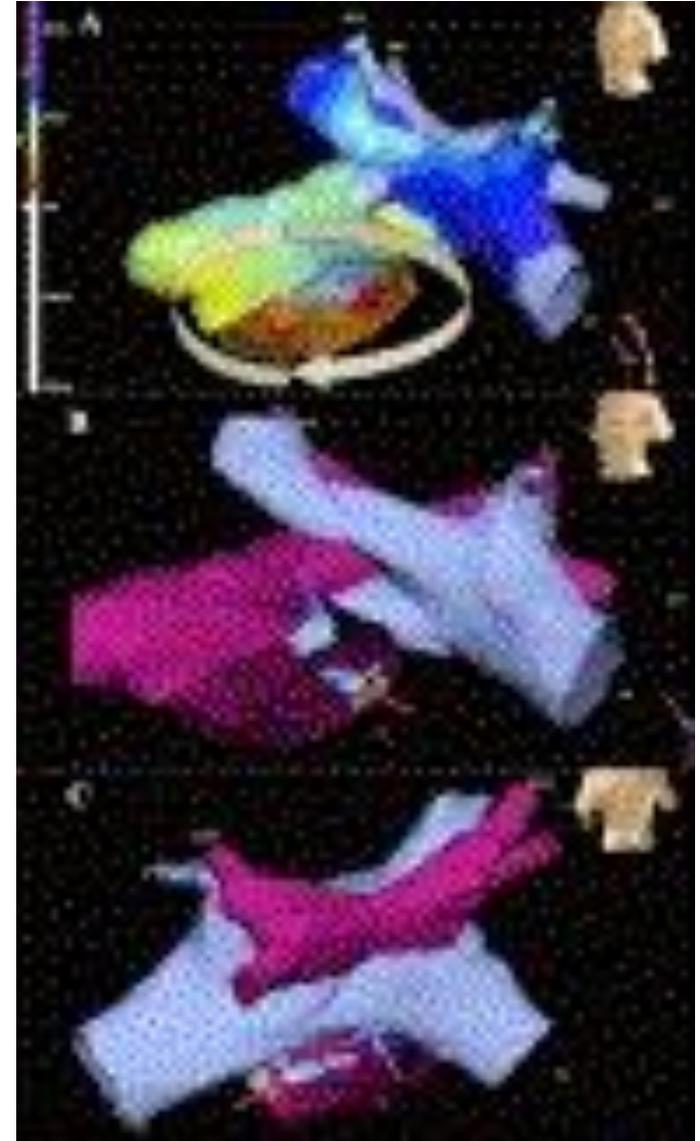
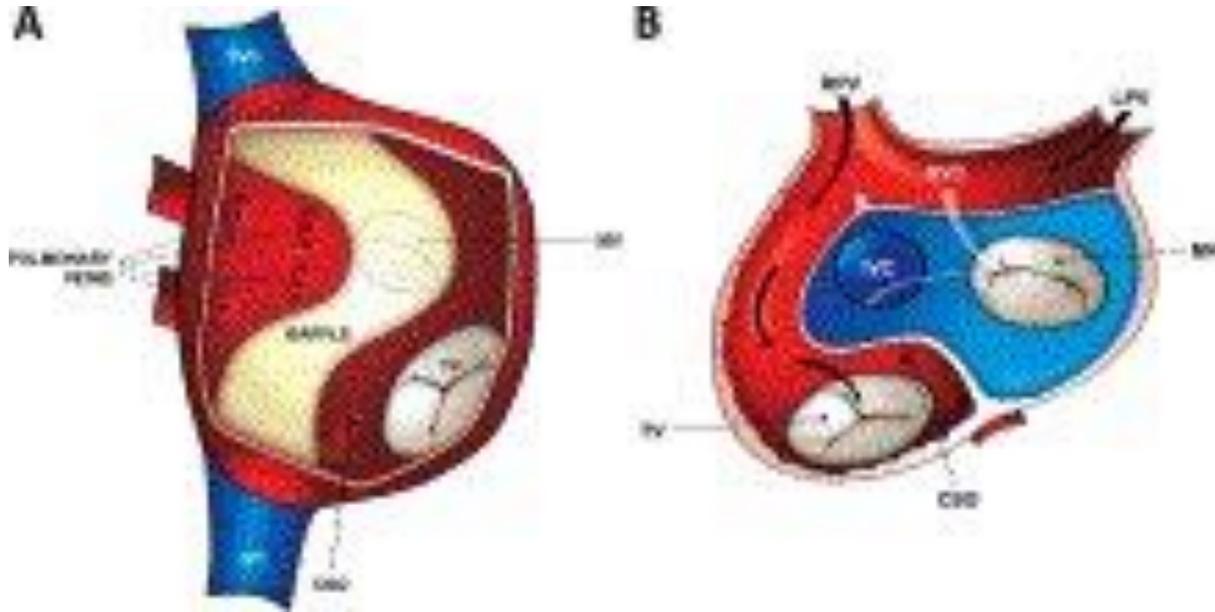
- Surcharge volémique induisant une dilatation VD morphologique, majoration de l'IT et du risque d'insuffisance cardiaque

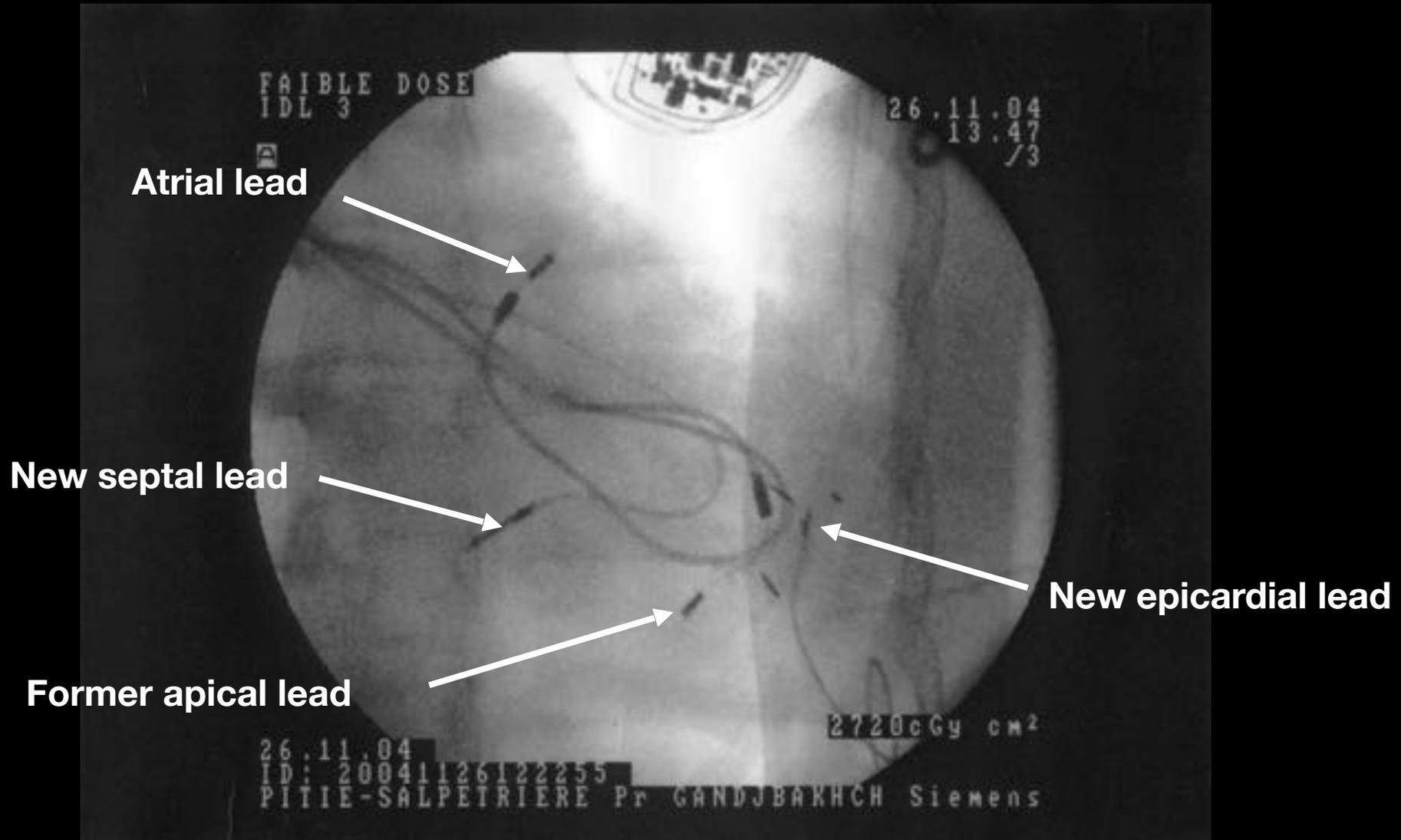


Suivi

- **REGULIER**
 - Annuel ou bi-annuel
 - ETT
 - IRM
 - Holter
 - Prophylaxie de l'endocardite: bof
 - Compte tenu de la CIV, des valvulopathies

Senning et Mustard : rythmologie de l'extrême





Senning resynchronisé

Background

The atrial switch operation for transposition of the Great Arteries (TGA) (Mustard or Senning Procedure) provides excellent short-term survival. Significant long-term concerns exist for these patients, however, including the ability of the right ventricle to maintain systemic perfusion and the risk of arrhythmia. We seek to describe long-term mortality and morbidity of this group of adult patients.

Methods

Consecutive patients who had undergone an atrial switch procedure, who were aged over 16 years and who were followed up at our tertiary level adult congenital heart disease (ACHD) service in Sydney, Australia since 2000 were included. We documented mortality using a National Death Index and analysed the prospectively defined composite endpoint of "Serious Adverse Events" including death, heart failure hospitalisation and/or documented ventricular arrhythmia.

Results

There were 83 patients included; mean age at most recent follow-up was 35 ± 5 years. Overall survival was 82% at 35 years and 22% of patients experienced a serious adverse event. Atrial and ventricular arrhythmias occurred in 45% and 7% of patients respectively. Eighteen (22%) patients required a pacemaker and six (7%) required implantable cardiac defibrillator (ICD) implantation. Significant right ventricular dysfunction was present in 26% of patients on their most recent visit and this, or requirement for permanent pacing, was associated with an increased risk of serious adverse events (OR 10.22, $p < 0.001$), (OR 4.998, $p = 0.04$) respectively.

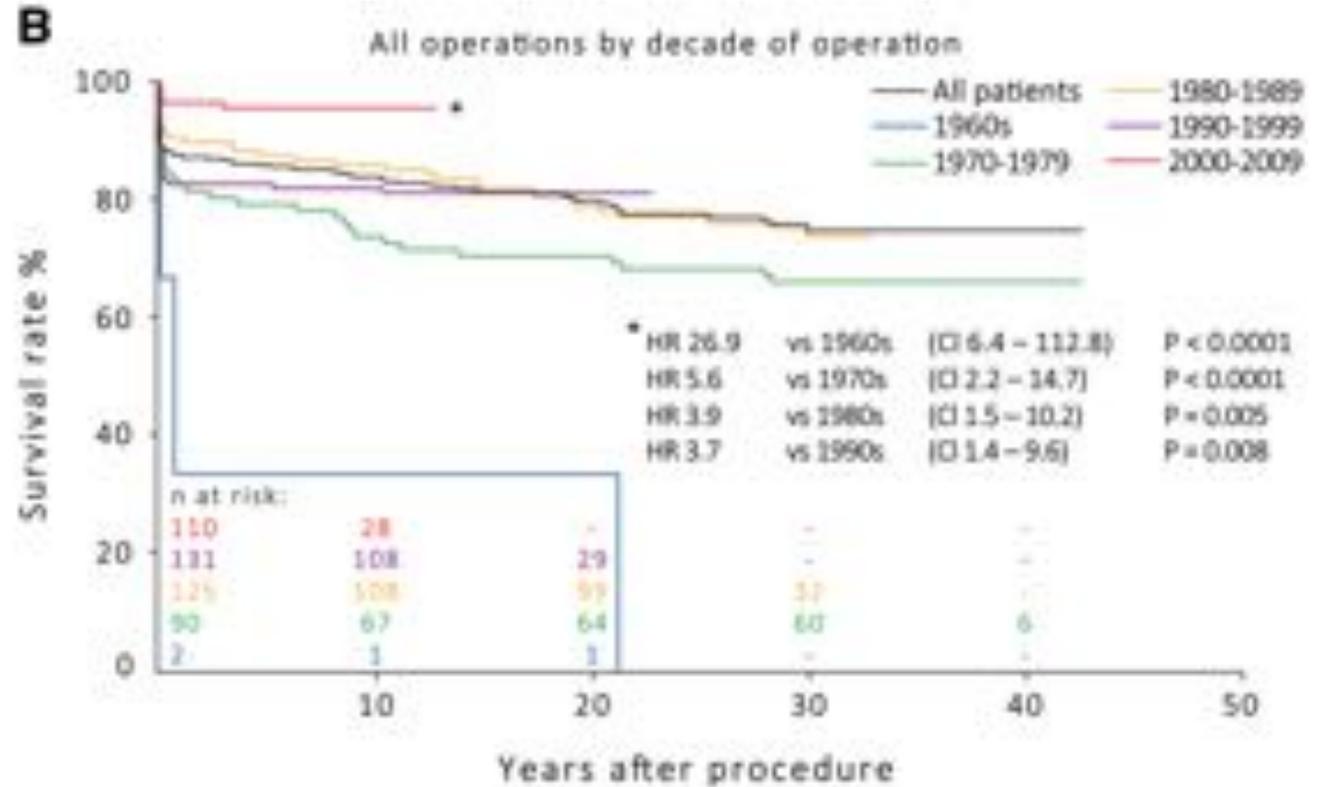
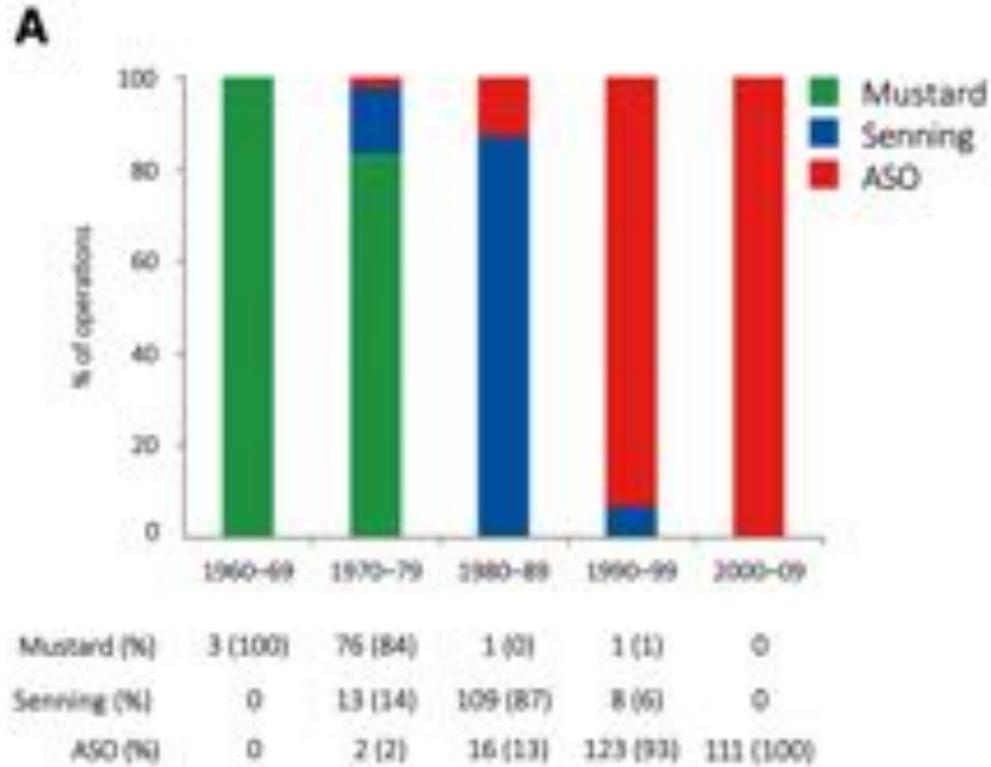
Conclusions

Significant mortality and morbidity accrues by mid-adult life after an atrial procedure for TGA. Right ventricular dysfunction and permanent pacing are associated with serious adverse events.

Keywords

Transposition of the great arteries • Congenital heart disease • Atrial switch

Outcomes after the Mustard, Senning and arterial switch operation for treatment of transposition of the great arteries in Finland: a nationwide 4-decade perspective

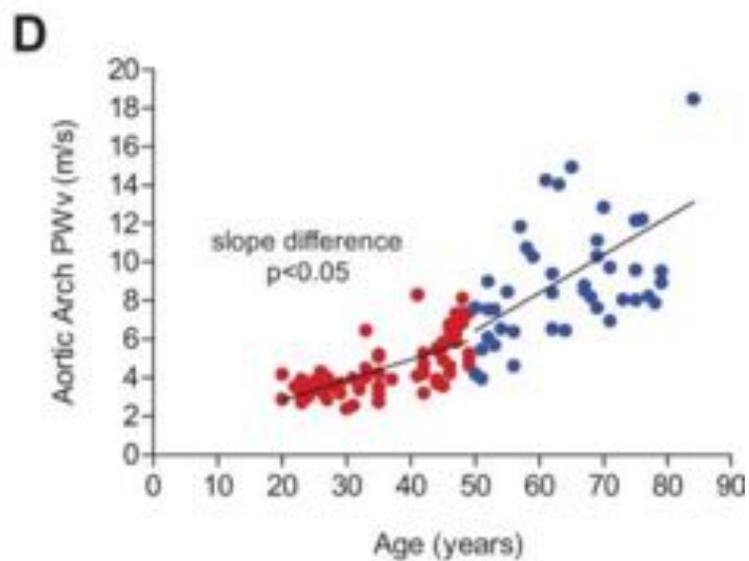
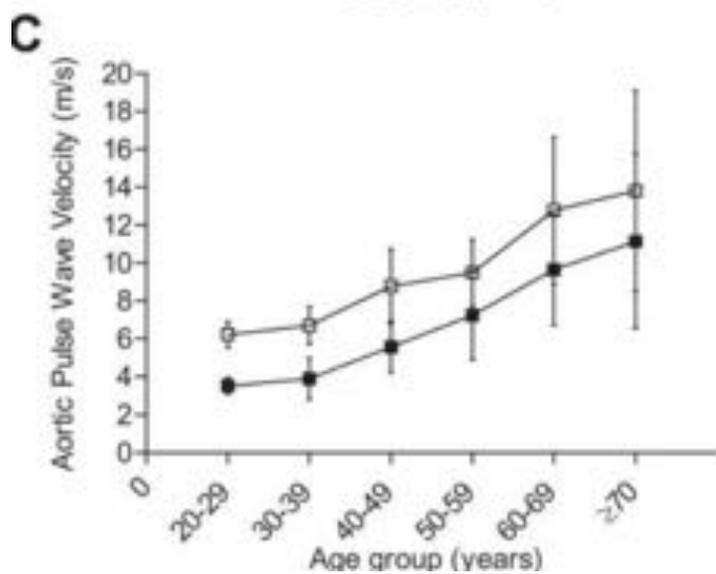
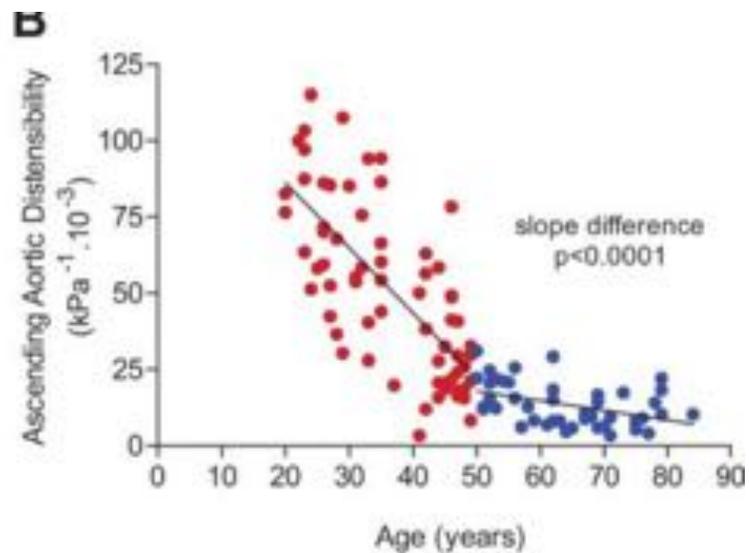
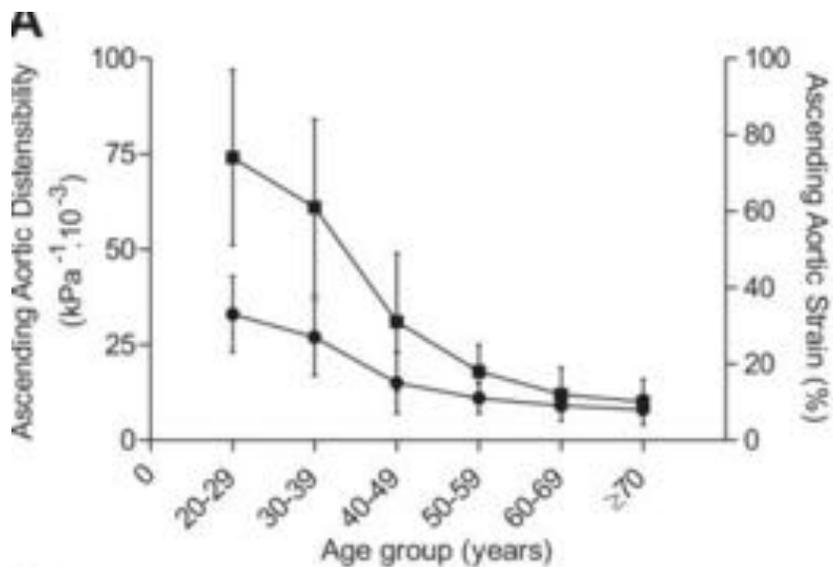


TGV switch Artériel: Goals of Imaging

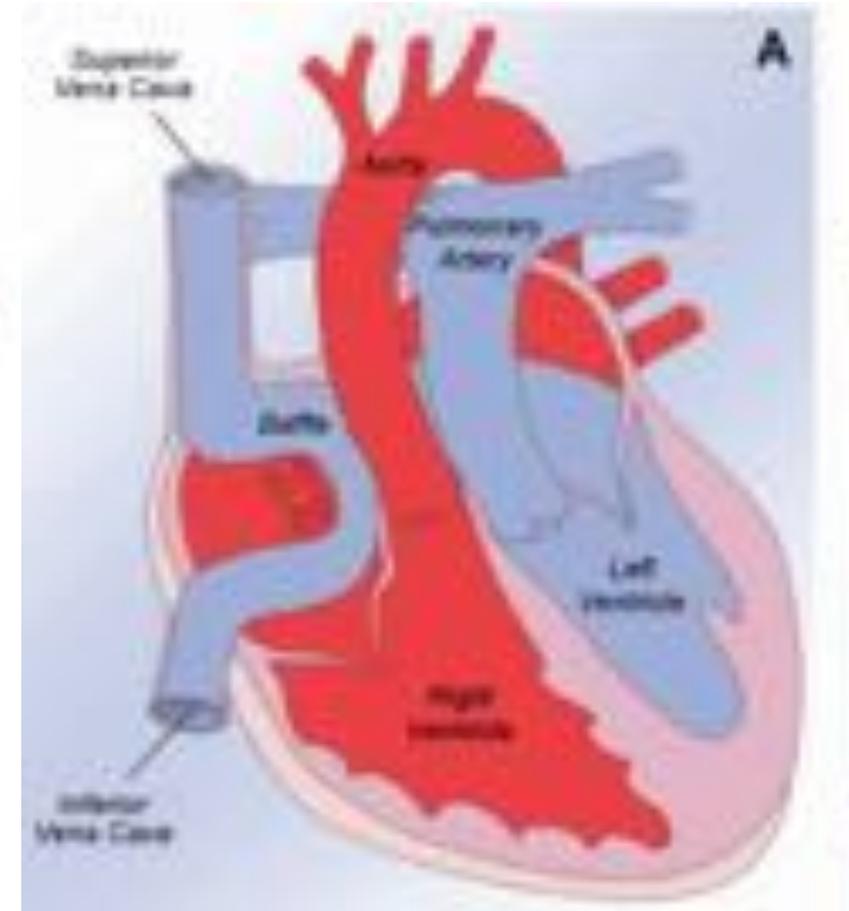
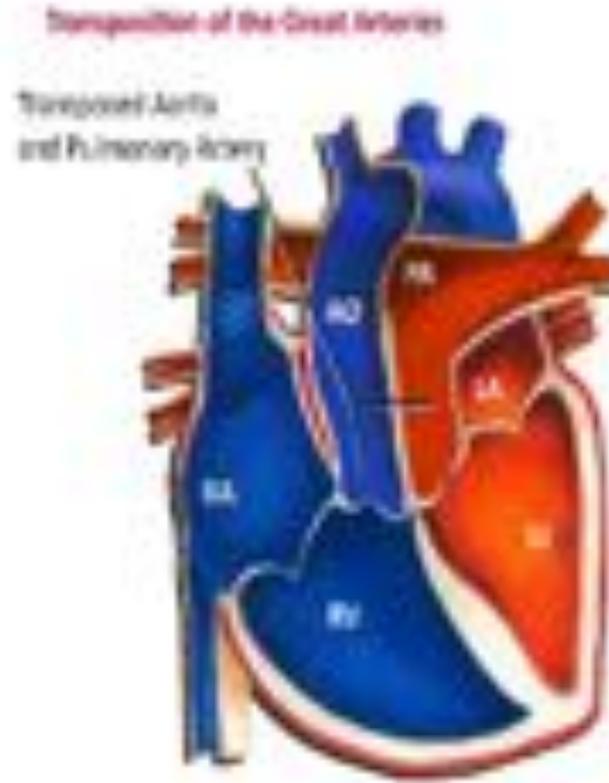
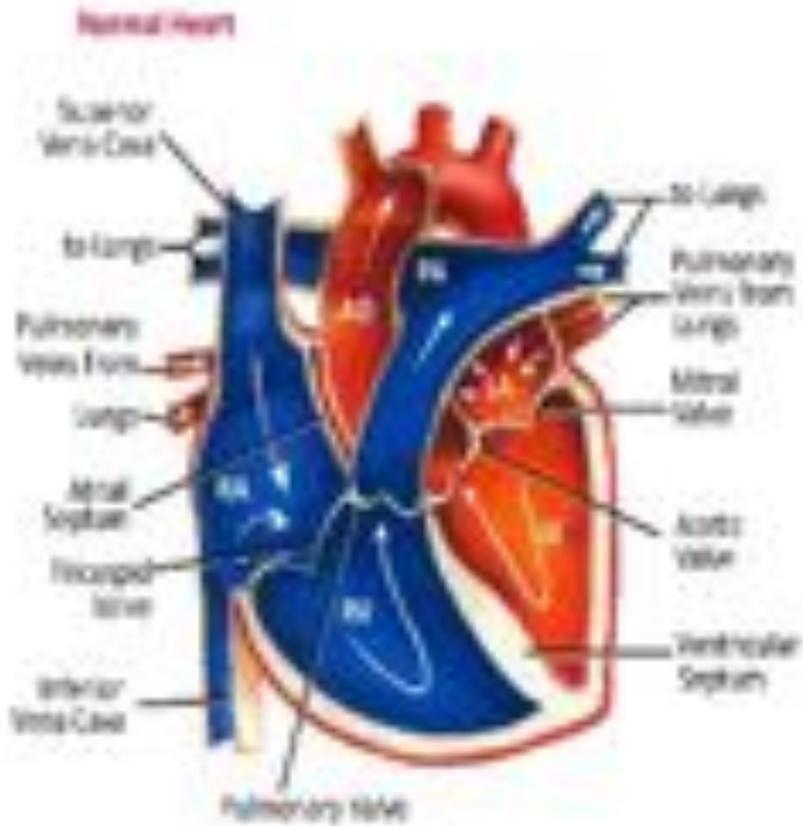
- Naissance et trajet proximal des coronaires
- Perfusion de stress (IRM)

- Etude de la dilatation de la Néo Racine Aortique / Fuite néo valve Aortique
- Anatomie AP et branches proximales
- Volumes et Fonction VG
- Shunt résiduel

Aging aorta



Transposition of great arteries: Atrial Switch

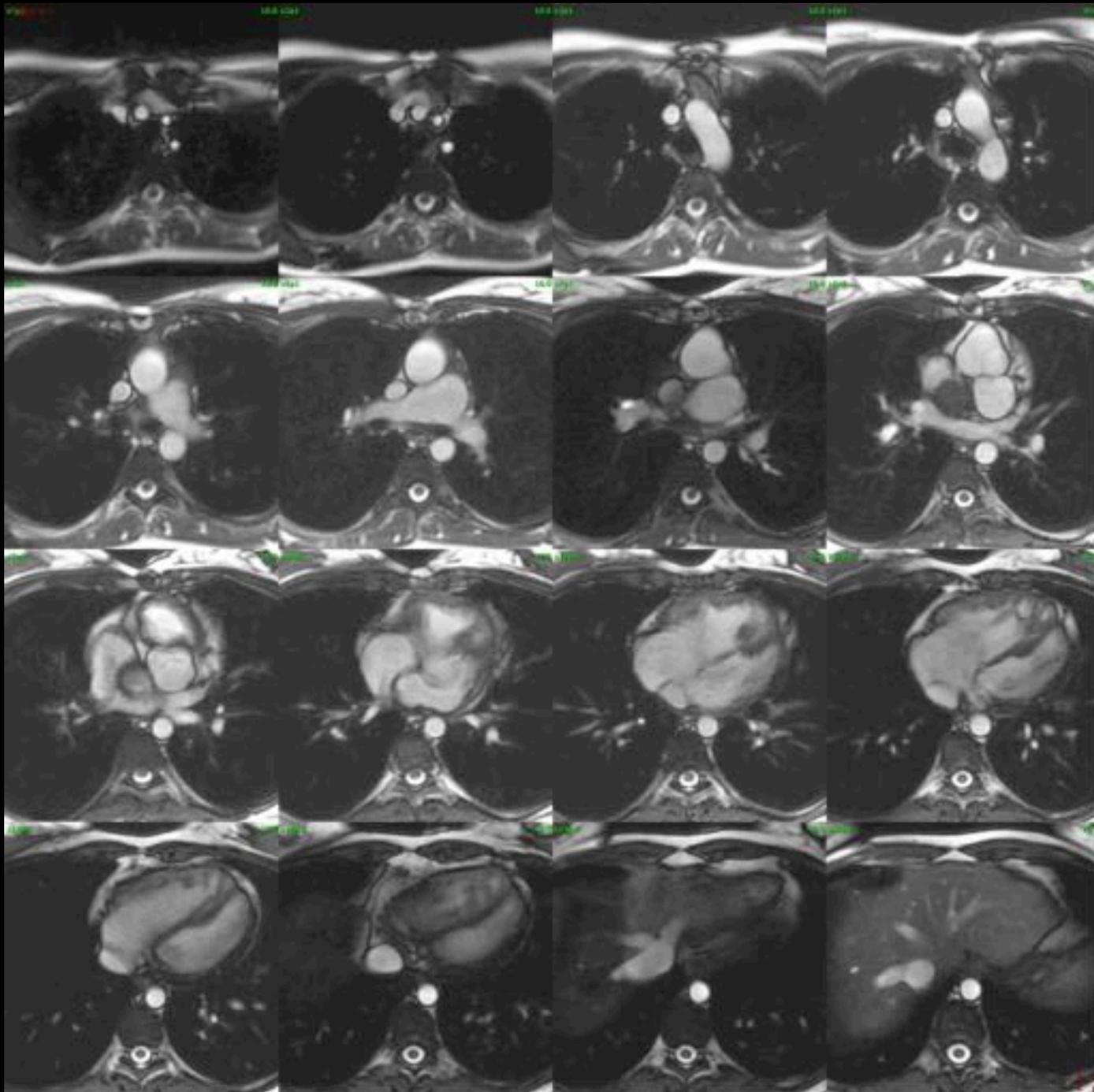


TGV switch Atrial: Goals of Imaging

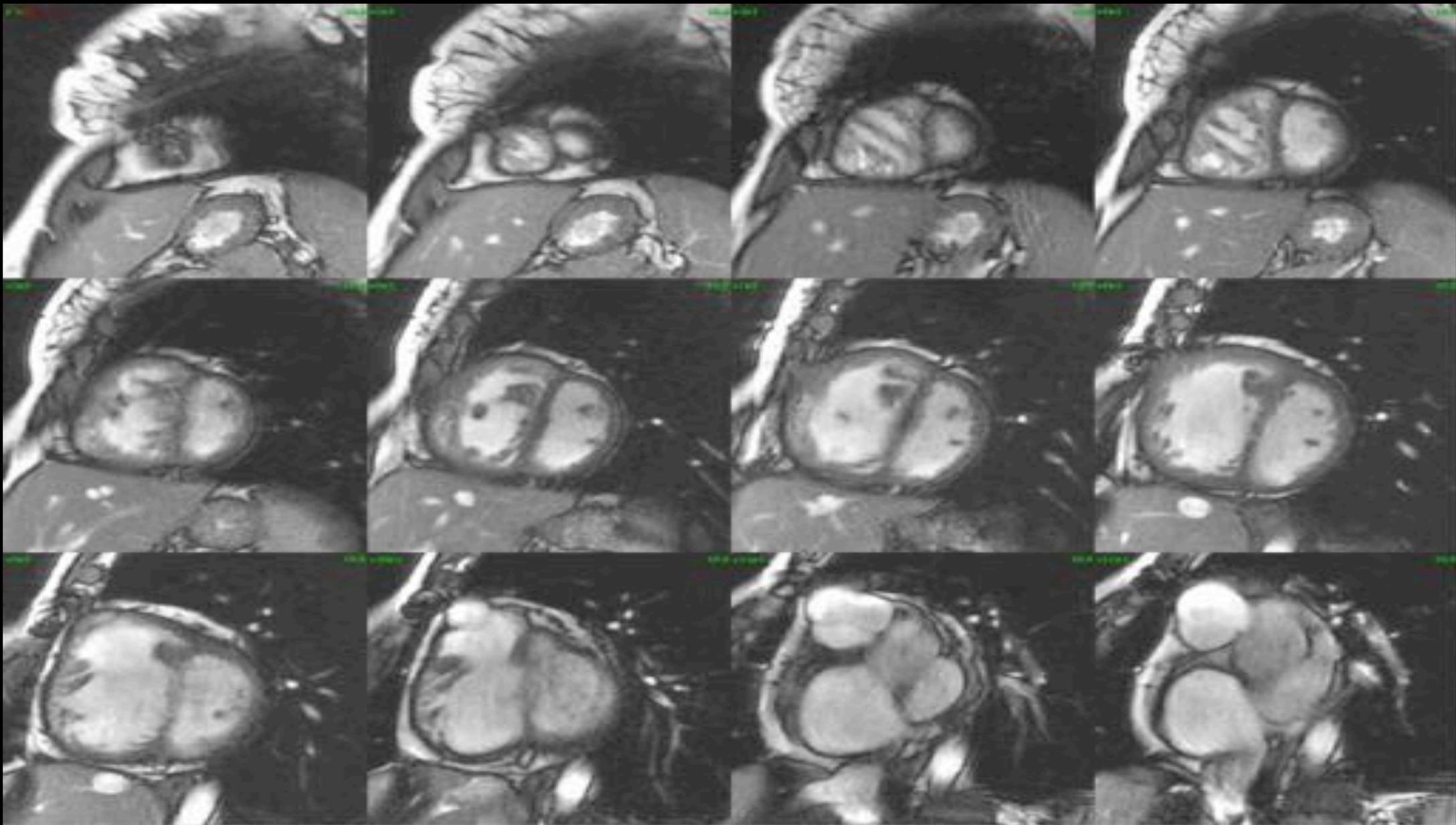
- Volumes et Fonction VD
- Anatomies chenaux veineux caves et pulmonaires (faux anévrysme, sténose)
- Shunt résiduel
- (Fuite tricuspide)
- Volume et fonction VG
- Naissance et trajet proximal des coronaires

- IRM +++
- Scanner si CI IRM

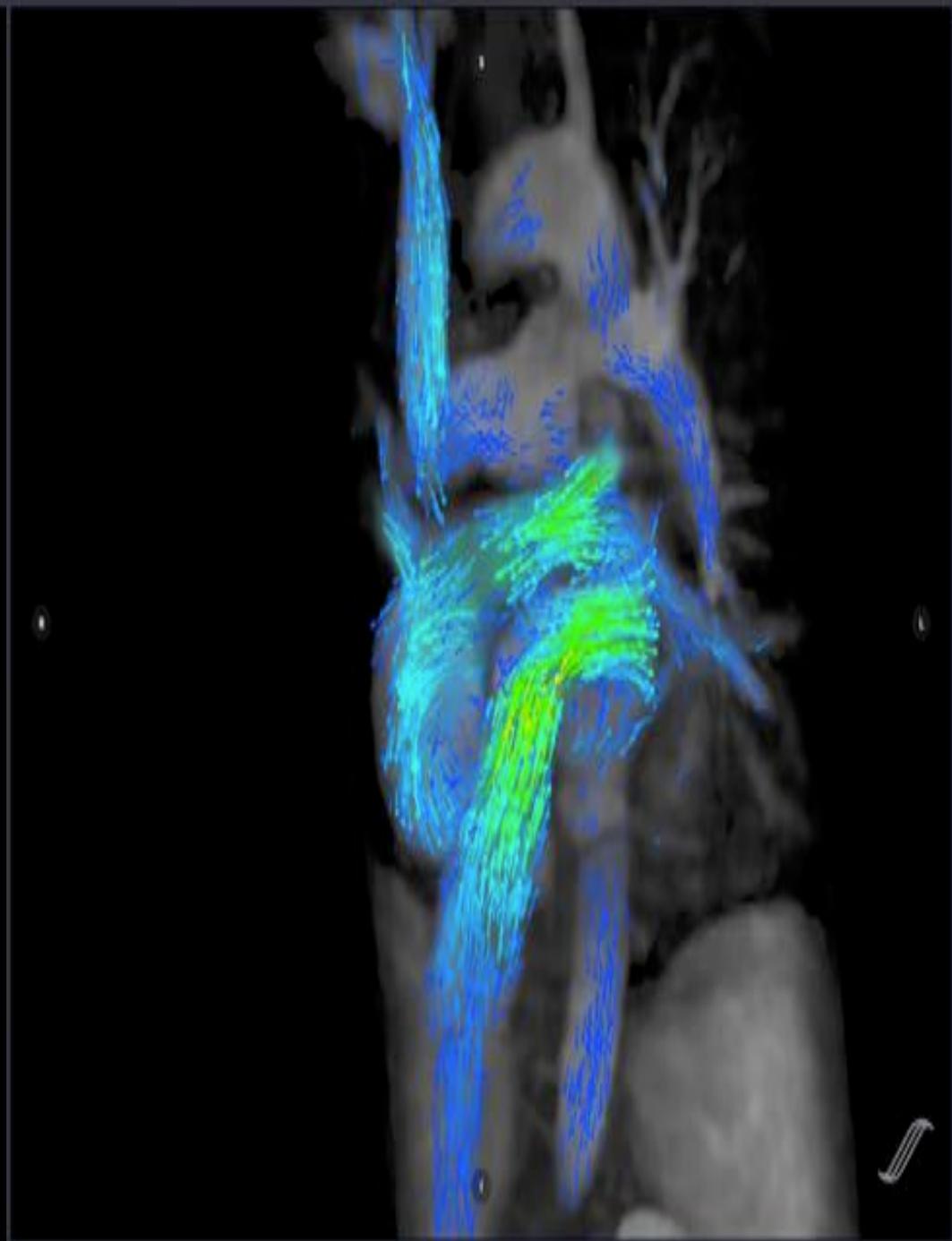
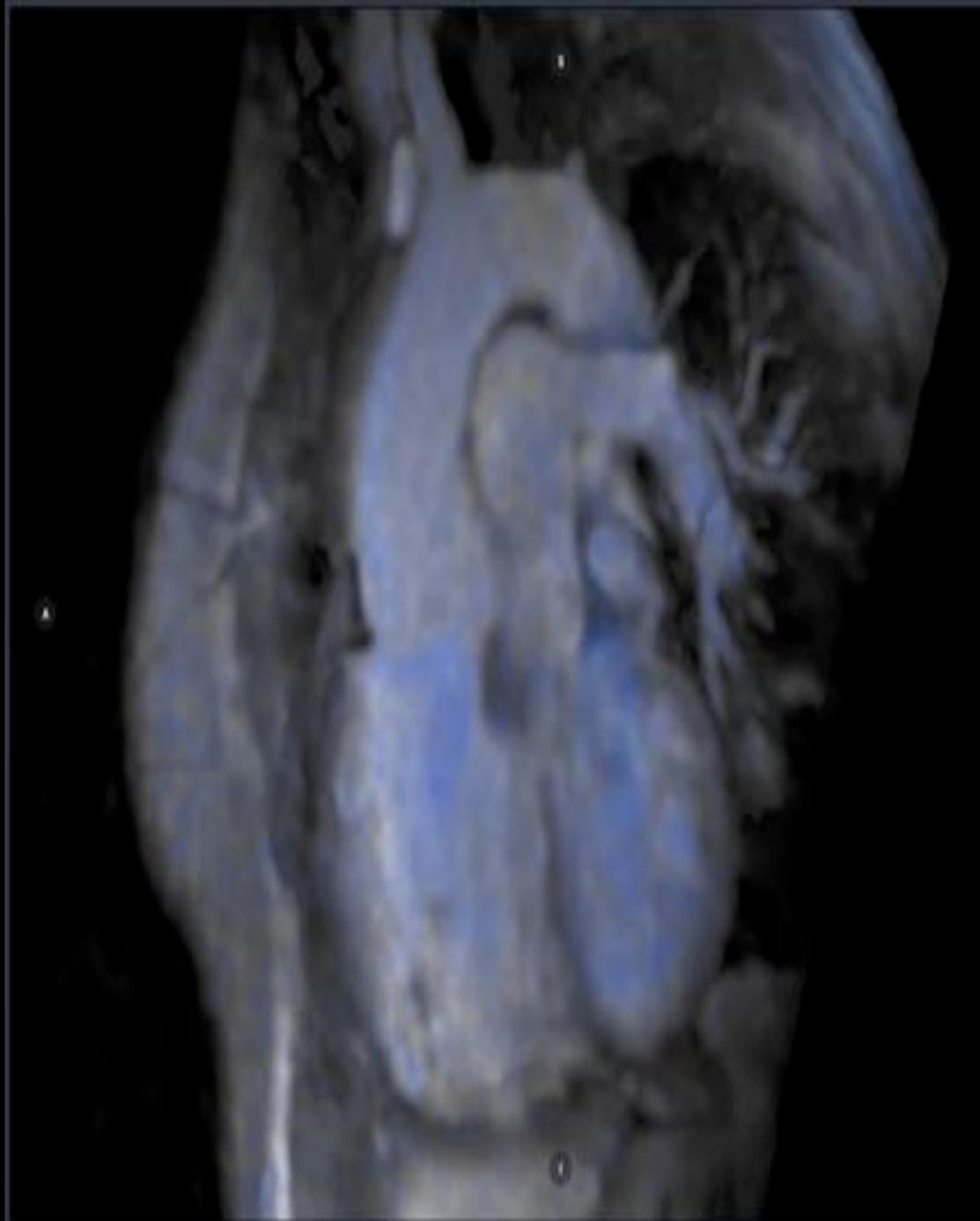
MRI senning



MRI senning

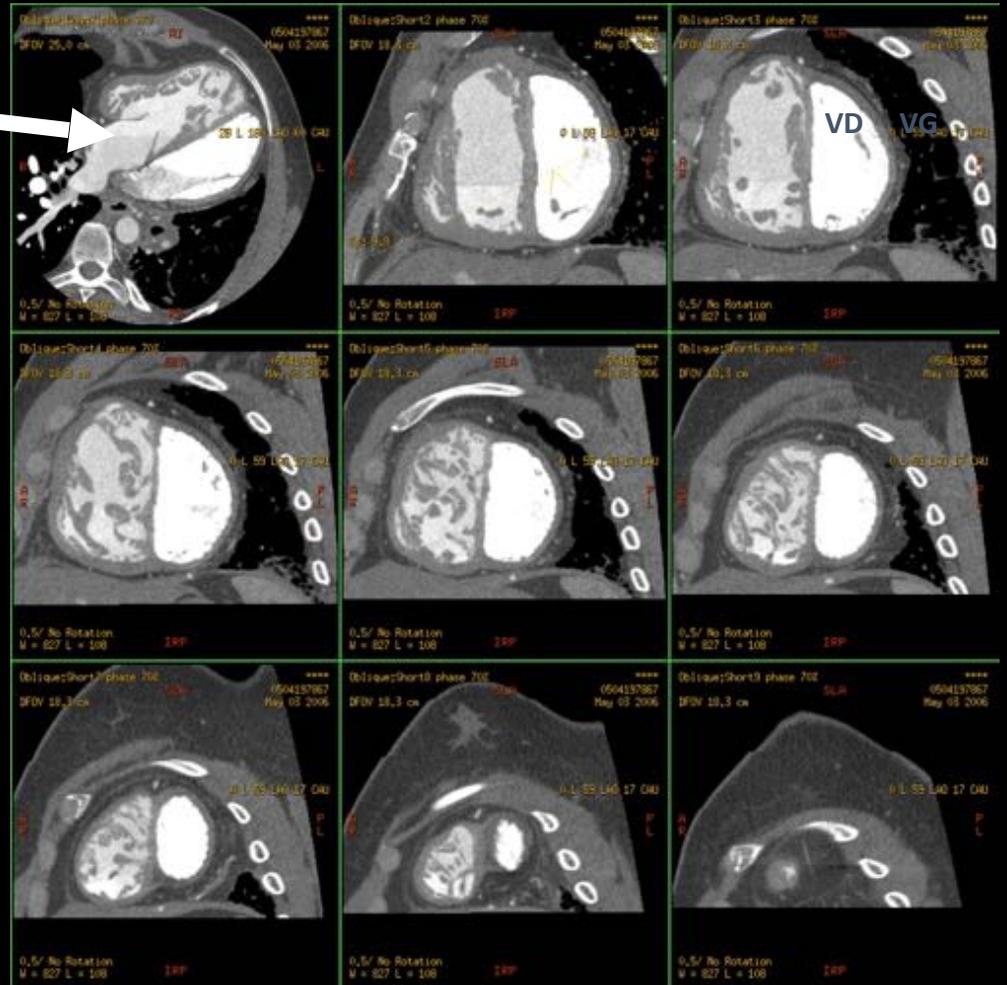


MRI senning



CT Scanner after Senning operation for TGA

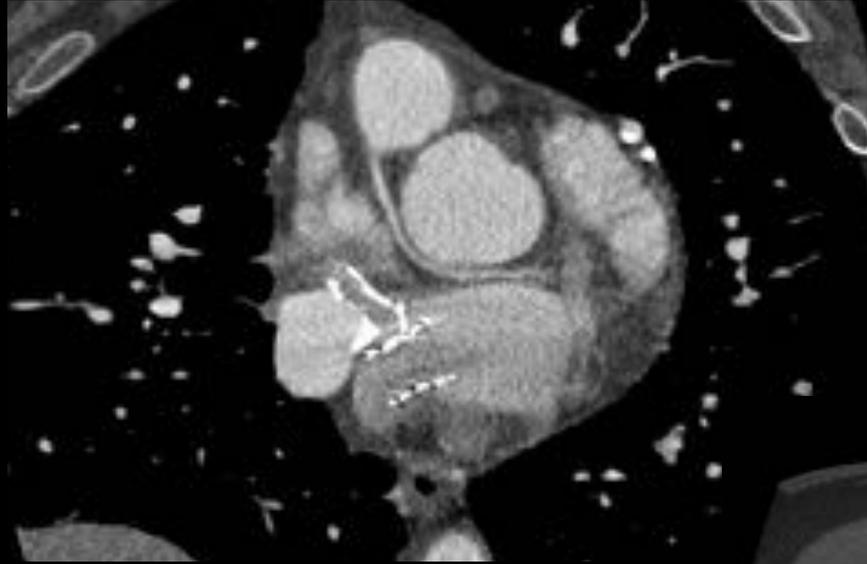
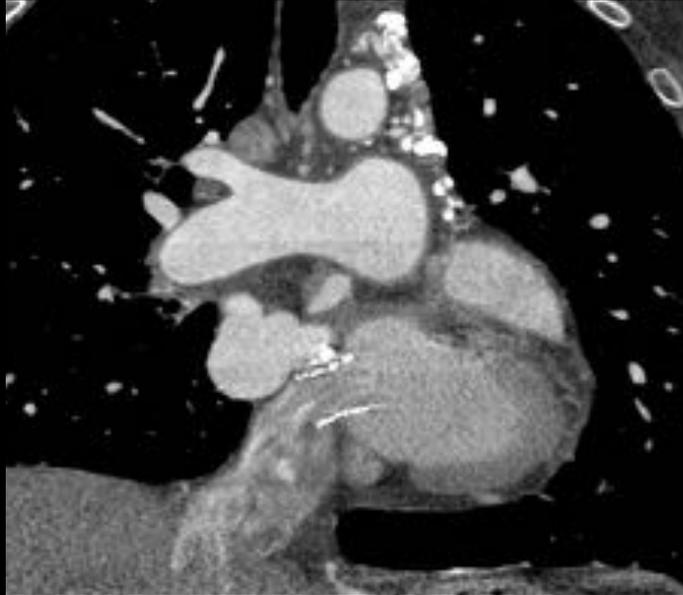
Pulmonary veins



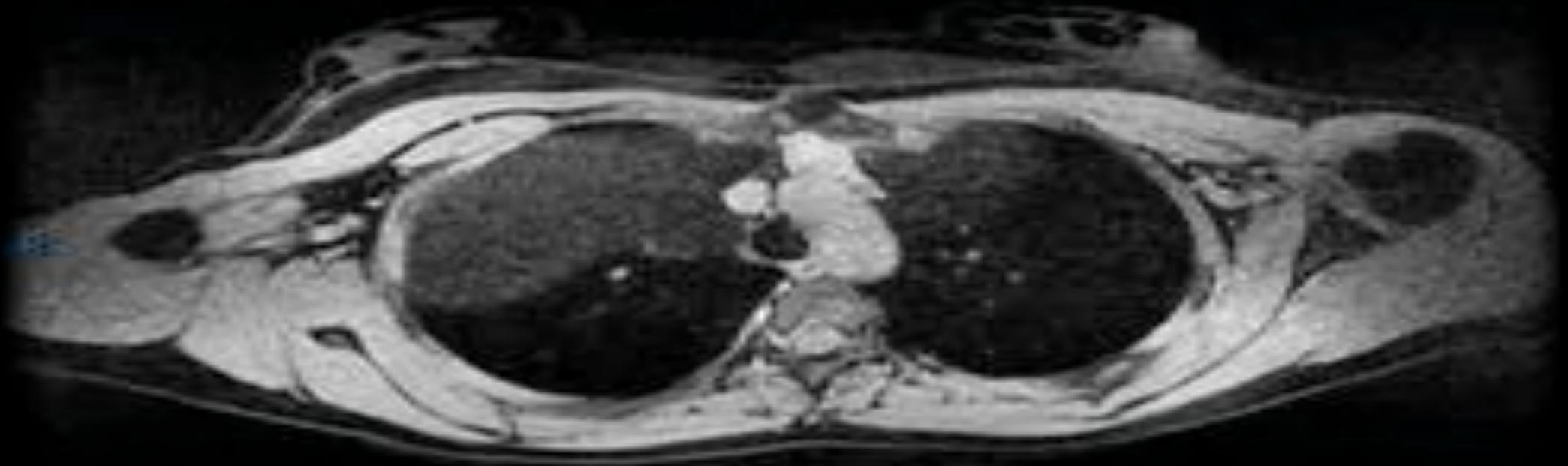
CT Scanner after Senning operation for TGA



CT Scanner after Senning operation for TGA



CT Scanner after Senning operation for TGA



RV function, systolic diastolic and strain ?

Abstract

Background

Systemic right ventricle (sRV) dysfunction in d-transposition of the great arteries following atrial switch (d-TGA) is associated with increased mortality. We aimed to characterize maladaptive sRV mechanisms in d-TGA patients, analyzing relation of echocardiographic parameters of sRV systolic function to objective measurements of exercise capacity.

Methods

Forty-seven adult patients with d-TGA and atrial switch (mean age 31.6 ± 4.2 years) underwent conventional echocardiography, bidimensional strain (2D-strain), cardiac magnetic resonance (CMR) imaging and cardiopulmonary exercise evaluation on the same day. Those with median peak oxygen uptake (VO_2) $>64.5\%$ ($n = 23$) constituted group A, those with $\text{VO}_2 \leq 64.5\%$ ($n = 24$) constituted group B and 23 healthy age and gender matched subjects constituted the control group.

Results

In group A, global longitudinal peak systolic 2D-strain (GLS) of sRV was significantly reduced compared to GLS of normal RV and LV in the healthy control group ($p < 0.01$), however peak longitudinal 2D strain was similar at basal and mid-segment of sRV free wall than normal LV. In group B, GLS was significantly reduced compared to group A ($-10.9 \pm 2.9\%$ vs $-13.1 \pm 2.3\%$, $p < 0.05$), mostly due to significant decrease of interventricular septum longitudinal strain. Other echocardiographic systolic parameters were not significantly different between groups A and B. Only sRV GLS showed significant correlation with functional capacity as measured by VO_2 ($r = 0.42$, $p < 0.01$), while CMR RVEF did not.

Conclusion

GLS of sRV predicts functional capacity and may be more sensitive than CMR RVEF in detecting early myocardial damage of sRV in patients with d-TGA and atrial switch.

Senning or Mustard repair for transposition of the great arteries

Standard transvenous pacing with an atrial lead in the systemic venous atrium and a ventricular lead in the morphologically left ventricle (LV) (subpulmonary chambers) is routinely accomplished. Pacing the systemic venous atrial appendage may cause phrenic nerve stimulation. The atrial lead should be placed at the roof of the systemic venous atrium accordingly. Lateral pacing of the morphologically LV may stimulate the diaphragm and induce ventricular dyssynchrony—the pacing lead should preferably be placed medially along the septum. Screw-in leads are preferred for use in the anatomically LV. For cardiac resynchronization therapy (CRT), epicardial RVP combined with transvenous left ventricular pacing is usually used. Transvenous CRT is technically feasible using a baffle leak or baffle puncture at the expense of an increased risk of systemic thromboembolism despite proper anticoagulation.²⁶⁵

American guidelines 2018

Recommendations for d-TGA With Atrial Switch		
Referenced studies that support recommendations are summarized in Online Data Supplement 45.		
COR	LOE	Recommendations
Diagnostic		
I	C-EO	1. Ambulatory monitoring for bradycardia or sinus node dysfunction is recommended for adults with d-TGA with atrial switch, especially if treated with beta blockers or other rate-slowing agents.
I	C-EO	2. Adults with d-TGA with atrial switch repair should undergo annual imaging with either echocardiography or CMR to evaluate for common long-term complications of the atrial switch.
IIa	C-LD	3. Assessment for a communication through the interatrial baffle or venous stenosis is reasonable for adults with d-TGA with atrial switch, particularly if transvenous pacemaker/ICD implantation is considered or leads are already present (S4.4.1.1-1).
Therapeutic		
I	B-NR	4. GDMT with appropriate attention to the need for anticoagulation is recommended to promptly restore sinus rhythm for adults with d-TGA with atrial switch repair presenting with atrial arrhythmia (S4.4.1.1-2).