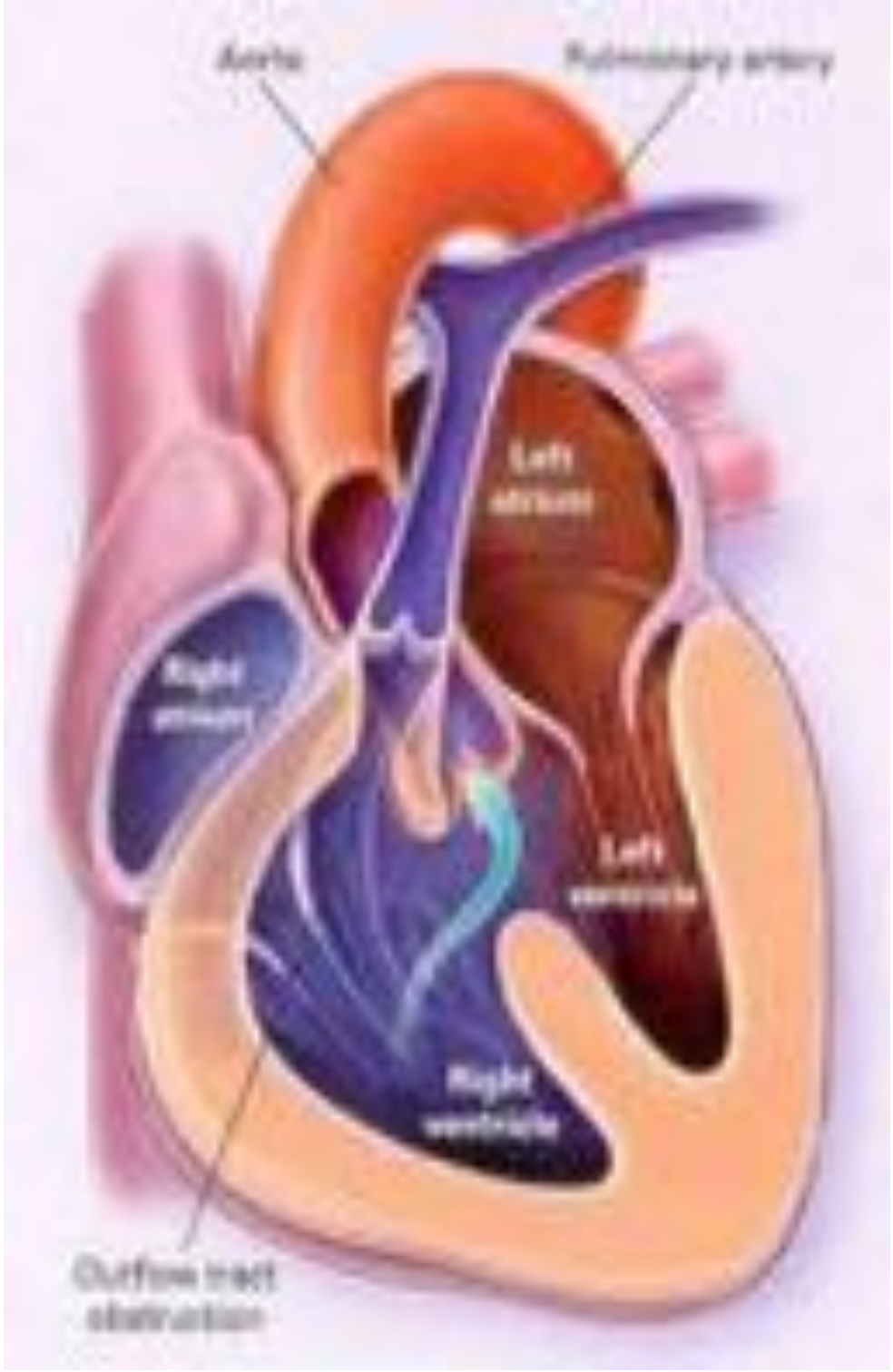
Tétralogie de Fallot à l ’age adulte quels sont les problèmes?

L Iserin

Necker, HEGP

fallot

- **Étienne-Louis Arthur Fallot** (1850-1911) was a French physician[1] born in Sète.
- Etienne Fallot attended medical school in Montpellier in 1867.
- While in residence in Marseille he wrote a thesis on pneumothorax. In 1888 he was made Professor of Hygiene and Legal Medicine in Marseille.
- In 1888 Etienne Fallot accurately described in detail the four anatomical characteristics of tetralogy of Fallot, a congenital heart defect.[2][3]

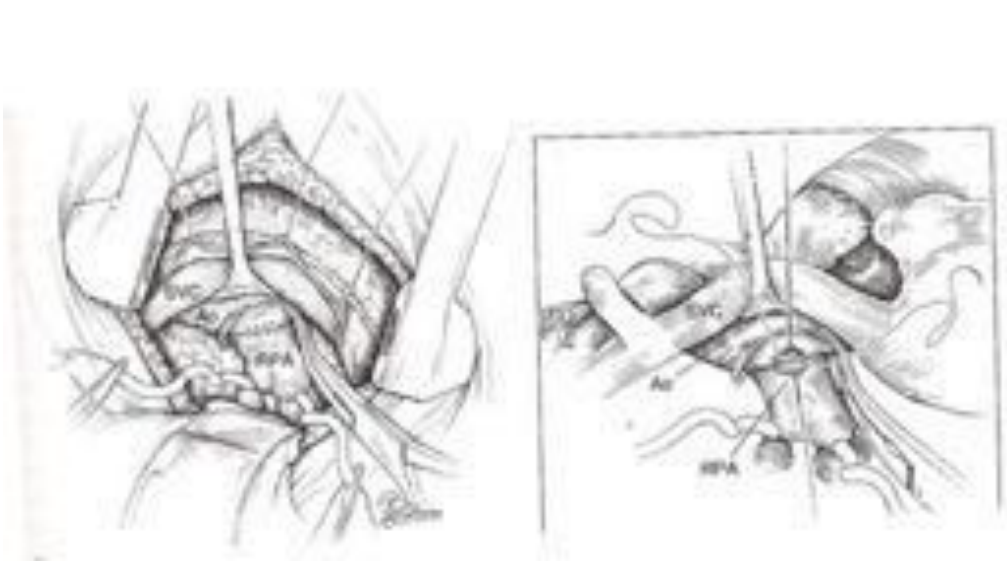


HEGP
Hôpital Européen Georges Pompidou

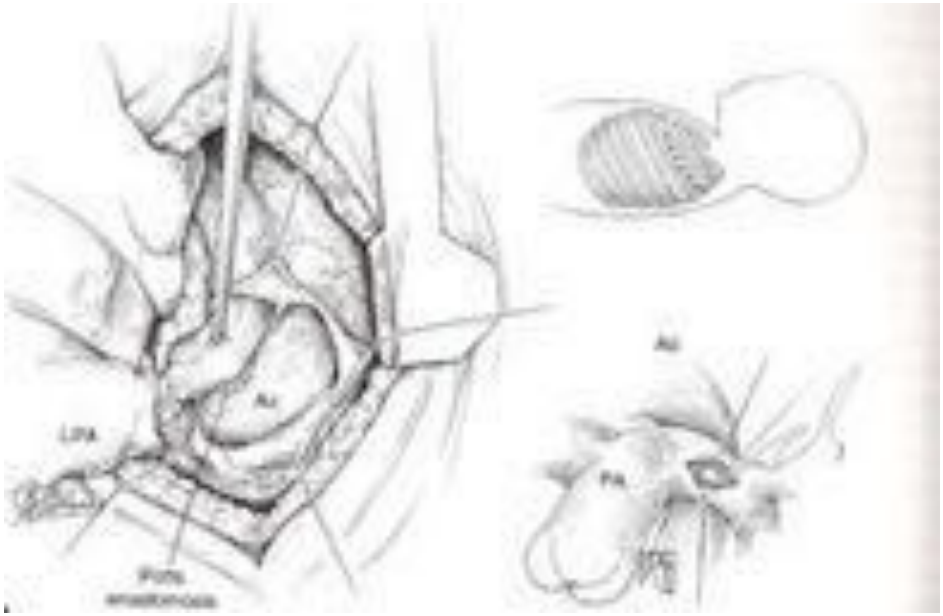
Tétralogie de Fallot

réparation chirurgicale en cure complète effectuée depuis plus de 40 ans

Anastomoses systémico pulmonaires

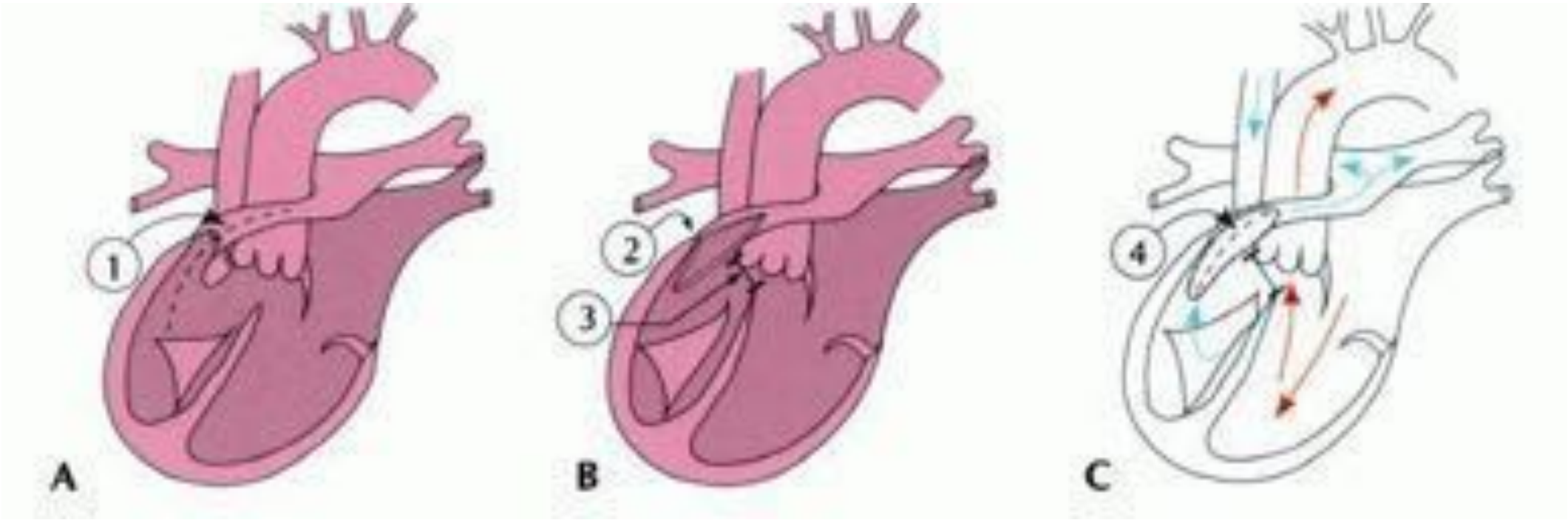


Waterston

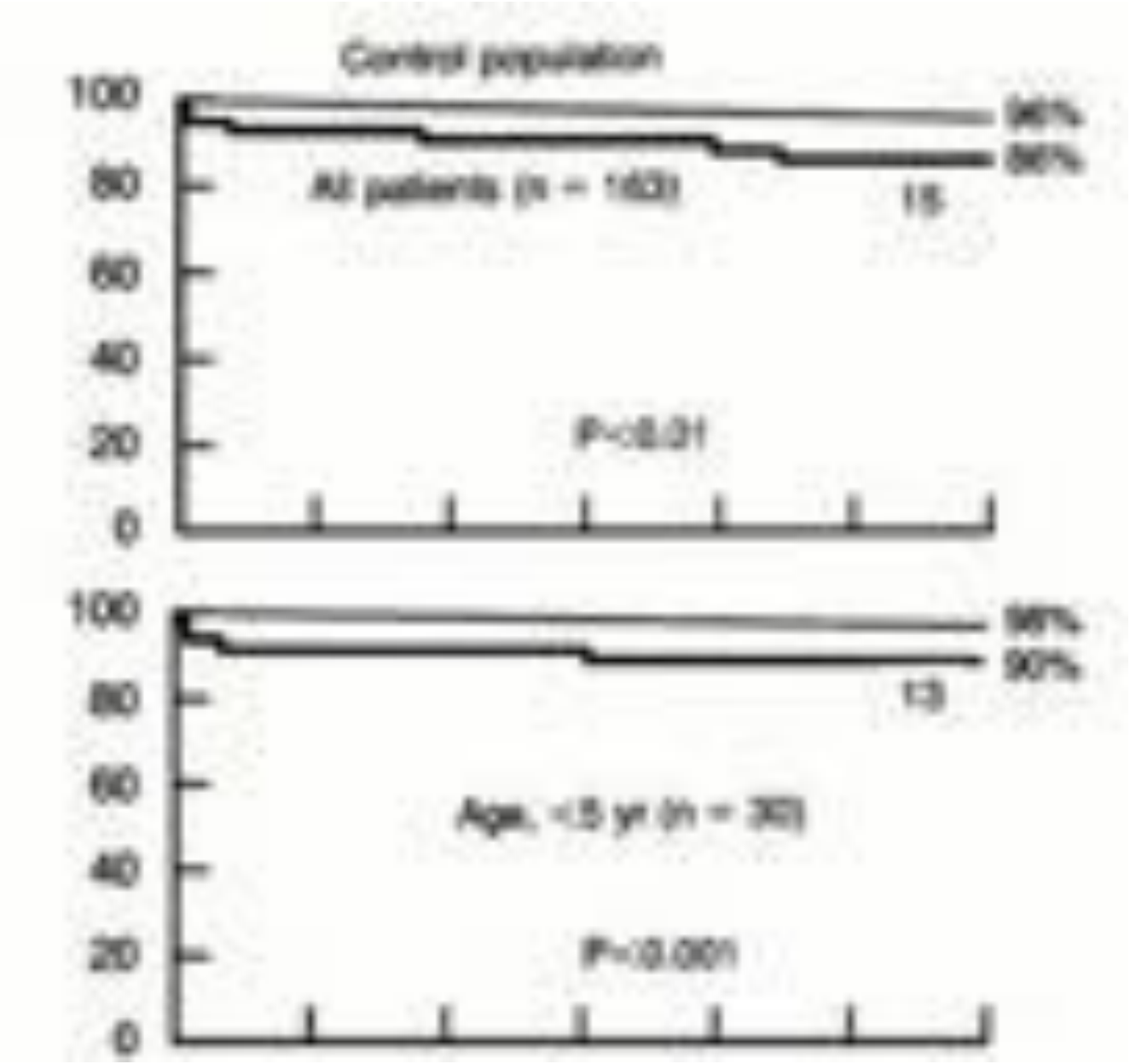


Potts

Tétralogie de Fallot réparation complète

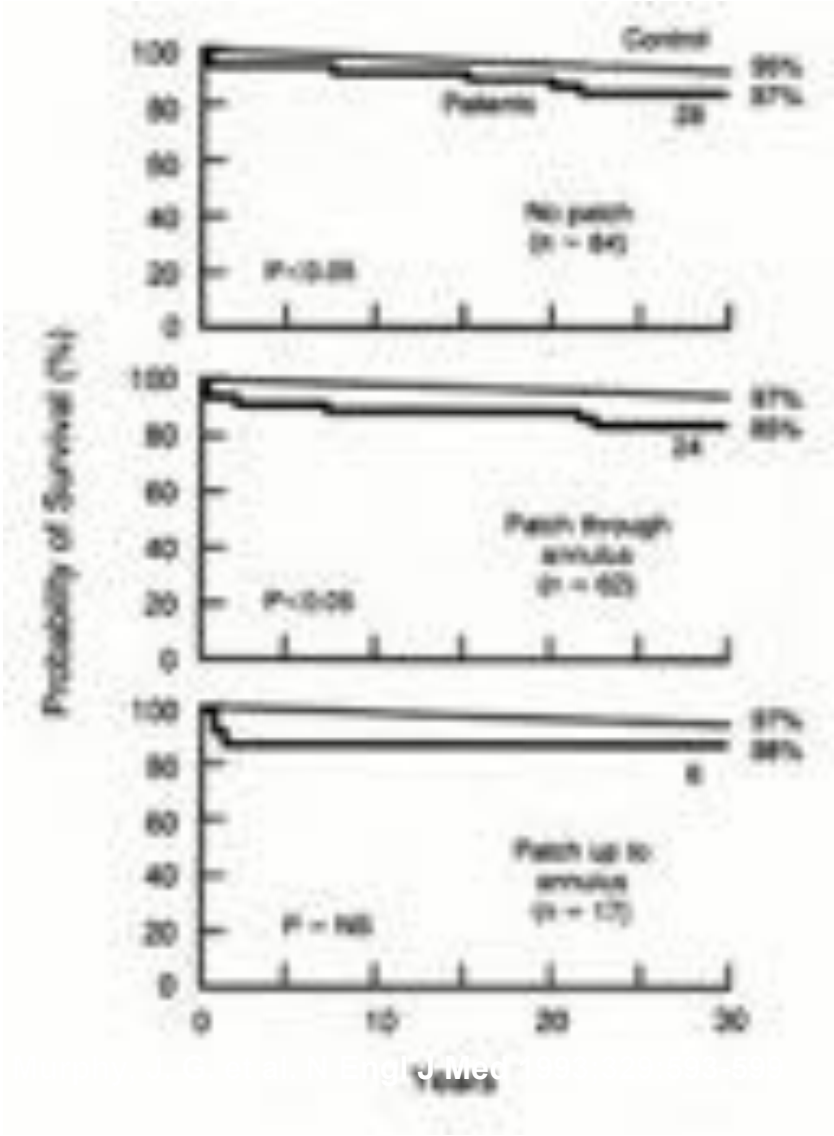


Survie à LongTerme des Patients opérés de Tetralogie ode Fallot



Murphy JG,
N Engl J Med. 1993,
26.

Survie à LongTerme des Patients opérés de Tetralogie de Fallot avec ou sans Patch transannulaire



Tétralogie de Fallot : excellents résultats : mais parfois mort subite

- Trouble du rythme ventriculaires sur VD fragilisé
- avec surcharge diastolique (fuite pulmonaire)
- et cicatrice d ’infundibulotomie

Problèmes posés

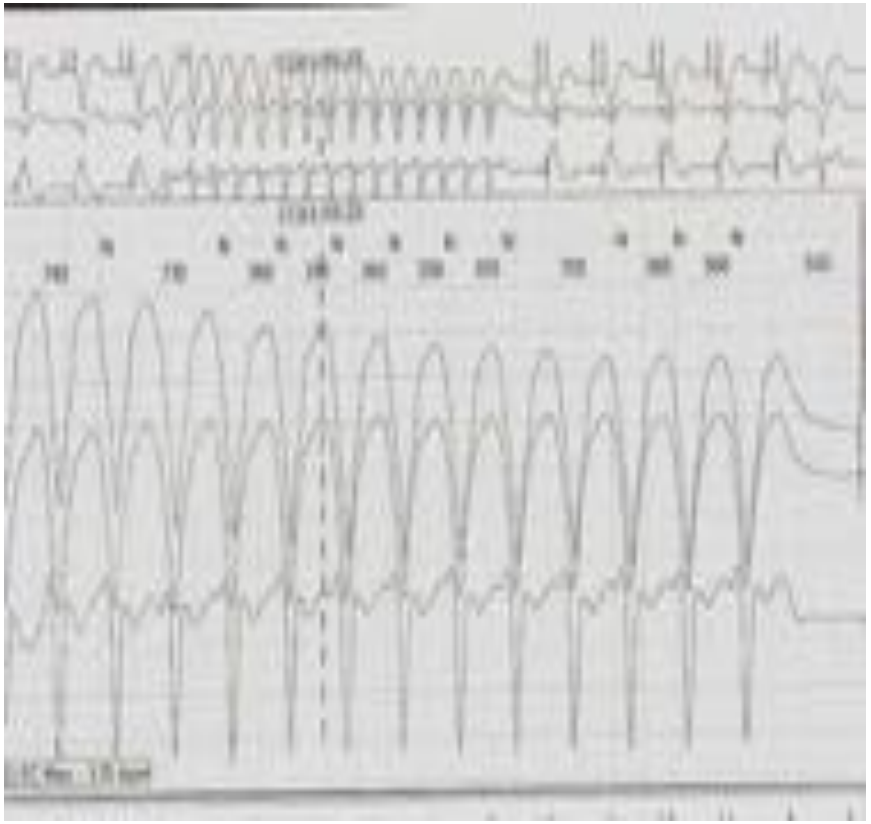
- Troubles du rythme / MS
- déterioration de la fonction VD par l ’IP
- le plus souvent asymptomatiques très longtemps
- détecter, évaluer les facteurs de risque
- traiter
- prévenir des événements rares

Troubles du rythme : TV et mort subite

TV soutenue

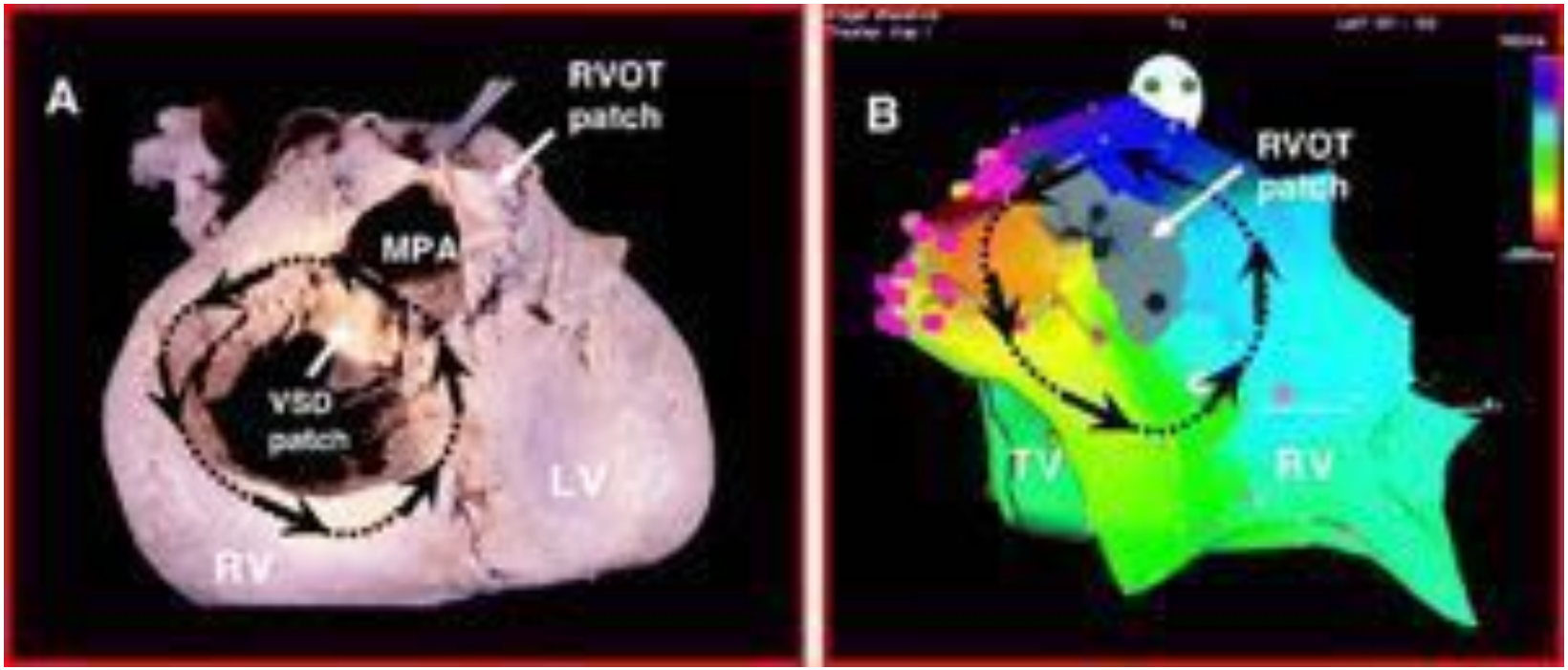
4 -7% des patients

Origine infundibulaire, BBG



Incidence de mort subite dans les Fallot

Study	Findings	Incidence per Decade, %
Murphy et al[6]	6% of 163 cases FU 30 years	2.0
Nollert et al[58]	3% of 490 cases FU 25 years	1.2
Silka et al[59]	2 Deaths per 1000 patient-years	2.0
Norgaard et al[60]	5.6% of 125 cases FU 25 years	2.2
Gatzoulis et al[55]	6% of 793 cases FU 21 years	3.0



Conduction channel between

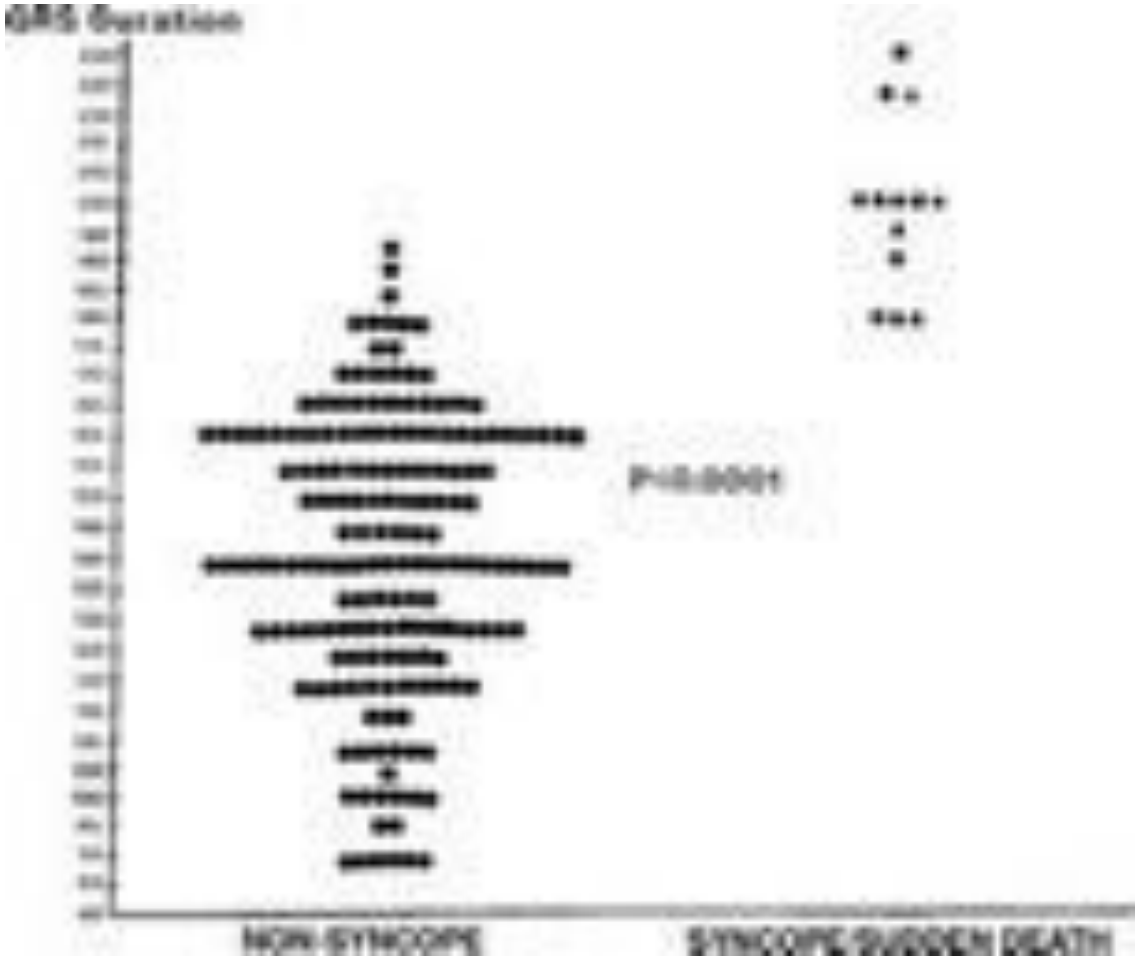
- rightward edge of the outflow patch scar (gray area)
- superior rim of the tricuspid valve.

Radiofrequency applications at this site (pink dots) closed off the channel and eliminated VT circuit

Facteurs prédictifs

Facteurs techniques	Taille de la cicatrice Patch infundibulaire Ventriculotomie
Facteurs temporels	**Age opératoire tardif** (fibrose myocardique) Suivi prolongé **Durée de CEC élevée** **Ère opératoire précoce**
Anomalies résiduelles Altérations Hémodynamiques	**Pression VD élevée** **Dysfonction V** **IP**
Anomalies ECG et électrophysiologiques	QRS QT, dispersion de la repolarisation BLOCS

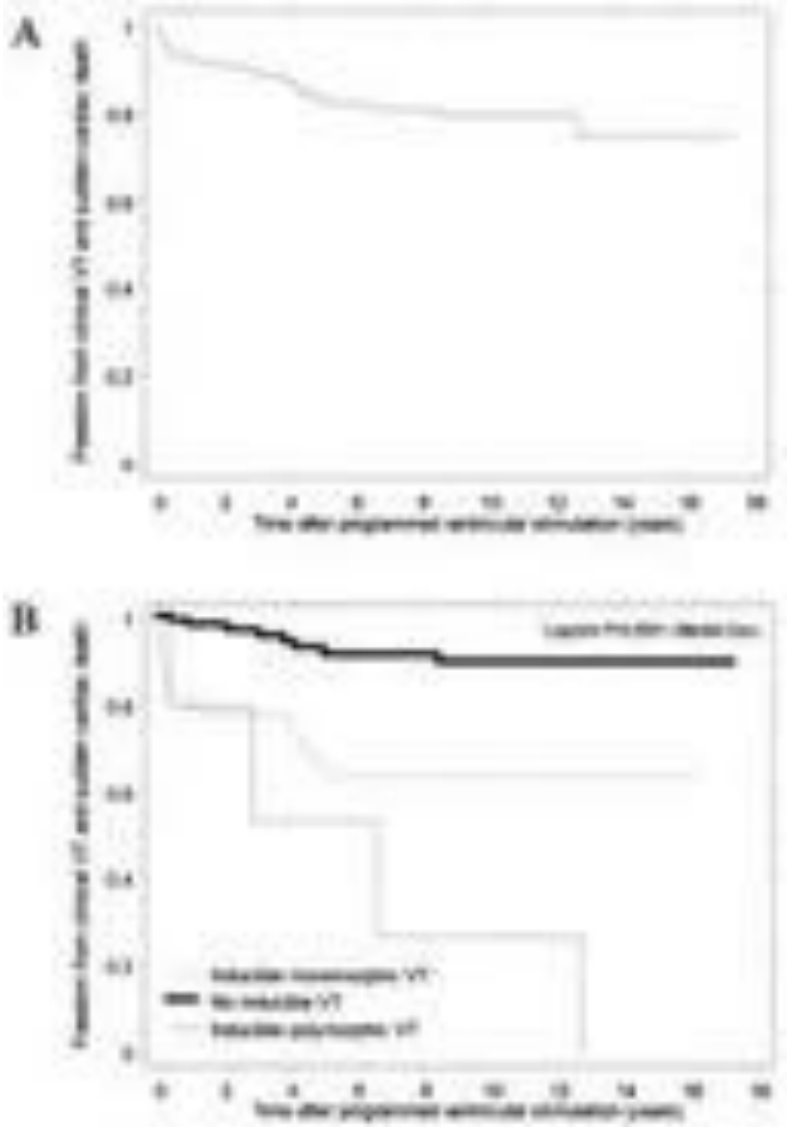
Interaction Mecanico électrique chez les patients opérés de Tetralogie de Fallot. Durée de QRS



Circulation. 1995.. Gatzoulis MA

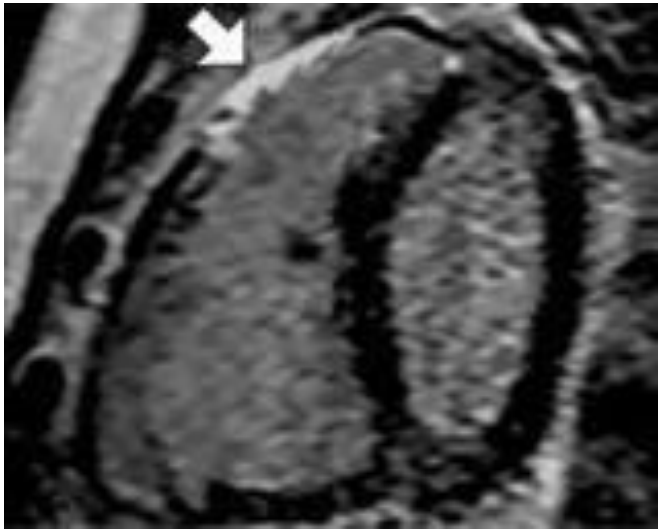
Valeur pronostique de la stimulation programmée

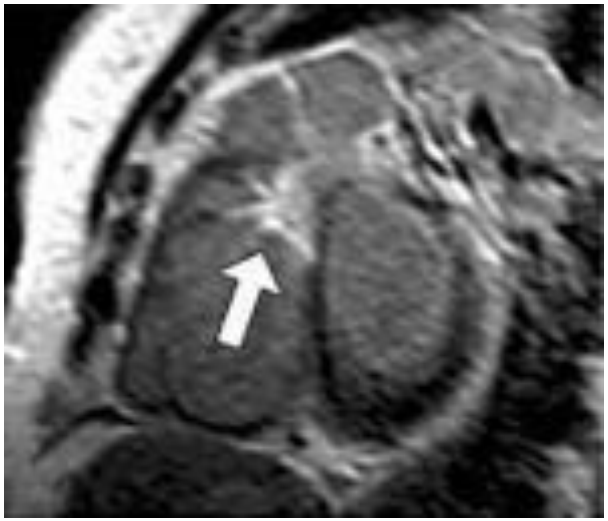
Kaplan-Meier curve of survival free from clinical VT and SCD in entire cohort

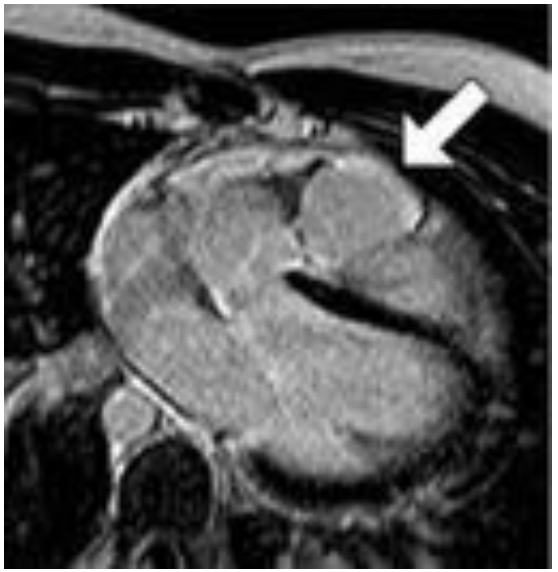


Khairy, P. et al. Circulation 2004;109:1994-2000

IRM : rehaussement tardif : fibrose





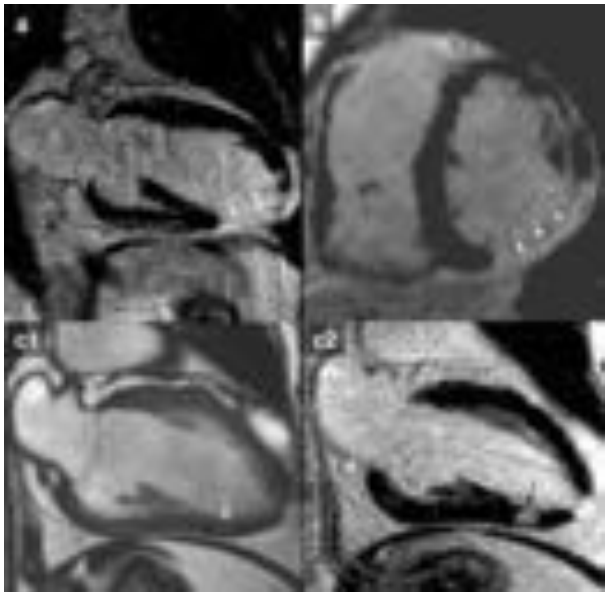


Oosterhof T, Mulder BJ, Vliegen HW, de Roos A.

Radiology. 2005

TABLE 3. Relationship of Documented Clinical Arrhythmia After Repaired TOF to Symptoms, Exercise Capacity, and RV LGE Score

	No Arrhythmia (n=76)	Arrhythmia* (n=16)	P
Age, y	31.2±10.6	36.9±12.3	0.043
New York Heart Association class	12/76	8/16	0.006
Redo surgery, n (%)	8/76 (11)	8/16 (50)	0.004
Peak heart rate, bpm	171 (162–186)	153 (134–171)	0.07
Exercise duration,	781±22	622±194	0.002
Maximum VO_2, mL·min^{-1}·kg^{-1}	27.1±7.6	23.4±4.4	0.079
QRS duration, ms	158 (148–167)	155 (143–164)	NS
QRS dispersion, ms	41±11	40±5	NS
QT dispersion, ms	85±30	76±32	NS
JT dispersion, ms	80±30	75±28	NS
Anterior RV LGE	19/76	9/16	0.017
RV LGE score	3.0 (3.0–5.0)	5.5 (3.8–12.0)	0.032
Non-apical vent LV LGE	7/76	5/16	0.017



Babu-Narayan SV, Kilner PJ, Li W,et al

Circulation. 2006

la fonction VG est aussi un facteur de risque

Comparaison de patients (12 MS, 125 contrôles vivants)

	MS	controles
• IP modérée ou sévère	92% vs.	51%,
• histoire de TV soutenue	42% vs	. 6%
• durée du QRS > 180 ms	56% vs.	13%.
• dysfonction VG modérée à sévère	42% vs.	9%

dysfonction VG modérée à sévère + QRS >180 ms
valeur prédictive positive 66%

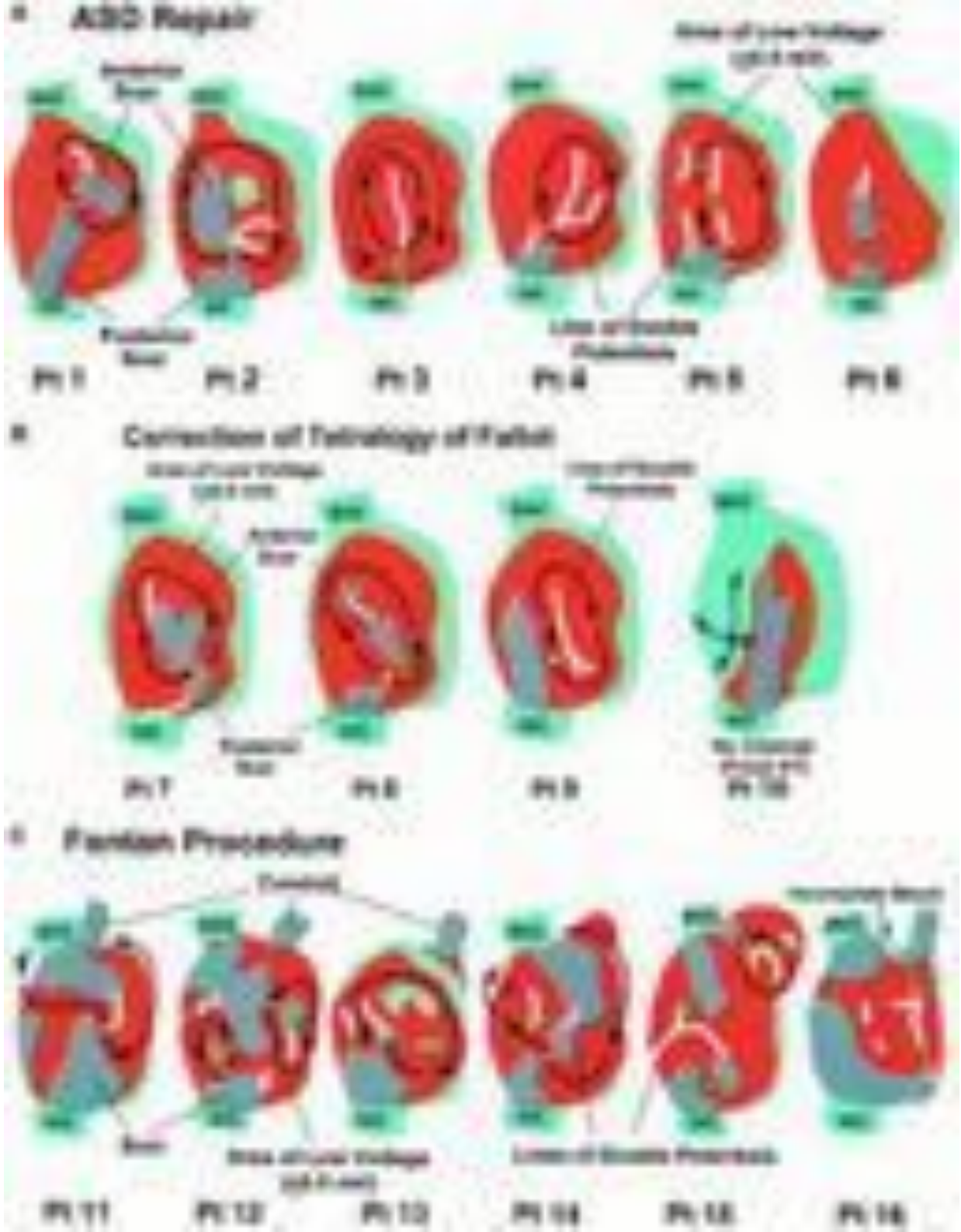
valeur prédictive négative 93%

Ghai, MD. Left Ventricular Dysfunction Is a Risk Factor for Sudden Cardiac Death in Adults Late After Repair of Tetralogy of Fallot JACC 2002

Troubles du rythme : arythmies atriales

plus fréquentes que TV !: 30% des adultes

Plus fréquents en cas de physiologie restrictive, et d ’IT

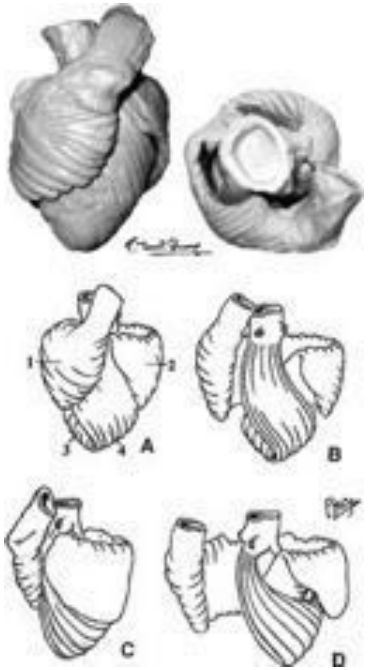

A ASD Repair
Pt 1
Pt 2
Pt 3
Pt 4
Pt 5
Pt 6
B Correction of Tetralogy of Fallot
Pt 7
Pt 8
Pt 9
Pt 10
C Fontan Procedure
Pt 11
Pt 12
Pt 13
Pt 14
Pt 15
Pt 16

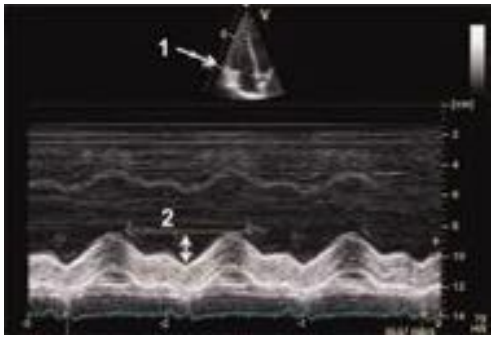
Fonction VD et IP

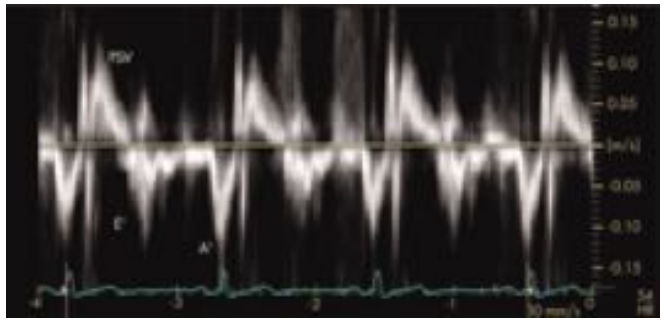
- Facteurs aggravant de l ’IP et dilatation VD
    - sténoses périphériques des AP, voire AP unique
    - résistances élevées (shunt préalable)
    - anévrisme ou akinésie infundibulaire
- Facteurs limitant l’ IP
    - physiologie restrictive du VD

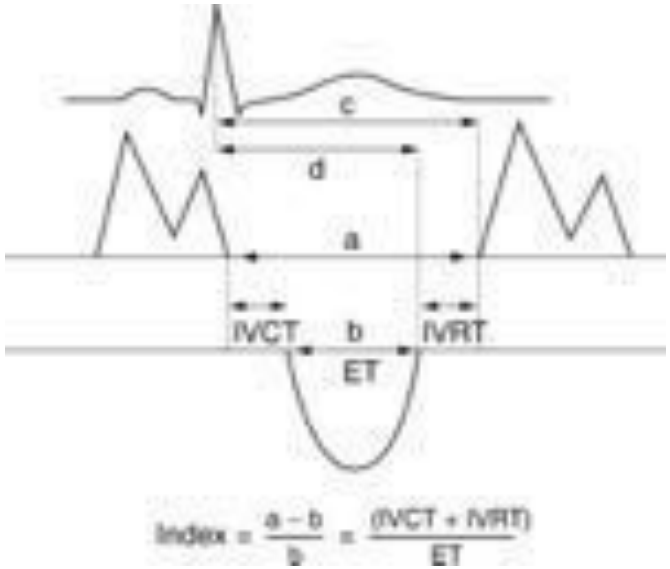
Evaluer la fonction ventriculaire droite par l'échographie est difficile

- Anatomie
  - Géometrie
  - valve Tricuspide
  - Septum
- Fonction
- lit d 'aval





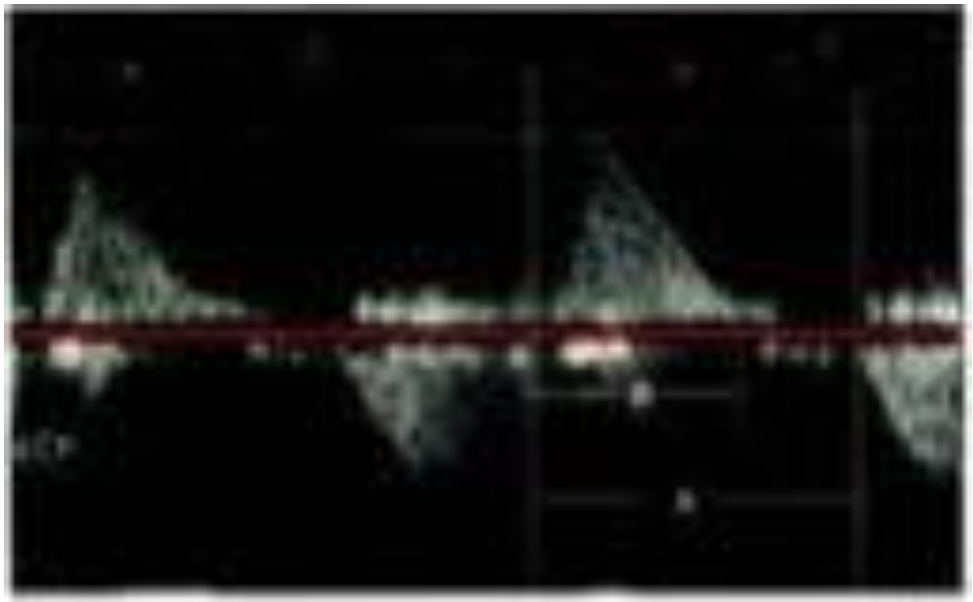


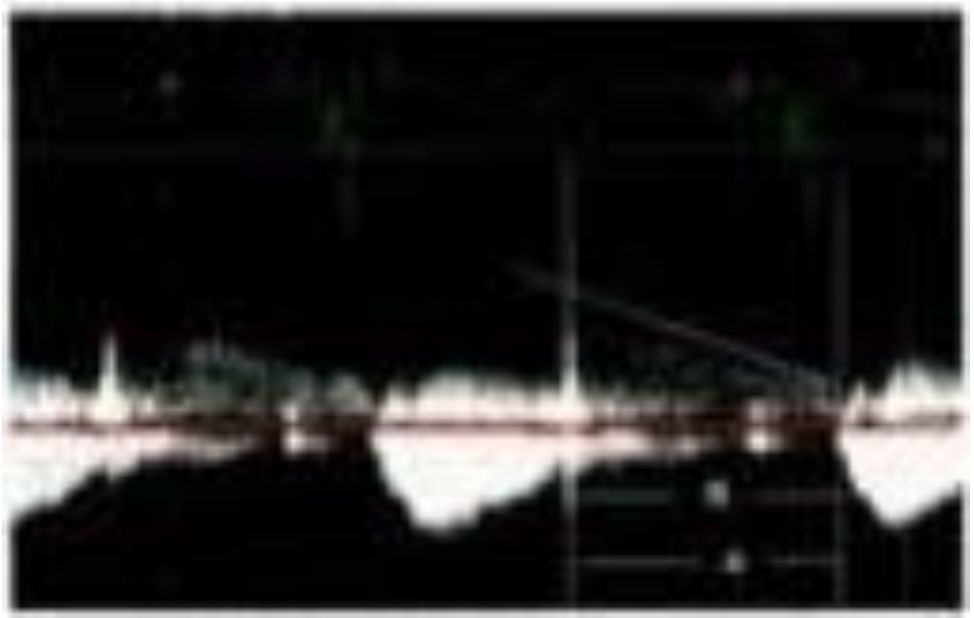


PR index:durée de l'IP /durée totale de la diastole
PRi < 0.77 relié à la FR en IRM

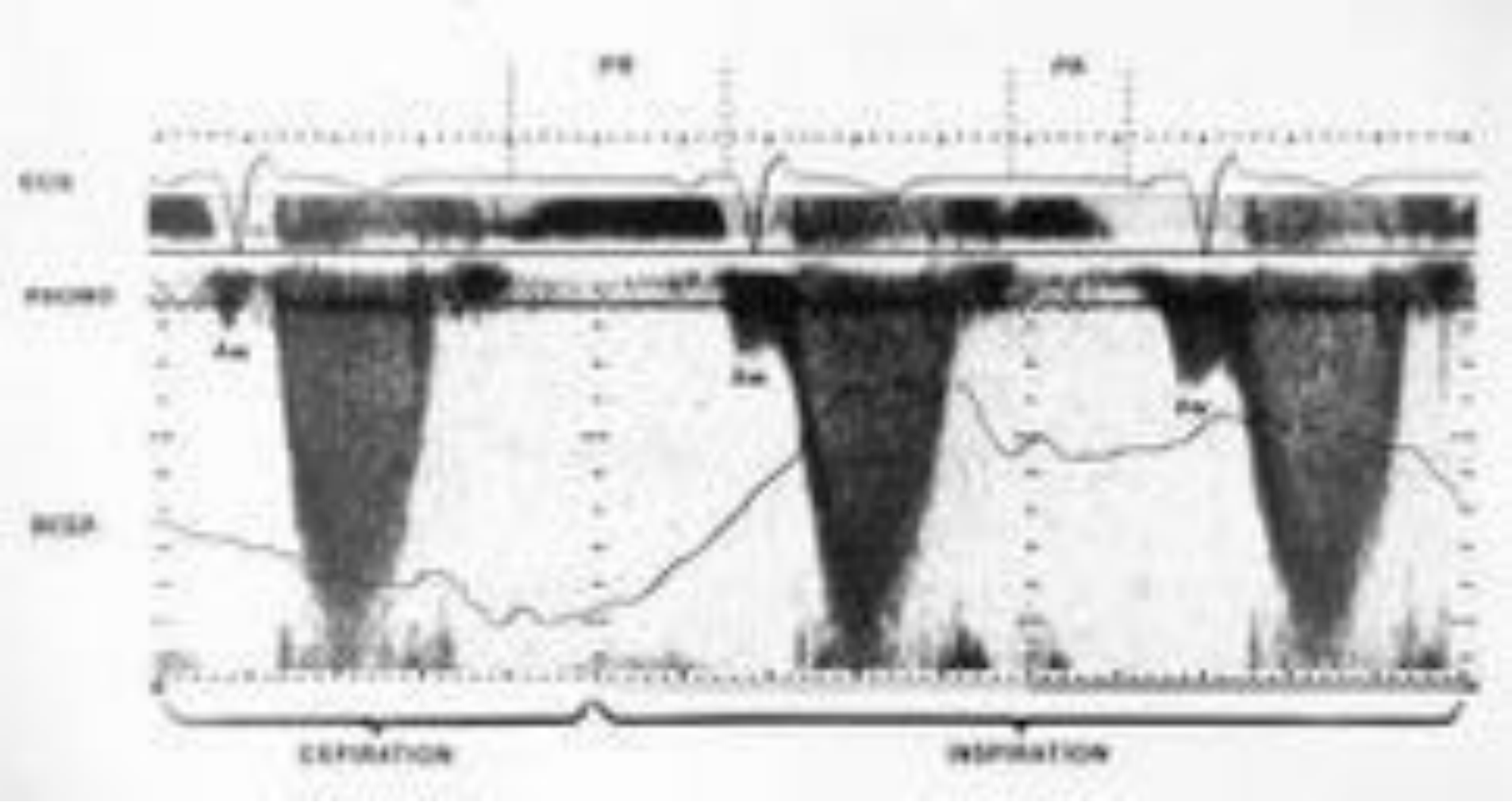
.

PHT <100ms





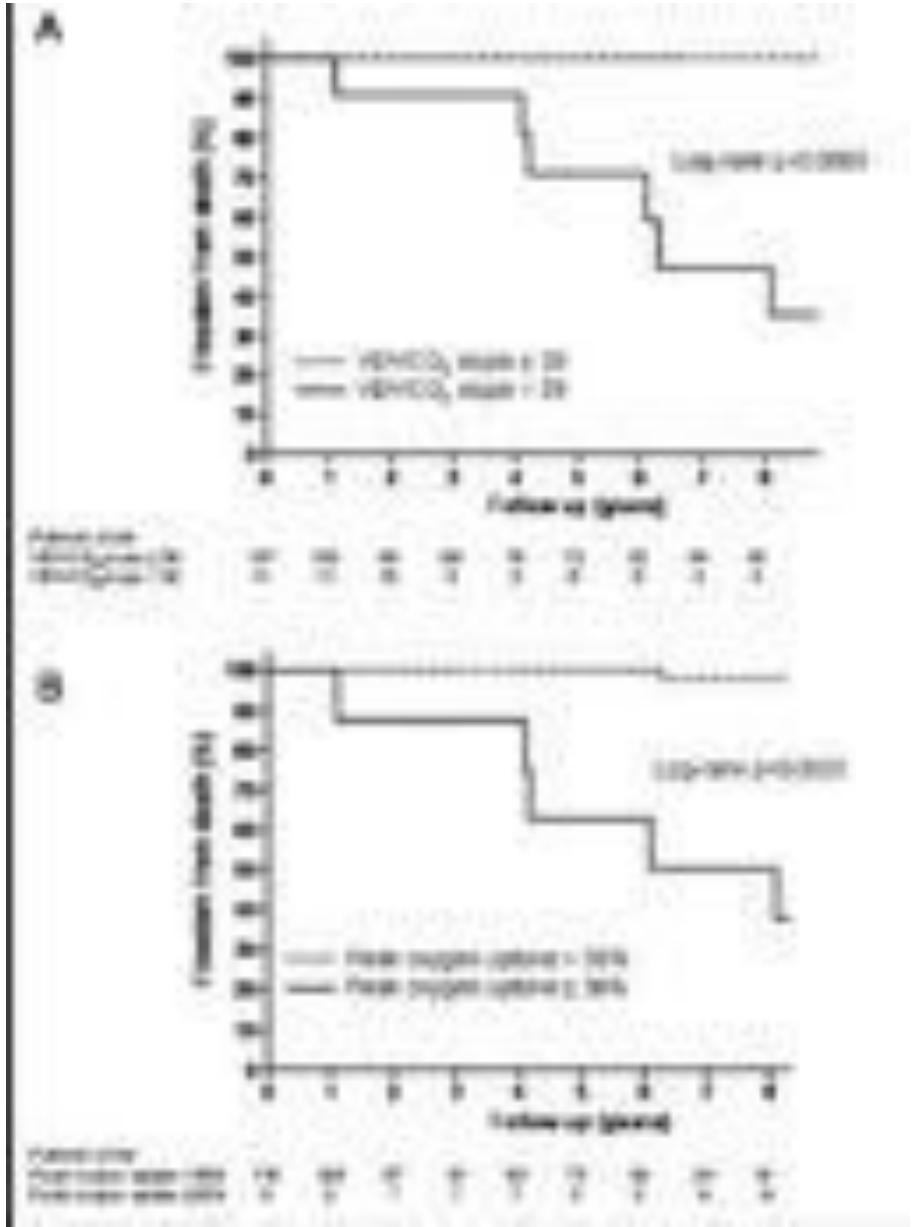
Physiologie Restrictive



Usefulness of Cardiopulmonary Exercise to Predict Long-Term Prognosis in Adults With Repaired Tetralogy of Fallot

Alessandro Giardini, MD[a,*], Salvatore Specchia, MD[b], Theresa Ann Tacy, MD[c],

Am J Cardiol 2007;99

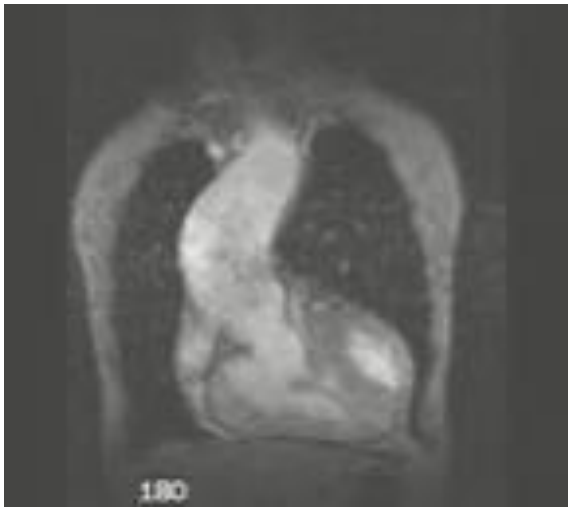


Evaluation de la fuite pulmonaire et de son retentissement

- évaluation de la largeur du QRS
- Echo dilatation VD, apparition d'une IT
- IRM
- épreuve d 'effort avec mesure de la VO2
- BNP
- Holter de peu de valeur si asymptomatique
- valeur des troubles du rythme symptomatiques

Pathologies acquises

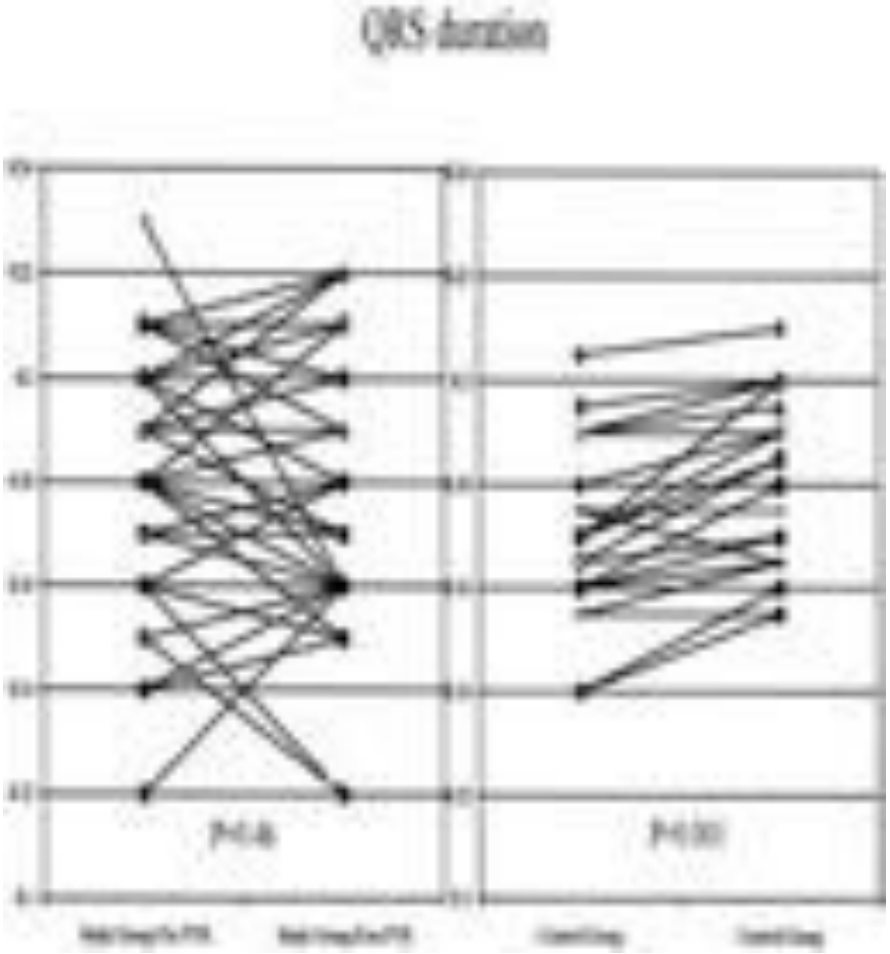
- Dilatation de l ’aorte ascendante
- IA
  - dissection : 3 cas dans la littérature



Pathologies acquises

- Collatéralité (hemoptysie)
- Pathologie coronaire
- Dysfonction gauche de mécanisme complexe
  - CEC sur myocarde épais , hypoxémique,
  - désynchronisation,
  - interaction VD VG,
  - trouble du rythme atriaux

Impact du replacement valvulaire pulmonaire sur la propension à faire des arythmies après reparation des tétralogies de Fallot.



Therrien J,Circulation. 2001

Approach to asymptomatic adult : unsettled.

Arrhythmias in Adult Patients With Congenital Heart Disease

Edward P. Walsh, MD; Frank Cecchin, MD , Circ 2007

Most clinicians : yearly evaluation with ECG, Holter or exercise testing

, echo and MRI to monitor the status of the RV.

Should nonsustained VT be detected in an asymptomatic patient,

or should RV function appear to be deteriorating,

Opinions still vary widely …….

Some clinicians would advocate EPS

Some would recommend surgery for PVR,

Some would prescribe antiarrhythmic drugs,

Some would implant a primary prevention ICD,

and some might refrain from any treatment .

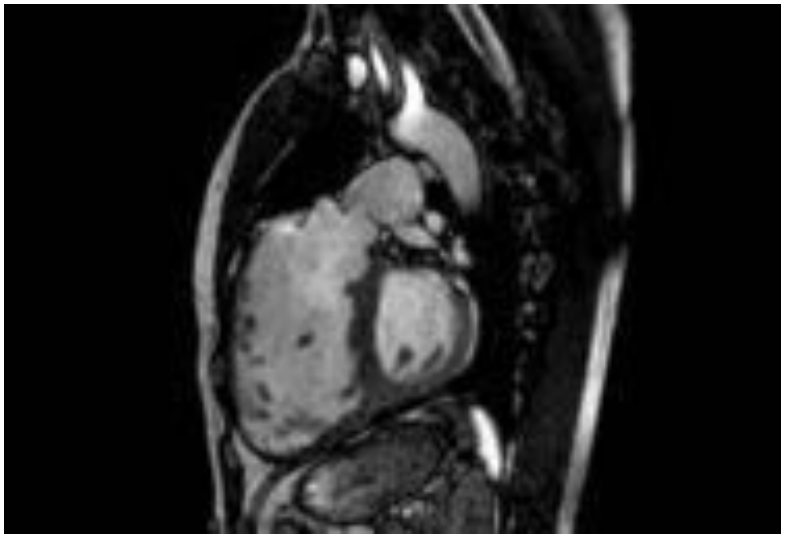
Therapy continues to be individualized for asymptomatic patients depending on institutional experience and *philosophy.*

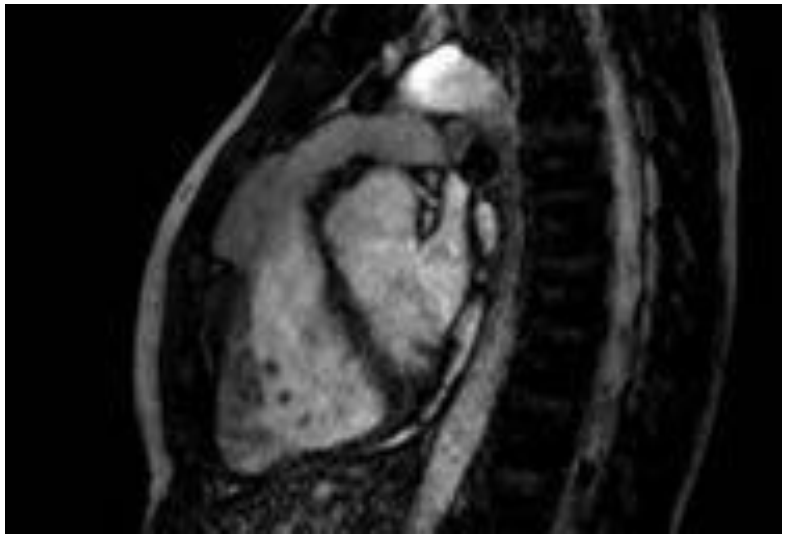
The lack of objective guidelines for VT prediction and treatment in CHD patients is *frustrating*

MRI : gold standard

- Si on réalise le RVP vers volume VD de 150 mL/m(2)
- réduction des volumes et recuperation de la FE VD*Buechel ER Eur Heart J. 2005;26,*
- **au delà de volume VD de 170ml/m2 le VD ne recupère pas une taille normale**.
- *Therrien J, Am J Cardiol. 2005, 95*

IRM : gold standard





IRM détermine les seuils de valvulation pulmonaire

- Remplacement valvulaire concernera 15% ou 50%???des patients
- Si volume télédiastolique VD >170 ml/m2
- ou volume télé systolique >85 ml/m2

les volumes VD ne se normalisent pas après chirurgie

- recommandations…… Valver quand VTDVD >150 ml/m2 ?

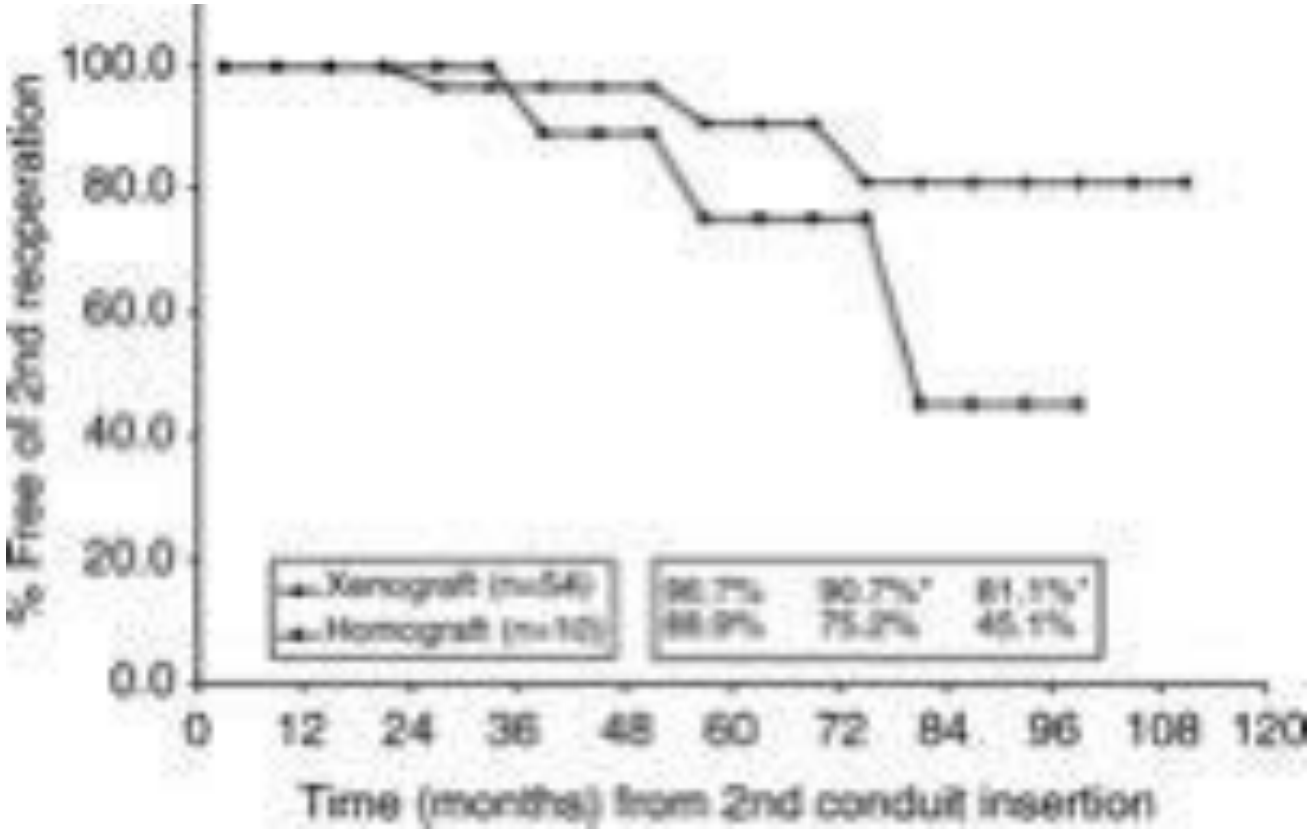
Therrien J, Am J Cardiol. 2005, 95

BNP

- BNP élevé par rapport à une population contrôle
- BNP augmente chez les patients avec une dilatation importante du VD
- correlé avec des index Doppler compliqués

Oosterhof T, Tulevski II, Vliegen HW Am J Cardiol. 2006 ;97

Brili, J Am Soc Echocardiogr. 2005;18.


% Free of 2nd reoperation
100.0
80.0
60.0
40.0
20.0
0.0
Xenograft (n=54)
Homograft (n=10)
0
12
24
36
48
60
72
84
96
108
120
Time (months) from 2nd conduit insertion

HEGP
Hôpital Européen Georges Pompidou

Long terme de RVP

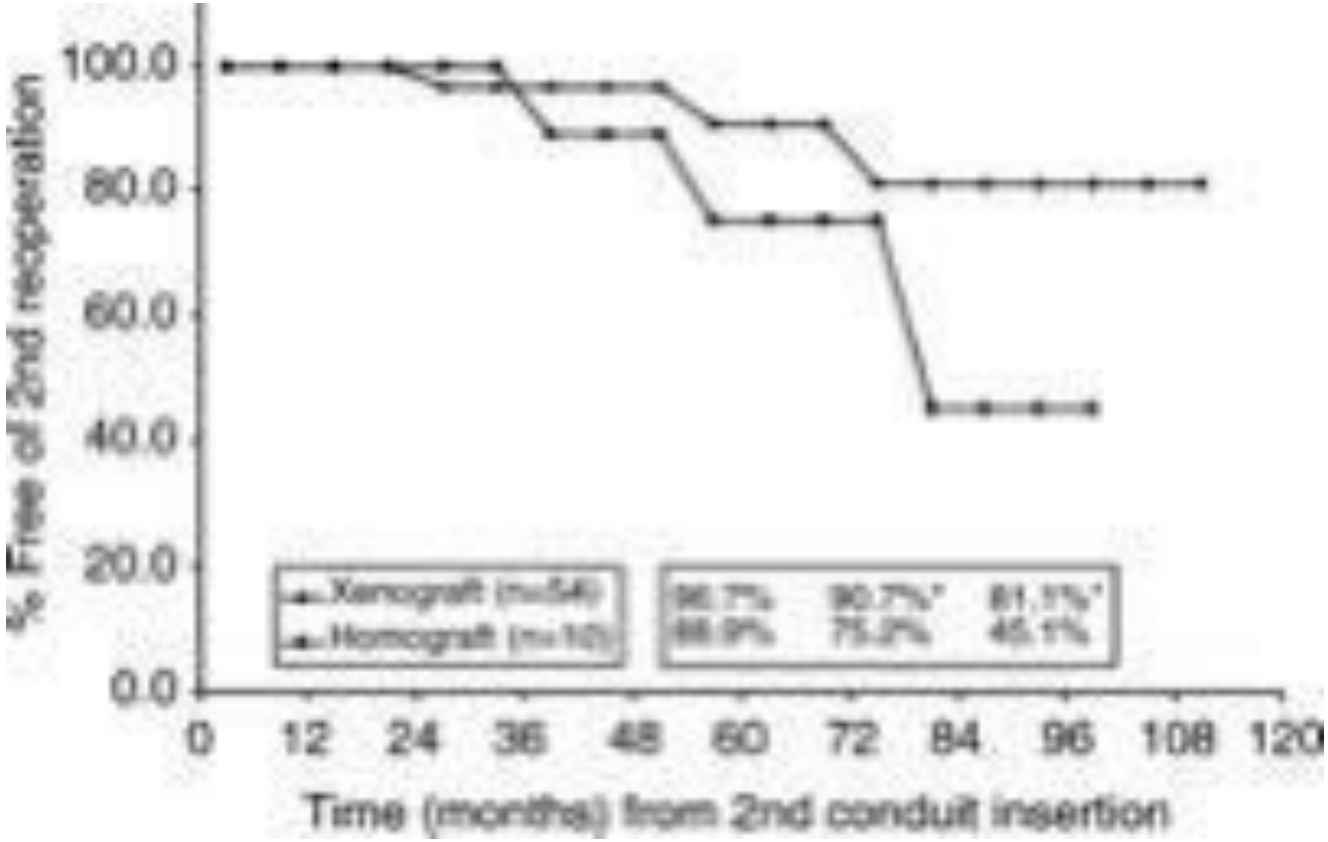
158 adultes (registre) opérés dans l ’enfance.

175 RVP entre 1986 et 2005.

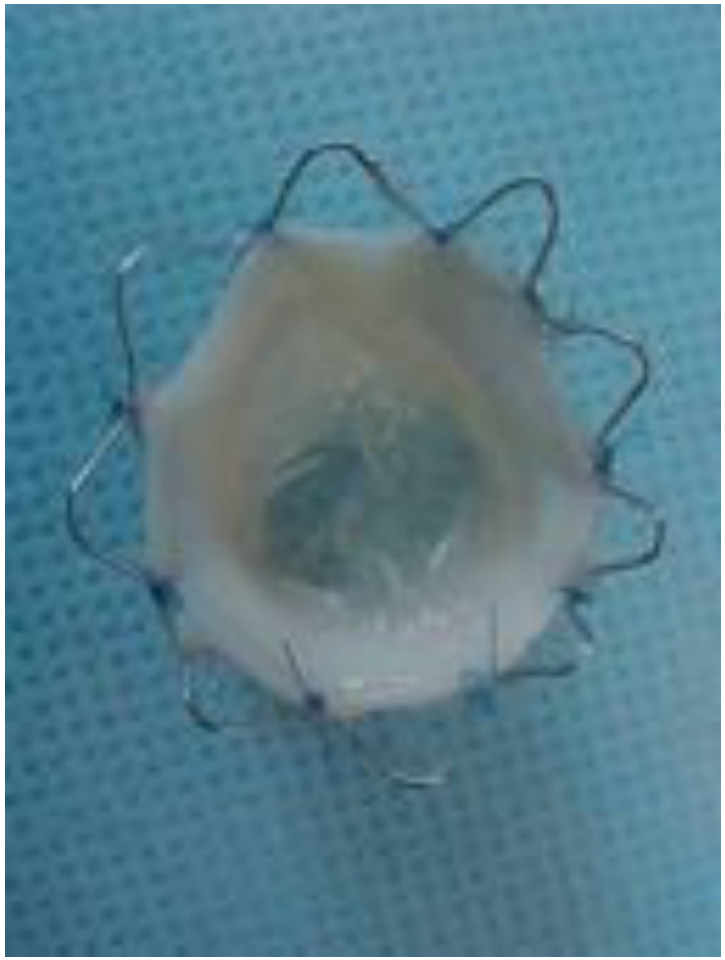
- 66% d ’absence de dysfonction de la valve à 5 ans et 47% à 10 ans.
- Survie sans évenements 78% à 10 ans 68% à 15 ans après RVP.
- IP et RP post op immédiate : FDR évenements.
- réalisation d ’une resection de patch ou d ’anévrisme → moins d ’IP post op
- Moins de SP pour les plus grandes tailles d ’ homogreffe

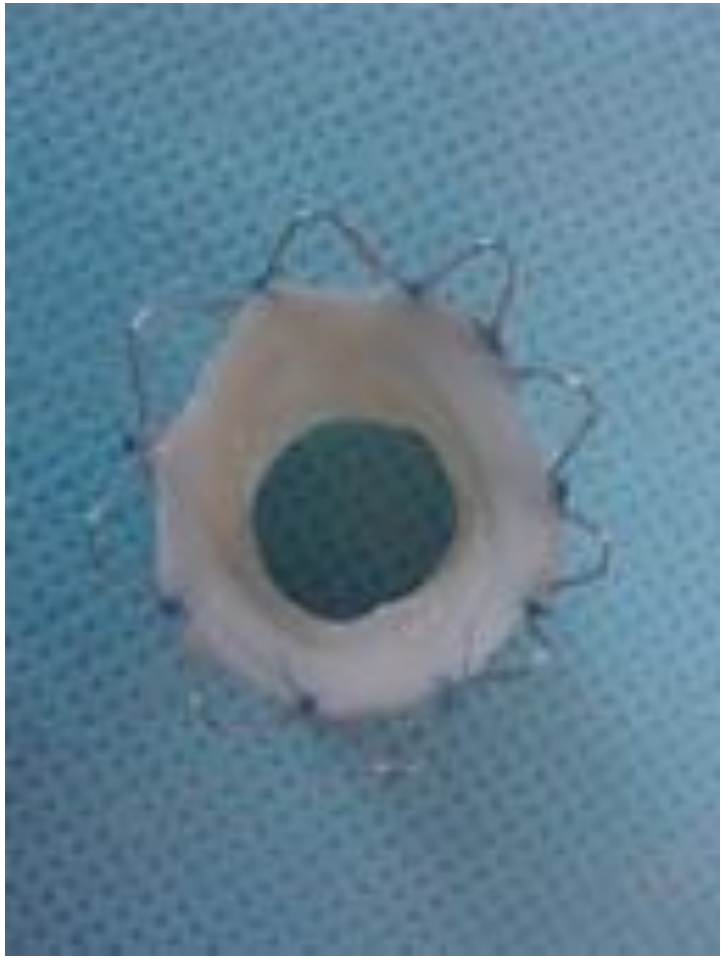
Oosterhof T,et al. Eur Heart J. 2006.

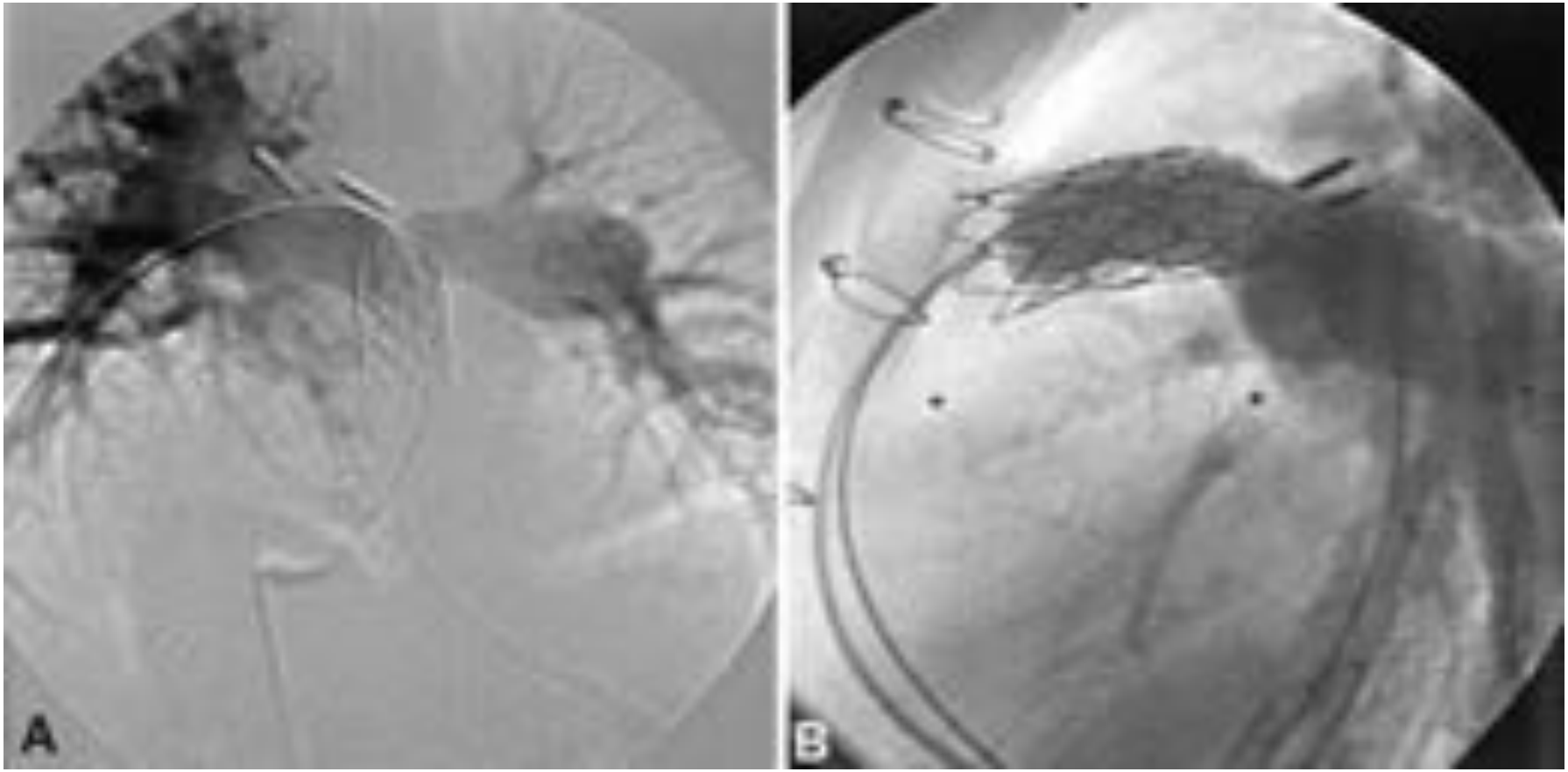
Quels substituts?



Espoir : valvulation percutanée pulmonaire






A
B

Review of SCD in ACHD
Fallot : rare event

Author	Findings	Mean F/U (years)	SCD incidence/ decade (%)
Murphy et al. [30]	SCD in 6% of 163 cases	30	2.0
Nollert et al. [34]	SCD in 3% of 490 cases	25	1.2
Silka et al. [47]	SCD in 2.6% of 445 cases	22	1.8
Norgaard et al. [35]	SCD in 5.6% of 125 cases	25	2.2
Gatzoulis et al. [12]	SCD in 6% of 793 cases	21	3.0

Silka, Pediatr Cardiol (2012)

Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

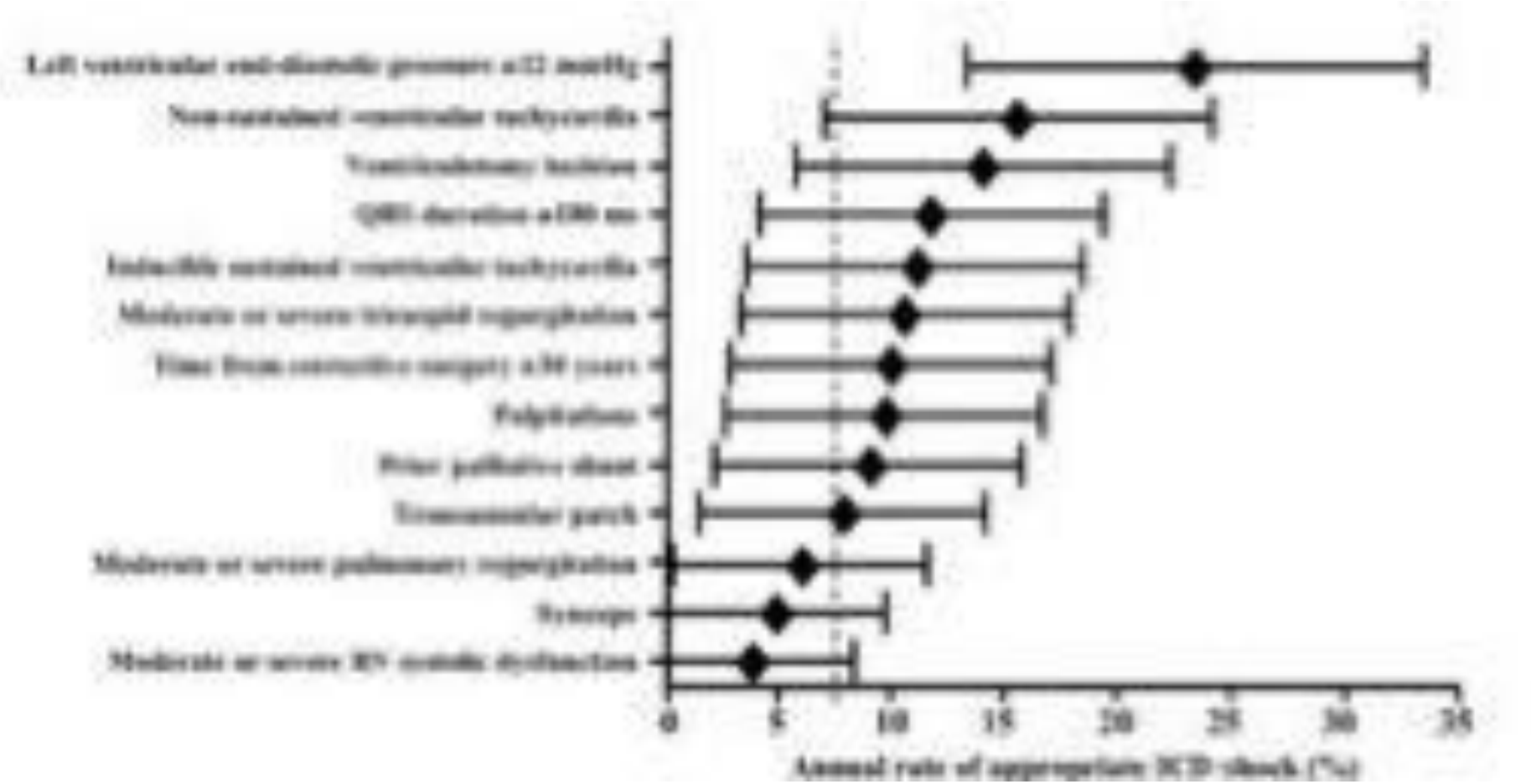
Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;

Methods and Results—We conducted a multicenter cohort study in high-risk patients with Tetralogy of Fallot to determine actuarial rates of ICD discharges, identify risk factors, and characterize ICD-related complications. A total of 121 patients (median age 33.3 years; 59.5% male) were enrolled from 11 sites and followed up for a median of 3.7 years. ICDs were implanted for primary prevention in 68 patients (56.2%) and for secondary prevention in 53 (43.8%), defined by clinical sustained ventricular tachyarrhythmia or resuscitated sudden death. Overall, 37 patients (30.6%) received at least 1 appropriate and effective ICD discharge, with a median ventricular tachyarrhythmia rate of 213 bpm. Annual actuarial rates of appropriate ICD shocks were 7.7% and 9.8% in primary and secondary prevention, respectively ($P=0.11$). A higher left ventricular end-diastolic pressure (hazard ratio 1.3 per mm Hg, $P=0.004$) and nonsustained ventricular tachycardia (hazard ratio 3.7, $P=0.023$) independently predicted appropriate ICD shocks in primary prevention. Inappropriate shocks occurred in 5.8% of patients yearly. Additionally, 36 patients (29.8%) experienced complications, of which 6 (5.0%) were acute, 25 (20.7%) were late lead-related, and 7 (5.8%) were late generator-related complications. Nine patients died during follow-up, which corresponds to an actuarial annual mortality rate of 2.2%, which did not differ between the primary and secondary prevention groups.

Conclusions—Patients with tetralogy of Fallot and ICDs for primary and secondary prevention experience high rates of appropriate and effective shocks; however, inappropriate shocks and late lead-related complications are common. (Circulation. 2008;117:363-370.)

Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;



ESC Guidelines for the management of grown-up congenital heart disease (GUCH)

Authors/Task Force Members:

Helmut Baumgartner, Chairperson (Germany)

Philipp Bonhoeffer (UK), Nataša De Groot (Netherlands), Fokko de Haan (Germany), John Erik Deanfield (UK), Nazzarena Gatzoulis (Italy), Michael Gatzoulis (UK), Christa Gohlke-Baerwolf (Germany), Harald Kaemmerer (Germany), Philip Kilner (UK), Folkert Meijboom (Netherlands), Barbara JM Mulder (Netherlands), Erwin Oechslin (Canada), Jose M Oliver (Spain), Alain Serraf (France), Andras Szatmari (Hungary), Erik Thaulow (Norway), Pascal R Vouhe (France), Edmond Walma (Netherlands)

www.escardio.org/guidelines European Heart Journal (2010) doi:10.1093/eurheartj/ehq249

Classes of recommendations

- Evidence and/or general agreement that a given treatment or procedure is beneficial, useful and effective
- Conflicting evidence and/or divergence of opinion about the usefulness/efficacy of the given treatment or procedure:
  - Weight of opinion/evidence is in favour of usefulness/efficacy
  - Usefulness/efficacy is less well established by evidence/opinion
- Evidence and/or general agreement that the given treatment or procedure is not useful/effective and in some cases may be harmful

Levels of evidence

- Data derived from multiple randomized clinical trials or meta-analyses.
- Data derived from a single randomized clinical trial or large non randomized studies.
- Consensus of opinion of the experts and/or small studies, retrospective studies, registries.

Indications for Intervention in Ventricular Septal Defect

Ventricular Septal Defect
Follow-up

- **FU evaluation** should include assessment of AR, TR, degree of (residual) shunt, LV dysfunction, elevation of PAP, development of DCRV and development of discrete subaortic stenosis by echocardiography.
- Possible development of complete AV block requires attention (patients who develop bifascicular block or transient trifascicular block after VSD closure are at risk).
- Patients with LV dysfunction, residual shunt, PAH, AR, RVOT or LVOT obstruction should be seen every year, small VSDs in 3-5 year intervals; after device closure, regular follow-up until 2 years and then depending on the result every 2-4 years is recommended. After surgical closure without residual abnormality 5-year intervals.

Sudden Cardiac Death (SCD) in GUCH

- SCD is of particular concern in GUCH patients.
- Five defects with greatest known risk:
  - repaired Tetralogy of Fallot,
  - TGA with atrial switch,
  - congenitally corrected TGA,
  - Aortic Stenosis,
  - Univentricular hearts
- Unexplained syncope is a warning event requiring careful evaluation of arrhythmia.
- Although various risk factors have been defined, algorithms for risk assessement and indications for ICD implantation have so far not been well established.

General Indications for EP and ICD Implantation in GUCH

- ICD implantation is indicated in survivors of cardiac arrest after exclusion of reversible causes.
- Patients with spontaneous sustained VT should undergo invasive haemodynamic and EP evaluation. Recommended therapy includes catheter ablation or surgical resection to eliminate VT. If that is not successful, ICD implantation is recommended.
- Invasive haemodynamic and EP evaluation is reasonable in patients with unexplained syncope and impaired ventricular function. In the absence of a defined and reversible cause, ICD implantation is reasonable.
- EP testing may be considered for patients with ventricular couplets or non-sustained VT to determine the risk of sustained VT.

www.escardio.org/guidelines

Review of SCD in ACHD
Fallot : rare event

Author	Findings	Mean F/U (years)	SCD incidence/ decade (%)
Murphy et al. [30]	SCD in 6% of 163 cases	30	2.0
Nollert et al. [34]	SCD in 3% of 490 cases	25	1.2
Silka et al. [47]	SCD in 2.6% of 445 cases	22	1.8
Norgaard et al. [35]	SCD in 5.6% of 125 cases	25	2.2
Gatzoulis et al. [12]	SCD in 6% of 793 cases	21	3.0

Silka, Pediatr Cardiol (2012)

Tetralogy of Fallot after repair
Follow-up (1)

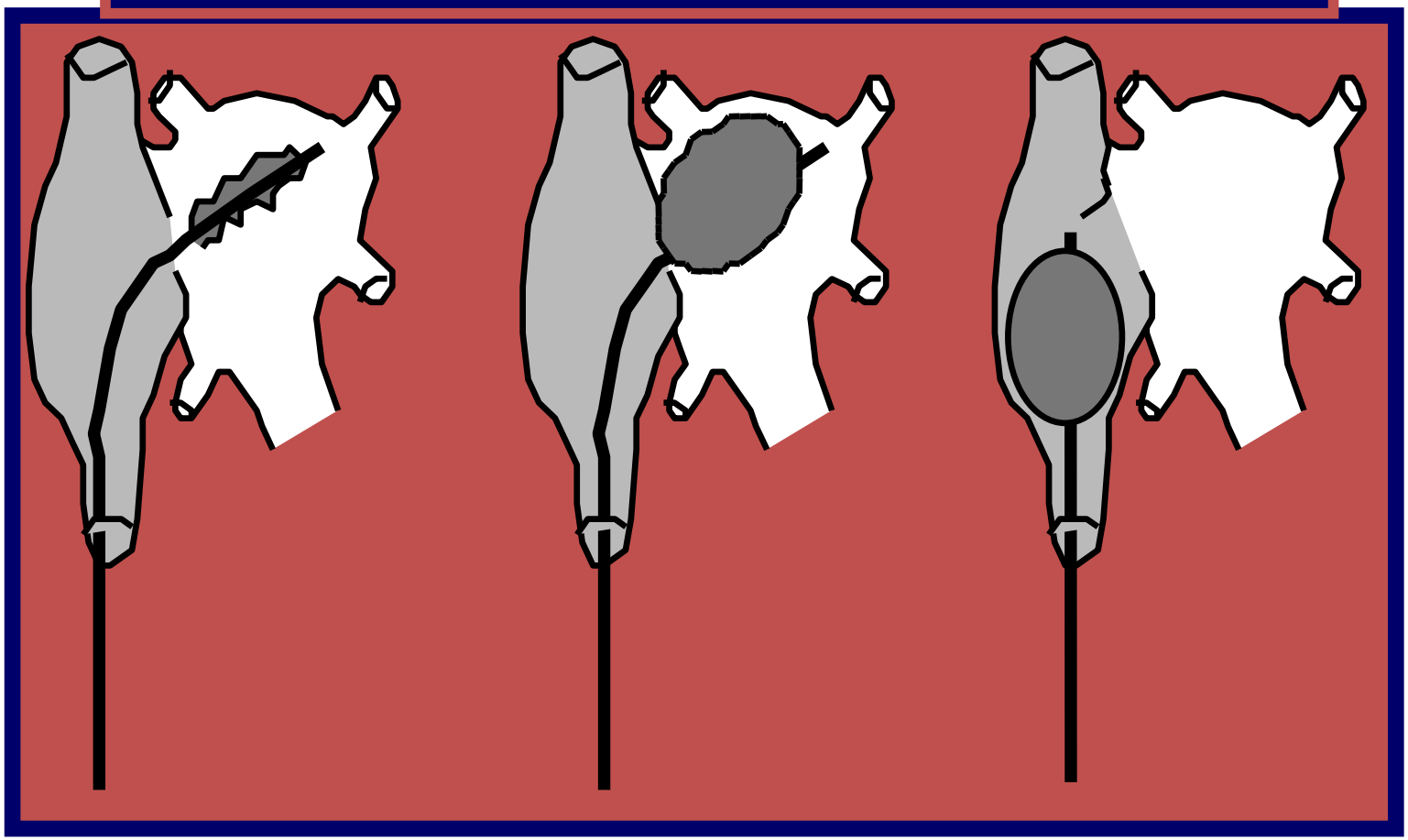
All patients should have regular follow-up in a specialized GUCH center, in general annually

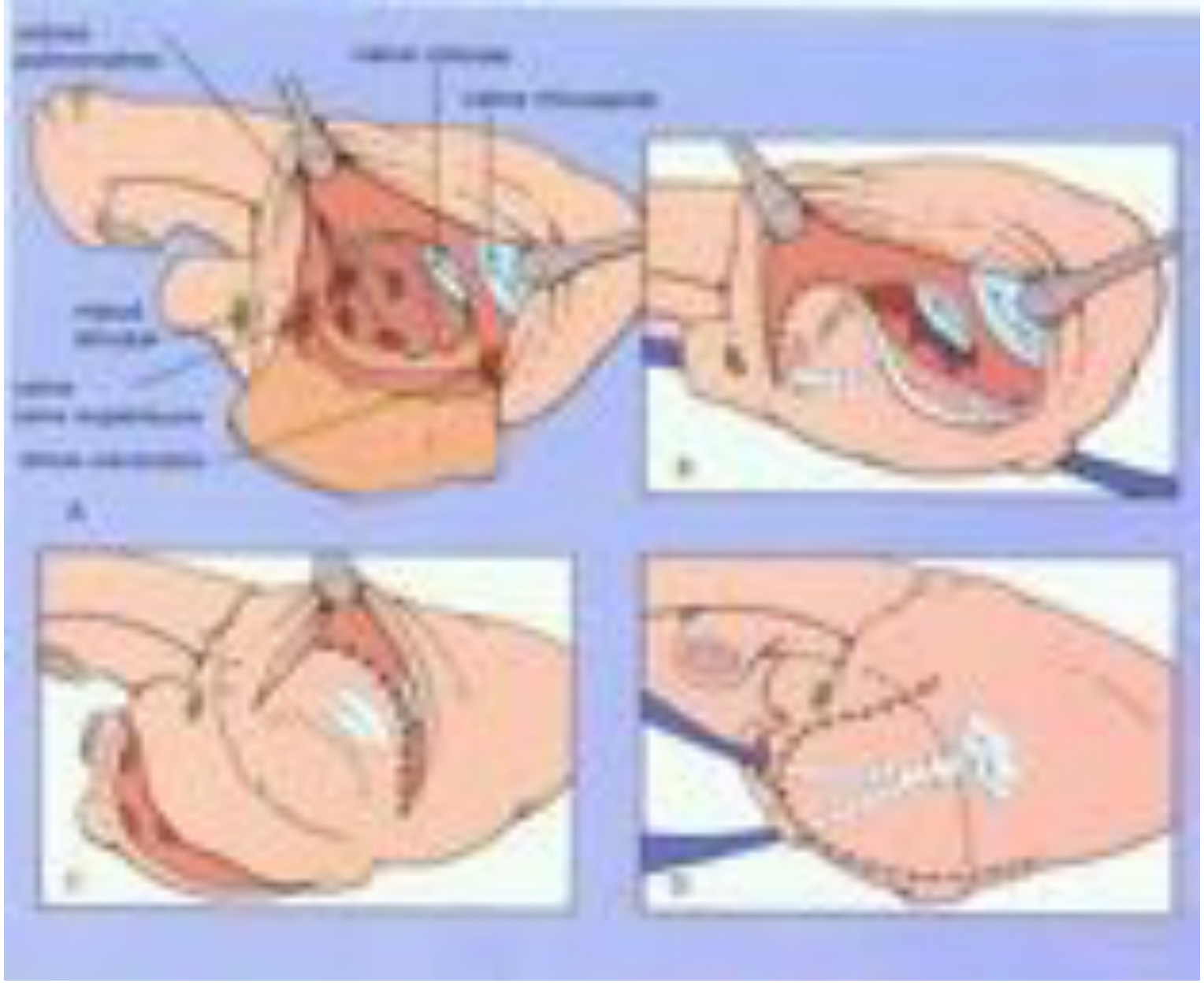
Late complications to look for:

- **Pulmonary regurgitation (PR):** significant PR is almost always encountered following a transannular patch repair. May eventually lead to symptomatic RV dilation and dysfunction.
- **Residual RVOTO:** can occur at the infundibulum, at the level of the pulmonary valve and main pulmonary trunk, distally, beyond the bifurcation and occasionally into the branches of the left and right PAs.

Transposition des gros vaisseaux

Procedure de Rashkind

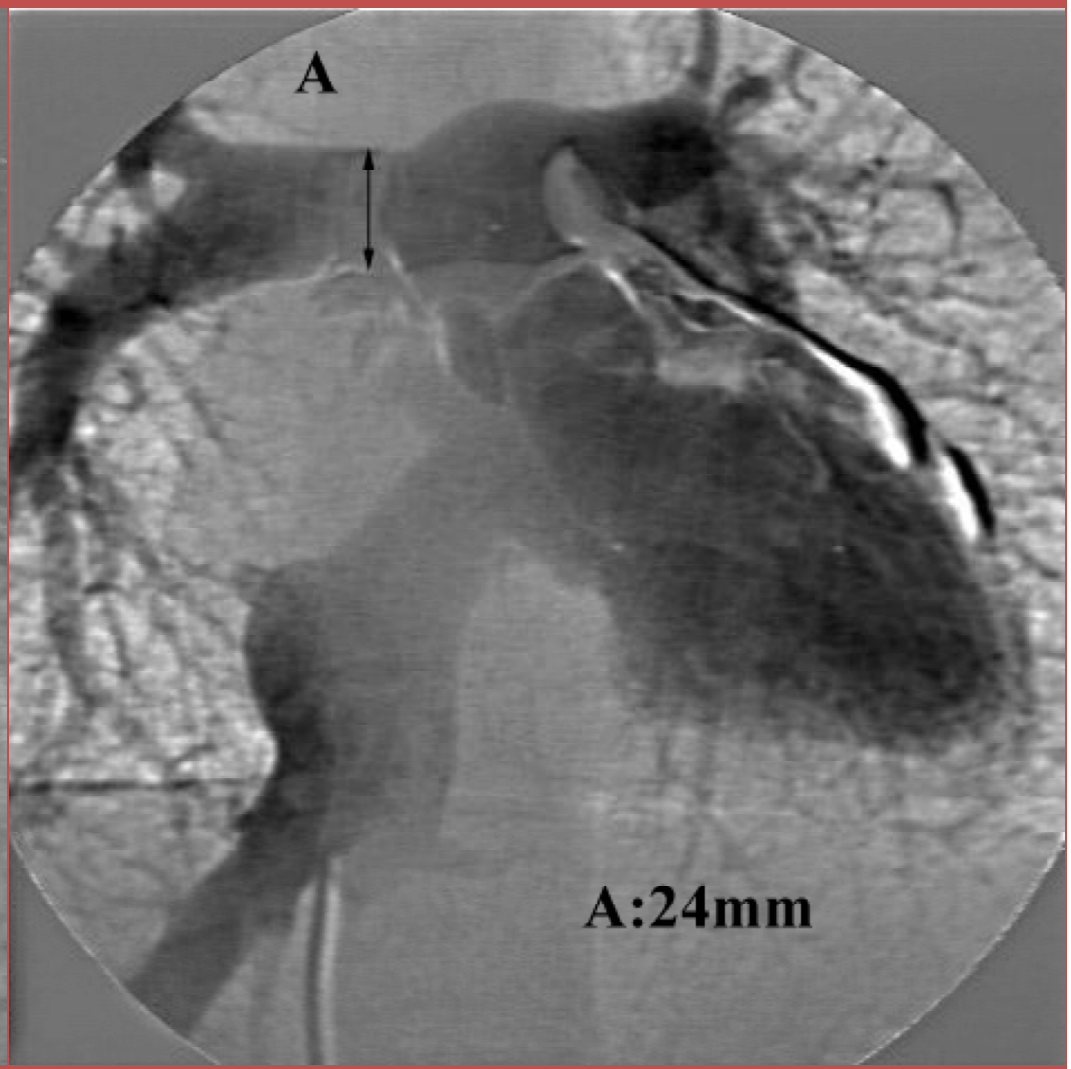




Transposition des gros vaisseaux switch atrial

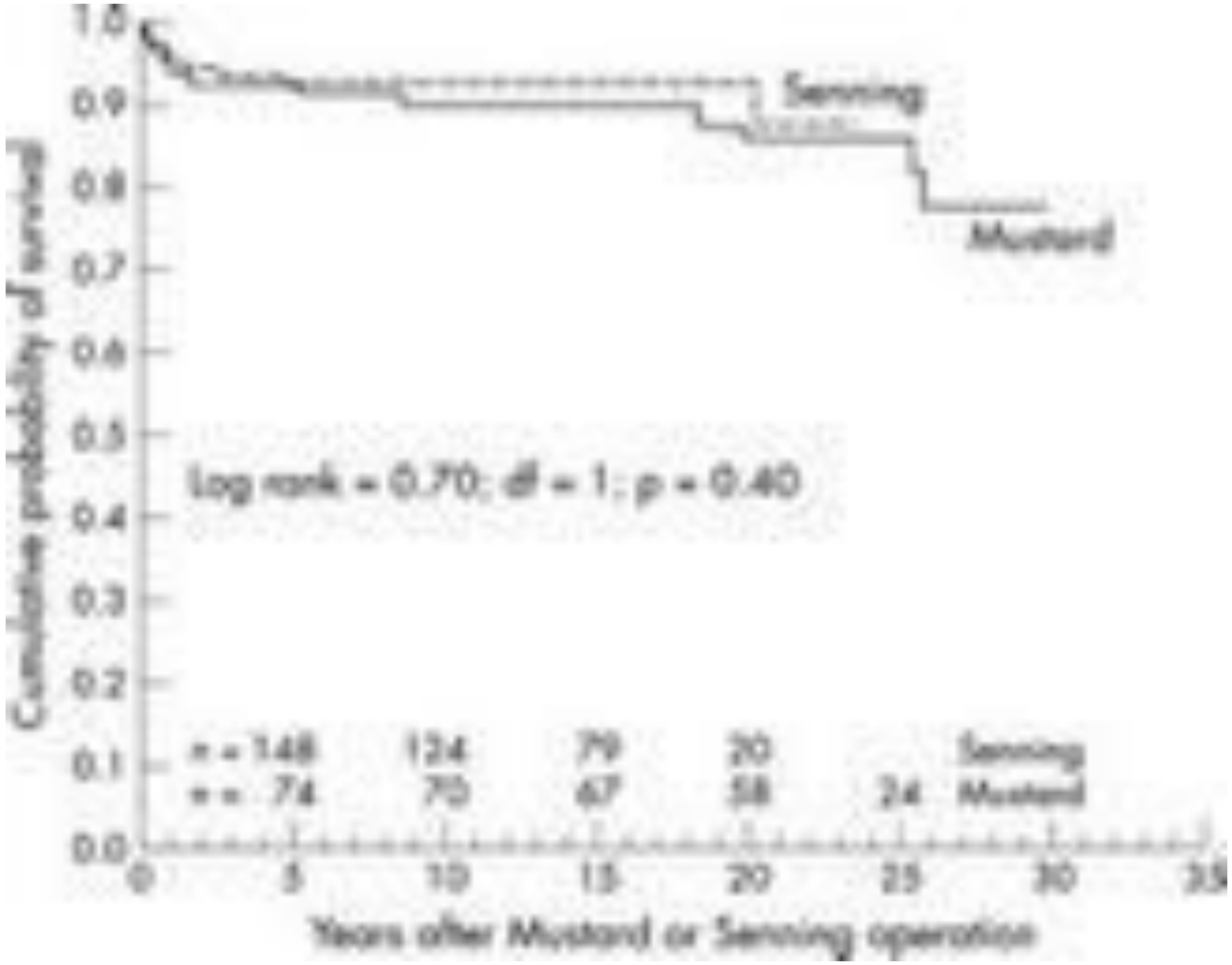

Chenal des VP

Intervention de Mustard



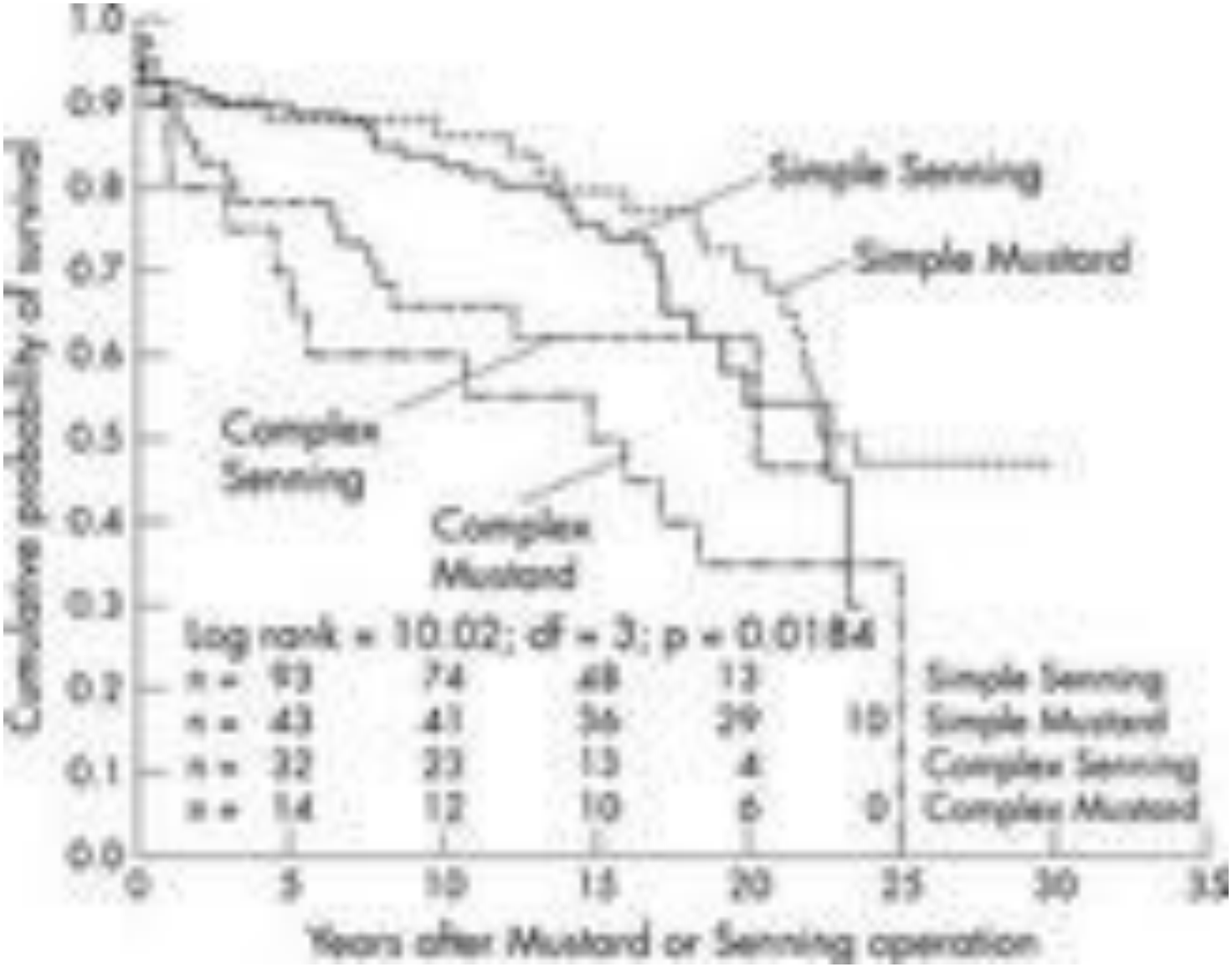
4 problèmes intriqués

- **Troubles du rythme auriculaires (flutter)**
- **dysfonction sinusale**
- **sténose ou thrombose des chenaux**
- **dysfonction du VD systémique - IT**
- **Médicaments (B-, cordarone)**
- **ablation radiofréquence**
- **PM : mais**
- **chenaux à vérifier**
- **cerclage transplantation cardiaque**

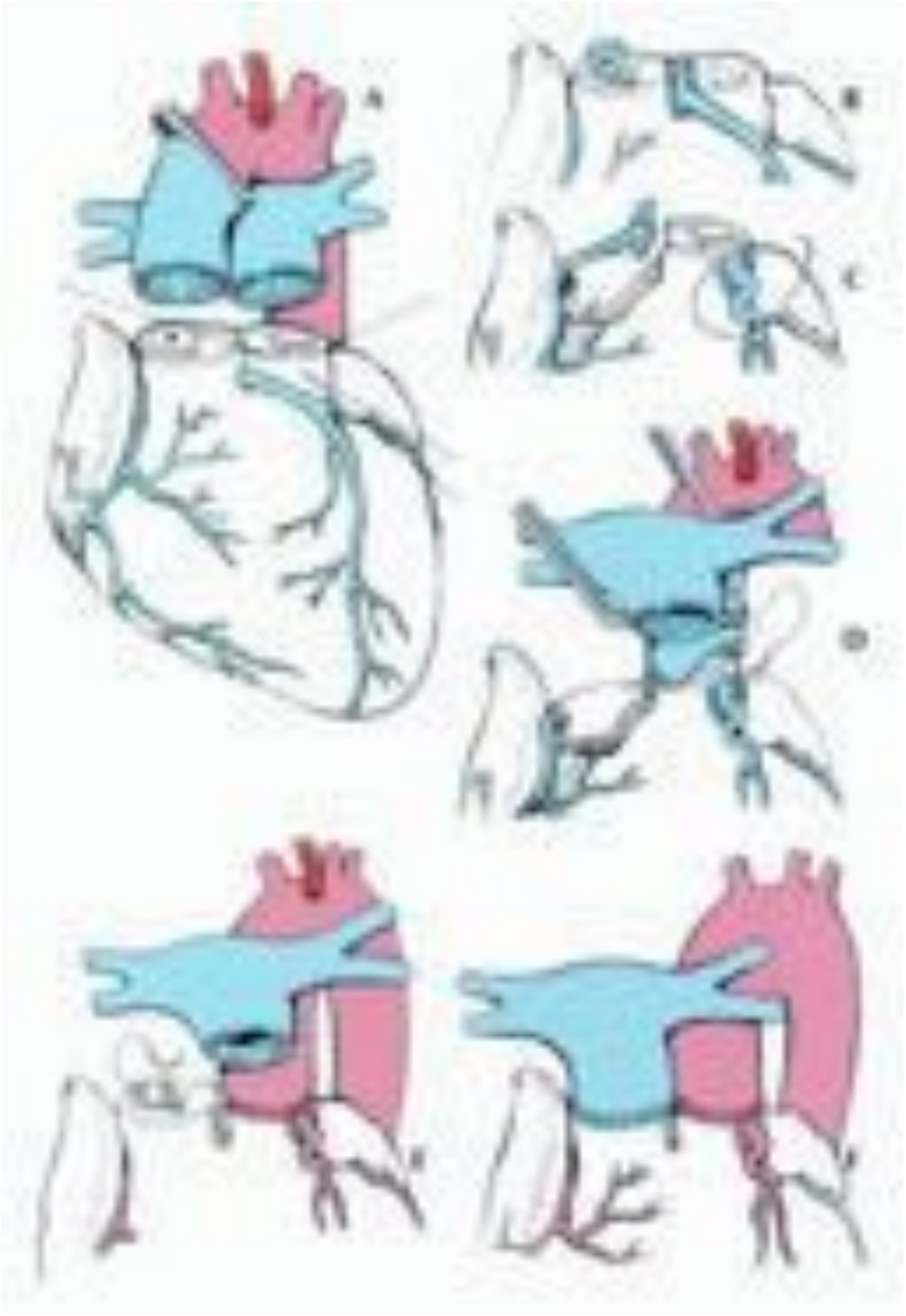


% survival for Senning and Mustard (survival after 30 days)

heart 2004 , Moons



% survival free of arrythmias for Senning and Mustard (survival 30 d post op)



Transposition des gros vaisseaux switch artériel

Arterial Switch Operation (ASO)
Long term results

Laurence Iserin

HEGP, M3C, Paris

2nd European Advanced Course in Grown up congenital heart patients Focus in Transposition

Paris

4th - 5th March 2011

History (1)

- Mustard and colleagues in 1954 unsuccessful ASO. (transfer only the left coronary artery).
- attempts to switch the great arteries without coronary transfer : failed.
- first successful arterial switch operation in a patient with TGA and a large VSD : Jatene 1975
- high early operative mortality in simple TGA
- modified techniques :
  - baffling of the coronary arteries to a surgically created aorto-pulmonary window
  - Lecompte and colleagues : transferring the distal pulmonary artery anterior to the ascending aorta (without conduit interposition) (1980)
-

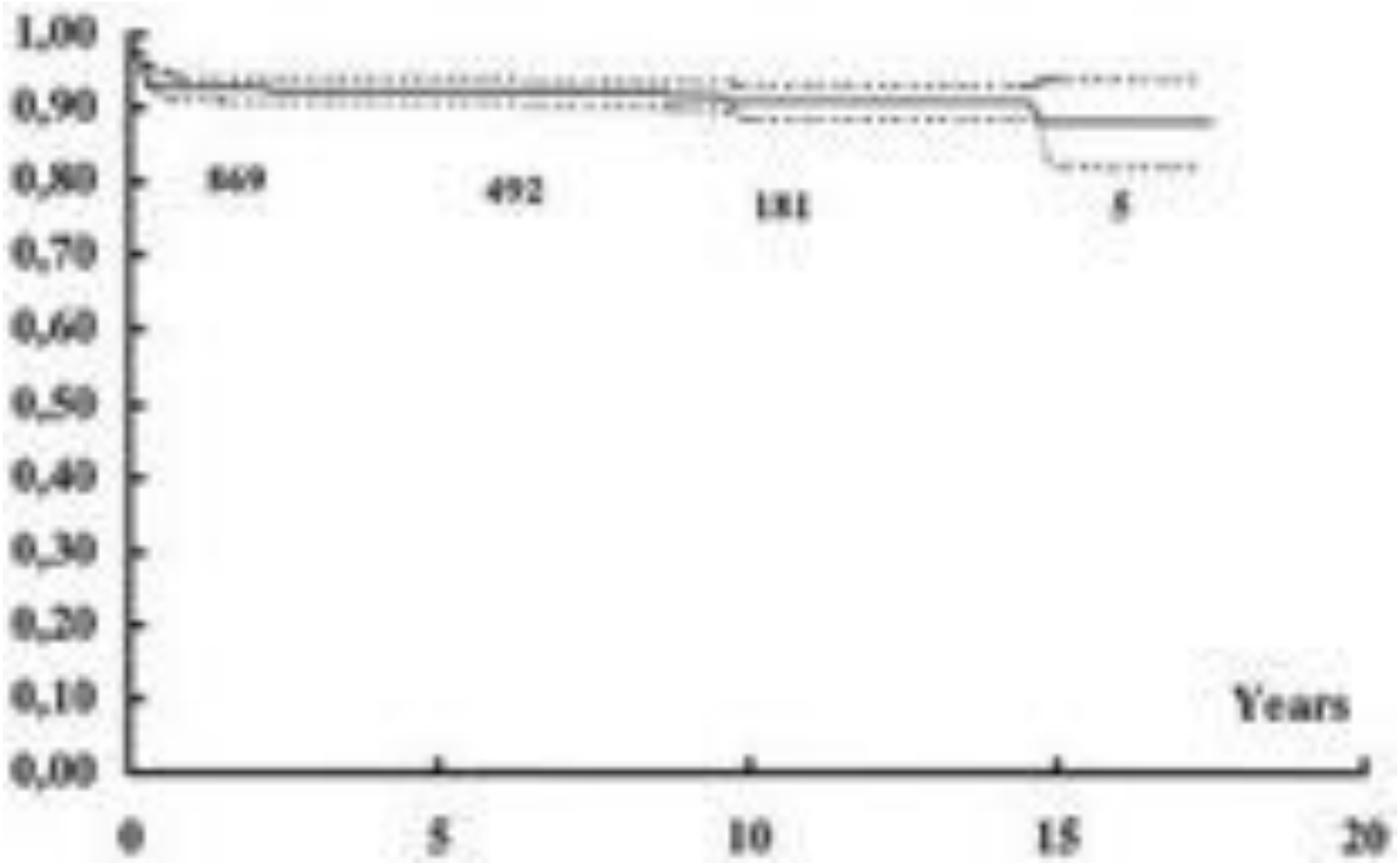
History (2)

- In TGA and intact ventricular septum (IVS), ASO failed because of left ventricular dysfunction.
- Yacoub and associates devised a" two-stage" pre PA banding (to increase LV Mass), followed by an ASO several months later.
- Rapid " two-stage" for TGA-IVS Children's Hospital in Boston PA banding and ASO within 7 days.
- ASO: surgical procedure of choice for correction of transposition of the great arteries (TGA) with or without (VSD).

  earlier eras: 15% early operative or hospital mortality

  Now : 5% early operative or hospital mortality in both simple TGA and TGA with VSD

Actuarial survival free of coronary events for 1304 patients



Legendre, A. et al. Circulation 2003;108:II-186-II-190

Circulation

Arterial Switch Operation (ASO): surgical technique

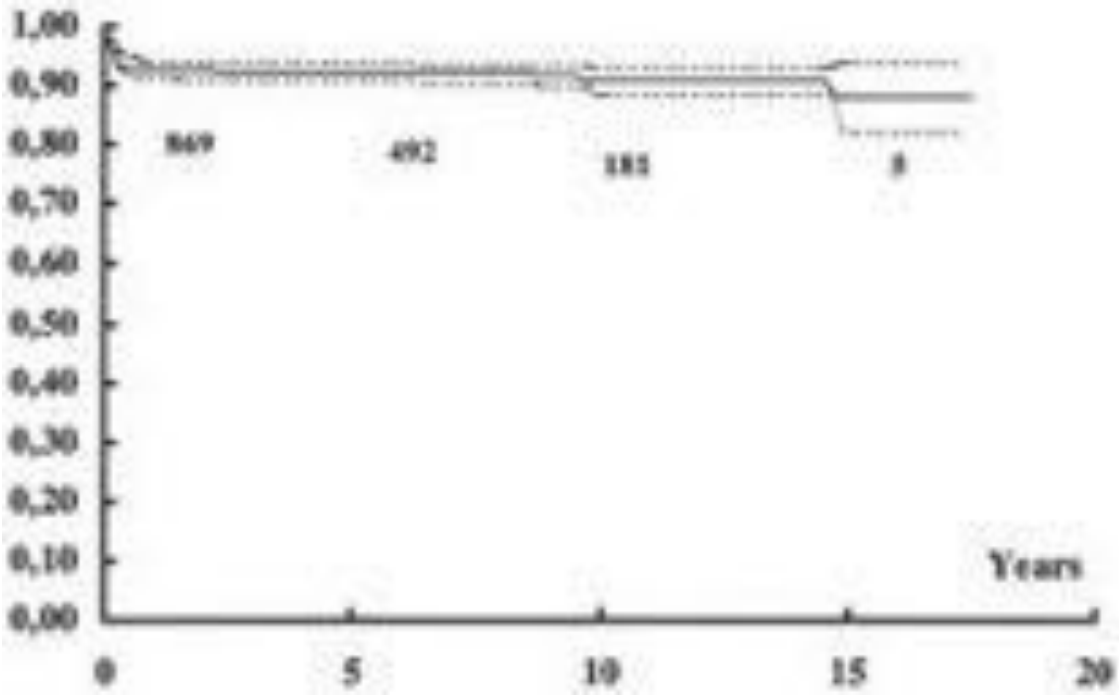
TGA

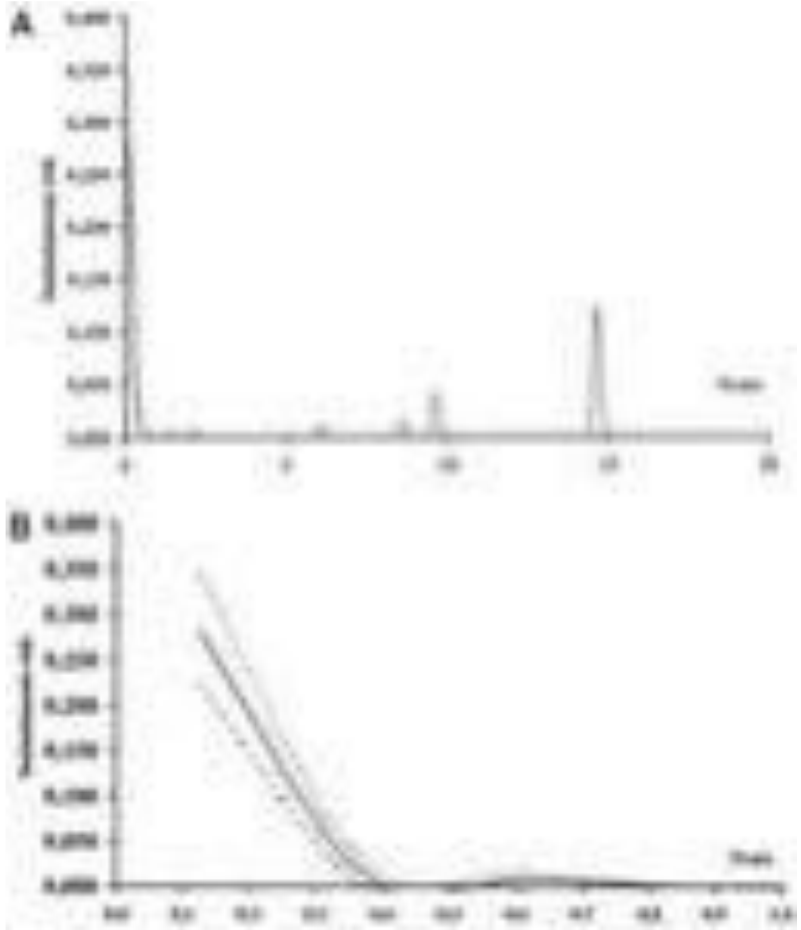
ASO

Normal heart

Coronary Events After Arterial Switch Operation for Transposition of the Great Arteries

A. Legendre, MD; J. Losay, MD; A. Touchot-Kone´ et al.





1982 - 2001

- •1304 ASO
- • 1 198 hospital survivors

Neoaortic valve regurgitation

- Underestimated complication of anatomic repair of TGA.
- Anatomic pulmonary valve (thin leaflets, little collagen and elastic tissue) : neoaortic valve after ASO.
- Mild regurgitation : 35% of patients
- Moderate to severe : 5% patients.
- Frequency of the regurgitation after ASO increases with time
- Isolated cases of valve replacement.
- In our serie (review of patients born before 4 AVR done at age (16, 18,22 , 23)

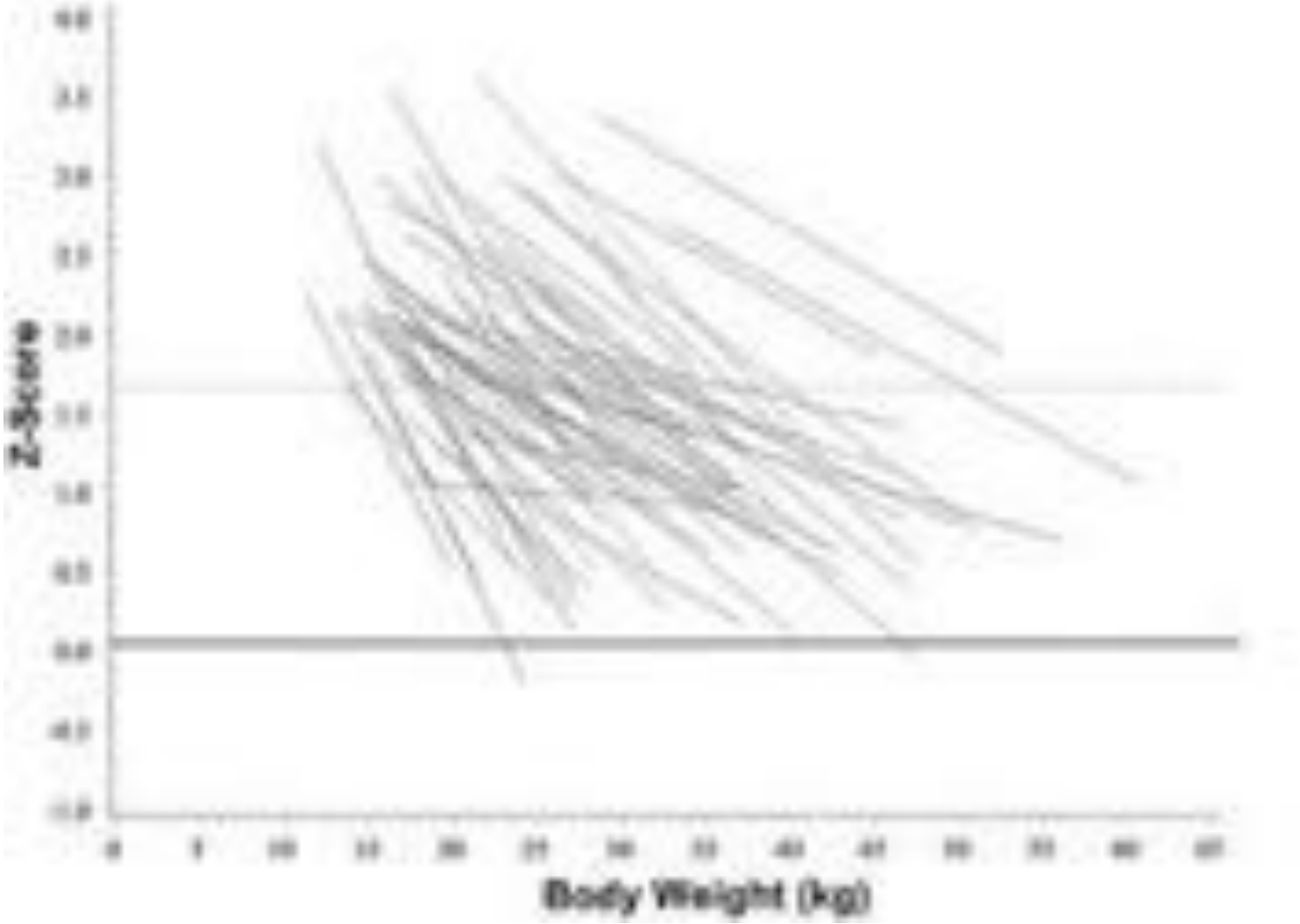
Aortic Root

- After ASO Neo-aortic valve and sinus are larger than normal.
- first year of life : rapid dilatation of the new aorta
- Further : active growth with tendency towards normalization of the valve and sinus size.

Echocardiographic end-systolic measurements of the neo-aortic root vs body weight

60 children (solid lines) after neonatal ASO

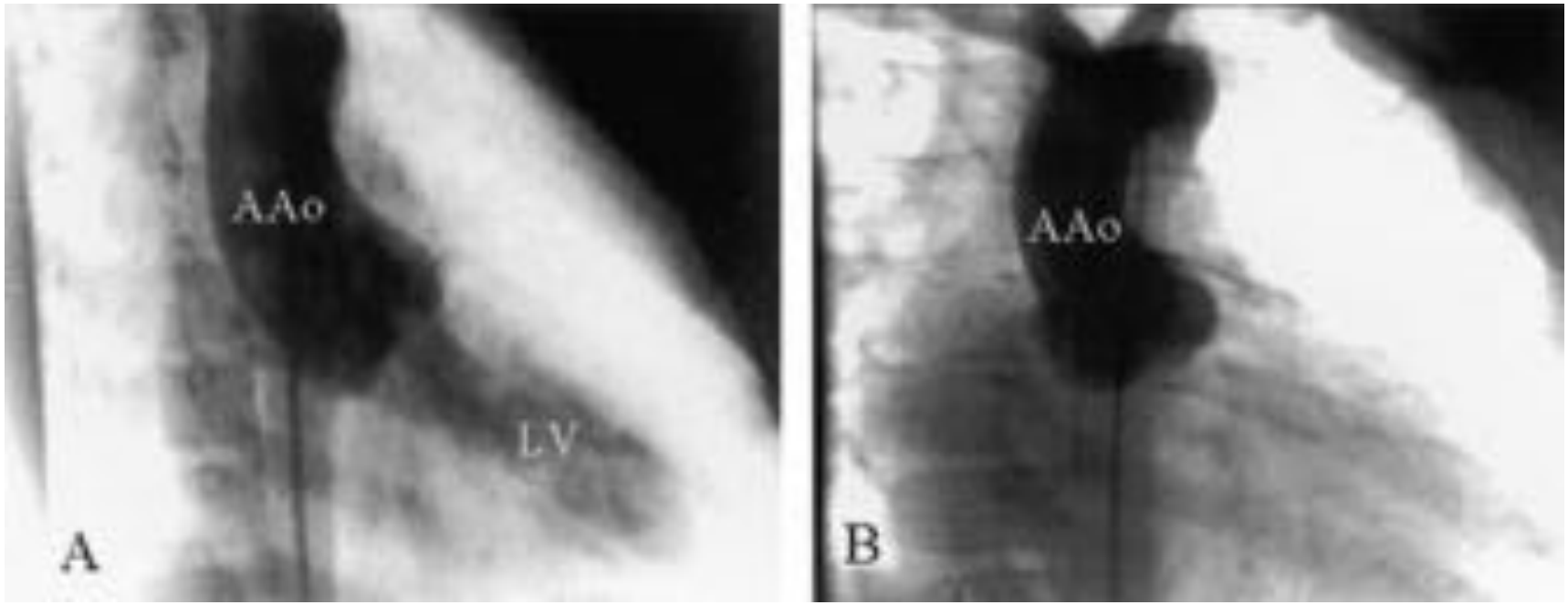
mean age 5.3 {+/-} 1.6 years and mean age 10.5 {+/-} 1.6 years



Hovels-Gurich H. H. et al.; Ann Thorac Surg 2003;75:935-943

THE ANNALS OF
THORACIC SURGERY

Morphology of the reconstructed Aorta: risk factor for Aortic regurgitation



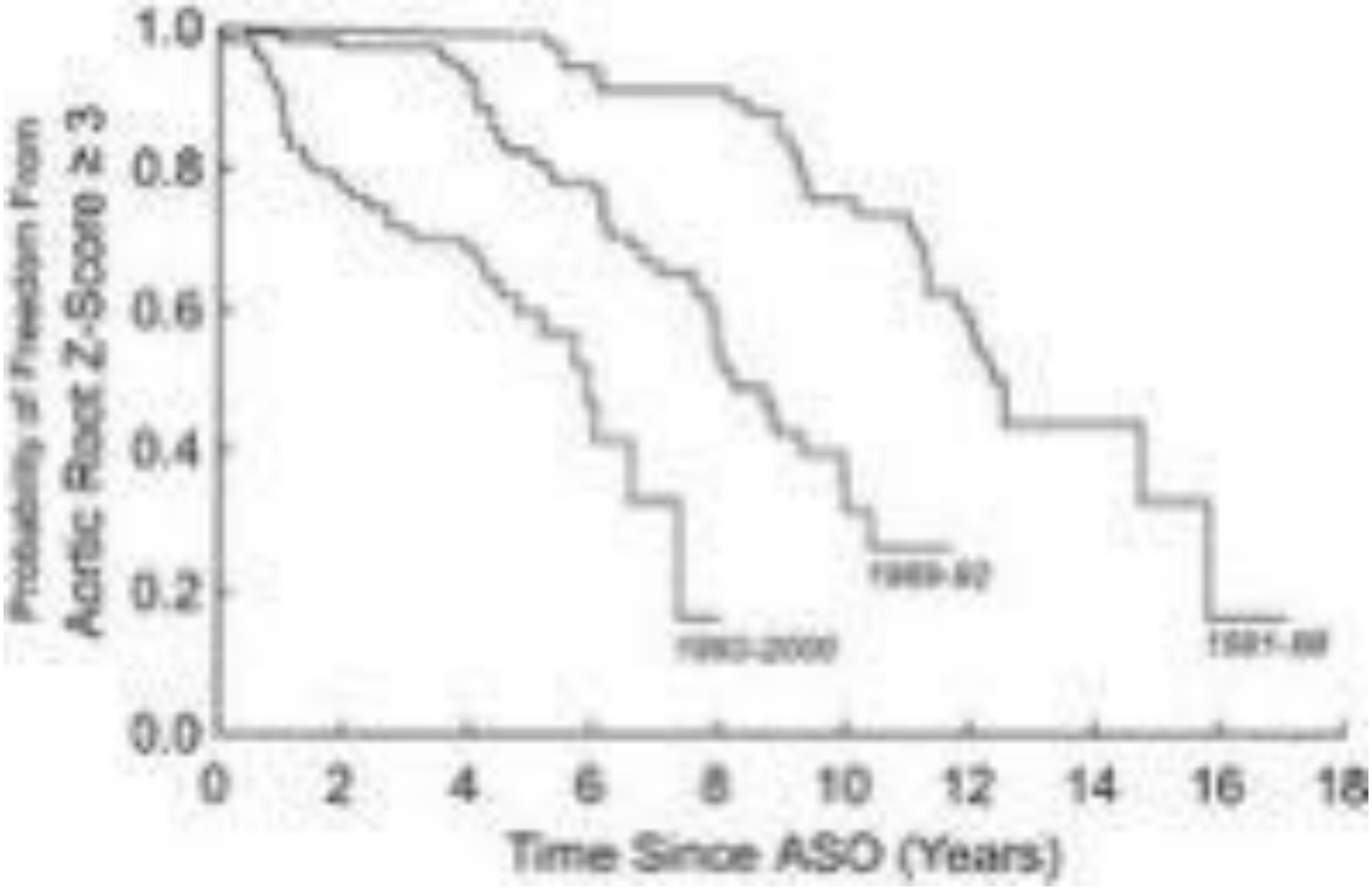
More AR NO AR

Formigari R. et al.; J Thorac Cardiovasc Surg 2003;126:1753-1759

The Journal of
THORACIC AND
CARDIOVASCULAR SURGERY

Freedom from aortic root dilation over time since ASO

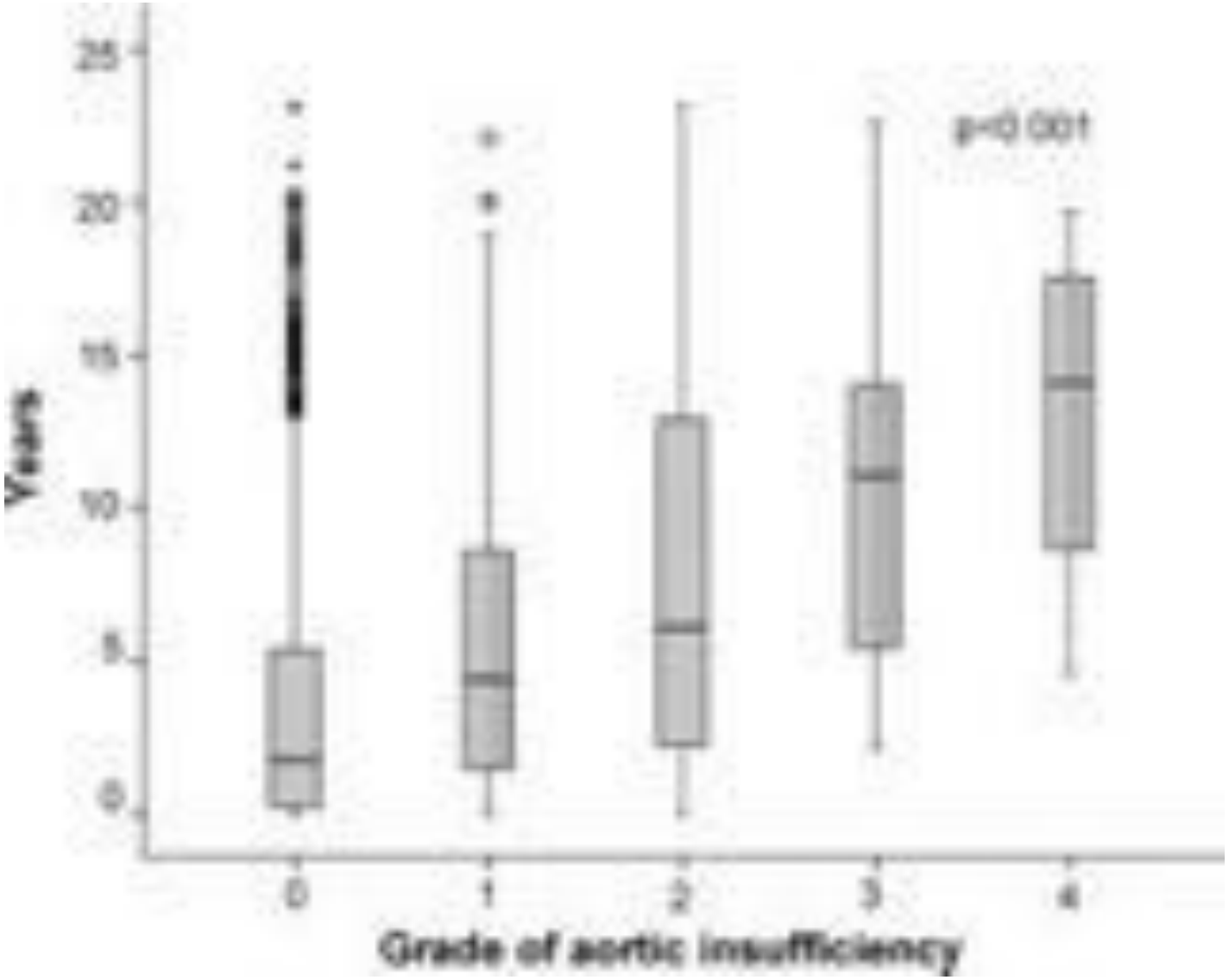
3 time periods : 1981 to 1988, 1989 to 1992, and 1993 to 2000 (P<0.001)



Schwartz, M. L. et al. Circulation 2004;110:II-128-II-132

Circulation

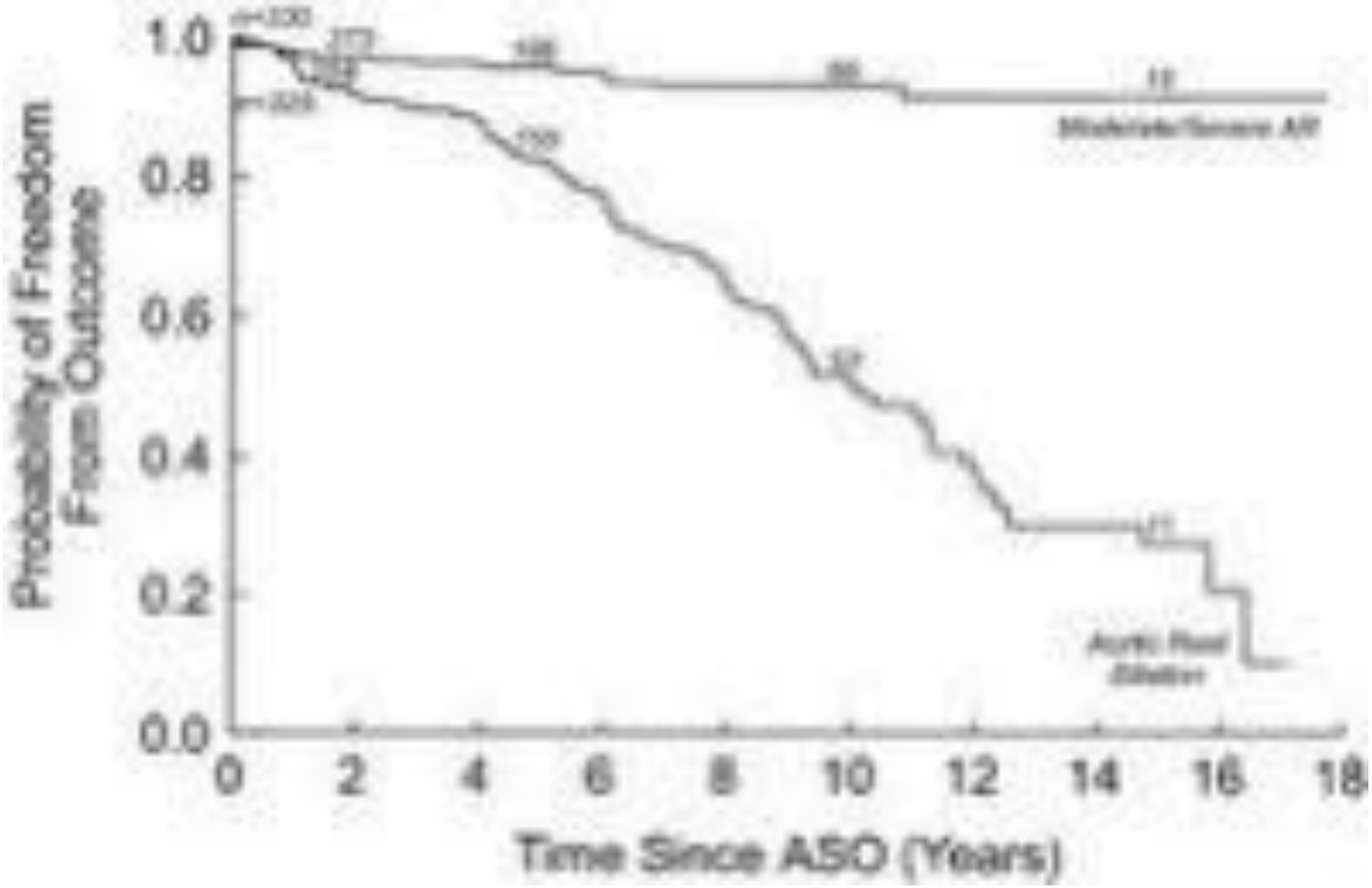
Development of Aortic Insufficiency over time



Lange R. et al.; Eur J Cardiothorac Surg 2008;34:711-717

EUROPEAN JOURNAL OF
CARDIO-THORACIC SURGERY

Freedom from neo-aortic root dilation (neo-aortic root z-score >=3.0) and probability from at least moderate neo-aortic regurgitation



Schwartz, M. L. et al. Circulation 2004;110:II-128-II-132

Aortic valve repair for AR after ASO?

Imamura M. et al.; Ann Thorac Surg 2000;69:607-608

THE ANNALS OF
THORACIC SURGERY

RV to PA obstruction

- Stenosis of the reconstructed pulmonary artery : distortion or retraction of the pericardial patch used to cover the defect left by the coronary artery harvest.
- Distortion of the main and peripheral pulmonary arteries as a result of the anterior placement of the bifurcation (Lecompte's maneuver)
- Prevention :
- . Extensive dissection of the pulmonary arteries to the hilum to avoid tension on the anastomosis
- Use of a larger patch, autologous pericardium (growth potential)
- Reoperation rate for a pulmonary artery stenosis at 5 years : 25% in early years Vs to 5% now.

Neopulmonary valve regurgitation

- incidence : 9% to 80%. (echo/MRI studies)
- Some patients will have the evolution of TOF
- (arrhythmias and RV dilatation)
- 1 Melody stent implanted for obstruction and PR with success

Coronary artery obstruction and myocardial perfusion

coronary events : 2% - 11%.

Early events

- coronary events most often occur immediately after the ASO main cause of death or morbidity
- early postoperative period : risk factors
  - coronary anatomy
  - surgical technique difficulties
- Causes

kinking or torsion

extrinsic compression by biological glue (immediate coronary revision or reoperation)

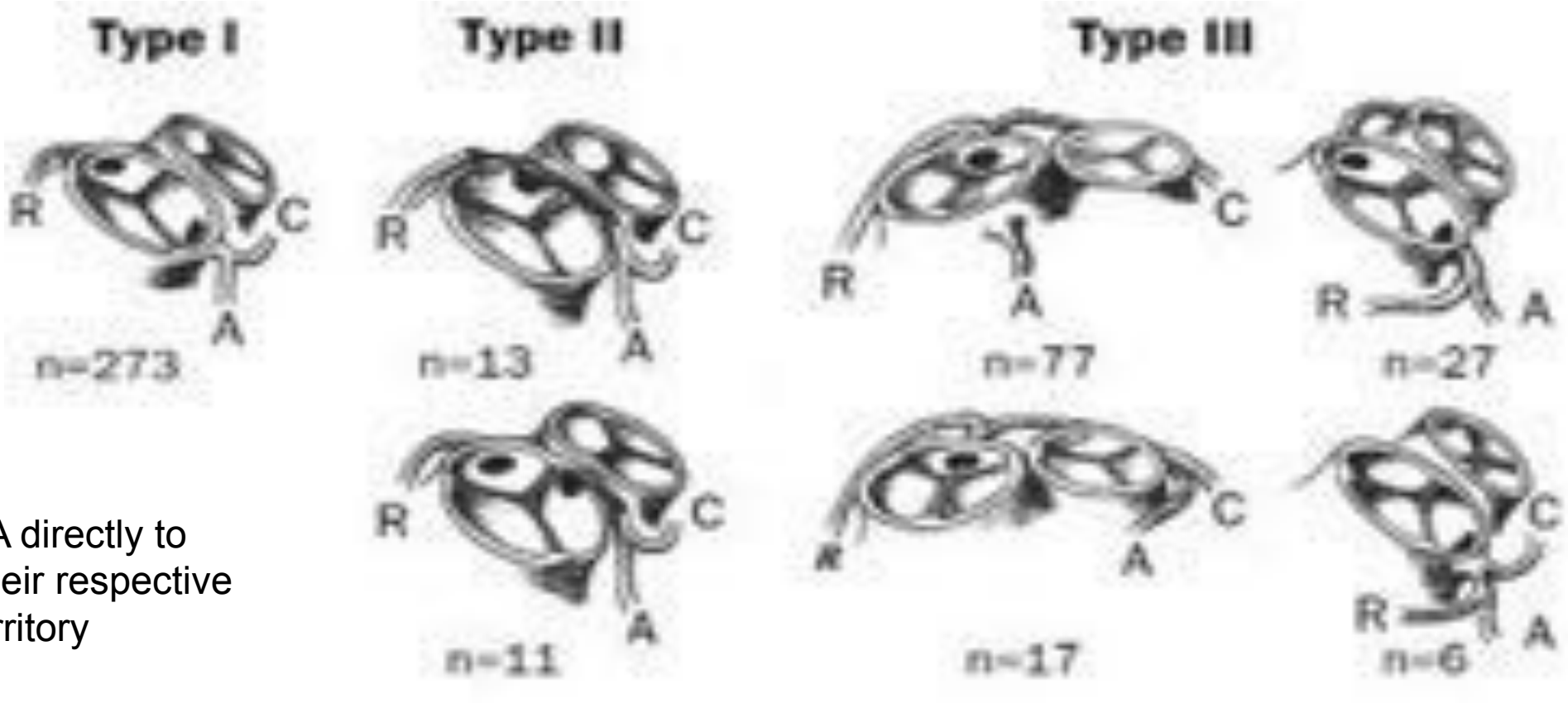
Coronary artery obstruction and myocardial perfusion

Late events

- Late coronary mortality and myocardial infarction : rare (prevalence < 2%)
- Coronary revascularizations occur late (most often > 3 years)
- Causes
  - progressive fibrocellular intimal thickening
  - stretching of the coronary artery with growth.
  - Position of the reimplanted coronary arteries

Coronary artery obstruction and myocardial perfusion

risk factors for early coronary complications (type 2 and 3)



CA directly to their respective territory

CA coursed between Ao and PA

CA make a loop around Ao or PA or both

Intramural course more frequent in type 2 and 3

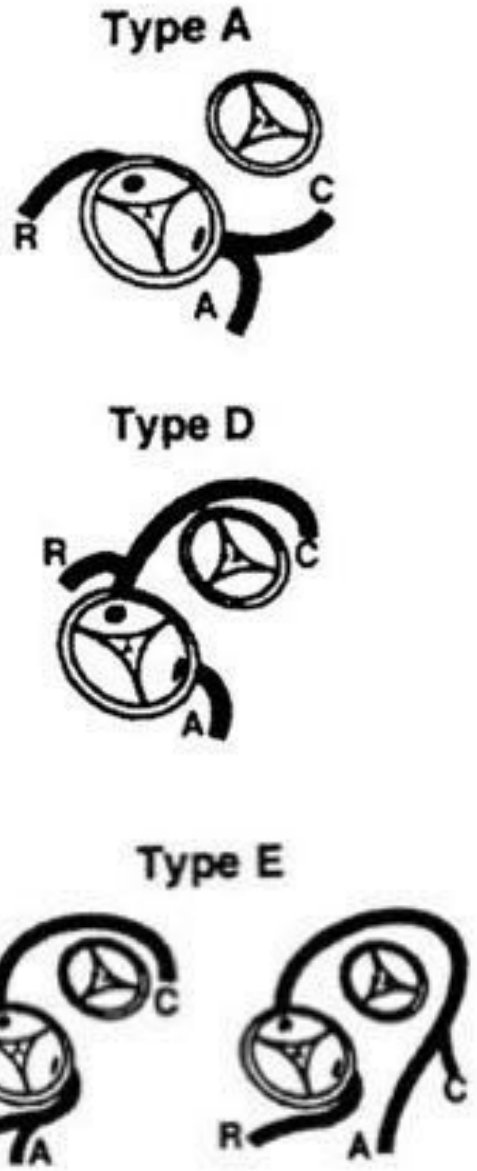
Lancet 2001; 357: 1826–30

Coronary artery lesions and coronary artery pattern

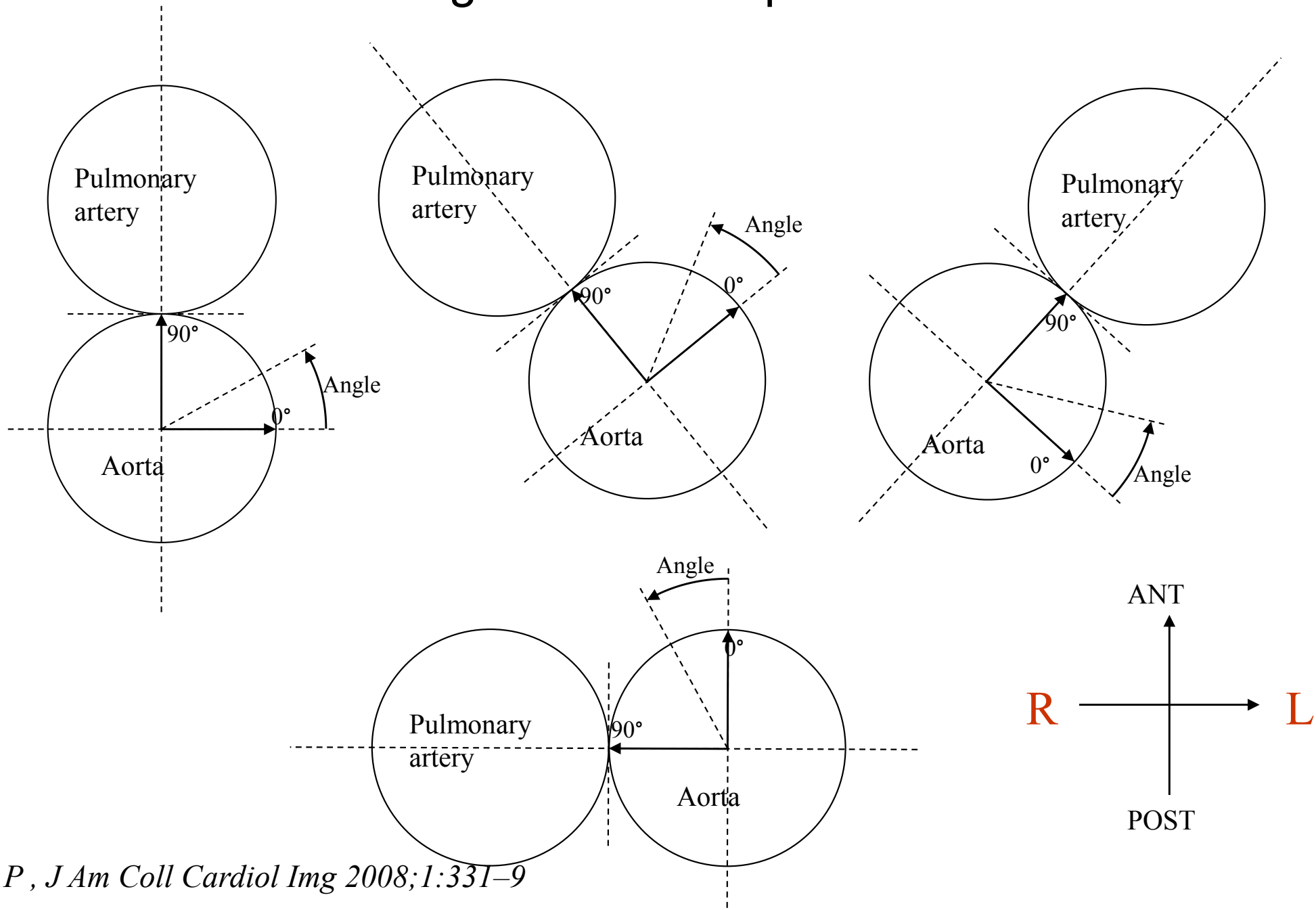
	Nb patients	Nb lesions	Nb LCA lesions	Nb RCA lesion	Nb retro-aortic Cx lesion
Type A	88	11 (12.5%)	10	1	
Type B	3	0			
Type C	12	2	2 (IVA intramurale)		
Type D	29	10 (34%)	8	0	2
Type E	14	3 (21%)	1	0	2

147 patients who underwent ASO;
mean age : 7 years

Ou P , J Am Coll Cardiol Img 2008;1:331–9



Possible Mechanism for progressive coronary lesions : Angle of the reimplanted ostia



Ou P , J Am Coll Cardiol Img 2008;1:331–9

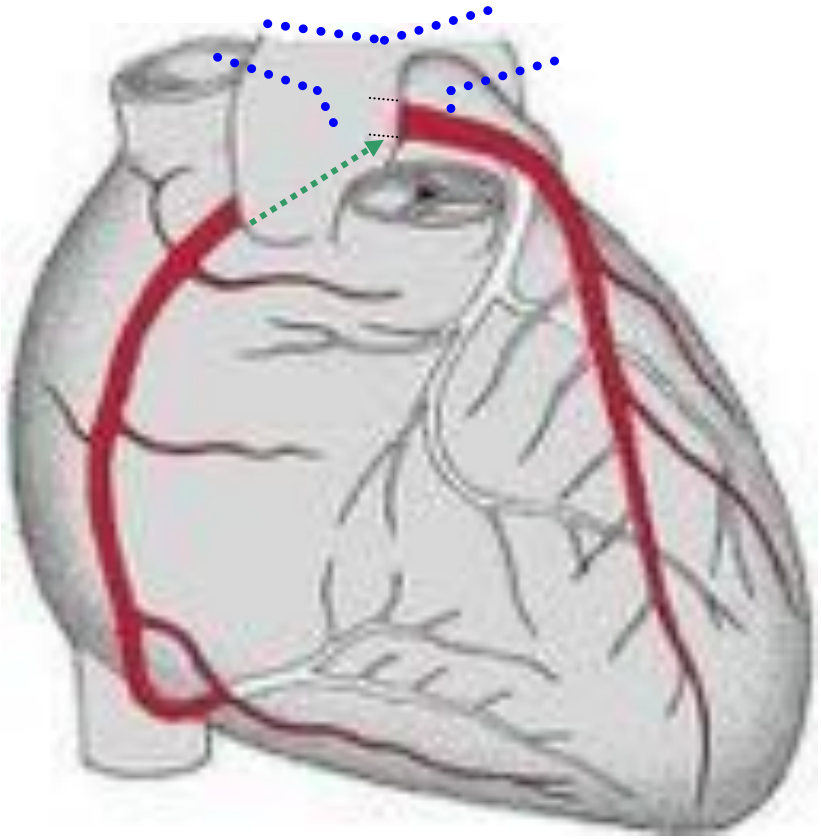
General condition

- **Functional status**
  exercise performance in this performance during exercise is generally excellent after the arterial switch operation. However, even though essentially all surviving patients are fully active and without limitation
- evidence of stress-induced myocardial ischemia continues to be a concern in this population.

- Continued monitoring of exercise performance, especially for ischemia, would appear to be warranted as this population ages and begins to participate in more vigorous athletic activities.

Mechanisms for progressive coronary lesions

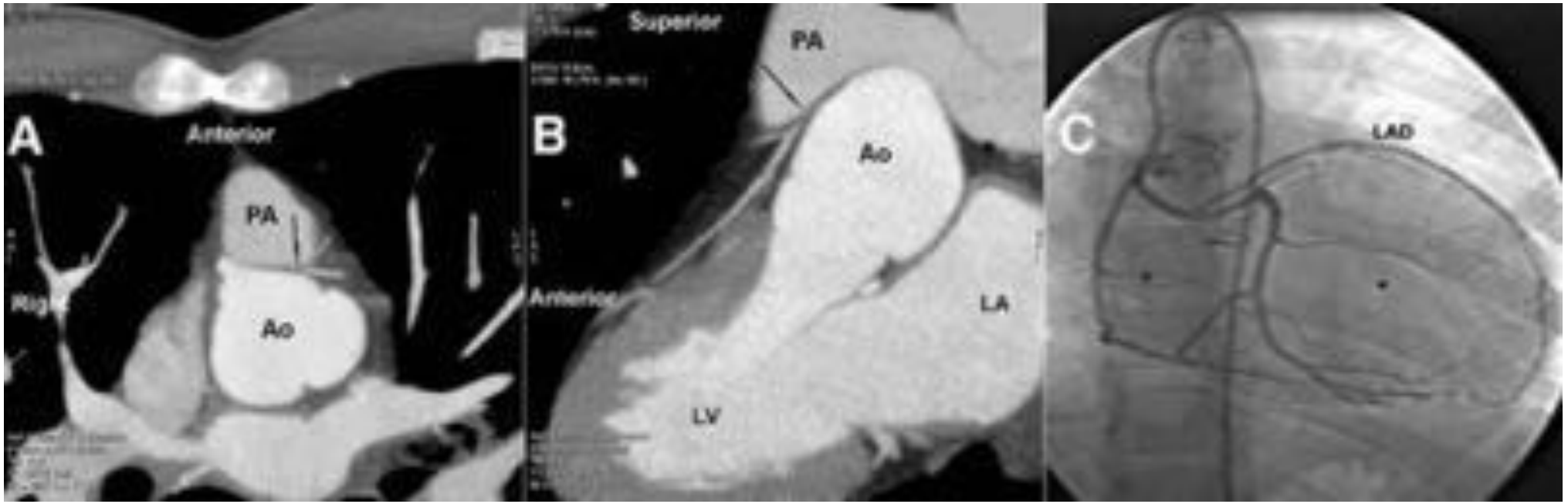
- Long distance between right ostium and pulmonary bifurcation (low implantation of the ostium of the retro aortic circumflex in the right sinus) increases the occurrence of retro-aortic circumflex artery lesion



Ou P , J Am Coll Cardiol Img 2008;1:331–9

Detection of coronary by CT coronary angiography

- 64-slice CT performs as well as invasive angiography for detecting significant coronary lesions in the majority of children who have undergone the arterial switch procedure for TGA
- CT angiography was able to detect 100% of the coronary artery lesions detected by the invasive angiography + 6 NS lesions.



Ou P , J Am Coll Cardiol Img
2008;1:331–9

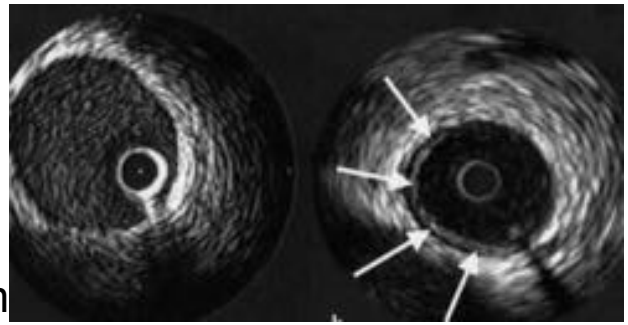
Spatial relations between coronary and great vessels

- Left ostium anterior location enhances occurrence of left coronary artery lesions : extrinsic compression by the Pulmonary Artery

Myocardial perfusion abnormalities

studies following the ASO : high incidence of perfusion defects.

- reduction of regional coronary flow reserve (ability of coronary flow to increase under hyperemic stimulation).
- rarely correlated with electrocardiographic, echocardiographic, or angiographic changes,
- clinical significance?.
- Rarity of regional left ventricular wall motion abnormalities : reassuring
- . Exercise testing appears
  - detect ischemic damage
  - exercise-induced ventricular arrhythmias
  - Uncertainty
  - IVUS study : proximal eccentric intimal proliferation
    - all children : coronary artery involvement
    - with 50% having moderate-to-severe lesions (0.3 mm)
- long-term development of coronary circulation after coronary reim
- evolution of atherosclerosis and coronary flow reserve.



coronary flow reserve ?

	Population	Imaging technique	coronary flow reserve
Bengel 1998	SA ealry/late-vs young adults	PET-scan	
Hauser 2001	SA and Ross -vs adults	PET-scan	
Oskarson 2002	SA (etude pts ferm CA)	Intra Coronary Doppler	Normal
Gagliardi 2005	SA-vs other CHD	Intra Cororary Doppler	
Turner 2010	SA-vs litterature	PET-scan	Normal
Falkenberg 2010	Piglets SA- vs piglets	In vitro: Langendorff	

Rhythm disturbances

- during ASO little intra atrial manipulation (except repair of ASD). One of the major theoretical advantages of anatomic correction is that the limited atrial procedure should result in a significantly improved rhythm status of the survivors.
- The reported incidence of arrhythmias after arterial switch is 3% to 7 (vs 13% - 100% in atrial switch)
- However, as the follow-up is relatively short, one must remain vigilant in this regard, since such problems may become apparent after many years.
- Pace maker implantation can be needed (in our series, all in TGA and VSD)
- But AV bloc can happen late in life .

Neuro development status

- ASO : combined circulatory arrest and low flow bypass
- At 8-year follow-up (Bellinger et al , J TCS 2003)
- overall physical and psychosocial health status was similar to that of general population
- But increased problems with attention, learning, speech, and developmental delay reported by parents .
- use of total circulatory arrest generally associated with greater functional deficits than is use of low-flow cardiopulmonary bypass

both strategies : increased risk of neuro developemental vulnerabilities.

Conclusion

- The incidence of moderate AR (15%) is relatively low even 20 years after ASO.
- The arterial switch operation has been universally recognized as the therapy of choice for children born with transposition of the great arteries.
- As experience with this operation has increased, mortality has been lowered and is consistently below 10% in most contemporary series.
- Most children who have undergone the ASO have also enjoyed normal growth, development, and cardiac function.
- . Incidence of late reintervention, whether in the form of therapeutic cardiac catheterization or operation, is not uncommon in this patient population. Therefore, it is important to realize that even though it is almost three decades since the first ASO was performed and morbidity and mortality has been lowered yet
- one must remain vigilant to identify newly emerging problems, since such unrecognized problems may become apparent only after follow-up of many years.