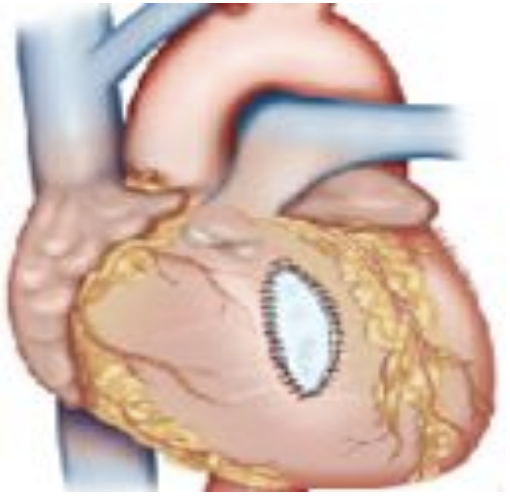
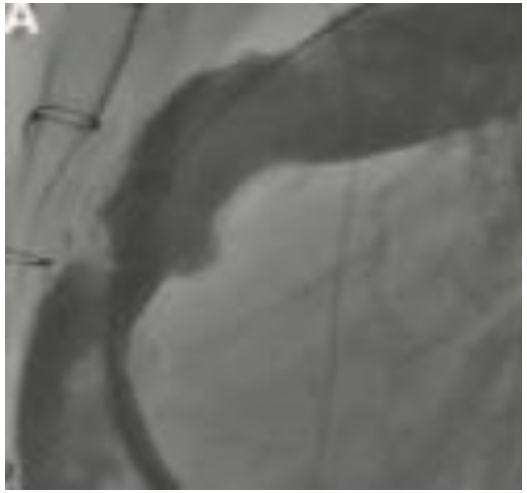
CARDIOPATHIES CONO-TRONCALES À L'ÂGE ADULTE

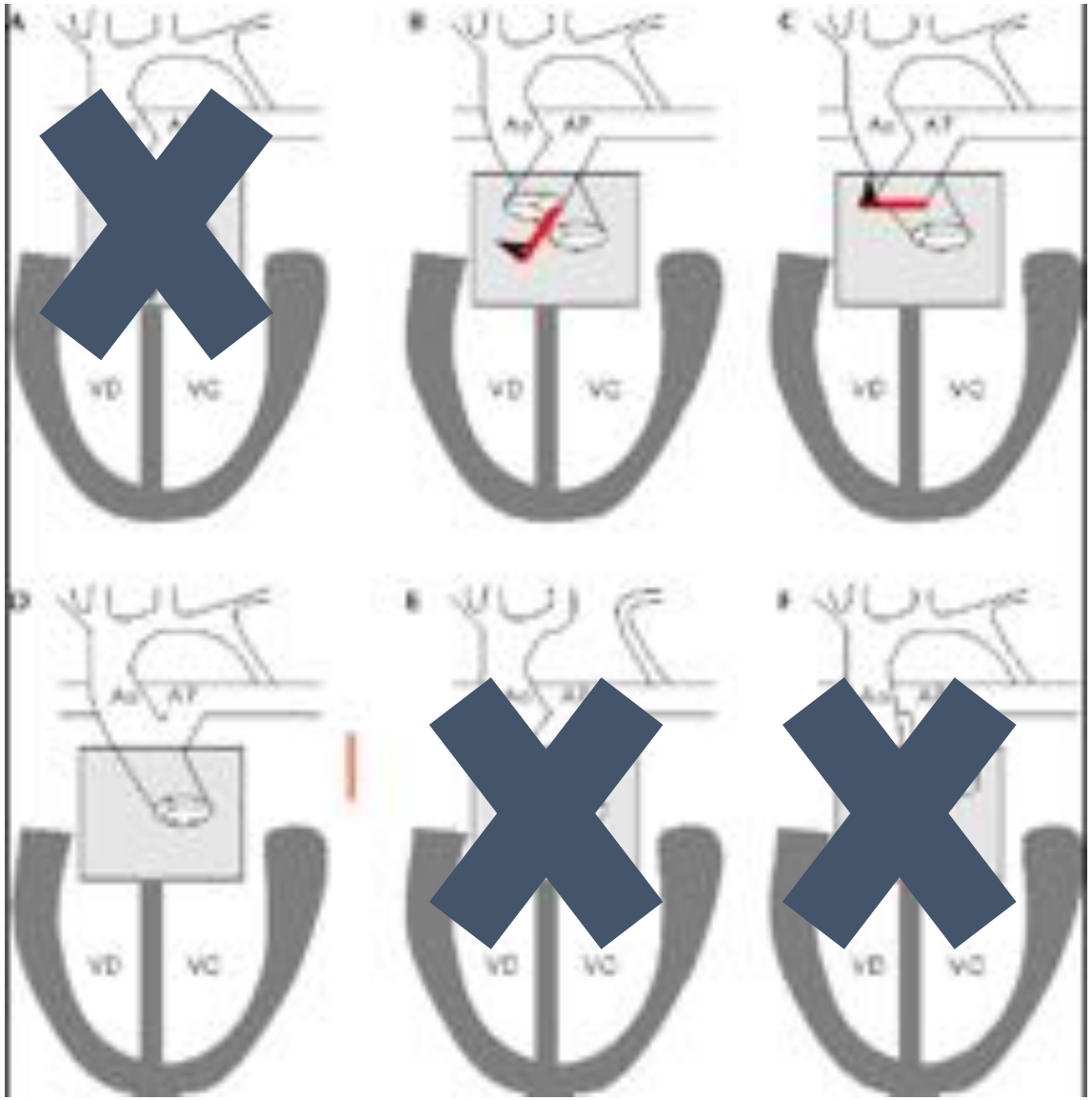




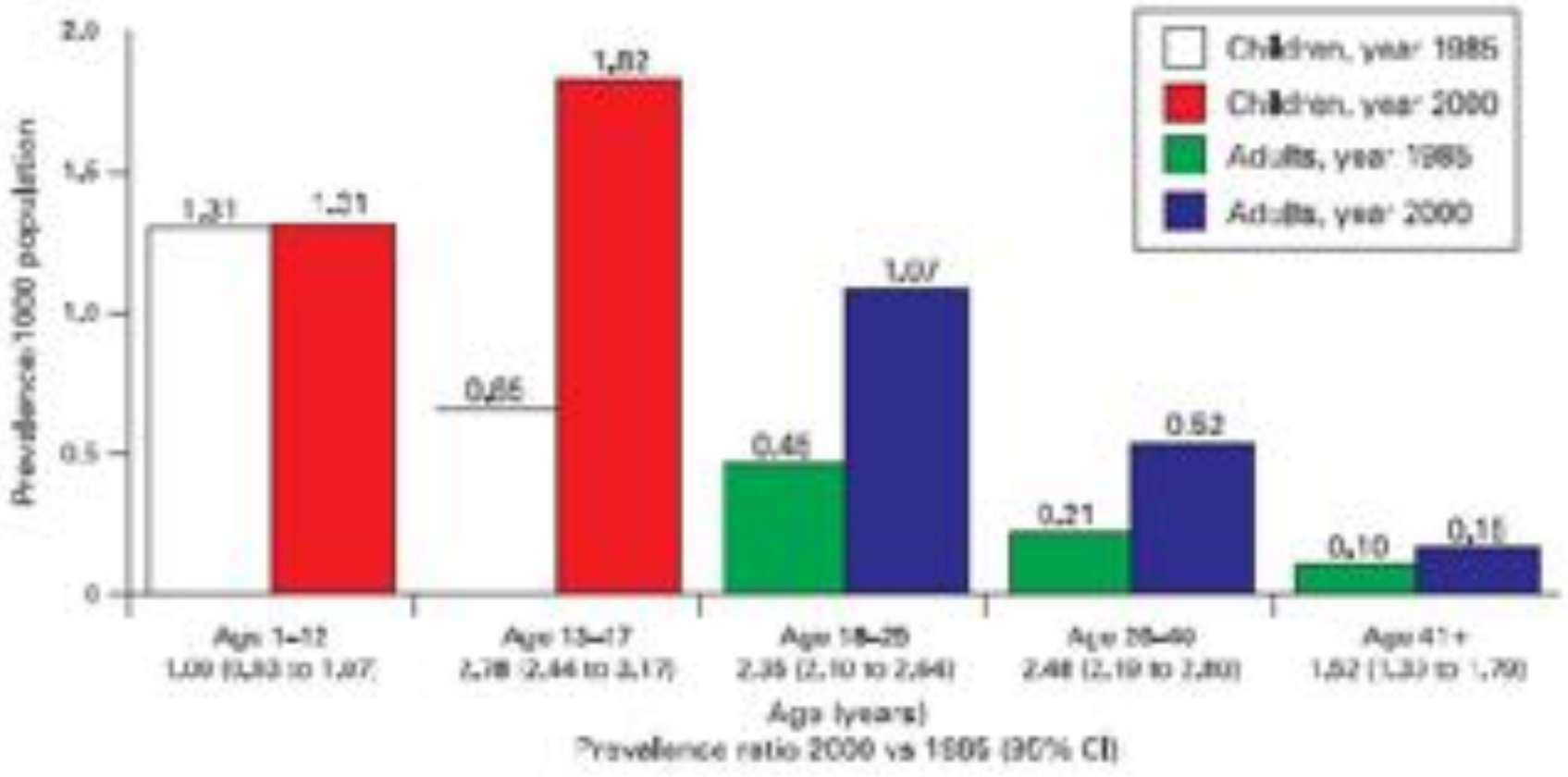
Dr. Zakaria Jalal

Service des cardiopathies congénitales de l'enfant et de l'adulte - Hôpital cardiologique Haut Lévêque - Bordeaux

INTRODUCTION



INTRODUCTION



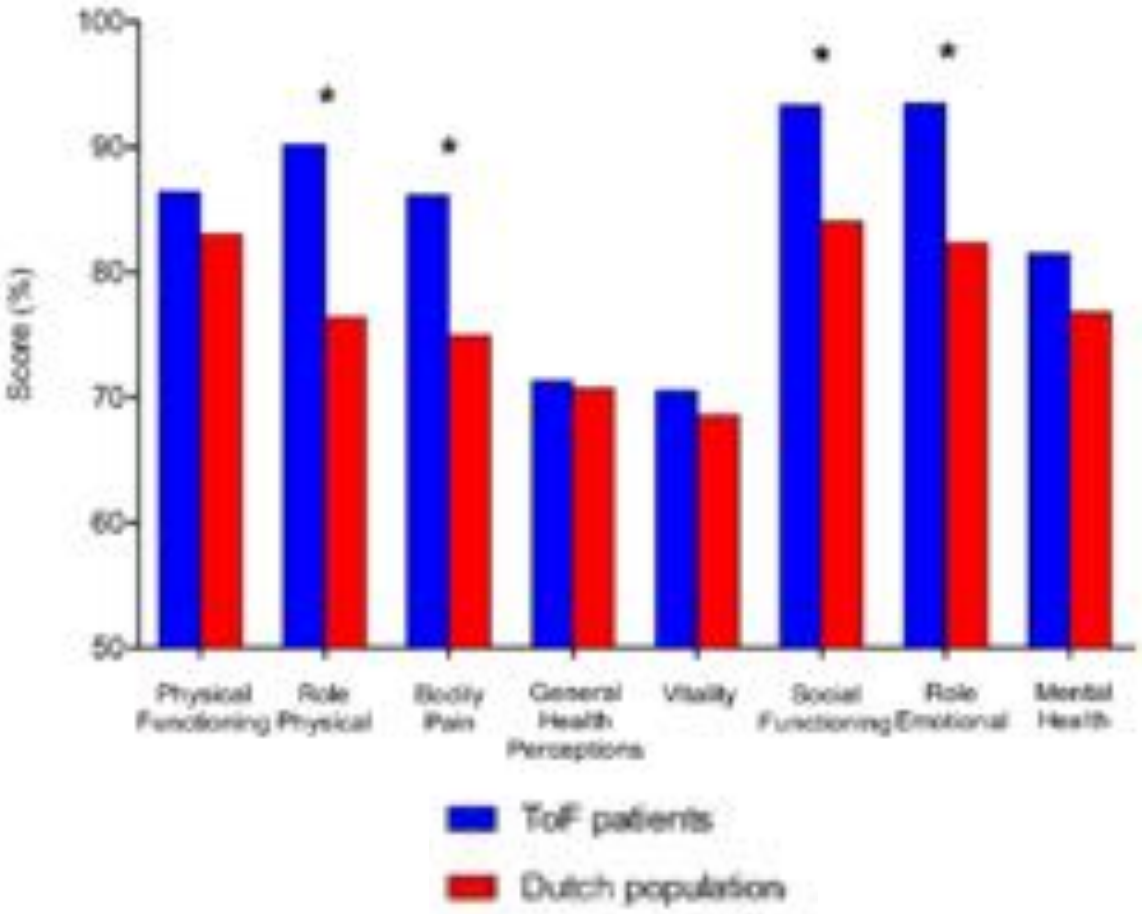
PRÉVALENCE CARDIOPATHIES CONO-TRONCALES

Stable chez les enfants.....En constante augmentation chez les adultes

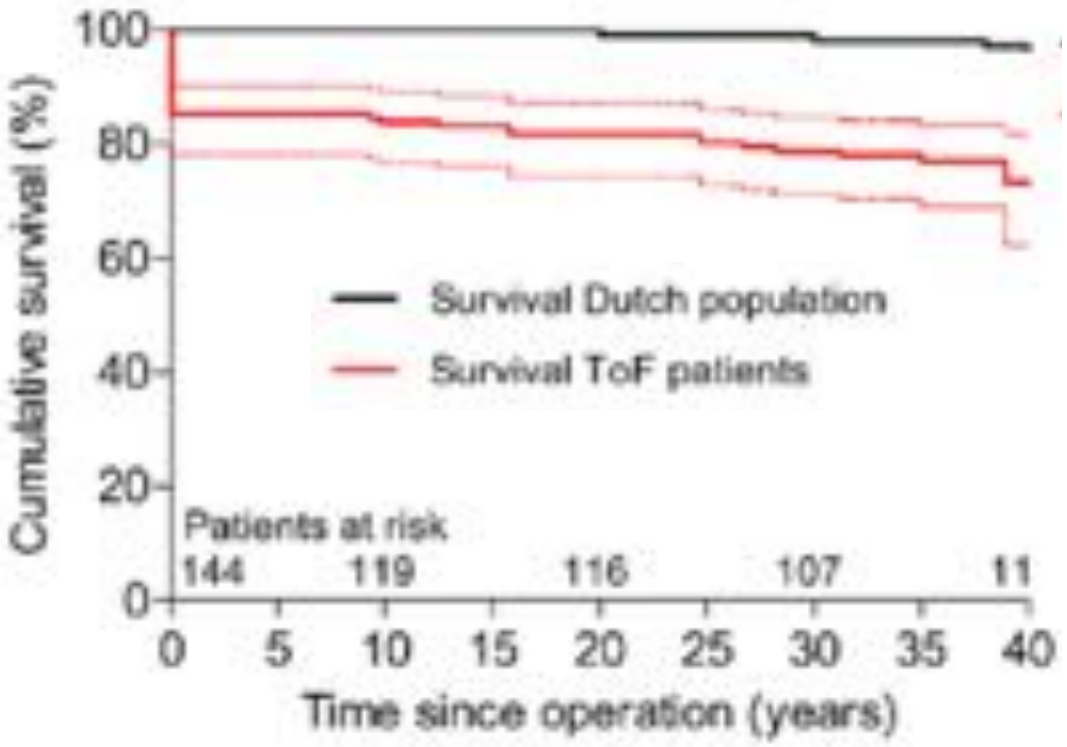
Marelli AJ et al. Circulation 2007

DONNÉES DEMOGRAPHIQUES

QUALITÉ DE VIE



SURVIE



Survie Cumulée: 81% et 72% à 20 et 40 ans,

Incidence Mortalité annualisée: 0.29/100 patient-années

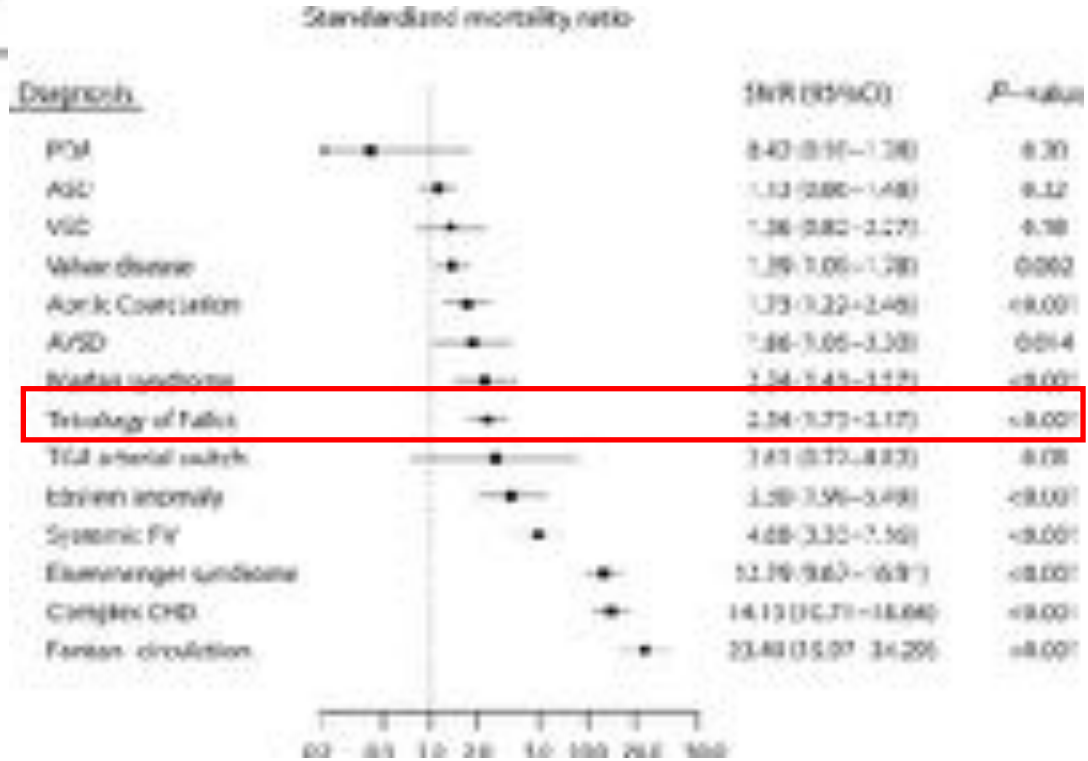
Cuypers et al. Circulation 2015

DONNÉES DEMOGRAPHIQUES

Monocentrique Rétrospectif (LONDON - UK)
6969 pts (1991-2013)
Analyse survie et causes décès

Table 1. Demographics and Baseline Characteristics

Diagnosis	n	Female, %	Deceased, n (%)	Age at Baseline, Years	Age at Death, Years Median [IQR]
ASD	1,080	61.2	66 (6.2)	36.8±18.3	72 [52–79]
PDA	117	77.8	2 (1.7)	32.8±16.4	74 [63–86]
VSD	213	50.6	19 (5.9)	26.1±12.5	48 [29–60]
Marfan syndrome	298	43.6	23 (7.8)	32.0±14.5	49 [33–60]
Valvar disease	1442	44.7	86 (5.9)	20.0±15.5	52 [45–77]
Aortic coarctation	860	41.2	39 (4.6)	28.9±14.3	48 [37–60]
Ebstein	150	54.9	19 (12.6)	34.5±16.2	52 [37–73]
AVSD	256	57.6	15 (5.9)	29.1±14.8	52 [48–63]
Tetralogy of Fallot	980	45.5	54 (6.3)	28.8±13.1	50 [35–66]



Diller et al. Circulation 2017

DONNÉES DEMOGRAPHIQUES

Table 2. Distribution of Causes of Death in Different Diagnostic Subgroups and Overall

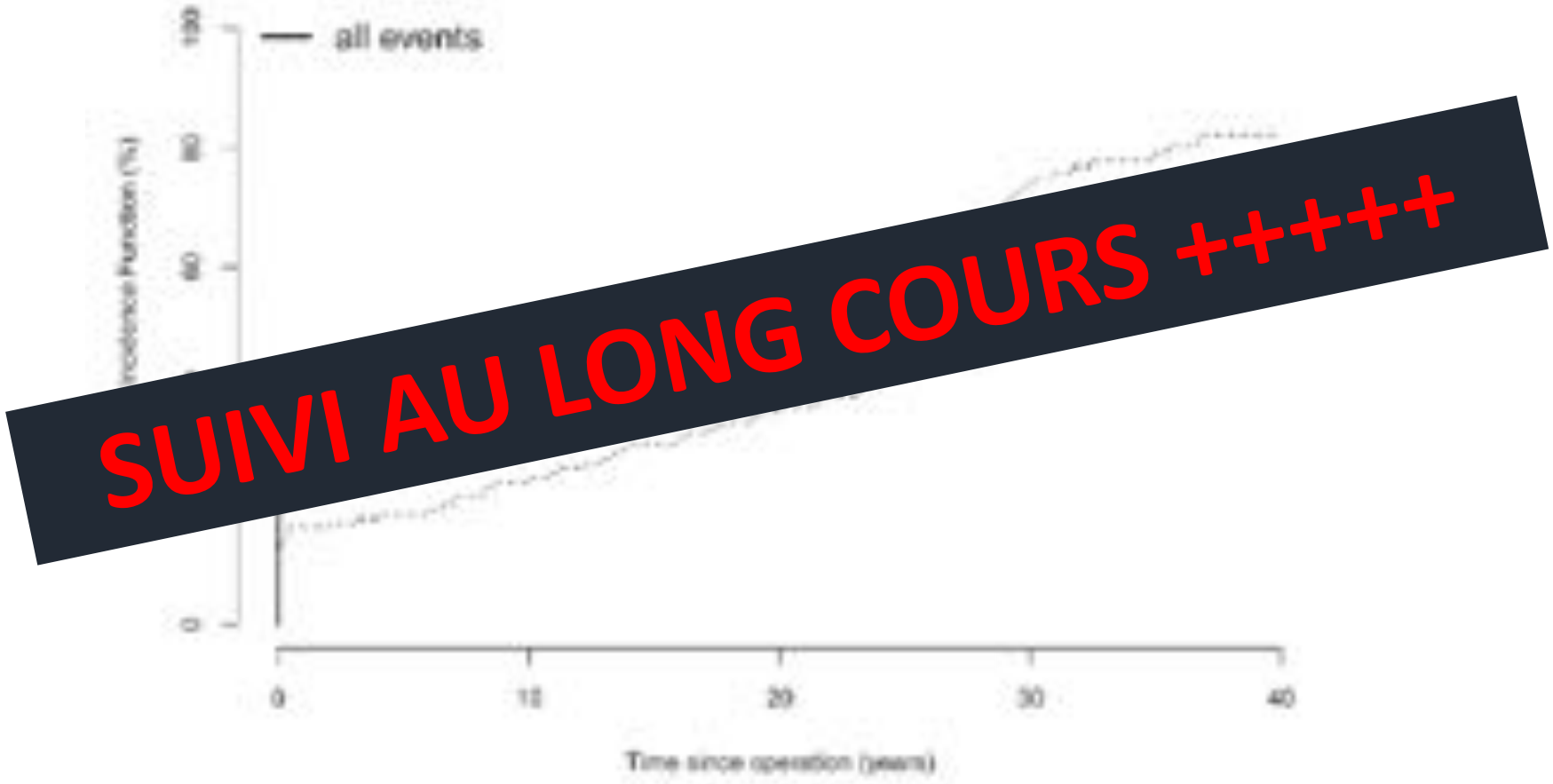
Rank	Cause of death	Aortic coarctation	ASD	AVSD	Complex CHD	Ebstein	Eisenmenger	Fontan	Marfan syndrome	PDA	Systemic RV	Tetralogy of Fallot	TGA arterial switch	Valvular disease	VSD	All patients
1	Heart failure	31%	28%	37%	57%	28%	45%	52%	30%	50%	60%	40%	—	43%	39%	42.5%
2	Pneumonia	8%	17%	7%	2%	—	16%	—	5%	—	—	18%	—	17%	6%	10.2%
3	Sudden cardiac death	—	—	14%	11%	—	6%	13%	5%	—	13%	6%	33%	8%	11%	7.9%
4	Cancer	11%	14%	7%	—	13%	—	3%	25%	50%	—	4%	—	2%	22%	6.3%
5	Cardiac Surgery/ Intervention	11%	2%	7%	5%	15%	2%	19%	5%	—	3%	12%	—	1%	—	6.1%
	(Cardiac surgery, post-op)	(5%)	(1%)	(7%)	(1%)	(17%)	(2%)	(18%)	(5%)	(—)	(3%)	(11%)	(—)	(1%)	(—)	(4.8%)
6	Hemorrhage	8%	5%	—	8%	—	9%	3%	5%	—	—	—	—	2%	11%	5.5%
7	Sepsis/infection	—	3%	—	6%	6%	3%	3%	—	—	—	8%	—	4%	11%	4.5%
8	Cerebrovascular	8%	3%	7%	—	6%	5%	—	—	—	3%	—	—	2%	—	4.1%
9	Acute myocardial infarction	8%	5%	—	3%	—	—	—	—	—	3%	8%	33%	—	—	2.7%
10	Endocarditis	—	—	—	2%	6%	3%	—	10%	—	—	4%	—	1%	—	1.5%
..	Aortic dissection	11%	2%	—	—	—	—	—	5%	—	—	—	—	1%	—	1.4%
..	Hepatic failure	—	3%	—	—	6%	—	3%	—	—	3%	—	—	1%	—	1.4%

Diller et al. Circulation 2017

DONNÉES DEMOGRAPHIQUES

SURVIE CUMULÉE SANS ÉVÉNEMENTS:

50% et 25% à 20 et 40 ans



SUIVI AU LONG COURS +++++

Diller et al. Circulation 2017

RECOMMANDATIONS

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

Follow-up recommendations

All patients with ToF should have periodic cardiac follow-up in a specialized GUCH centre, which in most patients should be done annually, but can be less frequent in those patients at the best end of the spectrum with minimal/stable haemodynamic disturbance. Follow-up evaluation needs to look for the complications listed above (see late clinical presentation). Echocardiography is performed as part of each visit. All patients should have CMR. The intervals for repeat studies depend on the pathology found.

Baumgartner et al. Eur Heart Jour 2010

AHA/ACC GUIDELINE

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

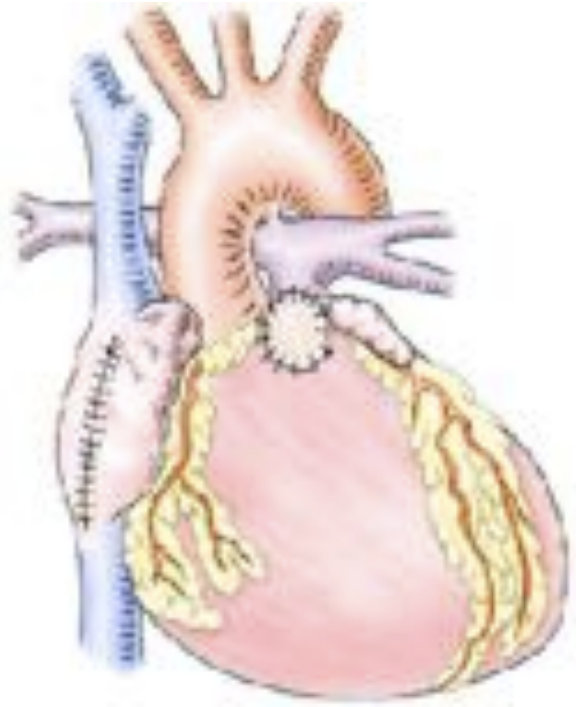
Table 27. TOF: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)
Outpatient ACHD cardiologist	12–24
ECG	24
TTE†	24
Pulse oximetry	As needed
Holter monitor	As needed
CMR‡/CCT§	36
Exercise test‖	36–60

Stout et al. Circulation 2019

COMPLICATIONS AU LONG COURS

- Dysfonction ventriculaire droite
- Fuite/Sténose voie pulmonaire (+/- prothétique)
- Sténoses des branches pulmonaires
- Dysfonction ventriculaire gauche
- Valvulopathie aortique
- Dilatation de l'aorte ascendante
- CIV résiduelles
- Arythmies
- Endocardite infectieuse



CAUSES INTRIQUÉES

CARDIOPATHIE INITIALE

CYANOSE PRÉ-OPÉRATOIRE

CEC

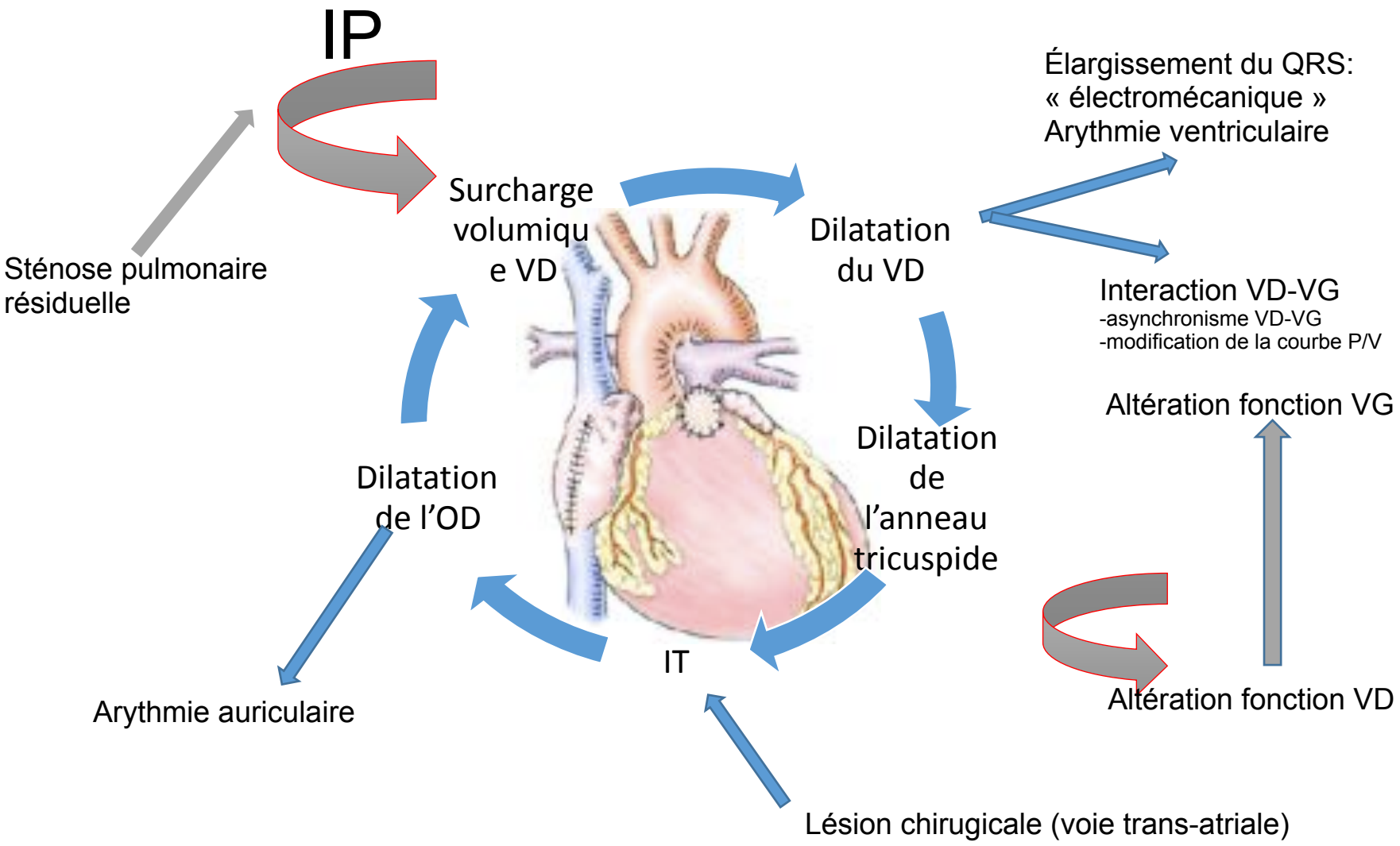
CICATRICES CHIRURGICALES

REMODELAGE TISSULAIRE

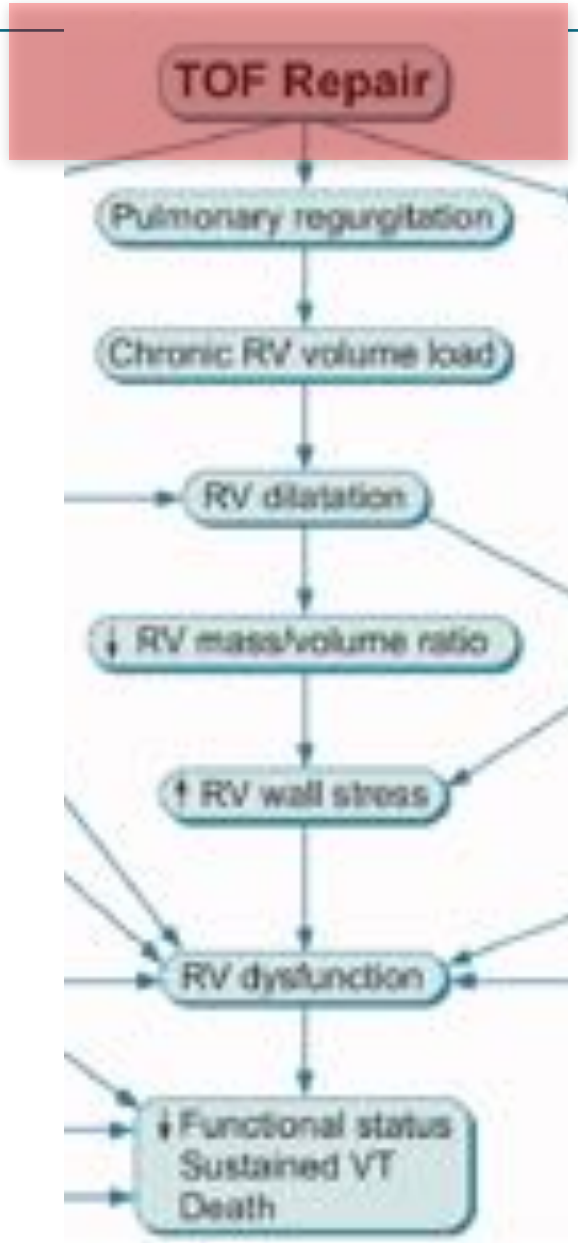
DURÉE DE VIE DES SUBSTITUTS

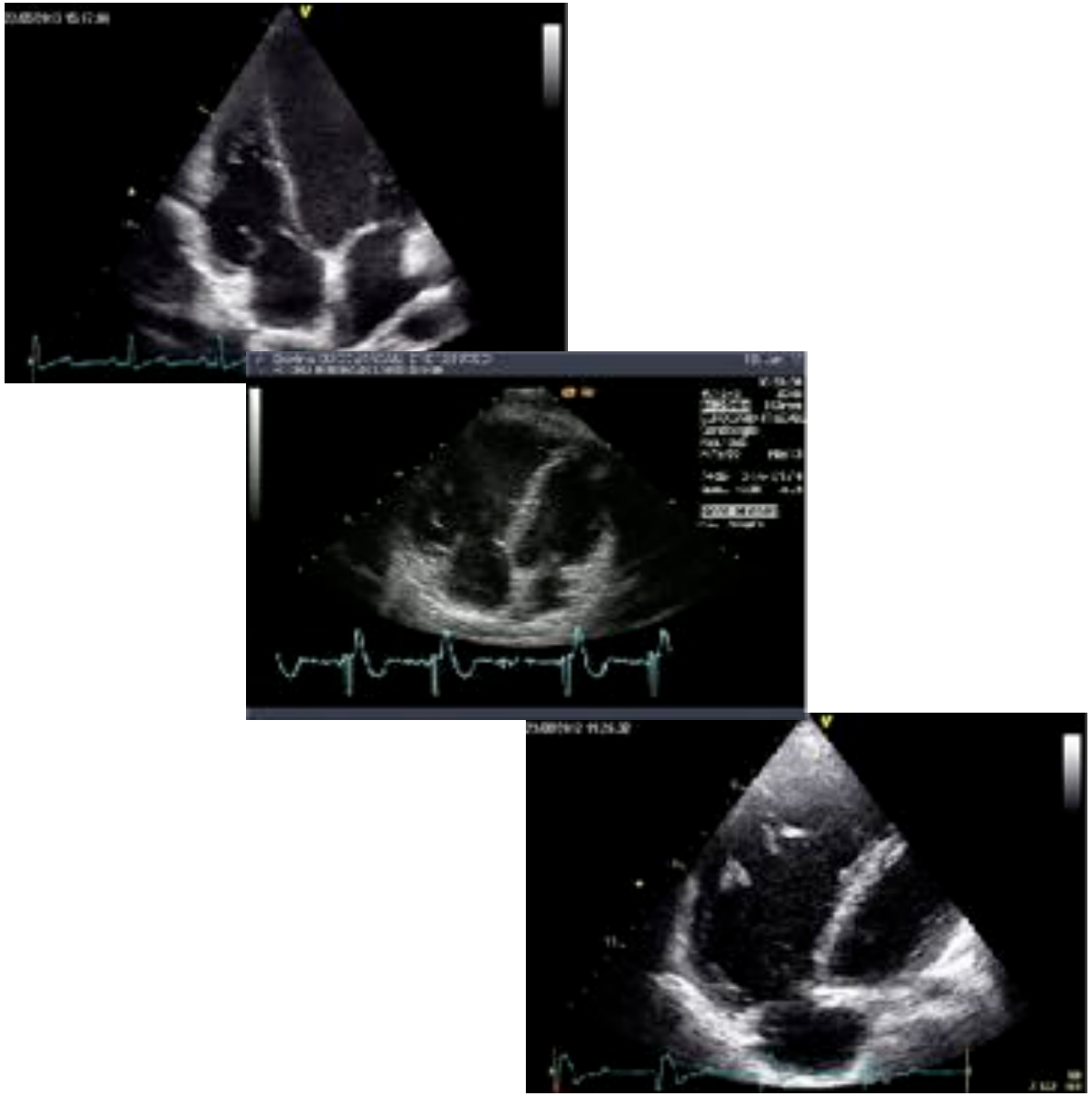
...

VALVULOPATHIE PULMONAIRE



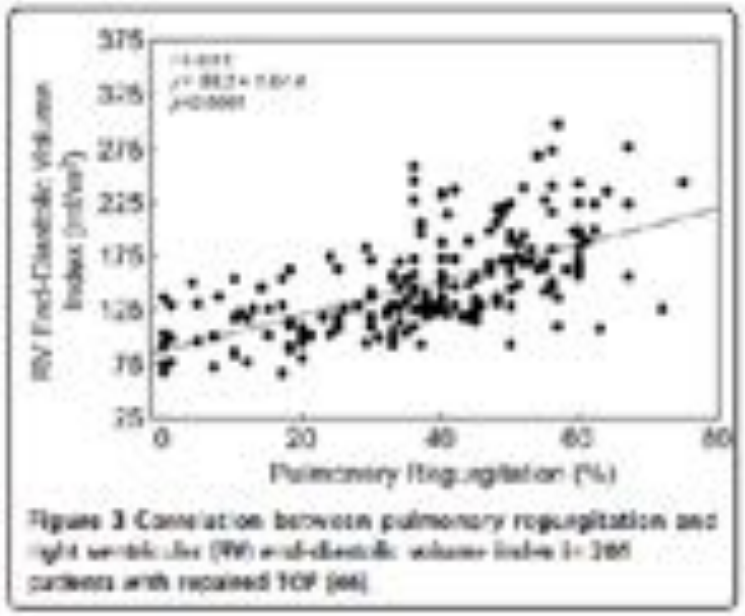
VALVULOPATHIE PULMONAIRE

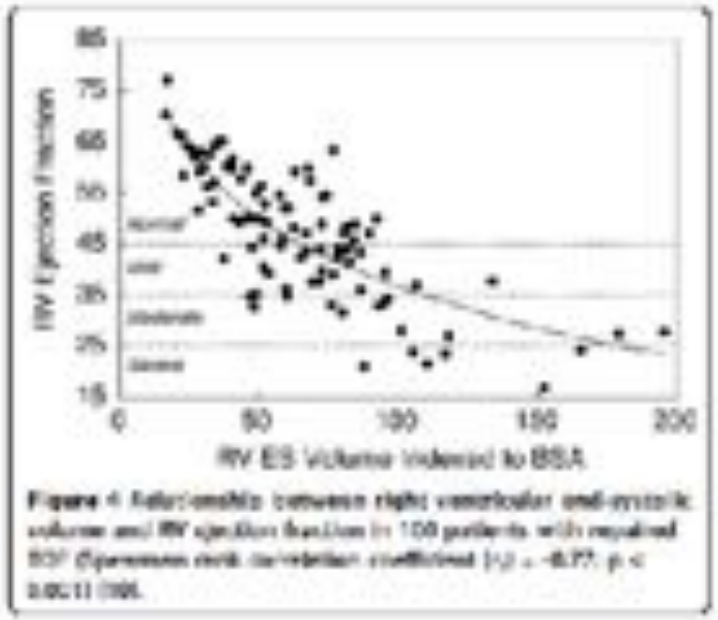


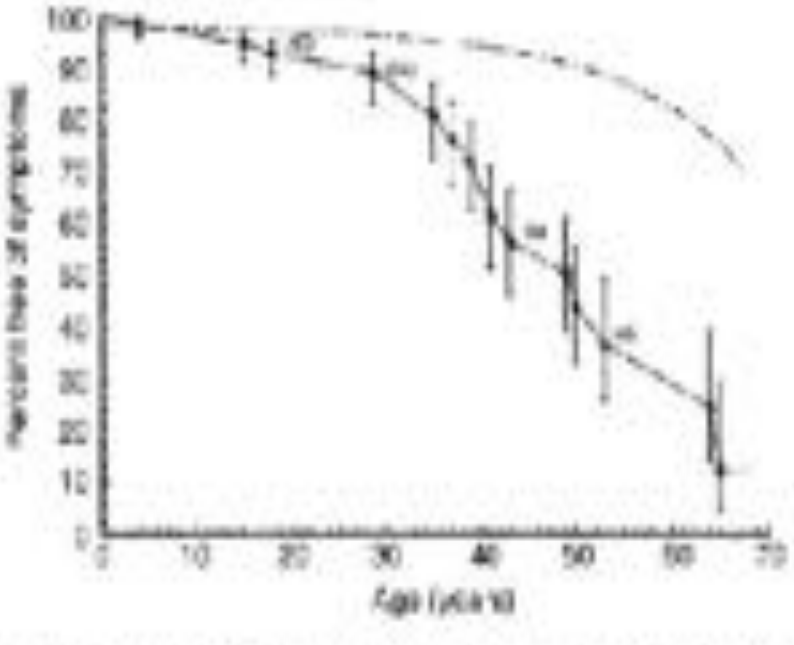


VALVULOPATHIE PULMONAIRE

- PHYSIOPATHOLOGIE DIFFÉRENTE DE L'IAO
- PATIENTS LONGTEMPS ASYMPTOMATIQUES
- PARAMETRES FONCTIONNELS CONSERVÉS
- MAIS DILATATION PROGRESSIVE DU VD +/- REVERSIBLE

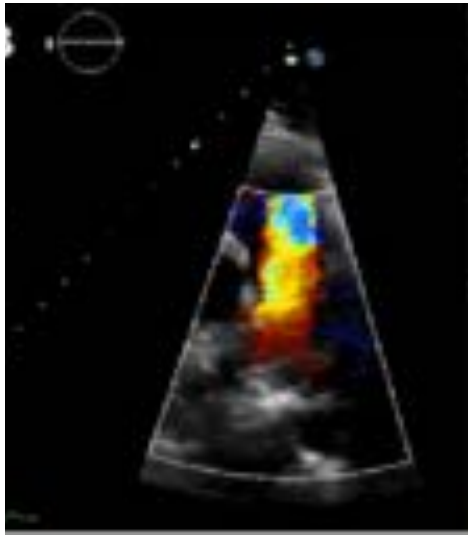


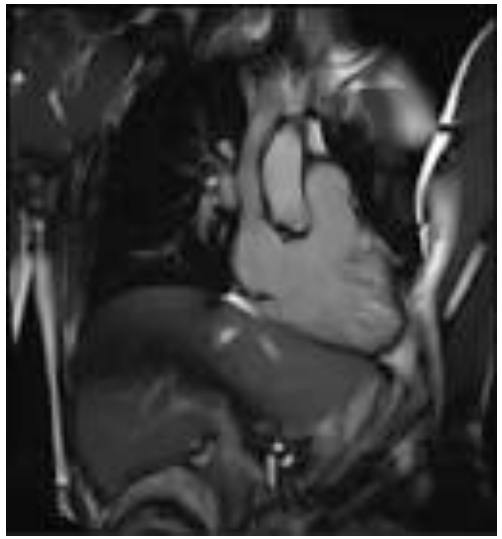




Shimazaki Y. Thorac cardiovasc Surgeon 1984
Samyn et al. J Magn Reson Imaging 2007
Geva et al. J Am Coll Cardiol 2004

VALVULOPATHIE PULMONAIRE





ÉVALUTATION MULTI-MODALE

ETT – IRM – TDM – CATHÉTÉRISME

OBJECTIFS:

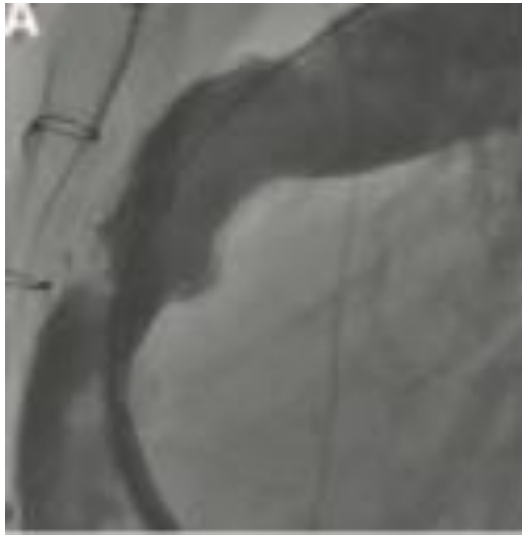
Type et sévérité de la lésion valvulaire

Retentissement VD (taille / fonction)

Retentissement VG (taille / fonction)

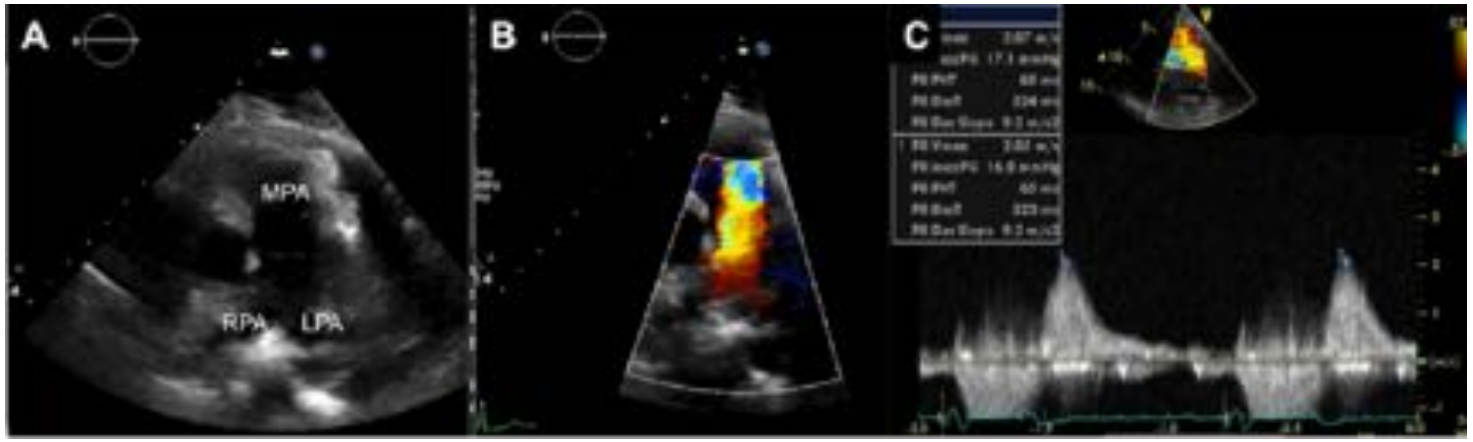
Lésions associées

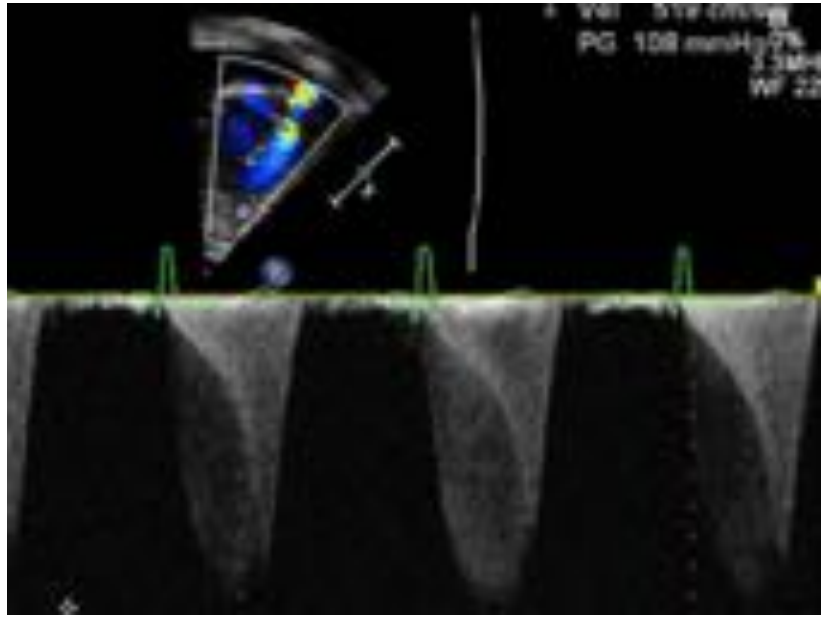
INDICATION THÉRAPEUTIQUE



VALVULOPATHIE PULMONAIRE

ÉVALUTATION ECHOGRAPHIQUE





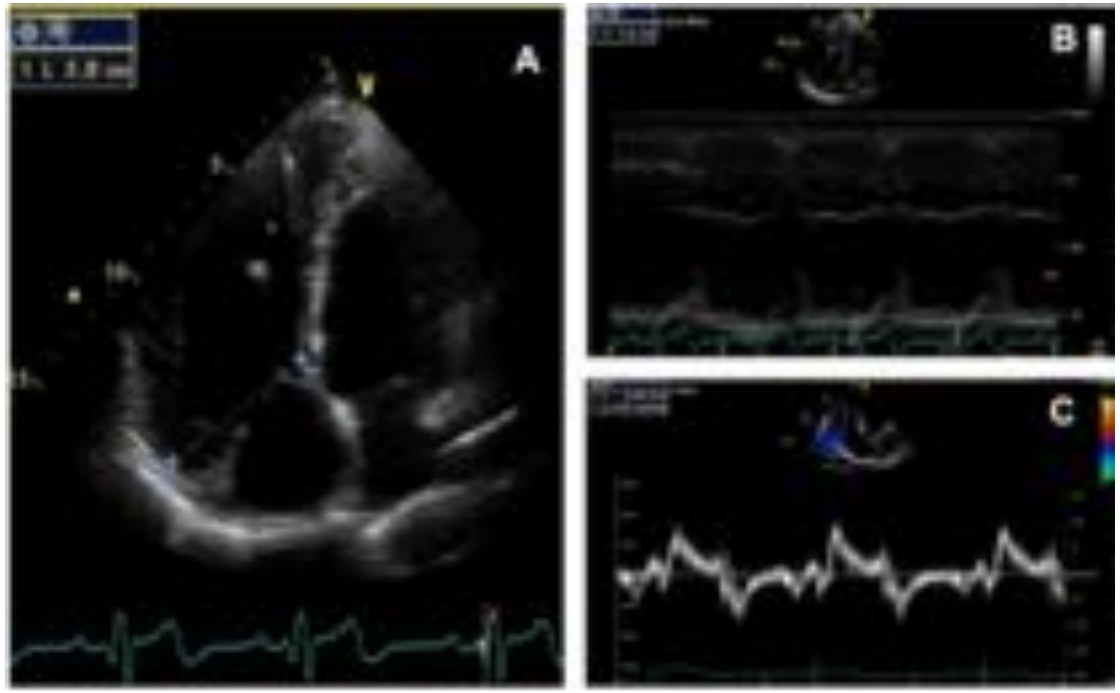
TYPE DE LÉSION VALVULAIRE

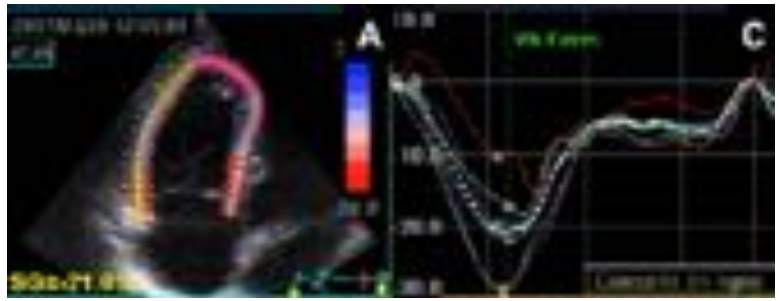
SIÈGE

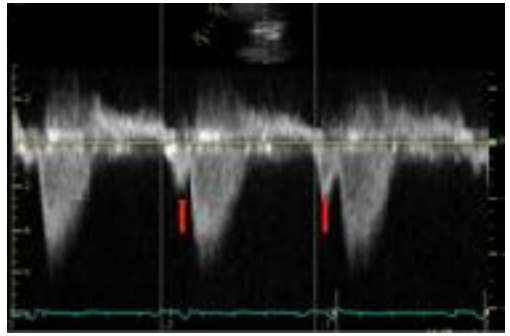
SÉVÉRITÉ

Parameter	Abnormality threshold
Pulmonary regurgitation PHT	<100 ms
Jet:Annulus width ratio	0.5
Diastolic flow reversal from the branch pulmonary arteries	From branch PAs
Termination of flow in mid-late diastole	No flow time >80 ms.

ÉVALUTATION ECHOGRAPHIQUE







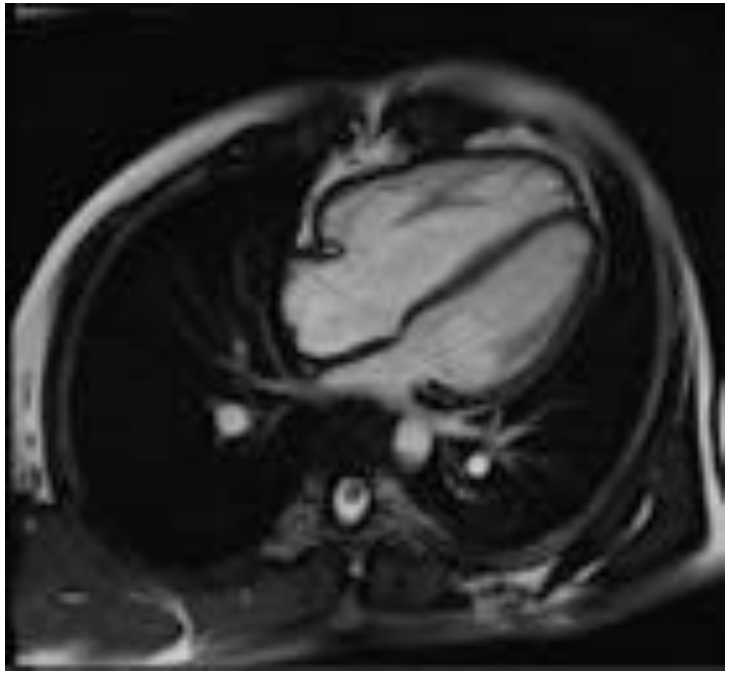
RETENTISSEMENT VENTRICULAIRE

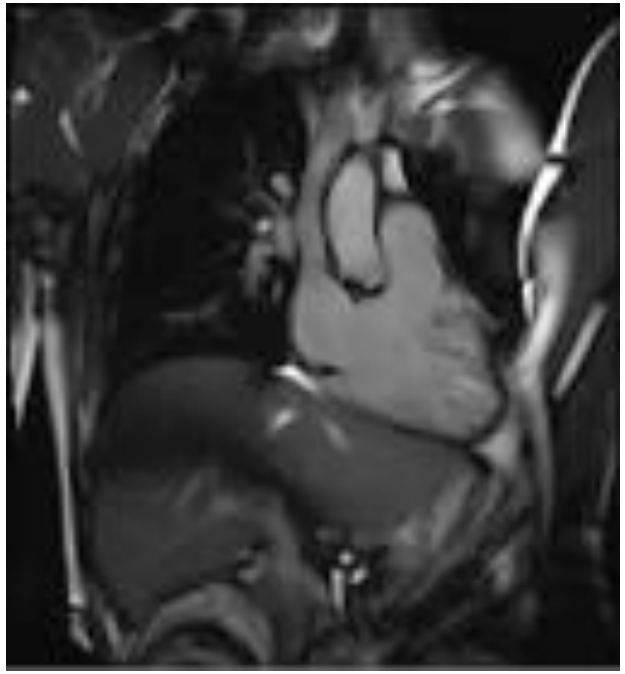
Table 5 Normal and abnormal valves for measures of RV function in adult patients (ASE/ESC recommendations) (13).

Parameter	Abnormality threshold
TAPSE (mm)	<17
RV FAC (%)	<35
RV free wall 2D strain (%)	≥−20
RV 3D EF (%)	<45
Pulsed Doppler S wave (cm/s)	<9.5

VALVULOPATHIE PULMONAIRE

ÉVALUTATION IRM





TYPE DE LÉSION VALVULAIRE

SIÈGE

SÉVÉRITÉ

VOLUMÉTRIE VENTRICULAIRE

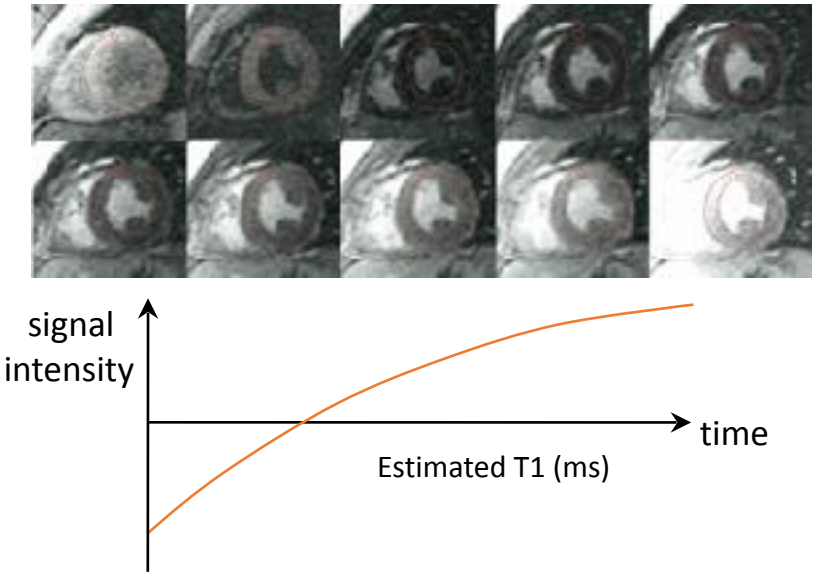
FONCTION VENTRICULAIRE

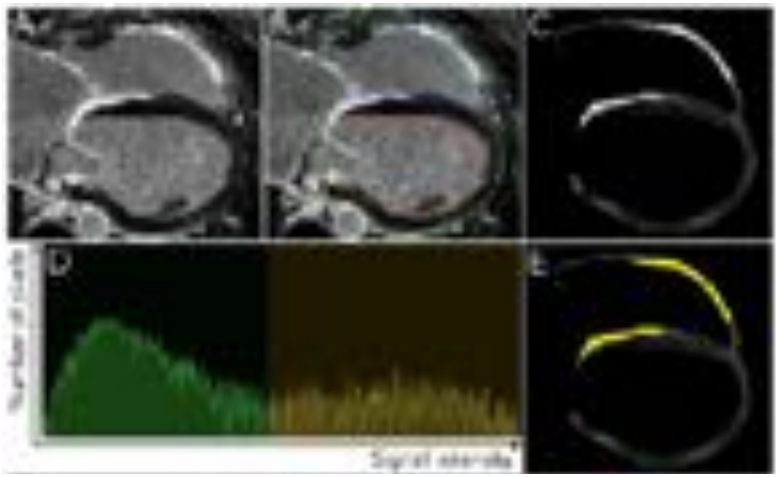
FIBROSE

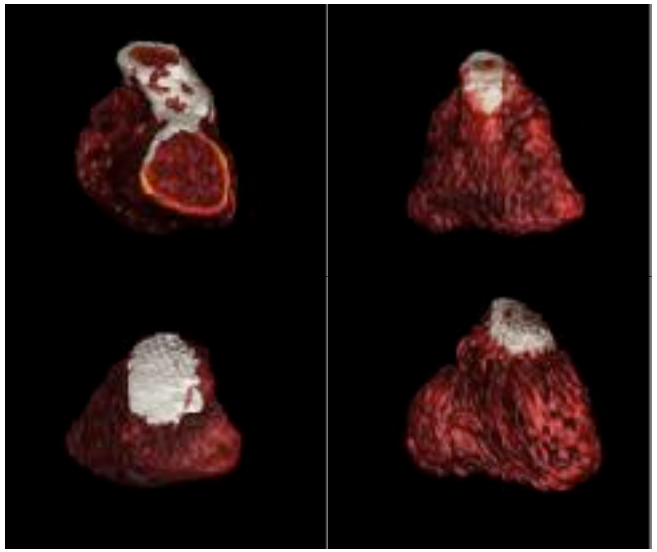
EVOLUTIVITÉ DES PARAMÈTRES

ÉVALUTATION IRM

QUANTIFICATION DE LA FIBROSE



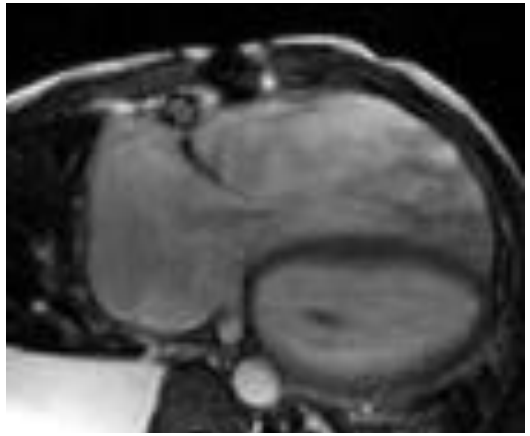




Chen C et al. JACC imaging 2016
Broberg CS et al. JACC imaging 2016
Cochet et al. Eur Heart JCVI 2019

VALVULOPATHIE PULMONAIRE

PULMONARY VALVE DYFUNCTION => THERAPEUTIC TARGET



IRREVESIBLE RV/LV DYSFUNCTION

SYMPTOMS OCCURENCE

INTERVENTION RISKS

PROSTHESIS DEGENERATION

REINTERVENTIONS

VALVULOPATHIE PULMONAIRE

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

Table 15 Indications for intervention after repair of tetralogy of Fallot

Indications	Class	Level
Aortic valve replacement should be performed in patients with severe AR with symptoms or signs of LV dysfunction	I	C
PVRep should be performed in symptomatic patients with severe PR and/or stenosis (RV systolic pressure >60 mmHg, TR velocity >3.5 m/s)	I	C
PVRep should be considered in asymptomatic patients with severe PR and/or PS when at least one of the following criteria is present: • Decrease in objective exercise capacity • Progressive RV dilation • Progressive RV systolic dysfunction • Progressive TR (at least moderate) • RVOTO with RV systolic pressure >80 mmHg (TR velocity >4.3 m/s) • Sustained atrial/ventricular arrhythmias	IIa	C

Baumgartner et al. Eur Heart Jour 2010

AHA/ACC GUIDELINE

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

Recommendations for TOF (Continued)

COR	LOE	Recommendations
Therapeutic		
I	B-NR	5. Pulmonary valve replacement (surgical or percutaneous) for relief of symptoms is recommended for patients with repaired TOF and moderate or greater PR with cardiovascular symptoms not otherwise explained.
IIa	B-NR	6. Pulmonary valve replacement (surgical or percutaneous) is reasonable for preservation of ventricular size and function in asymptomatic patients with repaired TOF and ventricular enlargement or dysfunction and moderate or greater PR.

Stout et al. Circulation 2019

QUAND VALVER UN PATIENT ASYMPTOMATIQUE?

Table 1
Summary of published guidelines for surgical PVR in the asymptomatic patient.

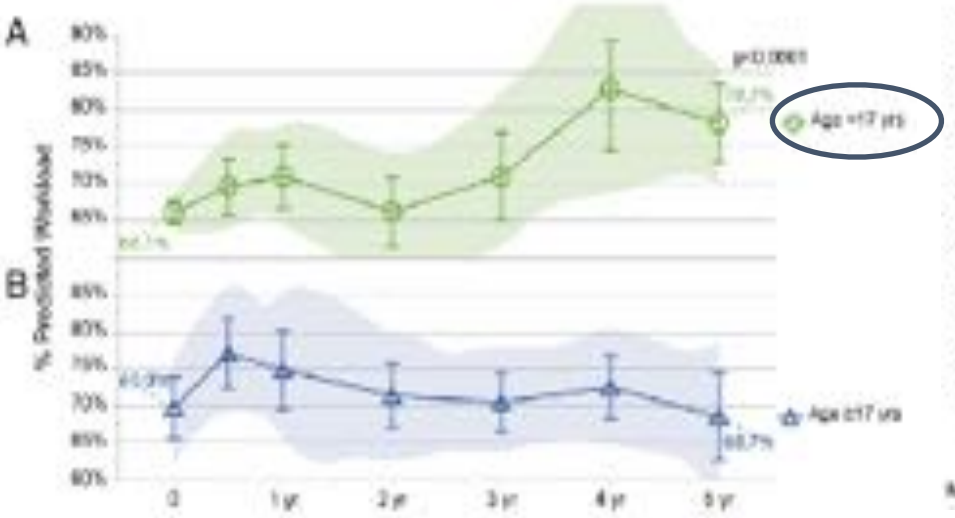
Criteria	AHA [13]	ESC [11]	CCS [12]	Geva [6]
RVEDVi	≥ Moderate	> 160 mL/m²	> 170 mL/m²	> 150 mL/m² or Z-score > 4 or RV:LV end-diastolic volume ratio > 2
RVESVi	Not specified	Not specified	Not specified	> 80 mL/m²
RV function	≥ Moderate RV dysfunction	Progressive RV dysfunction	≥ Moderate RV dysfunction	RV EF < 47%
RVOT obstruction*	PIG ≥ 50 mm Hg or RV:LV pressure ratio ≥ 0.7	PIG ≥ 80 mm Hg (4.3 m/s)	RV systolic pressure ≥ 2/3 systemic pressure	RV systolic pressure ≥ 2/3 systemic pressure
PR*	Severe	Severe	Free	≥ Moderate (PRF ≥ 25%)
TR	≥ Moderate	≥ Moderate	"Important"	≥ Moderate
QRS duration	Not specified	> 180 msec	Not specified	> 140 msec
Arrhythmia	Symptomatic or sustained AT or VT	Sustained AT or VT	AT or VT	Sustained tachyarrhythmia
Exercise cardiopulmonary function	Not specified	Objective decrease	Not specified	Not specified
Other considerations	Significant residual VSD or AR	Not specified	Significant residual VSD	LV EF < 55%, large RVOT aneurysm, severe branch pulmonary artery stenosis (< 30% flow to affected lung), significant residual left-to-right shunt, severe AR or aortic dilation

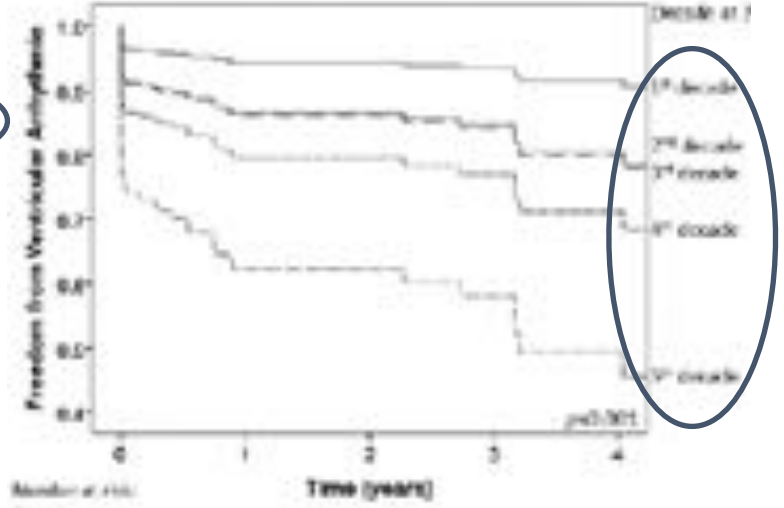
Tretter et al. Int Jour Cardiol 2016

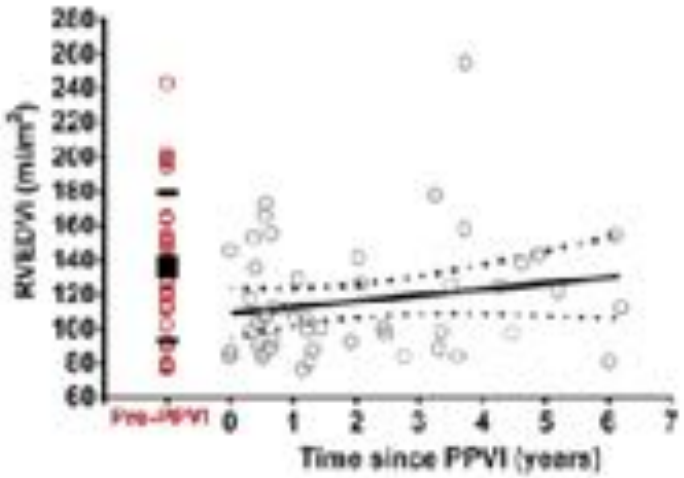
VALVULOPATHIE PULMONAIRE

Percutaneous Pulmonary Valve Implantation

Is Earlier Valve Implantation Better?







EXERCISE CAPACITY

ARRHYTHMIA BURDEN

VENTRICULAR SIZE AND FUNCTION

Priromprintr et al. Am Heart J 2019

Wadia et al. Heart 2019

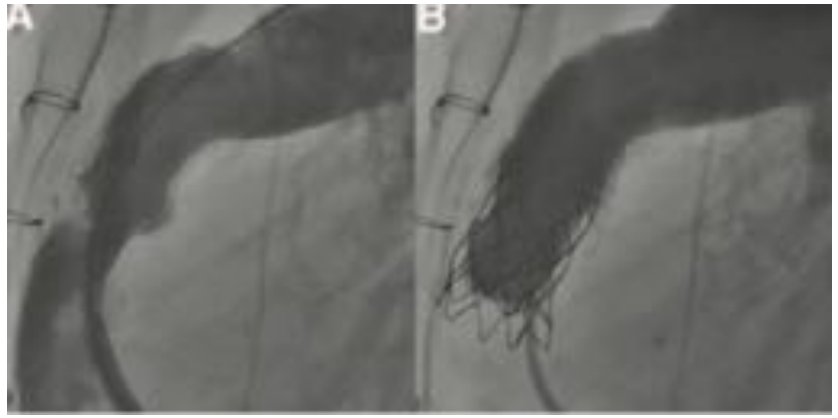
Borik et al. Circ Cardiovasc Interv. 2015

VALVULOPATHIE PULMONAIRE

COMMENT VALVER?

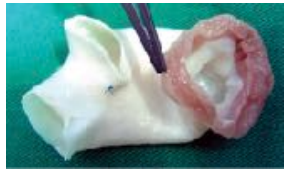
CHIRURGIE

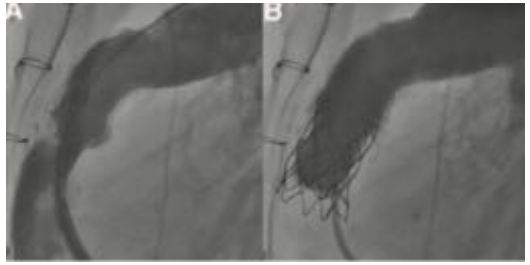
CATHÉTÉRISME



VALVULOPATHIE PULMONAIRE

COMMENT VALVER?





CHIRURGIE

AVANTAGES

- Possible quelque soit taille RVOT
- Lésions associées

INCONVÉNIENTS:

- Sternotomies répétées
- Risques CEC
- Durabilité
- Risque infectieux

CATHÉTÉRISME

AVANTAGES

- Moins invasif
- Valve-in-valve

INCONVÉNIENTS:

- Taille voie pulmonaire
- Complications liées à la technique
- Durabilité
- Risque infectieux

VALVULOPATHIE PULMONAIRE

Implantation chez l'homme

Faisabilité

Succès technique

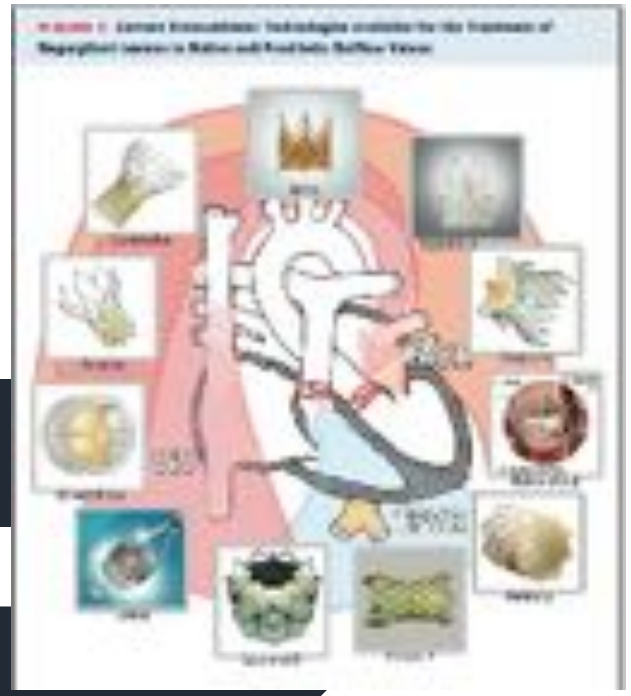
Excellent résultats hémodynamiques

Peu de complications aigues

2000

Marquage CE
FDA Approval

2008-2010



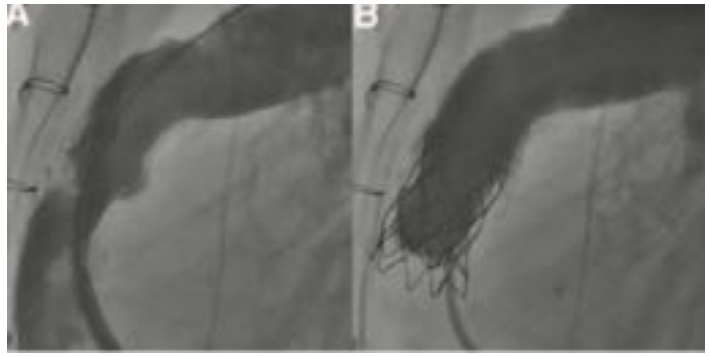
MELODY - Veine jugulaire de bœuf
16 – 22 mm ⦸

SAPIEN XT – Péricarde bovin
(20) – 23 – 26 – 29 mm ⦸

VALVULOPATHIE PULMONAIRE

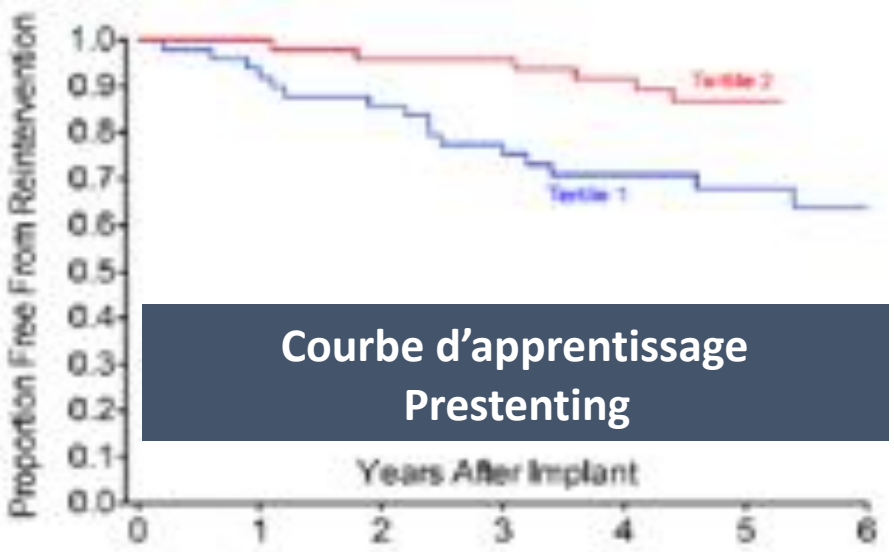
DÉVELOPPEMENT CROISSANT DE LA VALVULATION PERCUTANÉE

- Efficacité équivalente pour une approche moins invasive
- Indications élargies, limites techniques repoussées
- Futur radieux: nouveaux dispositifs, nouvelles technologies



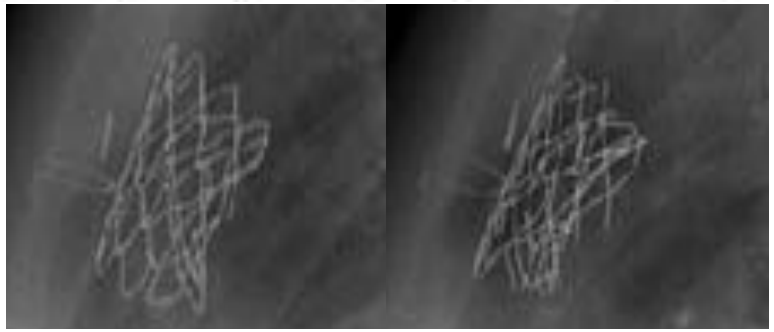
RESULTATS MELODY

- 150 pts, 2007 – 2010, multicentrique
- Succès 95%
- 2 décès per-procédure (rupture, dissection coronaire)
- Suivi médian 4.9 ans (0.4 – 7)



Courbe d'apprentissage Prestenting

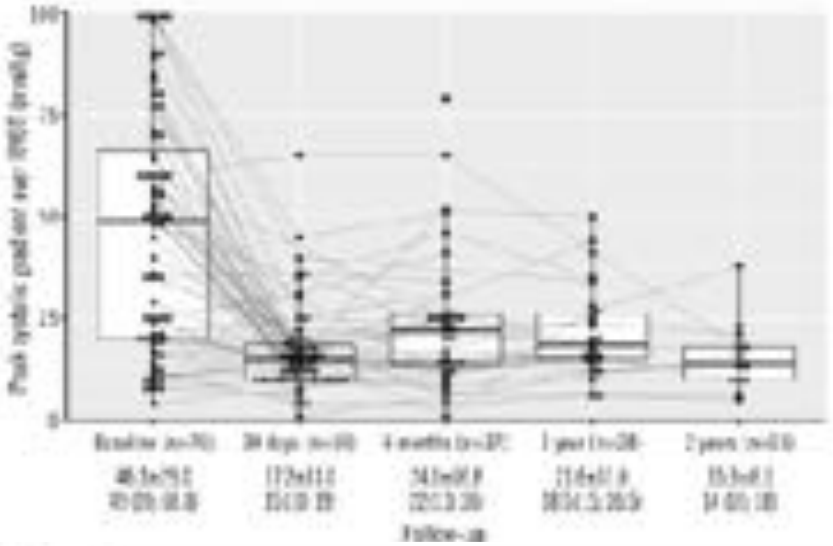
- 4 décès (1 EI, 3 non cardiaque)
- 14 bactériémie / endocardites
- 113 patients sans réintervention:
  - Grdt inchangé (15 vs 17 mmHg)
  - Fuite triviale ou absente: 112/113

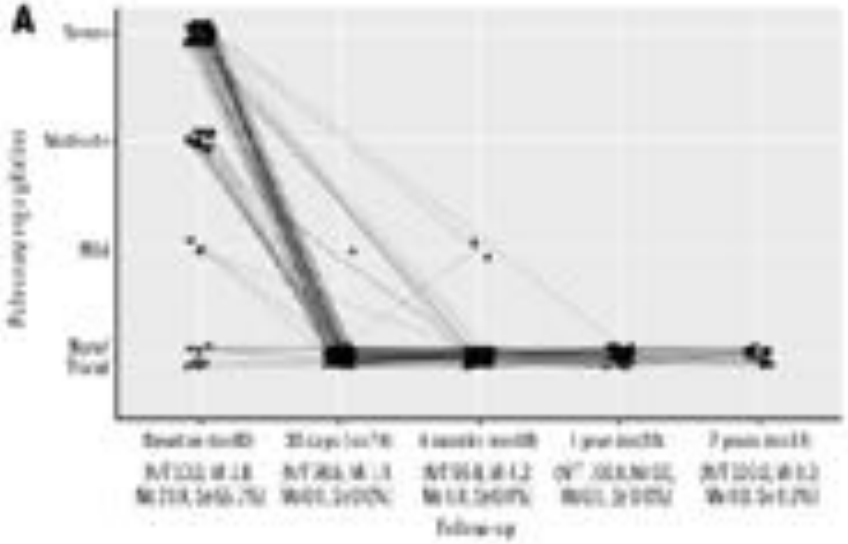


Survie sans réintervention à 5 ans
76%
↓
91% en excluant fracture de stents

Cheatham et al. Circulation 2015

VALVULOPATHIE PULMONAIRE

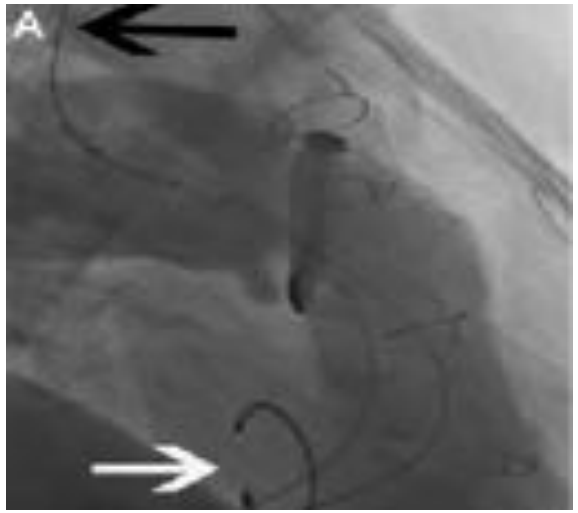




Hascoet et al. Eurointervention 2018

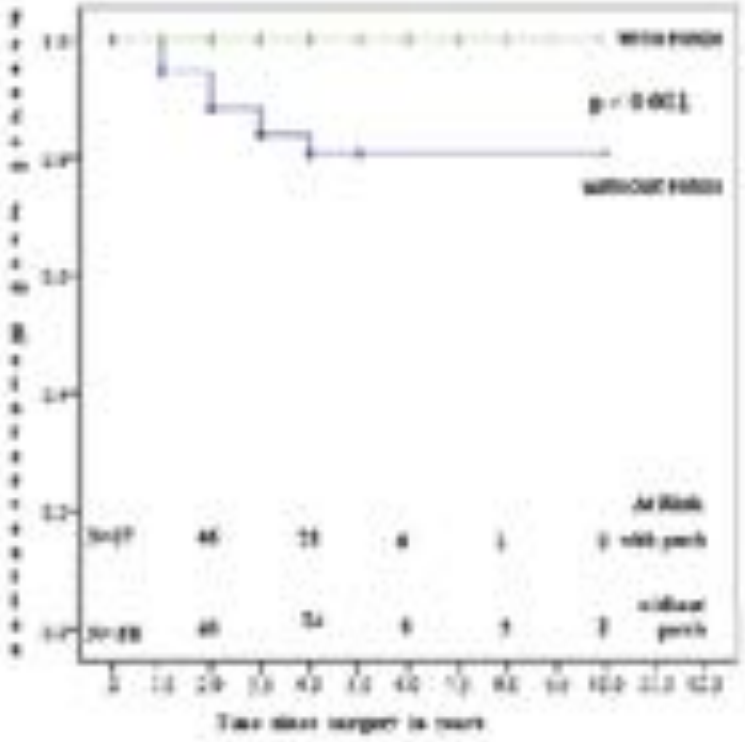
RESULTATS SAPIEN

- 82 pts, 2016 - 2018, registre multicentrique (Europe, Canada)
- 81/82 pts implantés avec succès
- 2 complications majeures (embolisation, effraction RVOT)
- Suivi médian 201 jours (IQR 69 – 365)
- 0 décès, 0 fracture de stent, 0 endocardite
- 2 thromboses de valve

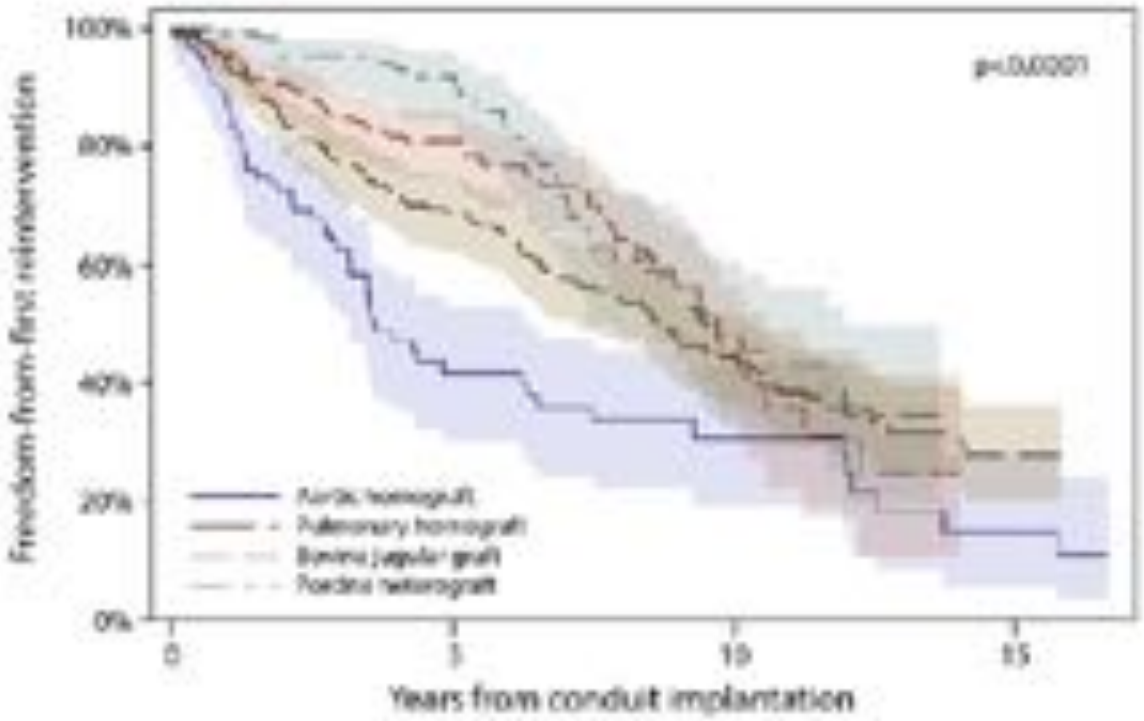


VALVULOPATHIE PULMONAIRE

SUBSTRATS CHIRURGICAUX



115 patients
Survie sans réintervention:
87% à 5 ans

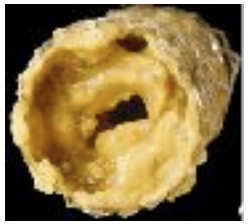


586 patients
Survie sans réintervention:
73% à 5 ans

Ramanan et al. Ann Thorac Surg 2015
Mery et al. JTCS 2016

VALVULOPATHIE PULMONAIRE

ENDOCARDITE INFECTIEUSE



Valve Melody : **0.8 – 3% pt-année**

Valves/conduits pulmonaire chir : **0.5 - 3% pt-année**

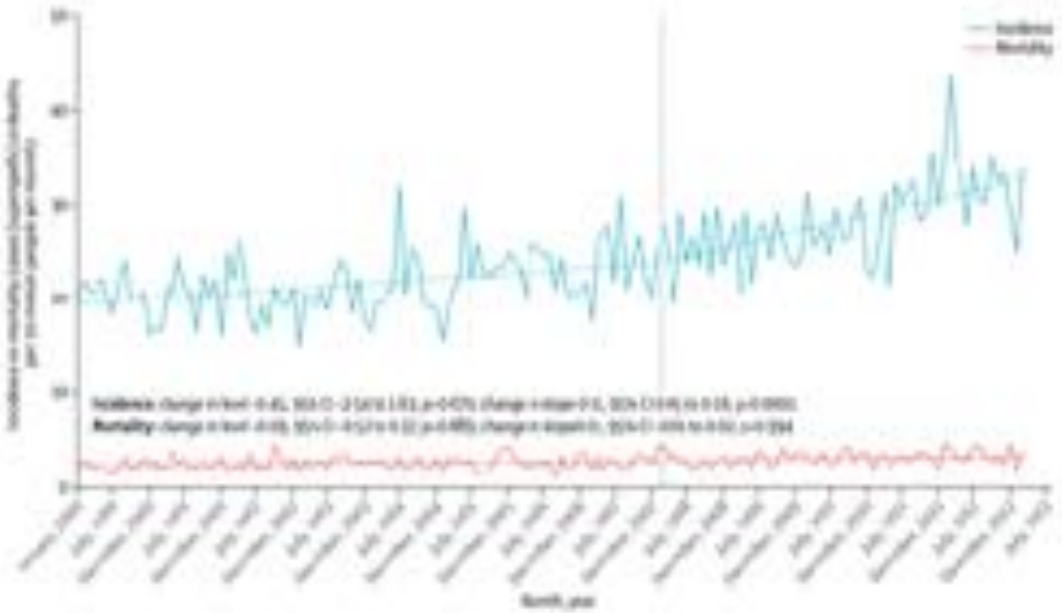
TAVI: **0.67 – 2.1% pt-année**

Valves Ao/mitrale chir : **0.3 – 1.2% pt-année**

Dispositifs electroniques implantables : **1.9/1000 device-année**

Patients avec CC**: 0.4 – 1.33 / 1000 pt-année**

Population générale : **30 -100/ million pt-année**



Miranda et al. Eur Heart Jour 2016
Wang et al. JAMA 2007
Rushani et al. Circulation 2013
Habib et al. Eur Heart Jour 2015
Dayer et al. Lancet 2015

VALVULOPATHIE PULMONAIRE

ENDOCARDITES DE LA VOIE DROITE

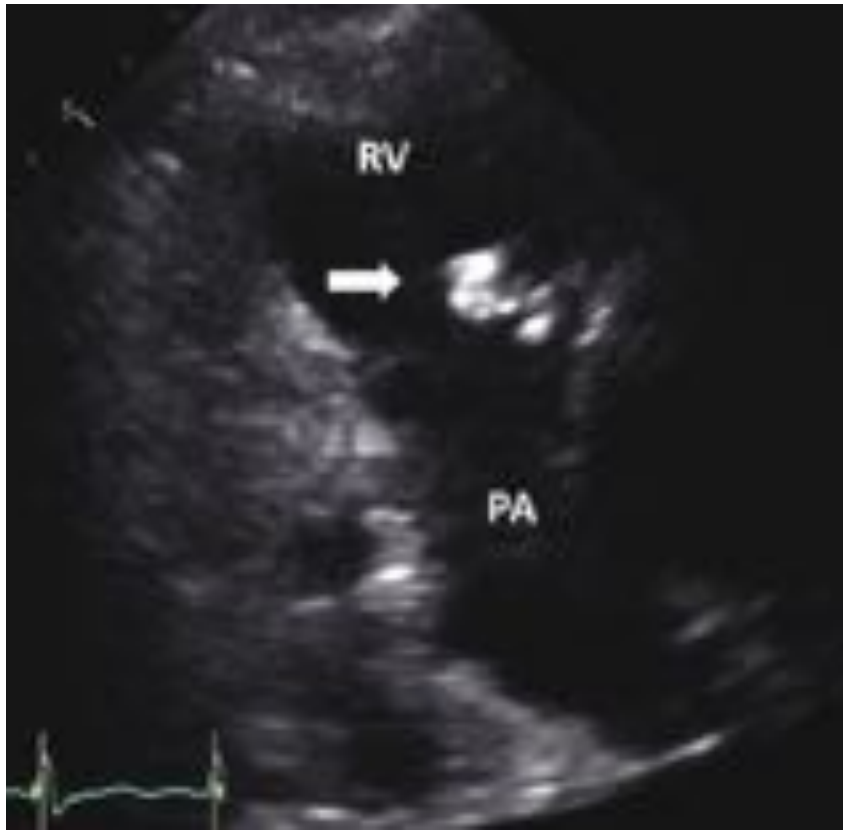
- Voie d'éjection droite?
- Valve Melody?
- Problème plus complexe?
  - Population à risque
  - Facteurs anatomiques
  - Facteurs hémodynamiques
  - Technique percutanée
  - Substrat valvulaire

VALVULOPATHIE PULMONAIRE

Infective Endocarditis Involving the Pulmonary Valve

William R. Miranda, MD[a,*], Heidi M. Connolly, MD[a], Daniel C. DeSimone, MD[b], Sabrina D. Phillips, MD[c], Walter R. Wilson, MD[b], Muhammad R. Sohail, MD[b], James M. Steckelberg, MD[b], and Larry M. Baddour, MD[a,b]

- Neuf patients 2000 - 2014
- 0.8% EI sur la période d'étude
- Age médian = 59 ans, 78% H
- 3 CC, 3 PM/DAI, 2 KT centraux
- Cinq patients (56%) RVP
- Pas de décès



Miranda et al. Am J Card 2015

VALVULOPATHIE PULMONAIRE

Infective Endocarditis in Children With Congenital Heart Disease

Cumulative Incidence and Predictors

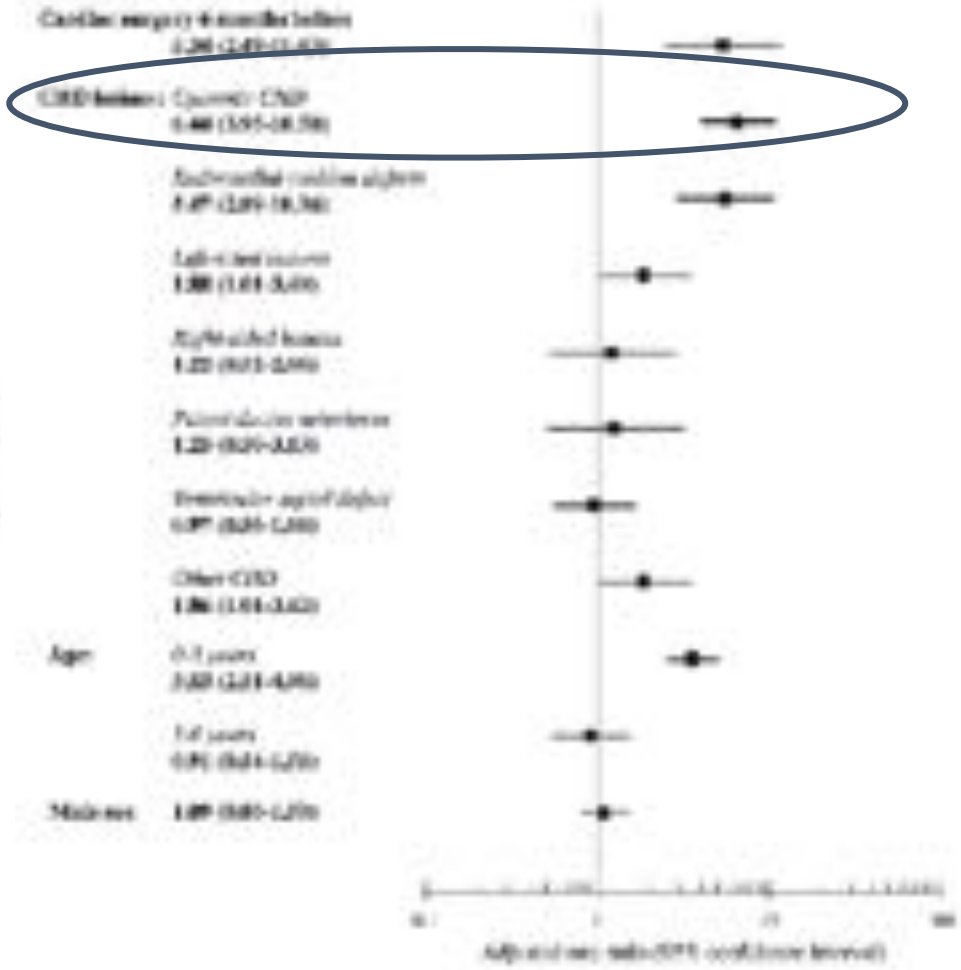
34 279 enfants avec CC suivis de 0 à 18 ans

Incidence annualisée = 4.1 / 10 000 pt-année

Table 2. Lesion Group–Specific Cumulative Incidence and Incidence Rate of IE in Children With CHD

CHD Lesions	Cumulative Incidence (95% CI) per 100 Children			Incidence Rate (95% CI) per 10 000 Person-Years
	0–6 y	0–12 y	0–18 y	
Cyanotic CHD	16.8 (11.9–23.8)	23.3 (17.0–31.8)	31.0 (22.5–42.7)	20.7 (15.4–27.7)
Endocardial cushion defects	5.5 (2.3–13.1)	8.7 (4.1–18.6)	11.1 (5.4–22.9)	7.7 (3.9–15.4)
Left-sided lesions	2.7 (1.3–5.7)	4.8 (2.6–8.7)	7.9 (4.4–14.0)	4.4 (2.6–7.4)
Right-sided lesions	2.5 (1.0–5.5)	2.3 (1.0–5.5)	4.2 (1.5–11.5)	2.8 (1.3–6.5)
Patent ductus arteriosus	3.2 (1.4–7.1)	3.2 (1.4–7.1)	3.2 (1.4–7.1)	3.5 (1.6–7.7)
Ventricular septal defect	2.0 (1.2–3.2)	2.4 (1.5–3.8)	3.2 (1.9–5.3)	2.4 (1.5–3.7)
Atrial septal defect	1.9 (1.3–2.9)	2.2 (1.5–3.4)	3.0 (1.9–4.8)	2.3 (1.6–3.4)
Other CHD	2.9 (1.4–5.8)	3.7 (1.8–7.3)	5.5 (2.9–10.6)	3.7 (2.0–6.7)
Overall	3.2 (2.6–3.9)	4.2 (3.5–5.1)	6.1 (5.0–7.5)	4.1 (3.5–4.9)

CHD indicates congenital heart disease; CI, confidence interval; and IE, infective endocarditis.



Rushani et al. Circulation 2013

VALVULOPATHIE PULMONAIRE

Table 4 Prediction model for developing IE, and score chart for the risk of developing IE up to 5 and 10 years

Predictor	HR(95% CI)	Points
Baseline valve-containing prosthetics	3.57(2.38–5.36)	3
Main defect[a]		
Pulmonary atresia with ventricular septal defect	4.05(1.85–8.86)	3
Double-outlet right ventricle	3.01(0.91–9.94)	2
Tetralogy of Fallot	1.81(0.99–3.33)	1
Univentricular heart	1.69(0.51–5.54)	1
Left-sided lesions	1.55(0.99–2.44)	1
Other	1	0
Multiple defects	1.68(1.15–2.46)	1
History of IE	2.21(1.22–4.01)	2
Male	1.89(1.28–2.81)	1

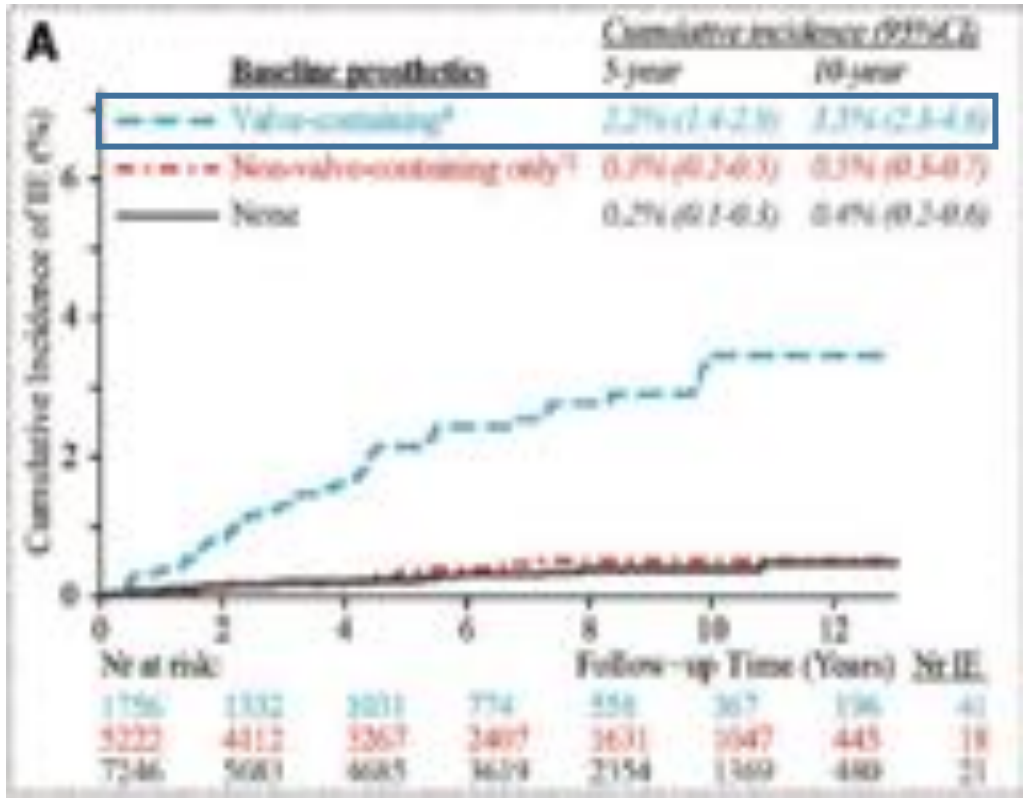
Score (sum points)

	Score									
	0	1	2	3	4	5	6	7	8	>8
Predicted 5 year risk (%)	<1	<1	1	1	1	2	3	4	7	9
Predicted 10 year risk (%)	<1	1	1	1	3	3	5	7	12	15

Registre CONCOR (14 224 patients>18 ans)

Incidence EI : **1.33/1000 pt-years**

Prothèse valvulaire: **HR=5.48(3.58–8.38)**



Kuijpers et al. Eur Heart Jour 2017

VALVULOPATHIE PULMONAIRE

Defect	Prosthetic aortic valve (-containing conduit)	N°	IE cases,n First	IE cases,n Total	Person-time,y	Incidence rate, cases per 1000py (95% CI)	Univariable HR	Univariable 95% CI	Univariable p value
LVOTO	No	670	4	4	4206	0.95 (0.30-2.21)	1		
	Yes	410	13	14	3854	4.91 (2.36-7.94)	5.35	1.74-16.45	0.003
BAV	No	1087	3	3	4957	0.61 (0.15-1.57)	1		
	Yes	315	10	10	1907	5.24 (2.53-9.20)	7.90	2.16-28.90	0.002
CoA	No	1299	9	11	8473	1.30 (0.67-2.22)	1		
	Yes	131	2	2	1075	1.86 (0.31-5.74)	1.83	0.39-8.52	0.447
Marfan	No	412	2	2	3242	0.62 (0.08-1.90)	1		
	Yes	185	9	9	1443	6.24 (3.00-11.24)	11.16	2.41-51.75	0.002
Other	No	99	1	1	489	2.05 (0.13-9.00)	1		
	Yes	71	0	0	409	0 (N/A)	N/A	N/A	N/A
All	No	3502	19	21	21370	0.98 (0.52-1.45)	1		
	Yes	1231	34	35	9687	4.55 (3.21-6.23)	5.23	2.99-9.18	<0.999

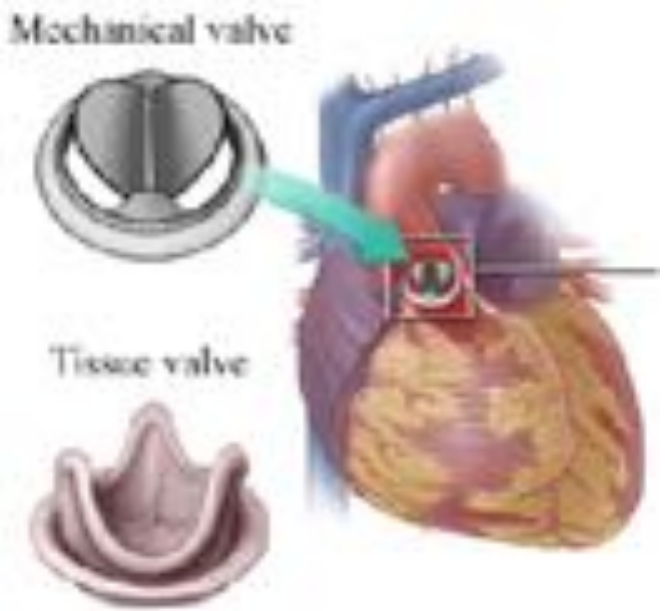


Table 1 Number of IE episodes according to main congenital heart lesion

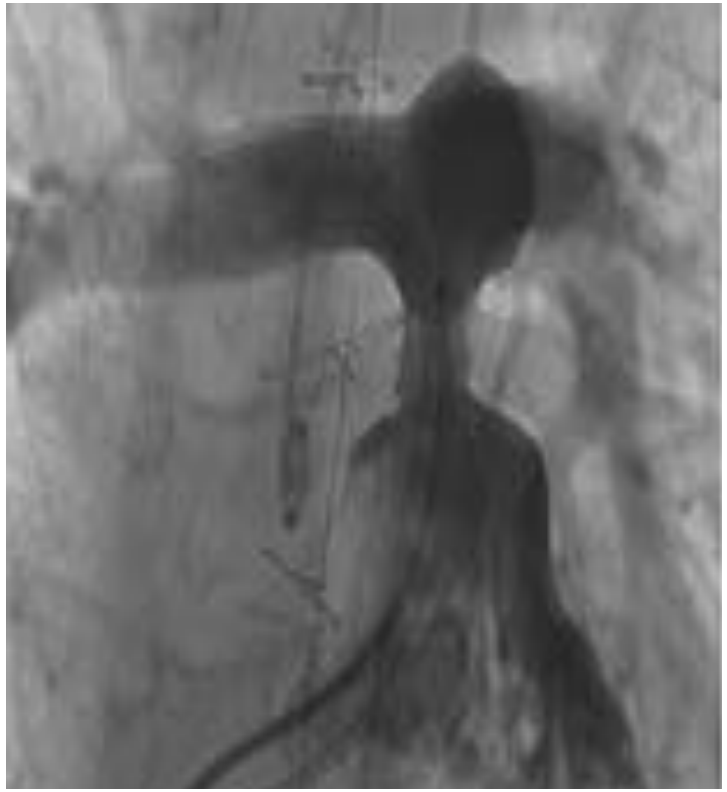
Diagnosis	Number of episodes (%)
Left ventricular outflow tract disease	44 (25.8)
Shunt	42 (25.6)
Tetralogy of Fallot/pulmonary atresia	36 (22.0)
TGA	20 (12.2)
Complex CHD	7 (4.3)
Single ventricle physiology	6 (3.7)
Mitral valve disease	3 (1.8)

Tutarel O, *et al*. *Heart* 2017
Kuijpers et al. Eur Heart Jour 2017

VALVULOPATHIE PULMONAIRE

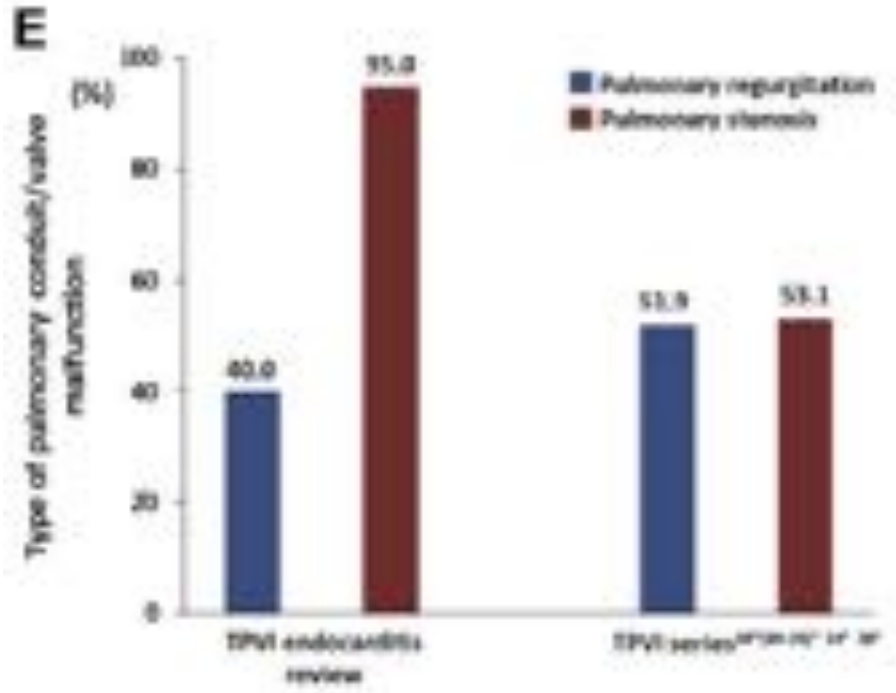
Invasiveness of left- and right-sided infective endocarditis: Does pressure explain pathology?

- Endocardites du cœur droit moins invasives
- Effet protecteur du régime à basse pression



Hussain et al. JTCS 2017
Bolt et al. JAMA ica 2013

VALVULOPATHIE PULMONAIRE



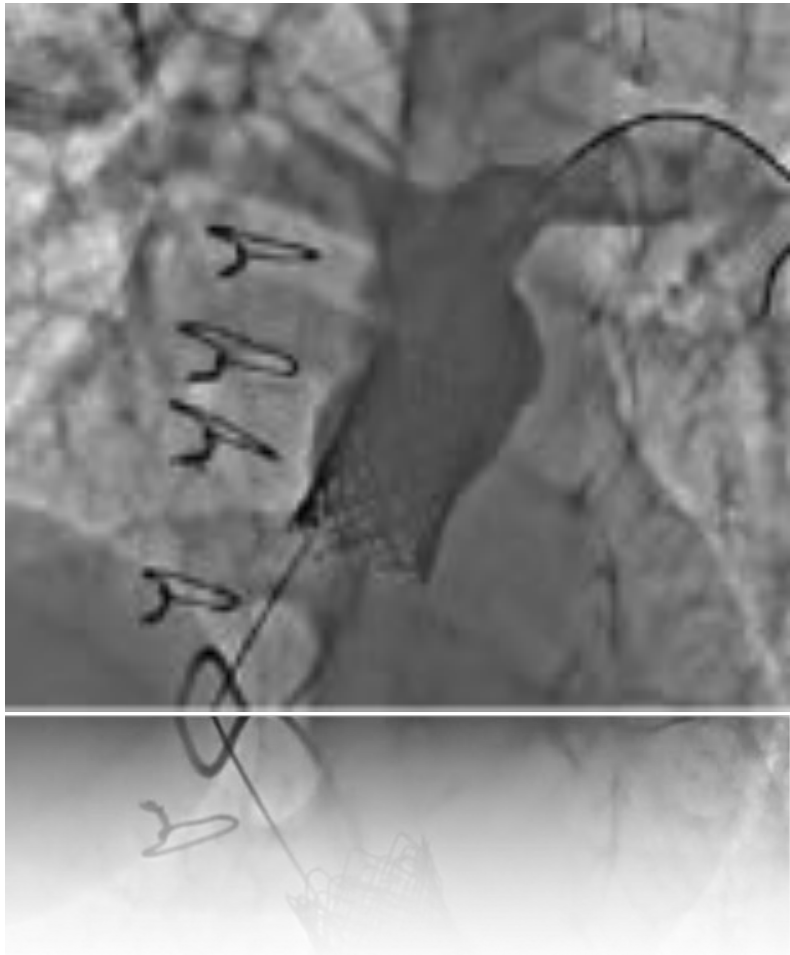
Lésions sténosantes

Tubes non expansibles

Préstenting

Gradient résiduel

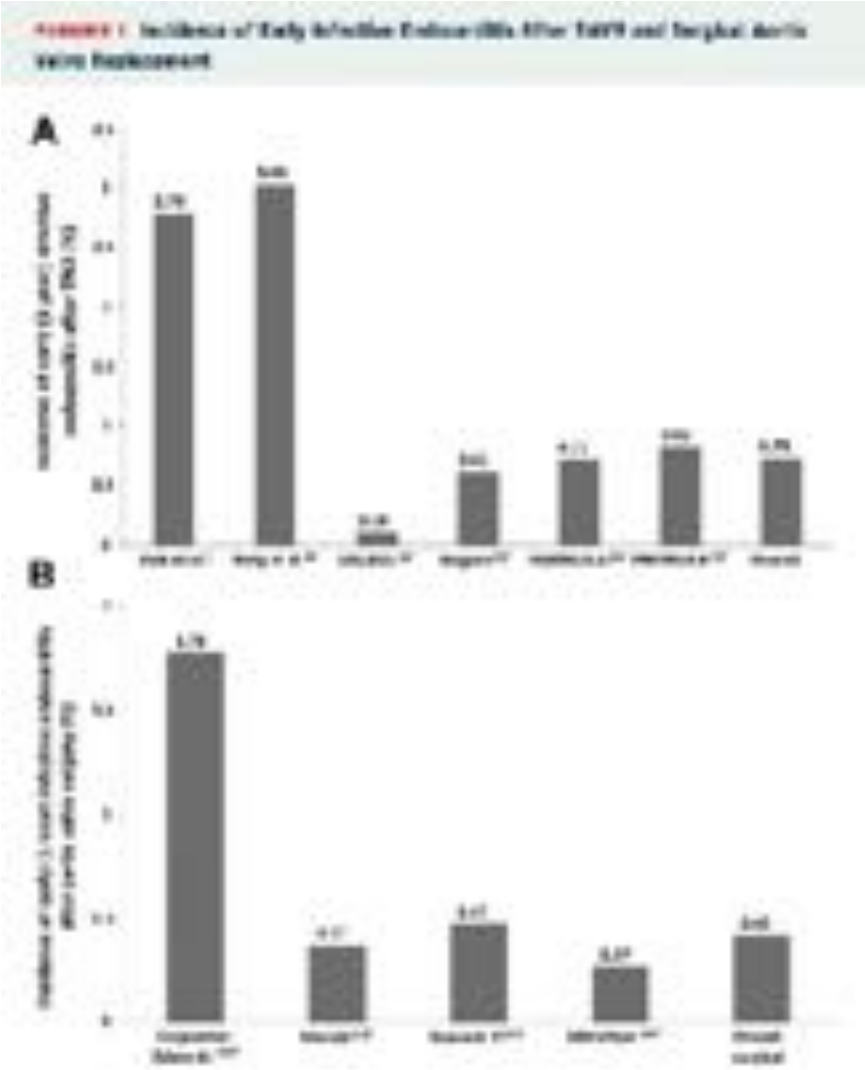
Irrégularités - Logettes



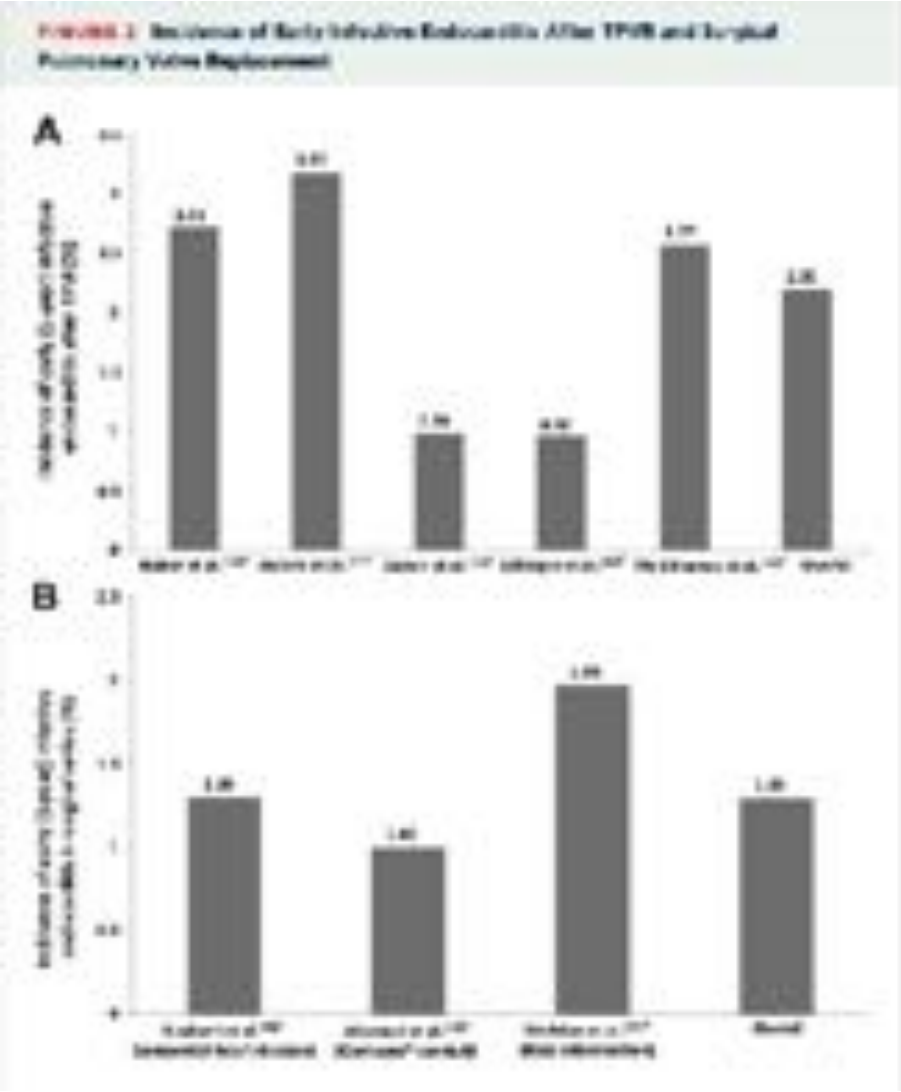
Buber et al. Circ Intrv 2013
Amat-Santos et al. JACC Intrv 2015

VALVULOPATHIE PULMONAIRE

TAVI

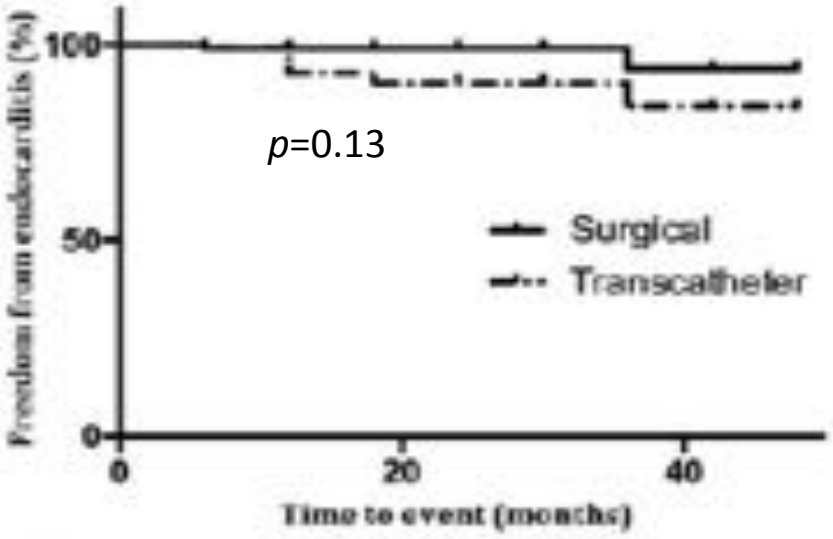


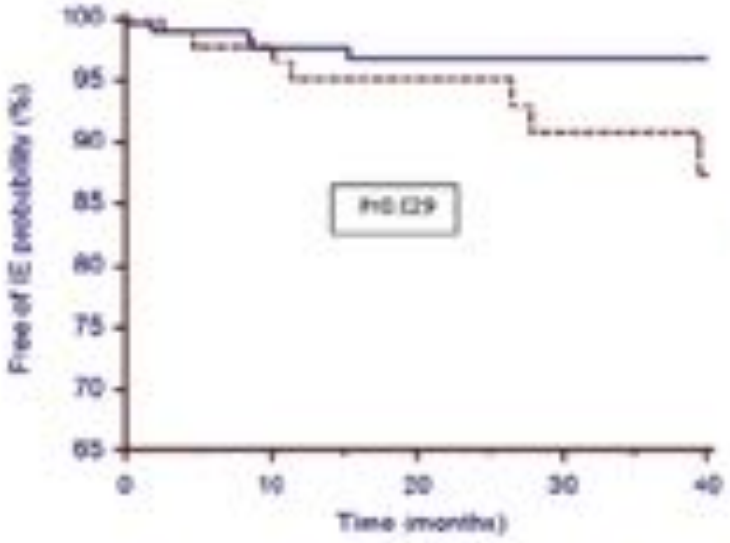
PPVI

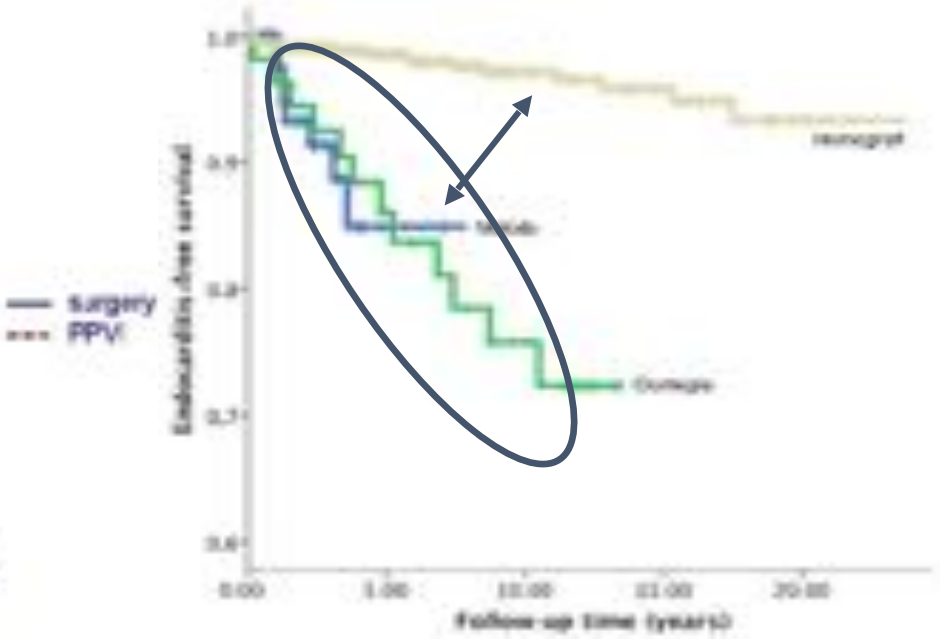


Amat-Santos et al. JACC Intrv 2015

VALVULOPATHIE PULMONAIRE







134 chir et 208 percut (33 Sapien)

Incidence IE: 0.5 vs 1.5 %pt/années

195 chir et 93 percut (0 Sapien)

Incidence IE: 1.2 vs 3.9 %pt/années

631 chir et 107 percut (0 Sapien)

Incidence IE: 0.8 vs 2.7 vs 3% %pt/années

Lluri et al. CCI 2017

Malekzadeh-Milani et al. JTCS 2014

Van Dijck et al. Heart 2014

VALVULOPATHIE PULMONAIRE

Author	Year	n	Substrate	EI Cumulative incidence	EI Annualized Incidence (% pt-year)	Median Follow-up (years)
Albanesi	2014	12/106	Contegra	11.3		7.6
Malekzadeh	2014	5/190	Homografts Contegra	2.6	1.2	2
Ramanan	2015	6/115	Freestyle	5.4	-	4.3
Mery	2016	23/586	Homograft Contegra Porcine valve	4	-	7
Ugaki	2016	21/298	Contegra Homograft	7	-	3.4

Tous les dispositifs valvulaires sont suseptibles d'etre le siège d'une EI
Avec une incidence variable mais significative

Albanesi et al. EJCTS 2014
Ramanan et al. Ann Thorac Surg 2015
Ugaki et al. Ann Thorac Surg 2016
Mery et al. JTCS 2016

VALVULOPATHIE PULMONAIRE

A Systematic Review of Infective Endocarditis in Patients With Bovine Jugular Vein Valves Compared With Other Valve Types

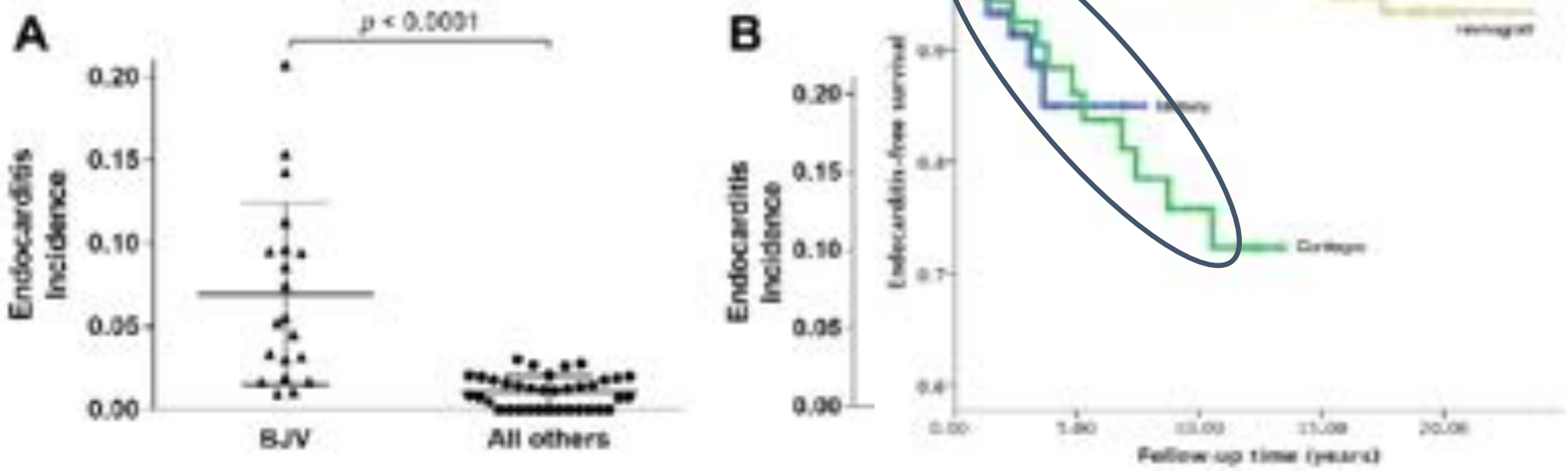
Méta-analyse sur IE chez les patients avec RVP chirurgical ou percutané

7063 patients

Incidence cumulative globale = 2.5%

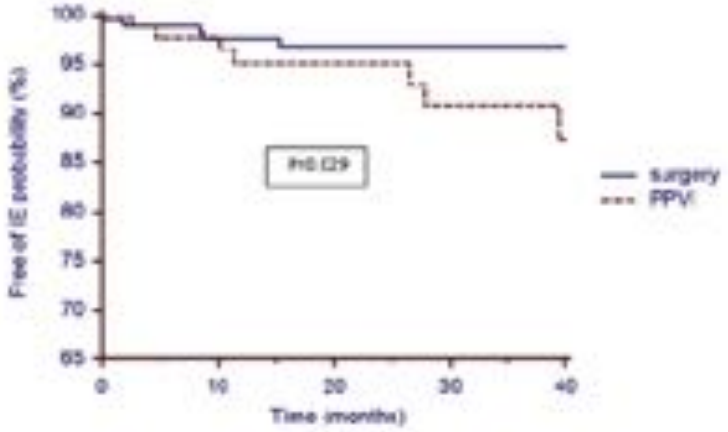
VJB vs autres substituts : 5.4% vs 1.2%; p < 0.0001

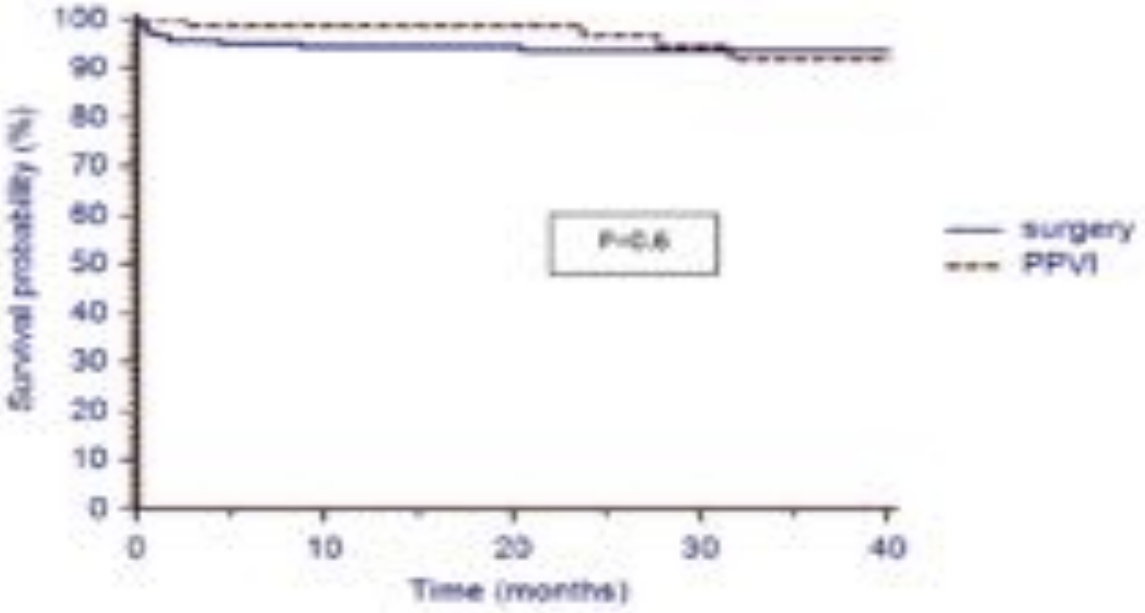
FIGURE 3 Incidence of Infective Endocarditis in BJV Compared to Other Valves

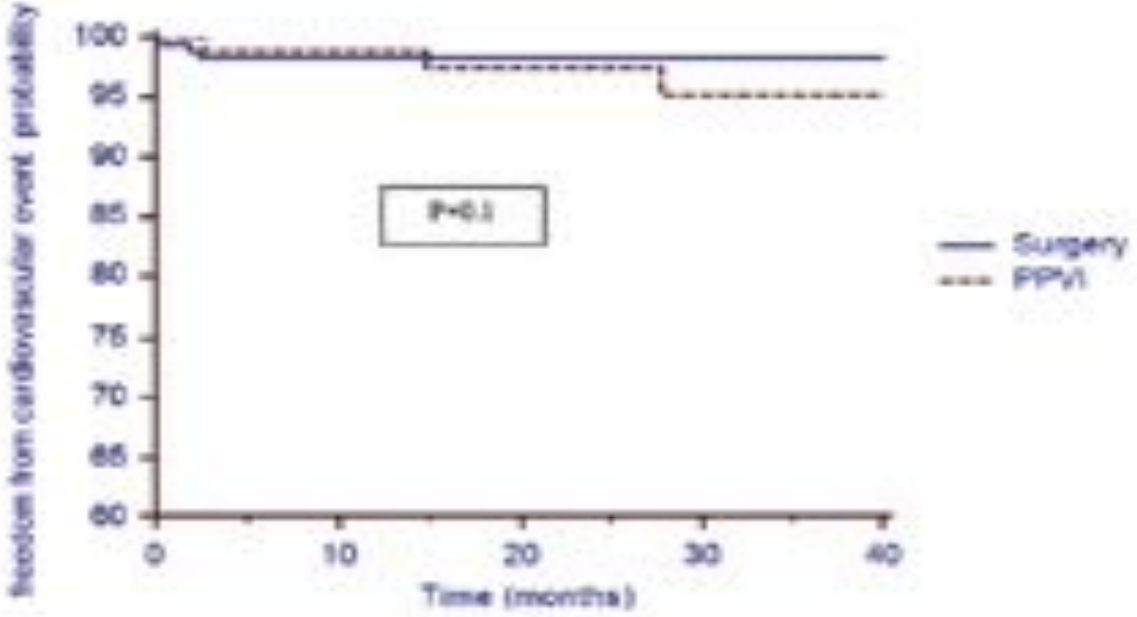


Sharma et al. JACC Int. 2017

VALVULOPATHIE PULMONAIRE







IMPACT ECONOMIQUE

Cost Analysis of Percutaneous Pulmonary Valve Replacement

Scott W. Gatlin, MD, Dennis W. Kim, MD, PhD, and William T. Mahle, MD*

patient). In conclusion, Melody valve implantation compares reasonably well to surgical conduit revision despite the added midterm costs, but ongoing analysis including the impact of nonsurgical options on quality-of-life measurements and improvement of reintervention rates for percutaneously placed valves needs to be considered. © 2011 Elsevier

Cost-Analysis of Percutaneous Pulmonary Valve Implantation Compared to Surgical Pulmonary Valve Replacement

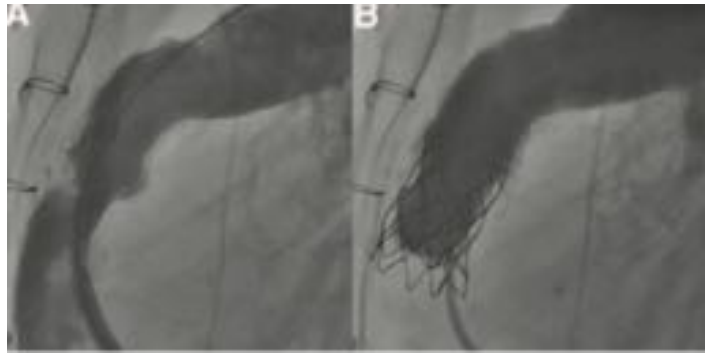
per year (or 93% at 10 years) to lose its cost advantage. Conclusions: PPVI holds a significant cost advantage over the surgical approach, fewer hospital days, and incurs less patient wage loss. Furthermore, it would need to have a very high failure rate at 10 years to lose its cost advantage. © 2013 Wiley Periodicals, Inc.

Gatlin et al. Am J Cardiol 2011

Vergales et al. Catheter Cardiovasc Interv. 2013

DÉVELOPPEMENT CROISSANT DE LA VALVULATION PERCUTANÉE

- Efficacité équivalente pour une approche moins invasive
- Indications élargies, limites techniques repoussées
- Futur radieux: nouveaux dispositifs, nouvelles technologies



VALVULOPATHIE PULMONAIRE

RECOMMANDATIONS ESC/AHA

≈ indications de RVP chirurgical

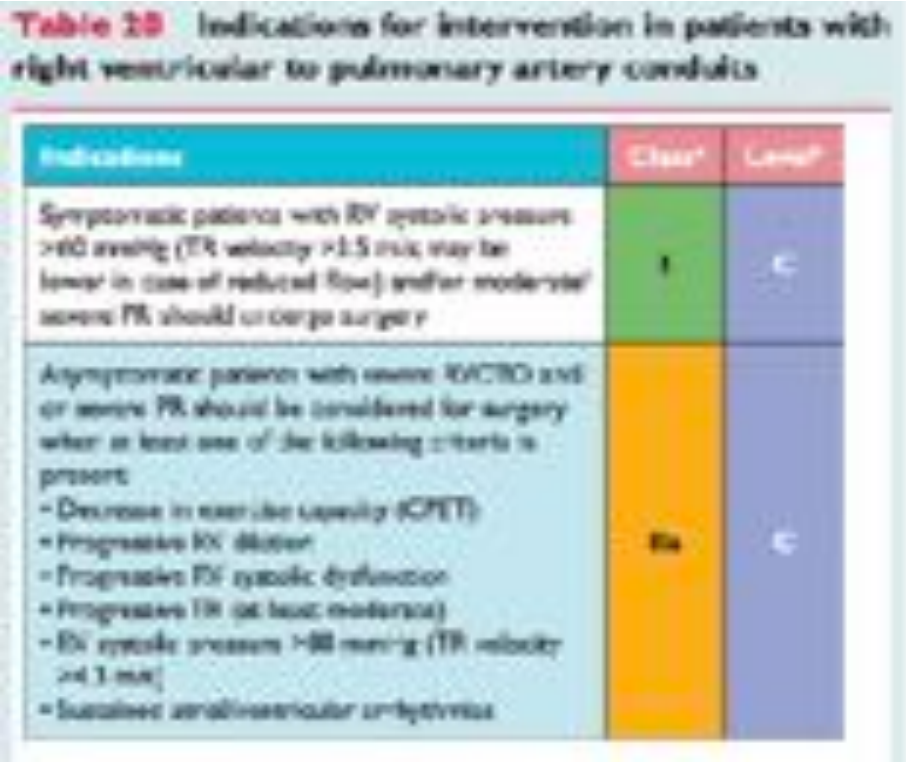


Table 29 Indications for intervention in patients with right ventricular to pulmonary artery conduits

Indications	Class*	Level*
Symptomatic patients with RV systolic pressure >60 mmHg (TR velocity >3.5 m/s; may be lower in case of reduced flow) and/or moderate/severe PR should undergo surgery	I	C
Asymptomatic patients with severe RVOTO and/or severe PR should be considered for surgery when at least one of the following criteria is present: - Decrease in exercise capacity (CPET) - Progressive RV dilation - Progressive RV systolic dysfunction - Progressive TR (at least moderate) - RV systolic pressure >80 mmHg (TR velocity >4.3 m/s) - Sustained ventricular arrhythmias	IIa	C

Baumgartner et al. Eur Heart J 2010
Feltes et al. Circulation 2011

« INSTRUCTIONS FOR USE »

- Conduit circonférentiel ≥ 16mm nominal ET
- Dysfonction de conduit symptomatique ET
  - Sténose avec Grdt moyen ≥ 35mmHg
  - Fuite ≥ modérée

15% des patients candidats à un RVP

OFF-LABEL

↓

Voies natives, patchées, bioprothèses

Schievano et al. J Cardiovasc Magn Reson 2007

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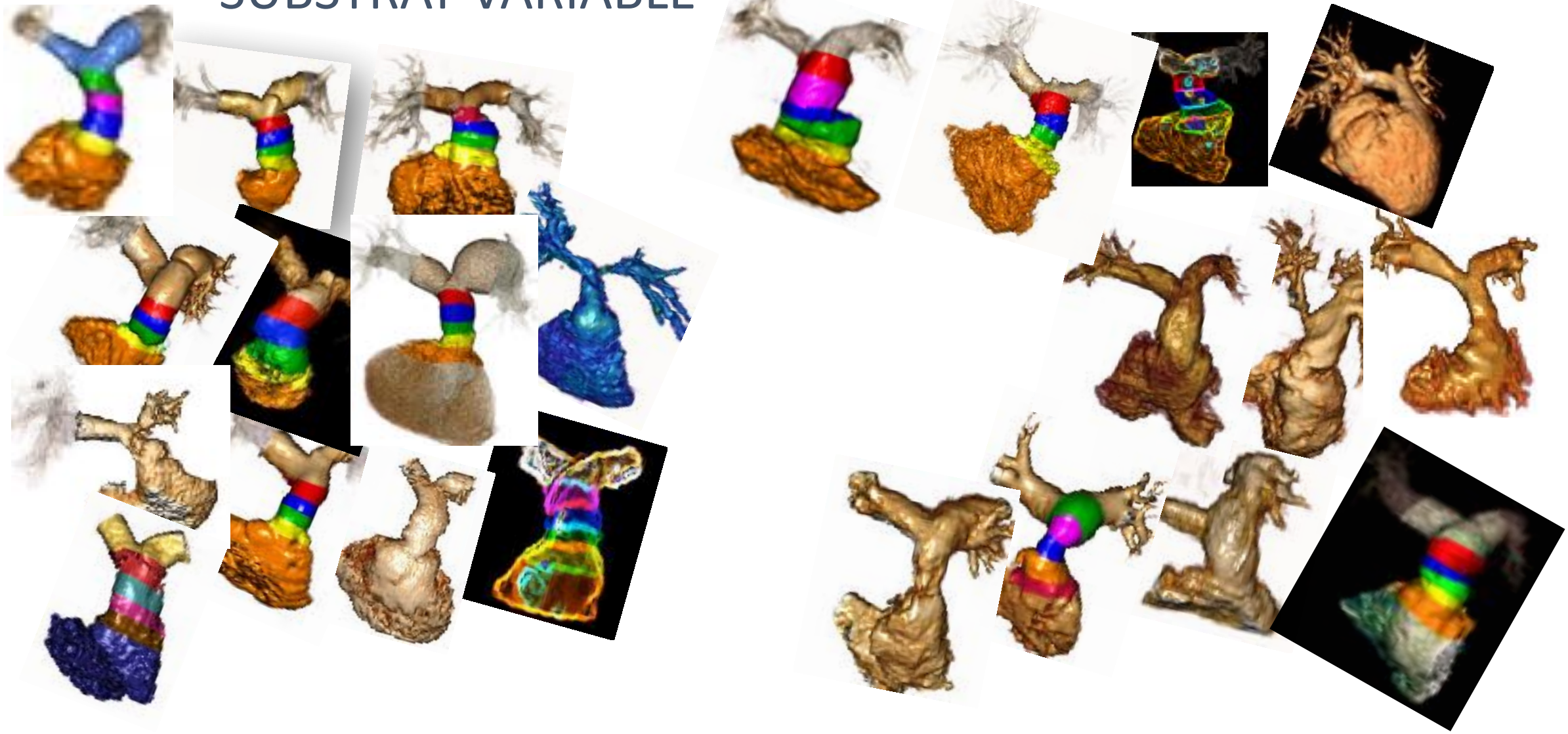
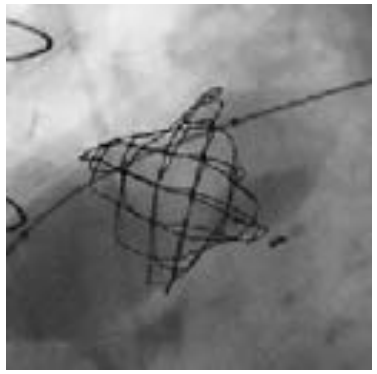
SUBSTRAT VARIABLE



VALVULOPATHIE PULMONAIRE

Transcatheter pulmonary valve replacement using the melody valve for treatment of dysfunctional surgical bioprostheses: A multicenter study

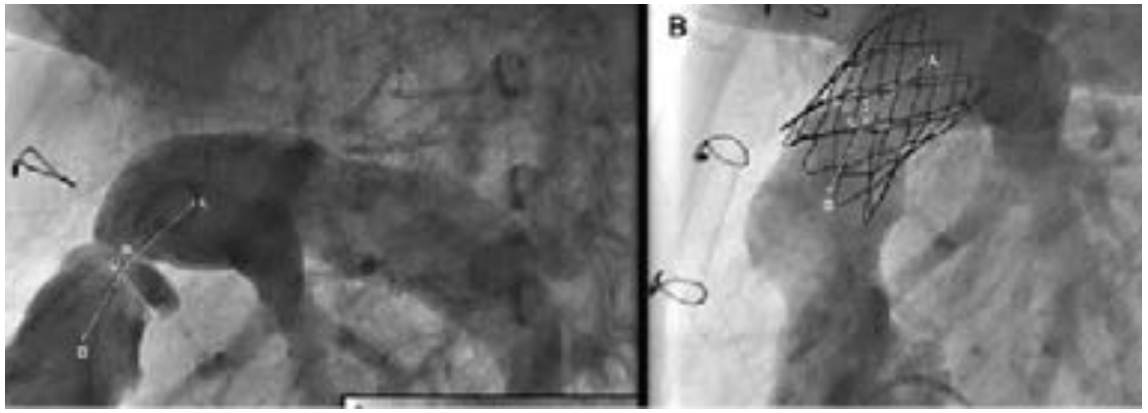
Transcatheter Pulmonary Valve Replacement With the Melody Valve in Small Diameter Expandable Right Ventricular Outflow Tract Conduits



Use and Performance of the Melody Transcatheter Pulmonary Valve in Native and Postsurgical, Nonconduit Right Ventricular Outflow Tracts

Percutaneous transcatheter pulmonary valve replacement in children weighing less than 20 kg

Mary Hunt Martin, MD[1] | Shabana Shahanavaz, MD[2] | Lynn F. Peng, MD[3] | Jeremy D. Asnes, MD[4] | Michelle Riley, NP[1] | William E. Hellenbrand, MD[4] | David T. Balzer, MD[2] | Robert G. Gray, MD[1] | Doff B. McElhinney, MD[3,5]



Cabalka et al. JTCVS 2018
Martin et al. CCI 2018

ELARGISSEMENT DES INDICATIONS

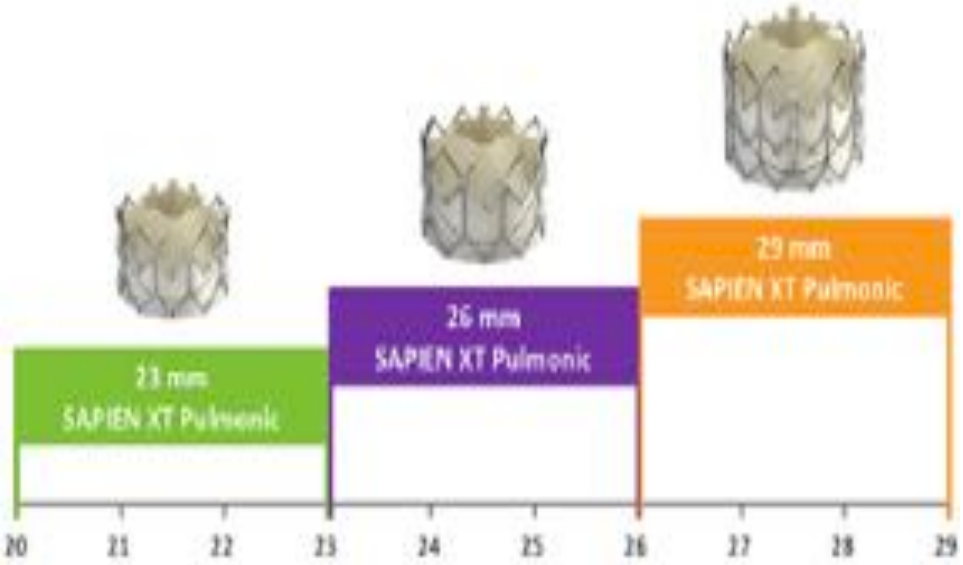
Shahanavaz et al. JACC Interv 2018
Meadows Circ Cardiovasc Interv. 2014

CHALLENGES TECHNIQUES: VOIES COMPLEXES

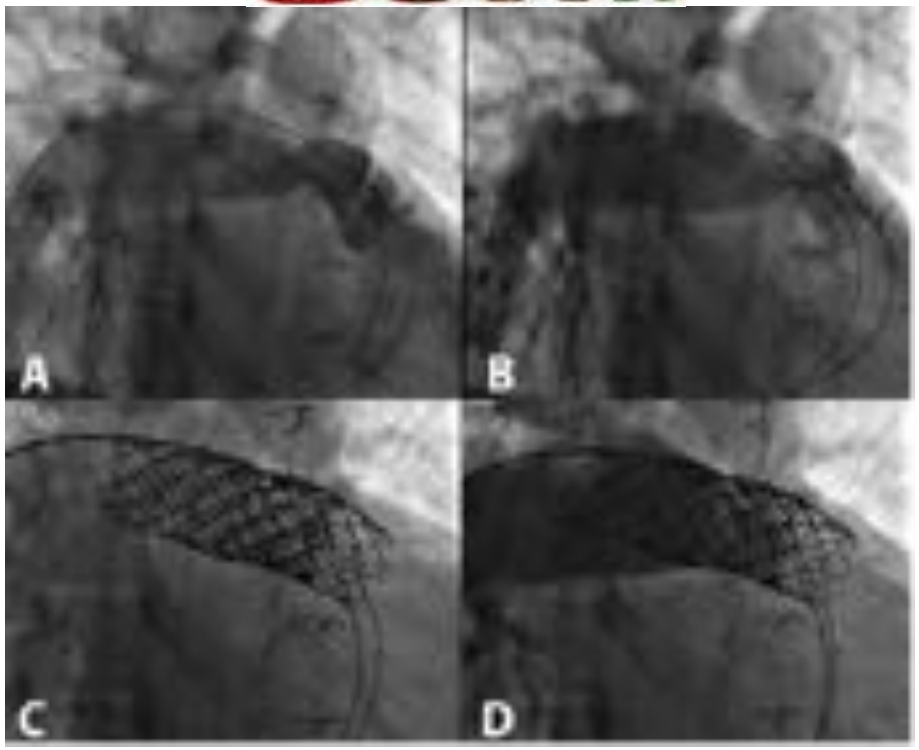
VOIES LARGES, VOIES COURTES, BIFURCATIONS PRECOCES, PROXIMITE CORONAIRE

TECHNIQUES ALTERNATIVES

Delivery System Size Inner Balloon / Outer Balloon	Corresponding Valve Outside Diameter (Balloon Inflated)
	mm
Size 18mm 9mm x 3.5cm/ 18mm x 4cm	20.1
Size 20mm 10mm x 3.5cm/20mm x 4cm	22.4
Size 22mm 11mm x 3.5cm/22mm x 4cm	24.1



VALVULOPATHIE PULMONAIRE



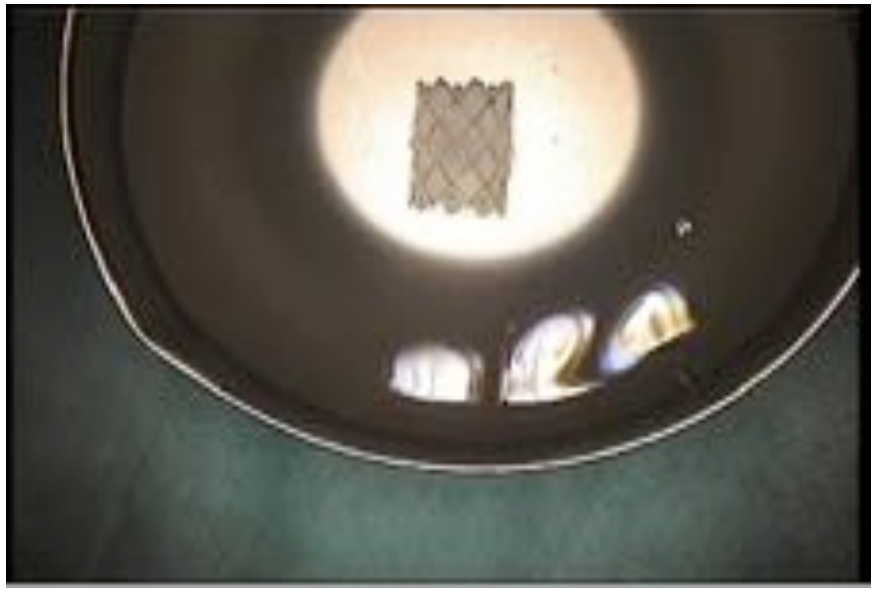
POUPEES RUSSES

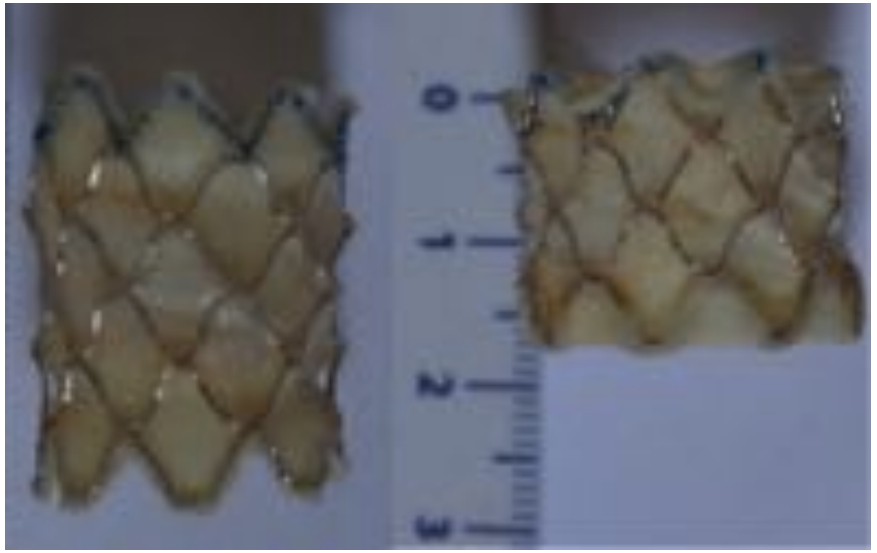
DIABOLO STENT

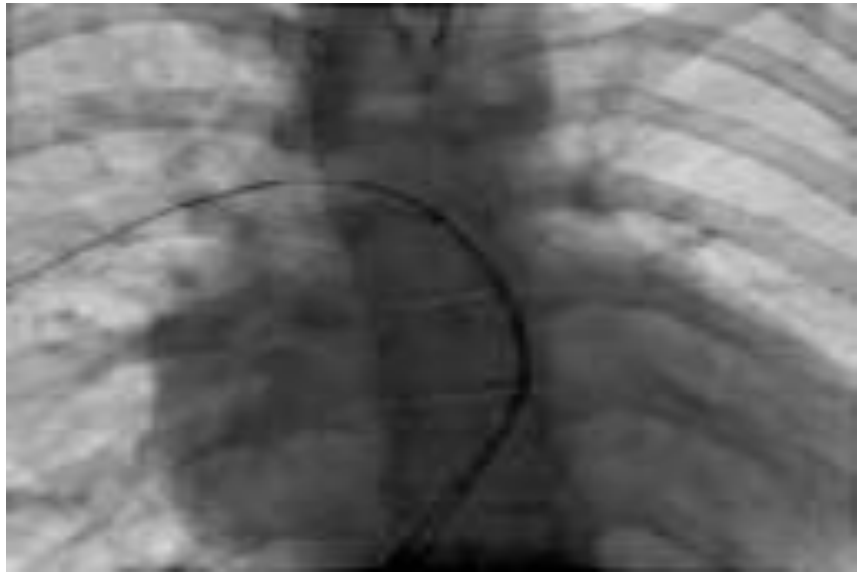
Boudjemline et al. ACVD, Circ Interv 2012
Malekzadeh-Milani et al. ACVD 2014

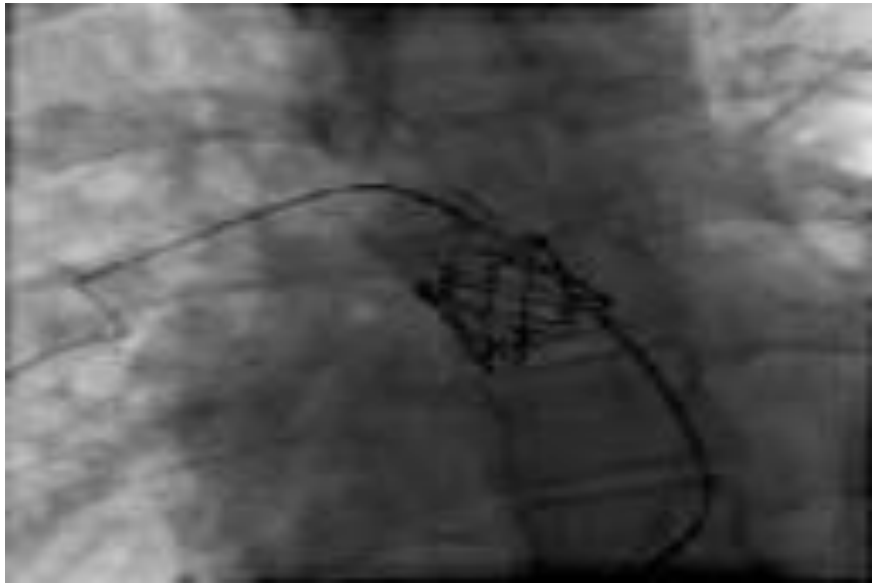
Malekzadeh-Milani et al. EuroIntervention 2016

VALVULOPATHIE PULMONAIRE



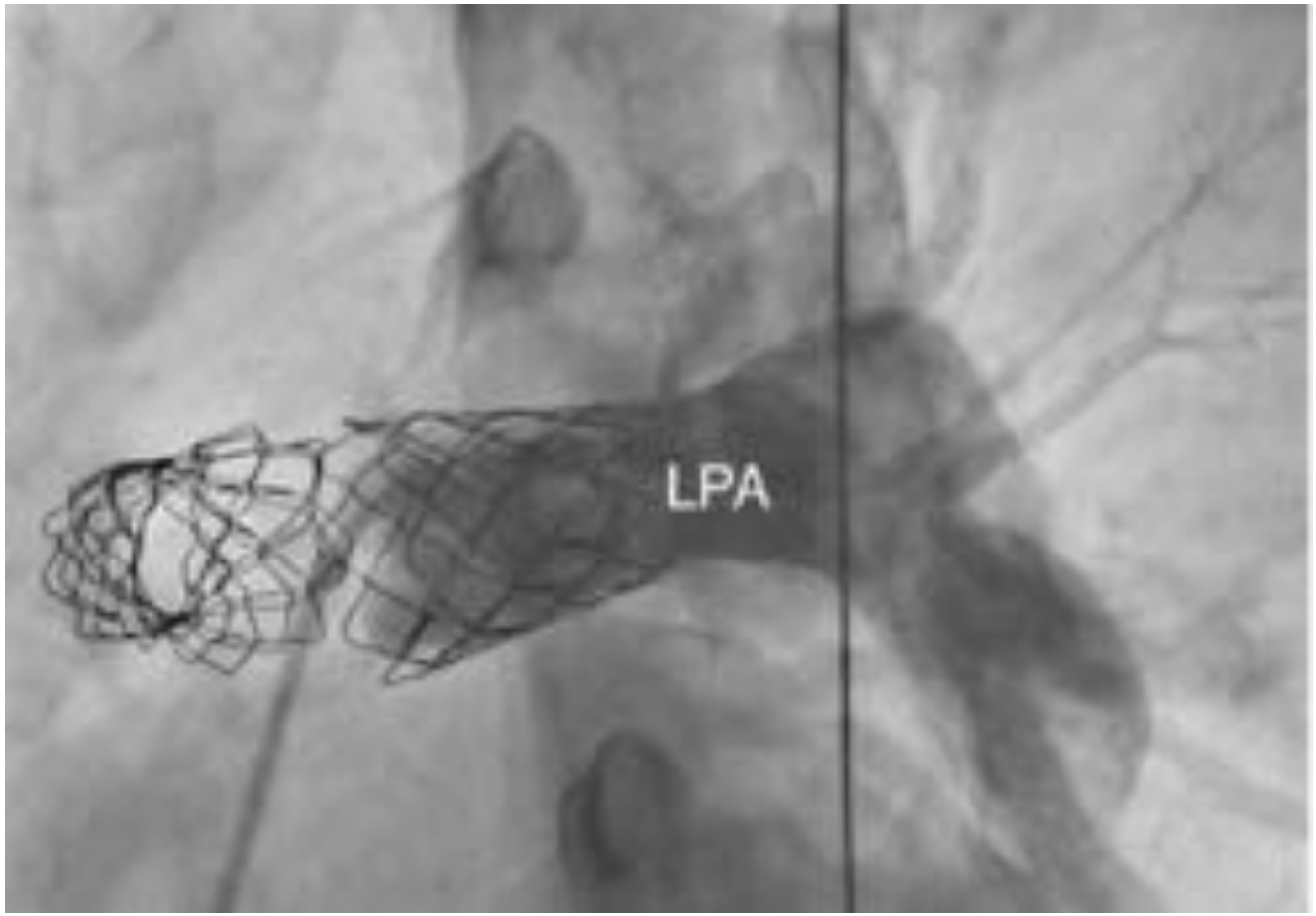






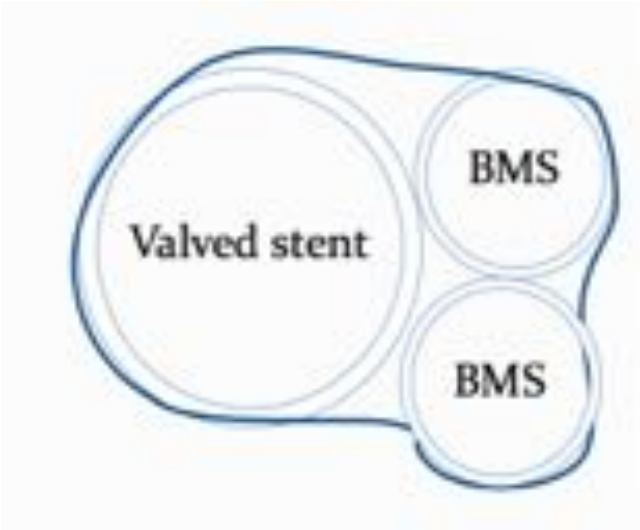
Jalal et al. CCI 2015, Malekzadeh-Milani et al. EuroInt 2015

CONCERTO POUR MELODYS

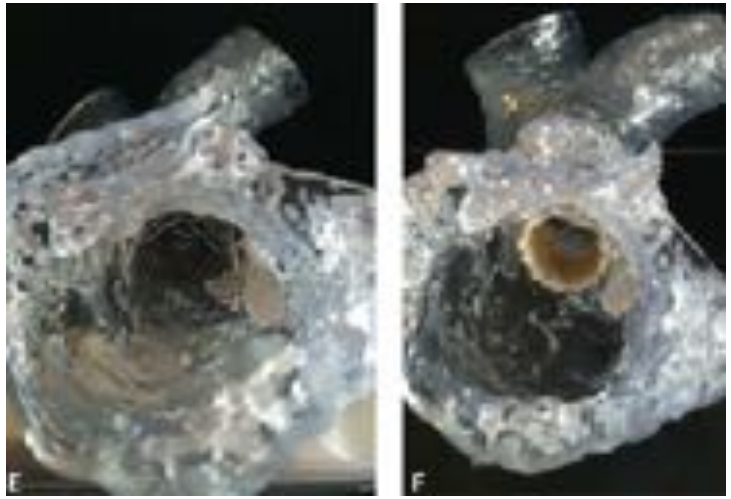


Maschietto et al. J Invasive Cardiol 2013
Gillespie et al. Circulation 2011

VALVULOPATHIE PULMONAIRE



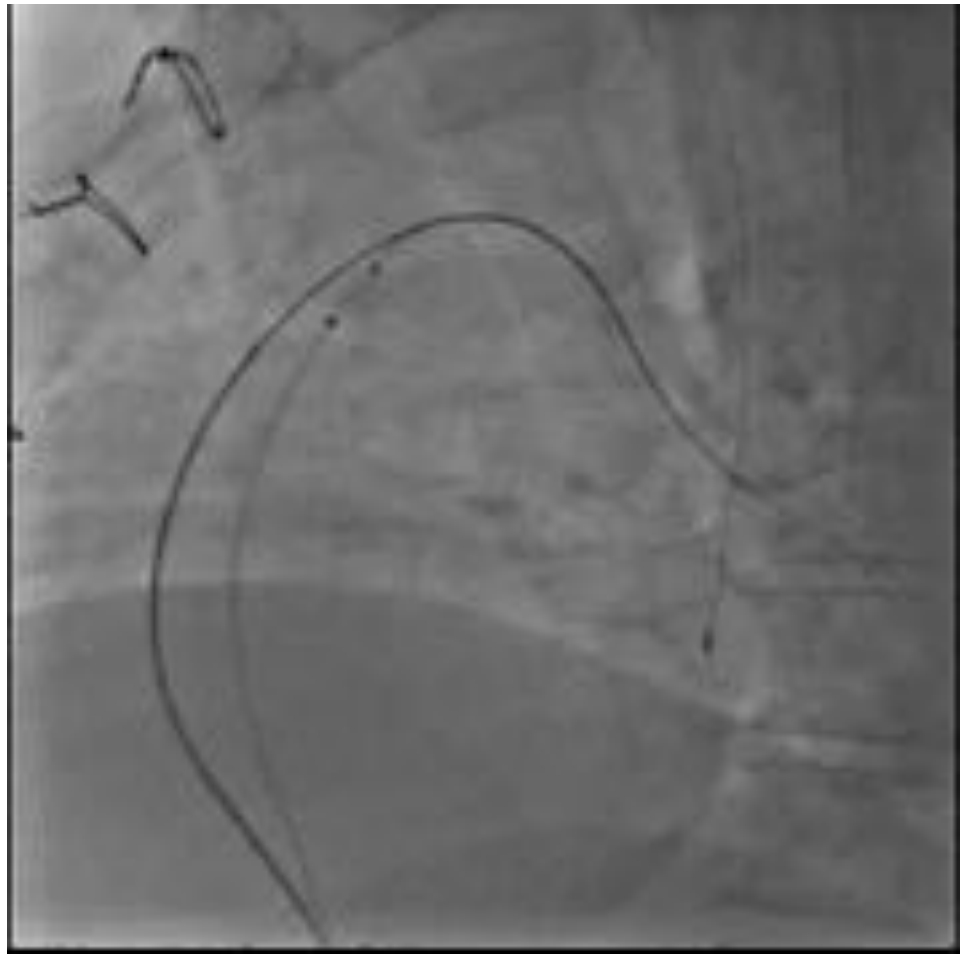
SIDE-BY-SIDE TECHNIQUE

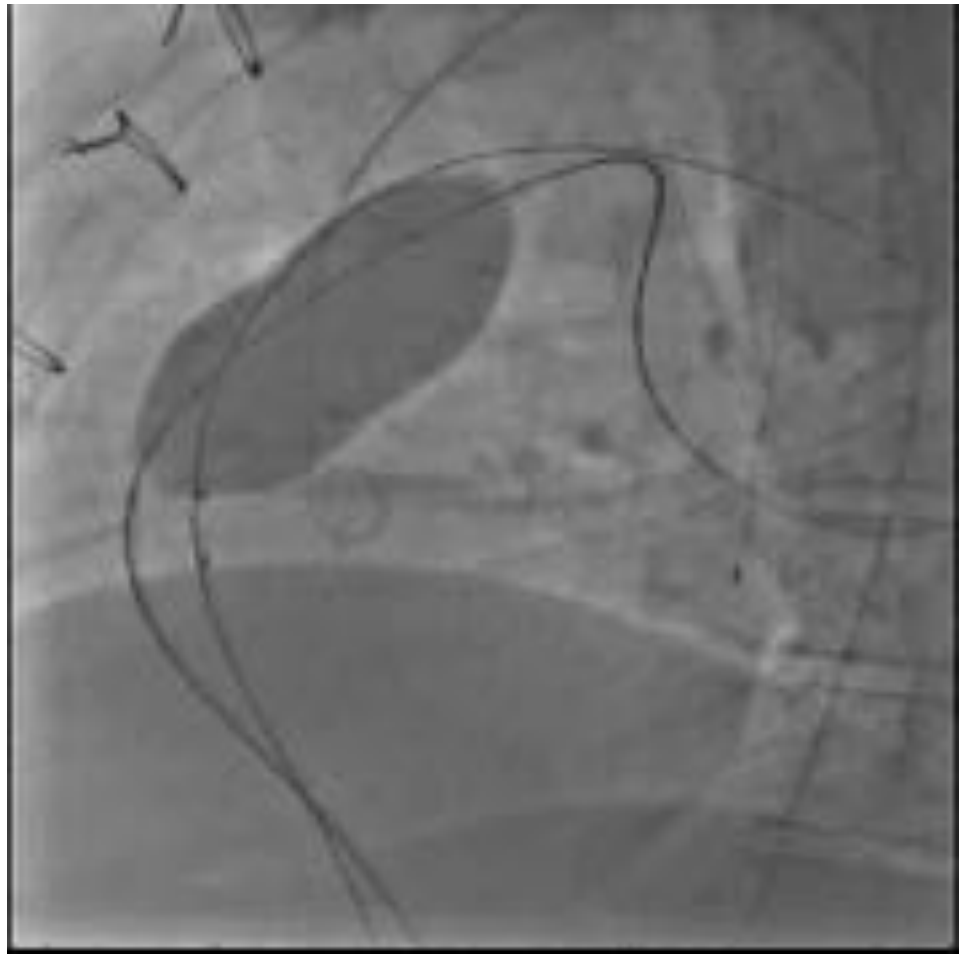


Phillips et al. CCI 2016

Courtesy of Younes Boudjemline

SIDE-BY-SIDE TECHNIQUE





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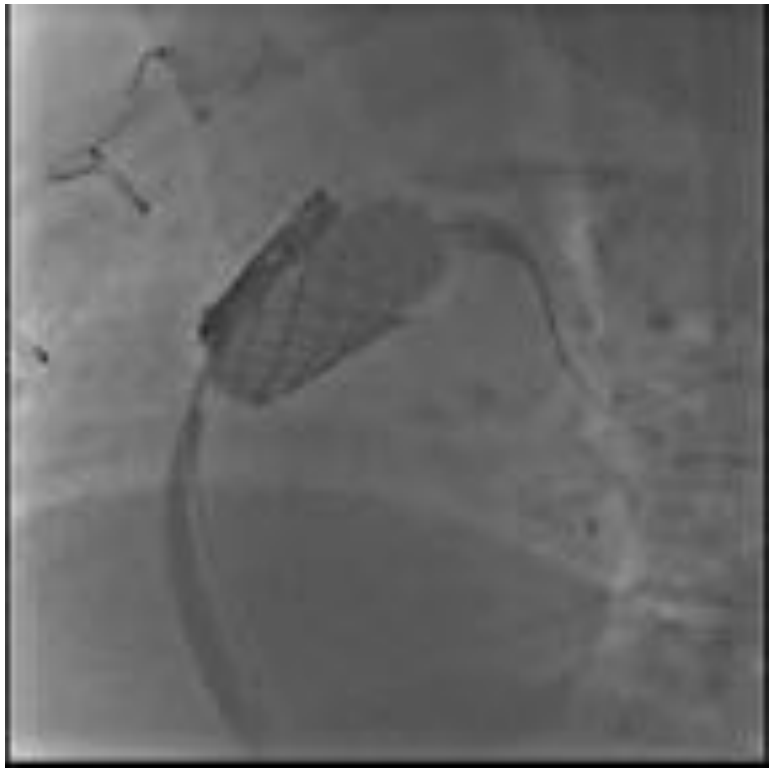
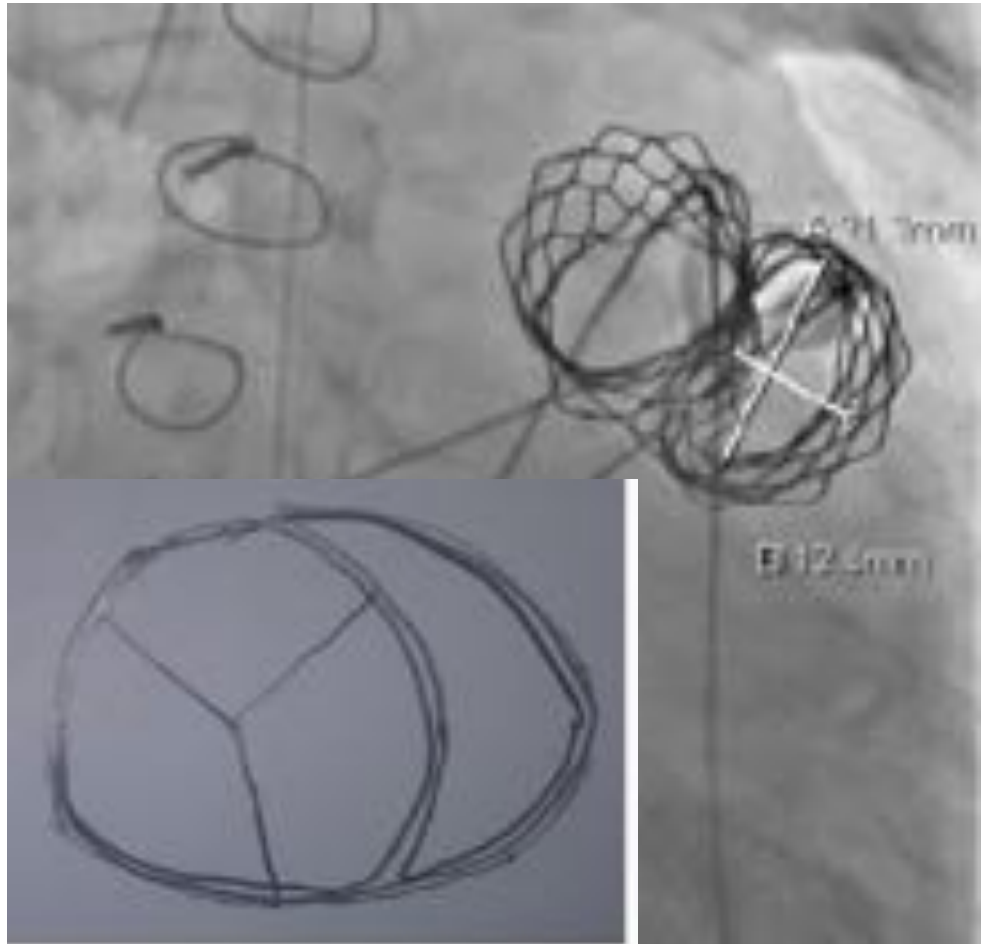
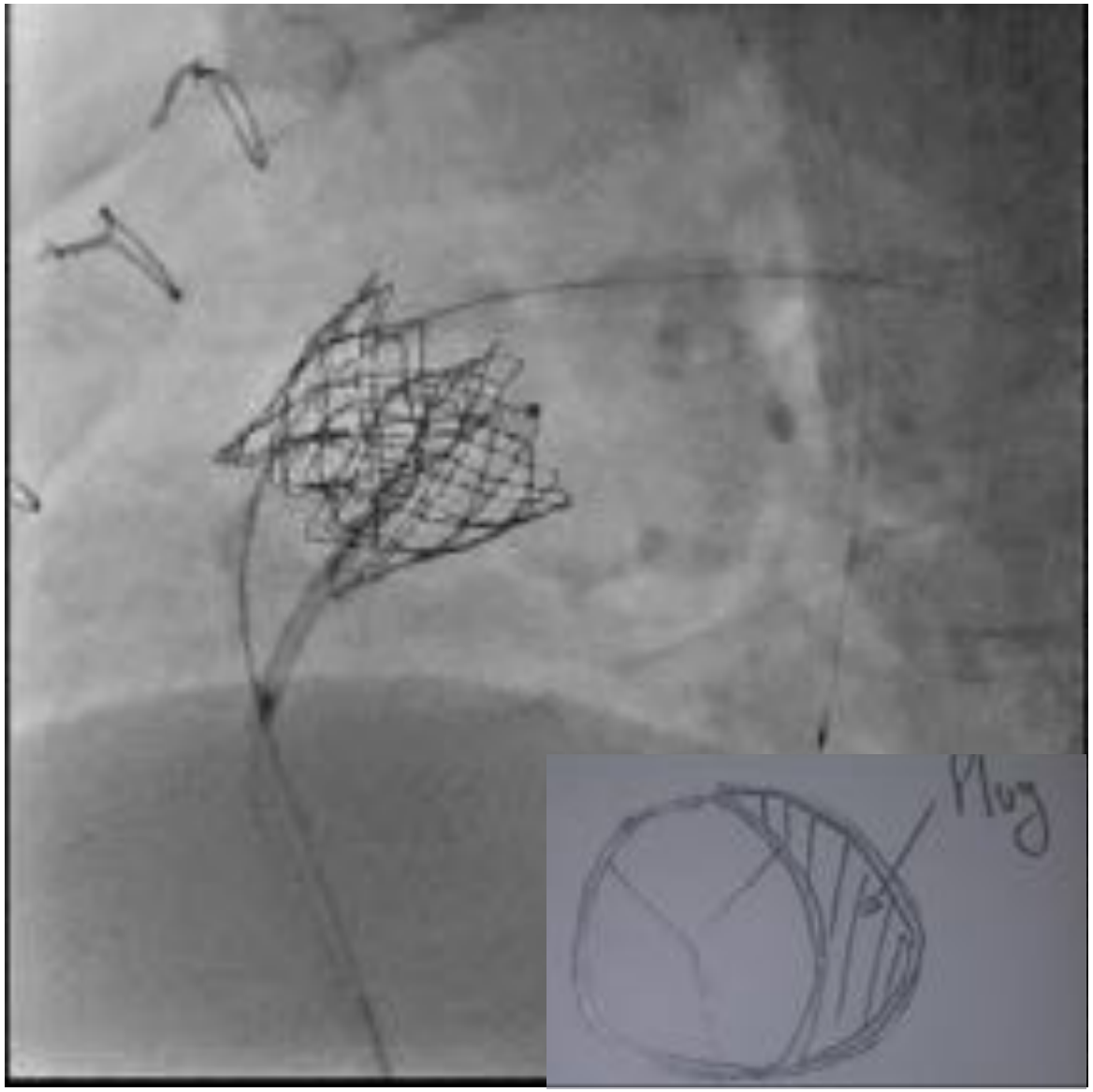
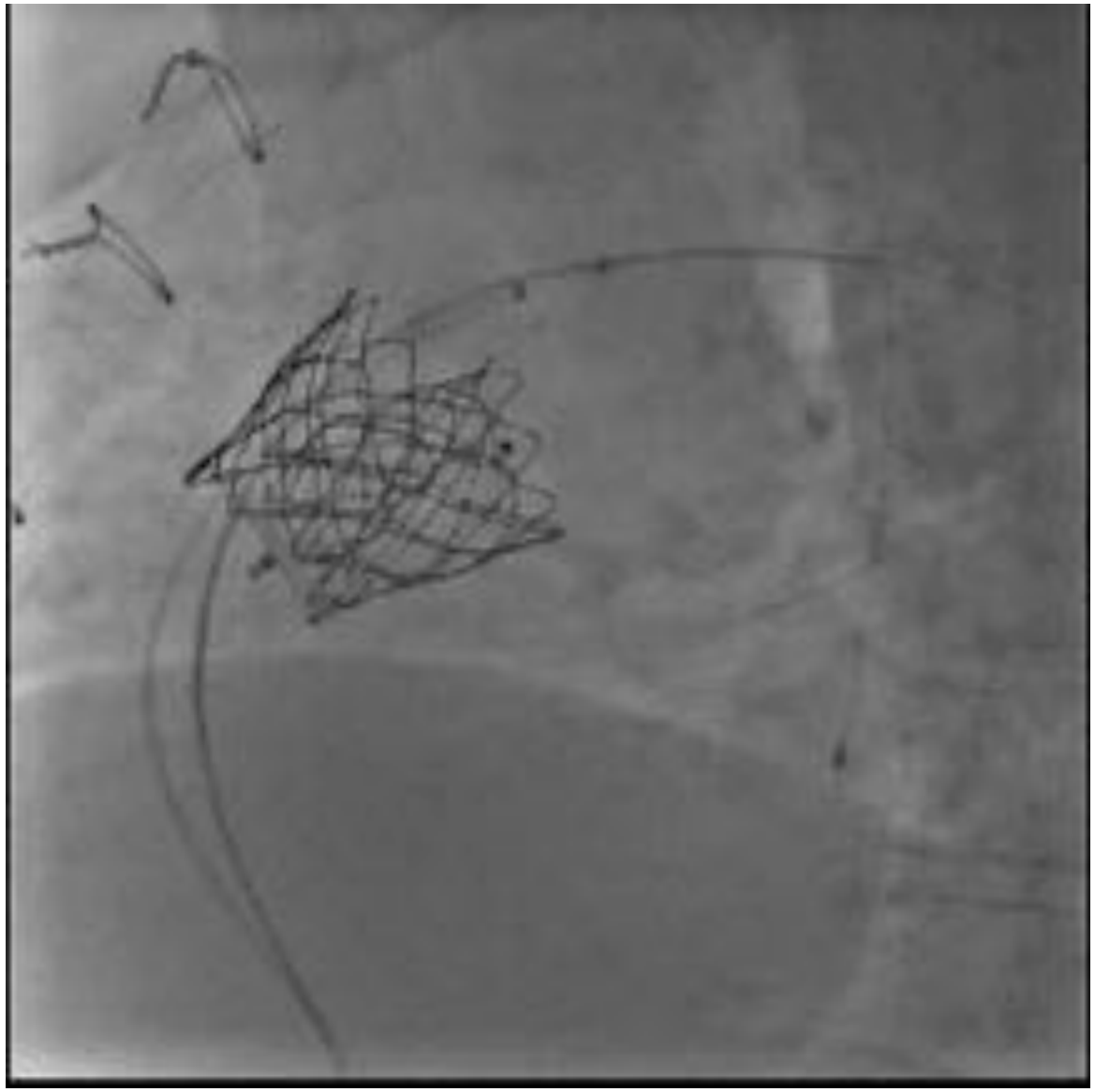


SIDE-BY-SIDE TECHNIQUE



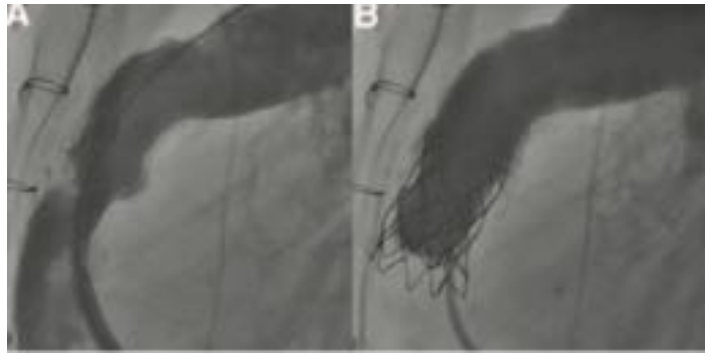
SIDE-BY-SIDE TECHNIQUE





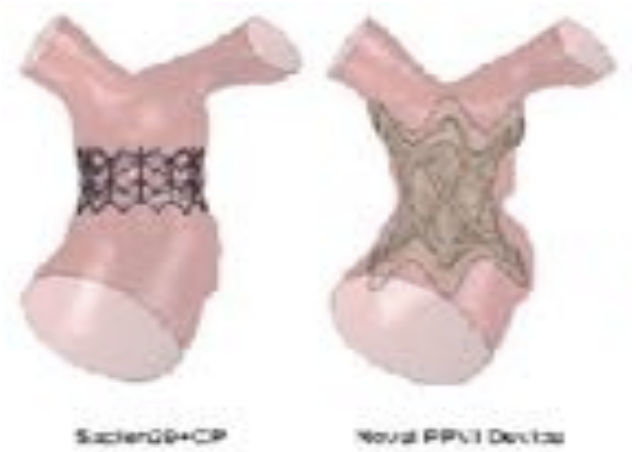
DÉVELOPPEMENT CROISSANT DE LA VALVULATION PERCUTANÉE

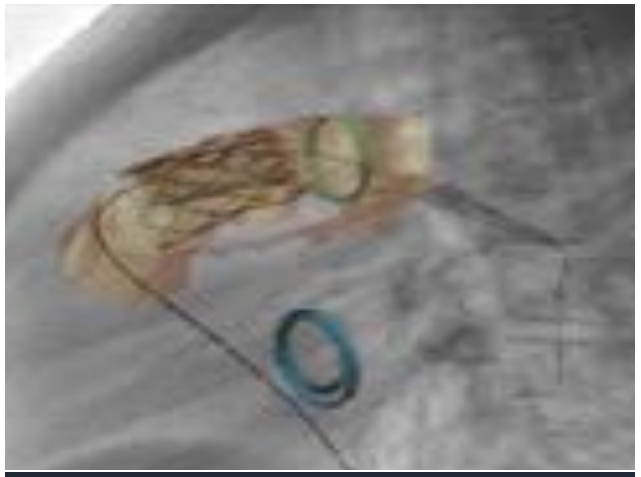
- Efficacité équivalente pour une approche moins invasive
- Indications élargies, limites techniques repoussées
- Futur radieux: nouveaux dispositifs, nouvelles technologies



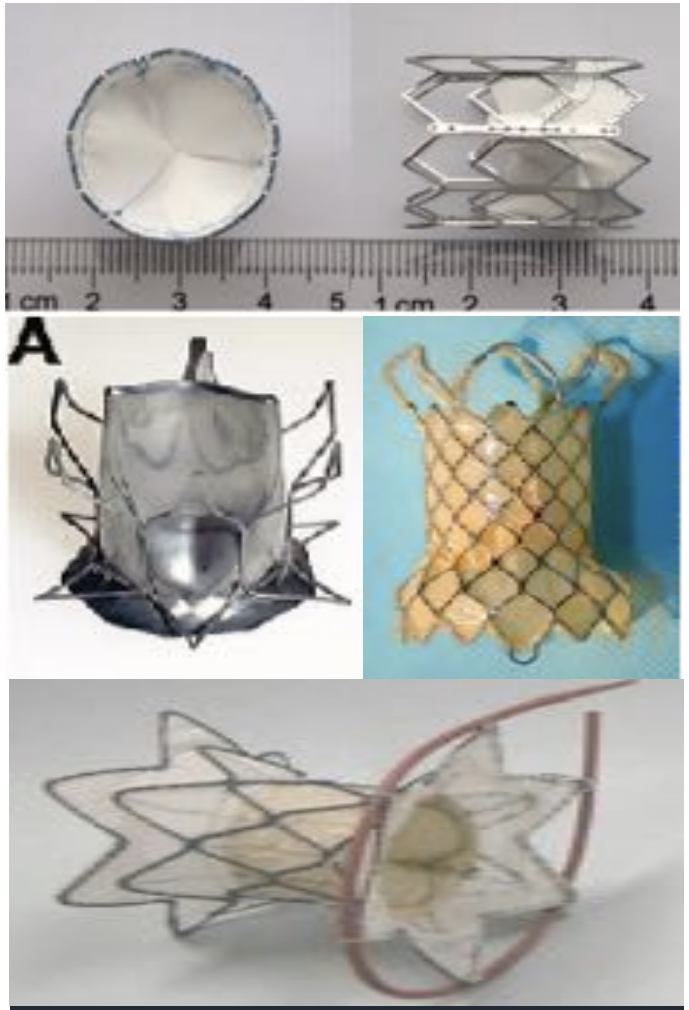
VALVULOPATHIE PULMONAIRE

PERSPECTIVES D'AVENIR





MODELISATION GUIDANCE

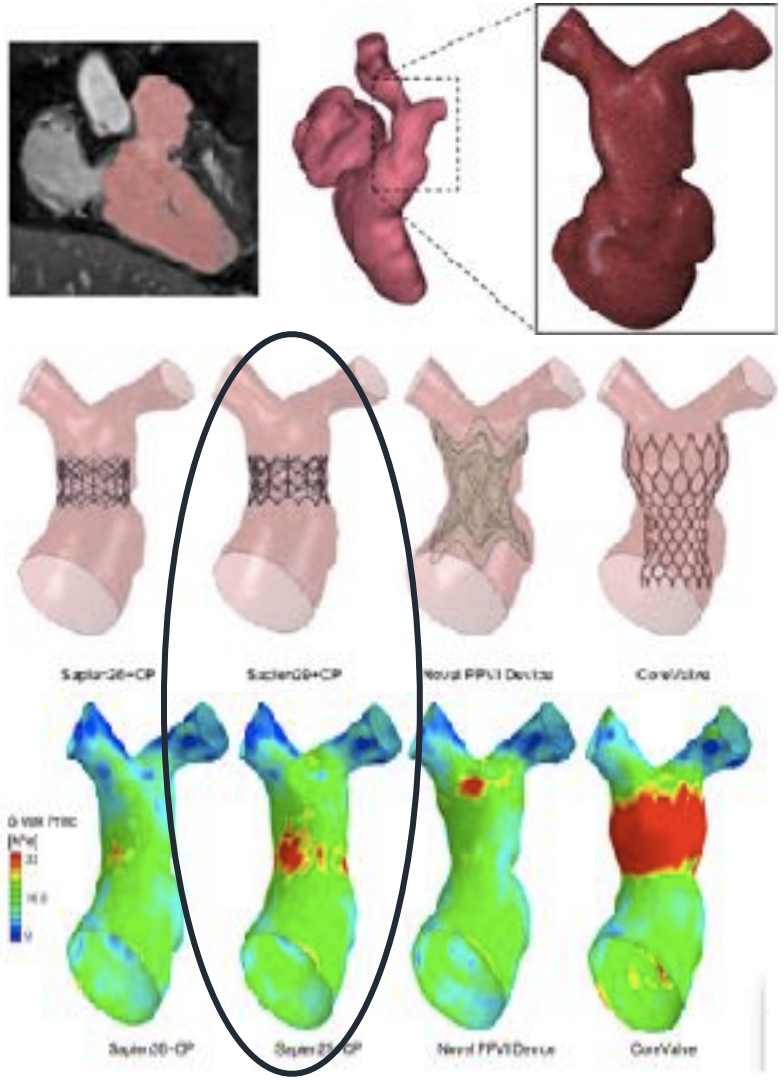


NOUVELLES PROTHESES

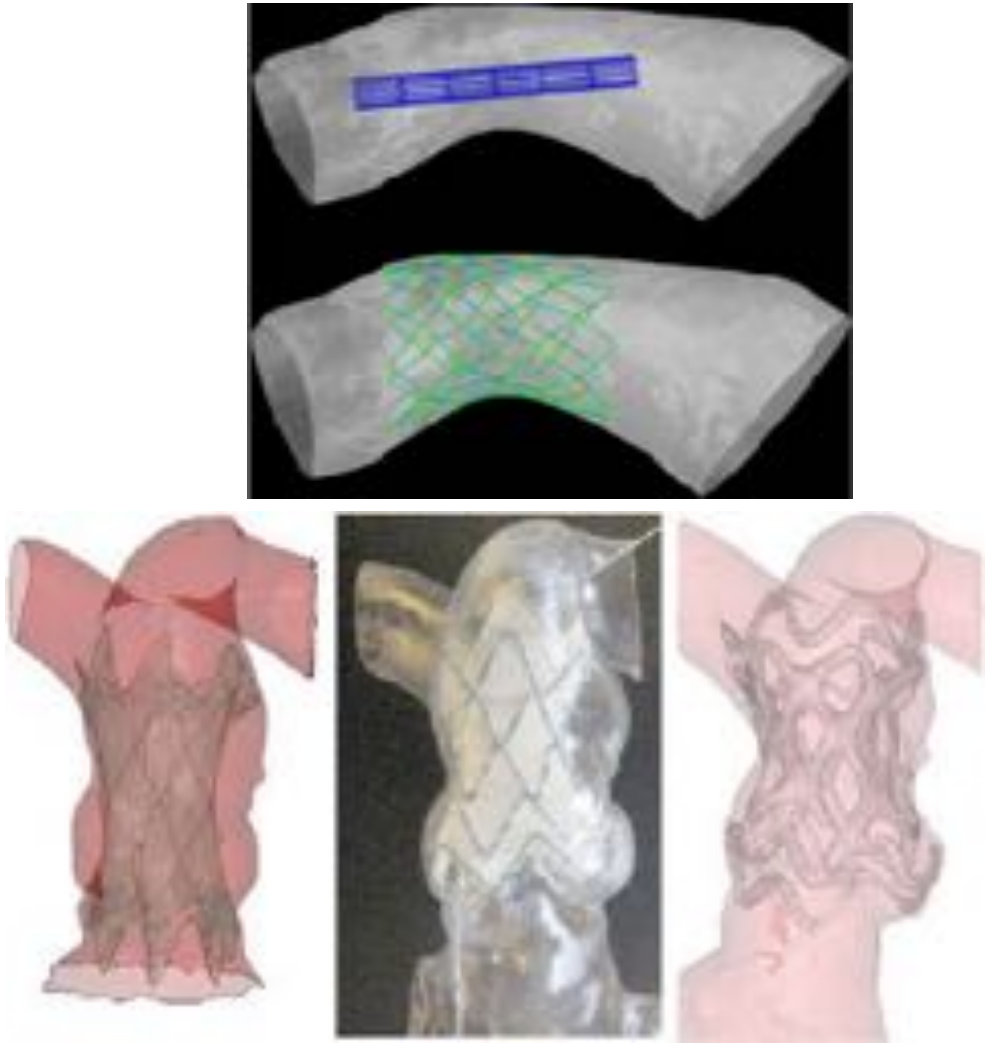
INGÉNIERIE TISSULAIRE

MODELISATION

VALVULOPATHIE PULMONAIRE

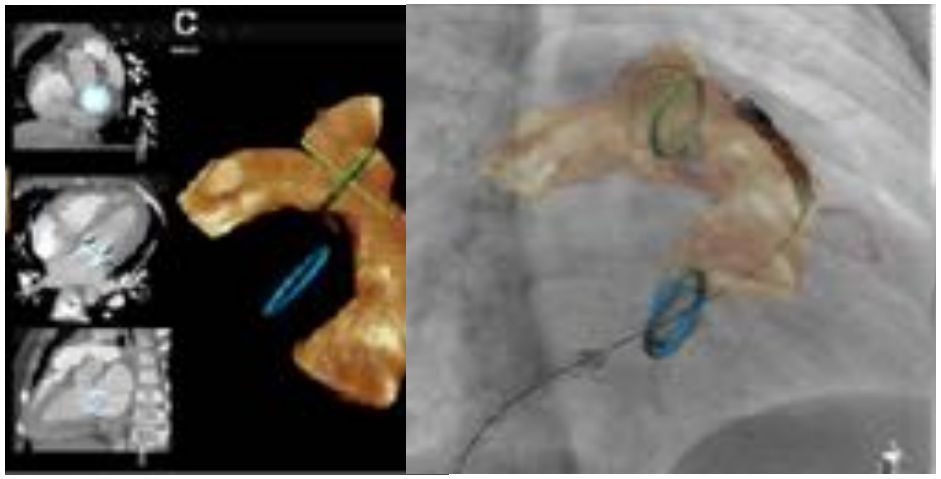


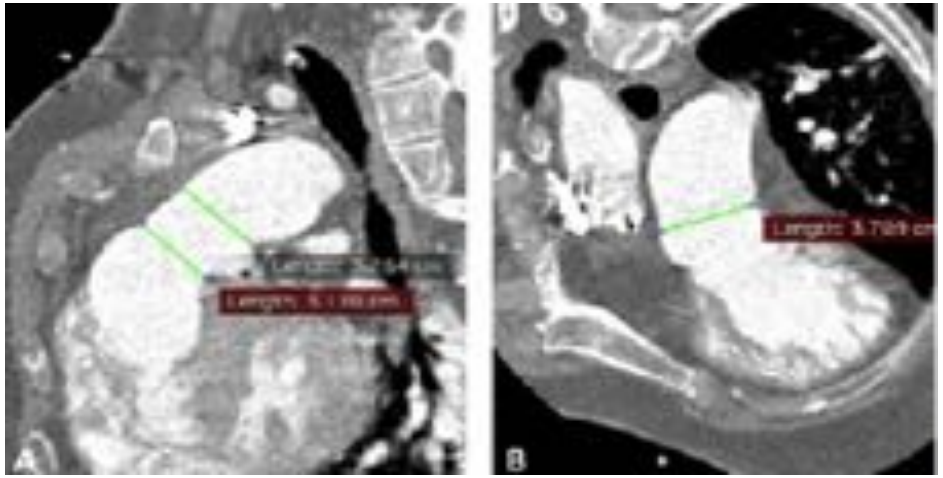
Bosi et al. CCI 2015

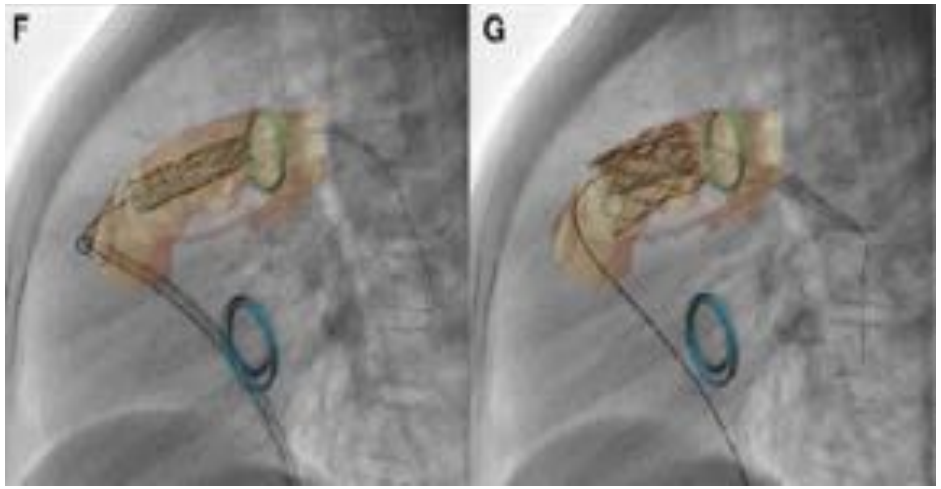


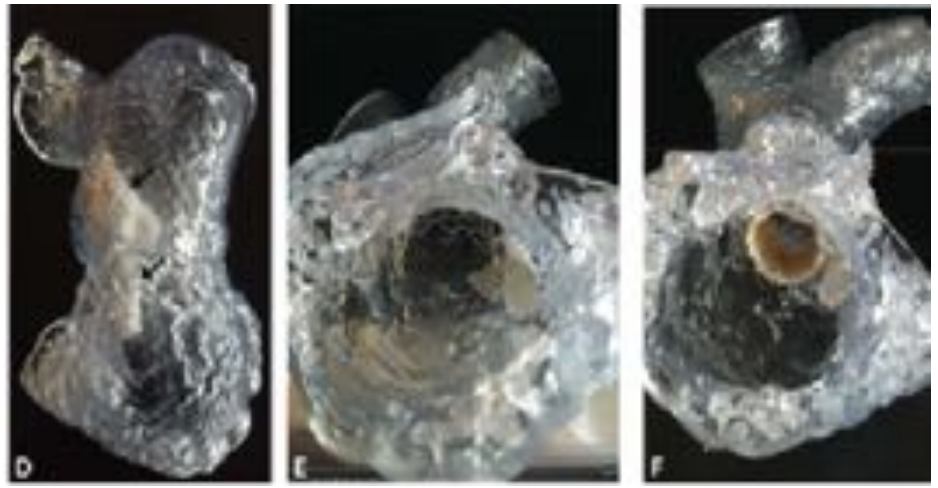
Chung et al. Circ Imaging. 2014

FUSION – IMPRESSION 3D







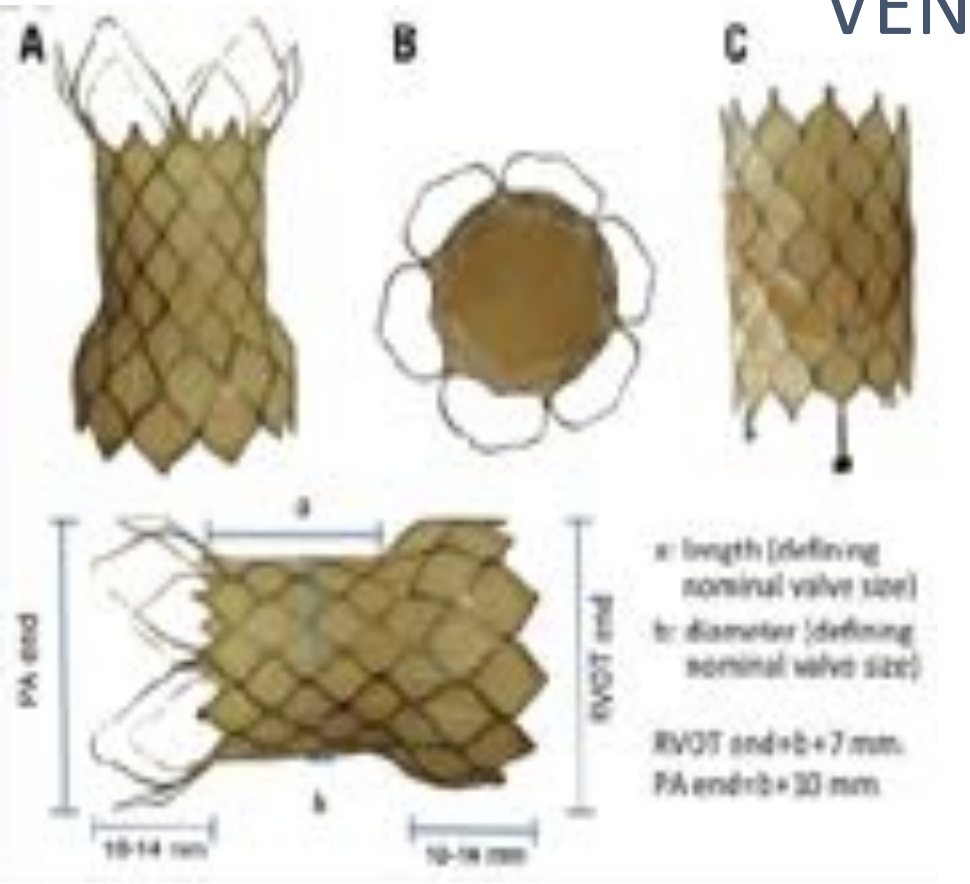


Goreczny et al. Circ Interv 2016

Phillips et al. CCI 2016

VALVULOPATHIE PULMONAIRE

VENUS VALVE



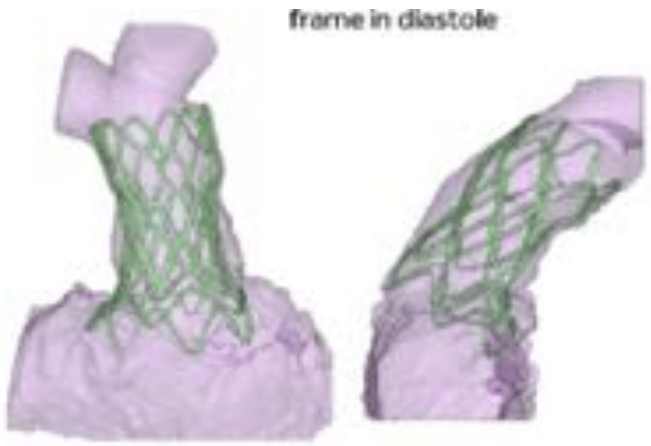
- Stent en Nitinol + Péricarde porcin
- 2 formes « flaired » et « straight »
- Tailles:
  - ⦸ = 16 – 32mm (2)
  - L = 20-35mm (5)
- Système de largage: 14 - 22 Fr

- **17 pts** (12 – 62 ans)
- **100% succès implantation**
- **Suivi 6 mois**
  - Bonne fonction valvulaire
  - Pas de complications

Promphan et al. Cardiol Young 2016
Husain et al. EuroIntervention 2016
Cao et al. CCI 2014

HARMONY

- Stent en Nitinol + Péricarde porcin
- Taille:24mm
- Système de largage: 25Fr

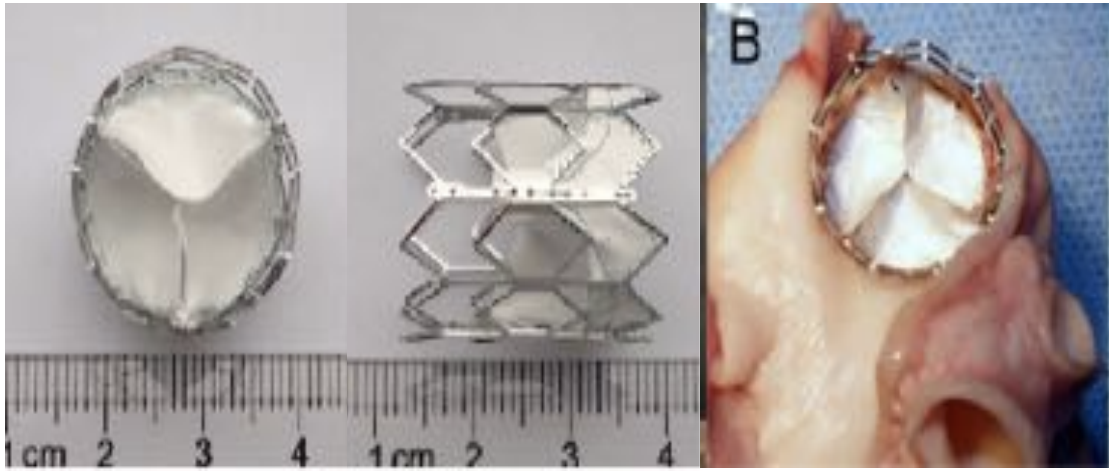


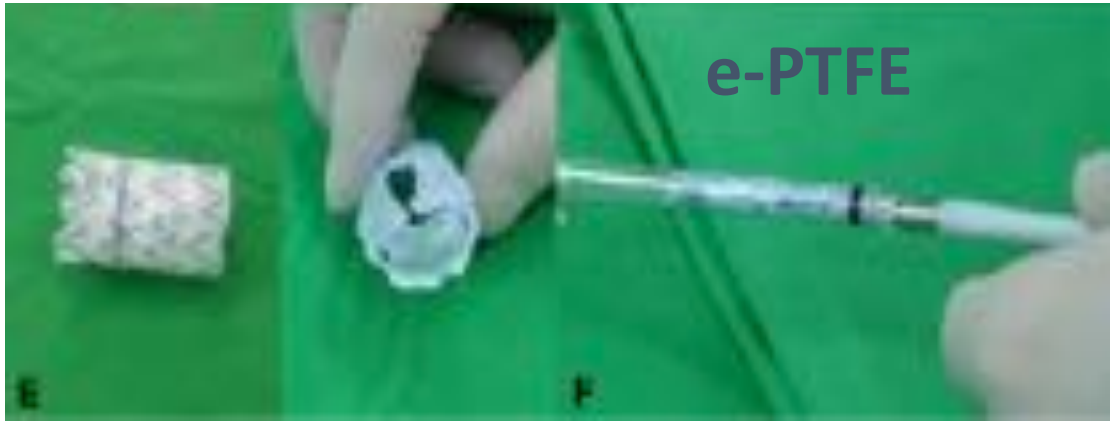
- **21 pts (29 ± 4 ans)**
- **95% succès implantation**
- **Suivi 1 mois**
  - Bonne fonction valvulaire
  - 4 patients avec arythmies ventriculaires

Bergensen et al. JACC Intrv 2017

VALVULOPATHIE PULMONAIRE

VALVES POLYMERIQUES

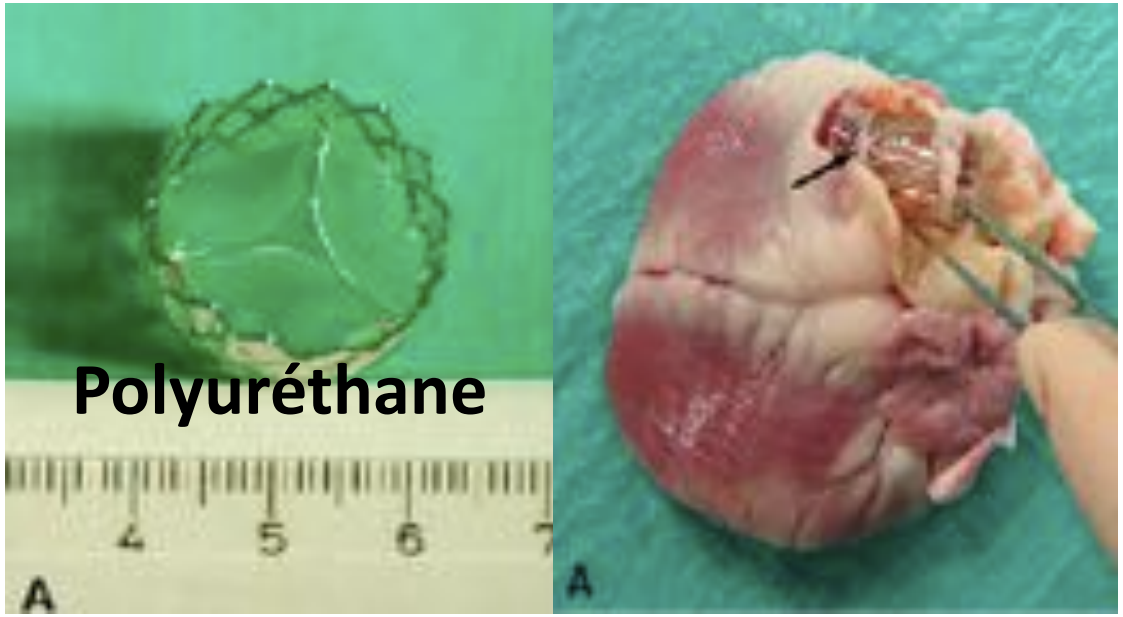




Lin et al. JTCS 2014
Claiborne et al. Expert Rev Med Devices. 2012

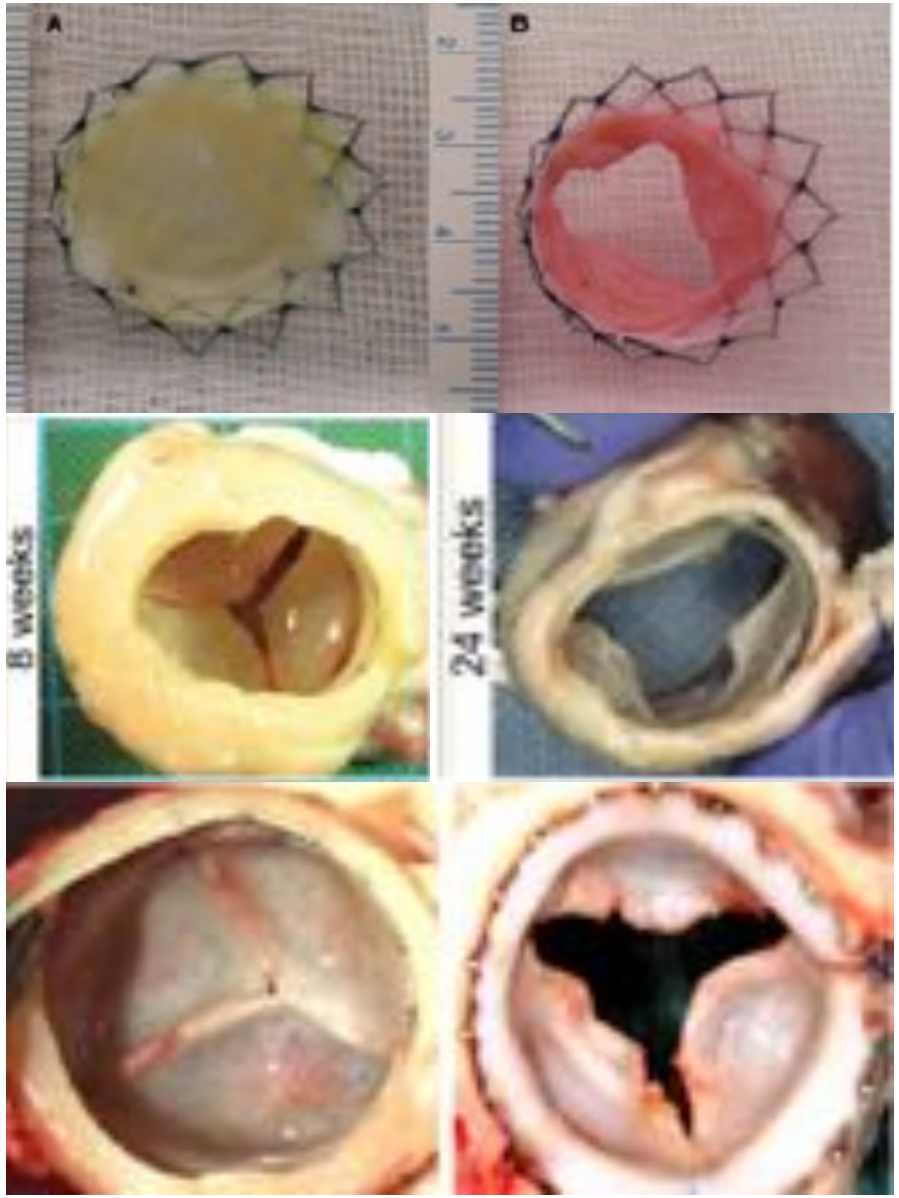
« Valves Biomécaniques »

- Durabilité des valves mécaniques mais pas d'AC
- Faisabilité/Court terme: satisfaisant
- Moyen terme: durabilité décevante



Zhang et al. PLoS ONE 2014
Metzner et al. JTCS 2010

VALVULOPATHIE PULMONAIRE



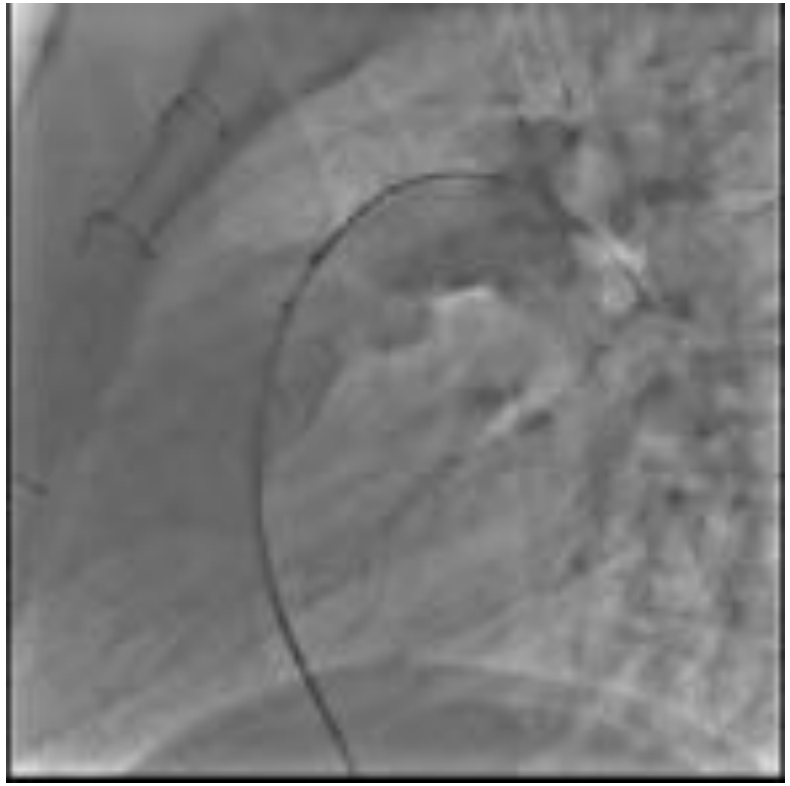
BIOENGINEERING

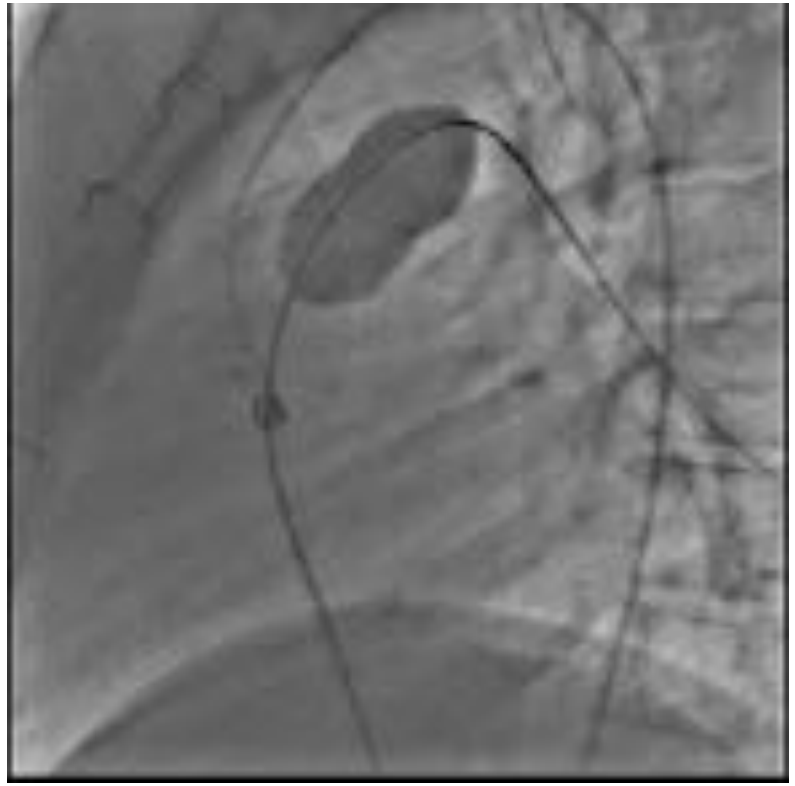
- <u>Cellules autologues ➔scaffold décellularisé</u>
  - valve porcine
  - polymère dégradable
- <u>Avantages théoriques:</u>
  - Biocompatibilité
  - Durabilité
  - Pas de thrombogénicité
- Bonne fonction initiale, endothélialisation
- Rétraction des feuillets ➔Fuite

Metzner et al. Eur. Heart J 2010
Driessen-Mol et al. JACC 2014
Schmitt et al. EuroIntervention 2016

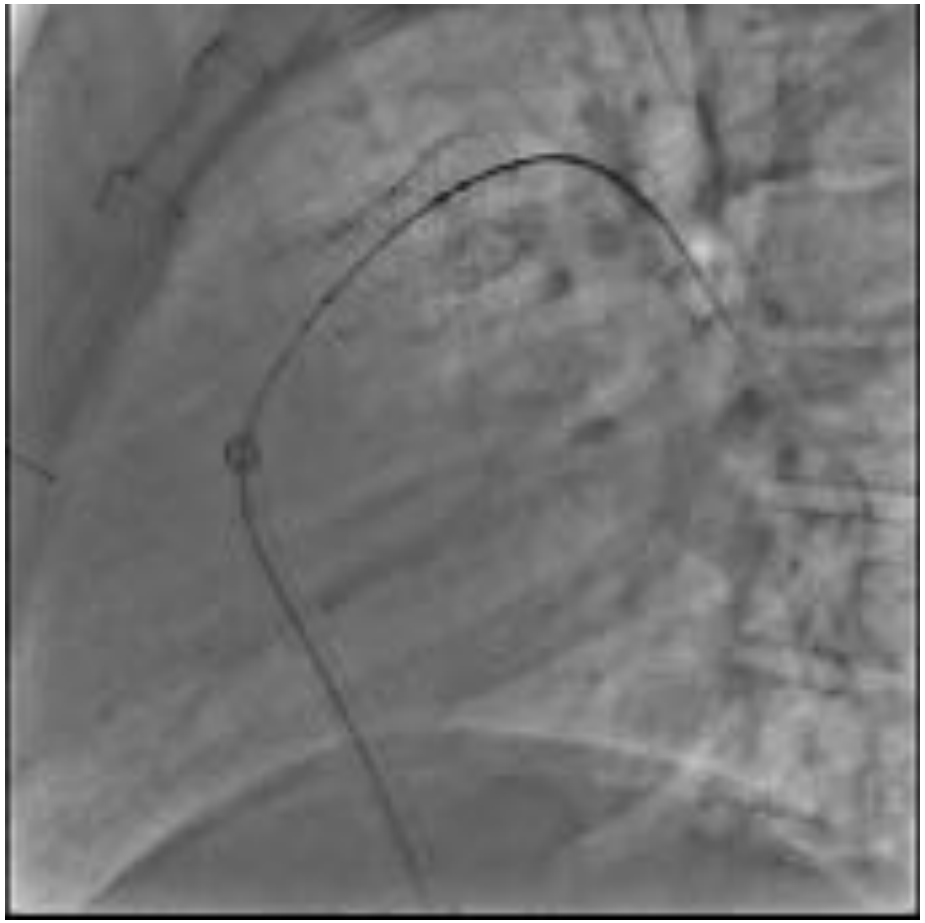
CAS CLINIQUE

- Jeune fille de 16 ans, ASPO multi opérée, Voie VP-AP patchée, Lésion mixte
- EEP pré-valvulation: Déclenchement d’une TV polymorphe rapide de 9s de rduction spontanée



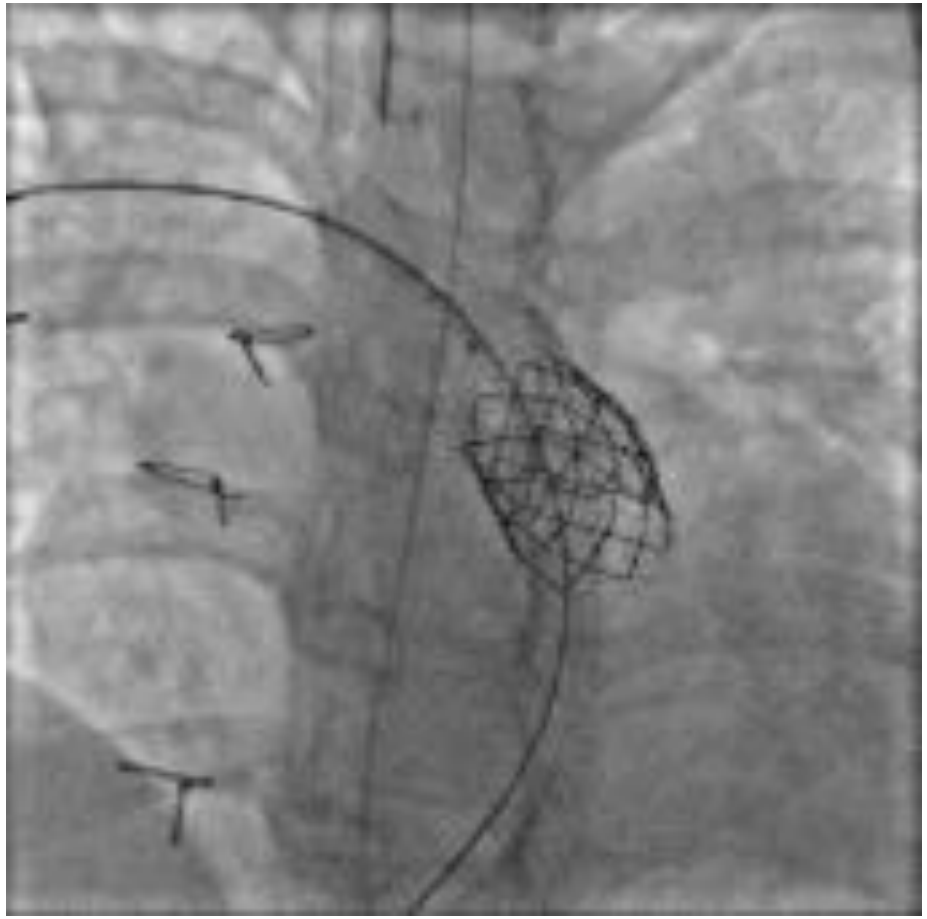


CAS CLINIQUE



Prestenting: EV3 36-mm sur BIB 22x40-mm

CAS CLINIQUE



Melody Ensemble 22mm

CAS CLINIQUE

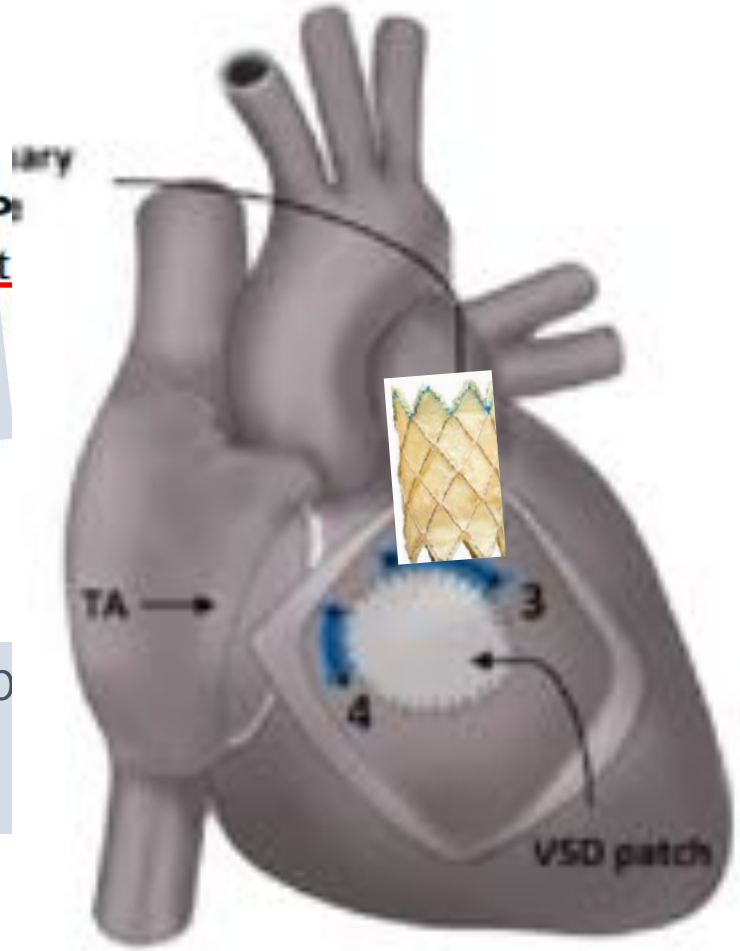
- M3: TV monomorphe à 210/min, origine RVOT, moyennement tolérée
- CEE, transfert à Bordeaux

Au total : non inductibilité en début de procédure malgré l'ISUP
polymorphe. Réalisation d'une ligne entre l'artère pulmonaire et
se trouve sous la MELODY et n'est pas
soit bloquée. En
ient résolu
nplanter un

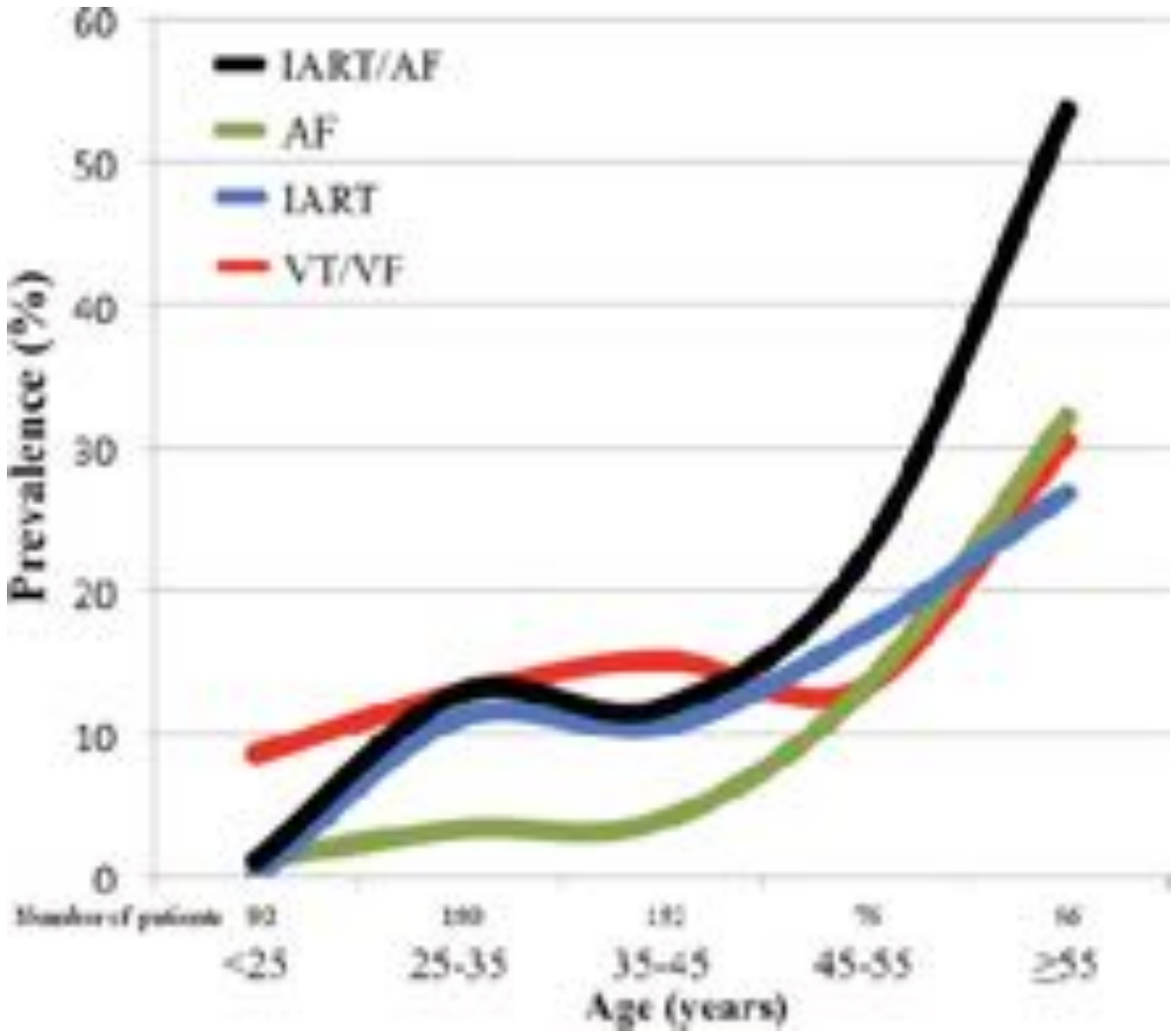
EFFET PROARRYTHMOGÈNE DU(DES) STENT?

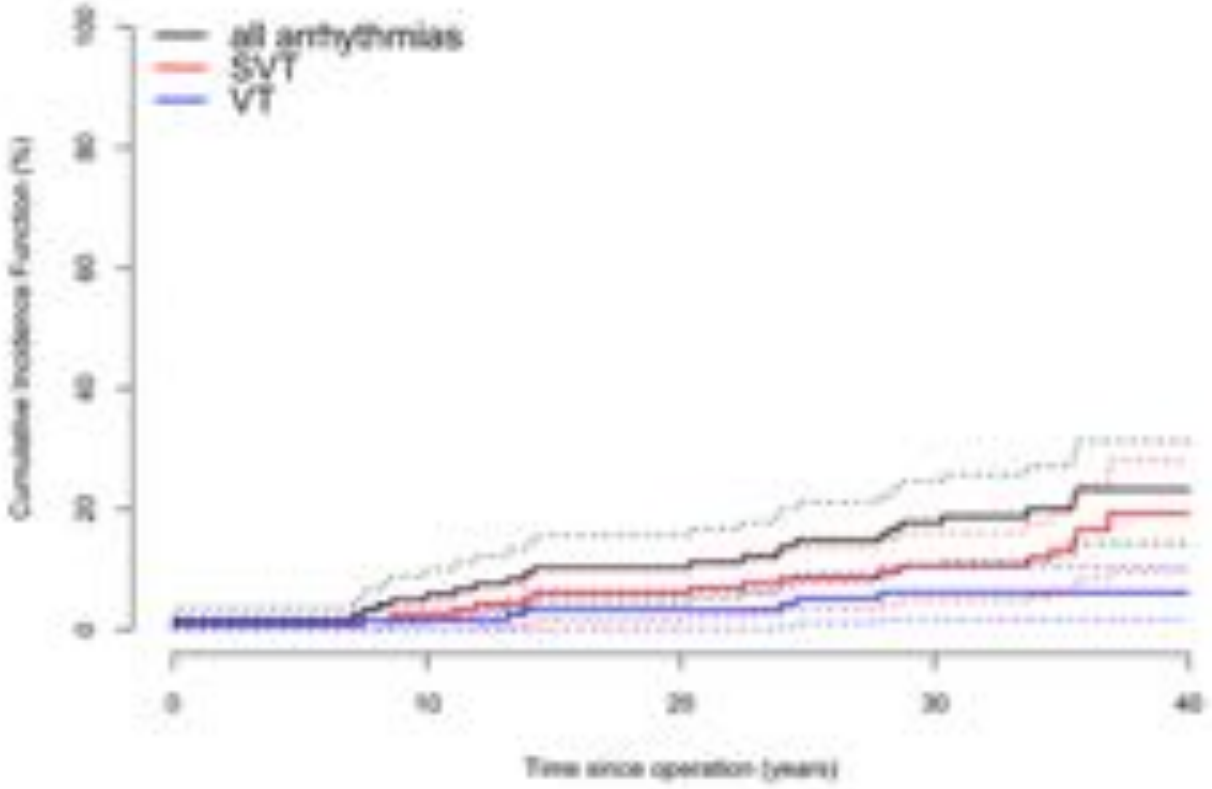
M6: CEE approprié pour TV monomorp

Renforcement du traitement médical



ARYTHMIES – MORT SUBITE

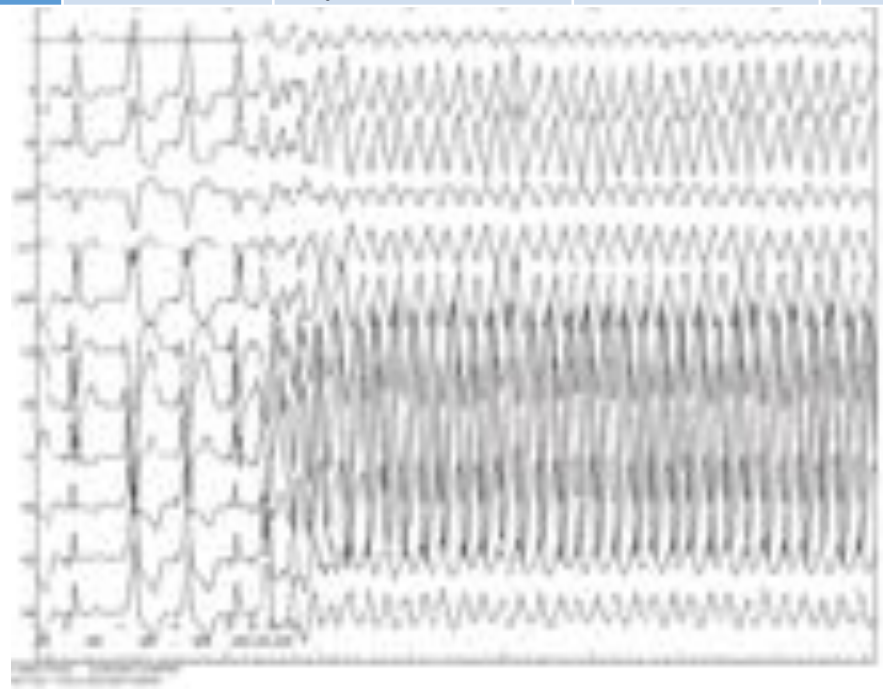




Khairy et al. Circulation 2010
Cuypers et al. Circulation 2014

ARYTHMIES – MORT SUBITE

Authors	patients	Mean follow-up	SCD	SCD incidence per decade
Murphy et al.[2]	163	30years	6%	2.0%
Nollert et al.[3]	490	25years	3%	1.2%
Silka et al.[7]	445	22years	2.6%	1.8%
Norrgaard et al.[4]	125	25years	5.6%	2.2%
Gatzoulis et al.[8]	793	21years	6%	3.0%



INCIDENCE CUMULÉE MORT SUBITE: 1–6%

Buber et al. Heart 2018

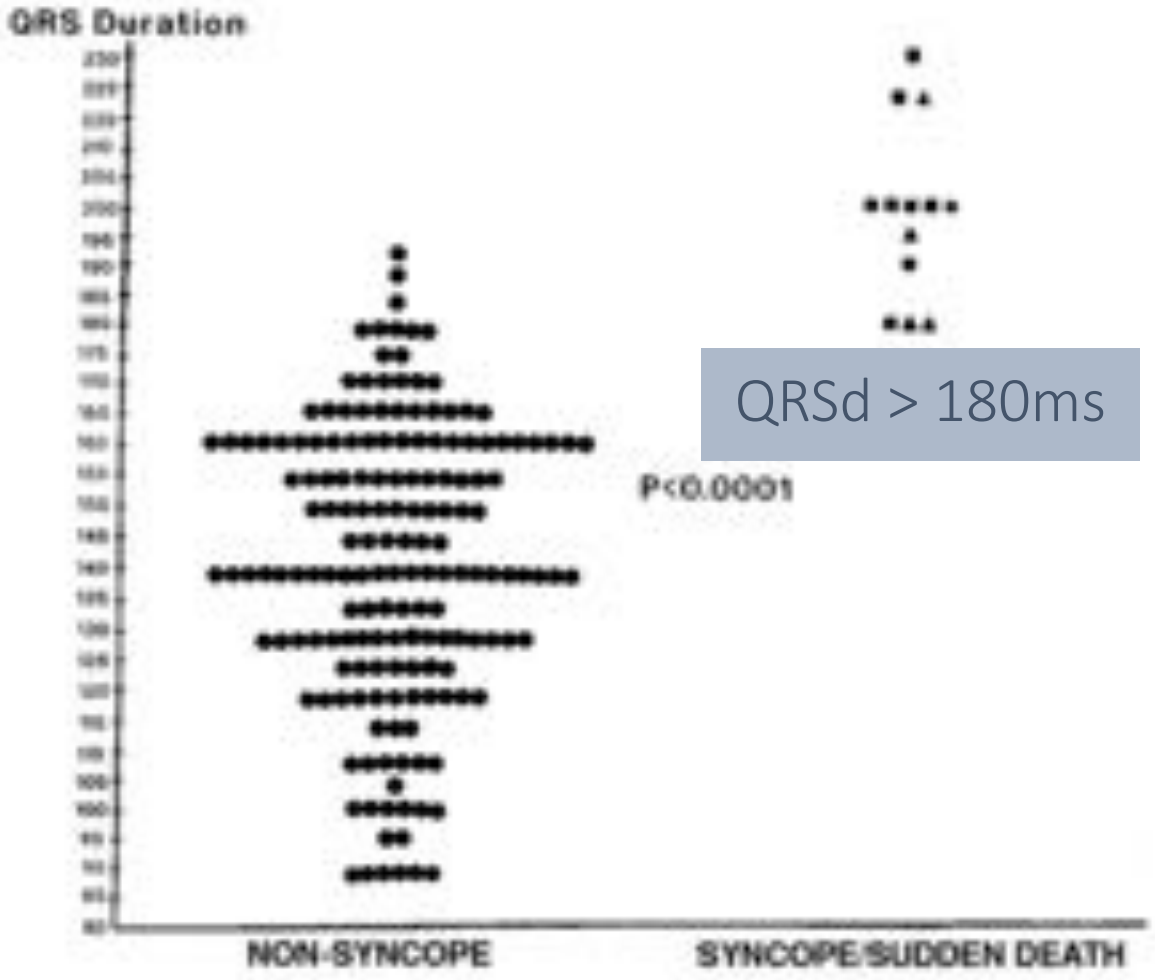
ARYTHMIES – MORT SUBITE

FACTEURS DE RISQUE

Table 5. Individual and Multivariable Factors Associated With Ventricular Tachyarrhythmias

Variable	OR	95% CI	P
Individual analysis			
LV ejection fraction, %	0.95	0.93–0.98	<0.001
Age, y	1.03	1.01–1.05	0.002
Age at repair, y	1.03	1.01–1.05	0.019
Prior cardiac surgeries, n	1.4	1.2–1.8	<0.001
Female sex, %	0.60	0.38–0.97	0.037
QRS duration, ms	1.02	1.01–1.03	<0.001
Hypertension, %	2.3	1.2–4.1	0.009
Diabetes mellitus, %	3.7	1.4–9.8	0.008
At least moderate RV dysfunction, %	1.9	1.1–3.5	0.025
RV outflow tract diameter, mm	1.35	1.02–1.79	0.034
At least moderate tricuspid regurgitation, %	2.2	1.3–3.9	0.005
Right atrial enlargement, %	2.0	1.2–3.3	0.009
Diastolic dysfunction, %	3.9	2.5–7.7	<0.001
LV end-diastolic dimension, mm	1.07	1.03–1.11	<0.001
LV end-systolic dimension, mm	1.08	1.04–1.11	<0.001
Left atrial enlargement, %	[illegible]	[illegible]	[illegible]
Multivariable analysis			
Prior cardiac surgeries, n	1.3	1.1–1.8	0.005
QRS duration, ms	1.02	1.01–1.03	0.014
LV diastolic dysfunction, %	3.3	1.5–7.1	0.002

Khairy P et al. Circulation 2010



QRSd > 180ms

Gatzoulis et al. Circulation 1995

ARYTHMIES – MORT SUBITE

FACTEURS DE RISQUE

Table 3. Risk Score for Appropriate ICD Shocks in Primary Prevention

Variable	Exp(β)	Points Attributed
Prior palliative shunt	3.2	2
Inducible sustained ventricular tachycardia	2.6	2
QRS duration ≥180 ms	1.4	1
Ventriculotomy incision	3.4	2
Nonsustained ventricular tachycardia	3.7	2
LVEDP ≥12 mm Hg	4.9	3
Total points	. . .	0–12

Table 5. Predictors of the Clinical End Points All-Cause Mortality, Arrhythmias/Pacemaker Implantation, and PVR

	Univariate Model			Multivariate Model		
End Point	HR	CI	P Value	HR	CI	P Value
Arrhythmias/PM implantation						
Early postoperative weight gain	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]
Temperature during surgery (per 1°C decrease)	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]
Prosthetic shunt	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]
Transmural patch	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]
Age at operation	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]
Operated after 1991	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]	[illegible]

STRATIFICATION DIFFICILE

Khairy P et al. Circulation 2008
Cuypers et al. Circulation 2014

DURÉE DU QRS

Postoperative Right Bundle Branch Block: Identification of Three Levels of Block

LEONARD N. HOROWITZ, M.D., JAMES A. ALEXANDER, M.D., AND L. HENRY EDMUNDS, JR., M.D.

SUMMARY It has been postulated that postoperative right bundle branch block (RBBB) may be produced by conduction block at any of several sites. In this study the site of block and resultant pattern of ventricular activation were documented in 20 patients in whom RBBB developed during repair of congenital cardiac defects. Intraoperative epicardial and endocardial mapping and recording from the right ventricular specialized conduction system were performed before and after repair in each patient. In eight patients right bundle branch (RBB) conduction was interrupted proximally in the area of the ventricular septal defect. Right ventricular (RV) activation in these patients was delayed at all sites. In five patients RBB conduction was interrupted distally in the area of the moderator band. RV activation in these patients was delayed at most sites; however, the apical septal sites were activated normally. In seven patients, RBB conduction was interrupted terminally in the area of the terminal fascicular network. In these patients RV activation was delayed only in basilar areas. We conclude that at least three distinct types of postoperative RBBB exist and can be identified by differences in RV activation.

Circulation 62, No. 2, 1980.

LÉSION CHIRURGICALE DE LA BRANCHE DROITE

Etiology of Right Bundle-Branch Block in Patients Undergoing Total Correction of Tetralogy of Fallot

By HENRY GELBAND, M.D., ALBERT L. WALDO, M.D., GERARD A. KAISER, M.D., FREDERICK O. BOWMAN, JR., M.D., JAMES R. MALAT, M.D., AND BRIAN F. HOFFMAN, M.D.

SUMMARY

The electrocardiographic (ECG) pattern of right bundle-branch block (RBBB) occurs routinely in patients after open-heart surgery for tetralogy of Fallot (TF). To determine the etiology of the RBBB pattern, 14 patients with TF, seven with ventricular septal defects (VSD), and one with pulmonary stenosis (PS) were studied during and after cardiac surgery. Bipolar electrograms from 10 selected right ventricular epicardial sites were recorded simultaneously with an ECG. Records were obtained before and immediately after vertical right ventriculotomy, after infundibular resection, and after repair of a ventricular septal defect (VSD). The vertical ventriculotomy alone was always associated with significant prolongation of the time of epicardial activation only to the recording sites lateral to the incision with prolongation of the QRS complex by an average of 30 msec, and with the appearance of an RBBB ECG pattern. Infundibular resection and VSD repair were not associated with any changes in the electrophysiologic parameters measured. A retrospective analysis of 251 patients with TF, VSD, and PS revealed a 100% incidence of RBBB pattern in the electrocardiograms of only those who had undergone ventriculotomy. It can be concluded that the RBBB pattern seen postoperatively in patients with TF is due to changes in right ventricular activation secondary to the vertical ventriculotomy.

Circulation, Volume XLIV, December 1971

ARYTHMIES – MORT SUBITE

DURÉE DU QRS

Right Ventricular Mechanics and QRS Duration in Patients With Repaired Tetralogy of Fallot

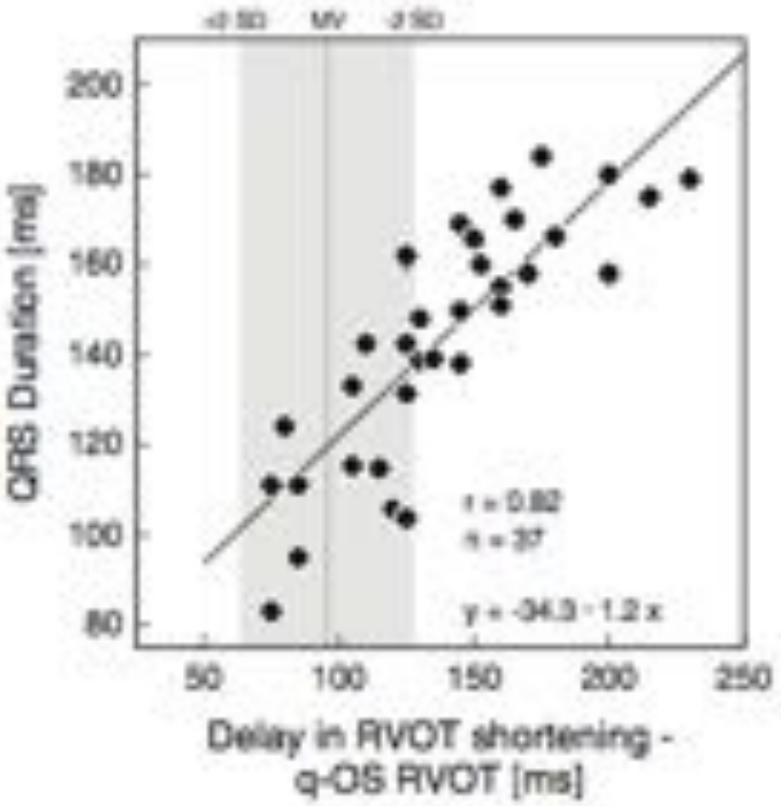
Implications of Infundibular Disease

Anselm Uebing, MD; Derek G. Gibson, MB, FRCP; Sonya V. Babu-Narayan, BSc, MRCP; Gerhard P. Diller, MD; Konstantinos Dimopoulos, MSc, MD; Oener Gokhoglan, MD; Mark S. Spence, MD, MRCP; Kai Andersen, MD, PhD; Michael Y. Henein, MD, PhD; Michael A. Gatzoulis, MD, PhD; Wei Li, MD, PhD

Background—Patients after repair of tetralogy of Fallot (ToF) frequently have right ventricular (RV) dysfunction and prolonged QRS duration (QRSd) and thus could be candidates for cardiac resynchronization therapy. We aimed to assess the relationship between QRSd and the timing of RV wall motion, including the RV outflow tract (RVOT), in these patients.

Methods and Results—Sixty-seven repaired ToF patients (median age, 34 years; interquartile range, 24 to 43 years) and 55 age-matched control subjects were studied by echocardiography and cardiovascular magnetic resonance (n=53 of 60 ToF patients). Time intervals of the RV cardiac cycle were measured from Doppler recordings. Long-axis M-mode recordings were acquired from the right ventricular (RV) free wall and RV outflow tract (RVOT), and the delay in onset of long-axis shortening was measured. ToF patients showed minor abnormalities of the RV cardiac cycle unrelated to QRSd. RV ejection time was prolonged and correspondingly filling time was reduced compared with control subjects (22.3±2.6 versus 20.0±2.9 s/min, $P<0.0001$; 29.0±2.8 versus 32.7±3.5 s/min, $P<0.0001$). Total isovolumic time was normal in ToF patients (8.7±4.0 versus 7.4±2.9 s/min; P=NS). QRSd correlated with the delay in RV free wall motion (r=0.55, $P<0.0001$) and more so with the delay in RVOT shortening (r=0.82, $P<0.0001$). QRSd also correlated with measures of RVOT abnormality such as long-axis RVOT excursion and akinetic area length (r=−0.46, P=0.004; r=0.33, P=0.01).

Conclusions—QRSd in postoperative ToF patients reflects mainly abnormalities of the RVOT rather than the RV body itself. Thus, prevention and treatment of mechanical asynchrony and malignant arrhythmia should focus on the RV infundibulum. Indications for cardiac resynchronization therapy after ToF repair warrant further investigation. (*Circulation*. 2007;116:1532-1539.)



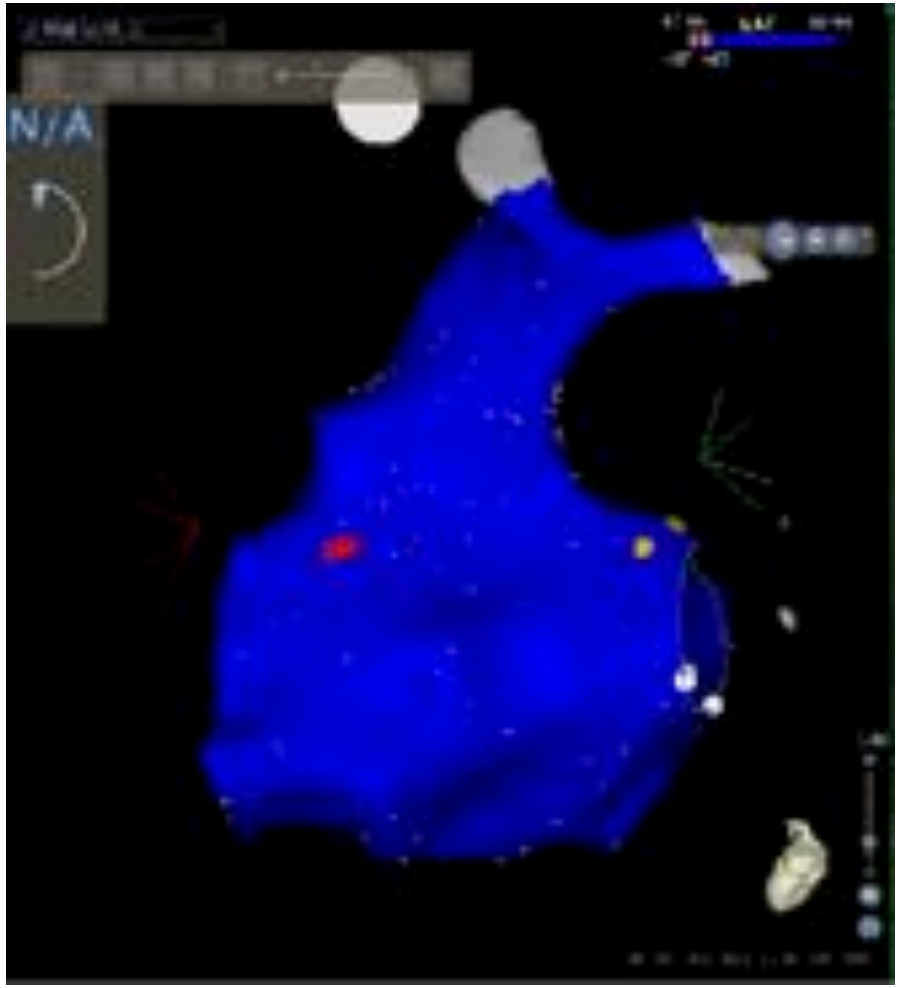
ARYTHMIES – MORT SUBITE

- 56 pts, 2009 - 2016, Prospectif CHU Bordeaux

- Cartographie électro-anatomique invasive
- Activation VD
- Conduction ralentie dans l'ensemble du VD

- Durée d'activation VD
  - Prolongée (médiane=134 ms (118-157))
  - Plus élevée lorsque QRSd ≥ 180 ms
  - Plus élevée lorsque VTDVD > 150mL/m^2
  - Peu corrélée à la durée du QRS (r=0.44; P<0.001)
- Séquence d'activation VD relativement homogène => Paroi libre

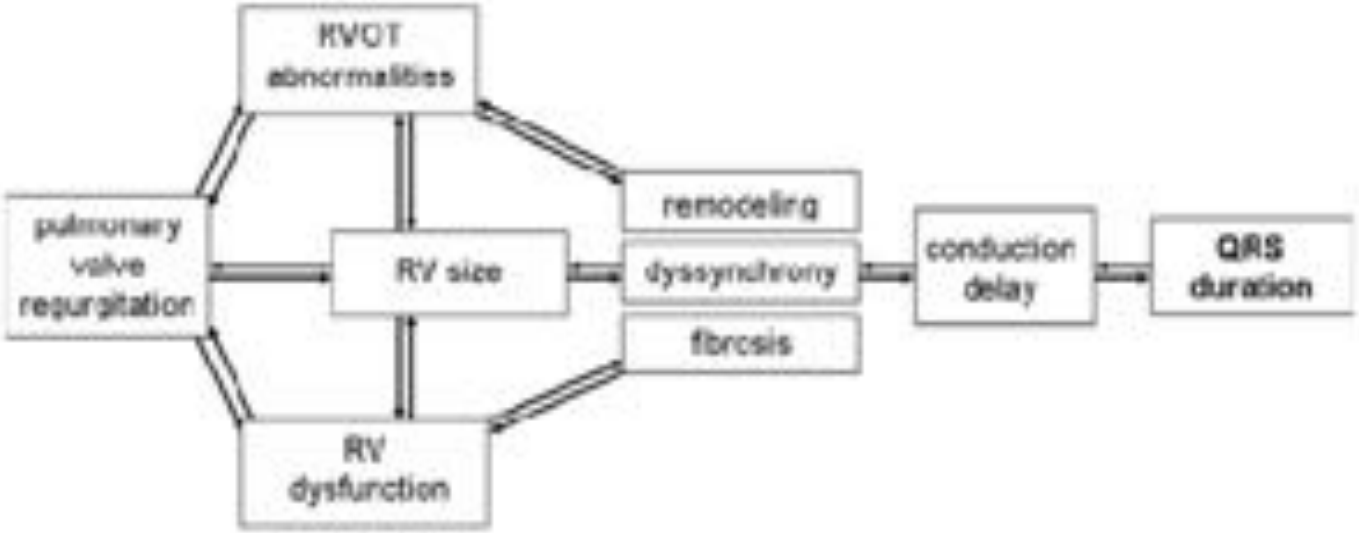
DURÉE DU QRS



Jalal et al. Circ EP 2019

ARYTHMIES – MORT SUBITE

DURÉE DU QRS



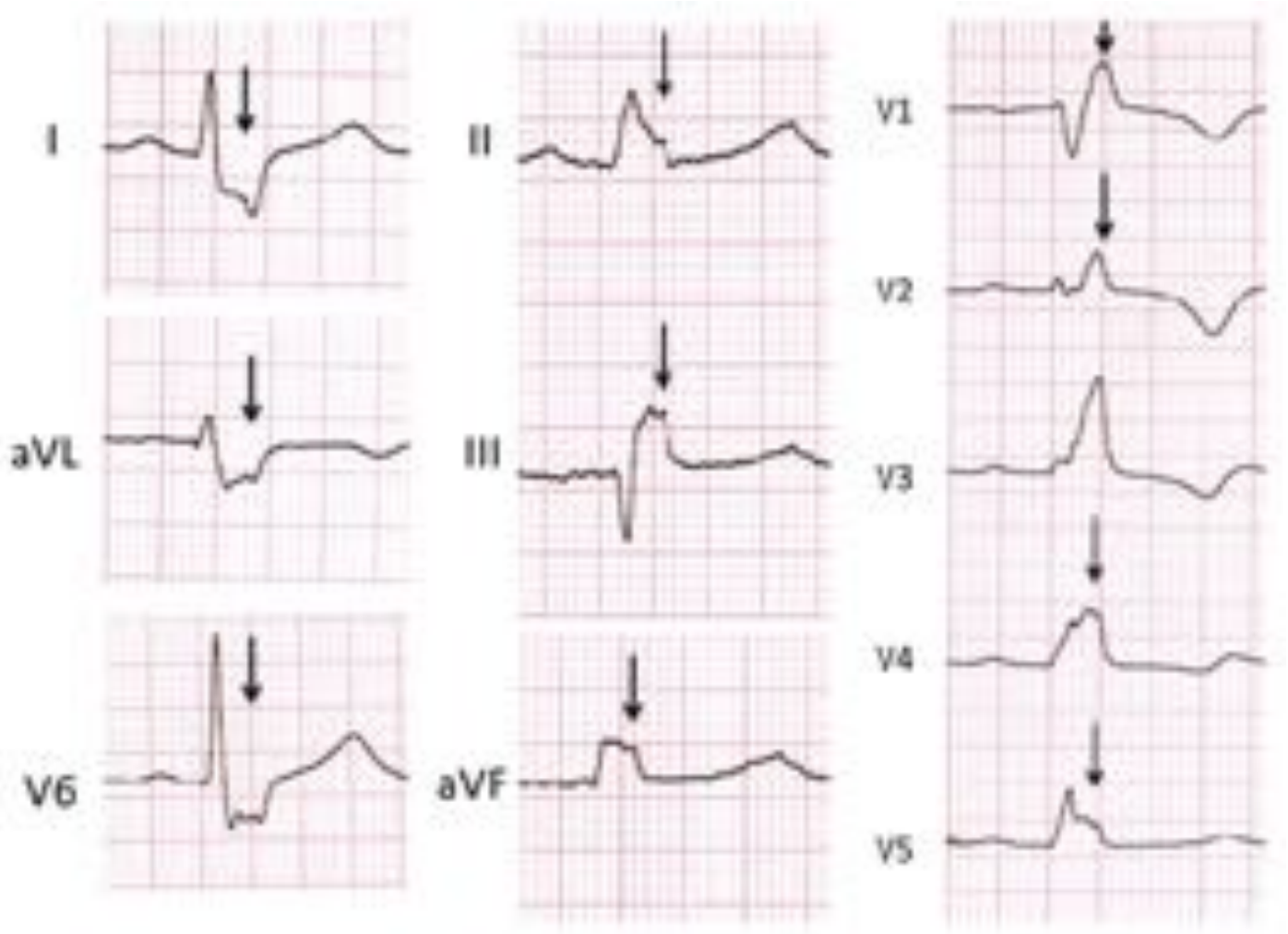
ECG Pré-op et 5 ans Post-op de Fallot

F.E.A. Udink ten Cate et al. Journal of Electrocardiology 2014

DURÉE DU QRS

À ANALYSER SOUS PLUSIEURS ANGLES

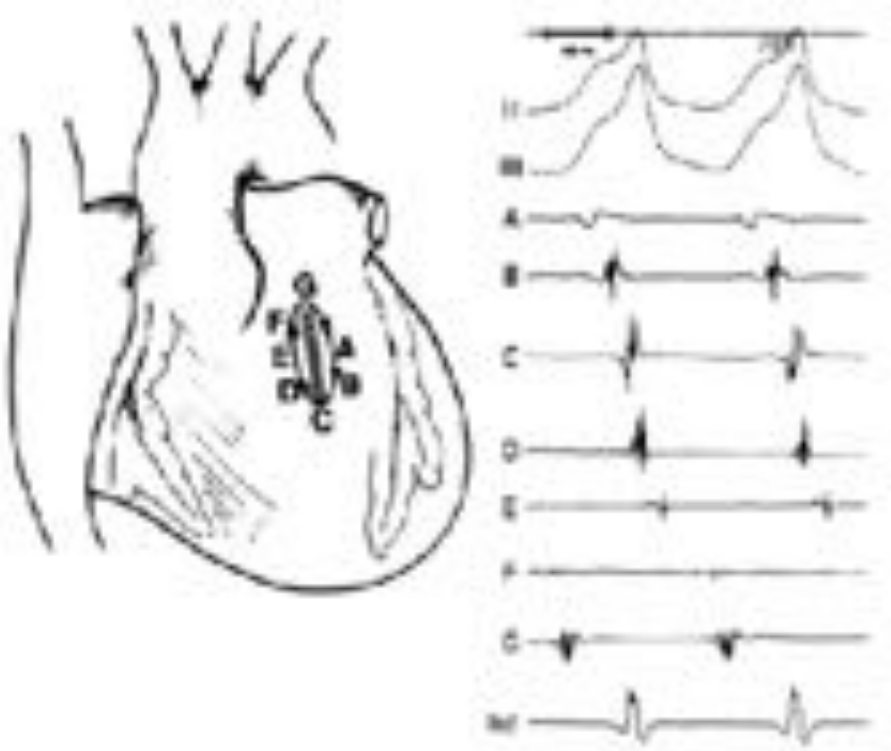
- Elargissement du QRS au cours du temps
- Durée ≥ 180 ms
- Fragmentation du QRS (dérivations Ant, Inf)

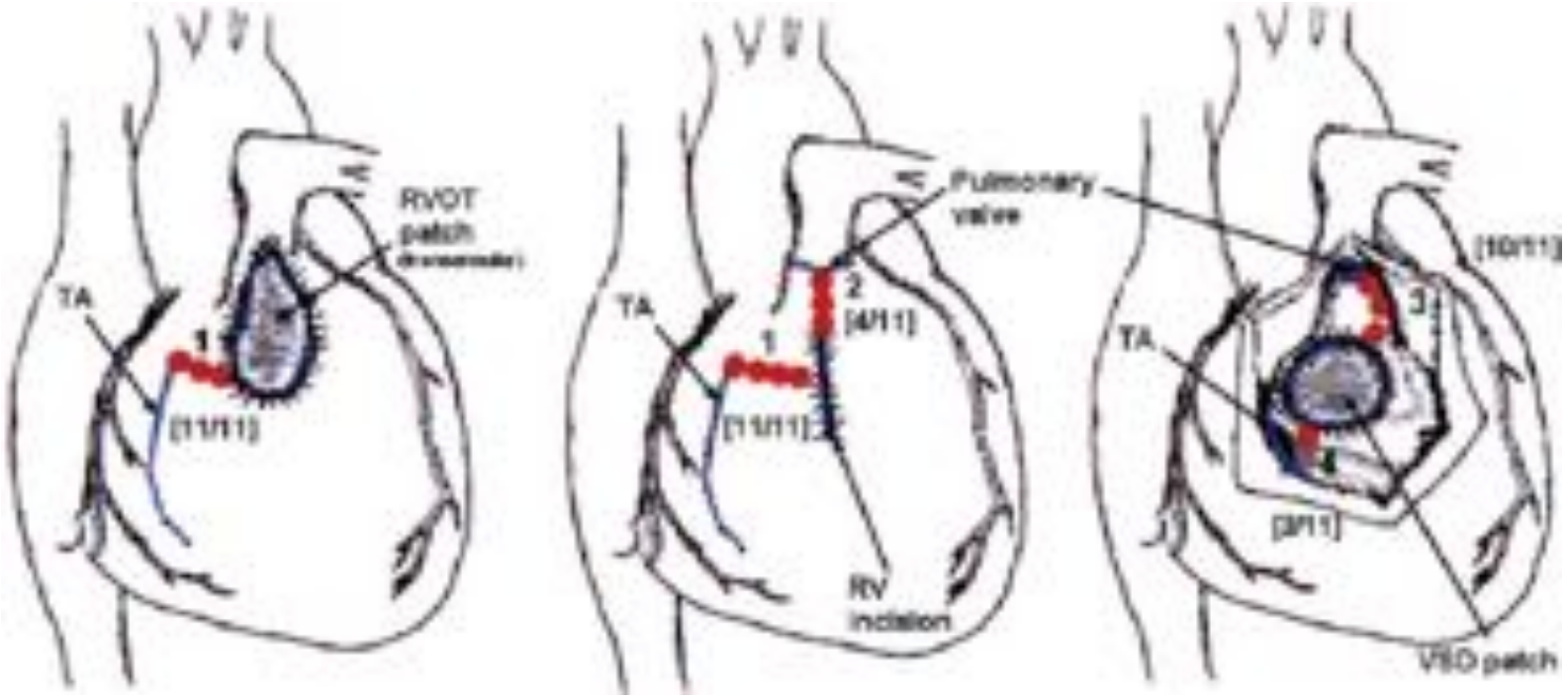


F.E.A. Udink ten Cate et al. Journal of Electrocardiology 2014
Shanmugam et al. Int J Cardiol 2013

ARYTHMIES – MORT SUBITE

TACHYCARDIES VENTRICULAIRES PAR RÉENTRÉE

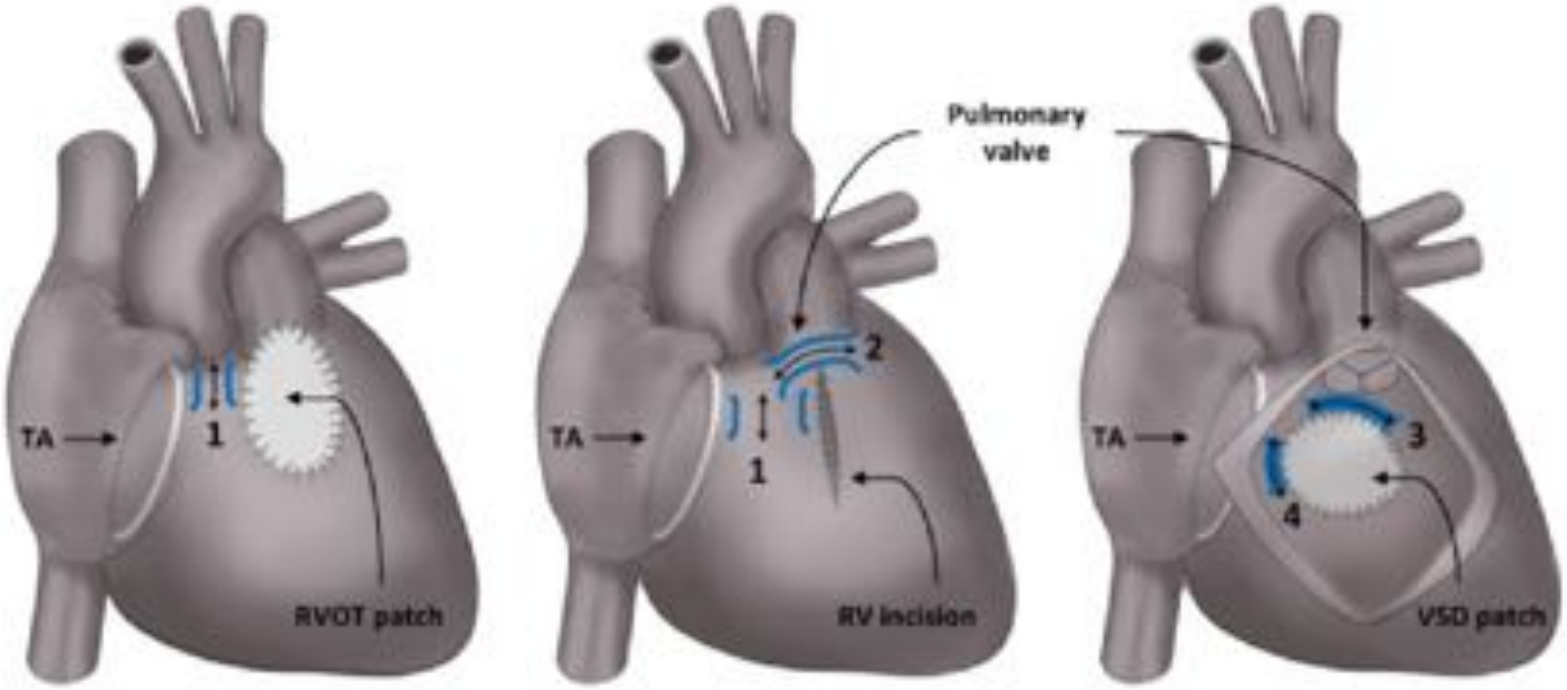


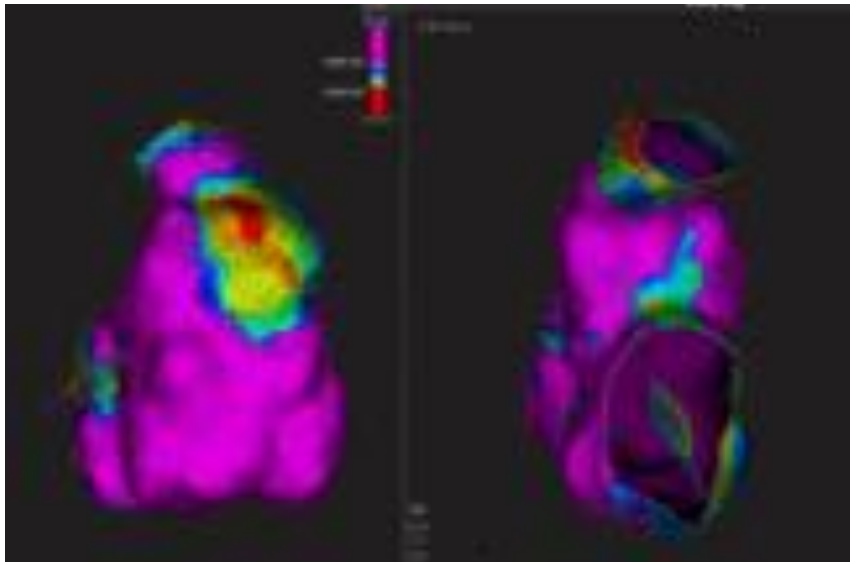


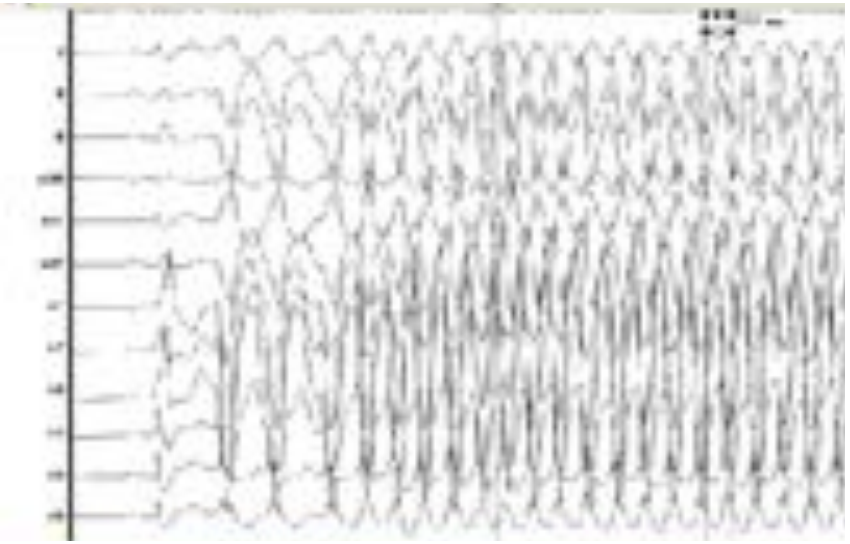
Zeppenfeld K et al. Circulation 2007
Mizaki et al. Circulation. 1994

ARYTHMIES – MORT SUBITE

TACHYCARDIES VENTRICULAIRES PAR RÉENTRÉE







Kapel et al. JACC EP 2018
Kapel et al. Eur Heart Jour 2016

TACHYCARDIES VENTRICULAIRES PAR RÉENTRÉE - RECOMMANDATIONS

IIa	B-NR	3. Programmed ventricular stimulation can be useful to risk-stratify adults with TOF and additional risk factors for SCD.[S4.3.5-3–S4.3.5-8]

7. Risk factors for SCD include:
   a. LV systolic or diastolic dysfunction
   b. Nonsustained VT
   c. QRS duration ≥180 ms
   d. Extensive RV scarring
   e. Inducible sustained VT at electrophysiological study

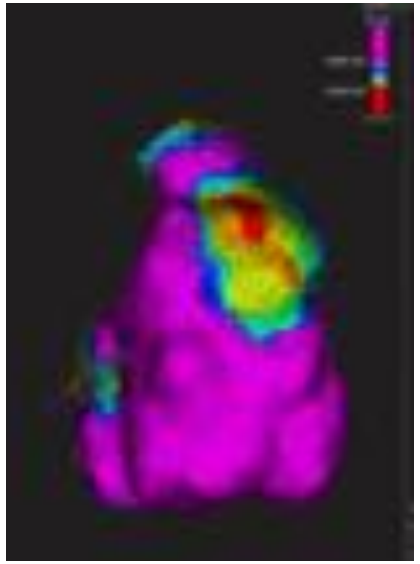
IIa	B-NR	7. Primary prevention ICD therapy is reasonable in adults with TOF and multiple risk factors for SCD.[S4.3.5-15–S4.3.5-17]

TACHYCARDIES VENTRICULAIRES PAR RÉENTRÉE - RECOMMANDATIONS

General Indications for EP and ICD Implantation in GUCH

	Class[a]	Level[b]
• ICD implantation is indicated in survivors of cardiac arrest after exclusion of reversible causes.	I	B
• 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.	I	C
• 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.	IIa	B
• EP testing may be considered for patients with ventricular couplets or non-sustained VT to determine the risk of sustained VT.	IIb	C

PROTOCOLE DE PRISE EN CHARGE



Indication d'ablation

Indication d'un RVP

EEP + Cartographie

Inductible

Non inductible

VALVULATION PERCUTANÉE CONSIDÉRÉE

ABLATION

Succès

Echec

Substrat?
Prestenting RVOT?

DAI

Cryoablation chirurgicale

Choix de la technique de valvulation

DÉCISION AU CAS PAR CAS

PLURIDISCIPLINAIRE (CARDIOPÉDIATRES, CHIRURGIENS, RYTHMOLOGUES)

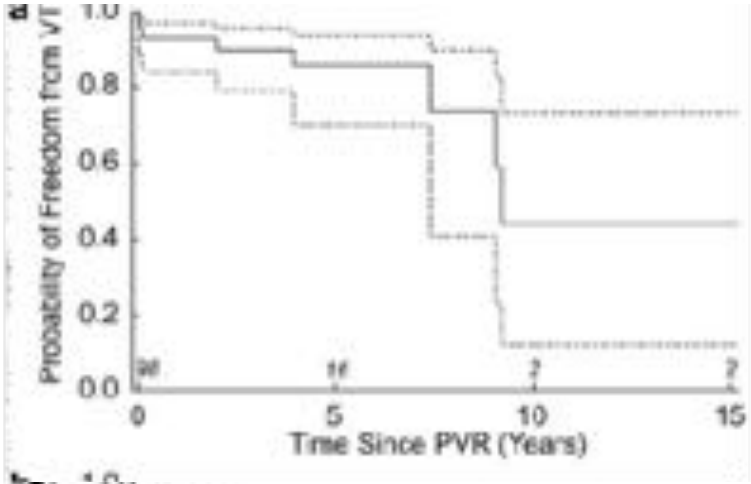
IMPLICATION ET EXPERTISE ÉQUIPE ÉLECTROPHYSIOLOGIE

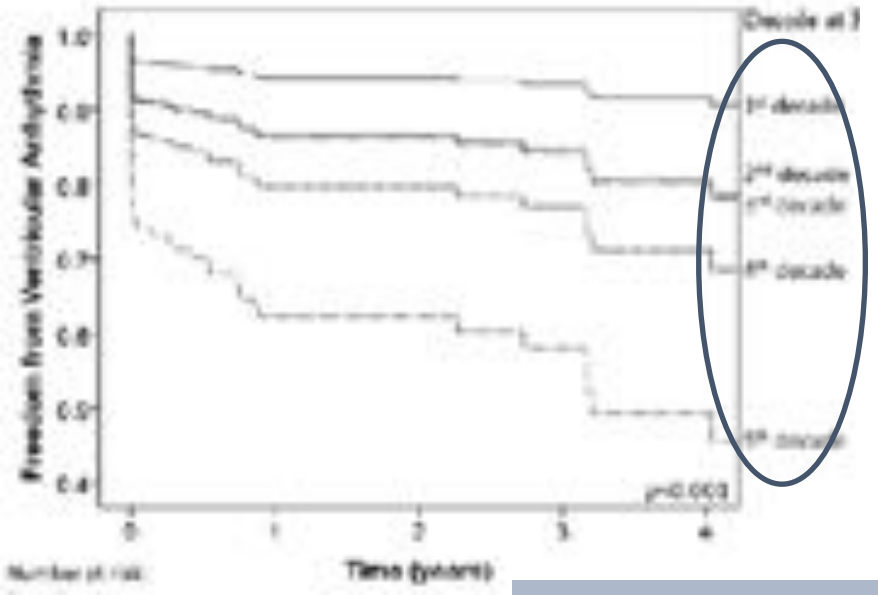
ble

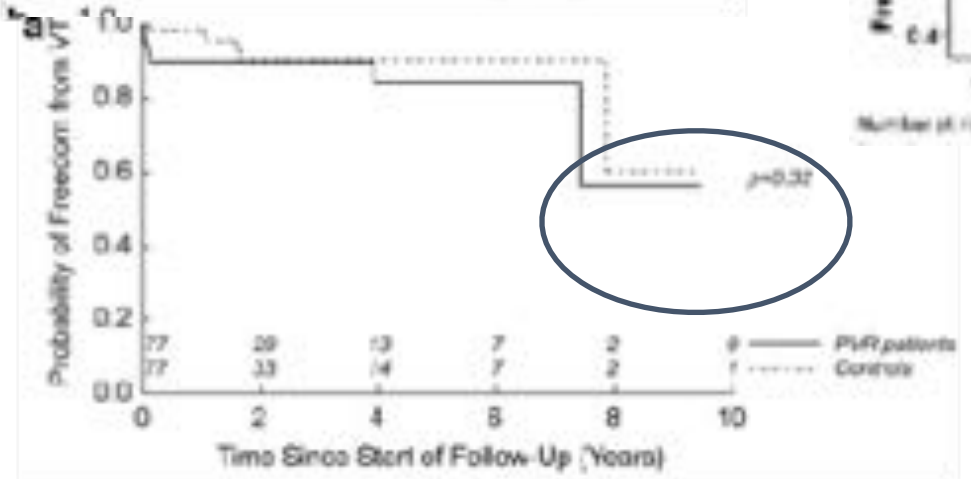
DAI

ARYTHMIES – MORT SUBITE

IMPACT VALVULATION PULMONAIRE







DÉPEND DU TIMING

DÉPEND DE LA TECHNIQUE

REMODELAGE+++

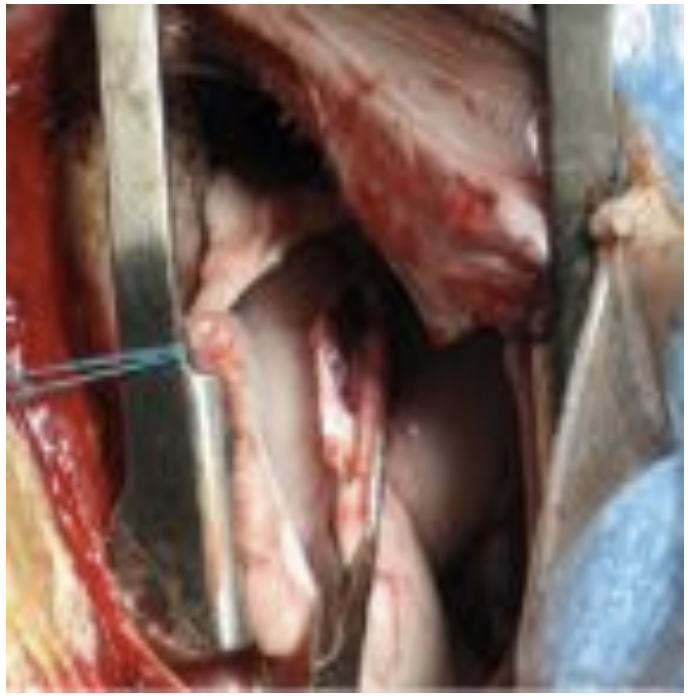
Harrild et al. circulation 2009

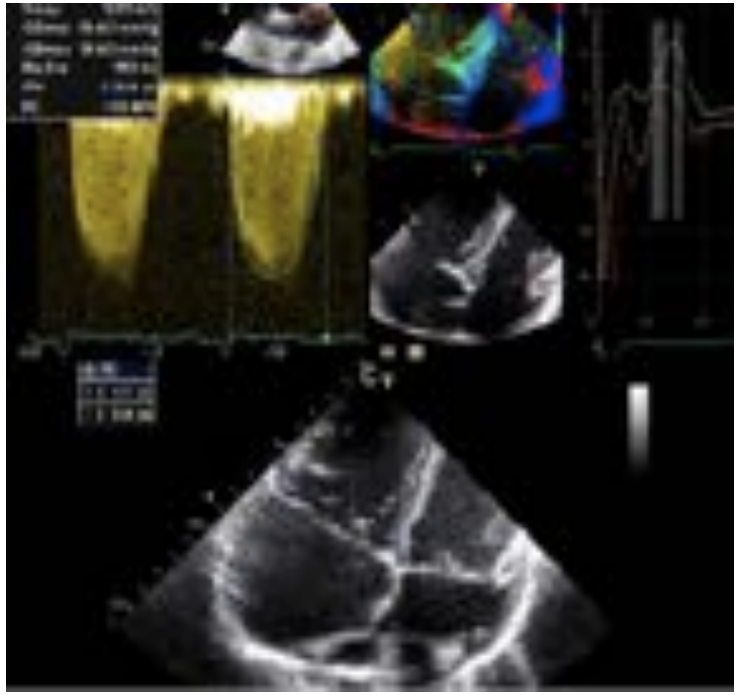
Wadia et al. Heart 2019

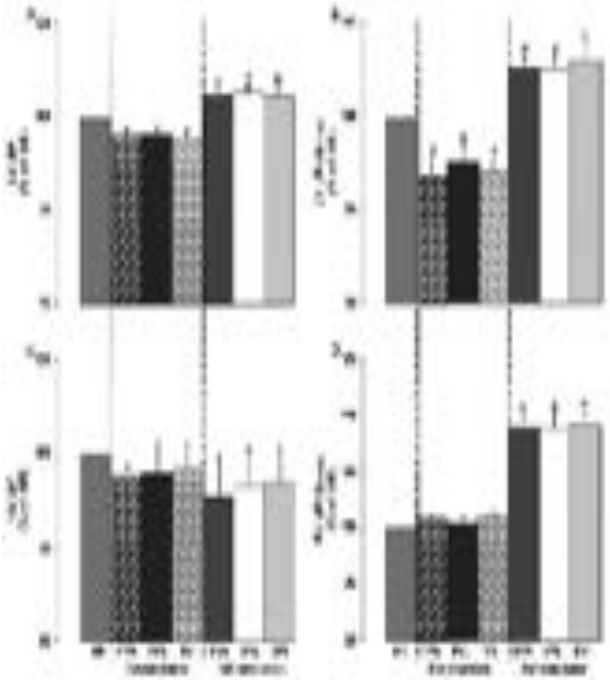
ARYTHMIES – MORT SUBITE

Validation of an animal model of right ventricular dysfunction and right bundle branch block to create close physiology to postoperative tetralogy of Fallot

Jean-Benoit Thambo [b,c,*], Francois Roubertie [b], Maxime De Guillebon [a,b], Louis Labrousse [b,c], Xavier Iriart [b], Hopewell Ntsinjana [d], Stephane Lafitte [b,c], Sylvain Ploux [a,b], Michel Haissaguerre [b], Xavier Roques [b], Pierre Dos Santos [a,b,c], Pierre Bordachar [b,c]



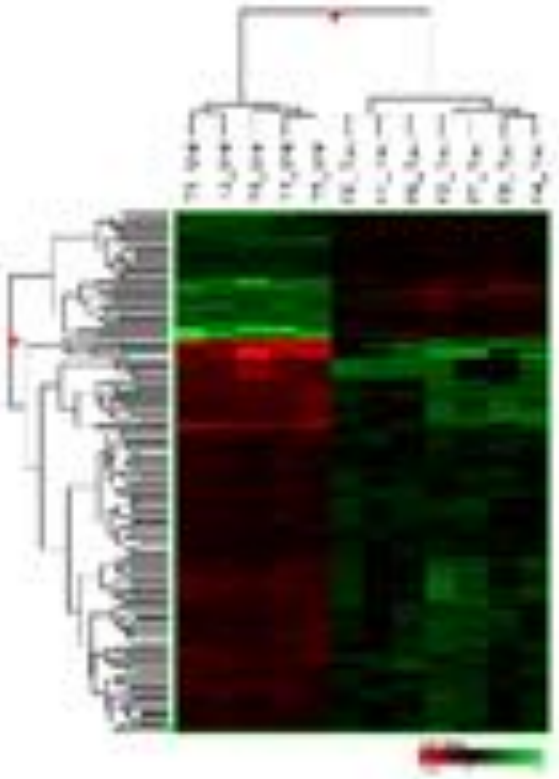




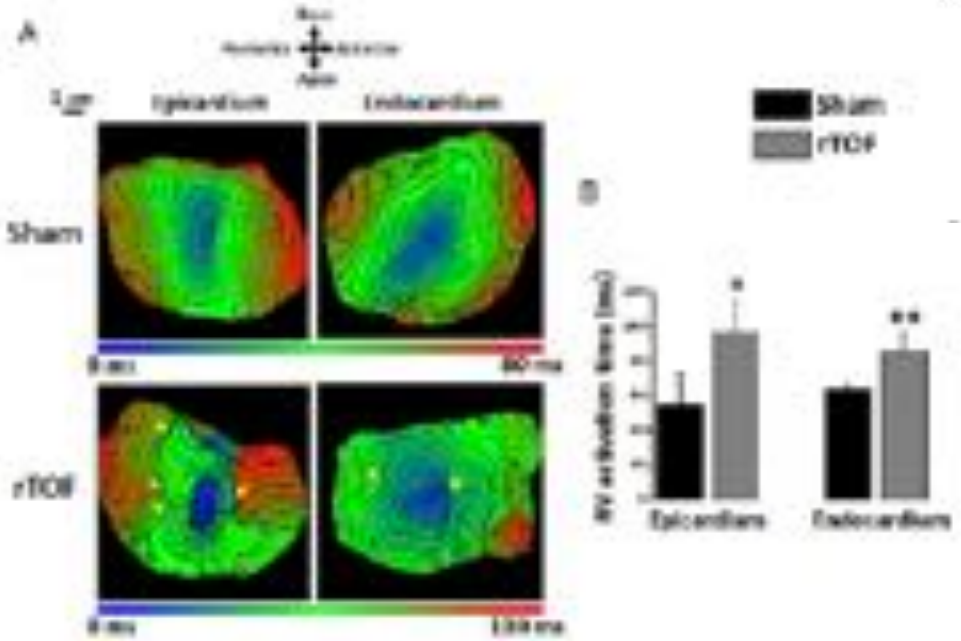
Thambo et al. Heart Rhythm 2011
Thambo et al. Int Jour Cardiol 2012

ARYTHMIES – MORT SUBITE

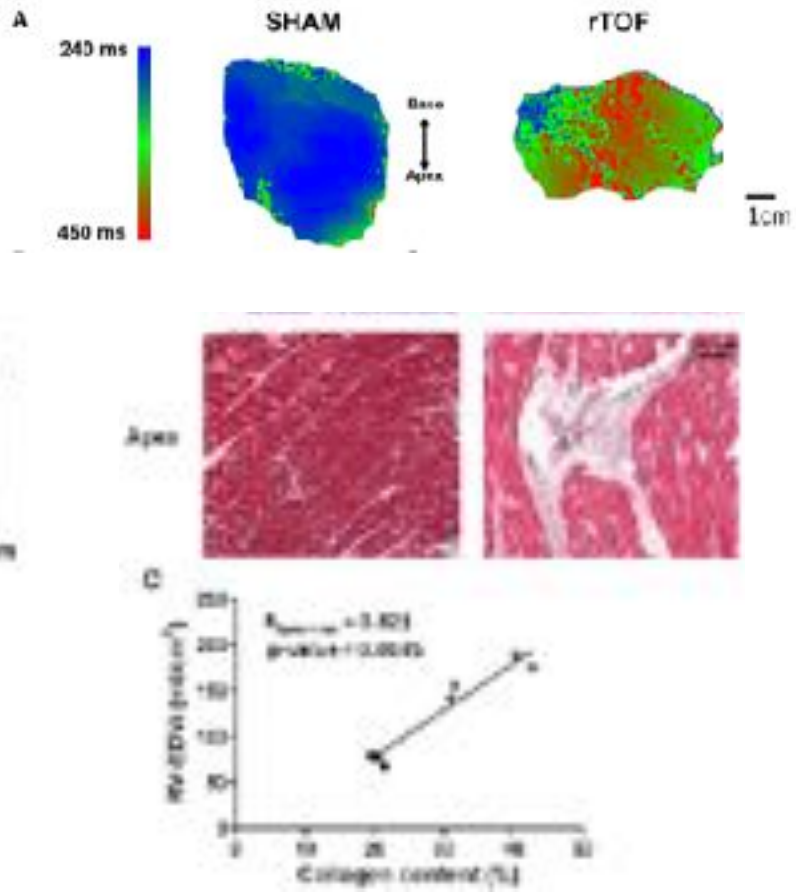
Dérégulation génétique



Modification électrophysiologique VD



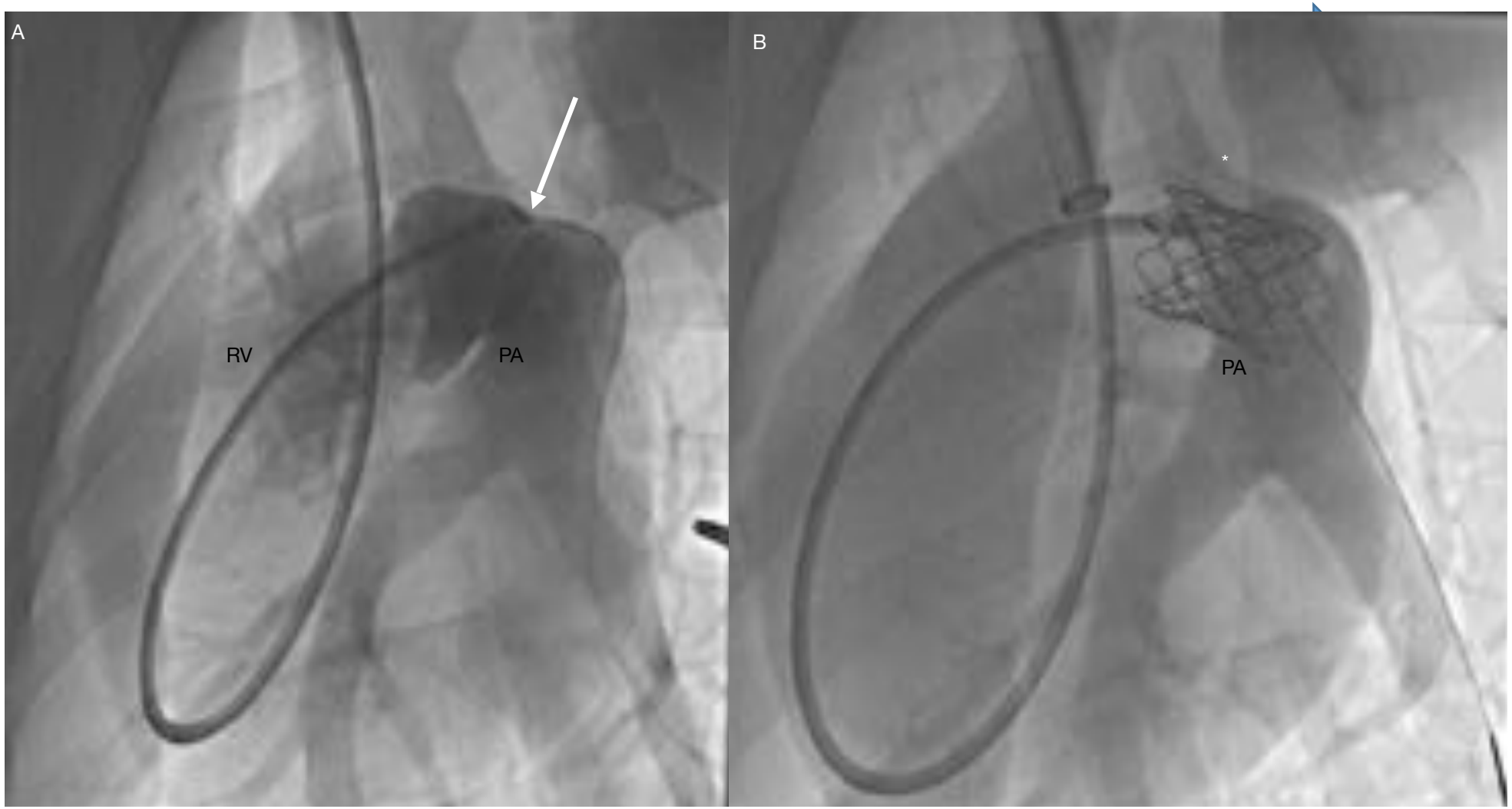
Modification électrophysiologique VG



Charron et al. PLOS One 2016
Benoist et al. Heart 2017
Dubes et al. Circ Arrhythm Electrophysiol. 2018

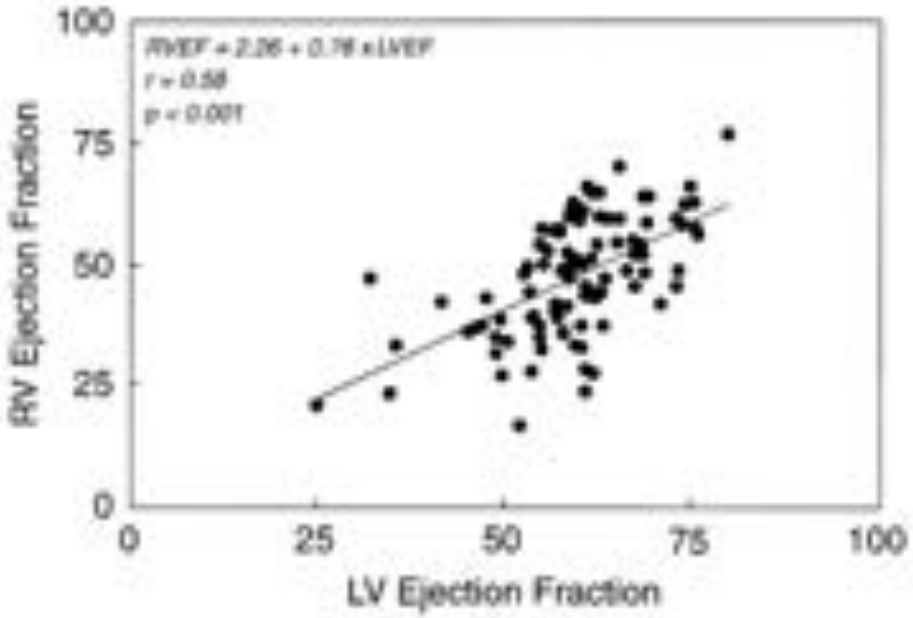
ARYTHMIES – MORT SUBITE

CE REMODELAGE TISSULAIRE ET ELECTROPHYSIOLOGIQUE EST-IL RÉVERSIBLE?



DYSFONCTION VENTRICULAIRE

INTERACTION VD/VG

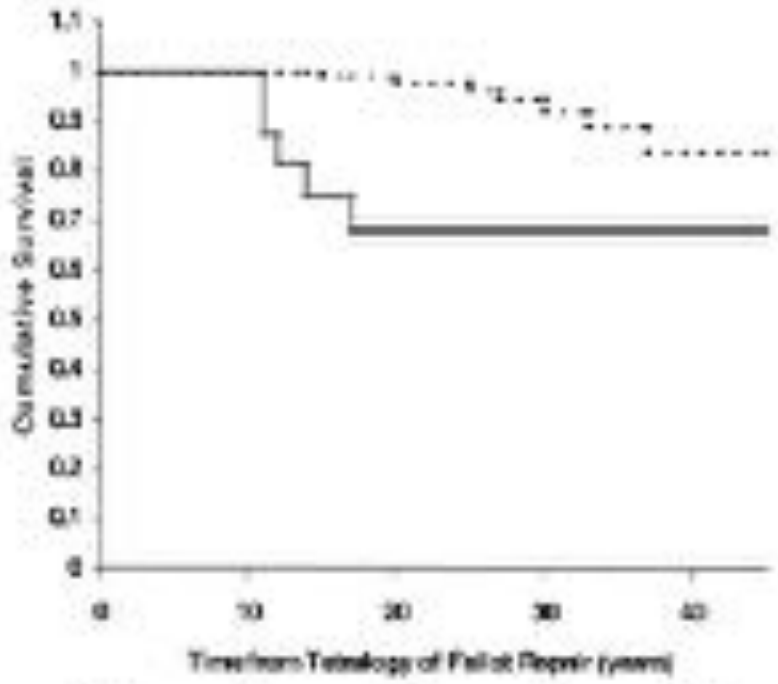


- Anatomique
  - Fibres superficielles communes (fct systolique)
  - SIV (modification de la courbe P/V VG) (fct systolodiastolique)
  - Péricarde (fct diastolique)
- Electrique
  - Asynchronisme interV, intraVD, intraVG
- Hémodynamique
  - Surcharge volumique
  - Surcharge Barométrique

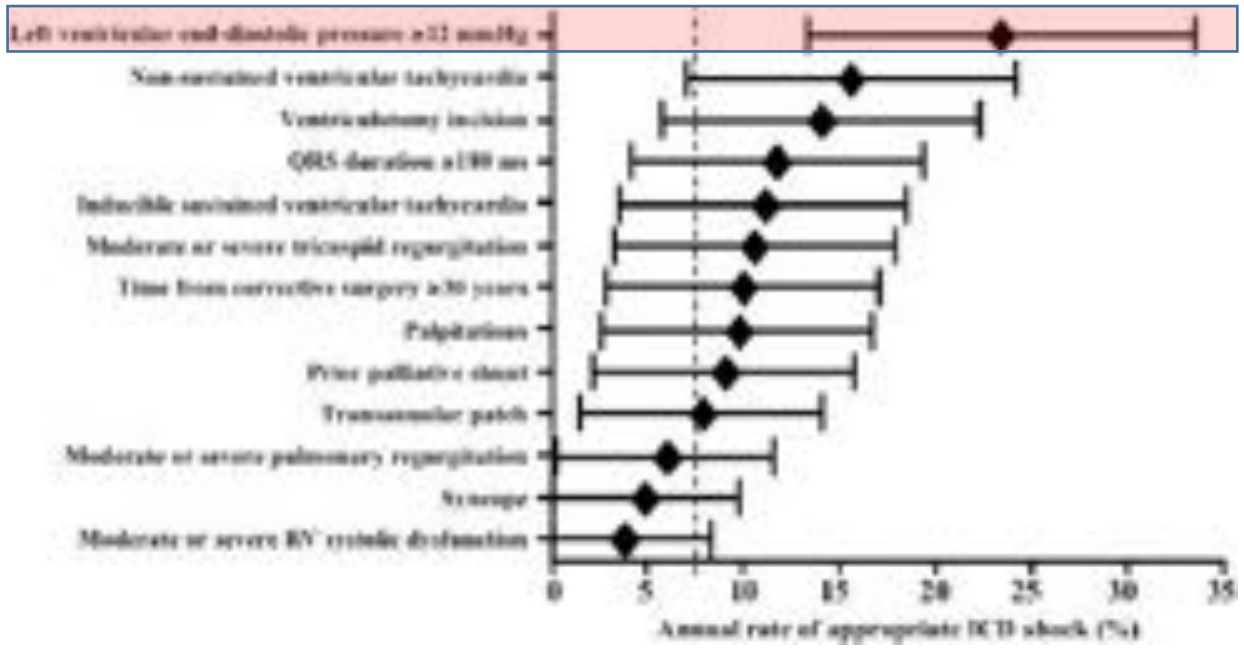
Geva T. JACC 2004

DYSFONCTION VENTRICULAIRE

IMPORTANCE DE LA FONCTION VENTRICULAIRE GAUCHE

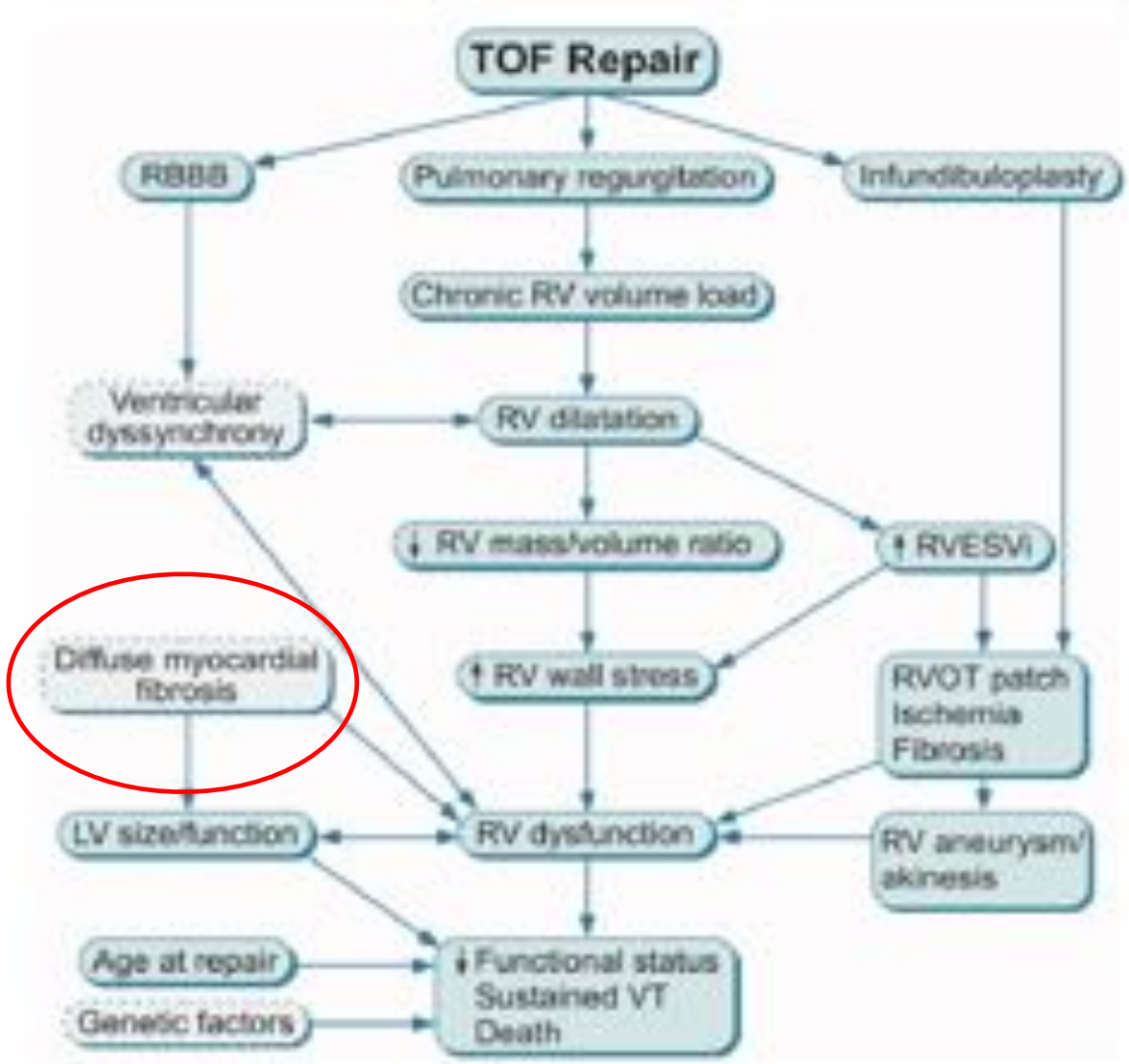


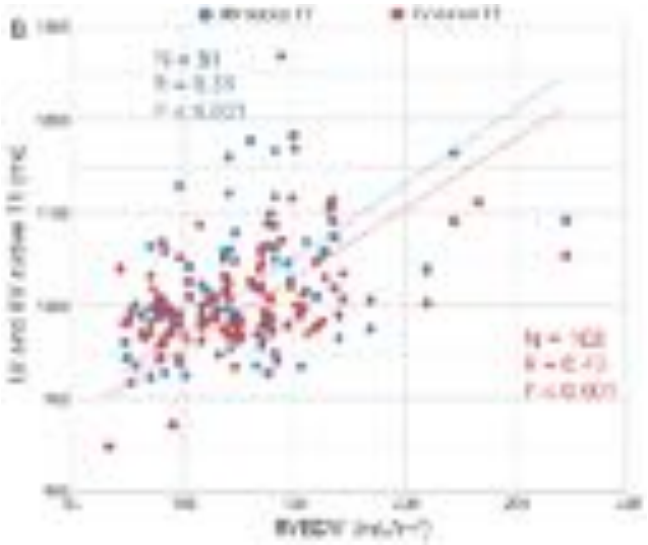
Ghai, A. et al. J Am Coll Cardiol 2002



Khairy P. Circulation 2008

DYSFONCTION VENTRICULAIRE



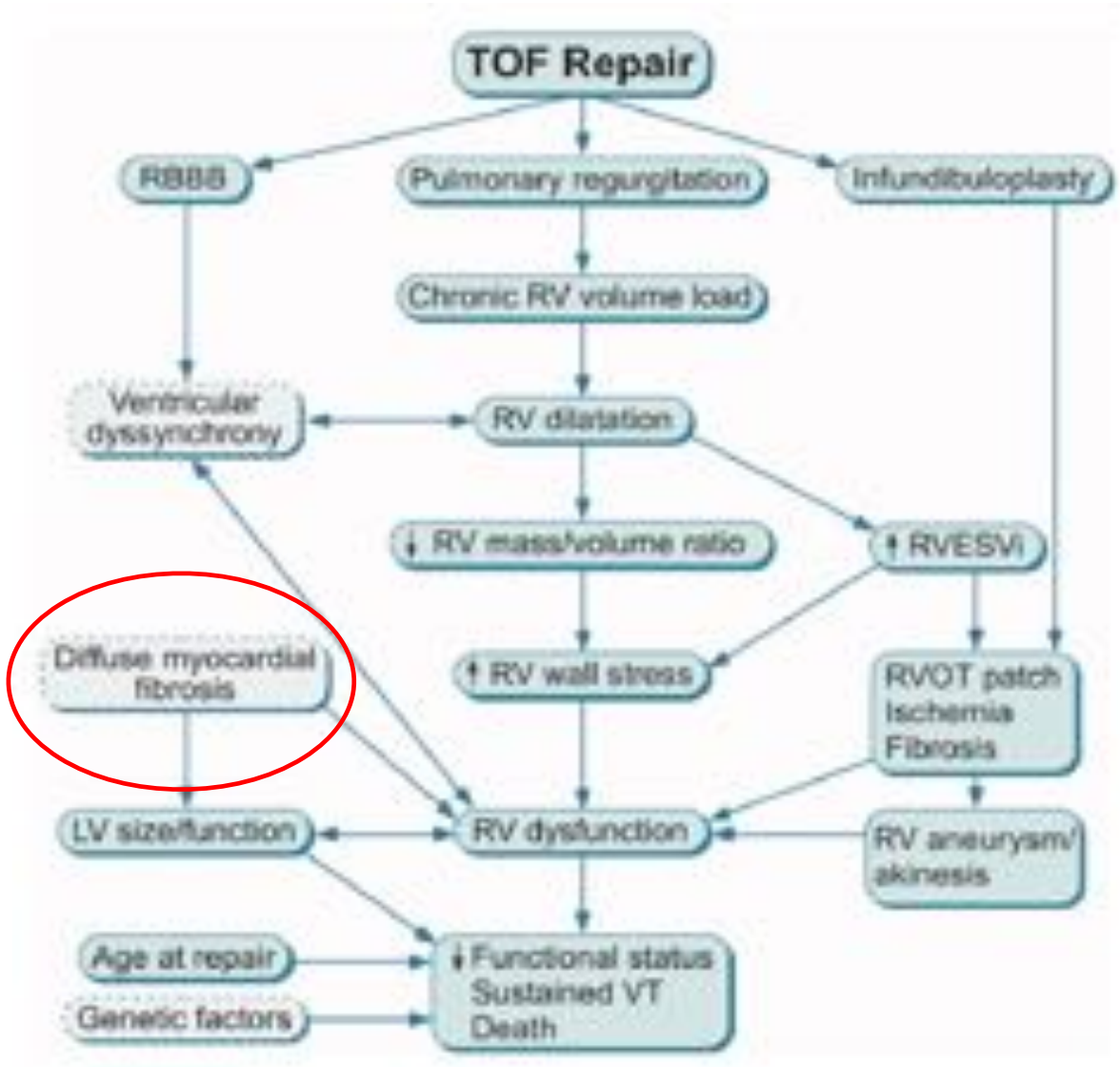


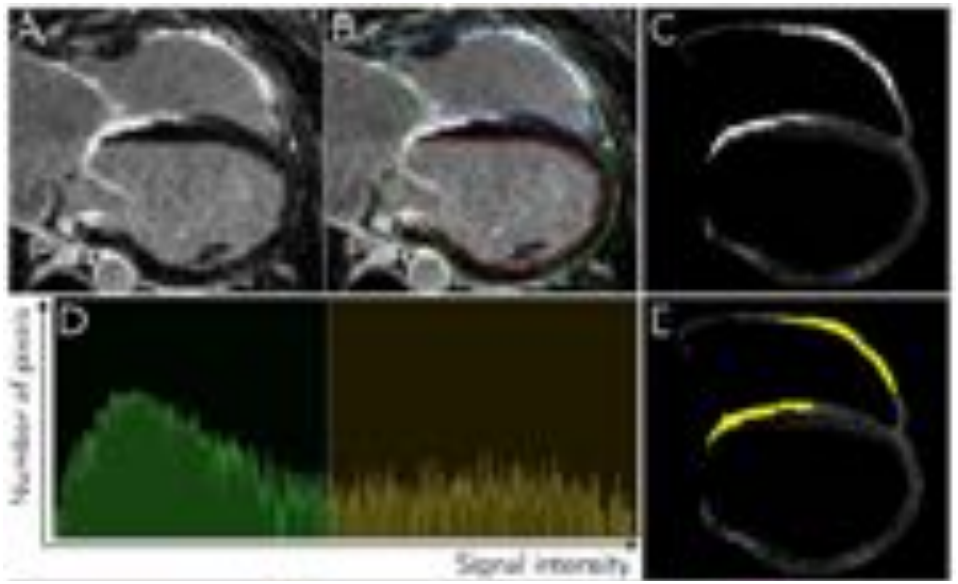
Diffuse Fibrosis associated with **RVEDV**

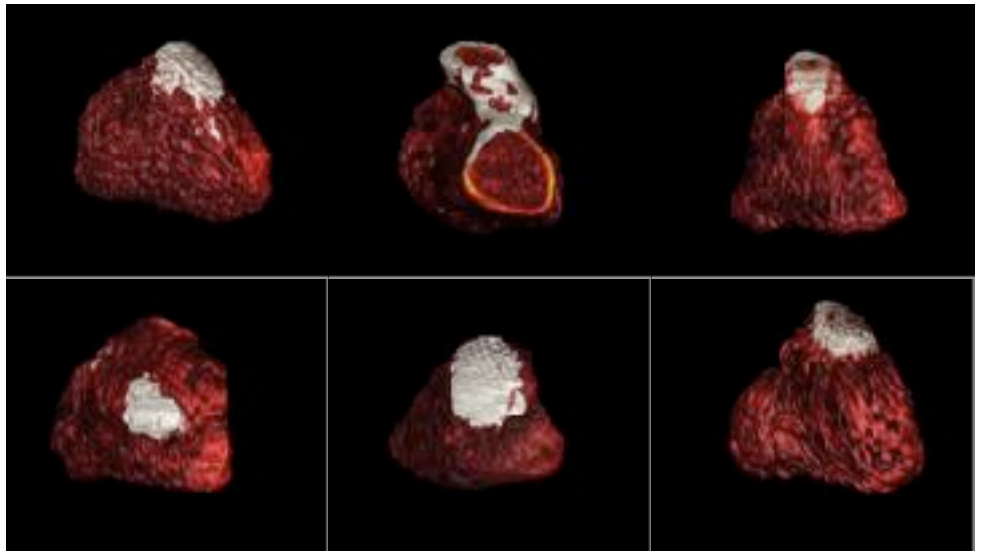
Table 6: Multivariable correlates of ventricular arrhythmia	R^2 value	P value
3-variable model:		
Total scar size, LV native T1, RVEF		
Total scar size	0.376	0.003
LV native T1	0.150	0.003
RVEF	0.021	0.23
Model's R^2	**0.547**	

Geva T et al. JCMR 2011
Cochet et al. EHJCVI 2019

DYSFONCTION VENTRICULAIRE

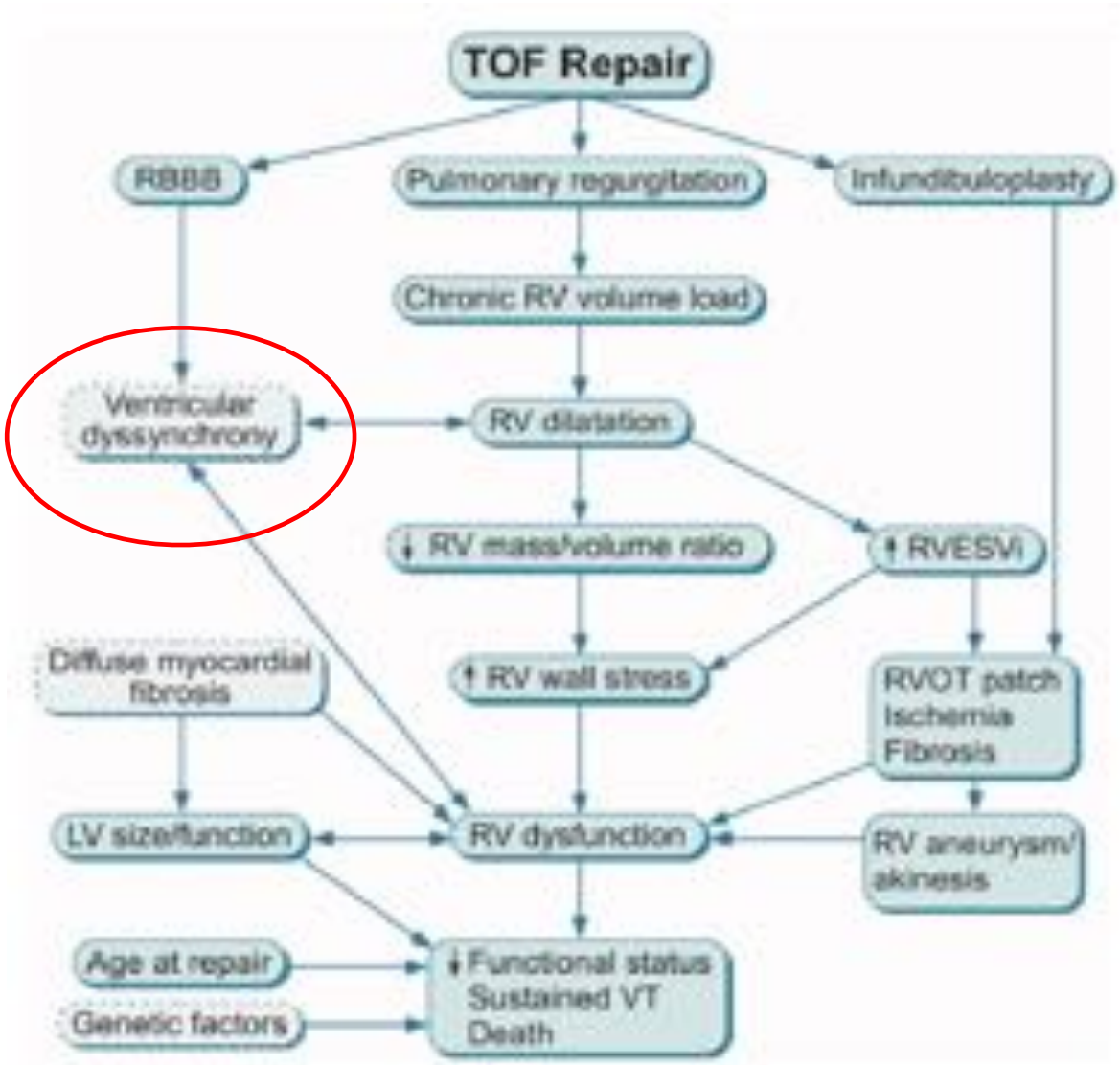






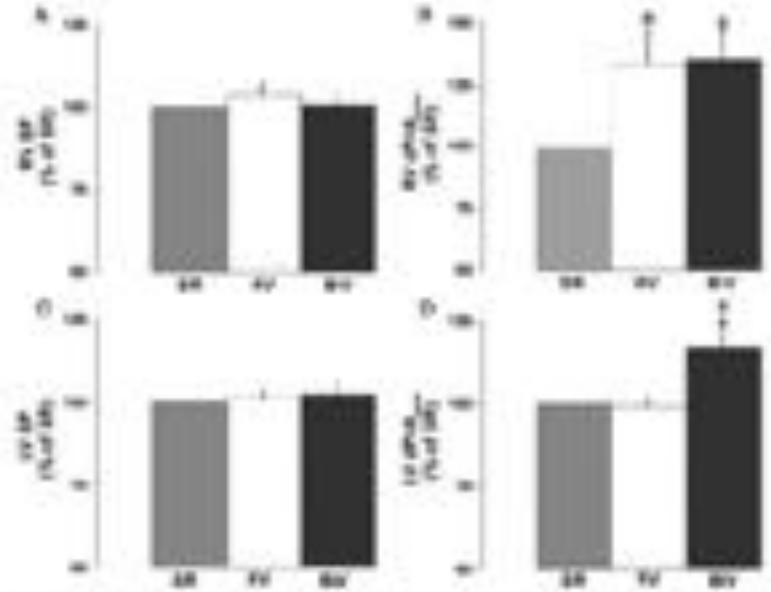
Geva T et al. JCMR 2011
Cochet et al. EHJCVI 2019

DYSFONCTION VENTRICULAIRE



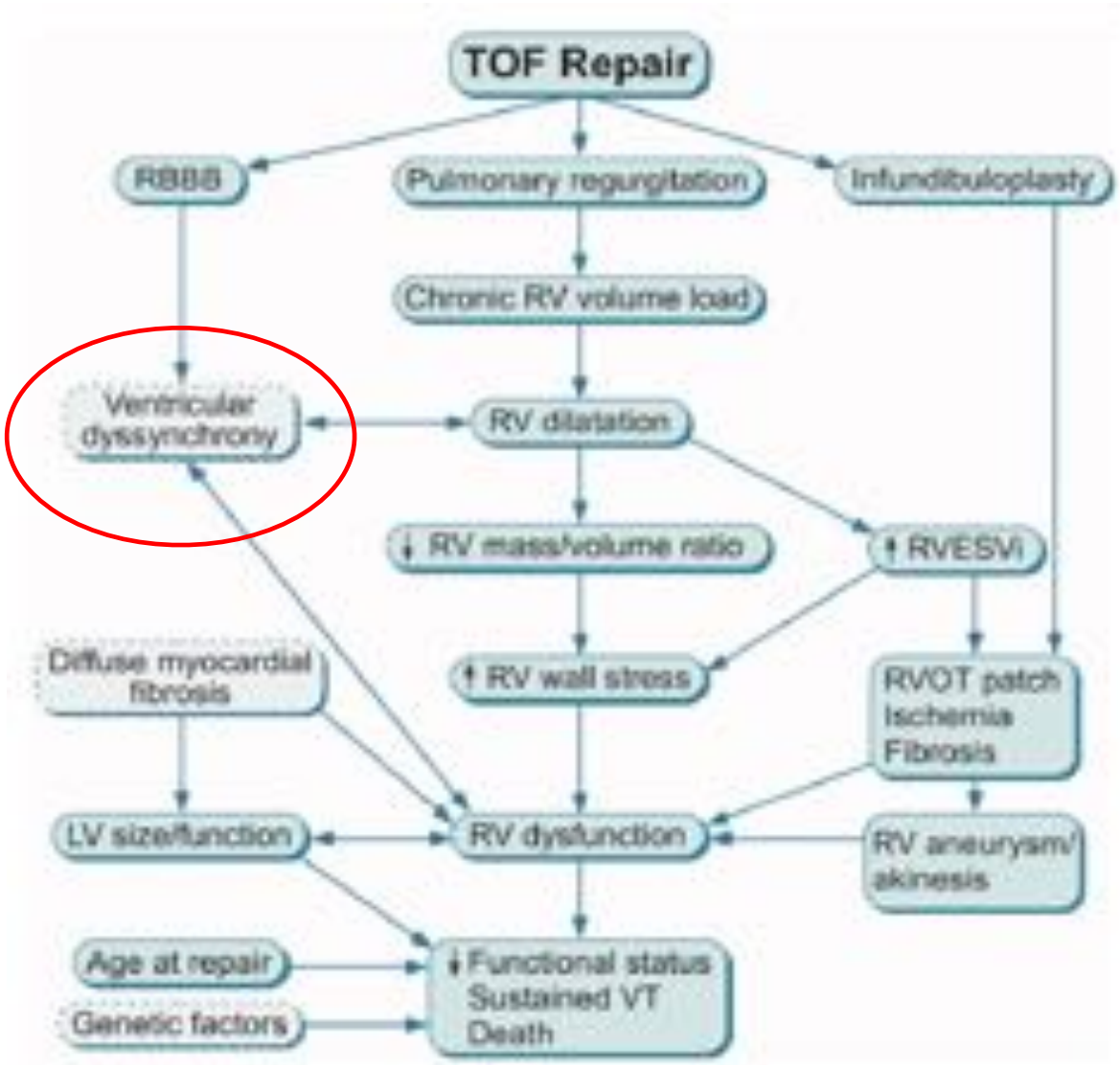
Biventricular stimulation improves right and left ventricular function after tetralogy of Fallot repair: Acute animal and clinical studies

Jean-Benoit Thambo, MD[†,‡], Pierre Dos Santos, MD, PhD[*,†,‡], Maxime De Guillebon, MD[*,†], Francois Roubertie, MD[†], Louis Labrousse, MD[†,‡], Frederic Sacher, MD[†], Xavier Iriart, MD[†], Stephane Lefitte, MD, PhD[†,‡], Sylvain Ploux, MD[*,†], Pierre Jais, MD[*,†], Xavier Roques, MD[†], Michel Haissaguerre, MD[*,†], Philippe Ritter, MD[†], Jacques Clementy, MD[*,†], Sanjiv M. Narayan, MD, FACC[¶], and Pierre Bordachar, MD[†,‡]



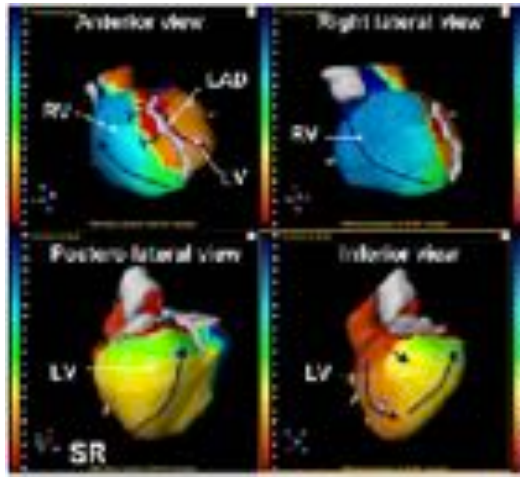
Geva T et al. JCMR 2011
Thambo et al. Heart Rhythm 2012

DYSFONCTION VENTRICULAIRE



Geva T et al. JCMR 2011
Thambo et al. Heart Rhythm 2012

RESYNCHRONISATION BI-VENTRICULAIRE



RÉSULTATS PROMETTEURS

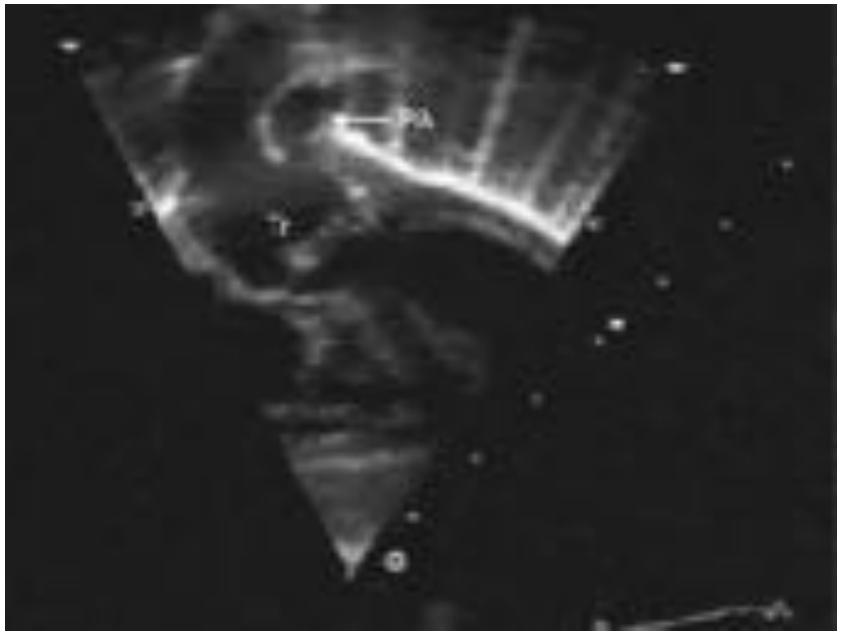
DONNÉES À LONG TERME RARES

SÉLECTION DES CAS

COMPLICATIONS LIÉES AUX DISPOSITIFS

PATHOLOGIE DE L'AORTE

VALVULOATHIE AORTIQUE (TAC+++)



DILATATION DE L'AORTE ASCENDANTE

15% des Fallots ont une dilatation de l'aorte ascendante (>40mm)

Phénomène progressif

Etiologies: hémodynamique, anomalies histologiques

Dilatation de l'anneau = > Fuite valvulaire

Indications chirurgicales rares (sauf TAC avec valve troncale anormale+++)

PATHOLOGIE DE L'AORTE

ASCENDING AORTA

Does the dilated ascending aorta in an adult with congenital heart disease require intervention?

- 81 patients
- Cardiopathie initiale:
  - APSO 41 patients (51%),
  - TF 19 (23%),
  - TAC 12 (15%),
  - VDDI 6 (7%),
- Diamètre médian aortique: 45mm (range23-80)
- Indications:
  - 74 valvulopathies,
  - 16 anévrysmes,
  - 1 dissection
- RVAo seul => 0 reprise pour l'aorte ascendante

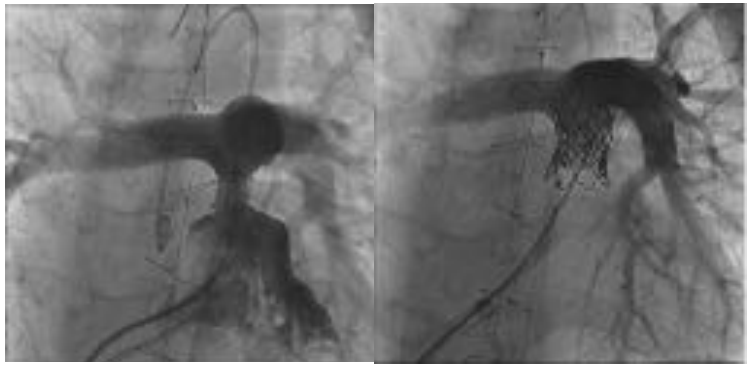
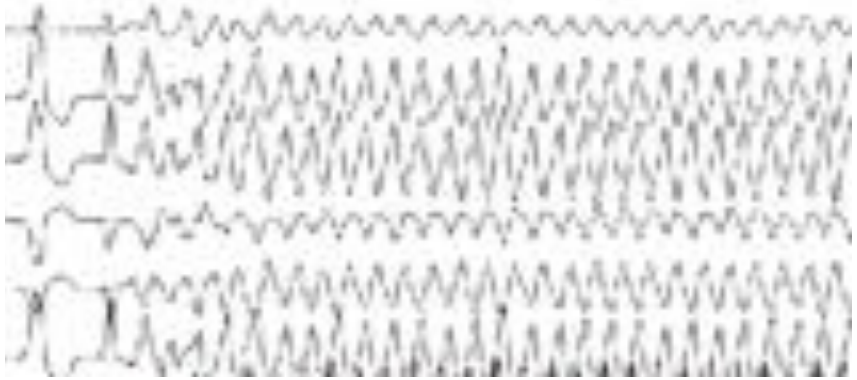
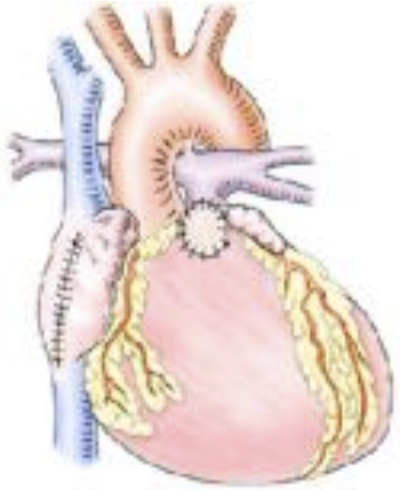
PATHOLOGIE DE L'AORTE

Table 15 Indications for intervention after repair of tetralogy of Fallot

Indications	Class[a]	Level[b]
Aortic valve replacement should be performed in patients with severe AR with symptoms or signs of LV dysfunction	I	C

Although patients with conotruncal abnormalities (TOF, dextrotransposition of the great arteries [d-TGA] after arterial switch,[512426-512428] pulmonary atresia with VSD, truncus arteriosus) commonly have aortic diameters of 40 mm to 50 mm, aortic complications are extremely rare (only 6 published case reports).[512429-512414] Therefore, there is no strong justification for empiric prophylactic surgery in such patients based solely on aortic diameter. Watchful

Prévention FDRCV, surtout HTA+++

CONCLUSION

- Cardiopathies réparées mais non guéries
- Bon pronostic au long cours incidence d’événements indésirable significative
- Suivi au long cours en cardiologue spécialisé en cardiopathies congénitales
- Complications au long cours multiples mais intriquées
- Evaluation multimodale
- Approche multidisciplinaire et personnalisée