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M3C Academy

Arrhythmias after Fallot Repair

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Pôle de cardiologie pédiatrique
et congénitale

Marie Lannelongue

Arrhythmias factors and Fallot patients

1) Factor associated with heamodynamic status before correction
- hypoxemia
- high output
- pressure overload

2) Factor associated with surgery :
- suture lines
- patches
- direct trauma of conducting system
- residual lesions

Genetic factors ?
. TBX5
. NKX 2.5
. Long QT genes

Goldmuntz E et al. Circulation 2001
Baban A et al.Am J Med Genet 2014
Chiu SN et al. Int J Cardiol 2017;249:156

3) Cardiovascular risk factors during life :
HBP, diabete melitus, tobacco use, dyslipidemia, SAS, aging

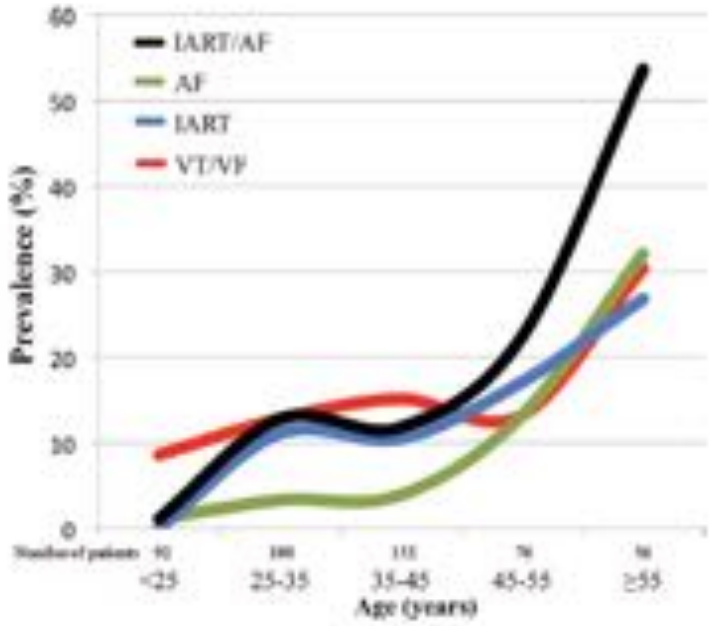
Hypertophy
Dilatation
Ischemia
Electrical dyssynchrony

Fibrosis

Substrate for arrhythmias :
Reentry +++

Arrhythmias epidemiology in Fallot patients

Arrhythmias up to 30% of Fallot patients with long follow up
Sustained ventricular arrythmias up to 10%
Sudden cardiac death incidence 0.2%/year



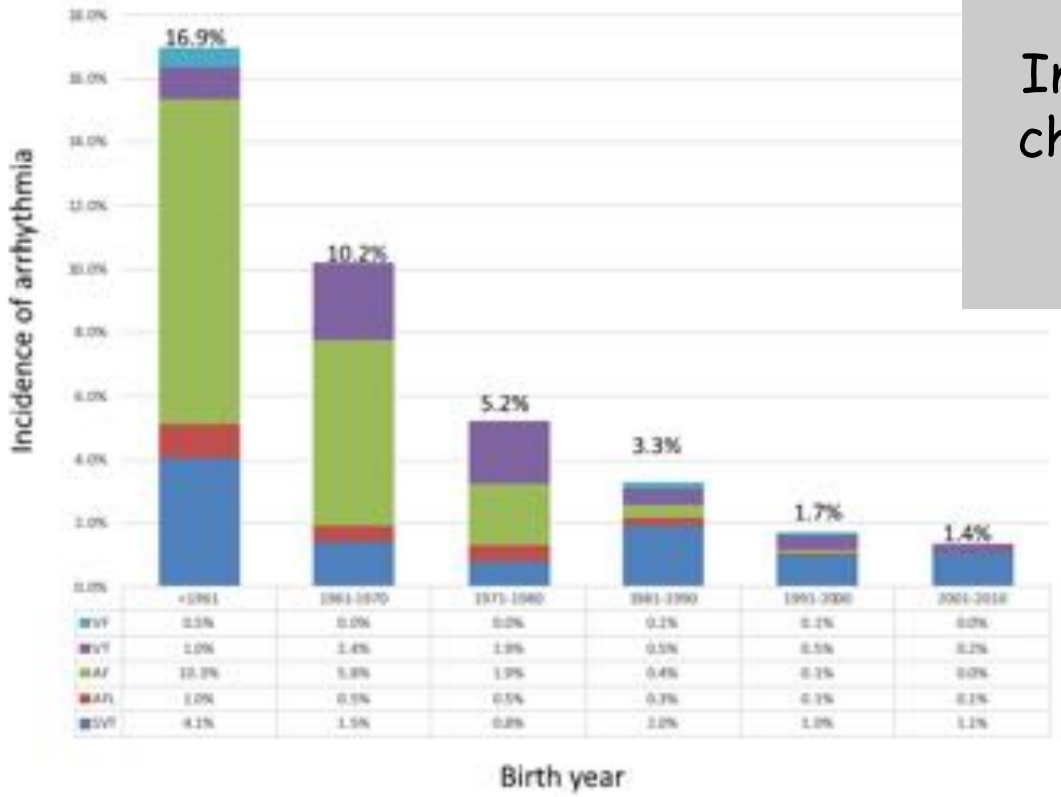
Khairy P et al. *Circulation* 2010;122:868

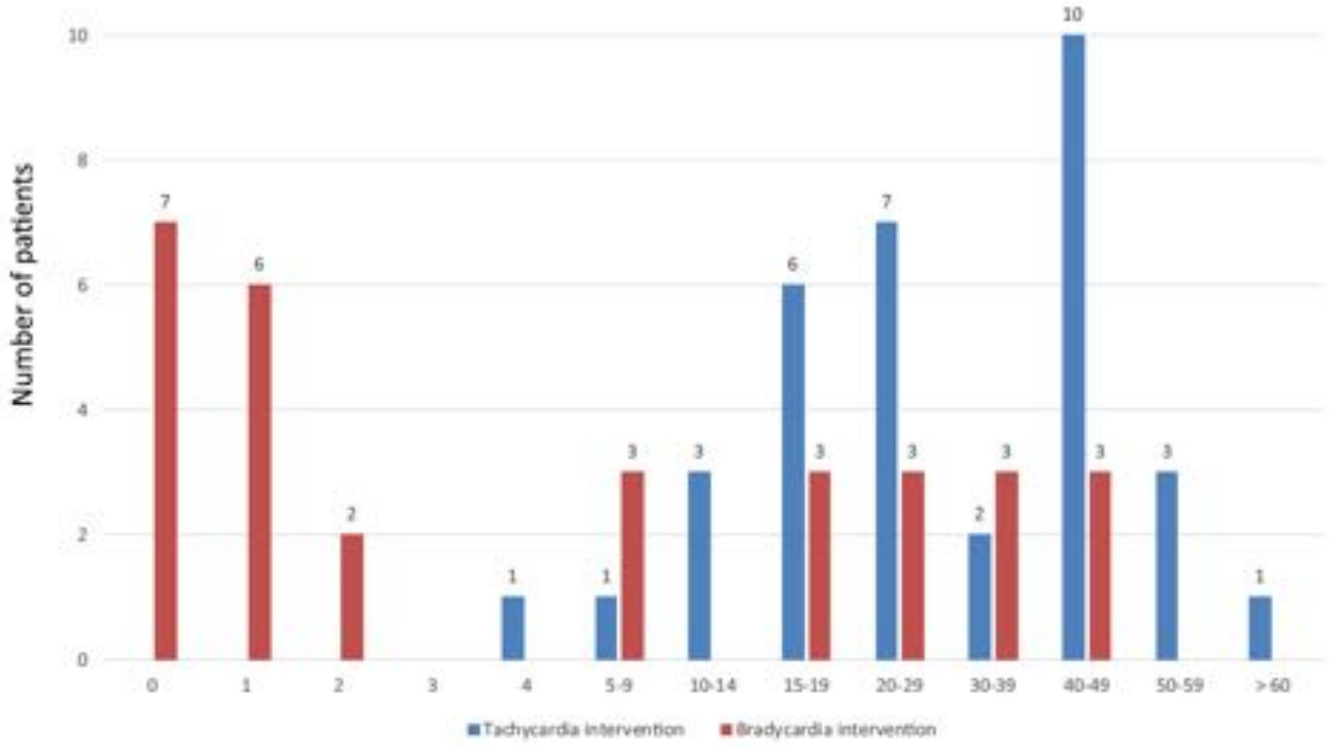
Sudden cardiac death related to:
1) Ventricular tachycardias
2) Rapid conducted supraventricular tachycardias
3) (acute AV block)

AV block essentially in the early post operative period (0.6% in the modern area)

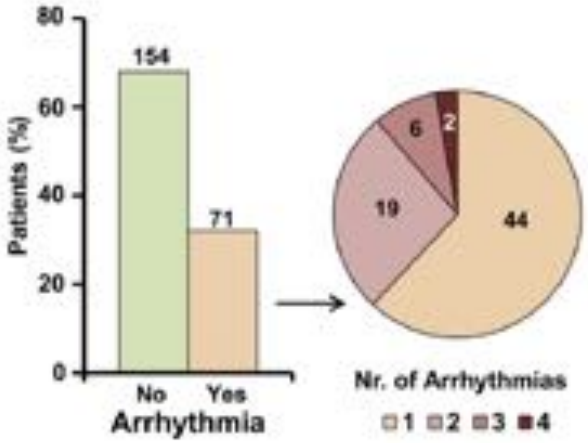
Increasing rate of supraventricular tachycardia with age changing from organized tachycardia to atrial fibrillation

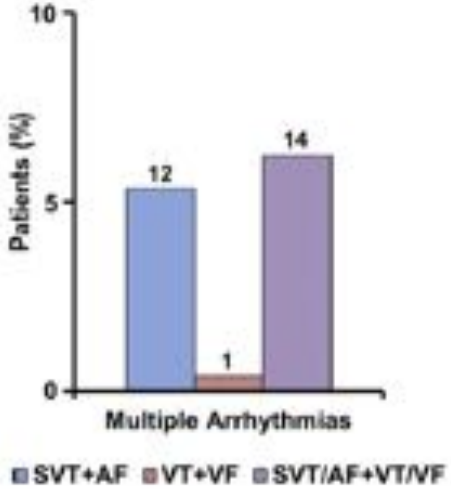
Interventionnal arrhythmia therapy in 20,4%

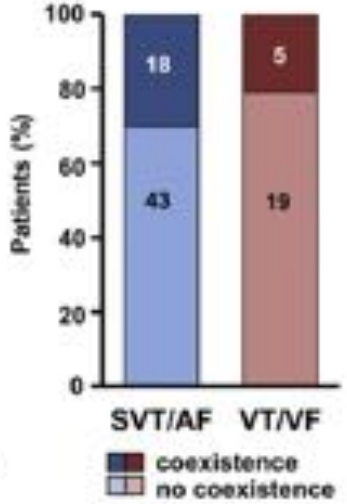




Wu MH et al. *Heart Rhythm* 2015;12:604

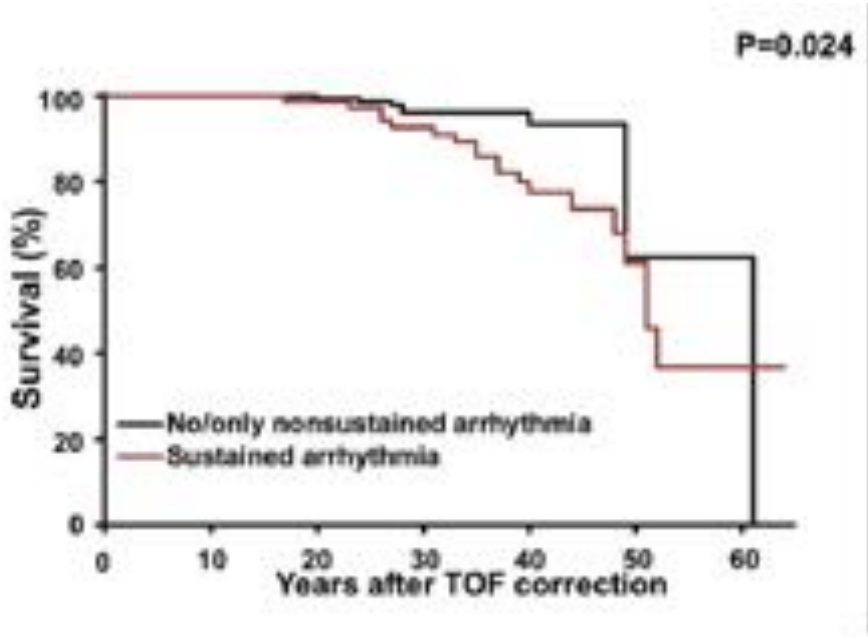


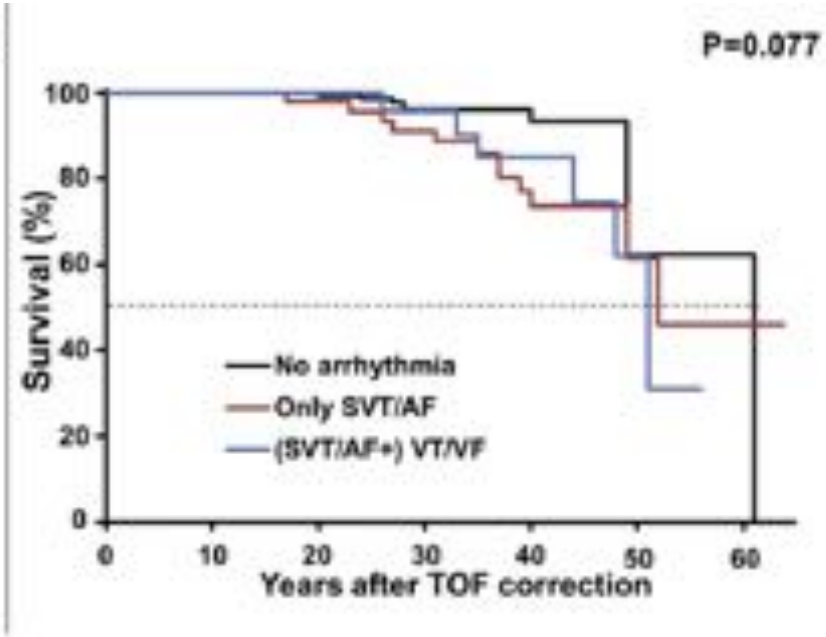




In adults, frequent association of multiple arrhythmias

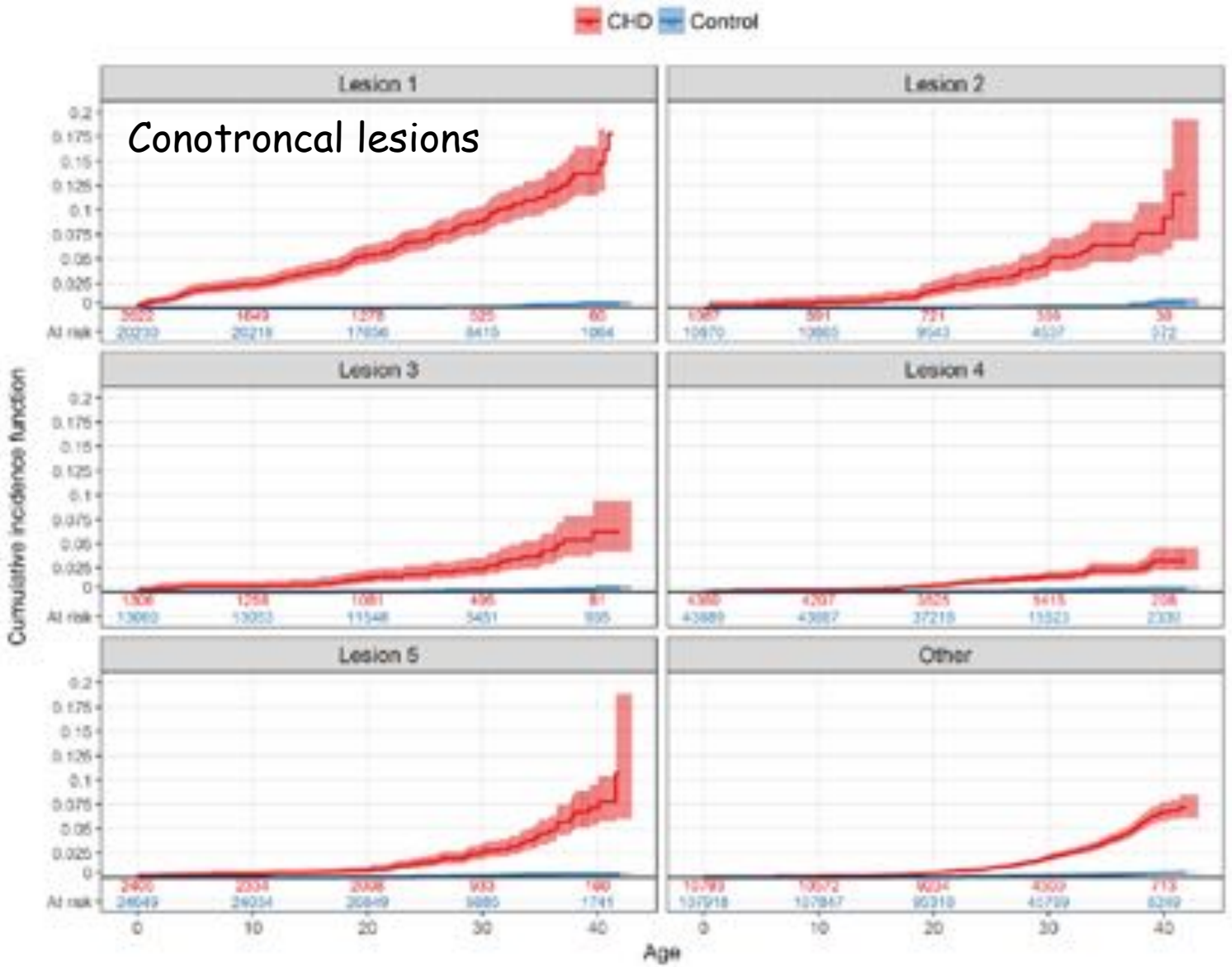
Association with mortality and heart failure





Mouws EM et al. *Heart Rhythm* 2018;15:503

High incidence of atrial fibrillation, X22 compare to control population



Mandalenakis Z et al. *Circulation* 2018;137:928

How to change this arrhythmia profile?

Changing surgical management?

- Early complete correction. Ideal timing?
- Increase cardiac protection, decrease aortic cross-clamp time?
- Limit ventricular incisions?
- Limit patches (indundibular)?
- Pulmonary valve sparing techniques to decrease redo procedures?
- Incisions orientations guided by potentials tachycardia isthmuses in infants?
- Arrhythmia prophylactic surgery in adults?
- Earlier redo surgery for residual lesions/pulmonary regurgitation?

Aggressive management of lifestyle and cardiovascular risk factors ?

Stimulation in Fallot

In cases of:

- Complete AV block: permanent/paroxysmal

- (Symptomatic sinus node dysfunction)

- Cardiac resynchronisation :
  . NYHA ≥ II, QRS > 150 ms (or > 120 ms with LBBB pattern), LV EF ≤ 35%
  . Indication for permanent ventricular stimulation and LV EF < 40%

How to stimulate?

Khairy P et al. Heart Rhythm 2014;11:e102
Hernandez-Madrid A et al. Europace 2018;20:1719

Stimulation in Fallot

Epicardial stimulation for:

- Children to spare venous access
- Pateints with residual shunts to avoid cardiac emboli

Mac Leod KA. *Heart* 2010;96:1502
Khairy P et al. *Circulation* 2006;113:2391
Gebauer RA et al. *Europace* 2009;11:1654
Kelvin CL et al. *Heart rhythm* 2015;12:566

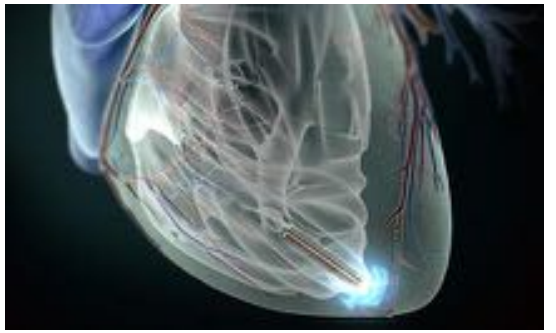
Mode of stimulation:

- Single left ventricular lead in infants with good haemodynamics
- Atrial and ventricular leads in other cases

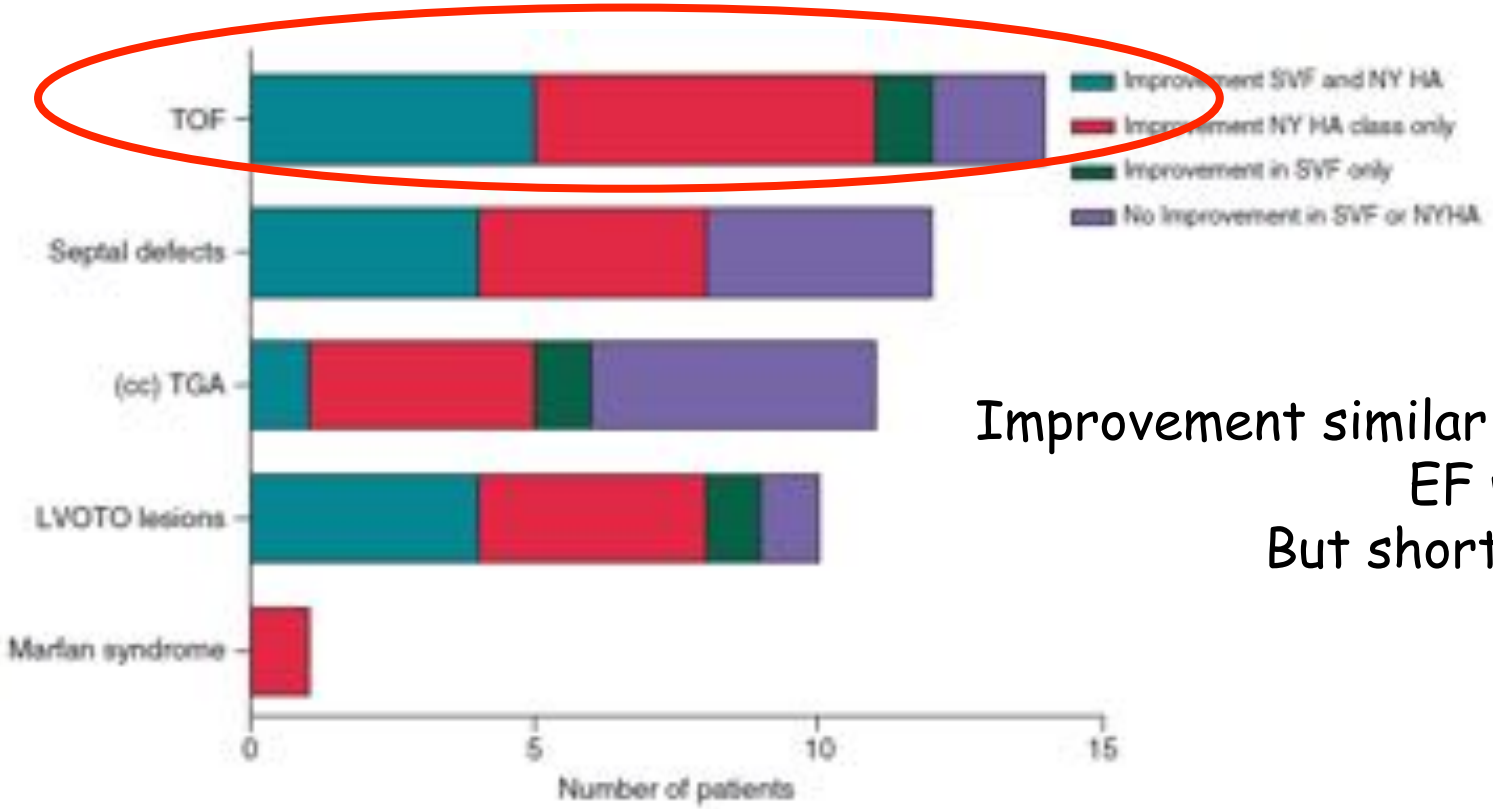
Contemporary problems:

- 30% lead failure at 10 years
- High threshold stimulation
- Difficult epicardial stimulation in patients with redo surgeries
- Devices related infections (endocardial ++)

Leadless pacemakers?
Long life batteries?



Cardiac resynchronisation in Fallot patients ?



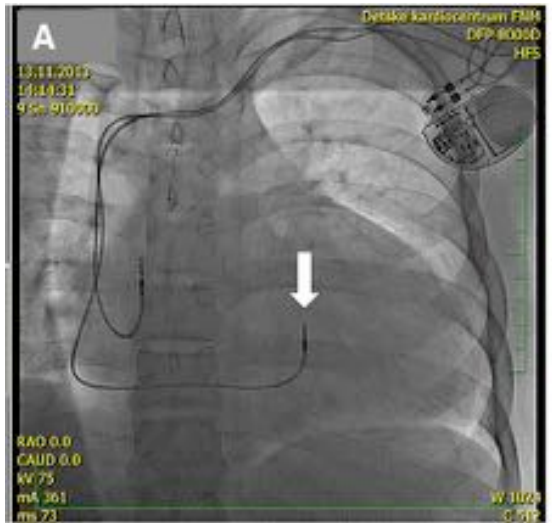
Improvement similar to cardiomyopathies with low LV EF with large QRS
But short series and follow-up

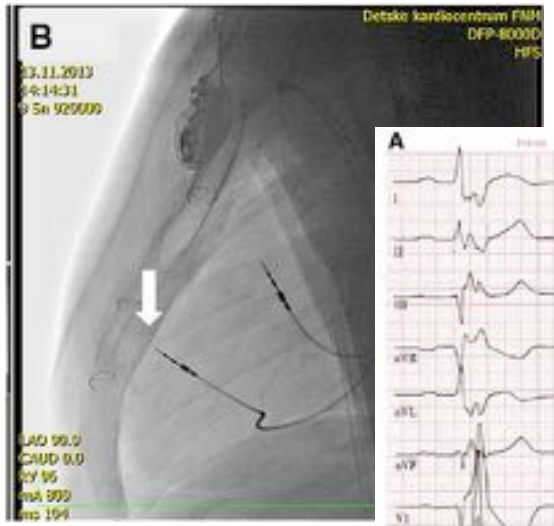
Koyak Z et al. *Europace* 2018

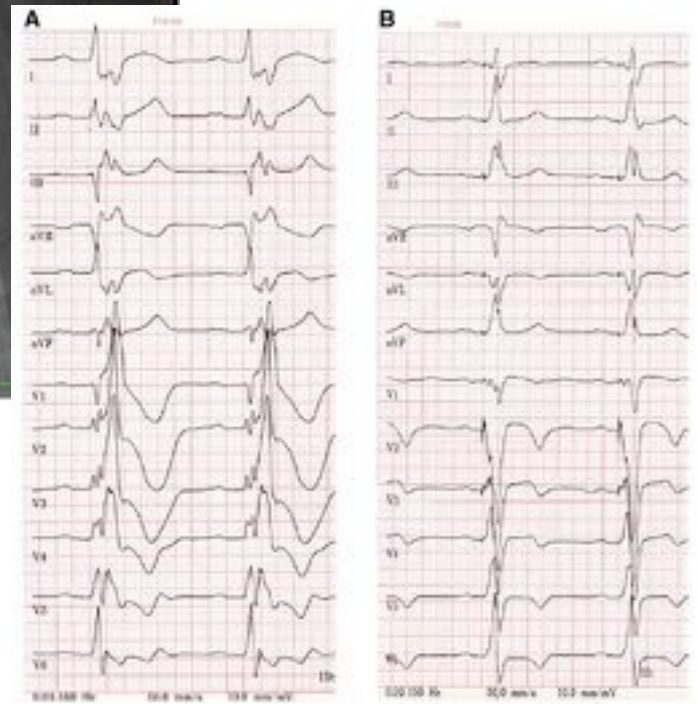
Merchant FM et al. *Pacing Clin Electrophysiol* 2014
Thambo JB et al. *Int J Cardiol* 2013
Thambo JB et al. *Heart Rhythm* 2010

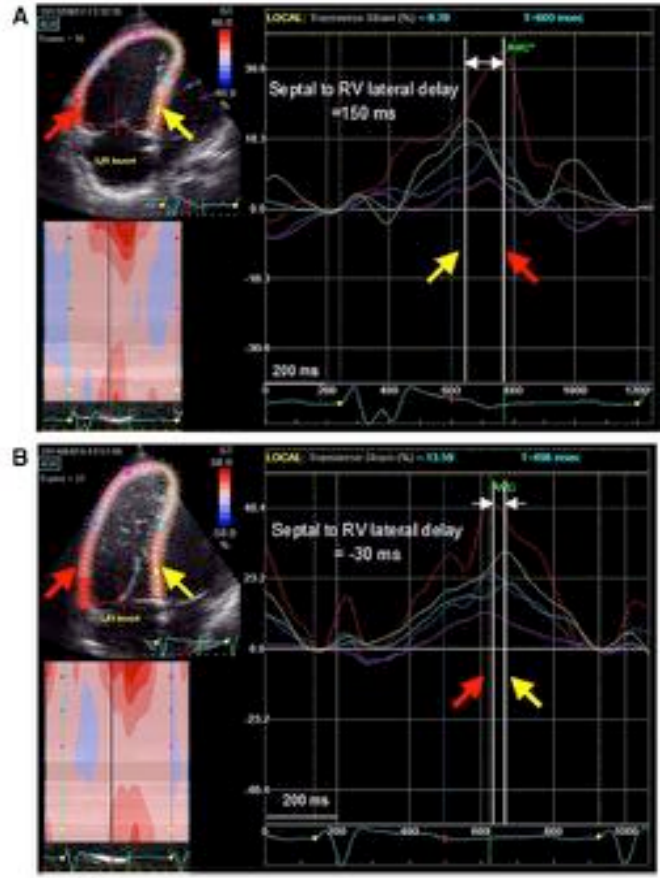
RV resynchronisation?

Possible haemodynamic improvement with stimulation of the most delayed RV wall in fusion with normal ventricular activation









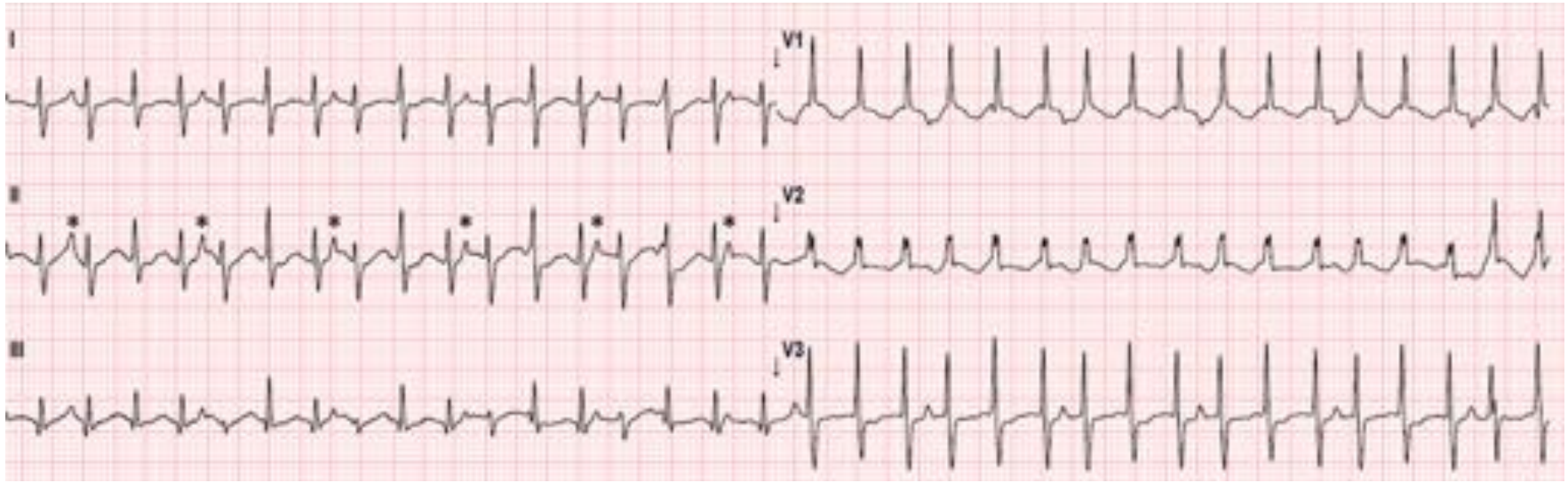
In selected patients
A need for multicentric and long term follow-up

Janousek J et al. *Circ Arrhythm Electrophysiol* 2019
Janousek J et al. *Circ Cardiovasc Imaging* 2017
Janousek J et al. *Circulation* 2014

Supraventricular arrhythmias

Early postoperative period

Junctional ectopic tachycardia (JET) the most frequent arrhythmia but rare
between 5 and 6% of patients



AV conduction tissue
inflammation/trauma

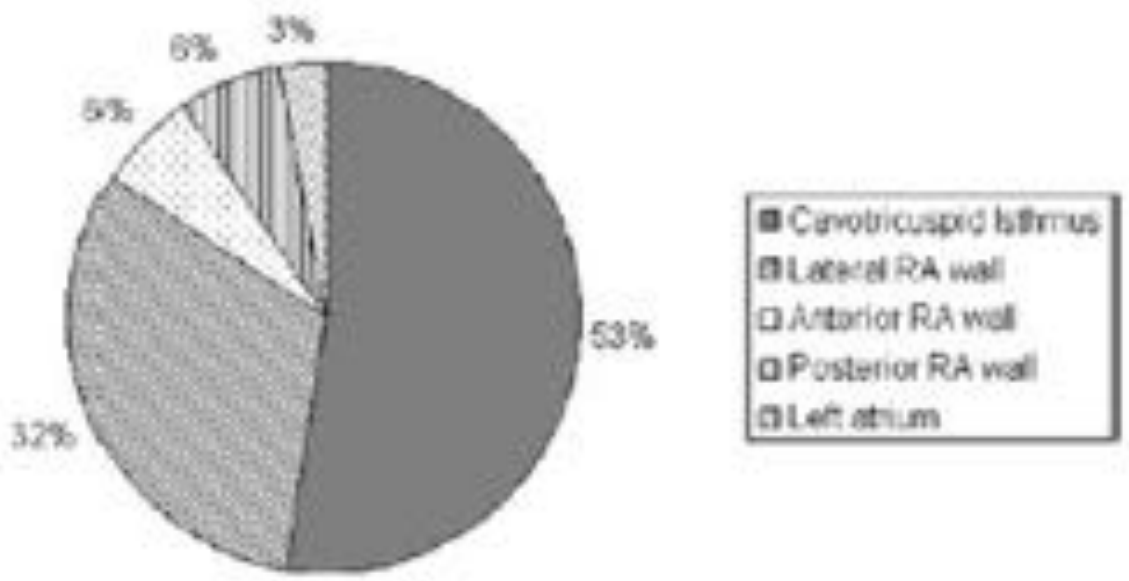
Association with younger age, longer aortic cross-clamping/by-pass time
Tendancy to increase mortality
No evolution to complete AV block

Paluszek C et al. *World J Ped Cong Heart Surg* 2019;10:50
Dodge K et al. *Thorac Cardiovasc Surg* 2002:123:624

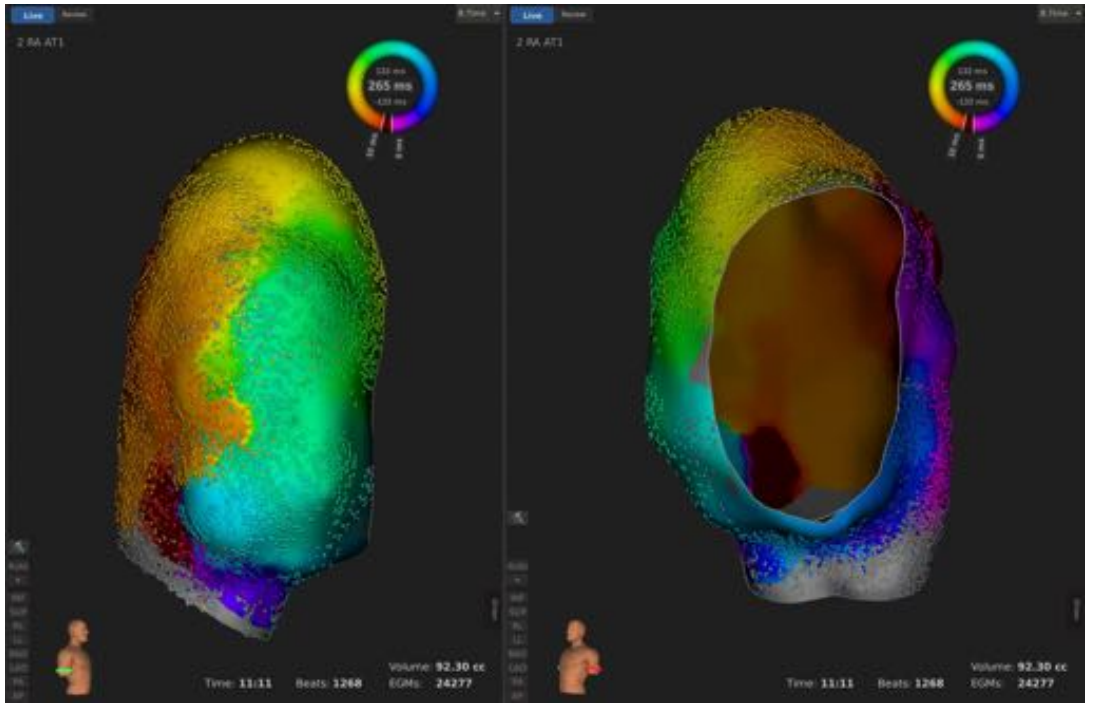
Supraventricular arrhythmias

Late postoperative period

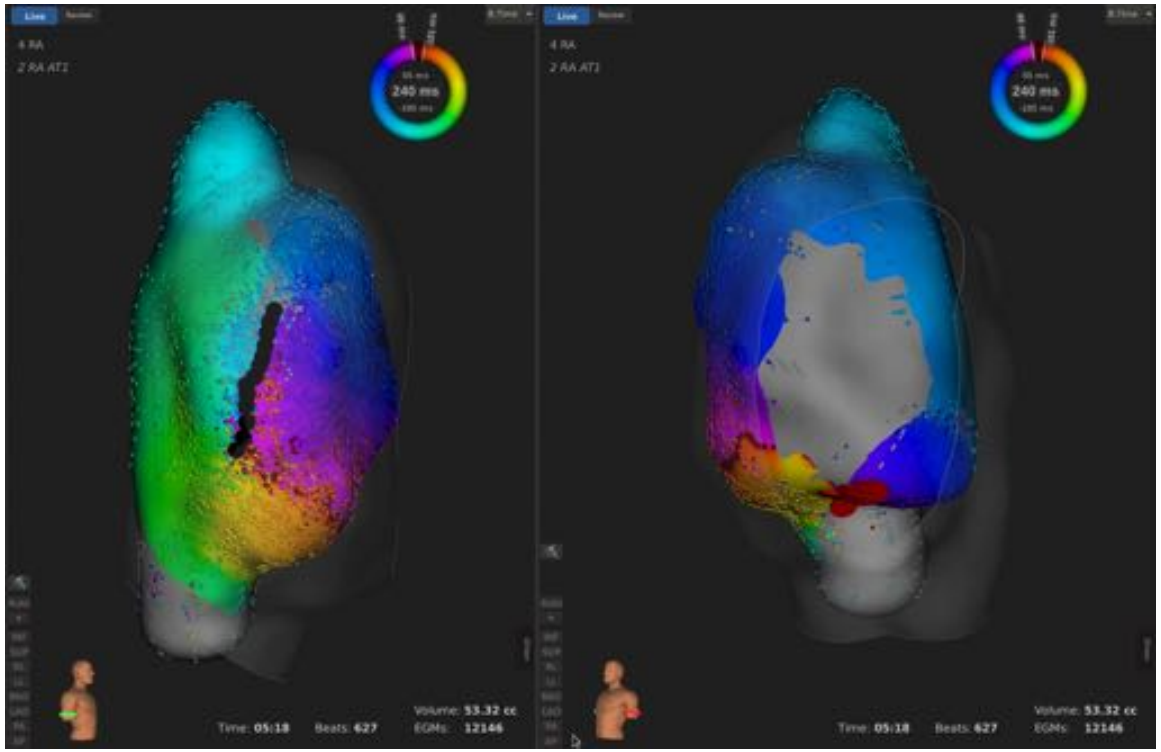
Arrhythmias from right atrium with a majority of macro-reentries
(++ cavo-tricuspid isthmus reentry ie common flutter)
Most of the time first arrhythmia after adolescence



Mah DY et al. *J Cardiovasc Electrophysiol* 2011;22:1013



Ross-Hesselink J et al. *Circulation* 1995;91:2214



Supra ventricular arrhythmias most of the time around scars/patches suture lines

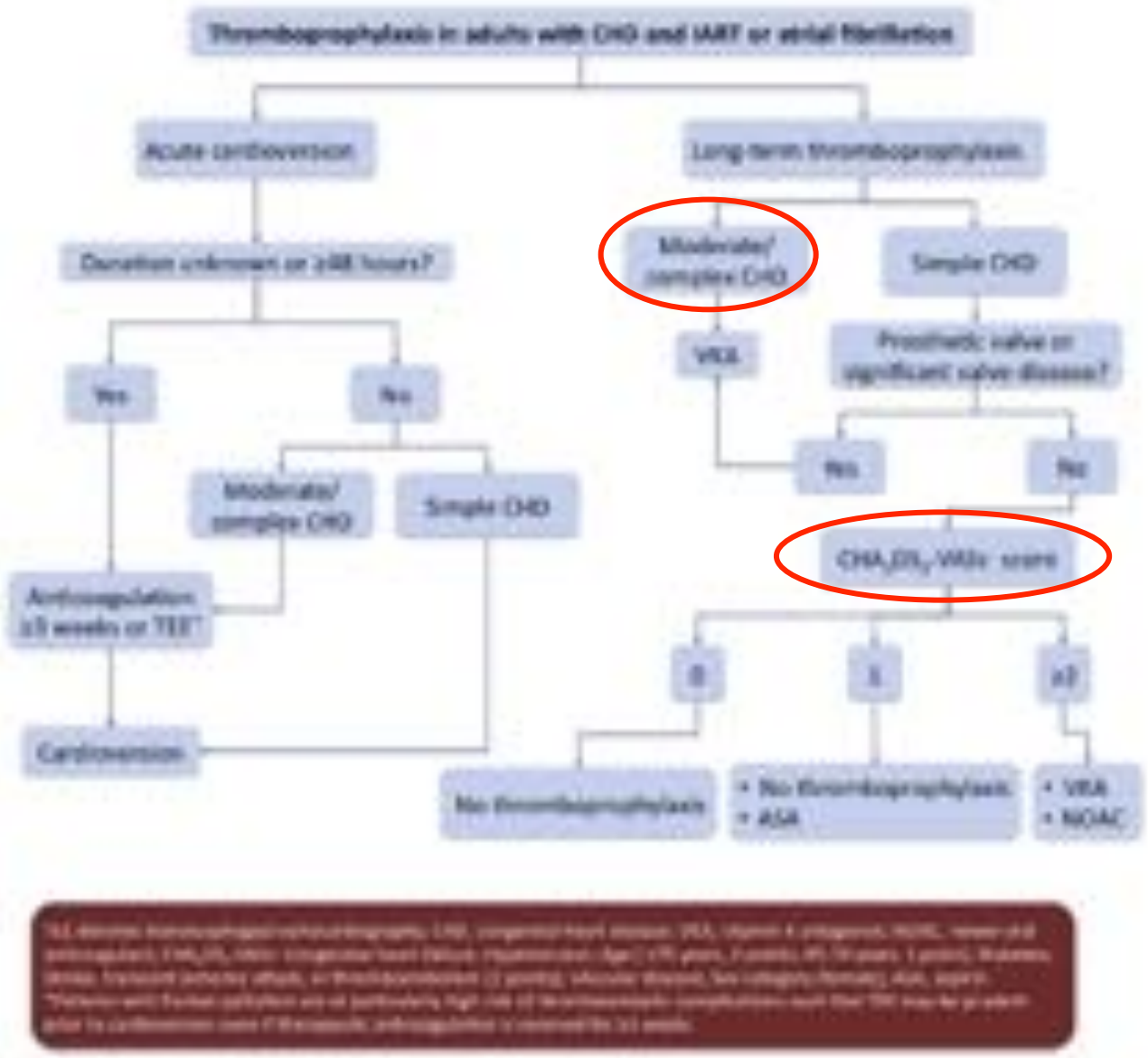
Clear goal for ablation
Good success rate of percutaneous ablation with contemporary tools (>90% acute success, 15% of recurrences)
Combes N et al. *Arch Cardiovasc Dis* 2017;110(5):334

Contemporary issues:

- Early detection to decrease related heart failure and mortality
- How to decrease incidence?

Large and early indication with avoidance of medical treatment
Abrams D et al. *HeartRyhthm* 2016;13:e251

Question of thromboembolic prevention in cases of supraventricular arrhythmias



Starting age?
NOAC? VKA? Other drug?
Duration after ablation?

Khairy P et al. Heart Rhythm 2014;11:e102

Atrial fibrillation: the last frontier

No sufficient data in CHD patients

Pranata R et al. *Indian Pacing Electrophysiol* J 2019;epub

Clear substrate? Right sided? Left sided with pulmonary veins?

Haemodynamic and common risk factors optimization +++

Poor efficacy and tolerance of anti-arrhythmic drugs

Percutaneous ablation with pulmonary veins isolation?

Simple

Low complication rate

Efficacy < 50%

Surgical treatment with Cox-Maze III?

Variable efficacy between 30 and 80%

High complication rate

Need for cardiac stimulation > 10%

Deal BJ, Mavroudis C.

Card Electrophysiol Clin 2017;9:329

Hybrid ablation?

Ventricular arrhythmias

Risk factors for ventricular tachycardia/sudden cardiac death

Standard clinical variables
Older age at time of repair
Prior large palliative shunts
Older chronologic age
Recurrent syncope
Pulmonary regurgitation
Residual pulmonary stenosis
Severe RV enlargement
Depressed RV function
Depressed LV function
High-grade ventricular ectopy on Holter or exercise test
Prolonged QRS duration on electrocardiogram (>180 ms)
Advanced testing
Positive ventricular stimulation at electrophysiology study
Large RV size on CMR
Large pulmonary regurgitant fraction on CMR

CMR = cardiac magnetic resonance; LV = left ventricular; RV = right ventricular.

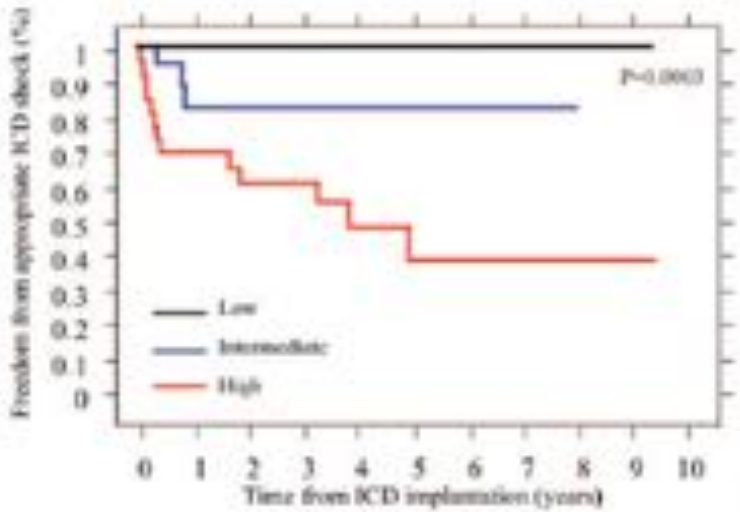
strong predictive negative accuracy but only fair predictive positive accuracy

Villafane J et al. *J Am Coll Cardiol* 2013;62:2155

No clear consensus for ventricular arrhythmias risk startification in these patients

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

Khairy P et al. *Circulation* 2008;117:363



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

Utility of invasive electrophysiological study for intermediate risk?

With MRI

Variable	Points
Prior palliative shunt	2
QRS duration ≥180 ms	1
Ventriculotomy incision	2
Previous VT	2
LV EF <45%[a]	2
RV EF <30%[a]	3
Total points	0-12

Bokma JP et al. *JAMA Cardiol* 2017;2:618

Cohort analysis : low arrhythmias rate in primary prevention with this approach

Probst J et al. *Int J Cardiol* 2018;15:269

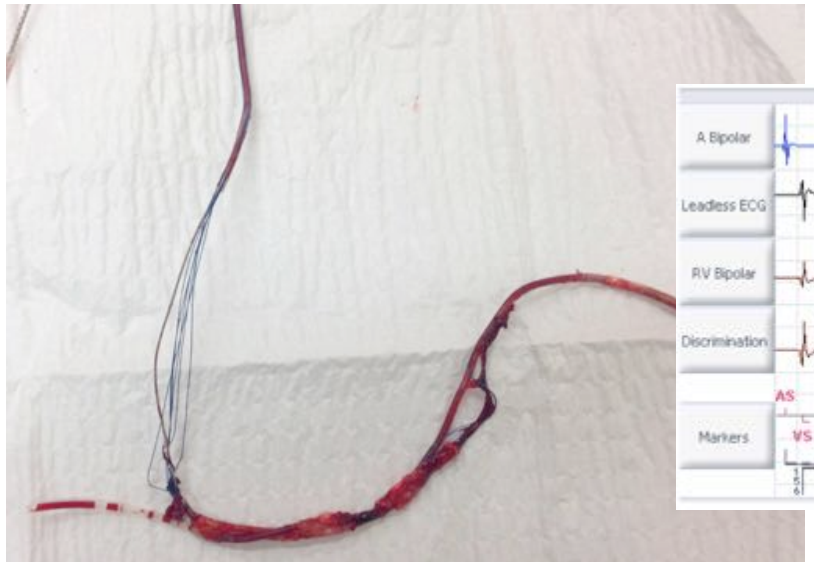
ICD in Fallot patients

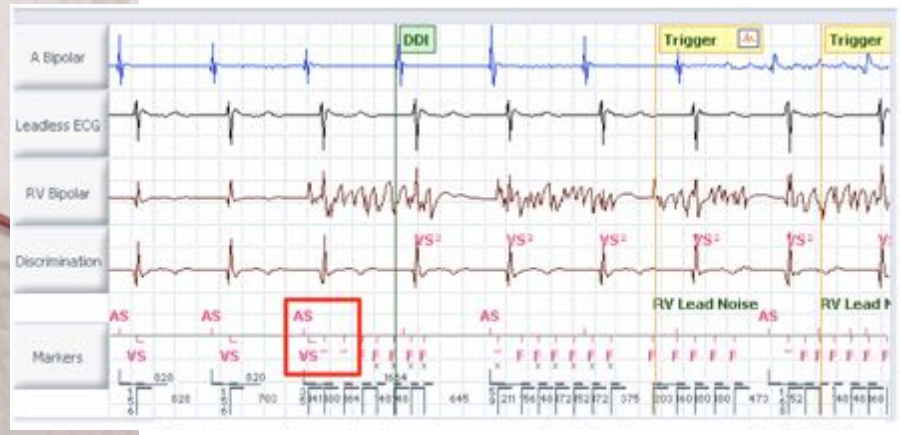
Secondary prevention : aborded SCD, VT with haemodynamic compromise

Primary prevention???

Problem of complication rate +++

- Technical problem (++ lead revision) : up to 30% after 4 years
- Inappropriate therapies: up to 6% after several years





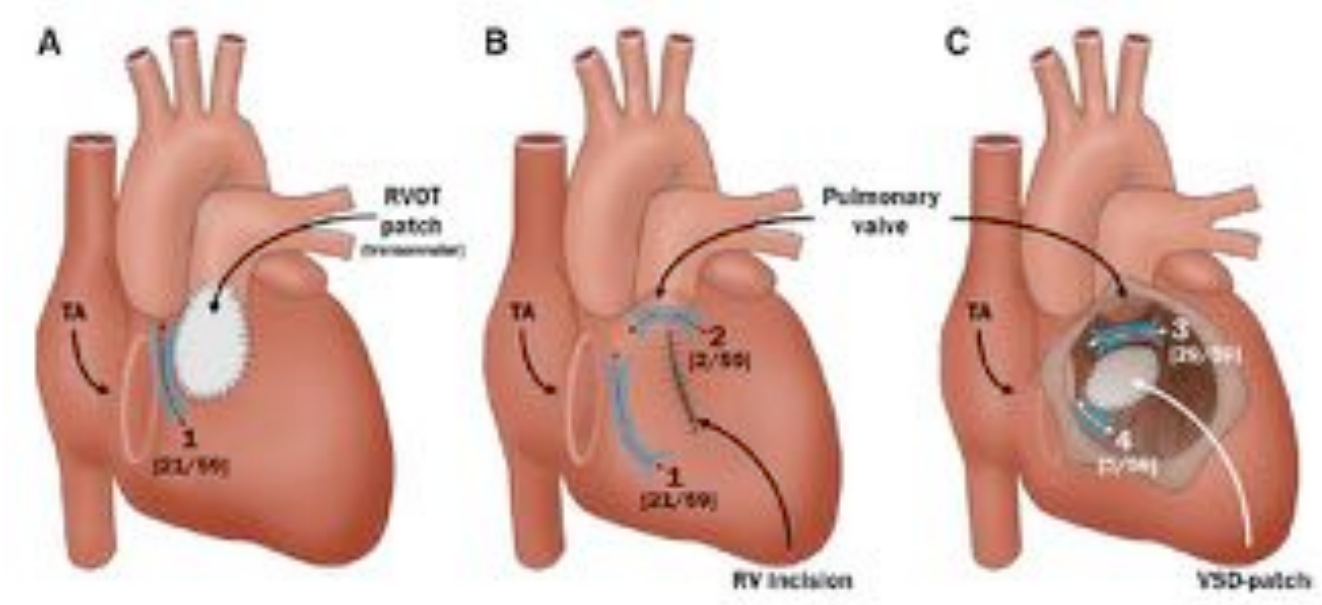
Khairy P et al. Circulation 2008;117:363
DAIT4F French registry in process

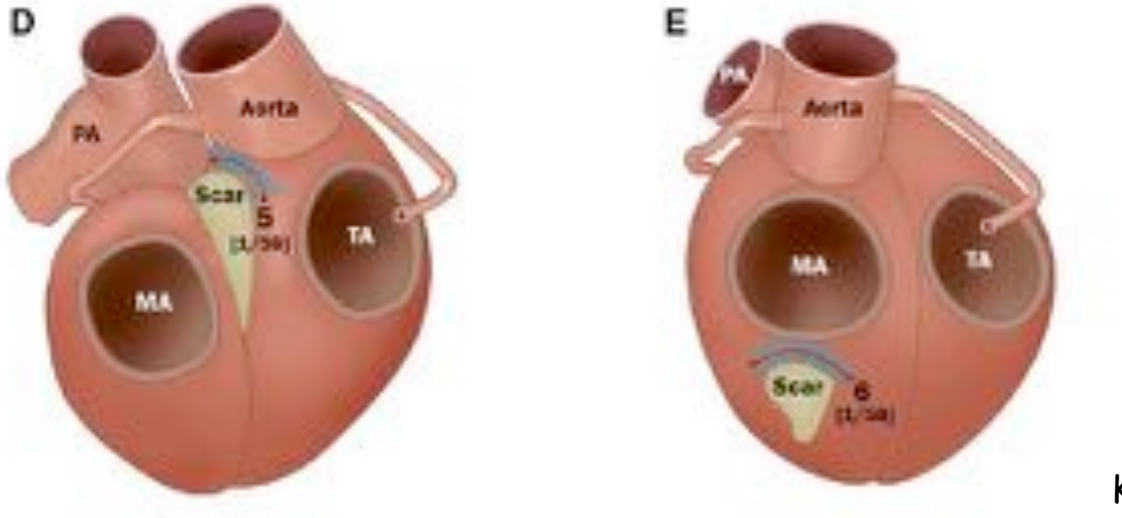
Decrease complication rate of ICD with subcutaneous ICD?

Actually no stimulation for bradycardia / anti-tachycardia pacing
Caution with low weights < 25 kgs
Risk of inappropriate shocks in young patients

Moore JP et al. *Circ Arrhythm electrophysiol* 2016;9

Mecanisms of ventricular arrhythmias in Fallot Patients



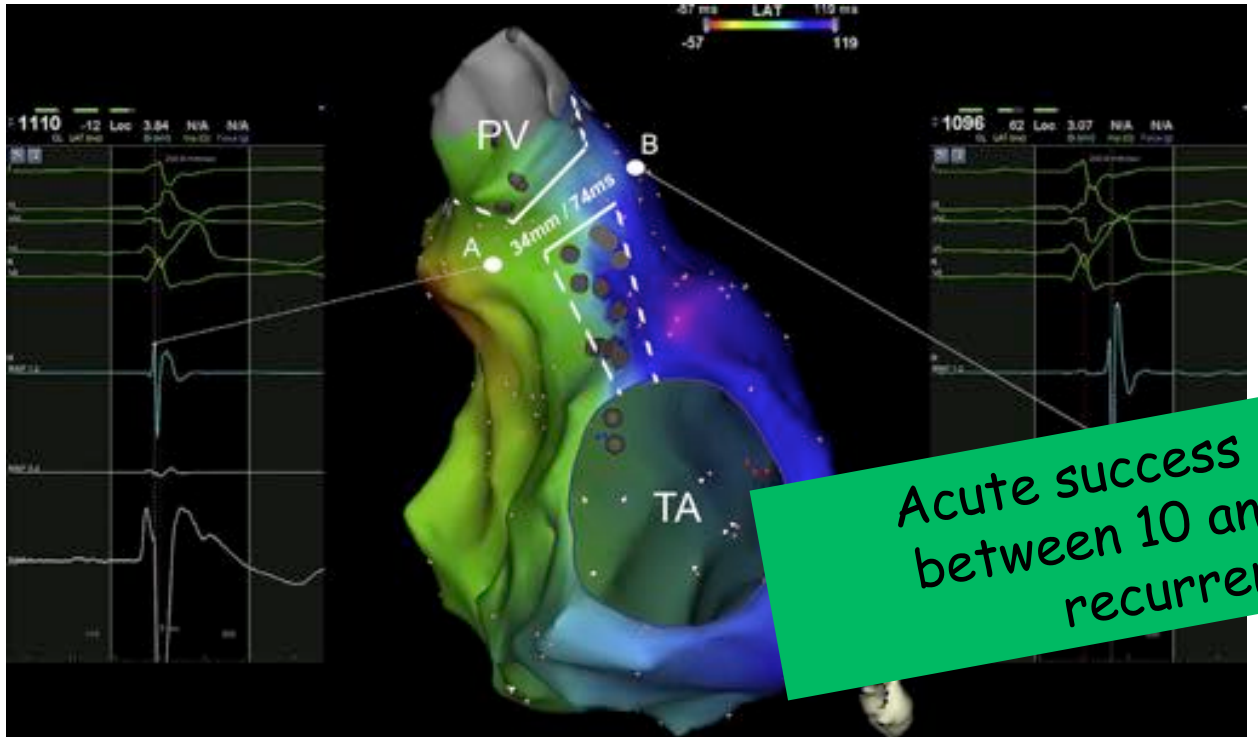


VT isthmuses are now well known
Most frequent : isthmus 3

Zappenfeld K et al. Circulation 2007;116:2241-2252
Kapel GF et al. Circ Arrhythm Electrophysiol 2015;8:102-109

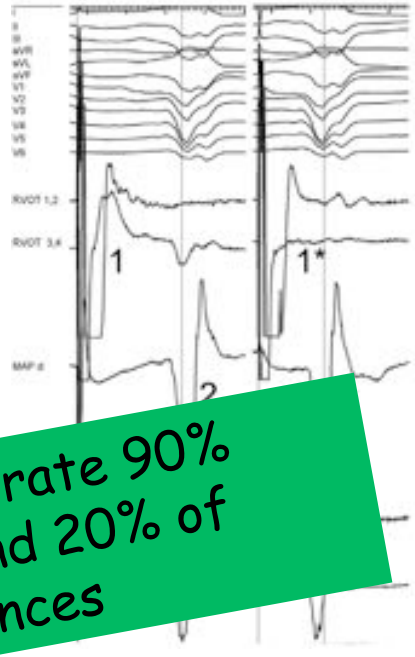
Utility of electrophysiologic study in the management of ventricular arrhythmias

Precise substrate location

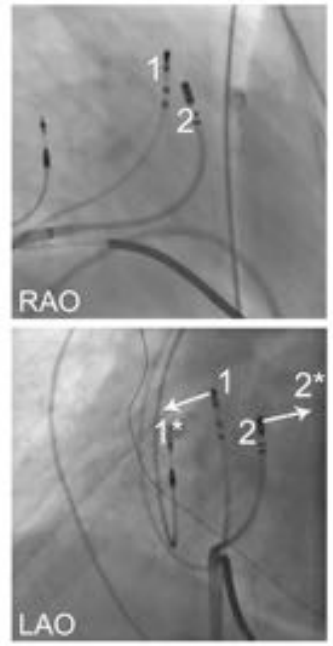


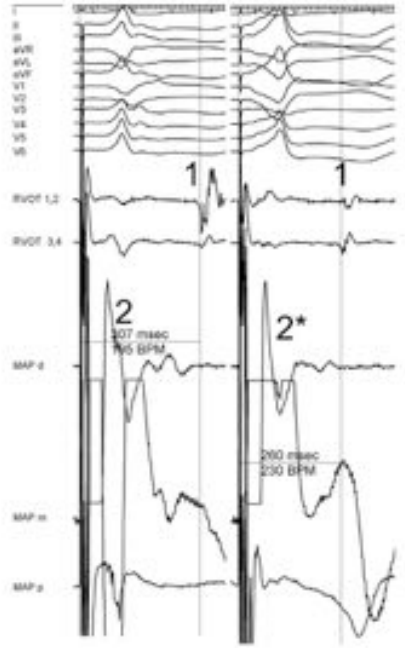
Scar / patches location
Slow conducting channel location
Activation map during VT

VT ablation and block lines validation



Acute success rate 90% between 10 and 20% of recurrences



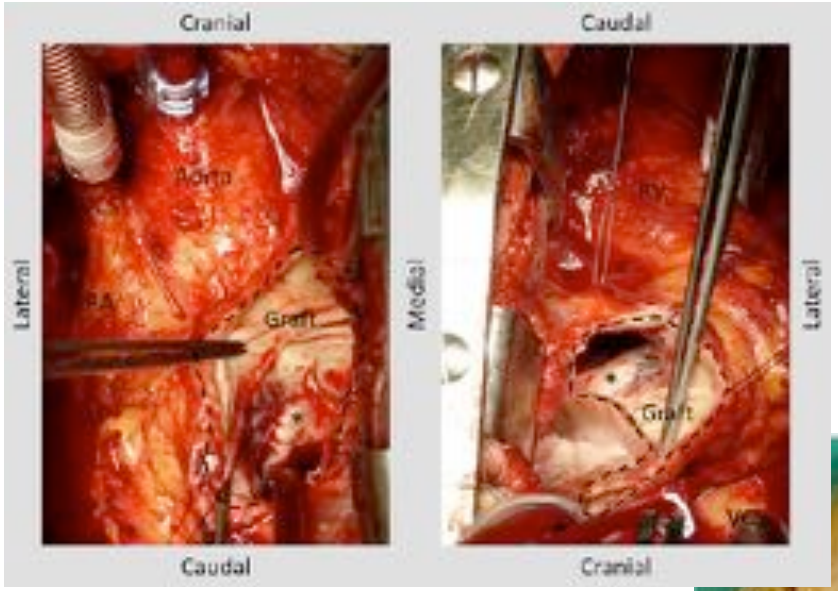


Zeppenfeld K et al. Card Electrophysiol Clin 2018

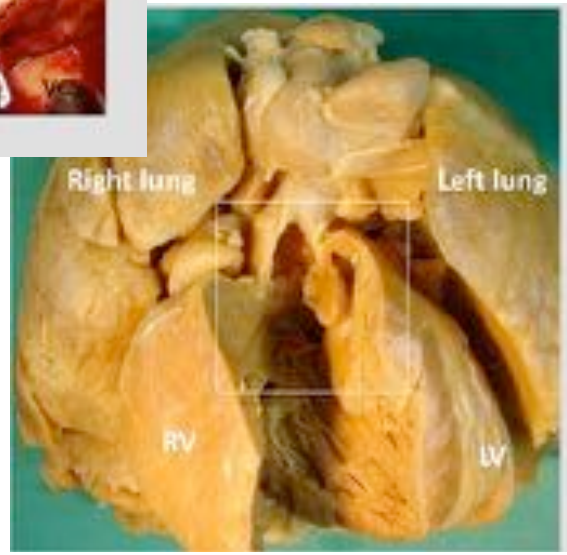
Gonska BD et al. Circulation 1996;94:1902-8
Morwood JG et al. Heart Rhythm 2004;1:301-8
Zeppenfeld K et al. Circulation 2007;116:2241-52
Kriebel T et al. Am Coll Cardiol 2007;50:2162-68
Schneider HE et al. J Cardiovasc Electrophysiol 2012;23:930-37
Kappel GF et al. Circ Arrhythm Electrophysiol 2015;8:102-9
Van Zyl M et al. Heart Rhythm 2016;13:1449
Larredo M et al. Arch Cardiovasc Dis 2017;110:292

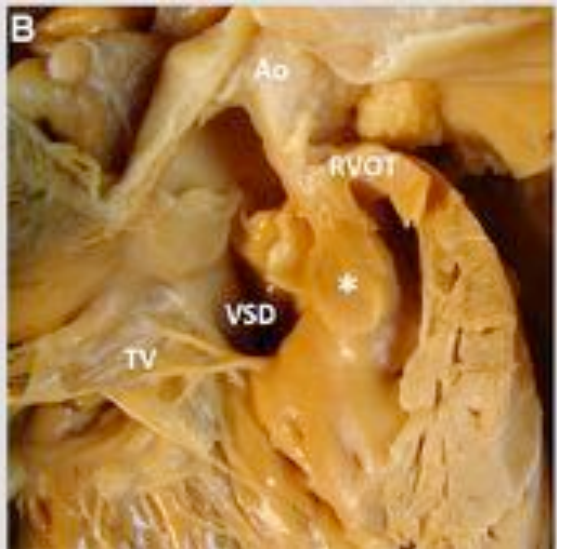
Causes of percutaneous ablation failure

Valvular prosthesis / homografts



Kapel GF et al. *Circ Arrhythm elctrophysiol* 2014;7:889



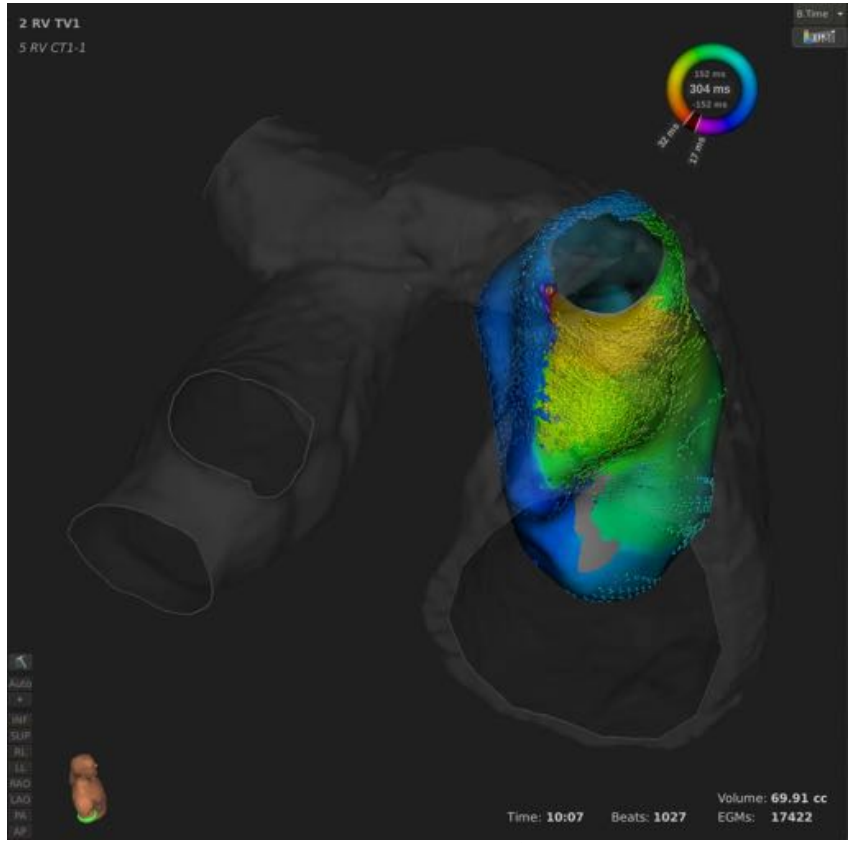


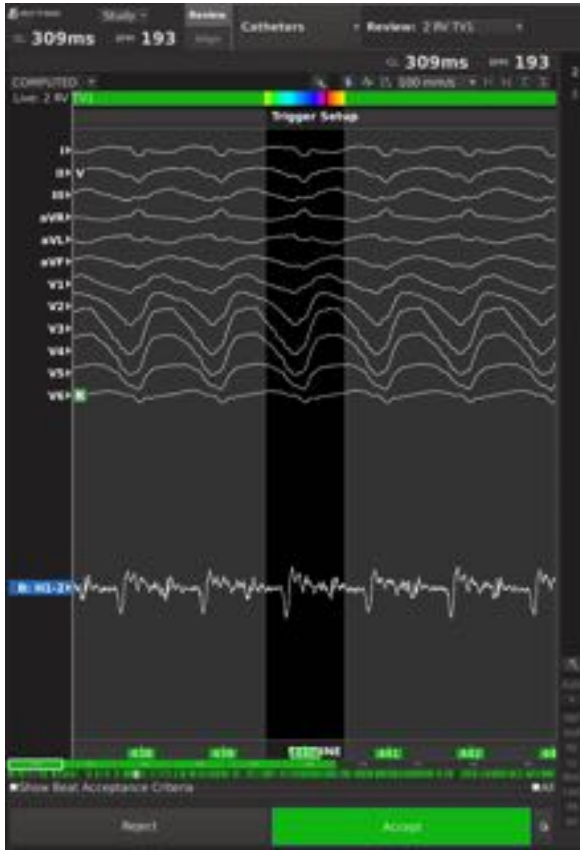
Conduction tissu proximity

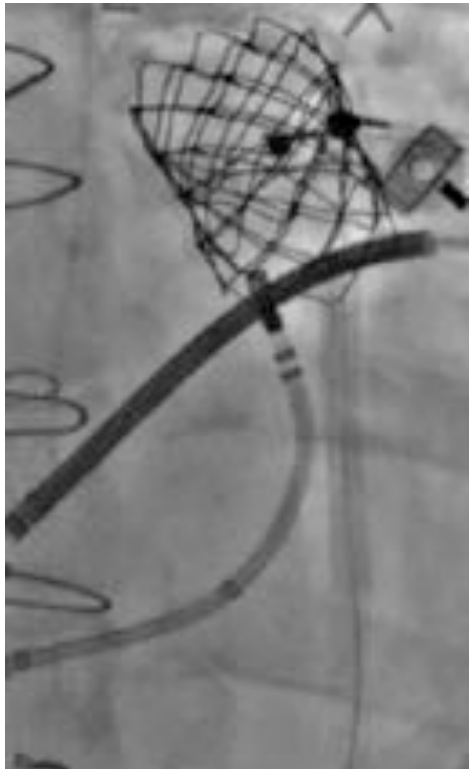
Local Hypertrophy, calcifications / epicardial source

Causes of percutaneous ablation failure

Caution with percutaneous pulmonary valves with possible arrhythmia « protected isthmus »







Combes N. et al. Eur Heart J 2019;epub

Discussion of arrhythmia evaluation before surgery in adults ++

Ventricular arrhythmias surgical treatment

With redo surgery with primary heamodynamic indication

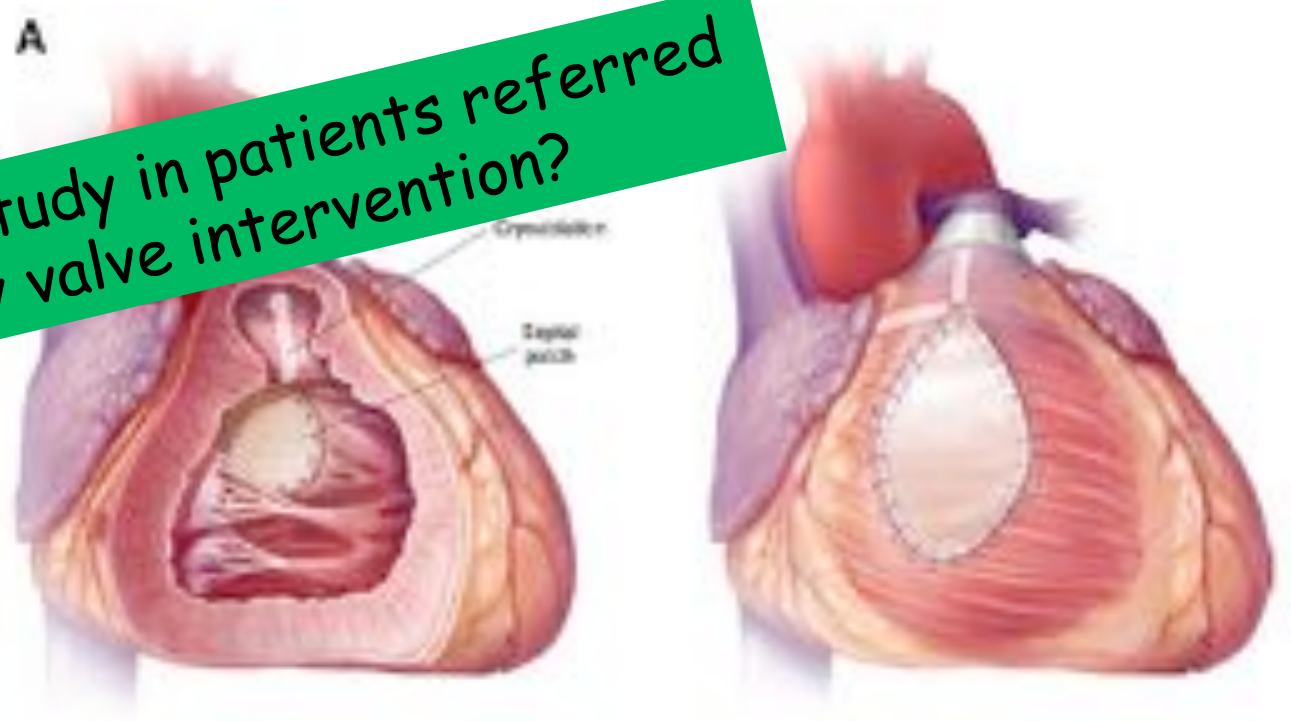
Ideally:

- guided by electrophysiological study before : isthmuses location
- With lesion control for achievement of complete block of couduct

When perform EP study in patients referred for pulmonary valve intervention?

Indic ion:

- Secondary prevention
- Primary prevention in selected patients?
  - Systematically?



Rotes AS et al. Circ Arrhythm Electrophysiol 2015;8:110-16

Conclusion

A life-long burden of arrhythmias in Fallot patients

An increasing knowledge of arrhythmias mecanisms

An increasing efficacy of interventional treatment with ablation therapies

A lot of improvement needed

- Early detection of arrhythmias
- Complications of PMK /ICD
- A better assessment of SCD risk

Muldisciplinary staffs with congenital cardiologists, cardiac surgeons, electrophysiologists to increase arrhythmia prognosis in CHD patients