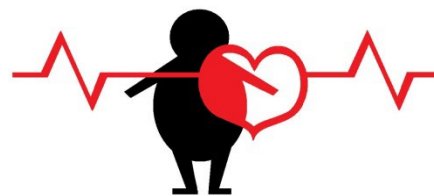


Ventricular arrhythmias ablation in congenital heart disease

Nicolas Combes
ncombes@clinique-pasteur.com



Département de rythmologie
et de stimulation cardiaque



Pôle de cardiologie pédiatrique
et congénitale



Conflict of interest

- Consultant : Meda Pharma, Medtronic, Microport.
- Clinical studies : Biotronik, Boston Scientific, Medtronic, Zoll


CHD associated with ventricular arrhythmias

Relative Risk for Specific Arrhythmias in Common Congenital Heart Defects

	IART	AF	WPW	VT/SCD	SA Node Dysfunction	Spontaneous AV Block	Traumatic AV Block
VSD	+			+			+
ASD	+	+					
TOF	++			++			+
AS		+		++			+
D-TGA (M&S)	+++			++	+++		
CAVC	+					+	++
SING V (F)	+++	+		+	+++		
L-TGA	+		++	+		++	+++
Ebstein's anomaly	++		+++	+			


AF indicates atrial fibrillation; WPW, Wolff-Parkinson-White syndrome; SCD, sudden cardiac death; SA, sinoatrial; VSD, ventricular septal defect; ASD, atrial septal defect; TOF, tetralogy of Fallot; AS, aortic stenosis; M&S, after the Mustard or Senning operation; CAVC, common AV canal defect; SING V (F), single ventricle after the Fontan operation; + + +, high risk; + +, moderate risk; and +, slight risk.

Specific ventricular arrhythmias in CHD



Macroreentrant monomorphic VT secondary to developmental and surgical abnormalities within ventricular muscle that create corridors of slow conduction:

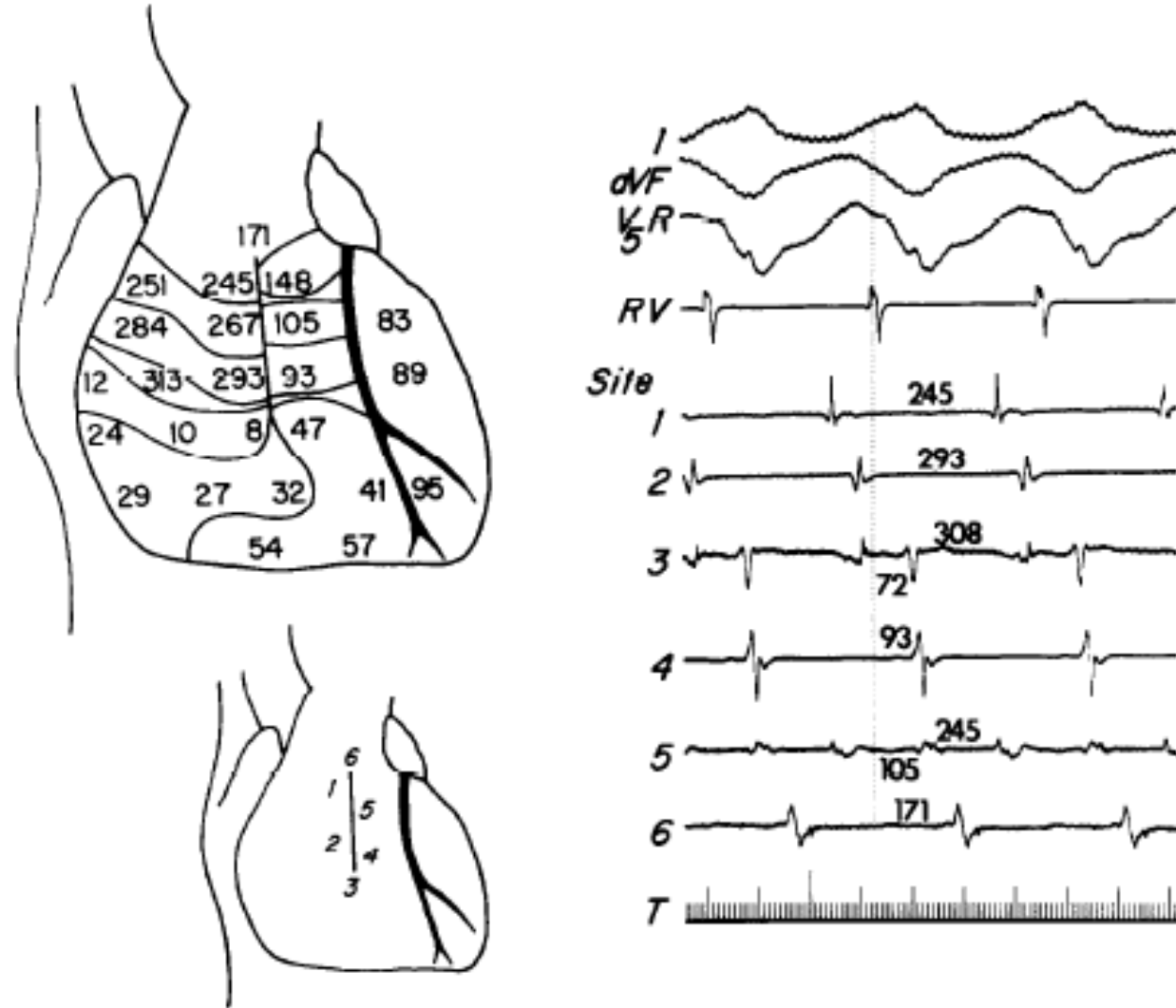
- Tetralogy of Fallot +++
- Ventricular septal defect after surgery
- Ebstein anomaly



Polymorphic VT and VF with diffuse abnormal myocardium (Long-standing Pressure and volume loads, cyanosis):

- Congenital aortic outflow tract obstruction
- Transposition of great arteries with atrial switch
- Tetralogy of Fallot with advanced ventricular dysfunction
- unrepaired ventricular septal defect with Eisenmenger physiology
- palliated single ventricle

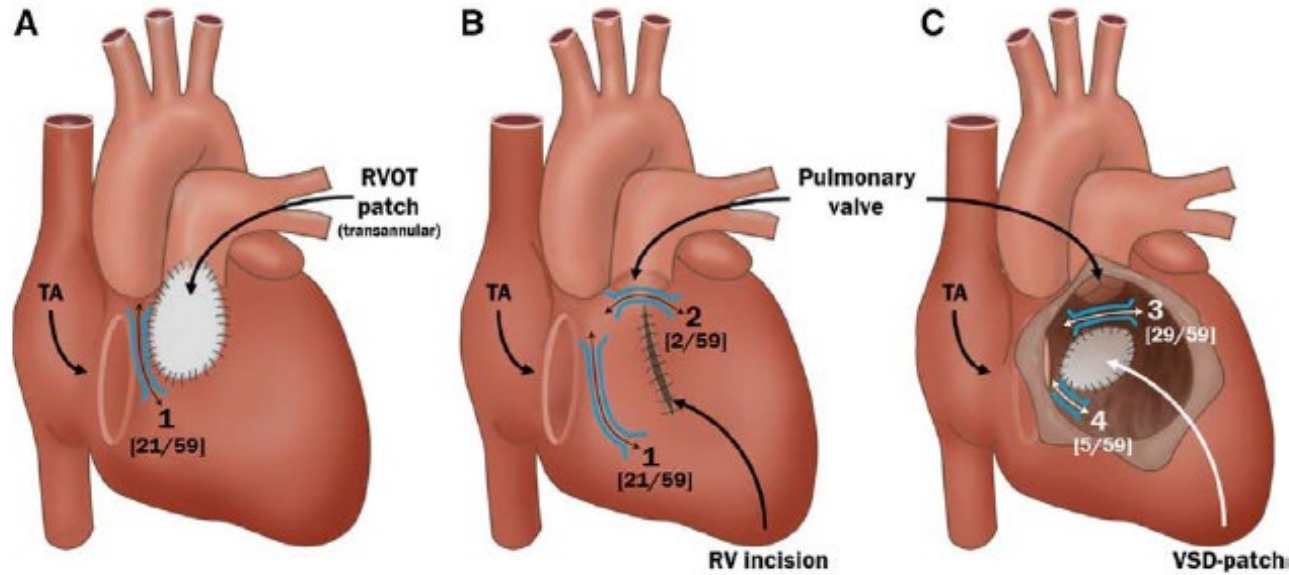
Substrate for monomorphic VT in CHD: scar related
Horowitz L et al. Am J Cardiol 1980;46:446-452



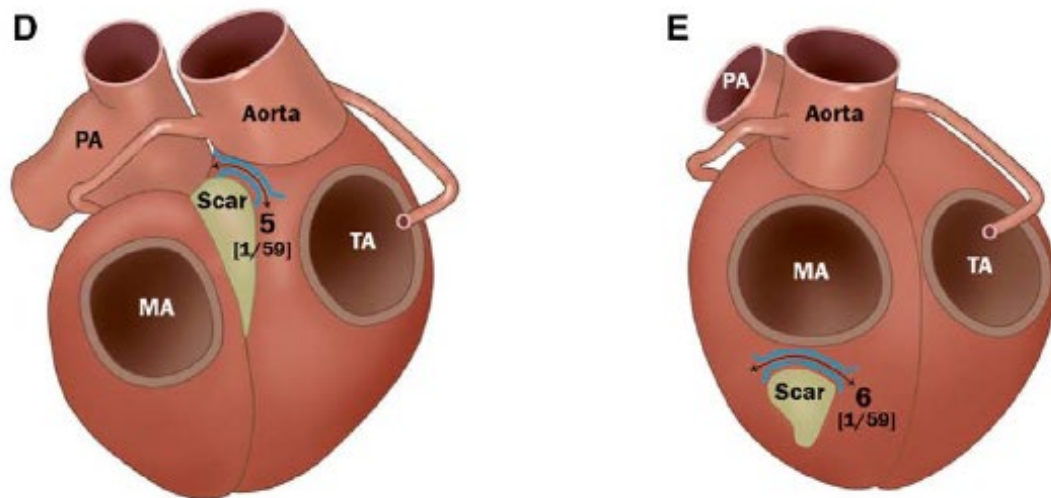
Reentry with right ventricular scar in a TOF patient

Location of critical VT isthmus?

Back to cardiopathy and cardiac surgeries



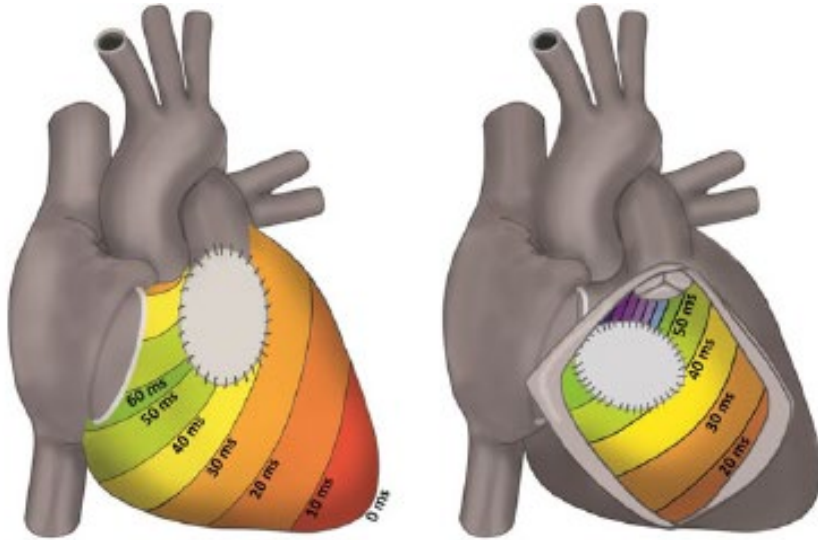
+++ position of the conal septum in TOF



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

Location of critical VT isthmus?

B



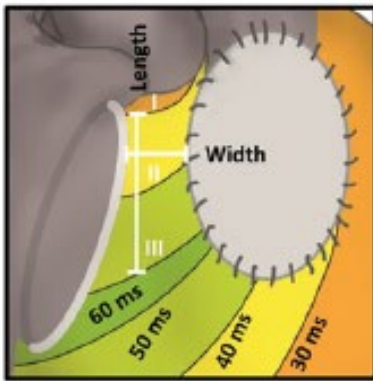
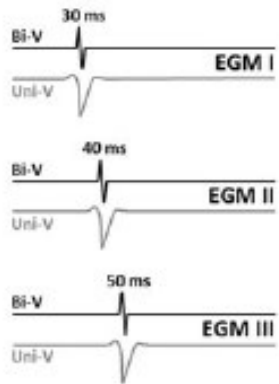
TOF patients isthmuses with monomorphic VT

Longer: 22 ± 7 vs 16 ± 7 mm $p=0.001$

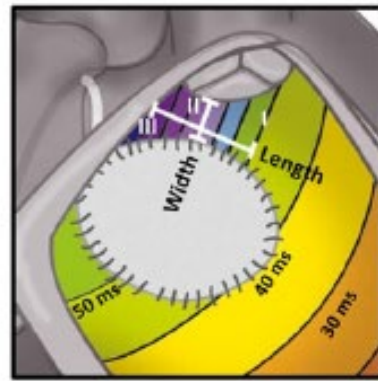
Narrower: 20 ± 8 vs 28 ± 11 mm $p<0.001$

Lower Conduction Velocity Index:
 0.36 ± 0.34 vs 0.78 ± 0.24 m/s

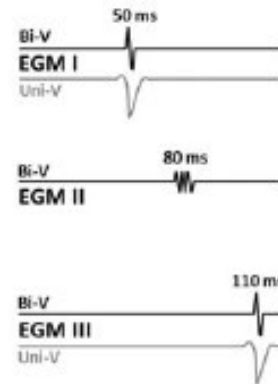
C



$CV_i = \text{length (mm)} \div \text{time (ms)}$
 $CV_i = 20 \text{ mm} / (50 - 30 \text{ ms}) = 1.0 \text{ m/s}$

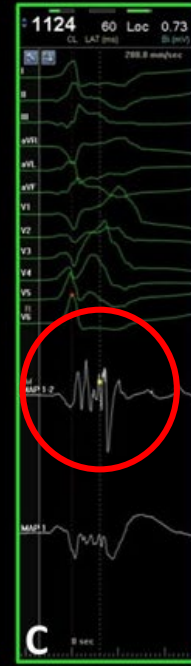
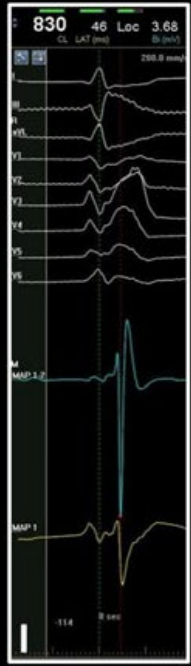


$CV_i = \text{length (mm)} \div \text{time (ms)}$
 $CV_i = 20 \text{ mm} / (100 - 50 \text{ ms}) = 0.33 \text{ m/s}$



Isthmus 1 & 2 - 'normal'

Isthmus 3 - 'abnormal'

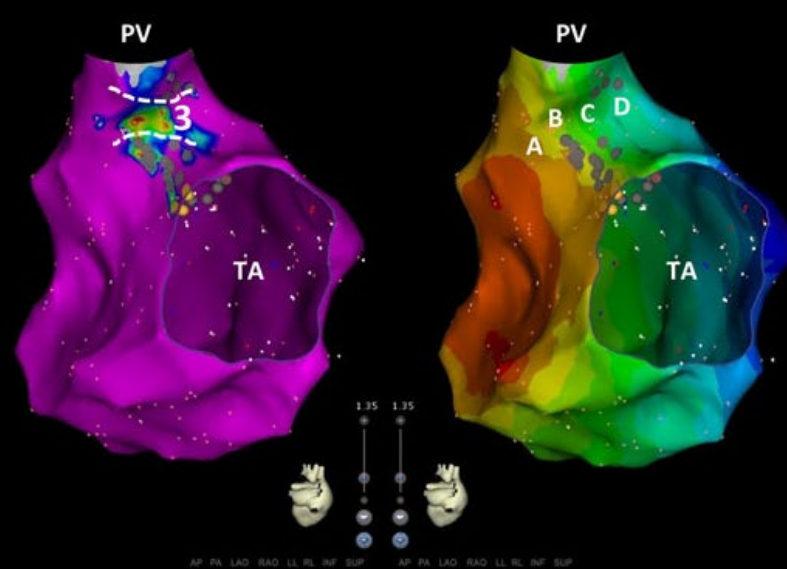
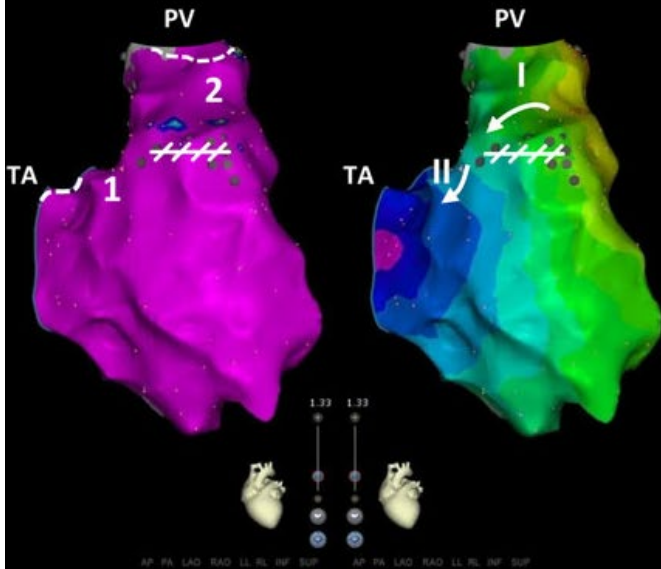


0.24 mV BI 11.86 mV
0.55 1.56

-37 ms LAT 126 ms

0.17 mV BI 12.61 mV
0.52 1.49

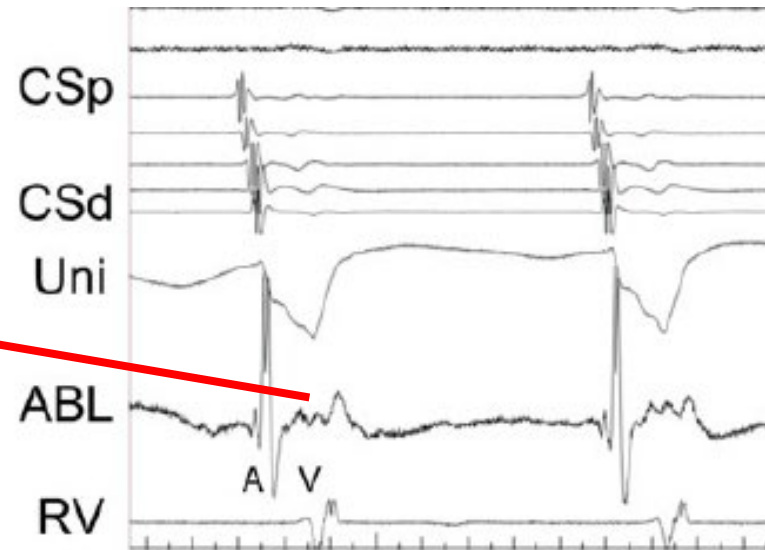
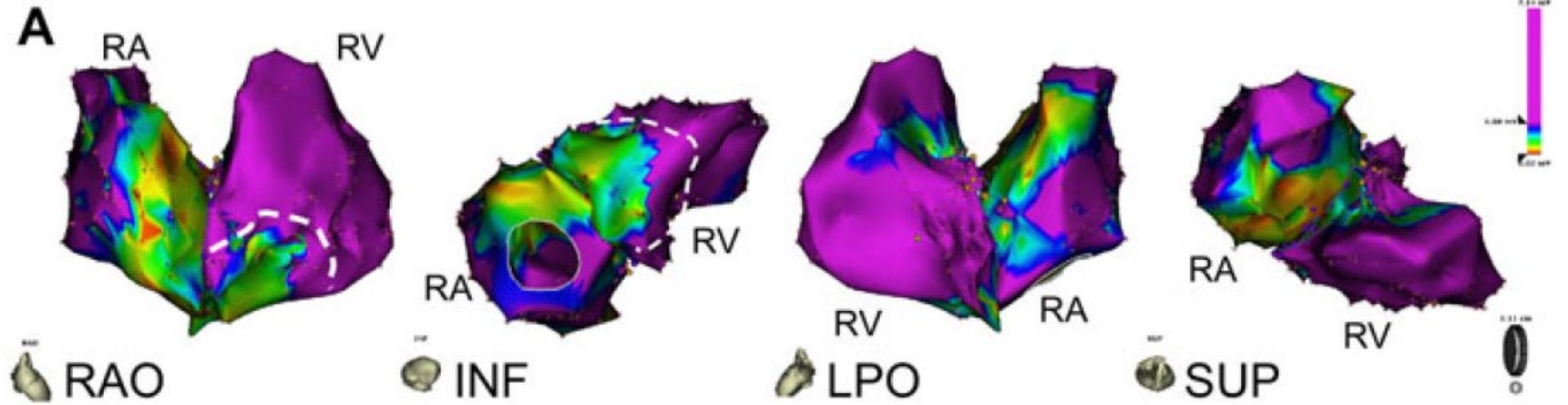
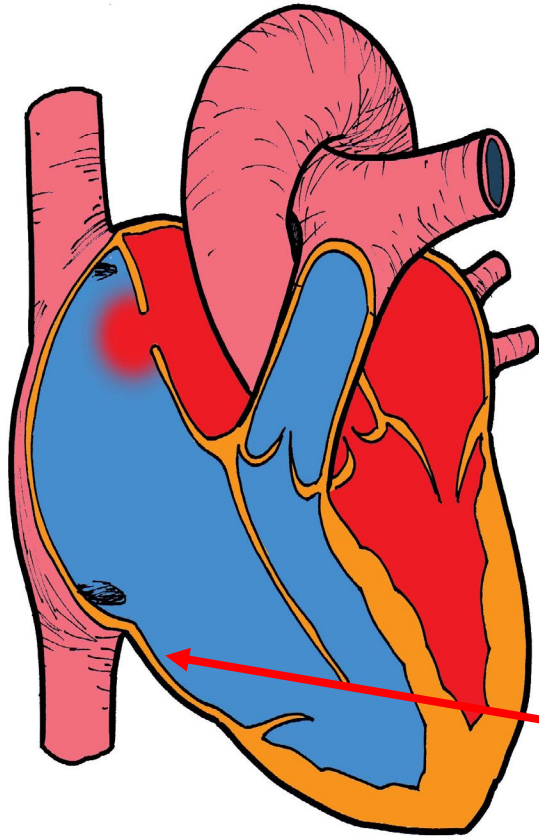
-16 ms LAT 140 ms



Slow conducting isthmus with abnormal local electrogram in sinus rhythm

Monomorphic VT substrate in Ebstein's anomaly

Abnormal ventricular voltage in atrialized right ventricle

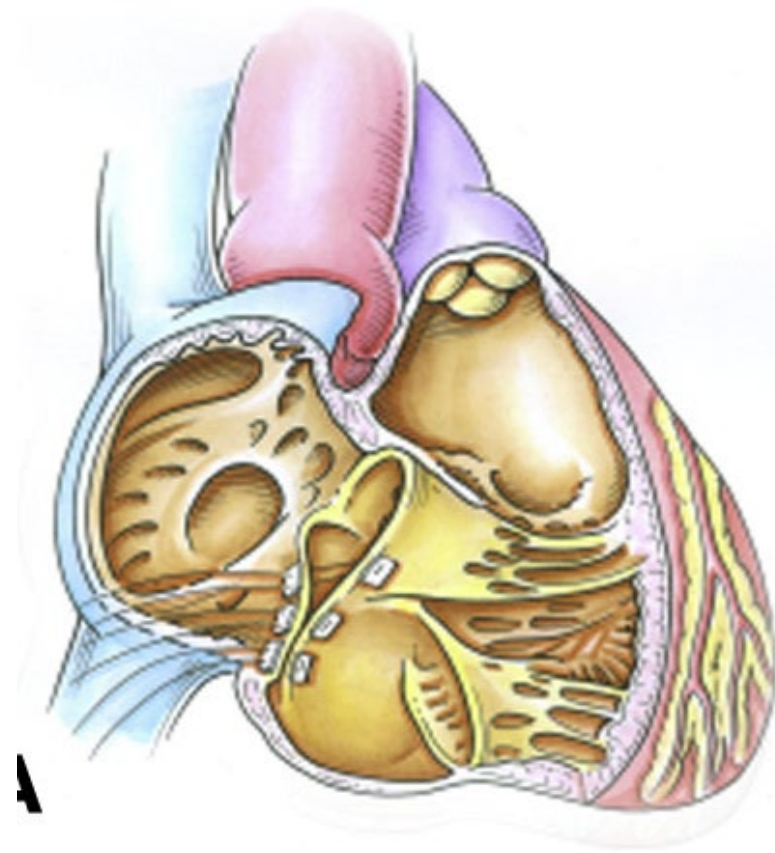
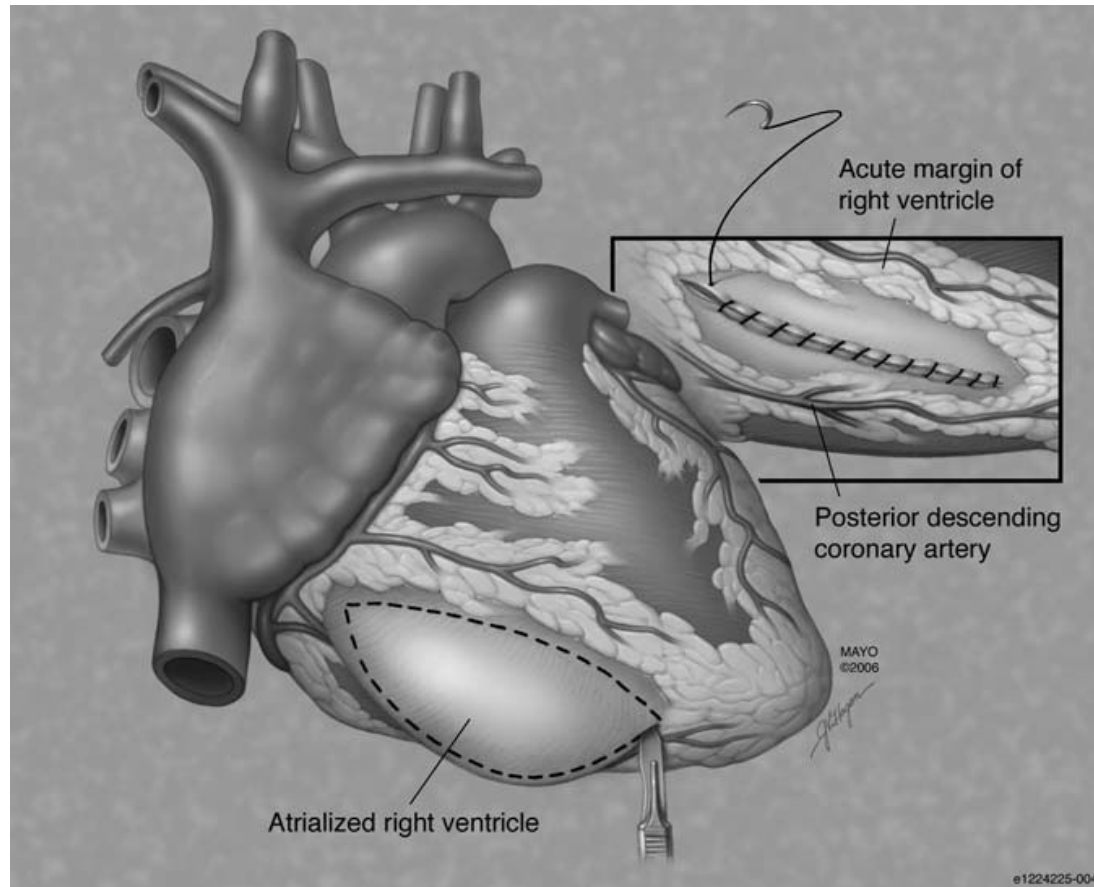


Guo XJ et al. J Cardiovasc Electrophysiol 2015;26:404

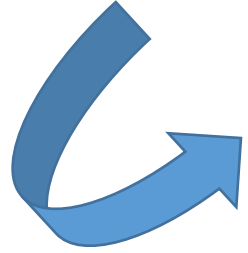
Monomorphic VT substrate in Ebstein's anomaly

After surgery: right ventricle plasty with excision or plication of the atrialized chamber

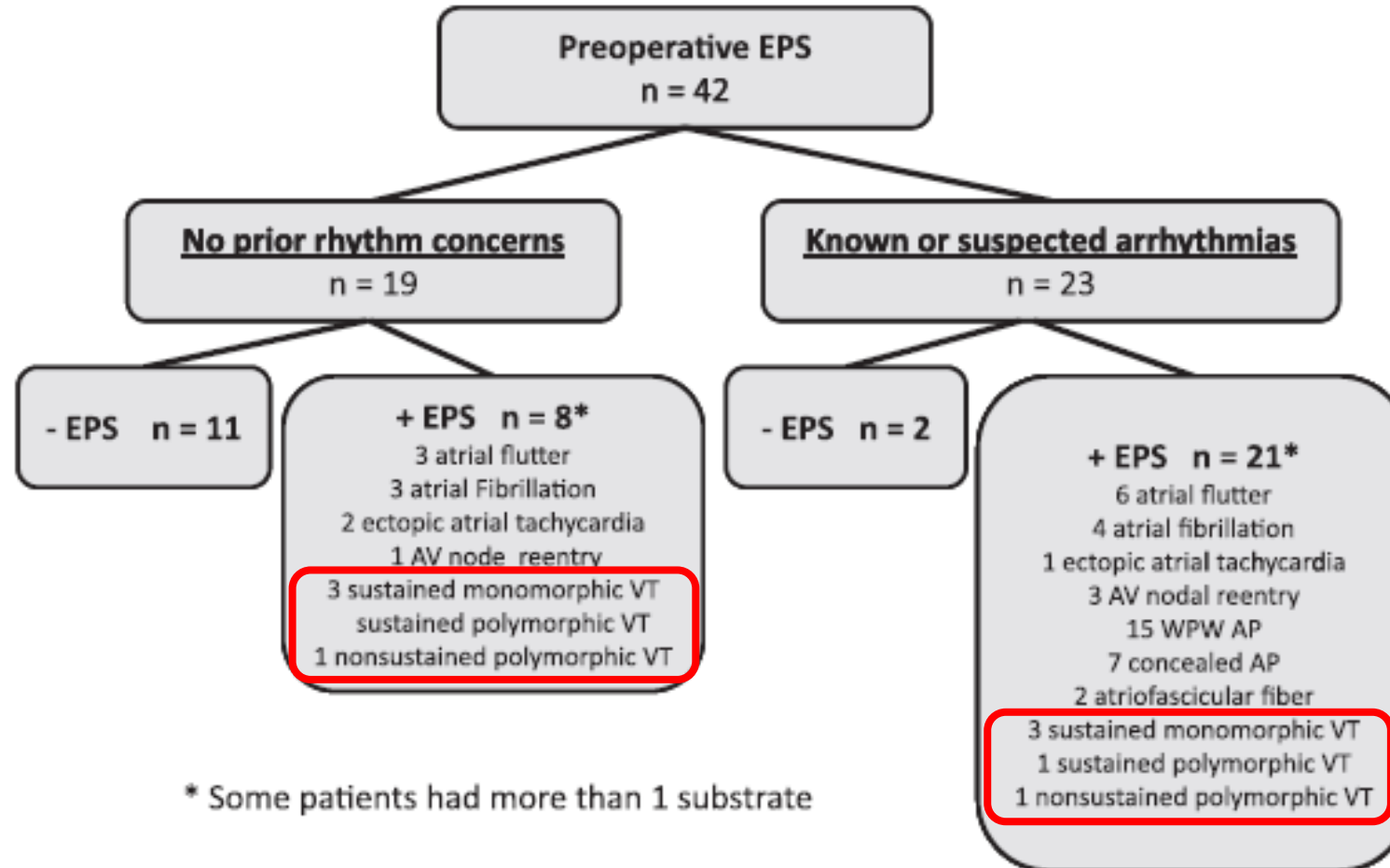
Important difficulties to reach the substrate with percutaneous access.



Monomorphic VT substrate in Ebstein's anomaly



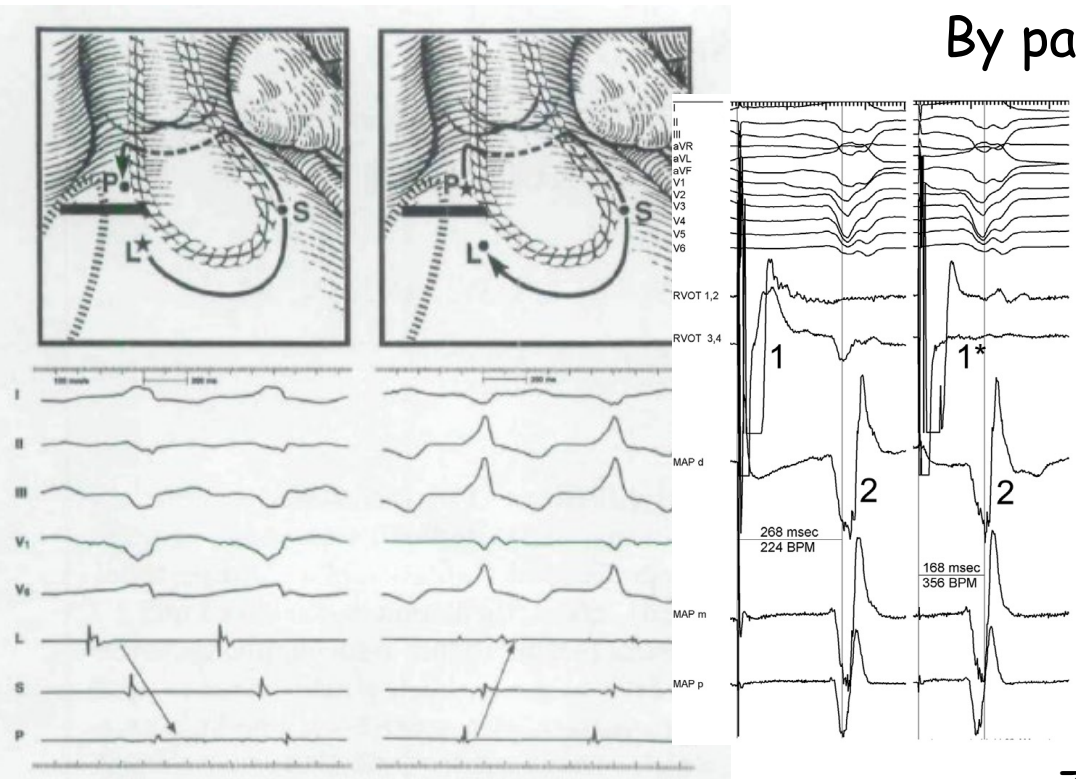
Preoperative evaluation +++ / ablation
Shivapour JK et al. Heart rhythm 2004;11:182-86



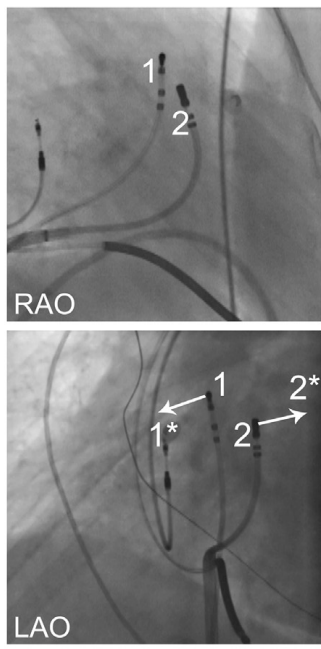
* Some patients had more than 1 substrate

After interruption of VT during ablation, validation of isthmus block is needed

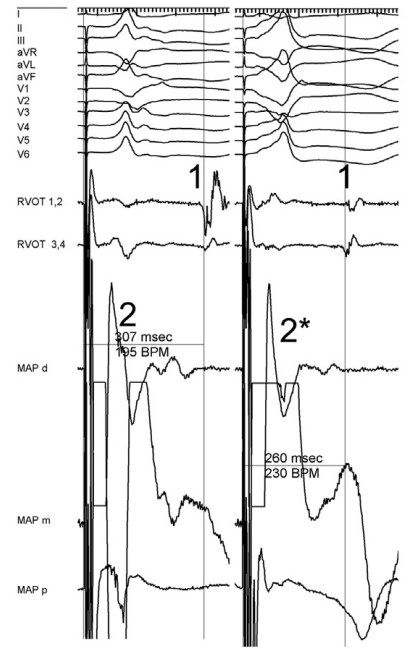
By pacing



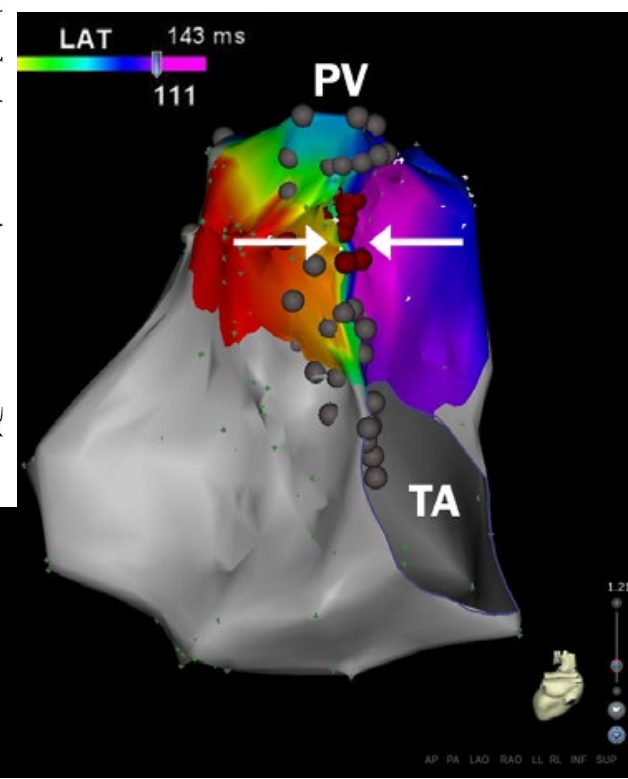
Horton RP et al.
J Cardiovasc Electrophysiol
1997;8:432-435



Zeppenfeld K et al.
Card Electrophysiol Clin 2018



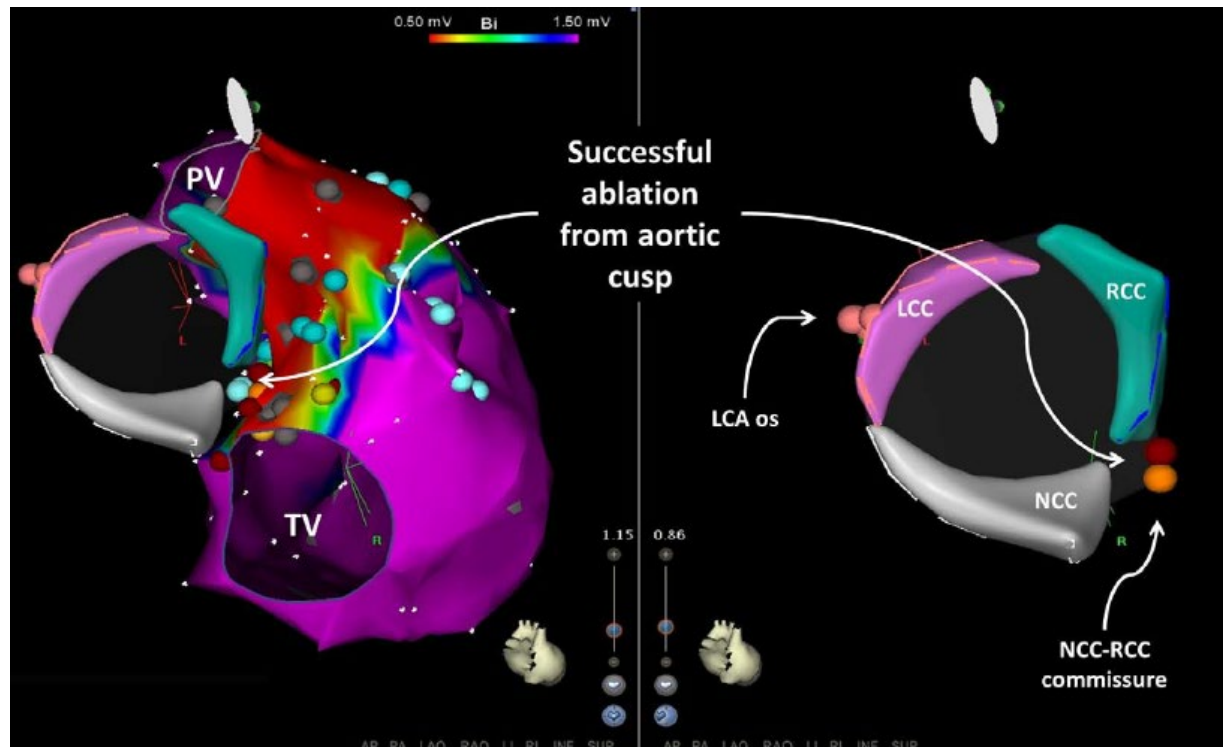
By 3D activation mapping



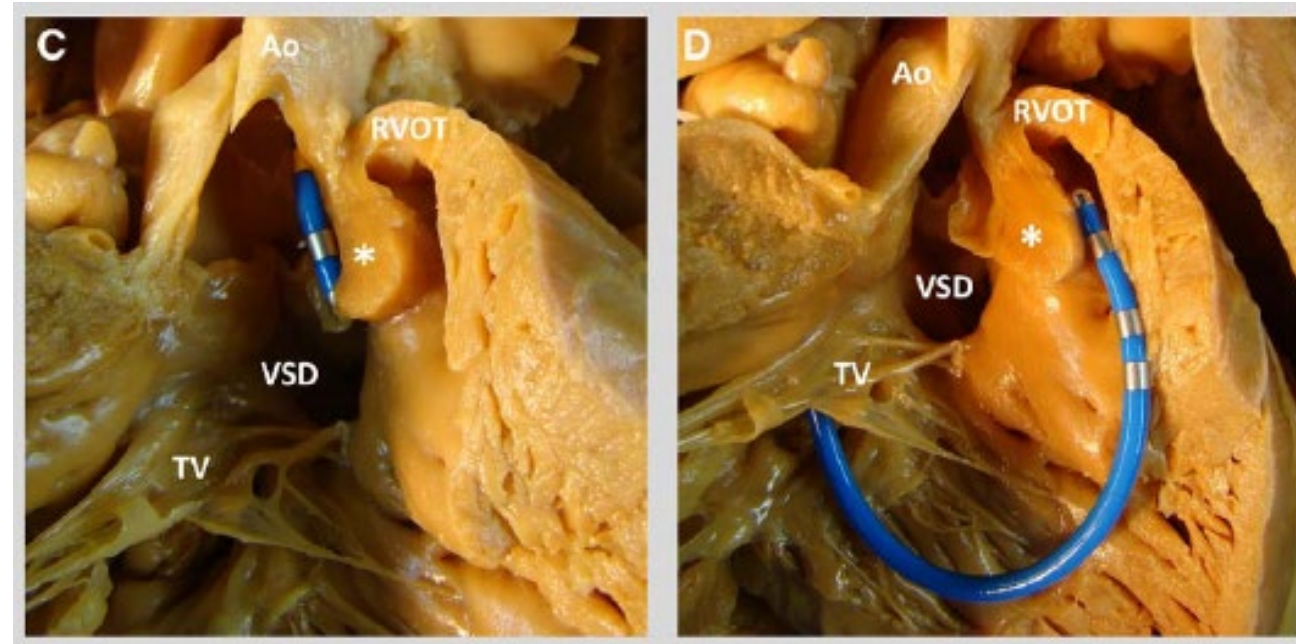
Kapel GF et al.
Circ Arrhythm Electrophysiol
2015;8:102-109

Causes of percutaneous ablation failure

Think about left side in right ventricular tachycardias

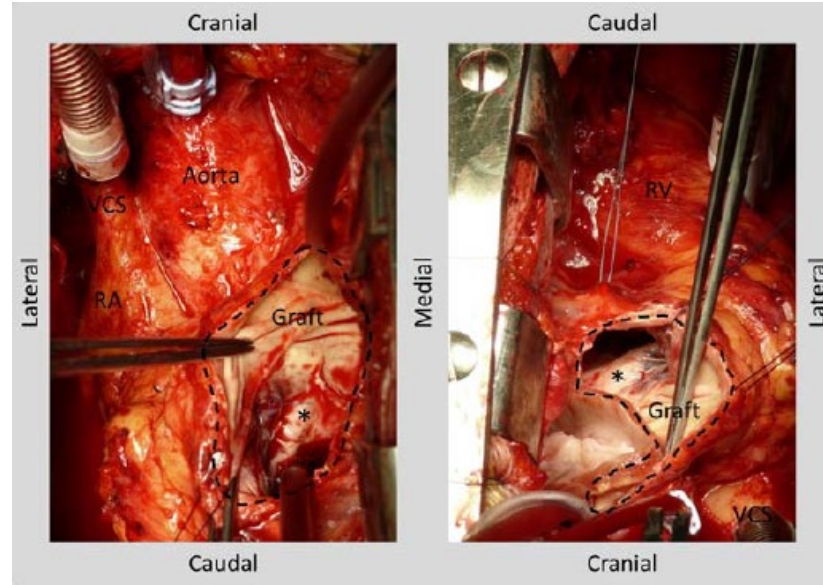
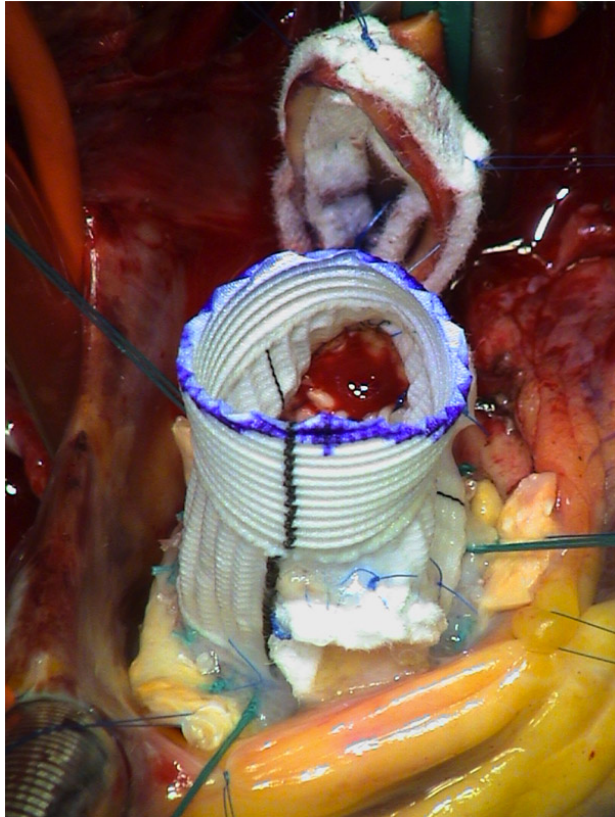


Kapel GF et al. *Circ Arrhythm Electrophysiol* 2014;7:889

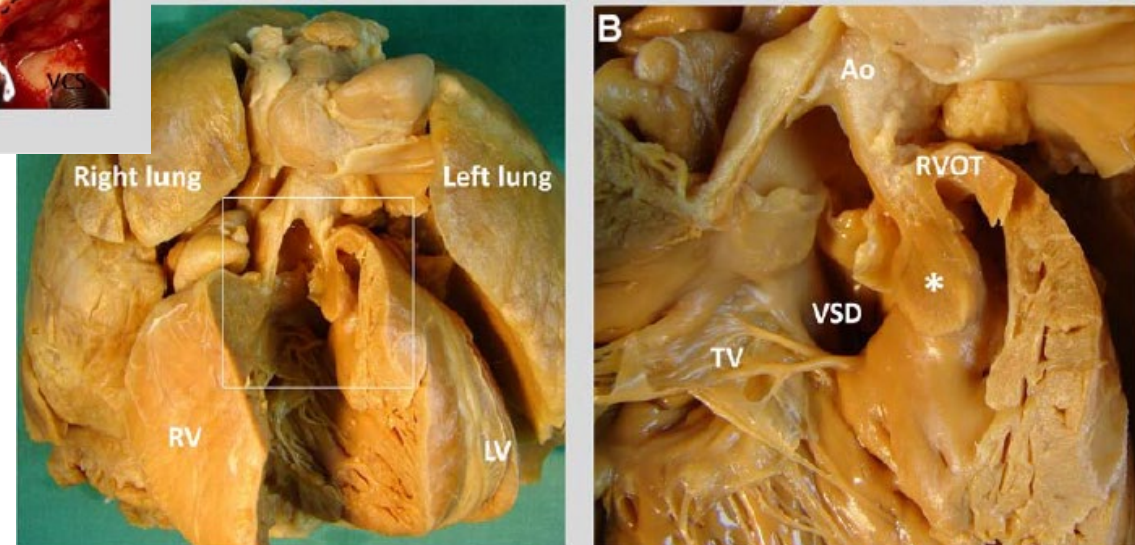


Causes of percutaneous ablation failure

Valvular prosthesis / homografts



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



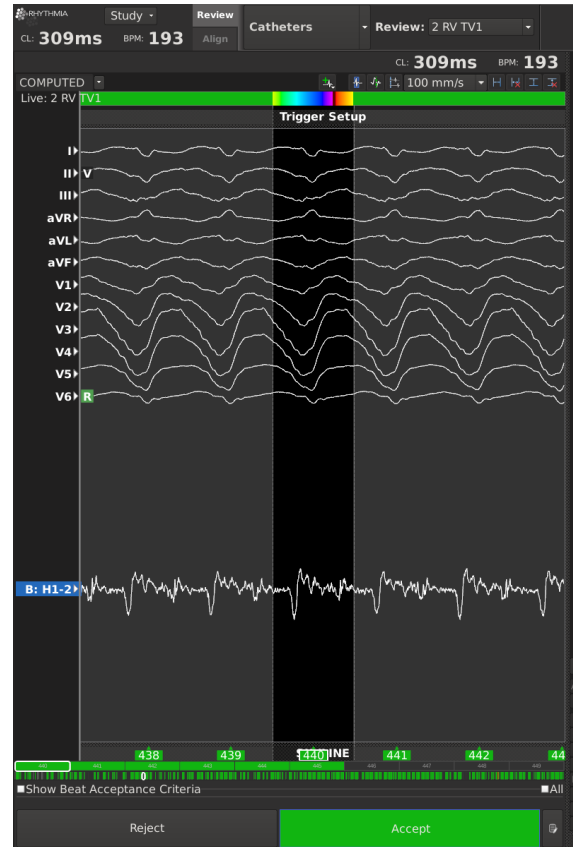
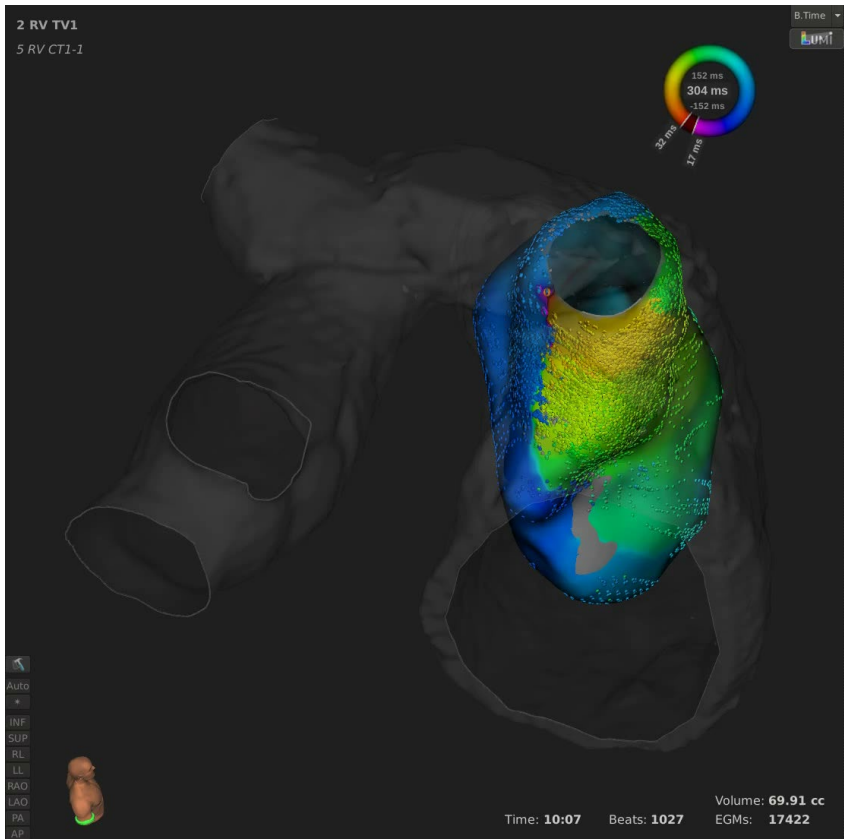
Conduction tissue proximity

Local Hypertrophy, calcifications / epicardial source

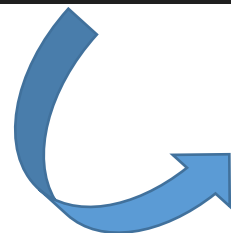
Access to cardiac chamber of interest

Causes of percutaneous ablation failure

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

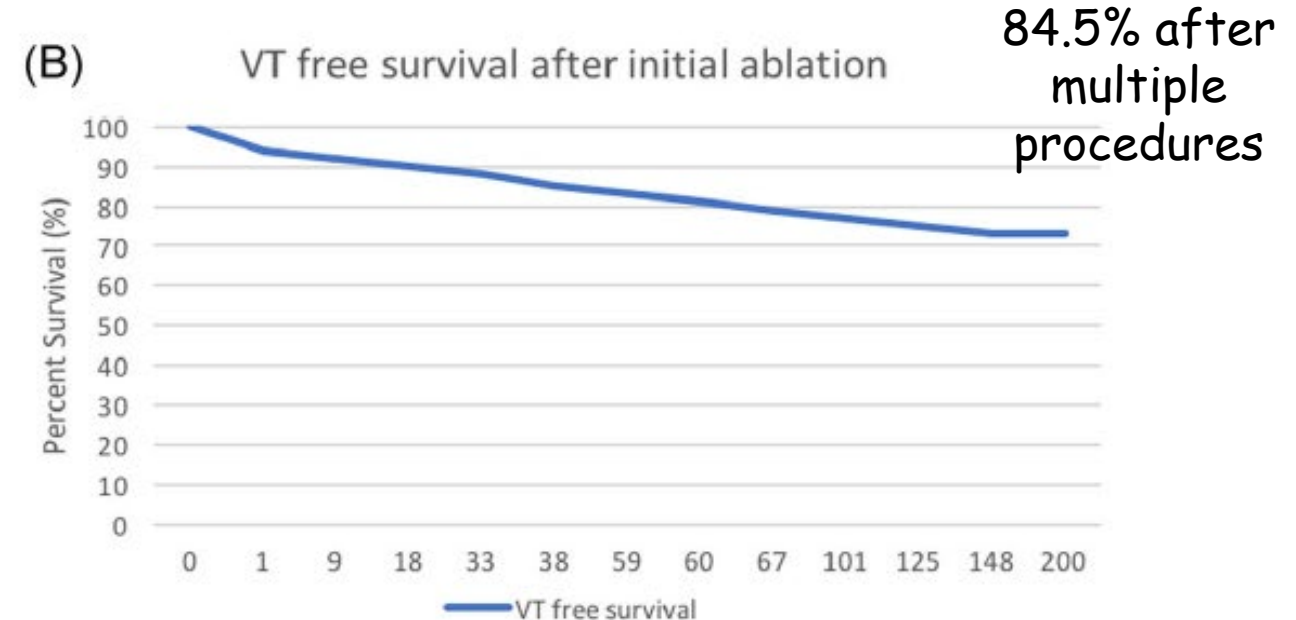
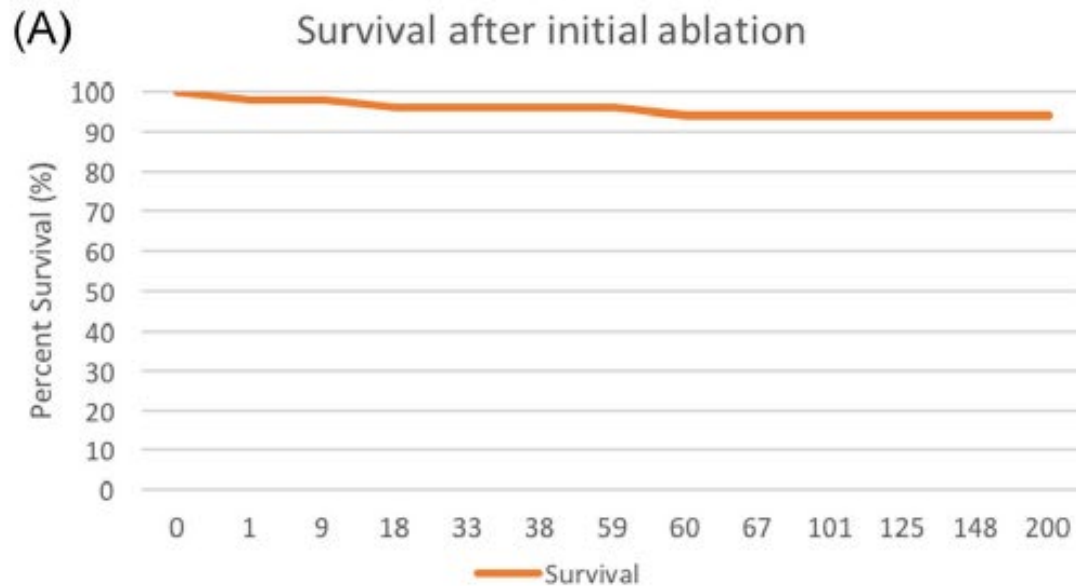


Combes N. et al. Eur Heart J 2019; epub



Discussion of arrhythmia evaluation before surgery in adults ++

Good results in long term follow up in tertiary centers



Yang J et al. *J Cardiovasc Electrophysiol* 2019;30:1560

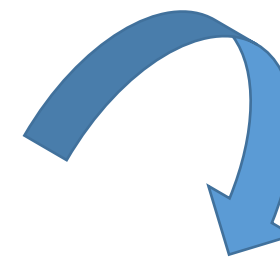
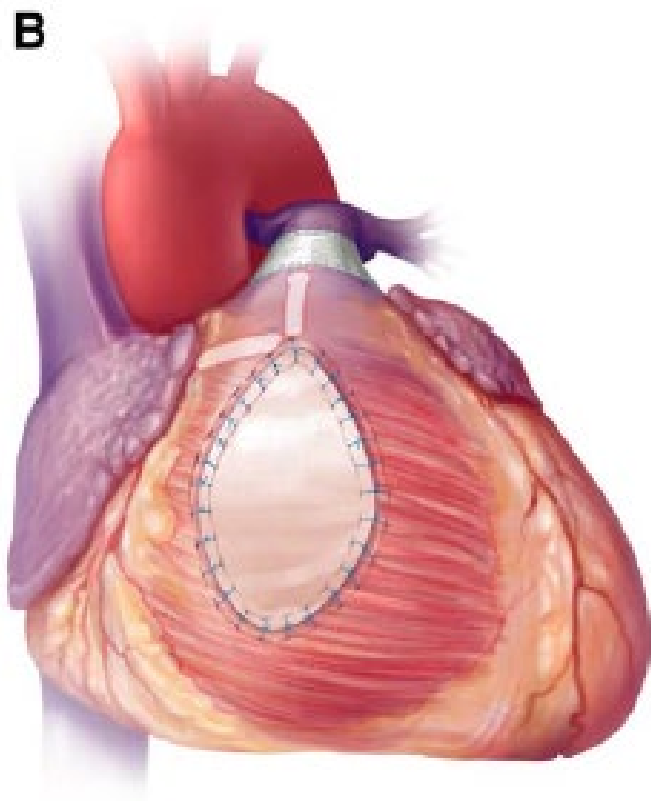
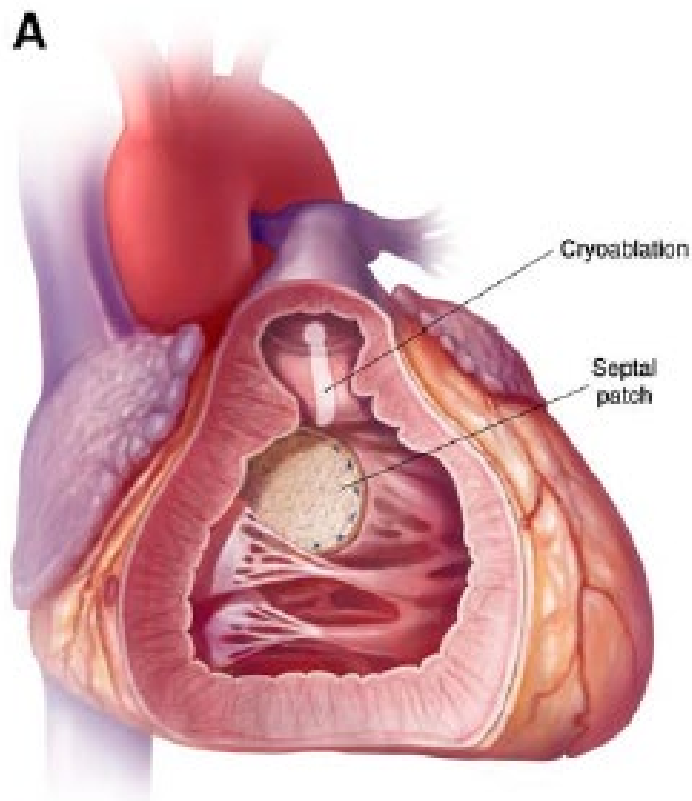
Global acute success rate 90% with 10 to 20% of recurrences at 2 years, complication rate 2%

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

Indications for surgical ablations

Few datas

In case of pulmonary valve replacement +++ but no systematic
(possibly pro arrhythmic if isthmus block is incomplete)
+++ EPS guided in selected patients with selected isthmus



Cryoablation lines +++

Indications for surgical ablations

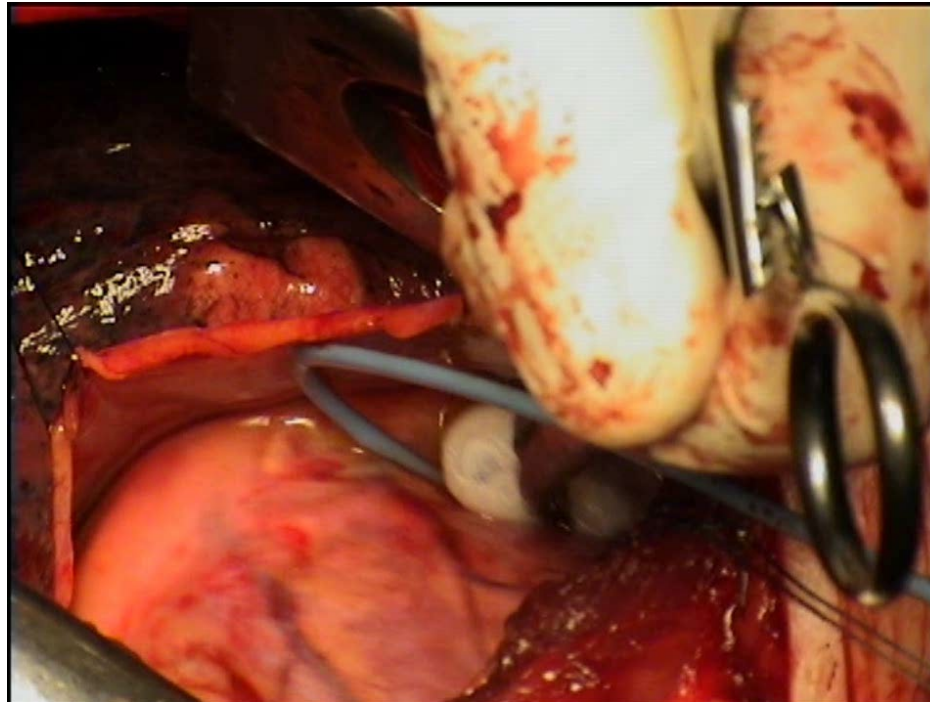
In cases of percutaneous failure and refractory symptomatic VTs



Resections of aneurysms/patches (infundibular ++)

Surgical lesions guided by EPS

Focal/linear epicardial ablation with intraoperative mapping during VT



Indication for VT ablation in CHD

Khairy P et al. HeartRhythm 2014;11(10):e102-65

Classe I

Level C

Catheter ablation is indicated as adjunctive therapy to ICD in adults with CHD, recurrent monomorphic ventricular tachycardia, and multiple appropriated shocks that are not managed by medical therapy or drug therapy

Classe IIa

Level B

Guidelines too restrictives for VT ablation with recent datas. A need for more aggressive treatment in dedicated multidisciplinary teams

In patients with monomorphic reentrant VT with good right and left function and criteria for complete isthmus ablation, first line therapy without ICD ?

and monomorphic alternative to drug

Classe IIa

Level C

is reasonable in adults with CHD and frequent associated with deteriorating ventricular function

Classe III

Level C

Catheter ablation alone is not considered appropriate prophylactic therapy in adults with CHD deemed to be at increase risk for sudden cardiac death

Conclusion

Most ventricular arrhythmias in CHD patients are reentrant.

Ablation in CHD is feasible with a good success rate and a low complication rate.

Ablation planning is crucial for good result with evaluation of haemodynamic status, review of surgical datas, check for substrate access, check for adapted tools (sheaths, guidewires, large ablation catheter curve, 3D mapping...).

Multidisciplinary team involved in CHD is mandatory for optimal management.