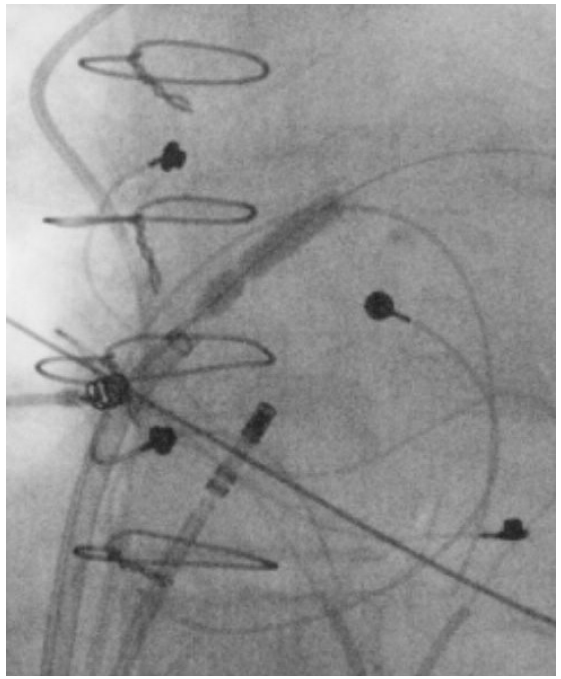
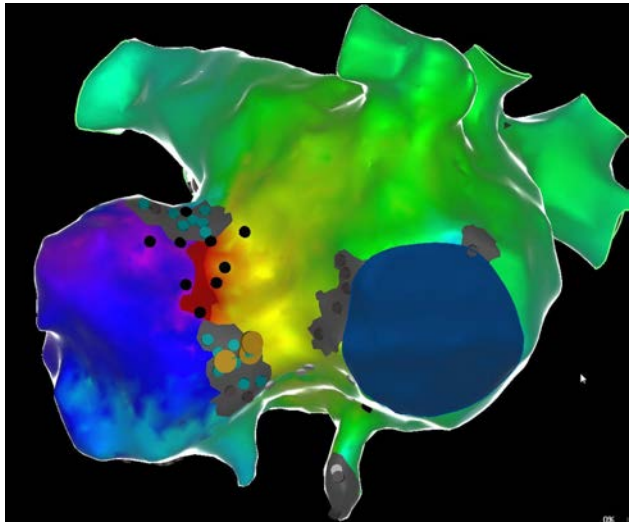
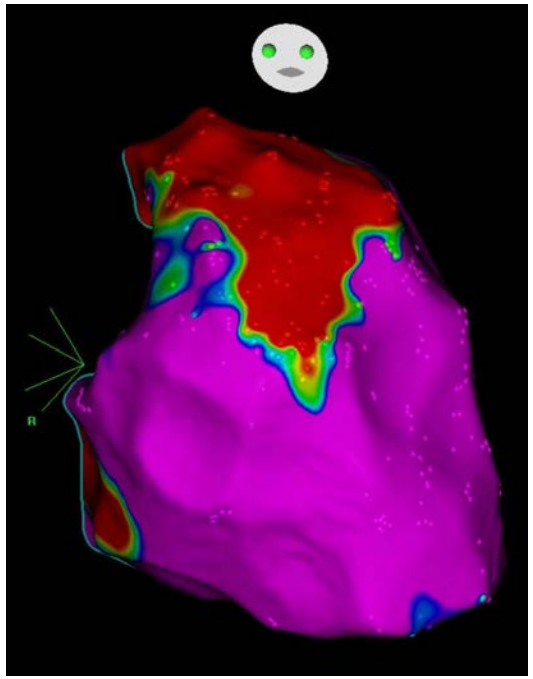
Cardiac Arrhythmias in Congenital Heart Diseases

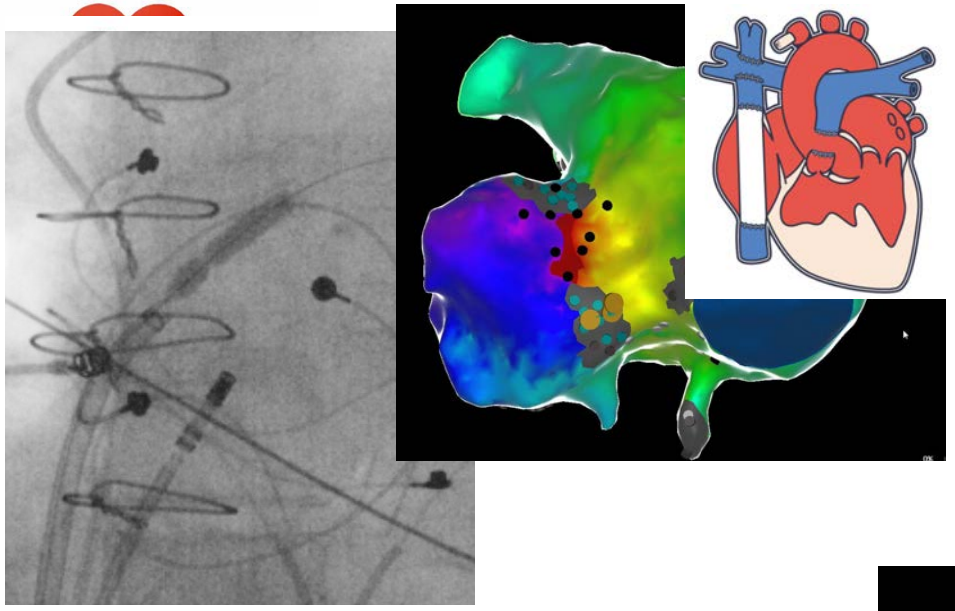


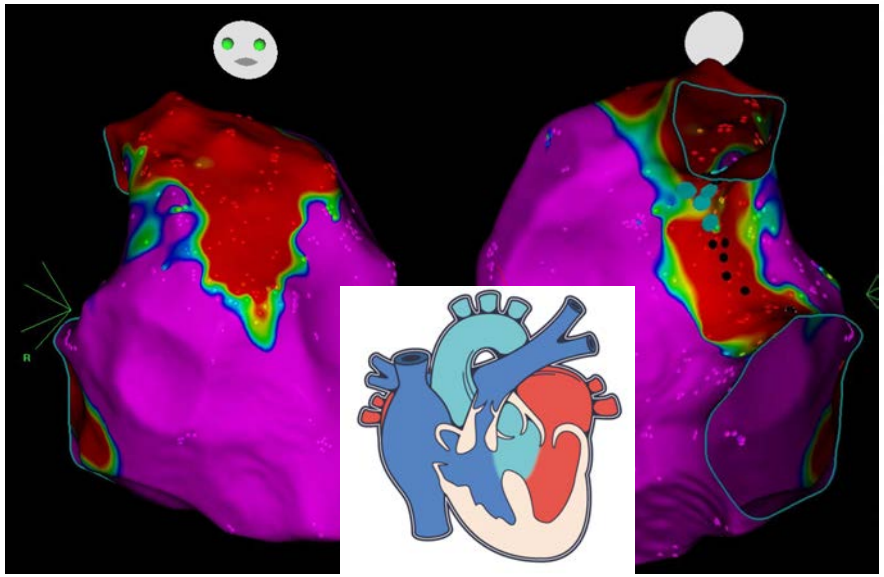


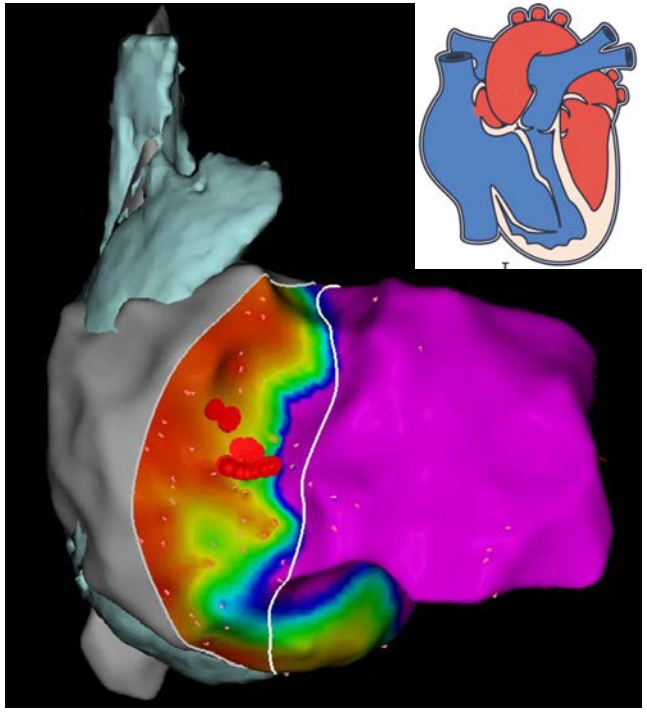


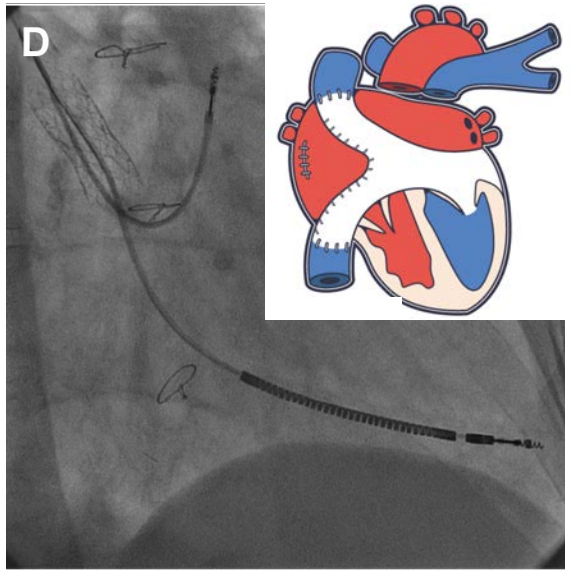
Victor WALDMANN

Hôpital Européen Georges Pompidou — Hôpital Necker





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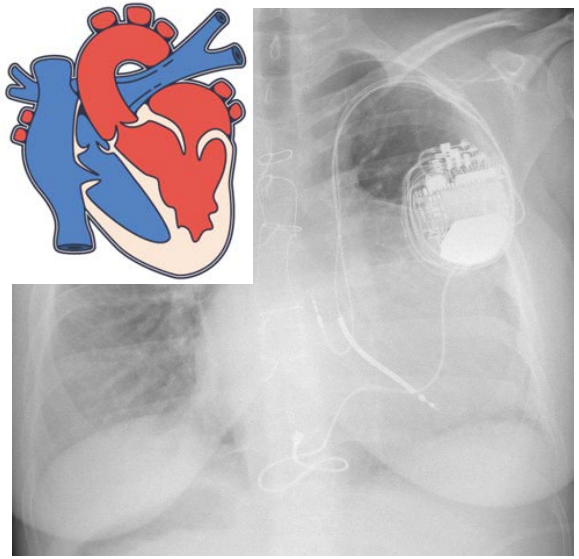
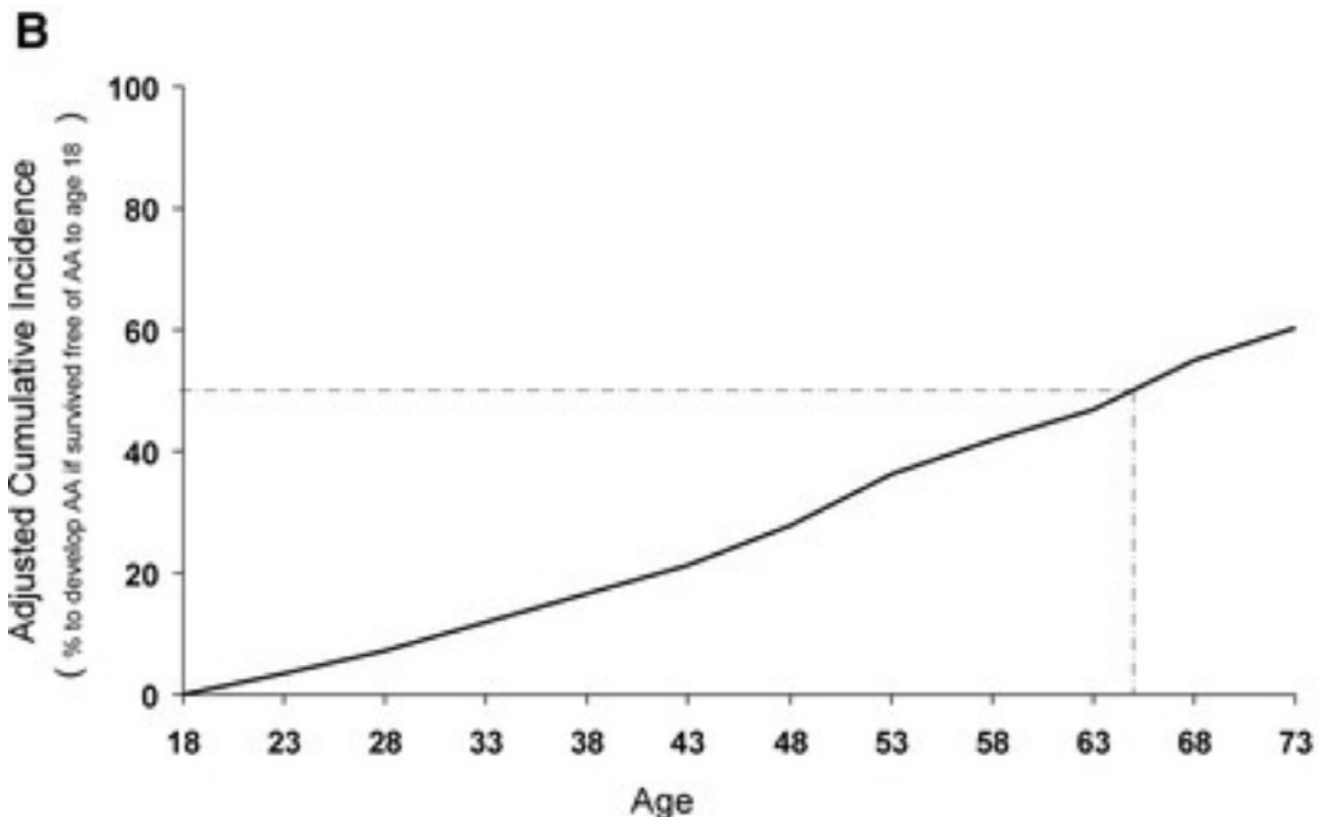


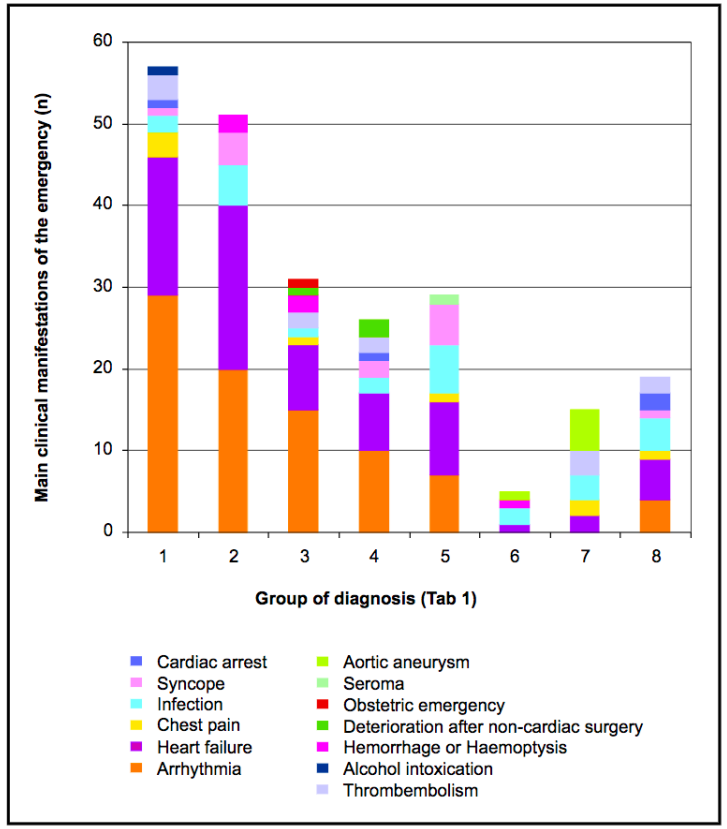
Atrial Arrhythmias

Prevalence 15%
Lifetime Incidence of 50%



Bouchardy J, et al. Circulation 2009

1st Cause of Urgent Admission



Kaemmerer H, et al. Am J Cardiol 2008

Sudden Death

2nd Cause of Death

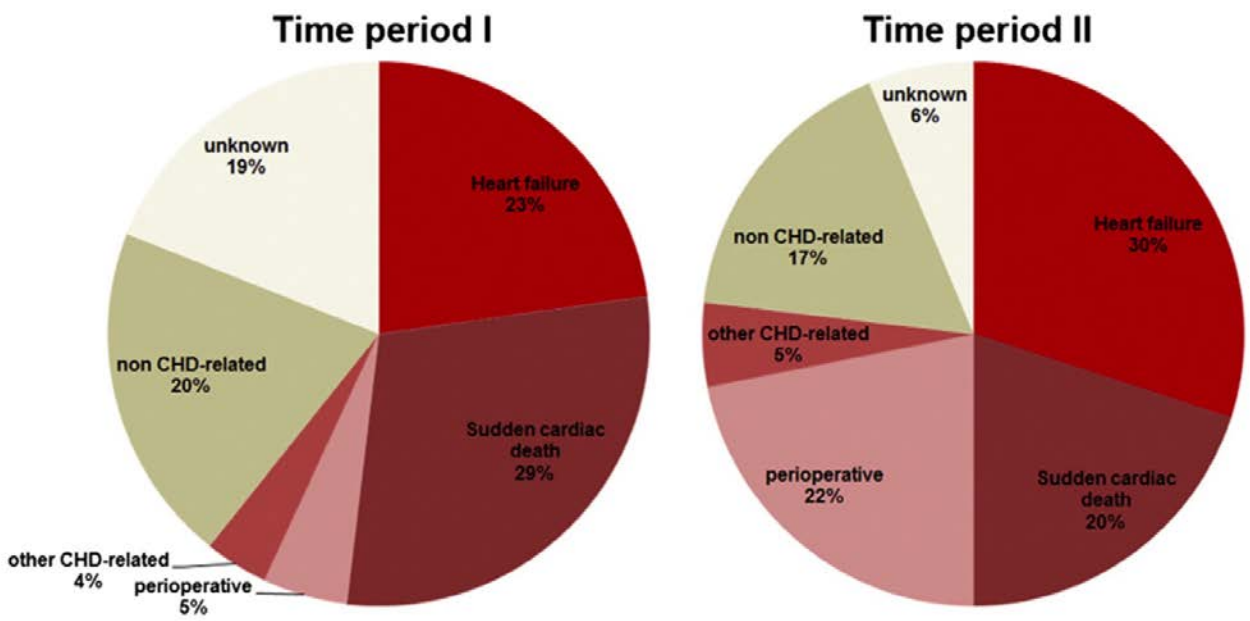
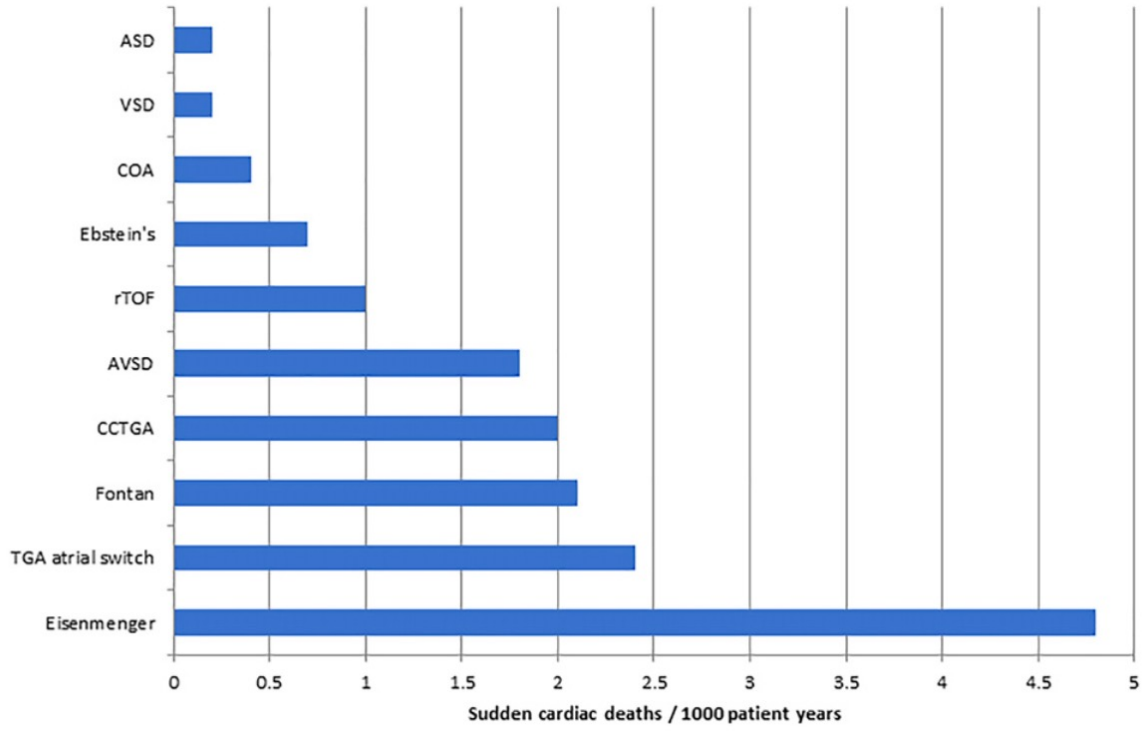


Fig. 2. Causes of death compared between time period I (January 2001–December 2008) vs. II (January 2009–January 2015).

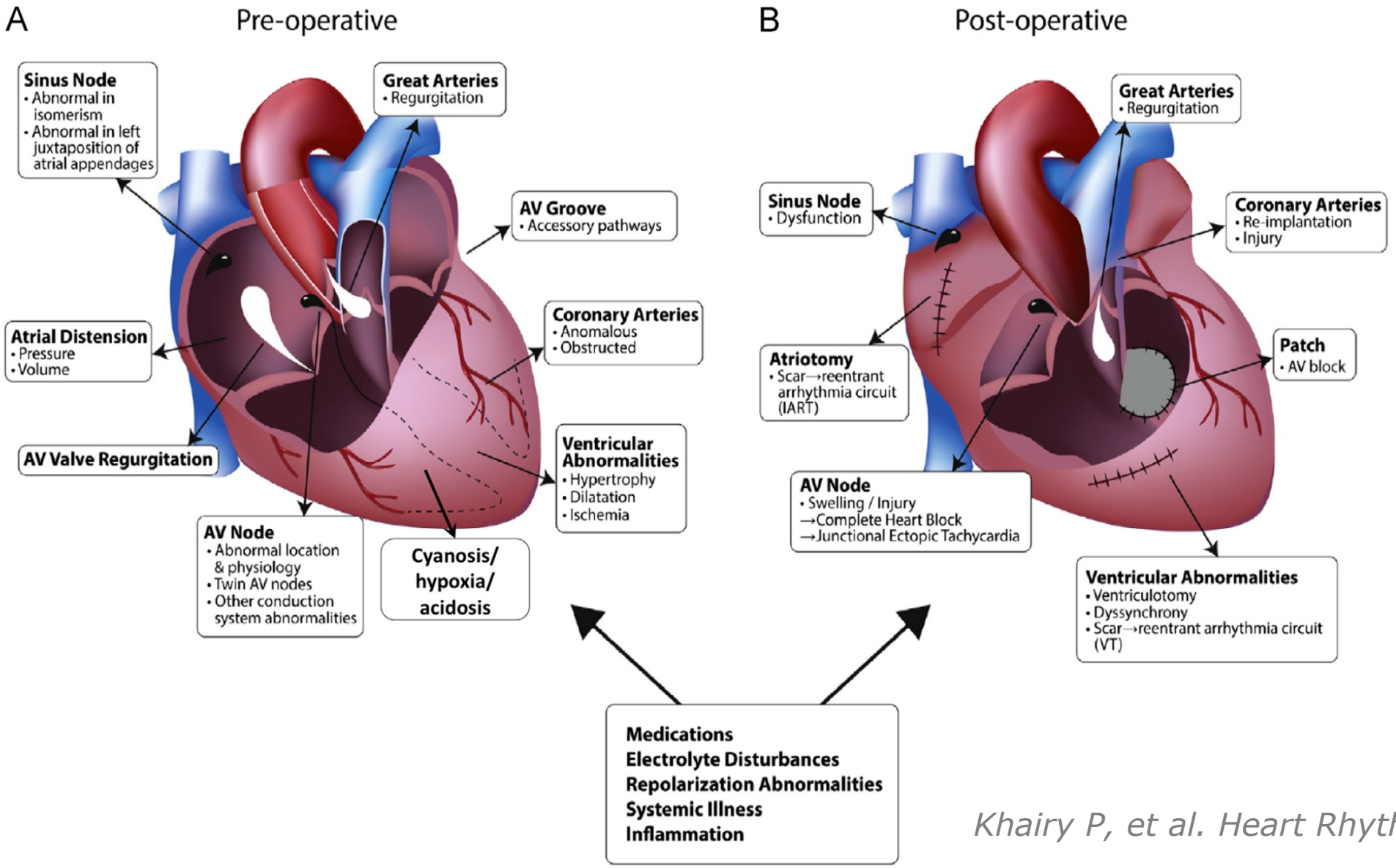
Engelings CC, et al. Int J Cardiol 2016

Incidence According to CHD



Moore B, et al. Am J Cardiol 2018

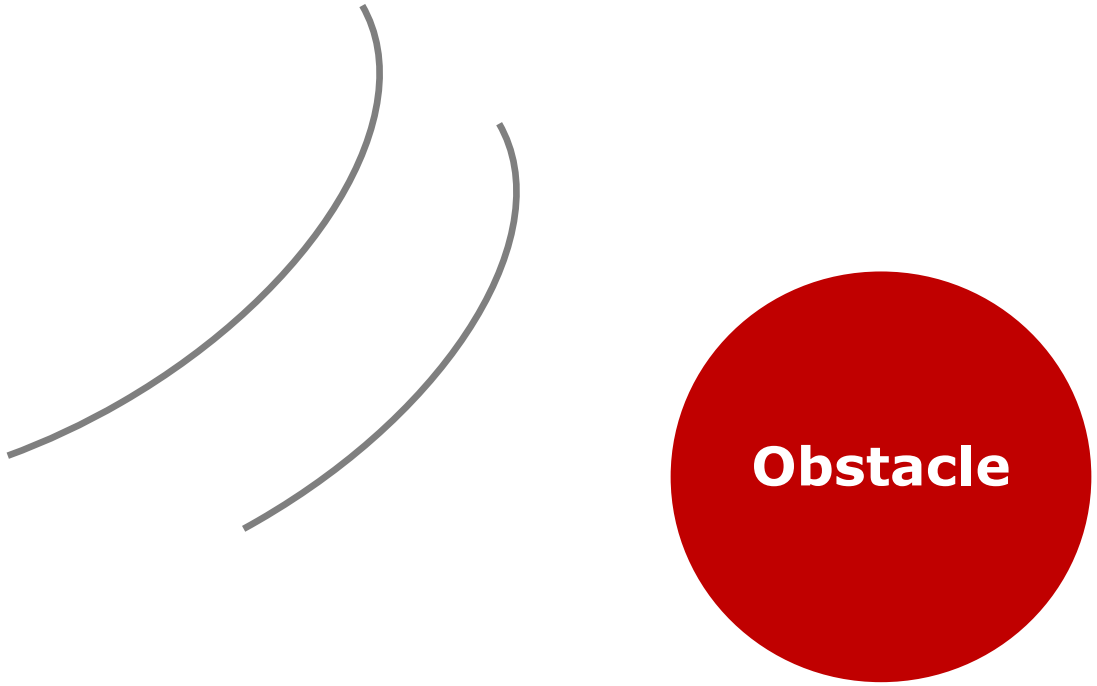
CHD — A Unique Substrate



Khairy P, et al. Heart Rhythm 2014

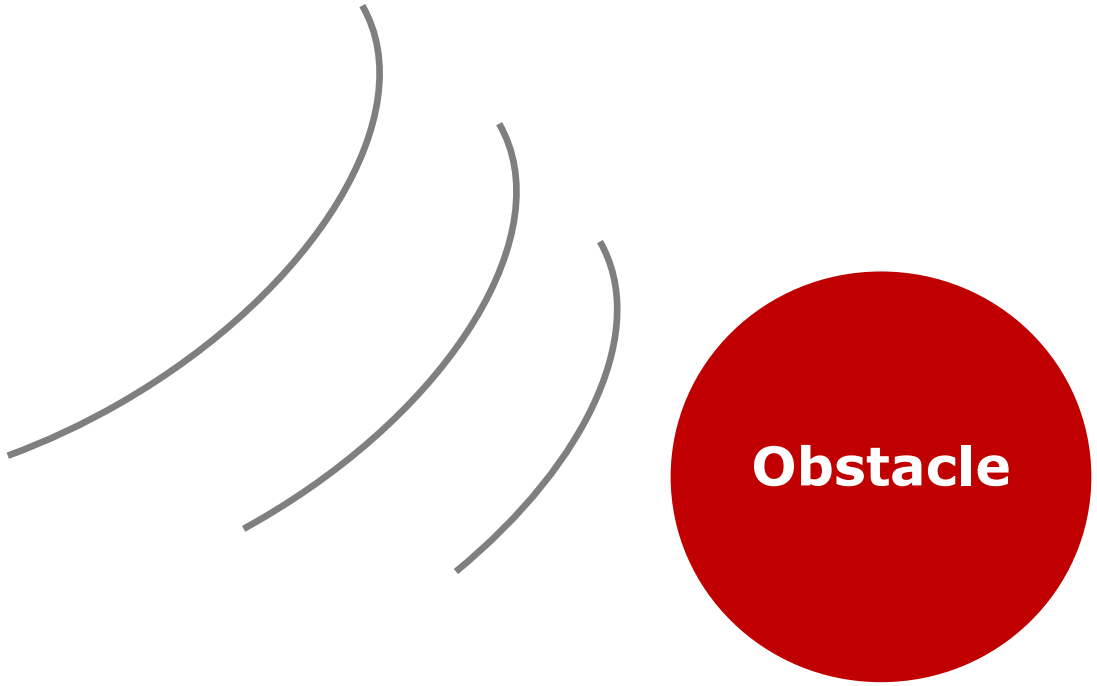
Tachyarrhythmia in CHD

Pathophysiology



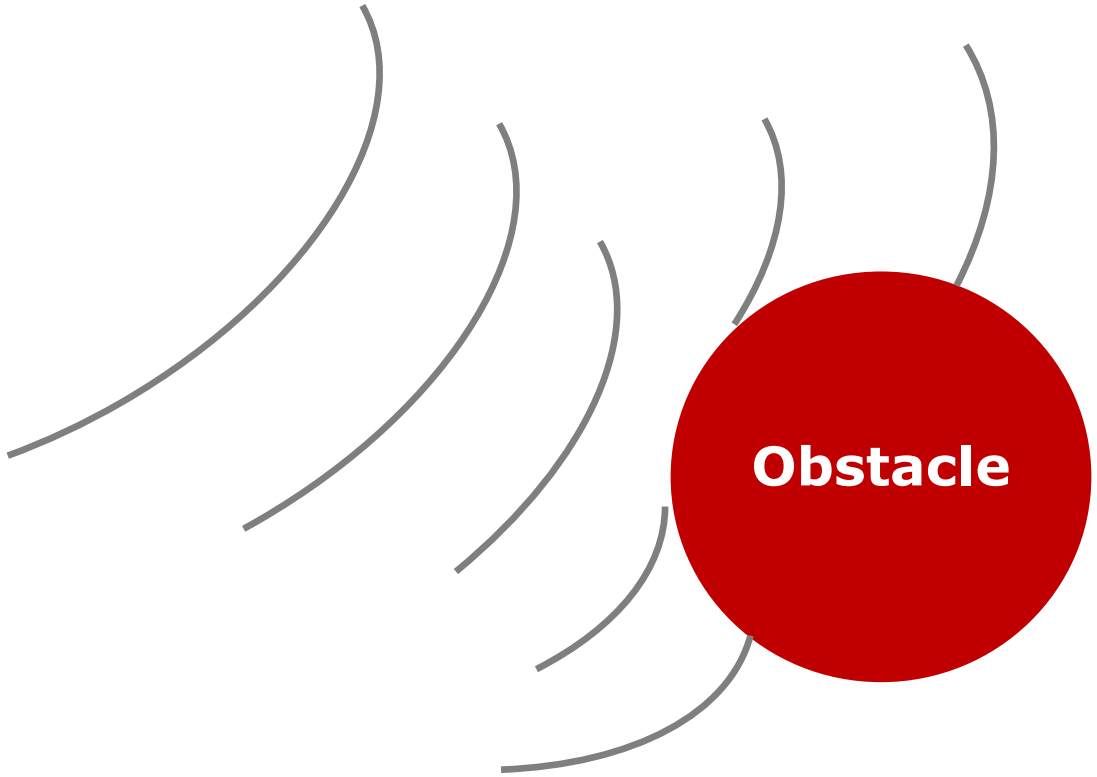
Tachyarrhythmia in CHD

Pathophysiology



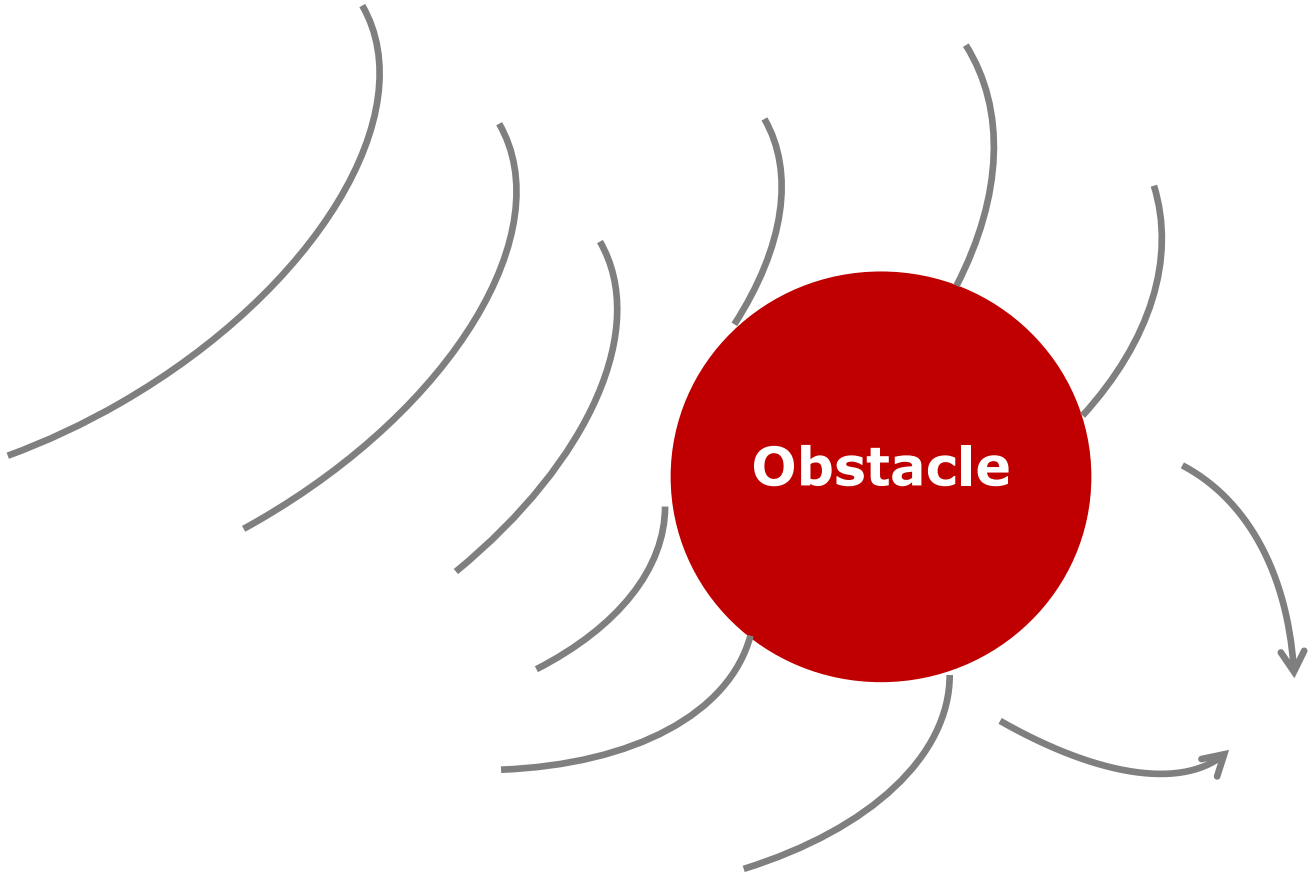
Tachyarrhythmia in CHD

Pathophysiology



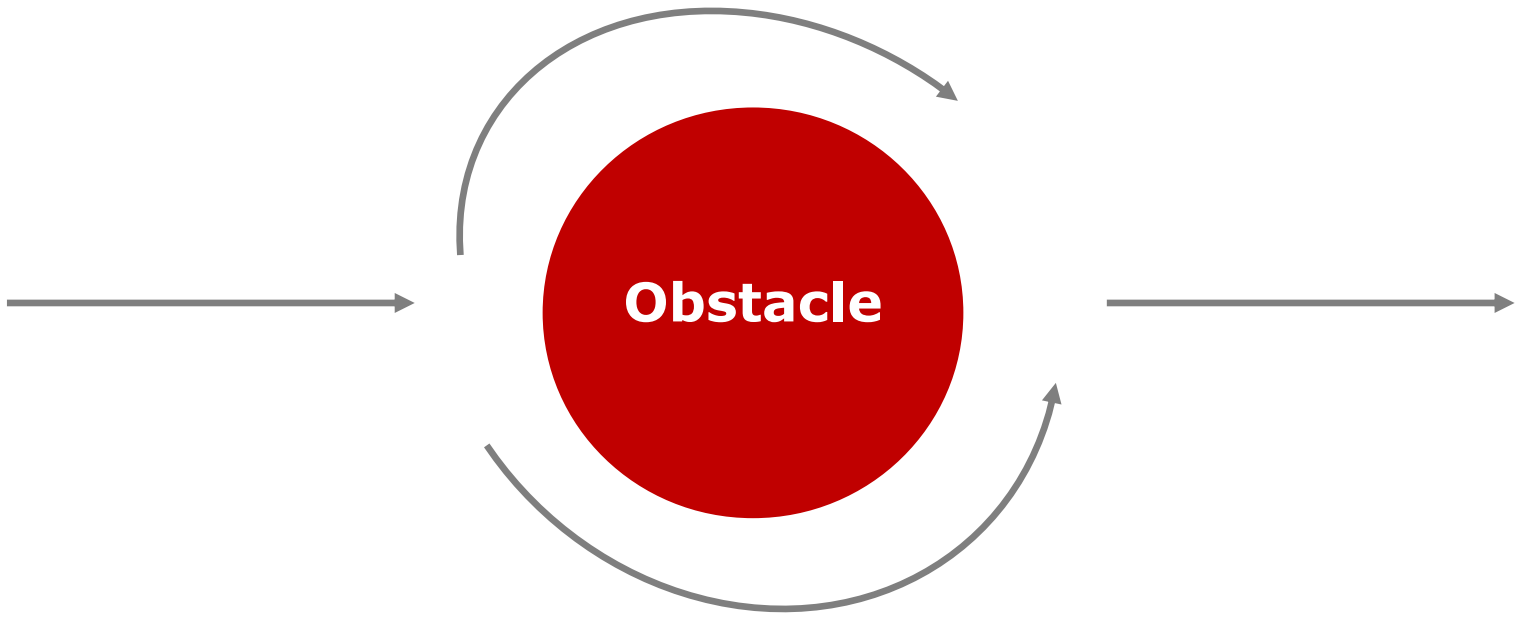
Tachyarrhythmia in CHD

Pathophysiology



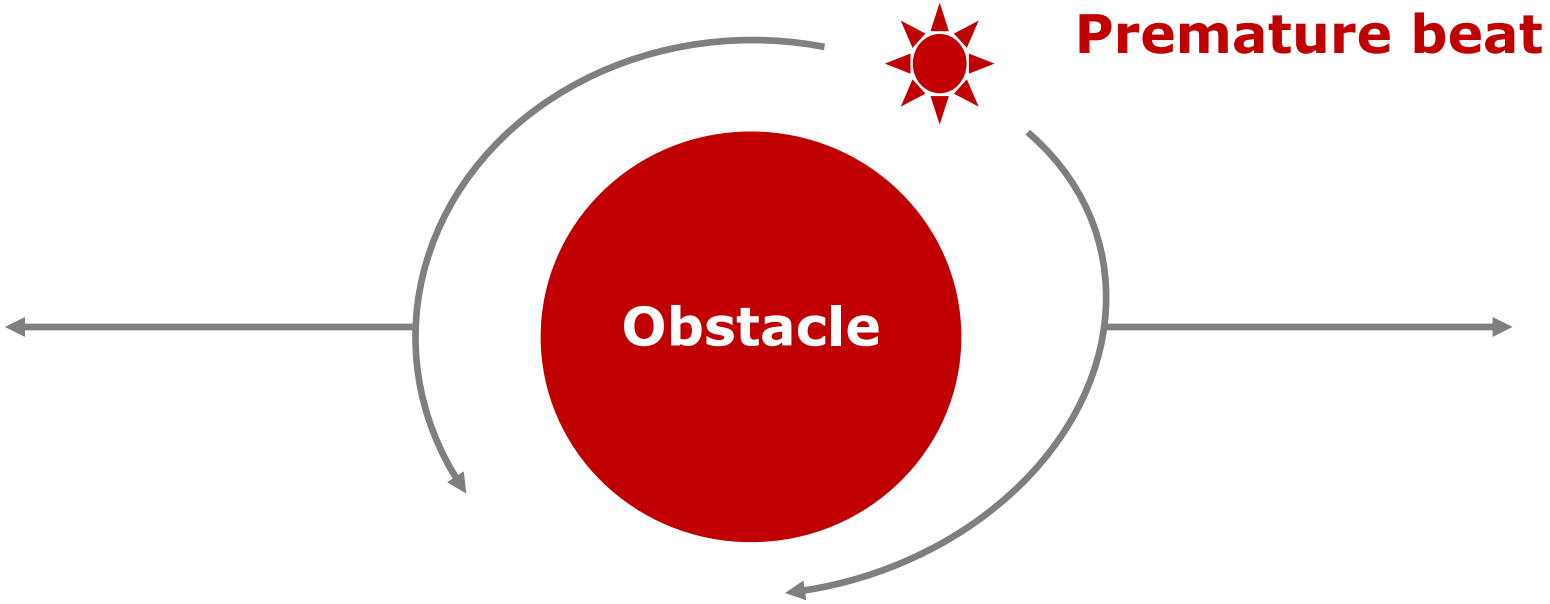
Tachyarrhythmia in CHD

Pathophysiology



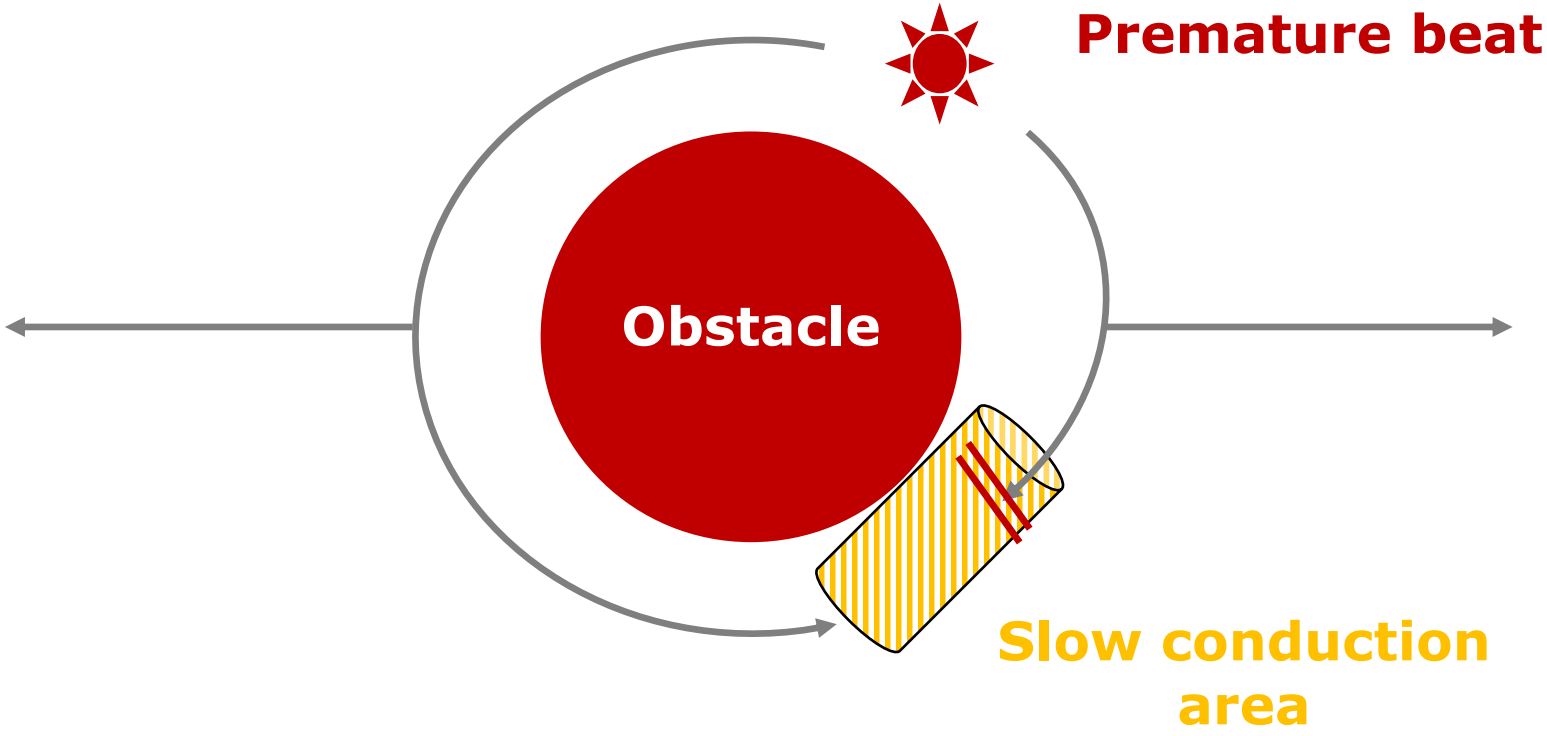
Tachyarrhythmia in CHD

Pathophysiology



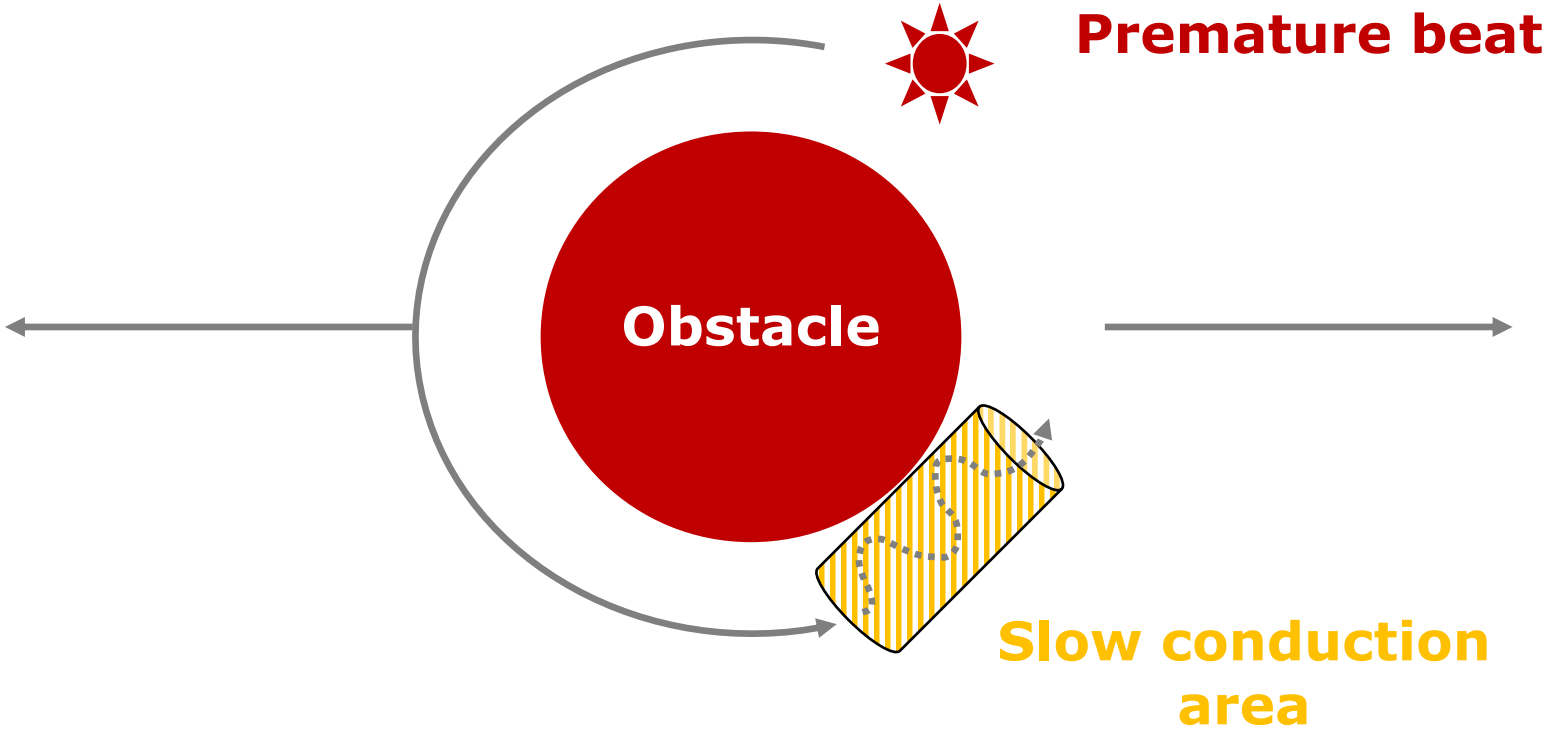
Tachyarrhythmia in CHD

Pathophysiology



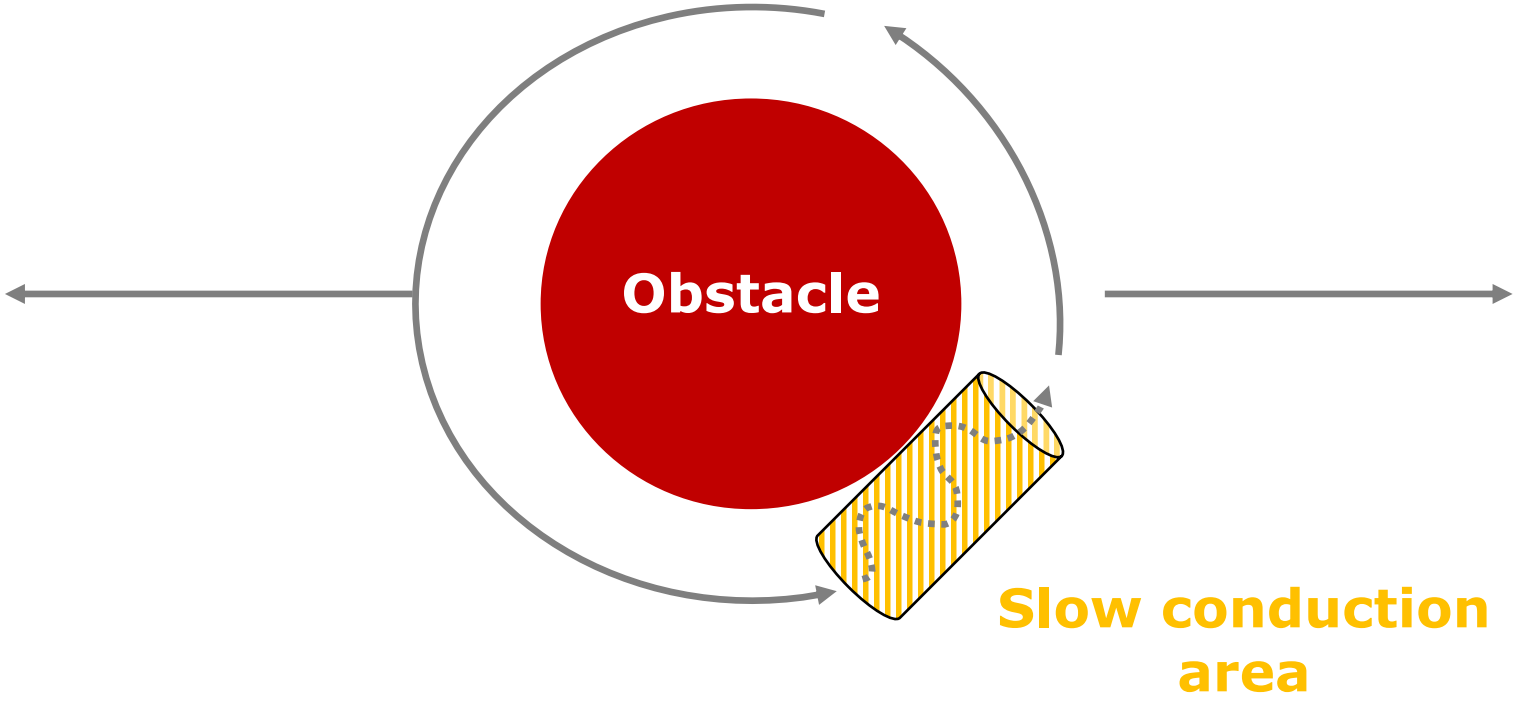
Tachyarrhythmia in CHD

Pathophysiology

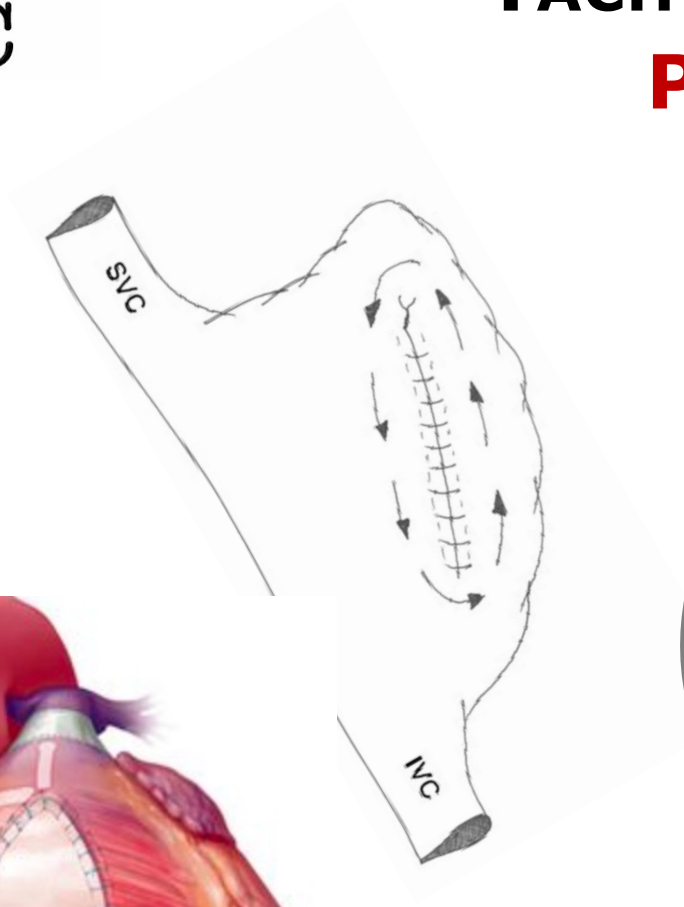


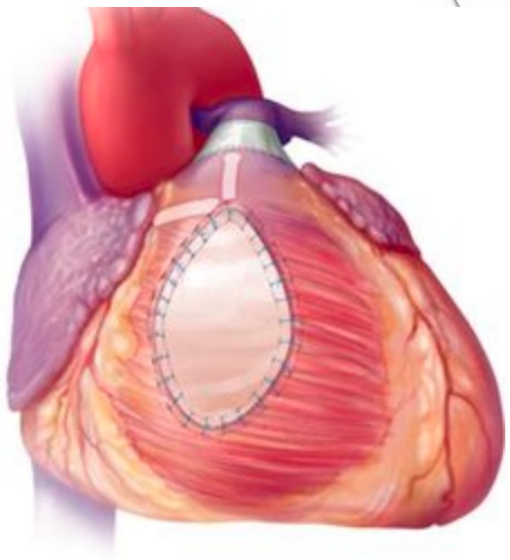
Tachyarrhythmia in CHD

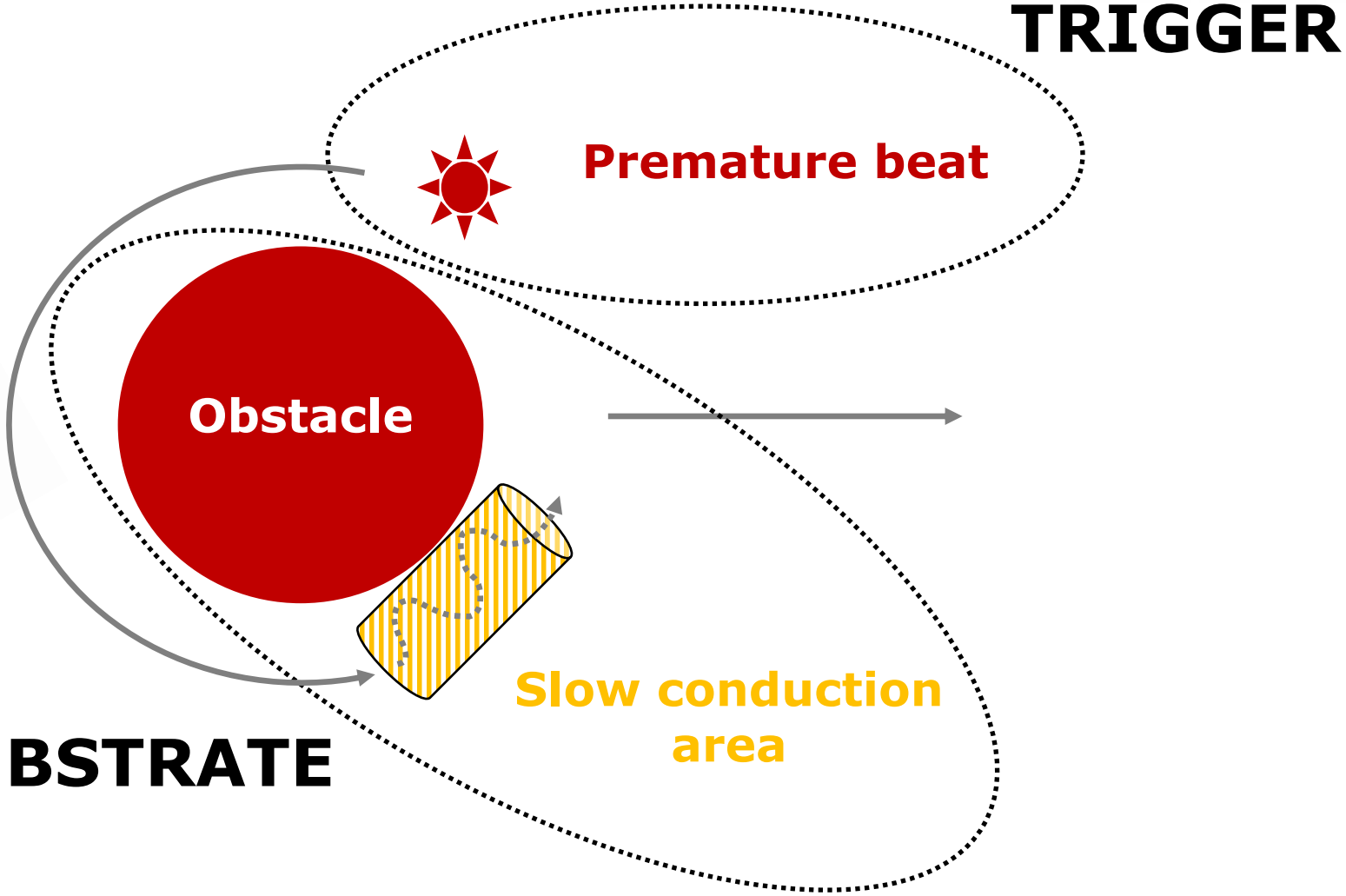
Pathophysiology



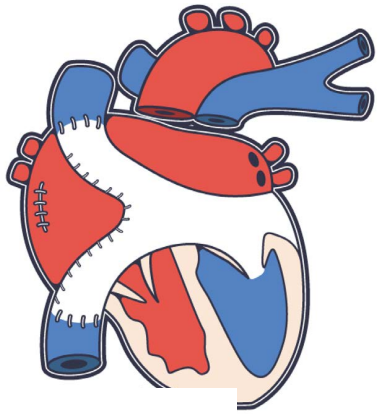
Tachyarrhythmia in CHD
Pathophysiology



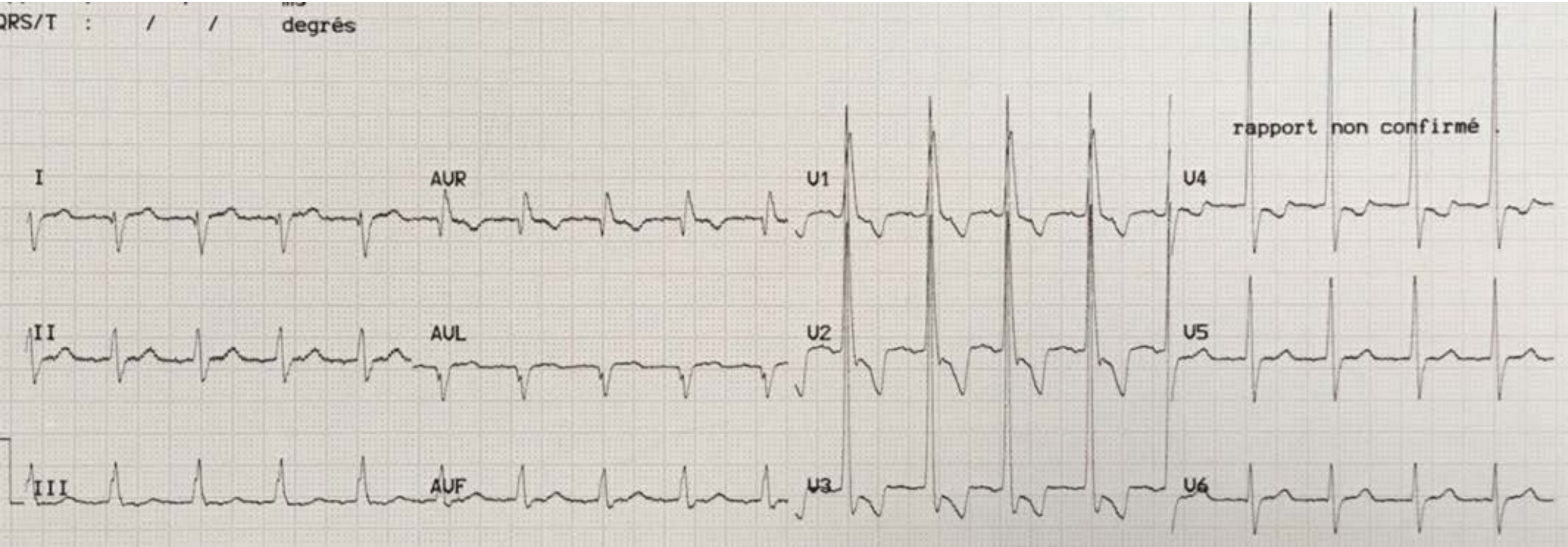




Case n°1

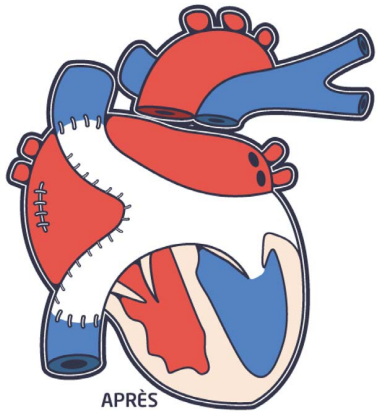


Man, 48 y
Mustard surgery at 2 y
Dyspnea NYHA III for 1 week

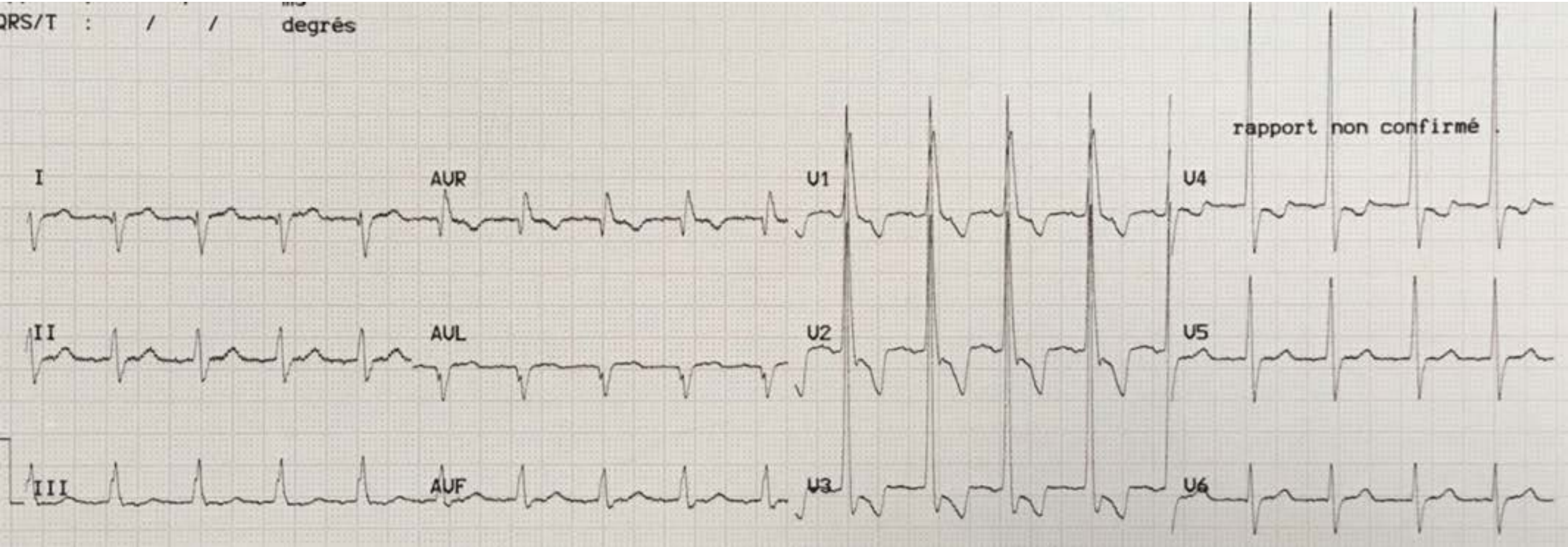


CASE N°1

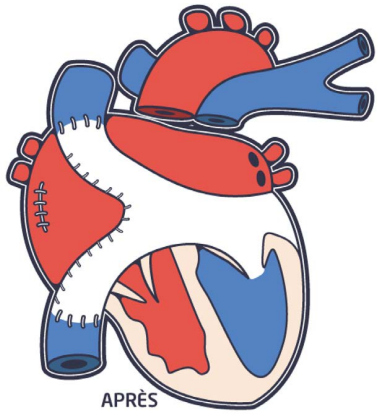
Man, 48 y
Mustard surgery at 2 y
Dyspnea NYHA III for 1 week



→ **Diagnosis ?**
→ **Management ?**



Case n°1



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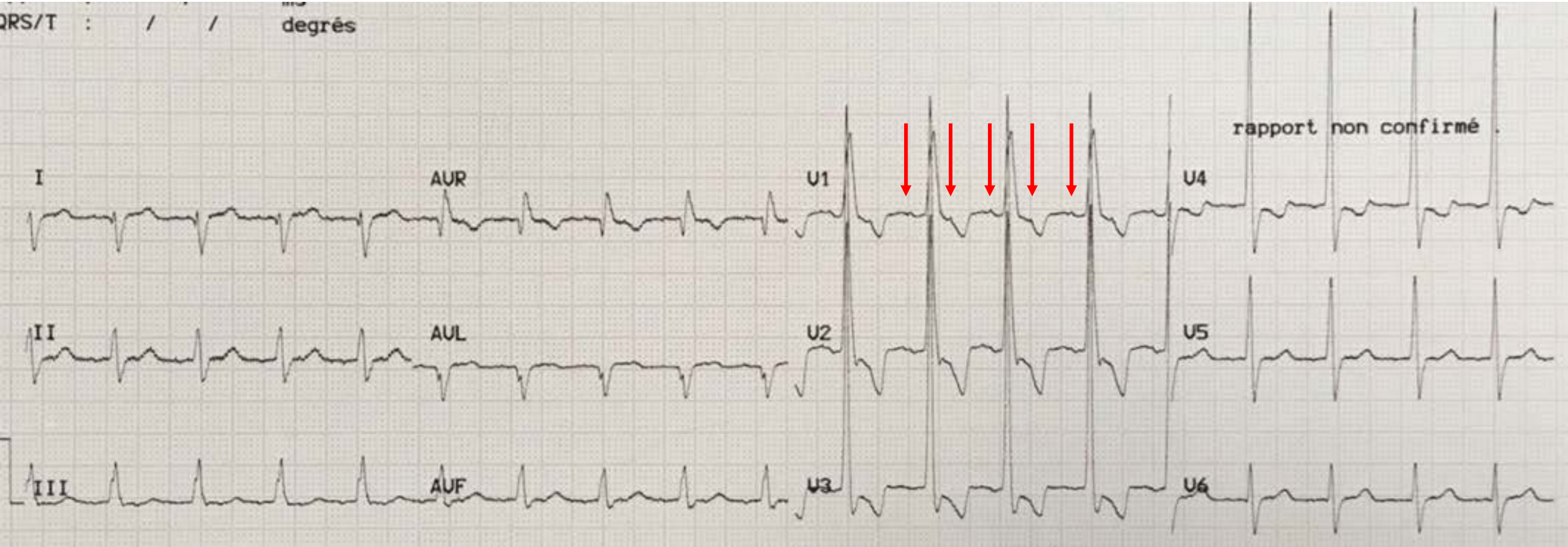
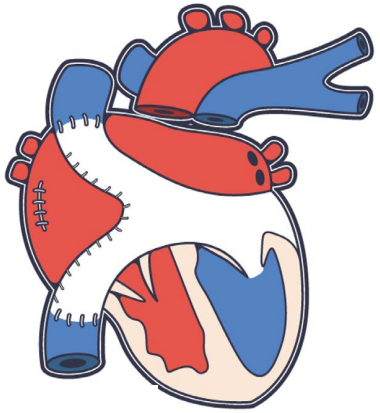


Case n°1



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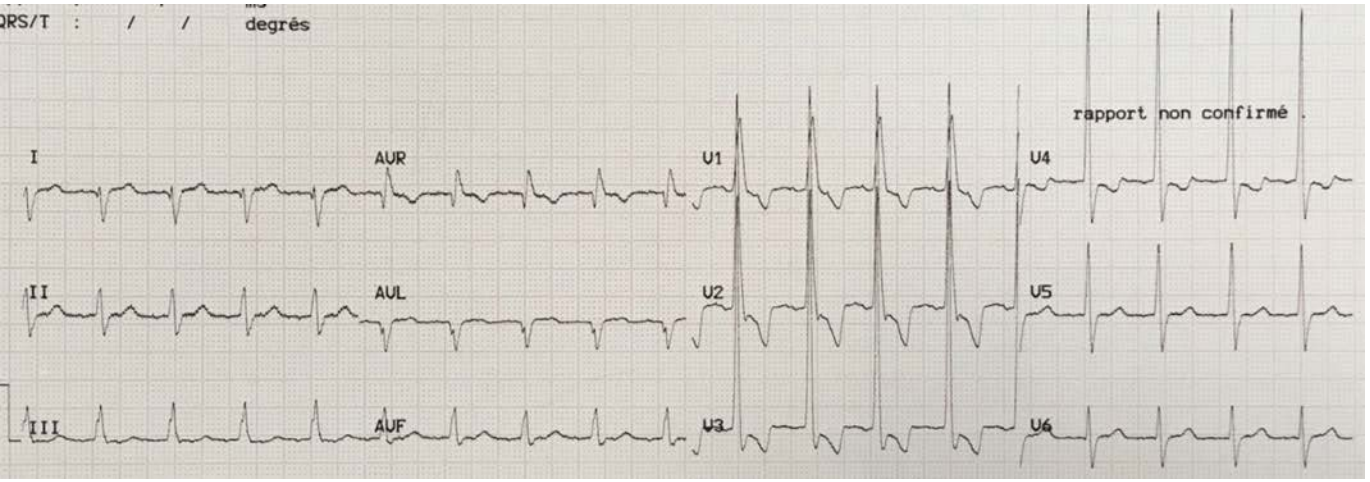


Ventricular function ?
Baffle stenosis/leak ?
Atrial dilatation ?

Management

1/ Anticoagulation
2/ B-blockers
3/ Cardioversion vs. ablation ++

Anticoagulation in Atrial Arrhythmias

Indications ?

- Simple defect = CHA2DS2-VASc
- Moderate or complex = anticoagulation !

VKA vs. NOAC ?

Anticoagulation in Atrial Arrhythmias

Indications ?

- Simple defect = CHA2DS2-VASc
- Moderate or complex = anticoagulation !

VKA vs. NOAC ?

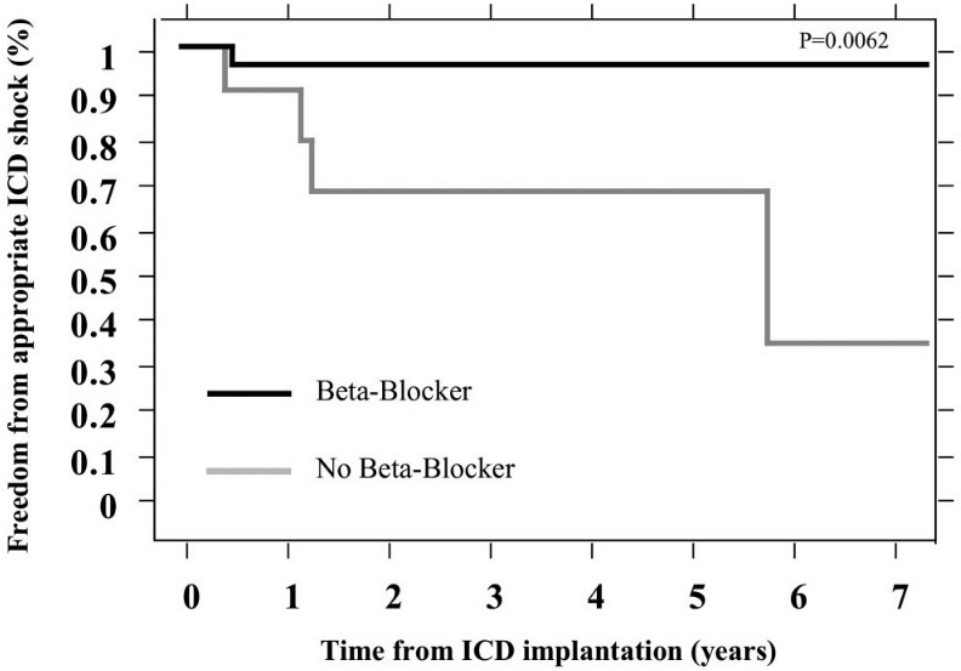
- Expert consensus (2014) discouraged use of NOAC in complex CHD
- Emergence of encouraging data (NOTE registry)
- NOAC use promoted by last ACC/AHA guidelines (2018)

Sudden Death and Defibrillators in Transposition of the Great Arteries With Intra-atrial Baffles

A Multicenter Study

Of the 18 adjudicated appropriate shocks, supraventricular tachyarrhythmias were recognized as preceding or coexisting with ventricular tachyarrhythmias in 9 patients (50.0%),

SVT may trigger sudden death in D-TGV patients with atrial switch



Khairy P, et al. Circ AE 2008

SVT may trigger sudden death in D-TGV patients with atrial switch

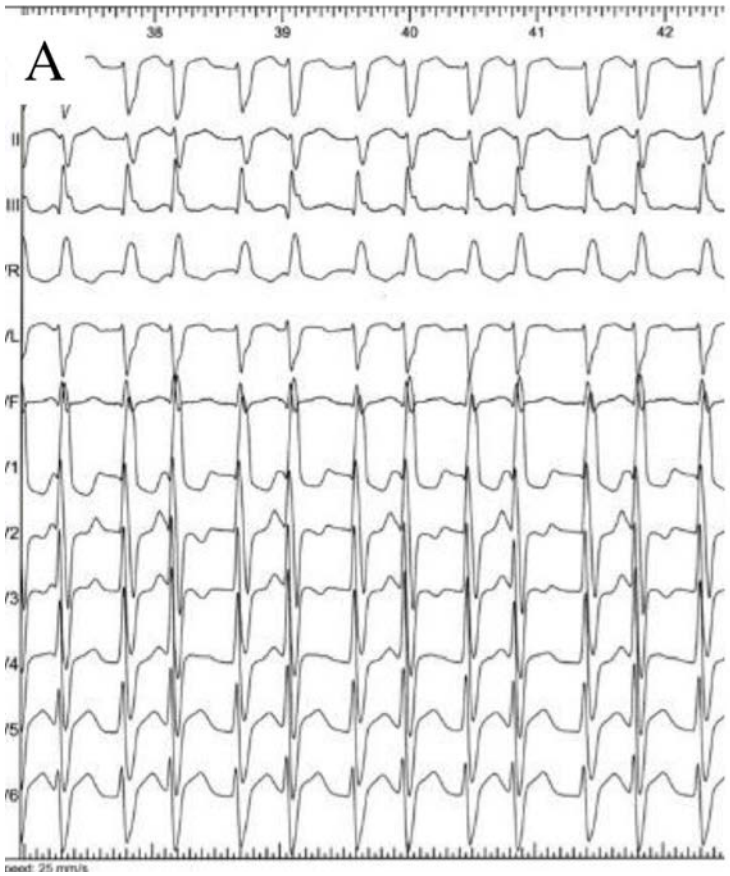
- Right systemic ventricle supplied by right coronary artery
- Ischemia
- Rapid conduction to ventricles (young age, risk of 1/1 conduction)
- Reduction in stroke volume due to poor atrial transport

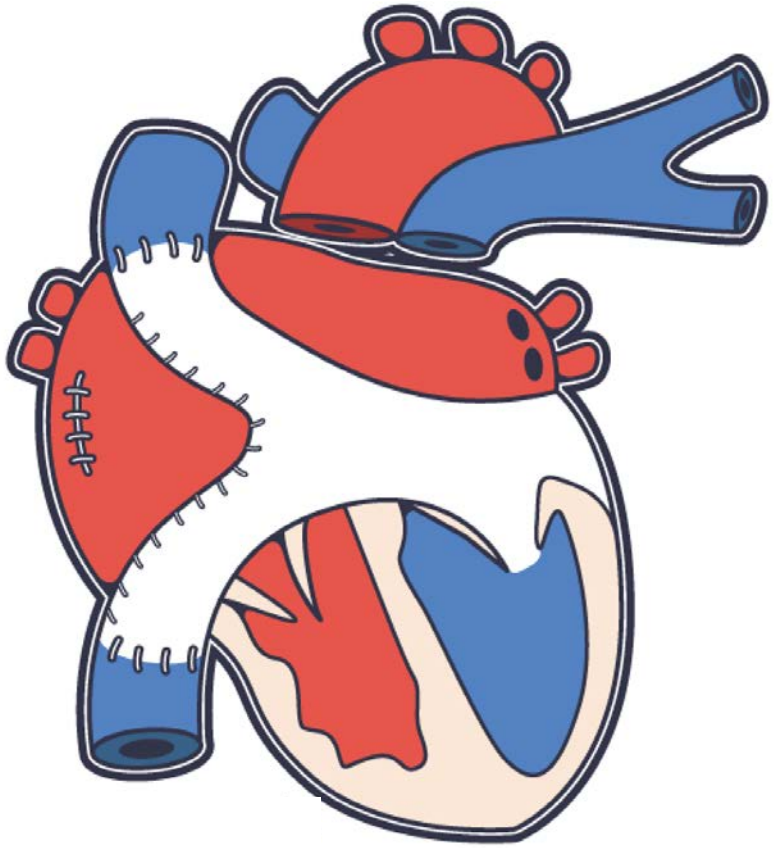
→ **Manage aggressively SVT in Senning/Mustard patients**
Efficacy of ablation >> pharmacological approach

Khairy P, et al. Circ AE 2008

Tachyarrhythmia in CHD

Atrial Arrhythmia <-> SCD

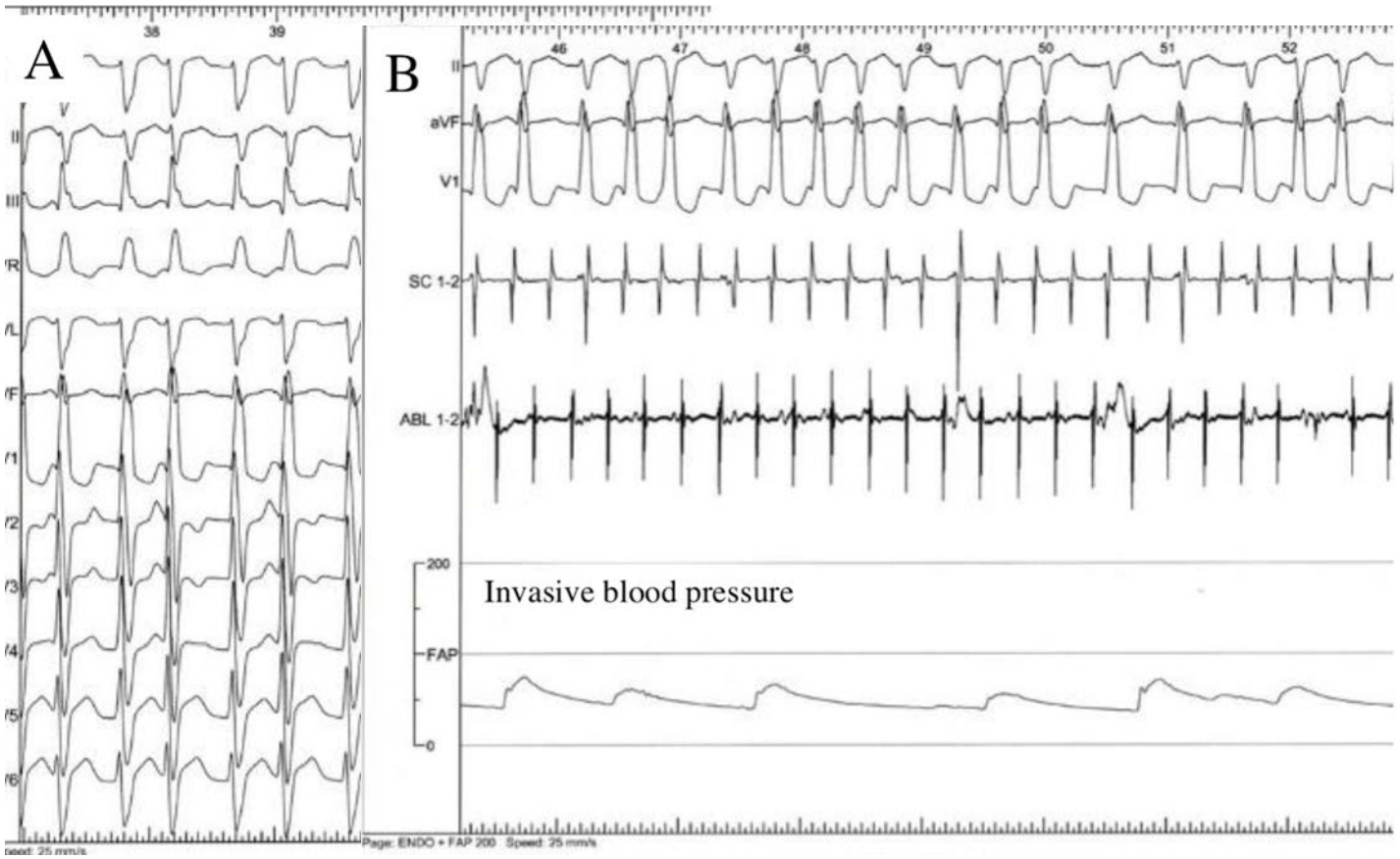




Combes N, et al. Europace 2019

Tachyarrhythmia in CHD

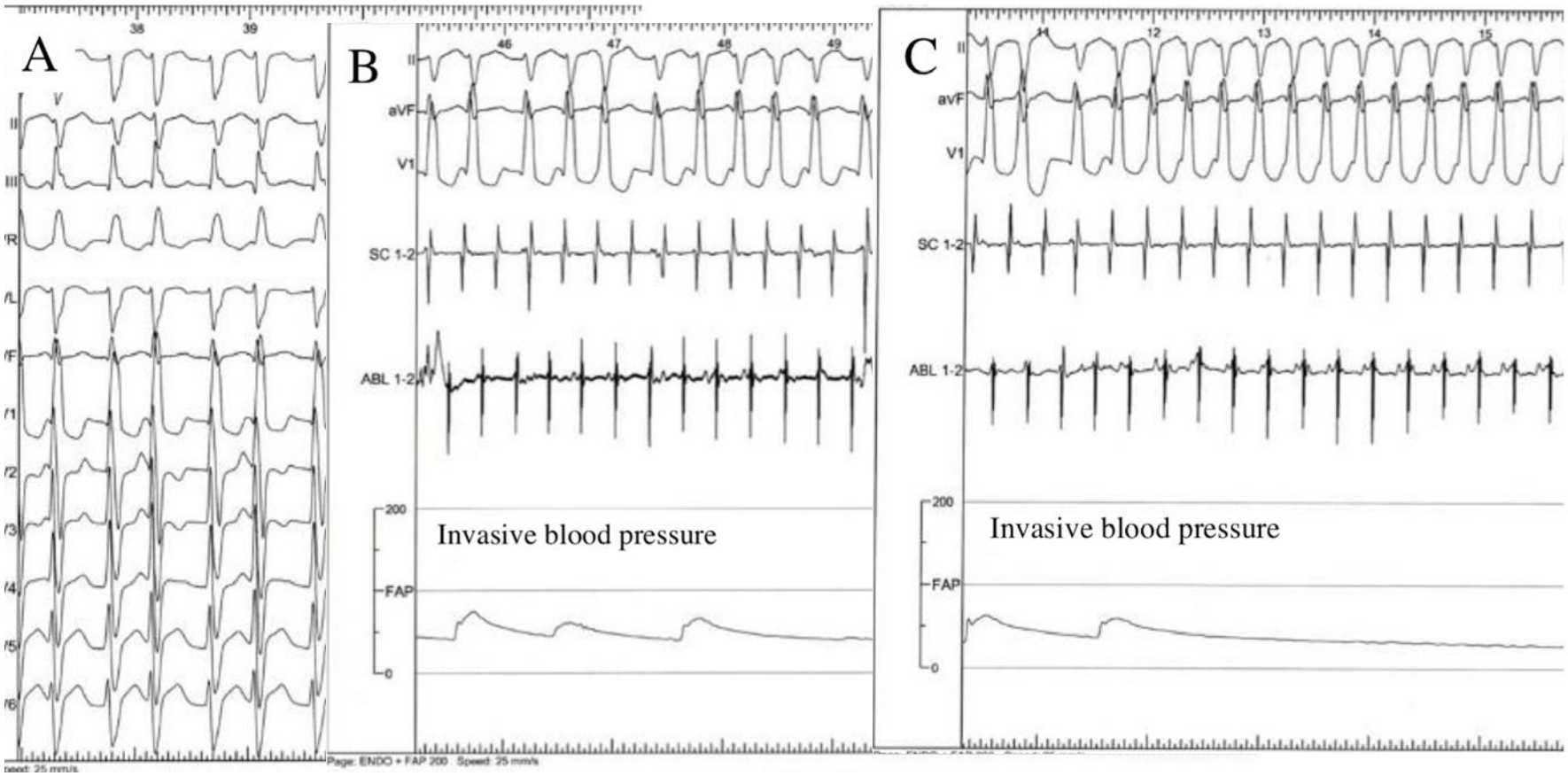
Atrial Arrhythmia <-> SCD



Combes N, et al. Europace 2019

Tachyarrhythmia in CHD

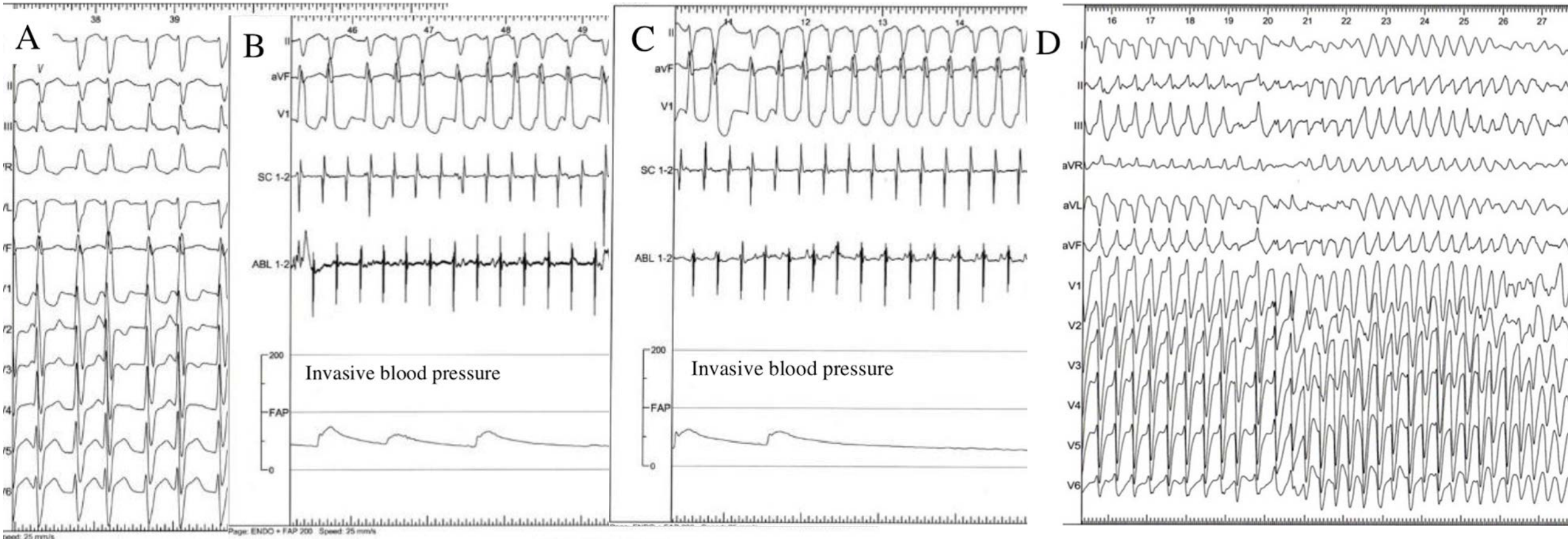
Atrial Arrhythmia <-> SCD



Combes N, et al. Europace 2019

Tachyarrhythmia in CHD

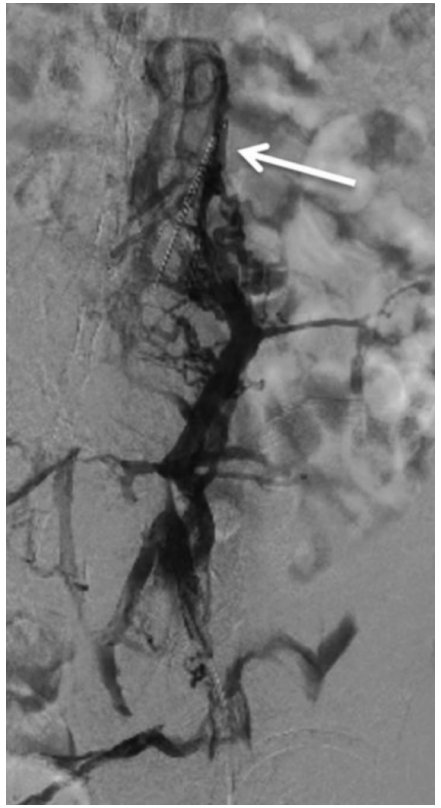
Atrial Arrhythmia <-> SCD

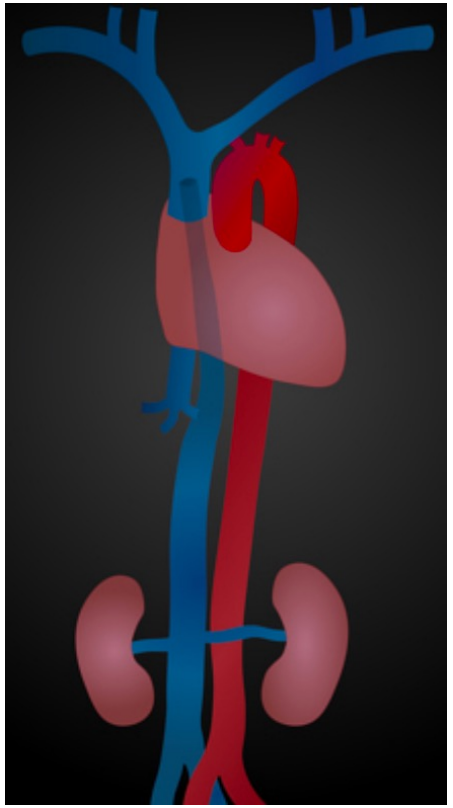


Combes N, et al. Europace 2019

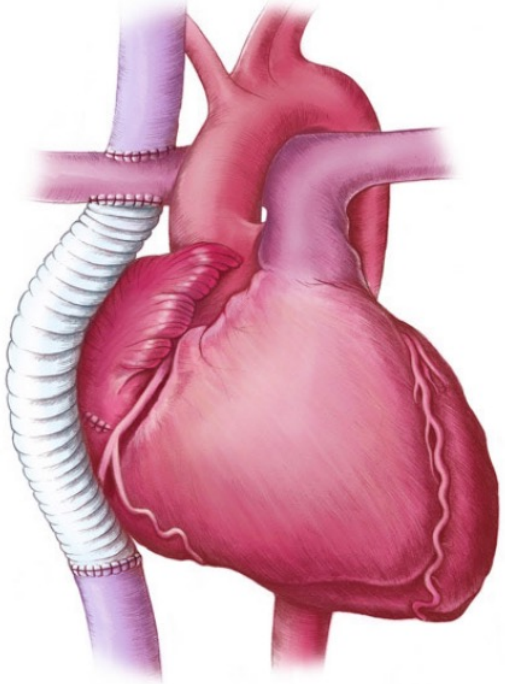
Catheter Ablation
Access Issues

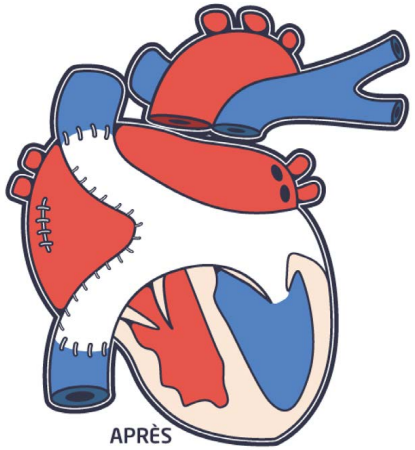
Vascular occlusion
Azygos continuation

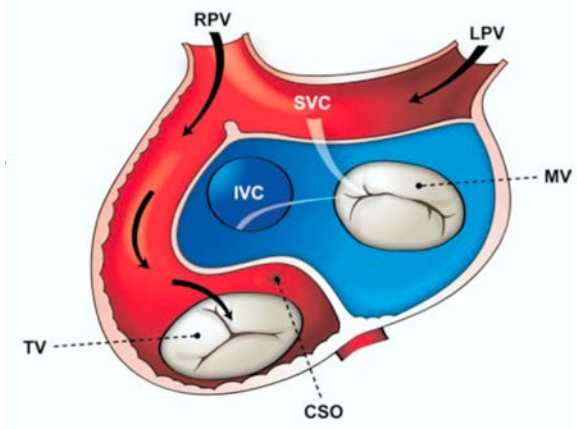


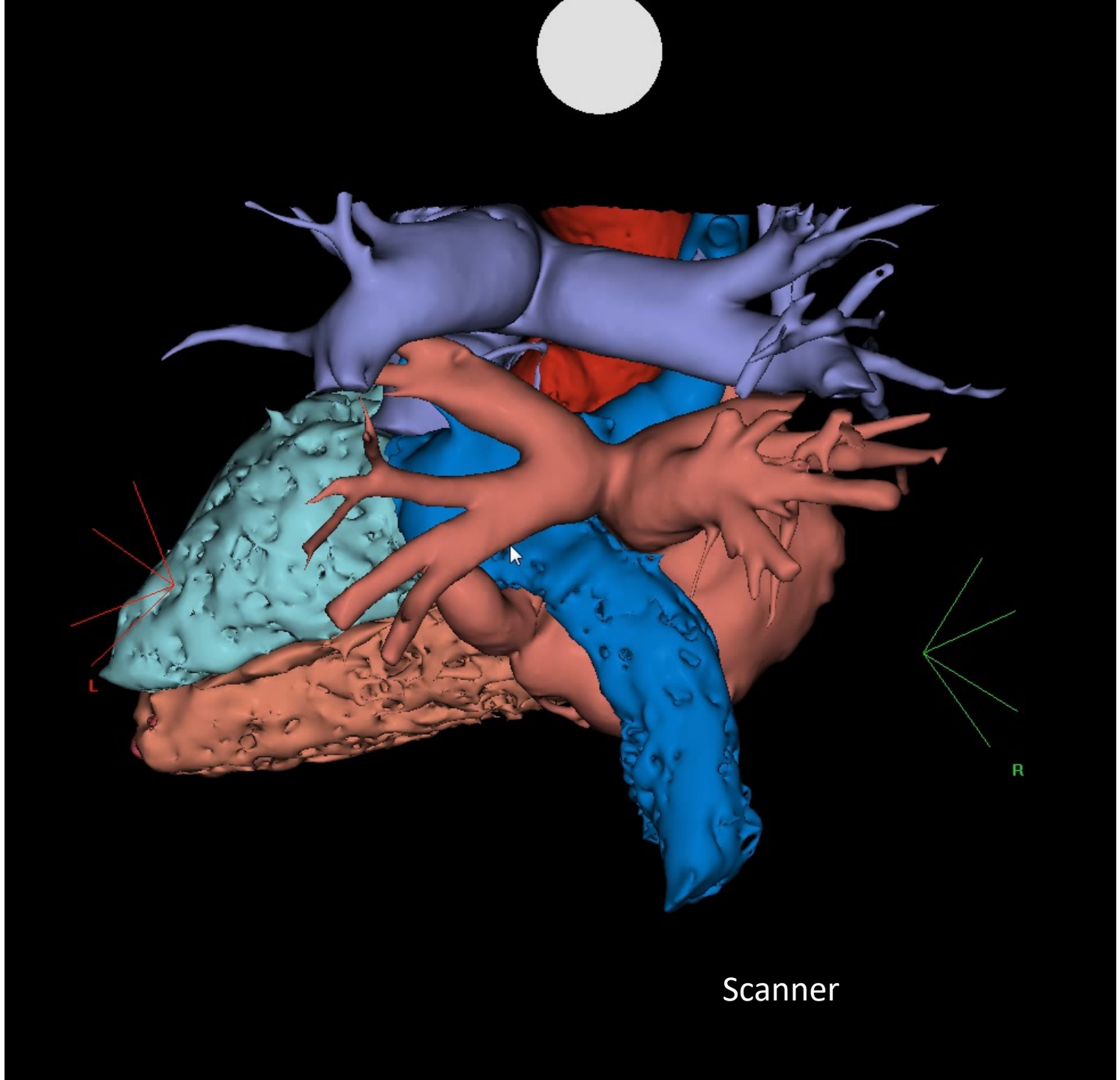


Complex anatomies

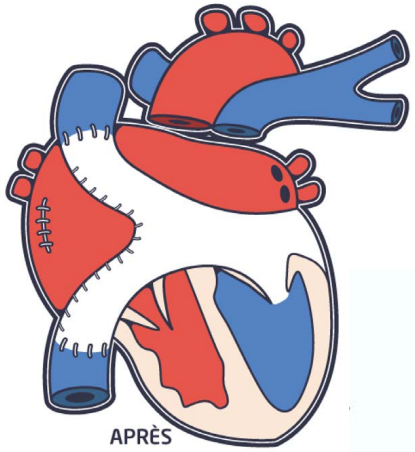


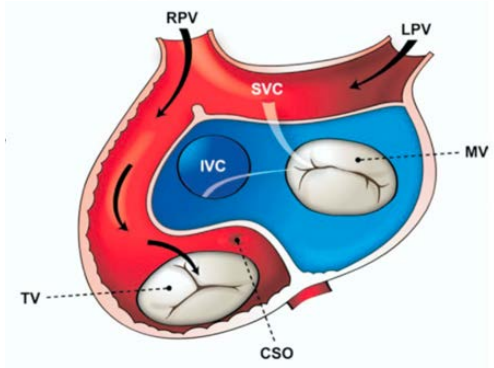


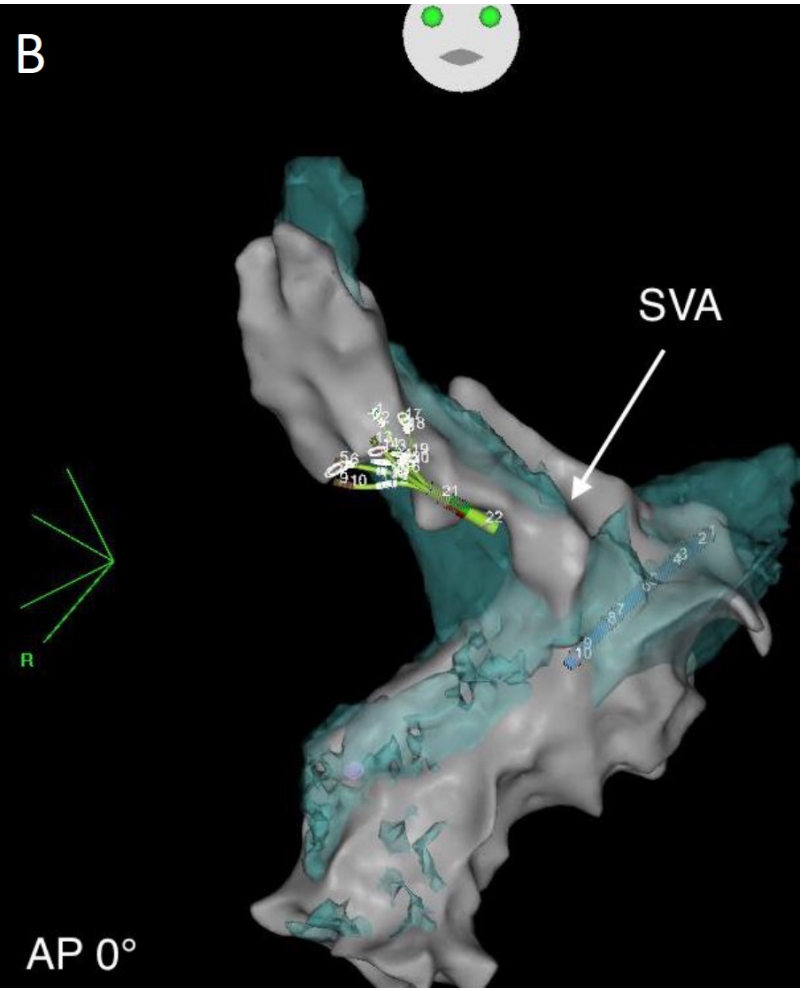



L
R
Scanner

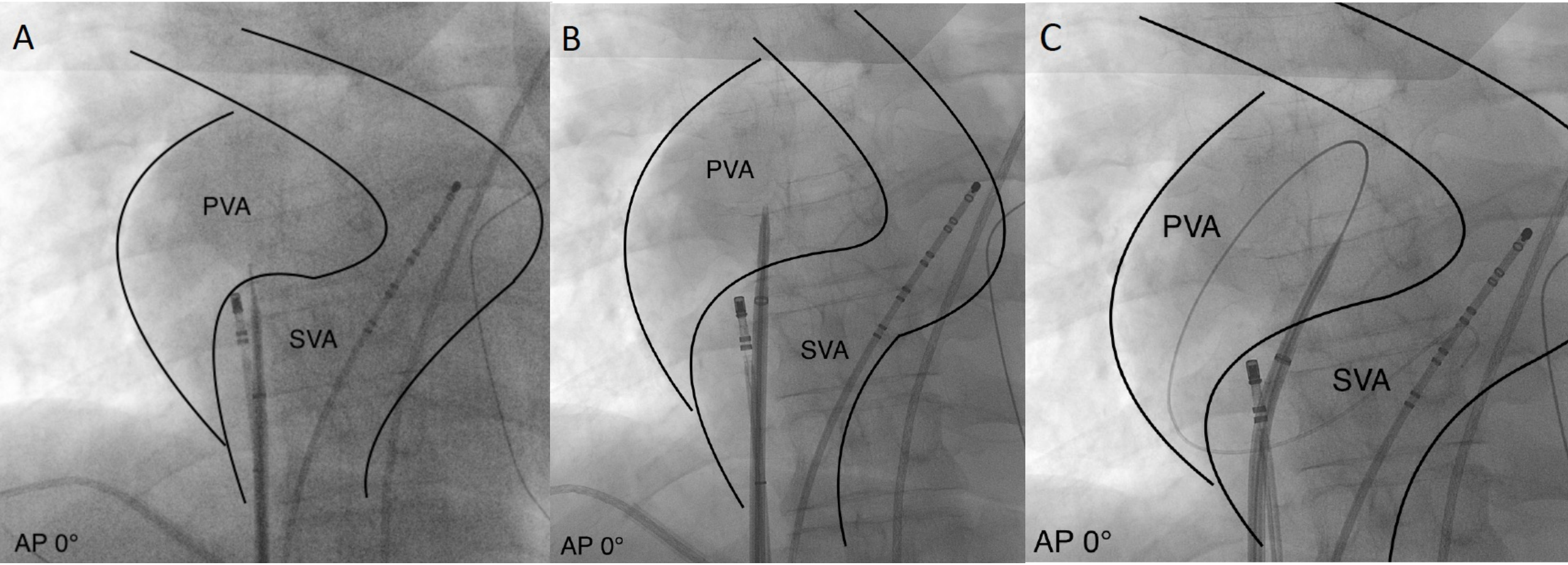
Catheter Ablation
Access Issues

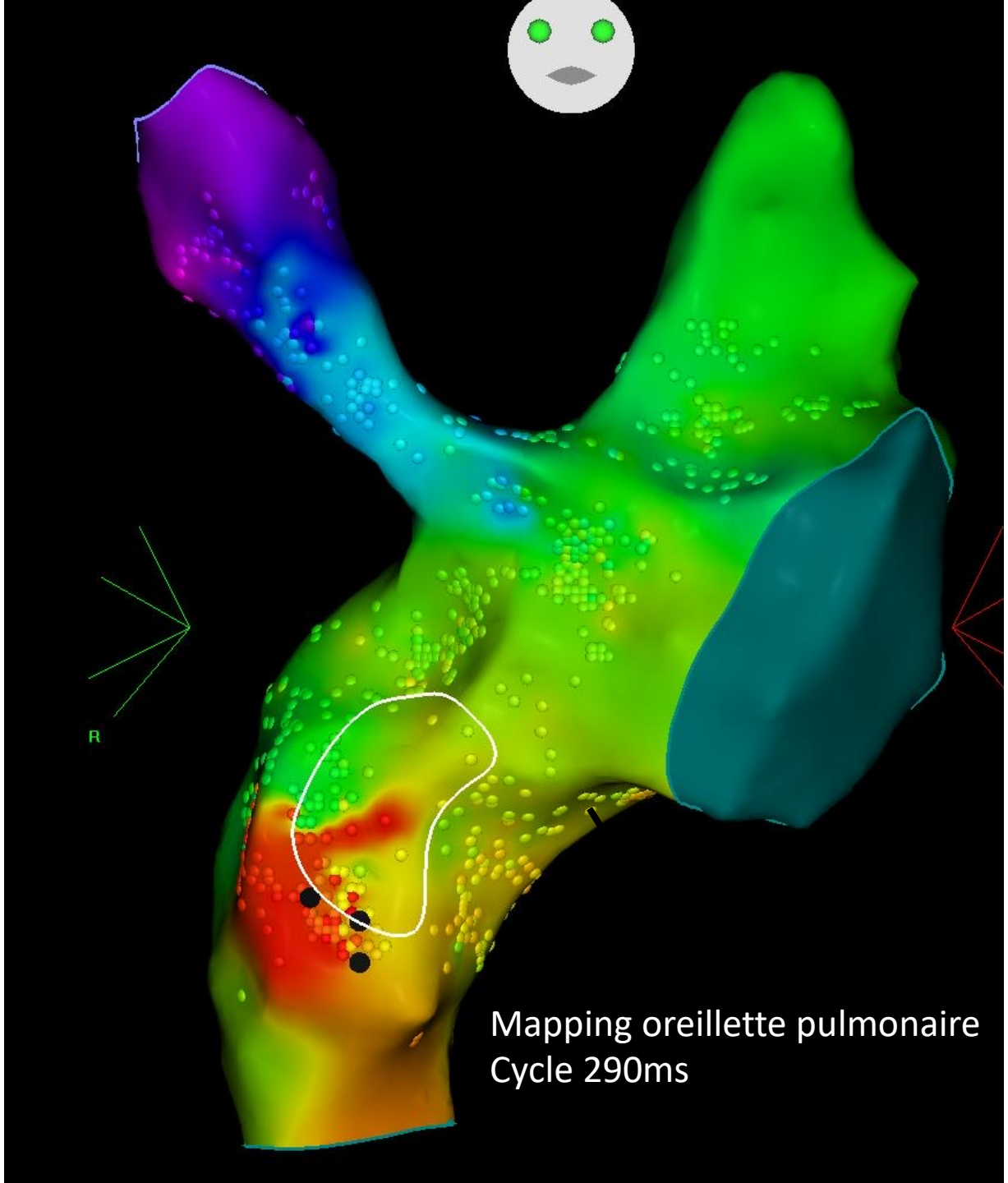






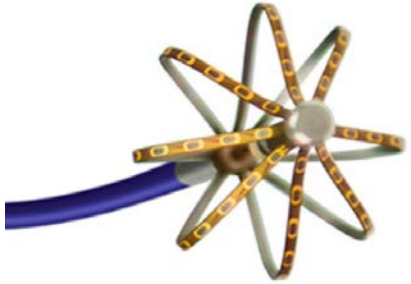
Catheter Ablation
Access Issues



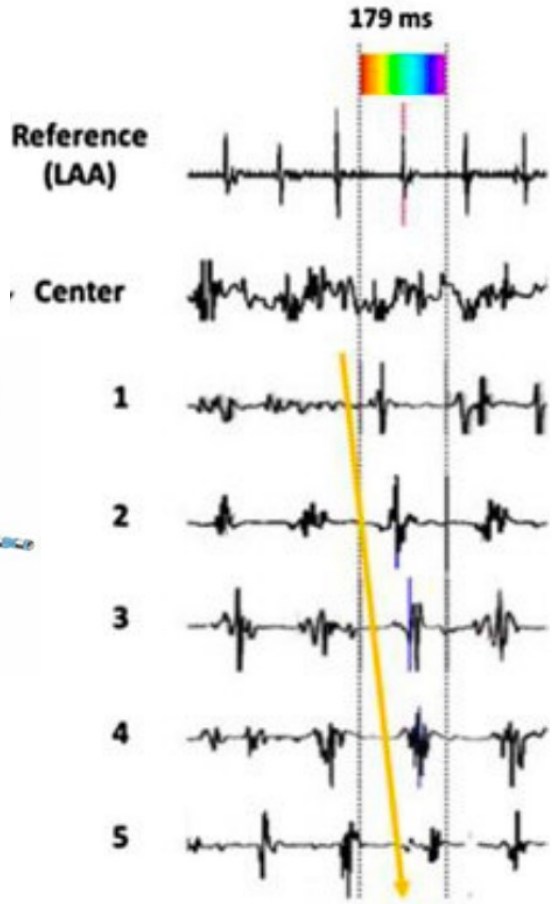


Mapping oreillette pulmonaire
Cycle 290ms

CARTO

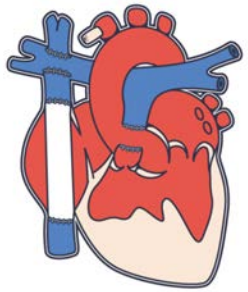


RHYTHMIA

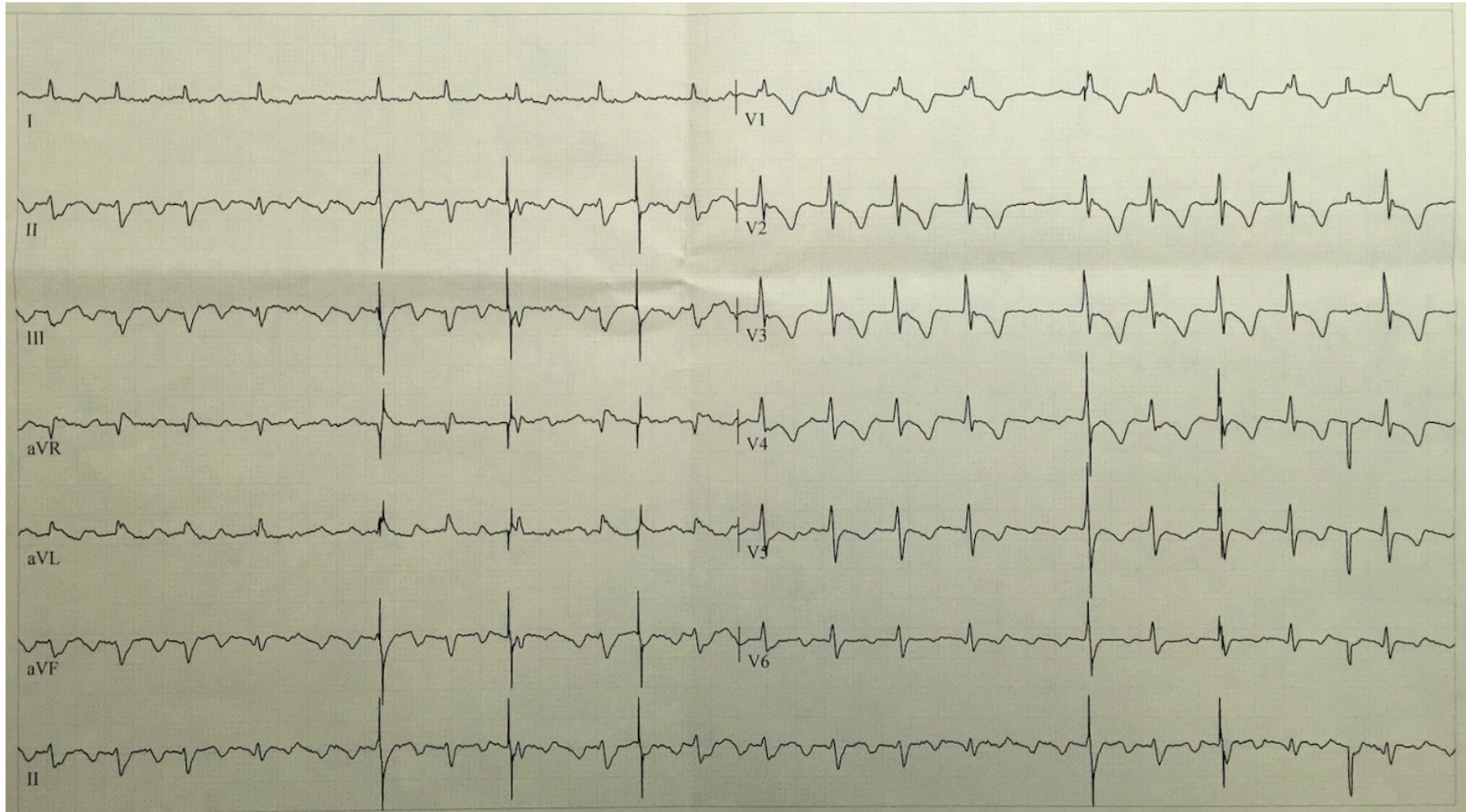


PRECISION

Case n°2

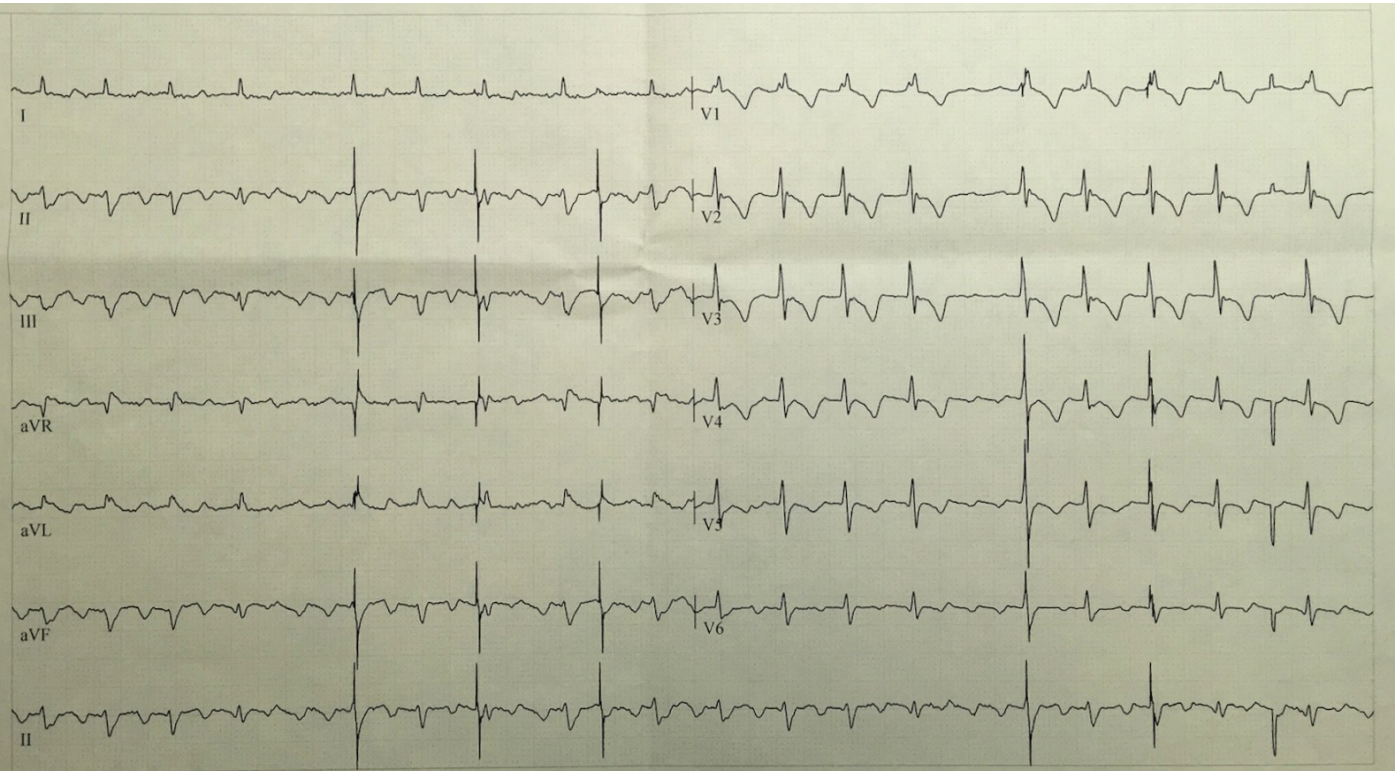


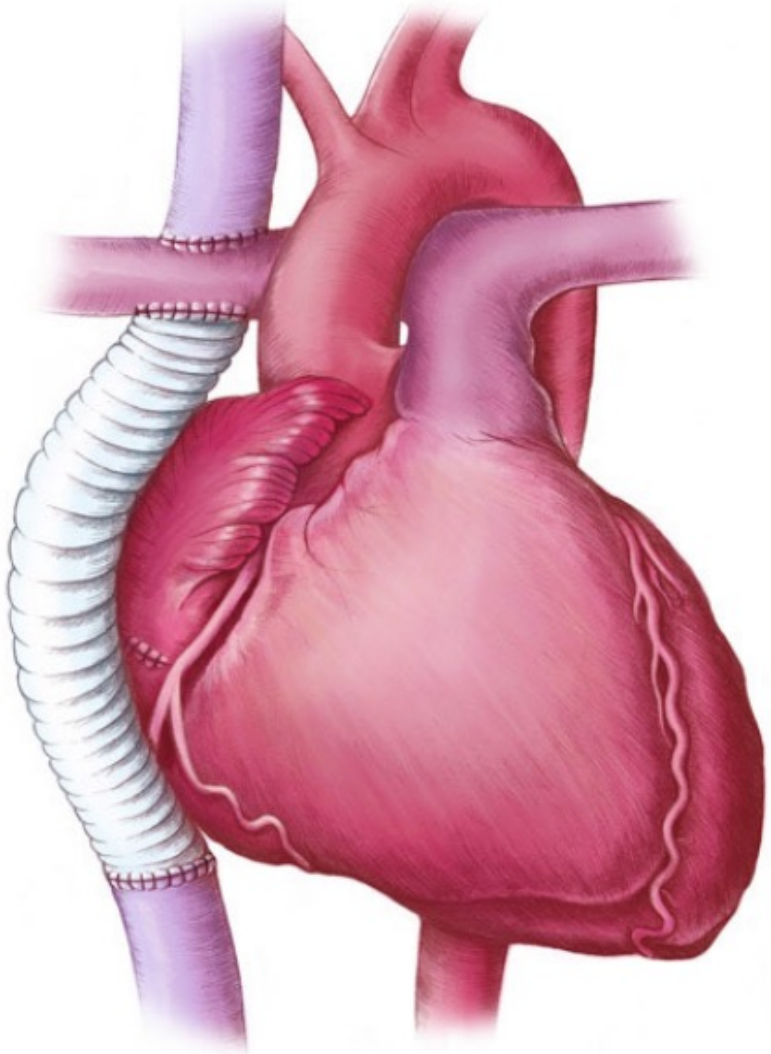
Woman, 46 y, tricuspid atresia
Extracardiac Fontan
Palpitations ++



Case n°2

Woman, 46 y, tricuspid atresia
Extracardiac Fontan
Palpitations ++

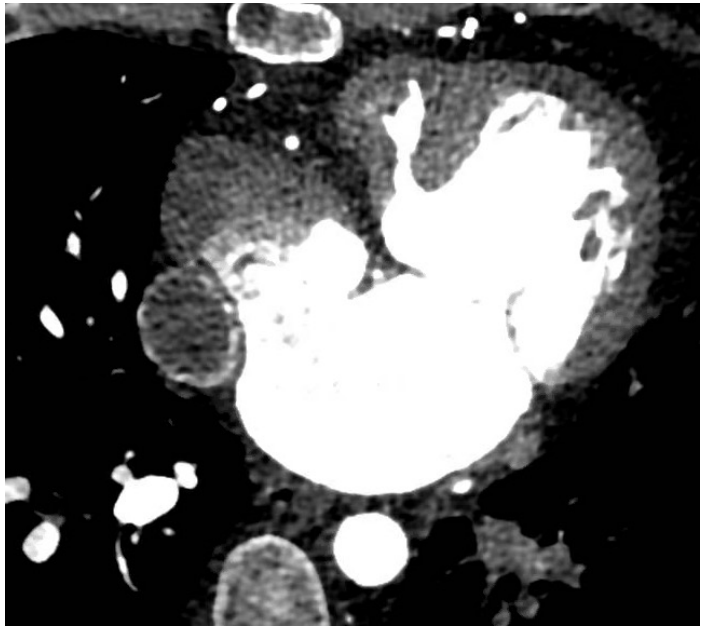


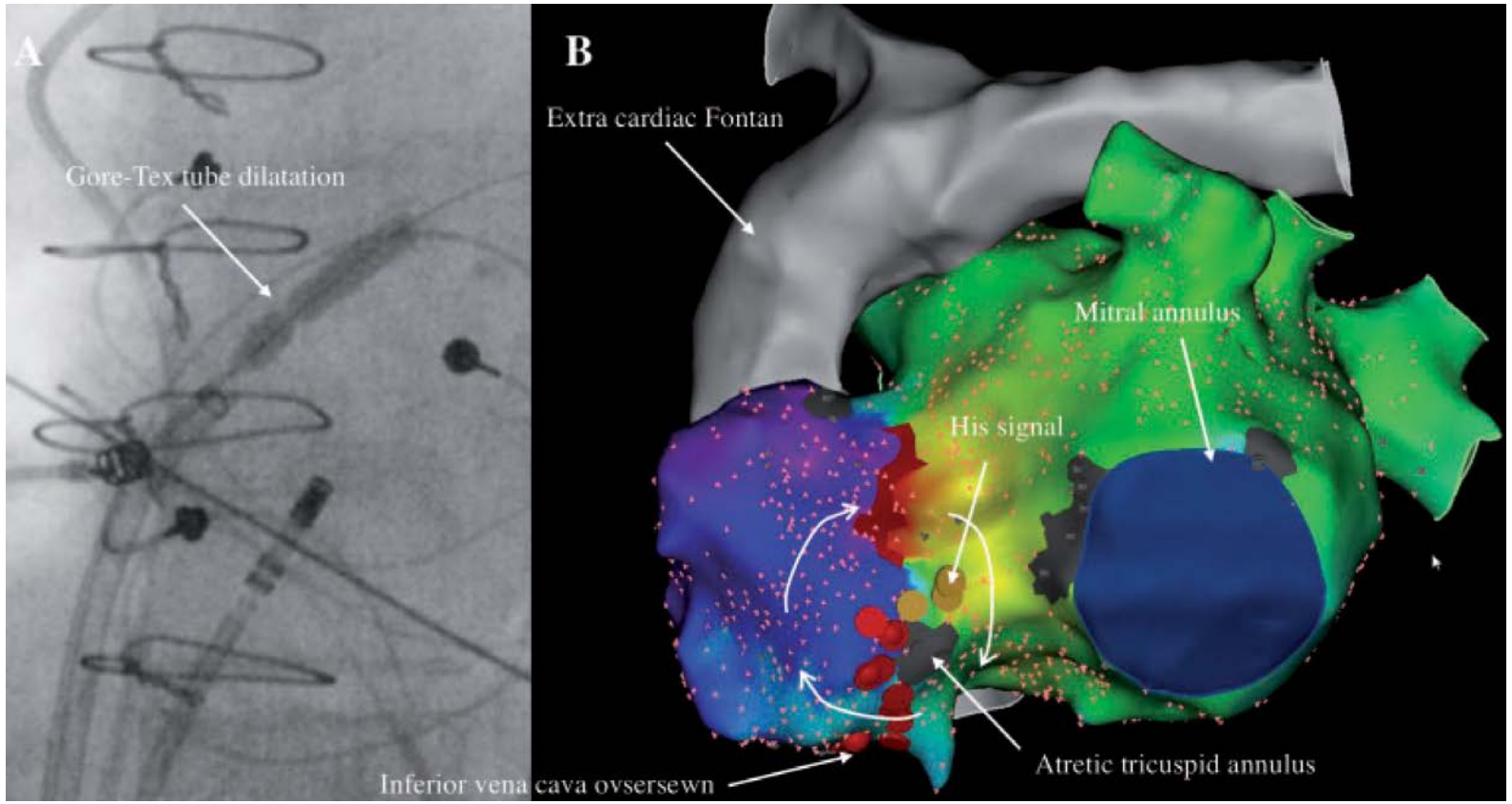


→ Ablation ?

Cardiac Access: Challenging Approaches

1/ Gore-Tex Tube Puncture



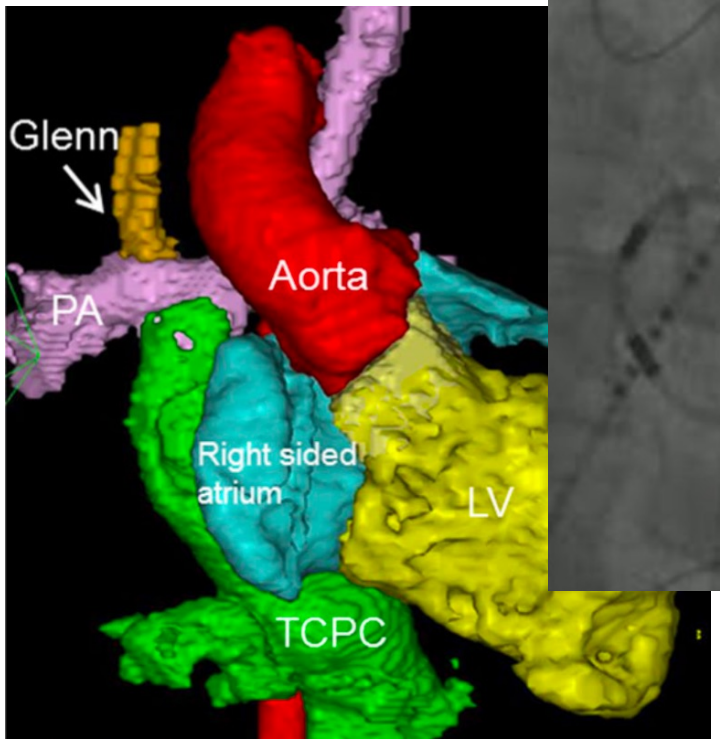


Waldmann V, et al. Europace 2019

Cardiac Access: Challenging Approaches

2/ Remote Magnetic Navigation

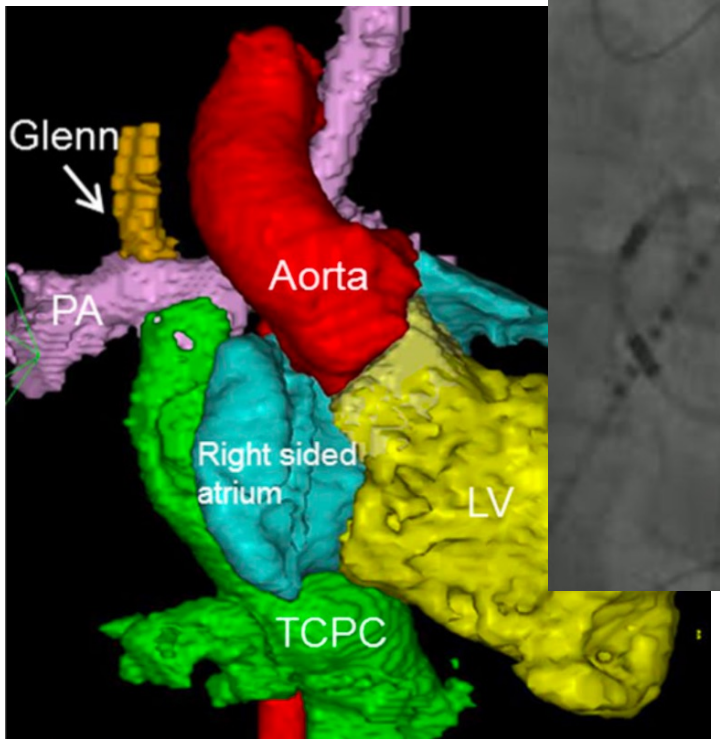
Retrograde aortic approach

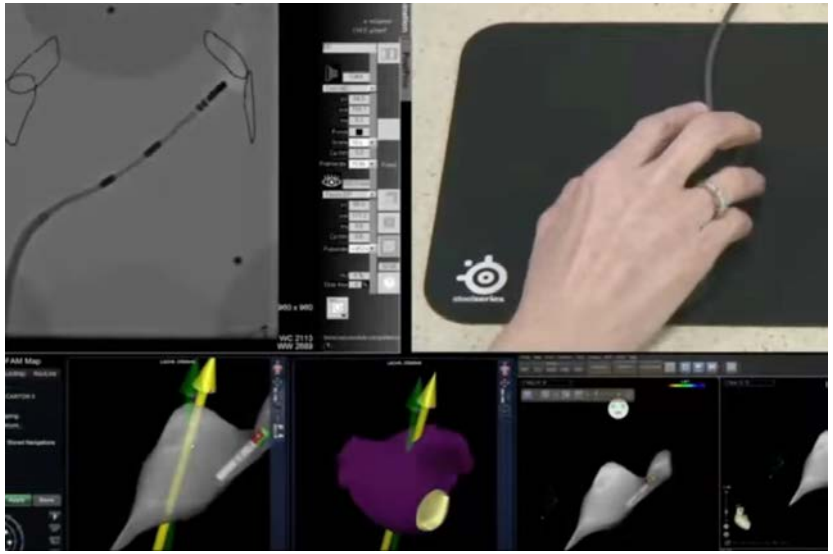


Cardiac Access: Challenging Approaches

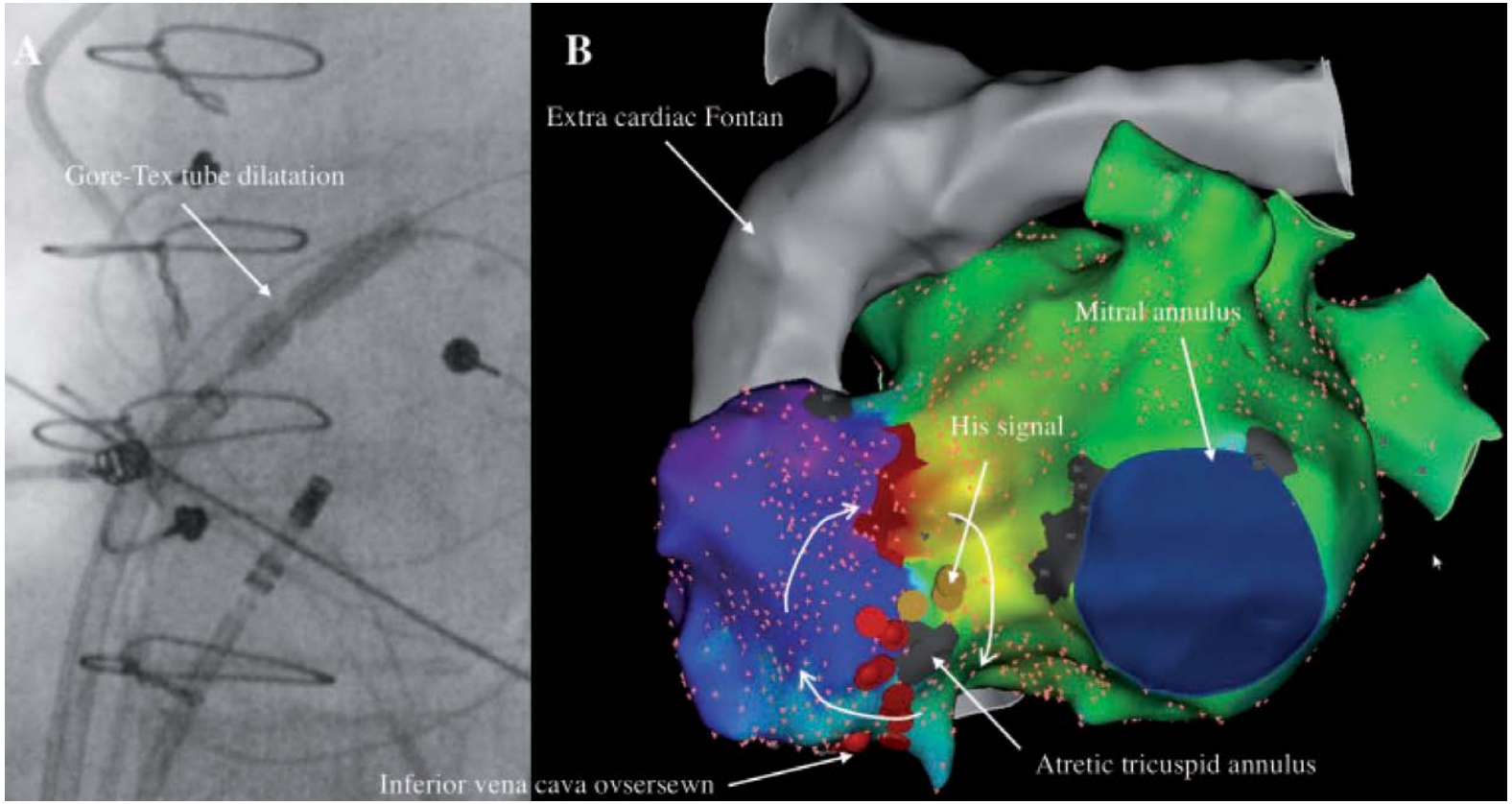
2/ Remote Magnetic Navigation

Retrograde aortic approach





Case n°2



Acute success
No reccurence at 6 months

→ Stop anticoagulation ?

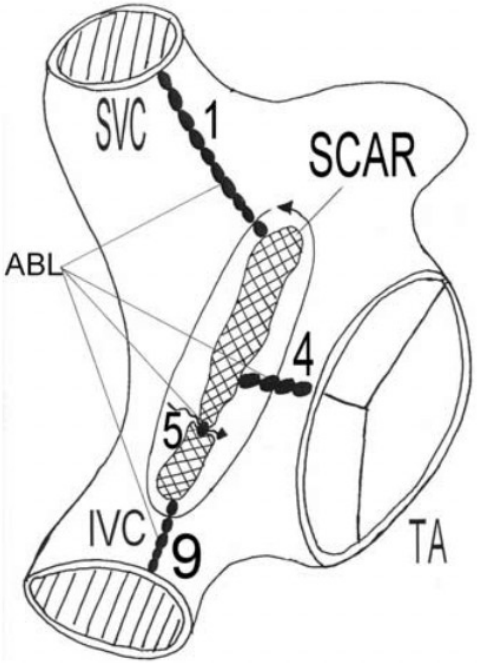
Waldmann V, et al. Europace 2019

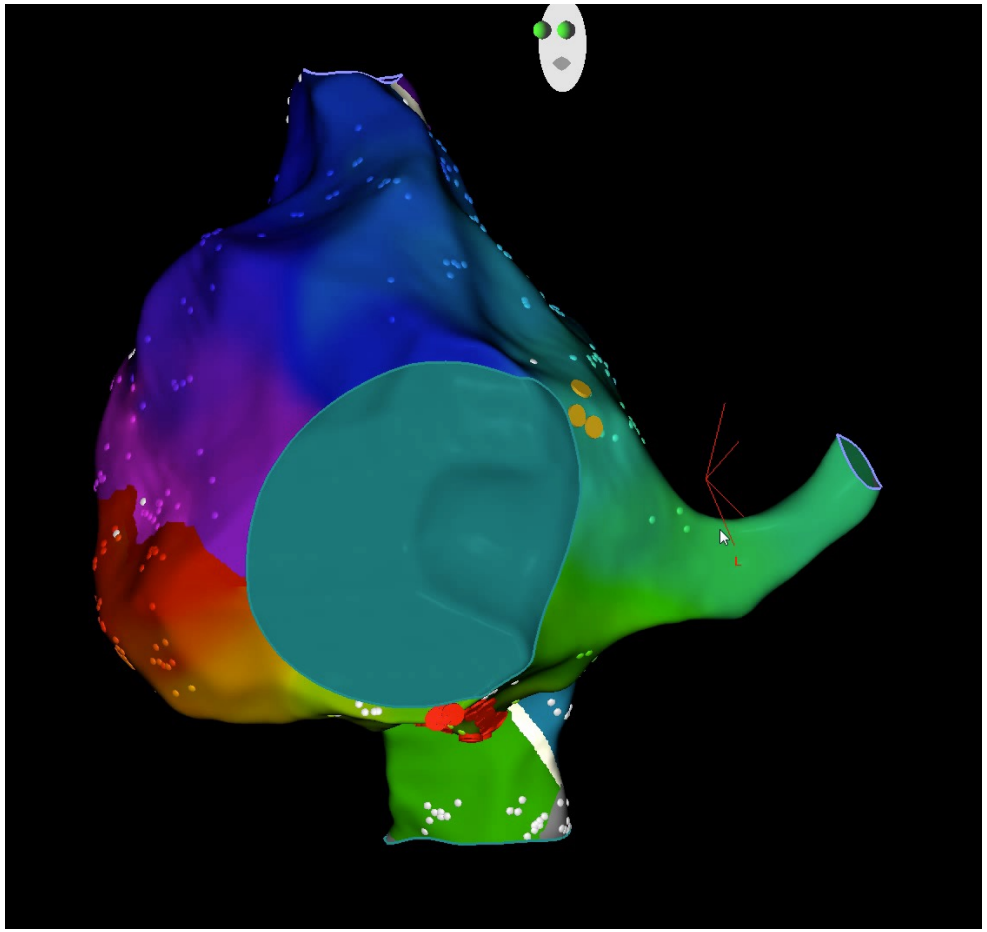
Stop Anticoagulation ?

- **No clear guidelines**
- **Depend on multiple factors**
  - History of atrial arrhythmia
  - CHD complexity
  - Importance of atrial remodelling
  - Number of inducible tachycardia
  - Arrhythmia type: cavotricuspid flutter or complex circuit
  - Associated thromboembolic factors
  - Symptoms during arrhythmia
- **Collegial discussion between experts ++**
- **Close follow-up (holter, PM/ICD, reveal…)**

Circuits of Atrial Arrhythmias

- **Intra-Atrial Reentrant Tachycardia >> Focal**
- **>50% = cavotricuspid isthmus**
- **Around atriotomy scar ++**

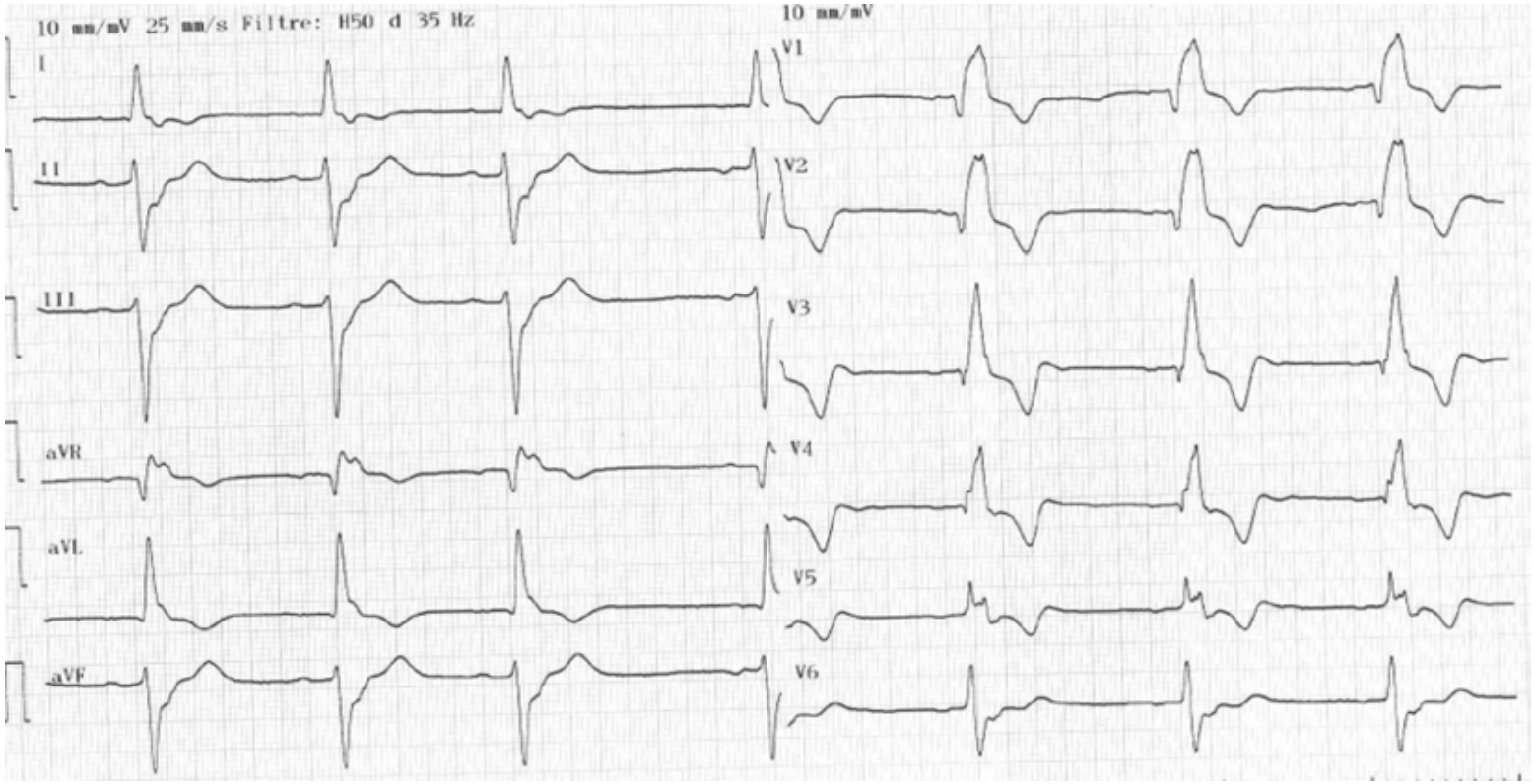




Lukac P, et al. Heart Rhythm 2005

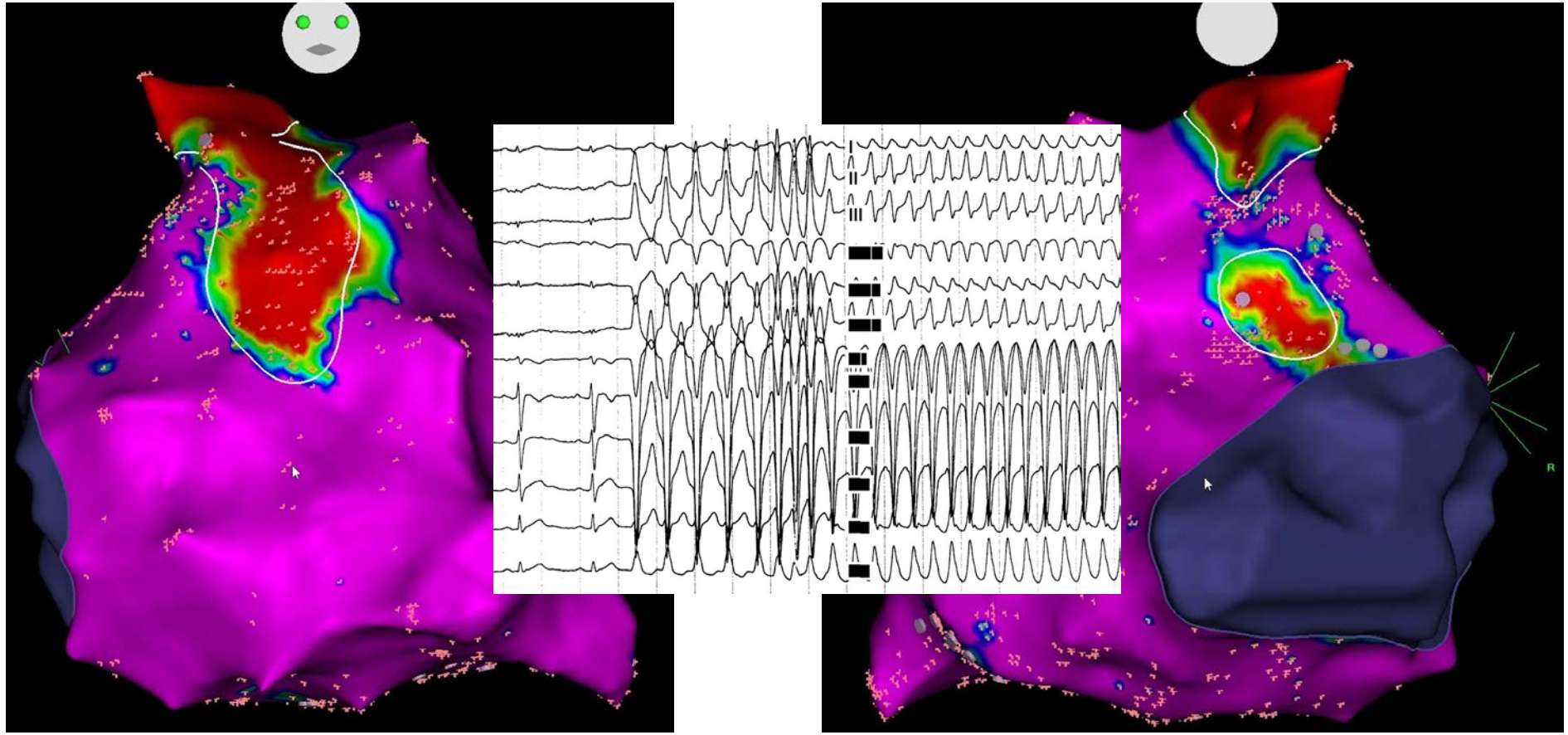
CASE N°3

Man, 38 y, tetralogy of Fallot
Severe pulmonary regurgitation, indication for PVR
Holter = NSVT



→ **What to plan before ?**

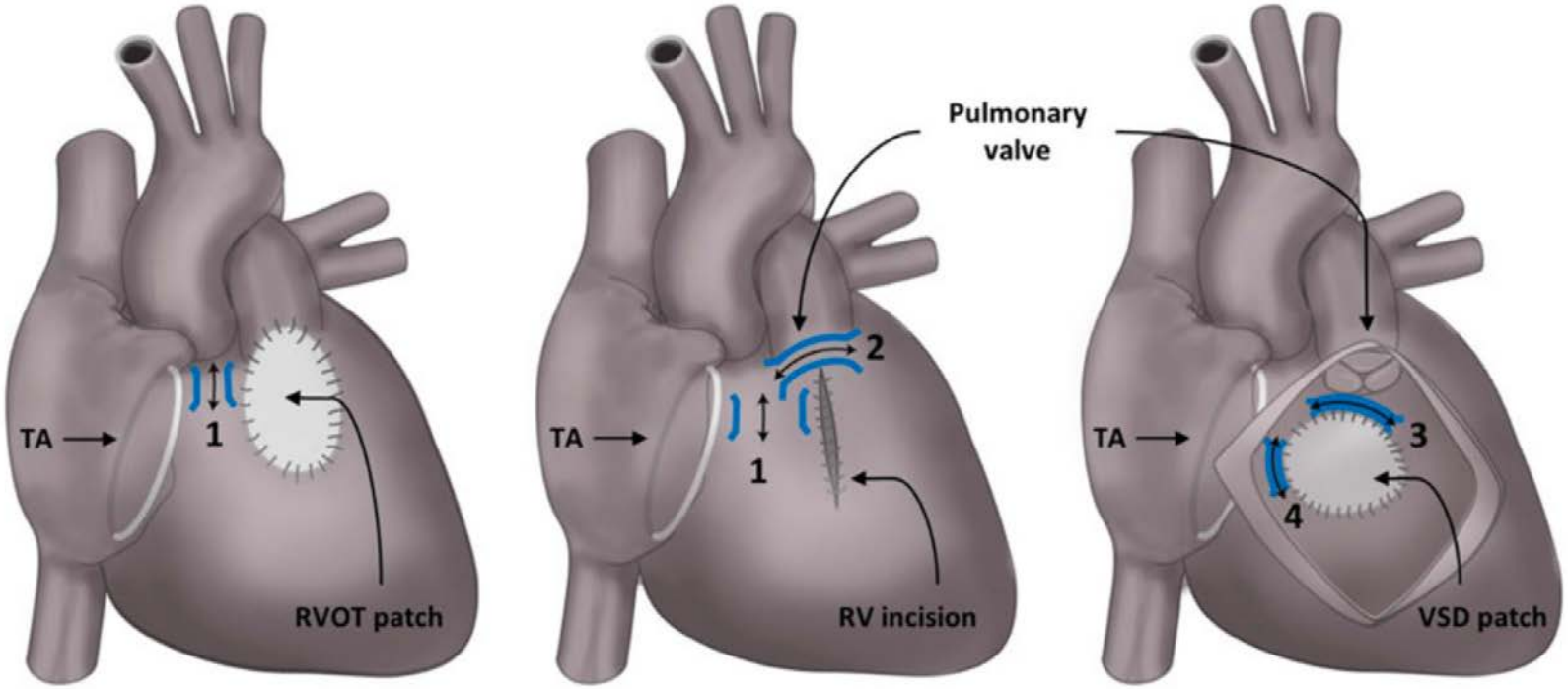
Programmed Ventricular Stimulation



Voltage Map and Ventricular Stimulation

Programmed Ventricular Stimulation

4 Anatomical Main Isthmus

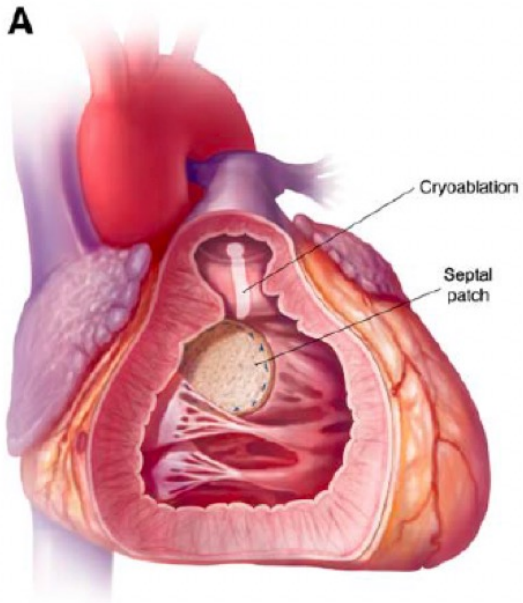


Kapel GFL, et al. European Heart Journal 2017

Programmed Ventricular Stimulation

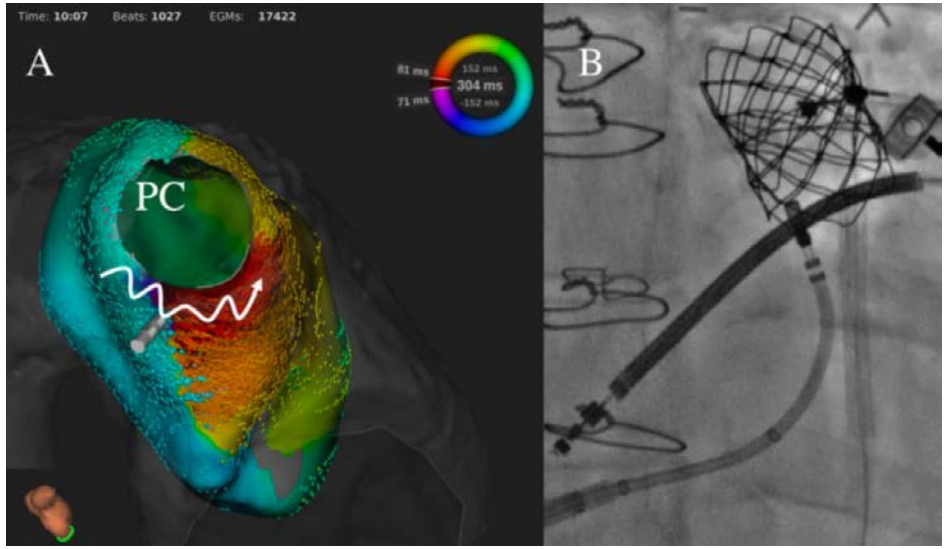
Before Surgery ?

- To guide surgical ablation if needed



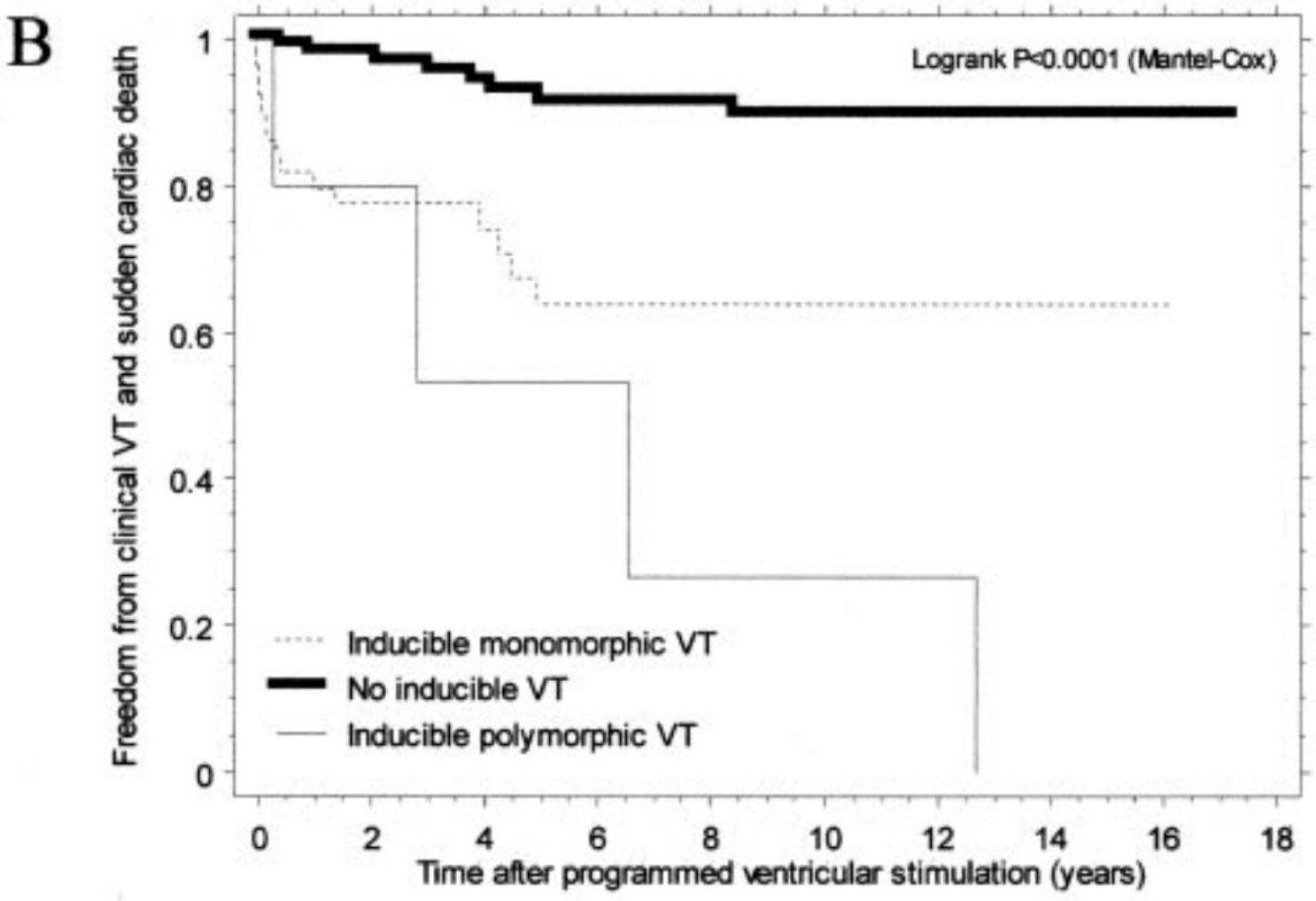
Before percutaneous approach ?

- Critical isthmus can be covered by valve
- To limit infectious risk (Melody ++)



Combes N, et al. European Heart Journal 2019

Programmed Ventricular Stimulation



- 252 ToF
- Median follow-up
  - 18.5 y after surgery
  - 6.5 y after PVS
- PVS positive in 35%
- Outcome in 62 (25%) patients

Khairy P, et al. Circulation 2004

Programmed Ventricular Stimulation

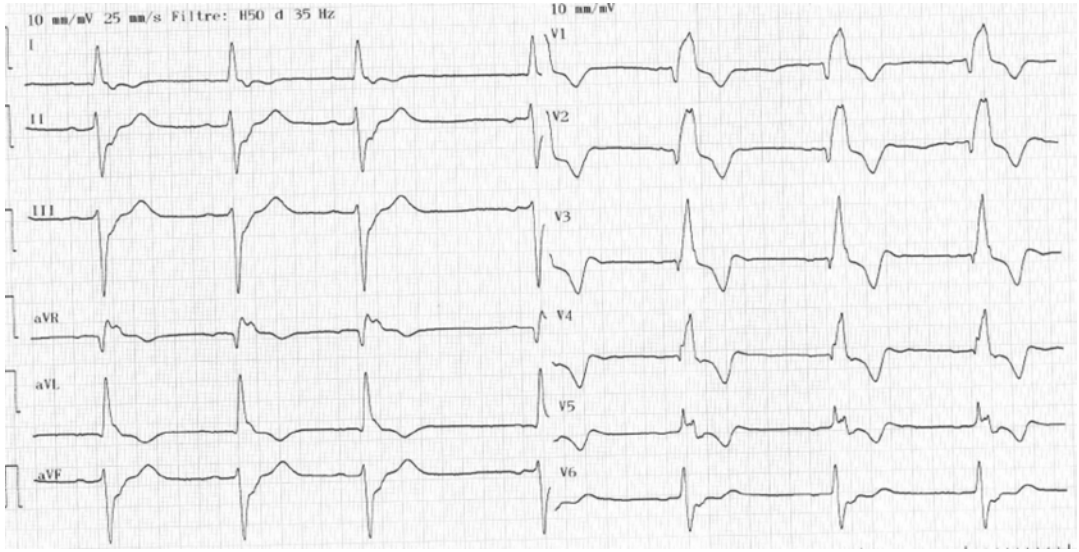
TABLE 6. Programmed Ventricular Stimulation in Routine Screening Versus Clinical Indication for Testing

	Routine Screening (n=93)	Clinical Indication (n=159)	*P*
Sensitivity, %	85.7±13.2	76.4±5.7	1.0000
Specificity, %	79.1±4.4	79.8±3.9	1.0000
Diagnostic accuracy, %	79.6±4.2	78.9±3.2	1.0000
Positive predictive value, %	25.0±8.8	66.7±5.9	0.0007
Negative predictive value, %	98.6±1.4	86.4±3.5	0.0084

Khairy P, et al. Circulation 2004

CASE N°3

Man, 38 y, tetralogy of Fallot
Severe pulmonary regurgitation, indication for PVR
Holter = NSVT
Wide QRS
PVS negative



→ **Indication for Implantable Cardioverter Defibrillator ?**

ICD in CHD

Recommendations	Consensus statement
ICD is recommended for patients with CHD who are survivors of an aborted cardiac arrest due to VF or haemodynamically unstable VT after evaluation to define the cause of the event and exclusion of any reversible causes.	
ICD is recommended for patients with CHD with symptomatic sustained VT who have undergone haemodynamic and electrophysiological evaluation.	
ICD is recommended in adults with CHD and a systemic LVEF ≤ 35%, biventricular physiology and NYHA functional Class II or III.	
ICD implantation should be considered in patients with CHD and syncope of unknown origin in the presence of either advanced ventricular dysfunction or inducible sustained VT or VF on VPS.	
ICD implantation should be considered in selected patients with TOF and multiple risk factors for SCD, including LV dysfunction, non-sustained VT, QRS duration ≥180 ms, or inducible sustained VT on VPS.	
ICD therapy may be considered in patients with advanced single or systemic RV dysfunction in the presence of risk factors such as non-sustained VT, NYHA functional Class II or III, QRS duration ≥ 140 ms or severe systemic AV valve regurgitation.	
ICD therapy may be considered for non-hospitalized adults with CHD awaiting heart transplantation.	

Hernandez-Madrid A, et al. Europace 2018

ICD IN CHD

Recommendations	Consensus statement
ICD is recommended for patients with CHD who are survivors of an aborted cardiac arrest due to VF or haemodynamically unstable VT after evaluation to define the cause of the event and exclusion of any reversible causes.	♥
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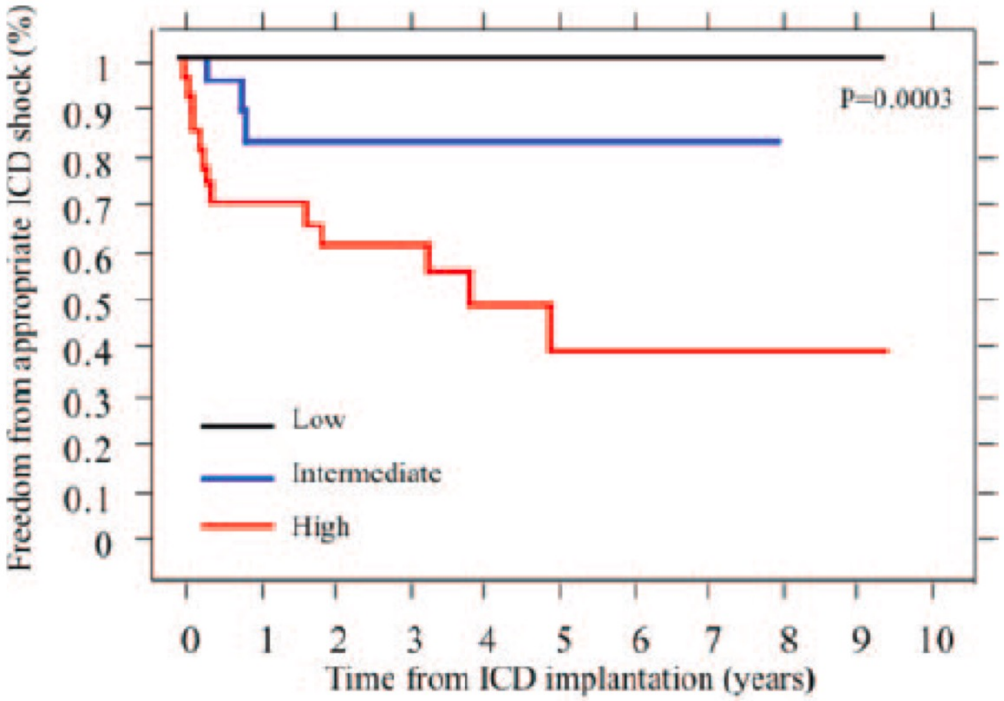
ICD therapy may be considered in patients with advanced single or systemic RV dysfunction in the presence of risk factors such as non-sustained VT, NYHA functional Class II or III, QRS duration ≥ 140 ms or severe systemic AV valve regurgitation.	♥
ICD therapy may be considered for non-hospitalized adults with CHD awaiting heart transplantation.	♥

- **Low level of evidence**
- **Complex decisions**
- **Need for collegial discussion in expert center**
- **Reveal monitoring in selected patients**

ICD in ToF

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



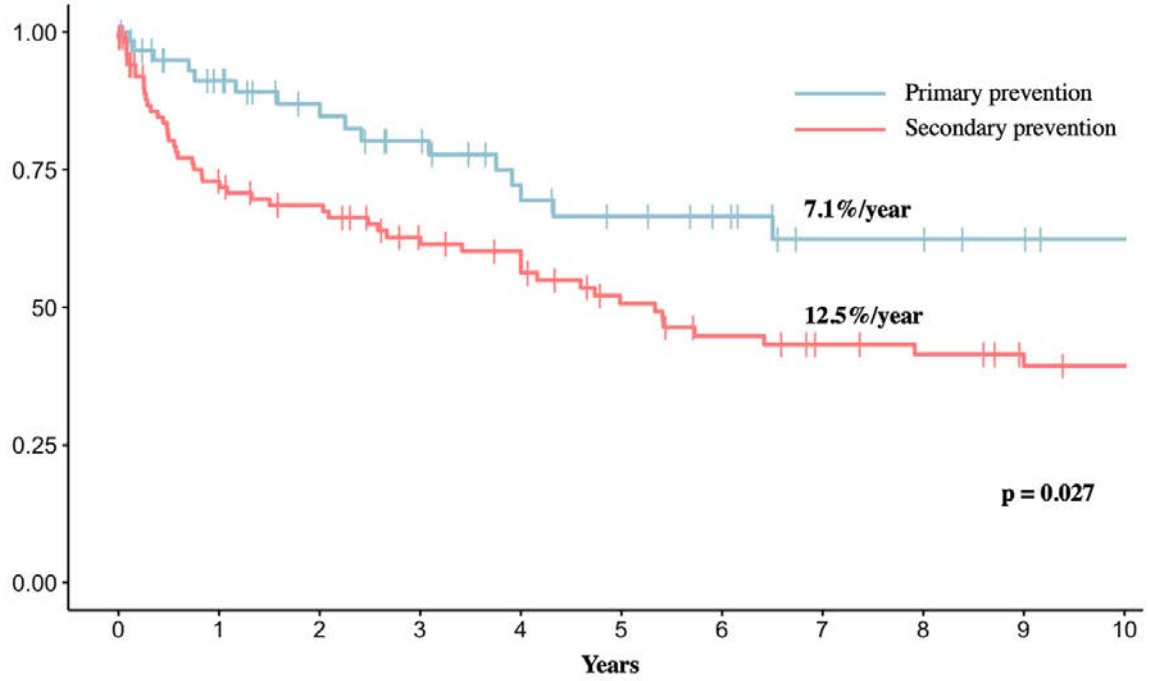
Risk score	Risk category	N	Annualized rate of appropriate shocks
0-2	Low	18	0%
3-5	Intermediate	24	3.8%
6-12	High	26	17.5%

- **Risk score**
- **Difficult to use in daily practice**
  Programmed ventricular stimulation, LVDEP
- **ICD therapies ≠ sudden death !!**

Khairy P, et al. Circulation 2008

ICD in ToF

- **DAI-T4F French National Registry**
- **> 165 patients already included**



Number at risk

Primary prevention	61	47	39	33	26	22	19	13	13	11	9
Secondary prevention	104	68	61	50	46	35	29	25	23	19	18

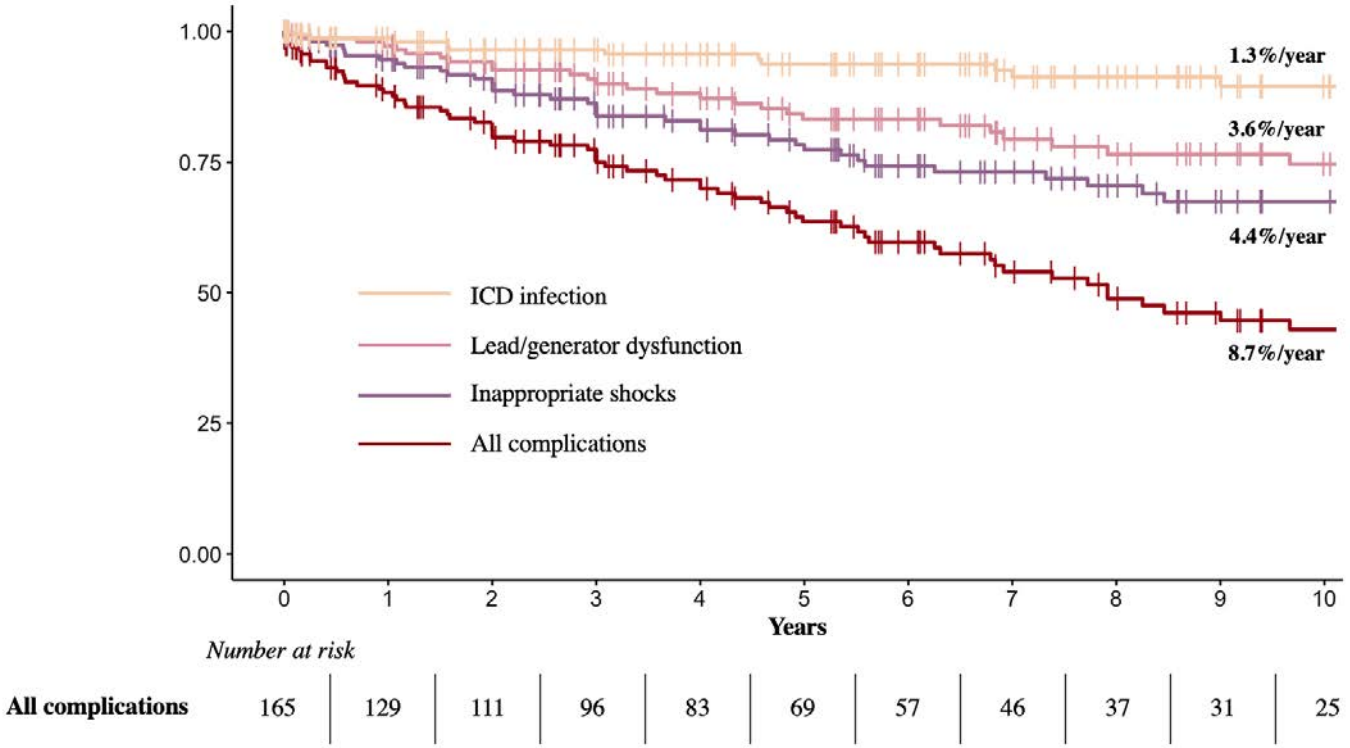
High rate of ICD therapies

BUT …

Waldmann et al. Circulation 2020

ICD in ToF

- **DAI-T4F French National Registry**
- **> 165 patients already included**



High rate of ICD therapies

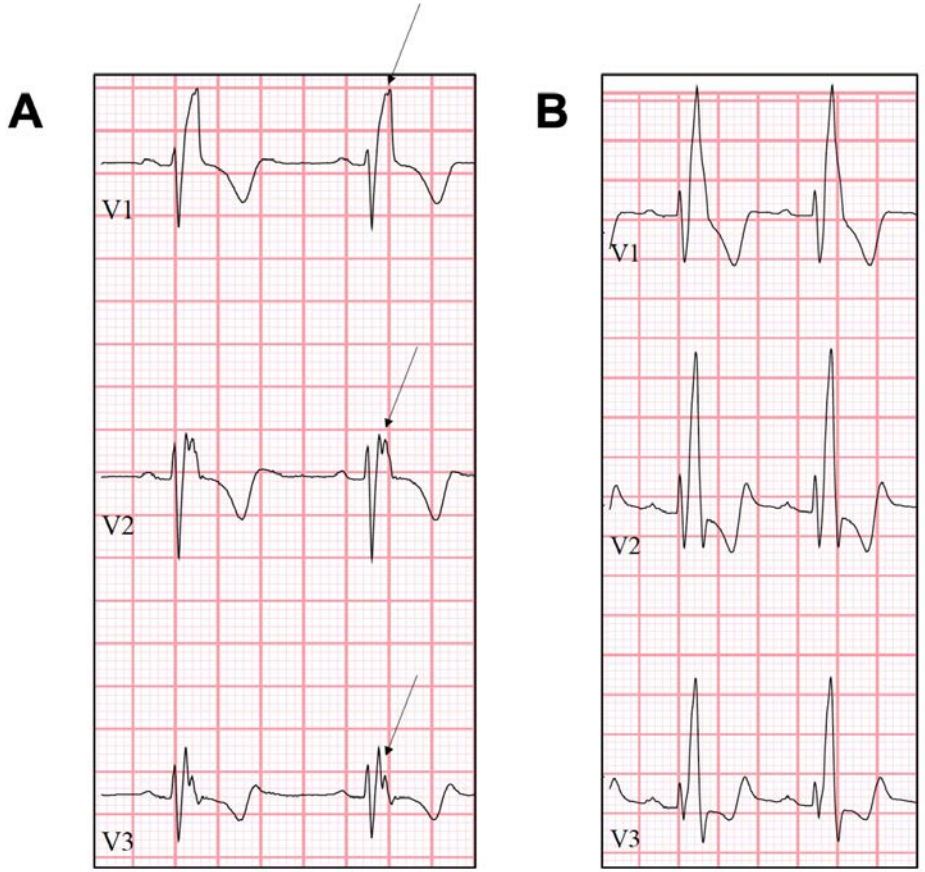
BUT …

High burden of complications ++

Waldmann et al. Circulation 2020

ICD in ToF

- **DAI-T4F French National Registry**
- **> 165 patients already included**

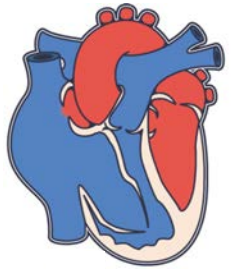


Interest of QRS fragmentation to improve risk stratification ?

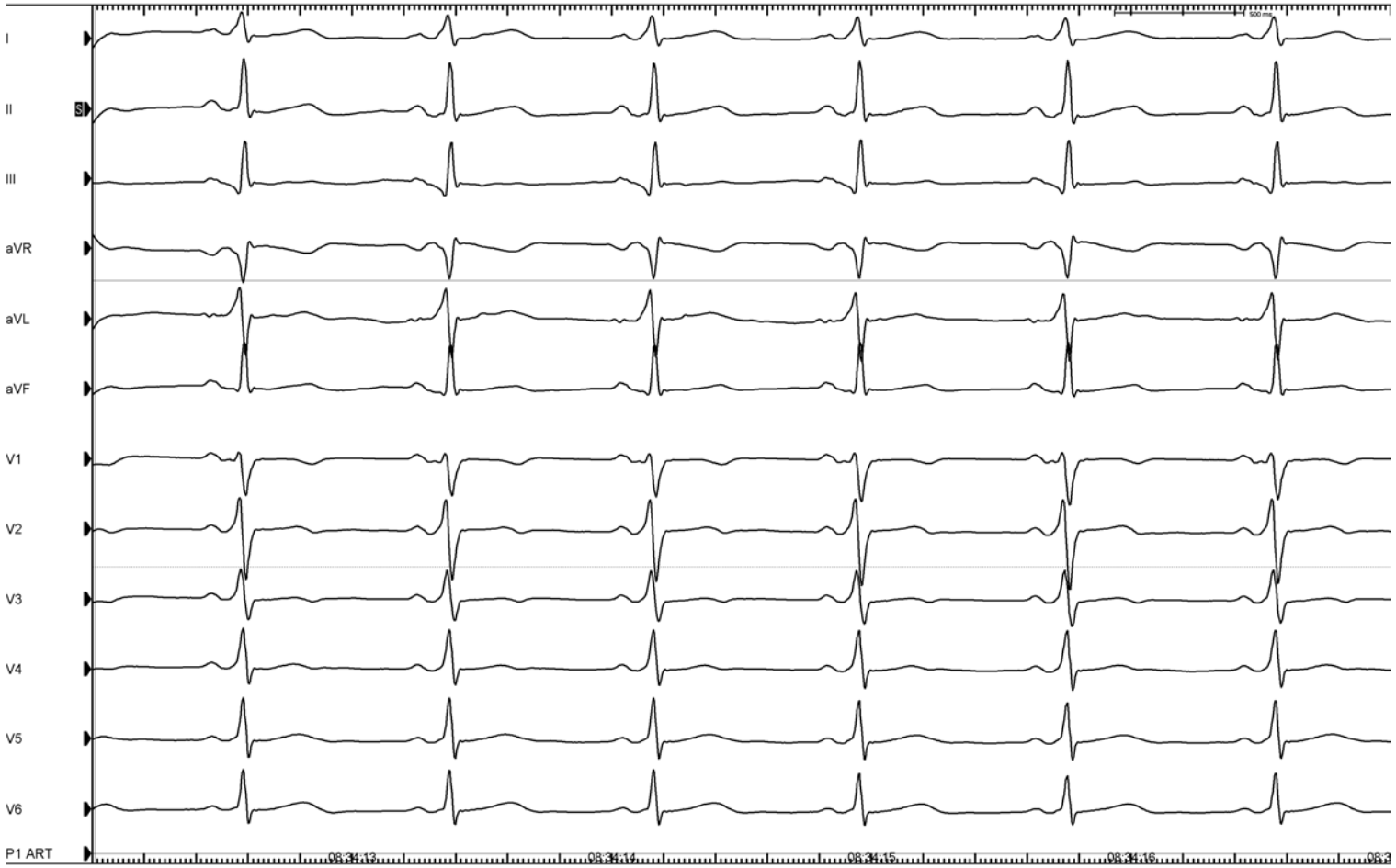
Adjusted HR 3.5 (95%CI 1.2-10.1)

Waldmann et al. Circulation 2020

Case n°4



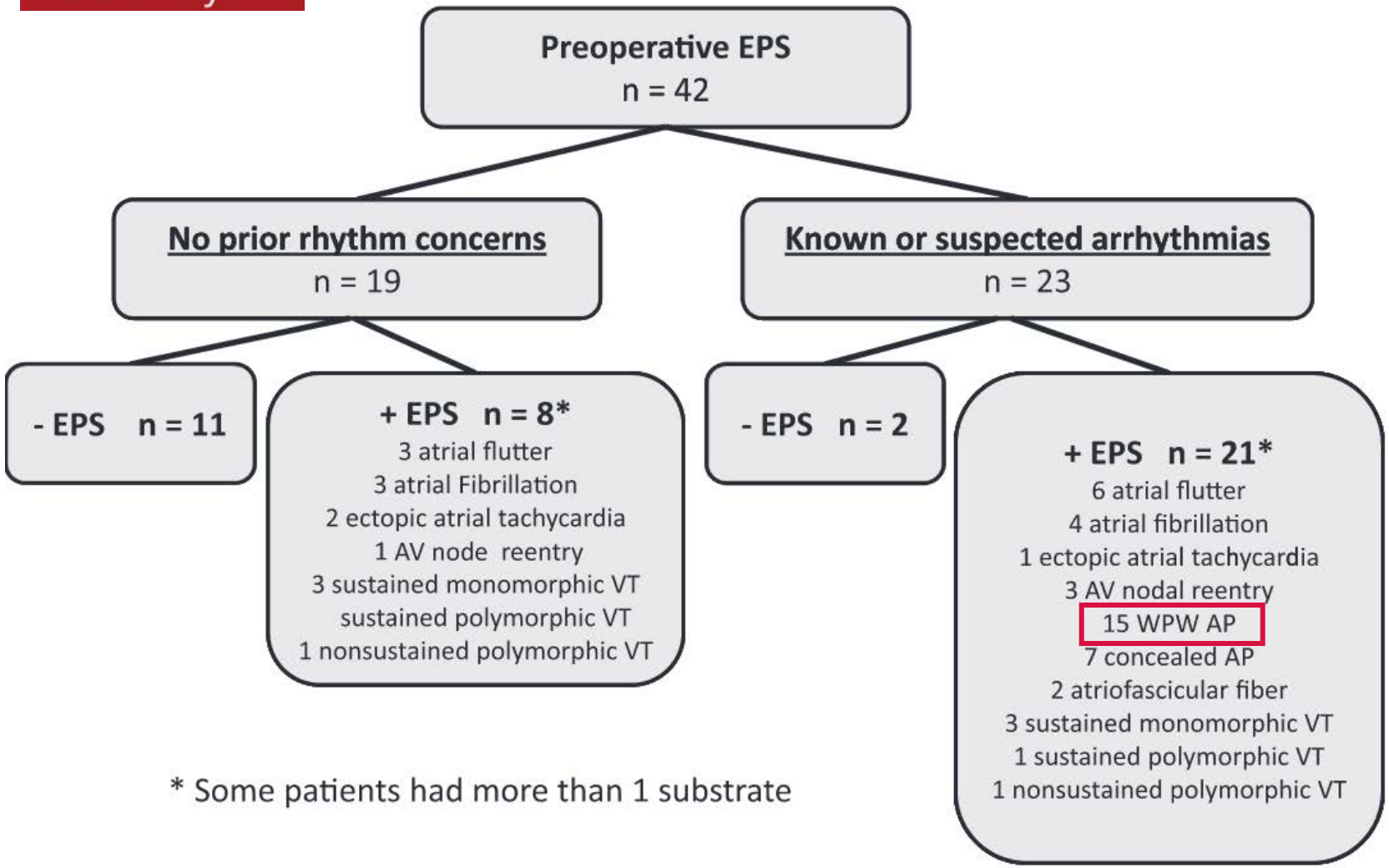
Man, 32 y, Ebstein anomaly
Severe tricuspid regurgitation, right ventricle dilatation
Rare episodes of palpitations
Referred for surgery



→ Do you plan electrophysiology study before surgery ?

Utility of preoperative electrophysiologic studies in patients with Ebstein's anomaly undergoing the Cone procedure

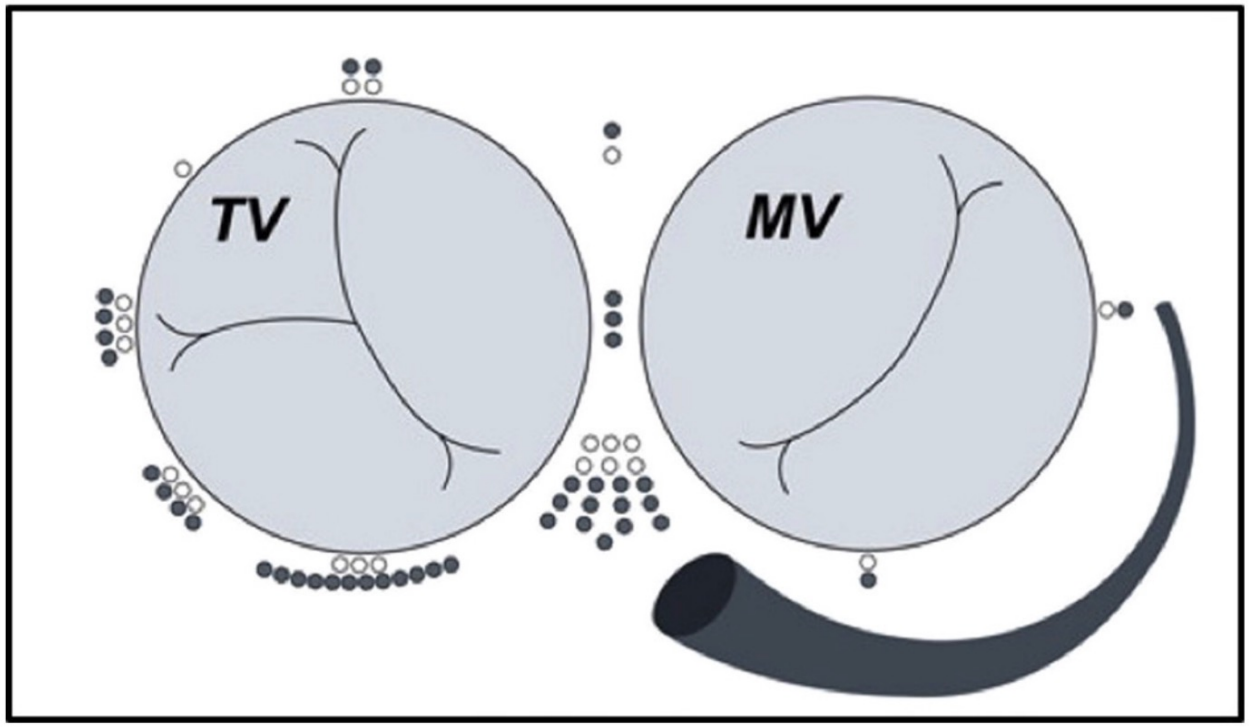
HeartRhythm

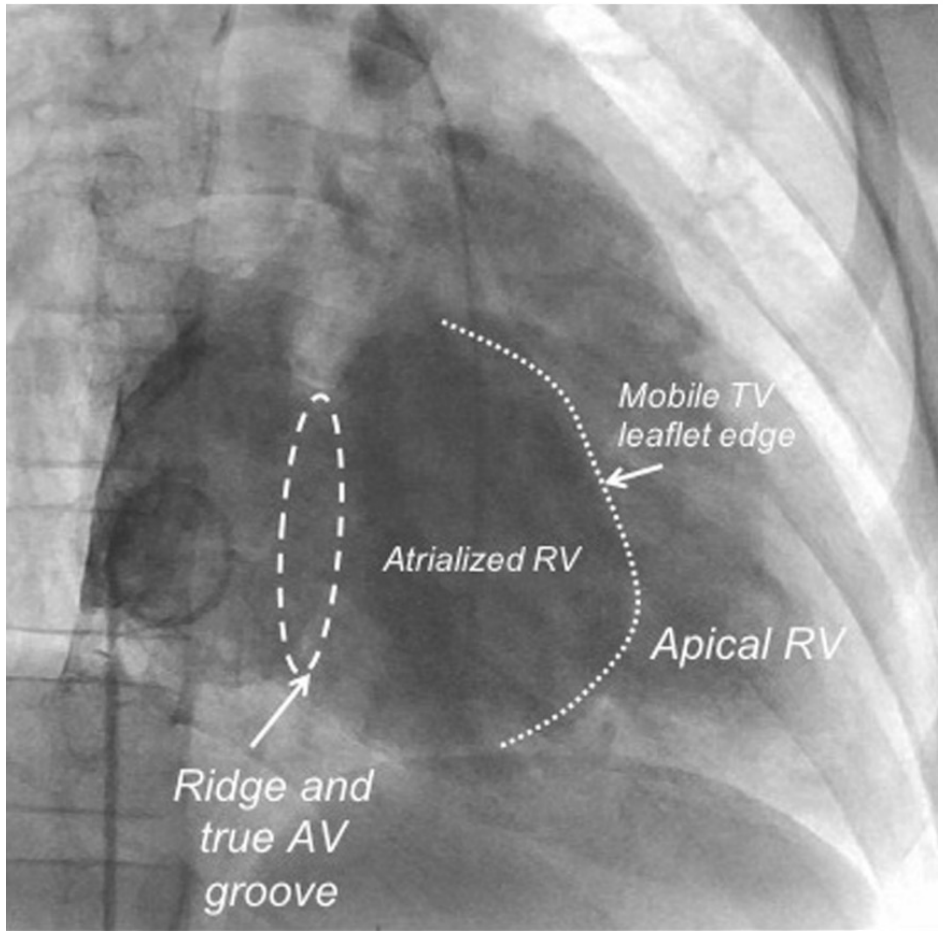


* Some patients had more than 1 substrate

Shivapour et al., Heart Rhythm 2014

Accessory Pathways in Ebstein

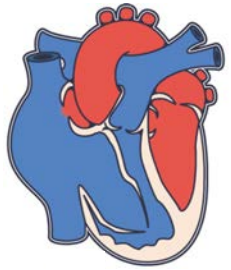


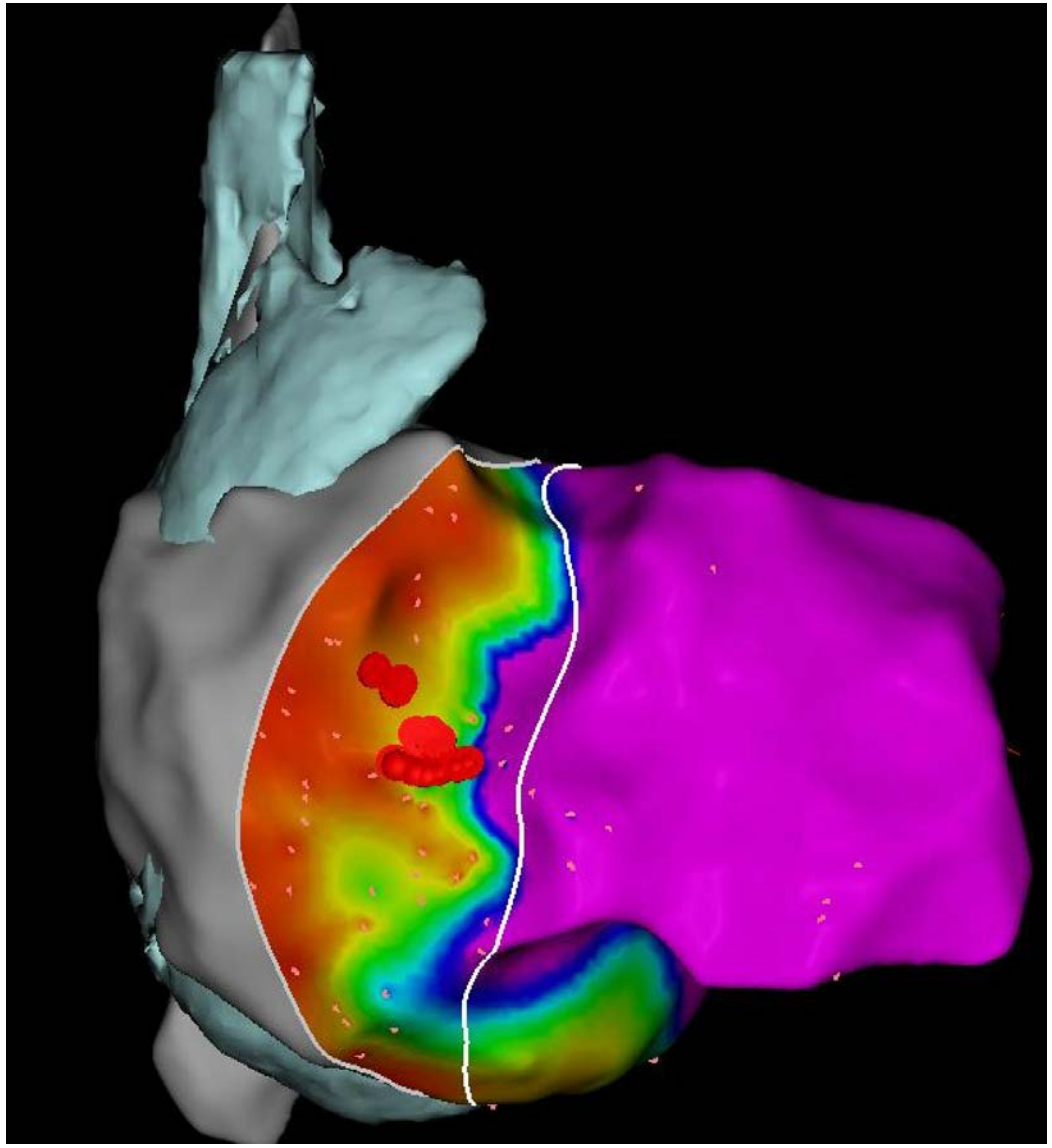


- ≈ 25 %
- Half have multiple AP
- Surgery can limit futur access

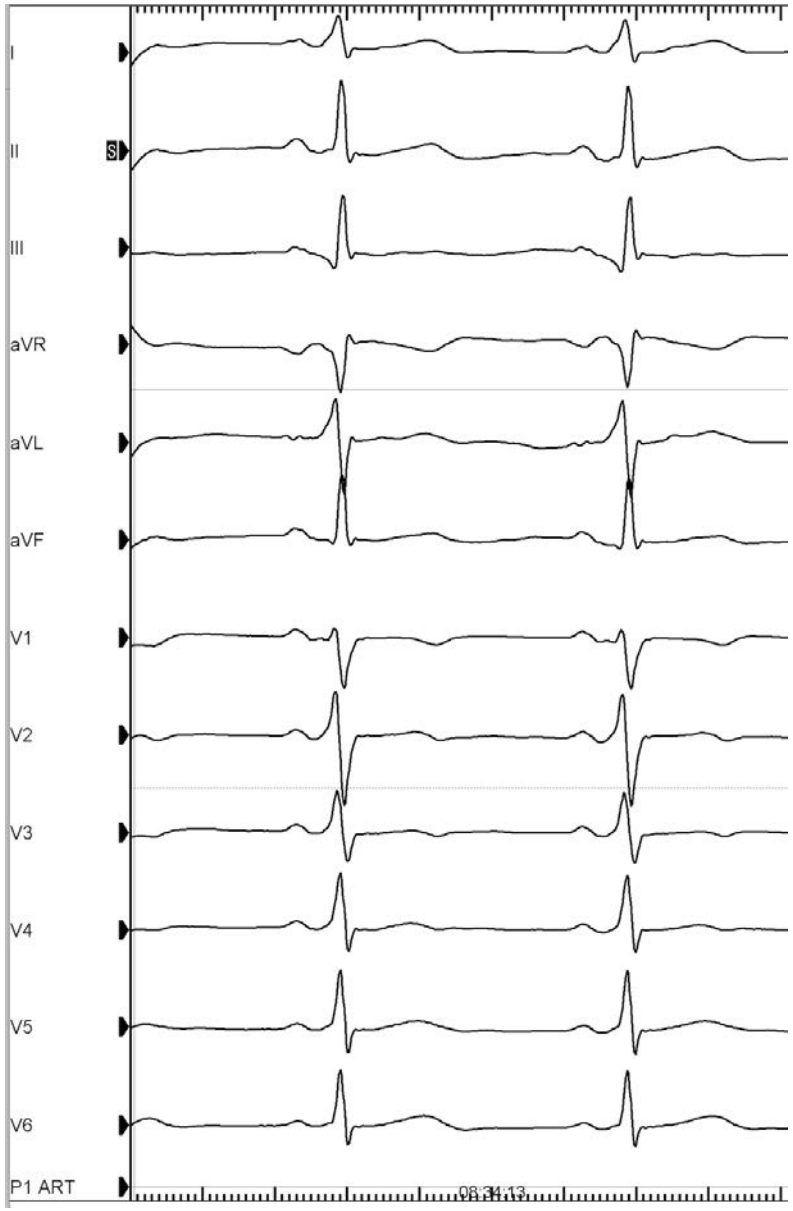
Walsh EP, JACC EP 2018

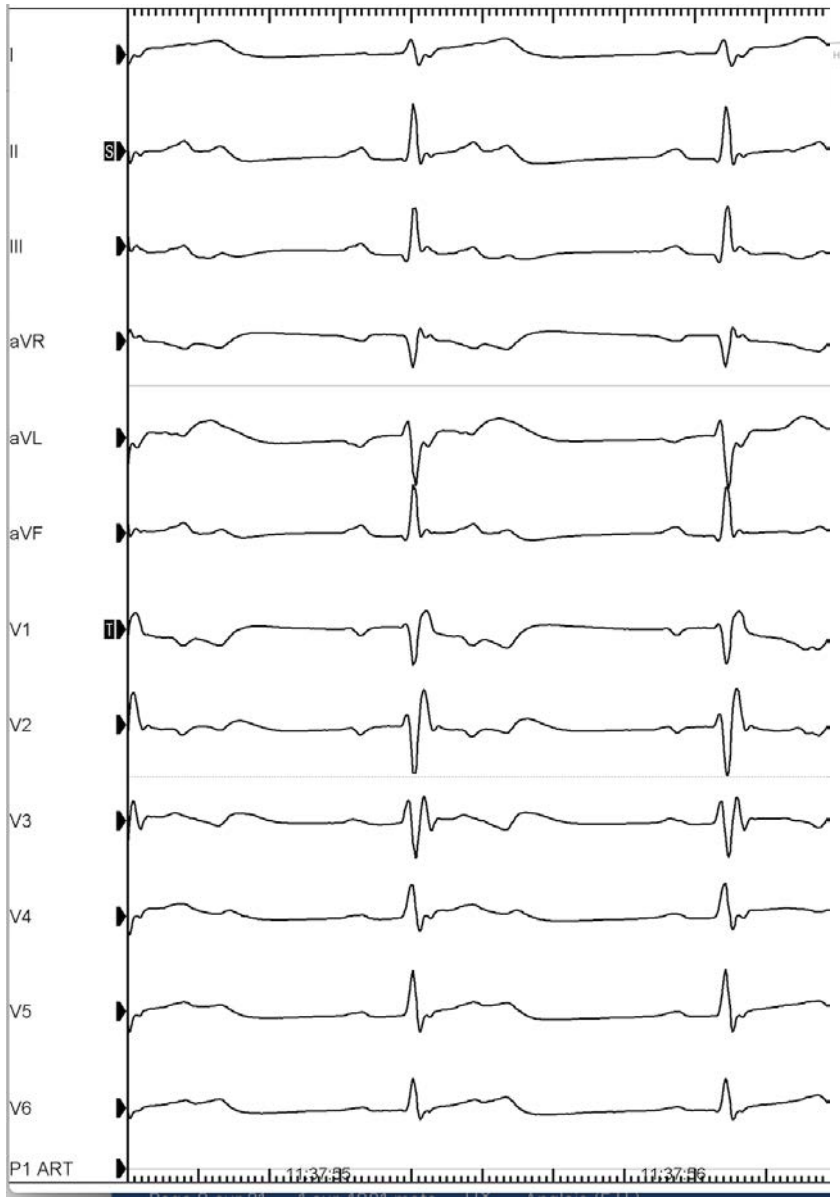
Case n°4





Case n°4

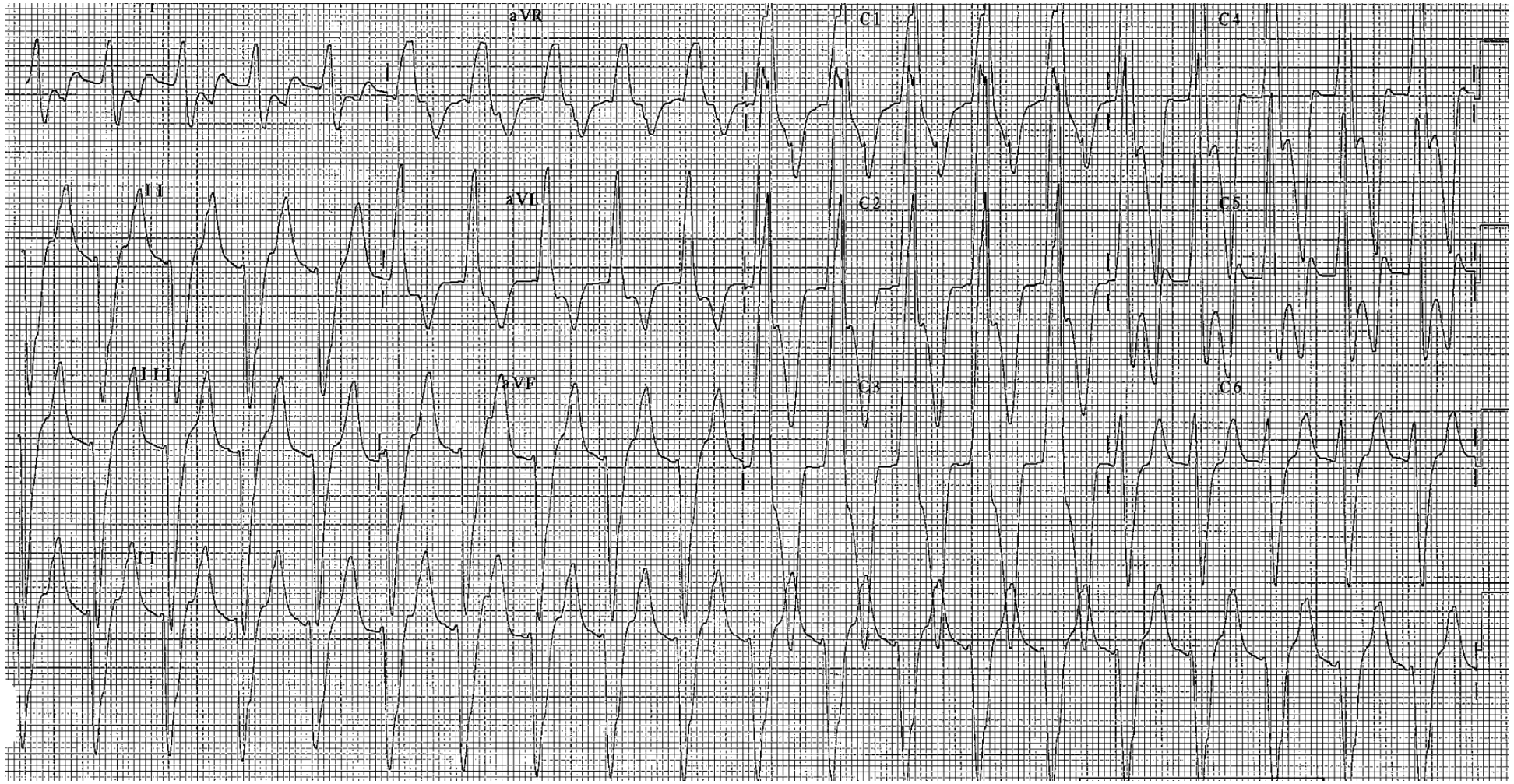

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
P1 ART

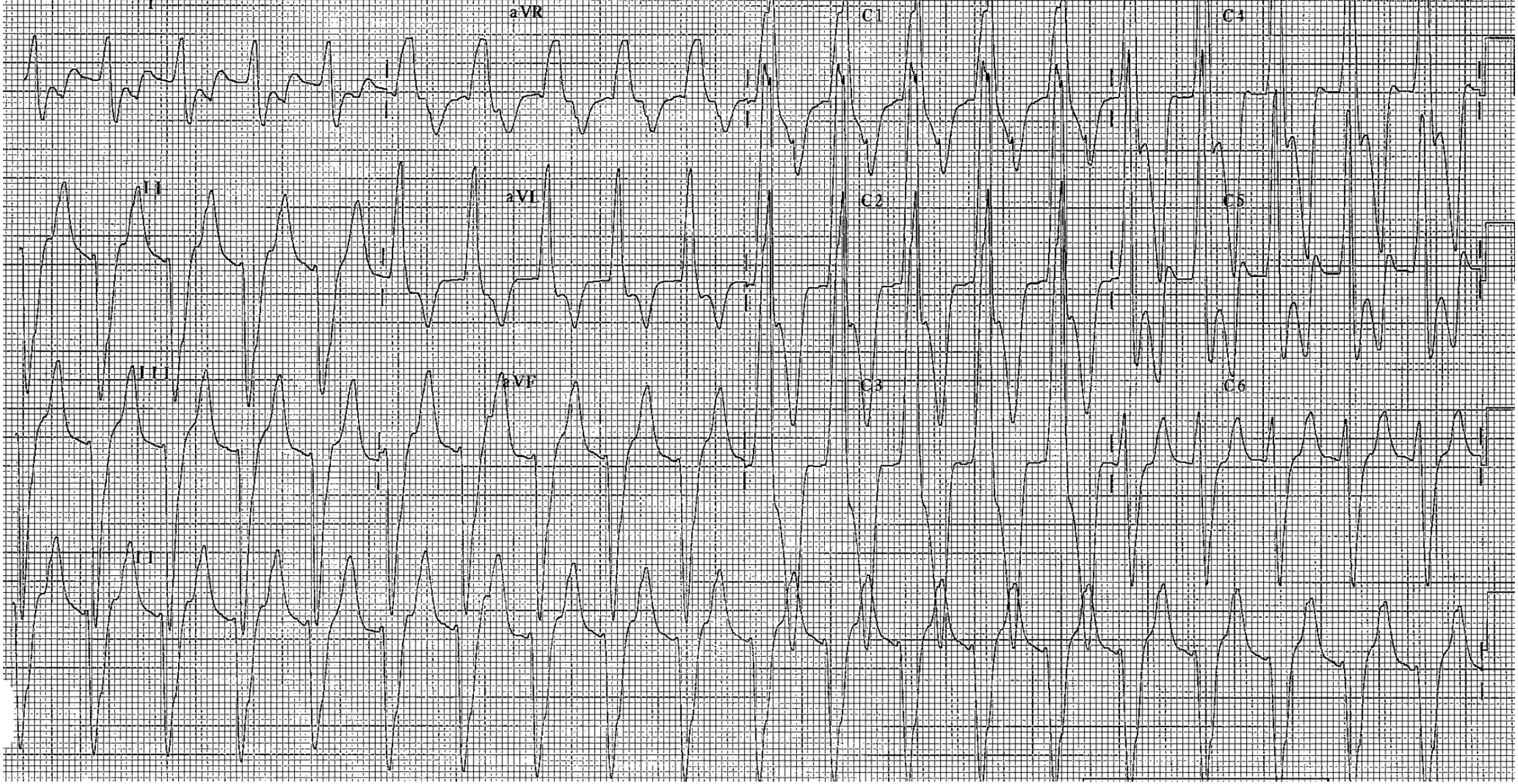

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
P1 ART

Cas n°5

32 ans, APSO non corrigée
Hospitalisé en salle avec télémétrie

Diagnostic ?



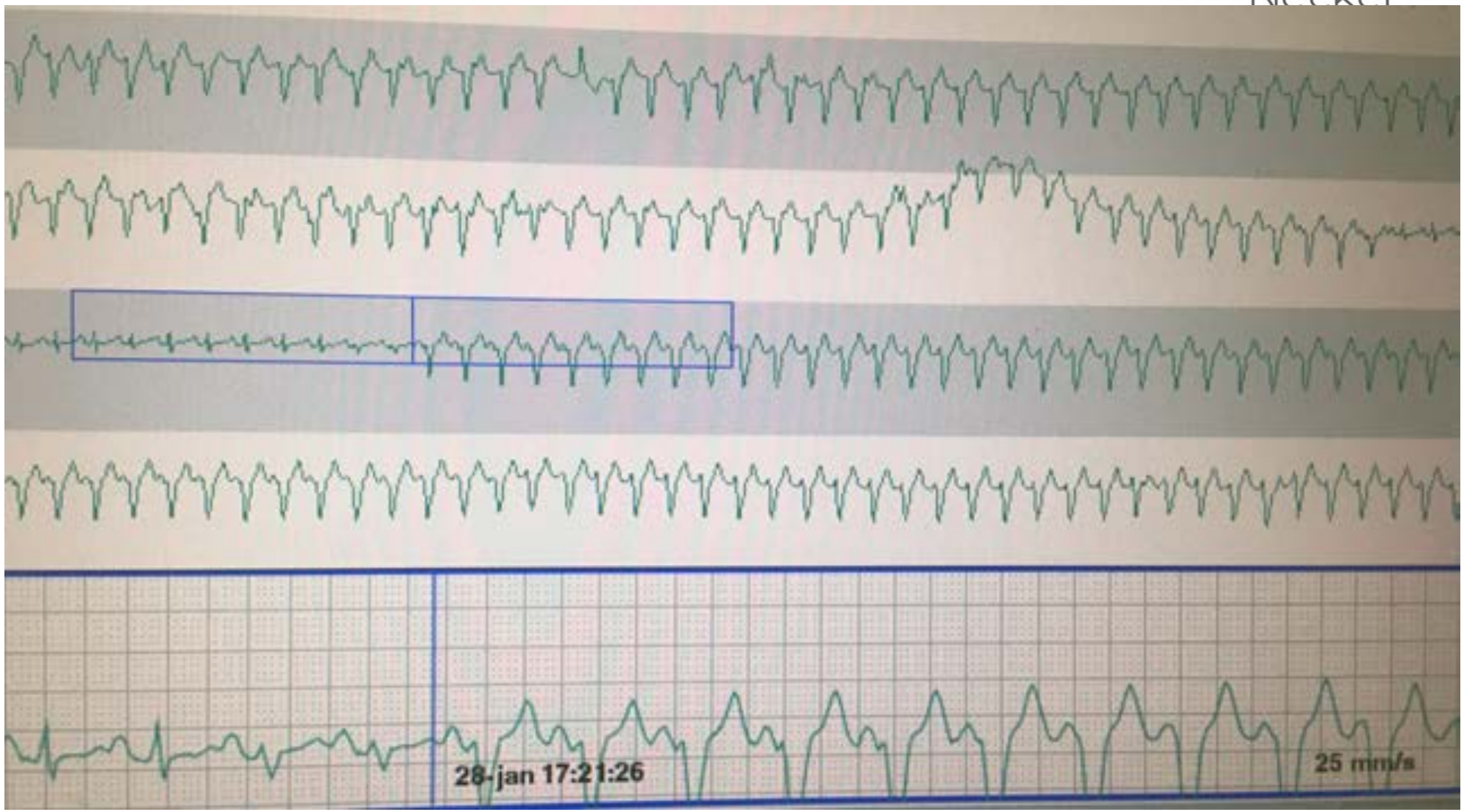

I
aVR
C1
C4
II
aVL
C2
C5
III
aVF
C3
C6

Cas n°5

32 ans, APSO non corrigée
Hospitalisé en salle avec télémétrie

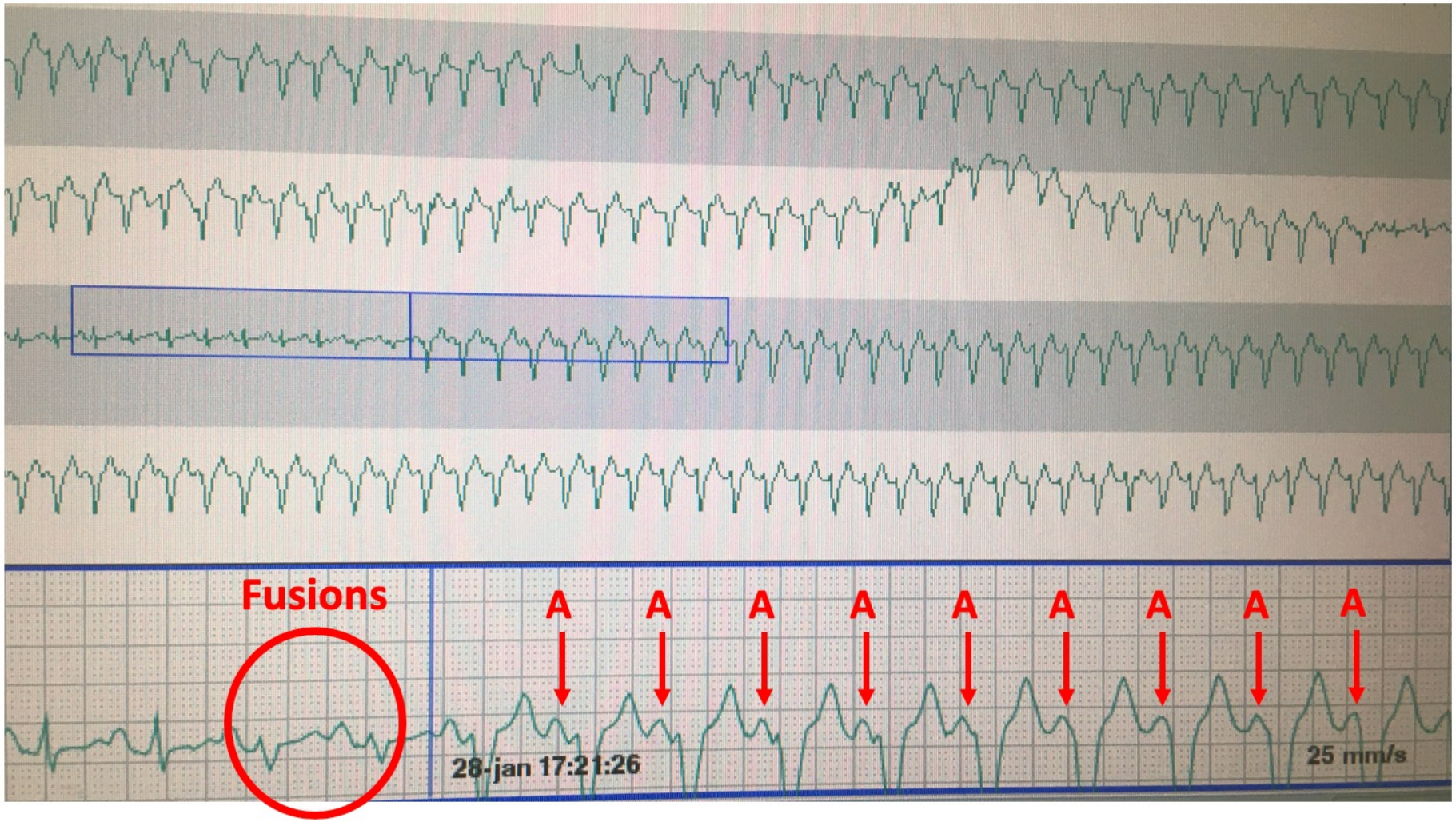
Regarder scope
En détail ++

Diagnostic ?



Cas n°5

32 ans, APSO non corrigée
Hospitalisé en salle avec télémétrie

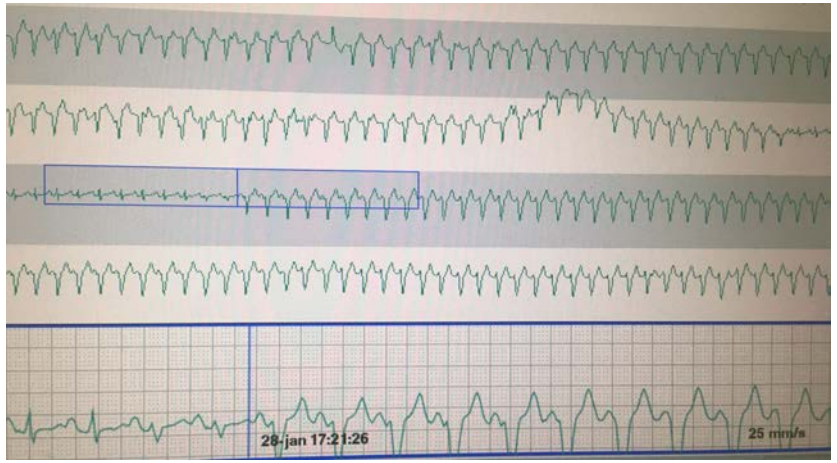


Dissociation VA avec V un peu plus rapide que A = TV

Regarder le scope +++

Début/fin tachy ++

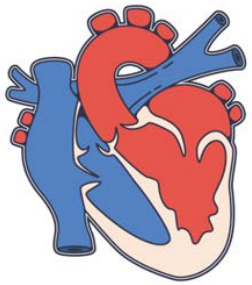
Débute sur un A ? Un V ?
Saut de conduction ?
S'arrête sur un A ?
Warmup/cool down
…



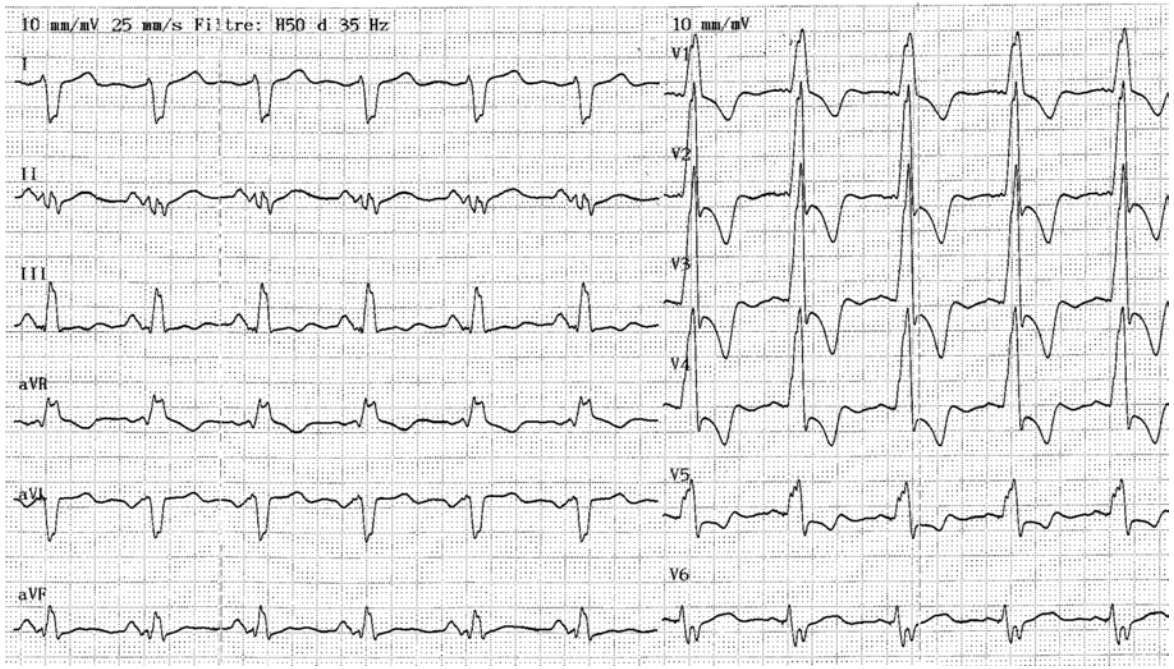
Recherche dissociation AV ++

V>A = TV ou tachy Hissienne
Conduction variable TA/Flutter ?

Case n°6

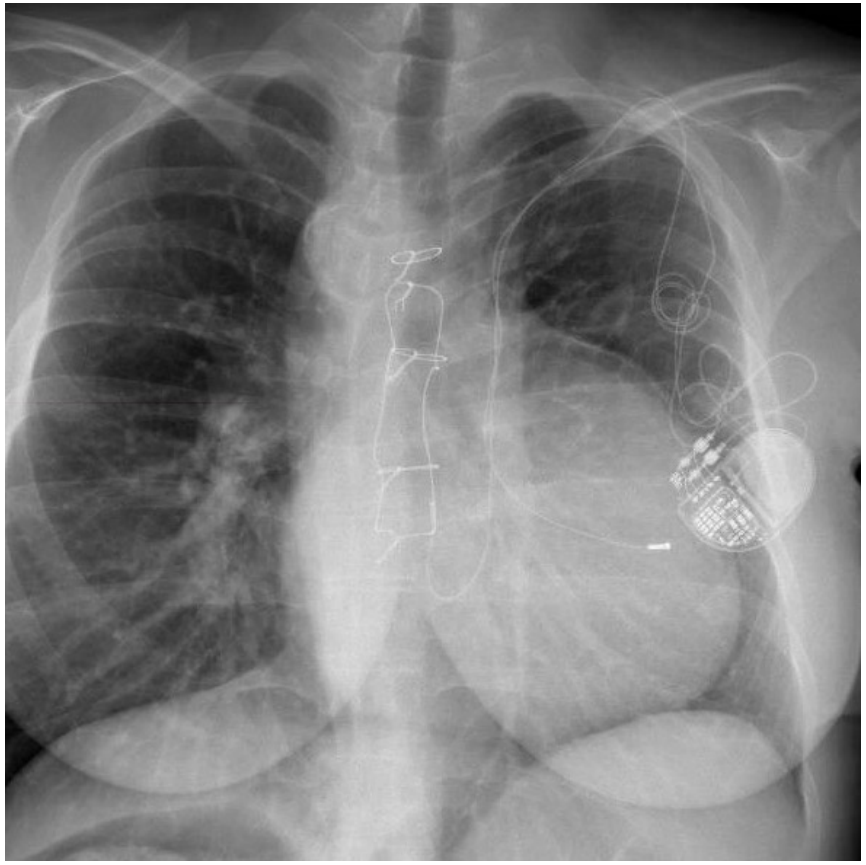


Woman, 44 y
Situs inversus, cc-TGA, levocardia
Endovenous pacemaker for complete AV block (QRS 180 ms)
Right systemic ventricular severe dysfunction
NSVT
NYHA 3
Optimal pharmacological therapy



→ **What do you propose ?**

CASE N°6



- **Anterior systemic RV**
- **No exploitable coronary sinus**

Resynchronization in CHD

Specific features

- **RBBB > LBBB**
- **Systemic RV dysfunction**
- **Subpulmonary RV dysfunction**
- **Univentricular heart dysfunction**

Circulation: Arrhythmia and Electrophysiology

SPECIAL REPORT

Cardiac Resynchronization Therapy for Treatment of Chronic Subpulmonary Right Ventricular Dysfunction in Congenital Heart Disease

Jan Janoušek, MD, PhD
Jan Kovanda, MD
Miroslav Ložek, MSc
Viktor Tomek, MD, PhD
Roman Gebauer, MD
Peter Kubuš, MD, PhD
Tammo Delhaas, MD, PhD

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Europace (2017) 0, 1–8
doi:10.1093/europace/euw386

CLINICAL RESEARCH

Cardiac resynchronization therapy in adults with congenital heart disease

Zeliha Koyak[1,2], Joris R. de Groot[1], Ahmed Krimly[3], Tara M. Mackay[1], Berto J. Bouma[1], Candice K. Silversides[3], Erwin N. Oechslin[3], Ulas Hoke[4], Lieselot van Erven[4], Werner Budts[5], Isabelle C. Van Gelder[6], Barbara J. M. Mulder[1,2]*, and Louise Harris[3]

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EXPRESS PUBLICATION

Cardiac Resynchronization Therap... A Novel Adjunct to the Treatmen... Prevention of Systemic Right Ve...

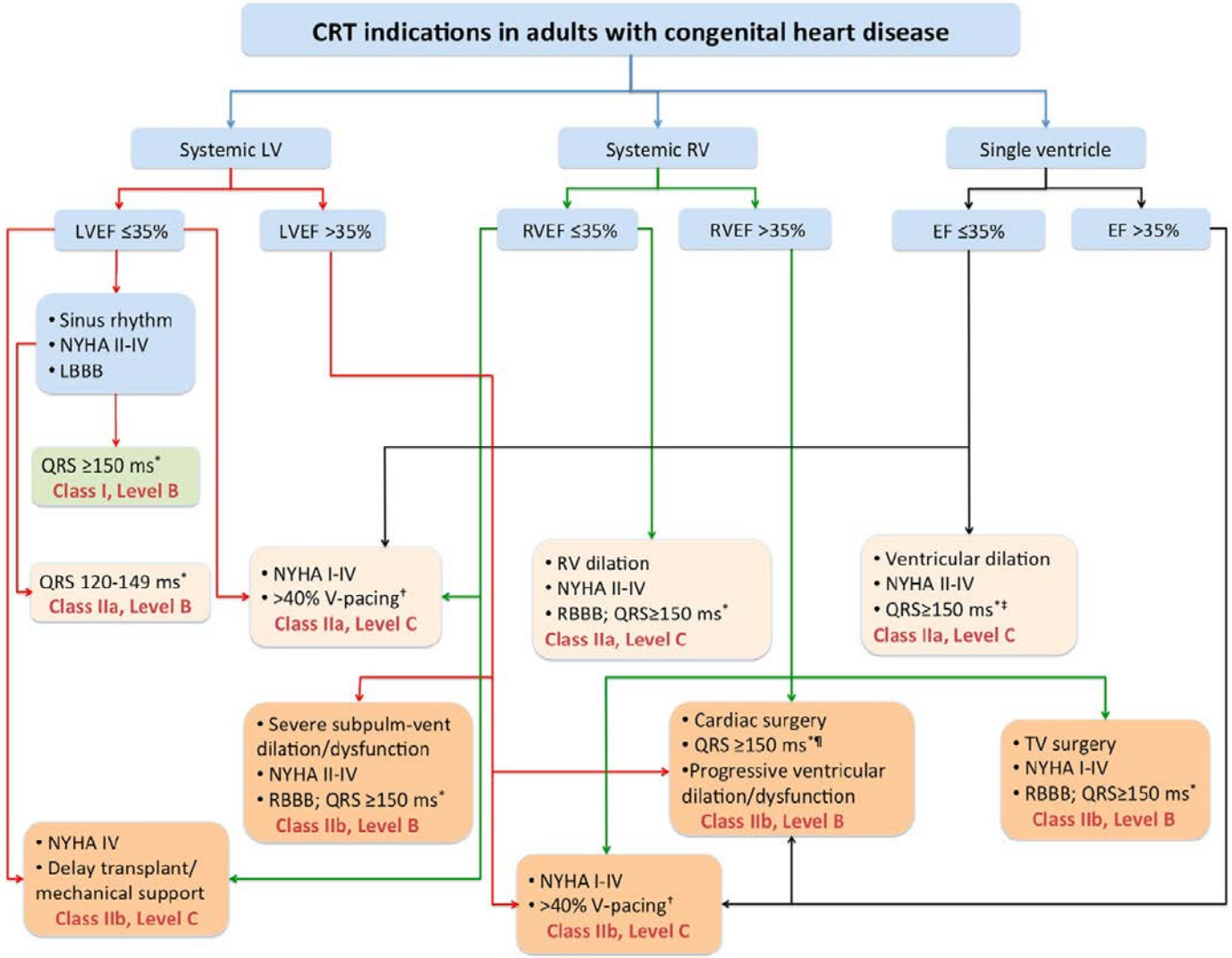
Jan Janoušek, MD,* Viktor Tomek, MD,* Václav Chalo... Roman A. Gebauer, MD,* Josef Kautzner, MD, PhD,†...

International Journal of CARDIOLOGY

Cardiac resynchronization therapy in congenital heart disease: Results from the German National Register for Congenital Heart Defects

Ann-Katrin Flügge[a], Kristina Wasmer[b], Stefan Orwat[a], Hashim Abdul-Khaliq[c], Paul C. Helm[d], Ulrike Bauer[d], Helmut Baumgartner[a], Gerhard-Paul Diller[a,*] for the German Competence Network for Congenital Heart Defects Investigators

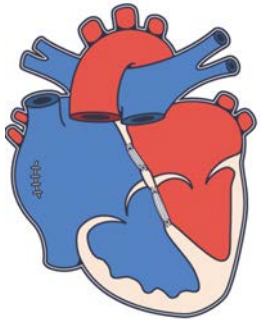
RESYNCHRONIZATION IN CHD



Khairy P, et al. Heart Rhythm 2014

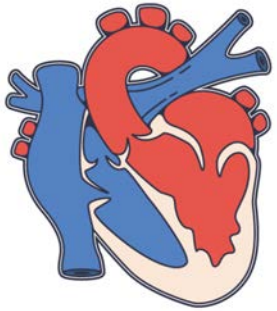
Resynchronization in CHD

Guidelines summary



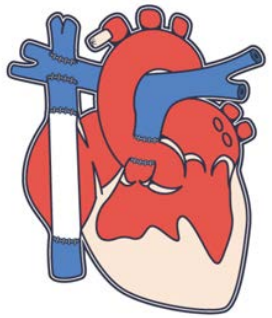
Systemic LV ≤ 35%

LBBB ≥ 120 ms



Systemic RV ≤ 35%

RBBB ≥ 150 ms



Single V ≤ 35%

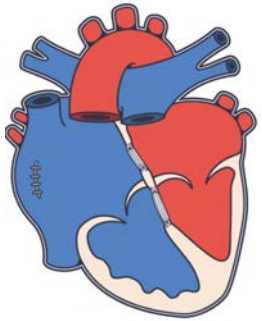
QRS ≥ 150 ms

Or V-pacing > 40%

Khairy P, et al. Heart Rhythm 2014
Hernandez-Madrid A, et al. Europace 2018

Resynchronization in CHD

Guidelines summary

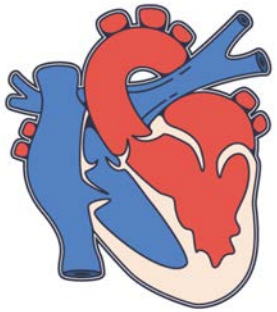


Systemic LV ≤ 35%

LBBB ≥ 120 ms

- Severe subpulm-vent dilation/dysfunction
- NYHA II-IV
- RBBB; QRS ≥150 ms*

Class IIb, Level B



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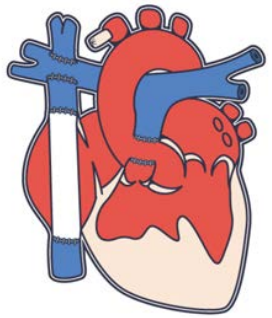

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Khairy P, et al. Heart Rhythm 2014
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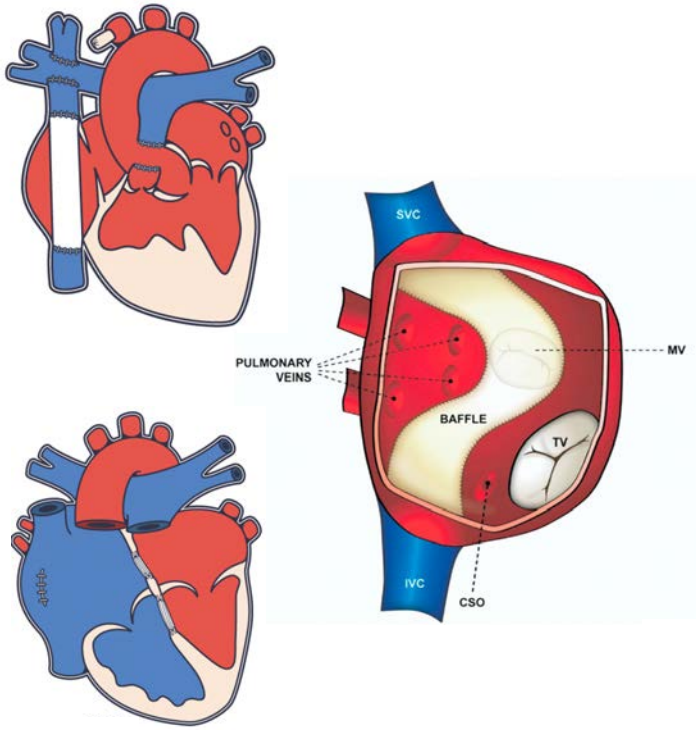
Resynchronization in CHD

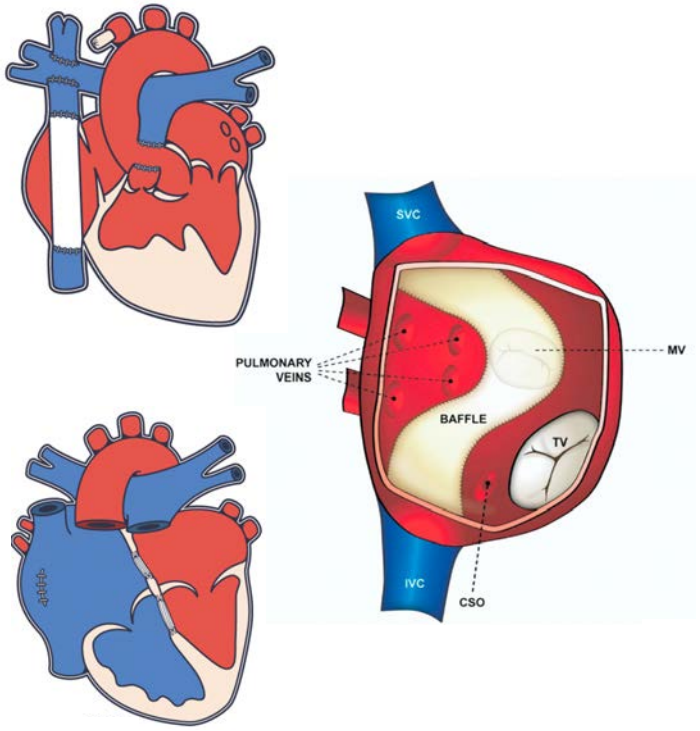
Technical aspects



Coronary sinus may be inaccessible

Resynchronization in CHD

Technical aspects



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FOCUS ISSUE: CARDIAC RESYNCHRONIZATION THERAPY

Congenital Heart Disease and Resynchronization

Resynchronization Therapy in Pediatric and Congenital Heart Disease Patients

An International MultiCenter Study

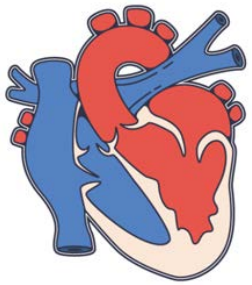
Anne M. Dubin, MD, FACC,* Jan Janousek, MD,† Edward Rhee, MD, FACC,‡

73 patients with CHD

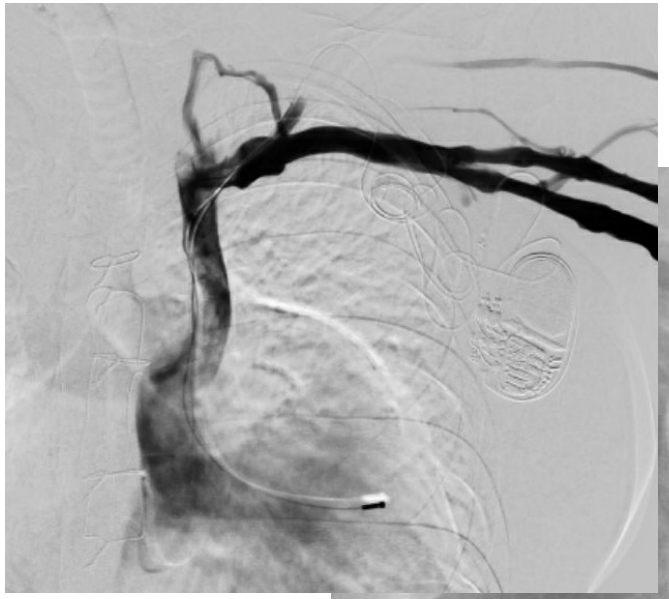
Coronary sinus may be inaccessible

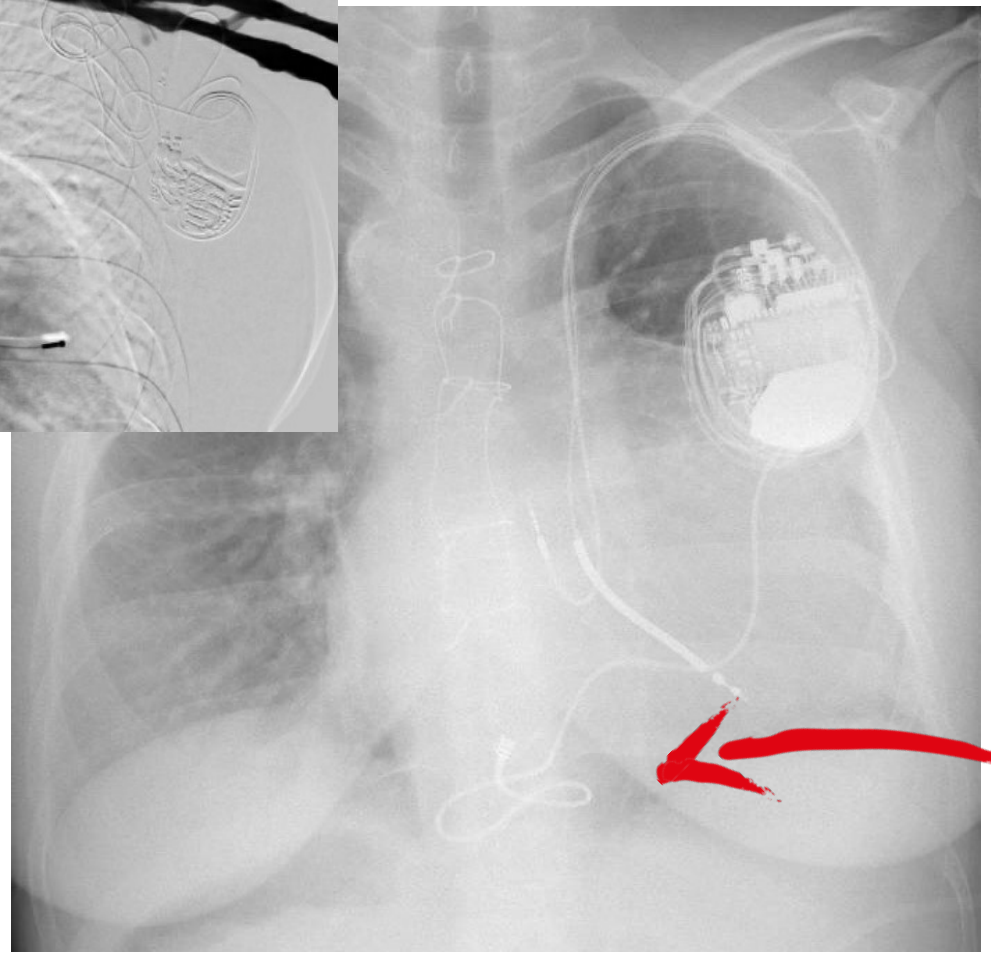
Transvenous System (%)	Epicardial/Mixed System (%/%)
26 (36%)	37/10 (51%/14%)

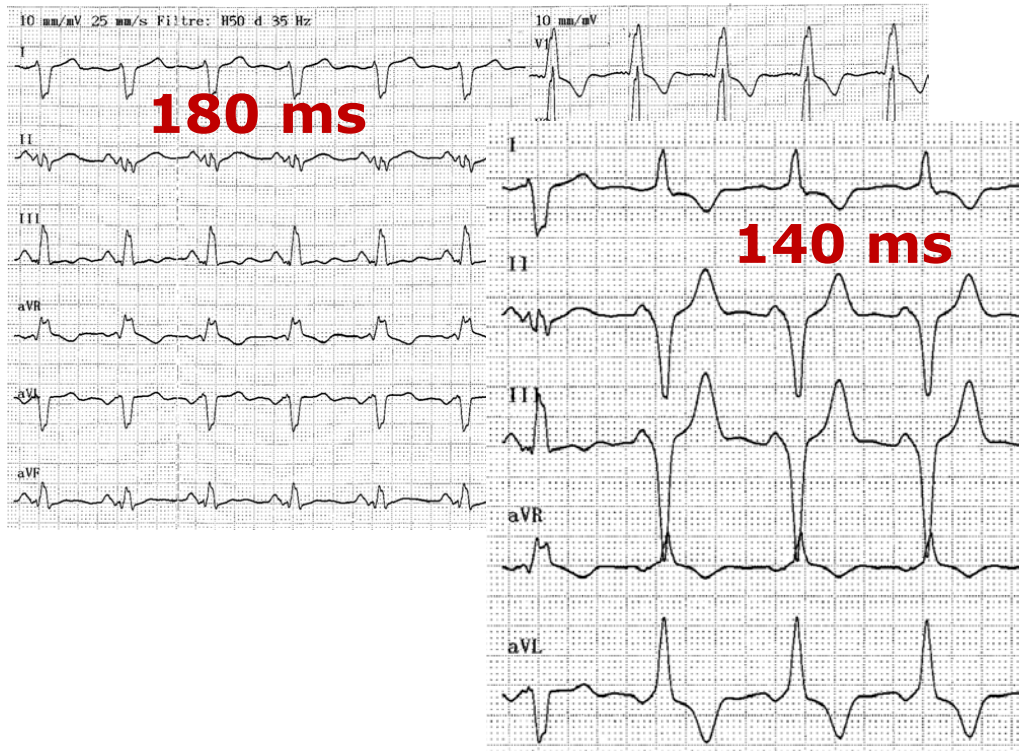
Case n°6



Hybrid approach

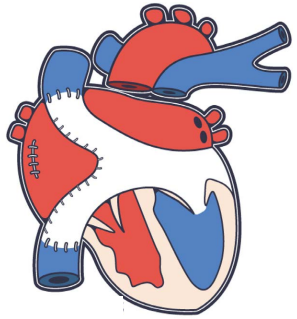




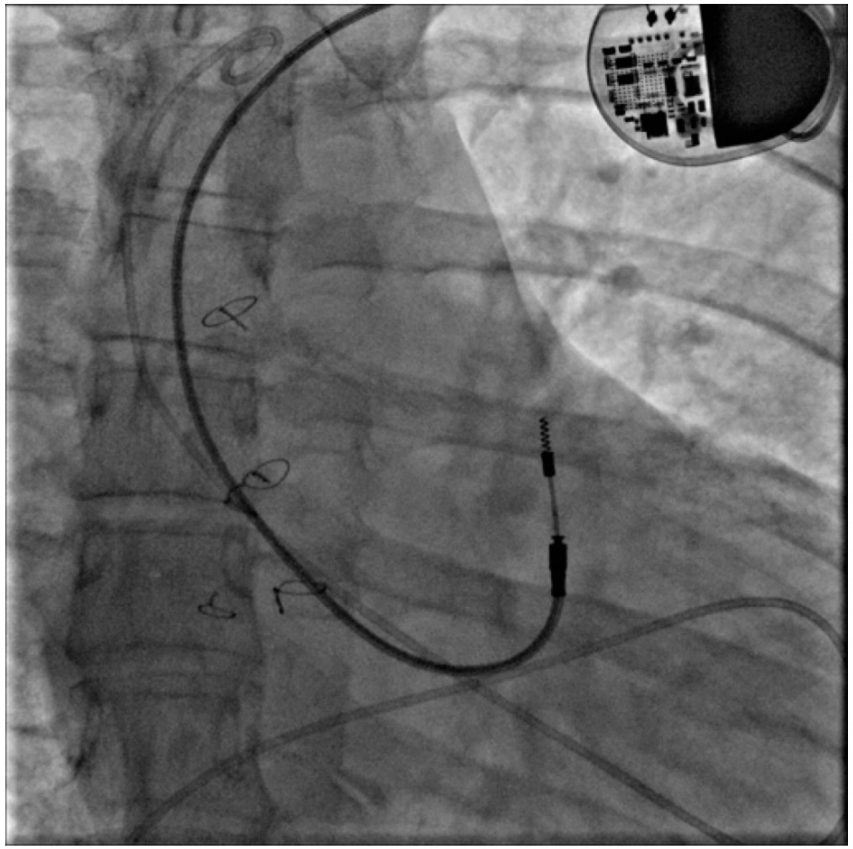


Epicardial « left » tunneled lead

Case n°7

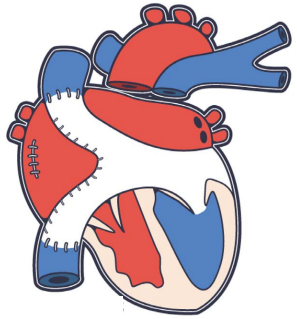


Man, 47 y, Mustard
Endovenous pacemaker for sinus node dysfunction
Hospitalization for sustained VT



→ **What to check before upgrade for ICD?**

Case n°7



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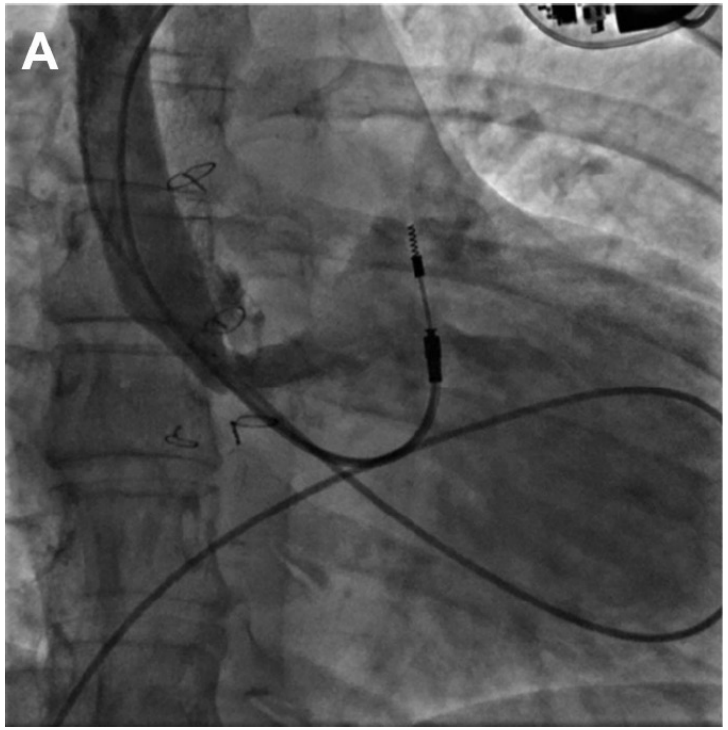
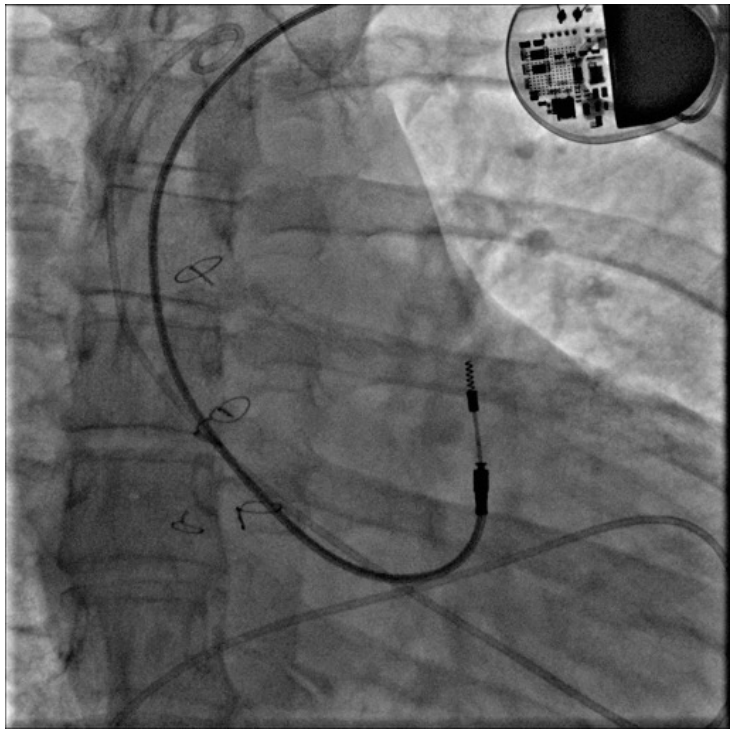
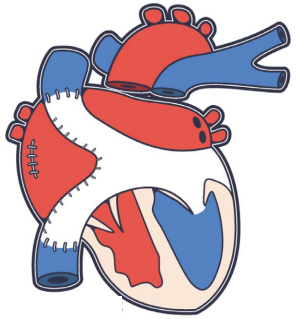
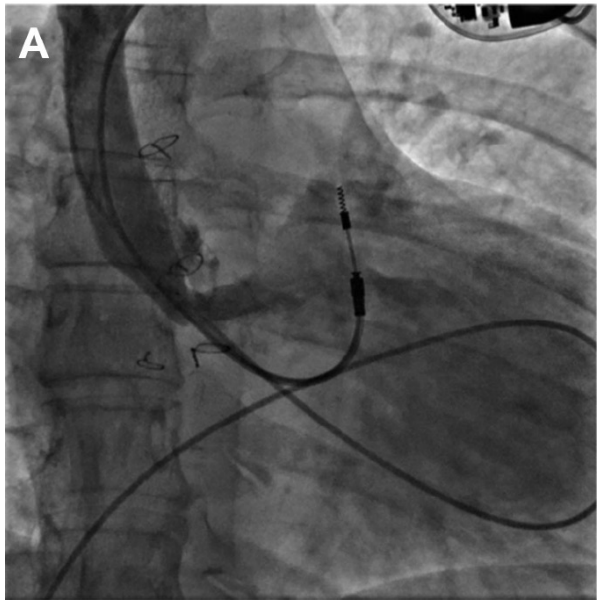


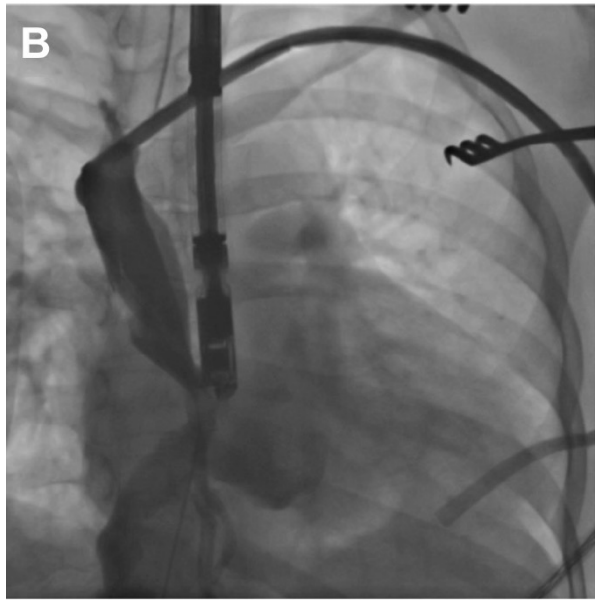


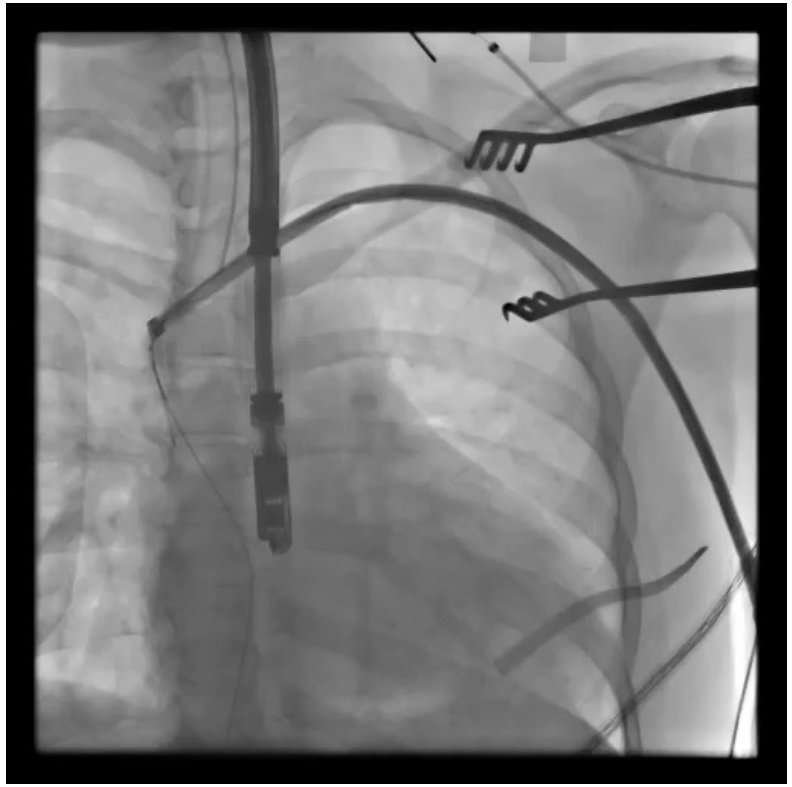
Laredo M, Waldmann V, et al. JACC EP 2019

Case n°7









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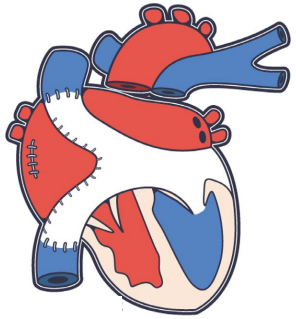
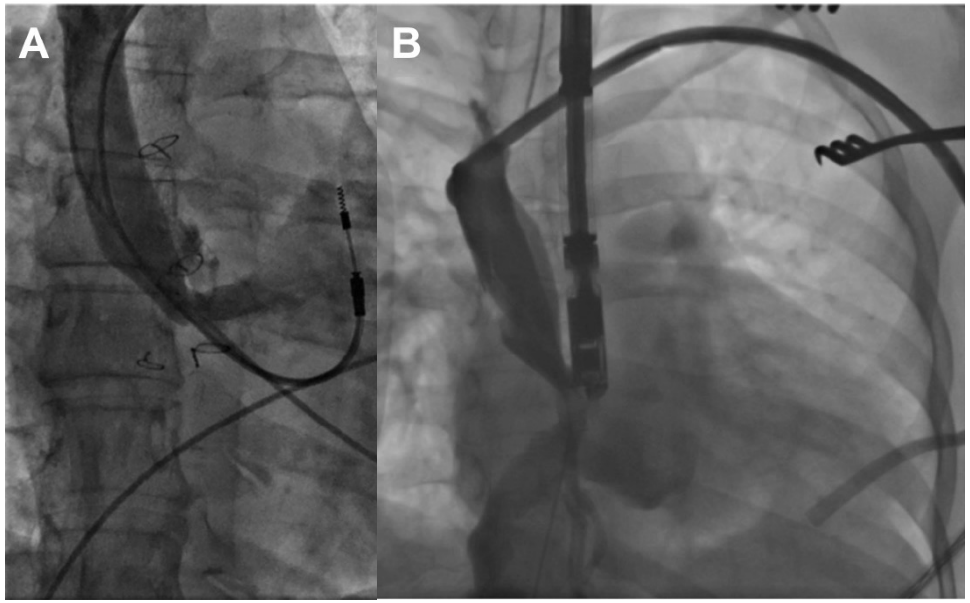
Case n°7





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Take home Messages

- Arrhythmias in CHD = 1st cause of urgent admission
- Poor tolerance: low threshold for hospitalization
- Complete/hemodynamic evaluation
- Ablation >> pharmacological therapy
- Need to improve selection of candidates for ICD / CRT
- Expert centers for collegial discussion
- Consider electrophysiological study before surgery (TOF, Ebstein)

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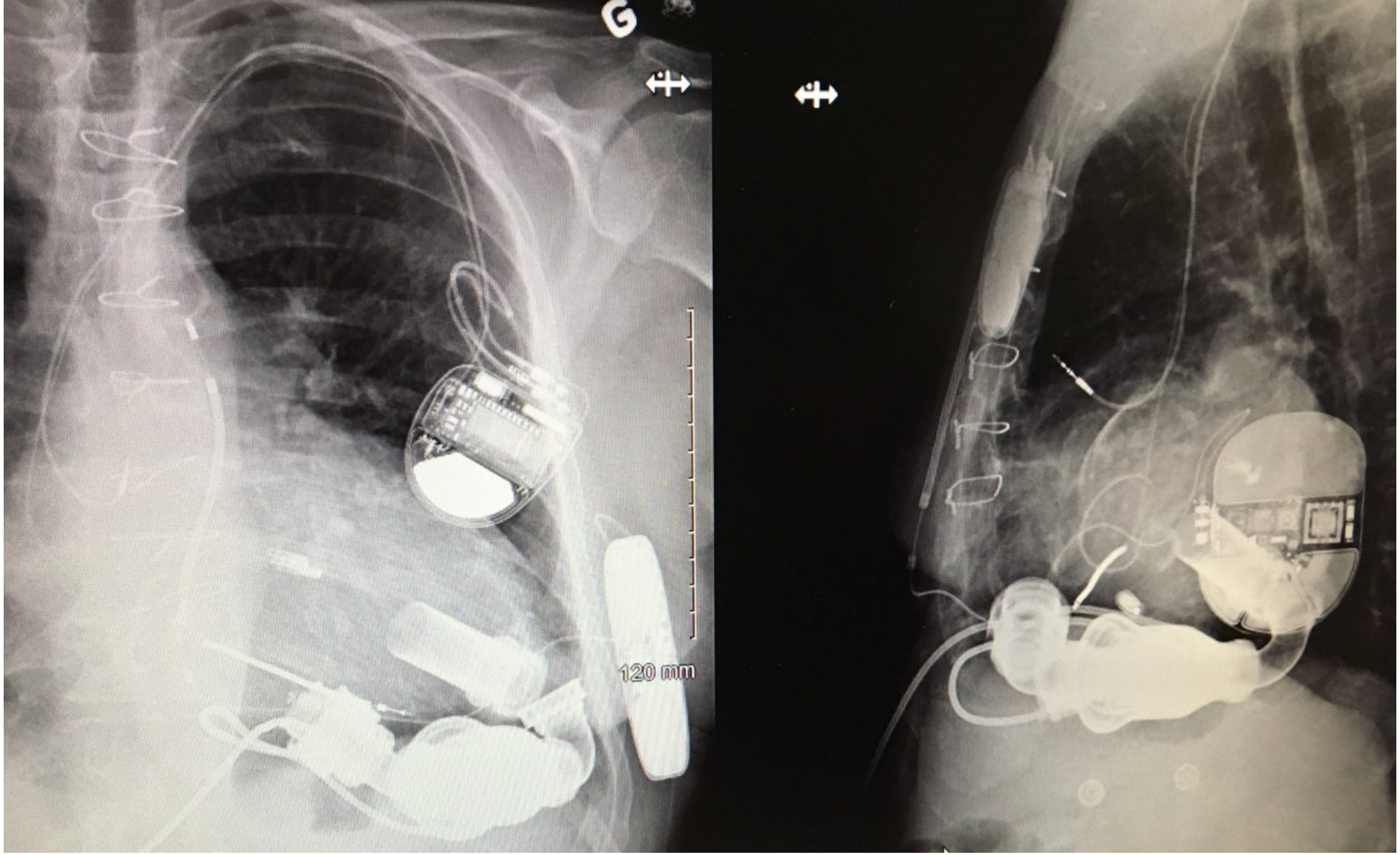
01 56 09 37 84



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