Tétralogie de Fallot, risque rythmique, pratique de l'activité physique et du sport

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Centre Universitaire de Rythmologie de Lyon

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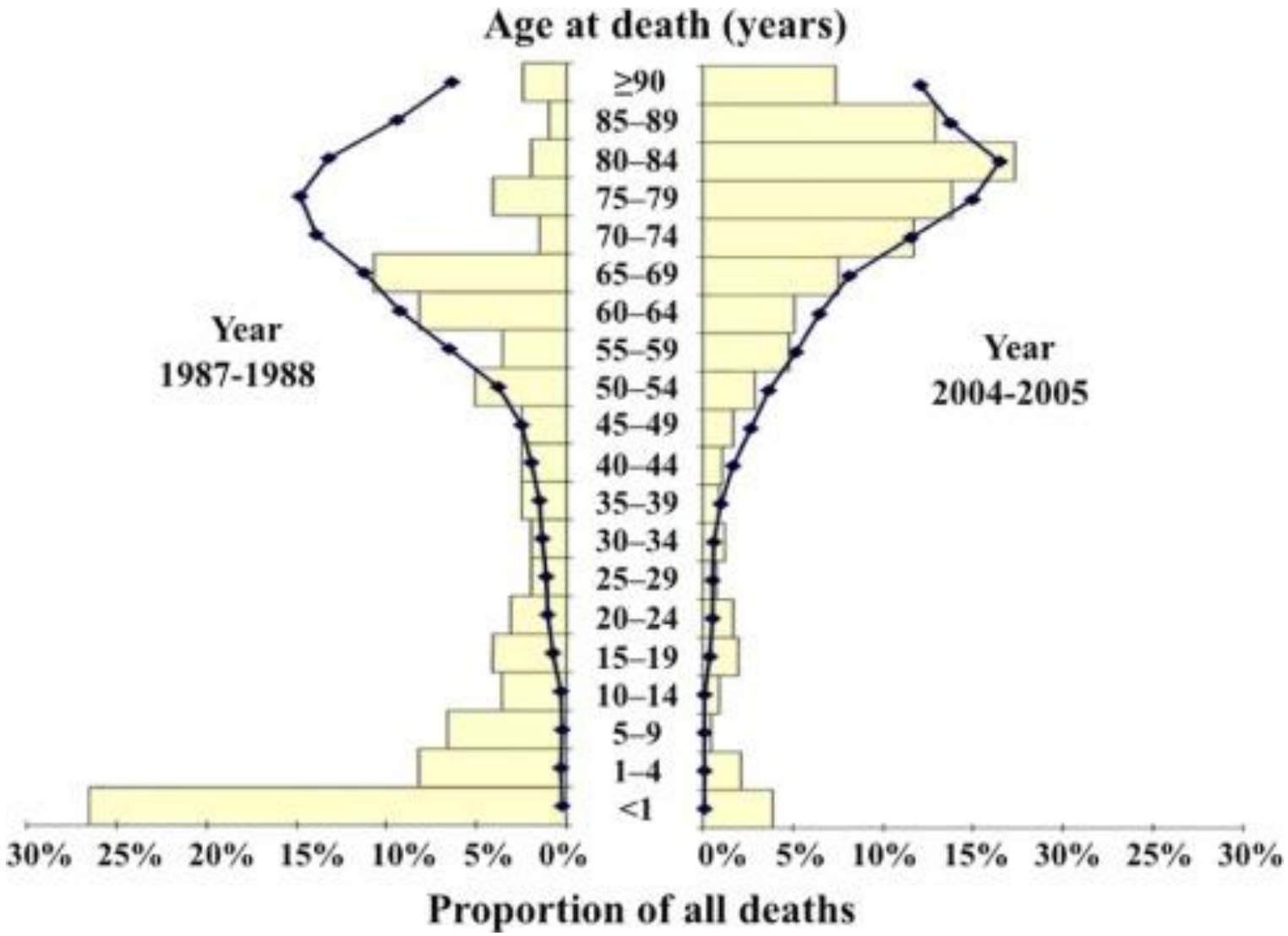
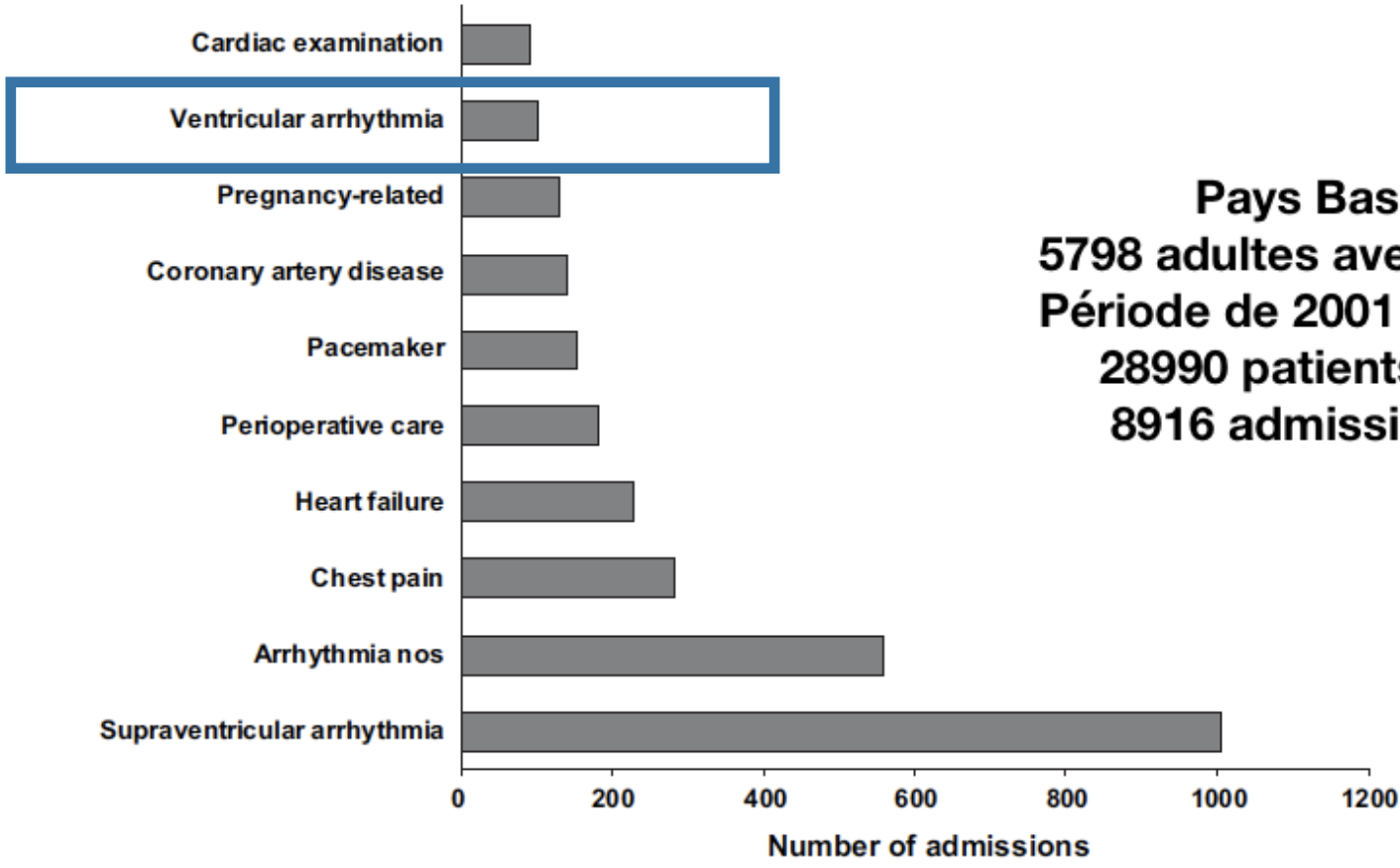
Epidemiology



Khairy JACC 2010

Epidemiology

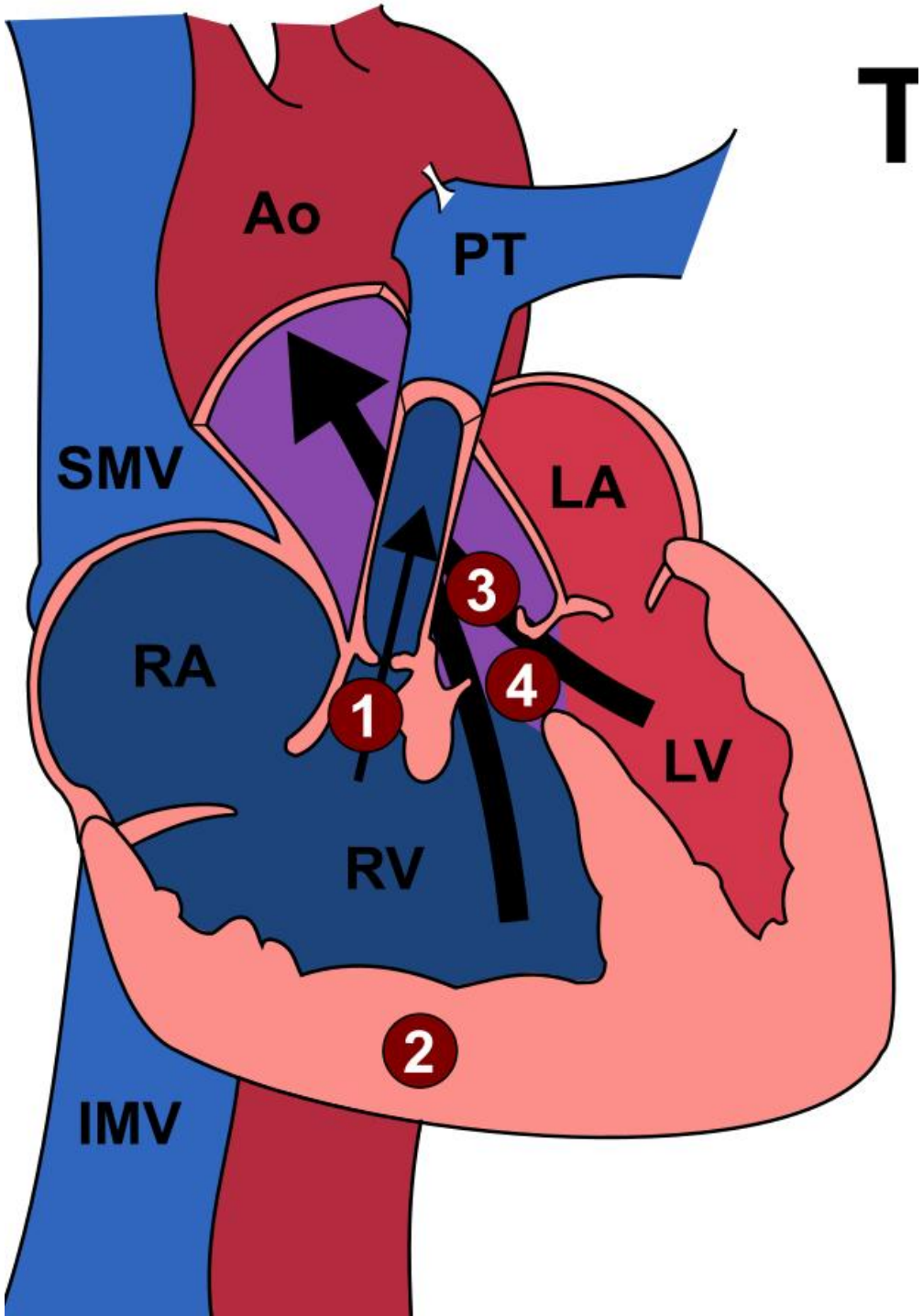


Pays Bas
5798 adultes avec CHD
Période de 2001 à 2006
28990 patients/an
8916 admissions

Verheugt L et al. Heart 2010;96:872-8

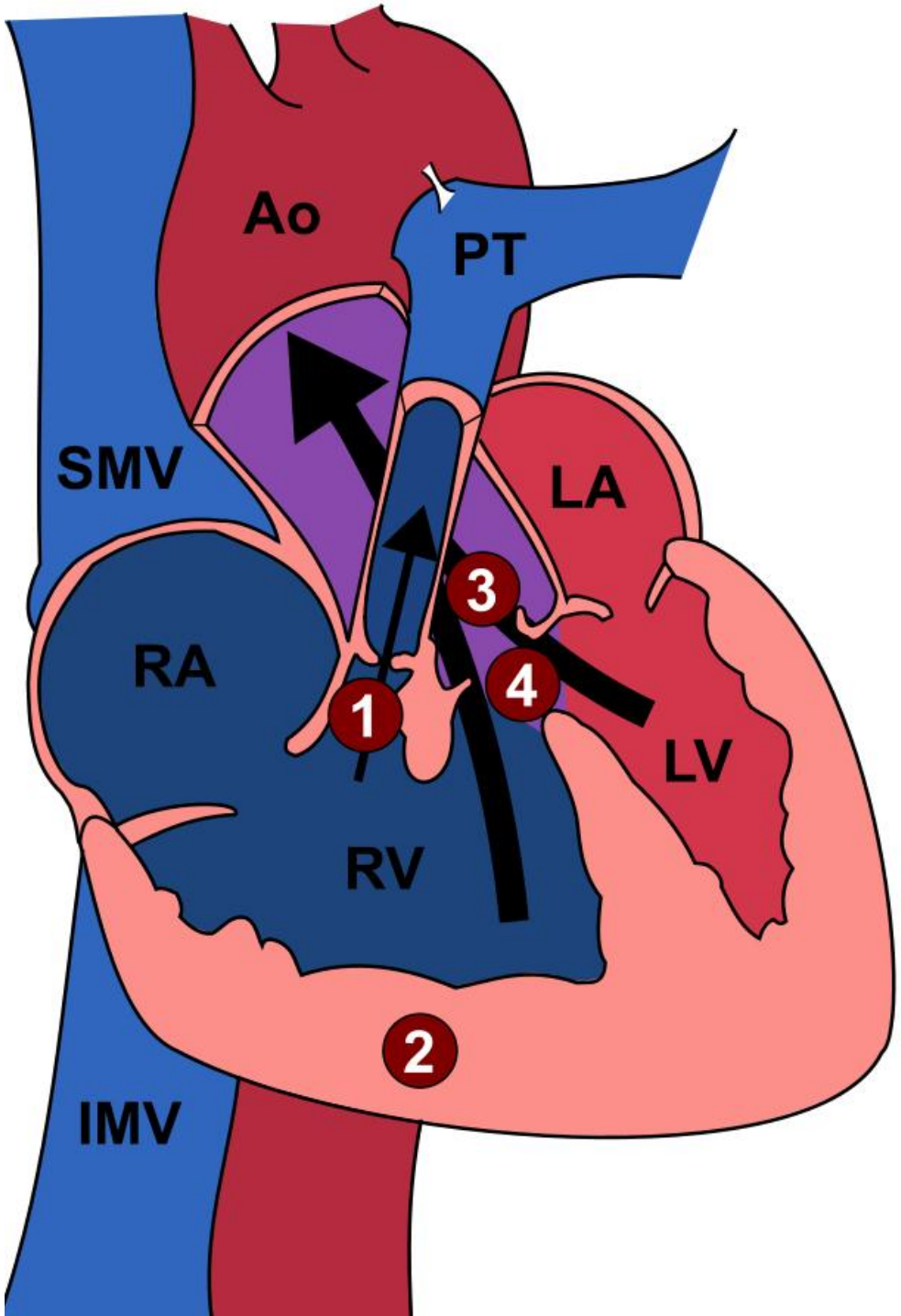
TOF

- 4.5% CHD
- 1/2800 living birth
- Conotruncal anomaly



TOF

- Definition
  - VSD
  - PVS
  - Dextro Ao
  - RV dilatation
- SCD rate : 0.15%/an (x200 pop g)
- Rate of VT: 14% after 30yo F.U
- Excepted surgeries
  - Palliative shunt
  - Corrective surgery
  - PVR



TOF

- SCD risk score

- + positive LGE MRI
- + Atrial arrhythmias

TABLE 1 Risk Score for Appropriate ICD Shocks in Patients With Tetralogy of Fallot

	Exp (B)	Point Attributed
Prior palliative shunt	3.2	2
Inducible sustained ventricular tachycardia	2.6	2
QRS >180 ms	1.4	1
Ventriculotomy incision	3.4	2
Nonsustained ventricular tachycardia	3.7	2
Left ventricular end-diastolic pressure ≥12 mm Hg	4.9	3
TOTAL POINTS		0–12

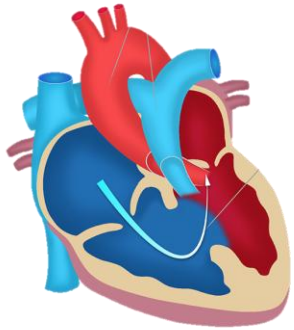
Adapted with permission from Wolters Kluwer Health Inc Khairy et al. (11).

Exp(β) = exponential of the beta-coefficient; ICD = implantable cardioverter-defibrillator.

Khairy et al. Circ 2007

VT ablation in ACHD: Lyon experience

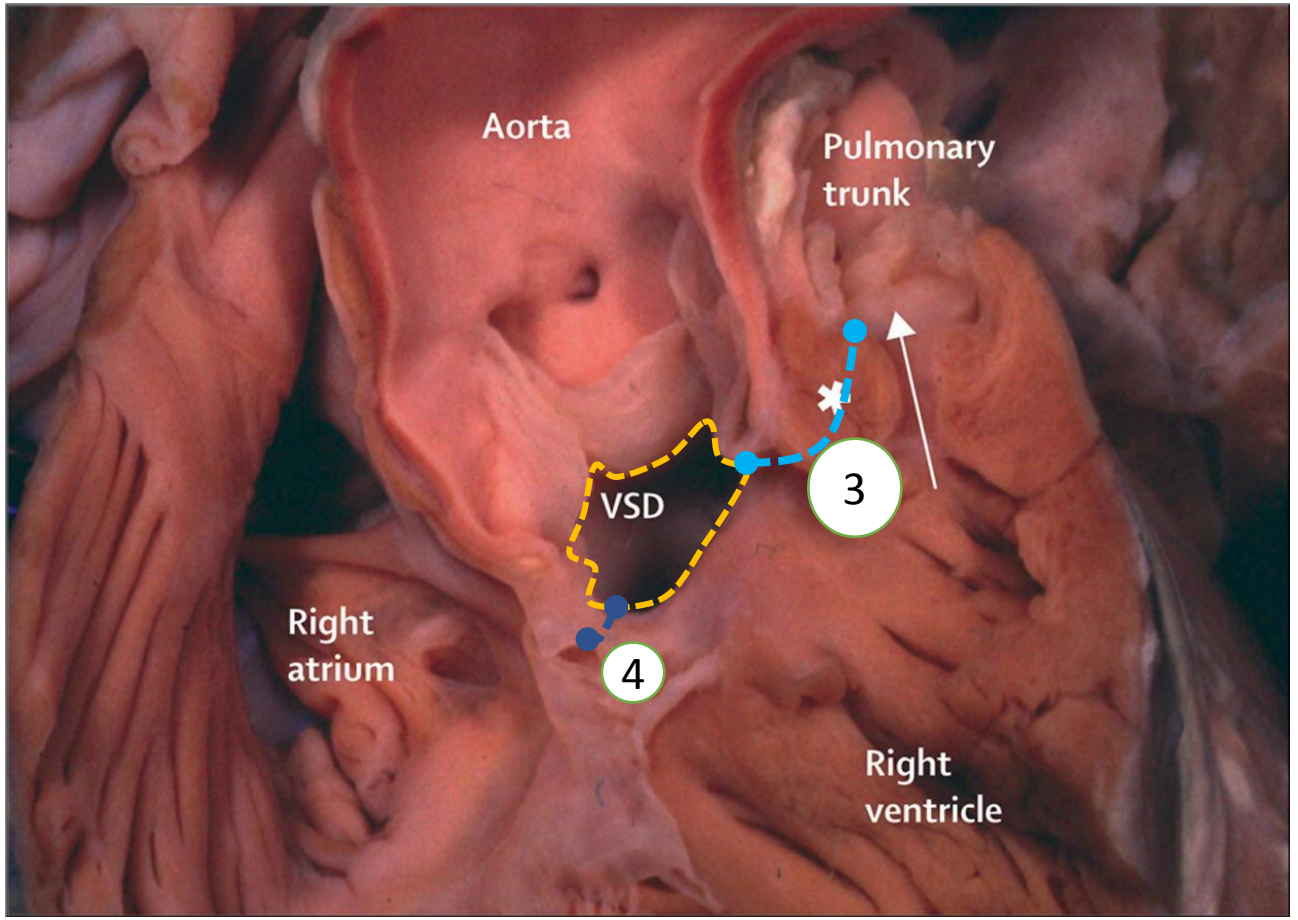
- From January 2020 to Dec 2022
- **34** VT CA in CHD / **31** patients
- including
  - 27 TOF
  - 1 DORV (Fallot like)
  - 1 DOLV
  - 2 VSD + D-TGA (arterial switch repair)



Common point?

- **VSD**
- In TOF or equivalent : **outlet**

- Creating 2 potential critical isthmus
  - **VSD to PV (3)**
  - **VSD to TV (4)**



Apitz et al. The Lancet, 2009

Ventricular arrhythmias in CHD

Type of CHD	Supraventricular arrhythmias			Ventricular arrythmias and SCD		Bradycardia			
						SND		AV block	
	AVRT	IART/ EAT	AF	Sustained VT	SCD	Congenital	Acquired	Congenital	Acquired
Secundum ASD		++	++			(+)	+		(+)
Superior sinus venosus defect		++	+				+		
AVSD/primum ASD		++	++	(+)		(+)		(+)	++
VSD		+	(+)	+	(+)[a]				+
Ebstein anomaly	+++	++	+	(+)	++[b]		++		
TOF		++	++	++	++		+		+
TGA									
Atrial switch		+++	+	++[c]	++[b]		+++		+
Arterial switch		+		+[c]	(+)		(+)		
ccTGA	++	+	+	(+)	++[b]			+	++
Fontan operation									
Atriopulmonary connection		+++	++		+[b]		++		
Intracardiac lateral tunnel		++	+		+[b]		++		
Extracardiac conduit		+	+		+[b]		+		
Eisenmenger physiology Incompletely palliated CHD		++	++		++[d]				

Empty cells indicate that although not specifically indicated, arrhythmic events may occur (no symbol).

(+) = minimal risk + = mild risk ++ = moderate risk +++ = high risk

©ESC

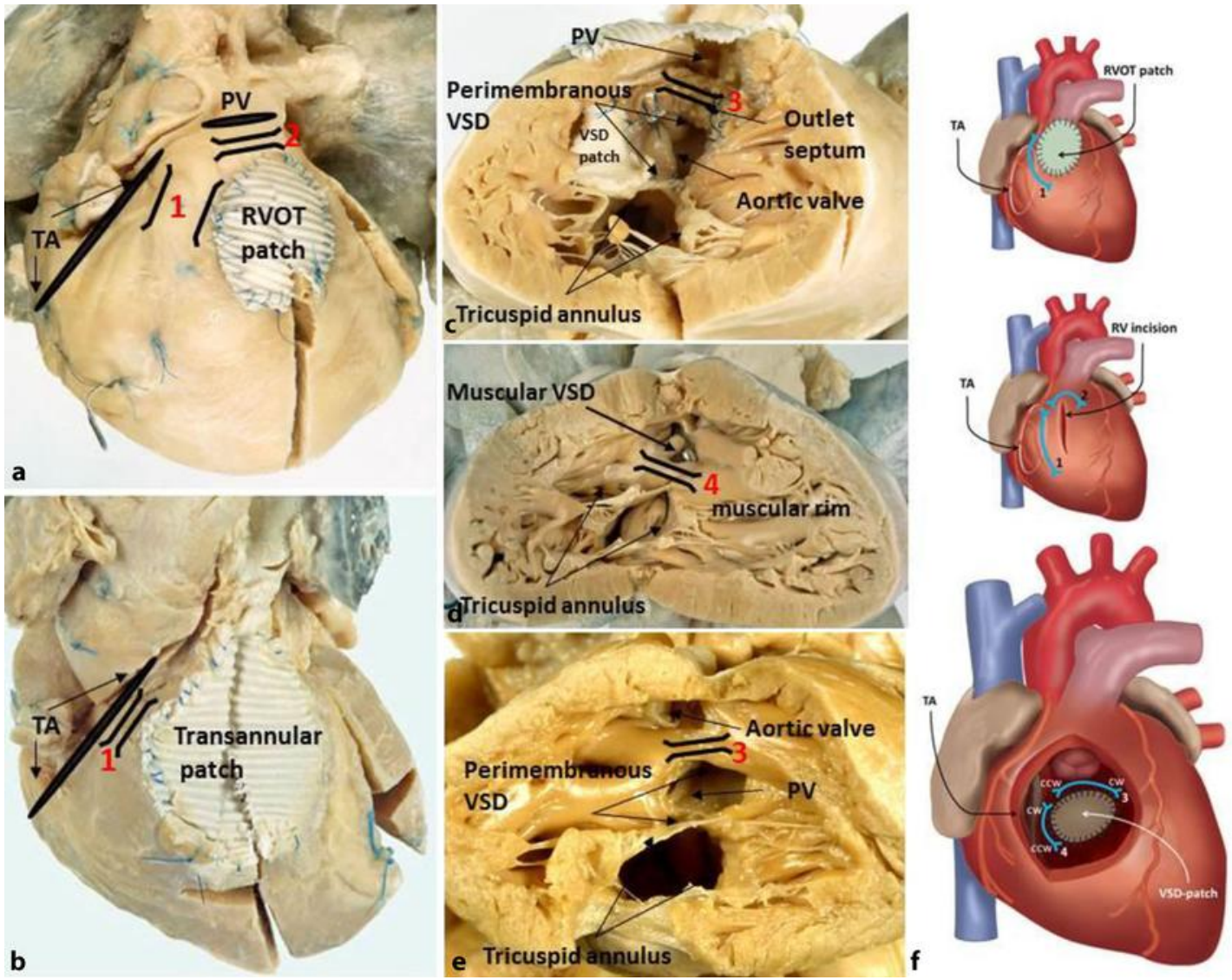
Monomorphic VT
Isthmus related

Polymorphic VT
Ischemic related

Eligible to ablation

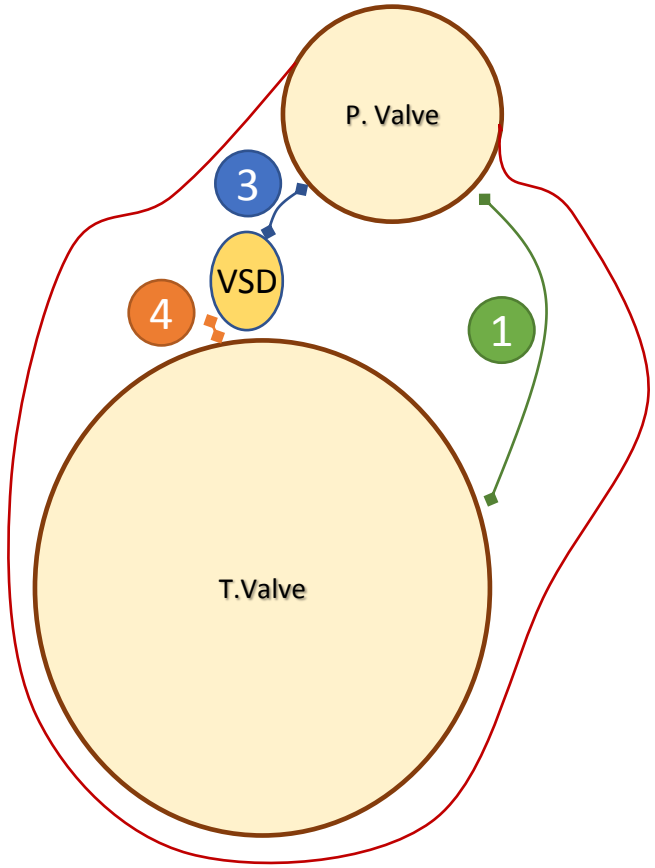
ESC
European Society of Cardiology

Zeppenfeldt classification (Circ. 2007)

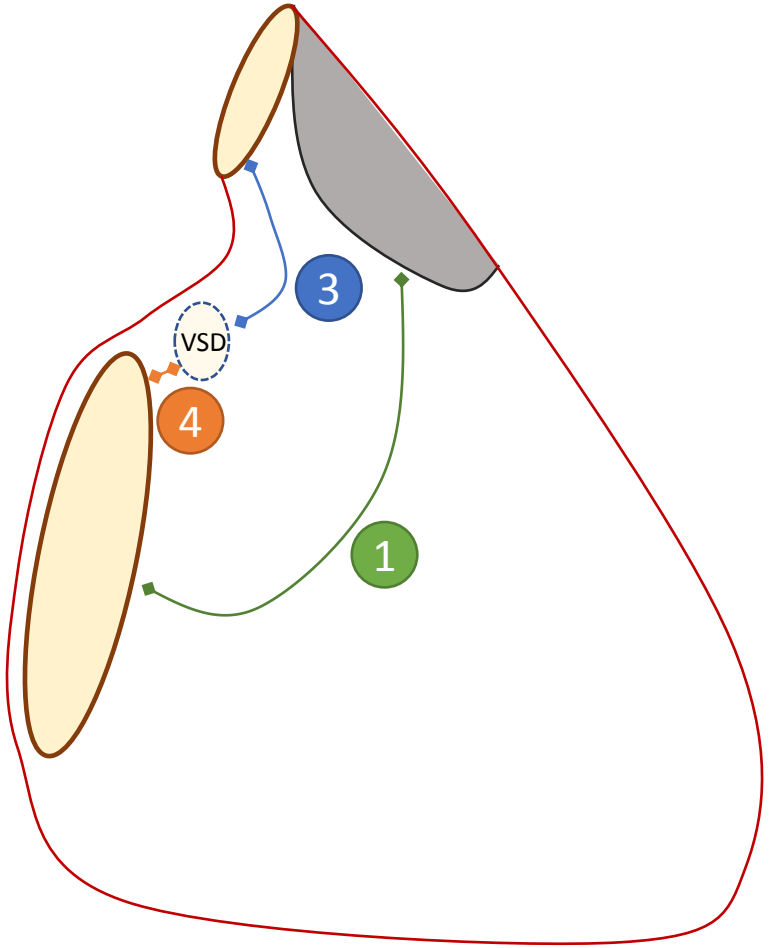


Zeppenfeld, Heart 2018, PMID 29305559

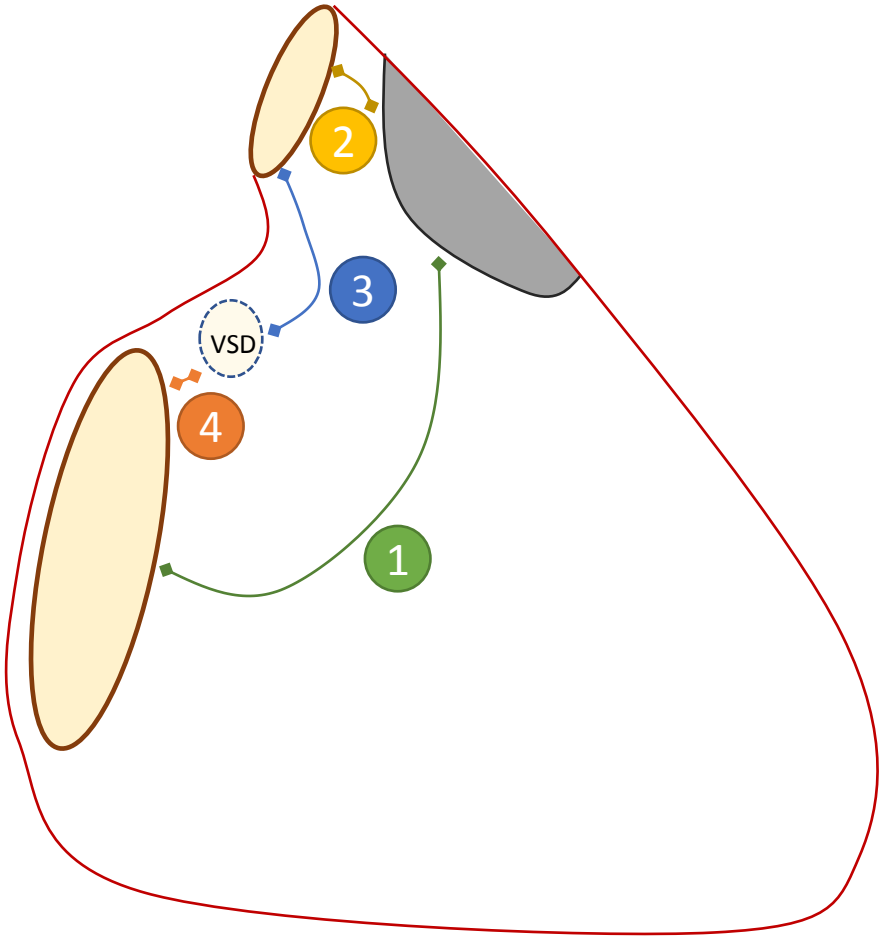
Different possible circuits…



PA view



RAO view
(Transannular patch)



RAO view
(RVOT patch)

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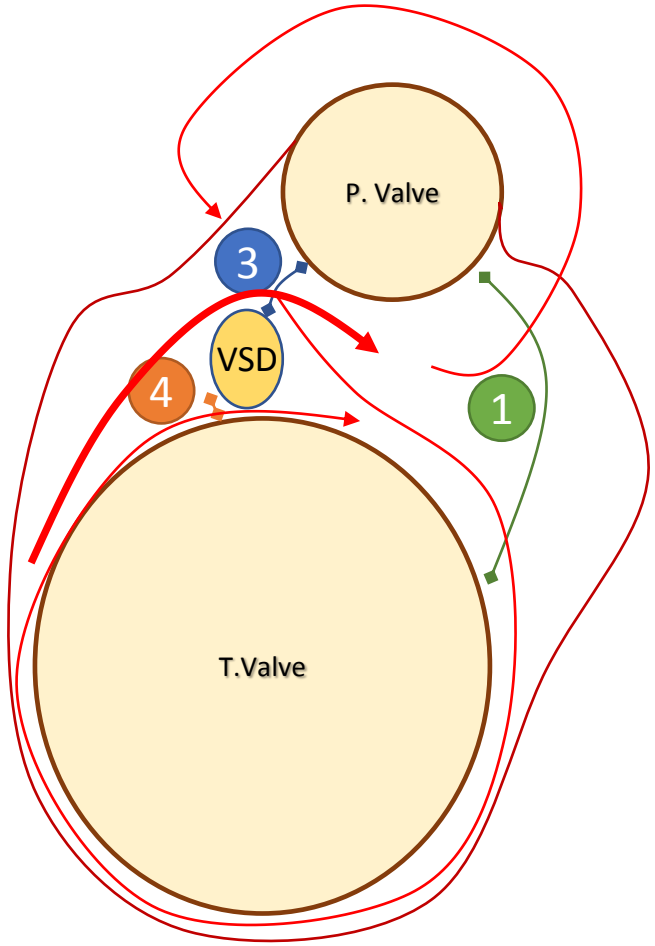

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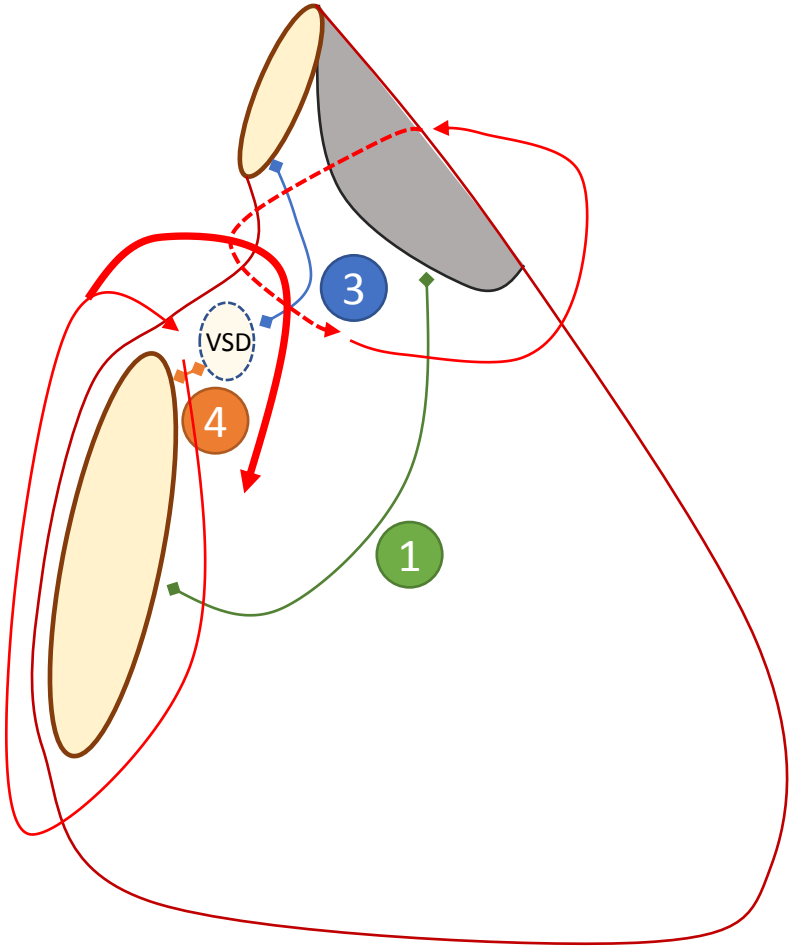

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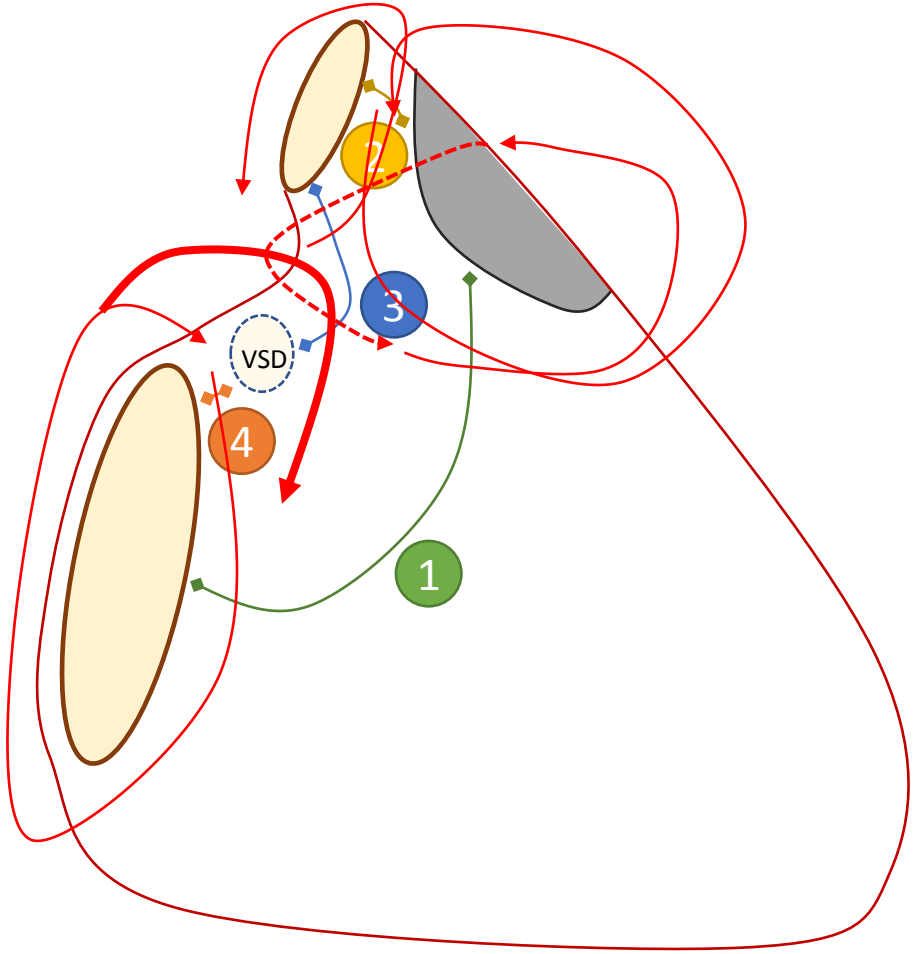

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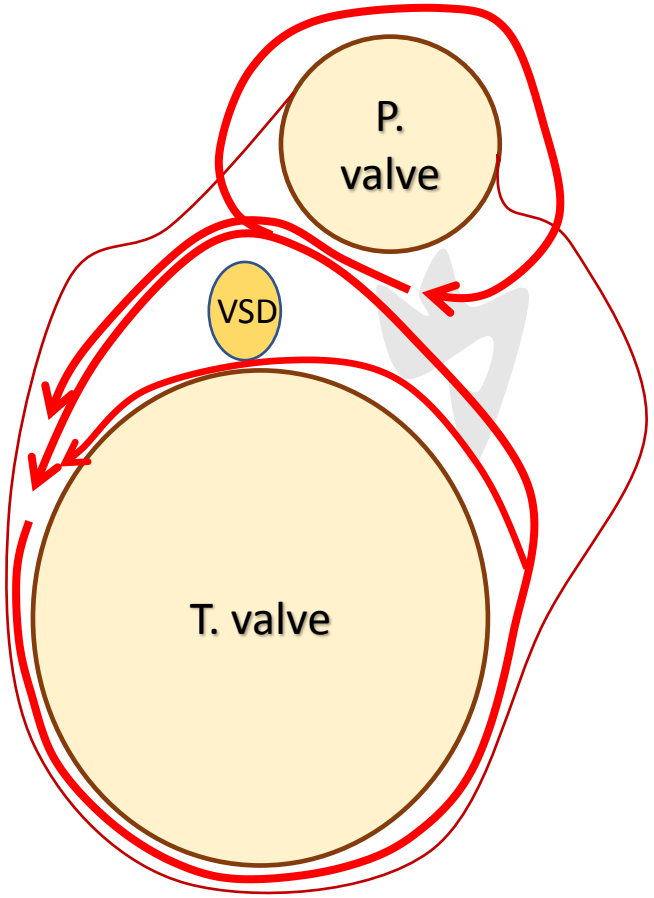
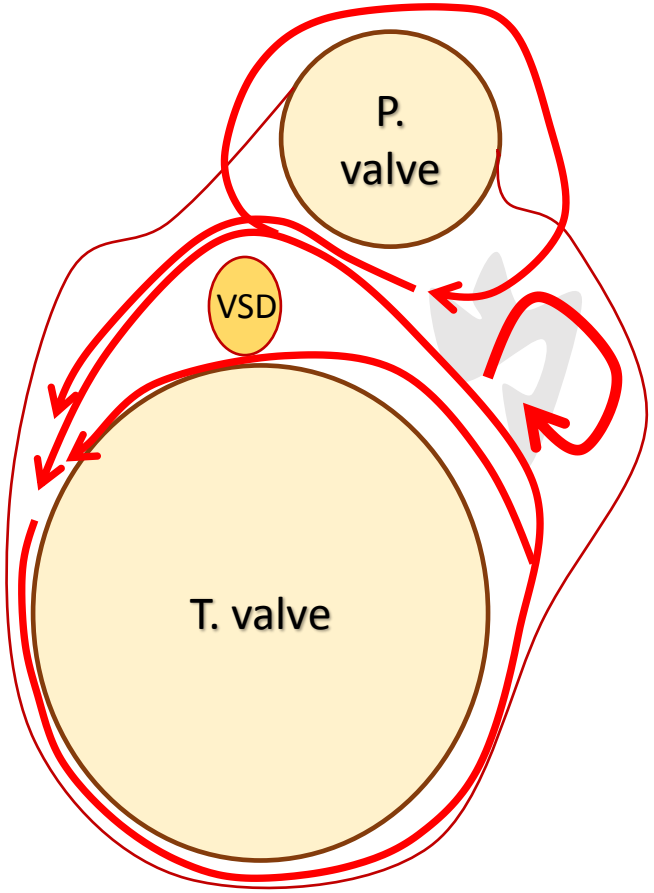
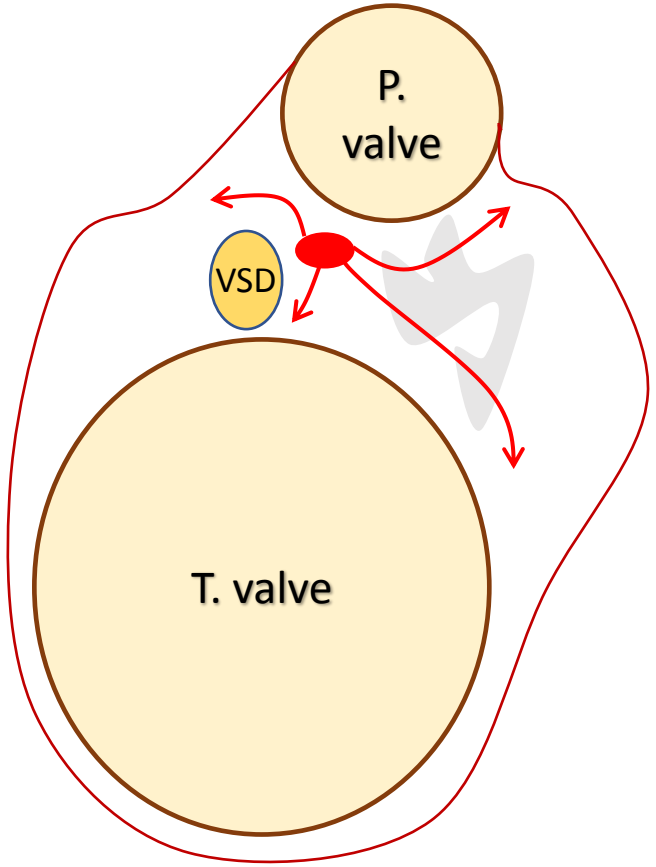


PA view

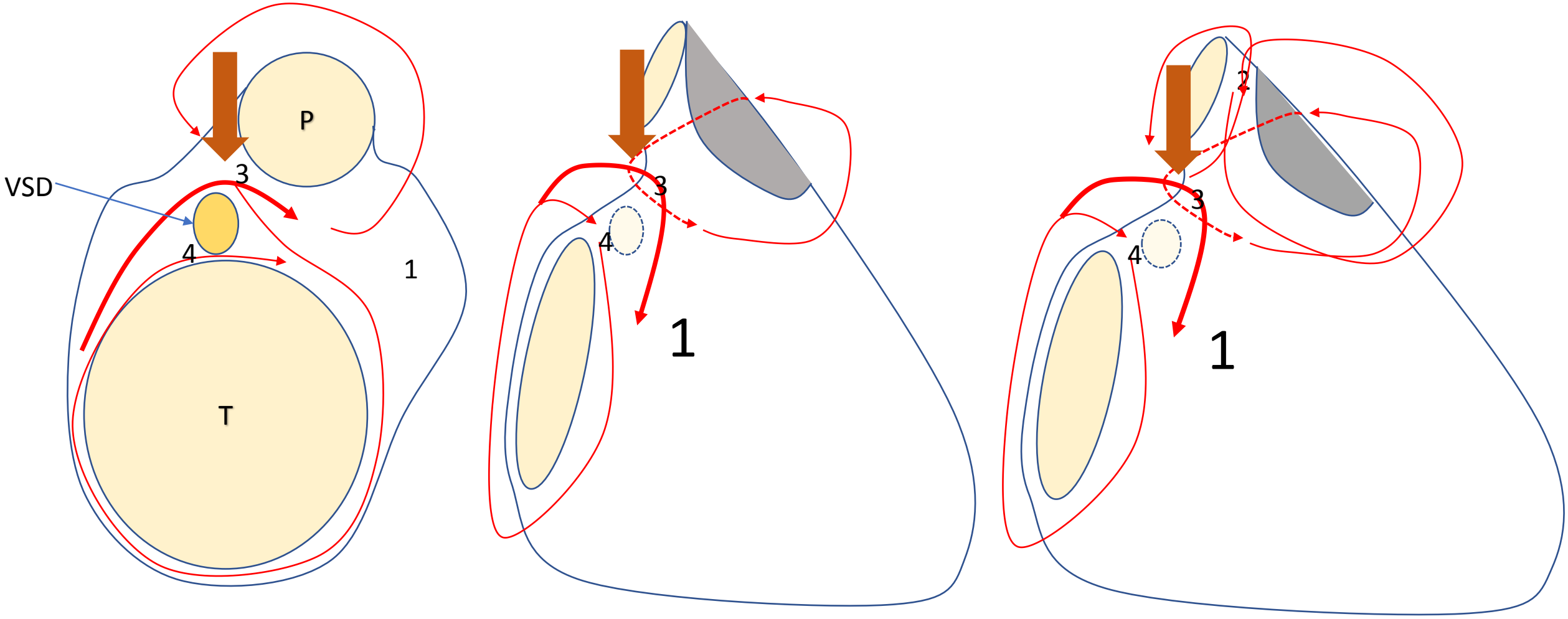


PA view



PA view

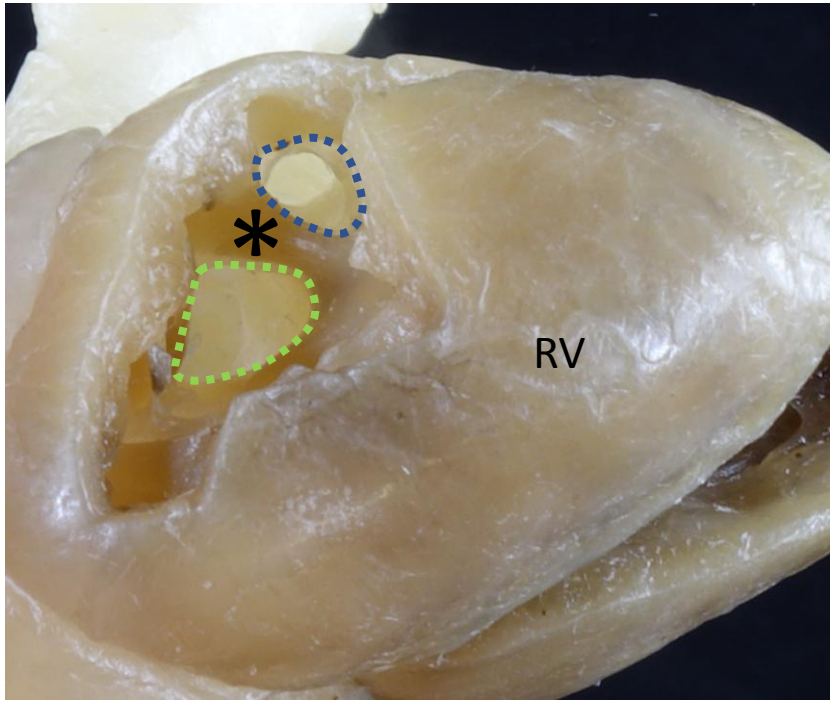
The majority of them **converge** to/through isthmus 3



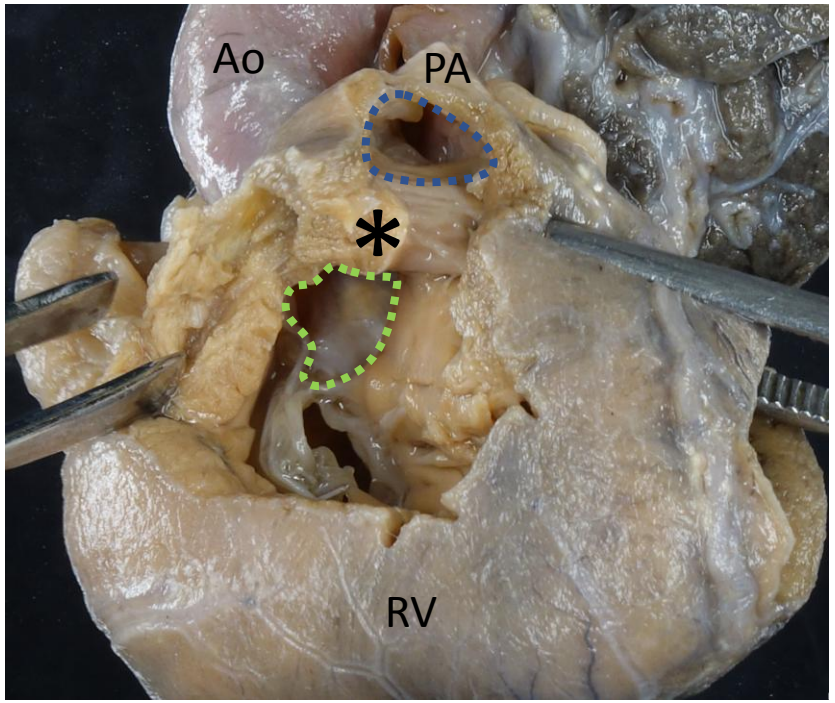
PA view

RAO view
Transannular patch

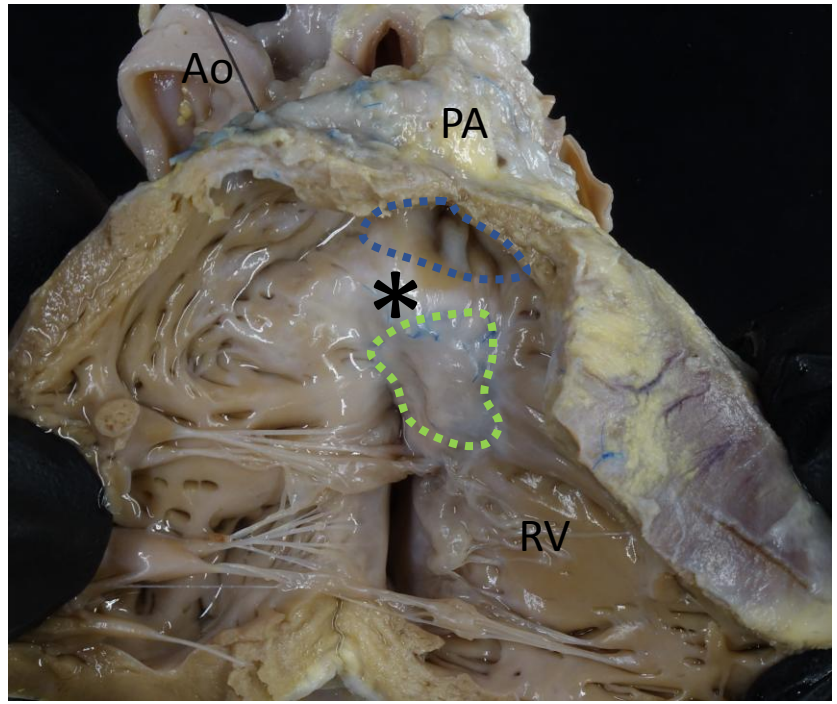
RAO view
(RVOT patch)



A



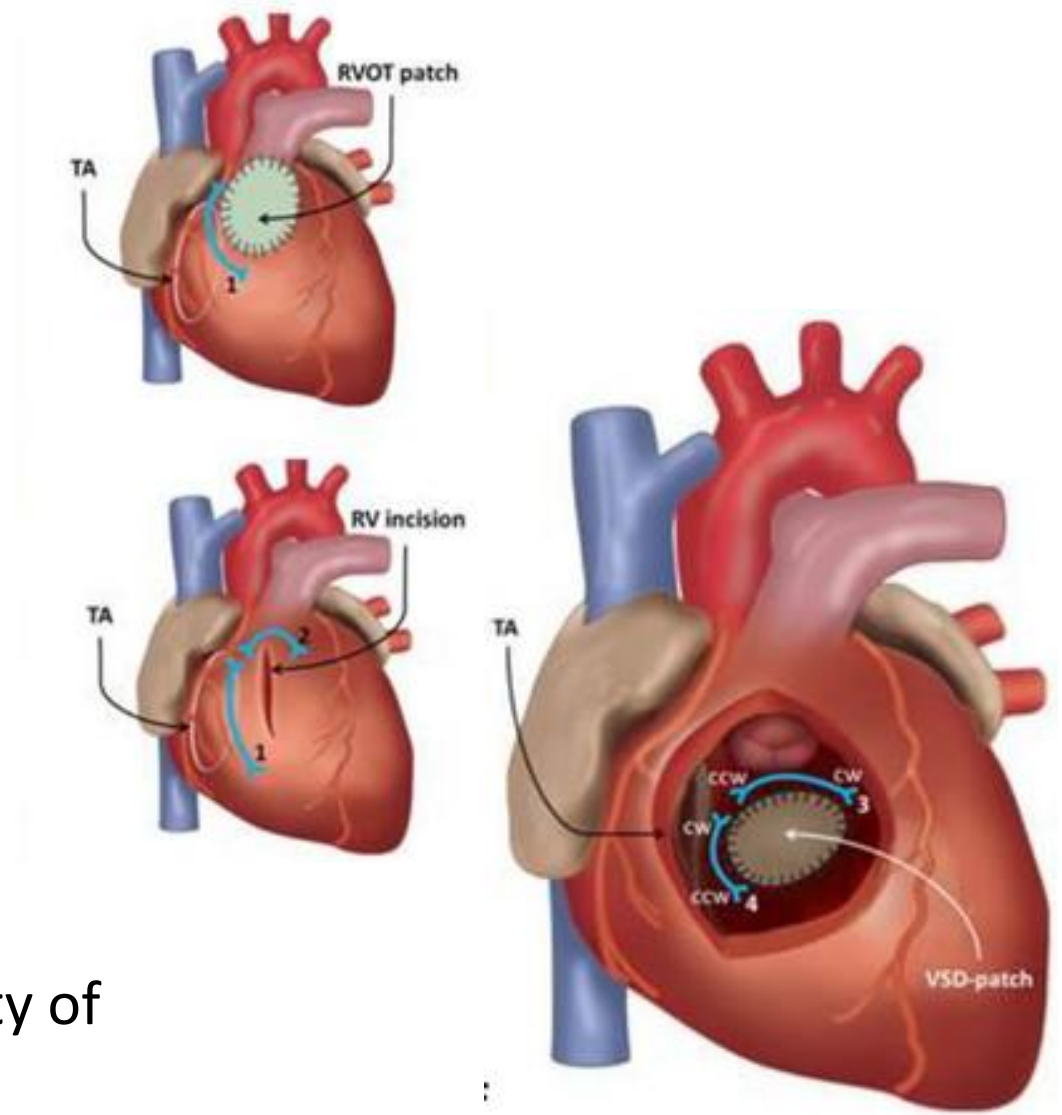
B



C

Isthmus to target?

- From anatomopathological point of view:
  - **Isthmus 1 : 99%**
  - Isthmus 2 : 42%
  - **Isthmus 3 : 99%**
  - Isthmus 4 : 6%
- From catheter ablation point of view:
  - **Isthmus 3** is easier to block
- From statistical point of view:
  - **Isthmus 3** is the meeting point of the majority of the circuits



Zeppenfeld, Heart 2018, PMID 29305559

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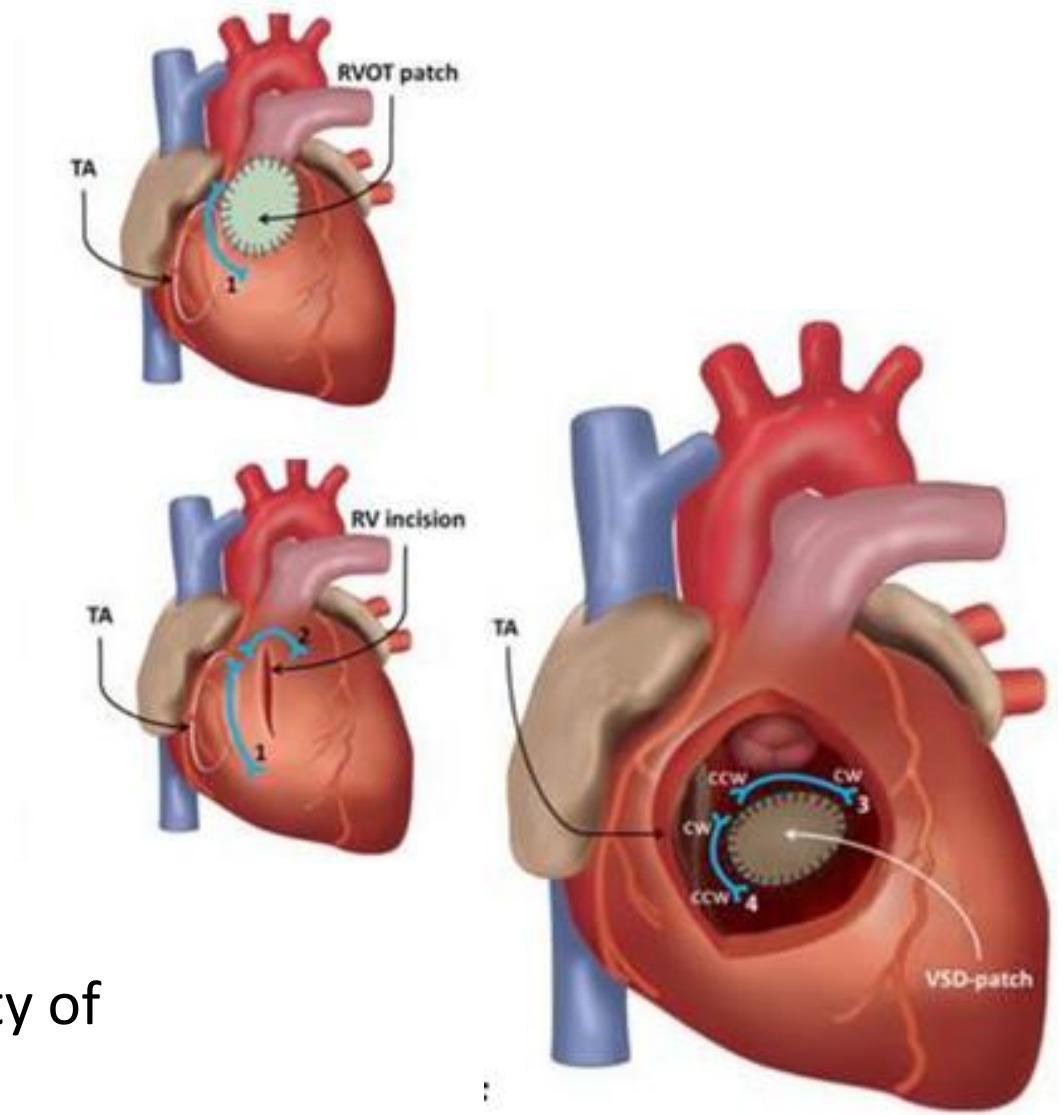
Isthmus 3 is THE TARGET++



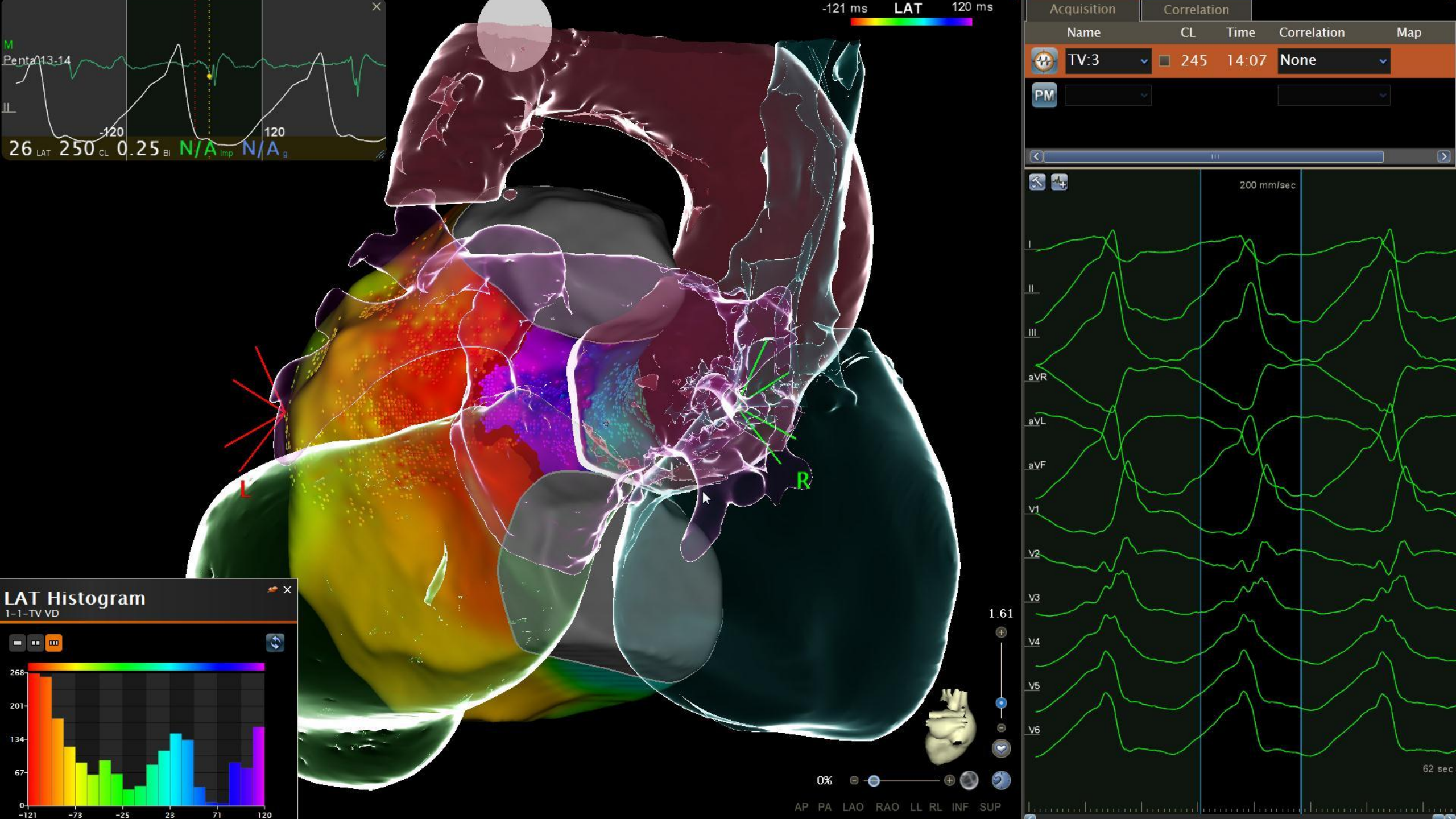
Zeppenfeld, Heart 2018, PMID 29305559

In real life…

- Lyon cohort : 27/27 patients = line between PV and TV = 100% isthmuses 3+4 targeted
- Kappel 2016 cohort :
  - 37 VT induced
  - => 24 in isthmus 3
  - => 10 in isthmus 1 (Isthmus 3 dependent circuit)
  - => 2 in isthmus 2
  - => 1 in isthmus 4

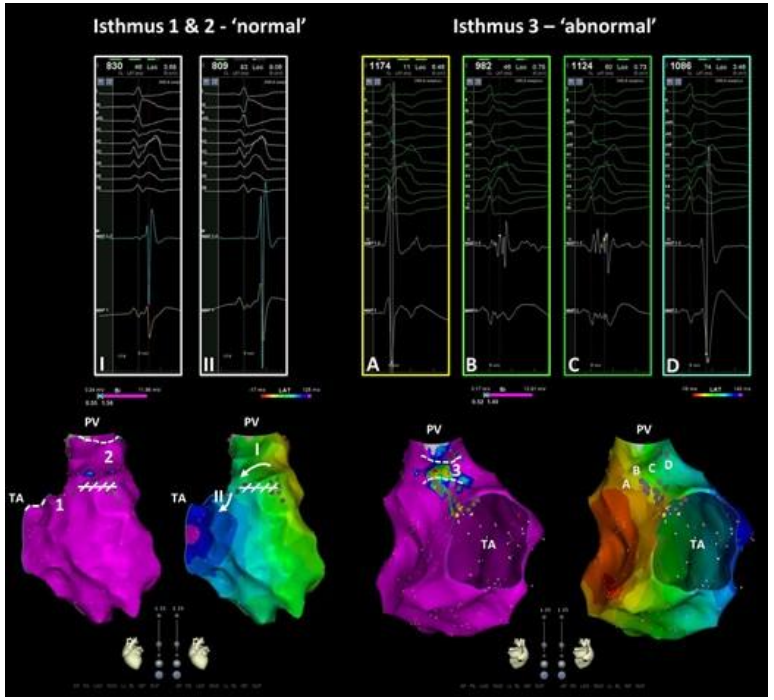
  - => blocking 3+4 would treat 35/37 VT in this cohort…

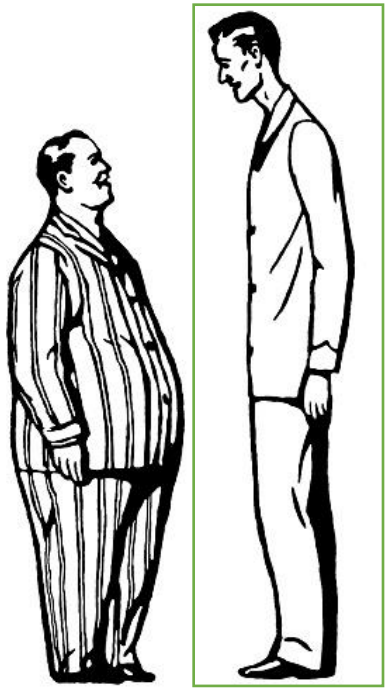
Kapel et al. EHJ 2016

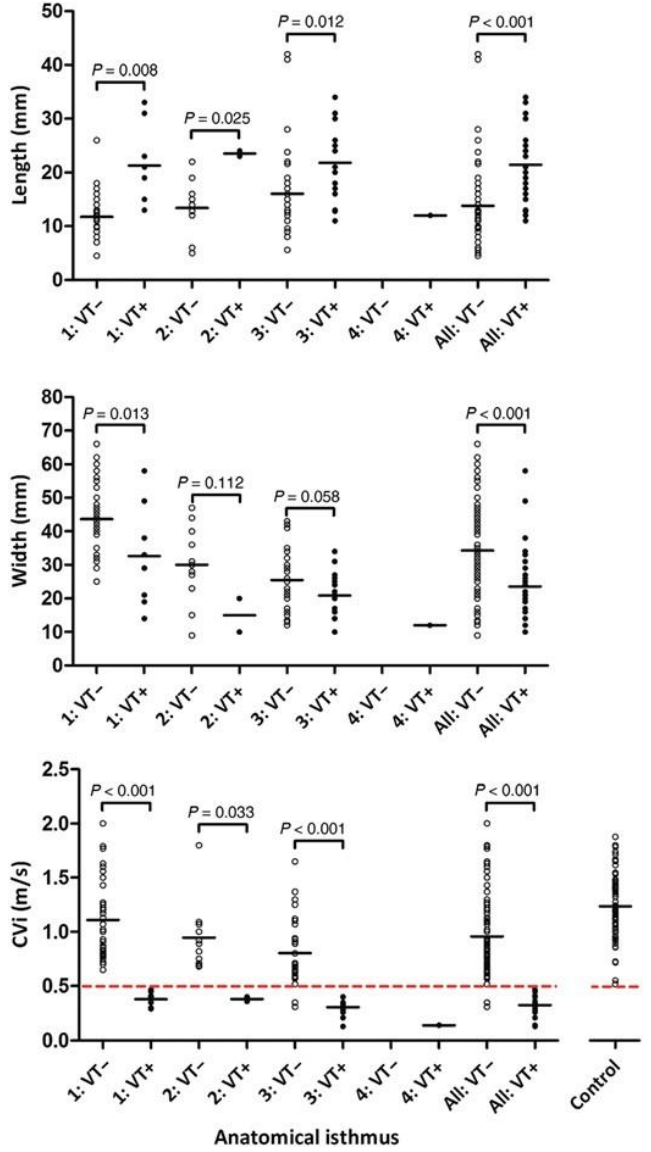

-121 ms
LAT
120 ms
Acquisition
Correlation
Name
CL
Time
Correlation
Map
TV:3
245
14:07
None
PM
M
Penta13-14
II
-120
120
26 LAT 250 CL 0.25 Bi N/A Imp N/A g
200 mm/sec
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
62 sec
L
R
LAT Histogram
1-1-TV VD
268
201
134
67
0
-121
-73
-25
23
71
120
1.61
0%
AP PA LAO RAO LL RL INF SUP

At risk isthmus…

- Conduction velocities < 0.5m/s
- Not too width (mean: 20mm vs 35mm)
- Relatively high (mean length 22mm vs 15mm)

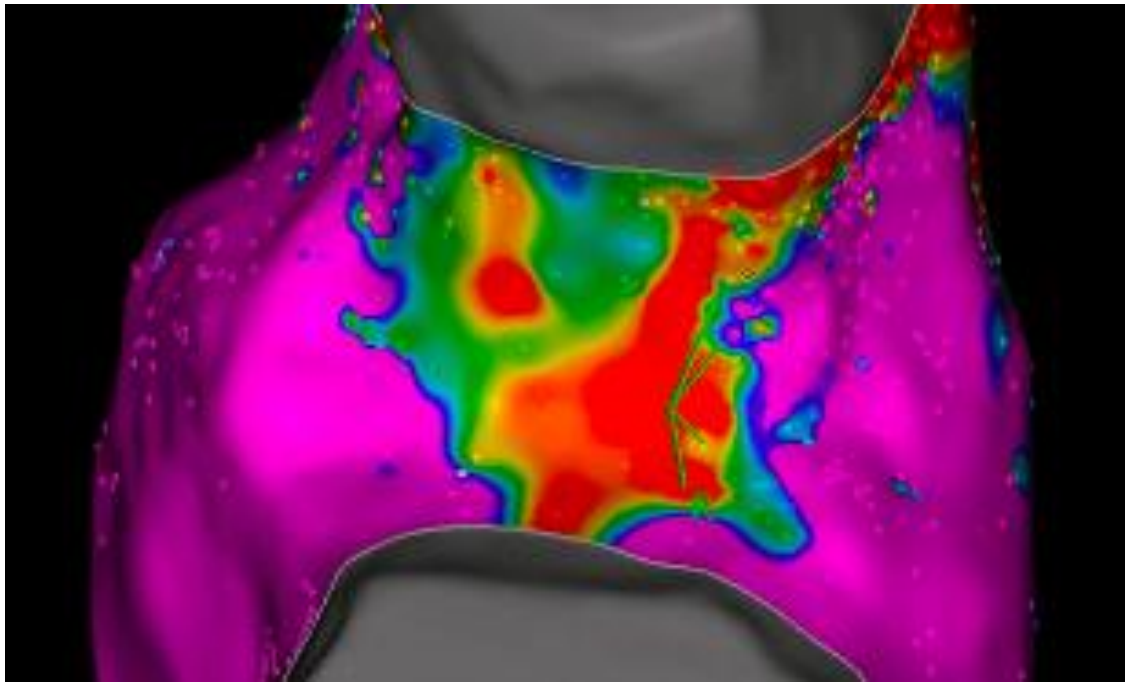




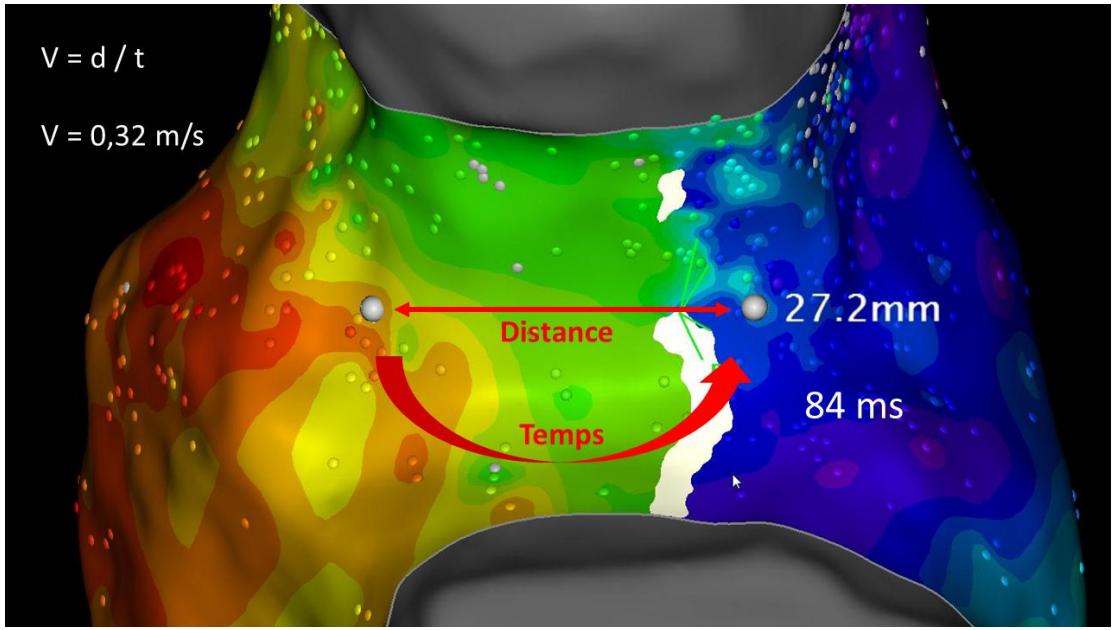


Kapel et al. EHJ 2016

In our case…



Width:27mm
Length (3): 21mm
V = 0.32m/s

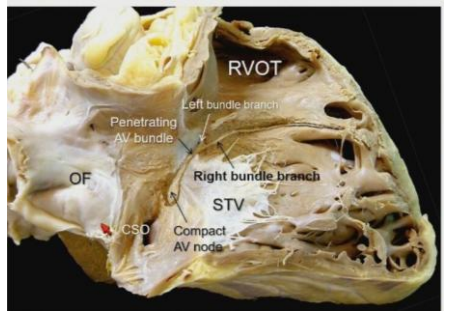


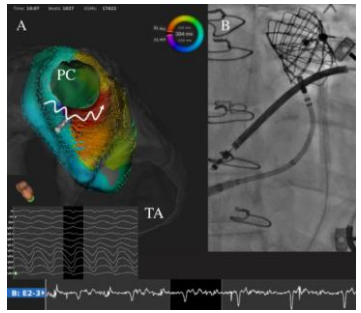
Challenges for VT ablation in TOF

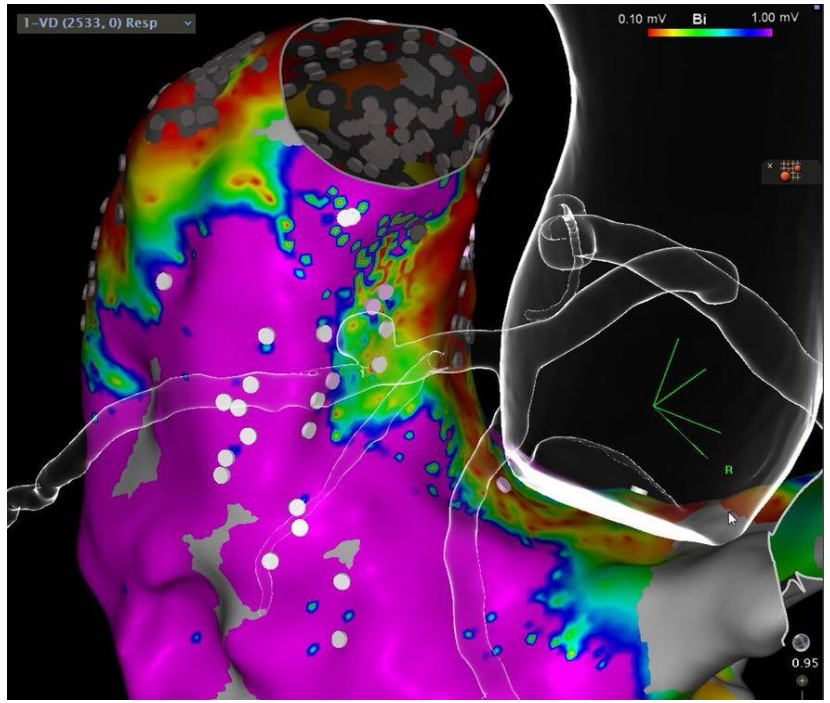
- Conduction system
- Isthmus 3 thickness, trabeculations, stability of catheters, prior PVR that would protect portions of anatomical isthmuses
- Vascular access

- Sometimes prevent RF lesions to block the line

=> Left side access sometimes required





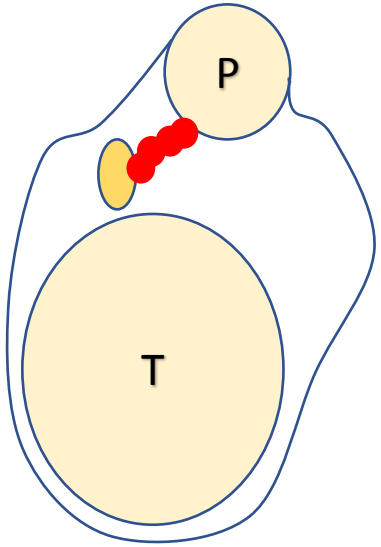


Combes et al, EHJ, doi:10.1093/eurheartj/ehz610

Cohen et al, JACC 2021 PMID 33573746

Is isthmus 3 should be blocked?

- Initially: preventive dissection of isthmus during the initial corrective surgery?
  - =>Too small (conduction system)(?)
  - BUT:
    - Atriotomy++ (to limit isthmus 2)
    - Combined isthmus 2 surgical ablation in case of ventriculotomy incision?
    - Repair ASAP? (to limit isthmus 1 size)



- At the time of PVR? (50% of the TOF population)
  - Preventively during surgical repair? (only with EP control+++, if CA failed)
  - Before surgical repair, after stratification of at risk patients?
  - Before any percutaneous replacement (valve skeleton on the isthmus 3)

Or should the evaluation be done…

- Before PVR
- In case of symptom
- On a regular basis

EP study before PVR: our results

- PVS in 122 patients before PVR *(HEGP/Necker/ML/ToulousePasteur/LyonLP)*
- 23 monomorphic VT (18%)
- Predictive factors:
  - **Age** (37.2 vs 46.3; p=0.009)
  - **Time to surgical repair** (31.8 vs 39.8; p=0.003)
  - Palliative shunt (34.7% vs 60.9%; p=0.026)
  - **History of SV arrhythmia** (17.2% vs 56.2%; p=0.001)
  - Previous history of PVC, VT (p=0.04)
  - NYHA >II (p=0.02)
  - **Ventriculotomy** incision (60% vs 88.8%; p=0.021)
  - **RVOT diameter** (27.1 vs 31.4mm; p=0.024) (but no RV size+++)

Bessière, Combes, Waldmann; To be published…

EP study before PVR: our results

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  - Age
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  - Palliative shunt
  - History of SV arrhythmia
  - Previous history of PVC, VT
  - NYHA III/IV
  - Ventriculotomy incision
  - RVOT diameter
  - RV dilatation

Similarities with SCD risk score

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Khairy et al. Circ 2007

Is PVR plays AAR role?

- **No**
  - Harrild et al Circulaion 2009
  - BUT… "A total of 98 patients with TOF and **late** PVR for RV dilation were identified"
- PVR indications for rhythm disorder are poor

- **Given our results, PVR should/may be discuss earlier+++**
  - Age (37.2 vs 46.3; p=0.009)
  - Time to surgical repair (31.8 vs 39.8; p=0.003)

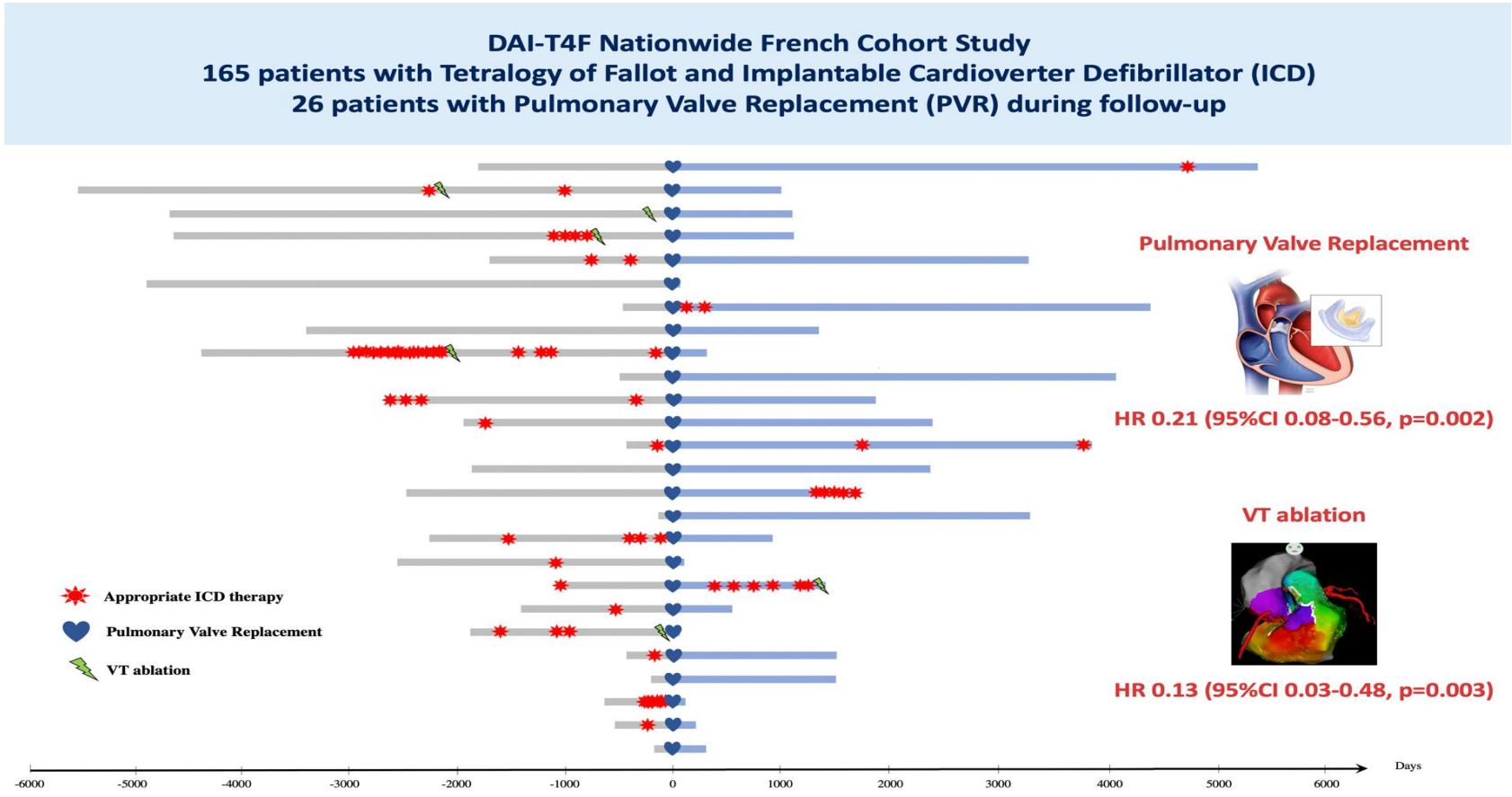
- !!: PVR improves symptoms and RVEF but no survival (yet…)

Indications
I. Asymptomatic patients with ≥2 of the following criteria:
a. RV end-diastolic volume index >150 mL/m^2 or z score >4. In patients whose body surface area falls outside published normal data: RV/LV end-diastolic volume ratio >2
b. RV end-systolic volume index >80 mL/m^2
c. RV ejection fraction <47%
d. LV ejection fraction <55%
e. Large RVOT aneurysm
f. QRS duration >160 ms
g. Sustained tachyarrhythmia related to right-sided heart volume load
h. Other hemodynamically significant abnormalities:
• RVOT obstruction with RV systolic pressure ≥0.7 systemic
• Severe branch pulmonary artery stenosis (<30% flow to affected lung) not amenable to transcatheter therapy
• Greater than or equal to moderate tricuspid regurgitation
• Left-to-right shunt from residual atrial or ventricular septal defects with pulmonary-to-systemic flow ratio ≥1.5
• Severe aortic regurgitation
II. Symptomatic patients fulfilling ≥1 of the quantitative criteria detailed above. Examples of symptoms and signs include:
a. Exercise intolerance not explained by extracardiac causes (eg, lung disease, musculoskeletal anomalies, genetic anomalies, obesity), with documentation by exercise testing with metabolic cart (≤70% predicted peak o_2 for age and sex not explained by chronotropic incompetence)
b. Signs and symptoms of heart failure (eg, dyspnea with mild effort or at rest not explained by extracardiac causes, peripheral edema)
c. Syncope attributable to arrhythmia
III. Special considerations:
a. Because of higher risk of adverse clinical outcomes in patients who underwent TOF repair at ≥3 years of age, PVR may be considered if they fulfill ≥1 of the quantitative criteria in section I
b. Women with severe PR and RV dilatation or dysfunction may be at risk for pregnancy-related complications. Although no evidence is available to support benefit from prepregnancy PVR, the procedure may be considered if fulfilling ≥1 of the quantitative criteria in section I

Geva. Circ 2013

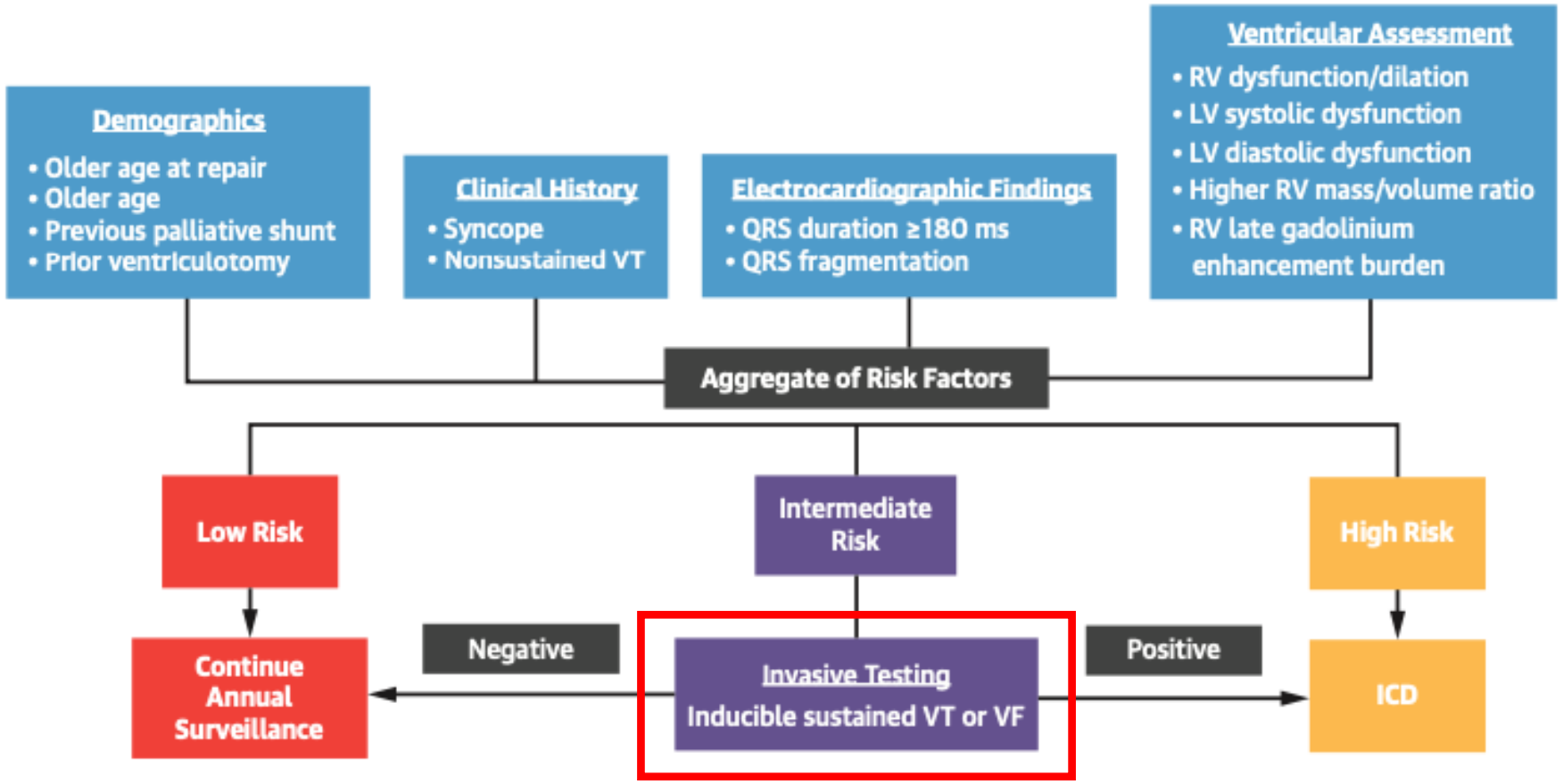
Is PVR plays AAR role?

- Possibly in <u>high risk selected patients</u>…



Bessière et al, JACC EP 2021

Invasive testing **is indicated to stratify** SCD risk



Cohen et al, JACC 2021 PMID 33573746

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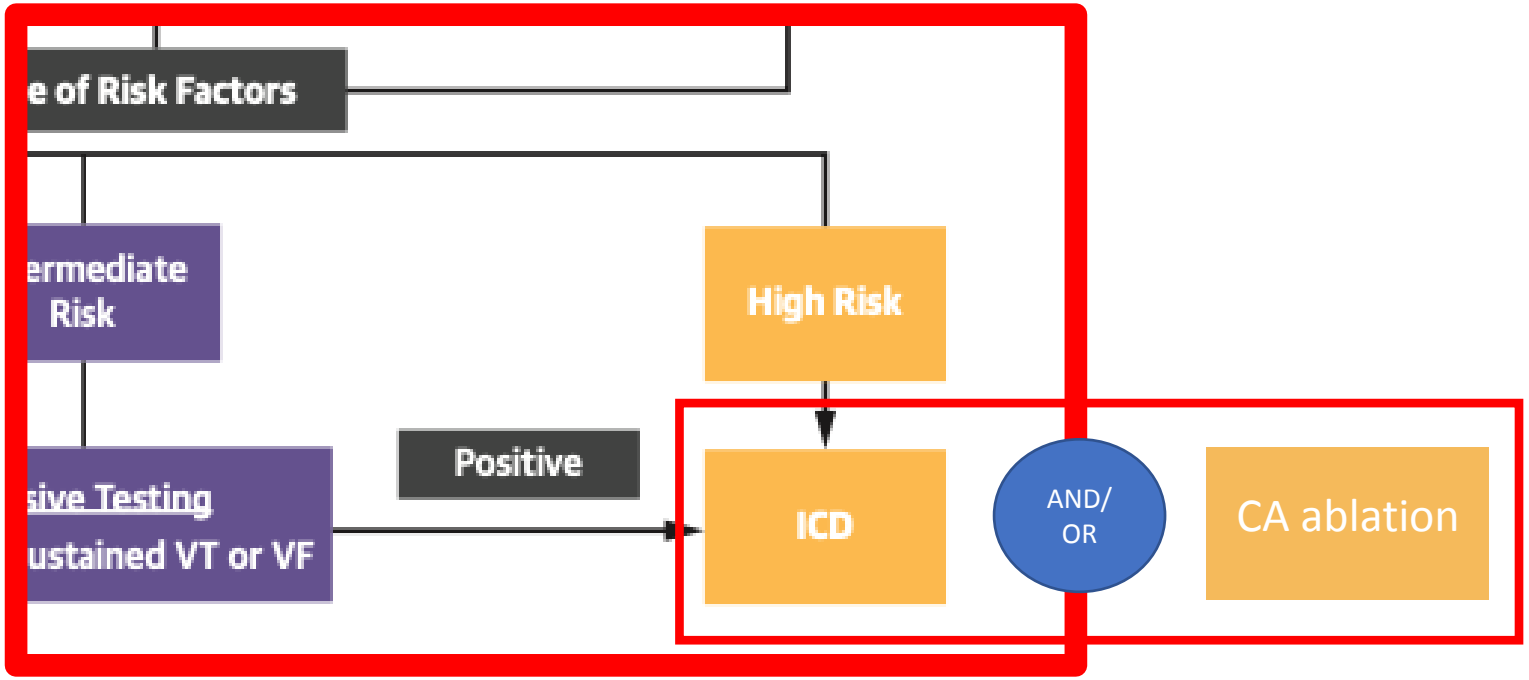
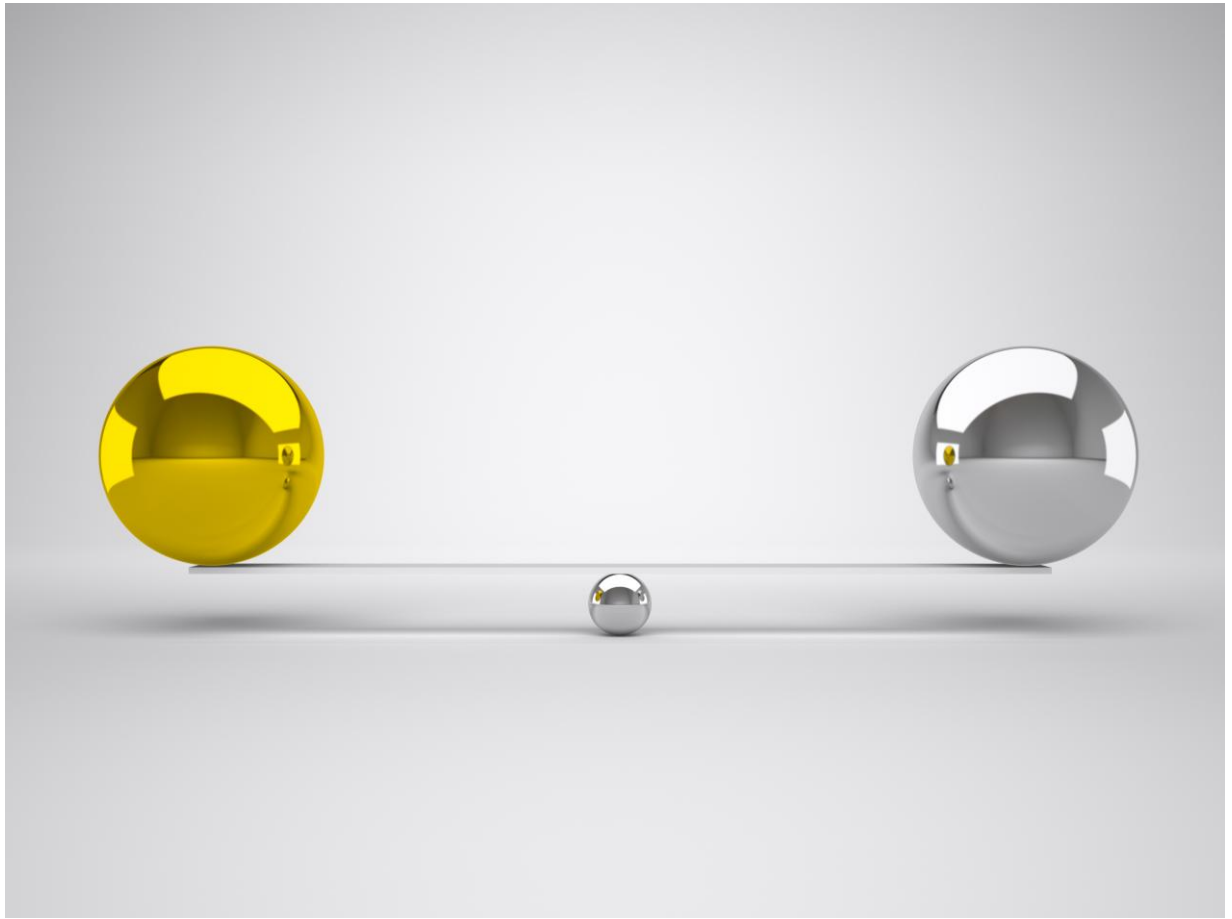
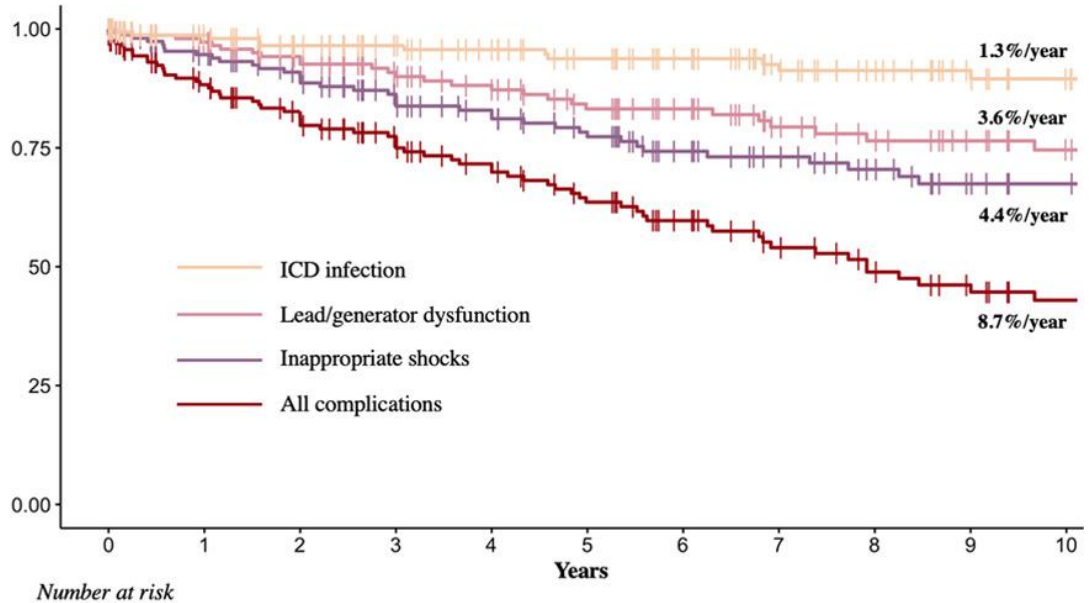


ICD and/or VT ablation to manage rhythm?



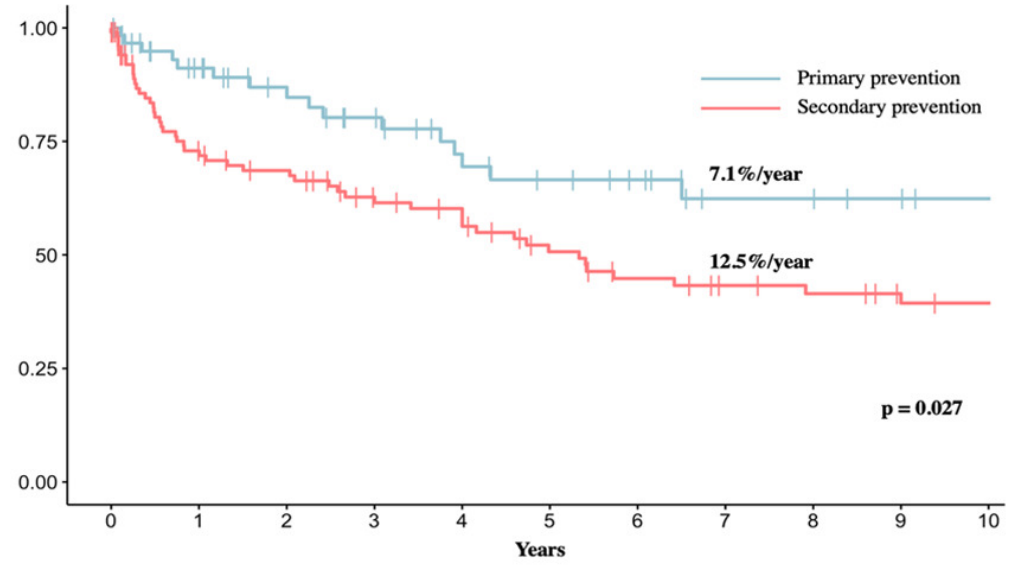
In case of ICD placement

- Device related complications : **8.7%!**
- Associated AAR drugs related complications
- Appropriate therapy rate (PP: 7.1%; SP: 12.5%)



Number at risk

All complications	165	129	111	96	83	69	57	46	37	31	25



Number at risk

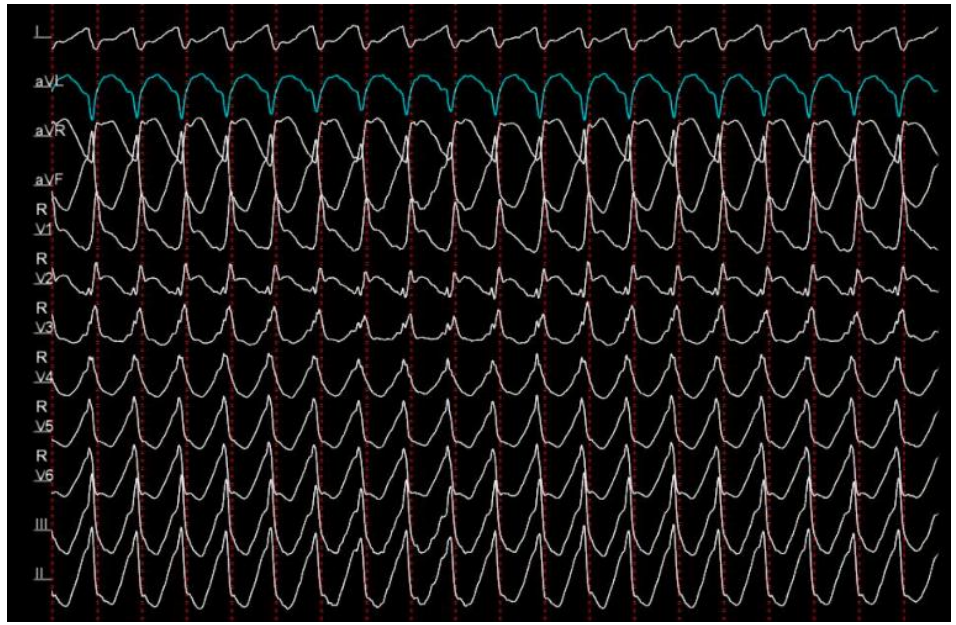
Primary prevention	61	47	39	33	26	22	19	13	13	11	9
Secondary prevention	104	68	61	50	46	35	29	25	23	19	18

Waldmann et al. Circ 2020

In case of VT ablation

- **Arrhythmia** characteristics
  - Induced or spontaneous VT =(Preventive or curative VT ablation)
  - Mono or polymorphic VT/VF
  - VT TCL / Hemodynamic tolerance
  - RV volume, RV voltage, scars
  - Isthmus properties (size, thickness, velocities…)

- **Clinical** characteristics



In case of VT ablation

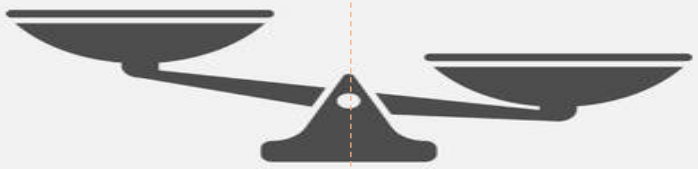
- Catheter ablation **expected efficacy**

*"**Catheter ablation is currently recommended as adjunctive therapy** to ICD patients with CHD who have recurrent monomorphic VT or appropriate ICD therapies that are not manageable by device reprogramming or drug therapy"*
Priori et al. Europace 2015

*"A combined endpoint of non-inducibility and conduction block was associated with **freedom of VT recurrence during 46 ± 29 months follow-up** in a recent series of 25 CHD patients"*
Kapel et al. Circ AE 2015

*"**Considering the high acute success rates and low recurrence rates VT ablation may offer a reasonable alternative to ICD therapy in carefully selected patients with preserved cardiac function**"*
Hernandez-Madrid et al. Europace 2018

Positive PVS or spontaneous VT…

	VT ABLATION +/- PVR	ICD
Young age Normal RVEF and LVEF Absence of ventriculotomy Minimal Scar Single monomorphic well tolerated VT Non inducibility post ablation Demonstration of bidirectional block across the targeted isthmus post ablation		
Older age Impaired RVEF or LVEF LV diastolic dysfunction or elevated LVEDP Ventriculotomy Prior palliative shunt Extensive scar More than one inducible VT Poorly tolerated VT Targeted isthmus difficult to block Inducibility post ablation		

Physical activity in CHD

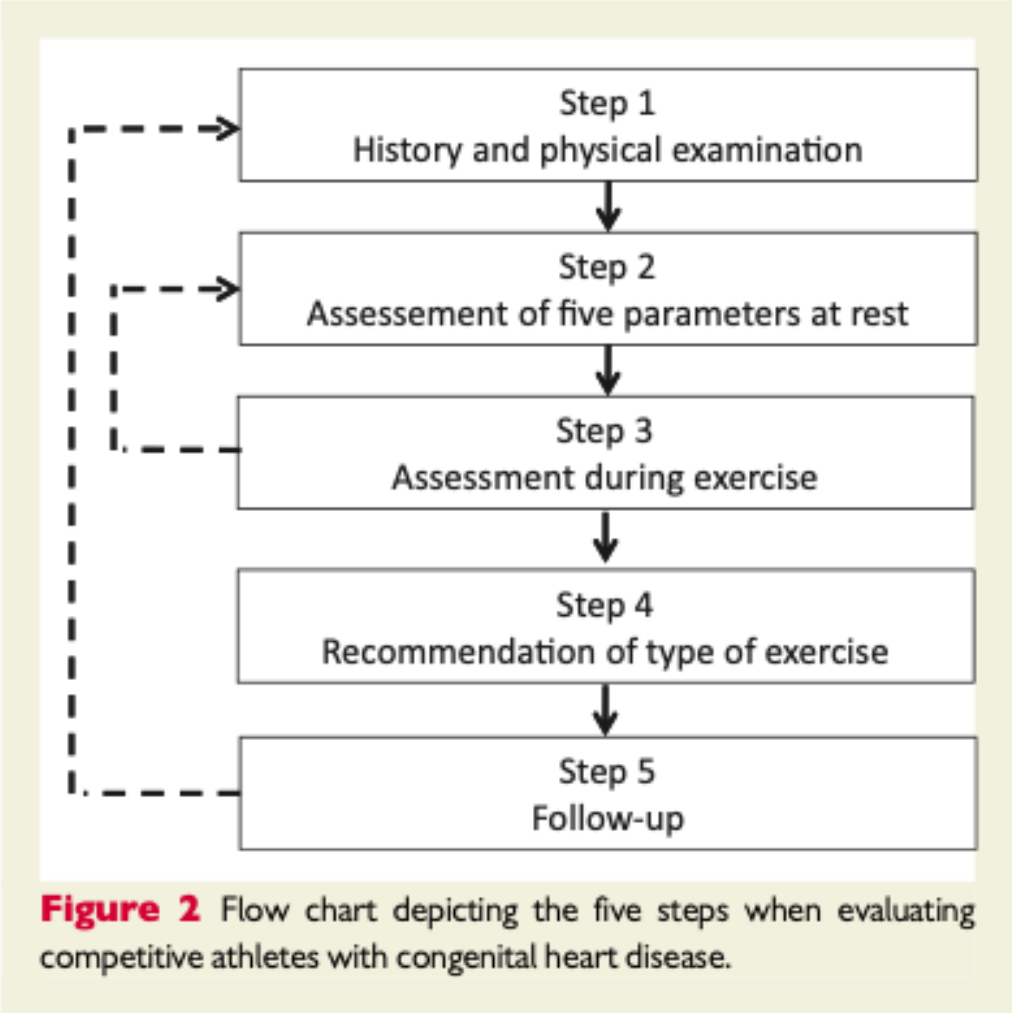


Figure 2 Flow chart depicting the five steps when evaluating competitive athletes with congenital heart disease.

- Ventricles structure and function
- Pulmonary artery pressure
- **Arrhythmia**
- Aorta
- Arterial O2 saturation

- Ventricles structure and function
- Pulmonary artery pressure
- **Arrhythmia**
- Aorta
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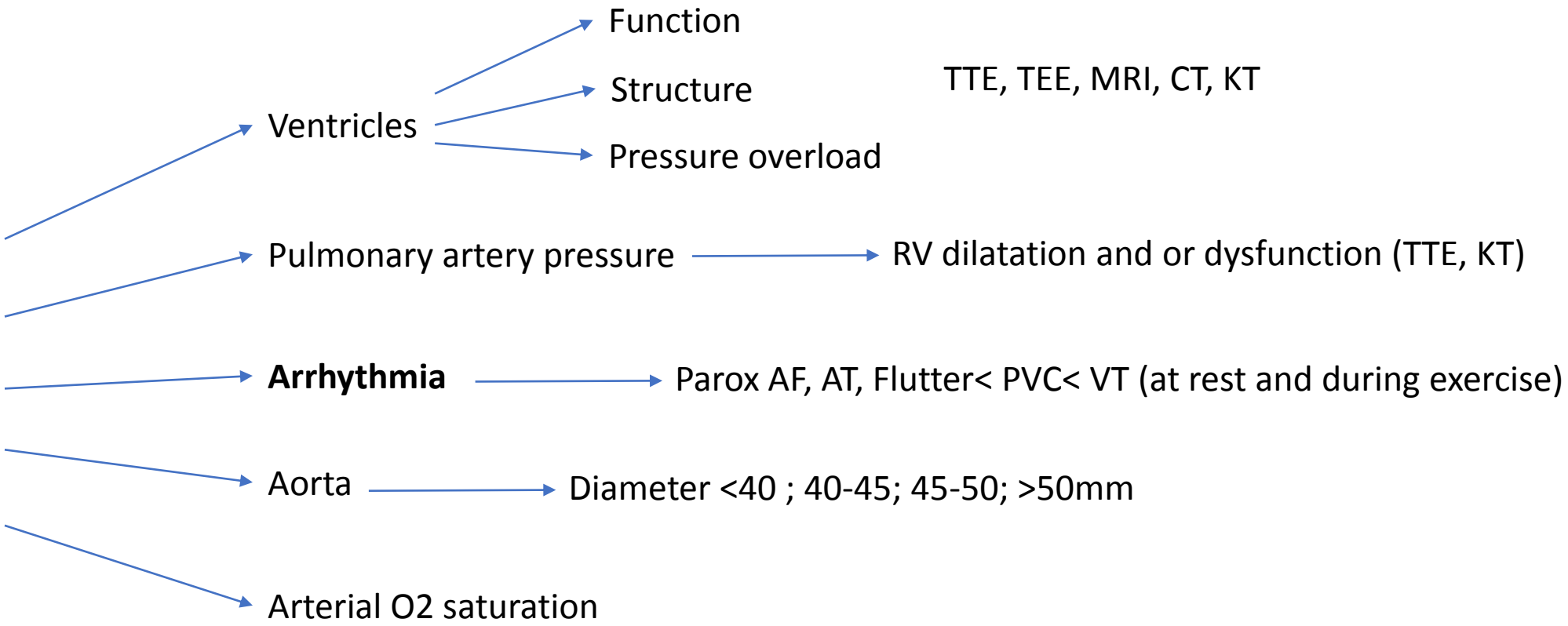
European Heart Journal (2020) 41, 4191–4199

Physical activity in CHD



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Physical activity in CHD

1. Ventricles	No systolic dysfunction No/mild hypertrophy No/mild pressure load No volume load	Mild systolic dysfunction Volume load without remodelling	Moderate systolic dysfunction Moderate hypertrophy Moderate pressure load Volume load with mild remodelling Single ventricle physiology Systemic right ventricle	Severe systolic dysfunction Severe hypertrophy Severe pressure load Volume load with severe remodelling
2. Pulmonary artery pressure	Low probability of pulmonary hypertension	PH without RV dilatation or dysfunction		PH with RV dilatation or dysfunction
3. Aorta	No/mild dilatation	Moderate dilatation	Severe dilatation	Dilatation approaching indication for repair
4. Arrhythmia at rest/during exercise	No arrhythmia	Mild arrhythmic burden Non-malignant arrhythmia		Significant arrhythmic burden Malignant arrhythmia
5. Saturation at rest/during exercise	No central cyanosis		Mild central cyanosis	Severe central cyanosis
	A	B	C	D
	When all applicable	When ≧1 parameters applicable AND no parameter falls within columns C or D	When ≧1 parameters applicable AND no parameter falls within column D	When ≧1 parameters applicable
Choice of competitive sport	All sports	Skill, Power, or Mixed sports	Skill sports only	NO COMPETITIVE SPORT

Figure 3 Flow chart depicting in detail Steps 2–4. Following assessment of the five variables at rest and during exercise, an individualized recommendation can be provided.

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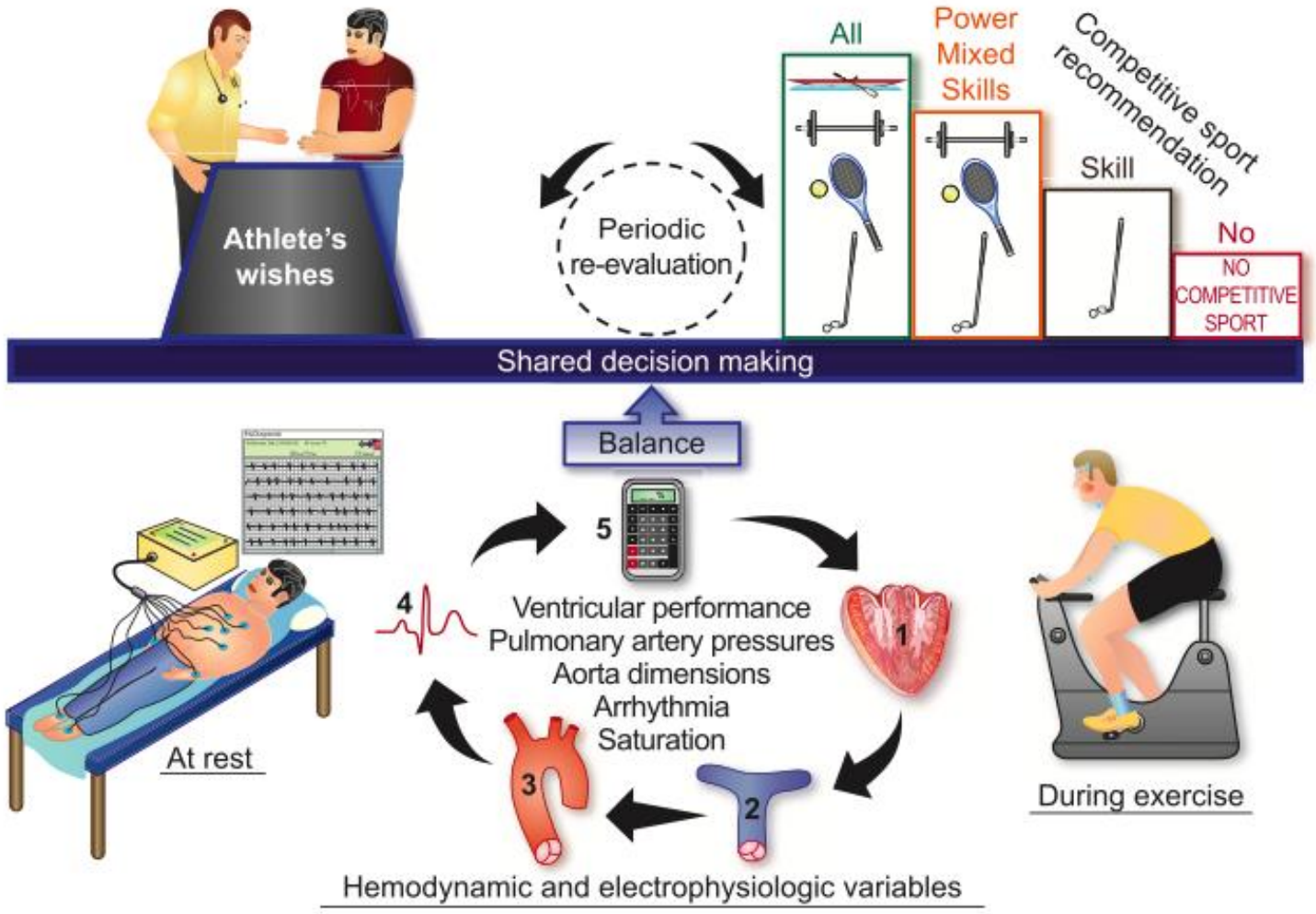
Physical activity in CHD



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In case of TOF

- Carefully evaluate (periodic re-evaluation)
  - RV++/LV: MRI/TTE
  - PV regurgitation: TTE, consider **PVR if needed**
  - Isthmus 3/SCD risk: ECG/ EP study/ stress test +/- **PVS**
  - General status: VO2max
- And consider
  - SCD risk stratification score
  - Holter monitoring (watches, Reveal…)
- Avoid “at-risk” situation (scuba dive, rock climbing)
- Patient education, cardiac resuscitation

In case of TOF

- If ICD: consider the risk of appropriate and inappropriate shock
- Altitude <1500m if cyanotic, high PA pressure…
- ACO: limit contact sports

Thank you

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