



Indications de Stimulation & Défibrillation Cardiopathies Congénitales de l'Adulte

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ASSISTANCE
PUBLIQUE



HÔPITAUX
DE PARIS



PM & ICD Guidelines



EUROPEAN
SOCIETY OF
CARDIOLOGY

**Pacing, SCD
ESC 2013, 2015**



**SCD
AHA/ACC 2017**



Heart
Rhythm
SocietySM



American Heart
Association



Canadian
Heart Rhythm
Society
Société canadienne
de rythmologie



**Arrhythmias in CHD
PACES/HRS 2014**



Pacing & ICD Scenarios

- **Pacing (SSS or AVB)**
 - Inherent to the underlying anatomic substrate
 - Immediate postoperative period secondary to injury to conduction system
 - Present years later as a result of a slow deterioration in the conduction system
- **Pacing (CRT)**
 - Challenge of the «response»
- **ICD**
 - Secondary prevention
 - Challenge of risk stratification (I^{ary} prevention)



PM & ICD Guidelines

**In non-ACHD,
easy!!**



European Heart Journal (2013) 34, 2281–2329
doi:10.1093/eurheartj/ehf150

ESC GUIDELINES

2013 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy

The Task Force on cardiac pacing and cardiac resynchronization therapy of the European Society of Cardiology and the European Heart Rhythm Association



European Heart Journal
doi:10.1093/eurheartj/ehv316

ESC GUIDELINES

2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC)

Endorsed by: Association for European Cardiology (AEPC)

Authors/Task Force Members: Silvia G. Priori, Carina Blomström-Lundqvist* (Co-chairpersons), Nico Blom^a (The Netherlands), Martin Borggrefe (Germany), Perry Mark Elliott (UK), Donna Fitzsimons (UK), Gerhard Hindricks (Germany), Paulus Kirchhof (Denmark), Karl-Heinz Kuck (Germany), Aron Klug (Denmark), Nikolaos Nikolaou (Greece), Tone M. Norekanger (Norway), and Dirk J. Van Veldhuisen (The Netherlands)

2017 AHA/ACC/HRS Guideline for Management of Patients With Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Rhythm Society

Developed in Collaboration With the Heart Failure Society of America

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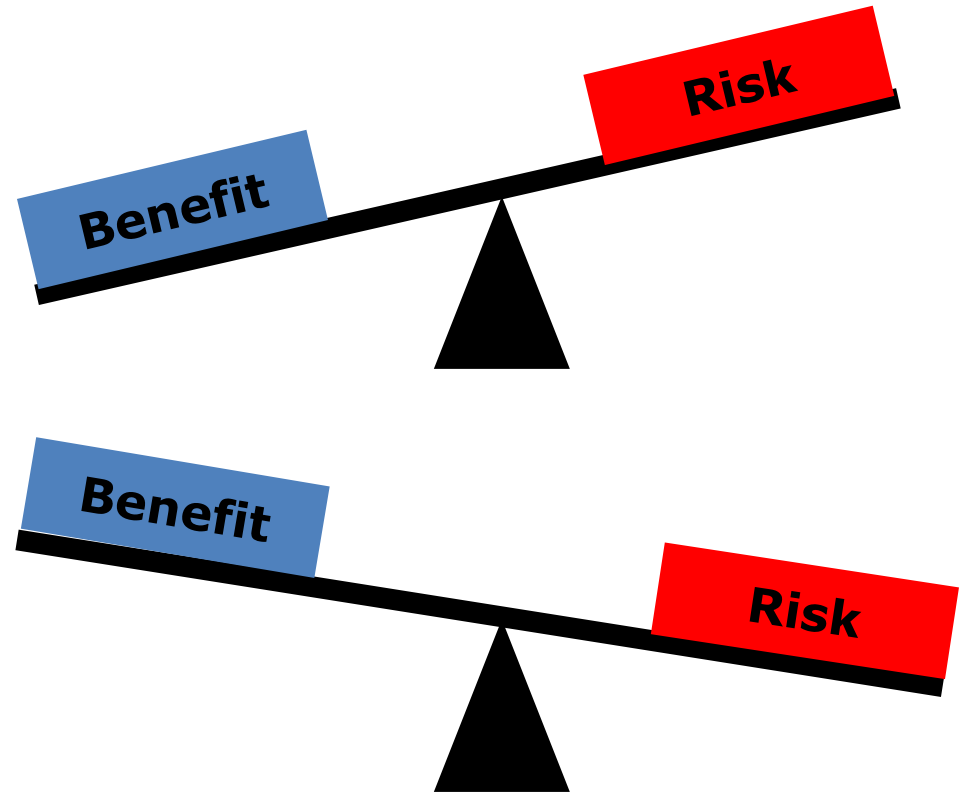
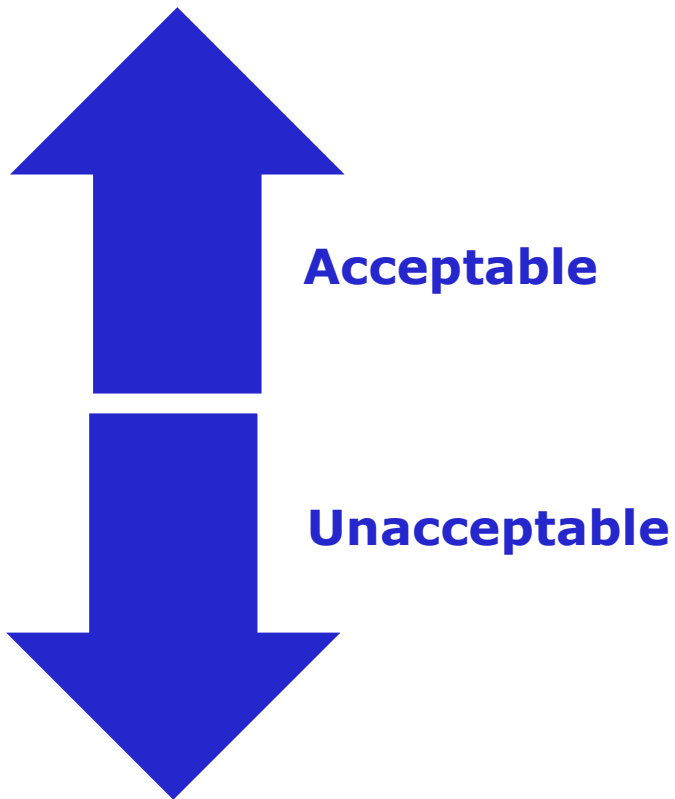
Usual Code...

Classes of recommendations	Definition	Suggested wording to use
Class I	Level of evidence A Data derived from multiple randomized clinical trials or meta-analyses.	Is recommended/is indicated
	Level of evidence B Data derived from a single randomized clinical trial or large non-randomized studies.	Should be considered
	Level of evidence C Consensus of opinion of the experts and/or small studies, retrospective studies, registries.	May be considered
Class III	Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.	Is not recommended

In ACHD, otherwise more challenging!!
(looking for the "box", reassuring time period?,...)



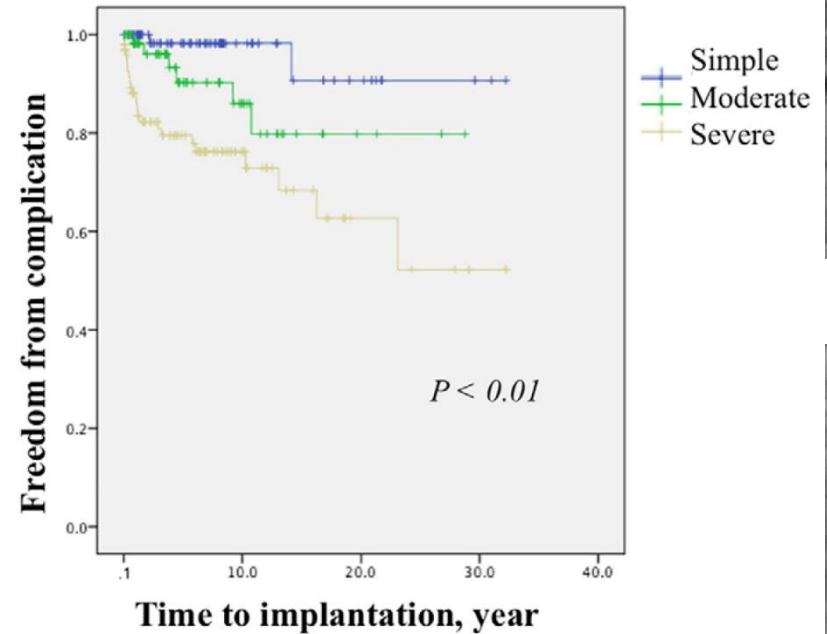
Benefit/Risk Balance



Benefit/Risk Balance

Ben

Ben



Royal Brompton Hospital 1978-2009



Midha D et al. Int J Cardiol 2017

Benefit/Risk Balance



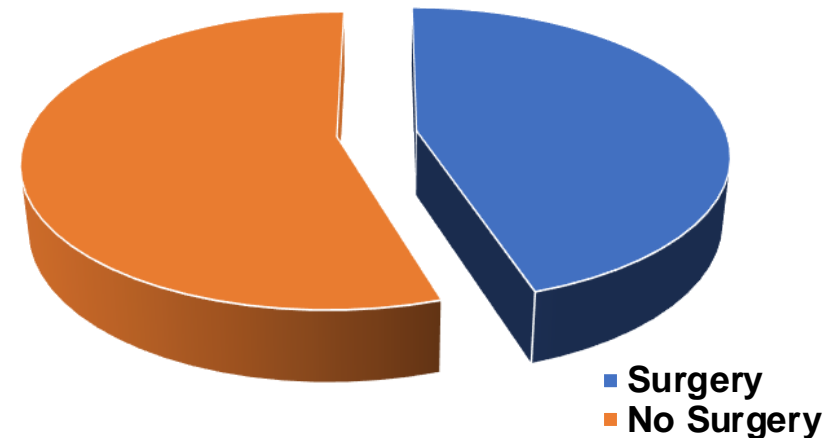
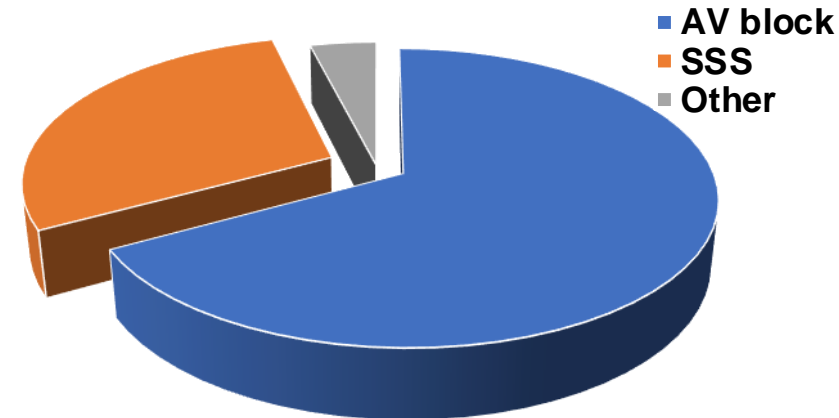
Pacing in ACHD

Complexity of CHD	Type of CHD	Prevalence (in CHD population)	Prevalence	
			SND	AV block
Simple	Patent ductus arteriosus	6-8%		
	Pulmonary stenosis	6-8%		
	Ventricular septal defect	30-32%		Light Blue
	Secundum atrial septal defect	8-10%	Light Blue	
Moderate	Aortic coarctation	5-7%		Light Blue
	Anomalous pulmonary venous return	0.5-2.5%	Dark Blue	
	Atrioventricular septal defect	3-5%		Dark Blue
	Aortic stenosis	3-5%		Light Blue
	Ebstein's anomaly	0.5-1.5%	Light Blue	Light Blue
	Tetralogy of Fallot	8-10%	Light Blue	
	Primum atrial septal defect	2-3%	Light Blue	Dark Blue
Severe	Truncus arteriosus	1.5-2%	Light Blue	Light Blue
	Pulmonary atresia	2-2.5%	Dark Blue	Light Blue
	Double outlet right ventricle	1.5-2%	Light Blue	Light Blue
	D-transposition of the great arteries	6-7%	Dark Blue	Light Blue
	L-transposition of the great arteries	1-2%	Light Blue	Dark Blue
	Hypoplastic left heart syndrome	3-4%	Dark Blue	Light Blue
	Other (heterotaxy, other single ventricles)	7-10%	Dark Blue	Dark Blue

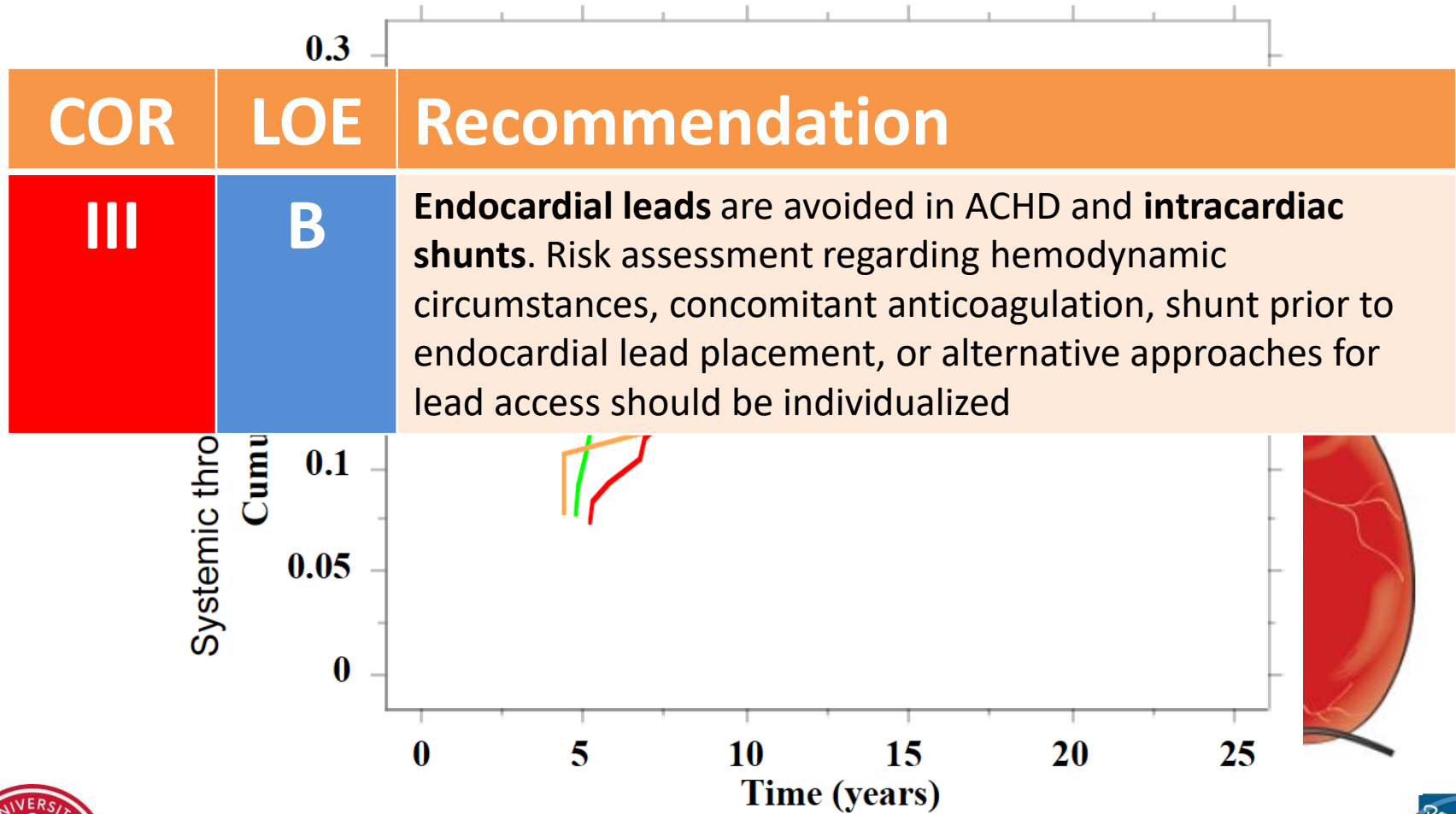
Khairy P et al. Heart Rhythm 2014

Pacing in ACHD

Congenital Anatomic Diagnosis	Number of Patients (%)
Complex anatomic DTGA	29 (17.3%)
Atrial septal defects	25 (14.8%)
Tetralogy of Fallot	24 (14.3%)
Ventricular septal defects	15 (8.9%)
LTGA	14 (8.3%)
Aortic valve stenosis	13 (7.7%)
Atrioventricular septal defect	10 (5.6%)
Ebstein's anomaly	7 (4.2%)
Coarctation of aorta	3 (1.8%)
Other	3 (1.8%)



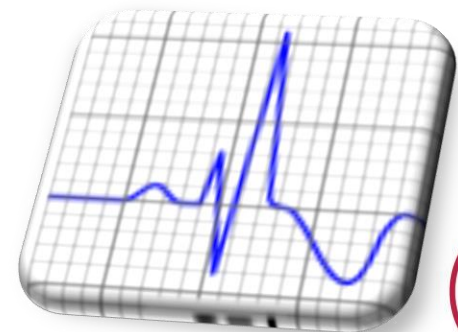
Pacing – Not Recommended



Khairy P et al Circulation 2008

Pacing – Not Recommended!

COR	LOE	Recommendation
III	B	Endocardial leads are avoided in ACHD and intracardiac shunts . Risk assessment regarding hemodynamic circumstances, concomitant anticoagulation, shunt prior to endocardial lead placement, or alternative approaches for lead access should be individualized
III	C	Pacing is not indicated in asymptomatic ACHD and bifascicular block with or w/o first-degree AVB in the absence of a history of transient complete AVB



Pacing – Recommended!

COR	LOE	Recommendation
I	B	Permanent pacing is recommended for ACHD and symptomatic SSS , including documented bradycardia or chronotropic incompetence that is intrinsic or secondary to required drug therapy
I	B	Permanent pacing is recommended in ACHD and symptomatic bradycardia in conjunction with any degree of AVB or with ventricular arrhythmias presumed to be due to AVB



Pacing – Recommended!

COR	LOE	Recommendation
I	C	Permanent pacing is recommended in ACHD and post operative high-grade second- or third-degree AVB that is not expected to resolve <i>[persisting 10 days]</i>



COR	LOE	Recommendation
I	B	Permanent pacing is recommended in adults with congenital AVB if associated to ventricular dysfunction, prolonged QTc interval, complex PVC, wide QRS escape rhythm, average daytime ventricular rate <50 bpm, ventricular pauses > 3-fold the cycle length of the underlying rhythm

Pacing – Should Be Discussed!

COR	LOE	Recommendation
IIa	C	Permanent pacing is reasonable for adults with complex CHD and an awake resting heart rate (sinus or junctional) <40bpm or ventricular pauses >3 seconds

COR	LOE	Recommendation
IIb	C	Permanent pacing may be reasonable in ACHD of moderate complexity and a awake resting heart rate (sinus or junctional) <40 bpm or ventricular pauses > 3 seconds
IIb	C	Permanent pacing may be considered in ACHD, a history of transient postoperative complete AVB, and residual bifascicular block



CRT

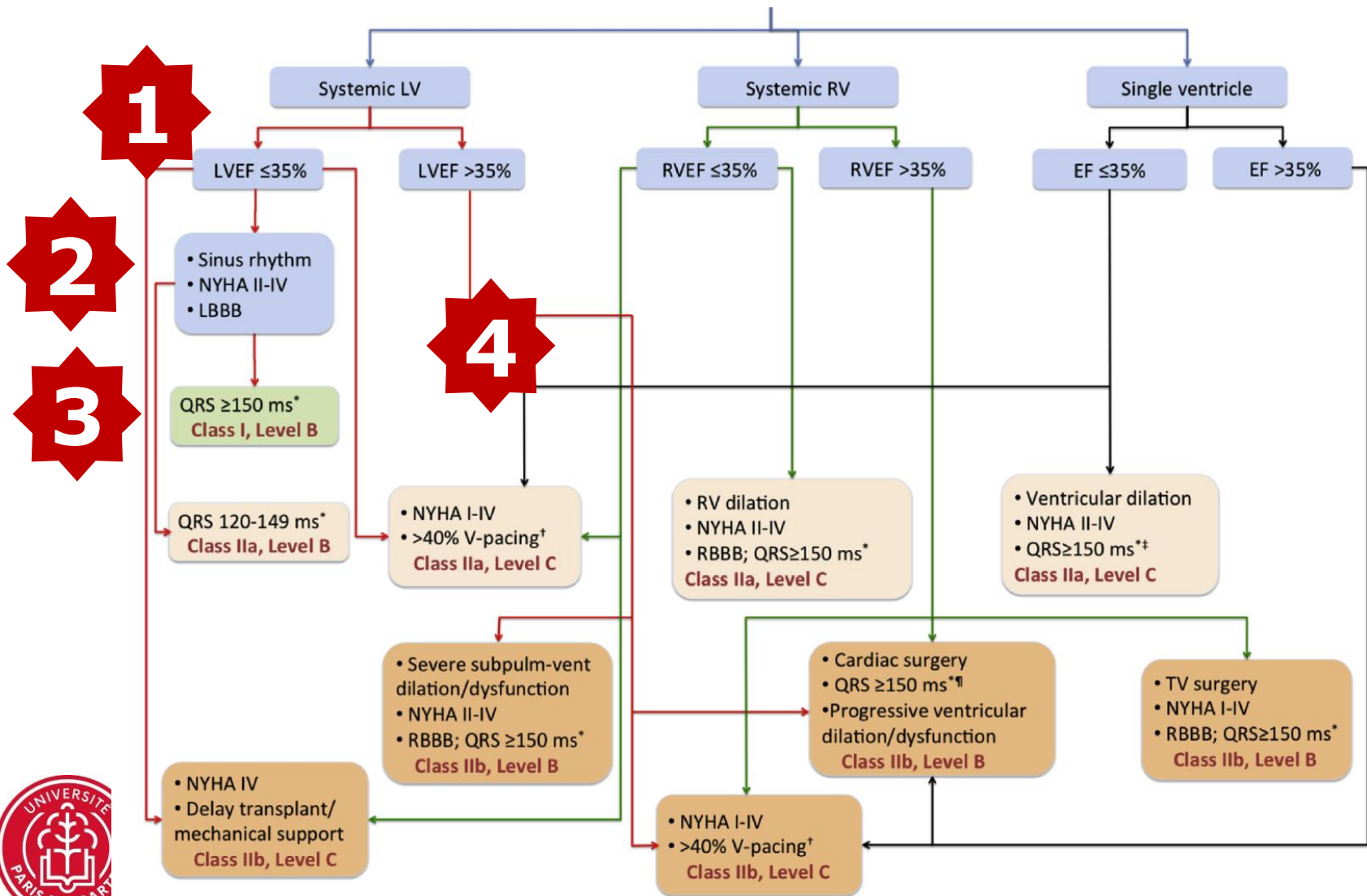


Demonstration of mechanical dyssynchrony is not a pre-requisite for CRT...

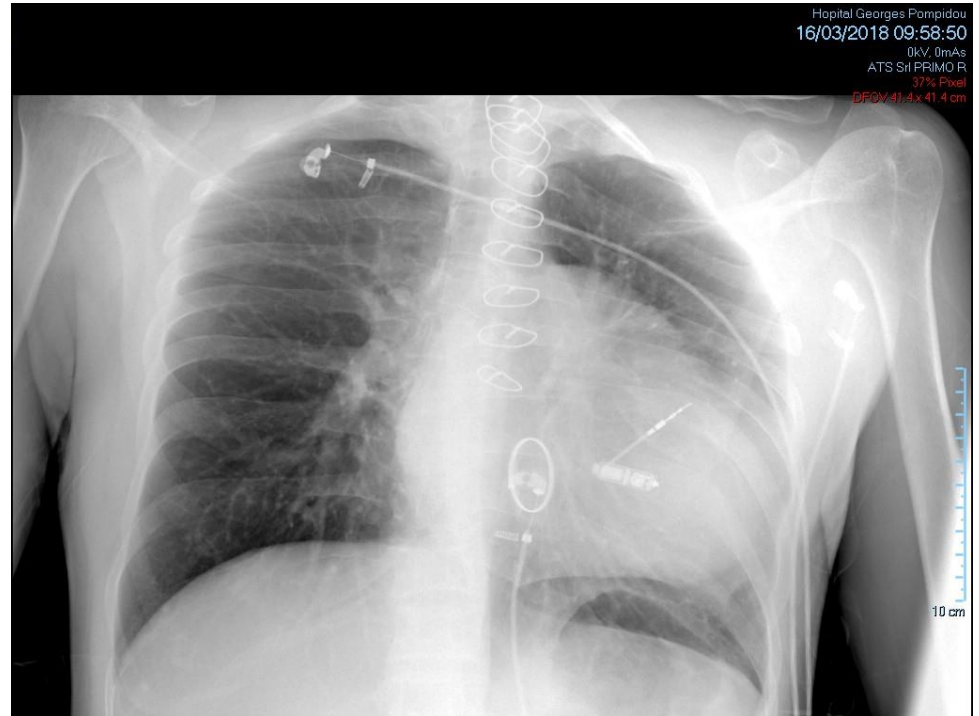
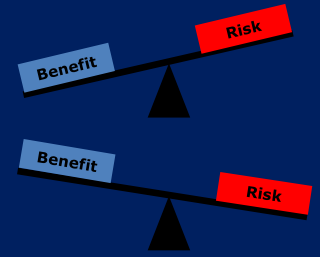
However, in which extent QRS duration is a good predictor of systemic ventricular dyssynchrony in ACHD given the diversity of structural and functional substrates (RBBB, systemic RV dysfunction, univentricular heart dysfunction...)

COR	LOE	Recommendation
III	B	CRT is not indicated in ACHD and a narrow QRS complex (<120 ms)
III	C	CRT is not indicated in ACHD whose co-morbidities and/or frailty limit survival with good functional capacity to less than 1 year

CRT Indication in ACHD

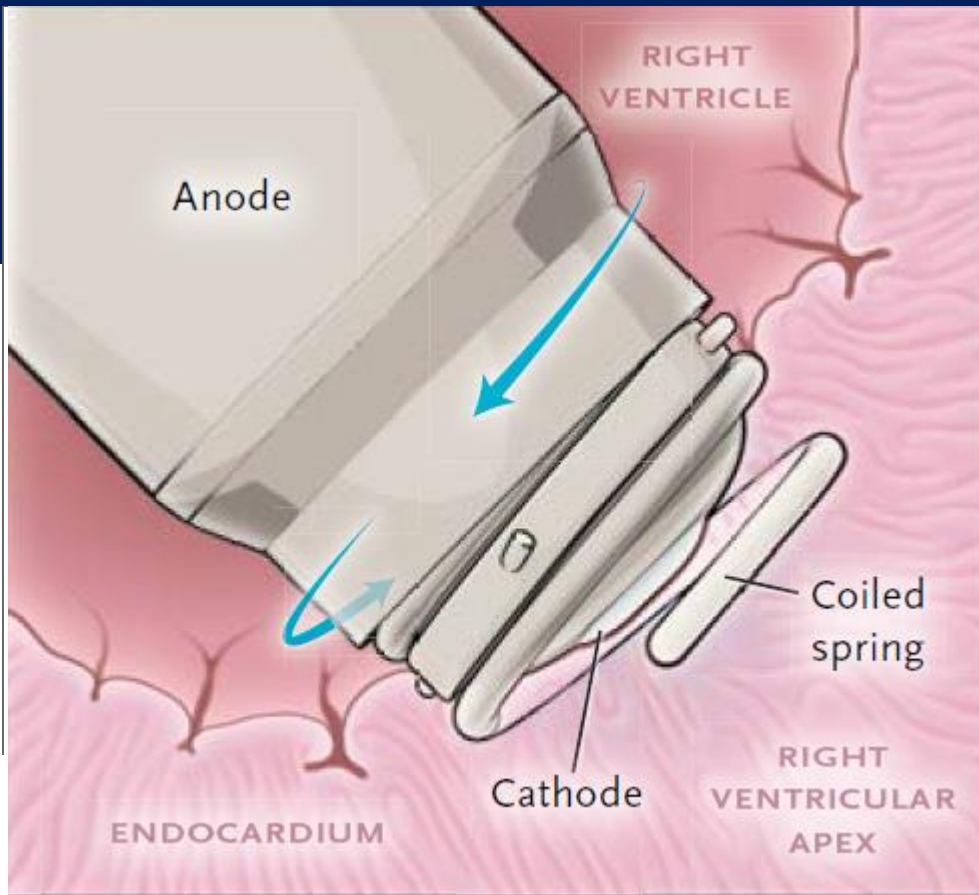


Leadless Pacing

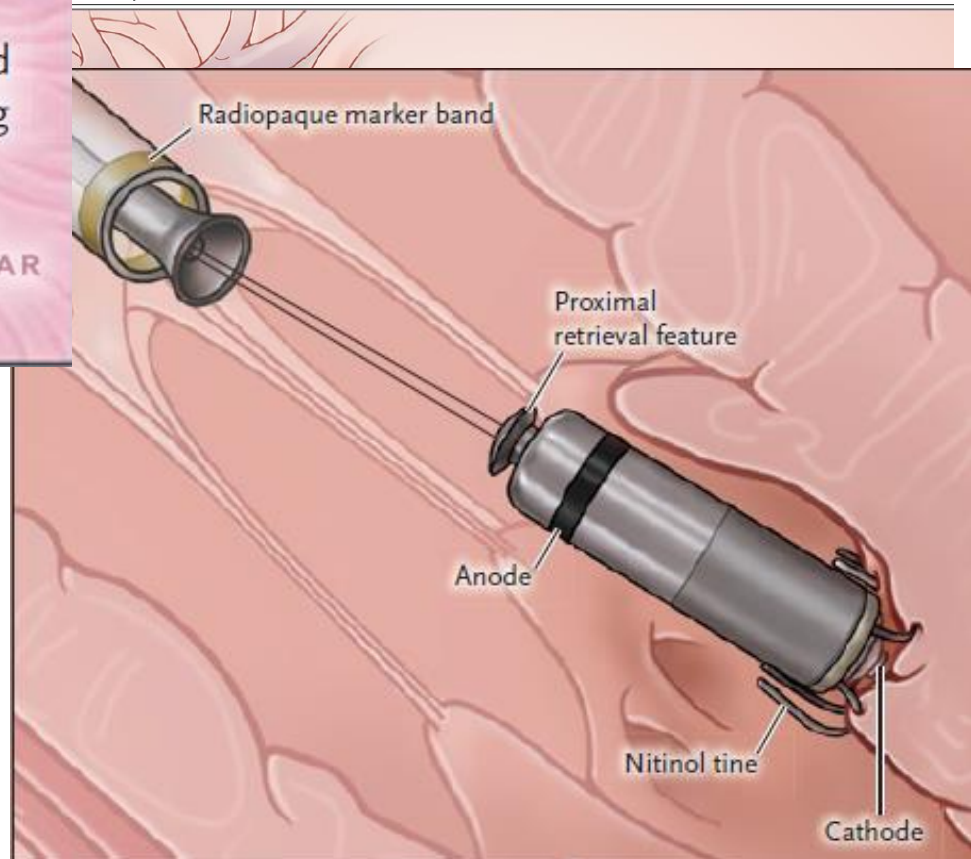


Leadless Pacing

**Micra
(Medtronic)
27 F**



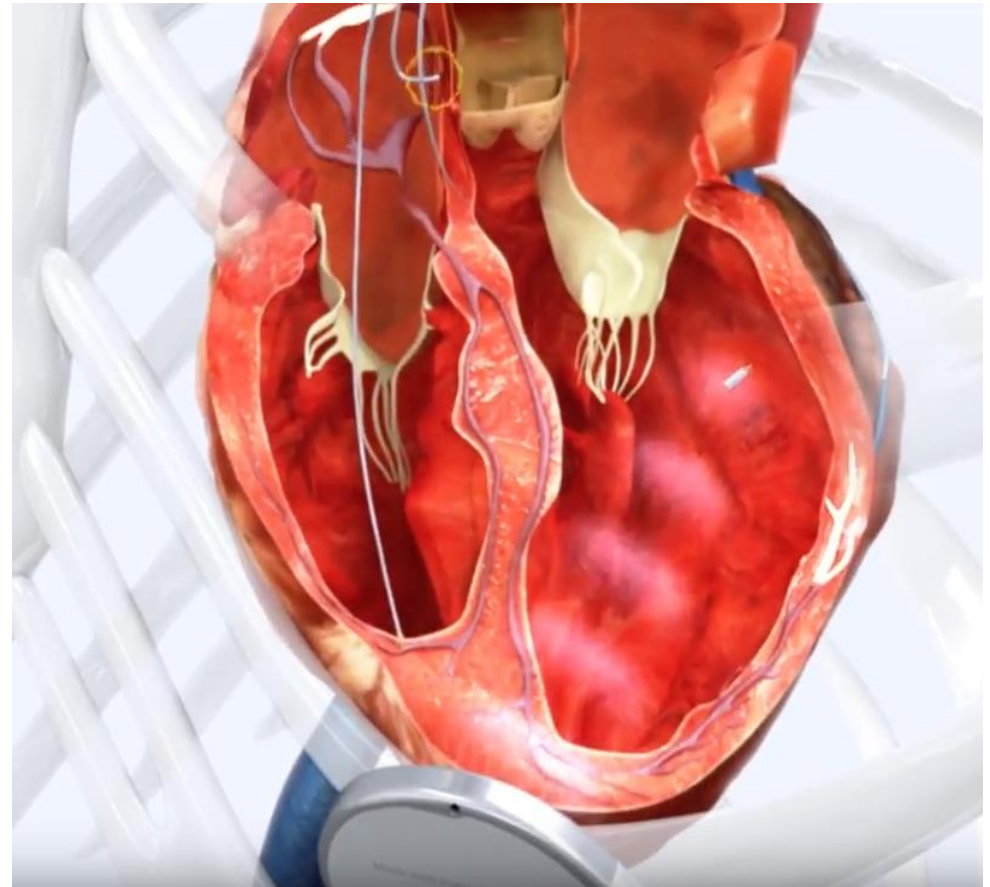
Nanostim (SJM) : 21F



**Eliminate: lead,
subcutaneous pocket...**

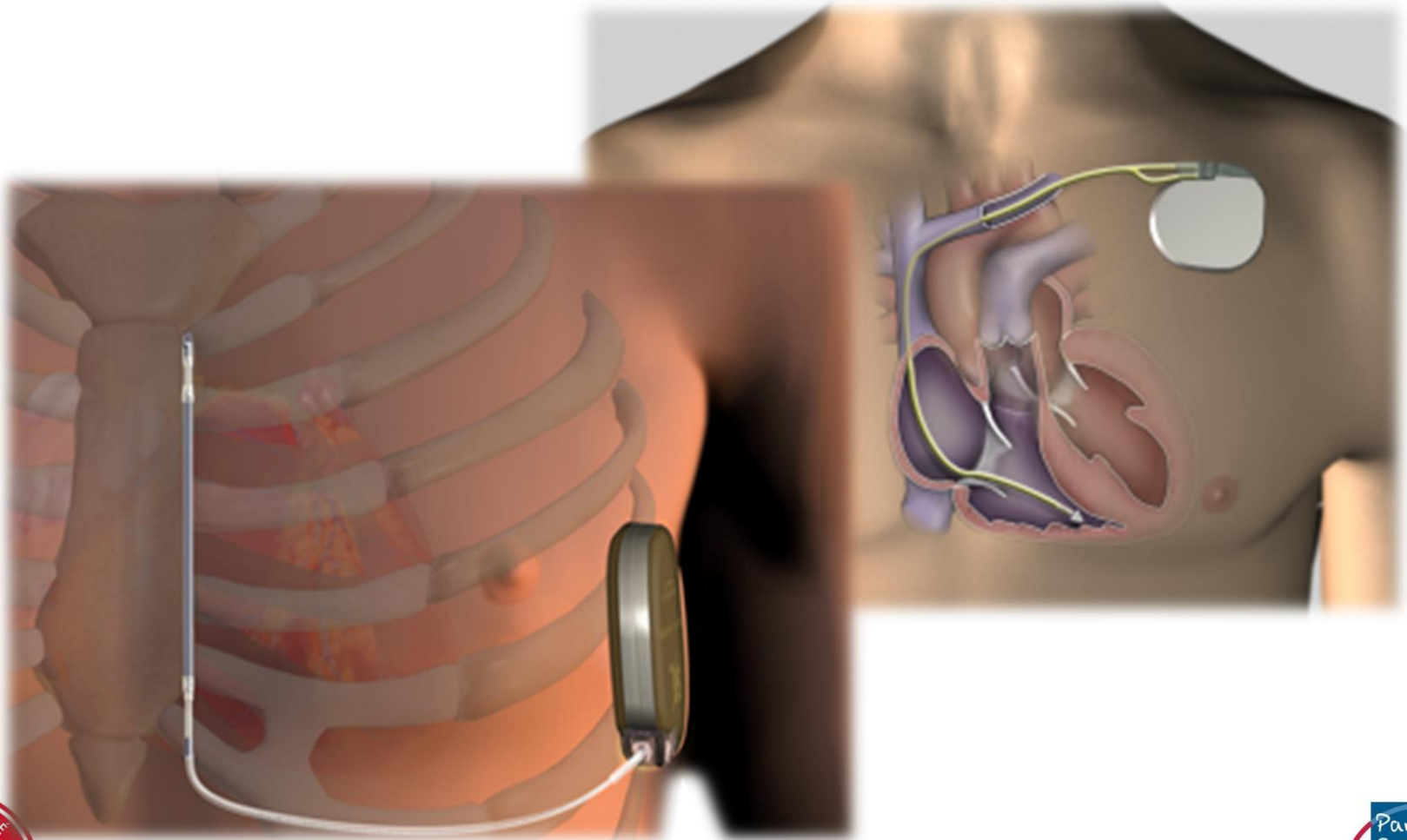
CRT With Wireless LV Endocardial Pacing

- Optimization of therapy (location)
- Control long-term complication...

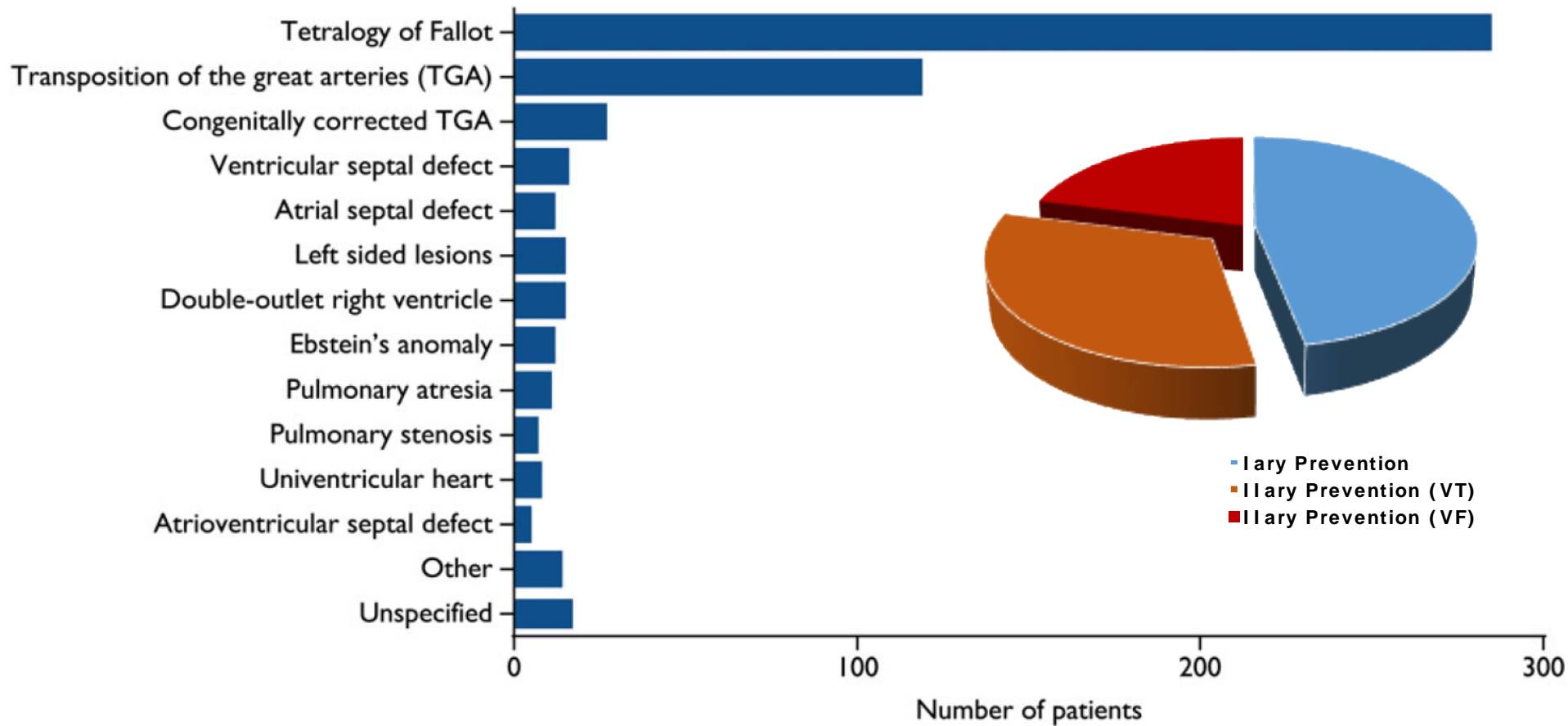


Reddy VY et al JACC 2017

Guidelines for ICD Therapy



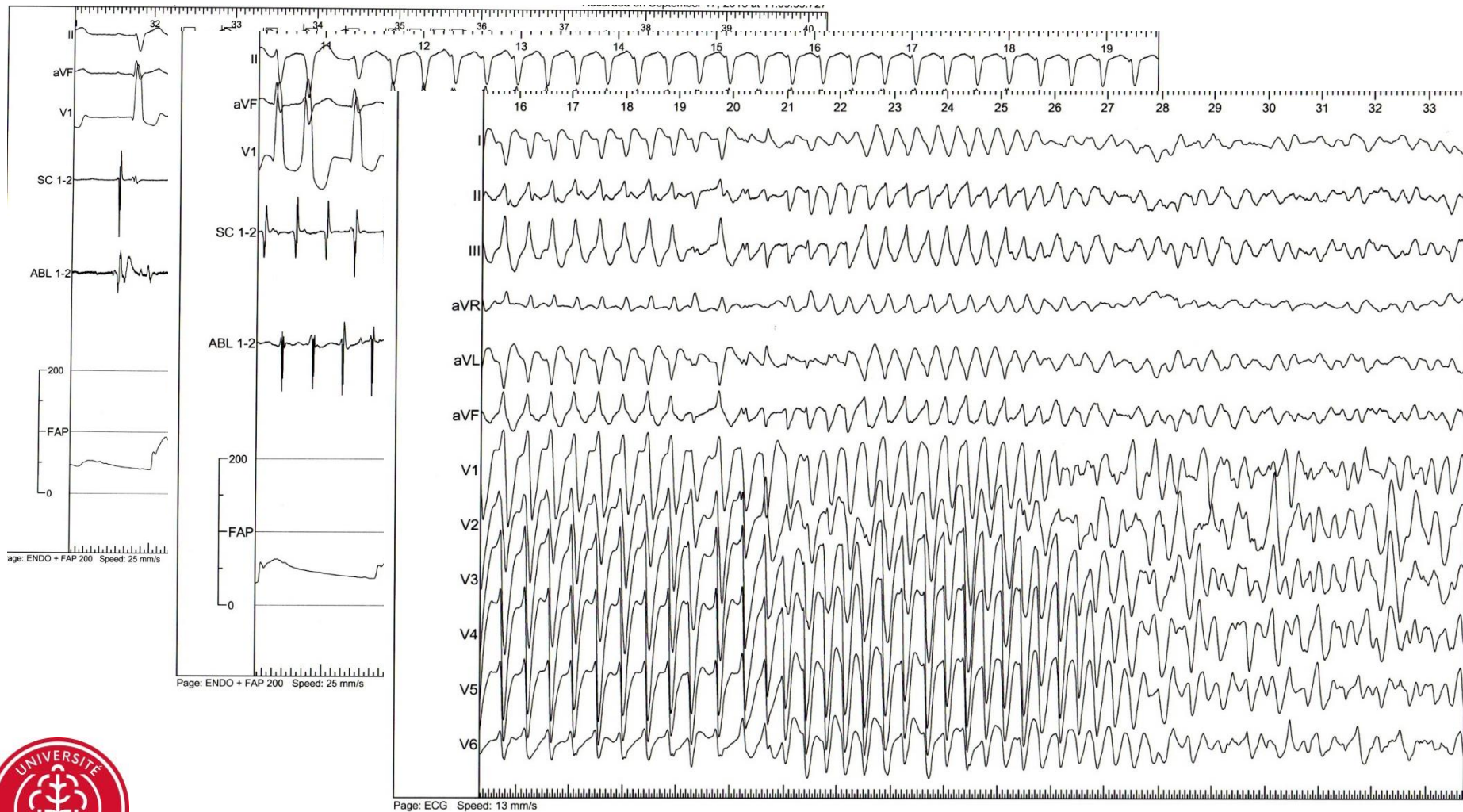
ICD in ACHD



Vehmeijer JT et al Eur Heart J 2016



ICD Complications & Bonus!



Courtesy Dr Nicolas Combes, CP, Toulouse



ICD – Not Recommended

COR	LOE	Recommendation
III	C	All Class III recommendations in adults w/o CHD: <ul style="list-style-type: none">–Life expectancy with acceptable functional status <1 year–Incessant VT/VF–Significant psychiatric illness that may be aggravated by ICD implantation or preclude systematic FU–Drug-refractory NYHA IV symptoms who are not candidates for cardiac transplantation or CRT
III	B	ACHD and advanced pulmonary vascular disease (Eisenmenger syndrome) are generally not considered candidates for ICD therapy
III	B	Endocardial leads are avoided in ACHD and intracardiac shunts .



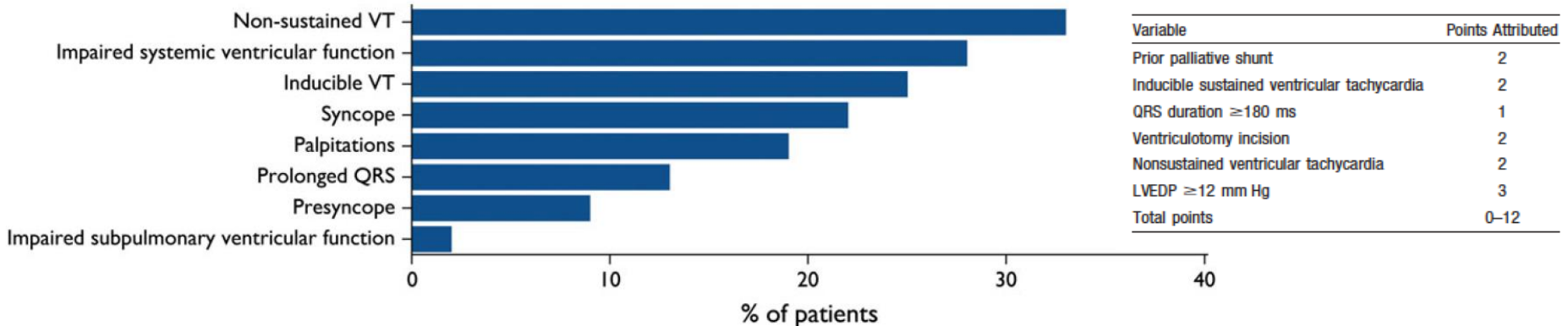
ICD – Recommended

COR	LOE	Recommendation
I	B	ICD therapy is indicated in ACHD who are survivors of cardiac arrest due to VF or hemodynamically unstable ventricular tachycardia after evaluation to define the cause of the event and <u>exclude any completely reversible etiology</u>
I	B	ICD therapy is indicated in ACHD and spontaneous sustained VT who have undergone hemodynamic and electrophysiologic evaluation.
I	B	ICD therapy is indicated in ACHD and a systemic left ventricular EF $\leq 35\%$, biventricular physiology, and NYHA II or III symptoms



ICD – Should Be Discussed!

COR	LOE	Recommendation
Ia	B	ICD is reasonable in selected adults with tetralogy of Fallot and multiple risk factors for SCD , such as left ventricular dysfunction, NSVT, QRS duration ≥ 180 ms, extensive right ventricular scarring, or inducible SVT at EPS



Vehmeijer JT et al Eur Heart J 2016
Khairy P et al. Circulation 2008



ICD – Should Be Discussed!

COR	LOE	Recommendation
IIb	C	ICD may be reasonable in adults with single or systemic right ventricular EF <35%, particularly in the presence of additional risk factors such as complex ventricular arrhythmias, unexplained syncope, NYHA II or III, QRS duration \geq 140ms, or severe systemic AV valve regurgitation
IIb	C	ICD may be considered in ACHD and syncope of unknown origin with hemodynamically significant sustained VT or VF inducible at EPS
IIb	C	ICD may be considered for non-hospitalized ACHD awaiting heart transplantation

Risk Stratification / Phenotype

- French DAI-T4F Registry
- DAI-PP NetWork



Registre DAI-T4F

CNIL # 2029070



Présentation

Comme vous le savez, il existe depuis 2010 un registre national incluant l'ensemble des patients avec une tétralogie de Fallot et porteur d'un défibrillateur implantés à partir de 2005. Ce registre, initialement limité à 17 centres, s'est étendu à plus de 50 centres, et a pour ambition de s'étendre à tous les centres autorisés à implanter des défibrillateurs en France. Cette collection de données a pour objectif de faire une synthèse des caractéristiques à l'implantation et des événements à moyen et long terme dans cette population bien particulière. Nous avons d'ores et déjà inclus 102 patients, aussi bien en prévention primaire que secondaire, avec un suivi moyen de 5.5 ans. L'actualisation du suivi se fait chaque année au cours du 1^{er} trimestre.

Promoteur

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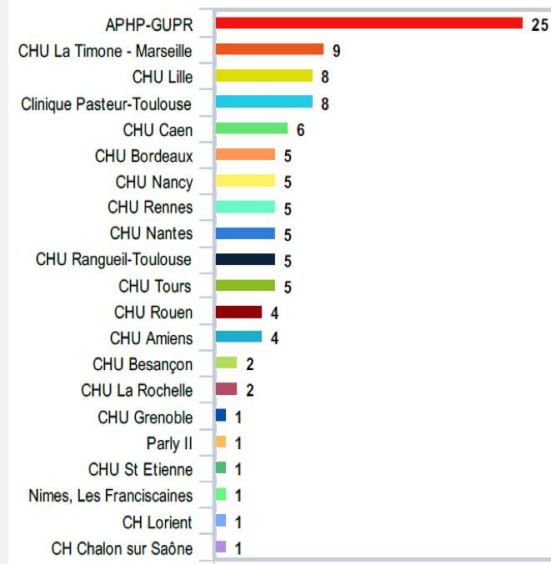
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Télécharger le CRF : Cliquez
ici [DAI-T4F](#)

Télécharger l'abstract :
Cliquez ici [ESC 2017](#)
Aperçu de la [Base](#)

INCLUSIONS DES PATIENTS PAR CENTRES MAI 2017



Nous aimerions former un groupe de travail pour participer à l'analyse des données parmi les investigateurs les plus investis dans cette thématique. N'hésitez pas à nous contacter. Une réunion sera prévue à Europeace.

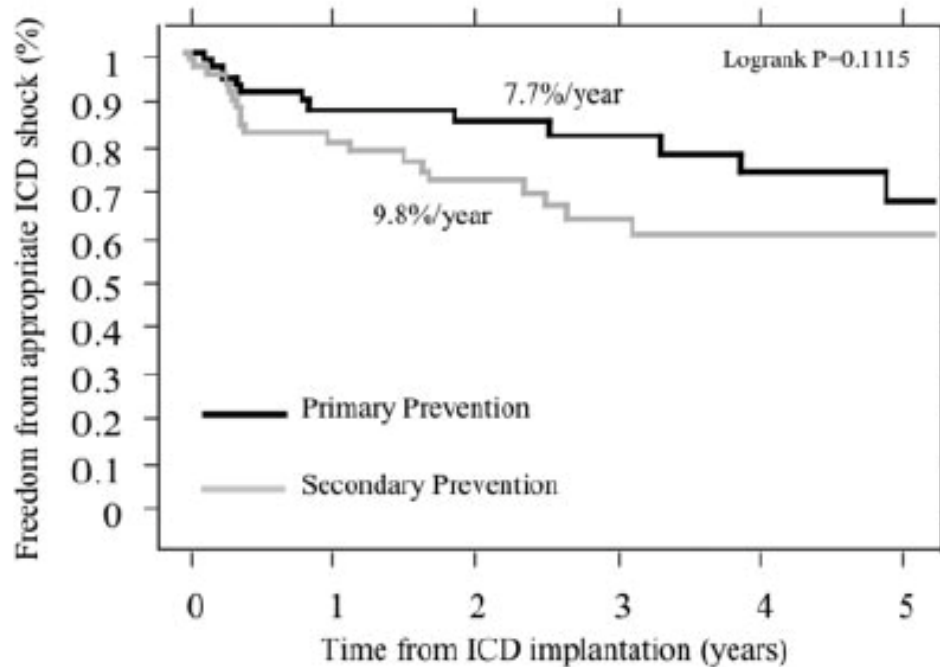
Implantable Cardioverter-Defibrillators in Tetralogy of Fallot

Paul Khairy, MD, PhD; Louise Harris, MD; Michael J. Landzberg, MD;

Sangeetha Viswanathan, MRCPCH; Amanda Barlow, MD; Michael A. Gatzoulis, MD;

Susan M. Fernandes, MHP, PA-C; Luc Beauchesne, MD; Judith Therrien, MD; Philippe Chetaille, MD;

Elaine Gordon, MD; Isabelle Vonder Muhll, MD; Frank Cecchin, MD



- Prévention I^{aire} :

- 23.5% chocs appropriés
- 7.7% par an
- 3.3% par an FV/TV polymorphe

- Prévention II^{aire} :

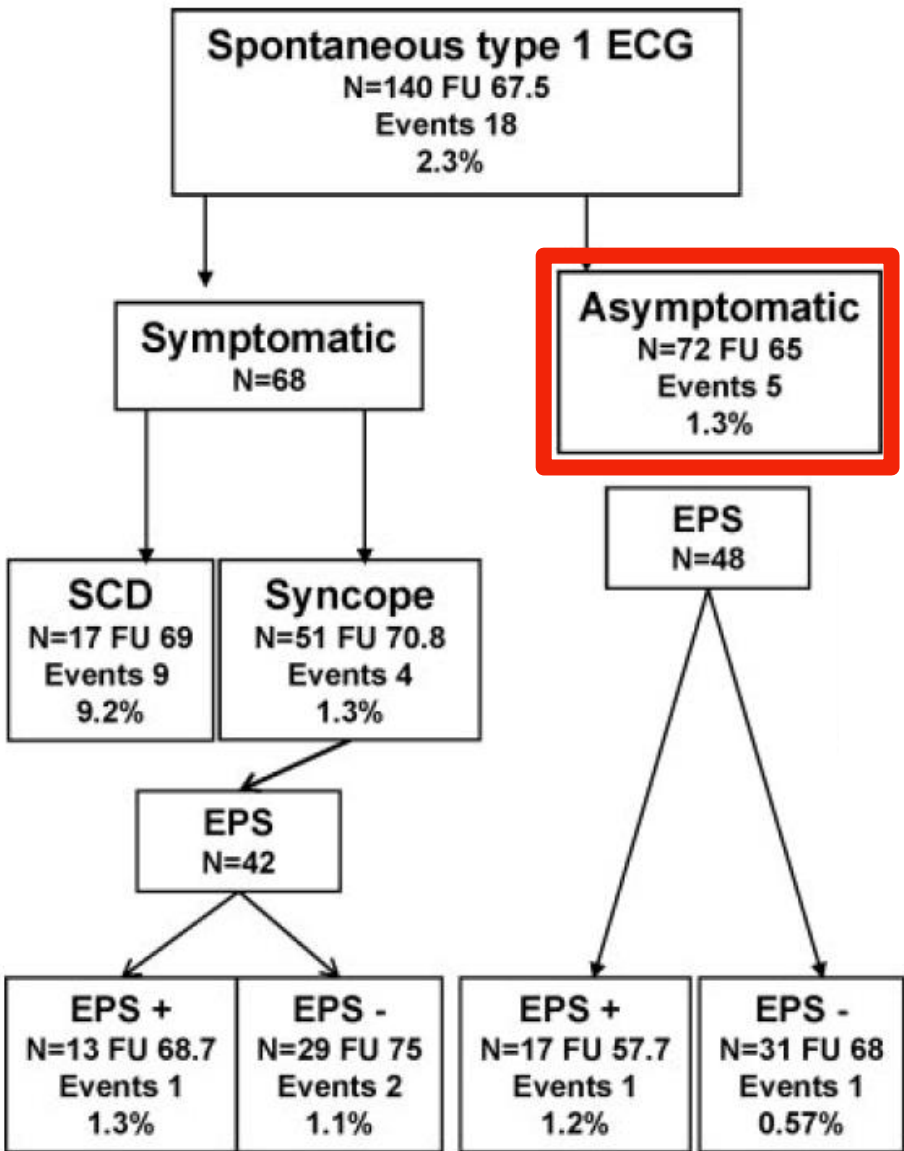
- 30.2% chocs appropriés
- 9.8% par an
- 4.2% par an FV/TV polymorphe

Khairy P et al. Circulation 2008



FINGER Registry

(1029 pts)



ICD implantation is recommended in patients with a diagnosis of Brugada syndrome who (a) Are survivors of an aborted cardiac arrest and/or (b) Have documented spontaneous sustained VT.	I	C
ICD implantation should be considered in patients with a spontaneous diagnostic type I ECG pattern and history of syncope.	IIa	C

Probst et al. Circulation 2010





HCM Risk-SCD Calculator

Age Years

Age at evaluation

Maximum LV wall thickness mm

Transthoracic Echocardiographic measurement

Left atrial size mm

Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation

Max LVOT gradient mmHg

The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernoulli equation: Gradient = $4V^2$, where V is the peak aortic outflow velocity

Family History of SCD No Yes

History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).

Non-sustained VT No Yes

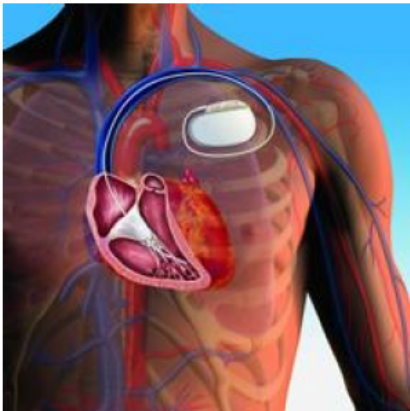
3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring

Risk of SCD at 5 years (%):

ESC recommendation:

S-ICD

TV-ICD



Documented need
Pacing or ATP

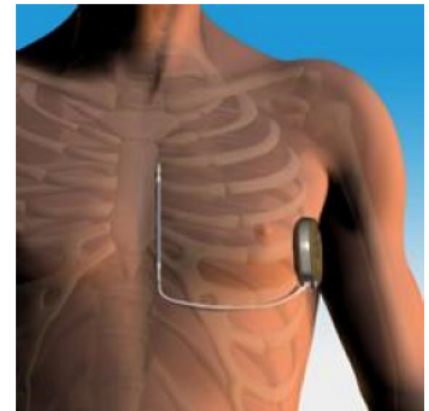


mCRM™ System*

EMBLEM™ S-ICD
EMPOWER™ Modular Pacing
System*

Potential need
for Pacing or ATP

S-ICD

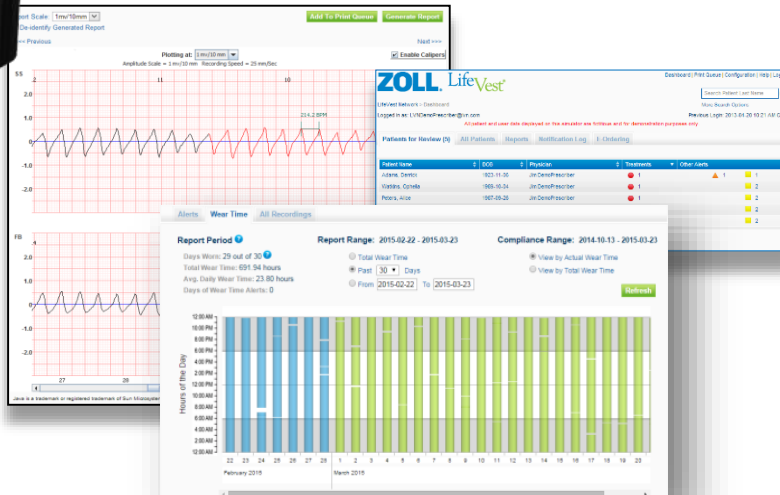


No need
for Pacing or ATP

Defibrillation for Transient Risk!!



Wearable Cardioverter Defibrillator?



Wear Time / Inappropriate Therapies



Wear Time™ Summary

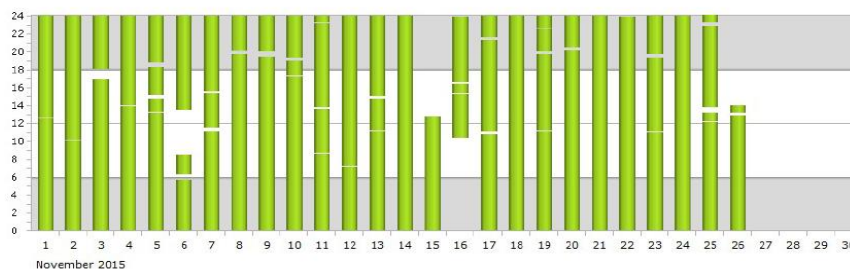
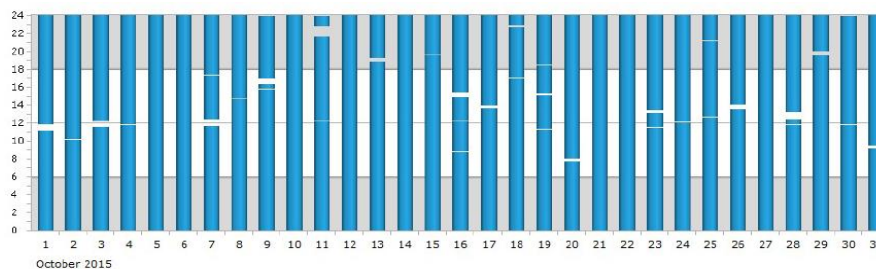
Total Days of Patient Use: 68 days

Total Patient Use Percent: 97.00%

Average Daily Patient Use:: 23.30 hours

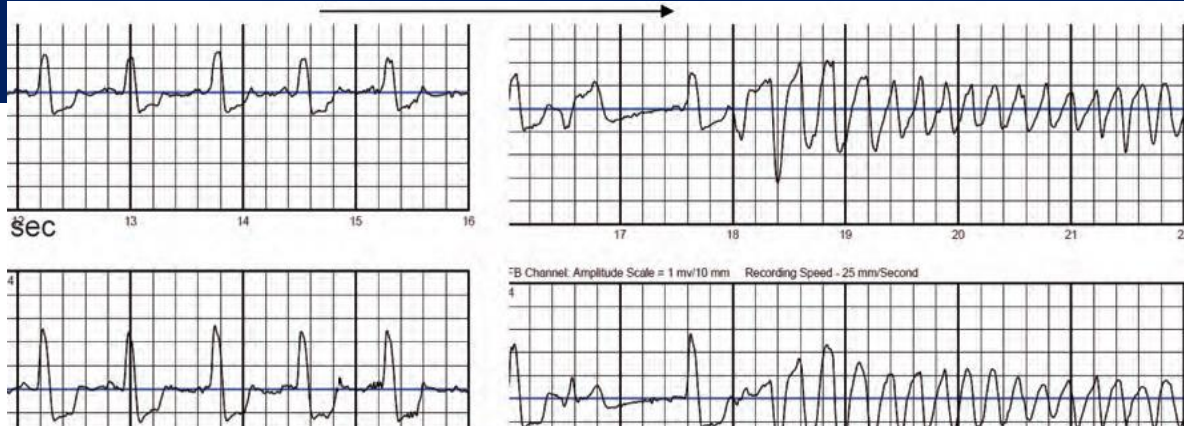
Number of Wear Time™ Alerts: 0

- Daily: 22.5h
- Total days: 90d
- Inappropriate therapies: 0.5% of patients

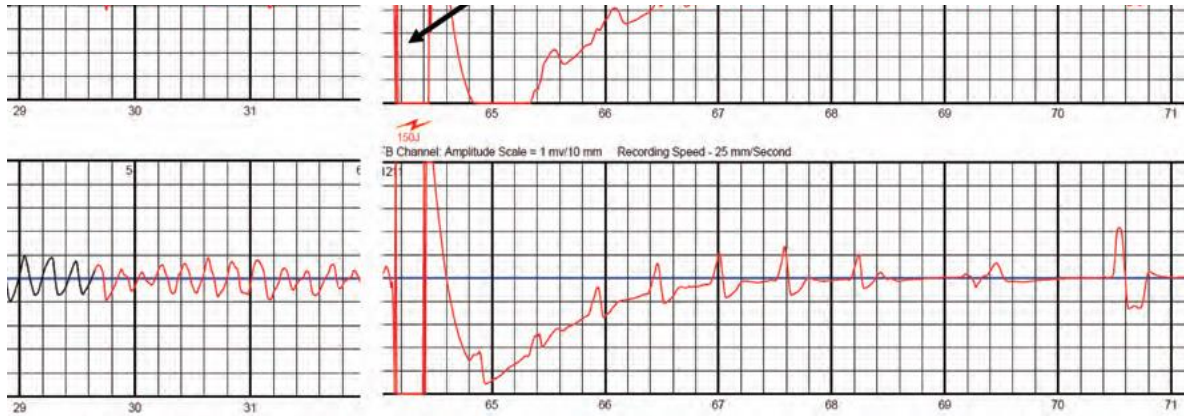


- Daily: 23.1h
- Total days: 59d
- Inappropriate therapies: 0.4% of patients





75 % Shock between 0-60 seconds ?? %
100 % Successful shock conversion 94 %



Life-Vest Indications

Recommendation	Class ^a	Level ^b
The WCD may be considered for adult patients with poor LV systolic function who are at risk of sudden arrhythmic death for a limited period, but are not candidates for an implantable defibrillator (e.g. bridge to transplant, bridge to transvenous implant, peripartum cardiomyopathy, active myocarditis and arrhythmias in the early post-myocardial infarction phase).	IIb	C

4 indications



- **Après explantation d'un système de défibrillation implantable** pour infection, de la loge ou des électrodes, jusqu'à la réimplantation (guérison de l'infection).
- **En attente de transplantation cardiaque.** L'indication doit être réévaluée tous les 3 mois (évaluation du rapport bénéfices/risques et de l'observance).
- **Après revascularisation myocardique si la fraction d'éjection ventriculaire gauche (FEVG) est inférieure à 30 %**, jusqu'à la réévaluation de la FEVG et discussion de l'indication d'un défibrillateur automatique implantable au 3^{ème} mois.
- **Au décours d'un infarctus du myocarde aigu si la FEVG est inférieure à 30 % après les 48 premières heures**, jusqu'à la réévaluation de la FEVG et discussion de l'indication d'un défibrillateur automatique implantable au terme du 1^{er} mois.



Take-Home Messages

- **Guidelines to help...**
- **Common challenges in arrhythmia devices for ACHD**
 - Selecting appropriate candidates (CRT/ICD)
 - Benefit/risk balance
- **Creative options hopefully**
 - Subcutaneous ICD
 - WCD
 - Leading pacing



Thank You!



Paris Sudden Death Expertise Center

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Paris Cardiovascular Research Center
56, rue Leblanc - 75015 Paris, France

