



RHYTHM 2017

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New Insights & Technological Advances

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Novotel PARIS Tour Eiffel



Implantable Cardiac Defibrillator in the Setting of Tetralogy of Fallot

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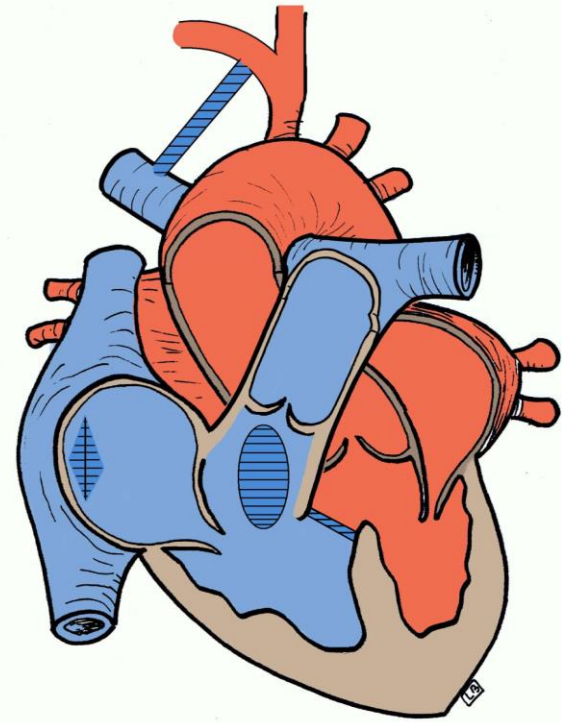
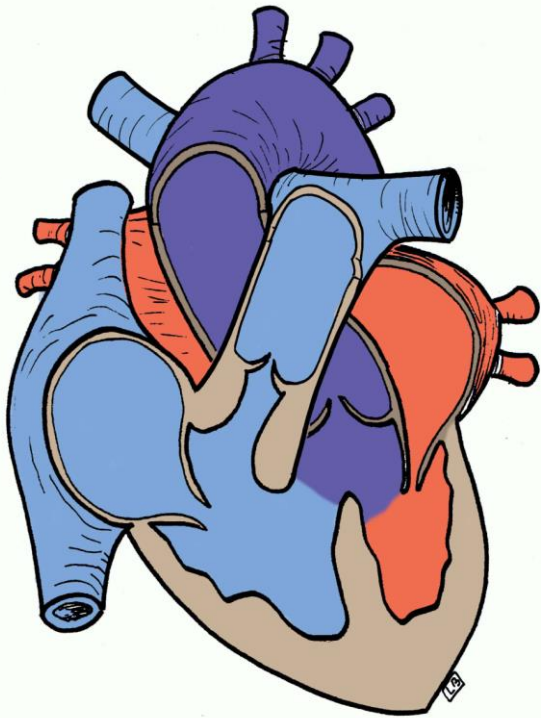
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Disclosure

I do not have any potential conflict of interest





A Population-Based Prospective Evaluation of Risk of Sudden Cardiac Death After Operation for Common Congenital Heart Defects

	No. (%) of Pts With Complete Follow-Up	Total Follow-Up (pt-yr)	Sudden Cardiac Death		Nonsudden Cardiac Death	
			No.	Incidence/ 1,000 Pt-yr	No.	Incidence/ 1,000 Pt-yr
ASD	622 (86%)	7,904	0	0	0	0
VSD	527 (87%)	6,354	1	0.2	8	1.2
AVSD	254 (87%)	2,217	2	0.9	15	6.7
PDA	623 (82%)	8,753	0	0	4	0.4
PS	241 (91%)	3,568	1	0.3	2	0.6
AS	169 (94%)	1,860	10	5.4	9	4.8
CoA	536 (92%)	6,706	9	1.3	17	2.5
TOF	445 (91%)	7,082	11	1.5	9	1.3
d-TGA	172 (95%)	1,413	7	4.9	10	6.9
Total	3,589	45,857	41	0.9	74	1.6

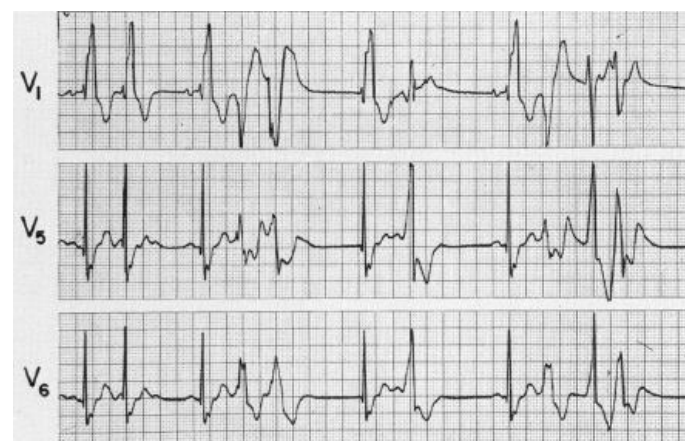
Conclusions. The risk of late sudden death for patients surviving operation for common congenital heart defects is 25 to 100 times greater than an age-matched control population. This

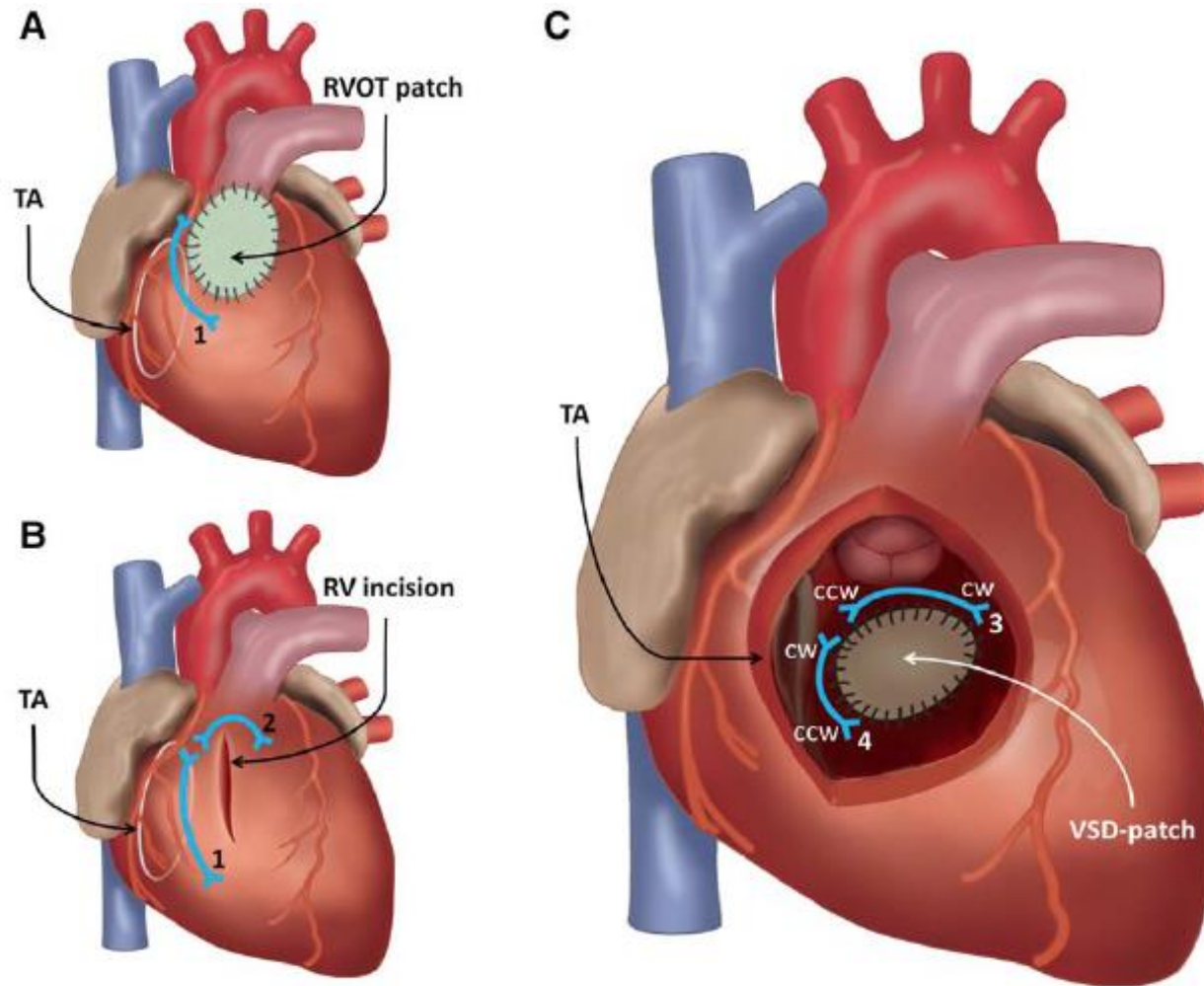
Unexpected Cardiac Arrest in Patients after Surgical Correction of Tetralogy of Fallot

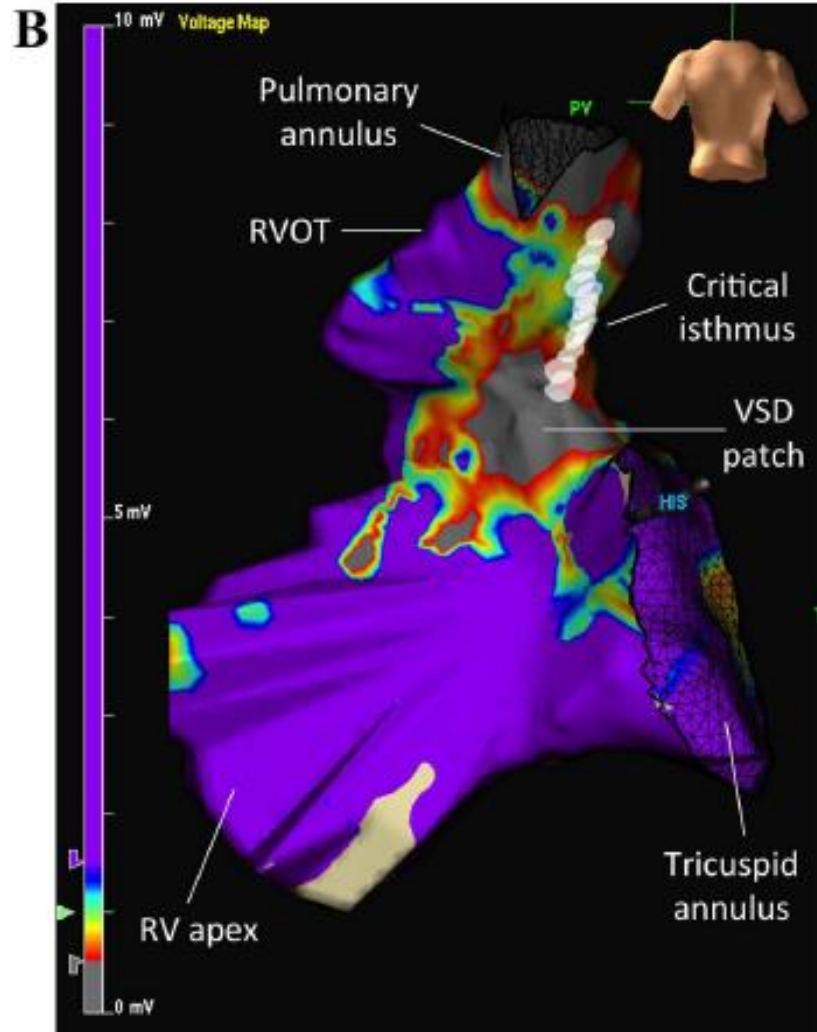
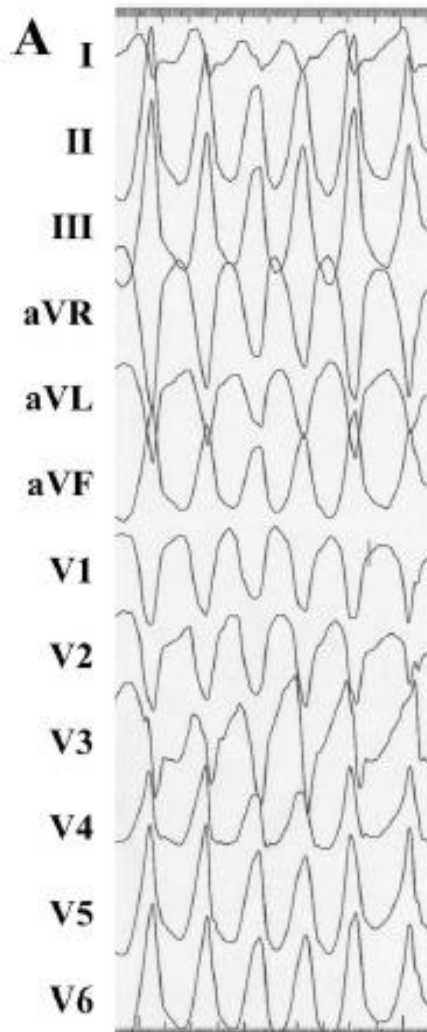
By FREDERICK W. JAMES, M.D., SAMUEL KAPLAN, M.D., AND TE-CHUAN CHOU, M.D.

Circulation, Volume 52, October 1975

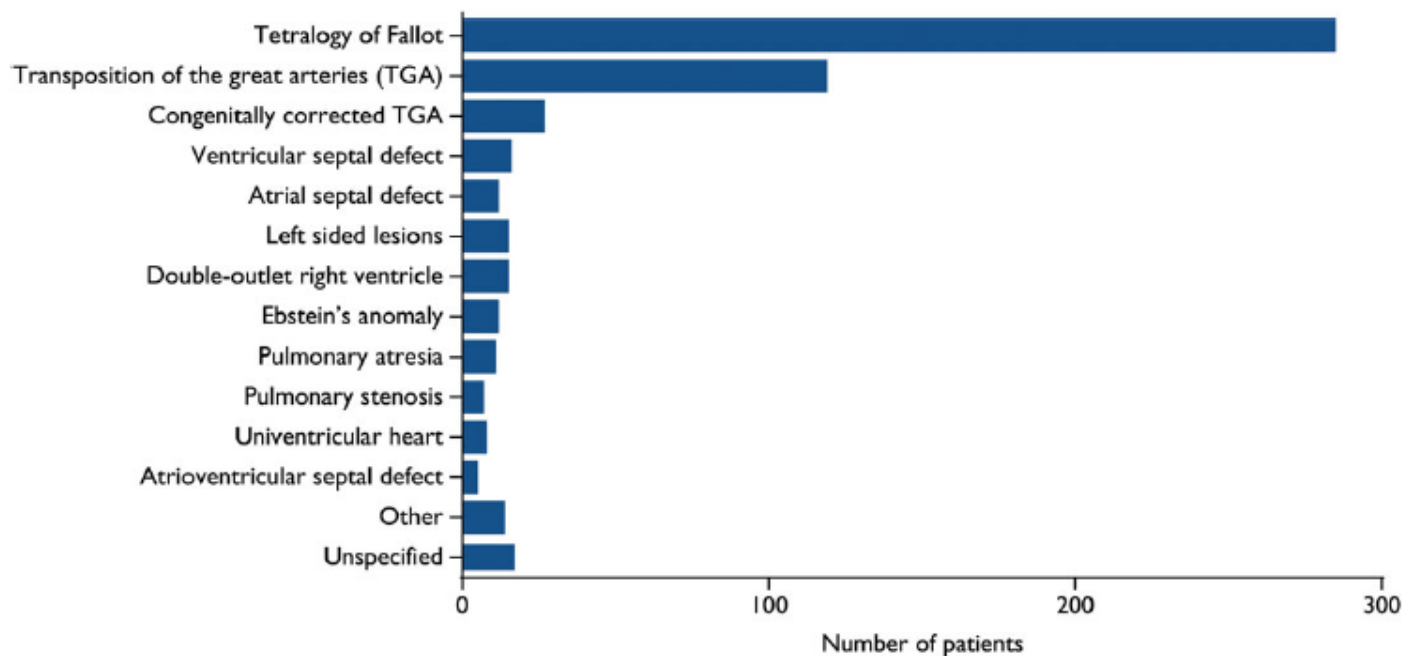
Although complete heart block cannot be excluded in these four patients, we reasoned that the cardiac arrests were probably preceded by ventricular tachyarrhythmia. Because of this experience, we believe that any patient who has had intraventricular surgery should be evaluated for ventricular arrhythmia. If frequent premature ventricular contractions or serious ventricular arrhythmias are documented, we seriously consider antiarrhythmic therapy in an attempt to prevent ventricular tachyarrhythmias and sudden death.



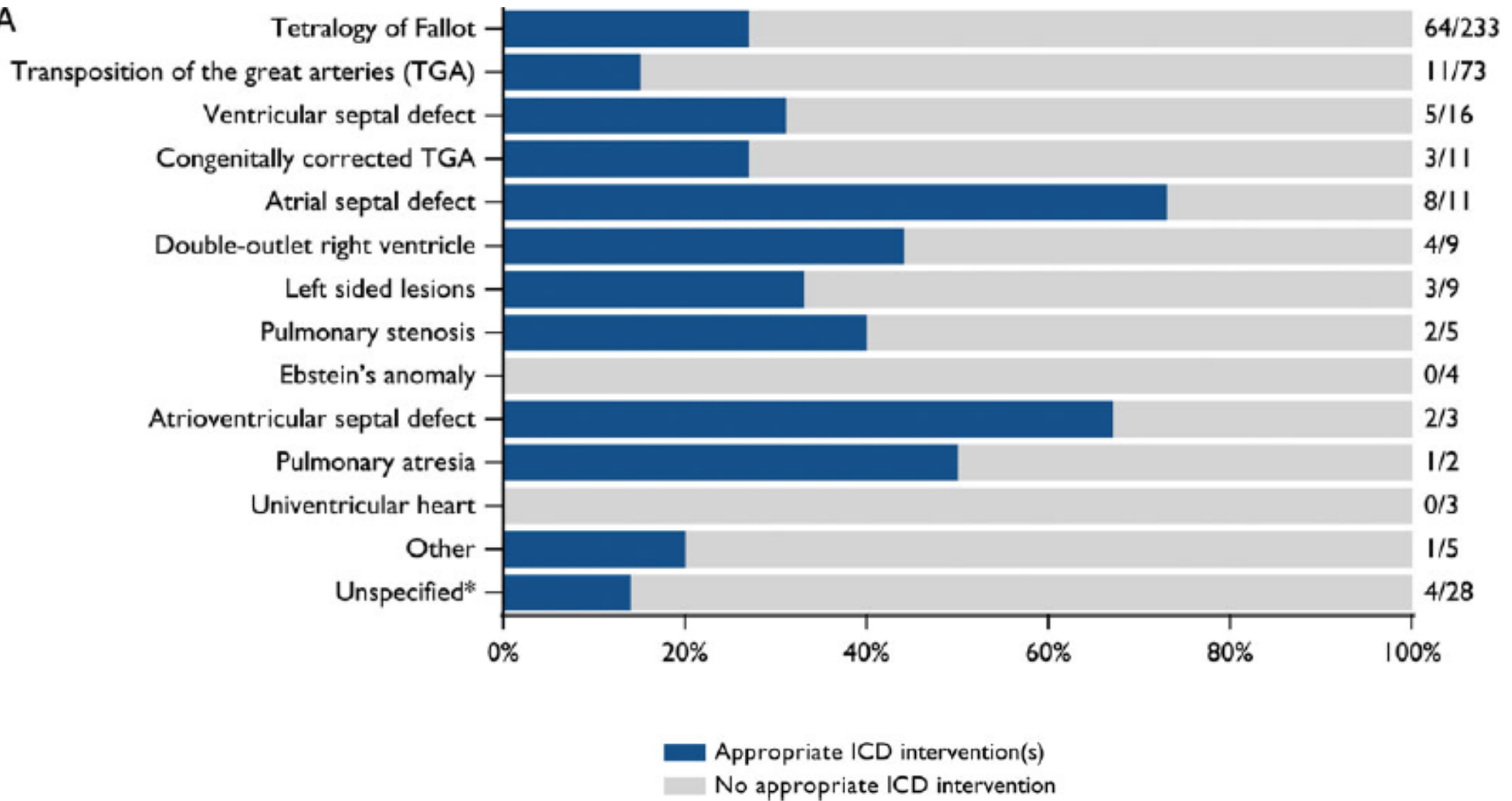




- **Tetralogy of Fallot (TOF)** is the most frequent form of congenital heart disease
- Sudden cardiac death (SCD) is the most common cause of mortality after late repair (0.2 to 0.3%/year)
- TOF is the most frequent form of congenital heart disease managed by EP physicians for potential implantable cardioverter defibrillator (ICD)



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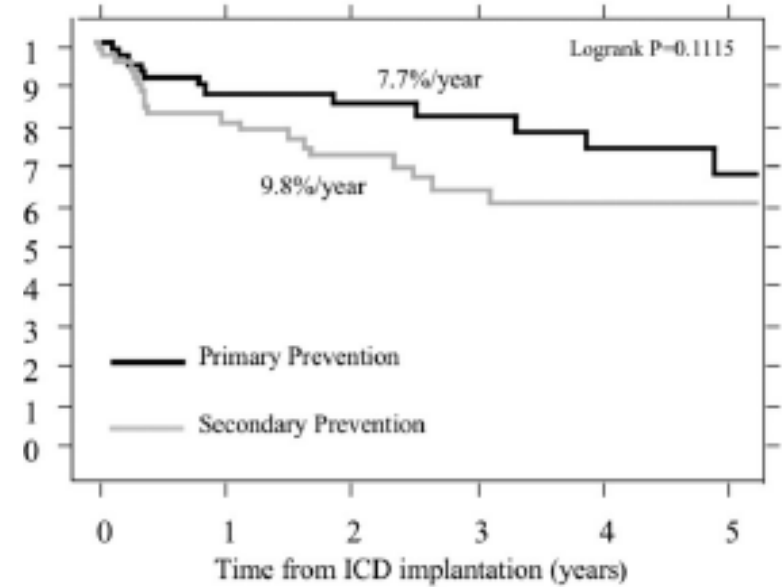
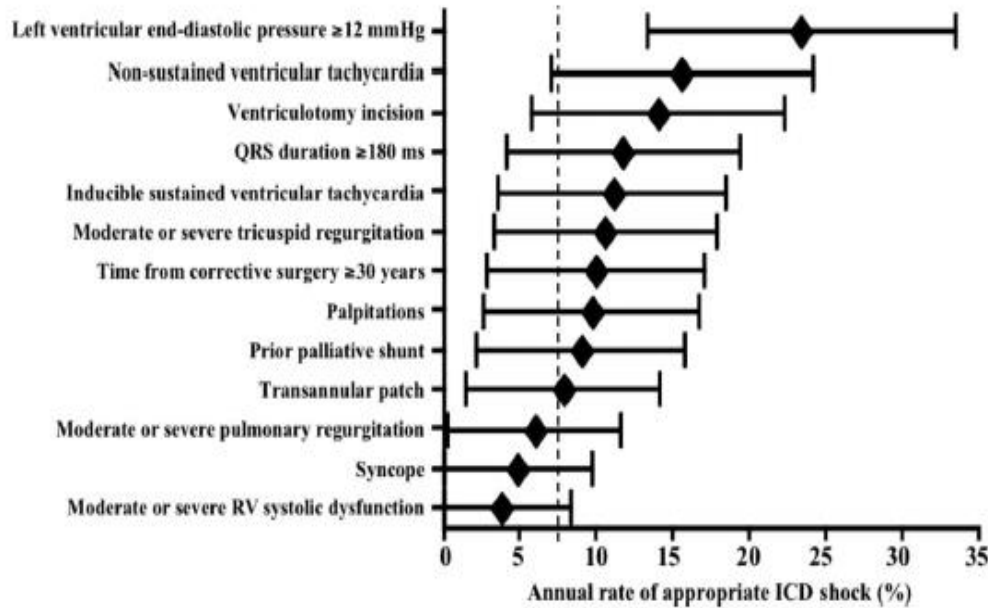
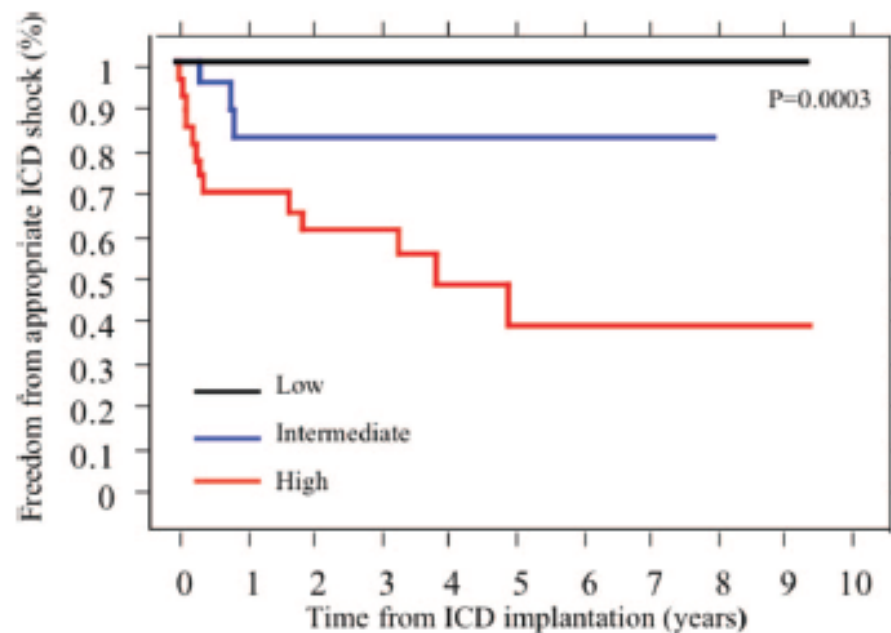
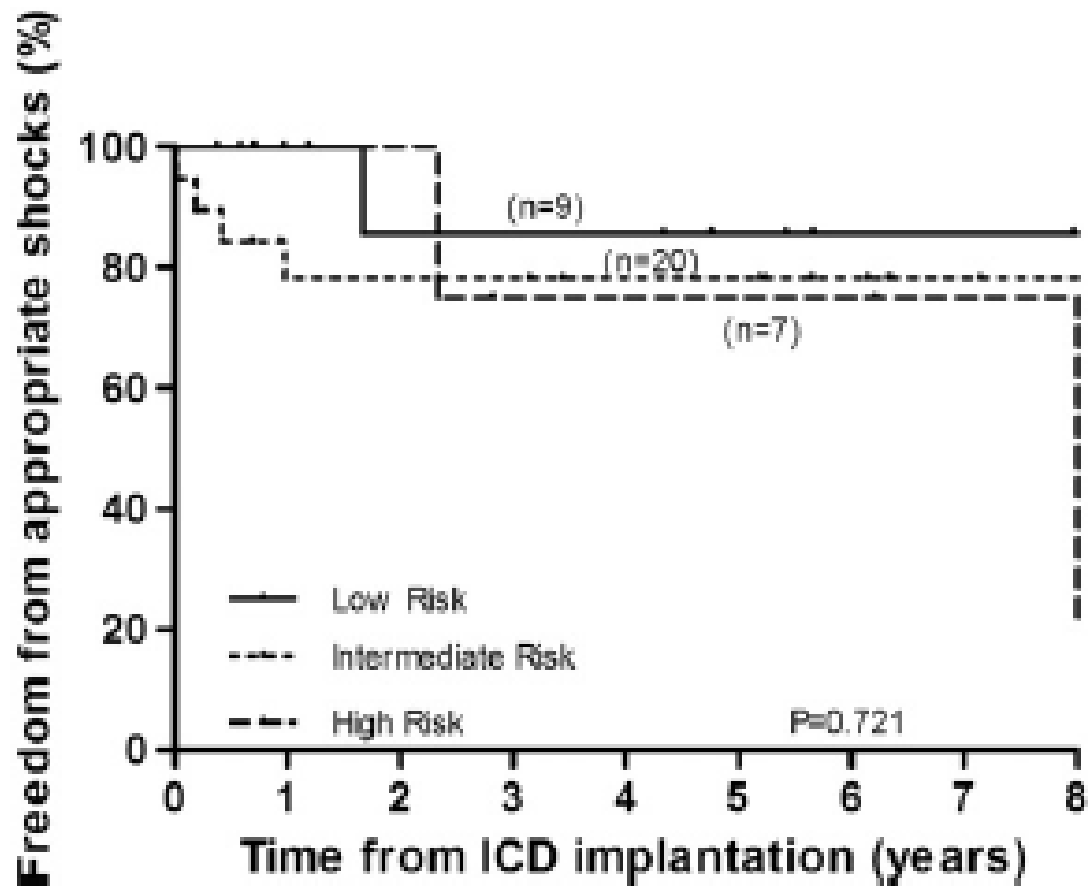


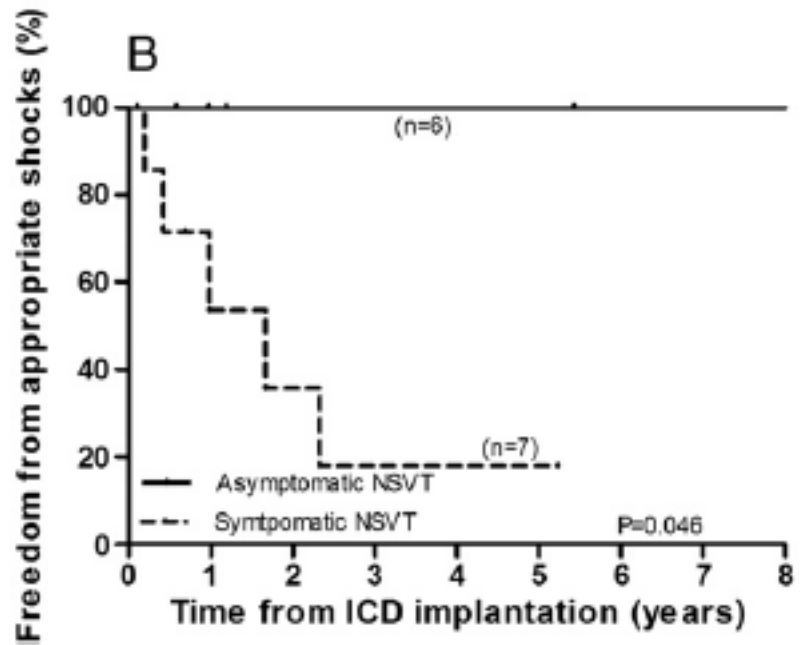
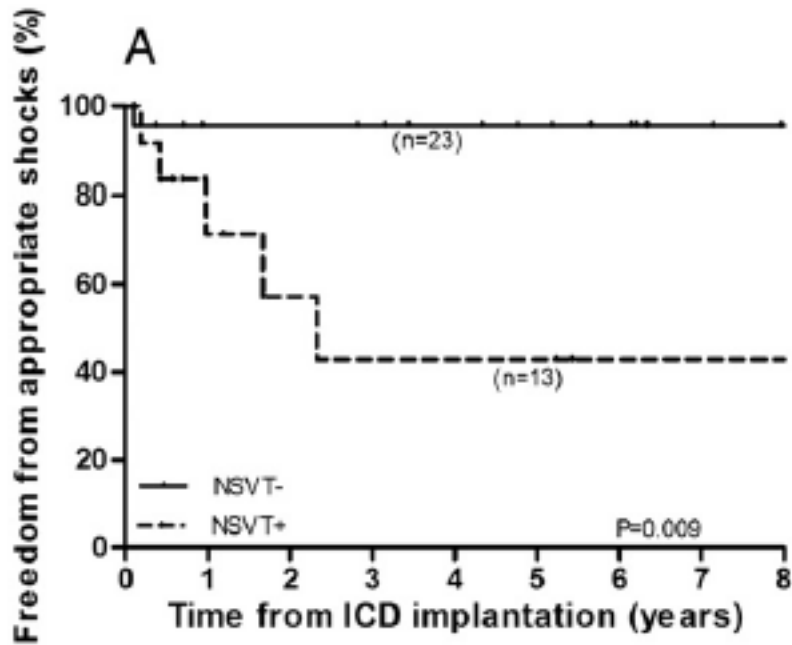
Table 3. Risk Score for Appropriate ICD Shocks in Primary Prevention

Variable	Exp(β)	Points Attributed
Prior palliative shunt	3.2	2
Inducible sustained ventricular tachycardia	2.6	2
QRS duration \geq 180 ms	1.4	1
Ventriculotomy incision	3.4	2
Nonsustained ventricular tachycardia	3.7	2
LVEDP \geq 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%







PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease

- Class I
1. ICD therapy is indicated in adults with CHD who are survivors of *cardiac arrest* due to ventricular fibrillation or hemodynamically unstable ventricular tachycardia after evaluation to define the cause of the event and exclude any completely reversible etiology (*Level of evidence: B*).^{40,46,460-462}
 2. ICD therapy is indicated in adults with CHD and *spontaneous sustained ventricular tachycardia* who have undergone hemodynamic and electrophysiologic evaluation (*Level of evidence: B*).^{40,46,97,426,460,461} Catheter ablation or surgery may offer a reasonable alternative or adjunct to ICD therapy in carefully selected patients (*Level of evidence: C*).⁴⁶³⁻⁴⁶⁵
 3. ICD therapy is indicated in adults with CHD and a *systemic left ventricular ejection fraction* $\leq 35\%$, biventricular physiology, and New York Heart Association (NYHA) class II or III symptoms (*Level of evidence: B*).^{97,111,428,433-435}

Class IIa ICD therapy is reasonable in selected adults with *tetralogy of Fallot* and multiple risk factors for sudden cardiac death, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration ≥ 180 ms, extensive right ventricular scarring, or inducible sustained ventricular tachycardia at electrophysiologic study (*Level of evidence: B*).^{31,40,76,84,101,313,439,445,466}

2. ICD therapy may be considered in adults with CHD and a *systemic ventricular ejection fraction* $< 35\%$ in the absence of overt symptoms (NYHA class I) or other known risk factors (*Level of evidence of: C*).^{36,97,467}
3. ICD therapy may be considered in adults with CHD and *syncope of unknown origin* with hemodynamically significant sustained ventricular tachycardia or fibrillation inducible at electrophysiologic study (*Level of evidence: B*).^{76,97,436}
4. ICD therapy may be considered for nonhospitalized adults with CHD *awaiting heart transplantation* (*Level of evidence: C*).^{97,468}

Data from the DAI-T4F National Registry



Objectives

- Characteristics and outcomes of patients
- Identification of risk factors of appropriate therapy discharge (ECG++)
- Evaluation of device related complications
- Overall mortality

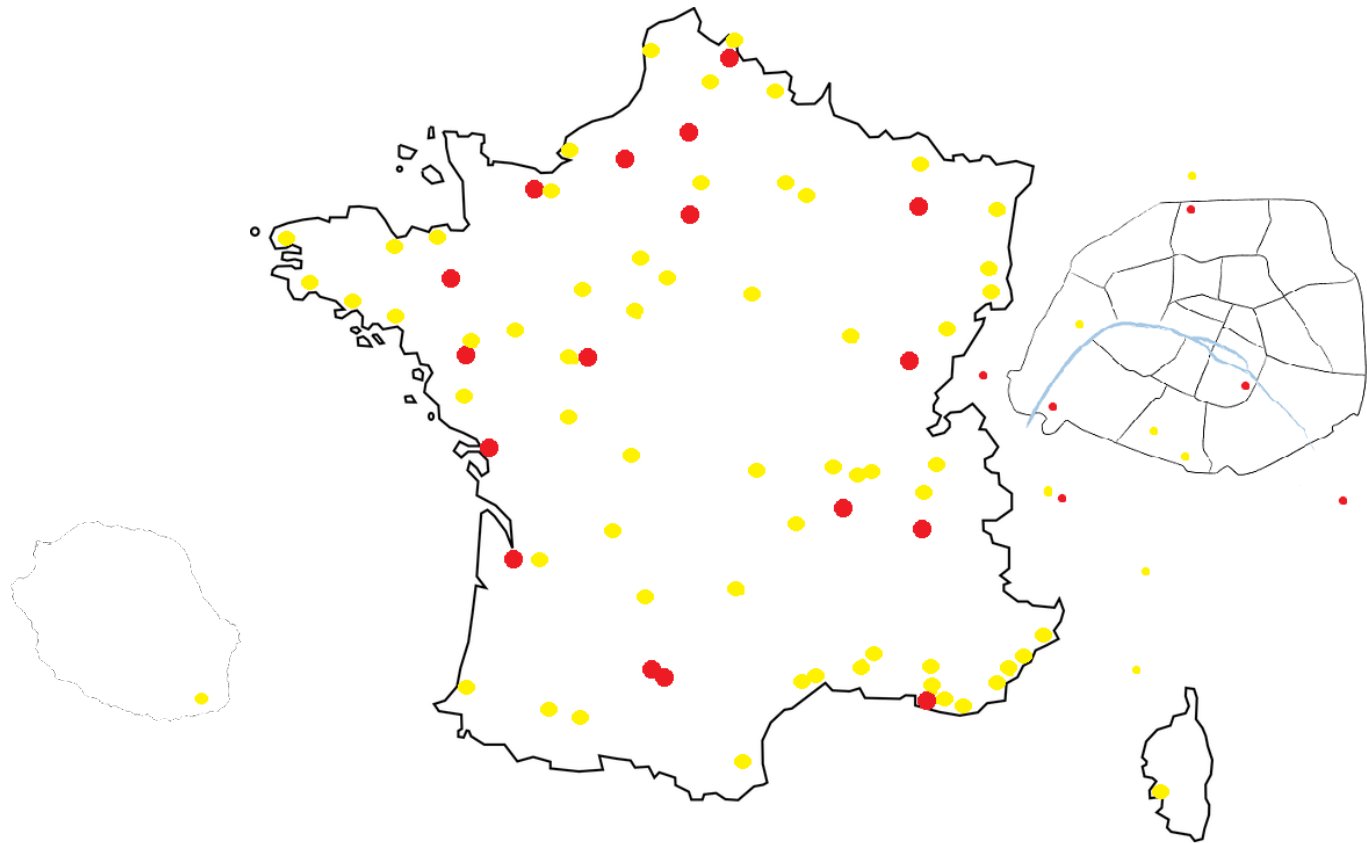
Methods

- Between 2005 and 2016
- Multicentric retrospective
- All TOF patients with ICD from 22 French centers were consecutively enrolled

Inclusion criteria

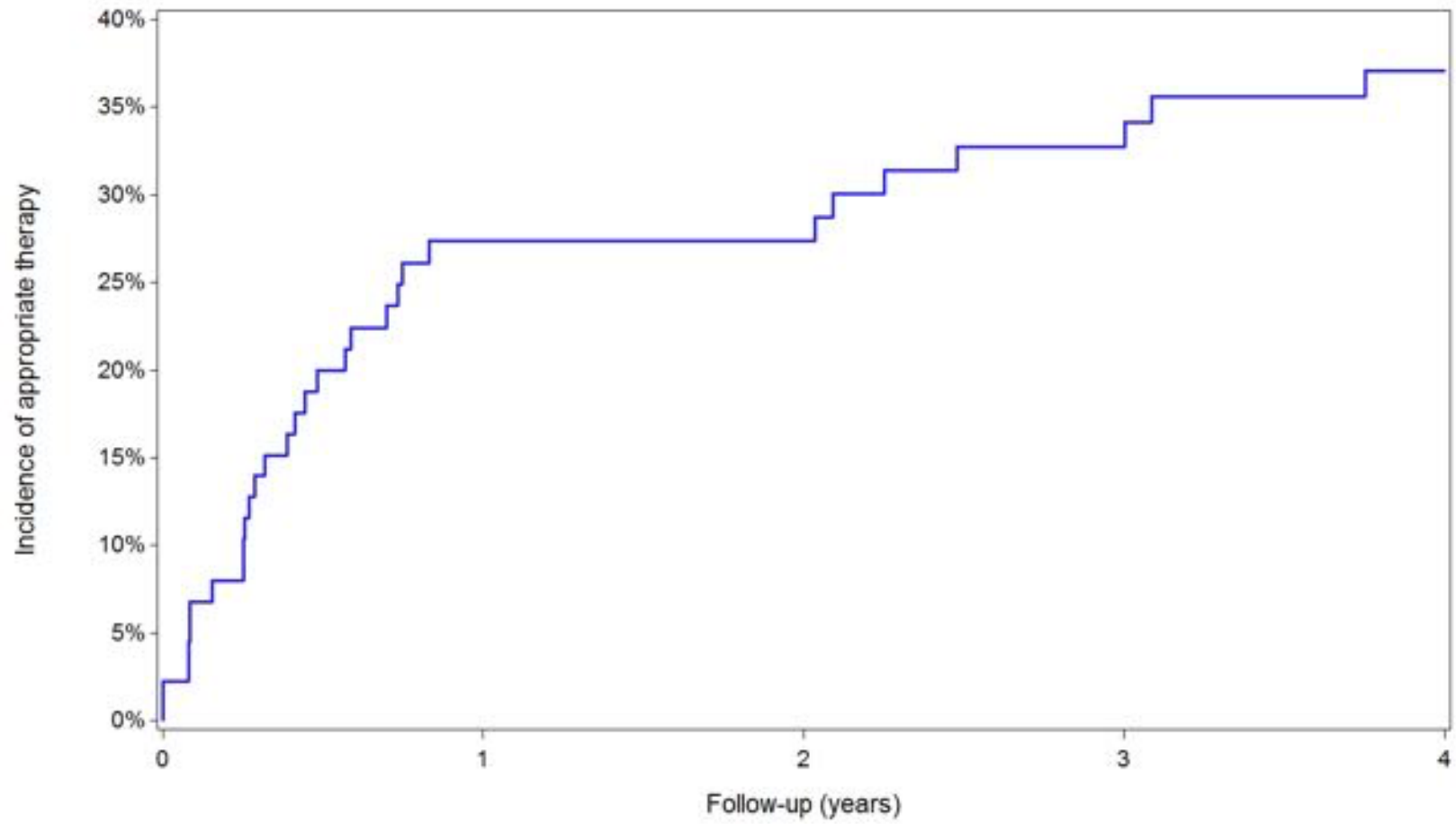
- > 18 yo
- TOF and ICD
- Primary or secondary prevention

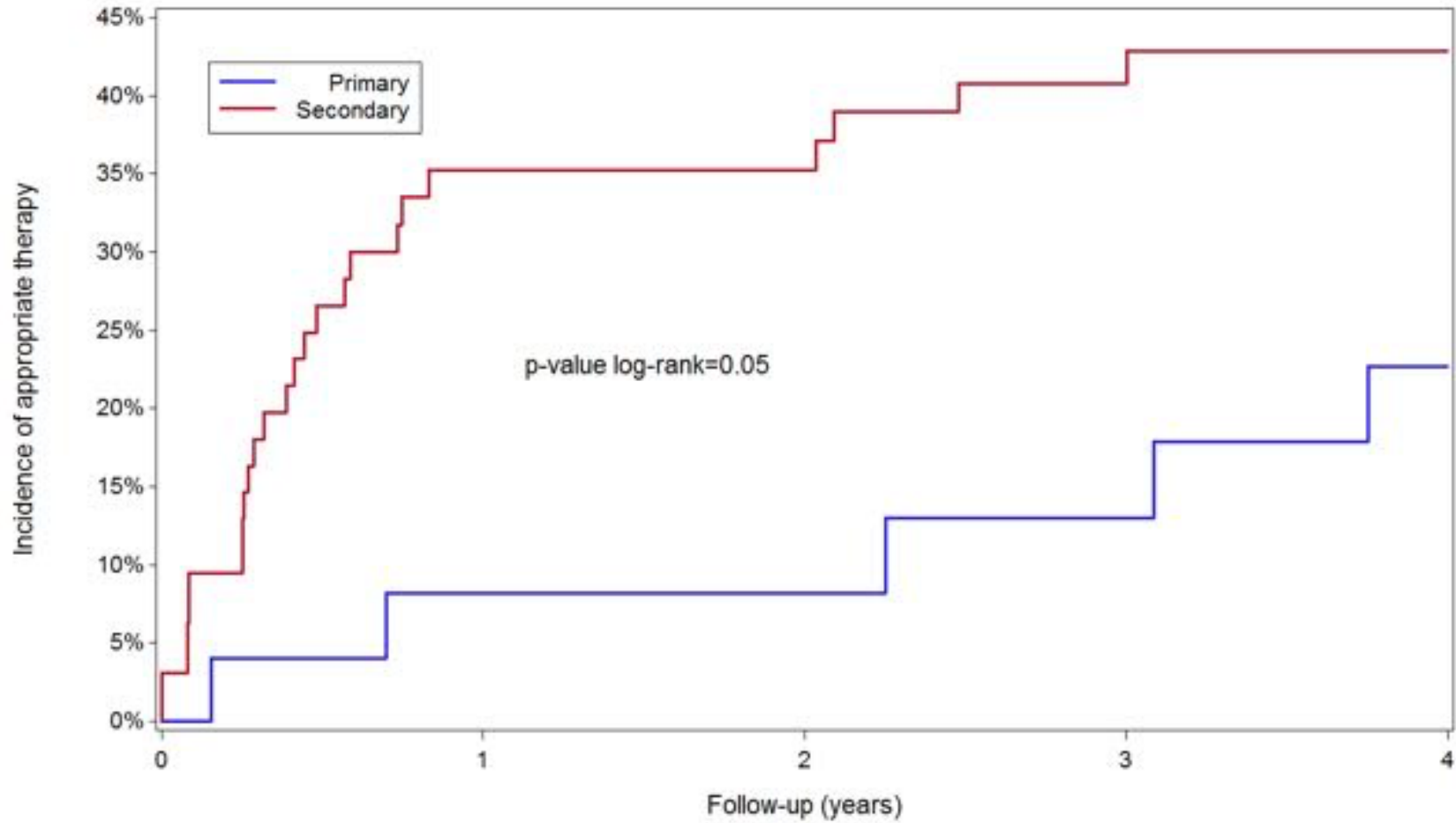
Geographical location of the centers



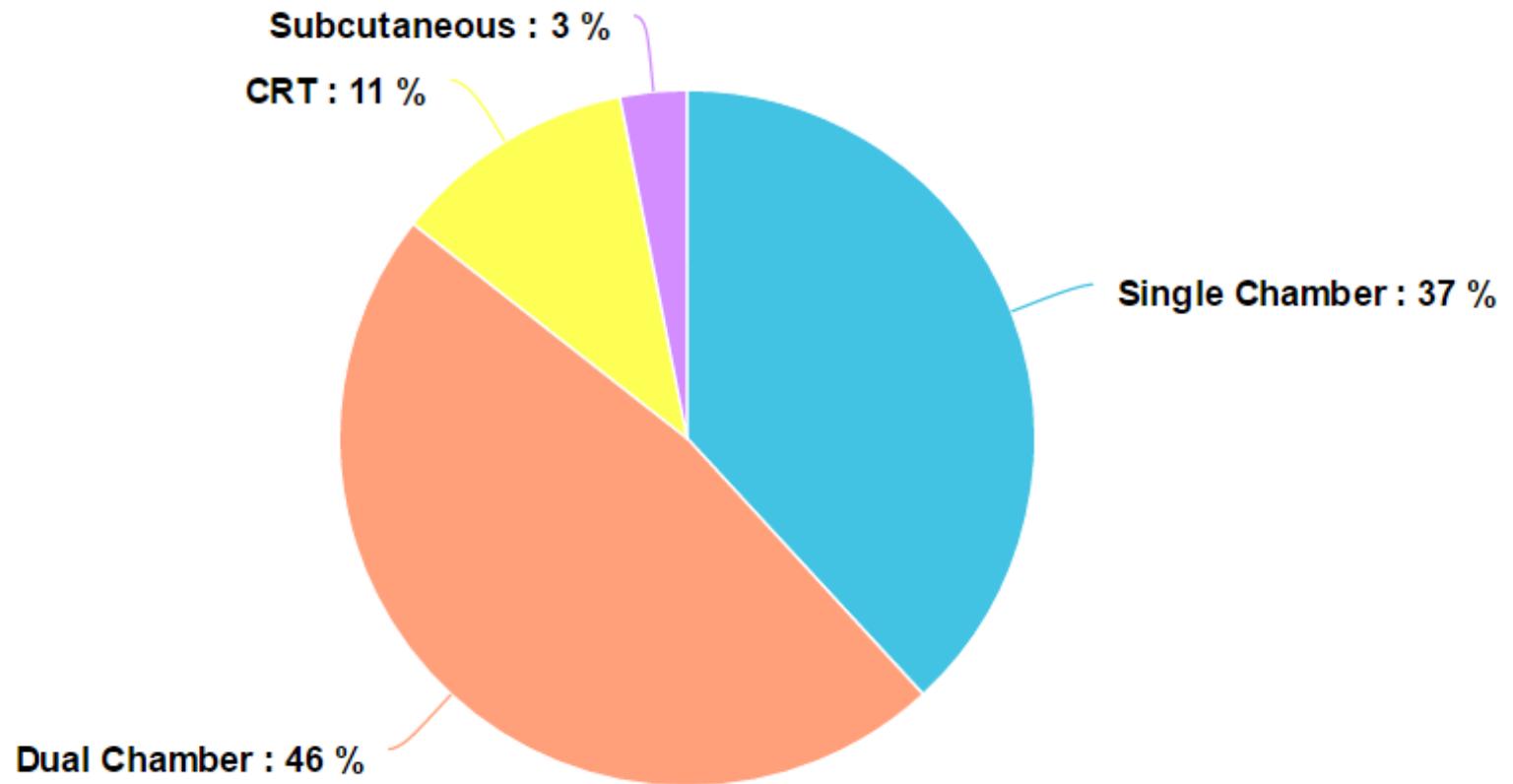
Preliminary results

- 101 patients : 43 ± 13 yo, 69% males
- Secondary prevention : 71%
- Primary prevention : 29%
- After a mean follow-up of 6.0 ± 4 years, 44 patients (43%) experienced at least one appropriate therapy
- \geq one ICD-related complication occurred in 40 patients (39%)





Annual-incidences of 4.95% vs 14.1% (P=0.009)



DAI – T4F : Take Home Message

- DAI – T4F : French National Registry ongoing
- High rates of appropriate ICD therapies
- Especially in secondary prevention
- Major ICD-related complications remain very high
- Selection for primary prevention remains challenging and can be potentially improved in the future

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