Mort Subite Cardiaque: Etiologies Electriques

J-Claude Daubert

Professeur émérite Université de Rennes 1, France

Liens d'intérêt:

Research grants-Consultant

- LivaNova
- Medtronic
- Novartis
- ST Jude Medical

Centre cardio-thoracique, CHU Rennes

Définitions - Prévalence

« **Sudden cardiac death** is an unexpected death due to cardiac causes that occurs in a short time period (<1 hour of symptom onset; <24 h for OMS) in a person with known or unknown cardiac disease »

RJ Myerburg Circulation 1997

« **Aborted cardiac arrest** is an unexpected circulatory arrest, occuring within 1-hr of onset of acute symptoms, which is reversed by successful resuscitation maneuvers »

2015 ESC Task Force

Prévalence et Incidence de la MS en France: Estimations

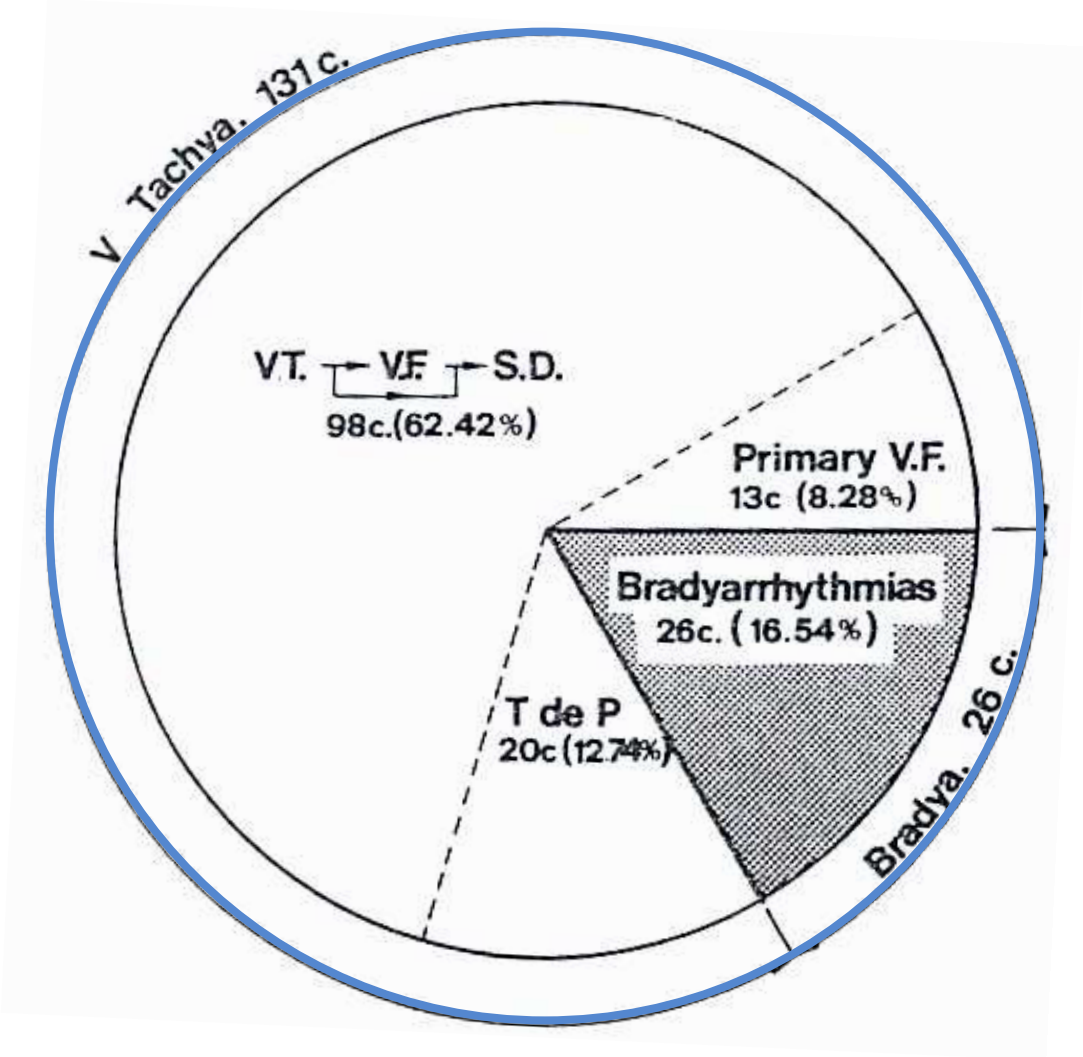
1/1000 hab./an = 60.000/an?
3H/1F; pic de fréquence: 45-75 ans

Mécanismes Rythmiques de l'Arrêt Cardiaque indépendamment de la cause

Revue de la littérature

N=231 décès subits sous Holter

Holters implantés
Mémoires PM/DAI



TDRV: 83.4%
Brady: 16%

A Bayès de Luna, Ph Coumel Am Heart J 1989

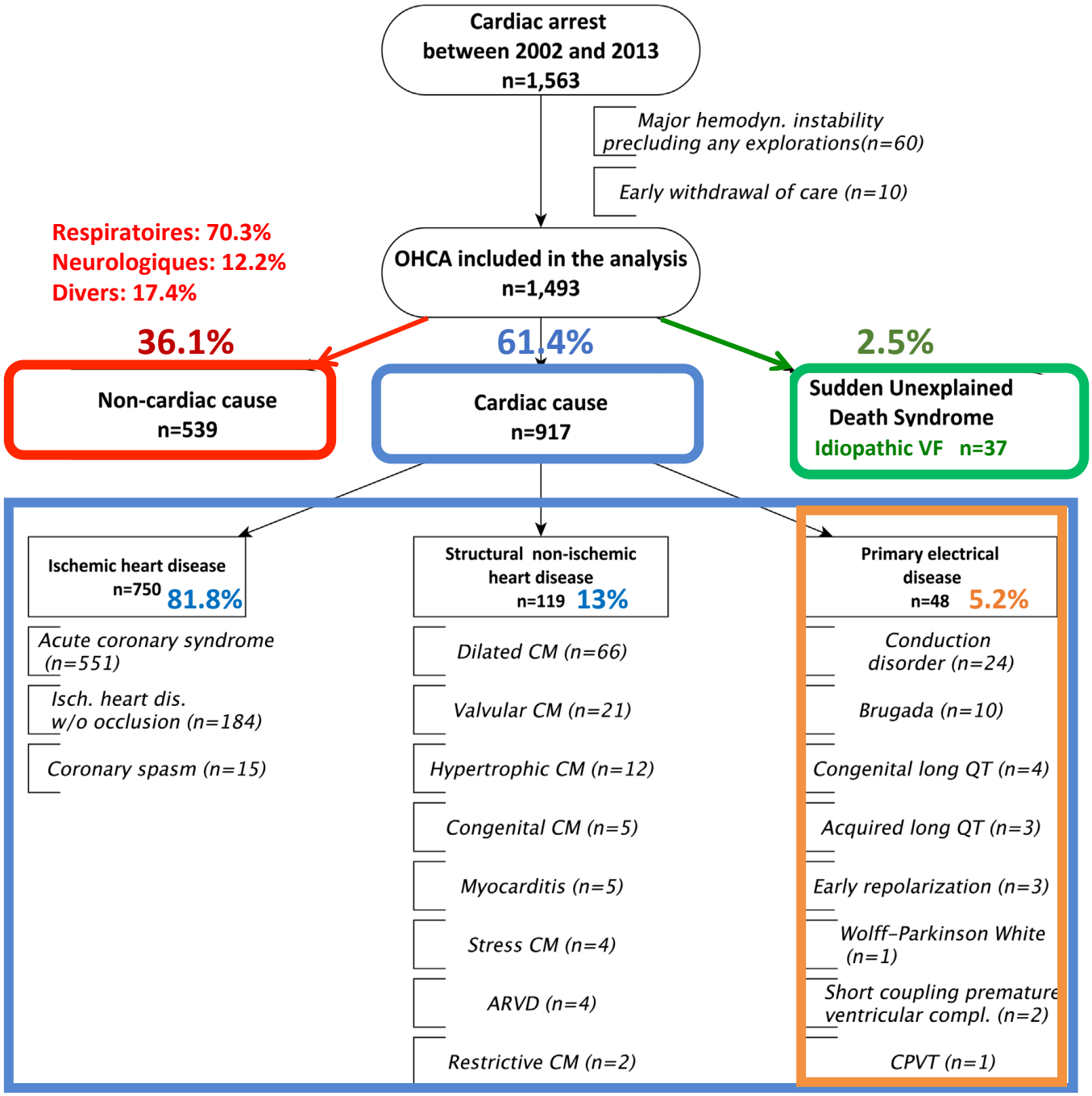
Etiologies de l'Arrêt Cardiaque

Arrêts cardiaques « resuscités » survenus en dehors de l'hôpital (2002-2014): 1563 pts; âge moyen: 60 ans

Bilan étiologique systématique comportant

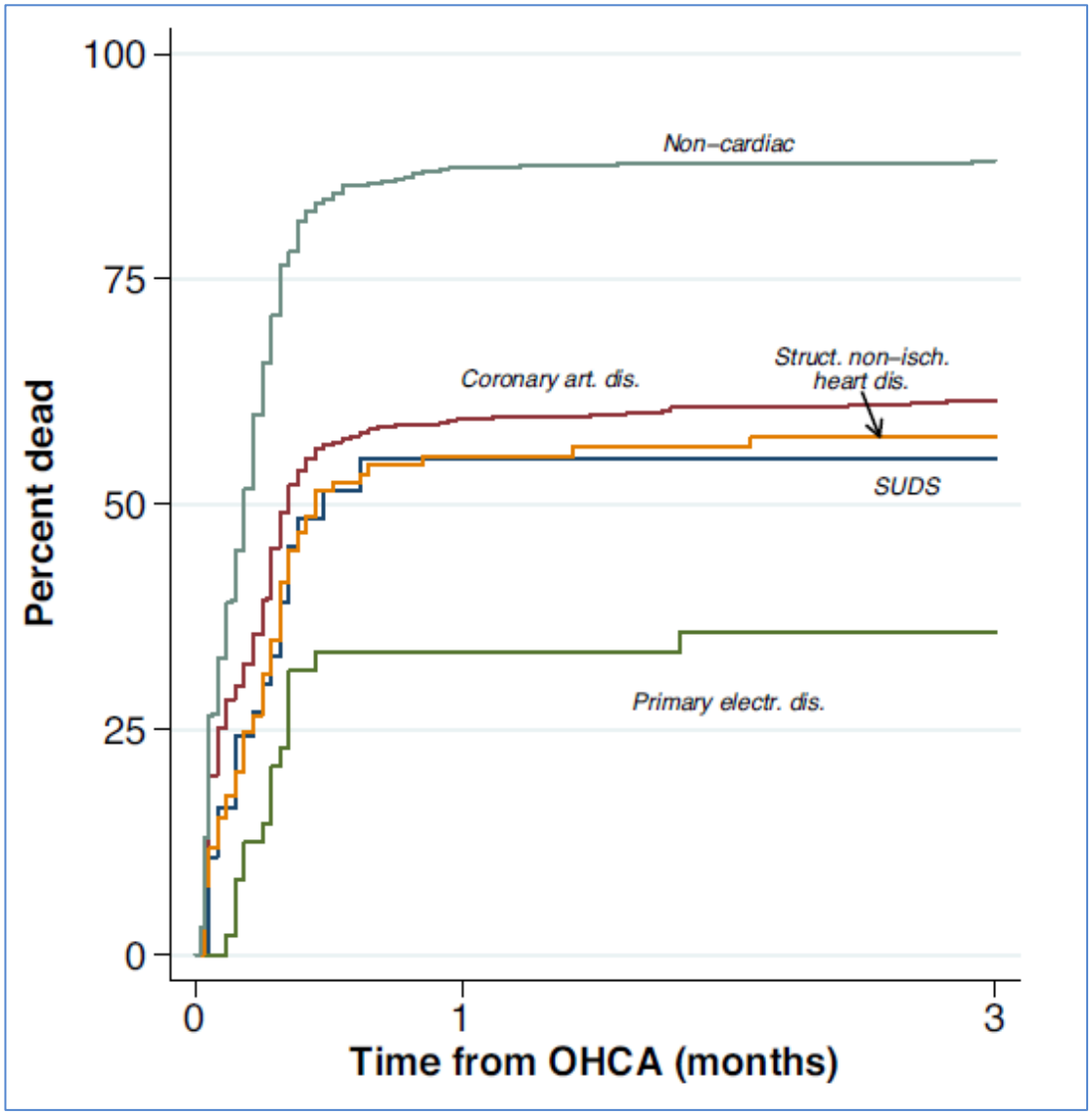
1. Dès l'admission, selon contexte:
   - coronarographie immédiate (70%)
   - et/ou scanner crânien (46%)
2. En USI:
   - histoire personnelle et familiale
   - ECG répétés
   - ETT/ETO
   - IRM
   - Bilan toxicologique
3. Autopsie en cas de décès

G. Geri et al. Resuscitation 2017; 117: 66–72



G. Geri et al. Resuscitation 2017; 117: 66–72

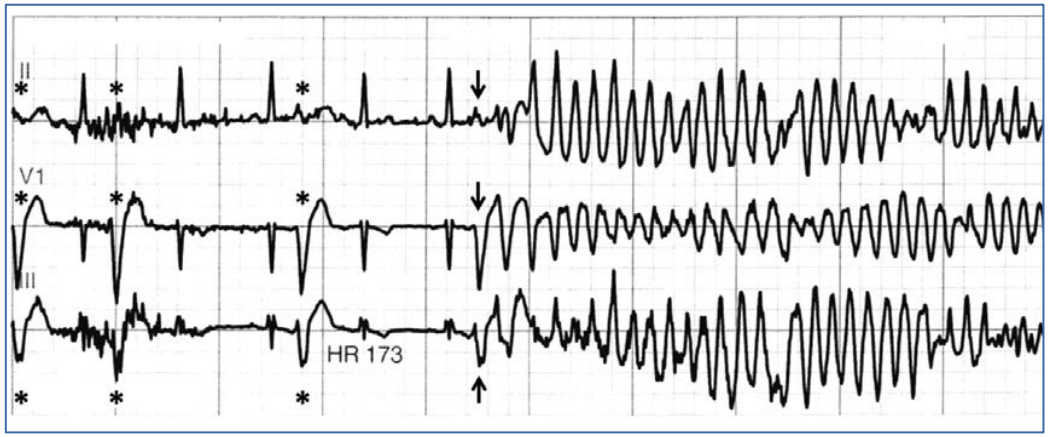
3-month Survival



G. Geri et al. Resuscitation 2017; 117: 66–72

SUDS: FV idiopathiques

- Diagnostic d'élimination après bilan exhaustif
- Incidence en diminution avec bilans plus complets
- Frontières incertaines avec autres syndromes: Onde J, TPCC…
- Patients asymptomatiques avant AC
- Risque élevé de récidive: 11-45% ; 31% à 5 ans *(méta-analyse Oyaidin)*
- Seul traitement recommandé: DAI
- Une place pour l'ablation (trigger)?



M Haissaguerre et al Circulation 2002;106:962-967

FV idiopathique: diagnostic d'élimination après bilan approfondi

Minimal required diagnostics

- ECG
- Blood chemistry
- Toxicological screening
- Holter monitoring
- Echocardiography
- Exercise ECG (including QTc posture test)
- Coronary angiography/ ventriculography*
- MRI†
- **No diagnosis**
- Sodium channel blocker provocation
- Ergonovine provocation

Independent of results provocation testing

- Targeted genetic testing based on phenotype‡

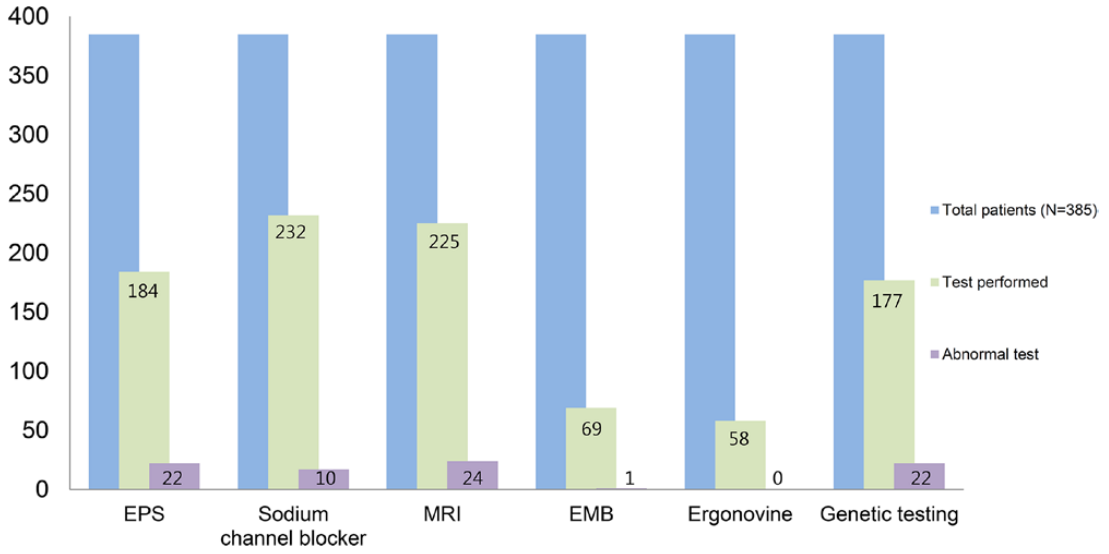
No pathogenic mutation

- Diagnosis IVF

↓

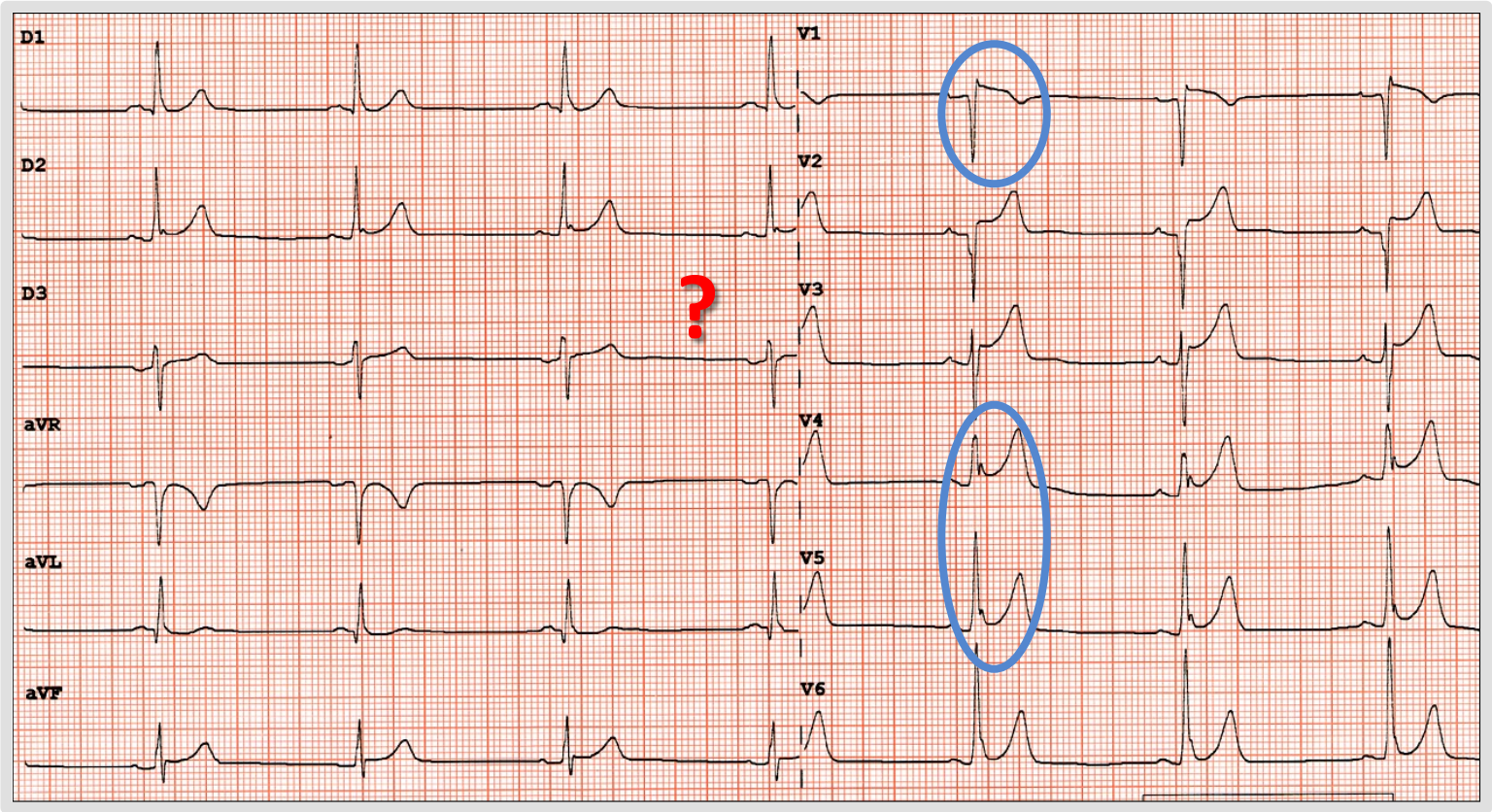
- FU: ECG/ echocardiography every 2 years
- After 6 years prolongation of FU interval
- Screening first degree family members: ECG, echocardiography, exercise ECG

Revue de litérature: 385 patients referés pour « FV idiopathique »



M Visser et al Circ Arrhythm Electrophysiol. 2016;9:e003817.

Repolarisation précoce: Syndrome de l'onde J



Haissaguerre M et al. Sudden cardiac arrest associated with early repolarization. N Engl J Med 2008; 358: 2016-2023

Beaucoup d'inconnues demeurent malgré une littérature prolifique et plusieurs conférences de consensus

Repolarisation précoce: Syndrome de l'onde J

Score de Shanghai	Points
I. Clinical History	
A. Unexplained cardiac arrest, documented VF or polymorphic VT	3
B. Suspected arrhythmic syncope	2
C. Syncope of unclear mechanism/unclear etiology	1
*Only award points once for highest score within this category	
II. Twelve-Lead ECG	
A. ER ≥0.2 mV in ≥2 inferior and/or lateral ECG leads with horizontal/descending ST segment	2
B. Dynamic changes in J-point elevation (≥0.1 mV) in ≥2 inferior and/or lateral ECG leads	1.5
C. ≥0.1 mV J-point elevation in at least 2 inferior and/or lateral ECG leads	1
*Only award points once for highest score within this category	
III. Ambulatory ECG Monitoring	
A. Short-coupled PVCs with R on ascending limb or peak of T wave	2
IV. Family History	
A. Relative with definite ERS	2
B. ≥2 first-degree relatives with a II.A. ECG pattern	2
C. First-degree relative with a II.A. ECG pattern	1
D. Unexplained sudden cardiac death <45 years in a first- or second-degree relative	0.5
*Only award points once for highest score within this category	
V. Genetic Test Result	
A. Probable pathogenic ERS susceptibility mutation	0.5

Peut-on prédire le caractère « pathogène » d'un syndrome de l'onde J?

Score (requires at least 1 ECG finding)

≥5 points: Probable/definite ERS

3-4.5 points: Possible ERS

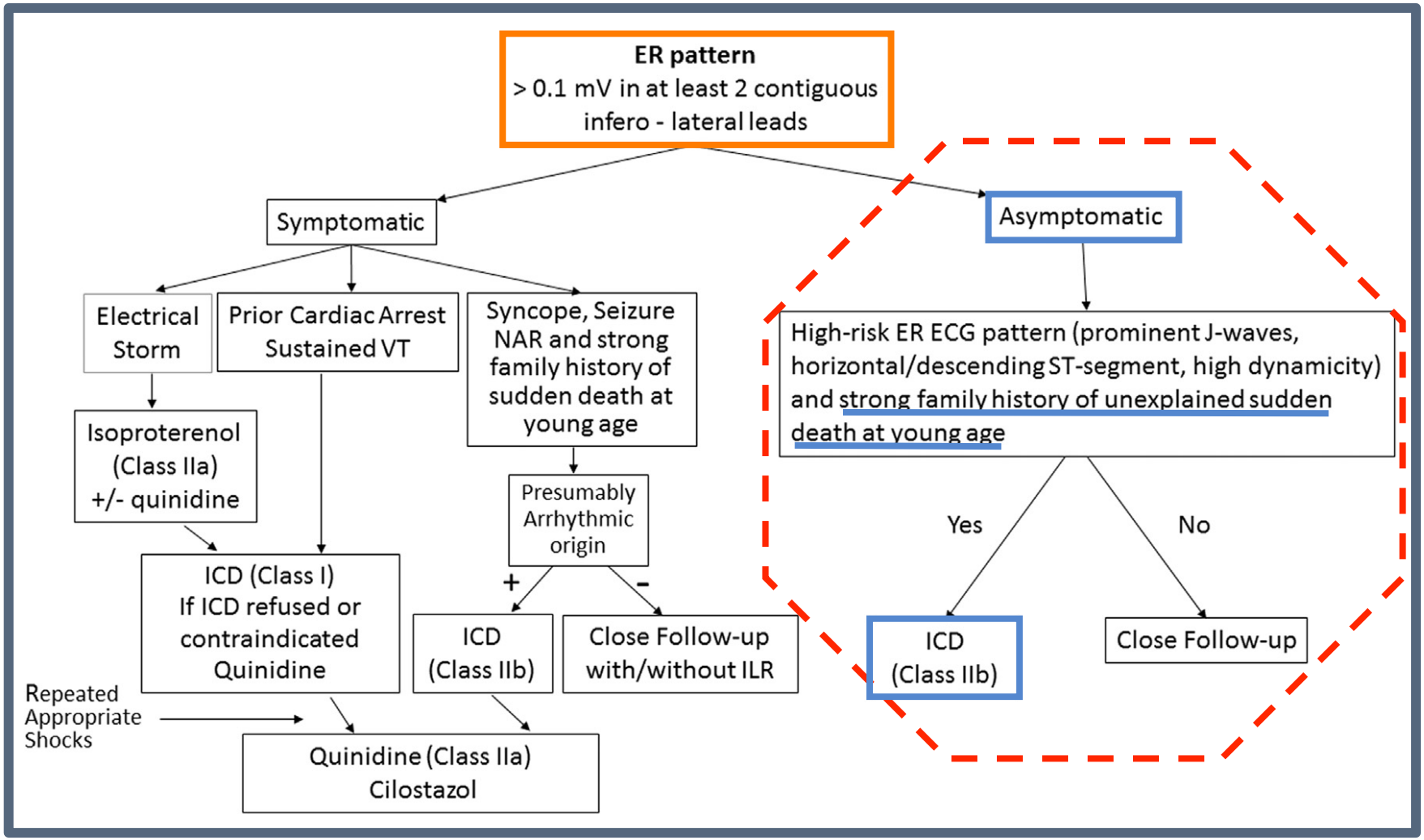
<3 points: Non-diagnostic

Pas de recommandations officielles des sociétés savantes

J-Wave Syndromes Consensus Report

C Antzelevitch et al Heart Rhythm 2016; 13: 295-324

Repolarisation précoce: Syndrome de l'onde J



J-Wave Syndromes Consensus Report

C Antzelevitch et al Heart Rhythm 2016; 13: 295-324

Brugada

Prévalence: 1/10.000? (1/1000 en Asie SE)

Transmission autosomique dominante

12 gènes associés, dont 2 seulement expliquent plus de 5% des cas

Pénétrance très variable, dépendant de l'âge et du sexe (evts rythmiques: x 8 H/F)

Age moyen de survenue des évènements rythmiques: 41±15 ans

Facteurs déclenchants: fièvre, alcool…

Diagnostic

Recommendations	Class[a]	Level[b]
Brugada syndrome is diagnosed in patients with ST-segment elevation with type 1 morphology ≥2 mm in one or more leads among the right precordial leads V1 and/or V2 positioned in the second, third, or fourth intercostal space, occurring either spontaneously or after provocative drug test with intravenous administration of sodium channel blockers (such as ajmaline, flecainide, procainamide or pilsicainide).	I	C

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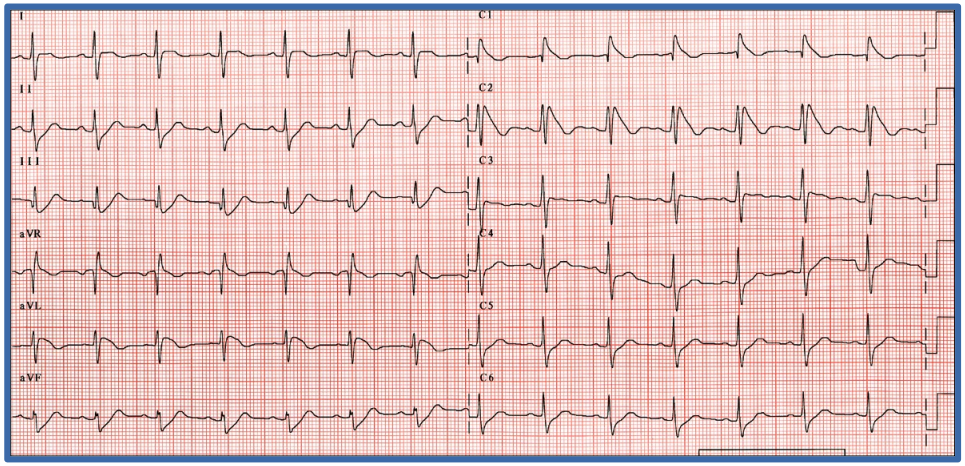

Brugada: Stratification du risque

Stratification du risque

Priorité aux évènements cliniques

- ATCD TV/FV/MS: 13.5%/an
- Syncopes: 3.2%
- Asymptomatique: 1%/an

ECG: type 1 spontané

Incertitudes:

- Fragmentation QRS
- SVP???
- Histoire familiale
- Génotypage

Shanghai score for Brugada syndrome	Point
I. ECG (12-Lead/Ambulatory)	
A. Spontaneous type 1 Brugada ECG pattern at nominal or high leads	3.5
B. Fever-induced type 1 Brugada ECG pattern at nominal or high leads	3
C. Type 2 or 3 Brugada ECG pattern that converts with provocative drug challenge	2
**Only award points once for highest score within this category. One item from this category must apply.*	
II. Clinical History*	
A. Unexplained cardiac arrest or documented VF/ polymorphic VT	3
B. Nocturnal agonal respirations	2
C. Suspected arrhythmic syncope	2
D. Syncope of unclear mechanism/unclear etiology	1
E. Atrial flutter/fibrillation in patients <30 years without alternative etiology	0.5
**Only award points once for highest score within this category.*	
III. Family History	
A. First- or second-degree relative with definite BrS	2
B. Suspicious SCD (fever, nocturnal, Brugada aggravating drugs) in a first- or second-degree relative	1
C. Unexplained SCD <45 years in first- or second-degree relative with negative autopsy	0.5
**Only award points once for highest score within this category.*	
IV. Genetic Test Result	
A. Probable pathogenic mutation in BrS susceptibility gene	0.5

C Antzelevitch et al Heart Rhythm 2016; 13: 295-324

Brugada: Prise en charge thérapeutique

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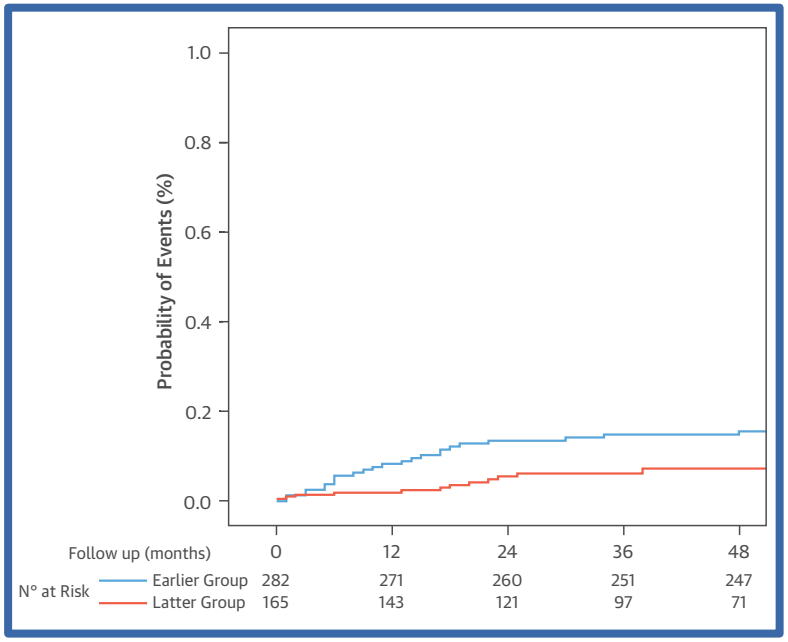
ICD implantation is recommended in patients with a diagnosis of Brugada syndrome who (a) Are survivors of an aborted cardiac arrest and/or (b) Have documented spontaneous sustained VT.	**I**	**C**
ICD implantation should be considered in patients with a spontaneous diagnostic type I ECG pattern and history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

Place de la quinidine et de l'ablation discutée

Une maladie moins grave qu'annoncée?



M Casado-Arroyo et J Brugada JACC 2016; 68: 614-623

QT long

Recommendations	Class[a]	Level[b]
LQTS is diagnosed with either – QTc ≥480 ms in repeated 12-lead ECGs or – LQTS risk score >3.[431]	I	C
LQTS is diagnosed in the presence of a confirmed pathogenic LQTS mutation, irrespective of the QT duration.	I	C
ECG diagnosis of LQTS should be considered in the presence of a QTc ≥460 ms in repeated 12-lead ECGs in patients with an unexplained syncopal episode in the absence of secondary causes for QT prolongation.	IIa	C

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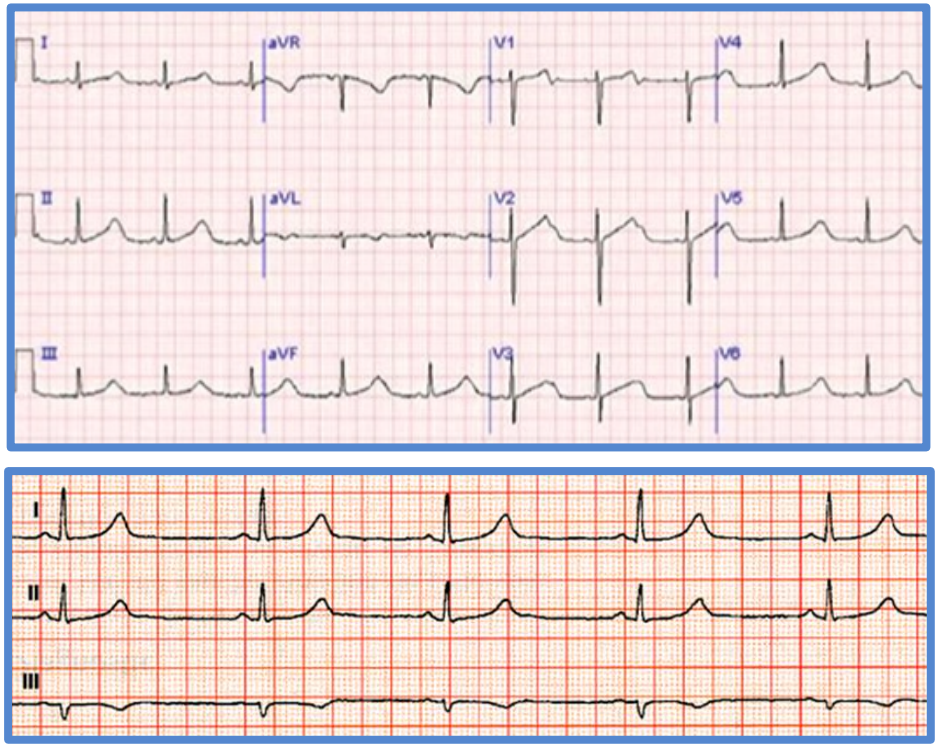


- Prévalence: 1/2500 (forme autosomique dominante)
- Age moyen de découverte: 14 ans
- 13 gènes associés (LQT1… LQT13)
- 3 expliquent 90% des cas mutés: KCNQ1, KCNH2, SCN5A)
- Génotypage recommandé chez les 1er degré en cas de mutation pathogène

QT long

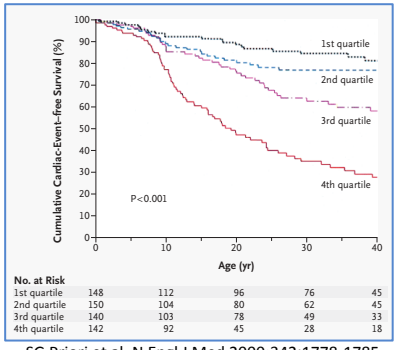
Stratification du risque

1) Symptômes: risque MS

- Très élevé en cas d'ACR, y compris sous BB
  14% à 5 ans
- Elevé en cas de syncopes: 5%/an
- Faible chez asymptomatiques: 1%/an

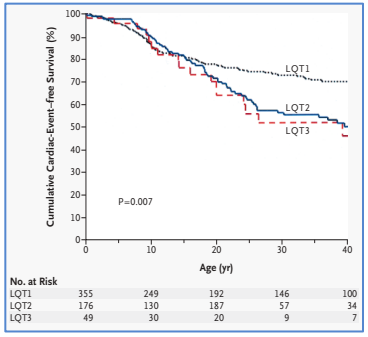
2) ECG: durée QT

Q1: ≤446 ms
Q2: 447-468 ms
Q3: 469-498 ms
Q4> 498 ms



SG Priori et al. N Engl J Med 2000;342:1778-1785

3) Mutation?



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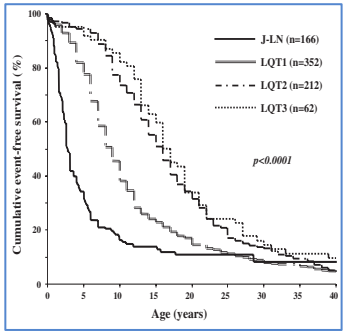

PJ Schwartz et al Circ AE. 2012; 5: 868-877

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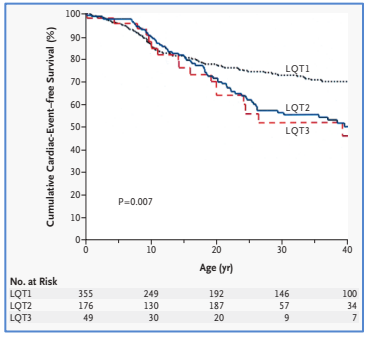
Beta-blockers are recommended in patients with a clinical diagnosis of LQTS.	I	B
ICD implantation with the use of beta-blockers is recommended in LQTS patients with previous cardiac arrest.	I	B
Beta-blockers should be considered in carriers of a causative LQTS mutation and normal QT interval.	IIa	B
ICD implantation in addition to beta-blockers should be considered in LQTS patients who experienced syncope and/or VT while receiving an adequate dose of beta-blockers.	IIa	B

Place de la sympathectomie cardiaque gauche et des bloqueurs Na^+ discutée

QT long

		Points
Electrocardiographic findings*		
A	QTc,† ms	
	≥480	3
	460–479	2
	450–459 (men)	1
B	QTc† 4th minute of recovery from exercise stress test ≥480 ms	1
C	Torsades-de-Pointes‡	2
D	T-wave alternans	1
E	Notched T wave in 3 leads	1
F	Low heart rate for age§	0.5
Clinical history		
A	Syncope‡	
	With stress	2
	Without stress	1
B	Congenital deafness	0.5
Family history		
A	Family members with definite LQTS‖	1
B	Unexplained sudden cardiac death younger than age 30 among immediate family members‖	0.5



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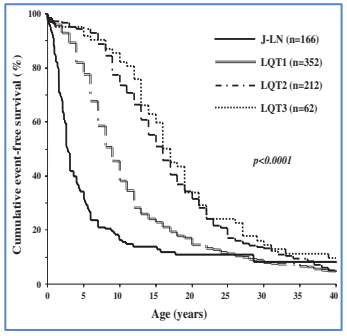

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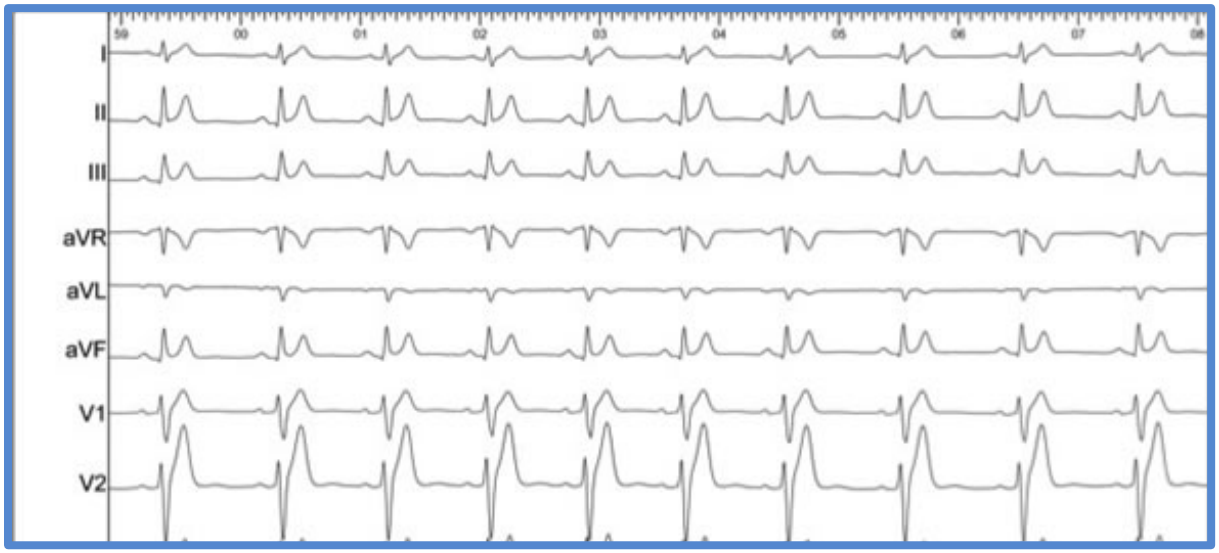
Beta-blockers are recommended in patients with a clinical diagnosis of LQTS.	I	B
ICD implantation with the use of beta-blockers is recommended in LQTS patients with previous cardiac arrest.	I	B
Beta-blockers should be considered in carriers of a causative LQTS mutation and normal QT interval.	IIa	B
ICD implantation in addition to beta-blockers should be considered in LQTS patients who experienced syncope and/or VT while receiving an adequate dose of beta-blockers.	IIa	B

Place de la sympathectomie cardiaque gauche et des bloqueurs Na^+ discutée

QT court



Recommendations	Class[a]	Level[b]
SQTS is diagnosed in the presence of a QTc ≤340 ms.	I	C
SQTS should be considered in the presence of a QTc ≤360 ms and one or more of the following: (a) A confirmed pathogenic mutation (b) A family history of SQTS (c) A family history of sudden death at age <40 years (d) Survival from a VT/VF episode in the absence of heart disease.	IIa	C

Recommendations	Class[a]	Level[b]
ICD implantation is recommended in patients with a diagnosis of SQTS who (a) Are survivors of an aborted cardiac arrest, and/or (b) Have documented spontaneous sustained VT.	I	C
Invasive EPS with PVS is not recommended for SCD risk stratification.	III	C

*Place de la Quinidine et du Sotalol discutée

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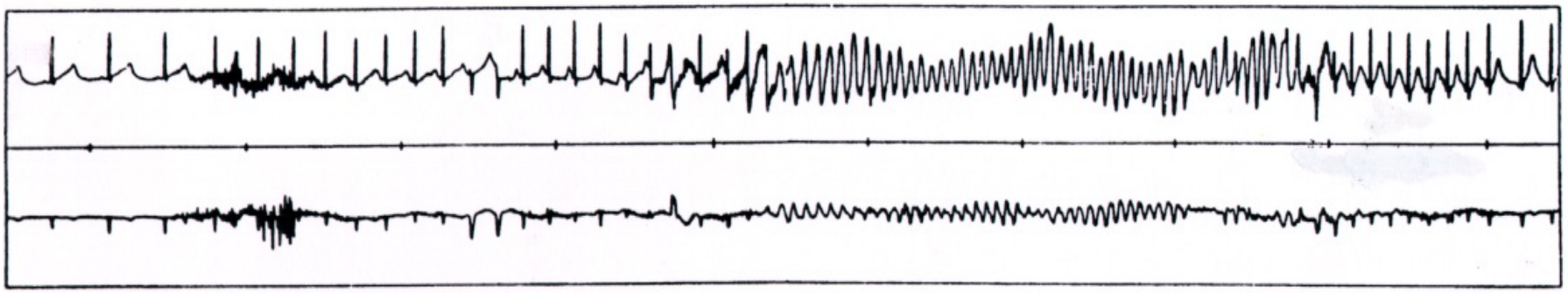
TV polymorphes catécholergiques



Recommendations	Class[a]	Level[b]
CPVT is diagnosed in the presence of a structurally normal heart, normal ECG and exercise- or emotion-induced bidirectional or polymorphic VT.	I	C
CPVT is diagnosed in patients who are carriers of a pathogenic mutation(s) in the genes *RyR2* or *CASQ2*.	I	C
Beta-blockers are recommended in all patients with a clinical diagnosis of CPVT, based on the presence of documented spontaneous or stress-induced VAs.	I	C
ICD implantation in addition to beta-blockers with or without flecainide is recommended in patients with a diagnosis of CPVT who experience cardiac arrest, recurrent syncope or polymorphic/bidirectional VT despite optimal therapy.	I	C
Therapy with beta-blockers should be considered for genetically positive family members, even after a negative exercise test.	IIa	C

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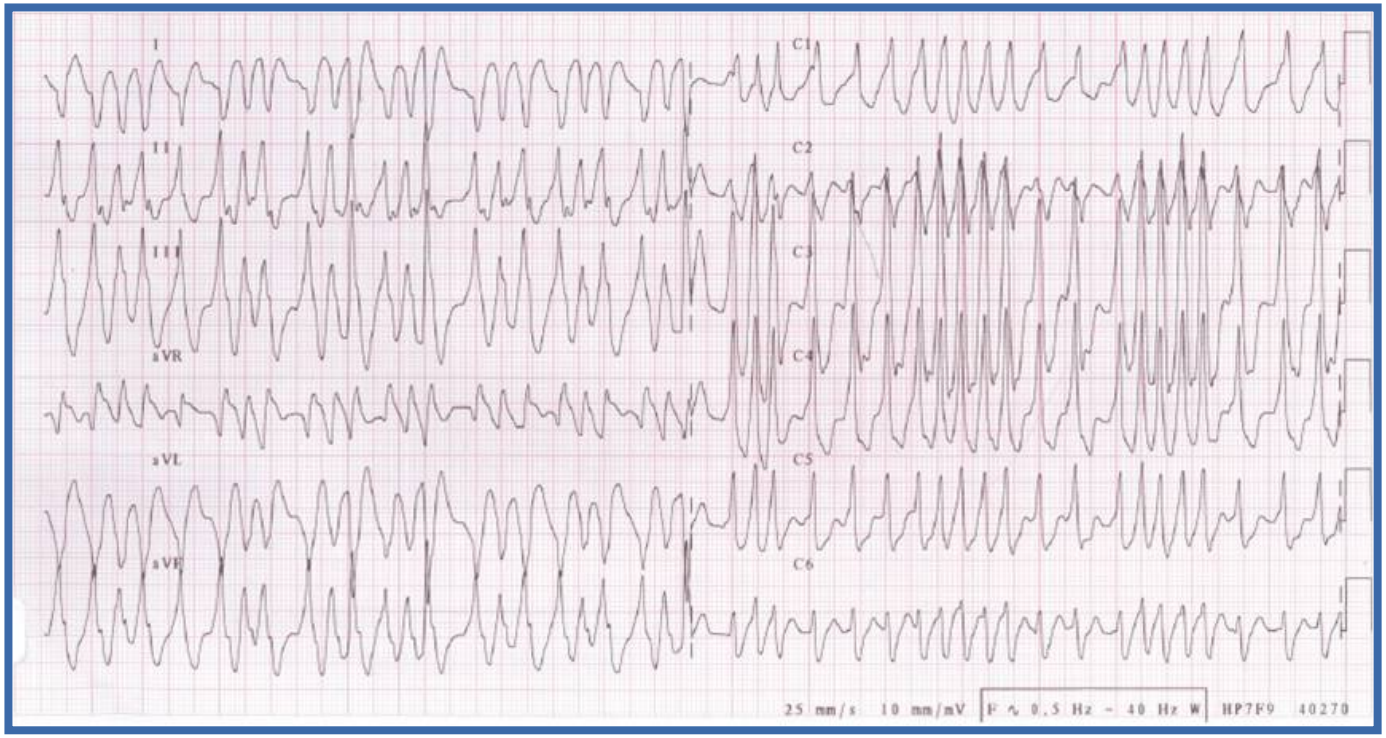
Place de la Flécaine?

WPW « malin »

- Transformation en FV d'une FA à cadence très rapide
- Evènement exceptionnel
- Cause curable par la seule ablation de la VA



Conclusions

- Dans la population générale, les causes électriques primaires n'expliquent que 5% environ des ACR
- La proportion est plus élevée chez l'enfant, l'adolescent et l'adulte jeune
- 3% des FV isolées restent encore sans explication: origine génétique? Lien avec syndrome de l'onde J?
- En dehors des très rares étiologies curables (WPW), un ACR de cause électrique primaire impose l'implantation d'un DAI
- La stratification du risque en prévention primaire reste délicate: priorité à l'histoire personnelle et aux symptômes