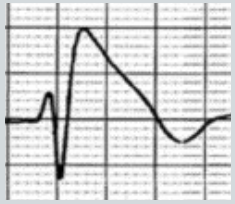
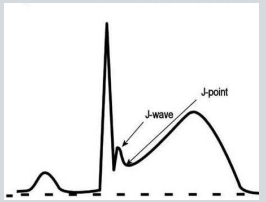




J-wave syndromes

Syndromes de Brugada et de repolarisation précoce

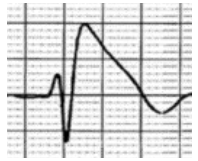
10émes journées du CMBCS

Chamonix, 08 octobre 2016

Didier IRLES

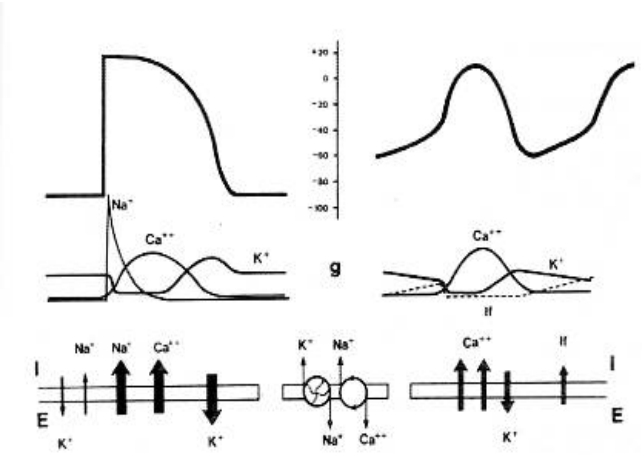
J-wave syndromes

De quoi parle-t-on ?



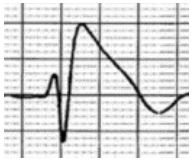
- Cœur échocardiographiquement sain

- Canalopathies
  - QT long
  - QT court
  - Brugada et syndrome de repolarisation précoce

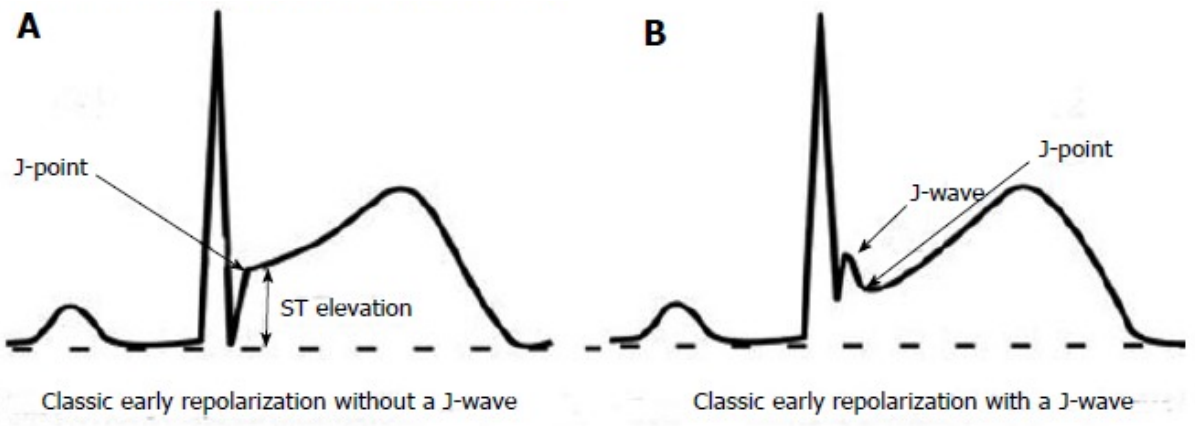


J-wave syndromes

De quoi parle-t-on ?

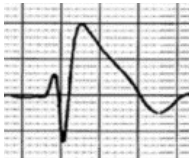


- Liée à la présence d'une onde J…

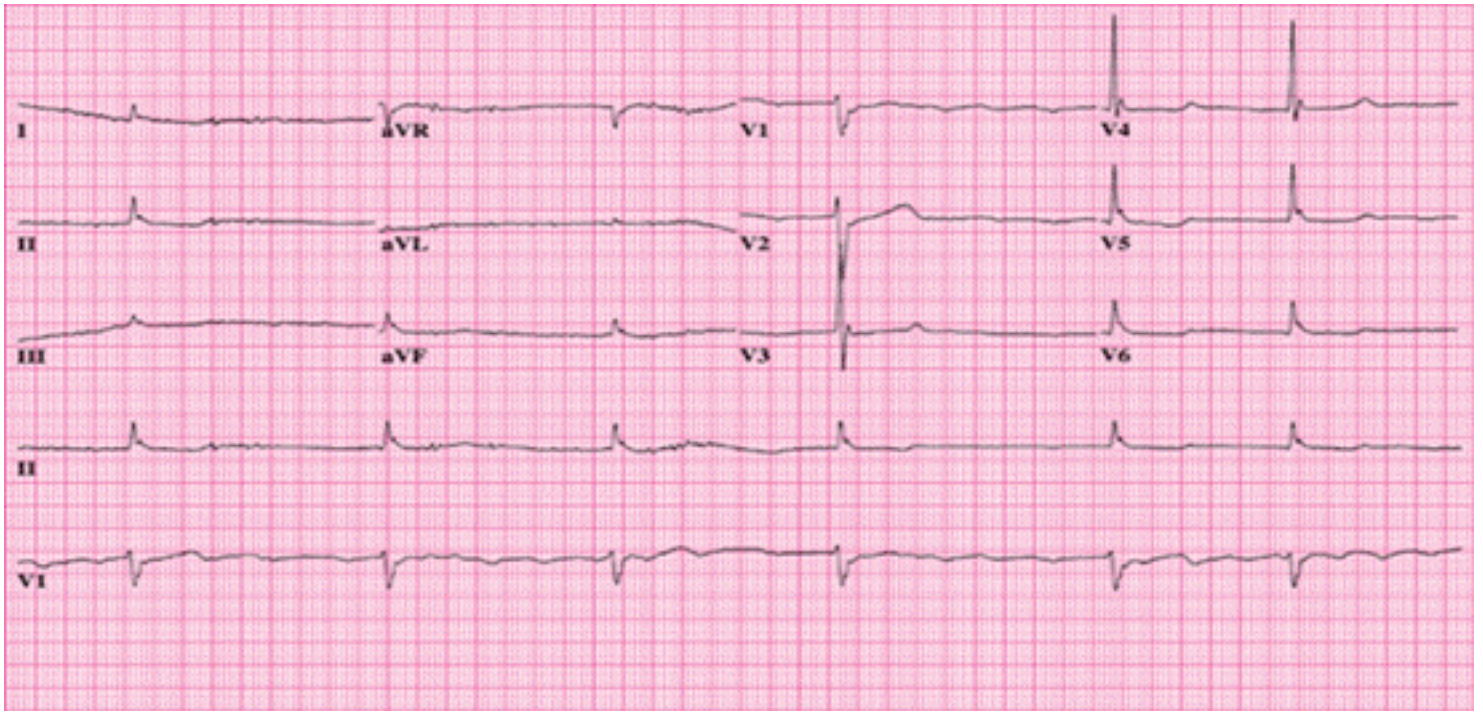


J-wave syndromes

De quoi parle-t-on ?

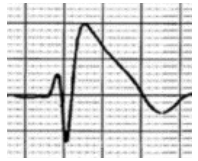


- Hypothermie, onde J d'Osborn…

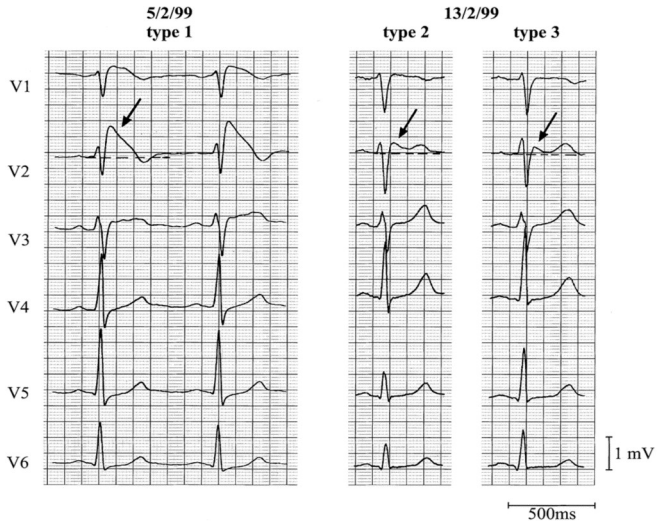


J-wave syndromes

De quoi parle-t-on ?



Syndrome de Brugada

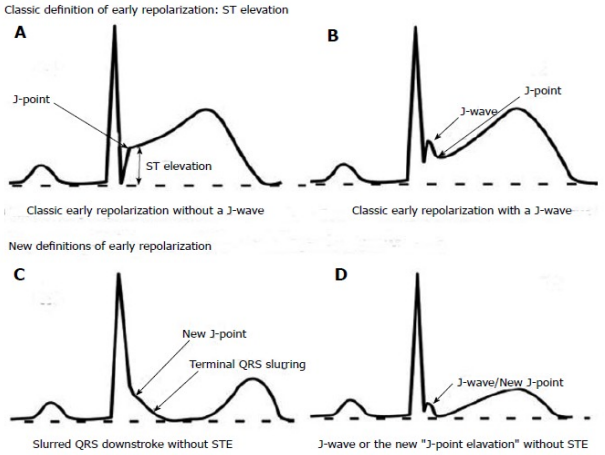


Pedro et Joseph Brugada, 1992

(dérivations V1-V2)

***J Am Coll Cardiol.* 1992 Nov 15;20(6):1391-6**

Syndrome de repolarisation précoce

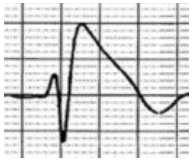


Michel Haissaguerre, 2008

(dérivations inférieures ou latérales)

***N Engl J Med.* 2008 May 8;358(19):2016-23**

Syndrome de Brugada



Right Bundle Branch Block, Persistent ST Segment Elevation and Sudden Cardiac Death: A Distinct Clinical and Electrocardiographic Syndrome

A Multicenter Report

PEDRO BRUGADA, MD, JOSEP BRUGADA, MD*†

Aalst, Belgium and Barcelona, Spain

Objectives. The objectives of this study were to present data on eight patients with recurrent episodes of aborted sudden death unexplainable by currently known diseases whose common clinical and electrocardiographic (ECG) features define them as having a distinct syndrome different from idiopathic ventricular fibrillation.

Background. Among patients with ventricular arrhythmias who have no structural heart disease, several subgroups have been defined. The present patients constitute an additional subgroup with these findings.

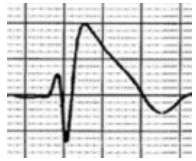
Methods. The study group consisted of eight patients, six male and two female, with recurrent episodes of aborted sudden death. Clinical and laboratory data and results of electrocardiography, electrophysiology, echocardiography, angiography, histologic study and exercise testing were available in most cases.

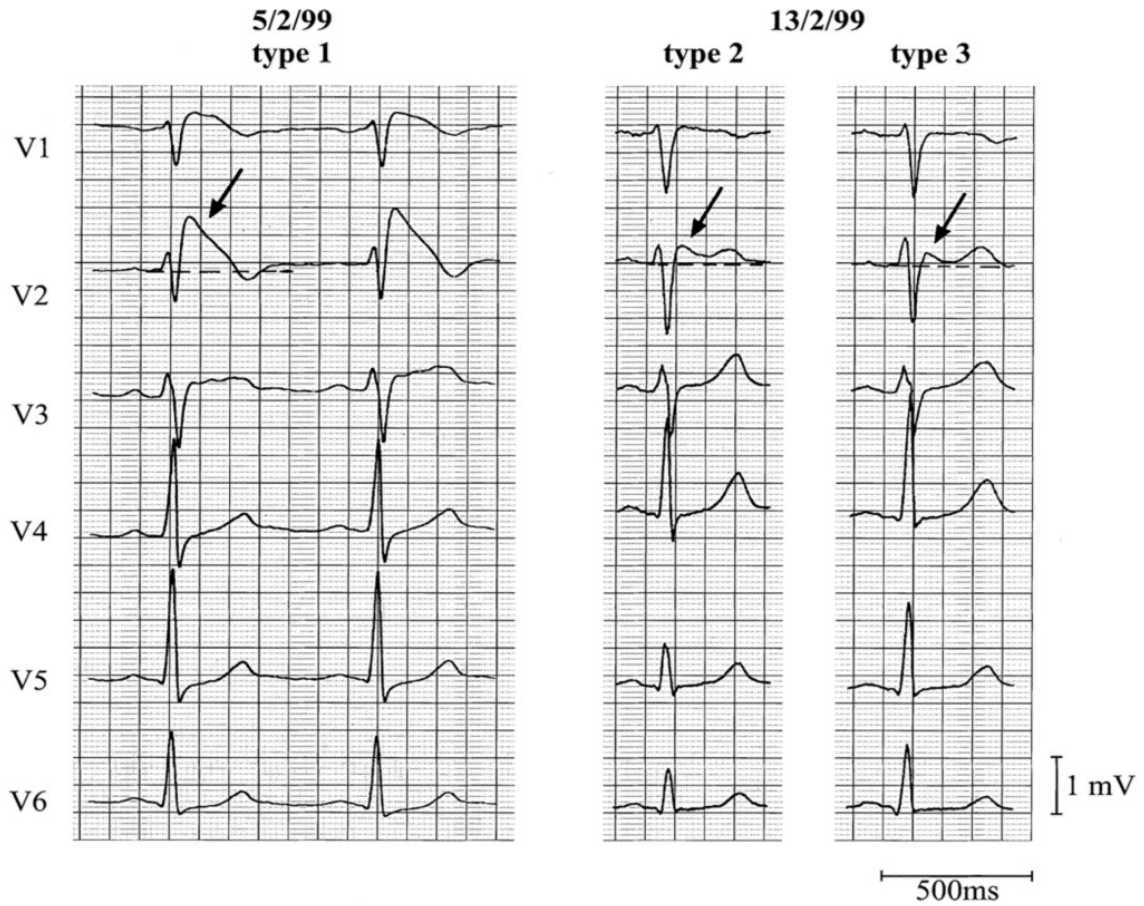
Results. The ECG during sinus rhythm showed right bundle branch block, normal QT interval and persistent ST segment elevation in precordial leads V_1 to V_2–V_3 not explainable by electrolyte disturbances, ischemia or structural heart disease. No histologic abnormalities were found in the four patients in whom ventricular biopsies were performed. The arrhythmia leading to (aborted) sudden death was a rapid polymorphic ventricular tachycardia initiating after a short coupled ventricular extrasystole. A similar arrhythmia was initiated by two to three ventricular extrastimuli in four of the seven patients studied by programmed electrical stimulation. Four patients had a prolonged HV interval during sinus rhythm. One patient receiving amiodarone died suddenly during implantation of a demand ventricular pacemaker. The arrhythmia of two patients was controlled with a beta-adrenergic blocking agent. Four patients received an implantable defibrillator that was subsequently used by one of them, and all four are alive. The remaining patient received a demand ventricular pacemaker and his arrhythmia is controlled with amiodarone and diphenylhydantoin.

Conclusions. Common clinical and ECG features define a distinct syndrome in this group of patients. Its causes remain unknown.

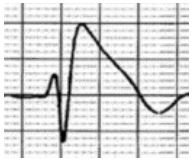
(J Am Coll Cardiol 1992;20:1391–6)

Syndrome de Brugada



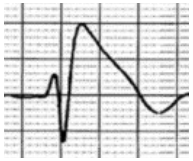


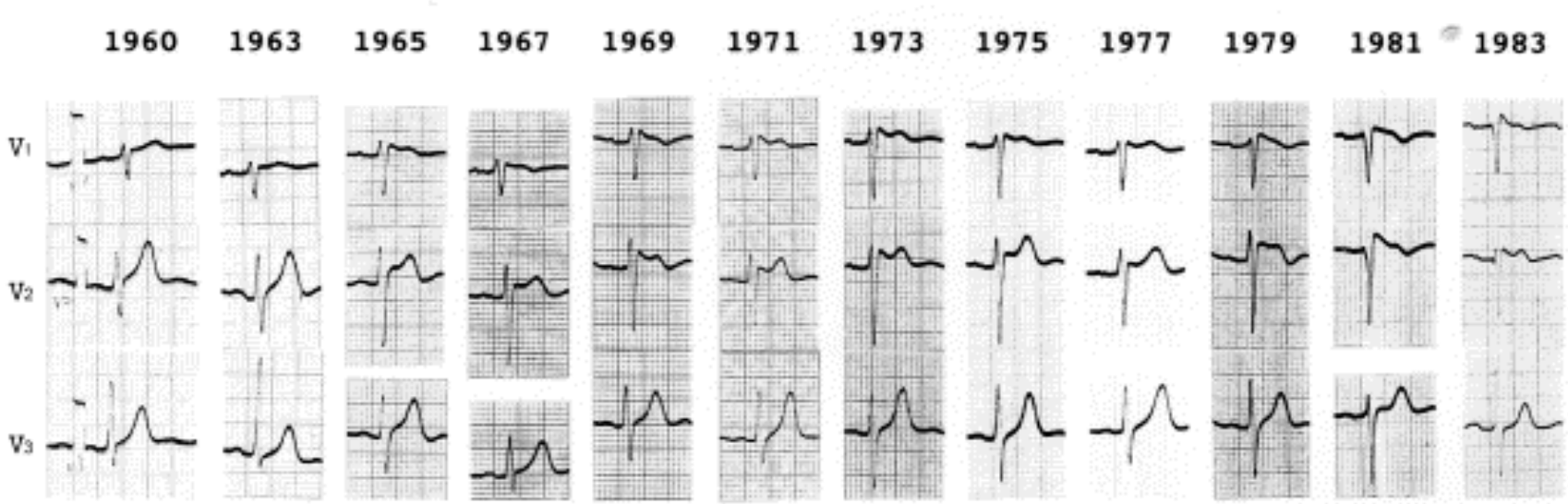
Syndrome de Brugada



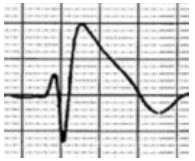
- Prévalence 5/10000
- Homme 80%
- Premier événement clinique entre 30 et 50 ans
- *Jusqu'à 20% des morts subites sur cœur sain ?*
- *Grande hétérogénéité inter-individuelle du risque…*

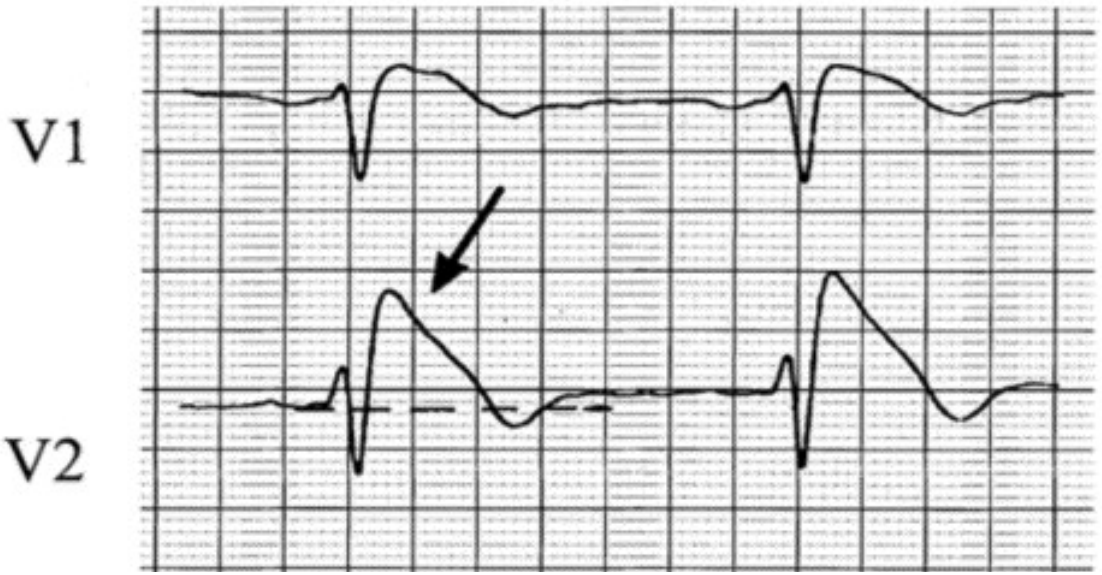
Syndrome de Brugada





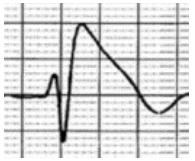
Syndrome de Brugada

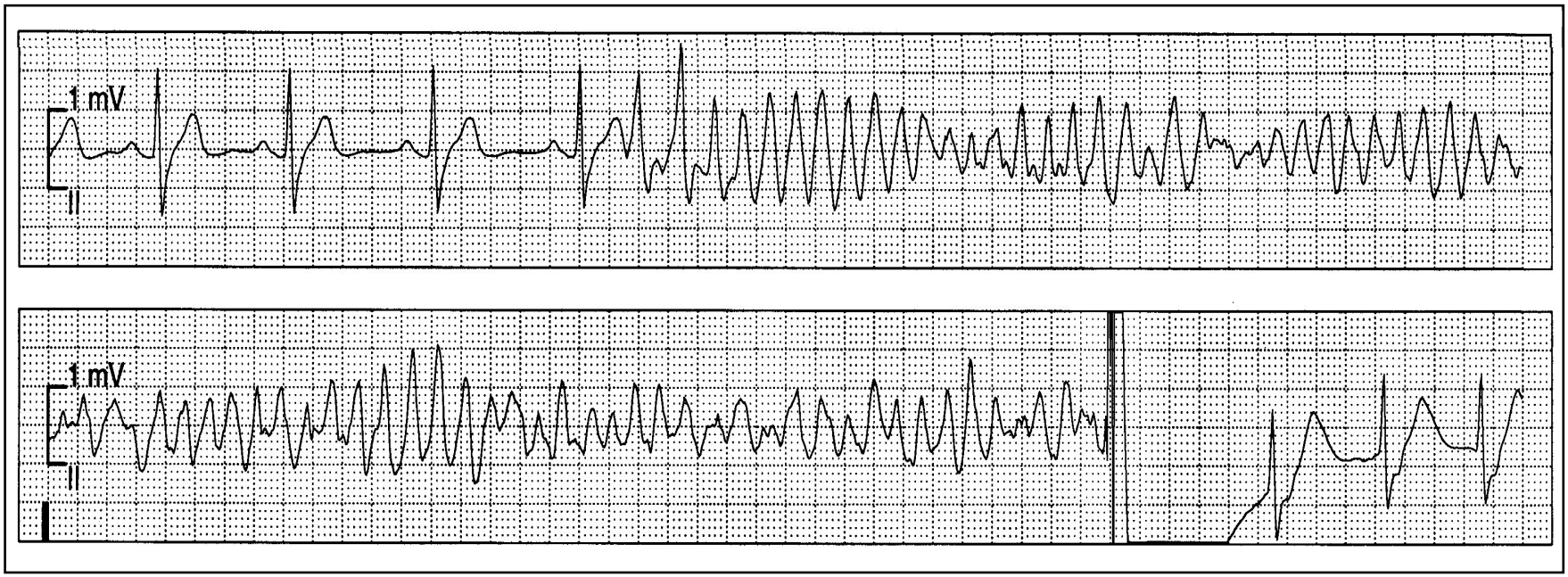




Syndrome de Brugada

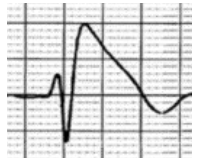
Quel est le risque ?



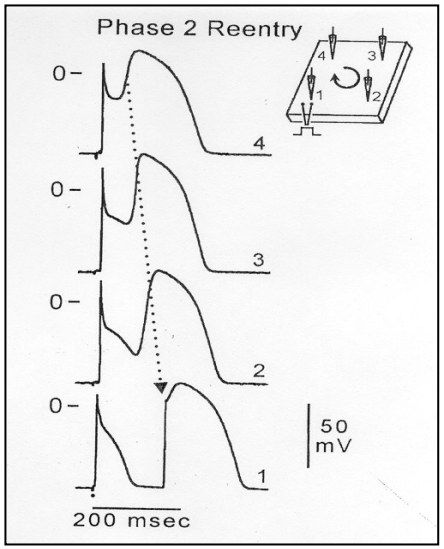


Syndrome de Brugada

Mécanismes physiopathologiques

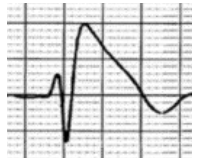


- Théorie de la dépolarisation
  - Potentiels fragmentés (chambre chasse VD)
  - Ablation épicardique du substrat *(Nademanee)*
- Théorie de la repolarisation
  - Gradient endo-épicardique
  - Réentrée de phase 2
  - ESV précoce
  - Efficacité de l'ablation du trigger *(Haissaguerre, Kakishita)*



Syndrome de Brugada

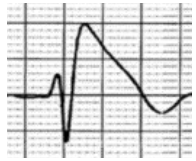
Qui puis-je rassurer ?



- Patient asymptomatique (pas d'ATCD de malaise avec PC, ni perte d'urine nocturne inexpliquée)
- Sans ATCD familial de mort subite, de pose de DAI, de Brugada avéré…
- Et sans ECG de type 1 spontané (ECG sensibilisé)

Syndrome de Brugada

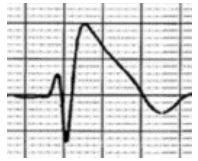
Qui est à risque ?



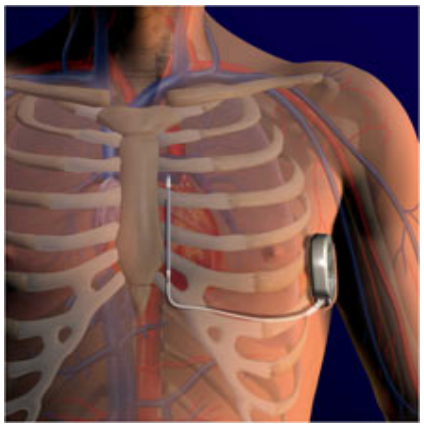
- Patient symptomatique = patient à haut risque
  - ACR récupéré
  - Syncope (y compris dans des conditions vagales)
  - Respiration agonique nocturne (perte d'urine)

Syndrome de Brugada

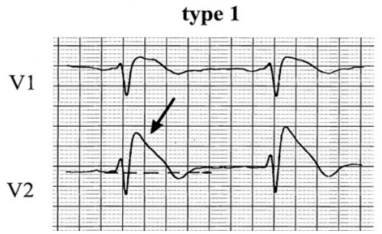
Qui est à risque ?



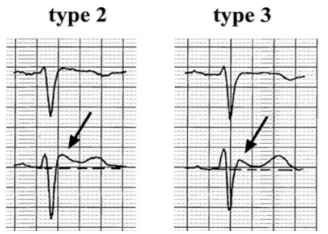
- Patient symptomatique = patient à haut risque
  - Type 1 spontané :



Défibrillateur
Automatique
Implantable

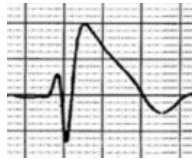


  - Suspicion de type 2 ou 3, ATCD familial : ***avis spécialisé*** (discussion de test pharmacologique)

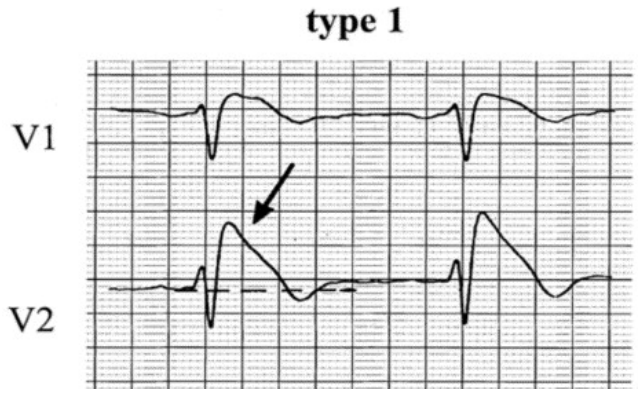


Syndrome de Brugada

Et le patient asymptomatique ?

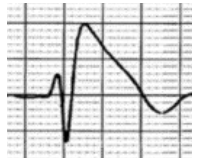


- ECG de Brugada type 1 spontané ou dans un contexte fébrile : ***avis spécialisé*** pour stratification du risque, organisation de la PEC

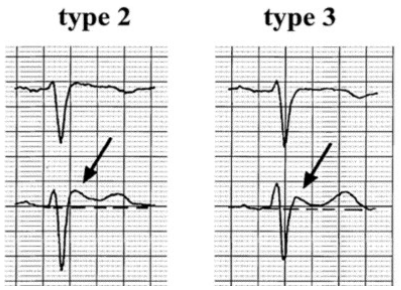


Syndrome de Brugada

Et le patient asymptomatique ?

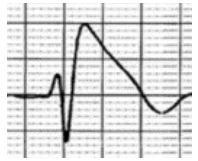


- ECG suspect de Brugada de type 2 ou 3 si ATCD familial : ***avis spécialisé***
- ECG suspect sans type 1 et sans histoire familiale : ***pas de bilan complémentaire***

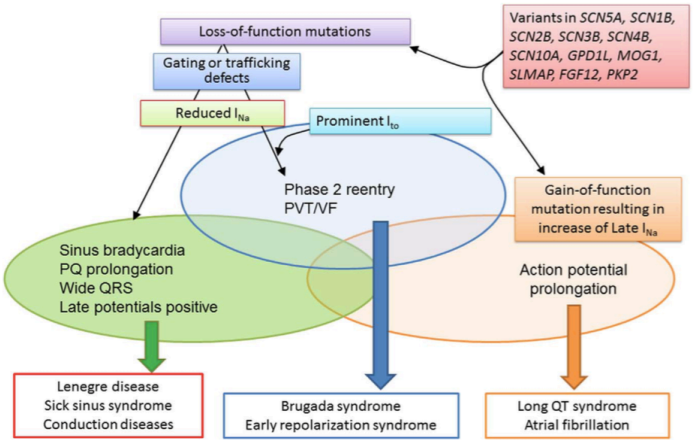


Syndrome de Brugada

Consignes (toujours)

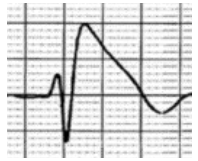


- Traitement de la fièvre
- Liste des médicaments contre indiqués
- Dépistage familial, enquête génétique
- Suivi (symptômes, évolution des PEC)

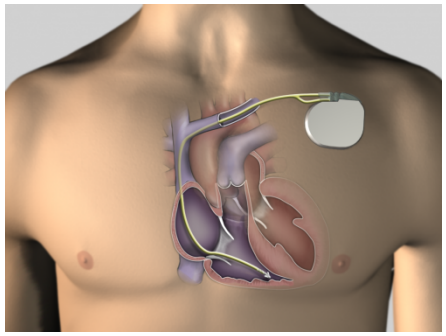


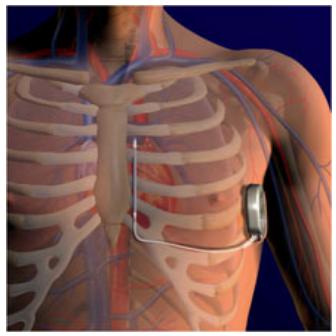
Syndrome de Brugada

Traitement (parfois)



- Haut risque : DAI

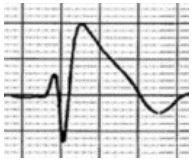


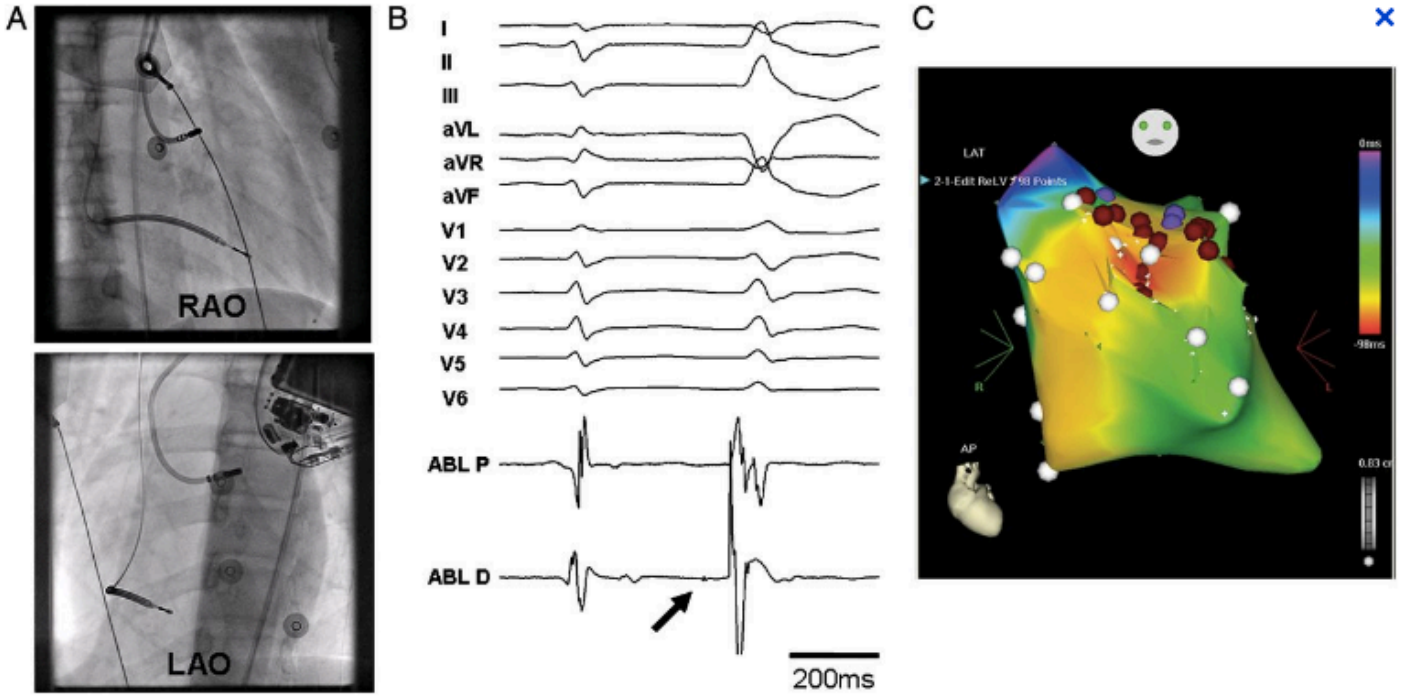


- Traitement pharmacologique : Quinidine, Hydroquinidine, Cilostazol, Bepridil…

Syndrome de Brugada

Traitement (parfois)

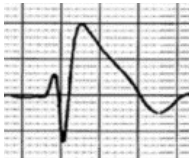


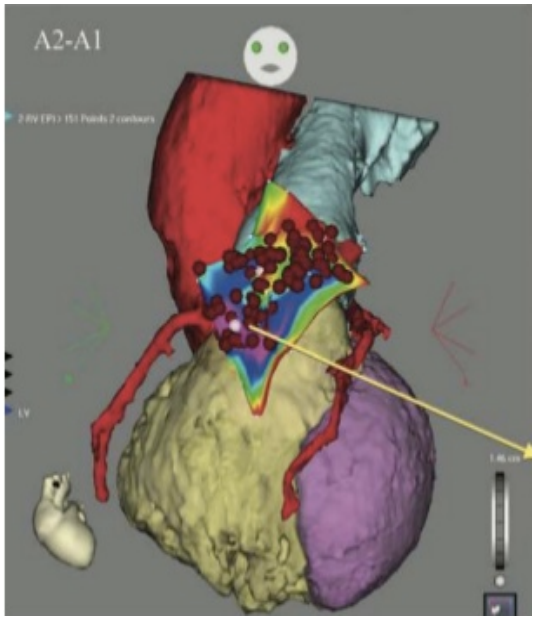


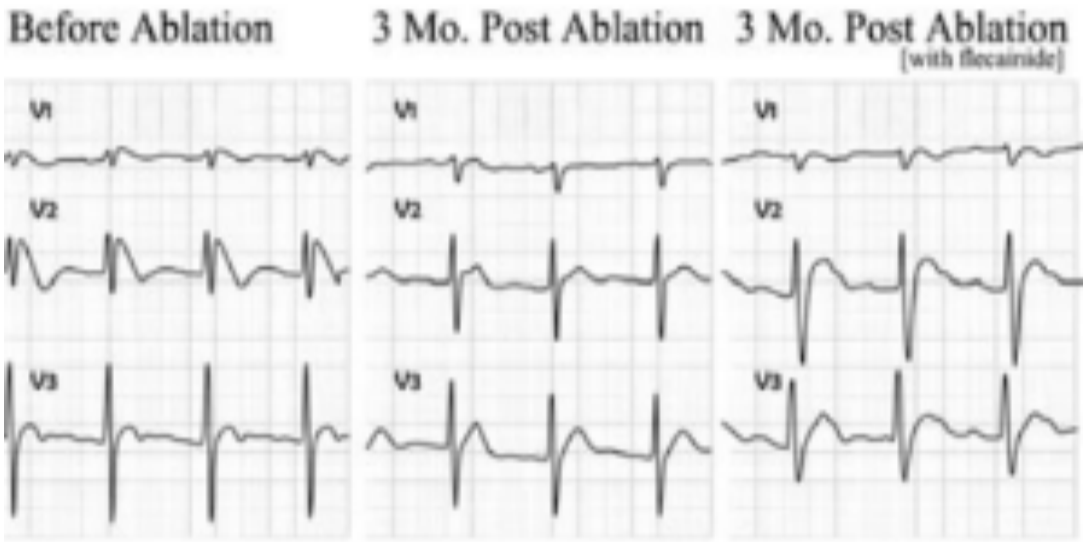
Ablation du trigger (ESV) : *Haissaguerre*

Syndrome de Brugada

Traitement (parfois)



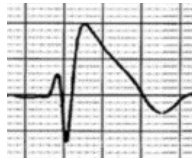




Ablation du substrat : *Nademanee*

Syndrome de Brugada

Stratégie de PEC (consensus 2016)



Type 1 Brugada pattern

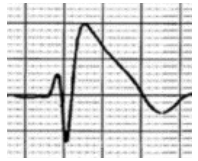
- Avoid drugs that may induce or aggravate ST segment elevation in right precordial leads (www.Brugadadrugs.org)
- Avoid cocaine and excessive alcohol intake
- Immediately treat fever with antipyretic drugs. (Class I)

Asymptomatic

Type 1 Brugada pattern induced by sodium channel blocker

Close Follow-up

Syndrome de repolarisation précoce



The NEW ENGLAND JOURNAL of MEDICINE

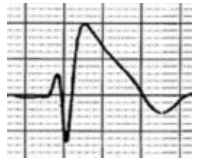
ORIGINAL ARTICLE

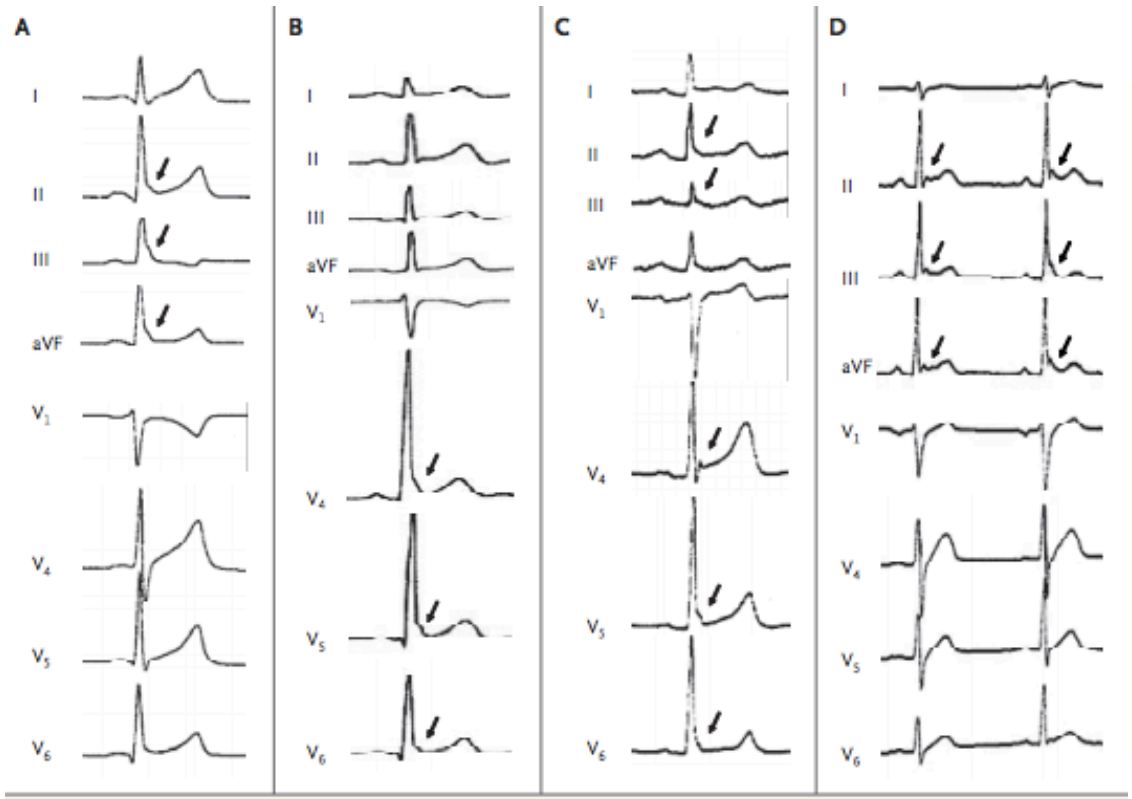
Sudden Cardiac Arrest Associated with Early Repolarization

Michel Haïssaguerre, M.D., Nicolas Derval, M.D., Frederic Sacher, M.D., Laurence Jesel, M.D., Isabel Deisenhofer, M.D., Luc de Roy, M.D., Jean-Luc Pasquié, M.D., Ph.D., Akihiko Nogami, M.D., Dominique Babuty, M.D., Sinikka Yli-Mayry, M.D., Christian De Chillou, M.D., Patrice Scanu, M.D., Philippe Mabo, M.D., Seiichiro Matsuo, M.D., Vincent Probst, M.D., Ph.D., Solena Le Scouarnec, Ph.D., Pascal Defaye, M.D., Juerg Schlaepfer, M.D., Thomas Rostock, M.D., Dominique Lacroix, M.D., Dominique Lamaison, M.D., Thomas Lavergne, M.D., Yoshifusa Aizawa, M.D., Anders Englund, M.D., Frederic Anselme, M.D., Mark O'Neill, M.D., Meleze Hocini, M.D., Kang Teng Lim, M.B., B.S., Sebastien Knecht, M.D., George D. Veenhuyzen, M.D., Pierre Bordachar, M.D., Michel Chauvin, M.D., Pierre Jais, M.D., Gaelle Coureau, Ph.D., Genevieve Chene, Ph.D., George J. Klein, M.D., and Jacques Clémenty, M.D.

N Engl J Med 2008; 358:2016-2023 | May 8, 2008 | DOI: 10.1056/NEJMoa071968

Syndrome de repolarisation précoce





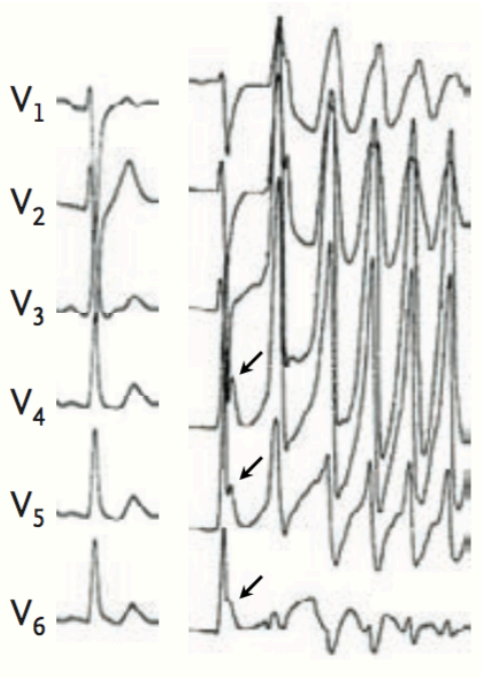
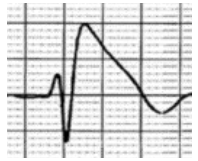


Figure 1. Baseline Electrocardiograms from Four Case Subjects.

In each panel, early repolarization is evident in the varying patterns of QRS slurring or notching in inferolateral leads (arrows). Panel D shows a beat-to-beat fluctuation in this pattern.

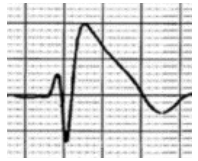
Syndrome de repolarisation précoce
Définition

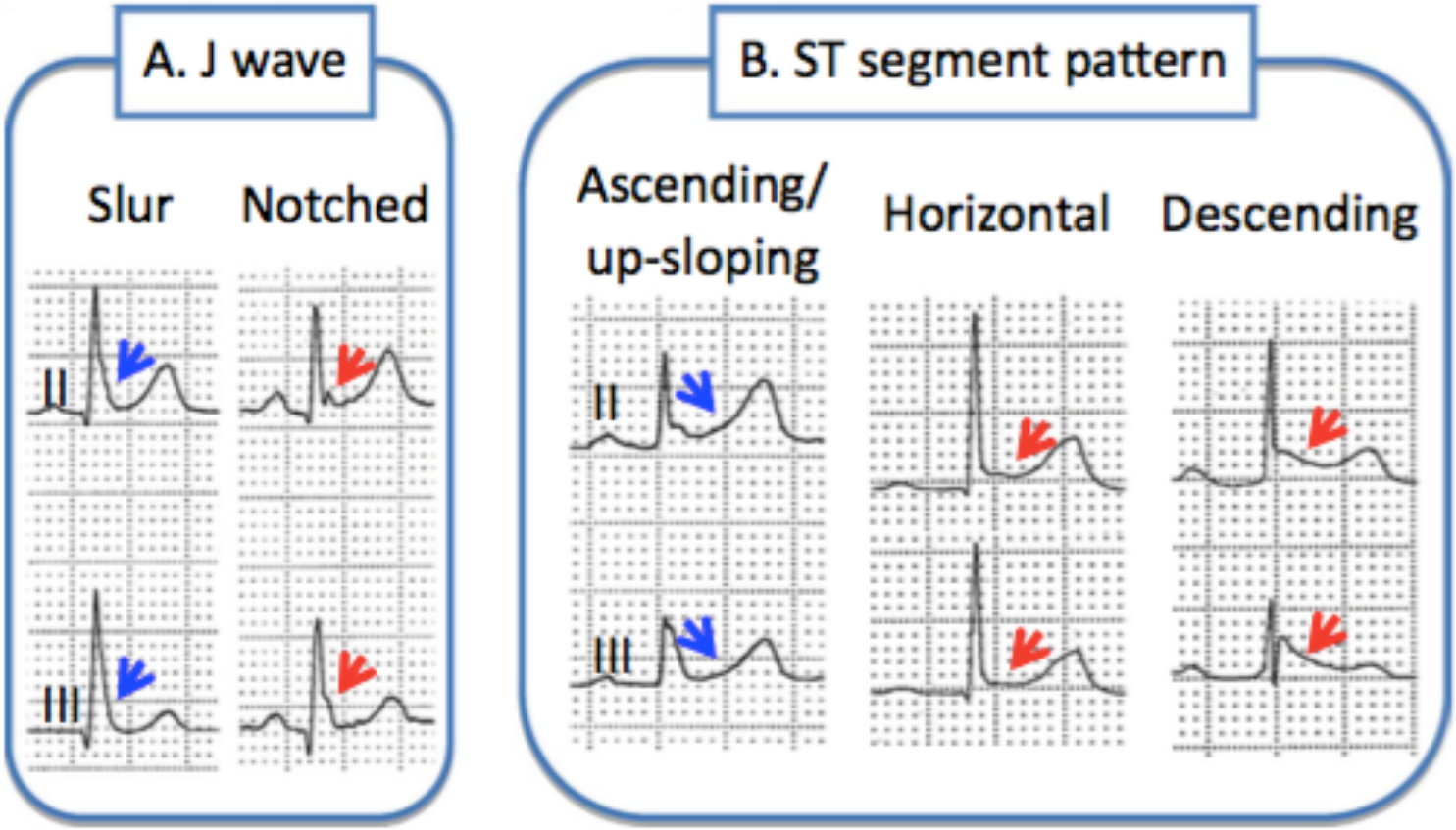


- ECG compatible (ER *pattern*) :
  - Aspect empâté ou crocheté du point J, avec onde J
  - Sus décalage du point J d'au moins 0,1mV (1mm) dans au moins deux dérivations contiguës
  - Associé ou non à un sus décalage du segment ST
- Syndrome de repolarisation précoce si association à des TV-FV en l'absence de cardiopathie sous jacente

Syndrome de repolarisation précoce

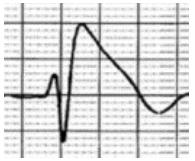
Définition

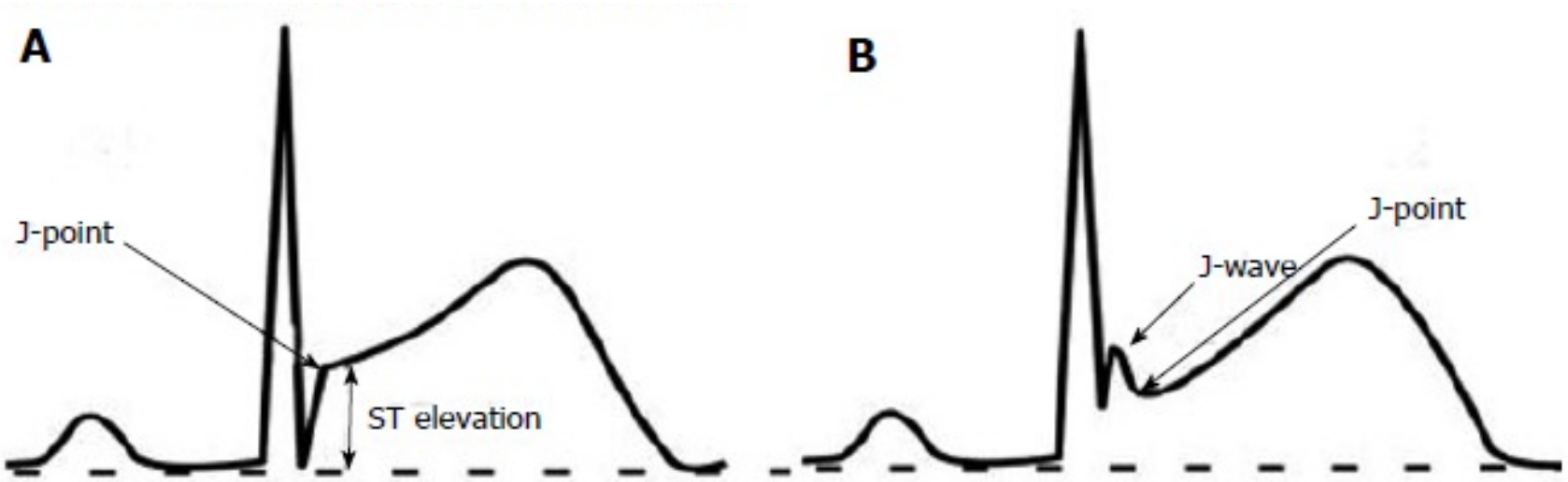




Syndrome de repolarisation précoce

Formes bénignes ou suspectes ?





Syndrome de repolarisation précoce

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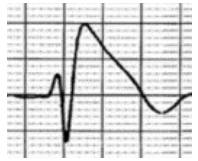
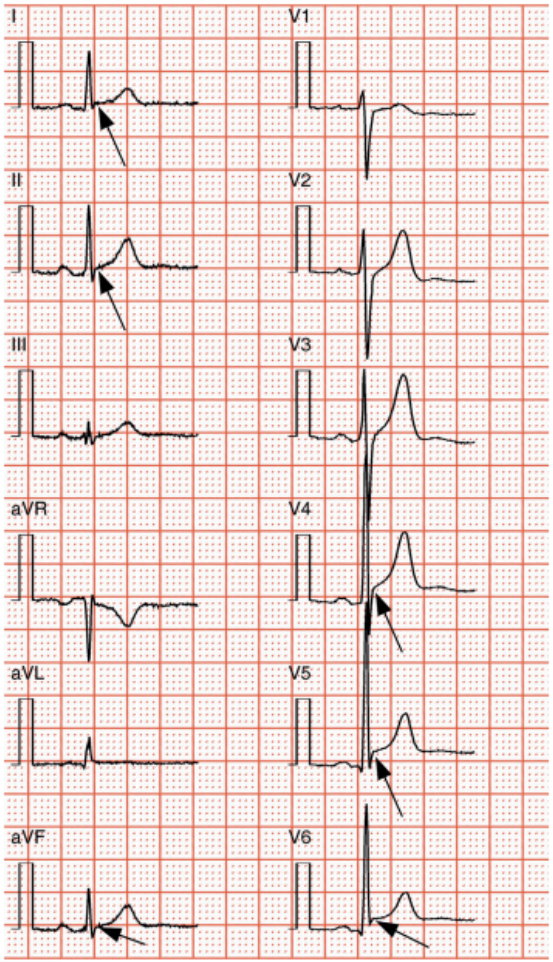
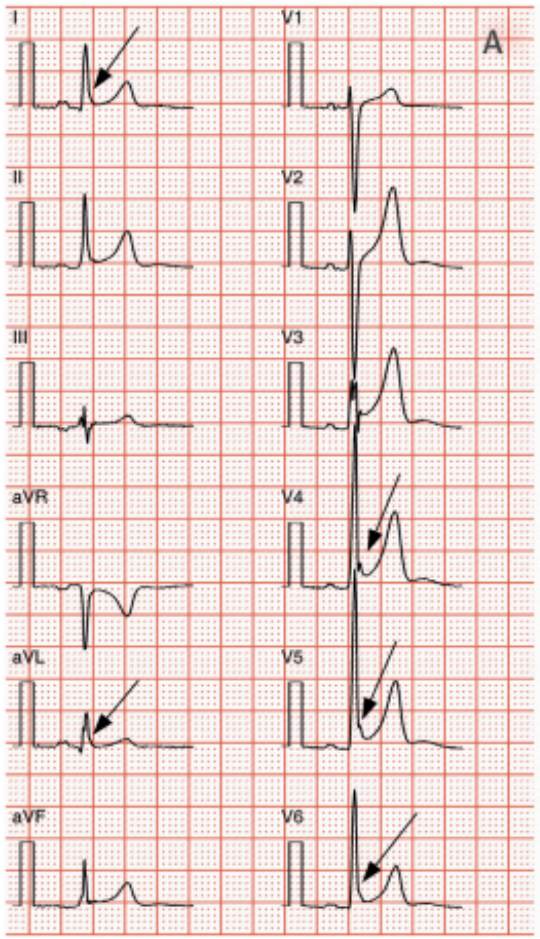
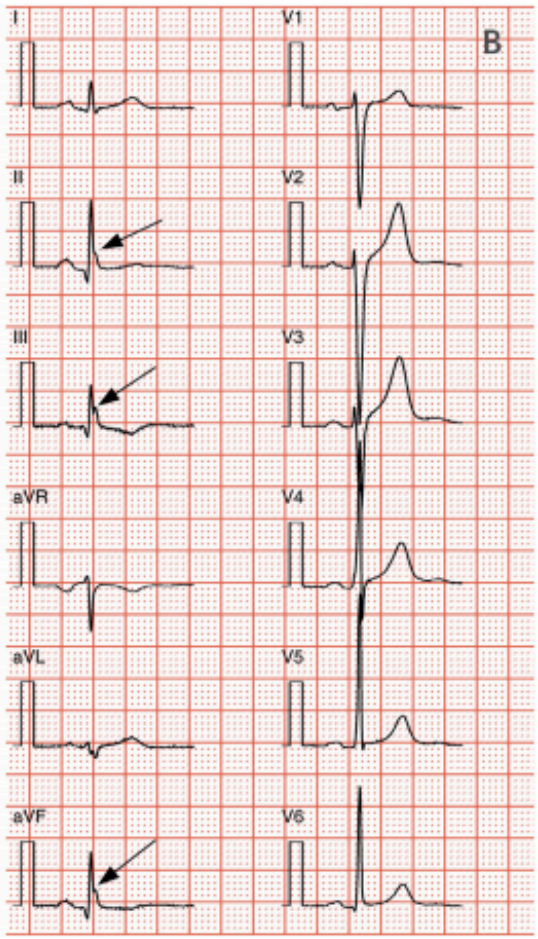


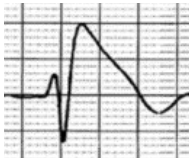


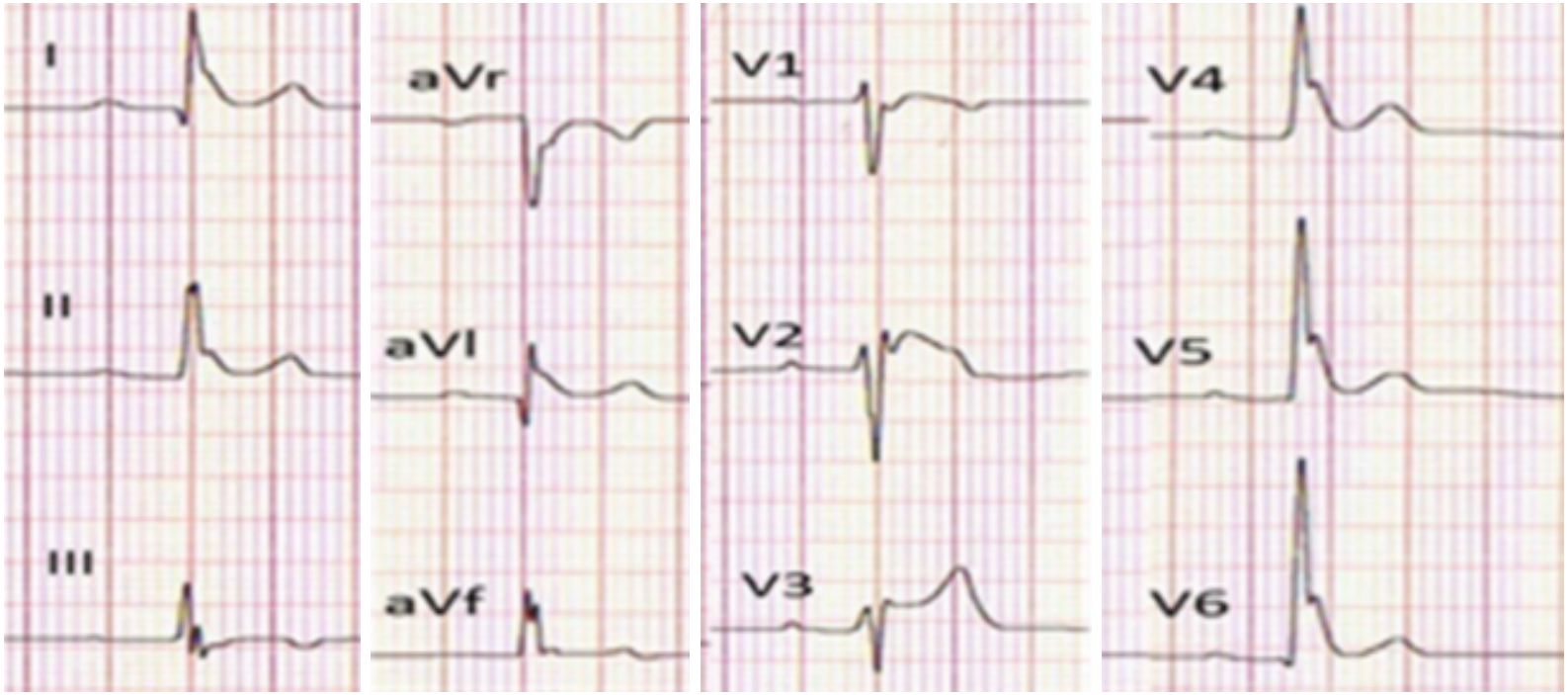




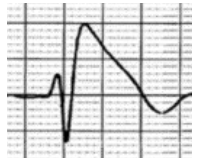
Syndrome de repolarisation précoce

Formes bénignes ou suspectes ?



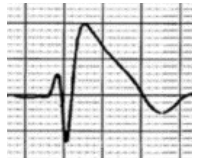


Syndrome de repolarisation précoce



- Similarités avec le syndrome de Brugada ++
  - Homme 80%
  - Premier événement clinique entre 30 et 50 ans
  - ESV précoce, risque de FV
  - Aggravation dans des conditions vagales
  - Réponse aux mêmes traitements

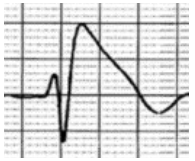
Syndrome de repolarisation précoce



- Différences Brugada – SRP
  - Ondes J en dérivations inférieures et latérales
  - Peu de sur-risque en cas de fièvre
  - Test ajmaline-flécaine peu contributif
  - Risque rythmique faible si asymptomatique

Syndrome de repolarisation précoce

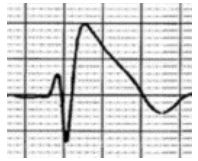
Quand l'évoquer ?

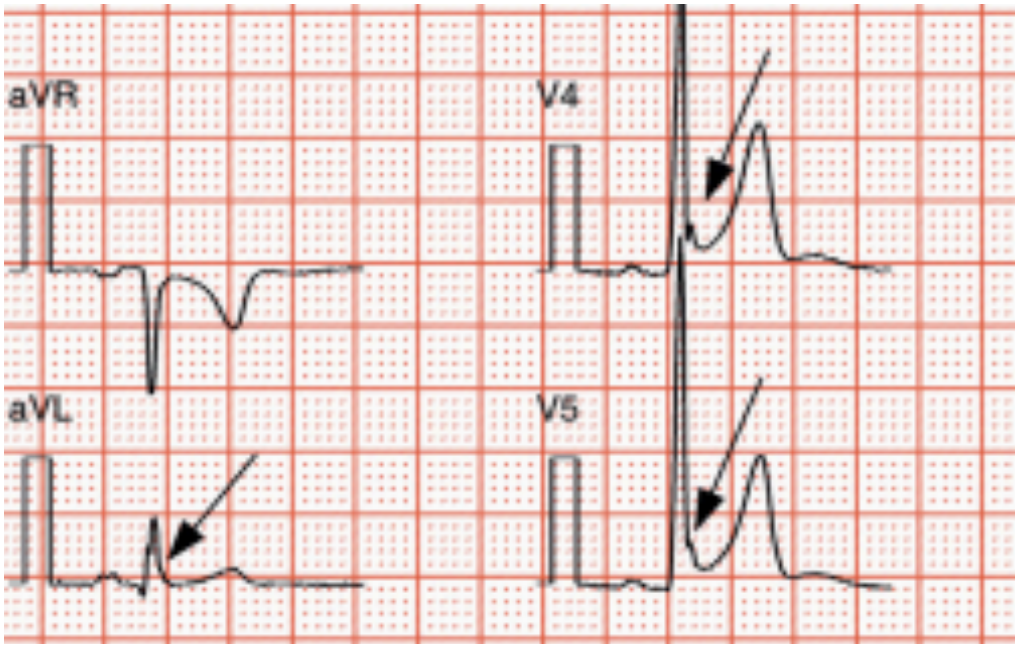


- Patient symptomatique : ACR récupéré, syncope
- Lourde hérédité familiale de mort subite

Syndrome de repolarisation précoce

Quand l'évoquer ?

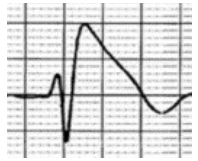




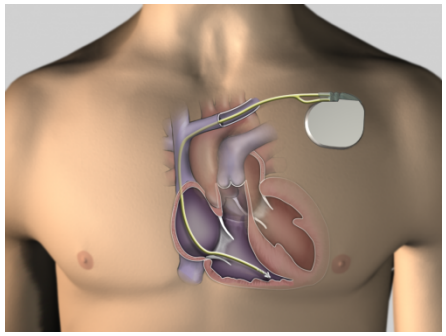
- Sus décalage du point J de 1-2mm
- ST descendant ou horizontal
- Dérivations inf

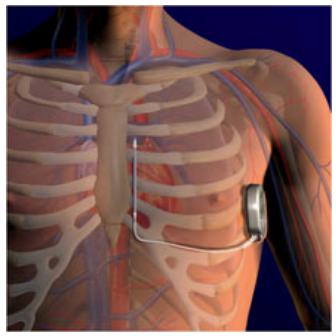
Syndrome de repolarisation précoce

Traitement



- Haut risque : DAI

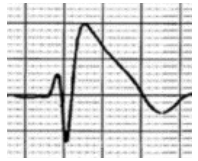


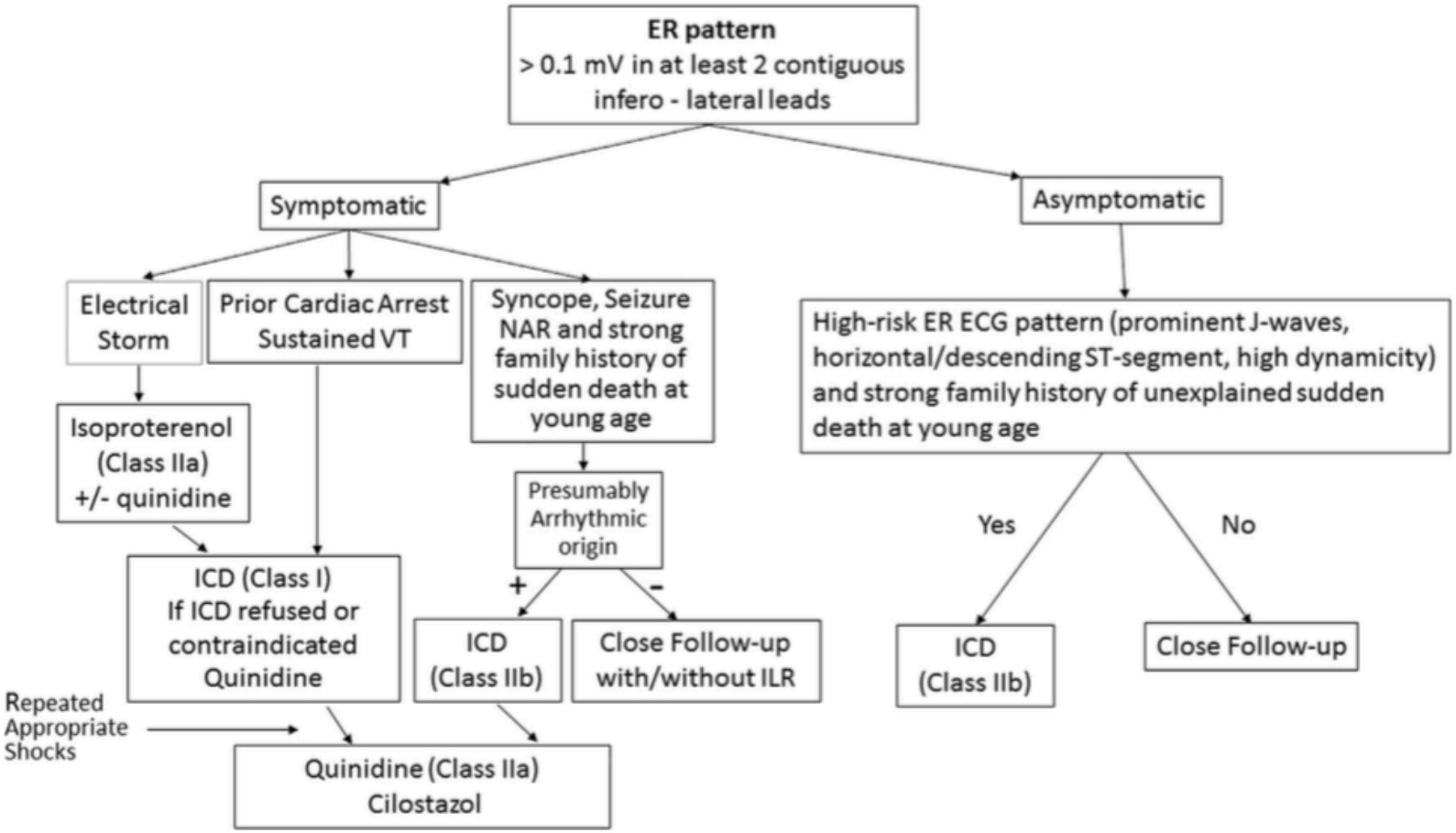


- Autres patients : pas de traitement, pas de consigne, vigilance en cas de malaise…
- Place du moniteur holter implantable ?

Syndrome de Brugada

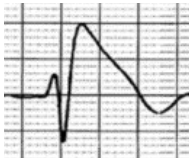
Stratégie de PEC (consensus 2016)





J-wave syndromes

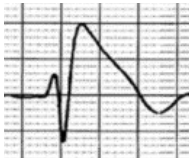
Et le sport ?



- Pas de sur-risque rythmique à l'effort
- Se méfier des hypertonies vagales post effort (pas d'arrêt brutal)
- Lutter contre l'hyperthermie (Brugada), bonne hydratation
- Contraintes liées à un éventuel DAI

J-wave syndromes

Quid de la compétition ? ESC 2005



European Heart Journal (2005) 26, 1422–1445
doi:10.1093/eurheartj/ehi325

ESC Report

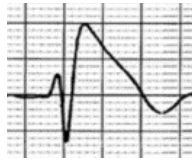
Recommendations for competitive sports participation in athletes with cardiovascular disease

A consensus document from the Study Group of Sports Cardiology of the Working Group of Cardiac Rehabilitation and Exercise Physiology and the Working Group of Myocardial and Pericardial Diseases of the European Society of Cardiology

Brugada syndrome Implanted PM	History, ECG, provocative test ECG, Echo, ET, 24 h Holter	Positive Brugada syndrome Normal heart rate increase during exercise, no significant arrhythmias, normal cardiac function	No competitive sports Low-moderate dynamic and low static sports (I A,B), except those with risk of bodily collision

J-wave syndromes

Quid de la compétition ? ACC-AHA 2015



AHA/ACC Scientific Statement

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 9: Arrhythmias and Conduction Defects

A Scientific Statement From the American Heart Association and American College of Cardiology

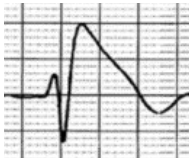
AHA/ACC Scientific Statement

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 10: The Cardiac Channelopathies

A Scientific Statement From the American Heart Association and American College of Cardiology

J-wave syndromes

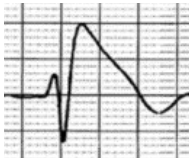
Quid de la compétition ? ACC-AHA 2015



- Pas de sport de compétition dans la phase d'évaluation du risque rythmique
- Patients porteurs de l'anomalie génétique mais sans expression phénotypique : consignes pour le sport (éviction médicamenteuse, traitement hyperthermie, hydratation…), pas de restriction de compétition. Formation des proches, acquisition d'un DSA… (IIA – C)

J-wave syndromes

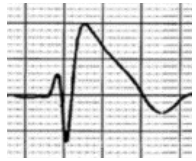
Quid de la compétition ? ACC-AHA 2015



- Patients ayant été symptomatiques ou type 1 de Brugada : compétition envisageable après 3 mois sans symptôme (IIB – C)
- Patients porteurs d'un DAI : compétition envisageable après 3 mois sans événement rythmique (IIB – C)
- Un DAI ne doit pas être posé dans le but de participer à un sport en compétition.

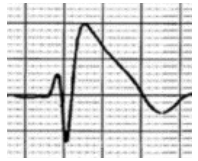
J-wave syndromes

Sport et DAI

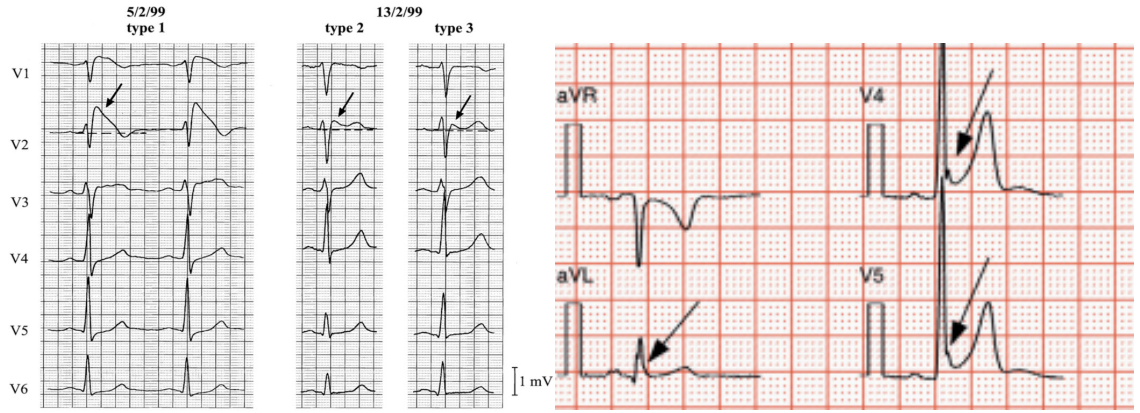


J-wave syndromes

Take Home Message

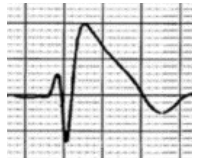


- A évoquer si ATCD familial de MS, ou patient symptomatique (syncope, même dans des conditions vagales)
- Avis spécialisé (canalopathies, DAVD, CMH…)

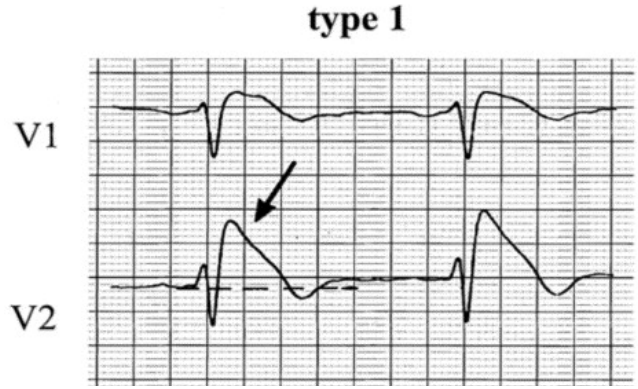


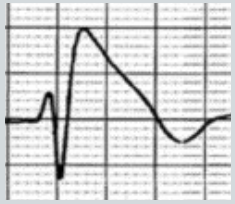
J-wave syndromes

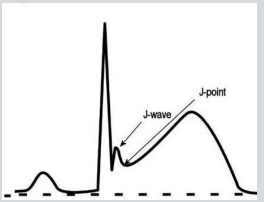
Take Home Message



- Patient asymptomatique, sans ATCD familial, éliminer un type 1 de Brugada







J-wave syndromes

Syndromes de Brugada et de repolarisation précoce

Club Mont Blanc
Coeur et Sport

CCS
CLUB DES CARDIOLOGUES DU SPORT

Merci de votre attention !

CH ANNECY GENEVOIS