

# Anomalies ECGS de la repolarisation ventriculaire

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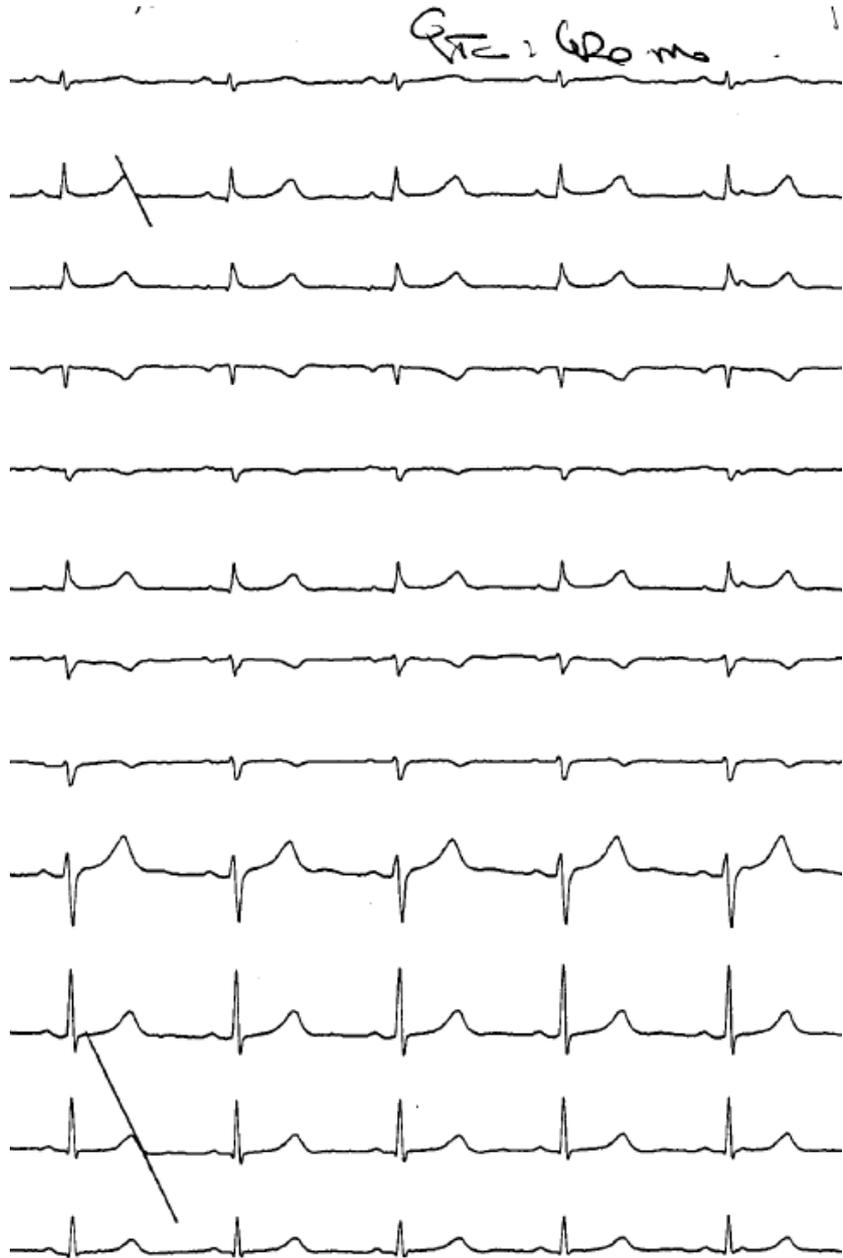
*Centre de Référence*

*Maladies Cardiaques Héritaires*

*Filière Cardiogen*

*Séminaire NEM 22 03 2018*





**Garçon de 15 ans**

**Syncope**

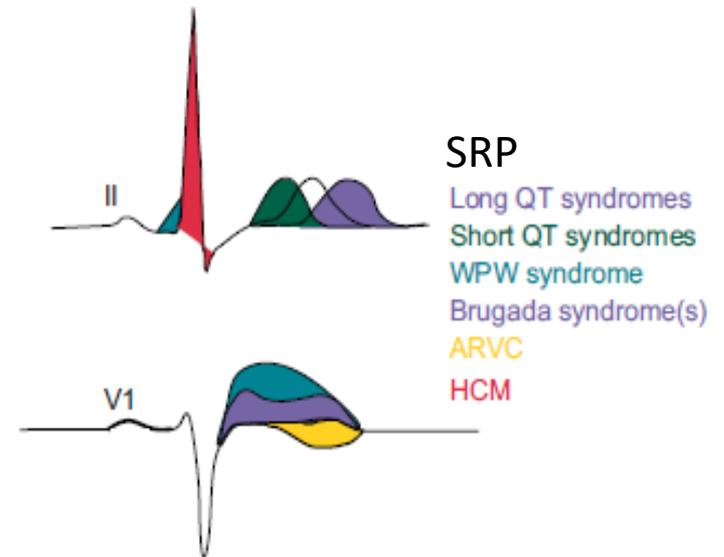
**Circonstances ?**

# Repolarisation ventriculaire

- Durée de l'intervalle QTc ?
- Morphologie de l'onde T ?

=> Moyens :

- Ecg 12 dérivations
- Holter
- Epreuve d'effort
- Tests de sensibilisation



**“a prolonged and a shortened mean QTc interval over 24 hours was associated with a more than twofold risk of sudden death compared with intermediate QTc values (400-440 ms)”**

<i>QTc duration (ms)</i>	<i>Sudden death rate at 2 years (%) (proportion)*</i>	<i>Relative risk† (95% confidence interval)</i>
<b>Lead V2:</b>		
<400	2.2 (26/1204)‡	1.3 (0.8 to 2.4)
400–440	1.6 (38/2365)	—
≥440	3.0 (37/1218)	1.9 (1.2 to 3.4)
<b>Lead V5:</b>		
<400	1.7 (22/1276)	1.0 (0.6 to 1.9)
400–440	1.7 (36/2150)	—
≥440	2.9 (36/1261)	1.7 (1.0 to 3.0)
<b>Leads I, II, and III:</b>		
<400	1.7 (25/1491)	1.0 (0.6 to 1.8)
400–440	1.7 (39/2350)	—
≥440	3.8 (35/917)	2.3 (1.4 to 4.1)

\*Denominator estimated as number of patients from random sample multiplied by inverse of sampling fraction, 6693/268, and corrected by 241/245 because four cases of sudden death were excluded.

†Ratio of sudden death rates in which the rate of a category was used as the reference relative risk was taken as reference.

<b>QT scale.</b>	
<b>Males</b>	<b>Females</b>
470	480
<b>Very long QT.</b> LQTS even if asymptomatic. Exclude II <sup>o</sup> causes	
450	460
<b>Long QT.</b> LQTS when supported by symptoms, family history or additional tests.*	
390	400
<b>Long QT possible.</b> Additional tests when indicated:* Repeated ECG, Holter, T-wave morphology, exercise, epinephrine-challenge, adenosine-challenge.	
360	370
<b>Normal QT.</b>	
330	340
<b>Short QT.</b> SQTS when supported by symptoms or family history. Additional tests: Repeated ECG, Holter, T-wave morphology (?), electrophysiologic studies (?)	
<b>Very short QT.</b> SQTS even if asymptomatic. Exclude II <sup>o</sup> causes	

**Sami Viskin. Heart Rhythm 2009.**

# Mesure de l'intervalle QT

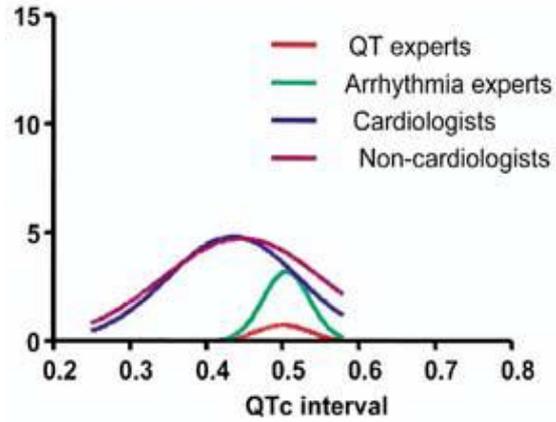


Mesure de l'intervalle QT corrigé (formule Bazett)

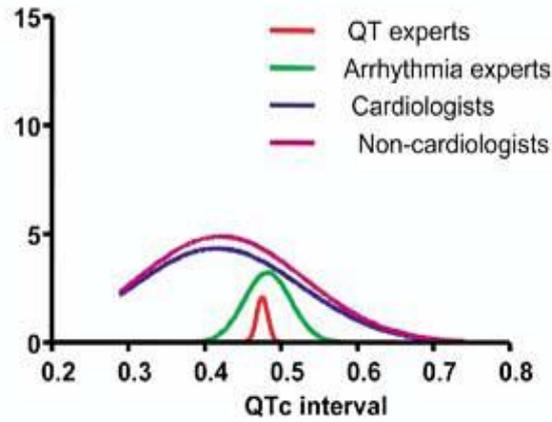
$$QT_c = \frac{(QT1 / \sqrt{RR1}) + (QT2 / \sqrt{RR2}) + (QT3 / \sqrt{RR3})}{3}$$

# QTc Measurement

Trace 1: Distribution of QTc values

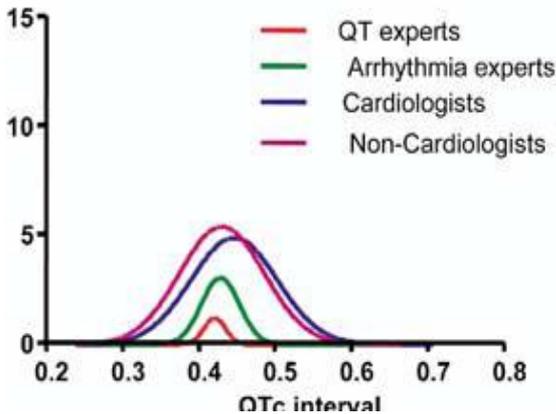


Trace 2: Distribution of QTc values

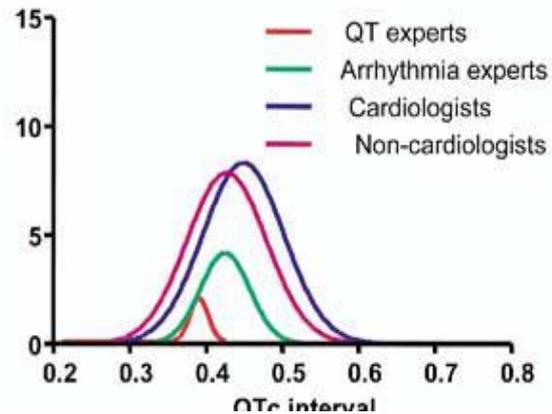


**Long QT syndrome :  
QTc under estimated**

Trace 3: Distribution of QTc values



Trace 4: Distribution of QTc values



**Normal QT :  
QTc over estimated**

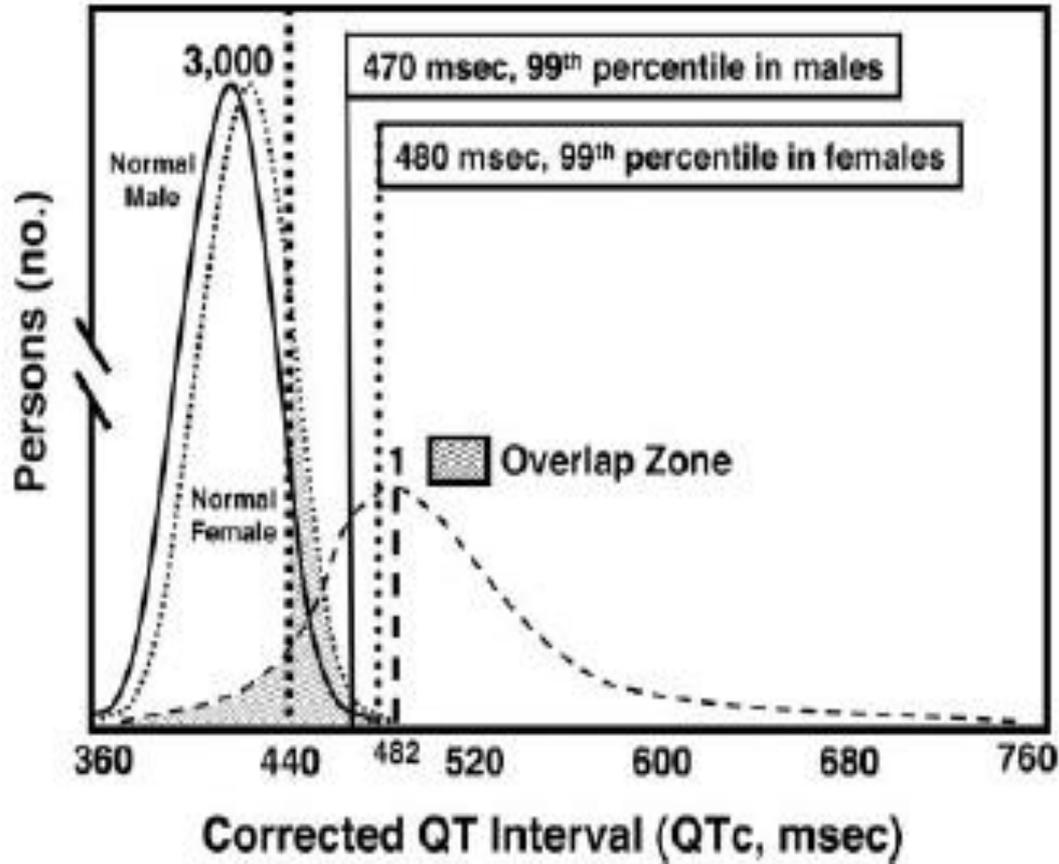
# Causes de QT allongé

- **Facteurs de risques non médicamenteux** :
  - sexe féminin, age avancé
  - troubles électrolytiques (hypokaliémie et hypomagnésémie)
  - insuffisance cardiaque
  - bradycardie
  - ischémie
  - **syndrome congénital du QT long**
  
- **Causes médicamenteuses** :

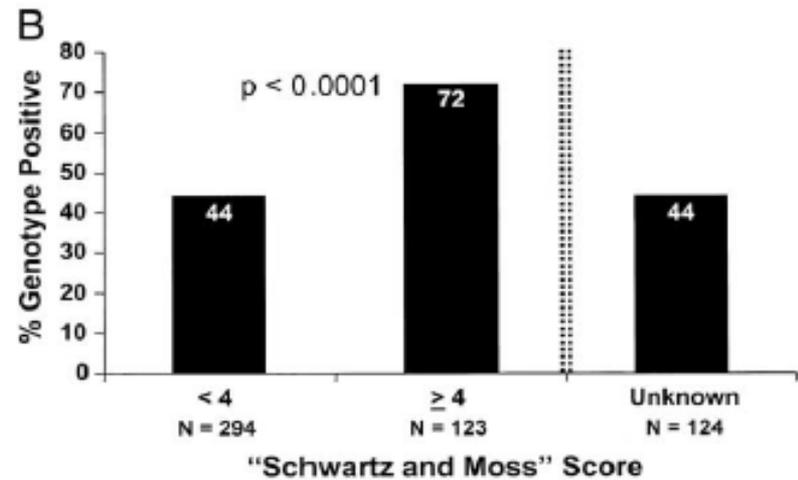
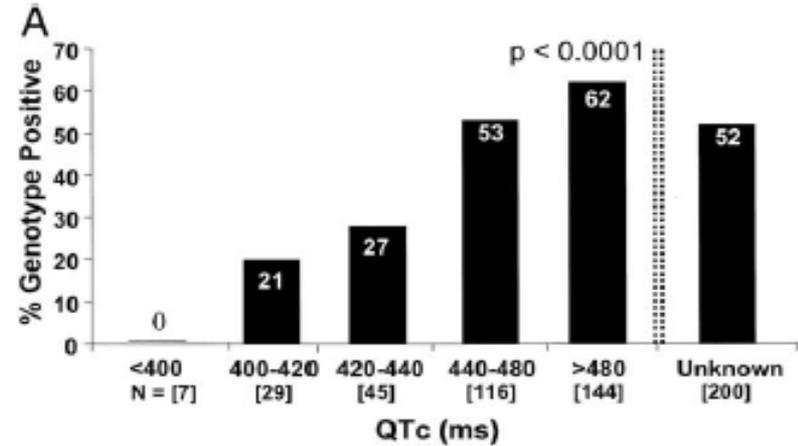
Agents anti-arythmiques : classe IA (quinidine, disopyramide, procaïnamide), classe II (sotalol et dans une moindre mesure amiodarone)

  - antidépresseurs : surtout tricycliques
  - neuroleptiques (halopéridol)
  - antibiotiques : macrolides, quinolones
  - antihistaminiques
  - agents antifongiques : fluconazole, itraconazole, kétaconazole

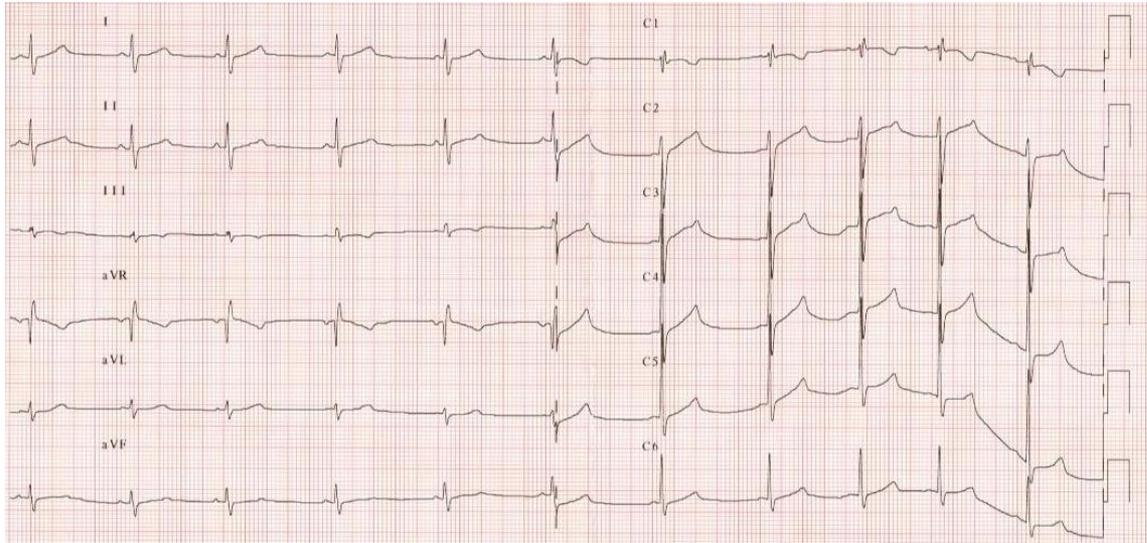
# Limitation of QTc



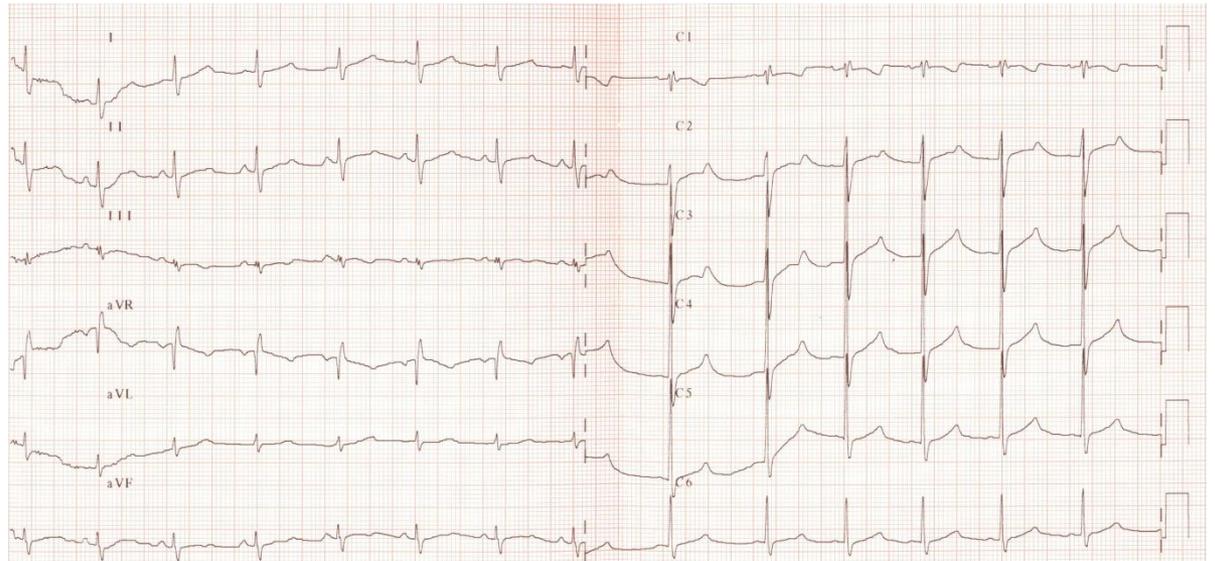
Taggart et al. Circulation  
2007;115



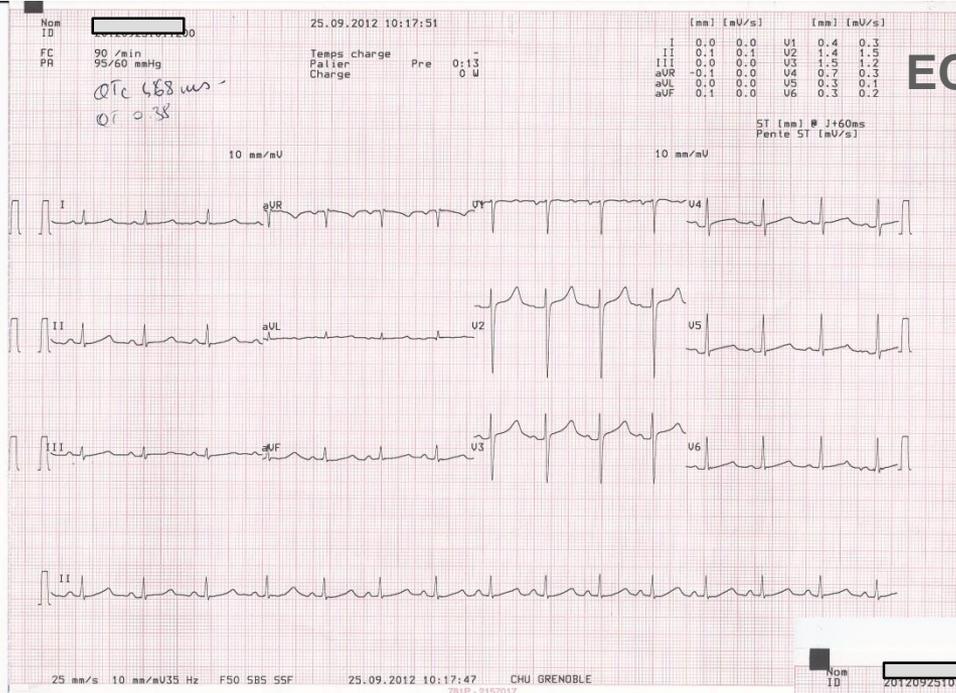
Tester et al.  
JACC 2006;47



**ECG allongé : QTc 440ms**

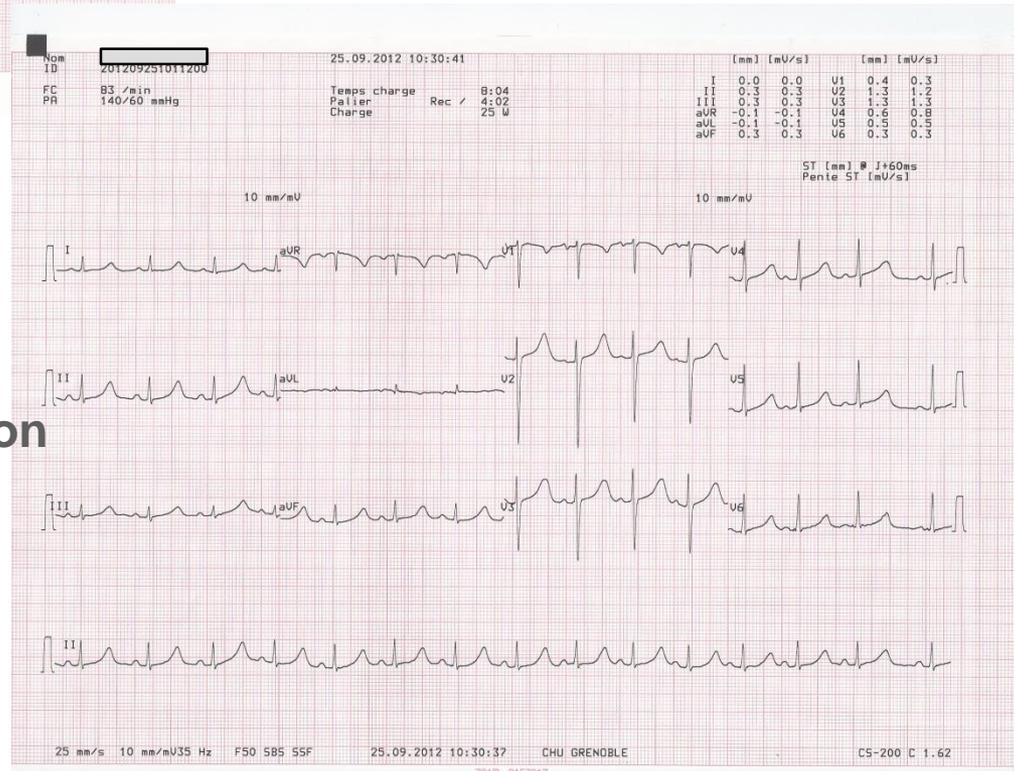


**ECG debout : QTc 470ms**



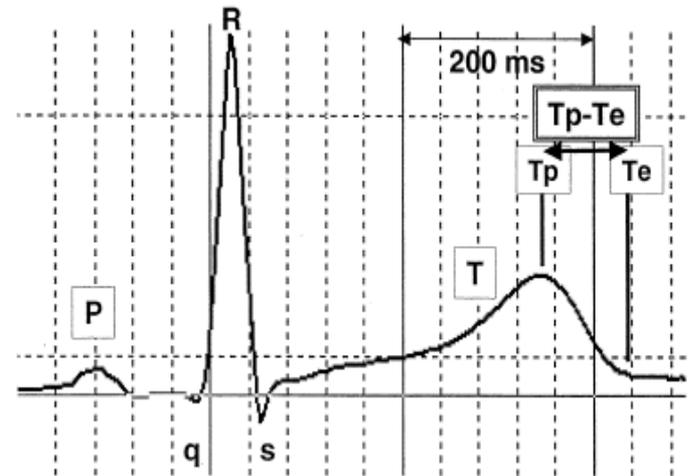
# ECG de base

## 4<sup>ème</sup> mn de récupération

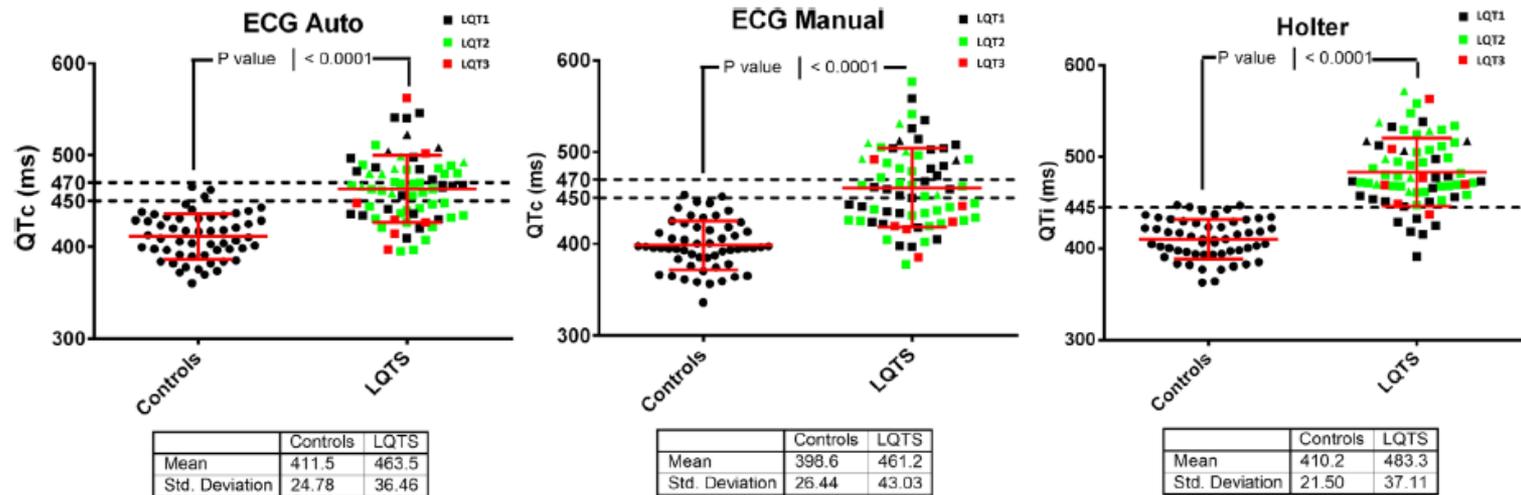


# Rechercher les anomalies de la repolarisation

- **Dynamique :**
  - Pentas QT/RR jour < nuit (Holter)
    - Merri et al, Circulation 1992
    - Neyroud et al, Eur Heart J, 1998
  - QTp/QTe ↗
    - Extramiana et al, Am J Cardiol 2005
    - Viitasalo et al, JACC 2006
- **Morphologie :**
  - ECG de surface
    - Moss et al, Circulation 1995
    - Zangh et al, Circulation 2001
  - Moyennage en fct FC (Holter)
    - Lupoglazoff et al, Circulation 2001



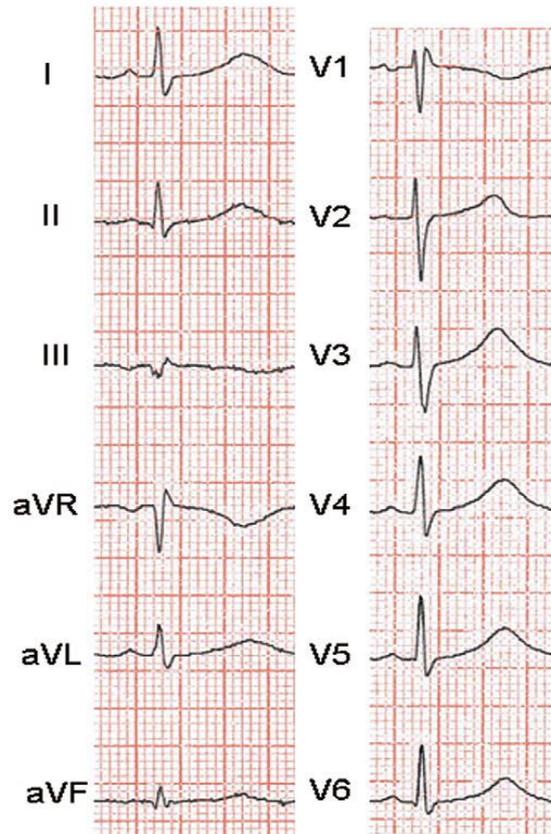
# Holter mieux que l'ECG



**Figure 2** Different QTc measurement techniques in controls and LQTS mutation carriers. Automated (left panel) and manual (middle panel) QT interval measurements with correction for heart rate using the Bazett formula and an individualized QT correction using a linear QT-RR regression obtained from 24-hour Holter data (right panel) in controls and LQTS mutation carriers. Error bars in red indicate mean and SD. In the LQTS group, colors indicate different genotypes (black = long QT type 1; green = long QT type 2; red = long QT type 3); triangles indicate symptomatic patients; and squares indicate asymptomatic patients. Dashed lines indicate cutoff values: 470 ms for women and 450 ms for men for QTc and sex-independent cutoff of 445 ms for QT<sub>i</sub>. LQTS = long QT syndrome; QT<sub>c</sub> = rate corrected QT interval using the Bazett formula; QT<sub>i</sub> = individualized corrected QT interval.

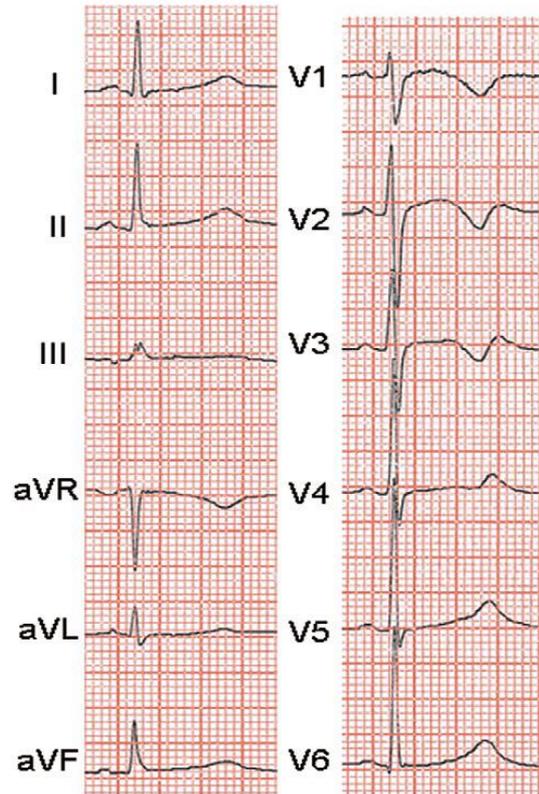
# Morphologies anormales onde T

**A**



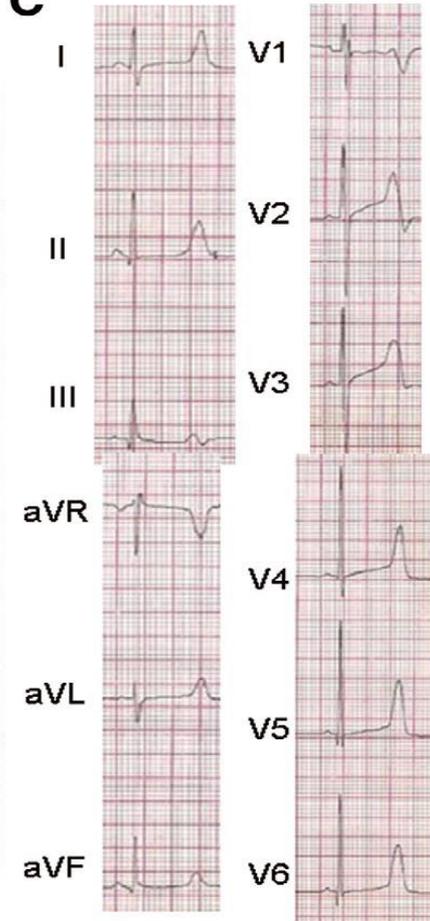
LQT1

**B**



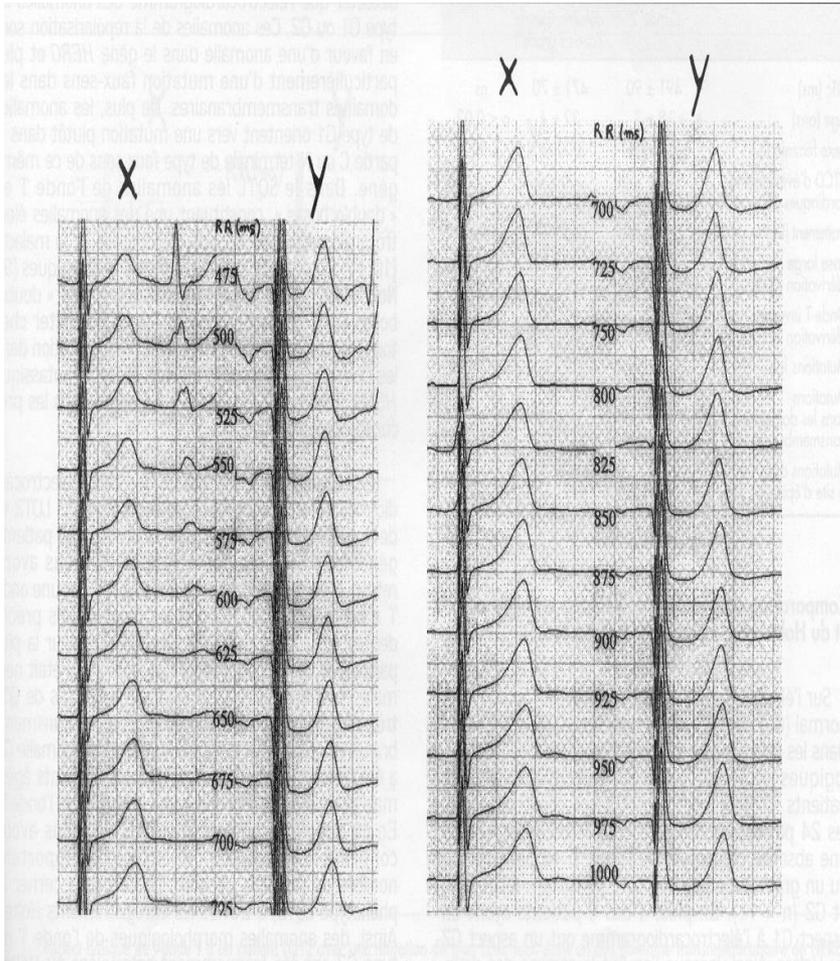
LQT2

**C**

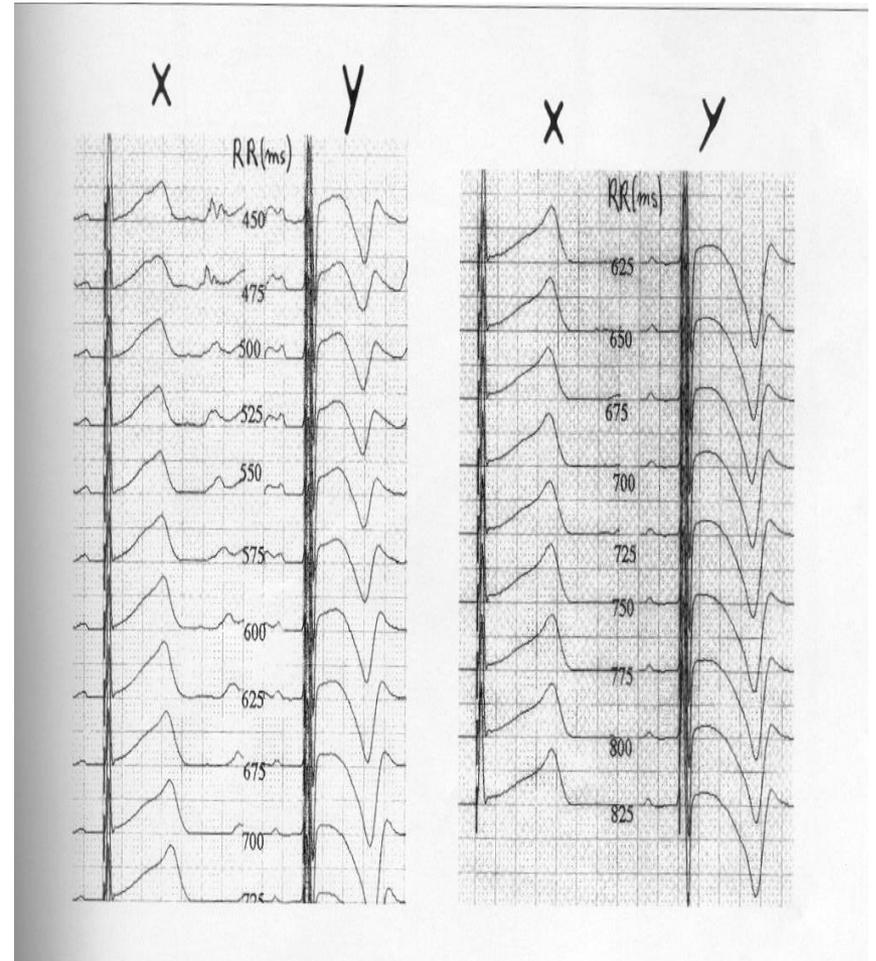


LQT3

# Morphologie de l'onde T : Holter

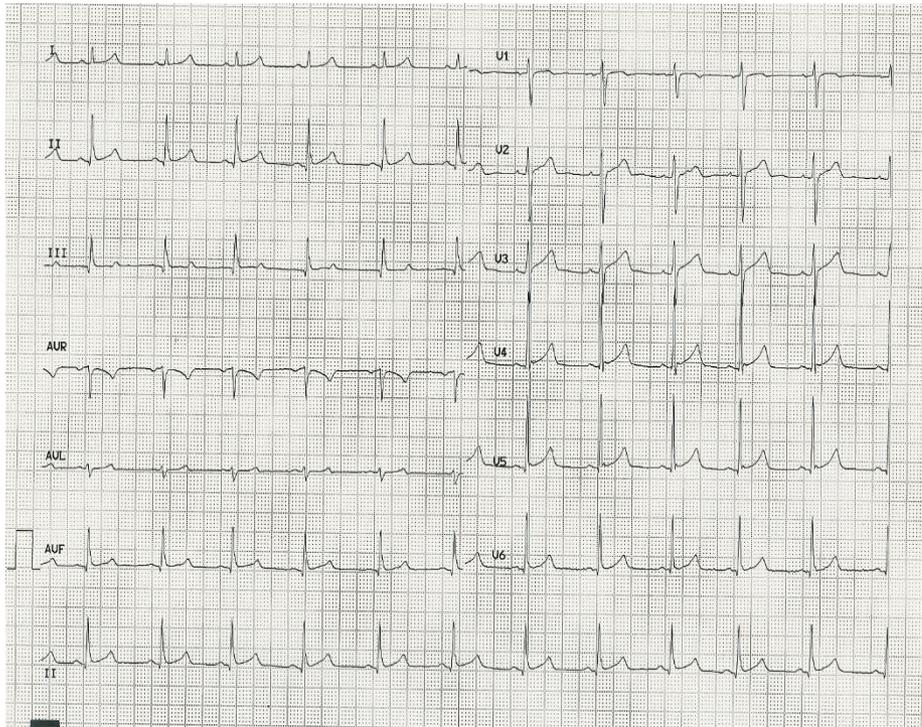


Forme LQT1

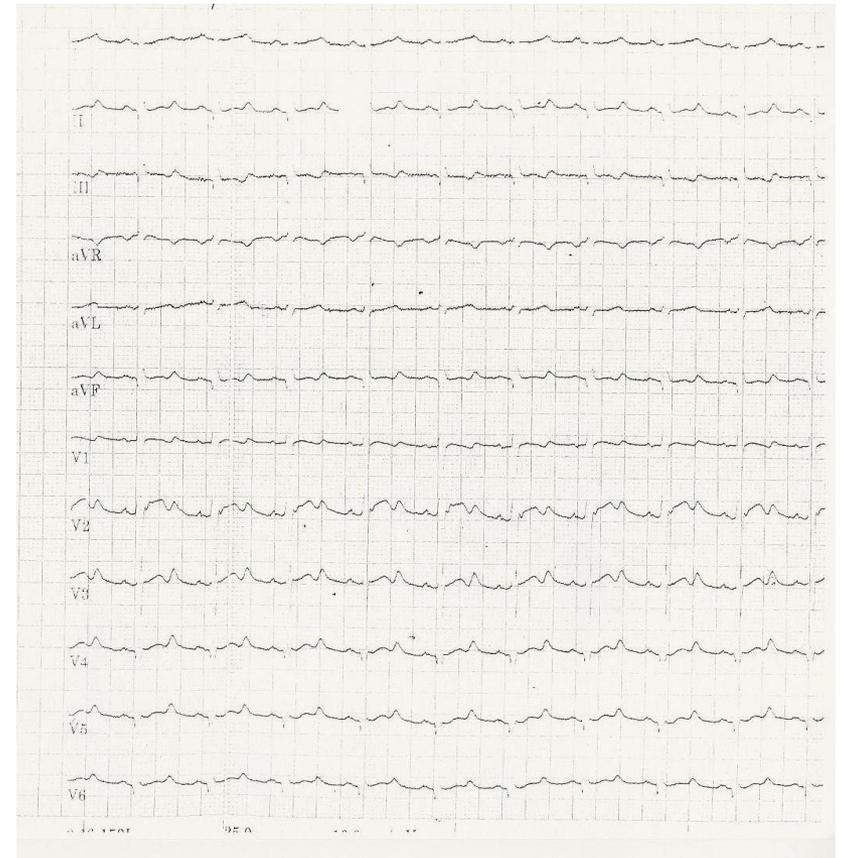


Forme LQT2

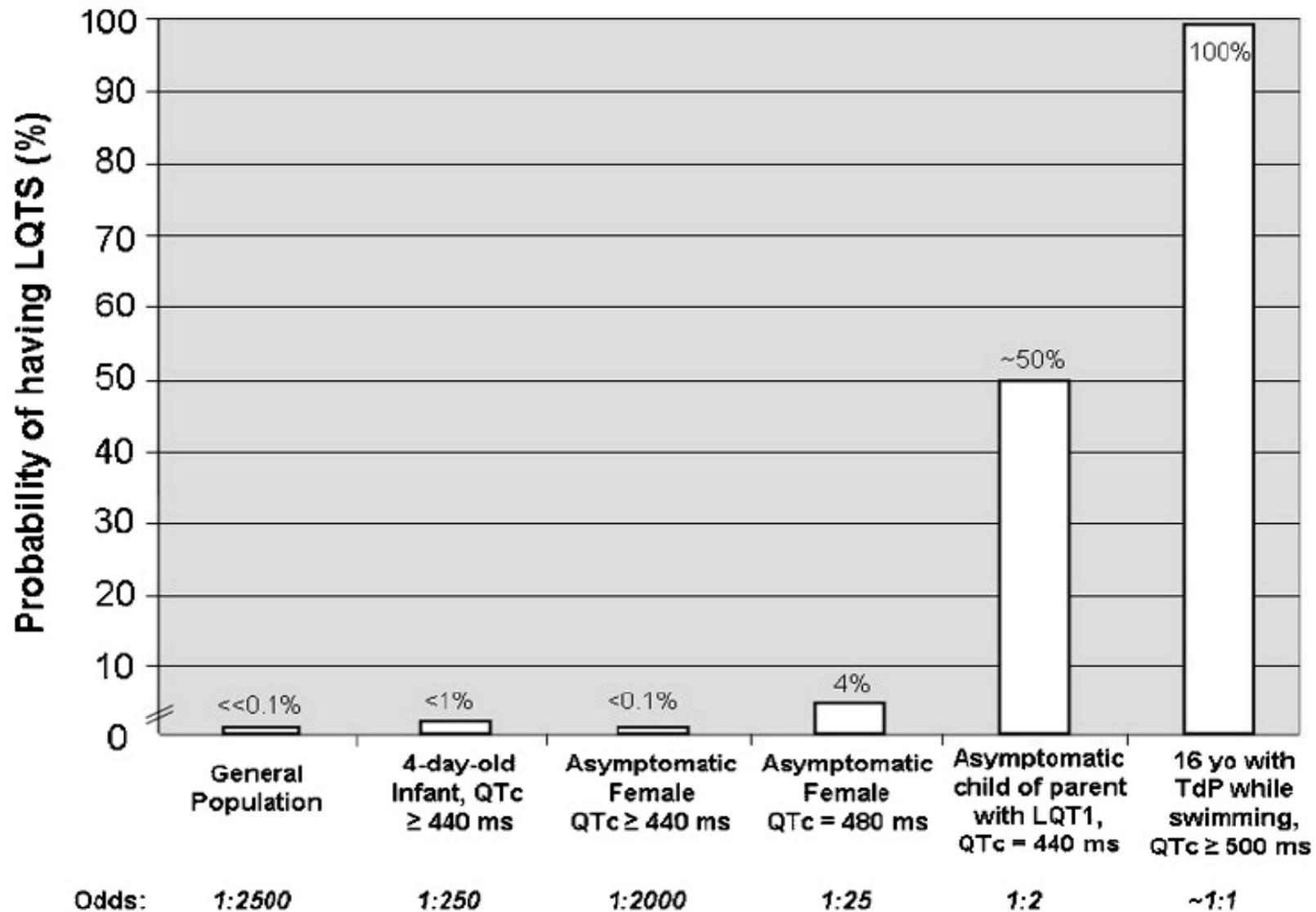
## Jeune fille de 12 ans : syncope brève



Ecg de repos : QTc 450 ms

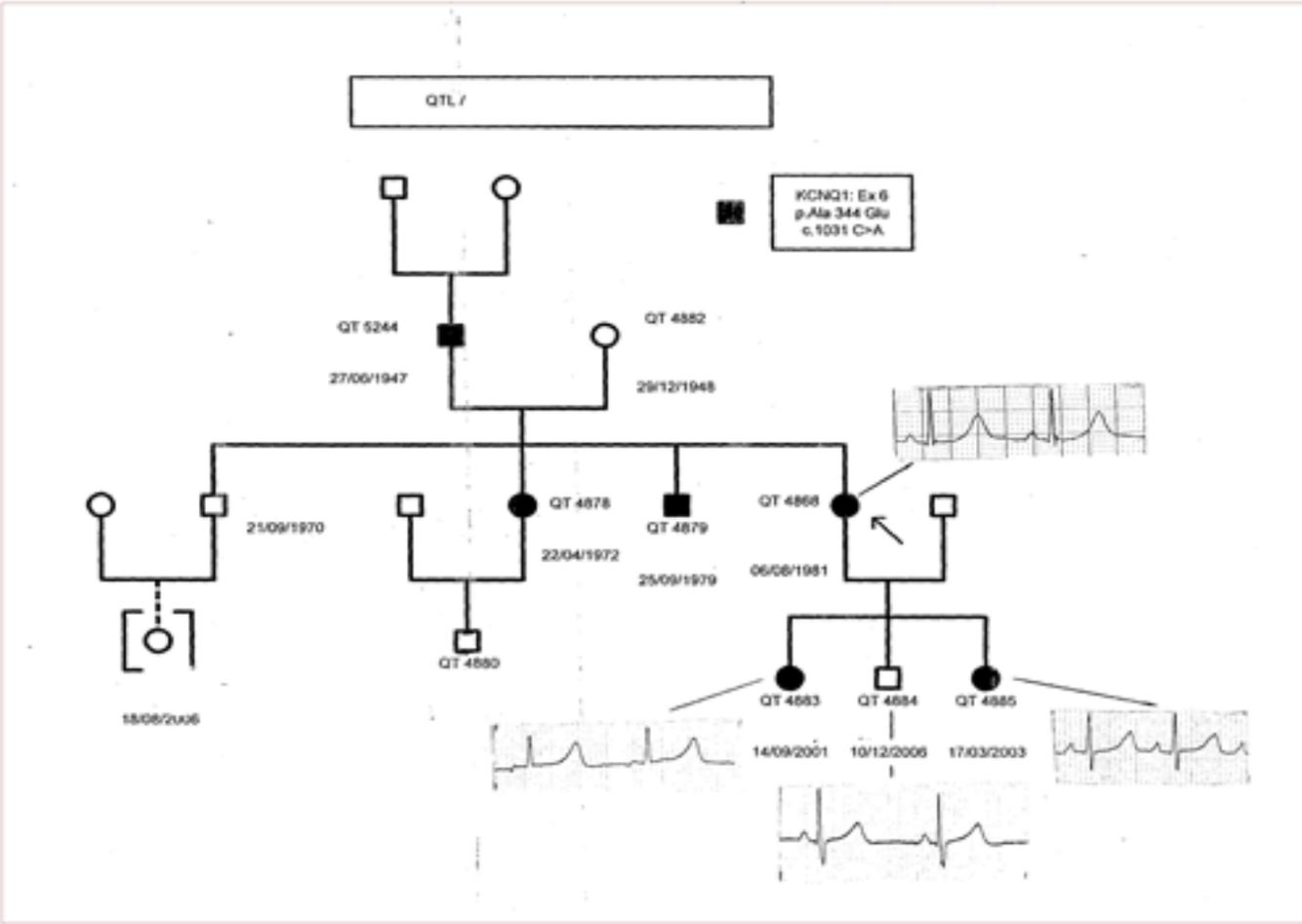


Test adrenaline : QTc 530 ms



*JN Johnson & MJ Ackerman Br J Sports Med 2009;43:657–62.*

# Etude familiale



# Diagnostic

**1. a: score  $\geq 3.5$**

**1. c: QTc  $\geq 500$  ms**

1. Le SQTL est diagnostiqué :

- a. En présence d'un score de risque  $> 3$  en l'absence d'une cause secondaire à l'allongement du QT, et/ou
- b. En présence d'une mutation pathogène dans un des gènes du SQTL quelle que soit la valeur du QTc, ou
- c. En présence d'un QTc  $\geq 480$  ms sur l'ECG 12 dérivation et en l'absence d'une cause secondaire d'allongement du QT.

2. Le SQTL peut être diagnostiqué en présence d'un QTc  $\geq 460$  ms sur l'ECG 12 dérivation chez un patient ayant présenté une syncope inexplicée en l'absence de cause secondaire à l'allongement du QT et en l'absence d'une mutation pathogène.

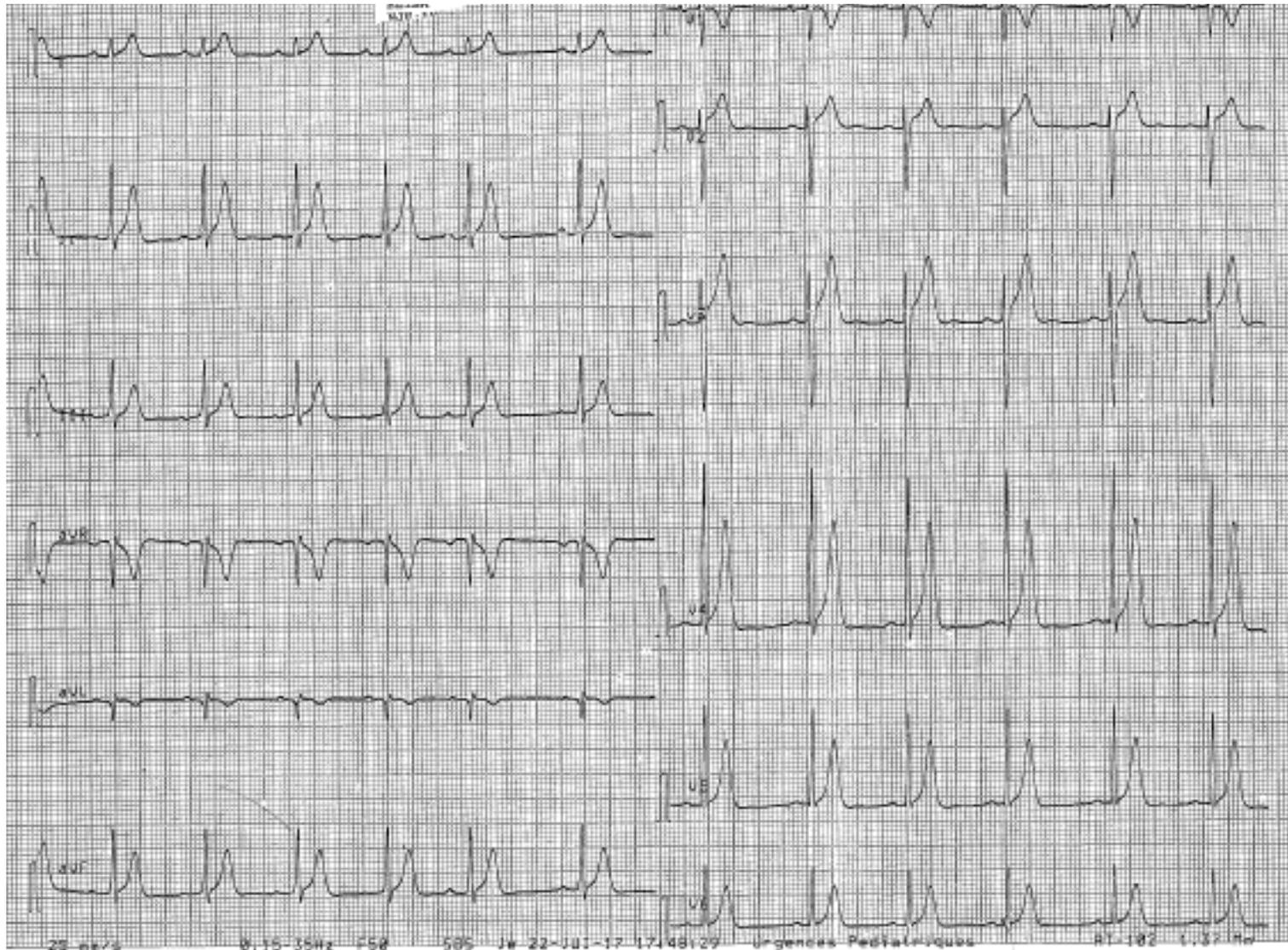
**2 . QTc entre 480 et 499 ms**

# Fille de 4 ans, ECG pour bradycardie



QTc = 320 ms; morphologie anormale T

## Fille de 10 ans, ECG pour le sport



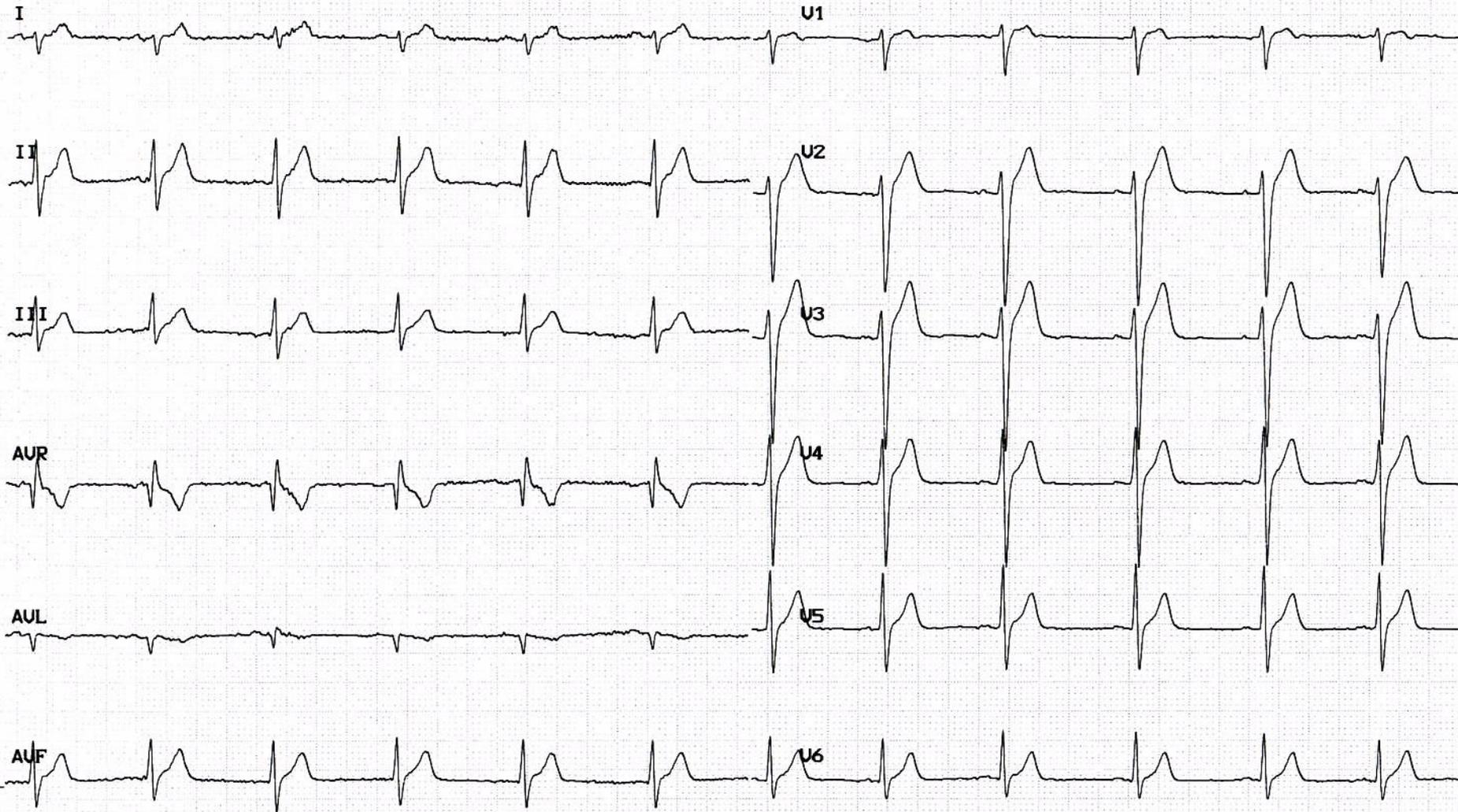
**QTc = 330 ms**

## Causes de QT court

- l'hypercalcémie, l'hyperkaliémie
- la tachycardie, l'acidose
- les catécholamines
- l'acétylcholine
  
- **le syndrome du QT court**

Éliminer causes secondaires : acidose, hyper K, hyper Ca, fièvre, tachycardie, dysautonomie, catécholamines, acétylcholine, imprégnation ou surtout surdosage en digitaliques

15 ans, asymptomatique

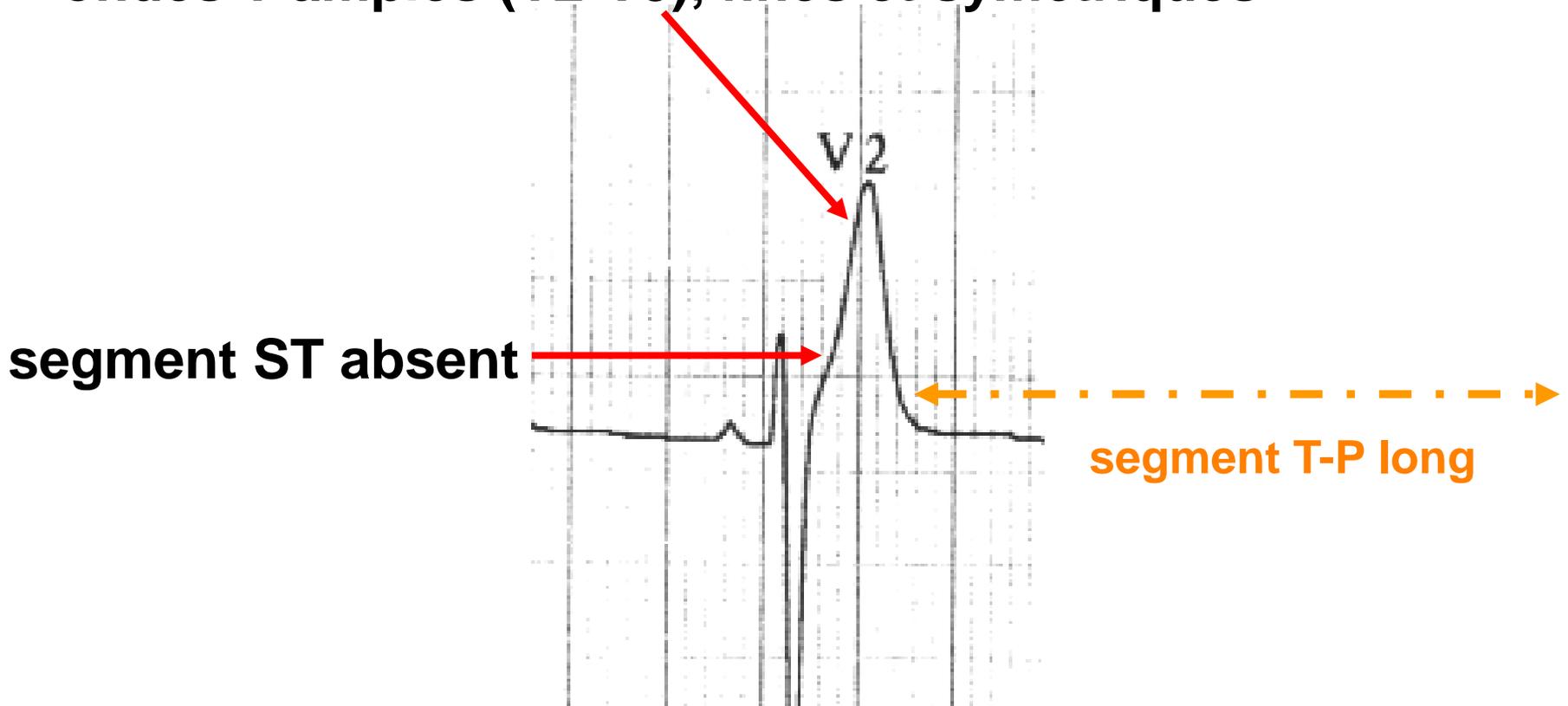


QT 290 ms, QTc 320 ms

*QT = 76 % QT théorique*

# Mesure QT court

ondes T amples (V2-V5), fines et symétriques



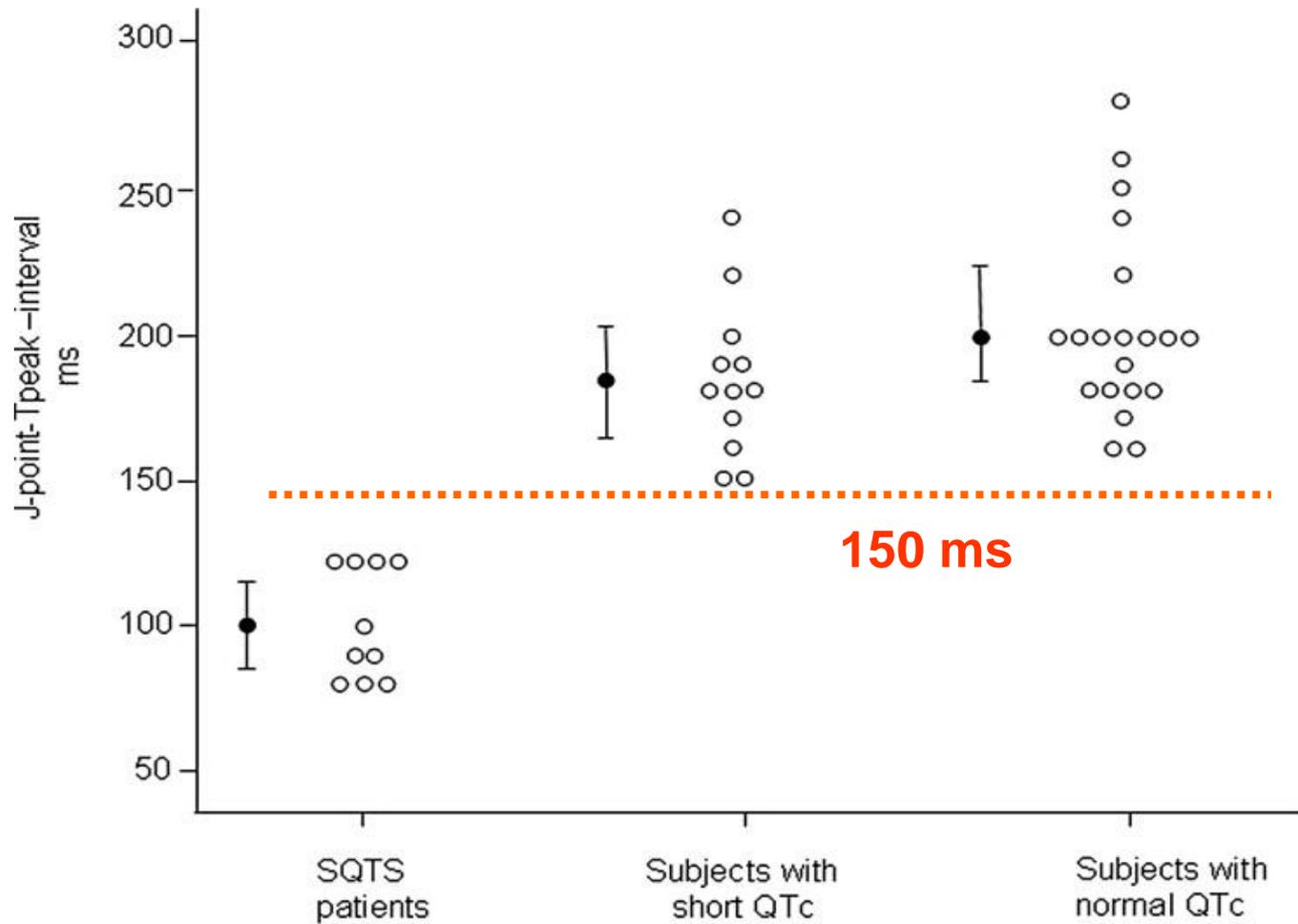
# Durée et morphologie

10 SQT avec MS

12 QT<320 ms  
asymptomatiques  
suivis 30 ans

20 témoins

<u>QTc</u>	317 ± 27ms	314 ± 14ms (biais?)	NS	405 ± 28 ms
<u>T amplitude</u>	1.2 ± 0.5 mV	1.1 ± 0,5 mV	NS	0.6 ± 0,3 mV
<u>T peak-T end/QT</u>	0.30 ± 0.04	0.24 ± 0.05	p=0,001	0.24 ± 0.04
			ns	
<u>J-T peak</u>	101 ± 18ms	184 ± 27ms	p< 0,001	203 ± 33ms
			ns	



**pas d'overlap**

# Anomalies morphologiques onde T



**SQTS 1**

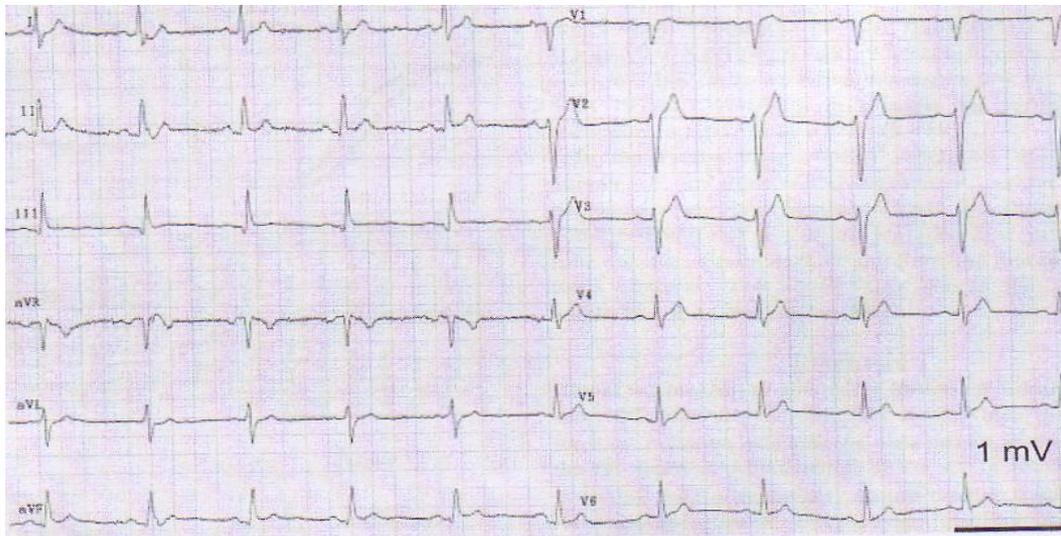
(QTc 320)



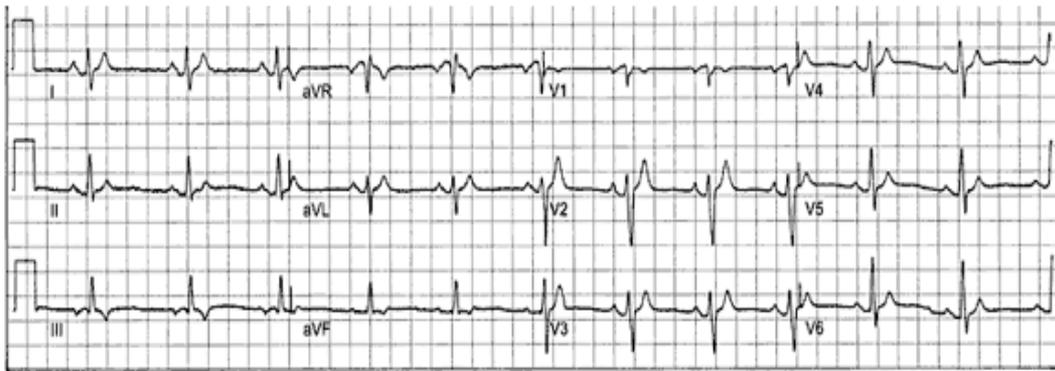
(QTc 315)



**SQTS 3**

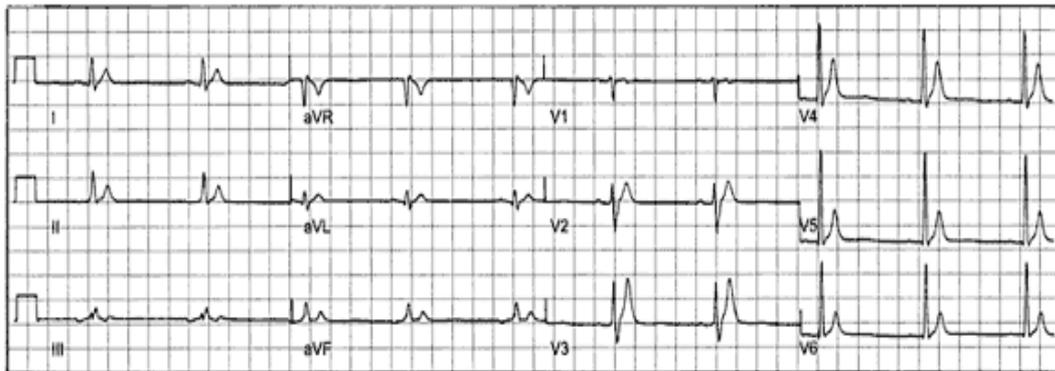


**SQTS 2**



a

**filles 17 ans, QTc 300 ms,  
syncope**



b

**frère 21 ans, QTc 267 ms,  
pas de symptôme**



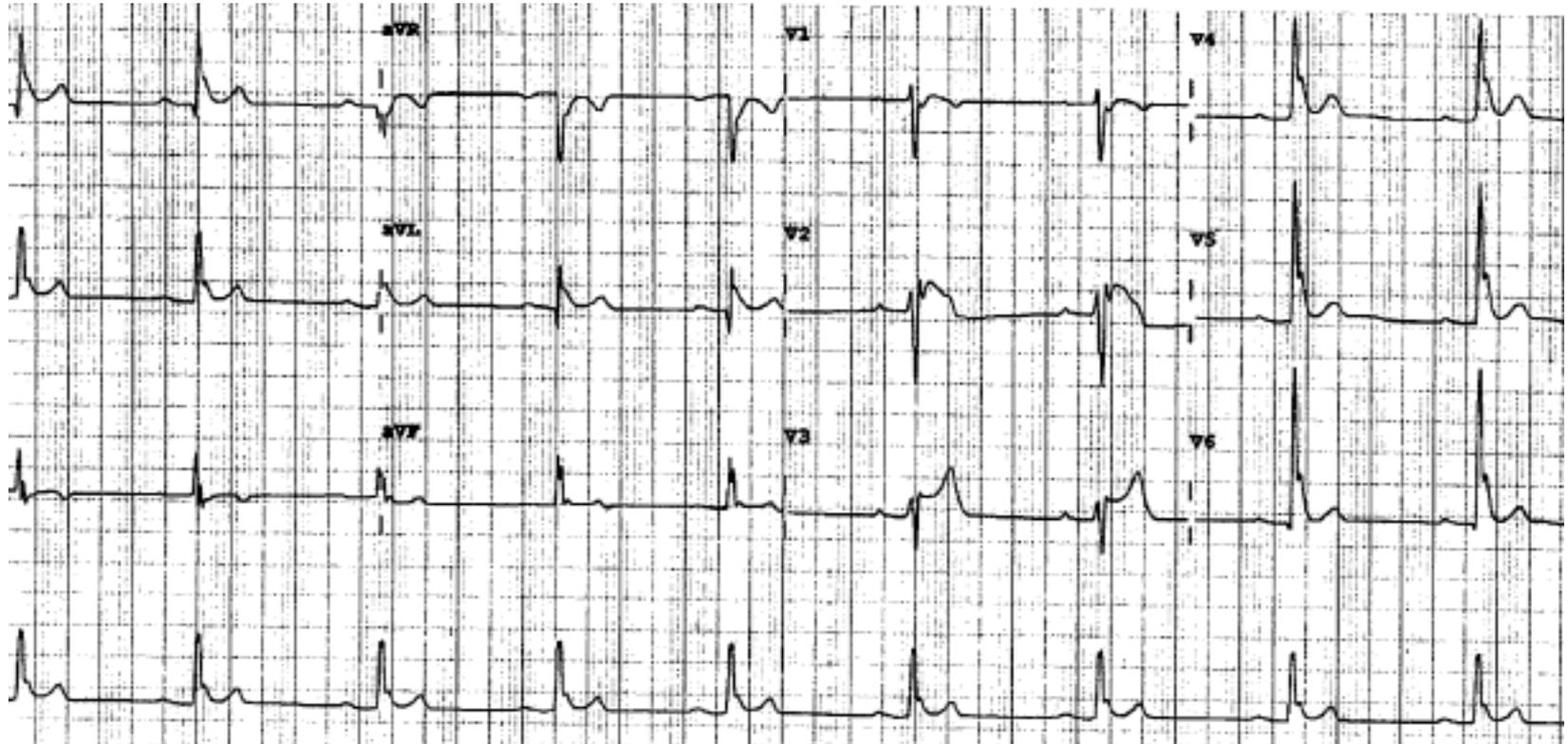
c

**mère 51 ans, QTc 289 ms,  
palpitations non documentées**

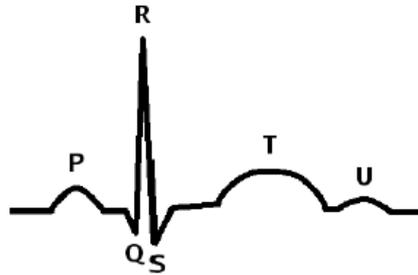
# Syndrome du QT court

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>	Ref. <sup>c</sup>
SQTS is diagnosed in the presence of a QTc $\leq$ 340 ms.	I	C	This panel of experts
SQTS should be considered in the presence of a QTc $\leq$ 360 ms and one or more of the following: <ul style="list-style-type: none"> <li>(a) A confirmed pathogenic mutation</li> <li>(b) A family history of SQTS</li> <li>(c) A family history of sudden death at age &lt; 40 years</li> <li>(d) Survival from a VT/VF episode in the absence of heart disease.</li> </ul>	IIa	C	This panel of experts

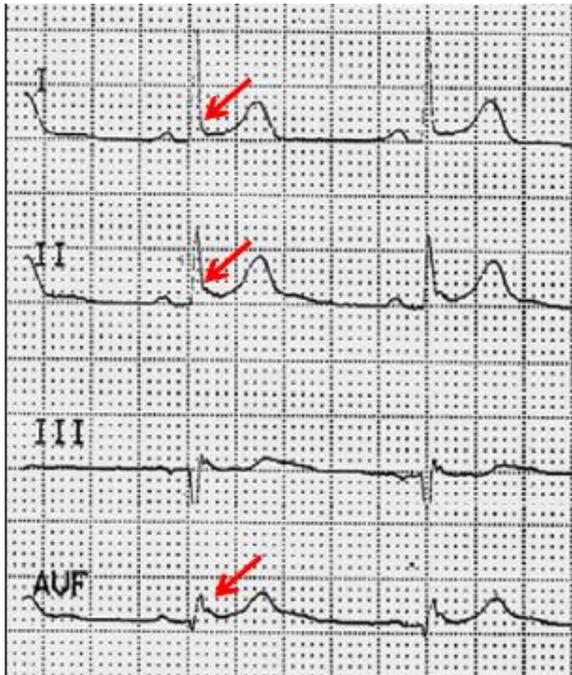
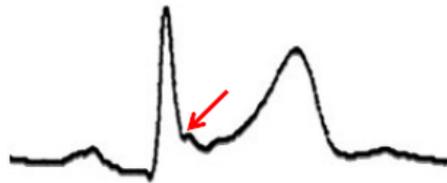
16 ans, MS récupérée



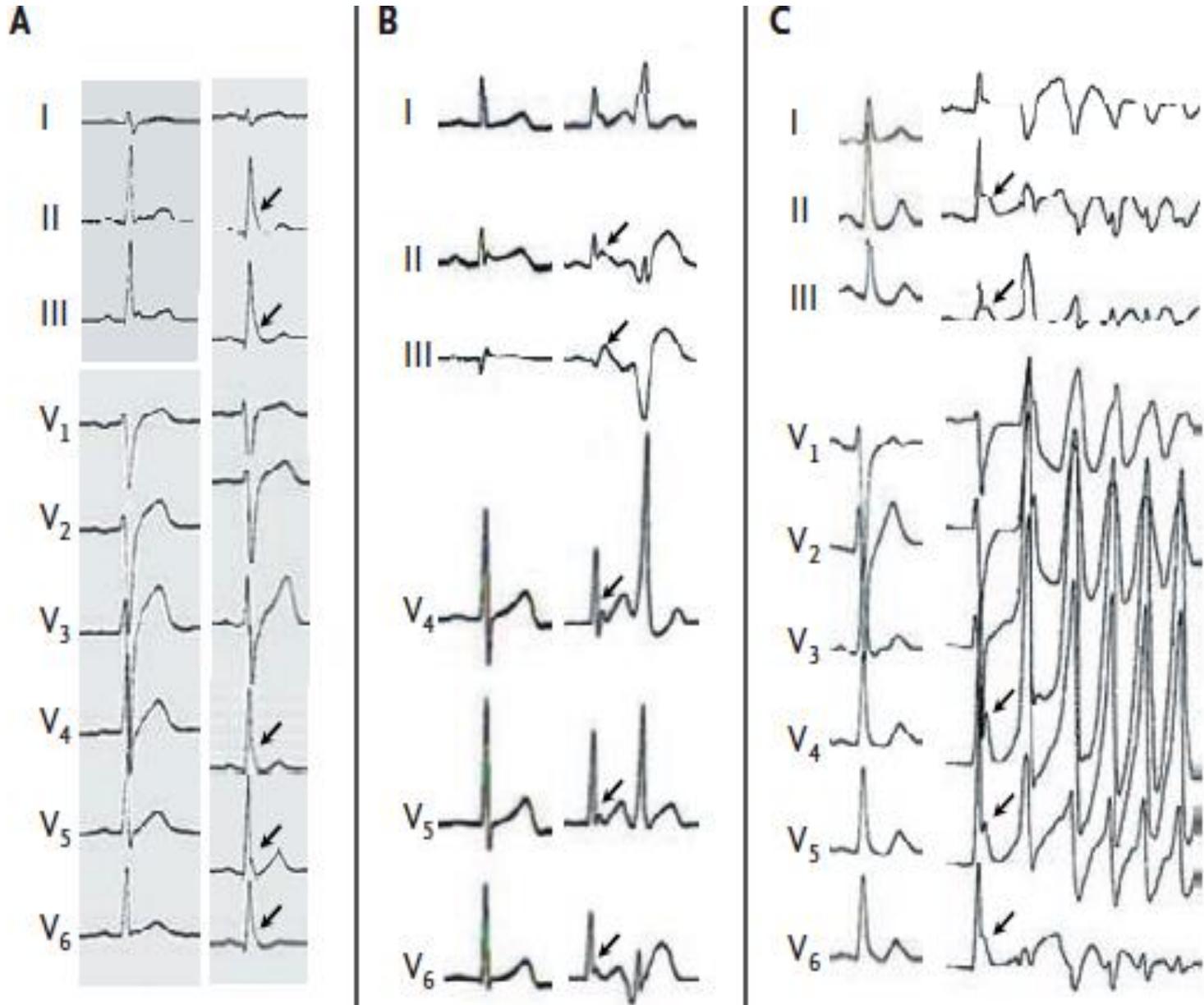
ECG normal



Repolarisation précoce

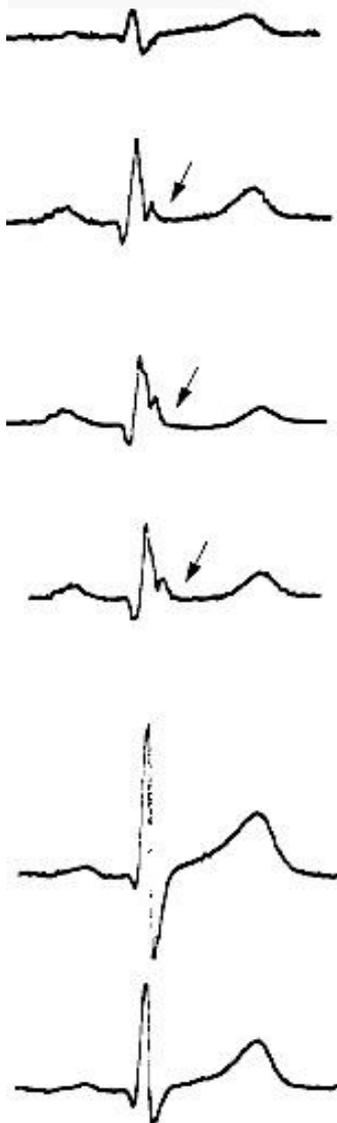


# Syndrome de repolarisation précoce

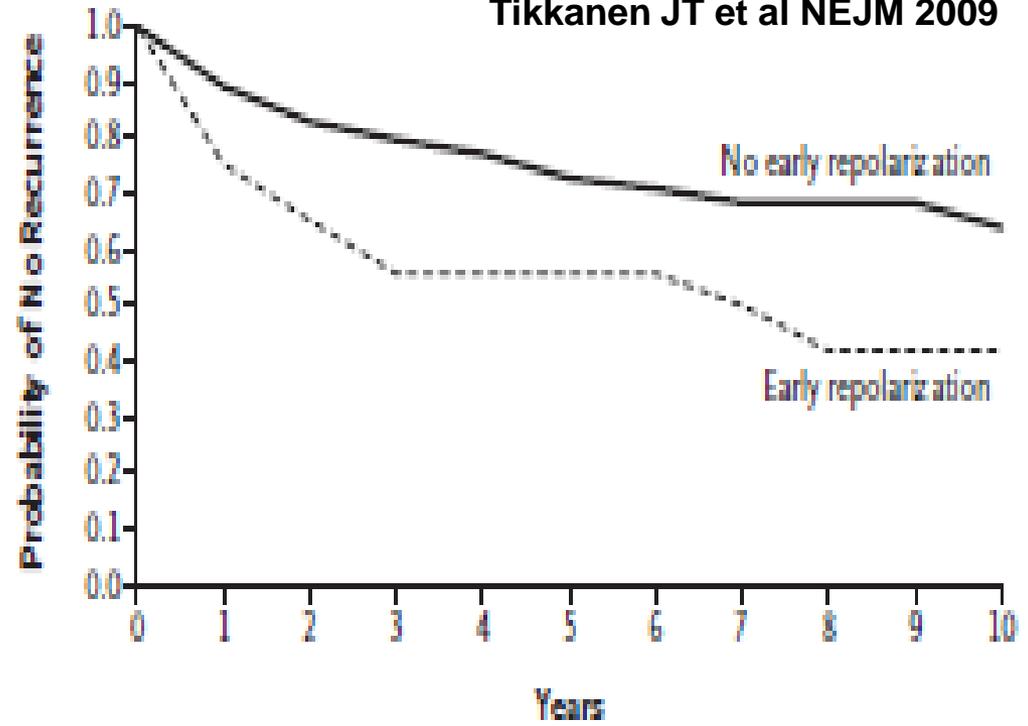
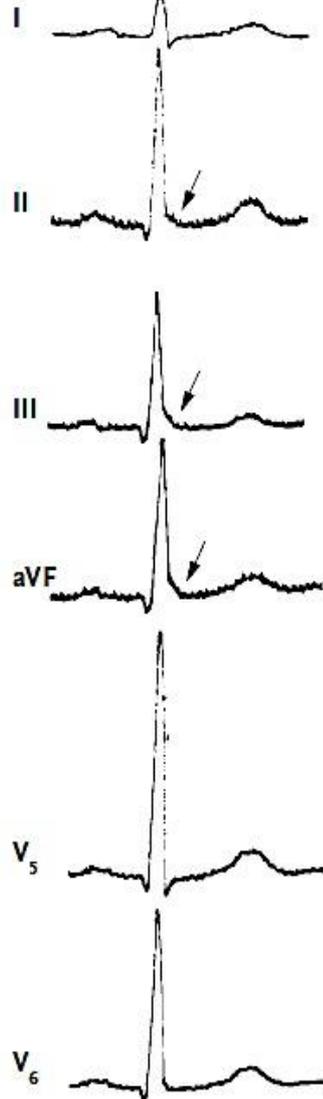


# Syndromes de repolarisation précoce (ondes J)

Notching



Slurring



TV idiopathique x 3.4 → 11/100 000 sujets

# Repolarisation précoce : diagnostic

## *Expert Consensus Recommendations on Early Repolarization Diagnosis*

1. ER ***syndrome is diagnosed*** in the presence of J-point elevation  $\geq 1$  mm in  $\geq 2$  contiguous inferior and/or lateral leads of a standard 12-lead ECG in a patient resuscitated from otherwise unexplained VF/ Polymorphic VT
2. ER syndrome ***can be diagnosed*** in a SCD victim with a negative autopsy and medical chart review with a previous ECG demonstrating J-point elevation  $\geq 1$  mm in  $\geq 2$  contiguous inferior and/or lateral leads of a standard 12-lead ECG
3. ER pattern ***can be diagnosed*** in the presence of J-point elevation  $\geq 1$  mm in  $\geq 2$  contiguous inferior and/or lateral leads of a standard 12-lead ECG

**Légère accentuation : bradycardie, bétabloquants, Valsalva**

**Diminution : effort / isoproterenol**

*HRS/EHRA/APHRs Expert Consensus Statement on the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes 2013*

# ST : ascendant/descendant

**ST ascendant**

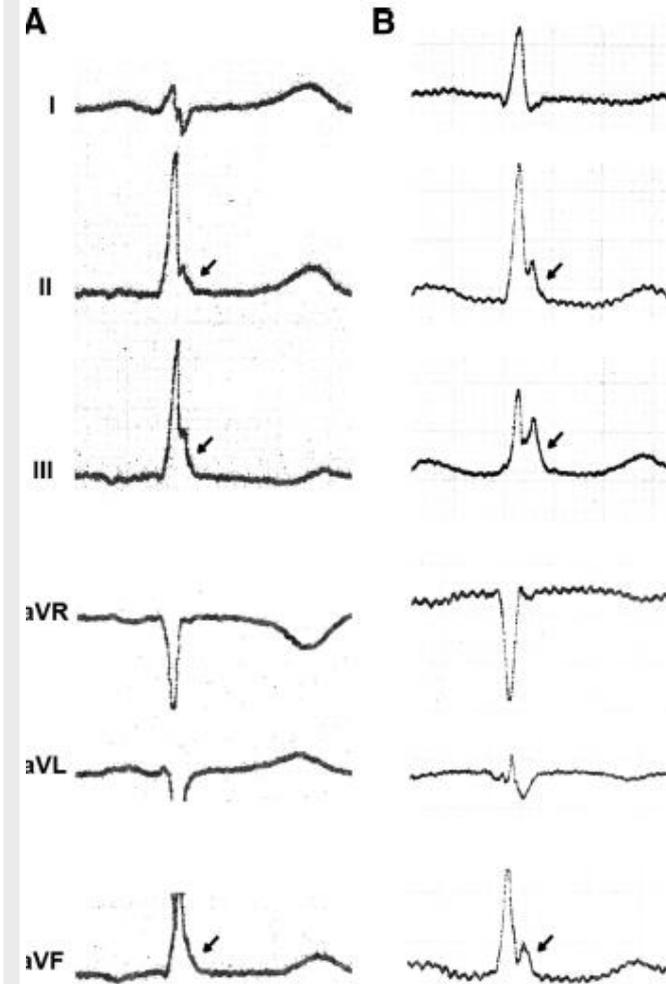
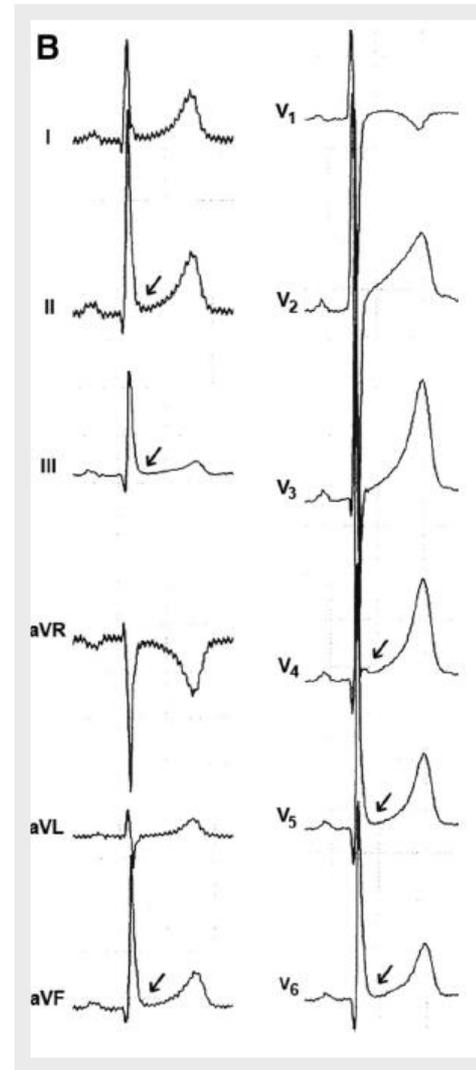
**→ bon pronostic**

**ST descendant**

**→ mauvais pronostic**

**Mortalité CV :**

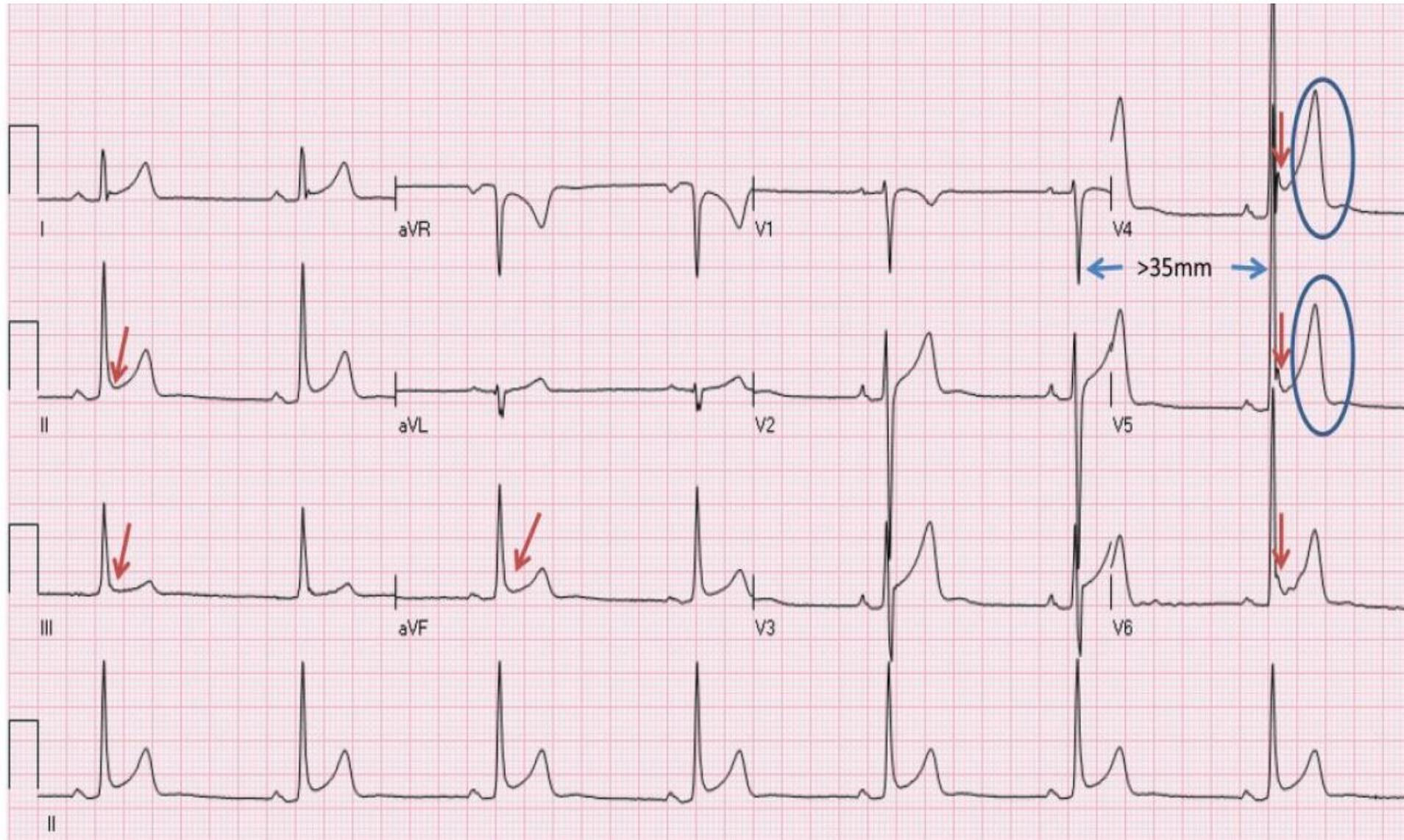
**HR: 8.75 (CI 3.48-22.0, p<0.0001)**



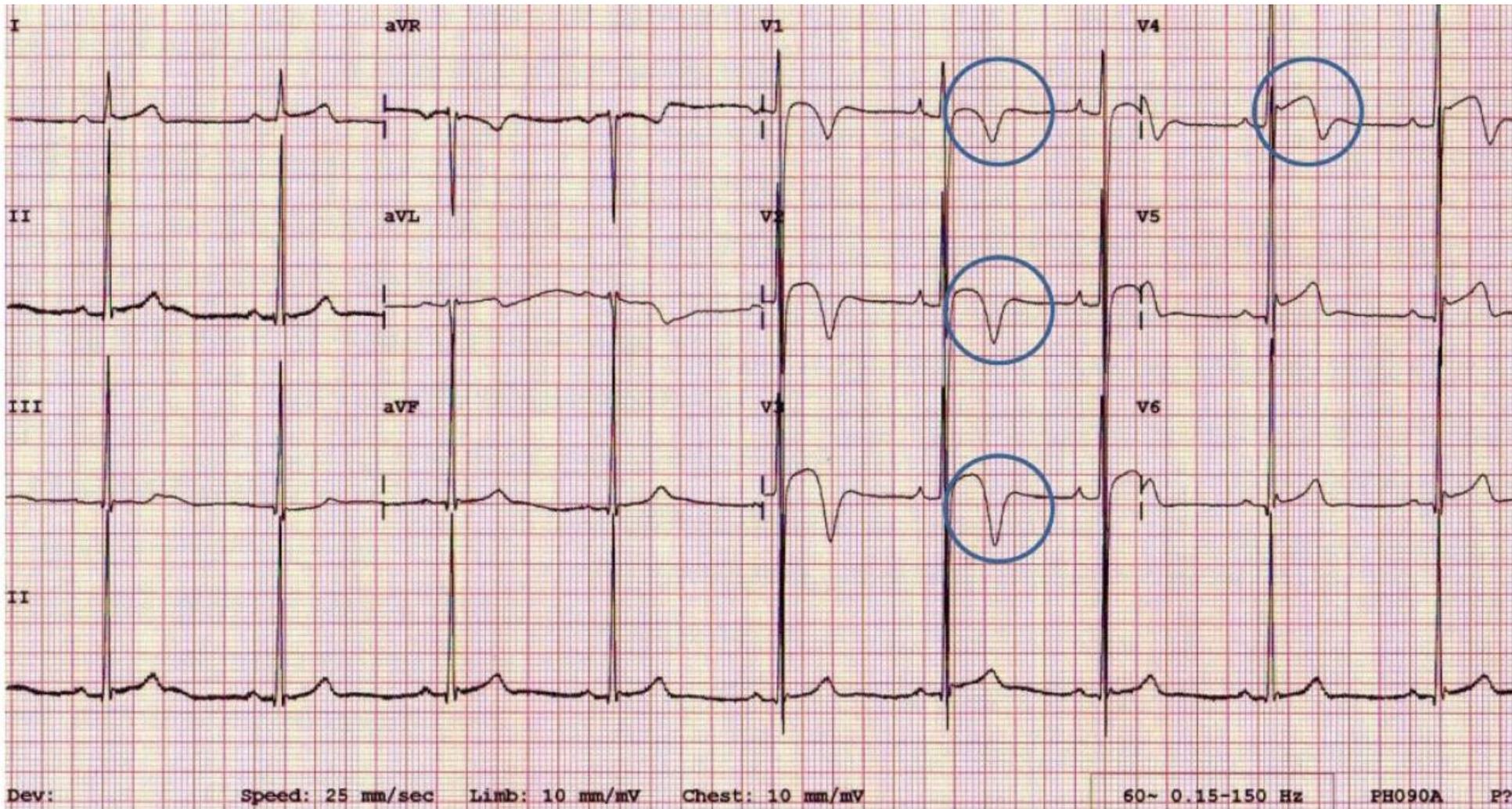
*Tikkanen J et al. Circulation 2011*

*Rosso R, Glikson E, Belhassen B, Katz A, Halkin A, Steinvil A, Viskin S. Heart Rhythm 2011*

# Sportif de 15 ans



# ECG athlete noir



# Conclusions

- Durée + morphologie anormales : suspect
- Contexte personnel et familial
- Tests de sensibilisation
  - Holter, EE, tests pharmacologiques