

Anomalies ECGS de la repolarisation ventriculaire

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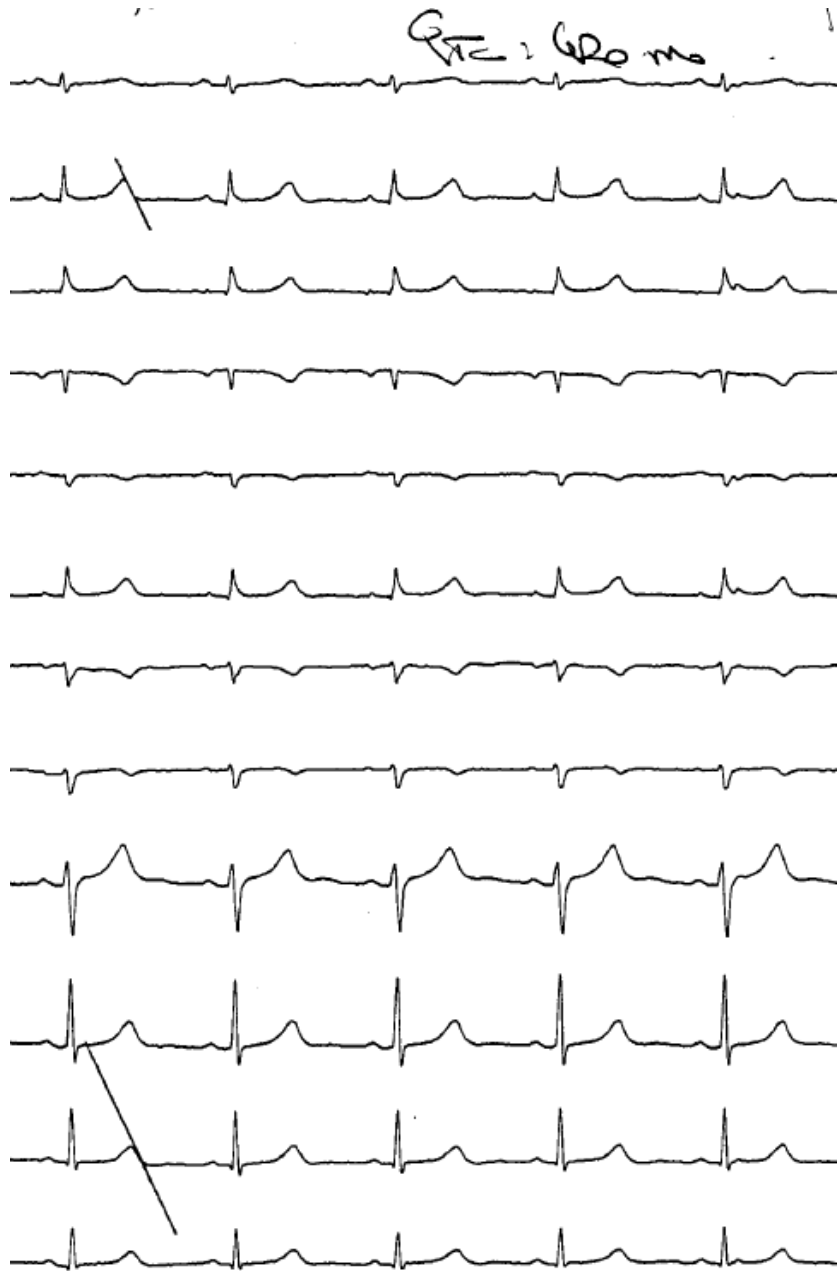
Centre de Référence

Maladies Cardiaques Héréditaires

Filière Cardiogen

Séminaire NEM 22 03 2018





Garçon de 15 ans

Syncope

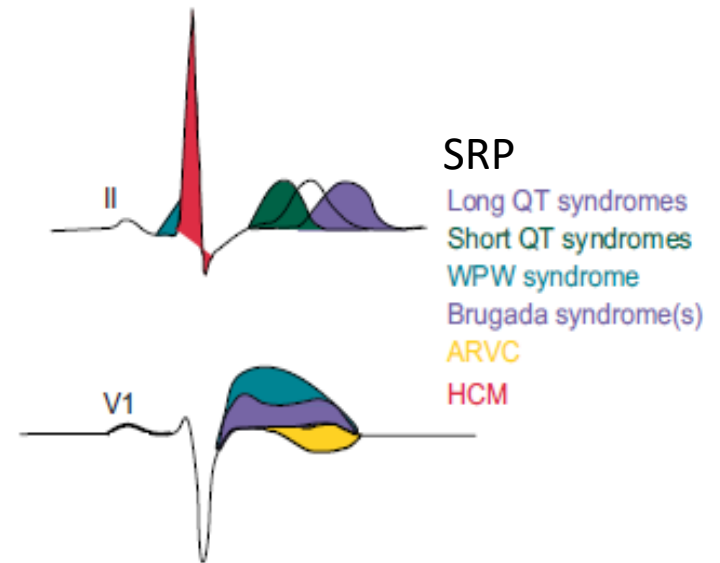
Circonstances ?

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>
Lead V2:		
<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)
Lead V5:		
<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)
Leads I, II, and III:		
<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.

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

Sami Viskin. Heart Rhythm 2009.

Mesure de l'intervalle QT

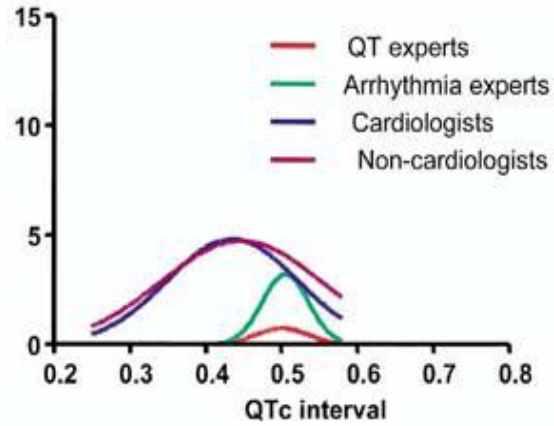


Mesure de l'intervalle QT corrigé (formule Bazett)

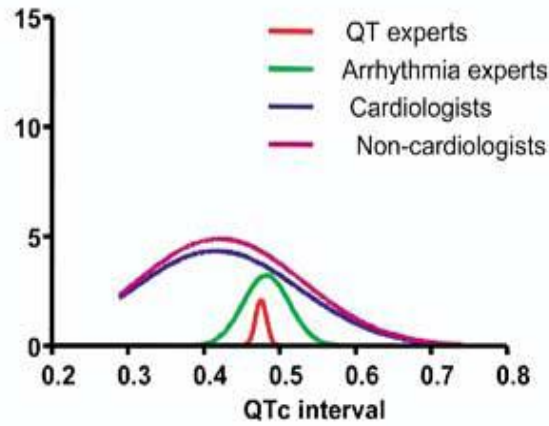
$$QTc = \frac{(QT1 / \sqrt{RR1}) + (QT2 / \sqrt{RR2}) + (QT3 / \sqrt{RR3})}{3}$$

QTc Measurement

Trace 1: Distribution of QTc values

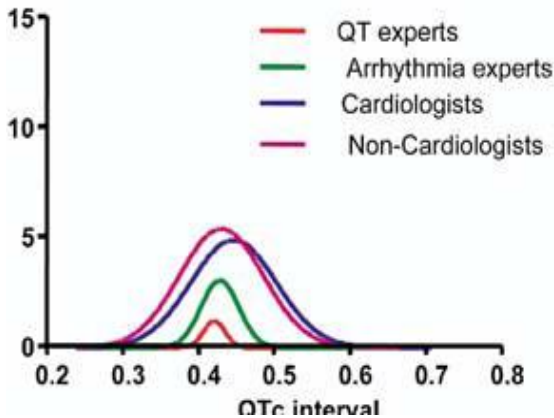


Trace 2: Distribution of QTc values

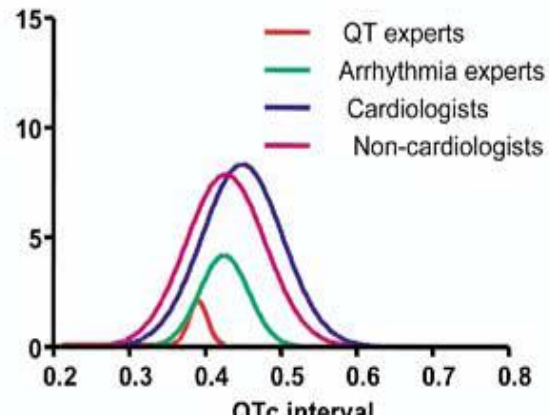


Long QT syndrome :
QTc under estimated

Trace 3: Distribution of QTc values



Trace 4: Distribution of QTc values



Normal QT :
QTc over estimated

Causes de QT allongé

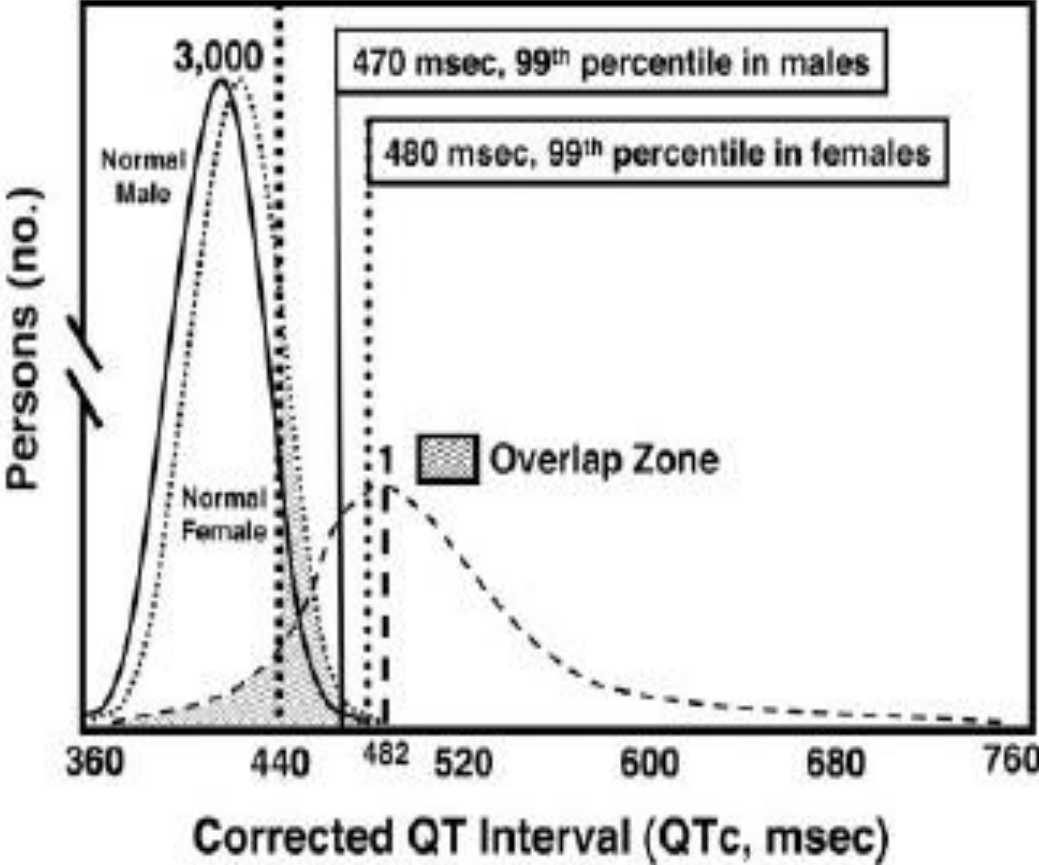
- **Facteurs de risques non médicamenteux** :
 - sexe féminin, age avancé
 - troubles électrolytiques (hypokaliémie et hypomagnésémie)
 - insuffisance cardiaque
 - bradycardie
 - ischémie
 - **syndrome congénital du QT long**

- **Causes médicamenteuses** :

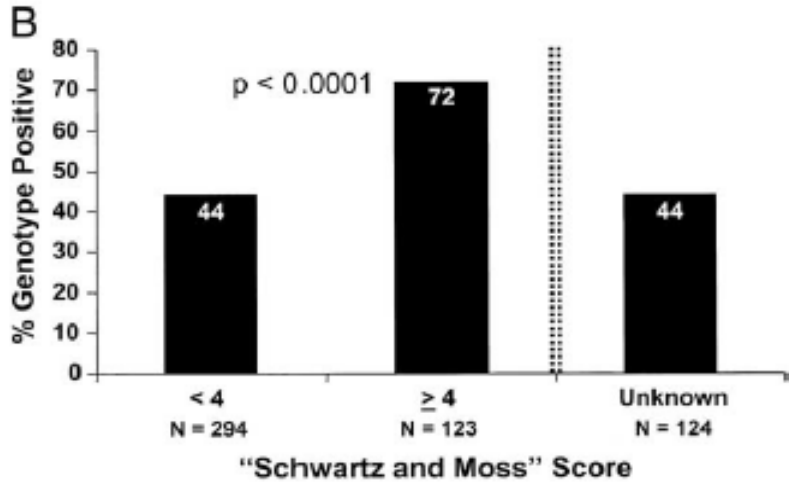
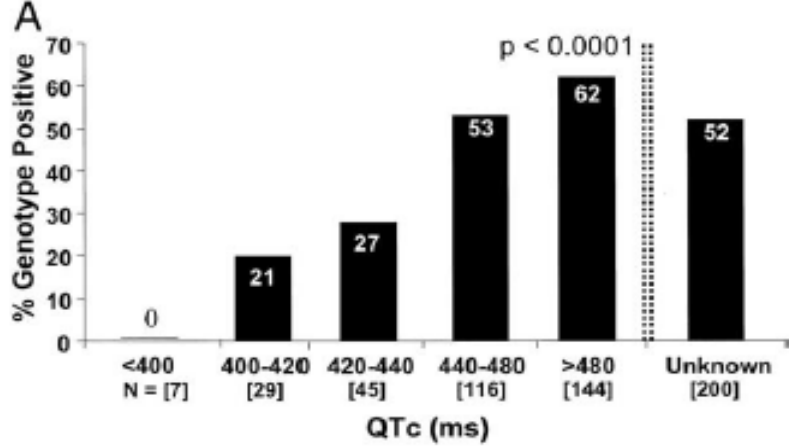
Agents anti-arythmiques : classe IA (quinidine, disopyramide, procaïnamide), classe II (sotalol et dans une moindre mesure amiodarone)

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

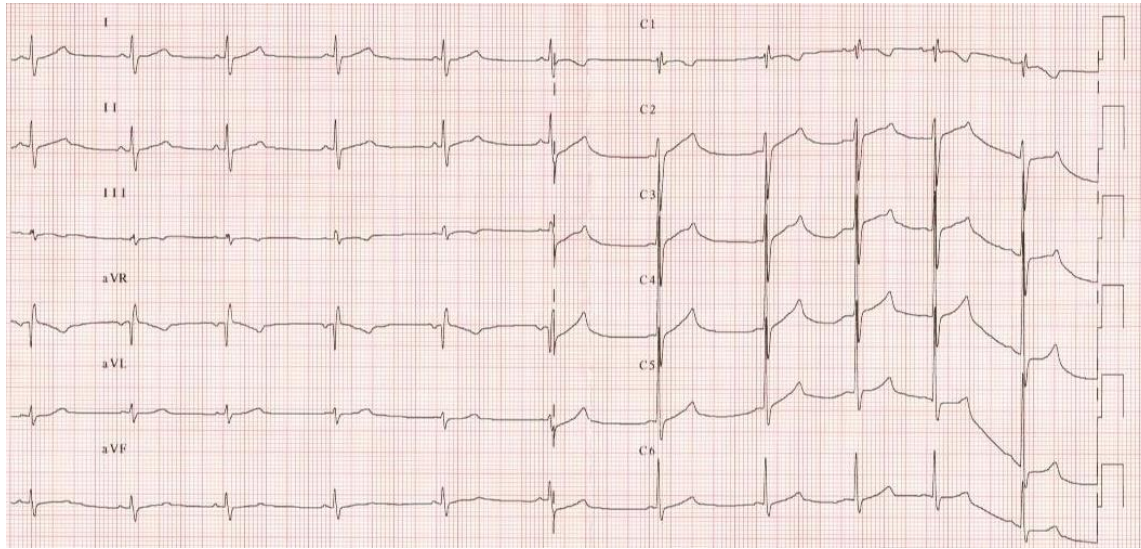
Limitation of QTc



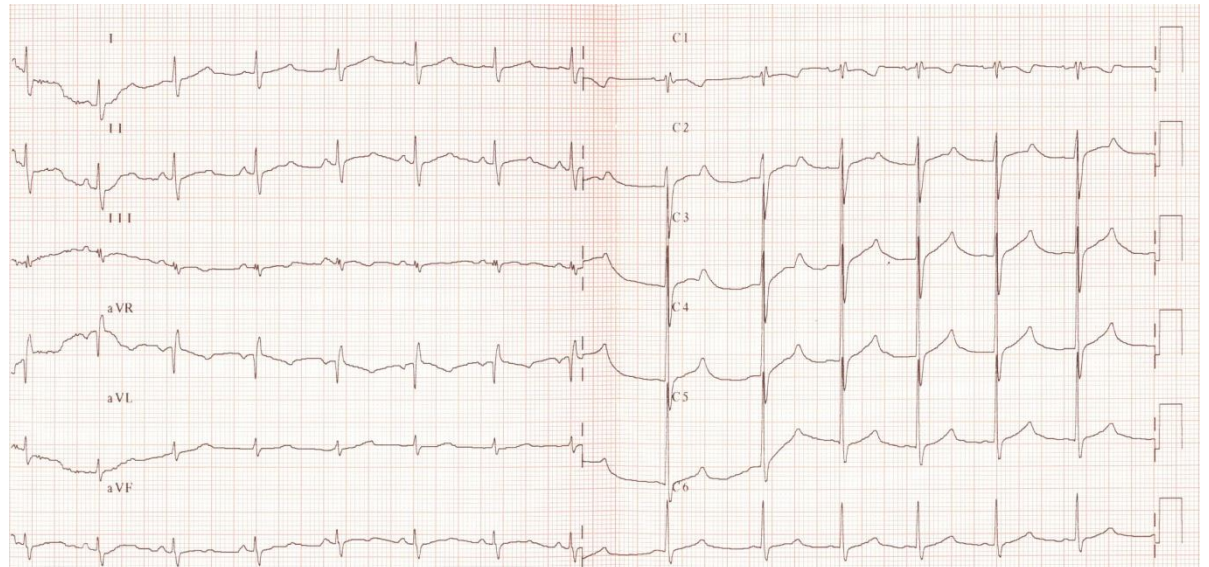
Taggart et al. Circulation
2007;115



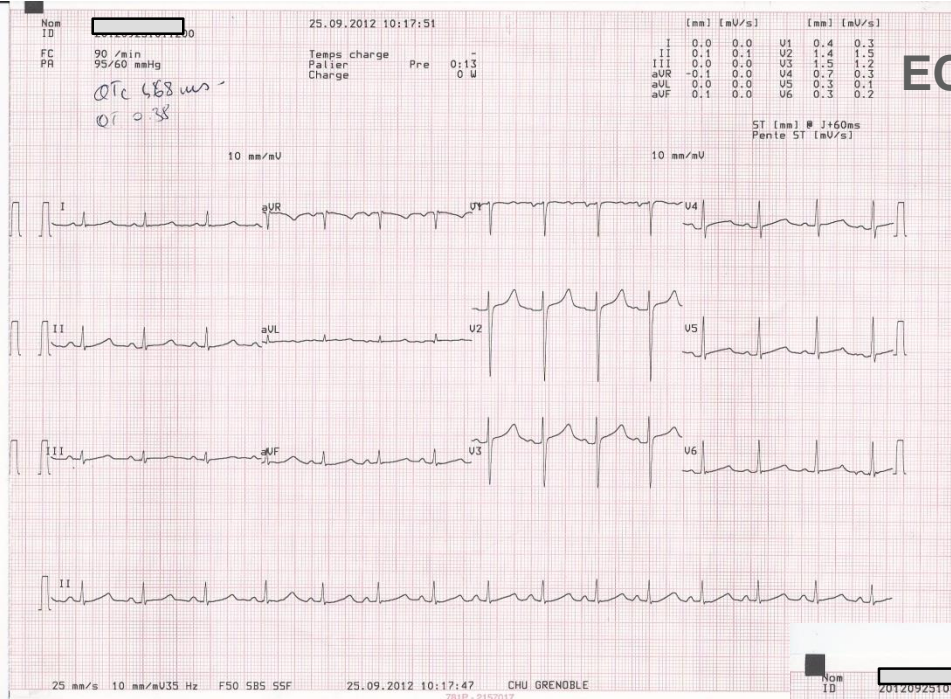
Tester et al.
JACC 2006;47



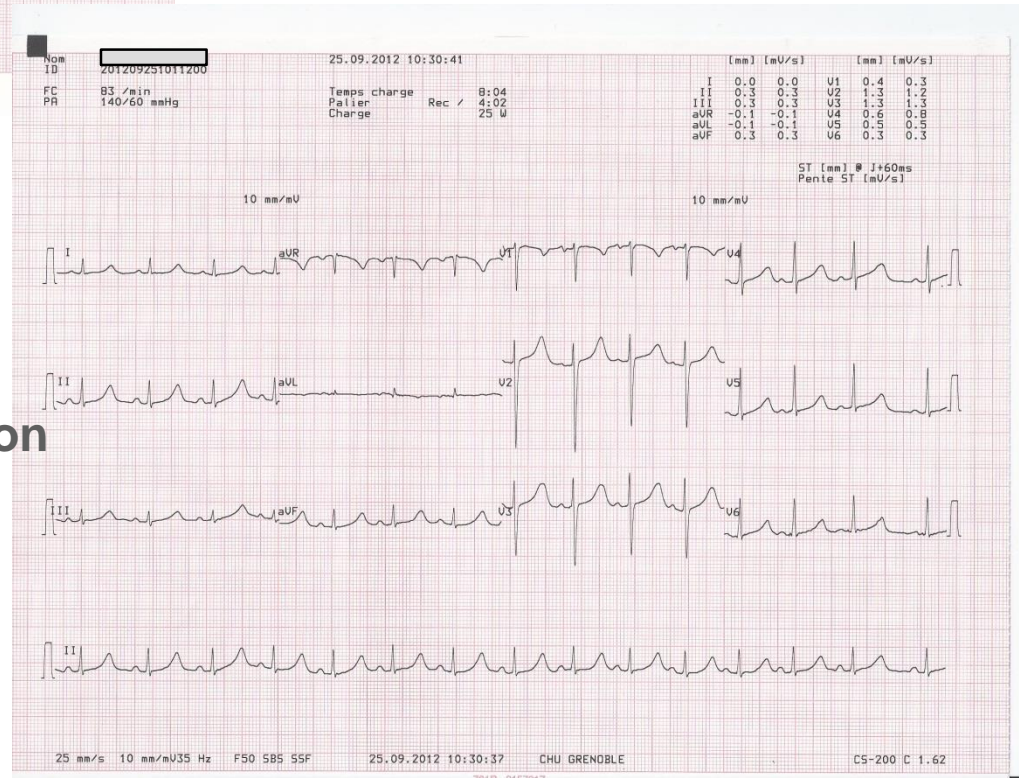
ECG allongé : QTc 440ms



ECG debout : QTc 470ms



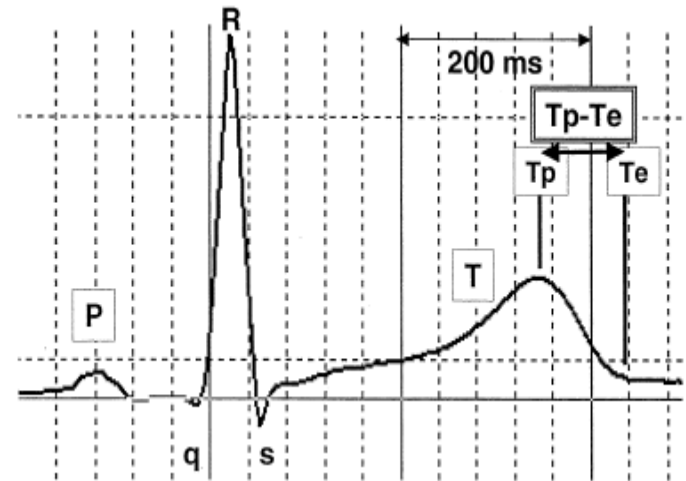
ECG de base



4^{ème} 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

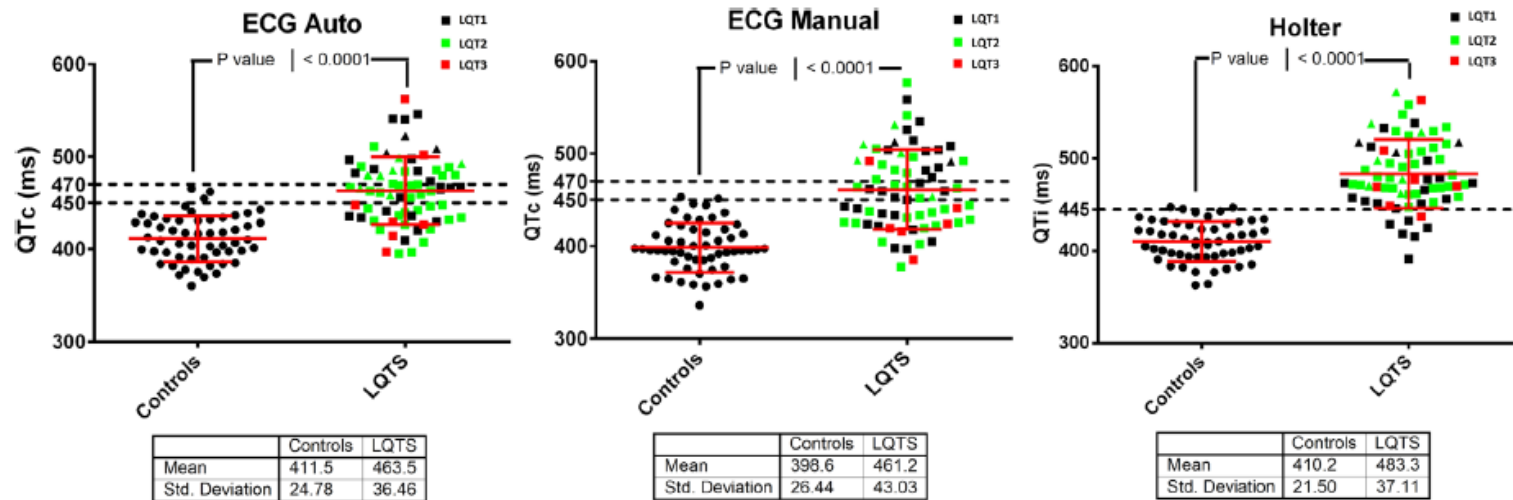
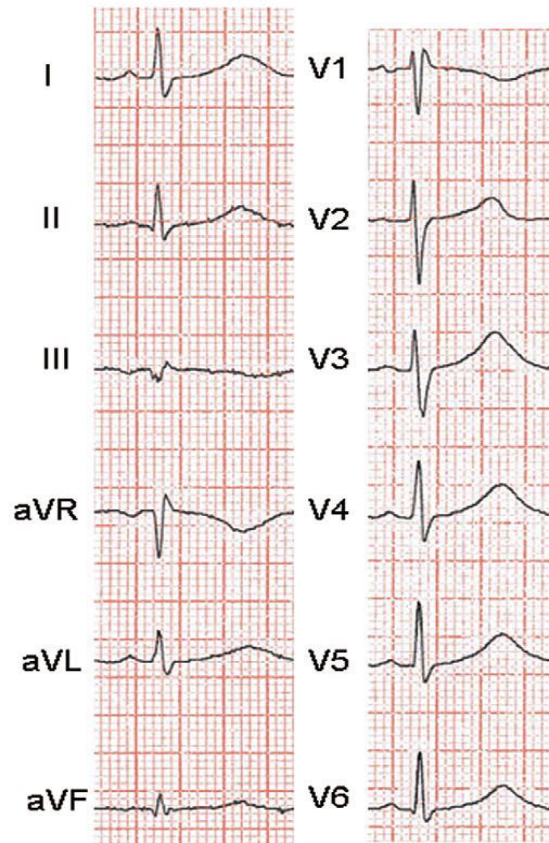


Figure 2 Different QTc measurement techniques in controls and LQTS mutation carriers. Automated (left panel) and manual (middle panel) QT interval measurements with correction for heart rate using the Bazett formula and an individualized QT correction using a linear QT-RR regression obtained from 24-hour 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 QTt. LQTS = long QT syndrome; QTc = rate corrected QT interval using the Bazett formula; QTt = individualized corrected QT interval.

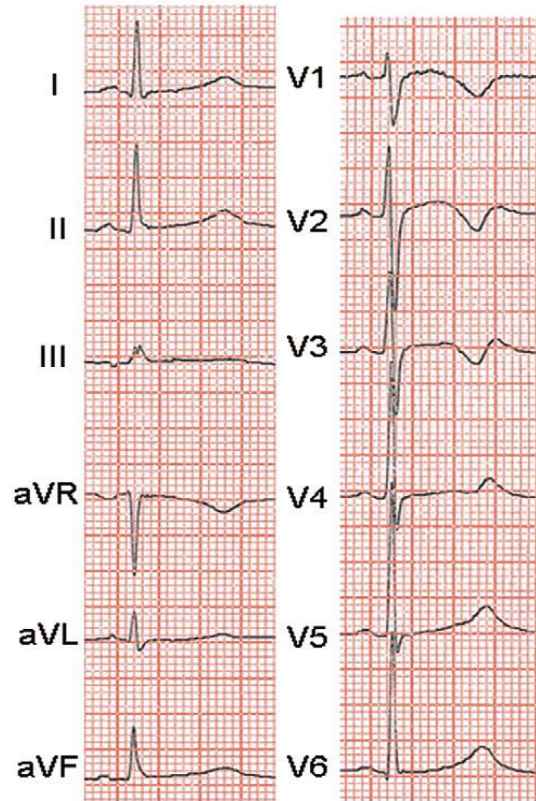
Morphologies anormales onde T

A



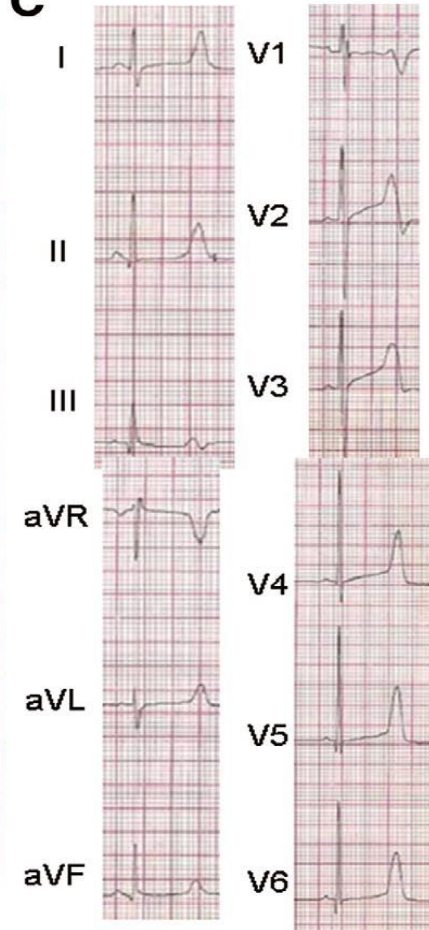
LQT1

B



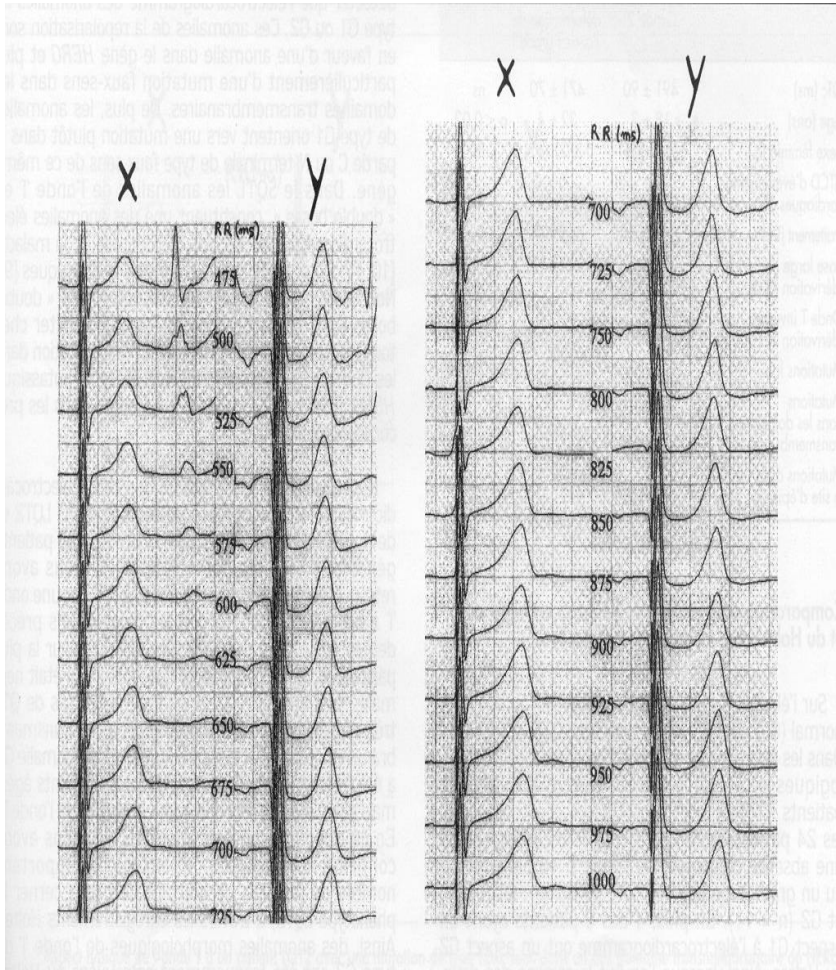
LQT2

C

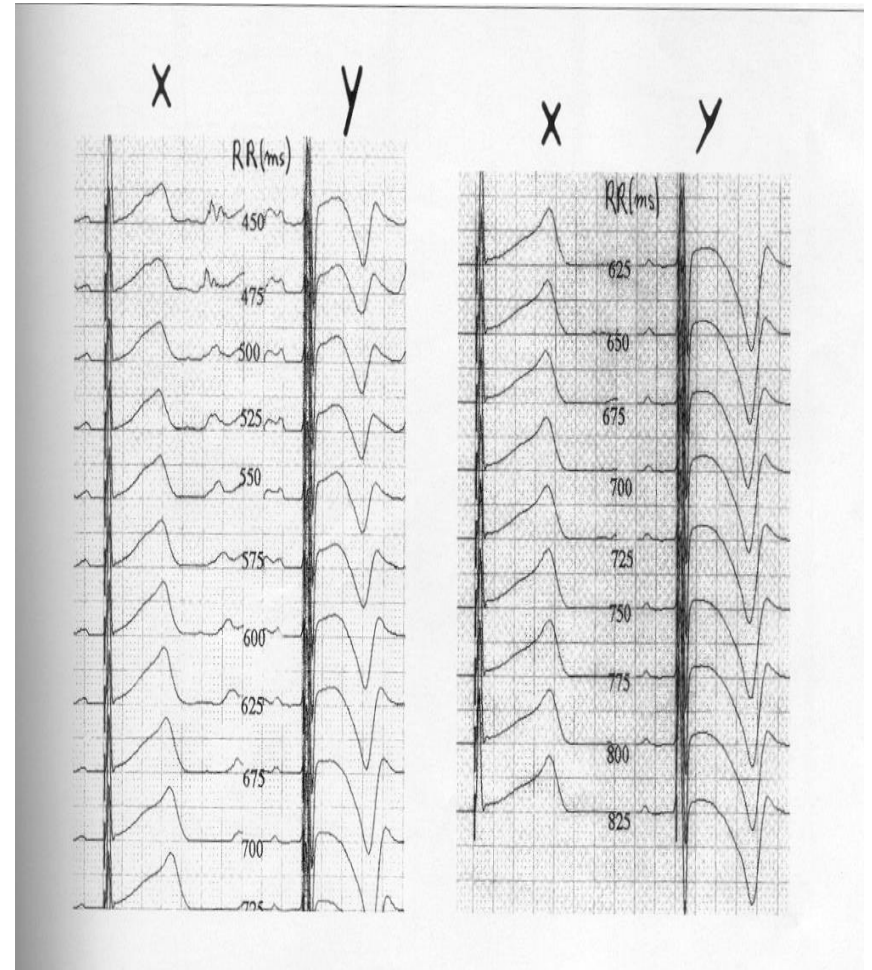


LQT3

Morphologie de l'onde T : Holter

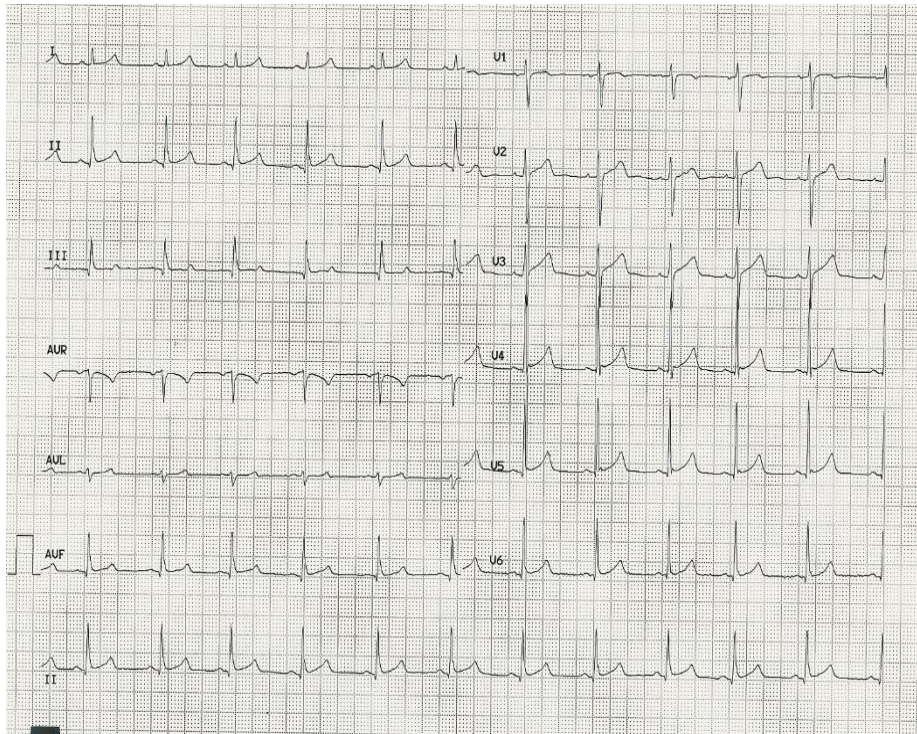


Forme LQT1

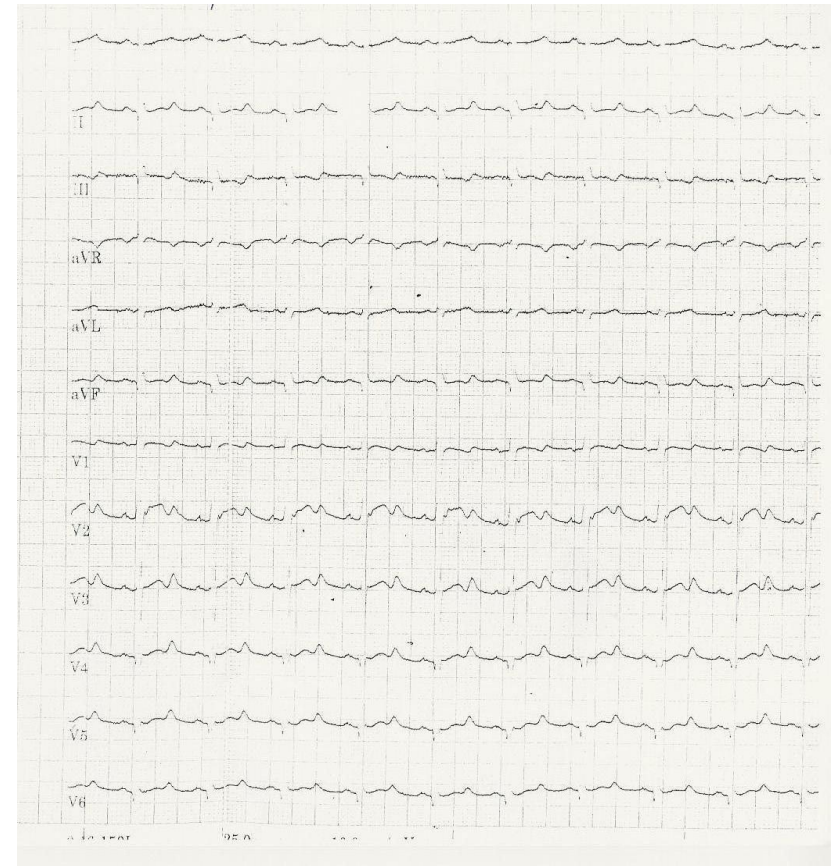


Forme LQT2

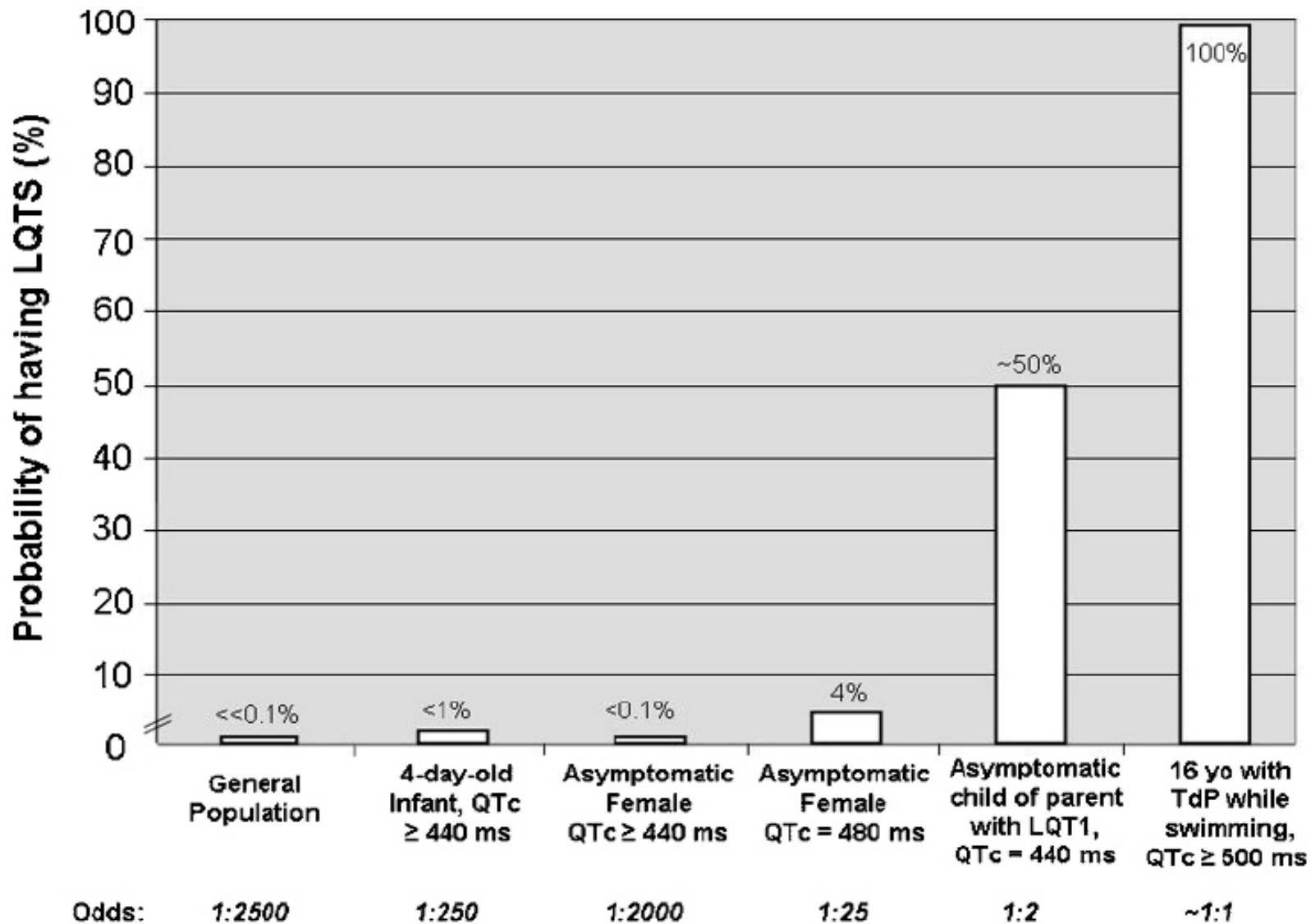
Jeune fille de 12 ans : syncope brève



Ecg de repos : QTc 450 ms

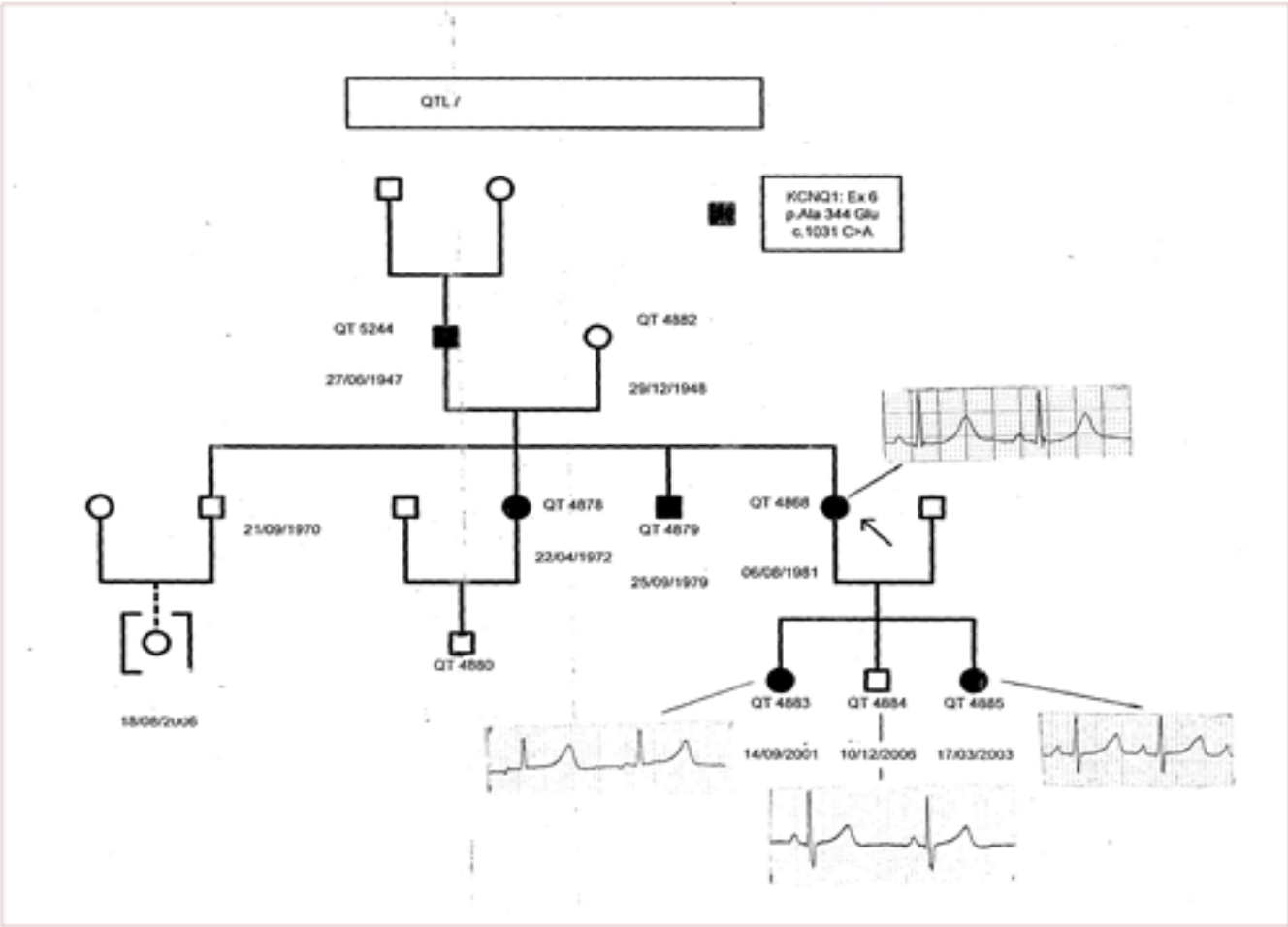


Test adrenaline : QTc 530 ms



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

Etude familiale



Diagnostic

- 1. a: score ≥ 3.5**
- 1. c: QTc ≥ 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 ≥ 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 ≥ 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.

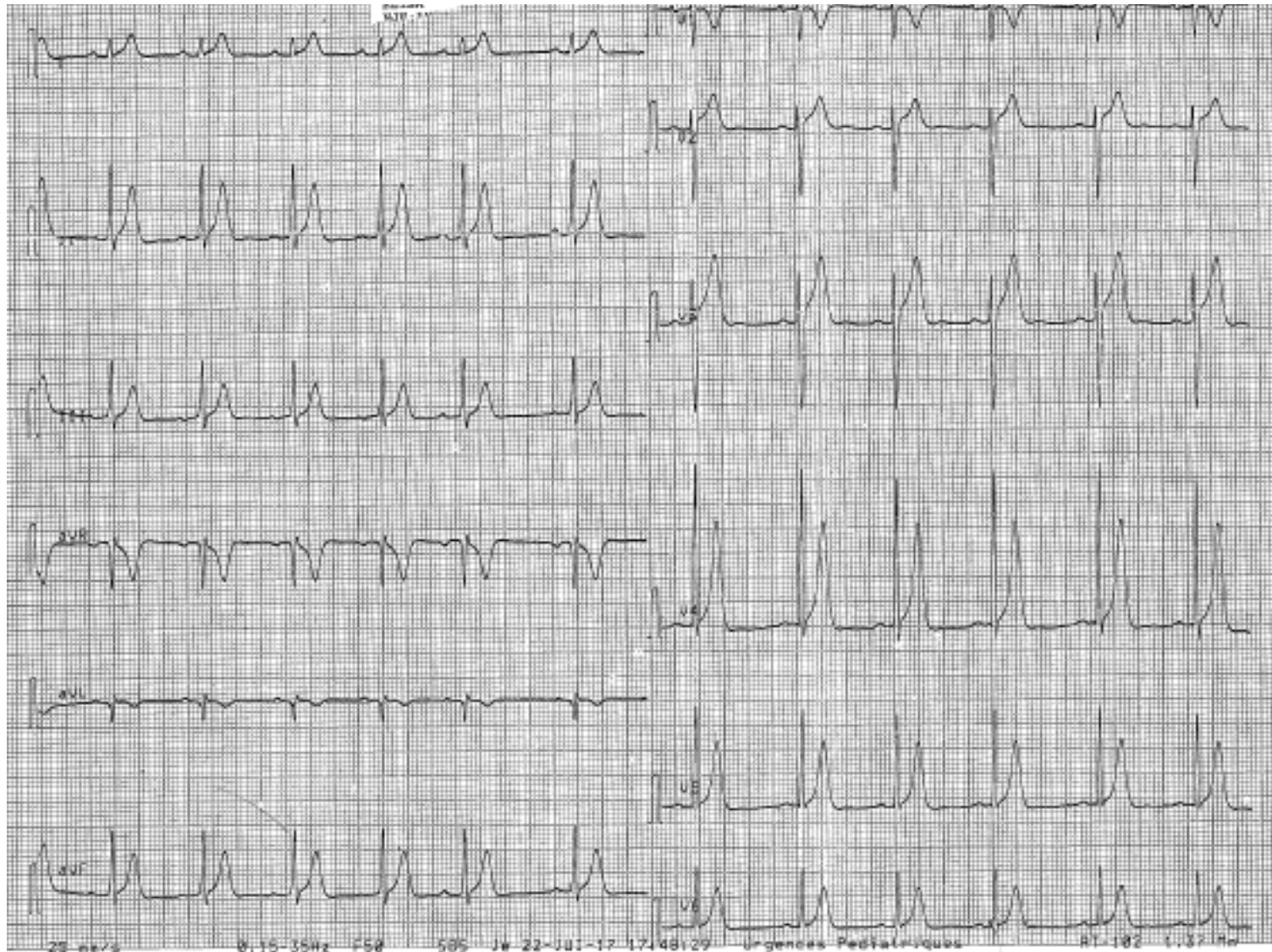
2 . QTc entre 480 et 499 ms

Fille de 4 ans, ECG pour bradycardie



QTc = 320 ms; morphologie anormale T

Fille de 10 ans, ECG pour le sport



QTc = 330 ms

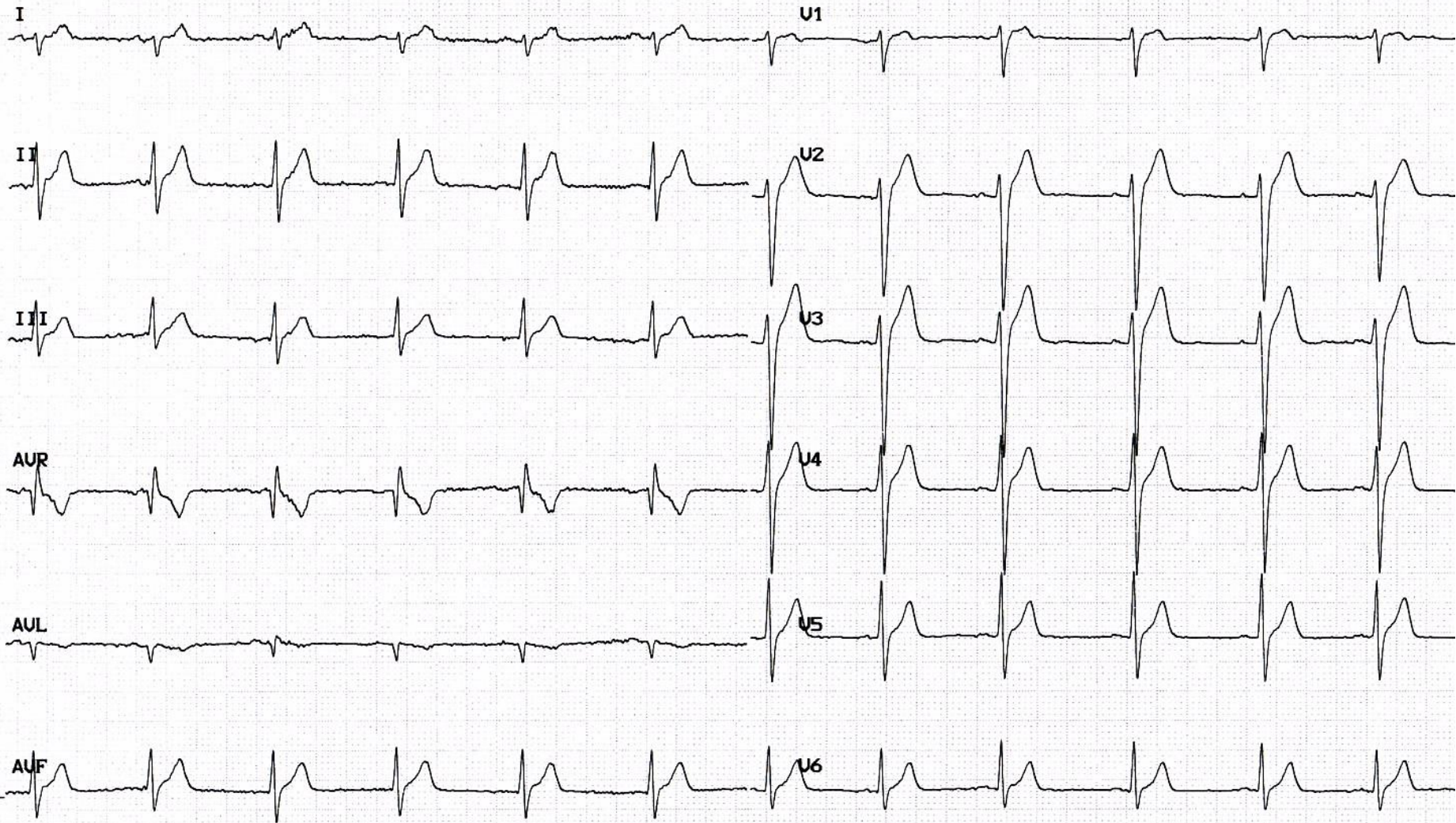
Causes de QT court

- 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

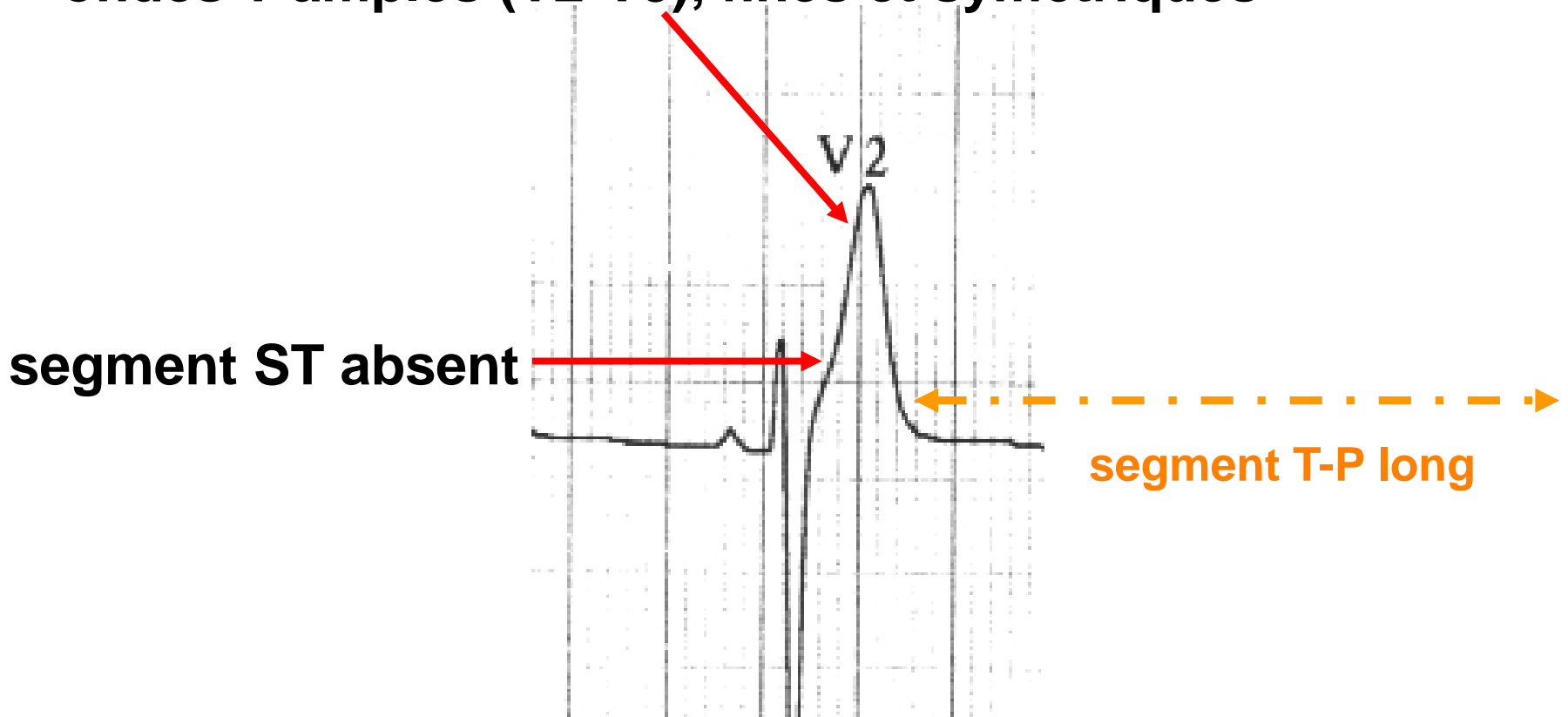


QT 290 ms, QTc 320 ms

QT = 76 % QT théorique

Mesure QT court

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



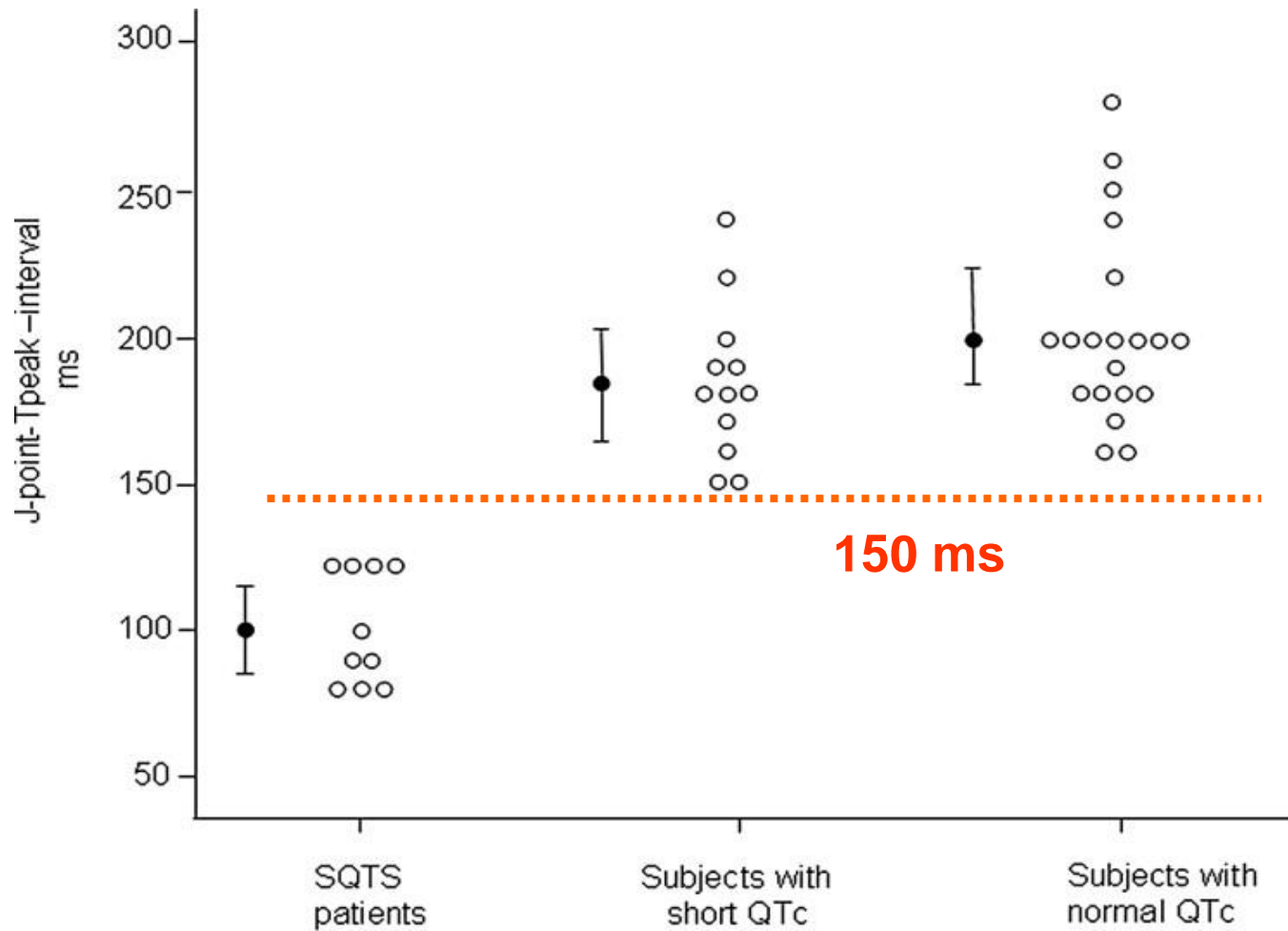
Durée et morphologie

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

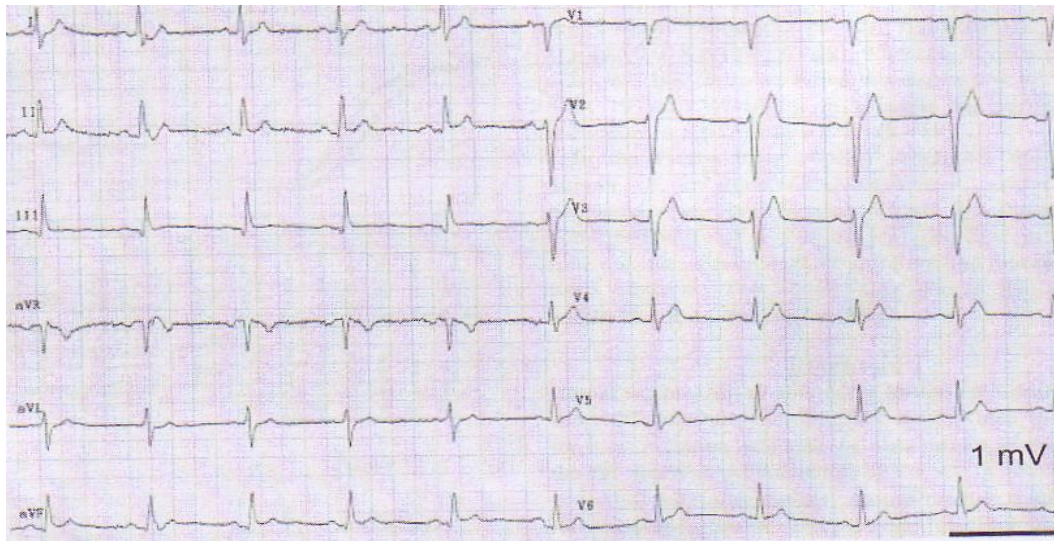
(QTc 320)



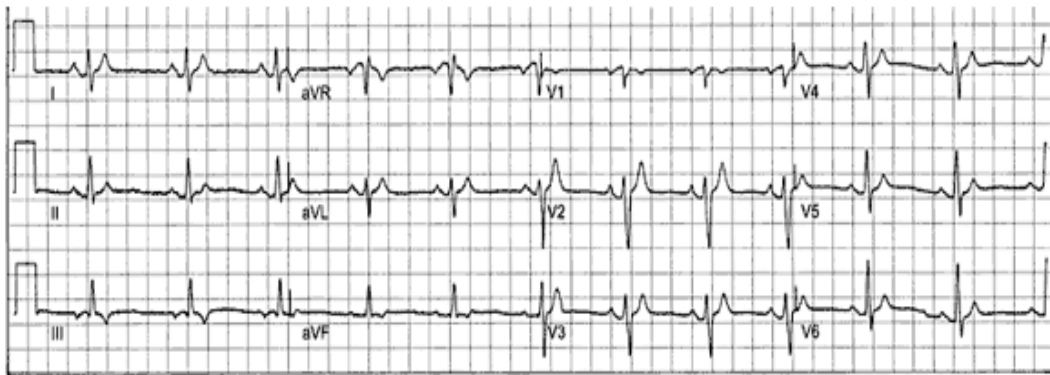
(QTc 315)



SQTS 3

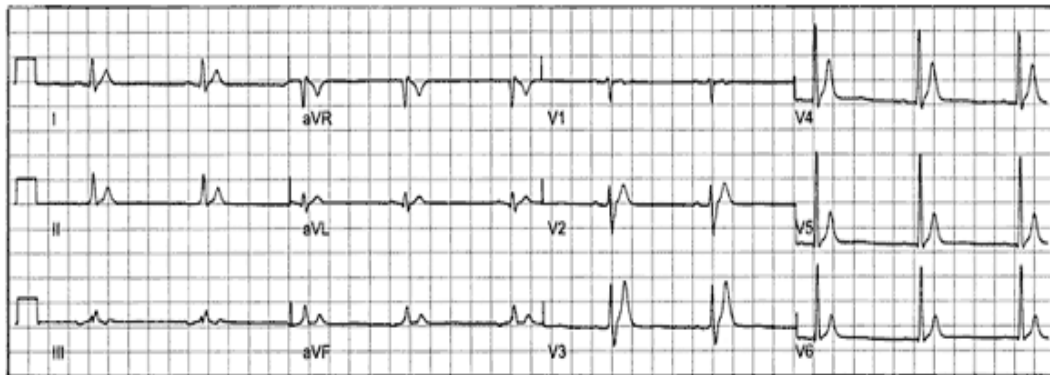


SQTS 2



a

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



b

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



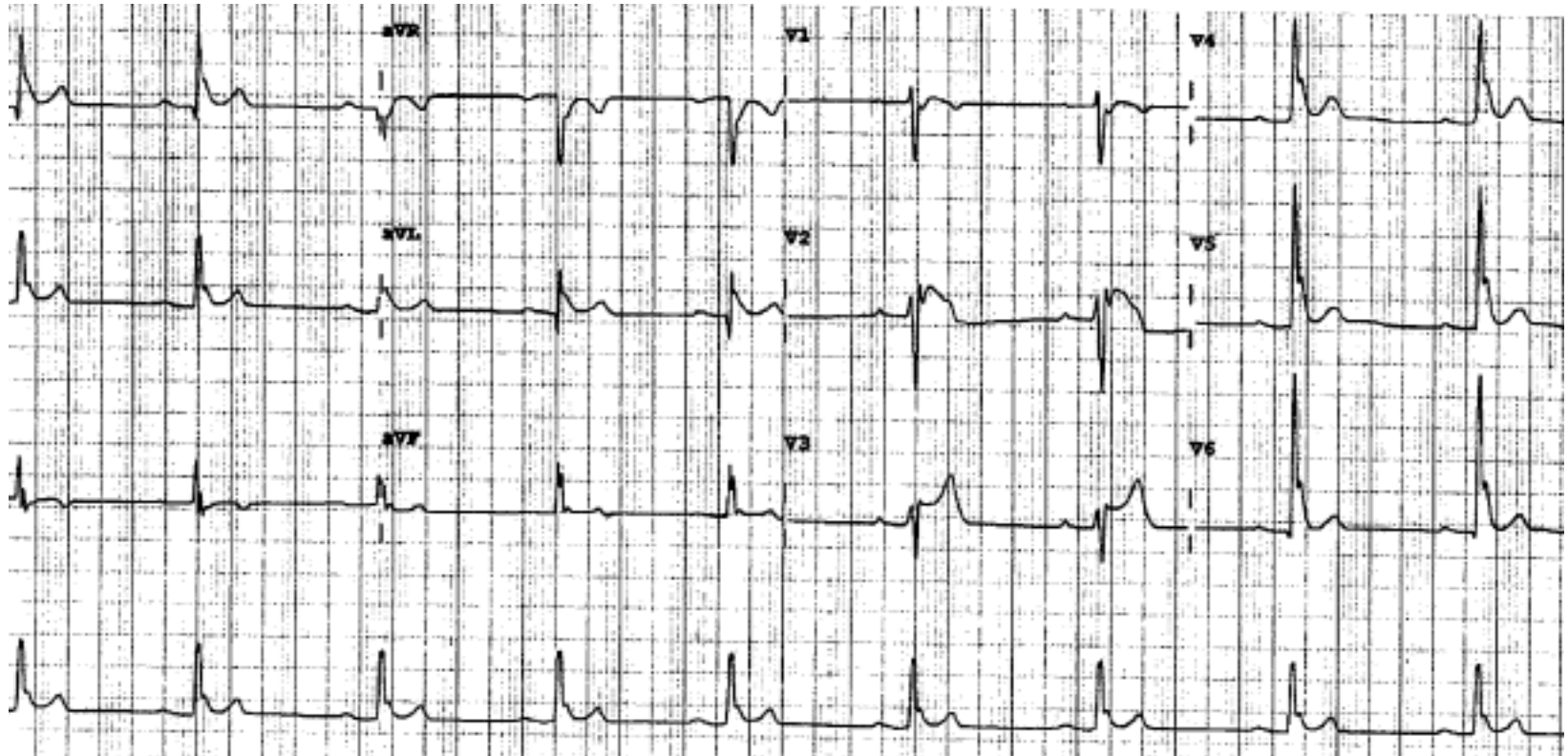
c

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

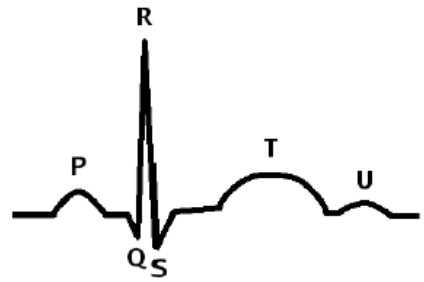
Syndrome du QT court

Recommendations	Class ^a	Level ^b	Ref. ^c
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"> (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	This panel of experts

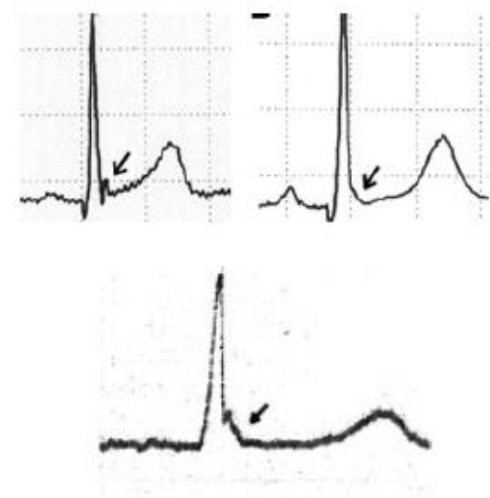
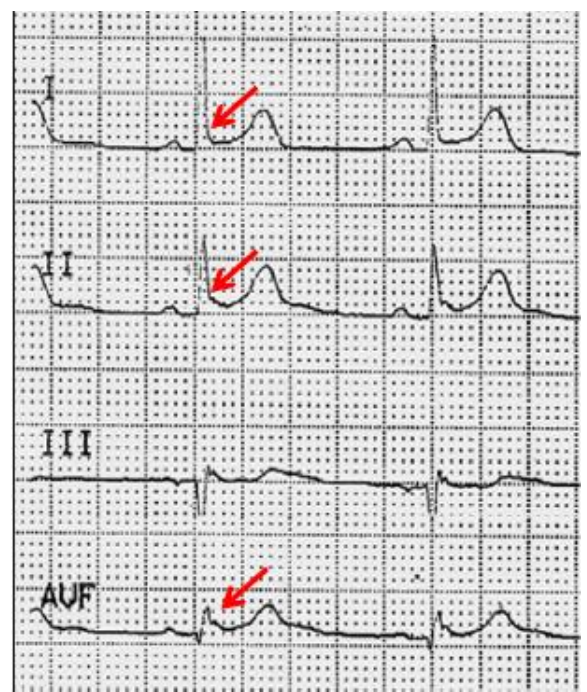
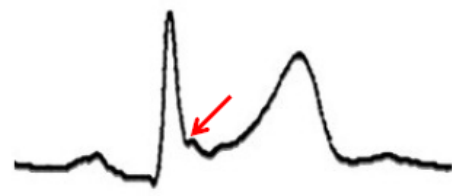
16 ans, MS récupérée



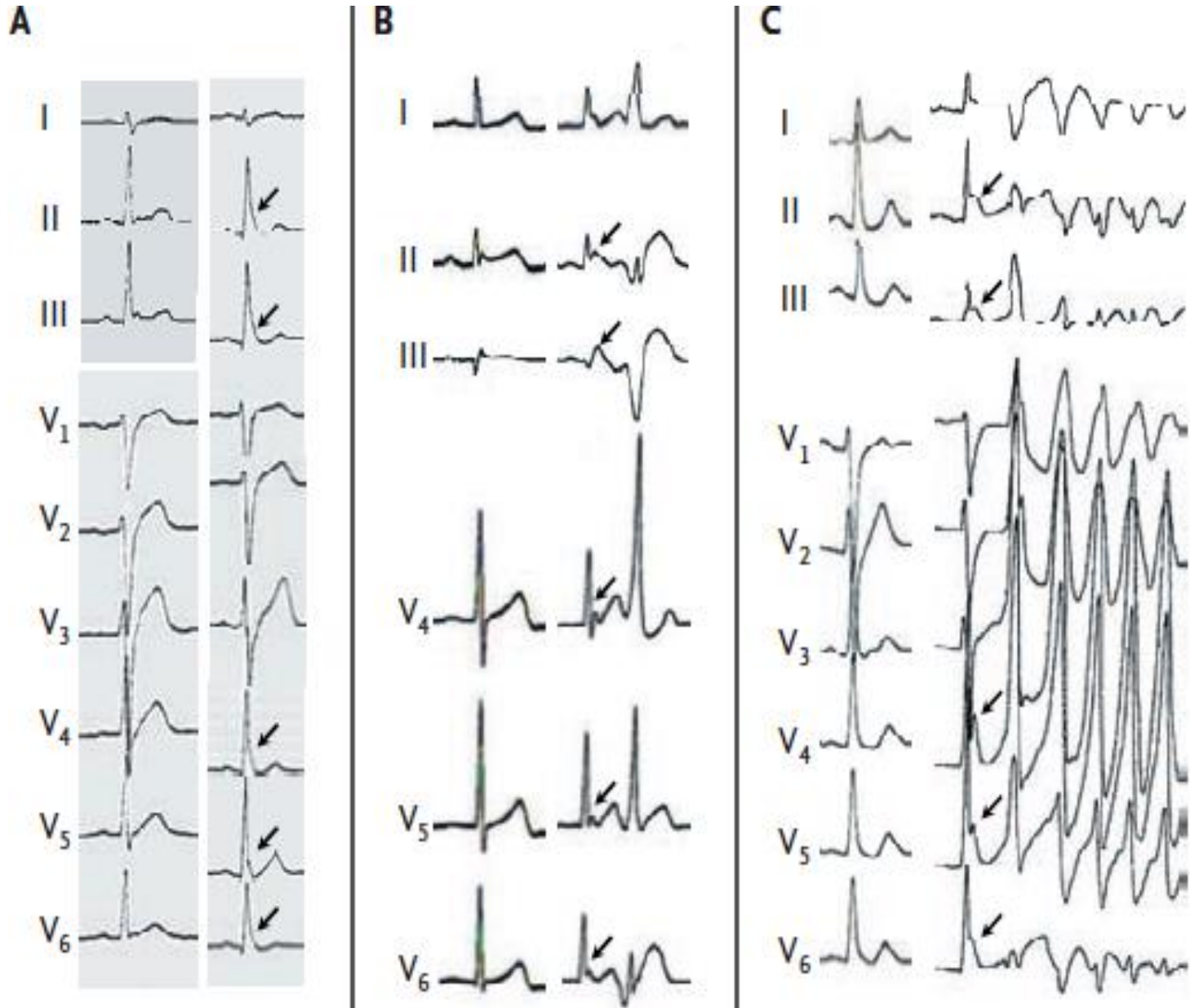
ECG normal



Repolarisation précoce

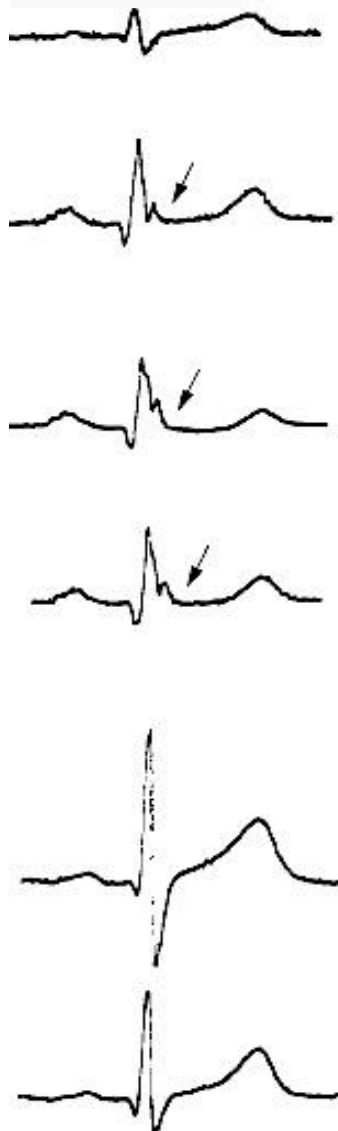


Syndrome de repolarisation précoce

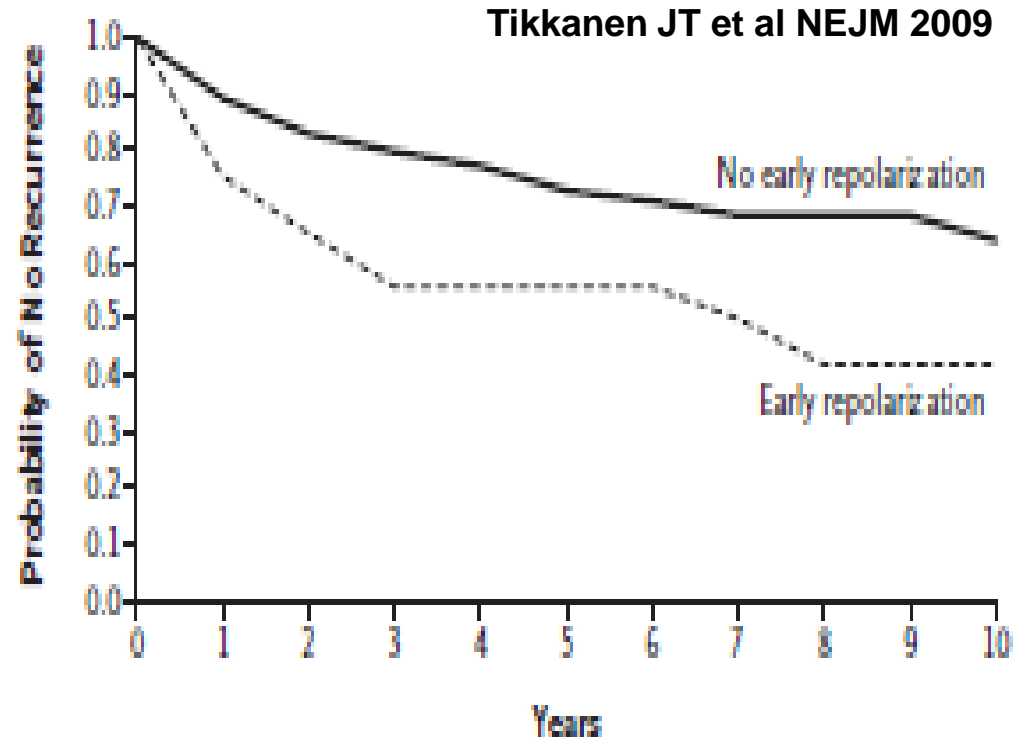
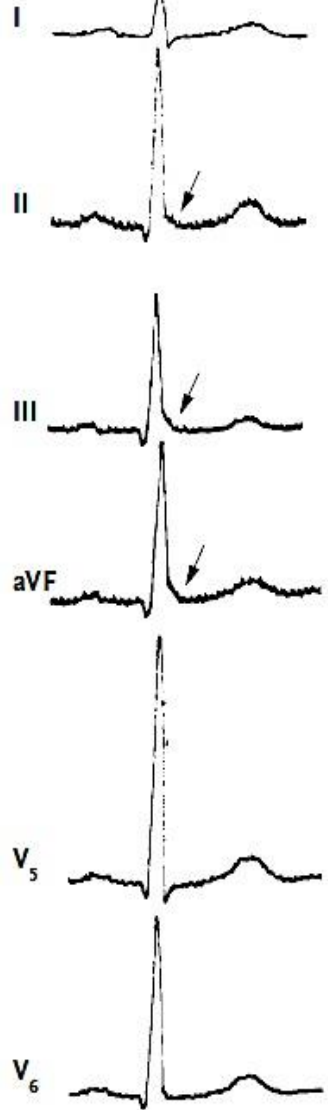


Syndromes de repolarisation précoce (ondes J)

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 ≥ 1 mm in ≥ 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 ≥ 1 mm in ≥ 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 ≥ 1 mm in ≥ 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

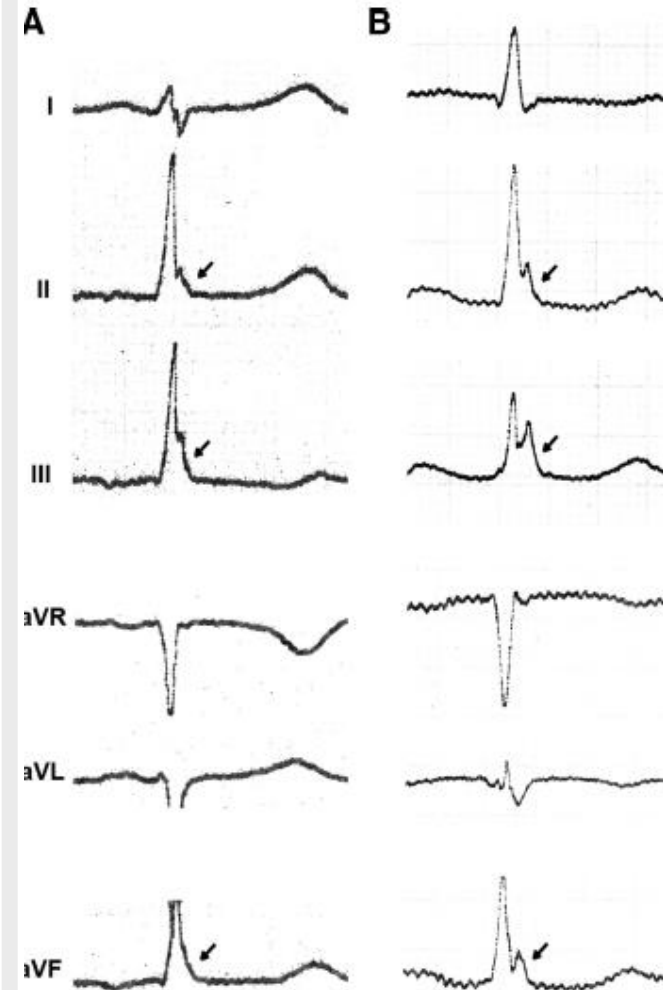
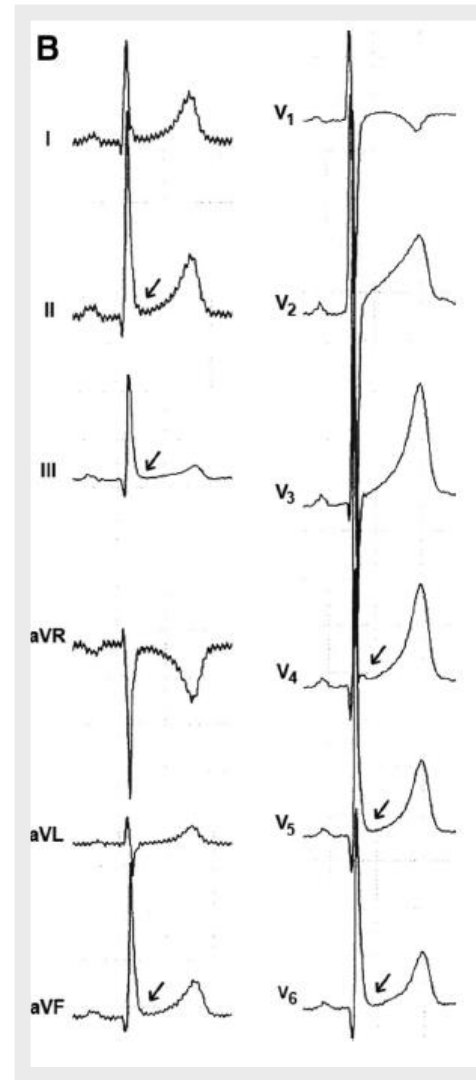
→ bon pronostic

ST descendant

→ mauvais pronostic

Mortalité CV :

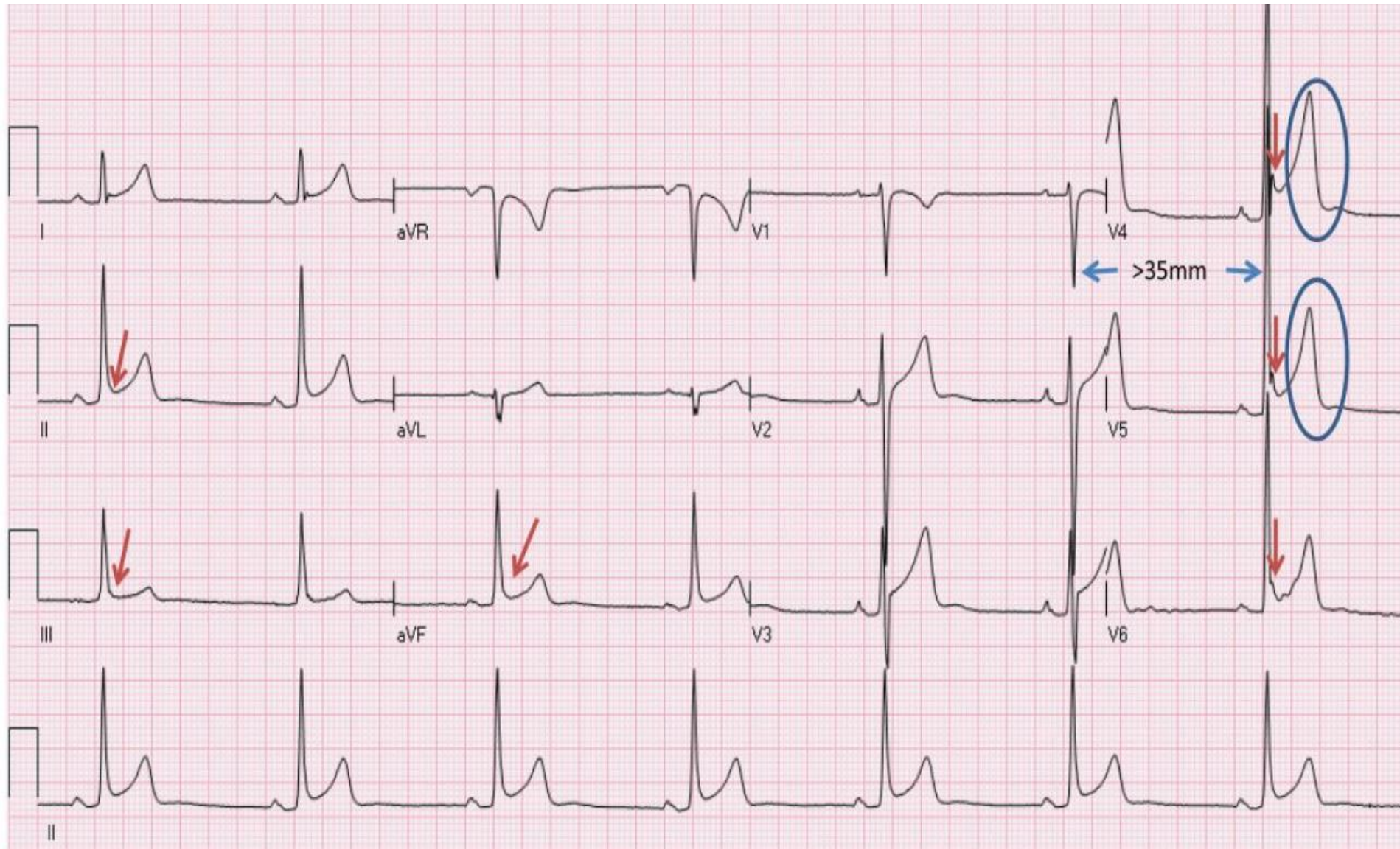
HR: 8.75 (CI 3.48-22.0, p<0.0001)



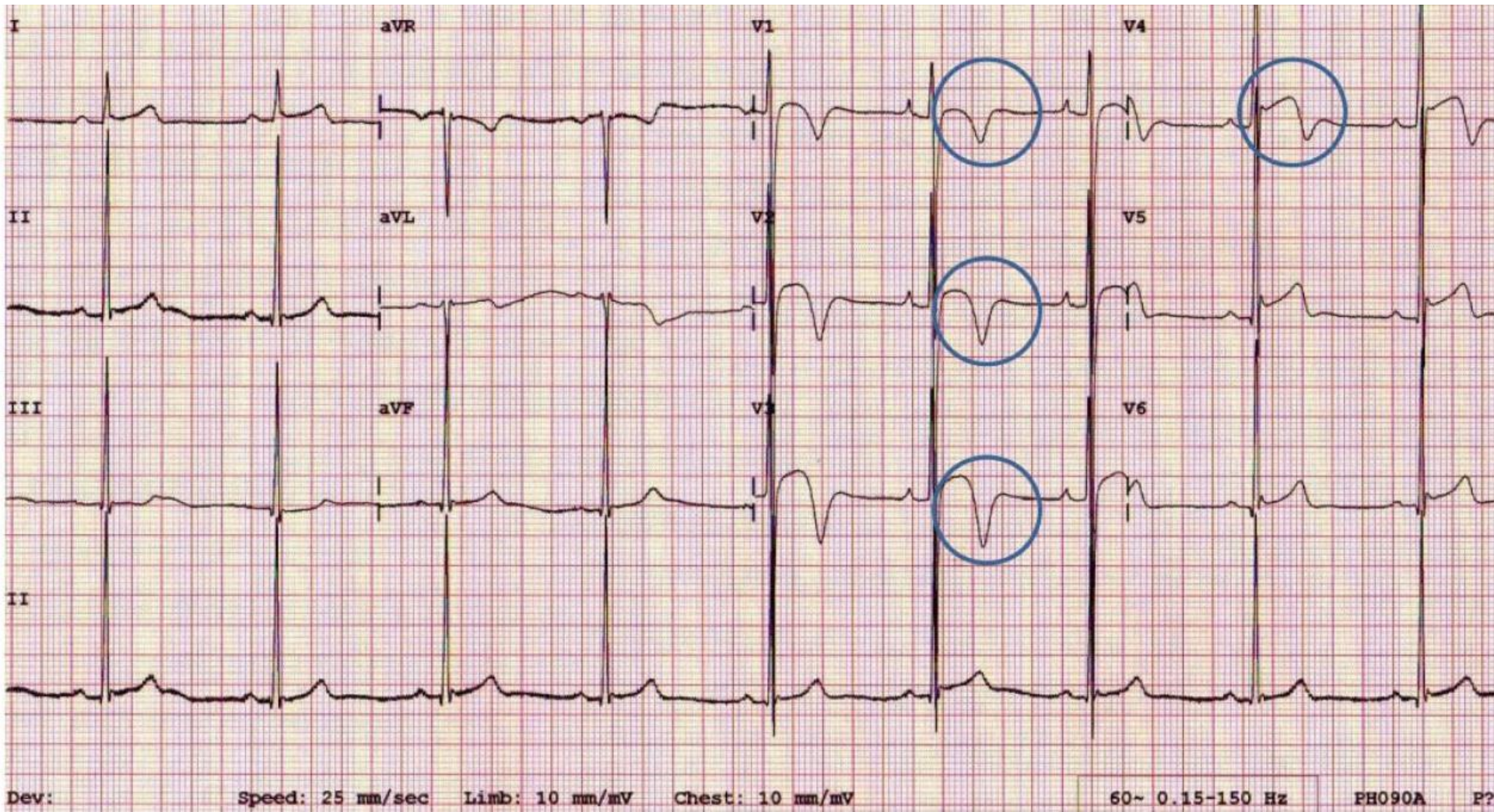
Tikkanen J et al. Circulation 2011

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