Cas cliniques en cardiologie du sport:

Canalopathie ?

Penser à la famille peut rapporter gros

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Conflits d’intérêts

Abvvie

Servier

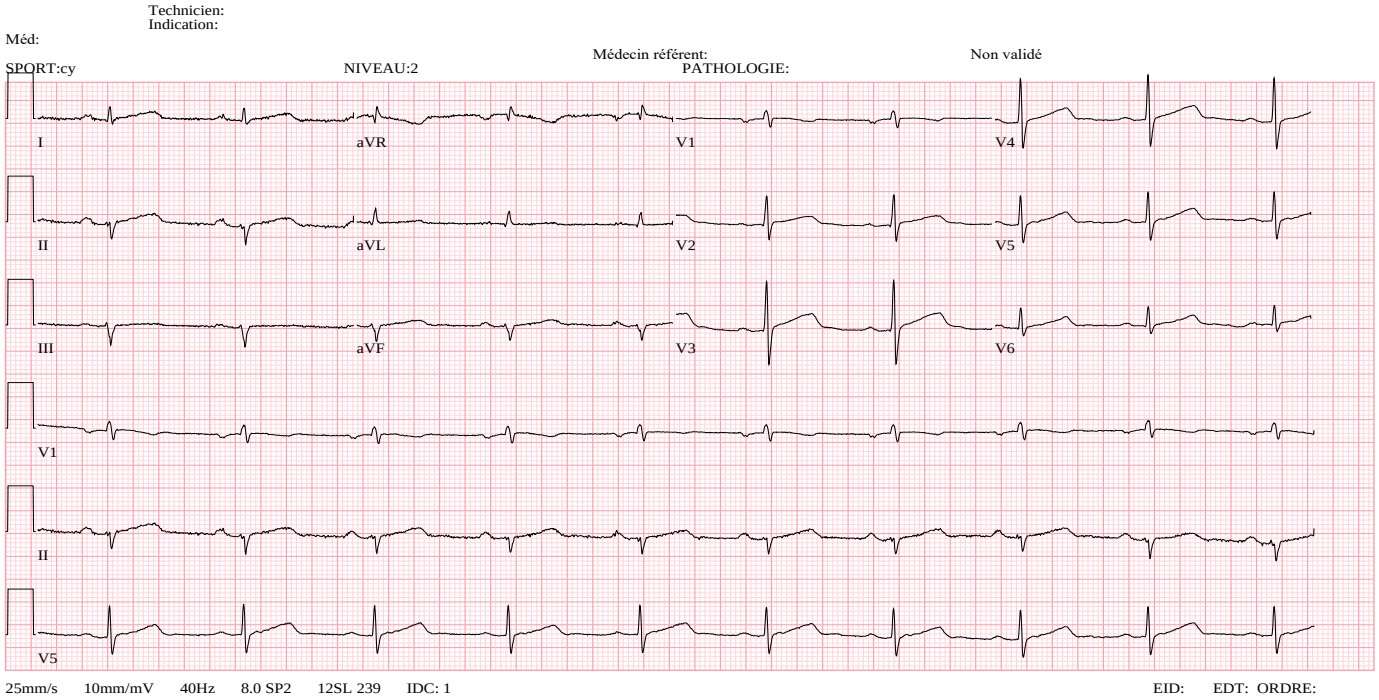
Sanofi

BMS

ECG d’un homme de 45 ans – cyclisme en compétition (entrainement fluctuant: entre 3 et 5 heures/semaine)

59	b. min-1	Bradycardie sinusale
198	ms	Hémi-bloc antérieur gauche
84	ms	QT allongé
490/485	ms	ECG anormal
47 -70	53	

QTc: 482 ms

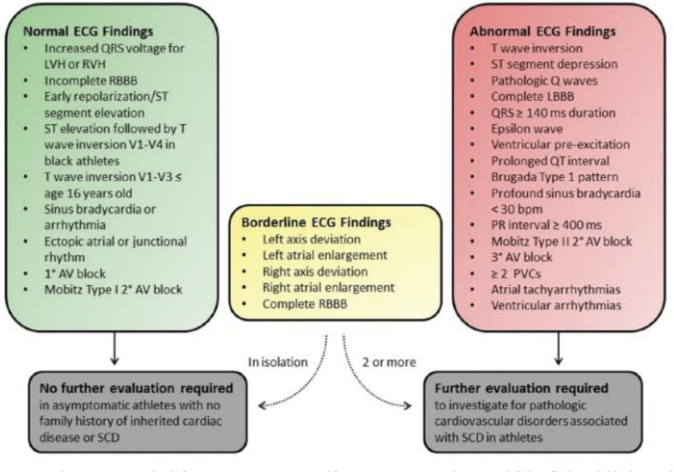


Quelle valeur seuil chez l'athlète ?

Valeur de références différentes chez l'athlète:

QTc prolongé si

≥ 470 ms (homme)
≥ 480 ms (femme)
≥ 500 ms (allongement marqué du QT)



Drezner J, et al. International recommendations for ECG interpretation in athletes. BJSM. 2017

Attention ne pas regarder que la durée mais aussi la morphologie

La morphologie de l'onde T peut également suggérer la présence d'un LQTS

« notched T wave in the lateral precordial leads where the amplitude of the second portion of the T wave following the notch is greater than the first portion of the T wave may represent LQT-2 even in the absence of overt QT prolongation »

« Un athlète avec un QTc prolongé sur un seul ECG ≠ diagnostic de SQTL MAIS cela nécessite d'une évaluation »

Drezner J, et al. International recommendations for ECG interpretation in athletes. BJSM. 2017

Schwartz Score		Points	Questionable in Athletes
Electrocardiographic findings			
A	QTc duration (ms) (Bazett formula)		
	≥480	3	
	460–470	2	x
	450 (in males)	1	x
B	Torsades de pointes*	2	
C	T-wave alternans	1	
D	Notched T wave in three leads	1	
E	Low heart rate for age	0.5	x
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 below the age of 30 among immediate family members	0.5	

Mutually exclusive. Source: Schwartz PJ, et al.[22] With permission from Wolters Kluver.

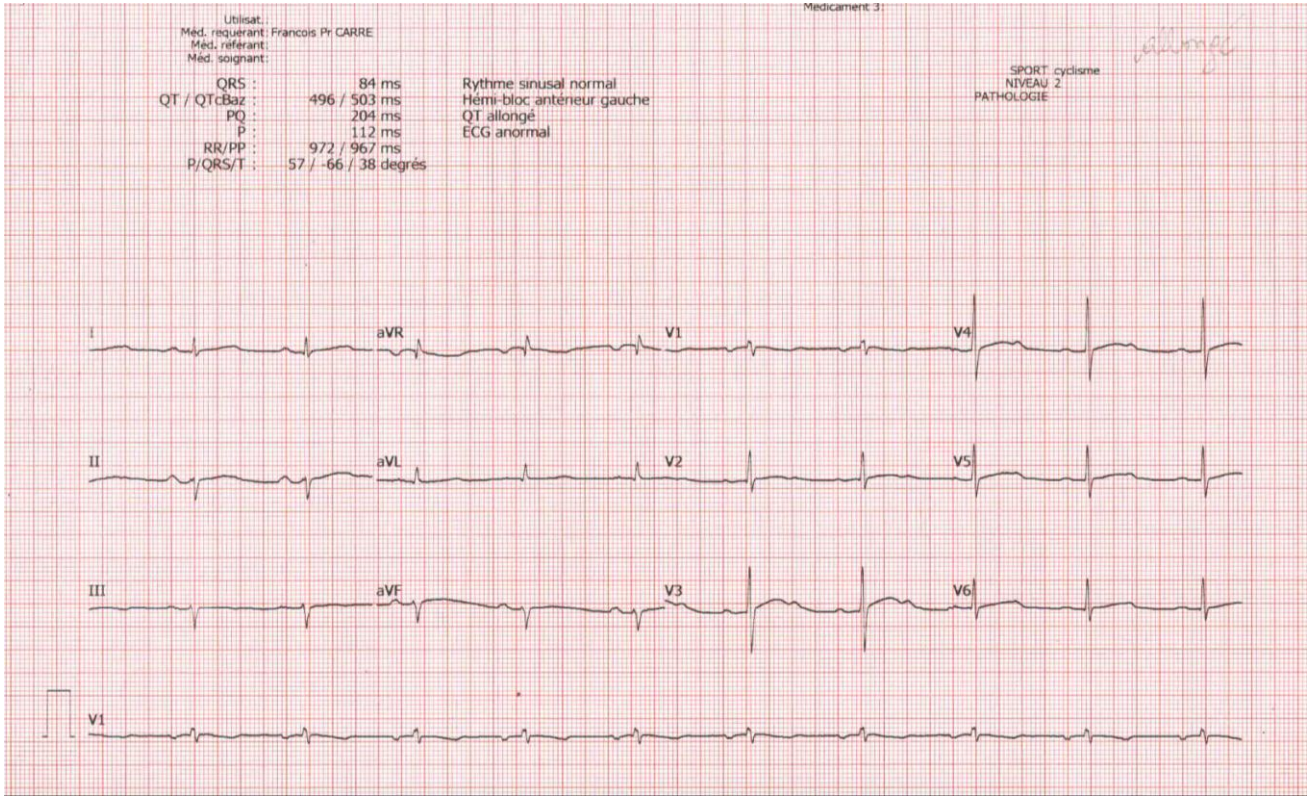
Autres évaluations

Médicaments prolongeant le QT ?

+ ionogramme

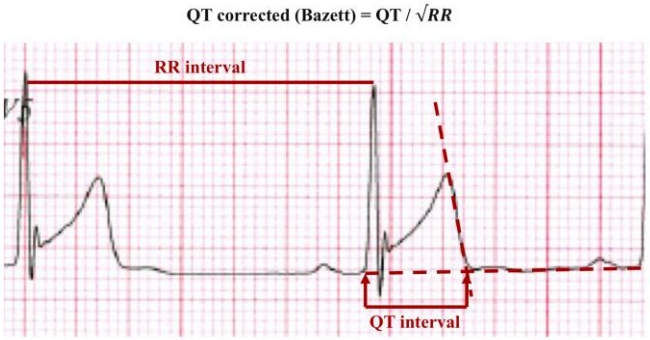
Refaire l’ECG après une pérode de désentrainement complet (2–4 semaines)

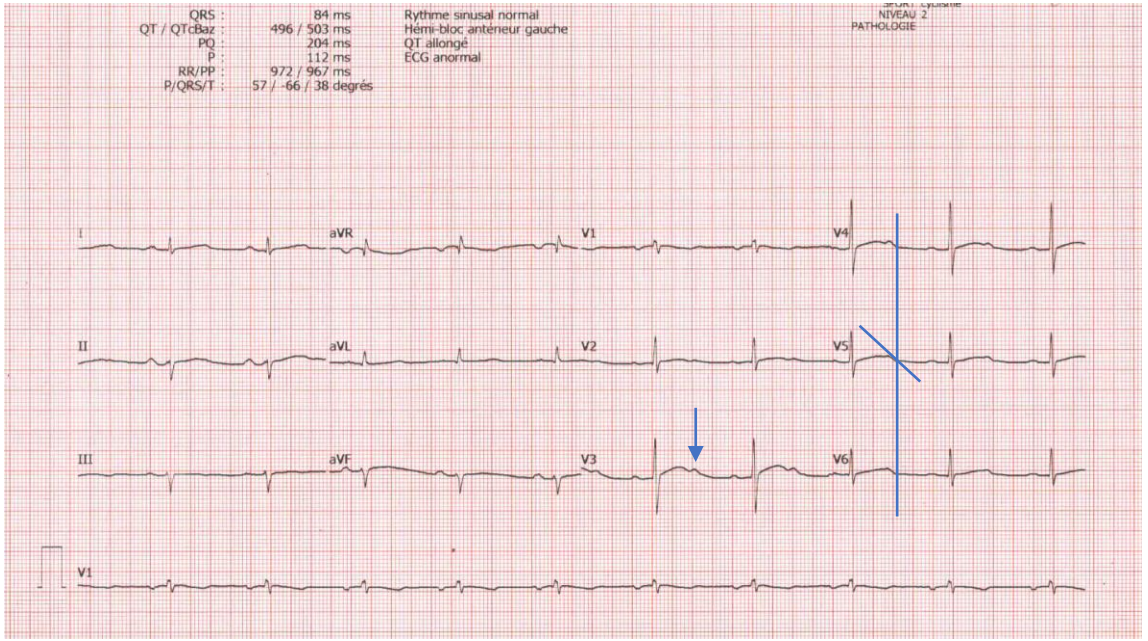
Consultation de suivi: Majoration de sa pratique: 7-8 heures d'entrainement / semaine



Difficulté de mesure du QT

QTc: 503 ms



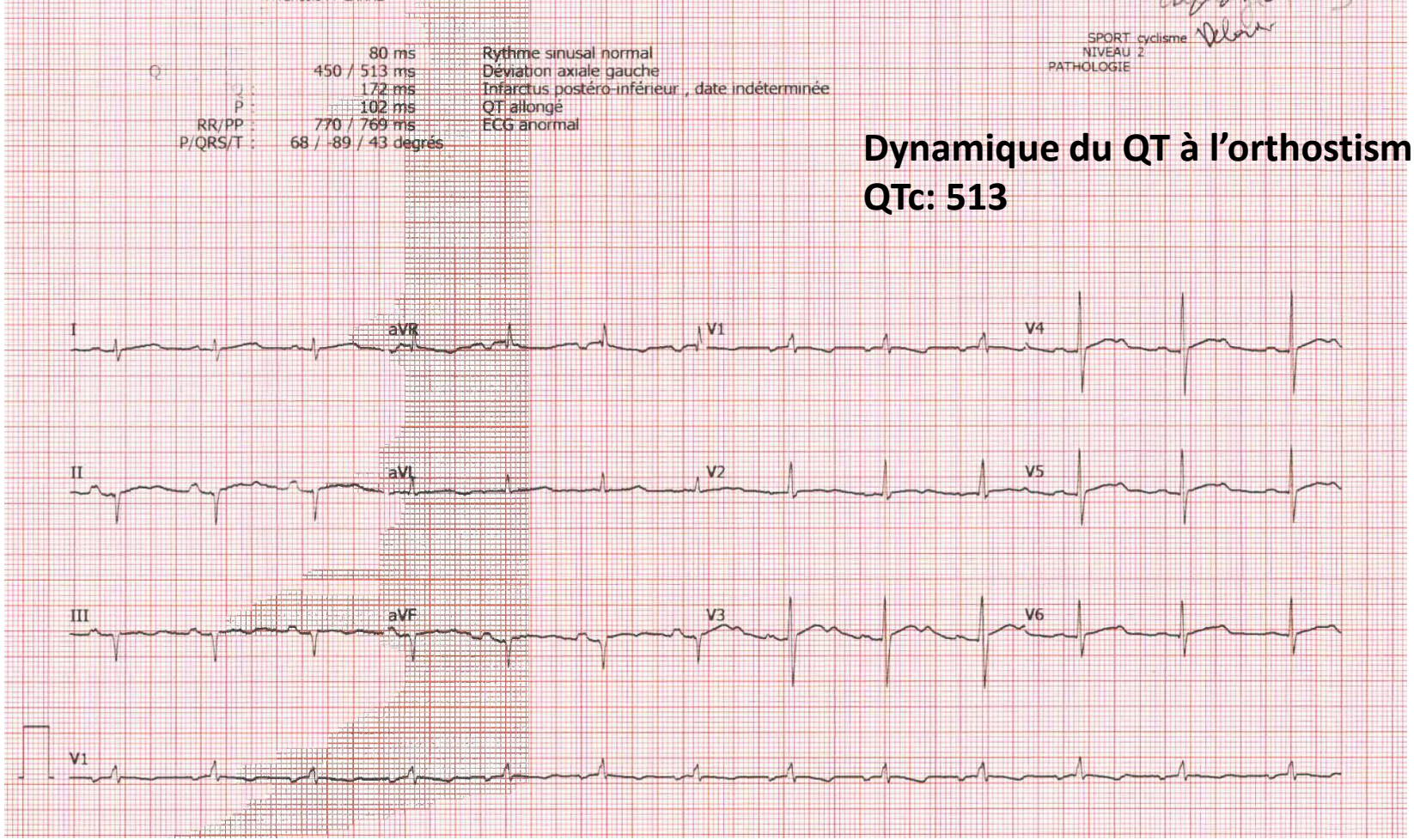


Méthode de la tangente
Ne pas inclure l'onde U

Dérivation II or V5

Autres évaluations

Evaluation de la dynamique du QT : Test d'effort (arythmie – évolution du QTc à 1 et 4 min de recupération), Holter ECG, test pharmacologique, test de stress mental



Dynamique du QT à l'orthostisme

QTc: 513

Autres évaluations

ECG chez les apparentés de 1er degré

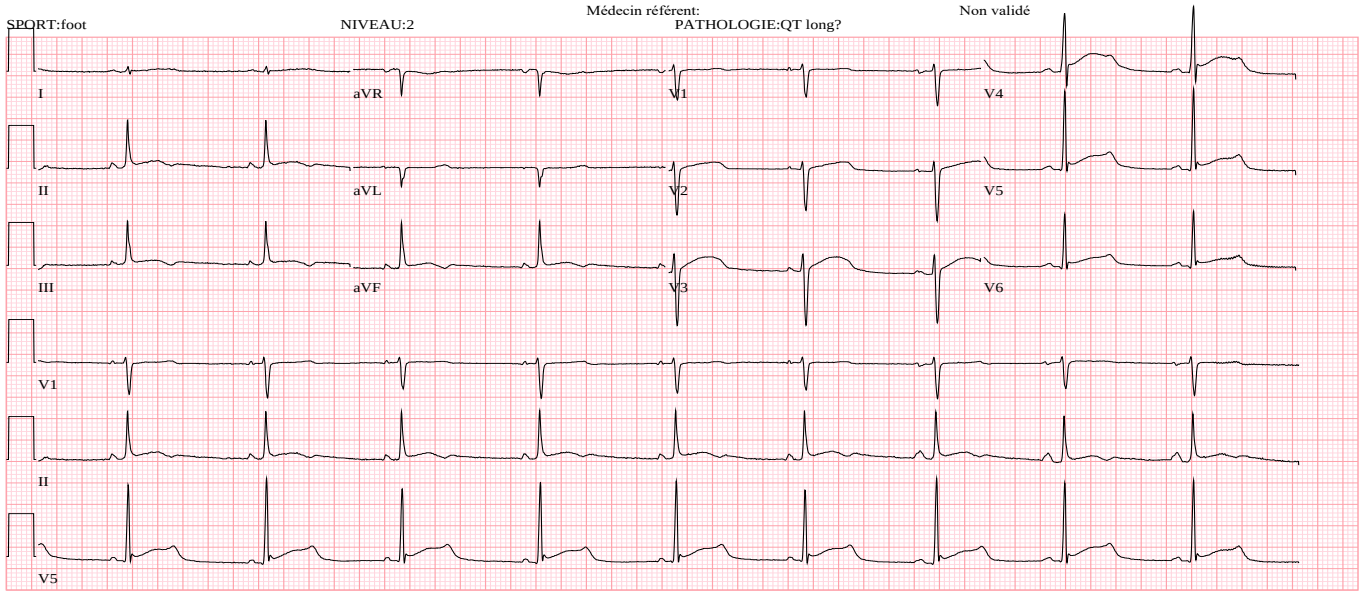
ECG de son plus grand fils 16 ans – cyclisme en compétition

ID :0000000000068293 21-mai-2019 09:57:07 THE FIRST SITE-DEFLT RECH. ORDINAIRE

57	b. min-1	** * Analyse de l'ECG pédiatrique * **
114	ms	Bradycardie sinusale
76	ms	Segment S-T sus-décalé ; repolarisation précoce, péricardite ou lésion
482/469	ms	Limite QT allongé
76 86	47	

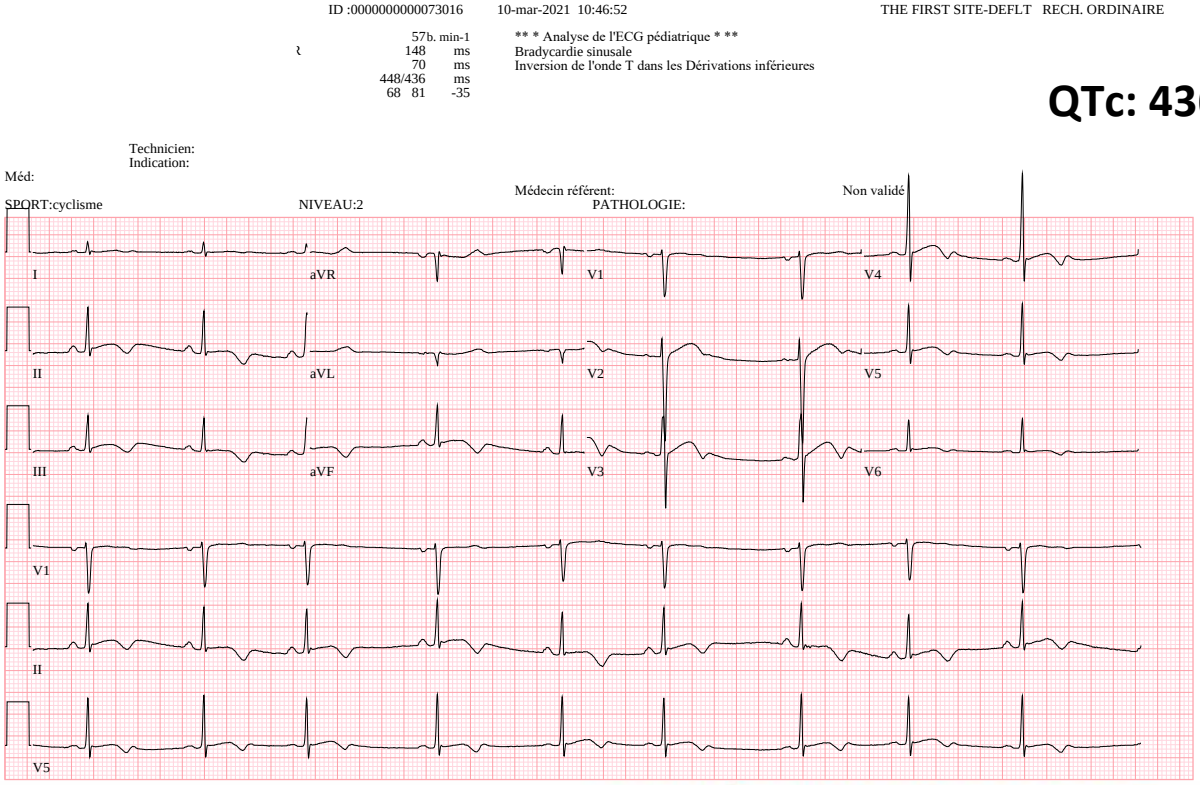
QTc: 407 ms

SPORT:foot NIVEAU:2 Médecin référent: PATHOLOGIE:QT long? Non validé



25mm/s 10mm/mV 40Hz 8.0 SP2 12SL 239 IDC: 1 EID: EDT: ORDRE:

ECG de son plus jeune fils (14 ans) - cyclisme en compétition

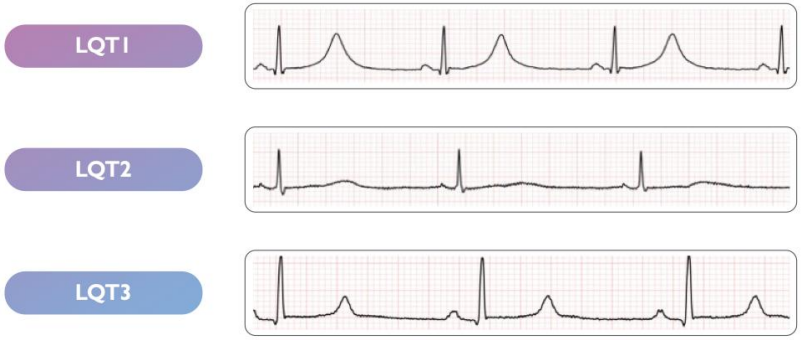


QTc: 436 ms ?

Autres évaluations

Test génétique si la suspicion clinique est élevée

Variant pathogène de niveau 5 dans le gène KCNH2 chez le père et ses 2 fils.



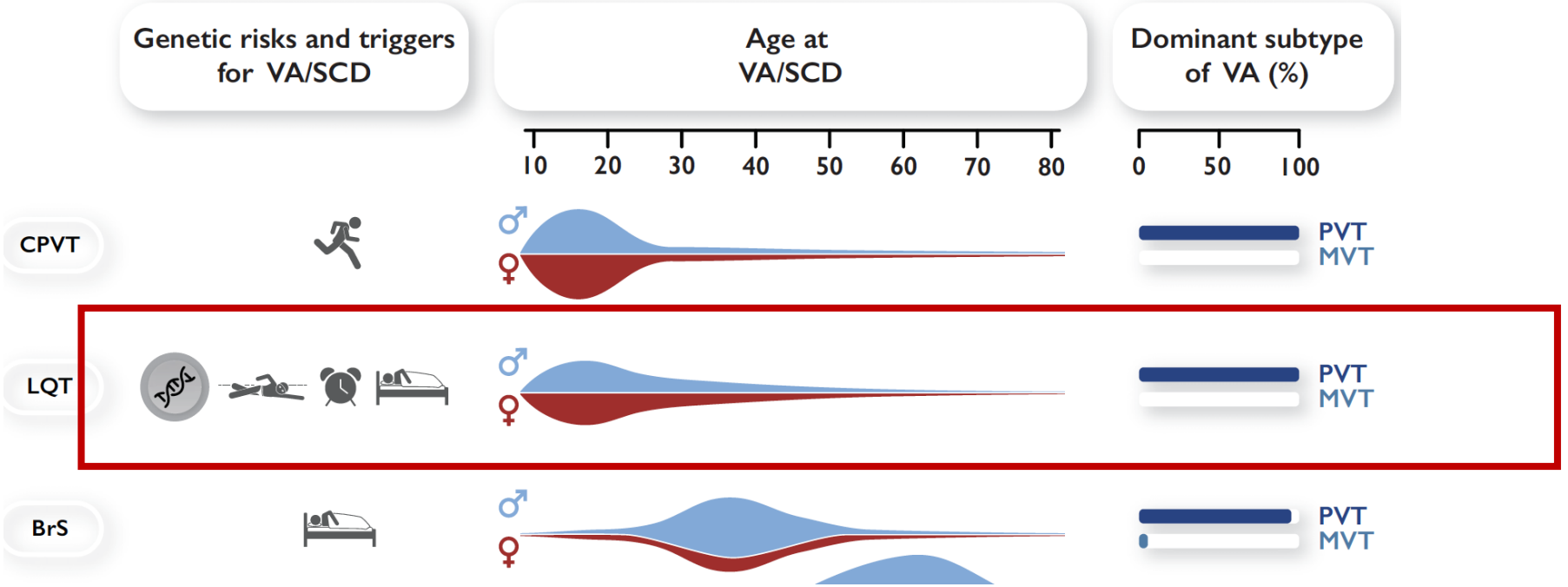
Jeppenfeld K, et al. 2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. EHJ 2022

Quelle prise en charge ?

General recommendations to prevent SCD		
The following is recommended in LQTS: • Avoid QT-prolonging drugs.[c] • Avoid and correct electrolyte abnormalities. • Avoid genotype-specific triggers for arrhythmias.[943]	I	C
Beta-blockers, ideally non-selective beta-blockers (nadolol or propranolol), are recommended in LQTS patients with documented QT interval prolongation, to reduce risk of arrhythmic events.[940,945,946]	I	B
Mexiletine is indicated in LQT3 patients with a prolonged QT interval.[948]	I	C
Beta-blockers should be considered in patients with a pathogenic mutation and a normal QTc interval.[82]	IIa	B

Jeppenfeld K, et al. 2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. EHJ 2022

Et le sport ?



Jeppenfeld K, et al. 2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. EHJ 2022

Différences en fonction du type de SQTL

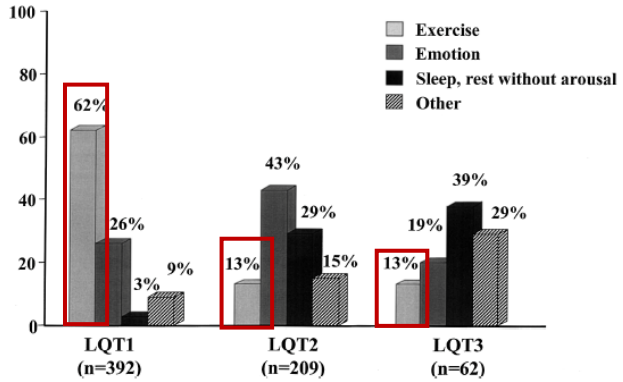


Figure 1. Triggers for cardiac events according to 3 genotypes. Numbers in parentheses indicate number of triggers, not number of patients.

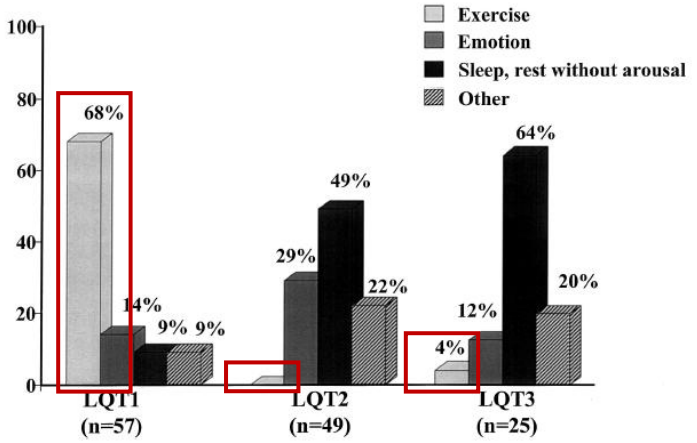


Figure 2. Lethal cardiac events according to 3 classified triggers in 3 genotypes. Numbers in parentheses indicate number of patients.

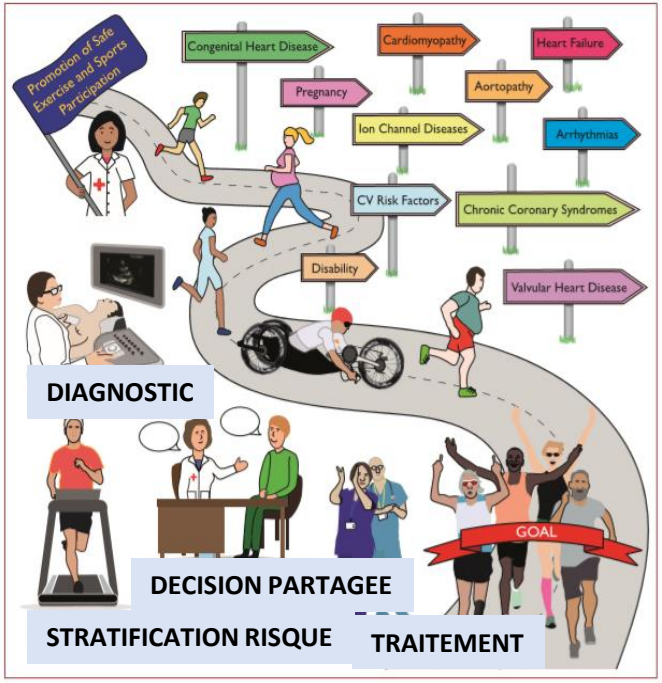
TABLE 3. Genotype and Specific Triggers

Genotype	
Auditory stimuli	
LQT1	7/320 (2%)
LQT2	45/176 (26%)
LQT3	4/58 (7%)
Swimming	
LQT1	107/320 (33%)
LQT2	1/176 (0.6%)
LQT3	0/58

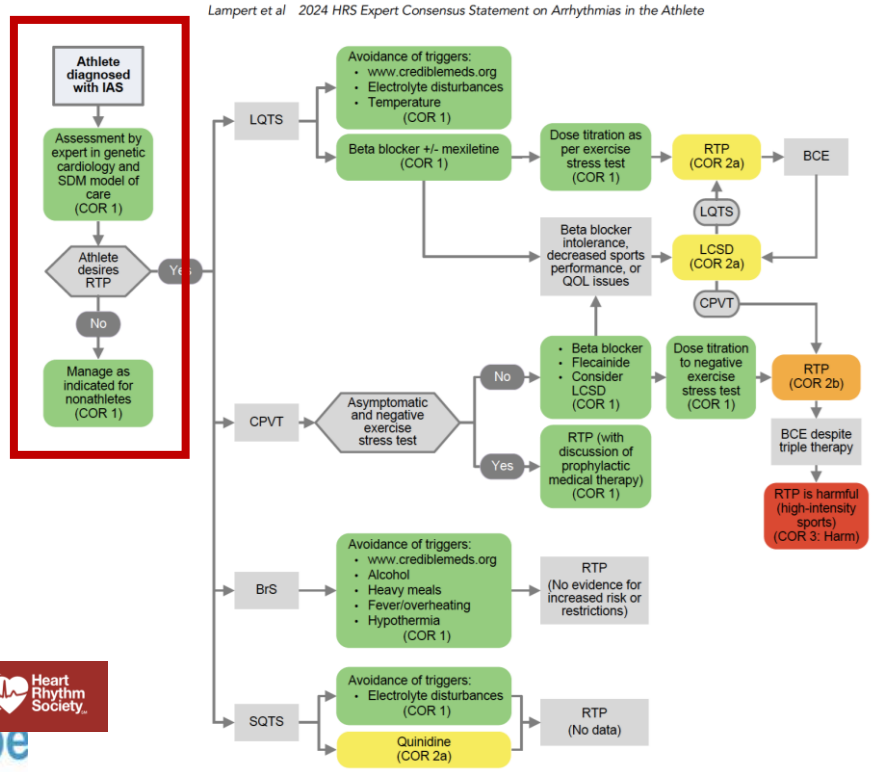
Attention au LQT1 à l'effort ++ surtout la natation

Schwartz PJ, et al. Genotype-phenotype correlation in the long-QT syndrome: gene-specific triggers for life-threatening arrhythmias. Circulation 2001

Reco ESC 2020 vs HRS 2024: les lignes directrices



faibles niveaux de preuves



HeartRhythm Heart Rhythm Society

www.forumeurope

Recommandations : sport ? ESC 2020

Recommendations	Class[a]	Level[b]
It is recommended that all exercising individuals with LQTS with prior symptoms or prolonged QTc be on therapy with beta-blockers at target dose.[529]	I	B
It is recommended that exercising individuals with LQTS should avoid QT prolonging drugs (www.crediblemeds.org) and electrolyte imbalance such as hypokalaemia and hypomagnesaemia.[529]	I	B
Shared decision making should be considered regarding sports participation in patients with genotype-positive/phenotype-negative LQTS (i.e. <470/480 ms in men/women). Type and setting of sports (individual vs. team), type of mutation, and extent of precautionary measures	IIa	C
Participation in high-intensity recreational and competitive sports, even when on beta-blockers, is not recommended in individuals with a QTc>500 ms or a genetically confirmed LQTS with a QTc≥470 ms in men or ≥480 ms in women.	III	B
Participation in competitive sports (with or without ICD) is not recommended in individuals with LQTS and prior cardiac arrest or arrhythmic syncope.	III	C

LQTS avec symptômes ou QT prolongé: **traitement béta bloquants**

Eviter médicaments allongeant QT, désordres hydroélectrolytiques

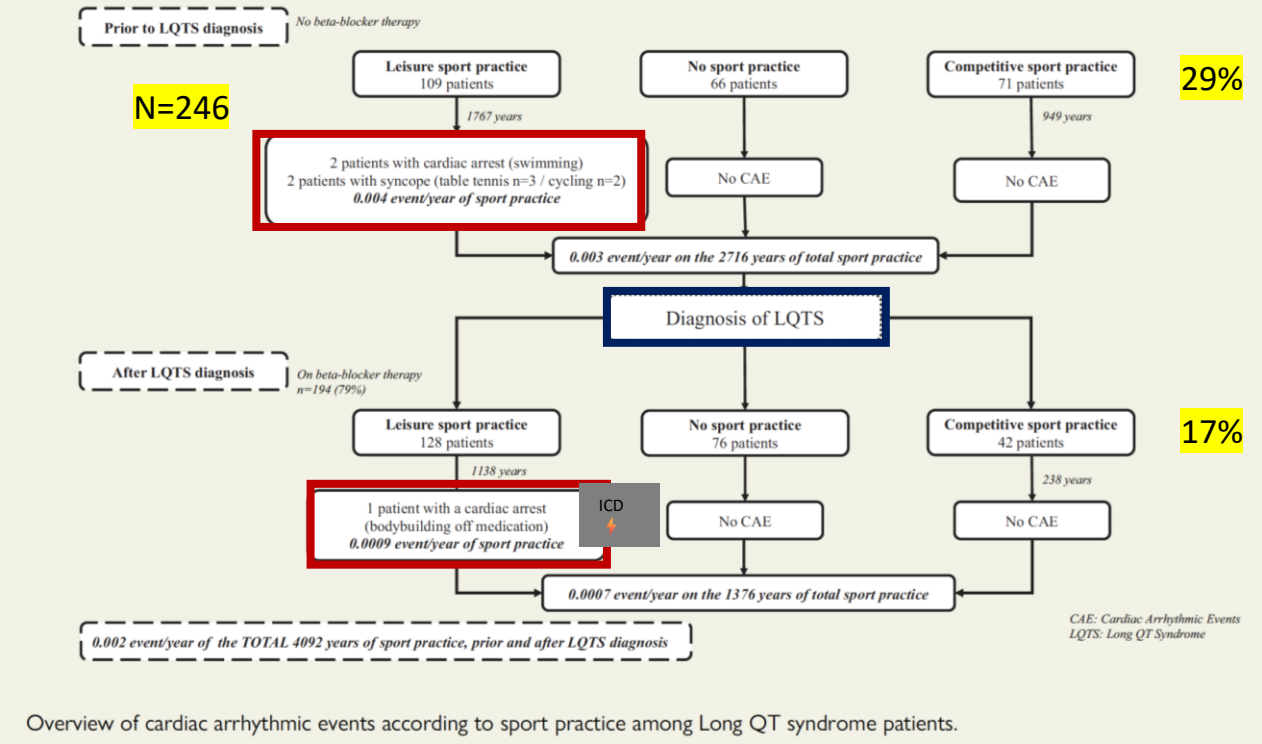
Décision partagée si génotype + / phénotype –
(type de sport, mutation, précautions..)

Sport en compétition **non recommandé** (même si sous BB)
si **QTc > 500 ms**
ou confirmé génétiquement **≥470ms chez ♂ ≥ 480 ms chez ♀**

Sport en compétition non recommandé si antécédent de MS ou syncope rythmique

Pelliccia A, et al. 2020 ESC Guidelines on sports cardiology and exercise in patients with cardiovascular disease. Eur HJ. 2020

Etude française



Overview of cardiac arrhythmic events according to sport practice among Long QT syndrome patients.

Davydoff C, et al. Does sports participation increase risk in patients with long QT syndrome? Results from a large French cohort. Europace 2022.

Table 1 General characteristics of the population

	Total ($n = 246$)	No sports practice after LQTS diagnosis ($n = 76$)	Sports practice after LQTS diagnosis ($n = 170$)	*P*-value
Female	141 (57%)	44 (58%)	97 (57%)	0.09
Age at survey (years) median (min–max)	43 (4.2–80)	51 (12–80)	38 (4.3–74)	**<0.001**
Proband	84 (34%)	18 (24%)	66 (39%)	**0.03**
QTc at diagnosis (ms)	457 (428; 482)	463 (424; 487)	453 (429; 470)	0.34
Mutations				0.24
KCNQ1	69 (28%)	14 (18%)	56 (33%)	
KCNH2	82 (33%)	29 (38%)	53 (31%)	
SCN5A	7 (3%)	2 (3%)	5 (3%)	
Rare mutations, no mutation, or NA	88 (36%)	31 (41%)	56 (33%)	
Circumstances of diagnosis				
Systematic ECG	31 (13%)	4 (5%)	27 (16%)	**0.02**
Family screening	159 (64%)	50 (66%)	109 (64%)	0.61
Symptoms	56 (23%)	22 (29%)	34 (20%)	0.12
Symptoms prior to the diagnosis				
Faintness	12 (5%)	4 (5%)	8 (5%)	1
Syncope	52 (21%)	24 (32%)	28 (16%)	**0.007**
Aborted cardiac arrest	6 (2%)	0 (0%)	6 (4%)	0.18
None	176 (72%)	48 (63%)	128 (75%)	0.051
Beta-blocker therapy	194 (79%)	59 (78%)	135 (79%)	0.75
Nadolol	161 (65%)	45 (59%)	116 (68%)	1
Inobservance	6 (3.1%)	4 (5%)	2 (1.2%)	0.08
Implanted cardiac devices				
ICD	15 (6%)	6 (8%)	9 (5%)	0.58
Pacemaker	13 (5%)	7 (9%)	6 (3.5%)	0.12
Sympathectomy	1 (0.4%)	1 (1.3%)	0 (0%)	0.31
Cardiac arrest prior to beta-blocker therapy	8 (3.3%)	2 (2.6%)	6 (3.5%)	0.74
Cardiac arrest on beta-blocker therapy	3 (1.2%)	1 (1.3%)	2 (1.2%)	1

Comparisons were made between the group with no sports practice after LQTS diagnosis vs. sports practice after LQTS diagnosis. LQTS, long QT syndrome; ICD, implantable cardioverter defibrillator; NA, non-available. Bold *p*-values denote statistical significance.

Table 6 Details of LQTS patients with CAE

Patient	Time of arrhythmic event	Type of event	Age	Sex	Mutation	QTc without/with beta-blocker (ms)	Type of sport (Mitchell's classification)	Medical therapy	Sports practice after CAE
1	Prior to LQTS diagnosis	Cardiac arrest	10	F	KCNQ1	540/490	Swimming (IIC) Leisure	None Nadolol after CAE	Yes Leisure
2	Prior to LQTS diagnosis	Cardiac arrest	11	M	None	455/465	Swimming (IIC) Leisure	None Nadolol after CAE	Yes Leisure
	After LQTS diagnosis	Cardiac arrest	16	M	None	455/465	Bodybuilding (IIIA) Leisure	Nadolol (non-compliance)	Yes Leisure
3	Prior to LQTS diagnosis	Syncope ($n = 2$)	51	F	None	535/450	Cycling (IIIC) Leisure	Hydroxychloroquine Nadolol after CAE	Yes Leisure
4	Prior to LQTS diagnosis	Syncope ($n = 3$)	6	M	KCNQ1	514/493	Table tennis (IB) Leisure	None Nadolol after CAE	Yes Leisure

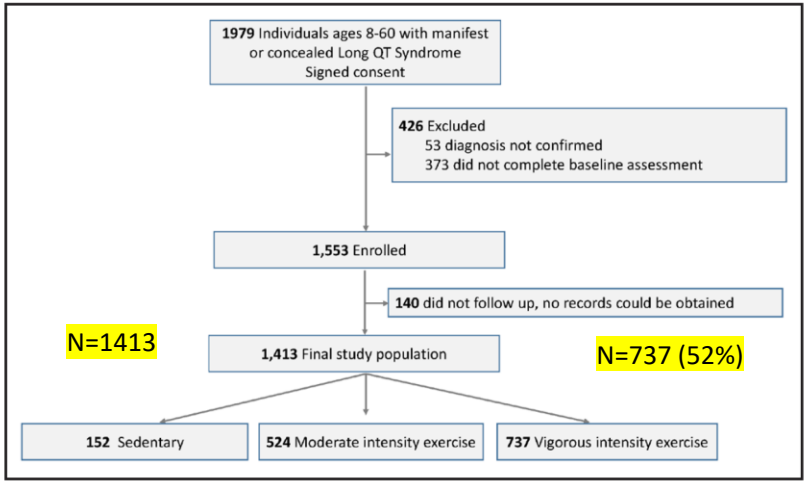
CAE, cardiac arrhythmic event; LQTS, long QT syndrome.

5 évènements – 4 patients – 1 évènement sous traitement

What's new?

- In patients with long QT syndrome (LQTS) and a low-risk profile, European 'real-life' competitive and leisure sports practice is associated with a very low rate of cardiac arrhythmic event (CAE).
- Good compliance to beta-blocker therapy was associated with no CAE during sports practice in LQTS patients.

Etude multicentrique prospective



Caractéristiques de la population
Génotype + : 90 % - SQTL1: 49%
Traitement (bêta-bloquants, dénervation sympathique et/ou un DAI): 91 %
DAI: 25%

Pratique sportive intense: 52%
Compétition: 29%

Événements cliniques : 37 événements composites (décès, MS récupérée, syncope d'origine rythmique, choc approprié) **dont 1 ACR et 1 MS dans groupe Sport non intense vs 1 ACR dans le groupe sport intense.**

Taux d'événements à 3 ans : 2,6 % pour le groupe exercices intenses et de **2,7 %** pour le groupe exercices moins intenses.

Lampert R, et al. Vigorous Exercise in Patients With Congenital Long QT Syndrome: Results of the Prospective, Observational, Multinational LIVE-LQTS Study. Circulation. 2024

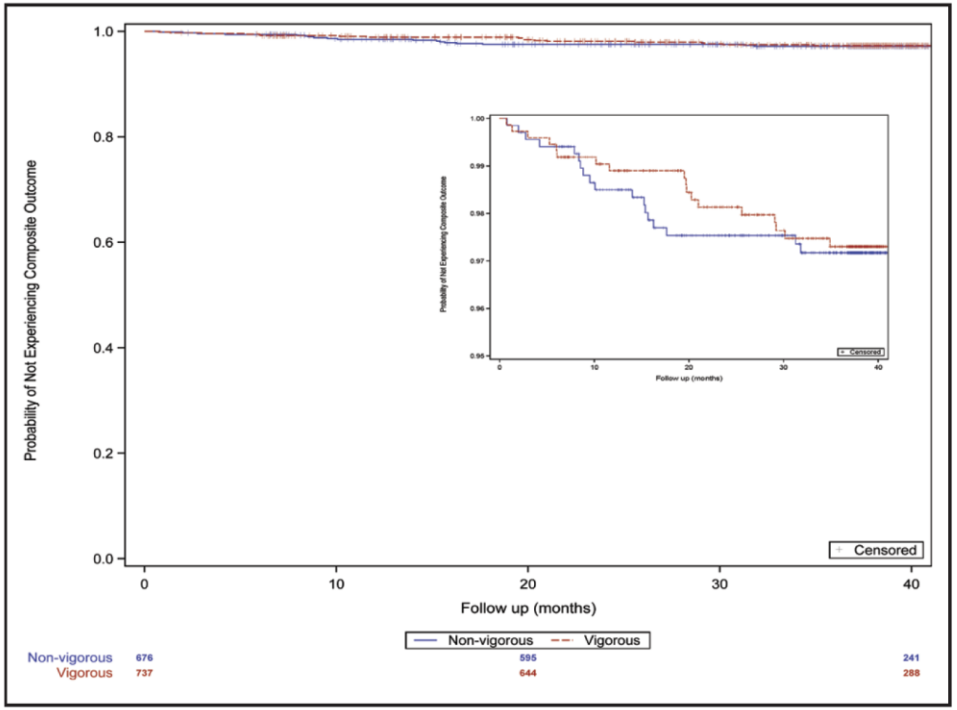


Figure 2. Kaplan-Meier survival curve for freedom from composite end point (death, sudden cardiac arrest, appropriate implantable cardioverter defibrillator shock, or arrhythmic syncope) by exercise group.

There was no statistically significant difference in freedom from composite end points between the those exercising vigorously and those exercising nonvigorously. Inset shows a magnified *y* axis.

Clinical Perspective

What Is New?

- This is the first prospective study to investigate whether vigorous exercise increases risk of arrhythmic events in individuals with congenital long QT syndrome.
- In this study, long QT syndrome–triggered cardiac events were low in both those exercising vigorously and those exercising nonvigorously, and there was no statistical difference in event rate. CIs were wide, and noninferiority not demonstrated.
- Findings were similar in vigorous-competitive athletes 14 to 22 years of age.

What Are the Clinical Implications?

- These findings will further inform shared decision-making discussions between patient and physician about exercise and competitive sports participation.

Lampert R, et al. Vigorous Exercise in Patients With Congenital Long QT Syndrome: Results of the Prospective, Observational, Multinational LIVE-LQTS Study. Circulation. 2024

Recommandations de pratique sportive ? HRS 2024

Recommendations for athletes with long QT syndrome

COR	LOE	Recommendations
2a	B-NR	1. In athletes with LQTS under expert assessment and supervision, return to play is reasonable in a shared decision-making model after risk assessment, education, and initiation of appropriate therapies.[26,194,379–382,398]
1	B-NR	2. In athletes with LQTS, review and/or cessation of medications known to prolong the QT interval is recommended, and whenever possible, prevention and correction of electrolyte disturbances are recommended.[399,400]
1	B-NR	3. In athletes with asymptomatic LQTS and a normal corrected QT interval (concealed variant-positive LQTS), initiation of QT-related preventative measures is recommended prior to return to play.[399,400]
2a	B-NR	4. In athletes with asymptomatic LQTS and a corrected QT interval < 470 ms, therapy with beta blockers can be useful.[387]
1	B-NR	5. In athletes with LQTS with symptoms and/or a corrected QT interval > 470 ms, guideline-directed and genotype/patient-tailored therapy with medications, LCSD, and/or device therapy should be optimized fully before return to play.[26,387,390,401]
1	B-NR	6. In athletes with LQTS on beta blocker therapy, nonselective beta blockers (especially nadolol and propranolol) are recommended, with dosing tailored to the patient's risk profile and response to therapy.[387]
2a	B-NR	7. For athletes with LQTS and severe bradycardia, other treatment configurations besides beta blockers (eg, alternative medical therapy, LCSD, and device therapy) are reasonable.[26,389–391]
2a	B-NR	8. In athletes with LQTS (including type 1), participation in swimming/diving is reasonable with appropriate precautions.[402]
1	B-NR	9. In athletes with LQTS who are unable to tolerate beta blockers or who have ongoing events on beta blockers, treatment intensification with medication, LCSD, and/or device therapy should be done and reoptimized fully prior to return to play.[26,390,401,403]

Lampert R, et al. 2024 HRS expert consensus statement on arrhythmias in the athletes: evaluation, treatment and return to play. HRS 2024

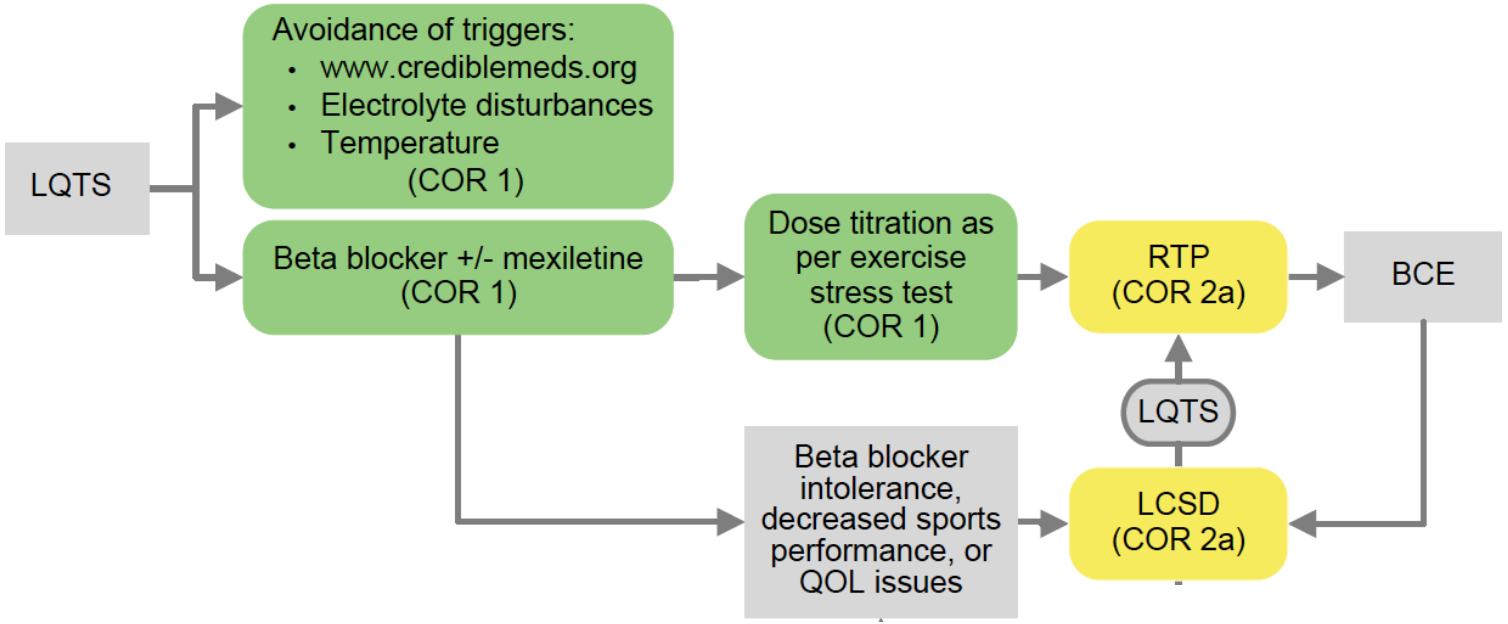
Centre spécialisé
Décision partagée
DAE accessible / discuter d'un DAE personnel
Plan d'Action d'Urgence

Pas donnée soutenant des restrictions chez les athlètes avec SQTL asymptomatiques avec QT corrigé normal

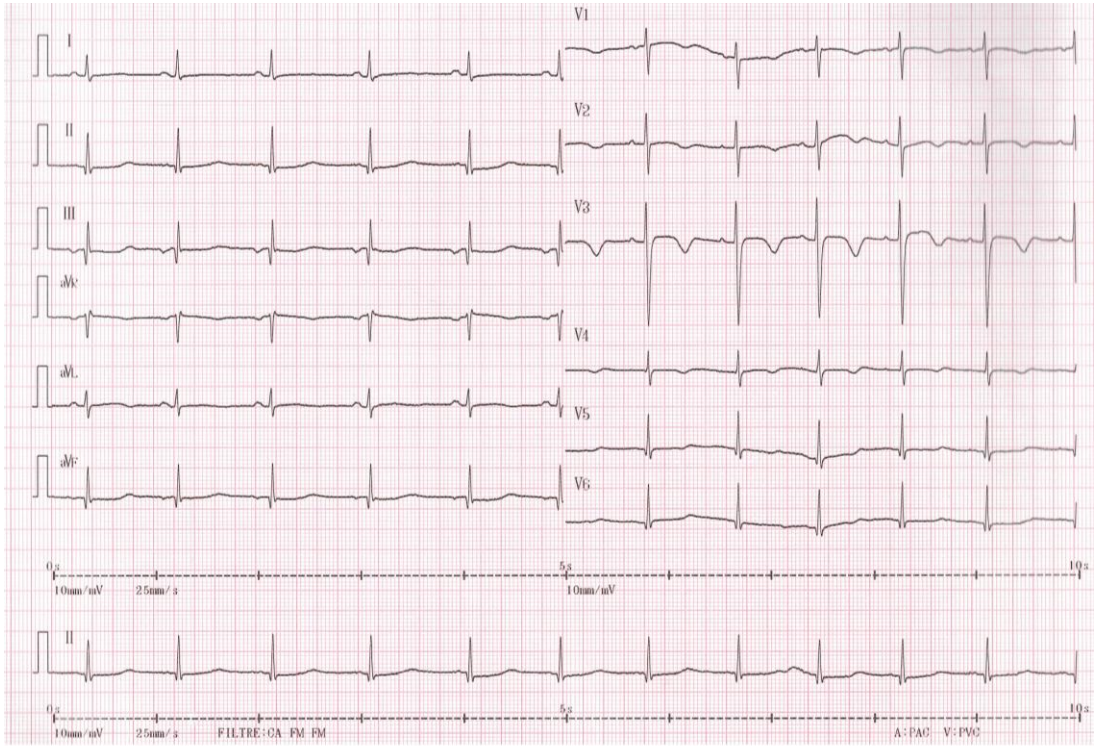
Titration BB: ↘ FC de 15-20% sur EE (3 mois)
Nadolol 0.8-1.5 mg/kg

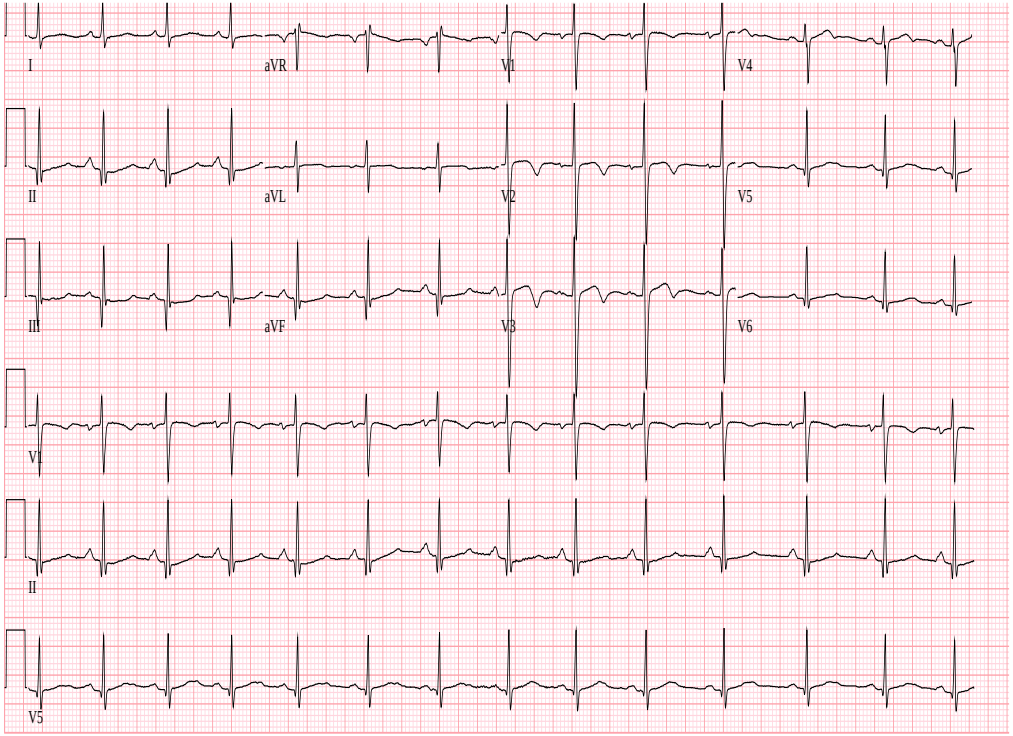
LQT1 + natation / plongée :
Plan de gestion personnalisé
DAE personnel à proximité
ne pas nager seul
nager en piscine.

Recommandations de pratique sportive ? HRS 2024



Jeune femme – kayak en haut niveau
ECG systématique réalisé dans son suivi





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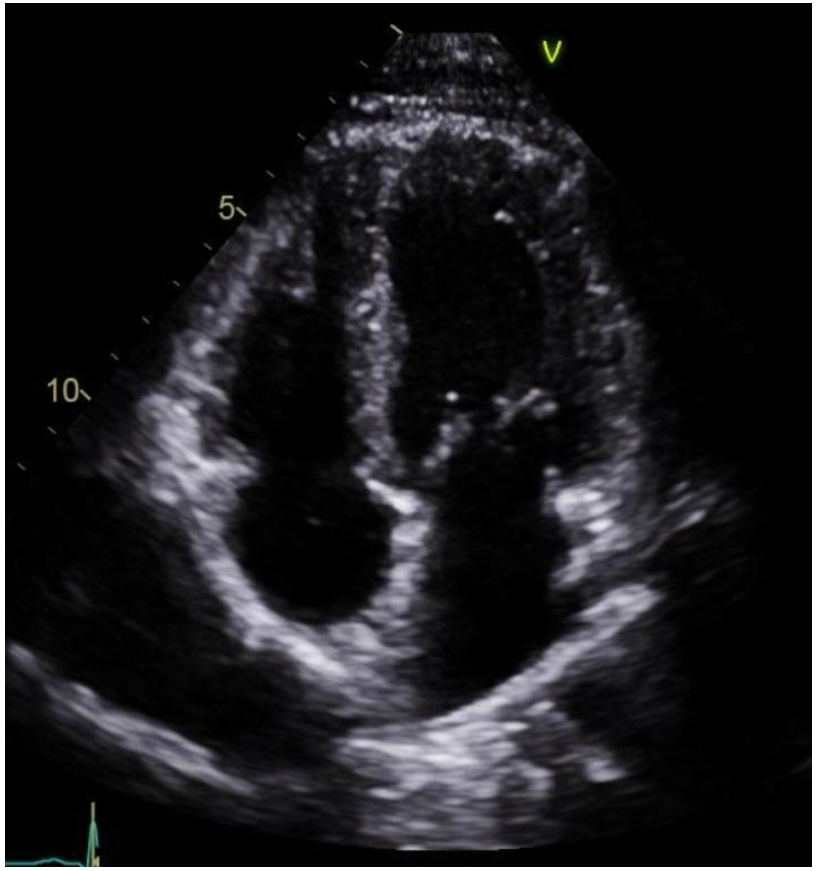
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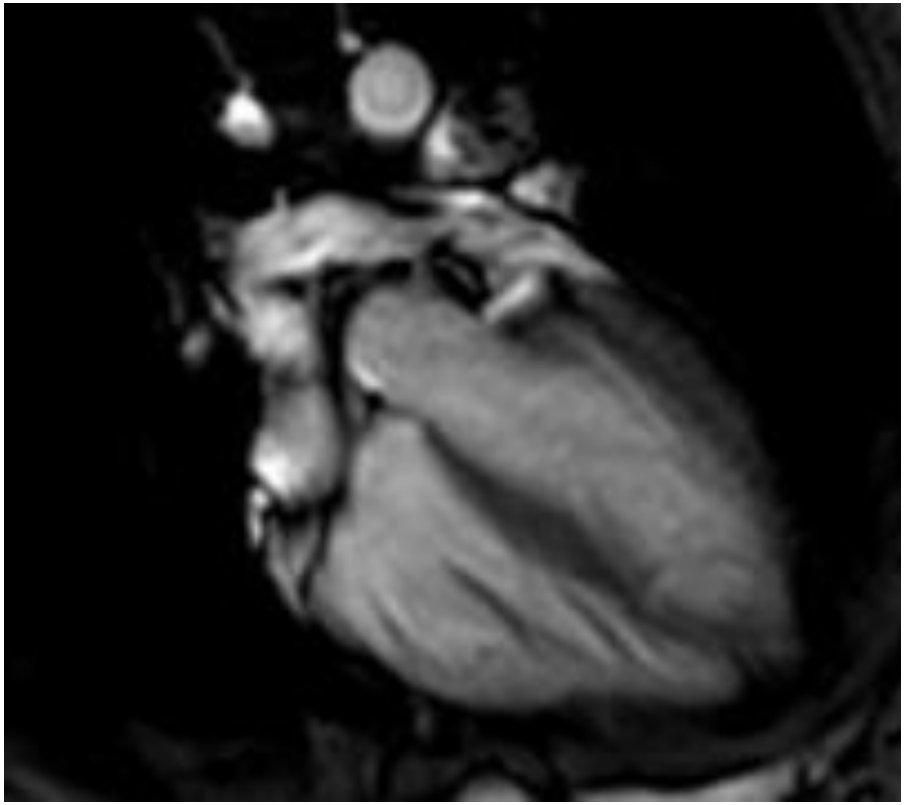
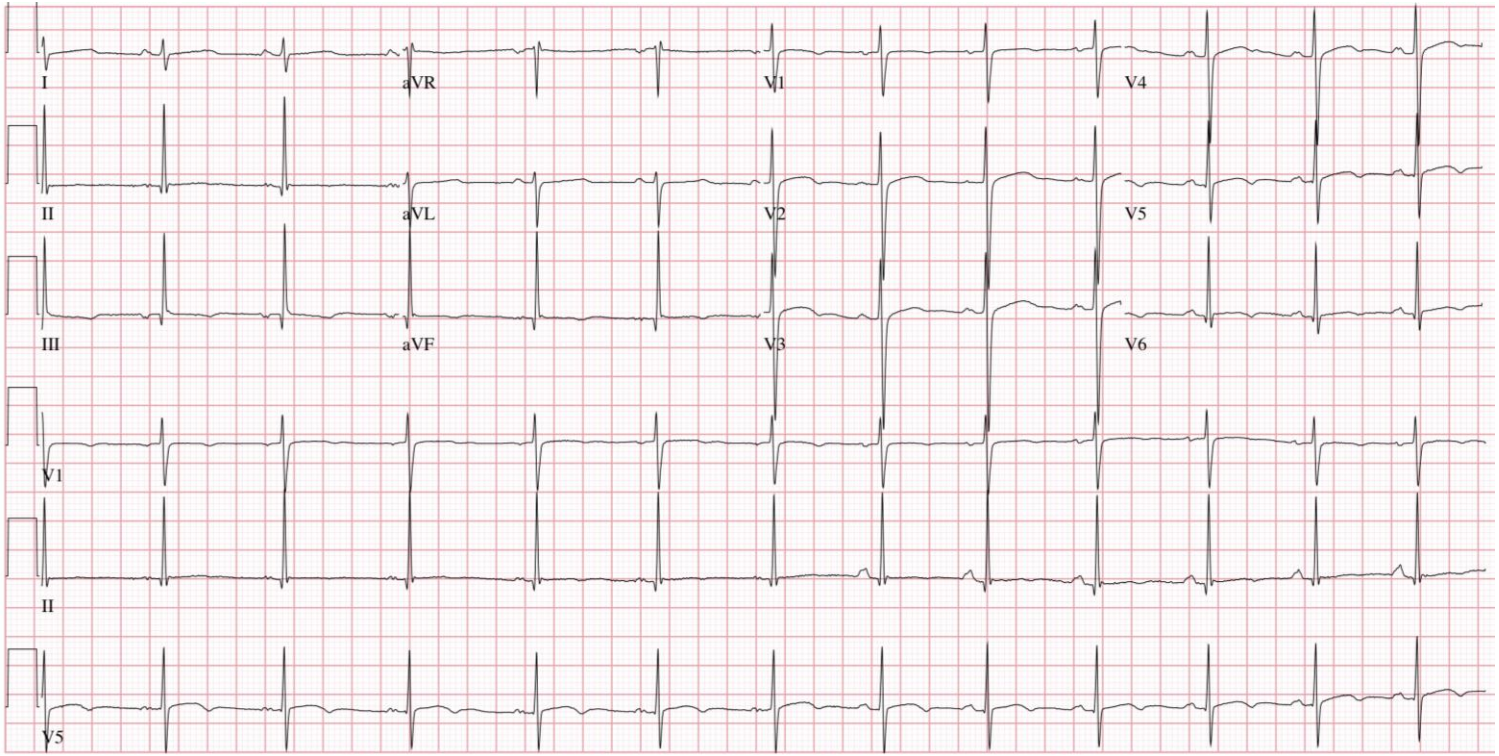


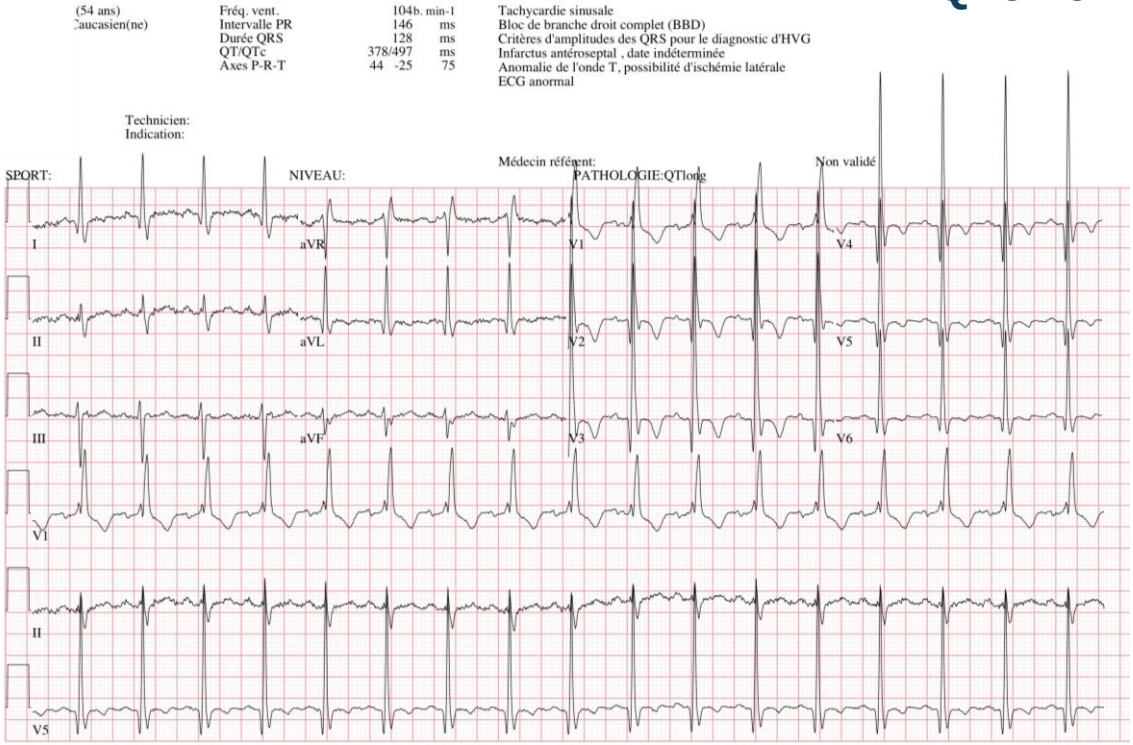
Soeur

QTc 447 ms



Père

QTc 497 ms



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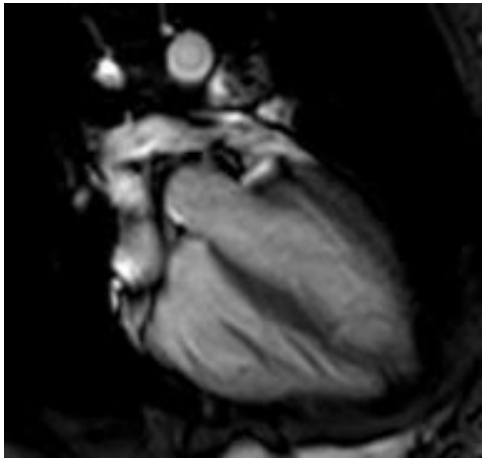
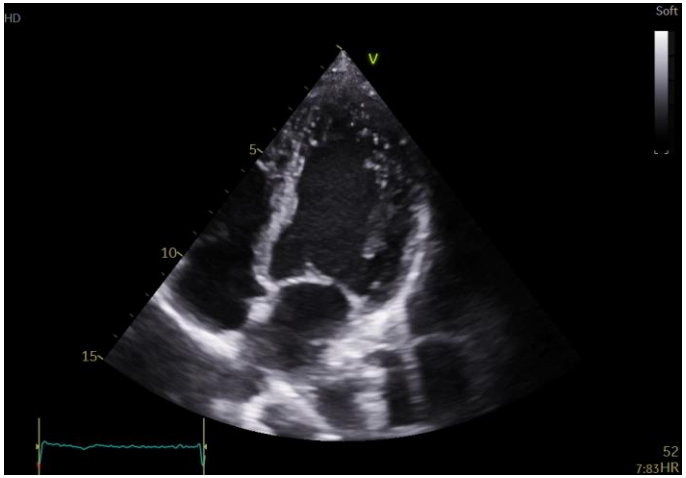
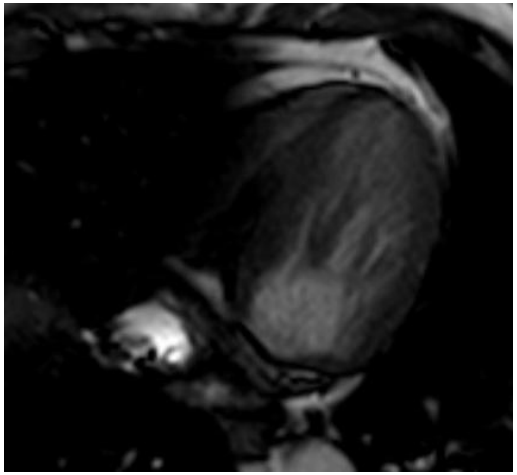
Cardiomyopathie structurelle



Cas index



Soeur



père

VG non compacté chez son père

(mutation génétique retrouvée chez les 3 dans **MYBPC3**)

CONCLUSIONS

Le diagnostic est parfois difficile - s'aider du bilan familial

RECOMMANDATION HRS 2024 beaucoup moins restrictives dans les canalopathies

Mais avec précautions importantes: DAE personnel !!

Cohérent avec la diminution générale des restrictions (ex. CMH)

Nécessité d'une stratification du risque ++

Participation du patient à la décision : DÉCISION PARTAGÉE

Être moins restrictifs pour les patients bien contrôlés sous traitement (?)