Congenital short QT syndrome: Main ECG/VCG features

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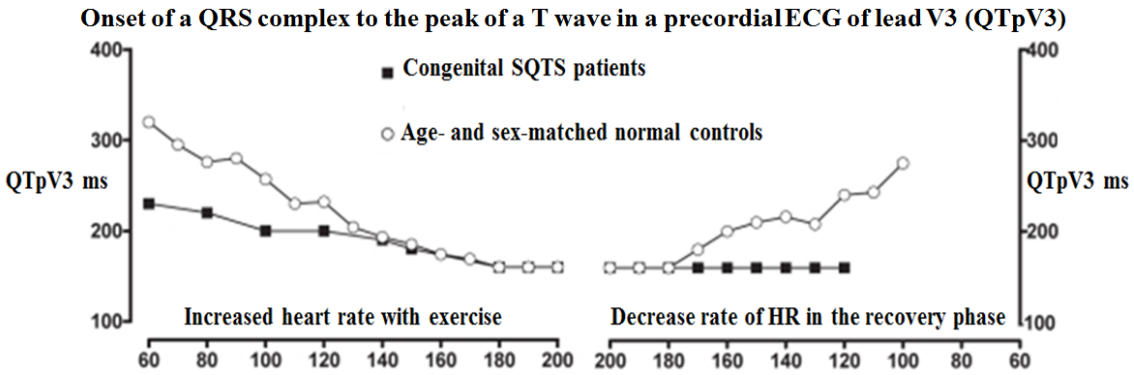
1) **Lack of normal changes in QT interval with HR variations**

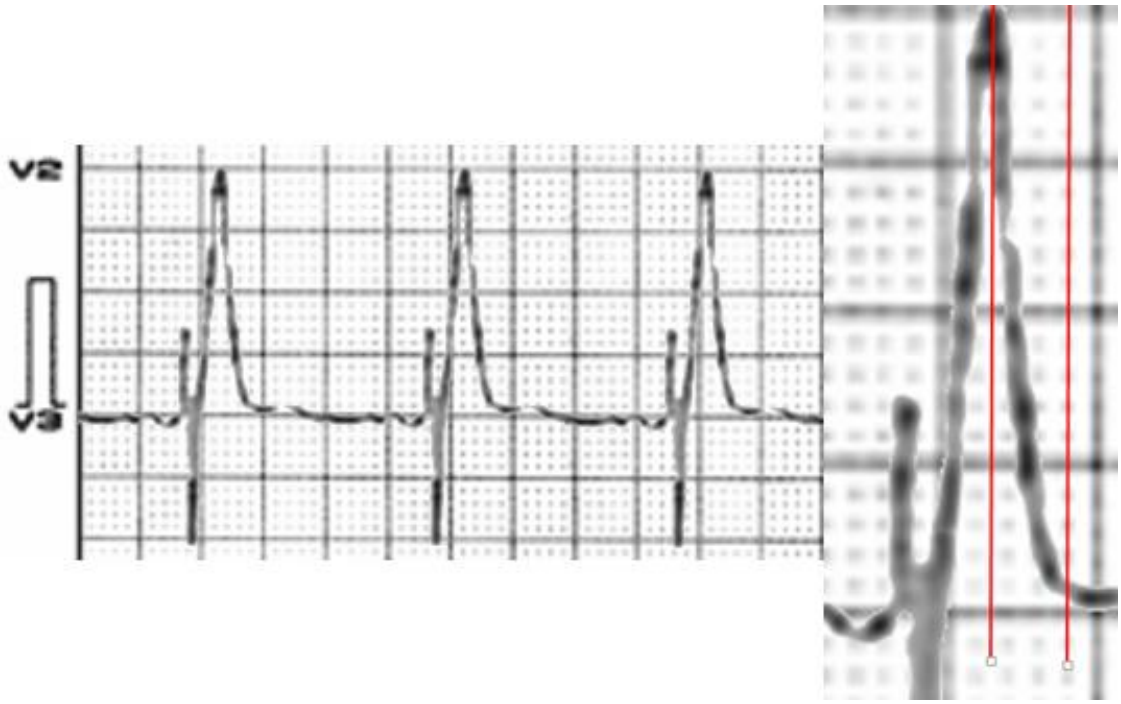
Fixed QT intervals which remain constant over a range of HRs. At fast HRs, the calculated QTc may appear normal (=”pseudonormal” QTc), however, as the HR slows, the QTc typically fails to prolong. serial strip of Holter monitoring at rest may be used to try and capture short QT intervals during periods of relative bradycardia (HR 60-80bpm). Finally, exercise testing demonstrates lack of adaptation of QT interval duration with different HRs. The QT interval, an index of ventricular repolarization, is heart rate (HR) dependent, in other words the QT interval shortens with exercise. Some of this shortening is due to an increase in HR, and some is due to other effects of exercise, probably mostly neuroendocrine effects. In normal hearts, two-thirds of exercise-induced QT interval shortening are due to an increase in HR, and one-third to other effects. Changes in plasma catecholamine levels on exercise are not closely related to changes in the QT interval on exercise (Davey 1999). Both exaggerated or lower rate dependence of repolarization are arrhythmogenic. Adaptation of the QT-interval to changes in HR reflects on the body-surface ECG the adaptation of action potential duration (APD) at the cellular level. The initial fast phase of APD adaptation has been shown to modulate the arrhythmia substrate. Whether the slow phase is potentially, proarrhythmic remains unclear. Patients with congenital SQTS have less variation of the QT interval in relation to the change in HR. Treadmill testing show a lack of adaptation of the QT interval, in congenital SQTS. Relative lack of adaptation of the QT interval (onset of a QRS complex to the peak of a T wave in a precordial ECG of lead V3) (QTpV3) to accelerated HR during exercise and lack of adaptation of the QT interval during decelerated HR in the recovery phase when compared with age- and sex-matched normal controls.



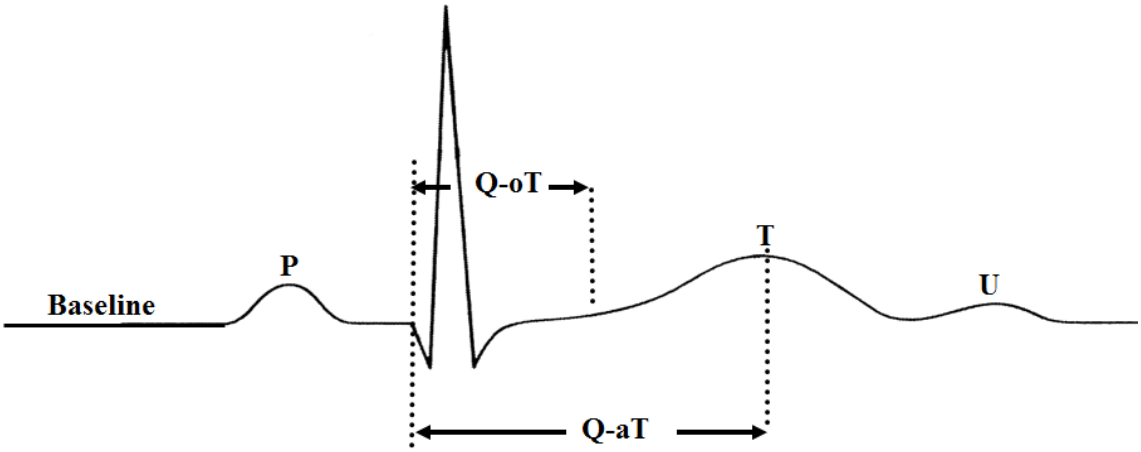
2) **ST segment usually absent or very short** (**Borggrefe M, Wolpert C, Antzelevitch C etal. Short QT syndrome. Genotype-phenotype correlations. *J Electrocardiol.* 2005; 38:75–80**).

This will result in the misinterpretation of the QTc interval with a faster HR and subsequently false-negative diagnosis of this possibly fatal syndrome. Holter monitoring can be helpful in this situation because it allows measurement of the QTc during a period of slower heart rate, such as sleep. Reduced rate-adaptation of the QT interval. The QT-RR relationship is less linear and its slope is less steep in SQTS patients as compared with control subjects. Quinidine restores the relationship toward control values. QRpV3 denotes the interval form the beginning of the QRS complex to the peak of T wave, measured in lead V3. Therefore, QTc corrected by any formula will fail to reflect the true QTc. At rapid rates, QTc will falsely approximate normal values leading to a false-negative diagnosis. This is particularly important for the diagnosis of SQTS in pediatric populations, where resting HR is >100 bpm. Sometimes, Holter monitoring shows impaired adjustment of QT interval with change in HR. Long-term ECG monitoring becomes necessary in such cases to make the correct diagnosis. The range of HRs is increased at baseline by using ambulatory electrocardiogram recordings in addition to those collected under semisupine, resting conditions (Garnett 2012). Quinidine is a Class IA antiarrhythmic drug –isomer of quinine found in the bark of the cinchona tree. The drug affects depolarization and repolarization by blocking Na^+ and K^+ channels respectively. Quinidine blocks the fast Na^+ current; I_{to1} channel or transient outward current, inward rectifier I_{K1}, delayed rectifier: I_{Ks}, I_{Kr} and I_{Kur}, I K_{ATP} or adenosine triphosphate ATP sensitive potassium channel, $IK_{\text{-Ach}}$, alpha 1 and alpha 2 adrenergic receptors: can cause orthostatic hypotension and reflex sinus tachycardia; M_2 muscarinic receptor. In the short QT syndrome, oral quinidine is effective in suppressing the gain of

function in I_{Kr} responsible for SQT1 variant with a mutation in HERG and thus restoring normal HR dependence of the QT interval and rendering VT/VF noninducible. Additionally, quinidine prolongs the QT interval into the normal range, restored the HR dependence of the QT interval toward a range of adaptation reported for normal subjects (**Wolpert C, Schimpf R, Veltmann C, Giustetto C, Gaita F, Borggrefe M. Clinical characteristics and treatment of short QT syndrome.Expert Rev Cardiovasc Ther. 2005 Jul;3(4):611-7. doi: 10.1586/14779072.3.4.611**.).

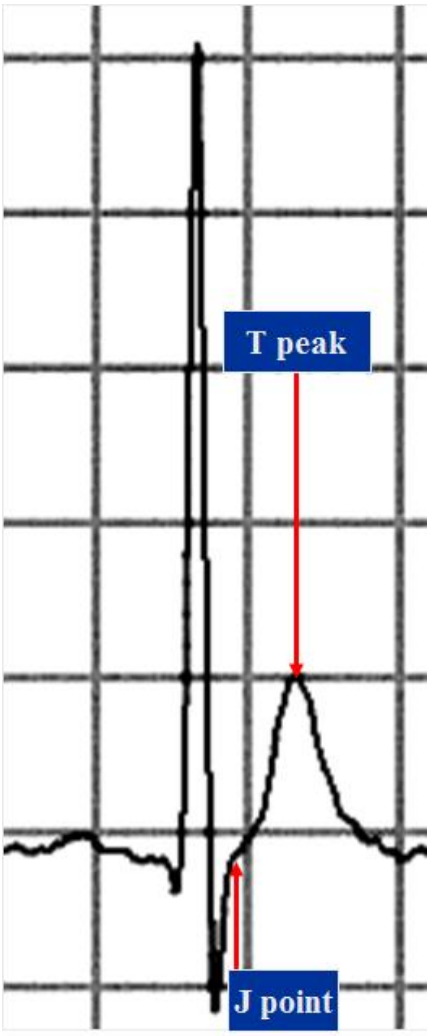


T-wave originates directly from QRS without an identifiable ST segment. Additionally, the distance between T-apex/T-end = 100 ms: transmural dispersion of repolarization. Short Q-oTc interval: interval that extends from the QRS onset up to the T wave onset, corrected by heart rate. Short Q-aTc interval: interval that extends from the QRS onset up to the T wave apex corrected by heart rate. Figure x



3) **Short J Point-T peak interval <120 ms.**

The J Point-T peak interval is the distance from the J point to the T peak. Values <120 ms are useful for the diagnosis of congenital SQTS (**Michael H Gollob 1, Calum J Redpath, Jason D Roberts The short QT syndrome: proposed diagnostic criteria J Am Coll Cardiol. 2011 Feb 15;57(7):802-12. doi: 10.1016/j.jacc.2010.09.048**.). Figure



J Point-T peak interval <120 ms: considered a criterion for diagnosis in the Gollob score; value = 1 point (Gollob 2011). The Short QT Syndrome diagnostic criteria is based on a point score system as follows:

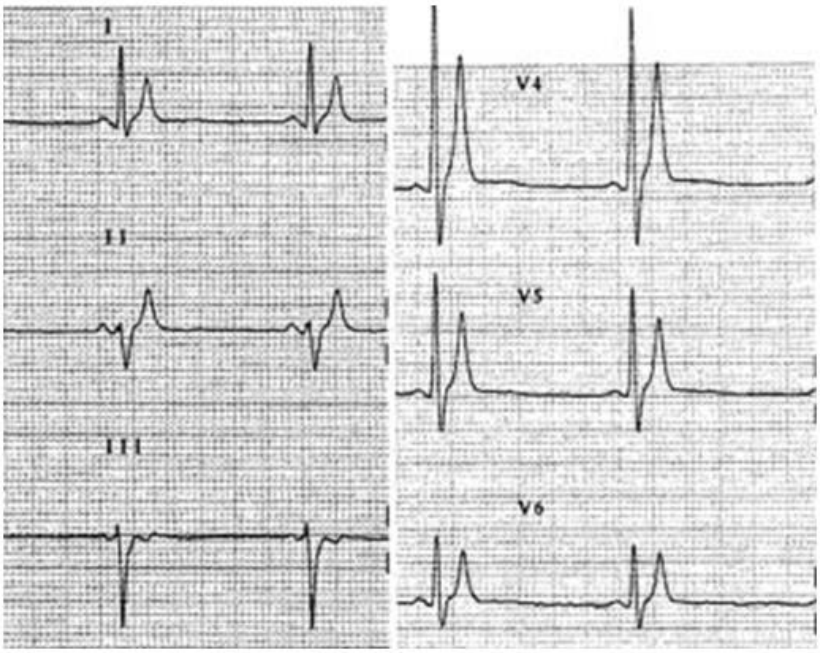
QTc in miliseconds	Score
< 370	1
< 350	2
< 330	3
J point – T peak interval	
< 120	1
Clinical history	
Sudden cardiac arrest	2
Polymorphic VT or VF	2
Unexplained syncope	1
Atrial fibrillation	1
Family history	
1st or 2nd degree relative with SQTS	2
1st or 2nd degree relative with sudden death	1
Sudden infant death syndrome	1
Genotype	
Genotype positive	2
Mutation of undetermined significance in a culprit gene	1

Interpretation: High-probability: ≥ 4 points; Intermediate probability: 3 points; Low probability: ≤ 2 points.

4) **Tall, peaked, symmetrical (pseudo symmetrical because the afferent limb of T-loop shows comets closer together in relation to efferent limb) and narrow-based T-waves** very similar to moderate hypokalemia (**Pérez Riera AR, Ferreira C, Dubner SJ, Schapachnik E, Soares JD, Francis J. Brief review of the recently described short QT syndrome and other cardiac channelopathies.Ann Noninvasive Electrocardiol. 2005 Jul;10(3):371-7. doi: 10.1111/j.1542-474X.2005.00632.**x.) (**Andrés Ricardo Pérez Riera 1, Adail Paixão-Almeida, Raimundo Barbosa-Barros, Frank G Yanowitz, Adrian Baranchuk, Sergio Dubner, Antônio Carlos Palandri Chagas. Congenital short QT syndrome: landmarks of the newest arrhythmogenic cardiac channelopathy. Cardiol J. 2013;20(5):464-71. doi: 10.5603/CJ.a2013.0052**.).

Difference of T waves morphologies in the potassium congenital SQTS variants

SQT1: The T waves in the precordial leads appear often tall, narrow and symmetrical, with a relatively prolonged Tpeak-Tend interval (**Gaita F, Giustetto C, Bianchi F, Wolpert C, Schimpf R, Riccardi R, Grossi S, Richiardi E, Borggrefe M. Short QT Syndrome: a familial cause of sudden death.Circulation. 2003 Aug 26;108(8):965-70. doi: 10.1161/01.CIR.0000085071.28695.C4**.).



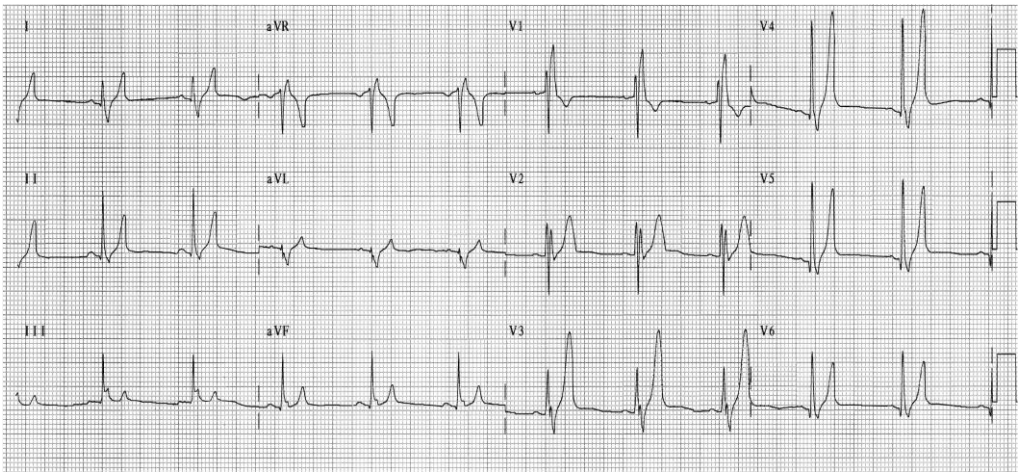
Twelve-lead ECG showing typical SQT1 features: tall, narrow and peaked T waves, QT 280 ms. Reproduced with permission from Gaita ((**Gaita F, 2003 Aug 26;108(8):965-70. doi: 10.1161/01.CIR.0000085071. 28695.C4**.).).

SQT2: The T waves appear to be symmetrical, but not as tall and narrow (Bellocq 2004; Hong 2005).

SQT3: Asymmetrical T waves with a rather normal ascending ramp and a remarkable rapid descending terminal ramp (**Priori SG, Pandit SV, Rivolta I, Berenfeld O, Ronchetti E, Dhamoon A, Napolitano C, Anumonwo J, di Barletta MR, Gudapakkam S, Bosi G, Stramba-Badiale M, Jalife J. A novel form of short QT syndrome (SQT3) is caused by a mutation in the KCNJ2 gene. Circ Res. 2005 Apr 15;96(7):800-7. doi: 10.1161/01.RES.0000162101.76263.8c**.).

Peaked and symmetrical, narrow-based T waves ("tent-shaped") are characteristic of mild hyperpotasssemia. Usually, this is the earliest sign of hyperkalemia. This morphological T waves are observed with slightly increased serum potassium levels (potassium level >5.5 mEq/L and <6.0 mEq/L). It is present only in 22% of the cases of hyperkalemia. Not too sensitive but quite specific. Similar T waves are registered eventually in congenital SQTS. Additionally, peaked and symmetrical with narrow-based T waves ("tent-shaped") are observed also in metabolic acidosis and without hyperkalemia (**Dreyfuss D, Jondeau G, Couturier R, Rahmani J, Assayag P, Coste F. Tall T waves during metabolic acidosis without hyperkalemia: a prospective study. Crit Care Med. 1989 May;17(5):404-8. doi: 10.1097/00003246-198905000-00005**.).Peaked, symmetrical T waves, with a broad base is an early sign of hyperacute phase of myocardial infarction (**Primeau R. Peaked, symmetrical T waves, an early sign of myocardial infarct].Union Med Can. 1969 Jan;98(1):104-5**.). Figure x

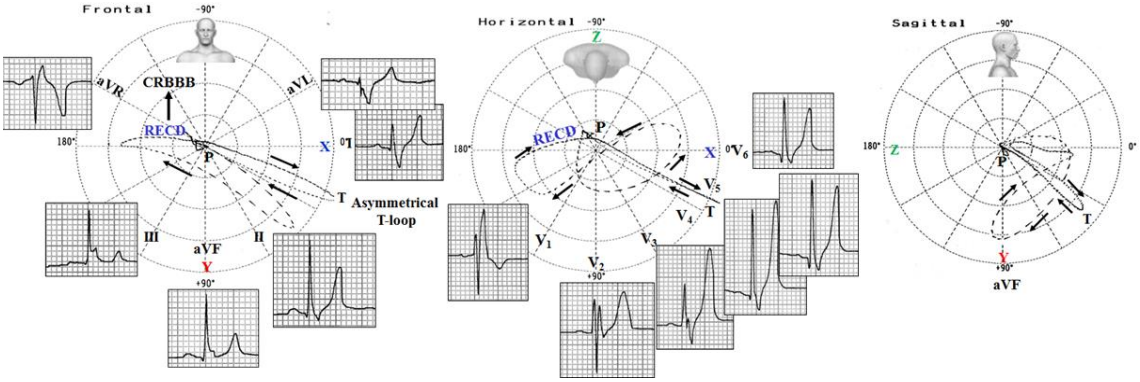
Name: JSVB; **Age**: 27; **Sex**: Male; **Race**: White; **Weight**: 67 Kg. **Height**: 1.72m. **Date**: 06/24/2004; **Medication in use**: none.



Rhythm: sinus; HR: 65 bpm; P wave: SAP axis: +54º in the FP and to the front in the HP; duration: 80 ms; voltage: 1 mm; PR interval: 134 ms; QRS: SÂQRS: +106º in the FP and to the front in the HP; QRS duration (QRSD): 120 ms; QRS morphology: triphasic rSR' pattern in V1 and broad S wave in left leads I, aVL V5 and V6 (right terminal forces); intrinsic deflection in V1 >50 ms.

T wave morphology: tall T wave from V3 through V5 with narrow base and a tendency to be symmetrical (the patient does not have serum potassium increase); SAT: +42º in the FP and discretely heading to the front and below in the HP; QT/QTc interval: 302/315: short for this rate (the inferior limit for a 67 bpm heart rate in men is 324 ms (**Sagie A, Larson MG, Goldberg RJ, Bengtson JR, Levy D.An improved method for adjusting the QT interval for heart rate (the Framingham Heart Study).Am J Cardiol. 1992 Sep 15;70(7):797-801. doi: 10.1016/0002-9149(92)90562-d**.).

ECG/VCG correlation



FP - QRS loop duration 120 ms. Right End Conduction Delay (RECD) located on the top right quadrant near the aVR lead. Asymmetrical T-loop SÂT +20º.

HP – Triphasic QRS pattern in V1-V2 and broad final S-wave in V5-V6: CRBBB. VCG Grishman-type of CRBBB: afferent loop behind the X line; triphasic rSR' pattern; short QT interval; tall T waves with narrow base from V_3 through V_5.

RSP – Asymmetrical T loop heading down and to the front.

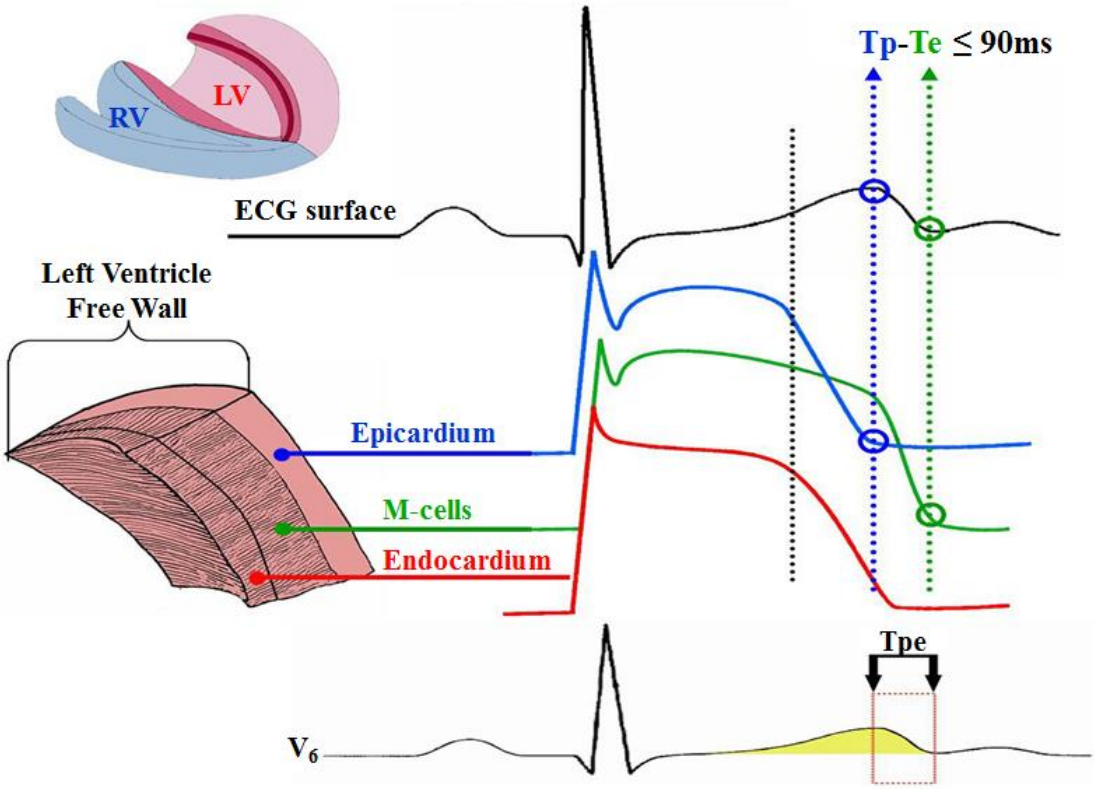
Note: The VCG is conclusive that the T wave is not symmetrical because the efferent limb has tears (or commets) very close to one another; on the other hand, the afferent limb has tears more separated from each other.

5) **Extremely short QT/QTc interval** QT < 320ms and a QTc < 340ms. SQTS genotypes 1-3 all had QTc intervals < 300-320 ms. Viskin suggested the following approach:QTc intervals < 330 ms in males or < 340 ms in females should be considered diagnostic of SQTS, QTc intervals < 360 ms in males or < 370 ms in females should only be considered diagnostic of SQTS when supported by symptoms and/ or positive family history.

6) **Frequent prominent U wave.** U waves are described as prominent if they are. >1-2mm or 25% of the height of the T wave. Maximum normal amplitude of the U wave is 1-2 mm. The normal voltage of the U wave is < 25% of the T-wave voltage. Prominent U waves most commonly found with bradycardia, severe hypokalemia, hypocalcaemia, hypomagnesaemia, hypothermia, raised intracranial pressure, LVH, HCM, drugs associated: digoxin effect,

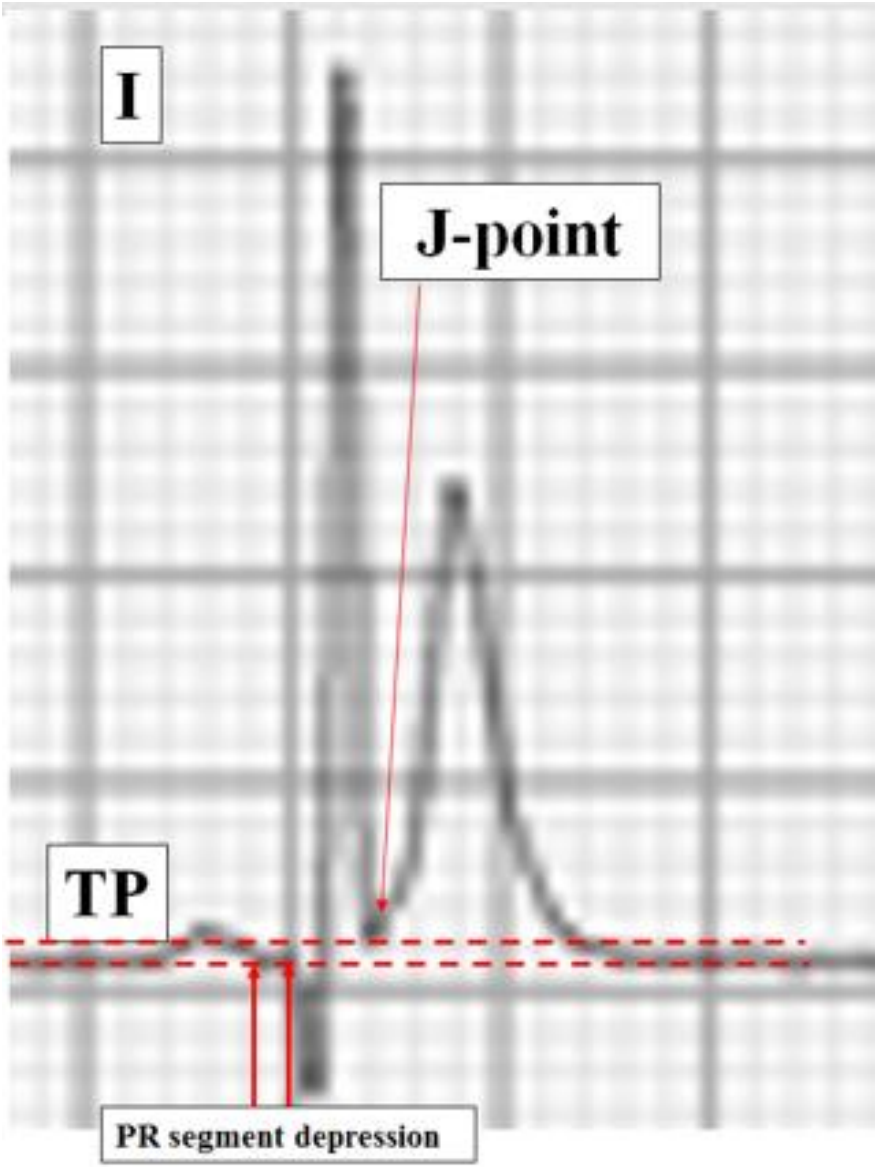
phenothiazines (thioridazine), class Ia antiarrhythmics (quinidine, procainamide), class III antiarrhythmics (sotalol), amiodarone, long QT interval, SQTS. (**Schimpf R, Antzelevitch C, Haghi D et al. Electromechanical coupling in patients with the short QT syndrome: further insights into the mechanoelectrical hypothesis of the U wave. *Heart Rhythm.* 2008;5:241–5**.) (**Pérez Riera AR, Ferreira C, Filho CF, Ferreira M, Meneghini A, Uchida AH, Schapachnik E, Dubner S, Zhang L The enigmatic sixth wave of the electrocardiogram: the U wave. Cardiol J. 2008;15(5):408-21**.).

7) **Eventual prolonged Tpeak-Tend interval indicative of augmented transmural dispersion of refractoriness.** (**Extramiana F, Antzelevitch C. Amplified transmural dispersion of repolarization as the basis for arrhythmogenesis in a canine ventricular-wedge model of short-QT syndrome. *Circulation.* 2004; 110:3661–6**.) The possible substrate for the development of ventricular tachyarrhythmias may be a significant transmural dispersion of the repolarization due to a heterogeneous abbreviation of the action potential duration. Normally T peak/Tend interval is 94 ms in men and 92 in women when measured in the V5 lead. In SQTS this parameter is prolonged >92 ms in women and >94 ms in men with the measurement in V5. In SQT1 patients the T waves in the precordial leads, appear often tall, narrow and symmetrical, with a relatively prolonged Tpeak-Tend interval (**Gaita F, Giustetto C, Bianchi F, Wolpert C, Schimpf R, Riccardi R, Grossi S, Richiardi E, Borggrefe M. Short QT Syndrome: a familial cause of sudden death.Circulation. 2003 Aug 26;108(8):965-70. doi: 10.1161/01.CIR.0000085071. 28695.C4**.).

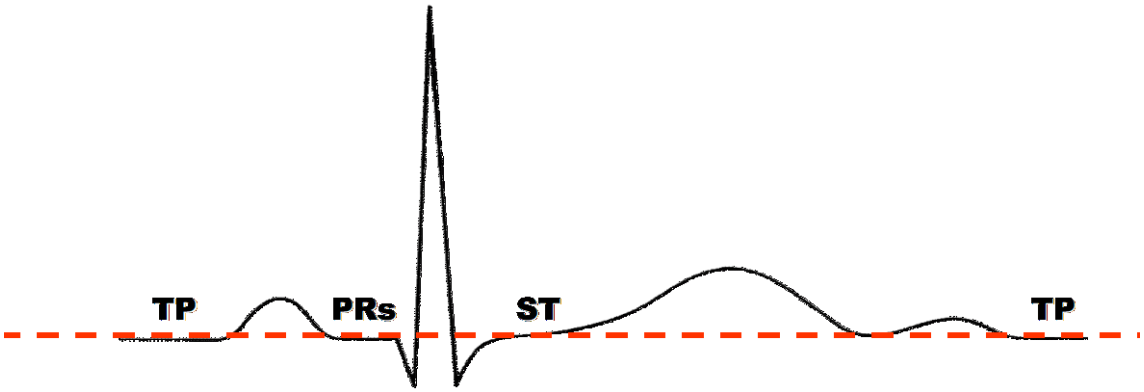


8) **Depression of the PR/PQ segment (PQD)**

It is consequence to a heterogeneous abbreviation of atrial repolarization, most prominently in inferior and anterior leads. Digitalized 12-lead ECGs of SQTS patients were evaluated for PR depression in all leads and for QT intervals in leads II and V5. PR depression was defined as 0.05 mV (0.5 mm) depression from the isoelectric line (TP segment of precedent beat). The study shows that 81% patients with SQTS reveal PQD. As PQD is rarely observed in healthy individuals, this ECG feature may constitute a novel marker for SQTS in addition to a very short QT interval (**Tulumen E, Giustetto C, Wolpert CC et al. PQ segment depression in short QT syndrome patients: A novel marker for diagnosing short QT syndrome? *Heart Rhythm.* 2014; 11:1024–30**.). The PQ or PR segment is the flat, usually isoelectric segment between the end of the P wave and the start of the QRS complex. Figure x



Regarding the level of PRs, in normal conditions is at the same level as ST segment (isoelectric) and TP segment of the precedent beat.



Usually, PR segment (end of P wave up to QRS complex onset), ST segment (from J point or the end of QRS up to the beginning of the T wave) and TP segment (from the end of the T wave up to the P wave of the following cycle) are at the same level. The figure shows a normal ECG and a line of dots pointing out the level of the three segments: PR, ST and TP.

PR (PRs) or PQ segment: it stretches from the end of P wave to the onset of QRS complex. The PR segment is leveled when it is at the same level of the PR segment of the beat being studied. If the PR segment falls below the baseline (TP segment of precedent beat), then it is said to be depressed.

ST segment: it stretches from the J point (union of ST with the end of QRS complex) up to the onset of the T wave.

TP segment: it stretches from the end of T wave to the onset of the P wave of the next cycle. TP segment is between the end of the T wave and the beginning of the next P wave. It is the true isoelectric interval in the electrocardiogram. In other words, the PR segment changes are relative to the baseline formed by the precedent TP segment of anterior beat.

Acquired clinical causes of PR segment depression

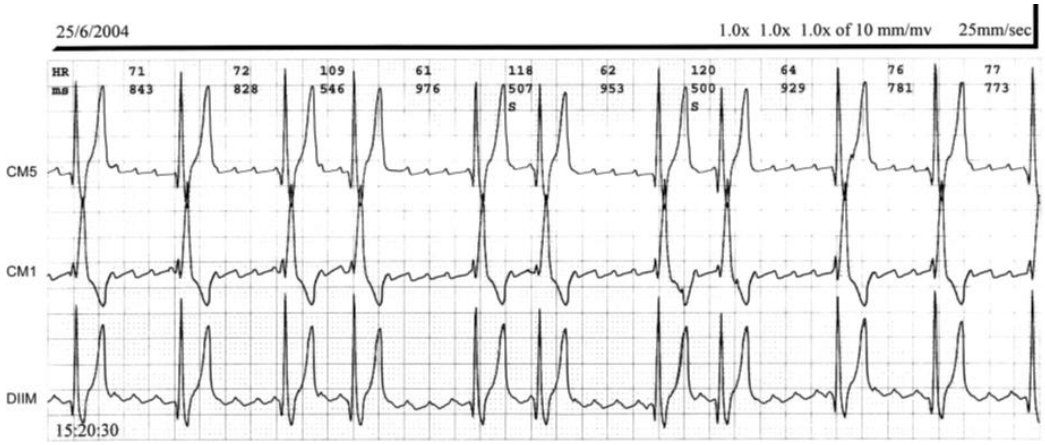
Acquired causes of PR segment depression are acute pericarditis and in acute myocardial infarction with concomitant atrial ischemia or atrial infarction which is indicative of poor outcomes consequence of development of atrioventricular block, supraventricular arrhythmias and cardiac free-wall rupture.

- Atrial fibrillation
- Acute inferior myocardial infarction (poor prognosis when present). PR segment depression or elevation in patient with acute myocardial infarction scenario indicates concomitant atrial ischemia or infarction. The ST segment indicates infarction in the ventricle, the PR segment indicates infarction in the atria. Diagnostic criteria for atrial infarction are: PR segment elevation >0.5 mm in V5 and V6 with reciprocal depression in V1 and V2, PR segment elevation >0.5 mm in I and depression in II and III, >1.5 mm PR segment depression in precordial leads and >1.2 mm PR segment depression in I, II or III in combination with atrial arrhythmias (Liu 1961). PR segment depression ≥1.2 mm in inferior leads was found in 1.9% of patients with acute inferior MI. This ECG sign represents a subgroup of patients with extensive atrial ischemia. This sign is associated with a high frequency of atrioventricular block, supraventricular arrhythmias and cardiac rupture, with high in-hospital mortality (**Jim MH, Siu CW, Chan AO, Chan RH, Lee SW, Lau CP. Prognostic implications of PR-segment depression in inferior leads in acute inferior myocardial infarction. Clin Cardiol. 2006 Aug;29(8):363-8. doi: 10.1002/clc.4960290809**.).
- PR segment depression in III and concomitant PR segment elevation in I and aVL are indicative of infarction on the posterior wall of the right atrium (**Radojevic N, Savic S, Aleksic V, Cukic D. Unusual case of right atrial reinfarction.J Forensic Leg Med. 2012 Feb;19(2):105-8. doi: 10.1016/j.jflm.2011.10.003. E**).
- Acute pericarditis: In these cases, there is also some concave-to-the-top ST segment elevation
- Acute myopericarditis: it occurs in the presence of diffuse inflammation of the pericardial sac and superficial epicardium from a multitude of infectious and inflammatory processes. This inflammation results in a current of myocardial injury resulting from the epicardial irritation manifested by a number of ECG findings: ST segment elevation with PR segment depression, normalization of the ST segment abnormality with T wave inversion, and eventual normalization of the ECG over a period of days to several weeks. In myopericarditis, the most common location for PR depression was lead II (55.9%), while this ECG finding least likely appeared in lead aVL (2.9%). PR depression in any lead had a high sensitivity (88.2%), but low specificity (78.3%) for myopericarditis. The combination of PR depressions in both precordial and limb leads had the most

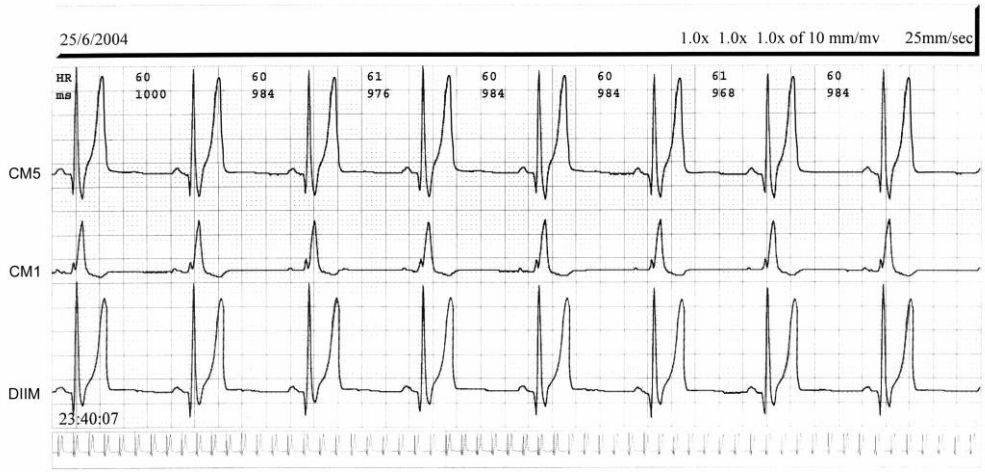
favorable predictive power to differentiate myopericarditis from STEMI (positive 96.7% and negative power 90%) (**Porela P, Kytö V, Nikus K, Eskola M, Airaksinen KE. PR depression is useful in the differential diagnosis of myopericarditis and ST elevation myocardial infarction. Ann Noninvasive Electrocardiol. 2012 Apr;17(2):141-5. doi: 10.1111/j.1542-474X.2012.00489.x**.).

- Extensive atrial impairment.
- During ablation of the left upper pulmonary vein in patient with atrial tumors.
- Cardiac tamponade: fluid collects in the pericardium. Because the pericardium is stiff, the heart is compressed, resulting in filling difficulties. Tamponade can be the result of pericarditis or myocarditis. After a myocardial infarction a tamponade can also develop; this is called Dresslers' syndrome. In case of cancer, increased pericardial fluid may develop. This is usually caused by carcinomatous pericarditis, meaning that the cancer has spread to the pericardium The ECG shows: sinus tachycardia, low QRS voltage, electrical alternans and PR segment depression.

9) **Familial arrhythmogenic disease associated with paroxysmal Familial Atrial Fibrillation (FAF)** (that terminates spontaneously or with intervention within 7 days of onset. Episodes may recur with variable frequency.) FAF and VF, syncope, and SCD (**Deo M, Ruan Y, Pandit SV, Shah K, Berenfeld O, Blaufox A, Cerrone M, Noujaim SF, Denegri M, Jalife J, Priori SG. KCNJ2 mutation in short QT syndrome 3 results in atrial fibrillation and ventricular proarrhythmia**. **Proc Natl Acad Sci U S A. 2013 Mar 12;110(11):4291-6. doi: 10.1073/pnas.1218154110**.) The SQTS is a channelopathy with remarkably abbreviated atrial an ventricular repolarization and a predisposition to supraventricular (Paroxysmal FAF) and ventricular arrhythmias in the absence of detectable SHD(***Patel C, Yan GX, Antzelevitch C* Short QT syndrome: from bench to bedside.*Circ Arrhythm Electrophysiol. 2010 Aug; 3(4):401-8***.). *Figure*



In this tracing we can see a short period of gross atrial fibrillation. The patient described palpitations. Congenital short QT syndrome is associated to a high incidence of paroxysmal atrial fibrillation, the electrophysiological mechanism of which would be caused by very short action potential with heterogeneous shortening of the cardiac potential and refractory period of atrial cardiomyocytes.



Sinus rhythm, tall/peaked, narrow-based T waves or pseudo symmetrical T-wave in a patient with SQTS. Approximately 8 hours later during the same test, the patient spontaneously reversed into sinus rhythm.

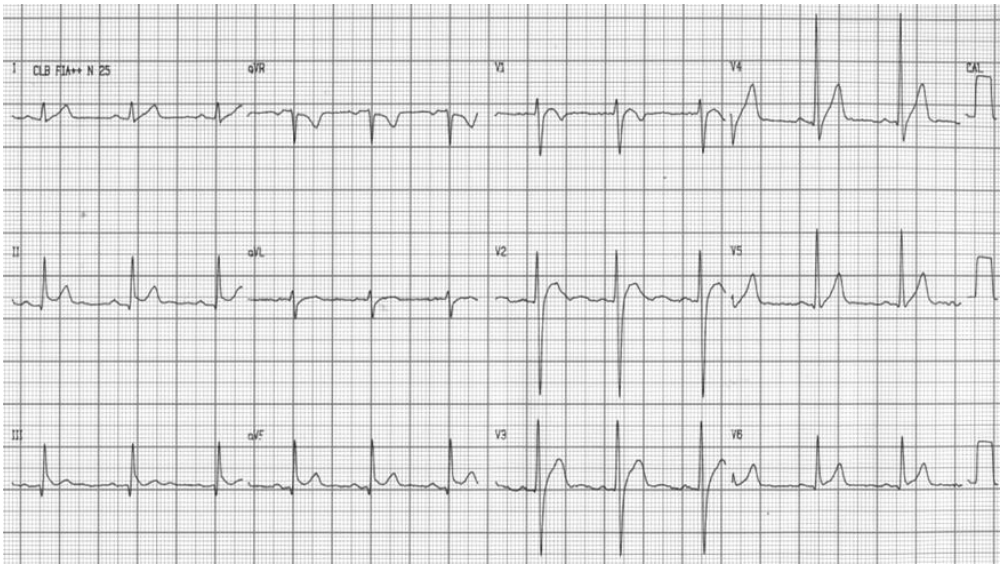
Villafañe et al, present a patient with congenital SQTS with AF and a slow ventricular response. Medical therapy has not been effective in maintaining sinus rhythm. The long-term outcome remains unknown for these children. This condition may present in utero as persistent bradycardia with postnatal ECG showing a very short QT interval (**Villafañe J, Fischbach P,**

Gebauer R. Short QT syndrome manifesting with neonatal atrial fibrillation and bradycardia. Cardiology. 2014;128(3):236-40. doi: 10.1159/000360758.).

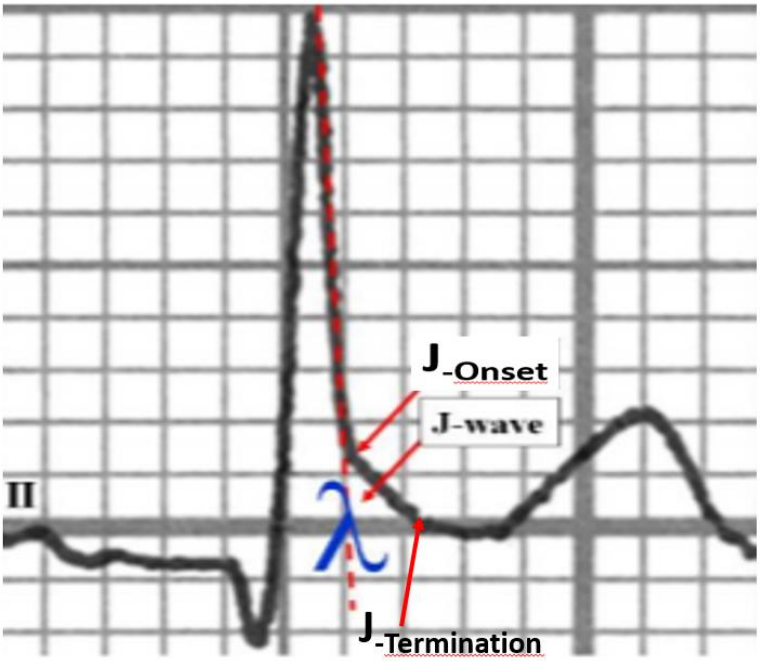
10) **High prevalence of early repolarization. It is associated with arrhythmic events**

There is a high prevalence of early repolarization in patients with congenital SQTS. Additionally, early repolarization may be useful in identifying the risk of cardiac events in SQTS High prevalence of early repolarization in SQTS. (**Watanabe H, Makiyama T, Koyama T, Kannankeril PJ, Seto S, Okamura K, Oda H, Itoh H, Okada M, Tanabe N, Yagihara N, Kamakura S, Horie M, Aizawa Y, Shimizu W. High prevalence of early repolarization in short QT syndrome. Heart Rhythm. 2010 May;7(5):647-52. doi: 10.1016/j.hrthm.2010.01.012**.)

Figure x Example of SQTS associated with early repolarization



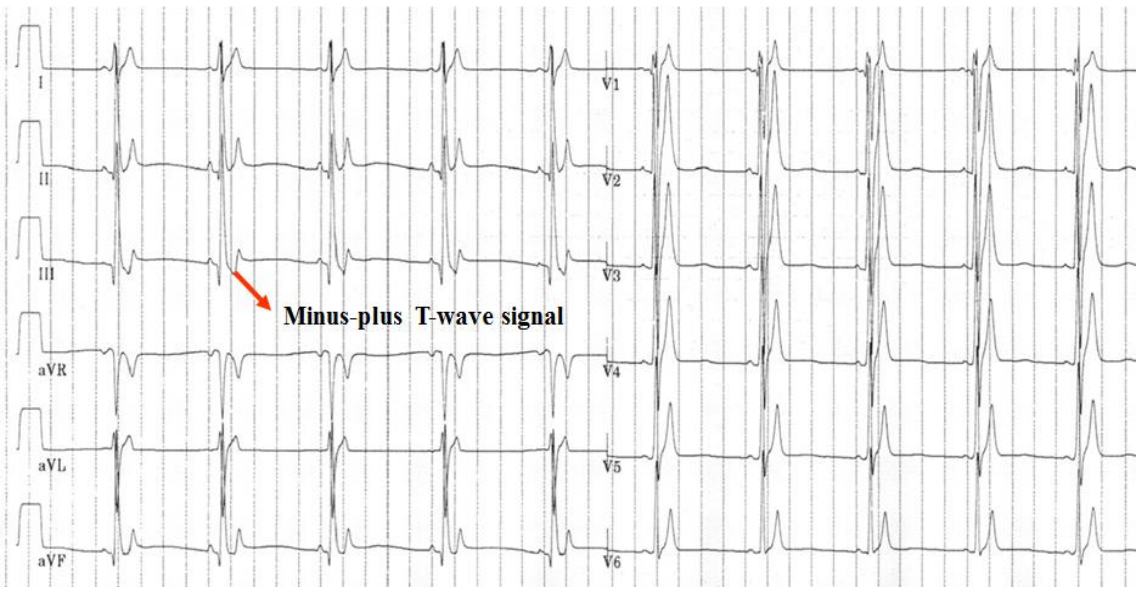
HR = 68 bpm; QT = 340 ms; RR = 880 ms; QTc = 362 ms



The first point of inflection of the R wave descendent ramp is considered the real J point. In these cases, the "tangent line" method is ideal. ST-segment elevation = 0.8 mm. We considered it an atypical C type variant of early repolarization pattern. The lambda aspect is a marker of fatal arrhythmias.

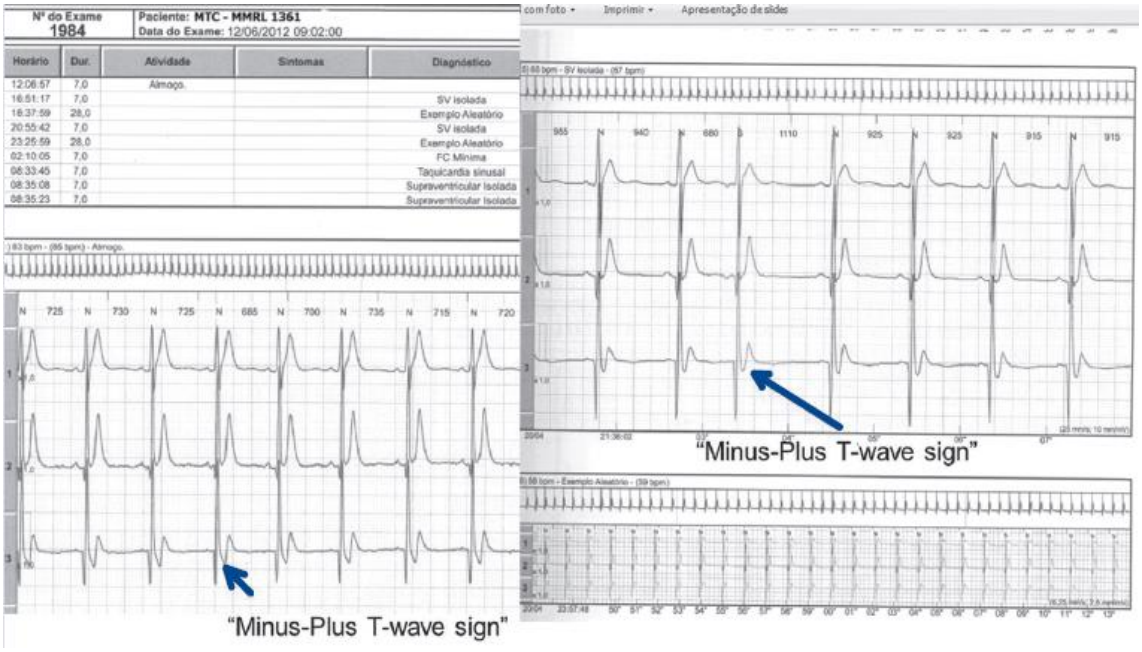
11) **Minus-plus T wave signal (see next ECG and Holter)**

This is and hallmark of congenital SQTS observed for our group a long time ago



The minus-plus T wave signal or negative-positive T wave without ST segment observed in III in a patient with SQT1 variant **(Schimpf R, Wolpert C, Gaita F,**

Giustetto C, Borggrefe M. Short QT syndrome. Cardiovasc Res. 2005 Aug 15;67(3):357-66. doi: 10.1016/j.cardiores.2005.03.026.)

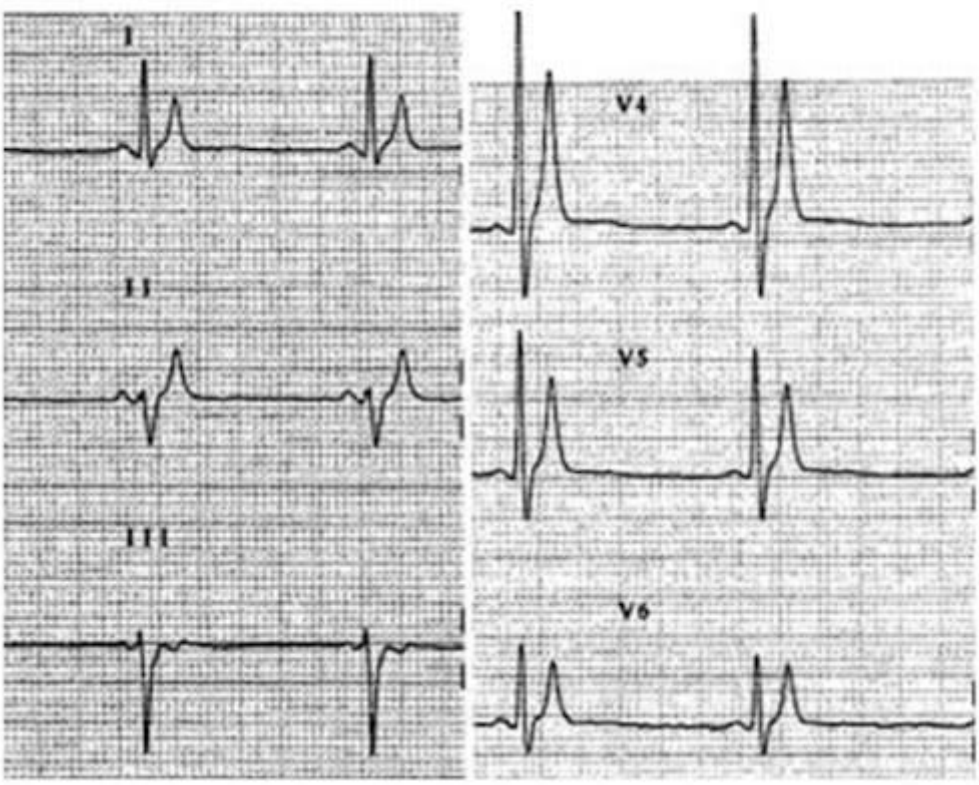


Brazilian patient with SQTS Holter monitoring. The “minus-plus T wave sign” observed in a Holter recording in a patient from Latin America (Brazil).

“Minus-plus T wave signal”: The initial part of the T wave is recorded immediately after the QRS complex (absence or minimal ST segment) shows a negative initial polarity in some leads (arrow) that we denominated "minus-plus" T wave signal (negative-positive). Coincidentally, the CAV3 mutation that causes gain-of-function of late I_{to} without affecting other cardiac ion channels corresponds to J point and the initial negative portion of the T wave on the surface ECG. There would be a genetypic/phenotypic relationship? Unpublished yet

12) **Electrophysiological studies(EPSs) in SQTS demonstrate: Extremely short atrial and ventricular refractory periods,** high rates of inducible AFl and VF and marked vulnerability to mechanical induction of VF. The role of EPSs in diagnosing and risk-stratifying patients with SQTS has not yet been established

HR is significantly slower in patients with SQTI than in those without, QT duration: 260-280ms consequence of abbreviates cardiac APs, which manifests on ECG as a very short QT interval, frequent early repolarization, tall T wave, and U wave presence, QT dispersion, FAF (atrial arrhythmias rate 30%) and ventricular arrhythmia (VA)/SCA are significantly more frequent in patients with SQTI than in those without. SQTI was significantly associated with FAF and VA/SCA. **Dae-Young Kim et al. Long-term prognosis of short QT interval in Korean patients: a multicenter retrospective cohort study BMC Cardiovasc Disord . 2021 Jan 6;21(1):17. doi: 10.1186/s12872-020-01824-3.** AF is common in patients with SQTS and in 50% of all cases it is the first clinical manifestation of the syndrome. (**Giustetto C, Di Monte F, Wolpert C et al. Short QT syndrome: clinical findings and diagnostic–therapeutic implications. *Eur Heart J.* 2006; 27:2440–7. doi: 10.1093/eurheartj/ehl185**. The physio-pathological mechanism responsible in SQTS for both atrial and ventricular arrhythmias is shortening of the refractory period due to a GOF of potassium channels. (***Bellocq C, van Ginneken AC, Bezzina CR, Alders M, Escande D, Mannens MM, Baró I, Wilde AA*** **Mutation in the KCNQ1 gene leading to the short QT-interval syndrome.*Circulation. 2004 May 25; 109(20):2394-7*.);**



Very short QT interval, minimal (or absent) ST segments, peaked tall near symmetric T waves and in two patients with SQTS 1 variant.