

## Congenital QT Syndrome Causing Syncope in Children, A Case to be proposed

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### Abstract

### Case Report

Congenital long QT syndrome (SQTL) is a group of genetically heterogeneous heart diseases characterized by repolarization disorders secondary to QT prolongation, leading to an increased risk of ventricular arrhythmia. The genes involved in these mutations encode for subunits of the ion channels responsible for cardiac electrical activity. SQTL should be evoked during the onset of syncope or sudden death in young subjects in the context of adrenergic stimulation. There is often a delay in diagnosis in this age group, because the symptoms are, wrongly, attributed to an epilepsy attack. The diagnosis is based on the electrocardiogram, a family survey and a genetic study. The treatment is based on beta-blockers and the implantable defibrillator. We report through an observation of a 3 years old girl who presented loss of consciousness following exertion. We will discuss her diagnosis and treatment.

**Keywords:** Congenital long QT syndrome (SQTL), heart diseases, syncope.

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## INTRODUCTION

Long QT syndrome (SQTL) is a possible factor in sudden death. It is characterized by an abnormality of ventricular repolarization on the surface electrocardiogram (ECG) (elongated QTc  $\geq 440$  ms) associated with a significant risk of syncope or sudden death due to ventricular arrhythmias of the torsade de pointes type or ventricular fibrillation.

We report the case of a girl who presented after an effort a faintness with loss of consciousness. We discuss the diagnostic means and the methods of treatment.

The interest of this observation is to insist on the need to perform an electrocardiogram for each child admitted to pediatrics or seen in consultation for ill-labeled discomfort with loss of consciousness.

## OBSERVATION

A little girl of consanguineous parents, born by caesarean section at 30 weeks with amenorrhea (AS). Her family's history is marked by sudden death in her older brother at the age of 3 months. She was with sensorineural hearing loss for which she had a cochlear implantation. At the age of 3, she fainted with a discomfort after doing some effort. The immediately performed EKG shows a very long QT interval with QT corrected to 0.527 seconds (Figure-1). She was put on b-blocker therapy and drug avoidance instructions were given and she received an indwelling pacemaker implantation. Since the discomforts are no longer reproduced. The family investigation revealed an asymptomatic congenital long QT syndrome in the father.



Fig-1: QTc prolonged (527 ms)

## DISCUSSION

The QTc is prolonged if it is greater than 440 ms in men and greater than 460 ms in women and children. However, a much higher QTc value is often reported in the literature, and Congenital long QT syndrome, defined by the corrected QT space (calculated according to Bazett's formula) greater than 0.44 seconds, is a rare condition.

An international multicenter study reported only 287 cases in seven countries [1]. Congenital long QT syndrome obeys two modes of transmission, a recessive mode of transmission, associated with hearing loss (Jervell-Lange-Nielsen syndrome) to which our observation corresponds and a dominant mode (Romano-Ward syndrome).

Family history and genetic origin have been known for many years. Thirteen genes encoding cardiac ion channel subunits (potassium and calcium) have been distinguished. In 75% of cases, the mutation is identified, and in 90% of cases, it is one of the following three genes: LQT1, LQT2 and LQT3 [2].

Mutations in genes coding for ion channel subunits are responsible for an increase in Na<sup>+</sup> and Ca<sup>2+</sup> + depolarizing currents, or decrease repolarizing potassium currents. The consequence is a prolongation of the ventricular action potential, resulting in a prolongation of the QT interval, which can degenerate into Torsade de Pointe. Our patient was not subjected to a genetic study.

It is usual to use Bazett's formula to correct the QT value for the heart rate ( $QTc = QT / \sqrt{RR}$ ). It is accepted that a threshold greater than 500 ms would be a factor of poor prognosis and risk of occurrence of torsade de pointes.

Clinically, these patients experience syncope triggered by exercise or intense effort, emotion, anesthesia or excessive noise. Another mode of revelation is represented by seizures, even by sudden death. In our observation, there was a sudden death in our patient's siblings. The mode of presentation was uneasiness and loss of consciousness.

Congenital long QT syndrome affects boys and girls without predilection; as in the case reported by Garson.

In the absence of secondary causes that can prolong the QT (drug, ionic), the diagnosis should be considered in the following cases:

- QTc measured greater than or equal to 480 ms in an asymptomatic subject or when the calculation of the "probability score" based on clinical criteria, ECG and family history is greater than 3 (2);

- Mutation identified in the patient, regardless of the length of his QTc;
- Patient with a history of syncope with no other cause found, with a QTc greater than or equal to 460ms, on ECG 12 repeated leads.

It is common to use Bazett's formula to correct the QT value based on heart rate. In our observations, the QT is abnormally long (greater than 500 ms). Given the variability of QTc in each individual and even in individuals with congenital long QT syndrome, a probability score has been proposed by Schwartz *et al.*, [3] which also takes into account other ECG abnormalities as well as a personal or family history of malaise.

According to this classification, our patient had a high probability of congenital long QT syndrome.

The introduction of beta-blockers in patients with prolonged QTc is a recommendation for class I, and class IIa for those with normal QTc with an identified mutation.

The recommended beta-blocker is naldolol at a dosage of 1 mg / kg / D, the protective effect is expected for LQT1 patients and certain LQT2.

For LQT3, the anti-arrhythmic treatment that has demonstrated its effectiveness is mexiletine, a sodium channel blocker at a dosage of 8 mg / kg / day (4).

The implantable defibrillator finds an indisputable indication in survivors of sudden death (class I) with syncope with beta-blocker (class IIa) and high-risk subjects like SQT2, SQT3, and QTc > 500 ms (class IIb) in the homozygous Jervell and Lange-Nielsen syndrome, and in symptomatic syncopal SQT3 or if the QTc exceeds 550 ms.

Survivors of sudden death (class I) with beta blocker syncope (class IIa) and high-risk subjects like SQT2, SQT3, and QTc > 500 ms (class IIb) [5-7].

The education of children is an integral part of therapeutic management with advices aiming to avoid any adrenergic stress, any competitive sport (especially SQT1), any noise stress (especially SQT2) and certain drugs liable to prolong the QT or to train torsades de pointes.

## CONCLUSION

Before of any unexplained discomfort, whether or not linked to the effort, in front of any atypical and unexplained comitality. The clinician should have the reflex to perform an electrocardiogram and calculate the corrected QT interval. Once the diagnosis is made, exploration of other family members is essential.

The patient prognosis was transformed by the b-blocker treatment and the implementation of measures to prevent risk factors for torsades de pointes.

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