CLINICAL VIGNETTE

Cardiac Arrest in a Middle-Aged Woman due to an Undetected Long QT Interval Syndrome

Anita Srinivasa, MD

Introduction

Long QT syndrome is a very rare heart rhythm disorder which affects repolarization of the heart after a heartbeat.[1] This increases risk of developing irregular heartbeats, palpitations and even sudden death. This electrical disturbance is often seen on an Electrocardiogram as a prolonged QT interval. It can either be a genetic disorder or acquired as in our patient from electrolyte imbalances or from the use of certain medications.[1]

Case Report

Patient is a healthy 53-year-old female with no active medical problems. She recently started a vegetarian diet and exercised at least four times a week with no problems. On the day prior to her hospitalization she developed abdominal discomfort and diarrhea. Over the night she had about ten loose stools. The following day, while talking with her husband, she collapsed and became unresponsive. Paramedics arrived within 4 minutes and full cardiac arrest with ventricular fibrillation. ACLS was initiated and she was shocked 3 times followed by an amiodarone bolus en route to the hospital. She was noted to have ROSC. Upon arrival at the hospital she had a pulse but was unresponsive and was intubated.

VITALS were as follows: HR 121 bpm.; BP 140/90.
LABS: Sodium 137, Potassium 3.3, Chloride 98, Bicarb 24, Magnesium 1.4. AST 188, ALT 134
EKG: NORMAL SINUS RHYTHM
PR: 174MS
QT: 316MS
QTc: 44ms

Hypothermia protocol was initiated and she underwent Cardiac Catherization which showed normal coronaries and an Ejection Fraction of 20 percent. The working diagnosis was cardiogenic shock with acute myocarditis and cardiac arrest. She remained intubated for 3 days after admission. Repeat echo showed improvement of Ejection Fraction to 40 percent. After completion of the hypothermia protocol, the EKG still continued to show a prolonged QT interval of 460 milliseconds.

She was evaluated by the electrophysiologist who confirmed long QT interval syndrome which was triggered by the hypokalemia from acute gastroenteritis.

She underwent placement of a dual chamber defibrillator.

Discussion

Long QT Syndrome is an electrical disturbance that is measured by a prolonged QT interval on an electrocardiogram. This is a rare condition with fewer than 200,000 cases diagnosed per year.[2]

This can be caused by an inherited genetic condition or acquired as a result of certain medications or electrolyte imbalances.

There are about 17 different genes associated with the LONG QT syndrome. About 20 percent of patients who definitively have the Long QT syndrome test negatively on the genetic assay. However, about 15-30 percent of relatives who test positive for the genetic assay have a normal QT interval.[2]

There are two types of inherited syndromes:

1) Romano-ward syndrome, found in patients who inherit only one genetic variant from one parent.
2) Jervell and Lange Nielsen syndrome, found in patients who inherit variants from both parents. This type tends to be more serious and children usually have congenital deafness.[3]

Causes of acquired prolonged QT include:

1) Electrolyte imbalances like hypokalemia
2) Antibiotics including the quinolones, including Levofloxacin, Ciprofloxacin, Macrolides)[4]
3) Antipsychotics and Antidepressants (Venlafaxine, Mirtazapine, Escitalopram, Haloperidol)[4]
4) Diuretics [5]

Risk Factors

People with higher risk of inherited or acquired QT include :

1) Children and young adults with unexplained fainting, near drownings, or unexplained seizures
2) Family members of children and young adults with unexplained fainting, unexplained near-drownings or seizures
3) First degree relatives of people with known LONG QT syndrome
4) People with electrolyte imbalances such as those with anorexia or bulimia nervosa[6]

Clinical Symptoms

1) FAINTING - the most common sign of long QT syndrome is fainting spells- that can occur when one is excited, angry, scared or even during exercise. This fainting is not associated with a prodrome or a warning like feeling lightheaded or having blurry vision, which can often occur with other types of syncope.[6]
2) PALPITATIONS
3) SEIZURES from continuous erratic heart beats, causing anoxia and seizures[7]
4) SUDDEN DEATH, which can occur if the heart beat does not return to its normal rhythm or if an external defibrillator is not used in time to convert the rhythm.

Complications

1) TORSADES DE POINTES, can present as syncope although the heart can correct itself if it lasts less than a minute. If it persists it can lead to ventricular fibrillation.
2) VENTRICULAR FIBRILLATION

Diagnosis

The diagnosis is made on a routine electrocardiogram that shows a prolonged QT interval. QT interval on an EKG represents the time of ventricular activity including depolarization and repolarization. It is measured from the beginning of the QRS complex to the end of the T WAVE. The normal QT interval measures 0.36-.0.44 milliseconds (or 9-11 boxes on the EKG). Not all patients will show changes on a single EKG tracing. Several EKG tests done over a period of time may be needed or a patient may have to wear a Holter monitor for 24 hours.[6]

Some patients only exhibit long QT when they exercise, so stress testing may be needed.

Management

Treatments are based on varying factors including[8]: whether a patient has had a cardiac arrest, what type of Long QT Syndrome, any prior fainting episodes.

Treatments include*:* Placement of defibrillator,[8] managing lifestyle - including immediate correction of hypokalemia with gastroenteritis and dehydration, avoiding medications that can trigger symptoms such as diuretics and certain antibiotics and avoiding strenuous exercises.

REFERENCES

1. Mayo Clinic: Long QT syndrome [Internet]. Available from: https://www.mayoclinic.org/diseases-conditions/long- \qt-syndrome/symptoms-causes/syc-20352518.
2. NHLBI: Long QT syndrome [Internet]. Available from https://www.nhlbi.nih.gov/health-topics/long-qt-syndrome.
3. Genetics Home Reference. NIH US National Library of Medicine. Jervell and Lange-Nielsen syndrome [Internet]. Available from https://ghr.nlm.nih.gov/condition/jervell-and-lange-nielsen-syndrome.
4. **Frothingham R**. Rates of torsades de pointes associated with ciprofloxacin, ofloxacin, levofloxacin, gatifloxacin, and moxifloxacin. *Pharmacotherapy*. 2001 Dec;21(12): 1468-72. PubMed PMID: 11765299.
5. **Bauman JL**. The role of pharmacokinetics, drug interactions and pharmacogenetics in the acquired long QT syndrome. *Eur Heart J Supplements*. 2001; 3(suppl K): S93-S100.
6. **Al-Khatib SM, LaPointe NM, Kramer JM, Califf RM**. What clinicians should know about the QT interval. *JAMA*. 2003 Apr 23-30;289(16):2120-7. Review. Erratum in: *JAMA*. 2003 Sep 10;290(10):1318. PubMed PMID: 12709470.
7. University of Rochester Medical Center: The heart-brain connection: The link between LQTS and seizures [Internet]. Available from: https://www.sciencedaily.com/releases/2016/07/160729132908.htm
8. **Schwartz PJ, Dagradi F, Castelletti S**. Evolution in Managing Long QT Syndrome: From Registries to Centers of Excellence. *J Am Coll Cardiol*. 2017 Jul 25;70(4):463-465. doi: 10.1016/j.jacc.2017.06.004. PubMed PMID: 28728691.

Submitted February 21, 2019