Romano-Ward syndrome

Romano-Ward syndrome is a condition that causes a disruption of the heart's normal rhythm (arrhythmia). This disorder is a form of long QT syndrome, which is a heart condition that causes the heart (cardiac) muscle to take longer than usual to recharge between beats. The term "long QT" refers to a specific pattern of heart activity that is detected with an electrocardiogram (ECG or EKG), which is a test used to measure the electrical activity of the heart. In people with long QT syndrome, the part of the heartbeat known as the QT interval is abnormally long. Abnormalities in the time it takes to recharge the heart lead to abnormal heart rhythms.

The arrhythmia associated with Romano-Ward syndrome can lead to fainting (syncope) or cardiac arrest and sudden death. However, some people with Romano-Ward syndrome never experience any health problems associated with the condition.

Fifteen types of long QT syndrome have been defined based on their genetic cause. Some types of long QT syndrome involve other cardiac abnormalities or problems with additional body systems. Romano-Ward syndrome encompasses those types that involve only a long QT interval without other abnormalities.

Frequency

Romano-Ward syndrome is the most common form of inherited long QT syndrome, which affects an estimated 1 in 2,000 people worldwide. Long QT syndrome may actually be more common than this estimate, however, because some people never experience any symptoms associated with arrhythmia and therefore may not be diagnosed.

Causes

Mutations in the *KCNQ1*, *KCNH2*, and *SCN5A* genes are the most common causes of Romano-Ward syndrome. These genes provide instructions for making proteins that form channels across the cell membrane. These channels transport positively charged atoms (ions), such as potassium and sodium, into and out of cells. In cardiac muscle cells, these ion channels play critical roles in maintaining the heart's normal rhythm. Mutations in any of these genes alter the structure or function of the channels, which changes the flow of ions in and out of cells. A disruption in ion transport alters the way the heart beats, leading to the abnormal heart rhythm characteristic of Romano-Ward syndrome.

Mutations in other genes involved in ion transport can also cause Romano-Ward syndrome; each of these additional genes is associated with a very small percentage of cases.
Inheritance Pattern
Romano-Ward syndrome follows an autosomal dominant pattern of inheritance, which means one copy of the altered gene in each cell is sufficient to cause the disorder. In most cases, an affected person inherits the mutation from one affected parent. A small percentage of cases result from new mutations in one of the associated genes. These cases occur in people with no history of Romano-Ward syndrome in their family. Some people who have an altered gene never develop signs and symptoms of the condition, a situation known as reduced penetrance.

Other Names for This Condition
• RWS
• Ward-Romano Syndrome
• WRS

Diagnosis & Management
Genetic Testing Information
• What is genetic testing? /primer/testing/genetictesting

Research Studies from ClinicalTrials.gov
• ClinicalTrials.gov https://clinicaltrials.gov/ct2/results?cond=%22Romano-Ward+syndrome%22+OR+%22Long+QT+Syndrome%22

Other Diagnosis and Management Resources
• MedlinePlus Medical Tests: Electrocardiogram https://medlineplus.gov/lab-tests/electrocardiogram/
Additional Information & Resources

Health Information from MedlinePlus

- Encyclopedia: Arrhythmias
  https://medlineplus.gov/ency/article/001101.htm
- Encyclopedia: Electrocardiogram
  https://medlineplus.gov/ency/article/003868.htm
- Health Topic: Arrhythmia
  https://medlineplus.gov/arrhythmia.html
- Health Topic: Congenital Heart Defects
  https://medlineplus.gov/congenitalheartdefects.html
- Medical Tests: Electrocardiogram
  https://medlineplus.gov/lab-tests/electrocardiogram/

Genetic and Rare Diseases Information Center

- Long QT syndrome
- Long QT syndrome 1
  https://rarediseases.info.nih.gov/diseases/3284/long-qt-syndrome-1
- Long QT syndrome 2
- Long QT syndrome 3

Additional NIH Resources

- National Heart, Lung, and Blood Institute
  https://www.nhlbi.nih.gov/health-topics/long-qt-syndrome

Educational Resources

- American Heart Association
- Centre for Genetics Education (Australia)
- KidsHealth from the Nemours Foundation
- MalaCards: long qt syndrome 1
  https://www.malacards.org/card/long_qt_syndrome_1
• MalaCards: long qt syndrome 2
  https://www.malacards.org/card/long_qt_syndrome_2

• Merck Manual Consumer Version: Long QT Syndrome

• Orphanet: Familial long QT syndrome
  https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=768

Patient Support and Advocacy Resources
• National Organization for Rare Disorders
  https://rarediseases.org/rare-diseases/romano-ward-syndrome/

• Resource List from the University of Kansas Medical Center
  http://www.kumc.edu/gec/support/conghart.html

• Sudden Arrhythmia Death Syndromes (SADS) Foundation: Long QT Syndrome
  https://www.sads.org/What-is-SADS/Long-QT-Syndrome#.Vds7xZdGdD8

Clinical Information from GeneReviews
• Long QT Syndrome

Scientific Articles on PubMed
• PubMed
  https://www.ncbi.nlm.nih.gov/pubmed?term=%28%28Romano-Ward+syndrome%5BTIAB%5D%29+OR+%28long+QT+syndrome%5BTI%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+360+days%22%5Bdp%5D

Catalog of Genes and Diseases from OMIM
• LONG QT SYNDROME 1
  http://omim.org/entry/192500

• LONG QT SYNDROME 2
  http://omim.org/entry/613688

• LONG QT SYNDROME 3
  http://omim.org/entry/603830

Medical Genetics Database from MedGen
• Long QT syndrome

• Romano-Ward syndrome
Sources for This Summary

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/20301308

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/27807201

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/22872816
  Free article on PubMed Central: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3392134/

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/26108145

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/22895603
  Free article on PubMed Central: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3461497/

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/24932360
  Free article on PubMed Central: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4051331/

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/11286954

  Citation on PubMed: https://www.ncbi.nlm.nih.gov/pubmed/15929461

Reprinted from Genetics Home Reference:

Reviewed: May 2017
Published: November 12, 2019

Lister Hill National Center for Biomedical Communications
U.S. National Library of Medicine
National Institutes of Health
Department of Health & Human Services