

Gilbert Gottfried had myotonic dystrophy before his death. What is the rare disease?

April 13 2022, by Mike Stunson

Comedian Gilbert Gottfried died on Tuesday after a long illness, his family announced.

His publicist revealed in a statement to multiple publications, including People, Gottfried had "recurrent ventricular [tachycardia](#) caused by [myotonic dystrophy](#) type 2."

Here's what to know about ventricular tachycardia and myotonic dystrophy.

What is myotonic dystrophy and what causes it?

"Myotonic dystrophy is a form of muscular dystrophy that affects muscles and many other organs in the body," the Muscular Dystrophy Association said. It comes from the word myotonia, which is "an inability to relax muscles at will."

Type 2 myotonic dystrophy is a milder version of the disease, which is prevalent in about 10 cases per 100,000 individuals, the association said.

It's caused by a mutation in the CNBP gene, whereas type 1 is caused by a mutation in the DMPK gene, according to the National Institutes of Health.

"Mutations in each of these genes involve a short segment of DNA that

is abnormally repeated many times, the NIH said. "This abnormal repetition forms an unstable region of the gene. These changes keep cells in the muscles and other [body tissues](#) from functioning normally, leading to signs and symptoms of myotonic dystrophy."

What are the symptoms of myotonic dystrophy type 2?

The neck and finger muscles are often the first ones affected in type 2 myotonic dystrophy. In addition to muscle weakness and pain, those who have the disease may suffer from myotonia or cataracts, according to the National Organization for Rare Disorders.

"For example, a person may have difficulty releasing their grip on a doorknob or handle," the National Institutes of Health said. "Also, affected people may have slurred speech; temporary locking of their jaw; and muscle pain and weakness that mainly affects the neck, shoulders, elbows, and hips."

Johns Hopkins Medicine refers to it as a "complex disorder," as it affects "many organ systems throughout the body."

There is no treatment to stop or slow myotonic dystrophy type 2, the NIH said, only methods to help handle the symptoms.

What is ventricular tachycardia?

Gottfried's myotonic dystrophy reportedly caused ventricular tachycardia, which is "a type of abnormal heart rhythm... (that) occurs when the lower chamber of the heart beats too fast to pump well and the body doesn't receive enough oxygenated blood," Johns Hopkins Medicine said.

It can be caused by some medications, excessive caffeine or alcohol, an electrolyte imbalance, or some genetically transmitted conditions, including myotonic dystrophy, according to Penn Medicine.

A normal resting heart beats at 60 to 100 times per minute, but with tachycardia, heart rates of 150 to 250 beats per minute are common, Stanford Health Care said.

What are the symptoms of ventricular tachycardia

According to Johns Hopkins Medicine, symptoms of ventricular tachycardia are [chest pain](#), dizziness, fainting, shortness of breath and cardiac arrest.

"During an episode of ventricular tachycardia, the heart is beating so fast that the [blood pressure](#) drops so the heart cannot pump enough oxygen to every part of the body, and this is what causes symptoms," Cleveland Clinic said.

Gottfried's ventricular tachycardia was reportedly recurrent, meaning it happened often. Episodes that last longer than a few seconds have potential to be life-threatening, the Mayo Clinic said.

To treat ventricular tachycardia, patients are encouraged to manage any diseases that may cause the condition, Cleveland Clinic said. In Gottfried's case, there was no treatment for myotonic dystrophy.

©2022 The Charlotte Observer.

Distributed by Tribune Content Agency, LLC.

Citation: Gilbert Gottfried had myotonic dystrophy before his death. What is the rare disease? (2022, April 13) retrieved 28 April 2024 from <https://medicalxpress.com/news/2022-04-gilbert-gottfried-myotonic-dystrophy-death.html>

This document is subject to copyright. Apart from any fair dealing for the purpose of private study or research, no part may be reproduced without the written permission. The content is provided for information purposes only.