

# Brain and heart link may explain sudden death in Rett

December 14 2011

---

Poets might scoff at the notion that heart and brain are closely related, but scientists led by those at Baylor College of Medicine ([www.bcm.edu](http://www.bcm.edu)) say a genetic defect that affects the brain can stop a heart.

In a study published today in the journal [Science Translational Medicine](#), BCM researchers and colleagues found that [heart problems](#) that occur in nearly 20 percent of children with Rett syndrome, a [neurological disorder](#), originate because the Rett gene ([MeCP2](#)) is lost in [nerve cells](#) – not in heart muscle cells.

"The finding in mice opens the door to tailoring treatments to prevent heart rhythm problems and even deaths in girls with the disorder," said Dr. Jeffrey Neul, associate professor of pediatrics at BCM, a researcher at the Jan and Dan Duncan Neurological Research Institute at Texas Children's Hospital and the assistant medical director of the Blue Bird Circle Rett Clinic.

"We began by looking at mice whose symptoms closely mimicked that of Rett syndrome. We found that those mice also exhibited electrical malfunctions in the heart, known as long QT syndrome," said BCM collaborator Dr. Xander Wehrens, professor of molecular physiology and biophysics and the Juanita P. Quigley endowed chair in cardiology at BCM. Long QT syndrome describes an interval usually measured on an electrocardiogram or ECG. These electrical malfunctions cause that interval to be too long, which can lead to a dangerous heart beat that can cause [sudden death](#) in people.

"Long [QT syndrome](#) is found in nearly 20 percent of people with Rett syndrome," said Neul. Sudden death also occurs in a significant percentage of Rett patients, and Neul and his colleagues wonder if the two might be linked.

The gene MECP2 occurs in the genomes of every cell, providing a code for the protein associated with it, MeCP2. It can have different activities in different cells. In this study, researchers created a mouse in which MeCP2 was missing only in the [brain](#) and [nervous system](#). Even though MeCP2 functioned normally in the heart, the heart beat abnormally.

"We showed that the heart problems were actually secondary to nervous system deficits and that cardiac activity can be remodeled by abnormalities in brain activity," said Neul. "Basically, the brain is sending the wrong message to the heart leading to the malfunction."

Taking a closer look at the heart cells affected by the nervous system deficits, researchers found an unusual persistent sodium current.

Sodium currents occur naturally in cell function. When there is cell activity, sodium travels in and out of the cell through a channel that acts like a window opening and closing. Unusual persistent sodium current means the "window" doesn't close properly, causing a slow trickle of sodium to flow through the channel for a longer period of time.

There are seizure medications that block this process, which also occurs in other neurological disorders. When Neul and his colleagues used an anti-seizure drug to treat these malfunctioning heart muscle cells in the lab, the sodium current problem improved. They then gave the common anti-seizure drug to the abnormal mice which corrected the [heart](#) rhythm problem and blocked sudden cardiac death.

The next step is to treat the animals long term, he said. At that same

time, they plan to look at what is going on in children with Rett who are already taking these kinds of medicines to treat their seizures.

"Are the people on these drugs less likely to have long QT intervals?" he said, "More research in the lab will help us reassess what we are doing in the clinic and determine if we need to change our treatment methods," Neul said.

Provided by Baylor College of Medicine

Citation: Brain and heart link may explain sudden death in Rett (2011, December 14) retrieved 24 April 2024 from <https://medicalxpress.com/news/2011-12-brain-heart-link-sudden-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.