MICHAEL ACKERMAN: Hello and welcome to this month's Heart of the Matter brought to you by Mayo Clinic and the SADS Foundation. I'm Dr. Michael Ackerman and I'm the Director of Mayo Clini's Long QT Syndrome Clinic, and the Windland Smith Rice Sudden Death Genomics Laboratory, and the president of the SADS Foundation.

This month's Heart of the Matter question asking about the relationship between long QT syndrome and genetic heart disease is like long QT syndrome and sudden infant death syndrome. Our laboratory at Mayo Clinic, and Peter Schwartz's program in Italy, have advanced our understanding of the relationship between the death of an infant during his or her first year of life that is otherwise unexplained, and the possibility that a cardiac channelopothy a genetic heart rhythm condition like long QT syndrome, or CPVT, or Brugada syndrome, could be to blame.

In the United States, about 3,000 apparently healthy infants will tragically fail to reach their first birthday. There won't be a reason at medical examination at their autopsy. It will be unexplained. It will get called SIDS, sudden infant death syndrome, or SUDI, sudden unexplained death during infancy. We now know that a small percentage of those SIDS deaths actually stem from, are the result of a lethal arrhythmia due to the presence of a genetic heart disease like long QT syndrome.

That number is about 10% to 15%. So it's very important for people to know that the vast majority of tragic, unexplained infant death or SIDS, is not due to a cardiac channelopothy like long QT syndrome. But the fact that we have observed that about 10% to 15% is-- with most of that 10% to 15% being due to long QT syndrome, and a small portion of that 10% to 15% being due to other genetic heart diseases like CPVT, or Brugada syndrome-- that should cause the family, when there has been such a tragedy, to ask, are there any other unexplained deaths in the family that might link the death of a three-month-old infant to a death many years ago of a 30-year-old, 40-year-old relative. That unexplained accident, that unexplained drowning, that person who is labeled with exercise-induced epilepsy, to learn clues that link this family together.

So because of that observation that about 10% of unexplained infant death could be due to long QT syndrome, it certainly is reasonable that when there has been such a tragedy, that these questions get asked of the family, and that the immediate relatives of that lost infant, the siblings, the parents, get a 12-lead electrocardiogram to further explore the possibility that long QT syndrome might have been to blame.

Now more recently we're learning that, underneath the umbrella of what is called unexplained infant death or SIDS, it's not all the same. Most SIDS deaths occur between two to four months of age. That's the majority. And so within that time period of infant death, long QT syndrome turns out to be not very common at all. But when that sudden infant death occurred in a six-month-old infant, or an eight-month-old infant, now it's possible that long QT syndrome is the root cause with a much higher likelihood.

How much higher? About a 20% to 25% chance of it being due to long QT syndrome when there has been a sudden death of a 6-, 7-, 8-month-old, which is a uncommon time period for what is called SIDS. And hopefully, I've been able to share with you that there is a relationship between SIDS and genetic heart rhythm diseases like long QT syndrome. But that importantly, we remember that the vast majority of SIDS is not due to these genetic heart rhythm diseases. So that is the question for this month's Heart of the Matter and I look forward to being back with you next month. Have a good day.