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Risk factors for sudden death for adult muscular dystrophy identified

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The largest assessment of people with adult muscular dystrophy has identified risk factors that can lead to sudden death for individuals with the most common form of this disease. The results of the multicenter study, led by the Indiana University School of Medicine, are reported in the June 19 issue of the *New England Journal of Medicine*.

Neurologists and cardiologists at 23 neuromuscular disease clinics nationwide affiliated with the Muscular Dystrophy Association assessed 406 adult patients with myotonic dystrophy type 1 using clinical history, genetic assessment and electrocardiograms (ECG) to determine the risk factors that cause arrhythmias and sudden cardiac death.

"The study has prospectively identified risk factors that predict a high risk of sudden death in people with myotonic dystrophy, the most common form of muscular dystrophy that we see in adults," said the study's principal investigator and lead author William J. Groh, M.D., M.P.H., associate professor of medicine at the Indiana University School of Medicine and the Krannert Institute of Cardiology. Sudden death is defined as a death that occurs in a stable patient within one hour of the onset of symptoms.

During the 10 years of the study, 20 percent of the people enrolled died, said Dr. Groh. Of those, one-third died of sudden death likely attributable to cardiac arrhythmia.

"Patients who had significant abnormalities on their ECG were at a 3.5

times higher risk of sudden death," said Dr. Groh. "Those with atrial (upper) chamber arrthymias had a 5 times higher risk."

In the future, physicians can use these risk factors to evaluate patients with myotonic dystrophy to hopefully prevent sudden death through further evaluation including electrophysiological studies (using catheters in the heart) or by surgically implanting a cardioverter-defibrillator.

Dr. Groh said another important outcome of the study was the discovery that pacemakers, commonly used to treat some forms of arrhythmia, did not help these patients prevent sudden death.

Myotonic muscular dystrophy, an inherited disease, is characterized primarily by progressive muscle weakness and muscle wasting. It affects approximately 1 of every 8,000 people in the United States. In many muscular dystrophies, the heart muscle is adversely affected. Those heart abnormalities can be as serious and debilitating as the skeletal muscle involvement more commonly associated with muscular dystrophy.

Source: Indiana University