Study helps predict which ARVD patients are at highest risk of sudden cardiac death

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(Medical Xpress) -- Johns Hopkins experts in arrhythmogenic right ventricular dysplasia (ARVD) have defined a set of criteria that could be used to assess a patient's need for an implanted defibrillator to prevent sudden death. In a study to be published in the September 27 issue of the Journal of the American College of Cardiology that is now online, the researchers report that using those criteria, they were able to separate the patients at high risk for a life-threatening irregular heart rhythm from those with low risk.

ARVD is an inherited cardiac disorder and one of the most common causes of sudden death in athletes and young, apparently healthy adults. ARVD creates scarring of the muscle, mainly on the right side of the heart. The scarring interrupts the normal electrical activity of the heart, causing very fast, abnormal heart beats that prevent the heart from pumping blood to the rest of the body. Without a shock from an external or an implanted defibrillator, the condition can be fatal.

Not everyone who has a diagnosis of ARVD will go on to have a life-threatening arrhythmia, but prior to the Johns Hopkins study, there was very little data to help determine who should have an implanted defibrillator for primary prevention of sudden cardiac death.

The study included 84 patients with definite or probable ARVD. None had a history of life-threatening irregular heartbeats from the lower chambers of the heart, known as sustained ventricular arrhythmias, but all had been given an implanted defibrillator as a precaution and were

followed for an average of five years. During that time, 48 percent needed either a shock or rapid pacing from the device to stop a dangerous abnormal rhythm while the remaining 52 percent did not experience an arrhythmia that required therapy from the defibrillator.

“Whether an ARVD patient should have an implantable defibrillator for primary prevention against sudden death is a critically important decision,” says Hugh Calkins, M.D., professor and head of cardiac electrophysiology. Dr. Calkins runs the Johns Hopkins ARVD program, which includes a large registry of ARVD patients in North America. “These are usually young patients with few or no symptoms, and putting in a defibrillator, which they will have for the rest of their lives, requires careful consideration,” adds Calkins, who is the senior author of the study.

Since ARVD runs in families, people with relatives who have died from sudden cardiac death and those who’ve had episodes of abnormally fast heartbeats coming from the lowers chamber of the heart need to be evaluated. For those who meet diagnostic criteria for ARVD, an implanted defibrillator is usually recommended to prevent sudden death.

Calkins and his team identified four criteria that should be considered to help determine which patients are at higher risk and most in need of the defibrillator. Patients in the study whose devices produced shocks or rapid pacing to stop abnormal rhythms generally met two or more of the criteria thought to put them at higher risk.

“We found that there was an incremental and additive risk to these patients the more criteria they met. So if a patient was found to match only one of the four criteria, that person was at lower risk. A patient who was positive on all four criteria was at highest risk,” says lead author Aditya Bhonsale, M.D.

One of those criteria was whether a sustained irregular heart rhythm could be induced by a procedure in the electrophysiology lab. Two of the other criteria related to findings on a Holter monitor, which patients wear for 24 hours to monitor the electrical activity of the heart. The fourth characteristic that was found to have higher value in predicting risk was whether the patient was the first in the family to be diagnosed with ARVD.

None of the study participants who had either zero or one risk factor needed shocks or rapid pacing from their defibrillator to stop a life-threatening arrhythmia during the study period. However, 23 percent of those with two risk factors needed defibrillator therapy. The number increased to 65 percent among those with three risk factors, and 78 percent for those with four risk factors.

“Our goal was to provide a good template for physicians to assess a patient’s risk,” says Bhonsale. “Although we followed some of the patients for as long as 10 years, we cannot say with complete certainty that their long-term risk would stay the same, but the majority of those who had appropriate shock or rapid pacing got it within one year of their defibrillator implantation,” he adds.

The researchers say that the study represents an important step in understanding the factors that predict which patients are at highest risk of sudden death and the need for an implanted defibrillator. ARVD is often a progressive disease and patients need to be followed on an ongoing basis. At Johns Hopkins, ARVD patients from across the country are part of the specialized program and are evaluated every one to three years.

Although about half of the study participants overall did not need defibrillator therapy to stop a potentially fatal arrhythmia during the study, Bhonsale points out that for every two ARVD patients with the

device, one patient's life was saved. He says that is a very favorable percentage when looking at the number of people who need to be treated in order to save one life.

"ARVD is a mysterious form of cardiomyopathy that kills young people. We are working hard to find answers and help patients and their physicians make decisions about therapy. We hope that this data will provide value to those who are engaging in those discussions," says Calkins.

Provided by Johns Hopkins University

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