Treatment outlook for adults with hypertrophic cardiomyopathy move from grim to good

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Newly published research led by the Minneapolis Heart Institute Foundation (MHIF) and Tufts Medical Center in Boston shows that implantable defibrillators (ICDs), along with other modern treatments, have reduced mortality rates and are helping patients with hypertrophic cardiomyopathy (HCM) live longer, including normal life expectancy. This research "changes our perceptions of HCM from a grim, unrelenting, and largely untreatable condition to a contemporary disease with effective treatment options and a low rate of death," states Dr. Barry Maron, lead researcher and Director of the HCM Center at MHIF. Dr. Maron will present the results of the study at the American College of Cardiology (ACC) conference in San Diego, CA on March 14.

HCM is a genetic heart condition in which the walls of the heart are abnormally thick. Despite the condition, many people with HCM live normal, healthy lives. But others develop cardiac problems that affect their quality of life or, in some cases, lead to early death. In particular, HCM can cause a dangerously rapid heart rhythm that can lead to sudden cardiac death (SCD).

With research and technology advances, treatment for adults with HCM has changed dramatically over the past 10-15 years. For example, ICDs, which were originally designed to prevent SCD in people with coronary artery disease, are now commonly prescribed for people with HCM who are at high risk for sudden death. If a dangerously rapid heart rhythm is

detected, ICDs work by delivering a shock that restores the heart's normal rhythm.

Dr. Maron wanted to know just how effective ICDs, along with other advances in care, were at changing the course of HCM patients from grim to good. To find the answer, he and a team of researchers at Tufts—led by his son Dr. Martin Maron—examined the long-term outcomes (death rates and causes of death) in 1000 adults with HCM from 1992-2011. The results? Mortality rates among adults with HCM were about the same as death rates among adults in the general population. "Today's treatment interventions, including ICDs for SCD prevention (an initiative started here at MHIF), have dramatically changed the outlook for these patients," said Dr. Maron. "They are living longer and deaths directly associated with the disease are much less common, especially those caused by SCD."

In the study, while 4% of patients had died from an HCM-related death at follow-up, 6% had survived a life-threatening event, most commonly attributed to an ICD (but also heart transplant, surgical myectomy or out-of-hospital defibrillation). Notably, of 17 HCM sudden deaths, 6 occurred in patients who declined a formal recommendation for ICD therapy and 11 were either initially evaluated in the 1990s prior to general employment of ICDs for HCM, or had no risk factors to justify an ICD decision.

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