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A Silent Killer Strikes The Hearts of the Young

By JESSICA KOVLER (New York Times)
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It was the morning of March 29 -- Good Friday, Michael Kress remembers -- when he entered the bedroom of his 14-year-old son, Timmy, to wake him for school.

Mr. Kress found Timmy on the floor. He yelled for his wife to dial 911, and he tried cardiopulmonary resuscitation. But Timmy had been dead for several hours.

Mr. Kress later learned that Timmy had a rare heart disease called hypertrophic cardiomyopathy, which often attacks without warning. In Timmy's case, it could have been detected and his death prevented, but the screening is expensive and is rarely recommended routinely by doctors.

In hypertrophic cardiomyopathy the growth of the heart muscle is excessive and sometimes blocks the flow of blood to the aorta artery.

The disorder, which is rarely detected early, is the most common cause of disease-related sudden death among young people. More than half of those with the disorder are genetically predisposed to it. Athletes in top physical condition are often among the victims.

"We wish we would have known sooner," Mr. Kress said. "If Timmy underwent some form of screening he might still be with us."

The disorder afflicts one in 500 people, though not always with fatal results.

A recent study by the University of California at San Francisco found that roughly half of the cases may be at least partly attributed to a mutation that disrupts at least two genes and interferes with the heartbeat.

Since Timmy's death, his four brothers have been tested for the disorder. Two do not have it; two are awaiting further testing.

The relevant genes contain the code for proteins that interact with the heart muscle. The mutations cause the muscle to grow excessively, thickening the heart and causing arrhythmia, which can lead to sudden death.

People with the the disorder fall into three categories: those with mild symptoms, those susceptible to episodic heart failure and those at high risk of sudden death.

A majority of these cases occur in otherwise healthy teenagers, often athletes, and researchers remain largely at a loss for an explanation for cases other than those associated with the genetic mutation.

"This disease comes totally out of left field," said Dr. Vinay Nadkarni, the chairman of the emergency cardiovascular committee of the American Heart Association.

Most people, but especially children, do not come to a doctor and demonstrate symptoms, Dr. Nadkarni said. "They collapse on the baseball diamond and die, and that's the end of it," he said.

Many doctors say exercise can strain the heart, aggravating the disorder, and actually contribute to cardiac arrest.

Because hypertrophic cardiomyopathy is relatively rare, many doctors are reluctant to recommend echocardiograms to screen young patients for the disorder. The screening is noninvasive, but it costs about $1,100.

No cures have been found for the disease, and doctors can promise nothing to protect patients completely from sudden death or even irregular heartbeats.

But several forms of therapy can treat symptoms effectively.

Those treatments include beta-blockers and calcium channel blockers. Pacemakers, implanted automatic defibrillators and surgery to remove a part of the overgrown muscle are also effective, though many doctors recommend them as a last resorts.

A procedure called alcohol septal ablasion, which uses 100-proof alcohol, is also available. It essentially causes a controlled, localized heart attack, killing many cells of the muscle. Experts say this reduces the likelihood of a severe heart attack.

Sharon Bates, whose son Anthony died in 2000 while playing football for Kansas State University, has begun crusading for more testing for young athletes.

"Prevention," Ms. Bates said, "is 10 times better than hopeless resuscitation."