



Study Gets Handle on Deadly Heart Condition in Kids

A survey finds the incidence of cardiomyopathy is higher in boys, blacks and Hispanics.

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WEDNESDAY, April 23, 2003 (HealthDayNews) -- A first-ever report from a data registry on the often-fatal childhood condition called pediatric cardiomyopathy confounds some widely held medical views.

However, it also points the way toward better diagnosis and perhaps even treatment, says the man who started the registry.

The report finds the rare condition is likelier to strike boys than girls, and is more common among blacks and Hispanics than whites.

Cardiomyopathy consists of a family of conditions that affect the heart muscle. About 1,000 children are born in the United States each year with one form or another of cardiomyopathy.

While enormous strides have been made in treating conditions caused by problems with the heart's blood vessels, little progress has been made on heart muscle problems. A transplant is the only hope for many young cardiomyopathy patients and "the time to transplantation or death for children with cardiomyopathy has not improved during the last 35 years," says a report in the April 24 issue of *The New England Journal of Medicine*.

That report offers no miracle cure or treatment. Instead, it lays out precise numbers about pediatric cardiomyopathy -- when it is diagnosed, its incidence in different ethnic groups, the percentage of the different kinds of cardiomyopathy, and so on -- for two regions of the United States. They are New England and the central southwest region, which includes Texas, Oklahoma and Arkansas.

Those numbers alone represent a major advance, says study author Dr. Steven E. Lipshultz, who started the Pediatric Cardiomyopathy Registry when he was at Boston Children's Hospital. He is now a professor of pediatrics at the University of Rochester School of Medicine in New York.

For example, they show that many textbooks are flat wrong in a lot of the things they say about the condition, Lipshultz says. "If you look at some of the current textbooks in pediatric cardiology, you will find they state that hypertrophic cardiomyopathy [in which the heart muscle is abnormally thickened] is rare in adolescents. That is not true."

The registry says 42 percent of cases are hypertrophic, while 51 percent are dilated, in which the heart muscle expands abnormally, with scattered other causes accounting for the remainder.

And while pediatric cardiologists said in a survey the condition could be detected at almost any age, the registry shows most cases being diagnosed in the first year of life, Lipshultz says. "That suggests that many of the causes are genetic, while in adults many cases are related to health habit issues," he says.

Other indicators that genetics is important are striking differences in incidence between the sexes and ethnic groups.

The incidence is 1.32 per 1,000 in boys compared to 0.92 per 1,000 in girls, in large part because several genes for neuromuscular disease that can also cause cardiomyopathy are more common in boys.

The condition is more common in black children (1.4 cases per 1,000) and in Hispanics (1.41 per 1,000) than in whites (1.06 per 1,000). The incidence was about 50 percent higher in the Southwest than in New England, a finding that leaves the researchers puzzled.

The effect of genetics can be important in early detection, Lipshultz says. It is important to screen other family members when a case is diagnosed, he says, to pick up cardiomyopathy that may be causing few or no symptoms.

What he also finds striking is that an epidemiological study done in Australia, and reported in the same issue of the journal, produces very similar numbers. "These two studies, done without knowledge of each other, have come up with almost the same results," Lipshultz says.

That study, led by Dr. Robert G. Weintraub of the Royal Children's Hospital in Melbourne, covered all known cases diagnosed in children 10 and under between 1987 and 1996. It found about the same overall incidence, 1.24 per 1,000, the same concentration of diagnoses in the first year of life and the same higher incidence in a minority group, in this case Australian aborigines (2.47 per 1,000).

"It is remarkable" the studies have similar results, but also fortunate, Weintraub says. "We think that the two studies serve as external validation to each other."

Weintraub is director of the heart transplant program at Royal Children's Hospital, which serves all of Australia. He says the study was undertaken "to better gain an appreciation of how to plan heart transplantation for those children who require it." The study has helped to give "a better appreciation of how these cases present and behave, so we can make long-term predictions about which children will require transplants."

More information

You can get detailed information about cardiomyopathy from the National Heart, Lung, and Blood Institute (www.nhlbi.nih.gov). You can also read about children and heart disease from the American Heart Association (www.americanheart.org).

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