CARDIOMYOPATHY AND HEART FAILURE

Cardiac death in children

Non-sudden cardiac death (SCD), which is caused by progressive heart failure, is as common as SCD in children with hypertrophic cardiomyopathy. "Therefore," says Susan Denfield, MD from the Texas Children's Hospital and Baylor College of Medicine, "identification of risk factors for non-SCD is at least as important as identifying risk factors for SCD."

Risk factors for cardiac death have been identified for adults with hypertrophic cardiomyopathy and clear treatment strategies have been established. Whether these risk factors are applicable to children remains uncertain. Consequently, no consensus exists for treatment of children with hypertrophic cardiomyopathy and current treatment of pediatric patients is based on risk factors for adults. Texas Children's Hospital and Baylor College of Medicine has treated a large population of children with various types of cardiomyopathies, including isolated hypertrophic cardiomyopathy, and has access to clinical information about these children. Researchers there conducted a retrospective review of children under the age of 18 years to establish whether risk factors for adults are indeed predictive of cardiac death in children and whether guidelines for treatment of adults are appropriate for the management of children. "Understanding these factors would hopefully allow us to improve survival," says Denfield.

Of 426 patients diagnosed with hypertrophic cardiomyopathy between 1985 and 2006, 96 were eligible for inclusion in the analysis on the basis of age <18 years at the time of diagnosis and having no other syndrome or apparent cause for left ventricular hypertrophy. A treatment algorithm devised by the group was associated with a survival rate of 80% over the 20-year follow-up period. Primary end points—transplantation or death—occurred in 11 patients (4 patients underwent transplantation, 3 died of non-sudden cardiac causes, 3 died suddenly and 1 died of unknown causes). Four previously undiagnosed children presented with SCD but were resuscitated and survived. None of the adult risk factors predicted SCD; however, extreme left ventricular hypertrophy, defined as a *z* score of >6 (ventricular septal or left ventricular posterior wall thickness at end diastole), and abnormal blood pressure responses to exercise were associated with non-SCD.

Earlier studies had established the importance of non-SCD in children under 1 year of age, whereas SCD has been the focus of research in older children. The findings of this study show that further research is warranted to identify risk factors associated with non-SCD in older children. The study has also highlighted the difficulties in identifying predictors for SCD, which was the presenting symptom in four children.

At present, little is known about pediatric heart failure in the setting of preserved systolic function. Thus, the researchers plan to further investigate heart failure deaths in patients with hypertrophic cardiomypathy to gain insight into how this condition leads to more severe symptoms. Patients will be encouraged to adhere to treatment guidelines, including exercise restrictions. Those with excessive left ventricular wall thickness and abnormal blood pressure responses to exercise—risk factors identified for non-SCD—will be monitored more closely now to ensure that worsening symptoms are detected early and treated.

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