

“Sudden Cardiac Death in the Young (SCDY)” - 2018

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“Sudden Cardiac Death in the Young (SCDY)” was a paper written in June 2016 by Dr James Yeh MD PhD commenting on a prospective study of SCD among children and young adults published in NEJM (**Bagnall RD, Weintraub RG, English J, et al. A Prospective Study of Sudden Cardiac Death among Children and Young Adults. N Engl J Med. 2016 Jun 23;374(25):2441-52**

Fatal heart conditions are shocking and tragic when they are sudden and unexpected, and especially when they occur in the young. Occasionally the public is reminded of this via a news story of a high school or professional athlete who collapses suddenly and dies during a sporting event. Often, the first manifestation of a serious heart condition in the young is also the last.

What are common causes of sudden cardiac death in the young?

A number of structural and arrhythmogenic heart conditions are associated with sudden cardiac death. The list includes hypertrophic, dilated, and arrhythmogenic right ventricular cardiomyopathies, myocarditis, congenital long-QT syndrome, Brugada syndrome and catecholaminergic polymorphic ventricular tachycardia. Most of these conditions have a genetic basis.

What is this study about?

Bagnal et al, present their findings from a prospective population-based observational study evaluating the incidence and cause of sudden cardiac death in individuals ages 1-35 years in Australia and New Zealand during a 2-year period from 2010 to 2012. The investigators collected autopsy information, demographic information, and clinical data, including genetic and laboratory information, about the deceased.

How is sudden cardiac death defined?

Various criteria have been used to define sudden cardiac death in the medical literature. However, the investigators defined sudden cardiac death as a sudden unexpected death in an otherwise healthy person within one hour of symptom onset, or within 24 hours of being seen well by others. Sudden unexplained death was defined as sudden cardiac death without clear cause of death identified after a complete and comprehensive autopsy examination.

What did they find?

Four hundred ninety sudden cardiac death cases were identified. The mean age was 24 years with a male predominance of slightly greater than 70% of the cases. Nearly 40% of the individuals died during sleep. The annual incidence was 1.3 per 100,000 persons aged 1-35 years. The age group at highest risk for sudden cardiac death was those in the 31-35 years group (3.2 per 100,000).

Forty percent of the cases had structurally normal hearts and were designated sudden unexplained deaths. The most common explained causes of sudden cardiac death were coronary artery disease (24%) and inherited cardiomyopathies (16%). For all age subgroups, unexplained death was the most common finding except for those 31-35 years, where coronary artery disease was most common.

In nearly 30% of the sudden unexplained death cases, a clinically relevant cardiac gene mutation was identified. During follow-up, a clinical diagnosis of an inherited cardiovascular disease was identified in 12% of the families.

What is my take-away?

Unexplained sudden cardiac deaths in the young account for 40% of the cases. While we often think about coronary artery disease as a disease occurring in the older population, nearly 25% of the cases in this study had coronary artery disease leading to sudden cardiac deaths with the highest risk in those in the older age group (31-35). Genetic testing was able to identify a clinically relevant cardiac gene mutation for nearly one-third of the sudden unexplained death cases and increased the likelihood of identifying a possible cause of death in individuals over autopsy alone. According to NEJM Deputy Editor Dr. John Jarcho, *“This study suggests that genetic testing is a useful addition to standard autopsy procedures in cases of sudden cardiac death in the young. However, there still remain many such cases for which no cause can be found.”*

