EDITORIAL

Approach to unexplained sudden death in the young: proactive during life and prospective at death

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This editorial refers to ‘Sudden cardiac death in 14- to 35-year olds in Ireland from 2005 to 2007: a retrospective registry’ by R. Margery et al., on page 1411.

From a societal perspective it is difficult to reconcile the occurrence of sudden and unexpected deaths among our young, especially in ostensibly healthy and active individuals with no prior warning. Even today, we must acknowledge that these situations also present a significant challenge to the clinician as well as the scientist, particularly since many of these cases remain unexplained. The paper from Margery et al.1 published in the current issue of Europace is one among a series of reports highlighting the conspicuously large subgroup of cases with sudden death and structurally normal heart in the young, also termed the sudden arrhythmic death syndrome. The authors, reporting results from a national retrospective post-mortem analysis of sudden cardiac deaths (SCDs) in the 15–34 years age group in the Republic of Ireland, have done justice to comparisons with existing studies. However, there is room for a brief conversation regarding two important messages from their findings as well as a comment on the methodology used to perform the study.

As expected for the age group of interest, SCD with structurally normal heart was the most common post-mortem diagnosis (27%), followed by coronary artery disease (21%) and hypertrophic cardiomyopathy (15%). However, what are we to make of the finding that at least 10% of all subjects had idiopathic left ventricular hypertrophy (LVH)? In fact similar findings have been reported from other autopsy series in both younger and older populations.2 The association between LVH and overall mortality as well as SCD has long been acknowledged but a direct causal relationship with SCD has been debated. Emerging data from community-based studies suggest that LVH or increased ventricular mass has at least some pathways to ventricular arrhythmogenesis and SCD that may be distinct from those involved with severe left ventricular systolic dysfunction.3 The significant prevalence of isolated, unexplained LVH in young individuals who suffer SCD is further evidence that when it comes to risk of fatal arrhythmia there is at least a subset of idiopathic LVH that is not to be trifled with. It is possible that this form of high-risk LVH distinct from hypertrophic cardiomyopathy could be identified by novel imaging or other methodologies but this form of screening does not exist at the present time. How then do we unravel the complexities of LVH in the young, especially when there could be overlap with ‘athlete’s heart’ and associations with multiple conditions such as early hypertension, sleep apnoea and renal disease?2 The study by Margery et al.1 was not designed for this purpose, but these issues will require systematic dissection in large studies. The possibility also remains that unexplained LVH is a bystander among subjects with familial arrhythmogenic syndromes and molecular autopsies should be performed, but especially in younger individuals, it still needs to be explained. In the meantime, there is a considerable literature that has long announced that hypertension and resultant LVH are now prevalent in much younger individuals in epidemic proportions likely due to the rising tide of obesity and the metabolic syndrome.5 This represents a call to action for health care providers for due diligence in the paediatric population with screening for hypertension and LVH especially prompted by a positive family history or diagnosis of obesity.6

The now undeniable finding from multiple and diverse populations that the largest subgroup of SCD in the young is comprised of individuals with sudden death and structurally normal heart merits urgent attention. It is likely that at least some of the unexplained SCD cases in the Margery et al.1 study were due to the primary arrhythmia syndromes, but since details of family history were not available and molecular autopsies were not performed we cannot comment on the prevalence of these conditions. While Margery et al. are to be commended for their efforts on a national scale, such retrospective efforts are unlikely to provide the comprehensive and complete evaluation of these patients.

The opinions expressed in this article are not necessarily those of the Editors of Europace or of the European Society of Cardiology.

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that is urgently needed. The overall approach must shift toward prospective studies in large populations. The Oregon Sudden Unexpected Death Study, now ongoing for 10 years is one proof of concept that such efforts are feasible.7,8 This study has been conducted prospectively in a community of ~1 million residents in the Portland, Oregon, USA metropolitan area as a collaborative project between cardiologists, the emergency medical services, local hospitals and the local coroner/medical examiner network, and continues to contribute clinical and genetic information regarding SCD occurrence at all ages.9 Furthermore, at a governmental level, public health policy needs to evolve such that the time-honoured coroner investigation incorporates the molecular autopsy10 as an integral part of the post-mortem examination.

In conclusion, the study from Margey et al.1 further highlights unexplained SCD as the most common form of SCD in the young with a significant subgroup having lone, idiopathic LVH. These difficult conditions will remain unsolved unless large prospective studies are conducted to comprehensively evaluate sudden deaths in the young learning from combining clinical and genetic information. While we await novel methods of predicting and preventing this devastating condition, health care providers need to be proactive and vigilant in the paediatric population with a special focus on childhood obesity, hypertension as well as a family history of high-risk disorders.

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