Cardiology in the Young (2017), **27(Suppl. 1)**, S1–S2
doi:10.1017/S1047951116002158

Preface

Prologue from the Guest Editor

Robert M. Campbell

Department of Pediatrics, Emory University School of Medicine, Atlanta, Georgia, United States of America

On the one hand, it is frustrating and disheartening to see that paediatric and young adult sudden cardiac death still occurs. On the other hand, however, much progress has been made over the past several decades to prevent these terrible events. Sudden cardiac death prevention can be classified into primary and secondary prevention strategies.

Primary prevention involves diagnosis, treatment, counselling, and appropriate restrictions to prevent sudden cardiac arrest. Below is a listing of only some of the recent advances in primary prevention:

- Education and awareness that warning signs and symptoms may help identify patients and families affected by rare cardiac disorders predisposing to sudden cardiac arrest.
- Standardised pre-participation evaluation forms and guidelines have been developed by the American Academy of Pediatrics and the American Heart Association, among others. These forms define a more comprehensive standardised template that emphasises history and physical examination.
- The role for mass electrocardiogram (ECG) screening of certain populations – for example, athletes, all paediatric-age patients, and neonates – continues to incite debate with the exact role for this screening technique still uncertain; however, this debate has led to the development of refined ECG criteria – the so called "Seattle Criteria" and the "Refined Criteria" – which may help enhance the accuracy of ECG screening, and specifically decrease the false-positive rate.
- The development of guidelines for athletic participation for individuals affected by cardiac disorders predisposing to sudden cardiac arrest have been recently described in a *2015 AHA/ACC Scientific Statement: Eligibility and Disqualification Criteria for Athletes with Cardiovascular Abnormalities*.
- Clinical genetic testing has evolved from the early days of research genetic testing, and is now widely and appropriately applied through paediatric and adult arrhythmia programmes and cardiology practices. The identification of disease causing class I genetic mutations associated with cardiomyopathies and channelopathies helps with diagnosis, prognostication, risk stratification, and patient management.
- The emerging role for genetic counsellors in association with arrhythmia and cardiomyopathy programmes enhances our ability to deliver accurate and timely cascade testing for thorough patient and family evaluation and management.
- The development of new scientific statements and guidelines, such as the Pediatric and Congenital Electrophysiology Society's consensus statement on asymptomatic young patients with Wolff–Parkinson–White and guidelines and on the management of children with catecholaminergic polymorphic ventricular tachycardia, increases our knowledge about appropriate risk stratification and clinical care decisions.
- Recent research and resultant publications define new strategies for the management of difficult patient populations including, but not limited to, long QTS type 3, catecholaminergic polymorphic ventricular tachycardia, Brugada syndrome, and hypertrophic cardiomyopathy. We have seen recent advancements in implantable cardioverter-defibrillator therapies, and the still evolving role for left cervical sympathetic denervation continues to hold promise for difficult patient subsets. Novel therapies such as wearable defibrillator vests may provide a temporary alternative to an implanted defibrillator.

Secondary prevention involves centres around emergency response, preventing sudden cardiac arrest

Correspondence to: R. M. Campbell, Department of Pediatrics, Emory University School of Medicine, 2835 Brandywine Road, Suite 300, Atlanta, GA 30341, United States of America. Tel: 404 785 1445; Fax: 404 785 1461; E-mail: campbellr@kidsheart.com

from becoming sudden cardiac death. The American Heart Association Chain of Survival, updated in 2015, describes the five-step process for out-of-hospital cardiac arrest. It begins with recognition and activation of the emergency response system and progresses rapidly to immediate high-quality bystander cardiopulmonary resuscitation (CPR), rapid defibrillation, and transport to cardiac care hospitals for basic and advanced emergency medical services and possibly advanced life support. The ultimate goal is hospital survival and discharge home, neurologically intact. Several key steps have occurred to advance secondary prevention, including the following:

- Collaboration among agencies such as the American Heart Association, Red Cross, Emergency Cardiovascular Care Update, and others have advanced education and awareness about sudden cardiac arrest emergency response throughout our country and within our healthcare provider networks.
- Data from registries such as the Cardiac Arrest Registry to Enhance Survival provide information about current-state outcomes and opportunities to provide more effective emergency response and patient management.
- Published data now document the impact of effective bystander CPR – using compressions-only CPR for witnessed cardiac arrest – and the appropriate timely application of automatic external defibrillation to enhance survival.
- Several communities such as Seattle have championed emergency preparedness strategies, resulting in dramatically increased survival from out-of-hospital cardiac arrest.
- School emergency action plans have been developed through efforts such as Project ADAM, and several states now have legislated school emergency action plans to provide for emergency response with CPR and automatic external defibrillations.

Organisations such as the Sudden Arrhythmic Death Syndrome Foundation and Parent Heart Watch tell devastating stories about sudden death in families; these stories drive us to improve primary and secondary prevention strategies. The National Institutes of Health and Centers for Disease Control recently instituted a joint Sudden Death in the Young Registry. This registry of deaths in young people from conditions such as heart disease and epilepsy will help investigators define the scope of the problem and understand the characteristics of youths who die suddenly. Through a better understanding of the epidemiology of these sudden death cases, we should be able to enhance feedback to our primary and secondary prevention strategies, lessening the chance that patients will suffer sudden cardiac arrest and sudden cardiac death.

Of course we have made progress, lots of progress; however, we continue to experience sudden cardiac death in our adolescents, children, and young adults, indicating that we have not solved this problem. The future holds promise, with the hope of even more refined genetic identification of affected patients and families, clarification of the variant of unknown significance conundrum, and potentially even the definition of 'no risk' subsets within disorders such as long QT syndrome and hypertrophic cardiomyopathy. Our basic researchers are looking to develop curative gene therapies and novel drugs to eliminate/minimise the risk of sudden cardiac arrest. In the meantime, we need to continue with a widespread adoption of emergency action plans and public-access defibrillation programmes.

Many hospitals now have quality and safety programmes that use mission or vision statements such as 'one is not zero'. Although no primary or secondary prevention strategy likely will ever be perfect, our goal should be zero paediatric and young adult sudden cardiac deaths. Our committed drive to perfection, which may not be achievable, will always lead us towards excellence.

The manuscripts included in this monograph are the result of a conference held in February, 2016, entitled 'Wolff-Parkinson-White Syndrome and Other Causes of Sudden Cardiac Arrest in the Young: The Fouad Mobassaleh Memorial Symposium' sponsored by Nicklaus Children Hospital in Miami, Florida, and by the Mobassaleh family. These manuscripts detail many of the important diagnostic and treatment aspects of the cardiac disorders predisposing to paediatric and young adult sudden cardiac arrest. Although current science helps establish published guidelines for many of these cardiac disorders, there is still plenty of uncertainty and controversy that mandates the use of the 'art of medicine' in the management of affected patients. It is the hope of all involved that multicentre collaboration will continue to advance the science, resulting in improved patient diagnostics and care delivery.

https://doi.org/10.1017/S1047951116002158 Published online by Cambridge University Press