J Wave Syndromes

Brugada and Early Repolarization Syndromes

Charles Antzelevitch Gan-Xin Yan *Editors*



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We dedicate this book to the members of the Heart Rhythm Society (HRS), the European Heart Rhythm Association (EHRA), and the Asian-Pacific Heart Rhythm Society (APHRS) who met in Shanghai, China, in April 2015 to generate an expert consensus document and formulate this text focused on the J wave syndromes. We are also proud to dedicate this compendium to the many investigators whose collective works have advanced us to this exciting juncture in the history of cardiac electrophysiology and arrhythmias, on whose shoulders we stand, as well as to our mentors, collaborators, and fellows who have assisted us in advancing the field. Last but certainly not least, we dedicate this book to our families, whose understanding and support permitted us to dedicate the time and effort needed to formulate this text.

> Charles Antzelevitch Gan-Xin Yan

Foreword

Sudden cardiac death remains the most important public health issue on the planet. Despite intense research efforts and remarkable progress in heart disease populations, we have not been able to impact its overall incidence in the community where the vast majority of victims have not been previously identified as being at risk. Particularly tragic is its continued occurrence in young and apparently healthy people. Although sudden death is far less common in this population, it is an unspeakable calamity, truncating productive lives, widowing spouses, and orphaning children around the world.

It is now clear that a particularly important mechanism for sudden death in the young is some form of the newly defined J wave syndromes. Though knowledge regarding these entities has advanced rapidly, many challenges remain. Drs. Antzelevitch and Yan, outstanding leaders in this area of research, convened an expert panel to review the state of the art in order to identify those issues requiring better definition. These include ionic mechanisms, genetics, and, most importantly, methods of diagnosis, risk stratification, and treatment that can be applied on a global basis. The aim of the exercise was to mark those areas in which research and educational methods can be most efficiently and expeditiously directed.

It is fair to say that this elite group of scientists succeeded beyond expectations. Not only have they produced an extremely valuable consensus document, but they have also put forth this book that provides amplification of the most complex issues. Most importantly, the information contained herein will help resolve confusion that exists in the clinical community and has led to inappropriate, ineffective, and frequently over-aggressive treatment.

As they have perennially, my colleagues Drs. Antzelevitch and Yan lead the way in helping us understand how we can translate learnings in the basic laboratory into the clinic, with the ultimate goal of mitigating, if not preventing, sudden cardiac death. For their stewardship of this Herculean effort, we are most grateful.

Wynnewood, PA, USA January 15, 2016 Peter R. Kowey, MD, FACC, FAHA, FHRS

Preface

The J wave syndromes, consisting of the Brugada (BrS) and early repolarization syndromes (ERS), have captured the interest of the cardiology community for over two decades, following the identification of BrS as a new clinical entity in 1992. Although experimental evidence for a heightened risk for sudden cardiac death (SCD) associated with early repolarization was reported at the turn of the century, the clinical impact of ERS was not fully appreciated until 2008 with seminal reports on the subject from the groups of Haissaguerre, Viskin, and Nam. A book dedicated to the Brugada syndrome was published by us and the Brugada brothers in 2005, emanating from the expert consensus conference dedicated to BrS in 2004. A book specifically focused on ERS has not previously been published other than the expert consensus document dealing with terminology of early repolarization by MacFarlane and coworkers.

This book stemmed from an expert consensus conference forum organized in 2015 to evaluate new information and highlight emerging concepts with respect to differential diagnosis, prognosis, and cellular and ionic mechanisms and approaches to the therapy of the J wave syndromes. Leading experts, including members of the Heart Rhythm Society (HRS), the European Heart Rhythm Association (EHRA), and the Asian-Pacific Heart Rhythm Society (APHRS) met in Shanghai, China, in April 2015. The task force was charged with a review of emerging concepts and assessment of new evidence for or against particular diagnostic procedures and treatments.

Members of this Task Force were selected to represent professionals involved with the medical care of patients with the J wave syndromes as well as those involved in research into the mechanisms underlying these syndromes. Selected experts in the field undertook a comprehensive review of the literature. Critical evaluation of methods of diagnosis, risk stratification, approaches to therapy, and mechanistic insights was performed, including assessment of the risk-benefit ratio.

This text is designed to supplement and complement the expert consensus document that will be simultaneously published dealing with the J wave Syndromes: Emerging Concepts and Gaps of Knowledge.

Wynnewood, PA, USA Wynnewood, PA, USA Charles Antzelevitch Gan-Xin Yan

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