Preface

Much has changed in the field of arrhythmias and sudden cardiac death in these last decades. Successful innovative catheter therapies and protective devices have been determinant in enhancing treatment and prevention strategies of individuals at risk. However, despite the advances, sudden cardiac death still remains a major contributor to mortality in our society. While most deaths occur in adult cases and are associated with ischemic heart disease, occasionally the youngest and the fittest, even those who have become our role models for their athletic abilities, may also die suddenly, usually from noncoronary cardiac causes. It has not been until the advent of molecular biology and genetics in cardiology when we have been able to further deepen in our knowledge of these dreadful events in the young.

In the last 20 years, genetic research in subjects and families with sudden cardiac death syndromes has brought a vast amount of information on genetic defects responsible for arrhythmogenesis, improving our understanding on how the abnormally codified proteins are involved in the pathogenesis of a disease and how this protein disrupts the myocyte electrical activity, generates a chaotic rhythm, and predisposes to ventricular fibrillation.

Inherited sudden cardiac death syndromes are indeed rare diseases, much rarer than hypertension or coronary artery disease. However, it is highly likely that as physicians we will at some point encounter a patient with one of these genetic diseases, and we have to be aware of at least two clinical implications.

First, the field of cardiac genetics has brought a new tool, genetic screening, which is presently standing out as a key diagnostic test, complementing the highly sophisticated, but often inaccurate, clinical instruments. With the use of genetic information in our practice, we have moved the information from the bench to the bedside, from research to clinical care, translational medicine at its best.

Second, cardiac genetics is also bringing a fundamental change for our clinical practice, which is not to be taken lightly. With the care for patients with inherited arrhythmias, we have gone from facing the single patient to facing the family, from one individual with signs and symptoms of a disease to several family members with a genetic defect. Familial global care is a tremendous and complex new task that includes genetic screening, treatment decisions especially difficult in children, childbearing choices, disease expression, and genetic penetrance. The family, with all its complexity, cannot be assumed by the lone physician but only by a multidisciplinary team of geneticists, cardiologists, psychologists, and genetic counselors.

Most cardiologists already appreciate that there is more to the sudden death of a young individual than just "natural" causes. Genetic information is changing the way we approach medical care in this genomic era. With this book, our goal is to provide

the latest information on sudden cardiac death and genetic syndromes, with the aim to guide the physician in this complex field.

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