Epidemiology and clinical aspects of sudden cardiac death in the young
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Citation for published version (APA):
van der Werf, C. (2013). Epidemiology and clinical aspects of sudden cardiac death in the young

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1 Outline of the thesis
The first part of this thesis addresses the epidemiology and clinical aspects of sudden death in the young with a focus on inherited cardiac diseases. These play a significant role in sudden death in the young, as detailed in Chapter 2. Chapters 3 and 4 present the rational, design and main results of the CAREFUL study. This study was undertaken to prospectively investigate the usual care after sudden death in the young aimed at identifying inherited cardiac disease as the cause of death, and to assess the efficacy of two interventions to improve this usual care. Chapter 5 reports the findings of a focus group study among general practitioners and coroners on factors that hinder the recommended strategy of autopsy and family evaluation in young sudden death cases. Chapter 6 delineates the information obtained by in-depth interviews with relatives from young sudden death victims who attended a cardiogenetics clinic. In Chapter 7, we describe the incidence, causes and outcome of out-of-hospital cardiac arrest in children. Chapter 8 reports on the yield of cardiologic and genetic examination of first-degree relatives of young sudden death victims who are autopsy-negative or in whom no autopsy is performed. While inherited cardiac diseases are identified in approximately a third of families, we present the outcomes of long-term follow-up in the diagnosis-negative families in Chapter 9.

In Part 2 we continue with a focus on catecholaminergic polymorphic ventricular tachycardia (CPVT). This rare inherited arrhythmia syndrome is characterized by adrenergically-mediated polymorphic ventricular tachyarrhythmias and linked to mutations in the cardiac ryanodine receptor in the majority of cases, as reviewed in Chapter 10. The clinical and electrophysiological characteristics of carriers of a mutation in the cardiac ryanodine receptor, identified by predictive genetic testing, are presented in Chapter 11.

In Chapters 12 and 13 the exciting new discovery of the efficacy of the antiarrhythmic drug flecainide in patients with CPVT is further explored in mutation-positive and mutation-negative patients, respectively.

Finally, Part 3 concentrates on new clinical diagnostic methods for the long-QT syndrome (LQTS), the most prevalent inherited arrhythmia syndrome. Chapter 14 presents an algorithm based on the corrected QT interval during exercise testing for prediction of genetic testing in relatives of patients with LQTS. The diagnostic accuracy of the so-called “standing-test” in patients with possible LQTS is explored in Chapter 15.

The thesis is summarized in Chapters 16 and 17 in English and Dutch, respectively.