

STATE-OF-THE-ART REVIEW

# Management of Arrhythmias in Cardiac Amyloidosis

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## ABSTRACT

Cardiac amyloidosis is characterized by extracellular protein fibril deposition in the myocardium leading to restrictive heart failure. Both atrial and ventricular arrhythmias, along with conduction disease, are common in cardiac amyloidosis, and are often highly symptomatic and poorly tolerated. Many commonly used therapeutics such as beta-blockers, calcium-channel blockers, and digoxin may be poorly tolerated and lead to clinical decompensation in this population, adding complexity to the co-management of these conditions. In addition, studies have shown that atrial fibrillation with cardiac amyloidosis carries a high risk of stroke and systemic embolism, making anticoagulation indicated in all patients regardless of CHA<sub>2</sub>DS<sub>2</sub>-VASc score. Ventricular arrhythmias are common, whereas an implantable cardioverter-defibrillator has not been shown to improve survival. Conduction disease is also common and permanent pacemaker placement is often needed. High-quality evidence and guideline recommendations are limited with regard to the management of arrhythmias in cardiac amyloidosis. Providers are often left to clinical experience and expert consensus to aid in decision-making. In this focused review, we outline current guideline recommendations, summarize both historical and contemporary data, and describe evidence-based strategies for managing arrhythmias and their sequelae in patients with cardiac amyloidosis. (J Am Coll Cardiol EP 2020;6:351-61) © 2020 by the American College of Cardiology Foundation.

Amyloidosis is characterized by extracellular protein fibril deposition throughout the body's organ systems (1). When involving the myocardium, it can often lead to a restrictive cardiomyopathy. Approximately 95% of cardiac amyloidosis is misfolded transthyretin (ATTR) or immunoglobulin light-chain (AL) types (2-4). The ATTR group is characterized by misfolding of transthyretin, a protein made in the liver. Both wild-type ATTR cardiomyopathy (wtATTR-CM) and hereditary transthyretin cardiomyopathy (hATTR-CM) have been described. The prevalence of wtATTR-CM is

estimated to be between 5.5% and 16.0% of people age >80 years of age, whereas hATTR-CM may be rarer and prevalence estimates range by specific gene mutation (5). AL amyloidosis is a clonal plasma cell proliferative disorder characterized by production of abnormal protein fibrils in the bone marrow. The estimated prevalence of AL amyloidosis in the United States ranges from 15.5 to 40.5 cases per million (4,6,7). Overall, cardiac amyloidosis is thought to be an under-recognized cause of heart failure. Recent trends have revealed a significant increase in the number of new diagnoses of ATTR-CM

From the Cardiac Electrophysiology Section, Division of Cardiology, Department of Medicine, University of California, San Diego, La Jolla, California. Dr. Hsu has received honoraria from Medtronic, Abbott, Boston Scientific, Biotronik, Amgen, Bristol-Myers Squibb, Alkermes Pharmaceuticals, and Biogen-Webster; has received research grants from Biogen-Webster and Biotronik; and has equity interest in Acute Medical and Vector Medical. Dr. Urey was a member of the Speakers' Bureau for Alkermes, Alnylam, and Pfizer, and has received honoraria from Alkermes Therapeutics, Alkermes Pharmaceuticals, and Pfizer. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose. The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the JACC Clinical Electrophysiology authors instructions page.

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**George Edward Billman**



## **Arrhythmias Therapies Arrhythmias Case Studies In Management Software Windowssingle User:**

**Books In Print 2004-2005** Ed Bowker Staff, Staff Bowker, Ed, 2004      Arrhythmias Edward P. Hoffer, G. O.

Barnett, 1998-01-01 The Arrhythmias Case Studies Program is designed to give the user practice in recognizing and treating a variety of common serious arrhythmias. It is aimed at the practicing internist and cardiologist as well as at family practitioners with an interest in cardiology and assumes a basic knowledge of ECG interpretation. Emphasis is on integrating clinical and ECG data to arrive at a rhythm diagnosis and etiology and then on selecting appropriate treatment. In each case the simulated patient may respond to therapy in predictable but not necessarily desirable ways. Included in the library of cases are patients with sick sinus syndrome in its varied manifestations, complex arrhythmias in the setting of myocardial infarction and supraventricular tachycardias. Windows Compatible      **Arrhythmias** Edward P. Hoffer, 1985-01-01 Assists in

making diagnostic management decisions in order to save the simulated patients as they experience a variety of cardiac arrhythmias      **Management of Cardiac Arrhythmias** Gan-Xin Yan, Peter R. Kowey, 2010-09-27 Few areas of medicine have evolved as rapidly as cardiac electrophysiology. What were only a short time ago seen to be lethal rhythm disturbances can now be treated with confidence in a diverse spectrum of patients. The first edition of Management of Cardiac Arrhythmias published over ten years ago has served clinicians not only as a practical guide to cardiac arrhythmias but also as a comprehensive reference source. The second edition builds upon the concise style and expert authorship of its predecessor to provide the most up to date information on the diagnosis and treatment of this group of diseases. The introductory chapters begin with historical perspectives of the field and move on to discuss the scientific basis of arrhythmogenesis and diagnostic testing. The book then devotes specific chapters to various arrhythmias including technical innovations in treatment and insights from clinical trials of and current guidelines for permanent pacemakers and implantable cardioverter defibrillators. Subsequent chapters focus on arrhythmias in specific populations including athletes, children and women during pregnancy. Syndromes such as syncope, long and short QT syndrome and J wave syndromes are also covered. Presenting complex information in a clearly structured and efficient format, this book is an incomparable asset to cardiologists and other physicians and health care professionals involved in the treatment of patients with cardiac arrhythmias      Arrhythmias

Krieger Publishing Company, 1994-09-01      **Arrhythmias Case Studies** Lester B. Jacobson, Nora Goldschlager, 1978

**Arrhythmias Case Studies** C. T. Kappagoda, 1985      Cardiac Arrhythmias M. Schlepper, B. Olsson, 2012-12-06 Asked to organize an international meeting on Propafenone Ryt monomer we had to question ourselves whether to accept this offer just as interested physicians or in our capacity as chairmen of the study group on arrhythmias and intracardiac electrography of the European Society of Cardiology. Considering the pros and cons and well aware that one can easily be regarded as product promoter we nevertheless choose to organize the Symposium in association with the study group. We felt justified to do so for a number of reasons. Arrhythmias and their treatment as well as the tools to detect the first and to control the latter

are well within the scope of the study group An international symposium provided the opportunity to have the state of the art in arrhythmias and antiarrhythmic treatment thoroughly reviewed by internationally renowned investigators thus serving an educational purpose Furthermore it was the intention and aim of the meeting to have what is known about a fairly new drug presented in a critical manner and to point out what is not known but should be known It is in that context the presentations of the free session should be judged To the careful reader it will become apparent that much which was not said can be read between the lines

*Cardiac Arrhythmias* Ambrose S. Kibos, Bradley P. Knight, Vidal Essebag, Steven B. Fishberger, Mark Slevin, Ion C. Tintoiu, 2013-12-13 This book covers all the major aspects associated with pathophysiological development of cardiac arrhythmias covering enhanced or suppressed automaticity triggered activity or re entry from basic concepts through disease association limitations of current pharmacotherapy and implant therapies and on going trials and analysis of new biomarkers based on current knowledge of cellular interaction and signalling The book describes novel and state of the art methods for differentiating between the major types of arrhythmia structural abnormalities and current practice guidelines and determination of risk stratification associated with sudden cardiac death A particular focus is on arrhythmias associated with atrial fibrillation and includes details of associations with cardiac disease current detection analysis and imaging and future perspectives

*Cardiac Arrhythmias and Device Therapy: Results and Perspectives for the New Century* I. Eli Ovsyshcher, MD, PhD, FESC, FACC, 2000-06-15 The final decades of this century have been a period of expansive growth for the electrophysiological management of cardiac arrhythmias Subsequently the rapid and widespread clinical application of so many technological advances has made the exchange of ideas in this field an urgent necessity *Cardiac Arrhythmias and Device Therapy Results and Perspectives for the New Century* addresses the issue by gathering current relevant information in a clear user friendly format This book covers a broad spectrum of subjects including basic electrophysiology impulse propagation subthreshold stimulation genetics and the molecular base of arrhythmias invasive and noninvasive clinical electrophysiology and the clinical aspects of cardiac pacing and defibrillation It provides a comprehensive review of various methods of treatment of atrial fibrillation and of the role of pacing in prevention of atrial fibrillation and in treatment of congestive heart failure dilated hypertrophic and restrictive cardiomyopathies The results of recent clinical trials in cardiac pacing and in the prevention and treatment of malignant ventricular arrhythmias are also presented Special sections have been dedicated to the various aspects of preventing sudden death including the use of internal and external defibrillation Finally the book offers a current review of hot topics with personal experiences and critical evaluations by the world renowned authors This book will be of interest to all cardiologists EP technicians and health care personnel involved in the diagnosis and management of arrhythmias who wish to gain a complete perspective on modern pacing and electrophysiology

*Basis and Treatment of Cardiac Arrhythmias* Robert E. Kass, Colleen E. Clancy, 2005-09-29 This book incorporates the most up to date information related to mechanisms and treatment of cardiac arrhythmia Many of the topics discussed in this

text reflect very recently undertaken research directions including genetics of arrhythmias cell signalling molecules as potential therapeutic targets and trafficking to the membrane These new approaches and implementations of anti arrhythmic therapy derive from many decades of research as outlined in the first chapter by distinguished Professors Michael Rosen Columbia University and Michiel Janse University of Amsterdam The text covers changes in approaches to therapy over time arrhythmias in multiple cardiac regions and over many scales from gene to protein to cell to tissue to organ Mechanisms and Treatment of Cardiac Arrhythmias H. Joseph Reiser, Leonard N. Horowitz, 1985

**Clinical Case Studies in Arrhythmia** Philip J. Podrid, 1994 Contains a compilation of case studies to demonstrate the diagnosis and management of arrhythmias Also contains questions and a discussion about each of the case studies

**Novel Therapeutic Targets for Antiarrhythmic Drugs** George Edward Billman, 2010-01-07 PROFILES POTENTIAL TREATMENT APPROACHES FOR CARDIAC ARRHYTHMIAS Cardiac arrhythmias of ventricular origin are responsible for the deaths of nearly half a million Americans each year while atrial fibrillation accounts for about 2 3 million cases per year a rate that is projected to increase 2 5 fold over the next half century Effectively managing these cardiac rhythm disorders remains a major challenge for both caregivers and the pharmaceutical industry Filling a gap in the current literature Novel Therapeutic Targets for Antiarrhythmic Drugs presents the latest treatments for cardiac arrhythmias alongside comprehensive presentations of basic cardiac physiology and pharmacology Written by leading experts in their research areas this invaluable resource offers both practitioners and researchers a one stop guide that brings together previously dispersed information The text consists of four sections Section One comprehensively reviews basic cardiac electrophysiology the mechanisms responsible for arrhythmias in the setting of ischemia and basic pharmacology of antiarrhythmic drugs Section Two addresses safety pharmacology including the concept of repolarization reserve safety challenges and regulatory issues for the development of novel antiarrhythmic drugs Section Three describes several novel pharmacological targets for antiarrhythmic drugs including both ion channel and non ion channel targets Section Four describes promising non pharmacological antiarrhythmic interventions including selective cardiac neural disruption or nerve stimulation aerobic exercise training and diet omega 3 fatty acids Offering an unparalleled look at the current state and future direction of cardiac arrhythmia treatment Novel Therapeutic Targets for Antiarrhythmic Drugs provides an important resource to advanced students working researchers and busy professionals alike

*Clinical Cases in Cardiac Electrophysiology: Supraventricular Arrhythmias* Lucian Muresan, 2022 This is the first of a three volume project aimed at providing unique case reports 20 cases per volume of a large number of supraventricular and ventricular arrhythmias encountered in clinical practice The book is focused on the treatment of supraventricular arrhythmias namely focal atrial tachycardias typical and atypical AVNRT and accessory pathways both manifest and concealed All presented cases were performed using the CARTO electro anatomical mapping system which allows the reader to better understand the arrhythmia features due to the high number of images provided A large number of

high quality figures which represent the core of the authors work enrich the contents all cases are built around suggestive figures acquired during the patients hospitalization which clearly illustrate important concepts used in catheter ablation of cardiac arrhythmias The figures are not only related to the catheter ablation procedure but also to the patient history thus helping the reader to place the invasive treatment of the arrhythmias into clinical context electrocardiograms Holter ECGs signal averaged ECGs chest X Rays transthoracic and transesophageal echocardiography images MRI CT scans coronary angiography images are provided when considered relevant in order to better present the patient s condition Short videoclips with activation maps of the atria or the ventricles during the studied arrhythmia complete the information provided Teaching oriented all chapters include questions and answers and key messages at the end of each case For this reason it will be an invaluable tool for cardiologists clinical cardiac electrophysiologists and interventional cardiac electrophysiologists in training but also for all those interested in learning more about the subject     Cardiac Arrhythmias: Interpretation, Diagnosis and Treatment, Second Edition George J. Klein, Eric N. Prystowsky, 2019-09-22 The acclaimed primer of electrophysiology from two leading experts Cardiac Arrhythmias Interpretation Diagnosis and Treatment Second Edition is a practical clinical guide for healthcare professionals who seek a basic knowledge of electrophysiology and how to interpret and treat patients with arrhythmias One of the main benefits of this book is the author s ability to take electrophysiology and break it down clearly and simply so internists and family practitioners who do not have an extensive background in this complex aspect of cardiology can understand it and its relationship to treating arrhythmias The Second Edition has been updated to include the latest treatment advances new and important clinical trials additional cases and coverage of new drugs and devices and is enhanced by full color tracings and an increased number of tables and illustrations     **Clinical Management of Cardiac Arrhythmias** G. Neal Kay, Rosemary S. Buben, 1992 A comprehensive discussion of arrhythmia diagnosis and treatment focusing on the principles indications and applications of electrophysiologic techniques in the treatment of specific arrhythmias Is presented in this excellent text Topics include programmed electrical stimulation interventional electrophysiology temporary and permanent antitachycardia and rate adaptive pacing techniques signal averaging and much more Patient care and nursing management are integrated throughout the text     **Arrhythmia Treatment - Medications, Procedures, and Devices** Recardim Smith Evalina, 2024-07-31 Arrhythmia Treatment Medications Procedures and Devices Discover the essential guide to managing arrhythmias with Arrhythmia Treatment Medications Procedures and Devices This book provides a clear overview of the treatment options available for arrhythmias including medications medical procedures and devices In this book you ll explore Medications Learn about antiarrhythmic drugs anticoagulants and other medications used to manage arrhythmias including their functions and side effects Medical Procedures Understand key procedures such as electrophysiology studies catheter ablation cardioversion and pacemaker or ICD implantation Devices Explore the use of pacemakers and ICDs in treating arrhythmias and maintaining heart rhythm

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 Resources Access information on support systems and resources available for individuals with arrhythmias Arrhythmia  
 Treatment Medications Procedures and Devices offers a concise and practical guide to understanding and managing heart  
 rhythm disorders     Cardiac Arrhythmias Wilbert S. Aronow, 2014-02-12 This book is useful for physicians taking care of  
 patients with cardiac arrhythmias and includes six chapters written by experts in their field Chapter 1 discusses basic  
 mechanisms of cardiac arrhythmias Chapter 2 discusses the chronobiological aspects of the impact of apnoic episodes on  
 ventricular arrhythmias Chapter 3 discusses navigation detection and tracking during cardiac ablation interventions Chapter  
 4 discusses epidemiology and pathophysiology of ventricular arrhythmias in several noncardiac diseases methods used to  
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 arrhythmias including indications for implantation of an AICD for primary and for secondary prevention in patients with and  
 without congestive heart failure Chapter 6 discusses surgical management of atrial fibrillation     Cardiac Arrhythmias 2005  
 Antonio Raviele, 2006-07-08 The year 1988 marked the beginning of the International Workshop on Cardiac Arrhythmias This  
 biannual series of meetings was initiated with the following goals 1 to present technological advances in the field of cardiac  
 arrhythmias 2 to publicise the results of current research and 3 to assess the impact of new diagnostic and therapeutic  
 approaches In addition by bringing together experts in this field controversial aspects in the diagnosis and treatment of  
 cardiac arrhythmias could be discussed allowing a consensus to be reached regarding the evaluation and management of  
 specific disease conditions The success achieved in reaching these goals and the utility of the workshops have been confirmed  
 by their increasing recognition and level of attendance The Proceedings of the Ninth Edition of the Workshop is a compilation  
 of the topics presented at the most recent meeting which was held in Venice at the Fondazione Giorgio Cini from the 2nd to  
 the 5th of October 2005 The book is divided into eight sections each addressing a different aspect of cardiac arrhythmia  
 Supraventricular Arrhythmia and Atrial Flutter Atrial Fibrillation Pathophysiology Clinical and Therapeutic Aspects Atrial  
 Fibrillation Catheter Ablation and Other Non pharmacological Therapies Hereditary Arrhythmogenic Syndromes Sudden  
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