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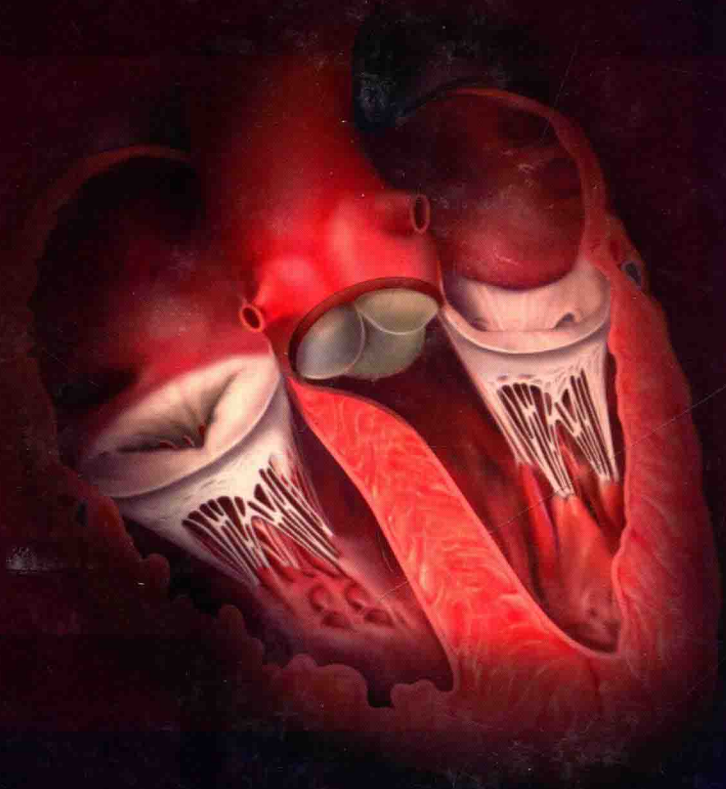
Valvular Heart Disease

A Companion to Braunwald's Heart Disease

FOURTH EDITION



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1600 John F. Kennedy Blvd.
Ste 1800
Philadelphia, PA 19103-2899

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CONTRIBUTORS

David H. Adams, MD
Marie-Josée and Henry R. Kravis Professor and Chairman
Department of Cardiothoracic Surgery
The Mount Sinai Medical Center
New York, New York

Thomas M. Bashore, MD
Professor of Medicine
Clinical Chief, Division of Cardiology
Duke University Medical Center
Durham, North Carolina

Helmut Baumgartner, MD
Professor of Cardiology/Adult Congenital Heart Disease
University of Muenster
Director, Division of Adult Congenital and Valvular Heart Disease
Department of Cardiovascular Medicine
University Hospital Muenster
Muenster, Germany

Alan C. Braverman, MD
Alumni Endowed Professor in Cardiovascular Diseases
Department of Medicine
Washington University School of Medicine
Saint Louis, Missouri

Ben Bridgewater, MB BS, PhD, FRCS (CTh)
Consultant Cardiac Surgeon
University Hospital of South Manchester
University of Manchester
Manchester Academic Health Science Centre
NICOR, UCL
London, United Kingdom

Charles J. Bruce, MBChB, FCP (SA)
Professor of Medicine, College of Medicine
Division of Cardiovascular Diseases
Mayo Clinic
Rochester, Minnesota

Blase A. Carabello, MD
The W.A. "Tex" and Deborah Moncrief, Jr. Professor of Medicine
Vice-Chairman, Department of Medicine
Baylor College of Medicine
Medical Care Line Executive
Veteran's Affairs Medical Center
Director, Center for Heart Valve Disease
St. Luke's Episcopal Hospital
Houston, Texas

John D. Carroll, MD
Professor of Medicine
University of Colorado Denver
Director, Interventional Cardiology
Medical Director, Cardiac and Vascular Center
University of Colorado Hospital
Denver, Colorado

Javier G. Castillo, MD
Department of Cardiothoracic Surgery
The Mount Sinai Medical Center
New York, New York

John B. Chambers, MD
Professor of Clinical Cardiology
Cardiothoracic Department
Guy's and St Thomas's Hospitals
London, United Kingdom

Heidi M. Connolly, MD
Professor of Medicine
Division of Cardiovascular Diseases
Mayo Clinic
Rochester, Minnesota

Arturo Evangelista, MD
Head of Cardiac Imaging Department
Director of Aortic Diseases Unit
Hospital Universitari Vall d'Hebron
Barcelona, Spain

Elyse Foster, MD
Professor of Medicine
University of California, San Francisco
Araxe Vilensky Endowed Chair
Cardiology
San Francisco, California

Benjamin H. Freed, MD
Assistant Professor
Division of Cardiology
Northwestern University Feinberg School of Medicine
Chicago, Illinois

Mario J. Garcia, MD
Chief of Cardiology
Professor of Medicine and Radiology
Co-Director, Montefiore-Einstein Center for Heart and Vascular Care
Montefiore Medical Center
Bronx, New York

Brian P. Griffin, MD
The John and Rosemary Brown Endowed Chair in Cardiovascular Medicine
Section Head, Cardiovascular Imaging
Cleveland Clinic
Cleveland, Ohio

Howard C. Herrmann, MD
Professor of Medicine
Perelman School of Medicine of the University of Pennsylvania
Director, Cardiac Catheterization and Interventional Cardiology
Hospital of the University of Pennsylvania
Philadelphia, Pennsylvania

Bernard Iung, MD
Professor of Cardiology
Cardiology Department
Bichat Hospital
Professor of Cardiology
University Paris 7 Diderot
Paris, France

Eric V. Krieger, MD
Assistant Professor of Medicine and Cardiology
University of Washington
Seattle, Washington

Amar Krishnaswamy, MD
Interventional Cardiology
Cleveland Clinic
Cleveland, Ohio

Roberto M. Lang, MD
Professor of Medicine and Radiology
Past President of the American Society of Echocardiography
Director, Noninvasive Cardiac Imaging Laboratories
Section of Cardiology
Department of Medicine
Chicago, Illinois

Grace Lin, MD
Assistant Professor
Division of Cardiovascular Diseases
Mayo Clinic
Rochester, Minnesota

Michael J. Mack, MD
Chairman, Cardiovascular Governance Council
Baylor Healthcare System
Chairman, Research Center
The Heart Hospital Baylor Plano
Dallas, Texas

S. Chris Malaisrie, MD
Assistant Professor of Surgery
Division of Cardiac Surgery
Northwestern University Feinberg School of Medicine
Chicago, Illinois

Patrick M. McCarthy, MD
Chief, Division of Cardiac Surgery
Director, Bluhm Cardiovascular Institute
Heller-Sacks Professor of Surgery
Northwestern University Feinberg School of Medicine
Chicago, Illinois

Jordan D. Miller, PhD
Assistant Professor
Mayo Clinic College of Medicine
Departments of Surgery and Physiology
Mayo Clinic
Rochester, Minnesota

Brad Munt, MD, FRCPC, FACC
Cardiologist
St. Paul's Hospital
Vancouver, British Columbia

Rick A. Nishimura, MD
Professor of Medicine
Cardiology
Mayo Clinic
Rochester, Minnesota

Kevin D. O'Brien, MD
Professor of Medicine
University of Washington
Seattle, Washington

Patrick T. O'Gara, MD
Director of Clinical Cardiology
Brigham and Women's Hospital
Professor of Medicine
Harvard Medical School
Boston, Massachusetts

David S. Owens, MD
Assistant Professor
Division of Cardiology
University of Washington
Seattle, Washington

Donald C. Oxorn, MD
Professor of Anesthesiology
University of Washington
Adjunct Professor of Medicine
Cardiology
University of Washington
Seattle, Washington

Rajni K. Rao, MD
Department of Medicine
Division of Cardiology
University of California, San Francisco
San Francisco, California

Raphael Rosenhek, MD
Department of Cardiology
Medical University of Vienna
Vienna, Austria

Ernesto E. Salcedo, MD
Professor of Medicine
University of Colorado Denver
Director of Echocardiography
University of Colorado Hospital
Denver, Colorado

Hartzell V. Schaff, MD
Professor of Surgery
Division of Cardiovascular Surgery Mayo Clinic
Rochester, Minnesota

David M. Shavelle, MD
Director, Cardiac Catheterization Laboratory
Director, Interventional Cardiology Fellowship
Los Angeles County + University of Southern California Medical Center
Associate Clinical Professor
Cardiovascular Medicine
University of Southern California
Los Angeles, California

Karen K. Stout, MD
Director, Adult Congenital Heart Disease Program
Associate Professor of Medicine and Cardiology
Adjunct Associate Professor of Pediatrics and Cardiology
University of Washington
Seattle, Washington

Pilar Tornos, MD
Professor of Cardiology
Servei de Cardiologia
Hospital Universitary Vall d'Hebron
Barcelona, Spain

Wendy Tsang, MD, FRCP(C)
Assistant Professor, Department of Medicine
Division of Cardiology
University Health Network–Toronto General Hospital
University of Toronto
Toronto, Ontario

Alec Vahanian, MD
Cardiology Department
Bichat Hospital
Paris, France

John G. Webb, MD
MacLeod Professor of Heart Valve Intervention
University of British Columbia
Director of Cardiac Catheterization
St. Paul's Hospital
Vancouver, Canada

FOREWORD

Valvular heart disease is an important clinical problem, responsible for an estimated 20,000 deaths and 100,000 hospitalizations each year in the United States alone. In recent decades valvular disease has been caught in two important cross-currents. The first is demographic. Despite the recent decline in the prevalence of rheumatic heart disease in North America, Western Europe, and Australia, the total number of patients with valvular heart disease in these regions is rising steadily because of the increase in degenerative valvular diseases that accompanies the aging of the population. The number of patients with valvular heart disease in developing countries is rising particularly rapidly. This is because the incidence of new cases of rheumatic heart disease has not (yet) fallen to the low levels observed in developed nations, but the numbers of elderly and accompanying degenerative valve diseases are increasing. About 17 million persons worldwide suffer from valve disease.

The second important cross-current relates to the changes in the diagnosis and management of valvular heart disease. Until relatively recently, the cardiac catheterization laboratory was the principal site at which the diagnosis and functional assessment of valvular heart disease were obtained, while the management of advanced valvular disorders took place in the operating room. Now noninvasive imaging techniques—echocardiography, including three-dimensional echocardiography, as well as cardiac magnetic resonance imaging and computed tomography—all provide rich anatomic and functional information. The cardiac catheterization laboratory is increasingly becoming the site of catheter-based correction of valvular disorders. This approach began 30 years ago with balloon mitral valvuloplasty and now involves growing efforts of transcatheter insertion of prosthetic aortic valves and corrections of severe mitral regurgitation.

The editors of *Valvular Heart Disease*, Drs. Otto and Bonow, are among the world's leaders in this field. They have selected outstanding authors, each an authority in the particular area that he or she covers. They discuss in depth the cross-currents mentioned above, which makes the understanding and management of valvular heart diseases more dynamic than ever. They also cover systematically the pathogenesis, pathophysiology, clinical findings, imaging, natural history, and therapeutic options. They describe challenges involved in the care of patients who have undergone valve replacement. There are new chapters on the epidemiology of valvular heart disease, transcatheter aortic valve implantation, imaging guidance of transcatheter valve procedures, and transcatheter mitral valve replacement and repair.

This fourth edition of *Valvular Heart Disease* is a classic, the leading textbook in the field, which builds on the previous editions. We congratulate the editors and authors for their important contributions and welcome this excellent book to our growing list of *Companions to Heart Disease*.

Eugene Braunwald, MD

Douglas L. Mann, MD

Douglas P. Zipes, MD

Peter Libby, MD

PREFACE

The scientific underpinnings, clinical evaluation, and treatment of valvular heart disease continue to advance at a startling rate. In the context of this rapidly expanding knowledge base, we are pleased to present the fourth edition of *Valvular Heart Disease: A Companion to Braunwald's Heart Disease,* which we believe will be a valuable, authoritative resource for practitioners of cardiology and surgery, physicians-in-training, and students of all levels.

In keeping with the previous editions of *Valvular Heart Disease*, the fourth edition covers the breadth of the field, providing the basics of diagnosis and treatment while highlighting new, exciting advances and their potential to transform outcomes for patients with heart valve disorders. With the help of our internationally recognized authors from the United States, Canada, and Europe, we have thoroughly revised this edition to keep the content vibrant, stimulating, and up-to-date. Eleven of the 27 chapters are entirely new, including 6 chapters that cover topics not addressed in earlier editions. We have added 24 new authors, all highly accomplished and recognized in their respective disciplines. The 16 chapters that have been carried over from the previous edition have been extensively updated by their authors, and new co-authors have been added to five of these chapters. These updated chapters cover topics ranging from diagnostic imaging to management of specific rheumatic, congenital, and degenerative diseases of the aortic valve, mitral valve, and right-sided valves.

The fourth edition follows the format of the previous edition. The initial section focuses on basic principles, epidemiology, mechanisms of disease, and diagnostic methods. This is followed by a second section covering aortic valve disease and a third covering mitral valve disease. The final section discusses diverse topics including intraoperative echocardiography, right-sided valve disease, endocarditis, prosthetic valves, and management of valvular heart disease during pregnancy.

Among the many enhancements found in the fourth edition are the four new chapters that open the basic principles section. These include a chapter on the global epidemiology of valvular heart disease by Drs. John Chambers and Ben Bridgewater, two entirely new chapters on the molecular mechanisms of calcific valve disease by Dr. Jordan Miller, and a chapter on the clinical, cellular, and genetic risk factors for calcific valve disease by Drs. Kevin O'Brien and David Owens. Drs. Roberto Lang, Wendy Tsang, and Benjamin Freed have together written a superb new chapter on the three-dimensional anatomy of the mitral and aortic valves; this chapter includes insights gleaned from their experience with the three-dimensional imaging of these structures.

New chapters also cover some of the more important aspects of valve disease diagnosis and management. Drs. Elyse Foster and Rajni Rao discuss the evaluation of and treatment options for the growing number of complex patients with secondary forms of mitral regurgitation stemming from ischemic left ventricular dysfunction (ischemic mitral regurgitation) and dilated cardiomyopathy (functional mitral regurgitation). Drs. Chris Malaisrie and Patrick McCarthy, coauthors of the chapter on mitral valve surgery in the last edition, have joined forces again in the current edition to write a comprehensive, expert chapter on surgical treatment of the aortic valve and ascending aorta. Dr. David Adams, coauthor of the chapter on aortic valve surgery in the last edition, has in this edition instead joined with Dr. Javier Castillo to write a superb chapter on surgical mitral valve repair and replacement. This latter chapter is enhanced by the new chapter on the important applications of intraoperative echocardiography during mitral valve surgery by Dr. Donald Oxorn.

New and updated chapters on transcatheter valve therapeutics cover this exciting and rapidly evolving field extensively. Because of the advent of transcatheter aortic valve implantation for patients who are at high risk for surgical valve replacement, Dr. John Webb has joined Dr. Brad Munt to update this important and topical chapter. Dr. Howard Herrmann has also joined our team of authors and contributed a new chapter on current and future transcatheter approaches to mitral valve repair and replacement. The transcatheter therapeutics discussions are further enhanced by two additional new and important chapters on this subject: Dr. Michael Mack has written a new chapter on the risk assessment of patients undergoing consideration for surgical versus transcatheter valve replacement or repair, and Drs. Ernesto Salcedo and John Carroll have provided an up-to-date view of the role of imaging in guiding the delivery of transcatheter valve devices and monitoring their results. These august authors, representing their respective fields of cardiac surgery, interventional cardiology, and cardiac imaging, have played an important role in the heart valve teams at their individual institutions. The expert commentary found in their chapters embodies the concept that such a collaborative, interdisciplinary valve team provides the needed expertise to make difficult management decisions regarding patients with complex illnesses and deliver the most appropriate treatments with optimal outcomes.

This edition of *Valvular Heart Disease* includes in the print version 388 full-color figures and 162 tables; additional figures and video content are available in the online version. The chapters continue to conform to current guideline recommendations of the American College of Cardiology/American Heart Association and the European Society of Cardiology/European Association of Cardio-Thoracic Surgery.

We are indebted to all of our authors for their commitment of considerable time and effort to ensure the high quality and authoritative nature of this edition of *Valvular Heart Disease.* We are also delighted that this book remains a member of the growing family of companion texts to *Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine.* As a member of the *Braunwald's* companion series, the book is also available online on the Expert Consult companion website. Figures and tables can be downloaded directly from the website for electronic slide presentations. In addition, there is a large portfolio of video content that supplements the print content of many of our chapters.

Despite the advances in diagnosis and treatments (both surgical and interventional), valvular heart disease remains a major cause of morbidity and mortality throughout the world. Rheumatic heart disease remains a scourge in developing countries, and congenital forms of aortic and mitral valve disease create a steady stream of young and middle-aged adults with aortic stenosis, aortic regurgitation, and mitral regurgitation in both developed and developing countries. The aging of the population in the United States and abroad results in an increasing number of elderly patients with degenerative forms of aortic stenosis and mitral regurgitation, who often present with age-related medical comorbidities that confound medical decision making. Unlike most other forms of cardiovascular disease, in which management decisions can be guided by the evidence base created by multiple large-scale randomized controlled clinical trials, the evidence base in valve disease is limited by a dearth of clinical trials.

In this field more than any other, expert clinical judgment and experience are the cornerstones of rational decision making and optimal patient management. We believe that the collective knowledge, experience, and expert clinical judgment of the accomplished authors of *Valvular Heart Disease* will serve as an invaluable resource for all of us who are called upon to provide care for our patients with these diseases.

Robert O. Bonow

Catherine M. Otto

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Sincere thanks are due to the many individuals who helped make this book a reality. In particular, we would like to express our deep appreciation to the distinguished chapter authors for their time and effort in providing excellent chapters. We also thank the publishing staff at Elsevier for their guidance and close working relationship. Finally and most important, we would like to thank our families for their constant understanding, encouragement, and support.

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CHAPTER 1

Epidemiology of Valvular Heart Disease

John B. Chambers and Ben Bridgewater

Key Points

- Rheumatic disease is the most common cause of valve disease worldwide, especially in the young, with an estimated prevalence of 15.6 to 19.6 million.
- Endomyocardial fibrosis is an underresearched disease common in equatorial Africa.
- In industrially developed regions valve diseases of old age predominate, particularly calcific aortic stenosis and functional mitral regurgitation.
- In the United States valve disease is most common in the elderly, with a prevalence of 13% in those older than 75 years.
- Drug-induced valve disease is increasing as a result of the use of 5-HT_{2B} receptor agonists.
- Infective endocarditis is increasingly related to medical devices and intravenous drug use.
- Failure of biological replacement valves is a major burden in all regions of the world.
- Substantial variation in access to health care exists in all countries, including those that are industrially developed.
- The main global challenge is to prevent chronic rheumatic disease, which will require collaborations among social, political, and medical programs.
- Valve care in industrially developed countries needs to be organized around specialist valve clinics that refer patients, as indicated, to specialist surgeons and interventional cardiologists.

Rheumatic fever is the most common cause of valve disease in the young,[1] but it predominates in industrially underdeveloped regions. These include Africa, India, the Middle East, South America, and parts of Australia and New Zealand, China, and Russia.[1] In developed countries, the incidence of rheumatic disease declined after the second half of the 20th century, although transient local resurgences still occur.[2] This decline was predominantly the result of improvements in living conditions and health care, as follows:

- Improvements in living conditions:[3]
  - Better housing to reduce overcrowding
  - Better nutrition
- Improved access to health care
- Treatment of streptococcal throat infections
- Use of secondary prophylaxis

In addition there was a spontaneous reduction in the virulence of streptococcal serotypes, but it occurred after the incidence of rheumatic fever had already fallen.[4]

These improvements in living conditions and health care have increased longevity so that valve conditions characteristic of old age now predominate (Figure 1-1). Some 2.5% of the U.S. population has moderate or severe valve disease, but the prevalence rises after age 64 (Figure 1-2) and is 13% in those older than 75.[5] Other studies confirm this age relationship.[6,7] The contribution of old age to the world prevalence of valve disease is difficult to estimate precisely but probably now rivals that of rheumatic disease (Table 1-1). The most common valve diseases in the elderly are:

- Calcific aortic valve disease
- Aortic dilation causing aortic regurgitation
- Functional mitral regurgitation as a result of left ventricular (LV) dysfunction

At the same time there has been a rise in new diseases induced by drugs or therapeutic irradiation. There has also been an increase in endocarditis related to drug use and device implantation. Reoperation for failing biological replacement valves is common in underdeveloped countries, where mechanical prostheses are avoided because of the difficulty of organizing anticoagulation. Reoperation is also a significant load in industrially developed regions mainly as a result of improved life expectancy.

Valve disease remains underdetected,[8] and there are major variations in the provision of health care in all countries of the world, including those that are industrially developed.[9,10] This chapter reviews the causes of valve disease, describes variations in clinical care, and discusses ways in which the worldwide burden of disease could be reduced.

Causes of Valve Disease

The principal causes of valve disease and approximate prevalence are shown in Table 1-1.

Rheumatic Disease

Rheumatic fever occurs in children aged 5 to 15 years from the immune response to group A beta-hemolytic streptococcal pharyngitis. The response occurs 1 to 5 weeks after the initial infection and is caused by molecular mimicries between streptococcal M protein and human myosin and between group A carbohydrate in the streptococcus and valve tissue.

Genetically determined immune markers affect susceptibility to the initial infection and help determine the risk for development of chronic rheumatic disease.[11,12] There is some evidence for disordered signaling mechanisms and reactivation of embryologic pathways.[13] Some streptococcal serotypes (emm types