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# Congenital Heart Disease in Adolescents and Adults

## **Endorsed by**

The ESC Working Group on Grown-up Congenital Heart Disease

AEPC Adult with Congenital Heart Disease Working Group

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The aim of this series is to cast light on the most significant aspects – whether still debated or already established – of congenital heart disease in adolescents and adults and its management. Advances in the medical and surgical management of congenital heart disease have revolutionized the prognosis of infants and children with cardiac defects, so that an increasing number of patients, including those with complex problems, can reach adolescence and adult life. The profile of the adult population with congenital heart disease (ACHD) is consequently changing, and in future many adult patients will present different hemodynamic and cardiac problems from those currently seen. A cure is rarely achieved, and provision of optimal care is therefore dependent on ongoing surveillance and management in conjunction with experts in this highly specialized field. Specialists in ACHD management need to have a deep knowledge not only of congenital cardiac malformations and their treatment in infancy and childhood, but of general medicine, too. A training in adult cardiology, including coronary artery disease, is also essential. Similarly, surgeons need to acquire expertise and good training in both adult and pediatric cardiosurgery. Readers will find this series to be a rich source of information highly relevant to daily clinical practice.

More information about this series at <http://www.springer.com/series/13454>

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Editors

# Intensive Care of the Adult with Congenital Heart Disease

 Springer

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*I dedicate this book to my lovely family, to my co-editors, and to the distinguished authors, and mostly to all the patients with congenital heart disease, those who have reached adulthood and those who will. Sending a strong message of hope and great expectations for the future.*

Eduardo da Cruz

*I thank my wife Anne, family, colleagues, and friends for their continuing support and forbearance, and Drs. Frank Shann and Michael Scallan for inspiring me through their professionalism and humility.*

Duncan Macrae

*Dedicated to the patients I have cared for, the colleagues I have worked with, and my loving family.*

Gary Webb

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## Preface to the Series

In Europe, we are currently faced with an estimated ACHD population of 4.2 million; adults with congenital heart disease now outnumber children (approximately 2.3 million). The vast majority cannot be considered cured but rather having a chronic heart condition that requires further surveillance and timely re-intervention for residual or consequent anatomical and/or functional abnormalities. ACHD patients have very special needs and the physicians taking care of them need expert training. Special health care organization and training programs for those involved in ACHD care are therefore required to meet the needs of this special population.

ACHD problems remain a small part of general cardiology training curricula around the world, and pediatric cardiologists are trained to manage children with CHD and may, out of necessity, continue to look after these patients when they outgrow pediatric age.

There are clearly other health issues concerning the adult with CHD, beyond the scope of pediatric medicine, that our patients now routinely face. Adult physicians with a non-CHD background are therefore increasingly involved in the day-to-day management of patients with CHD.

Experts in congenital heart disease should work to improve the health care system, so that teens and young adults have an easier time making the transition from receiving health care in pediatric cardiology centers to receiving care from specialists in adult cardiology.

The aim of this series is to cast light on the most significant aspects of congenital heart disease in adolescents and adults and its management, such as transition from pediatric to adulthood, pregnancy and contraception, sport and physical activities, pulmonary hypertension, burning issues related to surgery, interventional catheterization, electrophysiology, intensive care management, and heart failure.

This series wishes to attract the interest of cardiologists, anesthesiologists, cardiac surgeons, electrophysiologists, psychologists, GPs, undergraduate and post-graduate students, and residents, and would like to become relevant for courses of cardiology, pediatric cardiology, cardiothoracic surgery, and anesthesiology.

We thank both the wonderful groups of leading cardiovascular experts from around the world, for donating their precious time, producing excellent textbooks

and making this book series a reality, and the members of the two Working Groups (ESC and AEPC ACHD/GUCH Working Group) for the invaluable suggestions and support without which this work would not be possible.

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Münster, Germany  
Munich, Germany  
San Donato Milanese, Italy

Massimo Chessa  
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## Foreword

The care of children with congenital heart disease has been without doubt one of the greatest achievements of cardiovascular medicine and surgery in the twentieth century. As a result, the number of adults with congenital heart disease has grown exponentially; there are currently more adults with congenital heart disease alive than children. This number has now exceeded one million in North America, with similar increases in Western Europe. In parallel, there are now more patients with complex congenital heart disease, who—with improved care—also survive to adulthood.

The net effect of this successful story of modern medicine is that there are ever-increasing needs and pressures for tertiary expertise to provide for this growing adult population with congenital heart disease. Many patients require re-interventions, catheter and/or surgical, whereas late complications such as arrhythmias, heart failure, and pulmonary hypertension are common among them. The spectrum of anatomic substrate, the range of cardiovascular physiology, and the dynamic and continuously evolving nature of catheter and surgical interventions, both at presentation—early in life—and thereafter, means that adults with congenital heart disease represent a very heterogeneous patient group in need of tertiary expertise and multidisciplinary care.

Our approach to diagnosis and management of adults with congenital heart disease has made great progress over the past 2–3 decades, improving the prospects for our patients. There are now worldwide units and teams with medical and surgical expertise to provide for patients with congenital heart disease and their life-long needs. There are still areas, however, where a better understanding, more evidence, and a consensus to adult congenital heart care are lacking. Intensive care (IC) is one of them; patients coming for cardiac reoperations, high-risk catheter or other intervention, management of decompensated heart failure or pulmonary arterial hypertension, and/or for mechanical support (as a bridge to transplantation or destination therapy) require tertiary expertise in the best possible environment. The textbook *Intensive Care of the Adult with Congenital Heart Disease: Interdisciplinary Concepts* edited by Eduardo M da Cruz, Duncan Macrae, and Gary Webb comes to cover this need. The three eminent editors have brought together an international panel of experts in their respective fields to contribute to this effort. The ideal intensive care setting is discussed; there are chapters on nomenclature, classification and risk score assessment, general IC management, pharmacological considerations, anesthesia, psychosocial preparation, and follow-up of the patient. There are



specific challenges with this patient population requiring and receiving intensive care, namely ventricular fibrosis and dysfunction, low cardiac output, peri-procedural arrhythmia, cyanosis, and different adaptive mechanism/s to acute hemodynamic changes, compared to their pediatric counterparts. This necessitates intensive care adult expertise, ideally applied in an adult congenital heart disease health care environment. There are essential chapters on cardio-pulmonary interactions, critical care management of common conditions (tetralogy of Fallot, coarctation of the aorta, single ventricle, aortopathies, Eisenmenger syndrome), arrhythmias, endocarditis, heart failure, mechanical support, and transplantation. Last but not least, the textbook covers management of acquired cardiac disease, other noncardiac comorbidities, anticoagulation, nutrition, nursing, rehabilitation, and end of life issues. This rather comprehensive textbook, therefore, comes to fill in a knowledge gap and at the same time highlights research opportunities in this important field of IC, as pertains to the adult with congenital heart disease.

I believe that every physician, nurse, or health care professional—whether senior or junior—who works in our expanding field of adult congenital heart disease has much to gain from this book.

Royal Brompton and Imperial College  
London, UK

Michael A. Gatzoulis

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## Preface

There are currently more adults than children with congenital heart disease in the developed world, and this trend will continue owing to improvements in the management of pediatric patients and notably those with complex congenital heart disease. This number has now exceeded one million in North America reflecting similar data in other developed countries. This growing adult population with congenital heart disease requires highly specialized and multidisciplinary follow-up, surgical and interventional procedures, and management of late complications related to the natural history of their primary disease, comorbidities, and acquired ailments. Resources needed to successfully manage this very heterogeneous patient group are onerous as the expert community endeavors to implement consistent and efficient models that serve patients within the frame of excellent quality and safety. Although much progress has been accomplished, there are areas such as intensive care that still need to evolve as a core of interdisciplinary and transdisciplinary close collaboration between professionals trained in both pediatric and adult internal medicine, cardiology, anesthesia, intensive care, nursing, pharmacology, psychology, and surgery, to name but a few. The need to integrate experts from multiple backgrounds into a horizontal line of collaboration is vital, with the common objective of compassionately and effectively serving patients and their families. This textbook's title *Intensive Care of the Adult with Congenital Heart Disease: Interdisciplinary Concepts* illustrates the need for this interaction that is the only way to ultimately provide the best possible care to adults with congenital heart disease.

This comprehensive book discusses the many challenges faced in the management of these patients and attempts to provide the first universal review of the practical management of patients with these complex conditions. It advocates for a consistent interaction and collaboration between several disciplines while offering concise and pragmatic recommendations and basic and advanced concepts that allow caregivers to anticipate, prevent, and effectively treat such pathologies. While bringing together top international experts who are leading reference programs around the globe, this book is an indispensable teaching tool for clinicians and caregivers involved in the management of critically ill adults with congenital heart disease.

Aurora, CO  
London, UK  
Cincinnati, OH

Eduardo da Cruz  
Duncan Macrae  
Gary Webb

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## About the Editors

**Eduardo da Cruz** was born in Lisbon, Portugal, and has had an international life career, having lived in Portugal, Costa Rica, France, United Kingdom, Switzerland, and the United States of America. He trained in medicine and then in pediatrics at the Universidad de Costa Rica and the Hospital Nacional de Niños in San José, Costa Rica, and then pursued a fellowship in pediatric cardiology and intensive care in Paris, France (Hôpital Necker-Enfants Malades, Université René Descartes-Paris V). After completion of his training, he stayed in Europe as an attending physician in pediatric cardiology and intensive care until 2007, when he joined the Heart Institute at Children's Hospital Colorado, University of Colorado Denver, School of Medicine, in Denver, USA. He currently holds the title of Tenured Full Professor of Pediatrics, Pediatric Cardiology and Intensive Care, Associate Medical Director of the Heart Institute, and Head of the Pediatric Cardiac Critical Care Program and Inpatient Services. He has extensive experience in the medical and perioperative management of neonates, children, and young adults with complex congenital or acquired heart disease, including heart transplant, pulmonary hypertension, heart failure, mechanical assistance and quality improvement, safety, clinical effectiveness, stewardship, and crew resource management. He is very actively involved in clinical and translational research and teaching in the fields of pediatric cardiology and cardiac intensive care, has delivered hundreds of international lectures, and is a reviewer for 24 peer-reviewed journals, and the co-editor of seven CICU textbooks, and the editor-in-chief of the recently published reference entitled *Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care* (Springer-Verlag UK), a major textbook and e-book/e-reference with 6 volumes and close to 4000 pages. He has published more than 60 book chapters and close to 100 manuscripts in peer-reviewed journals. He is the founder and former chair of the Working Group on Pediatric Cardiac Intensive Care of the Association for the European Pediatric and Congenital Cardiology (AEPC), founder and chair of the Section of Pediatric and Congenital Cardiac Intensive Care and Mechanical Circulatory Assistance of the European Society of Pediatric and Neonatal Intensive Care (ESPNIC), a former board member of the Congenital Domain of the European Association for Cardio-Thoracic Surgery (EACTS), and member of the Society of Pediatric Research (SPR) and of multiple other international societies. Eduardo da Cruz is also an expert reviewer for the European Commission Horizon 2020 Project and the president and chair of the board of the Surgeons of Hope Foundation, a nongovernmental organization based in New York, USA.

**Duncan Macrae** is a pediatric intensivist at the Royal Brompton Hospital, London, UK, and also holds an academic appointment at Imperial College, London. He has worked in critical care for over 30 years, supporting the evolution of specialist congenital cardiac critical care. Dr. Macrae has made major contributions through research and teaching in areas such as ECMO, inhaled nitric oxide therapy, and glycemic control. He is a member of the editorial boards of several major journals including *Pediatric Critical Care Medicine*, *Intensive Care Medicine*, and the *World Journal for Pediatric and Congenital Heart Surgery*. He is a former president of the Pediatric Cardiac Intensive Care Society.

**Gary D. Webb, M.D.** is a professor of pediatrics and internal medicine at the University of Cincinnati College of Medicine and, until May 2016, the director of the Adult Congenital Heart Program at Cincinnati Children's Hospital Heart Institute. After receiving a B.Sc. and MDCM from McGill University in Montréal, Québec, he went on to intern at the Royal Victoria Hospital in Montréal, and then to training in internal medicine and cardiology at the University of Toronto. After 8 years as chief of cardiology at the Wellesley Hospital in Toronto, he moved back to Toronto General Hospital. Beginning in 1980, he was codirector and then director of the Toronto Congenital Cardiac Center for Adults. For several years, he directed the adult cardiology training program at the University of Toronto. He is a Fellow of the Royal College of Physicians and Surgeons of Canada in both internal medicine and cardiology. He is a Fellow of the American College of Cardiology and a life member of the European Society of Cardiology. In 2004, he relocated to Philadelphia, serving as director of the Philadelphia Adult Congenital Heart Center at the University of Pennsylvania. In 2009, he took up his position at Cincinnati Children's Hospital.