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JAMES JAGGERS
EDITORS

Pediatric and
Congenital
Cardiology,
Cardiac Surgery
and Intensive Care

Pediatric and Congenital Cardiology,
Cardiac Surgery and Intensive Care

Eduardo M. da Cruz • Dunbar Ivy
James Jagers
Editors

Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care

With 1377 Figures and 296 Tables

 Springer Reference

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*To Suzanne, Esteban, Tomás and all my dear family and
beloved ones*

To my inspiring mentors and mentees

To all the children of the World

Eduardo M. da Cruz

*To my loving and patient wife and daughters, Ellen, Madeline
and Meg*

Dunbar Ivy

*This book is dedicated to children and families with congenital
cardiac disease for whom I have been privileged to care for
and to the teachers and mentors who have provided me the
knowledge and skill to do so*

James Jagers

Foreword I

The last thirty years has seen some spectacular advances in the diagnosis and management of children with congenital and acquired heart disease. In the former instance, we have moved from an era of an early palliative surgical approach followed by later repair when mortality of 10 % or higher was common to the modern approach of surgical reconstruction in infancy with a mortality of less than 5 %. Underlying this success story are contributions from all the groups that are involved in the care of children with heart disease including pediatric cardiac nurses, cardiologists and cardiac surgeons, perfusionists, anesthesiologists and pediatric intensivists. They are all members of a team and teamwork is the key ingredient of high performing pediatric cardiac programmes.

My own area of intensive care medicine is an essential part of that team and has now developed into a specialty of pediatric cardiac critical care in it's own rite with an expectation that physicians should have a comprehensive knowledge of cardiac anatomy and physiology as well appropriate training in pediatric intensive care medicine. The newer generation of trainees will also be expected to have expertise in echocardiography, extracorporeal technology and mechanical support. This *Textbook of Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care* will be source material for all this because it covers every aspect of heart disease in children and will be an invaluable resource for all team members in the pediatric cardiac programme.

Desmond Bohn, MB FRCPC,
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Former Chief, Department of Critical Care Medicine
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Foreword II

To say that this text is a monumental undertaking is an understatement. The crux of this work is in the subtitle. It is *comprehensive, interdisciplinary*, and, perhaps most importantly, *interactive*. There are several excellent existing textbooks of pediatric cardiology, most of which follow a familiar format of chapters on anatomy and embryology, structural malformations, myocardial/inflammatory diseases, and perhaps heart failure and pulmonary hypertension. None that I know of, cover such a diverse range of subjects as this text: therapeutic hypothermia and tracheal reconstruction; venous thromboembolism and trypanosomiasis; Takayasu disease and Nursing. Authorship is likewise diverse, with contributors from all across the world.

Medicine is an extremely rapidly changing field. The time, often years, between the decision to publish a textbook and the actual publication frequently means that much of what is written is out of date by the time of publication. Thus the decision of the editors and publisher to create both a print and a web version, the latter to be updated frequently and read on a peripheral mobile device help make it both current and accessible. No one carries a large textbook into the ER in the middle of the night, but everyone carries a mobile phone.

Howard P. Gutgesell, MD
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Foreword III

Sixty years ago on May 6 1953 John Gibbon was the first to achieve successful repair of a congenital heart defect using a heart lung machine. What is often forgotten however is that Gibbon's subsequent five patients did not survive. Many other pioneers of cardiac surgery of that era had equally dismal results. Today the outlook for the child with congenital heart disease has improved dramatically. At the top of my list of reasons for that improvement has been the establishment of cohesive interdisciplinary teams of individuals dedicated to the care of patients with congenital heart disease. While these individuals may have expertise in an incredibly specialized area of cardiac surgery, cardiology, cardiac anesthesia, cardiac intensive care, perfusion, cardiac nursing or many of the essential ancillary healthcare support areas, their individual contribution to the successful care of a child with a complex congenital heart problem is only as good as the weakest link in the entire team. This stunning new textbook edited by Eduardo M. da Cruz, Dunbar Ivy and James Jagers brings together authors from all of the many components of the healthcare team devoted to care of the individual, both adult and pediatric, with congenital heart disease. The primary editors have assembled a team of specialist sub-editors who have each assembled a sub-team of contributors from around the globe. One of the great joys of working within the field of congenital heart disease is that in contrast to many other medical specialties, we are a relatively small family who have come to know each other on a global scale, symbolized by our quadrennial meeting at the World Congress of Pediatric Cardiology and Congenital Heart Surgery.

The authors have brought the reader into the new millennium of publishing by creating an electronic version of this textbook that includes access to videos demonstrating surgical technique as well as diagnostic modalities. The exhaustiveness of the coverage is truly breathtaking. Just reading the list of contents and appreciating the breadth of coverage draws attention to the very considerable care and effort that the editors have put into assembling this unique book. Whether a cardiologist, cardiac surgeon, cardiac intensivist,

a cardiac anesthesiologist or a nurse in the intensive care unit or any one of the many supporting allied healthcare workers in the field of congenital cardiac care, the reader will surely not be disappointed.

Richard Jonas, MD
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Preface

Pediatric and congenital cardiac patients, from the fetus to the adult, remain in the current century a complex challenge in an ever evolving discipline. Successful cardiovascular programs currently deal with the most critical patients by promoting a horizontal convergence of multiple specialties. Many, if not most, programs have multidisciplinary teams available, and yet the challenge remains to make these teams operate as *interdisciplinary*. Two key goals ought to be pursued in order to achieve successful short and long term outcomes with pediatric cardiac patients. Firstly, the driving force should always be *quality improvement and safety*, and secondly good *communication* shall prevail. There are two main pillars of the latter endeavors, namely individual-based and system-based principles. Individual-based principles are quite subjective and more difficult to manage, since they depend on personalities and behavior. In a balanced environment, individuals should be able to endorse a willingness to develop exhaustive knowledge, team work, trust, self-awareness, capacity to listen, common sense and respect. System-based practices are more objective, and although requiring cultural changes, promote consistency, harmonious interaction and better outcomes. Such practices allow the development of common paths for the team to follow, whilst enhancing good communication and reducing risks, optimizing prevention of complications, better handling fluctuations and identifying outliers, and predicting likely outcomes based on accurate data. The implementation and consolidation of efficient programs require systematic audits, development of sound database platforms, development of plans to address deficiencies, implementation of simulation and quality improvement and safety programs, and the eagerness for transparency and to share the available information with staff, patients and their families in a non-repressive and constructive atmosphere. Such models are not of the realm of utopia, although they are challenging to establish, at least whilst inducing the required cultural changes and promoting the conviction of their usefulness.

The textbook of Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care intends to achieve an ultimate objective beyond providing reliable scientific information. It endeavors to symbolize the imperative need for a cohesive and transparent interdisciplinary blend of expertise, by bringing together world renowned authors from different regions around the Globe, and representing the many specialties concerned by and involved in the management of pediatric patients with congenital and acquired cardiac

diseases. We have been privileged to gather many experts from reference programs to whom we will remain forever grateful. As much as we have endeavored to remain consistent, some authors may express personal opinions and hypothesis in a constructive manner, which -as we expect- may help readers understand the many facets and complexities of patient management at different levels. Very importantly, in this project readers will be able to access a website which we see as a source of constantly updated information, including videos dedicated to diagnosis and surgical interventions, and that should evolve overtime into a more interactive tool. We sincerely hope that the Textbook of Pediatric and Congenital Cardiology, Cardiac Surgery and Intensive Care will become a useful tool in the armamentarium of those caring for such complex patients; we aim to provide updated information to help caregivers become better practitioners and human beings. If so, this textbook would have achieved its *raison d'être*.

July 2013

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Last but not least, we thank Springer-Verlag, particularly Grant Weston, Mansi Seth and Navjeet Kaur for their advice and collaboration.

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Eduardo M. da Cruz is a Professor of Pediatrics (Pediatric Cardiology and Intensive Care) and the Head of the Pediatric Cardiac Critical Care Program & Inpatient Services at The Heart Institute, Department of Pediatrics, Children’s Hospital Colorado, University of Colorado Denver-School of Medicine. He trained in pediatrics, pediatric cardiology and intensive care and has been an attending physician for 20 years in Europe and in the USA. He has extensive experience in the medical and perioperative management of neonates, children and young adults with complex congenital or acquired heart, including heart transplant, mechanical assistance and quality improvement and safety. He is very actively involved in clinical research and teaching in the fields of pediatric cardiology and cardiac intensive care, has delivered hundreds of international lectures and is the editor of various Cardiology and CICU reference textbooks. Eduardo M. da Cruz has published 50 book chapters and dozens of manuscripts in peer-reviewed journals. He is the founder of the Working Group on Pediatric Cardiac Intensive Care of the

Association for the European Paediatric and Congenital Cardiology (AEPC), Deputy Chair and founder of the Section on Hemodynamics and Heart Disease of the European Society of Paediatric and Neonatal Intensive Care (ESPNIC), Board Member of the Congenital Domain of the European Association for Cardio-Thoracic Surgery (EACTS), member of the Society of Pediatric Research (SPR) and of multiple international Societies, and the President of Surgeons of Hope Foundation, based in New York.



Dunbar Ivy, MD, Chief of Pediatric Cardiology, Selby's Chair of Pediatric Cardiology, Co-Director of Children's Hospital Heart Institute, Director of Pediatric Pulmonary Hypertension Program Children's Hospital Colorado and Professor of Pediatrics, University of Colorado School of Medicine Aurora, CO, USA

Dr. Dunbar Ivy began his medical career at Tulane University School of Medicine following his premedical studies at Davidson College. While at Tulane, he became excited about a career in Pediatric Cardiology under the mentorship of Dr Arthur Pickoff. He then obtained training in General Pediatrics at the University of Colorado School of Medicine in Denver, Colorado. Early mentors in Pediatric Cardiology included Drs. Michael Schaffer and Henry Sondheimer. Interest in altitude related illness and pulmonary hypertension in congenital heart disease were fostered by Dr Robert Wolfe on the clinical side and Drs Steve Abman and John Kinsella in the fetal sheep laboratory while a fellow in Pediatric Cardiology at the University of Colorado. Following fellowship, he became a research instructor under the guidance of Dr Mark Boucek, who encouraged him to pursue a career as a clinician scientist. During his time as a Bugher fellow, he obtained early grants from the March of Dimes and American Heart Association regarding the role of endothelin in the perinatal pulmonary circulation. This work transitioned into a National Institutes of Health K-08 award to continue to study molecular derangements in the endothelin pathway in models of pulmonary hypertension. In 2003 Dr Ivy took the position of Chief of Pediatric Cardiology and Selby's Chair of Pediatric Cardiology. His research focus

became more clinical and translational. As Director of the Pediatric Pulmonary Hypertension Program, he began early clinical studies of medical therapy in children, including the use of intravenous epoprostenol, subcutaneous treprostinil, and oral bosentan. He began to work with Dr Robin Shandas regarding measurement of right ventricular afterload in children with pulmonary hypertension in an NIH sponsored Specialized Centers of Clinically Oriented Research grant headed by Dr Kurt Stenmark. Further work on ventricular vascular coupling has continued with NIH funding with Dr Shandas. Dr Ivy was the inaugural Chairman of the first Pediatric Pulmonary Hypertension taskforce at the World Symposium of Pulmonary Hypertension in Nice, France in 2013. Dr. Ivy is a member of multiple societies, and has published over 150 peer reviewed manuscripts.



James Jagers, MD, Barton-Elliman Chair and Chief of Pediatric Cardiac Surgery, Professor of Surgery, Children's Hospital Colorado, University of Colorado, Aurora, CO, USA

Dr. James Jagers began his medical career at the University of Nebraska Medical Center as a medical student. He then obtained training in general Surgery at the Oregon Health Sciences University in Portland Oregon and Thoracic Surgery training at the University of Colorado Health Sciences Center in Denver. During this period he also completed a Pediatric Cardiac Surgery Fellowship at the Children's Hospital in Denver. Following this he took a faculty position at Duke University Medical Center in Thoracic and Pediatric cardiac Surgery where he rose to the rank of Associate Professor with tenure. This tenure at Duke was interrupted by a very brief position at the University of Missouri and Mercy Children's Hospital in Kansas City. Following this, he returned to Duke to assume the position of Chief of Pediatric Cardiac surgery and Director of the Duke Pediatric Heart Institute. During his time at Duke, Dr. Jagers directed the pediatric cardiovascular surgery laboratory and mentored many research fellows. He was principal and co-principal investigator on two basic Science NIH grants and one Pediatric Heart Network NHLBI sponsored multicenter study. In 2010, Dr. Jagers

moved to the University of Colorado and Children's Hospital Colorado where he is now the Barton Elliman Chair of Congenital Cardiac Surgery and Professor of Surgery. His current research interests include Outcomes research in care of Congenital Heart Disease and investigation into the protein signaling of aortic stenosis and uncompensated cardiac hypertrophy and myocardial dysfunction. Clinical interests include complex neonatal heart surgery, cardiac transplantation and fetal intervention for cardiac defects. Dr. Jagers is a member of multiple Societies, and has published 117 peer reviewed manuscripts, published 21 book chapters and is a reputed international lecturer with 75 invited lectures.

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