Critical Care of Children with Heart Disease
To my wife Lina, my sons Rafael and Ricardo, and my grandson Daniel.
Ricardo Muñoz

To my wife Amy, and my children, Victor, Samuel and Oliver; for your love and support.
Victor O. Morell

To Tim, Jasmine, and Mom, for all of your patience...
Carol G. Vetterly

To Suzanne, Esteban and Tomas, as ever and forever...
Eduardo M. da Cruz

The Editors would also like to dedicate this book to all those caregivers who commit to care for children and young adults with critical congenital and acquired heart disease

We would like to express our sincere gratitude and appreciation for our illustrators, Steve Goldberg and Angelo Rutty, who created the exceptional surgical figures throughout the text. Their outstanding talents and contributions helped to make the book a valuable educational tool.
Ricardo Muñoz
Víctor O. Morell
Carol G. Vetterly
Eduardo M. da Cruz
Foreword
“Critical Care of Children with Heart Disease: Basic Medical and Surgical Concepts”

I am flattered to be invited to write a foreword for such a timely and comprehensive handbook of pediatric cardiac intensive care.

Having been intimately engaged in the management of congenital heart diseases for the greater part of the last 40 years, it has been rewarding to witness progress and interesting to reflect on what contributed to it. As in many endeavors, progress has been subtle and its underlying mechanisms have been multifactorial. There is little doubt that perioperative intensive care management has been a key component of this progress.

In many centers the overall mortality for pediatric cardiac surgery has fallen below the 2% mark. The morbidity, however, remains significant and the authors of this handbook must be congratulated for dedicating a large section to cardiac and extracardiac complications. Morbidity should become part of performance assessment.

It has been interesting to witness how postoperative morbidity has evolved. The postoperative capillary leak syndrome of the small infant in the 1970s has been replaced by the postoperative pulmonary hypertensive crises in the 1980s, to make room to the problems related to the imbalance between systemic and pulmonary blood flow in the Norwood patients.

The advent of the Norwood operation has been a cornerstone in the management of congenital heart defects. A successful Norwood implies accurate diagnosis, expert anesthesia and perfusion, impeccable surgery, deep understanding of the postoperative cardiopulmonary interactions, and taming of extracardiac problems, such as sepsis, gastrointestinal, and neurological impairments. The efforts deployed by many institutions to achieve excellence with the Norwood patients have uplifted their overall performance in the management of all congenital heart defects.

The impressive list of experts who have contributed to this book highlights the importance of multidisciplinarity in the perioperative management of the sick cardiac patient.

Multidisciplinarity is a two-edged sword. On the one hand, it provides knowledge, competence, commitment, and availability, which are key components of success. On the other hand, shared care can lead to conflicts and redundancy. It creates interfaces that require collaboration, hierarchy, and leadership. Too often those who are made responsible for problems arising within intensive care units do not have the authority to solve them. Several institutions over the past 30 years have faced difficulties in establishing the multidisciplinary structure of their cardiac intensive care units, which is most beneficial to their patients and most cost-effective to their institution. This is a delicate topic that often generates emotional if not legal argumentations. I am taking the liberty of alluding to this important issue even though it is not specifically addressed in this handbook.

Marc R de Leval, MD, FRCS
Professor of Cardiothoracic Surgery
Progress in medicine is compounded of two processes: small steps with intelligent “drift” toward best practice, and sea change with an occasional paradigm “shift.”

Ospina-Tascon, Buchele, and Vincent (Critical Care Medicine 2008; 36:1311–1322) were able to find in the literature only 72 large, randomized, controlled, intensive care trials with mortality endpoints. Of these, the trial intervention was a statistical success in ten. Often, progress in critical care takes place because reports that suggest but do not prove benefit, as well as lesser forms of evidence, become incorporated into clinical wisdom and medical practice by consensus. Ultimately, as norms drift, it may become impossible to test hypotheses by randomized, controlled trial because standard (control) treatment and trial intervention have become too alike. Outcome of new treatments must then be compared to historic controls because prospective trials are no longer possible.

In Pediatric Cardiology and Cardiac Surgery, the engine for change has seldom been the randomized controlled trial. Laboratory studies, case reports, small series, and staggering successes have fostered the “drift” and “shift” of cardiac care. Often, with little experimental foundation, one technique is modified or overthrown by another. New approaches are tried, and may become state-of-the-art virtually overnight.

The sea changes in pediatric cardiovascular surgery have been obvious. The first reported surgical ligation of a ductus arteriosus by Robert Gross, then 33 years old, was performed in 1938. Other pioneering “closed” procedures include the first excision of a coarctation of the aorta (Crafoord, 1944), the first subclavian to pulmonary artery shunt (Blalock and Taussig, 1944), and the first pulmonary artery band (Dammon-Muller, 1952). The surgical tradition is relatively receptive to steep “learning curves” in the adoption of new approaches. When cardiac bypass was introduced, between 1952 and 1954, 18 open heart operations were described in six separate reports, with an overall mortality of 94.5%. The bubble oxygenator was introduced in 1955. Then, in 1956, Lillehei, Cohen, Warden, and Varco reported the survival of a patient after total correction of tetralogy of Fallot. The sea changes have continued. More recent “paradigm shifts” include the introduction of deep hypothermic circulatory arrest, shelving of the Mustard and Senning procedures in favor of the arterial switch, Fontan’s approach to tricuspid atresia and the resulting sea change in the operative management of single ventricle defects, Norwood’s procedure for the hypoplastic left heart syndrome, and the introduction into postoperative care of extracorporeal membrane oxygenation and ventricular assist devices.

Some 50 years have elapsed since Lillehei’s report. Cardiac bypass has become a safe and well-established technique. Corrective open heart surgery is commonly
performed in small neonates. Most congenital cardiac defects can be either palliated or corrected, and many centers report overall mortality rates for congenital heart surgery at or below 2%. Much of this progress can be attributed to the more subtle process of “drift,” which has, by consensus, moved the age of corrective operation toward the first days of life, increased the specificity and accuracy of prenatal diagnosis, replaced routine cardiac catheterization with noninvasive diagnostic techniques like 2-D echocardiography and MR Angiography, and transformed many corrective procedures from surgical into transcatheter interventions.

In general, new discoveries and pioneering developments trigger sea changes. Intelligent adoption of a paradigm shift and cautious drift toward best practice occur by integration of the best evidence and by consensus building. That integration is the unwritten mission of a textbook such as this.

Consensus is a team activity and requires team work. The virtual exclusion of non-surgeons from postoperative care has ended. Interdisciplinary teams have been assembled to collaborate in this endeavor, improving outcome and accelerating the pace of progress. These teams include practitioners who first learn their craft as cardiologists, pediatric intensivists, pharmacists, and respiratory therapists, as well as cardiac surgeons who first learn their craft on adults (who are not just “big children”). It is the need for a literature to describe, explain, and teach critical care of the pediatric cardiac patient to trainees and collaborators of diverse backgrounds that has generated this textbook. In *Critical Care of Children with Heart Disease: Basic Medical and Surgical Concepts*, the authors embed the knowledge base of cardiology and congenital cardiac surgery into the context of intensive care, and advanced cardiac diagnostics and therapeutics. The product is not so much a cardiology text for intensive care physicians, as it is a text about the critical care of patients with pediatric heart disease.

Bradley P. Fuhrman, MD  
Chief, Pediatric Critical Care  
Children’s Hospital of Buffalo  
Professor of Pediatrics and Anesthesiology  
State University of New York at Buffalo  

Women and Children’s Hospital of Buffalo  
219 Bryant Street  
Buffalo, New York 14222  

Home:  
83 Ashland Avenue  
Buffalo, New York 14222
The development of intensive care units can be traced, in part, to the challenge of ventilating dozens of patients with respiratory failure simultaneously during the polio epidemic in Copenhagen (1952–1953). The advantage of cohorting patients with “intensive care” needs became apparent and the value of an intensive care unit was established. During this initial experience dozens of patients were hand ventilated simultaneously via tracheostomy tubes (or subsequently with negative pressure ventilators) before there was adequate clinical appreciation of acid base balance. Numerous deaths were attributed to “a mysterious alkalosis.” By 1966, with the advent of positive pressure ventilators and the appreciation of arterial blood gas analysis, patient care was transformed into the field of critical care.

There is a striking parallel and temporal overlap to this discovery of mechanical ventilation and innovation in critical care: the development of cardiopulmonary bypass (1952–1953) and the need for cardiac intensive care. The early experience with cardiopulmonary bypass was predominantly in children. The poor results were discouraging for most; 17 of the first 18 patients died. But Walt Lillehei’s demonstration of the feasibility of repairing congenital heart disease using a parent as the functional source of pump and gas exchange in a cross circulation experiment propelled the field forward and motivated others to refine cardiopulmonary bypass techniques and successfully apply them to the repair of congenital heart disease.

By 1966 (when Rashkin introduced the balloon atrial septostomy in infants with transposition of the great arteries) cardiac intensive care units for children were emerging in Boston, Toronto, Minneapolis and elsewhere. In a 1966 landmark paper by Brown, Johnston and Conn in Toronto, the early results of mechanically ventilated patients in a postoperative care unit for children with congenital heart disease were reported. Although mortality exceeded 50% for those patients requiring 24 h of postoperative mechanical ventilation, there were many successes. Pediatric cardiac intensive care units were born. At first wedged between adult intensive care and adult cardiac surgical units, then mingling, merging with, evolving, and emerging from pediatric intensive care units, this subspecialty now exists in its own right with dozens of highly specialized units throughout the world. The subspecialists who populate the clinical leadership of these units, represent its multifaceted heritage: pediatric anesthesiologists, cardiologists, cardiac surgeons, and intensivists. It is further defined by specialized nursing dedicated exclusively to caring for critically ill children with heart disease. This book, Critical Care of Children with Heart Disease: Basic Medical and Surgical Concepts, is a wonderful testament to the collaborative expertise that must be brought to bear on this relatively new subspecialty. The textbook, like the discipline, is truly a multidisciplinary and multinational effort.
I have been honored to work with Dr. Muñoz in his early years, along with many of the other authors involved in this textbook. They, like many of those in this new field, chose to blend multiple conventional training programs and an unconventional number of additional years of training to help define the curriculum, qualifications and content of pediatric cardiac intensive care. It is my pleasure now to read and learn from those who represent the next and now current authorities in this field.

David L. Wessel, MD  
Senior Vice President, Children’s National Medical Center  
Washington, D.C
Acknowledgements

Carol G. Vetterly has previously published under the name Carol G. Schmitt.
Preface

Pediatric cardiac intensive care patients pose special challenges to those practitioners caring for them.

The primary purpose of this textbook is to provide the health care practitioner with an overview of both the medical and surgical facets in caring for pediatric patients with congenital or acquired cardiac disease.

This book conceals a multitude of topics that may be encountered when caring for children in a cardiac intensive care setting. The first part of the text covers general aspects ranging from mechanical ventilation and cardiac anesthesia, sedation and pain management, to cardiopulmonary bypass, cardiac catheterization, echocardiography, in addition to describing the special monitoring required for pediatric cardiac patients. It also includes important recent developments in assessing and reporting risk factors.

The next sections address specific cardiac anomalies including acyanotic defects, right and left obstructive heart lesions, atrio-ventricular valve anomalies, vascular lesions, pulmonary hypertension, cardiomyopathies, pericardial diseases, and other complex heart defects. Specific chapters are dedicated to mechanical assistance, renal replacement therapy, transplant, arrhythmias, as well as the ethical and legal issues that involve the discontinuation of support of patients.

The last section reviews other challenging non-cardiac problems that caregivers in charge of pediatric cardiac patients have to face on a regular basis, including matters related to respiratory, gastrointestinal, nutrition, hematologic, renal and neurological systems, infectious disease, and skin protection.

A unique aspect of this text is the inclusion of drawings and descriptions of cardiac anomalies which serves as a valuable teaching and learning tool for cardiac intensivists, surgeons, fellows, residents, and nurses.

The editors sincerely acknowledge and express their gratefulness to the collaborating authors who were the successful artisans of this project.

It is our hope that practitioners will find this text useful in achieving the ultimate goal of providing the most superior quality of care to their pediatric cardiac intensive care patients.
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Contributors

Christine M. Angeletti, RN, BSN
Cardiac Intensive Care, Children’s Hospital of Pittsburgh, Pittsburgh, PA, USA

Gaurav Arora, MD
University of Pittsburgh School of Medicine, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Alison Artico, CNP
Pediatric Cardiology, Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA

Katherine A. Barsness MD
Division of Pediatric Surgery, Children’s Memorial Hospital, Chicago, IL, USA

Kristen P. Barton MD
Pediatric Cardiology, University of Florida, Jacksonville, FL, USA

Lee Beerman MD
Cardiology, University of Pittsburgh School of Medicine, Children’s Hospital of UPMC, Pittsburgh, PA, USA

Maurice Beghetti, MD
Pediatric Cardiology, Department of the Child and Adolescent, Children’s University Hospital of Geneva, Geneva, Switzerland

Shannon Buckvold, MD
Pediatric Cardiology and Cardiac Intensive Care, Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA

David Campbell, MD
Pediatric Cardiac Surgery, The Children’s Hospital of Denver, Aurora, CO, USA

Dana Casicato, MSN, CRNP
Cardiothoracic Surgery, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Constantinos Chrysostomou, MD
Cardiology & Critical Care Medicine, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Jessica Church, CNP
Pediatric Cardiology, Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA
**Erin L. Colvin, CRNP**  
Cardiothoracic Surgery, Children’s Hospital of Pittsburgh of UPMC,  
Pittsburgh, PA, USA

**Eduardo M. da Cruz, MD**  
Pediatric Cardiology and Cardiac Intensive Care, Department of Pediatrics,  
The Children’s Hospital of Denver, Aurora, CO, USA

**Jeffrey Darst, MD**  
Pediatric Cardiology and Cardiac Catheterization Laboratory,  
Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA

**Peter J. Davis, MD**  
Department of Anesthesiology, University of Pittsburgh Medical Center,  
Pittsburgh, PA, USA

**Denis Devictor, MD**  
Pediatric and Neonatal Intensive Care, Department of Pediatrics, Hôpital de Bicêtre,  
Assistance publique Hôpitaux de Paris, Le Kremlin-Bicêtre, France

**Yuliya A. Domnina, MD**  
Pediatric Cardiovascular Intensive Care Unit, Sanger Clinic, Levine Children’s Hospital, Charlotte, NC, USA

**Martin Elliot, MD**  
Cardiac Surgery, Great Ormond Street Hospital for Sick Children, London, UK

**Jennifer Exo, MD**  
Critical Care Medicine, Children’s Hospital of Pittsburgh of UPMC,  
Pittsburgh, PA, USA

**Rose Marie Faber, MSN/ED, RN, CCRN**  
Training and Education, Children’s Hospital of Pittsburgh of UPMC,  
Pittsburgh, PA, USA

**Thomas Fagan, MD**  
Pediatric Cardiology and Cardiac Catheterization Laboratory,  
Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA

**Brian Feingold, MD**  
Cardiology, Children’s Hospital of Pittsburgh of UPMC, University of Pittsburgh,  
School of Medicine, Pittsburgh, PA, USA

**Robyn A. Filipink, MD**  
Neurology, Children’s Hospital of Pittsburgh of UPMC,  
University of Pittsburgh, School of Medicine, Pittsburgh, PA, USA

**Rhonda J. Gengler, RN, BSN, CCRN**  
Pediatric Intensive Care Unit, Children’s Hospital of Pittsburgh of UPMC,  
Pittsburgh, PA, USA

**Steven P. Goldberg, MD**  
Pediatric Cardiac Surgery, The Children’s Hospital of Denver, Aurora, CO, USA

**Denise L. Howrie, PharmD**  
Department of Pharmacy, Children’s Hospital of Pittsburgh of UPMC,  
Pittsburgh, PA, USA
Jill Ibrahim, MD  
Pediatric Cardiology and Cardiac Intensive Care, Department of Pediatrics,  
The Children’s Hospital of Denver, Aurora, CO, USA

Dunbar Ivy, MD  
Pediatric Cardiology and Cardiac Intensive Care, Department of Pediatrics,  
The Children’s Hospital of Denver, Aurora, CO, USA

Jeffrey P. Jacobs MD FACS, FACC, FCC  
Department of Pediatrics, University of South Florida, Tampa, FL, USA

Edmund H. Jooste, MB, ChB  
Department of Anesthesiology, University of Pittsburgh Medical Center,  
Pittsburgh, PA, USA

Afksendiyos Kalangos, MD, PhD, DSc  
Cardiac Surgery, University Hospital of Geneva, Geneva, Switzerland

Jonathan Kaufman, MD  
Pediatric Cardiology and Cardiac Intensive Care, Department of Pediatrics,  
The Children’s Hospital of Denver, Aurora, CO, USA

Traci M. Kazmerski, BS  
University of Pittsburgh, School of Medicine, Pittsburgh, PA, USA

Kent Kelly CCP  
Svc, Cardiac Catheterization Laboratory, Cardiothoracic Surgery, ECMO,  
Children’s Hospital of Pittsburgh, Pittsburgh, PA, USA

Rukmini Komarlu, MD  
Pediatric Cardiology, Children’s Hospital of Pittsburgh of UPMC,  
Pittsburgh, PA, USA

Chokri Kortas, MD  
Department of Pediatric Cardiac Surgery, Centre Médico-Chirurgical Marie  
Lannelongue, Le Plessis Robinson, France

Deborah Kozik, DO  
Pediatric Cardiac Surgery, The Children’s Hospital of Denver, Aurora, CO, USA

Jacqueline Kreutzer, MD  
Cardiology, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Bradley A. Kuch, BS, RRT-NPS  
Department of Cardiothoracic Surgery, Children’s Hospital of Pittsburgh of UPMC,  
Pittsburgh, PA, USA

François Lacour-Gayet, MD  
Pediatric Cardiac Surgery, The Children’s Hospital of Denver, Aurora, CO, USA

Evelyn Lechner, MD  
Neonatal Intensive Care Unit, Children’s and Maternity Hospital Linz,  
Linz, Austria

Bertrand Léobon, MD  
Department of Pediatric Cardiac Surgery, Centre Médico-Chirurgical Marie  
Lannelongue, Le Plessis Robinson, France
Steven E. Litchenstein, MD  
Department of Anesthesiology, University of Pittsburgh Medical Center, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Kristyn S. Lowery MPAS, PA-C  
Cardiothoracic Surgery, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Mohammed Ly, MD  
Department of Pediatric Cardiac Surgery, Centre Médico-Chirurgical Marie Lannelongue, Le Plessis Robinson, France

Sara Mackie, PA-C  
Pediatric Cardiology, Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA

Tamara Maihle, MSN, CRNP  
Cardiothoracic Surgery, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Ana Maria Manrique, MD  
Department of Anesthesiology, University of Pittsburgh Medical Center, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Marian G. Michaels MD, MPH  
Division of Pediatrics Infectious Diseases, University of Pittsburgh School of Medicine, Pittsburgh, PA, USA

Nilanjana Misra, MBBS, MD  
Cardiology, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Max B. Mitchell, MD  
Pediatric Cardiac Surgery, The Children’s Hospital of Denver, Aurora, CO, USA

Shelley D. Miyamoto, MD  
Pediatric Cardiology, Department of Pediatrics, The Children's Hospital of Denver, Aurora, CO, USA

Victor O. Morell, MD  
Department of Cardiothoracic and Esophageal Surgery, University of Pittsburgh, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Michael Moritz, MD  
Division of Pediatric Nephrology, University of Pittsburgh School of Medicine, Pittsburgh, PA, USA

Diego Mouguillanski, MD  
Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Ricardo Muñoz, MD, FAAP, FCCM  
Department of Critical Care Medicine, University of Pittsburgh of UPMC, Pittsburgh, PA, USA

Richard A. Orr, MD  
Critical Care Medicine, University of Pittsburgh, School of Medicine, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA
Contributors

**Michael J. Painter, MD**
Neurology and Pediatrics, University of Pittsburgh, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

**Sang Park, MD**
University of Pittsburgh, School of Medicine, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

**Christina M. Phelps, MD**
Pediatric Cardiology and Cardiac Intensive Care, Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA

**Nicholas B. Pigott, MBBS, FRCPI, FRCPCH, FRACP**
Children’s Intensive Care Unit, Sydney Children’s Hospital, Sydney, Australia

**Eric S. Quivers, MD**
Cardiology, Children’s Hospital of Pittsburgh of UPMC, University of Pittsburgh School of Medicine, Pittsburgh, PA, USA

**Asrar Rashid, MD**
Queen’s Medical Centre, Nottingham, University Hospital NHS Trust, Nottingham, UK

**Jody Rutz, PA-C**
Pediatric Cardiology, Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA

**Alain Serraf, MD, PhD**
Department of Pediatric Cardiac Surgery, Centre Médico-Chirurgical Marie Lannelongue, Le Plessis Robinson, France

**Peter H. Shaw, MD**
Hematology/Oncology, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

**Dana Shiderly, BSN, RN, CCRN**
Cardiac Intensive Care Unit, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

**Michael K. Shoykhet, PA-C, MD, PhD**
Critical Care Medicine, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

**Daniel Sidi, MD**
Service of Pediatric Cardiology, Department of Pediatrics, Hôpital Necker-Enfants Malades, University of Paris V, Paris, France

**Joanne K. Snyder, PA-C**
Cardiothoracic Surgery, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

**Amy Stimmler, CNP**
Pediatric Cardiology, Department of Pediatrics, The Children’s Hospital of Denver, Aurora, CO, USA
Pierre Tissières, MD  
Pediatric and Neonatal Intensive Care, Department of Pediatrics, Hôpital de Bicêtre, 
Assistance publique Hôpitaux de Paris, Le Kremlin-Bicêtre, France

Cécile Tissot, MD  
Pediatric Cardiology and Cardiac Intensive Care, Department of Pediatrics, 
The Children’s Hospital of Denver, Aurora, CO, USA

Michael Tsifansky, MD  
Division of Critical Care Medicine, Department of Pediatrics, Advocate Lutheran 
General Children’s Hospital, Park Ridge, IL, USA

Shekhar Venkataraman, MD  
Critical Care Medicine and Pediatrics, Children’s Hospital of Pittsburgh of 
UPMC, Pittsburgh, PA, USA

Carol G. Vetterly, PharmD  
Children’s Hospital of Pittsburgh of UPMC, University of Pittsburgh, 
Pittsburgh, PA, USA

Pascal Vouhé, MD  
Service of Pediatric Cardiac Surgery, Department of Pediatrics, Hôpital 
Necker-Enfants Malades, University of Paris V, Paris, France

Peter D. Wearden, MD, PhD  
Cardiothoracic Surgery, Children’s Hospital of Pittsburgh of UPMC, 
Pittsburgh, PA, USA

Steven A. Webber, MBChB, MRCP  
Cardiology, Children’s Hospital of Pittsburgh of UPMC, Pittsburgh, PA, USA

Lisa B. Willis, MD  
Pediatric Cardiology, Department of Pediatrics, The Children’s Hospital of Denver, 
Aurora, CO, USA

Lisa Wise-Faberowski, MD  
Pediatric Anesthesiology and Cardiac Intensive Care, Department of Pediatrics, 
The Children’s Hospital of Denver, Aurora, CO, USA

Glenda V. Wright, MD  
Pediatric Infectious Diseases, Children’s Hospital of Pittsburgh of UPMC, 
Pittsburgh, PA, USA

Adel K. Younoszai, MD  
Pediatric Cardiology and Cardiac Imaging, Department of Pediatrics, 
The Children’s Hospital of Denver, Aurora, CO, USA