A Multidisciplinary Approach to Perinatal Cardiology
Volume 1

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Edited by

P. Syamasundar Rao,
Dharmapuri Vidyasagar

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Dedicated to my wife, Hymavathi, for her continued support of my academic activities for more than five decades; to my children, Vijay, Madhavi, and Radhika, for enduring and gracefully excusing my absences because of my academic vagaries; and to my grandchildren, Meghana, Anjali, and Rhea, our future!

PSR

Dedicated to my wife, Nagamani, for her continued professional and personal support through the last 50 years of my academic career; to my children, Sahana, Sadhana, and Sanjay, who have been a source of intellectual stimulation; and to my grandchildren, Kavi, Anika, and Maia, sources of joy!

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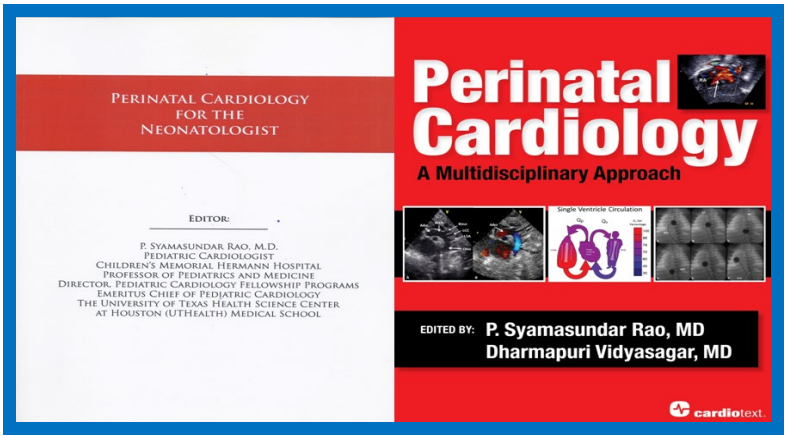
PREFACE TO THE CURRENT EDITION

Developments such as the early detection of neonates with serious heart disease and their rapid transport to tertiary care centers, availability of highly sensitive noninvasive diagnostic tools, advances in neonatal care and anesthesia, progress in transcatheter interventional procedures, and extension of complicated surgical procedures to the neonate and young infant have advanced to such a degree that almost all congenital cardiac defects can be diagnosed and "corrected". Defects that cannot be corrected can be effectively palliated. Cardiac defects that were once fatal in infancy are now treatable and patients are surviving into adulthood. The last five decades have seen a great many advances, as mentioned above, resulting in the increased survival of infants with congenital cardiac lesions. These advances occurred not only in the specialties of pediatric cardiology and pediatric cardiovascular surgery but also in several other disciplines such as neonatology, perinatology, interventional pediatric cardiology, and ultrasonography.

Over the last several years we have organized lectures to Pediatric Residents and Pediatric Cardiology, Neonatology, and Pediatric Intensive Care Fellows on prenatal and neonatal cardiac issues. Some of these presentations have been written up as reviews in publications such as Neonatology Today and Congenital Cardiology Today. The initial intent was to bring these together into a book format so that the material becomes available to all trainees and practitioners in Pediatric Cardiology, Neonatology and Pediatric Intensive Care. This resulted in the publication of a booklet entitled "Perinatal Cardiology for the Neonatologist" (Figure, left) for local distribution in 2011 and the book "Perinatal Cardiology: A Multidisciplinary Approach" (Figure, right) in 2015 for worldwide availability. These books and their contents were conceived in 2010 and some of the chapters were written in 2010 and 2011. However, the majority of chapters were written in 2013 and early 2014. Over these six to nine years, a number of advances in diagnostic studies, management concepts, methods of transcatheter intervention in the neonate, premature infant and fetus and concepts and methods of neonatal surgery, as well in anesthetic and post-operative management, have occurred. The purpose of this book, in addition to maintaining the objectives of the first book, is to

update and bring together the alluded advances to the attention of the reader.



This book attempts to bring the readers up-to-date evidence-based information in three major areas of neonatology and prenatal and neonatal cardiology. First, it provides an overview of the advances in the disciplines of Neonatology, Prenatal and Neonatal Cardiology, and Neonatal Cardiac Surgery in making early diagnosis and offering treatment options. Second, it describes the concept of a multidisciplinary approach to managing infants with congenital cardiac lesions. It provides evidence-based therapeutic approaches to successfully treating the fetus and the newborn with important neonatal issues and congenital cardiac lesions. Finally, it updates the authors' book on Perinatal Cardiology published in early 2015. Because of the large amount of material, the book is divided into two volumes.

P. Syamasundar Rao
Dharmapuri Vidyasagar

FOREWORD BY WILLIAM B. STRONG FOR THE FIRST EDITION

Pediatric cardiology has progressed as clinicians and scientists have had dreams of how it might be advanced. As advances in technology and miniaturization have occurred, clinicians have been able to apply their knowledge and skills to make the dreams reality. However, the technology would not have developed unless there were individuals capable of mastering it and courageous enough to apply it to the appropriate clinical situations. My forty-plus-year association with Dr. Syam Rao has provided me a great deal of opportunity to observe his mastery of new technologies, from M-mode echocardiography in the mid-1970s to the embracing of in the field of interventional pediatric cardiology in the latter years of the 20th century, in which he became a leader.

Dr. Rao's vision of the direction of pediatric cardiology has led to his acceptance that a great part of serious congenital heart disease is now managed in the perinatal period. It is for this reason that he and Dr. Vidyasagar are collaborating in the editing and writing of "Perinatal Cardiology." In addition to editing this multi-authored text, Dr. Rao has himself contributed numerous chapters based not only on his broad experiences, but also on the contributions of so many other colleagues who provide well-referenced texts.

This book is written not only for the pediatric cardiologist but for all those specialists and subspecialists who participate in a programmatic approach to the child with congenital heart disease, from the embryologist to the pediatrician or family practitioner to the hands-on caregivers in the tertiary care setting. The term programmatic is stressed because the care of the child with a significant congenital heart defect requires a team approach; no single specialist is capable of delivering all of the care these babies require. In addition, many of these children have other congenital abnormalities that will require other organ system specialists to participate in their care. Only a team approach with one physician being the coordinator will lead to these children being able to live "normal" and productive lives. The coordinating specialty physician, typically the

pediatric cardiologist, can then take control after the critical perinatal period to navigate the child through the remainder of infancy, into childhood, adolescence, and even into adulthood. The bonding of child and family with his or her caregivers is one of the greatest pleasures a physician can enjoy. That joy is highly dependent, however, on the professional teamsmanship of those early days of development and life. I believe that Drs. Rao and Vidyasagar and their colleagues have succeeded in illuminating this approach.

William B. Strong, MD,
Augusta, Georgia

PREFACE TO THE FIRST EDITION

Developments such as the early detection of neonates with serious heart disease and their rapid transport to tertiary care centers, availability of highly sensitive noninvasive diagnostic tools, advances in neonatal care and anesthesia, progress in transcatheter interventional procedures and extension of complicated surgical procedures to the neonate and young infant have advanced to such a degree that almost all congenital cardiac defects can be diagnosed and "corrected". The defects that cannot be corrected can be effectively palliated. Cardiac defects that were once fatal in infancy are now treatable and patients are surviving into adulthood. Because of the very optimistic prognosis of congenital heart defects, if diagnosed early on, a chapter devoted to the universal screening of all newborns using noninvasive pulse oximetry is included.

The last five decades have seen a great many advances, as mentioned above, resulting in the increased survival of infants with congenital cardiac lesions. These advances have occurred not only in the specialties of pediatric cardiology and pediatric cardiovascular surgery but also in several other disciplines such as neonatology, perinatology and ultrasonography. The growth of perinatology in tandem with neonatology has increased the survival of premature and extreme premature babies.

Advances in fetal echocardiography have enabled us to make in utero diagnoses of cardiac structural defects. The introduction of fetal surgery has led to successful in utero surgical correction of structural defects. Early postnatal diagnosis and advances in pediatric cardiac surgery supported by excellent postsurgical management by expert medical and nursing staff assure high survival in infants with many cardiac anomalies, including babies with hypoplastic left heart syndrome. Recent advances in medical and surgical therapy, particularly the application of "Fontan" principle, have markedly improved the long-term outlook of babies with single ventricle physiology. Palliative procedures to normalize the pulmonary blood flow (Blalock-Taussig or similar aorta-pulmonary shunt for patients with pulmonary oligemia, and pulmonary artery banding for patients with pulmonary plethora) and to relieve inter-atrial and/or inter-ventricular obstruction should be undertaken promptly. The Norwood

procedure or the Damus-Kaye-Stancel procedures are required for patients with single ventricle physiology. Staged total cavo-pulmonary connection (modified Fontan) to bypass the right atrium and right ventricle by bi-directional Glenn procedure initially, followed by extracardiac conduit diversion of inferior vena caval flow into the pulmonary arteries, appears to be the current procedure of choice in the surgical management of single ventricle lesions.

This book attempts to bring the readers up-to-date evidence-based information in three major areas of peri-neonatal cardiology. First, it provides an overview of the advances in the disciplines of Perinatology, Neonatology, Cardiology and Cardiac surgery in making early diagnosis and offering treatment options. Second, it describes the concept of a multidisciplinary approach to managing infants with congenital cardiac lesions. Finally, it provides evidence-based therapeutic approaches to successfully treat the fetus and the newborn with congenital cardiac lesions. Although every attempt was made to minimize the repetition, there are some areas, such as perinatal circulatory physiology and the management of babies with single ventricle physiology, that have had some degree of replication to preserve continuity of thought/discussion. Also, some issues are necessarily and completely neonatal in presentation, and they are presented for the sake of completeness.

The above discussed elements are important developments that obstetricians and pediatricians should be well aware of so that they can guide the prospective parents of infants with cardiac lesions to make the right decisions when faced with the diagnosis of an infant with congenital heart disease. In spite of the better prognostic outcome of infants with congenital heart defects, both parents and clinicians often face difficulties in making decisions in managing infants with critical congenital heart disease, raising ethical dilemmas; these are reviewed in a chapter of this book. Similarly, there is also a need for proper genetic counseling of parents undergoing a first-time experience of having a child with a congenital cardiac defect; this is discussed in another chapter.

For achieving excellence in cardiac care, however, close interaction and collaboration of the pediatric cardiologists with neonatologists, pediatricians and general/family practitioners (who care for children), anesthesiologists, pediatric intensivists, and cardiac surgeons is mandatory. The education of physicians caring for the newborn continues to be important in achieving optimal care for the neonate with heart disease. This book was designed

and produced with this in mind. We hope that this book will be useful not only to trainees, but also the Faculties in the Divisions of Pediatric Cardiology, Neonatology, Pediatric Cardiac Surgery and Pediatric Intensive Care. This would also serve as a resource to the Pediatric House-staff. Nursing and other para-medical staff in the Neonatal Intensive Care Unit, Pediatric Intensive Care Unit, and Pediatric Cardiac Intensive Care Unit and any pediatric cardiac unit will also find this book useful. The above discussions, we hope, will provide a fund of information to neonatologists, pediatricians and other physicians caring for newborns, which may help them provide better care for their babies.

P. Syamasundar Rao
Dharmapuri Vidyasagar

ACKNOWLEDGEMENTS TO THE FIRST EDITION

I have made generous use of case materials that I encountered over the years at the Medical College of Georgia, Augusta, Georgia; King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia; the University of Wisconsin, Madison, Wisconsin; Saint Louis University, Saint Louis, Missouri, and most recently at the University of Texas Medical School at Houston, Houston, Texas, in several chapters in this book. I wish to acknowledge the contributions of my pediatric cardiology, adult cardiology and cardiovascular surgery colleagues at these institutions and thank them for their patience and tolerance of many of my vagaries, particularly in my catheter-based interventional approaches.

It is of immense help to have Dr. Vidyasagar to help me edit this book; he brings a great neonatology perspective to our cardiology approaches. I sincerely thank Dr. Vidyasagar for co-editing the book with me; his contributions facilitated making this book more comprehensive.

Over the last four decades, I had the opportunity to train and guide several fellows and trainees and many of them not only contributed to the collected clinical materials, but also had stimulating queries which helped me to further focus on what is important. I also had the opportunity to take care of a number of patients and interacted with their parents, nurses and physicians. I thank these individuals for their willingness, courage and wisdom in participating in a number of clinical studies that I have undertaken; the knowledge so gained will serve generations of patients to come.

I would like to take this opportunity to thank the contributors of chapters in this book for their excellent reviews and perspective on the subjects they discussed. A number of echocardiographic pictures have been used as figures in many chapters of this book; a substantial portion of these studies were obtained at the Children's Memorial Hermann Hospital, Houston, Texas. I thank the sonographers for their diligence in securing high-quality pictures. A number of box diagrams of the heart illustrating the anatomy/physiology were reproduced from my publications in Neonatology Today and Congenital Cardiology Today. I sincerely thank Richard

Koulbanis, Publisher and Editor-in-Chief of Neonatology Today and Congenital Cardiology Today for granting us permission to reproduce these figures in this book.

P. Syamasundar Rao

SECTION I –

PERINATAL CIRCULATION

CHAPTER 1

PERINATAL CIRCULATORY PHYSIOLOGY

P. SYAMASUNDAR RAO, MD

Introduction

The fetal circulation uses the placenta for the exchange of respiratory gases while the lungs are utilized for gas exchange in the postnatal circulation. Consequently, the heart and lungs must adjust to these altering requirements. An appreciation of the fetal circulation and the alterations that it undergoes at birth is important for a better understanding of the postnatal adaptation of the circulation in various types of congenital cardiac defects (CHDs). In this chapter the author: 1) gives an outline of the fetal circulation; 2) discusses mechanisms that sustain fetal circulation; and 3) describes changes that take place immediately after birth.[1-5] The postnatal circulatory changes are likely to influence the presentation and neonatal course of important CHDs and will be reviewed in the next chapter. Normal circulations in the adult are shown in Figure 1-1 and in the fetus in Figures 1-2 through 2-4.