# Congenital Heart Disease in Pediatric and Adult Patients

## Anesthetic and Perioperative Management

Ali Dabbagh
Antonio Hernandez Conte
Lorraine Lubin
*Editors*

Springer

# Congenital Heart Disease in Pediatric and Adult Patients

Ali Dabbagh • Antonio Hernandez Conte
Lorraine Lubin
Editors

# Congenital Heart Disease in Pediatric and Adult Patients

## Anesthetic and Perioperative Management

Springer

*Editors*
Ali Dabbagh, MD
Cardiac Anesthesiology Department
Anesthesiology Research Center
Shahid Beheshti University of
Medical Sciences
Tehran
Iran

Antonio Hernandez Conte, MD, MBA
Kaiser Permanente Los Angeles Medical
Center
Division of Cardiac Anesthesiology
Los Angeles, CA
USA

Lorraine Lubin, MD
Department of Anesthesiology
Cedars-Sinai Medical Center
Los Angeles, CA
USA

ISBN 978-3-319-44689-9 ISBN 978-3-319-44691-2 (eBook)
DOI 10.1007/978-3-319-44691-2

Library of Congress Control Number: 2017933707



Printed on acid-free paper

This Springer imprint is published by Springer Nature
The registered company is Springer International Publishing AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

# Foreword

## Teach What You Practice!

There is an old educational aphorism, which has been used by educators for centuries*: Practice what you preach*. All too often teachers are somewhat distant from the educational frontlines. Although their opinions may be scientifically or evidence based, their edicts are not practical to clinicians treating complex patients in the real world (e.g., those with congenital heart disease). Rather, what the trainee (student, resident, or fellow) requires is an authoritative book edited and written by experts who manage these patients full time. They utilize evidence-based medicine to add to their clinical exposure and synthesize for the student an approach that "works." It is as if you are taking a master class with a master clinician whose "pearls" will make you a better doctor. Certainly, Drs. Ali Dabbagh, Antonio Hernandez Conte, and Lorraine Lubin represent an exemplary international composite of this type of physician. These are clinician-educators, who on a daily if not hourly basis interact with the patients with congenital heart disease.

From my own personal experience as a resident and cardiac anesthesia fellow, I desired to have a practical book that gave me a rapid overview of this field and allowed me to put my clinical observations in perspective. The editors have achieved just that. First, in 44 chapters, they take the reader from basic science to preoperative decision-making, monitoring and lesion-based intraoperative care, and finally care in the ICU setting. Second, the book is uniform in its presentation, showing the handiwork of the editors. This is the book you want available at 3 am in the morning when you called to manage the care of a patient undergoing an emergency surgical procedure or are facing a complex clinical dilemma in the ICU. Third, importantly, with the spectacular surgical results currently obtainable, pediatric patients are now living a productive life as adults. This means that many clinicians in nonacademic medical centers will be confronted by this reality. Thus, another group of physicians will benefit from the editors' and contributors' practical insights.

In this day of worldwide medical missions, the last chapter of this book deserves special mention. The coverage of *Pediatric Cardiac Anesthesia and Surgery in Developing Countries* is most likely unique to this book. This chapter will serve as an exceptional guide for use in settings where resources are limited.

In summary, the editors and contributors have skillfully fulfilled their mission as both *teachers* and *preachers*!

Paul Barash, MD
Professor of Anesthesiology
Yale University, School of Medicine
New Haven, CT, USA

# Introduction

Congenital cardiac disease encompasses a wide range of disorders that are usually diagnosed in the early infancy. The cardiac pathology may be life-threatening and require immediate intervention and/or surgery, or it may be less severe and allow for a series of interventions over the course of the patient's early childhood and into adulthood. In *Congenital Heart Disease in Pediatric and Adult Patients: Anesthetic and Perioperative Management*, we have collaborated with a vast array of leading cardiac clinicians from around the world who manage this patient population on a daily basis in order to highlight the subtle and not so subtle nuances. Their expertise allows for a practical approach in the management of a complex series of problems and issues.

*Congenital Heart Disease in Pediatric and Adult Patients: Anesthetic and Perioperative Management* is organized into six major sections. Each section describes a particular facet unique to this subspecialty and is designed to allow the clinician managing this patient population to rapidly become oriented with the specific pathologies and care issues.

*Part I* focuses upon the history of pediatric anesthesia as well as embryology and pediatric physiology and pharmacology.

*Part II* entails the technical requirements for diagnostic methods and for monitoring patients undergoing congenital corrective surgery.

*Part III* focuses upon the preoperative evaluation and considerations unique to patients with congenital heart disease.

*Part IV* describes in great detail the intraoperative care of patients with congenital heart disease with specific chapters on each of the congenital anomalies.

*Part V* expounds upon postoperative care of patients with congenital heart disease.

*Part VI* of the book addresses emerging trends and clinical care outside of the traditional operating room that is creating the new field of "hybrid" procedures for congenital issues. Also, the final chapter in this section discusses pediatric cardiac surgery in emerging countries.

We believe that *Congenital Heart Disease in Pediatric and Adult Patients: Anesthetic and Perioperative Management* will allow the reader to gain a general

and detailed knowledge base to optimally care for both pediatric and adult patients who undergo congenital heart surgery in the twenty-first century, both containing basic information and practical day-to-day data.

The editors would like to acknowledge the kind cooperation of all our contributors who have done their great work in preparing the chapters. Also, the kind cooperation and friendly patients of Springer authorities, especially Mr. Grant Weston and Mr. André Tournois, are highly acknowledged.

Tehran, Iran — Ali Dabbagh, MD
Los Angeles, CA, USA — Antonio Hernandez Conte, MD, MBA
Los Angeles, CA, USA — Lorraine Lubin, MD

# Contents

# Contributors

**Mohammad-Amin Abdollahifar, PhD** Department of Anatomy, School of Medicine, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Iki Adachi, MD** Division of Congenital Heart Surgery, Mechanical Circulatory Support, Texas Children's Hospital, Mechanical Circulatory Support, Baylor College of Medicine, Houston, TX, USA

**Felice Eugenio Agrò, MD** Anesthesia, Intensive Care and Pain Management Department, University School of Medicine Campus Bio-Medico of Rome, Rome, Italy

**Abdullah Amini, PhD** Department of Anatomy, School of Medicine, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Gerald A. Bushman, MD** Children's Hospital Los Angeles, Anesthesia Critical Care Medicine, Keck School of Medicine, Los Angeles, CA, USA

**Marcelo Cardarelli, MD, MPH** Pediatric and Congenital Cardiac Surgeon, Inova Children Hospital, Falls Church, VA, USA

William Novick Global Cardiac Alliance, Memphis, TN, USA

**Alessandro Centonze, MD** Department of Anesthesiology, Intensive Care and Pain Management, Campus Bio-Medico University Hospital of Rome, Rome, Italy

**Renzo Cifuentes, MD** Novick Cardiac Alliance, Universidad de Guayaquil, Guayaquil, Ecuador

**Antonio Hernandez Conte, MD, MBA** Division of Cardiac Anesthesiology, Kaiser Permanente Los Angeles Medical Center, Los Angeles, CA, USA

Cedars-Sinai Medical Center, Los Angeles, CA, USA

**Ali Dabbagh, MD** Cardiac Anesthesiology Department, Anesthesiology Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Filip De Somer, PhD** University Hospital Gent, Heart Center 5IE-K12, Gent, Belgium

**Elham Hashemi Dehkordi, MD** Pediatric Endocrinology, Child Growth and Development Research Center, Isfahan University of Medical Sciences, Isfahan, Iran

**Edmo Atique Gabriel, MD, PhD** Cardiac Surgeon, Coordinator and Professor of Medicine of União das Faculdades dos Grandes Lagos (Unilago), São José do Rio Preto, São Paulo, Brazil

**Sthefano Atique Gabriel, MD, PhD** Vascular and Endovascular Surgeon, Fellowship of Advanced Aortic Surgery at Ospedale San Raffaele, Milan, Italy

**Ruchira Garg, MD** Cedars-Sinai Medical Center, Los Angeles, CA, USA

**Mahmoud Ghasemi, MD** Pediatric Department, Kermanshah University of Medical Sciences, Kermanshah, Iran

**Dheeraj Kumar Goswami, MD** Anesthesiology and Pediatric Critical Care, Johns Hopkins Hospital, Baltimore, MD, USA

**Majid Haghjoo, MD** Department of Cardiac Electrophysiology, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran

**Mahin Hashemipour, MD** Pediatric Endocrinology. Endocrine and Metabolism Research Center, Isfahan University of Medical Sciences, Isfahan, Iran

**Alireza Imani, PhD** Department of Physiology, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

**Mohammadrafie Khorgami, MD** Department of Cardiac Electrophysiology, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran

**Lorraine Lubin, MD** Cedars Sinai Medical Center, Department of Anesthesiology, Los Angeles, CA, USA

**Stacey Marr, MSc, RN, NP** William Novick Global Cardiac Alliance, Memphis, TN, USA

**Ali Mazaheri, MD** Pediatric Department, Lorestan University of Medical Sciences, Lorestan, Iran

**Marion McRae, MScN** Cedars-Sinai Medical Center, Los Angeles, CA, USA

David Geffen School of Medicine, UCLA, Los Angeles, CA, USA

**Evelyn Carolina Monico, MD** Pediatric Anesthesia, Texas Children's Hospital, Baylor College of Medicine, Houston, TX, USA

**Neda Mostofizadeh, MD** Pediatric Endocrinology, Child Growth and Development Research Center, Isfahan University of Medical Sciences, Isfahan, Iran

**Pablo Motta, MD** Baylor College of Medicine, Texas Children's Hospital, Division of Pediatric Cardiovascular Anesthesiology, Houston, TX, USA

**Yamile Muñoz, MD** Cardiovascular and Thoracic Anesthesia, Novick Cardiac Alliance, Clínica Cardio VID, Medellín, Colombia

**Pooja Nawathe, MD** Cedars-Sinai Medical Center, Los Angeles, CA, USA

**William M. Novick, MD** Surgery and International Child Health, University of Tennessee Health Science Center, Memphis, TN, USA

William Novick Global Cardiac Alliance, Memphis, TN, USA

**Antonio Perez-Ferrer, MD** Department of Pediatric Anesthesiology and Intensive Care, La Paz University Hospital, Madrid, Spain

**Alistair Phillips, MD, FACC, FACS** Congenital Heart Program, the Heart Institute, Division of Congenital Heart Surgery, Division of Cardiothoracic Surgery, Department of Surgery, Cedars-Sinai Medical Center, Los Angeles, CA, USA

**Zoel Augusto Quiñónez, MD** Baylor College of Medicine, Texas Children's Hospital, Division of Pediatric Cardiovascular Anesthesiology, Houston, TX, USA

**Shahzad G. Raja, BSc, MBBS, MRCS, FRCS(C-Th)** Department of Cardiac Surgery, Harefield Hospital, London, UK

**Samira Rajaei, MD, PhD** Department of Immunology, School of Medicine, Tehran University of Medical Science, Tehran, Iran

**Michael A. E. Ramsay, MD, FRCA** Department of Anesthesiology and Pain Management, Baylor Research Institute, Baylor University Medical Center and Baylor Research Institute, Dallas, TX, USA

**Sri O. Rao, MD** Novick Cardiac Alliance, Memphis, TN, USA

**Noushin Rostampour, MD** Pediatric Department, Shahrekord University of Medical Sciences, Shahrekord, Iran

**Mohammad Ali Saghafi, MD** Anesthesiology Research Center, Cardiac Anesthesiology Department, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Jose E. Santoro, MD** Yale University School of Medicine, Yale New Haven Children's Hospital, New Haven, USA

**Jamie Wingate Sinton, MD** Anesthesiology Department, Baylor College of Medicine & Texas Children's Hospital, Houston, TX, USA

**Fahimeh Soheilipour, MD** Minimally Invasive Surgery Research Center, Iran University of Medical Sciences, Tehran, Iran

**Zahra Talebi, PharmD** Anesthesiology Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Ramin Baghaei Tehrani, MD** Pediatric Cardiac Surgery Department, Modarres Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

**Premal M. Trivedi, MD** Baylor College of Medicine, Texas Children's Hospital, Division of Pediatric Cardiovascular Anesthesiology, Houston, TX, USA

**David Freed Vener, MD** Department of Anesthesia and Pediatrics, Baylor College of Medicine and Texas Children's Hospital, Houston, TX, USA

**Marialuisa Vennari, MD** Department of Anesthesiology, Intensive Care and Pain Management, Campus Bio-Medico University Hospital of Rome, Rome, Italy

**Robert Wong, MD** Cedars-Sinai Medical Center, Los Angeles, CA, USA

**Ian Gavin Wright, MB ChB FCA(SA)** Department of Anaesthesiology, Intensive Care and Pain Management, Harefield Hospital, London, UK

# Part I
# History, Embryology, Physiology and Pharmacology

# Chapter 1
# History of Pediatric Anesthesia and Pediatric Cardiac-Congenital Surgery

**Antonio Hernandez Conte**

Pediatric surgery has a long history that dates back to the early twentieth century. In the early 1900s, pediatric medicine and surgery were undistinguishable from general adult surgical care, therefore, adult and pediatric patients were treated in a similar manner. Whereas in contemporary medicine, pediatric surgery is a completely separate specialty with different training pathways compared to a surgeon treating adult patients. It was soon discovered that the mortality rates in the younger population were extraordinarily high and that if improved results were expected, the pediatric patient would need a separate treatment approach.

Pediatric patients have traditionally been defined as patients under the age of 18; however, as will be discussed in later chapters, this age demarcation has once again become less defined as patients who manifested congenital heart disease grow into adulthood and require additional cardiologic or cardiac surgical interventions. Pediatric cardiac and congenital surgery focuses upon the surgical correction of major anomalies pertaining to the heart and surround vascular structures. As the subspecialty of pediatric surgery evolved in the mid-1900s, pediatric surgical care becomes more commonly based at children's specialty hospitals throughout Europe and the United States. In the period of less than 50 years, the initial development and evolution of medical and technical advances led by key scientists and physicians focusing upon care of the pediatric patient allowed the fields of pediatric surgery, pediatric cardiac/congenital surgery, and pediatric anesthesia to become a mainstay of modern-day medicine.

A. Hernandez Conte, MD, MBA
Division of Cardiac Anesthesiology, Kaiser Permanente Los Angeles Medical Center, Los Angeles, CA, USA

Cedars-Sinai Medical Center, Los Angeles, CA, USA
e-mail: sedated@bellsouth.net; antonio.conte@kp.org



A. Dabbagh et al. (eds.), *Congenital Heart Disease in Pediatric and Adult Patients*, DOI 10.1007/978-3-319-44691-2_1

## The Birth of Pediatrics and "Children's Hospitals" in the United States

The status of pediatric care in the 1800s and early 1900s was profoundly different than current standards. As mentioned earlier, pediatric patients were in essence treated in a manner similar to adults. Some examples of prevailing treatments and trends included lack of understanding of intravenous therapy, and fluid balance was based on adult models. Additionally, blood transfusions were not utilized, and appendicitis was the fourth common cause of death in children. The most common surgical procedures in children were abscess drainage, appendectomies, tumors, and hernia repairs. There was 90 % mortality for colostomies and intussusceptions. With the concomitant medical and surgical advances that had begun in the mid-1930s and continued thereafter in each decade, pediatric care was finally becoming entrenched in newly formed children's hospitals.

Dr. Abraham Jacobi is considered to be the father of pediatrics in the United States and offered the first lectures on pediatric disease in 1860. Generally speaking, adult medicine focused upon organ issues or technology. Dr. Jacobi believed that children warranted a broader approach with respect to child health and well-being and not just disease states. In 1880, Dr. Jacobi along with a few other physicians founded the American Medical Association's section on the diseases of children. They stressed the need for more children's hospitals and for the expansion of pediatric content in medical school curricula. By 1900, ten schools of medicine had full-time pediatricians.

In the pre-1850s, US hospitals generally had no place for children outside of the maternity ward. Childhood illnesses were therefore most often handled at home. When families sought medical care outside of their home, children were treated as tiny adults. This view lead to high infant and children mortality rates who had been admitted to hospitals, due to improper medical care. Dr. Francis West Lewis visits the Great Ormond Street Hospital for Sick Children in London. At the time this was the leading institution in the world for not only pediatric care but also the education of practitioners in the field. Dr. Lewis was inspired to bring what he saw in London to the United States. Lewis and his colleague Dr. Hewson Bache begin to work on what has now become the Children's Hospital of Philadelphia.

In 1850, the Children's Hospital of Pennsylvania (CHOP) in Philadelphia opened its doors. By 1871, CHOP was performing its first pediatric surgeries, and in 1871 the first pediatric-centered medical training program was established. Surgical clinics were created to train surgeons who were now working at the few emerging children's hospitals across the country. As early as the 1870s, physicians at the Children's Hospital of Philadelphia, for example, pressured the lay trustees who managed the hospital to increase patient turnover and accept more acutely ill children, especially orthopedic surgical patients who had something to offer physician education and on whom new surgical techniques and therapies could be tried. This new emphasis on the medical needs of patients and the experimental needs of doctors and nurses conflicted with the social welfare role children's hospitals saw themselves as performing.

The Children's Hospital in Boston was the second such specialty hospital in the United States and admitted its first patient in 1869. By the 1920s and 1930s, The Children's Hospital in Boston was becoming a major center for advanced care of the pediatric patient while also performing novel and innovative procedures for the first time. Advancements at The Children's Hospital in Boston have continued, and notable accomplishments include the first correction of hypoplastic left heart syndrome (1983) and the first pediatric open-heart transplant (1986).

## Surgical Pioneers in Pediatric Surgery

*William E. Ladd* is often referred to as the *father* of pediatric surgery. A Harvard-educated physician, Dr. Ladd's career path was dramatically altered after being dispatched by U.S President Lowell to treat the approximately 9000 victims who were innocent bystanders at the accidental collision of two ships in the Halifax Harbour in Nova Scotia in 1917. The explosion was the most powerful nonnuclear explosion that had ever occurred in history, and 4% of the population of Halifax was killed instantly. Dr. Ladd was sent on one of the first trains deployed to the accident site in Nova Scotia, and his experiences in Halifax had a profound effect upon him. After returning to Boston, Dr. Ladd devoted himself entirely to the surgical care of infants and children. Dr. Ladd recognized that children needed a very gentle and thorough physical evaluation, and surgeons needed to rely upon their own senses. Additionally, adult surgical instruments were not suitable for children, and he began to develop appropriately sized instruments.

In 1918, Dr. Ladd became an instructor in surgery at Harvard Medical School, and by 1927, he was named the chief of surgery. In 1931, Dr. Ladd became a full Harvard Medical School Professor, and in 1941, he published the seminal textbook entitled *Abdominal Surgery of Infancy and Childhood*. As quoted by Donald Watson, "Dr. Ladd brought the diagnosis and management of surgical lesions of infancy and childhood into new perspective." Indeed Dr. Ladd's pioneer efforts truly initiated pediatric surgery as a separate discipline in the Western Hemisphere." Physicians who trained under Dr. Ladd in Boston perpetuated the specialty and achieved their own independent successes to further cement and validate the emerging specialty. These physicians included Drs. Robert E. Gross, Theodore Jewett, Earle Wrenn, Donald Rooney, C. Everett Koop, and Monford Custer—all of them continued to advance the field of pediatric surgery in the twentieth century.

## Origins of Pediatric Anesthesiology

The subspecialty of pediatric anesthesia has significantly evolved since its origin in Jefferson, Georgia, when Dr. Crawford Long administered the first documented ether anesthetic to an 8-year-old boy for a toe amputation on July 3, 1842. From the very beginning, it was clear that children were at higher risk than adults for