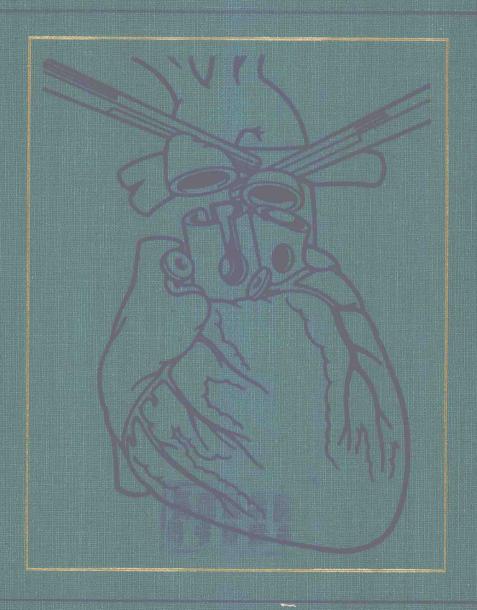
# CARDIAC ANESTHESIA for INFANTS and CHILDREN



Jay Kambam

# CARDIAC ANESTHESIA

# for INFANTS and CHILDREN

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Professor of Anesthesiology, Department of Anesthesiology, Vanderbilt University School of Medicine, Nashville, Tennessee Dedicated to my wife, Veni, and sons, Shravan and Praveen.

### Foreword

The surgical management of patients with congenital cardiac malformations had its infancy in the 1940s with the development of operative techniques to manage patent ductus arteriosus, coarctation of the aorta, and the creation of systemic pulmonary arterial shunts. From that time there has been rapid development of the specialty, particularly following the advent of successful extracorporeal circulation and the understanding of the benefits derived from hypothermic diminution in oxygen requirement during periods of circulatory arrest.

It has been my good fortune to be involved in the surgical management of patients with congenital cardiac malformation since the 1950s. The advances that have been made in the management of these desperately ill infants and children has been phenomenal, with tremendous improvement in survival figures for even the most complex intracardiac malformations. When one looks back at the evolution in surgical procedures and the management of patients in the perioperative period, it is difficult to single out specific changes that were instrumental in the resultant improvement in survival. Obviously important among those improvements were the better understanding of the pathologic anatomy of the various malformations and the ability to perform precise reparative procedures even in the smallest neonate. In addition to the obvious operative advances, the understanding and the ability to measure arterial blood gases, cardiac output, and electrolyte changes rapidly had a significant role in the perioperative management of these patients.

Commensurate with the development of children's cardiac surgery as a specialty has been the development of pediatric cardiac anesthesiology. The pediatric cardiac anesthesiologist's understanding of the pathophysiologic changes in infants with congenital cardiac malformations has been ex-

tremely important. The role of ventilation in the management of oxygen and CO<sub>2</sub> concentrations with respect to pulmonary vascular resistance is but one example. The drugs that are now available for the conduct of anesthesia and the management of patients in the postoperative period has had enormous impact on the survival of infants with complex cardiac malformations.

This textbook was written with the idea that it would be useful to students and house officers in anesthesiology. It was thought that it would provide a broad picture of the population of patients with congenital cardiac disease and the pathophysiologic changes related to the various specific abnormalities. As the book has evolved, it has become directed to a broader audience. The authors have managed to address each of the major groups of malformations with respect to the changes in pulmonary blood flow, changes in pulmonary vascular resistance, the degree or absence of cyanosis, and the degree or absence of congestive cardiac failure that may be present. In each chapter the authors have outlined the embryology, the pathologic anatomy and the changes in physiology that will have direct impact on the administration of anesthetic agents. As a result, this book will be of considerable interest to surgeons and surgical students as they approach patients with congenital heart dis-

I am confident that this book will be well received by pediatricians, surgeons, and anesthesiologists, and I am honored to be asked to write this foreword.

Harvey W. Bender, Jr., M.D.
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## Foreword

—Ah, what would the world be to us
If the children were no more?
We should dread the desert behind us
Worse than the dark before.—
H.W. Longfellow: "Children"

It has been said that as many as 1% of all infants are born with some degree of congenital heart anomaly, and that, uncorrected, these would result in approximately 40% mortality before the fifth year of life. Yet surgical correction of these anomalies was not attempted until the 1939 report by Gross and Hubbard of the surgical ligation of a patent ductus arteriosus, and even palliative surgery awaited the 1945 description by Blalock and Taussig of their shunting procedure.

The application to children of extracorporeal techniques approximately similar to current practice was not reported in a significant series until 1955 by Kirklin et al. However, by 1966 surgical mortality was still as great as 30% for relatively conservative repairs of major cardiac anomalies even at leading centers. The refinement process continued from approximately 1972 to the present until pediatric cardiac surgery reached a point where such ominous lesions as transposition of the great vessels have now been repaired in significant numbers of patients with a mortality rate less than 6%.

Today techniques as formidable as intentional deep hypothermic cardiac arrest have become so routine in children as to allow a series of nearly 160 cases to be collected within 26 months in the editor's hospital.<sup>12</sup>

Although they have been the leaders and the risk takers in achieving this progress, cardiac surgeons have been enthusiastic in enrolling help from a field of specialists ranging from physicians to engineers. Thus the flowering of congenital cardiac surgery has been a remarkable team effort. Anesthesiologists have contributed much to this progress, not only in the operating room, but also by their participation in the development of postoperative mechanical ventilation8 and other critical care support methods.4 However, specific advances in diagnosis such as enhanced radiographic and radionuclide procedures13 and expanded uses of echocardiography10; new pharmacology for preoperative, intraoperative, and postoperative medications such as prostaglandin<sup>11</sup> and indomethacin infusions<sup>7</sup>; the appearance of a variety of useful new catecholamines, positive inotropes, and receptor blockers and agonists; and a variety of advances in laboratory medicine, materials, engineering, extracorporeal life support disciplines<sup>14</sup>, and many other sources, have each made critical contributions.

More prominently than in many other areas of medicine, many of these advances have been made by the astute observations and expressed opinions of busy experienced clinicians in all of these fields, because of the relatively small numbers of patients and the intrinsic difficulty of controlled systematic research concerning alternative therapies. Perhaps fewer than 15,000 total surgical invasions for congenital heart problems requiring extracorporeal life support are required annually in the United States even now, and certainly far fewer were attempted per annum in the recent past. <sup>16</sup> Furthermore, objective research in the presence of a seriously ill child is frequently not technically difficult, but often is not ethically supportable.

Because of these limitations, the format chosen by the editor, Professor J.R. Kambam, is particularly appropriate and effective in delineating the problems of anesthesia in cardiac surgical patients. He brings to the task extensive practical personal experience in anesthesia for the surgery of congenital heart lesions gained in one of the busiest and most successful centers. In addition, he has chosen co-authors who, although many are internationally recognized authorities in their own field, are also busy clinicians, and most of whom are members of the same team in the same center. Thus the descriptions of the basic background, as well as the narration of diagnosis, therapy, and surgical procedures, are all formulated and influenced by the experience of a team of busy and practical clinician academics who have familiarity with the literature and with the actual practices of their coauthors in the care of these patients.

Professor Kambam has successfully coordinated the literary efforts of this authoritative group of friends into an enjoyable and concise, yet thorough, treatise. Enormous credit should also be acknowledged to the surgeon who has coordinated the efforts of this team through the past two decades, whose surgical achievements are enviable, and who has immeasurably influenced the philosophies expressed in this book, Professor H.W. Bender.<sup>2</sup> This book will be valuable not only to the anesthesiologist, but to surgeons, specialists, generalists, and nursing professionals who wish to become familiar with the many ways in which the anesthesiologist's involvement impacts the total care of patients with congenital heart defects. The editor, his co-authors, and colleagues are to be congratulated!

> Bradlev E. Smith, M.D. Professor of Anesthesiology Vanderbilt University Medical Center Nashville, Tennessee

#### REFERENCES

- 1. Bender HW Jr, Stewart JR, Merrill WH: Ten years' experience with the senning operation for transposition of the great arteries: physiological results and late follow-up, Ann Thorac Surg 47:218, 1989.
- 2. Bender HW Jr: Preparation, trust, and responsibility, Ann Thorac Surg 51:351, 1991.
- 3. Blalock A, Taussig HB: The surgical treatment of malformations of the heart in which there is pulmonary stenosis or pulmonary atresia, JAMA 112:189, 1945,
- 4. Gregory GA, Edmunds LH, Kellerman JA et al: Continuous positive pressure and pulmonary and circulatory function after cardiac surgery in infants less than three months of age, Anesthesiology 43:426, 1975.
- 5. Gross RE, Hubbard JP: Surgical ligation of a patent ductus arteriosus: report of first successful case, JAMA 112:729,

- 6. Hamilton DI: The surgery of congenital heart defects. In Lester J, Irving IM, editors: Neonatal surgery, London, 1990, Butterworths.
- 7. Heymann MA, Rudolph AM, Silverman NH: Closure of the ductus arteriosus in premature infants by inhibition of prostaglandin synthesis, N Eng J Med 295:530, 1976.
- 8. Kirby RR, Robinson EJ, Shultz J et al: A new pediatric volume ventilator, Anesth Analg 50:533, 1971.
- 9. Kirklin JW et al: Intracardiac surgery with the aid of a mechanical pump-oxygenator system (Gibbon type): report of eight cases, Mayo Clin Proc 30:201, 1955
- 10. Muhiudeen IA, Roberson DA, Silverman NH et al: Intraoperative echocardiography for evaluation of congenital heart defects in infants and children, Anesthesiology 76:165, 1992
- 11. Olley PM, Coceani F, Bodach E: E type prostaglandins: a new emergency therapy for certain cyanotic heart malformations, Circulation 53:728, 1976.
- 12. Phythyon JP: Deep hypothermia and circulatory arrest. In Kambam J, editor: Cardiac anesthesia for infants and children, St Louis, 1994, Mosby.
- 13. Rao PS, Wilson AD, Sideris EB: Transcatheter closure of patent ductus arteriosus with "buttoned" device: first successful clinical application in a child, Am Heart J 121:1799, 1991.
- 14. Roberts CP, McCarthy P, Wildman D et al: Left ventricular assist with the biomedicus pump on a 4-month-old infant with anomalous left coronary artery, J Ext Corp Tech 21:73, 1989
- 15. Strong MJ, Keats AS, Cooley DA: Anesthesia for cardiovascular surgery in infancy, Anesthesiology 27:257, 1966.
- 16. Vinas MS: Extracorporeal circulation. In Kambam J, editor: Cardiac anesthesia for infants and children, St Louis, 1994, Mosby.

## Preface

Anesthesia for surgery involving congenital heart defects has experienced tremendous breakthroughs over the past 15 years. The creation of *Cardiac Anesthesia for Infants and Children* has been based on several personal communications with friends in the field of anesthesiology, particularly cardiac anesthesiology. The purpose of this book is to provide a single source of the information necessary for optimal perioperative care of an infant or a child with congenital heart disease. As a cardiac anesthesiologist for more than 15 years, I have long felt the need for such a book.

Cardiac Anesthesia for Infants and Children was written by groups of pediatric cardiologists, cardiac surgeons, and pediatric anesthesiologists. It provides basic knowledge of the subject, as well as guidelines for perioperative management of a child with congenital heart disease undergoing cardiac or noncardiac surgery. The latest advances in pediatric cardiac surgery and anesthesia are described from several vital perspectives, including embryology, pathologic anatomy and physiology, medical and surgical management, and anesthesia management. Figures and drawings were designed for easy understanding of pathologic anatomy and physiology of complex congenital heart lesions.

Most chapters involving particular malformations were written by groups of pediatric cardiologists, surgeons, and anesthesiologists. Thus each chapter provides a balanced analysis of its subject, and each is presented in a coherent manner by experts familiar with the latest research. These chapters were deliberately designed to be read separately rather than in a generalized fashion (for example, L-to-R shunt lesions). Though this book was primarily written for anesthesiologists at all levels, I believe pediatricians and pediatric surgeons will also find it very useful.

We have written this book in a period of one year, and I wish to thank all of the contributors who have worked so hard to finish their chapters in such a short time. I also would like to thank Kathleen Finn and Steven Wasserman for their editorial advice, Paul Gross for the outstanding drawings he produced for this book, Susan Britt for her secretarial assistance, and all the publishers and their authors who gave us permission to reproduce figures and tables. A special thank you goes to Susan Gay and Sandy Clark of Mosby—Year Book publishers for their help and support in creating this book.

Finally, I must express my deep appreciation to my wife, Veni, for her patience and support during the hectic period of preparation of the book.

Jav Kambam

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## PART ONE

# **General Principles**