SURGERY FOR CONGENITAL HEART DEFECTS

Third Edition

Editors
Jaroslav F. Stark
Marc R. de Leval
Victor T. Tsang

Illustrations by Michael Courtney

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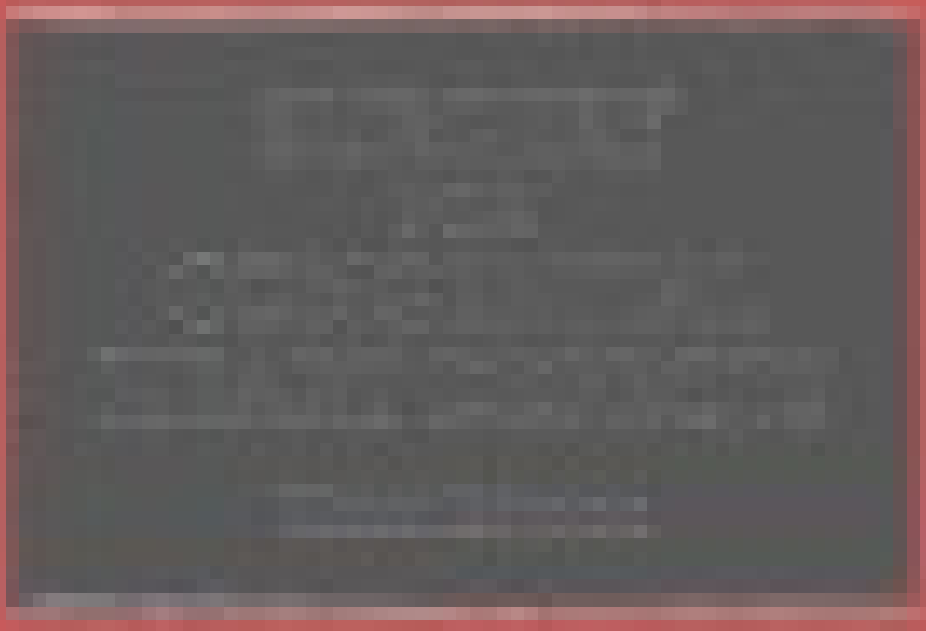

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Dedicated to the Children with Congenital Heart Disease

Preface to the Third Edition

The second edition of *Surgery for Congenital Heart Defects*, published in 1994, has been out of print for some time. We did not think that enough changes had occurred during the past 10 years to warrant another edition. However, our trainees and attendants of courses, particularly those attending the European School of Cardiothoracic Surgery in Bergamo, Italy, were urging us to do so.

When we reviewed the progress of the past decade, many new diagnostic techniques had been introduced, several aspects of anaesthesia, perfusion and postoperative care had been refined and a number of operative procedures had been modified, with emphasis on complex small infant surgery. For all these reasons we agreed to update the last edition.

Some chapters are completely new, such as those on MRI and CT scanning, interventional catheterization, electrophysiology of arrhythmias, paediatric cardiac mechanical support, lung transplantation and assessment of surgeons' performance. The last chapter was added to help readers to understand the difficulties and pitfalls of data collection and the importance of accurate data for everyday work.

All other chapters were revised to reflect the current practices of the editors and leading specialists in the field of congenital heart disease contributing to this edition. Major changes and updates were made in the chapters on echocardiography, invasive investigation and exercise testing, anaesthesia, postoperative care, vascular rings, anomalous pulmonary venous return, tetralogy of Fallot, pulmonary atresia with ventricular septal defect, surgery of left ventricular outflow tract atrio-ventricular discordance, hypoplastic left heart syndrome and paediatric heart and heart lung transplantation.

The format of the book follows that of the two previous editions. The emphasis remains on the details of operative procedures. Diagnostic criteria are briefly discussed in each chapter. Results are summarized for completeness. Fortunately the same artist, Michael Courtney, provided most of the additional illustrations. In the general section, readers should find information about nomenclature, anatomy, diagnosis, including invasive investigation, exercise testing, echocardiography and foetal echocardiography, anaesthesia, perfusion, postoperative care, pacing, electrophysiology, cardiac mechanical support and approaches to the heart. In the section on surgical procedures, we have kept some classical operations, even though some of them are currently performed only rarely or even exceptionally. Because of the rare use of these procedures, it is not easy for young surgeon to familiarize themselves with such procedures during their training.

We hope that this updated edition will again be helpful to trainees in paediatric cardiac surgery and allow them to start their independent careers without learning curves. It may also assist established surgeons when dealing with rare defects. Paediatric cardiologists, anaesthetists, intensivists, nurses and technicians may find the book a useful source of information.

Jaroslav F. Stark
Marc R. de Leval
Victor T. Tsang

Acknowledgements

We are grateful to all the contributors for their outstanding work. We are particularly indebted to Michael Courtney, whose illustrations are such an integral part of the book.

We would like to thank our research secretary Faith Hanstater, and in particular our secretary Juliet Aghion for her help with the manuscripts, organization of all the details of the work and her general support. Our researcher, Katy Burke, helped greatly with the sourcing and organisation of the literature.

We thank our team of cardiologists, anaesthetists, intensivists, nurses, physiotherapists and technicians, who have participated with us over the years in the care of our patients.

Finally, we are extremely grateful to Haymans Trust for their generous financial support, which helped with the secretarial support and additional illustrations required for this up-dated edition.

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Foreword to the Third Edition

The Hospital for Sick Children at Great Ormond Street (GOS) in London, England (currently Great Ormond Street Hospital for Children) has been at the forefront of paediatric cardiac surgery since its inception. Throughout much of GOS history, two of its surgeons, Jaroslav F. Stark and Marc R. de Leval, have contributed enormously to the clinical practice, research and teaching of congenital heart surgery.

Their textbook *Surgery for Congenital Heart Disease* was first published in 1983, was revised in 1994, and again revised in 2006 as this 3rd edition. The book has become a reliable and trusted friend. It has a proven ''track record'' and contains sage and practical advice for the present and future progress of our specialty.

The original publication grew from the authors' teaching sessions. In the preface to the 1983 edition, they state; ''*The successful outcome of many operations depends to a large extent on the careful planning and precise execution of a series of minor technical steps*''. Stark and de Leval provide a recipe for successful surgical outcomes by clearly defining the technical details that are often overlooked or assumed in other textbooks. In the current edition, Stark and de Leval have added a new editor, their younger colleague Victor Tsang, who continues their tradition of excellence in clinical surgery and clear unambiguous description of the essence of surgical care for children with congenital heart disease.

Core authorship of the book is the outstanding faculty at GOS. In addition, selected colleagues outside GOS have provided contributions in their areas of special expertise, thereby adding to the book's international scope and authority. The format of each chapter is consistent with emphasis on the practical. The artwork by Michael Courtney complements the text and greatly facilitates comprehension of the essential operative details which are the stated mission of the book.

The past few decades have produced remarkable improvements in the care for patients with congenital heart disease. Newborns with cardiac lesions that were rapidly fatal when the two previous editions of this textbook were conceived are now successfully palliated into adulthood. The functional ability and quality of life for all of our patients is improving. Yet much remains to be accomplished and will be, as we incorporate new science and technology into surgical practice. The essence of good surgical practice will remain the same and is clearly described in this text. The GOS tradition of excellence in clinical surgery and teaching lives on in this 3rd edition of *Surgery for Congenital Heart Disease*. It is a privilege to review the final draft of this valuable contribution to the care of children with congenital heart disease.

WILLIAM G. WILLIAMS, MD, FRCSC
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Foreword to the Second Edition

This second edition of the masterful *Surgery for Congenital Heart Defects* is a worthy successor to the first edition of that great work, which appeared in 1983. Drs Stark and de Leval have designated themselves 'editors,' but in fact they clearly are both the stimuli for the second edition and the main contributors to it. In fact, this important book could be considered a detailed summary of all aspects of the experience of their prestigious unit, The Hospital for Sick Children at Great Ormond Street, London, with congenital cardiac disease over a period of at least 40 years. This alone recommends this text to all serious students and practitioners of the art and science of managing patients with these conditions. The fact that the book describes far more than simply operations and their results emphasizes the multidisciplinary nature of all serious endeavors in this area; good surgery alone can no longer suffice as evidence of institutional expertise in the area of congenital cardiac disease.

The book is clearly more broadly based than it would be had it been written only by the staff at the Great Ormond Street hospital. In a sense it could be considered a contribution of London, England, to the expanding knowledge of all aspects of congenital heart disease; the major contributions from this area over a period of many years also make the text invaluable. In addition, however, Drs Stark and de Leval have selectively tapped the resources of the rest of the medical world in developing this superb text.

This book becomes available at a particularly important and probably pivotal time in the area of congenital heart disease. In some parts of the world, there now exists the capability of eliminating congenital heart disease from the population, using prenatal echocardiographic diagnosis and abortion of an affected fetus. Ironically, this comes at a time when the results of many types of interventions for congenital cardiac disease are outstandingly good. For some types of congenital heart disease, literal cures by surgery have been documented. In contrast, it is now known that some conditions (for example, coarctation of the aorta) that seemed to be cured by intervention are only palliated, but this palliation may last for 50 years or more. Certain congenital cardiac anomalies that only a few years ago were essentially untreatable, such as interrupted aortic arch and aortic atresia, now can be at least extremely well palliated by one of several methods, and this palliation can be expected to last for a number of years even though not for the usual lifetime of a normal individual. In view of the diversity in the time-related quality of the outcomes, the variety of the interventions available for various types of congenital heart disease, and the many decisions that currently must be made in managing patients with congenital heart disease, it now becomes essential to quantify and compare outcomes. Also, it may become necessary somehow to pick and choose between institutions because of the growing suspicion that a concentration of relatively large numbers of such patients in relatively few institutions may improve outcomes. This again should involve quantification of outcomes, sophisticated methods of analyses, and factual (rather than simply political) comparisons of institutions and physicians.

Were this not enough to justify the extensive material presented in this text, society in general has added quality of care, appropriateness of care, effectiveness of care, and cost of care to its jargon of qualifications to be met by all of us in our hospital wards, laboratories, operating rooms, and intensive care units. It is this, plus the alleged imperfections in the outcomes we obtain and their high costs, along with the changing lifestyles and philosophies of our times and the strong emotions that accompany all discussions of health care (let alone those of abortion, nonuseful extension of life, and physician-assisted death), that makes so necessary the gathering together of the varied and extensive information presented in this second edition of *Surgery for Congenital Heart Defects*.

I look forward to detailed study of each of the wonderful chapters that have been presented in this book. Indeed, I recommend such study to everyone involved with decisions and management for that part of our populations afflicted with congenital heart disease.

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Foreword to the First Edition

The entire discipline dealing with the surgical treatment of congenital heart disease is unique in retaining much of its original awesomeness. Perhaps this is because the act of opening a child's chest to manipulate in or about the heart still evokes the feeling that this is an incredibly impertinent thing to do. Even after all these years, the act seems to fly in the face of that which is natural and possible. Perhaps it is because of the great gravity of the procedure, the sometimes narrow margin between failure and success, with an entire unlived lifetime in the balance. Or perhaps it is because the surgical capability to invade the heart developed more recently than for any other organ—indeed, within the professional lifetime of some of its still-pioneering pioneers.

It is unquestionably true of each of the surgical disciplines that success or failure is related to the availability of fundamental knowledge, advanced technology and artistic skill. But perhaps those who are involved in surgery of congenital heart defects could be forgiven for observing that this axiom relates most particularly of all to this discipline. The structural complexity and functional intricacy of the cardiovascular system are exceeded by only one other, the central nervous system, which is so exalted in these respects as to remain relatively intractable to all but ablative surgical interventions. Of all other systems, with regard to complexity, none seem to compare embryologically, anatomically, physiologically, or electronically with the heart. And within the cardiovascular system itself, the subclass of conditions which has a congenital aetiology greatly surpasses other forms of heart disease with respect to multiplicity of pathological presentations, diversity of combinations of associated lesions, and requirements for ingenuity and precision of surgical technique and patient care.

The congenitally deformed heart poses special obstacles to corrective operation. The necessity for growth is particularly important, especially when prosthetic valves or conduits are required, and true also for cardiac transplantation or perhaps ultimately the use of an artificial heart. Pertinent also, is the problem of secondary effects of the congenital anomaly on pulmonary or peripheral vasculature, or on the myocardium itself.

A situation *par excellence* which demonstrates the necessity of sophisticated teamwork in modern medical practice is provided by groups which are successful in the management of patients with congenital heart disease. Collaboration, coordination, communication, cross-fertilization and mutual esteem should be in evidence among participants of this team, which includes a cardiologist, radiologist, physiologist, pathologist, anaesthetist, nurse, technician, surgeon and several other partners. Perhaps as well as being the most ''incisive'', the surgeon's role in this team is also highly decisive. It is necessarily upon the surgeon's shoulders that the principal burden of the operative intervention must primarily rest. He or she must feel the weight of having accepted from the parent that strongest of all possible human obligations—the profound, primordial, instinctive identification and responsibility for an offspring. Surely no more depth of sincerity can be imagined than that expressed in the ''Thank you'' of a parent for a safe and successful surgical endeavour, nor more pain, frustation and despair if the outcome should be the opposite. By virtue of the complexity of the many problems to be dealt with, the precision required during all phases of patient care, and the immense parental trust resting upon them, the team dealing with congenital heart disease sometimes finds the demands on reserves of physical, emotional and spiritual strength almost overwhelming. Yet the incomparable sense of reward which is frequently achieved compensates and justifies all.

Some 40–45 years are all that have been required for progression from that first adventure of cardiac surgery to the present when virtually every anomaly is amenable to some form of surgical intervention. During this time, the advance of one ingenious approach after another has stirred excitement and expectation, and the prospect of further refinements and innovations continues. However, at this point, one senses that a landmark may have been

reached. Techniques, approaches, results and remaining issues of concern all seem to have reached a tentative landing on the staircase of progress. This pause presents, perhaps for the first time, a splendid opportunity to survey the subject broadly and thoroughly. The authors of this book are well suited to the task from the standpoint of their multi-disciplinary diversity, their uniformly superb qualifications and their substantial individual contributions to this body of knowledge. The challenge of condensing the mass of information surrounding each aspect and anomaly, while retaining comprehensiveness, has, I think, been expertly accomplished.

It gives me honour to have the opportunity to present these reflections as a preliminary to this fine text. It gives me pleasure to congratulate all of those who contributed in either small or large measure to the remarkable achievements to which it attests. To its authors, I heartily extend my respect and gratitude.

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