Color Atlas of Congenital Heart Surgery

Second Edition

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To Judy
my wife and best friend
without whose love and companionship
even my career in congenital heart surgery
would be almost meaningless
and
to my daughters
Amy Jessica and Jill Andrea
whom I love and respect for their
intelligence, talent, inner and outer beauty,
and commitment to making a contribution
in their respective fields.

Foreword

The last three decades have witnessed enormous progress in the care of patients with congenital heart disease. Treatment of congenital heart disease is highly dependent on technology and much of the progress we have witnessed is attributable to technological advances we take almost for granted today. It would be difficult to overestimate the impact of these advances; noteworthy examples include the development of 2-D Doppler echocardiography resulting in increased diagnostic accuracy, improvements in preoperative management including the use of prostaglandins for maintaining ductal patency, better intraoperative support such as the development of cardiopulmonary bypass circuits specifically designed for neonates and infants and improvements in postoperative care too numerous to delineate. As the spectrum of congenital heart disease we can treat successfully has broadened and the results have improved much of the focus has shifted, properly, to long-term issues such as neurodevelopmental outcome and quality of life. Despite the current focus on long-term outcomes we must not forget that surgery is central to our treatment strategy. The word technology is derived from the Greek word "techne" meaning craft and before any late outcomes can be measured the "craft" of surgery must be performed with excellence.

Dr. Litwin's career has spanned these last three decades and he has witnessed and participated in the evolution of congenital heart surgery. The second edition of a "Color Atlas of Congenital Heart Surgery" is an outstanding contribution to the field by a master of the craft of congenital heart surgery. Most textbooks on congenital heart surgery rely on drawings to illustrate the operations. These drawings are the imperfect recollections of the author's experience; smoothed and cleaned of imperfections they sometimes oversimplify the operative technique and may bear only a passing resemblance to reality. A cliché has thus been coined "it never looks like it does in the book". In stark contrast, Dr. Litwin's atlas is a true-to-life depiction of the anatomy and steps required for each of the procedures. The photographs truly speak for themselves; they require little description as they demonstrate each step of the procedure. The images are testimony to the meticulous technique that Dr. Litwin is known for and which I have had the good fortune to witness and learn from first hand. In the present era when video sessions are an increasing part of surgical meetings, the "Color Atlas of Congenital Heart Surgery" is both avant-garde and historic. Operations that are part of the day to day activities of congenital heart surgery programs today such as the arterial switch and stage I palliation of hypoplastic left heart syndrome are illustrated but in addition the Senning procedure is also included. The current students of congenital heart surgery who must train and begin their careers in a complex environment marked by two opposing forces, one of increasing technical difficulty of the cases and the other a decreasing tolerance for any sort of learning curve, will find this book invaluable as it provides both detailed photographs of

real operations and a breadth of procedures that can be used either in isolation or in combination to manage virtually every anatomic variance encountered.

I recommend this book highly to anyone interested in congenital heart disease. It is the culmination of over thirty years of surgical experience from a dedicated surgical clinician and master technician. To those in nonsurgical specialties this atlas will provide insight into the procedures that drawings simply cannot match. For the surgeon the photographs provide not only outstanding illustrations of the procedures but also the inspiration to perform with the precision and care that would make each operation worthy of a photograph. Excellence in the craft of congenital heart surgery can only be achieved through careful study, practice and dedication. As surgeons we must remember that this skill is a necessary precursor to the pursuit of better long-term outcomes for our patients.

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Preface

Because of the wide variety of anomalies encountered in congenital heart surgery, a broad understanding of the pathologic anatomy of defects is vitally important to the surgeon. More than in many other fields of surgery, a feel for three-dimensional spatial relationships of anomalies is helpful in allowing the operating surgeon to improvise technical details of a procedure. Precisely shaping and sizing an intraventricular baffle or patch, or correctly placing a long intra-atrial suture can make the difference between a successful and an unsuccessful surgical outcome.

The congenital heart surgeon is a student during his or her entire career because he or she encounters so many different anomalies. Learning from the experience of others should always be part of the clinician's education; this is best done by personally observing an operation performed by another. Otherwise, the best record of a procedure is a good operative photograph.

For over 35 years it has been my practice to photograph most operations. These illustrations comprise a valuable part of each patient's record and are informative as a review of previous surgery and observed anatomy if future surgery is planned. The illustrations also serve to inform the referring doctor of details of surgery and this may also improve patient care.

The photographs have been an invaluable teaching aid for lectures, journal publications, and this book. I hope this atlas will be of interest to full-time or part-time congenital heart surgeons, pediatric and adult cardiologists, intensivists, pediatricians, internists, and all other students of congenital heart disease.

In this second edition, I have added many new sections, deleted a few obsolete sections, and in some areas changed the format. For example, the atrial switch operation has been moved to Chapter 16, l-Transposition of the Great Arteries (l-TGA). It is no longer used in the repair of d-transposition of the great arteries (d-TGA), but is applicable for the double-switch operation for l-TGA.

Photographs were taken with a Nikon F camera, usually using a Nikon 55-mm macro lens (Figure P-1). For some close-up pictures of the right and left ventricular outflow tracts, a Nikon 100-mm medical lens was used (Figure P-2). Pictures were taken at f8 to f11 at a distance of 9 to 12 inches from the field.

Lighting for most photographs was augmented with a side-mounted Honeywell Strobonar flash with a wide-angle neutral density filter (Figures P-1, P-2). Using a side-mounted flash, rather than a more traditional ring light (surrounding the lens), has resulted in some shadows in each picture, which improves the perception of depth of field. More recently, I have used a ring light mounted on the front of the lens. No special effort has been made to use the operating room lights or to move them out of the field.

Figure P-1. Nikon camera viewer from above. A 5-mm macro lens is attached, and the light source is positioned to the side of the lens. A waist-level viewer is attached for taking pictures from the head of the operating table.

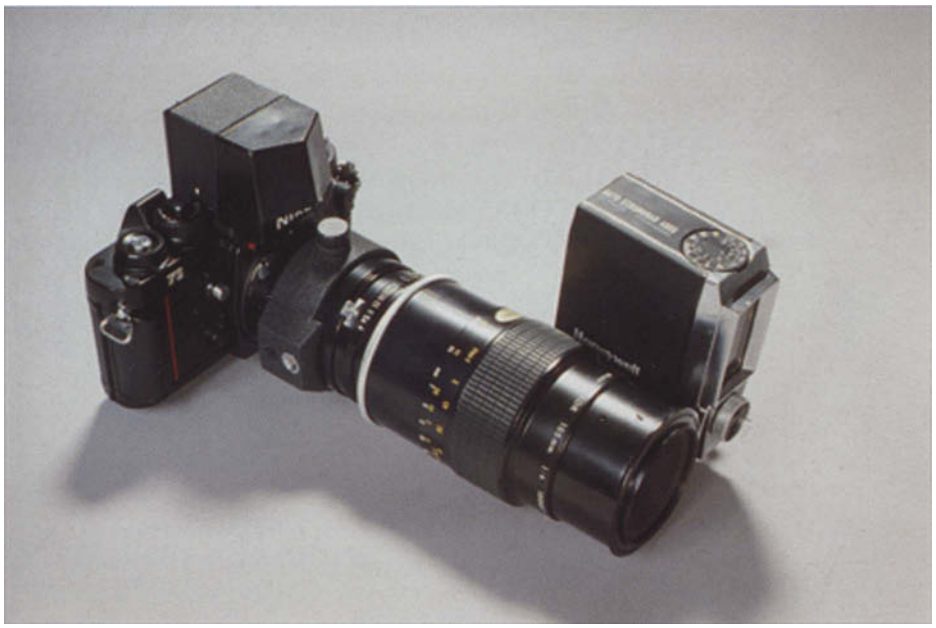

Figure P-2. Nikon camera with 100-mm macro lens attached for close-up views. A sports viewer is used, and the position of the light source is at the side of the lens.

Photographs were taken from behind the patient when surgery was performed through a lateral thoracotomy and from the head of the operating room table for a median sternotomy.

For orientation, pictures in this atlas are marked with *arrows* to indicate *R*, right side of patient; *L*, left side of patient; *Cep*, cephalad; *Caud*, caudad; *Ant*, anterior; and *Post*, posterior.

S. Bert Litwin, MD

Acknowledgments

It is an honor and a privilege to work in the field of congenital heart surgery. The workdays are long and the cases are complex and trying, but the rewards and satisfaction are enormous.

I am indebted to Linda Hamilton, RN, Nancy Stover, RN, Maryanne Kessel, RN, MBA, and Kathleen Mussatto, BSN, who have worked for me during successive years as cardiovascular surgical nurse clinicians. They catalogued photographs as one of many invaluable responsibilities that they tirelessly performed, in addition to carrying out various tasks in the care of many sick infants and children. Linda went on to be a senior administrator at various healthcare facilities in Vancouver, Canada. Nancy, sadly, is deceased. Maryanne is now Director of the Herma Heart Center and Kathy is Manager of Research of the Herma Heart Center, Children's Hospital of Wisconsin. I and all of my patients are forever in their debt.

Many surgeons had an influence on my career and, thus, the clinical experience that allowed this book to become a reality. Dr. Oliver Cope (deceased) taught me research techniques and an investigative approach to surgery, as well as humility and compassion for patients. Dr. W.G. Austen taught me the benefit of organizational skills and introduced me to cardiac surgery. Dr. Robert E. Gross (deceased) showed me the excitement of children's surgery and taught me the principles of congenital heart surgery. Dr. William F. Bernhard showed me the techniques of infant heart surgery, many of which have remained valid and can be applied to patient care today. Mr. R.H.R. Belsey showed me surgical technique par excellence and the value of excellent clinical judgment. Dr. Aldo Castaneda, with whom I had the opportunity to work for a short time, was the guiding light in the field of congenital heart surgery for many years and an example of the surgeon who can achieve the highest degree of success and innovation in this complex field of surgery.

Dr. Willis Williams, my long-time close friend, helped me to develop the photographic techniques that I have used for many years.

Tireless efforts of my cardiologists, anesthesiologists, intensivists, physician assistants, perfusionists, nurse clinicians, and many others are responsible more than I can say in helping to achieve any clinical success I have enjoyed.

I thank my associate, Dr. James Tweddell, for allowing me to photograph his patients undergoing repair of intramural coronary artery and stage I palliation for HLHS (Chapter 18, Sections 2 and 6, respectively).

My secretary Lisa Armitage helped enormously by typing the manuscript and cataloging photographic slides.

I am extremely grateful to the editors and previous and current staff of Springer Science+Business Media for encouragement in the preparation of this atlas and recognition of the value of operative photographs. This includes Laura Gillan, Beth Campbell, and Paula Callaghan. Springer Science+Business Media has done an outstanding job in reproducing so much material in an artistic, scholarly fashion.

S. Bert Litwin, MD

Contents

Introduction and Techniques

Some techniques and principles of operative exposure and cardiopulmonary bypass that I currently prefer are worth mentioning. No doubt different techniques used by other surgeons are just as effective in their hands.

Most open heart operations are performed through a standard median sternotomy with a longitudinal skin incision. For female patients of all ages who undergo simpler operations and in whom I do not anticipate a repeat sternotomy or the need to leave the sternum open, my preference is a transverse skin incision to approach the sternum for the sternotomy. The cosmetic advantage is obvious and I have encountered few complications with this incision. In order to avoid damage to the lateral and anterior cutaneous branches of the intercostal nerves that may result in loss of nipple sensation, the surgeon must restrict the subcutaneous flap dissection to a triangular area with the tip at the manubrium and the triangle base at the site of the transverse skin incision below the nipples.

For intra-atrial procedures, cardiopulmonary bypass can also be instituted by working through a right lateral thoracotomy in the fourth interspace. The cavae are readily exposed and the ascending aorta can be cannulated. Cardiopulmonary bypass through a left thoracotomy is more difficult. Aortic cannulation of the arch or descending thoracic aorta is simple, but a cannula for venous return has to be placed in the main pulmonary artery and, in older patients, through a femoral vein into the right atrium. When partial bypass through the left chest is used, the arterial cannula is placed in the descending thoracic aorta and a single venous cannula in the left atrial appendage. The head and neck are perfused by the beating heart, and the lower body is perfused by the partial cardiopulmonary bypass circuit.

Most closed heart surgery is performed through a lateral thoracotomy in the third or fourth intercostal space without rib resection. This includes isolated Blalock shunt and pulmonary artery banding operations. In this day of multiple mediastinal operations for staged repairs, it is not wise to add to mediastinal scarring with an initial simple palliative operation that can effectively be performed through a thoracotomy.

For cardiopulmonary bypass, a standard roller pump and hollow fiber membrane oxygenator are used. The arterial perfusion cannula I most often use is a plastic straight-tipped wire bound DLP cannula (Medtronic Inc., Minneapolis, MN) for small aortae and a right-angle metal-tipped Sarns Cannula (Terumo Cardiovascular Systems Corporation, Ann Arbor, MI) for older children.

In most cases, bicaval cannulation is performed because this allows the surgeon to work within the cardiac chambers during cooling and rewarming periods, even without aortic cross-clamping. Right-angle, metal-tipped DLP cannulae (Hospira, Inc., Lake Forest, IL) are usually placed directly in the superior vena cava and through the right atrial wall near the inferior vena cava. A single venous

cannula in the right atrial appendage is advantageous in small infants, but does not allow the surgeon to work within the atrium during cooling and rewarming.

A ventricular vent is placed, usually at the interatrial groove along the right heart border or through the left atrial appendage. I prefer large vents, because this is a major component of my de-airing technique that follows repair. A small, needle is placed in the proximal ascending aorta for delivery of cardioplegia and to help in later intracardiac de-airing.

Cardiopulmonary bypass indexed flow rates are at 2.2 to 3.4 L/min at mild hypothermia (32°C), 1.8 to 2.2 L/min at moderate hypothermia (26°C), and 0.5 to 1.5 L/min for temporary low flow (under 24°C). Low-flow cerebral perfusion (0.25–0.5 L/min indexed) has supplanted the need for circulatory arrest. Placement of central OPITCATH MVO_2 catheters (Hospira, Inc.) for online monitoring as a predictor of cardiac output, and the use of near infrared spectrometry (NIRS; Somanetics Corporation, Troy, MI) for continuous measurement of cerebral oxygenation are invaluable. The inclusion of amphiphylic, biopassive polymer coating including X-COATED (Terumo Cardiovascular Systems Corporation, Ashland, MA) and $SMAR_XT$ COATED (COBE Cardiovascular, Inc., Arvada, CO) coating on oxygenators and bypass tubing and the use of post-bypass modified ultrafiltration have been beneficial in reducing the inflammatory response to surgery. Control of systemic vascular resistance with the beta blocker phenoxybenzanine and use of the open sternum, particularly in small neonates after complex repairs, have helped to significantly reduce mortality. The value of EMCO support as a bridge to recovery for life-threatening ventricular failure in improving survival cannot be overstated.

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