BOOK REVIEW: CARDIAC CATHETERIZATION IN CONGENITAL HEART DISEASE - PEDIATRIC AND ADULT BY CHARLES E. MULLINS

By John W. Moore, MD

Cardiac Catheterization in Congenital Heart Disease - Pediatric and Adult is an exceptional book!

It is much more than I hoped for or expected. It is without doubt the most authoritative, the most comprehensive and the most detailed textbook about cardiac catheterization in congenital heart disease ever published.

There are 944 pages of text and 313 figures, divided among 35 chapters. Each chapter is well-referenced. Unlike most other familiar texts of this magnitude, Dr. Mullins wrote every chapter and every word. This is all his work. Most of the figures are instructive “free hand” line drawings contributed by one of Dr. Mullin’s patients.

The chapters cover the entire range of topics which are of interest to cardiologists and other professionals involved with the catheterization of infants, children, and older patients having congenital heart disease. The initial top-
cardeterization is fully discussed in chapters on right and left (transeptal and retrograde) heart catheterization, hemodynamics, and angiographic techniques. Finally, the bulk of the text and the majority of the chapters deal with interventional procedures and technologies, and follow in the order of their historical development. These chapters cover septostomy, valvuloplasty, angioplasty, stenting, occlusion procedures and devices, and cutting-edge technologies.

“It is obvious that with this book, Dr. Mullins gives each of us a huge gift. In the Preface he tells us that ‘the information contained in this text...represents the accumulation of knowledge, techniques, and procedures learned, utilized and/or developed by the author during the continued learning, practice and teaching, of cardiac catheterization procedures during ....four decades in the field.’”

Clearly, there are too many chapters to mention all of their topics. Each chapter is a comprehensive, detailed treatment of its subject matter. For example, in Chapter 24, entitled “Intravascular stents in venous stenosis,” Dr. Mullins examines this topic from every perspective. He provides a full discussion of appropriate equipment and stents, which complements a very detailed step-by-step description of how to do the procedures in a safe and reliable manner. His goal is to emphasize both the fundamentals and the details. There are also sections covering totally obstructed venous channels, peripheral vein obstruction, and post-implant care. Stent implants in Fontan circuits are considered separately, as are pulmonary vein stents and pulmonary venous baffle stenting. The chapter ends with a discussion of complications unique to venous stents. Any cardiologist seeking education or information about venous stenting, need only study this one source.

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I was especially fortunate to have my fellowship training under Dr. Mullins. I am one of many who know him as a gifted teacher and mentor. Cardiac Catheterization in Congenital Heart Disease - Pediatric and Adult codifies Dr. Mullins’ legacy in a definitive way. It will enable numerous present day and future cardiologists and their patients to benefit from his gifted teaching.

The book is dedicated to his wonderful wife Arlene; his mentor at Walter Reed Army Medical Center, Dr. Weldon Walker; and his friend, associate and chief for most of his tenure at Texas Children’s Hospital, Dr. Dan McNamara. It was published in December 2005 by Blackwell Publishing, Ltd., Oxford, UK. The book is available for purchase from the Blackwell Publishing online bookstore at www.blackwellcardiology.com for US$225.

~CCT~

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Between February 8th and 12th, 2006, The Cardiac Center at The Children's Hospital of Philadelphia will host Cardiology 2006: 9th Annual Post Graduate Course in Pediatric Cardiovascular Disease—State of the Art Management of the Neonate and Infant with Cardiac Disease. After eight successful meetings in Orlando, this year’s course will be held in Scottsdale, Arizona, at the Hyatt Regency Gainey Ranch and Spa. This course does not attempt to recreate the very successful subspecialty meetings for practitioners who care for children with heart disease; rather, a multidisciplinary approach combining neonatology, pediatric cardiology, cardiac surgery, nursing, anesthesia, critical care and perfusion will be taken. Over 90 faculty members from around the globe representing multiple disciplines and academic centers will present over 200 plenary and subspecialty lectures, and will feature three broad themes:

- common congenital heart disease: current management and controversies
- clinical trials and new research in pediatric cardiovascular disease, and
- ethical issues in the delivery of care to infants with cardiovascular disease.

Daily plenary sessions are designed to be of interest to all medical, nursing and allied health professionals, with supplementary specialty-specific breakout sessions as well. Individual ‘tracks’ have been designed for neonatology and intensive care, echocardiography, intraoperative care and perfusion, cardiac nursing, and administration.

The five comprehensive sessions on congenital heart disease will start with a review of anatomic specimens by Professors Robert Anderson and Paul Weinberg (including the controversies of nomenclature), preoperative imaging, surgical procedures, postoperative care and long term results. Lesions that will be reviewed include transposition of the great arteries, ventricular septal defect, tetralogy of Fallot, aortic valve disease and hypoplastic left heart syndrome. Current surgical approaches will be described by Drs. Vaughn Starnes (Los Angeles), Thomas Spray (Philadelphia), Mike Teodori (Phoenix), and John Mayer (Boston).

Three separate plenary sessions will be devoted to New Research and Clinical Trials. Dr. Skip Nelson will review regulatory issues in developing new drugs for children, Dr. Dave Wessel will review considerations in industry sponsored trials, and Dr. Gail Pearson will review the current status of the Pediatric Heart Network. New data on ICU pharmaceuticals (levosimendan, nesiritide, milrinone, esmolol), neurological outcomes after cardiac surgery, ABO incompatible heart transplantation, blood product utilization, and single ventricle surgery will be pre-
sented. Dr. Redmond Burke will provide an overview of information systems, research and clinical care, and Dr. Rick Ittenbach will review important statistical considerations for all practitioners.

On Friday, February 10th, the afternoon session will be devoted to controversial topics and ethical dilemmas in our practice. Two pro-con debates will take place: “Should Non-surgical Care be Offered to Parents of Neonates with HLHS”, and “Should Surgical Innovation be Monitored by Institutional Review Boards” (where Dr. Martin Elliot, a consultant surgeon from Great Ormond Street, will argue that new surgical procedures should be externally reviewed). End of life care, surgery in patients with lethal chromosomal defects and issues of informed consent will be reviewed. Finally, global perspectives on CHD care will be delivered by Dr. Daniel Penny (Australia) and Dr. Hiromi Kurosawa (Japan).

A special pre-conference seminar will be held (limited attendance) reviewing anatomic specimens and 3D echocardiography with Drs. Bob Anderson, Paul Weinberg and Girish Shirali. Dr. Shirali will also host a fabulous, two hour, hands-on session on 3D echo imaging on Friday, February 10th.

Over 40 abstracts will be presented in the young investigator award competition; the award will be given on Saturday, February 11th, in addition to the featured Rashkind lecture by Dr. Andrew Redington, the featured Lillehei lecture by Dr. Martin Elliot, and the featured Nursing Lecture by Ms. Kathy Mussatto.

Hope to see you in Arizona! Details and registration may be found at www.chop.edu/cardiology2006

~CCT~

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MEDICAL CONFERENCES

Clinico-Morphological Correlations Course
January 16-17, 2006; London, UK
www.ichevents.com

The 17th Scientific Session of the Saudi Heart Association
January 31- February 2, 2006; Khobar, Saudi Arabia
www.saudiheart17.com

25th Annual Scientific Meeting of the Belgian Society of Cardiology
February 2-4, 2006; Brussels, Belgium
www.bocardio.be

Cardiology 2006: Focus on the Neonate and Infant with Heart Disease
February 8-12, 2006; Scottsdale, AZ USA
www.chop.edu/cardiology2006

The 6th Annual International Symposium on Congenital Heart Disease with Echocardiographic, Anatomic, Surgical and Pathologic Correlation
February 17-22, 2006; St. Petersburg, FL USA
www.CHIF.us

16th Utah Pediatric Cardiovascular Disease Conference
February 18-21, 2006; Snowbird, UT USA
www.congenitalcardiology.com/Utah-PCD.pdf

ACC 55th Annual Scientific Session (American College of Cardiology)
March 11-14, 2006; Atlanta, GA USA
www.acc.org

Innovation in Intervention: i2 Summit 2006 (runs concurrently with ACC '06)
March 11-14, 2006; Atlanta, GA USA
www.acc.org

Doppler - Ultrasound in the Fetal Examination (14th International Symposium and Workshop)
March 14-19, 2006; Oberlech - Alberg, Austria
www.kinderherzzentrum.at/Lech2006

Visit www.CongenitalCardiologyToday.com to see a complete list of medical conferences, symposia and workshops for 2006

Cardiology 2006
February 8th-12th, 2006
Hyatt Regency Scottsdale at Gainey Ranch, Scottsdale, AZ, USA
Sponsored by The Cardiac Center at The Children’s Hospital of Philadelphia
www.chop.edu/cardiology2006/ or call +1.215.590.5263

Dr. Rossant has provided significant insights into how an embryo develops, how genes control development and how embryonic and other stem cells arise. Her research interests centre on understanding the genetic control of normal and abnormal development in the early mouse embryo, work that has shed light on how congenital anomalies in the heart, blood vessels and placenta arise. Her work on the genes that control blood vessel development has been of major importance in defining novel pathways for new drug interventions in cancer. Throughout her career, Dr. Rossant has been a pioneer and innovator of new techniques to manipulate the mouse genome, enabling the mouse to become the pre-eminent model for understanding the function of the human genome sequence.

As well as being the Chief of Research at SickKids and a senior scientist in Developmental Biology in the SickKids Research Institute, Dr. Rossant holds the Lombard Insurance Chair in Paediatric Research at The Hospital for Sick Children and is a University Professor in the Departments of Medical Genetics and Microbiology and Obstetrics and Gynecology at the University of Toronto. She is also the Deputy Director of the Canadian Stem Cell Network and the Director of the Centre for Modelling Human Disease in Toronto, which is developing new mouse models of human disease. She is actively involved in the international developmental biology community, serving as Editor of Development for many years and as President of the Society for Developmental Biology in 1996-97. Dr. Rossant also served as Chair of CIHR’s Working Group on stem cell research.

Dr. Rossant, trained at the Universities of Cambridge and Oxford in the United Kingdom, came to Canada in 1977. She is a Fellow of both the Royal Societies of London and Canada and a Distinguished Investigator of CIHR. She is also a two-time Howard Hughes Medical Institute International Scholar.

The Hospital for Sick Children, affiliated with the University of Toronto, is Canada’s most research-intensive hospital and the largest centre dedicated to improving children’s health in the country. Its mission is to provide the best in family-centred, compassionate care, to lead in scientific and clinical advancement, and to prepare the next generation of leaders in child health. For more information, please www.sickkids.ca.
Welsh Heart Research Cracking Sudden Death

Scientists have discovered for the first time that a genetic mutation linked to some cases of SCD makes a specialised heart muscle protein 'jittery'. This protein is important in controlling the rhythm of heartbeat. In sudden cardiac death the heartbeat can speed up out of control or become dangerously irregular.

Contraction and relaxation of the heart involves a system of coordinated transport of calcium in and out of the muscle cells. Mutations in the gene for the calcium release channel have recently been shown to destroy their ability to work properly following stress or exercise, possibly explaining why heart signalling is severely disrupted and leads to fatal heart rhythms in affected people. However, it was not known what was wrong with the mutated channels.

Now, a British Heart Foundation-funded research team in Cardiff have solved this mystery and pinpointed the changes in mutant channel proteins. They have shown that mutations make the activated channels ‘jittery’ and prevent them from closing properly, leading to the release of too much calcium.

Dr Christopher George, who led the research explained, “This is crucial new evidence that defects in the channel structure may cause these dangerous abnormalities in cardiac calcium release.

"Although there is a long way to go, this finding gives us vital clues that the precise stabilisation of these channels may be the best way to prevent this catastrophic disease in people carrying the faulty gene."

Work is already underway to develop a new therapeutic approach which is hoped will eventually restore proper channel closure and may help prevent SCD in susceptible individuals with the mutant gene.

Professor Jeremy Pearson, Associate Medical Director of the British Heart Foundation said, “Sudden cardiac death is particularly tragic as it can come out of the blue in seemingly healthy young people. It often even affects those taking the best care of their hearts by being fit and active.

“People can be prone to SCD if they have a number of underlying heart conditions. However, this research provides important evidence explaining how abnormalities in the calcium release system contribute to this devastating condition in people without another heart condition.

"We hope it will lead to further developments towards a way of preventing the problem in susceptible people, without stopping them from stressing their hearts through exercise."

For more information: www.bhf.org.uk
Camp Meridian: Seek, Explore, Discover

By Naomi S. Gauthier, MD

There are a few experiences in every physician’s career that stand out as especially poignant, and become deeply etched in our memories in surprising detail. I remember one such instance, from when I was a cardiology fellow. I sat in a small room facing two young, frightened, and completely overwhelmed parents, and listened as my attending broke the news that their baby had hypoplastic left heart syndrome. The options were outlined as: high risk surgery, await heart transplant, or choose no treatment and let the baby succumb. I remember how powerless I felt, frustrated by our medical shortcomings. I also recall wincing at how much bias crept into the conversation, the experiences of the cardiologist and neonatologist colored their language, the parent’s collective background affected their ability to hear clearly, and I had to admit that my own thoughts were slanted. A recent article from Canada reflected on this same slant or bias that affects the care plan chosen.[1] Pediatric cardiology does not have the luxury of large clinical trials, or carefully constructed evidence based medicine. Clinical experience and best judgment is a large part of how we all practice; bias, even if it is for the best, is still an inherent part of this. The positive aspect is that the field remains nimble, creative. It can, and has, advanced at a stunning rate. Flash forward over a decade later, and surgical palliation has become commonplace. My early experience with that neonate was sharply contrasted by my recent camp experience with a survivor, a nine year old boy who underwent a Norwood repair. He attended our first ever session of Camp Meridian, a free, non-profit camp for kids with heart defects. This boy, and the other children who attended, taught me to take a long hard look at my own biases, and has given me a fresh new outlook on the practice of pediatric cardiology not just in terms of survival, but in terms of living with their defect.

I founded Camp Meridian four years ago out of the old haunting frustration that modern medicine was not doing enough for these children. We have gotten quite good at diagnosing and treating heart defects, but still lag behind in knowing how to encourage the children to live their lives with a heart defect and not be defined by it. Camp Meridian was intended to be a highly structured, cooperative adventure to challenge these children to see themselves in a new way, and turn any thoughts of “I Can’t” into ones of “I Can.” What I did not realize was how much that philosophy applied to the staff as well.

To run Camp Meridian, we rent a beautiful, professional host camp facility on 600 acres in the mountains of New Hampshire. We devise a themed mystery program that transforms camp into a different exotic locale every year. This year we had a tropical theme, entitled “Searching the Sands of Bora Bora.” We spent the first afternoon doing get-to-know-you activities, and that evening we gathered around the fire to tell the fictional story of the Tikki Man who gave the power to the Wishing
vational gifts to point the campers toward their next activity.

Missions have varied through the years, including things like specialized obstacle courses, trying to get a group across a swinging platform without touching the ground, building a working raft while half the team is blindfolded, and all time favorite, scaling the climbing wall. Creative teamwork is encouraged, and the campers are allowed to figure out solutions without adult interference. As each mission was completed, the children gained a tremendous sense of accomplishment and bonded with their peers in overcoming trials and celebrating shared successes. With excitement mounting to a fever pitch, the weekend culminated with the children digging up buried treasure to find the Tikki Man’s lost pearls. One by one, the campers solemnly placed the individual pearls onto his necklace, together rebuilding and restoring his powers. They then received a necklace of their own to keep as a reminder of all that they achieved. To end on a high note, we had a surprise luau celebration, complete with music and food. As the parents arrived, we treated them to a slide show of the weekend’s events so they could see with their own eyes their children doing things they may have not thought possible.

Most of these children had never been away from home or slept anywhere other than their own beds. Just attending camp was in itself an accomplishment. We have only two and a half days to develop relationships and give the campers a whole new way of looking at themselves, and I remain amazed at how quickly this occurs. We take time throughout the weekend to reflect on each activity, and each mission group discusses their experience with the group at large what elements they used to succeed in their mission, such as teamwork, respect, and encouragement. The campers are surprisingly frank and open, and you can watch their confidence grow as the weekend progresses. This is helped by our special young adult counselors, who themselves have grown up with heart disease. We have no specified time to discuss heart defects, but invariably the children will decide to discuss their own medical experiences. When the counselors open up about their own heart disease, the looks on the faces of the campers are priceless. They quickly idolize their counselors, and when they find out they too have heart disease, it forces the children to reconcile their images of what it means to have congenital heart disease with what they see in their counselors.

Knowing what these children look like on paper and knowing their medical histories has turned out to be a two dimensional reality when faced with the full scope of what these children can do. This was most apparent to me when I was at the climbing wall. A nine year old boy with hypoplastic left heart syndrome, small for his age, almost dwarfed by the safety equipment, took a deep breath, looked up at the wall, and proceeded to climb all the way to the
top. His grin at the top, an undeniable accomplishment from his vantage point, made me flash back to the day I listened as the couple chose to take their infant son home to die. Had the boy at the top of the climbing wall been born a few years earlier, his fate may have been different. Had his parents not had the courage to drop him off at camp, he never would have had the opportunity to grin triumphantly from the top of a climbing wall, showing the world that a heart defect is in the eye of the beholder. I then watched a girl, nine years old with single ventricle physiology from double outlet right ventricle and mitral stenosis, approach the wall. She had a fenestrated Fontan and had suffered a stroke at age two years, leaving her with limited use of her left leg and paralysis of her left arm. She made it only a short distance, and slipped a few feet from the ground, caught by the safety ropes. I caught my breath, thinking about how she was on coumadin, finding myself worrying for no specific reason. I exhaled as I realized how close her feet were to the ground, how safely she and others were guided by the safety ropes, and how little real chance there was for any sort of injury. And this year, the red-headed girl with the Fontan and the left sided hemiparesis, slowly, carefully, and with assistance, made it safely all the way to the top of the wall. It made me wonder about how many times I had guided parents and kids away from activities because they somehow sounded risky, when I probably was acting more from nonspecific parental worry or bias than by any real understanding of kinesiology and pediatric cardiology.

In 21st century pediatric cardiology, we have become very successful at fixing or palliating heart defects. But what about living their lives once the surgery or intervention is complete? We fix their hearts, but what of their self esteem, their ability to take calculated risks in order to grow, their sense of safety margin that is not overly narrow? We have all seen the effects of the obesity epidemic, and the loss of physical activity in childhood. How do we fairly encourage our patients to have a healthy degree of exercise and a life long commitment to their cardiac health, without encouraging risk? Our adult colleagues have reaped the benefits of cardiac rehabilitation programs in their post-myocardial infarction patients, in what is certainly a higher risk population than most of our pediatric cardiology patients. Although we have some attention made to similar programs in youth, exercise prescription and formal exercise programs are sorely underutilized and we have much to learn. [2-10]

At my town’s recreational youth basketball game, I watched my eight year old son and his very competitive, very
talented teammate fly down the court and sink a perfect lay up. Both boys have older brothers, and were used to playing with more skilled players. Just a few days earlier, I had read the same boy’s MRI report confirming increasing left ventricular dimension from his severe aortic insufficiency, and knew that surgery was being recommended. My colleague had not restricted his activities, as he was only eight and this was “rec ball,” and common wisdom held that it was “safe.” I would have said the same. Sitting in the stands, I had to admit my own inconsistency, as I could see that the intensity of the play was no different from that of my older boys’ competitive travel basketball team. This conventional wisdom does have a basis; there is data on workload of different sports,[11,12] and there are the well-known Bethesda Guidelines.[12] On my review of the literature I could not find any case reports of children experiencing sudden death with exertion before age eight. However, the particulars of competition versus playground play, the skill level of the players, and the individual effort of the patient are harder to pin down, and I think our own biases come into play when we make, rightly or wrongly, the decisions we are asked to make. It remains a challenge to know how to encourage the patients; yet keep within a safe margin; how to allow for personal growth, yet not set unrealistic expectations; and how to avoid risks of obesity and inactivity, yet balance exertional risk. We have a lot to learn from the patients. For two and a half days each fall, when the foliage has reached its peak brilliance in the heart of New England, I am inspired by a group of children whose potential is only just beginning to be tapped. I try to re-learn the very goal we had for these children when we created Camp Meridian: to see that these children are much more than a diagnosis on a page, and that they are, first and foremost, developing children who happen to have heart defects as one aspect of themselves. One of the many reasons Camp Meridian has been so successful is because of the children themselves. They are remarkably accepting and inclusive; perhaps their own experiences have given them a sensitivity not always seen or encouraged at their age. Watching them blossom has been as deeply meaningful to them as it has been to the staff who supervise the weekend. Our motto is Seek, Explore, Discover: seek to be your best and help others, explore yourself and the world around you, and discover the pride of accomplishment and the joy of lasting friendships. The lessons learned transcend congenital heart disease, and are lessons for all.

References:
4. Kovacs AH, Sears SF, Saidi AS.

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If you have information to share with our readers on other camps for kids worldwide with CHD, or would like to submit an article about your camp, send an email to: CAMPS@CCT.bz