

HEART MURMURS



WINTER 2008/09 A PUBLICATION FOR PHYSICIANS, NURSES, INTERDISCIPLINARY COLLEAGUES, AND FRIENDS OF THE CARDIOVASCULAR PROGRAM AT CHILDREN'S



Vol. 11, No. 2

Children's Hospital Boston

TRAINING THE NEXT GENERATION

Training the next generation of clinicians is an important goal for the Cardiovascular Program at Children's Hospital Boston. We aim to attract and teach interdisciplinary trainees not only to become first-class clinicians, but also to help them become future leaders and innovators in the field. There are considerable pressures on training programs, and adaptations have been necessary to provide comprehensive training to achieve both our goals as a cardiovascular program and the requirements of post-graduate education. This edition of *Heart Murmurs* is devoted to training the next generation, with particular emphasis on the challenges and changes necessary to maintain a world-class training and education program for our interdisciplinary team.

Peter C. Laussen, MBBS

Patricia Hickey, MS, MBA, RN, NEA-BC

THE TRAINING OF CONGENITAL HEART SURGEONS

EMILE A. BACHA, MD, SENIOR ASSOCIATE IN CARDIAC SURGERY AND DIRECTOR, CONGENITAL HEART SURGERY FELLOWSHIP

Congenital heart surgery is a specialty characterized by low error tolerance, a high level of cognitive and technical performance, and coordinated efforts of multiple individuals operating within a sophisticated organizational structure. Training in adult and pediatric cardiac surgery is undergoing major changes. Nearly one-third of American Board of Thoracic Surgery–certified training positions have gone unfilled over the last year. Reasons are a shrinking patient pool (mostly due to percutaneous coronary interventions), the perception of poorer quality of life of cardiothoracic surgeons (as compared to other specialties), declining reimbursements and salaries, a shrinking job market, increased legal challenges, and increasing scrutiny by professional societies, payors, and government. Congenital heart surgery has been somewhat protected from these changes, although our “recruits” have traditionally come from the general ranks of cardiothoracic surgery trainees. In a recent survey, nearly one-quarter of cardiothoracic surgery training program graduates reported that they would not choose a career in cardiothoracic surgery again, and more than half would not strongly recommend cardiothoracic surgery to potential trainees.

THE (LONG) ROAD TO BECOMING A CONGENITAL HEART SURGEON

Until recently, one generally went through five years of general surgery training, two years minimum of cardiothoracic surgery residency, and one year of congenital heart surgery fellowship prior to becoming a practicing congenital heart surgeon (research years not included). One had to pass the board in Thoracic Surgery, and there was no “board” in Congenital Heart Surgery. In 2006, the American Board of Thoracic Surgery created a Congenital Cardiac Surgery Subspecialty Certificate. The following criteria must be met prior to becoming a candidate for this certificate:

- (1) The fellowship shall be for a minimum of 12 consecutive months and must follow satisfactory completion of thoracic surgery training in an American Council for Graduate Medical Education (ACGME)–approved program.
- (2) Operative experience must include a minimum of 75 major congenital cases and must encompass the following index cases: VSD, AVSD, TOF, Coarctation, Arterial Switch, Glenn, and Fontan.

Henceforth, all congenital heart surgeons will have to obtain this certificate, in addition to the board certification in Thoracic Surgery. On the other hand, reflecting on the very long time spent in training, the General Surgery board certification was made optional.

IN THIS ISSUE

- *A Tribute to Dr. Michael Freed* 3
- *Innovative Practices for Training CICU Nurses* 4
- *New Cardiac Neurodevelopmental Program* 7

CONTINUED ON PAGE 2

TRAINING IN CONGENITAL HEART SURGERY

Congenital heart surgery is a much-scrutinized specialty. Competence is difficult to measure. The commonly used metric of postoperative survival is at best an uncertain measure of individual surgeons' competence. A "mistake" during a heart surgery procedure is easily seen (and counted) by all, and can have devastating if not lethal consequences to the patient. Add to the required technical precision and skill the time pressure on cardiopulmonary bypass, and the needed situational awareness and cognitive demands, and you can easily see the need for highly trained and highly functioning individuals who must perform at this extreme end of performance on a daily basis. In addition, at Children's Hospital Boston (CHB) all pediatric cardiac surgery outcomes (including the major complications) are currently reported to the Society of Thoracic Surgery (STS) national database, as well as to the Commonwealth of Massachusetts.

The number and depth of skills required to be successful has also increased recently due to the shift toward earlier complete cardiac repairs; the emergence of subgroups of patients posing special demands, and challenges such as premature and low-birth-weight babies or adult congenital heart disease patients; the emergence of hybrid and endovascular techniques; and the increased sophistication of pediatric ventricular assist devices. In addition, trainees need to have in-depth knowledge of pediatric cardiac intensive care, physiology, immunology, and transplantation. Finally, they have to be able to work in a team with multiple specialists, and possess an understanding of reflective practice and patient safety principles. Thus, training to competency in congenital heart surgery is no easy task. The introduction of the special Certificate recognizes the need to shift from a practice-based curriculum (required number of years in training) to a performance-based one (required structured curriculum and mandatory standards and skills).

TRAINING IN CONGENITAL HEART SURGERY AT CHILDREN'S HOSPITAL BOSTON

Children's Hospital Boston's Cardiac Surgery Department employs an average of five to six congenital heart surgery trainees at any one time. About one-half to two-thirds are in the process of training in cardiothoracic surgery, and rotate through Children's via another program, such as Brigham and Women's Hospital or Massachusetts General Hospital. The others have finished their cardiothoracic surgery training in the United States or abroad, and are spending a year or more training in congenital heart surgery. Besides an intense operative schedule and many

weekly didactic sessions (which include hands-on pathology sessions in the Cardiac Registry), we have recently integrated simulation as a teaching technique into the structured curriculum. Cardiac residents and fellows are now an integral part of the monthly Crisis Resource Management (CRM) simulation session that integrates all cardiac subdisciplines at CHB in crisis scenarios.

In addition, with the help of the CHB Simulation Center and the Cardiac ICU, we have developed a special trainer for teaching of ECMO cannulation skills. High-fidelity simulation provides the opportunity to teach cannulation skills "on demand" within a safe and standardized environment without risk of harm to patients. The simulation suite is a dual-purpose simulation laboratory and procedure room with an adjacent control room and conference room. Closed-circuit links allow for immediate video-based debriefing as well as teleconferencing. The Embedded High Fidelity ECMO Skills Trainer is a simulated trainer with the look, feel, and anatomy of the actual tissue, and is embedded in the neck of a mannequin within the high-fidelity environment at the point of clinical care. Cannulation is performed with authentic "blood" flow rates in "vessels," as well as risks of "oozing" and bleeding from the operative site. Attending cardiac surgeons provide feedback and teaching.

THE FUTURE IN CONGENITAL HEART SURGERY TRAINING

In summary, training in congenital heart surgery remains a long and laborious endeavor. At CHB, we have strived to modernize and optimize the training to continue to attract and train the best surgeons in the world. The future of congenital heart surgery training will likely consist of increased mandatory performance metrics (i.e., a certain number of cases and minimally acceptable outcomes required during the first two years in practice prior to becoming a candidate for the Certificate), an increase in the use of simulation (development of cardiopulmonary bypass simulators, development of animal heart preparation for "in vivo" practice), and the use of virtual surgery based on 3-D MRI models to teach complex procedures. ♥

Reference

Bove E L. The American Board of Thoracic Surgery's perspective on the future of congenital heart surgery education. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu.* 2008;86-7



EMILE A. BACHA, MD

A TRIBUTE TO DR. MICHAEL FREED

BRIDGET STEWART, ADMINISTRATIVE DIRECTOR, CARDIOLOGY

On Saturday, June 14, 2008, family members, close friends, and colleagues joined Dr. Michael Freed in celebrating 35 years of dedicated service in the care and treatment of patients and families with congenital heart disease at Children's Hospital Boston. The event was held at the John F. Kennedy Library at Columbia Point in Boston.

More than 50 people responded with personal notes and words of wisdom and nearly 300 attended to pay tribute to Dr. Freed's notable career. Dr. Phyllis Pollack remarked, "Mike is the consummate clinical educator. His talk on differential diagnosis of agnostic congenital heart disease has been disseminated (exponentially) by decades of fellows to residents and medical students. His dedication, humor, and integrity remain an inspiration to us all."

Several distinguished guests spoke, including Drs. James Lock, Jane Newburger, Peter Lang, and Josh Robinson. Dr. Aldo Castañeda traveled from Guatemala to attend the celebration. He spoke about Dr. Freed's unique qualities as a pediatric cardiologist, but also about his broad vision in extracurricular interests and in literature.

Other speakers included Cardiology Nursing Director Cheryl O'Connell; his youngest son, Stephen Freed; and Arnie Kanter, a personal friend who attended Brandeis with Dr. Freed. Dr. Kanter recapped about their college days and reminisced about family trips with their children. He also talked about Dr. Freed's inherent flaws, such as a "keen intellect" and his "refusal to judge others."

Patricia Hickey, Vice President of Cardiovascular and Critical Care Services, presented Dr. Freed with a framed copy of an entry on his behalf into the United States *Congressional Record*, Senate #2288, on April 1, 2008, by Senator Edward M. Kennedy. Dr. Newburger provided the summary that Senator Kennedy read to the Senate. ♥

The excerpt from the *Congressional Record* reads as follows:

Mr. President, I welcome this opportunity to pay tribute, on the occasion of his retirement, to Dr. Michael David Freed of Children's Hospital Boston for his service to the hospital, and the thousands of children and young adults from Massachusetts and beyond who have benefited from his care.

Dr. Freed has had a long and distinguished career at Children's Hospital and Harvard Medical School, beginning in 1970 when he arrived to complete his fellowship training. At Children's Hospital, he rose to become Senior Associate in Cardiology in 1976, and Chief of the Division of Inpatient Cardiology in 1996. Dr. Freed is a physician's physician. His commitment to providing the best possible care for children with heart disease is unwavering. He has used his breadth and depth of knowledge, his clarity of thought, his empathy, and his sense of humor to train more than 200 pediatric cardiology fellows, and innumerable pediatric residents in the fundamentals of congenital heart disease. As a member of Sub-Board of the Pediatric Cardiology, he ensured the highest quality of care by setting standards for board certification for young pediatric cardiologists. At Children's Hospital, Dr. Freed has chaired or served on more than two dozen committees, projects, and task forces ranging from quality improvement and patient care, to graduate medical education and governance.

*His contributions extend well beyond Boston. He has served on the executive committees of all three major national organizations in his field — the American Heart Association, the American Academy of Pediatrics, and the American College of Cardiology, where he currently serves on the Board of Trustees. He is also a member of editorial boards in the field of cardiology, and regularly has been included on the lists of top physicians ranging from the book *The Best Doctors in America* to *Good Housekeeping* and *Boston Magazine*. He is consulted by other pediatric cardiologists from around the world who seek his opinion on the care of his patients. Dr. Freed has also written extensively in the field of pediatric cardiology and cardiac surgery, and is particularly recognized for his work in the newborn physiology of congenital heart disease, infective endocarditis, and valvular heart disease. He has authored more than 60 original articles, contributed more than 40 review chapters and editorials, and developed more than 25 clinical communications and instructive CD ROMS. His leadership in establishing clinical practice guidelines for early postoperative management of children in Boston undergoing open heart surgery was a model for the development of such guidelines nationally. In addition, he has been a member of national working groups to develop guidelines on optimal care of individuals with heart disease.*

I commend Dr. Freed for his outstanding career, and his achievement in improving the quality of care for children and young people with congenital heart disease in Boston and throughout the world, and I wish him well in retirement.

Signed, Senator Edward M. Kennedy



MICHAEL FREED, MD, JUNE 14, 2008

INNOVATIVE PRACTICES FOR TRAINING CICU NURSES

LISA McCABE, RN-BC, BSN, CCRN, AND ANNETTE IMPRESCIA, RN, CCRN,
EDUCATORS, CARDIAC INTENSIVE CARE UNIT

Increasing patient acuity, advances in technology, and the dynamic nature of our health-care environment present multiple challenges to nurses at all levels of practice in the Cardiovascular Intensive Care Unit (CICU). To prepare new staff for effectively meeting these challenges, the CICU orientation program has continued to evolve in creative and innovative ways to provide the knowledge, experience, and support these nurses need.

GAINING KNOWLEDGE AND SKILLS FOR SAFE AND COMPETENT CARE

The primary goal of the CICU orientation program is to support

HEART MURMURS

Editors

Patricia A. Hickey, MS, MBA, RN, NEA-BC
Peter C. Laussen, MBBS

Published twice a year
by the Cardiovascular Program
at Children's Hospital Boston
300 Longwood Ave.
Boston, MA 02115

Masthead drawing by Carolyn Lock

WE'D LIKE TO HEAR FROM YOU

If you wish to refer a patient, receive a consultation, or seek information about our program, please contact us by phone or e-mail.

Patricia A. Hickey, MS, MBA, RN, NEA-BC
617-355-5770
patricia.hickey@childrens.harvard.edu

Pedro del Nido, MD, 617-355-8290
pedro.delnido@childrens.harvard.edu

Peter C. Laussen, MBBS
617-355-5440
peter.laussen@childrens.harvard.edu

James E. Lock, MD, 617-355-7313
james.lock@childrens.harvard.edu

Donna Richardson
Children's Hospital Trust, 617-355-6890
donna.richardson@chtrust.org

Visit our web site
www.childrenshospital.org

 Printed on recycled paper

entry-level staff nurses with the knowledge and skills necessary to administer safe and competent care to our patients and their families. On average, experienced nurses receive 8 to 12 weeks of orientation, while 20 weeks are allotted to new graduate nurses. The trajectory of this training is outlined in the orientation syllabus. Expectations are further defined by standardized goals that allow both preceptors and nurses in training to keep the progress of orientation on target.

Our orientation program consists of 14 classroom days. Instruction is provided by cardiovascular staff nurses, a pharmacist, a child-life therapist, a respiratory therapist, and the medical director of the Cardiac Registry. Scheduling classes one or two days per week over the course of orientation allows new nurses to incorporate learned concepts into practice. Each class day includes time for formal nursing rounds or informal sessions during which nurses in training share their clinical experiences with their group to expand on classroom and clinical learning. Course content is further reinforced by the use of a variety of methods throughout the program, including Cardiac Jeopardy, Policy Jeopardy, Cardiac Anatomy Match Game, and "Make Me a Drip."

Similarly, our CICU New Graduate Internship Program was developed in 2007 to prepare newly licensed staff nurses with the knowledge and skills necessary to administer safe and competent care to our complex patients and their families. During the first phase of the program, new graduate nurses receive 20 weeks of classroom and clinical orientation similar to that outlined above. The second phase of the internship program consists of monthly "Building Nursing Practice" meetings designed to meet the learning

needs these nurses experience as they grow in their practice. Each session includes didactic content, simulation scenarios, nursing rounds, and professional/clinical discussions.

SIMULATOR TRAINING COMPLEMENTS CLASSROOM STUDY

The CICU Orientation Simulation Program was designed to facilitate the development of critical-thinking skills and confident decision making by reinforcing classroom learning in a non-threatening environment, where "mistakes" are an avenue for learning and questions are welcome. Simulator training is provided in the afternoon of most class days to complement the class curriculum. Through simulation scenarios, new CICU nurses can experience the low-volume high-risk situations that may not present during orientation, allowing the nurses to develop the critical-thinking skills necessary to identify potential deterioration of a patient's condition, respond appropriately to emergency situations, communicate effectively, and perform as a team member during resuscitation efforts.

CLINICAL EXPERIENCES LEAD TO INDEPENDENCE IN PRACTICE

Clinical experiences are facilitated by an orientation team of two to three preceptors and guided by a set of structured competencies. Feedback is provided regularly to the nurse in training at the end of the shift and in monthly meetings with the orientation team and a unit educator. Assessment of goal achievement and progress toward independent practice is documented weekly in an online tool, the Independence Rating Scale, developed by Greg Durkin, RN, Staff Development Specialist. This tool defines the key competencies for the entry-level staff

nurse based on the nursing department job document within the conceptual framework of the Synergy Model and our standards of care. An independence score of 70–80% in each of the Synergy domains is required for successful completion of orientation.

A key element of clinical orientation is library time, an hour set aside during each shift to allow the nurse in training to review the patient's physiology, care needs, and planned interventions; to look up information that is new or unfamiliar; and to record this information in a clinical journal to assure that the experiences of orientation are not lost to memory. To respond to limitations of skill practice within varied clinical assignments, the program also provides time for nurses in training to meet with the CICU Clinical Nurse Specialist (CNS) through a separate day free from patient-care responsibilities. During the Clinical Skills Day, the CNS works one-on-one with the nurse in training to hone clinical skills and provide individualized teaching on critical care concepts.

Firsthand knowledge of the patient experience across the continuum of the Cardiovascular Program is offered through observational experiences in the Catheterization Lab, the Cardiac OR, and the Cardiac Inpatient Unit.

SUCCESSFULLY INTEGRATING NEW STAFF INTO CICU ENVIRONMENT

Over the 10 months ending October 2008, 10 experienced nurses and 12 new graduate nurses have successfully completed the CICU orientation program. Feedback from our charge nurses, clinical nurse specialists, and staff nurses has confirmed the success of this program in fully integrating new staff into the CICU clinical environment. ♥



LISA MCCABE,
RN-BC, BSN, CCRN



ANNETTE IMPRESCIA,
RN, CCRN

DEVELOPING LEADERS IN CARDIOLOGY

PETER LANG, MD, SENIOR ASSOCIATE IN CARDIOLOGY AND
DIRECTOR, CARDIOLOGY FELLOWSHIP PROGRAM

The Department of Cardiology at Children's Hospital Boston enrolled its first trainee in 1951, three years before the first successful "open heart" operations were performed. Dr. Abraham Rudolf, the first fellow, was attracted by the promise of a new field, the challenge of discovery, and the offer of a salary. There were no funds available from hematology, Dr. Rudolf's initial choice for pediatric subspecialty training.

PROGRAM STRENGTHS FOUNDED ON GUIDING PRINCIPLES

Starting with Dr. Rudolf, more than 350 fellows have come to Boston to enroll in what is officially called the "Fellowship Training Program in Pediatric Cardiology and Cardiovascular Research." The fellows have been attracted by the strengths of the program, which have evolved over the years but are based on certain guiding principles. Dr. Alexander Nadas, the first chairman of our department and a true "father of pediatric cardiology," insisted that cardiology be established as an independent entity, which allowed the Department of Cardiology to set its own goals; the training of individuals prepared to be the leaders of the emerging discipline was of prime importance.

Dr. Nadas insisted that fellows have exposure to the science of cardiology, as well as clinical practice. Fellows were encouraged to explore many disciplines, including basic science in the Physiology Department at Harvard Medical School, and to explore emerging areas of interest, such as the physics of ultrasound at the Massachusetts Institute of Technology. Others were given the opportunity to participate in multicenter clinical studies, collaborating with pediatric cardiologists around the country. Today's fellows are required to complete a research project to successfully complete training.

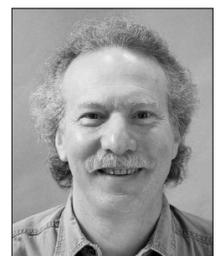
DEDICATION TO CLINICAL EXCELLENCE AND INNOVATION

Clinical excellence and innovation have been the leading factors that attract fellows to train in cardiology at Children's Hospital Boston. The strong alliance with Cardiac Surgery and Cardiac Anesthesia has allowed for many "firsts" in the care of children with heart disease.

Fellows came to the Department of Cardiology in two separate groups. The traditional fellowship is designed for trainees who have completed pediatric or medicine/pediatric residencies. They train for three years. During the first 18 months they learn the basics of pediatric cardiology, which include exposure to inpatient and outpatient management, echocardiography, cardiac catheterization, cardiac intensive care, and electrophysiology. The second 18 months allow fellows to explore areas of particular interest and complete research projects.

Another group of trainees come to Boston to serve as "Senior Clinical Fellows." They have already completed pediatric cardiology fellowships and work to develop skills in the subspecialty disciplines. The increasing complexity of pediatric cardiology makes it almost mandatory to complete this final level of training, which can last one or more years.

It would be a mistake to conclude that training in pediatric cardiology ends with the completion of a fellowship. The most important lesson that we give our fellows is the need to continue learning—from our colleagues and our students. ♥



PETER LANG, MD

INNOVATIVE CLINICAL MANAGEMENT OF SYNCOPE

NEW STANDARDIZED CLINICAL ASSESSMENT AND MANAGEMENT PLAN (SCAMP)

MARK E. ALEXANDER, MD, ASSOCIATE IN CARDIOLOGY

Syncope represents one of the truly sentinel symptoms that can bring the previously unrecognized or stable cardiac patient to medical attention. At its physiologic core, syncope represents an abrupt decline in cerebral perfusion either from exaggerated reflex mechanisms or acutely unstable, but self-limited, arrhythmias. Since the final common pathway between the essentially innocent but annoying common faints and life-threatening cardiac syncope is identical, cardiologists are regularly involved in evaluating these patients. While syncope is an important finding, the serious causes of syncope are very rare. These can include acute infectious causes, like Lyme disease and myocarditis, or familial diseases, like hypertrophic cardiomyopathy and Long QT syndrome. In contrast, common fainting, sometimes called neurocardiogenic syncope (NCS), occurs in nearly 25% of adolescents and young adults, with a peak incidence in adolescent girls. Cardiac syncope, primarily the result of self-resolving arrhythmias, is much less common. Effective triage is aimed at identifying the small number of patients with serious causes of syncope, while efficiently and appropriately reassuring and managing the much larger number with innocent syncope.

PATIENT EVALUATION FOR A VARIETY OF SCENARIOS

Syncope patients are evaluated throughout the cardiology program. Patients with a history of known heart disease, significant family history of cardiomyopathy or electrical disease, or concerning symptoms are generally rapidly triaged to more comprehensive evaluations. In addition to those with known heart disease, particular concerns include a trigger with exercise, brief convulsive events following collapse, injury, or urinary incontinence. These all raise suspicion of serious disease.

The adolescent with an isolated or infrequent typical syncopal event is usually recognized by the primary care physician, who can obtain an ECG and with clinical evaluation may not need additional evaluation. Cardiac evaluation is primarily aimed at reassuring the family that the symptoms are not the result of serious cardiac disease, while at the same time educating the child and family on ways to limit the symptoms. Often the dramatic response to increased fluids, increased sodium, and increased awareness of symptoms proves the diagnosis.

For those with more complicated scenarios more detailed investigations are available. In addition to the usual 24-hour and monthlong event monitors, there are

implantable arrhythmia monitors that permit several years of effective monitoring. Exercise testing is increasingly used as both a screen for some genetic arrhythmias and as a provocative test for those with neurally mediated syncope. These tests have assisted in proving that a number of highly competitive athletes with syncope do not have heart disease and permitted them to return to high-level competition. Genetic testing now allows more refined evaluation of families with familial arrhythmias. Those with repaired heart disease often require evaluation with diagnostic catheterizations. When therapy is ineffective or the diagnosis problematic, a limited number of patients will have head-up tilt testing done to correlate symptoms with hemodynamic and cerebral blood flow.

SCAMP TO STREAMLINE EVALUATION AND MANAGEMENT

A new Cardiovascular Program initiative developing Systematic Clinical Assessment and Management Plans (SCAMPs) aims to help streamline evaluation and management of high-frequency problems. The SCAMP process acknowledges both that (1) a reasonable data-driven approach to common clinical problems can be developed and (2) there inevitably will be choices where the proposed assessment and management plans are ineffective. By tracking actual clinical decisions, the SCAMP can be refined to reflect practice, opinion, and data.

The Syncope SCAMP will include input from our neurology colleagues. A typical faint will be defined as occurring at rest, while upright or sitting (but not supine) with a notable prodrome, recovery in < 1 minute, and little in the way of intercurrent symptoms. For these patients, a detailed history, orthostatic vital signs, normal examination, and normal ECG are sufficient for continued observation without drug therapy. Therapy options for different clinical scenarios will be standardized, with the critical notion that neither the physician nor the patient is obligated to follow those particular pathways, just to keep score.

The SCAMP process represents an exciting way of both offering more efficient care and collecting observational data regarding patients (How often is the ECG abnormal in syncope? How often is the echo abnormal?), and for exploring practice variability, answering questions such as, Why did we get an echo? ♥



MARK E. ALEXANDER, MD

NEW CARDIAC NEURODEVELOPMENTAL PROGRAM

IMPROVING OUTCOMES IN CHILDREN WITH CONGENITAL HEART DISEASE

JANE NEWBURGER, MD, MPH, ASSOCIATE CHIEF FOR ACADEMIC AFFAIRS

Advances in prenatal diagnosis, perioperative management, and postoperative care have reduced surgical mortality rates for virtually all forms of congenital heart disease. Whereas only a few decades ago, 20% of children born with a congenital heart defect survived to adulthood, by the 1980s this figure stood at 85% and has continued to increase. Indeed, the number of adult survivors of complex congenital heart disease has now surpassed the number of children. With improved survival, neurodevelopmental challenges have been increasingly recognized as children reach school age and adulthood. These problems can affect learning and development; interfere with the acquisition of academic, vocational, and social skills; and negatively affect adjustment and employability in adulthood.

MAXIMIZING DEVELOPMENT

Children who have undergone repair or palliation of congenital heart defects, compared to the general population, have significantly lower scores on IQ and achievement tests, delays in reaching motor milestones, and higher frequencies of learning disabilities, use of special services, and speech, language, and behavioral abnormalities. In general, outcomes after repair of simple lesions, such as atrial septal defects, are similar to those in the normal population, whereas developmental outcomes may be worse in some children who have had biventricular repair of more complex lesions. Developmental challenges are most common among those with various forms of single ventricle.

Many factors underlie neurological and developmental problems in patients with congenital heart defects. In addition to primary genetic and dysgenetic conditions, the brain may be affected in fetal life due to abnormal blood flow or growth disturbances related to insufficient oxygen or glucose supply. Other neurological abnormalities can be acquired after birth, either from the effects of heart disease itself (e.g., extremely low oxygen saturations, unstable blood pressure, or oxygenation before surgery) or from its therapies. Central nervous system injury may also be caused by events in the early postoperative period, when the body's usual mechanisms for adjusting blood flow to

the brain to compensate for low blood pressure or oxygenation may be temporarily disrupted by hypothermic cardiopulmonary bypass techniques. The brain may also be affected by aspects of the conduct of cardiopulmonary bypass, including acid-base management strategy, hematocrit, rate and depth of cooling, and methods of vital organ support (e.g., total circulatory arrest duration, total support time). Children's Hospital Boston has a long history of research in improving brain protection during cardiac surgery itself. We are now concerned with providing each infant who has had cardiac surgery with the support and interventions needed to reach full potential.

The Cardiac Neurodevelopmental Program has been established with the missions of enhancing the development of children with heart disease, providing support and assistance to their families, and maximizing their future contributions to society. These goals are based upon quantitative and qualitative data in children with other conditions, showing that early intervention can improve the development of the child and functioning of the family, and reduce requirement for special services later in life.

A child is eligible for participation in the program if he or she has had cardiac surgery before age one year of life or is referred by a cardiologist or other caretaker for testing to determine neurodevelopmental delay or disability. Developmental challenges of cardiac infants are very similar in severity and scope to those of premature infants. For this reason, we are currently providing developmental evaluation of infants who have had cardiac surgery before their hospital discharge. The evaluation includes screening for problems that would warrant a cardiac genetics consultation. After hospital discharge, we are offering follow-up evaluation at ages one, two, and three years to all infant graduates of our cardiac intensive care unit. Finally, select patients may be evaluated earlier than age one year if indicated by the in-hospital evaluation and family preference.

RESEARCH REGISTRY

Although we have come a long way toward improving outcomes in children with congenital heart disease, further research is needed to

improve the understanding and minimize the risk factors for adverse neurodevelopmental outcomes. For this reason, the program has a research arm, the Cardiac Neurodevelopmental Research Registry. The Research Registry serves as a source of information for research studies that explore risk factors for adverse neurodevelopmental outcomes in children with congenital heart disease. Specifically, the Research Registry will maintain a computer repository of medical information and neurodevelopmental test results, which can provide data for specific research projects. Children (and their families) are eligible to participate in the Research Registry if they were younger than age one year at their first surgery and have been evaluated in the clinical Cardiac Neurodevelopmental Program. The Research Registry also provides an infrastructure for specific research protocols with neurodevelopmental endpoints. The identity of subjects participating in the Research Registry is kept anonymous, and participation requires informed consent of parents and guardians, as well as assent of older children.

We are very excited about this opportunity to help our patients. Please don't hesitate to contact us for any questions. ♥



JANE NEWBURGER,
MD, MPH

Contact Information:

Donna Donati, Program Coordinator
Telephone: (617) 355-3401
E-mail: donna.donati@cardio.chboston.org

Developmental Psychology
Janice Ware, PhD
David Bellinger, PhD
Samantha Butler, PhD

Cardiology
Jane W. Newburger, MD, MPH

Cardiovascular Surgery
Frank Pigula, MD

Cardiovascular Genetics
Amy Roberts, MD

Nursing
Carolyn Dunbar-Masterson, RN
Ellen McGrath, RN
Bethany Trainor, RN
Judith Geva, MSW

HEARTS OF GOLD

When the phone rings in the middle of the night, it's usually not good. I always hope it's a wrong number, someone confused in Australia, unaware of the time difference, looking for his girlfriend. Nonetheless, most people have a minute of fear with that two a.m. ring, thinking the worst. This phone call came in the middle of the afternoon, routine to say the least. "Mom, we just came from the doctor and got some scary news. Travis has a hole in his heart and it can only be repaired by open heart surgery."

So begins *A Fragile Heart*, Barbara Trachtenberg's memoir about her family's poignant journey through a little boy's heart surgery. Together, Barbara and her husband Stan, daughter and son-in-law Julie and Paul D'Amours, and Paul's family, rallied around 18-month-old Travis.

A PARENT'S WORST FEAR

Even though Travis had a persistent cough and his growth had dropped to the fifth percentile, he otherwise appeared healthy. Barbara describes him as "an active, ruddy little guy who was the apple of our eye." While doctors were not overly concerned, Julie was. Her fears were well founded: At Travis's 18-month checkup, his doctor detected a very faint, but atypical, heart murmur. He referred Travis to a cardiologist, who discovered a hole in the little boy's heart that was unreachable by catheter. Since the D'Amours lived in Wyoming, the recommendation was for open heart surgery in Utah.

Over the following months, Julie and Paul researched options to ensure that Travis would receive the best care, regardless of where they had to go. After meeting a Wyoming family whose now-healthy daughter had undergone the same surgery at Children's Hospital Boston, Julie and Paul made their decision: They would travel to Children's to save their son.

SAVING TRAVIS

In June 2007, the D'Amours met with Children's cardiologist David Brown, MD, who discovered that the hole was larger than anyone realized—it had caused Travis's heart to enlarge, placing pressure on his ribcage. If Travis did not receive surgery soon, he risked developing permanent problems. A few months later, on September 25, 2007, cardiac surgeon Emile Bacha, MD, successfully repaired the hole in Travis's heart. Even though Dr. Bacha closed the

hole, the volume of one vessel was slightly abnormal. Because the vessel was close to the sinus node, which regulates heart rhythm, Dr. Bacha's team decided not to operate, reassuring the family that the vessel should not cause problems. And if it did, the problem was easily correctible without surgery. Not only was Travis's prognosis good, but the family had found compassionate advocates and friends in their Children's caregivers. As Barbara explains, "It's something you go away from a changed family."

Travis is now a thriving three-year-old. Says Barbara, "Travis is the most active child I've ever met. When I say to him, 'Get in your car seat, sweetheart,' and he says, 'No!' and refuses to budge, I don't find it frustrating, because look at the alternative. We count our blessings and know every moment matters."

GIVING BACK

To give back to the hospital that has given them so much, Barbara and Stan established the Travis Help a Heart Fund for Cardiac Surgery, which supports patients and their families from around the world who need cardiac surgery but have severe financial constraints. "I knew our family was fortunate to be in the position where we could fly to Boston to be with Travis," says Barbara. "I couldn't deal with the fact that because families didn't have the money, they didn't have this option. Or, even worse, their child wouldn't have access to life-saving surgery. It's just not fair." Barbara and Stan vowed to do their part to ensure these families receive the care they so desperately need.

Five-year-old Amber, born in the Dominican Republic with several heart defects, is a testament to the miracles possible through the Trachtenbergs' generosity. Dr. Bacha successfully operated on Amber this spring. "I'm amazed Amber survived as long as she did," Dr. Bacha says, "and were it not for the Travis Help a Heart Fund, she wouldn't have had access to crucial surgery—and she likely would have died within two years."

Barbara and Stan's goal is to help two children like Amber each year. Says Barbara, "Economies come and go, but little kids' health will always be important." Are you interested in championing the Trachtenbergs' mission and helping Children's save more lives? Please contact Sara Conahan at (617) 355-4007 or sara.conahan@chtrust.org. ♥



Children's Hospital Boston

CARDIOVASCULAR PROGRAM

300 LONGWOOD AVENUE
BOSTON, MASS 02115

NONPROFIT ORG.
US POSTAGE
PAID
BOSTON, MASS.
PERMIT NO.
59240