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Foreword

Cardiology 2007 – 10th Annual Update on Pediatric Cardiovascular Disease

Gil Wernovsky

The Cardiac Center at The Children's Hospital of Philadelphia hosted our 10th Annual Postgraduate Course at Disney's Yacht and Beach Club Resorts in sunny Orlando, Florida from February 21st-25th, 2007. Over 750 professionals and exhibitors (Fig. 1) gathered from around the globe to hear late breaking research, discuss controversial topics, review current practices and enjoy each others company and insight. I was privileged to learn from such a distinguished faculty and insightful group of course attendees, and we look forward to planning our 11th meeting next year in Scottsdale, Arizona.

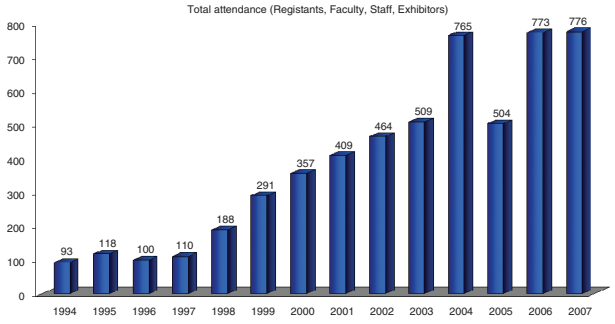

Fig. 1.
A chart showing the gradual increase in the numbers attending the meeting over the years since its inception.

What distinguishes this postgraduate from other excellent subspecialty meetings is the multidisciplinary group of course attendees (Fig. 2). Physicians made up approximately half of the attendees, and included representation from all disciplines involved in the care of children with cardiac disease, including cardiologists, intensivists, surgeons, anesthesiologist, neonatologists and maternal fetal specialists (Fig. 3). Approximately 50 physicians were fellows, residents or medical students. The remaining attendees included advanced practice, operating room, catheterization lab and bedside nurses, sonographers, physician assistants, respiratory therapists, and the largest group of perfusionists

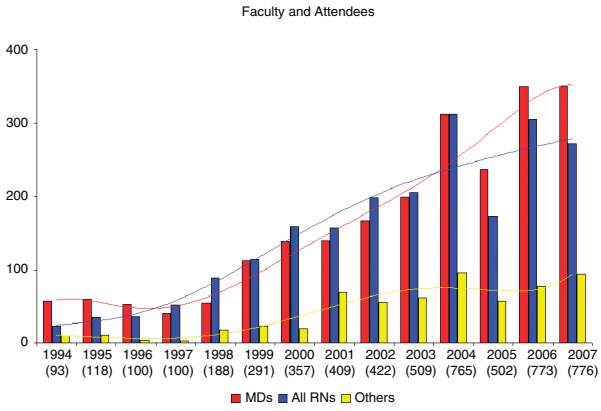

Fig. 2.
The breakdown of attendees over the years according to profession.

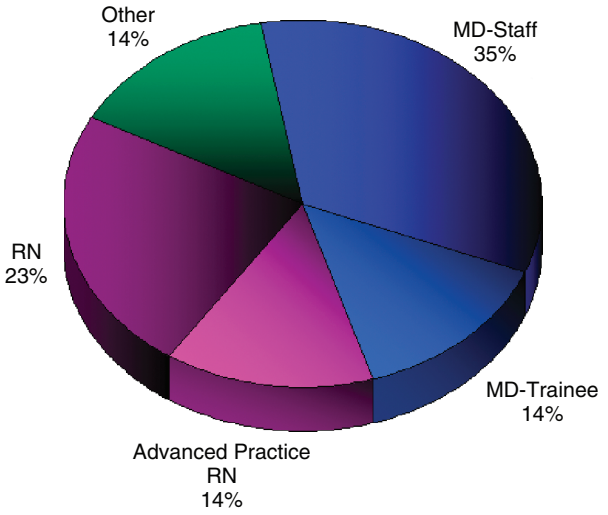

Fig. 3.
The breakdown of professions amongst those attending the event of 2007.

ever present at our meeting, specifically 48. A growing number of administrators also continue to partner with the providers of care on the front line in sessions devoted to delivery of care, fiscal responsibility, and patient safety.

Featured Research, Posters and Abstracts

From over 100 abstracts initially submitted, 65 were presented in two separate poster sessions from 40 institutions and 16 countries. Those that have not been previously published as abstracts are included later in this electronic supplement. The top three abstracts were presented as oral presentations by Kirsten Odegard of Children's Hospital, Boston, her title being "Prospective longitudinal study of coagulation profiles in children with hypoplastic left heart syndrome from stage I through Fontan completion", John Costello, also of Children's Hospital, Boston, who presented "A systematic initiative to reduce blood stream infections in a pediatric cardiac intensive care unit", and Jon Kaufman, from Children's Hospital of Denver, and the University of Colorado Health Science Center, who gave the talk entitled "Correlation of abdominal oximetry with gastric tonometry: measurement of splanchnic oxygenation in neonates and infants with congenital heart disease". In a very close vote by the entire faculty, the 4th Annual Outstanding Investigator Award was given to Dr. Kaufman and his colleagues.

Surgical innovations and practices in complex congenital heart disease were the focus of many of the posters. William Douglas, from Lexington, presented information on an adjustable flow device for systemic-to pulmonary arterial shunts; Daniel Nento, from Cleveland, presented improved outcomes in side-to-side, as opposed to end-to-side, techniques in augmenting the ascending aorta in patients with hypoplastic left heart syndrome, and Martin Zahorec, from Bratislava, discussed the role for closure of the arterial duct in patients undergoing placement of a modified Blalock-Taussig shunt. The most challenging aspects of hypoplastic left heart syndrome and other functionally univentricular diseases were tackled with researchers showing several "outcome challenges." Areas covered included results of the Fontan palliation in children with trisomy 21, given by Francis Moga, from Minneapolis; catheter intervention for hypoplastic left heart syndrome with intact atrial septum, by Chris Petit from Philadelphia, overall neurodevelopmental abnormalities by Anke Furck, from Kiel, and the role that inherent coagulation abnormalities may play, even prior to any surgery, in patients with functionally univentricular hearts, the latter presented by Nina Hakácová, from Bratislava. Lindsay Ryerson, from the University of Michigan, discussed the role of heparin therapy in protein losing enteropathy. Wasim Khan, from Rainbow Babies Hospital in Cleveland, described the physiological responses to exercise in children after the Fontan operation, while the group from The Children's Hospital of Philadelphia utilized magnetic resonance imaging to study the effects of exercise on power loss after Fontan palliation. Collaboration between institutions was evident in the efforts by the team from Duke University, presented by Maura Catherine Baldwin, to validate the follow-up program reported by The Children's Hospital of Wisconsin.

High fidelity simulators were shown significantly to improve the confidence of nurses working in a cardiac center in critical scenarios, an important support in times when rapid development of education and skills is an issue for many centers. This work was presented by Roberta Hales, from Philadelphia. Simulators were also shown to improve time for cardiac care teams to initiate extracorporeal membrane oxygenation, with this investigation presented by Cecilia St. George-Hyslop, from Toronto. Resuscitation scientists also showed improvements in cardiopulmonary resuscitation techniques with use of a Voice Advisory Manikin system, the research summarized by Robert Sutton, from Philadelphia.

Administrators, and others taking a "50,000 foot view" of our systems for care, were benefited by a cost analysis of various mechanical support techniques as a bridge to cardiac transplantation, the research presented by Andrei Morgan, and based on the experience from Newcastle-Upon-Tyne in the United Kingdom. Shabib Aldaheri, from Syracuse, discussed the role that virtual regionalization models can play in improving outcomes when providers can be mobile, but care cannot be given under one roof. Sonographers and their colleagues had their interests focused on the roles for the discharge echocardiogram in improving outcomes by Christa Barlow, from Cincinnati, and by Sarah Gelehter, from Ann Arbor, Michigan, on the fact that that pulmonary venous Doppler signals can correlate well with left atrial pressures.

The field of electrophysiology was investigated in five abstracts. Aimee Malpass and colleagues from the University of Mississippi showed that amiodarone is both safe and effective in the acute management of supraventricular tachycardia in neonates. The clinical and electrophysiologic characteristics of both left and right ventricular tachycardia in children with structurally normal hearts were presented by Nelangi Pinto from The Children's

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Hospital of Philadelphia. Armando Alfaro and colleagues from Costa Rica showed that ventricular single lead pacing is an effective and safe method to maintain atrioventricular synchrony in third degree atrioventricular block.

Fetal cardiology was also strongly represented. Paul Brooks studied the effects of antenatal diagnosis of pulmonary atresia with an intact ventricular septum and found that while there was an improvement in systemic oxygenation prior to the initiation of prostaglandin E1, there was no overall improvement in mortality. Kirsten Dummer reported from the Boston experience that prenatal diagnosis of tetralogy of Fallot is highly accurate and it is unusual to see progression to pulmonary atresia. Anita Szwast, from The Children's Hospital of Philadelphia, demonstrated that maternal hyperoxygenation in the fetus with hypoplastic left heart syndrome is a safe and useful procedure to assess the pulmonary vasculature. Lisa Wise-Faberowski, from the University of Colorado, presented two abstracts utilizing a chronic hypoxia model in the developing brain to describe the use of magnetic resonance spectroscopy, as well as the effects of isoflurane.

Several abstracts focused on patient care from the nursing perspective, and included a report from Stanford that emphasized the importance of continued nursing educational sessions as a mechanism to improved professional autonomy and enhanced patient safety, a similar point made by the nursing staff at The Children's Hospital of Philadelphia who routinely utilize the benefits of an educational resource nurse in the training of new cardiac intensive care nurses. Nursing and safety interventions were the highlight of many posters. An analysis of teams led by pharmacists in exploring the safety of paediatric medications was sufficient to win the award for the outstanding nursing poster for Jennifer Costello, from Newark. Deanna Edwards, from Little Rock presented on the checklists to ensure the safety of patients at handovers, and Diana Alexander, from Baltimore, discussed the targeted mining of administrative databases for identification of errors in cardiovascular medication. All of these were shown to play roles in improving outcomes and safety for our most vulnerable patients. Augmentation of the control of pain from sternotomy with continuous local infusions of anaesthetic, given by Marilyn Torres, from Miami, and programmes coordinated by nurses to improve the success of enteral feeding, given by Britt Elin Fredriksen, from Oslo, Norway, both drew comments and crowds in the busy session.

In the featured research session, Andrew Redington, from Toronto, 'teased' the audience with the study design of a randomized trial of aspirin and warfarin for patients with a Fontan circulation, and then told us that the results were still being analyzed....undoubtedly to garner an invitation to speak next year! Scott Bradley and colleagues from the Medical University of South Carolina performed a prospective study of a cohort of neonates undergoing midline reconstruction of the aortic arch, both for hypoplastic left heart syndrome and biventricular repairs, and reported a surprisingly high incidence of swallowing dysfunction, aspiration, and injury to the vocal cords. Barry Byrne, from the University of Florida in Gainesville, reported on important work into both the mechanism and treatment for inherited cardiomyopathies. Lynn Mahoney updated the group on the exciting work being conducted by the Pediatric Heart Network, sponsored by the National Heart, Lung and Blood Institute. See http://www.pediatricheartnetwork.com/ for more details.

Plenary Sessions

The opening session on cardiopulmonary resuscitation featured experts in the fields of resuscitation science, nursing, cardiac intensive care and pediatric cardiac anesthesia. Vinay Nadkarni, from Philadelphia, and Mary Fran Hazinski, from Nashville, reviewed the current state of the art and the science behind the guidelines provided by the American Heart Association for pediatric resuscitation. Sarah Tabbutt, from Philadelphia, gave excellent examples of why these guidelines are not always relevant to postoperative patients with congenital cardiac disease, and Ravi Thiagarajan, from Boston, showed us some practical and convincing work on the improvement gained with a simulation program for staff in an intensive care unit. George Hoffman, from Milwaukee, gave a comprehensive review of the literature on post-resuscitation cerebral protection, and speculated on the relevance to children following cardiac surgery. The first day ended with a spirited debate between Tom Spray, from Philadelphia, and Jim Tweddell, from Milwaukee, on the risks and benefits of open cardiac massage during resuscitation. The video provided by Tom, comparing the size of his hand to that of a typical neonatal heart, was quite visually "revealing" of the difficulties of direct massage in small neonates.

Two plenary sessions were given back-to-back on the second day of the conference, covering coronary arterial disease in children, and double outlet right ventricle. Jack Rome, from Philadelphia, moderated the session on coronary arterial disease in children, which began with Alan Friedman, from New Haven, reviewing normal coronary arterial anatomy, followed by a discussion on the anatomy,

haemodynamics, and clinical characteristics of abnormalities of coronary arterial origin, including anomalous coronary artery from the pulmonary trunk and intramural coronary arteries, as well coronary fistulas, and Kawasaki Disease. Michele Frommelt, from Milwaukee, discussed how to best image the coronary arteries by echocardiography, which was followed by Paul Weinberg, from Philadelphia, showing superb anatomic specimens of the previously discussed anomalies. Mark Fogel and Jeff Hellinger, both from Philadelphia, subsequently advocated use of cardiac magnetic imaging and computerized tomographic imaging to delineate these anomalies, showing beautiful images, including the "flythru" technique of three dimensional imaging. David Nykanen, from Orlando, illustrated the continued necessity for coronary angiography as an additional imaging modality, especially in evaluation and potential intervention of several coronary arterial anomalies. Paul Stephens, from Philadelphia, discussed the determinants of coronary flow, including how best to assess abnormalities of myocardial perfusion. The session concluded with the debate on whether children with an anomalous origin of a coronary artery from the 'wrong' sinus of Valsalva should undergo surgical correction. Tim Feltes, from Columbus, Ohio, argued that only those who have evidence of ischaemia should have surgery, as the true risk of a sudden catastrophic event is unknown. Jim Tweddell countered that surgery should occur irrespective of ischaemia, since there is an increased risk of ischaemia and sudden death. By a show of hands, the winner by a small margin was Tim Feltes.

The plenary session discussing double outlet right ventricle was moderated by Jack Rychik, from Philadelphia. Bob Anderson, from London, England, convinced the audience that double outlet right ventricle is not a single isolated defect, such as tetralogy of Fallot, but rather is a series of different relationships between the great arteries, where both are supported by the morphologically right ventricle, the interventricular communication being the most important feature. Paul Weinberg followed showing beautiful specimens, including those with mitral atresia and straddling of the mitral valve, as well as various relationships of the interventricular communication to the arterial trunks. Meryl Cohen, from Philadelphia, reviewed the key goals of the echocardiographic evaluation, emphasizing the relationship of the interventricular communication to the arterial trunks, the number of interventricular communications. the relationship of the great arteries to each other, the relative size of the ventricles, and the potential pathway from the left ventricle to one or other arterial trunk through the interventricular communication. This point was re-emphasized by Pedro del Nido, from Boston, who showed that the creation of a three-dimensional baffle from the left ventricle to the aorta or the pulmonary trunk is the greatest surgical challenge in these patients, and advocated for three-dimensional imaging as a routine part of the preoperative assessment.

For a change of pace, a 4th plenary session—"Improving Safety for Cardiovascular Patients"—proved to be an exciting testament to the work being done by so many colleagues in our field to improve the safety of our systems for delivery of care. Peter Laussen, from Boston, led off the session with a wonderful overview entitled "Changing culture for continuous quality improvement." Defining moments were described that required novel efforts to improve governance and management, and thus effect a change in culture. Peter showed the central role of nursing leadership in promoting change, and in leading to improved consistency in care of patients. Adequate support for change in culture can require significant financial resources, and emphasis was placed on the accountability of the attending physicians. We heard about the programme for quality and the safety of patients in place at Children's Hospital in Boston, the importance of audits in measuring and improving performance, and the imperative for leadership to promote a balance of work and personal goals. Karen Harrington, from St Justine Hospital, Montreal, gave an absolutely outstanding presentation "How may we improve communication during patient handover?" The audience was treated to a broad review with ideas of safety as a "dynamic non-event", communication breakdowns as a major cause of sentinel events, and that a knock-on effect of restrictions in working hours is an increased need for handovers, and an imperative to do them well. Karen implored our field to recognize handovers as both a transfer of responsibility and accountability, as well as information, and that improvements should be driven by local gradual change. Troy Dominguez, from Philadelphia, reviewed the risk factors for nosocomial infections, with an emphasis on infections of the blood stream associated with central lines, ventilator associated pneumonia, infections at the site of surgical incisions, the particular vulnerability of patients with congenital cardiac disease given their high number of risk factors, and the crucial role of adopting a team approach for better control and prevention. Richard Ohye, from Ann Arbor, in an effort towards "Ensuring patient safety during clinical trials, the ethics of human subjects' research" gave surely the most humorous presentation of the session.

The information given, though, was absolutely critical, and gave the audience plenty into which it could sink its teeth. Rick dared us to ask "Do trials help patients and are they relevant?" He reviewed definition of various clinical trials, problems with historically controlled trials, the benefits of randomization, and the clever way in which, given the inexorable progress in medicine, trials using historical controls always tend to favor the current intervention at question. We were implored for medicine to be based in and promoting of research – with its attendant requirements for respect for persons, beneficence, justice, and equipoise. John Charpie, also from Ann Arbor, followed with a presentation on "Developing and maintaining an effective quality assurance and improvement program." We learned the multiple aspects of quality, such as it being safe, effective, patient-centered, timely, and so on, and the details and rationale behind the "Plan-Do-Check-Act" cycle for improvement. Patricia Hickey, from Boston, riveted us with her bold opening that mandated ratios for nurse staffing are simply not the answer, as they can exacerbate work-related stress for staff nurses. Her presentation, "Rational work-hour assignments for bedside nurses: what's the rationale?" described that best practice in addressing the demands of nurse staffing depends on a multifactorial approach. The best models allow staff to respond to the demands of patients without having a negative impact on the staff themselves. Nurses should maintain control and choice over their own schedules. Introduced to the edict of Florence Nightingale to "put patient in best condition for nature to act," the audience was then led through the "Nightingale metrics" adopted by Patricia and her team, and their novel "continuity of care index." The session closed with a terrific debate between Martin Elliott, from London, England, and Alan Friedman on the pros and cons of restricted work hours for physicians. To most accounts, despite excellent arguments by both combatants, the contest was a draw, perhaps because all of the junior doctors had reached their work quota and were at the pool!. Patient safety, nonetheless, remains a crucial element in improving outcomes for our patients. Look forward to upcoming meetings from Children's Hospital, Boston (http://www.childrenshospital.org/clinicalservices/Site457/mainpageS457P15sublevel119Flevel120.html) the Hospital for Sick Children at Great Ormond Street in London, and The Children's Hospital of Philadelphia (www.chop.edu/cardiology2008) to promote the safety of cardiovascular patients.

The closing plenary session, devoted to Heterotaxy and Isomerism, was perhaps the most well attended Sunday morning session of the past 10 years. The session was dedicated to the work of, and was in honour of, Stella Van Praagh. She was lauded by all of the speakers for her insight into complex congenital cardiac disease, her compassion, her love of teaching her patients and her family. Bob Anderson and Paul Weinberg concisely reviewed a very complex topic where nomenclature is perhaps the most controversial issue. For those of us, like myself, who were taught and 'grew up' thinking of these diseases as "polysplenia" and "asplenia", Bob made a very convincing argument to stick to nomenclature based upon the anatomy of the atrial appendages. Paul Weinberg compared Heterotaxy syndrome to "Murphy's Law": if something can go wrong, it will! Peter Gruber, from Philadelphia, showed some new and exciting information regarding the genetic basis of abnormalities of sidedness being a 'proximal' event in cardiogenesis, with absolutely fascinating microscopic images of cilia directing cardiac cells to one side or another. Leo Lopez, from Miami, reviewed the anomalies as shown by echo, Jack Rome, from Philadelphia, did the same with angiography, and Mitchell Cohen, from Phoenix, showed some very elegant examples of the abnormalities of the conduction system, making total sense of a remarkably complex issue. The session closed with newly released data from the Pediatric Heart Network presented by Andy Atz, from Charleston, and relating to the recently completed Fontan cross-sectional study. Interestingly, despite having a higher complexity of disease, and most likely a higher initial risk of death, the current survivors appear to be no different than other patients with a Fontan circulation in terms of exercise performance, laboratory assessments of ventricular function, and importantly, patient and parent reported quality of life.

The featured lectures this year were given by Martha Curley, from Philadelphia, Philipp Bonhoeffer, from London, England, Pedro del Nido, from Boston, (Fig. 4), and Gavin Kerr, from Philadelphia. Martha gave an inspiring overview of what it takes to be a mentor, what, and how, we learn from them, and how they start a generational process of learning, teaching and compassion. Philipp reviewed percutaneous pulmonary valvar therapy from its humble beginning in the cow fields of France, where he collaborated with butchers, through design of the stents, involving collaboration with jewelers, through final implementation in patients, involving extensive collaboration with industry. His internationally known work was interspersed with funny anecdotes, and wonderful photos from his work in Africa. Pedro reviewed the pioneering work being done in Boston to 'recruit'

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Fig. 4.
The named lecturers, Martha Curley with Pedro del Nido to her left, and Philipp Bonhoeffer to her right. Gavin Kerr gave the fourth featured lecture.

two ventricles in the subgroup of patients with hypoplastic left heart syndrome and aortic and mitral stenosis. His presentation covered surgical technique, fetal intervention and the underpinnings of basic science, suggesting abnormalities of cell migration being the common denominator of such diverse diseases as pulmonary venous stenosis, mitral valvar pathology, and endocardial fibroelastosis. Finally, Gavin Kerr gave the first Keynote Lecture in Cardiovascular Administration, highlighting the partnership necessary between givers of care on the front line and those responsible for hospital administration.

Breakout sessions

Echocardiography breakout sessions were well attended throughout the meeting, including overviews of three dimensional imaging, provided by Girish Shirali, from Charleston, fetal imaging, by Jack Rychik, from Philadelphia, and intracardiac imaging, presented by Mike Brook, from San Francisco. Difficult cases were reviewed by the faculty in an informal session, and Leo Lopez, Michele Frommelt and Norman Silverman, also from San Francisco, gave wonderful overviews of specific cardiac lesions.

Similarly, the inpatient breakout sessions for intraoperative care, surgery, perfusion and intensive care were comprehensive and insightful. New strategies in monitoring using near infrared spectroscopy are becoming more ubiquitous in our field, and front line experts from Houston, Milwaukee and Charleston updated the group on current strategies and results. A particular focus of all of the breakout sessions was on reducing morbidity, particularly to the central nervous system. Dean Andropoulos and Chuck Fraser, Jr., from Houston, presented their current "best-practice" model to intraoperative perfusion and monitoring, followed by informal comments from practitioners in Philadelphia, Boston, Ann Arbor and Milwaukee. Lara Shekerdemian, from Melbourne, Australia, then updated the audience on their prospective studies following neonatal surgery in Australia. Andrew Redington once again showed some seminal work being done on remote ischaemic preconditioning, perhaps one of the best examples of translational research we have seen in our field.

The outpatient cardiology breakout sessions were also comprehensive and well attended, including topics such as pulmonary hypertension, routine screening for cardiovascular disease, cardiac transplantation, Barth's syndrome, Marfan syndrome, exercise testing and outpatient decision analysis, to name just a few. Smaller subspecialty breakouts were also held in developmental cardiology, electrophysiology, interventional catheterization and cardiovascular administration.

Nursing has traditionally been a particularly strong component of this meeting, especially in terms of the science presented by the faculty (Fig. 5), and the number of attendees from all disciplines in nursing. This year was no exception, with a featured nursing plenary session, a hands-on research planning session, and nearly 20 abstracts submitted by nurses from around the globe. Barbara Medoff-Cooper, from Philadelphia, and Mary Fran Hazinski, moderated the first nursing plenary session, which focused on the care of neonates with hypoplastic left heart syndrome. Eliot May, from Milwaukee, led a thorough discussion of low cardiac output in the early postoperative period and the

Fig. 5.
A representative group from the impressive and extensive Nursing faculty.

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usefulness of continuous monitoring of mixed venous oxygen saturation and near infrared spectroscopy in early detection and early intervention. Erika Wintering, from Phoenix, followed with an excellent review of preoperative and postoperative ventilation strategies, using examples and research to highlight key points. The remainder of the session addressed long term morbidities. Barbara Medoff-Cooper presented late breaking results from her study funded by the National Institutes of Health of patterns of feeding in newborns with congenital cardiac disease, and focused on some of the concerning results in this population—reduced length, weight, and head circumference at 3 and 6 months of age. Jo Ann Nieves, from Miami, reviewed the results from her international survey on practice variability, and Nancy Rudd, from Milwaukee, reviewed the results of their formalized programme for home-monitoring, which has resulted in a dramatic reduction of interstage mortality at that center. Kathy Mussatto, also from Milwaukee, closed the session with an insightful look at factors affecting quality of life for our patients and their families.

The second nursing plenary session focused on low cardiac output syndrome. Jo Ann Nieves and Erika Wintering reviewed several strategies for recognition and management of low cardiac output, which was followed by a comprehensive review of mechanical support of the failing circulation by Lisa Moore, from St. Petersburg. A special sunrise session was held with Barbara Medoff-Cooper and Kathy Mussatto on how to get nursing research off the ground, funded and published, followed by presentation of the top three nursing abstracts.

For the first time at this meeting, a full half-day session was devoted to the planning of Careers for Trainees and Junior Faculty, including rarely-discussed topics at scientific meetings, such as job interview skills, negotiating salary and benefits, comparisons of academic and private practice, work-life balance, managing funding and choosing an academic career and mentor. The session was graded very favourably by the trainees and junior faculty alike, and will most likely become a routine part of future meetings.

An opening plenary session featured a wonderful talk on why clinical practitioners of paediatric cardiology need to know about basic science from Jon Epstein, from Philadelphia, a self-flagellating talk by Martin Elliott on what he endured during his surgical training, and commentary by yours truly on how the theory of cognitive dissonance in social psychology pertains to the way we read, and sometimes dismiss, the literature, and respond to new and/or conflicting data.

Two pre-conferences were also held, one on cardiac anatomy and imaging, and one on resuscitation. The anatomic pre-conference was attended by nearly 150 physicians, nurses and sonographers, and featured anatomic specimens from the Cardiac Registry in Philadelphia and the Van Mierop collection in Gainesville. Wonderful correlations between the specimens and imaging examples brought it all together for the attendees.

The simultaneous session on 'mock codes' was coordinated by Stacie Peddy, representing the team from the Center for Simulation, Advanced Education and Innovation at The Children's Hospital of Philadelphia (Fig. 6). Small group didactics were held, but most of the experience was 'hands-on', with real life cardiac intensive care scenarios, and the use of the SimBaby from Laerdal. Working in small groups, the participants were taken through both predictable events, such as low cardiac output and tachyarrhythmias, as opposed to sudden and unanticipated events, such as respiratory failure, cardiac tamponade and shunt thrombosis, that characterize the 24 to 48 hours after cardiopulmonary bypass. In addition, preoperative scenarios, such as the hypercyanotic spell in the patient with unrepaired tetralogy of Fallot, and medical scenarios, like acute fulminant myocarditis, were utilized. Each SimBaby station was staffed with clinical and simulation facilitators, and teams of 3 to 4 participants "rotated" through all 7 scenarios. Through open communication and assignment of roles, each team actively worked through the clinical scenario presented to them. The learning objectives were reviewed, and an inclusive debriefing session was given at the completion of each scenario. The scientific sessions were rounded out by sunrise hands-on sessions on near infrared spectroscopy, cardiopulmonary bypass, automatic external defibrillators and permanent pacing.

Fig. 6.
The Faculty for the "Mock Codes" session including two "SimBabys".

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New learning technology and social events

No conference would be complete without a bit of fun and games. The weather could not have been better for the 3rd annual pre-conference golf tournament at the beautiful Lake Buena Vista golf course, one of Disney's finest. Twenty golfers of diverse talents journeyed from the farthest reaches of the earth, such as London, Michigan, and Wisconsin, to gather for their early morning tee times. Jim Tweddell, Mitch Cohen, Christine Anderson and Paul Stephens had 5 birdies and a single bogey for the winning score of 4 under, one a shot ahead of Jon Fleenor, Mike Mulreany, Scott Maurer and Bert Ross. Scott Maurer hit the longest drive on the par 4 ninth, which plays 360 yards. His tee shot landed only 30 yards from the green, so if my math serves me right, that is 330 yards of muscle and accuracy. Please plan on attending the 4th annual tournament next year in Scottsdale.

An audience response system was used for the first time to document anonymous responses by the audience to various clinical scenarios. In the physician session, a number of cases were shown, similar to the surgical conference as currently existing in most centers, and once again highlighting areas that require further investigation, such as optimal strategies for management for common disease, for example, small ventricular septal defect with aortic regurgitation, or management of atrial septal defect, and consensus guidelines for outpatient follow-up. In the afternoon session, nearly 500 respondents were led by Geoff Bird, from Philadelphia, in a series of clinical scenarios which highlighted the incredible diversity of management styles around the globe, including management of hyperglycemia, with as many respondents using insulin protocols as those who offered no treatment, types of inotropic support and the routine use of delayed sternal closure.

Mike Brook starred as Alex Trebek in Cardiology Jeopardy, simulating the popular game show (Fig. 7). Three teams competed on topics from geography, radiographs, boils and sores and historical facts. The 'home' team from The Children's Hospital of Philadelphia, made up of Tom Spray, Jack Rychik and Gil Wernovsky, was pitted against a team from the United States, comprising Norm Silverman, Tim Feltes and Jim Tweddell, and a Commonwealth team of Andrew Redington, Martin Elliott and Dan Penny. Despite cries of 'foul', and controversial answers, the team from the United States won in a close battle. Jeopardy is sure to become a standard offering in future meetings.

Fig. 7.
The gathered contestants in "Cardiology Jeopardy".

Fig. 8.
A gathering of attendees at "Pleasure Island".

Fig. 9.
Part of the faculty producing musical entertainment.

Finally, there were a number of networking receptions for nurses, trainees, staff physicians and perfusionists, and the Gala Reception (Fig. 8) was

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Fig. 10.
Mike Quartermain and Chris Petit during the Cardiology Ensemble.

Fig. 11.
Gil Wernovsky and Philipp Bonhoeffer discussing their approach to the theme from "Schindler's List".

followed by the 1st annual Cardiology Ensemble, featuring musical numbers from the faculty and attendees (Figs 9–11).

Future Directions

Cardiology 2008 is planned to take place from February 6 through 10, 2008 in Scottsdale, Arizona. Highlights from the meeting summarized in this foreword, as well as those from the 7th Annual International Symposium on Congenital Heart Disease held at St. Petersburg, Florida, will be published as a supplement to Cardiology in the Young in the Fall of 2007.

I am grateful to all of my colleagues at The Cardiac Center at The Children's Hospital of Philadelphia, who provided some of the material for this Foreword and helped plan and execute Cardiology 2007.

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Cardiology 2007

Cardiology in the Young

Abstracts from Cardiology 2007: 10th Annual Postgraduate Course in Pediatric Cardiovascular Disease, which took place in Orlando, Florida from February 21–25, 2007

Characterization of Cardiovascular Medication Errors in Children: An Analysis of the USP MedMarx Database: 2003–2004

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Background: Medication error rates are reported to be as high as 1 in 6.4 orders with more severe errors in children (31%) compared to adults (13%). Children are vulnerable given weight based dosing, wide ranges of therapeutic doses, "off label" use of medications, and lack of universal pharmacy compounding practices. We hypothesized that there would be a higher percent of harmful errors in pediatric inpatients using certain "high risk" cardiovascular drugs compared to other cardiovascular medications. Our second hypothesis was that children <1 year would have more error reports with a higher percent of them citing harm compared to older children.
Methods: We analyzed 2003–2004 data from the United States Pharmacopeia's (USP) MedMarx computer-based medication error reporting system used in all 50 States. Reports for patients <18 years old citing at least one medication from a cardiovascular therapeutic class were included. Facility characteristics and report demographics were categorized. Reports were stratified by USP harm score: "A": near miss, "B–D": error, no harm and "E–I": harmful error. Frequency of reports and percent of harmful reports were determined for each therapeutic class and age group. "High risk" drugs were defined a priori as antiarrthymics, antihypertensives, digoxin, and calcium channel blockers. Fisher's exact test was performed to assess differences between groups. Statistical significance was set at $p \leq 0.05$.
Results: 147 facilities submitted 821 error reports for children involving cardiovascular medications. Seventy one percent (71%) were community hospitals and 22% university and children's hospitals. Age ranged from 1 day to 17 years (mean: 4.14 years); median (0.90 years). The most common locations were NICU (22%), General Care Unit (20.5%), PICU (15.6%), Pediatrics (15.2%) and Inpatient Pharmacy (12%). Drug administration was the most commonly implicated step of the medication process accounting for 40% of reports. Delivery of an improper dose despite a correct medication order occurred in 26.5% of reports, with implication in 45% of harmful errors. Severity analysis showed 4.9% (40) "near misses", 91.4% (750) "error, no harm", and 3.8% (31) "harmful errors." 893 cardiovascular products were cited in 821 reports. Diuretics were cited most frequently (43%), then antihypertensives (11%), ACE inhibitors and angiotensin receptor blockers (9%), beta blockers (8.4%), digoxin (7.8%) and calcium channel blockers (4%). Drugs with highest percent harm were nesiritide (1/4–25%), calcium channel blockers (5/36–13.8%), intravenous phospodiesterase inhibitors (3/29–10.3%), antiarrhythmics (2/28–7.1%) and digoxin (5/70–7.1%). Differences between percent harmful reports were not statistically significant. Infants <1 year had the most reports (50%) ($p < 0.001$), with 45% in infants <6 months. The remainder of errors were evenly distributed among age groups. There was no significant difference in harmful events by age.
Conclusion: Infants <1 year are vulnerable to cardiovascular medication errors that reach the patient comprising 50% of all reports. Infants <6 months accounted for 45%. Harmful errors occurred with most cardiovascular classes. Most errors were related to administration of an improper dose despite a correctly written order, identifying an area in need of further research and intervention to decrease error.

VDD pacing in children and adolescents, a 4-year experience in Costa Rica

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VDD pacing in children combines physiological benefit of atrioventricular synchrony with the advantage of a single-lead system. This is a retrospective study to asses the results of the initial

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experience with the use of VDD pacemakers (PM) at the Hospital Nacional de Niños "Dr. Carlos Sáenz Herrera" of Costa Rica.
From July 2002 to August 2006, 55 patients received a PM in our hospital. 14 children had a VDD PM implanted and were retrospectively evaluated. PM implantations were done in the operating room under general anesthesia and fluoroscopy. The following parameters were evaluated: age, sex and etiology; clinical and electrocardiographic indications for PM implantation; venous access route, surgical technique and atrial sensing at the time of implantation; atrial sensing-ventricular pacing, symptoms and complications during follow-up; and final PM pacing mode.
VDD single lead pacing systems (Medtronic Sigma VDD 303 and Medtronic Kappa VDD 901 generators with Capsure VDD-2 5038 single pass lead) were implanted in 14 patients. The ages ranged from 5 years and 2 months to 16 years and 5 months (mean age 10.5 years), there were 4 boys (28.6%) and 10 girls (71.4%). All 14 cases had third-degree atrioventricular block (AVB) with normal sinus node function. In 9 patients (64.2%), the primary disease was congenital AVB. The rest developed complete AVB after heart surgery for congenital heart disease. Nine patients (64.2%) had an epicardial PM in VVIR mode before implantation of their VDD PM. The most commonly used venous access route was the right subclavian vein (57.1%) followed by a right cephalic vein puncture (28.5%). One patient required early repositioning of the lead because of lead displacement. Over the follow-up period of 1.5 to 52 months (mean 11 months), significant atrial undersensing did not occur and only one patient referred fatigability that improved with optimization of PM parameters. None had to be reoperated because of traction of the leads and all of them had 96 to 99.9% atrial sensing-ventricular pacing at the last follow-up visit.
VDD single lead pacing is an effective and safe method to maintain AV synchrony in children and adolescents with third degree AV block and normal sinus node function.

Home surveillance for hypoplastic left heart syndrome: validation of the experience at the Children's Hospital of Wisconsin

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Background: Despite recent advances in perioperative care for children with hypoplastic left heart syndrome (HLHS), interstage mortality continues to be a problem. The most successful intervention to date has been a Home Surveillance Program published by Milwaukee Children's Hospital. This study, in the Duke Pediatric Cardiology Clinic, is an attempt to validate the Milwaukee results in a single institutional cohort of patients with HLHS, with specific attention to feeding, growth and nursing parameters.
Methods: Infants were discharged home after their 1st stage palliation with a digital weight scale, pulse oximeter and a notebook for recording daily weights, feeding volume and oximetry readings. Detailed teaching was provided with specific call parameters in the event of abnormal home results. Patients were seen weekly/biweekly until the 2nd stage palliation by a dedicated Pediatric Cardiologist and Pediatric Cardiology Nurse. Retrospective analysis was performed on the infants followed in this Single Ventricle Clinic who had undergone the 2nd stage of surgical palliation since the start of the program.
Results: During the study period, there were many 1st stage palliative procedures performed at Duke University Medical Center. However, due to the tri-state referral area, only six of these patients were followed in the Duke Single Ventricle Clinic. Patients followed elsewhere were provided with similar equipment and teaching, but results were not comparable due to different follow-up regimens by the local Cardiologist. Of the six patients followed at Duke, there was no interstage mortality. At the time of 2nd surgery, half of the patients were at the 5th percentile for weight and half were below the 5th percentile, but all had maintained their growth trajectory. There were a total of 8 interstage admissions, of which only 2 were for cardiac reasons (both in the same patient). One patient was admitted at 5½ months of age for progressive cyanosis and underwent catheterization and subsequent semi-elective surgery. All others had elective 2nd stage surgery between 5–7 months of age. Although not documented by a formal survey, parents demonstrated benefit from the frequent monitoring program by having a greater understanding of the complexity and comfort with their child's heart disease.
Conclusion: Patients with HLHS between the 1st and 2nd surgical stages represent one of the most complicated patient groups in Pediatric Cardiology. Feeding issues are a common problem that can be successfully addressed through a team approach. This small cohort validates the published Milwaukee experience and demonstrates the importance of a hypervigilant team approach in the care of these patients.

Antenatal diagnosis in pulmonary atresia with intact ventricular septum: impact on early morbidity and mortality

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Background: Antenatal diagnosis of congenital heart disease with duct dependent systemic circulation or transposition of the great arteries has been shown to improve short term morbidity and mortality. Limited data exists for lesions with duct dependent pulmonary circulation. Pulmonary atresia with intact ventricular septum (PAIVS) is a lesion which historically has a poor long term outlook and one with potential for serious compromise in the neonatal period in the event of ductal closure.
Antenatal identification of the fetus with PAIVS should result in early initiation of prostaglandin to maintain ductal patency and present neonates in optimal condition for catheter or surgical intervention. This would be expected to improve long term neuro-developmental outcome and mortality.
Methods: A retrospective review of medical records, echocardiograms and catheters was performed for all children managed with PAIVS between 1990 and 2006.
Data analysis was performed with unpaired t-tests for non-parametric data, Chi squared test for categorical data and Kaplan-Meier survival curves.
Results: 93 newborns with PAIVS were identified over the period of review. 59 were local patients whilst 34 were referred from interstate or overseas.
38 (41%) patients with PAIVS were antenatally diagnosed. The mean follow up was 5.5 years (median 3.8 years, range 1 day–16.4 years). Overall survival was 79%, 76% and 76% at 1, 5 and 15 years respectively. In the antenatally diagnosed cohort survival was 66%, 66% and 66% versus 88%, 84% and 84% at 1, 5 and 15 years respectively in those postnatally diagnosed ($p = 0.03$)

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Statistically significant associations with antenatal diagnosis were higher saturation at initiation of prostaglandin therapy 82.2% vs 67.4% ($p < 0.0001$), earlier gestation at birth 37.0 vs 38.8 weeks ($p = 0.002$), the smallest ventricles ($p = 0.001$) and failure to achieve biventricular repair 26% vs 53% ($p = 0.011$). Features which did not reach statistical significance included birth weight ($p = 0.11$), acidosis at initiation of prostaglandin ($p = 0.31$), and tricuspid valve z score −1.5 vs −1.1 ($p = 0.26$).
Statistically significant associations with mortality included lower birth weight 2.59 vs 3.16 kg ($p = 0.004$), earlier gestation at birth 36.5 vs 38.5 weeks ($p = 0.003$), the smallest ventricles ($p = 0.01$), coronary sinusoids ($p = 0.001$), Ebstein's anomaly of the tricuspid valve ($p = 0.07$) and those not achieving a biventricular repair ($p = 0.001$). Features which did not reach statistical significance included oxygen saturation ($p = 0.16$) or presence of acidosis ($p = 0.28$) at initiation of prostaglandin therapy, tricuspid valve z score ($p = 0.20$), bidirectional cavopulmonary connection ($p = 0.34$) or Fontan circulation ($p = 0.10$).
Sub-group analysis for ventricular size confirmed no improvement in mortality in those antenatally diagnosed.
Conclusion: Antenatal diagnosis of PAIVS improves hypoxia prior to initiation of prostaglandin in the neonatal period. Our cohort showed a significantly greater mortality in those with an antenatal diagnosis due to identification of patients with the smallest ventricles and least chance of successful two ventricle repair. Our results confirm previously limited data that antenatal detection does not improve mortality in PAIVS.

A systematic initiative to reduce blood stream infections in a pediatric cardiac intensive care unit

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Background: Blood stream infections (BSI) cause morbidity and mortality, are costly, and are preventable. Children in dedicated cardiac ICUs (CICU) may be at greater risk for developing BSIs due to younger age, severity of illness, need for invasive surgical procedures and indwelling devices, and immunosuppression following cardiopulmonary bypass as compared to children in multidisciplinary ICUs. This study aimed to establish baseline rates for primary and CVC-associated BSIs in a large pediatric CICU, and to determine whether a systematic approach involving staff education, increased awareness, and practice change would decrease BSIs.
Methods: A primary blood stream infection was defined using CDC criteria for a laboratory-confirmed BSI. A CVC-associated BSI was defined as a laboratory-confirmed BSI in a patient with a central venous catheter used within the last 48 hours. A retrospective review conducted in July 2003 of all CICU admissions ($n = 1,106$) during calendar year 2002 at Children's Hospital Boston revealed 77 primary BSIs (11.6/1,000 patient-days). These concerning data stimulated the formation of a multidisciplinary Cardiovascular Program Nosocomial Infection Committee in October 2003. During a pre-implementation period (April 2004–December 2004), the Committee initiated prospective nosocomial infection surveillance in collaboration with the Infection Control Department. A pretest administered to CICU staff demonstrated knowledge deficits about nosocomial infection prevention, and educational tools were developed. During the implementation period (January 2005–March 2006), a comprehensive CVC-associated BSI prevention initiative was implemented including: establishment of a 0.75 FTE CICU-based infection control nurse position; mandatory education for physicians and nurses; establishment of a CVC insertion bundle including a kit for maximum sterile barrier precautions; implementation of CVC maintenance and access bundles; introduction of daily goals sheets emphasizing timely removal of CVCs; and participation in the Child Health Corporation of America's 2005 BSI collaborative. The Committee met monthly and implemented change in response to real-time BSI data, and provided monthly feedback to staff regarding current BSI rates and BSI prevention initiatives. Financial incentives to reduce CICU BSI rates were incorporated into the hospital's 2005 contract with a major third party payer. Rate ratios and 95% confidence intervals were generated to compare post-implementation BSI rates (April 2006–September 2006) to pre-implementation rates (April 2004–December 2004).
Results: The pre-implementation primary BSI rate was 6.5/1,000 patient-days, and 1.8/1,000 patient-days during the post-implementation period; rate ratio = 0.28 (95% CI 0.12, 0.63; $p = 0.002$). The pre-intervention CVC-associated BSI rate was 8.1/1,000 catheter-days, and 2.6/1,000 catheter-days during the post-intervention period; rate ratio = 0.32 (95% CI 0.11, 0.89; $p = 0.043$). Our post-intervention CVC-associated BSI rate is favorable when benchmarked against the CDC's National Nosocomial Infection Surveillance System pediatric ICU pooled mean rate of 6.6/1,000 catheter-days.
Conclusions: A systematic, multidisciplinary approach resulted in a significant reduction in primary and CVC-associated BSI rates in our pediatric CICU. We have expanded the initiative to include other ICUs within our institution. Ongoing studies will identify risk factors for BSIs in the CICU and outcomes for infected patients.

DiGeorge syndrome is not associated with heterotaxy syndrome

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Background: DiGeorge syndrome has been shown to be associated with cardiac conotruncal malformations. Patients with cardiac heterotaxy may have similar conotruncal defects and a DiGeorge probe may be part of their routine cardiac work-up though there is no data to show an association. The goal of this study was to determine if there was an association with patients with heterotaxy syndrome and DiGeorge syndrome.
Methods: Retrospective review of infants with diagnosis of heterotaxy syndrome was performed. Heterotaxy syndrome was defined if patients had complex cardiac malformations as well as abnormal abdominal situs findings. Fluorescence in situ hybridization using the DiGeorge syndrome chromosome region (LSI DiGeorge/Velocardiofacial syndrome, TUPLE1; Vysis) was used to detect deletion of the locus at band 22q11.2. Rule of threes was used to establish confidence intervals and Chi square was used to compare results with literature data.
Results: Searching echocardiographic and radiological data base from 1/1/03 to 10/1/06, 19 patients met criteria for heterotaxy syndrome. Three patients had no testing for DiGeorge syndrome. Of the remaining sixteen patients that were tested for DiGeorge syndrome, none were positive. 95th % percent confidence interval was 0 18.8%. Patients in this review had significantly less association

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with DiGeorge syndrome compared to patients with conotruncal abnormalities testing positive for DiGeorge syndrome in literature (Goldmutz E et al. Frequency of 22q11 Deletions in Patients with Conotruncal Defects. J Am Coll Cardiol 1998;32:492–498), Chi Square = 4.78, p = 0.03.
Conclusion: Patients with heterotaxy syndrome did not have an additional diagnosis of DiGeorge syndrome. DiGeorge probe analysis may not necessarily be needed in this patient population compared to patients with conotruncal defects.

Precise control of flow with an adjustable systemic-pulmonary artery shunt

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Objective: Control of pulmonary blood flow remains problematic after complex neonatal single ventricle reconstruction. Our laboratory has addressed this issue by creating a practical adjustable systemic-pulmonary artery shunt (AS).
Methods: An adjustable resistor was made using injection-molded acetal which snaps around a standard 3.5 mm polytetrafluoro-ethylene (PTFE) graft. A screw-plunger mechanism, driven by a stepper motor, was used to control flow. The AS was placed on a benchtop steady flow circuit. The relationship between flow and resistor setting was determined. The rotational displacement of the motor required to achieve a given flow was programmed into the motor's computer controller. The controller was then programmed to adjust the stepper motor to achieve target flow rates from 300 ml/min to 600 ml/min (inclusive) in 50 ml/min increments. The order of target flow rates was chosen at random (n = 10 determinations of each target flow rate).
Results: Actual flow rates differed from target flow rates by 1.60% ± 1.77% (mean ± standard deviation). Actual flow was closer to target flow at higher flow rates (.34% ± .20% difference at 600 ml/min target flow rate) than at lower flow rates (4.40% ± 1.46% difference at 300 ml/min target flow rate).
Conclusion: The AS controls flow through a 3.5 mm PTFE graft with excellent, predictable accuracy. The AS could allow clinicians to control pulmonary blood flow independent of factors affecting pulmonary and systemic vascular resistance. Independent control of pulmonary blood flow may improve survival after complex neonatal single ventricle reconstruction.

Implementation of a safety checklist at the time of nursing shift change in a pediatric Cardiovascular Intensive Care Unit

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Background: Medical errors constitute a major source of patient mortality and morbidity. In the setting of a pediatric cardiac intensive care unit, these errors can be magnified by the complexity of care required for these patients. We identified one source of errors to be at the time of nursing shift change. The verbal checkout was noted to result in the loss of vital pieces of information from shift to shift and led to sentinel events in our unit. We therefore introduced a systems-based format for checkout at shift change along with a checklist with important patient safety questions to be answered and signed off by both the outgoing and incoming nurse for each patient.
Methods: A pre and post survey was conducted 18 months a part to learn about nursing staff perceptions about the issues addressed in the patient safety checklist. The survey tool had 16 questions rated on a 5-point response scale ranging from *Never* to *Always*. A baseline survey was conducted and the changes were implemented in April 2005. The survey was repeated in October 2006. Forty two RN's on staff participated in the 2 surveys. The results were analyzed using the Fisher's exact test.
Results: The implementation of the new bedside report and checklist have resulted in a statistically significant improvement in the perceptions of the nursing staff as to the availability of bag and mask at the bedside ($p < 0.02$) and availability of working suction apparatus ($p < 0.0001$). There has been a major improvement in finding the correct IV fluids ($p < 0.0001$) running at the correct rate ($p < 0.0001$). It has also resulted in improved compliance with finding ID bands on patients ($p < 0.0001$). The nurses report that they are now more likely to fill out a medication error report if they find fluids or medications given at the wrong rate or wrong time ($p = 0.0001$). Finally, the nurses now increasingly feel that the adoption of this checklist helps them in maintaining a safer patient care environment ($p < 0.0001$).
Conclusions: The implementation of a systems-based bedside report and a checklist at the time of nursing shift change results in perceived improvements in patient safety environment. The widespread use of such checklists has the potential of significantly decreasing medical errors.

Utility of computed tomographic angiography in the preoperative planning for congenital heart surgery

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Background: Computed tomographic angiography (CTA) is an emerging modality for cardiovascular diagnosis in patients with structural heart disease. Its utility in preoperative planning for patients undergoing repair of congenital heart disease has not been well described.
Methods: Over a 16-month period, we prospectively identified 33 patients (children and adults) who were being evaluated for congenital cardiac surgery and in whom a CTA was obtained for diagnostic purposes prior to the planned surgery. Operations were classified according to the principal anatomic site of repair: aortic arch (n = 15), right ventricle to pulmonary artery conduits (n = 7), pulmonary arteries (n = 3), pulmonary veins, (n = 4), trachea/airway (n = 2), and other (n = 2). After the operation, the surgeon was polled to assess the study's overall utility in operative planning. Specifically, the surgeon was asked to grade the usefulness of the study on a 1 (not helpful) to 5 (essential) scale and comment on whether the CTA dictated the need for peripheral (femoral) cannulation and/or obviated the need for a cardiac catheterization. Comparisons among groups were assessed using a Fisher's Exact test.
Findings: Most studies (31/33; 94%) were classified as either very helpful or essential to preoperative planning. In the remaining patients, an echocardiogram provided sufficient anatomic information. Overall, CTA's proved to be equally useful in the pre-operative planning among diagnostic groups. However, they proved to be consistently useful to the surgeon for procedures involving the aorta (14/15, 93%) or the pulmonary veins (4/4, 100%). CTA

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obviated the need for pre-operative cardiac catheterizations in 14 patients (42%). In addition, CTA was important in determining the need for peripheral cannulation in many patients, but especially in those undergoing re-operations or conduit revisions compared to other diagnostic groups (6/7; 86%, $p = 0.02$).
Conclusions: CTA's were found to be very useful in the pre-operative planning of virtually all patients who were undergoing repair of congenital heart malformations at our institution, regardless of diagnosis. Specifically, the studies were essential in select populations, such as those undergoing repair of aortic arch or pulmonary vein anomalies, and helped determine cannulation sites in patients undergoing conduit replacement. In addition, CTA's reduced the need for invasive imaging in our patients.

The effect of isoflurane in a chronic hypoxia model of developing brain

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Introduction: The N-methyl-D-aspartic acid (NMDA) receptor subunits undergo age related changes during human hippocampal development. (1) This developmental effect is dependent on the partial pressure of oxygen and allows the fetus to tolerate hypoxic conditions in utero. (2) The effect of chronic hypoxia, as in children with cyanotic congenital heart disease, on NMDA 2B (NR2B) receptor subunit composition has not been evaluated. *We hypothesized that chronic hypoxia will increase the NR2B subunit composition and isoflurane will decrease the NR2B subunit composition.*
Methods: Hypoxia (10% oxygen) was administered via a hypoxic chamber and to PND2 rat pups and maintained for 2 days (PND4). At PND4, isoflurane was administered for 5 hours. After exposure to isoflurane, the animals were immediately sacrificed. NMDA receptor subunit composition of the isolated hippocampal region was determined using western blot analysis.
Results: The effect of *chronic hypoxia* exposure (2 day exposure to 10% oxygen) in a PND 4 rat pup demonstrates an increase in the NR2B subunit composition compared to a *normoxic* control PND4 rat pup (Fig. 1). Isoflurane, decreases the NR2B subunit composition to that of a *normoxic* control PND4 rat pup (Fig. 1).

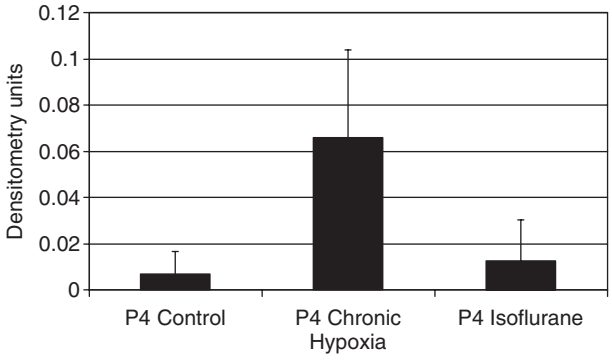

Figure 1. Data shows a change in NR2B subunit composition with respect to chronic hypoxia (2 day exposure to 10% oxygen). The data also demonstrates a NR2B receptor subunit composition change in response to a 5 hour exposure to 1.5% isoflurane in a rat pup exposed to chronic hypoxia (2 day exposure to 10% oxygen). The data is compared to a normoxic control (4 day exposure to room air, 21% oxygen). Data expressed in densitometry units as a ratio to ß-actin.

Conclusion: The response of the developing brain to *chronic* hypoxia is an increase in the NR2B subunit composition. This increase in the NR2B subunit composition is decreased by a prolonged exposure to isoflurane. The NR2B subunit composition is essential for synaptic plasticity. It is plausible that *chronic hypoxia* is protective against the neurodegenerative effects of isoflurane in the developing brain. Thus, the implications of isoflurane on neuronal cell death in the presence of chronic hypoxia will demand further investigation.

Nuclear magnetic resonance spectroscopy in a chronic hypoxia model of developing brain

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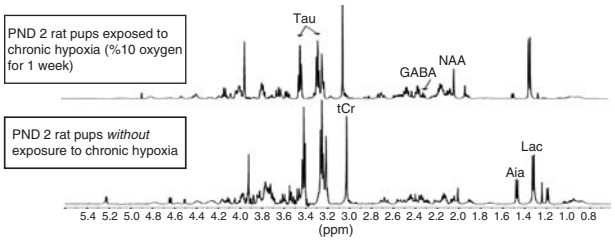
Introduction: Chronic hypoxia, as in children with congenital heart disease, may predispose these children to poor neurological outcome after surgical repair for congenital heart disease. Nuclear magnetic resonance spectroscopy (NMRS) of the brain has demonstrated no difference in low versus high hematocrit management during cardiopulmonary bypass in a limited number of patients. Concerns for the detrimental effects of chronic hypoxia on the developing brain have prompted early surgical repair for cyanotic congenital heart lesions. *We attempted to evaluate this concern by using NMR spectroscopy in a neonatal cyanotic rat pup model.*
Methods: Hypoxia (10% oxygen) was administered via a hypoxic chamber and to PND2 rat pups and maintained for 7 days (PND9). At PND9, the rat pups were decapitated and the brain tissue was immediately frozen and later evaluated using NMR spectroscopy for metabolic changes. These changes were compared to PND2 rat pups maintained in normoxic conditions for 7 days (PND9).
Results: The metabolic effects of chronic hypoxia as determined by NMRS in the developing brain demonstrate an increase in inositol, glutamate, GABA, lactate and N-acetyl-aspartate with a decrease in the level of taurine (Fig. 1).



Conclusions: These metabolic changes demonstrate a continued propensity towards normal myelinization and neuronal growth as expected in normal development but a decrease in astrocyte and glial development. These changes further demonstrate an increase in brain edema as reflected by the decreased taurine with exposure to chronic hypoxia. This would support normal brain development in children with cyanotic congenital heart disease but a propensity toward neurologic injury for lack of neurologic supporting structures.

Congenital heart surgery without the routine placement of temporary pacing wires

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Objective: Temporary pacing wires have been associated with serious postoperative complications. Recommendations for the routine use of cardiac pacing wires after open hearts surgery are decades old and may not reflect current surgical outcomes.
Methods: The electronic web-enabled records of all pts undergoing congenital heart surgery (CHS) from 1/1/02 through 12/31/05 were reviewed. Pts undergoing pacemaker implantation as a primary procedure or PDA ligation were excluded.
Results: There were 1193 CHS performed (1041 with cardiopulmonary bypass). Median age was 167 days (range 0 days to 54 yrs), weight 6.1 kg (range 1–114 kg). Mortality to discharge was 2.5% and median postoperative stay was 7 days. No deaths were attributed to arrhythmias. There were temporary pacing wires placed 14 times (1.3%). Indications for placement of temporary wires included preoperative diagnosis of sinus node dysfunction (SND) in 4, intraoperative evidence of SND in 4, high degree AV block in 4 and atrial flutter control 2. Four of these pts (0.4%) eventually underwent permanent pacemaker implantation, 2 for persistent sinus node dysfunction and 2 for persistent high degree heart block. Implantation occurred at an average of 6 days after their primary procedure (range 1–12 days). Average postoperative stay of these pts was 21.8 days. Postoperative junctional ectopic tachycardia requiring therapy occurred in 10 pts (1%). All recovered without incident and none required pacing.
Conclusions: Because of the diminished risks of unexpected postoperative arrhythmias in the current era, the routine placement of epicardial pacing wires may no longer be warranted. Meticulous surgery aimed at the preservation of the SAN, AVN and myocardial pump function alleviates the necessity for routine placement of temporary pacing wires after CHS. The need for temporary pacing wires can be predicted preoperatively or intraoperatively.

Enteral feeding and close follow-up: a key to early discharge and low incidence of NEC in HLHS patients

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Objectives: As survival increases after palliation for HLHS-syndrome, increasing attention has been focused on decreasing perioperative morbidity. Feeding problems represents a significant cause of morbidity in these patients. We have adopted a strategy of aggressive perioperative enteral feeding in all our patients with HLHS or related malformations, and a very tight follow-up by a nurse coordinator. We would like to present the preliminary results from this approach.
Methods: We reviewed the medical records of the entire experience of 58 consecutive neonates at a single institution undergoing palliation for HLHS or related malformations. In all patients, we adopted a strategy of aggressive perioperative enteral feeding regardless of the presence of umbilical lines, endotracheal tube, inotropic support or prostaglandin infusion. The survivors were discharged home with NG-tubes, if necessary, after parental counselling. Guidelines for feeding were given both written, and orally by the nurse coordinator to all persons involved. Weight control weekly was performed until stage 2.
Results: Preoperative oral (26/58) or NG (27/58) feeding was established in 91% (53/58) of patients. Survival to discharge was 69% (40/58). In survivors, chest tubes were required for 2.5 median days (range = 1–8 days) and enteral feeding was initiated or resumed at a median of 1 days (range = 0–2 days) after Stage 1. Chylothorax occurred in 3.4% (2/58). NEC occurred in 1.7% (1/58). Gastrostomy was not used in any patient. Survivors were discharged home on oral (4/40), combined oral/NG (10/55) or NG (26/40) feeding. Weight at surgery was 3470 grams, and at discharge 3775 grams. LOS was median 15 days. Interstage mortality occurred in three patients. Weight at surgery at stage 2 was 6200 grams. A total of three patients have required gastrostomy after stage 3.
Conclusions: A strategy of aggressive peri- and postoperative enteral feeding in patients undergoing staged repair of HLHS and related malformations appears to be associated with low incidence of NEC and feeding difficulties necessitating PEG. The role of supporting and teaching parents is thought to be of fundamental importance. The role of a nurse coordinator seems to be mandatory in this respect. The LOS and the low incidence of NEC in this material supports this approach.

Developmental outcome after three stage palliation for hypoplastic left heart syndrome

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Background: Treatment of patients with hypoplastic left heart syndrome (HLHS) implies a significant risk for psychomotor retardation.
Patients and Methods: 50 patients born between 1996 and 2002 were evaluated after their 3 staged palliation (Norwood-, Hemifontan-, Fontan operation) with the help of standardized tests for their cognitive (K-ABC) and visual motor integration (VMI) developmental stage and were compared with a control group of healthy children who were matched for their age and gender. A parent questionnaire was used to get information about the quality of life (Kindl test) and behaviour problems (CBCL).
A number of possible influence factors, for example the duration of the total cardiac arrest time, the pulmonary bypass time were checked of significant correlations with the test data.
Results: The scores of the intelligence quotient from the "scale of intellectual abilities" and "scale of proficiency" (K-ABC) were with 78.4 ± 14.7 respectively 76.1 ± 18.1 significantly ($p < 0.01$) lower as the control group (103.9 ± 9.1; 104.3 ± 11.4).
The patients showed also significantly lower scores in the area of visual motor integration (78.2 ± 23.0, $p < 0.01$; control group 105.3 ± 10.5). Particularly the influence factor cardiac arrest time showed a negative significance for both areas.
Both groups had equal good quality of life and no behaviour problems.
Conclusion: The children with HLHS successfully operated in the first years of our experience show after their stage palliation a worse cognitive and visual motor integration developmental stage than the control group. However, they have an equally good quality of life.
When we looked closer at the HLHS group all of the patients operated after 1999 had continuous cerebral perfusion during their Norwood operation with remarkable lower cardiac arrest time or no cardiac arrest time at all. This group is doing better in their cognitive (K-ABC) developmental stage, so far without statistical significance and remarkable better in their visual motor integration (VMI) developmental stage.

Does the preoperative use of inderal in patients with tetralogy of Fallot effect postoperative outcomes?

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Background: Beta-blockade is used in some patients with tetralogy of Fallot (TOF) as prophylaxis against hypercyanotic spells prior to surgical repair. Beta-blocker therapy has been linked to catecholamine resistance in postoperative patients. The aim of this study was to determine if preoperative inderal therapy had a deleterious effect on postoperative variables in patients with TOF.
Methods: Over an eight year period, 97 patients with TOF underwent complete repair. Patients with pulmonary atresia or absent pulmonary valve were excluded. Preoperative, intraoperative and postoperative variables were retrospectively reviewed. The decision for initiating preoperative inderal therapy was at the discretion of the primary cardiologist therefore no uniform criteria was utilized.
Results: See table

	Inderal (n = 32)	Non inderal (n = 65)	P value
Preoperative Variables			
Age (months)	5.0 (0.5–59.6)	6.5 (0.2–124.6)	0.08
Weight (kilograms)	5.8 (2.7–16.8)	6.8 (2.5–26.7)	0.35
Male:Female	18:14	34:31	0.88
Intraoperative variables			
CPB Time (minutes)	143 ± 24	152 ± 35	0.21
Cross clamp time (minutes)	75 ± 18	78 ± 20	0.41
Transannular patch	23 (72%)	37 (57%)	0.23
Postoperative variables			
Inotrope score (arrival)	8 (2–18)	5 (2–18)	0.10
Inotrope score (4 hours)	8 (3–18)	7 (1–18)	0.33
Inotrope score (8 hours)	9 (2–18)	8 (1–18)	0.11
Inotrope score (12 hours)	9 (1–30)	8 (0–22)	0.25
Inotrope score (24 hours)	8 (0–24)	5 (0–18)	0.05
Inotrope score (48 hours)	8 (0–13)	3 (0–18)	0.05
Pacing	5 (16%)	2 (3%)	0.04
Mechanical ventilation (hours)	54 ± 37	51 ± 49	0.68
Total inotrope (hours)	69 ± 36	66 ± 48	0.70
Total ICU stay (hours)	91 ± 42	98 ± 76	0.60
Total hospital stay (days)	8 ± 3	8 ± 5	0.72

Conclusion: Preoperative inderal therapy does not adversely affect postoperative variables in tetralogy of Fallot patients undergoing complete repair. There is a trend for increased inotrope score at 24 and 48 hrs postoperatively and an increase in the need for temporary pacing in the patients treated preoperatively with inderal, however this does not correspond with a longer time on mechanical ventilation, total time on inotropic medications, ICU or hospital stay. The efficacy of inderal therapy in prevention of preoperative hypercyanotic spells was not addressed by this study.

Coagulation factor abnormalities in neonates with single ventricle physiology precede surgery

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Background: Altered levels of coagulation factors have been reported in patients with functionally single ventricle before and following the second and third stage of single ventricle repair. The aim of the study is to test the hypothesis that coagulation factor abnormalities are present in neonatal age, before the first stage of the palliative surgery and are predispositional of hemocoagulation state disturbances.
Methods: Neonates with single ventricle physiology admitted to Children's Cardio-center in Slovakia were included into the study. Concentration of liver enzymes, serum albumin and complete blood count were measured and set as baseline characteristics of study population. Concentration of factor II, V, VII, VIII, Protein C, Protein S and Antithrombin were measured before the surgical procedure and evaluated as possible predictors of hemocoagulation state disturbances. Normal age matched reference intervals were determined as control values. Clinical signs (bleeding, thrombosis) and laboratory values (PT ratio, APTT ratio, D Dimmer, Fibrin Degradation Products) were analyzed as signs hemocoagulation disturbance.
Results: The study population consisted of 14 patients before the first stage of single ventricle palliative surgery. The age range was 1–30 days (median 7 days). No clinical signs of thrombosis or bleeding occurred before the surgery. Hematological variables, liver enzymes and concentration of serum albumin were within the normal range in all patients. Concentration of protein S ($p < 0.01$), protein C ($p < 0.03$), factor II ($p < 0001$), VII ($p < 0.01$) VIII ($p < 0.02$) and Antithrombin concentration ($p < 0.01$) were under the low limit for age, respectively. PT ratio ($p < 0.001$) and APTT ratio ($p < 0.001$) were prolonged in the study population.
Conclusions: Altered levels of both pro- and anticoagulation factors are evident in neonates before the course of staged single ventricle repair. Further study is required to determine, whether the reported abnormalities are predictive of coagulation abnormalities during the second and third stage of single ventricle repair.

High fidelity simulation training: improvement of self confidence of cardiac center nurses in crisis situations

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Introduction: Nationwide nursing shortages and staff retention is a big challenge in high stress patient care areas such as the cardiac intensive care unit (CICU). High-fidelity simulation training for pediatric cardiac nurses has recently been developed to improve training efficacy.
Methods: A 5-day high-fidelity simulation training session was conducted using 7 trained facilitators for cardiac center nurses. Primary session objectives included basic and advanced pediatric life support skills such as bag-valve-mask ventilation, chest compression, defibrillation, medication preparation and preparation for pericardial drainage; all required cardiac center nursing skills. Each session was 30–40 minutes, with 4–5 participants and 1 facilitator. All facilitators were trained for 2 hours by the Simulation Center Clinical Educator prior to the training session on the mechanics and function of the high fidelity simulator and simulation software.
Pre-training and post-training self assessment questionnaires assessed participant confidence level and both technical and

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non-technical skill improvement. An anonymous 38-item questionnaire with Likert scale (0–3) was analyzed. Descriptive and t statistics were used with two tails, alpha = 0.05. Median, mean, and standard deviation (SD) were reported when appropriate.
Results: 163 cardiac center nurses completed the training session and questionnaire. The majority (83/163:51%) had experience in pediatric *cardiac* nursing for <3 years (median 2 year, mean 4.7 year, SD 5.4). The majority (89/163: 55%) also had experience in nursing for <6 years. Participants evaluated the simulation training as effective in technical skills (mean 2.25, SD 0.70), and non-technical skills (mean 2.35, SD 0.82). At baseline prior to training, nurses with cardiac experience <3 years had significantly less self-confidence compared to those with cardiac experience >=3 years (Technical skills: mean 1.81 vs. 2.43, $p < 0.0001$; Non-technical skills: mean 2.10 vs. 2.75, $p < 0.0001$). Both groups evaluated the simulation training as effective in Technical (mean 2.16 vs. 2.34, $p = 0.10$), and Non-technical (mean 2.30 vs. 2.40, $p = 0.44$) skills.
Conclusion: Nurses with cardiac experience <3 years have less self-confidence in Technical and Non-technical skills in basic life and advanced pediatric life support skills required in the cardiac center. However, regardless of the experience, simulation training was effective to improve self-confidence in Technical and Non-technical skills.

Current practice patterns with respect to milrinone use in pediatric cardiac surgery patients: a Canadian survey

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Background: Milrinone is frequently used in the postoperative care of pediatric cardiac surgery patients for prevention and treatment of low cardiac output syndrome (LCOS). It is eliminated by the kidneys and is known to have a large volume of distribution. As such, published clinical studies recommend administering a bolus dose followed by a continuous infusion. Our hypothesis is that actual practice of milrinone prescription differs from published recommendations, and that the bolus dose is frequently omitted because of fear of hypotension. The objective of this study was to determine the actual practice patterns of milrinone use in pediatric cardiac surgery patients in Canada.
Methods: A questionnaire survey was sent to all intensivists, cardiologists, anesthesiologists and cardiac surgeons working in seven pediatric university-affiliated hospitals in Canada (n = 227). The survey was first sent in February 2006 with two subsequent reminders.
Results: The response rate was 67%. 56% (85/152) of physicians who returned the questionnaire are directly involved in the postoperative care of pediatric cardiac surgery patients; these 85 questionnaires were analyzed. 73.5% report using milrinone in more than half of their patients while 9.6% use it in all patients. Among respondents who administer milrinone in the operating room, 78.9% give a bolus followed by an infusion. In most instances (93.9%), it is given during cardiopulmonary bypass at a dose ranging from 25 to 100 μg/kg (25 (10.3%), 50 (48.3%), 75 (13.8%), 100 (24.1%), other (3.4%)) followed by an infusion varying from 0.25–0.75 μg/kg/min. Eleven different combinations of bolus and infusion doses were reported. Among respondents who initiate milrinone postoperatively in the ICU, 37% give a bolus dose before starting an infusion when milrinone is given for the prevention of LCOS and 50.9% do so for the treatment of LCOS. When a bolus is given, the most frequently used dose is 50 μg/kg followed by a infusion varying from 0.25 to 0.99 μg/kg/min. Eight different combinations of bolus and infusion doses were reported. Among respondents who start a milrinone infusion without a bolus dose, the infusion is most frequently started at 0.5 μg/kg/min (51.7% and 50% for prevention and treatment of LCOS, respectively). The most common reasons cited for not administering a bolus dose of milrinone were the presence of hypotension, concern about hypotension and intention to introduce milrinone gradually.
Conclusion: Significant variability exists among milrinone dosing regimens currently used in the postoperative care of pediatric cardiac surgery patients in Canada, and concern about hypotension prevents many physicians from administering a bolus dose. The omission of a bolus dose may result in sub-therapeutic milrinone concentrations for a number of hours following initiation of the drug. This may have important clinical consequences in patients at risk of, or with, LCOS. Administration of a bolus dose over a longer period of time may decrease the likelihood of hypotension and overcome physician reluctance to include a bolus dose. Milrinone therapeutic blood monitoring with dosing individualization should be considered in this specific population where, in addition to variable dosing regimens, many other factors affect milrinone kinetics.

High near infra-red spectroscopy: twice as many stickers, twice as many dollars; but how much more information?

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Background: Neurological impairment is the leading morbidity in pediatric cardiac surgery, with survivors manifesting highly prevalent patterns. Near infrared spectroscopy (NIRS) is increasingly used to identify and limit periods of potential mismatch between cerebral oxygen supply and demand, thought to be responsible for a component of the intraoperative neurological injury. One study demonstrated a 73% reduction in adverse neurological events following adoption of a neuromonitoring technique involving NIRS. However, design varies in that some studies have used a single probe placed centrally, to one side or bilateral probes. Thus far, three authors have published case reports indicating asymmetries. Two larger studies examined the data obtained from bilateral measurements. One was conducted in the context of neonatal arch repair; the other demonstrated that in only 8% of all asymmetric measures was the lower value significantly lower than baseline values. No study has approached the issue of overperfusion, nor attempted a cost-effectiveness analysis of the second probe.
Objective: We set out to examine the data from 65 consecutive pediatric cardiac surgery cases requiring cardiopulmonary bypass, to determine if having a second probe added significantly to information gathered intraoperatively. Secondary analysis examined the cost-effectiveness of adding the second probe.
Methods: A prospective cohort of 65 patients undergoing pediatric cardiac surgery involving cardiopulmonary bypass underwent standard intraoperative monitoring, with bilateral NIRS cerebral monitoring.
Demographic data was collected from the chart at the time of analysis.
NIRS values were deemed significant if they deviated from baseline values by 20% or more in either direction (standard in the

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literature). Bilateral information was declared discrepant if there was an unduplicated significant unilateral deviation from baseline. Frequencies of discrepancies were tabulated by case for either direction and analyzed separately. As 2 minutes was felt to represent a reasonable minimum time period in which permanent neurological injury could be sustained, further analyses considered only those discrepancies lasting 2 minutes or longer. Finally, a cost analysis was performed based on $72.80 CA/sensor.
Results: The cohort included 41 males and 24 females; 25 had non-cyanotic pathologies, 40 had cyanotic pathologies. Increases: Over the 65 cases, 144 important discrepancies occurred (average 2.2/case). Discrepancies occurred more often amongst cyanotic patients (0.79%, $p < 0.0001$) and males (72%, $p < 0.0001$). Decreases: Over the 65 cases, 93 important discrepancies occurred (average 1.4/case).
Discrepancies occurred more often amongst cyanotic patients (77%, $p < 0.0001$) and males (74%, $p = 0.01$). Cost Analysis: 26% of deviations were discrepant; thus bilateral monitoring accounts for 11% of the reduction in risk of adverse neurological events. Bilateral monitoring costs $19.97 CA/discrepancy captured.
Conclusion: A second probe offers significantly more (33%) information, particularly in cyanotic and male patients. 83% of patients manifest important discrepancies. For every extra event appreciated, there is an associated cost of $20 CA. Further study is needed to demonstrate that these NIRS events can be routinely resolved, and that this benefits patient outcome.

Physiological responses to exercise in children after Fontan operation, in the supine, semi recumbent and upright positions

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Objective: This study sought to measure hemodynamic responses to exercise in children and adolescents who underwent modified Fontan operation, during maximal and sub maximal exercise testing on a stationary bike.
Background and hypothesis: Several studies of the cardio respiratory response after Fontan operation have demonstrated subnormal/abnormal aerobic capacity and hemodynamic responses to exercise. Most of these studies have been conducted in older children during maximal exercise on a treadmill. There is no systematic data for cardio respiratory responses in Fontan patients during maximal and sub maximal exercises in supine, semi-recumbent and upright positions on a stationary bike. "*We hypothesize that in Fontan patients, the cardiorespiratory responses to exercise would be different in the supine, semi recumbent and upright positions on a stationary bike during constant work rate exercise. Specifically, we hypothesize that the time to attain steady state for VO_2 (oxygen consumption) and heart rate would be different in each position*".
Methods: The study included 10 randomly selected patients (aged 8-18 years) who underwent a modified Fontan operation between 1987 and 1999 at Rainbow babies and Children Hospital. Nine were in New York Heart Association (NYHA) functional class I and one patient was in class II. Each patient had a complete physical examination the day of testing. A transthoracic echocardiogram was performed at rest and during exercise. Gas exchange was measured using a breath-by-breath technique. Continuous 12 lead EKG monitoring, heart rate, oxygen saturation and blood pressure was performed. Peak exercise testing was performed in supine position on a previously calibrated stationary cycle ergometer and a 2 minute incremental cycle exercise protocol was adopted. Submaximal testing, defined as "exercise at a work rate equivalent to 60 to 70 percent of VO_2 max", was performed on a different day during supine, semi-recumbent and in upright positions.
Results: For the maximum exercise testing: VO_2 max ranged from 20 to 25.7 ml/kg/min (mean 21.7 ± 3.77 standard deviation), heart rate from 130 to 156 (137.8 ± 20.09), anaerobic threshold from 0.297 to 0.966 lit/min (0.55 ± 0.20), O_2-pulse 3.5 to 12.7 ml/beat (7.63 ± 3.20), respiratory quotient from 0.94 to 1.18 (1.05 ± 0.07) and METS from 3.42 to 7.34 (6.199 ± 1.07). There were no significant arrhythmias noted. Seven patients had mild atrioventricular valve regurgitation by transthoracic echocardiogram at rest and did not progress during peak or sub maximal exercises. For the submaximal exercise testing: there was no statistically significant difference in the VO_2, heart rate and O_2 pulse.
Conclusion: There was no statistically significant difference in the VO_2, heart rate and O_2 pulse in supine, semirecumbent and upright positions during submaximal testing. The maximal and submaximal exercise testing can be performed safely in Fontan patients.

Improved results with intraoperative extubation and minimally invasive techniques in infants with tetralogy of Fallot

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Background: Tetralogy of Fallot (TOF) is the 4th most frequent diagnosis reported to the Society of Thoracic Surgeons' (STS) congenital database, yet the optimal surgical approach for this lesion continues to evolve. Recent trends favor early intervention and primary correction, but suboptimal results in infants under six months of age have been reported. Here we evaluate the preliminary results of a minimally invasive surgical approach in this population. Our program combines early primary intervention with limited skin incisions and cardioscopy, along with intraoperative extubation, whenever feasible, for all patients regardless of age.
Methods: Our STS Congenital Database was queried for any patient who underwent complete repair of TOF between October, 2002 and February, 2007. A total of 14 patients were identified and these records were reviewed. Small surgical incisions were facilitated by cardioscopic visualization, and combined with generous local anesthesia to promote patient comfort. Intraoperative respiratory management included moderate doses of a short acting narcotic (fentanyl) and inhalational anesthetic (sevofluorane). Following extubation in the OR, the team allowed up to 30 minutes for the child to slowly awaken. Once breathing on their own, additional narcotics were titrated to respiratory rate. Upon arrival in intensive care, oxygen was delivered via hood and age appropriate pulmonary hygiene measures instituted.
Results: The STS classifications for these TOF procedures were: no ventriculotomy ($n = 9$), ventriculotomy with ($n = 4$) and without ($n = 1$) transannular patch, and additional concomittant repairs that included ASD closure ($n = 9$) and tricuspid valvuloplasty ($n = 3$). The mean age was 119 days (range 42–317), mean weight was 4.7 kg (range 2.8–6.98) with 10/14 patients under 6 months of age and weighing less than 5 kg. Mean time on cardiopulmonary bypass was 136 minutes (range 85–215), cross-clamp time was 64 minutes (range 38–117), with no use of deep

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hypothermic circulatory arrest or delayed sternal closure. There was no operative mortality, unplanned reoperation, neurologic complications, or sternal infection. Eleven of 13 patients (84.6%) were extubated intraoperatively (1 patient was chronically ventilated via tracheostomy). One patient required reintubation in the operating room, due to partial airway obstruction in a patient with Trisomy 21. Chylothorax developed in one patient, who also had Trisomy 21. Transient arrhythmias were noted in 3 patients, including junctional ectopic tachycardia (n = 2) and complete heart block (n = 1) and all resolved within 72 hours. Initial lactate levels averaged 2.8, and ranged from 1.7 to 4.0. Less than half of the patients (n = 6) required 2 or more inotropes beyond 24 hours. Median postoperative length of stay was 6 days.

Conclusions: Intraoperative extubation and minimally invasive techniques are feasible in even small infants with TOF, and contribute to excellent short-term outcomes. Avoidance of positive pressure ventilation and surgical trauma are particularly beneficial in this population to reduce right heart workload in the perioperative period. These initial results are encouraging and favor the pursuit of minimally invasive methodologies whenever feasible, regardless of patient age or size.

Extracorporeal membrane oxygenation (ECMO) versus ventricular assist devices (VADs) as a bridge to cardiac transplantation in children: a theoretical cost analysis

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Published indications for extracorporeal support now include sepsis, post in-hospital cardiorespiratory arrest and 'bridging to cardiac transplantation'. In the United Kingdom, ECMO access is limited to four centres, placing added pressure upon the optimal timing of referral, transportation and healthcare resource allocation. An expanding patient cohort and unpredictable supply of suitable donor hearts may necessitate protracted ECMO runs that risk complications and threaten outcome. Ventricular assist devices provide an alternative supportive strategy, available for patients with end stage, single organ cardiac failure. They offer extended support, fewer complications and an improved intact survival benefit. This is currently available in only two UK centres; experience is limited and capital outlay costs are high.

We compare the financial implications of both systems using hypothetical clinical scenarios based on our experience from November 2005 to date.

Using figures for the financial year of April 2005–March 2006, we calculated the costs of ECMO against BIVAD insertion with the Berlin Heart Paediatric ExCor device for 10 kg and 35 kg patients. Fixed, universal costs of monitoring and general hospital equipment were not included. Our scenarios are complication free, without mortality. Escalating costs are expressed graphically. To ascertain survival on ECMO, we consulted the ELSO Registry, splitting data into two groups to represent similar size patients to those included in the cost analysis.

ECMO requires a lower initial expenditure but has higher ongoing costs than BIVAD. This is due to continuing blood product usage and greater staffing requirements. There is a time when both systems are equal in cost. With no complications, BIVAD becomes cheaper after 30 days for a 10 kg patient and 32 days for a 35 kg patient.

Consultation with the ELSO Registry and review of our own data suggests smaller patients require longer ECMO support (mean 322.2 hrs 8–10 kgs; 260.5 hrs 30–40 kg, $p < 0.05$). The mortality rates of the two cohorts on ECMO are similar and increase with duration of support. BIVAD survival data is more favourable.

The differential morbidity and mortality rates of the two systems favour the VAD strategy. The financial benefits are two-fold:

1. With an associated higher incidence of complications, ECMO will preferentially escalate costs relative to BIVAD, displacing the point of intersection to an earlier time; hence, BIVAD becomes more cost effective sooner.
2. When considering the total cost of the service 'per survivor', the decreased mortality rate of the BIVAD system may incur fewer costs than ECMO from supporting patients who succumb to their illnesses.

Costing more complex scenarios demonstrates these conclusions. Further benefits of VADs may include decreased exposure to blood products, with lowered sensitisation of the panel reactive antibody, improving chances of transplantation. The improved morbidity on extracorporeal support with VAD will also shorten the length of inpatient hospital time post-transplantation, providing additional financial benefit.

Thus, there are financial and intact survival benefits of considering an elective VAD strategy for supporting patients with single organ, end-stage cardiac failure awaiting heart transplantation. This may not be applicable to all weight groups.

The incidence of pulmonary arterial hypertension in infants with bronchopulmonary dysplasia

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Broncho-pulmonary dysplasia (BPD) is a chronic lung disease affecting premature born babies and more particularly those with a very low birth weight. Incidence may vary from 35 to 57.2% in newborns of less than 1500 g. With today's higher quality of prenatal care, survival of very premature newborns has permitted us to diagnose BPD disease involving both the alveolar architecture and the pulmonary vascular tree. In these patients, pulmonary vascular pressures that are high in-utero and at birth decrease slowly and remain high. The incidence of pulmonary arterial hypertension is unknown in this population.

We reviewed 56 charts of patients that had a known diagnosis of BPD and that had left the hospital with oxygen treatment at home. These patients were followed in the pneumology clinic between 2003 and 2005. The charts were reviewed for perinatal history, cardiovascular evaluations, respiratory findings (number of ventilation days, ventilation pressures, oxygen, etc.), other treatments, growth, neonatal infections and rehospitalisations after discharge.

The average gestational age at birth was 28+2/7 weeks and their weight 1086 g. Antenatal steroids were given in 80.7% of mothers. Patients were ventilated for an average of 22.8 days, received CPAP for 21.8 days and total oxygen treatment of 190.4 days. Surfactant was given to 76.8% of patients. The average corrected age of weaning from oxygen is 6.23 months. Cardiovascular evaluations included EKG, echocardiogram and cardiac catheterization for respectively 71%, 95% and 2% of patients. The incidence of right ventricular hypertrophy (RVH) on EKG was 20.5%. If only one EKG criteria for RVH is included, the incidence increases to 33.3%. The incidence of increased right ventricular pressure diagnosed by echocardiogram was 14.6%. Only one child had a cardiac catheterization and it was normal. Overall, abnormalities on at

least one EKG or echocardiogram suggesting pulmonary arterial hypertension were found in 31.7% of our patients.
In conclusion, the incidence of pulmonary arterial hypertension in infants with BPD is non negligible. Close cardiovascular follow-up of this population is mandatory. Further studies are needed to assess risk factors and optimal therapeutic options in infants with BPD.

Improved outcome for the modified Norwood procedure using side-to-side ascending aorta to pulmonary artery anastomosis in children with diminutive ascending aorta

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Objective: End-to-side implantation or patch reconstruction techniques of the diminutive ascending aorta (<2.5 mm) frequently produce poor results, probably due to coronary insufficiency conferring significant risk for these patients during the Norwood procedure. Side-to-side anastomosis of the ascending aorta to pulmonary artery appears to reliably produce unobstructed coronary flow; we examined outcomes for this approach.
Methods: Since August 2003, 9 consecutive patients with hypoplastic left heart syndrome underwent a modified Norwood procedure with side-to-side anastomosis of a diminutive ascending aorta to the pulmonary artery. All repairs employed hypothermic circulatory arrest (HCA) and a modified Blalock-Taussig shunt. After transecting the main pulmonary artery, the diminutive ascending aorta was clipped and transected proximal to the arch with cardioplegia administered into the ascending aorta via hand-held catheter. The ascending aorta was opened longitudinally to the bulb between the left and right coronary arteries and anastomosed to the pulmonary artery just posterior to the facing commissure of the pulmonary valve (Figure). The circulation was then arrested and the arch was reconstructed in all cases with native tissue without the use of patch material (Figure inset). Results with this approach were compared to 9 consecutive preceding patients (encountered July 2000–July 2003) managed with end-to-side implantation of the diminutive ascending aorta to the pulmonary artery.
Results: 30-day and hospital survival were 100% and 88% for the side-to-side group versus 56% and 44% for the end-to-side implant group. There was no difference for total bypass or aortic cross-clamp times; however, HCA times were reduced for the side-to-side group (34 ± S.D. 3.9 minutes versus 50 ± S.D. 8.8 minutes; $p = 0.0001$). One death occurred after discharge in the end-to-side group (33% long-term survival); all eight hospital survivors in the side-to-side group have undergone a bidirectional cavopulmonary anastomosis [median follow-up 24 months (range 8–39 months)].
Conclusions: 1) In our institution, early survival and HCA times were substantially improved using side-to-side anastomosis of the diminutive ascending aorta to the pulmonary artery during the modified Norwood procedure. 2) Deaths from coronary insufficiency may be reduced with this technique compared to end-to-side implantation.
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Late presenters of transposition with intact interventricular septum: King Faisal Specialist Hospital and Research Centre-Jeddah Experience

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Introduction: The optimum management of patients presenting with TGA/IVS beyond the neonatal period remains controversial. The recommendations of management range from training of left ventricle with pulmonary artery banding and systemic-pulmonary shunt before the arterial switch operation (ASO) to intra-operative assessment of the ability of LV to tolerate an ASO.
We present our experience at KFSHRC-J in managing patients presenting beyond the neonatal period with TGA/IVS.
Methods: We reviewed the charts of all patients with TGA/IVS who presented after the neonatal period. We defined the late presenters as patients who were older than 4 weeks at the time of the first surgical intervention. The data collected included demography, ECHO and cardiac catheterization findings, surgical details, need for ECMO, the ICU course and outcome.
Results: From April 2001 to November 2006, 17 patients (10 males and 7 females) with median age 5 m (range 1.25–82 m) and median weight 5.13 kg (range 2.5–19 kg), were admitted. They were divided into 2 groups: LV training and No LV training group. Eleven patients were selected for LV training. This group had a median age 6 m (range 1.25–72 m) and median weight 5.4 kg (range 2.5–17.2 kg). ECHO showed LV compression with good function in all these patients. Cardiac Catheterization was performed in 7 of 11 patients and showed median LV to systemic pressure (LV/SP) ratio 0.53 (range 0.31–0.71). The median shunt size was 5 mm (range 4–6 mm) and the median LV pressure as percentage of systemic pressure was 67% (range 55–120%). Nine of 11 patients required inotropes (DOI) and mechanical ventilation (DOV) for median duration of 4 days (range 0–16 days) and 3 days (range 0.25 to 10 days) respectively. The median length of stay (LOS) in PICU was 5 days (range 1–11 days). Seven of the LV training group had ASO, 3 are awaiting ASO and 1 died a few hours after LV training. The median interval between LV training and ASO was 14 days (range 11–229 days). The median age of these 7 patients was 8.5 m (1.75–44 m) and median weight 6.5 kg (2.8-12.5 kg). Cardiac catheterization in 3 of the 7 patients showed median LV/SP ratio 0.78 (0.76–0.83). Two of 7 patients required ECMO. For the 7 patients the median DOI and DOV were 4 days (1–10days) and 3 days (14 hr–35 days) respectively. The median LOS was 4 days (14 hr–42 days). One patient died on ECMO. Six patients had no LV training. The median age was 4.75 m (range 1.5–82 m) and median weight was 4.2 kg (range 2.5–19 kg). ECHO showed LV compression in 3 of the 6 patients, with LVOTO in 2 and good cardiac function in all. Cardiac catheterization was performed in

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5 patients and showed median LV/SP ratio 0.67 (range 0.55–1.19). Three of 6 patients required ECMO. For the 6 patients the median DOI and DOV were 4 days (12 hr–11 days) and 4 days (12hr–7 days) respectively. The median LOS was 12 days (9–14 days). One patient on ECMO died.
Conclusion: The presence of LV compression on ECHO and a LV/SP ratio less that 2/3 were good guides for the need for LV training. The no LV training group was more likely to require ECMO compared to the LV training group although the final outcome was similar.

Reduced expression of atrial connexin43 and Sodium pump after pediatric heart surgery

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Background: Myocardial dysfunction and arrhythmias may be associated with congenital heart defects, and can also be induced by heart surgery with cardiopulmonary bypass (CPB). A differential expression of the gap junctional components connexin40 and connexin43 (Cx40 and Cx43) and the Sodium pump may contribute to post-operative arrhythmias and contractile dysfunction. We hypothesize that the expression of these components is influenced by the CPB and also by the type of heart defect.
Patients and methods: In sixteen pediatric patients undergoing corrective heart surgery mRNA expression of Cx40 and Cx43 and two isoforms of the Sodium pump (ATP1A1 and ATP1A3) was studied in right atrial myocardium, excised before and after CPB. Depending on the type of heart defect, the patients were assigned to a VO group (volume overloaded atrium, $n = 8$) and a NO group (not volume overloaded atrium, $n = 8$).
Results: Patients from the VO and NO group differed significantly regarding their ratio of right/left atrial size ($p = 0.0001$), whereas no difference was found regarding the age, gender and CPB variables. Following CPB, the expression of atrial Cx43 ($P = 0.008$) and both Sodium pump isoforms (ATP1A1 and ATP1A3, $p = 0.008$ and 0.038) decreased significantly in the VO group. However, no change was found in the NO group. Atrial Cx40 mRNA expression did not change significantly after CPB, in both groups. The differential expression of these molecules did not correlate with clinical outcome in terms of development of arrhythmia and myocardial dysfunction.
Conclusion: This study revealed a significant influence of the CPB on the expression Cx43 and isoforms of the Sodium pump. The volume overload itself had no influence on atrial expression of connexin or Sodium pump isoforms, but it changed the susceptibility of atrial myocardium to the effects of CPB. Although the level of expression of these molecules did not predict clinical outcome, they could potentially serve as markers for susceptibility to arrhythmia or myocardial dysfunction.

Mortality in the first year after discharge following congenital heart surgery

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High-risk follow-up for patients with hypoplastic left heart syndrome has been shown to diminish mortality after discharge following stage one palliation. Identifying other patients at high-risk for death after being discharged following congenital heart surgery (CHS) might lead to development of similar high-risk follow-up programs. We attempted to identify those patients at highest risk for death after CHS.
The web-based medical records of all patients undergoing CHS from February 2002 through May 2006 were reviewed. Patients who died during the first year after discharge were compared to those who lived.
There were 1301 patients who underwent CHS during this period. Of those, 1048 have a complete 1 year follow-up, with 253 not yet 1 year post discharge. There were 25 (1.9% of CHS pts) deaths after discharge, at an average of 128 days post discharge (range 2–341 d). Patients who died after discharge were younger and smaller at the time of their operation than those who did not 46.4 vs. 188 d ($p < 0.01$) and 3.65 vs. 6.3 kg ($p < 0.01$). Those dying after discharge had significantly longer postoperative stays (44 vs. 7 d, $p < 0.01$). Of the patients who died after discharge, 15 patients had single ventricle palliations, 9 patients had systemic to pulmonary artery shunts (5 s/p Norwood) and 5 patients died with bidirectional Glenn physiology. There were 14 (58%) sudden unexpected deaths and 9 (38%) deaths that occurred during a hospitalization. There were 4 patients who were noted to be either syndromic or had significant neurologic deficits prior to initial discharge after CHS.
Death in the first year after CHS is not uncommon, occurring in almost 2% of operated patients. Patients who are operated on at a younger age, those with prolonged postoperative hospital stays, those with single ventricle palliations and those with systemic to pulmonary artery shunts are at highest risk.

Utilizing a web-based medical record and a point of care testing device to achieve performance improvement by real-time performance measurement in a congenital heart surgical program

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Objective: Performance improvement is achieved only through the measurement of objective indicators of a group's process or procedure. Maintaining excellence and improving performance in a cardiac surgical program requires accurate and repeated objective measurement of a program's performance as a whole, and also at the individual patient level. Real-time performance assessment may allow a program to rapidly adapt to changing conditions to maintain or improve outcomes.
Methods: In July of 2001, a point of care testing device (i-Stat, Abbott) was introduced to the cardiac ICU to objectively measure the performance of individual patients as they progressed through their intensive care course. The blood lactate data would be used to guide the individual patient through their hospital course and later to evaluate the performance of an entire cohort of patients. In February 2002, Miami Children's Hospital helped develop software (i-Rounds, Teges) which allowed the team to track the ultimate outcome of each patient admitted to the cardiac program in real-time. The data was entered as the official hospital medical record and was stored as a sequel server database. This technique makes every piece of data entered available via query. Outcomes were then made available on the internet, in real-time, for internal review by team members or external review by the public, via the

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domain www.pediatricheartsurgery.com. The data was available in both tabular and graphic form and risk adjusted for age and complexity. This data was used to measure the cardiac team's performance as a whole.
Results: Currently, 88 members of the cardiovascular team have accessed the program at least once weekly for either data review or entry, reflecting team-wide acceptance of the program. From February 2002, through November 2006, the outcomes of 1440 congenital heart surgery procedures at MCH were accessed. Mortality at 30 days was 2.2% for these patients. Mortality from July 1995 through June 2001 was higher at 3.7% ($p = 0.02$). Operative performance improvement was measured by utilizing the last lactate value recorded in the OR. Lactate in pts less than a year of age decreased from 4.2 mmoles/l in 2001 to 3.0 in 2006 ($p < 0.01$).
Conclusions: There is uniform agreement that performance improvement can only be achieved through the use of accurate and timely performance assessment. We achieved performance improvement in a congenital heart program by measuring outcomes of both individual patients and the program's performance as a whole, in real-time.

Improving pain control in pediatric cardiac surgery: adding continuous local anesthetic infusion

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Background: Awareness and treatment of surgical pain in children has improved over the years, but continues to be challenging, especially in the post-operative period. With this knowledge at hand, adjunct therapies used to manage post-operative pain were investigated at a multi-center children's hospital. Our goal was to determine the safety and efficacy of the continuous infusion pump ON-Q Pain Buster in the pediatric patient undergoing cardiac surgery. Since local anesthetics can cause systemic toxicity, serum plasma levels where monitored during the post-operative period in Phase I of the study.
Methods: In a prospective, randomized and double blind study we compared either levobupivacaine 0.25% or bupivacaine 0.25% vs. saline in an elastomeric continuous infusion pump (ON-Q Pain Buster). A total of 72 children undergoing median sternotomy were enrolled after obtaining informed consent or assent. Infusion pump size and rate was determined by patient weight at time of surgery. Prior to incision closure, a 0.5 cc/kg bolus of local anesthetic was injected into the wound, catheter was placed; and incision closed. In addition to the continuous infusion pump, patients enrolled were treated with the institutions standard post-operative sedatives and analgesics; such as midazolam, lorazapam, morphine, ketorolac, acetaminophen and ibuprofen. Each child was monitored with the infusion pump for a total of 72 hours post operative. Plasma levels of levobupivacaine 0.25% or bupivacaine 0.25% were measured at 12, 24, 48 and 72 hours in the first 41 patients (Phase I). All patients were monitored for side effects of the medications. Age appropriate pain scores were recorded; along with any sedation or analgesics being administered (Phase II). Nursing personnel were in serviced on the study and standardize orders were created. Secondary parameters included time to first oral intake, use of anti-emetics, and length of stay.
Results: Phase I of the study evaluated the safety of the local anesthetic. Dosage of the local anesthetic ranged from 0.09–0.45 mg/kg/hr. The mean dose was 0.23 mg/kg/hr. There were no reported side effects from the local anesthetic and no levels were considered toxic (4 mcg/cc). Blood sampling was discontinued after safety had been established. Phase II evaluated analgesic efficacy. All subjects were included in this analysis. The total amount of morphine required over the three days was statistically less in the test group than the control group ($P = 0.05$). The number of patients requiring no morphine was greater in the test group versus the control group. The test group also required less midazolam and less ketorolac than the control group. Overall, pain scores were less in the test group, but statistical significance was not achieved. The usage of ondansetron, which was a marker for nausea and vomiting, approached statistical significance in the test group compared to the control group ($P = 0.07$). There was no difference in any of the other secondary parameters.
Conclusion: This study demonstrated that a continuous infusion of either 0.25% Bupivacaine or 0.25% Levobupivacaine was a useful adjunct therapy for patients undergoing a median sternotomy. It reduced the use of post-operative analgesics and sedation.

Anomalous coronary arteries in left ventricular lesions affecting surgical repair

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Coronary artery anomalies have been well described in association with complex congenital cardiac lesions such as pulmonary atresia with intact ventricular septum, transposition of the great arteries, and tetralogy of Fallot. Traditionally, there has been less concern regarding coronary anomalies with left-sided lesions. We present three cases where left-sided lesions with coronary anomalies profoundly affected surgical intervention and patient outcome.
Case 1: A two day old male with hypoplastic left heart syndrome variant underwent Norwood palliation. Pre-operative echocardiogram demonstrated mitral atresia with a hypoplastic left ventricle, aortic hypoplasia, and ventricular septal defect. Coronary arteries were not examined in detail. Intraoperatively, the left main coronary artery was inadvertently transected due to a long intramural course. Repair was difficult due to the extremely small caliber of the vessel. The patient required extracorporeal membrane oxygenation post-surgery, and later succumbed to multi-organ system failure.
Case 2: A two day old newborn male was transferred for intervention for Shone's complex. Echocardiogram demonstrated mild mitral hypoplasia, ventricular septal defect, mild aortic valve dysplasia, mildly hypoplastic aortic arch with periductal coarctation, and diminished left ventricular function. Coronary arteries were not dilated. However, images demonstrated the left coronary artery to arise from the undersurface of the right pulmonary artery. The patient underwent complete operative repair including reimplantation of the left coronary artery to the aorta. Discharge echocardiogram showed good biventricular systolic function with normal antegrade coronary flow, and no residual shunts or outflow tract obstruction.
Case 3: A newborn infant male with a prenatal diagnosis of complex heart disease was diagnosed postnatally to have double outlet left ventricle with the aorta to the right of the pulmonary artery, moderately hypoplastic tricuspid valve and right ventricle, and a small restrictive ventricular septal defect. Coronary artery origins were difficult to visualize but appeared normal. Surgical repair was planned with pulmonary artery ligation and placement

of a Blalock–Taussig shunt. Shortly after induction of anesthesia, the patient developed hypotension and ST segment changes, with left ventricular hypokinesis. Transesophageal echocardiogram could not demonstrate coronary artery origins in the usual location. External and internal examination of the heart demonstrated absence of coronary ostia and proximal coronary arteries. Cardioplegia was delivered retrograde through the coronary sinus, with what appeared to be return through sinusoids in direct communication with the right and left ventricles. Left ventricular function did not recover and the patient expired in the operating room.
Summary: Detailed coronary artery assessment, including careful evaluation of coronary artery origins and their course, should be part of the routine echocardiographic assessment of left-sided congenital lesions, in addition to those lesions typically known to be associated with coronary anomalies.

Long-term outcome of pediatric cardiosurgical patients requiring prolonged mechanical ventilation

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Objective: To assess one-year mortality and its risk factors in patients requiring prolonged mechanical ventilation (PMV) after pediatric cardiac surgery.
Methods: Retrospective cohort study of 41 patients requiring mechanical ventilation for 20 days or more during 2001–2005.
Results: One-year mortality rate was 49% (20 of 41 patients). Twelve patients died in Cardiac ICU, 8 patients died 6 to 331 days (median, 72 days) after transfer to another hospital. The reasons for PMV included low cardiac output (resulting from transient myocardial dysfunction, correctable residual cardiovascular defect or intractable residual cardiovascular defect), chronic lung disease, chylothorax, diaphragm paralysis, upper airway obstruction, bronchomalatia, sepsis, neurological complications (apalic syndrome, spinal lesion, cerebral edema following cardiopulmonary resuscitation), pulmonary hypertension, recurrent bronchospasms, and plastic bronchitis. Of these, intractable residual cardiovascular defect ($p < 0.0001$), long duration of inotropic support ($p = 0.016$) and mechanical ventilation ($p = 0.007$) were associated with patients death. Diaphragm paralysis ($p = 0.015$) and correctable residual cardiovascular defect ($p = 0.04$) were associated with survival. Seven patients with tracheostomy died. In patients with intractable residual cardiovascular disorder heart transplantation was not performed due to shortage of donor hearts.
Conclusion: One-year mortality in pediatric cardiosurgical patients requiring prolonged mechanical ventilation was high. Intractable cardiovascular defect was risk factor for mortality; diaphragm paralysis and correctable cardiovascular defect were associated with survival.

Nursing morbidity and mortality conference and journal club cycles: paving the way for nursing autonomy, enhanced patient safety, and evidence-based practice

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Background: The ability to critically review patient care is a prerequisite for autonomous nursing practice, and the ability to use science to guide care delivery a hallmark of professional practice. The nurses in our Cardiovascular Intensive Care Unit (CVICU) developed a nursing-focused Mortality and Morbidity (M & M) forum to advance their practice. The chief objectives of Nursing M & M were to improve error reporting, provide peer review, and promote the use of the best-available evidence to guide system and practice changes that would advance clinical excellence.
Methods: A team composed of CVICU bedside nurses, advanced practice nurses (APNs), the nurse manager, the CVICU medical director, and a quality manager joined together and created a process for three-month cycles of Nursing M & M conference, Journal Club, and a subsequent educational session in nursing staff meetings. The team identified a specific topic for each quarterly cycle and developed that topic through the entire cycle. The CVICU nurse practitioner and medical director worked collaboratively to present a clinical scenario at the M & M conference. Next, a CVICU APN worked with the Journal Club team to discuss a pertinent citation related to the cycle topic. In the final session of the series, a specific educational presentation was developed and lead by the bedside nurses and a CVICU APN incorporating the best-available evidence about the selected topic.
Results: Our team has performed two complete cycles. We found that interdisciplinary practice changes were indicated and that patient care could be improved by this process. For example, one cycle examined communication practices within the CVICU and led to formalizing interdisciplinary communication on daily work rounds and the implementation of SBAR. The second cycle examined resuscitation practices with a specific focus on patients with an open sternum and those with single ventricle physiology who decompensated following a stage I Norwood procedure.
Conclusions: Within our CVICU, cycles of Nursing M & M, Journal Club, and a focused educational session during nursing staff meetings supported patient safety, fostered professional autonomy, increased evidence-based practice, and facilitated collaborative, interdisciplinary patient care amongst our team members.

Short QT interval in children and adolescents

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Background: Short QT Syndrome (SQTS) is a rare familial electropathy that affects a wide range of subjects, from fetal life to old age. It involves gain-of-function in potassium channelopathies that lead to unique ECG findings and predisposes affected patients to atrial or ventricular fibrillation (VF) and sudden death (SD). It is unclear how short the QT interval (QT) should be to suggest SQTS.
Methods: We compared the ECG findings of 6 subjects (all male) with symptoms, dysrhythmias and/or a family history (FHx) compatible with SQTS (Group A) with those of 4 subjects (all male) with a short QT interval but a negative FHx and no dysrhythmias (Group B).
Results:

	Age (mean)	QT (mean)	QTc (mean)
Group A	7–19 yr (15 ± 7)	245–320 ms (298 ± 26)	286–320 ms (313 ± 13)
Group B	16–17 yr (17 ± 0.5)	317–360 ms (338 ± 23)	332–343 (334 ± 5)
p Value	0.20	0.02	0.01

In addition, all but one subject in Group A had atypical T waves (tall and peaked) and the absence of an isoelectric ST segment.

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Two of them had history of VF, one had aborted SD, one had ventricular tachycardia and four had positive FHx (SD or SQTS). Only four patients in Group A underwent electrophysiologic testing. All four had short atrial effective refractory period (190 ms or less) and ventricular effective refractory period (200 ms or less). Atrial fibrillation and/or VF was inducible in two, non-inducible in one and not attempted in a patient with VF.

Conclusion: In this small patient cohort, only the subjects with SQTS had atypical T waves with absent isoelectric ST segment. The QTc interval was shorter, less than 330ms, in our SQTS patients when compared with the group of subjects with only enhanced repolarization.

Abstract: Short QT syndrome (SQTS) is a rare familial electropathy. It involves gain-of-function potassium channelopathies that lead to enhanced repolarization. Affected subjects are predisposed to atrial and ventricular fibrillation. It is unclear how short the QT interval should be to suggested SQTS phenotype. We compared the ECG of 6 subjects with SQTS phenotype and/or positive family history (FHx) for SQTS (Group A), with 4 asymptomatic subjects with a relatively short QT interval, less than 360ms, and a negative FHx (Group B). Group A had a short QT (245–320 ms) and QTc (268–320 ms) than Group B (QT 317-360ms and QTc 332–343 ms). In this small cohort, subjects with a short QT interval ranging between 317–360 ms (and QTc 332–343 ms) were all asymptomatic, in spite of enhanced repolarization. Subjects with positive FHx and/or SQTS had ultra short QT (245–320 ms). A larger population and multicenter registry is recommended.

Myocardial tissue Doppler changes in patients with bronchopulmonary dysplasia

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Background: Bronchopulmonary dysplasia (BPD) occurs primarily when immature lungs are exposed to inflammation, oxygen, and ventilation. In the most severe forms, cor pulmonale can develop and the onset is often insidious. Recently, tissue Doppler imaging has been found to correlate with right ventricular (RV) dysfunction as measured by cardiac catheterization in adults. Minimal tissue Doppler data exists in infants and children. We hypothesized that tissue Doppler changes which reflect right ventricular function will worsen with increasing severity of BPD.

Methods: Limited echocardiograms were performed with standard M-mode measurements. Mitral and tricuspid inflow velocities were obtained from an apical four chamber view and tissue Doppler parameters were obtained from the lateral sides of the mitral and tricuspid annulus and interventricular septum. Myocardial performance index (MPI) was calculated from the tissue Doppler data obtained at the respective valve annulus. BPD severity was scored using the NICHD/NHLBI/ORD workshop rating scale by physicians blinded to the echocardiogram results.

Results: 21 patients (6 controls with no/mild BPD, 7 with moderate BPD and 8 with severe BPD) average gestational age of 26.5 ± 2.4 weeks and average corrected gestational age 44.9 ± 19.1 weeks. No significant differences in demographics between groups. When comparing controls to patients with moderate BPD, there was a significant difference in the early tricuspid inflow velocity to early tricuspid tissue Doppler velocity (E/E´) ratio (4.5 ± 1.5 vs. 6.2 ± 1.4, $p = 0.05$). A difference in the LV MPI index (0.43 ± 0.04 vs. 0.50 ± 0.06, $p = 0.04$), reflected impaired LV function in the moderate group. When comparing controls to severe BPD patients, early (48 ± 12 vs. 65 ± 13, $p = 0.03$) and late (53 ± 8 vs. 64 ± 11, $p = 0.05$) tricuspid inflow velocities were increased. There was a difference in the E/E´ ratio (4.5 ± 1.5 vs. 7.1 ± 1.9, $p = 0.02$). Patients with severe BPD had an abnormal LV MPI (0.43 ± 0.04 vs. 0.55 ± 0.12, $p = 0.03$). No significant difference was found when comparing patients with moderate BPD to those with severe BPD. Linear regression demonstrated a correlation between BPD category and RV E/E´ ($p = 0.007$, $R^2 = 0.33$) and LV MPI ($p = 0.02$, $R^2 = 0.28$).

Conclusion: Increasing RV E/E' ratio, suggestive of increased RV end-diastolic pressures, correlates with clinical severity of BPD. Abnormal LV MPI was noted to correlate with the grade of BPD. Further longitudinal studies are needed to assess the usefulness of tissue Doppler imaging in BPD.

Closure of the arterial duct during the modified Blalock-Taussig shunt operation: influence on early postoperative hemodynamics

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Background: The modified Blalock-Taussig (MBT) shunt is a procedure of choice for neonates with single ventricle physiology and ductus dependent pulmonary blood flow. The policy of closing vs. not closing patent ductus arteriosus (PDA) during MBT shunt surgery differs between institutions. The patency of PDA may adversely influence the early postoperative hemodynamics by increasing pulmonary blood flow. The objective of this study was to compare the early postoperative hemodynamic status of newborns with and without ductal closure during MBT shunt procedure.

Methods: Retrospective observational study of 74 newborns, who underwent primary MBT shunt surgery without associated intracardiac repair at our institution between January 1997 and November 2006. The early postoperative period was defined as first 24 postoperative hours. The preoperative and operative variables were analyzed as baseline characteristics of both groups. Mean arterial pressure (MAP), diastolic arterial pressure (DAP), central venous pressure, heart rate, arterial oxygen saturations, arteriovenous saturation difference, inotropic support, urinary output, blood losses and fluid balance recorded for the postoperative hours 6, 12, 18, 24 were analyzed as the main outcome measures.

Results: PDA was surgically closed in 27 patients (group 1) and was not closed during operation in 47 patients (group 2). Significantly less patients with pulmonary atresia were in group 1 then in group 2 ($P < 0.03$). Hospital mortality was 3.7% in the first group and 8.5% in the second group ($P = 0.42$). Early shunt failure did not occur in group 1 and was diagnosed in 5 patients in group 2 ($P = 0.08$). For group 1, significantly less positive fluid balance ($P < 0.01$) was recorded during first 6 postoperative hours, higher MAP ($P < 0.03$) and higher DAP ($P < 0.003$) were noted during first 12 postoperative hours, and lower heart rate ($P < 0.01$) was recorded during first 18 postoperative hours, comparing with group 2. Data analysis did not reveal significant differences in remaining outcome measures during the whole study period.

Conclusion: PDA closure during MBT shunt procedure influences positively hemodynamic status in very early postoperative period without adversely affecting hospital mortality.

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