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## CONGENITAL CARDIOLOGY TODAY

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## CONGENITAL HEART DISEASE IN THE DEVELOPING WORLD

*By R. Krishna Kumar, MD*

The specialties of pediatric cardiology and pediatric cardiac surgery have rapidly evolved in the last four decades, largely spurred by the dedicated efforts of pioneering professionals together with major technological advances. Today, they are good examples of medical specialties that largely depend on advanced training and sophisticated modern technology to make an impact on affected patients. The list of achievements in the last 40 years is truly impressive.<sup>1</sup> Either correction or some form of palliation is possible for the majority of congenital heart diseases in most advanced pediatric heart centers. The results are often dramatic in individual patients. The specialties continue to grow impressively with progressive sophistication and advancement of technology. The industry that has developed in connection with the technology enthusiastically supports this progress and works in close collaboration with pediatric cardiac surgeons, pediatric cardiologists and other providers of pediatric cardiac care. With the advent of globalization, this new technology has become widely available for patients all over the world.

While impressive strides have been made in the quality of care, the costs of care continue to be rather high.<sup>2,3</sup> Most interventional or surgical procedures are simply unaffordable to most families in the absence of some form of health insurance. Unlike other specialties, over the years the situation has not changed significantly. Most new technologies are expensive

when introduced. Over a period of time after introduction, the costs decline with increasing consumption and competition among manufacturers. Coronary angioplasty is a good example. Since its introduction the cost of angioplasty hardware has declined substantially. The cost reduction was made possible through an exponential growth in consumption. The large size of the potential market spurred a number of manufacturers to enter the fray and the competition forced them to focus on reducing costs.

***“Unfortunately ... very few high-quality institutions with comprehensive facilities to take care of children with congenital heart disease exist outside of the developed world.”***

The size of the market or the consumers of “pediatric cardiac technology” is perceived as small. This is because the number of children affected with congenital heart disease is relatively small when compared to acquired coronary artery disease. However, most manufacturers of “pediatric cardiac technology” have not been looking beyond industrialized nations, at least not until recently. The global burden of congenital heart disease is likely to be several times that of the industrialized or developed nations simply because of a much larger population. Unfortunately, however, very few high-quality institutions with comprehensive facilities to take care of children with congenital heart disease exist outside of the developed world.

It is not just the costs of consumables and equipment that make pediatric heart care expensive. A number of other factors contribute. They include infrastructure costs and costs of em-

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playing skilled manpower. The specialty consumes considerable material and human resources just to be able to deliver "basic" services. The manpower required for taking care of a typical child undergoing congenital heart surgery include: the pediatric cardiac surgeon, pediatric cardiologist, pediatric anesthesiologist, pediatric intensivists (ideally pediatric cardiac intensivists), pediatric perfusion technologists, pediatric nurses and respiratory therapists. The infrastructure includes a sophisticated operation theater with current cardiopulmonary bypass equipment, a modern pediatric intensive care facility, a cardiac catheterization laboratory, and a non-invasive imaging laboratory for echocardiography.

**What is the Developing World?**

The term "developing world" loosely encompasses a group of middle- and low-income countries classified by the World Bank largely on basis of the per-capita gross national income (see <http://www.worldbank.org/data/countryclass/countryclass.html>). Time and experience have shown, however, that social conditions and the general well being of people may not necessarily improve when a country's average income level increases. Considerable differences exist in health care delivery and health indices among these countries. The human development index (HDI) is a more comprehensive index that takes into account quality of life as measured by life expectancy, infant mortality and literacy. Nations are now ranked on basis of HDI and this list is revised periodically.

***"Given the current health care structure in most of the developing world, it is not surprising that the vast majority of children born with congenital heart disease receive no attention."***

cally. These definitions and classifications are essentially broad generalizations (see <http://hdr.undp.org/reports/global/2004>). Not only do the countries in the developing world differ markedly in their health statistics from developed countries, but there is also marked variability between countries of medium and low human development. Further, there is considerable variability even within countries.<sup>4</sup>

With the advent of globalization there are important changes taking place in the developing world. Selected, small urban pockets within the developing countries have become increasingly affluent and often cannot be distinguished from developed nations. The per capita income in these pockets is considerably higher and infrastructure is quite sophisticated in comparison with the rest of the country. Essentially, these are islands of "development" that are situated amidst an ocean of deprivation and poverty. These affluent pockets are looked upon as prospective, lucrative markets for the global industry and local entrepreneurs. The health care facilities in these areas are often very sophisticated, but care

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comes at a price that is not at all affordable to the average citizen of the country. There are sharp paradoxes: with areas of extreme poverty, malnutrition and ill health without access to the very basic health services in urban slums situated adjacent to affluent neighborhoods with access to the very best in tertiary health care. These paradoxes are visible even to the casual visitor to modern metros of the developing world. Today, there are examples of patients from United Kingdom who are now flying into selected centers in Indian Metros (such as Mumbai and Bangalore) for cardiac surgery that is offered at a considerably lower cost as compared to private centers in the UK. Yet, the vast majority of those living in the less privileged sections of the same city cannot afford care in these centers. The gap between the “haves” and “have-nots” only widened over the last few decades when developing nations have attempted to make the transition to emerging economies. Quality tertiary health care essentially can now only be afforded by a tiny fraction of the population. Sadly, however, very little efforts are being made to reach out to a larger fraction of

the population, as health care is becoming increasingly corporatized and driven by profit motives. The situation is particularly frustrating for patients and families who are often unable to afford quality care that is available in the cities in which they live.

**Congenital Heart Disease in the Developing World**

The majority of the world’s inhabitants live in developing countries. Although there are no reliable statistics, the number of affected children in the developing world is likely several times that of the developed world simply because of the large populations. Given the current health care structure in most of the developing world, it is not surprising that the vast majority of children born with congenital heart disease receive no attention. The situation can only change through establishment of regionalized centers of excellence within each country that focus on affordable care for children with congenital heart disease. The population, geography and available resources will determine the number of such centers. Simultaneously, institution of a number of other measures targeted at encouraging early diagnosis and prompt referral of children with congenital heart disease is required. It is also necessary to obtain an estimate of the absolute and relative magnitude of the problem of congenital heart disease through appropriate surveys. Formulation of health policies in the developing world for congenital heart disease in a complex and rapidly changing health care environment is likely to remain a for-

*“Formulation of health policies in the developing world for congenital heart disease in a complex and rapidly changing health care environment is likely to remain a formidable challenge.”*

**MAY CONFERENCE FOCUS**

**The 1st Annual Toronto Symposium: Contemporary Questions In Congenital Heart Disease 2005**

May 29-31, 2005; Toronto, Canada  
[www.sickkids.ca/cardiology](http://www.sickkids.ca/cardiology)

This symposium from Sick Kids Hospital in Toronto (a health care, teaching and research facility dedicated exclusively to children; affiliated with the University of Toronto) will aim to address hot topics in the field of child and adolescent cardiology.

Each lecture is framed to answer a specific and important question in pediatric cardiology. As such, the clinical management, evidence base, and current research understanding of very specific issues will be addressed in detail.

This program is planned to bring together experts in the field of congenital heart disease, answer important and provocative questions in the field of pediatric cardiology, develop a better understanding of the topics and provide opportunity for discussion and audience participation. Major topics include:

- Disease in the Normal Heart,
- Heart Failure/Cardiomyopathy, Catheter Intervention,
- Out-Patient Cardiology
- Surgical Outcomes
- Imaging, and Fetal Cardiology

There will be breakout sessions on Inpatient Care, Questions Parents Ask and How to Best Answer Them, and Genes and Development. There will also be debates on important topics.



midable challenge. Clearly in much of the developing world, the focus should be on health care delivery with an aim to reach out to the greatest proportion of affected patients. For example, it may not be appropriate to perform multi-staged palliative operations with a doubtful long-term survival or neurodevelopmental outcome (such as in hypoplastic left heart


*“In the coming issues of Congenital Cardiology Today, we hope to highlight these items systematically using the experience of selected centers that have been established in the developing world in the last 10 years.”*

syndrome) when there are many infants with relatively straightforward conditions (such as ventricular septal defect) that need a single corrective operation. Technology and human endeavor has to be appropriately directed towards this end. A list of action items that could be considered in order to establish a system for care of children with congenital heart disease in the developing world is shown in the table.

In the coming issues of Congenital Cardiology Today, we hope to highlight these items systematically using the experience of selected centers that have been established in the developing world in the last 10 years. The cumulative experience in terms of num-

Broad Category	Specific Issues
<b>Epidemiological Significance</b>	<ul style="list-style-type: none"> <li>● Global disease burden of congenital heart disease</li> <li>● The burden in developing world</li> <li>● Proportional infant and childhood mortality for CHD</li> <li>● Relative significance of CHD as compared to acquired heart disease such as rheumatic heart disease</li> <li>● Planning epidemiologic surveys in the developing world</li> </ul>
<b>Establishing a Center for Comprehensive Care of Patients with Congenital Heart Disease</b>	<ul style="list-style-type: none"> <li>● Personnel</li> <li>● Material resources; infrastructure</li> <li>● Support services and systems</li> <li>● Ensuring economic sustainability of the program</li> <li>● Overcoming philosophical hurdles (teamwork, multi-specialty institution-based care etc., infection control, etc.)</li> </ul>
<b>Resources</b>	<ul style="list-style-type: none"> <li>● Number of centers for comprehensive pediatric heart care in the developing world</li> <li>● Proportion of patients with CHD are likely to receive care in these centers</li> <li>● Health insurance for patients with CHD</li> <li>● Institutional and governmental support for individual patients</li> </ul>
<b>Special Challenges of the CHD Patient Population in the Developing World</b>	<ul style="list-style-type: none"> <li>● Malnutrition</li> <li>● Infections: Neonatal sepsis; lung infections</li> <li>● Consequences of late presentation:                             <ul style="list-style-type: none"> <li>● Pulmonary hypertension</li> <li>● Neurological complications and major neurodevelopmental sequelae</li> </ul> </li> <li>● Cardiac consequences: ventricular dysfunction, atrial enlargement and arrhythmias</li> </ul>
<b>Managing CHD Patients in Developing Nations: Practicalities</b>	<ul style="list-style-type: none"> <li>● Prioritization with respect to lesions</li> <li>● Cost-effective strategies for cardiac catheterization and interventions</li> <li>● Cost-effective strategies for congenital heart surgery and intensive care</li> <li>● The challenge of infection control</li> </ul>

Table 1. Issues involved in care of Patients with Congenital Heart Disease in Developing Countries.



**28th Annual Scientific Sessions**  
**and Melvin P. Judkins Cardiac Imaging Symposium**  
**May 4-7, 2005, Ponte Vedra Beach, Florida**

[www.scai.org](http://www.scai.org)

**The Society for Cardiovascular Angiography and Interventions**

Broad Category	Specific Issues
<b>Inexpensive and Indigenous Technology</b>	<ul style="list-style-type: none"> <li>● Occlusive Devices</li> <li>● Mechanical heart valves</li> <li>● Disposables for cardiac catheterization, congenital heart surgery and intensive care</li> <li>● Echocardiography machines</li> <li>● Cardiac catheterization laboratories</li> </ul>
<b>Training</b>	<ul style="list-style-type: none"> <li>● Pediatric Cardiology</li> <li>● Pediatric Cardiac Surgery</li> <li>● Anesthesiology and intensive care</li> </ul>
<b>Improving Awareness Through Education</b>	<ul style="list-style-type: none"> <li>● Pediatricians and primary care physicians</li> <li>● Adult cardiologists involved in the care of patients with CHD</li> <li>● Obstetricians, radiologists performing prenatal ultrasound</li> <li>● General public</li> </ul>
<b>Formulating Health Policies</b>	<ul style="list-style-type: none"> <li>● Defining the relative priority for congenital heart disease</li> <li>● Regulatory bodies on medical education</li> <li>● Policies on health insurance coverage</li> <li>● Support to institutions with pediatric heart programs</li> <li>● Research and education grants</li> <li>● Support for individual patients</li> </ul>
<b>Collaboration with Centers in the Developed World</b>	<ul style="list-style-type: none"> <li>● Exchange programs for training</li> <li>● Visits of teams from institutions in the developed world</li> </ul>
<b>Ethical Issues</b>	<ul style="list-style-type: none"> <li>● Bridging the gap between what is possible and what is feasible</li> <li>● Managing "complex" defects</li> <li>● Facilitating decision-making for families</li> </ul>

Table 1—(continued). Issues involved in care of Patients with Congenital Heart Disease in Developing Countries.

bers in these centers has been substantial. The challenges of providing care to a unique population with limited resources are many and worth sharing with the global community of providers of pediatric cardiac care.

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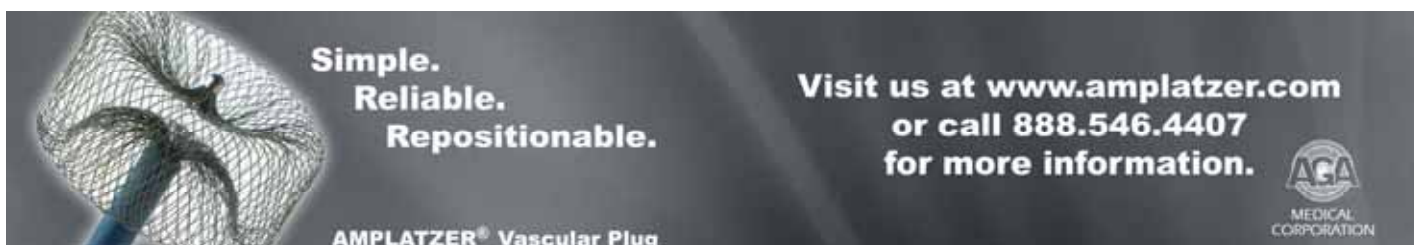
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## MEDICAL NEWS AND INFORMATION

**The Genetic Causes of One of the Most Common Types of Congenital Heart Defects are Being Studied by Academics at The University of Nottingham**

Professor David Brook in the University's Institute of Genetics is working in collaboration with colleagues at Leicester's Glenfield Hospital on the British Heart Foundation funded project, which will explore the molecular science behind the conditions Atrial Septal Defect (ASD) and Ventricular Septal Defect (VSD) — more commonly known as a 'hole in the heart' and accounts for around half of all congenital heart defects.

Until recently, ASD was thought to be sporadic and diagnosis for many patients came out of the blue. However, the team of researchers is studying patients — both adults and children — who have been diagnosed with ASD and other congenital heart disorders, and their families, to find out whether in some cases it could be an inherited condition.

Funded with a grant of £1,079,803 from the British Heart Foundation (BHF), the five-year project will be looking for more evidence to support the researchers' theory that the heart defect may be due to faulty genes, which helps to regulate the development of the heart.

The first breakthrough came when the researchers were studying the rare genetically-inherited condition Holt Oram Syndrome. Patients with the condition are born with limb defects, in some cases without any upper limbs. It is also characterized by a heart defect, most often a hole in the heart, either ASD or VSD.

A gene termed TBX5 was found to be responsible for the condition, which led the researchers to question whether it may also be a factor in cases of ASD or VSD that doctors previously believed to be sporadic.

In collaboration with researchers in Paris, the Nottingham group has now found that another gene called MYH6 is faulty in members of a family with inherited ASD. This work, published in a recent issue of the journal *Nature Genetics*, also shows that MYH6 is regulated by TBX5, which provides an important link between the two genes in the origins of heart defects.

Professor Brook said, "In patients with Holt Oram Syndrome their TBX5 doesn't work properly. It either doesn't tell the other genes that it controls what to do or there is not enough of it to make other 'down-stream- genes' behave as they should.

"We now know that MYH6 is an important downstream gene and we believe that a certain proportion of sporadic ASDs and VSDs may be due to an inherited predisposition. Thus we are testing patients for changes in their DNA to discover whether

either of these genes or two other related genes are implicated in sporadic heart abnormalities."

Learning more about how the genes work could eventually lead to a better diagnosis of the condition.

Professor Brook added: "Methods for DNA testing are improving all the time. It could be that in the future doctors could test just a single drop of blood to identify at birth whether a child has this faulty gene and warrants further examination for hole in the heart."

Professor Peter Weissberg, Medical Director of the BHF, concluded, "We are delighted to have funded this research, which provides a molecular explanation for why the wall that separates the left and right chambers of the heart (the atria) fails to form properly. It is a good illustration of how detailed research on a rare familial form of a condition can provide answers that may ultimately lead to treatments for a much larger population. Since ASDs can be readily detected at an early age by ultrasound imaging, this research marks the first step towards developing drugs that might induce the closure of a detected defect without the need to resort to surgery, something that the BHF is proud to be funding."

The British Heart Foundation is a major funder and authority in cardiovascular research. For more information: [www.nottingham.ac.uk/biology](http://www.nottingham.ac.uk/biology) or [www.bhf.org.uk](http://www.bhf.org.uk)

**International Heart and Sports Medicine Experts Call for Cardiovascular Screening Programme for Athletes**

(Source: *European Heart Journal*) International heart and sports medicine experts have called for a Europe-wide cardiovascular screening programme for all young athletes before they are allowed to take part in competitive athletics.

The aim is to pick up potentially life-threatening problems that put young athletes at risk and to cut the numbers collapsing and dying while participating in competitive sport.

A European Society of Cardiology consensus report<sup>1</sup> was published 2 February 2005 in Europe's leading cardiology journal, the *European Heart Journal*,<sup>2</sup> recommends that every young athlete involved in organised sport has a rigorous physical examination, a detailed investigation of their personal and family medical history and, most importantly, a 12-lead ECG.

The report's writers believe that screening using ECG has the potential to cut sports-related cardiac deaths in Europe by 50%-70% if it can be implemented in every country.<sup>3</sup>

Lead author Dr. Domenico Corrado from the Departments of

Cardiology and Pathology at the University of Padova, Italy, said, "We know very little about the risk of sudden death associated with exercise in young competitors, so the benefits versus the hazards of sports activity pose a clinical dilemma. However, we know from a study in the Veneto region of Italy that adolescents and young adults involved in competitive sport had a two and a half times higher risk of sudden death. The young competitors who died suddenly were affected by silent cardiovascular diseases, predominantly cardiomyopathies, premature coronary artery disease and congenital coronary anomalies."

He said it was not sport that directly caused the deaths, but rather that it triggered cardiac arrest in athletes with underlying diseases predisposing them to life threatening ventricular arrhythmias.

The consensus group was also drawing on the results of Italy's 25-year experience of systematic prescreening in reaching its conclusions. Italy has a mandatory eligibility test involving nearly six million young people every year and the test leans heavily on the use of 12-lead ECG. In one 17-year study by the Center for Sports Medicine of Padova involving nearly 34,000 athletes under 35, over 1,000 were disqualified from competing on health grounds, 621 (1.8%) because the tests revealed relevant cardiovascular abnormalities.

Dr. Corrado said that in the USA young athletes had physical examinations and personal and family history investigations, but 12-lead ECG was done only at the doctor's discretion. The American Heart Association previously assumed that ECG would not be cost-effective for screening because of low specificity.<sup>4</sup>

"In fact, this is not the case," said Dr Corrado "The Italian screening method has proven to be more sensitive than the limited US protocol. ECG is abnormal in up to 95% of patients with hypertrophic cardiomyopathy (HCM), which is the leading cause of sudden death in an athlete. ECG abnormalities have also been documented in the majority of athletes who died from other arrhythmogenic heart muscle diseases."

Comparisons between findings in Italy where ECG is used and research in the USA showed a similar prevalence of HCM in non-sport sudden cardiac death, but a significant difference – 2% versus 24% – in sports-related cardiovascular events.

"This suggests we have selectively reduced sports-related sudden death from HCM because our system, using ECG, identifies vulnerable young people," said Dr Corrado.

He said that a number of the conditions now being picked up by ECG had only recently been discovered, so diagnosis was increasing. Researchers would shortly be examining the impact on mortality of the increased detection of potentially lethal problems.

It was harder to detect premature hardening of the coronary arteries or abnormalities in the coronary artery in young com-

petitors, he said, because baseline ECG signs of blood flow (ischaemic) problems were scarce. However, his research team had earlier reported that about a quarter of the young athletes who had died from coronary artery diseases had displayed warning signs or ECG abnormalities during screening that could raise suspicions of a cardiac disease.

The consensus group recommends that screening should start around the age of 12 to 14 and be repeated at least every two years. It should involve complete personal and family history, a physical examination that includes blood pressure measurement and a 12-lead ECG. It should be performed by a physician with specific training who could reliably identify clinical symptoms and signs associated with cardiovascular diseases responsible for exercise-related sudden death. Those who tested positive according to set criteria should be referred for more extensive tests, and if that confirmed suspicions, barred from competition and training.

"From all the evidence that we have from 25 years experience in Italy, we can state unequivocally that screening is warranted," said Dr. Corrado. "It is ethically and clinically justifiable to make every effort to recognize in good time the diseases that put these athletes and risk, and to reduce fatalities.

"Screening of large athletic populations will have significant socio-economic impact and its implementation across Europe will depend on the different socio-economic and cultural backgrounds as well as on the specific medical systems in place in different countries. However, experience in Italy indicates that the proposed screening design is made feasible because of the limited cost of 12-lead ECG in a mass screening setting.

"The cost in Italy of screening, aside from equipment and training, is around €30 including the ECG and is covered by the athlete or his team, except for the under 18s for whom there is National Health System support. Although the protocol is at present difficult to implement in all European countries, we hope that the successful Italian experience will lead to its widespread adoption under European regulations."

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2. Cardiovascular pre-participation screening of young competitive athletes for the prevention of sudden death: proposal for a common European protocol. *European Heart Journal*. doi:10.1093/eurheartj/ehi108.
3. There are no estimates of the prevalence of sports-related sudden cardiac death in European countries apart from Italy, where sports-related sudden death occurs in approximately 2 per 100,000 athletes per year. Screening programmes on selected groups of athletes are used in the majority of European countries, but the screening is never systematic as it is in Italy.

4. Cardiovascular preparticipation screening of competitive athletes. *Circulation* 1996;94:850-856.

### **UCSD Team Discovers Specialized, Rare Heart Stem Cells In Newborns, With Potential for Replacing Damaged Tissue**

The first evidence of cardiac progenitor cells - rare, specialized stem cells located in the newborn heart of rats, mice and humans - has been shown by researchers at the University of California, San Diego (UCSD) School of Medicine. The cells are capable of differentiation into fully mature heart tissue.

Called *isl1+* cells, these cardiac progenitor cells are stem cells that have been programmed to form heart muscle during fetal growth. Until this new discovery, the cells were thought to be absent after birth. However, the UCSD team discovered a small number of the specialized stem cells remained embedded in a region of the newborn heart called the atrium. They also determined that the cells could be expanded into millions of progenitor cells by growing them on a layer of neighboring heart cells called fibroblasts.

Published in the 10 February 2005 issue of the journal *Nature*, the research identified the *isl1+* progenitor cells in the tissue of newborn rats and mice, and then in heart tissue taken from five newborn human babies undergoing surgery for congenital heart defects.

Study author Sylvia Evans, PhD, a member of the UCSD Institute of Molecular Medicine (IMM) and professor of pharmacology, and co-first author Alessandra Moretti, PhD, IMM member, explained that the cells are programmed to become spontaneously beating cardiac muscle cells simply by exposure to other neighboring heart cells.

Since these rare cardiac progenitor cells are found in regions of the atrium that are normally discarded during routine cardiac surgery, the discovery raises the possibility that an individual could receive their own cardiac stem cells to correct a wide spectrum of pediatric cardiac diseases, according to co-first authors Moretti and Karl-Ludwig Laugwitz, MD, a Heisenberg-Scholar of the German Research Foundation.

"Conceptually, these cells could provide a cell-therapy based approach to pediatric cardiac disease, which is new for cardiology," said the study's senior author, Kenneth Chien, MD, PhD, director of the UCSD Institute of Molecular Medicine. "Traditionally, pediatric cardiologists and cardiac surgeons have relied on mechanical devices, human and synthetic tissue grafts, and artificial and animal derived valves to surgically repair heart defects. While progenitor cells won't grow a whole new heart, our research has shown that they can spontaneously become cells from specific parts of the heart by simple co-exposure to other heart

cells, which could augment existing surgical procedures. If the cells maintain pacemaker function when placed in the intact heart, they might serve as biological pacemakers for infants born with heart block, which could also be valuable."

After the *isl1+* cells were found in newborn rats, the UCSD team used sophisticated genetic methods to tag the progenitor cells in living embryonic tissue and in the newborn heart of mice. With these techniques, they were able to show that the *isl1+* progenitor cells were spontaneously able to form cardiac muscle tissue.

"Furthermore, the cardiac muscle cells formed were totally mature and had the complete array of function that one would expect in completely differentiated heart tissue," said the study's co-first author Jason Lam, Ph.D. candidate in the IMM. The cells exhibited contractility, pumping ability, the correct electrical physiology and normal heart structure. In addition, the progenitor cells coupled with neighboring cardiac muscle cells with resulting normal electrical heart beats.

"Another important discovery was the ability to expand the few cells found in a newborn heart, into millions of cells in lab culture dishes," Laugwitz said. "This implies that the *isl1+* cells potentially could be harvested from an individual's heart tissue, multiplied in a laboratory setting, then re-implanted into the patient. Furthermore, the developmental lineage marker which identifies undifferentiated cardiogenic precursors suggests the feasibility of isolating *isl1+* cardiac progenitors from mouse and human embryonic stem cell systems during cardiogenesis."

"We think that these cells normally play an important role in the remodeling of the heart after birth, when the newborn heart no longer relies upon the mother's circulation and oxygenation," Chien said. "We believe the *isl1+* progenitor cells are left over from fetal development so that they can insure the closure of any existing small heart defects and the formation of a completely mature heart in newborns."

The UCSD team noted in the *Nature* paper that the next research steps with the *isl1+* cells will be cellular transplantation in living animals to study their role in endogenous repair after cardiac injury.

In addition to Chien, Evans, Laugwitz, Moretti and Lam, study authors included Peter Gruber, MD, PhD, Children's Hospital of Philadelphia; Yinhong Chen, MD, PhD, Sarah Woodard, BS, Lizhu Lin, PhD, and Chen-Leng Cai, PhD, UCSD Institute of Molecular Medicine; Min Min Lu, PhD, Department of Medicine, University of Pennsylvania; and Michael Reth, PhD, Max-Planck Institut für Immunbiologie, Universität Freiburg, Freiburg, Germany. The research was supported by the National Institutes of Health, the Fondation Leducq, and the German Research Foundation. For more information: <http://health.ucsd.edu/news/>.



HIGHLIGHTS FROM CARDIOLOGY 2005: 8TH ANNUAL POST GRADUATE COURSE IN PEDIATRIC CARDIOVASCULAR DISEASE

By Gil Wernovsky, MD

Between February 16th and 20th, 2005, The Cardiac Center at The Children's Hospital of Philadelphia hosted Cardiology 2005: 8th Annual Post Graduate Course in Pediatric Cardiovascular Disease at Disney's Yacht and Beach Club Resorts in Orlando, Florida.

Just over 500 professionals in pediatric and adult congenital cardiovascular disease attended the meeting from 45 states and 15 countries, including attendees from Japan, Taiwan, Australia, North and South America, Scandinavia and Europe.

At the opening sessions, Professor Robert Anderson (London, England) and Peter Gruber, MD, PhD (Philadelphia) discussed "Conception and Formation", in which Professor Anderson used common congenital heart examples to review the evidence based approach to cardiac morphology. Dr. Gruber, who is both a pediatric cardiac surgeon and developmental biologist, spoke about new molecular concepts on how the heart forms. These opening talks were followed by a mini-symposium on fetal cardiovascular disease which featured Drs. Larry Rhodes, Jack Rychik, Meryl Cohen and Gil Wernovsky (all from Philadelphia) and Dr. Wayne Tworetzky (Boston). Up-to-date results on fetal cardiovascular physiology during non-cardiac fetal surgery were presented by Dr. Rychik, as well as the exciting information on fetal cardiovascular intervention by Dr.

Tworetzky (see also the December issue of Pediatric Cardiology Today).

Later on the opening day, Dr. Ed Walsh (Boston) updated the audience on risk stratification and current experience with ICD therapy for young patients with hypertrophic cardiomyopathy, Katie Dodds, RN, MSN, PNP (Philadelphia) discussed the current roles and future directions in advanced practice nursing, and Dr. Neil Wilson (London) reviewed the exciting work on percutaneous replacement of the pulmonary valve. Finally, a series of controversial topics were presented on the future roles of cardiac MRI (Dr. Tal Geva, Boston), 3D echo (Dr. Girish Shirali, Charleston), interventional catheterization (Dr. Evan Zahn, Miami) and surgical solutions to interventional 'headaches' (Dr. Thomas Spray, Philadelphia). The evening was capped off with a special networking and informational session for residents and fellows entering a career in pediatric cardiovascular medicine. Over 30 young physicians attended this reception, organized by Dr. Alan Friedman (Yale), Dr. Beth Ann Johnson (Milwaukee) and Dr. Geoffrey Bird (Philadelphia). Topics included work-life balance, academic vs. private practice cardiology, salaries, benefits and long term career planning....presented by many of the course faculty. This session will be repeated at Cardiology

2006 in Scottsdale, Arizona (February 8th-12th, 2006).

On February 17th, a series of plenary lectures were held to review important topics in genetics (Dr. Betsy Goldmuntz, Philadelphia), heart failure (Dr. Robert Shaddy, Salt Lake City), coagulation (Dr. David Jobes, Philadelphia), cardiac intensive care (Dr. Sarah



Figure 1. Dr. Edward Bove (left), Dr. Scott Bradley (center) and Dr. Robert Anderson (right) following the Pro-Con Debate. Dr. Bove was also the 6th Annual C. Walton Lillehei Lecturer.

Tabbutt, Philadelphia) and implantable cardiac support (Dr. Elizabeth Blume, Boston). A mini-symposium on issues related to patient safety, with sentinel events described by Dr. Peter Laussen (Boston) and Dr. James Jagers (Chapel Hill) followed. Later in the day, breakout sessions were held in electrophysiology, heart failure and transplantation, echocardiography, cardiovascular nursing, ECMO and pediatric perfusion. The highlight of the day was a spirited debate between Dr.

Edward Bove (Ann Arbor) and his former trainee, Dr. Scott Bradley (Charleston). The session was moderated by Professor Robert Anderson, and debated the topic: "Center Volume is the Best Indicator of Surgical Results." Dr. Bradley, who took the "con" position, informally "won" the debate based upon a poll of the audience participants.

Friday, February 18th featured a morning of breakout sessions on interventional catheterization, cardiovascular nursing, rare cardiovascular diseases, cardiac surgery and anesthesia, outpatient cardiology, and work-hour restrictions. Larger sessions were held after lunch on "Essential Principles of Inpatient Care" and "Data Management, Outcomes and Access to Cardiovascular Services: Past, Present and Future". For the first time, a wireless audience response system was used both in the morning for the cardiovascular nursing session, and during general

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***"Just over 500 professionals in pediatric and adult congenital cardiovascular disease attended the meeting from 45 states and 15 countries, including attendees from Japan, Taiwan, Australia, North and South America, Scandinavia and Europe."***

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sessions, moderated by Dr. Ross Ungerleider (Portland). During this session, 4 different viewpoints on the management of hypoplastic left heart syndrome were presented, and difficult questions were generated about the differences in management options presented to families based upon so-



*Figure 2. Dr. Roberto Canessa held the audience spellbound during his presentation of Human Groups in Crisis Situations.*

cial situations. The groups learned quite a bit about the variability in management patterns, and the data generated will be published in a supplement of *Cardiology in the Young* late in 2005. Dr. Ungerleider then gave insightful comments about team composition and factors that build harmony in pediatric cardiovascular programs. At the end of the day, Dr. Roberto Canessa (Montevideo, Uruguay) presented his experience of crashing in the Andes in 1972, and his epic struggle to survive over 2 months (which was highlighted in the movie: *Alive!*). It was a compelling and moving experience for all 500+ people in attendance. Ironically, on the same day that Dr. Canessa presented this experience in Orlando, an American hiker in the Andes found the wallet of one of Dr. Canessa's teammates that had been buried for over 32 years!

Saturday, February 19th featured the named lectures that have been a part of this meeting since shortly after its inception. Dr. Edward Bove gave the Lillehei lecture on "Current Strategies

for the Surgical Management of AV Discordant Hearts: Can Two Wrongs Make a Right?" followed by featured nursing lecturer Elisabeth C. Smith, RGN, RSCN (London), speaking on Contemporary and Collateral Nurse Leadership in the United Kingdom". Finally, Dr. Norman Silverman gave the 4th Annual William Rashkind Memorial Lecture on "Ebstein's Anomaly: From the Fetus to the Adult."

The day was rounded out with minisymposia on pediatric perfusion, cardiovascular nursing, and a comprehensive review of pulmonary atresia with intact ventricular septum. The session featured anatomical review by Dr. Anderson, imaging by Dr. Friedman, perioperative care by Dr. Wernovsky and Dr. Chitra Ravishankar (Philadelphia) and diagnostic and interventional catheterization by Dr. Jacqueline Kreutzer (Philadelphia). A superb summary of the surgical management options was followed by a thoughtful review of the literature by Dr. James Tweddell (Milwaukee), and emphasized the current limitations in evidence-based surgical decision making.



*Figure 3. Rashkind Lecturer Dr. Norman Silverman (Stanford, left) and featured Nursing Lecturer Elisabeth C. Smith, RGN, RSCN (London, right).*

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Figure 4. Dr. Edward Bove (right) receives the 6th Annual Lillehei Award from Dr. Thomas Spray (left), Executive Director of The Cardiac Center at The Children's Hospital of Philadelphia

Throughout the course, 40 posters of original research were on display, representing work from 28 different cardiovascular programs, and covering the disciplines of perioperative care, cardiovascular nursing, perfusion, imaging, catheterization, surgery and many others. Following a vote by the faculty, Dr. Sarah Tabbutt was awarded the 2nd Annual Outstanding Investigator Award for her work on a contemporary comparison of shunt type in the surgical reconstruction of hypoplastic left heart syndrome.

The meeting ended with a terrific mini-symposium on "The Lost Generation: Challenges Facing Teenagers and Young Adults with CHD (and their providers)", chaired by Dr. Gary Webb (Philadelphia). The session included topics such as exercise physiology, the psychological impact of pacemakers and other implantable devices and long-term outcomes in patients with transposition of the great arteries, tetralogy of Fallot, and various forms of single ventricle. Presenters included

Dr. Mitch Cohen (Phoenix), Dr. Paul Stephens and Dr. Bernard Clark (Philadelphia), Dr. James Quintensenza and Dr. Michael Parpard, CCP (Tampa), Dr. Elizabeth Blume and Susan M. Fernandes, MHP, PA-C (Boston), and Marion McRae RN, MScN, CCRN, CCN (C) (Toronto). The session also included a presentation on administrative considerations in the delivery of services to adults in pediatric hospitals by Dr. Steven Altschuler (President and Chief Executive Officer of The Children's Hospital of Philadelphia).

Cardiology 2006 will be held February 8-12 at the Hyatt Gainey Ranch in Scottsdale, Arizona, and will feature three broad themes: (1) State of the art management of the neonate and infant with complex congenital heart disease, (2) Clinical trials in pediatric cardiovascular disease, and (3) Ethical issues in the delivery of care to infants with cardiovascular disease. See [www.chop.edu/cardiology2006](http://www.chop.edu/cardiology2006) for details.

For comments to this article, send email to: [APRGW@CongenitalCardiologyToday.com](mailto:APRGW@CongenitalCardiologyToday.com)

~CCT~



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The course director is Horst Sievert, MD, PhD of the Cardiovascular Center Frankfurt, Frankfurt, Germany with co-directors Neil Wilson, MD of the Royal Hospital for Sick Children in Glasgow, UK and Shakeel A. Qureshi, MD, Paediatric Cardiology of Guy's Hospital in London, UK. There will be a number of well-known guest and local faculty. Over the three days there will be ten major sessions plus step-by-step live sessions including:

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- Obstruction and Occlusions
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## SCREENING FOR SUDDEN CARDIAC DEATH USING THE PRE-PARTICIPATION PHYSICAL EXAM

By Tiffany J. Riehle, MD and Robert C. Campbell, MD

A pre-participation physical exam (PPE) is an important, often overlooked medical tool. Since it is one of the few healthcare contacts a child or adolescent may have, it should be used as a screening tool to identify conditions that may compromise the athlete's performance or health.<sup>1</sup> The pediatrician's role is not only to appropriately clear or restrict young athletes, but also to identify potential risk factors for sudden cardiac death in the family.

Unfortunately, there is variability in PPE form content and personnel performing the pre-participation exam. In the US, only 40% of states adequately screen high school athletes for sudden cardiac death (SCD) and only 17% of high schools use forms that have all the elements of a cardiac exam recommended by the AHA2. The 3rd Edition of the PPE monograph, published in November 2004 by Mc-Graw Hill, can be purchased from any medical website or bookstore. It was co-authored or sponsored by several organizations: the American Academy of Pediatrics, the American Academy of Family Physicians, the American College of Sports Medicine, the American Medical Society for Sports Medicine, the American Orthopaedic Society for Sports Medicine, and the American Osteopathic Academy of Sports Medicine<sup>1</sup>. It is easy to use, inexpensive, and comprehensive. If a

standard PPE is done thoroughly and consistently, by qualified individuals, it should be an effective tool in identifying important life-threatening cardiovascular conditions.

A local news report of a young, apparently healthy child's death can devastate a community and fill the pediatrician's office with worried families. Nearly half of patients who

<u>Structural/Functional</u>	
Hypertrophic Cardiomyopathy (HCM) ♥	
Coronary Artery Anomalies	
Aortic Rupture / Marfan ♥	
Dilated Cardiomyopathy (DCM) ♥	
Myocarditis, Endocarditis	
Left Ventricular Outflow Tract Obstruction	
Mitral Valve Prolapse (MVP)	
Coronary Artery Atherosclerotic Disease ♥	
Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) ♥	
Post-operative Congenital Heart Disease	
<hr/>	
<u>Electrical</u>	
Long QT Syndrome (LQTS) ♥	
Short QT Syndrome ♥	
Wolff-Parkinson-White Syndrome (WPW)	
Brugada Syndrome ♥	
Congenital Heart Block	
Catecholaminergic Ventricular Tachycardia ♥	
<hr/>	
<u>Other</u>	
Primary Pulmonary Hypertension ♥	
Drugs, Stimulants	
Commotio Cordis	
♥ indicate familial or genetic causes	

Table 1. Causes of Sudden Cardiac Death.



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succumb to SCD are asymptomatic or had unappreciated symptoms. The first sign of a life-threatening disease is often sudden death. Thus, care providers should screen for SCD risk factors at any opportunity, not just at times of heightened awareness.

SCD occurs infrequently, with a likely underestimated range from 1:100,000 – 1:200,000 athletes each year.<sup>2,3</sup> Although rare, 85% of SCD is caused by primary cardiac diseases. Of those, 36% are due to hypertrophic cardiomyopathy and 19% due to coronary anomalies, usually left main coronary artery arising for the right sinus of Valsalva.<sup>4</sup> Table 1 lists several other important causes of SCD, including structural, functional, and electrical conditions. All of these conditions may ultimately lead to ventricular fibrillation and sudden death. As indicated, many of these causes are genetic or familial. Since many have no discernable physical exam findings, close attention to patient and family history is the only way to raise suspicion of risk for SCD.

A thorough family history, including source documentation, can be obtained at any time and updated periodically. Often, parents or children will not volunteer information unless specifically asked. Nearly all Americans believe family health knowledge is important but only 1/3 actively organize a family history.<sup>5</sup> The Surgeon General launched the Family History Initiative in November 2004; a computerized tool, found at [www.hhs.gov/familyhistory/](http://www.hhs.gov/familyhistory/), aids in organizing a family history, especially during family gatherings where multiple family members are present.

Questions 5-16 of the History Form and Table 4 in the 3rd Edition of the standard PPE monograph relate to cardio-

<b><u>Patient History</u></b>	
Have you ever passed out or nearly passed out DURING exercise?	
Have you ever passed out or nearly passed out AFTER exercise?	
Have you ever had discomfort, pain, or pressure in your chest DURING exercise?	
Does your heart race or skip beats DURING exercise?	
Has a doctor ever told you that you have high blood pressure, high cholesterol, a heart murmur, or a heart infection?	
Has a doctor ever ordered a test for your heart (for example, ECG, echocardiogram)?	
<b><u>“Tell Me About Any Family Members Who. . . “</u></b>	
Died for no apparent reason (SIDS, car accidents, drowning)	
Has a heart problem	
Died of heart problems or of sudden death before age 50	
Has had syncope (fainting) or pre-syncope (nearly fainted)	
Has had unexplained seizures	
Has had significant arrhythmias or a pacemaker	
Has any of the following genetic disorders:	
	Hypertrophic Cardiomyopathy (HCM)
	Dilated Cardiomyopathy (DCM)
	Marfan Syndrome
	Ehlers-Danlos Syndrome
	Arrhythmogenic Right Ventricular Cardiomyopathy
	Early coronary artery disease
	Coronary Artery Anatomical Anomalies
	Brugada Syndrome
	Long QT Syndrome (LQTS)
	Short QT Syndrome
	Primary Pulmonary Hypertension
<b><u>Physical Exam:</u></b>	
Abnormal blood pressure or pulses	
Heart murmur	
Arrhythmias	
May be NORMAL!!	

Table 2. Warning Signs for Sudden Cardiac Death.

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***“In the US, only 40% of states adequately screen high school athletes for sudden cardiac death (SCD) and only 17% of high schools use forms that have all the elements of a cardiac exam recommended by the AHA2.”***

vascular problems.<sup>1</sup> This information is summarized in Table 2 of this article. Insight may be gained from open-ended questions regarding SCD warning signs and causes beginning with “tell me about anyone who. . .” High-risk patients include those with syncope or chest pain during or after exercise, or anyone with certain genetic disorders. The AHA recommends further cardiovascular evaluation for anyone with a suspected or detected cardiac disease. Furthermore, any unexplained deaths in the family should prompt comprehensive SCD screening for all first-degree relatives. Just one index patient who had SCD, or who has warning signs, may uncover an entire family at risk for SCD. Thus, taking extra time and care to perform family and patient history intake, especially as part of the PPE, may save many lives.

In a large retrospective study of SCD in athletes, Maron et al. found that only 3% of the 115 athletes who had undergone a pre-participation exam were suspected of cardiovascular disease.<sup>4</sup> However, the forms or personnel were not standard. There has been no prospective evaluation of the

impact of a standard, properly conducted PPE on SCD. Although imperfect, the AHA considers it the best screening tool available for large population screening.<sup>2</sup>

Since the prevalence of SCD in the general population is low, screening for SCD is difficult. Routine EKG and echocardiograms as screening tools are not cost-effective and can lead to misdiagnoses.<sup>2</sup> Many causes of SCD evolve over time and patients may be silent carriers; thus, a negative test may not clear a child for life. A standard PPE format can be applied inexpensively for screening at any age; positive findings on family or patient history or exam then warrant referral for a more targeted comprehensive cardiac evaluation.

A clear, comprehensive PPE exists and is the current standard of care. We advocate the proper use of the 3rd Edition PPE monograph as an excellent, inexpensive first line screening for cardiac causes of SCD in children of all ages. No opportunity to screen for families at risk for SCD should be overlooked.

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## RESPIRATORY FAILURE AFTER INTRATHORACIC SURGERY

By Aphrodite Tzifa, MD and Shakeel Qureshi, MD

### Introduction

Respiratory failure can be defined as the inability to maintain adequate gas exchange to match metabolic requirements. It can be characterized by abnormalities in oxygenation (hypoxic respiratory failure) or abnormal carbon dioxide clearance (hypercapnic respiratory failure). Categorization of respiratory failure into these two groups is arbitrary and obtained from the arterial blood gases. The cause of blood gas abnormalities following intrathoracic surgery (ITS) may vary and may be influenced by pre-existing lung disease or arise in the perioperative period.

### Hypoxic Respiratory Failure

The diagnosis of hypoxic respiratory failure is arbitrarily defined in terms of arterial blood gas abnormalities when PaO<sub>2</sub> <60 mmHg or oxygen saturations below 90%. Hypoxia may be caused by loss of alveolar surface area (e.g. pulmonary edema, ARDS) or abnormalities in ventilation/perfusion such as pulmonary embolism and atelectasis. These two mechanisms frequently co-exist. In addition patients undergoing ITS may have underlying cardiac disease which can lead to hypoxia secondary to intracardiac or intrapulmonary right to left shunts.

#### a) Loss of alveolar surface area

Conditions which reduce the surface

area of alveoli may reduce efficacy of gas exchange. This may include disease of the lung interstitial space, alveolar wall or alveolar space such as pulmonary edema or hemorrhage. Involvement of the alveolar air spaces can be focal, and confined to one or two lobes of the lung as in pneumonic consolidation/atelectasis or at the extreme end of the spectrum may involve four quadrant lung disease as in ARDS. The incidence of the latter, following cardiac surgery,

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*“The cause of blood gas abnormalities following intrathoracic surgery (ITS) may vary and may be influenced by pre-existing lung disease or arise in the perioperative period.”*

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is low (0.4%) but carries a high mortality (15%). Cardiogenic pulmonary edema may resemble ARDS but is associated with a high pulmonary wedge pressure (>18 mmHg). Cardiac conditions commonly associated with pulmonary edema in the postoperative period include residual VSDs, residual mitral valve stenosis or regurgitation, or conditions associated with excessive pulmonary blood flow such as large aortopulmonary shunts and inadequate PA banding. Treatment of cardiogenic pulmonary edema involves use of diuretics, inotropic support and correction of the underlying cardiac anomaly.

#### b) Ventilation perfusion mismatch

Ventilation and perfusion are not distributed homogeneously throughout the lung as the gravity effect causes increase of both ventilation and perfusion from the top of the lung to the bottom. However, blood flow increases more rapidly so that the V/Q ratio is inhomogeneous across the lung. Optimal gas exchange occurs when ventilation to perfusion matching occurs in every single alveoli.

In the postoperative patient V/Q mismatch may be due to pulmonary, pulmonary vascular or cardiac origin.

### Pulmonary causes

During V/Q mismatch areas of the lung with low V/Q ratios contribute blood with an abnormally low PO<sub>2</sub> to the pulmonary venous and systemic arterial blood and impair the removal of carbon dioxide.

Lobar atelectasis is a frequent cause of hypoxia following ITS. In ventilated patients it most often affects the right upper lobe and usually improves spontaneously. Risk factors for atelectasis include use of high inspired oxygen concentration (resorption atelectasis), underlying lung disease (bronchiectasis, tracheobronchomalacia) and anatomical lesions which obstruct the lumen of the airways such as vascular rings, dilated pulmonary arteries and enlarged left atrium.

Nosocomial pneumonia is a common complication in the patients who have undergone intrathoracic surgery and already have somewhat compromised



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cardiopulmonary function or prosthetic materials such as prosthetic valves, stents or coils inserted. Unusual or atypical organisms (acinetobacter, fungi) may occur in children with immune compromise (polysplenia in the left atrial isomeric patients, 22q deletion etc.).

**Pulmonary Vascular Causes**

Pulmonary hypertension can complicate the postoperative course of patients who had long increased pulmonary flow prior to their surgical repair. Commonly such patients are the ones with large VSDs, AVSDs, transposition of the great arteries with VSD, common arterial trunk, aortopulmonary windows, long standing large arterial ducts etc.

Increased pulmonary flow and the development of pulmonary hypertension lead to decreased lung compliance, and contribute to the development of atelectasis, with the latter potentially leading to loss of lung volume and increase of the intrapulmonary shunt.

The onset of PHT episodes post-operatively may be acute and are characterized by severe desaturations with hypotension and associated tachycardia.

The management of postoperative pulmonary hypertension involves increase of FiO<sub>2</sub>, hyperventilation with development of respiratory alkalosis, adequate sedation, neuromuscular paralysis and the use of medical therapy in the form of nitric oxide, MgSO<sub>4</sub> infusion, calcium channel blockers, phosphodiesterase inhibitors (sildenafil, enoximone) etc.

Pulmonary embolism is quite rare in the pediatric population but may occur in the postoperative patients with

complex heart disease, particularly with univentricular heart physiology. The clinical diagnosis of pulmonary embolism is highly non-specific and perfusion lung scanning will be required to confirm the diagnosis.

**c) Cardiac causes**

Hypoxic respiratory failure may be encountered in patients with right to left intracardiac shunts (at atrial or ventricular level) when the pulmonary artery pressure is elevated.

In particular, children with univentricular heart physiology are often desaturated after the Glenn operation for a few days until the pulmonary vasculature readjusts to the new circulation. Similarly, children after a fenestrated Fontan operation are occasionally hypoxic secondary to a right to left shunt that occurs through the fenestration when the pulmonary vascular resistance is elevated. Both groups of patients would benefit from early extubation, provided that the cardiac function is good.

Further investigations, such as cardiac catheterization or MRI, might be warranted in these patients in order to identify hemodynamically significant collateral vessels or baffle leaks that result in right to left shunts.

**Hypercarbic respiratory failure**

It may occur as a result of alveolar hypoventilation, airway obstruction, poor respiratory drive, acute muscle weakness or paralysis.

**Upper airway obstruction**

It is the most common postoperative airway problem, occurring in about 1% of the pediatric population. The length of ventilation, degree of air-leak around the ETT, changes of neck position, coughing whilst intu-

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bated and patient's age between 1-4 years are the main risk factors. It very rarely leads to irreversible respiratory insufficiency, though less rarely it may lead to the need for re-intubation.

**Central airway compression**

Vascular rings, dilated pulmonary arteries, left atrial enlargement and cardiomegaly may cause long term tracheal compression resulting in poor tracheal development. Tracheo-bronchomalacia may manifest as persistent wheeze postoperatively and can often go unrecognised for a long time until the patient eventually fails extubation repeatedly and chronic respiratory failure settles in.

The left lateral trachea, the superior aspect of the left mainstem bronchus, and the junction of the right intermediate bronchus with the right middle lobe bronchus are particularly vulnerable sites.

The diagnosis is made by the use of flexible bronchoscopy, where extrinsic compression becomes evident. These patients not uncommonly require tracheostomy for chronic mechanical ventilatory support and occasionally stenting of their airways or further surgical relief of the vascular airway compression (aortopexy, pulmonary artery plication) before extubation can be accomplished.

Furthermore, attention should be paid to the patient who fails extubation after aortic arch repair for aortic arch interruption or coarctation. Lower airway obstruction can arise from manipulation of the descending aorta during surgery and early recognition is crucial for the further management of the patient.

**Pleural effusions / Chylothorax**

They can potentially delay extubation or cause respiratory failure in the extubated patient. Common causes of pleural effusions include postoperative fluid overload, increased central venous pressure, as in patients with Fontan circulation, serous fluid leakage from extracardiac shunts, and chylothorax secondary to thoracic duct damage.

Chylothorax occurs in approximately 1% of cardiac operations, particularly extrapericardial procedures, the Fontan operation and Tetralogy of Fallot repair. It usually responds to conservative management with free drainage and exclusive medium chain triglyceride diet (MCT diet). More rarely total parenteral nutrition (TPN) or surgical thoracic duct ligation is required.

**Pneumothorax**

It is one of the most common causes of acute postoperative respiratory failure and should be immediately suspected and treated in all cases of acute respiratory deterioration of a postoperative patient. It develops most commonly secondary to partially obstructed endotracheal tube, barotrauma related to mechanical ventilation and chest drain obstruction.

**Diaphragmatic Paralysis**

It has been noted to be more frequent after operations that are conducted in the vicinity of the aortopulmonary trunk (arterial switch operation, coarctation repair, tetralogy of Fallot, augmentation of pulmonary artery etc).

Older children generally tolerate unilateral paralysis quite well, whereas infants are at particular risk of developing respiratory failure. This is attributed to the high compliance of

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***“Pulmonary hypertension can complicate the postoperative course of patients who had long increased pulmonary flow prior to their surgical repair.”***

---

their chest wall, and lack of respiratory reserve, particularly postoperatively.

Failure to wean the patient off the ventilator in the absence of other respiratory or cardiac causes should raise the suspicion of diaphragmatic palsy. The patient will typically tolerate low rate or continuous positive airway pressure (CPAP) well, but would fail extubation.

The definitive diagnosis is made by ultrasound or fluoroscopy of the diaphragms during spontaneous breathing, when paradoxical motion of the paralysed diaphragm is confirmed. Temporary diaphragmatic paresis may last up to 2 months. Assisted ventilation or CPAP should be applied until the diaphragmatic function recovers. However, if it persists, plication of the affected paralysed hemidiaphragm may be necessary to wean the patient from mechanical ventilation.

**Conditions Affecting Central Respiratory Drive**

These would include brain stem infarction or hemorrhage as a result of hypoxic ischemic injury post by-pass as well as excessive sedation with neuromuscular blockade.

**Postoperative respiratory failure of**



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### cardiac origin

Children with congenital heart disease often suffer from pulmonary congestion, edema and recurrent chest infections preoperatively. As a result, they are more likely to require a more prolonged ventilatory course postoperatively.

Cardiac causes of respiratory failure in the setting of operated congenital heart disease include:

a. residual left to right shunts, b. obstructive lesions c. excessive pulmonary flow and d. low cardiac output status.

#### Residual left to right shunt

These mainly include residual hemodynamically significant VSDs.

Diuretic therapy, positive pressure ventilation and inotropic support may be necessary to wean the child off the ventilator, whereas occasionally reoperation is required.

#### Obstructive lesions

Left sided obstructive lesions such as residual mitral or aortic valve stenosis, residual aortic arch obstruction after coarctation repair or Norwood I operation, or obstructed pulmonary venous drainage may lead to respiratory failure and poor cardiac output.

Similarly, right sided obstructive lesions, such as SVC obstruction after the Fontan operation or significant residual pulmonary stenosis after Fallot repair can result in pulmonary dysfunction and impaired cardiac output.

Correction of the underlying cause is imperative before successful extubation is achieved.

#### Excessive pulmonary flow

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***“In the fragile postoperative period early recognition of the causes of respiratory failure and clinical destabilization are crucial.”***

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Postoperative patients with large Blalock-Taussig or central aortopulmonary shunts may develop high pulmonary flow and pulmonary edema, particularly when the arterial duct has been left patent, or after concurrent atrial septectomy. Aggressive diuresis, whilst maintaining adequate cardiac output, is sometimes necessary in order to wean from mechanical ventilation.

#### Low cardiac output

Respiratory failure resulting from myocardial failure is common after intracardiac operations, particularly after prolonged bypass periods. As a result, pulmonary congestion develops with subsequent hypoxemia and hypercarbia. Inotropic support, vasoactive drugs and optimization of preload are useful maneuvers to assist the failing heart and improve the respiratory function.

#### Conclusions

Acute respiratory insufficiency is commonly seen in patients that have undergone intrathoracic surgery. Direct lung injury, bypass lung injury, adverse cardiorespiratory interactions, structural residual obstructions, infection etc. may complicate their postoperative course.

In the fragile postoperative period early recognition of the causes of respiratory failure and clinical destabilization are crucial. Increasing ex-

perience of managing these patients in the Paediatric Intensive Care setting has resulted in decreased mortality and morbidity and newer modes of ventilatory support have optimised their management and shortened their ICU stay.

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Selected questions and answers may be published in upcoming issues. Names will be withheld upon request.

## CONGENITAL CARDIOVASCULAR INTERVENTIONAL STUDY CONSORTIUM (CCISC)

By Tom Forbes, MD

At the end of 2003, sixteen institutions in North America and Europe participated in a retrospective review of results in stenting native and recurrent coarctation of the aorta. It became evident very early on, that there was no uniform consensus regarding how these patients should be followed up after the intervention. Some institutions would obtain a CT scan prior to discharge, at 6-8 months, and two to three years following the intervention. Other institutions would obtain an echocardiographic study once every two years. The group felt that a more organized and uniform approach in following up these patients would be helpful in providing important data. In addition, it also became apparent that this same approach should be performed in planning the intervention. This was the impetus for starting a consortium group called the Congenital Cardiovascular Interventional Study Consortium (CCISC). Since our first meeting at the SCAI Scientific Session in April of 2004, the number of participating institutions has expanded to over 40 in North America and Western Europe. The primary purpose of the group is to evaluate prospectively interventional procedures that are commonly performed in the catheterization lab. The first study to be undertaken is "Comparison of stent versus balloon angioplasty versus surgical repair of native and recurrent coarctation of the aorta in children and adults". In developing this study, the group came to a

consensus on what information needed to be collected and what type and frequency of imaging would be required to answer the questions and concerns we had in treatment of native and recurrent coarctation of the aorta. The patient follow up will last five years following the initial intervention. Our aim is to identify treatments for coarctation of the aorta which have optimal efficacy and safety.

A unique aspect of CCISC is that initially we will also have industry support. For the first time, participating institutions will be able to receive remuneration for participating in the consortium. The companies that have agreed to participate to date are: Siemens Corp, Cook Corp, NuMED Inc., B.Braun Medical Corp, Bayliss Medical Company, Inc. and Cordis Corp. This is in contrast to other registries, which require participating institutions to pay for participation to enter data sets into the registry. We are excited that industry is willing to step forward and assist us in this project. Though the CCISC will be initially funded by industry, the research undertaken by the Consortium will not be driven by industry. Protocols will be submitted by participating members of the Consortium to the scientific committee. This committee will select projects for the Consortium based on merit and importance. The amount of remuneration for participation will not completely cover the actual costs of obtaining and submitting the data. The main reason, and possibly the only reason, why this Consortium will be successful is that all participating institutions understand the

importance of gathering results of interventional procedures in an organized, prospective manner. In doing so, we hope to be better able to answer critical questions which will ultimately improve treatment in the patients we serve. Other registries such as the Congenital Heart Surgical Society (CHSS) and the Pediatric Heart Transplantation registry have been successful in giving us a better understanding in some of the issues we face in pediatric cardiology. Only by prospectively approaching issues in a focused manner, will we be able to answer questions in the interventional cardiology arena. The Consortium meets twice yearly at the SCAI and PICS meetings, with the next meeting planned for May 4, 2005 at Ponte Vedra, Florida. Any institution interested in participating is welcome to e-mail Tom Forbes, MD at [tforbes@dmc.org](mailto:tforbes@dmc.org).

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