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9008 Copenhaver Drive, Ste. M
Potomac, MD 20854 USA
www.CongenitalCardiologyToday.com

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.[1] 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.[2,3] Most interventional or surgical procedures are simply unaffordable to most families in the absence of some form of heath 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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ploying 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. 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.[4]

"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."

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

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.

"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."

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-

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

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 effected 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 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."

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
Epidemiological Significance	● Global disease burden of congenital heart disease ● The burden in developing world ● Proportional infant and childhood mortality for CHD ● Relative significance of CHD as compared to acquired heart disease such as rheumatic heart disease ● Planning epidemiologic surveys in the developing world
Establishing a Center for Comprehensive Care of Patients with Congenital Heart Disease	● Personnel ● Material resources; infrastructure ● Support services and systems ● Ensuring economic sustainability of the program ● Overcoming philosophical hurdles (teamwork, multi-specialty institution-based care etc., infection control, etc.)
Resources	● Number of centers for comprehensive pediatric heart care in the developing world ● Proportion of patients with CHD are likely to receive care in these centers ● Health insurance for patients with CHD ● Institutional and governmental support for individual patients
Special Challenges of the CHD Patient Population in the Developing World	● Malnutrition ● Infections: Neonatal sepsis; lung infections ● Consequences of late presentation: ● Pulmonary hypertension ● Neurological complications and major neuro-developmental sequelae ● Cardiac consequences: ventricular dysfunction, atrial enlargement and arrhythmias
Managing CHD Patients in Developing Nations: Practicalities	● Prioritization with respect to lesions ● Cost-effective strategies for cardiac catheterization and interventions ● Cost-effective strategies for congenital heart surgery and intensive care ● The challenge of infection control

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

Broad Category	Specific Issues
Inexpensive and Indigenous Technology	● Occlusive Devices ● Mechanical heart valves ● Disposables for cardiac catheterization, congenital heart surgery and intensive care ● Echocardiography machines ● Cardiac catheterization laboratories
Training	● Pediatric Cardiology ● Pediatric Cardiac Surgery ● Anesthesiology and intensive care
Improving Awareness Through Education	● Pediatricians and primary care physicians ● Adult cardiologists involved in the care of patients with CHD ● Obstetricians, radiologists performing prenatal ultrasound ● General public
Formulating Health Policies	● Defining the relative priority for congenital heart disease ● Regulatory bodies on medical education ● Policies on health insurance coverage ● Support to institutions with pediatric heart programs ● Research and education grants ● Support for individual patients
Collaboration with Centers in the Developed World	● Exchange programs for training ● Visits of teams from institutions in the developed world
Ethical Issues	● Bridging the gap between what is possible and what is feasible ● Managing "complex" defects ● Facilitating decision-making for families

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.

Acknowledgement

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For comments to this article, send email to: APRRKK@CongenitalCardiologyToday.com

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R. Krishna Kumar, MD, DM, FACC
Chief Pediatric Cardiologist
Amrita Institute of Medical Sciences and Research Center
Kochi, 682026, Kerala, India

Phone: 91-484-2801234
Fax: 91-484-2802020

rkrishnakumar@aimshospital.org

FIELD UPDATE #3: CONVOY TO BAGHDAD

By Major Joseph D'Angelo, MD

Major Joseph D'Angelo, MD is a pediatric cardiologist from Hawaii, who is currently serving a tour of duty as a combat physician in Iraq. This field update was originally sent to his family and friends on Feb. 17, 2005. It is edited here for space and reprinted with his permission.

Aloha from Baghdad, Iraq. I am finally where I am supposed to be, but not yet doing what I came here for. I have just done a little medicine at the Camp New York TMC (Troop Medical Clinic), and will likely do a bit more here at Camp Victory South until the unit we are replacing moves out and we take over, in a few weeks.

Convoy

The biggest and best news is that I survived the convoy. For the first time I have ever said "I survived..." something, I mean it literally, not figuratively. We had been receiving Convoy Operations and IED (Improvised Explosive Device) briefings in Hawaii, Texas, and at two or three different camps in Kuwait. We not only knew all the techniques the terrorists/insurgents had utilized, but which were most current and which they had abandoned. For example, a recent one was dropping IEDs from overpasses on convoys passing below. We had received the most current briefings about where recent IED and Small Arms Fire attacks had occurred. We knew there had been 7 or 8 attacks along our route over the past few weeks. They had been slowing down, but we expected it was to "save up" for the announcement of the election results, at which time it was predicted more violence would be unleashed. (I was not too up on the news, but I think I heard the day before we left there were multiple attacks in Baghdad to the tune of over 30 casualties). So, we were about as prepared as one could be.

Although I was interested in the experience, I was not looking forward to the duration of the trip (3 days) and I had preferred the thought of going by my third favorite mode of travel: helicopter. As it turns out, the remainder of the unit flew by plane, so it was just as well I drove. I also questioned the wisdom of sending a physician by such a dangerous mode of travel, and will raise this question for the docs that follow me, but did not want to make much of it now.

I have to say, it was a very strenuous trip. I was never much for long drives, such as the Southern-Northern California drive, or other lengthy road-trips. I generally found them uncomfortable and boring, unless there was stellar scenery. This was a different experience altogether on both accounts. Now that I ride a motorcycle, though, even though it gets physically tiring, the thought of doing this on a bike is a bit intriguing. Hmm, me in full leathers, body armor and fully armed, on a Kawasaki Vulcan 2000; now that would have been a convoy to remember, definitely right out of Mad Max! Somehow, I do not think I could have gotten the okay for that one, even with my rebellious nature. As for the discomfort, although it was so, I think it was more the claustrophobic and "sitting duck" thoughts I had when I first caught sight of where I would be: in the raised, middle seat of a cab of a truck, the kind with the windshield flush with the front of the vehicle (engine under the cab, not in front of). Given my height with an inch of head clearance, I was more concerned about getting a spinal compression injury.

In addition, as it turned out, ours was the first truck in the convoy (following anywhere from 1 to 5 hummers in various stages of armor and armament). At all times we were in full "battle rattle:" that is the body armor with all its accoutrements (mostly ammo), a 12 pound or so Kevlar helmet, kneepads, whatever "snivel-gear" (cold weather clothes) we had, goggles and ear plugs (my ears still ache from those). I also sat with my M4 rifle in my lap, and I did not like that my 9mm was not so easily accessible in my thigh holster, so I "tossed" it in my junk box, however, since it kept getting tossed about by either me searching for things, or just the motion of the truck, I thought it best in the final stretch to go back to wearing it. I just kept checking the safety frequently. I also kept up on the GPS and for about half the trip, I took over "commo" (communications), so I had a radio headset crammed under my helmet strap against me ear. Despite all this, and the various aches and pains, and the cramped quarters and occasional Charlie-horse, I was surprised I did not have more discomfort, and pleased to not have that spinal injury.

As for the boring, it was a most unique experience and anything but. As I said, it was very strenuous, but not just physically as noted above, but mentally. I cannot recall a time of such extreme vigilance over such a long period of time. Sure, I have to admit I dosed off briefly for a moment or two, as did my TC (Truck Commander), the guy who usually does the radio and navigation), but at least the driver stayed awake. Even though I was an officer and am usually spared guard duty, my TC and I traded off so the driver could sleep uninterrupted the entire night (which usually ended at about 3 am).

Time for a digression. I should introduce the cast of characters: there was a driver who is a specialist from my company whose specialty I was unaware, except afterward I was impressed that he was

great at the wheel of the rig and handled that truck like a race car driver. The whole undertaking reminded us of a Mad Max movie, and our driver was up to the task. Riding shotgun, although only figuratively, since it was actually a crew-served weapon (machine gun) that he had pointing out his window the entire ride, was our TC, although as I said, I liked being on commo and the last day, he asked if I could do it so he could focus on the ride. He is also a specialist (one rank above private first class, and one below a corporal), and he appears to be an excellent mechanic, caring for our ride like a baby. He also had a nice anal retentive streak akin to my own, preparing the cab for the ride, with our personal items and goodies (and ammo) well placed. Before we set out, he admitted to me "I don't know about you, but I'm scared." I was as reassuring as I could be and along the ride, I was quite impressed with him, representing the best in uniform now and in the past.

As for me, as I experienced before on drills, I could have felt like the payload, the valued but useless cargo; I guess I could have just sat there and allowed myself to go along for the ride. After all, not much is expected in the way of soldiering from the docs, so usually I have to say, "I want to do everything that you guys do." Anyway, I ended up taking a lot of time the day before plugging in grid coordinates into my GPS, and from what I could observe, was probably one of the best self-prepared TCs on the ride. I paid attention to where the "hotspots" were, so I could tell my crew to stay frosty and extra vigilant when we came upon a known area for IEDs or small arms fire. As the doc, I am usually protective about my soldiers, checking if they are okay, etc. It was a nice change on the morning of the last day when my TC, a specialist I way outranked, told me how it was going to go down if we took casualties... that is I was not to leave the safety of the vehicle and my security guard but to sit tight while the medics handled things until the area was secured. This makes sense anyway, because even the most experienced trauma doc is limited by the resources and on a convoy, basic medic/EMT procedures are all there is; the key is to call in the quick medivac. This is another reason why the docs are more cargo than actual practitioners, but I am sure any doc along for the ride would have a hard time just sitting by with injured around. Combat medicine is a whole different ball game though, and one of the first things we learned is the most important medicine is not the ABCs (airway, breathing and circulation of your basic CPR) or even stopping the bleeding (which takes precedence in combat even above the ABCs compared to civilian medicine), but superior firepower. You have to control and secure any area with casualties with that before you can start doing any care, otherwise the valued medic is at risk. Anyway, it was nice to feel taken care of by the very soldiers who I take care of in a different way.

Back to the vigilance. That was the word for the trip, and there are certain experiences in life that define words and concepts and for the first time I think I really understand to my core what vigilance means. We were alert, observant the entire time (except for those previously mentioned periods of exhaustion drop-off), constantly scanning from the up-close to the horizon. Everything was a potential threat: hills and berms along the side of the road, road debris, animal carcasses, oncoming traffic, pedestrians, bridges, any irregularity in the road or terrain, merging traffic, roadside structures, children (yes, sadly the tactics of terrorists know no bounds and are only limited by their imagination and I have learned there is NO lower level to which they can stoop, but beyond that it is all I will say on that matter). All of these things have been used as sources of attack. So, for roughly 8 hours each day we stared, scanned, observed and kept vigilant.

There was another aspect of the mental strain. That was the constant scenarios that ran through our minds; though we did not speak too often of it, I know we were constantly running the "what if..." in our

The Convoy Medics of the 2-299th Infantry Regiment.

thoughts. For a while, the most significant "ethical" question dominating my thoughts was at what point would I fire through the windshield.

The trip was not without some pleasant moments. As we got closer to our daily destinations, where the area was relatively secured, we took collective sighs of relief, cleared our weapons, and even left our vehicles for a while. I had a few nice experiences with two of my favorite things: birds and children, both of which I took plenty of pictures and some video. We even passed a few Italians in their convoy, as well as the Brits and a few other coalition forces.

Birds

As I said previously, where we were in Kuwait seemed relatively devoid of life. However, the more we moved closer to Baghdad and the hot zones, the more prolific (relatively speaking) the birdlife came. There were many ponds along the side of the road, and such watered areas are some of the best places to view birds. I do not wish to bore my non-birder readers, so here is just a quick list of some of what I saw: shorebirds of a few varieties, coots (like our American Coot, I believe this was the Eurasian Coot), stilts (also like our version, also I think called a Eurasian stilt... please feel free to correct me), swallows, terns and/or gulls, and a few small LBB (little brown birds), crows, and rock doves ("pigeons"). The star, however, was

a large bird I spotted at one of our stops. Those of you who "know" me could easily picture me, hopping out of the truck in this convoy having to wait again for hours (in a relatively safe zone), strolling over to the side of the road, binocs and camera in hand, checking out the birds. The only thing unfamiliar would be my 250+ pound total weight in battle rattle. Anyway, this star was a relatively large bird, with beautiful black and white plumage. At first I thought it might have been a Night Heron-like bird, but after it began to call, there was no mistaking it: it was some species of crow. I hope to look it up later. It was a nice moment away from the thoughts of war travel.

Children

Also at a relatively controlled stop, there were some Iraqi children along side of the road (actually, sporadically along the way we saw children, most waving, some gesturing to their mouths or for us to honk, and one naughty little girl flipping us off). These particular kids were playing on the other side of the concertina wire (large coils of barbed wire). One in particular, about a 5 year old, particularly drew my attention. I took a video clip of him loping around with his buds.

Unfortunately, I missed catching him doing a cute little dance moments before. Anyway, they just walked around, exploring stuff. I did not really want their attention, but eventually I caught his eye. Keep in mind, there was no language commonality at all (despite the booklet and CD provided courtesy of the Army), I just could not keep up on the language. Of course, it was not a typical language crash course... several years of Italian and one each of Spanish and French, and I never learned to say "stop or I'll shoot" at all, let alone in the first lesson. Sigh. The sad realities of human conflict. So, I started yelling to him, "you have nice birds here" and pointing to the birds. He looked back, but I think it was an older friend who actually understood. He said a word (too many syllables for me) I think for the birds, but he did not repeat it. We just did that classic talking to each other in our own language bit. The older boy then started to pick up rocks. Ah, children: you gotta love 'em. I laughed, but yelled (in a nice way), "no, you rascals, don't throw rocks at the birds." He understood and put the rocks down. Cute kids.

Astronomy

As I said in the last Update, the stars are the same, but I did have some cool experiences our second night. No sooner did I hop out of the truck, than I spotted a bright flash (only about as so as a bright star) and then again a few times on a straight trajectory. I thought it might have been a satellite, but it seemed too bright and was flashing. Maybe a plane, but its flash was too spaced out. I did shortly after see a real satellite tracking across the sky. Finally, since the ground was more gravelly than where I slept the night before in my sleeping bag, I chose to sleep on top of the steel container on the back of our truck. I was pretty high up (and with no edges... Mom, I can just hear you: "what the heck are you doing up there, you could fall off!" Good thing I do not roll around much at night) and had a nice view of the sky. Orion was obvious, but as usual I was disappointed I did not know more of the constellations.

Well, it is a real chore to put my uniform back on (since I am only in my PT (physical training) shorts and shirt), but I do not want to go to lunch in this, because not only are we required to carry our weapons, but we have to have ammo as well (and strapping my 9 to my bare thigh in my workout clothes to eat is definitely going too far! As Bilbo Baggins said, just before he bailed out of the Battle of the 5 Armies, "enough is enough." (Actually, I ended up neither wanting to change nor strap it on, so I just slung it over my shoulder.) Oh, that Tolkien reference reminds me of another thought I have experienced from that most inspirational of movies/books. In the 3rd, The Return of the King Pippin speaks to Gandalf about being less bothered by the thought of a battle that he cannot avoid, but waiting on the verge of it unable to do much. I thought of that a lot the days and nights before the convoy. I would like to think that in addition to good fortune and the prayers of loved ones, that by our own preparation, diligence, vigilance and sheer show of firepower, we were able to avoid an attack. Well, I have to get to lunch or miss it, and then clean my weapons. More on Victory South, Baghdad, and the reports of the mortars, later. Suffice it to say, I hear my building holds up nicely with the attacks, and from what I am told most of the mortars are of the kind too powerful and fly right over, or not enough so and hit the other camps short of ours. That whole last sentence sounds pretty darn weird and not something I could have ever imagined writing. But, I get a fair number of those fleeting thoughts lately: "what the heck am I doing here?" More on that later, too.

Aloha,

Joseph

P.S. I am still bald. Shaved it myself, already, twice. It is still going to take a little getting used to. When I used to scratch my head, there was some hair there to protect my tender scalp... not now . Always wearing cover (hats) in uniform, I finally got some sun on it today on my walk to lunch. I remind myself of Brando and Dr. Evil. Well, that is it from the End of the World Opposite to Paradise (in which lies my home); ironically, the Garden of Eden was supposed to be around here somewhere. Hmm, now probably buried under sand.

Major Joseph D'Angelo, MD
HHC, 2nd Bn-299th INF
Camp Victory, South
APO AE 09364

hiheart@shaka.com

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 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 Jaggers (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.

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.

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 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 social 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!

"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."

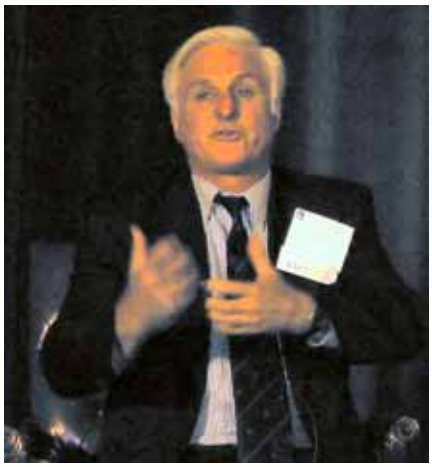

Figure 2. Dr. Roberto Canessa held the audience spellbound during his presentation of Human Groups in Crisis Situations.

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 mini-symposia 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).

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 for details.

For comments to this article, send email to: APRGW@CongenitalCardiologyToday.com

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Gil Wernovsky, MD, FACC, FAAP
Staff Cardiologist, Cardiac Intensive Care Unit
Director, Program Development; The Cardiac Center
Professor of Pediatrics at The Children's Hospital of Philadelphia
University of Pennsylvania School of Medicine

WERNOVSKY@email.chop.edu

JUNE CONGRESS FOCUS

8th International Workshop in Congenital and Structural Heart Disease
with Live Case Demonstrations and Hands-on Workshops
June 16-18, 2005;
Frankfurt, Germany
www.chd-workshop.org

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:

- Imaging
- ASD Closure
- Obstruction and Occlusions
- Step-by-Step and Tips and Tricks in PFO and LAA closures
- LAA Closure for Prevention of Cardioembolic Stroke
- Ventricular Septal Defects
- Complications – Prevention and Management
- And more...

In addition there will be Hands-on workshop in the catheterization lab at the CardioVascular Center Frankfurt.

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.[1] 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 Medicine1. 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

Structural/Functional
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
Electrical
Long QT Syndrome (LQTS) ♥
Short QT Syndrome ♥
Wolff-Parkinson-White Syndrome (WPW)
Brugada Syndrome ♥
Congenital Heart Block
Catecholaminergic Ventricular Tachycardia ♥
Other
Primary Pulmonary Hypertension ♥
Drugs, Stimulants
Commotio Cordis
♥ *indicate familial or genetic causes*

Table 1. Causes of Sudden Cardiac Death.

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.[2,3] 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.[4] 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.[5] The Surgeon General launched the Family History Initiative in November 2004; a computerized tool, found at 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-

Patient History
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)?
"Tell Me About Any Family Members Who. . . "
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
Ehrlers-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
Physical Exam:
Abnormal blood pressure or pulses
Heart murmur
Arrhythmias
May be NORMAL!!

Table 2. Warning Signs for Sudden Cardiac Death.

"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.[1] 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.[4] 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.[2]

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.[2] 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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For comments to this article, send email to: APRTJR@CongenitalCardiologyToday.com

~CCT~

Principal Author:

Tiffany J. Riehle, MD, MSE
Pediatric Cardiology Fellow
Sibley Heart Center at Children's Healthcare of Atlanta
Atlanta, GA 30329

riehlet@kidsheart.com

Co-Author:

Robert C. Campbell, MD, FAAP, FACC, CMO,
Sibley Heart Center at Children's Healthcare of Atlanta
Director, Sibley Heart Center Cardiology Division Director of Cardiology, Dept. of Pediatrics
Emory University School of Medicine

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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 (hypercapneic 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 PaO2 <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, 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 post-operative 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_2 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 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 devel-

opment 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 postoperatively may be acute and are characterized by severe desaturations with hypotension and associated tachycardia.

The management of postoperative pulmonary hypertension involves increase of FiO2, hyperventilation with development of respiratory alkalosis, adequate sedation, neuromuscular paralysis and the use of medical therapy in the form of nitric oxide, MgS04 infusion, calcium channel blockers, phospodiesterase 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 intubated and patient's age between 1-4years are the main risk factors. It very rarely leads to irreversible respiratory insufficiency, though less rarely it may lead to the need for reintubation.

Central airway compression

Vascular rings, dilated pulmonary arteries, left atrial enlargement and cardiomegaly may cause long term tracheal compression resulting in poor tracheal development. Tracheobronchomalacia 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 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 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

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 car-

diac 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 experience 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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For comments to this article, send email to: APRAT@CongenitalCardiologyToday.com

~CCT~

Address for correspondence:

Aphrodite Tzifa, MD, MRCP
Fellow in Pediatric Cardiology
Department of Congenital Heart Disease
Guy's & St Thomas' Hospital, London

aphrodite.tzifa@gstt.sthames.nhs.uk

Shakeel Qureshi, MD, FRCP
Department of Congenital Heart Disease
Guy's & St Thomas' Hospital, London

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.

"It became evident very early on, that there was no uniform consensus regarding how these patients should be followed up after the intervention."

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.

"Our aim is to identify treatments for coarctation of the aorta which have optimal efficacy and safety."

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

"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."

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.

> ***"The Consortium meets twice yearly … with the next meeting scheduled for May 4, 2005 during SCAI at Ponte Vedra, Florida."***

For comments to this article, send email to:
APRTJF@CongenitalCardiologyToday.com

~CCT~

Thomas J. Forbes, MD
Associate Professor Wayne State University
Director Catheterization
Children's Hospital of Michigan
Detroit, Michigan

tforbes@dmc.org

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DRUG-ERROR RISK AT HOSPITALS TIED TO COMPUTERS

By Scott Allen, Globe Staff - March 9, 2005

(Reprinted Courtesy of The Boston Globe): "Drug-error risk at hospitals tied to computers," by Scott Allen.) www.globe.com

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Publishing Management
Tony Carlson, Founder
Tel: +1.301.279.2005
Fax: +1.240.465.0692
TonyC@CongenitalCardiologyToday.com

Richard Koulbanis, Editor & Publisher
RichardK@CongenitalCardiologyToday.com

Virginia Dematatis, Editorial Consultant
VirginiaD@CongenitalCardiologyToday.com

Loraine Watts, Editorial Assistant

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Sales Office
CONGENITAL CARDIOLOGY TODAY
9008 Copenhaver Drive, Suite M
Potomac, MD 20854 USA
Tel: +1.301.279.2005
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Editorial Office
CONGENITAL CARDIOLOGY TODAY
19509 Pine Cone Court, Suite 100
Gaithersburg, MD 20879 USA
Edit@CongenitalCardiologyToday.com

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