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Lutembacher Syndrome Masquerading as Rheumatic Mitral Stenosis

*By Dinesh Kumar Yadav, MD; Sandeep
Choudhary, MD; Pankaj Gupta, MD; Pradeep
Kumar, MBBS*

Introduction

Lutembacher Syndrome refers to a congenital atrial septal defect (ASD) complicated by development of acquired mitral stenosis (MS).^{1,2} The incidence of MS in patients with ASD is 4 to 6%, whereas the incidence of ASD in patients with MS is 0.6%- 0.7%.^{1,3} In patients with MS who had undergone percutaneous balloon mitral valvuloplasty (PBMV) through the trans-septal approach, the latter procedure created an ASD during the procedure: the iatrogenic Lutembacher Syndrome which is rare in the pediatric age group.⁴ There is usually marked right ventricular hypertrophy and failure, and reduced blood flow to the left ventricle because blood flows back to the right atrium through the ASD.

Case Report

A 10-year-old female presented with a history of difficulty in respiration, cough, mild fever, chest pain and swelling in both the lower limbs for last 15 days. Difficulties in respiration was more in lying position and also during her routine work (NYHA-G-III), and within the last 7 months she developed

several episodes of respiratory distress. Difficulty in respiration was gradually progressive in nature. Cough was exaggerated in the lying down posture. It was not associated with a sore throat, sneezing, a runny nose or postural changes, and was nonproductive without any diurnal change. Chest pain was described as mere awareness of the heart beats against the chest wall. The fever was low grade, intermittent with no diurnal variation. There was no history of joint swelling, hemoptysis, cola or red-colored urine, rashes, loss of weight, weakness in any extremities and abnormal body movements.

“Lutembacher Syndrome refers to a congenital atrial septal defect (ASD) complicated by development of acquired mitral stenosis (MS).^{1,2}”

Examination at admission revealed a conscious thin child with a thin build, a weight 19.6 kilogram (less than 3rd centile as per WHO charts), with a height 124 centimeter (less than 3rd centile as per WHO charts), a mild pallor and no cyanosis, icterus,

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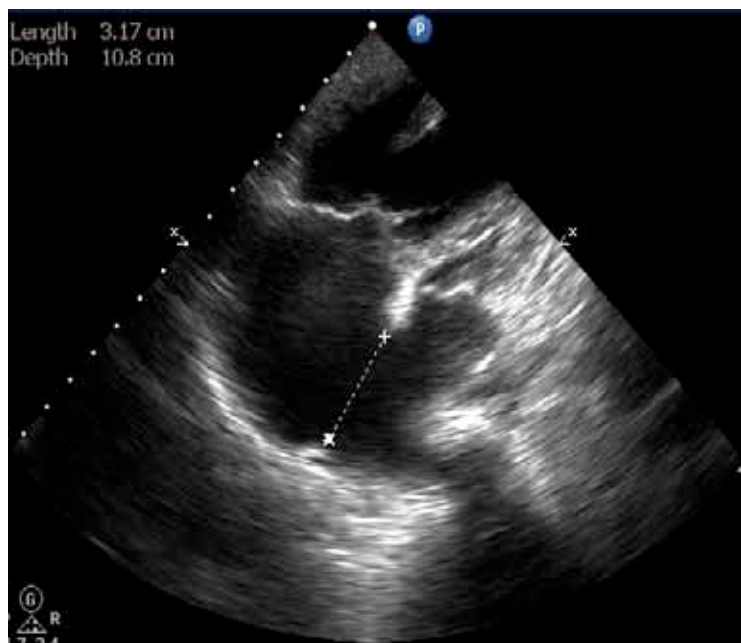


Figure 1. Showing atrial septum with severe mitral stenosis.

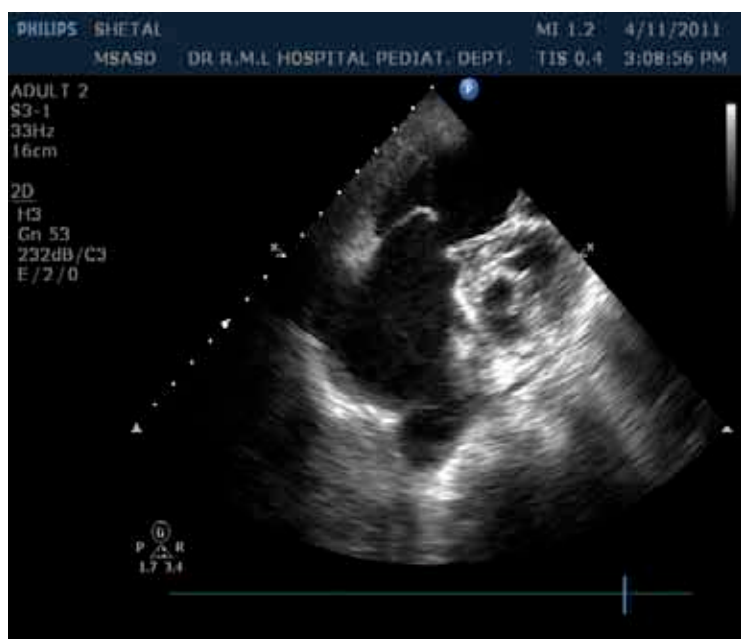


Figure 2. Showing severe MS with fibrosed tip of Tricuspid valve leaflet.

lymphadenopathy, splinter hemorrhages, subcutaneous nodules or rashes. She had bilateral pedal edema, which was pitting in nature. Jugular venous pressure (JVP) was elevated at 10 cm of water. Her blood pressure was 109/56 mm Hg and pulse 108/minute. She was tachypneic with a respiratory rate of 40 breaths per minute and oxygen saturation of 88 to 93% on room air. On precordial examination the apex beat was in the 6th intercostal space in anterior axillary line. There were prominent pulsations in the left parasternal area. Loud S1 and a mid-diastolic rumbling murmur grade II/IV was present in mitral area along with a pansystolic murmur of grade IV/VI along the left lower parasternal border. Also, an early to mid-systolic murmur grade II-III/VI was audible in the pulmonary area with a loud P2 and wide splitting of the second heart sound. Respiratory system examination revealed

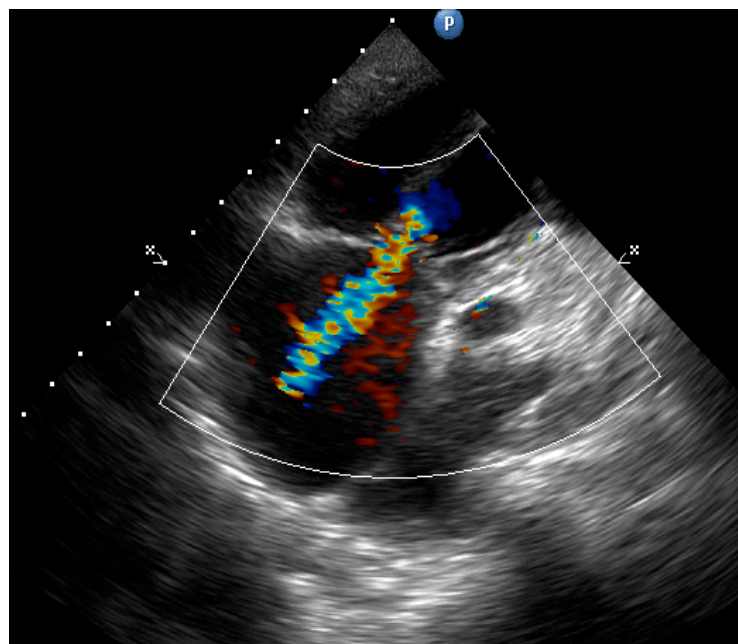


Figure 3. Showing tricuspid regurgitation.

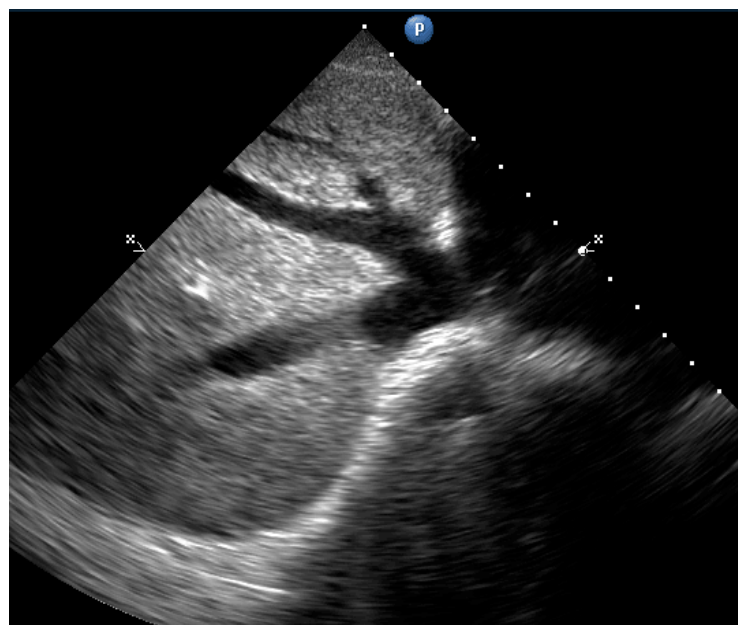


Figure 4. Showing dilated IVC.

bilateral basal crepts with rhonchi. Abdomen was soft, nontender and liver was palpable 7 centimeter below right costal margin. A provisional diagnosis of Rheumatic Heart Disease with active carditis, congestive heart failure, severe mitral stenosis, tricuspid regurgitation and pulmonary arterial hypertension without infective endocarditis was made. Atrial septal defect with mitral stenosis was kept as a remote possibility (Lutembacher Syndrome).

Investigations revealed a hemoglobin of 10.4 g/dl, total leucocyte count of 8200/mm³ (P70, L26, E04), erythrocyte sedimentation rate of 18 and a positive C-reactive protein. Her ASO titres were negative. Random blood sugar was 114 mg/dl whereas her serum sodium (134 mEq/L) and serum potassium (3.6 mEq/L) were normal. KFT (Blood urea and creatinine), LFT, CPK, LDH were normal. Her blood culture was sterile. X-ray chest revealed

plethoric lung fields and cardiomegaly with a C/T ratio of 0.70 and right ventricular hypertrophy with prominent pulmonary conus and right atrial enlargement. A twelve-lead electrocardiogram showed peaked P waves, normal PR intervals, with right axis deviation of the QRS, and rSR complex in right chest leads. Transthoracic echocardiography revealed a large ostium secundum atrial septal defect (size 3.2 cm) with grossly dilated right (size-6.1x5.3) and left (size-5.7x2.2) atrium. Her right ventricle was dilated, while the left ventricle was small with severe mitral stenosis (mitral valve area 0.738 cm²). Tricuspid regurgitation was present with a pressure gradient of 50 mm of Hg. Left ventricular systolic function was normal with an ejection fraction of 73%. Mitral valve leaflets were thickened with uniformly restricted movements and partly calcified with some fusion of adjacent chordae. Tricuspid leaflet was also thickened and partly calcified.

On final analysis of the patient, there was no history suggestive of Rheumatic Heart Disease, and the ASO titer was negative. The major and minor Jones criteria could not be fulfilled, and the acute phase reactants (ESR, leucocytosis) were also negative. Also, the patient's age was against the diagnosis of severe mitral stenosis due to Rheumatic Heart Disease.

The patient was treated with digoxin, diuretics, ACE inhibitor and ranitidine, and underwent successful open mitral valvulotomy, atrial septal defect closure and tricuspid valve annuloplasty.

Discussion

In 1916, Lutembacher first described a combination of congenital ASD with acquired mitral stenosis.⁵ Its incidence is very rare. Mitral stenosis may be acquired (Rheumatic Heart Disease) or congenital.

The mitral valve in secundum atrial septal defects has a morphological resemblance to both rheumatic and floppy valves, although it is quite different histo-pathologically. Histologically, in rheumatic disease of the mitral valve, vascularisation is a striking feature which is absent in Lutembacher Syndrome. The characteristic chordal elongation and myxomatous changes typical of the floppy valve are also absent. The consistent involvement of the medial half of the anterior cusp with total sparing of the posterior cusp is the hallmark of Lutembacher Syndrome. The valve changes in secundum atrial septal defect are caused by a mechanical secondary change due to abnormal movement of the medial half of the anterior cusp.⁶ This is supported by the fact that the apparent prolapse of the mitral valve disappears after closure of the defect.⁷ The changes in the anterior cusp of the tricuspid valve are produced by a similar mechanism and can be found in many cases of long standing pulmonary hypertension due to any cause.

The natural history and hemodynamic features of patients with Lutembacher Syndrome may vary and depend on the size of ASD, severity of mitral stenosis, pulmonary vascular resistance and the compliance of right ventricle.

The hemodynamic of ASD is altered by high left atrial pressure due to mitral stenosis. This, in turn, leads to an increase in the shunt across the ASD and in higher pulmonary flow than what

Left Ventricular Functions by M-mode at Variable Intervals

IVSd	IVSs	LVIDd	LVIDs	LVPWd	LVPWs	FS	EF(%)
1.18	1.20	1.72	1.04	0.816	0.952	39.5	73

otherwise would have been in the absence of mitral stenosis. When mitral stenosis is severe and atrial septal defect is small, it usually presents clinically as pure mitral stenosis. However, when the atrial septal defect is large, the signs and symptoms of ASD dominate the clinical picture, despite severe mitral stenosis. Thus, the severity of mitral stenosis may be underestimated in either case scenarios. It is prudent to exclude mitral stenosis before closure of ASD or else it results in increased morbidity and mortality from intractable pulmonary edema.

In our case the provisional diagnosis of Rheumatic Heart Disease with mitral stenosis was made due to increased prevalence, early presentation and incessant progression of rheumatic heart disease in the Indian subcontinent. Pediatricians and pediatric cardiologists have a great responsibility to acknowledge and rule out the possibility of Lutembacher Syndrome in any case

“This case report highlights the fact that the possibility of Lutembacher Syndrome should always be kept in mind in older children with atrial septal defect with prominent diastolic murmur and signs of pulmonary hypertension.”

presenting as mitral stenosis.

There are numerous case reports of Rheumatic Heart Disease in association with secundum defects.⁸⁻¹¹ The incidence of MS in patients with ASD is 4 to 6%, whereas the incidence of ASD in patients with MS is 0.6%-0.7%.^{1,3} Early diagnosis and surgical treatment bears a good prognostic value. If the patient is diagnosed at late stage, pulmonary hypertension and heart failure develops and the prognosis is poor.¹² If the patient is diagnosed earlier - before the development of pulmonary hypertension and heart failure - ASD closure with either mitral valvulotomy bears a good prognosis and prolongs survival.

This case report highlights the fact that the possibility of Lutembacher Syndrome should always be kept in mind in older children with atrial septal defect with prominent diastolic murmur and signs of pulmonary hypertension.

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Doctors, Patients and Families from Around the World Make Progress on a Unique Disorder

By Matthew J. Toth, PhD

Named after the Dutch neurologist Dr. Peter Barth, Barth Syndrome is a rare but underdiagnosed X-linked genetic disease. Barth Syndrome patients who experience a constellation of symptoms including cardiomyopathy, neutropenia, growth delay, and fatigue that present in varying degrees, have a very effective patient-driven organization in the Barth Syndrome Foundation (BSF). Like many patient-interest organizations the mothers of affected sons began to reach out by telephone and computer to find other parents of children with the same diagnosis. This reaching out and subsequent “kitchen table” meetings culminated in the formation of the BSF. Since that time and with the rise of the internet, the BSF has grown extensively with international affiliates in Canada, the UK, France, and South Africa as more and more patients are identified with this pan-ethnic disease. Established over 12 years ago, the BSF held its first scientific/medical/family conference on Barth Syndrome in Baltimore, Maryland in October, 2002 and has sponsored dynamic international meetings that focus on the scientific, medical and family issues of this unusual mitochondrial disease every two years since then.

On June 25-30, 2012 the 6th International Scientific, Medical & Family Conference on

Barth Syndrome will be held at the Don CeSar Hotel in St. Pete, Florida.

BSF conferences are organized in a way that has been cited by others as a model. The 2012 Conference will be another two-track event, bringing together in one location a scientific/medical meeting, running concurrently with a patient/family meeting. Scientists and affected families from all over the world are attracted to these gatherings. Knowledge about Barth Syndrome, especially in the scientific area, has increased enormously since that initial meeting. At the 2002 conference, there were only a few speakers, and presentations were less detailed because very little was known about Barth Syndrome. In contrast, for 2012 the number of invited speakers had to be limited to keep it to a two-day meeting. Much of this growth in knowledge about Barth Syndrome can be traced directly to the BSF Research Grant Program, which has directly funded over USD \$2 million of Barth Syndrome research since 2002. This biennial conference has become the premier meeting where updates on Barth Syndrome research and treatment are disseminated, and importantly, where investigators are energized, longitudinal data are collected, and new collaborations are initiated.

Like a microcosm of today's NIH-funded biomedical world and like other rare disease

research organizations, the BSF faces the same obstacles of “translating” the knowledge it has sponsored into effective clinical practice. Understanding the mechanism and effectively treating the cardiomyopathy of these patients is of paramount interest. Barth Syndrome boys and men often are physiologically stable for long periods of time; however, episodes of acute heart failure (which in some cases have led to death) punctuate the lives of these patients and their families and cause them to live in a cruel state of anxiety.

One way to tackle this issue was heralded at the 2010 Conference when the first reports of a mouse model of Barth Syndrome were presented—a major milestone for asking questions that could impact clinical practice. Since 2010, over eight additional laboratories have started working with this mouse model which is now available through Jackson Laboratories for worldwide distribution. For 2012, Drs. Phoon, Soustek, Khuchua, Kiebish and Chicco will talk about the amazing parallels of the mouse model with the human disease, especially in terms of the cardiac aspects. Dr. Phoon's work utilizing mouse echocardiographies, especially the in utero observations, are particularly relevant because cardiac complications are often found during the postnatal period of Barth Syndrome infants.



At the 2010 meeting, the clinically-oriented presentations focused on mitochondrial diseases in general, and the keynote speaker, Professor Douglas C. Wallace, gave a far-reaching talk that showed how mitochondrial diseases have a much greater impact on public health than is commonly thought. In 2012, there will be specific Barth Syndrome clinical reports from Drs. Kelley, Byrne, Cade, Pu, and Ren that relate directly to ongoing/future clinical care by covering areas such as: nutritional intervention, gene therapy, physical therapy, induced pluripotent stem cells, and potential pharmaceutical treatments. One prime example of the increased awareness of Barth Syndrome is the Barth Syndrome Service at the University Hospitals in Bristol, UK. This NHS Specialized Services program not only treats patients from the UK but also from mainland Europe. Dr. Steward, who heads this program, will report on his experiences and those of his team since beginning this special program in 2010.

The biochemical details of tafazzin mutations, the causative gene defect of Barth Syndrome, will not be neglected at the 2012 meeting. From the pioneering work of the late Dr. Peter Vrenken and colleagues, Barth Syndrome is uniquely associated with the special mitochondrial phospholipid, cardiolipin. For 2012, Drs. Claypool, Greenberg, Vaz, Schlame, McMaster, Hatch, Shi, and DeKroon will add to our understanding of tafazzin, phospholipid metabolism, and perhaps suggest therapeutic interventions. One of the few diseases directly associated with a lipid disorder, Barth Syndrome continues to present challenges to understand the mechanism(s) that gives rise to its varied symptomatology.

The conference features a valuable poster session (with poster travel stipends available), and for 2012 two poster presenters will be selected to speak about their work on the second day of the scientific-medical session. Also new for 2012 is a scholarship program for interested healthcare professionals, especially physicians. Treating Barth Syndrome patients, in fact even recognizing them clinically, often requires special attention. The BSF is offering scholarship stipends to help defray the expense of attending this conference for those healthcare

professionals who want to learn more about Barth Syndrome and improve patient care.

For 2012 the Director of the NIH's Office of Rare Disease Research, Stephen C. Groft, Pharm. D., will be the keynote speaker. Dr. Groft has been a great friend and advisor to the BSF over the years, and he will share with us his ideas about the future of rare disease research.

The presentation of the *Verner Award for Pioneers in Science and Medicine* will be part of the luncheon-keynote address on Friday, June 29th. In 2010, the award was presented to two pioneers -- Dr. Daniela Toniolo for her discovery that the tafazzin gene is responsible for Barth Syndrome, and posthumously to Dr. Peter Vreken for his work on cardiolipin and Barth Syndrome.

Because these meetings each time gather together the largest numbers of Barth Syndrome boys and men ever assembled, they are important opportunities to collect longitudinal data about this rare mitochondrial disease. In 2010, six IRB-approved protocols were participated in by most of the Barth Syndrome individuals who attended, and the 2012 conference will host its own data collection opportunities. The longitudinal data collected from previous conferences has been or will be published, which we hope will speed along advancement towards finding a specific treatment.

The cardiomyopathy of this rare disease can be deadly. By holding these conferences, the BSF hopes to stimulate awareness and generate interest in those physicians who deal with this as well as other important pathologies. The BSF wants to increase physician/scientist interest in order to turn the data collected into knowledge that can be translated into specific and effective treatments for Barth Syndrome individuals. There is no registration fee for this meeting, and the BSF invites all interested physicians and researchers to take advantage of our poster/scholarship programs and to attend the 2012 conference to learn more about Barth Syndrome. Detailed information is provided on our website, www.barthsyndrome.org, or can be obtained by contacting me directly.



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The 5th International Sudden Arrhythmic Death Syndromes (SADS) Foundation Conference: Preventing Unexpected Sudden Death in the Young

By Robert M. Campbell, MD

The 5th International Sudden Arrhythmic Death Syndromes (SADS) Foundation Conference: Preventing Unexpected Sudden Death in the Young, held October 1-2, 2011 in Atlanta, GA was a tremendous success. Greater than 175 attendees listened as speakers from around the country and world presented state-of-the-art presentations regarding channelopathies and sudden cardiac arrest. The objectives of the conference were to:

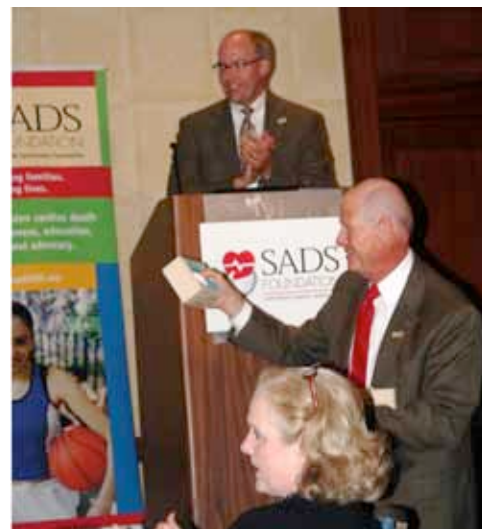
- 1) discuss the complexities and new developments and diagnosis of inherited channelopathies,
- 2) evaluate the importance of developing expanded family pedigree and prospective case findings and early treatment for the prevention of sudden cardiac death,
- 3) discuss the state-of-the-art ion channelopathy care with emphasis on new innovations and knowledge and current best practices,
- 4) enable affected individuals to live and thrive despite their SADS condition, and finally,
- 5) to analyze the pathophysiology, symptoms, presentation, diagnosis and management of channelopathies.

The Saturday, October 1st conference schedule was kicked off by Dr. Peter Fischbach,* President of the Pediatric and Congenital Electrophysiology Society (PACES); Children's Healthcare of Atlanta at Emory University School of Medicine who spoke about the challenges of diagnosing LQTS, CPVT, and Short QT Syndrome. Silvia Priori,# Director of the Cardiovascular Genetics program and Professor of Medicine at New York University, and the Director of Molecular Cardiology and Electrophysiology in Pavia, Italy, followed with an update on risk stratification for channelopathies. Most recent information gleaned from large channelopathy registries around the country confirms that these channelopathies, most commonly genetic, are less symptomatic and life threatening than previously thought. However, data for risk stratification, especially into a very low risk or no risk category, remains incomplete. Enhanced correlation between genotype and phenotype will allow the promise of enhanced risk stratification in the near future. Susan Etheridge, SADS Vice President and PACES Vice President, spoke about the challenges of living with SADS. While there are numerous risks, limitations, expenses, restrictions, and anxieties, there is now information detailing enhanced survival

and improved outcomes, with effective treatment. The state of pre and postmortem genetic testing for cardiac channelopathies was detailed by Dr. Michael Ackerman,* Director of the Long QT Syndrome/Inherited Arrhythmia Clinic and the Windland Smith Rice Sudden Death Genomics Laboratory at Mayo Clinic. Mike is also President of the SADS Foundation. He detailed the first ever expert opinion guidelines on genetic testing for these conditions, just published in *HeartRhythm* in August. Erin Demo, certified genetic counselor at Children's Healthcare of Atlanta Sibley Heart Center, described the role of genetic counseling.

"...the conference was regarded as a tremendous success, highlighted by direct face-to-face engagement between families affected with ion channelopathies and renowned ion channelopathy experts."

Dr. Priori led off the afternoon with an invigorating talk entitled "View Over the Horizon: What Will Cardiac Channelopathy Diagnosis and Management Look Like in 2020." It's daunting, and exciting to think how technological breakthroughs will enhance our ability to diagnose, risk stratify and treat channelopathies. Dr. Phil Saul,# Professor of Pediatrics at Medical University of South Carolina and immediate past President of PACES, discussed the upside and downside of implantable cardioverter defibrillators. A quick poll of attendees revealed more reporting adverse events (from implants, inappropriate shocks, etc) than appropriate lifesaving shocks. Dr. Michael Ackerman followed with a detailed discussion of latest treatment options for cardiac channelopathies, including new concepts regarding drug therapies for Long QT Syndrome, the antifibrillatory effect and role of left cardiac sympathetic denervation (LCSD) surgery, and the controversy that still persists regarding activity restriction recommendations versus published guidelines. Dr. Dan Roden,# Director of Personalized Medicine and



Dr. Michael Ackerman, current President of the SADS Foundation, honors Dr. Michael Vincent (foreground), the Founder and Member Emeritus of the SADS Foundation.



Marina Lesnitskaya (left) psychologist from the SADS Russia group with Dr. Silvia Priori (right), New York University.

Professor of Clinical Pharmacology at Vanderbilt University, defined for the audience the concepts behind Long QT Syndrome and Brugada drug lists. Many attendees were hearing for the first time how drugs interact with ion channels to enhance arrhythmogenesis.

An early evening healing wall ceremony with a candle lighting, for both those who have died from sudden cardiac arrest and those who have survived with their channelopathies, followed. This was a time of great emotion, but also bonding for families and among families.

The evening dinner gala included a special presentation from by Dr. Michael Vincent,



Children and teenagers attending the SADS Connect youth conference.

Professor Emeritus at University of Utah and the Founder and Member Emeritus of the SADS Foundation, who detailed his introduction to Long QT Syndrome, and the course that his career took guided by the involvement of a founder family affected by Long QT Syndrome in the Salt Lake City area. Katherine Timothy, an original member of the SADS Foundation staff and family, followed with a remembrance of her involvement with families and interactions with Dr. Vincent.

The Sunday, October 2nd session allowed attendees to hear from SADS volunteers and SADS staff, who spoke of their participation in education, awareness and fund-raising efforts. Marissa Smith, SADS volunteer and ion channelopathy patient herself, discussed her experience with the *Keeping Hearts Beating* campaign, a highly successful local fundraiser in her area. Dr. Stuart Berger,* Medical Director of the Herma Heart Center at Children's Hospital of Wisconsin, then followed with discussion of the Automatic External Defibrillator (AED) safety net and data describing successful implementation in schools and areas of high density public traffic. The morning session was highlighted

by the SADSConnect, during which a group of 24 children and teenagers described their experience during the youth conference (aka SADSConnect) to the larger SADS Foundation conference audience. A resounding theme for these young people was the positive impact that the conference had on their self-esteem and confidence. Many of them expressed that relating to others, similarly affected by ion channelopathies, made them feel "more normal," and "not alone."

Parents and families addressed their questions to speakers in a 1 ½ hour expert panel conference session. Results of a post conference attendee survey revealed that a direct interface with ion channelopathy experts, meeting with other families and hearing their stories, and the SADSConnect track were highlights of the meeting. Greater than ¾ of the attendees had traveled more than 250 miles to attend the conference.

Overall, the conference was regarded as a tremendous success, highlighted by direct face-to-face engagement between families

affected with ion Channelopathies and renowned ion channelopathy experts. Interface between the SADS Foundation and PACES continues to strengthen, which is critically important as new information regarding the diagnosis and management of individuals and their families with a possible cardiac channelopathy continues to explode at an almost exponential pace.

Footnotes

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Improving Communication and Outcomes for Children with Complex Congenital Heart Disease Using Parent Information-Seeking Behavior

By Debbie Hilton-Kamm, MBA

Introduction

The outcomes of children with Complex Congenital Heart Disease (CCHD) depend heavily upon good physician-parent communication, allowing parents to comply with physician's directions and adhere to follow-up protocols. Many studies have shown a correlation between physician-patient communication and improved patient health outcomes.¹ Parents of children with CCHD need to understand several aspects of their child's care including the diagnosis, pre/post-operative care, follow-up procedures and tests, medications, and the potential for developmental delays, physical limitations, and learning or behavioral issues. Helping parents understand how all of these factors impact their child requires effective and constant communication by the physician. The use of a communication strategy based upon parents' information-seeking behavior (ISB), reflective listening, and continual assessment of parents' understanding of these complex issues is essential in promoting successful communication.

Consequences of Ineffective Communication

Ineffective communication may include the complete lack of communication regarding a specific issue, communication that is partially or completely misunderstood, or that which is not retained by the parent. Any of these communication errors may result in parents unknowingly putting their child at increased risk of illness or death by not complying with certain instructions. Ineffective communication can also result in a parents' lack of hope for their child's future and damage to the physician-parent relationship.

Parents cannot act on information they do not comprehend, or that which they are not aware. Several studies have shown that a large percentage of parents of children with CCHD do not understand many aspects of their child's care such as the risk of infective endocarditis (IE) and the requirement for IE

prophylaxis.^{2,3} Awareness of the importance of dental hygiene was not associated with the parents' education level.³ Other studies report that many parents are unable to correctly name their child's defect.^{4,5} Cheuk, et al also reported significant knowledge gaps regarding the functions and potential side effects of medications, as well as a lack of understanding of the child's physical restrictions. A review of studies on respiratory syncytial virus (RSV) immunoprophylaxis reported compliance rates varied from only 25% to 100%. One of the major contributors to noncompliance was a lack of understanding the benefits of the vaccination against RSV.⁶

While the diagnosis of a CCHD carries significant risks and unknowns, physicians need to also convey information on positive outcomes to avoid negatively skewing information and diminishing parents' hope. In relation to Hypoplastic Left Heart Syndrome (HLHS) for example, physicians must be clear when citing the age of the eldest survivors (currently approaching age 30) so that it is not mistaken for life expectancy. Terminology regarding the condition must also be quantified whenever possible to avoid misinterpretations. A recent survey of parents of children with HLHS found that over a quarter of respondents interpreted the term "rare" to mean "few or no other survivors" or "it occurs in less than one in a million births." Similarly, more than 20% of respondents interpreted "rare" to mean "little or no chance of survival."⁷ This survey also found that only 18% of those receiving a prenatal diagnosis of HLHS and 8% receiving the diagnosis after birth received information on support networks from their pediatric cardiologist.⁷

Miscommunication can perhaps be the most detrimental when termination of pregnancy is presented as an option for those receiving a prenatal diagnosis of CCHD. Communication at this time is crucial to ensure parents understand the potential implications of their child's diagnosis and allow them to make critical decisions.⁸ Gaining a realistic perspective of the potential outcomes is essential for parents deciding whether or not to terminate the pregnancy; a decision which

has life-long implications no matter which they choose. Consequently, physicians need to listen carefully to the concerns and views of parents regarding termination to avoid misunderstandings.

Some parents may view the physician's mention of termination as a recommendation, and, if opposed to termination, parents may be offended at the suggestion.⁹ This can lead to irreparable damage to the physician-parent relationship; parents want physicians who are highly motivated to achieve the best outcomes for their child, and a "recommendation" to terminate the pregnancy may be viewed by some as counter to that objective. The mention of termination can also be interpreted as an indication that the child has a diminished chance of survival. A nationwide survey of parents of children with HLHS found that when termination was presented after the parents had declined, or if parents felt pressure to terminate from the pediatric cardiologist, they were less optimistic about their child's life expectancy. They were also more likely to find a new pediatric cardiologist than those who did not feel pressure or have termination mentioned after they declined.¹⁰

Communication Strategies Based on Information-Seeking Behavior

Effective communication must be flexible and adapted to parents with different backgrounds, communication styles, and personalities. Assumptions about parents based solely on demographic characteristics can be misleading, as those who share common demographics do not always think or behave in the same way. Patient participation in discussions with physicians has been found to be related to the physician's communication style more than the patients' demographic factors.¹¹ Psychographic variables - values, attitudes, beliefs and behavior - are a more meaningful way to segment populations than demographics alone. Values and beliefs may be difficult to assess quickly. However, parents' information-seeking behavior is readily apparent and can give physicians insight into the parents' motivations and level of understanding of critical information, helping



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physicians choose an appropriate communication strategy. This, in turn, can help parents feel more comfortable asking questions or clarifying information, leading to better understanding and better rapport with the physician.

Several models of information-seeking have been published illustrating the complex interactions between personal aspects of motivations, self-efficacy, beliefs and attitudes with external variables such as the environment and demographic factors.^{12, 13, 14} These variables interact to create a propensity to behave in a certain way such as active or passive information-seeking. Many recent studies now focus on information-seeking solely as it relates to internet use. However, ISB in a broader context includes asking questions, clarifying information presented by medical professionals, obtaining information and opinions from family and friends, reaching out to support networks or other parents with children with CCHD, and seeking out information on the diagnosis and treatment from the internet and other sources. The extent to which parents engage in this broader context of ISB (proactively, moderately or passively) can guide physicians to the most effective communication strategy (Diagram 1).

Proactive Information-Seekers

Behavior: Proactive information-seekers ask many questions, may reference what they have read or “heard” from other sources, and may even bring lists of questions to the appointment. Communication with these parents may be facilitated by their proactive nature; physicians have been found to better predict patients’ health beliefs when patients are more active participants.¹⁵

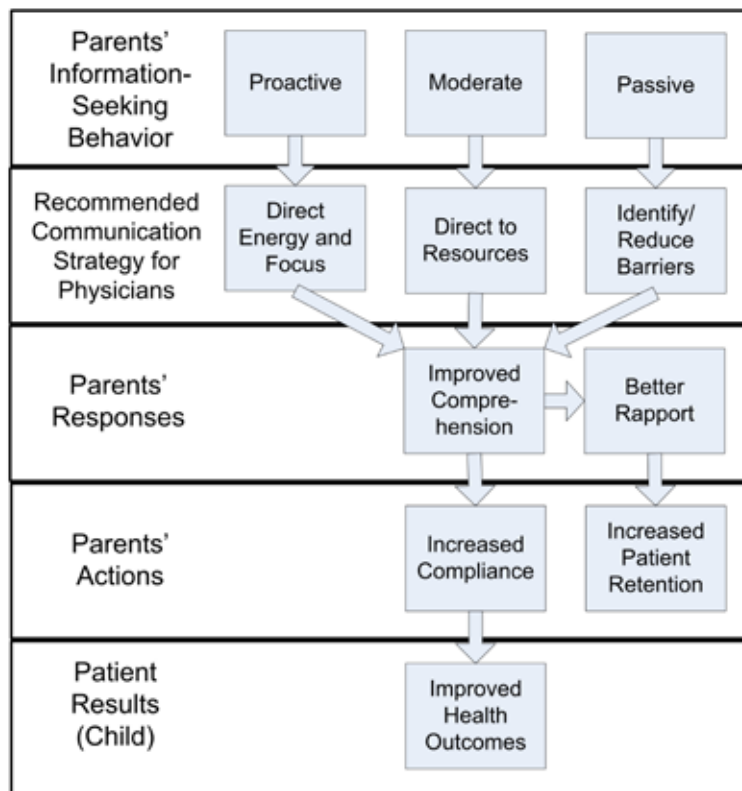
Characteristics: These are assertive “doers” who take an active role in learning about the condition and in the decision-making process. They are motivated by a need to feel that they did everything possible for their child to achieve the best possible outcome. Proactive information-seekers may be more factual and numbers-driven seeking information on survival rates and probabilities of various outcomes. Support networks are used as a source of emotional support and for factual information. Their demand for detailed information and the physicians’ time can be challenging; however, their high level of engagement indicates a motivation and willingness to comply with physicians’ instructions. Some in this segment are early adapters who are willing to participate in clinical trials and new treatments if they deem it to be beneficial to their own child and for other children with CCHD. Therefore, proactive information-seekers should be highly valued and effort should be made to effectively manage and retain these parents.

Socio-Economics: Those with formal educations may look to the physician as an expert in the specialty, but as a peer in decision-making, preferring shared control in the doctor-patient relationship.¹¹ They will be offended if they perceive the physician to be condescending or not completely forthright with information. Their economic status allows them more choices in where to seek treatment. If they have trouble establishing rapport or trust with the physician they will likely seek out other medical opinions and/or change physicians. Proactive information-seekers with lower socio-economic status may not have as many choices in where to seek treatment. However, the fact that they are proactive despite a lesser education or income level is an indication of a very high level of personal assertiveness and motivation.

Potential Influences: Proactive information-seekers utilize a variety of sources of information including medical literature via the internet,

friends or family members with medical backgrounds, and support networks. One study found that patients used an average of five different

Diagram 1: Results of Effective Communication Strategies Based on Information-Seeking Behavior



sources of information, but that the physicians were still the most trusted source.¹⁶

Communication Strategy: Direct Energy and Focus

Physicians can direct the parents’ energy and focus by giving them valid, prescreened resources (websites, printed materials), saving time and reducing confusion from conflicting or outdated information. Providing these additional resources gives them “permission” to continue their outside research, provides them with productive activities which reduces their anxiety level, and builds rapport with the physician.

Moderate Information-Seekers

Behavior: These consumers present with some questions, but may not have a good understanding of the condition or treatments. They may appear to be engaged, but are not clear what to ask.

Characteristics: These parents may be overwhelmed, may not appear to be very assertive, or may be worried about offending physicians with too many questions. They may be more socially-driven and want to feel comfortable with the physician. Moderate information-seekers may not seek second opinions as readily as proactive information-seekers. Their

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decision-making style may lean toward shared decision-making, but with an emphasis on the physician's recommendations.

Potential Influences: Friends and family members are their primary influences. They may also engage in some internet searches or seek out support groups primarily for emotional support.

Socio-Economics: Those from higher socio-economic levels will have more choices in providers; however, all moderate information-seekers can benefit by being given appropriate resources.

Communication Strategy: Direct to Resources

Showing compassion and understanding for their situation creates a warm, friendly atmosphere in which they feel more comfortable asking questions. Most of these parents simply need to know what questions to ask, and where to seek help. Support groups and community resources can provide them with a starting point where they can learn how other parents have handled the situation. The website CHDResources.org offers materials such as: a "Resource Guide," "What to Ask the Surgeon and Hospital," and "What to Ask the Insurance Company," among others.¹⁷ Providing basic information on the diagnosis and treatment options in writing is helpful to parents so they may review the information outside the physician's office. With appropriate guidance and resources, these parents can become more proactive in their information-seeking and more engaged in the decision-making process.¹¹

Passive Information-Seekers

Behavior: These parents may not ask many questions, or may ask questions that seem irrelevant or of minor consequence. Passive information-seekers are probably the most challenging for physicians because they provide little feedback on their level of understanding. Many different types of barriers exist that could result in passive ISB and physicians must try to identify and eliminate those barriers to improve the potential for good outcomes.

Characteristics: Passive information-seekers may have language, cultural, educational or emotional barriers to engaging and communicating with the physician. They may be intimidated by the physician, emotionally shell-shocked, or may not understand the severity of the situation. These parents may not feel confident in their abilities to make decisions and may want more physician-led decision-making. Compared with proactive information-seekers, these parents may appear at first to be "low-maintenance" – not demanding much time to answer questions. However, these parents actually require more time and attention to ensure they comprehend the information necessary to adequately care for their child.

Potential influences: Those with language or cultural barriers may be heavily influenced by family members and cultural norms. Those who lack those barriers but seem "shut off" emotionally or exhibit low health literacy may be reluctant to seek information from outside sources.

Communication Strategy: Identify and Reduce Barriers

Before true communication can begin, barriers must be identified and reduced or eliminated. Physicians first need to assess whether obvious language, cultural or educational barriers exist. Language barriers can be managed using formal interpreters or family members who can accurately communicate information to and from the parent. Cultural

barriers may include mistrust of physicians or the opposite - total reliance on the physician for decision-making. Some parents may be intimidated by the physician or think the physician will be offended if they ask questions. Cultural barriers can be mitigated by providing an inviting environment and letting the parents know that asking questions is OK and expected. Finding medical representatives, such as a nurse, with whom the parents may feel more comfortable, can also be helpful. Assessing the health literacy of parents, providing information in simple terms, and continually monitoring parents' understanding level of information can help those with low health literacy.¹⁸

Parents who present with none of the barriers listed above may have emotional barriers such as denial of the medical condition, or a belief that they are powerless in the situation. Physicians can emphasize the important role parents play in determining their child's outcome to help them feel empowered. If the parents are in denial, emotionally detached or overwhelmed, seeking help from a mental health provider or social worker may be beneficial in helping them become more engaged in their child's care. Breaking through to these parents is of utmost importance in helping them care for and bond with their child.

General Communication Strategies

Communication strategies that can benefit all parents should be used in conjunction with the more specific strategies outlined above. General strategies include: acknowledging parents' emotions, allowing time to digest information before making any decisions, providing written information including diagrams for review at a later time, offering prescreened internet sites and other resources, referring to support networks, and continually monitoring the parent's level of understanding.^{19,20} Stress level, emotional state, and perceived importance of information can affect recall of information. Some estimate that up to 40-80% of information may be lost immediately.²¹ Therefore, repeating information and presenting it in varying formats are necessary components of effective communication.

Reflective listening: Not all parents will express their emotions overtly, so it is up to the physician to gauge parents' emotional readiness to take in more information. Reflective listening is a technique whereby the physician identifies and acknowledges the parents' emotions by paraphrasing parents' comments. For example, if the parent says, "I just can't imagine my baby having heart surgery," the physician can respond with, "Thinking about your baby having surgery is frightening." Pausing after the reflective statement allows the parent to expand on the emotion, clarify the issue, or take the discussion in a different direction. The parents may give more detail about their concerns – perhaps expressing concern about the child dying, or apprehension about their ability to adequately care for the child. Allowing the parents to express these issues can help direct the discussion towards the parents' main concerns. This technique shows empathy towards the parents which leads to higher patient satisfaction. Despite the benefits of reflective listening and open-ended questions, one study found that these methods were only used 30-40% of the time by physicians.²²

Assessing understanding: To accurately gauge parents' comprehension, physicians need to continually assess the parents' level of understanding. The Teach-Back method, in which the physician asks the parent to repeat the information back, can be highly effective in helping physicians learn what information was understood and retained. It is best when the physician asks in a compassionate manner such as,



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"I'd like to make sure I explained things clearly. Can you tell me what you understood?" It is important to note that this method is not a test of the parents' knowledge, rather it is a test of how well the physician explained the material.

¹⁸ The use of open-ended questions such as, "What part of this is difficult to understand?" can also lead to more detailed discussions and deeper understanding by the parents. Unfortunately, these techniques may not be widely used. In a study of residents' counseling of patients the much less effective closed-ended questions and "OK?" were used frequently, while the Teach-Back method and open-ended questions were rarely used.²³

Conclusion

Using the communication strategies outlined above, physicians have the power to effectively manage and retain proactive information-seekers, and help moderate and passive information-seekers become more engaged in their child's care. Improved physician-parent communication saves time and leads to better understanding, compliance, and rapport. Most importantly, the use of these effective communication strategies can lead to better health outcomes for children with complex congenital heart disease.

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- Six-Page Resource Guide
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These free materials can help physicians:

- Save time and avoid misunderstandings
- Create clear written instructions quickly and easily
- Increase patient understanding, retention and compliance

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SCAI Monthly Column: Plan Now! Advance Registration Deadline for SCAI 2012 is March 29

The Society for Cardiovascular Angiography and Interventions (SCAI) is reminding those specializing in pediatric and adult congenital interventions to plan now to attend the *Congenital Heart Disease (CHD) Symposium at SCAI 2012 Scientific Sessions*. Taking place May 9-12, 2012 at the Mirage in Las Vegas, the meeting will celebrate 35 years of the best-of-the-best in interventional and invasive cardiology.

Be sure to beat the March 29th Advance Registration deadline by visiting www.scai.org/SCAI2012!

In addition to old favorites, CHD Symposium Chair Daniel Levi, MD, FSCAI and CHD Symposium Co-Chair Thomas Fagan, MD, FSCAI will be introducing several new sessions in May, each with an eye on creating a bridge between pediatric and adult interventionalists. Here's a preview:

Round Peg in a Square Hole

Outside-the-box thinking will take center stage at the new "Round Peg in a Square Hole" session. Moderated by Frank F. Ing, MD, FSCAI and Zahid Amin, MD, FSCAI, this session will focus on the overlooked role of the congenital interventionalist as innovator and how each attendee can be better at thinking outside-of-the box in applying existing devices to each individual's care.

"This is where you showcase your creative thinking," said Dr. Levi, "by sharing the innovative uses you've found for existing devices."

Safety First!

Imaging safety is an obvious, but often overlooked, concern in the pediatric cath lab. The issue is also one far too overlooked when it comes to physician education.

In order to fill this gap, *SCAI 2012's Congenital Heart Disease Symposium* will also be offering an all-new "Imaging Safely" Session. This session will feature a panel of physician experts sharing their strategies for reducing imaging radiation and introducing techniques that are likely to be the future of imaging.

Specifically designed to impact YOUR practice, attendees will come away with proven strategies they can employ

immediately to minimize radiation exposure for patients and the whole the care team.

The session will include presentations covering:

- Minimizing and Defining Radiation Exposure
- Multimodal Interventional Image Guidance
- XMRI Fusion
- Fluoroscopic Future

Brown-Bag Lunches

Eat while you meet with thought-leaders in congenital/pediatric/structural interventional therapies. Back by popular demand, SCAI's Brown-Bag Lunch series offers a unique opportunity for candid, forward-looking discussion of what's coming down the pipeline in Interventional Cardiology.

"I Blew It"

Learn from your colleagues' nightmares, or share your own. Now in its 13th year, the "I Blew It" sessions have become a favorite fixture of SCAI's Congenital Heart Disease Program, with off-the-chart evaluation ratings year after year. "Interventionalists love case-based learning, especially when the cases are about how to avoid or handle real-life complications," said Dr. Levi, "and the I Blew It" sessions are exactly that.

During the "I Blew It" sessions, attendees become the faculty and everyone participates in brainstorming around what went wrong, options for handling it, and – most importantly – how to avoid having it happen again.

Brain Scratchers

SCAI's Congenital Heart Disease program is continuing its popular "Brain Scratchers" session, where a series of unusual cases are presented and the audience and faculty work together on these brain scratchers.

"These are the puzzling cases that crop up in practice and you turn to your colleagues and ask what they would make of them," said Dr. Levi. "Attendees gave the 'Brain Scratchers' high marks last year, and we are pleased to bring them back."

Want to Learn More About SCAI 2012?

Want to review the schedule for *SCAI 2012* or want to take advantage of the advance



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- **Daily Breakout Sessions** dedicated to the care of adults with congenital and structural heart disease.
- **Hot Daily Debates** between cardiologists and surgeons on controversial issues in intervention for congenital and structural heart disease.
- **Breakout Sessions** for cardiovascular nurses and CV technicians.
- The popular session of **"My Nightmare Case in the Cath Lab"**
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Abstract Submission Deadline is December 1, 2011.

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RSNA 2011 Spotlight News

By Tony Carlson, Founder & Senior Editor

Benefits of Using a Cloud-Based Service to Share Medical Imaging Studies and Reports - Focus of a Podium Presentation at RSNA 2011

A podium presentation at *RSNA 2011*, "Benefits of Using a Cloud-Based Service to Share Medical Imaging Studies and Reports" by Drs. Mark D. Kovacs and Michael A. Trambert, reviewed data from Virginia Commonwealth University Medical Center (VCU), an early adopter of cloud-based medical data sharing. VCU uses eMix™, the industry-leading cloud-based medical data-sharing service.

Drs. Kovacs and Trambert described the old technical barriers to sharing patient data between healthcare providers with different, proprietary information technology systems. They also noted the problems associated with previous workaround solutions that were developed to overcome those barriers. These solutions include faxing, burning images to CDs, and sending files via virtual private networks. The inadequacy of these older solutions prompts redundant imaging studies 10%-to-20% of the time. Such unnecessary imaging exposes patients to excess radiation and adds tens of billions of dollars to national healthcare costs.

For more information, visit www.emix.com.

Siemens Second Annual International CT Image Contest at RSNA 2011

At *RSNA 2011*, Siemens announced the winners of its *2nd Annual International CT Image Contest*, which attracted more than 600 submissions from 43 countries in seven medical categories. A 3D reconstruction of a CT image created using Vizua's 3D scan visualization technology was selected as the winner of the pediatrics category.

Submitted by Dr. Jean Francois Paul from Centre Chirurgical Marie Lannelongue in France, the high-definition 3D image features a visualization of an aorta aneurysm in a 13-month old baby. It was recognized as an impressive example of how valuable diagnostic information can be obtained using low-dose scans.

"Vizua is the ideal bridge between the high-quality visual experience radiologists have today and what physicians and patients currently have access to," said Dean Lester, CEO at Vizua. "Vizua reconstructs 2D scans into 3D models that depict the true form of anatomy, enabling a deeper level of understanding of the patient's pathology without the need for mental gymnastics."

The winning image was created from CT scans that were uploaded to Vizua, a unique solution for creating, viewing, exploring and sharing large 3D images. Because Vizua is a cloud-based service, images are delivered through a web browser and can be shared instantly with anyone on any device, anywhere in the world. Affordably priced as a "pay-as-you-go" service, Vizua extends access to 3D renderings beyond the radiologist workstation out to physicians and patients, improving communication.

For more information visit vizua3d.com.

Digisonics Showcased Award-Winning CVIS at RSNA

Digisonics exhibited its DigiView Image Management and Structured Reporting System. The DigiView Image Management and Structured Reporting System has been ranked 2008, 2009 and 2010 *Best in KLAS in the Top 20 Best in KLAS Awards: Software & Professional Services* reports for the cardiology market segment. Digisonics solutions are standards-based and vendor-neutral, combining high-performance image analysis, professional reporting choices, an integrated clinical database and a powerful PACS into one comprehensive system.

The Digisonics CVIS also provides functionality designed specifically for congenital cardiology. Mullins and Mayer congenital heart diagrams can quickly be modified using graphical drawing tools and drop down labels. The Digisonics solution can also be interfaced with hemodynamics systems via HemoLink for autopopulation of the congenital diagram with pressures and flow measurements. Trend plots with z-scores enable clinicians to plot one or two specific measurements over time in order to track therapy or disease progression. Both the congenital heart diagrams and trend plots can be edited on-screen, printed in the report and stored in the cardiovascular database. A professional, structured report summarizing study findings can easily be generated using the configurable pediatric clinical comments and picklists and pediatric summary macro templates.

To learn more about Digisonics solutions, visit www.digisonics.com.

Toshiba's Aquilion™ PRIME CT Offers Fast, Low Dose Exams for Improved Patient Care

As the latest addition to the "ONE Family" of CT scanners, Toshiba America Medical Systems, Inc. showcased the Aquilion™ PRIME CT (pending 510(k) clearance). With double slice technology and coneXact™ reconstruction algorithm, Aquilion PRIME can generate 160 unique slices per rotation, achieving extremely accurate MPR and 3D-

rendered images. The Aquilion PRIME is designed for healthcare facilities that need to perform a wide variety of routine clinical examinations and produce the highest-quality clinical images with the least amount of radiation exposure, all while improving efficiency.

The Aquilion PRIME features an 80-row 0.5 mm detector, a 7.5-MHU large-capacity tube and 0.35 second scanning. This high-speed rotation allows rapid data acquisition and shortens scan times while the fast reconstruction unit further improves throughput. The system also features a 78 cm aperture gantry, the largest currently available in a high-end CT system. The combination of a 660 lb. patient weight capacity couch (option) with a large gantry bore makes the Aquilion PRIME an ideal system for use in non-standard conditions, such as emergency scanning or bariatric patient studies.

To reinforce the principle of As Low As Reasonably Achievable (ALARA) imaging, Adaptive Iterative Dose Reduction (AIDR) and NEMA XR 25 Dose Check Software are standard features on the Aquilion PRIME. AIDR technology is an iterative process that removes noise from the image to increase quality and helps to reduce radiation dose significantly. Toshiba's NEMA XR 25 Dose Check Software enhances user awareness of the radiation dose being administered to patients. This software includes Dose Alert and Dose Notification, addressing the two main components of MITA's CT Dose Check Initiative, as well as Tracking and Digital Imaging and Communications in Medicine (DICOM) to further enhance patient safety.

"The combination of the 0.35 second gantry rotation speed and the state-of-the-art reconstruction technology allows many examinations to be performed and reconstructed in seconds," said Joseph Cooper, Director, CT Business Unit, Toshiba. "That, along with standard patient safety features AIDR and NEMA XR 25 Dose Check Software, helps clinicians acquire high-quality images quickly, while minimizing radiation dose as much as possible."

For more information, visit www.medical.toshiba.com.

CCT

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Medical News, Products and Information

Rady Children's Researchers Find Heart Stent Effective in Very Young Patients

A widely accepted method of treating reduced blood flow to the heart – called percutaneous coronary intervention (PCI) – is a relatively common procedure in adult patients but not a generally accepted option for infants or toddlers with acute coronary syndrome. That could be changing, however, based on research conducted by the cardiology team at Rady Children's Hospital-San Diego.

In a new report published in the August issue of *Catheterization and Cardiovascular Interventions*, the official journal of The Society for Cardiovascular Angiography and Interventions (SCAI), Rady Children's researchers found coronary stent implantation to be a feasible and safe palliative option for children fifteen months and younger.

Studies have shown that in adults with reduced blood flow to the heart—known as acute coronary syndrome (ACS)—stent implantation significantly reduces mortality. While ACS is less common in children, there are congenital and acquired heart diseases that may compromise coronary circulation in the pediatric population. Due to increased risk of coronary interventions and difficulty of cardiac catheterization techniques in pediatric patients, PCI is not a recommended treatment strategy in this age group.

A research team led by Dr. András Bratincsák from Rady Children's conducted a retrospective review of seven children who had PCI between June 2006 and June 2010. Of those included in the study all were under 18 years of age, and four were 15 months or younger. PCI techniques included balloon coronary angioplasty and coronary stent implantation. Researchers analyzed patient data that included underlying diagnosis, comorbidities, catheterization technique, and outcomes.

In all seven cases, successful stent placement in the proximal portion of the left or right coronary arteries with excellent revascularization was achieved. The average diameter of the heart arteries was 0.65 mm prior to the intervention. Balloon angioplasty did not completely resolve the stenosis and bare metal stents were then implanted to a mean internal diameter of 2.5 mm. The team determined that the average intervention-free period was 434 days after stent implantation. Restenosis and thrombosis did not occur in cases where the implanted stent diameter was greater than 2.5 mm and patients received dual anti-platelet therapy.

"We provide evidence that stent implantation is a relatively safe option for pediatric patients with coronary stenosis, including those under the age of 15 months," said Dr. Bratincsák. "PCI offers a viable strategy for bridging infants and toddlers with blocked arteries or poor ventricular function to surgical revascularization or transplantation when they are at an older age."

Renowned Pediatric Cardiology Physician-Scientist Linda Cripe Joins Nationwide Children's Hospital

Linda H. Cripe, MD, a distinguished pediatric cardiologist, has joined the Heart Center at Nationwide Children's Hospital. Dr. Cripe will also be a member of the faculty at The Ohio State University College of Medicine.

Dr. Cripe is well-known for her continuing work in studying the care and treatment of cardiomyopathy associated with neuromuscular disease, especially Duchenne Muscular Dystrophy (DMD). She was a member of the Centers for Disease Control (CDC) National Steering Committee – Duchenne Muscular Dystrophy Standards of Care, and has been an invited lecturer nationally and internationally on cardiomyopathy related to DMD. She currently is a member of the Scientific Advisory Board for Parent Project Muscular Dystrophy.

Dr. Cripe was most recently the Chair of the Reappointment, Promotion and Tenure Committee and Coordinator for Medical Student Education in the Division of Cardiology at Cincinnati Children's Hospital Medical Center and served as an Associate Professor of Pediatrics at the University of Cincinnati. She completed her residency at the University of Iowa Hospitals and Clinics, and served as a pediatric cardiology fellow at the University of Iowa Hospitals and Clinics and at Children's Hospital Boston. She also has held faculty appointments at The Children's Hospital in Denver, Colorado, and at The University of Wisconsin.

"Linda Cripe is a preeminent physician-scientist and her recruitment to our clinical and research staff allows us to expand our capabilities in understanding and treating patients with cardiomyopathy and particularly those with neuromuscular disease," said Steve Allen, MD, Nationwide Children's CEO. "Recruiting staff of the caliber of Linda enables us to continue expanding our role as a leading institution in pediatric research and care."

Congenital Heart Patients to Benefit From Wireless, Battery-Free Cardiac Implant

A miniature, battery-free, wireless, cardiac implant being developed by a U-M researcher and the Ann Arbor company Integrated Sensing Systems, Inc. (ISSYS), has received important funding that could get it to patients more quickly.

A \$1.5 million grant from the National Institutes of Health (NIH), will help a research team, led by Martin Bocks, MD, and ISSYS, Inc., to complete the final preclinical testing required before seeking approval under Food and Drug Administration's Humanitarian Device Exemption pathway. Bocks is a pediatric cardiologist at the University of Michigan Congenital Heart Center and the U-M C.S. Mott Children's Hospital.

"We are extremely excited to continue working with ISSYS to develop a wireless, implantable pressure sensor for our patients with complex forms of congenital heart disease," says Bocks, the project's medical principal investigator.

"This pressure sensor has the potential to greatly improve the care we provide to our most complex patients and will provide us with unprecedented opportunities to learn more about their unique physiology. We appreciate the NIH and the National Heart Lung and Blood Institute (NHLBI) for their willingness to fund research on patients with rare, severe forms of congenital heart disease."

Bocks and his team at U-M have been working on this device – which will be used in infants and children – for three years. The 2.5-year grant will help test a device that will reduce the need for invasive cardiac catheterization procedures and provide a better understanding of congenital heart disease.

The device is a wireless, battery-free, miniature pressure sensor that is implanted within a heart chamber, Bocks says. Once implanted, doctors can measure pressure inside the heart while the patient is being seen in the outpatient clinic without having to do a heart catheterization. In the future, such monitoring may be performed remotely or from the home.

The device, which would stay in place permanently, is initially planned for use in patients with functional single ventricle conditions, such as Hypoplastic Left Heart Syndrome. It could be used in other forms of pediatric and adult heart disease in the future.

"We hope it would be going to market by year 2013," Bocks says. "We expect the device to decrease the number of heart catheterizations, help doctors better monitor the effects of medications, and provide early detection for conditions such as blood clots in lungs."

"We also hope that it will help us conduct more research," he adds. "We'll be able to obtain information from patients when they are sleeping, or exercising – information we had not been able to get to before."

The device is important because it will allow researchers to conduct more tests on these patients, which will lead to better treatments, says Nader Najafi, PhD, ISSYS President & CEO and the project's technical principal investigator.

"This grant helps us develop wireless, intelligent, miniature implants for patients with congenital and structural heart diseases. It paves the way for the start of clinical studies in infants and children with complex congenital heart defects," he says.

"The pediatric medical device field represents unique commercialization challenges due to its small size and stringent requirements," Najafi adds. "We are grateful to the NIH for this needed support for the challenging pediatric device field. The results of this effort will have important spillover effects for broader applications within the field of adult and pediatric cardiovascular medicine."

For information, visit: www.mems-issys.com.

Gladstone Scientists Identify Genetic Mechanism Linked to CHD

Scientists at the Gladstone Institutes have identified a finely tuned mechanism by which fetal heart muscle develops into a healthy and fully formed beating heart—offering new insight into the genetic causes of congenital heart disease and opening the door to one day developing therapies to fight this chronic and potentially fatal disorder.

In a paper published online in *Nature Genetics*, researchers in the laboratory of Gladstone, Senior Investigator Benoit Bruneau, PhD, described the roles that two genes—Ezh2 and Six1—played in embryonic heart development, while also uncovering how the genetic basis of embryonic heart formation can have profound health consequences later in life.

This research highlights the emerging importance of a biological process called "epigenetics," in which a genetic change that is inherited by a cell or organism early during development has long-term consequences.

Epigenetics is of particular interest in heart development, as the incorrect activation of genes in fetal development can lead to congenital heart disease into adulthood.

"Approximately 1.3 million children and adults in the US live with congenital heart disease—requiring daily medications, surgeries and for some, heart transplants," said Dr. Bruneau, who is also a professor of Pediatrics at the University of California, San Francisco, with which Gladstone is affiliated. "An understanding of the epigenetic regulation of heart development could someday bring us closer to improving the lives of these individuals."

At specific times during healthy heart development, Ezh2 acts as a "master regulator," shutting off genes that are no longer needed or that need to be kept off. In the past, the focus has been on which genes get switched on during normal heart development. But in this paper, Dr. Bruneau, along with Gladstone Postdoctoral Scholar Paul Delgado-Olguin, PhD, investigated which genes must remain off to ensure the development of a healthy heart.

In laboratory experiments, Drs. Bruneau and Delgado-Olguin removed Ezh2 from mice at various developmental stages, monitoring any ensuing genetic or physical changes and comparing them to mice whose Ezh2 remained intact. Surprisingly, mice without Ezh2 developed normally in the uterus. It wasn't until after birth that they began to show problems. Their hearts became enlarged and weakened and were unable to pump blood efficiently. An enlarged heart is a hallmark feature of cardiomyopathy, a form of congenital heart disease that afflicts thousands of children each year and for which the only manifestation may be sudden death.

Further analysis revealed that Six1 is normally on only for a brief period during heart development, after which Ezh2 shuts it off for good. But without Ezh2 to act as a regulator, Six1 remains on—leading to heart problems later in life.

This breakthrough may help researchers improve their understanding of the genetic causes of congenital heart disease while also pointing the way to potential therapies. For example, a type of congenital heart disease called dilated cardiomyopathy is

caused by mutations in Eya4, a gene that is also regulated by Ezh2 in the heart.

"Our next goal is to find out exactly how Ezh2 regulates these other genes, so that we can begin to develop a complete genomic blueprint of how a heart becomes a heart," said Dr. Bruneau.

Senior Research Technologist Yu Huang, MD, PhD, also participated in this research at Gladstone, which received funding from the National Institutes of Health, the California Institute for Regenerative Medicine, the DeGeorge Charitable Trust, the American Heart Association and William H. Younger.

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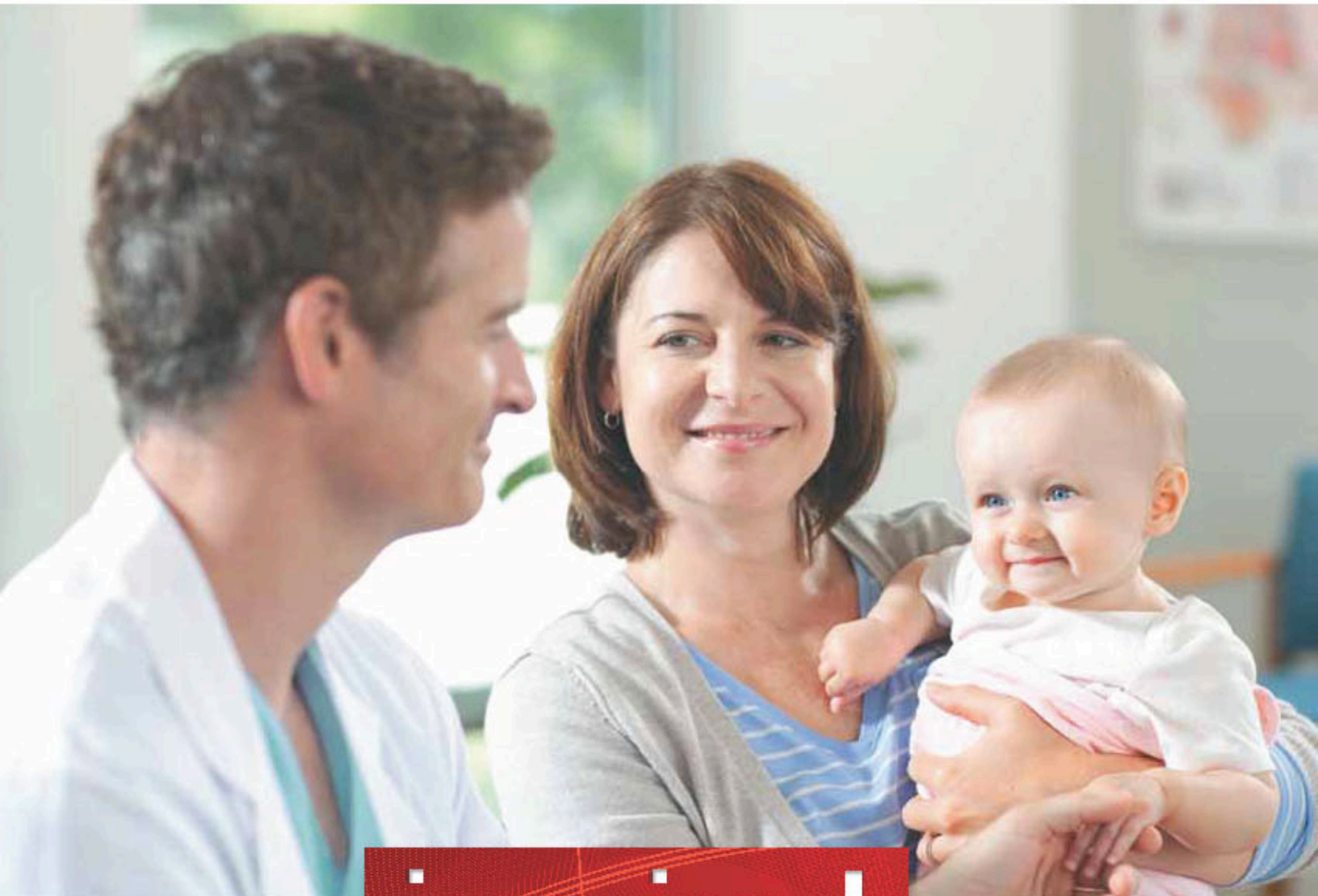
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