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www.israelheart.com

LAA Asia Pacific - How to Close the
Left Atrial Appendage
May 4, 2013; Hong Kong
csi-congress.org/laa-asia-pacific-workshop.php

SCAI 2013 Scientific Sessions
May 8-11, 2013; Orlando, FL USA
www.scai.org/SCAI2013

47th Annual Meeting of the Association for European
Paediatric and Congenital Cardiology
May 22-25, 2013, London, England
www.aepc-2013.org

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Big Data in Pediatric Cardiology: The Upcoming Knowledge Revolution

By Anthony C. Chang, MD, MBA, MPH

Big Data: An Introduction

We are immersed in a sea of data as a result of a proliferation of Internet-enabled communications: texts in books and publications, sensor data from monitors and self tracking devices, audiovisual data (recordings, pictures, video clips, etc), social media postings (Internet, Twitter, blogs, etc), and even global positioning system (GPS) signals. We are presently creating 2.5 quintillion (2.5 x 10¹⁸) bytes of data per day, and incredibly, 90% of all current data was produced only in the last two years with most of this data unstructured. In short, we are “dying of thirst (for knowledge) in an ocean of data”.¹

Big data is a recent data paradigm describing the coupling of this massive amount of data with sophisticated data analytics to acquire new knowledge or insight;² this phenomenon has taken place in a myriad of sectors such as business,³ finance,⁴ sports,⁵ and even the recent Presidential election (in which a data scientist, Nate Silver, handily proved his data analytical methodology to be superior to opinions of many political pundits in predicting the outcome of the election).⁶ Big data is thus, a mainstream data movement and knowledge revolution that emphasizes that “insight from data is more reliable than intuition from experts,” and is on the ascending “peak of inflated expectations” limb of the Gartner’s hype cycle for emerging technologies with an estimated 2-5 years to mainstream adoption (See Figure 1).



“You can have data without information, but you cannot have information without data.” ~ Daniel Keys Moran, science fiction writer/ computer programmer.”

In short, the advent of large amount of data coupled with the escalation of computer capability and data analytics has led to this data revolution. In simple layman terms, big data has been cleverly described by Yahoo Chief, Marissa Mayer, as “the planet developing a nervous system.”⁷ Big data has recently become such a high

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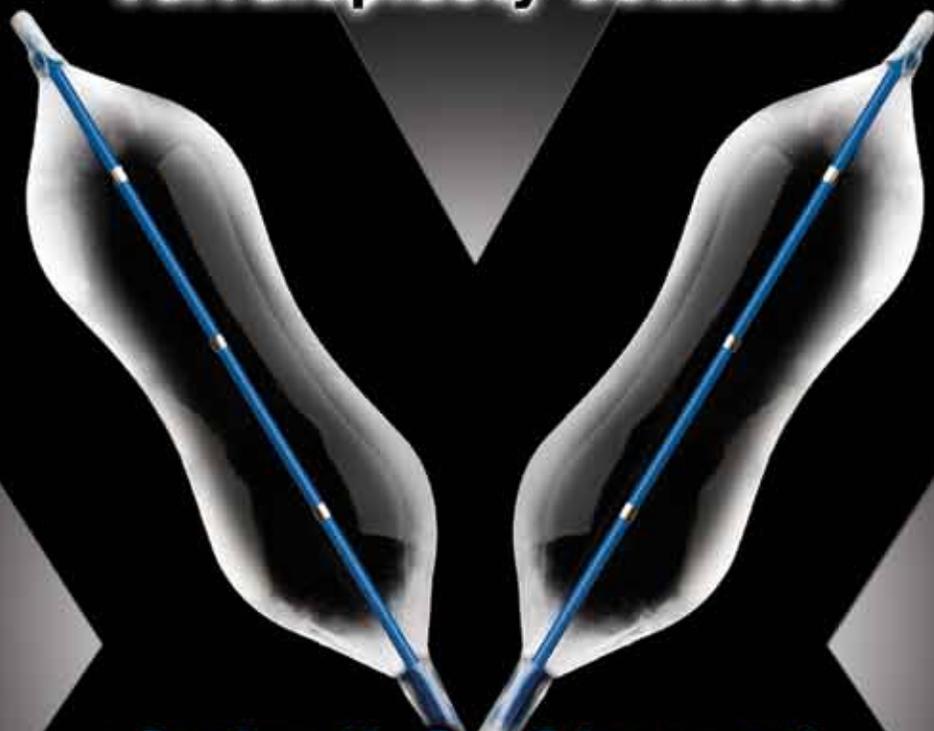
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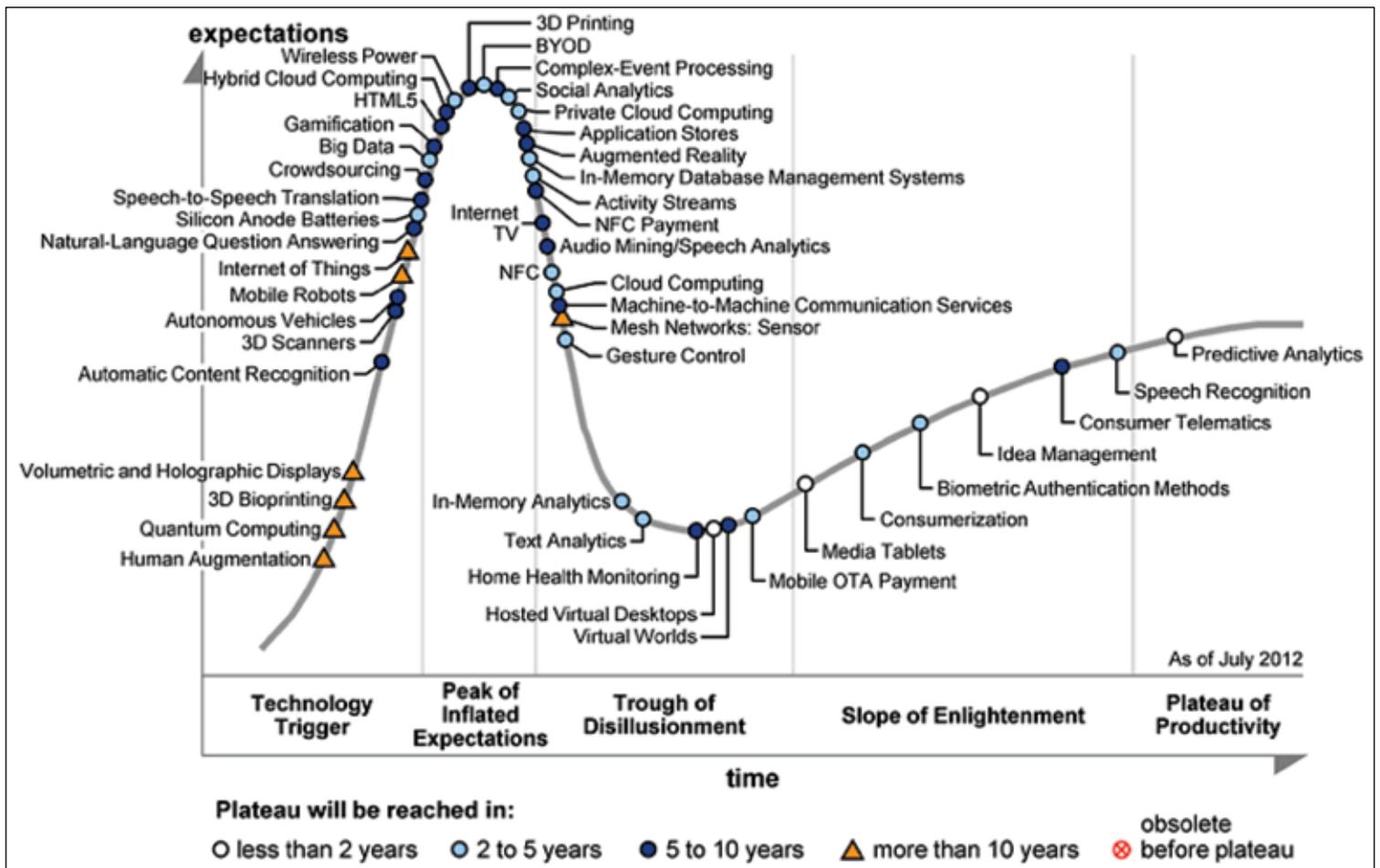


Figure 1. The Hype Cycle for Emerging Technologies, 2012 (From <http://www.forbes.com/sites/gartnergroup/2012/09/18/key-trends-to-watch-in-gartner-2012-emerging-technologies-hype-cycle-2/>).



Figure 2. The “V”s of Big Data (From <http://www.datasciencecentral.com/forum/topics/the-3vs-that-define-big-data>).

priority in the national strategic plan that President Obama approved a \$200 million Big Data Research and Development Initiative to glean discoveries from digital data.

Big data has important dimensions that all conveniently start with the letter “V” (See Figure 2), and these dimensions are:

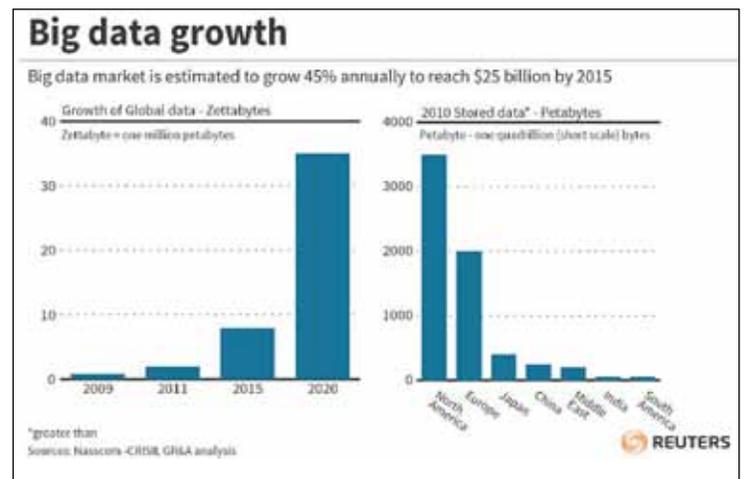


Figure 3. The Worldwide Data Growth (From <http://www.theopenstrategist.com/2012/10/big-data-growthchart.html>).

1. **Volume** - recent data volumes are calculated to be in petabytes (10^{15}) and exabytes (10^{18}) and will be in zettabytes (10^{21}) in the near future (highest unit is the yottabyte which is 10^{24}) (See Figure 3). In cardiology, these data include electronic medical records, home monitoring devices, genomic data, insurance claims, drug information, and imaging data (from echocardiograms, angiograms, and MRI/CT);



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2. **Variety** - both structured and unstructured data in numerous forms and combinations need to be considered; and
3. **Velocity** - from prior batch delivery, data transfer and analytics now need to be in fractions of seconds and even in real-time especially with imaging transfer of cardiac studies in telemedicine. Additional "V"s include:
4. **Veracity** - the accuracy and trustworthiness of big data and its analytics, and finally,
5. **Value** - ascertainment of how much this is worth to the institution.

Biomedical Big Data

Current biomedical big data, amassed by electronic medical records and digital image archiving (about 20 megabytes or MB per image), is reaching a staggering 100-250 exabytes with an annual growth rate of 1.2 to 2.4 exabytes,⁸ but remains extremely fragmented and disorganized. Our traditional "top-down" data approach up to now entails either healthcare databases or registries (that involve manual entry of data with its inherent limitations of accuracy and completeness, followed by data analysis with relatively basic statistical tools), or conventional hypothesis-driven research and randomized-controlled trials that have become prohibitively expensive, limited in scope, and often without definitive answers.

Recently, this new big data paradigm has been successfully applied to biomedical science mainly in the form of genomic medicine and its escalating genetic transcript big data.⁹ The vast magnitude and rapid acquisition of this genetic big data is absolutely vertiginous, as exemplified by Michael Snyder, a Stanford genetics PhD who has generated 30 terabytes of data of just his own biological data. Despite the daunting challenge, a few have met this challenge and successfully made strides that will have a positive impact on patient care.^{10,11} The capstone of this entire data transformation effort in genomic medicine is the ENCyclopedia Of DNA Elements (ENCODE project), an international collaboration of research groups funded by the National Human Genome Research Institute with the aim of delineating the entirety of functional elements encoded in the human genome.¹²

While this paradigm shift can also be applied to a clinical venue, such as pediatric cardiology and cardiac surgery, physicians and the healthcare system are collectively somewhat nescient in transforming the medical data imbroglio into better practice and usable knowledge (in other words, healthcare is "data rich, but information poor"). This glaring deficiency is primarily due to:

1. poor-quality data due to inaccuracy and incompleteness, as well as a lack of transparency and organization;
2. insufficient expertise in database management and subsequent data analysis as well as semantic analysis; and
3. a lack of healthcare organizational awareness and appreciation from the leadership down for the value of this data-derived information.

All of these inadequacies combined thus continually undermine the quality and outcomes of healthcare programs and organizations.

Interpreting Big Data

The new data paradigm in healthcare and in pediatric cardiology and cardiac surgery will be a "bottom-up" data management strategy that involves a three-step process after the data is acquired:

1. Data extraction with various data warehouses providing data;
2. Data transformation with data configured to a uniform format; and
3. Data loading with the data entered into an analytical system for final analysis.

One example of this data strategy was the tracking of the flu epidemic by Google (called "flu trends") and has been further refined in epidemiological studies using large data sets.¹³ The use of big data with real-time collection and analysis is facilitated by the new Hadoop database technology which minimizes the aforementioned extract-transform-load (or ETL) cycle of data processing.¹⁴

Big data can then strategically utilize methodologies in artificial intelligence to further gain essential information and to even discover new knowledge.^{15,16} MIT's Computer Science and Artificial Intelligence Laboratory (CSAIL) is leading some of the current efforts to combine big data and artificial intelligence to develop new techniques for big data interpretation that will lead to a genomic data-clinical medicine synergy. The advent of artificial intelligence methodologies such as natural language processing could enable the medical data manager



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This year the meeting will begin on Wednesday 22nd May with the Update course organised by the AEPC Imaging and Surgical Working Groups.

Followed by:

- The prestigious Edgar Mannheim lecture, by Professor John Hess from Munich, Germany on "Impact of changing patterns and technologies for congenital heart disease on long term outcomes."
- The main meeting includes: Plenary sessions and other sessions organised by various AEPC Working Groups, abstract presentations, moderated poster and other poster presentations and several industry sponsored symposia. An important scientific part of the meeting includes collaboration between different societies. These include the joint sessions between the AEPC and European Association of Cardiothoracic Surgery (EACTS), the AEPC and the Japanese Society of Pediatric Cardiology and Cardiac Surgery (JSPCCS) and AEPC and Asia Pacific Paediatric Cardiology Society (APPCS).
- There is a Young Investigators Award session during which the 6 best abstracts from European and Japanese researchers will be presented.
- This year's programme includes: the 9th Annual Multi-Societal Database Committee for Pediatric and Congenital Heart Disease meeting (23 May).
- The AEPC meeting will conclude with Keynote lectures on "the Past, Present and Future of Paediatric Cardiology" by Professors Michael Tynan and Shakeel Qureshi.

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"There is also some understandable concern that this paradigm shift will depersonalize medicine although the opposite may in fact be true as physicians are liberated from the burden of upkeep and stress of making the appropriate medical decision."

to not only organize, but also mine data from even unstructured electronic medical records automatically (without the tedious and inadequate manual inputting process). In addition, the impressive capability of the computer to perform word-sense disambiguation, or ability to put words into context, was displayed by the IBM computer Watson during its stint on the show Jeopardy! and is presently used effectively for cancer data analytics at Memorial-Sloan-Kettering Cancer Institute.

The perceived value of medical data should not be limited to data analysts and scientists in the IT department of the hospital but be extended to all healthcare workers in order to create a data-focused culture. In addition, big data is not merely about mining important information from the medical databases, but also demands executing practice changes and transformative innovations. The ultimate data transformation and medical knowledge in the future involves medical databases that will be "living" and "individualized" ongoing clinical research data and analysis with both static and dynamic data (via sensors) from the patient themselves. If embedded with conventional (such as data mining),¹⁷ or new (such as machine learning and artificial neural network)¹⁸ artificial intelligence techniques, these databases will finally enable "intelligence" into healthcare outcome determinations and decisions.

Implementation of Big Data

Big data is about quality, not quantity, of data. The implementation could involve a small project in a single hospital sector, such as the cardiology program, where data may be easier to be located. In addition, the focus should be on the problem that can be solved by big data, not big data or its analytics itself. The analytic skills even at a basic level should not rest with data analysts but should include everyone in the heart program. The project should undergo repetitive measurements and analyses to perfect the data analysis. Lastly, project execution should have a timeline and progress report throughout the project. Of course all of this data revolution will need adequate storage in the form of cloud computing.

There are some potential limitations of big data in biomedicine. First, there is the "signal-to-noise" problem: true disease data overwhelmed by data that do not truly reflect disease. This could result in over- or underdiagnosis/treatment of the disease. Second, the voluminous data needs to be relatable or at least understandable or visualizable by not just medical caretakers, but patients as well. Third, some caretakers may feel intimidated by the decision making aspects of big data when coupled with artificial intelligence in that in-



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telligent machines may replace certain caretakers. Perhaps the appropriate attitude is to form a human-computer synergy that will surpass the capabilities of either alone, not unlike how the GPS aids the driver. There is also some understandable concern that this paradigm shift will depersonalize medicine, although the opposite may in fact be true as physicians are liberated from the burden of upkeep and stress of making the appropriate medical decision. There is the potential issue of insufficient data storage capacity even with the cloud which can be solved by innovative solutions such as using DNA itself as a digital storage medium.¹⁹ Finally, there is the issue of ownership of the data with privacy of information that can lead to identification of patients (especially with a process called triangulation). This problem can be partly mitigated by anonymization mechanisms.

The Future: A Clinical-Digital Convergence

The U.S. excels in technology and innovation, but continually lags far behind other countries in healthcare data access, transparency, organization, management, and analysis. Our healthcare organizational and programmatic imperatives in this current era should embrace big data and data analytics/artificial intelligence expertise to harvest meaningful medical data and its hidden information to the fullest extent in order to improve quality and outcome.²⁰

In short, big data and strong data governance coupled with data analytics/ artificial intelligence will lead to a new wave of information/knowledge ("medical intelligence"). It therefore behooves us to rethink medical data in traditional forms of tedious databases/registries and even randomized clinical trials in order to innovate this valuable asset to create a clinical-digital convergence. Big data is, therefore, not about a collection of technological and analytical tools, but rather about a transformation in our healthcare ecosystem in which medical data, artificial intelligence, and personalized health are inextricably intertwined with human intellect and judgment.

"The end of theory: The data deluge makes the scientific method obsolete." ~ Chris Anderson, Wired magazine

References

1. Kohn, M (IBM Chief Medical Scientist). Personal communication (February, 2013).
2. McAfee A et al. Big Data: The Management Revolution. Harvard Business Review October 2012; 60-68.
3. Minelli M et al. Big Data, Big Analytics: Emerging Business Intelligence and Analytic Trends for Today's Businesses. John Wiley and Sons, Inc., Hoboken, New Jersey, 2013.
4. Cosentino T. Into the River: How Big Data, the Long Tail, and Situated Cognition are Changing the World of Market Insights Forever. Foundational Insights, LLC, Portland, Oregon, 2011.
5. The Baseball Prospectus Team of Experts. Baseball Between the Numbers: Why Everything You Know About the Game is Wrong. Prospetus Entertainment Ventures LLC/ Basic Books, New York, New York, 2006.
6. Silver, N. The Signal and the Noise: Why So Many Predictions Fail but Some Don't. The Penguin Press, New York, New York, 2012.
7. Hernandez, D. Big Data Is Transforming Healthcare. Wired Science, Oct 16, 2012.
8. Hughes G MD. How Big is Big Data in Healthcare? From A Shot in the Arm blog, October 21, 2011.
9. Butte A (Chief, System Medicine at Stanford School of Medicine). Personal communication (February, 2013).
10. Ashley EA et al. Clinical Evaluation Incorporating a Personal Genome. Lancet 2010; 375(9725): 1525-1535.
11. Butte A et al. Computational Translating Molecular Discoveries into Tools for Medicine: Translational Bioinformatics Articles now Featured in JAMIA. J Am Med Inform Assoc 2011; 18(4): 352-353.
12. The ENCODE (ENCyclopedia Of DNA Elements) Project. The ENCODE Project Consortium. Science 2004; 306(5696): 636-640.

13. Mark Reshef DN et al. Detecting Novel Associations in Large Data Sets. Science 2011; 334(6062): 1518-1524.
14. Mark McAfee A. What's the Big Deal about Big Data? Harvard Business Review webinar, October 4, 2012.
15. Mark Halevy A et al. The Unreasonable Effectiveness of Data. IEEE March/April, 2009, 8-12.
16. Mark Chang AC and Musen M. Artificial Intelligence in Pediatric Cardiology: An Innovative Transformation in Patient Care, Clinical Research, and Medical Education. Congenital Cardiology Today, 10(11): 1-15.
17. Hanson CW et al. Artificial Intelligence Applications in the Intensive Care Unit. Crit Care Med 2001; 29:427-435.
18. Hoefen R et al. In Silico Cardiac Risk Assessment in Patients with Long QT Syndrome. J Am Coll Cardiol 2012; 60:2182-2191.
19. Church GM et al. Next-Generation Digital Information Storage in DNA. Science 2012; 337(6102): 1628-1631.
20. Patel JL et al. The Coming of Age of Artificial Intelligence in Medicine. Artif Intell Med 2009; 46(1): 5-17.

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Review of PICS~AICS 2013 in Miami

By Ziyad M. Hijazi, MD, on behalf of all Course Directors & Co-Directors

With 800 attendees from 54 countries, the PICS/AICS meeting (Pediatric and Adult Interventional Cardiac Symposium) was a huge success and perhaps was the best ever! The City of Miami Beach opened its arms to the attendees with a personal letter from the Mayor of the city to the attendees.

Fifty percent of the attendees were from outside the US. One hundred-twenty-four faculty members gave over 120 talks; nine cardiac centers transmitted 20 live cases from North America, South America, and the Middle East! The quality of live cases this year was outstanding; everyone I talked to made special reference to the live cases and operators...congratulations to these operators, centers and their staff for making this possible.

PICS/AICS started Saturday January 19th at 8:15 am with a great session "Tips/Tricks," featuring three excellent speakers and demos of stents/valves/etc. Dr. Ing demonstrated stent techniques, Dr. Zahn demonstrated percutaneous valve techniques, and finally, Dr. Ilbawi demonstrated how to do a cutdown. This session was followed by taped cases where the presenters showed some cases, and directly interacted with the attendees. There were four taped cases presented by Drs. Ing, Zahn, Benson and Horlick.

Then over lunch break, there were three breakout sessions; the first one was "Stents in my Practice - When and Why." This session featured Tom Forbes, Jackie Kreuzer, Jo DeGiovanni and Marc Gewilg. The second session was "Device/Valve Design." In this session we had Hakan Akpinar and Jake Goble discuss "ASD devices" from an engineering point-of-view. Dr. Hieu, from Vietnam, talked about the "Lifetech Pulmonary Valve." Finally, the last session was about "Specialty Imaging," featuring Mario Carminati and Anthony Hlavacek. Then there was a debate between Craig Fleishman and Mark Fogel on the "Use of Echo vs. MRI for Percutaneous Pulmonary Valve Assessment."

The afternoon was rather busy with oral abstract presentations. This year, we decided to hold the oral presentations in the main room so that attendance would be maximum and it was. The quality of abstracts this year was exceptional. We received over 145 abstracts; a committee of 5 graders graded these abstracts. Finally, from 5-6 pm there was a session of "Meet the Expert," where we had the masters in our field discuss cases with the attendees. Chuck Mullins, Mario Carminati and John Bass moderated that session.

The exhibit opened its doors at 6 pm, and, with over 25 exhibitors showing the latest in medical technology, a crowd soon formed.

On Sunday, January 20th live cases were transmitted from Saudi Arabia with Dr. Tarek Momenah and his team performing three live cases, including a Melody valve.

Then Dr. Alejandro Peirone performed three excellent cases including: two ASD closures using the new PFM-R device, and one PDA closure using the new PFM-R PDA device. Finally, Dr. Pedra from São Paulo did three great cases including: two ASD cases with the new Occlutech Flex-II device. In one of the cases, the device had a fenestration, since the patient had elevated LVEDP. Dr. Peirone last case was coarctation stenting using the new Atrium Advanta covered stent.

There was a lot of significant and continuous interaction between the attendees and the operators regarding the technical aspects, as well as indications for interventions.

The afternoon was also busy with a few sessions. In the session, "Catheterization in the Developing World," a few speakers talked about pulmonary hypertension (Carlos Zabal), mitral valvuloplasty (Savi Shrivastava), available resources (Bharat Dalvi), VSD closure in China (Shiliang Jiang), PDA closure in South America (Raul Rossi). In another session, Dr. Kenny talked about the initiatives that have been implemented since the last PICS in Chicago, and the The Congenital Heart Intervention Mission Support (CHIMS), a charity foundation that was established to send help to developing countries.

There were two breakout sessions for the day. The first was "Adult Structural Heart Disease (LAA Closure)." In this session, we had the best experts in the world discuss various devices/techniques includ-





ing: Drs. Reisman, Turi, Kar, Walsh, Sievert and Ilbawi. Dr. Reisman showed a great videotape of pathological specimens of LAA and Dr. Sievert and Ilbawi had a spirited debate.

The second session has always been a popular one for nurses and technologists. This session was lead by Sharon Cheatham and Kathleen Nolan. The speakers included: Sharon Bradley-Skelton, Emily Kish, Elaine McCarthy, John P. Cheatham, Ruby Whalen, Karen Iacono, Richard Ringel, Gina Langlois and Kathleen Nolan.

The last session of the day was "Catheterization in HLHS," and in this session, we had Dr. Paul Weinberg show pathological specimens of HLHS. Dr. Marc Gewillig talked about stenting the septum, Dr. Holzer talked about coarctation, Dr. Stern talked about closure of APC's in these patients, Dr. Jo DeGiovanni talked about pulmonary artery interventions and, finally, Drs. Benson and Sano had a debate discussing the Sano operation vs. hybrid stage I for HLHS.



Monday January 21st, was also a busy day; it started with a new event sponsored by Siemens. Here I would like to thank Dan Digeorgio and Rob Dewey from Siemens for supporting this event. It was a "5K Run" at 6 am. We had over 70 runners, and I'm proud that people got up early and raced. Ralf Holzer was the winner in the event.

Monday we had three sites (Miami Children's, Orlando and Mississippi) transmit live cases. I had the pleasure to be a visiting operator/commentator with Darren Berman and Roberto Cubeddu, who did an excellent job in two cases: one was a Melody valve implantation, and the other one was coarctation stenting. Three-D rotational angiography was used in both cases. Dr. David Nykanen performed two cases: LPA angioplasty/stent and a VSD closure. Finally, Dr. Ebeid had Dr. Jones visiting with him as a guest operator/commentator, and they did two cases of transhepatic access, and a custom covered stent in a conduit in preparation for a Melody valve. The cases generated a lot of discussion and debate.

The didactic lectures in the afternoon were many including: a session on "new technologies that covered biodegradable stents" by Dietmar Schranz, Stem Cell Therapy for Percutaneous Valves by Massimo Caputo, and Transcatheter Ventricular Assist Device by Cliff Kavinsky. This session ended with a debate, Which is Better for Interventions in CHD...Europe or US, between Shak Qureshi and Larry Latson; of course we know who won the debate!

There were three breakout sessions on Monday. First, The PICES held their session this year at PICS. It was a very busy one and included: Tom Forbes giving a talk on CCISC, as well as case presentations moderated by Brent Gordon and Dan Gruenstein. Dan Levi talked about biodegradable stents in the context of getting ideas off the ground.

The second breakout session was about Mitral valve interventions. I attended this truly excellent session where: Mark Reisman again showed some specimens of the mitral valve, Scott Lim showed a taped case



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The end of the day was rather an emotional one, since we presented the 17th PICS Achievement Award to Dr. Savi Shrivastava. She did not know and we had her sister flown in from Boston secretly without Savi's knowledge. Shak did the presentation that he and I worked on for many months with the help of her sister, and her colleagues at Escort (Drs. Radhakrishnan and Iyer). This was followed by the Distinguished Service Award, which was given to Sharon Cheatham, who has supported PICS from the beginning and has developed the nursing and technologists sessions over the years. Dr. John Cheatham, her husband, and Dr. Cao helped me collect info/photos of Sharon from her early childhood to present.

After the awards, everyone joined us at the special dinner event, or what used to be the Gala night, which was held this year at Bongos Cuban Cafe; there was a lot of fun and camaraderie. We had over 550 people attend this event, and all had great fun to the degree they did not want to leave at the end of the night.

The last day of the meeting, Tuesday, January 22nd, was also a busy and very well-attended day. The live cases were transmitted from JFK Medical Center in Palm Beach, where Dr. Roberto Cubeddu and his team performed an excellent case of TAVI. I had the privilege to be the guest operator/commentator there. This was followed by a session that discussed ASD device erosions. In this one, Dr. Paul Weinberg showed specimens, followed by Girish Shirali talking on how to evaluate for erosions. Then the FDA joined the meeting via internet, and Nicole Ibrahim, PhD from the FDA, discussed the issue and recommendations of the FDA.

The other lectures in the session included an update on the RESPECT trial by Werner Budts; a talk by Joaquim Miro on ADP-II; a discussion of porto-systemic shunts in CHD led by Henri Justino; a talk by Elchanan Bruckheimer about covered stents; a talk by Tom Jones about RVOT conduit rupture during tPVR, and finally, Dr. Sano discussed tissue engineering in the management of HLHS.

Then live cases continued from Cedar Sinai with Dr. Zahn and his team, including Saibal Kar and Raj Makkar. They did great cases including: LAA closure with Lariat and a TAVR case (valve in valve). Live cases were then

"I hope to see you at our next meeting in Chicago, June 8-11, 2014 at the Marriott Downtown."

transmitted from the University of Colorado with Dr. Tom Fagan and his team, including John Carroll, who did an occlusion of anomalous vertical vein and a case of VSD closure. The last case was done using the new Philips system with MRI and echoNav.

The afternoon sessions were well-attended as well. The first was, "My Nightmare Case in the Cath Lab," where a few cases were presented. The winner was Masood Sadiq. This session was lively, and all attendees were very interested. The last session focused on the pulmonary valve and pulmonary arteries. In this session, we had Massimo Caputo talk about trans-apical pulmonary valve implantation; Lisa Bergersen talked about cutting balloon angioplasty; Phil Moore talked about high pressure balloon angioplasty to crack small stents; and the final two talks of the meeting were a debate between Emile Bacha and Zahid Amin about surgical arterioplasty. The meeting ended at around 5 pm.

In summary, the meeting was very successful and enjoyable. Miami offered a lot to the attendees and their families, and the scientific program was well-received.

I want to thank all who contributed to the success of the meeting, from attendees, organizers, coordinator, factoid specialists, live case operators, moderators, panelists, photographer, news editors and audiovisual staff who all helped make this meeting certainly one of the best in recent memory. I hope to see you at our next meeting in Chicago, June 8-11, 2014 at the Marriott Downtown. You can visit our website for more details and info at: www.picsymposium.com.

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of a Mitra Clip for MR, Ted Feldman talked about an update of the evolving mitral valve repair systems, Matt Gillepsie talked about percutaneous mitral valve replacement, David Reuter talked about mitral valve repair using the Mitral annuloplasty systems. Also in that session, we discussed paravalvar mitral leak by Omer Goktekin; this was followed by a debate between Bob Sommer and myself on the technique for paravalvar closure: transapical or endovascular?

The last breakout session was in Spanish, this is the first time we did such a session, and it was well-attended. This session was chaired by Horacio Faella and Carlos Pedra; speakers included: Joaquim Miro, Carlos Zabala, Felipe Heusser, Alejandro Peirone, Miguel Granja, and ended with a debate between Jackie Kreutzer and Raul Arrieta.

Successful Chronic Treatment with Sildenafil in Two Patients with Functionally Single Ventricle After Fontan / Hemi-Fontan Procedures

By Jacek Bialkowski, MD; Malgorzata Szkutnik, MD

Introduction

Inspiration to write this article came from "A Cautionary Tale for Pediatric Cardiologists" by Dr. John W. Moore, recently published in *Congenital Cardiology Today* (CCT). "A Cautionary Tale for Pediatric Cardiologists" by Dr. John W. Moore. In his excellent review of the book "Immortal Bird" by Doron Weber, he described the medical history and the history of medical relations of the family of the patient with functionally single ventricle, who had completed Fontan procedure.¹ After a few "good" years after surgery, the patient developed swollen testicles and had ruptured hernia due to development of Protein Losing Enteropathy (PLE). This patient had received albumin and IVIG infusions. He was also treated with steroids. Each of those therapies provided only temporary improvements, but PLE was relentless. Finally, he received a heart transplant. Unfortunately, the patient died shortly thereafter at the age of 16 years old because of fulminate Post-Transplant Lympho Proliferative Disorder. In this paper many important questions and problems related with communications between patient, his family and medical staff are described.

The aim of this paper is to describe two of our patients with single right ventricle after Fontan / Hemi-Fontan operation whom we treated successfully with Sildenafil. In the history of the child described above, this method of treatment was not mentioned, and in our opinion, it changed a primarily poor prognosis to a positive one.

Case Report 1

A twenty-one year old male from another center was admitted to our department because of severe heart failure and protein losing enteropathy (PLE) as a consequence of a failing Fontan. Initial diagnosis was mitral atresia, single ventricle and malposition of the great arteries. In infancy, pulmonary artery banding and surgical atrial septectomy (Blalock-Henlon) was performed. A second operation followed at the age of 4 years (hemi-Fontan), and one year later, completion of Fontan – total cavo pulmonary connection (TCPC), without fenestration was performed. During later follow-up the patient developed

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PLE with ascites. At the age of 15 years he was catheterized and the mean pulmonary artery pressure (mPAP) was 27 mmHg. Pulmonary artery anatomy was good. During the next 5 year his condition deteriorated – increasing peripheral swellings, ascites and cachexia. Because of his ascites, an abdominal hernia developed, which was closed surgically 1 month before his first admission to our department. At this moment there was still severe ascites with spontaneous drainage of peritoneal liquid from the post-surgical scar of the previously closed hernia. The circumference of abdomen at that time was 130 cm with body weight 65 kg. He also had a visible dilated jugular vein, hepatomegaly (6 cm) and peripheral oedema, desaturation (85%) and pleurothorax of the right lung. He was in NYHA class III. After examinations (ECHO, TC, MRI) good single ventricle function (EF 55%) and pulmonary anatomy were confirmed. During pleurocentesis 1400 ml of liquid was removed. Until that time he was treated with furosemid, hydrochlorothiazid, spironol, carvedilol and inhibace. Because of no clinical improvement, we decided to introduce sildenafil 3 x 25 mg daily. After 4 weeks of such therapy we observed spectacular clinical improvement (NYHA class II). He lost 5 kg of fluids, the circumference of his abdomen was smaller (85 cm), his post-surgical wound closed successfully, liver diminished (3 cm). He was discharged home on this therapy. After 3 months he lost another 14 kg of fluid, and his abdomen progressed to be a smaller size. An oxygen consumption

test consumption test improved from 14 ml/kg/min initially to 28 ml/kg/min one year after sildenafil therapy. This clinical improvement persisted after 4 years of follow-up with only mild ascites and pleurothorax. Now he can continue his university studies. He refused control cardiac catheterization for assessment of pulmonary pressure.²

Case Report 2

A seven year old boy with mitral atresia, single ventricle, malposition of great arteries was treated from the beginning in our center. His body weight was 17,5 kg. In infancy pulmonary artery banding and surgical atrial septectomy (Blalock-Henlon) was done. Second operation – Glenn procedure without closure of pulmonary outflow from common ventricle was performed. During the subsequent six years he was doing relatively well, without heart failure, with moderate desaturation (81%). At the age of 7 years old, he was admitted to our department for cardiac catheterization (qualification for Fontan completion). This examination revealed increased mean pulmonary artery pressure (mPAP) - 22 mmHg, good pulmonary artery anatomy, and increased pulmonary artery flow due to opened SV-PA outflow. QP/QS was calculated as 3/1. During the same catheterization pulmonary outflow (previous banding place) was closed with 5 mm Amplatzer Atrial Septal Occluder introduced through the right jugular vein. Control hemodynamic assessment revealed no change in pulmonary artery pressure after device outflow closure. After the procedure sildenafil was introduced (2 x 12,5 mg/day) to prepare the pulmonary vascular bed for Fontan operation (elevated mPAP pressure was the risk factor). He was also treated with captopril, spironol and carvedilol. After 6 months he was catheterized again. His pulmonary pressure dropped more than half (mPAP 10 mm Hg), QP/QS was 0.43, PVR 2 Wood units. After 3 months he had a TCPC operation performed with extracardiac tunnel (18 mm). The post-operative period was complicated with Low Cardiac Output Syndrome and right pleurothorax which needed 2 pleurocentesis. During the next few days his clinical condition improved and he was discharged home 17 days after operation on sildenafil (2 x 12,5 mg) and standard medication (captopril, furosemid, spironol, acenocumarol). He remains in good condition after 1 year of follow-up.³

Discussion

The administration of pulmonary vasodilators such as sildenafil has been shown to reduce elevated pulmonary artery pressure.⁴ Goldberg et al⁵ suggested that manoeuvres which increase cardiac output and lower central venous pressure can improve Fontan circulation. In case of PLE, with serum albumin level <2,0 g/dl, he recommended treatment with sildenafil, CD-budesonite or Fontan surgical revision or heart transplantation. Moreover, the same author in a recently published paper showed that sildenafil may be a useful therapy to improve or maintain ventricular performance in selected patients after the Fontan operation.⁶ Our observations in patients presented here, as well as those of Deal and Jacobs,⁷ suggest that chronic pulmonary vasodilator therapy, in addition to chronic diuretics, may become part of the routine long-term therapy in selected Fontan patients. Meadows and Jenkins⁸ in their comprehensive review summarized experience with evaluation, management and treatment of PLE in 18 pts from Boston Children Hospital (without application of sildenafil). Our cases and experience of others⁹) indicate that sildenafil can be used safely and effectively in the treatment of patients with failing Fontan circulations. Interestingly Ovaert et al¹⁰ in her study failed to show significant improvement after 3 months of treatment with bosentan (another type of pulmonary vasodilator) in 10 patients with failing Fontan.

References

1. Moore JW. A cautionary tale for pediatric cardiologists. CCT 2013,11, 1-4.
2. Bialkowski J, Rycaj J, Glowacki J et al. Successful chronic treatment with sildenafil in a patient with end-stage failure following Fontan procedure. Kardiol Pol 2011,69,302-4.
3. Salas Llamas JP, Szkutnik M, Fiszler R, Bialkowski J. Treatment of elevated pulmonary artery pressure in a child after Glenn procedure: pranscatheter closure of pulmonary artery banding with subsequent sildenafil therapy. Kardiol Pol 2012,70,201-3.
4. Lu X, Xiong C, Shan G et al. Impact of sildenafil therapy on pulmonary arterial hypertension in adults with congenital heart disease. Cardiovasc Ther 2010,28,350-5.

5. Goldberg DJ, Dodds K, Rychlik J. Rare problems associated with the Fontan circulation. Cardiol Young 2010, 20 (suppl 3), 113-9.
6. Goldberg DJ, French B, Szwast A et al. Impact of sildenafil on echocardiographic indices of myocardial performance after the Fontan operation. PediatrCardiol 2012,33,689-96.
7. Deal B, Jacobs M. Management of the failing Fontan circulation. Heart 2012, 98, 1098-1104.
8. Meadows J, Jenkins K. Protein-losing enteropathy: integrating a new disease paradigm into recommendations for prevention and treatment. Cardiol Young 2011, 21, 363-77.
9. Reinhardt Z, Uzun O, Bhole V et al. Sildenafil in the management of the failing Fontan circulation. Cardiol Young 2010, 20, 522-5.
10. Ovaert C, Thijs D, Dewolf D et al. The effect of bosentan in patients with failing Fontan circulation. Cardiol Young 2009, 19, 331-9.

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- Optionally, a picture of the author(s) may be submitted.
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- The main text of the article should be written in informal style using correct English. The final manuscript may be between 400-4,000 words, and contain pictures, graphs, charts and tables. Accepted manuscripts will be published within 1-3 months of receipt. Abbreviations which are commonplace in pediatric cardiology or in the lay literature may be used.
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Medical News, Products and Information

Children's Hospital of Philadelphia Experts Present Findings in Pediatric Heart Disease at the 2012 American Heart Association's Scientific Sessions in Los Angeles

Serious Blood Flow Reversal after Heart Surgery in Children -- Cardiac researchers have demonstrated that a combination of imaging techniques can identify a serious reversal of blood flow that occurs in some children after reconstructive surgery for complex heart defects. Using time-resolved gadolinium angiography (TWIST) and magnetic resonance phase contrast velocity mapping (PC-MRI), Kevin K. Whitehead, MD, PhD and colleagues describe their experience in identifying blood flow reversal in children who have undergone superior cavopulmonary connections (SCPC) as part of the Fontan procedure for single ventricle heart defects. Identifying flow reversal in the left pulmonary artery (LPA) and right upper lobe branch (RUL) pulmonary artery is important, because these manifestations of systemic to pulmonary arterial collateral flow (CollF) may result in poor outcomes for these patients. A failure to recognize LPA or RUL flow reversal may also cause clinicians to significantly underestimate CollF. The researchers reviewed 112 SCPC patients who had CollF quantified by MRI, and were able to readily identify LPA or RUL flow reversal in 7% of these patients. The CollF burden was much higher for patients with flow reversal than in those without flow reversal.

Decreasing Mortality of Ventricular Assist Devices at Children's Hospitals from 2000 to 2010: Improvement at a Cost -- Co-morbid Heart Failure Is Linked to Increased Mortality in Single-Ventricle Patients -- More than one in 10 children with single-ventricle heart disease may also be hospitalized with co-morbid heart failure. A retrospective review by Joseph W. Rossano, MD, and colleagues at CHOP found that this little-studied patient population has a higher mortality rate and longer length of stay (LOS) than single-ventricle children not hospitalized for heart failure. The researchers analyzed data from 2000, 2003 and 2006 in the Health Care Cost and Utilization Project Kids Inpatient Database, a nationwide sampling of pediatric hospitalizations. The analysis included 732 heart failure-related hospitalizations (HFRH) among single-ventricle patients in 2000 and 1,168 HFRH in 2006. HFRH patients had LOS nearly twice as long as non-HFRH patients, and their mortality was 50% higher. The HFRH group also had higher rates of morbidities, such as arrhythmias, sepsis and respiratory failure. Non-cardiac morbidities such as cerebrovascular disease, acute renal failure, sepsis and the use of extracorporeal membrane oxygenation (ECMO) were independently associated with hospital mortality.

New Website Helps Parents Manage Children's Pain After Surgery

When a young child has surgery, parents rely on doctors and nurses for advice on how to prepare and support children during the procedure and immediately afterwards. But once that child gets home, parents are left with little guidance on how to best help their children cope with pain.

A new website hopes to fill that information gap, and give parents the framework for how to be more effective caregivers for children after surgery.

Created with Linda Franck, RN, PhD, Chair of Family Health Care Nursing in UC San Francisco's School of Nursing, the website, *My Child is in Pain* (<http://mychildisinpain.org.uk>) was created for parents of children ages two to six who want to know how to help manage their child's post-operative pain.

"There are very few formal resources for parents to learn how to tell if their child is in pain and what they can do to relieve it," said Franck.

Over 80% of the more than 3 million children's surgeries in the U.S. are performed on an outpatient basis, leaving parents to manage post-operative pain at home. But returning home can be scary when parents aren't confident about how to determine if their child is in pain, and children aren't equipped with the language skills to fully articulate how they are feeling. Franck's research has shown that over 90% of children have pain two days after surgery and as many as 25% have pain four weeks after. Children with post-operative pain also were more likely to have problematic behaviors such as not sleeping, eating poorly, and being very anxious.

"Parents are not getting enough information or feeling comfortable using the information that's out there to manage pain at home," said Franck. "It became clear we needed to put together another resource using the best research evidence available and lots of parent input so that it was practical and useful for parents."

Franck has dedicated her career to pioneering pain assessment and management techniques for acutely and chronically ill infants and children. Her research highlights the information needs of parents when their children are in pain, and suggests innovative strategies for enhancing the partnership between parents and professionals to ensure children receive optimal pain care.

Through a series of videos, text and illustrations, the website helps parents understand how children respond to pain and guides them through how to tell if their child is in pain. It provides detailed information and practical advice on some of the simple but effective things they can do to provide effective comfort and manage their child's pain. The website also explains how pain is signaled to the brain, how common pain medications work, what times are best to administer the medication and what to do if it's not working. The site is funded by the United Kingdom based non-profit WellChild (www.wellchild.org.uk).

"When a child's pain is well-managed, he or she usually recovers more quickly," said Franck. "Simple comfort techniques can be very effective. Gently rocking and stroking their child can be soothing and help their child to relax, and when a child is relaxed and calm it can help the pain go away. Also, letting their child make some decisions can help them feel less anxious and more in control."

The project team was based in the UK, led by Bernie Carter, PhD from the School of Health at the University of Central Lancashire, and including Lucy Bray, PhD from Edge Hill University and Nic Blackwell, PhD, from OCB Media.

UCSF is a leading university dedicated to promoting health worldwide through advanced biomedical research, graduate-level education in the life sciences and health professions, and excellence in patient care.



Program topics will include management strategies of acute heart failure syndromes, methods of hemodynamic and physiologic monitoring, renal protective techniques, and updates on mechanical circulatory support in children.

When: October 10-12, 2013

Where: Houston Texas

Website: <http://www.texaschildrenshospital.org/phfs2013/>

ICUs for Newborns in Nine States See Sharp Drop in Bloodstream Infections

Newswise — Central Line Associated Bloodstream Infections (CLABSIs) in newborns were reduced by 58% in less than a year in hospital neonatal intensive care units (NICUs) participating in an Agency for Healthcare Research and Quality (AHRQ) patient safety program. Frontline caregivers in 100 NICUs in nine states relied on the program's prevention practice checklists and better communication to prevent an estimated 131 infections and up to 41 deaths and to avoid more than \$2 million in health care costs.

CLABSIs are Healthcare-Associated Infections (HAIs) that cause serious illness and death in infants as well as adults. A central line is a tube (catheter) that goes into a patient's vein or artery and ends in the central bloodstream. In newborns, especially premature infants, central lines can remain in place for weeks or months to provide nutrients and medications as babies become able to function on their own.

Health care teams in the project states, caring for a total of 8,400 newborns, used AHRQ's Comprehensive Unit-based Safety Program (CUSP) to improve safety culture and consistently implement catheter insertion and maintenance guidelines. CUSP is customizable and helps hospitals understand and apply the science of safety and take actions to improve teamwork and communications. This 11-month project used CUSP to help clinical teams focus on safe practices and appropriate steps when using central lines based on guidelines from the Centers for Disease Control and Prevention.

Each state-based team was led by a neonatologist who worked with the state's hospital association to implement the project. When the project began, participating NICUs had an overall infection rate of 2.043 per 1,000 central line days. At the end of the project, that rate was reduced to 0.855 per 1,000 central line days, a relative reduction of 58%. For more information on how NICUs achieved this reduction, visit www.ahrq.gov/qual/clabsi-neonatal/.

"The CUSP framework brings together safety culture, teamwork and best practices—a combination that is clearly working to keep these vulnerable babies safer," says AHRQ Director Carolyn M. Clancy, MD. "These remarkable results show us that, with the right tools and dedicated clinicians, hospital units can rapidly make care safer."

The nine-state project in NICUs is part of a larger AHRQ-funded effort to implement CUSP to prevent CLABSIs nationwide. Preliminary results of the larger project were announced in September 2012; final results from the national implementation project are now available and show that CLABSIs were reduced by 41 percent in adult ICUs. The final report is available at www.ahrq.gov/qual/clabsi-final/.

AHRQ provided funding to the Health Research & Educational Trust (HRET), the educational arm of the American Hospital Association (AHA), to conduct both projects. For the NICU project, HRET partnered with the Perinatal Quality Collaborative of North Carolina and the Missouri Center for Patient Safety to support Colorado, Florida, Hawaii, Massachusetts, Michigan, New Jersey, North Carolina, South Carolina and Wisconsin.

"The successes of the project are proof that a great deal of improvement can happen in a relatively short timeframe," says Maulik S. Joshi, DrPH, president of HRET and senior VP of the AHA. "We are excited by the outcomes of the collaboration, and we look forward to applying what we've learned about leveraging existing infrastructures to spread improvement in ongoing and future projects."

AHRQ's HAI Program contributes to the U.S. Department of Health and Human Services' National Action Plan to Prevent Healthcare-Associated Infections (www.hhs.gov/ash/initiatives/hai/index.html) and the Partnership for Patients (www.healthcare.gov/compare/partnership-for-patients), which offer a coordinated approach to making care

safer by drawing on the strengths and expertise of the HHS agencies.

Details about AHRQ's CUSP projects, including a report on the NICU project and the final report from the national implementation project, are available at www.ahrq.gov/qual/hais.htm. AHRQ's CUSP toolkit, which was developed from the national implementation project and used in the NICU project, is available at www.ahrq.gov/cusptoolkit/.

Hospital Readmissions: A Look at Pediatric Hospitals' Striking Differences in 30-Day Readmission Rates May Inform Prevention Efforts

Unintended hospital readmissions have become a key quality-of-care indicator, prompting penalties to adult hospitals with a high rate of patient readmissions within 30 days. Using national data, a study led by Boston Children's Hospital looked at readmission rates at dedicated pediatric hospitals and found great variability. Findings appear in the January 23/30 issue of *JAMA*, accompanied by an editorial.

The researchers believe there may be several reasons for the varied readmission rates, including differences in hospital care, follow-up care outside of the hospital, and community and family factors that may influence child health.

"Some hospitals and their local health systems had very low readmission rates for diseases that, on average, tend to have much higher rates," says first author Jay Berry, MD, MPH, a pediatrician in the Complex Care Service at Boston Children's Hospital. "We want to know whether there is something these hospitals and systems are doing to more effectively transition their children home."

Berry, senior investigator Mark Schuster, MD, PhD, Chief of General Pediatrics at Boston Children's Hospital, and their colleagues analyzed 568,845 admissions to 72 large tertiary-care children's hospitals from July 2009 through June 2010. They used data from the National Association of Children's Hospitals and Related Institutions (NACHRI) Case Mix, adjusting hospitals' readmission rates for chronic conditions that increase the risk of re-hospitalization. (NACHRI is part of what's now called the Children's Hospital Association.)

Overall, 6.5% of children had apparently unplanned readmissions to the hospital within 30 days of discharge, and of these, 39% were readmitted within 7 days. By contrast, reported readmission rates at adult hospitals range from 20 to 25%. Two-thirds of readmissions were in children with at least one chronic condition; for certain medical conditions, readmission rates were as high as 23%.

The 30-day readmission rates varied among the 72 hospitals, ranging from 4.6 to 8.5. Other findings:

- Readmission rates were 6.9% for patients with public insurance (e.g., Medicaid), 5.9% for those with private insurance and 4.5% for those with no insurance.
- Rates ranged from 5.4% for children with one chronic condition to 17% for those with four or more.
- Ten conditions accounted for the highest readmission rates: anemia/neutropenia, ventricular shunt procedures, sickle-cell crisis, seizures, gastroenteritis, upper respiratory infection, pneumonia, appendectomy, bronchiolitis and asthma.
- Children with the above 10 conditions accounted for 28% of all readmissions. Their readmission rates were 17 to 66% higher in hospitals with higher-than-average readmission rates than in low-readmission hospitals.
- Readmission rates were higher for patients with longer hospital stays, from 4.6% for patients with a 1- to 2-day stay to 11.2% for patients with stays of 7 days or longer.

"Some readmissions cannot be prevented, but various efforts to reduce readmissions by providing better supports for families have been successful at bringing rates lower," notes Schuster. "The varia-

tion we found in readmission rates at different hospitals suggests that there is room to improve. The effort, though, will involve more than just hospitals. Community clinicians and organizations have a role to play as well. Parents also need support in being able to stay home with their recuperating children.”

“Let’s put the child and family first,” says Berry. “There are some children with complicated medical needs who have really high readmission rates. Let’s figure out what’s going on and see if there is an opportunity to make their care transitions better.”

The study was funded by the Agency for Healthcare Research and Quality, the Centers for Medicare and Medicaid Services, and the Eunice Kennedy Shriver National Institute of Child Health and Human Development.

Abnormal Growth Regulation May Occur in Children with Heart Defects

The poor growth seen in children born with complex heart defects may result from factors beyond deficient nutrition. A new study by pediatric researchers suggests that abnormalities in overall growth regulation play a role.

“When compared with their healthy peers, children with congenital heart disease have impaired growth, as measured in weight, length, and head circumference,” said senior author Meryl S. Cohen, MD, a pediatric cardiologist in the Cardiac Center at The Children’s Hospital of Philadelphia. “We investigated patterns of poor growth in these children, as a starting point in guiding us toward more effective treatments.”

The study appeared as an online article in the January 2013 issue of *Pediatrics*.

The researchers performed a retrospective analysis of medical records of 856 children with Congenital Heart Disease (CHD), compared to 7,654 matched control subjects. All the children were measured up to age 3, and all were drawn from the healthcare network of The Children’s Hospital of Philadelphia.

Within weeks of birth, the children with CHD had significant deficits in weight, length and head circumference, compared to matched controls without CHD. The largest differences in weight occurred at 4 months of age. Among the 856 children with CHD, the 248 who required surgical repair were much more likely to be below the 3rd percentile in weight, length and head circumference during early infancy, and their growth by age 3 did not catch up with that of their healthy peers.

In the 608 children with CHD who did not require surgery, growth differences were not as pronounced, but even their growth patterns lagged behind those of healthy controls.

Findings suggest impaired growth in children with CHD at least partly affected by factors unrelated to nutrition.

Researchers already knew that children with CHD have an increased risk for poor growth, but this analysis provides a fuller picture of the problem. Cohen observed that in the general population, when caloric intake is insufficient, an infant’s weight is usually affected first, followed by length and head circumference. “The fact that all three parameters changed simultaneously rather than sequentially supports the idea that impaired growth in children with heart disease is affected at least in part by factors unrelated to nutrition.”

She added that further studies should investigate the possible roles of growth hormones and other physiologic factors that affect growth regulation in children with CHD.

Cohen’s co-authors were first author Carrie Daymont, MD, of the Uni-

versity of Manitoba, Ashley Neal, MD, of Children’s Hospital Boston, and Aaron Prosnitz, MD, of Yale-New Haven Children’s Hospital. All were at The Children’s Hospital of Philadelphia when the research was performed.

“Growth in Children with Congenital Heart Disease,” *Pediatrics*, Jan. 2013, pp. e236-e242.

Annual meeting of the Association for European Paediatric and Congenital Cardiology (AEPC), London 2013

The Association for European Paediatric and Congenital Cardiology (AEPC) was founded in Lyon in 1963 and over the years has created a network of specialists working in the same field. The mission of AEPC is to promote knowledge of the normal and diseased heart and circulation and to exchange knowledge and provide a forum for continuous education.

There are over 1000 members who include paediatric cardiologists and other specialists working in the field of paediatric cardiology and its related disciplines. AEPC is therefore the largest democratically administered global association in the field of congenital cardiology, and now has members from all the continents. The official journal of the Association is *Cardiology in the Young*. AEPC has encouraged collaboration with many associations around the world and has close relationship with European Society of Cardiology (ESC).

There are 11 Working Groups of the AEPC representing different subspecialties of paediatric and congenital cardiology (including a GUCH Task Force). These groups collaborate and facilitate research in closely related fields. They also work closely with the organisers of the annual AEPC meetings to develop a top quality scientific programme.

AEPC holds an annual meeting in the third week of May and the meeting rotates in different European countries. The meeting traditionally includes an Update/Teaching course organised by different Working Groups. This year, the 47th Annual AEPC meeting is being held on 22-25 May 2013 at the Hilton London Metropole Hotel, London, UK. London is a vibrant, multicultural city, with many tourist attractions. This is the first time that the AEPC meeting is being held in London and over 1000 attendees from all parts of the world are expected to attend, providing an excellent opportunity to share up to date knowledge and networking with colleagues.

This year the meeting will begin on Wednesday 22nd May with the Update course organising by the AEPC Imaging and Surgical Working Groups. This will be followed by the official start to the meeting with the prestigious Edgar Mannheimer lecture, given this year by Professor John Hess from Munich, Germany. He will be talking on “Impact of changing patterns and technologies for congenital heart disease on long term outcomes”. The main meeting includes Plenary sessions and other sessions organised by various AEPC Working Groups, abstract presentations, moderated poster and other poster presentations and several industry sponsored symposia. An important scientific part of the meeting includes collaboration between different societies. These include the joint sessions between the AEPC and European Association of Cardiothoracic Surgery (EACTS), the AEPC and the Japanese Society of Pediatric Cardiology and Cardiac Surgery (JSPCCS) and AEPC and Asia Pacific Paediatric Cardiology Society (APPCS). There is a Young Investigators Award session during which the 6 best abstracts from European and Japanese researchers will be presented. An addition to this year’s programme is the inclusion of the 9th Annual Multi-Societal Database Committee for Pediatric and Congenital Heart Disease meeting, which will be held on Thursday 23 May.

The AEPC meeting will conclude with Keynote lectures on “the Past, Present and Future of Paediatric Cardiology” to be given by Professors Michael Tynan and Shakeel Qureshi.

For more information can be obtained on www.aepc.org

Abnormal Growth Regulation May Occur in Children with Heart Defects

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The study appeared as an online article in the January 2013 issue of *Pediatrics*.

The researchers performed a retrospective analysis of medical records of 856 children with congenital heart disease (CHD), compared to 7,654 matched control subjects. All the children were measured up to age 3, and all were drawn from the healthcare network of The Children's Hospital of Philadelphia.

Deficits in weight, length and head circumference occurred within weeks of birth

Within weeks of birth, the children with CHD had significant deficits in weight, length and head circumference, compared to matched controls without CHD. The largest differences in weight occurred at 4 months of age. Among the 856 children with CHD, the 248 who required surgical repair were much more likely to be below the 3rd percentile in weight, length and head circumference during early infancy, and their growth by age 3 did not catch up with that of their healthy peers.

In the 608 children with CHD who did not require surgery, growth differences were not as pronounced, but even their growth patterns lagged behind those of healthy controls.

Findings suggest impaired growth in children with CHD at least partly affected by factors unrelated to nutrition

Researchers already knew that children with CHD have an increased risk for poor growth, but this analysis provides a fuller picture of the problem. Cohen observed that in the general population, when caloric intake is insufficient, an infant's weight is usually affected first, followed by length and head circumference. "The fact that all three parameters changed simultaneously rather than sequentially supports the idea that impaired growth in children with heart disease is affected at least in part by factors unrelated to nutrition."

She added that further studies should investigate the possible roles of growth hormones and other physiologic factors that affect growth regulation in children with CHD.

Cohen's co-authors were first author Carrie Daymont, MD, of the University of Manitoba, Ashley Neal, MD, of Children's Hospital Boston, and Aaron Prosnitz, MD, of Yale-New Haven Children's Hospital. All were at The Children's Hospital of Philadelphia when the research was performed. "Growth in Children with Congenital Heart Disease," *Pediatrics*, Jan. 2013, pp. e236-e242.

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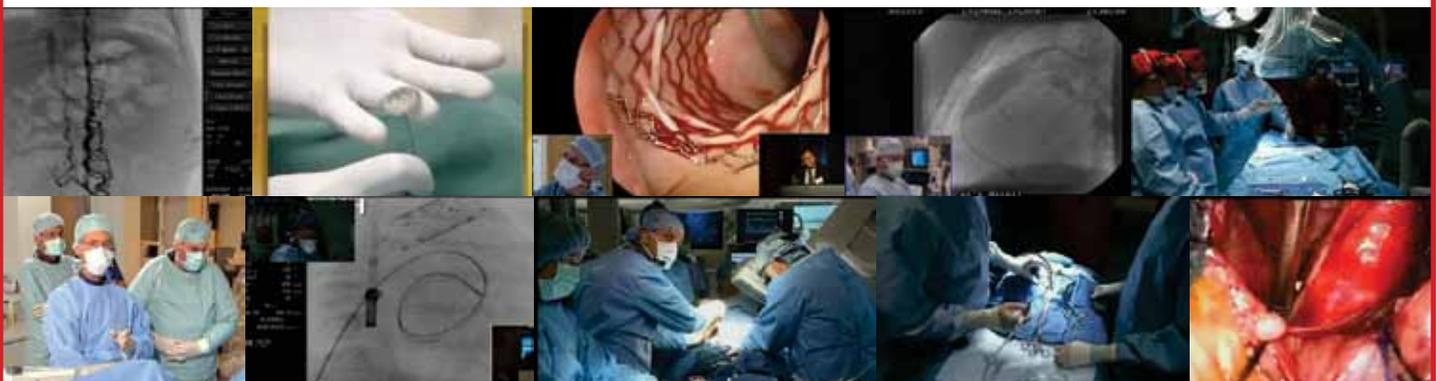
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