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Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

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Pulse Oximetry Screening for Unrecognized Congenital Heart Disease in Neonates

By John S. Hokanson, MD

Background

Congenital heart disease is the most common serious birth defect in humans. Many newborns with ductal-dependent heart disease will appear to be entirely well at the time of routine hospital discharge only to become critically ill a few days later.

Various estimates suggest that potentially life-threatening congenital heart disease is present in approximately 1:1,000 births¹⁻⁵ (Table 1). Unfortunately this heart disease will go unrecognized in some neonates until symptoms develop. These delayed or missed diagnoses can result in both disability and death. The incidence of a missed diagnosis of critical congenital heart disease can be defined in various terms and occurs in anywhere from 1 in 3,500 to 1 in 25,000 live births.^{1-4, 6} The low

incidence of missed congenital heart disease in our data from Wisconsin may be related to limiting the definition to death or readmission due to critical congenital heart disease occurring at less than 14 days of age.⁶

A retrospective analysis of patients admitted to the Children's Hospital of Philadelphia (CHOP) with critical congenital heart disease at less than 30 days of age suggested that 6.7% had a "significant physiologic compromise due to a missed diagnosis of critical congenital heart disease."⁷ This study could not evaluate the number of babies who died prior to diagnosis of congenital heart disease and transfer to CHOP and may underestimate the consequences of a missed diagnosis of critical congenital heart disease.

Studies of death due to unrecognized congenital heart disease suggest that the incidence of death due to missed congenital

Years			Table 1. Incidence of Missed Diagnosis of Critical Congenital Heart Disease										
Tears	Incidence of Critical Congenital Heart Disease	Missed or Delayed Diagnosis	Death Due to Mixed Dx	Deaths per Live Births	Location								
1985-2004	1:1,032	1:3,486	1:23,007	30/690,215	Northern Heath Region, UK								
1993-2001	1:1,135	1:6,899	Not Reported		Sweden								
1999-2004	1:971	1:14,261	Not Reported		New Jersey								
2002-2006	Not Reported	1:24,684	1:38,397	9/345,572	Wisconsin								
2004-2007	1:853	1:3,878	1:21,721	5/108,604	Sweden								
1	985-2004 993-2001 999-2004 002-2006 004-2007	Congenital Heart Disease 985-2004 1:1,032 993-2001 1:1,135 999-2004 1:971 002-2006 Not Reported	Congenital Heart Disease Delayed Diagnosis 985-2004 1:1,032 1:3,486 993-2001 1:1,135 1:6,899 999-2004 1:971 1:14,261 002-2006 Not Reported 1:24,684 004-2007 1:853 1:3,878	Congenital Heart Disease Delayed Diagnosis Mixed Dx 985-2004 1:1,032 1:3,486 1:23,007 993-2001 1:1,135 1:6,899 Not Reported 999-2004 1:971 1:14,261 Not Reported 002-2006 Not Reported 1:24,684 1:38,397 004-2007 1:853 1:3,878 1:21,721	Congenital Heart Disease Delayed Diagnosis Mixed Dx Live Births 985-2004 1:1,032 1:3,486 1:23,007 30/690,215 993-2001 1:1,135 1:6,899 Not Reported 999-2004 1:971 1:14,261 Not Reported 002-2006 Not Reported 1:24,684 1:38,397 9/345,572 004-2007 1:853 1:3,878 1:21,721 5/108,604								

* Does not include those dying before diagnosis

** Limited to missed diagnosis or death under the age of 14 days

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Texas Children's Hospital Section of Critical Care has 87 critical care beds, in three clinical areas: Cardiovascular Intensive Care Unit, the Pediatric Intensive Care Unit, and the Progressive Care Unit. The CVICU has recently expanded from 12 to 21 beds, and now admits all newborns with critical heart disease, as well as all post-operative patients, selected medical patients, and all children requiring acute mechanical support for cardiac disease, and VAD support as a bridge to cardiac transplantation. Each year, we care for about 900 children after cardiac surgery, of which approximately two-thirds have undergone open surgery requiring cardiopulmonary bypass. Texas Children's Hospital is the coordinating center for the FDA trial of mechanical cardiac support in pediatric patients. Since 2007, we have placed 40 long-term devices in children with end-stage heart failure, and currently we are the only institution using the Heartmate II device in children.

Texas Children's Hospital is the primary affiliated teaching hospital in pediatrics for Baylor College of Medicine. Closely affiliated in pediatric medicine since 1954, they are committed to driving the innovation that will transform the future of pediatric healthcare.

heart disease occurs in 1 in 20,000 to 1 in 40,000 births. If extrapolated to the US birth rate of roughly four million per year, somewhere between 100 and 200 deaths due to unrecognized heart disease in newborns would be expected each year. Although not indexed for the birth rate, a study by Chang⁸ suggested that there might be as many as 30 deaths per year due to unrecognized critical congenital heart disease in California alone.

Screening for Unrecognized Heart Disease in Other Settings

Significant efforts have been undertaken in recent years to screen for heart diseases that may result in sudden death in children, particularly in athletes and in those taking stimulant medications. The incidence of sudden death due to unrecognized heart disease in a child between 1 and 20 years of age has been reported in the range of 1:100,000 per year.⁹ The sudden death of a high school athlete due to unrecognized heart disease occurs in roughly 1:200,000 per year.¹⁰ Overall, the sudden death of a young athlete (up to the age of 39 years) during exercise occurs less than 100 times each year in the United States¹¹. Although an association has been suggested between the use of stimulant medications and sudden death in children, the available literature suggests that this occurs less than ten times per year in the United States¹²⁻¹⁴ (Table 2). Based in part on a small number of adverse effects reported in Canada, recommendations to consider ECG screening of children taking stimulant medications were made by the AHA and AAP in 2008.15

Table 2. Incidence of Death Due to Unrecognized Heart Disease				
Cause	Estimated Number of Deaths Per Year in US			
Unrecognized Critical Congenital Heart Disease in Neonates	100-200			
Sudden Death of a Young Athlete	<100			
Sudden Death Associated with Stimulant Medication Use	<10			

In de-Wahl Granelli's study⁴ there were 5 deaths due to unrecognized congenital heart disease in the 108,604 babies in the control arm and no deaths in the 38,429 babies in the population in which pulse oximetry screening was performed. Although not designed to test the hypothesis, her study suggests that the implementation of pulse oximetry screening decreases the risk of death due to a missed diagnosis of critical congenital heart disease. No such population based data exists for the implementation of screening strategies to decrease the incidence of death in athletes or those taking stimulant medications.

Detection of Congenital Heart Disease and the Cyanotic Blind Spot

Traditionally, congenital heart disease is detected prenatally with obstetric ultrasound or postnatally by physical examination or the development of symptoms. The prenatal diagnosis of congenital heart disease is the preferred mechanism, but data from both the US and UK suggests that most children with critical congenital heart disease are not detected prior to birth.16-17



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Physical examination of the newborn is the oldest method for detecting congenital heart disease prior to symptoms and remains invaluable, but has significant limitations. Certain types of critical ductal-dependent congenital heart disease will not be detected, even by experienced clinicians in the first days after birth. In the setting of valvar atresia or single ventricle physiology with systemic pulmonary pressures, there may not be a heart murmur to alert the clinician to the presence of heart disease. With a large PDA supporting the systemic circulation the femoral pulses may well be normal.

A major limitation of the newborn physical examination is the inability for the human eye to detect important degrees of cyanosis. The limits of visual recognition of cyanosis are well documented, but are frequently underappreciated. Nearly a century ago, it was suggested that between 4 and 6 grams of deoxygenated hemoglobin per deciliter of blood would be necessary for central cyanosis to be visible¹⁸ (Lundsgaard & Van Slyke 1923). Later reports suggested that only 3 grams of deoxygenated hemoglobin would be necessary to manifest central cyanosis¹⁹ (Lees 1970). Even if only three grams of deoxygenated hemoglobin need be present for the observation of central cyanosis, this still leaves a wide gap between normal saturation and visible cyanosis, the cyanotic blind spot (Figure 1). In a one day old term baby with a hemoglobin at the 50th percentile (17.5 g/dL),²⁰ cyanosis would be visible at or below approximately 83%. The cyanotic blind spot widens with anemia and in a one day old term baby with a hemoglobin at the 5th percentile (13.5 g/dL),20 cyanosis would not be visible until the saturation had dropped to 78% or below.

More recent work by O'Donnell²¹ suggests that both the ability to visually detect cyanosis and the inter-observer reliability of visual observations of cyanosis are poor even among neonatal intensive care personnel. This study was performed in the delivery room where a rapid increase in oxygen saturation is expected. The threshold for the resolution of cyanosis varied from 10% to 100% between the observers. Although the infants' hemoglobin concentrations were not reported in this study, the mean threshold saturation for the visible resolution of cyanosis was 69%.

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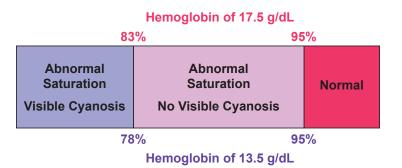


Figure 1: The Cyanotic Blind Spot

Pulse Oximetry Screening to Detect Unrecognized Ductal Dependent Heart Defects

The use of pulse oximetry to detect cyanosis in asymptomatic term neonates as a screening for critical congenital heart disease has been studied for several years. The use of pulse oximetry for this purpose has been viewed with scrutiny, as any intervention applied on a large scale should be. Although pulse oximetry could be considered an additional vital sign, the presence of a fixed cut-off value between normal and abnormal is not how other vital signs are usually considered. Pulse oximetry measurements differ from other vital signs in that there is data to suggest a relatively rigid differentiation between normal and abnormal in the baby greater than 24 hours old. Curiously, in many facilities the only inpatient population in which pulse oximetry is not routinely performed is the normal neonate.

Studies of Pulse Oximetry Screening

Although the concept supporting the use of pulse oximetry as a screening tool is elegant in its physiologic simplicity, implementation of pulse oximetry screening is another matter. The available data on the

subject suffers from a wide variety of study designs, study populations, and measures of outcome.^{2, 4-5, 22-31}

The sensitivity and specificity of pulse oximetry as a screening tool in these studies is highly variable and is influenced by the prenatal detection rate, the timing of screening, and the use of both pre- and post-ductal measurement, or of post-ductal measurements alone. In general, the earlier the screening is performed, the greater the sensitivity and the lower the specificity. In most studies, a post-ductal saturation between 94% and 96% has been used as the cut-off point, as the mean saturation in term neonates at 24 hours of age has been reported to be 97.2% +/- 1.6%.³² Other protocols have used a difference between preductal and post-ductal saturations as an additional indicator of critical ductal-dependent heart disease,^{4,23} although there is only one large scale study of such an approach.⁴

The false positive and positive predictive values are dependent on both the timing of the oximetry and whether or not a repeat measurement of abnormal values was performed. It appears that screening performed either primarily after 24 hours, or repeated after 24 hours will provide the lowest false positive rate and the highest positive predictive value. In comparing the two large studies of pulse oximetry after 24 hours of age, ^{4, 30} the value of pre-ductal oximetry is difficult to assess. One patient with interrupted aortic arch and aortopulmonary window had pre- and post-ductal oximetry alone. The authors of this study report no additional false positives based on the addition of the pre-ductal oximetry.⁴

Although a detailed discussion of individual study design is beyond the scope of this article, the false positive rates and positive predictive values of the more recent studies are presented in Table 3.

Public Policy Initiatives

In 2005, routine pulse oximetry was recommended by the Swiss Society of Neonatology and the Swiss Society of Pediatric Cardiology. By 2007, 85% of Swiss newborns were screened for congenital heart disease with pulse oximetry using a standardized protocol.³³ This protocol utilized post-ductal saturations measured on the first day with abnormal considered to <95%. Echocardiography was performed if repeat measurements remained less than 95%.

In 2005, mandated pulse oximetry screening was proposed in the State of Tennessee. A task force was assembled to determine the utility of such legislation. The available literature at the time comprised four studies of a total of less than 22,000 patients with wide variability in screening protocol and study design.²²⁻²⁵ Based on this limited data, the task force did not recommend pulse oximetry screening.³⁴

A probabilistic cost-effectiveness model reported from the UK in 2007 suggested that addition of pulse oximetry to the routine evaluation of newborns was likely to be cost effective.³⁵

In 2009, the American Heart Association and American Academy of Pediatrics published their scientific statement on the use of pulse oximetry in screening for congenital heart disease.³⁶ This review was completed prior to the publication of Riede's study of 41,445 neonates,³⁰ and concluded that further study was required prior to large scale implementation of routine pulse oximetry.

On September 17th, 2010, the Secretary's Advisory Committee for Hereditable Disorders in Newborns and Children recommended that pulse oximetry screening be added to the core panel for universal screening of newborns (www.hrsa.gov/heritabledisorderscommittee/ default.htm). The US Secretary of Health and Human Services, Kathleen Sebelius, will respond to these recommendations within 180 days. If approved, this recommendation will be forwarded to the individual states for implementation.

Implications of Population-Based Screening

Several factors must be considered regarding the population based implementation of pulse oximetry screening. The published data on this topic is gathered from different health care delivery systems with varying prenatal and postnatal detection rates. Much of the concern regarding pulse oximetry regards the impact of the false positive study. Except in Walsh's study,³¹ those children failing oximetry screening proceeded to echocardiography, which may not be immediately available in all settings.

No matter how screening is performed, false positive results will result in increased cost, delay in discharge, and anxiety. However, when echocardiography cannot be performed without transfer to another center the costs, delays, and anxieties associated with false positive studies will increase considerably. The application of pulse oximetry screening to rural settings with limited access to echocardiography may be challenging. In Wisconsin alone, half of the state's children were born in one of the 98 hospitals delivering less than 1250 babies per year and a quarter were born in one of the 80 hospitals delivering less than 675 babies per year.⁶

However, when performed after 24 hours and with repeat screening performed after an initial screening failure, the positive predictive value of pulse oximetry for potentially life-threatening heart disease is between 21% and 26%.^{4, 30} This data suggests that in settings where echocardiography is not available, it may be reasonable to extend the hospitalization of an asymptomatic newborn to allow for additional evaluation and to allow transitional circulation to resolve or to transfer the baby to a facility where echocardiography can be performed.

Table 3. Studies of Oximetry Screening									
Author	Year	Patients	Timing	Sites	Normal	False Positive Rate	Positive Predictive Value		
Sendelbach	2008	15299	4 hours repeat before d/c	foot	≥ 96%	1:15,233 for CHD	(0/1) 0% for CHD		
de-Wahl Granelli	2009	39821	38 hours repeated up to three times	hand and foot	≥ 95% OR ≤ 3% difference	1:557 for CHD 1:1601 for any disease	21% for CHD 72% for any disease		
Merberg	2009	50008	5 hours repeat in 2-3 hours if abnormal	foot	≥ 95%	1:178 for CHD 1:373 excluding transitional circulation	13% for CHD 56% for CHD or transitional circulation		
Walsh	2009	14564	>24 hours no repeat if abnormal	foot	≥ 94%	1:311 for CHD	1%		
Riede	2010	44240	>24 hours repeat in 1 hour if abnormal	foot	≥ 96%	1:1036 for CHD 1:3454 for any disease	26% for CHD 78% for any disease		



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Conclusion

When performed after 24 hours and repeated if abnormal, the use of pulse oximetry is a viable method of screening asymptomatic neonates for critical congenital heart disease. Based on the morbidity and mortality related to the missed diagnosis of congenital heart disease in the newborn and the growing body of evidence demonstrating the benefits pulse oximetry screening, the use of pulse oximetry is likely to become more widespread in the near future.

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Melody® Transcatheter Pulmonary Valve Ensemble® Transcatheter Valve Delivery System Indications for Use:

The Melody TPV is indicated for use as an adjunct to surgery in the management of pediatric and adult patients with the following clinical conditions:

- Existence of a full (circumferential) RVOT conduit that was equal to
 or greater than 16 mm in diameter when originally implanted and
- Dysfunctional RVOT conduits with a clinical indication for intervention, and either:
 - -regurgitation: ≥ moderate regurgitation, or -stenosis: mean RVOT gradient ≥ 35 mm Hg

Contraindications: None known.

Warnings/Precautions/Side Effects:

- DO NOT implant in the aortic or mitral position. Preclinical bench testing of the Melody valve suggests that valve function and durability will be extremely limited when used in these locations.
- DO NOT use if patient's anatomy precludes introduction of the valve, if the venous anatomy cannot accommodate a 22-Fr size introducer, or if there is significant obstruction of the central veins.
- DO NOT use if there are clinical or biological signs of infection including active endocarditis.
- Assessment of the coronary artery anatomy for the risk of coronary artery compression should be performed in all patients prior to deployment of the TPV.
- To minimize the risk of conduit rupture, do not use a balloon with a diameter greater than 110% of the nominal diameter (original implant size) of the conduit for pre-dilation of the intended site of deployment, or for deployment of the TPV.
- The potential for stent fracture should be considered in all patients who undergo TPV placement. Radiographic assessment of the stent with chest radiography or fluoroscopy should be included in the routine postoperative evaluation of patients who receive a TPV.
- If a stent fracture is detected, continued monitoring of the stent should be performed in conjunction with clinically appropriate hemodynamic assessment. In patients with stent fracture and significant associated RVOT obstruction or regurgitation, reintervention should be considered in accordance with usual clinical practice.

Potential procedural complications that may result from implantation of the Melody device include: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, and pain at the catheterization site.

Potential device-related adverse events that may occur following device implantation include: stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, and hemolysis. For additional information, please refer to the Instructions for

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Neonatal Cardiac Rhabdomyoma in Twin Boys

Bv Samir Atmani. MD

Introduction

Symptomatic cardiac rhabdomyomas and obstructive diffuse forms constitute a rare entity at birth. They are associated with tuberous sclerosis of Bourneville (TSB) in about two thirds of cases. The diagnosis is based on the ultrasound and the prognosis depends on their localization. We report a diffuse and obstructive form of TSB in a twin.

Observation

We reported cases of Ayman and Ahmed respectively full-term twin neonates of consanguineous parents. Upon birth, they presented with both perioral cyanosis and respiratory distress. At admission, SaO₂ was at 77% in the first twin, and 70% in the second, with signs of respiratory inconsistency. The cardiac examination revealed a continual murmur in both cases, in addition to pulmonary systolic murmur in the second case. Both infants had hypo-pigmented and achromic spots remarkable on the lower limbs, at the abdominal level and the chest (six spots on the first case, and 8 spots on second) [Figure 1]. After oxygenation treatment using oxygen mask, SaO₂ had improved and became respectively 98% in the first case, and 97% in the second. The Thoracic radiography objectified a diffuse bronchial infiltration both twins.



Figure 1: Numerous achromic spots on lower limbs.

In both twins, the ultrasound exploration demonstrated a PDA and numerous small lesions disseminate in the two ventricles with a large obstructive mass measuring 2 cm suspended in right outflow truck and the pulmonary valve within the second twin [Figure 2]. The cerebral CT-scan showed typical Bourneville tuberous sclerosis lesion.

Antibiotic and propanonol treatments were started immediately. Progressively, the first case status improved, whereas, the second infant died suddenly, probably by pulmonary trunk obstruction.

At three months of life, the surviving twin developed epilepsy, which is now controlled successfully by sodium vaproate. The clinical follow-up showed non-recurrent symptoms and serial ultrasound examination showed a decreased tumor mass.

Discussion

The cardiac tumors in child and fetus represent about 1% of cardiac disorders diagnosed inutero. The most frequent histological form in the fetus is the cardiac rhabdomyoma.^{1,2,3} Fortyfour cases were rhabdomyomas among a cardiac series of 56 tumors collected along several decades. 60-80% of children with rhabdomyoma are STB.4

The diagnosis is evoked by the presence of large echogenic masses, inserted in the cardiac cavities likely blocking the atrioventricular outflow and the aortic or pulmonary ejection.^{5,6} They might be single or multiple, either in the interventricular septum or in walls of the two ventricles, or exceptionally in the atrium walls.3,5,6

The prenatal diagnosis using antenatal ultrasound is possible after 22 weeks of amenorrhea.3

Hydrops foetalis, conductive disorders, or hypertrophic myocardiopathy by massive infiltration are the main prenatal manifestation. ⁶ Intrauterine death, as well as sudden death, immediately after birth has attributed to them. After delivery, these tumors are usually asymptomatic,^{6,7} and might be discovered during a routine ultrasound screening in TSB condition.6 However infrequently, they could initiate neonatal cardiac deficiency by obstruction or rate/rhythm disorders as Wolf-Parkinson White Syndrome. Also respiratory distress or thrombo-embolic stroke could be the revelator symptom.6, 7, 8

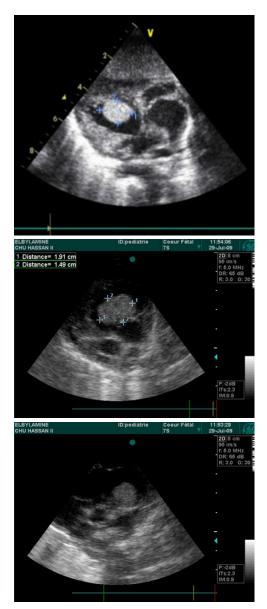


Figure 2 (top); Figure 3 (middle); Figure 4 (bottom): Enormous endocardic mass 2 cm size, appended to the pulmonary outflow truck.

The clinical examination could be normal, or associating TSB signs. The cardiac examination might show systolic pulmonary or aortic murmur such found in the case of the second twin. The thoracic radiography could be normal.





Doppler-ultrasound is the key diagnostic tool demonstrating the masses, their extension and localization, specifying their number, and assesses their hemodynamic characteristics.⁶

Although difficult to achieve in the neonate, the cardiac MRI allows a better study of the parietal infiltration.^{3,9}

Rhabdomyomas have been known to spontaneously regress. However, serious symptoms may precipitate the need need for surgical removal. Such as our second twin, who probably had, acute obstruction of the pulmonary trunk. 4,7,8,10

Conclusion

The clinical manifestations of cardiac rhabdomyomas depend especially on the localization in the heart; they are varied and polymorphic, and often asymptomatic. Particular forms of localization might impair the vital prognosis.

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ССТ

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Wanted: Pediatric Cardiologist with Primary Interest in Echocardiography

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We are seeking a pediatric cardiologist whose primary interest is in echocardiography, ideally with a focus in fetal echocardiography and other diagnostic imaging. General responsibilities include clinical care, teaching and research. A fourth year fellowship in echocardiography is desirable but not necessary. The candidate should be board certified or board eligible in pediatric cardiology.

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SCAI View - A Monthly Column: SCAI 2011 to Feature Expanded Congenital Heart Disease Symposium, Live Cases, and Late-Breaking Clinical Trials

By, Daniel S. Levi, MD

I hope you all have had a joyous holiday season spent with family and friends. Here's wishing you a prosperous 2011!

Speaking of prosperity, I am excited to announce an expanded *Congenital Heart Disease (CHD) Symposium* at *SCAI 2011* Scientific Sessions with four days of programming, including an all-new Thrombosis Workshop. Taking place May 4-7, 2011 in Baltimore, Md. the *CHD Symposium* will still feature uninterrupted, focused programming on interventional therapies for congenital and structural heart disease, only more of it.

Also new this year is the addition of the "Late Great Pediatric Trials" session. Richard Ringel, MD, FSCAI, of Johns Hopkins Children's Center will headline this program, presenting updated data on the COAST Trial.

Another great headliner, John P. Cheatham, MD, FSCAI, of Nationwide Children's Hospital in Columbus, Ohio, will be delivering the Mullins Lecture. There is much we can learn from this congenital pioneer and his work in developing new techniques and devices as well as hybrid therapies.

Of course, the CHD Symposium will also include your tried-and-true favorites. We'll be bringing back the enormously popular "Brain Scratchers" session to challenge you to solve hemodynamic, angiographic or interventional mysteries and to provide solutions for less than routine cases in the congenital catheterization laboratory. The "I Blew It" sessions will be returning for the 12th year to educate, entertain and shock with all the ways interventional cases can go awry, and with the creative ways that our colleagues manage these complications. More importantly, the session addresses how to avoid such events in the future. If you have a case that might be a good learning tool for either session, please contact Frank Ing, MD, FSCAI, at ing@bcm.edu.

To view the latest schedule – including information on live cases – simply visit www.scai.org/SCAI2011.

CALL FOR LATE-BREAKING CLINICAL TRIALS

SCAI 2011 is inviting you to submit your Late-Breaking Clinical Trials (LBCT's) online at www.scai.org/SCAI2011. Here are three reasons why your trial results belong at Interventional Cardiology's premiere education meeting:

- Your Findings Will Be Heard Loud and 1. Clear: SCAI will promote your clinical trial at its on-site newsroom, in its robust media kit and to its extensive list of mainstream and trade journalists. In recent years LBCT's presented at SCAI's Scientific Sessions were prominently featured in The Wall Street Journal, USA Today, The Washington Post, The L.A. Times and more! With the full attention of interventional cardiologists from all over the country, many from around the world, you can share information they will take back and immediately apply in their daily clinical practice.
- Focus on Congenital and Structural Interventional Therapies: SCAI's attendees are exactly those you want your results to reach in a forum where they won't be distracted by headliners from other subspecialties or general cardiology.
- Easy to Navigate: SCAI 2011 is just the right size to foster collegial education, dialogue and collaboration. You'll find yourself discussing your findings with colleagues and how they should be applied in their practice long after your presentation.

ССТ

Daniel S. Levi, MD, FSCAI Congenital Heart Disease Program Co-Chair, SCAI 2011 Mattel Children's Hospital at UCLA Westwood, CA

FEBRUARY MEETING FOCUS

Cardiology 2011 15th Annual Update on Pediatric ad Congenital Cardiovascular Disease Feb. 2-6, 2011; Scottsdale, AZ USA www.chop.edu/cardiology2011

Program Overview: Develop a systematic approach for all CHD-based on a standard approach in three common defects: Transposition of the Great Arteries, Tetralogy of Fallot and Single Ventricle Lesions; develop surveillance strategies; understand various risk factors; recognize important elements in team approach to caring for patients with CHD.

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Drawing will be held in March 2011. The winner will be notified by email or phone, and will be announced in the April issue of **Congenital Cardiology Today**.

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Medical News, Products and Information

Miami Children's Hospital (MCH) Announce Start of a New ACGME Accredited Pediatric Cardiology Fellowship Program

Miami Children's Hospital (MCH) is proud to announce the start of our newly ACGME accredited Pediatric Cardiology Fellowship Program. We are currently accepting applications for both classes starting July 2011 and July 2012. The three-year fellowship will train two fellows per year by an outstanding faculty of 13 cardiologists / cardiac intensivists, 3 cardiac anesthesiologists, and 2 cardiac surgeons. The goal of the fellowship is to graduate well-rounded pediatric cardiologists able to provide the highest quality of care to patients from infancy through adulthood. The fellowship will be a three-year program divided into 24 months of clinical rotations and 12 months of research. In addition, fellows will enjoy a robust clinical experience rotating through our dedicated 18 bed cardiac intensive care unit, inpatient floor, ambulatory offices, electrophysiology lab, non-invasive imaging lab and a new state-of-the art hybrid catheterization lab. Noninvasive imaging includes TTE (2D and 3D), TEE, fetal echo and cardiac MRI. The cardiac catheterization program led by Dr. Evan Zahn, Chief of Cardiology, performs over 450 cardiac catheterizations a year with a strong emphasis on interventional procedures. The cardiac surgical program led by Dr. Redmond Burke, Chief of Cardiothoracic Surgery, performs over 250 open-heart cases a year with one of the lowest mortality results in the country (real-time results on www.pediatricheartsurgery.com). Surgical procedures can be seen on The Congenital Heart Surgery Video Project at www.youtube.com/user/Redmond111.

Risk Gene for Severe Heart Disease Discovered

Research led by Klaus Stark and Christian Hengstenberg of the University of Regensburg identified a common variant of the cardiovascular heat shock protein gene, HSPB7, which was found to increase risk for dilated cardiomyopathy by almost 50%. Their paper appears on October 28 in the open-access journal PLoS Genetics.

Per year, about 6 in 100,000 individuals develop dilated cardiomyopathy (DCM), with a higher prevalence in men. This disease is characterized by an enlarged, weakened heart, subsequently affecting the pumping capacity and often leading to chronic heart failure.

Those cases of DCM that occur in certain family groups are associated with a number of mutations affecting muscle cells. However, most cases are of unknown cause. To identify risk alleles for non-familial forms of DCM, an international collaboration of scientists analyzed the contribution of common gene variants to the more frequent, sporadic form of dilated cardiomyopathy, by conducting a large-scale genetic association study with more than 5,500 subjects. Different study groups from Germany and France contributed both well-characterized DCM patients and healthy controls. The HSPB7 gene was strongly associated with susceptibility to DCM.

The researchers concluded that, while genetic testing for this variant is not suitable to date, the findings are a first step towards supporting



eleventh annual international symposium on Congenital Heart Disease February 10–13, 2011

Register at www.allkids.org/conferences

Renaissance Vinoy Resort and Golf Club • St. Petersburg, FL future preventive measures for this severe form of heart muscle disease.

CITATION: Stark K, Esslinger UB, Reinhard W, Petrov G, Winkler T, et al. (2010) Genetic Association Study Identifies HSPB7 as a Risk Gene for Idiopathic Dilated Cardiomyopathy. PLoS Genet 6(10): e1001167. doi:10.1371/journal.pgen.1001167.

Quebec City researchers Pave the Way for Novel Treatment of Pulmonary Hypertension

Montreal - A Heart and Stroke Foundation researcher has discovered what could be the first truly effective breakthrough in the diagnosis and treatment of pulmonary hypertension, a devastating, life-threatening condition which results in an enlargement of the heart.

"We have discovered an early warning system in a protein called PIM-1," Dr. Sébastien Bonnet told the Canadian Cardiovascular Congress 2010, co-hosted by the Heart and Stroke Foundation and the Canadian Cardiovascular Society.

Dr. Bonnet has established that the PIM-1 cells can be used as markers of pulmonary hypertension.

"Blood samples were taken from patients to measure PIM-1 expression in the blood," says Dr. Bonnet, who is a professor at Laval University and a researcher at Centre hospitalier universitaire de Québec. "We were surprised to find that the greater the PIM-1 levels, the more severe the pulmonary hypertension in the patient."

He says this opens the doors to using regular blood tests to look at PIM-1 levels. "If there is a slight increase in PIM-1, we will know that something is going on." This is important since the condition is underdiagnosed and often not discovered until it is in a late stage. Without earlier treatment it has a very poor prognosis. The condition has traditionally been diagnosed by a six-minute walking test.

PIM-1 also offers the opportunity to move beyond the diagnosis of pulmonary hypertension to treatment. By blocking the PIM-1 protein, researchers were able to reverse the condition.

"This is a remarkable finding," says Dr. Bonnet. "We have found that using gene therapy to inhibit the inappropriate activation of this protein is a novel and effective therapy that can reverse the disease altogether."

Before this discovery there had been no agent to reverse the disease. Current drug treatments can improve quality of life but to this date there has been nothing that can cure the disease.

Pulmonary hypertension is abnormally high blood pressure in the pulmonary arteries, the arteries which carry blood from the heart to the lungs. The condition makes it more difficult for blood to flow to the lungs, causing shortness of breath, fatigue, and swelling of the feet and ankles. It can make everyday tasks almost impossible.

The number of Canadians with pulmonary hypertension is difficult to estimate, because it is under-diagnosed and the early symptoms are common to other conditions such as asthma and general fatigue. In addition, few studies have been conducted.



Division of Pediatric Cardiology Saint Louis University School of Medicine **Cardinal Glennon Children's Medical** Center

UNIVERSITY

Saint Louis University, a Catholic, Jesuit institution dedicated to student learning, research, health care, and service is seeking an additional pediatric cardiologist to join an established group within the Division of Cardiology and the Department of Pediatrics at Cardinal Glennon Children's Medical Center. Applicants will be considered at the Assistant/Associate Professor rank, and must be board certified/eligible in Pediatric Cardiology. General responsibilities will include clinical care, teaching, and research.

Interventional Cardiologist

We are seeking a second invasive cardiologist to assist our current faculty with the growing number of cardiac catheterizations and interventional cases. A complete spectrum of pediatric interventional procedures is currently being performed. An interest in clinical research is encouraged. Academic rank will be commensurate with qualifications and experience.

The cardiology division is in a period of significant expansion, with the opening of the Dorothy and Larry Dallas Heart Center within Cardinal Glennon Children's Medical Center in January, 2009. An active congenital heart surgery program exists, and the hospital houses state-of-the-art operating rooms and a new 60bed neonatal intensive care unit. Construction of a new hybrid cardiac catheterization lab/operating suite is scheduled to begin in 2010. The Doisy Research Center, a new 10-story tower housing the Health Sciences Center Research laboratories, was opened in 2007.

Interested candidates must submit a cover letter, application, and current CV to http://jobs.slu.edu. Other correspondence regarding this position can be sent to:

Kenneth O. Schowengerdt, MD, Wieck-Sullivan Professor and Director of Pediatric Cardiology, Saint Louis University School of Medicine, 1465 South Grand Blvd, St. Louis, MO 63104. Telephone: (314)-577-5633 Fax: (314)-268-4035 email schowko@slu.edu

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10- ----For more information contact: Elizabeth Peña at Children's Hospital Los Angeles. 4650 Sunset Blvd., MS 34, Los Angeles, CA 90027



The University of Maryland Hospital for Children is developing a comprehensive Children's Heart Program to meet the cardiovascular healthcare needs of the children of Maryland. We are currently recruiting for a Director of Interventional Cardiology to work in our new Hybrid Catheterization Suite. We are also recruiting for Directors of Non-Invasive Imaging and Electrophysiology/Pacing.

Sub-specialty board certification or equivalent work experience is required for each position. The ideal candidates will have proven leadership and program development experience. Clinical duties will focus primarily in the respective subspecialty field of each faculty position, although all members of the program participate to varying degrees in the general pediatric cardiology and outpatient practices. The Children's Heart Program supports integrated quality enhancement and clinical research practices to improve patient outcomes and patient/ family experience.

The successful candidates will have faculty appointments in the Department of Pediatrics of the University of Maryland School of Medicine at academic levels to be determined by experience. The University of Maryland Medical Center is a major academic tertiary care center serving Baltimore, the state of Maryland, and the mid-Atlantic region. As the oldest public medical school in the United States, the University of Maryland School of Medicine has an established tradition of outstanding clinical care, education, and research. The Department of Pediatrics is deeply committed to promoting children's health in the community and across the state, while supporting innovative clinical programs and expanding research initiatives.

Located on the modern and urban campus of the University of Maryland at Baltimore, The School of Medicine is one of seven professional schools within the University of Maryland system. The campus is ideally located within walking distance to the Baltimore Inner Harbor, National Aquarium, Baltimore Convention Center, Hippodrome Theatre, Orioles Park at Camden Yards and Baltimore Ravens M & T Bank Stadium. The University of Maryland Hospital for Children is also close to historic Annapolis, the Chesapeake Bay, Washington D.C., and many residential communities with outstanding public and private schools. The area offers rich cultural fabric and many unique recreational opportunities.

The University of Maryland is an EOE/AA/ADA and encourages minorities to apply.

Interested applicants should send CV to:

Dr. Geoffrey L. Rosenthal Director, Pediatric & Congenital Heart Program University of Maryland Hospital for Children 22 S. Greene Street, N5W68 Baltimore, MD 21201 grosenthal@peds.umaryland.edu

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