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Evaluating Newborns for Critical Congenital Heart Disease: Expanding the Availability of Diagnostic Echocardiography

By Amy L. Peterson, MD; Shardha Srinivasan MBBS; and John Hokanson, MD

The term Critical Congenital Heart Disease (CCHD) is frequently used, but not always well-defined. Many clinicians define CCHD as those heart defects that are likely to result in death or disability without treatment in infancy. Modern surgical, catheterization, and anesthetic techniques, widespread access to echocardiography, and availability of prostaglandins have revolutionized the care of infants born with CCHD. However, Congenital Heart Disease (CHD) is still the most life-threatening condition in the first month of life, and as a group represents 20% of neonatal deaths.[1] In an effort to improve outcomes, much interest has been generated in facilitating earlier diagnosis.

Two important considerations limit the ability to demonstrate the value of early diagnosis of CCHD. If only those infants who survive to reach a tertiary care center are included for analysis, the adverse consequences of missed diagnoses are greatly understated and the value of early diagnosis underestimated. The failure to differentiate a timely postnatal diagnosis of CCHD (prior to hospital discharge or onset of symptoms) from an untimely postnatal diagnosis (after hospital discharge or onset of symptoms) also limits the available literature's ability to demonstrate the value of early detection of CCHD. Despite these inherent limitations, the available literature still demonstrates the benefit of early diagnosis. In a study of 309 infants with congenital heart disease who survived to reach a tertiary care center, those with a postnatal diagnosis had higher rates of potentially unnecessary intubation and mechanical ventilation. Twenty-nine percent of postnatally diagnosed infants required more than two

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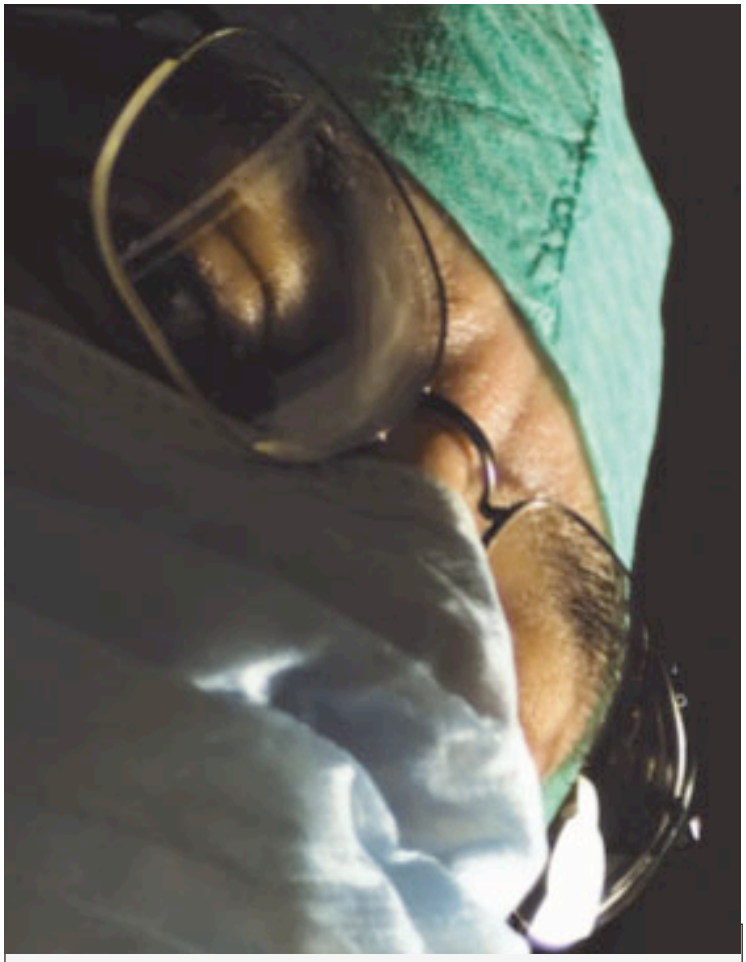

transports before they arrived at a facility equipped to treat their heart disease. Nineteen percent of infants had been discharged to home and were subsequently readmitted after presenting with symptoms.[2] Other studies have suggested similar improvements in outcome with earlier diagnosis.[3-5]

The detection of CCHD has evolved into a series of three mechanisms to detect these heart defects before the onset of symptoms and prior to hospital discharge: prenatal diagnosis, newborn physical exam, and pulse oximetry screening. Pulse oximetry screening has recently entered the field as the third safety net to detect CCHD that has not been recognized prenatally or identified by newborn physical examination. With the advent of pulse oximetry, a suspicion of CCHD prior to the onset of symptoms will be more frequently entertained prior to hospital discharge.

Prenatal Detection of Critical Congenital Heart Disease

What of prenatal diagnosis? Advances in routine obstetric ultrasound and the advent of fetal echocardiography have dramatically altered the approach to diagnosis of CCHD. However, even today, many infants with CCHD are not detected prenatally. Most infants born with CCHD are not from "high-risk" pregnancies that would trigger fetal echocardiography as a standard prenatal diagnostic test.[6] In order to prenatally detect the majority of CCHD, obstetric sonographers and physicians must be able to screen apparently healthy fetuses and identify possible congenital heart disease. This is heavily dependent on the skill level of the operator and the types of cardiac screening views obtained. A large congenital cardiac referral center found low prenatal detection rates for all forms of congenital heart disease amenable to prenatal diagnosis. The overall detection rate for all forms of CHD was 28%, with much higher rates at university-based practices (80%) than community-based practices (23%).[2] This rate does not differ substantially from rates in other series of reports.[7-10] Not surprisingly, detection rates varied enormously based on cardiac lesion, with higher prenatal detection rates among infants with heterotaxy, single ventricle physiology, or an atrioventricular canal defect (50-80%). Prenatal detection rates become disappointing with lesions such as Tetralogy of Fallot, mild Left Heart Obstructive Disease, or D-Transposition of the Great Arteries (19-31%).[2]

These low prenatal detection rates can be partly attributed to inadequate image acquisition technique or errors in interpretation, but this is not the entire explanation. Even with ideal images and interpretation, the expected detection rate for CHD in a low-risk population is only about 50% when a single 4-chamber view of the heart is used. The reason for this is obvious – many forms of CHD will in fact have a normal 4-chamber view. If this is the only view used to screen the heart, disease will be missed. The CHD prenatal detection rate climbs to 70-75% if a more comprehensive cardiac screen is used (4-chamber with outflow tracts and the "three vessel view").[9] Importantly, using the comprehensive cardiac screen will detect almost all forms of ductal-dependent CCHD. The importance of improving prenatal diagnosis rates cannot be over-emphasized; however, until the goal of universal prenatal detection is realized, additional tools to detect CCHD prior to the onset of symptoms are needed.

Postnatal Detection of Critical Congenital Heart Disease

Physical examination of the newborn is a vital tool in the detection of CCHD, but has significant limitations in its ability to detect CCHD prior to the onset of symptoms. Many infants with CCHD will not appear visibly cyanotic or have significant physical exam findings. Pulse oximetry screening for CCHD has recently emerged as an additional mechanism to detect CCHD prior to hospital discharge. When a postnatal diagnosis of CCHD is suspected by physical examination or pulse oximetry, echocardiography is the definitive diagnostic tool.

The technique to detect CCHD with pulse oximetry has been well-described.[11,12] New Jersey was the first state to legally mandate pulse oximetry screening and recently published their first 9 months of data. Of 75,324 births in 52 different facilities across the state, a total of 72,694 neonates were screened with pulse oximetry. Forty-nine failed (yielding a failure rate of 0.067%), and of those, 30 infants had no other signs or symptoms that would have prompted clinicians to evaluate them further. Of those 30, all of whom received a diagnostic evaluation, 3 had CCHD and 17 had another significant finding (sepsis, pneumonia, pulmonary hypertension, or a non-critical form of congenital heart disease). Of the 30 cases, only 6 infants required a transfer to a different facility that was

triggered solely by the failed pulse oximetry screen (and 5 of those infants had potentially significant echocardiogram findings).[13] Other studies have also demonstrated that when used appropriately, pulse oximetry screening has a very low false-positive rate.[11] For a state like New Jersey, with a relatively small geographical area, a high population density, and readily available on-site neonatal echocardiography, transportation of the small number of neonates a short distance to a facility with neonatal echocardiogram capabilities may be a good option.

Contrast those circumstances to those of Wisconsin, a state with a larger geographical area and lower population density. Ninety-nine hospitals in Wisconsin routinely deliver babies. Of 88 hospitals responding to a 2012 survey, 50 hospitals delivered less than one baby per day on average (Figure 1). A total of 55 Wisconsin hospitals, representing 25% of annual births, did not have same-day neonatal echocardiography available in house and would have to transport a baby to another institution to exclude CCHD, averaging 53 miles per transport.[14] Given the very low rate of pulse oximetry screening failures, even the highest-volume maternity hospitals might see very few screening failures per year. For the lowest-volume hospitals, there could be years between screening failures.

Expanding the Availability of Neonatal Echocardiography

How then does one provide the appropriate diagnostic tools for such a rare event? Under many circumstances, a baby could be transported to an institution with available neonatal echocardiography, often separating a new mother from her baby. In extremes of weather (a not-uncommon event in many less densely populated regions), transport of a potentially healthy baby for a diagnostic test may not be the safest option for all involved. If the echocardiogram findings did indicate CCHD, the infant may also require a second transport to an institution able to provide tertiary-level cardiac services. How will a neonatal echocardiogram be performed and interpreted under these circumstances?

Telemedicine is one potential solution. Webb et al reported infants with congenital heart disease born in an institution with access to telemedicine required less transports, had a shorter time to diagnosis, and had a shorter length of both hospital and ICU stay. In this study, echocardiography was performed by sonographers experienced in adult and pediatric echocardiography and interpreted by a pediatric cardiologist.[15] This intuitively makes sense. If a sonographer with substantial pediatric experience can obtain images and electronically send them to a pediatric cardiologist for interpretation, a diagnostic study can be performed without the cardiologist at the bedside. However in many centers that deliver babies, a sonographer with substantial pediatric experience is not available.

In most circumstances, the limiting factor in providing neonatal echocardiography is the availability of personnel who can obtain diagnostic images, not the technology required to transfer those images to a pediatric cardiologist for interpretation. Various solutions could be considered, including the utilization of personnel such as adult cardiac sonographers, general (non-cardiac) sonographers, physicians, or other individuals. However, given that a pulse oximetry screen failure in a smaller rural hospital will be such a rare event, a strategy of maintaining expertise in neonatal echocardiography among a group of people who do not routinely image the heart becomes impractical.

As a point of comparison, consider another application of cardiac ultrasound by non-sonographers. The use of Focused Cardiac Ultrasound (FCU) is endorsed by the American Society of Echocardiography (ASE). FCU is performed by ED and ICU physicians and is limited to a very specific evaluation of global myocardial function and pericardial effusion. To maintain proficiency in FCU, which is far less complex than a diagnostic neonatal echocardiogram, a three-part training approach is recommended, including didactic sessions, image acquisition, and image interpretation. The image acquisition portion of training generally takes the longest to achieve proficiency.[16] In their consensus statement, the ASE FCU committee recommended a formal training program for FCU because the interpretation of the study will have a "direct impact on patient care." The importance of regular performance of FCU to maintain competency is also emphasized. Maintaining proficiency in neonatal echocardiography among noncardiac sonographers responding to such rare events is not a

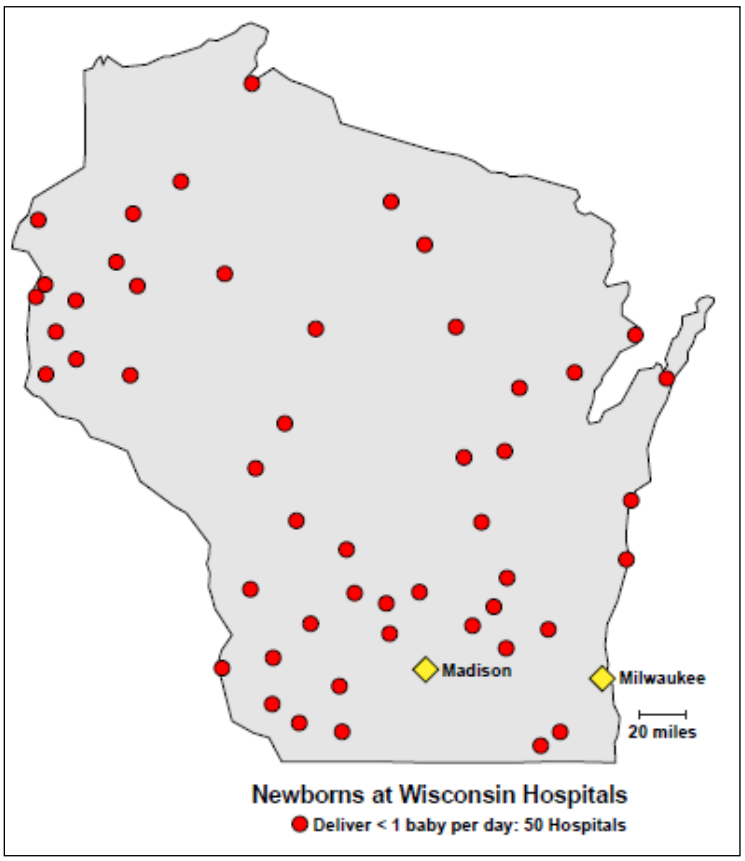

Figure 1. Geographic distribution of hospitals in Wisconsin. Circles show location of hospitals that deliver, on average, less than 350 babies annually. The diamonds identify Madison and Milwaukee, the two largest cities in Wisconsin. They are the only two locations in Wisconsin with pediatric cardiothoracic surgery and interventional cardiology services available.

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For a state like Wisconsin, if a sonographer experienced in adult echocardiography could be educated and coached to perform a neonatal echo on an infant who failed a pulse oximetry screen, the availability of in-house neonatal echocardiography expands considerably. A review of the ARDMS (American Registry for Diagnostic Medical Sonography) database shows that in Wisconsin in 2013, there are 415 registered sonographers RDMS-certified in adult echo and 58 sonographers certified in pediatric echo. Looking at their distribution across hospitals that deliver babies, 33/88 hospitals had the capability for same-day neonatal echocardiography already in place, with 75% of Wisconsin's babies born at one of these facilities. Forty-three additional hospitals had the capability for same-day adult echocardiography. If adult echocardiographers in those facilities could be trained to perform newborn studies, same day echocardiography would be available to 97% of Wisconsin newborns.[14] The availability of adult cardiac sonographers in all but the smallest hospitals is a resource that could be utilized to significantly expand the availability of same day neonatal echocardiography.

Echocardiographic Assessment of the Asymptomatic Newborn with a Failed Pulse Oximetry Screen

When a newborn fails pulse oximetry screening, once the possibilities of pulmonary disease or sepsis have been excluded, CCHD must be excluded by echocardiography. In this asymptomatic baby, the fundamental question to be answered is whether the baby needs immediate medical attention, needs outpatient cardiology follow-up, or can be sent home to receive normal newborn care. In a birth setting where definitive neonatal echocardiography performed by a pediatric trained cardiac sonographer is not available, the precise anatomic diagnosis may not be necessary as long as a distinction between normal and abnormal can be made. In a baby with no hemodynamic compromise, the risks of a missed diagnosis of CCHD must be balanced with the inconvenience, cost, and risk of transporting a newborn for echocardiography.

In our experience, a cardiac sonographer with adult training and the direct support of a remote pediatric cardiologist can perform echocardiography which answers the fundamental question of whether a baby has critical congenital heart disease. In nearly all situations, the pediatric cardiologist can be confident making one of three recommendations:

1) The echocardiogram demonstrates serious congenital heart disease or some other significant cardiac pathology necessitating immediate medical attention.
2) The echocardiogram is within normal limits and no further cardiovascular evaluation is necessary.
3) The echocardiogram demonstrates minor cardiac findings such as a small ventricular septal defect, a small patent ductus arteriosus, or a normally functioning bicuspid aortic valve that can be addressed with outpatient cardiology evaluation.

The need to transport an infant because the study could not confirm or exclude CCHD is quite rare.

Sonographer Education with the Wisconsin SHINE Project

This strategy, to educate adult sonographers in neonatal echocardiography technique, is one we have embraced at the Wisconsin SHINE (Screening Hearts In Newborns) project. A joint effort is being undertaken, combining the already-extensive cardiovascular imaging skill set of the adult sonographer with additional training and support from the pediatric cardiologist. To provide additional training, the Sonographer Education portion of the SHINE project is designed to assist sonographers who are not familiar with differences between congenital and adult echocardiography when faced with an asymptomatic neonate that has failed their pulse oximetry screen. This approach is not designed to be a comprehensive instruction in critical congenital heart disease, but to provide a basic framework that the adult cardiac sonographer can use to evaluate a newborn with remote support from a pediatric cardiologist. Sonographers are reminded that the goal of this echocardiogram is to assess for the presence of CCHD and determine whether or not advanced care is necessary for the neonate, not to make an exhaustive anatomic diagnosis.

The program is housed in the Wisconsin SHINE website (www.wisconsinshine.org) and will introduce sonographers to "The Terrible Ten" echo findings in critical congenital heart disease. These ten findings (Table 1) are ones that can be identified using the skill sets of an adult cardiac sonographer. The goal of this type of assessment is not to differentiate between the various forms of CCHD, but to determine if the infant requires immediate referral to a pediatric cardiac center for additional evaluation.

Additional defect-based segments are provided to foster better understanding of CCHD and echocardiography for CCHD. (Table 2).

Table 1:
"The Terrible Ten" Echo Findings that Could Indicate CCHD

- There is Retrograde Filling of the Ascending or Transverse Aorta
- The Aortic Arch Cannot Be Demonstrated in the SSN View
- There Is Exclusive Right-to-Left Shunting at Atrial or Ductal Level
- The Apical 4-Chamber View Is Abnormal
- An AV Valve Leaks... A Lot
- The Cardiac Apex Is Midline or Rightward
- A Normal Parasternal Long Axis View of the LV and Aorta Cannot Be Demonstrated
- A Normal Parasternal Short Axis View of the RV/PV/MPA Cannot Be Demonstrated
- The Aortic Valve and Pulmonary Valve Are Parallel
- Something Just Does Not Look Right

Table 2:
Types of Potentially Critical Congenital Heart Disease a Sonographer Might Encounter

- Cardiomyopathy
- Coarctation of the Aorta
- Critical Aortic Stenosis
- D-Transposition of the Great Arteries (including discussion of restrictive atrial septum)
- Ebstein's Anomaly
- Hypoplastic Left Heart Syndrome
- Interrupted Aortic Arch
- L-Transposition of the Great Arteries
- Single Ventricle Physiology
- Tetralogy of Fallot
- Total Anomalous Pulmonary Venous Return (including discussion of obstructed TAPVR)
- Tricuspid Atresia and Pulmonary Atresia with Intact Ventricular Septum

Sonographers are eligible for 2 SDMS CME credits free of charge upon completion of a pretest and posttest.

Conclusion

Under many circumstances, telemedicine may be an appropriate solution to providing diagnostic testing for an asymptomatic infant that has failed a pulse oximetry test. However, in the modern era of digital image acquisition and rapid electronic data transfers, image transfer technology is unlikely to be the limiting factor. The limiting factor is the skill of the sonographer performing the study and the ability of the physician to both remotely assist the sonographer and interpret the study. For routine, widespread use of pulse oximetry screening to be successful, both these human factors will need to be adequately addressed. Training adult sonographers to perform neonatal echocardiography under the guidance of a pediatric cardiologist appears to be the most viable solution.

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Supraventricular Tachyarrhythmia Secondary to Mahaim Syndrome in a 3-Year-Old Afrocaribbean

By Sandra Williams-Phillips, MB.BS, DCH, DM Paediatrics (UWI)

Abstract

Supraventricular Tachycardia (SVT) occurs in 40% of children with tachyarrhythmia. Mahaim Syndrome is a rare form of SVT and responds poorly to anti-arrhythmic medications leading to Arrhythmic Cardiomyopathy when uncontrolled. This index case is a 3-year-old boy presenting with sudden onset palpitations associated with cardiac decompensation responsive to vagal maneuvers. Electrophysiological studies and successful ablation confirmed the presence of a right-sided, mid-posteroseptal, concealed accessory pathway. The Index case had classical Electrocardiographic (ECG) patterns of LBBB and left axis deviation with wide QRS complex, captured during an episode of Mahaim Tachycardia. Minimal Pre-excitation was noted in L111 and V1 resting ECG. This is the 1st Case of documented Mahaim Syndrome in an Afro-Caribbean as far as the author is aware.

Keywords

Mahaim Syndrome, Supraventricular tachyarrhythmia, accessory fibres, ablation.

Introduction

Mahaim et al in 1938 described a rare concealed form of Supraventricular Tachycardia (SVT) which has no features of Pre-excitation on resting Electrocardiogram (ECG) and during tachycardia has a left bundle brand block morphology with left axis deviation and a delta wave following a normal PR interval.[1,2,3] It constitutes less than 3% of SVT's, and is usually resistant to anti-arrhythmic therapy.[3] The etiology as described by Mahaim et al has evolved over the past six decades. Initially described as being nodal in origin but later electrophysiological studies and ablations describe a right sided accessory pathway which in many case reports are adjacent to the Tricuspid annulus or adjacent septum. There has been confirmation that the position of Mahaim fibers can be in varied positions including originally described by Mahaim et al as nodal.[4-8] The accessory fibres are described as atriofascicular with decremental antegrade conduction properties, which connect the right atrium to right bundle branch or the HIS bundle or fasciculoventricular accessory pathway to the septum of right ventricle.[1-11] The ECG during tachycardia has a wide QRS complex with ventricular dysrrhythmia and LBBB pattern, with left axis deviation, which may make it difficult to differentiate from a primary Ventricular tachydysrhythmia.[7] Response to vagal manoeuvers makes the clinical differentiation easier being more likely to be a SVT.[3] Most of these patients usually have normal hearts as the index case does, but there have been documented cases with congenital heart disease such as Hypertrophic Cardiomyopathy[12] and other accessory pathways.[9] Resting ECG features of minimal Pre-excitation with a normal QRS complex duration, indicating the presence of an accessory pathway was noted in V1 and also in L111. Sternick et al (2004) found signs of minimal pre-excitation pattern in over 70% of patients with confirmed Mahaim fibres.[13]

Case Report

The index case is an Afro-Caribbean boy who had the first episode of palpitations occurring at 3 years and 11 months of age associated with chest pain and a heart rate between 200-250 beats per minute (bpm). The second episode of palpitations occurred four months later at 4 years and 3 months of age with Syncope and Cardiac decompensation, which was captured on Electrocardiogram (ECG) (Figure 1), followed by ten episodes occurring intermittently over the next 4 months, then weekly thereafter.. The palpitations occurred at rest, worsened by exertion and caffeine. One episode was terminated by one-sided carotid sinus massage. Duration of palpitations varied from 1 hour to 18 hours. He would function at NYHA 1V during episodes when he would normally function at NYHA 1. Despite compliance there was a poor response to Atenolol and total control with Flecainide at increasing dosage to 5.4 mg per kilogram per day in three divided dosages. There was associated dizziness, fainting and Syncope with palpitations. He had no Congenital Heart Disease, Cardiac Surgery, or history of deafness. He has a known case of Moderate Bronchial Asthma, is on prophylactic Singulair, and was advised about the possibility of exacerbation whilst on Atenolol, which did not occur.

There was no family history of arrhythmia, deafness, sudden death less than 50 years of age, use of Pacemaker, muscular dystrophy, Marfan's Syndrome, hypertrophic Cardiomyopathy or Arrhythmogenic Right Ventricular Disease.

On examination he was a 5-year-old obese young boy with no dysmorphic features. Arm spam height ratio was normal with negative thumb and wrist sign; the upper body and lower body segment ratio was normal.

- Weight: 42.3kg
- Height: 122cm.
- Saturation in air: 100%
- BP: 103/70
- Resting Pulse: 72/min
- Respiratory rate: 20/min

His pulse rate was 72 per minute with normal rhythm and volume, There was no collapsing pulse or pulse deficit. There was no palpable pulmonary component of the second heart sound, Apex beat was normal in the 5th left intercostal space mid clavicular line. Heart sounds 1 and 2 were normal with normal variable split of second heart sound and normal pulmonary component of the second heart sound. There were no murmurs.

There were no signs of Congestive Cardiac Failure or Pulmonary Hypertension.

Abdominal examination was normal with no hepatomegaly.

Respiratory system was normal, and there were no signs of Muscular Dystrophy, or Scoliosis with normal central nervous system examination.

Exercise was restricted with no competitive sports and sustained exertion advised.

Blood investigations revealed normal thyroid function tests, normal cardiac enzymes, normal urea and electrolytes, normal Calcium, Phosphorus and Magnesium levels.

One resting Electrocardiogram (ECG) before the captured tacharrhythmia was normal (Figure 1, first column). There were no overt signs of pre-Excitation Syndrome, minimal pre-excitation, Ion Channelopathy, Brugada Syndrome or Epsilon wave.

The 2nd resting ECG taken the day after the 2nd captured episode had normal P-R interval with minimal "rS" pre-excitation noted in L111 and a small pre-excitation "delta" wave on upstroke of R wave in V1, suggestive of Mahaim Syndrome (Figure 1, second column).

The ECG which captured tachyarrhythmia during the second episode at 4 years and 3 months showed Ventricular Tachycardia with wide QRS complexes and a LBBB morphology (Figure 1). All classical features of Mahaim Syndrome (Figure 1, third and fourth column). The ECG in the fourth column was taken stat before the ECG in the third column.

Chest X-Ray was normal with Normal Cardio-thoracic ratio, lung fields, normal right and left bronchi ratio and normal position of liver, spleen and stomach bubble ruling out isomerism.

Echocardiogram showed a Structurally Normal Heart with no signs specifically of Ebsteins Anomaly, Arrhythmogenic Right Ventricle, Arrhythmogenic Left Ventricle, Atrial Septal Aneurysn, Corrected Transposition, Atrial Septal Defects, Sinus Venosus Defects, Mitral Valve

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- Intended Intervention - COA Stent Using Atrium Advanta V12 Covered Stent - *Live Case*
- Intended Intervention - ASD Closure - *Live Case*
- Intended Intervention -Transcatheter VSD Device Closure - *Live Case* Intended Intervention - COA Stenting Using Premounted Advanta V12 Covered Sten - *Live Case*
- Stunning Revelation - The Medical System is Changing - What Can You Do To Show Patients That Your Practice Does It Right? Patient Perspective
- Percutaneous Paravalvular Leak Closure Outcomes
- Intensive Management of Critically Ill Infants Undergoing Catheterization
- Off Label Device Usage - Careful!
- Evolving Hybrid Programs Communications is the Key
- Anti-Coagulation in Pediatric Interventions - Evidence Please! Are we Flying Blind?
- Shaping Your Structural Heart Interventional Career: How Do I Get the Necessary Training and Start a Program?
- Closure of Post-Infarct VSDs Limitations & Solutions
- Percutaneous Interventions During Pregnancy
- Percutaneous PVL Closure Techniques & Outcomes
- Percutaneous Therapy for MR
- Update on Percutaneous Aortic Valve Replacement: Time to Open the Flood Gates?
- 2011 PICS/AICS Achievement Achievement Award Presented To Horst Sievert, MD.
- The Cases I Learn The Most From
- If I was Starting Now I Would... Contemplating the Question.
- 10 Tips for the Trainee
- The Perfect CV
- Why I Left The Cath Lab
- Intended Intervention Occlude Right High Flow Superior Segment PAVM
- Intended Intervention Edwards Sapien XT Transcatheter Heart Valve - 3D Guidance - *Live Case*
- Closure of the Silent PDA - Indicated or Not
- Transcatheter Tricuspod Valve Replacement
- Recommendations For Device Closure of MVSDs
- Intended Intervention Diagnostic Angio/First Intervention Occlude Left Lower Lobe - *Live Case*
- Intended Intervention PFO Closure using Gore Helex Septal Occluder Under GA and TEE Guidance - *Live Case*
- Pulmonary Valvuloplasty - Pulmonary Oedema Presentation
- Intended Intervention LAA Closure Using the Amplatzer Cardiac Plug - *Live Case*
- Transcatheter Alternatives in Sick Neonates With Coartation of the Aorta; Balloon Angioplasty vs Surgery for Native Coartation of the Aorta in Newborns
- **and many more…. Plus, live cases from other major meetings**

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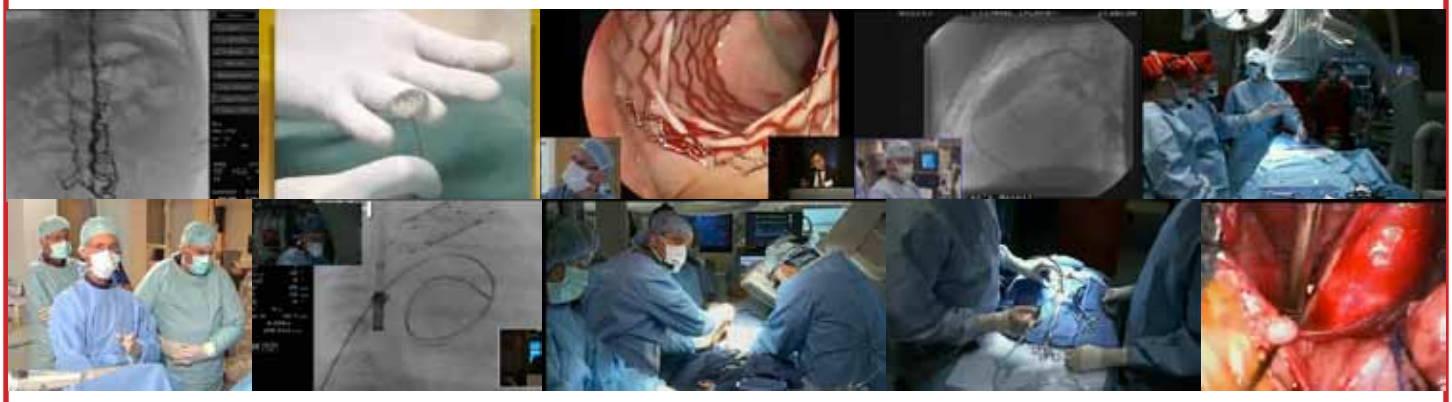

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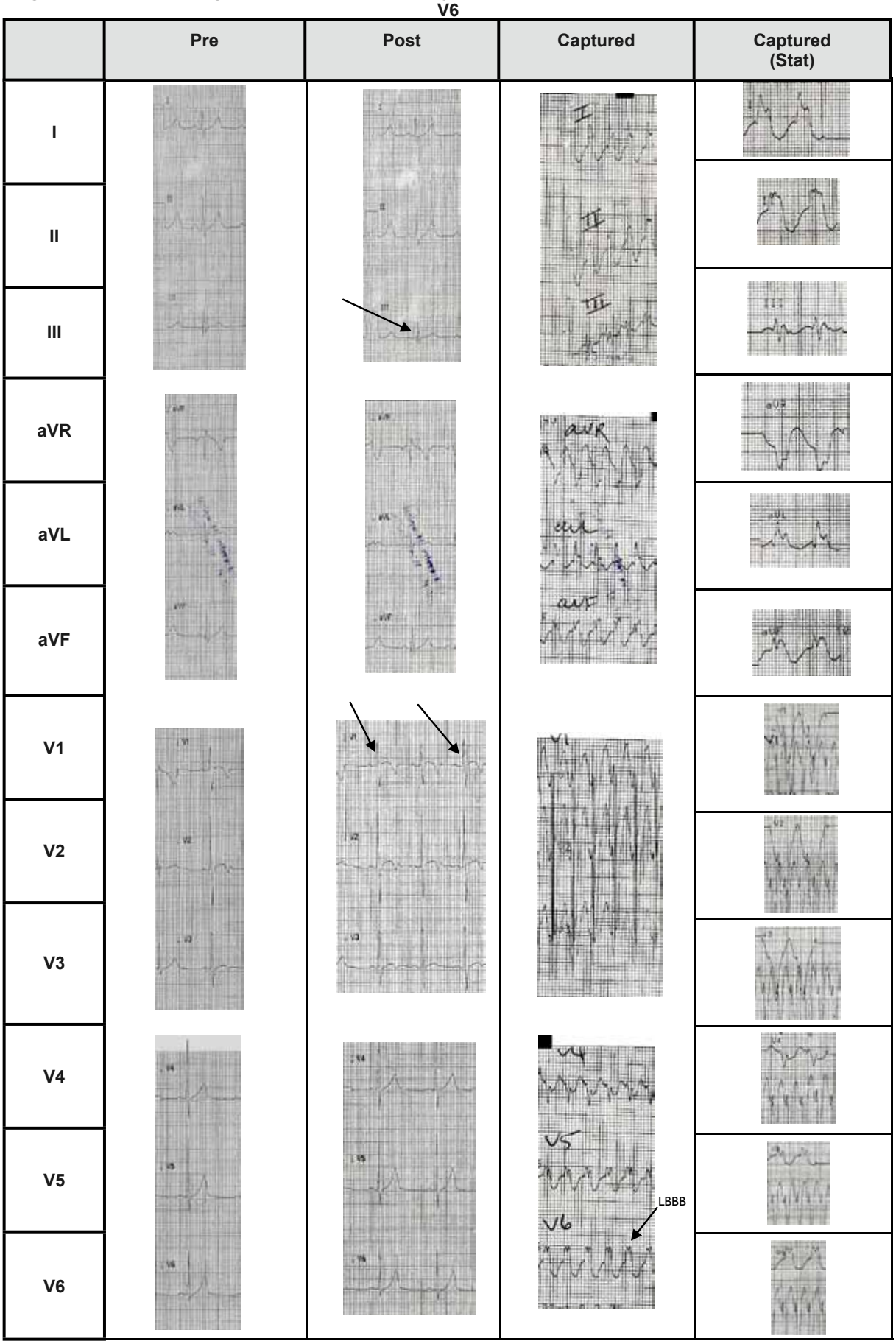
Figure 1. ECG's Showing Pre, Post, Captured, Tachycardia, Minimal Pre-excitation, Lead III, V1. LBBB in V6



Prolapse, Isomerism, Hypertrophic Cardiomyopathy or Pulmonary Hypertension.

Holter assessment was normal showing sinus arrhythmia, as no events occurred during the 24 hours.

Transoesophageal Pacing showed "abnormal pathway in right side of heart."

An uncomplicated Electrophysiological Study and Ablation overseas found and successfully ablated right sided mid-posteroseptal concealed accessory pathway.

In summary, we have a 6-year-old boy at Ablation with Supraventricular Tachyarrhythmia secondary to Mahaim Syndrome aborted by Vagal manoeuvers taught. He was controlled on Flecainide and cured by EP Study and ablation. He had Classical ECG features of Mahaim Syndrome when the 2nd episode of tachyarrhythmia was captured on ECG and signs of minimal pre-excitation on resting ECG following captured tachydysrhythmia.

Discussion

Sternick et al described minimal pre-excitation in Mahaim patients that were statistically significant in 60% of his group, of confirmed by Electrophysiological studies Mahaim fibre patients as "rS" pattern in L111, as noted in Index case.[13] The index case also had minimal pre-excitation in V1. The presence of pre-excitation was noted by other authors as low as 0% by Bardy et al to 50%. These minimal pre-excitations occurred when the duration of the QRS complex was of normal duration, less than 0.12 seconds.[13]

The Index case had Cardiac decompensation on two separate occasions as the Ventricular tachycardia lead to an inability to maintain cardiac output when he had the tachyarrhythmic episodes. Capturing the event via surface ECG as in this case or via Transoesophageal pacing and EP Study is important in helping to differentiate the type of SVT tachyarrhythmia. This provides a guide to suitable anti-arrhythmic therapy and future optimal management, as the technique of Ablation is dependent on pathway identified.[3,4,14-17] The response of his dysrhythmia's to vagal manoeuvers provided a clinical clue to a Supraventricular origin. A normal resting Electrocardiogram (ECG) and Echocardiogram and Holter assessment as in the Index case does not rule out this diagnosis. The anti-arrhythmic resistant nature of Mahaim type SVT, if not controlled quickly, can lead to an Arrhythmogenic Cardiomyopathy, which can be reversible, if dysrhythmia is treated early.[1,3,4] A Familial type of Mahaim Syndrome has been described in two brothers.[18] There was no family history of Sudden Infant Death Syndrome, Sudden Death under 50 years, dysrhythmia, or other Hereditary disorders associated suggesting a spontaneous mutation in this Index family.

The index case is the first documented case of a 3-year-old presentation of an Afro-Caribbean boy showing the classical ECG features of Mahaim Syndrome so defined over 6 decades ago by Mahaim et al in 1938 and significant minimal pre-excitation of "rS" in L111, as described by Sternick et al in 2004. Mahaim fiber tachyarrhythmia was cured over two years prior to documentation, by an uncomplicated Electrophysiological Study and Ablation.

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Recap of The 6th World Congress of Paediatric Cardiology and Cardiac Surgery, Cape Town 2013

By Christopher Hugo-Hamman, MD

The *6th World Congress of Pediatric Cardiology and Cardiac Surgery* (*WCPCCS*) was held in Cape Town, South Africa in February 2013. It was a seminal event, and the first time that a world congress in Cardiology or Heart Surgery has been held in Africa. We are pleased to report that the global economic recession, notwithstanding, and despite the long haul destination for North America and most of Asia, there were 3033 attendees in Cape Town. This is 700 more than in Cairns 4 years ago, and equal in number to Buenos Aires in 2005 (3000). There were delegates from 104 countries with over 100 more attendees from North America (571) than 4 years ago. Whilst we had hoped to grow the *World Congress* significantly, and had a figure of 4000 attendees in mind, the evidence suggests there is still life in international heart meetings in general and in the *WCPCCS* in particular.

We believe the *WCPCCS* is the pre-eminent meeting for professionals devoted to heart disease in children, and given its genuinely global footprint, an event which deserves respect and support from institutions and organisations across the globe. In a magnanimous gesture, the Children's Hospital of Philadelphia and the All Children's Hospital in St Petersburg cancelled "Heart Week" and their meetings for February 2013. "Cardiology in the Young" in London, did likewise. *The World Society for Pediatric and Congenital Heart Surgery* (*WSPCHS*) generously moved their *4th Congress and Scientific Meeting* from 2013 to 2014 to respect the *WCPCCS*. A few organisations, including the Association for European Paediatric and Congenital Cardiology (AEPC) and the Pan African Society of Cardiology went ahead with their own meetings and *PICS-AICS* moved its meeting to January 2013. Notwithstanding, we are pleased that with only one exception, all invited international organisations participated enthusiastically in the *World Congress*. The two existing international societies (*WSPCHS* and the International Society for Nomenclature in Pediatric and Congenital Heart Disease) had sessions in the programme. For the first time the world Heart Federation (WHF) participated in the *WCPCCS* with several key-note speakers and an exhibition booth and even co-hosted one of the most important closing seminars on, "Setting Millennium Development Goals for children with Heart Disease."

There were 29 industry symposia which were very well supported; in fact, many of them were oversubscribed. The Pediatric Cardiac Intensive Care Society, the AEPC, the Japanese Society of Pediatric Cardiology and Cardiac Surgery and the Pan Arab Congenital Heart Disease Association had symposia with their own subject choice. Eight North American children's hospitals (Philadelphia, Boston, Wisconsin, Texas, Toronto, St. Petersburg, Stollery, Cincinnati) and Mayo Clinic Rochester held their own symposia at the *WCPCCS* in Cape Town.

The scientific programme balanced the highest levels of achievement in research and technological innovation for complex congenital lesions, with health systems questions relevant to the distribution of cardiac care to needy children. Regenerative medicine, growing heart valves and stem cell therapies featured prominently, but so did the less frequently asked questions on how our patients are doing after they survive complicated heart surgeries. The surgical track plumbed the depths of left ventricular outflow tract obstruction with all manner of associated anomalies. This focus on a single lesion, as was the case with atrio-ventricular septal defect in Cairns, proved popular. On the other hand, and in a departure from previous congresses, one of the six tracks was devoted to "health systems and heart disease." For those in wealthier nations the cutting edge systems questions relate to organisation of services, rationing and rationalizing care and to quality assessment. However, in the less well-resourced countries few children with heart disease get the treatment they require and the *WCPCCS* strongly upheld the view that these children ought not to be denied the benefits of medical science. We fulfilled our vision - to highlight the challenges for paediatric cardiac care in less well-resourced countries and to engage government and civil society in finding solutions to the many systems deficiencies that lead to the neglect of children with heart disease. International and national humanitarian programmes were widely represented, participating in several sessions devoted to different models of service provision and system development. These included: The *Global Heart Network Foundation, Caring and Living as Neighbors CLAN, NCD Child, Medtronic Foundation, Chaine de L'Espoir, Save a Child's Heart, Global Heart Network, Children's HeartLink, World Heart Federation, Congenital Heart Disease Public Health Consortium* and the *International Quality Improvement Collaborative.*

WCPCCS was held in Africa for the first time; consequently, acquired heart disease in general and rheumatic heart disease (RHD) in particular were given an appropriately powerful platform. RHD ran in one of 11 parallel venues for the entire week, and was featured in the surgical, cardiology, imaging and health systems tracks. These sessions were very well-supported, recognising that over 35 million people are affected by RHD worldwide, and in most of the world, children with RHD represent half of the burden of heart disease. In another progressive development (50% of heart surgery in many developing countries is for RHD), the "Surgery for RHD" sessions were held jointly with the World Society for Pediatric and Congenital Heart Surgery.

Supported by a team from our partner "Congenital Structural Interventions (CSI) Frankfurt," the intervention programme ran live cases on three of the five days. On Day 1, Monday 18 February, transmission to the Cape Town International Convention Centre was by satellite from two venues in Europe (Frankfurt and Milano). Live transmission from 4 hospitals within South Africa was then streamed successfully via broadband internet later in the week. There had been scepticism about internet transmission, but it proved as reliable as satellite transmission with good quality images at a much reduced cost. We expect this experience will benefit other meetings running live case interventions. Indeed the live case programme brought forth many "firsts' in Africa, including trans-catheter pulmonary and tricuspid valve implantations, and was a wonderful showcase for what can be done on our continent. The intervention track showed lectures and debates in between live cases, and was not an appendage to, but a full and vital track within the *World Congress* programme. It was open to all attendees without an additional registration fee.

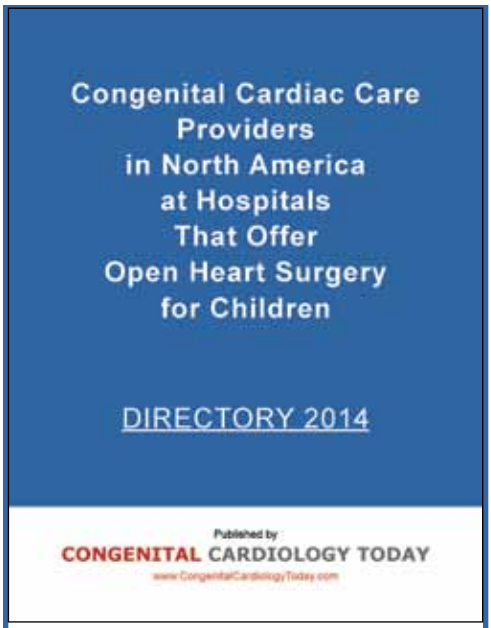
The nursing science track was well-attended with enthusiastic participation from a genuinely global audience. The ACHD track amongst other subjects, explored the challenges of care for Fontan and Tetralogy of Fallot patients and also women in their reproductive years, although the numbers of attendees at this track fell short of our expectations. This was possibly a consequence of competing and compelling attractions across the other five tracks; however, energizing the adult cardiac practitioners to the challenge and excitement of caring for adults with congenital heart disease remains difficult. This is a serious problem for healthcare planning and organisation, as there are now more patients older than 18 years with congenital heart disease than there are children with CHD in Europe, North America and other industrialized nations. Speaking to this burden of disease, were parent and patient support and advocacy groups like the Adult Heart Disease Association, and Congenital Heart Disease Consortium who participated in sessions on patient centered care and advocacy. To broaden interest and feature problems of "healthy" adolescents and young adults, "sudden death" amongst high performance athletes was highlighted in one of the 20 plenaries, and there was a special summit on sudden death featuring: Sudden Arrhythmia Deaths Syndrome (SADS) Foundation, PACE AFRICA and the Arrhythmia Alliance. Notwithstanding the popular debates, the highlights of the programme were without question the 20 plenaries and in particular, the lectures that honoured Jane Somerville (Innovation and fragility), Sir Donald Ross (Evolution of the pulmonary autograft aortic valve replacement) and the late Christiaan Barnard (Mechanical alternatives to transplantation).

The carbon footprint of an international conference of 3000 attendees is appalling and it is prohibitively expensive to offset the environmental impact of intercontinental air travel with "carbon credits." Through the use of recycled materials (for the delegate bags), the avoidance of bottled water and keeping printed material to a minimum, we were able to make useful contribution to partially offsetting the damage. Happily, technology assisted us, and we proved that the days of research scientists carrying paper and plastic posters across the world are now over. In the call for papers, 1405 abstracts were submitted; 1273 were accepted and 977 were finally shown at the Congress (123 orally by invitation and 854 as E-posters). Abstracts were submitted digitally and continuing this theme, "E-posters" were shown on-line and in the very well-attended "Abstract café." The format proved popular, and we feel our decision to go with digital E posters was vindicated.

It is not a simple matter designing a World Congress programme satisfying the curiosity of computational methodologies in CHD, integrating multimodal imaging, incorporating rotational angiography in the catheter laboratory and yet ensuring the more common problems confronting the office cardiologist are adequately covered. There is no formula or magic recipe, and many months of discussions, meetings, consultations and invitations were required. We feel the 6 tracks did justice to the diverse interests held within the *WCPCCS,* and under the leadership of Scientific Chairmen, John Lawrenson and John Hewitson, the International Scientific Committee did a commendable job. The "culture" of paediatric cardiac care is one in which we "work together" and this is possibly true for cardiac care more than for any other area of tertiary health care. The cohort of people from junior to senior, across specialities, across nursing-physician "barriers" and together with the NGOs, are marked by this common feature. The WCPCCS programme spoke for and to this reality. The presentations and E-posters can all be viewed at www.pccs2013.co.za.

Readers may find the challenges of financing large international meetings like *WCPCCS* of interest. We managed to attract a large industry exhibition with 74 companies exhibiting at 124 booths or stands. Most of these were South African subsidiaries of multinationals. Support for paediatric cardiology from the global pharmaceutical industry remains underwhelming and there were some glaring absences from medical device companies (despite the extraordinary 5 day structural heart disease intervention programme). One third of the budget was generated by educational grants secured years before the congress from South African hospital groups Netcare and MEDICLINIC, the Department of Health, Tropicana and the National Lottery of South Africa. This backing enabled a successful sixth *WCPCCS*, and we are profoundly grateful to these institutions for their support. Despite the parlous state of the global economy, delegate fees were below that charged at the *5th WCPCCS*. Considering this is a 5-day meeting which included live cases, the fees were competitive with similar international cardiac congresses. Although not financially viable, the very cheap "super early bird" registration brought in over 300 paying delegates 10 months before the Congress, and those delegates served as a registration vanguard for other people from their institutions. Offering reduced rates for attendees from the less well-resourced countries was an imperative. We followed the precedent set at the *5th WCPCCS* with the World Bank "low income country" being the criteria. We believe it would be more appropriate to have low and low middle income countries, and suggest this for the *7th World Congress.* Surgeons and cardiologists from most of the world do not have the luxury of a world class national meeting in their own country or own region every year, if ever. For them to learn from others and make their contributions to science, they need to travel. And of course most of them hail from countries characterised by low incomes and poorly valued currencies making international travel unaffordable. With assistance from PepsiCo, thirty-five *WCPCCS* scholarships and travel grants were offered to scientists from Africa and low income countries.

It was important for us that a project of this magnitude leaves a tangible legacy in South Africa. The Ministry of Health in our country, involved in many ways in the *WCPCCS*, has at last taken note of the fact that the majority of children with heart disease in South Africa never get the necessary care or surgery they require and it has joined a dialogue towards finding a solution. A web-based multi-lingual patient centered children's heart disease information system has already been launched thanks to the *WCPCCS* and will be an enduring footprint. A free provider portal for registered clinicians from Africa is the second component of this initiative. However, the most important achievements of a World Congress must surely flow from the meeting of minds with sharing of new ideas and information and this will be the intangible legacy. The outcomes develop slowly, out of reach, beyond awareness, often without any measure or acknowledgment. The *6th World Congress* held in Cape Town in February was the most important event in cardiac care to have ever been held in Africa. The numbers of delegates was three times greater than any other heart congress ever to have taken place on our continent and 1104 persons, a third of the attendees, were from Africa. The *WCPCCS* in Africa created opportunities for many surgeons and cardiologists who have never had the benefit of attending or presenting their data in a global scientific forum. Together with our faculty of 366 (from 36 countries), we achieved something significant: an exciting, diverse forum in which to share ideas and provoke thinking, renew old and make new friendships, learn new things and explore innovative ideas. All attendees had one



THE UPDATED 2014 DIRECTORY IS AVAILABLE ON CONGENITAL CARDIOLOGY TODAY'S WEBSITE

www.congenitalcardiologytoday.com /index_files/CCT-DIR-2014.pdf.

This free Congenital Heart Disease (CHD) resource can be viewed, downloaded/saved, and printed.

This professional resource will provide the Congenital Heart Disease community detailed information to better serve their patients.

It is an invaluable resource tool with tremendous benefit for you, your staff and for your patients. The information is arranged by State/Province and City, giving the Dept head, the Fellowship Director the pediatric cardiologists and congenital heart surgeons by hospital

"The success of a World Congress is measured in many different ways. It is not just about numbers, but about the simple and old values of friendship across boundaries of profession or borders of nation, fellowship, collegiality, getting together, cooperation and collaboration."

common goal: the health of children with heart disease. To this end, it was a satisfyingly successful week.

Over the years we have asked, why is there no global voice for children with paediatric and congenital heart disease? This was a recurrent theme across the health systems and heart disease track. Some have called for a "World Society for Paediatric and Congenital Heart Disease" and the *WSPCHS* commendably suggested the creation of a "Global Organisation." Consider that in 2011 the United Nations held a high level summit on non-communicable diseases (NCD's). There followed a September 2011 "Political Declaration of the High-level Meeting of the General Assembly on the Prevention and Control of Non-communicable Diseases." Although heart disease was recognised as one of the four NCD's, children's heart disease did not receive a mention. The same is true of the programmes of the other UN institutions that are active in child health. Whereas the World Heart Federation has lobbied intensely together with other advocacy groups for adult heart disease, it is our collective responsibility to admit we have failed to ensure an effective international coalition speaking out for children with heart disease. It is time to recognise and accept a responsibility to spread, develop and improve care for children and adults with congenital and acquired heart diseases. The Steering Committee of the "*World Congress of Paediatric Cardiology and Cardiac Surgery*" has now recognised it ought now to be at the forefront of developing an institution which is active and effective in global health and international public policy. Specifically, its goal should be to raise awareness about paediatric and congenital heart disease, with a mandate for international advocacy to improve availability of cardiac services for children and to drive campaigns with more specific disease based preventative and therapeutic objectives. This is our vision. Furthermore, we believe this should be a democratic and inclusive institution with representation from the humanitarian organisations and advocacy groups active in the provision of cardiac services as well as the paediatric and congenital heart disease societies active across the world.

The success of a *World Congress* is measured in many different ways. It is not just about numbers, but about the simple and old values of friendship across boundaries of profession or borders of nation, fellowship, collegiality, getting together, cooperation and collaboration. Personal contact will always remain an endearing and irreplaceable quality provided by these meetings. Hearing and meeting great people who have made new discoveries, invented wonderful things, or who have contributed to children's heart health in the most difficult of circumstances with meagre resources inspires us all. Having faith that this is what will ensure the survival of the *WCPCCS,* and based on our experiences in Cape Town, we expect that Istanbul, which is the destination for the *7th World Congress (http://wcpccs2017.org/en/)* in 2017, will be the best ever, and for those for whom numbers are important, reach the magical figure of 4000.

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Novel ACC Program Advises Hospitals on Building a Centralized Clinical Database

By Tim Feltes, MD, FACC

Over the last decade, patient data collection has become increasingly important to provide research and feedback reports to physicians which will ultimately lead to improved quality. However, several hospitals and practices are finding themselves in a situation where they maintain multiple databases that individually provide reports, and are looking for a simple, streamlined solution.

Creating a functional clinical database that lends itself to meaningful clinical research remains the Holy Grail in our field of medicine. There are often so many individual databases that need to interact on one platform.

In fact, Nationwide Children's participates in the NCDR's ICD Registry™, the IMPACT Registry®, an electronic health record system, and has several databases created by individual researchers or teams to provide feedback, capture data relevant to particular programs, or meet specific needs.

A work group from Nationwide Children's Hospital met with ACC staff during in person consulting sessions.
Top row: left-to-right: Anthony Orsini; Timothy Feltes, MD, FACC; Jamie Phillips, MHA, FACHE; Brendan Mullen; Soojin Lee; Laura Ritzenthaler, PA, MBA; and Nathan Glusenkamp, MA
Bottom row: left-to-right: Daniel Gomez; Maribeth Quinn; Naomi Kertesz, MD; Jeff Schmidt

My colleague, Jamie Phillips, MHA, FACHE, Vice President of Operations at The Heart Center, and I attended the ACC's *Cardiovascular Summit* in Las Vegas in January 2013. Here we both learned more about the ACC's NCDR PINNACLE Registry® and how it takes disparate data and provides one report back to clinicians. Jamie and I saw an opportunity to achieve our goal of developing one, uniformed database.

After gaining full support of hospital senior leadership, following the Cardiovascular Summit, Jamie reached out to the ACC's PINNACLE team to discuss consulting opportunities, and a consulting engagement agreement to help Nationwide Children's Hospital determine their reporting goals, re-focus their data elements, and create a centralized repository to increase the usefulness of the data for the researchers and physicians, was finalized.

Soon after, four members of ACC's PINNACLE staff flew to Columbus, OH, to meet with a Nationwide Children's Hospital workgroup comprised of physician leaders in various cardiovascular areas of expertise, including adult congenital heart disease, electrophysiology, catheterization/intervention, non-invasive imaging, inpatient and outpatient; The Heart Center's administration staff; nursing leadership; cardiothoracic surgery physicians; and the internal data warehouse team.

The ACC's PINNACLE team let us design what we wanted, but offered helpful insight as to the practicality and feasibility of implementation. This was useful for our whole Nationwide Children's Hospital team to experience and taught us to ask these questions throughout the process of developing our database

> ***"Creating a functional clinical database that lends itself to meaningful clinical research remains the Holy Grail in our field of medicine. There are often so many individual databases that need to interact on one platform."***

Between each meeting, the Nationwide Children's Hospital team was given assignments to help make the next meetings as productive as possible. Fortunately, we had an exceedingly motivated team both from the ACC and our internal team that made the partnership incredibly productive with our limited time together.

Throughout the course of the consulting engagement, the PINNACLE team helped us define exactly what we were looking to achieve. The exercise helped us focus on developing a manageable database and helped us define exactly how that database would be governed and managed in order to produce a reliable, high quality product that would be useful to our clinical investigators and ultimately positively influence our patient outcomes. PINNACLE staff also listened to what we wanted to develop rather than tell us how it's going to be. It was a great partnership

Congenital Heart Disease Clinical Practice Discussions Continue at ACC's 2014 CV Summit

By Robert Campbell, MD, FACC, and Maryanne Kessel, RN, MBA

The American College of Cardiology's (ACC) Adult Congenital and Pediatric Cardiology (ACPC) Section will again host a dedicated congenital heart disease workshop in conjunction with the American College of Cardiology's CV Summit. 2014 CV Summit: Solutions for Thriving in a Time of Change, January 16-18, 2014 in Las Vegas. As in years past, pediatric cardiologists and CHD professionals and administrators are invited to join the ACPC Section's Clinical Practice WG. The ACPC Clinical Practice WG is committed to encouraging discussions and aligning clinical practice goals between pediatric and adult congenital cardiology physician leadership and CHD administrators. As such the CHD discussions at the CV Summit will be focused on financial issues, data structure, governance models and service line structure.

Additionally, Timothy Feltes, MD, FACC, Jamie Phillips and the ACC PINNACLE Team will present the Centralized Clinical Database at the CHD workshop.

Information about registration and the overall CV Summit program is available at: www.CardioSource.org/CVSummitE3 or email StephanieMitchell at: smitchel@acc.org for more details.

Robert Campbell, MD, FACC and Maryanne Kessel, RN, MBA are Co-chairs of the American College of Cardiology's Adult Congenital and Pediatric Cardiology Section's Clinical Practice Work Group.

between Nationwide Children's and the ACC NCDR PINNACLE team.

As of July 2013, Nationwide Children's Hospital has completed their in-person meetings, and is now providing comprehensive reports in a centralized database that will be self-sufficient moving forward.

The energy that this exercise created for us not only benefited our development of our clinical research database, but will carry over in other team-developed initiatives. Consider the fact that an adult cardiology registry could be adapted to a congenital heart disease registry. That's quite a leap! I believe that the future of improved care for our congenital heart disease patients throughout childhood and into adulthood depends upon meaningful, accurate, and reliable data. No single congenital heart program is large enough to

"That's quite a leap! I believe that the future of improved care for our congenital heart disease patients throughout childhood and into adulthood depends upon meaningful, accurate, and reliable data."

conduct the clinical trials needed to advance our field. We are hopeful that a critical mass of congenital heart programs could potentially collaborate to create an even more robust clinical research database. This may be the start of something big.

For more information about the ACC NCDR PINNACLE Registry, please contact Samantha Risch at srisch@acc.org.

*The NCDR® is the American College of Cardiology's worldwide suite of data registries helping hospitals and private practices measure and improve the quality of cardiovascular care they provide. The NCDR encompasses six hospital-based registries and one outpatient registry.

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Dr. Feltes' article also appears in the Summer 2013 issue of ACC'S *Cardiology magazine - http://www.cardiosource.org/Cardiology and is republished here with permission.*

Letters to the Editor

Congenital Cardiology Today welcomes and encourages Letters to the Editor. If you have comments or topics you would like to address, please send an email to: LTE@CCT.bz, and let us know if you would like your comment published or not.

Medical News, Products & Information

New Technique May Help Regenerate Heart Cells to Treat Heart Disease

Researchers have developed a new technique that might one day be used to convert cells from heart disease patients into heart muscle cells that could act as a personalized treatment for their condition. The research was published online on August 22nd in the *Journal of the International Society of Stem Cell Research, Stem Cell Reports*, published by Cell Press.

The investigators previously reported the ability to convert scar-forming cells in the heart (called fibroblasts) into new, beating muscle in mice that had experienced heart attacks, thereby regenerating a heart from within. They accomplished this by injecting a combination of three genes into the animals' fibroblast cells. "This gene therapy approach resulted in new cardiac muscle cells that beat in synchrony with neighboring muscle cells and ultimately improved the pumping function of the heart," explains senior author Dr. Deepak Srivastava of the Gladstone Institutes and its affiliate, the University of California, San Francisco.

In this latest research, Dr. Srivastava and his colleagues coaxed fibroblasts from human fetal heart cells, embryonic stem cells, and newborn skin grown in the lab to become heart muscle cells using a slightly different combination of genes, representing an important step toward the use of this technology for regenerative medicine. Two other groups recently reported similar results using human fibroblasts.

The team envisions that introducing these genes into damaged hearts by gene therapy might convert fibroblasts into new muscle, thereby improving the function of the heart. "Over 50% of the cells in the human heart are fibroblasts, providing a vast pool of cells that could be harnessed to create new muscle," says Dr. Srivastava. However, additional research is needed to improve the process of reprogramming adult human cells in this way. Ultimately, replacing the genes with drug-like molecules that produce a similar effect would make the therapy safer and easier to deliver.

'Virtual Heart' Precision-Guides Defibrillator Placement in Children With Heart Disease

Newswise - The small size and abnormal anatomy of children born with heart defects often force doctors to place lifesaving defibrillators entirely outside the heart, rather than partly inside - a less-than-ideal solution to dangerous heart rhythms that involves a degree of guesstimating and can compromise therapy.

Now, by marrying simple MRI images with sophisticated computer analysis, a team of Johns Hopkins researchers says it may be possible to take the guesswork out of the process by using a virtual 3-D heart model that analyzes a child's unique anatomy and pinpoints the best location for the device before it is implanted.

A description of the team's work was published ahead of print in *The Journal of Physiology.*

"Pediatric cardiologists have long sought a way to optimize device placement in this group of cardiac patients, and we believe our model does just that," says lead investigator Natalia Trayanova, PhD, the Murray B. Sachs Professor of Biomedical Engineering at Johns Hopkins. "It is a critical first step toward bringing computational analysis to the pediatric cardiology clinic."

If further studies show the model has value in patients, it could spare many children with heart disease from repeat procedures that are sometimes needed to re-position the device, says co-investigator Jane Crosson, MD, a pediatric cardiologist and arrhythmia specialist at the Johns Hopkins Children's Center.

"It's like having a virtual electrophysiology lab where we can predict best outcomes before we even touch the patient," Crosson says.

In adults and in children with normal size and heart anatomy, one part of the device lies under the collar bone, while the other end is inserted into one of the heart's chambers, a standard and well-tested configuration. But in children with tiny or malformed hearts, the entire device has to be positioned externally, an often imperfect setup. Such less-than-precisely positioned defibrillators can fire unnecessarily or, worse, fail to fire when needed to shock a child's heart back into normal rhythm, experts say. In addition, devices that are not positioned well can pack a punch, delivering ultra-strong, painful jolts that frighten children and could even damage heart cells.

"These are lifesaving devices but they can feel like a horse kick to the chest and really traumatize children," Crosson says.

With the Johns Hopkins heart model, scientists say they can find exactly where in relation to a patient's heart the device would be best able to reset the heart by using the least amount of energy and gentlest shock. This translates into longer battery life for the device as well, Trayanova says.

To build the model, the Johns Hopkins team started out with simple, low-resolution MRI heart scans of a child born without a tricuspid valve and right ventricle. Based on these images, the researchers developed a 3-D computer model that allowed them to simulate a dangerous rhythm disturbance during which the heart's strong, regular beats degenerate into weak quivers that, if uninterrupted, could kill in minutes. The model predicted how effectively the defibrillator would terminate this dangerous rhythm when located in each one of 11 positions around the heart. Based on the model, the scientists determined that two particular positions rendered therapy optimal.

A particular advantage of the model is its true-to-life complexity. The model was built using digital representations of the heart's subcellular, cellular, muscular and connective structures - from ions and cardiac proteins to muscle fiber and tissue. The computer model also included the bones, fat and lungs that surround the heart.

"Heart function is astounding in its complexity and person-to-person variability, and subtle shifts in how one protein interacts with another may have profound consequences on its pumping and electric function," Trayanova says. "We wanted to capture that level of specificity to ensure predictive accuracy."

Trayanova and her team also have designed image-based models that pinpoint arrhythmia-triggering hot spots in the adult heart muscle and can help guide therapeutic ablation of such areas. The new pediatric virtual heart, however, is the team's first foray into pediatric cardiology.

Co-investigators on the research included: Lukas Ratner, Fijoy Vadakkumpadan and Philip Spevak, all of Johns Hopkins.

The research was funded by the National Institutes of Health under grant number R01HL103428.

Your Finger's Pulse Holds the Key to Your Heart's Health New Technique from UI Study use Finger's Pulse to Measure Stiffness in the Aorta

Newswise - A University of Iowa physiologist has a new technique to measure the stiffness of the aorta, a common risk factor for heart disease. And it can be as simple as measuring the pulse in your finger.

The new procedure developed by Gary Pierce, Assistant Professor in the Department of Health and Human Physiology, works by placing an instrument called a transducer on the finger or over the brachial artery, located inside the arm just beneath the elbow. The readout, combined with a person's age and body mass index, lets physicians know whether the aorta has stiffened.

Currently, physicians see whether a patient has a hardened aorta by recording a pulse from the carotid artery, located in the neck, and the femoral artery, which is located in the groin. Taking a pulse from the finger or on the arm is easier to record and nearly as accurate, Pierce says. It also works better with obese patients, whose femoral pulse can be difficult to obtain reliably, he adds.

"The technique is more effective in that it is easy to obtain just one pulse waveform in the finger or the brachial artery, and it's less intrusive than obtaining a femoral waveform in patients," says Pierce, first author on the paper, published in the American Journal of Physiology Heart and Circulatory Physiology. "It also can be easily obtained in the clinic during routine exams similar to blood pressure tests."

Heart disease is the leading cause of death for both men and women in the United States, killing about 600,000 people every year, according to the federal Centers for Disease Control and Prevention.

One key to a healthy heart is a healthy aorta. A person's heart has to work harder when the aorta, the large artery that leaves the heart and delivers blood to the body's tissues, stiffens due to aging and an inactive lifestyle. The harder a person's heart needs to work, the higher risk he or she has for developing high blood pressure, stroke and a heart attack.

Since people can live for years without any knowledge of existing cardiovascular problems, this new measurement tool is especially important. It can provide useful diagnostic information for middle-aged and older patients, who are most susceptible to having hardened arteries that can lead to heart disease.

Regular assessments of the aorta may help reduce those risks. Pierce's instrument measures notes the speed, called aortic pulse wave velocity, at which the pulse moves between two points. The UI team validated the new instrument's performance against the carotid-femoral-artery pulse wave velocity tests, considered the gold standard for determining aortic stiffness.

"Finding simple noninvasive methods to measure aortic pulse wave velocity in the clinic may help physicians to better inform middle-aged and older adults about their level of cardiovascular risk," Pierce says, noting that past studies have shown that regular exercise protects the aorta from hardening in those age groups.

The paper's corresponding author is Harald Stauss, Associate Professor in Health and Human Physiology. Other authors from the UI include: Darren Casey, Jess Fiedorowicz, and DeMaris Wilson. Douglas Seals from the University of Colorado-Boulder, and Timothy Curry and Jill Barnes from the Mayo Clinic in Rochester, Minn. also contributed to the paper.

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Surgical Mortality Percentage Categorized by Complexity of Procedure				
RACHS-1* Category	CM** 1 Year Average	STS*** 1 Year Average	CM 4 Year Average	STS 4 Year Average
1 (least complex)	0.0	0.5	0.0	0.7
2	0.0	1.1	1.1	1.1
3	2.9	3.3	3.7	3.7
4	5.1	7.1	8.2	7.1
5 & 6 (most complex)	15.4	14.3	20.8	16.4

*Risk Adjustment for Congenital Heart Surgery classification

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