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Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide

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Full-Term Newborn With Tachypnea at Twenty-Four Hours of Life May Be Sign of Complex Underlying Problem

By Kamlesh K. Jha, MBBS, MD, FAAP; Saranya Ravichandran, MD, PGY3; Jovina Benedito, BSN, RN; Pragya Jha

Case Report

A 40-week, appropriate for gestational age male with Apgar score 9 and 9 at one and five minutes was born vaginally to a 26-year-old, G2P1 mother. His birth weight, length and head circumference 3620 grams, 50.8cm and 35cm, respectively. The mother's blood type was A positive; HIV, HBsAg, RPR, GBS and urine toxicology were all negative. Mom received adequate antenatal care and had two antenatal ultrasounds at 21 and 29 weeks of gestation. Mom had no medical problems. Five minutes after birth a paucity of left-upper extremity movement, specifically abduction at the shoulder, was observed; flexion at the elbow and weak grasp reflex compared to the right was also observed. A chest X-ray revealed no broken bones. The baby received routine newborn medications, namely; Erythromycin eye ointment, 1mg IM of vitamin K injection and 10 micrograms of Hepatitis B vaccine, which was given intramuscularly. The baby stayed with the mother and tolerated feeding without any problem.

At twenty-four hours of age, the baby was screened for Critical Congenital Heart Disease (CCHD), and it was negative (pre-ductal 95% and post-ductal 97%). One hour after screening for CCHD, the registered nurse (RN) noted intermittent tachypnea and tachycardia with a missing beat on the monitor without having desaturation. The RN reported this new development to the neonatologist. On evaluation, the neonatologist noted the vitals were within normal range except for the intermittent tachypnea for which the neonatologist decided to monitor baby in the special care unit with continuous cardiorespiratory monitoring and continuous clinical examinations. The neonatologist ordered an electrocardiogram which was reported normal for age. CBC, CXR and a blood culture were done and the baby was started on IV Ampicillin and Gentamicin. The CXR reported by the radiologist was unremarkable. The neonatologist discussed the findings with the radiologist and spoke about the possibility of performing a fluoroscopy to observe diaphragmatic movement. Based on the chest X-ray (Figure 1), in which the position of both domes of the diaphragm was in a normal position, the radiologist came to the conclusion that doing fluoroscopy and exposing the baby to unnecessary radiation should be avoided.

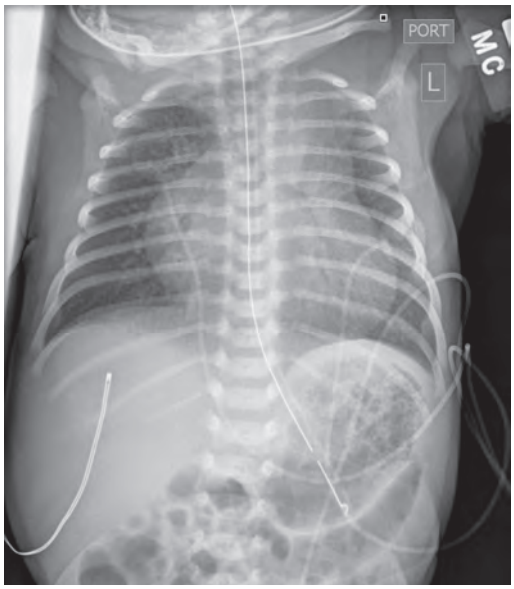

Figure 1

The baby continued to tolerate feeding and remained hemodynamically stable. The neonate was evaluated by the rehabilitation physician and at discharge the mother was advised to schedule a follow-up appointment with Schwab Rehabilitation Center.

At 48 hours of age, a systolic heart murmur was heard and the on-call neonatologist decided that he would consider doing an echocardiogram if the baby deteriorated or did not improve. At 66 hours of life, a capillary blood gas was done in room air and was unremarkable. At 72 hours of life, (DoL4), the morning RN, before getting a sign-out from the night RN, observed the monitor beeping frequently. The morning RN saw the baby and thought the baby appeared mottled, breathing fast with retractions. The morning RN asked the night RN to call the neonatologist to evaluate. After the neonatologist evaluated the baby, he ordered HFNC 3.5L 30% FiO_2 and decided to transfer the baby to the tertiary care center for further evaluation and management. The morning RN saw the baby saturating 99-100% and reduced FiO_2 to 21% and flow to 2L. The morning RN said she did not want to close the PDA (Patent Ductus Arteriosus) by giving high oxygen. ABG at 74 hours of life, on 3L flow with 21% FiO_2, was 7.34/37/45/20. Before the neonate was transferred, the morning RN repeated the screen for CCHD. It was a positive screen with pre-ductal saturation of 70% and post-ductal saturation of 100%. Later, the neonate was transported in the ambulance with HFNC.

On the way to the tertiary care center, the transport team, which had a nurse and respiratory therapist, along with EMS personnel, increased FiO_2 to 100%. The baby did not deteriorate, but immediately on arrival to the baby suffered a hemodynamic collapse. Subsequently, the neonate was intubated and placed on SIMV pressure support ventilation. A Stat Echocardiogram was ordered. ABG was done which revealed metabolic acidosis with high PaO_2 (7.16/19/192/6.8/-19.7). The EKG revealed Vent rate of 144bpm, atrial rate of 144bpm, P-R interval 110ms, QRS duration 086ms, QT interval 284ms, P-R-T axes 066-119-043 degrees, and QTc interval 439ms. Right atrial enlargement and right ventricular hypertrophy were observed. Absent q waves and prominent p waves suggested severe right-sided hypertrophy. An echocardiogram reported a hypoplastic left heart with moderately restricted ASD, restrictive PDA, all right-to-left with a gradient of 30-40mmhg and aortic and mitral valve atresia. Contractility of heart was satisfactory. A PGE infusion was started after the echo report and the patient was then transferred to the cardiac center where he underwent cardiac surgery.

Discussion

Congenital Heart Disease is one of the most common major newborn anomalies, occurring at a rate of eight cases per thousand live births. Almost 20% of neonatal deaths are due to congenital heart anomalies[1]. Hypoplastic Left Heart Syndrome (HLHS) occurs in one in 5,000 live births[2]. HLHS includes a spectrum of left-side heart anomalies in which aortic and mitral valves are stenosed or atretic, plus hypoplastic or atretic ascending aorta and left ventricle. The Hypoplastic Left Heart Syndrome is a total mixing lesion where pulmonary venous return shunted from the left atrium via an Atrial Septal Defect (ASD) or Foramen Ovale to the right side where it mixes with the systemic venous return. Ideally, all congenital malformations should be diagnosed antenatally to plan the optimal route and place to deliver the baby in order to optimize care and improve outcome. Clinically, cyanosis may not be evident with the naked eye in the first 48 hours, but because of hypoperfusion, clinicians may suspect HLHS where oxygen saturation measured by pulse oximeter is low. In any newborn with cyanosis, if Congenital Heart Disease is one of the differentials, and even if echo is not possible, the clinician should start the baby on prostaglandin infusion. The infant should be transferred to a tertiary care institute for an echocardiogram, further evaluation, and management. Surgical reconstruction is done after a diagnosis of HLHS is confirmed by echocardiogram.

Recommendations

1. A Critical Congenital Heart Disease screen should be repeated in all newborns at 48 hours of age to increase the sensitivity.
2. Any newborn with respiratory distress with a normal chest x-ray should have an echocardiogram.
3. Do not give 100% supplemental oxygen to any newborn with respiratory distress with normal lung parenchyma on chest x-ray.
4. Pediatricians and neonatologists who work at Level I & II newborn nurseries should always remember the standard of care to start prostaglandin E1 (PGE1) infusion in a newborn where suspicion of underlying Cyanotic Congenital Heart Disease is a possibility.

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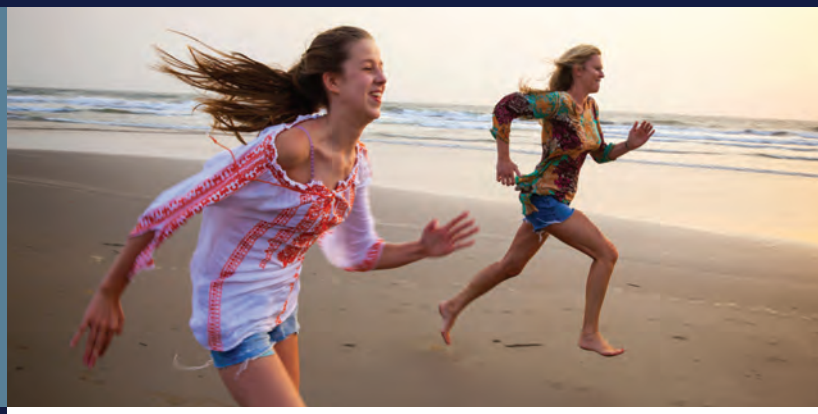

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Archiving Working Group of the International Society for Nomenclature of Pediatric and Congenital Heart Disease: Image of the Month #15

By James D. St. Louis, MD; Jorge M. Giroud, MD; Charles W. Shepard, MD; Allen D. Everett, MD; Robert H. Anderson, MD; Vera D. Aiello, MD; Diane E. Spicer, BS; Jeffrey P. Jacobs, MD

Common Arterial Trunk

- ***International Paediatric and Congenital Cardiac Code (IPCCC):*** 09.01.01
- ***AEPC Derived Terms:*** Common arterial trunk (*truncus arteriosus*)
- ***EACTS-STS Derived Terms:*** Truncus arteriosus
- ***ICD11 Derived Terms:*** Common arterial trunk (*truncus arteriosus*)

Commentary

The present column shows in figures the major morphological features of the congenital malformation known as Common Arterial Trunk, or *Truncus Arteriosus Communis*. It represents one form of single outlet of the heart, defined as a common artery originating from the base of the heart, and supplying directly the systemic, the pulmonary, and the coronary circulations (Figure 1). The presence of a sub-truncal interventricular communication, usually termed a Ventricular Septal Defect (VSD), is the rule. In more than two-thirds of the cases, the defect has a muscular postero-inferior border (Figure 1), interposing between the hinges of the leaflets of the tricuspid valve and truncal valves. It also separates the defect from an intact membranous septum. The circumference of the truncal root may be supported predominantly by one or other of the ventricles, but more usually takes a balanced form. Usually the defect is non-restrictive. Rarely the common trunk arises exclusively from the right ventricle in association with an intact ventricular septum. The leaflets of the truncal valve can also close on the crest of the muscular septum during ventricular diastole, thus removing the potential for interventricular shunting.

Differential diagnosis should include the solitary aortic trunk arising from the base of the heart in cases of pulmonary atresia when the remnants of the pulmonary trunk cannot be identified and the arterial supply to the lungs comes from extrapericardial arteries, usually major aorto-pulmonary collateral arteries from the descending aorta.

The anatomy of the aorta and central pulmonary arteries, as components of the common arterial trunk, is quite variable, and is generally used to classify the anatomical variants. Collett and Edwards, in 1949[1] proposed a system of classification that is still widely used. It is based on the pattern of origin of the central pulmonary arteries. Their "Type I" was described as a common trunk from which emerged a short confluent pulmonary component, which then branched to give rise to the right and left pulmonary arteries. In "Type II" and "Type III", there was not confluent pulmonary component, but separate origins of the pulmonary arteries, which arose either from the dorsal aspect (Type II) or from each side (Type III) of the common arterial trunk. Their Type IV was defined on the basis of absence of intrapericardial pulmonary arteries. This variant is now recognized as a solitary arterial trunk, rather than a common trunk as described above.

Van Praagh and Van Praagh, in 1965, modified this system[2], grouping together the Types II and III of Collet and Edwards. They then introduced their own types 3 and 4, to account for discontinuous pulmonary arteries, and hypoplasia of the aortic component, respectively. More recently, Russell and co-workers proposed a categorization that expanded an initial suggestion by van Praagh and Van Praggh, namely that the cases could be distinguished according to the dominance of the aortic or the pulmonary components of the common trunk[3]. This approach was argued to comply with the needs of the surgical approach to the anomaly[4].

When there is pulmonary dominance, it is common to find obstructive lesions of the aortic arch, such as coarctation or interruption of the aortic arch (Figure 2). In such cases, there is a widely patent arterial duct, which laces the pulmonary arterial confluence in continuity with the descending aorta. In the setting of aortic dominance, usually the pulmonary arterial branches take a confluent origin from the left and posterior aspect of the common trunk (Figures 3 and 4). Very recently, it has been noted that, in a significant number of cases, the pulmonary component can arise from a truncal arterial sinus[5]. It is also important to note occasionally crossing of the pulmonary arteries as they arise from the common trunk (Figure 1).

The semilunar leaflets of the truncal valve vary in number from two to six, with the valve being trifoliate in more than half of the cases. In one anatomical series, dysplastic valves were present in three-fourths of the specimens[6], and were usually present when there were four leaflets. Such leaflets show uneven sizes and irregular thickening (Figure 5), and may cause valvar incompetence or stenosis.

"The present column shows in figures the major morphological features of the congenital malformation known as Common Arterial Trunk, or Truncus Arteriosus Communis. It represents one form of single outlet of the heart, defined as a common artery originating from the base of the heart, and supplying directly the systemic, the pulmonary, and the coronary circulations (Figure 1)."

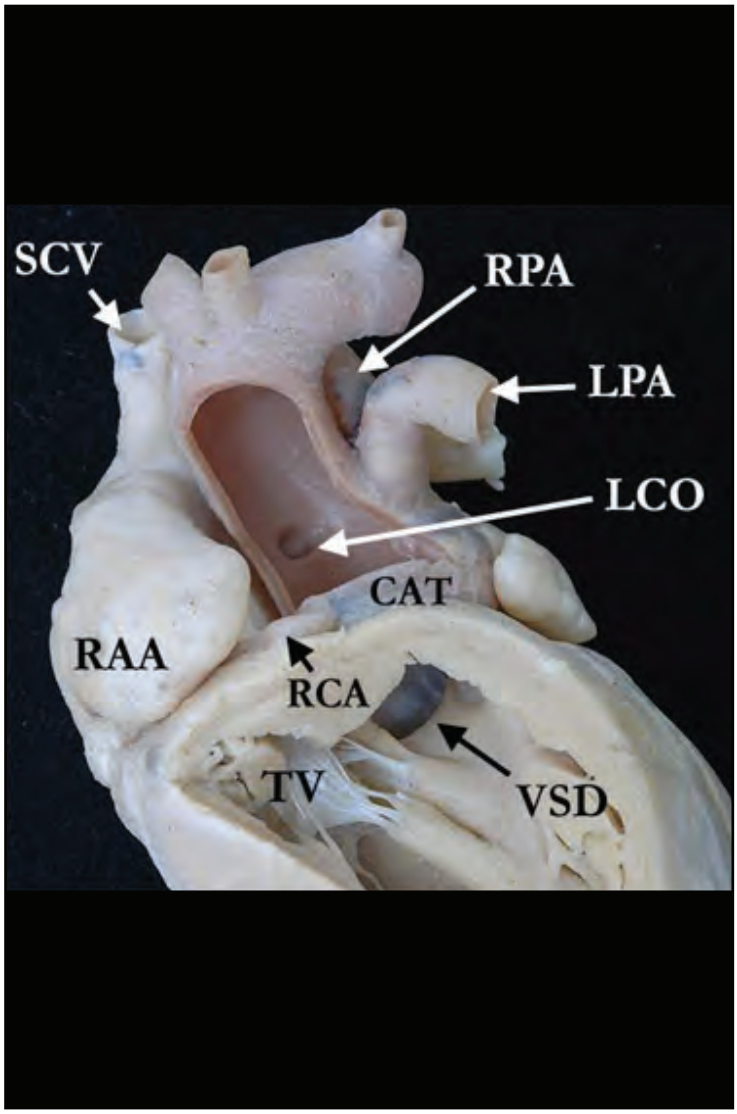


Figure 1

Modality: Anatomic specimen

Orientation: Right anterior view

Description: This figure illustrates a heart with a common arterial trunk supplying the aortic, pulmonary and coronary circulations. The right pulmonary arteries bifurcate well above the sinutubular junction at the lateral aspect of the common trunk. There is an unusually high take off of the left coronary artery (LCO). The right coronary artery (RCA) arises normally and can be visualized on the epicardial surface. The right ventricle (RV) has been windowed, demonstrating the ventricular septal defect (VSD) beneath the truncal valve with a muscular rim along its posterior inferior aspect. This muscular rim separates the leaflets of the tricuspid (TV) and truncal valves. Note the crossing of the right and left pulmonary arteries. (SCV-superior caval vein, RAA-right atrial appendage).

Contributor: Diane E. Spicer, BS

Institution: University of Florida, Gainesville, FL USA

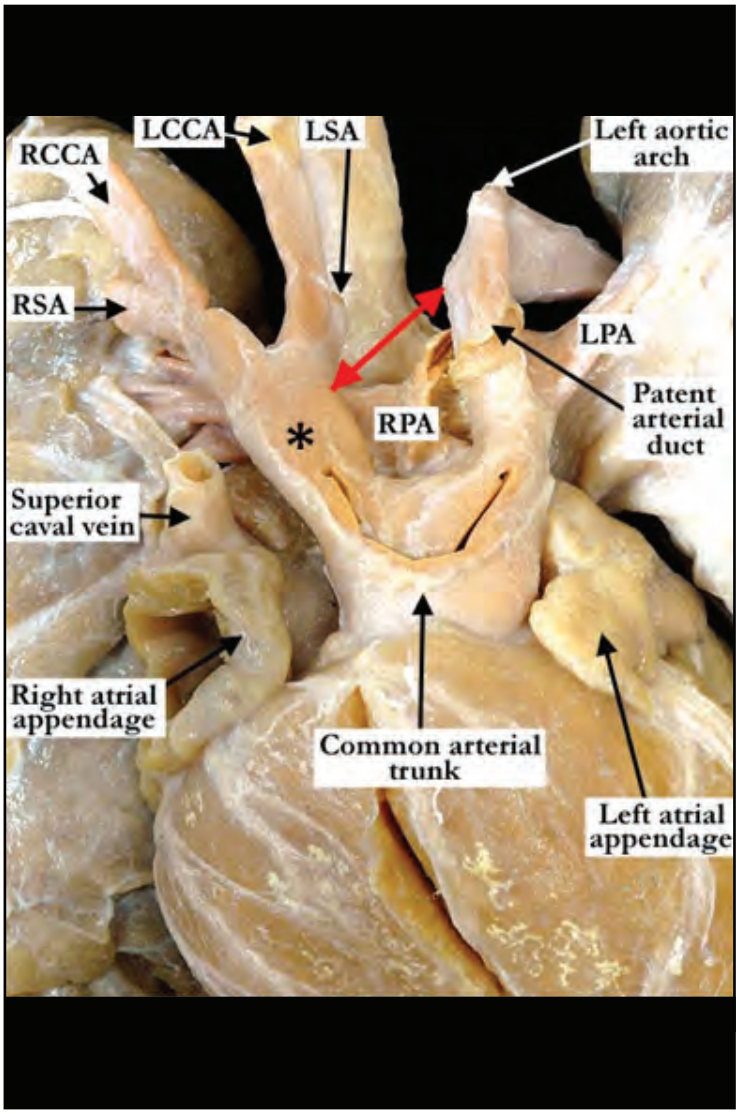


Figure 2

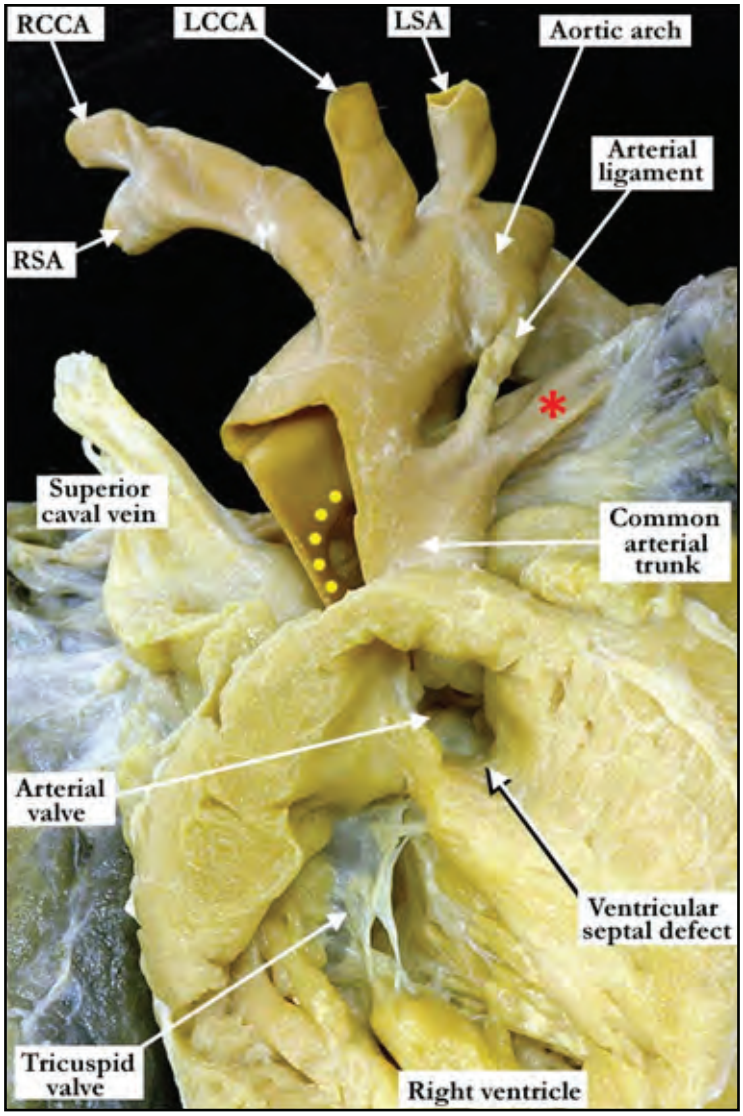
Modality: Anatomic specimen

Orientation: Frontal view

Description: This heart specimen illustrates a common arterial trunk with an interrupted aortic arch and pulmonary dominance. The ascending aorta (asterisk) extends from the common trunk to give rise to the brachiocephalic vessels with a normal branching pattern and a left-sided arch. The aortic arch is interrupted (red double headed arrow) distal to the left subclavian artery (LSA) and proximal to the arterial duct. The patent arterial duct connects the common arterial trunk to the descending aorta. The right (RPA) and left (LPA) pulmonary arteries bifurcate from a confluent pulmonary component that arises from the posterior aspect of the common trunk. (RSA-right subclavian artery, RCCA-right common carotid artery, LCCA-left common carotid).

Contributor: Diane E. Spicer

Institution and Image Source: Medical University of South Carolina, Charleston, SC, USA



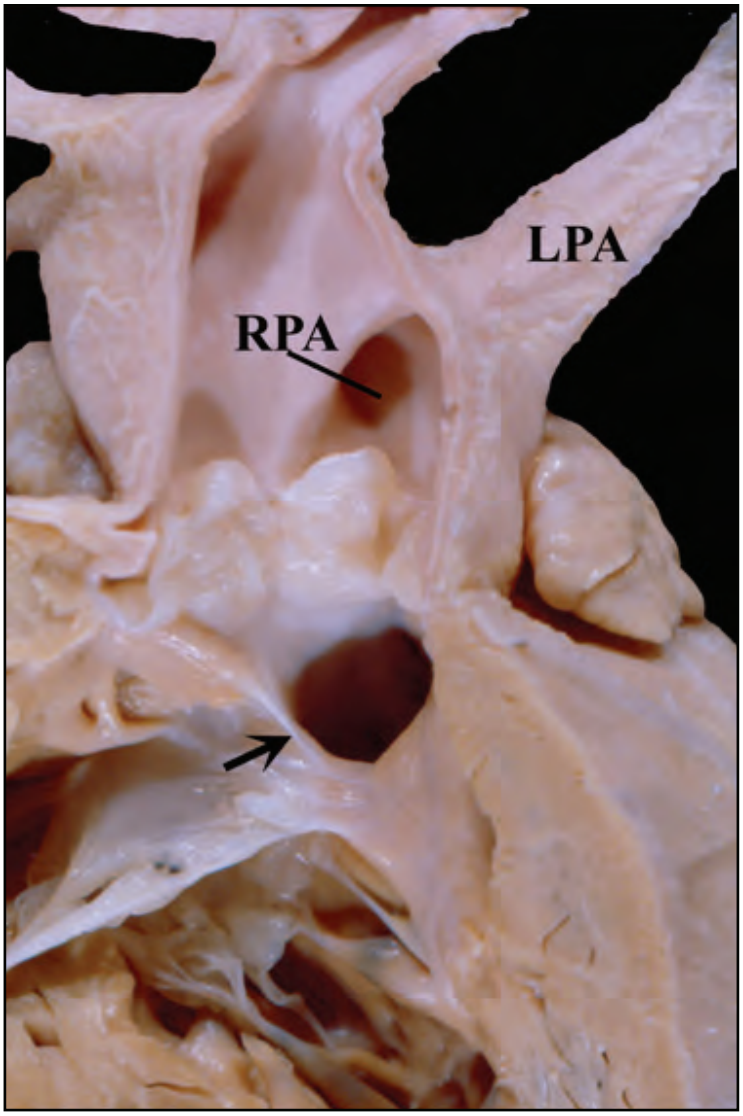


Figure 3

Modality: Anatomic specimen

Orientation: Right-anterior view

Description: The right ventricular free wall has been windowed to show the origin of the common arterial trunk and the subarterial interventricular communication. There is aortic dominance and a normal left aortic arch, with the usual branching pattern of the brachiocephalic arteries. The right (yellow dots) and left (red asterisk) pulmonary arteries have a separate, although closely approximated, origin from the posterior aspect of the common arterial trunk. An arterial ligament arises at the point where the left main pulmonary artery bifurcates from the common trunk.

Contributor: Diane E. Spicer

Institution and Image Source: Medical University of South Carolina, Charleston, SC, USA

Figure 4

Modality: Anatomic specimen

Orientation: Frontal view with opened right ventricular outflow tract

Description: This specimen illustrates a common arterial trunk arising from the base of the heart. There is aortic dominance and the pulmonary arteries take origin separately from the posterior and left margin of the common trunk. The valvar leaflets are mildly dysplastic. There is fibrous continuity postero-inferiorly between the leaflets of the mitral and tricuspid valves (arrow). LPA- left pulmonary artery; RPA- orifice of the right pulmonary artery.

Contributor: Vera D. Aiello

Institution and Image Source: Heart Institute (InCor), University of Sao Paulo School of Medicine, Brazil

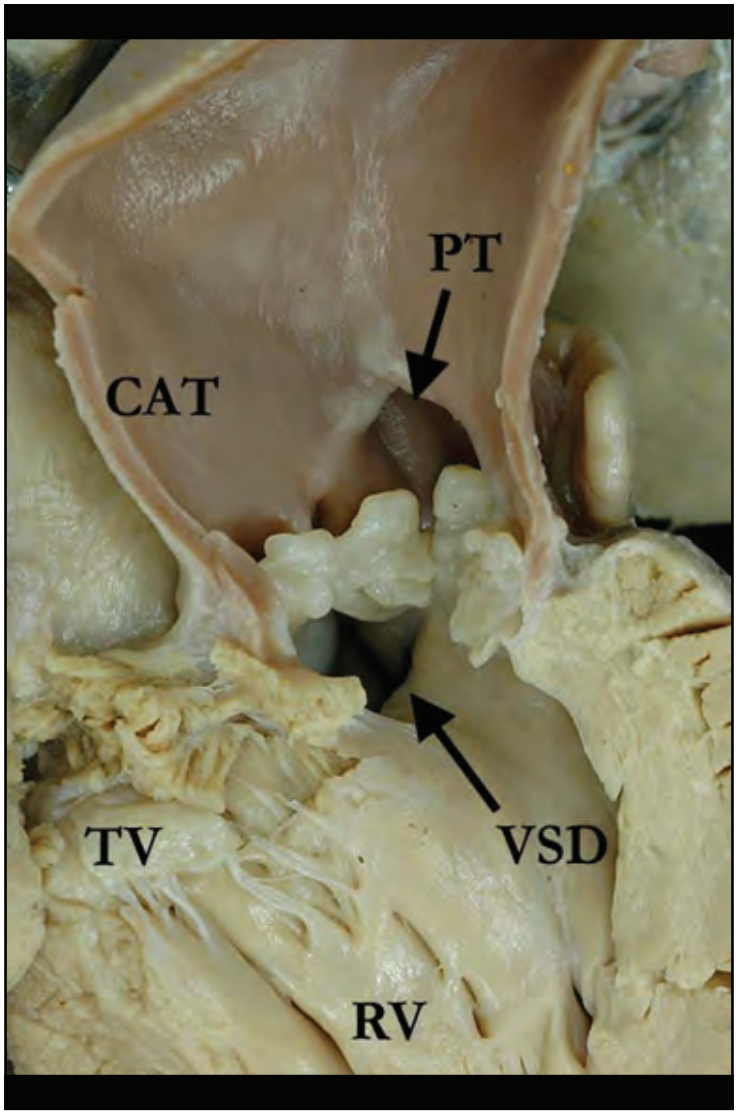


Figure 5

Modality: Anatomic specimen

Orientation: Anterolateral view

Description: This figure demonstrates the right ventricular (RV) aspect of the common arterial trunk (CAT). It shows the low take-off of the pulmonary trunk (PT), just at the sinutubular junction. The valve is markedly dysplastic and is quadricuspid. The ventricular septal defect appears restrictive.

Contributor: Diane E. Spicer, BS

Institution and Image Source: University of Florida, Gainesville, FL, USA

"The semilunar leaflets of the truncal valve vary in number from two to six, with the valve being trifoliate in more than half of the cases. In one anatomical series, dysplastic valves were present in three-fourths of the specimens[6], and were usually present when there were four leaflets. Such leaflets show uneven sizes and irregular thickening (Figure 5), and may cause valvar incompetence or stenosis."

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Please visit the website of the *International Society for Nomenclature of Pediatric and Congenital Heart Disease*, www.ipccc.net, where the Virtual Atlas illustrates terms found in ICD-11. It is under construction but a 2015 prototype is available to view.

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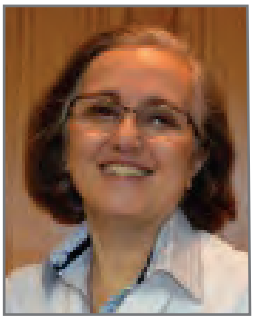

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The Archiving Working Group is a committee of the *International Society of Nomenclatures for Pediatric and Congenital Heart Disease* (ISNPCHD).

The purpose of AWG is to identify, develop and maintain a virtual encyclopedia of representative images that illustrate the codes and definitions of the Working Groups of the ISNPCHD.

www.ipccc.net

After Losing a Son - A Family's Journey

By Darren Sudman

I was ready. It was my second child. Just two years earlier, I had become an "expert" expectant parent. I read all of the books. I had all of the product guides. I was a big fan of the Baby Bjorn. I knew that Britax made a really good car seat. Alimentum was a great formula for colic. I preferred Pampers over Huggies. My Baby Einstein DVD collection was complete. What could go wrong?

Simon arrived on October 21, 2004. He scored an 8 and 9 on his APGAR test. He was average for height and weight. He smiled for the first time at 47 days; and died 43 days later. He was taking a nap after dinner and never woke up.

What did I do? Was there something that I didn't do? His tests were normal! They told us he was healthy!

Our pediatricians came to the hospital that night. I don't remember much except that they told us to get our hearts checked because babies don't just die. I would later learn that this was very unique and profound advice. This curiosity would define our journey and ultimately impact the lives of so many children.

Phyllis and I made appointments. We also made an appointment for our two-year old daughter, Sally. We started researching Sudden Infant Death Syndrome (SIDS). We had read about this "boogie man" in those books. Don't smoke around the baby. Don't sleep with the baby. Don't use fluffy blankets or bumpers. No stomach sleeping. The cause is unknown.

Phyllis was diagnosed with Long QT Syndrome, an arrhythmia. She had never fainted or felt like her heart was racing. In fact, she had never gotten her heart screened. Childbirth is the most physically stressful event of a woman's life. Should we check their hearts? Would a NASCAR driver ever enter a race without checking the car engine?

Now, we had to research Long QT Syndrome too. We met with the late Dr. Arthur Moss in Rochester. He maintained a Long QT Registry. Phyllis got genetic testing, and was entered into the registry. The testing came back negative.

Dr. Michael Ackerman at the Mayo Clinic found that up to 15% of all SIDS deaths could be linked to Long QT Syndrome. We had to meet with him too. But wait. The heart was never mentioned in any of the SIDS books or articles. We knew about all of the environmental stuff that were believed to contribute to SIDS.

Legacy Youth Tennis and Education Center receives an AED from Simon's Heart.

If the heart could be the reason that one out of every 10 infants die suddenly, shouldn't we be talking about that too? Shouldn't this warrant parents and surviving siblings to get their hearts checked after any SIDS death?

On May 16, 2005, the boat from Boston College finished first at the Dad Vail Regatta. Shortly thereafter, Scott Laio collapsed and died. We never knew that seemingly healthy student athletes could just die. We were learning that sudden cardiac arrest was not just an adult thing and that our society was not doing much to prevent it.

Recognizing there were two ways to prevent sudden cardiac death - primary intervention and secondary intervention - we opted for primary and launched Simon's Fund (which is now called Simon's Heart). We began providing free heart screenings for students. The screening included a medical and family history, a physical exam, an ECG exam and an echocardiogram (for about 10% of the students). Since we began in 2006, we have screened over 17,000 and helped 123 discover heart conditions.

We partnered with the Pennsylvania Chapter of the American Academy of Pediatrics to educate families about the warning signs. Our posters were displayed in offices throughout the Commonwealth. We authored legislation - the Sudden Cardiac Arrest Prevention Act - that became law in Pennsylvania in 2012. Since, it has been adopted by eleven other states. It requires parents, coaches and student athletes to learn about the warning signs of cardiac arrest and mandates that students who exhibit symptoms be removed from play and cleared by a licensed medical professional. The law reaches two million students annually, and has educated over 500,000 coaches nationwide.

Our biggest contribution to the primary prevention effort is the development of a nationwide digital cardiac registry of seemingly healthy kids. In 2014, we launched HeartBytes. Our heart screenings are now conducted online and all of the data (including digital images) is stored in the cloud. The data is deidentified and made available to qualified researchers, free of charge. HeartBytes is used by several screening organizations around the country, and it contains over 10,000 electronic health records. www.simonsheart.org/protect-this-heart/heartbytes/

Our goal is to change the standard of care so that students get their hearts screened, just like their sight and hearing. This approach will save lives and help us better understand the seemingly healthy pediatric heart, which in the long run, should reduce costs and improve the cardiac health of adults.

We also need a backup plan because watching for warning signs and screenings will not eliminate sudden cardiac arrest. Simon's Heart embraces secondary prevention with awareness campaigns and a crowdfunding website. GotAED is dedicated to placing AED devices where kids learn and play. It empowers youth leagues, youth facilities and schools to raise funds and obtain an AED device. AED Madness is an annual campaign to educate college basketball fans around Philadelphia. We appear at five home games during the college basketball season, educate the fans with a message from the coach, and donate an AED device to a local youth facility. The Overtime Challenge is a partnership with the Philadelphia Flyers. Every time the Flyers win a game in sudden death, an AED device is donated to a local youth facility.

Simon's Heart was not created to solve the sudden cardiac arrest problem. It exists to fill a void left by the medical community. It brings awareness to the problem, inspires curiosity and innovation, and proves concepts. Ultimately, the solution to this problem lies with the medical community.

Phyllis and Darren Sudman, co-founders of Simon's Heart, surrounded by students who have discovered heart conditions at a screening.

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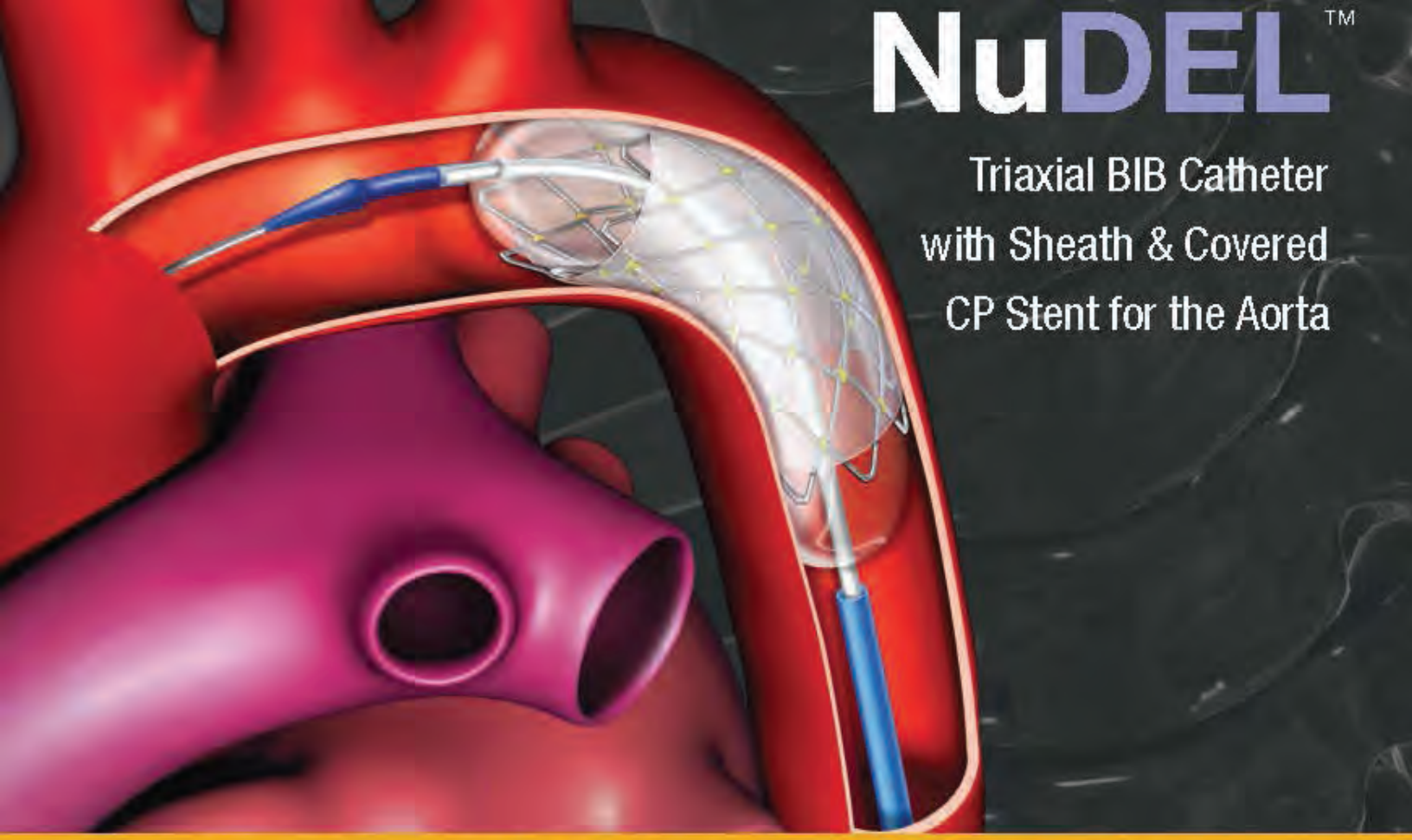

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The First International PDA Symposium – Memphis, Tennessee, May 18-19, 2018

By Ranjit Philip, MD; Mark Weems, MD; Ajay Talati, MD; Leah M. Apalodimas, MSN, APN, CCRN, CPNP; Shyam Sathanandam, MD, FCSAI

The *International PDA Symposium* was the first dedicated international conference strictly geared towards management of the patent ductus arteriosus (PDA) with a special focus on trans-catheter techniques for closure of the PDA in extremely low birth weight preterm infants. Having come a long way (80 years) since the first surgical ligation of a PDA by Dr. Robert Gross, there still remains no consensus on whether a PDA needs to be closed in children born premature and if so, when and how it needs to be closed. This meeting provided a unique opportunity for different sub-specialties to come together in one forum to discuss and debate variations in management practices. State-of-the-art presentations were given by world-class experts in the field of interventional cardiology, neonatology, cardiac surgery, and cardiac anesthesia among others (Figure 1). Being the first meeting of its kind, there was an overwhelming response with registrations surpassing the cap of 180 (pediatric cardiologists, neonatologists, cardiac surgeons, cardiac anesthesiologists, CRNAs, cardiac perfusionists, nurse practitioners, nurses, sonographers, trainees from cardiology and neonatology, and industry partners) from across the United States, and seven other countries and four continents (Figure 2). Registrants left the meeting not only with the thrill of the live case of transcatheter PDA occlusion in a 700 grams, 24 weeks' gestation premature neonate but also with an enhanced insight into which PDAs may benefit from closure and what might be the optimal timing for closure. There was a hands-on echocardiography workshop geared towards neonatologists (Figure 3). Better insight was provided on the different methods of closure (medical therapy, surgical ligation, trans-catheter closure) that may have different biological effects on the neonate and that the short and long-term outcomes from one technique may be different from the other and hence must be evaluated separately. There was also a commitment from most speakers to offer continuing education and consultation for registrants throughout the year.

Figure 1. International PDA Symposium – State of the art presentations given by world-class experts.

The symposium aptly began with a comprehensive overview on ductal physiology by Dr. John Jeffrey Reese (Vanderbilt University). Dr. Shahab Noori (Children's Hospital Los Angeles) then discussed "What is a hemodynamically significant PDA?" This was followed by talks on the effect of PDA on BPD, the heart, and pulmonary vascular disease as well as echocardiographic methods to assess the PDA in a preterm infant.

The focus of the much awaited second session was: Neonatology and Cardiology perspectives of management of PDA in the premature infant and who better than Dr. William Benitz (Stanford University) to deliver an outstanding presentation on, "What the current evidence suggests". The participants were equally amazed with the captivating performance by Dr. Matthew Gillespie (Children's Hospital of Philadelphia) on the cardiologist's perspective.

The next session transitioned to the discussion of techniques of PDA closure including pharmacotherapy, surgical techniques and the introduction of trans-catheter PDA closure by none other than Dr. Evan Zahn (Cedars-Sinai Heart Institute). The registrants were also enlightened by an excellent hemodynamic talk and review of post ligation syndrome by Dr. Patrick McNamara (The Hospital for Sick Children, Toronto).

The afternoon session was dedicated to trans-catheter PDA closure in extremely low birth weight infants. After presentations on transport and anesthesia management of the extremely low birth weight infant in the catheterization lab and echocardiographic guidance for trans-catheter PDA closure, Dr. Shyam Sathanandam (LeBonheur Children's Hospital, University of Tennessee) showcased the mounting Memphis experience with trans-catheter PDA closure in neonates < 1000 grams. This was followed by an enchanting presentation by Dr. Neil Wilson (Children's Hospital Colorado), one of the pioneers of bedside trans-catheter PDA closure. In the true spirit of being an international conference, Dr. Guiti Milani (The Necker University Hospital for Sick Children in Paris) ended the session discussing the French Multicenter Registry Data on trans-catheter PDA closure in premature infants. Future directions deliberating the importance of thoughtful designing of randomized control trials were then nicely summed up by Dr. Carl Backes (Nationwide Children's Hospital) and Dr. Rush Waller (LeBonheur Children's Hospital, University of Tennessee).

There were three breakout sessions. Breakout session #1 focused on ductal dependent pulmonary circulation. After a good overview

Figure 2. Registrant map from across the United States and four continents.

of ductal dependent pulmonary circulation lesions, Dr. Athar Qureshi (Texas Children's Hospital) gave a wonderfully crafted talk on ductal stenting which led to a riveting, yet entertaining debate on the Blalock-Taussig shunt versus ductal stenting between Dr. Christopher Knott-Craig (LeBonheur Children's Hospital, University of Tennessee) and Dr. Andrew Glatz (Children's Hospital of Philadelphia). Breakout session #2 focused on targeted neonatal echocardiography (TnECHO) and was an extremely popular session and a big draw to many of the registrants as they were made privy to our state-of-the-art simulation laboratory for hands-on echocardiography on actual patients

Figure 3. Hands-on Echocardiography workshop at the Simulation Center.

with PDAs as well as simulation models. Breakout session #3 was devoted to nursing which also had a significant turn out with interactive case studies and specific nursing care modules. The day ended with a heavy hors d'oeuvres reception and a trip to the Memphis barbecue festival.

The highlight of the symposium undoubtedly was the live transcatheter PDA closure case in the morning on Day 2 (Figure 4). Participants were at the edges of their seats as Dr. Shyam Sathanandam performed a transcatheter PDA closure in an extremely low birth weight, 700 grams, 24 weeks preterm infant. As seamless and as quickly as the procedure went, it had its share of theatrical twists, nonetheless concluding well with an appropriately placed device in a hemodynamically stable neonate. This was followed by an eloquently delivered talk by Leah Apalodimas, APN, PNP on "Elements of a Comprehensive PDA Program" where the main take-home points were teamwork of the multiple specialties, the importance of meticulous patient selection and the learning curve involved with the individual operator. The oral abstract session then followed. It was a testament to the reputation of the symposium that there was an abstract presenter who traveled from Israel for the meeting.

Dr. Athar Qureshi then shared results of a survey that we conducted electronically with an equally distributed (neonatology and cardiology) participant pool to evaluate practice variations in the management of PDA. This was well received and instigated a lot of discussion that blended nicely into the focal point of this meeting which was the panel discussion. The panelists included all the eminent invited speakers from cardiology and neonatology mentioned above (Figure 5) as well as Dr. Ron Clyman (via teleconference). The ultimate goal of the panel discussion was to formulate a joint statement and recommendations from this symposium. What was agreed upon was that as a medical community we need to be more intelligent and mindful and stop doing trials with crude measures of significance and no outcome. What most panelists also agreed on was that if PDA closure via the trans-catheter route was to be attempted, the ideal timing is between 2-4 weeks of age. With the advent of transcatheter PDA closure, this is our moment of opportunity to do a well thought out trial on the appropriate patient population (i.e. babies with the biggest shunt with at least physiologic consequences with well identified morbidities) without lumping all treatments together.

The symposium kept registration fees well below the national average through the generous support of our sponsors. We have had glowing reviews from the participants

Figure 4. Live Broadcast of Transcatheter PDA Closure in a 700 Grams ELBW Infant during the International PDA Symposium.

and the invited speakers themselves on the symposium and the content. The discussions from the *First International PDA Symposium* will be published in a special issue of Congenital Heart Disease as 24 separate review articles written by experts from the field. The *International PDA Symposium* will continue to serve as a platform to bring the leaders of the field together to improve care and outcomes of premature infants. Based on the feedback, plans are already underway for the *Second International PDA Symposium* to be held in Memphis, Tennessee in April 2019.

Figure 5. Panel discussion - panelists included all the eminent invited speakers from cardiology and neonatology.

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[1] "International Multicentre Clinical Device Investigation on Safety and Effectiveness of the Nit-Occlud® Lê VSD Spiral Coil System for VSD Occlusion" (clinicatrials.gov identifier NCT00390702).

[2] "The Nit-Occlud® Lê VSD Registry", publication in preparation.

Medical News, Products & Information Showcased at the Radiological Society of North America's (RSNA) Annual Meeting 2018

Compiled and Reviewed by Kate Baldwin and Tony Carlson

DiA Imaging Analysis, an AI-Powered Ultrasound Analysis Company, Demonstrated its Automated Cardiac Toolbox at the *2018 RSNA* Meeting

DiA Imaging Analysis Ltd. (DiA), a provider of artificial intelligence (AI)-powered ultrasound analysis tools, offers fully automated, easily implemented tools that enable quick, objective and accurate ultrasound analysis. These tools are based on advanced, proprietary pattern recognition and sophisticated machine learning algorithms, offering ultrasound user supporting tools as an integrated part of their workflow. Part of DiA's audience are point-of-care clinicians, such as cardiologists, ED physicians, GP's, etc., who use portable and mobile ultrasound for early diagnosis and patient follow-up. During the *RSNA* summit, DiA presented its "LVivo Toolbox" that brings a new era of automation and efficiency to cardiac ultrasound analysis, allowing clinicians to automatically measure, track and evaluate cardiac ultrasound images.

Currently, most ultrasound evaluation is done visually, manually or semi-automatically -- which is subjective, time-consuming, error-prone, cumbersome and highly dependent on the user's experience.

DiA's LVivo Toolbox supports the clinician's decision by providing objective, fast and accurate AI tools anywhere, anytime as part of their ongoing workflow.

At *RSNA*, the company presented its "LVivo EF" tool, fully integrated as part of the GE Healthcare's VSCAN Extend handheld mobile ultrasound and demonstrated its cloud- based automated abilities with Google Cloud.

LVivo EF fully and automatically calculates Ejection Fraction and end-systolic, end-diastolic left ventricular volumes by automated edge detection of the left myocardial wall using apical 4- and/or 2-chamber views based on DiA's AI algorithms that imitate how the human

eye identifies borders and motion. LVivo EF is used to evaluate patients with CHF, cardiogenic shock, etc., monitoring patient fluid balance in the cardiac unit and emergency departments, ICU and anesthesia departments.

"As the use of point-of-care ultrasound devices expands across the industry, and AI plays a growing role in ultrasound analysis, our cardiac toolbox is the beginning of transforming point-of-care ultrasound to a more consistent and patient-centered process," said DiA CEO and Co-Founder Hila Goldman-Aslan. "We already see clinicians across point-of-care settings recognizing how LVivo Toolbox empowers their real-time decision-making and enhances efficiency."

An additional tool that was demonstrated at *RSNA* was DiA's LVivo SAX (Parasternal Short Axis) which provides an automated global and segmental LV function analysis from the parasternal Short Axis (SAX) view, allowing clinicians to assess global cardiac function in CHF and shock patients, as well as segmental abnormalities in patients with ACS (acute coronary syndrome). LVivo SAX compliments DiA's LVivo SWM and LVivo Strain tools that automate the evaluation of the 17 LV muscle segments' wall motion and segmental and global LV strain calculation from the three apical LV views.

Using novel image processing algorithms, together with machine learning and deep learning technologies, DiA is the market leader in ultrasound AI.

www.youtube.com/watch?time_continue=6&v=_fPHWr6SSTE

www.dia-analysis.com

Digisonics Showcases Radiology Reporting Workflow Solutions at *RSNA 2018*

Digisonics, a leading provider of clinical image management and structured reporting systems for over 40 years, showcased their solutions to streamline radiology reporting workflows, including integration with PowerScribe 360, at last year's annual *RSNA* Conference in Chicago, IL.

The Digisonics Solution transforms the radiology workflow into a streamlined, fully automated electronic process, from data capture during the imaging exam to the fast creation of a structured report, through distribution to the patient's medical record. Radiology workflow inefficiencies and manual entry errors are eliminated, improving the overall quality of patient care. Users can be confident that all patient care documentation is captured in an accurate, consistent format, satisfying the requirements for reimbursement and accreditation.

Bi-directional integration of Digisonics with PowerScribe 360 offers further workflow efficiency. Users have the option of either sending the PowerScribe dictated report impression directly to Digisonics or auto exporting of the Digisonics sonographer worksheet to PowerScribe. Both options allow clinicians to retain their comfort with dictation while automating workflows and eliminating manual steps where possible.

Seamless integration with PACS for automated export of the sonographer worksheet or finalized report eliminates manual procedures to print, scan and upload to PACS, saving valuable time that can be better spent on direct patient care.

www.digisonics.com

Image Diagnostics' Featured Disruptive Products

Image Diagnostics Inc., a leading manufacturer of specialized diagnostic imaging equipment, showcased products at this year's *RSNA* that are expected

to radically improve occupational health and efficiency for interventional surgery. Two featured products located at the Image Diagnostics booth were the Protego Radiology Shield, currently in development, and the ilex55, available now for purchase. Key products featured at the Image Diagnostics booth during *RSNA 2018*:

Protego Radiology Shield

Image Diagnostics recently entered into a product development agreement with ECLS, Inc., a company founded by James Goldstein, MD, to design a groundbreaking radiation shield aiming for apron-free imaging that is now in development and undergoing testing. Dr. Goldstein, a cardiologist at Royal Oak, Michigan's Beaumont Hospital, has been an innovator in his field for over 35 years. The radiation shield is expected to mark a new era for the safety of interventional suite personnel, addressing the adverse health effects of radiation exposure, including orthopedic injuries caused by long-term wear of protective lead, along with cataracts and cancers.

"Our system currently in development is designed with a radiation shielding apparatus to facilitate performance of fluoroscopic catheterization procedures with unprecedented radiation reduction," said Dr. James Goldstein. "Decreasing the risk of cancer and cataracts and eliminating the need of wearing protective lead that often causes orthopedic burdens, we expect it to cause a paradigm shift in the occupational health of interventionalists."

Ilex55

Also at the Image Diagnostic's RSNA booth was the world's first large field 4k mobile and multi-modality monitor system, ilex55. This product provides revolutionary surgical imaging capabilities in a mobile platform for hospitals and surgery centers. Its breakthrough technology optimizes clinical images during GI, general, orthopedic, spine and vascular, and hybrid surgery. It can be rolled into the OR when needed and easily moved from room to room.
"Image Diagnostics Inc. introduced portable video integration in 2013 with the MDS." said Mark Hansen, Image Diagnostics' Vice President of Product Development. "The ilex55 represents our commitment to the evolution of lower cost, high value video integration solutions. The platform displays multi-modality images from a wide variety of sources in a single, side by side or quad view. Providing state of the art imaging capabilities, maximum flexibility and economic advantages, ilex55 is a trusted alternative to fixed equipment/ monitor booms or high-cost hybrid OR equipment."

A large part of what distinguishes Ilex55 from other options are its features, designed for optimal imaging capabilities, ergonomic flexibility and functionality. Below are features and advantages of the innovative mobile monitor system:

Ilex55 Imaging Features

- 55" UHD 4K monitor on an easy-to-move mobile platform with 20" vertical travel
- Displays one 4K image, two side-by-side images or up to four 26" HD images at one time
- Remote control screen layout and sourcing selection
- Calibrated surgical color
- Integrated speaker and USB charging station
- Displays analog and digital signals from multiple sources including: C-Arm, ultrasound, echocardiogram, hemodynamics, cameras, image guidance, & PACS

Ilex55 Key 4K Ultra-High Definition Advantages

- *Expanded Color Pallet* - 4K systems have the capacity to produce more shades of color and greyscale with or without a 4K input source. Naturally, this expanded color pallet

allows higher accuracy and visibility than current HD technology.
- Dose Reduction - Ilex55 customers observe and state they will use the C-Arm Mag function less, as the image quality is improved and so much larger.
- Image Clarity – Ilex55 provides an increase in sharpness and clarity. More pixels and colors make a cleaner picture.

www.imagediagnostics.com

Bring Peace of Mind to Medical 3D Printing with Materialise Certified Solutions

Partnerships surrounding Mimics inPrint software deliver broad benefits for patient-specific care.

Materialise NV (Nasdaq: MTLS), a world-leading 3D printing solutions provider, is working with partners to remove uncertainty and confusion for hospitals and physicians who are incorporating 3D printing into their diagnostic and surgical planning processes.

Materialise has launched a certification program that allows printer manufacturers that partner with the company to have products tested and validated as being fully compatible with Materialise Mimics inPrint software. inPrint is the first and only software to gain clearance from the FDA to develop 3D printed anatomical models for diagnostic and surgical planning uses. The inPrint software is printer- and material-agnostic, allowing Materialise to develop partnerships and solutions to meet a range of hospital and clinician requirements.

Materialise is working to address potential challenges for health care providers by ensuring their 3D operations are fully compatible and able to meet quality standards for developing and printing patient-specific 3D anatomical models. As the backbone of the 3D printing industry, Materialise incorporates nearly three decades of 3D printing experience and expertise to help medical professionals deliver more personalized patient care.

Stratasys and Ultimaker are the first two 3D printing hardware partners to participate in this program and to have their products tested by Materialise to certify compatibility.

"These partnerships offer health care providers the benefits of our open and flexible 3D printing solutions, while eliminating potential compatibility challenges with third-party hardware providers," said Bryan Crutchfield, Vice President and General Manager of Materialise North America.

Materialise's testing certifies that, when used in combination with Mimics inPrint software and in compliance with relevant quality assurance standards and in accordance with manufacturer instructions, a printer is capable of printing models suitable for use in surgical planning and multidisciplinary communication.

www.materialise.com

Emory University Study Demonstrates that Educational Videos Significantly Improve Patient Consent Process and Save Time - Results Suggest That Patient-Facing Videos Could Help Decrease Physician Burnout

The School of Medicine at Emory University has published a study suggesting that educational videos significantly improve the patient consent process versus traditional face-to-face methods. The physicians who participated in the study also believed that they spent less time during the face-to-face portion of the consent process following video consent while still being able to answer all of the patient's questions and concerns. The time-saving element of the study is noteworthy, given that long workdays are a contributing factor to the rising physician burnout epidemic, which affects more than half of U.S. physicians.

The study at Emory University was performed on 80 patients in the Interventional Radiology Department using educational videos created by the Holvan Group, a San Luis Obispo-based company that enhances patient experience through advanced communication. The research was commissioned to address the fact that patients do not always achieve the ideal level of understanding during the consent process. For the initial phase of the study, the consent process was performed unaltered. The patients were then given a survey, which was graded on a scale of 1 to 10 about their knowledge of the procedure, including the benefits, risks and alternatives available – the essential elements of an informed consent. Following this, patients were shown educational videos containing relevant content about the procedure they were about to undergo, after which another survey was completed.

The study produced the following results:

- There was significantly improved understanding of the risks with video-aided consent (standard 7.54, video-aided, 8.36).
- There was significantly improved understanding of the benefits with video-aided consent (standard 7.43, video-aided 8.45).
- There was a trend towards improved understanding of the procedure category (7.42 v 8.07), approaching statistical significance.
- The alternatives category saw an increase in the mean, for 6.8 to 7.5, however this did not reach statistical significance.

Overall, it was found that video-aided consent showed promise in the consent process. Across all categories, there was an increase in understanding after initiation of the video consent process. There was an anecdotal perception among the physicians that there was also a significant decrease in time spent during the face-to-face portion of the consent process following video consent.

These results were echoed in a separate nine-month pilot of educational videos provided by The Holvan Group at Sierra Vista Regional Medical Center, where 97 percent of patients felt prepared for their procedure after watching the videos before speaking with a physician. Physicians piloting the videos also reported that the videos freed up their time, as patients arrived with most of the preliminary questions answered. This enabled the doctors to shorten their workdays and spend more time getting to know their patients before the procedure.

www.theholvangroup.com

Arterys Demonstrates Suite of AI-Powered, Cloud-based Medical Image Analysis Solutions at RSNA 2018

Arterys, the leader in intelligent, 100% web-based medical imaging software, demonstrated its wide-ranging suite of AI powered solutions that support fast, efficient and accurate analysis of medical images at *RSNA* 2018, November 25-30 in Chicago.

Driven by deep learning and cloud computation, the Arterys platform opens a new era for medical imaging and clinical diagnosis with the power of the internet to enhance clinician workflow, streamlining and speeding analysis of breast, heart, liver and lung images to deliver improved patient outcomes for key workflows.

"Arterys is taking medical imaging analysis to new heights with a powerful U.S. FDA-cleared system that makes it easier for clinicians to detect and diagnose tumors and heart problems," said Arterys CEO, Fabien Beckers. "We are excited to highlight our latest advancements and demonstrate multiple web-based solutions for the radiology community, and to sponsor the Machine Learning Showcase at *RSNA*."

Arterys provided demonstrations of its AI-powered, web-based solutions, including:

- *Arterys Cardio AIMR*, a program that combines the power of deep learning and cloud computing to automate analysis of cardiac MR images. By eliminating many tedious, manual tasks, Arterys Cardio AI enables clinicians to quickly and easily identify, determine treatment for and track heart problems. It is the first and only commercial solution to offer deep learning-based semi-quantitative perfusion and quantitative delayed enhancement analysis.
- *Arterys Viewer*, a program that is powered by AI to increase speed, efficiency and accuracy of reading medical images. Offering multi-modal support, including for MRI, CT, X-ray and ultrasound images, Arterys Viewer is designed to deliver the best patient outcome by enabling clinicians to share images, collaborate and consult via a shared work space.

The Arterys HIPAA- and EU GDPR-compliant solutions integrate with existing Electronic Health Records (EHRs), PACS and RIS environments and dictation solutions.

Arterys CEO Fabien Beckers, along with Michael Poon, MD, Northwell Health cardiologist, presented "The Potential of a Web Platform to Transform Medical Imaging with AI and Cloud Computation" in the RSNA Machine Learning Showcase.

www.arterys.com

FujiFilm Presents Comprehensive Synapse Imaging Information and Enterprise Imaging Portfolio at *RSNA* 2018

FUJIFILM Medical Systems USA, Inc. showcased its complete line of innovative Enterprise Imaging and Informatics solutions at the *Radiological Society of North America's (RSNA)* annual meeting held from November 25th-30th, 2018 at McCormick Place in Chicago, IL. Attendees had the opportunity to participate in hands-on demonstrations of Fujifilm's robust technologies including Synapse® 5 PACS, VNA, Enterprise Information System, Cardiology, 3D, and Enterprise Viewer. In addition to presenting its comprehensive enterprise imaging product portfolio, Fujifilm highlighted its Cloud Services offering and featured an in-booth Artificial Intelligence (AI) lab to demonstrate REiLi, Fujifilm's innovative AI platform in development within Synapse 5 PACS.

"Fujifilm's Medical Informatics and Enterprise Imaging solutions are designed to transform radiology workflow and ultimately, patient care," said Johann Fernando, PhD, Chief Operating Officer of Fujifilm Medical Systems USA, Inc. "Our best in breed Enterprise Imaging solutions drive operational efficiency across and beyond the healthcare institution—ensuring true interoperability and the secure accessibility of patient records which spurs collaborative care."

The following Fujifilm technologies were showcased:

- *Synapse 5 PACS*— Designed with speed and functionality in mind, Synapse 5 PACS is a next-generation, secure server-side technology that enables instant access of massive data sets, working within the most popular browsers while using less bandwidth throughout the enterprise. Synapse 5 PACS will include: Synapse Communications which features an enhanced suite of tools including a native chat, emergency department findings, results escalation and tracking, and radiologist peer review—all of which help create a tailored workflow and a closed loop of communication between the radiology department and enterprise clinical departments.
- *Synapse VNA*— At the core of Fujifilm's comprehensive Enterprise Imaging portfolio, the TeraMedica division of Fujifilm's patented Synapse VNA is an industry leader in the vendor-neutral archive market by integrating more specialties, more devices, and more data than any other VNA. Through industry standards, healthcare organizations can achieve cost savings, impact clinical outcomes, optimize workflow efficiencies, securely manage all departmental data and experience true imaging interoperability.
- *Synapse EIS*— Synapse Enterprise Information System (EIS), is one of the most comprehensive radiology information systems and informatics workflow managers on the market. Synapse EIS enables providers to engage with their patients, improving quality of patient care and accuracy while also reducing costs.
- *Synapse Cardiology*— Making its RSNA debut on the latest Synapse platform, this next-generation, secure server-side rendering technology enables instant access of massive cardiology data sets. Synapse Cardiology Viewer works within the most popular browsers while using less bandwidth throughout the enterprise. Developed with ongoing direction from cardiologists, Synapse Cardiology offers capabilities and tools that help streamline image review and reporting across cardiac catheterization, ECG management, echocardiography, nuclear cardiology, 3D imaging and vascular ultrasound workflows.
- *Synapse 3D*— Designed for use across multiple specialties including radiology, cardiology, surgery and more, Synapse 3D is an enterprise-wide solution for quickly performing advanced visualization workflows and accessing 3D rendered images. Full integration with Synapse 5 PACS and Synapse Cardiology means seamless access from any Synapse diagnostic viewer.
- *Synapse Cloud Services*— A scalable environment shaped to host and manage the Synapse Enterprise Imaging portfolio. Critical-access hospitals, imaging centers, and teleradiology providers can take advantage of Fujifilm's cloud-based infrastructure and interoperability of software to maximize efficiency and cost savings.

"With Fujifilm healthcare IT solutions on their side, today's radiologists are better equipped than ever before to achieve diagnostic accuracy, operational efficiency, and improved patient care", said Bill Lacy, Vice President of Medical Informatics at FUJIFILM Medical Systems, USA, Inc. He invited attendees to visit the Fujifilm Medical Systems booth to see for themselves how the company's robust enterprise imaging technologies can help them thrive in today's value-based, collaborative care environment.

www.fujifilmusa.com/products/medical/

Canon Medical Debuts All-New Alphenix 4D CT at RNSA 2018

Innovative New Combination Pairs Alphenix Sky + C-arm with Aquilion ONE / GENESIS Edition CT

Canon Medical Systems USA, Inc. introduces a new, innovative angiography configuration featuring its Alphenix Sky + C-arm and Hybrid Catheterization Tilt/Cradle Table for interventional procedures with its innovative Aquilion™ ONE / GENESIS Edition CT system. The new pairing, called the Alphenix 4D CT, allows clinicians to efficiently plan, treat and verify in a single clinical setting. The flexible hybrid system enables streamlined workflow and outstanding range of patient access and coverage.

Benefits include:

- DoseRite technology designed to help clinicians minimize patient X-ray exposure while maintaining optimum image quality.
- Outstanding patient access, enabling clinicians to move the system, not the patient, and utilizing the new tilting cradle table.
- Greater control for the clinician with the tableside Alphenix Tablet to deliver a fast, seamless and rich workflow experience (optional).
- Unprecedented flexibility with innovative C-arm flip, right or left lateral flexibility, speed, and full body 3D imaging capability.
- Boosted productivity with the new Alphenix Workstation which integrates applications to help clinicians plan, analyze and perform interventional procedures.
- The new 4D CT configuration is part of the new family of Alphenix angiography systems which delivers images with clarity and precision. The new family combines industry-leading dose optimization technologies, enhanced workflow and innovative features to help clinicians provide patients with safe and fast treatment.

"The all new Alphenix 4D CT was designed based on the feedback we've received from clinicians. With the Hybrid Catheterization Tilt/Cradle Table and workflow enhancements enabled by the new Alphenix Workstation, we are confident this new system delivers on our customers' unmet needs," said Casey Waldo, director, Vascular Business Unit, Canon Medical Systems USA, Inc. "The new system expands the capabilities of 4D CT even further and creates new opportunities for innovation through intervention."

www.us.medical.canon

Corrections from the January 2019 Issue

In the article "A Congenital Heart Disease Patient 'Dreams Big' and Writes 'If My Heart Could Talk & Other Poems'" on p. 16, the caption under the first picture incorrectly identified the woman as David's mother. The caption is corrected below and in the electronic issues.

David with his ICU nurse after his first surgery.

In the article "Preview of NeoHeart – Cardiovascular Management of the Neonate, March 27th to 29th, 2019, at Hyatt Regency Hotel, Huntington Beach, California" on p. 19 the last name of the first author, John P. Cleary, MD, was misspelled in the author box and in his email. The correct email is jcleary@choc.org. This has been corrected in the electronic issues.

CCT apologizes for the mistakes and for any confusion they may have caused.

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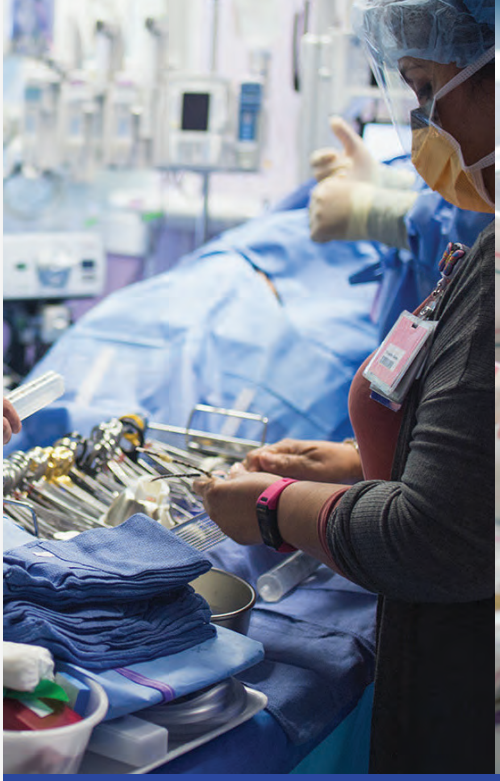

NeoHeart

Cardiovascular Management of the Neonate

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