# CONGENITAL CARDIOLOGY TODAY

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

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North American Edition


IN THIS ISSUE



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**accscientificsession.acc.org/acc.aspx**

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Recruitment adds, pages:
4, 5, 7, 15, 18, 19, 20, 23


# Scimitar Syndrome in a 3-Month-Old Infant

*By Howard Weber, MD, FSCAI; Khushboo Parikh, MD*

**Introduction**

Scimitar Syndrome is a rare congenital cardiac anomaly characterized by partial or complete anomalous pulmonary venous drainage of the right lung to the Inferior Vena Cava (IVC), either above or below the diaphragm.[1, 2] It accounts for 3%–6% of all the partial anomalous pulmonary venous returns.[3] Scimitar Syndrome is frequently associated with hypoplasia of the right lung, dextroposition of the heart, hypoplasia or malformations of the right pulmonary artery, anomalous systemic supply of the lower lobe of the right lung from the abdominal aorta or its branches and pulmonary hypertension.[5]

Patients diagnosed before the first year of life were reported to have major associated cardiac defects and a worse prognosis.[2] Management of the scimitar vein with respect to timing and type of intervention remains controversial.[1]

We describe a case of 3-month-old infant with Scimitar Syndrome and pulmonary hypertension, diagnosed during the evaluation of a cardiac murmur in the setting of respiratory distress.

**Case Presentation**

A 3-month old previously healthy infant presented to an outside hospital with a cough, congestion and fussiness. She was noted to have saturations in the 80s in the Emergency Department. $O_2$ supplementation was offered and she was admitted to the hospital. Chest X-ray demonstrated an enlarged cardiothoracic silhouette and increased interstitial edema (Figure 1). During the hospitalization the patient was

***"Patients diagnosed before the first year of life were reported to have major associated cardiac defects and a worse prognosis.[1]"***

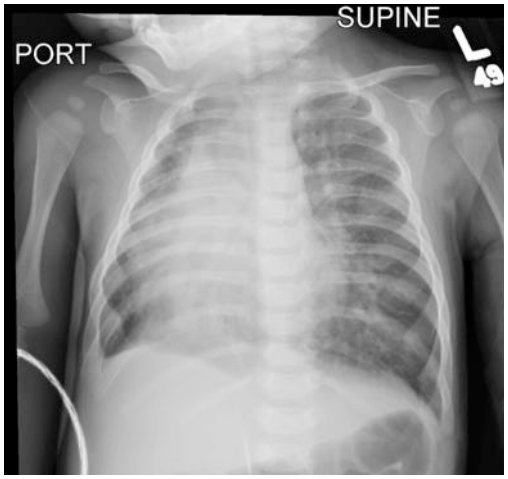
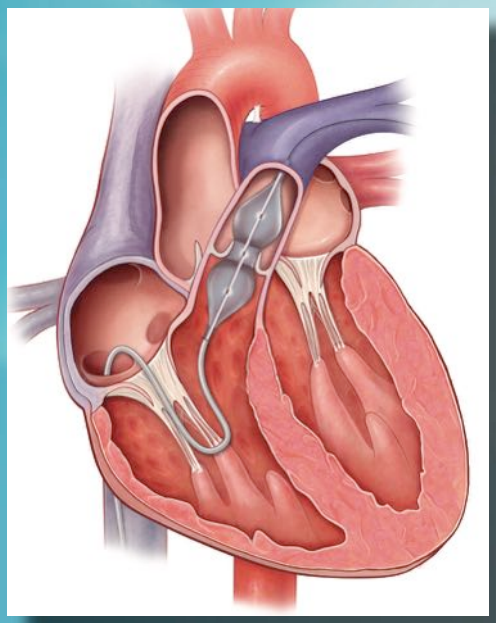



*Figure 1. Enlarged cardiomediastinal silhouette. Increased interstitial edema and subsegmental airspace opacification throughout the right lung.*



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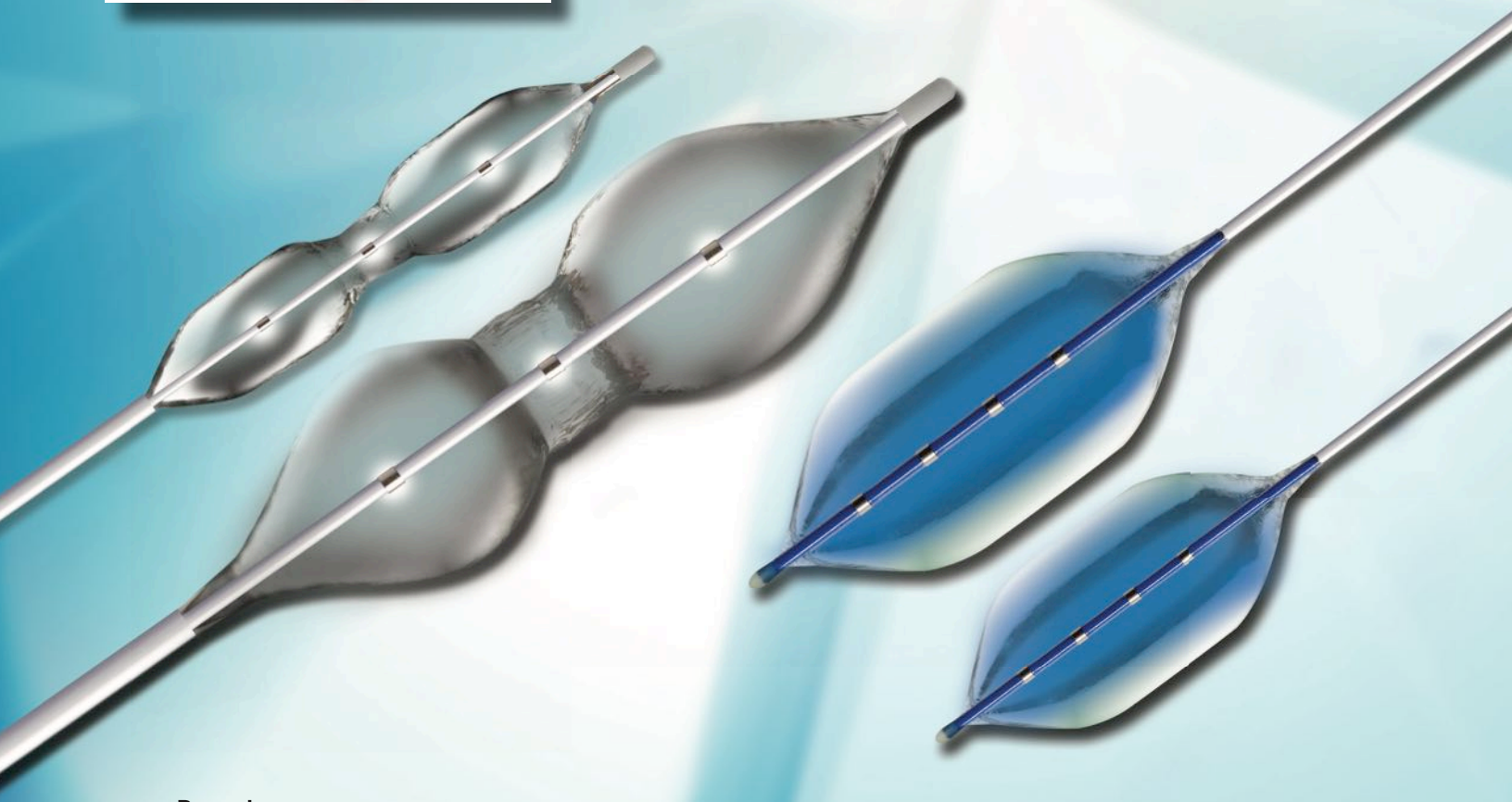

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noted to have a murmur warranting an echocardiogram. The study revealed a moderately-enlarged right atrium and right ventricle with suprasystemic right ventricular pressures. A moderate high secundum Atrial Septal Defect (ASD) with left-to-right shunting was noted. Only the left pulmonary veins were seen draining into the left atrium, and the right pulmonary veins were not well-defined. The decision was made to transfer the patient to a tertiary care center.

The following morning a cardiac CT scan was obtained to better delineate the underlying cardiac anatomy with special attention to the pulmonary veins. The CT scan showed complex right-partial anomalous pulmonary venous return, with two separate right pulmonary veins draining into IVC. An associated right aortopulmonary collateral arising from the celiac artery, and a mildly hypoplastic right pulmonary artery were noted. The scan also confirmed the echocardiographic finding of marked right atrial and ventricular enlargement. This confirmed the diagnosis of Scimitar Syndrome.

The patient was weaned off the oxygen and she remained stable on room air. Three days later the patient was taken to the catheterization lab. Angiograms performed in the catheterization lab revealed a 3.3 mm tortuous collateral arising from the abdominal aorta, which drained into the posterior aspect of the right lower lobe via two separate branches (Figures 2 and 3). The aortopulmonary collateral was coil embolized using a total of 3 Gianturco coils (one 4 mm diameter x 5 cm long, one 3 mm diameter x 4 cm long and one 2 mm diameter x 2.5 cm long). Repeat angiograms demonstrated complete occlusion of the aortopulmonary collateral.

Following a discussion with a cardiothoracic surgeon, and 3 days post-cardiac catherization, the patient underwent suture closures of the Atrial Septal Defect and tricuspid valve annuloplasty. Post-operative measured right ventricular pressures off cardiopulmonary bypass were 1/2 systemic.

Postoperative follow-up echocardiograms revealed moderately depressed RV function, moderate tricuspid regurgitation and systemic right ventricular pressures. The patient was placed on IV milrinone and supplemental oxygen with improvement of RV systolic function, although the pulmonary artery pressures remained at a systemic level. The patient was started on sildenafil and increased to 1 mg/kg q8hr; ultimately the pulmonary artery pressures decreased to 1/2 systemic prior to hospital discharge.

The patient was discharged home on 1 L $O_2$ supplementation and sildenafil with plans to repair the scimitar vein at a later date.

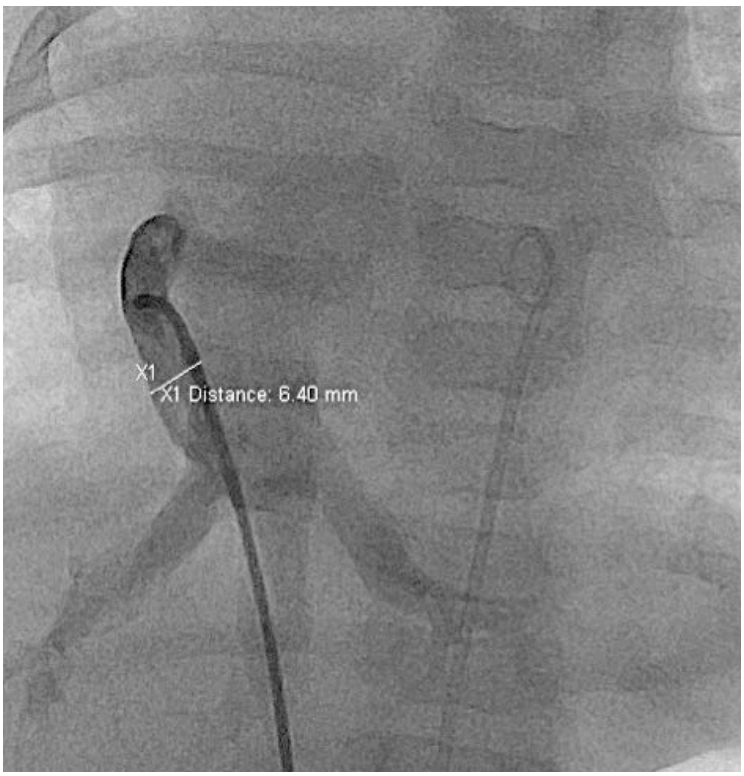


*Figure 2. Right pulmonary veins entering the IVC above the diaphragm.*

**Discussion**

In 1836 Chassinat and Cooper independently described anatomic features of a developmental complex in pathological specimens, which was later described as Scimitar Syndrome by Neill et al.[5] Dubuis and colleagues identified two forms of Scimitar Syndrome: the infantile form and adult form based on the age of presentation. The infantile form is associated with frequent symptoms, coexisting congenital heart defects and pulmonary artery hypertension and have a worse prognosis.[1] On the contrary, the adult form may present with recurrent pulmonary infections, dyspnea, or could be totally asymptomatic.[4]

Pulmonary artery hypertension, as noted in our patient, is identified as a risk factor for death in Scimitar Syndrome.[1, 3] It is attributed to multiple causes including: stenosis of the scimitar vein with pulmonary venous congestion; large aorta to right lung collaterals; right lung hypoplasia and pulmonary overcirculation.[1, 7]

In isolation, the anomalous venous drainage to the systemic venous atrium is unlikely to produce symptoms, and may even result in less RV volume overload when compared to other cases of partial anomalous pulmonary venous return. The hypoplastic right lung and right pulmonary artery may, in fact, limit the magnitude of the left-to-right shunt.[6]

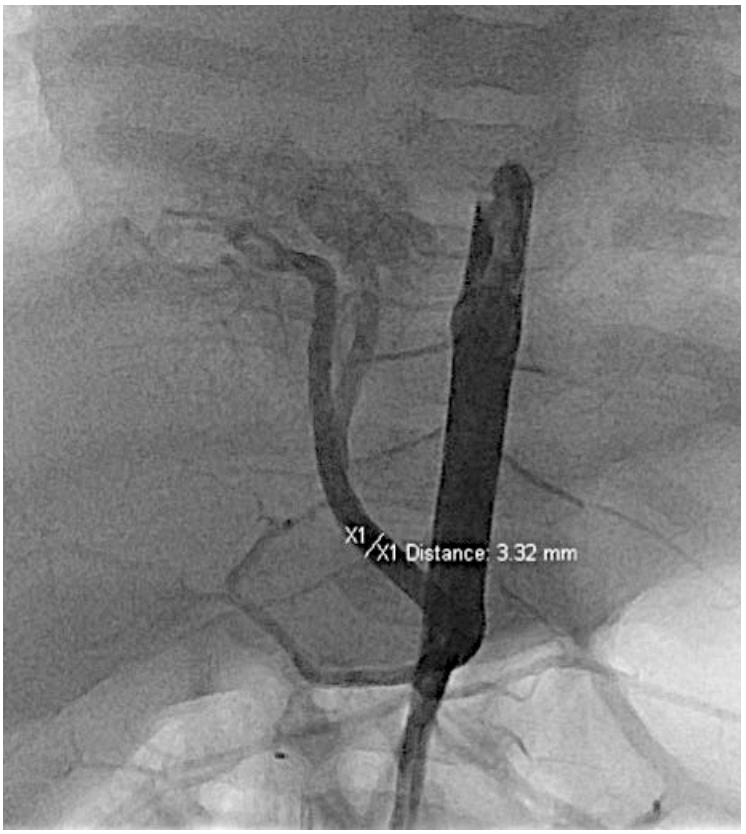


*Figure 3. Aortopulmonary collateral measuring 3.3 mm seen arising from the abdominal aorta and terminating in the lower lobe of the right lung.*

Haworth and colleagues proposed that the pulmonary hypertension was likely secondary to the failure of the pulmonary vasculature to adapt to the extra-uterine life in the presence of large pulmonary blood flow.[5] Dickinson et al and Levine at al demonstrated significant improvement in heart failure symptoms after ligation or coil embolization of the aortopulmonary collaterals.[6]

Definitive repair of the scimitar vein primarily involves redirecting the anomalous pulmonary venous drainage to the left atrium, either by baffling the anomalous drainage to the left atrium via a tunnel or by transecting the scimitar vein at its entrance to the IVC and reimplanting it to the left atrium.[3] Dusenbery and colleagues from Boston, however, reported high frequency of pulmonary vein stenosis in infants who underwent either the baffle or reimplanatation procedure.[1] This was supported by the European Congenital Heart Surgeons Association multicentric study that reported relatively high operative mortality and complication rate in patients who underwent repair before the age of 1 year when compared to patients who underwent repair at an older age.[3] Hence, they recommend conservative management in patients with no pulmonary hypertension, aortopulmonary collaterals, other intracardiac defects and only mild RV dilation. In patients with pulmonary arterial hypertension, aortopulmonary collaterals and intracardiac defects, a staged assessment plan was proposed. Initially, the aortopulmonary collaterals should be embolized, followed by repair of coexisting congenital heart defects, and reevaluation for any significant pulmonary artery hypertension or clinical symptoms.[1] This was the approach adopted in our case as well. We also instituted supplemental oxygen and Sildenafil for its potential pulmonary vasodilator effects, which appears to have improved based on serial echocardiograms.

## Conclusion

Scimitar Syndrome is a complex lesion, which comprises anomalies of the lung and the heart. The infantile form is particularly complex and repair of the anomalous vein in infancy has not been reported to be successful.[1, 2] Hence, our patient with anomalous right-pulmonary venous return, pulmonary arterial hypertension, aortopulmonary collateral and ASD only underwent coil embolization of the aortopulmonary collateral and surgical repair of the Atrial Septal Defect. She remains on sildenafil and $O_2$ supplementation for her pulmonary arterial hypertension with plans to definitely repair the scimitar vein at a later date.

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*"Scimitar Syndrome is a complex lesion, which comprises anomalies of the lung and the heart. The infantile form is particularly complex and repair of the anomalous vein in infancy has not been reported to be successful.[1, 2] Hence, our patient with anomalous right pulmonary venous return, pulmonary arterial hypertension, aortopulmonary collateral and ASD only underwent coil embolization of the aortopulmonary collateral and surgical repair of the Atrial Septal Defect."*


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**CCT**

**Brief Introduction of Principal Author**

Howard Weber, MD is a Professor of Pediatrics at the Penn State Hershey Children's Hospital and Director of the Pediatric Catheterization Lab. He completed his fellowship at Yale New Haven Hospital in 1989, and is a fellow of the American Academy of Pediatrics and the Society for Cardiac Angiography and Intervention.


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**Article Update - November 2015**

The article "Pattern of Grown-Up Congenital Heart Disease in Lagos, Nigeria" by By A.D. Olusegun-Joseph, MD; A.E. Okobi, MD; E.Y. Nkanor, MD; J.A. Mokwunyei, MD; F.E. Eto-Abasi, MD; A.C. Mbakwem, MD; J.N.A. Ajuluchukwu, MD (page 10-13) has been updated with an Addendum including a new table on Page 13).

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Fig. 5-IId

# Review of PICS~AICS 2015 in Las Vegas

*By Karim Diab, MD*

*The 19th Annual Pediatric and Adult Interventional Cardiac Symposium* (*PICS~AICS*) meeting was held in the "City of Lights" at the Aria Hotel, Las Vegas from September 18th-21st, 2015. With almost 850 attendees from sixty countries, the meeting continues to be relentlessly successful!

Thirty-nine percent of the attendees were from outside the US. One hundred- thirty-two faculty members gave over 120 talks; nine cardiac centers transmitted 20 live cases from North America, South America, Asia and the Middle East! There were 190 abstracts submitted this year to the meeting, the best ever!

The meeting featured a comprehensive program that covered various aspects of interventional therapies together with imaging modalities applied during interventional procedures in Congenital and Structural Heart Disease, from the neonate to the adult. This year's meeting focused on how to overcome greater complexity through collaboration with sessions that focused on outlining opportunities for greater integration with adult structural heart disease specialists and congenital surgeons. Hence, sessions discussed both congenital and structural interventions to try to learn from specialists in both fields. This included: discussing echocardiographic imaging done by congenital specialists during structural interventions, blood preservation, trans-septal techniques and antiplatelet therapy from the structural guys, review of various devices, as well as other topics.

Due to the increased focus on imaging modalities, and the importance of imaging for the success of any intervention, this time PICS dedicated a separate session for an imaging symposium preceding the meeting on Thursday that focused entirely on Innovations in Imaging for Congenital and Structural Heart Disease Interventions. Topics discussed included: the use of 3DRA and Computational Fluid Dynamics (CFD) in the cath lab, the use of intracardiac echo (ICE) and its advantages for imaging during interventions, the role of multimodality fusion and 3D Infinix Toshiba imaging technologies in congenital cardiac catheterizations and interventions, the use of the new Philips AlluraClarity system for radiation reduction, and the use of multimodality 3D image integration during congenital cardiac catheterization.

*PICS~AICS 2015* started officially on Friday and kicked off with live cases transmitted from major centers around the world. Unlike previous years, live cases started on the first day of the meeting! Live cases were beamed from nine national and international venues with experienced operators who demonstrated the latest in medical device technology using approved and investigational devices/valves/stents. The live cases this year were transmitted live via satellite from: Argentina, Saudi Arabia, Brazil, India, Las Vegas, Seattle, Columbus, Houston and San Diego.

Six cases were transmitted on the first day of the meeting. From Riyadh, Saudi Arabia, Drs. M Aljufan, F. AlFadley and M. AlAhmadi attempted a hybrid PPIV, and also performed a case of percutaneous Fontan completion in a SV patient. From São Paolo, Brazil, Drs. C. Pedra, S. Pedra and R. Costa performed closure of an interesting case of Aorto-Pulmonary Window using a 7mm Flex II ASD Occluder. They also performed a case of branch PA stenting and ASD closure using the Figulla Flex II ASD Occluder. From Cordoba, Argentina, Drs. Peirone and Ferrero and their team performed an ASD closure using the Nit-Occlud device in a 7-year-old, whose previously attempted ASD closure in 2012 had resulted in 1st degree heart blockage and was aborted. This time the closure went successfully with no complications, and the patient was in sinus rhythm. They also performed a case of PFO closure using the new pfm Nit Occlud PFO device.

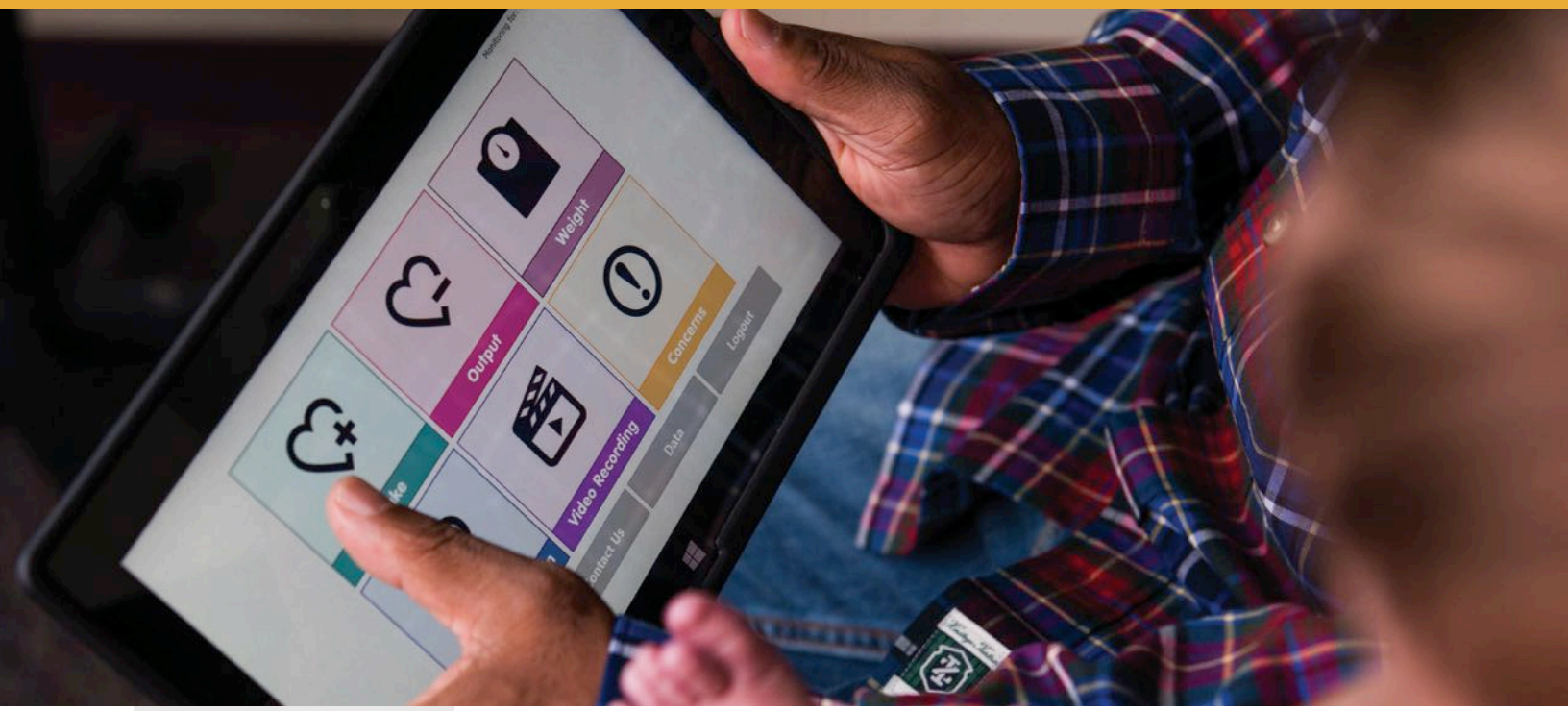
The popular Taped Case Sessions were presented during lunch with three taped cases discussed. Dr. B. Gordon from Loma Linda University, presented a case of pulmonary perforation in a case of pulmonary atresia with intact septum. Dr. B. Goldstein from Cincinnati Children's Hospital presented a case of branch PA stenosis in a 30-year-old adult with pa/ivs s/p Fontan, and Dr. J. Asnes from Yale University Medical Center, presented a case of a PVR and CAD in a 62-year-old with hx of TOF s/p repair and s/p ASD device closure.

The afternoon sessions on Friday were quite busy and included a session that focused on QA/QI issues for the cath lab and the new accreditation benchmarks for the cath lab that are evolving including review of the effect of IMPACT data, Multicenter QI initiatives and QA in developing countries. The highlight of the afternoon was the session focusing on the collaboration between specialists in both congenital and structural interventions. Dr. R. Hahn discussed the use of TEE, and what the congenital echocardiographers can show to the structural interventionalist. Dr. C. Kavinsky went over hemostasis devices, transseptal puncture and dual antiplatelet therapy. Dr. J. Aboulhosn discussed measuring shunts and flows. Dr. H. Sievert talked about the evolution of devices from PDA to VSD devices, and those for LAA and paravalvar leak, as well as TAVR valves. He highlighted improvements done throughout the years, as well as the still awaited improvements in many of the currently available devices, as there is always room for better technologies including: bioresorbable devices and better valves. Dr. F. Berger went over the use of stents and advanced angioplasty, and how it has altered the management of CHD. He talked about the risk of intimal hyperplasia followed by stenting, as well as the use of cutting balloons as a pretreatment approach, followed by stent implantation and the role of absorbable polymer stents in neonates. Dr. T. Jones discussed the evolution of interventional transcatheter valve techniques and the challenges in developing those revolutionary ideas and getting them into real practice.

*PICS/AICS 2015* also featured a break-out session held by the Pediatric/Congenital Interventional Cardiology Early-Career Society (PICES). PICES was established in July 2011, and is currently a subcommittee under the umbrella of the Congenital Heart Disease Council of SCAI. This group was created to support and advance the careers of young interventionalists in the fields of pediatric and adult congenital heart disease. The goals of the PICES group include:



Fig. 5-IId

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**—Girish S. Shirali, MBBS, FACC, FASE, Co-Director, the Ward Family Heart Center**

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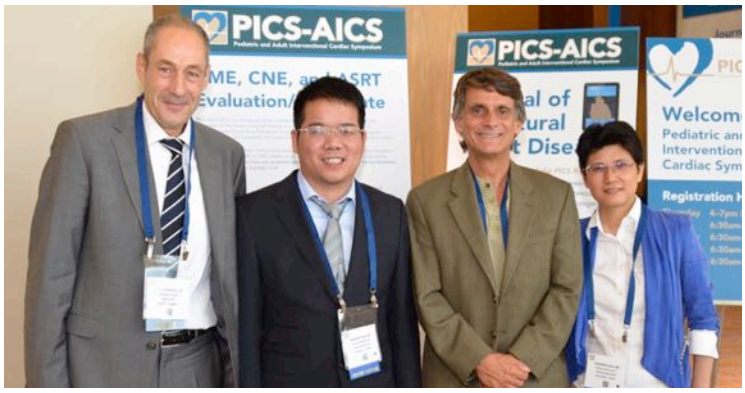
promoting clinical education and multi-center research collaboration, improving transcatheter treatment of Congenital Heart Disease in developing countries, and creating a professional network of young interventionalists and investigators. The PICES group currently has over 120 members with representatives from the United States and around the world. This year topics discussed included: holographic imaging in the cath lab, trachea-endotracheal stenting, and research updates.

Friday finished with the Oral Abstract Presentations with the best abstracts being presented on Sunday. The day ended with the





*PICS~AICS Achievement Award* honoring Dr. Joseph De.Giovanni. This award is designed to encourage and recognize investigators who have contributed exceptionally to the field of interventional cardiology in congenital and structural heart disease.

The Exhibit doors were opened in the evening together with the welcoming reception and attendees were able to browse more than 32 exhibitors showing the latest in medical technology.

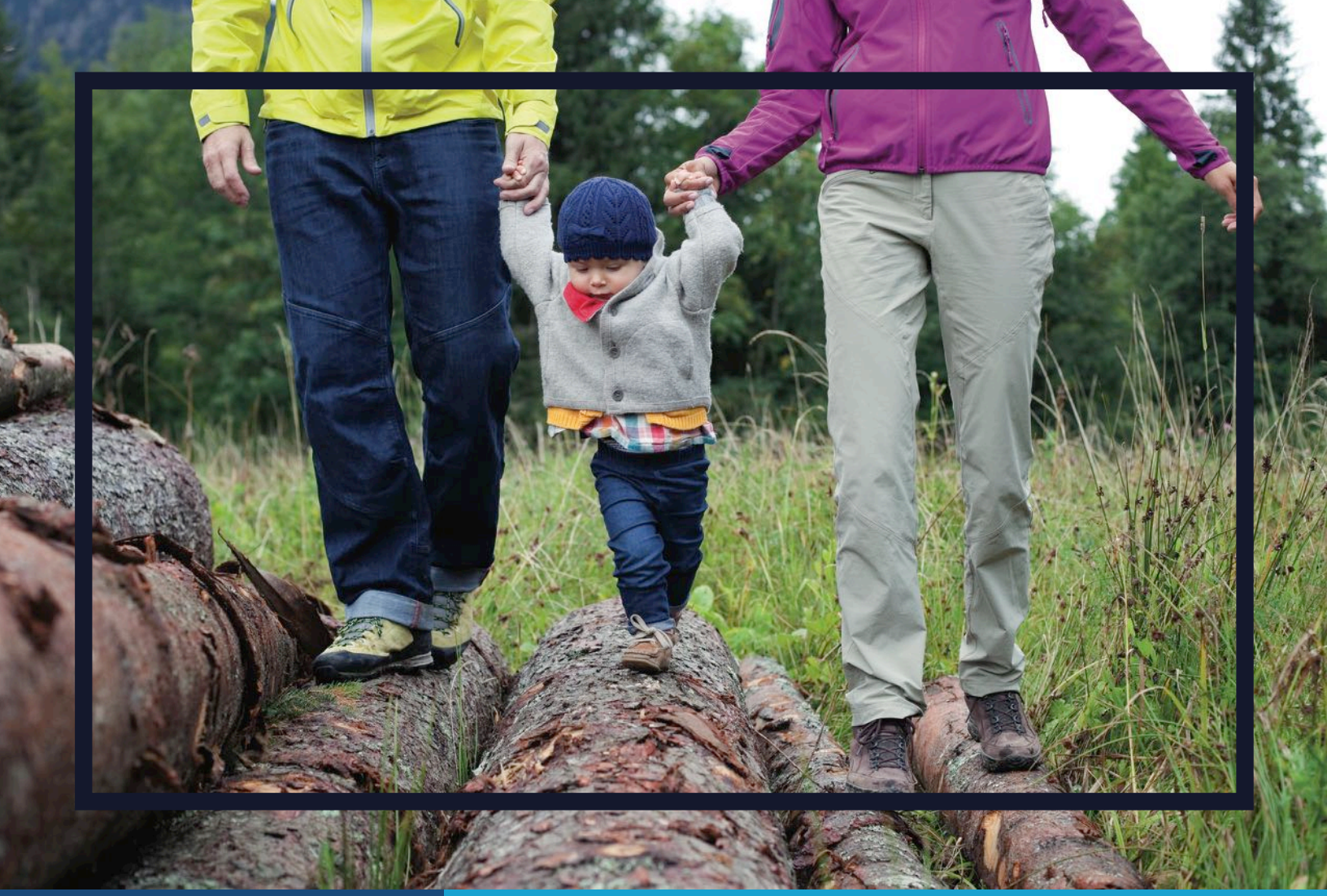
*PICS~AICS 2015* continued with a busy and informative program throughout the day Saturday. The morning started with Dr. Terry King talking about ASD closure, marking the 40th anniversary of this revolutionary idea and technique.

The live case transmissions then continued in the morning with another set of 6 cases. The live cases included:

- From Navati Hospital in Mumbai, India: Drs. B. Dalvi, P. Kerkar, R. Pinto performed a case of ASD closure using the Amplatzer Septal Occluder with Balloon Assisted Technique, and a case of closure of a ruptured sinus of Valsalva with Amplatzer Duct Occluder.
- From Children's Heart Center of Nevada in Las Vegas, Nevada: Drs. A .Galindo and A. Rothman, and joined by Dr. Z. Hijazi performed a case of percutaneous placement of an Edwards Sapien valve, and a case of closure of perimembranous VSD using the Amplatzer Duct Occluder II.
- From Rady Children's Hospital in San Diego, California: Drs. J. W. Moore and
- H. Al Said implanted a Melody Valve in a 15-year-old with DORV s/p Rastelli repair, and performed cases of occlusion of arterial feeders of AVM.

More taped cases were presented during the lunch break. These included a case of transfemoral TAVR presented by Dr. Mark Osten from Toronto General Hospital, a case of presented by Dr. Christopher Petit from Emory University, and a case by Dr. Frank Ing from the University of Southern California.

The afternoon session continued, with some of the highlighted breakout sessions at *PICS* including: the nursing and associated professionals breakout session and another joint session about the collaboration between interventionalists and surgeons in building a Hybrid program. There were discussions regarding several issues pertaining to Hybrid interventions. This included topics such as: myths and truths about CPB (Dr. Caputo), building the Hybrid room (Dr. M. Galantowicz), Hybrid interventions in the neonate with critical outflow tract obstruction (Dr. D. Berman), perventricular VSD closure (Dr. Z. Amin), exit angiography (Dr. R. Holzer), PVR (Drs. Ing and Philips), carotid cutdown for ductal stenting in the neonate (Dr. M. Alwi), perventricular closure of Perimembranous VSD (Dr. A. Omelchenko) and the Hybrid procedure for branch PAs and for HLHS (Drs. Ilbawi and Benson).



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The day also featured a session on Innovations in Structural Heart Disease. The session was divided into topics focusing on Mitral Valve/Left Atrial Appendage Disease and Tricuspid Valve Disease. Within the Mitral Valve/LA appendage session, different devices that are being tested were presented including: some preclinical devices, such as the Heart stitch, the Middle Peak Medical device used to replace the posterior leaflet, and the Mistral device. Dr. M. Reisman talked about the mitral valve apparatus, and showed beautiful cadaveric pictures from the wet lab. Dr. Z. Turi talked about the WATCHMAN device. As for Tricuspid Valve Disease, Dr. Riesman again kicked off the session with an anatomical lesson of the RV and the tricuspid valve using cadaveric images. Dr. J. Carroll then presented a brief case of Tricuspid Valve Disease in an adult patient. Dr. Hans Figulla talked about Heterotopic valve replacement for severe TR in patients who are not candidates for surgical intervention. Dr. Scott Lim then gave a talk about his institution's experience in percutaneous valve repair for chronic TR using the Mitralign device. Finally, Dr. D. McElhinney talked about the valve-in-valve tricuspid technology for failed bioprostheses.

The session also focused on Aortic Valve Disease with Dr. R. March talking about how to prevent catastrophic complications during TAVR, Dr. S. Bailey discussing emerging TAVR technologies, and Dr. C. Spies talking about predictors of early discharge after TAVR. There was also discussion of PFO closure in high-risk patients and percutaneous treatment of paravalvular leaks.

The second day ended with the "Meet the Experts" session, which allowed attendees to discuss cases with the experienced faculty in an interactive format.

Sunday early morning marked the Third Annual *PICS~AICS* 5km Run/1 Mile Walk co-sponsored by Siemens. This run/walk is to support the Congenital Heart Intervention Mission Support project that was launched during *PICS 2013*. The organization has been very active in providing a coordinated and sustainable benefit to interventional catheterization for structural heart disease in developing countries through centralizing and consolidating pre-existing charitable mission work. The work at CHIMS is far from imaginary and many steps have already taken place on the ground. Since CHIMS inception, over 20 centers have shipped equipment to a centralized repository in Memphis through a cost-free shipping service supported by the International Children's Heart Fund. To date, CHIMS has been able to support eight mission trips and over 50 catheterization procedures. This year, one hundred and six people signed up and over seventy participated in the run and Dr. R. Holzer was the winning champion!!

Sunday sessions at *PICS~AICS 2015* were marked by a day full of didactic lectures and updates in various areas of interventions for structural and congenital heart disease. The morning started with an update on clinical trials with discussion on Device Development and EFS by Dr. Nicole Ibrahim, Atrial Decompression for Left Heart Failure by Dr. Hans Figulla and PARCS and COAST II trials by Dr. Richard Ringel. The latter showed that covered stents are highly effective for relieving coarctation and repairing aortic wall injury with low incidence of serious complications (serious arterial access site injury <1%), and with a common reintervention for stent redilation (17%). Dr. Ringel also talked about the Pulmonary Artery Repair with Covered Stents (PARCS) study which showed that about 88% of patients with RVOT tears had homografts, and 43% developed tears during redilation. This was followed by an update on occlusion device studies such as: the St. Jude ASO, the GSO, NitOclud PDA device, the LAA devices, Mitraclip, Medtronic self-expanding PV and the


Advanta V12 covered stent. Dr. Zahid Amin talked about the SJM post-approval study that showed 0.3% erosion events with a 0.152 erosion incidence per 100 patient years. He also went over the ASO 522 study, the FDA-mandated erosion study to further evaluate the potential risk factors associated with erosion. He mentioned that currently, only 46 of the 450 sites in the US are activated and he encouraged operators to enroll. So far, no erosions have been reported by the study participants. Dr. John W. Moore then went over the results of the combined U.S. Multicenter Pivotal Study and the Continuing Access Study of the Nit-Occlud PDA device for percutaneous closure of Patent Ductus Arteriosus which showed that closure of small- and medium-sized PDA with the Nit-Occlud PDA is effective and safe when compared with objective performance criteria, with no deaths or significant serious adverse events, and a total adverse event rate of 4.7%.

The morning was marked by a keynote lecture given by Sir Magdi Yacoub during the session on the future in Congenital and Structural Interventions talking about "Innovations at What Price?"

Following up on innovative ideas, Dr. Yoav Dori talked about catheterizing the lymphatic system in complex CHD, which is barely studied. This included interventions including stenting the lymphatic system, and others, he mentioned building an interventional program for the lymphatic system. Dr. Dan Levi went over bioresorbable stents and occlusion devices, which are quite needed in the field. He mentioned the challenges in the field, and how advances will be driven by material combinations, creative designs and interest, initiatives and financial support. Dr. Massimo Caputo talked about advances in stem cell technology in the use for CHD. Dr. John Cheatham went over transcatheter Fontan completion and future advances still needed. Dr. Dietmar Shranz discussed the transcatheter systemicpulmonary shunt in PAH. He noted that TPS creation is feasible, and may offer symptomatic relief to select patients with refractory PAH. Dr. Thomas Jones talked about treating CHF in the cath lab, including eliminating shunts, fixing leaking valves and supporting pump failure. Dr. Elchanan Bruckheimer talked about 3D holography, and how it will shape interventions in the next 15 years.

The mid-day sessions focused on access options during catheterization, with techniques including carotid access in infants for interventional procedures, vessel rehabilitation, acute arterial ischemia post-catheterization, and simple modifications for successful Fontan fenestration. There was a debate about interventional procedures pre- and post- CPB, and whether there should be a time limit or not. Dr. Daniel Gruenstein argued that there should be no limit as to doing urgent interventions pre- or immediately post-surgical cath interventions, pointing out a few recently published studies. Dr. Zahid Amin argued the opposite, mentioning that there needs to be a barred time for doing


such interventions, as it was recently shown it can be associated with a higher rate of complications, as interventions are often at the suture line and cause stents to migrate.

The afternoon session focused on Current Controversies in Congenital Heart Interventions. This included topics such as: Palliation in Newborns With Tetralogy of Fallot: Infundibular vs. Ductal Stenting by Dr. JV DeGiovanni; Complex Aortic Arch Narrowing: Stent vs. Surgical Treatment by Dr. David Nykanen, and a hot debate on the risk of erosion after ASD closure (Pro: John Rhodes; Con: Joaquim Miro). Dr. Mark Fogel gave the last talk in this session about when we should close the Aorto-Pulmonary Collaterals.

The afternoon also featured the Spanish breakout session which covered fetal cardiac interventions, long-term outcome in PA/IVS and others. The last session of the afternoon focused on updates on Valve interventions in CHD, such as updates on the Melody Valve, risk for endocarditis, an update on "Next Generation" Native RVOT Valve Studies, valve-in-valve interventions, and the COMPASSION data.

The last day at PICS~AICS 2015 was busy as well! More live cases were transmitted from national and international sites all morning. The live cases that day included cases beamed from:

- Columbus, Ohio: Drs. D. Berman, A. Armstrong and C. Daniels performed a case of transcatheter PV implantation using the Melody Valve, transcatheter CardioEMS implantation
- Houston, Texas: Drs. H. Justino and A. Qureshi performed percutaneous splenic puncture and possible placement of device in portosystemic shunt, balloon dilatation and stenting of Pulmonary vein stent and a case of transcatheter VSD closure
- Seattle, Washington: Drs. T. Jones, A. Rubio, T. Johnston and B. Morray performed two cases of transcatheter PV implantation, one using a 3D reconstruction of RVOT conduit.

This was then followed by a special session that focused on: new interventions in extremely premature infants such as PDA device closure, Hybrid for HLHS in low birth weight newborns, special imaging during interventions in this high risk population, transcatheter treatment of the atretic RVOT in low birth weight newborns and CPB issues specific to premature and low birth weight newborns. The lunch session provided a competitive feel as the girls challenged the boys to a "Best Case." Other popular sessions included: "My Nightmare Case in the Cath Lab" also took place in the afternoon and helped ensure the opportunity for discussion and learning from each other's experiences. The final afternoon featured a session labeled "Battle of the Continents," a quiz-based session on all aspects of catheterization!

*PICS~AICS 2015* then ended with closing remarks by Dr. Z. Hijazi and the drawing of the passport winner!!

The next *PICS-CSI Asia* will be taking place in Dubai, UAE on March 3-5, 2016! Then, the next *PICS~AICS* will be back to Miami, Florida on January 16-19, 2017! See you in the "Magic Kingdom" for another amazing, educational and fun symposium! (For registration check the website at www.picsymposium.com.

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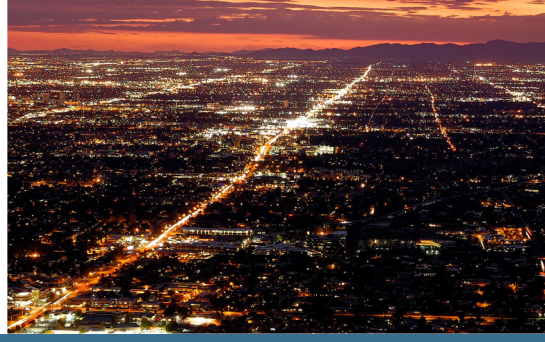

# Fetal Cardiology Opportunity

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Help grow our already large fetal heart program and collaborate with maternal-fetal medicine and neonatology practices in the Phoenix metro area. As a leader in fetal cardiology, our practice sponsors the Annual Phoenix Fetal Cardiology Symposium and is a participating site in multiple fetal cardiology multi-center trials.

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**5** fetal sonographers

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**5** office locations

More than **1,500** fetal echocardiograms performed last year

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Arizona Pediatric CARDIOLOGY

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# Forty Years of the Fontan Operation - Parma Meeting 2016 - Rastelli Lecture

*By Frank Cetta, Jr., MD*

In October of 2015 the group from the Mayo Clinic reported the long-term outcomes of 1,052 patients who had a Fontan operation. This publication covered the institution's entire experience dating from 1973 through 2012. Thirty-year survival was <50% for this cohort of patients However, in the recent era, 10-year survival improved to 95%. The Fontan operation was paradigm shifting in that it allowed for definitive palliation for children born with complex single ventricle physiology. It has resulted in a functional lifestyle into adulthood for many of these patients. However, despite the successes of the last 40 years, these patients are not "cured" or "repaired." At best, the Fontan operation should be considered "definitive palliation," and these patients demand lifelong, meticulous follow-up at centers equipped to handle their multifactorial medical problems. While it is encouraging that the more recent cohort has a favorable 10-year survival rate compared to those operated in the 1970s and 1980s, the long-term morbidity related to the Fontan operation is significant.

More than 50% of patients had rhythm issues that required medication at 20-year follow-up, and at least 20% needed pacemaker implantation. Fontan revision or conversion was needed in 10% and late repair/replacement of an atrioventricular valve was required in 5% of patients. Protein-losing enteropathy is one of the most devastating issues related to post-Fontan physiology. The mortality from PLE was 72%. Five-year survival after diagnosis of PLE for patients with Fontan physiology has been 50%. There are newer data that suggest with modern treatment regimes this may be improved, but when one looks at an overall cohort study, the long-term survival in patients after Fontan with PLE is poor. Cardiac transplantation in the modern era offers a treatment that may reverse PLE for these patients. A small minority of these patients also suffers with plastic bronchitis. There are new treatment strategies to identify lymphatic abnormalities that may be amenable to embolization strategies. Ten to twenty years after Fontan operation, issues related to liver pathology have been noted in these patients. Imaging techniques of the liver have improved over the years. However, the ability to treat post-Fontan hepatic fibrosis and cirrhosis is still being investigated.

In another analysis, patients who were referred to Mayo and had their initial Fontan elsewhere were also included. This brought the total cohort up to 1,138 patients. In this group, 195 patients had postoperative liver imaging or laboratory data available for analysis. In this subgroup of 195 patients, the freedom from cirrhosis at 10, 20 and 30 years was 99%, 94% and 57%. One-fifth of these patients were diagnosed with cirrhosis. The occurrence of cirrhosis was incremental. This is an interesting subset of patients.

Previous work from Mayo has demonstrated that patients with single ventricle physiology may actually have onset of Hepatic Disease prior to Fontan operation. Over the last 5-10 years, there has been increased surveillance of liver function and liver imaging in patients after Fontan operation. Some of this imaging has become quite sophisticated. Magnetic Resonance Elastography (MRE) in patients who are able to have MRI performed is an emerging technique for these patients. MRE evaluates hepatic stiffness, and helps to identify lesions that may be targets for biopsy. Some institutions have routinely performed liver biopsy on all patients at 10 years after Fontan operation. The proper diagnostic algorithm is still debatable. Even more dubious at this time is the proper treatment regime for hepatic disease in patients after Fontan. There is some suggestion that chronic anticoagulation with warfarin may be protective from developing cirrhosis in patients after Fontan. However, the data thus far are too limited to make this conclusion. The next decade of research will need to focus on improved therapy for this subset of patients. The vast majority of post-Fontan patients show some signs of increased hepatic stiffness or early fibrosis. Predicting which ones will progress to cirrhosis and even more importantly, hepatocellular carcinoma, is the question that will need to be answered.

Contraceptive practices and pregnancy outcomes in this cohort were also reviewed. There were 138 women with available contraceptive data; 44% used no contraception. The remainder used a variety of barrier methods, hormone therapies, sterilization and invasive devices. Interestingly, only 6 women suffered thrombotic complications and only one was utilizing oral contraceptives. 35 women had available pregnancy data. As has been shown in other studies of post-Fontan pregnancies, miscarriage rates were high (50%) and preterm deliveries were common. The mean gestational age of newborns born to women included in this study was 33 weeks and mean birth weight was only 2.1 kg. In select women after Fontan operation, pregnancy can be successful. The hemodynamic challenges posed by the placenta and possible placental insufficiency towards the end of pregnancy may contribute to preterm births and low birth weights. In this cohort, no viable pregnancies occurred in women with systemic oxygen saturations <90% or ejection fraction <40%.

***"At best, the Fontan operation should be considered "definitive palliation" and these patients demand lifelong, meticulous follow-up at centers equipped to handle their multifactorial medical problems."***

In summary, one has to wrestle with the fact that Fontan physiology is still a "man-made" form of chronic heart failure, and the sequelae of it can be substantial. More data from these studies and discussion will be presented during the *24th Parma International Echo Meeting*, Parma, Italy, May 28th, 2016. For meeting information, contact Professor Umberto Squarcia, MD- squarciaumberto@gmail.com or Professor Donald J Hagler, MD - hagler.donald@mayo.edu

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# Medical News, Products & Information

*Compiled and Reviewed by Tony Carlson, Senior Editor*

**New Findings on Embryonic Heart Valves May Prevent Congenital Heart Defects in Newborns**

Newswise - Cornell biomedical engineers have discovered natural triggers that could reduce the chance of life-threatening, congenital heart defects among newborn infants. Those triggers can override developmental, biological miscues, leading to proper embryonic heart and valve formation.

“The heart is the first organ to form in the embryo. It morphs dynamically and rapidly all the while pumping nutrients to the developing body,” said senior author Jonathan Butcher, associate professor at Cornell’s Nancy E. and Peter C. Meinig School of Biomedical Engineering.

The early embryonic heart originates as a looped tube, without valves or pumping chambers. During the last few weeks of the first trimester, these heart chambers form, but need something to maintain one-way blood flow.

“Wispy globular masses (called cushions because of their shape in the heart wall) need to condense and elongate to form thin robust leaflets capable of fast and resilient opening and closing,” Butcher said. “It is this maturation process that’s likely disrupted in many clinical cases.”

Until this study, scientists did not know how – or if – mechanical forces drove the biological remodeling of cushions into valves. Medical science understood that the embryonic heart needed blood flow to grow, but the valve component’s role was not entirely understood, Butcher said.

The researchers found that cyclic stretches and stressing forces activate sensitive enzymes called GTPases, specifically RhoA and Rac1, which coordinate the embryonic heart’s maturation. Without the enzymes activating at the proper times, heart valves do not form correctly.

“We identified a mechanism that transduced – or translated – a mechanical force into a biological response,” Butcher said. “That biological response over time creates these thin, flexible, formative leaflets. If this tissue fails to get thinner, that’s a problem. If the tissue fails to elongate, that’s a problem. And these are all problems we see in the clinic.”

This work lays a foundation for hemodynamically informed surgical interventions to potentially retard valve malformation – or to restore it, Butcher said.

The research was published in *Current Biology,* and supported by the National Science Foundation, the National Institutes of Health and the American Heart Association.

**Nurse Staffing and Work Environments Affect Survival after In-hospital Cardiac Arrest**

Patients with in-hospital cardiac arrest (IHCA) have low survival rates, but why do some hospitals achieve higher survival than others? Higher nurse staffing levels and better working conditions may be part of the answer, reports a study in the January issue of *Medical Care*, published by Wolters Kluwer.

"These results add to a large body of literature suggesting that outcomes are better when nurses have a more reasonable workload and work in good hospital work environments," according to the new research study, led by the University of Pennsylvania School of Nursing's Matthew McHugh, PhD, JD, MPH, RN, Associate Director, Center for Health Outcomes and Policy Research. "Improving nurse working conditions holds promise for improving survival following IHCA."

*Results Help Explain Hospital Variations in IHCA Survival:* Despite the chance of early intervention, less than one-fourth of patients with IHCA are discharged from the hospital alive. Some hospitals have been much more successful than others at improving survival after IHCA. Because nurses are the first link in the "Chain of Survival" in IHCA, factors related to nurse staffing might help to explain these variations.

Dr. McHugh and colleagues analyzed 2005-07 data on more than 11,000 adults with IHCA at 75 hospitals in four states, drawn from the American Heart Association's "Get With The Guidelines-Resuscitation" database. Data from national surveys of hospital characteristics and nurse staffing were used to analyze how these factors affected hospital survival rates after IHCA.

Only 15% of the patients with IHCA survived to hospital discharge. Most of the IHCAs occurred in an Intensive Care Unit (ICU), and 80% were witnessed. Eighty-eight percent of patients were on cardiac monitoring equipment when their cardiac arrest occurred.

Several factors affected the chances of survival--including whether the patient had a "shockable" heart rhythm that can potentially be reversed by an electric shock. Patients who were being monitored were also more likely to survive.

But even after taking these and other factors into account, hospitals with higher nurse staffing levels had higher IHCA survival rates. On general medical-surgical units, each additional patient per nurse was associated with a 5% relative reduction in the odds of survival.

In addition, the likelihood of survival was 16% lower at hospitals with poor work environments. That classification was based on a survey evaluating key areas for professional nursing practice, such as nurse participation, leadership, and support.

Nurse staffing levels in ICUs did not significantly affect the chances of survival after IHCA. That likely reflected limited variation in ICU staffing levels due to increasing standardization .

By comparison, staffing on general medical-survival units varies considerably between hospitals. Dr. McHugh and coauthors note, "Nearly half of IHCAs occur on medical-surgical units, which also have the most variable staffing levels and the most problematic work environments." The authors suspect that having too many patients to manage interferes with nurses' ability to effectively monitor patients closely, identify changes in patient condition, and intervene with lifesaving efforts quickly when seconds count.

The results add to a growing body of evidence that improving hospital work environments may be a promising approach to reducing preventable deaths--particularly after IHCA. "Adequate hospital nurse staffing may be an important strategy in efforts aimed at achieving excellent patient outcomes," the researchers write. But improving staffing may be difficult for some hospitals because of costs--nurses already account for more than 40% of direct care costs for hospitalized patients.

Dr. McHugh and colleagues believe that simply adding more nurses without considering the work environment may be a poor investment. They write, "Improvement of work environments...requires a change of inter-professional culture and extended delegation of care management to those care providers who are closest to patients."

Article: "Better Nurse Staffing and Nurse Work Environments Associated With Increased Survival of In-Hospital Cardiac Arrest Patients" (doi: 10.1097/MLR.0000000000000456)

### In Child Heart Patients, Gene Effects Overlap in Cardiac, Brain Development

Newswise — Some of the same gene mutations that cause heart defects in children also lead to neurodevelopmental delays, including learning disabilities. A large study of Congenital Heart Disease (CHD) reveals overlapping genetic influences during early childhood development.

"This research suggests that among infants with heart disease, we may be able to identify those at higher risk for neurodevelopmental problems, and to intervene earlier with therapy during a crucial stage of brain development," said study co-author and pediatric cardiologist Elizabeth Goldmuntz, MD, of The CHildren's Hospital of Philadelphia (CHOP). She added that follow-up research must be done before the findings could be used in early screening tests.

The collaborative federally sponsored study, with authors from Mt. Sinai Hospital, Yale, Harvard, and Columbia Universities, and other centers, appeared in the Dec. 4th issue of the journal *Science.*

Occurring in nearly 40,000 births each year, CHD is the most common birth defect in the U.S., and includes a variety of heart conditions with a broad range of severity. The new study analyzed DNA from over 1200 CHD patients, along with DNA from their parents. Some of the children had isolated heart disease, some also had neurodevelopmental delays, and a third group had CHD plus a birth defect in another organ.

Researchers have long known that a significant group of children with CHD have neurodevelopmental delays, often detected after they begin school. Some of these neurodevelopmental problems have been attributed to abnormal prenatal blood flow stemming from the original heart defect, and to the side effects from life-saving surgical and medical interventions during infancy.

However, said Goldmuntz, "over the years, all those non-genetic factors couldn't fully explain the neurological disabilities in these patients. This new study supports what many of us suspected—that some genes control both heart and brain development."

Goldmuntz, who has studied the genetic basis of CHD for the past 20 years, was the principal investigator at CHOP within the Pediatric Cardiac Genomics Consortium, established by the National Heart, Lung and Blood Institute. CHOP provided data for nearly one quarter of the children with CHD in this study.

The study team focused on "de novo" damaging mutations, those not found in the parents, but occurring first in a child. These mutations, found within the exome—the portion of DNA that carries codes for proteins—are predicted to alter protein function. The research found that these damaging de novo mutations occurred in 6% to 20% of children with CHD and other birth defects, but in only 2% of children with isolated CHD.

Moreover, said Goldmuntz, "this was not a random group of genes. They were genes that regulate gene transcription and chromatin remodeling, two biological processes that play key roles in early development by affecting whether other genes become active." Ultimately, she added, these genes act on biological pathways that might eventually be targeted with specific drugs, but such targeted therapies will require further research.

In all, the study team discovered 21 genes harboring protein-damaging mutations in more than one patient, and many other genes each in just one patient having a damaging mutation. Goldmuntz expects many more genes will be discovered that contribute to CHD, either in isolation or in combination with other developmental defects. In the meantime, said Goldmuntz, she and other cardiology researchers continue to face "the challenge of translation," applying genetic findings to clinical treatments.

The NIH provided grant support for this study. In addition to her CHOP position, Goldmuntz also is a Professor of Pediatrics in the Perelman School of Medicine at the University of Pennsylvania. For more information, visit www.chop.edu.

**EchoPixel Launches Brings Interactive Virtual Reality to Healthcare**

Marketwired - EchoPixel has introduced a new generation of medical visualization solutions that will transform the practice and study of healthcare. The company announced 510(k) FDA clearance of its True3D Viewer, the first platform to convert anatomical data from patients into fully interactive, three-dimensional virtual reality images. Early clinical trials with True3D have shown extraordinary results, with immediate impact. Across a range of procedures in surgery and radiology, preliminary results show that EchoPixel's technology can change the ways doctors work and dramatically improve patient outcomes.

Working hand-in-hand with top surgeons and radiologists across the country, EchoPixel has created a pioneering technology with clinically relevant applications. True3D enables a new world of patient care, allowing medical professionals and students to interact with patient-specific body parts in an open 3D space emanating from a desktop display. EchoPixel's True3D Viewer brings data from MRI scans, CT scans, and all DICOM imaging to life, letting doctors examine and interact with anatomical structures in an intuitive and non-invasive way. By presenting patient data in lifelike 3D, EchoPixel's technology has been shown to dramatically improve both speed and efficacy across a broad range of medical operations.

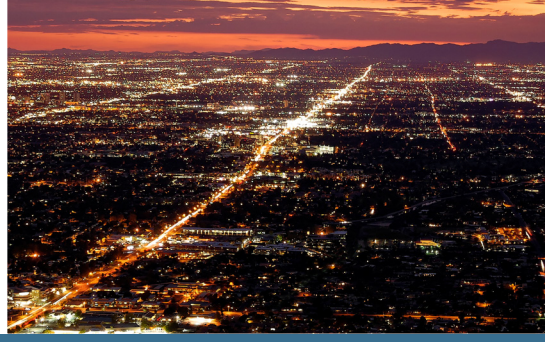
"Since CT scans were invented in the 1970s, doctors have learned about patient anatomy by mentally piecing together multiple images from flat screens," said Ron Schilling, CEO of EchoPixel. "That's not what the inside of a patient looks like. When working with doctors, we found they were wasting energy trying to solve imaging problems instead of clinical ones. Using virtual reality, we can provide an interactive, three-dimensional view of patient data that is far clearer and more realistic."

"We're taking virtual reality technology, the kind that's previously been restricted to entertainment, and applying it to medicine," said Sergio Aguirre, founder and CTO of EchoPixel. "This gives doctors a fully immersive, accurate representation of patient anatomy. The results we're seeing, from trials with real patient data, have confirmed the urgent need for this kind of imaging in medicine."

Dr. Frandics Chan, Associate Professor of Radiology at Stanford, has used EchoPixel in preliminary clinical surgical planning trials for children with pulmonary atresia (PA) with major aortopulmonary collateral arteries. Using True3D, detection sensitivity has increased, even as interpretation time has decreased. "Every patient is unique. I would previously have to verbally describe the path to the surgeon, but that's not adequate," said Dr. Chan. "EchoPixel presents an opportunity to see the world, and the pathology of the patient, as it is."

Beyond its immediate clinical impact, True3D technology promises a completely new standard for sharing medical data and expertise in the 21st century. With proprietary protocols, EchoPixel can formulate and distribute expert-derived methodologies for surgical and radiological procedures, in a richly annotated 3D format. This allows medical professionals to share information, collaborate, and leverage best practices for particular procedures. Moving forward, patients themselves will gain access to these images, and obtain a clearer and more realistic understanding of their own anatomy. For more information, www.echopixeltech.com.





# Opportunity for BC/BE Pediatric Cardiologist in Phoenix, Arizona

## About the practice:

- ✓ Collaborative effort of Arizona Pediatric Cardiology and Phoenix Children's Hospital
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- ✓ Large full-service practice with 26 pediatric cardiologists, 8 office locations and numerous outreach clinics throughout Arizona
- ✓ Services and programs include interventional and diagnostic catheterization, electrophysiology/ablations, cardiac imaging (MRI), fetal cardiology, adult congenital heart disease, anticoagulation program, interstage monitoring program, preventive cardiology, heart transplant/heart failure, pulmonary hypertension program, and a metabolic stress lab
- ✓ 35-year history in the community
- ✓ Covers 20+ hospitals

## About the location:

- ✶ Fifth largest city in the United States
- ✶ Averages 310 days of sunshine per year
- ✶ Variety of outdoor activities, as well as cultural and sporting events
- ✶ Family-oriented community with excellent school districts and unlimited activities for children
- ✶ Diverse geography offers recreational opportunities for desert, lake and mountain activities
- ✶ California coast within a day's drive

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# Siemens Introduces A New Generation of Flat Panel Detectors

*Allan Berthe, Contributing Editor*

*Congenital Cardiology Today* continues its series of articles dedicated to informing its readers about technology, products and techniques that aid in reducing radiation dose in the interventional catheterization suite.

Cardiac Interventionalists today are more aware of dose management, and are applying As Low As Reasonably Achievable (ALARA) and Image Gently (a campaign focused on practices to lower dose exposure to pediatric patients) techniques to their practice. Clinicians apply learned dose reduction fundamentals to manage dose, and create a safe environment for patient and staff. Today new technology is being added to systems that focuses on reducing the amount of radiation used, while producing an effective image, thereby, lowering radiation exposure to the patient and clinician.

Regarding technology, flat panel detectors (FPD) capable of fluorographic and radiographic imaging began to gain a foothold in the interventional system marketplace just over a decade ago. Gradually, users moved away from Image Intensifier systems and embraced FPD systems. Today, virtually all new fixed mounted C-arm systems sold are equipped with various sized Flat Panel Detectors.

The conversion to FPDs continued and system offerings expanded, including biplane systems that are routinely used while treating congenital patients. One of the leading organizations providing flat panel detector systems is Siemens Healthcare. They, along with other equipment manufacturers, have coupled FPDs with more system computing and imaging power, enhancing the clinician's tool box to aid with treatment planning, therapeutic intervention and dose management. Siemens recently introduced a new line of FPD systems, the Artis Q.zen which utilizes an enhanced imaging chain and new generation flat panel detector that continues to push new technology forward to improve image quality and dose management.


*Figure 1. Artis Q.zen Crystaline Silicon FPD designed to allow ultra-low dose imaging.*

Typically, FPDs are constructed in layers utilizing amorphous silicon, a thin-film transistor (T-F-T), a structured cesium iodide scintillator and a protective coating, When X-rays strikes the scintillator, visible light is emitted and converted to an electrical signal in the T-F-T array. Each pixel is then read out by on board amplifiers, which is converted to a digital value forming an image.

Siemens Artis Q.zen (www.siemens.com/artis-q-zen) created a unique FPD by changing one of main components, the amorphous silicon has been replaced by crystiline silicon. This change allows the electric signal to be amplified directly on the individual detector pixel. An electronic component generates electronic noise, the less distance a signal has to travel the less electronic noise is created. The Artis Q-zen on-pixel amplification cuts the distance between the signal generation and the amplification, thereby, reducing the noise.

Siemens made this FPD construction change considering the imaging needs of the congenital interventionalist and patient, to ultimately deliver a sharper image allowing the visibility of more detail in a wide variety of imaging conditions. The overall lower dose benefit comes in two forms:

- The system can be set at lower dose frames rates and deliver a better quality image as compared to previous generation systems/FPDs.
- If a clinician can see better, they should be able to work more confidently and efficiently, thereby reducing total procedure time. Reducing imaging time reduces dose.

*Figure 2. Artis Q.zen Biplane System*

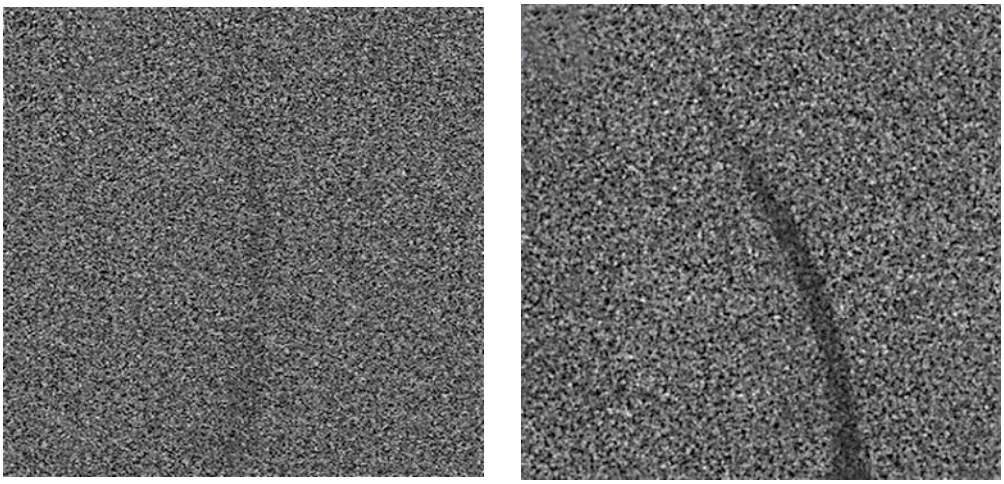

*Figure 3. Comparative clinical images of conventional (left) and Crystalline Silicon (right) flat panel detectors at the same acquisition dose of 6 nGy/frame. Notice the performance of the Crystalline Silicon FPD and the enhanced view of the Trans-septal needle.*

Any technology that permits a reduction in dose exposure to the congenital patient is appreciated by the clinician, as congenital patients may undergo a variety (from a variety of modalities utilizing ionizing radiation) of diagnostic and interventional procedures. Since children's bodies are more susceptible to radiation and they may face a lifetime of procedures and radiation exposure, the clinician's ability to better manage and reduce exposure is extremely beneficial.

**Seattle Children's Hospital Experience with the Artis Q.zen System**

Siemens is delivering Artis Q.zen systems worldwide to adult and congenital / pediatric hospitals. One of the early users of the system was Seattle Children's Hospital, which serves the Washington State area and beyond and performs hundreds of complex procedures per year on congenital patients from newborns to adults.

Seattle Children's interventional cardiologist Tom Jones, MD, Professor of Pediatrics and Medicine at the University of Catheterization Laboratories have been utilizing the Artis Q.zen Biplane system for about 18 months now.

The Seattle Children's Hospital team has collected data, and have noted a significant reduction in radiation dose as compared to previous generation systems. Lowering system dose output and ultimately exposure to the patient while maintaining appropriate image quality has made an impact.

Dr. Jones indicates, "Sometimes it just takes a while to get into some of these more difficult to reach locations; in the past, we would become increasingly nervous about exposure to the patient, as well as staff. Having low dose technology enables us to be that much more thorough in getting the job done."

Dr. Jones, nicely sums up their experience with the Artis Q.zen system. "In pediatric interventional catheterization, the efficiency afforded by improved image quality allows you to move much more quickly through procedures. With high-quality imaging, we can accomplish the same procedures with a whole lot less fanfare and less stress to the staff and the patients."

Clearly, clinicians and industry providers (such as Siemens), continue to make progress, and to focus on important areas, such as procedure efficiency and dose management technology, especially when it comes to pediatric patients.

**CCT**

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- ✓ Vacation and one weekend per month coverage provided by affiliated practice in San Antonio

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