

# CONGENITAL CARDIOLOGY TODAY

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## **CONGENITAL CARDIOLOGY TODAY**

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## **A Rare Fetal Diagnosis: Aortico-Left Ventricular Tunnel**

By Brie Ann Muller, MD; Karim A. Diab, MD

### **Introduction**

Aortico-Left Ventricular Tunnel (ALVT) is a rare congenital anomaly, occurring in less than 0.1-0.5% of patients with Congenital Heart Disease, and is usually diagnosed in the postnatal period.<sup>1,2</sup> It is comprised of a tunnel-like communication from the ascending aorta to the left ventricle. There have been reported cases of communication with the right ventricle, but these remain much less frequent. Associated coronary anomalies have occasionally been reported as well. Only a few cases of prenatal diagnoses have been previously reported, as this can be a challenging lesion to diagnose in the fetus. However, advancements in fetal imaging are likely to lead to more antenatal diagnosis of ALVT. Of the reported cases with a prenatal diagnosis, only a few have had a favorable outcome after surgery. We report a case of ALVT, that was diagnosed at 26 weeks of gestation and successfully managed surgically in the newborn period.

### **Case Presentation**

A 30 year-old, G3P2, female was referred to our center at 26 weeks of gestation for a fetal cardiac evaluation due to suspicion of a cardiac anomaly on routine screening ultrasound. A fetal echocardiogram revealed ALVT resulting in severe regurgitation and dilation of the left ventricle with increased wall thickness and mildly decreased systolic

function (Figure 1). Additional fetal echocardiogram findings included thickened and dysplastic aortic valve leaflets with mild aortic valve regurgitation and dilated ascending aorta with significant flow reversal across the aortic arch (Figure 2). There was cardiomegaly with a CT ratio of 0.6. The patient was monitored with serial echocardiograms with normalization of ventricular function and no evidence of hydrops, and was delivered at 38 weeks by normal vaginal delivery.

Postnatal echocardiogram confirmed the prenatal diagnosis of ALVT (Figure 3) with the additional suspicion of a coronary artery anomaly, as there was no flow visualized in the right coronary artery (RCA). Cardiac computed tomography angiography confirmed the findings with a tunnel seen from the aortic root, extending anteriorly to the aortic valve and inserting into the LV outflow tract. The left coronary artery origin was normal and the right coronary artery appeared to originate from the right aortic root adjacent and posterior to the insertion of the tunnel. The infant underwent surgical patch repair on Day of Life 3 with patch placement at the aortic end of the tunnel. The right coronary ostium, and RCA were not visualized intra-operatively, but this did not impact the repair. Post-operative course was complicated by a large pericardial effusion which was drained, and the patient was discharged home at 14 Days of Life. On serial follow-up visits, the patient was growing well, and an echocardiogram revealed initial mild

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residual patch leak at the aortic end of the tunnel that subsequently resolved at 5 months of age, and normal left ventricular systolic function.

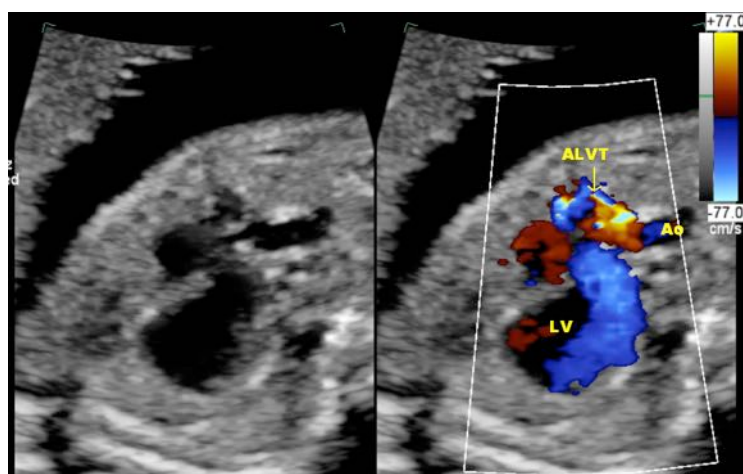


Figure 1. Fetal echocardiogram with 2D and color Doppler showing the Aortico-Left Ventricular Tunnel from the aortic root, extending anteriorly to the aortic valve and inserting into the LV; Ao = aorta; LV = left ventricle; RA = right atrium; ALVT = Aortico-Left Ventricular Tunnel.

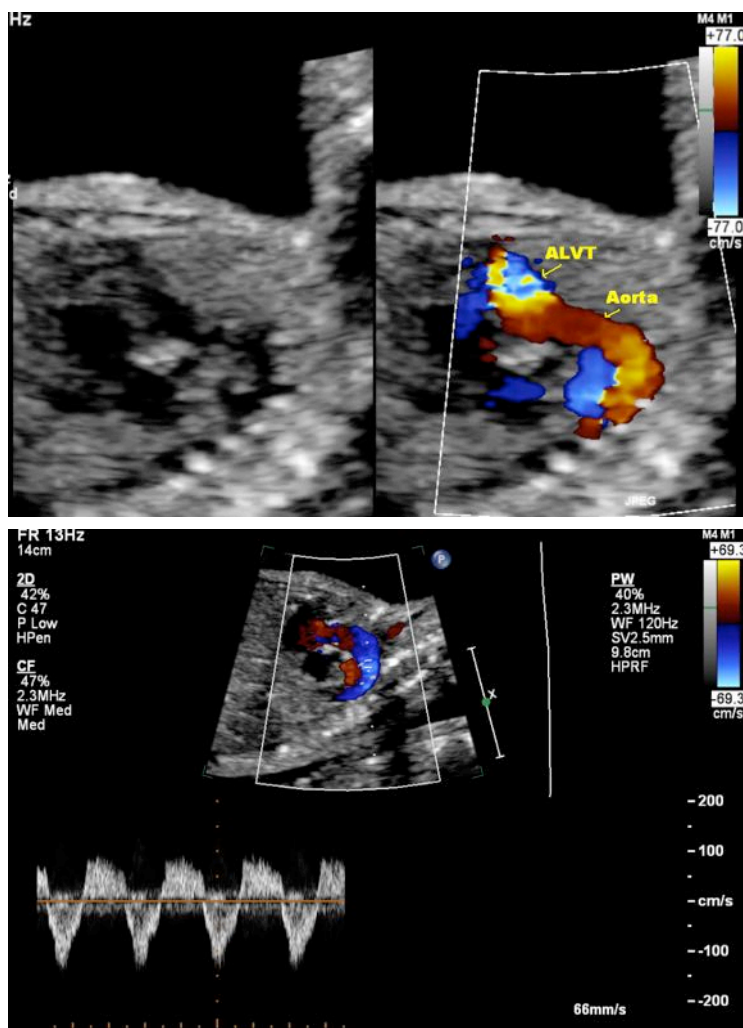


Figure 2. (A-Top) Fetal echocardiogram demonstrates dilated ascending aorta with retrograde flow into the ascending aorta on color Doppler, and (B-Bottom) by pulse Wave Doppler. ALVT = Aortico-Left Ventricular Tunnel.

## Discussion

Since Levy et al. first described ALVT in 1963<sup>3</sup>, approximately 130 cases have been reported; of those, fetal diagnoses occurred in less than 20%. Fetal diagnosis of ALVT was first reported in 1995<sup>4</sup> and appears to be on the rise in recent years, likely due to improvements in routine ultrasound screening. Historically, survival for patients with fetal diagnosis of ALVT has been lower than for those diagnosed in infancy or later in life. This was likely because those diagnosed in utero had severe aortic regurgitation, dilated left ventricles with poor function and hydrops, which prompted their fetal diagnosis. Whether or not survival is improving for those patients with fetal diagnosis of ALVT remains uncertain. In 1996, Sousa-Uva described 3 antenatal cases of ALVT, all of which died (1 was due to pregnancy termination).<sup>5</sup> In recent years, multiple cases have been reported with more favorable outcomes.<sup>6,7</sup> A more hopeful report by Singh et al. described 4 antenatal cases, 3 of which survived.<sup>8</sup>

Signs of ALVT by routine obstetric ultrasound include: dilated and/or dysfunctional left ventricle, enlarged cardiothoracic ratio and Doppler color flow mapping suggesting a regurgitant flow into the left ventricle. Other signs more likely detected by fetal echocardiography include: LV hypertrophy, reversal of

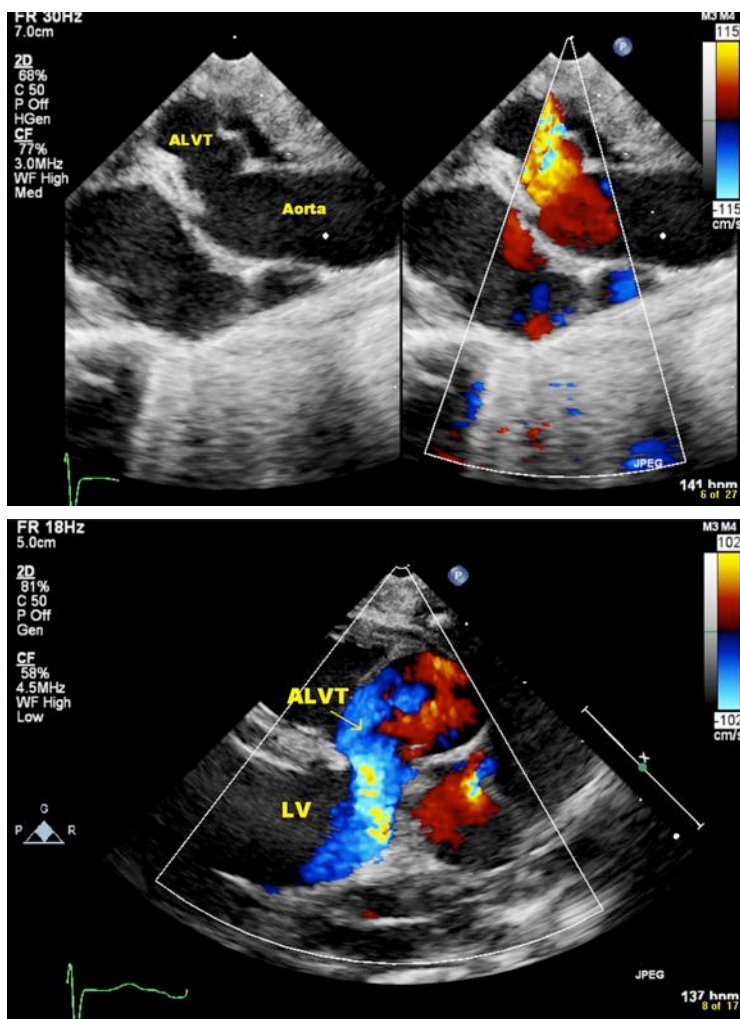


Figure 3. (A-Top) Postnatal transthoracic echocardiogram showing the parasternal long-axis view of the Aortico-Left Ventricular Tunnel extending from the aortic root, and (B-Bottom) inserting into the LV outflow tract.

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

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flow in the ascending and descending aorta, dilated ascending aorta, increased pulsatility of the ascending and descending aorta in combination with no or minimal regurgitation through the aortic valve itself. The presence of a “cockade sign” by 2D and color Doppler demonstrating the tunnel as a double ring structure around the aortic valve annulus can help suggest the diagnosis as well.<sup>7</sup>

Although fetal echocardiography will likely distinguish ALVT from isolated aortic valve regurgitation, a few other diagnoses may remain on the differential list -- mainly, ruptured sinus of Valsalva and coronary artery-LV fistula, in addition to an associated VSD. By postnatal echocardiogram, the differential diagnosis can be narrowed even further when the coronary artery anatomy, ventricular septum and ascending aorta can be studied in detail. ALVT can be distinguished from ruptured sinus of Valsalva by the more anterior and superior origin of the tunnel and the absence of a dilated sinus of Valsalva. Normal right and left coronary artery anatomy will eliminate the possibility of a coronary artery-LV fistula, although one must take into account the possibility of coronary artery abnormalities in association with ALVT. Cases have been reported of single left coronary artery and anomalous origin of the right coronary artery from the ALVT.<sup>2</sup> Therefore, precise assessment of the coronary anatomy must be done in those with suspected ALVT and additional imaging modalities may be necessary in these patients, specifically cardiac catheterization with angiography, cardiac CT or MRI. Lastly, the absence of left-to-right shunt across the LV septum will exclude an associated VSD.

Further complicating the diagnosis, aortic valve pathology is often seen with ALVT. Bicuspid aortic valve with varying degrees of aortic stenosis have been reported.<sup>9</sup> In one extreme case, the tunnel was mistaken for the LVOT and aortic valve due to aortic atresia.<sup>10</sup> More severe forms of aortic stenosis will lead to LV hypertrophy, myocardial fibrosis and LV dysfunction. These are likely the patients with ALVT who present with fetal hydrops and in utero demise.

The patient with signs of ALVT on fetal echocardiogram should be followed frequently after diagnosis due to the possibility of progressive LV dilation and subsequent dysfunction with the ultimate scenario of fetal hydrops. After delivery, the diagnosis should be confirmed by transthoracic echocardiogram, and any further imaging necessary, as mentioned above. Once complete evaluation of the cardiac anatomy has been performed and ALVT confirmed, repair is usually prompt, pending any other comorbidities.

Treatment for this lesion is primarily surgical, with patch or suture closure at either or both ends of the tunnel. Transcatheter device closure has been described for the initial closure of the defect, as well as for residual defects after surgical patch closure.<sup>11,12</sup> Survival has improved to nearly 100% following surgical closure.<sup>13</sup> The risk factors for increased mortality appear to be: size of the



## OPPORTUNITY IN PEDIATRIC CARDIOLOGY LAREDO, TEXAS

Driscoll Children's Hospital is advancing a comprehensive Heart Center to meet the healthcare needs of congenital heart patients in South Texas. The Center is recruiting a physician to support outpatient clinic activities in Laredo, TX. Sub-specialty board eligible or certification is required. Spanish speaking is preferred.

Pediatric Cardiology has been an integral part of Driscoll Children's Hospital since 1962. The Hospital and the Heart Center are committed to bringing state-of-the-art technology and quality service to 31 counties in South Texas. In 2013, the Heart Center saw 9,500 outpatient and satellite visits; 6,121 echocardiograms, including 500 fetal echos, and 192 heart catheterizations (82% interventional). The Laredo Clinic is a major clinic affiliated with Driscoll Children's Hospital. The Heart Center employs 8 physicians including 1 Electrophysiologist, 2 Interventional cardiologists, 1 MRI Imaging cardiologist, and 1 fetal cardiologist. Three pediatric cardio-thoracic surgeons deliver all aspects of surgical service including hybrid procedures.

The Laredo Clinic is a major clinic affiliated with Driscoll Children's Hospital. The cardiologist in Laredo will share a 1:2 call rotation. Physicians will see challenging, complex patients in a beautiful, well-staffed clinic with 2 sonographers. The qualified physician will enjoy a young, fast growing patient base.

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tunnel and resulting amount of regurgitation, concomitant aortic valve abnormalities, coronary artery anomalies and resultant myocardial ischemia and ventricular dysfunction.

In conclusion, ALVT is a surgically treatable cardiac lesion with good long-term results. Patients diagnosed in utero tend to have greater risk for early mortality. Prenatal diagnosis of ALVT is possible, and can aid in decisions regarding postnatal care and timely surgical treatment.

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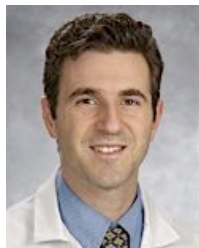
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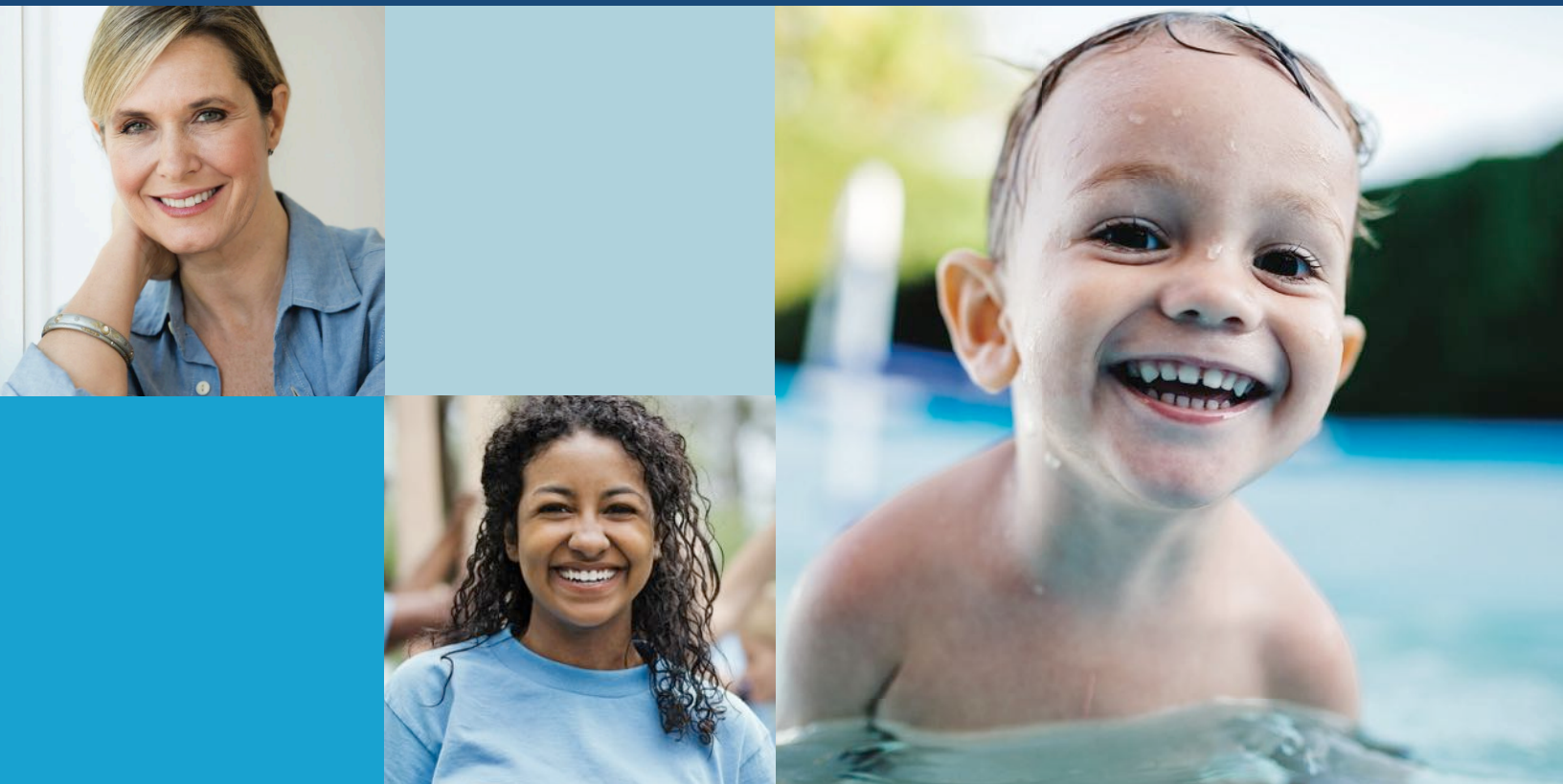
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# Pulmonary Arterial Hypertension with Atrial Septal Defect in a Newborn Baby with Goldenhar Syndrome

Preeti Srivastava, DNB; Asit Kumar Mishra, MD; Md Waseem Uddin, MD; Mrigendra Nath Tudu, MD

## Introduction

Goldenhar Syndrome (Oculo-Auriculo-Vertebral Syndrome - OAVS) with hemifacial microsomia is a rare congenital anomaly involving the first and second branchial arches.

It is a disorder where the patient's facial features are incompletely developed on one side, resulting in eye, ear, and jaw abnormalities. In 85% of patients with Goldenhar Syndrome, only one side of the face is affected. Cervical spine vertebral deformities are part of the collection of symptoms.<sup>1</sup>

The syndrome was first described in 1952 by the French ophthalmologist Maurice Goldenhar.<sup>2</sup> The incidence of Goldenhar Syndrome has been reported to be between 1:3500 and 1:5600, with a male:female ratio of 3:2(3). The exact etiology is not known. However, it is possible that abnormal embryonic vascular supply, disrupted mesodermal migration or

some other factor leads to defective formation of the branchial and vertebral systems.<sup>3,4</sup> Most of the cases have been sporadic. Autosomal dominant, autosomal recessive and multifactorial modes of inheritance have also been suggested.<sup>3</sup>

Ingestion of drugs such as thalidomide, retinoic acid, tamoxifen, and cocaine by the pregnant mother may be related to the development of this syndrome. Maternal diabetes, rubella, and influenza have also been suggested as etiologic factors.<sup>5</sup>

The classic features of this syndrome include: ocular changes such as microphthalmia, epibulbar dermoids, lipodermoids, and coloboma; aural features such as pre-auricular tragi, hearing loss, and microtia; and vertebral anomalies such as scoliosis, hemivertebrae, and cervical fusion.<sup>2,5</sup>

Systemic features are cardiac and renal malformations.<sup>6, 7</sup> Congenital heart defects have been reported in 5%–58% of patients with OAVS. Tetralogy of Fallot and Ventricular Septal Defects are the most common cardiovascular anomalies associated with OAVS.<sup>5</sup>

Here we report a rare association of Atrial Septal Defect with Pulmonary Arterial Hypertension in a newborn with Goldenhar Syndrome presenting with congestive cardiac failure.

## Case Report

A 5-day-old, male baby, weighing 2.4 kg was admitted with complaints of breathing difficulty and refusal to feed for 3



Figure 1.



Figure 2.



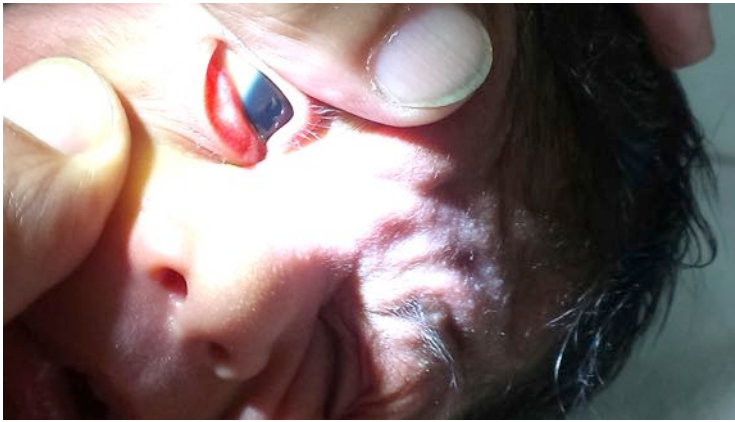


Figure 3.

days. The baby was born at 38 weeks gestational age to a 32 year old, gravida 3, para 2, abortion 1, mother by normal delivery. The baby had cried immediately after birth, and was started on breastfeeds soon after birth. Ante-natal history was insignificant.

On examination, the baby was having respiratory distress, mild icterus: head-to-foot examination revealed several anomalies in form of abnormal pinna of right ear (Figure 1), right mandibular hypoplasia, right-sided facial paralysis (Figure 2), bilateral coloboma of iris (Figure 3) and retinochoroidal coloboma. Systemic examination showed normal lungs, hyperdynamic precordium with normal heart sounds, abdomen was soft with palpable liver 4 cm below the subcostal line. A detailed work-up was done to rule out other anomalies. CECT brain scan showed posterior falx, tentorial and bitemporal minimal cortical hemorrhage (Figure 4). Echocardiography revealed 12 mm ASD, L-R shunt, 3 mm PDA with moderate PAH (Figure 5). USG abdomen was normal. Chest X-ray and OAE were normal. X-rays of thoracic and lumbar spine were normal.

Baby was managed with antibiotics, fluid, and oxygen. Cardiac decongestive measures were added after ECHO findings. Baby gradually stabilized, and was put on feeds. He was discharged after 7 days on decongestive measures, and advised to attend a cardiac centre.

## Discussion

Review of literature reveals that patients with Goldenhar Syndrome usually present in childhood, adolescence or adulthood with audiovisual or skeletal problems. They might present in infancy due to facial dysmorphism or complications arising due to Congenital Heart Disease. Frequency of cardiovascular malformations in this syndrome varies between 5-58%.<sup>8,9</sup> Cardiovascular developmental anomalies in Goldenhar Syndrome are



Figure 4.

usually severe, such as Tetralogy of Fallot, Ventricular Septal Defect, Wolf-Parkinson-White Syndrome and other vascular anomalies.<sup>8, 9</sup> In our case, the baby presented in early neonatal period (on Day 5) with signs of congestive heart failure. Echocardiography revealed a large ASD with small PDA and moderate PAH. The occurrence of features of CCF with the identified cardiac lesions could not be



**Barth Syndrome**  
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## Barth Syndrome (ICD-10: E78.71)

Symptoms:  
**Cardiomyopathy, Neutropenia, Muscle Weakness,  
 Exercise Intolerance, Growth Delay, Cardiolipin Abnormalities**

[www.barthsyndrome.org](http://www.barthsyndrome.org)



Name: B/O KAJAL

Ref. By:

Age:

Sex:

Id: 4889/14

Date: 22/10/2014

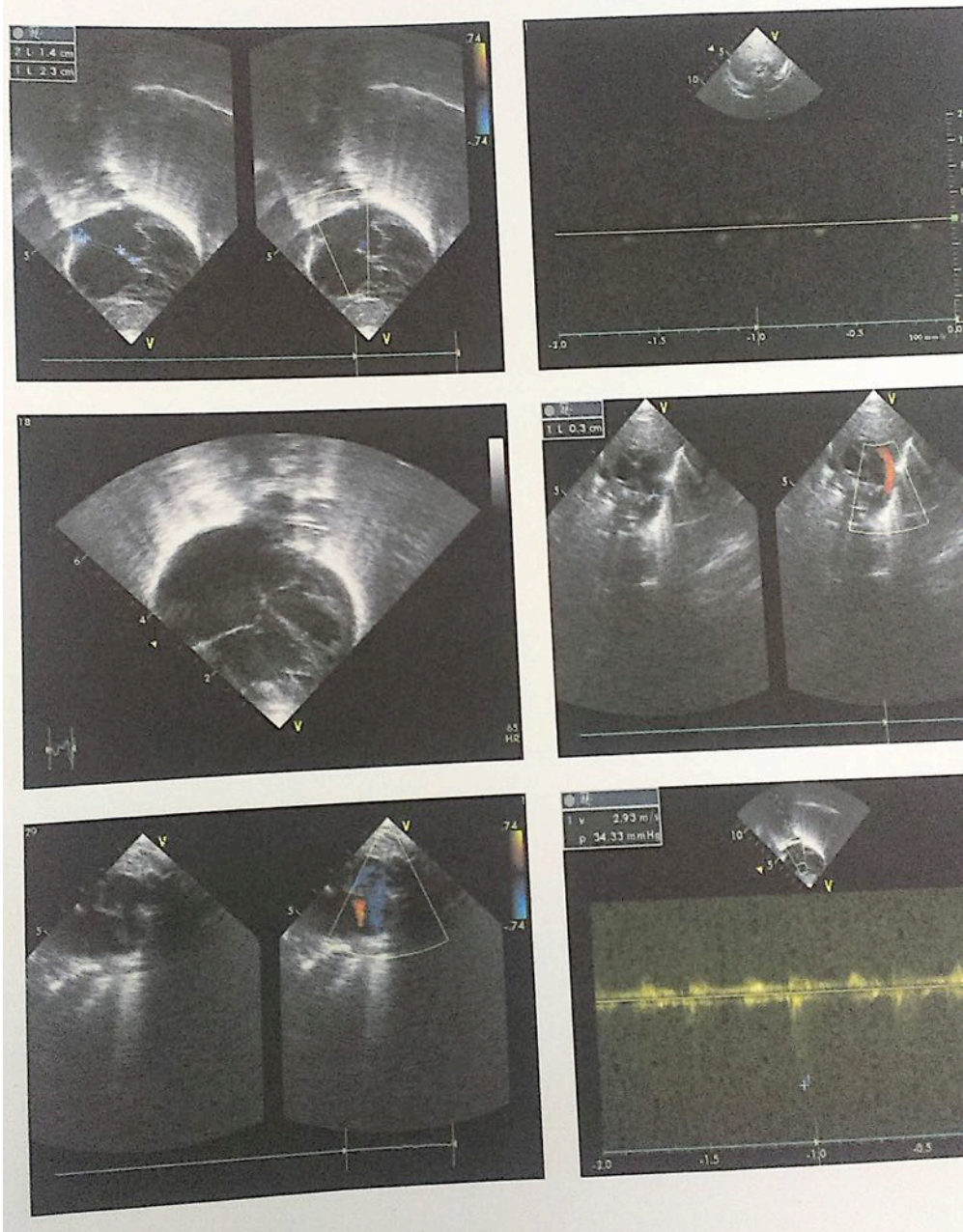


Figure 5.

explained and hence baby was referred to higher cardiac centre for further evaluation.

This case is being reported due to the rare presentation of Goldenhar Syndrome with ASD, PAH and CCF in the early neonatal period.

***“Review of literature reveals that patients with Goldenhar Syndrome usually present in childhood, adolescence or adulthood with audiovisual or skeletal problems. They might present in infancy due to facial dysmorphism or complications arising due to Congenital Heart Disease. Frequency of cardiovascular malformations in this syndrome varies between 5-58%.<sup>8,9</sup>”***

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**CCT**

#### Profile of Principal Author

Preeti Srivastava, DNB (Paediatrics)

Currently working as Registrar in Dept. of Paediatrics, Tata Main Hospital, Jamshedpur. The first author has had four and half years of post PG experience and is currently working in the neonatal unit for two and a half years. The author firmly believes in serving the paediatric community with competence and compassion and has a special interest in Pediatric Neurology subspecialty.

#### Principal Author



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Jamshedpur*

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Tata Main Hospital  
Jamshedpur*

*Mrigendra Nath Tudu, MD  
Dept. of Paediatrics  
Tata Main Hospital  
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**E-mail comments to:  
CCT-Mar17-GS@CCT.bz**

## BC/BE FT & PT GENERAL PEDIATRIC CARDIOLOGIST

The Pediatric Cardiology Center of Oregon is recruiting a BC/BE FT & PT GENERAL PEDIATRIC CARDIOLOGIST to join the private practice of four Pediatric Cardiologists and one PA located in Portland, OR.

The Pediatric Cardiology Center of Oregon works with Legacy's Randall Children's Hospital and their pediatric CV Surgery team and are associated with multiple hospital systems throughout the state of Oregon and Washington. We have a comprehensive team that includes a cardiac interventionist providing transcatheter valve replacement (Melody Valve), ASD and VSD devices, an EP specialist experienced in ablations and device implants, a large fetal program and a Cardiac CT/MRI program.

In addition to our main office in Portland, our physicians provide outreach to multiple sites across Oregon and SW Washington. The group and surgical program also provides services to children from Alaska and has an active bloodless surgery program.

Candidates must be able to read and perform transthoracic and transesophageal echocardiograms. The group is looking for a part-time, salaried, clinic only position without call responsibilities. Potential to become a full-time position with added call responsibilities. The group is also looking for a full-time, salaried position with call and hospitalist responsibilities. Visit our site at [www.pccoforegon.com](http://www.pccoforegon.com) for more information about our clinic.

**Please send curriculum vitae, cover letter, recommendation letters and questions to:**

**[amandal@pccoforegon.com](mailto:amandal@pccoforegon.com)**



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Funded by Cincinnati Children's Heart Institute



# Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

## **Surgical Ablation Found to Be Effective in Reducing Atrial Fibrillation and Improving Quality of Life**

Newswise — CHICAGO—New clinical practice guidelines have been issued by The Society of Thoracic Surgeons (STS) that include major recommendations for the use of surgical ablation when treating Atrial Fibrillation (Afib), the most common type of irregular heartbeat. The guidelines, posted online on December 13<sup>th</sup>, 2016 in *The Annals of Thoracic Surgery*, and appeared in the January 2017 print issue.

“These guidelines represent nearly 2 years of effort by some of the nation’s leading experts in the surgical treatment of atrial fibrillation,” said guidelines co-authors Vinay Badhwar, MD and Gordon F. Murray Professor and Chair of the West Virginia University Heart & Vascular Institute. “This important document highlights the increasing global evidence on the safety and efficacy of surgical ablation for the treatment of Afib.”

STS believes that the practice of summarizing current scientific evidence into clinical practice guidelines and recommendations may contribute importantly to improving surgical outcomes, as well as the quality of patient care. In this case, the literature revealed that surgical ablation as a treatment option for Afib has experienced continued development over the last 30 years, with its frequency and success steadily increasing. The guideline writing committee merged these findings into a singular consensus paper to shape practice, concluding that surgical ablation is effective in reducing Afib and improving quality of life, and so deserves a more prominent role in adult cardiac surgery.

In patients with Afib, rapid, disorganized electrical signals cause the two upper parts of the heart (the atria) to quiver. The quivering upsets the normal rhythm between the atria and the lower parts of the heart (the ventricles). As a result, the ventricles may beat fast and without a regular rhythm.

Afib can lead to blood clots, stroke, heart failure, and other heart-related complications. According to the American Heart Association (AHA), untreated Afib doubles the risk of heart-related deaths and is associated with a significantly increased risk for stroke. At least 2.7 million Americans are living with Afib.

“It is recognized that surgical ablation impacts long-term outcomes with improvements in normal heart rhythm, quality of life, and stroke reduction,” said Dr. Badhwar. “Current evidence reveals that surgical ablation can be performed without significant impact to major complications or death.”

One option for Afib treatment—as the new clinical guidelines recommend—is surgical ablation, also known as the maze procedure. When performing surgical ablation, the surgeon makes very specific and defined lesions in the heart. Scar tissue forms, blocking the abnormal electrical signals while also creating a controlled path for electricity in the heart to follow. The heartbeat should eventually normalize.

Surgical ablation can be done as a standalone procedure or in combination with another heart surgery. In developing these new guidelines, the authors assessed the safety of performing surgical ablation for three surgical approaches: primary open atrial operations where the left atrium, or top chamber of the heart, is already being opened, such as mitral valve repair or replacement and/or tricuspid valve repair; primary closed atrial operations when the left atrium would not otherwise be open, such as Coronary Artery Bypass Grafting (CABG) and/or Aortic Valve Replacement (AVR)

operations; and standalone operations when the only goal is to perform surgical ablation to treat Afib.

The new clinical practice guidelines offer evidence-based recommendations that include:

- Surgical ablation for Afib at the time of concomitant mitral operations to restore cardiac rhythm;
- Surgical ablation for Afib at the time of concomitant isolated AVR, isolated CABG, and AVR+CABG operations to restore cardiac rhythm; and
- Surgical ablation as a primary standalone procedure to restore cardiac rhythm for symptomatic Afib that is resistant to medication or catheter ablation.

The authors also recommend a multidisciplinary heart team assessment, treatment planning, and long-term follow-up in order to optimize patient outcomes in the treatment of Afib.

“These guidelines may help guide surgeons when faced with a challenging decision on the management of Afib,” said Dr. Badhwar. “The guidelines represent an assimilation of the world’s literature; they do not supersede the final medical decision of the surgeon. It is important to remember that the ultimate choice of any therapy remains between the patient and their doctor.”

Badhwar V, Rankin JS, Damiano RJ, Gillinov AM, Bakaeen FG, Edgerton JR, Philpott JM, McCarthy PM, Bolling SF, Roberts HG, Thourani VH, Suri RM, Shemin RJ, Firestone S, and Ad N. The Society of Thoracic Surgeons 2017 Clinical Practice Guidelines for the Surgical Treatment of Atrial Fibrillation. DOI: 10.1016/j.athoracsur.2016.10.076.

Link to Guideline: [http://www.annalsthoracicsurgery.org/article/S0003-4975\(16\)31615-0/fulltext](http://www.annalsthoracicsurgery.org/article/S0003-4975(16)31615-0/fulltext)

*The Annals of Thoracic Surgery* is the official journal of STS and the Southern Thoracic Surgical Association.

## **Therapy for Abnormal Heartbeats May Cause Brain Injury UCSF Researchers Find Link Between Ablation and Brain Lesions**

A common treatment for irregular heartbeats known as catheter ablation may result in the formation of brain lesions when it is performed on the left side of the heart, according to new research at UC San Francisco. Importantly, there also is evidence these lesions may be associated with cognitive decline, meaning they may not be benign.

In a small study of patients undergoing catheter ablation for common abnormal heartbeats from the lower chamber of the heart (Premature Ventricular Contractions (PVCs)), researchers found a significantly higher rate of seemingly asymptomatic brain injury due to embolism among the patients whose therapy occurred on the left ventricle of the heart, which supplies blood to the brain, compared to patients whose therapy was conducted on the right ventricle, which pumps blood to the lungs.

The researchers recommend further study on the impact of these lesions and strategies to avoid them. Their study appeared online Jan. 24, 2017, in the American Heart Association journal *Circulation*.

“The rate of asymptomatic emboli in similar procedures for other types of heart rhythm disturbances tends to be 10%-20%,” said study senior author Gregory Marcus, MD, MAS, a UCSF Health Cardiologist and Director of Clinical Research in the UCSF Division of Cardiology.

"Our study finding is relevant to a large number of patients undergoing this procedure and hopefully will inspire many studies to understand the meaning of and how to mitigate these lesions," Marcus said. "This also will become an important consideration as we think about how to optimally help the large number of people out there with PVCs."

PVCs are extra, abnormal heartbeats originating in the ventricles. They disrupt the heart's regular rhythm and typically have not been a reason for concern.

However, recent research by Marcus and his colleagues demonstrated that PVCs are an important predictor of heart failure and mortality and can cause very bothersome symptoms. Further, such early beats occurring continuously for more than 30 seconds is a potentially serious cardiac condition called ventricular tachycardia (VT).

Given growing recognition of all these phenomena, catheter ablation for PVCs and VT is mainstream and becoming even more common, with well more than 235,000 such procedures performed annually. It is also increasingly used for patients with heart failure due to a weak heart muscle that may improve after frequent PVCs are eradicated.

In this minimally invasive procedure, thin, flexible wires called catheters are inserted into a vein and threaded into the heart. The tip of the catheter either delivers heat or extreme cold to destroy tissue responsible for starting or maintaining the abnormal heart rhythm. The procedure can result in the complete and permanent cessation of the PVCs/VT that are targeted and is generally considered low risk.

An "embolism" occurs when an object moves through the bloodstream from one part of the body to another. Catheters placed in the left side of the heart may lead to brain injury if something that can occlude a blood vessel is either formed, such as a blood clot, or dislodged by the catheter and travels to the brain. Because the right side of the heart leads to the lungs, not the brain, brain emboli generally are not a concern.

Data from previous left heart-based procedures has shown that brain injury



## Heart Program



**PEDIATRIC  
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Miami Children's Health System

### Two Physicians Wanted to Join Our Heart Program's Division of Cardiac Critical Care Medicine

Nicklaus Children's Hospital (formerly Miami Children's Hospital), a 289-bed freestanding children's hospital and Level III trauma center, and Pediatric Specialists of America (PSA), the physician-led group practice of Miami Children's Health System, have an exceptional opportunity for two physicians to join our esteemed Heart Program's division of cardiac critical care medicine.

Our Cardiac Intensive Care Unit (CICU) was the first in the Southeast and provides care for newborns and children receiving treatment for congenital heart defects. With a long-standing tradition of excellence, our cardiac critical care team is currently comprised of six full-time attending physicians and six full-time nurse practitioners. We have an illustrious cardiology fellowship and have offered advanced training in cardiac critical care medicine for more than 20 years. The desired candidates should be board certified or eligible in pediatric critical care medicine or pediatric cardiology. Preference will be given to individuals with dual training in pediatric critical care and cardiology or those board eligible in either cardiology or pediatric critical care who have completed a minimum of one year of advanced training in cardiac intensive care medicine. Applicants should exhibit a strong interest in clinical care, education and academics. Nicklaus Children's Hospital is an affiliate of the Florida International University Herbert Wertheim College of Medicine. Candidates possessing all levels of experience shall be considered.

In October 2016, we will move into our new state-of-the-art Advanced Pediatric Care Pavilion, which will house a 34-bed cardiac in-patient unit with an adjustable acuity model that allows all rooms to accommodate critically ill patients with heart disease. The Heart Program offers a full range of services, including the management of patients following congenital heart surgery, interventional catheterization and invasive electrophysiology. Open heart surgical services are offered to patients as small as one kilogram through young adulthood. Our cardiac surgical program, led by Dr. Redmond Burke, is one of the most innovative in the world and the most transparent. It remains the only cardiovascular surgical program to offer real-time outcomes reporting (<http://www.pediatricheartsurgery.com/realtimeoutcomes/cvperformance.aspx>). Nicklaus Children's also plans to open a birthing center for at-risk fetuses with congenital heart disease. Construction began last fall.

Founded in 1950, the rebranded Nicklaus Children's Hospital is renowned for excellence in all aspects of pediatric medicine and has numerous subspecialty programs that are routinely ranked among the best in the nation. It is also home to the largest pediatric teaching program in the southeastern U.S. Many of the physicians on our staff have trained or worked at other leading medical institutions. Join a phenomenal team that brings lifelong health and hope to children and their families through innovative and compassionate care.

Nicklaus Children's Hospital is located in Miami, Florida, and offers all of the advantages of a tropical, diverse, metropolitan community. Competitive compensation and benefits package.

**Interested candidates please send inquiries to:**

Anthony Rossi, MD

Section Chief, Cardiovascular Medicine

Pediatric Specialists of America/Miami Children's Health System

[anthony.rossi@mch.com](mailto:anthony.rossi@mch.com)

The ACHA website offers resources for ACHD professionals as well as for patients and family members.

Explore our website to discover what ACHA can offer you.

[www.achaheart.org/home/professional-membership-account.aspx](http://www.achaheart.org/home/professional-membership-account.aspx)



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Heart Association  
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thought to be due to embolism rarely occurs. Those emboli generally have been attributed to either issues with the particular patient populations studied or the risks inherent to treating another common heart rhythm disturbance, atrial fibrillation, with ablation.

In this *Circulation* study, Marcus and his colleagues enrolled 18 patients scheduled for VT or PVC ablation over a nine-month period. The average patient age was 58, with half being men, half having a history of hypertension, and a majority having no known vascular disease or heart failure. Most patients were generally healthy.

Left Ventricular (LV) ablation was performed in 12 patients compared to a control group of six patients who underwent Right Ventricular (RV) ablation. Pre- and post-procedural brain magnetic resonance imaging (MRI) was performed on each patient within a week of the ablation, along with a complete neurological examination.

Overall, seven of the 12 patients (58%) who underwent LV ablation experienced 16 brain embolisms combined, compared to zero patients who underwent the RV ablation. Seven of 11 patients (63%) who underwent a retrograde approach to their LV ablation developed at least one new brain lesion.

"Further research is important to understanding the long-term consequences of these lesions and determining optimal strategies to avoid them," said lead author Isaac Whitman, MD, UCSF Cardiac Electrophysiology Fellow.

Other UCSF contributors to the *Circulation* study were Rachel Gladstone, Cardiology Research Assistant; Nitish Badhwar, MD, Professor of Medicine; Henry Hsia, MD, Professor of Medicine; Byron Lee, MD, Professor of Medicine; S. Andrew Josephson, MD, Carmen Castro Franceschi and Gladys K. Mitchell Neurohospitalist Distinguished Professor of Neurology; Karl Meisel, MD, Assistant Professor of Neurology; William Dillon Jr., MD, Elizabeth A. Guillaumin Professor of Radiology; Christopher Hess, MD, PhD, Professor of Radiology; and Edward Gerstenfeld, MD, Professor of Medicine and holder of the Melvin M. Scheinman Endowed Chair in Cardiology. Financial support was provided by the Joseph Drown Foundation.

## Upcoming Medical Meetings

### NeoHeart: Cardiovascular Management of the Neonate

Mar. 22-25, 2017; San Diego, CA USA  
www.choc.org/neoheart

### 51<sup>st</sup> AEPIC Annual Meeting

Mar. 29 - Apr. 1 2017; Lyon, France  
www.aepic2017.org/en/

### CSI Dubai

Apr. 20 - 22, 2017; Dubai, UAE  
www.csi-congress.org/csi-dubai.php

### Catheter Interventions in Congenital, Structural & Valvular Heart Disease

Jun. 28 - Jul. 1, 2017; Frankfurt, Germany  
csi-congress.org

### 7<sup>th</sup> World Congress of Pediatric Cardiology & Cardiac Surgery

Jul. 16 - 21, 2017; Barcelona, Spain  
wcpccs2017.org/en

### CSI-UCSF

Sep. 8 - 9, 2017; San Francisco, USA  
www.csi-congress.org/csi-ucsf.php

### 27<sup>th</sup> International Symposium on Adult Congenital Heart Disease

Sep. 14-16, 2017; Cincinnati, OH USA  
www.cincinnatichildrens.org/  
ACHDSymposium

### LAA 2017

Nov. 17-18, 2017; Frankfurt, Germany  
www.csi-congress.org/laa-workshop.php?go=0

### Devices for Heart Failure

Dec. 15-16, 2017; Berlin, Germany  
www.csi-congress.org/dhf.php?go=0



## Interventional Cardiologist

The Johns Hopkins School of Medicine seeks a pediatric interventional cardiologist to join the Division of Pediatric and Congenital Cardiology at The Charlotte R. Bloomberg Children's Center of the Johns Hopkins Hospital. The applicant must be board certified/eligible in pediatric cardiology and have completed advanced training in congenital interventional catheterization or be an active (>120 cases/yr) practicing pediatric interventionalist, with experience as an independent operator.

The rank of this position is at the level of Assistant Professor or Associate Professor, based on qualifications and academic achievements. The candidate will be responsible for cardiac catheterization, as well as some general cardiology inpatient and outpatient duties. The applicant should have the anticipation of becoming cath lab director.

The Bloomberg Children's Center has a state-of-the-art, dedicated pediatric catheterization laboratory as part of the Johns Hopkins Cardio Vascular Interventional Laboratory (CVIL). The Pediatric suite is both hybrid and 3DRA enabled. Our cath lab is staffed by team of pediatric catheterization-dedicated technicians and nurses, performing the full range of pediatric and congenital cardiac interventional procedures.

### Interested candidates should forward their CV to:

Richard E. Ringel, MD  
Professor of Pediatrics  
Johns Hopkins School of Medicine  
Taussig Heart Center – M2201  
Bloomberg Children's Center  
1800 Orleans Street  
Baltimore, MD 21287

Email: [rringel@jhmi.edu](mailto:rringel@jhmi.edu)

## CONGENITAL CARDIOLOGY TODAY

### CALL FOR CASES AND OTHER ORIGINAL ARTICLES

Do you have interesting research results, observations, human interest stories, reports of meetings, etc. to share?  
Submit your manuscript to: [RichardK@CCT.bz](mailto:RichardK@CCT.bz)



UC San Francisco (UCSF) is a leading university dedicated to promoting health worldwide through advanced biomedical research, graduate-level education in the life sciences and health professions, and excellence in patient care. It also includes UCSF Health, which comprises three top-ranked hospitals, UCSF Medical Center and UCSF Benioff Children's Hospitals in San Francisco and Oakland, and other partner and affiliated hospitals and healthcare providers throughout the Bay Area. For more information, visit [www.ucsf.edu/news](http://www.ucsf.edu/news).

#### **What Patients Say and What Doctors Document - Comparison of Medical Record to Self-Report of Eye Symptoms Shows Wide Variation**

Compare a patient's self-reported eye symptoms to their electronic medical record, and clear discrepancies can be seen.

A study from the University of Michigan Kellogg Eye Center revealed wide disparities in the content of pre-appointment patient questionnaires and what a clinician wrote down to document the visit.

"We found pretty noticeable differences between the two," says Maria Woodward, MD, MS, an Assistant Professor of Ophthalmology and Visual Sciences at U-M. "I think certainly the biggest takeaway is when people are presented things in different ways, they tell you different things."

The study, published Jan. 26<sup>th</sup> in *JAMA Ophthalmology*, analyzed the symptoms of 162 Kellogg patients. Each completed a 10-point survey while waiting to see a physician; questions came from sources including the National Institutes of Health Toolbox.

The doctors treating these patients weren't told about the surveys, or that their record-keeping would be reviewed for comparison.

The contrast was stark: of the study's group, "exact agreement" between an individual's survey, and what appeared on their medical record occurred in only 38 patients.

#### **A need for focus**

Symptom reporting drove the inconsistencies between surveys and medical records, the study found.

The top discordant issue: glare. Of patients reporting concern about glare on their surveys, 91% didn't have it on their medical record.

Eye redness was second-most common; (80% had no medical record mention, followed by eye pain (74.4%). Blurry vision was only the symptom to tilt the scales -- with more instances of inclusion in medical records than in questionnaires.

As a result, other doctors treating the same patient in future visits could have an incomplete picture of their symptoms.

Perhaps more risky: because digital medical records are increasingly used to guide clinical practice or research, the collective data may be shortsighted or misleading in some scenarios, Woodward says.



## **NATIONWIDE CHILDREN'S**

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### **Congenital Cardiac Intensivist**

The Heart Center (THC) at Nationwide Children's Hospital, the primary pediatric teaching facility for The Ohio State University in Columbus Ohio, is recruiting an attending physician, at any academic level, for the Cardiothoracic Intensive Care Unit (CTICU). This individual would join a group of seven multi-background academic cardiac intensivists and eight dedicated nurse practitioners devoted to the CTICU providing 24/7 in house coverage.

Our current independently-managed free-standing CTICU is a 20 bed unit with approximately 800 total admissions per year (medical and surgical) and an average daily census of 12. Candidates must have completed fellowship training in pediatric cardiac anesthesia, critical care and/or cardiology that includes advanced cardiac intensive care training. Preference will be given to those with experience and are boarded in pediatric cardiology. THC embraces a culture of patient safety and quality, transparency, translational/outcome research, education, cost-containment and public health awareness. These create ample participation and leadership opportunities for the candidate's professional growth.

THC is comprehensive with services including an active hybrid palliation center, a comprehensive single ventricle program, thoracic organ transplantation program, blood conservation strategies, and cardiac mechanical support team. The CTICU is supported by world-class and innovative interventional cardiology, cardiac imaging, cardiothoracic surgery and adult congenital heart experts. Current annual clinical metrics for THC includes: over 500 cardiothoracic surgeries, over 700 cardiac catheterizations and EP procedures, and over 13,000 cardiology outpatient visits. We have a pediatric and pediatric/adult combined cardiology fellowship programs. We participate in numerous multicenter clinical trials and quality initiatives including the JCCHD QI Collaborative. We are directly linked to our Center for Cardiovascular and Pulmonary Research which has an NIH T-32 training grant.

**Interested candidates are encouraged to submit their curriculum vitae to:**

Janet Simsic, MD

Director of the Cardiothoracic Intensive Care Unit,  
Nationwide Children's Hospital

T2296

700 Children's Drive  
Columbus, OH 43205

or [janet.simsic@nationwidechildrens.org](mailto:janet.simsic@nationwidechildrens.org)



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"Many parties in health care use the electronic health records now and they expect the data to accurately reflect the interaction with the doctor," says Woodward, also a member of the Institute for Healthcare Policy and Innovation.

Explanations for the medical record-doctor-patient disconnect are understandable, she adds, with neither party at fault. The doctor-patient relationship is more nuanced than what is reflected in the medical record. A patient might not choose to mention all of their symptoms. Doctor dialogue may follow a conversational path versus a point-by-point checklist.

Time constraints of record keeping in the electronic chart can also be an issue. And not every detail of a given appointment -- particularly minor concerns -- is necessarily worth documenting.

Still, notes Woodward, "The concern highlighted by this research is that important symptoms may be overlooked. If a patient has severe symptoms, all of those symptoms should be documented and addressed."

#### *Gaining greater clarity*

The study highlights an opportunity to improve lines of communication between patients and doctors.

For example, implementing pre-appointment eye symptom questionnaires, similar to those in the study, could be simple and effective, Woodward says. A similar pilot program, she notes, is underway in her clinic. "This is definitely a pathway I see as very feasible to resolving this disconnect in the near future; the infrastructure is already there," she says.

The concept also could help bring more clarity to what ends up on a patient's medical record.

Because the surveys Woodward and her team used asked participants to assess their conditions' severity on a numeric scale, results could help practitioners better evaluate the depth of a patient's symptoms -- and even identify concerns that might have gone unnoticed.

The use of a self-report system before seeing the doctor could "really change the conversation between the doctor and the patient," says Woodward. Rather than spending time to identify symptoms, doctors and patient could be talking about how to manage severe symptoms.

#### **How Medical Simulation Save Lives**

Newswise — Medical simulation, the use of manikins, virtual reality, live action scenarios designed to train first responders, physicians, anesthesiologists, nurses and other medical professionals, provides life-saving techniques.

Through a series of education sessions, keynote addresses and an exhibit floor with many of the newest technologies, simulation was on full display at IMSH, the International Meeting for



## Memorial is creating leaders.

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### **Manager Pediatric Echocardiography**

We are currently seeking a Manager Pediatric Echocardiography to manage the daily operations of multiple departments within pediatric imaging to ensure alignment with departmental and organizational objectives. Duties will include:

- Assisting with developing specific departmental goals, standards, and objectives which directly support the strategic plan and vision of the organization.
- Managing staff relations including performance management, staff satisfaction, and conflict management. Performs and oversees scheduling, recruitment, payroll, and student engagements.
- Developing and monitoring departmental budgets, regulatory compliance, departmental contracts and vendor relations. Determines and justifies needs for systems/equipment/supplies purchases, monitors usage, and oversees proper working order and/or stock supplies.
- Assessing the quality of patient care delivered and coordinates patient care services with patients, staff, physicians, and other departments.

#### Qualifications:

- Graduate of an accredited Imaging Technologist program.
- Bachelor's degree in management or related field.
- 3 years of pediatric, imaging congenital heart defect and surgical repair at an accredited ICAEL institution required.
- Registered as a diagnostic medical sonographer PEDS, or diagnostic cardiac sonographer Peds - RDCS (Peds) by the American Registry for the Diagnostic Medical Sonography (ARDMS) or as a technologist (RT) by the American Registry of Radiographic Technologists (ARRT).
- Florida state licensure.
- Critical thinking skills, effective communication skills, decisive judgment and the ability to work with minimal supervision.
- Must be able to work in a stressful environment and take appropriate action.

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**Barth Syndrome (ICD-10: E78.71)**  
**Symptoms:**  
 Cardiomyopathy, Neutropenia, Muscle Weakness,  
 Exercise Intolerance, Growth Delay, Cardiolipin Abnormalities  
[www.barthsyndrome.org](http://www.barthsyndrome.org)

Simulation in Healthcare in January at the Hyatt Regency Orlando.

The International Meeting on Simulation in Healthcare (IMSH) is the world's largest conference dedicated to healthcare simulation learning, research and scholarship, offering 250 sessions in various formats, from large plenary sessions to small, interactive immersive courses. It is also considered to be the educational and networking event for the simulation world; in fact, 92% of last year's attendees rated networking opportunities the most valuable or strongest value of attending.

The Society for Simulation in Healthcare (SSH) seeks to improve performance and reduce errors in patient care through the use of simulation. Established in 2004 by professionals using simulation for education, testing, and research in health care, SSH membership includes nearly 3,200 physicians, nurses, allied health and paramedical personnel, researchers, educators and developers from around the globe. SSH fosters the improvement and application of simulation-based modalities such as human patient simulators, virtual reality, standardized patients and task trainers.

### Soft Robots Hug the Heart to Help Pump Blood

An implantable soft-robotic device could help failing hearts pump blood by giving the organ gentle squeezes, mimicking the natural motion of cardiac muscle, a new study reveals. The silicon-based device, which stiffens or relaxes when inflated with pressurized air, could prove to be a promising strategy for the development of assistive devices for heart failure – a serious condition afflicting 41 million people worldwide and over five million in the U.S., costing the nation an estimated \$32 billion each year. While ventricular assist devices (VADs) are currently used as a life-prolonging therapy, they are in constant contact with the blood – increasing a patient's risk for infection, coagulation and stroke, and requiring the use of long-term blood-thinning medications.

VADs also interfere with the normal curvature of the heart and its contraction mechanics. Seeking to pioneer a more effective device, Ellen Roche and colleagues developed a novel apparatus designed to augment cardiac function by closely replicating normal heart muscle behavior, instead of disrupting it. In ex vivo experiments, the device successfully conformed to porcine heart surfaces, synchronizing with native heart motion. It also restored normal blood flow after acute cardiac arrest in six living pigs. The researchers were able to "fine tune" the device by selectively twisting and compressing either the right or left ventricle of explanted pig hearts – a key finding, as chronic heart-failure often only affects a portion of the organ. With further investigation, the device could be tailored for individual patient needs, to better target cardiac rehabilitation or recovery. Additional work is needed to make this technology suitable for longer-term implantation in the body, the authors say.

Article #3: "Soft robotic sleeve supports heart function," by E.T. Roche; M.A. Horvath; A. Alazmani; S.-E. Song; W. Whyte; C.J. Payne; D.J. Mooney; C.J. Walsh at Harvard University in

Cambridge, MA; E.T. Roche; M.A. Horvath; A. Alazmani; S.-E. Song; W. Whyte; C.J. Payne; J. Weaver; D.J. Mooney; C.J. Walsh at Wyss Institute for Biologically Inspired Engineering in Boston, MA; E.T. Roche at National University of Ireland Galway in Galway, Ireland; M.A. Horvath at Technische Universität München in Garching, Germany; I. Wamala; A. Alazmani; S.-E. Song; Z. Machaidze; J. Kuebler; N.V. Vasilyev; F.A. Pigula at Boston Children's Hospital in Boston, MA; A. Alazmani at University of Leeds in Leeds, UK; S.-E. Song at University of Central Florida in Orlando, FL; W. Whyte at Royal College of Surgeons in Ireland in Dublin, Ireland; W. Whyte at Trinity College Dublin in Dublin, Ireland. For a complete list of authors, please see the manuscript.

To view the papers abstract, go to: <http://stm.sciencemag.org/lookup/doi/10.1126/scitranslmed.aaf3925>

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# Congenital Cardiac Care Providers in North America at Hospitals That Offer Open Heart Surgery for Children

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