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CSI at UCSF - Catheter Interventions in Structural, Valvular & Congenital Heart Disease, Atrial Fibrillation & Heart Failure

Sep. 8-9, 2017; San Francisco, CA USA
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choa.org/medical-professionals/professional-events/50th-anniversary-southeast-pediatric-cardiovascular-society-meeting

11th IPC Workshop

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Oct. 8-11, 2017; Rochester, MN USA
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Rare Case of a Left Superior Vena Cava Connection to Left Upper Pulmonary Vein

By Sinéad L. Murphy; Kenneth Fang, MD; Tabitha Moe, MD

Keywords: Left superior vena cava, Congenital Heart Disease (CHD), Anomalous left upper pulmonary venous return, Thromboembolic stroke

Abstract

A persistent Left Superior Vena Cava (LSVC) is a relatively common, quiescent congenital anomaly in the general population, often with drainage into the coronary sinus and return to the right atrium. We present a rare case of persistent LSVC with connection to the Left Upper Pulmonary Vein (LUPV), creating potential for right-to-left shunting, in the absence of other congenital anatomical anomalies. This connection can result in cyanosis, increased risk for endocarditis and, as seen in this patient, paradoxical thromboembolism. A discussion of morphologic forms, diagnostic imaging, clinical significance, and potential corrective procedures is reviewed.

Case Report

We present the case of a 64-year-old female who initially presented to care in the Bahamas for an acute middle cerebral artery stroke, thought to be thromboembolic in nature. TPA was administered, and she was transferred to Arizona for management and further workup. In the search for thromboembolic origin, a transthoracic echocardiogram was performed, showing

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severe mitral regurgitation, mitral valve

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signs of shunting were observed on echocardiogram. With the working diagnosis of undetected atrial fibrillation, the patient was scheduled for operative replacement of the mitral valve.

Pre-operatively, a left internal jugular central venous catheter was placed for hemodynamic management during surgery. Initially, the catheter transduction demonstrated venous waveforms as anticipated, but subsequently changed to demonstrate an arterial pattern. The patient was hemodynamically stable, and the surgical team chose to proceed as planned. Careful dissection showed a persistent LSVC which drained into the LUPV which then returned normally to the left-sided atrium (Figure 1). The other pulmonary veins were identified and found to return to the left atrium. The right-sided superior vena cava was intact and patent. The central venous catheter was withdrawn into the venous system, and the anomalous systemic-pulmonary venous connection was ligated.

Post-operatively, the patient developed advanced atrio-ventricular block and profound bradycardia, which precipitated torsades de pointes on three occasions. As a result, she underwent transvenous placement via right subclavicular approach of a permanent pacemaker without further incident. She was discharged to inpatient rehabilitation with plans to return to the Bahamas.

Discussion

A persistent Left Superior Vena Cava (LSVC) is a relatively common, incidentally found congenital anomaly in the general population, with estimated prevalence of from 0.3% to 2%.¹ In patients with other forms of CHD the prevalence increases to 4.4%, and has been reported in association with heterotaxy syndromes, primum-type Atrial Septal Defects, clefting of the left-sided atrioventricular valve, atrioventricular canal defects, and coarctation of the aorta.² Most patients are asymptomatic, with 80%-90% of reported anomalies draining into the coronary sinus and right atrium.¹

The condition presents more significant risk and sequelae, rarely, when the LSVC connects to a left heart structure, creating the potential for a shunting defect. This can occur with persistent LSVC drainage through the left atrium via a direct connection to the left atrium, or through the Left Upper Pulmonary Vein (LUPV). These types of anomalies are

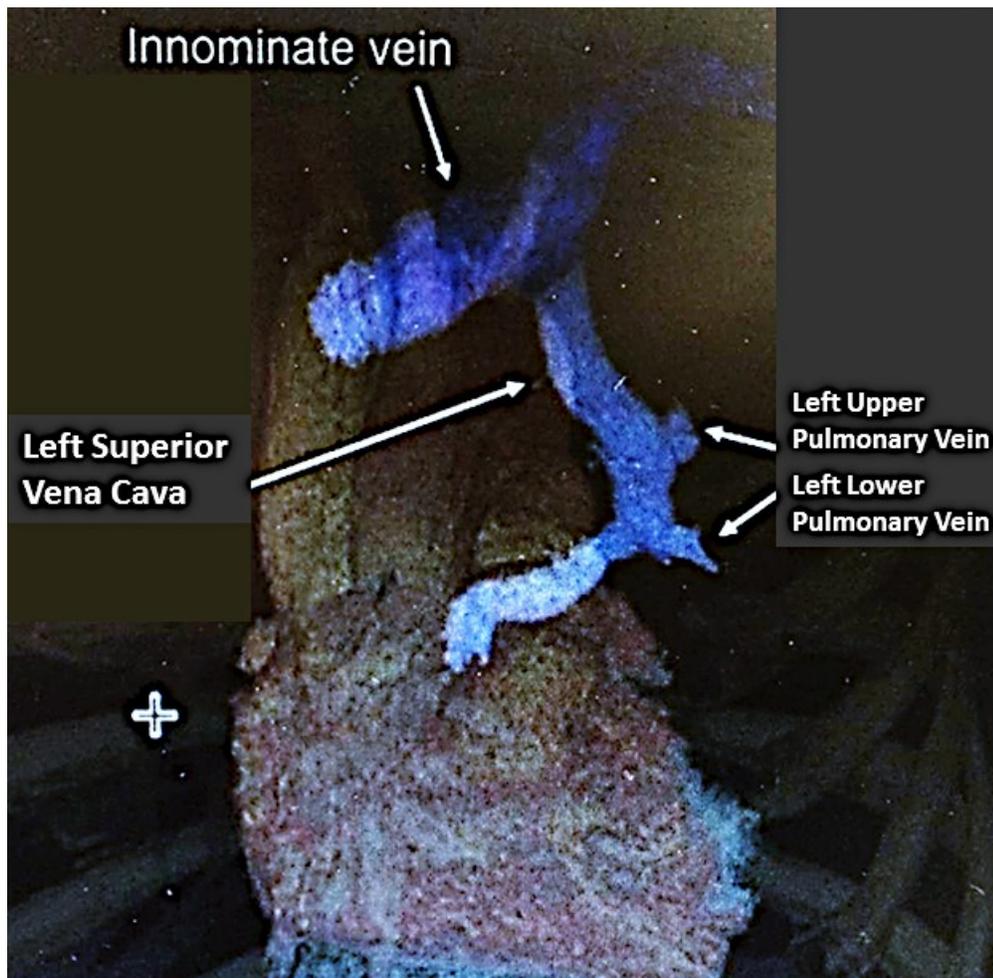


Figure 1. Angiography of left superior vena cava to left upper pulmonary vein connection. Informed consent: Informed consent was obtained from the individual participant included in the study.

associated with an increased risk of cyanosis, heart failure, intracerebral abscess,^{3,4} and embolic cerebrovascular events.⁵ Reports of these anomalies often follow embolic stroke work up, as seen in this patient.

The case we present is a very rare morph of persistent LSVC to LUPV connection with only a half dozen other cases reported in the literature to our knowledge.^{3, 5-8} Of these reported cases, only three have been in the absence of other congenital cardiac anomalies. Diagnostic modalities previously reported include chest radiograph,⁴ angiography⁴ and digital subtraction angiography,⁵ contrast enhanced computer tomography scan,⁵ transesophageal echocardiography with microbubbles,⁷ and open dissection. With the current imaging

modalities, the authors believe cardiac magnetic resonance imaging would also be a viable method of diagnosis. Intervention is not absolutely indicated, with medical management to avoid thromboembolic events a reasonable alternative, but necessary due to right-to-left shunting potential. Reported methods of closure include: surgical diversion of the LSVC to the right atrium via baffle in the case of absent right SVC,⁴ occlusion of the LSVC with the Amplatzer Vascular Plug,⁷ and surgical ligation of the LSVC. Post-operative side effects have included Superior Vena Cava Syndrome,⁴ and arrhythmia.

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- The main text of the article should be written in informal style using correct English. The final manuscript may be between 400-4,000 words, and contain pictures, graphs, charts and tables. Accepted manuscripts will be published within 1-3 months of receipt. Abbreviations which are commonplace in pediatric cardiology or in the lay literature may be used.
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Position Details: The Pediatric Nurse Practitioner/Physician Assistant in Pediatric Interventional Cardiology works in collaboration with the supervising physician and assists in performing diagnostic and interventional procedures for cardiology patients with critical acuity. The ACP will work with our two interventionalists and one transplant cardiologist and be the liaison between the Pediatric Cardiologist, patients and families and staff. Our new state of the art two Cath Lab suite opened in February 2017 right beside our CVICU. The cath lab volume has been growing at a rate of 10-15% per year, and we will be developing a 3-D printing program in the next 6 months. Active industry sponsored research is ongoing, affording many opportunities for clinical research projects.

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Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

Children's Hospital of Philadelphia and Mount Sinai Health System Mark Milestone in Fetal Medicine and Children's Heart Programs

Newswise — Children's Hospital of Philadelphia (CHOP) and the Mount Sinai Health System took another step forward at the end of May in their 18-month-old alliance with the official opening of their Fetal Medicine Program and the affiliation of the Mount Sinai Children's Heart Center with the Cardiac Center at CHOP.

Officials from both hospitals participated in a ribbon-cutting ceremony held today at the new facility at The Mount Sinai Hospital's Annenberg Building.

The Fetal Medicine Program will offer access to an unprecedented scope of services. This program provides mothers carrying fetuses at risk for or identified with possible anomalies a "one-stop experience" that includes a comprehensive diagnostic evaluation and consultation. Specifically, in a single-day visit, the patient will undergo state-of-the-art diagnostic testing, which may include ultrasound, echocardiography, and fetal magnetic resonance imaging (MRI), and then meet with a team of Mount Sinai and CHOP experts to discuss the presumptive diagnosis and options for treatment. All diagnostic testing will be performed at The Mount Sinai Hospital; images will be read by specialists in fetal radiology at CHOP and Mount Sinai in coordination with Mount Sinai Maternal Fetal Medicine Specialists using telemedicine video links. Once the diagnostics are reviewed, and depending on the presumptive diagnosis, a conference with each patient and her family may include a maternal fetal medicine specialist, pediatric cardiologist, pediatric surgeon, geneticist, and other relevant pediatric subspecialists. CHOP subspecialists will participate in these family meetings by video conferencing. The program is the only one in New York City offering such convenience and level of services.

The new affiliation of the Mount Sinai Children's Heart Center and the CHOP Cardiac Center brings to New York access to unparalleled expertise and resources from one of the nation's leading pediatric cardiac centers. The affiliation includes the Fetal Heart Program, which aims to diagnose Congenital Heart Disease (CHD) as early as 12-14 weeks gestation utilizing fetal echocardiography. The results of the fetal imaging can then allow the clinical care team, comprised of providers at Mount Sinai and CHOP collaborating via telemedicine technology, to develop an optimal plan for care. The Children's Heart Center at Mount Sinai Hospital—overseen by the Divisions of Pediatric Cardiology and Cardiac Surgery—provides a continuum of care from fetal through adult life, and will now have access to the expertise of CHOP's Cardiac Center in areas such as pediatric cardiac intensive care and pediatric cardiothoracic surgery. The Children's Heart Center offers exceptional pediatric cardiology and cardiac surgical services, including echocardiography, exercise testing, Holter monitoring, interventional cardiology and angiography, and cardiovascular genetics. The Mount Sinai and CHOP teams can provide joint expertise at all points of treatment through direct consultation and use of telehealth technology.

The alliance between Mount Sinai and Children's Hospital of Philadelphia was announced in the fall of 2015, and includes three services: Fetal Medicine, Pediatric Cardiac Care, and Pediatric Oncology. This collaboration is intended to give patients and their families access to the most advanced diagnostics and treatments delivered by Mount Sinai and CHOP experts close to home at select Mount Sinai locations.

"Mount Sinai is pleased to collaborate with CHOP, a world-renowned institution, to ensure excellent patient care," said Kenneth L. Davis, MD, President and Chief Executive Officer of the Mount Sinai Health System. "Our goal is to offer the highest quality maternal, fetal, and pediatric care to patients—especially those with complex needs—across a large health system and a fast-growing ambulatory care network. And together, we will be uniquely positioned to recruit and retain the best faculty in the region."

"CHOP is pleased to mark this milestone in the relationship between two health care institutions that are totally dedicated to caring for the health of children and their families," said Madeline Bell, President and Chief Executive Officer of Children's Hospital of Philadelphia. "The alliance with the Mount Sinai Health System continues to evolve and reflects both a shared vision to better serve families in the New York region and a mutual professional respect and admiration between our organizations," Bell said.

The Mount Sinai Health System is an integrated health system committed to providing distinguished care, conducting transformative research, and advancing biomedical education. Structured around seven hospital campuses and a single medical school, the Health System has an extensive ambulatory network and a range of inpatient and outpatient services—from community-based facilities to tertiary and quaternary care.

The System includes approximately 7,100 primary and specialty care physicians; 12 joint-venture ambulatory surgery centers; more than 140 ambulatory practices throughout the five boroughs of New York City, Westchester, Long Island, and Florida; and 31 affiliated community health centers. Physicians are affiliated with the renowned Icahn School of Medicine at Mount Sinai, which is ranked among the highest in the nation in National Institutes of Health funding per investigator. The Mount Sinai Hospital is in the "Honor Roll" of best hospitals in America, ranked No. 15 nationally in the 2016-2017 "Best Hospitals" issue of *U.S. News & World Report*. The Mount Sinai Hospital is also ranked as one of the nation's top 20 hospitals in Geriatrics, Gastroenterology/GI Surgery, Cardiology/Heart Surgery, Diabetes/Endocrinology, Nephrology, Neurology/Neurosurgery, and Ear, Nose & Throat, and is in the top 50 in four other specialties. New York Eye and Ear Infirmary of Mount Sinai is ranked No. 10 nationally for Ophthalmology, while Mount Sinai Beth Israel, Mount Sinai St. Luke's, and Mount Sinai West are ranked regionally. Mount Sinai's Kravis Children's Hospital is ranked in seven out of ten pediatric specialties by *U.S. News & World Report* in "Best Children's Hospitals." For more information, <http://www.mountsinai.org>.

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All Heart Patients Have Some Liver Disease After Fontan Surgery - CHOP Researchers Report Universal Liver Fibrosis in Survivors of Operation for a Severe Heart Defect

Newswise — Patients who undergo the Fontan operation as children for a Complex Congenital Heart Defect are at risk of developing progressive liver fibrosis, a buildup of fibrous deposits, as a result of the circulation created by the surgery, according to a new study. A research team says their findings underscore the importance of improving ongoing medical surveillance, so that physicians can develop the most appropriate care for their patients.

The Fontan operation is a series of three-staged reconstructive surgeries in children born with single-ventricle disease, a life-threatening condition characterized by a severely underdeveloped ventricle, one of the heart's two pumping chambers. The palliative surgeries re-route blood to the lungs, but result in an abnormal physiology that puts many organ systems at risk. One such organ is the liver, in which fibrosis develops in response to elevated pressure within the veins. Over time this can lead to liver cirrhosis, a condition of significant liver impairment.

"We have known for some time that liver fibrosis is a complication of Fontan surgery, but this was an important study that demonstrated that the length of time after the Fontan operation is a significant contributor to the degree of fibrosis," said study leader David J. Goldberg, MD, a pediatric cardiologist at Children's Hospital of Philadelphia (CHOP).

Goldberg and colleagues published their study online April 26th in the *Journal of the American Heart Association* (<https://doi.org/10.1161/JAHA.116.004809>).

The research is a retrospective analysis of 67 children and adolescents (with a mean age of 17 years) evaluated at CHOP between 2009 and 2014 under the hospital's Single Ventricle Survivorship Program. Most of these patients were evaluated 10 to 15 years after their original Fontan operation.

Most of the patients had no overt symptoms of liver disease, which progresses very gradually, and were generally considered to be in good health. All 67 patients showed evidence of some liver fibrosis, measured by the quantity of collagen deposits found after needle biopsies of the patients' livers. The only risk factor the researchers found was time from Fontan—the degree of fibrosis increases over time. The researchers added that there are likely other risk factors for fibrosis not measured in their analysis.

"Our finding that liver fibrosis is universal after the Fontan operation reinforces the need for ongoing surveillance of the liver in this patient population," said Goldberg. "However, it is important to find a less invasive method than liver biopsy to measure liver fibrosis, as well as to investigate possible medications that may reduce the rate of fibrosis. As we work to develop clinical guidelines for liver surveillance, we can better provide care tailored to our individual patients."

Children's Hospital of Philadelphia: Children's Hospital of Philadelphia was founded in 1855 as the nation's first pediatric hospital. For more information, visit www.chop.edu



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Heart Center

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The Department of Cardiology at Boston Children's Hospital seeks an experienced interventional cardiologist to join a thriving, well-established, academic practice at a Harvard teaching hospital. The applicant will be Board certified/eligible in pediatric cardiology, have completed advanced training in congenital interventional catheterization, will be an active (>200 cases/yr) practicing pediatric interventionalist with prior experience as an independent operator, and a record of academic accomplishment.

Applicants with unique areas of clinical expertise, or a successful record of program development, will be given priority as potential additions to a diverse existing staff of interventional providers. Robust clinical and translational resources and opportunities exist within the Department. In addition to procedural responsibilities, the position entails participation in fellowship training, quality initiatives, and limited inpatient services.

The catheterization facilities at Boston Children's Hospital consist of 3 state-of-the-art, mixed-vendor, biplane cardiac catheterization laboratories with rotational angiography capability, in addition to a 4th biplane lab and a single plane lab for accommodation of invasive electrophysiology procedures. These procedural areas are supported by a central, 10 bed recovery and post-anesthesia unit, in close proximity to a dedicated cardiac MRI suite and inpatient echocardiography lab.

Interested candidates should forward their CV to:

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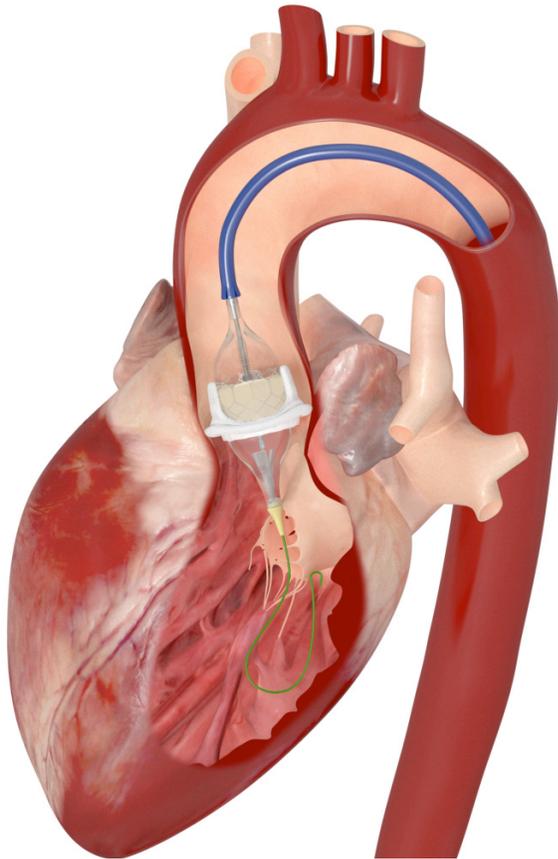
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Edwards SAPIEN 3 Valve Receives FDA Approval For Aortic, Mitral Valve-In-Valve Procedures

PRNewswire - Edwards Lifesciences Corporation, a global leader in patient-focused innovations for structural heart disease and critical care monitoring, announced June 5th it has received U.S. Food and Drug Administration (FDA) approval for aortic and mitral valve-in-valve procedures using the Edwards SAPIEN 3 transcatheter heart valve. The SAPIEN 3 valve is the first transcatheter heart valve approved in the U.S. for the treatment of both aortic and mitral



Credit Edwards Lifescience

Top: (Anatomical image) An image of the deployed SAPIEN 3 transcatheter heart valve inside a surgical valve

Bottom: The SAPIEN 3 transcatheter heart valve is now approved for aortic and mitral valve-in-valve procedures in patients at high risk of a subsequent surgery.



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The Department of Pediatrics at the University of North Carolina School of Medicine is recruiting at an open rank, fixed term clinical faculty position for the Division of Pediatric Cardiology. We are seeking a faculty member to join our Division to augment our team of non-invasive cardiologists in the expansion of subspecialty services. The ideal candidate will be an individual with advanced training or experience in all aspects of non-invasive imaging including fetal echocardiography and cardiac MRI. The candidate should also be skilled and interested in the practice of ambulatory and inpatient pediatric cardiology. Primary responsibilities will include interpretation of non-invasive imaging, outreach and telemedicine, and inpatient care. Clinical research and quality improvement interests will be encouraged. The faculty member will be involved in the education of medical students and house staff.

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Timothy M. Hoffman, MD
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patients who are at high risk for a subsequent open-heart surgery to replace their bioprosthetic valve.

"Expansion of the SAPIEN 3 device indication to include valve-in-valve procedures is a meaningful advancement for patients at high risk of an additional open-heart valve procedure, particularly for those in need of a safe alternative for mitral valve

replacement," said Larry L. Wood, Edwards' Corporate Vice President, transcatheter heart valves.

This anticipated FDA approval of the indication expansion was supported by real-world data collected from the Society of Thoracic Surgeons and American College of Cardiology (STS/ACC) Transcatheter Valve Therapy (TVT) Registry. The TVT Registry includes information and outcomes on patients undergoing transcatheter valve replacement and repair procedures in the United States

"This approval brings a safe and effective transcatheter therapy to patients who would do very poorly with repeat open-heart surgery," said John Carroll, MD, Professor of Cardiology at the University of Colorado School of Medicine and Director of Interventional Cardiology at the University of Colorado Hospital, Denver and member of the TVT Registry Steering Committee. "I am pleased to see that the FDA recognizes the value of the high-quality evidence generated by the STS-ACC TVT Registry and its ability to play an important role in assessing 'real-world' clinical results in specialty indications, such as valve-in-valve, and for particular patient groups, such as those needing replacement of a bioprosthetic mitral valve."

The Edwards SAPIEN 3 valve was approved by the FDA in 2015 for severe, symptomatic aortic stenosis patients at high risk for open-heart surgery, and, in 2016, received approval for the treatment of patients who are at intermediate risk for open-heart surgery. The SAPIEN 3 valve builds on Edwards' decades of experience in the development of tissue heart valves, and the proven benefits of the Edwards SAPIEN valves.

For more information on Edwards Lifesciences Corporation, visit www.Edwards.com.

ACC Opposes Current Approaches to Health Care Reform ACC President Urges Senate to Work in 'Open, Bipartisan Process'

WASHINGTON (July 25, 2017) — American College of Cardiology President Mary Norine Walsh, MD, FACC, made the following statement on the upcoming Senate vote to proceed to health care reform legislation, "In light of new analysis from the Congressional Budget Office (CBO), it is clear that the health reform measures being contemplated in the Senate, whether the Better Care Reconciliation Act or the Obamacare Repeal Reconciliation Act, would result in the loss of insurance coverage for millions of Americans, limiting access to care for those who need it most."

"The American College of Cardiology opposes both of these approaches to health reform. Instead, we urge Senate leaders and members on both sides of the aisle to work in an open, bipartisan process to advance concepts that align with the ACC's Principles for Health Reform. These principles prioritize patient access to meaningful insurance coverage and protection for individuals with pre-existing conditions, goals that would advance the outcome we all strive for: a healthcare system that strengthens and secures the highest quality care for all Americans."

For more information, visit acc.org.

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Our Division of Cardiology (<https://healthsciences.ucsd.edu/som/pediatrics/Divisions/cardiology>) is recruiting for 1-2 full-time general cardiologists at any career level. The successful candidate(s) will be responsible for seeing 6-8 half day clinics sessions weekly in the Rady Children's Hospital Heart Center and in satellite clinics located in San Diego County and regionally. The successful candidate will play a major role in our clinical care, and in the education of students, residents and fellows.

Candidates for this position must have an M.D. and be board-certified or board-eligible in pediatric cardiology. Candidates must also be licensed or licensable to practice medicine in the state of California. Preferred candidate(s) will be proficient in speaking, reading, and writing Spanish for our Spanish-speaking population of patient families. Additional training in preventive pediatric cardiology is highly desirable.

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garywebb6@gmail.com



The CHIP Network, the Congenital Heart Professionals Network, is designed to provide a single global list of all CHD-interested professionals.

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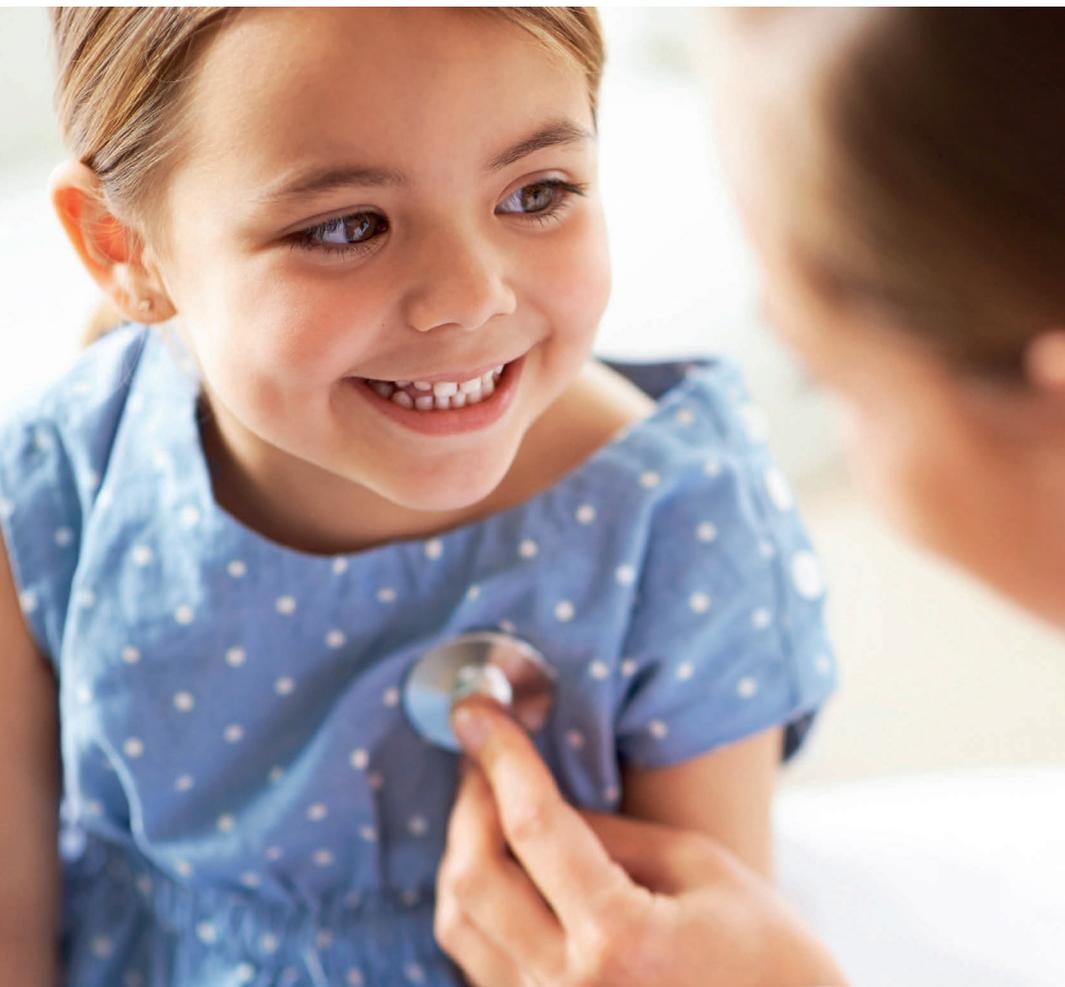
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