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3D Printing of Interrupted Aortic Arch Type C with Isolation of Right Subclavian Artery: The First Reported Case

By Samantha Gilg, MD; Gabe Linke; James Hammel, MD; Scott Fletcher, MD

Introduction

Three-dimensional (3D) printing has become a valuable imaging modality for Complex Congenital Heart Disease. 3D modeling of the heart has been described in the literature and its importance in the diagnosis and treatment of Congenital Heart Disease is rapidly expanding¹.

Interrupted Aortic Arch (IAA) is a rare congenital defect, accounting for about 1% of all Congenital Heart Disease^{2,3}. The most common type of interruption is Type B, which occurs between the left common carotid and the left subclavian artery. Type A is less common and occurs beyond the left subclavian artery origin. Type C is the most rare, accounting for only 3% of cases of IAA, and occurs between the innominate artery and the left carotid artery⁴.

Isolation of the right subclavian artery from the pulmonary artery is another rare congenital cardiac anomaly. It is, however, frequently associated with other aortic arch or intracardiac anomalies, which are reported in up to 68% of patients with isolated right subclavian⁵.

Case Report

Our patient was diagnosed prenatally with suspicion of Interrupted Aortic Arch and Ventricular Septal Defect (VSD). The patient

was born via C-section at 38 weeks and an echocardiogram showed large outlet malalignment VSD and IAA of uncertain type due to inability to accurately visualize the aortic arch branching pattern and the continuity between ascending and descending aorta.

A cardiac CT scan was performed to clarify anatomy; the scan demonstrated prior known intracardiac anatomy and bilateral Patent Ductus Arteriosus (PDA). The right-sided PDA provided blood flow to an isolated right subclavian artery and the left-sided PDA provided flow to the descending aorta, distal to the right carotid artery, with Type C-IAA.

A classic Edward's diagram, which represents the abnormal embryologic development of our patient, is shown (Figure 1 on the following page). The patient had normal ascension of the 7th intersegmental artery on the right followed by distal involution, but also abnormal involution of the proximal right 4th arch early on in development, which led to formation of the isolated right subclavian artery receiving blood flow from the right-sided PDA (persistent ductal portion of the right 6th arch). The patient also had abnormal regression of the ventral portion of the left 3rd and 4th arches leading to the Type C-IAA and required the left-sided PDA to provide blood flow to the descending aorta.

On Day of Life 4 the patient was taken for complete repair which included re-implantation of the anomalous right subclavian artery from the right pulmonary artery to the right carotid artery, Yasui arch repair, and Rastelli right

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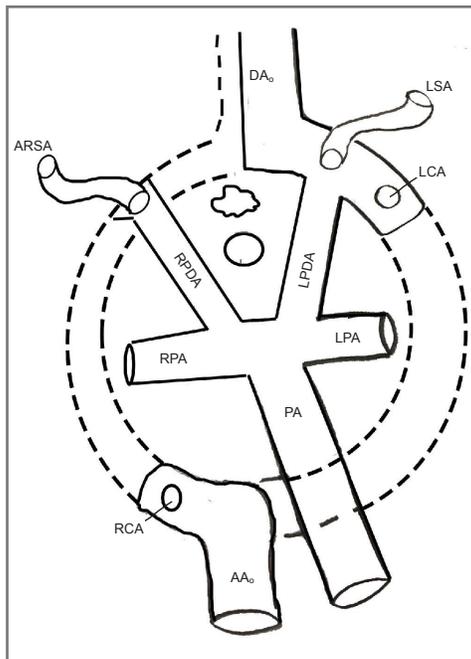


Figure 1. Edward's Diagram. ARSA – Aberrant Right Subclavian Artery. DA_o – Descending Aorta. LSA – Left Subclavian Artery. LCA – Left Carotid Artery. AA_o – Ascending Aorta. RCA – Right Carotid Artery. RPDA – Right Patent Ductus Arteriosus. LPDA – Left Patent Ductus Arteriosus. RPA – Right Pulmonary Artery. LPA – Left Pulmonary Artery. PA – Pulmonary Artery.

ventricle-to-pulmonary artery homograft. The patient has had a very good result clinically and based on echocardiograms.

Discussion

While cardiac CT imaging has allowed for significant advancements in pre-surgical anatomic diagrams, 3D printing has expanded on this technology, allowing for creation of a precise anatomic model which optimizes pre-operative planning and intraoperative strategy.

In our case, the 3D printed heart model was used to accurately delineate the origin of the right subclavian artery and the specific type of IAA for our patient's rare anatomical variant. This allowed the team to accurately plan for the complex operative repair. Being prepared led to an excellent repair and post-operative outcome.

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3D reconstruction from high-resolution CT images and 3D printing was utilized to determine the exact relationship of great vessels prior to surgical intervention (Figures 2 and 3).

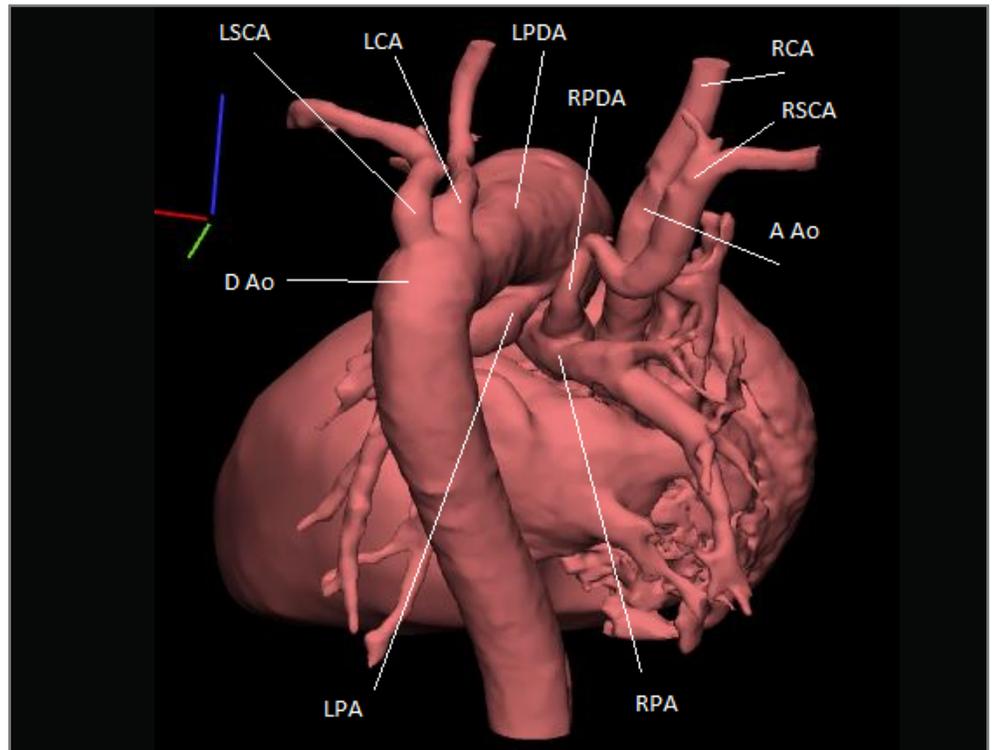


Figure 2. 3D Model Reconstruction. AA_o – Ascending Aorta. DA_o – Descending Aorta. LPA – Left Pulmonary Artery. RPA – Right Pulmonary Artery. LCA – Left Carotid Artery. RCA – Right Carotid Artery. LSCA – Left Subclavian Artery. RSCA – Right Subclavian Artery. LPDA – Left Patent Ductus Arteriosus. RPDA – Right Patent Ductus Arteriosus.

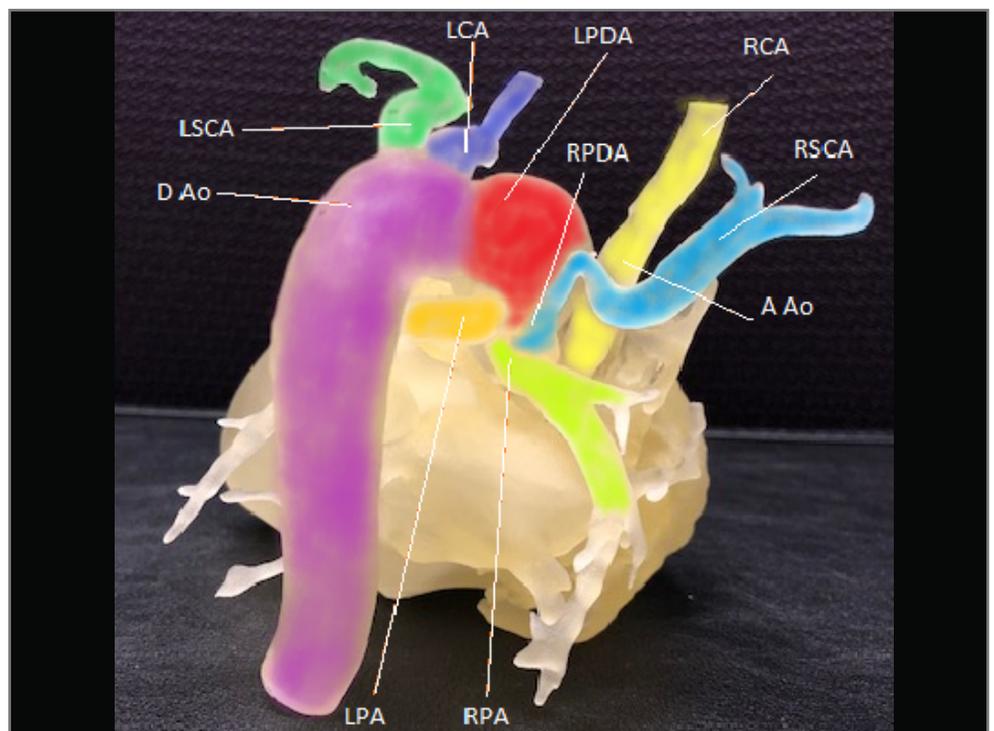


Figure 3. 3D Model Reconstruction. AA_o – Ascending Aorta. DA_o – Descending Aorta. LPA – Left Pulmonary Artery. RPA – Right Pulmonary Artery. LCA – Left Carotid Artery. RCA – Right Carotid Artery. LSCA – Left Subclavian Artery. RSCA – Right Subclavian Artery. LPDA – Left Patent Ductus Arteriosus. RPDA – Right Patent Ductus Arteriosus.

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"The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

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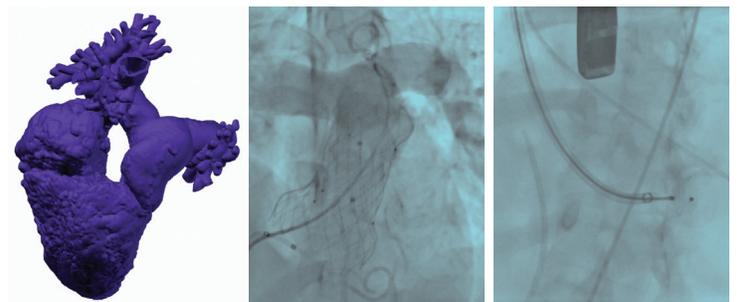
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The Heart Summit Reaches New Heights in Multi-Modality Medical Education

By Jeanne James, MD

The Heart Summit, held by the Herma Heart Institute at Children's Hospital of Wisconsin, is quickly becoming a must-attend event for pediatric cardiologists, researchers and surgeons. Building from our experience with the second *Heart Summit* in October 2018, this year's event will be expanded to three days.

A Rare Opportunity

The 2018 *Heart Summit* had a clinical focus on Left Ventricular Outflow Tract Obstruction (LVOT). Attendees witnessed a surgical procedure live-streamed from the OR and presented in 3D. Lead surgeon Viktor Hraska, MD, PhD, Medical Director of Cardiothoracic Surgery and Surgical Director of the Herma Heart Institute, performed an operation to relieve supra-valvar aortic stenosis in a 4-year-old child. Throughout the procedure, he described his approach and responded to Heart Summit attendees' questions in real time.

Dr. Hraska is a champion of this style of interactive presentation, having developed a similar program when he was Cardiac Surgeon-in-Chief at the Pediatric Heart Center Sankt Augustin, Germany. The 3D video provides unparalleled visibility for attendees, and the dialogue between the surgical team and the audience enhances learning for all.

Learning Moments

A second intervention was also scheduled to be live-streamed from the cardiac catheterization laboratory. The interventional cardiology team, led by Dr. Susan Foerster, anticipated balloon valvuloplasty to relieve aortic stenosis judged to be severe by echocardiography. However, direct measurement of the aortic valve gradient at the beginning of the procedure demonstrated much less severe obstruction, so balloon intervention was not indicated. For participants, this change in plan provided a forum to discuss both the risk-to-benefit ratio of balloon valvuloplasty and indications for intervention. It was a further demonstration that plans can change in real-time, and that the best action is the one offering the best possible functional outcome for the patient.



Knowing that interventional plans can change in the catheterization laboratory, the interventional team had already selected a previously recorded valvuloplasty to demonstrate the procedure. All in all, the catheterization experience and ensuing discussions were "value added" to the *Heart Summit*.

While watching actual procedures was a major learning opportunity for attendees, there were additional presentations on the genetic and embryological origins of LVOT; diagnostic techniques, including 3D echocardiography; assessment of the "borderline" left ventricle and the effects of LVOT over the lifetime of a patient.



Looking Forward

The *2019 Heart Summit* will be held October 3–5 on the topic “Management of the Failing Fontan: A Look into the Future.” The Fontan procedure serves as definitive palliation for patients born with single ventricle physiology, but there is an inescapable multisystem burden on other organs in the second decade of life and beyond. Because contemporary single ventricle management now allows survival into adulthood of 85–90% of patients, pediatric

cardiologists will be treating more and more Fontan complications as we continue to accrue teenaged and older survivors.

We are pleased to have two nationally recognized experts on complications of the Fontan circulation join us from Children’s Hospital of Philadelphia for our *2019 Heart Summit*. Dr. Jack Rychik will present “The Fontan Conundrum (setting the stage and explaining the complex disease),” and Dr. Yoav Dori will present a lecture on the evaluation and intervention of abnormal lymphatic



channels using MRI and cardiac catheterization. In addition, Dr. Hraska will demonstrate live and in 3D a cutting-edge Fontan modification: lymphatic diversion surgery.

This year, we are adding a Single Ventricular Morphology and Imaging Course, to be co-directed by Robert Anderson, MD, PhD, FRCPath and Peter Frommelt, MD. Dr. Anderson is a world expert in embryological cardiac development and a professor at the Institute of Child Health, University College London. Dr. Frommelt is the Medical Director of the Herma Heart Institute Echocardiography Laboratory and Core Imaging Research Laboratory. Together, Drs. Anderson and Frommelt will explore the embryology of the univentricular heart, including isomerism. Didactic presentations will be supplemented with focused sessions on imaging complex anatomy and demonstrations of anatomic specimens.

We are very excited to include demonstrations of the Stanford Virtual Heart* by the Herma Heart Institute and Marquette Visualization Laboratory. The Virtual Heart is an innovative virtual reality tool that allows one to examine complex anatomy in 3D from both outside and within the heart, a significant leap forward in understanding Congenital Heart Disease.

Finally, poster sessions and workshops on a variety of topics will round out this year's *Heart Summit*.

The Children's Hospital of Wisconsin is committed to providing the best in patient care through research, innovation and education. We look forward to learning more from acknowledged leaders in the field to enhance treatment of patients with complex single ventricle cardiac disease across the United States and worldwide.

Some travel awards are available to defray costs of attending the conference. For more information about the 2019 *Heart Summit*, please visit chw.org/theheartsummit.

Disclaimer

The Stanford Virtual Heart was created by Lighthouse Inc. with the support of The Betty Irene Moore Children's Heart Center at Stanford and Oculus VR (a Facebook subsidiary).



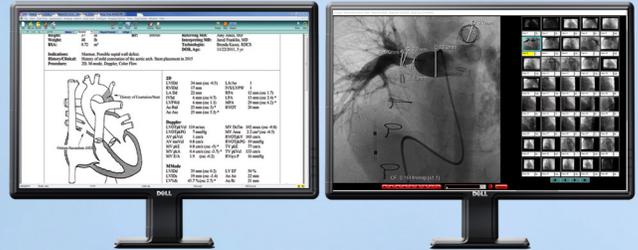
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Medical News, Products & Information

Compiled and Reviewed by Kate Baldwin and Tony Carlson

Bohr Scientists Figure Out How to Measure Electrical Activity in a Fetal Heart

Faculty of Science - University of Copenhagen: Electrodes are placed on the patient's chest area to record cardiac electrical activity - e.g. to determine whether the heart rhythm is so irregular that treatment is required; a type of medical examination for which ECG serves well as a diagnostic tool.

Not quite so when it comes to examining fetal cardiac electrical activity - for the obvious reason that it is impossible to place electrodes on a fetus's chest area, which makes ECG a no-go in this context. Instead, doctors will typically try to get an impression of the cardiac electrical activity by conducting an ultrasound scan; which, however, will not provide precise answers as to what is wrong should the heart e.g. be beating too fast or too slow.

In a foreseeable future these problems regarding the examination of fetal cardiac electrical activity are about to be solved - thanks to the joint effort of two groups of scientists from University of Copenhagen: from Quantum Optics (Quantop) at the Niels Bohr Institute (NBI) and from Department of Biomedical Sciences, respectively.

In a research paper - which the two groups have just published in the journal *Scientific Reports* - they describe an experiment which demonstrates that it is indeed possible to get a detailed read-out of fetal cardiac electrical activity; that is, if you ally yourself with a cloud of caesium atoms locked up in a hermetically closed glass cell.

"Our next challenge will be to incorporate this technique in a diagnostic sensor - and that is doable," says Assistant Professor Kasper Jensen, Quantop.

Kasper Jensen and Professor Eugene Polzik, head of Quantop, have been in charge of the experiment as far as tests conducted via the locked-up caesium atoms go, while Bo Hjorth Bentzen, Associate Professor at Department of Biomedical Sciences, has coordinated the biological part of the experiment that includes the use of isolated guinea-pig hearts.

Clouds of Atoms

The locked up cloud of caesium atoms is the cornerstone of a technique tailored for observations and measurements which Eugene Polzik and his team at Quantop have refined over a number of years - and applied to a number of tasks.

Put simply, the technique allows extremely precise observations and measurements at quantum level - if laser light at certain wavelengths are transmitted through the locked-up atom cloud. One project, which Quantop is currently involved in, thus aims at boosting the capacity of gravitational wave detectors through the 'cloud of atoms-principle'.

"The locked-up caesium atoms are capable of detecting very small magnetic fields. That is the reason why we also started to study this technique as a possible way of measuring fetal cardiac electrical activity - through the pregnant woman's belly. And our experiments demonstrate that this is indeed possible - which we also conclude in our article in *Scientific Reports*", says Kasper Jensen.

Guinea-Pig Hearts

In order to conduct the experiments, the Quantop-scientists needed hearts which they could measure - and these hearts were provided by associate professor Bo Hjorth Bentzen and his team at Department of Biomedical Sciences.

They chose guinea-pig hearts which are similar in size to that of a human fetus at the gestational age of approximately 20 weeks - and in a number of other respects also are well-suited for these kinds of

Archiving Working Group



A committee of the International Society of Nomenclatures for Pediatric and Congenital Heart Disease (ISNPCHD).

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

www.ipccc-awg.net

experiments, says Bo Hjorth Bentzen, who specializes in heart rhythm analysis. "Guinea-pigs have a heart rhythm fairly close to that of a human fetus, and a number of the proteins which regulate heart functions in guinea-pigs resemble the corresponding proteins in humans."

During the experiment, the scientists at Department of Biomedical Sciences euthanized a total of six guinea-pigs - in accordance with protocols approved by the Danish Veterinary and Food Administration. The hearts were surgically removed from the animals, cooled down, then transported to the Quantop lab at NBI just a few hundred meters away.

At Quantop the hearts were gradually warmed up to body temperature and subsequently placed in a Plexiglas chamber with a constant supply of oxygen and water in the form of a salty solution. This environment made the guinea-pig hearts start beating - which they would typically do for the next three-to-four hours.

The equipment was placed behind a magnetic shield in order to keep all outside electromagnetic activity away - and while the heart was beating, the scientists measured the electrical activity from the organ through the Plexiglas wall.

By measuring in this fashion - at a distance of approximately one centimeter and without attaching electrodes to the heart - the Quantop scientists mimicked a situation where fetal cardiac electrical activity is recorded via an instrument placed directly on top of the pregnant woman's belly.

In order to show that the equipment is capable of detecting electrical signals stemming from heart problems, the team of scientists from Department of Biomedical Sciences added a chemical to the salty solution that was continuously pumped into the Plexiglas chamber. This chemical changes the electrical signal in the heart - (triggering a reaction similar to what is seen in association with Long QT Syndrome, a hereditary heart condition) - which the system was also fully able to detect.

Future Treatment

New equipment which can conduct ECG-examinations of fetuses based on the NBI-method could have a significant impact on future treatment, says Niels Vejlstup, MD, PhD, and a specialist in treatment of fetal heart problems at Department of Cardiology at Rigshospitalet in Copenhagen. "Such equipment could make a difference in relation to e.g. AV-block - a rare condition which blocks certain electrical pulses in the heart. AV-block can develop in a fetus if the mother suffers from Lupus or Sjogren's Disease and, if doctors suspect that a fetus is developing AV-block, they will start treating the mother medically in an attempt to protect the fetus. However, at present we only have one option when it comes to evaluating how severely damaged a fetus's heart conduction system actually is - namely, doing an ultrasound scan. This method is encumbered with uncertainty - which is not the case when you conduct a direct measurement of fetal cardiac electrical activity," says Dr. Vejlstup.

Rigshospitalet is keen on participating in clinical trials in order to develop the new method, says Niels Vejlstup, adding that the method will be equally beneficial when it comes to diagnosing all other types of fetal heart rhythm disturbances.

At Room Temperature

Around the world, groups of scientists are developing advanced measuring systems - in some cases based on superconductors or on the use of rubidium, a chemical element. These methods, however, require extreme temperatures - close to absolute zero at -273.15 C, or in the vicinity of +200 C.

"In both cases the temperature bars the technique from 'just' being incorporated in equipment designed to detect e.g. fetal heart rhythm. Our equipment, on the other hand, operates at room temperature which is an advantage in this context. We estimate that within three years doctors can start using our equipment to measure fetal cardiac electrical activity," says Kasper Jensen.

The principle behind the method will also be applicable to other forms of biological

registrations and examinations, he says: "E.g. measuring brain activity when looking for signs of epilepsy."

Kawasaki Disease: One Disease, Multiple Triggers

Recent Clustering of Kawasaki Disease in San Diego Points to Environmental Causes

University of California - San Diego: Researchers at University of California San Diego School of Medicine, Scripps Institution of Oceanography, and international collaborators have evidence that Kawasaki Disease (KD) does not have a single cause. By studying weather patterns and geographical distributions of patients in San Diego, the research team determined that this inflammatory disease likely has multiple environmental triggers influenced by a combination of temperature, precipitation and wind patterns. Results were published in the November 12, 2018 online edition of Scientific Reports.

"We are seeing firsthand evidence of these weather patterns in San Diego, where eight children have recently been diagnosed with Kawasaki Disease. Recent low pressure systems in San Diego have been associated with two distinct clusters of the disease," said Jane C. Burns, MD, pediatrician at Rady Children's Hospital-San Diego and director of the Kawasaki Disease Research Center at UC San Diego School of Medicine. "Our research is pointing towards an association between the large-scale environment, what's going on with our climate on a large scale, and the occurrence of these clusters."

Kawasaki Disease is the most common acquired heart disease in children. Untreated, roughly one-quarter of children with KD develop coronary artery aneurysms -- balloon-like bulges of heart vessels -- that may ultimately result in heart attacks, congestive heart failure or sudden death.

Burns and her team examined 1,164 cases of KD treated at Rady Children's Hospital over 15 years. Noticeable clusters of KD cases were often

associated with distinct atmospheric patterns that are suspected to transport or concentrate agents that result in KD. Days preceding and during the KD clusters exhibited higher than average atmospheric pressure and warmer conditions in Southern California, along with a high pressure feature south of the Aleutian Islands.

“For the first time, we have evidence that there is more than one trigger for Kawasaki Disease. Up until now, scientists have been looking for one ‘thing’ that triggers KD,” said Burns. “Now we see that there are distinct clusters of the disease with different patterns suggesting varying causes.”



Jane C. Burns, MD, Director of The Kawasaki Disease Research Center at UC San Diego School of Medicine

Gene expression analysis further revealed distinct groups of KD patients based on their gene expression pattern, and that the different groups were associated with certain clinical characteristics. “Our data suggest that one or more environmental triggers exist, and that episodic exposures are influenced at least in part by regional weather conditions. We propose that characterization of the environmental factors that trigger KD in genetically susceptible children should focus on aerosols inhaled by patients who share

common disease characteristics,” said Burns who has studied KD for more than 35 years.

Although KD is estimated to affect fewer than 6,000 children in the U.S. each year, the incidence is rising in San Diego County. While the average incidence per 100,000 children less than five years of age residing in San Diego County was approximately 10 for the decade of the 1990s, the estimate from 2006 to 2015 was 25.5. This increase may be attributed to the efforts of the KD team at Rady Children’s Hospital to teach local physicians how to diagnose KD. Or it may be due to increasing exposure to the environmental triggers of the disease.

Prevalence rates of KD are increasing among children in Asia. Japan has the highest incidence rate, with more than 16,000 new cases per year. One in every 60 boys and one in every 75 girls in Japan will develop KD during childhood.

Incidence rates in the U.S. are approximately 19 to 25 cases per 100,000 children under age five -- but are higher in children of Asian descent. Predictive models estimate that by 2030, one in every 1,600 American adults will have been affected by the disease.

Study Finds Phone App Effectively Identifies Potentially Fatal Heart Attacks with the Near Accuracy of a Standard ECG

Can your smart phone determine if you’re having the most serious – and deadly – form of heart attack? A new research study says it can – and may be a valuable tool to save lives.

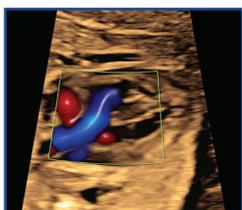
Newswise — The international study, led by researchers from the Intermountain Medical Center Heart Institute in Salt Lake City, found that a smartphone app to monitor heart activity and determine if someone is having an ST-Elevation Myocardial Infarction (STEMI), a heart attack in which the artery is completely blocked, has nearly the same accuracy as a standard 12-lead electrocardiogram (ECG), which is used to diagnose heart attacks.

Researchers say the findings are significant because the speed of treatment after a STEMI heart attack helps save lives.

“The sooner you can get the artery open, the better the patient is going to do. We found this app may dramatically speed things up and save your life,” said J. Brent Muhlestein, MD, lead investigator of the study and cardiovascular researcher at the Intermountain Medical Center Heart Institute.

In the study, 204 patients with chest pain received both a standard 12-lead ECG and an ECG through the AliveCor app, which is administered through a smartphone with a two-wire attachment. Researchers found the app with the wire set-up effective in distinguishing STEMI from non-STEMI ECGs accurately and with high sensitivity compared to a traditional 12-lead ECG.

“We found the app helped us diagnose heart attacks very effectively — and it didn’t indicate the presence of a heart attack when one wasn’t occurring,” Dr. Muhlestein said.



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A STEMI is a very serious type of heart attack during which one of the heart's major arteries — which supplies oxygen and nutrient-rich blood to the heart muscle — is blocked. ST-segment elevation is an abnormality that's detectable on the 12-lead ECG.

Researchers presented results from the study at the American Heart Association's 2018 Scientific Session in Chicago.

Researchers conducted the study, called the ST LEUIS International Multicenter Study, at five international sites that are all associated with the Duke University Cooperative Cardiovascular Society (DUCCS), with Intermountain Medical Center Heart Institute serving as the coordinating institution where they collected and collated data.

Other participating research centers include: Duke University, Integris Heart Hospital (Oklahoma City), Catholic University (Argentina), Mayo Clinic, Stanford University, Erlanger Institute for Clinical Research (Tennessee), University of Utah Health, AliveCor Corporation, and the Rocky Mountain University of Health Professions.

The idea for this kind of ECG set-up perhaps came from the use of treadmills for personal fitness development, said Dr. Muhlestein. Many people using treadmills wear a simple device that can detect their heart rate, through a single ECG lead,

more accurate than just checking the pulse. "It's a simple jump from there to putting it on a smartphone, and then recording the same ECG lead from several body positions," he said.

The new Apple 4 smartwatch also comes with a single-lead ECG. A typical ECG has 12 leads, which improves the accuracy of a diagnosis because heart attacks happen in different parts of the heart, and each lead looks at a different part. With the AliveCor app, the two wire leads are moved around the body in order to record all 12 parts.

The findings of the study are important for two reasons, said Dr. Muhlestein. The first is it could speed up the urgent treatment a patient needs after suffering a STEMI. American College of Cardiology/American Heart Association guidelines recommend that the "door-to-balloon time" — or the time from when a patient enters the hospital to when a catheter with a balloon on the tip is inserted into the patient's blocked artery, then inflated to flatten plaque against the wall of the artery — be less than 90 minutes.

"If somebody gets chest pain and they haven't ever had chest pain before, they might think it's just a bug or it's gas and they won't go to the emergency room," Dr. Muhlestein said. "That's dangerous, because the faster we open the blocked artery, the better the patient's outcome will be."

The app can take the electrocardiogram on the spot, send the results into the cloud where a cardiologist reviews it immediately and, if a STEMI is found, tell the person so they can be rushed to the hospital.

Secondly, the price of the app with the two-wire extension is low, which could put the power of an ECG into the hands of anyone with a smartphone or smartwatch, and make ECGs accessible in places like third world countries where people have smartphones but where expensive ECG machines are hard to find, if they're available at all.

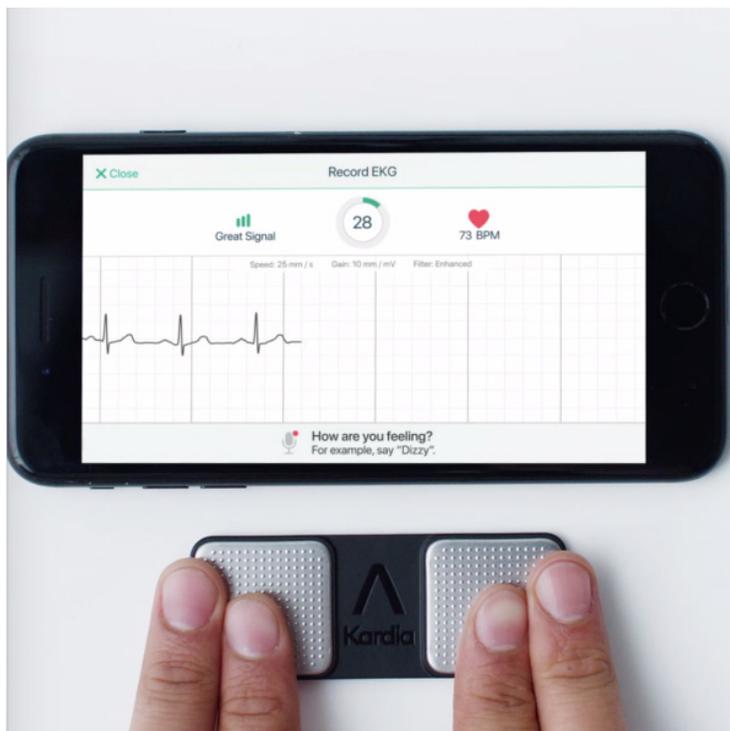


Photo credit: Intermountain Medical Center Heart Institute.



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