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

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Rare Association of Retroaortic Innominate Vein and Coarctation of Aorta with Hypoplastic Left Aortic Arch: Embryology Review of Retroaortic Innominate Vein

Siddharth Patel, MD; Pooja Makhija, MD; Steffan Sernich, MD

Introduction

The retroaortic course of the left innominate vein is a rare entity which can be misinterpreted during echocardiography for other abnormal vascular structures under the aortic arch. We reported the case of a 2-week-old female infant whose echocardiogram showed coarctation of the aorta and 2 vascular structures beneath the hypoplastic left aortic arch, one of which was traced to be a retroaortic left innominate vein. Right aortic arch is a common association.1 More than 80% of the patients with anomalous retroaortic left innominate vein have obstruction of the right ventricular outflow tract (Tetralogy of Fallot).[2] However, our patient has a left sided aortic arch with hypoplastic transverse arch and juxtaductal coarctation of aorta without any right ventricular outflow tract obstruction.

Case

A newborn girl with prenatal diagnosis of hypoplastic aortic valve and hypoplastic aortic arch on fetal echocardiogram was delivered at a referring hospital. Her initial postnatal echocardiogram was read as normal for age with a patent ductus arteriosus; the patient was discharged home with a cardiology follow-up appointment. However, after missing her appointment, she presented to the Emergency Department in cardiogenic shock at 2 weeks of age. She was hypotensive, tachycardic and tachypneic with cold extremities on presentation. Pulses were diminished in lower extremities with poor perfusion. She was intubated and started on mechanical ventilation due to worsening respiratory distress. Coarctation of aorta was suspected based on physical exam. Her initial echocardiography showed juxtaductal Coarctation of the aorta, tiny Patent Ductus Arteriosus (PDA) and mildly impaired left ventricular function. The patient was immediately started on intravenous prostaglandin E1, inotropic support, and metabolic acidosis was corrected.

Once patient was stabilized, a repeat complete echocardiogram demonstrated bicuspid aortic valve, juxtaductal coarctation of the aorta, patent ductus arteriosus, hypoplastic left aortic arch and two retroaortic vessels; the upper vessel being smaller in size was identified as the left innominate vein (Figure 1). Typically, there is only one vessel under the aortic arch, the right pulmonary artery. The differential diagnosis of an additional vessel in the suprasternal windows include an aorto-pulmonary collateral, a PDA arising from aortic arch, persistent fifth aortic

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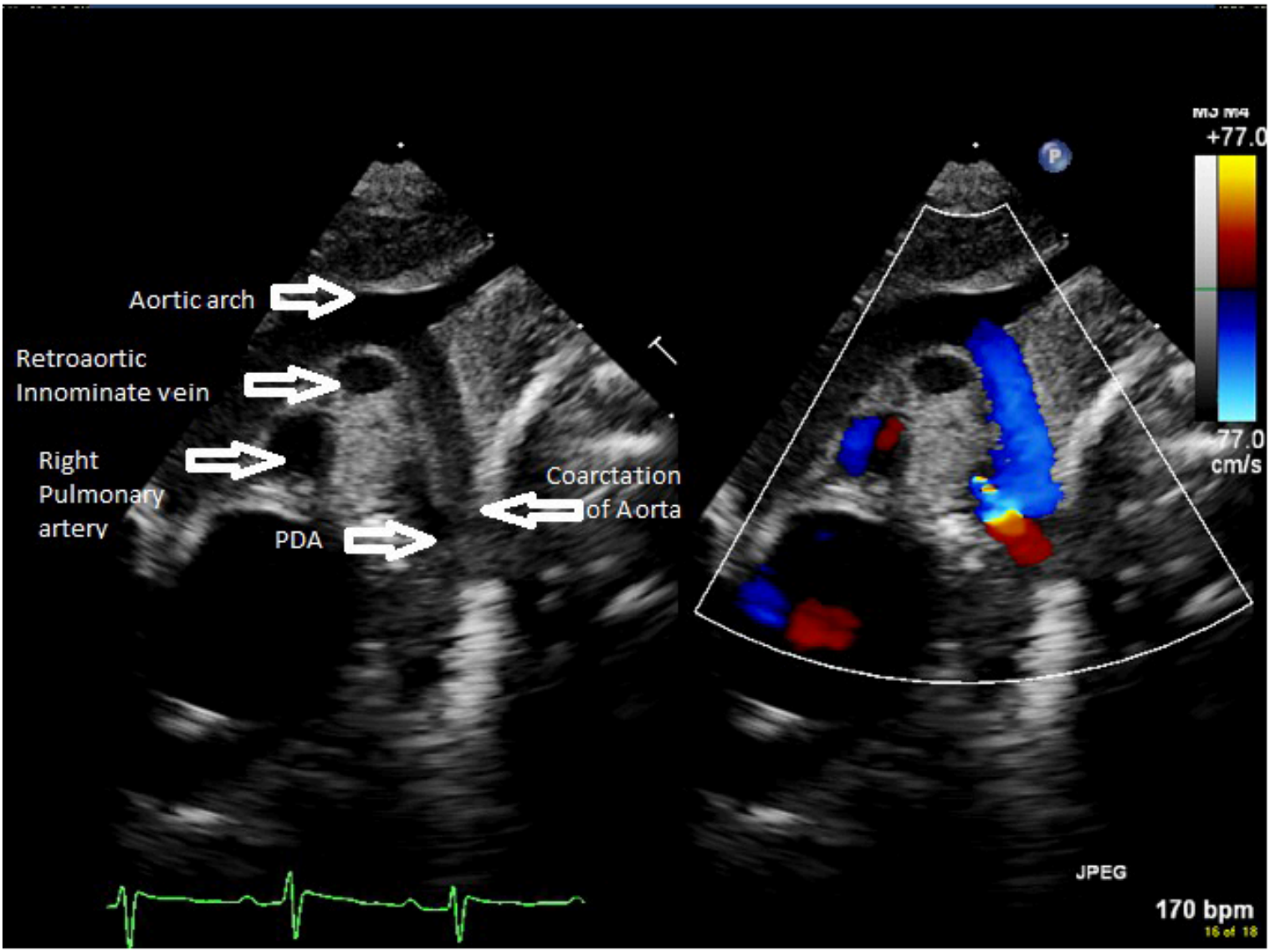

Figure 1. Suprasternal view shows retroaortic left innominate vein, hypoplastic arch, coarctation of aorta and patent ductus arteriosus.

arch, the vertical vein in supracardiac-type of Total Anomalous Pulmonary Venous Connection (TAPVC), and venous structures such as a retro-aortic innominate vein. Doppler interrogation demonstrated continuous low velocity pattern suggestive of venous structure. The probe was tilted leftward and anteriorly to trace the vessel which revealed venous flow from the left side of the neck passing below the aorta, directed to the right side and draining into the superior vena cava (Figure 2), confirming the diagnosis of retro-aortic left innominate vein.

The patient underwent a computerized topographic angiogram to assess the aortic arch which showed diffuse left aortic arch hypoplasia (3 mm) beyond the origin of the innominate artery with mild focal discrete juxtaductal coarctation of the aorta (2.5 mm) just distal to the left subclavian artery. Also, poststenotic dilation of the descending thoracic aorta (7 mm) and low origin of the left subclavian artery was noted (Figure 3).

The patient underwent coarctation of the aorta and hypoplastic aortic arch repair with bovine pericardium via a left lateral thoracotomy. The patient had an uneventful postoperative course.

Discussion

The left innominate vein normally courses obliquely downward to the right passing superoanterior to the aortic arch, and in front of its branches to drain into superior vena cava. Retro-aortic innominate vein is a rare entity where the innominate vein takes an anomalous course below the aortic arch with an incidence of 0.2-1.0% among congenital cardiac anomalies.[1] Right aortic arch is also a common association.1 More than 80% of the patients with anomalous retroaortic left innominate vein have obstruction of the right ventricular outflow tract (Tetralogy of Fallot).[2] However, our patient has a left-sided aortic arch with a hypoplastic transverse arch and juxtaductal coarctation of the aorta, without any right ventricular outflow tract obstruction. One such case is described in literature so far.[6]

Embryologically, as most of the left cardinal veins disappear, the venous drainage from the left side of the head and neck and the left arm is directed into the right anterior cardinal vein by 2 transverse capillary plexi above and below the fourth aortic arch. Normally, the aortic arch shortens during the embryological development and occupies the space of the inferior transverse capillary plexus, thus causing its regression. The rest of the venous blood shunts into the superior transverse capillary plexus and forms the normal supraortic course of the left innominate vein.[2] In contrast, reduced shortening of the aortic arch (right-sided or

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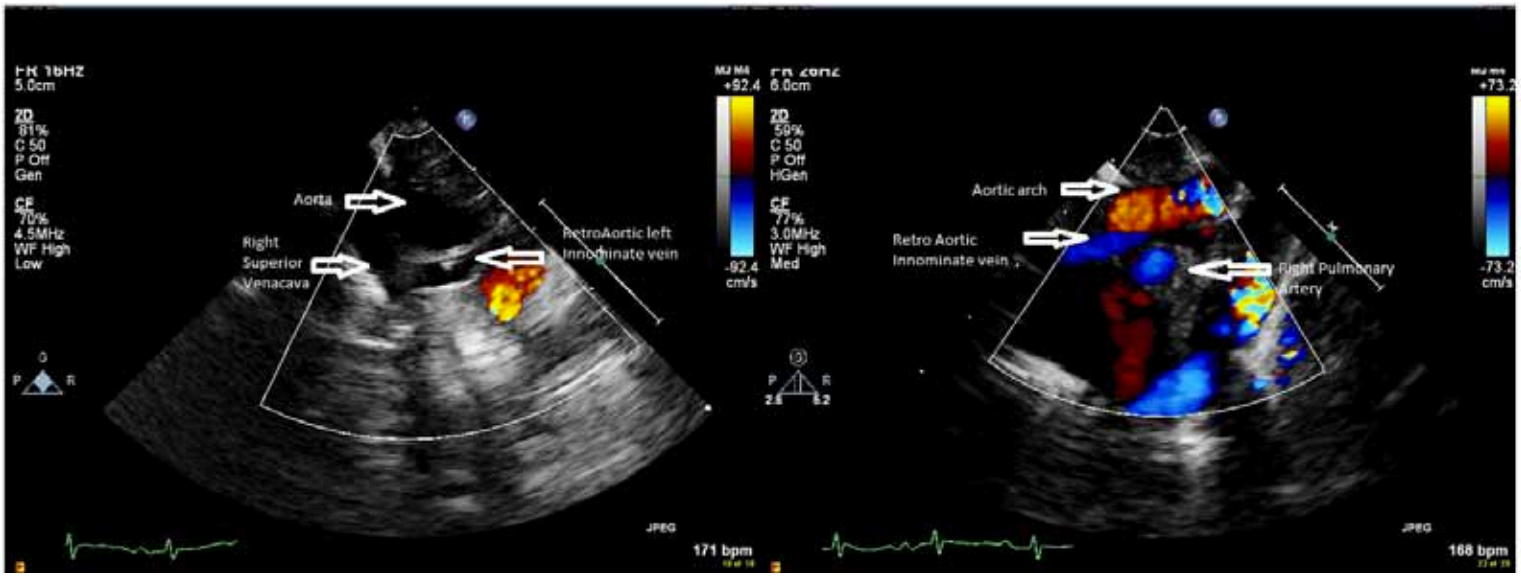

Figure 2. Suprasternal view showing retroaortic left innominate vein venous flow from the left side of the neck passing below the aorta to the right side.

high aortic arch) may cause regression of the superior capillary plexus and results in preservation of the inferior capillary plexus and formation of the retroaortic vein (Figure 4). Abnormal development of the pulmonary arteries (pulmonary atresia or pulmonary stenosis) encourages the sparing of the lower dorsal plexus, leading to formation of an anomalous innominate vein.[1]

The retroaortic innominate vein can usually be traced by echocardiography in the suprasternal long axis view to the left side of neck when the transducer is tilted leftwards and anteriorly, Doppler color flow and Doppler spectral analysis shows low velocity venous flow into the superior vena cava.[3] An aorto-pulmonary collateral and PDA arising from the aortic arch show a high velocity continuous flow.

Usually, the retroaortic innominate vein in isolation has no clinical importance. The anomalous innominate vein may cause technical difficulties during pacemaker insertion or central venous line placement through the left arm approach. For patients undergoing cardiac surgery, the superior vena caval cannulation for cardiopulmonary bypass has to be done more caudally than usual to avoid obstruction of the retroaortic innominate vein.[1] The anomaly may complicate exposure of the pulmonary arteries while creating systemic vein to pulmonary artery anastomosis during Glenn shunt. Also, it

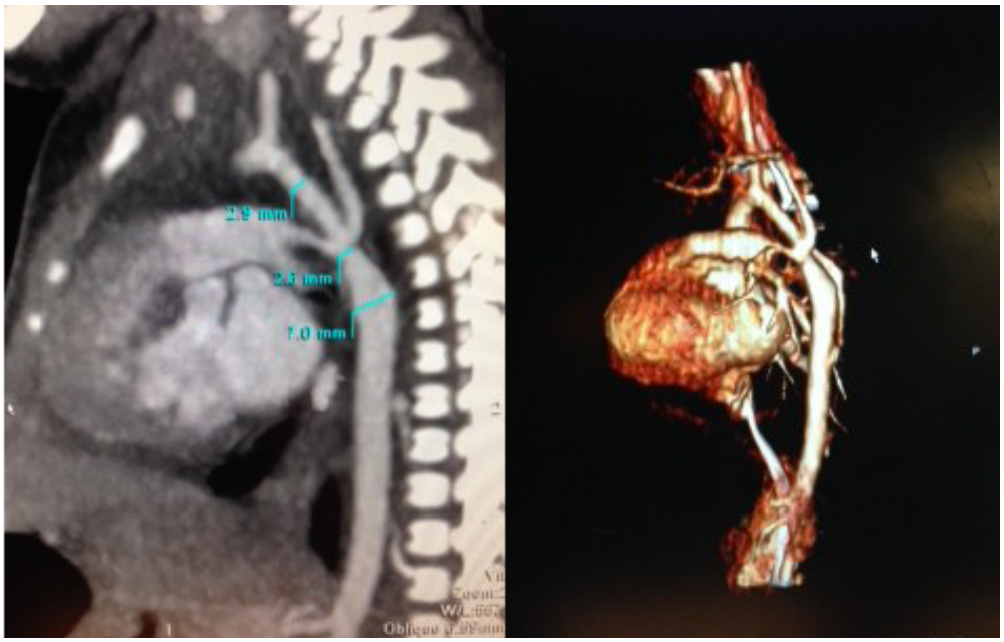

Figure 3. CT angiogram, Figure 3a-lateral view and Figure 3b-3D reconstruction showing hypoplastic left aortic arch, juxtaductal coarctation of aorta, patent ductus arteriosus and low origin of left subclavian artery just proximal to the coarctation site.

may obscure the surgical field in the construction of a subclavian to pulmonary artery shunt and the ligation of a patent ductus arteriosus.[4] Retroaortic innominate vein has been used in pulmonary artery reconstruction by creating a wide side-to-side anastomosis.[5]

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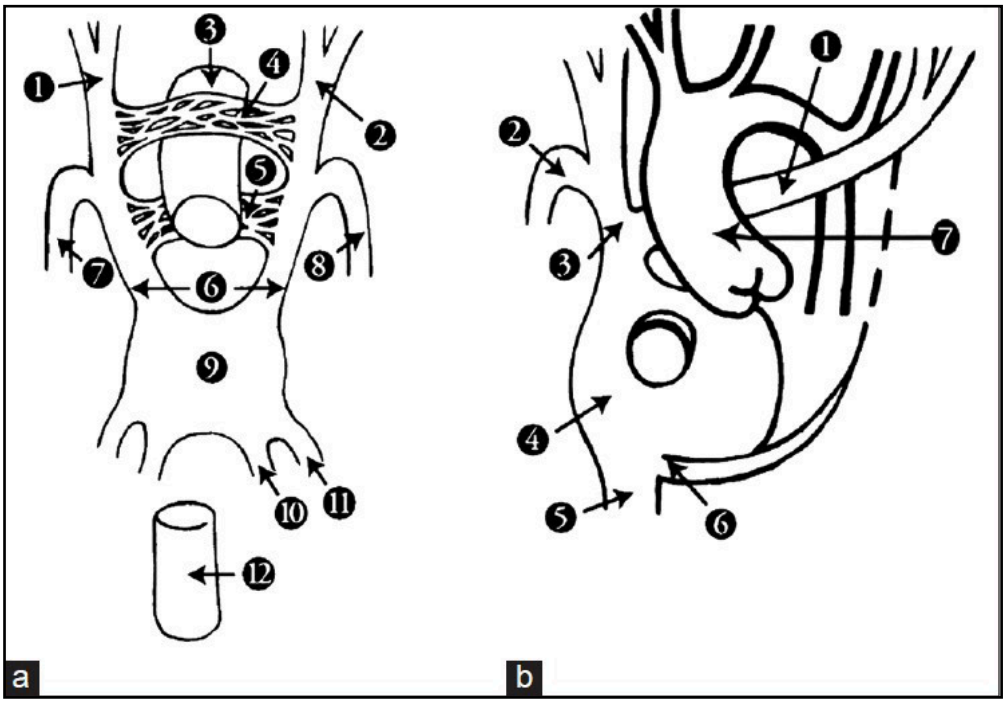

Figure 4. Schematic diagrams illustrating the development of retroaortic left innominate vein (RLIV) (a) In the sixth week of gestation: 1 - right anterior cardinal vein; 2 - left anterior cardinal vein; 3 - primitive aorta; 4 - superior transverse capillary plexus; 5 - inferior transverse capillary plexus; 6 - common cardinal veins; 7 - right posterior cardinal vein; 8 - left posterior cardinal vein; 9 - sinus venosus; 10 - vitelline vein; 11 - umbilical vein; 12 -developing inferior vena cava. (b) Retroaortic left innominate vein: 1 - innominate vein; 2 - azygos vein; 3 - superior vena cava; 4 - right atrium; 5 - inferior vena cava; 6 – coronary sinus; 7- ascending aorta.

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Letters to the Editor

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CHiP Network Update

By Gary Webb, MD

The basic idea of the CHiP Network (he Congenital Heart Professionals Network) is that congenital heart and pediatric cardiac professionals can subscribe to the service, indicating the type of information they would like to receive. This builds a list that is global, and that can be used by approved/appropriate organizations to communicate with professional members of this community. As a result, these organizations will be able to communicate with a larger readership. There is no charge for the service.

The CHiP Network has the capacity to send information only to certain types of professionals, in certain countries or cities, and to subscribers requesting messages in languages other than English. The potential of the system is still being explored. The CHiP Network is all-inclusive and is comprised of everyone who considers themselves a congenital heart or pediatric cardiac professional or administrator, including:

- Pediatric cardiologists
- ACHD cardiologists
- Cardiac surgeons
- RNs and APNs
- Physician assistants
- Cardiac care associates
- Trainees/fellows
- Administrators
- Social workers
- Psychologists and mental health professionals
- Researchers/scientists
- Transition medicine specialists
- Intensivists
- Anesthetists
- and Industry representatives.

This will make it possible for these various groups to communicate with people who identify themselves as being in the same profession. Of course, individuals in those groups will need to initiate the communications and consider how best to do this.

This will enable the circulation of various types of information:

- pending events
- *Journal Watch*
- recruitment opportunities

Geographic Distribution by Continent		
Continent	**#**	**%**
North America	1720	60%
Europe	420	15%
Asia-Pacific	335	12%
Middle East	171	6%
Central & South America	127	4%
Africa	106	4%

Profession Distribution		
Work Name Type	**#**	**%**
Cardiologist	1420	58%
Cardiac Care Associate / PA / RN	308	13%
Cardiac Surgeon	231	9%
Cardiac Anesthetist	120	5%
Nurse Practitioner	102	4%
Cardiac Intensivist	95	2%
Echo Technician	56	2%
Researcher / Scientist	54	2%
Administrator	49	2%
Industry Representative	28	1%

- research opportunities
- educational resource announcements
- news stories
- and professional society news reports and membership recruitment.

Undoubtedly, other applications will be developed.

To this point, the CHiP Network has circulated a monthly newsletter and a monthly *Journal Watch* service to its subscribers. The Journal Watch includes abstracts from the previous month for six different subspecialty areas: pediatric cardiology; interventional CHD; EP CHD; CHD surgery; fetal cardiology; and ACHD cardiology. The section editors identify the most important abstracts for the month, and offer a featured commentary on them. *Journal Watch* is housed on the ACHD Learning Center, so users can examine abstracts on a monthly basis going back 2-3 years.

CHiP's challenge, and the challenge of congenital heart professionals with an interest in the success of this resource, is to grow the network of subscribers and to

"The CHiP Network has the capacity to send information only to certain types of professionals, in certain countries or cities, and to subscribers requesting messages in languages other than English. The potential of the system is still being explored."

Table 3. Geographic Distribution by County Distribution							
Countries		**Countries**		**Countries**		**Countries**	
United States	1485	Malaysia	16	Greece	4	Zambia	2
Canada	240	Belgium	15	Poland	4	Libyan Arab Jamahiriya	2
United Kingdom	137	Denmark	14	Slovakia	4	Cameroon	2
Australia	94	China	14	Slovenia	4	Netherland Antilles	2
Saudi Arabia	58	Israel	13	Sudan	4	Bolivia	1
South Africa	50	Korea, Republic of	13	Ghana	4	Cuba	1
Brazil	46	Turkey	12	Ethiopia	4	El Salvador	1
India	46	Venezuela	10	Morocco	4	Panama	1
Netherlands	37	Colombia	10	Bangladesh	4	Guatemala	1
Iran, Islamic Republic of	28	United Arab Emirates	10	Unspecified	4	Ecuador	1
Sweden	28	France	10	Costa Rica	3	Syrian Arab Republic	1
Argentina	24	Pakistan	10	Trinidad and Tobago	3	Iceland	1
Egypt	24	Philippines	10	Iraq	3	Albania	1
Switzerland	24	Qatar	9	Oman	3	Malta	1
Japan	24	Hungary	9	Bulgaria	3	Croatia	1
Germany	21	Russian Federation	9	Finland	3	Senegal	1
New Zealand	21	Austria	7	Tanzania, United Republic of	3	Zimbabwe	1
Thailand	21	Chile	6	Singapore	3	Mozambique	1
Italy	20	Kenya	6	Nepal	3	Georgia	1
Norway	20	Lithuania	5	Mongolia	3	Uzbekistan	1
Indonesia	19	Ukraine	5	Jamaica	2	Azerbaijan	1
Nigeria	17	Portugal	5	Bahrain	2	French Polynesia	1
Taiwan	17	Uganda	5	Jordan	2	Brunei Darusssalam	1
Mexico	16	Hong Kong	5	Ireland	2	Bhutan	1
Spain	16	Kuwait	4	Cyprus	2		

"CHiP's challenge, and the challenge of congenital heart professionals with an interest in the success of this resource, is to grow the network of subscribers and to enhance the value of the service."

enhance the value of the service. CHiP has over 25 partner organizations. Each of these is asked to invite its members to join CHiP, and to show informational material about CHiP at their meetings.

To this point, we are grateful that financial support has been provided by Cincinnati Children's Hospital Heart Institute. We are also introducing a new category of partners, Institutional Partners. The first of these to be identified is Toronto's Hospital for Sick Children, with 58 subscribers to date.

Join CHiP - membership is free - www.chipnetwork.org

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A Case Series of Unfortunate Gerbode Defects

By Lucky R. Cuenza, MD; Anna Christina Adora,MD; Thessie Minelli O. Valdez, MD; Cheryl P. Fomaneg, MD; Efren R. Vicaldo, MD

Introduction

Gerbode Defect is a very rare congenital anomaly described as a communication between the left ventricle and the right atrium. It was named after Frank Gerbode, a surgeon from Stanford University who described his surgical findings in 1958. He reported two types (direct and indirect) depending if the defect lies above or below the tricuspid valve along with the presence of a true ventriculo-atrial communication.[1] The pathology may be due to a congenital defect, or can result from trauma, myocardial infarction, endocarditis or even aortic valve replacement.[2] It can appear as an isolated lesion, but it has been reported to accompany other congenital lesions. We describe 3 cases of patients who had Gerbode Defect along with other findings noted on echocardiography also with other findings not previously reported.

Case 1

A 31-year-old female came in due to frequent symptoms of palpitations of five months duration accompanied by dyspnea on exertion. Physical examination showed a blood pressure of 100/60 with a heart rate of 82 beats per minute. Auscultation of the chest revealed a 4/6 holosystolic murmur at the 4th intercostal space left sternal border with a thrill. Doppler studies showed a mosaic color flow display going from the left ventricle to the left atrium (Figure 1). This was confirmed by transesophageal echocardiogram (Figure 2). Peak systolic gradient between the left ventricle and the right atrium was measured at 89mmHg. There was no chamber dilatation noted. The patient was started on diuretics and eventually underwent surgical closure of the defect. She was discharged stable and improved.

Case 2

A 25-year-old female was referred to our institution for difficulty of breathing, exertional dyspnea and bipedal edema. Physical examination revealed a 4/6 holosystolic murmur at the 3rd left intercostal space. Doppler color flow on transthoracic echocardiogram showed an abnormal color flow display across the perimembranous portion of the interventricular septum during systole from the left ventricle to the right atrium. The defect was noted above the tricuspid valve (Figure 3). There was dilatation of the right atrium and the right pulmonary artery with moderate tricuspid regurgitation. There was also note of Patent Ductus Arteriosus (PDA) with a bidirectional shunt (Figure 4). She was started on Furosemide 40mg once a day which relieved her symptoms. She was initially scheduled for transesophageal echo and further evaluation, but was lost to follow-up.

Case 3

A 29-year-old female diagnosed with Congenital Heart Disease since 9-years-old complained of palpitations and easy fatigability a week postpartum. Physical

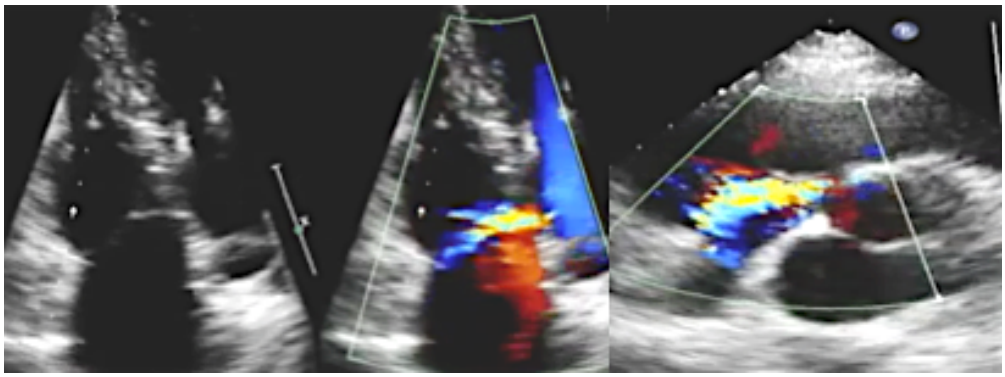


Figure 1. Transthoracic echocardiography apical four chamber view showing color flow directed towards the right atrium from the left ventricle (left and middle). Short axis view at the level of the aortic valve shows the jet situated above the tricuspid valve.

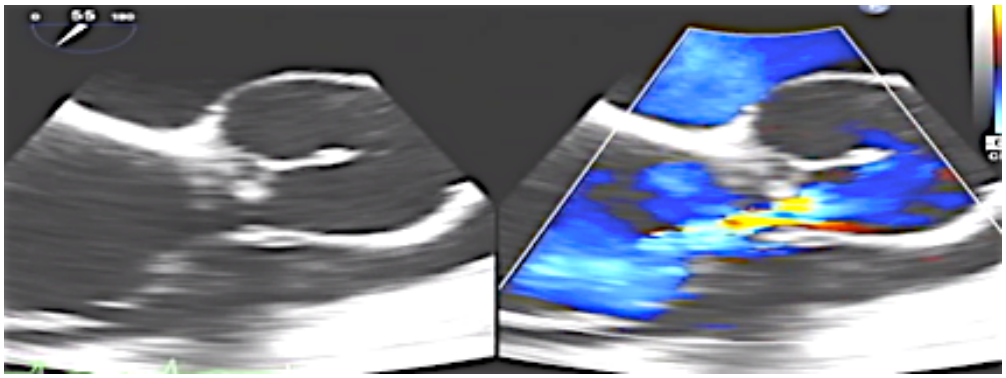


Figure 2. Transesophageal echocardiography mid esophageal short axis view showing an echo dropout at the ventriculoatrial septum which measures around 0.45cm (left). The flow is directed towards the right atrium from the left ventricle (right).

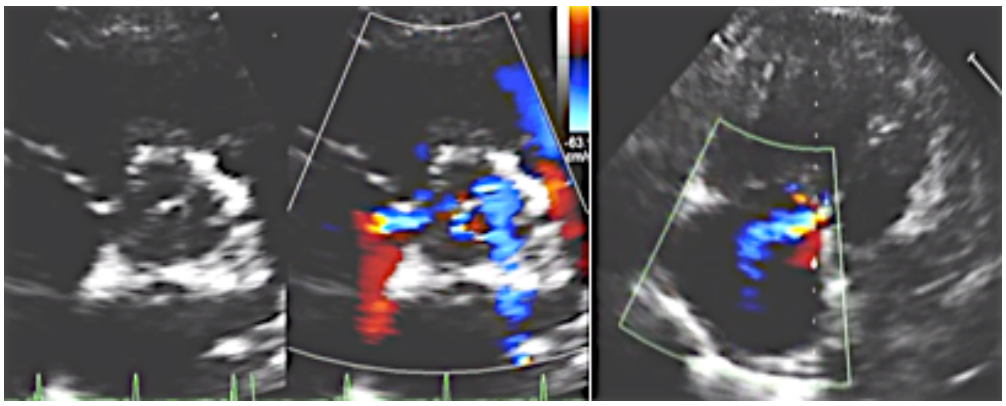


Figure 3. Transthoracic echocardiogram at the short axis view shows the ventriculoatrial communication on Color flow Doppler. Apical four chamber view shows a mosaic color flow from the left ventricle to the right atrium situated above the valve.

examination showed a blood pressure of 130/80 and tachypnea at 32 cycles per minute with an oxygen saturation of 95% at room air. She was tachypneic at 32 cycles per minute with an oxygen saturation of 95% at room air. She was tachycardic but with regular rhythm, with right ventricular heave, single S2, with 4/6 holosystolic murmur at the 2nd left intercostal space, (+) thrill, a 4/6 holosystolic murmur at the 4th left intercostal space and positive for Carvallo's sign. There was electrocardiographic and x-ray evidence of right atrial enlargement and right ventricular hypertrophy. There was a suspicious mosaic color flow during systole from the left ventricle to the right atrium on transthoracic echocardiogram. Two dimensional and 3D transesophageal echocardiogram confirmed the findings of a direct Gerbode Defect above the tricuspid valve (Figure 5). Transthoracic echocardiogram showed a Patent Ductus Arteriosus (Figure 6) with severe pulmonary hypertension. The left atrium, right atrium, main pulmonary artery and right ventricular had signs of volume and pressure overload and there was mitral valve prolapsed with moderate regurgitation. Additional findings included a mild valvar pulmonic stenosis (Figure 7) with a peak gradient of 20 mmHg. A flap-like structure was likewise noted at the interatrial septum suggestive of Patent Foramen Ovale (PFO). Contrast study demonstrated passage of microbubbles from the RA to the LV and LA and from the PA to the descending aorta indicative of the presence of Gerbode defect, PFO and PDA, respectively.

Discussion

Gerbode defects are very rare abnormalities with an incidence of 0.08% of congenital defects.[3] It is primarily caused by an anatomic deficiency of the membranous septum which can be either congenital or acquired. Riemenschneider and Moss devised a classification[4] based on the anatomical relationship of the left ventricle to the right atrium shunt with the tricuspid valve. This anatomical arrangement allows the septal leaflet to divide the membranous ventricular septum into two portions: a supravalvular portion, and an infravalvular portion. A defect in the supravalvular portion results in a direct left ventricle to right atrium communication and a defect below the tricuspid valve represents a perimembranous ventricular septal defect and would normally result in a communication between the left ventricle and the right ventricle then through a defective tricuspid valve into the right atrium. This is termed as an indirect Gerbode Defect. The true (or direct) Gerbode defect is a true left ventricular to right atrial communication and is rarer than the indirect type.[5] All the patients in our series had the true or direct form. In both defects left ventricular to right atrium communication allows shunting of blood to the right atrium during systole. If the communication is large enough, the patient may become symptomatic, and may present with signs of volume overload.[6]

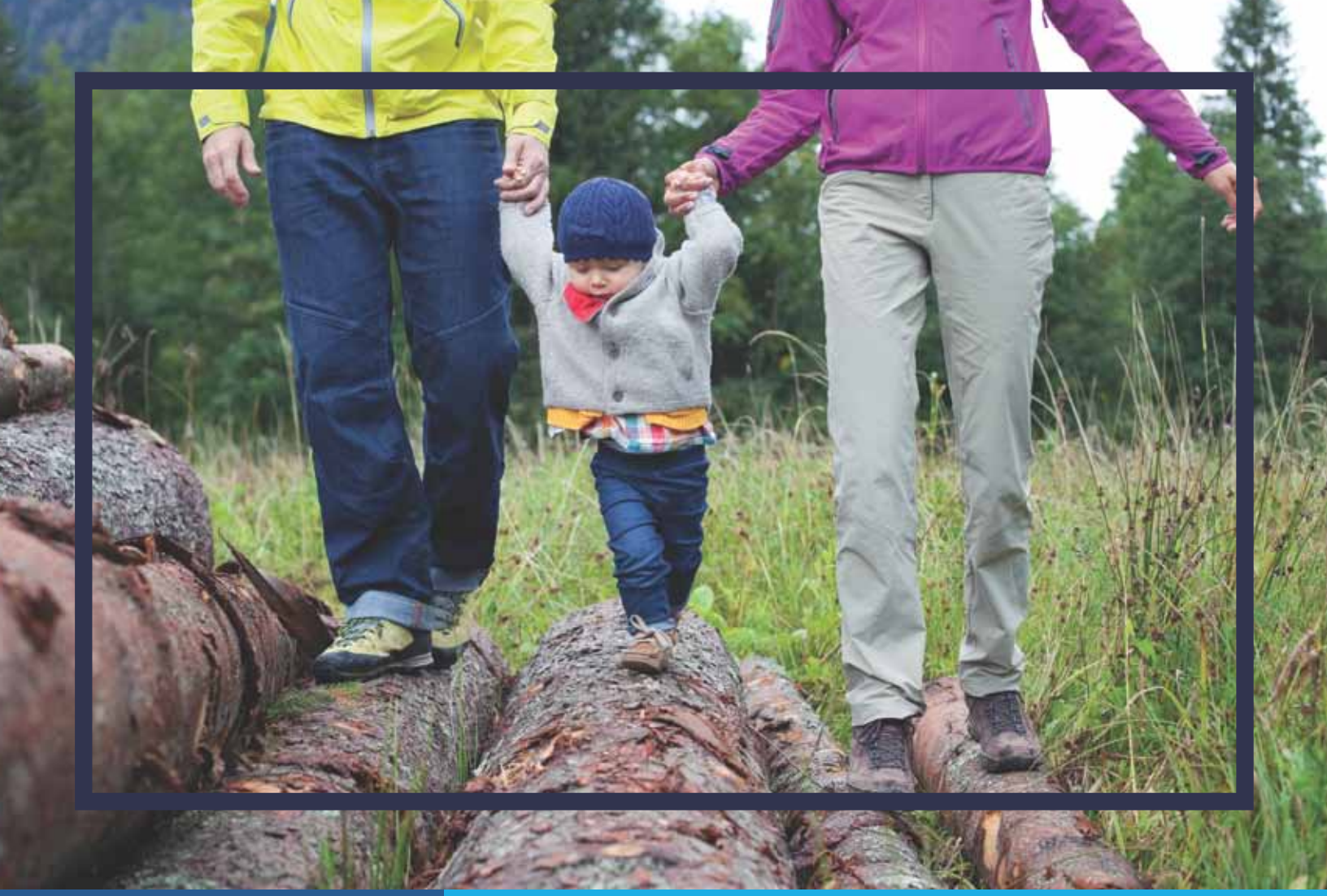
Typical findings are similar to that of a ventricular septal defect, with a harsh systolic murmur accompanied by a thrill. A disproportionate enlargement of the right atrium may be found on radiograph along with features of right atrial abnormality on electrocardiogram.[7] However, definitive diagnosis may not be possible based on these methods only. Two of the cases also presented with other shunt abnormalities, as well as varying degrees of pulmonary hypertension, making the diagnosis clinically challenging.

Echocardiography identifies the location of the defect and color flow imaging can identify the flow pattern from the left ventricle to the right atrium. It is also used to locate the relationship of the defect with respect to the tricuspid valve. Transesophageal echocardiography can reliably distinguish the two types and was also found to impact operative repair.[8] One of the hallmarks of Gerbode ventriculo-atrial defect is a high Doppler gradient between the left ventricle and the right atrium accompanied by right atrial dilation. The echocardiographer must be vigilant as the jet from the Gerbode Defect may be confused with a tricuspid regurgitant jet and can be mistakenly associated with pulmonary hypertension.[9] The presence of normal diastolic pulmonary arterial pressure using pulmonic regurgitation jet is very useful to distinguish the true pulmonary arterial hypertension from high velocity jet in the right atrium caused by Gerbode-type defect. Our second patient had a bidirectional shunt with severe pulmonary hypertension and the tricuspid regurgitation jet was delineated properly from the jet coming from the Gerbode Defect. Our third patient had multiple congenital heart defects. The Gerbode Defect was only suspected on transthoracic echo, while integrating clinical findings of right-sided volume overload. Transesophageal echocardiogram revealed the Gerbode Defect, to go along with the patent ductus arterosus, a patent foramen ovale and severe pulmonary hypertension. Other imaging modalities such as 3D transesophageal echocardiogram may add incremental diagnostic value by delineating the morphologic nature of the ventricular septal defect and its shape, and showing other structures in their realistic spatial distribution.[10] This may have implications in surgical planning.

Surgical repair is indicated for symptomatic patients with significant-sized shunts and is usually associated with favorable outcomes.[11] Gerbode defects may or may not be associated with other cardiac abnormalities. In the study by Anderson et al they noted concomitant congenital anomalies in six children. One patient had a right aortic arch, one patient had a patent ductus arteriosus, three patients had a left superior vena cava, two patients had an anomalous left hepatic vein.[12] In this series, the first patient had an isolated Gerbode Defect with good functional capacity. The second patient similarly had a patent ductus arteriosus with a bidirectional shunt as well as moderate tricuspid regurgitation and severe pulmonary hypertension. Our third patient had multiple congenital defects. Aside from the Gerbode Defect she also had a Patent Ductus Arteriosus with a PFO, with severe tricuspid regurgitation and mild supravalvar pulmonary stenosis. The findings from the latter two patients have not been previously described in literature. Mechanistically the shunt from the Gerbode Defect may contribute to more right-sided volume overload and pulmonary hypertension. The study by Anderson et al was done in children and had no reported operative or late mortality.[12] The outcomes in patients with multiple and advanced concomitant congenital heart disease is uncertain. We recommended surgical closure for our first patient and further work up (hemodynamic studies) on the latter two cases to determine operative prognosis.

Conclusion

Gerbode's Defect is a very rare congenital anomaly which can occur in isolation or may be associated with other congenital heart defects. It may complicate these conditions and even lead to misinterpreted diagnosis. We reported three cases in our institution,



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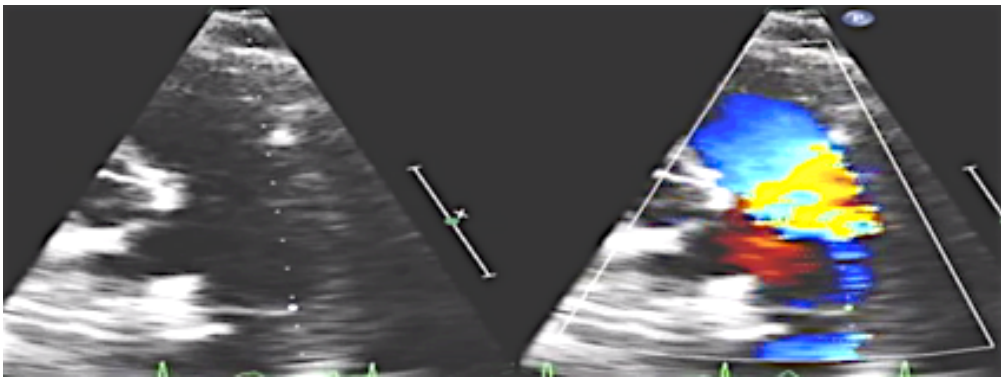

Figure 4. Transthoracic echocardiogram short axis view at the level of the pulmonary artery bifurcation tract showing a connection between the pulmonary artery and the aorta.

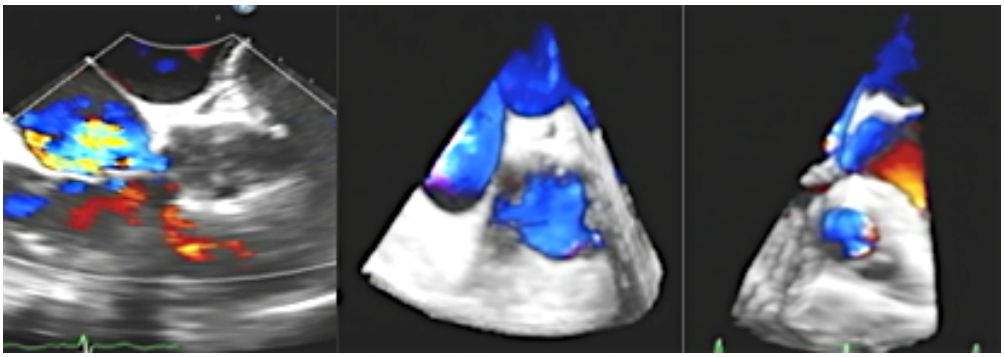

Figure 5. Transesophageal Echocardiography mid-esophageal short axis view showing an echo drop out at the ventriculoatrial septum between the LVOT and right atrium measuring 0.46 cm in diameter (left) with mosaic color flow display across the ventriculoatrial septum and tricuspid valve. 3D TEE mid-esophageal short axis view showing an echo drop out at the ventriculoatrial septum between the LVOT and right atrium with mosaic color flow display across the ventriculoatrial septum and tricuspid valve.

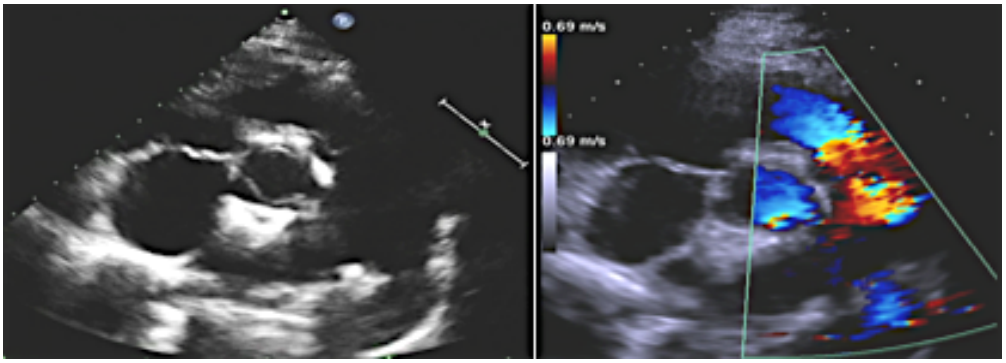

Figure 6. Transthoracic Echocardiography showing the pulmonic valve in parasternal short axis view with systolic doming motion (left) and continuous flow from the aorta to the pulmonary artery (right).

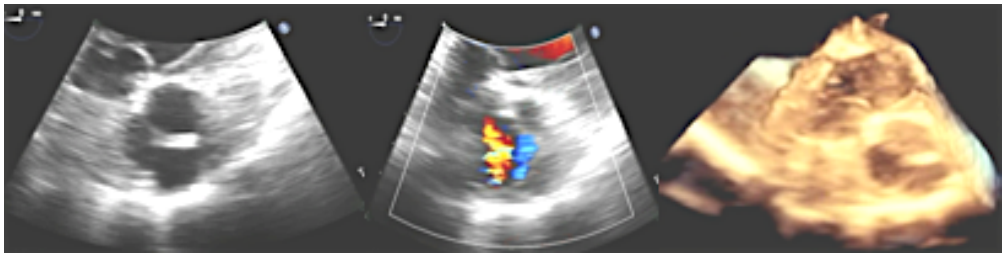

Figure 7. Transesophageal Echocardiography showing the pulmonic valve in mid-esophageal long axis view during diastole(left)and mosaic color flow display across the pulmonic valve during systole (middle). 3D TEE showing the pulmonic valve in mid-esophageal short axis view during systole (right).

which had a direct type of ventriculoatrial communication. These cases underlie the importance of integrating sound clinical assessment, as well as meticulous echocardiographic investigation in order to properly diagnose this unusual anomaly, facilitating surgical planning and determination of optimum management.

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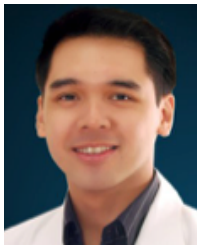

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2nd Annual Basic & Advanced Fetal Cardiac Symposium and Workshop
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This symposium will discuss various concepts and common CHD malformations encountered in the fetus, with emphasis on imaging and practical tips for diagnosis. In particular, it will include sessions focusing on the following areas:

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- Evaluation of Fetuses with Borderline Cardiac Findings
- Diagnosis and Management of Fetal Arrhythmia
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Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

The William Novick Global Cardiac Alliance

Announcing the William Novick Global Cardiac Alliance - a new non-profit charity launched in November 2014 by 18 professional colleagues formerly of the International Children's Heart Foundation (ICHF).

Prof. William Novick is a Pediatric Cardiothoracic Surgeon and Paul Nemir Professor of International Child Health at the University of Tennessee Health Science Center.

Prof. Novick and colleagues left the ICHF and launched the Novick Cardiac Alliance (NCA) as a new, completely separate entity after a 22 year experience operating on over 7500 children, and developing over 25 independent pediatric cardiac surgical programs in 5 continents. The NCA was founded with the continued goals of collaboratively operating on children with heart disease, educating teams in their own environment, innovating new solutions to complex problems and fostering sustainable solutions to pediatric cardiac care.

With this announcement, the NCA is extending an invitation to professionals in pediatric congenital cardiac care to volunteer for our 2 week cardiac teams. We need Cardiac Surgeons, Cardiologists (ECHO and Cath lab), Anesthesiologists, Perfusionists, Intensivists, PICU nurses and Respiratory Therapists. Volunteers are not required to pay for accommodation or travel. The NCA has 30 trips scheduled for 2015.

The NCA is by design non-sectarian and provides comprehensive care to all children with congenital or acquired heart disease regardless of gender, ethnicity, religion, political ideation, genetic factors or economic means.

Founding Board of the William Novick Global Cardiac Alliance are: William Novick, MD, MS; Brian Forsberg, MPH CCP; David Wieduwilt, BME CE; Elizabeth Novick, BSN/RN; Eugene Suslin, MD; Farzana Shah, RN; Frank Molloy, RN MSc; Humberto Rodriguez, MD; Igor Polivenok, MD, PhD; Jean Towne; Karen Bowtell, RN, PGDip; Kathleen Fenton, MD; Marcelo Cardarelli, MD, MPH; Martina Pavanic, BSN/RN; Pavel Shauchenka, MD; Siarhei Liauchonak, MD; Sri Rao, MD; and Stacey Marr, RN, MSc.

For more information got to: facebook.com/cardiacalliance or www.cardiac-alliance.org

Acute Kidney Injury can be a Serious Complication Following Heart Surgery

Newswise — The anti-inflammatory drug dexamethasone helps prevent serious kidney complications that can arise following heart surgery, according to the results of a randomized clinical trial. The findings, which appeared in an upcoming issue of the *Journal of the American Society of Nephrology* (JASN), could lead to a change in care for patients during cardiac operations.

Acute Kidney Injury (AKI) is one of the most devastating complications following cardiac surgery. Approximately 1% of patients undergoing cardiac surgery require dialysis to treat severe AKI that arises after surgery, and the incidence is higher among patients with pre-operative chronic kidney disease. These patients experience strikingly high death rates while in the hospital that exceed 40%. "One percent sounds like a small percentage; however, given the fact that each year, over half a million people undergo heart surgery in the United States alone; this means that an estimated 5000 patients develop renal failure, and of those about 2500 die as a result of this complication," said Kirolos Jacob, MD (University Medical Center, Utrecht, The Netherlands). He noted that these figures are rising due to the aging population.

Because heart surgery initiates an inflammatory reaction in the body that can have negative effects on the kidneys, Dr. Jacob and his colleagues wondered whether giving patients dexamethasone, an anti-inflammatory drug, could decrease the risk of severe AKI following cardiac surgery. The team analyzed the results of a large randomized controlled trial called the Dutch Dexamethasone for Cardiac Surgery (DECS) trial, which included 4465 patients undergoing cardiac surgery who were randomized to receive placebo or dexamethasone during surgery. The original trial tested whether dexamethasone could reduce the risk of a variety of major postoperative complications. In this analysis, the investigators specifically examined kidney failure and focused on the most severe form: AKI requiring dialysis.

Dexamethasone appeared to protect against the development of severe AKI. Patients who received the drug had about a 2.5-times lower risk of developing AKI requiring dialysis compared with those receiving a placebo.

"The beneficial effects of dexamethasone were particularly present in those who already had pre-existing kidney disease before heart surgery," said Dr. Jacob. "This reinforces the fact that this drug could be of major importance for the increasing elderly population with pre-existing kidney disease undergoing a heart operation."

The study is the largest randomized, placebo-controlled trial showing a potential benefit of any therapeutic drug for the prevention of severe kidney injury following heart surgery. A single dose of dexamethasone during a heart operation is inexpensive, straightforward, painless, and safe for patients. "These advantages make the intervention very accessible and cost-effective, especially since the costs for dialysis are very high," said Dr. Jacob.
Study co-authors include David Leaf, MD, MMSc, Jan Dieleman, MD, Diederik van Dijk, MD, PhD, Arno Nierich, MD, PhD, Peter Rosseel, MD, PhD, Joost van der Maaten, MD, PhD, Jan Hofland, MD, PhD, Jan Diephuis, MD, Fellery de Lange, MD, PhD, Christine Boer, PhD, Jolanda Kluin, MD, PhD, Sushrut Waikar, MD, MPH, for the Dexamethasone for Cardiac Surgery (DECS) Study Group.

Disclosures: The DECS study was sponsored by grants from the Netherlands Organisation for Health Research and Development (ZonMw) and the Dutch Heart Foundation.

The article, entitled "Intraoperative High-Dose Dexamethasone and Severe Acute Kidney Injury after Cardiac Surgery," appeared online at http://jasn.asnjournals.org/ on May 7, 2015.

Founded in 1966, and with more than 15,000 members, the American Society of Nephrology (ASN) leads the fight against kidney disease by educating health professionals, sharing new knowledge, advancing research, and advocating the highest quality care for patients.

Hospital to Create Emergency Experiences Using Virtual Reality: Walk Around Inside Crisis Situations, Educate Medical Professionals

Next Galaxy Corp (OTC: NXGA) recently announced the signing of an agreement with Miami Children's Hospital. Next Galaxy will develop immersive Virtual Reality medical instructional content for patient and medical professional education using the Company's VR Model. Per the multi-year agreement, Next Galaxy and Miami Children's Hospital are jointly creating VR Instructionals on cardiopulmonary resuscitation (CPR) and other lifesaving procedures, which will be released as an application for smartphones.

Incorporating eye gaze control, gestures, and voice commands while "walking around" inside an emergency medical experience or crisis, Next Galaxy's Virtual Reality Model engages participants far beyond today's methodology of passively watching video and taking written tests.

“Assessments are incorporated directly into the medical VR models. We will design situations where participants are required to make the appropriate decisions about proper techniques. The Virtual CPR instructional will measure metrics and provide real-time feedback ensuring participants accurately perform CPR techniques. Further, the instructional will explain any mistake and prompt users to try again when errors are made. Supportive messages are delivered upon success,” states Mary Spio CEO, Next Galaxy Corp.

The medical VR models will be viewable through smartphones and desktops as 3D, and via VR devices such as Google Cardboard, VRONE and Oculus Rift.

For further information, visit www.nextgalaxycorp.com.

Higher Activity Levels Increase Survivability of ICD Patients

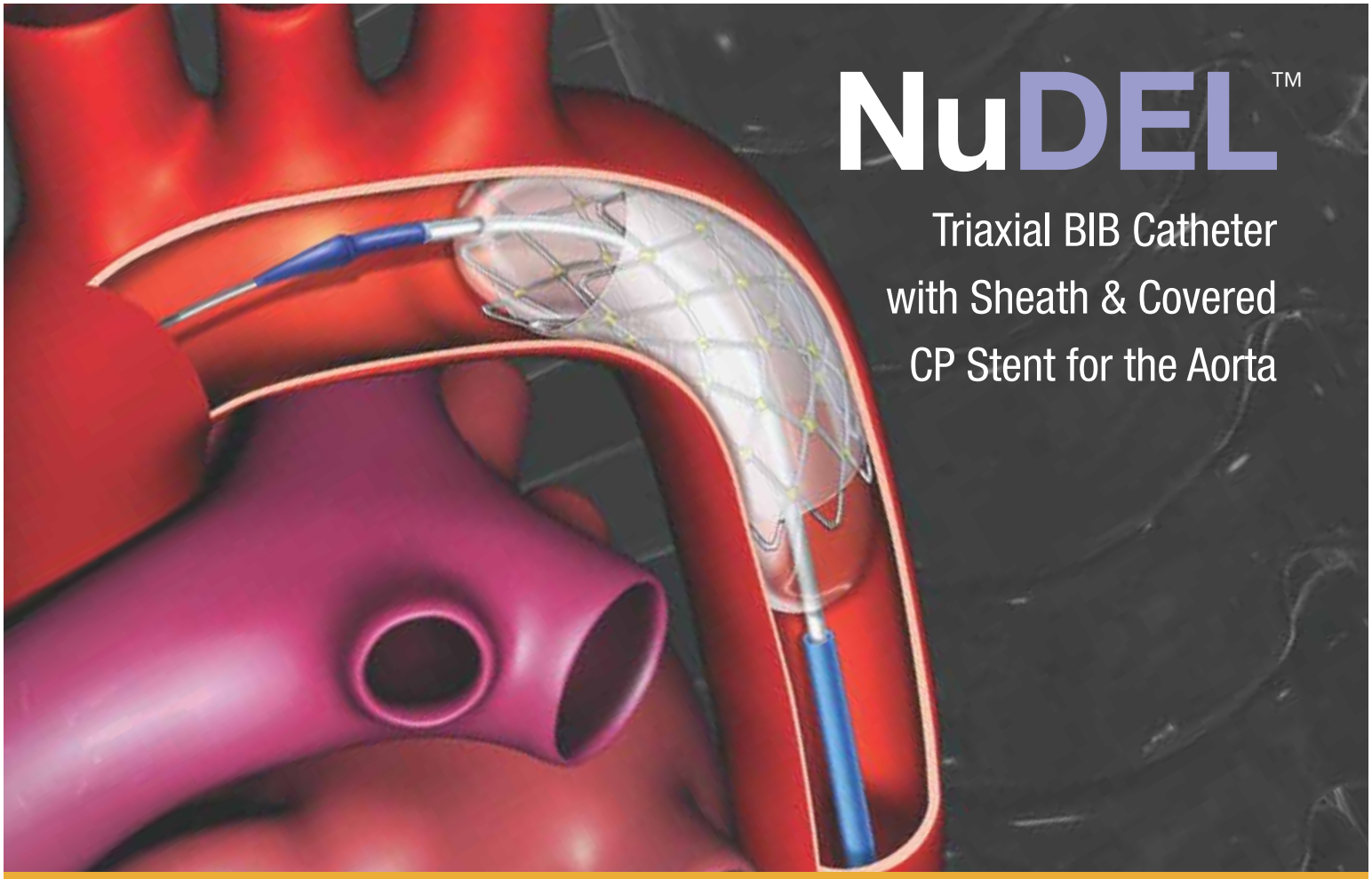
Patients who had higher activity levels following ICD implantation had better survival, according to research in the *Journal of the American Heart Association*. The research will be simultaneously presented at the Heart Rhythm Society 2015 Scientific Sessions.

An implantable cardioverter-defibrillator (ICD) is a battery-powered device that typically combines a "generator" placed under the skin near the shoulder with a wire that is inserted into the heart through the vein beneath the collarbone. ICDs are very effective at recognizing fast, potentially life-threatening heart rhythm disorders and providing timely shocks that restore a normal rhythm. However, many patients who receive ICDs may still be at risk of dying from progression of their underlying heart disease or other problems.

In the largest study on the relationship between activity and survival in ICD patients, researchers analyzed how active participants were in the first 30-60 days after implantation and then over time up to four years. ICDs collect patients' daily activity automatically using sensors embedded in the device itself, which determines whether patients are active or not on a minute-by-minute basis. "Active" measured in this way means approximately a walking speed of 2 miles an hour.

Researchers found that patients in the most active group after getting an ICD had a 40% absolute lower risk of death at four years compared to patients who had engaged in the least activity. Regardless of age, gender or device type, lower average activity during the first 30-60 days was independently associated with a 44% higher risk of death over time. A similar analysis looking at activity over several years demonstrated a similar risk of death for each 30 minutes' difference in activity. "We expected to see a difference, but we were struck by the magnitude of these results," said Daniel B. Kramer, MD, MPH, lead author of the study and Assistant Professor of Medicine at Harvard Medical School in Boston, MA.

"Patients' functional status clearly predicts survival. Our hope would be to use activity as a factor in not just predicting outcomes but also to guide strategies that may improve outcomes. But that is much further down the line."



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Researchers studied the ALTITUDE registry, a nationwide database that involved 98,437 patients enrolled in a remote monitoring program. About 57% of the patients had received a new or replacement ICD and 43% had received cardiac resynchronization therapy (CRT-D) devices in 2008-12. CRT-D therapy combines an ICD with cardiac resynchronization therapy. Patients were followed for a median 2.2 years.

"What is intriguing about our results is that looking at just one piece of information collected automatically after a device is implanted provides very powerful prognostic information about how they are likely to do over the next several years," Kramer said.

The major limitation of the analysis, researchers noted, is that this study design only supports describing an association between activity and survival. Further studies are needed to test strategies aimed at promoting activity or using this information to change patients' treatment.

Co-authors are: Susan L. Mitchell, MD, MPH; Joao Monteiro, PhD; Paul W. Jones, MS; Sharon-Lise Normand, PhD; David L. Hayes, MD; and Matthew R. Reynolds, MD, M.Sc. Author disclosures and funding information are on the manuscript. The ALTITUDE study is supported by Boston Scientific.

Tiny Heart, Big Promise - Understanding How Cells Become Coronary Vessels May Lead to Advances in Repairing Heart Damage

Newswise — The heart has its own dedicated blood supply, with coronary arteries that supply oxygen-rich blood to the heart and cardiac veins that remove deoxygenated blood. This system of vessels nourishes the heart, enabling it to pump blood to all the other organs and tissues of the body. Yet despite their critical importance, the process and molecules required for coronary vessel development have not been fully determined.

Studying zebrafish, investigators at The Saban Research Institute and the Heart Institute of Children's Hospital Los Angeles discovered a new source for cells that can develop into coronary vessels and have identified the signaling protein, a chemokine called CXCL12, which guides this process. Results of the study was published online May 26*th* by the journal *Developmental Cell*.

Zebrafish have emerged as an important vertebrate model for cardiovascular research for a number of reasons, including the ability to regenerate its heart if damaged, and because the transparency of the embryos allows easy observation of internal processes like blood vessel development. Using confocal and time-lapse imaging, the investigators were able to visualize coronary vessels developing from the endocardium, or the inner lining of the heart – specifically from the atrioventricular canal, the structure that divides the heart into compartments.

"This furthers our efforts into heart regeneration to repair human hearts," said Ching-Ling (Ellen) Lien, PhD, principal investigator at The Saban Research Institute of CHLA and senior author on the paper. "We have now found a novel source of cells that can differentiate into coronary vessels and have identified the factors required."

Lien and her team observed that zebrafish with a mutation at the CXCR4 receptor survive, but are not able to form coronary vessels or undergo heart regeneration following injury. Since fish without this mutation are able to do both, the investigators concluded that an interaction between CXCR4 receptors on endothelial cells and the CXCL12b protein expressed by the myocardium regulate the process. In addition to providing basic information about the developing heart, this finding may also have clinical relevance.

"Children or young adults may not be aware of having abnormal coronary vessels because their circulation is adequate until the heart is stressed by increased demands, for instance when participating in strenuous sports," explains Lien, who is also an assistant professor at the Keck School of Medicine and an investigator at the Cardiovascular Thoracic Institute, both at the University of Southern California. "Then suddenly, an apparently healthy, young person dies. Alternatively, a person with abnormal coronary vessels might have higher risk of experiencing heart attacks later on in life. Our findings will guide future study toward understanding these devastating conditions in order to be better able to diagnose them and develop interventional strategies."

The first author, Dr. Michael R.M. Harrison is a CIRM scholar and Saban RCDF fellow. Additional contributors include Ying Huang and Arthela Osorio, The Saban Research Institute of CHLA; Jeroen Bussmann and Arndt F. Siekmann, Max Planck Institute for Molecular Biomedicine, Muenster, Germany; Long Zhao, C. Geoffrey Burns and Caroline E. Burns, Harvard Medical School; and Henry M. Sucov, Broad Center for Regenerative Medicine and Stem Cell Research at USC.

The study was supported in part by the National Heart, Lung and Blood Institute R01HL096121, The Saban Research Institute Career Development Award, and a California Institute for Regenerative Medicine (CIRM) postdoctoral fellowship TG2-01168.

For more information, visit CHLA.org.

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Fetal Cardiology Opportunity

Health care is transforming. We're transforming health care.

Join Us

If you are an experienced fetal cardiologist interested in joining a progressive practice, consider Arizona Pediatric Cardiology (APC). APC, in affiliation with Phoenix Children's Hospital (PCH), provides state-of-the-art, comprehensive inpatient and outpatient pediatric cardiology services throughout the state of Arizona. Phoenix Children's Hospital, ranked 16th by the U.S. News and World Report, has 24 CVICU beds and two cardiothoracic surgeons who perform over 500 surgeries annually.

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

Strength in numbers

25 pediatric cardiologists, including **5** fetal cardiologists

5 fetal sonographers

2 advanced practice fetal cardiac nurse coordinators

5 office locations

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

This is an excellent opportunity to be a part of a team of echocardiographers who perform and interpret transthoracic and transesophageal echocardiograms in an IAC-accredited lab. Clinical responsibilities also include inpatient and outpatient general pediatric cardiology care.

In addition to fetal cardiology, APC provides a wide range of services and programs including interventional and diagnostic catheterization, electrophysiology/ablations, cardiac imaging (MRI), adult congenital heart disease program, anti-coagulation program, preventive cardiology, heart transplant/heart failure program and a metabolic stress lab.

About Phoenix

- ✶ Wide variety of cultural and sporting events
- ✶ California coast within a day's drive
- ✶ Diverse geography offers desert, lake and mountain recreational activities
- ✶ 310 days of sunshine per year
- ✶ Family-oriented community
- ✶ Excellent school districts
- ✶ Unlimited activities for children

Candidate requirements:

- ✓ A minimum of three years' experience
- ✓ A fourth year of fellowship in advanced imaging training, including fetal echocardiography, preferred

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