Intrapericardial Teratoma in Newborn Infant Revealed by Cardiac Tamponade

By Amadou Gabriel Ciss, MD; Momar Sokhna Diop, MD; Pap Adama Dieng, MD; Pap Salmame Ba, MD; Assane N’diaye, MD, PHD; Mouhamadou N’diaye, MD, PHD; Magaye Gaye, MD; Oumar Diarra, MD, PHD; Mohamed Leye, MD; Souleymane Diatta, MD; Arame Diallo, MD; Fall Lamine, MD; Etienne Birame Sène, MD; Oumar Kane, MD, PHD; Abdou Magib Gaye, MD

Summary
A 15-day-old boy presented with signs of heart failure and tamponade. Two-dimensional echocardiography revealed a complex intrapericardial mass (47 mm x 36 mm) with a large pericardial effusion compressing the heart. A CT scan defined the relationship with great vessels, pericardium and myocardium. Complete surgical resection was performed without complication. Histology of the tumor confirmed the presumptive imaging diagnosis of teratoma. Intrapericardial teratomas are rare primary cardiac tumors usually diagnosed in neonates and infants. They contain endodermic, mesodermic, and neuroectodermic germinals layers. Intrapericardial teratomas are usually benign tumors but may be life-threatening because of pericardial effusion and heart compression. Echocardiography was used to make a diagnosis by showing a intrapericardial heterogeneous mass compressing the heart. Bi-dimensional echocardiography was a performed exam in primary cardiac tumors diagnosis, but tomodensitometry (CT scan) and magnetic resonance imaging (RMI) have advantages in large tumors assessing the relationship between the tumor and adjacent tissues.

Keys Words: tumor, teratoma, surgery, tamponade, cardiac

Introduction
Intrapericardial teratoma is a rare primary cardiac tumor usually diagnosed in newborns. This tumor realized a compression of the cardiac cavities, the respiratory system and pericardial effusion. The authors report a case of intrapericardial tumor in a newborn revealed by cardiac tamponade.
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Case Presentation

A 15-day old male newborn with 3.5 kg weight was admitted with signs of heart failure and tamponade. A two-dimensional echocardiography revealed a complex intrapericardial mass (47 mm x 36 mm) with a large pericardial effusion compressing the heart (Figure 1). An electrocardiogram of the heart was normal. Fluid was removed (150ml of serofibrinous fluid) to prevent cardiac tamponade. A biopsy of the pericardium revealed a benign fibro hyalin tissue proliferation. A CT scan realized after one week showed a intrapericardial multi-cystic mass with a capsule, adhering to the wall of the ascending aorta and compressing the right atrium and the right ventricle, involving of the lateral wall of left ventricle (Figure 2). Alpha fetal protein and beta H-C-G were normal. Under a median sternotomy the tumor was explored. Extra corporeal circulation was on standby. The thymus gland was preserved. The pericardium was opened and a small quantity of fluid was aspirated. A mass, around 60 mm x 50 mm x 30 mm of size was revealed, overlaying the great vessels, with its volume covering the larger part of the right ventricle and pressing on all the heart (Figure 3). The mass was completely removed; the attachment of aortic wall was excised without artery injuries. After meticulous haemostase of the aortic implantation area, the pericardial cavity was rinsed with normal saline solution and drained with a chest tube in a usual way (Figure 4). The post-operative course was simple; the endotracheal tube was removed two hours after surgery, he was discharged from the intensive care unit on the second post-operative day and from the hospital on October 10-12, 2013
Where: Houston Texas
Website: http://www.texaschildrenshospital.org/phfs2013/
the 7th post-operative day. Histopathology exam showed a benign mature teratoma with multiple cysts surrounded by conjunctive tissues, including endodermic, mesodermic, and neuroectodermic germinal layers.

Three months later, the child was asymptomatic with normal echocardiogram.

**Discussion**

Intrapericardial teratoma is a rare primary cardiac tumor. It is an embryologic tumor observed in newborns. Intrapericardial teratomas are usually benign tumors, but may be life-threatening because of pericardial effusion and heart compression. Today the majority of Intrapericardial teratomas are diagnosed in utero by prenatal echography. In this case, tamponade is due to late diagnosis, because echography is not commonly administered to low-income pregnant women.

Two-dimensional echocardiography is the primary diagnostic imaging modality. In under-developed countries, explorations including magnetic resonance (MRI) or CT Scan are not accessible immediately for the majority of population. The only alternative for us in this emergency case of cardiac tamponade is pericardial drainage. MRIs and CT scans have advantages in assessing the relationship between large tumors and adjacent tissues.

The complete excision of the tumor was done 37 days after a diagnostic ultrasound. Surgery was performed with a beating heart. The bypass pump was on standby. In a few cases, these excisions are done with cardiopulmonary bypass and cross-clamping the aorta. These tumors are usually removable and recurrences in mature teratomas are exceptional.
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A newborn with a teratoma has an excellent prognosis after excision. The diagnosis in utero with prenatal echography and resonance magnetic imaging in newborns prevents cardiac adiastoly and improves the care.

References


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Corresponding Author
Amadou Gabriel Ciss, MD
Cardiovascular Surgeon; Specialist, Congenital Heart Diseases; Maître Assistant, Dakar University; Member, African Society of Thoracic and Cardiovascular Surgeons; Member, French Society of Thoracic and Cardiovascular Surgeons
University Hospital of Fann
BP 5035 Dakar Sénégal West Africa
Tel: 00 221 70 600 12 25
Cissgaby@yahoo.fr

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Ventricular septal defect (VSD) + malaligned outlet septum anteriorly (07.10.17)
Pulmonary trunk band (PA band) (12.14.02)
Ligation of patent arterial duct (PDA) (12.24.23)

EACTS-STS Derived Term:
VSD, Type 2 (Perimembranous) (Paramembranous) (07.10.01)
VSD-modifier for infundibular septal morphology, VSD + malaligned outlet septum, Anterior deviation of infundibular septum (07.10.17).
PA Banding, Band on main pulmonary artery (12.14.02).
PDA closure, Surgical therapy, Ligation (12.24.23)

ICD10 Derived Term: Q21.0 Congenital malformations of the cardiac septa. Ventricular septal defect.

Commentary: Hearts with antero-cephalad malalignment of the muscular outlet, or conal septum are usually found in the setting of Tetralogy of Fallot. In the specimen shown here, however, the malalignment of the outlet septum did not cause sub-pulmonary obstruction. This arrangement, with an unobstructed outflow tract, is the lesion illustrated by Victor Eisenmenger at the end of the nineteenth century, and known by many as the “Eisenmenger VSD”. The very existence of this type of heart, with an unobstructed subpulmonary outlet, showed that something extra is needed over and above the anteroccephalad deviation of the outlet septum so as to produce the phenotypic morphology of Tetralogy of Fallot. The additional feature in Tetralogy is an abnormal arrangement of the septoparietal trabeculations, which in most instances are additionally hypertrophied. It is the combination of the deviated outlet septum and the abnormal septoparietal trabeculations that produces the “squeeze” at the mouth of the subpulmonary infundibulum that is the essence of tetralogy of Fallot. We will discuss additional morphological aspects of the consequences of overriding of arterial valvar roots to the definition and description of holes between the ventricles is a future column from the Archiving Working Group.

Please visit us at the AWG Web Portal at http://ipccc-awg.net/ and help in the efforts of the Archiving Working Group and the International Society for Nomenclature of Paediatric and Congenital Heart Disease.

AWG Web Portal link for this series of images: http://ipccc-awg.net/VSD_Page/VSD_07_10_17/VSD_07_10_17.html

Description: This view of the great vessels as they exit the ventricular mass shows the aorta arising slightly to the right of its usual position. The pulmonary trunk is dilated with a pulmonary artery band in place and a ligated arterial duct. The right atrium is dilated and there is right ventricular hypertrophy. Contributor: Diane E. Spicer, BS

Description: An image of the same heart demonstrating on the anterior apical view of the right ventricle the antero-superior malalignment of the outlet septum into the right ventricle with an unobstructed subpulmonary outlet. The aorta overrides the ventricular septum, although the aortic valve is not appreciated in this view. This defect is consistent with an Eisenmenger type of ventricular septal defect. Contributor: Diane E. Spicer, BS

Description: In this anatomic view of the opened left ventricle, the free wall has been lifted away to demonstrate the aortic root. The aorta clearly overrides the interventricular septum with a good portion of the aortic valve supported in the right ventricle. Contributor: Diane E. Spicer, BS
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Vera D. Aiello, MD
Co-chairman, Archiving Working Group
Heart Institute (InCor), Sao Paulo University School of Medicine, Brazil

Jeffrey P. Jacobs, MD
Archiving Working Group
Congenital Heart Institute of Florida, St. Petersburg & Tampa, USA

Corresponding Author

Jorge M. Giroud, MD
Co-chairman, Archiving Working Group
Congenital Heart Institute of Florida & Pediatrix Medical Group
St. Petersburg & Tampa, USA
jorgemgiroud@gmail.com

Diane E. Spicer, BS
Senior Archivist, Archiving Working Group
University of Florida, Department of Pediatrics-Cardiology, Gainesville, Florida and the Congenital Heart Institute of Florida, St. Petersburg & Tampa, FL USA

Robert Anderson, MD
Co-chairman, Archiving Working Group
Institute of Genetic Medicine, Newcastle University, Newcastle upon Tyne, United Kingdom

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An Unusual Case of Prenatal Diagnosis of a Large Left Ventricular Diverticulum and Its Intermediate Term Outcome

By Resham Kaur, MD; Javier Paiz, RDCS; John Brownlee, MD; Umang Gupta, MBBS, DCH

Introduction

Congenital Ventricular Aneurysm or Diverticulum are very rare conditions. There are few reported cases of these lesions in the literature. Here we describe a rare case of Wide-mouthed Left Ventricular (LV) Free Wall Diverticulum that was identified by fetal echocardiography and subsequently followed in our clinic.

Case Report

A 24 week pregnant G4P2012 was referred for fetal echocardiography due to abnormality noted in the Left ventricular LV free wall and mild pericardial effusion.

The echocardiogram done in the clinic showed large LV free wall outpouching with a wide mouth and some synchronous contractility with the rest of LV free wall. A very small pericardial effusion was seen. The LV systolic function appeared to be depressed with marked reduction in the contractility in and around the lesion. No arrhythmias were seen. No intracardiac thrombi were identified.

She was worked up extensively for conditions that could lead to dilated cardiomyopathy in fetus with no cause detected.

The patient was followed in the fetal clinic every 4 weeks by serial echocardiograms, the last being performed at 36 weeks of gestation (Figure 1). No changes were noted either in the size or the function of the LV and the diverticulum.

The fetus continued to show normal growth on prenatal examination and didn’t develop hydrops and showed no arrhythmias. No mitral valve regurgitation was seen.

The baby was subsequently delivered through an uneventful spontaneous vaginal delivery weighing 3420 grams. At birth no other congenital defects were identified.

He was subsequently transferred to a transplant center for evaluation and underwent an extensive work up to rule out any genetic/metabolic abnormalities all of which were negative. He was started on carvedilol and enalapril and after a period of observation was discharged home. At the time of discharge patient was feeding very well, was very vigorous and was growing normally. No arrhythmias were identified at any time during the hospital stay.

He has subsequently been followed very closely in the cardiology clinic and at the time of his last visit was 7 months old and showed normal growth and development.

He has had no episodes of arrhythmias. His diverticulum continues to decrease in size relative to his LV chamber size (Figure 2) with continued improvement in contractility.

Discussion

Congenital Ventricular wall aneurysm or diverticulum is a very rare condition. There are few reported cases of congenital ventricular aneurysm in the literature. The finding of congenital diverticulum is even rarer. Many of these have been diagnosed after birth and only a handful have been diagnosed to date using fetal echocardiography.

The reasons for referral for fetal echocardiography in most cases have varied from abnormalities on the four-chamber views to hydrops/pericardial effusion to arrhythmias (including atrial arrhythmias) seen during routine prenatal ultrasound examination.

Our case was referred to us due to “abnormality” detected on the LV free wall and small pericardial effusion.

Most of these congenital diverticula are described as showing synchronous contractility with the rest of the chamber and usually have a narrow mouth, characteristics, that help differentiate them with aneurysms.

In our case, the mouth of the lesion was wide, which had initially confounded us to believe it to be an aneurysm (Figure 1). Later imag-
ing proved it to have synchronous systolic contraction (Figure 3 & 4). The subsequent improvement in function after birth made the diagnosis of diverticulum more obvious.

Ever since they were first described a common association has been found between these diverticula and other congenital defects, most notably midline thoraco-abdominal defects.12

In our case, we didn’t identify any other congenital defect.

A lot of focus has been placed on the outcome. However, literature remains inconclusive about the course and prognosis of these anomalies. Reported experience has varied from a relatively benign course to a more ominous presentation and outcome.1,2,3,4,5,6,7,8,9,10,11 The most common causes of death that have been reported seem to be arrhythmias or severe fetal hydrops.1,7

Our patient remained stable throughout the pregnancy and showed normal intrauterine growth. Her delivery remained uneventful and he has shown progressive improvement in his left ventricular function with decrease in the size of the diverticulum and improved contractility of the involved region.

References


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Corresponding Author
Resham Kaur, MD
Driscoll Children’s Heart Center
Driscoll Children’s Hospital
3533 S. Alameda St.
Corpus Christi, TX 78411, USA
resham_thethi@yahoo.com
Tel: 36.877.9474; Fax: 361.855.9518
Corresponding email: author-umanggupta@yahoo.com

Javier Paiz, RDCS
Driscoll Children’s Heart Center
Driscoll Children’s Hospital
Corpus Christi, TX 78411, USA

John Brownlee, MD
Driscoll Children’s Heart Center
Driscoll Children’s Hospital
Corpus Christi, TX 78411, USA

Umang Gupta, MBBS, DCH
Driscoll Children’s Heart Center
Driscoll Children’s Hospital
Corpus Christi, TX 78411, USA

Figure 3 & Figure 4. Showing the diastolic and systolic frame of LV and diverticulum in parasternal short axis view. Note the synchronous contractility of the diverticulum with the rest of the LV.
The foramen ovale, a remnant of the fetal circulation remains patent in approximately 25% of the population based on autopsy and echocardiographic studies.1,2 Though population-based longitudinal studies do not support a higher risk of stroke in the presence of a patent foramen ovale (PFO),3,4 patients with cryptogenic stroke are more likely to have a PFO (at least 40%) than the general population.5 Moreover, passage of thrombus originating from the venous system via a PFO into the systemic circulation is physiologically plausible and has been demonstrated in a number of case reports unequivocally demonstrating paradoxical embolism via an interatrial communication.6-9 Intuitively, anticoagulation, antiplatelet therapy or closure of a PFO should abolish or reduce the risk of stroke from paradoxical embolism. Nevertheless, significant controversy remains as to how a patient should be treated who has experienced a stroke in the absence of an identifiable cause with the exception of a PFO.

For the affected individual, there are four options: no therapy at all, anticoagulation, antiplatelet therapy and surgical or percutaneous closure. Unfortunately, a study that provides us with a definitive answer does not exist.

Let us briefly examine the evidence for either of the available approaches. For the first option, no therapy, we do not know what the risk of a recurrent event is because the natural history (no therapy) of individuals with a PFO and cryptogenic stroke is unknown. Regarding the second and third options, we do not know if either anticoagulant or antiplatelet therapy reduces the rate of recurrent events, as neither has ever been compared with no therapy in a randomized controlled trial. Likewise, we do not have any conclusive evidence to support anticoagulant therapy over antiplatelet therapy or vice versa. Finally, percutaneous or surgical closure has never been compared to no therapy in a randomized controlled fashion. To conclude, in the absence of a randomized controlled trial comparing therapy to no therapy, we do not know if any therapy is better than no therapy. Does this imply that our response to the individual with a cryptogenic stroke and PFO should be that no therapy is required?

If we believe that a patient who has experienced a cryptogenic stroke in the absence of a PFO should be treated with antiplatelet therapy because the most likely mechanism of the event (with the exception of the rare pure atherosclerotic debris, fat, tumor or air embolism) is platelet aggregation and thrombus formation with or without embolization, then we should, likewise, favor antiplatelet therapy for the patient in question because these mechanisms may also be present in patients with cryptogenic stroke in the presence of a PFO. The data in support of this approach stems from trials in patients with strokes regardless of mechanism who benefit from aspirin administration. Hence at least an aspirin should be recommended. However, some would argue that, in patients with cryptogenic stroke and PFO, aspirin might not be sufficient because it does not provide adequate protection against thrombus formation in the venous circulation and paradoxical embolization via a PFO. On a theoretical basis, only PFO closure or anticoagulation may have merit in preventing venous thrombus and paradoxical embolism. No dedicated randomized trials comparing aspirin and anticoagulation with warfarin in patients with PFO and cryptogenic stroke exist. Available data stems from subgroup analysis of patients with PFO in stroke trials comparing aspirin and warfarin. Though there was a trend favoring warfarin over aspirin, this was not statistically significant.10,11 However, the trials were underpowered to detect a difference. Hence, in the absence of more convincing data, there is no conclusive evidence to favor warfarin over aspirin for the prevention of recurrent events in patients with PFO and cryptogenic stroke. How about PFO closure? There are non-randomized studies comparing percutaneous PFO closure to medical therapy suggesting a lower stroke recurrence rate after closure12,13 and there is surgical data that suggests a very low rate of event recurrence after surgical PFO closure.14,15 More importantly, there are now three trials comparing percutaneous PFO closure to medical therapy, CLOSURE 1 (Evaluation of the STARFlex Septal Closure System in Patients with a Stroke and/or Transient Ischemic Attack Due to Presumed Paradoxical Embolism through a Patent Foramen Ovale),16 PC (Clinical Trial Comparing Percutaneous Closure of the Patent Foramen Ovale Using the Amplatzer PFO Occluder with Medical Treatment in Patients with Cryptogenic Embolism)17 and RESPECT (Randomized Evaluation of Recurrent Stroke Comparing PFO Closure to Established Current Standard of Care Treatment)18 in patients with cryptogenic stroke and PFO.

In CLOSURE 1, patients were randomized to closure with the STARFlex device (NMT Medical) and in PC and RESPECT to closure with the Amplatzer PFO occluder (St. Jude Medical) versus medical therapy. In CLOSURE 1, 1,909 patients (mean age 45 years) were enrolled. After 2 years, though the rate of the primary event (stroke, transient ischemic attack and 30-day all cause and 30-day to 2 year neurologic mortality) and rate of recurrent strokes were numerically lower in patients who underwent PFO closure, the difference was not statistically significant.16

Three important aspects regarding CLOSURE I are worth mentioning. First, the closure rates were lower with the STARFlex device (no or minimal residual shunt: 86%) than with the Amplatzer PFO occluder in both PC (96%) and RESPECT (94%). Second, the follow-up was limited to 2 years. Third, the rates of thrombus formation and atrial fibrillation appear to be higher after STARFlex implantation than after implantation of the most commonly used Amplatzer devices.18 This may reduce or (in the case of device associated thrombus or atrial fibrillation) offset the potential benefits of PFO closure. In PC, 414 patients (mean age 44-45 years) were enrolled. After a mean of 4 years of follow-up, there was no difference in the primary endpoint (composite of death, nonfatal stroke, transient ischemic attack or peripheral embolism) in an intention-to-treat analysis.17 However, there was a trend toward a lower rate of strokes (0.5% versus 2.4%) in patients treated with PFO closure (p=0.13) by the definition used in the trial and a strong trend towards a reduction in
stroke risk based on the definition used in RESPECT (p=0.07). Nine-hundred and eighty patients (mean age 46 years) were enrolled in RESPECT.\(^1\) Once again, there was a strong trend towards a reduction in stroke recurrence in patients treated with PFO closure compared to those treated medically (after a mean follow-up time of approximately 2 years) in an intention-to-treat analysis. In addition, there was a significant reduction in stroke recurrence in an as-treated analysis (p=0.007). Though this type of analysis may be confounded by non-random factors leading to deviation from the treatment protocol, it is worth mentioning that among the nine patients who experienced a recurrent stroke assigned to the device group, three had not undergone device closure, one of whom had a stroke after enrollment but before scheduled device implantation. The second patient, after treatment assignment, decided not to proceed with device implantation. The third patient underwent bypass surgery and surgical PFO closure after assignment to the device group but prior to planned implantation. Importantly, in subgroup analyses, there was a significant reduction in the primary endpoint in favor of PFO closure (compared with medical therapy only) in patients with an interatrial septal aneurysm or large right-to-left shunt.

Particularly the stronger treatment effect with a larger shunt size is in line with the observed higher event rate in patients with cryptogenic strokes and large shunts,\(^{19,20}\) both of which support the notion that PFO’s promote strokes via paradoxical embolism and the rationale of its closure. In addition, there was a strong trend toward a reduction in the primary endpoint in patients whose index infarct had been located more superficially favoring an embolic etiology.

The event rates continued to separate further in favor of closure with time. For example, the stroke rates were 1.3%, 1.6% and 2.2% in the closure group and 1.7%, 3.0% and 6.4% in the medically treated group after one, two and five years, respectively. Though the absolute difference in event rates is small, the relative difference at 5 years is 65%. Assuming that the difference between the treatment arms is a true difference and not a result of chance, in young patients this may translate into a large number of strokes prevented. In addition, no device erosion or thrombus formation directly related to the device occurred in either the PC or RESPECT trial supporting the safety of closure with the Amplatzer PFO occluder.

If one focuses on strictly predefined intention-to-treat findings of these trials and allows only results with a p-value of <0.05 to be meaningful, discounts subgroup and as-treated analyses and ignores potential differences among PFO devices, withholding PFO closure from patients with a cryptogenic stroke may be the logical consequence. However, based on above described data, it appears more reasonable to conclude that the risk of harm due to device closure (i.e. device associated thrombus formation and device erosion) is very low. Further, there may be a benefit of closure compared with medical therapy particularly in certain patient subgroups. Therefore, though PFO closure in an unselected population is not appropriate, categorically closing the door to PFO closure is equally inappropriate. For example, mandating no therapy or aspirin therapy only in a young individual with a cryptogenic stroke, PFO with a large interatrial shunt or atrial septal aneurysm and superficial infarct seen on brain imaging seems to be premature. Rather, careful patient selection and a fair discussion with the patient outlining the therapeutic options, risks and limitations to current knowledge, but ultimately respecting the patient’s decision seem to be most appropriate.

In this context, we would like to draw attention to our upcoming ICI/CSI Conference in Frankfurt Germany this June, where topics such as the above and many others related to congenital and structural
heart disease will be discussed at length by experts in the respective subjects. For more information visit: www.ici-congress.org and www.csi-congress.org.

References


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Philips Announces FDA Clearance for its EchoNavigator Live Image-Guidance Tool

Royal Philips Electronics announced that it has received 510(k) clearance from the US Food and Drug Administration (FDA) to market its innovative EchoNavigator live image-guidance tool (EchoNavigator). This world-first technology helps interventional cardiologists and cardiac surgeons perform minimally-invasive structural heart disease repairs by providing an intelligently integrated view of live X-ray and 3D ultrasound images. Following the CE marking of EchoNavigator in Europe, Philips will now be able to introduce the system globally.1 The first systems have already been installed in Europe and the US.

Philips’ EchoNavigator has been developed in response to a clear upward trend in the use of both X-ray imaging and 3D cardiac ultrasound imaging (also known as echocardiography, or echo for short) during structural heart disease procedures - an area of interventional cardiology that is growing at around 40% per year. During such procedures, ultrasound imaging provides critical insights into the heart’s soft tissue anatomy, while X-ray imaging has particular strengths in visualizing the catheters and the heart implants.

Working in collaboration with partner hospitals in Europe and the US, Philips designed EchoNavigator to address the unique challenges associated with working with live X-ray and 3D ultrasound images simultaneously. Accurately recognizing the heart structures from these medical images takes years of training and experience, and the use and interpretation of both imaging techniques during the procedures can be challenging, especially when manipulating and steering the catheters that carry the implants. Moreover, the communication during the critical moment of the procedure between the interventional cardiologist or surgeon steering the catheters, and the echocardiographer operating the 3D ultrasound equipment, has been identified as particularly demanding.

“Together with Philips, we set out to bring two separate medical imaging techniques together in a way that provides clear visual guidance,” said Professor John Carroll, MD, Interventional Cardiologist, University of Colorado Hospital, Denver, US. “A world-first, EchoNavigator is enabling us to use X-ray images combined with real-time 3D ultrasound images to navigate catheters and deploy implants in the right position in the heart, making such treatments more straightforward.”

Philips’ EchoNavigator will enable clinicians to perform procedures more efficiently by providing intelligently integrated X-ray and 3D ultrasound images into one intuitive and interactive view, as well as providing easy-to-use system navigation and better communication between the multidisciplinary team carrying out the procedure. As a result, EchoNavigator helps save valuable time and enhances patient care.

“We have learned that ideally two live imaging technologies are needed to guide catheter-based repairs to the heart and a multidisciplinary team is needed to perform it,” said Professor Roberto Corti, MD, Interventional Cardiologist, University Hospital Zurich, Switzerland. “This adds to the complexity of such procedures. The development of a more sophisticated imaging technology such as EchoNavigator will definitely provide us with a better understanding of the complex structures of the heart and its repair.”

“As the global market leader in interventional cardiology, we have worked with our partners to lead the way with pioneering solutions such as our real-time 3D ultrasound technology and more recently our HeartNavigator navigation tool,” said Gene Saragnese, CEO for Imaging Systems at Philips Healthcare. “EchoNavigator is further evidence of our commitment to transforming healthcare through the introduction of innovations that enables best-in-class minimally-invasive procedures.”

Philips offers a comprehensive interventional cardiology portfolio that includes hybrid operating room solutions and imaging solutions, plus advanced interventional tools that work smoothly in sync with them. In 2011, Philips introduced HeartNavigator, a procedure planning and image guidance tool optimized for minimally invasive aortic valve replacements. Philips’ new EchoNavigator unites the company’s strengths in interventional X-ray and ultrasound, as well as its strengths in clinical information solutions. Philips introduced Live 3D Trans Esophageal Echo (Live 3D TEE) technology2 as an industry-first in 2007 on its iE33 ultrasound system and last year introduced CX50 xMATRIX - the world’s first compact portable ultrasound system to incorporate Live 3D TEE technology.3

“By the emerging field of complex structural heart disease interventions, the information obtained by merging imaging technologies, as now possible with HeartNavigator and EchoNavigator, will be of tremendous value to the interventionalist, and in turn to the patient,” said Dr. Carlos Ruiz, MD, Director of the Structural and Congenital Heart Disease program, Department of Interventional Cardiology at Lenox Hill Hospital, New York (US).

1 Not available for sale in Brazil, Saudi Arabia, Serbia, and Australia pending clearance by the relevant regulatory authorities.
2 With this technology an ultrasound scan is taken by placing a special 3D ultrasound transducer into the patient’s esophagus so that it is positioned close to the heart valves.
3 Both iE33 xMATRIX and CX50 xMATRIX support EchoNavigator.

Lower-Profile SAPIEN XT Transcatheter Heart Valve Associated With Improved Procedural Outcomes

Preliminary Results From Inoperable Cohort of The PARTNER II Trial Presented at ACC 2013

SAN FRANCISCO, CA—(Marketwire - March 10, 2013) - Edwards Lifesciences Corporation (NYSE:EW), the global leader in the science of heart valves and hemodynamic monitoring, announced that preliminary results from The PARTNER II Trial demonstrated similar one-year outcomes in mortality and major clinical events between the Edwards SAPIEN XT transcatheter aortic valve and the Edwards SAPIEN valve, yet fewer vascular events with the lower-profile SAPIEN XT valve. These data from The PARTNER II Trial studying transcatheter aortic valve replacement (TAVR) in inoperable patients with severe, symptomatic aortic stenosis were presented today as a late-
The PARTNER II Trial, Inoperable Cohort (Cohort B) (i)

<table>
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<tr>
<th>Outcome</th>
<th>SAPIEN XT</th>
<th>SAPIEN XT</th>
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<th>SAPIEN XT</th>
<th>SAPIEN XT</th>
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<td>All-cause mortality - %</td>
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<td>22.5</td>
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<td>0.706</td>
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<td>Stroke: all - %</td>
<td>4.3</td>
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<td>0.88</td>
<td>5.9</td>
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<td>Vascular events: major - %</td>
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<td>15.5</td>
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<tr>
<td>Vascular events: minor - %</td>
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<td>7.4</td>
<td>0.23</td>
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(i) All percents for mortality and stroke data in this press release are Kaplan-Meier estimates.

Edward's anticipates submitting data from the inoperable cohort (Cohort B) of The PARTNER II Trial to the United States Food and Drug Administration (FDA) in the second quarter. The company expects to complete enrollment in the intermediate risk cohort (Cohort A) of The PARTNER II Trial mid-year.

Background Information on The PARTNER Trial, Inoperable Cohort (Cohort B)

The PARTNER Trial studied 358 patients with severe, symptomatic aortic stenosis deemed inoperable for traditional open-heart surgery, and enrolled between April 2007 and March 2009. Patients were evenly randomized to receive either the Edwards SAPIEN valve or standard therapy. Please note that the results from The PARTNER Trial and The PARTNER II Trial are not directly comparable.

ECG Screening for Competitive Athletes Would not Prevent Sudden Death

The risk of cardiovascular sudden death was very small and only about 30% of the incidence were due to diseases that could be reliably detected by pre-participation screening, even with 12-lead ECGs, according to research in a US high school athlete population presented March 10th at the American College of Cardiology (ACC) Scientific Sessions.

Sudden death in young competitive athletes due to cardiovascular disease is an important community issue, which could impact the design of population-based screening initiatives. The frequency with which these tragic events occur impacts considerations for selecting the most appropriate screening strategy. Currently, athletes are assessed through a healthcare professional performing a physical exam and reviewing the individual's clinical history.

“Screening initiatives for high school-aged athletes have the potential to impact 10-15 million young adults in the US,” says the study’s lead author, Barry J. Maron, MD, Director of the Hypertrophic Cardiomyopathy Center at the Minneapolis Heart Institute Foundation in Minneapolis. “This is a controversial issue because some are suggesting that all young competitive athletes should be screened with a 12-lead ECG screening, which would be a massive and costly undertaking. Also, we do not have any evidence to show whether this is clinically necessary.”

To assess this need, Maron and his colleagues interrogated the forensic case records of the U.S. National Registry of Sudden Death in Athletes over a 26-year period (1986-2011) to identify those events judged to be cardiovascular in origin occurring in organized competitive interscholastic sports participants in Minnesota. There were more than 4.44 million sports participations, including 1,930,504 individual participants among 24 sports.

There were 13 incidences of sudden deaths in high school student-athletes related to physical exertion during competition (7) or at practice (6). The ages were 12 to 18 and each was a white male. Most common sports involved were basketball, wrestling or cross-country running. Sudden deaths occurred in 1 out of 150,000 participants.

Autopsy examination documented cardiac causes in 7 of the 13 deaths. In only 4 athletes (31%) could the responsible cardiovascular...
cular diseases be reliably detected by history, physical exam or 12-lead ECG, which is equivalent to 1 in 1 million participants.

“This very low event rate does not warrant changing the current national screening strategy, especially because only one-third of the deaths would have been detectable through additional screening,” says Maron. “These findings demonstrate that these tragic events are rare. In addition to these data, no evidence in the medical literature has shown that ECGs reduce mortality in a broad-based screening effort.”

Close-to-the-Heart Catheters Safer for Hospitalized Children

Location, location, location. A new Johns Hopkins Children’s Center study shows the real-estate mantra also holds true when it comes to choosing correct catheter placement in children.

The research findings, described online March 18th in JAMA Pediatrics, show that catheters in children inserted in a vessel in the arm or leg and not threaded into a large vein near the heart are nearly four times as likely to dislodge, cause vein inflammation or dangerous blood clots as are catheters advanced into major vessels near the heart.

A peripherally inserted central venous catheter, or PICC line, is a tube placed into a small blood vessel, usually in the arm, and threaded toward a major blood vessel near the lungs and heart to serve as a temporary portal for medications, nutrients or fluids. However, clinicians sometimes forego threading close to the heart and leave the PICC line in a peripheral vein in the arm or leg instead — a choice dictated by the ease and speed of placement or a child’s overall condition or anatomy.

The study findings, however, suggest that leaving the device in a non-central vein should only be done as last resort, the researchers say.

“Clinicians should carefully weigh the ease and speed of non-central vein placement against the higher complication risk that our study found goes with it,” says senior investigator and pediatric infectious disease specialist Aaron Milstone, MD, MHS.

Non-central, smaller veins, especially those in the arm, are narrower, thinner and more prone to injury than major vessels near the heart, the researchers say. Thus, a catheter can easily damage the protective coating on the walls of such veins and encourage the formation of blood clots that, in the worst-case scenario, can dislodge and travel to the lungs or heart, causing a pulmonary embolism or heart damage.

Conducted among more than 1,800 pediatric patients hospitalized at Johns Hopkins over six years, the study found that such non-centrally positioned catheters accounted for a mere 16% of the central lines, but for 44% of all complications that led to catheter removal.

Children in the study cumulatively underwent more than 2,500 catheter insertions, of which more than 500 — one in five — had to be removed due to complications. Three-quarters of problems stemmed from mechanical malfunction such as device breakage or dislodgement, clot formation or blood vessel inflammation. The rest were due to infection, which traditionally has been the greatest worry with central lines. Vein location, however, played no role in infection risk, the research showed.

Despite the higher risks seen with non-centrally position catheters, overall complications rates dropped significantly over the six-year study period, a trend that should get a further boost by emerging technologies, the investigators say.

“We are already adopting new technologies that render PICC placement near the heart easier, safer and faster, and which will drive complications rates further down,” says Leslie Gosey, RN, MS, leader of the pediatric catheter-insertion team at Johns Hopkins.

The study was funded by the National Institute of Allergy and Infectious Diseases (K23 AI081752) and the National Institute of Nursing Research (R03 NR012558).

Co-investigators included: Ketan Jamani, BDS, MPH; Sonali Advani, MBBS, MPH; and Nicholas Reich, PhD, all from Johns Hopkins.

Depression in Kids Linked to Cardiac Risks in Teens

Teens who were depressed as children are far more likely than their peers to be obese, smoke cigarettes and lead sedentary lives, even if they no longer suffer from depression.

The research, by scientists at Washington University School of Medicine in St. Louis and the University of Pittsburgh, suggests that depression, even in children, can increase the risk of heart problems later in life.

The researchers reported their findings March 15th at the annual meeting of the American Psychosomatic Society in Miami, FL.

“Part of the reason this is so worrisome is that a number of recent studies have shown that when adolescents have these cardiac risk factors, they’re much more likely to develop heart disease as adults and even to have a shorter lifespan,” says first author Robert M. Carney, PhD, a professor of psychiatry at Washington University. “Active smokers as adolescents are twice as likely to die by the age of 55 than nonsmokers, and we see similar risks with obesity, so finding this link between childhood depression and these risk factors suggests that we need to very closely monitor young people who have been depressed.”

Researchers have known for years that adults with depression are likely to have heart attacks and other cardiac problems, but it hasn’t been clear when risk factors for heart disease such as smoking, obesity and sedentary lifestyle join forces with depression to increase the risk for heart problems.

“We know that depression in adults is associated with heart disease and a higher risk of dying from a heart attack or having serious complications,” Carney says. “What we didn’t know is at what stage of life we would begin to see evidence of this association between depression and these cardiac risk factors.”

The researchers studied children who had participated in a 2004 study of the genetics of depression. At the time, their average age was 9. The investigators surveyed 201 children with a history of clinical depression, along with 195 of their siblings who never had been depressed. They also gathered information from 161 unrelated age- and gender-matched children with no history of depression.

In 2011, when the study participants had reached the age of 16, the researchers surveyed them again, looking at rates of smoke...
ing, obesity and physical activity in all three groups of adolescents.

“Of the kids who were depressed at age 9, 22% were obese at age 16,” Carney says. “Only 17% of their siblings were obese, and the obesity rate was 11% in the unrelated children who never had been depressed.”

Carney and his colleagues found similar patterns when they looked at smoking and physical activity.

“A third of those who were depressed as children had become daily smokers, compared to 13% of their nondepressed siblings and only 2.5% of the control group,” he says.

In terms of physical activity, the teens who had been depressed were the most sedentary. Their siblings were a bit more active, and members of the control group were the most active.

When the researchers took a closer look and used statistical methods to eliminate other factors that potentially could have influenced smoking or obesity rates in the depressed children, Carney’s team found that the effects of depression grew even more pronounced.

“The siblings of depressed children were five times more likely to smoke than members of the study’s control group, so depression wasn’t the only risk factor for smoking,” he explains. “But the depressed children in the study were another 2½ times more likely to smoke than their nondepressed siblings.”

And the heart disease risk factors were more common in formerly depressed children whether or not they still were clinically depressed at the time of the second survey. In fact, Carney says, for most of the adolescents, depression was in remission by the time the second survey was conducted in 2011, with only 15% of them reporting depression.

The results suggest that any history of depression in childhood appears to influence the presence of cardiac risk factors during adolescence, according to Carney.

“Depression seems to come first,” he says. “It’s playing an important, if not a causal, role. There may be some related genetic influences that give rise to both depression and to heart disease, or at least to these types of cardiac risk behaviors, but more study will be required before we can draw any firm conclusions about that.”


Washington University School of Medicine’s 2,100 employed and volunteer faculty physicians also are the medical staff of Barnes-Jewish and St. Louis Children’s hospitals. The School of Medicine is one of the leading medical research, teaching and patient care institutions in the nation, currently ranked sixth in the nation by U.S. News & World Report. Through its affiliations with Barnes-Jewish and St. Louis Children’s hospitals, the School of Medicine is linked to BJC HealthCare.

**Novel Approach To Treating Children With Irregular Heart Beat**

Newswise — A new retrospective study from The Children’s Hospital at Montefiore (CHAM) found similar safety and efficacy at lower cost using a novel three-catheter approach for ablation in children with Wolff-Parkinson-White (WPW) Syndrome, a condition where an extra, abnormal electrical pathway in the heart causes rapid heart rate or tachycardia. The current standard of care for ablation of left-sided accessory pathways in children with WPW is a five-catheter approach and patients treated with this approach served as the age and gender control match in this study. These data were presented March 10th at The American College of Cardiology’s 62nd Annual Scientific Session in San Francisco.

“With increased concern over healthcare expenditures, it is paramount that we maintain quality, efficacy and safety while also maintaining or reducing costs,” said Robert H. Pass MD, Director of the Pediatric Cardiac Catheterization Laboratory, CHAM, and Associate Professor of Pediatrics at Albert Einstein College of Medicine of Yeshiva University. “Our study showed that using a three-catheter approach in this patient population can reduce the cost by more than 20% with comparable outcomes.”

Wolff-Parkinson-White affects one to three people in every 1,000 worldwide. The long-term treatment is catheter ablation, where radiofrequency energy is applied to the extra electrical pathway, disabling the heart’s ability to beat irregularly. Typically five catheters are used to monitor both sides of the heart, but in patients with WPW who have a left-sided pathway, an electrocardiogram test prior to the procedure enables surgeons to accurately predict the location of the pathway. That information allows for reduction in the number of catheters used on the right side of the heart.

The four-year study included 56 children with WPW who were treated with catheter ablation. One group (n=28) received the new approach, where three catheters were inserted through the patient’s groin and neck and threaded through to the heart where the extra electrical pathway was identified and disabled, and had a 100% success rate. The control group (n=28) received the standard, five-catheter approach and had a 96% success rate. The average catheter cost was lower in the three-catheter group ($1,940 vs. $2,620).

“In Wolff-Parkinson-White ablation cases where we can predict that a pathway is located on the left side of the heart, we are confident in this method and believe it should be applied more often,” said Dr. Pass. “Most importantly, with the growing cost of healthcare, we believe any such effort to provide outstanding and consistent outcomes while garnering significant savings is paramount.”

The Pediatric Cardiac Catheterization Laboratory is part of the Pediatric Heart Center at CHAM. It was the first hybrid catheterization lab in the New York metropolitan region, allowing for maximal efficiency and coordination of care for children requiring both surgical and interventional procedures. The Pediatric Heart Center is recognized as a world leader in providing advanced cardiovascular care for young patients with congenital heart diseases, treating patients of all ages, from newborns to adults, from minor arrhythmias to heart transplantation.

For more information please visit www.montefiore.org and www.montekids.org.
Beware: Newly Recognized Heart Cardiomyopathy Is Not Always Benign - Largely Present in Women, ‘Broken Heart Syndrome’ is Often Triggered by Stress

Even though a newly recognized cardiomyopathy, which mainly impacts women, is typically treatable, Tako-tsubo cardiomyopathy can also be deadly when compounded by other co-morbidities, such as heart failure, according to a study was presented March 9 at the American College of Cardiology (ACC) Scientific Sessions.

This condition, formally known as Tako-tsubo Cardiomyopathy (TTC) and informally known as stress cardiomyopathy or Broken Heart Syndrome, has abrupt onset of symptoms and is characterized by a distinctive left ventricular (LV) contraction profile. Ninety percent of the time, this condition affects women, who are usually middle-aged and older, and the condition usually is triggered by a stressful event.

“Although TTC is typically reversible and considered to have favorable clinical outcomes, we have identified an important subset of patients, particularly those with severe heart failure and hypotension, who can have a substantial mortality risk,” says the study’s lead author Scott W. Sharkey, MD, a research cardiologist at the Minneapolis Heart Institute Foundation and a physician at the Minneapolis Heart Institute® at Abbott Northwestern Hospital in Minneapolis. “It’s also important that physicians are aware this is not a rare a condition, as it is present in nearly 10% of women who present to the hospital with suspected heart attacks.”

MHIF researchers reviewed 250 TTC patients who presented to the Minneapolis Heart Institute at Abbott Northwestern Hospital between 2001 and 2012. Then, they segregated those TTC patients presenting with particularly severe heart failure and very low pressure (< 100 mm Hg), who required supportive treatment.

They found that severe hypertensive heart failure occurred in 45 patients. In this subset, 9 female patients died in-hospital despite aggressive treatment intervention, representing the only TTC-related hospital deaths in the 250 patient cohort.

Therefore, Sharkey and his colleagues concluded that TTC is not necessarily a benign condition. Severe hypertensive heart failure of severity necessitating vasopressor and/or intra-aortic balloon pump occurs in nearly 20% of patients. Also, all TTC-related hospital deaths occurred in the hypertensive heart failure subgroup with an overall mortality of 3.5%.

Importantly, triggering physical stressors related to severe co-morbid non-cardiac conditions (8) or advanced age (1) were present in all 9 non-survivors, Sharkey notes.

“To raise additional awareness and improve care of these patients, he adds that guidelines would be helpful at this time, in order to standardize diagnosis and treatment across varied healthcare settings.

“Unfortunately, there are not any guidelines or criteria to instruct diagnosis and treatment of these patients at this time,” says Sharkey. “Therefore, this study could be a starting point for this process, as it provides a more complete profile of the clinical spectrum of TTC and provides useful guidance for the effective management of these acutely ill patients.”

To: LTE@CCT.bz

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

Letters to the Editor

Congenital Cardiology Today welcomes and encourages Letters to the Editor. If you have comments or topics you would like to address, please send an email to: LTE@CCT.bz, and let us know if you would like your comment published.
Fixing a heart from birth through adulthood takes big teams working together. So we examined the needs of leading clinicians when designing our hybrid solutions. The result: our Infinix™-i with 5-axis positioners and low profile detectors, stays out of the way, but right where needed, providing the best possible access to patients. To lead, you must first listen. medical.toshiba.com