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PERCUTANEOUS CLOSURE FOR ANEURYSMS OF PERIEMBRANOUS VENTRICULAR SEPTAL DEFECTS: A NOVEL STRATEGY FOR TECHNOLOGY AND APPLICATION

By Liu Hanmin, MD, Hua Yimin, MD; Zhou Tongfu, MD; Zhu Qi, MD; Hua Jiping, MD; Wang Yibin, MD; Shi Xiaoqing, MD

Introduction

Ventricular septal defect with aneurysm of the membranous ventricular septum (AMS) is an aneurismal protrusion toward the right ventricle, and is usually identified in the pars membranacea, a relatively small, thin, oval-shaped fibrous structure in the interventricular septum. It is a less common anomaly generally diagnosed by echocardiography and angiography. Since Lev and Saphir reported the first 70 cases in 1826, studies have been carried on the formation and clinical diagnoses of AMS. At the end of the 20th century, Amplatzer device developed by AGA Medical Corporation (U.S.A.) for perimembranous VSDs came into clinical use[1-5]. With the understanding of anatomic formation and hemodynamic changes of AMS and the advancement of interventional cardiology, the management strategy to close AMS continues to improve. Recently, interventional strategies have attracted a great interest in the therapy of AMS. In this retrospective study, we reviewed percutaneous occlusion of AMS performed at our medical

center over the past two years and proposed some applicable suggestions on device selection and placement.

Materials and Methods

Definition

For the purpose of this study, AMS is defined as an angiographic evidence of any deposition of tissue in any form around the rim of the

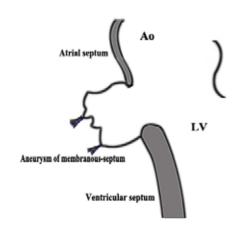


Figure 1.

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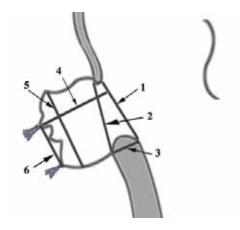


Figure 2.

membranous VSDs, which produces a structure protruding into the right ventricle with each systole and occluding the blood flow through the associated defect partially or completely (Fig 1).

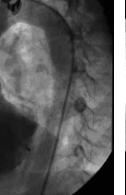
Based on the definition, we reviewed 96 VSD patients who were admitted in our medical center and accepted for percutaneous closure from June, 2004 to April, 2005. All patients were screened by twodimensional Doppler transthoracic echocardiography (TTE, VIVID-7, General Electric Company) with four-chamber view, five-chamber view and parasternal short-axis view. The number, size and exact location of VSD were assessed before intervention. All patients' legal representatives (parents or grandparents) had read the details about the benefits, complications and procedures of the device closure and signed the agreements before the intervention.

Parameter Measurements of AMS

In order to describe the AMS clearly, left ventricular angiography in single plane (60°LAO/ 20°cranial) was performed at the beginning of the intervention. We defined six segments as follows:

- the diameter of the defect on the left ventricle:
- 2. the diameter of the defect on the right ventricle;
- the thickness of ventricular septum; 3.
- the distance from the farthest end of the aneurysm to the defect;





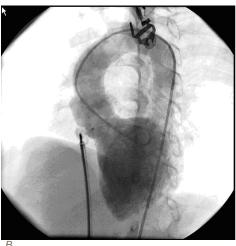


Figure 3 (A-B).

- 5. the diameter of the widest part of the aneurysm;
- the distance between the two farthest orifices on the aneurysm (Figure 2).

After reviewing the angiography in the left ventricle carefully, we chose an appropriated image to do the measurement. Two specialists did the measurements separately and the measurement was reassessed if the difference was apparent until an agreement was reached.

Device and Delivery System Conformation

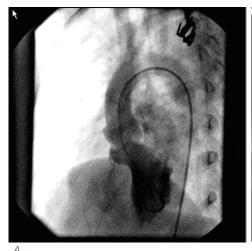
Two kinds of devices were adopted in our study: Amplatzer PMVSD occluder from AGA Medical Corp., and a PMVSD device from Shanghai Sharp Memory Alloy Co, Ltd, China(SHMA). Recent publications have described the Amplatzer PMVSD device in detail. The device contains a dissymmetrical left ventricular disk and a round right ventricular disk, with a selfexpandable short cylindrical connecting waist. The delivery system consists of a delivery cable, a long braided sheath with a dilator. Compared to the Amplatzer device, the device from SHMA contains two symmetrical disks and a self-expandable cylindrical connecting waist that is 0.5 mm longer than the Amplatzer device.

Procedure

The technique of transcatheter closure of VSD was similar to that recommended by AGA Medical Corporation[6]. Under gen-

eral anesthesia, cardiac catheterization and angiography were performed before occlusion. Firstl after identifying the left arterial femoral access and the right venous femoral access, a 6 Fr NIH catheter was advanced from the right femoral vein to the pulmonary artery. A 5 Fr pigtail catheter was inserted into the left ventricle. The pressure was recorded separately and blood samples were collected to evaluate the pulmonary resistance. Second, left ventricular angiography in single plane (60°LAO/ 20°cranial) was performed to define the location, size and the number of openings of the AMS in the right ventricular surface.

The closure procedure was performed in two steps. First, crossing the VSD from the left ventricle. A 5 Fr Judkins right coronary catheter (JR) was inserted into the left ventricle to point toward the defect, into which a 0.035" 260 mm exchange glide wire was then inserted and delivered into the right ventricle, followed by pushing the JR catheter to slide into the right ventricle along the wire. An Amplatzer noodle wire was inserted from the left femoral artery to replace the exchange guide wire and was advanced into the superior vena cava or branch pulmonary artery through the right ventricle. An Amplatzer snare was placed into the corresponding site to exteriorize the noodle wire from the femoral vein. This procedure provided a stable arteriovenous loop to allow the long delivery sheath to advance from the



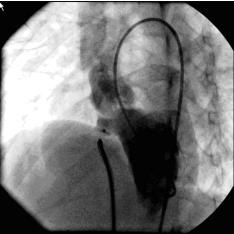


Figure 4 (A-B).

Table 1. Relationship between Orifices and the Shape of Aneurysms

	Orifices of Aneurysms		
	1	2	≥3
Cystiform aneurysms	6	3	7
Tubiform aneurysms	16	2	1

В.

femoral vein and position into the ascending aorta. Second, positioning the delivery sheath into the left ventricle, the sheath was placed from the right femoral vein over the noodle wire to reach the left ventricle. The noodle wire was removed and a corresponding device was advanced along the delivery sheath. With the guidance of ultrasound and X-rays in single plane (60°LAO/ 20° cranial), the device was precisely deployed like a flower bud and pulled back into the left ventricle. Then the device was positioned and released as usual. Doppler detecting and left ventricle angiography were performed to exclude residual leaks or aortic valve regurgitation. The patient was observed in the cardiac intensive care unit overnight.

All patients had a transthoracic color Doppler echocardiography and routine twelve-

lead electrocardiography at 24h post closure and were discharged four or five days later.

Statistical Analysis

Data are expressed as means ± standard deviations or range, as appropriate.

Results

Patient Data

Thirty-seven patients from June, 2004 to April, 2005 were enrolled in this study. Twenty-one of 37 (56.8%) patients were male. The median patient age at the time of the catheterization was 5.3 years (range 2.0 to 14 years). The median weight was 17.6kg (range 8.5 to 28.0 kg). All patients were diagnosed as having significant isolated perimembranous VSD with septal aneurysm via physical examinations, electrocardiography, radiology, transthoracic

echocardiography, and left ventricular angiography.

Echocardiography Findings

Twenty-two cases were diagnosed as aneurysms by transthoracic echocardiography (60% positive). Evaluation of the VSDs' location in the parasternal shortaxis view revealed that two VSDs were at 9 o'clock position, four at 9 to 10 o'clock position, eleven at 10 o'clock position, three at 10 to 11 o'clock, and two at 11 o'clock position. The size of the maximal orifice ranged from 1.2 mm to 7 mm with the average of 2.9 mm. The rim from defect to the aortic valve ranged from 2 to 7mm. With color Doppler echocardiography, the maximal velocity of the jet flow through the shunt ranged from 3.94 to 6.40 m/sec with the average of 4.57 m/sec.

Aneurysm Measurements

The left ventricle angiograms were performed in the single plane (60°LAO /20°cranial) before the occlusion. After choosing a clear and proper image, the aneurysms were assessed as follows:

Morphological Issues of AMS

Angiography revealed variations in AMS morphology, size and location. According to our definition, aneurysms could be classified into two types: cystiform and tubiform aneurysms. Cystiform aneurysm referred to one that had irregular shapes with a diameter of the widest part of the aneurysm 2 mm greater than that of the defect on the left ventricle. Tubiform aneurvsm was defined as a tube - like shape with a diameter of the widest part of the aneurysm equal to or smaller than that of the defect on the left ventricle, a type also including the infundibular aneurysm. Table 1 shows the relationship between the orifice and shape of AMS. Multiple orifices were more common in cystiform aneu-



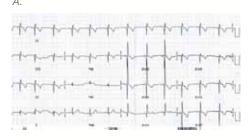
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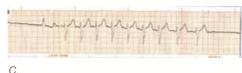
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B.



C.



D



E.

Figures 5 (A-E).

rysms than those in tubiform, and single orifice was more common in tubiform aneurysms than in cystiform. The detailed morphological measurements were described in Table 2. No statistical differences were found in parameters between

Table 2. Measurements of AMS Parameters

	Cystiform aneurysm	Tubiform aneurysm
the diameter of the defect on the left ventricle	7.38±2.32	5.62±2.35
the diameter of the defect on the right ventricle	5.13±1.86	3.69±1.46
the thickness of ventricular septum	3.67±1.06	3.60±0.76
the distance from the farthest end of the aneurysm to the defect	5.76±1.84	6.11±1.35
the diameter of the widest part of the aneurysm	9.00±2.87	4.03±2.13*
the diameter of the largest orifice	3.39±0.98	2.99±0.63

^{*} Compare to cystiform group, p<0.05

the cystiform and tubiform AMS patients except the diameter of the widest part of the aneurysms.

<u>Technical Issues of the Device Choice</u> and Deposition

Successful implantation of the device was achieved in thirty-four patients. The fluoroscopy and procedure times were 13.58±5.72 (8-30) and 63.8±29.3 (30-155) min, respectively. The left disk of the device was deployed in two sites: the left ventricular septum or the left side of the exit orifice. Figure 3 and Figure 4 showed the deployment of the device. Thirty-one of 34 successful implantations had the left disk deployed on the left ventricular septum. Three of 34 had the left disk deployed on the left side of the exit orifice. Table 3 shows the morphological assessment of these three patients. Three patients of this group failed the closure. The girl who failed the intervention suffered a third degree atrioventricular block during the procedure, and the procedure was aborted. In the two boys, the left ventricle diameter of the defect was too large and/or the distance between the two farthest orifices in the aneurysm was to long for the devices to deploy, and the procedures were aborted.

Device selection was mostly based on the diameter of the defect on the left ventricle. In our study of those deployed the left disk on the left side of the orifice, eight were selected with the size equal to the diameter of the defect on the left ventricle; six were selected with a size 1 mm larger waist than the diameter of the defect on the left ventricle; fourteen were selected with 2 mm larger waist than the diameter of the defect on the left ventricle. In patients 9 and 13, the angiography showed the distance from the farthest point of the aneurysm to the defect was very long (9.2 and 10.0 mm), so we selected a 12 mm and a 14 mm device to close the defect, respectively.

Complication Issues of the Closure

Screening of patients with echocardiography and electrocardiography three times after the occlusion revealed neither residual shunt nor regurgitation of the tricuspid valve or aortic valve. Hemolysis and device shift were not detected in our group. Arrhythmia was presented as the most common complication. For the purpose of this study, arrhythmia was defined as abnormal ECG which lasted for over one hour during or after the occlusion. Those who did not have the procedure were excluded. Eleven of thirty-five patients presented arrhythmia, including one whose ongoing procedure was aborted. The occurrence rate was 31.4%. The classification of the arrhythmia is shown in Table 4. The left axis deviation appeared in seven patients' ECG. Five of them presented LABBB or LBBB or High degree AVB, respectively.

For the arrhythmia occurrence in different stages during perioperative period, two of them occurred when setting up the arteriovenous loop using an ex-

Table 3. Morphological Parameters of AMS and Device Used

Patient number	2	10	32
Classification of AMS	cystiform	tubiform	tubiform
diameter of the defect on the left ventricle (mm)	10.0	8.9	6.2
distance from the farthest end of the aneurysm to the defect	4.8	3.3	3.4
diameter of the widest part of the aneurysm	3.9	2.3	3.8
diameter of the largest orifice	2.1	2.9	2.3
number of the exit orifice	1	1	1
device size	4	6	4

Table 4. Classification of the Arrhythmia

Arrhythmia	Case Number	ECG Sequel when discharged
Accelerated junctional escape rhythm	1	normal
Nonparoxysmal junctional tachycardia	1	normal
Incomplete right bundle branch block	3	no change
Complete right bundle branch block	2	no change
Left anterior bundle-branch block	3	no change
Complete left bundle branch block	1	no change
High degree atrioventricular node block	1	LABBB
third degree atrioventricular node block	1	normal

Notes: Two patients presented both incomplete right bundle branch block and left anterior bundle-branch block.

change-length 0.035' J-tipped guidewire, three occurred when transferring the sheath through the defect, one occurred after releasing the device and four occurred 1 day after the closure. One case suffered high degree atrioventricular node block 5 days after the closure.

Therapeutic modes were conducted as follows. In those who presented sinus tachycardia, single incomplete bundle branch block was observed clinically. Prednisone was given to those who pre-

sented sinus bradycardia, accelerated junctional escape rhythm, nonparoxysmal junctional tachycardia, complete right bundle branch block and double bundle branch block. Methylpredni- solone and isoproterenol were given intravenously to the third degree atrioventricular block and the high degree atrioventricular block patients.

No mortality report was recorded with this group. All patients with the sinus abnormalities and ectopic rhythms recovered

three to seven days after the intervention. The aborted patient recovered after three days with a normal ECG. The patient with the third degree atrioventricular block showed left anterior block and left axis deviation ECG 3 days after closure. Three months later, he presented with normal ECG. The ECG series appears in Figure 5. The other 12 patients with different kind of blocks showed no change in ECG when discharged from the hospital. Follow-up is still ongoing.

Discussion

Aneurysm of the perimembranous portion of the ventricular septum has been an intense research and debate for many years. Several hypotheses are proposed to explain the formation and the attention is mostly focused on the incomplete spontaneous closure of a perimembranous ventricular septal defect[7]. There is no accurate report on morbidity of AMS. Recent data coming from our left ventricular angiography of 200 children with VSD showed aneurysm-like formations in 24% of the patients. In 1970s, studies on AMS were focused on pathogenesis and echocardiography evaluation[8-10]. With the development of intervention, reports on closing AMS are being documented. AMS usually presents with a variety of complex structures and exit orifices as the consequence of blood thrusting and tissue adhesion. It is therefore technically challenging to perform AMS intervention. In order to explore the morphological characteristics and the interventional rules of AMS in detail, we defined six new lineal measures based on the left ventricular angiography. Two measurements establish the diameters of the defect on the left ventricle and of the widest part of the aneurysm. Four measurements defined the position of the device released, involving the diameter of the defect on the right ventricle, the thickness of ventricular septum, the distance



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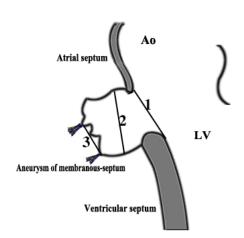


Figure 6.

from the farthest end of the aneurysm to the defect and the distance between the two farthest orifices on the aneurysm. These measurements were utilized to guide our studies.

No unified classification of AMS existed in the past and descriptive terms were of common use. In order to describe AMS effectively in clinics, we classified AMS into two categories: cystiform and tubiform. Cystiform AMS was defined as that with irregular shape and the diameter of the widest part of the aneurysm was 2 mm or greater than the diameter of the defect on the left ventricle. Tubiform AMS was defined as that with regular tube-like shape and the diameter of the widest part of the aneurysm was equal to the diameter of the defect on the left ventricle. Based on this classification. we found that nearly all AMS with multiple exit orifices were cystiform, which presented complex shapes, irregular profiles and various thicknesses of the aneurysm wall. One exit orifice was more common in tubiform and the aneurysm wall was thick.

Selection of an appropriate device and positioning the device were the two major technical issues, and the latter was more critical. The left disk of the device could be placed at positions shown in Figure 6. The left disk could be positioned adjacent to the left side of the defect (line 1), or at the widest part of the aneurysm (line 2), or at the left side of the exit orifice (line 3). Line 2 was inappropriate because there was no foundation to fix the disk. Line 3 was in the right ventricle and could be af-

fected by several right ventricular structures, such as the outlet tract, valves and chordae tendineae. When the aneurysm was long, it was not a proper placement to choose from. The left side of the defect located in the left ventricle had a smooth surface and could be measured clearly and precisely by the angiography. The residual shunt was less frequent with multiple exit orifices when placing the left disk on line 1. So the left side of the defect was the primary position where we placed the device.

The formation of the right disk was the secondary issue we focused on when positioning the left disk on line 1. A proper formation of right disk could close all exit orifices and shorten the length of an aneurysm so as not to interfere with the structures in the right ventricle. As shown in Figure 7, when the body of an aneurysm was long, the length of the device upon the left disk released (segment 1) should be considered to assure the right disk could form properly. A long diameter of the defect on the left ventricle or a long distance between the farthest end of the aneurysm to the defect would prevent a proper formation of the right disk. Under these conditions, we had to select the left side of the exit orifice to position the left disk. There were still limitations to the procedure when the device was larger than 6 mm, or the aneurysm wall was not thick enough, or the multiple orifices spread around the aneurysm in which case, we had to abort the intervention. In our experience. the distance between the two farthest orifices on the aneurysm should be less than 4 mm.

The size selection of the device usually depended on the position of the left disk of the device. In our practice, the size of the device was usually equal to or 1-2 mm longer than the diameter of the left side of the defect when placing the left disk on the left side of the defect. If the body of an aneurysm was too long and the margin from the defect to the aortic valve was enough, we could choose a larger device to make the right disk well formed. When positioning the left disk on the left side of the exit orifice and there was only one exit orifice, the diameter of the device should be equal to or 1 mm greater than that of the orifice. If there were multiple exit orifices, the diameter of the selected device

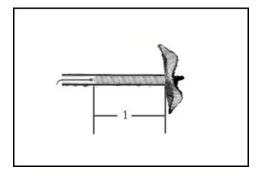


Figure 7.

should be the one minimally required to block all of the orifices.

Compared to the traditional cardiac surgery, the cardiac intervention of VSD was performed without cardiac incision or suture so that it was unlikely to damage or cut off the conductive bundles of the heart. But recent reports showed the tendency of an increased occurrence of arrhythmia during or after the intervention. The morbidity ranged from 12.5% to 47.7%, most of which had different degrees of conductive blocks. There was no direct report of the morbidity of the arrhythmia during the intervention of AMS in the past. Our data showed a 31.4% occurrence of arrhythmia. We surmised that the arrhythmia was caused by congestion and edema around the rim of the defect or direct pressure of the device on the conductive tissue. In our patient data, we found that the highest morbidity occurred at the step of setting up the arteriovenous loop using an exchange-length 0.035' J-tipped guide wire, which might be caused by a relative long time stimulation by the duct or the guide wire to the conductive tissue, because of the complex structure of the aneurysm full of fibers or muscular bundles. When an arrhythmia occurred, it could last for a long time even when the operation stopped. Thus we thought the operation around the ventricular septum and the aneurysm might be one of the reasons causing the higher morbidity of the arrhythmia in AMS.

Another interesting observation was the axis change in ECG. The left axis deviation was common during or after the intervention. We thought it represented to some degree pathological changes because all of the patients were children and manifested no left axis deviation in ECG before the intervention. Some ECGs

showed a correlation between the left axis deviation and the left anterior bundle-branch block. So we hypothesized that the left axis deviation was caused by a stimulation of the left anterior bundle during the interventional procedure. In our patient group, a short and limited follow-up showed that the arrhythmia tended to stabilize after 7 to 10 days, and might be recovered after a longer time period of acclimatization. In our opinion, there was no direct damage of the conductive bundles during the intervention, so the prognosis might be benign. The long-time follow-up results will be reported in the future.

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JOURNEY TO TOMSK, SIBERIA WITH HEART TO HEART INTERNATIONAL CHILDREN'S MEDICAL ALLIANCE

By Janet M. Simsic, MD, Paul M. Kirshbom, MD, Valery O. Kisselev, MD; Evgeny V. Krivochekov, MD

Heart to Heart

Heart to Heart is a non-profit organization founded in 1989 with the goal of developing self-sustaining pediatric cardiac centers of excellence in Russia. Russia is the largest country in the world but only 10-15% of Russian children have access to the medical and surgical care of congenital heart disease that we have throughout the United States. Since 1989. Heart to Heart has sent more than 35 medical missions to Russia, all staffed by volunteer physicians, nurses and related technicians from across the United States. The first pediatric cardiac center of excellence is in St. Petersburg. Russian pediatric cardiac surgeons there, trained by Heart to Heart volunteers. have now performed over 4,000 congenital cardiac surgeries, with outcomes comparable to American standards.

Heart to Heart's ultimate vision is to develop seven such centers, one in each region of Russia's heartland. Heart to Heart makes a 7-year commitment to develop each center; increased funding levels enable development of more than one center at a time. In 2003, Heart to Heart made its first medical mission to Samara, the largest city in the Volga Region (population 33 million). The fourth Samaran medical mission will take place this fall, and pediatric cardiac surgeries at the Samara Regional Cardiac Care Center are already exceeding Heart to Heart's amibitious projections. Most recently (May 2006), Heart to Heart took its first medical mission to Tomsk, Siberia, the largest geographic region of Russia, with a population of 29 million.

History of Cardiac Surgery in Tomsk

The first successful operation on the heart in Tomsk, Siberia was repair of an external cardiac wound performed in 1907 by Professor Tichov. The first operation with cardiopulmonary bypass in Tomsk in a young lady with congenital heart disease (atrial septal defect) was performed in 1986 by surgeon Vikenty Pekarsky. Before that surgical procedures were performed with hypothermia and cardiac arrest, because early devices for cardiopulmonary bypass in Siberia were not adequate.

In 1988, Professor Pekarsky gathered a team of physicians interested in cardiac surgery and established the first cardiac surgical department and the first center of pacing in Eastern Russia at the Institute of Cardiology. Professor Vladimir M. Shipulin became chief of the cardiac surgical department after the death of Professor Pekarsky. Professor Shipulin supported the continuing advancement of congenital heart disease surgery in Tomsk. Since the late 1980's, more than 700 cardiac surgical operations and interventional catheterization procedures have been performed in pediatric patients in Tomsk. Over half of those procedures for congenital heart disease have been in the last 5 years.

Over 200 cardiac surgical operations have been performed over the past 3 years, 60 of them in children under 1 year of age. In 2005, the cardiac surgical team performed 150 cases (130 with cardiopulmonary bypass) and 92 interventional catheterization procedures. Unfortunately the overall mortality is high compared with other centers in the world; mor-

tality in children less than 5 kg is even higher. While the pediatric experience is not extensive and limitations, both technical and medical, are significant, the pediatric cardiac team in Tomsk are dedicated, motivated, enthusiastic and, most importantly, believe in themselves, their abilities, and their future. Heart to Heart is working with the Tomsk team to develop the capacity to perform 300 cases per year, with a goal that ultimately includes a high percentage of newborns and infants.

May 2006, Tomsk, Siberia

A team of 15 physicians, nurses and nonmedical personnel traveled to Tomsk, Siberia to volunteer their time and talents to care for children with congenital heart disease and provide education and training for the Tomsk physicians and nursing staff in preoperative, operative and postoperative management strategies. Our medical team came from hospitals in three different cities: Paul Kirshbom, MD (cardiothoracic surgeon, Emory University/Children's Healthcare of Atlanta Sibley Heart Center), Elizabeth Wilson, MD (cardiac anesthesiologist, Emory University/Children's Healthcare of Atlanta Siblev Heart Center): Janet Simsic. MD (cardiac intensivist, Children's Healthcare of Atlanta Sibley Heart Center); Frank Cetta, MD (cardiologist, Mayo Clinic) Andy Pelech, MD (cardiologist, Children's Hospital of Wisconsin); Matt Brown, MD (cardiology fellow, Children's Hospital of Wisconsin), Brenda Jarvis and Lyndsey Piland (cardiac intensive care nurses Children's Healthcare of Atlanta Sibley Heart Center); Kim Crews (OR scrub technician Children's Healthcare of Atlanta); and AnnMarie McGoldrick (perfusionist Chil-



PediHeartNet is a free internet-based discussion group exclusively for health care professionals interested in Pediatric Cardiology and Cardiac Surgery.

For information or to sign up please visit:

www.Pediheart.net



dren's Healthcare of Atlanta). Our nonmedical team consisted of: Josie Everett (Executive Director, Heart to Heart); Maryanne Kessel (Director, Herma Heart Center at Children's Hospital of Wisconsin); Rose Glickman (program coordinator, Heart to Heart); Joye Leventhal (volunteer, photographer) and Brad Corcoran (volunteer, videographer).

The goal of this journey halfway around the world was to provide education, training, and supplies, as well as to identify areas where Heart to Heart could be of assistance in patient care strategies and program development. The team of Americans worked side-by-side with our cardiology, surgical, anesthesia and nursing colleagues in Tomsk to evaluate 34 children with congenital heart disease, performing 5 cardiac catheterizations and 9 cardiac surgical operations. Operations included repair of Tetralogy of Fallot, repair of aortic coarctation, repair of ventricular septal defect, and repair of transitional atrioventricular canal defect. All of the children who underwent cardiac surgery did well and were transferred to the pediatric floor the following week. They will remain in the hospital for several weeks as is the standard of care in Russia post cardiac surgery.

Our educational highlights included the introduction of a preoperative conference attended by the cardiologists, surgeon, and anesthesiologists to discuss the children prior to cardiac surgery with review of their echocardiograms and angiograms. In the operating room, a new cardioplegia solution was introduced and the technique of modified ultrafiltration was enhanced. In the intensive care unit, educational opportunities included: the importance of vigilance, anticipating potential postoperative problems and, attention to detail following cardiac surgery. Management strategies of Tetralogy of Fallot including the use of a

patent foramen ovale, transannular patch, and right ventricular compliance were also reviewed. Our American team members learned a great deal from their respective Russian counterparts, whose dedication, motivation, enthusiasm, and desire for continued education are truly remarkable -- and inspirational for the entire American team. Our trip to Tomsk is the beginning of a beautiful friendship - and the launch of a 7-year program leading to a self-sustaining resource giving children of this region access to quality pediatric cardiac care for the first time. Our Russian colleagues will save the lives of hundreds of children in the development phase, with the potential for exponential success once the center is established.

~CCT~

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MEDICAL CONFERENCES

The World Congress of Cardiology 2006; Joint Congresses of the European Society of Cardiology and the World Heart Federation Sep.2-6, 18-22, 2006; Barcelona, Spain www.worldcardio2006.org

PICS 2006 (Pediatric and Adult Interventional Cardiology Therapies for Congenital and Valvular Heart Disease)

Sep. 11-14, 2006; Las Vegas, NV USA www.picsymposium.com

Problem Solving in Adult Congenital Heart Disease: A Case-Based Approach

Sep. 16, 2006; San Francisco, CA USA www.ucsfcme.com/blast2006/adultcongenital2.htm

39th Southeast Pediatric Cardiovascular Society Meeting September 28-30, 2006; Stone Mountain, GA USA www.sepcs.org

CHSS (Congenital Heart Surgeon's Society) Annual Meeting 2006
Oct. 8-9, 2006; Chicago, IL USA
www.chss.org

Acute Cardia Care 2006 Oct. 21-24, 2006; Prague, Czech Republic www.escardio.org

Fetal Echocardiography for the Obstretrician and Sonographer Oct. 27-28, 2006; London, UK mailto:Morphology@rbht.nhs.uk

The First Asia- Pacific - Pediatric Cardiology and Cardiac Surgery 2006 Nov. 1-4, 2006; Bangkok, Thailand www.PCCS2006.com

Congreso Latino de Cardiologia Pediatrica

Nov. 2-4, 2006; Lima, Peru www.cardiologiapediatricia.org

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HIGHLIGHTS FROM PEDIATRIC CARDIOLOGY REVIEW 2006

By Anthony C. Chang, MD

This inaugural comprehensive review course of pediatric cardiology, held at the spectacular new five-star Ritz-Carlton Resort in South Beach, was attended by over 200 attendees from 25 countries. While most of the audience were pediatric cardiologists taking either the board or recertification examinations, there were also many attendees from other specialties (pediatric intensive care, cardiac anesthesiology, adult cardiology, cardiac surgery, and nursing) who attended the course as an opportunity to review the entire field of pediatric cardiology in less than one week with the outstanding faculty, which was speciallyselected based on their teaching and expertise. In short, it was a "dream team" faculty.

Each of the comprehensive morning sessions included didactic lectures covering core cardiology concepts (such as molecular biology of cardiology, pulmonary circulation, cardiac pharmacology, etc), followed by comprehensive review of congenital heart lesions (including embryology, anatomy, physiology, and surgery). The second half of the intense morning sessions included select cardiology topics (such as genetic disorders, systemic diseases, Kawasaki disease, etc) as well as 90 minute subarea reviews (including exercise physiology). A fast-paced 15 to 30 minute question and answer period concluded the morning sessions.

There were two lunch sessions: one on tips for examinations and advice for careers, and the other on statistics. The remaining two afternoons were left free for attendees to enjoy the summer weather in Miami, either by the pool or on the beach. The breaks were much needed after the intense morning sessions.

The late afternoon sessions included a one-hour workshop to emphasize the concepts of the subareas reviewed earlier in the day and a 90-minute board question review session that covered twenty questions per session (with answers explained).

The feedback from both the attendees as well as the faculty was overwhelmingly positive. Some of the best-liked aspects of the week were the question and answer periods with the senior faculty members. Over 200 questions from the audience (most written on cards) were fielded during the weeklong course by the faculty. The evening board question sessions were also popular as it utilized an audience response system that enabled the attendee to track his/her own score and cumulative response time. A prize was given for each night's winner (best score and lowest response time) and the overall top three winners were given book prizes of up to one thousand dollars.

The entire course syllabus (with succinct summaries from the presentations as well as MiniTopics in pediatric cardiology) and accompanying CD-ROM (with close to 2000 slides) are available for purchase (please contact me at my email address below).

The next review course (Pediatric Cardiology Review 2008) will be in May of 2008, most likely back in Miami. Hope to see you there!

~CCT~

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SEPTEMBER SYMPOSIUM FOCUS

PICS 2006
(Pediatric and Adult Interventional
Cardiology Therapies for Congenital
and Valvular Heart Disease)
Sep. 10-13, 2006; Chicago, IL USA
www.picsymposium.com

PICS 2006 will feature more than 40 live case demonstrations which will be transmitted via satellite from many centers in the US, Europe, South America and Canada. The emphasis of these live cases will be to demonstrate step-by-step basic and advanced techniques for both common and unusual case scenarios.

This will be a comprehensive symposium worldwide in the burgeoning field of interventional therapies for congenital and structural heart disease in children and adults. This symposium will give the attendees a unique opportunity to interact with a distinguished faculty during didactic lectures, during the live case demonstrations, and smaller group breakout sessions. For the first time, this year's symposium will feature a revised format which is meant to maximize the educational experiences of the attendees. Day 1 will include industrysponsored symposia, oral abstract presentations, operator workshops, and "Meet the Expert" sessions. Day 2 will be a combined session with all attendees and includes topics of general interest and multi-site pediatric and adult interventional live cases. The third and fourth days will be separated into pediatric and adult arenas with specific didactic presentations and live cases transmitted simultaneously to each arena involving different techniques that are applicable to children and adults. This unique format is meant to allow both pediatric and adult interventionalists to interact on common topics and derive individualized educational activities most relevant to their particular interests.

Special sessions during this course will provide in-depth focus on septal defect closure (PFO, ASD, VSD and PDA), coarctation stenting, aortic valve disease (including the new percutaneous aortic valve implantation techniques), mitral valve disease, RVOT and pulmonic valve disease, hybrid intervention for hypoplastic left heart syndrome, and fetal interventions.

Listen to a Congenital Cardiology Today Podcast Interview with Dr. Ziyad Hijazi on the upcoming PICS 2006 at www.CongenitalCardiology.com/z.mp3

HEARTS WITH HOPE MEDICAL MISSION TO AREQUIPA, PERU: INGREDIENTS FOR A SUCCESSFUL MEDICAL MISSION

By Daniel Levi, MD and Ilan Levi, PhD, MD

On January 15, 2006 an enthusiastic group of pediatric healthcare professionals and volunteers arrived in Arequipa. Peru's second largest city. Their mission: Help as many children with congenital heart defects as possible by performing heart surgeries and using transcatheter procedures that will minimize risk, costs and recovery time and maximize the benefits to the children. The mission, staffed predominately by doctors and nurses from Mattel Children's Hospital at UCLA, did most of their work at the Nacional Carlos Alberto Seguin Escobedo Hospital in Arequipa. Peru. Two weeks later the happy, but tired, team boarded a plane homebound to the USA, realizing that their success was the result of not only good fortune, but also good planning. Using the Hearts with Hope Mission as an example, this paper will detail some of the key ingredients for successful medical missions to third world countries.

The Los Angeles based Hearts with Hope Foundation (also known as Corazones con Esperanza) was founded in 2003 and is supported by individual donors and by major cardiac device manufacturers. It is headed by Dr. Juan Carlos Alejos, an interventional and transplant cardiologist at UCLA. The mission of Hearts with Hope is to provide medical and humanitarian aid to children with congenital heart disease and their families throughout Latin America. Dr. Alejos has led three previ-

ous missions to South America. Direct contacts with the health care community led to the fourth mission taking place in Areguipa. This is the fourth and largest team that has been led on behalf of the foundation. At the end of the two week stay in Areguipa, the team had performed 23 transcatheter interventions (in children aged from 2 months to 15 years) ranging from valvuloplasty to ASD and PDA closures to stenting of native coarctations. During the second week, ten surgical cases were performed by Dr. Christian Pizarro. To screen, select and follow-up patients, over 200 echoes were performed.

All the procedures were completed as planned and all patients were discharged in excellent condition with minimal hospital stays. The mission marked the first time for transcatheter closure of ASDs (six ASDs were closed with devices donated by AGA) and PDAs (NitOcclud devices were donated by pfm Inc) to be performed on children in southern Peru. Pediatric open heart surgery had also never before been performed in this region. The milestones were widely reported on the front page of local papers.

Mission Team

The cadre of workers on the Hearts with Hope Team included pediatric cardiologists Drs. Daniel Levi, Greg Perens, Juan Alejos and Josephine Isabel-Jones, who performed most of the interventional procedures and echoes; pediatric anesthesiologists Drs. Mike Sopher and Swati Patel; cardiac perfusionist Juan Obado; cardiac ICU nurses Denise Brown, Jennifer Correa, Ai-Jin



Figure 1. Local surgeons learn techniques for repair of Tetralogy of Fallot from Dr Pizzarro.

Lee and Giselle Mata; and OR and cath lab technicians and assistants Samuel Olmeda. Chris Gatica and Annie Mever. As mentioned above. Dr. Christian Pizzarro, a congenital cardiac surgeon from the Nemours Cardiac Center at the Dupont Hospital for Children performed all of the surgical procedures. Finally, Dr. Jessica Greenwood coordinated the movement of patients from the echo diagnostics to the Catheter Lab and occasionally the OR. Yolanda Peneda, a family liaison with the Medical Home Project at UCLA recreated her role in Arequipa and dealt with families, insurance issues and coordinating catheterization lab procedures.

The UCLA team also included medical students, a photographer and several volunteers without specific medical expertise. Having a large and diverse team, over 20 in total, the Hearts with Hope Team was able to benefit every pediatric hospital and orphanage in Arequipa through a variety of charitable, educational and medical activities. By



having more than "just enough" time and personnel, there could be an emphasis on education as well as patient care. The doctors, nurses and perfusionists were able to hold seminars on the caring for children with congenital heart disease at both Hospital Nacional and Goyeneche Hospital. Each team member had a well defined primary role and each one also worked hard to accomplish specific goals during the time in Arequipa. Without volunteers and students in addition to doctors, nurses, surgeons and other specialists, our primary and secondary goals could not have been accomplished.

Domestic Financial and Industrial Support

The willingness of all companies approached to help with our mission was perhaps the most impressive aspect of the Hearts with Hope Mission. Even given last minute requests, pfm-Ag Inc. donated eleven NitOcclud devices, AGA Medical donated six ASO devices and delivery systems, and Johnson and

"Using the Hearts with Hope Mission as an example, this paper will detail some of the key ingredients for successful medical missions to third world countries."

Johnson/Cordis Corporation donated a wide range of equipment for use in both surgeries and transcatheter procedures. In addition to donating pacemakers, computers to interrogate pacemakers, prosthetic Hancock II heart valves and pacemaker leads, Medtronic allowed one of their present employees (Annie Meyer) to come on the mission.

Because of generous cooperation of the airline LAN Peru, the UCLA medical team was able to transport directly to Arequipa about 50 massive duffel bags full of medicines, medication, transcather and surgical materials as well as

a dozen carefully packed boxes with sensitive instruments. Although the team's plane tickets were purchased with money raised by the Hearts with Hope foundation silent auction fundraiser, all of the catheters, syringes, wires, transcatheter devices, medicines, pacemakers, monitoring equipment and other supplies needed for the planned procedures were transported to Peru on the commercial airliner without additional charge. (All unused supplies were inventoried and left in the hospital - we came back with nothing but gifts for our families).

International Staffing

International support is most important for the identification of potential patients and for the follow-up of these patients after the mission. It is important for a pediatric interventionalist or cardiothoracic surgeon to prepare for a mission with detailed knowledge of the patients to be treated.

In the case of the Hearts with Hope mission, a dedicated team of adult cardiologists, intensivists, nurses and operating room technicians were available, motivated and prepared to assist in the Hearts with Hope Mission. Drs. Pedro Torres, Alejandro Basso and Nassip Llerena were able to send a DVD of ECHOs to us. Having a database of ECHOs several months prior to the trip was invaluable in preparing the equipment and personnel needed for every potential candidate for intervention by our team. In addition to screening more patients for the next mission, these same doctors (adult cardiologists by training) are now following up on children who had surgical and transcatheter interventions.

Given the proper financial support and cooperation from an international airline, it is possible to bring nearly all the equipment and medicines needed for a medical mission in pediatric cardiology. However, no one wants to bring "sand to the beach." Valuable resources can be saved if the appropriate contacts exist at the international site. To further insure that only necessary equipment is transported while no critical items are



Figure 2. The town of Arequipa is surrounded by snow covered volcanoes.

excluded, an advance trip by a knowledgeable "scout" can prove invaluable. The scout would be well advised to take many pictures and even video footage to share with the volunteer team prior to its departure. These visual aids and available equipment lists will allow each physician to thoroughly visualize their planned procedures and list all the devices, facilities, medicines and support required to successfully complete each intervention.

Properly Equipped Location

In addition to the dedicated and competent staff at the hospital in Arequipa Peru, the hospital was adequately equipped to support the mission. Anesthesia carts, ventilators, single plane fluoroscopy and a basic cardiac operating room were available and reserved for our use. Missions like the Hearts with Hope can maximize their impact by serving regions where transcatheter and surgical congenital cardiac interventions are not available for children, yet where some basic OR facilities and diagnostic and imaging equipment are in place. The Arequipa hospital was well equipped for cardiac surgery for adults and the resident cardiologists and surgeons limited their work to adult patients. The adult facilities were easily modified to provide the basic equipment needed for pediatric ICU care and pediatric catheter-based and cardiothoracic surgical interventions.

Charitable/Good Will Component

The Hearts with Hope team developed special ties to several children's hospi-

tals and orphanages. One volunteer, Bill Sears, himself a heart transplant recipient five years ago, was the medical team's goodwill ambassador. Prior to the Areguipa mission, Sears collected several large duffels full of small toys, coloring books with crayons, yo-yos and other common toys. With help from local volunteers, Sears delighted the local children by handing out hundreds of toys at several hospitals and orphanages. Dr. Alejos and team coordinator Jolanda Peneda took the time to assess the most urgent equipment needs of the hospitals in the area, with the intent of supplying the critical equipment with help from US companies and the Hearts with Hope Foundation. Three more large duffels were filled with freshly laundered hand-me-down children's' clothes donated by American families. These were also distributed and fitted to grateful children by local associates of the Foundation. Medical missions certainly have the power to spread goodwill into even remote parts of our world.



Figure 3: Teaching in the cardiac catheterization laboratory.

Education

Local surgeons participated in most of the interventional cardiac procedures that were performed at The Hospital Nacional Carlos Alberto Seguin Escobedo. The cardiac surgeons who routinely operated on adult hearts were eager to observe pediatric repair of congenital lesions including, Tetralogy of Fallot, Double Outlet Right Ventricle, Ventricular Septal Defect with a SubAortic Membrane and Atrial Septal Defects. While the teaching extended into the ICU and nursing staff as well, it always involved both teams of professionals learning from one another.

Our team was also fortunate to have eager UCLA undergraduate pre-med students on our trip. While these students were a great help to our mission (they consistently brought pizza to the on-call doctors), they were also provided with a unique educational experience. Dr. Greg Perens, a UCLA fellow in pediatric cardiology, described his experience as the best learning experience of his fellowship, "These missions are the ultimate cultural and medical learning opportunities."

Fun and Recreation

The mission to Arequipa was not all work for the Americans. A series of parties, dinners, receptions, shopping excursions and an optional weekend sidetrip to Cusco and Machu Picchu all added an element of fun and bonding among the Americans and their Peruvian hosts. We can assume that these recreational and social activities were an extra bonus to the team members adding to the satisfaction they all felt with the impact of their medical endeavors. The social and professional links established between the American volunteers and the Peruvian doctors and nurses will surely serve as a strong foundation for future, perhaps annual missions from Los Angeles to Arequipa.

Upon arrival the team was treated to a tour of the town. Perched in the Andes foothills at an elevation of 7700 ft and surrounded by snow capped volcanic peaks, the town is known for the pearly white volcanic rock, known as sillar, used in many of its buildings. The team was treated to both welcome and farewell barbegues. Complete with traditional Inca music, each of these festive meals included dancing, singing and "pisco sours" (the local drink). While shopping for bargains, drinking Chilean wine and karaoke were favorite evening activities, seventeen members of the mission traveled over the weekend to Cuzco with a visit to the extensive archeological site of Machu Picchu.

Perhaps most memorable is the friend-liness and gratefulness of the parents, children and the entire Arequipa community. Tears of gratitude flowed freely at the EsSalud Hospital as Dr. Alejos and his team bid farewell to the dozens of children and parents whose lives they improved and who can now not only hope but actually realize life for their children with healthy hearts.

~CCT~

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Congenital Cardiology Today

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MEDICAL NEWS, PRODUCTS AND INFORMATION

Request for Applications: Biomedical Research on Pediatric Cardiomyopathy (Dilated, Hypertrophic, Restrictive or Arrhythmogenic Right Ventricular Cardiomyopathy)

<u>Description of Opportunity</u>: The Children's Cardiomyopathy Foundation (CCF) is inviting investigator-initiated research applications for innovative basic, clinical, or translational studies relevant to the cause or treatment of cardiomyopathy in children under the age of 18 years. CCF's grant program is designed to provide seed funding to investigators for the testing of initial hypotheses and collecting of preliminary data to help secure long-term funding by the NIH and other major granting institutions.

Application Deadline & Review Process: Grant award decisions are made through a careful and detailed peer-review process led by CCF's Medical Advisors and reviewed by the Board of Directors. Grant guidelines and the application form are available online at www.childrenscardiomyopathy.org/site/grants.php This year's deadline for grant submissions is October 6, 2006 with final award decisions and funding made available by December 31, 2006.

Eligibility Requirements: Principal investigator must hold an MD, PhD or equivalent degree and reside in the United States. The investigator must have a faculty appointment at an accredited U.S. institution and have the proven ability to pursue independent research as evidenced by original research in peer-reviewed journals.

<u>Available Funding & Award Duration</u>: Funding is available in the range of US\$25,000 to US\$50,000 for one year of total direct costs. For grant renewals, CCF funding is limited to two years (consecutive or otherwise) of support.

For more information contact: Lisa Yue, Executive Director; Children's Cardiomyopathy Foundation; Phone: 201-227-8852, ext 901; E-mail: lyue@childrenscardiomyopathy.org

European Society of Cardiology: Routine ECGs for Newborns Would Identify Life-Threatening Heart Condition Screening

Italian heart specialists are calling on health care providers throughout Europe to give urgent consideration to introducing ECG screening for all babies at around three to four weeks of

age to pick up a life-threatening genetic condition called long QT syndrome.

Evidence they have gathered from ECGs in a just completed and still unpublished prospective study of 45,000 infants, coupled with a cost-effectiveness analysis, has demonstrated that significant numbers of sudden deaths could be prevented for less than €12,000 per year-of-life saved.

The findings from the cost analysis study were published on-line (Thursday 13 July 2006) in the European Heart Journal, the Journal of the European Society of Cardiology.

The study, led by Professor Peter J. Schwartz of the University of Pavia and IRCCS Policlinico San Matteo, focused on the inherited condition long QT syndrome (LQTS), a disorder of the electrical rhythm of the heart[2]. It rarely produces symptoms but is a leading cause of sudden death in children and young adults, and when it occurs in infants their deaths are usually mislabeled as sudden infant death syndrome (SIDS).

In fact, Professor Schwartz and associates have recently completed a study on over 200 victims of SIDS from Norway and have found genetic mutations diagnostic for long QT syndrome in almost 10%. This indicates that early treatment of LQTS would also prevent some deaths regarded as SIDS because, once detected, it can be successfully treated in most cases with medication by betablockers, or occasionally by sympathetic denervation[3] or the use of implantable defibrillators.

Although the focus of the 45,000-infant study was to identify babies with LQTS, the research also unexpectedly revealed four cases involving two other life-threatening heart conditions – coarctation of the aorta and anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA)[4] – which had gone undetected in initial neonatal checks. This finding led directly to the researchers extending their cost-effectiveness analysis to include these two congenital heart diseases.

Professor Schwartz, who is chairman of the department of cardiology, said: "Our study clearly demonstrates that neonatal ECG screening is highly cost-effective and that a significant number of lives can be saved – possibly up to



November 29-Dec 2, 2006

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230-250 a year, for example, in the 15 countries of the pre-enlarged EU – for an objectively small cost. The time is ripe for those involved in the administration of public health to consider the implementation by the National Health Services of such a programme, with the objective of reducing the number of preventable sudden cardiac deaths in infants, children and young adults."

He said that although there could be some differences in specific costs in different countries, their calculation for its introduction in Italy varied all probabilities and costs by plus or minus 30% so as to be valid for most European countries.

There have been various estimates in the medical literature in the past about the prevalence of LQTS, with figures ranging from one in 5,000 to one in 20,000, but none was based on actual data. The Pavia team's research is the first large prospective study on an unselected population. They performed ECG between days 15 and 25 in 45,000 babies and found a markedly prolonged QT interval in almost one in 1,000 (0.9). In more than 50% of these infants Professor Schwartz has identified gene mutations responsible for LQTS. "Therefore this means that the prevalence is probably around one in 2,500 - higher than previous estimates," he said.

"The cost per year-of-life saved by identifying and treating LQTS is really very low – €11,740: for saving one entire life of 70 years we are looking at just €820,000. When, to account for what happens in the real world, we also consider the two other congenital heart diseases that routine ECG screening can pick up, then the cost per year-of-life saved is only just over €7,000 or around €490,000 over a 70-year lifetime."

He said that 220 out of 550,000 new-

borns per year (in Italy) are affected by LQTS. Without screening 13.5% (30) would die early. But screening would cut this to 3.2% (7), saving 23 lives a year. Additionally, screening would prevent four more deaths due to the two congenital heart diseases. In total therefore, screening in Italy would save an estimated 27 lives a year from the three conditions combined.

Professor Schwartz outlined the potential clinical benefits of screening for LQTS:

- we can treat the baby, protecting it from life-threatening arrhythmias, thus reducing deaths in childhood and later years as well
- we can identify other family members affected by LQTS who are at risk of sudden death as well
- by treating all the infants who show manifest LQTS, we would also protect those at risk of dying in the first few months of life and whose deaths would be labelled 'SIDS'
- in addition, our unexpected finding of these two other conditions means we have an opportunity to pick up the few, but life-threatening cases of congenital heart diseases that might escape routine medical checks, for which surgery can radically change the prognosis.

"Parents of a newborn child have the right to know about the existence of an uncommon disease, which is potentially lethal but for which effective and safe treatments exist, and which can be diagnosed by a simple ECG. European citizens and taxpayers also need to be informed about what is possible to do, and at what cost, to reduce the tragic burden of sudden deaths in the young," Professor Schwartz concluded.

For more information: www.escardio.org.

CONGENITAL CARDIOLOGY TODAY

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