Tetralogy of Fallot (TOF) with hemitruncus is a rare congenital heart malformation involving - in addition to TOF - anomalous origin of one of the pulmonary arteries from the ascending aorta. Early recognition of this lesion is essential, as surgical repair needs to be planned early in infancy. Prognosis is otherwise poor due to the high risk for development of significant pulmonary vascular disease in the unprotected lung. The surgical repair could also be disastrous if the anomaly is not diagnosed pre-operatively. Typically, this lesion is diagnosed by echocardiography and angiography in suspected cases. In this article, we report a case of tetralogy of Fallot with left hemitruncus and right aortic arch diagnosed in utero and we present the associated postnatal findings on cardiac Computed Tomography (CT).

Case report

A pregnant mother was referred to our center for evaluation of her fetus at 19 weeks of gestation due to abnormal cardiac findings on obstetrical ultrasound screening. Fetal echocardiogram revealed the diagnosis of tetralogy of Fallot with a large malalignment ventricular septal defect, right ventricular hypertrophy, overriding aorta and pulmonary stenosis. The branch pulmonary arteries were noted to be hypoplastic, and a patent ductus arteriosus was not found. In addition, the left pulmonary artery was noted to be anomalous in that it arose from the ascending aorta rather than from the main pulmonary artery (hemitruncus) (Figures 1 A-B).

The patient was delivered at term by normal vaginal delivery with a weight of 2.855kg. Heart rate was 121 bpm and oxygen saturations were 95% on room air. Physical examination revealed normal first and second heart sounds and a grade 2/6
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systolic ejection murmur over the pulmonary area. Peripheral pulses were normal. Postnatal echocardiogram revealed the classic findings of tetralogy of Fallot, presence of a persistent left-superior vena cava to a dilated coronary sinus, right aortic arch with mirror image branching and the presence of the anomalous origin of the left pulmonary artery from the ascending aorta (Figure 2). There was mild pulmonary stenosis with a peak pressure gradient of 30 mmHg.

Cardiac CT with angiography was performed using an EKG-gated multidetector CT scanner with modulation technique followed by 3D reconstructions. CT scan further documented the anomalous origin of the left pulmonary artery which measured approximately 5 mm in diameter (figures 3 A-C). Additional work-up included a karyotype that was normal and FISH for 22q11.2 deletion that was negative.

The patient underwent surgical repair at 2 months of age with complete tetralogy repair and re-implantation of the left pulmonary artery into the main pulmonary artery by end to end anastomosis. The patient had an uneventful postoperative course. The left pulmonary artery, however, remained patent with no evidence of stenosis at 9 months follow-up.

Discussion

Hemitruncus was first described by Fraentzel in 1868. Although this is usually an isolated anomaly, it is often associated with other congenital heart disease malformations including: tetralogy of Fallot, interrupted aortic arch type A, aorto-pulmonary window and coarctation. The largest series and review of this lesion was published by Kutsche and Van Mierop in 1988 summarizing a total of 108 cases that included 89 patients with anomalous right pulmonary artery and 19 with anomalous left. It is important to note that although left hemitruncus is less common than right hemitruncus, the former is the more commonly associated lesion with either tetralogy of Fallot or right aortic arch, as was the case in our patient. To date, 17 cases of tetralogy of Fallot with left hemitruncus have been described in the literature.
It is important to distinguish this anomaly from discontinuous pulmonary arteries, where blood supply to one or both pulmonary arteries originates from major aorto-pulmonary collaterals or from the ductus arteriosus. The latter lesion is commonly associated with tetralogy of Fallot and is a common lesion in patients with DiGeorge Syndrome. To the contrary, hemitruncus is extremely rare in patients with this syndrome.

Early repair of this lesion is important to improve survival, which has been reported to be as low as 30% if left untreated. In addition, recognition of this lesion prior to tetralogy repair is essential to prevent disastrous outcomes. If left untreated, the pulmonary bed is vulnerable to early onset of pulmonary vascular obstructive disease due to the large blood supply to both lungs: one because it receives blood at systemic pressure from the aorta and the other because it receives the entire right ventricular cardiac output, unless it is protected by significant pulmonary stenosis. Therefore, we believe that early detection of this anomaly on fetal echocardiography was very helpful in contemplating early repair in our case and for providing appropriate counseling for the family at the time of diagnosis.

Cardiac CT using an EKG-gated multidetector scanner is a useful noninvasive imaging modality to evaluate this lesion. We currently prefer this modality to cardiac catheterization unless a potential intervention is required.

Nathan et al. reported the results of repair of hemitruncus on a series of 16 patients with 93% survival at 20 years and low incidence of re-operation or re-intervention. Similar favorable long-term results were also reported even in those with associated lesions including Tetralogy of Fallot. We believe these patients should be referred for early repair and hence the importance of early detection of this rare malformation.

References
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Do you or your colleagues have interesting research results, observations, human interest stories, reports of meetings, etc. that you would like to share with the congenital cardiology community?

Submit a brief summary of your proposed article to: RichardK@CCT.bz

The final manuscript may be between 400-4,000 words, contain pictures, graphs, charts and tables.

www.CongenitalCardiologyToday.com
This year the International Workshop on Interventional Pediatric Cardiology, directed by Mario Carminati, Gianfranco Butera and Massimo Chessa, and the International Symposium on the Hybrid Approach to Congenital Heart Disease, directed by John Cheatham, Mark Galantowicz and Sharon Hill, held a joint meeting in Milan, Italy.

At the event, more than 600 participants from all over the world gathered. Europe was the most representative region; followed by America, Asia and Africa.

Within Europe, Italy provided the highest number of participants (320), followed by the United Kingdom, Germany and France.

Among the other countries, the United States sent the most significant number of participants, followed by Brazil and Canada. As per the 83 faculty members, they came mostly from Italy and the United States, although over 16 countries were represented.

Due to the different approaches to congenital heart disease, the Joint Meeting directors agreed to keep the two meetings separated: the IPC, focusing on the percutaneous approach, took place during the first three days while the ISHAC, focusing on the hybrid approach, took place on the fourth day.

As in the previous editions, the first day of the IPC was dedicated to the training of junior doctors, nurses and technicians. The Sim-Suite Catheter Simulation System was available for all juniors and fellows who, under the close supervision of Marco Papa, had the chance to experience the simulation of ASD/PFO closure.

The Workshop for Physicians investigated all interventional procedures: transesophageal and intracardiac echocardiography (Ornella Milanesi, Marteen Witsenburg), 3D Echo (Achi Ludomirsky), MRI-CT scan (Pierluigi Festa), PDA closure (Caroline Ovaert) with a focus on premature babies (Neil Wilson), ASD/PFO closure (Jo DeGiovanni), VSD closure (Massimo Chessa), aortic (Sandra Giusti) and pulmonary (Jochen Weil) valvuloplasty, RF perforation in pulmonary atresia (Gianfranco Butera), pulmonary arteries angioplasty/stenting (Marc Gewillig), coarctation/angioplasty stenting...
(Shakeel Qureshi), embolizations (Marteen Witsenburg), percutaneous pulmonary valve (Mario Carminati), and retrieval techniques (Andras Szatmari).

The Workshop for Nurses and Technicians focused on: the preparation of patient candidates for interventional and procedures (Viorika Zarnescu), patient’s monitoring (Antonella Villa), sedation/anaesthesia (Marco Ranucci), x-ray exposure/protection (Marta Pasquato), materials for the most common procedures (Marina Battista), logistics of materials in the catheterization lab (Eugenio Opipari), the scrub nurse (Manuela Bonci), best angiographic projections for different procedures (Angelika Grundler), contrast media (Roberto Cristinelli), preparation of pediatric population for catheterization procedures (Tiffani MacDougall), and the hybrid approach from the nurse point-of-view (Sharon Hill).

This year, the IPC directors particularly stressed the importance of the psychosocial support which must be given to patients (both children and adults) with congenital heart disease as well as to their parents.

Later on in the day, the sponsoring companies presented to the audience their recent achievements and products, and the Italian Charity Association for Children with Congenital Heart Disease (Associazione Bambini Cardiopatici nel Mondo) gave a presentation of its successful projects based worldwide.

The work day ended with the traditional “Cheese & Wine,” with fine Amarone wine from the famous Valpolicella region.

Day 2 was the official start of the Congress: besides lectures, 10 live cases were performed from San Donato, Warsaw, Massa and Sant’Ambrogio in Milan.

The first scientific session focused on fetal interventions for aortic stenosis and for RVOT obstructions (Gerald Tulzer), neonatal critical aortic stenosis (Gabriella Agnoletti), and pulmonary atresia with IVS (Lee Benson). During Session 2, the topic was obstruction of pulmonary arteries,
pulmonary veins and systemic veins. In this context the focus was the technique for cutting balloons (Marc Gewillig), complications and how to prevent or to avoid stenting of pulmonary arteries (Frank Ing), stenting pulmonary veins (Neil Wilson) and stenting systemic veins (Frank Ing).

ICE monitoring of ASD/VSD closure (Zyiad Hijazi: he was not physically present but sent his presentation), how to close complex ASD (Carlos Ruiz), an analysis of long term follow up of ASD closure (Zahid Amin), congenital VSD closure (John Bass), and different ways to close a PDA (Carlos Pedra) were the content of Session 3 lectures.

Session 4 focused on catheter interventions in adults (Kevin Walsh), late post surgical sequelae for different congenital heart defects (Massimo Chessa) and catheter interventions in ACHD patients (Michael Landzberg).

The IPC traditional session on “Nightmares in the Cath Lab” ended the second working day; the audience had the chance to hear from Gabriella Agnoletti, Paolo Guccione, Caroline Ovaert, Evan Zahn, and Kevin Walsh the presentation of a particularly difficult experience faced in the cath lab.

The evening social event for all faculty members was a magnificent dinner at the “Terrazza Martini”, the most exclusive restaurant in Milan located on the 15th floor of a skyscraper right in the historic centre of the city.

The restaurant atmosphere was simply speechless!

The extraordinary location of this restaurant allows its guests to have a 360° view of the city’s major monuments as the Duomo, the Vittorio Emanuele Gallery, the Sforzesco Castle and the Velasca Tower.

All IPC faculty members wrote short greeting messages to Mario Carminati on a classic white table cloth, where later a specialized craftsman hand embroidered them.

On Day 3, Session 6 started with a session on PFO: the first two lectures focused on “PFO and Stroke” with both the neurologist’s (Gianpaolo Anzola) and the
interventionist’s (Neil Wilson) point-of-view; the last two lectures
focused on “PFO and Migraine” with the neurologist (Domenico
D’Amico) and the interventionist (Jo DeGiovanni) point of view. An
update on available devices (Horst Sievert) and the presentation of
the Italian PFO Registry (Gianfranco Butera) were also given.

Nine live cases were performed from San Donato, Berlin and
Naples.

Session 7 focused on RVOT dysfunction with lectures on: timing for
percutaneous/surgical treatment (Alessandro Frigiola),
percutaneous pulmonary valve replacement (Mario Carminati),
pulmonary valve implantation as a palliative procedure (Philipp
Lurz), the surgical point of view (Alessandro Giamberti),
engineering aspects (Silvia Schievano) and future perspectives
(Philip Bonhoeffer).

Session 8 focused on aortic and mitral valves: valve Edwards
(Giuseppe Sangiorgi), valve CoreValve (Horst Sievert),
percutaneous mitral valve repairs (Carlos Ruiz), paravalvular leaks
(Donald Hagler) and, finally, the surgeon’s point-of-view (Ottavio
Alfieri).

During Session 9 the theme was the treatment of aortic coarctation:
balloon angioplasty vs stenting in children (Jochen Weil), results
and complications after stenting (William Hellenbrand), Middle
Aortic Syndrome and other unusual coarctation (Grazyna
Brzezinska-Rajszys, who did her presentation directly from
Warsaw).

Finally, a completely new session: the IPC Award. The Committee
had selected beforehand five presentations on interventional
procedures among those submitted by worldwide junior doctors
and fellows. The winner, Claudio Capelli from the University
College of London, lectured on new percutaneous pulmonary valve
device: he will be part of the 2011 IPC Faculty.

The IPC & ISHAC Gala Dinner location was the magnificent Villa
San Carlo Borromeo: new this year was the amazing show by the
Sonic, the acrobats who performed during the Tourin 2006 Olympic
Games.

Also, a great violinist who’s also a model for major fashion stylists,
played her music throughout the dinner.

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2009 Annual Meeting of the Western Society of Pediatric Cardiology

By Carl Y. Owada, MD

The Western Society of Pediatric Cardiology (WSOPC) recently convened for a three-day conference hosted by the cardiology group at Children’s Hospital Central California. Over 120 participants were treated to world class presentations provided by 26 faculty drawn from the wealth of talent within the society membership, in addition to three invited guest speakers. The WSOPC is a vibrant organization representing cardiologists and surgeons involved in the care of children, adolescents and young adults with congenital heart disease in 12 western states.

The meeting was held at Tenaya Lodge at the gate of Yosemite National Park. With the exception of several rogue surgeons, unseasonable rain kept the audience focused on science instead of the hiking trails.

Dr. Antonella Rastelli (Washington University, St. Louis) delivered the keynote address entitled “My Father Dr. Giancarlo Rastelli.” Through her journey of family discovery we came to know Dr. Rastelli not only as a prolific and honored surgeon whose pioneering work continues to shape pediatric cardiology, but also as a loving husband, devoted father, accomplished theater actor, and passionate researcher up until his untimely death when Antonella was only 4 years old. In his honor, Antonella has established an educational and training fund (www.italiausa.net/Rastelli/main.htm) to encourage young and talented investigators.

During the surgical session we learned about current concepts of the hybrid approach to congenital heart disease palliation. Dr. Mark Galantowicz (Nationwide Children’s Hospital) focused on the hybrid staged treatment of Hypoplastic Left Heart Syndrome. In his impressive series there were over 100 patients who had undergone the hybrid stage 1 procedure, 60 completed the comprehensive stage 2 procedure and 25 completed the final stage Fontan. In selected patients, stage 1 survival exceeded 97%. The learning curve has been steep with challenges in identifying potential retrograde aortic arch stenosis and providing sufficient atrial communication. Dr. Galantowicz stressed the need for collaboration between surgeons and cardiologists for a successful hybrid approach.

Drawing from the experience of over 400 extracardiac Fontan procedures performed at Stanford and affiliated programs, Dr. Frank Hanley presented their current concepts and outcomes. The procedure has evolved to one performed “off pump” with passive IVC decompression without an oxygenator. There is no
need to fenestrate. The majority of patients are extubated in the operating room. Mid-term results are very promising with very low mortality and Fontan failure rates.

Dr. Mark Rodefeld (Indiana University School of Medicine) challenged the concept of the staged univentricular repair. He proposed the conceptual model of an infant Fontan. The Fontan circulation is augmented by the Rodefeld bilateral, bidirectional spinning disk pump. This pump, which could conceivably be introduced percutaneously, would provide just enough forward flow into the lungs to guard against systemic venous congestion and augment ventricular preload. The infant Fontan avoids years of hypoxemia, volume overload, and repeated myocardial insult.

The interventional session was highlighted by considering percutaneous valves, ductal stents, and occlusion devices, all brought together in a hybrid lab of the future. Although we continue to rely on “off-label” use of existing devices and medications to treat our patients, the future holds promise for product development specific to the treatment of congenital heart disease. Interventionalists are limited only by their imagination.

The fellows’ abstract competition started the second day. Six papers were presented:

1. Christopher Talluto (Lucille Packard Children’s Hospital at Stanford) Aortic regurgitation in patients with Tetralogy of Fallot, pulmonary atresia and major aorto-pulmonary connections.
2. Sarena Teng (Denver Children’s Hospital) Experience with Propofol as a bridge to extubation in children with congenital heart disease.
3. Phillip Chang (Children’s Hospital Los Angeles) Amiodarone versus Procaainamide for acute treatment of recurrent supraventricular tachycardia.
5. Nadine Choueiter (Seattle Children’s Hospital) A prospective open-label trial of etanercept as adjunctive therapy for Kawasaki Disease.
6. Darren Berman (Children’s Hospital Los Angeles) Prenatal screening for major congenital heart disease: superiority of the outflow tract views over the four-chamber view.

After careful consideration of scientific merit, study design, and quality of delivery, the 1st place was awarded to Dr. Choueiter and the 2nd place to Dr. Chang.

We learned from Dr. Mike Schaffer (University of Colorado, Denver) that if you’re not feeling well and have chest pain, you probably DON’T need an ICD. More importantly as we come to understand mechanisms of rhythm disturbances, genomic characterization more and more will come to serve as a proxy for predicting disease severity.

For information on PFO detection go to: www.spencertechnologies.com
There is a reason we have a conduction pathway. Dr. Anne Dubin (Stanford) showed very eloquently that the closer we can mimic this pathway with biventricular pacing the better chance we have at preserving myocardial function.

The who, when, where and how of ablation by the 411 of electrophysiologists was presented by Kevin Shannon (University of California, Los Angeles). It was made very clear that issues of frying, freeze thawing, or shocking parts of the heart should be left to the experienced operator.

We reviewed the roll of MRI, CT, 2-D and 3-D echocardiography and good old fashioned angiography on the characterization of congenital heart disease. We challenged the presenters to find which modality is favored by the surgeons. I am in agreement with Dr. David Teitel (University of California, San Francisco) that catheter-based angiography is best in that subtle anomalies can be identified and therapeutically addressed using a single modality. Furthermore, the ability to assess hemodynamics cannot be overstated. Contrary to popular belief the dinosaur lives!

Understanding the molecular and genetic basis of heart disease will guide us to more specific therapy. Exciting research in multi-genetic interaction in the mouse model has provided insight into a spectrum of congenital malformations. As an example Ching-Pin Chang (Stanford) presented his findings of Pbx1-null mutation causing persistent truncus arteriosus, and aberrant carotid and subclavian arteries in the mouse. The ability to turn off or turn on genes to arrest cono-truncal abnormalities may be standard therapy for tetralogy of Fallot in the near future. Bringing the bench-top to the bedside, Michael Portman (Seattle Children's Hospital) presented his work on targeted therapy for Kawasaki Disease. It is well recognized that TNFa is elevated in children with KD. The idea is to provide a safe antagonist that may reduce coronary artery inflammation. Early results suggest that etanercept reduces the refractory and re-treatment rate following IVIG therapy in acute KD. These findings have spawned a randomized, placebo controlled, prospective study to evaluate etanercept in KD. This is the first novel treatment for KD in almost 20 years.

During the gala dinner Dr. Kavin Desai (Kaiser, Hayward, CA) challenged us with the notion that outcome measures need to be more than just mortality and morbidity rates. He presented the success of camps dedicated to patients combating congenital heart disease; a place where kids come to discover that they are not alone, and where they are empowered with knowledge and self confidence. These camps also provide a venue for parents and siblings to share their mutual experiences, fears, frustrations and coping mechanisms. We heard first hand from Camp Taylor kids who are not a ‘Fontan,’ not a ‘Tetralogy,’ and not a ‘Pacemaker,’ but individuals with great dreams and aspirations.

The challenges we face in the treatment and management of adults with congenital heart disease are daunting but not insurmountable. Focusing on the adult Fontan, Jamil Aboulhosn (University of California, Los Angeles) stressed the need for vigilant follow-up and monitoring for arrhythmias, thromboembolic risk, liver damage and protein-losing enteropathy. Adjunctive therapy with multi-site single ventricle pacing, aggressive anticoagulation, and use of pulmonary vaso dilators to reduce Fontan pressures may improve outcome. The transition of the repaired or palliated patient from the pediatric setting to the adult setting cannot start too early. Alison Meadows (University of California, San Francisco) suggested that patient transition educational can start as early as 14 to 15 years old.

The meeting ended with Dr. Julien Hoffman’s vision of the future. The time of observation is coming to an end and the time of understanding is just beginning.

Through genetic and microbiologic research the understanding of disease mechanism will be made more clear thus facilitating targeted therapy. Tissue engineering will allow us to grow our own replacement valve or even a replacement heart. These exciting advances in understanding mechanisms and technology are minuscule in the shadow of a looming public health crisis.

Unless we make a dramatic change toward a healthier lifestyle, there will be an entire generation of young patients battling coronary artery disease, diabetes and heart failure. We need to move from a primarily reactive discipline to a proactive/preventative discipline.

We are looking forward to the 2010 Annual Meeting of WSOPC which is in the early planning phase for Phoenix, Arizona.
These DVDs are two little gems! For reference and for teaching, most angiographers are accustomed to thumbing through the pages of one or the other editions of the Toronto Group’s atlas. Taking nothing away from that fine work, these DVDs from Dr. Nihill and the Houston Group raise the bar. The main reason is hundreds of high quality still frames and videos of angiograms! It’s like visiting “Catheterization Conference” in Houston!

The Atlas is the primer. For cardiology and radiology fellows, it provides an introduction and an overview to angiography of congenital heart disease. The Atlas is also full of detailed information and commentary of interest to experienced angiographers. Furthermore, it provides a quick, interactive, visual reference demonstrating most congenital defects (both common and rare). Basic chapters cover: Principles of Angiography, Contrast Agents, Normal Anatomy, and Anomalies. Anomalies are divided into sub-chapters on Systemic Veins, Right Atrium, Tricuspid Valve, Right Ventricle, Ventricular Septum, Pulmonary Valve, Tetralogy of Fallot, Pulmonary Atresia/VSD, Pulmonary Arteries, Pulmonary Veins, Left Atrium/Mitral Valve, Left Ventricle, Aortic Stenosis, Coronary Arteries, Aorta & Branches, Truncus Arteriosus, Transposition of the Great Arteries, Congenitally Corrected Transposition, Single Ventricle, A-V Malformations, and Conjoined Twins. Another truly outstanding feature of the Atlas is the accessibility of all the angiogram files to be copied for personal use. Every still frame and every video may be “borrowed” for use on personal power point or other educational projects. This feature alone more than justifies picking up a copy of the Atlas for your professional library!

The DVD focusing on congenital heart disease in the adult is a similarly valuable reference. It is aimed at cardiology fellows and internist cardiologists who care for adults with congenital heart disease. It provides excellent initial chapters introducing both the patient population and the unique angiographic techniques appropriate for this population’s structural heart defects and post-operate anatomy. There are excellent chapters on the anomalies: systemic veins, pulmonary veins, tricuspid valve, pulmonary valve, atrial septal defects, ventricular septal defects, patent ductus arteriosus, aortic stenosis, coronary anomalies, tetralogy of Fallot, The Rastelli operation, transposition of the great arteries, congenitally corrected transposition, single ventricles, The Fontan operations, Pulmonary hypertension, and shunts.

Both DVDs are intuitive and easy to navigate. Both have a similar search capability allowing the viewer to select a key work or a page, and go directly to the portion of the DVD devoted to the related materials. These DVD’s are designed for viewing on personal computers. The Atlas requires Windows 95 or later, and the Adult Congenital Heart Disease DVD requires Windows 98 or later.

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Medical News, Products and Information

FDA Panel Recommends Approval of Medtronic Heart Valve under Humanitarian Device Exemption

The US Food and Drug Administration (FDA) panel unanimously vote (12-0) to recommend approval with conditions of a novel medical device that addresses the effects of a dysfunctional pulmonary valve without opening the chest for surgery. The pulmonary valve connects the heart to the lungs, where blood is enriched with oxygen before being pumped to the rest of the body.

The FDA’s Circulatory System Devices Panel recommended conditional approval of a Humanitarian Device Exemption (HDE) for the Melody Transcatheter Pulmonary Valve, by Medtronic, Inc. for the treatment of children and adults with congenital heart disease. The FDA usually follows the recommendations of its expert panels. Devices approved under HDE are intended to serve patient populations of fewer than 4,000 per year in the United States.

“The Melody valve is an enormous breakthrough – and an enormous relief – for patients with congenital heart disease,” said pediatric cardiologist Dr. William E. Hellenbrand of the New York-Presbyterian Morgan Stanley Children’s Hospital and Professor of Clinical Medicine at Columbia University Medical Center. “The device offers the potential to break the cycle of open-heart surgery after open-heart surgery. If the FDA follows the advice of its expert panel, physicians and patients will have a powerful new option for these very sick and hard-to-treat patients.”

Transcatheter valve (TCV) technology provides a less invasive means to replace a failing heart valve. It is designed to allow physicians to deliver replacement valves via a catheter through the body’s cardiovascular system, eliminating the need to open the chest. TCV technology can thereby delay open-heart surgery for valve replacement. Medtronic is committed to leading the development of TCV technology for all four valves of the heart: aortic, mitral, pulmonic and tricuspid.

The Melody Transcatheter Pulmonary Valve is the first transcatheter heart valve to be approved for commercial use anywhere in the world and the first to be reviewed by an FDA panel. It received Conformité Européenne (CE) mark, the European regulatory approval, in October 2006, and has been approved for sale in Canada since December 2006. To date, nearly 1,000 patients worldwide have benefited from the Melody Transcatheter Pulmonary Valve. With today’s panel recommendation, the device is on track to become the first transcatheter heart valve of any kind to receive FDA approval for use in the United States.

The panel’s recommended conditions of approval include a post-approval study, revision of the patient brochure and instructions for use, and implementation of a physician training and education program.

Patients with a dysfunctional pulmonary valve tire easily, as the heart over-exerts itself trying to get oxygenated blood throughout the body. The condition has traditionally required opening the chest for heart surgery, which is associated with discomfort and significant risks for the patient.

The Melody Transcatheter Pulmonary Valve gives physicians and patients the option to delay the next open-heart surgery. The device is intended to ensure blood flow from the right ventricle to the lungs, and ultimately to the rest of the body. The Melody system may also reduce the cost of treatment by avoiding surgical complications, postoperative intensive care and extended hospital stays.

“We are very pleased with the FDA panel’s recommendation today and look forward to working with the FDA to address labeling and any remaining issues as quickly as possible so that more congenital heart disease patients in the US can benefit from this technology,” said cardiac surgeon Dr. John Liddicoat, VP & General Manager of the Structural Heart division, part of the CardioVascular business at Medtronic. “By providing a less invasive option and therefore delaying even one open-heart surgery, we can improve the lives of thousands of congenital heart disease patients.”

Following the panel recommendation, the Melody Transcatheter Pulmonary Valve remains an investigational device in the US where its use is limited to a clinical trial approved by the FDA. Approval of the Melody system as an HDE is under review by the FDA.

For more information, visit www.medtronic.com.

Cardiology 2010 - The 13th Annual Update on Pediatric and Congenital Cardiovascular Disease, An Innovative Track for Trainees and Junior Faculty

When Cardiology 2010 convenes February 10-14, 2010 at Disney’s Contemporary Resort in Orlando, it will feature a unique curriculum designed for fellows and junior faculty. Expanding this popular feature from previous years, the “Early Career Track” is being offered in addition to the usual symposia, research presentations and small-group breakout sessions. Each day, breakout sessions will be dedicated to both the professional and personal aspects important in the early stages of a career in cardiovascular or intensive care medicine. Program directors and division chiefs from across the country will lead the small-group discussions.

Guidance will be offered on rarely discussed topics such as job interviews, crafting academic, research or private practice careers, financial security, work-life balance, choosing a mentor and being a good mentee. On Wednesday, Feb. 10, a special (optional) pre-conference will be followed by a plenary session on aortic valve disease and a meet-and-greet reception for residents, fellows and junior faculty. The informal reception will allow trainee track attendees to get to know the internationally acclaimed faculty who are established members in the field. This unique opportunity, open only to early career track attendees, will provide an opportunity for one-on-one conversation in a relaxed setting.

The Young Investigator Abstract Competition is back for another year. Selected abstracts will be presented at poster sessions throughout the conference, and the top six abstracts will be presented as oral abstracts. The author of the abstract chosen as most outstanding will receive free tuition to Cardiology 2011 in Scottsdale, AZ, and a $500 travel grant.

Course Highlights:

• Cardiac pathology sessions led by Paul M. Weinberg, MD, Cardiac Center, The Children’s Hospital of Philadelphia, and Robert H. Anderson, MD, Medical University of South Carolina
• A plenary session highlighting clinical trials from around the world including the latest cardiac research from the Pediatric Heart Network
• Hands-on electrophysiology and echocardiography sessions
• And, of course, the now famous annual Cardiology Jeopardy
Embryo’s Heartbeat Drives Blood Stem Cell Formation

Newswise - Biologists have long wondered why the embryonic heart begins beating so early, before the tissues actually need to be infused with blood. Two groups of researchers from Children’s Hospital Boston, Brigham and Women’s Hospital, and the Harvard Stem Cell Institute (HSCI) — presenting multiple lines of evidence from zebrafish, mice and mouse embryonic stem cells — provide an intriguing answer: A beating heart and blood flow are necessary for development of the blood system, which relies on mechanical stresses to cue its formation.

Their studies, published online by the journals Cell and Nature, respectively, on May 13, together offer clues that may help in treating blood diseases such as leukemia, immune deficiency and sickle cell anemia, suggesting new ways scientists can make the types of blood cells a patient needs. This would help patients who require marrow or cord blood transplants, who do not have a perfect donor match.

One team, led by Leonard Zon, MD, of the Division of Hematology/Oncology at Children’s and Director of its Stem Cell research program, used zebrafish, whose transparent embryos allow direct observation of embryonic development. Publishing in Cell, Zon and colleagues discovered that compounds that modulate blood flow had a potent impact on the expression of a master regulator of blood formation, known as Runx1, which is also a recognized marker for the blood stem cells that give rise to all the cell types in the blood system.

Confirming this observation, a strain of mutant embryos that lacked a heartbeat and blood circulation exhibited severely reduced numbers of blood stem cells. Further work showed that nitric oxide, whose production is increased in the presence of blood flow, is the key biochemical regulator: Increasing nitric oxide production restored blood stem cell production in the mutant fish embryos, while inhibiting nitric oxide production led to reduced stem cell number.

Zon and colleagues went on to demonstrate that nitric oxide production was coupled to the initiation of blood stem cell formation across vertebrate species. Suppression of nitric oxide production in mice, by either genetic or chemical means, similarly reduced the number of functional Runx1-expressing blood stem cells.

“Nitric oxide appears to be a critical signal to start the process of blood stem cell production,” says Zon, who is also affiliated with the HSCI. “This finding connects the change in blood flow with the production of new blood cells.”
The second team, publishing in Nature, was led by George Q. Daley, MD, PhD, Director of the Stem Cell Transplantation Program at Children's Hospital Boston, and Guillermo García-Cardeña, Director of the Laboratory for Systems Biology of the Center for Excellence in Vascular Biology at Brigham and Women's Hospital, along with scientists from the Indiana University School of Medicine. Intrigued by the appearance of blood progenitors in the wall of developing aorta soon after the heart starts beating, they investigated the effects of mechanical stimulation on blood formation in cultured mouse embryonic stem cells.

They showed that shear stress — the frictional force of fluid flow on the surface of cells lining the embryonic aorta — increases the expression of master regulators of blood formation, including Runx1, and of genetic markers found in blood stem cells. Shear stress also increased formation of colonies of progenitor cells that give rise to specific lineages of blood cells (red cells, lymphocytes, etc.). These findings demonstrate that biomechanical forces promote blood formation.

Daley, García-Cardeña and colleagues also studied mouse embryos with a mutation that prevented initiation of the heartbeat. These embryos had a sharp reduction in progenitor blood cell colonies, along with reduced expression of genetic markers of blood stem cells. When specific cells from the mutant embryos were exposed in vitro to shear stress, markers of blood stem cells and numbers of blood cell colonies were restored.

Finally, the team showed that when nitric oxide production was inhibited, in both cell cultures and live mouse embryos, the effects of shear stress on blood progenitor colony formation were reduced.

“In learning how the heartbeat stimulates blood formation in embryos, we’ve taken a leap forward in understanding how to direct blood formation from embryonic stem cells in the petri dish,” says Daley, who is also affiliated with the HSCI.

“These observations reveal an unexpected role for biomechanical forces in embryonic development,” adds García-Cardeña. “Our work highlights a critical link between the formation of the cardiovascular and hematopoietic systems.”

The authors of the two papers speculate that drugs that mimic the effects of embryonic blood flow on blood precursor cells, or molecules involved in nitric oxide signaling, might be therapeutically beneficial for patients with blood diseases. For example, nitric oxide could be used to grow and expand blood stem cells either in the culture dish or in patients after transplantation.

Trista North, PhD, and Wolfram Goessling, MD, PhD, now principal faculty at HSCI and Assistant Professors at Beth Israel Deaconess Medical Center and Brigham and Women’s Hospital, respectively, were first authors on the Cell paper. Luigi Adamo, MD, of Brigham and Women’s Hospital and Olaia Naveiras, MD, PhD of Children’s Hospital Boston and Brigham and Women’s Hospital were first authors on their study.

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