

**May 2010****In This Issue**[Regional News from the United States](#)[Regional News from Asia Pacific](#)[Regional News from Europe](#)[Regional News from Latin America](#)[Regional News from Canada](#)[Nursing Network News](#)[Case Report](#)[Journal Watch](#)**Join ISACHD Today!****JOIN NOW****International Society for Adult Congenital Heart Disease****ISACHD May 2010 Newsletter****President's Message**

The ISACHD newsletters will be distributed monthly. This will allow members to know what's going on in the global-world of congenital heart disease. Every month the newsletter will include a President's message, regional news from all parts of the world, especially for announcement of upcoming symposia and congresses and a Journal Watch.

Furthermore, case reports, announcements, requests for participation or ideas on various topics will be included, depending on your input and your wishes.

We would like to have your opinion about the way you would like to see these upcoming newsletters. For example, what format would you prefer for the Journal Watch? Are you interested in the abstracts? Do you prefer a short selection of all manuscripts that have been published or would you like to see them all (probably around 200 articles)? Are you interested in comments from experts? How do you read the newsletters? Do you print the newsletters to read them in the train, car, airport or wherever on hard copy? Or would you prefer a short overview of announcements with links to larger documents?

In order to improve and optimize the monthly newsletters your input is extremely important.

Please fill out the short questionnaire at <http://www.surveymonkey.com/s/SKG2Y6D> to provide us your feedback.

**Regional News from the United States**

1. The upcoming [Skamania Lodge ACHD Symposium](#) June 6-9, 2010 co-chaired by David Sahn, Erwin Oechslin, Gary Webb, Craig Broberg and Pamela Miner and co-sponsored by the OHSU and the ACC. This is one of the longest running, best-established ACHD Symposia in the US dating back to origins in San Diego in the early nineties. The venue is on the Columbia River gorge on the Washington



State side but not too far east of Portland, Oregon. International travelers could choose to fly into Portland and either rent a car or take a shuttle to Skamania, or fly into Seattle and rent a car if more touring in the Pacific NW is a consideration. Skamania and The Toronto ACHD Symposium have alternated years, more recently a third site was introduced, Philadelphia in 2007 and now Cincinnati in 2011. The scheduled Skamania Lodge Symposium for 2009 was delayed by a year to facilitate attendance at the 4th World Congress of Pediatric Cardiology and Cardiac Surgery in Australia last June. For all details the website address is <http://www.ohsu.edu/xd/education/schools/school-of-medicine/gme-cme/cme/ACHD.cfm>.

2. Vision 2020 of the ACHA for planning US ACHD care for the next decade had a productive Forum in Washington, DC Friday 4/23. About 50 attended with round table input received from all 50 attendees who came from the entire U. S. and Canada. Working groups were on: 1. Program Accreditation, 2. Regional Care Planning, 3. Workforce Planning, and 4. Business Model Development. This finishes Phase I of the Vision 2020 process. In Phase II some of the documents will be revised according to input received and a major emphasis will be placed on fundraising to support Vision 2020 proposals.

## Regional News from Asia Pacific

During the next World Congress of Cardiology to be held in Beijing, China, from June 16 to 19, 2010 there will be several sessions related to Adult Congenital Heart Disease with participation of physicians and nurses members of ISACHD.



### Thursday 17th June

- **Saint Lawrence Level 4 10:30-12:00**  
Symposium 27.04 Adult congenital heart disease and surgery  
Catheter interventions for adult congenital heart disease
- **Saint Lawrence Level 4 14:00-15:30**  
Symposium 27.04 Adult congenital heart disease and surgery  
ACHD: a rapidly growing population
- **Mississippi Level 4 14:00-15:30**  
Symposium 3.04 Surgery for valve disease  
Percutaneous intervention in valve disease

### Friday 18th June

- **Saint Lawrence Level 4 14:00-15:30**  
Symposium 27.04 Adult congenital heart disease and surgery  
Atrial septal defect (ASD)

- **Thames Level 3 16:00-17:30**  
Symposium 4.04 Pulmonary circulation, other  
Pulmonary circulation: 2010
- **Saint Lawrence Level 4 16:00-17:30**  
Clinical Seminar 27.06 Percutaneous intervention  
Catheter interventions update in congenital heart disease

### Saturday 19th June

- **Niger Level 2 14:00-15:30**  
Clinical Seminar 28.01 Acute nursing care  
Acute care of adults with congenital heart disease

### Publications

#### ***Left ventricular noncompaction associated with hypertrophic cardiomyopathy and Wolff-Parkinson-White syndrome***

Luis Alday, Eduardo Moreyra, Eva Bruno, Norma Rossi, Hector Maisuls

[Abstract](#) | [Reference](#) Full Paper: [PDF](#) (Size:303K), pp.200--203

DOI: 10.4236/health.2010.23029

### Regional News from Europe

*By Folkert Meijboom*

The first Advanced Course on Grown Up Congenital Heart Disease organized by the working group on GUCH of the ESC was held in London on Wednesday 14th and Thursday 15th of April, organized by professor John Deanfield. It was held back-to-back with Cardiology in the Young, a very succesful annual course on pediatric cardiology that has been held for many years already. Despite the fact that it was not widely advertized, it was fully booked. Among the many very good presentations one stood out: the first Jane Somerville lecture, held by professor Deanfield, on Fontan; 40 years of failure or success?



Both participants and organizers were very happy with the course and it is the idea to have an annual course on GUCH in Europe organized by the ESC WG. Paris will be the next host of the event in April 2011. Until now there have been separate basic training courses on GUCH, like 2008 and 2009 in Antalya and October 2010 in Lund, Sweden and now an advanced course in London. From 2011 on there will be no longer separate meeting but one large GUCH meeting. For catheter interventions there is the annual EuroPCR, for echocardiography there is EuroEcho and the idea is to come to an annual EuroCongenital-Heart-Disease. Details of the Paris meeting will follow soon.

There are still places left for the GUCH teaching course in 22 and 23 October in Lund. It has an attractive program and an international faculty of speakers, who will assist the local organizers Johan Holm and Ulf Thilen. The website is <http://www.>

[malmokongressbyra.se/guch2010](http://malmokongressbyra.se/guch2010)

For this year's ESC meeting in Stockholm there will be an unprecedented number of 8 sessions on Congenital Heart Disease, including one Focus Session on the "dilating aortic root" organized by prof Barbara Mulder from Amsterdam, and one session entirely dedicated to the new GUCH guidelines, arranged by the chairman of the writing task force of the new guidelines, prof Helmut Baumgartner of Münster, Germany.

For those who come to the ESC there also will be a social event of the GUCH WG og the ESC; all ISACHD are very welcome to this event. Details will be announced later. For those who are planning to come to Stockholm: it is advisable to book early for the hotel accomodation, because in recent edition in Stockholm those who booked late often had to stay outside Stockholm, up to 60 km form this beautiful capital of Sweden, where the Congress venue is. Also in Stockholm, during the business meeting of the WG, Pedro Trigo Trindade from Zurich, Switzerland, will replace Folkert Meijboom form Utrecht, The Netherlands as chairman of the WG for the coming 2 years. Johan Holm from Lund, Sweden is elected as new vice-chairman; According to the new ESC policy they will be elected for a duration of 4 years (instead of 6 years), in order to enhance active particiaption of more members of the WG's nucleus.

## Regional News from Latin America



During the XXXVI Argentine Congress of Cardiology, to be held in Buenos Aires, Argentina, from October 7th to 9th, 2010, there will be a Joint Session between the ISACHD and the Argentine Society of Cardiology on "Adult Congenital Heart Disease" with the participation of Drs. Jane Somerville and Michael Landzberg, as special guests. Endorsement of EURO GUCH has also been requested.

## Regional News from Canada



Download Canadian guidelines from [www.isachd.org](http://www.isachd.org) or from [www.cachnet.ca](http://www.cachnet.ca)

## Nursing Network News

Thanks to all who submitted abstracts for the International Symposium on ACHD. Abstract submissions for International Symposium on ACHD to be held in June, 2010, Skamania resulted in 3 oral presentations and 4 poster presentations. Poster presentations will be moderated and occur during the nursing Dinner.

### Case Report

#### **Cyanosis, Clubbed Finger and Toes, and Normal Cardiovascular Findings.**

*Monica Benjamin, Marie Jose Cabrere and Luis Alday*

Section of Adult Congenital Heart Disease and Department of Cardiovascular Interventions, Sanatorio Allende, Cordoba, Argentina

A 33 year-old female was referred because of severe progressive dyspnea starting 6 months earlier, cyanosis, and marked clubbing of fingers and toes which she had had since childhood. The maternal grandmother also had clubbing.

She had a history of 3 pregnancies, the first one ended in miscarriage, and the last 2, were complicated by cyanosis and congestive heart failure near term and in the immediate postpartum period. An ischemic cerebro-vascular accident with right sided hemiparesis with complete recovery occurred following the first delivery. She also had a history of nose bleeds. A cardiological examination was advised 8 years before her first visit. She was then told that her heart was normal and received no explanation for the cyanosis.



Figure 1

On physical examination she had marked cyanosis (78% systemic saturation by pulse oximetry) and clubbed fingers and toes. (Figure 1) There were no other abnormal findings but a mild immediate diastolic murmur in the pulmonary area and along the left sternal border. A chest X-ray showed dilated and tortuous pulmonary vessels in the right upper lung lobe. There was also a small rounded imagen in the left mid lung. The heart and great arteries appeared normal. (Figure 2) The EKG was normal and a color Doppler echocardiogram revealed rapid pulmonary venous return to the left atrium. The clinical diagnosis of pulmonary arteriovenous malformation was then made.

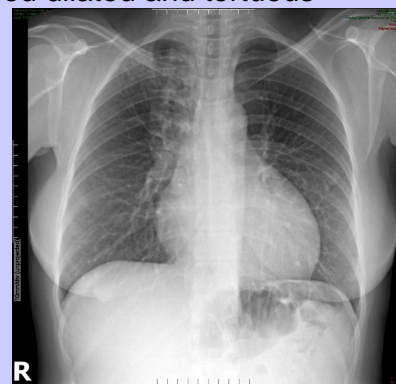
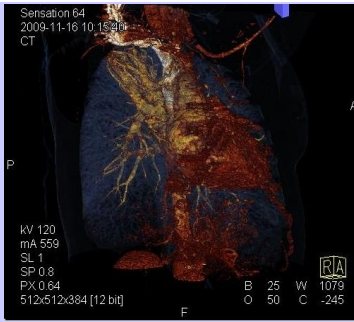


Figure 2

Multislice computed tomography and cardiac catheterization and cineangiography showed multiple severe arteriovenous pulmonary fistulae in the right upper lung

lobe and a small one in the left lung. (Figures 3 and 4)



**Figure 3**  
*Multislice computed tomography reconstruction showing a big pulmonary arteriovenous malformation in the right upper lung lobe with multiple dilated pulmonary arteries and a large pulmonary vein (outer vessel) draining into the left atrium*

Embolization of the fistulae was performed with Amplatzer vascular plugs II occluding 3 pulmonary artery branches in the right lung. In the smaller ones (5 mm diameter), we used 8 mm plugs, and in one of the larger ones (10 mm diameter) we deployed a 16 mm plug. (Figures 5 and 6) Systemic O2 saturation breathing room air increased from 78 to 91%. She was given low molecular weight heparin for 5 days and started on ASA.



**Figure 4**  
*Selective left pulmonary angiography to show the small, round, pulmonary artery malformation in the left mid lung. Reflow to the right pulmonary artery shows the big fistula as described in the computed tomography.*



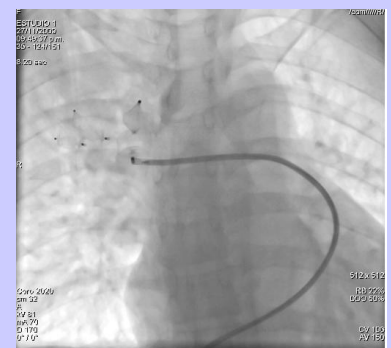
**Figure 5**  
*Following occlusion of three right pulmonary artery branches with vascular plugs the distal flow is much slower and there is no rapid filling of the pulmonary vein.*

She was discharged the day after and was seen in the outpatient clinic a week later. She felt dramatically improved. Another session of embolizations will be performed in the near future.

Pulmonary arteriovenous fistula is a rare congenital anomaly producing different degrees of cyanosis. It may complicate with paradoxical embolism, bacterial endocarditis, ruptured aneurysm and congestive

heart

failure. Our patient's history of epistaxis points to the diagnosis of hereditary hemorrhagic telangiectasia, also known as Rendu-Osler-Weber syndrome, a genetic autosomal dominant disease. The pulmonary malformation was unnoticed until adulthood despite the family history, symptoms, and signs because of the lack of cardiac physical findings. A similar situation may occur when a systemic vein drains into the left atrium. Nowadays, the preferred treatment is catheter embolization. The procedure is usually long lasting, especially if the malformations are multiple and big like in our patient, and more than a session may be required. It is convenient to occlude as many fistulae as possible because they tend to enlarge with time.



**Figure 6**  
*Vascular plugs in place in the right lung.*

## Journal Watch

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