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# ISACHD Newsletter

## President's Message

by Curt J Daniels



Dear ISACHD Members,

ISACHD ACC 2013 is history but the outstanding presentations provided not only a phenomenal educational experience but also a clear opportunity and a path to move ISACHD forward over the next several months.

Michael Maves, MD, Executive Vice President for Project Hope [www.projecthope.org](http://www.projecthope.org), delivered an informative and gripping presentation highlighting the international work of Project Hope. Please visit their website to learn more. He

closed his presentation with potential opportunities for ISACHD to partner with Project Hope to build international ACHD programs. We have follow up scheduled with Dr. Maves over the coming months to explore these opportunities.

Barbara Mulder, MD (Amsterdam, The Netherlands, ISACHD Past -President), Ariane Marelli, MD (Montreal, Canada) and I presented ACHD Program Accreditation From Around the World. We compared and contrasted program accreditation and cardiology certification criteria between Europe (contributions by Kate English, MD for the UK, Helmut Baumgartner, MD for Germany), Canada and the United States. This sparked lively discussion amongst the audience and lead to a need for follow up discussion. Each country and health care system continues to look for ways to improve the quality of care we deliver and recognize setting standards for training cardiologists and developing criteria for programs will impact how we deliver ACHD services.

Els Pieper, MD (Groningen, The Netherlands) presented an update of the ISACHD Global Education Work Group (Chair Erwin Oechslin, MD Toronto, Canada). Els highlighted the tremendous work being done to develop the ACHD eLearning Center, lead by Gary Webb, MD (Cincinnati, USA). Work is being done to develop an ACHD basic teaching course and a repository for online ACHD education. This initiative fits nicely with the Global Health agenda to support the educational development of international ACHD programs.

Lastly, Adrienne Kovacs, PhD (Toronto, Canada) presented an update from the Global Research Work Group. Adrienne provided an update for a new international multi-center study entitled Assessment of Patient-Reported Outcomes in Adults with Congenital Heart Disease - International Study (APPROACH-IS). The study design has been developed and is coordinated by Philip Moons, PhD, Koen Luyckx, PhD, Silke Apers, (Leuven, Belgium), and Adrienne Kovacs, PhD. The study utilizes a number of patient surveys to assess international differences in perceived health, psychosocial functioning, behavioral outcomes and quality of life. Sites interested in participating please contact Philip Moons.

ISACHD continues to move forward its agenda for improving ACHD Global Health, Global Education, and Global Research. Please join ISACHD, become a

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member and contribute to the work and the effort to improve ACHD care, education and research around the world.

Please visit the our website at [www.isachd.org](http://www.isachd.org) to learn more about ISACHD, visit the journal watch page and find the latest ACHD publications, view upcoming conferences endorsed by ISACHD and register to become a member of ISACHD.

Look forward to seeing you soon,

Curt J Daniels, MD, President

**Regional News****News from USA**

*By Bill Davidson*

Please take note of the **next congress in Skamania**, the 27th International Symposium on ACHD. June 2-5. Beautiful setting, great program.

Information and further details are provided at the following URL:

[www.ohsuheart.com/congenital-cme](http://www.ohsuheart.com/congenital-cme)

The **AHA** scientific sessions will be in Dallas this November 16-20. There was enough interest that the CVDY got extra CHD/ACHD sessions. So this might be of interest to some (especially if they like barbecue).

We are expecting the **first ACHD subspecialty exam** to be given in 2015.

Also, the ACHA Clinic Directory now lists 110 clinic sites in US and Canada, almost double the initial 2006 listing of 62 sites!

**News from Europe**

*By Massimo Chessa,*

The European Meeting of the GUCH- Working group, was held in Milan on March 22nd and 23rd.

Alessandro Giamberti, Mario Carminati and I, were the local organizers of this event, and we are very happy about its successful turnout, which was also the

result of an intense collaboration between all the members of the Nucleus of the Working Group supported by the presence in the Scientific Committee of Johan Holm, Chairman of the Working Group.

On the first day, the focus was on the Right Heart, including topics about the atrial septal defects, the problems of the Tricuspid valve, the RVOT, and the pulmonary arteries.

In order to improve the educational possibilities four lunch symposia were organized, which were dedicated to very precise topics, such as recreational sport activities, or particularly difficult clinical cases. During the day live interventional catheterizations were transmitted both from the Policlinico San Donato Hospital and also from other European places connected by satellite. This was possible because the Meeting was partly organized in collaboration with the now consolidated Workshop on Interventional Pediatric and Adult Congenital Cardiology that organized every two years in Milan.

The first day was concluded by the usual Jane Somerville lecture that was given by Davide Cirelli, the Vice President of the Italian GUCH Association (Davide is also a patient with a Fontan). This was a worthy tribute from the patient's point of view to the person who did a lot for GUCH/ACHD in Europe, Jane Somerville, who was present during the entire course.

The focus of the second day was the Univentricular Heart with an attempt to cover all the most significant diagnostic and therapeutic aspects with various talks. Attention was also given to burning topics such as heart transplantation in Fontan patients, the antiaggregation/anticoagulation therapy with the new drugs, and the important psychosocial aspects pertaining to these patients.

The Lunch Symposia of the day were dedicated to pulmonary hypertension and to other particularly complexed clinical cases, which were discussed in an interactive manner.

During the two days, two Lunch Symposia were organized with the Working Group of Interventional Cardiology of the AEPC, which were dedicated to the young fellows, but since topics proposed proved to be very interesting our more experienced colleagues have offered their additional insight.

The talk of Helmut Baumgartner resulted as being particularly interesting to the young fellows who are still in training as he presented the milestones, which are ideally the most linear in order to build an educational program in Interventional Procedure in GUCH/ACHD patients.

In my opinion, the main aspects of this meeting's edition should include the following:

1. The involvement of colleagues with other specific competences (interventional congenital cardiologists)
2. The collaboration with other scientific societies (AEPC)
3. The tight collaboration in the organization with surgeons who are directly involved with the handling of GUCH/ACHD, which represents a useful method to be repeated for a more institutionalized manner in the future.

The good attendance (about 400 attendees) on the first day, thanks to the contemporary presence of interventionalist colleagues, and of about 180 in the audience on the second day, the excellent quality of the talks proposed by 56 members of faculty from the EU, USA, Canada and South America, and last, but not least, the quality of the social events, have contributed to make this edition a success as the previous ones in the past.

Finally, it is noteworthy to mention the importance of this event as it received a direct endorsement by the President of the Italian Republic, Giorgio Napolitano.

All the presentations and live clinical cases will be available soon on this website <http://www.workshopipc.com/main.php>



**The 5th European GUCH course** next year will be held in Amsterdam, April 10 to 11.

**The ESC meeting** this year will be from August 31st to September 4th, and also in Amsterdam.

Please take note of the planned social event on Monday, September 2nd, which will start at Barbara Mulder's house near the Congress center, beginning with cocktails at 6:00 PM. Our dinner will be held on the canal boat.

### **News from South America**

*By Luis Alday*

The XXIV Inter-American Congress of Cardiology along with the XXXIX Argentine Congress of Cardiology will be held at La Rural Convention Center in Buenos Aires in October 18-20 this year. A Joint ISACHD - Inter-American Society of Cardiology Session is being planned.



## WG on Education

By Ervin Oechslin, Toronto (Canada)

The following meetings have been endorsed:

- **23rd International Symposium on Congenital Heart Disease in the Adult: 'Improving Long-Term Outcomes'**. June 1-5, 2013, at the Skamania Lodge outside of Portland, Oregon. This symposium is organized by Oregon Health & Sciences, University, Portland, Oregon. Organizers: Drs. Craig Broberg, David Sahn and Karen Stout. Please visit <http://www.ohsu.edu/xd/health/services/heart-vascular/for-healthcare-professionals/continuing-medical-education/heart-disease-symposium.cfm>
- **PACES Pre - HRS 2013 Symposium: Management of Arrhythmias in Adult Congenital Heart Disease**. Tuesday, May 7, 2013, Parkside Mansion, 1859 York Street, Denver, CO; Organizer: Dr. Paul Khairy, Montreal. Program available at [www.isachd.org](http://www.isachd.org) or [www.cachnet.ca](http://www.cachnet.ca)

[PACES: Pediatric & Congenital Electrophysiology Society]

Please contact me at [erwin.oechslin@uhn.ca](mailto:erwin.oechslin@uhn.ca) if you want to endorse your ACHD meeting.

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## Case Report

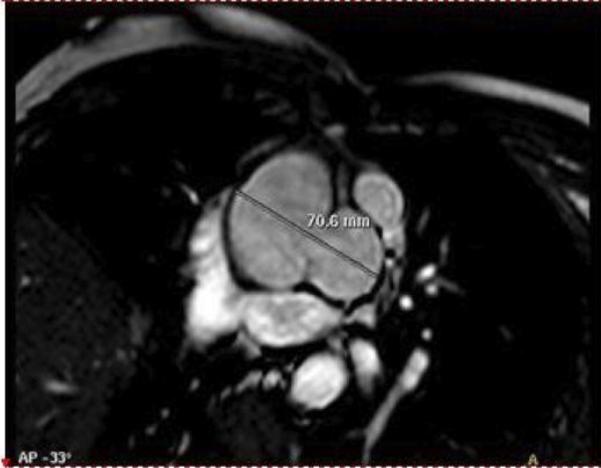
### **Unruptured giant sinus of Valsalva aneurysm long after surgery for severe congenital aortic valve stenosis with bicuspid aortic valve.**

*Ivana Seia\*, Juan Sánchez\*, Roque Córdoba\*, Leandro Marani\*, Javier Courtis#, Fernando Daghero\*, Luis Alday\**

From the \*Section of Adult Congenital Heart Disease and Cardiovascular Unit, Sanatorio Allende, Córdoba, Argentina; and the #Section of Computed Tomography and Magnetic Resonance Imaging, Oulton Institute, Córdoba, Argentina.

A 37-year-old asymptomatic male who had surgical valvotomy for severe aortic valve stenosis at the age of 5 years was seen for a routine follow-up visit. Relevant clinical features on physical examination were a BMI of 24.8, normal blood pressure, a systolic ejection click, and ejection systolic and early diastolic murmurs 3 and 1/6 on intensity respectively in the aortic area. The EKG showed left atrial enlargement and the chest x-ray a normal size heart with a dilated ascending aorta. A color flow mapping echocardiogram showed a 15 mmHg gradient across a bicuspid aortic valve and there was also trivial aortic regurgitation. The left sinus of Valsalva was markedly dilated with a width of 46 mm. There was also dilatation of the ascending aorta and mild mitral incompetence. Cardiac magnetic resonance imaging confirmed the diagnoses of a large aneurysm of the left sinus of Valsalva with a maximal length of 70.6 mm and marked dilatation of the ascending aorta.

**Fig. 1: Cardiac MR axial view of the aortic root showing the large left sinus of Valsalva aneurysm**

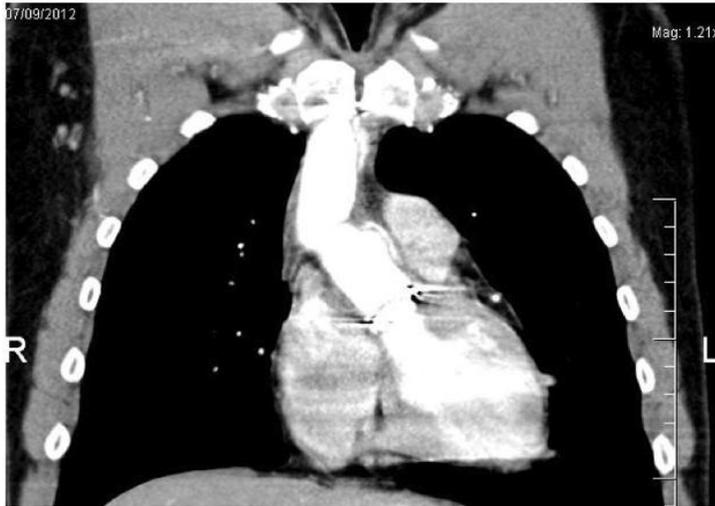


**Fig. 2: CMR reconstruction of a sagittal view of the aorta depicting the giant left sinus of Valsalva aneurysm and dilated ascending aorta**



With such diagnoses, the patient was referred to surgery undergoing resection of the aneurysm, replacement of the aortic valve and a markedly dilated ascending aorta (47 mm at the aortic root) with coronary reimplantation (Bentall-De Bono technique), with a synthetic valved aortic graft (St Jude 23) . The left anterior descending and circumflex coronary arteries had separate ostia The postoperative period was uneventful the patient being discharged seven days after surgery on oral anticoagulants, 100 mg aspirin and beta-adrenergic blockers. Nine months after surgery, the patient was completely asymptomatic and a follow-up multislice computed tomography study of the thoracic aorta, showed and excellent mid-term surgical result.

**Fig. 3: Multislice CT shows mild angulation at the site of the anastomosis of the valved graft and the normal diameter distal ascending aorta**



## Discussion

Congenital aneurysms of the sinuses of Valsalva are rare cardiac anomalies caused by a defective aortic wall media at its union with the annulus fibrosus of the aortic valve. (1) Aneurysms of the sinus of Valsalva are located underneath the aortic valve leaflets, most frequently affecting the right coronary sinus followed by the non-coronary and only in 6% of cases the left sinus. (3) Not infrequently, they are associated with other anomalies like supravalvular ventricular septal defect, bicuspid aortic valve, and other aortic lesions. (4) They are usually unrecognized until rupture following continuous growth up to the 3rd or 4th decade of life. They may present with chest pain, symptoms of heart failure, and a continuous murmur. They usually rupture into the right ventricle though they might do it in any of the other cardiac chambers. Left sinus of Valsalva aneurysms may rupture into the left pulmonary artery. (5) Nowadays the diagnosis can be readily made with non-invasive multimodality imaging.

Unruptured aneurysms may be found by chance during a diagnostic study though they may present with symptoms and signs of compression of cardiac structures resulting in ischemia, arrhythmias or murmurs. (1) Though asymptomatic, patients with unruptured aneurysms are prone to suffer adverse events, therefore, surgical treatment is usually advised. Rupture, infective endocarditis, thrombosis and embolism, fistulous communications, aortic regurgitation and other infrequent complications, have been described (3). Several anomalies were associated in our patient with bicuspid aortic valve. Aside from the unruptured left sinus of Valsalva, he also had aortic valve stenosis, separate origins of the left anterior descending and circumflex coronary arteries, and aneurysmal dilatation of the ascending aorta. This widely spread arteriopathy is known as bicuspid aortic valve syndrome secondary to a connective tissue disarrangement. (6) He underwent successful surgical treatment with both aortic valve and ascending aorta replacement with reimplantation of the coronary arteries (Bentall-De Bono procedure). (7)

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## Journal Watch

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