

November, 2012

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[In This Issue](#)

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International Society for  
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## ISACHD Newsletter

### President's Message

by Curt J Daniels



Dear ISACHD Members,

Those able to attend AHA in LA and our ISACHD meeting enjoyed an exceptional program highlighted by an elegant lecture by Dr Barbara Mulder and her team from the Netherlands. The group reported their multi-center study (Programs from Amsterdam, Toronto and Leuven) to define clinical parameters, and cause and circumstance leading to SCD in ACHD. Over 25,000 patients were evaluated and risk factors for arrhythmic SCD were reported and a risk score was presented. A lively discussion followed and future studies were discussed. Additional information will be forthcoming.

Please stay tuned.

Philip Moons, PhD, and his team from Belgium presented their proposal for A Multi-International Study on Patient Reported Outcomes in ACHD. The question raised by this study; What are the differences in perceived health, psychosocial functioning, behavioral outcomes and quality of life of adults with congenital heart disease who are living in different areas of the world? The initial goal is to enlist several programs from around the world to participate. Philip requested, ACHD programs interested in participating in this study, please contact him for more information.

Also, the ISACHD Global Health Work Groups provided updates, all of which are moving forward with great progress.

ISACHD Global Health: A small exploratory team (Mike Landzberg, Boston, MA, USA ISACHD Past-President; Paul Khairy, Montreal, Quebec, CA, ISACHD President-Elect, Disty Pearson, Boston, MA, USA, and yours truly) met, in September 2012, with the leadership of Project Hope [www.projecthope.org](http://www.projecthope.org) at their home base in Millwood, Virginia, USA. A proposal was recently sent to Project Hope outlining a partnership to provide training and develop ACHD programming in undeveloped countries/regions of the world.

ISACHD Global Education: Erwin Oechslin (Toronto, CA, ISACHD Canadian Regional Representative and Chair ACHD Global Education WG), Gary Webb (Cincinnati, OH, USA, ISACHD Past president) and their team of ACHD educators continue their development of a web-based ACHD educational program to eventually be utilized to communicate and educate our international colleagues, particularly those in underserved areas less able to attend symposia and conferences. This effort will coordinate with our global health agenda as we develop international health partners ie Project Hope.

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Additionally, our ISACHD regional representatives provided updates regarding local and regional ACHD Programming; Dr Luis Alday (South America), Dr Helmut Baumgartner (Europe), Dr Koichiro Niwa (Asian Pacific), Dr Bill Davidson (USA), and Dr Erwin Oechslin (Canada).

As described above, we are in early phases of organizing the priorities of the work groups. We will need YOU and many volunteers once we move into implementation of the initiatives. Please stay informed and contact me or any of the work group chairs to discuss your ISACHD thoughts and ideas.

If you haven't renewed your membership for this year, please do so, now is the time.

Please visit the ISACHD 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  
President

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**New Case Report**

**Infective endocarditis and systemic emboli in a 68 year-old man with bicuspid aortic valve long after coarctation of the aorta repair.**

*Ivana Seia, Marcelo Urinovsky, Alberto Marangoni, Eduardo Moreyra, Luis Alday*

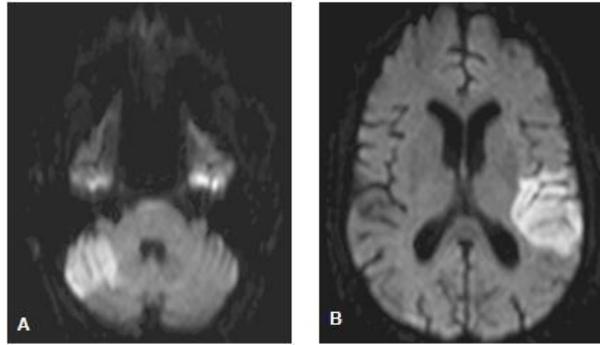
Section of Adult Congenital Heart Disease and Cardiovascular Unit, Sanatorio Allende, Cordoba, Argentina.

A 68-year-old male with a 2-week history of fever of unknown origin and back pain, presented to a hospital of a neighboring province with the clinical diagnosis of transient ischemic attack following an episode of dysarthria lasting about 5 hours. A brain CT scan and Doppler study of the neck vessels were normal. TTE and TEE ruled out the presence of heart valve vegetations and intracardiac thrombi. Blood and urine cultures were negative. The 1st hour erythrocyte sedimentation rate was 74 mm. Spinal MRI showed degenerative lumbar disc disease and a bone centellogram mild hyperdensity at 5 L level.

His past history included resection of aortic coarctation at age 13 years, systemic hypertension, tabaquism, and surgical resection of prostate cancer and lymphadenectomy followed by radiation therapy at age 57 years. Six years earlier he had an episode of paroxysmal atrial fibrillation without recurrences.

Since no diagnosis was made for the origin of his persistent fever, he was referred to an institution in our city where all studies were repeated with no changes regarding the original ones. He was put on levofloxacin 500 mg daily which he discontinued on the 3rd day.

**Figure 1. Brain MRI displaying large ischemic lesions in the right cerebellar hemisphere (A) and in the left fronto-temporal cerebral region (B)**



Two weeks later he was admitted in our hospital because of vertigo, dysarthria, and expressive aphasia. There was no motor involvement of the rest of the body. He was afebrile and had poor dental health. The heart rate and blood pressure were 75/bpm and 140/80 mmHg respectively. Femoral pulses were normal. A harsh 3/6 ejection systolic murmur was heard at the aortic area. Pertinent laboratory studies showed 89% neutrophils for a total white cell count of 6.8/mm<sup>3</sup>, erythrocyte sedimentation rate 85 mm/hour and a CRP of 4.03 mg%. The EKG showed left atrial enlargement, left anterior hemiblock, and ST-T changes compatible with left ventricular overload and/or ischemia. The chest x-ray showed left ventricular hypertrophy and dilation of the ascending aorta. The aortic knuckle was prominent and the descending aorta was tortuous. There were no lung abnormalities. A brain MRI showed two recent ischemic lesions, in different stages, located in the basal right cerebellar hemisphere and the left fronto-temporal insular region. (Fig. 1) A TEE study showed mild mitral regurgitation and vegetations on the anterior mitral valve leaflet with 8 mm maximal diameter and a smaller one on the posterior leaflet. (Fig. 2) The aortic valve was bicuspid with severe calcific stenosis (Doppler velocity 4.22 m/second, mean gradient 35 mmHg) and mild aortic insufficiency. (Figure 3) The aortic root measured 4.0 cm and there was left ventricular hypertrophy with 80% ejection fraction. The left atrium was moderately enlarged with a volume index of 40.5 ml.

Intravenous antibiotic treatment was initiated with 1.0g vancomycin plus 2.0g ceftriaxone bid which then was rotated to 24 million IU G penicillin plus 80 mg gentamicin tid when viridans streptococci were isolated from 3 blood cultures. The patient's course during admission was uneventful with improvement of the expressive aphasia. Continued medical treatment was advised. He was discharged 2 weeks later to continue ambulatory treatment with ceftriazone 2.0g bid for two more weeks. A new TEE showed no changes and laboratory acute phase-reactants were improved. Ten months later, he still had expressive aphasia recovering slowly with rehabilitation. He had no cardiovascular symptoms but he was leading a sedentary lifestyle. His last TTE showed a 0.5 mm vegetation at the anterior mitral valve leaflet, mild mitral regurgitation, left ventricular hypertrophy (145g mass index), left atrial enlargement (33 ml index volume), and severe calcific aortic stenosis and mild aortic incompetence on a bicuspid aortic valve (4 m Doppler velocity and 34 mmHg mean gradient) Aortic valve replacement was advised.

**Figure 2. TEE showing a large vegetation at the anterior mitral valve leaflet (arrow)**



**Figure 3. TEE study shows a bicuspid, calcific and stenotic aortic valve**



## Discussion

On admission to the hospital, our patient fulfilled the Duke criteria for *Definite Infective Endocarditis* with

2 major criteria present. He had echocardiographic evidence of endocardial involvement and positive blood cultures with viridans streptococcus isolation as the causative organism.

Besides, there was evidence of systemic embolization to the brain which is frequently the target organ when this complication takes place. Viridans streptococci account for about 20% of cases of infective endocarditis. They are very sensitive to penicillin which is frequently used with gentamicin because of their synergistic action. (1) This combination therapy was used successfully in our case. The port of entry was most likely the oral cavity since he had very poor dental health.

Despite having a calcified stenotic bicuspid aortic valve, which is a well known threat for infective endocarditis (2), the involved valve was the mitral. The presence of long lasting mild aortic incompetence might have caused a jet lesion on the mitral valve accounting for the valve involvement. Medical treatment was chosen since there was no hemodynamic compromise and the vegetation had less than 10 mm in diameter. Surgical replacement of the aortic valve has recently been advised.

Finally, this patient illustrates the need for transitioning and continuous care of patients with ACHD in a tertiary center. He had a nearly fatal event 55 years following aortic coarctation repair without appropriate follow-up. (3)

## References

1. Karchmer AW. Infective endocarditis. In: Bonow RO, Mann DL, Zipes DP, Libby P, eds. *Braunwald's Heart Disease*. 9th ed. Philadelphia: Elsevier; 2012 pp: 1540-58.
2. Lamas CC, Eykyn SJ. Bicuspid aortic valve-a silent danger: analysis of 50 cases of infective endocarditis. *Clinic Infect Dis* 2000; 30: 336-41.
3. Patel MS, Kogon BE. Care of the adult congenital heart disease patient in the United States: A summary of the current system. *Pediatr Cardiol* 2010; 31: 511-4.

## Regional News:

### News from United States of America

by Bill Davidson, Jr.

The PATCH program is a funded initiative of the ACHA and the ACC. It seeks to educate general cardiologists about ACHD and about where they can send patients for expert consultation within their state or nearby. The Program works with state chapters. The pilot program targets 5 states which had applied to do this: Massachusetts, Ohio, Georgia, Michigan and California.

The American Board of Medical Specialties has approved an ACHD subspecialty.

This will lead in the next 2 years or so to ACGME approved ACHD fellowships and to an ACHD subspecialty board exam with pathways for fellows graduating from ACGME-approved ACHD fellowships and for all current ACHD specialists via a 'practice pathway' for the first 3 exams which will be given every other year. This is a fairly standard approach for new subspecialty board exams. Our present and twice past ISACHD presidents had a big hand in this effort and should be congratulated for this culmination of their hard work.

The ACC (3/9-11/13) in San Francisco and the Skamania Lodge ACHD meeting in Washington State (6/2-5/13) are already on the ISACD website I believe.

### **News from Canada**

*by Erwin Oechslin*

CACH Network held its annual Business Meeting on October 28, 2012, during the Canadian Cardiovascular Congress in Toronto. In addition to the usual business items, Erwin Oechslin reported about educational activities and national and international collaboration and relationships. The CACH Network Website continues to be a great success with more than 9,000 visitors and more than 120,000 hits every month.

CACH Network has decided, in collaboration with the Canadian Cardiovascular Society (CCS) to develop a **Position Statement about Training and Maintenance of Competency in Adult Congenital Heart Disease**. We will submit the application to CCS at the beginning of 2013.

The Royal College of Physicians and Surgeons of Canada (RCPSC) introduced the Area of Focused Competence (AFC) Diploma Program in 2011 to recognize new subspecialties and established disciplines of medicine that enhance scope of practice. CACH Network is evaluating the application process for the **Area of Focused Competence (AFC) Diploma in Adult Congenital Heart Disease**.

Many Canadian Congenital Heart Disease Centres use CACH codes that were developed by Dr. Gary Webb and do not match entirely to international codes. **IPCCC** (International Pediatric and Congenital Cardiac Code) is a master code developed to translate two different pre-existing coding systems for congenital heart disease which were published in 2000: EPCC (European Pediatric Cardiac Code) and Society of Thoracic Surgeons and European Association for Cardio-Thoracic Surgery Code (STS-EACT).

The former was more robust for diagnostic terms, the latter was more robust for procedural terms. An International Nomenclature Working Group combined the terms from the EPCC and STS-EACT nomenclature lists and other nomenclature lists (e.g. CACH Network Codes) to produce a comprehensive, single nomenclature tree for pediatric and congenital heart disease: this list is called IPCCC (=master code). Terms were added to the EPCC and STS-EACT codes, now called EPCC-derived IPCCC and STS-EACT derived IPCCC.

Dr. Jack Colman (Toronto), in collaboration with Dr. Gary Webb (Cincinnati) and Graham Jericho (Toronto, developer of CAPS) has translated the CACH coding system via IPCCC to EPCC-derived IPCCC codes. The matched IPCCC are being evaluated in a test version of the CAPS database in Toronto with more than 14,000 patients. We will switch to IPCCC in Toronto by the end of December 2012.

Please visit [www.cachnet.ca](http://www.cachnet.ca) and read the President's message for more information.

## WG on Education

by Erwin Oechslin

The Editorial Board of the **ACHD e-Learning Centre** (Chairman of the Editorial Board: Dr. Gary Webb, Cincinnati) is evaluating existing educational resources, and assessing suitability of the education material for the collection on the Website. This Website will be the e-hub for educational material on ACHD, will be a great resource to find educational material and a great educational experience for care providers.

The speaker list for the **basic teaching course in ACHD** for trainees adult cardiology is being finalized under the leadership of Dr. Els Pieper, Groningen (NL).

If you have not provided your link, yet, and if you want to link **your teaching/education website to the ACHD e-Learning Centre**, please email the link to [gary.webb@cchmc.org](mailto:gary.webb@cchmc.org) and to [erwin.oechslin@uhn.ca](mailto:erwin.oechslin@uhn.ca).

## Endorsement of ACHD Meetings

The following meetings have been endorsed:

- **Sport and Heart Disease: November 30 - December 1, 2012**, German Heart Centre, Lazarettstrasse 36, Munich (Germany). This symposium is organized by the German Heart Centre, Munich; Scientific Chair: Alfred Hager, MD. Registration at [www.sport-CHD.de](http://www.sport-CHD.de)
- **9th DHZB Lange Symposium: Left Heart under Pressure: January 19/20, 2013, Berlin**. This symposium is organized by the German Heart Centre, Berlin; Organizers: Prof. F. Berger, Dr. S. Ovrutskiy, and PD Dr. L. Schmitz. Registration at [kikasymp2013@dhzb.de](mailto:kikasymp2013@dhzb.de) or [www.dhzb.de/kikasymp2013](http://www.dhzb.de/kikasymp2013)

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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## Journal Watch

J Interv Card Electrophysiol. 2012 Oct 19. [Epub ahead of print]  
[Cardiac resynchronization therapy for patients with congenital heart disease: technical challenges](#). Manchanda M, McLeod CJ, Killu A, Asirvatham SJ.

### Source

Division of Cardiovascular Diseases, Mayo Clinic, 200 First Street SW, Rochester, MN, 55905, USA.

### Abstract

Cardiac resynchronization therapy (CRT) is a commonly used procedure to help patients with drug refractory heart failure (HF) symptoms. More patients with congenital heart disease (CHD) survive to adulthood with the improvements that have occurred as a result of surgical and medical care of these patients. However, patients with CHD may develop ventricular dysfunction and HF and thus be considered for CRT. In this review, we discuss the unique features of CRT in the adult CHD population. We examine the existing data on utilization of CRT in patients with HF and CHD and specifically discuss the limitations in terms of benefit as well as data availability. Finally, we review the specific coronary sinus anatomy and technical considerations for placing a left ventricular lead in patients with CHD.

Circ J. 2012 Oct 16. [Epub ahead of print]

[Fate of Preoperative Atrial Fibrillation After Correction of Atrial Septal Defect.](#)

Wi J, Choi JY, Shim JM, Uhm JS, Hwang HJ, Kim JY, Pak HN, Joung B, Lee M.

**Source**

Cardiology Division, Department of Internal Medicine, Yonsei University College of Medicine.

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