

Is Anybody Listening?

Jack M. Colman, MD



Just five years ago, at the adult hospital housing the Toronto ACHD program, a program founded in 1959 and located 20 yards across a side street from one of the largest Pediatric Cardiology programs in the world, the hospital administration were not really aware of what ACHD was, though they knew that the patients cost too much and stayed in the ICU too long. Today, in contrast, ACHD has been recognized as one of the Five Fundamental Pillars of our Cardiology Program, and receives committed support at all levels.

Why is this Toronto story important? Because it reminds us that we have a great product and an important mission. We can teach people about our amazing patients. We can keep faith with the promise made by pediatric cardiology and congenital surgery when these infants and children were offered a future. We can convince people who are not adult congenital physicians or surgeons, who are not pediatric cardiologists, who are not family members of children with CHD, that they too have a stake in one of the greatest triumphs of modern medicine – the defeat of congenital heart disease.

This story is not so unusual today. It is the one I know best because I saw it happen up close; I lived through the paradigm shift. But surely that paradigm is shifting where you live too. When both houses of the US Congress recognize the needs of ACHD in legislation, there is a paradigm shift. When the ACC and AHA and the ESC and the CCS and the JCS devote resources to producing guideline documents on a par with guidelines on ischemic or valvular heart disease or arrhythmia, that is a paradigm shift. When patients and families come together to create dynamic and successful support organizations, some thing special has happened. When nurses and CCA's devote their careers to the field, and are visibly excited and pleased about what they do every day, we are all stronger. When bright young people begin to direct their training toward ACHD while still in medical school, they are following a path, a path that they have been shown by us, that those who came before them could not possibly have found. When colleagues from around the world join this organization, continue to support it, and want to build it, we learn again that we are not alone, not just a few.

There are huge challenges. Most patients are still not under proper care. Many natural and unnatural histories are yet to be written. Much to learn, much to do, lack of resources, lack of training programs, lack of research funding, lack of insurance support, lack of space, lack of time.

But lack of interest? Never! We have a great story to tell. We are telling it well, and as long as we do, we will find those who will listen. And we will move from strength to strength.

Mission Statement

The purpose of the International Society for Adult Congenital Cardiac Disease is to promote, maintain and pursue excellence in the care of adults with congenital cardiac disease.

The Society is dedicated to the advancement of knowledge and training in medical disciplines pertinent to congenital heart disease in adults.

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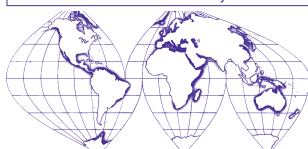
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International Society for



Adult Congenital Cardiac Disease

Good News from Asia

Heung Jae Lee, MD, PhD, President, Korean Society of Adult Congenital Heart Disease and Koichiro Niwa MD, FACC, PhD, President of Japanese Society of Adult Congenital Heart Disease – Co-Directors, Symposium & Gathering of Asia-Pacific GUCH

We are very happy to tell you good news from Asia. There is a new movement in the Asian-Pacific area among doctors in the field of adult congenital heart disease.

We are planning to have an Asia-Pacific Adult Congenital Heart gathering together with 'The 1st Asia-Pacific Adult CHD Symposium' on May 29, 2008. This will be a satellite symposium of 'The 2nd Congress of Asian-Pacific Pediatric Cardiology and Cardiac Surgery Society (PCCS 2008)', a bi-annual official meeting of the Society, which will be held in Jeju Island, Korea, May 28-30, 2008. The Society is an official body & society for pediatric cardiology and congenital heart disease in Asia and Australia-New Zealand.

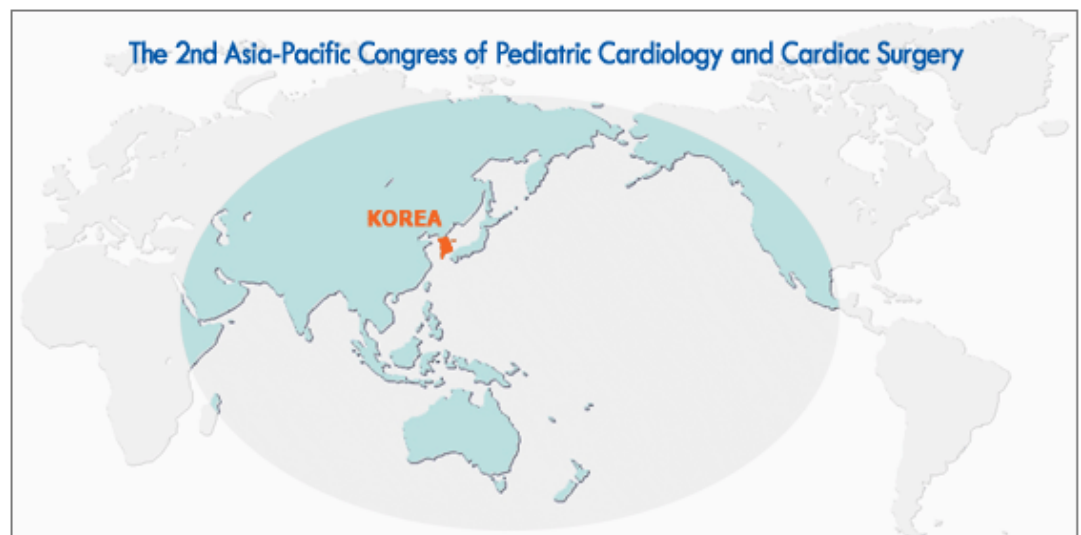
The Korean GUCH Society and the JSACHD (Japanese Society of Adult CHD), initially proposed having a joint meeting of these 2 Societies, together with ISACCD and the Euro GUCH working group of the European Society of Cardiology, at PCCS 2008. But we have now expanded our plan to host an Asia-Pacific ACHD gathering and possibly establish the first meeting of a new Asian-Pacific Adult Congenital Heart Disease Society. The presidents and executive committees/nucleus members of the ISACCD and Euro GUCH have been supportive of our new movement. Also many colleagues in Asia Pacific have already agreed to participate in this program.

For this new ACHD meeting in the Asia-Pacific region, Jack Colman MD, President of ISACCD, and Harald Kaemmerer MD. Past Chair of the Euro GUCH Working Group will join the meeting to represent their Societies. Also, representatives from India, Singapore, Thailand, Taiwan, China, Australia and New Zealand, together with Korea and Japan, have already agreed to join.

We hope this meeting provides momentum for the future development of an Asia-Pacific Society of ACHD and facilitates increasing international collaboration among ISACCD, Euro GUCH and Asian-Pacific ACHD groups.

Future updates of the program and other details, including venue, will be provided as soon as possible, when the program is finalized. All the members of ISACCD and other people who are interested in ACHD in the Asia-Pacific region are welcome to join this fascinating new meeting.

For further information please go to the website for the
Asia-Pacific PCCS at: www.pccs2008.com



Coronary Anomalies of Consequence – Anomalous Coronary Artery from the Opposite Sinus of Valsalva

Melvin D. Cheitlin, MD, Emeritus Professor of Medicine, University of California, San Francisco

Coronary artery anomalies are uncommon in the adult, found in 0.3% of all autopsies¹ and 1% of all coronary arteriograms². Anomalous origin from the wrong Sinus of Valsalva (AOWSV) comprises 0.15% of all coronary anomalies. These anomalies came to prominence when they were recognized as the cause of sudden death in young people during or immediately after vigorous exercise, often during competitive athletics. Subsequently, this anomaly has been found to be the cause of death in 12% of sports-related sudden deaths³.

The first extensive review of these anomalies was in 1974⁴ where 51 cases from the Armed Forces Institute of Pathology (AFIP) were reported. Of 36 cases of left coronary artery from the right sinus of Valsalva (LCRSV), 9 died suddenly during or after vigorous physical activity. Of 18 cases of right coronary artery from the left coronary sinus of Valsalva (RCLSV), none died suddenly. The only distinguishing feature of those dying suddenly during exercise from those not dying suddenly was their young age (mean age 20 [range 13-36] vs mean age 53 [range 24-87]). In a later autopsy study of 12 sudden deaths in patients with AOWSV, there were 4 sudden deaths in patients with RCLSV, 2 during exercise⁵. Of the 8 with LCRSV, all died during vigorous exercise. With the exception of passage between the aortic root and the pulmonary outflow tract, there were no anatomic features that significantly distinguished those who died suddenly from those who did not, and only the young age of those who died suddenly was significantly different from those who did not.

Non-invasive diagnostic modalities such as echo-Doppler, contrast enhanced AT and MRI all can identify the origin and initial course of the coronary arteries from the aorta. In the adult a routine 2-D trans-thoracic echocardiogram (TTE) infrequently identifies the origin of both coronary arteries. In the adult, even when specifically sought, it is often not possible to be sure of the coronary origins. In a study of 2,388 consecutive echocardiograms in children, with specific attention to coronary origin, 4 were found to have anomalous origin⁶.

The proposed mechanisms of arterial closure are:

1. Slit-like orifice from the acute coronary take-off with flap-like closure.
2. First segment of anomalous artery tunneling in the aortic media.
3. Compression or kinking of the anomalous coronary due to expansion of aorta and pulmonary artery during exercise.
4. Intimal damage and coronary spasm with sympathetic stimulation due to exercise.

In a registry of 27 sudden deaths in young athletes, Basso and colleagues⁷ described 25 who died during or immediately after exercise, 23 with LCRSV and 4 with RCLSV. Fifteen (55%) had had no clinical cardiovascular manifestations or tests and 12 (45%) had clinical data, 10 having premonitory symptoms (syncope in 4, chest pain in 5, palpitations in 1). An ECG in 9, maximal stress test in 6 and echocardiogram in 2 were all normal. It appears that the sudden ischemic event that causes sudden death is episodic and not predictable by exercise testing.

Certain facts are evident:

1. The commonest circumstance where sudden death occurs is with or after exercise.
2. The patient has done the same or more exercise many times in the past without difficulty.
3. Frequently the patient has had a previous episode of exercise-associated syncope or chest pain.

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If you are not receiving this newsletter via e-mail, please contact the society at info@isaccd.org or (919) 861-5578, so that we can update our records with your current e-mail address.

International Society for Adult Congenital Cardiac Disease to Meet in Orlando



Join Us!

Sunday November 4th, 2007
Buffet Breakfast from 6:30 AM
Meeting 7:00-9:00 AM
The Peabody Orlando
9001 International Drive
Orlando FL 32819
Orlando Room

Highlights of ISACCD Meeting at AHA 2007

Featured Address:



Dr Joseph Perloff, on the subject: Complex Determinants of Ventricular Function in Congenital Heart Disease. Professor Perloff, author of many textbooks and scientific papers, and one of the founders of the field of Adult Congenital Heart Disease, will provide perspective on this very important subject. He will challenge us to think broadly and deeply about how ventricular function in congenital heart disease differs from that seen in acquired heart disease. His subject has important

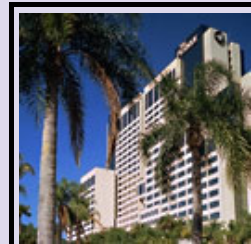
implications for the investigation and management of the increasing numbers of ACHD patients with failing ventricles. Don't miss this meeting and don't miss Dr. Perloff's talk!

Updates on ISACCD projects

- ACHD Research Round table follow-up
- World-Wide ACHD Training Program Update
- ACHA/ISACCD Clinic Directory Status Report and Future Plans
- ISACCD Multi-centre research initiatives

Election of ISACCD Executive Committee 2008-2010 - Suggested Slate Proposed By Nominating Committee will be Circulated in Fall Newsletter Shortly - watch for it!!

The ISACCD Meeting will start with a Continental Breakfast. All are Invited, whether ISACCD members or not. Please bring along colleagues and trainees interested in ACHD.



Coronary Anomalies of Consequence

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4. The patients who die suddenly are almost exclusively under age 35, most often under age 30.
5. Ischemia is not easily revocable on exercise tests.
6. Sudden ischemia resulting in death occurs when the proximal anomalous coronary artery is transiently occluded.

With a prevalence of the anomaly of 0.02-0.11% of 4 million live births/year in the US, about 2000 children are born each year with this coronary anomaly. Recognizing that there are 250,000 – 500,000 sudden deaths per year, half < 35 years of age, the number of sudden deaths due to this anomaly must be exceedingly small compared to the population with this anomaly.

An approach to the management of the patient found to have a AOWSV:

Surgical correction:

1. Patients with AOWSV with symptoms (exertional chest pain or syncope)
2. Patients at any age with stress test showing ischemia in the appropriate myocardial region.
3. ? asymptomatic patients <35 years old serendipitously found to have AOWSV without demonstrable ischemia.

Observation without surgery:

1. Asymptomatic patient > age 35 without demonstrable ischemia.

References:

1. Alexander RW, Griffith GC. Circulation 1956; 14:800.
2. Yamanaka O, Hobbs RE. Cathet Cardiovasc Diagn 1990; 21:28.
3. Burke AP, et al. Am Heart J 1991; 121: 568.
4. Cheitlin MD, et al Circulation 1974; 50:780.
5. Taylor AJ, et al. Am Heart J 1997: 133:428.
6. Davis JA, et al. J Am Coll Cardiol 2001; 37:593.
7. Basso C, et al. J AM Coll Cardiol 2000; 35:1493.



**18th International Symposium
on Adult Congenital Heart Disease**

Congenital Heart Disease 2008 & Beyond

TORONTO, June 4 – 7, 2008

Four Seasons Hotel

Course Directors:
Dr. Erwin Oechslin, Chair
Dr. Peter Liu
Jeanine Harrison, MN, APN
Marion McRae, APRN, BC

Co-Directors:
Dr. David Sahn
Dr. Gary Webb


University of Toronto


Congenital Cardiac
Centre for Adults


Peter Munk
Cardiac Centre
University Health Network


SickKids
The LaBrett Family
Heart Centre

www.uhn.ca/ACHDConference2008.asp

Nominating Committee Announces “Slate” for ISACCD Executive Committee 2008-2010

The ISACCD Nominating Committee, Drs. Michael Gatzoulis, David Sahn and Gary Webb, recently announced a suggested slate of officers for the new ISACCD executive committee, for a two year term. The official election occurs at the Winter 2008 meeting of ISACCD, scheduled for March 29, 2008 in Chicago (coincident with ACC 2008). Additional nominations can be sent to Dr. Gary Webb, and should include the names of two seconders and agreement of the candidate to stand. If there are additional nominations, a further mailing will go out to members well before the March 2008 meeting.

Here is the proposed slate. Brief biographical sketches of the nominees follow:

President: Dr Michael Landzberg, Boston, MA, USA
 President-elect: Dr. Barbara JM Mulder, Amsterdam, the Netherlands
 Secretary: Dr. Koichiro Niwa, Chiba, Japan
 Treasurer: Dr. William R. Davidson, Hershey, PA, USA
 Past-president: Dr. Jack M. Colman, Toronto, ON, Canada

Mike Landzberg MD, FACC

Director, Boston Adult Congenital Heart (BACH) and Pulmonary Hypertension Group, Children’s Hospital, Brigham and Women’s Hospital, and Beth Israel Deaconess Medical Center; President-Elect, ISACCD; ACHA Medical Board Member; ACC-AHA ACHD Guidelines Committee; AARCC Research Alliance guide; NHLBI ACHD Research Working Group; Assistant Professor, Harvard Medical School; jogger, basketball wannabe, poor karaoke singer, groom to Abby (endocrinologist), father of 3 (Nicole, Judah, Ethan).

Plan for ISACCD: As a major fan of the growth and progress within ISACCD over this past decade and since its inception, I hope to further and strengthen the objectives and mission statement that ISACCD has pursued under Jack Colman’s tenure: to increase advocacy, education and international cooperation for all involved in ACHD care-giving, with emphasis on increasing and extending constituency participation, web-based materials, round-tables and working groups, all at the international level, with application for all.

Barbara JM Mulder, MD, PhD, FESC

Professor of Cardiology, Academic Medical Center Amsterdam, the Netherlands

Director of the Department of Adult Congenital Heart Disease, AMC Amsterdam

Consultant Cardiologist Adult Congenital Heart Disease, University Medical Center Utrecht, NL

1998-2006	Founder and Chairman of the Working Group on Adult Congenital Heart Disease in the Netherlands (Netherlands Society of Cardiology)
2000-2005	Member of the Committee for Cardiovascular Databases, Registries and Surveys of the ESC
2000-2005	Chairman European Registry on Adult Congenital Heart Disease. The Euro Heart Survey. (ESC)
1994-present	Member of the Working Group on Grown-up Congenital Heart Disease (European Society of Cardiology, ESC)
2000-present	Project leader “National Registry and DNA-bank on Adult Congenital Heart Disease”, CONCOR, (Interuniversity Cardiology Institute of the Netherlands)
2001-present	Spokesperson on Adult Congenital Heart Disease for the ESC
2004-present	Member of the Public Relations Committee of the ESC
2006-present	Project leader of WP1 of the European project “Heart Failure and cardiac Repair”: Genes for Heart repair and plasticity.
2007-present	National Ambassador for the Netherlands Heart Foundation

Editor of books:

- 1) Aangeboren hartafwijkingen bij volwassenen. (Congenital Heart Disease in Adults) Bohn Stafleu van Loghum, Houten, 1999. (2nd edition 2006)
- 2) Adult Congenital Heart Disease in the Netherlands. Guidelines 2000. Working Group - Adult Congenital Heart Disease of the Netherlands Society of Cardiology. The Netherlands Heart Foundation, The Hague, 2000.

Plan for ISACCD:

Focus on international collaboration, i.e. to:

- broaden the basis of ISACCD and expand more deeply in Europe and Asia
- stimulate exchange of students and fellows between North America, Europe and Asia
- apply for joint sessions in ACC/AHA/ESC/APSC during annual congresses

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- increase the number of newsletters
- organize 3 meetings a year at: ACC, ESC and one dedicated congress
- make ACHD cardiologists feel uncomfortable without an ISACCD membership

Koichiro Niwa MD, PhD, FACC

Director of the Department of Adult Congenital Heart Disease and Pediatrics, Chiba

Cardiovascular Center, Chiba Japan

President of the Japanese Society of Adult CHD

Co-director 1st Asian Pacific Adult CHD Meeting

Treasurer, ISACCD, 2006-2008

Member, European Society of Cardiology, Working group on Grown-Up CHD

Author of Japanese text-books:

1. Niwa K, Nakazawa M, eds. Adult Congenital Heart Disease. Medical View Co, Tokyo, 2005
2. Niwa K, ed. Guidebook of Pregnancy and Delivery in Women with CHD for Patients. Chuouhouki Co, Tokyo, 2006

Plan for ISACCD: I joined ISACCD from an Asian country, and I would strongly like to step forward the international collaboration in the field of ACHD. Also, we have to educate the next generation and I am sure they will grow rapidly through our support.

William R. Davidson, Jr., MD, FACC

Graduated from the University of Pennsylvania School of Medicine in 1979 and completed his Fellowship in Cardiovascular Disease at the University of Rochester. A significant portion of his fellowship was spent involved in pediatric cardiology under the direction of Dr. James Manning. In 1985 he joined the cardiology faculty of the Milton S. Hershey Medical Center, the university hospital of the Pennsylvania State University College of Medicine, to direct the echocardiography laboratory which has grown to 12,000 cases annually. He still directs the Lab and has risen to Full Professor. In 1991, at the encouragement of Dr. John Waldhausen, Dr. Davidson founded the Program for Adults with Congenital Heart Disease (PACHD). Originally, one clinic half day was held per month and the program has since grown to 1,000 visits annually with seven half day clinic sessions weekly, and expanding faculty.

Dr. Davidson has been active in ISACCD for many years and has served as Project Committee chair. He has contributed to ISACCD projects including the CCTGA natural history study, the Fontan study and the PVR in Tetralogy of Fallot study. With Dr. George Warren and the ACHA, he worked to develop the ACHD Clinic Directory and presented its original findings at the AHA last year. He has served on the ACHA's Medical Advisory Board and has just become one of the original members of the ACHA MAB's Executive Committee. He participated in the 32nd Bethesda Conference on the Care of the Adult with Congenital Heart Disease.

Along with multicenter ACHD projects, investigative interests have coupled echocardiography and ACHD and include diagnosis of LV-RA communications, stress echo in the assessment of the right ventricle in tetralogy of Fallot and Doppler assessment of pulmonic insufficiency. Current projects include diagnosis of right aortic arch and long term consequences of subclavian flap aortoplasty for coarctation of the aorta.

Future plans include ongoing participation in the ACHA Clinic Directory Project, encouraging growth of the ISACCD membership, gaining cardiac MRI experience, expansion of ACHD basic and clinical research at Hershey and development of an ACHD fellowship at Hershey.

Jack M. Colman MD, FRCPC, FACC

Associate Professor of Medicine (Cardiology), University of Toronto. Staff Cardiologist, Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, Toronto. Staff Cardiologist and Co-Director, Heart Disease in Pregnancy Program, Mount Sinai Hospital, Toronto. President, ISACCD, 2006-2008.

I have been a member of ISACCD since the organization was founded and have been honoured to serve on the executive for the past 4 years. During this time we have begun to broaden horizons and define more precisely the role we in ISACCD can take in the world-wide development of ACHD, while we at the same time, wearing other hats, define the best roles for the other organizations in which we participate that are also dedicated to this important and fascinating field. I look forward to continuing to witness, contribute to, and passionately advocate for the success of this now not-so-new but still emerging calling.



6th Advanced
Symposium
on Congenital
Heart Disease
in the Adult

Host Institution

Royal Brompton Hospital
and the National
Heart & Lung Institute
London, UK

Programme Directors

Prof Michael A. Gatzoulis
Mr Darryl F. Shore

26 & 27
September 2008

Thessaloniki - Greece

Makedonia Palace Hotel

F I R S T A N N O U N C E M E N T

For more information, visit: www.isaccd.org/news/FIRSTANNOUNCEMENT-ForSept08.pdf