The official newsletter of the
International Society for Adult Congenital Cardiac Disease

ISACCD update

President's Message

by Michael A. Gatzoulis

It is with honour and a strong sense of responsibility that I introduce this report as the first President of the International Society of Adult Congenital Cardiac Disease (ISACCD) based outside North America. This speaks for the Society's strength and its international/global perspective and commitment.

There has been tremendous progress since ISACCD's conception in 1992. The increased workload of caring for adults with congenital heart disease (CHD) and the need for representation through a professional body was recognized early by its pioneer officers. Their first meeting was convened during the Annual Scientific Sessions of the American College of Cardiology in Dallas, in April 1992. The foresight of the steering committee, including Drs Joseph Perloff, Gary Webb, Dan Murphy, Tim Garson, Richard Liberthson, David Skorton, Carole Warnes and others, led to a new professional body, the ISACCD. The purpose of the society was summarized in its *"mission statement 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."*

ISACCD has grown and flourished since, with a current full membership of 236 members and a truly worldwide representation and has achieved many of its goals over the past 12 years. ISACCD has promoted advocacy and education through its own meetings during the Scientific Sessions of the American College of Cardiology and the American Heart Association, more recently extended to the Annual European Society of Cardiology Conference and other International Symposia in Adult CHD around the world. ISACCD has facilitated the publication of educational material including its own monograph and the building of its own active website http://www.isaccd.org with a wealth of information for professionals and patients alike, including training opportunities and the availability of its members around the world. ISACCD has pioneered collaborative research in the field and supported an international database, thus enhancing our understanding of CHD and enabling smaller unit participation. ISACCD officers and members were instrumental in the two Bethesda Conferences addressing the needs of the Adult with CHD and actively participated in national and international advisory bodies that influence health policy around the world. Over the past 12 months, ISACCD has established a strong link with the International Journal of Cardiology, which has since become its affiliated Journal. This affiliation has secured more publishing space for Adult CHD work, including free page allocation for the Societies news and activities and has facilitated greater exposure of our work to a more general cardiology audience. Do take advantage of this partnership and consider your academic work for the Journal. Also let us know of your local successes and challenges, write to us with your thoughts on ways of improving patient care and tell us of initiatives that you would like to see being implemented by ISACCD. We make a pledge to make your contribution public through our website, the ISACCD Newsletter and/or the Journal.

One of the common misconceptions which does not serve the patients or our objectives (anymore) is that Adult CHD is a small and thus an exclusive area within cardiovascular medicine. While this may have been the case in the past -and there are obvious ongoing challenges in managing these patients- clearly our field is not a small field any longer. With a worldwide CHD incidence of about 1% and 85% survival rates to adulthood it should not come to a surprise that there are approximately

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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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International Society for Adult Congenital Cardiac Disease

Current Status of 3-Dimensional Echocardiography Use in Adult Congenital Heart Disease

from the ACC Scientific Session in March 2004

"David Sahn, who is Director of the Clinical Care Center for Congenital Heart Disease and Director of the OHSU Interdisciplinary Program for cardiac imaging, has gradually made a larger investment of his time in implementing MRI studies in adults with Congenital Heart Disease.

Dr. Sahn reviewed some of the advantages of real-time 3D echocardiography for quantification of flow and muscle mechanics in 3D space and for RV and LV function. These methods are robust and can provide automatic boundary tracking and can provide tissue Doppler and strain rate functionalization in 3D space. They are excellent not only ventricular function, but for fine details of atrial-ventricular and semi-lunar valve anatomy where anatomy and flow can be approximated and understood in 3D space. There are opportunities for real-time, closed-chest guidance of cardiac surgery.

MRI however, is not window-limited and can provide robust characterization of cardiac function, both by velocity encoded wall motion studies as well as tissue tagging for quantitation of aortic and pulmonary flows and quantitation of shunts can be obtained by velocity encoded MRI and of course it is known to be exquisite for extra-cardiac anatomy, both pulmonary arteries and the aortic arch using gadolinium enhanced 3D angiography.

Recent studies of aortic flow propagation and mechanical characterization represent elegant opportunities for learning about the natural history in diseases, for instance, like Marfan's Syndrome.

Myocardial perfusion can be reliably done by MRI and late gadolinium take-up appears to represent a way of identifying scarred and/or non-viable myocardium. MRI and 3D echo are complimentary and should be used alternately over the course of follow-up of adults with Congenital Heart Disease. Rather than automatically using one or the other, a rational question should be asked as to what information is needed for this particular visit.

Finally, Dr. Sahn emphasized that MRI is best implemented either in a collegial multi-disciplinary environment or it must be done by the Congenital Heart Disease cardiologist themselves, since they know the anatomy and the clinical questions to be addressed.

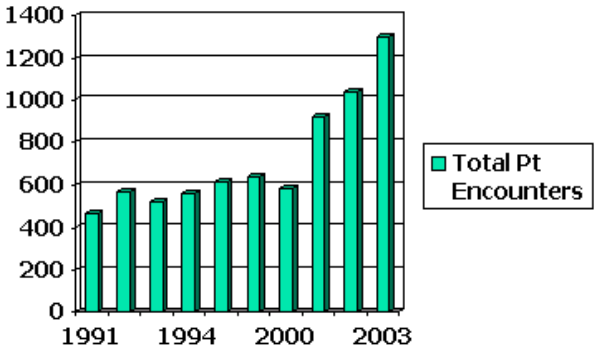
Upcoming Meetings

The Fifteenth Annual International Symposium on Congenital Heart Disease in the Adult will be held at the Four Seasons Hotel in Toronto May 26-28, 2005, directed by Drs. Peter Liu, Gary Webb, and David Sahn. An outstanding international faculty will be present. The meeting is sponsored by the American College of Cardiology. Course details and registration can be accessed at www.acc.org (click on "Programs") after November 1, 2004.

For those interested in further expanding their learning in Toronto in 2005, watch for details about an **International Pediatric Cardiology meeting** directed by Dr. Andrew Redington and others, which will take place in Toronto commencing May 29, 2005 the day after the ACHD Symposium ends.

The Ahmanson/UCLA ACHD Centre experience

from the ACC Scientific Session in March 2004



At the Semi-Annual ISACCD Meeting on Sunday 07 March 2004 (held jointly with the ACC) in New Orleans, Dr. John S. Child, the Director of the Ahmanson-UCLA Adult Congenital Heart Disease Center (ACHDC) shared the center's experience. The ACHDC, established formally in 1981 with Dr. Joseph K. Perloff (now emeritus professor) has a multimission profile- foremost being refined patient care and advanced training of fellows, residents, and medical students, buttressed by clinical and basic research to advance scientific knowledge in the field. Dr. Child noted that, as of end 2003, the registry included 2,097 patients (male 928, female 1169), of which 317 were currently cyanotic, 1142 operated, and 172 deceased. This center combines the collaborative resources of the adult and pediatric cardiology programs with their colleagues in cardiothoracic surgery, radiology, pathology, obstetrics and gynecology, as well as a host of other disciplines.

The rate of growth of the center was presented using the period from 1991-2003(see chart), which highlighted the need for more such centers in North America and more graduate trainees to enter the field. An advanced ACHDC fellowship program has provided training since 1991, with 11 such fellows to date, with several from other countries. Research directions include systemic disorders in cyanosis, pulmonary hypertension, myocardial function in systemic ventricles, electrophysiology of congenital heart disorders, great arterial wall characteristics and vascular biology, developmental biology, and outcomes analysis.

Don't Miss the ISACCD Semi-annual Meeting on Sunday, November 7, 2004 in New Orleans!

AHA Scientific Sessions in New Orleans
Wyndham New Orleans Hotel at Canal Place
Sunday, November 7, 2004 • 7:00-8:30 am in the River Room

Agenda

a. Welcome - Dr. Gatzoulis
b. Introduction of Speakers - Dr. Graham

   Lecture A: " What is new in Pulmonary Arterial Hypertension?", Dr. Michael Landzberg, 40 minutes-includes discussion

   Lecture B: "ACHD Centers Experience: Zurich and Swiss WATCH (Working Group on Adults and Teenagers with Congenital Heart Disease)", Dr. Erwin Oechslin, 10 minutes-includes discussion
c. *International Journal of Cardiology* Associate Editor Report, Dr. Gatzoulis
d. Treasurers report of membership and finances, Dr. Oechslin
e. Website report - New Website officer Dr. Frank Cetta
f. Newsletter report - Dr. Child
g. ACC meeting planning for March 05 - Dr. Murphy
h. Joint American Academy of Pediatrics/ISACCD Plenary session - Friday, October 7, Washington DC - Dr. Gatzoulis
i. Project Committee report - Dr. Bill Davidson
j. Joint Council for Congenital Heart Disease; Update from the September Chicago Meeting- Dr. Jack Colman
k. Educational/Training grant. Website development on education? - Dr. Liberthson
l. Report from European GUCH Working Group - Dr. Oechslin
m. Membership committee Report - Drs. Colman/Gatzoulis
   - Electronic membership registration
   - ISACCD/IJC cards
n. ACHD Centers/Access to ACHD Patient Associations - Dr. Graham and Amy Verstappen
o. Next ISACCD meeting: Orlando(ACC) - suggestions for the program
p. Other business

New Directions for the Adult Congenital Heart Association

by Amy Verstappen, ACHA President

2004 brought a number of exciting firsts for the Adult Congenital Heart Association (ACHA). In February, our national board had its first strategic retreat and rewrote our mission statement as follows:

The Adult Congenital Heart Association (ACHA) is a nonprofit organization which seeks to improve the quality of life and extend the lives of adults with congenital heart defects. Through education, outreach, advocacy, and promotion of research, ACHA serves and supports the more than one million adults with congenital heart defects, their families, and the medical community.

With this mission as the centerpiece, we went on to articulate ACHA's core values, which include confidentiality, professionalism, compassion, integrity, inclusion, activism, and commitment to excellence. Our goal is to be a forward-looking, vocal organization that effectively generates change on behalf of the adult congenital heart community.

In the intervening months, we have hired an office management service, increased our fundraising activity, and are exploring new approaches to capacity building. Through media contact and outreach, ACHA's credibility continues to rise, and we are increasingly invited to represent the patient perspective at national and international venues, including the first working group on adults with congenital heart defects convened by the National Heart, Lung, and Blood Institute.

Our focus continues to be on the huge barriers to care that currently exist for the adult congenital heart patient, particularly those with complex disease. First and foremost is the common misperception of being "fixed", both by the patient him or herself, and by the medical community. Additional barriers include lack of knowledge about one's defect, lack of knowledge about cardiac symptoms and risks, intermittent or absent cardiac care, lack of specialized care, and insurance barriers. Unfortunately, many in our community go without appropriate care until they arrive in the emergency room with serious cardiac issues. This reflects the existing data, which suggests that as few as 10% of all adults living with complex heart defects currently receive appropriate follow up.

To address these barriers, ACHA seeks to give every individual living with a heart defect access to the ACHA "Toolkit". This consists of:

- Knowledge of one's defect(s)
- Ability to recognize cardiac symptoms
- Understanding of need for on-going care
- Understanding of risks particular to defect(s)
- Access to medical records
- Awareness of "life style" issues
- Information on finding appropriate care
- Access to support and on-going information

We are currently planning a number of new initiatives to address these needs, including new publications, dissemination of a CHD passport, and a more extensive on-line clinic directory.

Throughout these changes, we have continued quarterly publication of our newsletter, which has now changed its name to "Heart Matters", and our website continues to receive thousands of hits a month from individuals seeking support and information. In April, more than two hundred individuals with heart defects, their families, and medical professionals gathered in Rochester, MN for our largest-ever national conference. In 2005, we are planning three regional events, in Orlando, New York, and Los Angeles. ACHA also continues to work with ISACCD to identify areas in which we can more fully collaborate and support each other's efforts. For more information about ACHA and our activities, or to join as a professional member, please visit our website at www.achaheart.org. Working together, we can preserve the health and save the lives of our unique, pioneering, and highly endangered community – adults living with congenital heart disease.

President's Message

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1,000,000 adults with CHD in the USA, 200,000 in the UK with similar numbers of patients in most countries around the world. Sadly, the vast majority of these patients have been lost to follow-up. The need for bringing them back to appropriate care cannot be overemphasized. Periodic outpatient assessment combined with timely intervention –often necessary- are essential for safeguarding long-term outcomes for CHD and for improving survival and importantly quality of life. We are very fortunate today with more knowledge and are well equipped with advanced technology and skilful interventions and this must lead to better outcomes for our patients. We need to practice proactive and not reactive medicine, however, and we need to engage our patients in this process. The data is compelling that making decisions based on overt symptoms compromises our patient's outcome and their ability for complete heart remodelling after late reoperations (which themselves carry a higher risk, if performed late). Nobody doubts that there are difficult decisions to be made, but a multi-disciplinary team approach combined with a life-long partnership with the patient and a zest for ongoing critical review and reporting of our practices are bound to assist this process. I trust that we all agree on the principle that while most of our patients have had life saving operations and continue to enjoy good quality of life their hearts were not fixed. We need to get this message across to patients and their families and to a broader professional base so that we can begin seeing that the full potential of every single patient born with CHD is ultimately realized.

On behalf of all of us I would like to take the opportunity here to thank all previous officers of the Society, the Past President Dr Thomas Graham for the other members of the recent executive Drs Elyse Foster, Michael Landzberg and Daniel Murphy Jr for their time and many contributions towards advancing the causes of ISACCD and for their personal encouragement and support from my first days in Adult CHD.

There is clearly much to do. The following challenges are ahead of us all and I look forward to working together to:

- Continue to expand our membership; encourage our colleagues from our own establishment to join and make colleagues from other institutions aware of ISACCD and our causes
- Establish a closer interaction and partnership with patient support groups; together, we have a much stronger voice and lobbying power and we are more likely to be successful in our common goals
- Increase awareness of Adult CHD through campaigns to health care professionals, patients themselves and to the public; utilize general and specialized conferences, the scientific and general press, media and the web to advertise our work and the tremendous need that exists
- Secure much needed resources for supporting clinical needs, training and education and for facilitating research; remain persistent in engaging funding bodies, philanthropy, government and industry alike to achieve these goals
- Last but not least, accept the fact that Adult CHD is a wonderful but also grossly under funded (and in that sense neglected) field of modern cardiovascular medicine, hence the need for all of us to take a long-term view for meeting the challenge and the privilege of caring for the adult patient with CHD.

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.