

ANNUAL SCIENTIFIC REPORT 2005 - 2006
ESC WORKING GROUP ON GROWN-UP CONGENITAL
HEART DISEASE (WG #22)

1. OBJECTIVES OF THE WG ON GROWN-UP CONGENITAL HEART DISEASE:

- To promote, to maintain and to pursue excellence in the care of adults with congenital heart disease
- To improve the quality of care by endorsing, supporting and organizing educational programs in adult congenital heart disease
- To establish close links with other groups/societies dealing with adult congenital heart disease patients
- To promote research in the field of adult congenital heart disease
- To establish guidelines for the treatment and management of adults with congenital heart disease.

The ambition of the ESC Working Group on Grown-Up Congenital Heart Disease (WG 22) is to serve as the leading organization in the field of adult congenital heart disease in Europe and to attract all physicians to foster the development of this area of expertise.

2. STRUCTURES AND ORGANISATION

2.1. During the year 2005/2006, the **Nucleus** of the WG has been composed as follows:

- **Chairman:** Harald Kaemmerer, Munich (DE)
- **Vice-Chairman:** Helmut Baumgartner, Vienna (AT)
- **Past-Chairman:** Erwin Oechslin, Toronto (Can)
- **Secretary:** Jana Popelova, Praha (CZ)
- **Treasurer:** Folkert Meijboom, Rotterdam, (NL)
- **Nucleus Members:** Luciano Daliento, Padova (IT)
John E. Deanfield, London (GB)
Rafael Hirsch, Petah Tikva (IL)
Ulf Thielén, Lund (SE)
Fernando Picchio, Bologna (IT) – AEPC representative
(appointed by the AEPC)
- **Honorary Member:** Jane Somerville, London (GB)

2.2. Membership

The total number of WG 22 membership has increased to more than 206 active members at the date of August 2006.

3. ACTIVITIES OF THE WORKING GROUP 2005 – 2006

During the year 2005-2006, a main focus of our activities was again to further strengthen the existing ties among the different WGs / Associations and Societies and to create new ties with people who are interested in the care of adult congenital heart disease.

Throughout the year WG 22 was involved in several collaborative meetings and projects.

Special thanks to Prof. Öztekin OTO, Izmir (TR), our long-acting cluster-coordinator who presented our proposals to the Congress Programme Committee for many years.

During the ESC spring summit it was decided to propose to change the clustering in order to make a more logical clustering, with working groups that have more in common with each other than it has been the case until now. During the ESC council meeting in Barcelona this proposal will be discussed and possibly decided.

Also, we appreciate the excellent support given by the president of the AEPC Prof. András SZATMARI, Budapest (HU), and all Congress Programme Committee members who are aware of the increasing importance of our field.

- Of outstanding importance is, that Prof. Erwin Oechslin, MD, and past-chairman of our WG left Zürich (CH) in order to follow Gary Webb as the new director of the Adult congenital heart disease center in Toronto, a worldwide leading institution. Congratulations !

3.1. INTERNATIONAL CONGRESSES and MEETINGS

3.1.1. Annual ESC-Congress in Stockholm 2005

The congress of the ESC in Stockholm confirmed the increasing importance and popularity of the Adult congenital heart disease or GUCH (grown-up congenital heart disease) sessions organized by the WG 22 among the cardiologists.

There were 6 pre-arranged Clinical Seminars or Symposia organized by WG 22 (or organized in a co-operation with other WGs), three abstract sessions and one poster session concerning the topic of adult congenital heart diseases.

The Symposia and Clinical Seminars:

- Of mice and men (279-282)
- Cardiovascular magnetic resonance of right ventricle in congenital heart disease (283-286)

- Diagnostic and therapeutic dilemmas in adult congenital heart disease (938-941)
- Antithrombotic strategies in adults with congenital heart disease (942-945)
- Do we treat Marfan patients properly? (991-994)
- New steps forward in interventional techniques in congenital heart disease (1495-1498)

Most sessions were very well attended and evoked often long and lively discussions. They showed many burning questions and subjects to be solved on the basis of international collaborative multicenter studies. The discussions showed the importance of exchanging experience in the diagnostics and treatment of this very special population of adult cardiac patients.

Even in the other sessions of other WGs the subject of adult congenital heart disease played an important role, as in the pulmonary hypertension, atrial fibrillation and arrhythmia, cardiac anatomy and morphology and pediatric cardiology.

1.1.2. Joint Meetings With Other Organizations And Societies

Several members of WG 22 were heavily represented as speakers and chairmen in quite a lot of international and national meetings, e.g.:

- **EUROECHO 9**, 7-10 December 2005, Florence (IT)
- **4th World Congress of Pediatric Cardiology & Cardiac Surgery** 18-22 September 2005, Buenos Aires, Argentina
- **41st Annual Meeting of the Association for European Paediatric Cardiology (AEPC)** 24-27 May 2006, Basel (CH)
- **6th Annual Congenital Heart Disease in the Adult: A Combined International Symposium**, 25-28 June 2006, Skamania Lodge, Stevenson, Washington (US).

3.2. UPCOMING MEETINGS

- **World-Congress in Barcelona, 2 – 6 September 2006**
- **Echocardiography in Pediatric and adult Congenital Heart Disease**, Leuven, 20 – 24 September 2006
- **5th Advanced Symposium on Congenital Heart Disease in the Adult**, London (UK), 25-27 September 2006
- **The second international congress “Arrhythmias, from neonate to adult”**, University Medical Center Groningen, Netherlands, 12-13 October 2006
- **Cardiac Disease and Pregnancy**, London, 20 November 2006
- **EuroEcho 10**, Prague, CZ, 6-9 December, 2006
- **Basic Course on Paediatric Cardiac Morphology**, Leiden University Medical Center, 25-26 January 2007

- **D-A-CH-Symposium on GUCH**, Vienna, 4 - 5 May 2007
- **17th Annual Congenital Heart Disease in the Adult: A Combined International Symposium**, Philadelphia (US) 20-23 June 2007.

4. **ACTIVITIES RUN OUTSIDE THE ESC ANNUAL CONGRESS**

4.1. **ESC Pocket Guidelines: Management of Grown Up Congenital Heart Disease**

The first European Task Force Document on the Management of Grown-Up Congenital Heart Disease published in 2003 (*Eur Heart J* 2003; 24:1035-1084) is a key manuscript for high quality care of adults with congenital heart disease, not only in Europe!

An editing committee (A. Hoffmann, Basel (CH); J. Deanfield, London (GB); H. Kaemmerer, Munich (DE); E. Oechslin, Zurich (CH)) composed the ESC pocket guidelines, which appeared during the Stockholm meeting, 2005.

4.2. **Collaboration with ESC-WGs, AEPC and ISACCD**

The fruitful cooperation of WG 22 with ESC-WG's and other international societies has been strengthened in order to transfer knowledge and expertise !

Association for European Pediatric Cardiology (A.E.P.C.): The very successful collaboration with AEPC has been continued. Prof. Fernando Picchio, Bologna (IT) is still the AEPC representative within our Nucleus. Prof. J. Deanfield is responsible for relations to AEPC. There were several very fruitful and stimulating discussions with the AEPC-President Prof. A. Szatmary (HU).

Harald Kaemmerer (DE) and Helmut Baumgartner (AT) have been invited to act as councilors in the GUCH-task force of the A.E.P.C..

Next year, the annual AEPC meeting, to be held in Warsaw in May 2007, will be preceded by a one day teaching course adult congenital heart disease.

International Society for Adult Congenital Cardiac Disease (ISACCD): The tight cooperation between ISACCD and ESC-GUCH representatives persists. Michael A. Gatzoulis (WG 22 Member), London (GB) is the first European ISACCD president in charge (2004-2006) and Erwin Oechslin (Past-Chairman WG 22), Toronto (CAN) is ISACCD treasurer (2004-2006).

Canadian Congenital Heart Network (CACH Network): Also this very close and successful cooperation continues!

4.1. **European Heart Survey on Adult Congenital Heart Disease:**

Members of WG 22 formed the Scientific Committee of the **Adult Congenital Heart Disease** Survey under the chairmanship of Barbara Mulder (NL). The Euro Heart Survey on CHD was finished in April 2004.

During the past year several articles were prepared and published on data from the Euro Heart Survey. Presently, four papers have been published, one is in press, six papers are still in preparation. Altogether, this means that the EHS on congenital heart disease has been a great success. We want to congratulate all who participated with this success, but especially Barbara Mulder. We also thank her for all her time and effort she put into this survey and for the inspiring way she chaired this.

Papers that have been published or are in preparation are listed below.

1. The spectrum of adult congenital heart disease in Europe Eur Heart J 2005 (Engelfriet et al)
2. Adherence to guidelines Eur Heart J 2006 (Engelfriet et al)
3. The organisation of care Eur Heart J 2006 (Moons et al)
4. The distal aorta in Marfan syndrome Heart 2006 (Engelfriet et al)
5. Radiation exposure in ACHD patients Int J Cardiol (In Press) (Hoffmann et al)
6. PAH in septal defects Submitted (Engelfriet/Möller et al.)
7. Epidemiology of ASD in adults Submitted (Engelfriet/Meijboom et al)
8. Fontan patients in the EHS In preparation (Thilén et al)
9. Risks of smoking in CHD In preparation (Drenthen et al)
10. LV dysfunction in TOF patients In preparation (Bouzas/Gatzoulis et al)
11. Pregnancy and heart function in CHD In preparation (Budts et al).

Continued follow-up: For a continued follow-up of the EHS cohort, an addition to the electronic CRF has been posted on the web, collecting only few (!) data (Deceased, Date of death, Cause of death, Lost to follow-up, Date of last contact. Unfortunately the response rate so far is only $\pm 25\%$.

This means that it's doubtful whether a continued follow-up of the cohort is possible. This means that it's doubtful whether a continued follow-up of the cohort is possible.

5. **UPCOMING PROJECTS**

Several study groups dealing with specific problems regarding adults with congenital heart disease have been established. Their task is:

- To establish specific registries in patients with congenital heart disease
- To manage the database
- To analyze the data and to report the results

- To report about the activities and the progress of the registry at the annual WG business meetings.

Registry on Arrhythmias in Adult Congenital Heart Disease

A task force has been established to create a registry on Arrhythmias in adults with congenital heart disease to collect data regarding arrhythmias, management strategies and outcome in adults with congenital heart disease.

Members of this task force: Urs Bauersfeld, Zurich (CH) (Project leader); Joachim Hebe, Bremen (DE); Günter Stix, Vienna (AT), Bernhard Zrenner, Munich (DE).

Registry on Pregnancy in Adult Congenital Heart Disease

A task force has been established to create a registry on pregnancy in patients with congenital heart disease to collect data regarding course, management strategies and outcome in adults with congenital heart disease.

Members of this Study Group: Jörg Stein, Innsbruck (AT) and J. Roos-Hesselink, Rotterdam (NL) (project leaders). They decided to have develop the registry as a joint project together with Roger Hall.

Register of Adult Congenital Heart Disease-Centers in Europe

A task force has been established to collect data regarding the localization and infrastructure of Adult Congenital Heart Disease-Centers in Europe. For patients who travel through Europe or on vacation it is helpful to know in which countries and cities they can find specialized GUCH-units and who they can be contacted.

Members of this Study Group: Harald Kaemmerer, Munich (DE) (project leader); Helmut Baumgartner, Vienna (AT); Erwin Oechslin, Zurich (CH).

A first inventarization has been made and has been sent by e-mails to the members of the working group in 2005. An update is scheduled for 2007.

Radiation doses during Follow-Up of Adults with Congenital Heart Disease

Andreas Hoffmann, Basel (CH) and the“ European Heart Survey on Grown-Up Congenital Heart disease (GUCH)”-Group performed this study.

Study Group on ‘Pulmonary hypertension in adult congenital heart disease’

Our working group has established a study group on ‘Pulmonary hypertension in congenital heart disease. Members of this Study Group: Ingram Schulze-Neick, Berlin (DE) and Werner Buds, Leuven (BE) (project leaders), Maurice Beghetti, Geneve (CH), Rolf Berger, Groningen (NL), N. Galie, Bologna (IT), Michael Gatzoulis, London (UK), Harald Kaemmerer, Munich (DE), Marc Humbert (F).

Almost all had participated in the Breathe 5 study, the study on effect of Bosentan in patients with Eisenmenger syndrome, published in Circulation in July 2006. The authors should be congratulated with this. For most of the members of our WG22, who see many patients with Eisenmenger syndrome in daily clinical practice, it means that evidence-based medical therapy can be offered to these patients, thanks to international multicenter study. A good example of how multicenter cooperation leads to enough patients and data within an acceptable to allow solid statistical analysis, supplying evidence as the basis for medical treatment. In many, if not in almost all other fields in our part of Cardiology we still lack evidence. Therefore, this example is one to follow.

Retrospective multicenter study on cardiac resynchronization therapy in children (-19 years) and patients with congenital heart disease (any age).

This Retrospective multicenter study on cardiac resynchronization therapy in children (-19 years) and patients with congenital heart disease (any age) is performed by Jan Janousek, Leipzig (DE).

Accreditation in Congenital Heart Disease Echocardiography

On December 7, 2006, there will be the first examination in congenital echocardiography during EUROECHO 10 in Prague. This examination is jointly organized by the European Association in Echocardiography (EAE), Association for European Paediatric Cardiology (AEPC) and the WG on Grown-Up Congenital Heart Disease (WG GUCH) of the European Society of Echocardiography. For our Working group Helmut Baumgartner (AU) and Folkert Meijboom (NL) were involved. This joint effort stresses the importance of special skills and training in the examination of congenital heart disease patients and the need for continued education and practical involvement to maintain the quality of care. One of the goals of accreditation are to protect children and adults with congenital heart disease from undergoing echocardiographic examinations performed by unqualified sonographers and to set a European standard for competence and excellence in congenital echocardiography. Further information: www.esccardio.org/bodies/associations/EAE/accreditation/CHD/?hit=home

"Incidence of late cerebrovascular complications in Fallot-patients who underwent a Blalock shunt".

This Collaborative study is initiated by A. Hoffmann, Basel (CH).

6. ACKNOWLEDGEMENT

We appreciate all activities of our WG members to improve the care for adults with congenital heart disease and we are indebted to the Nucleus Members for their stimulating discussions and excellent support !

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