### September 2011

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**ISACHD Newsletter**

**In This Issue**

**Regional News**

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**President’s Message**

by Barbara J.M. Mulder

Dear ISACHD members,

ISACHD has become an affiliate member of WHF, the World Heart Federation. During the ESC congress in Paris, with more than 30,000 attendees the largest cardiology congress worldwide, Jack Colman accompanied your president to the WHF meeting. Sidney C. Smith, President of WHF, chaired the meeting with around 40 attendees from various organizations. Currently unknown faces, but hopefully there are some future partners among them. The main focus of the meeting was the upcoming World Congress of Cardiology, the official congress of the WHF, which is held every two years. The next World Congress of Cardiology Scientific Session will take place in 2012 from 18-21 April in Dubai, United Arab Emirates. The preliminary program contains quite some sessions on congenital heart disease, which can be found on the WHF website. At the meeting, no other WHF activities were discussed where ISACHD could join at present. We need to stay alert and wait for appropriate opportunities.

From 12-16 November 2011 the next congress of the American Heart Association will take place in Orange County Convention Center Orlando. You will be informed about the exact time and location of the ISACHD meeting. Latest developments of the newly installed ISACHD working groups on Global Health Care, Education and Research will be discussed. You can still join one of these working groups by sending an email to Curt Daniels (curt.daniels@nationwidechildrens.com), Erwin Oechslin (erwin.oechslin@uhn.on.ca) or Koichiro Niwa (kniwa@aol.om). The ISACHD executives are waiting for your enthusiastic participation!
Regional News:

News from Latin America

By Luis Alday

The XXXVII and XIV Argentine Congresses of Cardiology and Pediatric Cardiology respectively will take place in Buenos Aires at the Buenos Aires Sheraton Hotel from Sunday 2 till Tuesday 4 October 2011. An International Society for Adult Congenital Heart Disease (ISACHD) and Argentine Society of Cardiology (SAC) Joint Session is being planned with the participation of Drs Roberta Williams (UCLA, Los Angeles, US) and Andrew Reddington (Hospital for Sick Children, Toronto, Canada). These sessions were inaugurated in 2009 with a great success and attendance by doctors from Argentina and neighboring countries.

News from Europe

By Helmut Baumgartner

The ESC congress in Paris was a big success. With more than 30.000 attendees ESC has become the largest cardiology meeting worldwide. Adult congenital heart disease was well represented with 11 sessions. Besides 7 pre-arranged sessions, 50 abstracts were presented in 4 oral sessions and poster sessions. During the general assembly of the ESC working group on Grown-up Congenital Heart Disease Pedro Trigo Trindade - current chairman of the WG - summarized the recent activities and future plans of the working group. The 2nd European GUCH meeting (March 4-5, 2011 in Paris) was well attended and will be followed by the 3rd meeting next year in Munich (March 16-17, 2012). The 6th European Echo Course on Congenital Heart Disease will also take place in Munich from November 16th to 19th, 2011. In addition, Congenital Heart Disease will be an important part of EuroEcho 2011 held from December 7th to 10th in Budapest, Hungary.

After publication and presentation of the new ESC guidelines on the management of grown-up congenital heart disease last year, the new ESC guidelines on the management of cardiovascular disease during pregnancy could be presented at this years ESC congress and simultaneously published in the European Heart Journal. Congenital heart disease is of course an important part of this document and working group members contributed essentially.

After the recommendations for cardiovascular magnetic resonance in adults with congenital heart disease (published 2010 in the European Heart Journal) the working group is now preparing two new position papers that should be ready for publication next year: "Recommendations for the organization of care and the training in GUCH" and "Recreational activities and GUCH". The European registry for ICD and CRT devices in pediatrics and adults with congenital heart disease and the European registry on pregnancy and heart
disease are still ongoing.

**Journal Watch**

Int J Cardiol. 2011 Aug 22. [Epub ahead of print]

**Detrimental impact of socioeconomic status on exercise capacity in adults with congenital heart disease.**


Source

Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital, Sydney Street, London, UK; National Heart and Lung Institute, Imperial College School of Medicine, London, UK.

Abstract

**OBJECTIVES:**

To evaluate the relationship between socioeconomic status (SES), access to physical activity resources, urban-rural dwelling, levels of pollution and exercise capacity in adult congenital heart disease (ACHD) patients.

**BACKGROUND:**

Exercise intolerance is prevalent in ACHD and the contributing factors are poorly understood.

**METHODS:**

A total of 1268 ACHD patients living in England who underwent cardiopulmonary exercise testing at our center were included. Neighborhood deprivation (English Indices of Deprivation), urban-rural dwelling, availability of green space, distance to the closest gym/fitness center and levels of pollution were estimated based on administrative data.

**RESULTS:**

Urban-rural dwelling, availability of green space and levels of pollution were unrelated to exercise capacity. Lower SES was associated with a significantly lower peak oxygen consumption (P<0.002) and heart rate reserve (P<0.004). This association was non-linear and most pronounced in ACHD patients with cardiac defects of medium complexity living in the most socioeconomically disadvantaged communities. Low SES was associated with higher prevalence of diabetes (P=0.015) and smoking (P=0.01). Coronary artery disease was rare in this young population and low SES was found to be related to exercise capacity independently of the presence of coronary artery disease.

**CONCLUSIONS:**
Living in poorer areas is associated with exercise intolerance in contemporary ACHD patients. Although low SES is linked to traditional cardiovascular risk factors, the deleterious effects of SES on exercise capacity seem to be only partially mediated via coronary artery disease. Reducing social inequalities in ACHD patients may have a positive effect on quality of life and long-term prognostic implications.

Am J Cardiol. 2011 Aug 20. [Epub ahead of print]

**The Cardiac Malpositions.**
Perloff JK.

Source
Ahmanson/UCLA Adult Congenital Heart Disease Center, Los Angeles, California.

Abstract

Dextrocardia was known in the 17th century and was 1 of the first congenital malformations of the heart to be recognized. Fifty years elapsed before Matthew Baillie published his account of complete transposition in a human of the thoracic and abdominal viscera to the opposite side from what is natural. In 1858, Thomas Peacock stated that "the heart may be congenitally misplaced in various ways, occupying either an unusual position within the thorax, or being situated external to that cavity." In 1915, Maude Abbott described ectopia cordis, and Richard Paltauf's remarkable illustrations distinguished the various types of dextrocardia. In 1928, the first useful classification of the cardiac malpositions was proposed, and in 1966, Elliott et al's radiologic classification set the stage for clinical recognition. The first section of this review deals with the 3 basic cardiac malpositions in the presence of bilateral asymmetry. The second section deals with cardiac malpositions in the presence of bilateral left-sidedness or right-sidedness. Previous publications on cardiac malpositions are replete with an arcane vocabulary that confounds rather than clarifies. Even if the terms themselves are understood, inherent complexity weighs against clarity. This review was designed as a guided tour of an unfamiliar subject.

Cardiol Young. 2011 Aug 19:1-8. [Epub ahead of print]

**Clinical application of a micro multiplane transoesophageal probe in congenital cardiac disease.**
Pushparajah K, Miller OI, Rawlins D, Barlow A, Nugent K, Simpson JM.

Source
Department of Congenital Heart Disease, Evelina Children's Hospital, Guy's & St Thomas' NHS Foundation Trust, London, United Kingdom.

Abstract

Aim
To assess the quality of imaging modalities of a new micro multiplane transoesophageal echocardiogram probe.

Method
This is a prospective study of micro transoesophageal echocardiogram S8-3t
probe used at a single institution between 15 December, 2009 and 15 March, 2010. The images were compared with standard paediatric or adult probes where possible. Assessors prospectively rated imaging quality - two dimensional, colour flow imaging, pulse wave, and continuous wave Doppler - with a subjective 4-point scale (1 = poor to 4 = excellent).

RESULTS:
A total of 24 studies were performed on 23 patients, with a median weight = 11.7 kilograms (2.6-72 kilograms) and a median age of 3 years (0.16-60 years). Of the 23 patients, one neonate (2.8 kilograms) had transient bradycardia on probe insertion. Imaging in patients less than 10 kilograms was of full diagnostic value and new information was obtained in eight out of ten patients. Pulse wave and continuous wave Doppler was consistently good across all weight groups. There were high frame rates and good imaging quality to a depth of 4-6 centimetres in all studies. A comparison with a larger alternative probe was available for 12 studies (weight 11.9-72 kilograms). The median micro transoesophageal two-dimensional image quality score was 3 (2-4) and 4 (3-4) with the comparative probe. For the 10- to 30-kilogram group, image quality with the micro transoesophageal echocardiogram probe was judged as inferior to larger standard probes. Adult sized patients had good imaging of near the field, allowing guidance for percutaneous device closure of the atrial septum.

Conclusion
The micro multiplane transoesophageal echocardiogram probe provides imaging of diagnostic quality in neonates. In larger patients, it offers good imaging of near field structures. In the intermediate-sized child (10-30 kilograms), standard paediatric probes provide better imaging.


**Education and support needs of the older adult with congenital heart disease.**
Riley JP, Habibi H, Banya W, Gatzoulis MA, Lau-Walker M, Cowie MR.

Source

Jillian P. Riley PhD RN NFESC Head of Post-graduate Education (Nursing and Allied Professionals) Royal Brompton & Harefield NHS Foundation Trust, London, UK Hajar Habibi MSc RN Staff Nurse Royal Brompton & Harefield NHS Foundation Trust, London, UK Winston Banya MSc Statistician Royal Brompton & Harefield NHS Foundation Trust, London, UK Michael A. Gatzoulis MD PhD FACC Professor of Cardiology Imperial College, London, UK Margaret Lau-Walker PhD BA RN Lecturer in Nursing King's College, London, UK Martin R. Cowie MD MSc FESC Professor of Cardiology Imperial College, London, UK.

Abstract

riley j.p., habibi h., banya w., gatzoulis m.a., lau-walker m. & cowie m.r. (2011) Education and support needs of the older adult with congenital heart disease. Journal of Advanced Nursing 00(0), 000-000. doi:10.1111/j.1365-2648.2011.05809.x

ABSTRACT:

Aim.
This article is a report of a study exploring health-related quality of life in adults with congenital heart disease and the extent to which it is associated with
patients' illness beliefs and emotional health.

Background.
A reduction in mortality in patients with congenital heart disease has led to an increasingly older population that faces new challenges. Studies in a younger adult population have reported inconsistent findings regarding health-related quality of life. Factors such as, the complexity of the congenital heart defect, have not been found to be associated with quality of life. The association between illness beliefs and health-related quality of life has not previously been reported.

Method.

Results.
The mean age of the study population was 37.2 years. Participants reported poorer physical functioning, role functioning and general health than a general population. High levels of anxiety were reported in 38% and high levels of depression in 17%. In multivariate analysis, higher levels of anxiety and depression were associated with poorer mental functioning and higher levels of depression with poorer physical quality of life.

Conclusion.
We have reported that high levels of anxiety and depression in an older population of patients with congenital heart disease are associated with poorer quality of life. This highlights the need to routinely assess anxiety and depression in this patient group and to provide psychological support appropriately.

Int J Cardiol. 2011 Aug 10. [Epub ahead of print]

Pathophysiology of chronic venous insufficiency in adults with a Fontan circulation (a pre-defined substudy of the CALF investigation).

Source
Children's Hospital Boston, Department of Cardiology, Boston, MA, United States; Brigham and Women's Hospital, Division of Cardiology, Boston, MA, United States.

Abstract
OBJECTIVES:
To investigate the pathophysiology of chronic venous insufficiency in adults with a Fontan circulation.

BACKGROUND:
Chronic venous insufficiency (CVI) is prevalent in adults with a Fontan circulation, however the underlying pathophysiology has not been identified. We defined the prevalence of venous reflux and obstruction in a Fontan population, identified associated factors, and assessed the hypothesis that venous reflux correlates with clinical CVI in adults with a Fontan circulation.
METHODS:

Fifty-one adult Fontan subjects from the Congenital Heart Disease in Adults: Lower Extremity Systemic Venous Health in Fontan Patients (CALF) study, ten control subjects without cardiac disease, and ten comparator subjects with repaired tetralogy of Fallot underwent lower extremity Doppler venous ultrasound. We investigated 10 venous segments in each subject for venous obstruction and reflux.

RESULTS:

The prevalence of venous reflux was significantly greater in the Fontan population compared to healthy controls (51% vs. 10%, p=0.03). Venous flow patterns demonstrated a range of pulsatility and biphasic flow in many subjects. There was no evidence of venous obstruction in any subject. Venous reflux did not correlate with clinical venous insufficiency; 49% of Fontan subjects with observed venous reflux demonstrated no clinical signs of venous disease. Predictors of venous reflux in multivariate analysis were single right ventricle (p=0.03), use of anti-arrhythmic medications (p=0.04), and family history of venous disease (p=0.003).

CONCLUSIONS:

Venous reflux is highly prevalent and venous obstruction is absent in adults with a Fontan circulation. Clinical CVI underestimates pathophysiologic venous insufficiency in adults with a Fontan circulation.


Association of atrial tachyarrhythmias with atrial septal defect, Ebstein's anomaly and Fontan patients.

Loomba RS, Chandrasekar S, Sanan P, Shah PH, Arora RR.

Source

James A Lovell Federal Health Center/Chicago Medical School, 3001 North Green Bay Road, North Chicago, IL 60064, USA.

Abstract

The number of adults with congenital heart disease is increasing as medical and surgical palliation for congenital heart lesions improves. With this comes long-term complications of congenital heart disease such as the increased risk of atrial tachyarrhythmias. Atrial septal defect, Ebstein's anomaly and post-Fontan patient subsets are particularly important to focus on due to their unique characteristics and association with atrial tachyarrhythmias. Reviews, randomized controlled trials, and meta-analyses were obtained using electronic search strategies such as Medline and the Cochrane Library. References of electronically obtained studies were then used to obtain additional relevant studies. Sources were deemed relevant if they discussed the relationship between atrial septal defects/Ebstein's anomaly/Fontan procedure and atrial tachyarrhythmias in respect to incidence, mechanism, recurrence or treatment. Selected sources were then stratified on the basis of quality. Patients in these subsets of congenital heart disease are at increased risk of atrial tachyarrhythmias for a variety of reasons when compared with the general
population. It is necessary for pediatric and adult cardiologists alike to understand these differences, as well as their implications in diagnosis and management of such occurrences.


**Development of an international research agenda for adult congenital heart disease nursing.**
Goossens E, Fleck D, Canobbio MM, Harrison JL, Moons P; on behalf of the International Society of Adult Congenital Heart Disease (ISACHD) Nursing Network.

Source

Center for Health Services and Nursing Research, Katholieke Universiteit Leuven, Leuven, Belgium.

Abstract

**BACKGROUND:**
Since the population of adults with congenital heart disease (CHD) is growing, the role of nurse specialists is expanding. In order to advance ACHD nursing, the establishment of an international nursing research agenda is recommended. We aimed to investigate research priorities as perceived by nurse specialists and researchers in ACHD.

**METHODS:**

We applied a sequential quan-qual design. In the quantitative phase, a two-round Delphi study was conducted, in which 37 nurse specialists and nurse researchers in ACHD care participated. Respondents assessed the level of priority of 21 research topics using a 9-point rating scale (1=no priority at all; 9=very high priority). In the qualitative phase, semi-structured interviews were performed with six selected Delphi panelists, to scrutinize pending research questions.

**RESULTS:**

This study revealed that priority should be given to studies investigating knowledge and education of patients, outcomes of Advanced Practice Nursing, quality of life, transfer and transition, and illness experiences and psychosocial issues in adults with CHD. A low priority was given to post-operative pain, sexual functioning, transplantation in ACHD, and health care costs and utilization. Agreement about the level of priority was obtained for 14 out of 21 research topics.

**CONCLUSION:**

Based on this study, we could develop an international research agenda for ACHD. Researchers ought to focus on these areas of highest priority, in order to expand and strengthen the body of knowledge in ACHD nursing.

Int J Cardiol. 2011 Jul 27. [Epub ahead of print]

**Therapy for pulmonary arterial hypertension due to congenital heart disease and Down’s syndrome.**

Source

Department of Cardiology, Second University of Naples, Monaldi Hospital, Naples, Italy.

Abstract

BACKGROUND:

Oral bosentan is effective in pulmonary arterial hypertension (PAH) related to congenital heart disease (CHD). In patients with Down's syndrome, the effect of bosentan is largely unknown. Aim of the study was to evaluate the long-term effects of bosentan in adult patients with CHD-related PAH with and without Down's syndrome.

METHODS:

WHO functional class, resting oxygen saturation, 6-minute walk test (6MWT) and hemodynamics were assessed at baseline and after 12 months of bosentan therapy in patients with CHD-related PAH with and without Down's syndrome.

RESULTS:

Seventy-four consecutive patients were enrolled: 18 with and 56 without Down's syndrome. After 12 months of bosentan therapy, both with and without Down's syndrome patients showed an improvement in WHO functional class (Down: 2.5±0.5 vs 2.9±0.6, p=0.005; controls: 2.5±0.5 vs 2.9±0.5, p=0.000002), 6-minute walk distance (Down: 288±71 vs 239±74 m, p=0.0007; controls: 389±80 vs 343±86 m, p=0.0003), and hemodynamics (pulmonary flow, Down: 4.0±1.6 vs 3.5±1.4 l/m² (2), p=0.006; controls: 3.5±1.4 vs 2.8±1.0 l/m² (2), p=0.0005; pulmonary to systemic flow ratio, Down: 1.4±0.7 vs 1.0±0.4, p=0.003; controls: 1.1±0.7 vs 0.9±0.3, p=0.012; pulmonary vascular resistance index, Down: 15±9 vs 20±13 WU/m² (2), p=0.007; controls: 20±10 vs 26±15 WU/m² (2), p=0.002). No differences in the efficacy of therapy were observed between the two groups.

CONCLUSIONS:

Bosentan was safe and well tolerated in adult patients with CHD-related PAH with and without Down's syndrome during 12 months of treatment. Clinical status, exercise tolerance, and pulmonary hemodynamics improved, regardless of the presence of Down's syndrome.


Diagnosis of pulmonary hypertension in the congenital heart disease adult population impact on outcomes.

Lowe BS, Therrien J, Ionescu-Ittu R, Pilote L, Martucci G, Marelli AJ.

Abstract

OBJECTIVES:

The aim of this study was to assess the impact of the diagnosis of pulmonary
hypertension (PH) on mortality, morbidity, and health services utilization (HSU) in an adult congenital heart disease (CHD) population.

BACKGROUND:

Although PH is a well-recognized complication of CHD, population-based studies of its significance on the survival and functional capacity of patients are uncommon.

METHODS:

A retrospective longitudinal cohort study was conducted in an adult CHD population with 23 years of follow-up, from 1983 to 2005. The prevalence of PH was measured in 2005. Mortality, morbidity, and HSU outcomes were compared between patients with and without diagnoses of PH using multivariate Cox (mortality and morbidity) and Poisson (HSU) regression models within a subcohort matched for age and CHD lesion type.

RESULTS:

Of 38,430 adults alive with CHD in 2005, 2,212 (5.8%) had diagnoses of PH (median age 67 years, 59% women). The diagnosis of PH increased the all-cause mortality rate of adults with CHD more than 2-fold compared with patients without PH (hazard ratio [HR]: 2.69; 95% confidence interval [CI]: 2.41 to 2.99). Morbid complications including heart failure and arrhythmia occurred with a 3-fold higher risk compared with patients without PH (HR: 3.01; 95% CI: 2.80 to 3.22). The utilization of inpatient and outpatient services was increased, especially cardiac catheterization, excluding the index diagnostic study (rate ratio: 5.04; 95% CI: 4.27 to 5.93) and coronary and intensive care hospitalizations (rate ratio: 5.03; 95% CI: 4.86 to 5.20).

CONCLUSIONS:

A diagnosis of PH in adults with CHD is associated with a more than 2-fold higher risk for all-cause mortality and 3-fold higher rates of HSU, reflecting high morbidity.

Opinions of physicians regarding problems and tasks involved in the medical care system for patients with adult congenital heart disease in Japan.

Ochiai R, Murakami A, Toyoda T, Kazuma K, Niwa K.

Source
Department of Adult Nursing/Palliative Care Nursing, School of Health Science and Nursing, Graduate School of Medicine, Tokyo, Japan. rochiai-tky@umin.ac.jp

Abstract

BACKGROUND:
The aim of this study is to summarize the opinions of physicians regarding problems and tasks involved in the medical care system for patients with adult
congenital heart disease (ACHD) in Japan.

METHODS AND RESULTS:
We conducted a semistructured interview with 30 subjects consisting of 13 pediatric cardiologists, 11 cardiovascular surgeons, and six cardiologists who were selected from among the randomly sampled medical facilities meeting each of the following institutional criteria: (1) facilities with ≥50 ACHD outpatients; (2) facilities with ACHD-specialized outpatient clinic; (3) facilities that are members of the Japanese Association of Children's Hospitals and Related Facilities. The interview time was 27-91 minutes (mean, 70.0). The age of the subjects ranged from 36 to 62 years (mean, 46.7), and they had 0.5-34 years (mean, 16.2) of experience in treating congenital heart disease. From an analysis of interview details, the following four themes were extracted (in descending order of the number of comments): "(1) Who should treat ACHD" (comments by 29 subjects), "(2) Centralization of medical care" (comments by 29 subjects), "(3) What is the role of children's hospitals in ACHD?" (comments by 24 subjects), and "(4) Psychosocial problems" (comments by 24 subjects).

CONCLUSIONS:
Regional ACHD centers need to be established to promote centralization of patients, physicians, and educational function. This will provide higher quality medical service to more patients in the near future.


Congenital heart defects in adulthood.
Diller GP, Breithardt G, Baumgartner H.

Source
Kardiologisches Zentrum für Erwachsene mit angeborenen und erworbenen Herzfehlern (EMAH), Münster, Germany. Gerhard.Diller@ukmuenster.de

Abstract
BACKGROUND
More than 90% of children with congenital heart defects now survive into adulthood; just a few decades ago, survival was rare, particularly among patients with complex defects. The new population of adults with congenital heart disease presents a special challenge to physicians from all of the involved specialties.

METHODS:
Selective literature review.

RESULTS AND CONCLUSION:
A complete cure of the congenital heart defect in childhood is exceptional, and most adult patients continue to suffer from residual problems and sequelae. Further surgery or catheter interventions may be needed. Potential late complications include arrhythmias, heart failure, pulmonary hypertension, endocarditis, and thromboembolic events. The management of these patients during pregnancy or non-cardiac surgery remains a challenge. If this evolving patient population is to receive the best possible care, the adequate provision of specialized medical services is a necessary, but not sufficient, condition:
patients and their referring physicians will also need to be aware that these services are available, and then actually make use of them. Moreover, optimal communication among all of the involved physicians is essential.

Int J Cardiol. 2011 Jul 14. [Epub ahead of print]

**Treatment of segmental pulmonary artery hypertension in adults with congenital heart disease.**
Schuuring MJ, Bouma BJ, Cordina R, Gatzoulis MA, Budts W, Mullen MP, Vis JC, Celermajer D, Mulder BJ.

Source
Academic Medical Centre, Department of Cardiology, Amsterdam, The Netherlands; Interuniversity Cardiology Institute of the Netherlands, Utrecht, The Netherlands.

Abstract

INTRODUCTION:
Pulmonary arterial hypertension (PAH) in patients with congenital heart disease (CHD) usually has a homogeneous pressure distribution. More rarely, complex CHD patients have segmental PAH. This is often post-surgically. The characteristics of these patients and their responsiveness to specific pulmonary vasodilator therapy have not been described.

METHODS:
Seven adults with segmental PAH complicating CHD were treated at 3 specialized adult CHD centers between January 2006 and December 2010. Clinical characteristics, six minute walking distances (6MWD), laboratory tests and images were obtained from medical records and the responses to Bosentan, an endothelin-1 receptor antagonist, were assessed.

RESULTS:
All patients (mean age 32 (23-42) years, five females) had a primary diagnosis pulmonary atresia (PA), four with major aortopulmonary collateral arteries (MAPCAs). Four segmental PAH patients had a right pulmonary artery stenosis, two a left pulmonary artery stenosis and one a unilateral MAPCA stenosis. All patients were symptomatic (functional class II or III) and bosentan was started empirically. Bosentan treatment led to a significant improvement in functional class compared to baseline (1.7±0.5 versus 2.4±0.5; p<0.01). Mean 6MWD (available in 6 patients) increased by 62m (22-150m) from 386±135 to 448±133m (p=0.03) after 12months treatment. Most improvement was seen in patients with low baseline 6MWD. Higher baseline exercise heart rate was significantly associated with lesser improvement in 6MWD (r=-0.91 p=0.01). Laboratory results did not change after initiation of bosentan treatment.

CONCLUSION:
This small retrospective case series suggested a significant improvement of functional class and exercise capacity after bosentan treatment in patients with segmental PAH. These findings warrant a prospective study of the potential benefit of selective pulmonary vasodilator therapy in these complex patients. Therefore, we call on treating physicians to share similar cases.

**Status and future needs of regional adult congenital heart disease centers in Japan.**

Source
Department of Adult Nursing/Palliative Care Nursing, School of Health Science and Nursing, Graduate School of Medicine, The University of Tokyo.

Abstract
Background: Although the prevalence of adult congenital heart disease (ACHD) in Japan continues to rise, the number and geographic distribution of facilities potentially serving as regional ACHD centers remains unknown. We examined trends in ACHD care in Japan to identify needs and to determine potential regional responses to this growing patient population.

Methods and Results: A descriptive, cross-sectional, nationwide survey was conducted to assess the status and needs of cardiology specialists related to providing ACHD care. Questionnaires were mailed to 138 cardiology departments located in 8 geographical regions throughout Japan; respondents were asked to document the status and future direction of ACHD care for each facility. Of the 109 facilities that responded, approximately one-third currently treat or plan to treat all ACHD patients. Fourteen facilities (12.8%) fulfilled all criteria for becoming regional ACHD centers. Although each regional center was projected to serve a population of 9.1 million, in 2 regions, no centers possessed the necessary care structure.

Conclusions: Our findings revealed a shortage of adult cardiologists dedicated to ACHD care. Moreover, basic as well as formal fellowship ACHD training was deemed necessary. In Japan, the number of potential regional ACHD centers has just reached international standards. However, based on the geographic gaps documented here, a strategy other than regional centralization might be required to deliver adequate ACHD care to rural areas. (Circ J 2011; 75: 2220-2227).


**Growth differentiation factor 15: an additional diagnostic tool for the risk stratification of developing heart failure in patients with operated congenital heart defects?**

Source
Division of Paediatric Cardiology, Department of Paediatrics, London Health Sciences Centre, University of Western Ontario, Canada.
kambiz.norozi@lhsc.on.ca

Abstract
BACKGROUND: Many young adults who have congenital heart defects develop heart failure despite corrective surgeries. Growth differentiation factor 15 (GDF-15) has an established role as a marker for risk stratification and mortality both in patients after acute myocardial infarction and in patients with heart failure. Our aim was
to establish a role for GDF-15 for monitoring heart failure in operated congenital heart defects (ACHD). This potential biomarker was validated through comparison with maximal oxygen uptake (VO\(_{2\text{max}}\)) and to another biomarker, N-terminal pro-brain natriuretic peptide (NT-proBNP).

**METHODS:**
A total of 317 ACHD patients (129 females) with an average age of 26.5 ± 8.5 years (mean ± SD) enrolled in the study. We studied the relation between GDF-15 and NT-proBNP and VO\(_{2\text{max}}\)% (percent predicted for age and gender). The cutoffs for the groups were as follows: NT-proBNP <100, 100 to 300, and >300 pg/mL; VO\(_{2\text{max}}\)% <65%, 65% to 85%, and >85% of predicted normal.

**RESULTS:**
Significant differences in mean GDF-15 levels were found between the NT-proBNP <100 and NT-proBNP >300 groups, as well as between the 100 to 300 and the >300 groups. For VO\(_{2\text{max}}\)% significant differences were found in GDF-15 levels between <65% and >85% and between <65% and 65% to 85%, respectively. The lowest mean GDF-15 was found in groups with NT-proBNP <100 pg/mL and VO\(_{2\text{max}}\)% >85%. The highest mean GDF-15 was found in the groups with NT-proBNP >300 pg/mL and VO\(_{2\text{max}}\)% <65%. A subgroup analysis, including 82 patients with operated tetralogy of Fallot, showed that patients in the New York Heart Association I class have significantly lower NT-proBNP and GDF-15 level and markedly higher VO\(_{2\text{max}}\) compared with the patients in higher New York Heart Association class.

**CONCLUSION:**
Growth differentiation factor 15 might be used as a surrogate marker for latent heart failure and could help to identify patients with ACHD who are at risk for developing heart failure, even if they are clinically asymptomatic.


*Affective functioning and social cognition in Noonan syndrome.*

Wingbermuhle E, Egger JI, Verhoeven WM, van der Burgt I, Kessels RP.

Source
Vincent van Gogh Institute for Psychiatry, Venray, The Netherlands.

Abstract

**BACKGROUND:**
Noonan syndrome (NS) is a common genetic disorder, characterized by short stature, facial dysmorphism, congenital heart defects and a mildly lowered IQ. Impairments in psychosocial functioning have often been suggested, without, however, systematic investigation in a clinical group. In this study, different aspects of affective processing, social cognition and behaviour, in addition to personal well-being, were assessed in a large group of patients with NS.

**Method**
Forty adult patients with NS were compared with 40 healthy controls, matched with respect to age, sex, intelligence and education level. Facial emotion recognition was measured with the Emotion Recognition Task (ERT), alexithymia with both the 20-item Toronto Alexithymia Scale (TAS-20) and the Bermond-Vorst Alexithymia Questionnaire (BVAQ), and mentalizing with the Theory of Mind (ToM) test. The Symptom Checklist-90 Revised (SCL-90-R) and the Scale for Interpersonal Behaviour (SIB) were used to record aspects of psychological well-being and social interaction.
RESULTS:
Patients showed higher levels of cognitive alexithymia than controls. They also experienced more social distress, but the frequency of engaging in social situations did not differ. Facial emotion recognition was only slightly impaired.

CONCLUSIONS:
Higher levels of alexithymia and social discomfort are part of the behavioural phenotype of NS. However, patients with NS have relatively intact perception of emotions in others and unimpaired mentalizing. These results provide insight into the underlying mechanisms of social daily life functioning in this patient group.


**Risk factors and early outcomes of multiple reoperations in adults with congenital heart disease.**
Holst KA, Dearani JA, Burkhart HM, Connolly HM, Warnes CA, Li Z, Schaff HV.

Source
Division of Cardiovascular Surgery, Mayo Clinic and Foundation, Rochester, Minnesota 55905, USA.

Abstract

BACKGROUND:
Advances in treatment of congenital heart disease (CHD) have resulted in most patients surviving to adulthood. Despite surgical "correction," the need for reoperation(s) persists, and there are few outcome data. This study examined early postoperative results to determine risk factors for cardiac injury and early death in adults with CHD undergoing repeat median sternotomy.

METHODS:
Data from the most recent median sternotomy of 984 adults (49% male) with CHD were analyzed. Mean age at operation was 36.4 years. Diagnoses were conotruncal anomaly, 361 (37%); Ebstein/Tricuspid valve, 174 (18%); pulmonary stenosis/right ventricular outflow tract obstruction, 92 (9%); single ventricle, 71 (7%); atrioventricular septal defect, 64 (7%); subaortic stenosis, 62 (6%); aortic arch abnormalities, 23 (2%); anomalous pulmonary vein, 21 (2%); Marfan syndrome, 14 (1%); and other, 102 (10%).

RESULTS:
Overall early mortality was 3.6%; including 2%, 6%, 7%, and 0% at sternotomy 2 (n=597), 3 (n=284), 4 (n=72), and 5+ (n=31), respectively. Cardiac injury occurred in 6%. Independent predictors of cardiac injury were single-ventricle diagnosis and increased number of prior sternotomies. Increased time from previous sternotomy decreased the incidence of cardiac injury. Independent risk factors for early death were urgent operation, single-ventricle diagnosis, and longer bypass time. Increased preoperative ejection fraction decreased early mortality.

CONCLUSIONS:
Subsequent sternotomy showed increased early mortality, yet neither sternotomy number nor cardiac injury was an independent predictor of early death. Two variables were protective: early mortality was reduced with
increased ejection fraction and cardiac injury was less likely with increased interval from the previous sternotomy.


**Cheatham-Platinum stent for native and recurrent aortic coarctation in children and adults: immediate and early follow-up results.**

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Source
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Abstract

**OBJECTIVE:**
To present our institutional experience of endovascular Cheatham-Platinum stent implantation in children and adults with native and recurrent aortic coarctation.

**METHODS:**
Between August 2007 and November 2009, 45 patients had aortic coarctation treated with 47 stents implantation. We preferred primarily stent implantation in adult patient with coarctation, in children more than five years-old it is preferred in cases of aneurysm, subatretic or blind coarctation and coarctation with patent ductus arteriosus or in restenosis. Files of stent-implanted patients were retrospectively analyzed in terms of patients' demographic features, echocardiographic and angiographic findings both before and after procedure. Patients were grouped as Group 1: native coarctation and Group 2: recoarctation developed after either surgery or balloon angioplasty. Findings of the cases' were compared using paired and unpaired Student's t, Mann-Whitney U and Chi-square tests.

**RESULTS:**
Sixteen covered and 31 bare totally 47 balloon expandable stents were implanted in 45 patients. The mean follow up duration was 12.1 ± 7.1, median 11 months (2-29 months). There was no procedure related death. In two patients two stents were implanted in tandem. While the coarctation of the aorta was native in 26 patients (functionally interrupted aortic arch in one), recoarctation was detected in 7 patients after surgery, in 8 patients after balloon angioplasty, in 4 patients both after surgery and balloon angioplasty. One patient had functionally interrupted aortic arch perforated with guide wire and then covered stent implanted. The mean age 12.2 ± 5.9 years (5-33 years) and mean body mass index was 21 ± 3.7 kg/m² (14.8-31 kg/m²). Considering all cases, a statistically significant decrease in both the invasive and echocardiographic gradients (p<0.001 for both) and statistically significant increase in lesion diameter (p<0.001) were detected. The decrease in invasive and echocardiographic gradients and increase in lesion diameter is statistically significant in each group also (p<0.001, <0.001 and <0.001 for both groups, respectively). Before the procedure, the invasive gradient was significantly higher and the lesion diameter was significantly lower in group I than in group II (p=0.002 and p=0.005, respectively). Also the percentage of decrease in gradient and increase in diameter was statistically higher in group 1 than in...
CONCLUSION:
Our early and short-term follow-up results indicate that stent implantation is safe and very effective in reducing coarctation gradient and increasing lesion diameter both in native coarctation and recoarctation.


Risk factors for death after adult congenital heart surgery in pediatric hospitals.
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Source
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Abstract

BACKGROUND:
Despite the central role that pediatric hospitals play in the surgical treatment of congenital heart disease, little is known about outcomes of adult congenital cardiac surgical care in pediatric hospitals. Risk factors for inpatient death, including adult congenital heart (ACH) surgery volume, are poorly described.

METHODS AND RESULTS:
We obtained inpatient data from 42 free-standing pediatric hospitals using the Pediatric Health Information System data base 2000 to 2008 and selected ACH surgery admissions (ages 18 to 49 years). We examined admission characteristics and hospital surgery volume. Of 97 563 total (pediatric and adult) congenital heart surgery admissions, 3061 (3.1%) were ACH surgery admissions. Median adult age was 22 years and 39% were between ages 25 to 49 years. Most frequent surgical procedures were pulmonary valve replacement, secundum atrial septal defect repair, and aortic valve replacement. Adult mortality rate was 2.2% at discharge. Multivariable analyses identified the following risk factors for death: age 25 to 34 years (adjusted odds ratio [AOR], 2.1; P=0.009), age 35 to 49 years (AOR, 3.2; P=0.001), male sex (AOR, 1.8; P=0.04), government-sponsored insurance (AOR, 1.8; P=0.03), and higher surgical risk categories 4+ (AOR, 21.5; P=0.001). After adjusting for case mix, pediatric hospitals with high ACH surgery volume had reduced odds for death (AOR, 0.4; P=0.003). There was no relationship between total congenital heart surgery volume and ACH inpatient mortality.

CONCLUSIONS:
Older adults, male sex, government-sponsored insurance, and greater surgical case complexity have the highest likelihood of in-hospital death when adult congenital surgery is performed in free-standing pediatric hospitals. After risk-adjustment, pediatric hospitals with high ACH surgery volume have the lowest inpatient mortality.


Adults with congenital heart disease: patients' knowledge and concerns
about inheritance.
van Engelen K, Baars MJ, van Rongen LT, van der Velde ET, Mulder BJ, Smets EM.

Source
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Abstract
With recent advances in medical and surgical management, most patients with congenital heart disease (CHD) survive to reproductive age. Current guidelines recommend counseling about inheritance and transmission of CHD to offspring. We evaluated whether adult CHD patients recalled having received information about the inheritance of their CHD, patients' knowledge about inheritance and their concerns in this regard. A questionnaire was sent to 486 non-syndromic CHD patients aged 20-45 years. We received 332 useful questionnaires (response rate 68%). One-third (33%) of patients recalled receiving information about inheritance of CHD from their cardiologist, and 13% had consulted a clinical geneticist. Eight percent of patients who were considering having children estimated the recurrence risk for their own offspring to be 1% or lower, whereas one-fourth (25%) estimated it to be higher than 10%. According to our classification, 44% estimated the recurrence risk in a correct range of magnitude. Additional information about inheritance of CHD was desired by 41% of patients. Forty-two percent of patients considering having children reported concerns about transmitting CHD to offspring. We conclude that a substantial proportion of adult CHD patients lacks knowledge and desires more information about inheritance, indicating a need for better patient education. Current guidelines and/or their implementation do not seem to meet the needs of these patients. A dedicated program of counseling for adults with CHD has to be developed to optimize knowledge and satisfaction with information provision and to reduce or manage concerns regarding inheritance of CHD.


Preventing sudden death in the adult with congenital heart disease.
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Abstract
Adults with repaired congenital heart disease represent a complex and heterogeneous group of patients that are increasingly surviving beyond childhood. Patients have a variety of diagnoses that include specific structural anomalies, assorted physiologic derangements, and unique techniques for surgical repair. During long-term follow-up, even the most excellent surgical outcome may result in anatomic stenosis and insufficiency, cardiac rhythm disturbance, and myocardial dysfunction. Any of these abnormalities, alone or in combination, may result in significant morbidity and mortality. Sudden death is commonly due to a cardiac etiology and arrhythmias are frequently suspected
to be the cause. Unfortunately, arrhythmias are difficult to predict and may be potentially lethal at their initial presentation. In addition, a wide spectrum of arrhythmias, both supraventricular and ventricular, are possible, depending on the specific diagnosis and type of repair performed. This review will focus primarily on arrhythmias as a cause of sudden cardiac death in this patient population. Particularly important considerations regarding arrhythmias in adults with congenital heart disease include the fact that supraventricular arrhythmias may be as lethal as ventricular arrhythmias, arrhythmia substrates develop in a unique manner when compared with other adult cohorts, and the electrophysiologic status of patients with repaired congenital heart disease must be considered together with the hemodynamic result.


Outcomes of hospitalization in adults in the United States with atrial septal defect, ventricular septal defect, and atroventricular septal defect.

Source

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Abstract

Atrial septal defect, ventricular septal defect (VSD), and atroventricular septal defect (AVSD) are among the most common congenital heart lesions, with most children surviving to adulthood. However, the clinical course of these patients is largely unknown, particularly pertaining to inpatient care. The purpose of this study was to assess hospitalizations for septal defects in adults with congenital heart disease (CHD) and risk factors associated with significant morbidity and mortality. The 2007 Nationwide Inpatient Sample was used to assess national prevalence of hospitalizations in adults with CHD with septal defects. Co-morbidities and risk factors for mortality were also determined. There were 84,308 adult CHD admissions in the United States in 2007. Fifty-four percent of adult CHD admissions had diagnoses of septal defects, with 48% having atrial septal defect, 7% having VSD, and 0.4% having AVSD. Overall in-hospital mortality was 2.1%. Common co-morbidities included arrhythmias (31%), heart failure (20%), and diabetes mellitus (18%). On multivariable analysis, independent risk factors for mortality included presence of VSD (odds ratio 3.1, 95% confidence interval [CI] 1.5 to 6.5), trisomy 21 (odds ratio 2.9, 95% CI 1.1 to 7.5), and pulmonary hypertension (odds ratio 1.5, 95% CI 1.0 to 2.4). In conclusion, this study of hospitalizations in adults with septal defects found that admissions are common and associated with significant co-morbidities. Overall mortality is low but is increased in patients with VSD. Cardiac and noncardiac co-morbidities are commonly encountered. Many noncardiac conditions, including trisomy 21 and the youngest and oldest groups, are associated with an increased risk of death.

Genetic counseling in the adult with congenital heart disease: what is the role?
Burchill L, Greenway S, Silversides CK, Mital S.

Source
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Abstract
New discoveries using high-resolution methods for detecting genetic aberrations indicate that the genetic contribution to congenital heart disease has been significantly underestimated in the past. DNA diagnostics have become more accessible and genetic test results are increasingly being used to guide clinical management. Adult congenital heart disease specialists seeking to counsel adults with congenital heart disease about the genetic aspects of their condition face the challenge of keeping abreast of new genetic techniques and discoveries. The emphasis of this review is on the genetic basis of structural cardiovascular defects. A framework for identifying adult congenital heart disease patients most likely to benefit from genetic testing is suggested, along with a summary of current techniques for genetic testing. The clinical and ethical challenges associated with genetic counseling are highlighted. Finally, emerging technologies and future directions in genetics and adult congenital heart disease are discussed.


Monitoring the patient with transposition of the great arteries: arterial switch versus atrial switch.

Roche SL, Silversides CK, Oechslin EN.

Source
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Abstract
Sufficient time has passed that adult congenital heart disease (ACHD) specialists now frequently encounter survivors born with complete transposition of the great arteries and palliated with an atrial or arterial switch procedure. To ensure the ongoing health of these patients, it is of paramount importance that their surgeries are understood and that physicians are aware of and remain vigilant for potential late complications. Adult survivors should be assessed annually in a regional ACHD center. Clinical assessment, electrocardiogram, and multimodality imaging are the mainstay of routine monitoring. Doppler echocardiography is the first-line imaging modality; other diagnostic tests are tailored to seek specific long-term complications. Clinicians, specialists in cardiovascular imaging, nurses and others involved in the delivery of care need special training and expertise. Care for these complex patients is best provided by multidisciplinary teams located in regional ACHD centers with access to adequate human and structural resources.

Parameters of arterial function and structure in adult patients after coarctation repair.

Source
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Abstract
Regardless of a successful operation, patients with coarctation of aorta (CoAo) are exposed to the risk of hypertension and a propensity to vascular and end-organ damage. The aim of this study is to evaluate the influence of residual aorta stenosis as well as the age at the operation on the parameters of arterial function and structure in patients after CoAo repair. Eighty-five patients after CoAo repair (53 males; mean age: 34.6 ± 10.3 years, mean age at the repair: 10.9 ± 8.2 years) were enrolled in the study. The control group consisted of 30 healthy subjects (18 males; mean age: 33.6 ± 8.2 years). Indices of systemic arterial remodeling [flow-mediated dilatation (FMD), nitroglycerine-mediated vasodilatation (NMD), carotid intima-media thickness (IMT), pulse wave velocity (PWV)] were analyzed in all study patients. In normotensive patients after CoAo repair (47/55%), a significantly increased PWV was observed in comparison to the control group (6.8 ± 1.2 vs. 5.4 ± 0.9 m/s; p = 0.003), with no difference in IMT values (0.53 ± 0.1 vs. 0.51 ± 0.1 mm; p = 0.06). Mean FMD (4.8 ± 2.8 vs. 8.5 ± 2.3%; p = 0.00003) and NMD (11.3 ± 4.6 vs. 19.8 ± 7.2%; p = 0.00001) were lower than in the controls. In patients with a residual aorta stenosis (46/54%), defined as an arm-leg pressure gradient ≥ 20 mmHg, no differences were found within the scope of both systolic and diastolic blood pressure and of all of the examined vascular parameters. No significant correlations were revealed between the vascular parameters and the gradient across descending aorta as well as the age at the operation. Residual stenosis in the descending aorta does not affect the arterial vasodilatation nor stiffness in patients after CoAo repair. An early surgery does not influence the remodeling of the vessels, which supports the thesis that CoAo is a generalized vascular disease and that even an early operation cannot prevent the progressive and vascular changes and end-organ damage.


Adult congenital heart care in a pediatric setting-a patient’s perspective.

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Source
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Abstract
BACKGROUND:
Congenital heart disease (CHD) affects approximately 1% of all live births today. With improvements in diagnostic, medical, surgical, and interventional procedures, 85% or more of all infants with CHD will reach adulthood. The
number of adults living with CHD has been estimated to exceed 1 million and now exceeds the number of pediatric CHD patients. Because residual problems following intervention for congenital heart lesions can present during the adult years and complicate the well-being of these patients, ongoing care is warranted. Adult care providers have a limited knowledge of the complexities of CHD. The American College of Cardiology recommends the establishment of adult CHD clinics with both pediatric and adult cardiologists. Although our clinic is located in a pediatric hospital, little attention has been placed on where these clinics should be located—an adult setting or a pediatric setting. We sought to determine the adult CHD patient's perspective on being seen in a pediatric setting.

OBJECTIVE:
The objective of the study was to determine the perspective of adults with CHD receiving follow-up care in a pediatric setting.

METHODS:
A pilot 11-question anonymous patient satisfaction survey with no personal identifiers and no diagnoses was sent to all patients who had attended the adult congenital heart clinic at our pediatric hospital medical center during a 2-year period.

RESULTS:
From our respondents, 96% did not have any concerns with being seen in a pediatric setting for adult congenital heart care, and 98% would recommend our clinic to other patients.

CONCLUSION:
Care for the adult with CHD involves multiple care providers. The most important finding from the patient's perspective is knowledge of the complexities of congenital heart lesions and possible future complications. There was little impact from being seen in a pediatric hospital setting.