January, 2012

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In This Issue

International Society for Adult Congenital Heart Disease

ISACHD Newsletter

President's Message

by *Barbara J.M. Mulder*
Dear ISACHD members,

What can you expect from ISACHD in 2012? A membership certificate is being prepared for all ISACHD members. It is a proof of membership which you can use for congresses endorsed by ISACHD. Our aim is to achieve a reduced registration fee for (paying only!) ISACHD members at all endorsed ACHD meetings worldwide. Erwin Oechslin, as the chairman of the education working group, is leading the process of endorsement. On the ISACHD website you may find a list of endorsed meetings for the next coming months.

The education working group is very active in preparing a web-based teaching course. Els Pieper from Groningen has prepared an excellent preliminary program, which is now being finalized. Erwin foresees that all talks will have been recorded by the end of this year. In addition, the working groups on Global Health, led by Curt Daniels, and on Global Research, led by Koichiro Niwa, are proceeding rapidly with their activities. Collaboration with The World Heart Federation and Project Hope will offer opportunities for health care projects in underserved areas.

Several general topics for joined research worldwide were chosen at the AHA meeting in Orlando last November: sports, QoL, lost to follow-up, and ACHD care facilities. The working groups have conference calls regularly, and at the ACC in Chicago the actual status of their activities will be presented. The ISACHD meeting in Chicago is scheduled for Sunday evening, March 25th, from 7-9 pm. More details will follow.
In Chicago, ISACHD's leadership will change, and Curt Daniels will become the new president. This will undoubtly bring new ideas and plans for the rest of the year.

The ISACHD executives wish you all a happy and healthy 2012!
We would love to welcome you at our meetings. And we hope to welcome as many new ISACHD members as we did last year!

Barbara J.M. Mulder

Representative, South America
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Hospital de Niños
Córdoba, Argentina

Representative, Europe
Helmut Baumgartner, M.D.
Adult Congenital And Valvular Heart Center
Universitatsklinikum Muenster
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Representative, Asia-Pacific
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Chiba, Japan

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FPMR
Raleigh, NC, USA

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Gary Webb, MD
1994-1996

Carol Warnes, MD
1996-1998

Richard Liberthson, MD

President

Click here for example certificate.

News from the psychosocial section

Adrienne Kovacs

Ethics of the heart II: Ethical and policy changes in pediatric and adult congenital heart disease. March 16th-17th, Philadelphia.

The purpose of this educational activity is to convene a multidisciplinary panel of nationally known experts to identify and discuss the most important ethical issues in congenital heart disease, providing a forum for clinicians, ethicists, chaplains, social workers, public health workers, and trainees to discuss developments in this field. Additional goals of this course are to incorporate the psychosocial, religious, and spiritual needs of congenital heart disease patients and their families into care plans. In addition, this course aims to explore issues related to quality of life, advance care planning, and palliative care in pediatric and adult populations.

Click here to see the flyer for Ethics of the Heart.

Regional News:

News from Europe:
Helmut Baumgartner

-8th DHZB Lange Symposium: Transposition of Great Arteries, Old Story--New Aspects January 21-22, 2012, Berlin, Germany.
-The 3rd Advanced course on Adult Congenital Heart Disease, March 16,17th in Munich.

1998-2000

Daniel Murphy, Jr., MD
2000-2002

Thomas P. Graham, Jr., MD
2002-2004

Michael A. Gatzoulis, MD, PhD, FACC
2004-2006

Jack M. Colman, MD
2006-2008

Michael J. Landzberg, MD
2008-2010

-2nd International Congress on Cardiac Problems in Pregnancy, May 17-20th Berlin, Germany.
-ESC meeting in August 2012, August 25-29th, again in Munich, with a joint ESC-ISACHD session.

News for Asia-Pacific:
Koichiro Niwa

-14th Congress of Japanese Society for Adult CHD (JSACHD) will be held January 14-15, 2012, in Tokyo (http://www.jsachd.org). There are 31 orals, 36 posters, 5 symposia, and special talks (Joseph K Perloff MD, Philip Steer MD, Ad Boger MD, Eun Jun Bae MD, etc). The concept of the meeting is establishment of ACHD facility in Japan.
-The 3rd Congress of APSACHD (Asia Pacific Society for ACHD) will be held April 7th, 2012, in Taipei combined with APPCS (Asia Pacific Cardiac Society).

News from Canada

Erwin Oechslin

- I would like to invite you to participate in the **22nd Annual International Symposium on Adult Congenital Heart Disease in Toronto.** This Symposium reflects a long partnership between the faculties of Oregon Health Sciences University, Cincinnati Children's Hospital Medical Centre/University of Cincinnati, and University of Toronto. It is a unique forum where medical professionals can meet their colleagues from around the world and exchange ideas and information in the growing field of Adult Congenital Heart Disease (ACHD). The meeting will be held at Toronto Marriott Downtown close to the famous Eaton Centre **May 30 to June 2, 2012.**
The following **hot topics** will be discussed: *the Fontan disease*--diagnostic and therapeutic challenges of a systemic disease, *end of life and supportive care* (advanced care)--an ignored challenge and urgent need in our growing population, *pulmonary hypertension* in congenital heart disease, *arrhythmias, catheter-based interventions, new techniques in imagin, heart disease and pregnancy,* amongst others.
Special Registration Fee: Paying ISACHD and or CACH Network members will be offered a 10% reduction on the registration fee. The website www.uhn.ca/ACHHDConference2012.asp will open next week.

-**CACH Network News**
Please visit www.cachnet.ca for CACH Network News and visit the President's message page.

Journal Watch

ARTICLE OF THE MONTH: January 2012
Commentary on paper 'Fontan conversion with concomitant arrhythmia surgery for the failing atriopulmonary connections: mid-term results from a single centre' by Luke Burchill.

Atriopulmonary (AP) to total cavopulmonary extracardiac Fontan conversion with arrhythmia surgery is a treatment option for patients with an AP Fontan and impaired functional status secondary to refractory atrial arrhythmias, right atrial enlargement and/or Fontan pathway obstruction [1-4]. Heart transplantation is increasingly common among patients with failing Fontan physiology but unfortunately wait list and early operative mortality remains high. By reinstating a low-pressure, unobstructed venous circuit, Fontan conversion offers an 'intermediate' palliative treatment strategy that can reduce arrhythmia burden and improve functional class, particularly when combined with arrhythmia surgery and valve repair/replacement. Patient specific risk factors for increased mortality after Fontan conversion have been previously reported and include protein losing enteropathy, significant AV valve regurgitation and a ventricle of right ventricular or ambiguous morphology [3]. Whether Fontan conversion is superior to continuing medical treatment is not known.

Two recent articles provide important insights into clinical outcomes after Fontan conversion. The first, by Dr Carl Backer, summarises the Chicago experience and reports clinical outcomes in patients who underwent Fontan conversion, comprising bidirectional superior cavopulmonary anastomosis, right or biatrial maze procedure, and epicardial dual-chamber antitachycardia pacemaker insertion [5].

Comprising the largest series to date, the Chicago group report outcomes after Fontan conversion in 133 patients between 1994 and 2011. Mean age at operation was 23.3 years +/- 8 yrs and the mean interval from patients' original Fontan operation to conversion was 16.3 +/- 5.4 yrs. The main advantages of Fontan conversion were freedom from arrhythmia recurrence (85% at 10 years) and improved NYHA functional class in the majority of patients. Given the low operative mortality of 1.5% and the substantial improvements in arrhythmia burden and functional class reported by the Chicago group, many centres now recommend Fontan conversion to patients with an AP Fontan complicated by symptomatic atrial arrhythmia.

By comparison, the study by Sridhar et al reports medium-term outcomes in a small cohort of 15 patients who had Fontan conversion in a single Italian centre between 2002-2009 [5]. Similar to the Chicago group, the median age of Fontan conversion patients was 26.2 years and the median interval from the original Fontan operation to conversion was 16 years. The reduction in arrhythmia burden after Fontan conversion also mirrored the Chicago experience with 85% being arrhythmias free 53 months post conversion. Despite these similarities, there was a notable difference in mortality between the two studies. Compared to a operative mortality of 1.5% in the Chicago cohort, Sridhar et al report a mortality rate of 13.3%. One death resulted from intractable atrial arrhythmia precipitating low output cardiac / multiorgan failure while a second death occurred secondary to fatal hematemesis from esophageal varices. Both deaths occurred in patients who had pre-operative arrhythmias for more than 10 years, although the cohort was too small to assess for any statistical association between arrhythmia duration and operative mortality. Further studies are required to confirm this potential association.

The significant differences in operative mortality after Fontan conversion are consistent with prior reports. It is expected that operative mortality will fall as the experience with Fontan conversion grows. Determining candidacy for Fontan conversion requires ACHD clinicians to weigh up the potential benefits and risks according to the anticipated benefits of surgery, the presence of risk factors known to increase operative mortality, and institiutional outcomes.

1. Mavroudis C, Backer CL, Deal BJ, Johnsrude C, Strasburger J. Total cavopulmonary conversion and maze procedure for patients with failure of the Fontan operation. J Thorac Cardiovasc Surg, 122(5), 863-871 (2001).
2. Mavroudis C, Backer CL, Deal BJ, Johnsrude CL. Fontan conversion to cavopulmonary connection and arrhythmia circuit cryoblation. J Thorac Cardiovasc Surg, 115(3), 547-556 (1998).
3. Mavroudis C, Deal BJ, Backer CL et al. J. Maxwell Chamberlain Memorial Paper for congenital heart surgery. 111 Fontan conversions with arrhythmia surgery: surgical lessons and outcomes. Ann Thorac Surg, 84(5), 1457-1465; discussion 1465-1456 (2007).

4. Morales DL, Dibardino DJ, Braud BE et al. Salvaging the failing Fontan: lateral tunnel versus extracardiac conduit. Ann Thorac Surg, 80(4), 1445-1451; discussion 1451-1442 (2005).
5. Backer CL. 12th Annual C. Walton Lillehei Memorial Lecture in Cardiovascular Surgery: Fontan conversion - the Chicago experience. Cardiol Young, 21 Suppl 2, 169-176 (2011).

Commentary:
Luke Burchill, Fellow Adult Congenital Heart Disease
Toronto General Hospital, Divison of Cardiology
Toronto, Ontario, Canada

Cardiol Young. 2011 Dec;21(6):665-9. Epub 2011 May 27.
Fontan conversion with concomitant arrhythmia surgery for the failing atriopulmonary connections: mid-term results from a single centre.
Sridhar A, Giamberti A, Foresti S, Cappato R, García CR, Cabrera ND, Micheletti A, Negura D, Bussadori C, Butera G, Frigiola A, Carminati M, Chessa M.
Source
Department of Pediatric Cardiology & Adult with Congenital Heart Disease, IRCCS-Policlinico San Donato, Cardiovascular Center 'E. Malan', University of Milan, Italy.
Abstract
Objectives: Classical Atriopulmonary Fontan connections tend to fail in the long term due to progressive anastomotic site obstruction, right atrial enlargement, and refractory atrial arrhythmias. Conversion to total cavopulmonary connection with concomitant arrhythmia surgery is a promising treatment but optimal timing of the procedure remains controversial.
Methods: Between the years 2002 and 2009, 15 patients with a median age of 26.2 (12-43) years underwent Fontan conversion operation with concomitant arrhythmia surgery. All were symptomatic and 14 out of the 15 patients had refractory arrhythmias. The duration of pre-operative arrhythmia and the outcome of surgery were correlated to study the impact of delay in surgical intervention on post-operative survival and arrhythmia control.
Results: There were two patients who died in the early post-operative period (13.3%). At the mid-term follow-up, 53 (20-86) months, late atrial arrhythmias had recurred in two of the 13 surviving patients (15.30%) and one patient developed late sinus node dysfunction. The need for anti-arrhythmic drugs decreased considerably from 93.5% to 15.3% on mid-term follow-up. There was no late death or need for cardiac transplantation. The duration of arrhythmia before surgery was prolonged for more than 10 years in patients who died as well as in those who had complications like late recurrence of arrhythmias, dependence on anti-arrhythmic medications, and worsening of ventricular dysfunction.
Conclusions: Fontan conversion is a well-established treatment option for salvaging the failing atriopulmonary connections. Concomitant arrhythmia surgery effectively resolves the refractory atrial arrhythmias

and improves survival, but we need to optimise the timing of Fontan conversion to improve the long-term outcome.

Intern Med J. 2011 Dec 29. doi: 10.1111/j.1445-5994.2011.02708.x. [Epub ahead of print]

Congenital Heart Disease Associated Pulmonary Arterial Hypertension: Preliminary Results From a Novel Registry.

Rose M, Strange G, King I, Arnup S, Vidmar S, Kermeen F, Grigg L, Weintraub R, Celermajer D.

Source

Royal Children's Hospital, Melbourne, Australia; Murdoch Children's Research Institute, Melbourne, Australia; Department of Cardiology, Royal Prince Alfred Hospital, Sydney, Australia; Department of Epidemiology and Preventative Medicine, Monash University, Melbourne, Australia; Department of Paediatrics, University of Melbourne, Melbourne, Australia; Pulmonary Hypertension and Transplant Unit, The Prince Charles Hospital, Brisbane, Australia; Department of Cardiology, Royal Melbourne Hospital, Melbourne, Australia; Department of Medicine, The University of Sydney, Sydney, Australia.

Abstract

Objectives: Pulmonary arterial hypertension (PAH) frequently accompanies childhood congenital heart disease (CHD) and may persist into adult life. The advent of specific PAH therapies for PAH prompted formation of a national ANZ registry in 2010 to document the incidence, demographics, presentation and outcomes for these patients. **Design:** This multicentre, prospective, web-based registry enrolls patients with CHD associated PAH being followed in a tertiary centre. The inclusion criteria stipulated patient age >16 years, a measured mPAP >25mmHg at rest or echocardiographic evidence of PAH or a diagnosis of Eisenmenger syndrome, and followed since 1/1/2000. A single observer collected standardised data during a series of site visits. **Results:** Of the first 50 patients enrolled, 30 (60%) are female The mean age (SD) at the time of PAH diagnosis or confirmation in an adult centre was 27.23 (10.07) years and 32 (64%) patients are currently aged >30 years. 14 (28%) patients were in WHO functional Class II and 36 (72%) in Class III at the time of diagnosis. 47 of 50 (94%) had congenital systemic-pulmonary shunts and 36 (72%) never underwent intervention. 13 (26%) had Down's syndrome. Confirmation of PAH by recent cardiac catheterization was available in 30 (60%) subjects. During follow-up a total of 32 (64%) patients received a PAH specific therapy. **Conclusions:** CHD associated with PAH in adult life has resulted in a new population with unique needs. This registry will allow documentation of clinical course and long-term outcomes for these patients.

Heart Vessels. 2011 Dec 28. [Epub ahead of print]

Comparison of stroke work between repaired tetralogy of Fallot and normal right ventricular physiologies.

Lee N, Das A, Banerjee RK, Gottliebson WM.

Source

Mechanical Engineering, School of Dynamics Systems, University of Cincinnati, 593 Rhodes Hall, ML 0072, Cincinnati, OH, 45221, USA.
Abstract
Adult patients who underwent tetralogy of Fallot repair surgery (rTOF) confront life-threatening ailments due to right ventricular (RV) myocardial dysfunction. Pulmonary valve replacement (PVR) needs to be performed to restore the deteriorating RV function. Determination of correct timing to perform PVR in an rTOF patient remains subjective, due to the unavailability of quantifiable clinical diagnostic parameters. The objective of this study is to evaluate the possibility of using RV body surface area (BSA)-indexed stroke work (SW(I)) to quantify RV inefficiency in TOF patients. We hypothesized that RV SW(I) required to push blood to the lungs in rTOF patients is significantly higher than that of normal subjects. Seven patients with rTOF pathophysiology and eight controls with normal RV physiology were registered for this study. Right ventricular volume and pressure were measured using cardiac magnetic resonance imaging and catheterization, respectively. Statistical analysis was performed to quantify the difference in SW(I) between the RV of the rTOF and control groups. Right ventricular SW(I) in rTOF patients (0.176 ± 0.055 J/m(2)) was significantly higher by 93.4% (P = 0.0026) than that of controls (0.091 ± 0.030 J/m(2)). Further, rTOF patients were found to have significantly higher (P < 0.05) BSA normalized RV end-systolic volume, end-systolic pressure, and regurgitation fraction than control subjects. Ejection fraction and peak ejection rate of rTOF patients were significantly lower (P < 0.05) than those of controls. Patients with rTOF pathophysiology had significantly higher RV SW(I) compared with subjects with normal RV physiology. Therefore, RV SW(I) may be useful to quantify RV inefficiency in rTOF patients along with currently used clinical end points such as RV volume, pressure, regurgitation fraction, and ejection fraction.

Eur Heart J. 2011 Dec 23. [Epub ahead of print]
Reference values for exercise limitations among adults with congenital heart disease. Relation to activities of daily life--single centre experience and review of published data.
Kempny A, Dimopoulos K, Uebing A, Moceri P, Swan L, Gatzoulis MA, Diller GP.
Source
Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital, Sydney Street, SW3 6NP London, UK.
Abstract
Aims: We aimed to investigate the distribution of exercise capacity across the spectrum of adult congenital heart disease (ACHD) using own data and the published experience and to provide diagnosis, gender-, and age- specific reference values.
Methods and results: Publications describing exercise capacity in ACHD patients using cardiopulmonary exercise testing (CPET) were identified (n = 2286 patients in 23 papers). In addition, we included 2129 patients who underwent CPET at our own institution. The majority of patients (80%) had reduced peak oxygen uptake (peak VO(2))

compared with normal values (defined as <90% of predicted peak VO(2)). There were significant differences in peak VO(2) between subgroups of patients, with the lowest values seen in patients with Eisenmenger syndrome and complex heart disease. However, even in patients with simple lesions, peak VO(2) was on average significantly reduced compared with normal values. Based on a large number of observations we herewith provide gender- and age-specific peak VO(2) centile plots for the most common lesions (Tetralogy of Fallot, systemic right ventricle, Ebstein anomaly and Fontan-palliation) and relate disease-specific exercise capacity to that required for specific activities of daily life, sports, and occupations.
Conclusion: We provide age-, gender-, and diagnosis-specific data on peak VO(2) levels across the spectrum of ACHD allowing to compare the exercise capacity of individual patients with that of their peer patients. These data should be helpful in interpreting CPET results, guiding therapy, and advising patients on activities of daily life, sports participation, and choice of occupation.

Congenit Heart Dis. 2011 Dec 21. doi: 10.1111/j.1747-0803.2011.00606.x. [Epub ahead of print]
Efficacy and Safety of Bosentan in Adults with Simple and Complex Eisenmenger's Syndrome.
Williams R, Houser L, Miner P, Aboulhosn J.
Source
Ahmanson/UCLA Adult Congenital Heart Disease Center, David Geffen School of Medicine, Los Angeles, Calif, USA.
Abstract
Background: Eisenmenger's syndrome (ES) is associated with decreased longevity and reduced functional capacity. Targeted pharmacologic therapies improve functional capacity and survival in these patients. We sought to compare the response of patients with simple vs. complex ES following initiation of bosentan.
Methods: ES patients with a history of bosentan use were identified by chart review. Simple ES was defined as ES associated with atrial septal defect, ventricular septal defect, or patent ductus arteriosus. Complex ES consisted of patients with truncus arteriosus and single ventricle congenital heart disease. Six-minute walking distance (6MWD), maximal oxygen consumption (VO(2) max), brain natriuretic peptide (BNP), and resting oxygen saturation were compared between simple and complex ES patients before and after bosentan treatment.
Results: Twenty-four patients were included (11 simple, 13 complex). Resting oxygen saturation, 6MWD, VO(2) max, and BNP were not significantly different between the two groups prior to bosentan initiation. Ten patients received bosentan monotherapy, and bosentan was used in combination with sildenafil in 13 (five simple, eight complex). One patient received bosentan with iloprost. Mean duration of therapy was 38±14 months in the simple group and 40±8.1 months in the complex group (P= NS). Posttreatment, 6MWD increased from 274±135m to 326±106m in simple ES patients (P= .32). 6MWD in patients with complex ES increased from 332±51m to 364±109 (P= .028). VO(2) max

improved from 13.4±3.8 to 17±6 (P= .54) in the simple group, while VO(2) max in the complex group improved from 12.7±2.3 to 15.5±2.2 (P= .17). There was minimal change in BNP or resting oxygen saturation between the groups.
Conclusions: Treatment with bosentan is both safe and effective in patients with both simple and complex forms of ES.

Cardiol Rev. 2011 Dec 16. [Epub ahead of print]
Cardiac Manifestations of Neonatal Lupus: A Review of Autoantibody Associated Congenital Heart Block and its Impact in an Adult Population.
Capone C, Buyon JP, Friedman DM, Frishman WH.
Source
Dept. of Pediatrics, Children's Hospital of Montefiore, Bronx, NY **Dept. of Medicine, Div. of Rheumatology, New York University Medical Center, NY Dept. of Pediatrics, Div. of Pediatric Cardiology, New York Medical College, Valhalla, NY §Dept. of Medicine, New York Medical College/Westchester Medical Center, Valhalla, NY.
Abstract
Within the last decade the prevalence of adult patients living with congenital heart disease equals that seen in children. This expanding population poses a challenge to clinical cardiologists who will be caring for patients with the clinical manifestations of this condition. Neonatal lupus is a model of passively acquired auto-immunity and is responsible for the majority of clinical cases of congenital heart block. This review will focus on the presentation, pathophysiology, and the long-term follow up of congenital heart block associated with neonatal lupus, as well as discuss important diagnostic tests, familial implications, and pacemaker issues associated with the care of an adult with congenital heart block.

Cardiol Young. 2011 Dec 14:1-7. [Epub ahead of print]
Tetralogy of Fallot in men: quality of life, family, education, and employment.
Bygstad E, Pedersen LC, Pedersen TA, Hjortdal VE.
Source
Department of Cardiothoracic and Vascular Surgery, Aarhus University Hospital, Aarhus, Denmark.
Abstract
Introduction: Little is known about the quality of life, health, family, education, and employment status among adult men with repaired tetralogy of Fallot.
Material and methods: A total of 68 men who underwent repair of tetralogy of Fallot between 1971 and 1991 were studied. Fifty-three patients answered the SF-36 health survey and additional questions regarding offspring, education, and employment status. The men with repaired tetralogy of Fallot were compared with 32 healthy men and 40 women who also underwent repair of tetralogy of Fallot in the same period.
Results: The patients scored lower than healthy men in the SF-36 categories physical functioning, general health, and physical component

summary. There were no statistically significant differences in the scores from male and female patients except a lower score in bodily pain among women. Educational level for men operated for tetralogy of Fallot was similar to the general male population, whereas fewer were employed and more were retired, undergoing rehabilitation or receiving social benefits. The reproduction rate was lower compared with the general population (0.65 versus 1.02 children per man) but relatively higher than the rate among women with tetralogy of Fallot (0.88 versus 1.84 children per woman). The risk of having a child with congenital heart disease was 8.3%.
Conclusion: Men operated for tetralogy of Fallot have good quality of life and educational status. They start a family, although their reproduction rate is two-thirds that of the general population. The risk of having a child with congenital heart disease is higher compared with the background population. The overall quality of life is similar for men and women operated for tetralogy of Fallot.

Am J Med Genet A. 2011 Dec 7. doi: 10.1002/ajmg.a.34392. [Epub ahead of print]
A comparison of the Ghent and revised Ghent nosologies for the diagnosis of Marfan syndrome in an adult Korean population.
Yang JH, Han H, Jang SY, Moon JR, Sung K, Chung TY, Lee HJ, Ki CS, Kim DK.
Source
Department of Medicine, Cardiac and Vascular Center, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea.
Abstract
Recently, a revised Ghent nosology has been established for the diagnosis of Marfan syndrome (MFS) that puts more weight on the aortic root aneurysm and ectopia lentis. We compared the application of the Ghent and revised Ghent nosologies in adult Korean patients for whom there is suspicion of MFS. From January 1995 to June 2010, we enrolled 106 patients older than 20 years for whom there was suspicion of MFS, and who had undergone genetic analysis of the fibrillin-1 gene (FBN1). Of 106 patients, 86 patients (81%) fulfilled the criteria of the Ghent nosology, and 84 patients (79%) met the criteria of the revised Ghent nosology. The two patients who met the Ghent nosology criteria, but not the criteria of the revised Ghent nosology were diagnosed with Loeys-Dietz syndrome and MASS phenotype. The level of agreement between both nosologies was very high (K=0.94, 95% confidence interval: 0.86 to 1.0). Marfan-like syndromes were diagnosed in 30% (6/20 patients) with negative Ghent and revised Ghent criteria and no FBN1 mutations. These results suggest that adult Korean patients who fulfill the old Ghent criteria almost all fulfill the new criteria for the diagnosis of MFS. © 2011 Wiley Periodicals, Inc.

Int J Cardiol. 2011 Dec 8. [Epub ahead of print]
Dobutamine stress MRI in repaired tetralogy of Fallot with chronic pulmonary regurgitation A comparison with healthy volunteers.Parish V,

Valverde I, Kutty S, Head C, Qureshi SA, Sarikouch S, Greil G, Schaeffter T, Razavi R, Beerbaum P.
Source
King's College London BHF Centre, Division of Imaging Sciences, NIHR Biomedical Research Centre at Guy's & St Thomas' NHS Foundation Trust, UK; Adult Congenital Heart Disease Service, Guy's & St Thomas' NHS Foundation Trust, London, UK.
Abstract
Background: To compare the ventricular response to dobutamine stress between adult patients with chronic pulmonary regurgitation (PR) after repair of tetralogy of Fallot (r-TOF) and healthy volunteers using a staged dobutamine stress MR (DS-MR) protocol.
Methods: Eighteen r-TOF patients (median age 31.9years, range 16.2-60.1) with severe PR and 10 healthy controls (median age 40.6years, range 23.9-51.8) completed staged DS-MR (baseline, 10 and 20µg/kg/min) with ventricular volumetry and pulmonary flow quantification. Comparative analysis involved 3-way ANOVA, t-test, regression analysis, and coefficient of variance.
Results: All controls had significant increase of ejection fraction (EF) at each stress level for both ventricles (normal contractile reserve, all $p<0.05$). In r-TOF patients (RV-EDV 126±27ml/m(2), RV-EF 55±7%, LV-EF 58±6%, PR-fraction 43±15%), low-dose DS-MR at 10µg/kg/min demonstrated normal biventricular contractile reserve as seen in volunteers. On increase from 10 to 20µg/kg/min a subgroup showed worsening ejection fraction ($n=8$, $p<0.05$), mainly due to lack of reduction or even increase of RV-ESV, while the remainder responded with further reduction of RV-ESV and RV-EDV ($n=10$, $p<0.05$) and a non-significant trend to increased EF. This different response could not be predicted at baseline.
Conclusions: In r-TOF patients with chronic PR, DS-MR at 10µg/kg/min showed normal biventricular systolic response compared with controls. Increase to 20µg/kg/min provoked abnormal RV-ESV response in some r-TOF patients, suggesting presence of ventricular systolic dysfunction not evident at rest.

Arch Cardiovasc Dis. 2011 Dec;104(12):627-35. Epub 2011 Nov 21.
Intravascular stenting for the treatment of coarctation of the aorta in adolescent and adult patients.
Godart F.
Source
Faculté de médecine, université de Lille Nord-de-France, IFR114, EA 2693, Lille, France; Service de cardiologie infantile et congénitale, hôpital cardiologique, CHRU de Lille, boulevard du Professeur-Leclercq, 59037 Lille cedex, France.
Abstract
In the past 10 years, stent implantation has become a real alternative to surgery in the management of both native and recurrent coarctation of the aorta in adolescents and adults. The purpose of this report is to provide a detailed review of stent implantation techniques, including pre-

procedure imaging, technical aspects and results. The success rate is usually high (around 90%), and the procedure results in an increase in the diameter of the coarcted segment, a decrease in the transisthmic systolic gradient and a better control of systemic hypertension. The most serious complication, rupture of the aorta, can be fatal, but is rare (< 2%). Aneurismal dilatation is another potential problem that occurs in around 5-9% of cases, and may be related to overstretching and pre-stent dilatation, so these should be avoided. It is necessary to point out that most of these aneurysms are small and conservatively managed. Restenosis is another complication that may result from neointimal proliferation, stent recoil and stent fracture. Balloon dilatation with a higher inflating pressure or repeat stent implantation is proposed herein. A comparison with surgery is also discussed, and a follow-up protocol is proposed to capture late complications. Owing to good initial and intermediate results, stent implantation is nowadays considered as a first-line therapy in most adolescents and adults with (re)coarctation of the aorta.

J Cardiovasc Magn Reson. 2011 Dec 8;13(1):78. [Epub ahead of print]
3D Echo systematically underestimates right ventricular volumes compared to cardiovascular magnetic resonance in adult congenital heart disease patients with moderate or severe RV dilatation.
Crean AM, Maredia N, Ballard G, Menezes R, Wharton G, Forster J, Greenwood JP, Thomson JD.

Abstract
Background: Three dimensional echo is a relatively new technique which may offer a rapid alternative for the examination of the right heart. However its role in patients with non-standard ventricular size or anatomy is unclear. This study compared volumetric measurements of the right ventricle in 25 patients with adult congenital heart disease using both cardiac magnetic resonance (CMR) and three dimensional echocardiography.
Methods: Patients were grouped by diagnosis into those expected to have normal or near-normal RV size (patients with repaired coarctation of the aorta) and patients expected to have moderate or worse RV enlargement (patients with repaired tetralogy of Fallot or transposition of the great arteries). Right ventricular end diastolic volume, end systolic volume and ejection fraction were compared using both methods with CMR regarded as the reference standard
Results: Bland-Altman analysis of the 25 patients demonstrated that for both RV EDV and RV ESV, there was a significant and systematic under-estimation of volume by 3D echo compared to CMR. This bias led to a mean underestimation of RV EDV by -34% (95%CI: -91% to + 23%). The degree of underestimation was more marked for RV ESV with a bias of -42% (95%CI: -117% to + 32%). There was also a tendency to overestimate RV EF by 3D echo with a bias of approximately 13% (95% CI -52% to +27%).
Conclusions: Statistically significant and clinically meaningful differences in volumetric measurements were observed between the two techniques. At the current time, three dimensional echocardiography is

not interchangeable with CMR for volumetric assessment of ACHD patients who have more than mild RV dilatation.

Int J Cardiol. 2011 Nov 30. [Epub ahead of print]
Cardiothoracic ratio from postero-anterior chest radiographs: A simple, reproducible and independent marker of disease severity and outcome in adults with congenital heart disease. Dimopoulos K, Giannakoulas G, Bendayan I, Liodakis E, Petracco R, Diller GP, Piepoli MF, Swan L, Mullen M, Best N, Poole-Wilson PA, Francis DP, Rubens MB, Gatzoulis MA.

Source

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

Abstract

Objective: The wide spectrum of intracardiac anatomy and reparative surgery available for adults with congenital heart disease (ACHD) makes uniform measurement of cardiac size and disease severity challenging. The aim of this study was to assess the prognostic potential of cardiothoracic ratio, a simple marker of cardiomegaly, in a large cohort of ACHD.

Patients and Setting: Chest radiographs from 3033 ACHD patients attending our institution between 1998 and 2007 and 113 normal controls of similar age were analyzed blindly.

Design: Cardiothoracic ratio derived from plain postero-anterior chest radiographs, was compared between ACHD patients and controls, different diagnostic subgroups and different functional classes. Relationship between cardiothoracic ratio and survival was assessed using Cox regression.

Results: Average cardiothoracic ratio in ACHD was 52.0±7.6% (over 50% in 56.4%), significantly higher in all ACHD diagnostic subgroups compared to controls (42.3±4.0%, $p<0.0001$) and highest in the "complex" cardiac anatomy, Ebstein's anomaly and Eisenmenger subgroups. Cardiothoracic ratio related to functional class, but was high even in asymptomatic patients. During a median follow-up of 4.2years, 164 patients died. Patients with a cardiothoracic ratio >55% had an 8-fold increased risk of death compared to those in the lowest tertile (<48%). Even patients with mildly increased cardiothoracic ratio (48-55%) had an adjusted 3.6-fold increased mortality compared to the lowest tertile.

Conclusions: Cardiothoracic ratio derived from postero-anterior chest radiographs is a simple, and reproducible marker, which relates to functional class and predicts independently mortality risk in ACHD patients.

Pediatrics. 2011 Dec;128(6):e1489-e1495. Epub 2011 Nov 28.
Parental Knowledge Regarding Lifelong Congenital Cardiac Care.
Fernandes SM, Verstappen A, Ackerman K, Adams EE, Barton C, Breitinger P, Crumb S, Dummer K, Harada K, Khairy P, Landzberg MJ,

Linstead-Goldsmith R, Meadows AK, Nieves JA, Saidi A, Takahashi M, Zhou J, Ziniel S, Williams R; on behalf of the Adult Congenital Heart Association; the Adult Congenital Cardiac Care Associate Research Network.

Source

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Abstract

Objective: To assess parental knowledge regarding lifelong congenital cardiac care (LLCCC).

Background: National guidelines recommend that nearly 50% of adult survivors with congenital heart disease (CHD) receive LLCCC; the number of adults who receive such care seems far less. Inadequate parental knowledge of LLCCC might contribute to care interruption.

Methods: In this multicenter study, we administered a questionnaire to parents of children with moderate and complex CHD to assess knowledge of LLCCC.

Results: A total of 500 parents participated; the median age of their children was 10 years (range: 2-18 years). Most parents (81%) understood that their child would need LLCCC, but only 44% recognized that their child's cardiology care should be guided by an adult congenital heart specialist in adulthood. More than half (59%) of the parents stated that their current cardiology team had never spoken to them about LLCCC, but 96% wished to learn more. Variables associated with parental LLCCC knowledge included previous discussions regarding LLCCC, underlying cardiac surgical diagnosis, and level of parental education.

Conclusions: A substantial number of parents of children with moderate and complex CHD lack knowledge about LLCCC, but almost all of them have a desire to learn more about the care their child will need as an adult.

Expert Rev Cardiovasc Ther. 2011 Dec;9(12):1547-56.

Pregnancy in women with Fontan physiology.

Le Gloan L, Mercier LA, Dore A, Marcotte F, Mongeon FP, Ibrahim R, Asgar A, Poirier N, Khairy P.

Source

Adult Congenital Heart Center, Montreal Heart Institute, 5000 Belanger St E, Montreal, QC, H1T 1C8, Canada.

Abstract

Advances in the care of patients with a univentricular heart have enabled the majority to survive well into adulthood. Consequently, an increasing number of women with Fontan palliation are contemplating pregnancy. This review summarizes physiologic modifications related to pregnancy in the setting of single-ventricle physiology, discusses potential maternal cardiovascular concerns and describes commonly encountered obstetrical and neonatal complications. Management issues are addressed, including counseling, anesthetic considerations

and the need for multidisciplinary specialized care.

J Cardiovasc Surg (Torino). 2011 Dec;52(6):873-6.
Hypertension in adult after operation of aortic coarctation.
Palma G, Giordano R, Russolillo V, Cioffi S, Palumbo S, Mucerino M, Poli V, Vosa C.

Source

Department of Clinical Medicine and Cardiovascular Sciences, University of Naples Federico II, Naples, Italy.

Abstract

Aim: The benefit of coarctation repair on the resolution of systolic hypertension in adults has been questioned.
Methods: Between March 1997 and July 2009, 65 consecutive adult patients (≥ 16 years) underwent repair of aortic coarctation. There were 40 men (65%) and 25 women (35%) with a mean age of 22.3 ± 4.8 years (range, 16 to 34 years). All patients had critical systolic blood hypertension (SBP ≥ 140 mmHg). SBP ranged from 140 to 205 mmHg, with a mean of 163.5 ± 17.6 mmHg. The mean diastolic BP was 95.1 ± 18.3 mmHg (range, 70 to 120 mmHg). Most patients (41/65, 74%) were on a regimen of at least one antihypertensive drug.
Results: The patients were followed up after coarctation repair for 2 to 144 months (mean, 68 ± 39 months). There was no death. No other major complications occurred. There have been no repeat interventions during follow-up. Four patients were lost to follow-up. Of the 61 patients with preoperative hypertension, 53 (87%) were normotensive (SBP <140 mmHg) at the most recent follow-up visit. The remaining eight patients showed substantial improvement versus the preoperative status. The mean SBP after operation was 122.5 ± 12.4 mmHg. Mean diastolic blood pressure was 79.5 ± 11.6 mmHg. Forty-one (67%) patients were taking no medication at the last follow-up.
Conclusion: Surgical repair of coarctation of the aorta in adults can lead to regression of systolic hypertension and a decreased requirement for antihypertensive medication.

J Am Soc Echocardiogr. 2011 Dec;24(12):1392-1399.e1. Epub 2011 Oct 14.
Biventricular Performance in Patients with Marfan Syndrome without Significant Valvular Disease: Comparison to Normal Subjects and Longitudinal Follow-Up.
Scherptong RW, Vliegen HW, van der Wall EE, Hilhorst-Hofstee Y, Bax JJ, Scholte AJ, Delgado V.

Source

Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands.

Abstract

Background: The presence and progressive nature of primary myocardial involvement in Marfan syndrome are debated. The aim of this study was to evaluate the clinical relevance of left ventricular (LV) and right ventricular (RV) strain in adult patients with Marfan syndrome without significant valvular disease.

Methods: Adult patients with Marfan syndrome (n = 50; mean age, 35.2 ± 12.9 years) were followed prospectively. Echocardiography was performed annually and consisted of comprehensive assessment of ventricular and valvular function. Using speckle-tracking imaging, the baseline strain values of the Marfan population were calculated and compared with the values of normal controls. The follow-up evaluations were used to assess changes in ventricular strain. The association between the incidence of adverse events (heart failure, [supra]ventricular arrhythmias, and proximal aorta surgery) and baseline strain values was investigated.
Results: Compared with controls, patients with Marfan syndrome had significantly lower peak longitudinal LV strain (-18.9 ± 2.3% vs -20.1 ± 1.9%, P < .01) and RV strain (±26.9 ± 5.2% vs ±29.3 ± 4.25%, P < .01). The absolute changes in LV longitudinal, radial, and circumferential strain and RV longitudinal strain during a median 4 years of follow-up were 0.1 ± 2.8%, 1.12 ± 7.6%, 0.3 ± 3.7%, and 0.9 ± 5.5%, respectively, which was not statistically significant. Cox regression demonstrated that reduced LV or RV strain was not associated with adverse outcome (supraventricular arrhythmias, n = 3; proximal aorta surgery, n = 4).
Conclusions: This study suggests that patients with Marfan syndrome show lower ventricular strain and strain rate values compared with healthy controls. However, no relevant changes in LV and RV function occurred during midterm follow-up in patients with Marfan syndrome without valvular disease at baseline. Although ventricular strain and strain rate were mildly reduced in patients with Marfan syndrome, this did not affect outcomes negatively in the present study.

Pacing Clin Electrophysiol. 2011 Dec;34(12):1621-7. doi: 10.1111/j.1540-8159.2011.03226.x. Epub 2011 Sep 28.
Pediatric and Adult Congenital Endocardial Lead Extraction or Abandonment Decision (PACELEAD) Survey of Lead Management.
McCanta AC, Schaffer MS, Collins KK.
Source
Department of Pediatric Cardiology, The University of Colorado Denver School of Medicine, Children's Hospital Colorado, Aurora, Colorado Department of Pediatric Cardiology, Duke University Medical Center, Durham, North Carolina.
Abstract
Background: Nonfunctional, dysfunctional, recalled, or additional endocardial leads in pediatric and congenital heart disease patients pose significant challenges for management. There are no set standards for lead extraction in this patient population.
Methods: Physician members of the Pediatric and Adult Congenital Electrophysiology Society (PACES) were contacted via e-mail and invited to respond to a 33-question online Pediatric and Adult Congenital Endocardial Lead Extraction or Abandonment (PACELEAD) survey.
Results: Responses were received from 75 of 138 (54%) physician members of PACES. Institutional volumes of device placement (<25 devices/year for 51% of responders), patients with abandoned leads (<25 patients for 71%), and lead extractions (<10 extractions/year for

51% and no extractions for 29%) were low for the majority of responders. Personal experience with lead extraction was also minimal with 49% not performing the procedure and 39% with less than 40 leads extracted as primary operator. Most responders (54, 72%) refer their lead extractions to another practitioner or facility with more experience. Responders were more likely to recommend lead extraction (>70%) for class IIa indications such as bacteremia, chronic pain that is not medically manageable, and functional leads with ipsilateral venous occlusion. Lead abandonment was favored (>70%) for one class IIb indication, a functional lead that is not currently being used.
Conclusions: Optimal lead management is challenging in pediatric and congenital heart disease patients, and considerable variability of practice is reported in their care. Low institutional and personal volumes may account for this variability. (PACE 2011; 34:1621-1627).

Eur Heart J. 2011 Dec;32(24):3137-46. Epub 2011 Sep 4.
Acute pulmonary vasodilator response in paediatric and adult pulmonary arterial hypertension: occurrence and prognostic value when comparing three response criteria.
Douwes JM, van Loon RL, Hoendermis ES, Vonk-Noordegraaf A, Roofthooft MT, Talsma MD, Hillege HL, Berger RM.
Source
Center for Congenital Heart Diseases, Department of Paediatric Cardiology, Beatrix Children's Hospital, University Medical Center Groningen, University of Groningen, PO Box 30 001, 9700 RB Groningen, The Netherlands.
Abstract
Aims: To assess the occurrence and prognostic value of acute vasodilator response (AVR) in paediatric vs. adult pulmonary arterial hypertension, and idiopathic/hereditary pulmonary arterial hypertension (iPAH/HPAH) vs. pulmonary arterial hypertension associated with congenital heart disease (PAH-CHD) using three different response criteria.
Methods and results: Ninety-nine PAH patients underwent AVR testing (37 children, 62 adults; 70 iPAH/HPAH, 29 PAH-CHD). Three response criteria from clinical practice were used to define AVR. The number of responders was evaluated separately in subgroups based on age, diagnosis, and presence of a non-restrictive post-tricuspid shunt. Numbers of responders varied importantly using the different criteria but were always higher in iPAH/HPAH, compared with PAH-CHD. The number of responders did not differ between paediatric and adult iPAH/HPAH. No responders were identified in patients with a post-tricuspid shunt. Acute vasodilator response was associated with improved survival using all three criteria. Low baseline mean right atrial pressure (mRAP) was associated with improved survival in adults ($P < 0.001$). High baseline mean pulmonary arterial pressure (mPAP)/mean systemic arterial pressure (mSAP) and pulmonary vascular resistance (PVR)/systemic vascular resistance (SVR) were associated with worse survival, statistically independent from age, diagnosis, and the presence of a post-tricuspid shunt.

Conclusion: The proportion of patients with AVR highly depends on the used criteria, but did not differ between paediatric and adult iPAH/HPAH. Current response criteria are not suitable for patients with a post-tricuspid shunt. In both children and adults without post-tricuspid shunts, AVR was associated with improved survival independent of the used criteria. Nevertheless, prognostic value in the individual patient was limited. Baseline mRAP showed a good correlation with survival for adult PAH patients, but not for children. High baseline mPAP/mSAP and PVR/SVR was associated with worse prognosis independent from age, diagnosis, or the presence of a post-tricuspid shunt.

Cardiol Young. 2011 Dec;21(6):639-45. Epub 2011 May 24.
Adult congenital cardiac surgery in Indonesia.
Wilamarta KV, Yuniadi Y, Rachmat J, Fakhri D, Hakim T, Anwar M.
Source
Department of Cardiac Surgery, Faculty of Medicine, University of Indonesia, Jakarta, Indonesia.
Abstract
Background: Successful paediatric cardiac surgery and cardiology treatment has resulted in an increase in the use of surgery as a method of treatment of congenital cardiac disease in adult population. However, late detection and lower socio-economic condition in developing countries might change patients' characteristics by the time they come for treatment. This study aimed to elaborate the long-term surgical results of adult congenital cardiac disease in Indonesia as a developing country.
Methods and Results: We reviewed retrospectively all adult congenital cardiac disease patients with a mean age of 28 years plus or minus 9.5 years, who underwent surgery at National Cardiovascular Center. The types of procedures used were corrective in 338 patients (89.2%), palliative in 10 patients (2.6%), and re-operations in 31 patients (8.2%). The overall hospital mortality rate was 2.6% but as high as 20% with palliative surgery. Post-operative New York Heart Association class III-IV is the only independent predictor of death at 60 months (hazard ratio 61.48, 95% confidence interval 9.41-401.69, $p<0.001$). The survival rates were 96.3% and 95% for overall and non-atrial septal defect in patients at 60 months, which was highest in corrective procedures (97.6%). The percentage of patients free of re-operation at 5 years' follow-up was 85.4% and 42.7% at 10 years.
Conclusion: In developing countries, surgical treatment of adult congenital cardiac disease is effective and safe, with an overall survival rate of 96.3% at 60 months. Due to high mortality rate, palliative surgery of a non-atrial septal defect patient is recommended to be discontinued. The independent predictor of mortality was post-operative New York Heart Association functional class III-IV.