April, 2012

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International Society for Adult Congenital Heart Disease

ISACHD Newsletter

Presidents Message

by Curt J Daniels

Dear ISACHD Members,

Just a few weeks ago we met once again for our annual ISACHD meeting at ACC, this year in Chicago. The meeting held Sunday evening, March 26th, was well attended exceeding expectations--standing room only, and we even had to ask the hotel staff for extra chairs!

We were honored to see Dr. Dave Skorton join the meeting and provide insight as we presented our current ISACHD initiatives. Dr. Skorton is the former founder and director of ACHD at the University of Iowa, and a lead author of *Bethesda 32 Task Force 5 ACHD: Access to Care*. In 2006 he became president of Cornell University. We were pleased to see Dr. Skorton and look forward to his future collaboration with ISACHD.

During our meeting, the work group chairs presented an update of activities and invited comment. I presented an update on the Global Health initiative. We continue to gather information and will begin to look for international health partners to join with ISACHD members to build an ACHD global health plan. Erwin Oechslin presented an update on the ACHD Global Education program and along with the WG members is preparing web-based ACHD lectures. Erwin has also developed a successful model to provide ISACHD endorsement for international ACHD conferences. Several meetings are currently endorsed and advertised on the ISACHD website. ISACHD members receive a 10% discount for ISACHD endorsed meetings (Join ISACHD today!).

Koichiro Niwa updated the audience regarding ISACHD Global Research. Koichiro handed the research agenda to Philip Moons, who presented two exciting proposals for multi-center, multi-international research: "International Study on Lost To Care" and "International Study

Chiba, Japan

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Chiba, Japan

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Dr. Erwin N Oechslin
Toronto General Hospital
Toronto,
Ontario, Canada

on Quality of Life." There will be more to come from Philip regarding the proposed international studies.

We ending the evening by introducing the 2012 ISACHD Executive Committee highlighted by President -Elect, Paul Khairy, Montréal, QC, Canada. Paul will bring energy, enthusiasm, and a wealth of knowledge to ISACHD, and we look forward to Paul's contributions over the next several years.

We also look forward to seeing many ISACHD members in Dubai at the World Heart Federation World Congress of Cardiology, April 18-21, 2012;in Toronto for the 22nd International Symposium on ACHD, May 30- June 2, 2012; and in Munich for the ESC Congress 2012, August 25-29, 2012.

I am honored, excited, and humbled to serve as president of ISACHD and look forward to continuing the Mission with all of you as colleagues and members of ISACHD. A significant number of you as ISACHD members have indicated your desire to volunteer for the various ISACHD initiatives, and we look forward to providing opportunities to utilize your talents and expertise.

Curt J. Daniels
President

News from the Psychosocial Section

Adrienne Kovacs

The importance of a thoughtful and coordinated transition from pediatric to adult-focused cardiology care is well-recognized, although there are few formal educational opportunities in this area. We were therefore excited to learn that the University of Florida will offer an

Online Graduate Certificate in Education and Health Care Transition (EdHCT), beginning in Summer 2012. This certificate is designed for graduate students and professionals in medicine, nursing, education, social work, law, public health and public policy, physicians' assistants, and other health-related areas. The program allows participants to learn new skills for integrating education and health care transition and building expertise in this emerging discipline. The program consists of a total of 12 graduate credit hours that are completed online. The flexible schedule is ideal for busy practitioners, although the program can be completed in one year. For more information, please contact Arwa Saidi (ACHD cardiologist) at asaidi@ufl.edu or visit the website at http://education.ufl.edu/education-healthcare-transition.

Regional News:

News from Latin America

Luis Alday

During the next Argentine Congress of Cardiology to be held at the Buenos Aires Sheraton Hotel from 5 to 7 October 2012, a new ISACHD-SAC joint session will take place with Dr. Ariane Marelli, from Montreal, Canada, as the guest speaker. The Argentine Congress of Cardiology is usually a large meeting with attendance of nearly 10000 physicians from Argentina and neighbouring countries with guest speakers from all over the world.

News from Asia Pacific
Koichiro Niwa

The 3rd Congress of APSACHD Meeting was held on Saturday, April 7th, in Taipei, Taiwan. It was held combined with the 4th Congress of APPCS (Asia Pacific Pediatric Cardiology). The total attendance for APPCS and APSACHD was 700 individuals from 33 countries; the largest number of attendants were from Taiwan (130), followed by Japan (100), Korea (60), China (50), and others.

Various topics were presented and discussed. At the business meeting of APSACHD, 30 delegates from 14 countries were gathered. The governing body has been fixed (President: Koichiro Niwa MD; Executive committee: Jae-Kon Ko MD (Korea), Gu Hong MD (China), Geetha Kandavella MD (Malasia), Clare O'Drnell MD 8NZ), Ju-Le Tan MD (Singapore), Mei Hwan Wu MD (Taiwan), Jou-Kou Wang MD (Taiwan), Jun Huh MD (Korea), and Shigeru Tateno MD (Japan). A web page was also created (http://www.apsachd.org/).

The next Congress of APSACHD, 4th APSACHD Meeting, will be held on March 20-23, 2014, New Delhi, India. The program committee, research committee, and education committee have been assigned. APSACHD is taking big steps forward to a bright future.

News from Europe
Helmut Baumgartner

The 3rd European Meeting on Adult Congenital Heart Disease--the official annual European meeting of the ESC working group on Grown-Up Congenital Heart Disease--took place in Munich from March 16 to 17, 2012. Hosted by John Hess, Andeas Eicken, and Harald Kaemmerer in the German Heart Center Munich, this meeting was well-attended and very well-accepted. A distinguished faculty discussed a broard spectrum of adult congenital heart disease topics with a highly interested audience. The tradition will be continued next year in Milan, where along with the 9th International Workshop on Interventional Paediatric and Adult Congenital Cardiology, the 4th European Meeting on Adult Congenital Heart Disease is planned from March 20-23, 2013.

News from Canada
Erwin Oechslin

22nd ACHD Symposium in Toronto - May 30 to June 2, 2012:
ACHD Meeting Toronto2012 or
www.uhn.ca/ACHDConference2012.asp

Paying ISACHD/CACH Network Members Save Money!
Membership dues paying ISACHD and/or CACH Network members are offered a special registration fee until **April 15, 2012**.

Physicians	$765.00 CDN
Trainees, Nurses, Cardiac Care Associates	$585.00 CDN

Non-members early registration fee ends April 15, 2012

Physicians	$850.00 CDN
Trainees, Nurse, Cardiac Care Associates	$650.00 CDN

Hotel Reservation
The Symposium will be held at theToronto Marriott Downtown Eaton Centre Hotel
located in the heart of Downtown Toronto.
Special room rates are available until April 30, 2012: single and double room for only $189.00 CDN.

Online registration and hotel reservation is available at ACHD Meeting Toronto 2012 or www.uhn.ca/ACHDConference2012.asp. Please click on Location & Accommodation and follow instructions.

Hot Topics Are Discussed!
'Beyond Saving Lives' is the theme of the Symposium. Thousands of lives of children have been saved, but we are facing new challenges in the growing and aging congenital heart disease population. In addition to the common long-term complications, we are challenged with end of life questions and the provision of advanced/supportive care, which has been ignored and neglected in our young patients so far. A plenary session **(Beyond Saving Lives or Code 'Palliative': Beyond DNR, Quality of Death and Dying Experience)** and a Workshop **(Advanced Care Planning in Adult Congenital Heart Disease)** will cover this hot topic. Management of the failing Fontan circulation and heart failure are discussed amongst other challenging questions.

The program is available at ACHD Meeting Toronto 2012 or www.uhn.ca/ACHDConference2012.asp.

Accreditation and Endorsement
The symposium is an accredited group learning activity for Section 1

Credits as defined by the Royal College of Physicians and Surgeons of Canada and approved by the Canadian Cardiovascular Society for a maximum of 27 credits. The symposium is endorsed by ISACHD, APSACHD, and CACH Network.

Sixty-six (66!) Abstracts Have Been Submitted
Abstract submission has been a great success. The top three abstracts will be presented as oral presentations on May 31 and June 1, 2012. **The Winners of the best abstracts in the Nursing and Physician section** will receive the Investigator Award and free access to the 23rd International Symposium at Skamania Lodge, Stevenson, WA (USA) in 2013.

Please feel free to contact Carole Ryan at carole.ryan@uhn.ca if you have any questions.

Cardiac Pathology & Imaging Course in Calgary
June 21-23, 2012

The 3rd Resident Canadian Cardiac Pathology and Imaging Course will be held in Calgary, Alberta from June 21 until June 23, 2012. Prof. Robert Anderson will be the distinguished speaker and support the internationally renowned faculty from the University of Alberta (Stollery Children's Hospital and Alberta Children's Hospital), and University of Toronto (SickKids). The program is available at http://www.isachd.org or at http://www.cachnet.ca. The course is endorsed by ISACHD and paying ISACHD Members will receive a special registration fee (10% discount).

WG on Education

Erwin Oechslin
Chair, WG on Education, Toronto (Canada)

Els Pieper, UMS Groningen (NL) has finalized the program for a Web-based, 2-day teaching course for trainees with a background in adult cardiology. Our goal was to assign speakers to the different topics by mid March 2012, which hasn't happened, yet. We discussed options how to record the presentations at the semi-annual meeting during the ACC meeting in Chicago. We hope to have the presentations recorded by the end of this year.

Under the leadership of Dr. Gary Webb, Cincinnati, we are collecting information of educational tools and resources, which should be made available to the ACHD community. **Please send links of educational resources** to gary.webb@cchmc.org; these links will be posted at www.isachd.org.

Endorsement of ACHD Meetings
The following meetings have been endorsed:

* **NEW**: Pediatric & Adult Interventional Cardiac Symposium (PICS & AICS 2012), Chicago, Illinois, April 15-18, 2012 (www.isachd.org)
* **NEW**: the 3rd Resident Canadian Cardiac Pathology and Imaging Course, Calgary, Alberta; June 21-23, 2012 (www.isachd.org or www.cachnet.ca)
* 2nd International Congress on Cardiac Problems in Pregnancy, Berlin, Germany; May 17-20, 2012 (www.cachnet.ca)
* 22nd International Symposium on Adult Congenital Heart Disease, Toronto, May 30 - June 2, 2012 (www.uhn.ca/ACHDConference2012.asp)

Paying ISACHD members receive a 10% discount on the registration fee!

Please be proactive and contact me at erwin.oechslin@uhn.ca if you want to endorse your ACHD meeting.

Journal Watch

Am J Cardiol. 2012 Mar 29. [Epub ahead of print]

Incidence and Predictors of Sudden Cardiac Arrest in Adults With Congenital Heart Defects Repaired Before Adult Life.

Gallego P, Gonzalez AE, Sanchez-Recalde A, Peinado R, Polo L, Gomez-Rubin C, Lopez-Sendon JL, Oliver JM.

Source

Adult Congenital Heart Disease Unit, La Paz University Hospital, Madrid, Spain; Department of Cardiology, Virgen Macarena University Hospital,

Seville, Spain.

Abstract

Many adult survivors of repaired congenital heart disease (CHD) are at premature risk of death. Sudden cardiac arrest (SCA) is 1 of the leading causes of death but little is known about determinants for SCA in adults with repaired lesions. We sought to determine incidence and risk factors for SCA in a study population of 936 adults with previously repaired CHD who had completed follow-up at a single tertiary center during a mean period of 9 ± 7 years. Mean age at first examination in our institution was 21 ± 7 years. Diagnostic categories included tetralogy of Fallot (216), coarctation of the aorta (157), transposition complexes (99), single ventricle (55), and other CHD (409). During a total follow-up of 8,387 person-years, 22 patients (2.6 per 1,000 person-years) presented with SCA. Incidence of SCA varied widely between specific lesions; the highest incidence was observed in transposition complexes (10 per 1,000 person-years). Independent predictors of SCA were retrospectively identified using multivariate Cox proportional hazard modeling. Age at initial examination and severely impaired subaortic ventricular systolic function were independent risk factors for SCA (severe subaortic ventricular systolic dysfunction, adjusted hazard ratio 29, 95% confidence interval 11 to 72, p <0.001). SCA occurred in 23% of patients with severe subaortic ventricular systolic dysfunction versus 0.7% of patients with nonsevere decreased subaortic ventricular function (p <0.001). In conclusion, severe subaortic ventricular systolic dysfunction is a dominant multivariate predictor of SCA in an unselected population of adult survivors after surgery for CHD. Our data support the consideration of primary prevention strategies in these patients.

Clin Transplant. 2012 Mar 30. doi: 10.1111/j.1399-0012.2012.01611.x. [Epub ahead of print]

Transplantation for complex congenital heart disease in adults: a subanalysis of the Spanish Heart Transplant Registry.

Paniagua Martín MJ, Almenar L, Brossa V, Crespo-Leiro MG, Segovia J, Palomo J, Delgado J, González-Vílchez F, Manito N, Lage E, García-Guereta L, Rodríguez-Lambert JL, Albert DC.

Source

Department of Cardiology, Hospital Universitario A Coruña, A Coruña, Spain.

Abstract

Background: Congenital heart diseases (CHDs) have high infant mortality in their severe forms. When adulthood is reached, a heart transplant (HTx) may be required. Spanish adult population transplanted for CHD was analyzed and compared with the most frequent causes of HTx and between different subgroups of CHD.

Materials and Methods: A total of 6048 patients (HTx 1984-2009) were included. Pediatric transplants (<15 yr), combined transplants, reHTx, and HTx for heart diseases other than idiopathic dilated cardiomyopathy (IDCM) and ischemic heart disease (IHD) were excluded. Total patients

included: 3166 (IHD = 1888; IDCM = 1223; CHD = 55). Subgroups were studied as follows: (1) single ventricle with pulmonary stenosis (n = 18), (2) single ventricle with tricuspid atresia and Glenn/Fontan surgery (n = 10), (3) congenitally corrected transposition of the great vessels (TGV) or with switch atrial surgery (n = 10), and (4) CHD with right ventricle overload (n = 17).
Results: Survival probability was different between groups (p = 0.0001). Post hoc analysis showed some differences between groups (CHD vs. IHD, p = 0.05; CHD vs. IDCM, p = 0.5; IHD vs. IDCM, p = 0.0001). Early mortality was different between CHD subgroups (group 1 = 19%, group 2 = 40%, group 3 = 0%, group 4 = 29%; p < 0.001); however, overall mortality did not show differences between subgroups (p = 0.5).
Conclusions: The percentage of Spanish adult HTx patients for CHD is low (1%). The survival curve is better than for other HTx causes (IHD). Nevertheless, early mortality was higher, particularly in some subgroups (Fontan).

Am J Med Genet A. 2012 Mar 27. doi: 10.1002/ajmg.a.35343. [Epub ahead of print]
Diagnostic yield in adults screened at the Marfan outpatient clinic using the 1996 and 2010 Ghent nosologies.
Aalberts JJ, Thio CH, Schuurman AG, van Langen IM, van der Pol BA, van Tintelen JP, van den Berg MP.
Source
Department of Cardiology, University Medical Centre Groningen, University of Groningen, The Netherlands. jjj_aalberts@hotmail.com.
Abstract
Marfan syndrome (MFS) is diagnosed according to the Ghent nosology, which has recently been revised. In the Netherlands, evaluation for possible MFS is performed in specialized Marfan outpatient clinics. We investigated the diagnostic yield in our clinic and the impact of the 2010 nosology. All adult patients (n=343) who visited our clinic between 1998 and 2008 were included. We analyzed their reasons for referral, characteristics, and established diagnoses. In addition, we applied the 2010 nosology to all patients and compared the outcomes to those obtained with the 1996 nosology. Diagnoses that were made using the 1996 and the 2010 Ghent nosology included MFS (44/343 vs. 47/343), familial thoracic aortic aneurysm and/or dissection (22/343 vs. 22/343 patients), Loeys-Dietz syndrome (4/343 vs. 4/343 patients), and (familial) mitral valve prolapse (MVPS; 5/343 vs. 28/343 patients). In both nosologies, 77% of MFS patients had an FBN1 mutation. The 2010 nosology led to an increase in the number of diagnoses made: 4 additional cases of MFS were identified (one patient was "lost" who no longer fulfilled the criteria) and 23 additional cases of MVPS were diagnosed. The diagnostic yield of patients with aortic root dilatation was 65% using the 1996 nosology and 70% using the 2010 nosology. The change in diagnoses did not lead to a difference in clinical follow-up. We conclude that the diagnostic yield of our

specialized clinic was high, in particular in patients with aortic root dilatation. Further more the 2010 Ghent nosology led to a significant increase in the number of diagnoses made, mainly due to lowering of the diagnostic threshold for MVPS. © 2012 Wiley Periodicals, Inc.

Am J Cardiol. 2012 Mar 27. [Epub ahead of print]
Knowledge of and Preference for Advance Care Planning by Adults With Congenital Heart Disease.
Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach SL, Kovacs AH.

Source

Division of Cardiology, University Hospital of Basel, Basel, Switzerland; Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Toronto, Ontario, Canada.

Abstract

Congenital heart disease (CHD) is a chronic illness. Few adults with CHD are cured and those with disease of moderate or great complexity remain at risk of premature death. Current adult CHD guidelines recommend that providers encourage their patients to complete advance directives. We evaluated the prevalence of completed advance directives by and the preference for information about life expectancy of outpatients at a large adult CHD program. Two hundred patients with CHD (52% men, 35 ± 15 years old, range 18 to 79, 81% with disease of moderate or great complexity) completed a survey that assessed knowledge of advance directives and nature of and preferences for advance care planning. Only 5% of patients reported that they had completed advance directives; 56% had never heard of them. However, most patients (87%) reported that they would prefer to have an advance directive available if they were dealing with their own dying and were unable to speak for themselves. Patients who had formally identified substitute decision makers (n = 34) were typically older (47 ± 16 vs 33 ± 13 years, p <0.001) and more likely to have partners (30% vs 6%, p <0.001). Most patients (70%) reported that they wanted general information about the average life expectancy for patients with their heart condition. In conclusion, in contrast to recommendations from published guidelines, advance care planning documents are infrequently completed by outpatients. Health care providers caring for patients with CHD should educate their patients about advance directives and assist them in preparing formal end-of-life-planning documents.

Heart Vessels. 2012 Mar 29. [Epub ahead of print]
Complications of cardiac catheterization in adults and children with congenital heart disease in the current era.
Mori Y, Takahashi K, Nakanishi T.

Source

Department of Pediatric Cardiology, The Heart Institute, Tokyo Women's Medical University, Tokyo, Japan, y.mori@sis.seirei.or.jp.
Abstract
The number of adults with congenital heart disease (CHD) requiring diagnostic and/or therapeutic cardiac catheterization has been increasing. However, there have been few studies on the complications of performing cardiac catheterization in adults with CHD. The aim of this study was to determine the incidence of complications during congenital cardiac catheterization in both adults and pediatric patients. A total of 2134 consecutive cardiac catheterizations performed between 2003 and 2008 were prospectively analyzed. Complications were graded from 1 to 5 based on severity and these, with ≥grade 3 being defined as major. During the study period, 576 procedures (393 diagnostic, 90 interventional, and 93 electrophysiological) were performed in adult patients (≥18 years). Complex heart disease was present in 435 of 576 procedures (75.6 %). A total of 65 complications (11.3 %) with 13 major complications including 1 death (2.3 %) were encountered. The most common complications were arrhythmias. The majority of complications were successfully treated or temporary, and all but one of the patients were without residua. Of the 1558 pediatric procedures performed during the same period, we found a total of 229 complications (14.7 %), of which 89 (5.7 %) were major complications including 5 deaths. The safety of performing cardiac catheterization for adult CHD appears to be similar to that for pediatric patients. The complication rates in adults with CHD are low, but not negligible.

Clin Res Cardiol. 2012 Mar 28. [Epub ahead of print]Interventional closure of atrial septal defects without fluoroscopy in adult and pediatric patients.
Schubert S, Kainz S, Peters B, Berger F, Ewert P.
Source
Department of Congenital Heart Disease and Pediatric Cardiology, Deutsches Herzzentrum Berlin, Augustenburger Platz 1, 13353, Berlin, Germany, sschubert@dhzb.de.
Abstract
Background: Interventional closure of atrial septal defects (ASDs) with a transcatheter device is the preferred strategy in children and adults. This procedure has been proven in numerous studies, but X-ray and contrast agent exposure is still a major side effect. The aim of this study was to clarify whether the interventional closure of ASDs is possible and safe if it is guided by transesophageal echocardiography (TEE) alone.
Methods and Results: We retrospectively selected and studied pediatric and adult patients with interventional closure of ASDs at the Deutsches Herzzentrum Berlin (DHZB) without fluoroscopy between 1999 and 2010. We included 330 out of 1,605 patients; 254 had an

ASD II, 30 a PFO and 46 multiperforated atrial septum. Median age was 8.92 (0.96-76.3) years and median body weight 32.6 (8.3-156) kg. Median stretched defect size was 13 (5-29) mm. Median procedure time was 50 (20-170) min. Closure was performed in the majority of patients with the Amplatzer(®) septal occluder or Amplatzer(®) PFO occluder. The procedure succeeded in 98.2 % of cases and closure rate was 94.9 % after 48 h. Complication rate was low and procedure time was similar to that necessary with studies using fluoroscopy.
Conclusion: Interventional closure of ASDs is safe and effective if guided with TEE alone. The results can compete with those with the use of fluoroscopy. TEE-guided closure of ASD should be considered in more catheter laboratories to avoid unnecessary radiation exposure for the patient and the examiner.

Am J Cardiol. 2012 Mar 21. [Epub ahead of print]
Relation of Prolonged Tissue Doppler Imaging-Derived Atrial Conduction Time to Atrial Arrhythmia in Adult Patients With Congenital Heart Disease.
van der Hulst AE, Roest AA, Holman ER, Vliegen HW, Hazekamp MG, Bax JJ, Blom NA, Delgado V.
Source
Department of Pediatric Cardiology, Leiden University Medical Center, Leiden, The Netherlands.
Abstract
Atrial arrhythmia (AA) is common in adult patients with congenital heart disease (CHD). To enable the prevention of AA or its complications, timely identification of adult patients with CHD at risk of AA is crucial. Long total atrial activation times have been related to AA. Tissue Doppler imaging (TDI) permits noninvasive evaluation of the total atrial conduction time (PA-TDI duration). The present study evaluated the association between the PA-TDI duration and the development of AA in adult patients with CHD. A total of 223 adult patients with CHD were followed up for the occurrence of AA after PA-TDI duration assessment. The PA-TDI duration was defined as the interval from the onset of the P wave on the electrocardiogram to the peak of the A' wave at the lateral atrial wall on TDI tracings. Among the various clinical and echocardiographic parameters, the association between the PA-TDI duration and AA occurrence was investigated. The median follow-up was 39 months (interquartile range 21 to 57). A PA-TDI duration of ≥126 ms was associated with AA during follow-up (log-rank, $p < 0.001$). On multivariate analysis, a PA-TDI duration >126 ms (hazard ratio 2.25, 95% confidence interval 1.21 to 4.19) and history of AA (hazard ratio 4.89, 95% confidence interval 2.75 to 8.71) were independently associated with the occurrence of AA. In conclusion, PA-TDI duration and a history of AA were independently associated with the occurrence of AA in adult patients with CHD. The PA-TDI duration is a useful tool to identify

patients with CHD at risk of AA during follow-up.

Am J Cardiol. 2012 Mar 23. [Epub ahead of print]
Social Burden and Lifestyle in Adults With Congenital Heart Disease.
Zomer AC, Vaartjes I, Uiterwaal CS, van der Velde ET, Sieswerda GJ, Wajon EM, Plomp K, van Bergen PF, Verheugt CL, Krivka E, de Vries CJ, Lok DJ, Grobbee DE, Mulder BJ.

Source

Department of Cardiology, Academic Medical Center, Amsterdam, the Netherlands; Julius Center for Health Sciences and Primary Care, University Medical Center Utrecht, Utrecht, the Netherlands; InterUniversity Cardiology Institute of the Netherlands, Utrecht, the Netherlands.

Abstract

We aimed to evaluate how the presence and severity of congenital heart disease (CHD) influence social life and lifestyle in adult patients. A random sample (n = 1,496) from the CONgenital CORvitia (n = 11,047), the Dutch national registry of adult patients with CHD, completed a questionnaire on educational attainment, employment and marital statuses, and lifestyle (response 76%). The Utrecht Health Project provided a large reference group (n = 6,810) of unaffected subjects. Logistic regression models were used for subgroup analyses and to adjust for age, gender, and socioeconomic status where appropriate. Of all patients 51.5% were men (median age 39 years, interquartile range 29 to 51) with mild (46%), moderate (44%), and severe (10%) CHD. Young (<40-year-old) patients with CHD were more likely to have achieved a lower education (adjusted odds ratios [ORs] 1.6 for men and 1.9 for women, p <0.05 for the 2 comparisons), significantly more often unemployed (adjusted ORs 5.9 and 2.0 for men and women, respectively), and less likely to be in a relationship compared to the reference group (adjusted ORs 8.5 for men and 4.5 for women). These poorer outcomes were seen in all severity groups. Overall, the CHD population smoked less (adjusted OR 0.5, p <0.05), had more sports participation (adjusted OR 1.2, p <0.05), and had less obesity (adjusted OR 0.7, p <0.05) than the reference group. In conclusion, there was a substantial social disadvantage in adult patients with CHD, which was seen in all severity groups and primarily in young men. In contrast, adults with CHD had healthier lifestyles compared to the reference group.

Ann Thorac Surg. 2012 Mar 17. [Epub ahead of print]
Functional Health Status of Adults With Tetralogy of Fallot: Matched Comparison With Healthy Siblings.
Knowles R, Veldtman G, Hickey EJ, Bradley T, Gengsakul A, Webb GD, Williams WG, McCrindle BW.

Source

Division of Cardiology, Department of Pediatrics, University of Toronto, The Hospital for Sick Children, Toronto, Ontario, Canada.

Abstract
Background: Survival prospects for adults with repaired tetralogy of Fallot (TOF) are now excellent. Attention should therefore shift to assessing and improving functional health status and quality of life. We aimed to assess late functional health status of adults surviving TOF repair by matched comparison to their healthy siblings.
Methods: All 1,693 TOF repairs performed at our institution between 1946 and 1990 were reviewed. A matched comparison was undertaken whereby presumed survivors and their healthy sibling were contacted and asked to complete the Ontario Health Survey 1990 and the 36-Item Short Form Health Survey (SF-36) questionnaire.
Results: Both questionnaires were completed by 224 adult survivors and their sibling closest in age. Adults with repaired TOF had lower scores for self-perceived general health status ($p < 0.001$), were less likely to rate their health as good or excellent ($p < 0.001$), and had lower SF-36 scores for physical functioning and general health ($p = 0.001$) than their siblings. However, patients reported similar satisfaction with their lives, similar levels of social participation and support, and were as likely to be in long-term partnerships. Worse physical and mental health scores were associated with older age at surgery and at time of questionnaire completion and recent requirement for noncardiac medication.
Conclusions: Although reporting lower functional health status then their siblings, quality of life and life satisfaction for adults who underwent surgery for TOF during childhood is comparable to that of their siblings without heart defects. Follow-up of younger adults is required to understand current health outcomes attributable to improvements in the management of TOF.

Catheter Cardiovasc Interv. 2012 Mar 14. doi: 10.1002/ccd.23470. [Epub ahead of print]
Percutaneous interventions in high-risk patients following mustard repair of transposition of the great arteries.
Hill KD, Fleming G, Curt Fudge J, Albers EL, Doyle TP, Rhodes JF.
Source
Division of Pediatric Cardiology, Duke University Medical Center, Durham, North Carolina. kevin.hill@duke.edu.
Abstract
Objectives: To assess safety, efficacy, and intermediate term outcomes of percutaneous interventions in Mustard patients.
Background: Baffle leaks and obstruction are present in 20% of Mustard survivors. Surgical reintervention is associated with high mortality.
Methods: Retrospective review of percutaneous interventions performed at three adult congenital catheterization programs.
Results: Overall, 26 catheterizations and 29 interventions were

performed in 22 patients (mean age 32.4 ± 8.3 years). Previous laser pacemaker lead extraction was successful in seven of seven procedures where the lead was at risk. Stent placement was successful in all 18 patients with systemic venous baffle (SVB) obstruction (mean gradient: 6.2 ± 3.4-0.6 ± 1.0 mm Hg; P < 0.01, narrowest diameter 4.5 ± 4.5-17.1 ± 3.9 mm; P < 0.01). Balloon angioplasty was performed in two patients for pulmonary venous baffle (PVB) obstruction with mixed results. Baffle leak interventions included device occlusion (n = 6), coil occlusion (n = 1), and covered stent occlusion (n = 3). Postprocedural residual leaks were demonstrated in three of eight. In two of the three the residual leak was not appreciable at 1-year follow-up. No patient experienced leak or obstruction related symptom recurrence (mean follow-up: 33.4 ± 29.5 months). Complications included one death secondary to ventricular arrhythmia 2 days after PVB angioplasty and device related inferior SVB obstruction with resolution following stent placement.
Conclusions: Stent placement for SVB obstruction following Mustard repair is effective and likely safer than surgical intervention. Baffle leak occlusion can be safely accomplished but residual leaks are common in the short term.

Eur J Cardiovasc Nurs. 2012 Mar 13. [Epub ahead of print]
Patients with a congenital heart defect and Type D personality feel functionally more impaired, report a poorer health status and quality of life, but use less healthcare.
Schoormans D, Mulder BJ, van Melle JP, Pieper EG, van Dijk AP, Sieswerda GJ, Hulsbergen-Zwarts MS, Plokker TH, Brunninkhuis LG, Vliegen HW, Sprangers MA.
Source
Academic Medical Centre, Amsterdam and Interuniversity Cardiology Institute of the Netherlands, Utrecht, the Netherlands.
Abstract
Background: Type D personality, characterized by high levels of negative affectivity and social inhibition, is related to mortality, morbidity, poor health status, quality of life (QoL) and less healthcare utilization in various cardiovascular patient groups. To date, studies in patients with congenital heart disease (CHD) are lacking.
Aims: (1) To examine the prevalence of Type D personality in CHD patients; (2) to compare Type D to non-Type D patients with regard to disease severity, functional status, health status and QoL; and (3) to examine the extent to which Type D personality is independently related to healthcare utilization.
Methods: A total of 1109 adult CHD patients were included in a questionnaire survey. Due to missing data, 302 patients were excluded.
Results: The prevalence of Type D personality was 20.4%. Type D patients reported a poorer functional status, health status and QoL

than non-Type D patients (p<0.05). Type D patients reported less healthcare use than non-Type D patients (primary and cardiac outpatient healthcare: adjusted OR=0.56, 95% CI=0.35-0.90; inpatient healthcare: adjusted OR=0.38, 95% CI=0.17-0.83). Results of a post-hoc analysis showed a high prevalence of Type D personality in patients with a poor functional status who did not consult their cardiologist.
Conclusion: Type D patients report a poorer functional status, health status and QoL, but less healthcare utilization. In clinical practice, patients should be screened for Type D personality, since social inhibition may prevent them from contacting a healthcare provider in the event of symptom aggravation.

Future Cardiol. 2012 Mar;8(2):329-42.
Heart transplantation in adults with end-stage congenital heart disease.
Burchill LJ, Ross HJ.
Source
University of Toronto, Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, Canada.
Abstract
Residual abnormalities in cardiac structure and function predispose adults with congenital heart disease to late-onset heart failure and its complications. Evaluation of this population requires collaboration between adult congenital and heart failure specialists. In addition to assessing heart transplant eligibility, clinicians must balance the risks of premature listing against progressive heart failure and increased waiting list mortality. Following heart transplantation, adults with congenital heart disease have higher mortality due to an increased risk of bleeding, infection and donor right heart failure secondary to pulmonary hypertension. Concerns relating to increased early mortality should be balanced against superior long-term survival in adult congenital heart disease patients surviving beyond the first year after heart transplantation.

Future Cardiol. 2012 Mar;8(2):305-13.
Changing lesion demographics of the adult with congenital heart disease: an emerging population with complex needs.
Stuart AG.
Source
Congenital Heart Unit, Bristol Royal Hospital for Children/Bristol Heart Institute, Upper Maudlin St, Bristol, BS2 8XW, UK. graham.stuart@nhs.net.
Abstract
The demography of congenital heart disease is changing. Largely as a consequence of successful cardiac surgery in childhood, there are an increasing number of adults with congenital heart disease with a prevalence of more than four per 100 adults. The type of disease in adults is also changing with an increasing number of survivors with

complex disease. These patients have a significantly increased healthcare requirement in comparison to healthy adults and this includes noncardiac, multisystem morbidity. The adult congenital heart disease population are now developing problems associated with aging and there is a new population of geriatrics with congenital heart disease. As survival continues to improve, increased healthcare resources need to be directed towards the management of the adult with congenital heart disease.

Future Cardiol. 2012 Mar;8(2):297-304.
Looking ahead: clinical trial design in adult congenital heart disease.
Khairy P.
Source
Adult Congenital Heart Center, Montreal Heart Institute, 5000 Belanger St. E., Montreal, QC, H1T 1C8, Canada. paul.khairy@umontreal.ca.
Abstract
Continued growth in the field of adult congenital heart disease has given rise to a critical mass of patients, investigators and resources dedicated to advancing knowledge through research. Impressive gains over the past decade have largely been driven by successful observational studies. Despite the privileged role of clinical trials in the hierarchy of evidence-based medicine, few such studies have thus far been conducted in adult congenital heart disease. Obstacles and challenges particular to clinical trials in adult congenital heart disease are addressed in this review. In looking ahead, examples of creative methodological solutions to maximizing the efficiency of clinical trials for rare diseases are discussed, including Bayesian analyses, outcome-adaptive randomization and internal pilot studies.

Future Cardiol. 2012 Mar;8(2):285-96.
Robotics and imaging in congenital heart surgery.
Vasilyev NV, Dupont PE, Del Nido PJ.
Source
Department of Cardiac Surgery, Children's Hospital Boston, MA, USA.
Abstract
The initial success seen in adult cardiac surgery with the application of available robotic systems has not been realized as broadly in pediatric cardiac surgery. The main obstacles include extended set-up time and complexity of the procedures, as well as the large size of the instruments with respect to the size of the child. Moreover, while the main advantage of robotic systems is the ability to minimize incision size, for intracardiac repairs, cardiopulmonary bypass is still required. Catheter-based interventions, on the other hand, have expanded rapidly in both application as well as the complexity of procedures and lesions being treated. However, despite the development of sophisticated devices, robotic systems to aid catheter procedures have not been commonly applied in children. In this article, we describe new catheter-like robotic delivery platforms, which facilitate safe navigation and enable complex repairs, such as tissue approximation and fixation, and tissue removal, inside the beating heart. Additional features including the

tracking of rapidly moving tissue targets and novel imaging approaches are described, along with a discussion of future prospects for steerable robotic systems.

Future Cardiol. 2012 Mar;8(2):171-7.
Changing epidemiology and mortality in adult congenital heart disease: looking into the future.
Greutmann M, Tobler D.
Source
Department of Cardiology, University Hospital Basel, Petersgraben 4, CH-4031 Basel, Switzerland.
Abstract
Advances in surgical and medical treatment of children born with congenital heart disease have led to a growing number of adult survivors, particularly to a growing number of adults with complex congenital heart disease. Childhood mortality has continuously decreased over the last few decades and mortality has shifted almost entirely to adulthood. However, most patients are not cured and many remain at risk of premature death. The extent of excess mortality among individual congenital disease entities is not well defined. In this article we outline the current demographics of adults with selected congenital heart lesions. Based on these contemporary patient cohorts, we delineate future changes in patient demographics. A better understanding of these trends may help in the optimal planning of future resource allocation for medical care and optimal planning of multicenter research, for this novel and growing population of young, chronically ill adults.

Heart. 2012 Mar 7. [Epub ahead of print]
B-type natriuretic peptide concentrations in contemporary Eisenmenger syndrome patients: predictive value and response to disease targeting therapy.
Diller GP, Alonso-Gonzalez R, Kempny A, Dimopoulos K, Inuzuka R, Giannakoulas G, Castle L, Lammers AE, Hooper J, Uebing A, Swan L, Gatzoulis M, Wort SJ.
Source
Royal Brompton Hospital, London, UK.
Abstract
Objective: To assess the relationship between elevated levels of B-type natriuretic peptide (BNP) and outcome in patients with Eisenmenger syndrome.
Design: Retrospective study.
Setting: Tertiary centre for adult congenital heart disease.
Patients: All patients with Eisenmenger syndrome (n=181, age 36.9±12.1 years, 31% with Down syndrome) in whom BNP concentrations were measured as part of routine clinical care were included.
Main Outcome Measures: The study end point was all cause mortality.
Results: During a median follow-up period of 3.3 years, 20 patients (7 with Down syndrome) died. Higher BNP concentrations were predictive of all cause mortality on univariate analysis in patients with or without

Down syndrome. On multivariable Cox proportional hazard analysis, BNP predicted survival independently of renal function, Down syndrome, or 6 min walk test distance (p=0.004). Temporal increases in BNP concentration were also found to predict mortality. Treatment with disease targeting therapies was associated with a significant reduction in BNP concentrations.
Conclusions: BNP concentrations predict outcome in contemporary Eisenmenger patients. Increases in BNP concentrations over time are also of prognostic significance. In addition, disease targeting therapies may help to reduce BNP concentrations in this population, while treatment-naïve patients have static or rising BNP concentrations.

Int J Cardiol. 2012 Mar 3. [Epub ahead of print]
Disease targeting therapies in patients with Eisenmenger syndrome: Response to treatment and long-term efficiency.
Diller GP, Alonso-Gonzalez R, Dimopoulos K, Alvarez-Barredo M, Koo C, Kempny A, Harries C, Parfitt L, Uebing AS, Swan L, Marino PS, Wort SJ, Gatzoulis MA.
Source
Adult Congenital Heart Disease Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital, London, UK; National Heart and Lung Institute, Imperial College School of Medicine, London, UK.
Abstract
Objectives: To examine long-term efficacy of disease targeting therapies (DTT) in patients with Eisenmenger syndrome.
Methods: All adult patients with Eisenmenger syndrome treated with DTT at our institution were included. Functional class (FC), oxygen saturation and 6-minute walk test distance (6MWTd) were analysed retrospectively.
Results: Between 2002 and 2010, 79 Eisenmenger patients (21 males, 16 with Down syndrome) aged 34±10years (range 17-68years) were included. Median follow-up was 3.3years (range 0.2 to 8.9years). 6MWTd increased early after initiation of DTT, with a plateau after approximately 3years and no obvious trend towards a deterioration on average during longer-term follow-up. Two patients died during follow-up and escalation of treatment was required in 18 patients after a median period of 2.5years. Escalation of therapy was also associated with an increase in 6MWTd. In addition, FC improved on DTT and oxygen saturation, increased, both at rest and peak exercise. This effect was more pronounced in the patients with the lowest baseline oxygen saturation at rest.
Conclusions: Long-term DTT is safe and improves objective exercise capacity and subjective symptoms. Response to DTT was typically observed early after initiation of DTT and was, on average, maintained longer-term. However, 1 in 5 patients required escalation of DTT, with time, due to symptomatic deterioration and this was associated with an afresh improvement in 6MWTd.

BJOG. 2012 Mar 6. doi: 10.1111/j.1471-0528.2012.03295.x. [Epub ahead of print]

Pulmonary hypertension and pregnancy-a review of 12 pregnancies in nine women.

Curry R, Fletcher C, Gelson E, Gatzoulis M, Woolnough M, Richards N, Swan L, Steer P, Johnson M.

Source

Academic Department of Obstetrics and Gynaecology, Faculty of Medicine, Imperial College London, Chelsea and Westminster NHS Foundation Trust Adult Congenital Heart Disease Unit, Royal Brompton and Harefield NHS Foundation Trust, and the National Heart and Lung Institute at Imperial College, London Magill Department of Anaesthesia, Intensive Care and Pain Management, Chelsea & Westminster NHS Foundation Trust, London, UK.

Abstract

Please cite this paper as: Curry R, Fletcher C, Gelson E, Gatzoulis M, Woolnough M, Richards N, Swan L, Steer P, Johnson M. Pulmonary hypertension and pregnancy-a review of 12 pregnancies in nine women. BJOG 2012; DOI: 10.1111/j.1471-0528.2012.03295.x. Objective To report outcomes in a recent series of pregnancies in women with pulmonary hypertension (PH). Design Retrospective case note review. Setting Tertiary referral unit (Chelsea and Westminster and Royal Brompton Hospitals). Sample Twelve pregnancies in nine women with PH between 1995 and 2010. Methods Multidisciplinary review of case records. Main outcome measures Maternal and neonatal mortality and morbidity. Results There were two maternal deaths (1995 and 1998), one related to pre-eclampsia and one to arrhythmia. Maternal morbidity included postpartum haemorrhage (five cases), and one post-caesarean evacuation of a wound haematoma. There were no perinatal deaths, nine live births and three first-trimester miscarriages. Mean birthweight was 2197g, mean gestational age was 34weeks (range 26-39), and mean birthweight centile was 36 (range 5-60). Five babies required admission to the neonatal intensive care unit, but were all eventually discharged home. All women were delivered by caesarean section (seven elective and two emergency deliveries), under general anaesthetic except for one emergency and one elective caesarean performed under regional block. Conclusions Maternal and fetal outcomes for women with PH may be improving. However, the risk of maternal mortality remains significant, so that early and effective counselling about contraceptive options and pregnancy risks should continue to play a major role in the management of such women when they reach reproductive maturity.

Hepatology. 2012 Mar 2. doi: 10.1002/hep.25692. [Epub ahead of print]

Congenital heart disease and the liver.

Asrani SK, Asrani NS, Freese DK, Phillips SD, Warnes CA, Heimbach J, Kamath PS.

Source

Division of Gastroenterology and Hepatology, Mayo Clinic College of Medicine, Rochester, Minnesota.

Abstract

There are approximately one million adult patients with congenital heart

disease (CHD) in the United States and the number is increasing. Hepatic complications are common and may occur secondary to persistent chronic passive venous congestion or decreased cardiac output due to the underlying cardiac disease, or as a result of palliative cardiac surgery; transfusion or drug related hepatitis may also occur. The unique physiology of Fontan circulation is particularly prone to development of hepatic complications and is in part related to the duration of the Fontan procedure. Liver biochemical test abnormalities may be related to cardiac failure, due to intrinsic liver disease, secondary to palliative interventions, or drug-related. Complications of portal hypertension and rarely, hepatocellular carcinoma may also occur. Abnormalities such as hypervascular nodules are often seen; in the presence of cirrhosis surveillance for hepatocellular carcinoma is necessary. Judicious perioperative support is required when cardiac surgery is performed in patients with advanced hepatic disease. Traditional models for liver disease staging may not fully capture the severity of disease in patients with CHD. The effectiveness or safety of isolated liver transplantation in patients with significant CHD is limited in adults; combined heart-liver transplantation may be required in those with decompensated liver disease or hepatocellular carcinoma, but experience is limited in the presence of significant CHD. The long term sequelae of many reparative cardiac surgical procedures are not yet fully realized; understanding the unique and diverse hepatic associations and the role for early cardiac transplantation in this population is critical. As this population continues to grow and age, consideration should be given to developing consensus guidelines for a multidisciplinary approach to optimize management of this vulnerable population. (HEPATOLOGY 2012.).

Int J Cardiol. 2012 Feb 25. [Epub ahead of print]
Increased risk for ascending aortic dilatation in patients with complex compared to simple aortic coarctation.
Luijendijk P, Franken RJ, Vriend JW, Zwinderman AH, Vliegen HW, Winter MM, Groenink M, Bouma BJ, Mulder BJ.
Source
Department of Cardiology, Academic Medical Centre, Amsterdam, The Netherlands; Interuniversity Cardiology Institute of the Netherlands, Utrecht, The Netherlands.
Abstract
Aims: Aortic coarctation (CoA) occurs as a "simple" isolated disorder, and in a more "complex" form, combined with associated congenital cardiac abnormalities. Long term outcome of all CoA patients may be complicated by dilatation of the thoracic aorta. The aim of this study was to quantify progressive aortic dilatation, and identify determinants for progressive aortic dilatation.
Methods and Results: Cardiovascular Magnetic Resonance Imaging (CMR) and echocardiographic data of 93 CoA patients were analyzed retrospectively on the progression, and determinants, of progressive thoracic aortic dilatation. Outcome of simple- versus complex CoA patients were compared. 93 CoA patients (mean age 39±12years, male

59%) were followed with CMR (follow-up 5.3±1.8years). Twenty-eight patients were classified as simple- and 68 as complex CoA. The mean progression rate of thoracic aortic dilatation was highest in the ascending aorta with 2.2±2.0mm/5years (range 0-7.2mm/5years). History of VSD (β=1.77, P=0.004) and an increased left ventricular mass index (β=0.02, P=0.04) were associated with progressive ascending aortic dilatation. Complex CoA patients show an increased progression rate compared to simple CoA patients with 2.4mm/5years versus 1.5mm/5years respectively. (P=0.03).
Conclusion: Adult post-coarctectomy patients show an increased mean progression rate of ascending aortic dilatation with 2.2mm/5years. The progression rate of ascending aortic dilatation is increased in complex CoA patients, as compared to simple CoA patients. These findings point towards a more comprehensive genetic subset of patients with an increased risk for progressive ascending aortic dilatation.

Int J Cardiol. 2012 Feb 26. [Epub ahead of print]
Mirror image atrial dilatation in adult patients with atrial fibrillation and congenital heart disease.
Bouchardy J, Marelli AJ, Martucci G, Bottega N, Therrien J.
Abstract
Background: Atrial fibrillation (AF) is largely regarded to be initiated from left atrial (LA) dilatation, with subsequent dilatation of the right atrium (RA) in those who progress to chronic AF. We hypothesized that in adult patients with right-sided congenital heart disease (CHD) and AF, RA dilatation will predominate with subsequent dilatation of the left atrium, as a mirror image.
Methods: Adult patients with diagnosis of right-sided, ASD or left-sided CHD who had undergone an echocardiographic study and electrocardiographic recording in 2007 were included. RA and LA area were measured from the apical view. AF was diagnosed from a 12-lead electrocardiogram or Holter recording. A multivariate logistic regression model was used to identify predictors of AF and linear regression models were performed to measure relationship between RA and LA area and AF.
Results: A total of 291 patients were included in the study. Multivariate analysis showed that age (p=0.0001), RA (p=0.025) and LA area (p=0.0016) were significantly related to AF. In patients with pure left-sided pathologies, there was progressive and predominant LA dilatation that paralleled the development of AF from none to paroxysmal to chronic AF. In patients with pure right-sided pathologies, there was a mirror image of progressive and predominant RA dilatation with the development of AF.
Conclusion: We observed a mirror image atrial dilatation in patients with right sided disease and AF. This may provide novel mechanistic insight as to the origin of AF in these patients and deserves further studying in the form of targeted electrophysiological studies.

Congenit Heart Dis. 2012 Feb 23. doi: 10.1111/j.1747-0803.2012.00633.x. [Epub ahead of print]

Does Anticoagulation in Eisenmenger Syndrome Impact Long-term Survival?

Sandoval J, Santos LE, Córdova J, Pulido T, Gutiérrez G, Bautista E, Martinez Guerra ML, Peña H, Broberg CS.

Source

Cardiopulmonary Department, Ignacio Chavez National Institute of Cardiology, Mexico City, Mexico Adult Congenital Heart Program, Division of Cardiology, Oregon Health and Science University, Portland, Ore, USA.

Abstract

Objective: To determine the impact of anticoagulation on survival in Eisenmenger syndrome.

Background: The use of anticoagulation for primary prevention of adverse events in patients with Eisenmenger syndrome has been proposed but not studied. Strong arguments have been made both for and against anticoagulation based on the known risk of hemoptysis and pulmonary vascular thrombosis.

Design and Setting: Retrospective cohort study at a tertiary referral hospital.

Patients and Interventions: One hundred forty-four patients with established Eisenmenger physiology all underwent initial laboratory, echocardiographic, and catheterization evaluation after initial referral. We retrospectively identified patients who were started on anticoagulation (AC) and compared them to patients who did not receive anticoagulation therapy (non-AC). Baseline variables were compared between groups, as well as between survivors and nonsurvivors. Analyses of prognostic factors and survival were done using Cox and Kaplan-Meier methods. Outcome Measures. The primary outcome was death since time of baseline evaluation.

Results: We identified 48 anticoagulated and 44 nonanticoagulated patients with Eisenmenger physiology (oxygen saturation 82±9%, PaO(2) 48±8mmHg, hemoglobin 18.6±4g/dL). More atrial septal defect patients were in the AC group, but there were no other baseline differences in clinical, functional, or hemodynamic data. After mean follow-up of 7±5.4years (range 1-31), 11 patients died in the AC and 10 died in the non-AC group. There was no survival difference between groups (log rank test=1.78; P is not significant). For the entire cohort, mortality was significantly associated with New York Heart Association class 3-4 (hazard ratio=4.2), evidence of right heart failure (hazard ratio=13.6), and a mean corpuscular volume <80fL (hazard ratio=3.8). Use of anticoagulation did not impact survival. Bleeding complications occurred in seven (16%) of AC patients, including two fatalities.

Conclusions: Anticoagulation had no impact on long-term survival in this limited study. These data may be useful in considering future studies addressing this question.

Congenit Heart Dis. 2012 Feb 20. doi: 10.1111/j.1747-0803.2012.00635.x. [Epub ahead of print]

The Safety and Effects of Bosentan in Patients with a Fontan Circulation.

Bowater SE, Weaver RA, Thorne SA, Clift PF.

Source

University Hospital Birmingham Foundation NHS Trust, Birmingham, United Kingdom.

Abstract

Objective. Adult patients with a Fontan circulation tend to have diminished exercise capacity. The principal objective of this study was to investigate the safety of the endothelin receptor antagonist bosentan in Fontan patients, and, secondarily, to assess effects on cardiovascular performance, New York Heart Association functional classification (NYHA FC), and ventricular function. Design. A 6-month prospective, single-center, pilot, safety study of bosentan in Fontan patients. Setting. Adult Congenital Heart Disease referral center. Patients. All patients ≥18 years old with a Fontan circulation and in NYHA FC ≥II were invited to enroll. Interventions. Patients started on 62.5mg bid of bosentan, uptitrating to 125mg bid after 2 weeks. Outcome Measures. Safety was assessed by the incidence of anticipated and unanticipated adverse events during the 6-month study period; specifically those relating to hepatic, renal, or hematological dysfunction as measured by monthly blood tests. Other outcome measures included cardiopulmonary exercise test, 6-minute walk distance test, Borg dyspnea index, NYHA FC, and ventricular function parameters using transthoracic echocardiography. Results. Of the eight patients enrolled, six completed the study. Two patients withdrew from the study (one for non-trial related reasons, one due to adverse events). No clinically significant adverse events relating to bosentan therapy occurred during this study and, in particular, no significant abnormalities in hepatic function tests were observed. Three patients reported transient adverse events. Improvements in NYHA FC and systolic ventricular function were observed after 6 months of bosentan treatment. Conclusions. The small number of patients with a Fontan circulation in our study was able to tolerate bosentan for 6 months. The safety and tolerability of bosentan in a larger patient population remains unknown. The results presented here justify further investigation in larger studies.

Am J Cardiol. 2012 Mar 15;109(6):873-80. Epub 2012 Jan 13.

Functional health status in adult survivors of operative repair of tetralogy of fallot.

Hickey EJ, Veldtman G, Bradley TJ, Gengsakul A, Webb G, Williams WG, Manlhiot C, McCrindle BW.

Source

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Abstract

We aimed to determine late functional health status of the growing adult population with repaired tetralogy of Fallot (TOF). We studied all 840 patients with TOF born from 1927 through 1984 who survived to adulthood (> 18 years of age). Clinical follow-up was by chart review, telephone interview (n = 706), and echocardiographic reports (n = 339).

Functional health status was assessed using Short Form-36 (SF-36) surveys (n = 396) indexed to normative data. Risk of reoperation was low (≈ 1%/year) but increased beyond age 40 years. At latest follow-up moderate or severe pulmonary regurgitation was common (54%) and right ventricular outflow tract stenosis presented in 1/3. Consequently, evidence of right ventricular dilatation and dysfunction and tricuspid regurgitation was typical. Left-sided abnormalities were also common: hypertrophy ($p < 0.0001$) and outflow tract dilation ($p < 0.0001$) with at least mild aortic regurgitation in > 50%. Cardiorespiratory symptoms were reported in 45% (palpitations 27%, dyspnea 21%, chest pain 17%). SF-36 scores were significantly below normal for 4 physical domains ($p < 0.001$). Decrements in physical functioning were associated particularly with older age at follow-up ($p < 0.0001$), associated syndromes/lesions, reoperations, ventricular dysfunction, tricuspid regurgitation, residual septal defects, and cardiorespiratory symptomatology. Echocardiographic abnormalities were more common in older patients ($p < 0.0001$). All 3 SF-36 domains specific to psychosocial well-being were normal. In conclusion, despite excellent survival prospects, physical compromise is common in adults with repaired TOF. Greater decrements in older patients may reflect late deterioration with advancing age or cohort effects related to historical management. Efforts to limit ventricular and outflow tract dysfunction may translate into improved late functional status.

Contemp Clin Trials. 2012 Mar;33(2):410-6. Epub 2011 Nov 13.
Rationale and design of a trial on the effect of high dose statins on cardiovascular risk in adults after successful coarctation repair.
Luijendijk P, Bouma BJ, Vriend JW, Groenink M, Vliegen HW, de Groot E, Pieper PG, van Dijk AP, Sieswerda GT, Veen G, Zwinderman AH, Mulder BJ.
Source
Department of Cardiology, Academic Medical Centre, Amsterdam, The Netherlands.
Abstract
Background: HMG-coA-reductase-inhibitors (statins) have been proven to reduce atherosclerosis progression as observed by carotid intima-media thickness in patients with known coronary heart disease, independent of lipid lowering. Cardiovascular complications are common in patients after successful coarctation repair. The effect of statins on cardiovascular risk in adults after successful coarctation repair has not yet been established.
Methods: We designed a multicentre, prospective, randomised, open label trial to evaluate the effect of the HMGcoA-reductase-inhibitor (Atorvastatin) on atherosclerotic progression in adult post-coarctectomy patients. The primary endpoint in this study is the carotid intima-media thickness as measured by Bmode ultrasonography of the carotid arteries.
Conclusion: This large prospective, randomised, open label trial will establish the effect of HMG-coA-reductase inhibitors (Atorvastatin) on cardiovascular risk in adult patients after successful coarctation repair.

J Interv Card Electrophysiol. 2012 Mar;33(2):143-9. Epub 2011 Oct 21.
Systemic venous anatomy in congenital heart disease: implications for electrophysiologic testing and catheter ablation.
Cordina RL, Celermajer DS, McGuire MA.

Source

Royal Prince Alfred Hospital, Sydney, Australia.

Abstract

Introduction: Cardiac arrhythmias are a significant problem in patients with congenital heart disease. Many patients with congenital heart disease have abnormal systemic venous anatomy which can complicate electrophysiologic testing, catheter ablation and pacemaker and defibrillator implantation. We reviewed the systemic venous anatomy in a cohort of patients undergoing electrophysiologic testing and catheter ablation.
Methods and results: We reviewed all electrophysiologic studies performed in patients with adult congenital heart disease (n=80) at our institution between January 1998 and October 2009. Ten patients (13%) had a congenital systemic venous anomaly. Of these, seven (9%) had a left superior vena cava and four (5%) had infrahepatic interruption of the inferior vena cava (two had both anomalies). One patient's inferior vena cava was connected to a left-sided atrium; she had right atrial isomerism. In four patients (40%), systemic venous abnormalities were discovered at the time of electrophysiologic testing.
Conclusions: Systemic venous anomalies occur frequently in the congenital heart disease population and may complicate electrophysiologic testing and catheter ablation. Pre-procedural imaging may assist in facilitating a successful procedure.

Congenit Heart Dis. 2012 Mar;7(2):103-10. doi: 10.1111/j.1747-0803.2011.00567.x. Epub 2011 Oct 20.
Five-year Follow-up of Intracardiac Echocardiography-assisted Transcatheter Closure of Complex Ostium Secundum Atrial Septal Defect.
Rigatelli G, Dell' Avvocata F, Cardaioli P, Giordan M, Vassiliev D, Nghia NT, Chen JP.

Source

Section of Adult Congenital and Adult Heart Disease, Cardiovascular Diagnosis and Endoluminal Interventions, Rovigo General Hospital, Rovigo, Italy Interventional Cardiology, National Heart Institute, Sofia, Bulgaria Interventional Cardiology Department Cho Rey Hospital, Ho Chi Minh City, Vietnam Saint Joseph's Heart and Vascular Institute, Atlanta, Ga, USA.

Abstract

Objective. We sought to prospectively evaluate long-term follow-up results of intracardiac echocardiography-aided transcatheter closure of complex atrial septal defects (ASD) in the adults. Design and Settings. Prospective multicenter registry in tertiary care hospitals. Patients and Interventions. Over a 5-year period, we prospectively enrolled 56 patients (mean age 49±16.7 years, 24 females) who have been referred

to our center for catheter-based closure of complex secundum ASD (>25mm diameter, deficiency of ≥1 rim, multiple secundum ASD, multiperforated ASD, associated incomplete floor of the fossa ovalis with or without aneurysm, embryonic remnants of incomplete atrial septation). All patients were screened by means of transesophageal echocardiography before the operation. Eligible patients underwent intracardiac echocardiography study and closure attempt. Results. Forty patients underwent a transcatheter closure attempt: transesophageal echocardiography-planned device type and size were modified in 32 patients (64%). Rates of procedural success, predischarge occlusion, and major complications rate were 100%, 90%, and 2%, respectively. On mean follow-up of 5.4±1.8 years, the follow-up occlusion rate was 98%. During follow-up, only one case of permanent atrial fibrillation was observed. There were no cases of aortic/atrial erosion, device thrombosis, or new atrioventricular valve dysfunction. Conclusions. Intracardiac echocardiography-guided complex secundum ASD transcatheter closure is safe and effective and appears to have excellent long-term results, thus minimizing potential complications resulting from the complex anatomy.

J Thorac Cardiovasc Surg. 2012 Mar;143(3):569-75. Epub 2011 Sep 9.
Exercise capacity and cardiac reserve in children and adolescents with corrected pulmonary atresia with intact ventricular septum after univentricular palliation and biventricular repair.
Romeih S, Groenink M, Roest AA, van der Plas MN, Hazekamp MG, Mulder BJ, Blom NA.

Source

Department of Cardiology, Academic Medical Center, Amsterdam, The Netherlands.

Abstract

Objective: Management of pulmonary atresia with intact ventricular septum is challenging and depends on the severity of right ventricular hypoplasia. Clinical outcomes of biventricular repair seem favorable to univentricular palliation, but data on superiority of biventricular repair regarding exercise capacity are conflicting. We investigated the response to physical and pharmacologic stress in patients with surgically corrected pulmonary atresia with intact ventricular septum.

Methods: Sixteen patients (7 patients after univentricular palliation, age 11.8 ± 2.6 years; 7 patients after biventricular repair, age 12.9 ± 2.7 years; and 2 patients after 1.5 ventricular repair, age 12 and 19 years) underwent cardiopulmonary exercise test, dobutamine stress magnetic resonance imaging, and delayed contrast-enhanced magnetic resonance imaging.

Results: The univentricular group showed impaired exercise capacity in contrast with normal exercise capacity in the biventricular group. Left ventricular ejection fraction increased in both groups. With dobutamine, left ventricular stroke volume increased only in the biventricular group (+11.3 ± 5.0 mL, $P < .001$) and not in the univentricular group (-0.04 ± 3.6 mL, $P = .9$). Heart rate increase was inadequate in the univentricular group. Maximum oxygen consumption and oxygen pulse were strongly

correlated with left ventricular stroke volume during stress but not at rest. The results of the 2 patients after 1.5 ventricular repair were comparable to those of the univentricular group. No myocardial fibrosis was detected.
Conclusions: Impaired exercise capacity in children and adolescents with pulmonary atresia with intact ventricular septum after univentricular palliation is related to decreased cardiac reserve and inadequate chronotropic response. Young patients with pulmonary atresia with intact ventricular septum after biventricular repair show normal exercise capacity and cardiac reserve.

Int J Cardiol. 2012 Mar 22;155(3):383-7. Epub 2010 Nov 20.
End-of-life in adults with congenital heart disease: a call for early communication.
Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach LS, Kovacs AH.
Source
Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Toronto, Canada.
Abstract
Background: We investigated preferences of adults with congenital heart disease (CHD) and their health care providers regarding end-of-life (EOL) communication.
Methods: Adult CHD outpatients and health care providers completed surveys about preferences for and experiences with EOL communication. Responses were compared between patients and providers.
Results: Two hundred patients (18-79 years) and 48 CHD health care providers (primarily cardiologists) completed surveys. Only 2 patients (1%) indicated that they had discussed EOL planning with their medical team. In contrast, 50% of providers reported that they typically discuss issues including life expectancy, advance planning, and resuscitation preferences with their outpatients. Seventy-eight percent (156/199) of patients wanted their medical team to raise EOL issues; this preference was independent of disease complexity and socio-demographic factors. In contrast, providers reported that their EOL discussions increase in accordance with disease complexity ($p<0.001$). Early initiation of EOL discussions, before diagnosis with life-threatening complications, was favored by 62% of patients but only 38% of providers ($p<0.001$).
Conclusion: Health professionals caring for adults with CHD should explore preferences of their patients for EOL discussions earlier in the disease course, and not only with patients facing life-threatening complications and/or with complex conditions. When EOL discussions do occur, health care providers should attempt to ensure that patients better understand these conversations. Increased attention to EOL issues is proposed in order to improve the care of patients with CHD across the lifespan.

Int J Cardiol. 2012 Mar 22;155(3):378-82. Epub 2010 Nov 16.

Bosentan-sildenafil association in patients with congenital heart disease-related pulmonary arterial hypertension and Eisenmenger physiology.

D'Alto M, Romeo E, Argiento P, Sarubbi B, Santoro G, Grimaldi N, Correra A, Scognamiglio G, Russo MG, Calabrò R.

Source

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Abstract

Objectives: The aim of the present study was to evaluate the safety, tolerability, clinical and haemodynamic impact of add-on sildenafil in patients with congenital heart disease (CHD)-related pulmonary arterial hypertension (PAH) and Eisenmenger physiology after failure of oral bosentan therapy.

Methods: Thirty-two patients with CHD-related PAH (14 male, mean age 37.1 ± 13.7 years) treated with oral bosentan underwent right heart catheterization (RHC) for clinical worsening. After RHC, all patients received oral sildenafil 20mg thrice daily in addition to bosentan. Clinical status, resting transcutaneous oxygen saturation (SpO(2)), 6-minute walk test (6MWT), serology and RHC were assessed at baseline (before add-on sildenafil) and after 6 months of combination therapy.

Results: Twelve patients had ventricular septal defect, 8 atrio-ventricular canal, 6 single ventricle, and 6 atrial septal defect. Twenty-eight/32 had Eisenmenger physiology and 4 (all with atrial septal defect) did not. All patients well tolerated combination therapy. After 6 months of therapy, an improvement in clinical status (WHO functional class 2.1 ± 0.4 vs 2.9 ± 0.3; P=0.042), 6-minute walk distance (360 ± 51 vs 293 ± 68 m; P=0.005), SpO(2) at the end of the 6MWT (72 ± 10 vs 63 ± 15%; P=0.047), Borg score (2.9 ± 1.5 vs 4.4 ± 2.3; P=0.036), serology (pro-brain natriuretic peptide 303 ± 366 vs 760 ± 943 pg/ml; P=0.008) and haemodynamics (pulmonary blood flow 3.4 ± 1.0 vs 3.1 ± 1.2l/min/m(2), P=0.002; pulmonary vascular resistances index 19 ± 9 vs 24 ± 16 WU/m(2), P=0.003) was observed.

Conclusions: Addition of sildenafil in adult patients with CHD-related PAH and Eisenmenger syndrome after oral bosentan therapy failure is safe and well tolerated at 6-month follow-up, resulting in a significant improvement in clinical status, effort SpO(2), exercise tolerance and haemodynamics.