February, 2012

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In This Issue
Program Summary: ACC-ACHD 2012
22nd International Symposium on ACHD
Journal Watch

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ISACHD Newsletter

President's Message by Barbara J.M. Mulder
Dear ISACHD members,
Within one month we will have the opportunity to meet again at the ISACHD Spring meeting:

Sunday, March 25th, from 7-9 pm
Blackstone Renaissance Hotel
636 S. Michigan Avenue
Chicago, IL

Preliminary Agenda
* Welcome: Barbara Mulder
* Working groups
Global Health Care: Curt Daniels
Education: Erwin Oechslin
Research: K. Niwa, Ph. Moons
* Regional & nursing news: D. Fleck K Niwa, E. Oechslin, L. Alday, B. Davidson, H. Baumgartner
* Financial overview: K. Niwa
* Transition of presidency
* Upcoming meetings & other business
We will proceed with our discussions on global health, education, and global research. The working group chairmen will present the updates, and your input is more than welcome! The new ISACHD president, Curt Daniels, will be installed. Please be there to have a drink and toast with him!

Bill Davidson has made a summary for you of all ACHD sessions during the ACC meeting (see below, under Regional News). Bill: "The dedicated ACHD sessions are clearly marked, and there are many! Sunday is so rich we’ll need to wear our running shoes." Planning website is at the ACC website under Meetings/Plan Your ACC or at this link.

Jane Somerville will be giving the Dan McNamara lecture Sunday afternoon, speaking on "Fifty Years with Cardiac Surgeons." The session is at 2:30 PM, Sunday the 25th, and is titled "Legends in Cardiovascular Medicine-2012 Dan G. McNamara Lecture." The session will meet in McCormick Place North, N228. The international ACHD community should make a special effort to be there to honor Jane, our ACHD legend. Her talk will be followed by a panel discussion on lifelong care of the CHD patient.

The executives hope to welcome you all at the ISACHD meeting at 7 pm in Blackstone Renaissance hotel, in the very center of Chicago!

Barbara J.M. Mulder
President

Program Summary ACC-ACHD 2012 Chicago

By Bill Davidson

Mar 24, 2012
8:00AM - 9:30AM
### ACC Symposium

McCormick Place South, S105a  
10:30AM - 12:00PM

**ACC Oral Contributions**

**ACC Oral Contributions.901.From Targeting Errors to UNOS: How Quality and Databases Can Impact Clinical Care**  
McCormick Place South, S105a  
2:00PM - 3:30PM

**ACC Symposium**

**ACC Symposium.616.Quality, Safety and Resources Enhancing Pediatric Cardiovascular Care**  
McCormick Place North, N231  
4:30PM - 6:00PM

**ACC Symposium**

**ACC Symposium.625.Updates on Care in the Congenital Heart Disease Patients**  
McCormick Place North, N231  
Mar 25, 2012

**ACC Symposium**

**ACC Symposium.634.Preoperative Assessment of Patients on the Single Ventricle Pathway: Special Problems Creative Solutions**  
McCormick Place North, N231  
9:30AM - 10:30AM

### ACC Moderated Poster Contributions

**ACC Moderated Poster Contributions.1133.Congenital Cardiology Solutions: The Single Ventricle**  
McCormick Place South, Hall A

**ACC Moderated Poster Contributions.1135.Congenital Cardiology Solutions: Surgical Considerations**  
McCormick Place South, Hall A

**ACC Moderated Poster Contributions.1137.Congenital Cardiology**
Solutions: Contemporary Considerations from the Fetus to Adolescence
McCormick Place South, Hall A

ACC Moderated Poster Contributions.1139. Congenital Cardiology Solutions: Therapy
McCormick Place South, Hall A

ACC Moderated Poster Contributions.1140. Congenital Cardiology Solutions: Adult I
McCormick Place South, Hall A

ACC Moderated Poster Contributions.1142. Congenital Cardiology Solutions: Adult II
McCormick Place South, Hall A

10:45AM - 12:15PM
ACC Symposium
ACC Symposium.637. Post-Operative Glenns and Fontans: Navigating Turbulent Waters
McCormick Place North, N231

ACC Oral Contributions
ACC Oral Contributions.920. Adults with Congenital Heart Disease: Imaging Predictors, Evolving Therapies and Outcomes
McCormick Place South, S402

11:00AM - 12:00PM
ACC Moderated Poster Contributions
ACC Moderated Poster Contributions.1134. Congenital Cardiology Solutions: Roles of Imaging
McCormick Place South, Hall A

ACC Moderated Poster Contributions.1136. Congenital Cardiology Solutions: Interventional Solutions
McCormick Place South, Hall A

ACC Moderated Poster Contributions.1138. Congenital Cardiology Solutions: It's Not All Congenital
McCormick Place South, Hall A

ACC Moderated Poster Contributions.1141. Congenital Cardiology
Solutions: Adult III
McCormick Place South, Hall A

ACC Moderated Poster Contributions.1143.Congenital Cardiology
Solutions: Adult IV
McCormick Place South, Hall A

12:30PM - 1:45PM
ACC Special Session
ACC Special Session.301.Career & Mentoring Session for Pediatric and Congenital Cardiologists
McCormick Place North, N231

2:00PM - 3:30PM
Legends in Cardiovascular Medicine
Legends in Cardiovascular Medicine.402.Legends in Cardiovascular Medicine -- 2012 Dan G. McNamara Lecture
McCormick Place North, N228

4:30PM - 6:00PM
ACC Symposium
ACC Symposium.667.The Great Debates
McCormick Place North, N231

ACC Symposium.668.ACHD for the General Cardiologist
McCormick Place North, N230

Mar 26, 2012
8:00AM - 9:30AM
ACC Symposium
ACC Symposium.678.Pulmonary Artery Stenosis Current Therapy and Future Directions: Live Case
McCormick Place North, N231

ACC Symposium.679.Women with Congenital Heart Disease: Fertility, Pregnancy and Menopause
McCormick Place South, S501a

10:30AM - 12:00PM
ACC Symposium
ACC Symposium.692.Pulmonary Vein Stenosis: What Are Realistic Expectations?  
McCormick Place North, N229

ACC Symposium.693.Challenging Imaging Issues in Congenital Heart Disease  
McCormick Place North, N231

12:15PM - 1:45PM  
ACC International Lunch Symposium  
ACC International Lunch Symposium.506.Joint Session of the Egyptian Society of Cardiology and the American College of Cardiology: Adult Congenital Heart Disease: Where We Came From and Where We Are  
McCormick Place North, N231

2:00PM - 3:30PM  
ACC Meet the Experts  
ACC Meet the Experts.247.Acquired Heart Disease in Childhood Epidemiology: Current and Future Management  
McCormick Place South, S501a

3:45PM - 5:15PM  
ACC Meet the Experts  
ACC Meet the Experts.252.Golden Moments: When Is the Right Time to Intervene in Congenital Heart Disease?  
McCormick Place South, S501a

Mar 27, 2012  
8:00AM - 9:30AM  
ACC Symposium  
ACC Symposium.724.Complex Conotruncal Malformations: What the Surgeon Needs to Know and How to Get the Information  
McCormick Place North, N229

ACC Symposium.725.Coming of Age: The HLHS Turns 21  
McCormick Place North, N231

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 Canada
Erwin Oechslin, Toronto, Canada

22nd ACHD Symposium in Toronto:
Website is open - Register and Submit Your Abstract!
ACHD Meeting Toronto 2012 or www.uhn.ca/ACHDConference2012.asp

It is my great pleasure to invite you to the 22nd Annual International Symposium on Adult Congenital Heart Disease, which will be held at the Toronto Marriott Downtown Eaton Centre Hotel from May 30 - June 2, 2012. This Symposium, organized in partnership with the Oregon Health & Science University in Portland (Oregon), Cincinnati Children’s Hospital Medical Centre, University of Cincinnati (Ohio) and University of Toronto, is a unique forum where medical professionals can meet their colleagues from around the world, and exchange ideas and information in the field of Adult Congenital Heart Disease.

'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 such as heart failure, arrhythmias, the Fontan disease, pulmonary arterial hypertension and others, 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': Quality of Death and Dying Experience) and a Workshop (Advanced Care Planning in Adult Congenital Heart Disease) will cover this hot topic amongst many others.

I expect the Symposium to provide the best of science and education in this area of practice. The program is designed for all healthcare professionals working with Adult Congenital Heart Disease (ACHD) and should familiarize participants with the latest clinical information on the diagnosis and management for ACHD. Nursing and Congenital Cardiac Care Associates will also have a major educational role within this program.

Please visit ACHD Meeting Toronto 2012 or www.uhn.ca/ACHDConference2012.asp to get more information.

Information at a glance:
Abstract: Submission is open. Submission Deadline: March 19, 2012 (6 am EST)

* Abstracts will be considered for all aspects of care for adults with congenital heart disease (ACHD), including cardiac issues in the inpatient and outpatient areas, ACHD nursing, surgical, anesthesia and perfusion research, interventional procedures, and diagnostic testing. The Faculty Members will select the accepted abstracts and notify the submitting author of acceptance by April 8, 2012.

Work that has been previously presented at local, national or international meetings will be considered for presentation at the Toronto Symposium. However, material that has been already published as a manuscript in a peer-reviewed journal will not be considered.

* The top three abstracts will be presented as oral presentations on May 31 and June 1, 2012. Winner of the best abstracts in the Nursing and Physician section will get the Investigator Award and free access to the 23rd International Symposium at Skamania Lodge, Stevenson, WA (USA) in 2013.

Registration:

* Early Registration until April 15, 2012. Special Rates for Paying ISACHD and/or CACH Network Members

Registration Fees:
Early Registration (until April 15, 2012)
* Physicians $850.00 CDN
* Trainees, Nurse, Cardiac Care Associates $650.00 CDN

Special Rate for Paying ISACHD and/or CACH Network Members (until April 15, 2012)
* Physicians $765.00 CDN
* Trainees, Nurses, Cardiac Care Associates $585.00 CDN

Hotel Reservation:
* Special room rates are available until April 30, 2012: single and double room for only $189.00 CDN.
* Online Registration is available at ACHD Meeting Toronto 2012 or www.uhn.ca/ACHDConference2012.asp

Please feel free to contact Carole Ryan at carole.ryan@uhn.ca if you have any questions.
WG on Education
Erwin Oechslin
Chair, WG on Education, Toronto (Canada)

Els Pieper, UMS Groningen (NL), has the final version of the program for a Web-based, 2-day teaching course for trainees with a background in adult cardiology. We have to assign speakers to the different topics, which we set as a goal for the ACC meeting in Chicago. There has been some delay from my end because of the organization of the Toronto meeting.

Under the leadership of Dr. Gary Webb, Cincinnati, we are collecting information on 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:
Please be proactive and contact us if you want to endorse your ACHD meeting.

The following meetings have been endorsed:
* NEW: 3rd Congress of the Asia Pacific Society for Adult Congenital Heart Disease (APSACHD), in Taipei, Taiwan, April 4-7, 2012 (http://www.appcs2012.org)
* 3rd European Meeting: Adult Congenital Heart Disease, German Heart Centre, Munich; March 16-17, 2012 (www.isachd.org)
* 2nd International Congress on Cardiac Problems in Pregnancy, Berlin, Germany; May 17-20, 2012 (www.cppcongress.com)

Journal Watch
ARTICLE OF THE MONTH: February 2012
Commentary on the article "Functional health status in adult survivors of operative repair of Tetralogy of Fallot" by Dounya Schoormans

Hickey et al conducted a retrospective study examining late functional status of 840 adult survivors of operative repair of Tetralogy of Fallot (TOF). The objectives of this study were (a) to examine clinical follow-up (i.e. number of re-operations and experienced symptoms) after reaching adulthood, (b) to compare patients' functional status to a gender- and age matched reference group, and (c) to relate patient-specific features to patients' functional status. All 840 patients were clinically follow-up by means of either a chart review (16%) or a telephone interview on clinical history and experienced symptoms (84%). In addition, functional status was assessed in 396 patients (47%) by the Short Form-36 (SF-36).

Clinical follow-up
Of the 840 adult patients with TOF, 68 patients died (8.1%). Almost 15% of patients were re-operated. At the ages 30, 40 and 50 years respectively, 90 ± 1%, 79 ± 2%, and 62 ± 4% were free from re-interventions. Factors associated with re-operation were operating details from the first correctional surgery (i.e. use of conduct or patch to the right ventricular outflow tract that was not transannular, and duration of cardiopulmonary bypass) and the presence of a right aortic arch. Results of the follow-up interview (n=706) showed that cardiorespiratory symptoms were present in 45% of patients. Palpitations (27%), exertional dyspnea (21%), and chest pain were the most common symptoms.

Functional status
Results showed that patients were impaired on the physical functional status domains (i.e. physical functioning, role physical and general health) when compared to a gender- and age matched reference group. In contrast, for the mental domains, patients were only impaired regarding the vitality domain. Older age, the presence of coexisting syndromes or cardiac abnormalities, having undergone cardiac re-operations, and the presence of cardiorespiratory symptoms were associated with impairments in physical functional status domains. Differences in TOF types were not related to impairments in functional status. More in detail, lower general health status scores were associated with an increased incidence in re-operations.

The current study concludes that although adult patients with TOF have excellent survival prospects, their physical functional status is impaired. Mixed results have been reported on this topic. Van Rijen and colleagues report similar results concluding that patients with TOF have a physically impaired functional status.[1] On the contrary, other research groups conclude that TOF-patients have a good functional status compared to the general population.[2] This is in line with a study...
in patients with various congenital heart defects who show no impairments in functional status.[3] Overall, the level of impairment increases with age, suggesting that TOF is a progressive illness, which is in line with studies relating age to functional status in patients with various congenital heart diseases.[4] Given the found negative relation between cardiorespiratory symptoms and functional status, treatment of cardiorespiratory symptoms is of utmost importance when improving patients' physical functioning. In depth studies examining patients' symptomatology and the relative impact of specific symptoms on functional status is of paramount importance to guide treatment strategies aiming to improve patients' functional status.


Commentary: Dounya Schoormans, MPhil
PhD student
Academic Medical Center
Dept. of Cardiology and Dept. of Medical Psychology
Meibergdreef 9
1105 AZ Amsterdam, The Netherlands

Am J Cardiol. 2012 Jan 12. [Epub ahead of print]

Functional Health Status in Adult Survivors of Operative Repair of Tetralogy of Fallot.

Source
Division of Cardiovascular Surgery, Department of Surgery, University of Toronto, Hospital for Sick Children, Toronto, Ontario, Canada.

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.

Pediatr Cardiol. 2012 Feb 10. [Epub ahead of print]
Risk Factors Associated With Morbidity and Mortality After Pulmonary Valve Replacement in Adult Patients With Previously Corrected Tetralogy of Fallot.
Source
Department of Cardiothoracic Surgery, Emory University School of Medicine, Atlanta, GA, USA.
Abstract
Patients with palliated tetralogy of Fallot (TOF) often require pulmonary valve replacement in adulthood, yet the data regarding their outcomes are scarce. This study aimed to identify risk factors associated with postoperative complications in these patients and to establish long-term survival data for this patient group. A retrospective cohort study investigated 153 consecutive patients with a history of TOF repair who underwent pulmonary valve replacement at a single large academic center between March 1996 and March 2010. In part 1 of the study, logistic models were constructed to assess demographic, medical, and surgical risk factors for operative mortality; occurrence of a major adverse event (stroke, renal failure, prolonged ventilation, deep sternal infection, reoperation, or operative mortality); and prolonged hospital stay (>7 days). Risk factors with a p value less than 0.10 by univariate
analysis were included in the subsequent multivariate analysis. In part 2 of the study, long-term, all-cause mortality was determined by construction of a Kaplan-Meier curve for the cohort. Seven patients died (4.5%). Significant risk factors for mortality in the multivariable analysis included age older than 40 years (odds ratio (OR) 9.89) and concomitant surgery (OR 6.65). A major adverse event occurred for 22 patients (14.4%). The only significant risk factor in the multivariable analysis for an adverse event was concomitant surgery (OR 6.42). The hospital stay was longer than 7 days for 31 patients (20.3%). The significant risk factors for a prolonged hospital stay included the presence of preoperative arrhythmias (OR 4.17), New York Heart Association class 3 (OR 4.35), and again, concomitant surgery (OR 4.2). Among the 146 hospital survivors, only 5 patients died in the intervening period. The predicted survival rates were 98.5% at 1 year, 96.7% at 5 years, and 93.5% at 10 years. Pulmonary valve replacement in adults with palliated TOF is a safe procedure with excellent long-term survival, but there remain important risk factors for postoperative mortality, prolonged hospital stay, and major adverse events. Awareness and modification of important risk factors may help to improve outcomes.

Pediatr Cardiol. 2012 Feb 9. [Epub ahead of print]

Sudden Cardiac Death and Malignant Arrhythmias: The Scope of the Problem in Adult Congenital Heart Patients.
Perry JC.

Source
Electrophysiology and Adult Congenital Heart Programs, UCSD/Rady Children's Hospital San Diego, San Diego, CA, USA, jerry@rchsd.org.

Abstract
A key component of recognizing sudden cardiac death (SCD) risk in the adult congenital heart disease (ACHD) patient is the recognition of heart failure risk for each physiology. The risk of SCD is an accrued phenomenon, representing the influences of anatomy, genetics, surgical and catheter interventions, and long-term sequelae of residual hemodynamic issues. These all lead to a substrate for tachyarrhythmia. It is beneficial in thinking about all of the potential combinations of CHD anatomy and physiologies to categorize SCD risk for the ACHD patient in terms of systemic left-ventricular failure, systemic right-ventricular failure, subpulmonary ventricular failure, the dyssynchronous contractility states due to bundle branch block, and single-site ventricular pacing. This article reviews important issues in arrhythmogenesis for ACHD patients with all of these physiologies and discusses potential cardiac rhythm device-management needs.

Pediatr Cardiol. 2012 Feb 7. [Epub ahead of print]

Sex and Age Differences in Body-Image, Self-Esteem, and Body Mass
Index in Adolescents and Adults After Single-Ventricle Palliation.

Pike NA, Evangelista LS, Doering LV, Eastwood JA, Lewis AB, Child JS.

Source
School of Nursing, University of California, Factor Building Room 3-938, Los Angeles, CA, 90095-6919, USA, npike@sonnet.ucla.edu.

Abstract
Single-ventricle congenital heart disease (SVCHD) requires multiple palliative surgical procedures that leave visible surgical scars and physical deficits, which can alter body-image and self-esteem. This study aimed to compare sex and age differences in body-image, self-esteem, and body mass index (BMI) in adolescents and adults with SVCHD after surgical palliation with those of a healthy control group. Using a comparative, cross-sectional design, 54 adolescent and adult (26 male and 28 female) patients, age 15-50 years, with SVCHD were compared with 66 age-matched healthy controls. Body-image and self-esteem were measured using the Multidimensional Body-Self Relations Questionnaire-Appearance Scale and Rosenberg Self-Esteem Scale. Height and weight were collected from retrospective chart review, and BMI was calculated. Female adolescents and adult patients with SVCHD reported lower body image compared with male patients with SVCHD and healthy controls (p = 0.003). Specific areas of concern were face (p = 0.002), upper torso or chest (p = 0.002), and muscle tone (p = 0.001). Patients with SVCHD who were <21 years of age had lower body image compared with healthy controls (p = 0.006). Self-esteem was comparable for both patients with SVCHD and healthy peers. There were no sex differences in BMI; BMI was higher in subjects >21 years of age (p = 0.01). Despite the similarities observed in self-esteem between the two groups, female patients with SVCHD <21 years of age reported lower perceived body-image. Our findings support the need to recognize poor psychological adjustment related to low self-esteem in patients with SVCHD; female patients warrant increased scrutiny. Strategies to help patients with SVCHD cope with nonmodifiable aspects of body-image during the difficult adolescent-to-young adult years may potentially enhance self-esteem and decrease psychological distress.
Real-time three-dimensional (3D) echocardiography allows us to measure right ventricular (RV) end-diastolic volume irrespective of its shape. Tissue Doppler imaging (TDI) and speckle tracking imaging (STI) are new tools to assess myocardial function. We sought to evaluate RV function by 3D echocardiography and myocardial strain imaging in adult patients with atrial septal defect (ASD) before and 6 months after transcatheter closure in order to assess the utility of these new indexes in comparison with standard two-dimensional (2D) and Doppler parameters. Thirty-nine ASD patients and 39 healthy age- and sex-matched controls were studied using a commercially available cardiovascular ultrasound system. 2D-Doppler parameters of RV function (fractional area change, tricuspid annular plane systolic excursion, myocardial performance index) were calculated. 3D RV volumes were also obtained. RV peak-systolic velocities, peak-systolic strain, and peak systolic and diastolic strain-rate were measured in the basal, mid and apical segments of lateral and septal walls in apical 4-chamber view by TDI and STI. In open ASD, RV ejection fraction (3D-RVEF) and global and regional RV longitudinal strain were significantly higher than control group and decreased significantly after closure. By multivariate analysis 3D-RVEF, apical strain and strain rate were independent predictors of functional class. ROC analysis showed 3D-RVEF and apical strain to be more sensitive predictors of unfavorable outcome after defect closure compared to 2D-Doppler indexes. 3D echocardiography and myocardial strain imaging give useful insights in the quantitative assessment of RV function in ASD patients before and after closure.


Totally endoscopic robotic ventricular septal defect repair in the adult.

Source
Department of Cardiovascular Surgery, People’s Liberation Army General Hospital, Beijing, China.

Abstract

Objective: We have previously reported total endoscopic ventricular septal defect repair in the adult using the da Vinci S Surgical System. The optimal results encouraged us to extend the use of this technology to more complicated patients with ventricular septal defect.

Methods: From January 2009 to July 2010, 20 patients underwent total endoscopic robotic ventricular septal defect repair. The average patient age was 29.0 ± 9.5 years (range, 16-45). Of the 20 patients, 9 were female and 11 were male. The echocardiogram demonstrated that the average diameter of the ventricular septal defect was 6.1 ± 2.8 mm (range, 2-15), and 4 patients had concomitant patent foramen ovale. Ventricular septal defect closure was directly secured with interrupted mattress sutures in 14 patients and patched in 6 patients. All the procedures were completed using the da Vinci robot by way of 3 port
incisions and a 2.0- to 2.5-cm working port in the right side of the chest.

**Results:** All patients were operated on successfully. The mean cardiopulmonary bypass and mean crossclamp time was 94.3 ± 26.3 minutes (range, 70-140) and 39.1 ± 12.9 minutes (range, 22-75), respectively. The mean operation time was 225.0 ± 34.8 minutes (range, 180-300). The postoperative transesophageal echocardiogram demonstrated an intact ventricular septum. No residual left-to-right shunting and no permanently complete atrioventricular dissociation was found postoperatively. The mean hospital stay was 5 days. No residual shunt was found during a mean follow-up of 7 months (range, 1-22). The patients returned to normal function within 1 week without any complications.

**Conclusions:** Total endoscopic robotic ventricular septal defect repair in adult patients is feasible, safe, and efficacious.


**Endovascular Repair of Aortic Isthmus Coarctation With a Self-Expanding Covered Stent.**

Oberhuber A, Muehling BM, Orend KH, Schelzig H.

**Source**

Department of Thoracic and Vascular Surgery, University of Ulm, Ulm, Germany.

**Abstract**

**Background:** Coarctation is one of the most often seen congenital aortal defects. In the majority, diagnosis will be made in newborns. Endovascular repair is critical in children owing to their growth, but in adult patients, it is an interesting alternative.

**Methods:** A 31-year-old man presenting with hypertension of upper extremities and pulseless lower extremities was admitted to our hospital. Systolic blood pressure was 190 mm Hg, although a triple antihypertensive medication was administered. Computed tomographic angiography showed a nearly total occlusion of the aortic isthmus. Coarctation was treated by an endovascular approach with a self-expanding covered stent-graft (Medtronic Talent; Medtronic World Medical, Sunrise, FL) after predilatation with a Reliant balloon (Medtronic World Medical, Sunrise, FL).

**Results:** After a follow-up of 40 months, the patient is normotensive and antihypertensive medication could be reduced. Lower extremities showed normal pulses and normal ankle-brachial index. Computed tomographic scans showed unchanged stent-graft position, with constant slight waist.

**Discussion:** Endovascular repair of atresia or coarctation of the thoracic aorta is a minimal invasive debatable option. Not only reduction of blood pressure but also reduction of left ventricular mass and prolongation of life expectancy can be achieved by endovascular treatment.
Surgical outcome of partial Shone complex.


Source
Cardiovascular and Thoracic Department, San Raffaele Scientific Institute and Università Vita-Salute San Raffaele, Milan, Italy.

Abstract
Partial forms of Shone complex are rare. Surgical outcomes of the complete forms have generally been poor, whereas there is a lack of data on long-term follow-up of surgically treated adult partial complex. Between 2001 and 2011, nine patients (age: 38 ± 8 years; six males, 67%) were referred for valvular heart disease. Transthoracic and transoesophageal echocardiography was performed. Data were confirmed by intra-operative findings and reports. Patients were diagnosed as partial Shone complex and presented with mitral stenosis (MS) (45%) or mitral regurgitation (22%) or aortic regurgitation (22%). All but one patient (89%) reported previous surgery: coarctation of the aorta repair (87.5%) and aortic valvulotomy (12.5%). Redo intervention included: mitral valve replacement (25%), mitral repair (25%), aortic valve replacement (37.5%) and subvalvular aortic ridge resection (25%). One patient refused surgery. Patients surgically treated before the age of 5 (87.5%) showed favourable outcome (survival rate: 100%) and a 23.6 (± 4.6)-year follow-up free from events. The patient who underwent first intervention at the age of 50 and the patient with MS who refused surgery showed a 45 (± 7)-year follow-up free from major morbidity. Patients with partial Shone complex, properly diagnosed and treated, show favourable surgical outcome free from major clinical events.

Evaluation of the adolescent or adult with some features of Marfan syndrome.

Pyeritz RE.

Source
Perelman School of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania, USA.

Abstract
Disclaimer: ACMG standards and guidelines are designed primarily as an educational resource for medical geneticists and other health care providers to help them provide quality medical genetic services. Adherence to these standards and guidelines does not necessarily ensure a successful medical outcome. These standards and guidelines should not be considered inclusive of all proper procedures and tests or
exclusive of other procedures and tests that are reasonably directed to obtaining the same results. In determining the propriety of any specific procedure or test, the geneticists should apply their own professional judgment to the specific clinical circumstances presented by the individual patient or specimen. It may be prudent, however, to document in the patient's record the rationale for any significant deviation from these standards and guidelines. Individuals who are suspected of having Marfan syndrome are often referred to a medical geneticist for further evaluation and diagnosis. However, there are a number of conditions that share physical manifestations with those of Marfan syndrome; therefore, an approach to diagnosis and evaluation is crucial to the proper long-term follow-up of these individuals. This practice guideline provides guidance for the approach to this cadre of individuals. Genet Med 2012;14(1):171-177.


**Tricuspid Valve Replacement through a Left Atriotomy and Transseptal Approach in a Congenitally Malformed Heart.**

Kogon BE, Grudziak J, McConnell M, Book WM.

**Source**

Departments of Cardiothoracic Surgery Cardiology and Sibley Cardiology, Emory University, Atlanta, Ga, USA.

**Abstract**

More and more children with congenital heart disease are surviving into adulthood. These patients are forcing adult congenital cardiac surgeons to develop innovative approaches to correct their complex anatomy and physiology. This report describes a patient with a congenitally malformed heart necessitating a novel approach to access the tricuspid valve—a left atriotomy and transseptal incision. Three-dimensional preoperative imaging allowed for successful surgical planning.


**The adult congenital and pediatric cardiology section increasing the opportunities for the congenital heart disease community within the american college of cardiology.**

Martin GR, Mitchell S, Beekman RH 3rd, Feinstein JA, Jenkins KJ, Landzberg M, Webb G.

**Source**

Children's National Medical Center, Washington, DC.

**Abstract**

The Adult Congenital and Pediatric Cardiology (AC/PC) Section was established to develop a clear voice within the American College of Cardiology and address the myriad issues facing the congenital heart
disease profession. The Section is governed by the AC/PC Council, which includes pediatric cardiologists, adult congenital cardiologists, a cardiac care associate, and a fellow-in-training member. The Council is responsible for bidirectional communication between the College’s Board of Trustees and the AC/PC Section members. Since its founding in 2004, Section objectives have been defined by the College's mission: to advocate for quality cardiovascular care through education, research promotion, and the development and application of standards and guidelines and to influence health care policy. The pillars of the College-advocacy, quality, education, and member engagement-serve as the defining template for the Section's strategy. The Section has developed work groups in advocacy, clinical practice, education and training, quality, and publications. A separate leadership group has been developed for adult congenital heart disease. Work groups are open to all Section members. Recognition of the importance of lifelong care in congenital heart disease led Section leaders to incorporate pediatric cardiology and adult congenital heart disease content into each of the work groups. There are more than 1,200 Section members, with nearly 400 members actively contributing to Section activities. This article outlines Section efforts to date and highlights significant successes to date.

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Source
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Abstract
Background: The Society of Thoracic Surgeons (STS) and the American Association for Thoracic Surgery (AATS) have intermittently surveyed their combined membership. These manpower surveys have provided snapshots of thoracic surgery, documenting practice changes over time. At this critical time in US health care reform the physician workforce is of critical importance. This survey updates the data obtained from the 2000 and 2005 surveys.

Methods: The survey instrument was updated from the 2005 survey. It was received by 5,265 surgeon members of the STS/AATS during November and December 2009. There was a superb 50% return rate. The data were entered into a comprehensive database. Perception Solutions, Inc, independently performed the analysis.

Results: The median age of the active US thoracic surgeons is 52.9 years. Women comprise 3.4% of adult cardiac, 5.2% of congenital heart, and 7.9% of general thoracic surgeons. The decision to pursue a career in thoracic surgery was made before or in medical school by 45.3% of surgeons. The majority of survey respondents had a mean of 8.7 years
of residency training after medical school graduation. The cumulative average educational debt was $56,000. Overall career satisfaction was 46% (very or extremely satisfied). Database participation was 84%. Operative volume over the past 12 months decreased for 30% of surgeons. Malpractice premiums have steadily increased over the past 5 years from $55,947 to $59,673. The number of additional years the currently active US cardiothoracic surgeon plans to practice is 12.6 years. Therefore, the projected retirement age of the thoracic surgery workforce will be 65. This is consistent among all surgeons: adult cardiac, 66 years; congenital heart, 65 years; and general thoracic, 67 years.

**Conclusions:** These data give a clear profile of the specialty at this time. The major challenges remain length of training and educational debt of the thoracic surgeon. Case volume, scope of practice, malpractice costs, and career satisfaction remain major elements to provide a positive environment to recruit new surgeons in to the specialty. The resident pool has contracted while the workforce ages and retirement looms. Significant shortages may develop as the US population ages in the environment of health care reform.


**Comprehensive use of cardiopulmonary exercise testing identifies adults with congenital heart disease at increased mortality risk in the medium term.**


**Source**

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**Abstract**

**Background:** Parameters of cardiopulmonary exercise testing were recently identified as strong predictors of mortality in adults with congenital heart disease. We hypothesized that combinations of cardiopulmonary exercise testing parameters may provide optimal prognostic information on midterm survival in this population.

**Methods and Results:** A total of 1375 consecutive adult patients with congenital heart disease (age, 33±13 years) underwent cardiopulmonary exercise testing at a single center over a period of 10 years. Peak oxygen consumption (peak $\text{Vo}(2)$), ventilation per unit of carbon dioxide production ($\text{Ve/Vco}(2)$ slope), and heart rate reserve were measured. During a median follow-up of 5.8 years, 117 patients died. Peak $\text{Vo}(2)$, heart rate reserve, and $\text{Ve/Vco}(2)$ slope were related to midterm survival in adult patients with congenital heart disease. Risk of death increased with lower peak $\text{Vo}(2)$ and heart rate reserve. A higher $\text{Ve/Vco}(2)$ slope was also related to increased risk of death in...
noncyanotic patients, whereas the Ve/Vco(2) slope was not predictive of mortality in cyanotic patients. The combination of peak Vo(2) and heart rate reserve provided the greatest predictive information after adjustment for clinical parameters such as negative chronotropic agents, age, and presence of cyanosis. However, the incremental value of these exercise parameters was reduced in patients with peak respiratory exchange ratio <1.0.

**Conclusions:** Cardiopulmonary exercise testing provides strong prognostic information in adult patients with congenital heart disease. Prognostication should be approached differently, depending on the presence of cyanosis, use of rate-lowering medications, and achieved level of exercise. We provide 5-year survival prospects based on cardiopulmonary exercise testing parameters in this growing population.


**Coronary artery disease in adult congenital heart disease: outcome after coronary artery bypass grafting.**

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**Source**
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**Abstract**

**Background:** Atherosclerotic coronary artery disease may be seen during repair of adult congenital heart disease (ACHD). There are few data outlining outcomes of concomitant coronary artery bypass grafting (CABG) in these patients.

**Methods:** Between February 1972 and August 2009, 122 patients (77 men) underwent concomitant CABG at the time of ACHD repair; median age was 64 years (range 40 to 85 years). Thirty patients (25%) had preoperative angina, 7 patients (6%) had previous myocardial infarction (MI), and 6 patients (5%) had previous percutaneous intervention. Most common primary cardiac diagnoses included secundum atrial septal defect (ASD) in 73 patients (60%), Ebstein’s anomaly in 14 patients (11%), and partial anomalous pulmonary venous connection in 8 patients (7%).

**Results:** Operations included ASD repair in 78 patients (64%), tricuspid/pulmonary valve procedures in 23 patients (19%), and ventricular septal defect repair in 10 patients (8%). One bypass graft procedure was performed in 69 patients (57%), 2 bypass graft procedures were performed in 32 patients (26%), 3 bypass graft procedures were performed in 14 patients (11%), 4 bypass graft procedures were performed in 5 patients (4%), and 5 bypass graft procedures were performed in 2 patients (2%). There were 4 early deaths (3.3%). During a median follow-up of 6 years (maximum follow-up, 32 years), actuarial survival was 76% at 5 years and 56% at 10 years. In patients with left anterior descending (LAD) artery disease,
survival was higher when a left internal mammary graft (LIMA) was used (5 years, 86% versus 66%; 10 years, 66% versus 36%; p < 0.05).

**Conclusions:** Concomitant CABG may be required at the time of correction of ACHD. Survival is higher when a LIMA graft is used, and late functional outcome is good, with a low incidence of late angina and need for reintervention.


**Maternal cardiovascular events during childbirth among women with congenital heart disease.**


**Source**

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**Abstract**

**Objectives:** To define the epidemiology of adverse cardiovascular events among women with congenital heart disease (CHD) hospitalised for childbirth in the USA. Design and setting The 1998-2007 Nationwide Inpatient Sample, an administrative dataset representative of overall US hospital admissions, was used to identify hospitalisations for delivery.

**Main outcome measures:** Logistic regression was used to estimate ORs for cardiovascular outcomes (arrhythmia, heart failure, cerebrovascular accident, embolism, death or a combined outcome) for women with and without CHD. Covariates included age, number of medical comorbidites, pulmonary hypertension, hospital teaching status, insurance status and method of delivery.

**Results:** Annual deliveries for women with CHD increased 34.9% from 1998 to 2007 compared with an increase of 21.3% in the general population. Women with CHD were more likely to sustain a cardiovascular event (4042/100,000 vs 278/100,000 deliveries, univariate OR 15.1, 95% CI 13.1 to 17.4, multivariable OR 8.4, 95% CI 7.0 to 10.0). Arrhythmia, the most common cardiovascular event, was more frequent among women with CHD (2637/100,000 vs 210/100,000, univariate OR 12.9, 95% CI 10.9 to 15.3, multivariable OR 8.3, 95% CI 6.7 to 10.1). Death occurred in 150/100,000 patients with CHD compared with 8.2/100,000 patients without CHD (multivariable OR 6.7, 95% CI 2.9 to 15.4). Complex CHD was associated with greater odds of having an adverse cardiovascular event than simple CHD (8158/100,000 vs 3166/100,000, multivariable OR 2.0, 95% CI 1.4 to 3.0).

**Conclusions:** Maternal CHD is associated with a markedly increased risk of adverse cardiovascular events and death during admission for delivery.
Independent risk factors for cardiac operations in adults with congenital heart disease: a retrospective study of 543 operations for 500 patients.


Source
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Abstract
Adults with congenital heart disease (CHD) are an increasing population requiring cardiac operations. To date, the perioperative risk factors for this group have not been identified. This study aimed to identify clinical, morphologic, and hemodynamic risk factors for an adverse outcome. This study retrospectively analyzed a cohort of 500 patients (ages >16 years) who underwent 543 operations between January 2004 and December 2008 at a single center. The composite end point of an adverse outcome was in-hospital death, a prolonged intensive care exceeding 4 days, or both. The composite end point was reached by 253 of the patients (50.6%). Of the 500 patients, 13 (2.6%) died within 30 days after the operation. After logistic regression analysis, the following eight items remained significant: male gender (P = 0.003; odds ratio [OR] 1.8; 95% confidence interval [CI] 1.2-2.6), cyanosis (P > 0.006; OR 3.7; 95% CI 1.5-9.4), functional class exceeding 2 (P = 0.004; OR 2.2; 95% CI 1.3-3.7), chromosomal abnormalities (P = 0.004; OR 3.3; 95% CI 1.4-7.7), impaired renal function (P = 0.019; OR 3.8; 95% CI 1.2-11.5), systemic right ventricle (RV) in a biventricular circulation (P = 0.027; OR 3.3; 95% CI 1.1-9.5), enlargement of the systemic ventricle (P = 0.011; OR 1.7; 95% CI 1.1-2.6), and operation with extracorporeal circulation (P = 0.002; OR 4.3; 95% CI 1.7-11.4).

Early mortality in the current adult CHD population is low. Morbidity, however, is significant and influenced by the patients' conditions (male gender, chromosomal abnormalities), history (cyanosis, New York Hospital Association [NYHA] class), and underlying morphology (systemic RV). This information for a large cohort of patients could help progress toward more adequate counseling for adults with a congenital heart defect.

Safety and efficiency of chronic ACE inhibition in symptomatic heart failure patients with a systemic right ventricle.

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Source
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Abstract

**Background:** ACE inhibition is an established treatment regimen in patients with congestive heart failure due to left ventricular dysfunction which improves morbidity and mortality. However, little is known about the beneficial effects of ACE inhibition in adult patients after Mustard procedure for transposition of the great arteries with heart failure symptoms. Therefore, we investigated the effects of ACE inhibition in these patients on heart failure symptoms, echocardiographic diameters, NT-proBNP and exercise capacity.

**Methods:** In 14 patients (age 25.2 ± 3.5 years), after Mustard procedure for transposition of the great arteries (age at operation 1.1 ± 1.3 years) with heart failure NYHA II (New York Heart Association class), an ACE inhibition was initiated. At baseline and 13.3 ± 4.0 months after treatment with enalapril (10mg twice a day), echocardiography, exercise test and NT-proBNP measurements were performed and compared to an age- and sex-matched control group.

**Results:** Maximum oxygen uptake and echocardiographic parameters did not change significantly in both groups. However, NT-proBNP showed a significant decrease in the treatment group (242 ± 105 vs. 151 ± 93 ng/l, p=0.004), while in the control group a significant increase (120 ± 89 vs. 173 ± 149 ng/l, p<0.05) was observed. Furthermore, ACE inhibitor treatment did not result in a deterioration of heart failure symptoms or renal function.

**Conclusions:** Thus, ACE inhibitor treatment of heart failure symptoms in patients with a systemic right ventricle is safe and reduces NT-proBNP levels significantly as a marker for ventricular overload. Nevertheless, larger scale trials are warranted to show effects on morbidity and mortality in this highly selected patient group.

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