

November, 2011

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International Society for Adult Congenital Heart Disease

## ISACHD Newsletter

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### President's Message

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



From November 12th to 16th the AHA Scientific Sessions take place in Orlando, Florida. We'll have our ISACHD meeting on Sunday, November 13th, 6:30-9pm, in Salon 13, at the Rosen Plaza Hotel, 9700 International Drive, Orlando, Florida 32819.

The ISACHD executives hope to welcome you all!



### Agenda

Welcome

Barbara Mulder

Working groups

- Global Health Care Curt Daniels
- Education Erwin Oechslin
- Research and Journal Watch Koichiro Niwa, Philip Moons

Break-out working groups for discussion (30 min), followed by report of plans

Regional + Nursing News

Desiree Fleck, Luis Alday, Koichiro Niwa, Erwin Oechslin

Upcoming Meetings

GUCH Training Course, Munich, March, 2012  
3rd Congress - Asia Pacific Soc for Adult CHD, Taipei, Taiwan, April, 2012  
22st ACHD Symposium, Toronto, May, 2012

Other business

### AHA Congress

A rough summary of the scientific program on congenital heart disease at the AHA Congress is shown below. It shows some very interesting sessions, like

Muenster Germany	extracardiac manifestations of the Fontan circulation, what to do with asymptomatic coronary artery abnormalities, a session on cardiac fibrosis, biomedical engineering insights into congenital heart disease and heart failure in congenital heart disease.
<b>Representative, Asia-Pacific</b>	
Koichiro Niwa, M.D. Chiba Cardiovascular Center Chiba, Japan	<b><u>ACHD Activities at AHA 2011, by Curt Daniels</u></b> <b><u>Sunday</u></b> Sun 11/13 5:30 PM- 6:45 PM, room W203c <b>CVS.302.Extracardiac Manifestations of the Fontan Physiology</b>
<b>Representative, Canada</b>	
Dr. Erwin N Oechslin Toronto General Hospital Toronto, Ontario, Canada	Hideo Ohuchi, Osaka, Japan: Liver, Lipids, and Glucose Manifestations Gruschen R Veldtman, Southampton, United Kingdom; Intestinal Manifestations William Mahle, Atlanta, GA: Vascular Manifestations Candice Silversides, Toronto, ON: Manifestations of the myocardium
<b>Representative, IACHD Nursing Network</b>	
Desiree Fleck, PhD, CRNP Children's Hospital of Philadelphia Philadelphia, USA	<b><u>Monday</u></b> Mon 11/14 7:30-8:45 AM Room W110a Ast the Experts <b>ATE.301.Heart Failure in the Adult with Congenital Heart Disease</b> Candice Silversides, Toronto, ON: Biomarkers and Medical Heart Failure Therapy in Adults with Congenital Heart Disease Edward Walsh, Boston, MA: Drugs and Pacemakers in Arrhythmia Management of Adults with Congenital Heart Disease and Heart Failure Jacqueline M Lamour, Bronx, NY: Challenges of Mechanical Assist and Heart Transplantation in Adults with Congenital Heart Disease
<b>Journal Watch Editor</b>	
Philip Moons, PhD, RN Catholic University Leuven Leuven, Belgium	Mon 11/14 Starting at 2:00PM <b>AOS.300.002YN.Alexander S. Nadas Lecture, Best of CVDY and the Latest from the Single Ventricle Reconstruction Trial</b> 3:45PM-4:00PM Room W110a 10098 Richard G Ohve, University of Michigan, Ann Arbor, MI: for the Pediatric Heart Network Investigators, Cause, Timing and Location of Death in Single Ventricle Reconstruction Trial
<b>Executive Director</b>	
Peter Kralka FPMR Raleigh, NC, USA	Mon 11/14 5:30-6:45PM, Room W 110a Cardiovascular Seminar <b>CVS.306.Cardiac Fibrosis in Pediatric Heart Disease: Finding It and Fixing It</b>
<b><u>Past Presidents</u></b>	
Gary Webb, MD 1994-1996	Tal Geva, Boston, MA: Imaging Fibrosis from Histology to Novel Imaging Techniques Seema Mital, Toronto, ON, Canada: Cardiac Fibrosis in Congenital Heart Disease
Carol Warnes, MD 1996-1998	Michael D Puchalski, Salt Lake City, UT: Significance of Fibrosis in Cardiomyopathies Steven D Colan, Boston, MA: Can We Prevent or Treat Cardiac Fibrosis?
Richard Liberthson, MD 1998-2000	Mon 11/14 5:30-6:45PM, Room W110b Cardiovascular Seminar <b>CVS.308.Frontiers in Congenital Transcatheter Cardiovascular Intervention</b>
Daniel Murphy, Jr., MD 2000-2002	Doff B McElhinney, Boston, MA Transcatheter Pulmonary Valve Implants: Patient Selection and Outcomes Andreas Eicken, Munich, Germany: The European Experience with Transcatheter Pulmonary Valve Implants John P Cheatham, Columbus, OH: Hybrid Procedures for Complex Congenital Heart Disease Pedro J Del Nido, Boston, MA: Surgical Strategies to Incorporate Future Transcatheter Revisions
Thomas P. Graham, Jr., MD 2002-2004	
Michael A. Gatzoulis, MD, PhD, FACC 2004-2006	
Jack M. Colman, MD 2006-2008	<b><u>Tuesday</u></b> Tues 5:30-6:45PM Room W110b Cardiovascular Seminar <b>CVS.310.Biomedical Engineering Insights into Congenital Heart Disease</b>
Michael J. Landzberg, MD 2008-2010	Ajit Yoganathan, Atlanta, GA: Applying Biomechanical Principles to Valve Design Kirk R. Kanter, Atlanta, GA: Can We Engineer a Better Cardiac Operation Using 3D Imaging Reconstruction Jeffrey A Feinstein, Palo Alto, CA: Fluid Dynamics and Engineering Insights for the Fontan Circulation John LaDisa, Milwaukee, WI: The Success of Aortic Arch Surgery: An Engineer's Perspective
	Tues 5:30-6:45PM, Room W104 Cardiovascular Seminar <b>Genetic Testing for Arrhythmia and Hypertrophic Cardiomyopathy: Current Role in Diagnosis, Treatment and Prognosis</b>

Silvia G Priori, Pavia, Italy: Other Channelopathies: Brugada, Catecholaminergic Polymorphic Ventricular Tachycardia, etc, and Arrhythmogenic Right Ventricle  
Hugh C Watkins, Oxford, United Kingdom: Hypertrophic Cardiomyopathy

### **Wednesday**

Wed 7:30-8:45 Room W110b How to Session

### **HTS.304.How to Manage Ventricular Tachycardia in Repaired Congenital Heart Disease**

Philip Saul, Charleston, SC: Endocardial and Epicardial Ablation  
Michael J Silka, Los Angeles, CA: Drugs and ICD  
Francis Fynn- Thompson, Boston, MA: Surgical Options

Barbara J.M. Mulder  
President

## **Regional News:**

### **News from Asia-Pacific**

#### ***By Koichiro Niwa***

1. Korea GUCH Satellite Symposium was held on September 3rd in Seoul. Drs. Carole A Warnes, Rick Nishimura, Mei Hwan Wu, and myself, were invited to give a presentation update on ACHD.

The Symposium celebrated the retirement of Heung Jae Lee MD, Samsung Med Center, the President of Korean GUCH Society. Dr Lee has been working for a long time in this field and had many great contributions on the development of the ACHD field in Korea, Asia and Internationally. He continues to stay in this field and still works with us.

2. The fourth Meeting of Adult CHD Seminar was held in Osaka, Japan, on October 22nd. The purpose of this seminar is primarily education. Wide range of basic and updated topics were discussed. About 200 attendants, including co-medicals, were gathered. This seminar will be held twice a year, first in Tokyo, and then in Osaka.

### **News from Latin America**

#### ***By Luis Alday***

The Joint ISACHD-SAC session during the Argentine Congress of Cardiology in Buenos Aires was a great success. Roberta Williams gave an excellent talk about drop out prevention of adolescents with CHD. The other speakers also delivered excellent talks. In the evening, everyone was taken to a tango show. Roberta impressed us all with her new ability - tango dancing. In the picture below you can see her dancing with a pro.



### **News from Canada**

#### ***By Erwin Oechslin***

#### **CACH Network Business Meeting in Vancouver**

CACH Network held its annual Business Meeting on October 23rd during the Canadian Cardiovascular Congress 2011 in Vancouver Canada. In addition to the usual business agenda, Erwin Oechslin reported on educational activities, national and international collaboration and relationships and the advantage of being an Affiliate of the Canadian Cardiovascular Society (CCS). The CACH Network continues to be a great success with more than 10,000 website visitors and more than 140,000 hits every month.

Dr. Marla Kiess delivered a very stimulating and inspiring talk entitled "How to set

up a combined ACHD/PHT clinic: the "Vancouver Experiences". She highlighted the benefits of a combined ACHD/PHT clinic which was established in Vancouver in 2008. This combined clinic has a major impact on patient care and has a great potential for education (for staff physician, fellows and residents). Toronto opened their combined ACHD/PHT clinic in 2011.

### **Patient Information on Adult Congenital Heart Disease and Pulmonary Arterial Hypertension**

This is a patient brochure with important and very helpful information for patients with pulmonary arterial hypertension associated with congenital heart disease. Doreen Fofonoff (lead; Vancouver), Yvonne Balon (Calgary) and Pamela Heggie (Edmonton) prepared the brochure content with excellent diagram illustrations. Actelion Pharmaceuticals Canada Inc., supported the project with an unrestricted grant. The brochure will be delivered to all CACH Network Centres and Pulmonary Hypertension Centres in Canada. An electronic version will be made available on the CACH Network website site (<http://cachnet.org>).

### **22st International ACHD Symposium in Toronto: May 30 - June 2, 2012**

After 2008, Toronto will become again the home city of this International Meeting, which was held at Skamania Lodge, Stevenson, WA, in 2010, and in Cincinnati in 2011. The meeting will be held at the Toronto Marriott Downtown hotel close to the famous Eaton Centre (please see the below flyer).

The following hot topics, amongst others, will be discussed at the 2012 meeting: the Fontan Disease - Diagnostic and Therapeutic Challenges of a Systemic Disease; End of Life and Supportive Care (Advanced Care) - an Ignored Challenge and Urgent Need for the Growing ACHD Population; Aortopathies, Arrhythmias, Heart Disease and Pregnancy. Breakout sessions with case presentations tailored to didactic presentations during plenary sessions (e.g. Fontan), will give the attendees the opportunity to discuss open questions with international experts in details. ACHD nurses will also play a major role at the Toronto meeting.

The necessary expertise and the multidisciplinary strategies required for caring for Congenital Heart Disease in the Adult will be stressed in a manner that allows practicing cardiologists, surgeons and nurses to appreciate and plan for managing their patients.

*Abstract Presentations:* There will be an opportunity for abstract submissions for ACHD providers from all backgrounds (physicians, nurses). The top three abstracts will be presented orally.

### **Heart Disease and Pregnancy Website**

This website was launched and is a great educational resource for physicians and patients!

The Heart Disease and Pregnancy website provides information for physicians and patients about the risks associated with pregnancy and strategies for management of pregnant women with heart disease. The website was developed by an international panel of physicians including internists, cardiologists, obstetricians as well as other medical specialists with expertise in pregnancy and heart disease. A drop-down menu for different acquired and congenital heart diseases makes for easy access to information about maternal and fetal complications, management strategies, references, and other subtopics for a specific disease. Key links to national and international societies, teaching tools are also listed and easily accessible.

I congratulate the contributors for their outstanding, globally available educational tool. Special thanks to the editors Dr. Candice Silversides, Dr. Jack Colman, Angela Kennie and web designer Ray Ganesh.

Visit the website at:

[www.heartdiseaseandpregnancy.com](http://www.heartdiseaseandpregnancy.com)

(there is also a link on the CACH Network Website [www.cachnet.org](http://www.cachnet.org))



## Journal Watch

Am J Cardiol 2011 Sept 21. [Epub ahead of print]

[Efficacy and Safety of Bosentan for Pulmonary Arterial Hypertension in Adults With Congenital Heart Disease.](#)

Monfredi O, Griffiths L, Clarke B, Mahadevan VS.

### Source

Cardiovascular Research Group, School of Biomedicine, University of Manchester, Manchester, United Kingdom.

### Abstract

The dual endothelin receptor antagonist, bosentan, has been shown to be well tolerated and effective in improving pulmonary arterial hypertension (PAH) symptoms in patients with Eisenmenger syndrome but data from longer-term studies are lacking. The aim of this study was to retrospectively analyze the long-term efficacy and safety of bosentan in adults with PAH secondary to congenital heart disease (PAH-CHD). Prospectively collected data from adult patients with PAH-CHD (with and without Down syndrome) initiated on bosentan from October 2007 through June 2010 were analyzed. Parameters measured before bosentan initiation (62.5 mg 2 times/day for 4 weeks titrated to 125 mg 2 times/day) and at each follow-up (1 month and 3, 6, 9, 12, 18, and 24 months) included exercise capacity (6-minute walk distance [6MWD]), pretest oxygen saturation, liver enzymes, and hemoglobin. Data were analyzed from 39 patients with PAH-CHD (10 with Down syndrome) who had received  $\geq 1$  dose of bosentan (mean duration of therapy  $2.1 \pm 1.5$  years). A significant ( $p < 0.0001$ ) average improvement in 6MWD of 54 m over a 2-year period in patients with PAH-CHD without Down syndrome was observed. Men patients had a 6MWD of 33 m greater than women ( $p < 0.01$ ). In all patients, oxygen saturation, liver enzymes, and hemoglobin levels remained stable. There were no discontinuations from bosentan owing to adverse events. In conclusion, patients with PAH-CHD without Down syndrome gain long-term symptomatic benefits in exercise capacity after bosentan treatment. Men seem to benefit more on bosentan treatment. Bosentan appears to be well tolerated in patients with PAH-CHD with or without Down syndrome.

## Article of the Month: September 2011

**Commentary on paper 'Efficacy and safety of Bosentan for Pulmonary Arterial Hypertension in Adults with Congenital Heart Disease.'**

**Monfredi O et al. Am J Cardiol 2011'**

**by Gabriele Egidy Assenza**

Pulmonary hypertension complicates natural history of congenital heart disease (CHD) in about 6% of adult congenital heart patients and is associated with increased mortality and morbidity (1). Of even greater

concern are adult patients with CHD and elevated pulmonary vascular resistance (pulmonary arterial hypertension or PAH). Sustaining greatest health and functional capacity in this population of patient is both clinically very relevant and challenging. Monfredi et al. show that the nonselective endothelin receptor antagonist Bosentan is associated with improved functional capacity in 39 patients with congenital heart disease and pulmonary arterial hypertension

(2). In this study the long term administration of Bosentan (to a target dose of 125 mg twice a day) improved 6 minute walking test distance by 54 meters from the baseline. Such improvement was sustained after two years from the beginning of the treatment and was not associated with either systemic desaturation or significant adverse effect. Bosentan was originally tested in the Bosentan Randomized Trial of Endothelin Antagonist Therapy-5, BREATHE-5 trial. In this randomized, placebo-controlled trial, focused on patients with Eisenmenger physiology and WHO functional class III, Bosentan administration was not associated with systemic desaturation and did induce a placebo-corrected improvement of 53 meters in 6 minute walking test performed 16 weeks after randomization (3). Moreover, in the treatment arm of the trial, pulmonary vascular resistance presented a placebo-corrected decrease of 427 dyne\*s\*cm-5 at the end of the follow-up. Such decrease challenged the classic notion which considered Eisenmenger physiology as characterized by a "fixed" pulmonary vascular bed with minimal or no possibility of dynamic modulation. After the results of this trial were made available, Bosentan rapidly became the first line treatment (though under utilized) for patients with Eisenmenger physiology and (although this was not tested in the trial) often for patients with PAH associated with CHD, even in the absence of Eisenmenger physiology. Concerns have been raised regarding the persistence of the drug benefit after the very short time of observation of the trial. In addition, Eisenmenger physiology represents a very specific sub-group of the PAH associated with CHD and no strong data had been provided suggesting any benefit of Bosentan in patients with CHD and PAH without Eisenmenger physiology. Diller et al. reported sustained improvement in functional capacity after a mean follow-up of 29 months in 14 patients with PAH associated with CHD, started on chronic Bosentan therapy (4). In addition, D'Alto and colleagues showed in a similar population of patients that the functional improvement was coupled with stable and favorable hemodynamic modulation after 12 months of therapy (5). However, the notion of stable and sustained benefit has been questioned by other groups (6-7), and in the setting of this conflict, the study of Monfredi et al. seems to suggest that such long term improvement could be a real clinical phenomenon and would imply to continue this therapy indefinitely, in the absence of significant adverse reactions. In line with these data, Gatzoulis et al. (as reported in the discussion section of the present study) reported improved survival in a retrospective cohort of patients with Eisenmenger physiology started on advanced therapy for pulmonary hypertension (73.5% of the patients in the treatment group were started on Bosentan) compared to a propensity-score matched cohort of similar patients who did not receive such treatments (8). Without doubt, our better understanding of the pathobiology of PAH associated with CHD, coupled with good clinical research, is improving our ability to care for this very challenging population of patients. Two national initiatives are underway in the United States, the first to assess adherence to guidelines in optimal care of adult patients with Eisenmenger physiology and the other to assess risk factors for development of PAH in adults after closure of systemic arterial to pulmonary arterial shunts.

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7. Apostolopoulou SC, Manginas A, Cokkinos DV, Rammos S. Long-term oral bosentan treatment in patients with pulmonary arterial hypertension related to congenital heart disease: a 2-year study. *Heart* 2007;93:350-4.
8. Dimopoulos K, Inuzuka R, Goletto S et al. Improved survival among patients with Eisenmenger syndrome receiving advanced therapy for pulmonary arterial hypertension. *Circulation* 2010;121:20-5.

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*Pacing Clin Electrophysiol*. 2011 Sep 28. doi: 10.1111/j.1540-8159.2011.03226.x. [Epub ahead of print]

[Pediatric and Adult Congenital Endocardial Lead Extraction or Abandonment Decision \(PACELEAD\) Survey of Lead Management.](#)

McCanta AC, Schaffer MS, Collins KK.

#### Source

Department of Pediatric Cardiology, The University of Colorado Denver School of Medicine, Children's Hospital Colorado, Aurora, Colorado  
Department of Pediatric Cardiology, Duke University Medical Center, Durham, North Carolina.

#### Abstract

Background: Nonfunctional, dysfunctional, recalled, or additional endocardial leads in pediatric and congenital heart disease patients pose significant challenges for management. There are no set standards for lead extraction in this patient population.

Methods: Physician members of the Pediatric and Adult Congenital Electrophysiology Society (PACES) were contacted via e-mail and invited to respond to a 33-question online Pediatric and Adult Congenital Endocardial Lead Extraction or Abandonment (PACELEAD) survey.

Results: Responses were received from 75 of 138 (54%) physician members of PACES. Institutional volumes of device placement (<25 devices/year for 51% of responders), patients with abandoned leads (<25 patients for 71%), and lead extractions (<10 extractions/year for 51% and no extractions for 29%) were low for the majority of responders.

Personal experience with lead extraction was also minimal with 49% not performing the procedure and 39% with less than 40 leads extracted as primary operator. Most responders (54, 72%) refer their lead extractions to another practitioner or facility with more experience. Responders were more likely to recommend lead extraction (>70%) for class IIa indications such as bacteremia, chronic pain that is not medically manageable, and functional leads with ipsilateral venous occlusion. Lead abandonment was favored (>70%) for one class IIb indication, a functional lead that is not currently being used.

Conclusions: Optimal lead management is challenging in pediatric and congenital heart disease patients, and considerable variability of practice is reported in their care. Low institutional and personal volumes may account for this variability. (PACE 2011; 1-7).

Int J Cardiol. 2011 Sep 17. [Epub ahead of print]

[Structure and function of the ascending aorta in palliated transposition of the great arteries.](#)

Ladouceur M, Kachenoura N, Lefort M, Redheuil A, Bonnet D, Celermajer DS, Iserin L, Mousseaux E.

**Source**

INSERM, UMRS 678, Université Pierre et Marie Curie, Paris, France; Université Paris Descartes, Centre de référence des Malformations Cardiaques Congénitales Complexes, M3C, APHP-Hôpital Necker, Paris, France; Adult congenital heart diseases Unit, Department of Cardiology, APHP-Hôpital Européen Georges Pompidou, Paris, France.

**Abstract**

**BACKGROUND:**

In transposition of the great arteries (TGA), the right ventricle (RV) is subaortic and abnormal aortic structure or function could adversely affect the capacity of the RV to supply the systemic circulation. Our aim was to assess aortic dimensions and distensibility and RV function in patients with palliated TGA using cardiovascular magnetic resonance imaging (CMR).

**METHODS:**

We studied 29 patients (22 males; age 29±4years) with simple TGA, who underwent an atrial switch procedure, and 29 age and sex matched controls. All subjects had cine and phase contrast CMR to evaluate aortic function and global RV function.

**RESULTS:**

TGA patients had significant dilatation of the aortic annulus (21.0±3.6mm vs. 17.6±4.1mm, p=0.002) and the sinus of Valsalva (30.0±4mm vs. 26.8±4.2mm, p=0.005,) compared to controls. These findings were associated with reduced distensibility of the ascending aorta in patients with TGA (3.5±1.6 vs. 5.3±2.4mmHg(-1).10(-3), p=0.0009). We could not show a significant correlation between aortic stiffness indices and RV size, function, mass or presence of fibrosis.

**CONCLUSION:**

The aortic root dilates and the ascending aorta stiffens in TGA, during young adult life. Although these proximal aortic changes did not show adverse effects on the RV in our young TGA sample, they might have important long-term physiopathological consequences in these patients.

Jpn J Radiol. 2011 Oct;29(8):576-82. Epub 2011 Sep 17.

[Interatrial shunt: diagnosis of patent foramen ovale and atrial septal defect with 64-row coronary computed tomography angiography.](#)

Kosehan D, Akin K, Koktener A, Cakir B, Aktas A, Teksam M.

**Source**

Department of Radiology, Fatih University School of Medicine, Alparslan Turkes Cad. No. 57, Emek, Ankara, 06510, Turkey, [drdilekoz@gmail.com](mailto:drdilekoz@gmail.com).

**Abstract**

**PURPOSE:**

The aim of this study was to investigate the frequency with which interatrial shunts are found during routine coronary computed tomography (CT) angiography and to describe imaging characterizations of patent foramen ovale (PFO), atrial septal defect (ASD), and atrial septal aneurysm (ASA).

**MATERIALS AND METHODS:**

A total of 1081 adult patients were evaluated retrospectively for interatrial shunting; 77 were excluded from the study. CT diagnosis of PFO was defined as (1) a channel-like appearance of the interatrial septum (IAS) and (2) a contrast agent jet flow from the left atrium (LA) to the right atrium (RA). ASD was defined as (1) the IAS resembling a membrane with a hole and (2) a contrast jet flow between the two atria. ASA was identified by detecting a minimum 10-mm protrusion of the LA beyond the IAS into the RA.

**RESULTS:**

Among 1004 patients, 86 patients (8.6%) were diagnosed to have PFO. Another 23 patients (2.3%) had a hole in the IAS and were diagnosed as having ASD: 21 with an ostium secundum-type ASD and 2 with the sinus venosus type. ASA accompanied ASD in three patients.

**CONCLUSION:**

Electrocardiography-gated CT using the saline-chaser contrast injection technique that is routinely used for coronary arterial imaging can be used to detect interatrial shunts. The technique can also serve as a method for differentiating PFO, ASD, and ASA.

JAMA. 2011 Sep 14;306(10):1104-12.

**[Incidence of aortic complications in patients with bicuspid aortic valves.](#)**

Michelena HI, Khanna AD, Mahoney D, Margaryan E, Topilsky Y, Suri RM, Eidem B, Edwards WD, Sundt TM 3rd, Enriquez-Sarano M.

**Source**

Division of Cardiovascular Disease, Mayo Clinic, 200 First St SW, Rochester, MN 55905, USA. [michelena.hector@mayo.edu](mailto:michelena.hector@mayo.edu)

**Abstract****CONTEXT:**

Bicuspid aortic valve (BAV), the most common congenital heart defect, has been thought to cause frequent and severe aortic complications; however, long-term, population-based data are lacking.

**OBJECTIVE:**

To determine the incidence of aortic complications in patients with BAV in a community cohort and in the general population.

**DESIGN, SETTING, AND PARTICIPANTS:**

In this retrospective cohort study, we conducted comprehensive assessment of aortic complications of patients with BAV living in a population-based setting in Olmsted County, Minnesota. We analyzed long-term follow-up of a cohort of all Olmsted County residents diagnosed with definite BAV by echocardiography from 1980 to 1999 and searched for aortic complications of patients whose bicuspid valves had gone undiagnosed. The last year of follow-up was 2008-2009.

**MAIN OUTCOME MEASURE:**

Thoracic aortic dissection, ascending aortic aneurysm, and aortic surgery.

**RESULTS:**

The cohort included 416 consecutive patients with definite BAV diagnosed by echocardiography, mean (SD) follow-up of 16 (7) years (6530 patient-years). Aortic dissection occurred in 2 of 416 patients; incidence of 3.1 (95% CI, 0.5-9.5) cases per 10,000 patient-years, age-adjusted relative-risk 8.4 (95% CI, 2.1-33.5; P = .003) compared with the county's general population. Aortic dissection incidences for patients 50 years or older at baseline and bearers of aortic aneurysms at baseline were 17.4 (95% CI, 2.9-53.6) and 44.9 (95% CI, 7.5-138.5) cases per 10,000 patient-years, respectively. Comprehensive search for aortic dissections in undiagnosed bicuspid valves revealed 2 additional patients, allowing estimation of aortic dissection incidence in bicuspid valve patients irrespective of diagnosis status (1.5; 95% CI, 0.4-3.8 cases per 10,000 patient-years), which was similar to the diagnosed cohort. Of 384 patients without baseline aneurysms, 49 developed aneurysms at follow-up, incidence of 84.9 (95% CI, 63.3-110.9) cases per 10,000 patient-years and an age-adjusted relative risk 86.2 (95% CI, 65.1-114; P < .001 compared with the general population). The 25-year rate of aortic surgery was 25% (95% CI, 17.2%-32.8%).

**CONCLUSIONS:**

In the population of patients with BAV, the incidence of aortic dissection over a mean of 16 years of follow-up was low but significantly higher than in the general population.

Neth Heart J. 2011 Sep 14. [Epub ahead of print]

**[Per-operative stent placement in the right pulmonary artery; a hybrid technique for the management of pulmonary artery branch stenosis at the time of pulmonary valve replacement in adult Fallot patients.](#)**

Windhausen F, Boekholdt SM, Bouma BJ, Groenink M, Backx AP, de Winter RJ, Mulder BJ, Hazekamp MG, Koolbergen DR.

**Source**

Department of Cardiology, Academic Medical Center, PO Box 22660, 1100 DD, Amsterdam, Netherlands.

**Abstract**

After having undergone surgical correction at an early age, many patients with tetralogy of Fallot develop long-term complications including progressive pulmonary regurgitation and peripheral pulmonary stenosis. A high percentage of these patients need to undergo a second operation in their adolescence or early adulthood. If simultaneous treatment of both pulmonary regurgitation and peripheral pulmonary stenosis is warranted, a complete surgical approach has several disadvantages. We describe four cases of Fallot patients with severe pulmonary regurgitation and peripheral pulmonary stenosis who were treated using a hybrid approach involving surgical implantation of a pulmonary homograft and peroperative stenting of the pulmonary artery.

J Cardiovasc Nurs. 2011 Sep 9. [Epub ahead of print]

[Quality of Life, Health Status, and Depression: Comparison Between Adolescents and Adults After the Fontan Procedure With Healthy Counterparts.](#)

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

**Source**

Nancy A. Pike, PhD, RN, CPNP-AC Assistant Professor, School of Nursing, University of California, Los Angeles. Lorraine S. Evangelista, PhD, RN Associate Professor, Program of Nursing Science, University of California, Irvine. Lynn V. Doering, PhD, RN Professor, School of Nursing, University of California, Los Angeles. Jo-Ann Eastwood, PhD, RN Assistant Professor, School of Nursing, University of California, Los Angeles. Alan B. Lewis, MD Pediatric Cardiologist, Division of Pediatric Cardiology, Children's Hospital Los Angeles. John S. Child, MD Cardiologist and Director, Adult Congenital Heart Disease Center, Ahmanson-University of California, Los Angeles.

**Abstract**

**BACKGROUND:**

Quality of life (QOL) in adolescents and adults who have undergone the Fontan procedure and are living with only 1 ventricle is presumed to be diminished.

**OBJECTIVES:**

This study aimed to compare QOL, health status, and prevalence of depression in adolescents/adults after the Fontan procedure with healthy counterparts and to identify predictors of QOL in the Fontan group.

**METHODS:**

Using a comparative, cross-sectional design, 54 adolescents and adults with single ventricle congenital heart disease who have undergone the Fontan procedure were compared with 66 age-matched healthy counterparts. Quality of life, health status, depression, and social support were measured using the Satisfaction With Life Scale, Short Form Survey Version 2, Patient Health Questionnaire Depression Module, and Multidimensional Scale of Perceived Social Support. Clinical variables were abstracted from medical records. Predictors of QOL were determined using multiple linear regression.

**RESULTS:**

Adolescents and adults in the Fontan group reported lower physical health status (mean [SD] = 46.5 [9.3] vs mean [SD] = 55.9 [5.1],  $P < .001$ ) and were more depressed (mean [SD] = 7.3 [5.9] vs mean [SD] = 4.5 [4.3],  $P < .004$ ) than their healthy counterparts. There were no differences in QOL, mental health status, or social support between the 2 groups. Functional status (New York Heart Association class), depression, and social support accounted for 55% of the variance in QOL in the Fontan group.

**CONCLUSIONS:**

Despite lower levels of physical health, the QOL of Fontan patients was comparable with that of their healthy counterparts; this finding contradicts previous proxy reports, self-reports, and assumptions that QOL is lower in patients with complex single ventricle congenital heart disease. However, because Fontan patients were more depressed than their healthy counterparts, the need for early screening and detection is

warranted.

J Thorac Cardiovasc Surg. 2011 Sep 10. [Epub ahead of print]  
[Outcomes in adult congenital heart surgery: Analysis of the Society of Thoracic Surgeons Database.](#)

Mascio CE, Pasquali SK, Jacobs JP, Jacobs ML, Austin EH 3rd.

**Source**

Division of Thoracic and Cardiovascular Surgery, Department of Surgery, University of Louisville, Louisville, Ky.

**Abstract**

**OBJECTIVE:**

Outcomes data for adults undergoing congenital heart surgery are limited. Previous analyses used administrative data or focused on single-center outcomes. We describe the most common operations, patient characteristics, and postoperative outcomes using a multicenter clinical database.

**METHODS:**

The study included adults (aged  $\geq 18$  years) listed in the Society of Thoracic Surgeons Congenital Heart Surgery Database (2000-2009). We describe patient characteristics and morbidity and mortality, and examine congenital procedures in the Society of Thoracic Surgeons Adult Cardiac Surgery Database to permit consideration of the primary dataset within a broader context.

**RESULTS:**

A total of 5265 patients (68 centers) from the Society of Thoracic Surgeons Congenital Heart Surgery Database were included. Patients' median age was 25 years (interquartile range, 20-35). Common preoperative risk factors included noncardiac abnormalities (17%) and arrhythmia (14%). Overall, in-hospital mortality was 2.1%, 27% had 1 or more complication, and median length of stay was 5 days. Common operations included right ventricular outflow tract procedures (21%) and pacemaker/arrhythmia procedures (20%). We further evaluated cardiopulmonary bypass procedures in more than 100 patients. Mortality ranged from 0% (atrial septal defect repair) to 11% (Fontan revision/conversion). Separate evaluation of the Society of Thoracic Surgeons Adult Cardiac Surgery Database revealed 39,872 adults undergoing congenital heart operations.

**CONCLUSIONS:**

Most adult congenital heart operations listed in the Society of Thoracic Surgeons Congenital Heart Surgery Database are performed in the third to fourth decades of life; approximately half are for right heart pathology or arrhythmia. Many patients have complications, but mortality is low with the exception of those undergoing Fontan revision/conversion. Many more adults undergoing congenital heart surgery are entered into the Society of Thoracic Surgeons Adult Cardiac Surgery Database.

Pediatr Cardiol. 2011 Sep 8. [Epub ahead of print]

[Independent Risk Factors for Cardiac Operations in Adults With Congenital Heart Disease: A Retrospective Study of 543 Operations for 500 Patients.](#)

Vogt MO, Hörer J, Grünewald S, Otto D, Kaemmerer H, Schreiber C, Hess J.

**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.

Eur Heart J. 2011 Sep 4. [Epub ahead of print]

[Acute pulmonary vasodilator response in paediatric and adult pulmonary arterial hypertension: occurrence and prognostic value when comparing three response criteria.](#)

Douwes JM, van Loon RL, Hoendermis ES, Vonk-Noordegraaf A, Roofthoof MT, Talsma MD, Hillege HL, Berger RM.

#### Source

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#### Abstract

**Aims**To assess the occurrence and prognostic value of acute vasodilator response (AVR) in paediatric vs. adult pulmonary arterial hypertension, and idiopathic/hereditary pulmonary arterial hypertension (iPAH/HPAH) vs. pulmonary arterial hypertension associated with congenital heart disease (PAH-CHD) using three different response criteria.  
**Methods and results**Ninety-nine PAH patients underwent AVR testing (37 children, 62 adults; 70 iPAH/HPAH, 29 PAH-CHD). Three response criteria from clinical practice were used to define AVR. The number of responders was evaluated separately in subgroups based on age, diagnosis, and presence of a non-restrictive post-tricuspid shunt. Numbers of responders varied importantly using the different criteria but were always higher in iPAH/HPAH, compared with PAH-CHD. The number of responders did not differ between paediatric and adult iPAH/HPAH. No responders were identified in patients with a post-tricuspid shunt. Acute vasodilator response was associated with improved survival using all three criteria. Low baseline mean right atrial pressure (mRAP) was associated with improved survival in adults (P < 0.001). High baseline mean pulmonary arterial pressure (mPAP)/mean systemic arterial pressure (mSAP) and pulmonary vascular resistance (PVR)/systemic vascular resistance (SVR) were associated with worse survival, statistically independent from age, diagnosis, and the presence of a post-tricuspid shunt.  
**Conclusion**The proportion of patients with AVR highly depends on the used criteria, but did not differ between paediatric and adult iPAH/HPAH. Current response criteria are not suitable for patients with a post-tricuspid shunt. In both children and adults without post-tricuspid shunts, AVR was associated with improved survival independent of the used criteria. Nevertheless, prognostic value in the individual patient was limited. Baseline mRAP showed a good correlation with survival for adult PAH patients, but not for children. High baseline mPAP/mSAP and PVR/SVR was associated with worse prognosis independent from age, diagnosis, or the presence of a post-tricuspid shunt.

Headache. 2011 Sep;51(8):1297-304. doi: 10.1111/j.1526-4610.2010.01824.x. Epub 2011 Jan 26.

[Headache in 25 consecutive patients with atrial septal defects before and after percutaneous closure-a prospective case series.](#)

Riederer F, Baumgartner H, Sándor PS, Wessely P, Wöber C.

#### Source

From the University Hospital Zurich, Department of Neurology, Zurich, Switzerland (F. Riederer and P.S. Sándor); University Hospital Munster, Adult Congenital and Valvular Heart Disease Center, Department of Cardiology and Angiology, Münster, Germany (H. Baumgartner); RehaClinic, Bad Zurzach, Switzerland (P.S. Sándor); Medical University Vienna, Department of Neurology, Waehringer Guertel, Vienna, Austria (P. Wessely and C. Wöber).

**Abstract**

(Headache 2011;51:1297-1304) In contrast to patent foramen ovale that is highly prevalent in the general population, atrial septal defect (ASD) is a rare congenital heart defect. The effect of ASD closure on headache and migraine remains a matter of controversy. The objectives of our study were (1) to determine headache prevalence in consecutive patients with ASD scheduled for percutaneous closure for cardiologic indications, using the International Classification of Headache Disorders and (2) to compare headache characteristics before and after closure of ASD. In this observational case series no a priori power analysis was performed. Twenty-five consecutive patients were prospectively included over 27 months. Median duration of follow-up was 12 months [interquartile range 0]. Prevalence of active headache seemed to be higher compared with the general population: any headaches 88% (95% confidence interval 70-96), migraine without aura 28% (14-48), migraine with aura 16% (6-35). After ASD closure, we observed a slightly lower headache frequency. In patients with ongoing headaches, a significant decrease in headache intensity was reported. Three patients reporting migraine with aura before the intervention noted no migraine with aura attacks at follow-up, 2 of them reported ongoing tension-type headache, 1 migraine without aura. In summary, this prospective observational study confirms the high prevalence of headache, particularly migraine, in ASD patients and suggests a possible small beneficial effect of ASD closure.

Circ J. 2011 Sep 22;75(10):2287-95. Epub 2011 Sep 1.

[Recent advances in adult congenital heart disease.](#)

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

**Source**

Adult Congenital Heart Center, Montreal Heart Institute, Université de Montréal.

**Abstract**

As a result of major achievements in pediatric cardiac care, a growing number of patients with congenital heart disease (CHD) are flourishing well into adulthood. This heterogeneous and aging population of patients, many of whom represent the first generation of middle-age survivors, faces unique issues and challenges. As a field, adult CHD has evolved markedly during the past decade on several fronts, including imaging, arrhythmia management, percutaneous interventions, surgical techniques, research, and multidisciplinary care that extends beyond the cardiac realm. This review highlights recent advances across the wide spectrum of key issues encountered by adults with CHD. (Circ J 2011; 75: 2287-2295).

Eur J Pediatr. 2011 Sep;170(9):1111-27. Epub 2011 Jul 20.

[Infective endocarditis in congenital heart disease.](#)

Knirsch W, Nadal D.

**Source**

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**Abstract**

Congenital heart disease (CHD) has become the leading risk factor for pediatric infective endocarditis (IE) in developed countries after the decline of rheumatic heart disease. Advances in catheter- and surgery-based cardiac interventions have rendered almost all types of CHD amenable to complete correction or at least palliation. Patient survival has increased, and a new patient population, referred to as adult CHD (ACHD) patients, has emerged. Implanted prosthetic material paves the way for cardiovascular device-related infections, but studies on the management of CHD-associated IE in the era of cardiovascular devices

are scarce. The types of heart malformation (unrepaired, repaired, palliated) substantially differ in their lifetime risks for IE. Streptococci and staphylococci are the predominant pathogens. Right-sided IE is more frequently seen in patients with CHD. Relevant comorbidity caused by cardiac and extracardiac episode-related complications is high. Transesophageal echocardiography is recommended for more precise visualization of vegetations, especially in complex type of CHD in ACHD patients. Antimicrobial therapy and surgical management of IE remain challenging, but outcome of CHD-associated IE from the neonate to the adult is better than in other forms of IE. Conclusion: Primary prevention of IE is vital and includes good dental health and skin hygiene; antibiotic prophylaxis is indicated only in high-risk patients undergoing oral mucosal procedures.

J Clin Endocrinol Metab. 2011 Sep;96(9):E1517-26. Epub 2011 Jul 13. [Standardized multidisciplinary evaluation yields significant previously undiagnosed morbidity in adult women with turner syndrome.](#)

Freriks K, Timmermans J, Beerendonk CC, Verhaak CM, Netea-Maier RT, Otten BJ, Braat DD, Smeets DF, Kunst DH, Hermus AR, Timmers HJ.

**Source**

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**Abstract**

Context: Besides short stature and gonadal dysgenesis, Turner syndrome (TS) is associated with various abnormalities. Adults with TS have a reduced life expectancy, mainly related to structural abnormalities of the heart and aorta, and an increased risk of atherosclerosis. Objective: Our objective was to investigate the yield of an initial standardized multidisciplinary screening in adult TS patients. Design and Setting: This was an observational study at a multidisciplinary care unit for adult women with TS. Participants: Participants were adult women with TS (n=150). Mean age was 31.0±10.4 yr, with 47% karyotype 45,X. Interventions: All women were consulted by an endocrinologist, a gynecologist, a cardiologist, an otorhinolaryngologist, and when indicated, a psychologist. The screening included magnetic resonance imaging of the heart and aorta, echocardiography, electrocardiogram, dual-energy x-ray absorptiometry, renal ultrasound, audiogram, and laboratory investigations according to international expert recommendations. Main outcome measures: New diagnoses and prevalence of TS-associated morbidity were evaluated. Results: Thirty percent of patients currently lacked medical follow-up, and 15% lacked estrogen replacement therapy in the recent last years. The following disorders were newly diagnosed: bicuspid aortic valve (n=13), coarctation of the aorta (n=9), elongation of the transverse aortic arch (n=27), dilation of the aorta (n=34), osteoporosis (n=8), osteopenia (n=56), renal abnormalities (n=7), subclinical hypothyroidism (n=33), celiac disease (n=3), glucose intolerance (n=12), dyslipidemia (n=52), hypertension (n=39), and hearing loss warranting a hearing aid (n=8). Psychological consultation was needed in 23 cases. Conclusions: Standardized multidisciplinary evaluation of adult women with TS as advocated by expert opinion is effective and identifies significant morbidity. Girls with TS benefit from a careful transition to ongoing adult medical care.

Curr Opin Support Palliat Care. 2011 Sep;5(3):291-6.

[Supportive and palliative care for adults dying from congenital heart defect.](#)

Tobler D, de Stoutz N, Greutmann M.

**Source**

Department of Cardiology, University Hospital Basel, Switzerland.

**Abstract**

PURPOSE OF REVIEW:

Adult survivors with congenital heart disease (CHD) are a novel group of adults with chronic disease. Due to the nature of their surgical repair, the majority is not cured. This review focuses on changing demographics and

mortality of adults with CHD, available knowledge and data on end-of-life (EOL) and palliative care in this patient group and outlines needs for future clinical practice and research.

**RECENT FINDINGS:**

Recent findings underscore that survival to adulthood has become the rule for the majority of patients born with CHD. Many remain at increased risk of premature death from their disease. Chronic circulatory failure has been identified as one of the leading causes of death and, thus, patients may benefit from incorporation of palliative care principles in their management. Recent studies show that this practice has not yet been widely adopted. Nonetheless a survey of adult outpatients has shown that many patients prefer to be involved in EOL decisions early in the course of their disease.

**SUMMARY:**

More research is needed to learn how we can best implement principles of palliative care in the growing population of adult survivors with CHD. This includes improved communication about EOL issues and a more comprehensive approach for symptom control.

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