

December, 2011

**Do you need
to update
your Member
Information?**

[Click here](#)



**International Society for
Adult Congenital Heart Disease**

ISACHD Newsletter

In This Issue

[Regional News](#)

[22nd
International
Symposium on
ACHD](#)

[Journal Watch](#)

President

Barbara Mulder,
M.D.
Academic Medical
Center
Amsterdam, The
Netherlands

President-Elect

Dr. Curt J Daniels
Columbus
Children's Hospital
Columbus, OH,
USA

Secretary

Philip Moons, PhD,
RN
Catholic University
Leuven
Leuven, Belgium

Treasurer

Koichiro Niwa,
M.D.
Chiba
Cardiovascular
Center
Chiba, Japan

Past President

Michael J.
Landzberg, M.D.

President's Message

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



During the AHA congress in Orlando, the ISACHD meeting was held on Sunday evening, November 13th. With 82 attendees it was well visited and there were quite some new faces. The ISACHD meeting was preceded by an ACHA reception in honor of Gary Webb, leaving the ACHA Board. Mike Landzberg and Amy Verstappen held an emotional speech, and Gary was well prepared with a humorous response.

The official ISACHD meeting began with an optimistic view on numbers of members and financial status. With a positive net income, ISACHD is not threatened by a bankruptcy, and even seems to flourish with the three new working groups, led by Curt Daniels, Erwin Oechslin, and Koichiro Niwa. But there is a large amount of "free-riders" who forget to pay their dues. In the next coming months we will make some more efforts to remind these members to fulfill their duty.

The main issue we discussed was the progress of the newly formed working groups on Health care (Curt Daniels), Education (Erwin Oechslin) and Research (Koichiro Niwa). After a short overview of the goals and of the first steps that have been made, the attendees were split to form three break-out sessions. It was discussed how to proceed with the next steps in the coming two years.

Curt and his group thought it would be wise to look for other international organizations, already working in the field of health care in under-served areas. Project Hope seems an attractive organization, already involved in many areas around the world. Curt has recently met the president of Project Hope to discuss possibilities for collaboration. Also the new ISACHD membership of the World Heart Federation will create opportunities to find partners, allowing realistic and feasible plans for health care projects.

Erwin was happy to have Gary on board with his large experience on education and web based material from the Cincinnati meeting in June.

Children's Hospital
Boston, MA, USA

**Representative,
USA**

William Davidson,
M.D.
The Pennsylvania
State University
Hershey, PA, USA

**Representative,
South America**

Luis Alday, M.D.
Hospital de Niños
Córdoba,
Argentina

**Representative,
Europe**

Helmut
Baumgartner, M.D.
Adult Congenital
And Valvular Heart
Center
Universitätsklinikum
Muenster
Germany

**Representative,
Asia-Pacific**

Koichiro Niwa,
M.D.
Chiba
Cardiovascular
Center
Chiba, Japan

**Representative,
Canada**

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

**Representative,
IACHD Nursing
Network**

Desiree Fleck,
PhD, CRNP
Children's Hospital

Els Pieper from Groningen, the Netherlands, did an excellent job by developing a draft for a basic teaching course on the internet. The easiest and cheapest way to proceed would probably be to record the presentations at home by the various speakers. The teaching course needs fine-tuning, but this goal could be for a large part be achieved within half a year. The second plan of this working group is to collect links of high quality meetings and education slides of existing resources. All members are asked to provide as much information as possible. The collected education material will be organized and placed on the ISACHD website. Finally, endorsement of ACHD related meetings by ISACHD has already started with two courses. It will increase visibility (flyers, ISACHD logo) and endorsement will include a reduced registration fee for ISACHD members.

Koichiro and his group started discussing topics which could be suitable for global research. Quality of life, patients lost to follow-up, sports and exercise, and research on health care provision in various regions were suggested as good starters. They would make an attractive "product" when looking for financial support.

Regional news was reported by the regional representatives. At the CACH website (www.cachnet.ca) information can be found on pulmonary hypertension (patient information) and pregnancy (risk stratification and management) in ACHD patients. Koichiro mentioned the tremendous progress made in the Asian-Pacific area. Nursing news came from Desiree Fleck.

At 9 pm we had to close a very busy and active meeting of ISACHD. Many promises were made for the next ISACHD meeting in April 25th 2012 in Chicago, where hopefully again many members will be welcomed.

Please, let us know when you want to join one of the three working groups. All support is welcome and needed to achieve the desired goals.

Barbara J.M. Mulder
President

Regional and Working Groups Update:

• **Research Working Group**

By Koichiro Niwa

Members of the Research Working Group at this point are as follow: Clare O'Dornell MD (NZ), Vaikom Mahadevan MD (UK), Mary Heitschmidt (US), Barbara Mulder (NL), Philip Moons (Belgium), Desiree Fleck (US), Dimitra Kalimanovska-Ostric (Serbia), Ann Gianola (US), Nancy McCabe (US), Ann Marie Valente (US).

The studies we are going to have are divided into two groups: Disease Specific and General (non-disease specific). This time we selected four general topics.

of Philadelphia
Philadelphia, USA

**Journal Watch
Editor**

Philip Moons, PhD,
RN
Catholic University
Leuven
Leuven, Belgium

Executive Director

Peter Kralka
FPMR
Raleigh, NC, USA

Past Presidents

Gary Webb, MD
1994-1996

Carol Warnes, MD
1996-1998

Richard Liberthson,
MD
1998-2000

Daniel Murphy, Jr.,
MD
2000-2002

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

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

Jack M. Colman,
MD
2006-2008

Michael J.
Landzberg, MD
2008-2010

Those are:

- 1) International ACHD care facility survey: establishment and education
- 2) Survey: Lost Follow-up
- 3) EX and Exercise training in ACHD
- 4) QOL measures, Psychological issues international difference

We will select the leader and fix the protocol of each study shortly.
And regarding the leadership of Phillip - it is excellent! Nancy McCabe (nancym.mccabe@gmail.com) comes from Emory University; she is very much interested in the QOL research.

- **WG on Education - News from Orlando**

Erwin Oechslin welcomed 10 participants (eight of them were new members) in the breakout session of the WG on education at the IACHD meeting in Orlando. Els Pieper, UMC Groningen (NL) put together an excellent preliminary program for a Web-based 2-day teaching course for trainees.

The group had some discussion about the content of the course and course module: should there be a module for ACHD trainees with a pediatric background and one for trainees with an adult cardiology background? There was the agreement that this teaching course should focus on trainees with an adult cardiology background.

Members of the group provided important input on educational tools about (adult) congenital heart disease, which are already available (e.g. www.cardiacmorphology.com). Erwin Oechslin is collecting information of such tools so that the core group under the lead of Els Pieper can evaluate them and avoid duplications.

The group set the following timelines:

- December 31, 2011 Program finalized
- ACC 2012 Speakers identified
- AHA 2012 Talks recorded

Erwin Oechslin, Toronto
Chair, WG on Education

- **News from Canada**

By Erwin Oechslin

Special Rate for the Toronto Meeting 2012:

Paying ISACHD members and CACH Network members will be offered a 10% reduction on the registration fee of the 22nd International Symposium on Adult Congenital Heart Disease in Toronto which will be held from May 30 to June 2, 2012.

Save the Date

**22nd International Symposium on
Adult Congenital Heart Disease**
"BEYOND SAVING LIVES"

Toronto, May 30 to June 2, 2012

University of Toronto Congenital Cardiac Centre for Adults Peter Munk Cardiac Centre University Health Network Scl Kids The Labatt Family Heart Centre

Journal Watch

Postgrad Med. 2011 Nov;123(6):32-45.

[Adult congenital heart disease and pulmonary arterial hypertension: the Texas Adult Congenital Heart Program experience.](#)

Franklin WJ, Parekh DR, Safdar Z.

Texas Children's Hospital, Baylor College of Medicine, Department of Medicine, Cardiology Section and Department of Pediatrics, Cardiology Section, Houston, TX. wjf@bcm.edu.

Congenital heart disease (CHD) is a common structural defect of the heart or major blood vessels. Patients with adult congenital heart disease (ACHD) have medical needs that are distinct from those of pediatric patients with CHD, and the transition into adult health care is important for management of the patient with ACHD. A large proportion of patients with CHD develop diseases and complications associated with the long-term stress of intracardiac shunts. Pulmonary arterial hypertension (PAH) is a significant complication of some CHD lesions. The treatment of these patients remains challenging due to their combined heart and lung disease, and multidisciplinary care is often necessitated for a variety of secondary conditions. A number of treatment options are available for the management of PAH associated with CHD, including prostanoids, phosphodiesterase type-5 inhibitors, and endothelin receptor antagonists. This article discusses the diagnosis and management of such ACHD patients with PAH.

Ann Thorac Surg. 2011 Nov 16. [Epub ahead of print]

[Coronary Artery Disease in Adult Congenital Heart Disease: Outcome After Coronary Artery Bypass Grafting.](#)

Stulak JM, Dearani JA, Burkhart HM, Ammash NM, Phillips SD, Schaff HV.

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

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.

Cardiology. 2011 Nov 17;120(1):36-42. [Epub ahead of print]

[Patient-Reported Outcomes in Adult Survivors with Single-Ventricle Physiology.](#)

Overgaard D, Schrader AM, Lisby KH, King C, Christensen RF, Jensen HF, Idorn L, Søndergaard L, Moons P.

The Heart Centre, Copenhagen University Hospital, Copenhagen, Denmark.

Objectives: Data on patient-reported outcomes (PROs) in patients with single-ventricle physiology (SVP) are scarce. We sought (1) to describe

the perceived health status, quality of life, symptoms of anxiety and depression, and sense of coherence in adult survivors with SVP, (2) to compare PROs across functional classes, and (3) to compare PROs between patients and controls. Methods: A case-control study in two adult congenital heart programmes with 62 adult survivors with SVP were matched to 172 healthy controls. A wide range of PROs were measured using validated questionnaires. The treating physician classified patients according to the Ability Index. Results: Patients with SVP have a good functional status. Patients in Ability Index class I consistently reported the best scores, similar to those of healthy controls. Negative associations were found between functional class and outcomes of perceived health and quality of life. For patients in Ability Index class II and III, PROs were poorer. Conclusions: PROs in patients with SVP are generally good.

Heart. 2011 Nov 10. [Epub ahead of print]

[Effects of atorvastatin on endothelial function and the expression of proinflammatory cytokines and adhesion molecules in young subjects with successfully repaired coarctation of aorta.](#)

Brili S, Tousoulis D, Antonopoulos AS, Antoniadis C, Hatzis G, Bakogiannis C, Papageorgiou N, Stefanadis C.

Hippokraton Hospital, Athens University Medical School, Athens, Greece.

Objective: To investigate the effects of atorvastatin on endothelial function and low-grade systemic inflammation in subjects with successful surgery for aortic coarctation repair (SCR). Design Open-label study. Setting Outpatients visiting the adult congenital heart disease department of our hospital. Patients 34 young people with SCR. Interventions Patients with SCR received atorvastatin 10 mg/day (n=17) or no treatment (n=17) for 4 weeks. At baseline and at 4 weeks, endothelial function was assessed by flow-mediated dilatation (FMD) of the right brachial artery, and blood samples were obtained. Serum levels of interleukin (IL) 1b, IL-6 and soluble vascular cell adhesion molecule-1 (sVCAM-1) were determined by ELISA. Main outcome measures Effects of treatment on FMD and serum levels of IL-1b, IL-6 and sVCAM-1. Results FMD in the atorvastatin group was significantly improved after 4 weeks (from 6.46±0.95% to 11.24±1.38%, p<0.01), while remaining unchanged in the control group (from 6.74±0.58% to 6.95±0.53%, p=NS). Even though atorvastatin had no effect on serum IL-6 levels (0.62 (0.37-0.88) pg/ml to 0.53 (0.28-0.73) pg/ml, p=NS), it significantly reduced circulating levels of IL-1b (from 1.17 (0.92-1.77) pg/ml to 1.02 (0.75-1.55) pg/ml, p<0.05) and sVCAM-1 (from 883.4 (660.3-1093.1) ng/ml to 801.4 (566.7-1030.2) ng/ml, p<0.05). No changes were seen in serum levels of IL-6, IL-1b and sVCAM-1 in the control group after 4 weeks compared with baseline (p=NS for all). Conclusions Atorvastatin treatment for 4 weeks in subjects with SCR significantly improved endothelial function and suppressed systemic inflammatory status by decreasing circulating

levels of IL-1b and sVCAM-1.

Cardiol Young. 2011 Nov 9:1-9. [Epub ahead of print]

[Total isovolumic time relates to exercise capacity in patients with transposition of the great arteries late after atrial switch procedures.](#)

Tay EL, Gibson D, Inuzuka R, Josen M, Alonso-Gonzalez R, Giannakoulas G, Li W, Dimopoulos K, Gatzoulis MA.

1Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital, London, United Kingdom.

BACKGROUND:

Systemic right ventricular systolic dysfunction is common late after atrial switch surgery for transposition of the great arteries. Total isovolumic time is the time that the ventricle is neither ejecting nor filling and is calculated without relying on geometric assumptions. We assessed resting total isovolumic time in this population and its relationship to exercise capacity.

METHODS:

A total of 40 adult patients with transposition of the great arteries after atrial switch - and 10 healthy controls - underwent transthoracic echocardiography and cardiopulmonary exercise testing from January, 2006 to January, 2009. Resting total isovolumic time was measured in seconds per minute: 60 minus total ejection time plus total filling time.

RESULTS:

The mean age was 31.6 plus or minus 7.6 years, and 38.0% were men. There were 16 patients (40%) who had more than or equal to moderate systolic dysfunction of the right ventricle. Intra- and inter-observer agreement was good for total isovolumic time, which was significantly prolonged in patients compared with controls (12.0 plus or minus 3.9 seconds per minute versus 6.0 plus or minus 1.8 seconds per minute, p-value less than 0.001) and correlated significantly with peak oxygen consumption (r equals minus 0.63, p-value less than 0.001). The correlation strengthened (r equals minus 0.73, p-value less than 0.001) after excluding seven patients with exercise-induced cyanosis. No relationship was found between exercise capacity and right ventricular ejection fraction or long-axis amplitude. **Conclusion** Resting isovolumic time is prolonged after atrial switch for patients with transposition of the great arteries. It is highly reproducible and relates well to exercise capacity.

Int J Cardiol. 2011 Nov 1. [Epub ahead of print]

[Self-reported health status \(EQ-5D\) in adults with congenital heart disease.](#)

Berghammer M, Karlsson J, Ekman I, Eriksson P, Dellborg M.

The Sahlgrenska Academy at University of Gothenburg, Institute of Health and Care Sciences, Box 457, SE-40530 Gothenburg, Sweden.

PURPOSE:

Today, more patients with congenital heart disease (CHD) reach

adulthood. There are conflicting findings concerning the relationship between quality of life (QoL) or health state for adults with CHD and the complexity of their CHD. The aim of the study was, firstly, to compare the reported health status and health perception of adult patients with CHD and, secondly, to investigate what variables influenced the patients' health status and health perception.

METHODS:

Data from 1435 patients completing the EQ-5D questionnaire, which includes reported health status and health perception, were analyzed.

RESULTS:

Valid EQ-5D data were reported by 1274 patients, showing overall results indicating a good health status. Problems were most frequently reported in the dimension "pain/discomfort" (31.9%) and "anxiety/depression" (29.8%). Higher occurrence of problems were reported by patients with complex disease i.e. single ventricle ($p < 0.001$) and by female patients ($p < 0.0001$). Symptomatic patients reported a lower health status ($p < 0.0001$) and a lower perceived health on EQ-VAS ($p < 0.0001$). Of the asymptomatic patients, 20.5% nevertheless reported problems in "pain/discomfort" and 22.2% in the "anxiety/depression" dimension.

CONCLUSION:

The health status of adults with CHD is influenced by symptoms, NYHA-classification, age and gender. Adults with CHD report a lower occurrence of problems in comparison to previously published results from a general population, but the importance of actively asking about the patient's experience is demonstrated by the high degree of asymptomatic patients reporting problems on EQ-5D.

J Card Fail. 2011 Nov;17(11):957-63

[Left ventricular function in adult patients with atrial septal defect: implication for development of heart failure after transcatheter closure.](#)

Masutani S, Senzaki H.

Department of Pediatric Cardiology, International Medical Center, Saitama Medical University, Saitama, Japan.

Despite advances in device closure for atrial septal defect (ASD), post-closure heart failure observed in adult patients remains a clinical problem. Although right heart volume overload is the fundamental pathophysiology in ASD, the post-closure heart failure characterized by acute pulmonary congestion is likely because of age-related left ventricular diastolic dysfunction, which is manifested by acute volume loading with ASD closure. Aging also appears to play important roles in the pathophysiology of heart failure through several mechanisms other than diastolic dysfunction, including ventricular systolic and vascular stiffening and increased incidence of comorbidities that significantly affect cardiovascular function. Recent studies suggested that accurate assessment of preclosure diastolic function, such as test ASD occlusion, may help identify high-risk patients for post-closure heart failure. Anti-heart failure

therapy before device closure or the use of fenestrated device appears to be effective in preventing post-closure heart failure in the high-risk patients. However, the long-term outcome of such patients remains to be elucidated. Future studies are warranted to construct an algorithm to identify and treat patients at high risk for heart failure after device closure of ASD.

Eur Heart J. 2011 Oct 27. [Epub ahead of print]

[Exercise training improves exercise capacity in adult patients with a systemic right ventricle: a randomized clinical trial.](#)

Winter MM, van der Bom T, de Vries LC, Balducci A, Bouma BJ, Pieper PG, van Dijk AP, van der Plas MN, Picchio FM, Mulder BJ.

Department of Cardiology, Academic Medical Center, Meibergdreef 9, 1105 AZ Amsterdam, The Netherlands.

Objective: To assess whether exercise training in adult patients with a systemic right ventricle (RV) improves exercise capacity and quality of life and lowers serum N-terminal prohormone brain natriuretic peptide (NT-proBNP) levels. **Design** Multi-centre parallel randomized controlled trial. **Participants** Patients with a systemic RV due to congenitally or surgically corrected transposition of the great arteries. **Methods** Fifty-four adult patients with a systemic RV, were randomized using unmarked opaque envelopes to an intervention group (n = 28) with three training sessions per week for 10 consecutive weeks, and a control group (n = 26). Randomization was stratified by participating centre. At baseline, and follow-up, we determined maximal exercise capacity ($\dot{V}O_{2peak}$), serum NT-proBNP levels, and quality of life by means of the SF-36, and the TAAQOL Congenital Heart Disease questionnaires. The final analysis was performed by linear regression, taking into account the stratified randomization. **Results** Forty-six patients were analysed (male 50%, age 32 ± 11 years, intervention group n = 24, control group n = 22). Analysis at 10 weeks showed a significant difference in $\dot{V}O_{2peak}$ (3.4 mL/kg/min, 95% CI: 0.2 to 6.7; P = 0.04) and resting systolic blood pressure (-7.6 mmHg, 95% CI: -14.0 to -1.3; P = 0.03) in favour of the exercise group. No significant changes were found in serum NT-proBNP levels or quality of life in the intervention group or in the control group nor between groups. None of the patients in the intervention group had to discontinue the training programme due to adverse events. **Conclusion** In adult patients with a systemic RV exercise training improve exercise capacity. We recommend to revise restrictive guidelines, and to encourage patients to become physically active. (Trial registration: The study was registered at <http://trialregister.nl>. Identifier: NTR1909.).

J Interv Card Electrophysiol. 2011 Oct 21. [Epub ahead of print]

[Systemic venous anatomy in congenital heart disease: implications for electrophysiologic testing and catheter ablation.](#)

Cordina RL, Celermajer DS, McGuire MA.

Royal Prince Alfred Hospital, Sydney, Australia.

INTRODUCTION:

Cardiac arrhythmias are a significant problem in patients with congenital heart disease. Many patients with congenital heart disease have abnormal systemic venous anatomy which can complicate electrophysiologic testing, catheter ablation and pacemaker and defibrillator implantation. We reviewed the systemic venous anatomy in a cohort of patients undergoing electrophysiologic testing and catheter ablation.

METHODS AND RESULTS:

We reviewed all electrophysiologic studies performed in patients with adult congenital heart disease (n=80) at our institution between January 1998 and October 2009. Ten patients (13%) had a congenital systemic venous anomaly. Of these, seven (9%) had a left superior vena cava and four (5%) had infrahepatic interruption of the inferior vena cava (two had both anomalies). One patient's inferior vena cava was connected to a left-sided atrium; she had right atrial isomerism. In four patients (40%), systemic venous abnormalities were discovered at the time of electrophysiologic testing.

CONCLUSIONS:

Systemic venous anomalies occur frequently in the congenital heart disease population and may complicate electrophysiologic testing and catheter ablation. Pre-procedural imaging may assist in facilitating a successful procedure.

Cardiol Young. 2011 Oct 21:1-9. [Epub ahead of print]

[Risk factors for loss to follow-up among children and young adults with congenital heart disease.](#)

Mackie AS, Rempel GR, Rankin KN, Nicholas D, Magill-Evans J.
1Division of Cardiology, Department of Pediatrics, University of Alberta, Edmonton, Alberta, Canada.

OBJECTIVE:

To identify risk factors for loss to cardiology follow-up among children and young adults with congenital heart disease.

METHODS:

We used a matched case-control design. Cases were born before January, 2001 with moderate or complex congenital heart disease and were previously followed up in the paediatric or adult cardiology clinic, but not seen for 3 years or longer. Controls had been seen within 3 years. Controls were matched 3:1 to cases by year of birth and congenital heart disease lesion. Medical records were reviewed for potential risk factors for loss to follow-up. A subset of cases and controls participated in recorded telephone interviews.

RESULTS:

A total of 74 cases (66% male) were compared with 222 controls (61% male). A history of missed cardiology appointments was predictive of loss to follow-up for 3 years or longer (odds ratio 13.0, 95% confidence interval 3.3-51.7). Variables protective from loss to follow-up were higher

family income (odds ratio 0.87 per \$10,000 increase, 0.77-0.98), cardiac catheterisation within 5 years (odds ratio 0.2, 95% confidence interval 0.1-0.6), and chart documentation of the need for cardiology follow-up (odds ratio 0.4, 95% confidence interval 0.2-0.8). Cases lacked awareness of the importance of follow-up and identified primary care physicians as their primary source of information about the heart, rather than cardiologists. Unlike cases, controls had methods to remember appointments.

CONCLUSIONS:

A history of one or more missed cardiology appointments predicted loss to follow-up for 3 or more years, as did lack of awareness of the need for follow-up. Higher family income, recent catheterisations, and medical record documentation of the need for follow-up were protective.

Congenit Heart Dis. 2011 Oct 20. doi: 10.1111/j.1747-0803.2011.00567.x. [Epub ahead of print]

[Five-year Follow-up of Intracardiac Echocardiography-assisted Transcatheter Closure of Complex Ostium Secundum Atrial Septal Defect.](#)

Rigatelli G, Dell' Avvocata F, Cardaioli P, Giordan M, Vassiliev D, Nghia NT, Chen JP.

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

Objective: We sought to prospectively evaluate long-term follow-up results of intracardiac echocardiography-aided transcatheter closure of complex atrial septal defects (ASD) in the adults.

Design and Settings. Prospective multicenter registry in tertiary care hospitals. **Patients and Interventions.** Over a 5-year period, we prospectively enrolled 56 patients (mean age 49±16.7 years, 24 females) who have been referred to our center for catheter-based closure of complex secundum ASD (>25mm diameter, deficiency of ≥1 rim, multiple secundum ASD, multiperforated ASD, associated incomplete floor of the fossa ovalis with or without aneurysm, embryonic remnants of incomplete atrial septation). All patients were screened by means of transesophageal echocardiography before the operation. Eligible patients underwent intracardiac echocardiography study and closure attempt. **Results.** Forty patients underwent a transcatheter closure attempt: transesophageal echocardiography-planned device type and size were modified in 32 patients (64%). Rates of procedural success, predischage occlusion, and major complications rate were 100%, 90%, and 2%, respectively. On mean follow-up of 5.4±1.8 years, the follow-up occlusion rate was 98%. During follow-up, only one case of permanent atrial fibrillation was observed. There were no cases of aortic/atrial erosion, device thrombosis, or new

atrioventricular valve dysfunction.

Conclusions. Intracardiac echocardiography-guided complex secundum ASD transcatheter closure is safe and effective and appears to have excellent long-term results, thus minimizing potential complications resulting from the complex anatomy.

Circ Cardiovasc Qual Outcomes. 2011 Nov 1;4(6):634-9. Epub 2011 Oct 18.

[Resource use among adult congenital heart surgery admissions in pediatric hospitals: risk factors for high resource utilization and association with inpatient death.](#)

Kim YY, Gauvreau K, Bacha EA, Landzberg MJ, Benavidez OJ.

Divisions of Cardiology, Hospital of the University of Pennsylvania and Children's Hospital of Philadelphia, University of Pennsylvania School of Medicine, Philadelphia, PA.

Background- Pediatric hospitals frequently perform congenital heart surgery in adults with congenital heart disease. The impact of these admissions on pediatric hospital resources is unknown. Our goals were to examine resource use by adults undergoing congenital heart surgery in pediatric hospitals, explore the association between high resource use (HRU) and inpatient death, and identify HRU risk factors.

Methods and Results- We obtained inpatient data from 42 pediatric hospitals from 2000 to 2008 and selected adult congenital heart (ACH) surgery admissions. We defined HRU admissions as those exceeding the 90th percentile for total hospital charges. We performed multivariable analyses using generalized estimating equations to identify risk factors for HRU. Of 97 563 congenital heart surgery admissions to pediatric hospitals, 3061 (3.1%) were adults, accounting for 2.2% of total hospital charges. The threshold for HRU was total hospital charges \geq \$213 803. Although HRU admissions comprised 10% of admissions, they accounted for 34% of charges for all ACH surgery admissions. Mortality rate was 16% for HRU admissions and 0.7% for others ($P < 0.001$). Multivariable analysis demonstrated higher case complexity: risk category 2 (adjusted odds ratio [AOR], 3.6; $P = 0.02$), risk category 3 (AOR, 13.7; $P < 0.001$), and risk category 4+ (AOR, 30.7; $P < 0.001$) as compared with risk category 1; DiGeorge syndrome (AOR, 4.2; $P = 0.006$); depression (AOR, 3.1; $P < 0.001$); weekend admission (AOR, 2.6; $P < 0.001$); and government insurance (AOR, 2.0; $P < 0.001$) as risk factors for HRU.

Conclusions- High resource use ACH surgery admissions are associated with significantly greater mortality rates. ACH admissions with greater surgical complexity, government insurance, DiGeorge syndrome, weekend admission, and depression were more likely to result in HRU.

Child Care Health Dev. 2011 Nov;37(6):841-51. doi: 10.1111/j.1365-2214.2011.01317.x.

['Down the rabbit hole': enhancing the transition process for youth with](#)

[cystic fibrosis and congenital heart disease by re-imagining the future and time.](#)

Moola FJ, Norman ME.

Graduate Department of Exercise Sciences, The University of Toronto, Toronto, ON, Canada. fiona.moola@utoronto.ca

BACKGROUND:

Although the transition from paediatric to adult clinical care is an important one, the process is far from seamless. Little is known about the transition experiences of youth with cystic fibrosis (CF) and congenital heart disease (CHD).

METHOD:

Informed by the new social studies of childhood, this qualitative study adopted a thematic analytical approach in order to explore how 50 youth and 28 parents affected by CF and CHD at a large children's hospital in Canada negotiate constructions of 'normal developmental time'--in both anticipating and dealing with the transition from adolescence to adulthood.

RESULTS:

Illness appeared to render the future as an uncertain terrain for youth living with CF and CHD. Concerns related to deteriorating health and occupational restrictions in the future were paramount for these youth. For young women with CF and CHD, the loss of 'normal' gendered roles--such as motherhood--was also a distressing future concern. For youth living with CF and their parents in particular, time was thought to be stolen and the future was abbreviated. Despite these seemingly anxiety-inducing experiences, youth and their parents demonstrated considerable creativity as they devised strategies to deal with the future and stolen time.

RECOMMENDATIONS AND CONCLUSION: In addition to challenging ideological assumptions about developmental time which may alienate youth with chronic illnesses, the results from this study suggest that attending to youth's temporal anxieties and future concerns may ultimately enhance the transition process for youth with CF and CHD.

J Am Soc Echocardiogr. 2011 Dec;24(12):1392-1399.e1. Epub 2011 Oct 14.

[Biventricular Performance in Patients with Marfan Syndrome without Significant Valvular Disease: Comparison to Normal Subjects and Longitudinal Follow-Up.](#)

Scherptong RW, Vliegen HW, van der Wall EE, Hilhorst-Hofstee Y, Bax JJ, Scholte AJ, Delgado V.

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

BACKGROUND:

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

disease.

METHODS:

Adult patients with Marfan syndrome (n = 50; mean age, 35.2 ± 12.9 years) were followed prospectively. Echocardiography was performed annually and consisted of comprehensive assessment of ventricular and valvular function. Using speckle-tracking imaging, the baseline strain values of the Marfan population were calculated and compared with the values of normal controls. The follow-up evaluations were used to assess changes in ventricular strain. The association between the incidence of adverse events (heart failure, [supra]ventricular arrhythmias, and proximal aorta surgery) and baseline strain values was investigated.

RESULTS:

Compared with controls, patients with Marfan syndrome had significantly lower peak longitudinal LV strain ($-18.9 \pm 2.3\%$ vs $-20.1 \pm 1.9\%$, $P < .01$) and RV strain ($\pm 26.9 \pm 5.2\%$ vs $\pm 29.3 \pm 4.25\%$, $P < .01$). The absolute changes in LV longitudinal, radial, and circumferential strain and RV longitudinal strain during a median 4 years of follow-up were $0.1 \pm 2.8\%$, $1.12 \pm 7.6\%$, $0.3 \pm 3.7\%$, and $0.9 \pm 5.5\%$, respectively, which was not statistically significant. Cox regression demonstrated that reduced LV or RV strain was not associated with adverse outcome (supraventricular arrhythmias, n = 3; proximal aorta surgery, n = 4).

CONCLUSIONS:

This study suggests that patients with Marfan syndrome show lower ventricular strain and strain rate values compared with healthy controls. However, no relevant changes in LV and RV function occurred during midterm follow-up in patients with Marfan syndrome without valvular disease at baseline. Although ventricular strain and strain rate were mildly reduced in patients with Marfan syndrome, this did not affect outcomes negatively in the present study.

Am J Cardiol. 2011 Nov 15;108(10):1483-8. Epub 2011 Sep 21.

[Efficacy and safety of bosentan for pulmonary arterial hypertension in adults with congenital heart disease.](#)

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

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

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 ≥ 1 dose of bosentan (mean duration of therapy 2.1 ± 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.

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.

Department of Radiology, Fatih University School of Medicine, Alparslan Turkes Cad. No. 57, Emek, Ankara 06510, Turkey. drdilekoz@gmail.com

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.

J Thorac Cardiovasc Surg. 2011 Nov;142(5):1090-7. Epub 2011 Sep 10.
[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.
Division of Thoracic and Cardiovascular Surgery, Department of Surgery,
University of Louisville, Louisville, KY, USA.
cmascio@louisvilleheartsurgery.com

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 ≥ 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.

Am J Cardiol. 2011 Nov 1;108(9):1352-61. Epub 2011 Aug 20.

[The cardiac malpositions.](#)

Perloff JK.

Ahmanson/UCLA Adult Congenital Heart Disease Center, Los Angeles, California, USA. josephperloff@earthlink.net

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

Cardiol Young. 2011 Oct;21(5):556-61. Epub 2011 Apr 19.

[Adolescents' drawings of their cardiac abnormality.](#)

Wang Q, Hay M, Clarke D, Menahem S.

School of Psychology and Psychiatry, Monash University, Melbourne, Australia. qfting@hotmail.com

Following advances in overall management and improved outcomes, an increasing number of adolescents with cardiac disease are reaching adult age. Patients in general, including adolescents, seem to have a poor knowledge of their illness, which may further reflect in a less optimal quality of life. As a guide to their knowledge of their cardiac condition, adolescents were asked to draw a diagram of their cardiac abnormality. Relatively well adolescents aged 12-20 years with a cardiac abnormality were consecutively recruited from an ambulatory setting. All were asked to draw a picture of their cardiac abnormality and describe their condition. A total of 120 patients were recruited and had conditions varying from a hyperplastic right ventricle to a small ventricular septal defect. Only 60 (50%) of the patients completed a drawing, of which one-third did so at the time of attendance. Nevertheless, there was no difference between the accuracy of the adolescents' drawings completed at home or at the clinic. Only three patients drew an accurate diagram of their congenital cardiac abnormality. A further nine patients drew a reasonably correct diagram, 13 patients a partially correct diagram, whereas 35 patients submitted

incorrect diagrams. Adolescents with congenital cardiac disease, many having been cared for since infancy with regular cardiological reviews, had a poor anatomical knowledge of their cardiac lesion, as reflected by their inability to correctly draw their abnormality. These findings suggest the need for improved strategies in developing appropriate education programmes for this patient population.

Pediatr Cardiol. 2011 Oct;32(7):891-5. Epub 2011 May 8.

[Patients with single-ventricle physiology: prognostic implications of stress testing.](#)

Bauer BS, Aboulhosn JA, Williams RJ, Child JS.

Ahmanson/UCLA Adult Congenital Heart Disease Center, David Geffen School of Medicine at UCLA, Los Angeles, CA 90095, USA.

This study used a retrospective analysis of adults with single-ventricle physiology to ascertain the predictive power of cardiopulmonary stress-testing parameters in determining patients at increased risk of suffering from adverse clinical outcomes. We found that the specific parameters of percent of maximum predicted heart rate achieved and maximum oxygen consumption were significantly correlated with adverse clinical outcomes in patients with single-ventricle congenital heart disease.

J Cardiovasc Nurs. 2011 Nov;26(6):497-503.

[Healthcare needs of adults with congenital heart disease: study of the patient perspective.](#)

Harrison JL, Silversides CK, Oechslin EN, Kovacs AH.

Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Canada.

jeanine.harrison2@uhn.on.ca

BACKGROUND:

More than 90% of infants born with congenital heart disease reach adulthood. International medical recommendations outline patient care needs in an effort to optimize patient health. There are, however, limited data focusing on the patient perspective.

OBJECTIVES:

This study investigated adult congenital heart disease patient-reported (1) barriers to medical care, (2) healthcare behaviors, and (3) concerns regarding medical, psychosocial, and lifestyle matters.

METHODS:

In this cross-sectional study, a questionnaire was distributed to all patients who attended a patient education conference.

RESULTS:

There were 123 adult congenital heart disease participants (58% female; mean age, 37 [SD, 13] years). The most common self-reported cardiac diagnoses were tetralogy of Fallot and transposition of the great arteries. Most patients did not report transportation or financial barriers to care, but did report the following: not wanting further surgery even if it was

recommended (18%), not liking to think or talk about one's heart (17%), and not understanding doctors' information; 8% of patients inaccurately considered themselves to be "cured." With regard to healthcare behaviors, more than 80% of patients reported annual family physician and dentist visits, but 34% of patients were unaware when to seek urgent medical attention. Patients reported moderate to extreme concern about the following medical topics: heart rhythm problems (82%), infections (74%), and understanding treatment options (71%). Patients most often reported moderate to extreme concern about the following lifestyle and psychosocial topics: physical activity (77%), insurance (72%), assuming increased health responsibility (73%), diet (71%), mental health (60%), and death and dying (57%).

CONCLUSIONS:

This study provides important information about 3 specific areas. First, there are potential barriers to care beyond financial and transportation challenges. Second, many patients require education regarding when to seek urgent medical attention. Third, the concerns of this patient population are not limited to medical information. A patient-centered educational program is recommended.

Int J Cardiol. 2011 Oct 20;152(2):171-6. Epub 2010 Aug 1.

[The prevalence and risk factors for cholelithiasis and asymptomatic gallstones in adults with congenital heart disease.](#)

Shiina Y, Toyoda T, Kawasoe Y, Tateno S, Shirai T, Matsuo K, Mizuno Y, Ai T, Niwa K.

Department of Adult Congenital Heart Disease and Pediatrics, Chiba Cardiovascular Center, 575 Tsurumai, Ichihara, Chiba 290-0512, Japan.

PURPOSE:

Cyanosis is considered to be a risk factor for cholelithiasis which is an important complication of cyanotic congenital heart disease (CCHD) in adults. In this study, the prevalence of cholelithiasis and asymptomatic calcium bilirubinate gallstones was evaluated in adults with congenital heart disease (CHD). Furthermore, risk factors for this potentially high risk complication were assessed.

MATERIALS AND METHODS:

Subjects were derived from 114 consecutive congenital patients who visited our center from May 2008 to January 2009. For analyses of risk factors, we divided them into 4 groups: group A, 15 CCHD patients without reparative surgery (7 men, 31.8 ± 7.0 years old); group B, 41 CCHD patients rendered acyanotic by reparative surgery (21 men, 32.5 ± 11.8 years old); group C, 23 unoperated acyanotic CHD patients (11 men, 42.4 ± 16.4 years old); and group D, 35 patients who were acyanotic before and after operation (18 men, 36.3 ± 14.8 years old). Gallstones were identified by abdominal ultrasound and risk factors were analyzed by a multivariate logistic regression model.

RESULTS:

Cholecystectomy was performed in 5/114 (4.3%), asymptomatic gallstones were seen in 16/114 (14%), and symptomatic gallstones except for patients after cholecystectomy were seen in 7/114 (6.1%). In group A, 4 (27%) with gallstones underwent cholecystectomy ($p < 0.01$). Non-cholesterol gallstones were observed in 5 patients (33%) in group A, 12 patients (29%) in group B, nobody in group C, and 3 patients (8.6%) in group D. By a multivariate logistic regression model, CCHD by nature regardless of repair, prolonged cyanosis periods, higher frequency of cardiopulmonary bypass (CPB), and lower platelet counts were significant factors predicting gallstones (odds ratio 4.48, 1.08, 3.96, and 0.87, 95% CI, 1.14-17.5, 1.00-1.18, 1.65-9.54, and 0.75-0.99, respectively).

CONCLUSIONS:

The prevalence of cholelithiasis and asymptomatic gallstones is significantly high in CCHD patients regardless of cardiac repairs. CCHD by nature, prolonged cyanosis durations, high frequency of CPB and low platelet counts have influences on gallstone formation in adults with CHD.