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# ISACHD Newsletter

## President's Message

by Curt J Daniels

Dear ISACHD Members,

With summer 2012 coming to a close, ISACHD is ramping up its activities centered around ACHD Global Health Initiatives. Erwin Oechslin (Toronto, CA, ISACHD Canadian Regional Representative and Chair ACHD Global Education WG), Gary Webb (Cincinnati, OH, USA, ISACHD past president), and their team of ACHD educators have been developing web-based ACHD educational programming to eventually be utilized to communicate and educate our international colleagues, particularly those in underserved areas less able to attend symposia and conferences. We look forward to an update on their tremendous work and progress at our upcoming ISACHD meeting AHA in November 2012 in Los Angeles.

The ACHD Global Health WG continues to move forward and has scheduled upcoming meetings with international health organizations to develop partnerships and eventually plan missions to underserved CHD programs needing ACHD guidance and care of their patients. We will be asking work group members who have volunteered toward this effort to assist with planning once we solidify our international health care partners and establish a working relationship--more to come at ISACHD meting AHA in November 2012.

Philip Moons (Leuven, Belgium, ISACHD Secretary) and Koichiro Niwa (Tokyo, Japan, ISACHD Treasurer and Asian Pacific Regional Representative) continue their efforts developing an international ACHD multi-center research. Soon, programs interested and able to participate will be invited to join this collaborative effort.

ESC Congress 2012 is just a few weeks away (August 25-29th) in Munich Germany. There are several GUCH sessions planned and will provide updated information regarding the following:

- GUCH and Pulmonary Hypertension
- GUCH and Heart Failure
- Imaging Modalities in GUCH
- ISACHD session on the 27th, 1630-1800: GUCH for the General Cardiologist

We look forward to seeing you in Munich!

Please visit the ISACHD website at www.isachd.org to learn more about ISACHD, visit the journal watch page and find the latest ACHD publications, view upcoming conferences endorsed by ISACHD, and register to become a member of ISACHD.

Academic Medical Center
Amsterdam, The Netherlands

**Representative, USA**
William Davidson, MD
The Pennsylvania State University
Hershey, PA, USA

**Representative, South America**
Luis Alday, MD
Hospital de Niños Córdoba, Argentina

**Representative, Europe**
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**Representative, Asia-Pacific** Koichiro Niwa, MD
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**Representative, Canada**
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Ontario, Canada

**Representative, IACHD Nursing Network** Desiree Fleck, PhD, CRNP
Children's Hospital of Philadelphia
Philadelphia, USA

**Journal Watch Editor**
Philip Moons, PhD, RN

Looking forward to seeing you soon,

Curt J. Daniels
President

## Invitation to the Social Event in Munich, Monday August 27, for All ISACHD Members

It is our great pleasure to welcome you to Munich during the ESC-Congress. For all "GUCH-friends" and members of the ESC Working Group on Adults with Congenital Heart Disease, we have arranged an evening reception and a guided tour in the Ayinger Brewery, on Monday 27 August 2012 starting at 20:00.

Aying is a small village southeast of Munich, just half an hour from the Bavarian capital.The Ayinger Brewery is a "typical piece of Upper Bavaria." To learn more about the specialty beers, we have organised a guided tour for you in the brewery.

We hope that you will enjoy the charming atmosphere of a traditional Bavarian brewery. This place blends nicely into its natural surroundings and will allow attendees to meet old friends in a relaxed atmosphere.

Due to the ever-increasing financial constraints, we unfortunately cannot take over the entire costs and are forced to charge a fee of 30 Euros per person. Note that transportation from the fairground to Aying and return to Munich Central City is provided.

Further information regarding the location and a time table will follow shortly. We are looking forward to hearing from you and welcoming you to Munich soon.
Warmest regards,

Andreas Eicken Pedro Trigo Trindade Harald Kaemmerer

# ESC Congress

## WG on GUCH Evening in Munich: You Can Still Register!

Still not registered yet? Do not miss the opportunity to meet your friends and colleagues in the charming atmosphere of a traditional Bavarian brewery. Read the invitation and send back the coupon (here attached).

Kind regards,

ESC Office for Working Groups
Membership Service Department
European Society of Cardiology (ESC)

Email: workinggroups@escardio.org
Visit our ESC Web Site: http://www.escardio.org

## ESC Congress Program:

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|---|---|---|---|---|
| 25 Aug 2012 | New insights into the longterm outcome of congenital heart disease | 11:00-12:30 | Rome - Village 8 | Grown-up congenital heart disease and surgery |
| 26 Aug 2012 | Multi-modality image guidance in 2D and 3D for structural interventions | 09:00-10:30 | Room B12 - Hands-On Tutorials | Aortic valve disease |
| 26 Aug 2012 | Poster session 2: Clinical impact of imaging in congenital heart disease | 14:00-18:00 | Posters - Village 10 | Congenital heart disease imaging |
| 26 Aug 2012 | Role of imaging in interventions on structural heart disease | 16:30-18:00 | Helsinki - Village 8 | Congenital heart disease imaging |
| 26 Aug 2012 | Poster session 2: New predictors of outcome in congenital heart disease | 14:00-18:00 | Posters - Village 10 | Grown-up congenital heart disease and surgery |
| 26 Aug 2012 | Ebstein's malformation: recent advances | 08:30-10:00 | Helsinki - Village 8 | Grown-up congenital heart disease and surgery |
| 26 Aug 2012 | The new 2012 ESC/EACTS Guidelines on the management of valvular heart disease | 11:00-12:30 | Tirana - Central Village | Valvular heart disease, other |
| 26 Aug 2012 | Advances in tricuspid regurgitation | 14:00-15:30 | Sarajevo - Village 8 | Valvular heart disease, other |
| 27 Aug 2012 | How to manage pulmonary hypertension in valvular heart disease | 08:30-10:00 | Helsinki - Village 8 | Valvular heart disease, other |
| 27 Aug 2012 | Heart failure in adult congenital heart disease | 11:00-12:30 | Helsinki - Village 8 | Grown-up congenital heart disease |

| | | | | |
|---|---|---|---|---|
| | | | | and surgery |
| 27 Aug 2012 | Decision making in patients with multivalvular disease | 12:45-13:45 | Helsinki - Village 8 | Valvular heart disease, other |
| 27 Aug 2012 | Arrhythmias in congenital heart disease: new insights | 14:00-15:30 | Rome - Village 8 | Grown-up congenital heart disease and surgery |
| 27 Aug 2012 | Difficult echocardiography in adult congenital heart disease | 15:40-16:20 | Helsinki - Village 8 | Congenital heart disease imaging |
| 27 Aug 2012 | Congenital heart disease: new developments for the general cardiologist | 16:30-18:00 | Rome - Village 8 | Grown-up congenital heart disease and surgery |
| 29 Aug 2012 | Poster session 7: Exercise capacity and quality of life in congenital heart disease | 08:30-12:30 | Posters - Village 10 | Grown-up congenital heart disease and surgery |

## Regional News:

### News from Latin America

*By Luis Alday*


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

### News from United States of America

*By Bill Davidson*

Registration is now open for AHA Scientific Sessions 2012 in Los Angeles, CA.

### WG on Education

*By Erwin Oechslin, Toronto (Canada)*

The WG on Education of the ISACHD has the following objectives:

- To establish an Internet-based ACHD Learning Centre;
- To develop a basic teaching course in ACHD for adult cardiology trainees;
- To endorse national/international conferences/courses related to

ACHD.

We have established an Editorial Board for the ACHD Learning Centre, which consists of a balanced group of individuals representing Europe, North America, Asia, physicians, non-physicians, and ACHD trainees.

Members of the Editorial Board for the ACHD learning Centre Committee include the following:

- Gary Webb (USA), Chair
- Els Pieper (NL)-Lead for the ACHD basic teaching course
- Richard Krasuski (USA)
- Jonathan Windram (CA)
- Naser Ammash (USA)
- Kate English (UK)
- Helmuth Baumgartner (DE)
- David Drajpuch (USA)
- Philip Moons (BE)
- Teiji Akagi MD, Okayama (JP)
- Erwin Oechslin (CA)

The Objectives of the Editorial Board are as follows:

- To identify and to collect existing teaching resources and education material in ACHD, which will be made available through the ACHD Learning Centre (simple ACHD learning address);
- To develop an internet-based basic teaching course in ACHD (target audience: trainees in adult cardiology), led by Dr. Els Piper, Groningen (NL);
- To seek input from both education and IT professionals.

Our past requests to submit links of teaching resources were not successful at all. We changed the strategy and sent an email to more than 3000 recipients to introduce the concept of an Internet-based ACHD Learning Centre. The responses were very encouraging: we received excellent feedback and links to existing teaching resources.

**If you know an existing teaching resource or if you want to link your teaching/education website to the Internet-based ACHD Learning Centre, please email the address of existing web-based teaching resources to gary.webb@cchmc.org and to erwin.oechslin@uhn.ca.**

**Endorsement of ACHD Meetings**
The following meetings have been endorsed:

- Adult Congenital Heart Disease: The Imaging and Intervention Revolution; September 21-22, 2012, Luxe Sunset Boulevard Hotel, Los Angeles, California. This course is organized by the Office of Continuing Medical Education, David Geffen School of Medicine at UCLA, and the Ahmanson/UCLA Adult Congenital Heart Disease Center.. Course Chair: Jamil Aboulhosn, MD. Online registration at www.cme.ucla.edu/courses (click on Adult

Congenital Heart Disease, and click the registration button).
- Sport and Heart Disease: November 30 - December 1, 2012, German Heart Centre, Lazarettstrasse 36, Munich (Germany). This symposium is organized by the German Heart Centre., Munich; Scientific Chair: Alfred Hager, MD. Registration at www.sport-CHD.de.

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

## Journal Watch

Eur J Cardiothorac Surg. 2012 Aug 14. [Epub ahead of print]
Closure of secundum atrial septal defects in the adult and elderly patients.
Nyboe C, Fenger-Grøn M, Nielsen-Kudsk JE, Hjortdal V.
**Source**
Department of Cardiothoracic Surgery, Aarhus University Hospital, Aarhus, Denmark.
**Abstract**
**Objectives:** Treatment of atrial septal defect (ASD) in adults is still controversial, and with older age the likelihood of treatment is decreased. The aim of this study was to investigate the effect of ASD closure in adults and especially in the elderly in our institution in a retrospective review.
**Methods:** Adult patients (n = 220) underwent surgical or catheter closure for an isolated ASD at Aarhus University Hospital from 1990 to 2008. Eleven were lost to follow-up and 13 had cardiac comorbidity, and thus 196 were eligible for analysis in the study. Hospital records were reviewed and symptoms and echocardiographic findings registered preoperatively and at 3-month follow-up. Patients were divided into Group I (n = 117): between 18 and 50 years old and Group II (n = 79): older than 50 years. Symptoms and echocardiographic findings before and 3 months after closure were compared within and between the two groups.
**Results:** One patient (0.5%) died during follow-up. Complications occurred in 16% in Group I and 22% in Group II. There was an absolute risk reduction of -62 and -52%, respectively in right ventricle (RV) dilation after operation. Atrial fibrillation was noticed preoperatively in 6% of the young and 47% of the elderly, with an absolute risk reduction after treatment of -20% in Group II ($P < 0.0001$). Subjective symptoms occurred in 75% in Group I and 99% in Group II with a postoperative reduction to 43 and 67%, respectively. In Group I, 70% felt an improvement of symptoms while this was true for 89% in Group II.
**Conclusions:** Symptoms and RV dilation are more pronounced in the elderly (>50 years), but reversibility is the same as in the young (<50 years) patients. The elderly benefit substantially from ASD closure. Based on these data, ASD closure is recommendable even after the fifth decade.

Congenit Heart Dis. 2012 Aug 14. doi: 10.1111/j.1747-0803.2012.00708.x. [Epub ahead of print]
Iron Deficiency Anemia Detection from Hematology Parameters in Adult Congenital Heart Disease Patients.
Martínez-Quintana E, Rodríguez-González F.
**Source**

Cardiology Service, Insular-Materno Infantil University Hospital, Las Palmas de Gran Canaria, Spain.

**Abstract**

**Introduction:** Iron deficiency anemia is the most common single cause of anemia worldwide. The purpose of our study was to estimate the prevalence of anemia in adult congenital heart disease (ACHD) patients, compare different hematology parameters between hypoxemic and nonhypoxemic ACHD patients, and determine which parameters detect iron deficiency anemia in hypoxemic ACHD patients.

**Methods:** ACHD patients were studied and blood samples collected for determination of hemoglobin, derived red cell indices, serum iron, apoferritin, total iron-binding capacity, transferrin saturation index, C-reactive protein (CRP), and N-terminal proB-type natriuretic peptide (NT-proBNP) levels.

**Results:**Two hundred seventy-eight ACHD patients, mean age 31.6 ± 14.3 years old, were studied. One hundred sixty-seven (60%) patients were male. Two hundred forty-five patients were nonhypoxemic and 33 patients were hypoxemic. Hypoxemic ACHD patients had significant higher hemoglobin concentration (g/dL) (17.5 ± 3.5 vs. 14.6 ± 1.7, P <.001), red cell distribution width (RDW) (%) (17.0 ± 3.3 vs. 14.1 ± 7.6, P <.034), apoferritin (ng/mL) (19.8 [4.1-147.2] vs. 38.0 [6.7-191.2], P =.019), CRP (mg/dL) (0.50 [0.0-3.8] vs. 0.12 [0.0-1.4], P <.001), and NT-proBNP (pg/mL) (409.3 [33.3-9830.8] vs. 5.2 [0.0-1068.4], P <.001) levels than nonhypoxemic ACHD patients. Serum iron, total iron-binding capacity, and transferrin saturation index were not statistically significant between hypoxemic and nonhypoxemic ACHD patients. In the hypoxemic group, 15 (45%) patients had apoferritin levels <20 ng/mL and eight (24%) patients developed microcytosis and hypochromia. A RDW above the normal range (>14.5%) in hypoxemic ACHD patients allowed the detection of an apoferritin level <20 ng/mL with a sensitivity of 93%.

**Conclusions:** RDW seems to be a useful and economic tool to detect low serum apoferritin levels in hypoxemic ACHD patients.

Congenit Heart Dis. 2012 Aug 14. doi: 10.1111/j.1747-0803.2012.00707.x. [Epub ahead of print]

[Hemodynamic Characteristics of Cyanotic Adults with Single-ventricle Physiology without Fontan Completion.]

Saab FG, Aboulhosn JA.

**Source**

David Geffen School of Medicine, University of California, Los Angeles, Calif, USA.

**Abstract**

**Objective:** The aim of the current study is to describe the long-term clinical and hemodynamic characteristics of adult patients with single-ventricle physiology who have not undergone the Fontan operation and consequently have remained cyanotic.

**Design:** Adult patients at the Ahmanson/UCLA Adult Congenital Heart Disease Center with non-Fontan single-ventricle physiology who had undergone cardiac catheterization between 2005 and 2011 were included. Echocardiographic and cardiac catheterization data were reviewed.

**Results:** Mean estimated single ejection fraction was 56±8%. Eight of 13 subjects had documented E/E' data with a mean of 6.44. Seven subjects had both A' and E' data documented, of which two subjects exhibited A'>E'. Mean ventricular end-diastolic pressure (MVEDP)

was15.77±4.91mm Hg, and was> 12 mm Hg in eight of the 13 patients (62%). MVEDP was also analyzed by age, and in the single-ventricle patients was 13.55±4.12 mm Hg in those <50 years of age, compared with 20.75±1.89 mm in those >50 years of age (P=.003). MVEDP prior to inhaled pulmonary vasodilator administration was 14.75±5.5 mm Hg, compared to15.00±6.78 mm Hg in the postvasodilator group (P=.48). Subjects with end-diastolic pressure (EDP) <12 had a mean brain natriuretic peptide (BNP) of 108±187 pg/mL , while subjects with EDP >12 had a mean BNP of 234.5±127.36 pg/mL (P=.11). **Conclusions:** Cyanotic adult single-ventricle patients not palliated with Fontan completion have preserved single-ventricle systolic function but develop elevated ventricular filling pressure with increasing age. Only invasive hemodynamic measurements demonstrated elevated ventricular filling pressures, while traditional echo/Doppler criteria for diastolic dysfunction were not met. Aging with cyanotic single-ventricle physiology is associated with a greater degree of filling pressure elevations than in the general population. Single-ventricle patients with EDP >12 exhibited markedly elevated BNP compared to those with normal EDP.

Catheter Cardiovasc Interv. 2012 Aug 6. doi: 10.1002/ccd.24594. [Epub ahead of print]

Off-label use of percutaneous pulmonary valved stents in the right ventricular outflow tract: Time to rewrite the label?

Boshoff D, Cools B, Heying R, Troost E, Kefer J, Budts W, Gewillig M.

**Source**

Paediatric and Adult Congenital Heart Unit, University Hospitals KU Leuven and UC Louvain, Belgium.

**Abstract**

**Introduction:** Percutaneous pulmonary valve implantation is now considered feasible and safe. "Native" right ventricular outflow tract (RVOT), small diameter conduits (less than 16mm) and relatively large RVOT with a dynamic outflow aneurysm are currently considered off-label uses. Extending indications creates concerns of safety, ethics, reimbursement and liability.

**Aim of Study:** To report the safety and feasibility of off-label application of percutaneous pulmonary valve implantation.

**Design:** Retrospective analysis of prospectively collected data.

**Patients and Methods:** Off-label indications: conduit-free RVOT or patients with an existing but undersized conduit.

**Results:** Twenty-one Melody® valves and 2 Sapien® valves were successfully implanted in 23 patients (16.9 years; range 6.1 to 80.5 years). In 22 patients, pre-stenting was performed 4.8 months (range 0 to 69.2) before valve implantation (15 covered and 13 bare stents). Stent endothelial ingrowth was allowed for at least 2 months prior to implantation of the percutaneous valve if stent stability or sealing by the covering was presumed to be insufficient. Group 1 patients (n=8) had a "conduit-free" RVOT after transannular/infundibular patch and after pre-stenting underwent PPVI, with a final RVOT diameter of 21.5mm [range 16 to 26mm]. Group 2 patients consisted of 2 elderly patients with pulmonary valve stenosis and severe RVOT calcifications. Group 3 (n=13) had an existing conduit (nominal 15.9±3.2mm, range10 to 20mm). The conduit was augmented from 14.7±3.5 mm to 20±1.6mm with PPVI. The RVOT preparation and valve implantations were uneventful.

**Conclusions:** PPVI is safe and feasible in selected patients with an off-

label indication. Creating an adequate "landing zone" by pre-stenting makes the procedure safe and predictable. Updating the indications for PPVI should be considered. © 2012 Wiley Periodicals, Inc.

Int J Cardiol. 2012 Aug 9. [Epub ahead of print]
Outcome in adult patients after arterial switch operation for transposition of the great arteries.
Kempny A, Wustmann K, Borgia F, Dimopoulos K, Uebing A, Li W, Chen SS, Piorkowski A, Radley-Smith R, Yacoub MH, Gatzoulis MA, Shore DF, Swan L, Diller GP.
**Source**
Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton and Harefield NHS Foundation Trust, London, UK.
**Abstract**
**Background:** The arterial switch operation (ASO) is currently the treatment of choice in neonates with transposition of the great arteries (TGA). The outcome in childhood is encouraging but only limited data for long-term outcome into adulthood exist.
**Methods and Results:** We studied 145 adult patients (age>16, median 25years) with ASO followed at our institution. Three patients died in adulthood (mortality 2.4/1000-patient-years). Most patients were asymptomatic and had normal left ventricular function. Coronary lesions requiring interventions were rare (3 patients) and in most patients related to previous surgery. There were no acute coronary syndromes. Aortic root dilatation was frequent (56% patients) but rarely significant (>45mm in 3 patients, maximal-diameter 49mm) and appeared not to be progressive. There were no acute aortic events and no patient required elective aortic root surgery. Progressive neo-aortic-valve dysfunction was not observed in our cohort and only 1 patient required neo-aortic-valve replacement. Many patients (42.1%), however, had significant residual lesions or required reintervention in adulthood. Right ventricular outflow tract lesions or dysfunction of the neo-pulmonary-valve were frequent and 8 patients (6%) required neo-pulmonary-valve replacement. Cardiac interventions during childhood (OR 3.0, 95% CI 1.7-5.4, P<0.0001) were strong predictors of outcome (cardiac intervention/significant residual lesion/death) in adulthood.
**Conclusions:** Adult patients with previous ASO remain free of acute coronary or aortic complications and have low mortality. However, a large proportion of patients require re-interventions or present with significant right sided lesions. Life-long cardiac follow-up is, therefore, warranted. Periodic noninvasive surveillance for coronary complications appears to be safe in adult ASO patients.

Ann Thorac Surg. 2012 Aug 9. [Epub ahead of print]
Impact of Single-Ventricle Physiology on Death After Heart Transplantation in Adults With Congenital Heart Disease.
Karamlou T, Diggs BS, Welke K, Tibayan F, Gelow J, Guyton SW, Slater MS, Broberg C, Song HK.
**Source**
Congenital Cardiac Center, Pediatric Cardiac Surgery, Seattle Children's Hospital, Seattle, Washington.
**Abstract**
**Background:** Prevalence of univentricular (1V) anatomy over time and whether 1V anatomy is associated with early death after heart transplant (HTx) among recipients with adult congenital heart disease (ACHD) is unknown. We investigated changes in case-mix over time, 1V vs

biventricular (2V) status, and the effect of 1V anatomy on death after HTx among ACHD recipients.
**Methods:** The Nationwide Inpatient Sample (NIS) was used to identify ACHD HTx recipients in the United States aged 14 years or older from 1993 to 2007, divided into era 1 (1993 to 2000) and era 2 (2001 to 2007). In-hospital death was compared among recipients with 1V and 2V anatomy. Multivariable determinants associated with an increased risk of in-hospital death were sought with logistic regression models.
**Results:** From a national estimate of 509 ACHD recipients, 143 were 1V and 366 were 2V. Overall, 1V in-hospital mortality (23%) was higher than for 2V (8%; $p < 0.001$) and remained associated with in-hospital death after adjustment for other factors (odds ratio, 3.9; 95% confidence interval, 1.29 to 11.74; $p = 0.02$). All 1V diagnoses had higher mortality than all 2V diagnoses. Despite minor fluctuations, the proportion of 1V patients did not increase over time (era 1, 36%; era 2, 30%; $p = 0.46$).
**Conclusions:** Overall case-mix of ACHD recipients (1V vs 2V) has not changed over time. Initial 1V anatomy increases post-HTx death among ACHD recipients, whereas 2V patients have mortality rates similar to non-CHD recipients. National and international transplant registries should include specific CHD diagnoses because this factor plays such a large role in determining early outcomes.

Int J Cardiol. 2012 Aug 7. [Epub ahead of print]
Rates and determinants of progressive aortic valve dysfunction in aortic coarctation.
Luijendijk P, Stevens AW, de Bruin-Bon RH, Boekholdt SM, Vriend JW, Vliegen HW, Bouma BJ, Mulder BJ.
**Source**
Department of Cardiology, Academic Medical Centre, Amsterdam, The Netherlands; Interuniversity Cardiology Institute of The Netherlands, Utrecht, The Netherlands.
**Abstract**
**Purpose:** Aortic valve dysfunction is common in coarctation patients(CoA). Bicuspid aortic valve (BAV) in CoA is associated with aortic valve stenosis (AS), aortic valve regurgitation (AR), and ascending aortic dilatation. The aim of this study was to evaluate the progression of and predictors for aortic valve dysfunction in CoA.
**Methods:** 96 CoA patients prospectively underwent echocardiography twice between 2001 and 2010. AS was defined as an aortic valve gradient ≥20mmHg, AR as none/minor, or moderate/severe. Aortic dilatation as an ascending aortic diameter ≥37mm.
**Results:** All patients (median age 28.0years, range 17-61years; male 57%) were followed with a median follow-up of 7.0years. Sixty patients (63%) had BAV. At baseline 10 patients had AS (10%, 9 BAV), 6 patients AR (6%, 3 BAV) and 11 patients aortic dilatation (11%, 11 BAV). At follow-up 15 patients had AS (15%, 13 BAV) and 12 patients AR. (13%, 8 BAV). Median AS progression was 1.1mmHg/5years (range - 13-28). Determinants for AS at follow-up were age (ß=0.20, P=0.01), aortic dilatation (ß=4.6, P=0.03), and baseline aortic valve gradient (ß=0.93, P<0.001). BAV was predictive for AR. (ß=0.91, P=0.049).
**Conclusion:** Progression of AS in adult CoA patients is mild in this young population. Older age, aortic dilatation and the baseline aortic valve gradient are determinants for AS at follow-up. BAV is predictive for AR. These findings point towards a common embryological pathway of both valvular and aortic disease in CoA.

Heart. 2012 Aug 6. [Epub ahead of print]
Exercise capacity and stroke volume are preserved late after tetralogy repair, despite severe right ventricular dilatation.
O'Meagher S, Munoz PA, Alison JA, Young IH, Tanous DJ, Celermajer DS, Puranik R.

**Source**

The University of Sydney, Sydney, Australia.

**Abstract**

**Objectives:** To assess if exercise capacity and resting stroke volume are different in tetralogy of Fallot (TOF) repair survivors with indexed RV (right ventricle) end-diastolic volume (RVEDVi) more versus less than 150 ml/m(2), a currently suggested threshold for pulmonary valve replacement (PVR).
**Design:** Cross-sectional study.
**Setting:** Single-centre adult congenital heart disease unit.
**Patients:** 55 consecutively eligible patients with repaired TOF (age at repair 2.3±1.9 years; age at evaluation 26.2±8.8 years; NYHA Class I or II).
Interventions: Cardiovascular MRI (1.5T) and cardiopulmonary exercise test.
**Main Outcome Measures:** Biventricular volumes and function; exercise capacity.
**Results:** 20 patients had RVEDVi below, and 35 had RVEDVi above 150 ml/m(2), at time of referral. In the >150 ml/m(2) group, fractional pulmonary regurgitation was higher (41±8 vs 31±8%, p<0.001). Although RV ejection fraction (EF) was lower (47±7 vs 54±6%, p=0.007), indexed RV stroke volume was higher (87±14 vs 64±10 ml/m(2), p<0.001) in the >150 ml/m(2) group. There were no significant differences in LVEF, indexed LV stroke volume or exercise capacity (% predicted peak work: 90±17 vs 89±11% and; % predicted VO(2) peak: 84±17 vs 87±12%).
**Conclusions:** Exercise capacity and stroke volume are maintained with RVEDVi above compared with below a commonly used cut-off for PVR surgery. Optimal timing for PVR, thus, remains unclear.

Am J Cardiol. 2012 Aug 2. [Epub ahead of print]
Usefulness of Serum Brain Natriuretic Peptide to Predict Adverse Events in Patients With the Eisenmenger Syndrome.
Reardon LC, Williams RJ, Houser LS, Miner PD, Child JS, Aboulhosn JA.

**Source**

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

**Abstract**

The aim of this study was to evaluate the prognostic value of brain natriuretic peptide (BNP) in outpatients with the Eisenmenger syndrome (ES). BNP is often elevated in patients with cyanotic congenital heart disease. The clinical utility of BNP in patients with cyanotic congenital heart disease and the ES has not been clearly delineated. Records of adults with ES who had undergone serum BNP measurement were reviewed. The primary end point was death or heart failure admission. Fifty-three patients were included, with 15 patients (28%) meeting the primary end point (death in 7, heart failure hospitalization in 8). Mean and median baseline BNP in patients meeting the primary end point were 322 ± 346 and 179 pg/ml, compared to 100 ± 157 and 41 pg/ml in those not meeting the primary end point (p = 0.0029). A Cox proportional-hazards model using baseline BNP between the 2 groups

yielded a hazard ratio of 1.84 (95% confidence interval [CI] 1.19 to 2.85, p = 0.006). The relative risk for baseline BNP level >140 pg/ml was 4.62 (95% CI 1.80 to 11.3, p = 0.008). Patients who met the primary end point increased their BNP levels by 42.5 pg/ml per year (95% CI 12.09 to 72.95, p = 0.006) compared to 7.2 pg/ml per year (95% CI 2.01 to 12.47, p = 0.007) in patients who did not meet the primary end point. In conclusion, elevated BNP levels are predictive of death or heart failure admission in patients with the ES. A serum BNP level >140 pg/ml is a useful tool in identifying high-risk patients.

Am J Cardiol. 2012 Aug 1. [Epub ahead of print]
Comparison of Aortic Root Diameter to Left Ventricular Outflow Diameter Versus Body Surface Area in Patients With Marfan Syndrome.
Shiran H, Haddad F, Miller DC, Liang D.

**Source**

Department of Cardiovascular Medicine, Stanford University, Stanford, California.

**Abstract**

Aortic root dilation is important in the diagnosis of familial aortic syndromes, such as Marfan syndrome, and an important risk factor for aortic complications, such as dissection or rupture. Transthoracic echocardiography reliably measures the absolute aortic root size; however, the degree of abnormality of the measurement requires correction for the expected normal aortic root size for each patient. The expected normal size is currently predicted according to the body surface area (BSA) and age. However, the correlation between root size and BSA is imperfect, particularly for older patients. A potential exists to improve the diagnosis and treatment of patients with aortic disease, with an improved estimation of normal aortic root size. A reference size derived from within the cardiovascular system has been hypothesized to provide a more direct correlation with the aortic root size. Images from the Stanford echocardiography database were reviewed, and measurements of the aortic root and internal dimensions were performed in a control cohort (n = 150). The measurements were repeated in adult patients with Marfan syndrome (n = 70) on serial echocardiograms (145 total studies reviewed). Of the 150 control patients, excellent correlation was found between the aortic root and left ventricular outflow tract diameters, r(2) = 0.67, and r(2) = 0.34 with BSA (p <0.0001, for both). More importantly, using the left ventricular outflow tract to predict the normal aortic root size, instead of the BSA and age, improved the diagnostic accuracy of aortic root measurements for diagnosing Marfan syndrome. In conclusion, an internal cardiovascular reference, the left ventricular outflow tract diameter, can improve the diagnosis of aortic disease and might provide a better reference for the degree of abnormality.

J Interv Cardiol. 2012 Jul 23. doi: 10.1111/j.1540-8183.2012.00755.x. [Epub ahead of print]
Safety and Long-Term Outcome of Modified Intracardiac Echocardiography-Assisted "No-Balloon" Sizing Technique for Transcatheter Closure of Ostium Secundum Atrial Septal Defect.
Rigatelli G, Dell'avvocata F, Cardaioli P, Giordan M, Dung HT, Nghia NT, Daggubati R, Nanjiundappa A.

**Source**

From the Section of Adult Congenital and Adult Heart Disease, Cardiovascular Diagnosis and Endoluminal Interventions, Rovigo

General Hospital, Rovigo, Italy From the Interventional Cardiology, Thonh Nhat Hospital, Ho Chi Minh, Vietnam From the Interventional Cardiology Department, Cho Rey Hospital, Ho Chi Min City, Vietnam From the Brody School of Medicine at East Carolina University, Greenville, North Carolina From the CAMC Vascular Center of Excellence, West Virginia University, Charleston, West Virginia.

**Abstract**

**Background:** The need for sizing the secundum atrial septal defect (ASD) with the balloon sizing technique is still debated at least in adult patients. We sought to prospectively evaluate the effectiveness of intracardiac echocardiography (ICE)-aided sizing technique for transcatheter closure of secundum ASD, without using a balloon sizing.

**Methods:** In a prospective 5-year registry, we enrolled 81 patients (mean age 48 ± 13.7 years, 54 females) who had been referred to three different centers for catheter-based closure of secundum ASD. Eligible patients underwent ICE study and closure attempt. In a preliminary group of 21 patients, sizing balloon was performed under ICE guidance to assess the value of rim thickness necessary for device anchorage. In the remaining 60 patients, the retrieved value of the rim thickness was measured on ICE and used as key points to measure the defect and select the device.

**Results:** In the preliminary group of patients, the value of thickness at point of initial deflection by the balloon was 1.23 ± 0.1 mm. ASD diameter in the study group was measured at the point of rim with at least 1.2 mm and the mean ASD diameter was 26.2 ± 10.1 mm. Rates of procedural success, predischarge occlusion, and major complications rate were 100%, 93.3%, and 0%, respectively. On mean follow-up of 5.4 ± 1.8 years, the occlusion rate was 98.7% with no long-term complications.

**Conclusions:** Our novel ICE-sizing technique appears to be safe and effective in adult patients, thus eventually minimizing overestimation, costs, and potential complications of balloon sizing. (J Interven Cardiol 2012;**:1-7).

Eur Heart J. 2012 Jul 19. [Epub ahead of print]

Cardiac outcomes in adults with supravalvar aortic stenosis.

Greutmann M, Tobler D, Sharma NC, Vonder Muhll I, Mebus S, Kaemmerer H, Schuler PK, Deanfield JE, Beauchesne L, Salehian O, Hoffmann A, Golovatyuk V, Oechslin EN, Silversides CK.

**Source**

Adult Congenital Heart Disease Program, University Hospital Zurich, Zurich, Switzerland.

**Abstract**

**Aims:** Supravalvar aortic stenosis is a rare form of left ventricular outflow tract obstruction that is often progressive in childhood. Little data are available on outcomes in the adult population. Our aim was to define cardiac outcomes in adults with supravalvar aortic stenosis.

**Methods and results:** This is a multicentre retrospective study of cardiac outcomes in adults (≥18 years) with supravalvar aortic stenosis. We examined: (i) adverse cardiac events (cardiovascular death, myocardial infarction, stroke, heart failure, sustained arrhythmias, and infective endocarditis) and (ii) the need for cardiac surgery in adulthood. One hundred and thirteen adults (median age at first visit 19 years; 55% with Williams-Beuren syndrome; 67% with surgical repair in childhood) were identified. Adults without Williams-Beuren syndrome had more severe supravalvar aortic stenosis and more often associated left

ventricular outflow tract obstructions (P < 0.001). In contrast, mitral valve regurgitation was more common in patients with Williams-Beuren syndrome. Eighty-five per cent of adults (96/113) had serial follow-up information (median follow-up 6.0 years). Of these patients, 13% (12/96) had an adverse cardiac event and 13% (12/96) had cardiac operations (7 valve repair or replacements, 4 supravalvar aortic stenosis repairs, 1 other). Cardiac surgery was more common in adults without Williams-Beuren syndrome (P = 0.007). Progression of supravalvar aortic stenosis during adulthood was rare.
**Conclusion:** Adults with supravalvar aortic stenosis remain at risk for cardiac complications and reoperations, while progression of supravalvar aortic stenosis in adulthood is rare. Valve surgery is the most common indication for cardiac surgery in adulthood.

Cardiol Clin. 2012 Aug;30(3):383-94. Epub 2012 Jun 6.
Congenital heart disease in pregnancy.
Franklin WJ, Gandhi M.
Source
Texas Adult Congenital Heart Disease Program, Texas Children's Hospital, Departments of Pediatrics and Medicine, Baylor College of Medicine, 6621 Fannin Street, 20th Floor West Tower, MC 19-345C, Houston, TX 77030, USA.
**Abstract**
Heart disease is a main cause of maternal mortality in the United States and the United Kingdom. Most deaths are from acquired conditions. However, due to the increased survival of children born with congenital heart disease (CHD) over the past 30 years, the population of adults with congenital heart disease in the U.S. now exceeds 1 million. Thus, there are now more adults with CHD than children with CHD. Many of these adult survivors of pediatric heart disease are of childbearing age and are considering pregnancy. This article reviews the literature concerning pregnancy and CHD.

J Am Coll Cardiol. 2012 Jul 17;60(3):224-9.
The immediate and long-term impact of pregnancy on aortic growth rate and mortality in women with marfan syndrome.
Donnelly RT, Pinto NM, Kocolas I, Yetman AT.
**Source**
Division of Cardiology, Department of Pediatrics, Primary Children's Medical Center, University of Utah, Salt Lake City, Utah.
**Abstract**
**Objectives:** The study sought to assess the impact of pregnancy on the rate of aortic growth as well as on short- and long-term clinical outcomes in women with Marfan syndrome.
Background: There is a paucity of data on peripartum and long-term clinical outcomes in women with Marfan syndrome who are followed prospectively during pregnancy.
**Methods:** Echocardiographic, demographic, and surgical data review of all adult females with a confirmed diagnosis of Marfan syndrome was performed.
**Results:** Of the 98 women identified, 69 (72%) experienced a total of 199 pregnancies resulting in 170 (86%) live births. The median number of pregnancies per women was 3 (interquartile range: 1 to 12). Obstetrical complications occurred in 17 (10%) and adverse fetal outcomes in 22 (13%). No woman experienced aortic dissection or required cardiac surgery during pregnancy. Aortic growth rate increased

during pregnancy and did not return to baseline following pregnancy completion. Despite the lack of catastrophic peripartum complications, the prevalence of both aortic dissection and elective aortic surgery during long-term follow-up was higher in those women who had a prior pregnancy. Risk factors for adverse cardiac outcome included greater aortic diameter, greater rate of aortic growth during pregnancy, increased number of pregnancies, lack of beta-blocker use during pregnancy, and lack of prospective pregnancy follow-up.
**Conclusions:** There is a low incidence of aortic complications during pregnancy in women with Marfan syndrome and an aortic diameter <4.5 cm. However, pregnancy does increase the risk of aortic complications in the long-term in this group of patients.

Genes Brain Behav. 2012 Jul 11. doi: 10.1111/j.1601-183X.2012.00821.x. [Epub ahead of print]
Cognitive functioning of adults with Noonan syndrome: a case-control study.
Wingbermühle E, Roelofs RL, van der Burgt I, Souren PM, Verhoeven WM, Kessels RP, Egger JI.
**Source**
Vincent van Gogh Institute for Psychiatry, Venray; Donders Institute for Brain, Cognition and Behaviour, Centre for Cognition; Behavioural Science Institute.
**Abstract**
Noonan syndrome (NS) is a genetic disorder characterised by short stature, facial dysmorphia, congenital heart defects and mildly lowered intellectual abilities. Research has mainly focused on genetic and somatic aspects, while intellectual and cognitive functioning has been documented scarcely. Also, to date studies have been primarily performed in children. This is the first study in which functioning within the major cognitive domains is systematically evaluated in a group of adults with NS and compared with a control group. Extensive neuropsychological assessment, including the domains intelligence, speed of information processing, memory (working memory, immediate recall and delayed recall), executive function and visuoconstruction, was performed in a sample of 42 patients with NS and 42 healthy controls, matched on age, sex and education level. In addition, subjective cognitive complaints were assessed with self-report questionnaires. On the domain speed of information processing patients performed worse than controls ($P<0.05$). Furthermore, except for slightly better results on delayed recall in the patients with NS ($P<0.05$), none of the other cognitive domains showed between-group differences. On the questionnaires, patients reported substantially more complaints about their own cognitive abilities than controls ($P<0.05$). A lowered speed of information processing and relatively intact functioning in other cognitive domains characterises the cognitive profile of adult patients, in contrast to previous findings in children with NS, who seem to have more generalised cognitive deficits.

Genet Med. 2012 Jun 28. doi: 10.1038/gim.2012.66. [Epub ahead of print]
Functional outcomes of adults with 22q11.2 deletion syndrome.
Butcher NJ, Chow EW, Costain G, Karas D, Ho A, Bassett AS.
**Source**
1] Clinical Genetics Research Program, Centre for Addiction and Mental Health, Toronto, Ontario, Canada [2] Institute of Medical Science,

University of Toronto, Toronto, Ontario, Canada.

**Abstract**

**Purpose:** The 22q11.2 deletion syndrome is a common multisystem genomic disorder with congenital and later-onset manifestations, including congenital heart disease, intellectual disability, and psychiatric illness, that may affect long-term functioning. There are limited data on adult functioning in 22q11.2 deletion syndrome.
**Methods:** We used the Vineland Adaptive Behavior Scales to assess functioning in 100 adults with 22q11.2 deletion syndrome (n = 46 male; mean age = 28.8 (standard deviation = 9.7) years) where intellect ranged from average to borderline (n = 57) to mild intellectual disability (n = 43).
**Results:** More than 75% of the subjects scored in the functional deficit range. Although personal, vocational, and financial demographics confirmed widespread functional impairment, daily living skills and employment were relative strengths. Intelligence quotient was a significant predictor ($P < 0.001$) of overall and domain-specific adaptive functioning skills. A diagnosis of schizophrenia was a significant predictor ($P < 0.05$) of overall adaptive functioning, daily living skills, and socialization scores. Notably, congenital heart disease, history of mood/anxiety disorders, sex, and age were not significant predictors of functioning.
**Conclusion:** Despite functional impairment in adulthood that is primarily mediated by cognitive and psychiatric phenotypes, relative strengths in activities of daily living and employment have important implications for services and long-term planning. These results may help to inform expectations about outcomes for patients with 22q11.2 deletion syndrome.Genet Med advance online publication 28 June 2012.

Eur Heart J Cardiovasc Imaging. 2012 Jun 26. [Epub ahead of print]

Certification in echocardiography of congenital heart disease: experience of the first 6 years of a European process.

Mertens L, Miller O, Fox K, Simpson J.

**Source**

Division of Cardiology, The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada.

**Abstract**

**Aims:** Certification in congenital heart disease echocardiography presents unique challenges compared with certification of adult transthoracic and transoesophageal echocardiography. We report our experience in collaboratively developing an exam process that covers the size and age range of congenital heart patients, the varying professional backgrounds of echocardiography practitioners across the field and our approach to the challenge of introducing a pan-European certification endorsed by the major stakeholder groups; the European Association of Echocardiography (EAE), the Association for European Paediatric and Congenital Cardiology (AEPC) and the Grown Up Congenital Heart Working Group of the European Society of Cardiology (ESC).
**Methods and Results:** Since its inception in 2006 the exam has been held seven times; 137 candidates from 27 countries have sat the exam, 107 candidates (78%) have passed the exam components and 60 candidates have successfully completed the logbook submission and have been certified in echocardiography of congenital heart disease echocardiography by the EAE. In addition to the certification process, a comprehensive curriculum, teaching programme, and teaching courses

have been developed.
**Conclusion:** The instititution of a European certification process for echocardiography of congenital heart disease has proved feasible.

Eur J Cardiothorac Surg. 2012 Jun 24. [Epub ahead of print]
Is heart transplantation for complex congenital heart disease a good option? A 25-year single centre experience.
Seddio F, Gorislavets N, Iacovoni A, Cugola D, Fontana A, Galletti L, Terzi A, Ferrazzi P.
**Source**
Paediatric Cardiovascular Surgery Unit, Bergamo Hospital, Bergamo, Italy.
**Abstract**
**Objectives:** Heart transplantation (HTx) in patients with complex congenital heart disease (CHD) is a challenge because of structural anomalies and multiple previous procedures. We analysed our results in adult and paediatric patients to evaluate outcome and assess risk factors affecting mortality.
**Methods:** Between 1985 and 2011, among 839 patients who underwent HTx, 85 received transplantation for end-stage CHD. Patients were divided into four age subgroups: <1 year (8 patients, Group I), 1-10 years (20 patients, Group II), 11-18 years (24 patients, Group III) and >18 years (33 patients, Group IV) and into two time periods: 1985-2000 (47 patients) and 2001-2011 (38 patients). Anatomical diagnoses were single-ventricle defect in 37 patients (44%) and two-ventricle defect in 48 patients (56%). Seventy-three patients (86%) had undergone one or more cardiac surgical procedures prior to HTx (mean 2.4 ± 0.9). Twenty-two of them were suffering from Fontan failure. Mean pulmonary artery pressure was 25.2 ± 14.2 mmHg. Mean transpulmonary gradient was 9.4 ± 6.9 mmHg.
**Results:** Mean follow-up after HTx was 7.8 ± 6.8 years. Survival at 1 month was 37.7% in Group I, 85.8% in Group II, 96.8% in Group II and 98.4% in Group IV and was significantly worse in younger recipients. Overall 30-day mortality was 17.6%. Currently 56 patients (65.8%) are alive. Overall survival at 1, 5, 10 and 15 years is 83-, 73-, 67- and 58%, respectively. There were 14 late deaths. Univariate analysis found that risk factors for early and late death were those related to recipient illness, such as pre-transplant creatinine, intravenous inotropic drugs, intravenous diuretics, mechanical ventilation and presence of protein-losing enteropathy (PLE). Multivariate analysis for all events (early and late deaths) identified preoperative mechanical ventilation as an independent risk factor for mortality. Number of previous procedures did not influence survival. Previous Fontan procedure did not increase mortality. We documented the reversibility of PLE in survivors.
**Conclusions:** We demonstrated that heart transplantation for patients with CHD can be performed with the expectation of excellent results. Previous procedures, including the Fontan operation, do not reduce survival. Mortality is related to preoperative patient condition. We advocate early referral of complex CHD patients for transplant assessment and for inclusion in waiting lists before the detrimental effects of end-stage failure manifest themselves.

Int J Cardiol. 2012 Jun 22. [Epub ahead of print]
Fibrinogen function is impaired in whole blood from patients with cyanotic congenital heart disease.
Jensen AS, Johansson PI, Bochsen L, Idorn L, Sørensen KE, Thilén U,

Nagy E, Furenäs E, Søndergaard L.

**Source**

Department of Cardiology, Rigshospitalet, Copenhagen, Denmark.

**Abstract**

**Background:** Patients with cyanotic congenital heart disease (CCHD) have haemostatic abnormities associated with bleeding and thrombo-embolic events. The haemostatic abnormalities are not fully understood, but recent studies indicate that elevated haematocrit and fibrinogen function may be of importance. The aim of this study was to characterise the haemostatic profile and examine the potential role of haematocrit on clot formation and strength in CCHD patients. Furthermore to examine whether CCHD patients with history of haemoptysis have diminished fibrinogen function compared to those without haemoptysis.

**Methods:** In a prospective study 75 adult CCHD patients had haematocrit, platelet count, and plasma fibrinogen concentration examined. Furthermore thrombelastography(TEG) as well as TEG Functional Fibrinogen(TEG FF) assay evaluating fibrinogen function(FLEV) was performed. Data were compared with historical data regarding previous haemoptysis in CCHD patients.

**Results:** Haematocrit was 57±8% and platelet counts in the lower normal range. TEG revealed a hypocoagulable condition with impaired clot formation. TEG values were correlated to haematocrit, indicating that elevated haematocrit causes impaired clot formation and strength. Despite high levels of plasma fibrinogen, TEG FF demonstrated that FLEV was diminished and negatively correlated to haematocrit. Furthermore CCHD patients with previous history of haemoptysis had significantly lower FLEV compared to CCHD patients without haemoptysis.

**Conclusion:** Patients with CCHD are hypocoagulable mainly due to impaired fibrinogen function. Despite a low platelet count, platelet function does not seem to be severely affected in CCHD patients. Haemostasis, and especially fibrinogen function, is negatively affected by elevated haematocrit, and fibrinogen function is diminished in CCHD patients with haemoptysis.

Congenit Heart Dis. 2012 Jun 13. doi: 10.1111/j.1747-0803.2012.00677.x. [Epub ahead of print]

Reproductive and Contraceptive Counseling Received by Adult Women with Congenital Heart Disease: A Risk-based Analysis.

Hinze A, Kutty S, Sayles H, Sandene EK, Meza J, Kugler JD.

**Source**

Joint Division of Pediatric Cardiology, University of Nebraska Medical Center College of Medicine/Creighton University School of Medicine, Children's Hospital & Medical Center Department of Biostatistics, College of Public Health, University of Nebraska Medical Center College of Medicine, Omaha, Neb, USA.

**Abstract**

**Background/Objective:** Our aim was to study the prevalence of counseling received by adult women with congenital heart disease to determine from whom they received such counseling and to describe their contraceptive and reproductive knowledge.

**Methods/Design:** Using a cross-sectional survey, information was collected from 83 women, ≥19 years of age with congenital heart disease from a group of 404 women followed in our adult congenital heart disease clinic. Women were stratified into combined hormonal contraceptive and pregnancy World Health Organization risk classes 1-4

based on cardiac lesion.
**Results:** We hypothesized that >50% of women had not received both contraceptive and reproductive counseling that addressed their heart condition; indeed, 59% of women reported they had not received such counseling (P=.05). Women who had received heart-specific contraceptive counseling were in higher risk combined hormonal contraceptive World Health Organization classes (P=.02). Similarly, women who reported receiving counseling regarding risks of pregnancy were also in higher pregnancy World Health Organization risk classes (P=.002). Fifty-two of 77 women (63%) did not know if there was a contraindicated contraceptive method given their underlying heart condition; 16 of these 52 women (31%) were combined hormonal contraceptive class 3 or class 4.
**Conclusions:** This adult congenital heart disease survey study demonstrates an opportunity to improve individualized contraceptive and reproductive counseling with a goal toward minimizing each patient's risk of potentially avoidable adverse events. A stronger collaboration among health care professionals is needed to increase the prevalence of heart-specific counseling and to increase the quality of counseling these women are receiving.

Pediatr Cardiol. 2012 Jun 7. [Epub ahead of print]
Renal Dysfunction is Common Among Adults After Palliation for Previous Tetralogy of Fallot.
Buelow MW, Dall A, Bartz PJ, Tweddell JS, Sowinski J, Rudd N, Katzmark L, Earing MG.
**Source**
Department of Pediatrics, Medical College of Wisconsin, 9000 W Wisconsin Avenue, Milwaukee, WI, 53226, USA.
**Abstract**
Long-term survival after tetralogy of Fallot (TOF) repair is excellent. However, little is published regarding late noncardiac complications. This study aimed to determine the prevalence and risk factors for renal dysfunction among adults after TOF repair. For this study, 56 adult patients with complete repair of TOF were identified, and their charts were retrospectively reviewed. An estimated glomerular filtration rate (eGFR) for each patient was calculated using the Modification of Diet in Renal Disease formula (MDRD). Using each patient's eGFR, he or she was classified into stages based on the National Kidney Foundation chronic kidney disease (CKD) staging. Clinical parameters were compared among patients with and those without renal dysfunction to identify risk factors for renal impairment. The median estimated eGFR rate for the cohort was 78 ml/min/1.73 m(2). Based on the National Kidney Foundation CKD staging system, 54 % of the patients had at least stage 2 chronic renal disease. The risk factors identified were hypertension ($p < 0.01$), type 2 diabetes mellitus ($p < 0.05$), longer follow-up evaluation ($p < 0.005$), older age at complete repair ($p < 0.05$), and use of daily diuretics ($p < 0.05$). After repair of TOF, renal dysfunction is common at late follow-up evaluation. The study findings show the importance of routine assessment of renal function and the need to limit or avoid future episodes of acute kidney injury in this at-risk population.

Echocardiography. 2012 Jun 5. doi: 10.1111/j.1540-8175.2012.01750.x. [Epub ahead of print]
Echocardiographic Guidance of Percutaneous Patent Foramen Ovale

Closure: Head-to-Head Comparison of Transesophageal versus Rotational Intracardiac Echocardiography.

Vigna C, Marchese N, Zanchetta M, Chessa M, Inchingolo V, Pacilli MA, Amico C, Fanelli M, Fanelli R, Loperfido F.

**Source**

Department of Cardiology, Casa Sollievo della Sofferenza Hospital IRCCS, San Giovanni Rotondo, Italy Department of Cardiovascular Disease, Ospedale Civile, Cittadella, Italy Department of Pediatric Cardiology and Adult with Congenital Heart Defect, Policlinico San Donato IRCCS, San Donato Milanese, Italy Department of Neurology, Casa Sollievo della Sofferenza Hospital IRCCS, San Giovanni Rotondo, Italy Department of Cardiology, University of Foggia, Foggia, Italy Department of Cardiology, Catholic University Medical School, Rome, Italy.

**Abstract**

**Background:** Transesophageal (TEE) and intracardiac (ICE) echocardiography are commonly used to guide percutaneous patent foramen ovale (PFO) closure. The study aim was to perform a head-to-head comparison between TEE and rotational ICE echocardiography in the measurement of the fossa ovalis and device selection.

**Methods:** In 45 patients with cryptogenic stroke or peripheral embolism and PFO with large right-to-left shunt, fossa ovalis dimensions were assessed preoperatively by TEE and intraoperatively by rotational ICE. The Amplatzer devices, deployed on the basis of ICE, were compared with those that would have been selected by TEE.

**Results:** A good correlation between TEE and rotational ICE was observed for both longitudinal and transverse fossa ovalis dimensions (TEE four-chamber vs. ICE four-chamber: $r = 0.75$; TEE bicaval vs. ICE four-chamber: $r = 0.77$; TEE aorta vs. ICE aorta: $r = 0.59$; $P < 0.001$ for all). However, no such correlation was found in 13 patients with atrial septal aneurysm (ASA) (TEE four-chamber vs. ICE four-chamber: $r = 0.33$; TEE bicaval vs. ICE four-chamber: $r = 0.49$; TEE aorta vs. ICE aorta: $r = 0.05$; $P = NS$ for all). At Bland-Altman analysis, slight systematic differences with wide limits of agreement for each comparison were observed, particularly in patients with ASA, suggesting that the two imaging modalities cannot be used interchangeably. As regards device selection, a moderate agreement was found between TEE- and ICE-guided device size (72%, $\kappa = 0.53$, $P < 0.001$), except in patients with ASA (36%, $\kappa = 0.02$, $P = NS$).

**Conclusions:** Our study suggests a significant disagreement between TEE and rotational ICE in measuring fossa ovalis and selecting the device for PFO closure, particularly in patients with ASA. (Echocardiography, 2012;**:E1-E8).

Heart. 2012 Jul;98(13):1014-9.

Bicuspid aortic valve and associated aortic dilation in the young.

Fernandes S, Khairy P, Graham DA, Colan SD, Galvin TC, Sanders SP, Singh MN, Bhatt A, Lacro RV.

**Source**

Department of Cardiology, Children's Hospital, Boston, MA 02115, USA.

**Abstract**

**Background:** The aorta in patients with bicuspid aortic valve (BAV) is larger and grows more rapidly than in patients with tricommissural aortic valve. Young patients with BAV can have significant aortic dilation that places them at risk for morbidity and mortality.

**Objective:** The aims of this study were to determine the rate of growth

of the aorta in young patients with BAV and to identify predictors of significant dilation and rapid aortic growth.
**Methods:** 333 patients were randomly selected from an inception cohort of 1192 patients with BAV identified between 1986 and 1999.
**Results:** Median age at the most recent study was 13.5 (0-30) years, 74% were male. Moderate/severe (Z>4) aortic root and ascending aortic dilation was present in 14/333 (5%) and 53/333 (16%) of patients, respectively. In longitudinal follow-up, only a minimal change in aortic Z-score was noted. Predictors of moderate/severe aortic root dilation included moderate/severe aortic regurgitation, absence of moderate/severe aortic stenosis and fusion of the right and left coronary leaflets. Predictors of moderate/severe ascending aortic dilation included moderate/severe aortic regurgitation and absence of aortic coarctation.
**Conclusion:** Moderate/severe dilation of the ascending aorta is common in young patients with BAV, but moderate/severe dilation of the aortic root is less common. The Z-scores for both remained relatively constant over time even in patients with significant dilation, implying that young children with moderate/severe aortic dilation may be at the highest risk for dilation-related complications as adults.

Heart Lung Circ. 2012 Aug;21(8):433-8. Epub 2012 May 11.
Does pregnancy contribute to systemic right ventricular dysfunction in adults with an atrial switch operation?
Zentner D, Wheeler M, Grigg L.
**Source**
Department of Cardiology, The Royal Melbourne Hospital, Parkville, Vic 3050, Australia.
**Abstract**
**Background:** To determine whether pregnancy might impact adversely on long-term outcomes in adults post an atrial switch repair on the background of data demonstrating an increased rate of heart failure and death in these adults with systemic right ventricles.
**Methods:** We retrospectively analysed our adult population with an atrial switch repair for transposition of the great arteries to see whether any differences in outcomes (sudden cardiac death, heart failure admissions, use of heart failure medications) existed between women who had and women who had not undergone pregnancy. Controls from the remaining population (transposition of the great arteries and atrial switch operation women) were elected as long as their year of birth fell into the year of birth range seen in the patient group.
**Results:** In women with transposition of the great arteries who have had an atrial switch repair, the long-term occurrence of sudden cardiac death and clinical heart failure (defined as a need for prescription of anti-failure medications or heart failure admissions) appears to be increased.
**Conclusion:** Pregnancy may have an adverse effect on long-term outcomes in women with systemic right ventricles.

Gen Thorac Cardiovasc Surg. 2012 Jun;60(6):341-4. Epub 2012 May 8.
Pulmonary valve replacement long after repair of tetralogy of Fallot.
Shiokawa Y, Sonoda H, Tanoue Y, Nishida T, Nakashima A, Tominaga R.
**Source**
Department of Cardiovascular Surgery, Kyushu University, 3-1-1 Maidashi, Higashiku, Fukuoka 812-8582, Japan. s-yuchi@kyudai.jp
**Abstract**
**Purpose:** Pulmonary valve replacement long after repair of tetralogy of

Fallot can improve cardiac function, functional status, and arrhythmia propensity. This has not been reported in Japan. We aim to evaluate the effects of pulmonary valve replacement in repaired tetralogy of Fallot.
**Methods:** Nineteen patients underwent pulmonary valve replacement after repair of tetralogy of Fallot, excluding Rastelli type operation, between August 1981 and August 2011. The results of the pulmonary valve replacement were assessed by analyzing preoperative and postoperative cardiothoracic ratio, cardiac function, functional status, QRS duration and durability of the prosthetic valves.
**Results:** There were neither operative nor late deaths. The Cardiothoracic ratio significantly improved from 61.0 ± 5.2 % preoperatively to 56.2 ± 4.8 % postoperatively ($P < 0.001$). The New York Heart association functional class significantly improved from 2.4 ± 0.8 preoperatively to 1.2 ± 0.4 postoperatively as well. Left ventricular ejection fraction showed significant improvement. QRS duration showed significant reduction. The freedom redo pulmonary valve replacement at 20 years was 100 %.
**Conclusion:** Pulmonary valve replacement long after repair of previous tetralogy of Fallot had clinical benefits with low mortality. We recommend bioprosthesis for pulmonary valve replacement when adult-sized valve can be accommodated.

Congenit Heart Dis. 2012 Jul;7(4):344-8. doi: 10.1111/j.1747-0803.2012.00658.x. Epub 2012 Apr 27.
Electrophysiology procedures in adults with congenital heart disease.
Ermis P, Franklin W, Kim J, Moodie D, Parekh D.
**Source**
Department of Pediatric Cardiology, Texas Children's Hospital, Baylor College of Medicine, Houston, Tex, USA.
**Abstract**
**Background:** In adult congenital heart disease (CHD), arrhythmias contribute significantly to morbidity and mortality. Often, these adult patients are treated at a freestanding pediatric facility. Limited data exist looking at this cohort.
**Methods:** A retrospective review was performed of all electrophysiology (EP) procedures performed in adults at our institution during a 5-year period from January 1, 2006 through December 31, 2010.
**Results:** There were 99 cases performed in a total of 87 adults with CHD during this time period. The mean patient age was 27.1 years (18-51 years). The most common congenital cardiac diagnoses were: 27% with D-transposition of the great arteries (n=27)-of which 85% (n=23) have had a previous atrial switch procedure, 20% with tetralogy of Fallot (n=20), and 16% with previous Rastelli repair (n=16). Overall, 37 EP studies were performed, with the majority done in patients with complex CHD. There were 74 additional cases. These procedures consisted of: 38 pacemakers (51%), 26 implantable cardiac defibrillators (36%), six laser lead extractions (8%), two loop recorders (3%), and two pocket revisions (3%). During this 5-year period, there was one major complication (1%) and seven minor complications (7%).
**Conclusions:** The complex care of adults with CHD requiring EP procedures can be safely and effectively accomplished in a freestanding pediatric hospital with low complications, provided institutional support of an adult CHD program.

Respirology. 2012 Aug;17(6):957-63. doi: 10.1111/j.1440-1843.2012.02180.x.

Partial anomalous pulmonary venous connection and pulmonary arterial hypertension.

Sahay S, Krasuski RA, Tonelli AR.

**Source**

Department of Medicine, Akron General Medical Center, Akron Adult Congenital Heart Disease Services, Department of Cardiovascular Medicine, Heart and Vascular Institute Department of Pulmonary, Allergy, and Critical Care Medicine, Respiratory Institute, Cleveland Clinic, Cleveland, Ohio, USA.

**Abstract**

**Background and Objective:** Isolated partial anomalous pulmonary venous connection (PAPVC) has been implicated as a cause of pulmonary arterial hypertension (PAH); however this condition is often overlooked in the diagnostic work up of patients with PH. We studied the prevalence of PAH both in patients with isolated PAPVC or associated with other congenital heart diseases (CHD) such as atrial septal defect (ASD). We also aimed to identify factors related to the presence of PAH in these patients.

**Methods:** We retrospectively analyzed data from the Adult CHD database at the Cleveland Clinic, USA between October 2005-2010. We included all patients diagnosed with PAPVC with or without other CHD. We excluded all patients with previous corrective surgeries.

**Results:** We identified 14 (2.5%) patients with PAPVC. Group I included patients with PAPVC (with or without patent foramen ovale (PFO)). Group II included patients with PAPVC associated with other CHD. PAH was seen in six (6/14, 42.8%) patients, two (2/7, 28.5%) in group I and four (4/7, 57.1%) in group II (P=0.3). The mean pulmonary artery pressure in all patients (n=14) was 29.5±13.8 mmHg. Group I had a mean PAP of 23.6±6.6 mmHg as compared to 33.7±16.5 mmHg for group II (P=0.34). The two patients in group I with PAH had either two anomalous pulmonary veins or a condition (sickle cell disease) that could potentially explain the haemodynamic findings.

**Conclusions:** Patients with PAPVC (with or without PFO) in the absence of other CHD had normal pulmonary arterial pressure (PAP) unless they have two pulmonary veins with anomalous return or associated conditions known to cause PAH.

Am J Cardiol. 2012 Jul 1;110(1):109-17. Epub 2012 Mar 29.

Incidence and predictors of sudden cardiac arrest in adults with congenital heart defects repaired before adult life.

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

**Source**

Adult Congenital Heart Disease Unit, La Paz University Hospital, Madrid, Spain.

**Abstract**

Many adult survivors of repaired congenital heart disease (CHD) are at premature risk of death. Sudden cardiac arrest (SCA) is 1 of the leading causes of death but little is known about determinants for SCA in adults with repaired lesions. We sought to determine incidence and risk factors for SCA in a study population of 936 adults with previously repaired CHD who had completed follow-up at a single tertiary center during a mean period of 9 ± 7 years. Mean age at first examination in our institution was 21 ± 7 years. Diagnostic categories included tetralogy of Fallot (216), coarctation of the aorta (157), transposition complexes (99), single ventricle (55), and other CHD (409). During a total follow-up of

8,387 person-years, 22 patients (2.6 per 1,000 person-years) presented with SCA. Incidence of SCA varied widely between specific lesions; the highest incidence was observed in transposition complexes (10 per 1,000 person-years). Independent predictors of SCA were retrospectively identified using multivariate Cox proportional hazard modeling. Age at initial examination and severely impaired subaortic ventricular systolic function were independent risk factors for SCA (severe subaortic ventricular systolic dysfunction, adjusted hazard ratio 29, 95% confidence interval 11 to 72, p <0.001). SCA occurred in 23% of patients with severe subaortic ventricular systolic dysfunction versus 0.7% of patients with nonsevere decreased subaortic ventricular function (p <0.001). In conclusion, severe subaortic ventricular systolic dysfunction is a dominant multivariate predictor of SCA in an unselected population of adult survivors after surgery for CHD. Our data support the consideration of primary prevention strategies in these patients.

Am J Cardiol. 2012 Jun 15;109(12):1797-800. Epub 2012 Mar 28.
Knowledge of and preference for advance care planning by adults with congenital heart disease.
Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach SL, Kovacs AH.

**Source**

Division of Cardiology, University Hospital of Basel, Basel, Switzerland.

**Abstract**

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

Am J Cardiol. 2012 Jun 1;109(11):1657-63. Epub 2012 Mar 23.
Social burden and lifestyle in adults with congenital heart disease.
Zomer AC, Vaartjes I, Uiterwaal CS, van der Velde ET, Sieswerda GJ, Wajon EM, Plomp K, van Bergen PF, Verheugt CL, Krivka E, de Vries CJ, Lok DJ, Grobbee DE, Mulder BJ.

**Source**

Department of Cardiology, Academic Medical Center, Amsterdam, the

Netherlands.

**Abstract**

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

Ann Thorac Surg. 2012 Jul;94(1):124-32. Epub 2012 Mar 17.

Functional health status of adults with tetralogy of fallot: matched comparison with healthy siblings.

Knowles R, Veldtman G, Hickey EJ, Bradley T, Gengsakul A, Webb GD, Williams WG, McCrindle BW.

**Source**

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

**Abstract**

**Background:** Survival prospects for adults with repaired tetralogy of Fallot (TOF) are now excellent. Attention should therefore shift to assessing and improving functional health status and quality of life. We aimed to assess late functional health status of adults surviving TOF repair by matched comparison to their healthy siblings.

**Methods:** All 1,693 TOF repairs performed at our institution between 1946 and 1990 were reviewed. A matched comparison was undertaken whereby presumed survivors and their healthy sibling were contacted and asked to complete the Ontario Health Survey 1990 and the 36-Item Short Form Health Survey (SF-36) questionnaire.

**Results:** Both questionnaires were completed by 224 adult survivors and their sibling closest in age. Adults with repaired TOF had lower scores for self-perceived general health status (p < 0.001), were less likely to rate their health as good or excellent (p < 0.001), and had lower SF-36 scores for physical functioning and general health (p = 0.001) than their siblings. However, patients reported similar satisfaction with their lives, similar levels of social participation and support, and were as likely to be in long-term partnerships. Worse physical and mental health scores were associated with older age at surgery and at time of

questionnaire completion and recent requirement for noncardiac medication.
**Conclusions:** Although reporting lower functional health status then their siblings, quality of life and life satisfaction for adults who underwent surgery for TOF during childhood is comparable to that of their siblings without heart defects. Follow-up of younger adults is required to understand current health outcomes attributable to improvements in the management of TOF.

Pediatr Cardiol. 2012 Jun;33(5):705-12.
Sex and age differences in body-image, self-esteem, and body mass index in adolescents and adults after single-ventricle palliation.
Pike NA, Evangelista LS, Doering LV, Eastwood JA, Lewis AB, Child JS.
**Source**
School of Nursing, University of California, Factor Building Room 3-938, Los Angeles, CA 90095-6919, USA. npike@sonnet.ucla.edu
**Abstract**
Single-ventricle congenital heart disease (SVCHD) requires multiple palliative surgical procedures that leave visible surgical scars and physical deficits, which can alter body-image and self-esteem. This study aimed to compare sex and age differences in body-image, self-esteem, and body mass index (BMI) in adolescents and adults with SVCHD after surgical palliation with those of a healthy control group. Using a comparative, cross-sectional design, 54 adolescent and adult (26 male and 28 female) patients, age 15-50 years, with SVCHD were compared with 66 age-matched healthy controls. Body-image and self-esteem were measured using the Multidimensional Body-Self Relations Questionnaire-Appearance Scale and Rosenberg Self-Esteem Scale. Height and weight were collected from retrospective chart review, and BMI was calculated. Female adolescents and adult patients with SVCHD reported lower body image compared with males patients with SVCHD and healthy controls ($p = 0.003$). Specific areas of concern were face ($p = 0.002$), upper torso or chest ($p = 0.002$), and muscle tone ($p = 0.001$). Patients with SVCHD who were \21 years of age had lower body image compared with healthy controls ($p = 0.006$). Self-esteem was comparable for both patients with SVCHD and healthy peers. There were no sex differences in BMI; BMI was higher in subjects[21 years of age ($p = 0.01$). Despite the similarities observed in self-esteem between the two groups, female patients with SVCHD\21 years of age reported lower perceived body-image. Our findings support the need to recognize poor psychological adjustment related to low self-esteem in patients with SVCHD; female patients warrant increased scrutiny. Strategies to help patients with SVCHD cope with nonmodifiable aspects of body-image during the difficult adolescent-to-young adult years may potentially enhance self-esteem and decrease psychological distress.

QJM. 2012 Jun;105(6):527-35. Epub 2012 Feb 1.
A simple clinical model to estimate the probability of Marfan syndrome.
Sheikhzadeh S, Kusch ML, Rybczynski M, Kade C, Keyser B, Bernhardt AM, Hillebrand M, Mir TS, Fuisting B, Robinson PN, Berger J, Lorenzen V, Schmidtke J, Blankenberg S, von Kodolitsch Y.
**Source**
Department of Cardiology/Angiology, University Hospital Hamburg - Eppendorf, Hamburg, Martinistrasse 52, 20246 Hamburg, Germany.
**Abstract**
**Background:** Marfan syndrome is a heritable connective tissue disease.

Definitive diagnosis is complex, and requires sequencing of a large gene, FBN1.
**Aim:** We aimed to develop a simple model to estimate the pre-test probability of Marfan syndrome.
**Design:** Prospective cross-sectional study.
**Methods:** We applied diagnostic standards for definitive diagnosis or exclusion of Marfan syndrome in 329 consecutive persons. In 208 persons with random assignment to our derivation group, we performed multivariate logistic regression to assess 14 clinical variables for inclusion in a prediction model with derivation of score points from the estimated coefficients. We created cut-offs to classify low, moderate and high probability of Marfan syndrome. For validation, we applied the model to the remaining 121 persons.
**Results:** We identified seven variables for inclusion in the final model, where we assigned four score points to ectopia lentis, two points to a family history of Marfan syndrome, and one point to previous thoracic aortic surgery, to pectus excavatum, to a wrist and thumb sign, to previous pneumothorax, and to skin striae. In the derivation group 12, 42 and 92% of persons with low (≤1 point), moderate (>1-3.5 points) or high pre-test probability (>3.5 points) had Marfan syndrome, compared to 12, 57 and 91%, respectively, in the validation group. Positive likelihood ratios were 13.96 and 8.54 in the high probability group of the derivation and validation group, respectively.
**Conclusion:** A simple prediction model provides evidence for Marfan syndrome. This model can be used to identify patients who require definitive diagnostic work-up.

Eur Heart J. 2012 Jun;33(11):1386-96. Epub 2011 Dec 23.
Reference values for exercise limitations among adults with congenital heart disease. Relation to activities of daily life--single centre experience and review of published data.
Kempny A, Dimopoulos K, Uebing A, Moceri P, Swan L, Gatzoulis MA, Diller GP.
**Source**
Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital, Sydney Street, London, UK. kempny@gmail.com
**Abstract**
**Aims:** We aimed to investigate the distribution of exercise capacity across the spectrum of adult congenital heart disease (ACHD) using own data and the published experience and to provide diagnosis, gender-, and age- specific reference values.
**Methods and Results:** Publications describing exercise capacity in ACHD patients using cardiopulmonary exercise testing (CPET) were identified (n = 2286 patients in 23 papers). In addition, we included 2129 patients who underwent CPET at our own institution. The majority of patients (80%) had reduced peak oxygen uptake (peak VO(2)) compared with normal values (defined as <90% of predicted peak VO(2)). There were significant differences in peak VO(2) between subgroups of patients, with the lowest values seen in patients with Eisenmenger syndrome and complex heart disease. However, even in patients with simple lesions, peak VO(2) was on average significantly reduced compared with normal values. Based on a large number of observations we herewith provide gender- and age-specific peak VO(2) centile plots for the most common lesions (Tetralogy of Fallot, systemic right ventricle, Ebstein anomaly and Fontan-palliation) and relate

disease-specific exercise capacity to that required for specific activities of daily life, sports, and occupations.
**Conclusion:** We provide age-, gender-, and diagnosis-specific data on peak VO(2) levels across the spectrum of ACHD allowing to compare the exercise capacity of individual patients with that of their peer patients. These data should be helpful in interpreting CPET results, guiding therapy, and advising patients on activities of daily life, sports participation, and choice of occupation.

Cardiol Young. 2012 Aug;22(4):417-23. Epub 2011 Dec 14.
Tetralogy of Fallot in men: quality of life, family, education, and employment.
Bygstad E, Pedersen LC, Pedersen TA, Hjortdal VE.
**Source**
1 Department of Cardiothoracic and Vascular Surgery, Aarhus University Hospital, Aarhus, Denmark.
**Abstract**
**Introduction:** Little is known about the quality of life, health, family, education, and employment status among adult men with repaired tetralogy of Fallot.
**Material and methods:** A total of 68 men who underwent repair of tetralogy of Fallot between 1971 and 1991 were studied. Fifty-three patients answered the SF-36 health survey and additional questions regarding offspring, education, and employment status. The men with repaired tetralogy of Fallot were compared with 32 healthy men and 40 women who also underwent repair of tetralogy of Fallot in the same period.
**Results:** The patients scored lower than healthy men in the SF-36 categories physical functioning, general health, and physical component summary. There were no statistically significant differences in the scores from male and female patients except a lower score in bodily pain among women. Educational level for men operated for tetralogy of Fallot was similar to the general male population, whereas fewer were employed and more were retired, undergoing rehabilitation or receiving social benefits. The reproduction rate was lower compared with the general population (0.65 versus 1.02 children per man) but relatively higher than the rate among women with tetralogy of Fallot (0.88 versus 1.84 children per woman). The risk of having a child with congenital heart disease was 8.3%.
**Conclusion:** Men operated for tetralogy of Fallot have good quality of life and educational status. They start a family, although their reproduction rate is two-thirds that of the general population. The risk of having a child with congenital heart disease is higher compared with the background population. The overall quality of life is similar for men and women operated for tetralogy of Fallot.

J Thorac Cardiovasc Surg. 2012 Jun;143(6):1279-85. Epub 2011 Dec 10.
Power loss and right ventricular efficiency in patients after tetralogy of Fallot repair with pulmonary insufficiency: clinical implications.
Fogel MA, Sundareswaran KS, de Zelicourt D, Dasi LP, Pawlowski T, Rome J, Yoganathan AP.
**Source**
Division of Cardiology, Department of Pediatrics, Children's Hospital of Philadelphia, University of Pennsylvania School of Medicine, Philadelphia, PA 19104, USA. fogel@email.chop.edu

**Abstract**
**Objectives:** To quantify right ventricular output power and efficiency and correlate these to ventricular function in patients with repaired tetralogy of Fallot. This might aid in determining the optimal timing for pulmonary valve replacement.
**Methods:** We reviewed the cardiac catheterization and magnetic resonance imaging data of 13 patients with tetralogy of Fallot (age, 22 ± 17 years). Using pressure and flow measurements in the main pulmonary artery, cardiac output and regurgitation fraction, right ventricular (RV) power output, loss, and efficiency were calculated. The RV function was evaluated using cardiac magnetic resonance imaging.
**Results:** The RV systolic power was 1.08 ± 0.62 W, with 20.3% ± 8.6% power loss owing to 41% ± 14% pulmonary regurgitation (efficiency, 79.7% ± 8.6%; 0.84 ± 0.73 W), resulting in a net cardiac output of 4.24 ± 1.82 L/min. Power loss correlated significantly with the indexed RV end-diastolic and end-systolic volume (R = 0.78, P = .002 and R = 0.69, P = .009, respectively). The normalized RV power output had a significant negative correlation with RV end-diastolic and end-systolic volumes (both R = -0.87, P = .002 and R = -0.68, P = .023, respectively). A rapid decrease occurred in the RV power capacity with an increasing RV volume, with the curve flattening out at an indexed RV end-diastolic and end-systolic volume threshold of 139 mL/m(2) and 75 mL/m(2), respectively.
**Conclusions:** Significant power loss is present in patients with repaired tetralogy of Fallot and pulmonary regurgitation. A rapid decrease in efficiency occurs with increasing RV volume, suggesting that pulmonary valve replacement should be done before the critical value of 139 mL/m(2) and 75 mL/m(2) for the RV end-diastolic and end-systolic volume, respectively, to preserve RV function.

Cardiol Young. 2012 Aug;22(4):381-9. Epub 2011 Nov 9.
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.
**Source**
1 Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital, London, United Kingdom.
**Abstract**
**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.

Eur Heart J. 2012 Jun;33(11):1378-85. Epub 2011 Oct 27.
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.
**Source**
Department of Cardiology, Academic Medical Center, Meibergdreef 9, Amsterdam, The Netherlands.
**Abstract**
**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 (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 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.).

Cardiol Young. 2012 Jun;22(3):307-15. Epub 2011 Oct 21.
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.
**Source**

Division of Cardiology, Department of Pediatrics, University of Alberta, Edmonton, Alberta, Canada. andrew.mackie@ualberta.ca
**Abstract**
**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.

Eur J Cardiovasc Nurs. 2012 Jun;11(2):239-47. Epub 2012 Mar 5.
A review of the management of pulmonary arterial hypertension associated with congenital heart disease.
Harries C, Armstrong I.
**Source**
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**Abstract**
Approximately 5-10% of adolescent and adult patients with congenital heart disease (CHD) will develop pulmonary arterial hypertension (PAH). Patients with PAH associated with CHD (PAH-CHD) exhibit several similarities to those with idiopathic and other associated forms of PAH, especially with regards to their non-specific, cardinal symptoms. The development of PAH-CHD can lead to lifelong impairment although, paradoxically, survival may be better versus idiopathic PAH. Patients with PAH-CHD may experience social limitations, and emotional and psychological issues, arising from their disease burden. Nurses and other allied healthcare professionals are well placed to deliver the individually-tailored care that patients with PAH-CHD require. Activities known to be of particular benefit include patient engagement and education, patient empowerment, colleague training, and ensuring effective communication across the multidisciplinary team. Recent developments in the management of PAH-CHD have led to changes in the medical needs and optimal care of this patient population. This review aims to provide an overview of the natural course, diagnosis, symptoms and impact of PAH-CHD. We also aim to communicate the

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| | | current standards in management of patients with PAH-CHD, and how their outlook can be improved in the future. |