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[In This Issue](#)

[Regional News](#)

[Journal Watch](#)

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International Society for
Adult Congenital Heart Disease

ISACHD Newsletter

President's Message

by Curt J Daniels



Dear ISACHD Members,

I hope everyone had a nice summer and was able to spend some time with the family. At ISACHD headquarters, we are in the early stages of planning the ISACHD meeting for AHA 2012 in Los Angeles, California, USA. At the present time, we expect the ISACHD meeting to be scheduled for Sunday evening, 7:00 pm, November 4th, at a site TBD. Our work groups are continuing to make progress.

ISACHD Global Health

A small exploratory team (Mike Landzberg, Boston, MA, USA ISACHD Past-President; Paul Khairy, Montreal, Quebec, CA, ISACHD President-Elect; Disty Pearson, Boston, MA, USA; and yours truly) met this past week with the

leadership of [Project Hope](#) at their home base in Millwood, Virginia, USA.

I encourage you to visit their website and learn more about Project Hope. The purpose of the meeting was to exchange ideas, set priorities, and begin moving toward a partnership between ISACHD and Project Hope with the potential to provide to underserved or under-structured CHD areas of the world in the following areas:



- ACHD Education
- Guidance for ACHD Health Care Delivery
- ACHD Training
- International ACHD Database

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- International ACHD Research

Meeting with Project Hope

We will need volunteers to accomplish our initiatives, so please stay informed.
There will be more to come and discuss at our ISACHD AHA meeting 2012.

ISACHD Global Education

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. This effort will coordinate with our global health agenda as we develop international health partners, i.e., Project Hope. We look forward to an update on their tremendous work and progress at our upcoming ISACHD AHA meeting 2012.

ISACHD Research

Philip Moons (Leuven, Belgium, ISACHD Secretary), Koichiro Niwa (Tokyo, Japan, ISACHD Treasurer and Asian Pacific Regional Representative), and the work group 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. We look forward to Philip presenting his proposal for multi-international research at the ISACHD AHA meeting.

As described above, we are in the early phases of organizing the priorities of the work groups. We will need YOU and many volunteers once we move into implementation of the initiatives. Please stay informed and contact me or any of the work group chairs to discuss your ISACHD thoughts and ideas.

If you haven't renewed your membership for this year, please do so. Now is the time!

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.

Looking forward to seeing you soon,

Curt J. Daniels
President

**Regional News:
News from Europe**

By Helmut Baumgartner

The ESC Annual Congress 2012 took place in Munich, Germany from August 25th to 29th, 2012. This meeting has currently become the largest cardiovascular forum in the world. The WG GUCH of the ESC was very well represented and able to offer an attractive scientific program, which attracted large audiences. For the first time ever, the program included nine pre-arranged oral sessions, touching upon all aspects of ACHD (Arrhythmias, Heart Failure, Imaging, Interventions, Outcomes, Pregnancy, and Surgery). Invited speakers

came from Europe and North America. The number of oral and poster sessions is also increasing. During this meeting Dr. Johan Holm, from Lund, Sweden (a former fellow of Dr. Gary Webb), became chairperson of the WG GUCH ESC for the period 2012-2014.

News from South America

By Luis Alday

Dr. Ariane Marelli from Canada is the special guest for the "ISACHD - Argentine Society of Cardiology Joint Session" that will address "Adult Congenital Heart Disease and Genetic Cardiomyopathies in the Present Century" at the Buenos Aires Sheraton Hotel as a part of the 28th Argentine Congress of Cardiology on Sunday October 7th, 2012.

The following program has been planned:

- 12:00 Demographic changes in congenital heart disease in the XXI century:
Impact in management and outcomes.
Dr. MARELLI, Ariane (CANADA)
- 12:15 Left ventricular noncompaction
Dr. ALDAY, Luis (ARGENTINA)
- 12:30 Learned lessons from the residency and registry for adults with congenital heart disease in the province of Cordoba
Dr. MAISULS, Hector (ARGENTINA)
- 12:45 Quality of care in the adult with congenital heart disease: What does it mean?
Dr. MARELLI, Ariane (CANADA)

WG on Education

By Erwin Oechslin, Toronto (Canada)

The WG on Education is establishing an Internet-based ACHD Learning Centre and developing a basic teaching course in ACHD for adult cardiology trainees. If you have not provided your link yet, and 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:

- Chicago Adult Congenital Heart Network: October 20, 2012. The West Chicago Lakeshore, 644 North Lake Shore Drive, Chicago, Illinois 60611. President CATCH Executive Committee: Joel Hardin, Division Director, Pediatric Cardiology, Loyola University Health System. To register please email to Natalie.pitts@gmail.com and ask for a registration form. There is no registration fee.
- 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.

From the Nursing Network

By Desiree Fleck, PhD, CRNP

From ISACHD APC, formerly ISACHD Nursing Network:

ACHD Toronto: During the 22nd International Symposium on ACHD, there were about 37 nursing/PA members in attendance, attending four breakout sessions. During the breakout sessions, we learned about Congenital heart defects (Marion McCrae), Fontan physiology and approaches (Marion McCrae), "What should I have in my tool box to care for ACHDers" (Qunyu Li, Laura Lee Walker, Steve Crumb), What is Blue in ACHD, Eisenmenger's, and pulmonary hypertension (Disty Pearson and Desiree Fleck). In the main sessions, Philip Moons, Steve Crumb, Disty Pearson, and Qunyu Li also presented on the Top 10 questions for APNs in ACHD, How to Optimize the Role of Nurses/Congenital Cardiac Care Associates in Heart Failure Management, Advanced Care Planning, and Quality Death and Dying. In addition, there was a networking dinner for 23 CCAs in ACHD. Much discussion was shared about Doctoral preparation, networking, patient education, and future networking ideas. Over eighteen programs and five countries were represented!

The ISACHD Nursing Network is now the ISACHD Advanced Practice Clinicians, fondly referred to as ISACHD APC. We are reformatting our group to be more inclusive to other non-physician members. Currently, members of the board include Mary Canobbio, Lori Newman, Michelle Nickolaus, Philip Moons, Disty Pearson, and Desiree Fleck. We are looking for new members to our group to identify two objectives to achieve. If you are interested, please let any of us know (fleckd@email.chop.edu).

Journal Watch

Commentary of "Outcomes of pulmonary valve replacement in 170 patients with chronic pulmonary regurgitation after relief of right ventricular outflow tract obstruction: implications for optimal timing of pulmonary valve replacement" by Pia Schuler.

Background: Severe pulmonary regurgitation (PR) has been recognized to have major impact on right ventricular (RV) function and the outcome of patients repaired tetralogy of Fallot. Beneficial effects of pulmonary valve replacement (PVR) with this regard have, however, to be weighed against the operative risk and repeat interventions. Optimal timing of PVR remains a challenge. Magnetic resonance imaging (MRI) is now accepted to be the gold standard for RV volume measurement and is considered to provide important information for the decision of intervention.¹

Summary of the paper:

Objectives: The main purpose of this study was to evaluate outcomes of PVR in patients with chronic pulmonary regurgitation and to determine the optimal timing of PVR.

Method: A cohort of 170 patients who underwent surgical PVR between 1998 and 2011 for chronic PR was retrospectively analyzed. The underlying heart defects comprised tetralogy of Fallot (139 patients) or similar physiology (31 patients). Pre- and postoperative MRI data was analyzed in 67 patients. Postoperative MRI was carried out at a median of 9.7 months. MRI parameters included indexed biventricular endsystolic (RVESVI, LVESVI) and enddiastolic volumes (RVEDVI, LVEDVI), ejection fraction (RVEF, LVEF) and pulmonary regurgitation (PR RF).

The median follow-up duration was 5.9 years. Outcome measurements

compromised all-cause deaths, pulmonary valve failure (need for redo PVR or interventional catheter procedure), pulmonary valve dysfunction (peak pressure gradient ≥ 40 mmHg or at least moderate PR on echocardiography) and arrhythmias (sustained atrial fibrillation, atrial flutter or ventricular tachycardia). Optimal outcome was defined as normalization of RV volume and function by MRI.

Results: Overall survival at 10 years was $97.5 \pm 1.6\%$. Twelve patients underwent redo PVR, and 2 patients underwent interventional balloon valvuloplasty. Freedom from redo PVR at 10 years was $74.7 \pm 10\%$, and freedom from prosthetic valve failure and dysfunction at 10 years was $50.3 \pm 10.1\%$. There were 2 early deaths (1.2%) due to ischemic brain injury and RV dysfunction. 3 patients developed arrhythmias. QRS duration decreased significantly ($p=0.002$). Overall event-free survival at 10 years was $69 \pm 10.5\%$. In addition, the number of patients with tricuspid regurgitation of at least moderate grade decreased significantly (10% vs 0%, $p<0.001$). Subgroup analysis ($n=67$) in patients with pre- and post-MRI measurements showed a significant reduction in RV volumes, improvement in biventricular function and increase in LVEDVI. RVEDVI was normalized in 65% and RVESVI in 52%. Optimal outcome in term of MRI parameters was achieved in 51%. No patient with optimal MRI outcome had a poor clinical outcome (death, arrhythmia). Cutoff value of pre-operative RVEDVI for normalization of RVEDVI was 168 ml/m^2 (sensitivity 74%, specificity 74%), and cutoff value of pre-operative RVESVI for normalization of RVESVI was 80 ml/m^2 (sensitivity 68%, specificity 68%). Cutoff values for optimal outcome (normalized RV volumes and function) were 163 ml/m^2 for RVEDVI and 80 ml/m^2 for RVESVI. Higher pre-operative RVESVI was identified as a sole independent risk factor for suboptimal outcome.

Comments:

This study confirms the results of Knauth et al² that ventricular size and function assessed by cardiac MRI predict major adverse clinical longterm outcome after tetralogy of Fallot repair. No patient with normalized RV volume and function had a poor clinical outcome, when normalization was achieved by restoration of pulmonary valve function. Lee et al also showed that there is a limit in RV dilatation where PVR does no longer lead to normalization of RV volumes and function.

The MRI RV volume threshold for postoperative RV normalization has already been studied by several groups of investigators. Therrien et al³ suggested $\text{RVEDVI} \leq 170 \text{ ml/m}^2$, Oosterhov et al⁴ $\leq 160 \text{ ml/m}^2$, Buechel et al⁵ $\leq 150 \text{ ml/m}^2$ and Frigiola et al⁶ $<150 \text{ ml/m}^2$ for optimal postinterventional outcome. The indexed pre-operative systolic volume for optimal outcome was $\leq 85 \text{ ml/m}^2$ by Therrien et al³ and $< 90 \text{ ml/m}^2$ by Geva et al⁷.

Explanation for these differing values might be varying measurement techniques. There is little agreement on the most suitable imaging plane for RV volumetric analysis in the setting of abnormal RV physiology. The recently published data by Clarkes et al⁹ revealed a statistically significant difference in interobserver reliability of RV end-systolic volume (ESV) measurements favouring the axial method ($p = 0.047$). In subjects with RV EDV 150 ml/m^2 , RV SV measured using axial contours yielded better agreement with forward flow measured in the pulmonary trunk (CCC = 0.63) than measurements made using short-axis contours (CCC = 0.56; $p = 0.007$).

In addition, differences in volume parameters can be explained by whether pure endocardial RV borders are outlined, or careful contouring around each trabeculation is performed. Fallot patients present with increased trabeculation and therefore, measurements with or without trabeculation will produce different

results.

As diastolic volumes are profoundly influenced by surgical repair (RVOT aneurysms or akinesia by patch plastic), and as the global systolic function decreases late in the pathophysiologic response to severe pulmonary regurgitation⁸, the endsystolic RVESVI may be a more sensitive parameter than the enddiastolic RVEDVI to predict suboptimal outcome as suggested in this paper. However, the reduced sensitivity and specificity for RVESVI (68%) in this study needs to be taken into account.

Overlapping numbers in RVEF and QRS duration in both, the optimal and suboptimal group, are another concern. Geva et al⁷ suggested a pre-operative RVESVI < 90 ml/m² and QRS-duration of < 140 msec as predictors of optimal post-operative result (normalization of MRI-based RV volume and function). The number of patients with QRS measurements in this study was low (n=14, n=20). Conflicting results may also be due to different characteristics of

the study population.

Lee et al report a pulmonary valve failure or dysfunction of 50% after 10 years. This implies, the earlier the valve is replaced the more often the patient has to undergo further valve interventions. With the advent of transcatheter valve implantation, the number of re-operation may however be significantly reduced. This may allow to change strategies in the future and justify early decision for PVR. The fact, that the present study could not find an improvement of arrhythmias and of oxygen consumption by cardiopulmonary stress testing after PVR may support that intervention is required even earlier.

Conclusion:

MRI assessment of the right ventricle plays an important role for the optimal timing for pulmonary valve repair after repair of tetralogy. Additional measures such as symptoms, QRS duration, history of arrhythmia, exercise capacity and progression of disease continue to be important for the pre-operative evaluation. Further studies are needed to refine optimal criteria for the timing of PVR.

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Article: Lee C, Kim YM, Lee CH, Kwak JG, Park CS, Song JY, Shim WS, Choi EY, Lee SY, Baek JS. Outcomes of pulmonary valve replacement in 170 patients with chronic pulmonary regurgitation after relief of right ventricular outflow tract obstruction: implications for optimal timing of pulmonary valve replacement. *J Am Coll Cardiol.* 2012 Sep 11;60(11):1005-14.

Pia K Schuler, M.D.

Adult Congenital & Valvular Heart Disease Center, Cardiology & Angiology, University Hospital

Commentary: Muenster,
Muenster,
Germany

J Am Coll Cardiol. 2012 Sep 11;60(11):1005-14. Epub 2012 Aug 22.

[Outcomes of pulmonary valve replacement in 170 patients with chronic pulmonary regurgitation after relief of right ventricular outflow tract obstruction: implications for optimal timing of pulmonary valve replacement.](#)

Lee C, Kim YM, Lee CH, Kwak JG, Park CS, Song JY, Shim WS, Choi EY, Lee SY, Baek JS.

Source

Department of Thoracic and Cardiovascular Surgery, Cardiovascular Center, Sejong General Hospital, Bucheon, Republic of Korea.

Abstract

Objectives: The objectives of this study were to evaluate outcomes of pulmonary valve replacement (PVR) in patients with chronic pulmonary regurgitation (PR) and to better define the optimal timing of PVR.

Background: Although PVR is effective in reducing right ventricular (RV) volume overload in patients with chronic PR, the optimal timing of PVR is not well defined.

Methods: A total of 170 patients who underwent PVR between January 1998 and March 2011 for chronic PR were retrospectively analyzed. To define the optimal timing of PVR, pre-operative and post-operative cardiac magnetic resonance imaging (MRI) data (n = 67) were analyzed.

Results: The median age at the time of PVR was 16.7 years. Follow-up completeness was 95%, and the median follow-up duration was 5.9 years. Overall and event-free survival at 10 years was 98% and 70%, respectively. Post-operative MRI showed significant reduction in RV volumes and significant improvement in biventricular function. Receiver-operating characteristic curve analysis revealed a cutoff value of 168 ml/m² for non-normalization of RV end-diastolic volume index (EDVI) and 80 ml/m² for RV end-systolic volume index (ESVI). Cutoff values for optimal outcome (normalized RV volumes and function) were 163 ml/m² for RV EDVI and 80 ml/m² for RV ESVI. Higher pre-operative RV ESVI was identified as a sole independent risk factor for suboptimal outcome.

Conclusions: Midterm outcomes of PVR in patients with chronic PR were acceptable. PVR should be considered before RV EDVI exceeds 163 ml/m² or RV ESVI exceeds 80 ml/m², with more attention to RV ESVI.

Circulation. 2012 Sep 18. [Epub ahead of print]

[Sudden Cardiac Death in Adult Congenital Heart Disease.](#)

Koyak Z, Harris L, de Groot JR, Silversides CK, Oechslin EN, Bouma BJ, Budts W, Zwinderman AH, Van Gelder IC, Mulder BJ.

Source

1 Academic Medical Center, Amsterdam & ICIN, Utrecht, the Netherlands;

Abstract

Background: Sudden cardiac death (SCD) is a major cause of mortality in adults with congenital heart disease (CHD). The aim of this study was to determine the adult CHD population at risk of SCD and the clinical parameters associated with SCD.

Methods and results: We performed a multicenter case-controlled study. Patients who died suddenly due to proven or presumed arrhythmia were included (cases). For each case, two controls matched on diagnosis, type of surgical intervention, age and gender were included. From three databases including 25790 adults with CHD, 1189 deaths (5%) were identified, of whom 213 patients (19%) died suddenly. Arrhythmic death occurred in 171 of 1189 patients. The underlying cardiac lesions were mild, moderate and severe CHD in 12%, 33% and 55% of the SCD cases, respectively. Clinical variables associated with SCD were supraventricular tachycardia (SVT, OR 3.5, 95% CI 1.5-7.9, P=0.004), moderate to severe systemic ventricular dysfunction (OR 3.4, 95% CI 1.1-10.4, P=0.034), moderate to severe subpulmonary ventricular dysfunction (OR 3.4, 95% CI 1.1-10.2, P=0.030), increased QRS duration (OR 1.34 (per 10 ms increase), 95% CI 1.10-1.34, P=0.008), and QT

dispersion (OR 1.22 (per 10 ms increase), 95% CI 1.22-1.48, P=0.008).

Conclusions: The clinical parameters found to be associated with SCD in adults with a broad spectrum of CHD, including systemic right ventricles, are similar to those in ischemic heart disease. Moreover, even those patients with mild cardiac lesions, are potentially at risk for SCD. This highlights the need for further prospective studies as well as vigilant ongoing follow-up of the adult with CHD.

Am J Ther. 2012 Sep 12. [Epub ahead of print]

[Acute Care Medicine: Perioperative Management of Adult Congenital Heart Disease.](#)

Howard-Quijano K, Schwarzenberger JC.

Source

Department of Anesthesiology, David Geffen School of Medicine, University of California at Los Angeles, Los Angeles, CA.

Abstract

Since the advent of neonatal cardiac surgery in the 1970s, an increasing number of patients suffering from congenital heart disease (CHD) have survived into adulthood. In 2010, it is estimated that 1.2 million or 1 in 150 of young adults have some form of CHD in the United States. Current birth, incidence, and survival rate predict an increase in the CHD population between 10,000 and 300,000 patients per year. Data from large adult CHD (ACHD) centers (UCLA, Toronto, Mayo Clinic) show that as many as 50% of these patients with complex physiology are 40 years of age or older and that two-thirds of them can be categorized as medium or high risk to demonstrate signs and symptoms of low cardiac output. As this population ages, it is very likely for hospital-based physicians to encounter such patients in their procedural and/or surgical practice. Risk stratification and interdisciplinary approach in the care of these patients will assure a safe outcome. The assessment of the patient must consider the variable expression of CHD. Because most lesions are "fixed but not cured," the periprocedural practitioner must consider the altered physiology of the heart in context of the physiological challenges of percutaneous and open surgical interventions. The 2008 American Heart Association/American College of Cardiology guidelines for the Management of Adults with CHD establish for the first time a streamlined approach to the care of these patients. This review will apply these guidelines to the patients with ACHD undergoing noncardiac surgery and intervention.

Congenit Heart Dis. 2012 Sep 12. doi: 10.1111/j.1747-0803.2012.00717.x. [Epub ahead of print]

[Impact of Central Venous Pressure on Cardiorenal Interactions in Adult Patients with Congenital Heart Disease after Biventricular Repair.](#)

Ohuchi H, Ikado H, Noritake K, Miyazaki A, Yasuda K, Yamada O.

Source

Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, Osaka, Japan.

Abstract

Background: Cardiorenal interactions adversely impact the prognosis in heart failure patients an effect which crucially involves increased central venous pressure (CVP). However, it is unclear whether the same pathophysiology operates in adults with congenital heart disease (CHD).

Purpose: The present study was designed to assess cardiorenal interactions in adults with CHD after biventricular repair.

Methods and results: We measured the kidney length (KL, cm/m) and renal resistive index (RI) in 77 consecutive patients and 30 controls. We also measured hemodynamics, plasma B-type natriuretic peptide level, 24-hour creatinine clearance, and peak oxygen uptake in each patient. The CVP correlated with the KL ($r = 0.44$, $P < .001$) and the RI was greater in the patients ($P < .0001$). The high RI was independently determined by the CVP, aortic pressure, and cardiac index ($P < .05$ -.001), and correlated with the 24-hour creatinine clearance ($r = -0.30$, $P < .05$). The RI correlated closely with the neurohumoral activations and peak oxygen uptake ($|r| = 0.45$ - 0.50 , $P < .0001$), and the patients with a traditional criteria of high RI (≥ 0.70) had a higher incidence of cardiovascular events that required unscheduled hospitalization (hazard ratio = 2.78, 95% confidence interval 1.26-6.10, $P < .05$). Multivariate Cox model with the cutoff values of $KL \geq 68$ cm/m and $RI \geq 0.74$ revealed that a greater KL (hazard ratio = 4.03, 95% confidence interval 1.46-11.1, $P < .01$) as well as B-type natriuretic peptide ($P < .001$) independently predicted the events.

Conclusions: Hemodynamics, especially a high CVP, independently predicted the enlarged kidney and abnormal intrarenal flow dynamics that are closely associated with heart failure severity and cardiovascular events in adults with CHD after biventricular repair.

Congenit Heart Dis. 2012 Sep 12. doi: 10.1111/chd.12001. [Epub ahead of print]

[Long-term Outcome Following Pregnancy in Women with a Systemic Right Ventricle: Is the Deterioration due to Pregnancy or a Consequence of Time?](#)

Bowater SE, Selman TJ, Hudsmith LE, Cliff PF, Thompson PJ, Thorne SA.

Source

Department of Cardiology, Queen Elizabeth Hospital NHS Foundation Trust.

Abstract

Introduction: The right ventricle (RV) supports the systemic circulation in patients who have had an intraatrial repair of transposition of the great arteries or have congenitally corrected transposition. There is concern about the ability of a systemic RV to support the additional volume load of pregnancy, and previous studies have reported deterioration in RV function following pregnancy. However, conditions with a systemic RV are also associated with progressive RV dysfunction over time. To date, no study has examined whether the deterioration associated with pregnancy is due to the physiological changes of pregnancy itself, or is part of the known deterioration that occurs with time in these patients.

Methods: Women who had undergone pregnancy under the care of the Adult Congenital Heart Disease Unit at the Queen Elizabeth Hospital were retrospectively identified and matched to separate male and nulliparous female controls. Functional status (New York Health Association [NYHA]), RV function, and systemic atrioventricular valve regurgitation were recorded for each group at baseline, postpregnancy (or at 1 year for control groups) and at latest follow-up.

Results: Eighteen women had 31 pregnancies (range 1-4) resulting in 32 live births. There were no maternal but one neonatal death. At baseline, there was no significant difference in NYHA class or RV function between pregnancy and control groups. In postpregnancy, there was a significant deterioration in the pregnant group alone for both NYHA class ($P = 0.004$) and RV function ($P = 0.02$). At latest follow-up, there was a significant deterioration in RV function in all three groups. There was still a reduction from baseline in NYHA of women who had undergone pregnancy ($P = 0.014$), which again was not seen in the controls groups.

Conclusion: This study suggests that pregnancy is associated with a premature deterioration in RV function in women with a systemic RV. These women are also more symptomatic, with a greater reduction in functional class compared with patients with a systemic RV who do not undergo pregnancy. This study will allow this cohort of women to be more accurately counseled as to the potential long-term risks of pregnancy.

J Interv Cardiol. 2012 Sep 7. doi: 10.1111/j.1540-8183.2012.00764.x. [Epub ahead of print]

[Use and Performance of Premounted Stents Compared to Nonpremouted Stents in Pediatric and Adult Congenital Cardiac Catheterization.](#)

Meadows J, Teitel D, Moore P.

Source

From the Department of Pediatrics, Division of Pediatric Cardiology, University of California, San Francisco, California.

Abstract

Objectives: We sought to characterize the use, efficacy, and performance characteristics of premounted stents relative to nonpremouted stents when used during congenital cardiac catheterization.

Background: Endovascular stent implantation is an effective means of relieving vascular obstruction in patients with congenital heart disease. However, stent implantation is technically challenging and important complications occur. Premounted stents appear to offer many advantages relative to their nonpremouted counterparts, and it has been suggested that the use of premounted stents is associated with fewer complications. However, translation of these potential benefits into procedural or clinical success has been poorly examined and the data are conflicting.

Methods: All stent placements performed between January 1, 1999 and December 31, 2009 were reviewed. Analysis of technical success, hemodynamic success and complications was performed.

Results: 416 stents were placed over the 10 year period. 158 (38%) were premouted. There was no apparent trend in the frequency of use of premouted stents over the study period. Implanted premouted stents were smaller in diameter than nonpremouted stents 4.9 mm +/- 1.8 versus 13.9 mm +/- 3.7, and the site of stent placement differed significantly. Unadjusted and adjusted analysis of technical success with respect to the precision of stent placement, hemodynamic success, and complications showed no difference between premouted and non-premouted stents.

Conclusions: We found no difference between premouted and nonpremouted stents with respect to procedural and hemodynamic success or complications. Nevertheless, there remain practical advantages to

the use of premounted stents that may justify their expanding role in congenital cardiac catheterization. (J Interv Cardiol 2012;**:1-4).

Am J Cardiol. 2012 Aug 27. [Epub ahead of print]

[Comparison of Outcomes in Adults With Congenitally Corrected Transposition With Situs Inversus Versus Situs Solitus.](#)

Oliver JM, Gallego P, Gonzalez AE, Sanchez-Recalde A, Brett M, Polo L, Gutierrez-Larraya F.

Source

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Abstract

The long-term outcome of patients with congenitally corrected transposition of the great arteries is mainly determined by progressive morphologically tricuspid valve regurgitation, heart block, atrial arrhythmias, and/or systemic ventricular dysfunction. Situs abnormalities have been reported in $\leq 34\%$ of cases, but whether clinical differences exist between a situs inversus and situs solitus arrangement has not yet been studied. The clinical records of 38 adults with congenitally corrected transposition of the great arteries (mean age 40 ± 15 years) followed for a mean period of 7.4 years were reviewed. Of these 38 patients, 8 presented with situs inversus and 30 with situs solitus. No significant differences were found between the 2 groups in age, gender, ventricular septal defect, pulmonary tract stenosis, previous surgical repair, or duration of follow-up. However, none of the patients with situs inversus presented with an Ebstein-like anomaly of the morphologically tricuspid valve and none developed nonoperative-related complete atrioventricular block compared to 15 (50%; $p = 0.013$) and 11 (42%; $p = 0.032$) of the patients with situs solitus, respectively. At follow-up, 2 patients with situs inversus (25%) presented with sustained atrial arrhythmia, severe tricuspid regurgitation, or severe systemic right ventricular systolic dysfunction compared to 22 (73%) of 30 those with situs solitus ($p = 0.034$). No patient with situs inversus presented with cardiac death or severe heart failure compared to 12 (40%) of 30 with situs solitus ($p = 0.038$). In conclusion, Ebstein-like anomaly or spontaneous complete atrioventricular block are rare in patients with congenitally corrected transposition of the great arteries with situs inversus, and late complications are uncommon. The long-term outcome of patients with situs inversus was significantly better than that for patients with situs solitus.

Clin Res Cardiol. 2012 Aug 30. [Epub ahead of print]

[Percutaneous implantation of the Edwards SAPIEN \(TM\) pulmonic valve: initial results in the first 22 patients.](#)

Haas NA, Moysich A, Neudorf U, Mortezaeian H, Abdel-Wahab M, Schneider H, De Wolf D, Petit J, Narayanswami S, Laser KT, Sandica E.

Source

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Abstract

Background: Percutaneous pulmonary valve implantation (PPVI) was introduced in 2000 as an interventional procedure for the treatment of right ventricular outflow tract (RVOT) dysfunction. The new Edwards SAPIENTM pulmonic valve has reached CE certification at the end of 2010 thus offering an attractive alternative with extended sizes (23 and 26 mm) to the conventional Melody[®] valve (sizes 18, 20 and 22 mm).

Patients: Over a 1-year period, PPVI using the Edwards SAPIENTM pulmonic valve was performed in 22 patients using a standardized procedure. Primary diagnosis was tetralogy of Fallot ($n = 11$), pulmonary atresia ($n = 2$), Truncus arteriosus ($n = 3$), TGA/PS-Rastelli ($n = 1$), Ross surgery ($n = 2$), double outlet right ventricle ($n = 2$) and absent pulmonary valve syndrome ($n = 1$). The character of the RVOT for PPVI was transannular patch ($n = 4$), bioprosthesis ($n = 2$), homograft ($n = 5$) and Contegra[®] conduit ($n = 11$). The leading hemodynamic problem consisted of a pulmonary stenosis (PS) ($n = 2$), pulmonary regurgitation (PR) ($n = 11$) and a combined PS/PR lesion ($n = 9$).

Results: In 21/22 patients, PPVI was performed successfully (10×23 and 11×26 mm). There were 9 female and 13 male patients; the mean age was 21.7 years (range 6-83 years), the mean length was 162 cm (range 111-181 cm) and the weight 56.5 kg (range 20-91 kg). Invasive data showed a decrease of RV-systolic pressure from 61.2 mmHg (± 23.1) to 41.2 mmHg (± 8.6) and reduction of RV-PA gradient from 37.3 mmHg (± 23.2) to 6.9 mmHg (± 5.3). The PA-systolic pressure increased from 25.8 mmHg (± 8.6) to 33.9 mmHg (± 9.3) as did the PA diastolic pressure (from 6.0 mmHg (± 5.6) to 14.6 mmHg (± 4.3)). There was a substantial reduction of pulmonary regurgitation from before (none/trivial $n = 0$, mild $n = 2$, moderate $n = 9$, severe $n = 11$)

to after PPVI (none/trivial n = 20, mild n = 1). During the short-term follow-up of 5.7 months there was no change in the immediate results.

Conclusion: PPVI using the Edwards SAPIEN™ pulmonic valve can be performed safely in a wide range of patients with various diagnoses and underlying pathology of the RVOT and enables the restoration of an adult-size RVOT diameter. Although the immediate and short-term results seem promising, the long-term effects and safety have to be assessed in further clinical follow-up studies.

Catheter Cardiovasc Interv. 2012 Aug 25. doi: 10.1002/ccd.24635. [Epub ahead of print]

[Incidence of patent foramen ovale and migraine headache in adults with congenital heart disease with no known cardiac shunts.](#)

Volman M, Mojadidi MK, Gevorgyan R, Kaing A, Agrawal H, Tobis J.

Source

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Abstract

The purpose of this study was to understand why patients with adult congenital heart disease (CHD) but no obvious shunt have an increased frequency of migraine headaches (MH). CHD patients with No Known Cardiac Shunts (CHD-NKS), based on their echocardiographic or angiographic procedures, were tested for a right-to-left shunt using agitated saline contrast transcranial Doppler (TCD). Medical records of 2920 patients from the UCLA Adult CHD Center were screened to participate in a study to evaluate the prevalence of MH in adults with CHD. 182 patients (6.23%) had CHD-NKS ; of these, 60 (30%) underwent a TCD. 23 (38%) tested positive and 37 (62%) tested negative for a right-to-left shunt ($p=0.01$ compared to controls). The frequency of MH was 43% in CHD-NKS compared to 11% in controls ($p<0.0001$). TCD demonstrated right-to-left shunting in approximately 2/3 of patients with pulmonary stenosis, the Marfan syndrome and congenitally corrected transposition of great vessels, 1/4 of patients with bicuspid aortic valve, 1/5 of patients with mitral valve prolapse and all patients with Ebstein's anomaly. Approximately half of these experienced MH. Patients who had MH did not show a higher frequency of right-to-left shunt when compared to patients without MH [$p=0.57$]. In conclusion, CHD patients with conditions usually not associated with a shunt have a higher than expected prevalence of PFO which permits intermittent right-to-left shunting undetected by standard non-contrast TTE and TEE; the increased prevalence of right-to-left shunting may partially explain the higher than expected frequency of migraines.

J Thorac Cardiovasc Surg. 2012 Aug 23. [Epub ahead of print]

[Anticoagulation early after mechanical valve replacement: Improved management with patient self-testing.](#)

Thompson JL, Burkhart HM, Daly RC, Dearani JA, Joyce LD, Suri RM, Schaff HV.

Source

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Abstract

Objective: Self-testing to determine the international normalized ratio improves management with warfarin and reduces the risks of adverse events. Self-testing usually begins several weeks after hospital dismissal after valve replacement. We aimed to compare the in-hospital initiation of international normalized ratio self-testing with usual care in mechanical heart valve recipients.

Methods: A total of 200 adult mechanical heart valve recipients were randomly assigned to in-hospital international normalized ratio self-testing instruction or usual care. Instruction for self-testing patients began on the fourth postoperative day. The patients were followed up for 3 months to compare the number of international normalized ratio tests, percentage of time in the therapeutic range, and adverse events.

Results: The baseline characteristics were similar between the 2 groups. During the first 3 postoperative months, the usual-care group underwent an average of 10 international normalized ratio tests, and the self-testing group completed 14 international normalized ratio tests. The mean \pm SD percentage of international normalized ratio tests within the therapeutic range was 45% \pm 22% for the usual-care group and 52% \pm 22% for the self-testing group ($P = .05$). Within 90 days after dismissal, transient ischemic attack occurred in 1 patient in the usual-care group and 2 patients in the self-testing group. Bleeding complications occurred in 3 patients in the usual-care group and 5 patients in the self-testing group.

Conclusions: Management of anticoagulation with warfarin after mechanical valve replacement is improved with self-testing, even during the early postoperative phase when international normalized ratio testing is performed frequently. Although the incidence of adverse events was similar in the 2 groups, better control of

the international normalized ratio would be expected to improve outcome in large populations of patients.

Congenit Heart Dis. 2012 Aug 16. doi: 10.1111/j.1747-0803.2012.00699.x. [Epub ahead of print]

[Evaluation of Silent Thrombus after the Fontan Operation.](#)

Grewal J, Al Hussein M, Feldstein J, Kiess M, Ellis J, Human D, Leipsic J.

Source

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Abstract

Background: Thromboembolic complications have been noted after the Fontan operation. However, the prevalence of silent events among an adult contemporary population is not known. Noninvasive screening by any method including computed tomography (CT) has been technically limited to date.

Objectives: The objective of this study was to evaluate a novel dual-energy CT (DECT) protocol in determining the prevalence of "silent" intracardiac thrombus and thrombus in the Fontan and pulmonary circulations among adults after the Fontan operation.

Methods: All post-Fontan patients attending the Pacific Adult Congenital Heart Clinic were approached for study participation. Those agreeable underwent a full clinical assessment, cardiopulmonary stress testing, transthoracic echocardiogram, and DECT low kilovoltage imaging protocol.

Results: Twenty-three patients were included in the study (30 ± 10 years, 26% women). Three (13%) patients had evidence of silent thrombi detected on DECT. All three of these patients had an extracardiac conduit and mural thrombus was found within the conduit. Older age at the time of the Fontan operation was associated with the presence of thrombus (21 ± 14 vs. 11 ± 6 years, $P = .05$).

Conclusions: Thirteen percent of adult patients post-Fontan procedure have clinically silent thrombi. These were all found among patients with an extracardiac conduit traditionally thought to be at low risk for thromboembolism. Given the significant risk of thromboembolic complications, large randomized prospective studies looking at anticoagulation therapy in all Fontan patients are urgently needed. In the meanwhile, given the important rate of silent thrombi, a systematic robust screening protocol that includes noninvasive low radiation methods such as DECT methods should be considered.

Pacing Clin Electrophysiol. 2012 Aug 16. doi: 10.1111/j.1540-8159.2012.03494.x. [Epub ahead of print]

[Importance of Tachycardia Cycle Length for Differentiating Typical Atrial Flutter from Scar-Related in Adult Congenital Heart Disease.](#)

Uhm JS, Mun HS, Wi J, Shim J, Hwang HJ, Sung JH, Kim JY, Pak HN, Lee MH, Joung B.

Source

Division of Cardiology, Department of Internal Medicine, Yonsei University College of Medicine, Seoul, South Korea Division of Cardiology, Department of Internal Medicine, Bundang CHA Medical Center, CHA University, Seongnam, South Korea.

Abstract

Background: Radiofrequency catheter ablation (RFCA) for intraatrial reentrant tachycardia (IART) in congenital heart disease (CHD) remains difficult. Methods: Thirty-four consecutive adult patients (age, 37.6 ± 12.8 years; male, 21) with previously repaired CHD and IART underwent an electrophysiological study and RFCA. CHD included atrial septal defect (ASD, $n = 14$), tetralogy of Fallot ($n = 11$), ventricular septal defect ($n = 4$), pulmonary atresia ($n = 2$), atrioventricular septal defect ($n = 1$), transposition of the great arteries ($n = 1$), and double-outlet right ventricle ($n = 1$).

Results: Duration of CHD repair to IART onset was 19.1 ± 8.5 years. Thirty and four patients had single- and double-loop reentrant tachycardia, respectively. Among the total of 38 IARTs, which were mapped, 22 (57.9%) and 13 (34.2%) IARTs were cavotricuspid isthmus (CTI)-dependent atrial flutter (AFL) and scar-related AFL, respectively. Typical AFL electrocardiography findings including definite sawtooth appearance in inferior leads and positive F wave in lead V1 were observed in only 12 of 21 patients (57.1%) with CTI-dependent AFL. CTI-dependent AFL had a significantly longer tachycardia cycle length (TCL) than scar-related AFL (267.6 ± 34.4 ms and 235.9 ± 37.0 ms, respectively; $P = 0.031$). $TCL > 250$ ms had 79% sensitivity as the cutoff value for differentiating CTI-dependent from scar-related AFL. The acute success rates of RFCA in CTI-dependent and scar-related AFLs were 85.7% and 90.0%, respectively. The recurrence rates in CTI-dependent and scar-related AFLs were 11.1% and 11.1%, respectively, during a follow-up of 21.2 ± 28.3 months.

Conclusions: CTI-dependent AFL was the most common IART in adult patients with repaired CHD and was easily manageable by RFCA. TCL might help to differentiate CTI-dependent AFL from other IARTs. (PACE 2012;XX:1-10).

Pacing Clin Electrophysiol. 2012 Aug 16. doi: 10.1111/j.1540-8159.2012.03489.x. [Epub ahead of print]

[A Review on Advanced Atrioventricular Block in Young or Middle-Aged Adults.](#)

Barra SN, Providência R, Paiva L, Nascimento J, Marques AL.

Source

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Abstract

Complete atrioventricular block is a relatively uncommon arrhythmia that is nonetheless increasingly seen in elderly people of developed countries, due to the increase in life expectancy. Congenital and degenerative etiologies are the most commonly seen among young and old patients, respectively. However, scientific literature is surprisingly scarce regarding the etiology of complete atrioventricular block in the asymptomatic otherwise healthy young and middle-aged adult population. Coronary artery disease, autoimmune disorders such as systemic lupus erythematosus or rheumatoid arthritis, history of acute or chronic infectious or hypersensitivity myocarditis, infiltrative processes, hypothyroidism, congenital cardiopathies such as left ventricular noncompaction or Ebstein anomaly, lamin A/C mutations, and pathologic hypervagotony and idiopathic degenerative scleroatrophy of the atrioventricular junctional specialized tissue (Lenegre-Lev disease) are among the most frequent etiologies of complete atrioventricular block in young or middle-aged adults. To our knowledge, no comprehensive review on the specificities of the investigation warranted in this age group has ever been developed, nor have the implications of particular diagnoses on treatment modalities been appropriately addressed. We aim at reviewing the most frequent differential diagnoses of advanced atrioventricular block in otherwise healthy asymptomatic or mildly symptomatic young or middle-aged adults and their impact on therapeutic options. Additionally, we suggest a diagnostic algorithm that may be helpful in this group of patients. (PACE 2012; 00:1-11).

J Cardiol. 2012 Sep;60(3):151-9. doi: 10.1016/j.jjcc.2012.06.014. Epub 2012 Aug 11.

[Catheter intervention for adult patients with congenital heart disease.](#)

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Source

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Abstract

Adult congenital heart disease is one of the most important clinical issues not only for pediatric cardiologists but also adult cardiologists. After the introduction of catheter intervention for atrial septal defect in the pediatric population, therapeutic advantages of this less invasive procedure now focused on even geriatric patients. The most valuable clinical benefit of this procedure is the significant improvement in symptoms and daily activities, which result from the closure of left to right shunt without thoracotomy or cardiopulmonary bypass surgery. Although currently available therapeutic options for device closure for congenital heart disease in Japan are limited to atrial septal defect, patent ductus arteriosus, or some vascular abnormalities such as coronary arteriovenous fistula, various new techniques or devices such as ventricular septal defect device, pulmonary valve implantation, are going to be introduced in the near future. To perform safely and achieve good procedure success, real time imaging plays an important role in interventional procedures. Real time three-dimensional transesophageal echocardiography can provide high quality imaging for anatomical evaluation including defect size, surrounding rim morphology, and the relationship between device and septal rim. In adult patients, optimal management of comorbidities is an important issue, including cardiac function, arrhythmias, pulmonary function, and renal function. In particular, atrial arrhythmias are key issues for long-term outcome. Because the interventional procedures are not complication-free techniques, the establishment of a surgical back-up system is essential for achieving a safe procedure. Finally, the establishment of a team approach including pediatric and adult cardiologists, cardiac surgeons, and anesthesiologists is the most important factor for a good therapeutic outcome. Their roles include pre-interventional hemodynamic evaluation, good imaging technique for anatomical evaluation, management of comorbidities, and surgical back up.

Ann Thorac Surg. 2012 Sep;94(3):751-8. Epub 2012 Jun 16.

[Open, hybrid, and endovascular treatment for aortic coarctation and postrepair aneurysm in adolescents and adults.](#)

Roselli EE, Qureshi A, Idrees J, Lima B, Greenberg RK, Svensson LG, Pettersson G.

Source

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Abstract

Background: Open, hybrid, and endovascular procedures are used for grown-up patients with aortic coarctation and complications after repair, an expanding population. We sought to characterize patients and procedures, assess early and late outcomes, and describe indications to guide treatment of these complex patients.

Methods: Between May 1999 and January 2011, 110 patients underwent open (n = 40), hybrid (n = 11), or endovascular (n = 59) repair of coarctation (n = 43), recurrent aortic coarctation (n = 42), or postrepair aneurysm (n = 25). Mean age was 38 ± 14 years. Sixty-eight had previous repairs (median 27 years earlier; range, 1 to 50). Twenty-two had prior cardiovascular operations other than coarctation and 50% had bicuspid valve. Fifty-nine concomitant procedures were performed in 45 patients (40%). Data were from the prospective database, chart review, and Social Security Death Index.

Results: Technical success was achieved in 100%, with no hospital deaths, no strokes, and no paraplegia. Complications were uncommon and included respiratory failure (n = 2, 1.8%), and temporary renal failure (n = 2, 1.8%). Twenty-two patients required reinterventions, but half of those were planned. There was no difference in occurrence of unplanned reintervention between approaches (endovascular 12%, hybrid 18%, open 12.5%). Length of stay was 4.8 ± 4.8 days. Transcoarct gradient fell from 37.6 ± 18 mm Hg preoperatively to 7.0 ± 6.9 mm Hg in coarctation patients. Postrepair aneurysm patients had no late ruptures, and maximum diameter shrunk from 5.9 ± 1.3 cm preoperatively to 4.8 ± 1.3 cm. Estimated survival at 1, 5, and 8 years was 95%, 95%, and 90%, respectively.

Conclusions: Coarctation, recurrent coarctation, and postrepair aneurysm/pseudoaneurysm in adolescent and adult patients can be safely and effectively managed with open, hybrid, or endovascular techniques. Optimal results are achievable in this complex population of patients with a multimodality approach tailored to surgical indication and anatomy. All survivors of coarctation repair require lifelong surveillance.

Echocardiography. 2012 Sep;29(8):E197-200. doi: 10.1111/j.1540-8175.2012.01721.x. Epub 2012 May 29.

[Not all obstructive cardiac lesions are created equal: double-chamber right ventricle in pregnancy.](#)

Murthy S, Lui G, Raiszadeh F, Boxt L, Taub C.

Source

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Abstract

Double-chambered right ventricle (DCRV) is a rare form of right ventricular outflow tract (RVOT) obstruction accounting for approximately 1% of patients with congenital heart disease. It consists of an anomalous muscle bundle that divides the right ventricle usually between the sinus (inlet) and the infundibulum (outlet). This division creates a proximal chamber with high pressure and a distal chamber with low pressure. The hemodynamic obstruction of the RVOT is usually an acquired phenomenon, however the substrate for the anomalous muscle bundle is likely congenital. The diagnosis of DCRV should be considered in the young patient with an elevated right ventricular systolic pressure and intracavitary gradient. Echocardiography and cardiac MRI are the principal diagnostic tools for the assessment of DCRV. This entity is often misdiagnosed as pulmonary hypertension in the young patient, and can often go overlooked and untreated for many years. Definitive therapy involves surgical resection of the muscle bundle. This can often be curative and if done in a timely fashion, may prevent right ventricular remodeling. We describe the unique diagnostic dilemma, the course and management of a young adult with DCRV during pregnancy.

Clin Res Cardiol. 2012 Sep;101(9):691-700. Epub 2012 Mar 28.

[Interventional closure of atrial septal defects without fluoroscopy in adult and pediatric patients.](#)

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Source

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Abstract

Background: Interventional closure of atrial septal defects (ASDs) with a transcatheter device is the preferred strategy in children and adults. This procedure has been proven in numerous studies, but X-ray and contrast agent exposure is still a major side effect. The aim of this study was to clarify whether the interventional closure of ASDs is possible and safe if it is guided by transesophageal echocardiography (TEE) alone.

Methods and results: We retrospectively selected and studied pediatric and adult patients with interventional closure of ASDs at the Deutsches Herzzentrum Berlin (DHZB) without fluoroscopy between 1999 and 2010. We included 330 out of 1,605 patients; 254 had an ASD II, 30 a PFO and 46 multiperforated atrial septum. Median age was 8.92 (0.96-76.3) years and median body weight 32.6 (8.3-156) kg. Median stretched defect size was 13 (5-29) mm. Median procedure time was 50 (20-170) min. Closure was performed in the majority of patients with the Amplatzer(®) septal occluder or Amplatzer(®) PFO occluder. The procedure succeeded in 98.2 % of cases and closure rate was 94.9 % after 48 h. Complication rate was low and procedure time was similar to that necessary with studies using fluoroscopy.

Conclusion: Interventional closure of ASDs is safe and effective if guided with TEE alone. The results can compete with those with the use of fluoroscopy. TEE-guided closure of ASD should be considered in more catheter laboratories to avoid unnecessary radiation exposure for the patient and the examiner.

Eur J Cardiovasc Nurs. 2012 Sep;11(3):349-55. Epub 2012 Mar 13.

[Patients with a congenital heart defect and Type D personality feel functionally more impaired, report a poorer health status and quality of life, but use less healthcare.](#)

Schoormans D, Mulder BJ, van Melle JP, Pieper EG, van Dijk AP, Sieswerda GJ, Hulsbergen-Zwarts MS, Plokker TH, Brunninkhuis LG, Vliegen HW, Sprangers MA.

Source

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Abstract

Background: Type D personality, characterized by high levels of negative affectivity and social inhibition, is related to mortality, morbidity, poor health status, quality of life (QoL) and less healthcare utilization in various cardiovascular patient groups. To date, studies in patients with congenital heart disease (CHD) are lacking. **Aims:** (1) To examine the prevalence of Type D personality in CHD patients; (2) to compare Type D to non-Type D patients with regard to disease severity, functional status, health status and QoL; and (3) to examine the extent to which Type D personality is independently related to healthcare utilization. **Methods:** A total of 1109 adult CHD patients were included in a questionnaire survey. Due to missing data, 302 patients were excluded.

Results: The prevalence of Type D personality was 20.4%. Type D patients reported a poorer functional status, health status and QoL than non-Type D patients ($p < 0.05$). Type D patients reported less healthcare use than non-Type D patients (primary and cardiac outpatient healthcare: adjusted OR=0.56, 95% CI=0.35-0.90; inpatient healthcare: adjusted OR=0.38, 95% CI=0.17-0.83). Results of a post-hoc analysis showed a high prevalence of Type D personality in patients with a poor functional status who did not consult their cardiologist.

Conclusion: Type D patients report a poorer functional status, health status and QoL, but less healthcare utilization. In clinical practice, patients should be screened for Type D personality, since social inhibition may prevent them from contacting a healthcare provider in the event of symptom aggravation.

Hepatology. 2012 Sep;56(3):1160-9. doi: 10.1002/hep.25692. Epub 2012 Aug 2.

[Congenital heart disease and the liver.](#)

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

Source

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Abstract

There are approximately 1 million adult patients with congenital heart disease (CHD) in the United States, and the number is increasing. Hepatic complications are common and may occur secondary to persistent chronic passive venous congestion or decreased cardiac output resulting from the underlying cardiac disease or as a result of palliative cardiac surgery; transfusion or drug-related hepatitis may also occur. The unique physiology of Fontan circulation is particularly prone to the development of hepatic complications and is, in part, related to the duration of the Fontan procedure. Liver biochemical test abnormalities may be related to cardiac failure, resulting from intrinsic liver disease, secondary to palliative interventions, or drug related. Complications of portal hypertension and, rarely, hepatocellular carcinoma (HCC) may also occur. Abnormalities such as hypervascular nodules are often observed; in the presence of cirrhosis, surveillance for HCC is necessary. Judicious perioperative support is required when cardiac surgery is performed in patients with advanced hepatic disease. Traditional models for liver disease staging may not fully capture the severity of disease in patients with CHD. The effectiveness or safety of isolated liver transplantation in patients with significant CHD is limited in adults; combined heart-liver transplantation may be required in those with decompensated liver

disease or HCC, but experience is limited in the presence of significant CHD. The long-term sequelae of many reparative cardiac surgical procedures are not yet fully realized; understanding the unique and diverse hepatic associations and the role for early cardiac transplantation in this population is critical. Because this population continues to grow and age, consideration should be given to developing consensus guidelines for a multidisciplinary approach to optimize management of this vulnerable population. (HEPATOLOGY 2012;56:1160-1169).

Int J Cardiol. 2012 Sep 6;159(3):211-6. Epub 2011 Mar 22.

[Neurohormonal activity and vascular properties late after aortic coarctation repair.](#)

Moutafi AC, Alissafi T, Chamakou A, Chryssanthopoulos S, Thanopoulos V, Dellos C, Xanthou G, Tousoulis D, Stefanadis C, Gatzoulis MA, Davos CH.

Source

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Abstract

Background: Coarctation of aorta (CoA) patients present cardiovascular complications late after repair the causes of which are not fully understood. Our study investigates the neurohormonal and immune activation and the elastic properties of the aorta and peripheral vessels in adult patients with coarctation of aorta (CoA), late after repair.

Methods: Nineteen adult patients with repaired CoA and 29 matched healthy controls underwent aortic distensibility, stiffness index, a study of the elastic properties of peripheral vessels proximal to the coarctation site and measurement of plasma cytokine and neurohormone levels.

Results: Distensibility index was reduced ($p=0.02$) and stiffness index was increased ($p=0.005$) in CoA patients compared to control. Augmentation index ($p=0.0007$) and augmented pressure ($p=0.001$) were higher in CoA patients and Forearm Blood Flow (FBF) index was reduced ($p=0.009$). Plasma levels of sICAM-1 ($p=0.01$), sVCAM-1 ($p=0.05$), E-selectin ($p=0.01$), sFas-ligand ($p=0.02$) and IL-10 ($p=0.01$) were also elevated in CoA patients vs control. TNF- α , IL-6, Endothelin-1 and NT-pro-BNP levels were not.

Conclusions: Adults with repaired CoA seem to develop a late inflammatory reaction, which reflects a functional problem in all vessels, regardless of the initial lesion. This may explain the late complications of the disease despite early repair and improved surgical procedures.