

June, 2011

**Its Time to Renew
Your Membership!**



International Society for
Adult Congenital Heart Disease

*Electronic Invoices will be
coming soon*

ISACHD Newsletter

[Click here](#) to renew your
membership now!

In This Issue

[Nursing News](#)

[Journal Watch
Commentary](#)

[Journal Watch](#)

President

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

President-Elect

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

Secretary

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

Treasurer

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

Past President

Michael J. Landzberg, M.D.
Children's Hospital
Boston, MA, USA

Representative, USA

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

Representative, South America

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

President's Message- Cincinnati: The place to be from June 19-22!!

by *Barbara J.M. Mulder*

Dear ISACHD members,



Please join the International symposium on ACHD in
Cincinnati from June 19-22!

It is the 21st annual course in this comprehensive,
state-of-the-art series of programs on Congenital
Heart Disease in the Adult. A distinguished faculty of
pediatric and adult cardiologists, surgeons, and

other specialists with expertise in medical treatment, diagnosis, nursing,
cardiac imaging, and operative/postoperative care of adults with congenital
heart disease has been assembled to provide a comprehensive four-day
program. The live program will be enhanced by the planned use of
audience response systems and the availability of time for questions and
discussion.



There will be an all-day **ACHD Echo Symposium** on **Sunday, June 19**,
organized by Dr. Tom Kimball of Cincinnati Children's. On Sunday, June
19, there will also be an all-day **ACHD Nursing Symposium**.

The **main program** will be held **Monday, June 20-Wednesday, June 22**
inclusive. There will be two main meeting rooms running concurrently
during the main program. One room will feature case-based teaching. This
will be led by major ACHD teams from North America and Europe. The
second room will feature an academic and clinical agenda, including
lectures, panel discussions, research competitions, and discussions of
controversial issues.

North American ACHD Course Planning Committee:
Craig Broberg, MD, Oregon Health Sciences University

Representative, Europe

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

Representative, Asia-Pacific

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

Representative, Canada

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

Representative, IACHD Nursing Network

Desiree Fleck, PhD, CRNP
Children's Hospital of
Philadelphia
Philadelphia, USA

Journal Watch Editor

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

Executive Director

Peter Kralka
FPMR
Raleigh, NC, USA

Past Presidents

Gary Webb, MD
1994-1996

Carol Warnes, MD
1996-1998

Richard Liberthson, MD
1998-2000

Daniel Murphy, Jr., MD
2000-2002

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

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

Jack M. Colman, MD
2006-2008

Michael J. Landzberg, MD

Erwin Oechslin, MD, University of Toronto
David Sahn, MD, Oregon Health Sciences University
Gary Webb, MD, University of Cincinnati

Barbara J.M. Mulder
President

Nursing News

By: Desiree Fleck

Nursing Symposium for Cincinnati International ACHD Symposium, June 19.

The Nursing network has established the research priorities for ACHD nursing.

Several presentations on Adult Congenital Heart Disease were presented at the Spring meeting of the Cardiovascular Council of nursing and allied health professionals in Belgium this past spring.

ACHD Nursing Education Day will be presented at the Children's Hospital of Philadelphia.

Journal Watch

Commentary on, "Usefulness of Exercise-Induced Hypertension as Predictor of Chronic Hypertension in Adults After Operative Therapy for Aortic Isthmic Coarctation in Childhood" by Robert M. Radke



Robert M. Radke

The prevalence of residual arterial hypertension in adult patients with aortic coarctation remains high despite repair in early life and this is associated with an increase in cardiovascular morbidity and mortality (1). As many as 50% of patients aged between 30 and 40 years remain hypertensive or require antihypertensive medication (2). Re-Coarctation provides a substrate for hypertension in some patients and should be promptly identified. But arterial hypertension often occurs even in patients without significant stenosis. Thus, blood pressure monitoring and management is essential in clinical practice.

The value of exercise testing in patients after coarctation repair still is unclear (3). In the past the measurement of maximal exercise systolic blood pressure has been proposed both as a possible tool to identify patients with re-coarctation (4) as well as patients at increased risk of developing chronic hypertension in the future (5). While the former has

become more straightforward due to the increased availability of MRI, the latter still remains challenging today. Exercise induced hypertension in these patients has been shown to be associated with the mean daytime systolic blood pressure (4) and increased common carotid intima-media thickness (6). But others have found no difference in maximal exercise systolic blood pressure between patients and controls and have questioned the value of this test (7). The prognostic value for the development of arterial hypertension in the future in adult patients after coarctation repair has not been studied in a longitudinal fashion.

In a prospective follow up study with 74 adult patients after successful coarctation repair and without re-coarctation Luijendijk et al aimed to evaluate the predictive value of exercise induced hypertension for chronic hypertension. They conducted ambulatory blood pressure measurements, exercise testing as well as standardized echocardiography at baseline and 6.3 +/- 0.8 years later. At baseline the prevalence of chronic hypertension and exercise hypertension was 36% and 15% respectively. At follow up 55% showed hypertension and 22% exercise induced hypertension. 64% of the patients with exercise induced hypertension at baseline developed chronic hypertension at follow up. This was true for only 19% of patients without any hypertension at baseline. On multivariate analysis, baseline maximum exercise systolic BP was independently associated with the

mean systolic BP at follow-up ($b=0.13$, $p=0.005$). Calculations of sensitivity and specificity of exercise hypertension for the prediction of chronic hypertension after 6 years are not shown in the paper but can be calculated as sensitivity of only 50% and specificity of 87,9%.

The current study finds a strong correlation between maximal exercise systolic blood pressure and 24h ambulatory blood pressure at follow up. This suggests a possible value of this test for risk stratification and could have implications in clinical practice: Patients at higher risk for hypertension might benefit from more frequent blood pressure monitoring, closer follow up and early modification of other risk factors. However, the value of this parameter for clinical practice remains uncertain. Further studies are needed to analyze the long-term consequences and to thoroughly evaluate the predictive value of this parameter and the possible benefit of early treatment.

1. Toro-Salazar OH, Steinberger J, Thomas W, Rocchini AP, Carpenter B, Moller JH. Long-term follow-up of patients after coarctation of the aorta repair. *Am J Cardiol.* 2002 Mar. 1;89(5):541-547.
2. Hager A, Kanz S, Kaemmerer H, Schreiber C, Hess J. Coarctation Long-term Assessment (COALA): significance of arterial hypertension in a cohort of 404 patients up to 27 years after surgical repair of isolated coarctation of the aorta, even in the absence of restenosis and prosthetic material. *J Thorac Cardiovasc Surg.* 2007 Sep.;134(3):738-745.
3. Baumgartner H, Bonhoeffer P, de Groot NMS, de Haan F, Deanfield JE, Galie N, Gatzoulis MA, Gohlke-Baerwolf C, Kaemmerer H, Kilner P, Meijboom F, Mulder BJM, Oechslin E, Oliver JM, Serraf A, Szatmari A, Thaulow E, Vouhe PR, Walma E, Vahanian A, Auricchio A, Bax J, Ceconi C, Dean V, Filippatos G, Funck-Brentano C, Hobbs R, Kearney P, McDonagh T, Popescu BA, Reiner Z, Sechtem U, Sirnes PA, Tendera M, Vardas P, Widimsky P, McDonagh T, Swan L, Andreotti F, Beghetti M, Borggrefe M, Bozio A, Brecker S, Budts W, Hess J, Hirsch R, Jondeau G, Kokkonen J, Kozelji M, Kucukoglu S, Laan M, Lionis C, Metreveli I, Moons P, Pieper PG, Pillosoff V, Popelova J, Price S, Roos-Hesselink J, Uva MS, Tornos P, Trindade PT, Ukkonen H, Walker H, Webb GD, Westby J. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010): The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC). *Eur Heart J.* 2010 Jul. 27;
4. Markel H, Rocchini AP, Beekman RH, Martin J, Palmisano J, Moorehead C, Rosenthal A. Exercise-induced hypertension after repair of coarctation of the aorta: arm versus leg exercise. *JAC.* 1986 Jul.;8(1):165-171.
5. Vriend JWJ, Mulder BJM. Late complications in patients after repair of aortic coarctation: implications for management. *Int J Cardiol.* 2005 Jun. 8;101(3):399-406.
6. Vriend JWJ, de Groot E, Bouma BJ, Hrudova J, Kastelein JJP, Tijssen JGP, Mulder BJM. Carotid intima-media thickness in post-coarctectomy patients with exercise induced hypertension. *Heart.* 2005 Jul.;91(7):962-963.
7. Swan L, Goyal S, Hsia C, Hechter S, Webb G, Gatzoulis MA. Exercise systolic blood pressures are of questionable value in the assessment of the adult with a previous coarctation repair. *Heart.* 2003 Feb.;89(2):189-192.

Article: Paul Luijendijk, Berto J Bouma, Joris W J Vriend, Hubert W Vliegen, Maarten Groenink, and Barbara J M Mulder. *Usefulness of Exercise-Induced Hypertension as Predictor of Chronic Hypertension in Adults After Operative Therapy for Aortic Isthmic Coarctation in Childhood.* *Am J Cardiol.* 2011 May 6. [Epub ahead of print]

Commentary: Robert M. Radke
Adult congenital and valvular heart disease center (EMAH-

Zentrum)
University Hospital of Muenster
Department of Cardiology and Angiology
Albert-Schweitzer-Str. 33
48149 Muenster
Germany

Am J Cardiol. 2011 May 6. [Epub ahead of print]

[Usefulness of Exercise-Induced Hypertension as a Predictor of Chronic Hypertension in Adults After Operative Therapy for Aortic Isthmic Coarctation in Childhood.](#)

[Luijendijk P, Bouma BJ, Vriend JW, Vliegen HW, Groenink M, Mulder BJ.](#)

Source

Department of Cardiology, Academic Medical Centre, Amsterdam, The Netherlands; Interuniversity Cardiology Institute of the Netherlands, Utrecht, The Netherlands.

Abstract

Chronic hypertension is a major concern in adults who have undergone resection of coarctation of the aorta (CoA) in childhood. In otherwise healthy subjects, exercise-induced hypertension is prognostic for chronic hypertension; however, the prognostic value in patients with CoA remains unknown. The aim of the present study was to evaluate the predictive value of exercise-induced hypertension for chronic hypertension in these patients. In the present prospective follow-up study, 74 patients with CoA (58% men, age 30.9 ± 9.5 years) underwent ambulatory blood pressure (BP) monitoring and exercise testing twice from 2001 to 2009 with a follow-up period of 6.3 ± 0.8 years. Hypertension was defined as a mean systolic BP ≥ 140 mm Hg and/or mean diastolic BP ≥ 90 mm Hg or the need for antihypertensive treatment. Exercise-induced hypertension was defined as a mean systolic BP of < 140 mm Hg and peak exercise systolic BP of ≥ 200 mm Hg. At baseline, 27 patients (36%) were hypertensive, 11 (15%) had exercise-induced hypertension, and 36 (49%) were normotensive. At follow-up, all 27 hypertensive patients remained hypertensive. Of the 11 with exercise-induced hypertension, 7 (64%) had developed chronic hypertension, and 4 (36%) continued to have exercise-induced hypertension. Of the 36 normotensive patients, 7 (19%) had developed hypertension, 12 (33%) had developed exercise-induced hypertension, and 17 (47%) remained normotensive. On multivariate analysis, baseline maximum exercise systolic BP was independently associated with the mean systolic BP at follow-up ($\beta = 0.13$, $p = 0.005$). In conclusion, the maximum exercise systolic BP was a predictor for chronic hypertension in patients with CoA. These findings demonstrate the clinical importance of exercise-induced hypertension and warrant additional study into the long-term consequences of exercise-induced hypertension and the potential beneficial role of early antihypertensive treatment in adult patients after CoA repair with exercise-induced hypertension.

Cardiol Young. 2011 May 27:1-5. [Epub ahead of print]

[Fontan conversion with concomitant arrhythmia surgery for the failing atriopulmonary connections: mid-term results from a single centre.](#)

[Sridhar A, Giamberti A, Foresti S, Cappato R, Rubio-Iglesias Garcia C, Cabrera ND, Micheletti A, Negura D, Bussadori C, Butera G, Frigiola A, Carminati M, Chessa M.](#)

Source

Department of Pediatric Cardiology & Adult with Congenital Heart Disease, IRCCS-Policlinico San Donato, Cardiovascular Center 'E. Malan', University of Milan, San Donato Milanese, Milan, Italy.

Abstract

OBJECTIVES:

Classical Atriopulmonary Fontan connections tend to fail in the long term due to progressive anastomotic site obstruction, right atrial enlargement, and refractory atrial arrhythmias. Conversion to total cavopulmonary connection with concomitant arrhythmia surgery is a promising treatment but optimal timing of the procedure remains controversial.

METHODS:

Between the years 2002 and 2009, 15 patients with a median age of 26.2 (12-43) years underwent Fontan conversion operation with concomitant arrhythmia surgery. All were symptomatic and 14 out of the 15 patients had refractory arrhythmias. The duration of pre-operative arrhythmia and the outcome of surgery were correlated to study the impact of delay in surgical intervention on post-operative survival and arrhythmia control.

RESULTS:

There were two patients who died in the early post-operative period (13.3%). At the mid-term follow-up, 53 (20-86) months, late atrial arrhythmias had recurred in two of the 13 surviving patients (15.30%) and one patient developed late sinus node dysfunction. The need for anti-arrhythmic drugs decreased considerably from 93.5% to 15.3% on mid-term follow-up. There was no late death or need for cardiac transplantation. The duration of arrhythmia before surgery was prolonged for more than 10 years in patients who died as well as in those who had complications like late recurrence of arrhythmias, dependence on anti-arrhythmic medications, and worsening of ventricular dysfunction.

CONCLUSIONS:

Fontan conversion is a well-established treatment option for salvaging the failing atriopulmonary connections. Concomitant arrhythmia surgery effectively resolves the refractory atrial arrhythmias and improves survival, but we need to optimise the timing of Fontan conversion to improve the long-term outcome.

Echocardiography. 2011 May 25. doi: 10.1111/j.1540-8175.2011.01434.x. [Epub ahead of print]

[Right and Left Ventricular Strain and Strain Rate in Young Adults before and after Percutaneous Atrial Septal Defect Closure.](#)

[Bussadori C, Oliveira P, Arcidiacono C, Saracino A, Nicolosi E, Negura D, Piazza L, Micheletti A, Chessa M, Butera G, Dua JS, Carminati M.](#)

Source

Pediatric Cardiology and Adult with Congenital Heart Disease Department Center for Diagnosis and Treatment of Valvular Diseases, IRCCS San Donato Hospital, Milan, Italy
Manchester Heart Centre, Central Manchester University Hospitals, NHS Foundation Trust, Manchester, UK.

Abstract

To evaluate acute change of right and left ventricle after percutaneous closure of isolated atrial septal defect (ASD) 21 adult patients (13 F; 8 M) aged 28 ± 9.5 range 18-49 years have been examined by echocardiography before and 24 hours after percutaneous closure of ASD. Twenty-one normal adult subjects, as control group were included. A MyLab25 echo machine equipped with a multifrequency 2.5-3.5 MHz transducer was used. Offline computer-based analysis for strain and SR were performed using XStrain software based on a feature tracking algorithm. All patients had ASD OS2 with right ventricular dilatation and diastolic areas were larger than in controls: $P = 0.0158$. Global right ventricular longitudinal strain was higher $P = 0.0438$. Twenty-four hours after ASD closure, right ventricular diastolic and systolic areas were significantly reduced. Right ventricular global longitudinal systolic strain decreased: $P = 0.00016$, as well as global right ventricular longitudinal SR $-1.56/\text{sec} \pm 0.57$ vs. $-1.28/\text{sec} \pm 0.31$, $P = 0.02646$. At the mean time left ventricular end diastolic volume and left ventricular cardiac output

measured by two-dimensional echocardiography both increased significantly $P = 0.002145$ and 0.013409 . Global circumferential strain at mitral level augmented significantly $-20.3\% \pm 4.64$ vs. $-25.39\% \pm 5.22$, $P = 0.00003$. Longitudinal strain of the right ventricle works as indicator of right ventricular function dependent on loading conditions while SR seems to be less dependent on it. Circumferential strain could be used as an indicator of left ventricular response to normalized loading conditions. (Echocardiography ****; **:1-8).

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

[Clinical outcomes of adult survivors of pulmonary atresia with intact ventricular septum.](#)

John AS, Warnes CA.

Source

Division of Cardiovascular Diseases, Internal Medicine, and Pediatric Cardiology, Mayo Clinic, Rochester, MN, United States; Division of Cardiology, Children's National Medical Center, George Washington University, Washington, DC, United States.

Abstract

BACKGROUND:

There are no studies on the long term clinical outcomes and complications in the adult patient with pulmonary atresia with intact ventricular septum (PA/IVS). This study reviews our experience with a limited group of adult survivors of PA/IVS seen in our adult congenital clinics.

METHODS:

Twenty adult patients with PA/IVS (1998 to 2009) were identified from Mayo Clinic adult congenital heart disease databases. Surgical history and clinical outcomes were reviewed.

RESULTS:

Mean age at last evaluation was 29 years (19-39years). There were five deaths within the study period (1998-2009). Median age at death was 32years (30-37years). Seven patients underwent the Fontan operation, eight patients had a biventricular repair, and five patients remained with palliative shunts. All patients required re-interventions in adulthood. Tricuspid valve (TV) (n=5), pulmonary valve (PV)/conduit (n=6), and mitral valve (n=2) replacements were the most frequent re-intervention in the biventricular repair subset. Atrial arrhythmias were present in 80% of the total cohort, the highest rate among Fontan repairs (n=7) and biventricular repairs (n=7). Ventricular arrhythmias occurred in 15% of the cohort.

CONCLUSIONS:

Although limited in number, the adult PA/IVS patients in this series continue to have high rates of morbidity and mortality, with arrhythmias and need for re-operations as the major causes. Patients with biventricular repairs had the highest re-intervention rate in adulthood. While this subset of patients might not be representative of all adult PA/IVS survivors, continued follow-up at centers with expertise in adult congenital cardiology is recommended for all patients.

Curr Cardiol Rep. 2011 May 11. [Epub ahead of print]

[Preventing Sudden Death in the Adult with Congenital Heart Disease.](#)

Tanel RE.

Source

Division of Pediatric Cardiology, UCSF Benioff Children's Hospital, 521 Parnassus Avenue, Box 0632, San Francisco, CA, 94143, USA, ronn.tanel@ucsf.edu.

Abstract

Adults with repaired congenital heart disease represent a complex and heterogeneous group of patients that are increasingly surviving beyond childhood. Patients have a variety of diagnoses that include specific structural anomalies, assorted physiologic derangements, and unique techniques for surgical repair. During long-term follow-up, even the most excellent surgical outcome may result in anatomic stenosis and insufficiency, cardiac rhythm disturbance, and myocardial dysfunction. Any of these abnormalities, alone or in combination, may result in significant morbidity and mortality. Sudden death is commonly due to a cardiac etiology and arrhythmias are frequently suspected to be the cause. Unfortunately, arrhythmias are difficult to predict and may be potentially lethal at their initial presentation. In addition, a wide spectrum of arrhythmias, both supraventricular and ventricular, are possible, depending on the specific diagnosis and type of repair performed. This review will focus primarily on arrhythmias as a cause of sudden cardiac death in this patient population. Particularly important considerations regarding arrhythmias in adults with congenital heart disease include the fact that supraventricular arrhythmias may be as lethal as ventricular arrhythmias, arrhythmia substrates develop in a unique manner when compared with other adult cohorts, and the electrophysiologic status of patients with repaired congenital heart disease must be considered together with the hemodynamic result.

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

[Patients With Single-Ventricle Physiology: Prognostic Implications of Stress Testing.](#)

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

Source

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

Abstract

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

Am J Cardiol. 2011 May 3. [Epub ahead of print]

[Outcomes of Hospitalization in Adults in the United States With Atrial Septal Defect, Ventricular Septal Defect, and Atrioventricular Septal Defect.](#)

Rodriguez FH 3rd, Moodie DS, Parekh DR, Franklin WJ, Morales DL, Zafar F, Graves DE, Friedman RA, Rossano JW.

Abstract

Atrial septal defect, ventricular septal defect (VSD), and atrioventricular septal defect (AVSD) are among the most common congenital heart lesions, with most children surviving to adulthood. However, the clinical course of these patients is largely unknown, particularly pertaining to inpatient care. The purpose of this study was to assess hospitalizations for septal defects in adults with congenital heart disease (CHD) and risk factors associated with significant morbidity and mortality. The 2007 Nationwide Inpatient Sample was used to assess national prevalence of hospitalizations in adults with CHD with septal defects. Co-morbidities and risk factors for mortality were also determined. There were 84,308 adult CHD admissions in the United States in 2007. Fifty-four percent of adult CHD admissions had diagnoses of septal defects, with 48% having atrial septal defect, 7% having VSD, and 0.4% having AVSD. Overall in-hospital mortality was 2.1%. Common co-morbidities included arrhythmias (31%), heart failure (20%), and diabetes

mellitus (18%). On multivariable analysis, independent risk factors for mortality included presence of VSD (odds ratio 3.1, 95% confidence interval [CI] 1.5 to 6.5), trisomy 21 (odds ratio 2.9, 95% CI 1.1 to 7.5), and pulmonary hypertension (odds ratio 1.5, 95% CI 1.0 to 2.4). In conclusion, this study of hospitalizations in adults with septal defects found that admissions are common and associated with significant co-morbidities. Overall mortality is low but is increased in patients with VSD. Cardiac and noncardiac co-morbidities are commonly encountered. Many noncardiac conditions, including trisomy 21 and the youngest and oldest groups, are associated with an increased risk of death.

Congenit Heart Dis. 2011 May;6(3):241-6. doi: 10.1111/j.1747-0803.2011.00522.x. Epub 2011 May 5.

[The impact obesity on early postoperative outcomes in adults with congenital heart disease.](#)

Zaidi AN, Bauer JA, Michalsky MP, Olshove V, Boettner B, Phillips A, Cook SC.

Source

The Adolescent and Adult Congenital Heart Disease Program, The Heart Center, Nationwide Children's Hospital, Columbus, Ohio, USA The Ross Heart Hospital, Ohio State University, Columbus, Ohio, USA The Research Institute, Nationwide Children's Hospital, Columbus, Ohio, USA Surgical Director, Healthy Center for Weight and Nutrition, Nationwide Children's Hospital, Columbus, Ohio, USA Department of Cardiothoracic Surgery, Nationwide Children's Hospital, Columbus, Ohio, USA.

Abstract

Background. As the prevalence of obesity continues to increase, it now includes the growing number of patients with congenital heart disease (CHD). This particular obese patient population may pose additional intraoperative as well as postoperative challenges that may contribute to poor outcomes. Our aims were to determine the influence of obesity on morbidity and mortality in adults with CHD undergoing surgical repair at a free standing children's hospital.

Methods. A retrospective analysis of adult (≥ 18 years) CHD surgery cases from 2002 to 2008 was performed. Congenital heart lesions were defined as mild, moderate, or complex. Patients were categorized by body mass index (BMI): underweight (BMI $< 20\text{kg/m}^2$), normal (BMI $20\text{--}24.9\text{kg/m}^2$), overweight (BMI $25\text{--}29.9\text{kg/m}^2$), and obese (BMI $\geq 30\text{kg/m}^2$). Demographics, incidence of mortality, or specific morbidities were statistically compared using Fisher's exact test and analyses of variance (anovas).

Results. In this population ($n = 165$), overweight (29%) and obese (22%) patients were prevalent. Hypertension (HTN) and pre-HTN were more prevalent in obese and overweight patients. Postoperative renal dysfunction was observed in obese patients with complex CHD ($P = .04$). Mortality was not different among groups.

Conclusions. Obesity is becoming increasingly common among adults with CHD. Despite marginal evidence of postoperative renal complications in obese patients with CHD of severe complexity, the overall presence of obesity did not influence mortality or short term postoperative morbidities.

Congenit Heart Dis. 2011 May;6(3):219-227. doi: 10.1111/j.1747-0803.2011.00521.x. Epub 2011 May 5.

[The Perspective of Patients with Congenital Heart Disease: Does Health Care Meet Their Needs?](#)

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

Source

Department of Medical Psychology, Academic Medical Center, Amsterdam, The Netherlands Department of Cardiology, Academic Medical Center, Amsterdam, The Netherlands Interuniversity Cardiology Institute of the Netherlands, Utrecht, The Netherlands Department of Cardiology, University Medical Center Groningen, Groningen, The Netherlands Department of Cardiology, Radboud University Nijmegen Medical Center, Nijmegen, The Netherlands Department of Cardiology, University Medical Center Utrecht, Utrecht, The Netherlands Department of Cardiology, TweeSteden Hospital, Tilburg and Waalwijk, The Netherlands Department of Cardiology, Sint Antonius Hospital, Nieuwegein, The Netherlands Department of Cardiology, Orbis Medical Center, Sittard, The Netherlands Leiden University Medical Center, Leiden, The Netherlands.

Abstract

Objective. A first step in the delivery of tailored care is answering the following question: does health care meet the needs of patients? Therefore patients' perspective on health care use and their needs was examined. The design used was cross-sectional questionnaire study.

Patients. A total of 1109 adult congenital heart defect (CHD) patients attending one of eight Dutch hospitals were randomly selected from a national database (10% of all registered patients).

Main Outcome Measures. Patient reported questionnaires on in- and outpatient health care use during the past year and need for additional care.

Results. A total of 66% and 40% of patients had contact with their cardiologist and general practitioner, respectively. Six to 10 percent were hospitalized, operated upon, or visited the emergency room. For the majority, the amount of contact was sufficient. Most patients indicated that the communication skills and expertise of the cardiologist and general practitioner were sufficient, and health care improvements were not necessary. Frequent health care users had a poor functional status and frequent contact with their cardiologist and general practitioner. Patients who want more contact with their cardiologist rated the communication skills of the cardiologist as insufficient.

Conclusions. For most patients, the amount and quality of care are both sufficient. Patients who rate the communication skills of the cardiologist as insufficient have need more contact. In addition to the recommended training program as described in the American College of Cardiology/American Heart Association (ACC/AHA) and European Society of Cardiology (ESC) guidelines, we recommend the incorporation of communication training. This is the first study to provide insight into health care use and needs of CHD patients in countries with a compulsory health insurance system from the patient perspective.

Curr Cardiol Rep. 2011 May 3. [Epub ahead of print]

[Genetic Counseling in the Adult with Congenital Heart Disease: What is the Role?](#)

Burchill L, Greenway S, Silversides CK, Mital S.

Source

Department of Medicine, Division of Cardiology, Toronto General Hospital, Toronto, ON, Canada.

Abstract

New discoveries using high-resolution methods for detecting genetic aberrations indicate that the genetic contribution to congenital heart disease has been significantly underestimated in the past. DNA diagnostics have become more accessible and genetic test results are increasingly being used to guide clinical management. Adult congenital heart disease specialists seeking to counsel adults with congenital heart disease about the genetic aspects of their condition face the challenge of keeping abreast of new genetic techniques and discoveries. The emphasis of this review is on the genetic basis of

structural cardiovascular defects. A framework for identifying adult congenital heart disease patients most likely to benefit from genetic testing is suggested, along with a summary of current techniques for genetic testing. The clinical and ethical challenges associated with genetic counseling are highlighted. Finally, emerging technologies and future directions in genetics and adult congenital heart disease are discussed.

Am J Cardiol. 2011 Apr 27. [Epub ahead of print]

[Prevalence and Determinants of Incomplete Right Atrial Reverse Remodeling After Device Closure of Atrial Septal Defects.](#)

[Fang F, Yu CM, Sanderson JE, Luo XX, Jiang X, Yip GW, Lam YY.](#)

Source

Division of Cardiology, Department of Medicine and Therapeutics, Prince of Wales Hospital, Chinese University of Hong Kong, Hong Kong, China; Translational Medicine Research and Development Center, Institute of Biomedical and Health Engineering, Shenzhen Institutes of Advanced Technology, Chinese Academy of Sciences, Shenzhen, China; Ultrasound Department, Beijing Anzhen Hospital, Capital Medical University, Beijing, China.

Abstract

Whether the relief of chronic right atrial (RA) volume load by device closure of an atrial septal defect (ASD) normalizes RA size is unknown. The present study evaluated the prevalence and determinants of incomplete RA reverse remodeling (RAR) after ASD closure in adults. Transthoracic echocardiography was performed in 44 consecutive patients with secundum ASD (age 43 ± 17 years, 10 men) without a history of atrial arrhythmia shortly before and at 3 months after device closure of ASD. The pulmonary/systemic flow ratio was derived using invasive oximetry. The RA size had significantly decreased at 3 months of follow-up (RA volume index [RAVI] 52 ± 29 to 27 ± 17 ml/m², $p < 0.001$). Incomplete RAR (defined as a RAVI of ≥ 21 ml/m²) was detected in 25 patients (57%) after closure. They were older, had a larger pulmonary/systemic flow ratio, a higher pulmonary arterial systolic pressure, more tricuspid regurgitation, and larger RA, left atrial, and right ventricular sizes before closure than those with a normalized right atrium. Before closure, RAVI was the only independent determinant for incomplete RAR (odds ratio 1.115, 95% confidence interval 1.019 to 1.220; $p = 0.018$). A cutoff value of RAVI of ≥ 40 ml/m² has a sensitivity of 84% and specificity of 72% in the receiver operating characteristic curve. The preclosure RAVI correlated moderately with the shunt-duration index, calculated by multiplying the age to pulmonary/systemic flow ratio ($r = 0.64$, $p < 0.01$). In conclusion, incomplete RAR occurred in $>1/2$ of the adult patients at 3 months after ASD device closure and was related to excessive preclosure RA dilation.

J Card Surg. 2011 May;26(3):254-8. doi: 10.1111/j.1540-8191.2011.01249.x. Epub 2011 Apr 11.

[Surgical rescue of embolized amplatzer devices.](#)

[Amanullah MM, Siddiqui MT, Khan MZ, Atiq MA.](#)

Source

Cardiothoracic Surgery and Pediatric Cardiology, Aga Khan University Hospital, Karachi, Pakistan University of Karachi, Karachi, Pakistan.

Abstract

Background and Aim: Transcatheter closure of atrial septal defect (ASD) and patent ductus arteriosus (PDA) with Amplatzer septal/duct occluder (ASO/ADO) is an established, safe, and efficient procedure with high success. However, device embolization remains a major complication requiring immediate intervention (either

percutaneous or surgical) for retrieval and correction of the heart defect. The aim of this study is to share the experience of managing embolized ASO/ADO. Methods: Of the 284 cases of device closure performed from October 2002 to December 2010, four patients (1.4%) had device embolization requiring immediate surgical retrieval. Two adult female patients with secundum ASD had ASO device implanted. One embolized to the right ventricle and the other into the ascending aorta. An eight-month-old boy and a four-year-old girl with hypertensive PDA had device closure. Device embolization occurred into the descending aorta and right pulmonary artery, respectively. Results: All four devices were retrieved and the defects closed successfully with a low morbidity and no mortality. Conclusion: Careful consideration should be given to surgical or transcatheter closure of a heart defect. Life-threatening complications although rare can occur. Our experience strongly suggests that these devices should only be inserted in facilities where cardiac surgical support is immediately available. (J Card Surg 2011;26-254-258)

Congenit Heart Dis. 2011 May;6(3):190-201. doi: 10.1111/j.1747-0803.2011.00504.x. Epub 2011 Mar 28.

[Liver disease in the patient with fontan circulation.](#)

Wu FM, Ukomadu C, Odze RD, Valente AM, Mayer JE Jr, Earing MG.

Source

Boston Adult Congenital Heart Service, Department of Cardiology, Children's Hospital Boston, Boston, Mass Division of Cardiology Division of Gastroenterology, Department of Medicine Department of Pathology, Brigham and Women's Hospital, Boston, Mass Department of Cardiovascular Surgery, Children's Hospital Boston, Boston, Mass Medical College of Wisconsin, Milwaukee, Wis, USA.

Abstract

The Fontan procedure has undergone many modifications since first being performed on a patient with tricuspid valve atresia in 1968. It is now the procedure of choice for individuals born with single-ventricle physiology or for those in whom a biventricular repair is not feasible. Forty years of experience with the Fontan procedure have gradually revealed the shortfalls of such a circulatory arrangement. Sequelae related to the underlying congenital anomaly or to the altered physiology of passive, nonpulsatile flow through the pulmonary arterial bed can result in failure of the Fontan circulation over time. Liver abnormalities including abnormalities in the clotting cascade have been well documented in Fontan patients. The clinical significance of these findings, however, has remained poorly understood. As Fontan survivors have increased in age and number, we have begun to better recognize subclinical hepatic dysfunction and the contribution of liver disease to adverse outcomes in this population. The purpose of this review is to summarize the existing data pertaining to liver disease in the Fontan population and to identify some questions that have yet to be answered.

Eur J Cardiothorac Surg. 2011 May;39(5):699-704. Epub 2010 Sep 29.

[Outcomes of ventricular assist device support in young patients with small body surface area.](#)

Fan Y, Weng YG, Xiao YB, Huebler M, Franz N, Potapov E, Hetzer R.

Source

Department of Cardiovascular Surgery, Chongqing Xinqiao Hospital, Third Military Medical University, Chongqing, China; Department of Cardiothoracic and Vascular Surgery, Deutsches Herzzentrum Berlin, Berlin, Germany.

Abstract

Objective: Although the ventricular assist device (VAD) has been a well-established therapy for larger adolescents and adult patients with advanced heart failure, current experience with the use of VAD for mechanical circulatory support in infants and young children with small body surface area is still limited. Methods: Between January 1999 and December 2009, 56 small children with body surface area 1.2m^2 were implanted with Berlin Heart EXCOR pediatric VAD in Germany Heart Institute Berlin. The etiology of end-stage myocardial failure included non-congenital (75%) and congenital heart disease (25%); the median age at implant was 1 year (12 days to 14 years), and the median support time was 55 days (1-432 days). Results: Of the 56 pediatric patients, 24 were bridged to heart transplantation, 12 were explanted following myocardial recovery, two continued to receive support, and the other 18 died on support. The accurate rate of survival on VAD support was $81.1\% \pm 5.8\%$ and $51.4\% \pm 9.3\%$ at 30 days and 1 year after EXCOR implantation. Patients receiving biventricular support had a trend towards higher post-implantation mortality as compared with children implanted with left ventricular assist device (LVAD) ($P=0.09$). Lower post-implantation survival was also observed in patients with congenital cardiac disease compared with children with a non-congenital etiology ($P<0.001$). Conclusions: Berlin Heart EXCOR pediatric VAD could provide satisfactory and safe circulatory support for small children with end-stage heart disease.