April & May, 2011

In This Issue

International Society for Adult Congenital Heart Disease

ISACHD Newsletter

President's Message

by *Barbara J.M. Mulder*

Dear ISACHD members,

In New Orleans it was decided to form working groups on 3 important strategic topics:

-Global healthcare delivery, including possible affiliation with WHF (chaired by Curt Daniels)
-Global education in ACHD (chaired by Erwin Oechslin)
-International research (chaired by Koichiro Niwa)

In this link you may find more information.

It is quite tremendous the level of experience, expertise and enthusiasm we are developing on these working groups. We have tapped into issues many feel passionate about and have experience to go along with the enthusiasm. Presently, the following members have volunteered:

Global Health Care:
Curt Daniels, Mike Landzberg, Jack Colman, Erik Meijboom, Mary Canobbio, Isabelle vonder Muhl, Luis Alday, Roberta Williams, Helmut Baumgartner, Disty Pearson, Kathi Kinnett, Peggy Powers, Clare O'Donnell, Marla Mendelson, Mahadevan Vaikom

Global Education:
Erwin Oechslin, Helmut Baumgartner, Els Pieper, Desiree Fleck, David Drajpuch, Harold Kaemmerer, Richard A. Krasuski, Isabelle vonder Muhll, Gary Webb
For a summary of activities and presentation at ISACHD meeting in New Orleans, click here.

International Research:
Koichiro Niwa, Mahadevan Vaikom, Philip Moons and Clare O'Donell

If you want to join one of these working groups, please contact the chair of the working group.

Barbara J.M. Mulder
President

21st Annual Congenital Heart Disease in the Adult: An International Symposium June 19-22, 2011 Cincinnati, OH

By Gary Webb

Program Overview

This 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.

Abstracts will be presented by ACHD providers from all backgrounds (physicians, nurses, sonographers). The top three physician abstracts and nursing/sonographer abstracts will be presented orally. Prizes will be awarded. The remaining abstracts will be presented in poster sessions, and there's time on the program for all registrants to review and discuss the posters.

We will record the program and to put it on the Internet by September 2011. As a result, registrants will be able to experience the programs they were interested in that they were not able to attend in person. Furthermore, the Internet version will be enhanced through the use of pretest and posttest questions to measure the effectiveness of the teaching experience.

Program Description

There will be an all-day **ACHD Echo Symposium on Sunday, June 19,** organized by Dr. Tom Kimball of Cincinnati Children's. At this program, consensus recommendations as to how best to perform echoes on adult patients with congenital heart defects will be presented. This program will appeal not only to the ACHD community, but more broadly to adult cardiologists, pediatric cardiologists, and sonographers.

Additionally, on **Sunday, June 19**, there will also be an all-day **ACHD Nursing Symposium** for nurse practitioners and other nonphysicians who work at ACHD clinics in North America and beyond.

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. Each will emphasize teaching points to the attendees and will be equipped with an audience response system to encourage audience engagement.

One room will feature case-based teaching. This will be led by major ACHD teams from North America and Europe. Each team will include four faculty members. Case

presentations will also be invited from other participants, as has been the case traditionally.

The second room will feature an academic and clinical agenda, including lectures, panel discussions, research competitions, and discussions of controversial issues.

Target Audience
The program is intended for pediatric and adult cardiovascular specialists and nurse practitioners/CCAs ***with a commitment to ACHD care***.

Regional News:

News in Asia-Pacific
by Koichiro Niwa

-13th South China Congress of Cardiology was held in Ganzhou, and there was a session on ACHD.
-4th ACHD Seminar (2 days teaching course of ACHD) will be held at Auditorium in St Luke's Hospital, Tokyo Japan (May 14, 15). Click here for more information.
Around 200 attendants will be expected.

News from Canada
by Erwin Oechslin

-The 5th *Cardiac Symposium: Hypoplastic Left Heart Syndrome* will be held in the Four Seasons Hotel from June 5-7, 2011.
-The *Pre-Symposium Course: 3 Dimensional Anatomy of the Heart* will be held from June 1-3, 2011.
More information and registration can be found here.

News from US
by Bill Davidson

ACHA 6th national conference this Fri-Sun atLAX Marriot in Los Angeles, has a 2 day professional track.

Cincinnati International ACHD Symposium in June. All day echo session Sun the 19th, 3 day symposium Mon- WEd (20-22)

AHA this Fall: Abstract submission closes June 1, 11, Registration opens June 22, 11 (earlier for FAHA and professional members), Sessions 11/11-16/11 in Orlando, FL

AHA website for Scientific Sessions: click here

Journal Watch

Article of the Month

Europace. 2011 Mar 8. [Epub ahead of print]

Cardiac outcome of pregnancy in women with a pacemaker and women with

untreated atrioventricular conduction block.

Thaman R, Curtis S, Faganello G, Szantho GV, Turner MS, Trinder J, Sellers S, Stuart GA.
Source: 1Bristol Congenital Heart Centre, Adult congenital Heart Unit, Bristol Royal Infirmary, Level 4 Dolphin House, King Edward Building, Bristol, UK.

Abstract
Aims
The natural history and outcome of pregnancy in patients with a pacemaker or those presenting with atrioventricular conduction block in pregnancy are unknown with only a limited number of case reports published. Methods and results This study examines the progress and outcome of 25 pregnancies in 18 women who were either paced or presented with untreated atrioventricular conduction block during pregnancy. All patients were seen in a single referral centre between 1998 and 2008 and were evaluated at regular intervals with ECG, echocardiography, and 24 h Holter. Four women (4 pregnancies) had new-onset atrioventricular block, 3 women (5 pregnancies) had previously diagnosed atrioventricular block who had not undergone pacing, and 11 women (16 pregnancies) had known atrioventricular block with a pacemaker prior to pregnancy. Of the four patients presenting for the first time in pregnancy, the frequency or severity of atrioventricular conduction block increased during pregnancy. One required pacing during and one after pregnancy. In two patients the conduction disturbance resolved postpartum. In the three patients who had known but untreated atrioventricular block before pregnancy, this progressed during each pregnancy but did not require pacing. In patients paced before pregnancy, there were no complications as a result of the pacemaker, but maternal complications were seen in patients with underlying structural heart disease. Conclusions Atrioventricular block in pregnancy is progressive; pacing is not always required but all patients should be closely monitored during and after pregnancy. In patients paced before pregnancy, pacing is well tolerated.

Commentary on Article of the Month

Commentary on paper 'Cardiac outcome of pregnancy in women with a pacemaker and women with untreated atrioventricular conduction block'
by Monica Benjamin

Nowadays, is not uncommon that female patients with a pacemaker or with atrioventricular conduction block reach childbearing age and contemplate undergoing pregnancy. [1] Striking advances in the management of conduction disturbances have been achieved over the past half century, mainly since permanent pacing became available in 1962 and widely implemented in the following years. This allowed survival in children with heart rhythm problems, especially those patients who had undergone open heart surgery for congenital heart disease and presented postoperative atrioventricular (AV) conduction block. [2] The real incidence and outcome of AV conduction block during pregnancy is unknown, being this a rare finding. There are only limited numbers of published case reports of patients with these features in the literature. [1,3-5] The aim of the retrospective study conducted by Thaman et al. is to describe the natural history and outcome of pregnancy in patients with a pacemaker or those presenting with AV conduction block in pregnancy.

The authors examined the course and outcome of 25 pregnancies in 18 women either paced or presenting with untreated AV conduction block during pregnancy. Two different groups were evaluated: the first group (11 patients, 16 pregnancies) included those patients who had a pacemaker implanted prior to pregnancy and were paced for a variety of reasons related to sinus node dysfunction or AV conduction disease. Five of these had underlying structural heart disease. The majority had prepectoral, followed by subpectoral transvenous pacemakers and abdominal epicardial pacemakers. There were no pacemaker or lead related complications during pregnancy. Maternal complications were seen in 3 patients who presented heart failure and atrial fibrillation, all of them with underlying structural heart disease. The second group comprised those patients with

untreated bradyarrhythmia or AV conduction disease at the time of pregnancy and was further divided into those presenting for the first time (4 patients, 4 pregnancies) and those with known conduction disorders but not previouslypaced (3 patients, 5 pregnancies). In this group, the severity of the bradyarrhythmia increased during the course of pregnancy. Two of these required pacemaker implant, one during pregnancy and the other following delivery. The remaining two were followed closely, not requiring pacing with resolution of conduction disorder postpartum. In those who had known but untreated atrioventricular conduction disease prior to pregnancy, the frequency and/ or severity of these also appeared to increase but in all cases regressed to pre pregnancy levels in the postpartum period. All had successful pregnancies without need of cardiac intervention.

Since there are no established guidelines for the management of pregnancy in these conditions, especially in those patients with untreated conduction disease, this study provides important information in this particular clinical scenario. Pacemakers were well tolerated in pregnancy according to data from previous studies. [1,3-5] This is also the first report showing that AV conduction block during pregnancy may resolve postpartum. The factors that could exacerbate conduction delay in pregnancy are unknown but may relate with normal hemodynamic changes, leading to an increase in the size in the cardiac chambers. The authors hypothesized that the increase in atrial stretching might be enough to provoke conduction disturbance in patients with a pre-existing substrate, also accounting for the resolution in the postpartum period. Therefore, not all the patients presenting with atrioventricular block during pregnancy require pacing. [1,5] Finally the outcome and course of pregnancy in the study were more likely determined by the underlying cardiac disorders. Despite the small number of patients, this study provides a significant contribution in the field. However further investigation is needed to provide guidelines for the proper management of theses patients.

1. Tateno S, Niwa K, Nakazawa M, Akagi T, Shinohara T, Yasuda T. Arrhythmia and conduction disturbances in patients with congenital heart disease during pregnancy: multicenter study. Circ J 2003; 67:992-7
2. Weindling SN, Saul JP, Gamble WJ, Mayer JE, Wessel D, Walsh EP. Duration of complete atrioventricular block after congenital heart disease surgery. Am J Cardiol 1998; 82:525-527.
3. Dalvi VB, Chanduri A, Kulkarni HL, Kale PA. Therapeutic guidelines for congenital complete heart block presenting in pregnancy. Int J Gynecol 1992;79: 802-4.
4. Sharma JB, Malhotra M, Pundit P. Successful pregnancy outcome with cardiac pacemaker after complete heart block. Int J Gynecol Obstet 2000;68:145-6.
5. Hidaka N ,ChibaY, Kurita T., Satoh S. and Nakano H:Is intrapartum temporary pacing required for women with complete atrioventricular block? An analysis of seven cases. BJOG 2006 ;113(5):605-7.

Article: *Thaman R., Curtis S. Faganello G, Szantho G.V,TurnerM. S., Trinder J.,Sellers S., and Stuart G.A:Cardiac outcome of pregnancy in women with a pacemaker and women with untreated atrioventricular conduction block. Europace 2011 Mar 8. [Epub ahead of print]*

Commentary: *Monica N. Benjamin, M.D.*
Senior Fellow
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Echocardiography. 2011 Apr:28(4):416-30. doi: 10.1111/j.1540-8175.2010.01359.x.
Impaired biventricular deformation in marfan syndrome: a strain and strain rate study in adult unoperated patients.

Kioteskoglou A, Saha S, Moggridge JC, Kapetanakis V, Goyindan M, Alpendurada F, Mullen MJ, Nassiri DK, Camm J, Sutherland GR, Bijnens BH, Child A.

Source

Department of Cardiac and Vascular Sciences, St George's University of London, London, UK Department of Clinical Physiology, Thoracic and Vascular Division, Karolinska Institutet at Sundsvalls Hospital, Sundsvall, Sweden MRC Biostatistics Unit, Institute of Public Health, Cambridge, UK Royal Brompton and Harefield Hospital NHS Trust, London, UK ICREA-Universitat Pompeu Fabra (CISTIB) and CIBER-BBN, Barcelona, Spain.

Abstract
Objective: To investigate the presence of any regional myocardial deformation abnormalities in Marfan syndrome (MFS) and determine the benefits of using advanced echocardiography compared to conventional techniques. Background: Myocardial dysfunction in MFS may be caused by extracellular matrix remodeling thus, resulting in uniform reduced functionality. However, increased aortic stiffness may cause segmental ventricular abnormalities. Strain rate imaging (SRI) constitutes a validated technique to assess regional deformation in various clinical conditions. With this in mind, we aimed to investigate biventricular function in MFS using SRI. Methods: Forty-four MFS patients (mean age 30 ± 12 years, 26 men) and 49 controls without valvular disease were examined using SRI. Ejection fraction (EF) was calculated by the Simpson's biplane method. Biventricular deformation was assessed by measuring strain/strain rate. Strain values were divided by left ventricular (LV) end-diastolic volume to adjust LV deformation for geometry changes providing a strain index (SI). Aortic stiffness was evaluated using the β-stiffness index. Results: EF (%) was reduced in MFS patients (59 ± 5 vs 72 ± 4, $P < 0.001$), whereas β-stiffness was increased ($P < 0.001$). LV radial and LV and right ventricular (RV) long-axis strain values (%) were reduced in the patient group (70 ± 17 vs 93 ± 10; 19 ± 2 vs 25 ± 2; 30 ± 9 vs 36 ± 8, respectively, $P < 0.001$). Strain rate measurements were also reduced ($P < 0.001$). In a multiple regression analysis, MFS diagnosis was negatively associated with LV SI (-0.262 [-0.306, -0.219], $P < 0.001$). β-Stiffness was negatively associated with SI obtained from the septum, inferior and anterior walls. ROC analyses demonstrated that SRI, when compared with conventional echocardiography, had higher sensitivity and specificity in predicting biventricular dysfunction in MFS. Conclusions: Our study showed a uniform reduction in biventricular deformation in MFS. These findings suggest that assessment of myocardial function using advanced echocardiographic techniques could be more accurate in MFS patient evaluation than conventional echocardiography alone. (Echocardiography 2011;28:416-430).

Arch Cardiovasc Dis. 2011 Mar;104(3):155-60. Epub 2011 Mar 25.

Early single clinical experience with the new Figulla ASD Occluder for transcatheter closure of atrial septal defect in adults.

Cansel M, Pekdemir H, Yağmur J, Tasolar H, Ermis N, Kurtoglu E, Acıkgoz N, Atas H, Ozdemir R.
Source: Faculty of Medicine, Inonu University, Malatya, Turkey.

Abstract

BACKGROUND:
Recently, the Occlutech Figulla ASD Occluder (FSO) has been introduced for transcatheter closure of atrial septal defects. This device can be used for transcatheter closure of small as well as large atrial septal defects.
AIMS:
To evaluate the feasibility and short-term results of transcatheter closure of secundum type atrial septal defects using the FSO device in adult patients.
METHODS:
Seventy-four consecutive adult patients were referred for transcatheter closure of secundum large atrial septal defects ("stretched" diameter>20mm and/or invasive pulmonary/systemic flow [Qp/Qs] ratio>1.5) using the FSO device.
RESULTS:
The FSO device was successfully implanted in 68 patients (mean±SD [range] age: 31.8±12.3 [17-64] years; weight: 71.5±18.4 [49-98]kg). All patients had right atrial and ventricular volume overload with a mean Qp/Qs ratio of 2.5±0.6 (range 1.5-3.8). Mean atrial septal defect diameter was 22.3±4.8 (range 12-33)mm and the size of the implanted FSO was 24.1±4.9 (range 12-36)mm. Two patients had trivial (jet width<1mm in diameter) residual shunts and one patient had a small (1-2mm) residual shunt. There were no moderate or severe residual shunts. No device embolization or other serious complication occurred during either the procedure or the follow-up.
CONCLUSION:
The present study found that transcatheter closure of isolated secundum atrial septal defects using the novel design of the FSO device was safe, effective, and had an excellent outcome during the 6month follow-up period.

Curr Cardiol Rep. 2011 Apr 12. [Epub ahead of print]

Monitoring the Patient with Transposition of the Great Arteries: Arterial Switch Versus Atrial Switch.

Roche SL, Silversides CK, Oechslin EN.
Source: Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network/Toronto General Hospital, 585 University Avenue, Toronto, ON, M5G 2N2, Canada, lucy.roche@uhn.on.ca.

Abstract
Sufficient time has passed that adult congenital heart disease (ACHD) specialists now frequently encounter survivors born with complete transposition of the great arteries and palliated with an atrial or arterial switch procedure. To ensure the ongoing health of these patients, it is of paramount importance that their surgeries are understood and that physicians are aware of and remain vigilant for potential late complications. Adult survivors should be assessed annually in a regional ACHD center. Clinical assessment, electrocardiogram, and multimodality imaging are the mainstay of routine monitoring. Doppler echocardiography is the first-line imaging modality; other diagnostic tests are tailored to seek specific long-term complications. Clinicians, specialists in cardiovascular imaging, nurses and others involved in the delivery of care need special training and expertise. Care for these complex patients is best provided by multidisciplinary teams located in regional ACHD centers with access to adequate human and structural resources.

J Card Surg. 2011 Apr 11. doi: 10.1111/j.1540-8191.2011.01249.x. [Epub ahead of print]

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

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.

Catheter Cardiovasc Interv. 2011 Mar 31. doi: 10.1002/ccd.22964. [Epub ahead of print]

Hybrid aortic reconstruction for treatment of recurrent aortic obstruction after stage 1 single ventricle palliation: Medium term outcomes and results of redilation.

Kutty S, Burke RP, Hannan RL, Zahn EM.
Source: University of Nebraska/Creighton University Joint Division of Pediatric Cardiology, Children's Hospital and Medical Center, Omaha, Nebraska.

Abstract

OBJECTIVE:
We describe a hybrid approach to the treatment of aortic obstruction after stage 1 palliation (S1P) of hypoplastic left heart syndrome.
BACKGROUND:
Recurrent aortic obstruction is a common problem after S1P of hypoplastic left heart syndrome. Even mild aortic obstruction is poorly tolerated so early and definitive therapy is desirable. Although stent implantation is an effective treatment for aortic obstruction in older children and adults, technical issues due to small vessels and concerns regarding future potential for expansion have generally precluded the use of stents in this setting.
METHODS:
Six patients underwent hybrid aortic reconstruction (HAR) in the operating room or catheterization laboratory, with the interventional cardiologist and cardiac surgeon working in cooperation.
RESULTS:
Patients had a mean weight of 5.8 kg (2.9-7.7) and a mean age of 5.6 months (0.5-12.9) at the time of HAR. Five patients had undergone prior balloon angioplasty at a mean age of 2.8 months (2.1-3.5), and five had moderately depressed single ventricular function prior to HAR. The balloons used had a diameter of 7-10 mm and introducer sheath size ranged from 6 to 10 F. There were no immediate or late procedure related complications. Stent redilation was performed in 5 patients for relief of recurrent obstruction or to keep pace with somatic growth. At a median follow up of 4.8 years (0.2-7.9), there were 3 patients progressing well after Fontan palliation and 3 deaths.
CONCLUSIONS:
HAR allows for placement of stents that can ultimately reach adult size in small infants who have recurrent aortic obstruction after balloon angioplasty following S1P. Advantages include freedom from delivery sheath constraints when determining stent type/size, facilitation of precise stent position, and avoidance of vascular damage or hemodynamic compromise during the procedure. Longer follow-up and larger experience are required to determine if this therapy will provide a long-term solution to this difficult problem.

Congenit Heart Dis. 2011 Mar 28. doi: 10.1111/j.1747-0803.2011.00504.x. [Epub ahead of print]

Liver Disease in the Patient with Fontan Circulation.

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

J Card Fail. 2011 Apr;17(4):265-71. Epub 2011 Jan 21.

Response to inhaled nitric oxide predicts survival in patients with pulmonary hypertension.

Krasuski RA, Devendra GP, Hart SA, Wang A, Harrison JK, Bashore TM.
Source: Director of Adult Congenital Heart Disease Service, Division of Cardiovascular Medicine, Cleveland Clinic Foundation, Cleveland, OH.

Abstract
OBJECTIVE:
To examine the ability of vasodilator response to predict survival in a diverse cohort of patients with pulmonary hypertension (PH).
PATIENTS & METHODS:
A total of 214 consecutive treatment-naive patients referred for invasive PH evaluation were enrolled between November 1998 and December 2008. Vasoreactivity was assessed during inhalation of 40 parts per million nitric oxide (iNO) and vasodilator responders were defined as those participants who achieved a mean pulmonary artery pressure (PAP) of ≤ 40 mm Hg and a drop in mean PAP ≥ the median for the cohort (13%). Kaplan-Meier analysis and Cox proportional hazards modeling were used to identify predictors of survival.
RESULTS:
There were 51 deaths (25.9%) over a mean follow-up period of 2.3 years. Kaplan-Meier analysis demonstrated that vasodilator responders had significantly improved survival ($P < .01$). Vasodilator responders had improved survival regardless of whether or not they had idiopathic or nonidiopathic PH ($P = .02$, $P < .01$) or whether or not they had Dana Point class 1 or non-Dana Point class 1 PH ($P < .01$, $P = .01$). In multivariate modeling, advanced age, elevated right atrial pressure, elevated serum creatinine, and worsened functional class significantly predicted shorter survival ($P = .01$, $P = .01$, $P = .01$, $P < .01$), whereas vasodilator response predicted improved survival ($P = .01$).
CONCLUSIONS:
Vasodilator responsiveness to iNO is an important method of risk stratifying PH patients,

with results that apply regardless of clinical etiology.

Congenit Heart Dis. 2011 Mar 25. doi: 10.1111/j.1747-0803.2011.00493.x. [Epub ahead of print]

Interventional Closure of Atrial Septal Defects in Adult Patients with Ebstein's Anomaly.

Jategaonkar SR, Scholtz W, Horstkotte D, Kececioglu D, Haas NA.
Source: Departments of Cardiology Congenital Heart Defects, Heart and Diabetes Center North Rhine-Westphalia, Ruhr University Bochum, Bad Oeynhausen, Germany.

Abstract
Ebstein's anomaly is frequently associated with interatrial communications. In patients with severe tricuspid regurgitation standard treatment is the surgical repair or replacement of the tricuspid valve and patch closure of the atrial septal defect. We sought to evaluate the feasibility and short-term outcome of interventional device closure of interatrial communications in Ebstein patients with mild to moderate tricuspid regurgitation and various degrees of clinical symptoms. In this case series of 9 patients the device closure could be performed safely and 8 of 9 patients improved in their exercise capacity or clinical condition. However, the patients need to be selected carefully and appropriately for this palliative method. In those with predominant left-to-right shunting, ASD-closure reduces the volume load of the right ventricle and can be performed according to routine procedures. In those patients with cyanosis and right-to-left shunting however, test occlusion of the interatrial communication with adequate balloon size followed by careful examination of the hemodynamics at rest and under catecholamine stimulation is compulsatory to evaluate the feasibility of device closure. The tricuspid regurgitation should not exceed moderate level, the right atrial and ventricular pressure should be within normal range for an adequate time during test occlusion and the systemic blood pressure and cardiac output maintained safely.

Int J Cardiol. 2011 Mar 22. [Epub ahead of print]

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: Cardiovascular Research Laboratory, Biomedical Research Foundation, Academy of Athens, Athens, Greece.

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-a, 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.

Obstet Gynecol. 2011 Apr;117(4):886-91.

Effect of maternal heart disease on fetal growth.

Gelson E, Curry R, Gatzoulis MA, Swan L, Lupton M, Steer P, Johnson M.
Source: From the Academic Department of Obstetrics and Gynaecology, Imperial College London, Chelsea and Westminster Hospital, London, United Kingdom; Adult Congenital Heart Centre, National Heart and Lung Institute, Imperial College London, Royal Brompton Hospital, London, United Kingdom; and Chelsea and Westminster Healthcare NHS Foundation Trust, London, United Kingdom.

Abstract
OBJECTIVE:
To estimate the effect of maternal heart disease on fetal growth and neonatal outcomes.
METHODS:
A retrospective cohort study of all women with congenital and acquired heart disease admitted at Chelsea and Westminster Hospital between 1994 and 2010 was performed. The women who delivered immediately before and immediately after each index pregnancy were used as controls. Data were obtained from medical and obstetric notes. Birth weight percentiles were calculated using a customized birth weight percentile program, and neonatal complications (preterm birth, perinatal mortality, and recurrence of congenital heart disease) were noted.
RESULTS:
Median birth weight percentile was significantly lower in the heart disease group (n=31) compared with the control group (n=49; P<.001 Mann-Whitney U test). The rate of neonatal complications was significantly higher in the heart disease group (34% compared with 15%). Preterm birth occurred in 42 (13%) pregnancies, of which 67% were iatrogenic. Eighty-one (25%) newborns in the heart disease group were small for gestational age, and there were four stillbirths and four neonatal deaths (perinatal mortality rate 20 per 1,000).
CONCLUSION:
This cohort study suggests a significant reduction in fetal growth rates associated with maternal heart disease, which is also associated with preterm delivery and reduced birth weight. The presence of maternal cyanosis and a reduced cardiac output are the most significant predictors.

Congenit Heart Dis. 2011 Mar 21. doi: 10.1111/j.1747-0803.2011.00503.x. [Epub ahead of print]

Hyponatremia and Its Association with the Neurohormonal Activity and Adverse Clinical Events in Children and Young Adult Patients after the Fontan Operation.

Ohuchi H, Negishi J, Ono S, Miyake A, Toyota N, Tamaki W, Miyazaki A, Yamada O.
Source: Departments of Pediatric Cardiology, National Cardiovascular Center, Osaka, Japan.

Abstract

Background: Hyponatremia (HN) is relatively common in adults with congenital heart disease and is a powerful predictor of mortality. However, the precise relationship of HN to the Fontan pathophysiology remains unknown.
Purpose: Our study aimed to clarify the association of HN to the Fontan pathophysiology. We measured the plasma sodium (Na) level in 169 consecutive Fontan patients (78 children) and HN (<137 mEq/L) was observed in 50 patients (30% of the total patients, 31% of the children). The HN patients showed a lower peak oxygen uptake (VO(2)) with a greater New York Heart Association class (P < .0001). The plasma level of norepinephrine (NE), rennin activity (PRA), arginine vasopressin, central venous pressure (CVP) and medications were associated with the Na levels and the NE, PRA, and diuretic use were the independent determinants (P < .01 - .0001). The plasma B-type natriuretic peptide was

not correlated with the Na levels. In the children, diuretic use and the PRA independently determined the Na levels without any association to the CVP or peak VO(2). During a median follow-up of 2.1 years, the HN in addition to the CVP and peak VO(2) independently predicted the unscheduled hospitalizations in all patients, while the HN was the only independent predictor of the hospitalizations in the adult patients (hazard ratio: 3.1, 95% confidence interval 1.2-8.0, P= .021).
Conclusions: Child and adult Fontan patients exhibited a high prevalence for HN that closely reflected some neurohumoral activation and predicted adverse clinical events, especially in adult Fontan patients.

Congenit Heart Dis. 2011 Mar;6(2):134-8. doi: 10.1111/j.1747-0803.2011.00498.x. Epub 2011 Mar 21.

Assessment of Electronic Health Information System Use and Need in US Adult Congenital Heart Disease Centers.

Weiss JB, Grant A, Marelli A, Khairy P, Maurais T, Rehel S, Chetaille P, Broberg CS.
Source: Adult Congenital Heart Disease Program, Oregon Health and Science University, Portland, Ore, USA Collaboration in Research for Effective Diagnostics (CRED), Université de Sherbrooke, Sherbrooke MAUDE Unit (McGill Adult Unit for Congenital Heart Disease), McGill University Health Center, Montreal Adult Congenital Heart Centre, Montreal Heart Institute, University of Montreal, Montreal Quebec Heart and Lung Institute, Quebec City, QC, Canada.

Abstract
Objectives.Efforts to improve care for adult congenital heart disease (ACHD) patients necessitates collection of accurate, detailed, longitudinal data. We sought to document what electronic health record systems are currently available at ACHD centers and to assess national interest in a uniform ACHD-focused system.
Design.Directors of ACHD centers in the United States were invited to complete an online questionnaire regarding current health information systems at their institution both for general cardiology and for ACHD. Topics that were surveyed included utility and perceived limitations of currently available systems. The survey also assessed the level of interest in an ACHD-specific system, and its optimal functions.
Results.Thirty-four centers responded, representing both pediatric and adult institutions that care for patients with ACHD. Of these, 80% reported using a variety of commercially supported electronic medical record products, whereas only 50% employed an ACHD-specific noncommercial database to supplement their institutional system. Comparison of the two systems revealed that most clinical activities are pursued through the institutional electronic medical record system. Research and tracking of clinical activities were the primary uses of ACHD-specific systems, which have several noted limitations. Strong interest in an integrated ACHD-specific system was found among responders.
Conclusions.There is both an unmet need and a strong interest in an ACHD-oriented electronic health record that could facilitate research, outcome tracking, quality assurance, and inter-institutional collaboration, all functions that are lacking in electronic health systems currently in use.

Congenit Heart Dis. 2011 Mar;6(2):147-56. doi: 10.1111/j.1747-0803.2011.00497.x. Epub 2011 Mar 21.

Pregnancy in women with congenital heart disease: the impact of a systemic right ventricle.

Jain VD, Moghbeli N, Webb G, Srinivas SK, Elovitz MA, Paré E.
Source: Division of Maternal Fetal Medicine, Christiana Hospital, Newark, Del Women's Cardiovascular Center, Pennsylvania Hospital Division of Maternal Fetal Medicine, Center for Reproductive Research and Women's Health (CRRWH) Division of Maternal Fetal Medicine, University of Pennsylvania, Philadelphia, Pa Cincinnati Children's Hospital, Adolescent and Adult Congenital Heart Disease Program, Cincinnati, Ohio, USA.

Abstract

Objective. Individuals with a systemic right ventricle develop cardiac complications earlier in life. Limited data exists regarding the effect of a maternal systemic right ventricle on cardiac events during pregnancy. We sought to assess the effect of a systemic right ventricle on cardiac events and pregnancy outcomes.
Design. The study was designed as a retrospective cohort study of pregnant women with maternal congenital heart disease. Setting.The study was set in a university, academic tertiary care referral center. Patients.Study subjects were identified by International Statistical Classification of Diseases and Related Health Problems-9 codes. Women with mitral valve prolapse only or noncongenital cardiac disease were excluded. The exposure was defined by systemic ventricle.
Outcome Measures.The primary outcome was a composite of congestive heart failure, arrhythmia, stroke, cardiac arrest/death during pregnancy or postpartum (CARDCOMP). The secondary outcome (PREGCOMP) was a composite of preterm delivery, preeclampsia, growth restriction, and stillbirth/pregnancy loss (PREGCOMP). Student's t-test or chi-square/Fisher's exact tests were used for comparison of continuous/categorical variables. Multivariable logistic regression was performed to control for possible confounders.
Results.One hundred forty-six pregnancies in 114 women were included; 15 (10.3%) pregnancies involved a systemic right ventricle. CARDCOMP complicated 12.3% of these pregnancies. Women with a systemic right ventricle were more likely to develop CARDCOMP even after adjustment for confounders (odds ratio [OR] 6.32 [1.7-23.5], P= .006). PREGCOMP complicated 40.4% of all pregnancies. Women with a systemic right ventricle were also more likely to develop PREGCOMP (OR 5.37 [1.4-20.7], P= .015) compared with women with a systemic left ventricle after controlling for confounders.
Conclusion.In women with congenital heart disease, a systemic right ventricle is associated with adverse cardiac and pregnancy outcomes. This information is critical for counseling and caring for these women. Further investigation is warranted regarding the effect of pregnancy on long-term health for this unique cohort of women.

Congenit Heart Dis. 2011 Mar;6(2):139-46. doi: 10.1111/j.1747-0803.2011.00490.x. Epub 2011 Mar 21.

Effects of Pulmonary Vasodilator Therapy on Ventilatory Efficiency during Exercise in Adults with Eisenmenger Syndrome.

Yang-Ting S, Aboulhosn J, Sun XG, Child JS, Sietsema KE.
Source: Kaiser Permanente Medical Center, Downey, Calif, USA David Geffen School of Medicine at UCLA, Los Angeles, Los Angeles, Calif, USA Harbor-UCLA Medical Center, Torrance, Calif, USA.

Abstract
Objective. Eisenmenger syndrome, characterized by systemic-level pulmonary arterial resistance with resultant right-to-left shunt, is associated with low exercise capacity and hyperpnea at rest and exercise. Because ventilatory requirements are augmented by right-to-left shunting, we hypothesized that if pulmonary vasodilator treatment improved pulmonary perfusion in this condition, this would also improve ventilatory efficiency during exercise.
Design. To investigate this, data from incremental cardiopulmonary exercise tests performed by Eisenmenger patients before and after beginning therapy with pulmonary hypertension medications were retrospectively analyzed.
Setting. Ahmanson/University of California, Los Angeles Adult Congenital Heart Disease Center.
Patients. Ten adults with Eisenmenger syndrome treated with either bosentan or sildenafil.

Outcome Measures. The primary analysis was comparison, before and after treatment, of the efficiency of exercise ventilation as reflected in the ratio of ventilation () to carbon dioxide output () measured at the anaerobic threshold (AT), the slope of during incremental exercise, and end tidal partial pressure of CO(2) (PETCO(2)) at the AT. Secondary measures included peak oxygen uptake () and AT.
Results. Following treatment there were significant reductions in the slope (59.5 +- 12.9 vs. 50.0 +- 7.2, P= .003), and significant decrease in ratio (56.9 +- 6.2 vs. 50.2 +- 5.9, P=

.00004) and increase in PETCO (2) (21.12 +- 2.43 vs 23.9 +- 2.62 torr, P= .0092) measured at the AT. Increases in peak (0.73 +- 0.25 vs 0.78 +- 0.32 L/min, P= .333) and AT (0.61 +- 0.20 vs. 0.68 +- 0.25 L/min, P= .154) were not significant.
Conclusions. These findings are consistent with reduction in right-to-left shunt due to improved pulmonary blood flow, though attenuation of ventilatory drive is not excluded. Treatment of adult Eisenmenger patients with pulmonary the pulmonary vasodilators bosentan or sildenafil leads to improvement in parameters of ventilatory efficiency during exercise.

J Interv Cardiol. 2011 Mar 17. doi: 10.1111/j.1540-8183.2011.00638.x. [Epub ahead of print]

Medium-Term Outcomes for Peripheral Pulmonary Artery Stenting in Adults with Congenital Heart Disease.

Kenny D, Amin Z, Slyder S, Hijazi ZM.
Source: From the Rush Center for Congenital and Structural Heart Disease, Rush University Medical Center, Chicago, Illinois.

Abstract
Objective: We describe our medium-term outcomes for peripheral pulmonary artery stenting in adults with congenital heart disease. Background: Improved outcomes in congenital heart disease have lead to the evolution of transcatheter therapies in adults aimed at maintaining normal hemodynamics. Stenting for pulmonary artery narrowing is effective in children, however little is known about outcomes in adults.
Methods: Retrospective data analysis and follow-up review of our complete experience with peripheral pulmonary artery stenting in adults.
Results: Over a 9-year period we carried out 15 procedures, implanting 23 stents (11-left pulmonary artery) in the pulmonary arteries of 12 adult patients (7 female). Eleven patients had previous cardiac surgery. Median age at implantation was 32.5 years (range, 18.7-56.7 years) with median weight of 71.3 kg (range, 44.5-95 kg). Six patients underwent bilateral pulmonary artery stenting. Median procedure time was 140 minutes (range, 76-263 minutes) and 4 patients had other interventions performed during the same procedure. Median systolic pressure gradient across the narrowing's of 24 mmHg (range, 11-61 mmHg) was reduced to 3 mmHg (range, 0-17 mmHg) postprocedure ($P < 0.001$). Three patients had acute stent embolization, one of whom required surgical removal. No aneurysm formation or significant stent fractures have been noted on median follow-up of 27.4 months (range, 1-97 months). Two patients required reintervention with further stent implantation.
Conclusions: Pulmonary artery stenting provides effective relief of narrowing in adults with congenital heart disease. Bilateral and/or multiple stenting are often required. Stent embolization may occur particularly in patients with associated significant pulmonary regurgitation.

J Am Soc Echocardiogr. 2011 Mar 8. [Epub ahead of print]

Reference Values for Myocardial Two-Dimensional Strain Echocardiography in a Healthy Pediatric and Young Adult Cohort.

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Source: Children's Heart Centre, Radboud University Nijmegen Medical Centre, Nijmegen, The Netherlands.

Abstract
BACKGROUND:
The accurate evaluation of intrinsic myocardial contractility in children with or without congenital heart disease (CHD) has turned out to be a challenge. Two-dimensional strain echocardiographic (2DSTE) imaging or two-dimensional speckle-tracking echocardiographic imaging appears to hold significant promise as a tool to improve the

assessment of ventricular myocardial function. The aim of this study was to estimate left ventricular myocardial systolic function using 2DSTE imaging in a large cohort consisting of healthy children and young adults to establish reference strain values.
METHODS:
Transthoracic echocardiograms were acquired in 195 healthy subjects (139 children, 56 young adults) and were retrospectively analyzed. Longitudinal, circumferential, and radial peak systolic strain values were determined by means of speckle tracking. Nonlinear regression analysis was performed to assess the effect of aging on these 2DSTE parameters.
RESULTS:
There was a strong, statistically significant second-order polynomial relation ($P < .001$) between global peak systolic strain parameters and age. Global peak systolic strain values were lowest in the youngest and oldest age groups.
CONCLUSION:
This is the first report to establish age-dependent reference values per cardiac segment for myocardial strain in all three directions assessed using 2DSTE imaging in a large pediatric and young adult cohort. There is a need to use age-specific reference values for the adequate interpretation of 2DSTE measurements.

Mol Cell Biomech. 2011 Mar;8(1):21-42.

Comparison of hemodynamic endpoints between normal subject and tetralogy patient using Womersley velocity profile and MR based flow measurements.

Das A, Gottliebson WM, Karve M, Banerjee R.
Source: School of Dynamic System, University of Cincinnati, Cincinnati, Ohio, USA.

Abstract
Right ventricular (RV) enlargement and pulmonary valve insufficiency (PI) are well-known, unavoidable long term sequelae encountered by patients who undergo tetralogy of Fallot (TOF) surgery. Despite their lifelong need for cardiac surveillance and occasional re-intervention, there is a paucity of numerical data characterizing blood flows in their pulmonary arteries (PA). Specifically, although PA regurgitation is well-known to be ubiquitously present in adult repaired TOF (rTOF) patients yet, there have been only limited numerical studies to fully characterize this process. The few studies available have utilized idealized, simplistic geometric models or overly simplistic boundary conditions that fail to account for flow reversals near the arterial walls as observed in in-vitro and MRI based in-vivo studies. The objective of this study was to establish and validate a numerical methodology of PA blood flow using actual patient specific geometry and flow measurements obtained using phase-contrast MRI, employing Womersley type velocity profiles that model flow reversals near walls. The results from computation were validated with measurements. For the normal subject, the time averaged right PA pressure from computation (13.8 mmHg) and experiment (14.6 mmHg) differed by 6%. The time-averaged main PA pressure from computation (16.5 mmHg) and experiment (16.3 mmHg) differed by 1%. The numerically computed left PA regurgitant fraction was 89% compared to measured 77.5%, while the same for the rTOF was 43% (computation), compared to 39.6% (measured). We conclude that the use of numerical computations using the Womersley boundary condition allows reliable modeling of the pathophysiology of PA flow in rTOF.

J Cardiovasc Nurs. 2011 Mar 2. [Epub ahead of print]

Health Care Needs of Adults with Congenital Heart Disease: Study of the Patient Perspective.

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Source: Jeanine L. Harrison, MN, NP Nurse Practitioner, Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Canada. Candice K. Silversides, MD Cardiologist, Research Director, Toronto

Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Canada. Erwin N. Oechslin, MD Director, Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Canada. Adrienne H. Kovacs, PhD Psychologist, Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Centre, University Health Network, University of Toronto, Canada.

Abstract

BACKGROUND:
More than 90% of infants born with congenital heart disease reach adulthood. International medical recommendations outline patient care needs in an effort to optimize patient health. There are, however, limited data focusing on the patient perspective.
OBJECTIVES:
This study investigated adult congenital heart disease patient-reported (1) barriers to medical care, (2) health care behaviors, and (3) concerns regarding medical, psychosocial, and lifestyle matters.
METHODS:
In this cross-sectional study, a questionnaire was distributed to all patients who attended a patient education conference.
RESULTS:
There were 123 adult congenital heart disease participants (58% female; mean age, 37 [SD, 13] years). The most common self-reported cardiac diagnoses were tetralogy of Fallot and transposition of the great arteries. Most patients did not report transportation or financial barriers to care, but did report the following: not wanting further surgery even if it was recommended (18%), not liking to think or talk about one's heart (17%), and not understanding doctors' information; 8% of patients inaccurately considered themselves to be "cured." With regard to health care behaviors, more than 80% of patients reported annual family physician and dentist visits, but 34% of patients were unaware when to seek urgent medical attention. Patients reported moderate to extreme concern about the following medical topics: heart rhythm problems (82%), infections (74%), and understanding treatment options (71%). Patients most often reported moderate to extreme concern about the following lifestyle and psychosocial topics: physical activity (77%), insurance (72%), assuming increased health responsibility (73%), diet (71%), mental health (60%), and death and dying (57%).
CONCLUSIONS:
This study provides important information about 3 specific areas. First, there are potential barriers to care beyond financial and transportation challenges. Second, many patients require education regarding when to seek urgent medical attention. Third, the concerns of this patient population are not limited to medical information. A patient-centered educational program is recommended.

J Invasive Cardiol. 2011 Mar;23(3):120-4.

Initial experience with the Amplatzer Vascular Plug IV in congenital heart disease: coronary artery fistula and aortopulmonary collateral artery embolization.

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Source: Department of Paediatric Cardiology and Adult Congenital Heart Disease, Policlinico San Donato IRCCS, Via Morandi 30, San Donato Milanese (MI), Italy.

Abstract

BACKGROUND:
A number of percutaneous devices are available to embolize anomalous vessels in congenital heart disease. We report our initial single-center experience with the new Amplatzer Vascular Plug IV (AVP IV) in congenital heart disease to embolize a coronary artery fistula and aortopulmonary collateral arteries in 4 cases.
METHODS:
From August 2009 until April 2010, 7 AVP IV devices were deployed in 4 patients, age range 5 months to 9 years, weight 3.5-27.7 kg. One patient had a large coronary artery

fistula, the others had anomalous aortopulmonary collaterals; 2 patients had tetralogy of Fallot with pulmonary atresia, with the other having dextrocardia, anomalous pulmonary venous drainage and pulmonary atresia.
RESULTS:
In all 4 patients, vessels intended to be closed with the AVP IV were closed successfully with minimal residual shunting and no device failures. Deployed devices ranged from 4-8 mm in diameter. One patient had 4 devices, closing large branching infradiaphragmatic aortopulmonary collaterals. The other 3 patients had single devices. Complete vessel embolization was seen with no device embolization or implantation complication.
CONCLUSION:
This preliminary experience illustrates the utility of the AVP IV in congenital heart disease, occluding a coronary artery fistula and aorto-pulmonary collaterals, with devices between 4 mm and 8 mm in diameter, demonstrating its safety and effectiveness. It is particularly useful in embolizing difficult-to-reach large, tortuous vessels with a small-sized catheter in a single procedure. Longer-term follow up in a larger cohort of patients will be required to establish long-term efficacy and device safety.

Am J Cardiol. 2011 Apr 15;107(8):1215-20. Epub 2011 Feb 23.

Prevalence of left ventricular systolic dysfunction in adults with repaired tetralogy of fallot.

Broberg CS, Aboulhosn J, Mongeon FP, Kay J, Valente AM, Khairy P, Earing MG, Opotowsky AR, Lui G, Gersony DR, Cook S, Ting JG, Webb G, Gurvitz MZ; Alliance for Adult Research in Congenital Cardiology (AARCC).
Source: Oregon Health and Science University, Portland, Oregon.

Abstract

Left ventricular (LV) systolic dysfunction has been observed in patients with repaired tetralogy of Fallot (TOF), although its clinical associations are unknown. Adults with repaired TOF were identified from 11 adult congenital heart disease centers. Clinical history was reviewed. Patients with pulmonary atresia were excluded. Echocardiograms were reanalyzed to estimate LV ejection fraction. LV function was defined as normal (LV ejection fraction ≥55%) or mildly (45% to 54%), moderately (35% to 44%), or severely (<35%) decreased. Right ventricular (RV) and LV dimensions and Doppler parameters were remeasured. Function of all valves was qualitatively scored. Of 511 patients studied, LV systolic dysfunction was present in 107 (20.9%, 95% confidence interval 17.4 to 24.5). Specifically, 74 (14.4%) had mildly decreased and 33 (6.3%) had moderately to severely decreased systolic function. Presence of moderate to severe LV dysfunction was associated with male gender, LV enlargement, duration of shunt before repair, history of arrhythmia, QRS duration, implanted cardioverter-defibrillator, and moderate to severe RV dysfunction. Severity or duration of pulmonary regurgitation was not different. In conclusion, LV systolic dysfunction was found in 21% of adult patients with TOF and was associated with shunt duration, RV dysfunction, and arrhythmia.

Circulation. 2011 Mar 1;123(8):896-903. Epub 2011 Feb 14.

Comparison of the structure of the aortic valve and ascending aorta in adults having aortic valve replacement for aortic stenosis versus for pure aortic regurgitation and resection of the ascending aorta for aneurysm.

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Abstract

BACKGROUND:
There is debate concerning whether an aneurysmal ascending aorta should be replaced when associated with a dysfunctioning aortic valve that is to be replaced. To examine this

issue, we divided the patients by type of aortic valve dysfunction-either aortic stenosis (AS) or pure aortic regurgitation (AR)-something not previously undertaken.
METHODS AND RESULTS:
Of 122 patients with ascending aortic aneurysm (unassociated with aortitis or acute dissection), the aortic valve was congenitally malformed (unicuspid or bicuspid) in 58 (98%) of the 59 AS patients, and in 38 (60%) of the 63 pure AR patients. Ascending aortic medial elastic fiber loss (EFL) (graded 0 to 4+) was zero or 1+ in 53 (90%) of the AS patients, in 20 (53%) of the 38 AR patients with bicuspid valves, and in all 12 AR patients with tricuspid valves unassociated with the Marfan syndrome. An unadjusted analysis showed that, among the 96 patients with congenitally malformed valves, the 38 AR patients had a significantly higher likelihood of 2+ to 4+ EFL than the 58 AS patients (crude odds ratio: 8.78; 95% confidence interval: 2.95, 28.13).
CONCLUSIONS:
These data strongly suggest that the type of aortic valve dysfunction-AS versus pure AR-is very helpful in predicting loss of aortic medial elastic fibers in patients with ascending aortic aneurysms and aortic valve disease.

Heart. 2011 Mar;97(5):394-9.

Efficacy of percutaneous closure of patent foramen ovale: comparison among three commonly used occluders.

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Abstract
BACKGROUND:
Percutaneous closure of patent foramen ovale (PFO) is standard treatment for patients with paradoxical embolism but studies examining the efficacy of the various occluders are lacking.
OBJECTIVE:
To evaluate short- and medium-term closure rates of three common occluders.
METHODS:
One hundred and sixty-six adults (47±12 (18-81 years)) were evaluated with transthoracic bubble echocardiography before and after PFO closure. Only patients with large PFOs were included (>30 bubbles in the left heart after Valsalva).
RESULTS:
Three occluders were used: Amplatzer (AGA Medical Corporation) (n=80, 48%), Gore Helex (n=48, 29%) and Premere TM (St Jude Medical) (n=38, 23%). One (0.6%) neurological event occurred during follow-up. At 6 months significant residual shunting after Valsalva was highest in the group that received the Helex (58.3%), and lower for Premere (39.5%) and Amplatzer (32.5%). At final follow-up residual shunting remained higher in patients with the Helex (33.3%) than in Premere (18.5%) and Amplatzer (11%). Amplatzer had a significantly lower residual shunt rate than Helex ($p<0.05$ at 6 months and final follow-up). The Premere had an intermediate residual shunt rate. Septal aneurysm also predicted residual shunting (RR=24.7, 95% CI: 8.2 to 74.4, $p<0.0001$).
CONCLUSIONS:
Percutaneous PFO closure is an efficacious progressive treatment but closure rates also depend on the presence of aneurysm and differ between occluders.

J Thorac Cardiovasc Surg. 2011 Mar;141(3):631-6. Epub 2011 Jan 13.

Intraoperative device closure of atrial septal defects with inferior vena cava rim deficiency: a safe alternative to surgical repair.

Chen Q, Chen LW, Cao H, Zhang GC, Chen DZ, Zhang H.
Source: Department of Cardiovascular Surgery, Union Hospital, Fujian Medical University, Fuzhou, People's Republic of China.

Abstract
OBJECTIVE:
Our objective was to evaluate the safety and feasibility of intraoperative device closure of atrial septal defects with inferior vena cava rim deficiency.
METHODS:
From January 2005 to December 2008, we enrolled 65 patients who had a secundum atrial septal defect with inferior vena cava rim deficiency closure in our institution. Patients were divided into 2 groups: 35 patients in group I underwent intraoperative device closure with a right lateral minithoracotomy and 30 in group II underwent open cardiac repair with a right lateral thoracotomy and cardiopulmonary bypass. Intraoperative device closure involved a minimal intercostal incision that was performed after full evaluation of the atrial septal defect by transthoracic echocardiography and the insertion of the device through the delivery sheath to occlude the atrial septal defect.
RESULTS:
The procedure was successful in all patients. In group I, the diameter of the atrial septal defect ranged from 30 to 44 mm (mean, 35.3 ± 3.9 mm), and the size of the implanted occluder ranged from 34 to 48 mm (mean, 40 ± 2.1 mm). The total occlusion rate was 82.9% immediately after the operation, 97.1% at 3 months, and 100% at 12 and 24 months of follow-up. In group II, all patients had successful closure. A follow-up period of 12 to 24 months was obtained in both groups. During the follow-up, there was no recurrence, thrombosis, or device failure. In our comparative studies, group II had significantly longer operative time, intensive care unit stay, and hospital stay than group I ($P < .001$). The cost of group I was less than that of group II (20,450.9 ± 840.8 RMB vs 25,884.9 ± 701.8; $P < .001$).
CONCLUSIONS:
Intraoperative device closure of atrial septal defects with inferior vena cava rim deficiency is a safe and feasible technique. It has the advantages of cost savings, cosmetic results, and less trauma than surgical closure. Early and midterm results are encouraging.

J Heart Lung Transplant. 2011 Apr;30(4):395-401. Epub 2010 Oct 29.

Would access to device therapies improve transplant outcomes for adults with congenital heart disease? Analysis of the United Network for Organ Sharing (UNOS).

Everitt MD, Donaldson AE, Stehlik J, Kaza AK, Budge D, Alharethi R, Bullock EA, Kfoury AG, Yetman AT.
Source: University of Utah, Salt Lake City, Utah; Utah Transplant Affiliated Hospitals (U.T.A.H.) Cardiac Transplant Program, Salt Lake City, Utah.

Abstract
BACKGROUND:
Patients with congenital heart disease (CHD) now survive into adulthood and often present with end-stage heart failure (HF). HF management and approach to orthotopic heart transplant (OHT) may differ from adults without CHD. We sought to compare OHT waitlist characteristics and outcomes for these 2 groups.
METHODS:
The Organ Procurement and Transplantation Network (OPTN)/United Network for Organ Sharing (UNOS) database was used to identify adults (≥18 years) listed for OHT from 2005 to 2009. The cohort was divided into those with or without CHD.
RESULTS:
Of 9,722 adults included, 314 (3%) had CHD. Adults with CHD were younger (35 ± 13 vs 52 ± 12 years, $p < 0.01$) and more often had undergone prior cardiac surgery (85% vs. 34%, $p < 0.01$). Patients with CHD were less likely to have a defibrillator (44% vs 75%, $p < 0.01$) or ventricular assist device (5% vs 14%, $p < 0.01$) and were more likely to be listed at the lowest urgency status than patients without CHD (64% vs 44%, $p < 0.01$). Fewer CHD patients achieved OHT (53% vs 65%, $p < 0.001$). Although overall waitlist mortality did not differ between groups (10% vs 8%, $p = 0.15$), patients with CHD were more likely to experience cardiovascular death (60% vs 40%, $p = 0.03$), including sudden in 44% and due to HF in 16%.
CONCLUSIONS:
Despite lower urgency status, patients with CHD have greater cardiovascular mortality

awaiting OHT than those without. Increased defibrillator use could improve survival to OHT, because sudden death is common. VAD support may benefit select patients, but experience in CHD is limited. Referral to specialized adult congenital heart centers can enhance utilization of device therapies and potentially improve waitlist outcomes.

J Thorac Cardiovasc Surg. 2011 Mar;141(3):637-44, 644.e1-3. Epub 2010 Sep 29.

Increased postoperative and respiratory complications in patients with congenital heart disease associated with heterotaxy.

Swisher M, Jonas R, Tian X, Lee ES, Lo CW, Leatherbury L.
Source: Laboratory of Developmental Biology, National Heart, Lung, and Blood Institute, National Institutes of Health, Bethesda, MD, USA.

Abstract

OBJECTIVE:
Patients with heterotaxy and complex congenital heart disease underwent cardiac surgery with high mortality and morbidity. Recent studies have revealed an association among heterotaxy, congenital heart disease, and primary ciliary dyskinesia. We undertook a retrospective review of patients undergoing cardiac surgery at Children's National Medical Center between 2004 and 2008 to explore the hypothesis that there is increased mortality and respiratory complications in heterotaxy patients.
METHODS:
Retrospective review was performed on postsurgical outcomes of 87 patients with heterotaxy and congenital heart disease exhibiting the full spectrum of situs abnormalities associated with heterotaxy. As controls patients, 634 cardiac surgical patients with congenital heart disease, but without laterality defects, were selected, and surgical complexities were similar with a median Risk Adjustment in Congenital Heart Surgery-1 score of 3.0 for both groups.
RESULTS:
We found the mean length of postoperative hospital stay (17 vs 11 days) and mechanical ventilation (11 vs 4 days) were significantly increased in the heterotaxy patients. Also elevated were rates of tracheostomies (6.9% vs 1.6%; odds ratio, 4.6), extracorporeal membrane oxygenation support (12.6% vs 4.9%: odds ratio, 2.8), prolonged ventilatory courses (23% vs 12.3%; odds ratio, 2.1) and postsurgical deaths (16.1% vs 4.7%; odds ratio, 3.9).
CONCLUSIONS:
Our findings show heterotaxy patients had more postsurgical events with increased postsurgical mortality and risk for respiratory complications as compared to control patients with similar Risk Adjustment in Congenital Heart Surgery-1 surgical complexity scores. We speculate that increased respiratory complications maybe due to ciliary dysfunction. Further studies are needed to explore the basis for the increased surgical risks for heterotaxy patients undergoing cardiac surgery.

Eur J Cardiothorac Surg. 2011 Apr;39(4):490-4. Epub 2010 Sep 16.

Risk stratification for adult congenital heart surgery.

van Gameren M, Putman LM, Takkenberg JJ, Bogers AJ.
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Abstract

OBJECTIVE:
At this moment, no risk stratification models are available for adult congenital cardiac surgery. This study aims to identify a suitable stratification tool for the adult congenital heart surgery population. Pediatric congenital cardiac surgery score models were therefore tested in an adult congenital population. In addition, an age component was added to these models and performance was compared with the original score systems.
METHODS:

The Risk Adjustment in Congenital Heart Surgery (RACHS-1), Basic Aristotle Score, Society of Thoracic Surgeons (STS)-European Association for Cardiothoracic Surgery (EACTS) Score and Comprehensive Aristotle Score were calculated for all adult patients who underwent congenital cardiac surgery between January 1990 and January 2007 in a single center (N=963). In addition, an age component was added to these models. Discrimination was then tested for all models with and without the age component.
RESULTS:
Application of the original pediatric risk scores resulted in c-statistics for 30-day mortality of 0.60, 0.60, 0.60, and 0.66 respectively. Combining these models with the age component resulted in significantly higher c-statistics of 0.69, 0.70, 0.69, and 0.76 respectively. Age as a sole predictor already resulted in a c-statistic of 0.67. Comparable results were found for 1-year mortality.
CONCLUSIONS:
The discriminatory power of the pediatric risk scores was suboptimal, but increased when adding age as a score component. The best performance was achieved by the combination of age and the Comprehensive Aristotle Score, for both 30-day and 1-year mortality.

Catheter Cardiovasc Interv. 2011 Mar 1;77(4):564-9. doi: 10.1002/ccd.22713.

Premere occlusion system for transcatheter patent foramen ovale closure: mid-term results of a single-center registry.

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Abstract
BACKGROUND:
Transcatheter closure of patent foramen ovale (PFO) with rigid devices may be problematic in patients with long channel PFO: alternative devices with asymmetrical opening and more physiological positioning may be preferable in such cases. We present the mid-term results of transcatheter closure of PFO with Premere Occlusion System, a device studied for this specific anatomy, in a single-center registry of adults with previous cerebral ischemia.
METHODS:
During a 53-months period (July 1, 2005 to December 1, 2009) 70 patients (48 females and 22 males, mean age 38 ± 6.7 years) with previous stroke were admitted in our center for transcatheter closure of PFO with Premere Occlusion System on the basis of absence of moderate or severe atrial septal aneurysm (ASA) on Transesophageal echocardiography and intracardiac echocardiography (< 3RL or 3LR ASA and length of PFO channel >10 mm).
RESULTS:
The procedure was successful in all of the patients with no peri-operative and in-hospital complications. Forty-six 20 mm and twenty-four 25 mm Premere devices were implanted. Rates of procedural success, predischarge occlusion, and complication were: 100%, 95.7% and 0%, respectively. On mean follow-up of 40 ± 10.9 months (range 6-54), the follow-up occlusion rate was 98.5%. During follow-up, no cases of permanent atrial fibrillation, aortic/atrial erosion, device thrombosis, or atrioventricular valve inferences were noted.
CONCLUSION:
The mid-term outcomes of our registry suggests that the Premere Occlusion System may be an excellent device for patients with long-channel PFO and absence of moderate/severe ASA, offering a physiological and anatomically respective closure of PFO also in patients with hypertrophic rims.

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Gender differences in angiotensin II and aldosterone secretion in patients with pressure overloaded systemic right ventricles are similar to those observed in

systemic arterial hypertension.

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Abstract
BACKGROUND:
There are very few and inconclusive data concerning the renin-angiotensin-aldosterone system activity in adults with systemic right ventricles, compared to classic heart failure patients. Therefore, we prospectively evaluated angiotensin II and aldosterone levels in a series of patients following Mustard or Senning procedures for complete transposition of the great arteries.
METHODS:
Forty-two patients (31 male and 11 female, mean age 20.8 ± 3.7 years), 18.2 ± 2.8 years following atrial switch procedures, were included in the analysis. All the patients underwent comprehensive echocardiographic examinations. Angiotensin II and aldosterone levels were measured with immunoradiometric assays.
RESULTS:
The mean angiotensin II level was 11.9 ± 9.4 pg/mL; 15 patients (35.7%) had angiotensin II levels exceeding the upper limit of normal values. There was a negative correlation between angiotensin II levels and treatment with angiotensin enzyme inhibitors (r = -0.33, P = 0.03). The mean aldosterone level was 217.7 ± 160.2 pg/mL; 26 patients (61.9%) had aldosterone levels exceeding the upper limit of normal values. Female patients had significantly higher aldosterone levels than male patients (321 ± 248 vs 180 ± 95 pg/mL, P = 0.01). A negative correlation between angiotensin II levels and fractional area change (r = -0.65, P=0.03), and a positive correlation between aldosterone levels and right ventricular end-diastolic area (r = 0.66, P = 0.03) were observed in female but not in male patients.
CONCLUSIONS:
Renin-angiotensin-aldosterone axis activation in patients with systemic right ventricles was similar to reported values in other studies of stable heart failure. The gender differences in aldosterone levels in patients with systemic right ventricles were similar to that associated with left ventricular remodeling in systemic arterial hypertension.

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Exercise intolerance in patients with congenitally corrected transposition of the great arteries relates to right ventricular filling pressures.

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Abstract
BACKGROUND:
Patients with congenitally corrected transposition of the great arteries (ccTGA) have significantly reduced exercise tolerance. Progressive right ventricular (RV) dysfunction with tricuspid regurgitation (TR) and other haemodynamic lesions are common among them. We hypothesised that interaction of these factors may result in increased systemic RV filling pressure, which in turn impact on exercise capacity.
METHODS:
Patients with ccTGA in functional class I or II, able to perform treadmill exercise and without resting cyanosis were enrolled. All patients underwent cardiopulmonary exercise testing and transthoracic echocardiographic examination. RV filling pressure was estimated using tissue Doppler imaging (TDI) techniques by measuring early annular diastolic velocity (Ea) and the ratio of the transtricuspid inflow to the early annular diastolic velocity (E/Ea).
RESULTS:

A total of 27 patients (mean age 41 years, 48% female) were assessed, the majority (63%) asymptomatic. Many patients had coexistent haemodynamic lesions including shunts, pulmonary stenosis, TR and systemic ventricular dysfunction. Average percentage predicted peak oxygen consumption, VE/VCO(2) slope and heart rate reserve were abnormal in this population. Patients with moderately/severely impaired exercise capacity (≤ 60% predicted peak VO(2)) had significantly higher E/Ea ratios compared to those with normal/mildly impaired exercise capacity (septal E/Ea = 17.1 ± 9.7 vs 8.8 ± 1.6 and lateral E/Ea = 11.5 ± 5.8 vs 6.6 ± 1.3, p = 0.007 and 0.01 respectively).

CONCLUSION:

Reduced exercise capacity is common in adults with ccTGA even among asymptomatic patients and relates to increased RV filling pressures assessed by TDI. This index could potentially be used to optimize therapy or prognosticate adverse events in ccTGA patients.