July, 2011

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In This Issue

International Society for Adult Congenital Heart Disease

ISACHD Newsletter

President's Message

by *Barbara J.M. Mulder*

Dear ISACHD members,

From June 19-22 the 21st annual course on Congenital Heart Disease in the Adult was held in Cincinnati organized by Gary Webb, Craig Broberg, Erwin Oechslin and David Sahn. It was the largest ACHD symposium ever! A huge facility of pediatric and adult cardiologists, surgeons and nurses provided a four-day program with a large variety of topics.

On Sunday the symposium started with an all-day ACHD Echo Symposium and an ACHD Nursing Symposium in the Cincinnati Netherland Plaza hotel. The main program started at Monday. There were two main meeting rooms running concurrently. One room featured case-based teaching, led by major ACHD teams from North America and Europe. Gary won a T-shirt from Amsterdam, but unfortunately it was undersized. In the second room clinical lectures, panel discussions, research competitions, and discussions of controversial issues were visited by a large and active audience. Monday evening, participants were spread all over Cincinnati; both the cruise on the Ohio river and the baseball match between the Red and the Yankees were quite exciting!

On Tuesday even four parallel sessions offered high quality talks and lectures.People had some difficulties with choosing between different interesting topics. Fortunately, the whole program was recorded and will be put on the Internet by September 2011. As a result, registrants will be able to experience the programs they were interested in and 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. At Tuesday evening all participants were invited for a reception and a gala diner. Live music filled the room and created a very special atmosphere. Gary held an inspiring speech and showed his enthusiasm about this unique meeting in Cincinnati!

Wednesday continued with parallel sessions and it was the last day of a wonderful meeting where all caregivers for adults with congenital heart disease could meet each other. The ISACHD executive had a lunch

meeting to discuss the progress of the newly installed working groups on Health Care, Education and Research. The chairs, Curt Daniels, Erwin Oechslin and Koichiro Niwa, respectively, will contact all volunteers for these working groups within due time. If you are interested in participating, please send an email to one of these chairs or to b.j.mulder@amc.uva.nl, president of ISACHD.

Please don't forget to renew your membership now and go to www.isachd.org.
It has become simple and easy!

Barbara J.M. Mulder
President

WHF: ISACHD Joins the World Heart Federation (WHF)

The International Society of Adult Congenital Heart Disease (ISACHD) has been accepted as an Associate International Member of the World Heart Federation. The WHF Organization is looking forward to working with the ISACHD Members on CVD. The mission of WHF is to unite their members and lead the global fight against heart disease and stroke, with a focus on low and middle-income contries. For more information and member benefits for the World Heart Federation, please visit:
http://www.world-heart-federation.org

Case Report: An elite athlete with persistent atrial firbrillation

Monica Benjamin, Marcelo University, Jose Luis Serra, Luis Alday Section of Adult Congenital Heart Disease and Cardiovascular Unit, Sanatorio Allende, Crodoba Argentina.

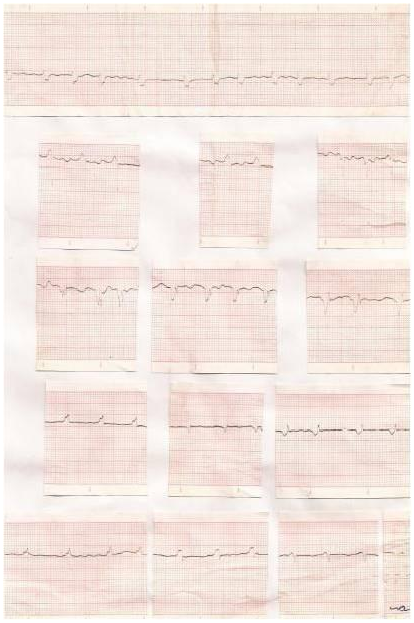

Figure 1

A 60 year-old man, asymptomatic until 5 years ago, was referred for frequent episodes of paroxysmal atrial fibrillation (AF). At the age of 55 with rapid ventricular response. Aside from arrhythmia, the EKG showed left axis deviation and right bundle branch block. (Figure 1) A chest x-ray revealed moderate cardiomegaly with a CT ratio of 60%, a prominent pulmonary artery and increased pulmonary vascular markings. (Figure 2) A color Doppler TTE study concluded that the left atrium and right side chambers were enlarged while the left ventricular size and function were normal. Mild tricuspid regurgitation was present. There was no mention of any other abnormalities.

2008-2010

The initial treatment strategy was rate control with digoxin and oral anticoagulation. He had good response and a few days later he was discharged with heart failure treatment including enalapril, carvedilol, furosemide, and spironolactone with the diagnosis of persistent AF.
He had been an elite athlete in his youth with no symptoms whatsoever. Several medical examinations through his long career failed to demonstrate any abnormality.

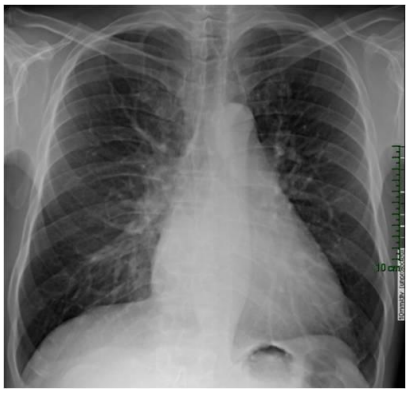

Figure 2: Chest x-ray on Admission

Following discharge, he did well on digoxin and oral anticoagulation. Four weeks later, digoxin was discontenued and hew as started on amidaron. Sinus rhythm was restored with DC cardioversion with no AF recurrences for almost three years.

Following this period of time, he began having frequent AF recurrences that required admissions for congestive heart failure.On his last admission, he had AF with a ventricular rate of 100 bpm and a blood pressure of 100/68 mmHg. The heart sounds were distant and there was a widely split 2nd heart sound and a mild (2/6) ejection systolic murmur along the upper left sternal border. A new TTE was performed which besides the findings shown in the initial study, disclosed a moderate size (13 mm) secundum atrial septal defect (ASD) which was suitable for device closure. The estimated pulmonary artery systolic pressure was 30 mmHg. A TEE confirmed the TTE findings.

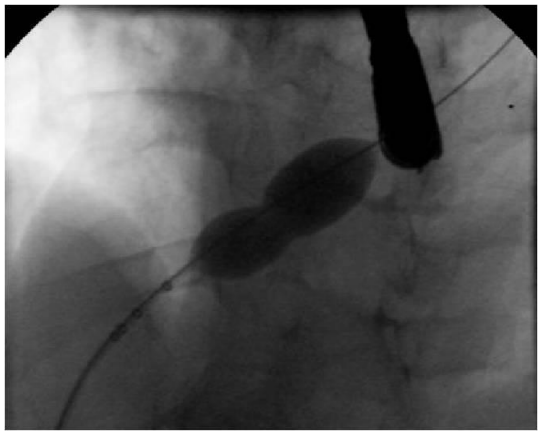

Figure 3: Balloon sizing confirming ASD diameter

Cardiac catheterization was performed for device closure of the defect which was occluded uneventfully. (Figure 3) Four weeks later, he underwent successful electrical cardioversion and heart failure treatment was continued. (Figure 4) The patent remained symptom free avoiding strenuous exercise.

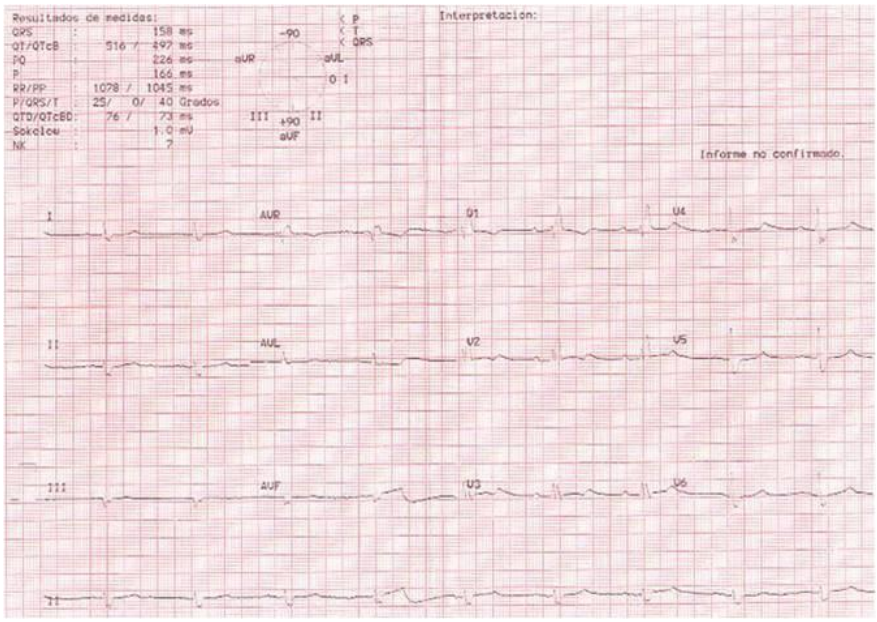

Figure 4: EKG following cardioversion

Comment

ASD is the most common adult congenital heart disease with a prevalence of 30% among all cardiac anomalies presenting at this age. Patients with small to moderate defects may have no symptoms until the 4th or 5th decade of life that along with subtle clinical findings often lead to delayed diagnosis. (1-3) Systemic hypertension, coronary heart disease, acquired valvular heart disease, or just aging, may reduce the left ventricular compliance increasing the left to right shunt across the defect leading to pulmonary hypertension and congestive heart failure. Mitral regurgitation is frequently present since untreated ASD patients often have mitral valve prolapse. A volume overloaded left atrium is an ideal myocardial substrate for atrial arrhythmias. (2-4) Our patient was an elite athlete with an overstanding career, a winner of the world championship in his discipline. He never had symptoms until after retirement. When he developed symptoms related to AF, the initial diagnostic studies failed to demonstrate the ASD. However, retrospectively, the EKG and x-ray were compatible with an ASD and a more thorough echocardiographic study made the diagnosis of a moderate ASD. The defect was closed with a percutaneous device and normal sinus rhythm was restored though we do not know for how long.

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Regional and Nursing News:

News from Latin America
by Luis Alday

The XXXVII and XIV Argentine Congresses of Cardiology and Pediatric Cardiology respectively will take place in Buenos Aires at the Buenos Aires Sheraton Hotel from Sunday 2 till Tuesday 4 October 2011. An International Society for Adult Congenital Heart Disease (ISACHD) and Argentine Society of Cardiology (SAC) Joint Session is being planned with the participation of Drs Roberta Williams (UCLA, Los Angeles, US) and Andrew Reddington (Hospital for Sick Children, Toronto, Canada). These sessions were inaugurated in 2009 with a great success and attendance by doctors from Argentina and neighboring countries.

News from Europe
by Helmut Baumgartner

The yearly European congress of Cardiology will be held in Paris, France,from August 27th to August 31st. Joint session ESC/ISACHD August 2011, ESC congress Paris. At Sunday afternoon, there will be a joint session ESC/ISACHD:

Title: Controversies in Adult Congenital Heart Disease
Chair: BJM Mulder, GD Webb
Is identification of early RV dysfunction possible, by P. Beerbaum, London
Can we help the failing RV with drugs? By N.M. Ammash, Rochester
Implantable defibrillatorin ACHD: who benefits? By F. Cecchin, Boston
Optimal pacing sites in CHD, by T Delhaas, Maastricht

Monday evening ISACHD will have a joint social event with the ESC GUCH Working group.

Nursing Network
by Desiree Fleck

We had 60 attendees at the nursing symposium and had a wonderful session. We were also together for a breakfast and reception. New members were identified for the nursing network with representatives from Denmark, Ireland, Canada and the US. The nursing network is reorganizing. There were 3 abstract winners from the nursing section who presented their projects at the conference. Catriona King from Copenhagen, Denmark, won the abstract competition. she presented her abstract on Single Ventricle. Marty Tomlin, CRNP from Cincinnati Children's Hospital came in second and presented on the Adults satisfaction being seen in a pediatric institution. finally, Mia from Copenhagen Denmark presented her abstract on The Cognitive status in adults with Eisenmenger's Syndrome.

There is an ACHD Education Day being sponsored by the Children's Hospital of Philadelphia geared to the inpatient nurses on September 12, 2011.

Article of the Month:

J Am Coll Cardiol. 2011 Jun 7:57(23): 2368-74

Transfer of adolescents with congenital heart disease from pediatric cardiology to adult health care an analysis of transfer destinations.

Goossens E, Stephani I, Hilderson D, Gewillig M, Budts W, Van Deyk K, Moons

P; SWITCH(2) Investigators.

Source Center for Health Services and Nursing Research, Katholieke Universiteit Leuven, Leuven, Belgium.

Abstract

OBJECTIVES: The transfer of adolescents with **congenital heart** disease from pediatric to **adult** care was examined. The aims were to investigate where these adolescents received **adult**-centered care to determine the proportion of patients with no follow-up and with no appropriate follow-up after leaving pediatric cardiology, and to explore the determinants of no follow-up and no appropriate follow-up.

BACKGROUND: Even after successful treatment, many patients require lifelong cardiac surveillance by specialized practitioners. Although guidelines describe the most appropriate level of follow-up, this is not always implemented in practice.

METHODS:

A descriptive, observational study was performed, including 794 patients with **congenital heart** disease examined and/or treated at a tertiary care center.

RESULTS:

Overall, 58 of the 794 patients included (7.3%) were not in follow-up. Cessation of follow-up was found in 2 of 74 patients with complex (2.7%), 31 of 448 patients with moderate (6.9%), and 25 of 272 patients with simple (9.2%) heart defects. Moreover, 684 patients (86.1%) remained in specialized follow-up. According to international guidelines, 81 patients (10.2%) did not receive the minimal level of cardiac care. Multivariable logistic regression revealed that male sex and no prior heart surgery were associated with no follow-up. Male sex, no prior heart surgery, and greater complexity of **congenital heart** disease were associated with no appropriate level of cardiac follow-up.

CONCLUSIONS: The proportion of patients in this study lost to follow-up was substantially lower than in other Western countries. Because only patient-related factors were examined with respect to loss to follow-up, further examination of patient-related, hospital-related, and healthcare-related determinants of lack of follow-up is needed.

Commentary on the paper 'Transfer of adolescents with congenital heart disease from pediatric cardiology to adult health care' by Yumi SHINA

Yumi Shina

Owing to advances in surgical and medical management, most patients with congenital heart disease (CHD), event complex CHD, can be expected to reach adulthood [1]. There have been several excellent reports about the estimated number of adults with CHD in Europe, US and Asia [2-4] and the prevalence of adults with CHD is 3.92-4.09 per 1000 adults. Various studies revealed 21-76% of patients with CHD failed to receive regular follow-up in adulthood [5][6]. These data are not comparable, because the studies differed substantially in definition of 'loss of follow-up' and, in most studies, patients were recruited at ACHD

clinic.

The aims of this study were 1)to investigate where these adolescents received adult-centered care, 2)to determine the proportion of patients with no follow-up and with no appropriate follow-up after leaving pediatric cardiology, and 3)to explore the determinants of no follow-up and no appropriate follow-up. A descriptive, observational study was performed, including 794 patients with CHD examined and/or treated at a tertiary care center in Leuven, Belgium. Overall, 58 of the 794 patients included (7.3%) were not in follow-up. Cessation of follow-up was found in 2 of 74 patients with complex (2.7%), 31 of 448 patients with moderate (6.9%), and 25 of 272 patients with simple (9.2%) heart defects. Moreover, 684 patients (86.1%) remained in specialized follow-up. In multivariable logistic regression, 1)male sex, 2) no prior heart surgery, and 3) greater complexity of congenital heart disease were associated with no appropriate levels of cardiac follow-up.

The proportion of patients lost to follow-up in this study was lower than in other Western countries. Those of the male sex, with no prior heart surgery, and with greater complexity of congenital heart disease have a tendency to attend inappropriate care centers and, obviously, some of them are in the high risk group suffering from infective endocarditis. They may have no information on prophylactic antibiotics. Further examinations of the detailed reasons (number of tertiary care centers, the age and the way of transfer, and social status etc) for failing to receive appropriate follow-up are required. We should especially focus on the potential high risk patients who drop out of regular follow-up when they transfer from the pediatric department.

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Article: *Goossens E, Stephani I, Hilderson D, Gewillig M, Budts W, Van Deyk K, Moons P; SWITCH² Investigators. Transfer of adolescents with congenital heart disease from pediatric cardiology to adult health care - an analysis of transfer destinations. J Am Coll Cardiol. 2011 Jun 7;57(23):2368-74.*

Commentary: *Yumi SHIINA, M.D. PhD*
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Journal Watch

Int J Cardiol. 2011 Jun 30. [Epub ahead of print]

Prolonged beneficial effect of bosentan treatment and 4-year survival rates in adult patients with pulmonary arterial hypertension associated with congenital heart disease.

Vis JC, Duffels MG, Mulder P, de Bruin-Bon RH, Bouma BJ, Berger RM, Hoendermis ES, van Dijk AP, Mulder BJ.

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

Abstract
Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) due to systemic to pulmonary shunting is associated with a high risk of morbidity and mortality. In this study we evaluated 4years treatment effect of bosentan on exercise capacity and quality of life and survival rates in 64 adult patients with PAH associated with CHD, including patients with Down syndrome (DS). All patients were evaluated at baseline and during follow-up with laboratory tests, 6-minute walk test, quality of life questionnaires, and Doppler echocardiography. In total, 13 patients (20%) died during 4-years of follow-up; 4 patients with DS and 9 patients without DS. Mean follow-up of all patients treated with bosentan was 3.5±1.2year. We analyzed treatment efficacy separately within patients without DS (n=34) and patients with DS (n=30). Mean 6-minute walking distance (6MWD) in patients without DS significantly increased at 6months from 417±108 to 458±104m (+41m; p=0.002) and significant improvement continued to exist during at least 2.5years of follow-up (p=0.003). Moreover, stroke volume increased significantly (p=0.02). In the patients with DS, 6-MWD, stroke volume and quality of life remained stable during treatment. In this study we demonstrate a prolonged beneficial effect of bosentan treatment on exercise capacity, stroke volume and quality of life in patients without DS. However the mortality rate of 20% of patients after 4years of follow-up remains high.

Anadolu Kardiyol Derg. 2011 Jun 28. doi: 10.5152/akd.2011.112. [Epub ahead of print]

Cheatham-Platinum stent for native and recurrent aortic coarctation in children and adults: immediate and early follow-up results.

Erdem A, Akdeniz C Saritas T, Erol N, Demir F, Karaci AR, Yalcin Y, Celebi A.

Source
Department of Pediatric Cardiology, Dr. Siyami Ersek, Thoracic and Cardiovascular Surgery Center, Istanbul-Turkey. drabdullaherdem@hotmail.com.

Abstract
OBJECTIVE:
To present our institutional experience of endovascular Cheatham-Platinum stent implantation in children and adults with native and recurrent aortic coarctation.

METHODS:
Between August 2007 and November 2009, 45 patients had aortic coarctation treated with 47 stents implantation. We preferred primarily stent implantation in adult patient with coarctation, in children more than five years-old it is preferred in cases of aneurysm, subatretic or blind coarctation and coarctation with patent ductus arteriosus or in

restenosis. Files of stent-implanted patients were retrospectively analyzed in terms of patients' demographic features, echocardiographic and angiographic findings both before and after procedure. Patients grouped as Group 1: native coarctation and Group 2: recoarctation developed after either surgery or balloon angioplasty. Findings of the cases' were compared using paired and unpaired Student's t, Mann-Whitney U and Chi-square tests.

RESULTS:
Sixteen covered and 31 bare totally 47 balloon expandable stents were implanted in 45 patients. The mean follow up duration was 12.1±7.1, median 11 months (2-29 months). There was no procedure related death. In two patients two stents were implanted in tandem. While the coarctation of the aorta was native in 26 patients (functionally interrupted aortic arch in one), recoarctation was detected in 7 patients after surgery, in 8 patients after balloon angioplasty, in 4 patients both after surgery and balloon angioplasty. One patient had functionally interrupted aortic arch perforated with guide wire and then covered stent implanted. The mean age 12.2±5.9 years (5-33 years) and mean body mass index was 21±3.7 kg/m2 (14.8-31 kg/m2). Considering all cases, a statistically significant decrease in both the invasive and echocardiographic gradients ($p<0.001$ for both) and statistically significant increase in lesion diameter ($p<0.001$) were detected. The decrease in invasive and echocardiographic gradients and increase in lesion diameter is statistically significant in each group also ($p<0.001$, <0.001 and <0.001 for both groups, respectively). Before the procedure, the invasive gradient was significantly higher and the lesion diameter was significantly lower in group I than in group II ($p=0.002$ and $p=0.005$, respectively). Also the percentage of decrease in gradient and increase in diameter was statistically higher in group 1 than in group 2 ($p=0.04$ and $p=0.04$).

CONCLUSION:
Our early and short-term follow-up results indicate that stent implantation is safe and very effective in reducing coarctation gradient and increasing lesion diameter both in native coarctation and recoarctation.

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

A guide for identification and continuing care of adult congenital heart disease patients in primary care.

Ellison S, Lamb J, Haines A, O'Dell S, Thomas G, Sethi S, Ratcliffe J, Chisholm S, Vaughan J, Mahadevan VS.

Source
Cheshire and Wirral Partnership NHS Foundation Trust, United Kingdom.

Abstract
BACKGROUND:
Surgical and other advances in the treatment and care of congenital heart disease have resulted in a significant increase in the number of adults with congenital heart disease (ACHD), many of whom have no regular cardiology follow-up. Optimised care for ACHD patients requires continuity of specialist and shared care and education of practitioners and patients. The challenges for managing ACHD were identified by a Health Needs Assessment in the North West and are addressed within the UK Department of Health's ACHD Commissioning Guide.

MATERIALS AND METHODS:
An ACHD model of care was recommended in the North West of England and developed by the three North West Cardiac & Stroke Networks. Within this, a Task Group focused on the role of primary care in the identification and continuing care of ACHD patients. A feasibility study demonstrated that existing diagnostic Read Codes can identify ACHD patients on general practice registers. An ACHD Toolkit was developed to provide algorithms to guide the appropriate management of ACHD patients through primary, secondary and/or specialist ACHD care and to improve education/knowledge amongst primary care staff about ACHD and its wider implications.

RESULTS:
Early findings during the development of this Toolkit illustrate a wide disparity of provision between current and optimal management strategies. Patients lost to follow-up have already been identified and their management modified.

CONCLUSIONS:
By focusing on identifying ACHD patients in primary care and organising/delivering ACHD services, the ACHD Toolkit could help improve quality, timeliness of care, patient experience and wellbeing.

Expert Rev Respir Med. 2011 Jun;5(3):363-76

Atrial septostomy in patients with pulmonary hypertension: should it be recommended?

Lammers AE, Haworth SG, Diller GP.

Source
Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hosptial, Sydney Street, London SW3 6NP, UK.

Abstract
Treatment options for patients with advanced pulmonary hypertension (PH) are limited. Iatrogenic creation of an interatrial communication (i.e., atrial septostomy [AS]) has been suggested as a possible treatment option or as a bridge to transplantation in selected patients and has been incorporated into current PH guidelines. Uptake of the procedure has been slow and the worldwide experience with AS is limited to approximately 280 published cases, over a period of more than 25 years. The rationale for creating an AS has been provided by the observation that patients with congenital heart disease, shunt lesions and PH have a better survival compared with patients with idiopathic PH. We review pathophysiologic data and the published clinical experience and discuss the rationale, indication and potential pitfalls of AS in patients with severe PH.

Circ Cardiovasc Qual Outcomes. 2011 Jun 21. [Epub ahead of print]

Risk Factors for Death After Adult Congenital Heart Surgery in Pediatric Hospitals.

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

Source
Divisions of Cardiology, Hospital of the University of Pennsylvania and Children's Hospital of Philadelphia, University of Pennsylvania School of Medicine, Philadelphia, PA; the Department of Cardiology, Children's Hospital Boston, Harvard Medical School, Boston, MA; Boston Adult Congenital Heart
Program, the Department of Cardiology, Children's Hospital Boston, Boston, MA; the Division of Cardiology, Brigham and Women's Hospital, Harvard Medical School, Boston, MA; and the Department of Surgery, Morgan Stanley Children's Hospital of New York, Columbia University College of Physicians and Surgeons, New York, NY.

Abstract
Background Despite the central role that pediatric hospitals play in the surgical treatment of congenital heart disease, little is known about outcomes of adult congenital cardiac surgical care in pediatric hospitals. Risk factors for inpatient death, including adult congenital heart (ACH) surgery volume, are poorly described. Methods and Results We obtained inpatient data from 42 free-standing pediatric hospitals using the Pediatric Health Information System data base 2000 to 2008 and selected ACH surgery admissions (ages 18 to 49 years). We examined admission characteristics and hospital surgery volume. Of 97 563 total (pediatric and adult) congenital heart surgery admissions, 3061 (3.1%) were ACH surgery admissions. Median adult age was 22 years and 39% were between ages 25 to 49 years. Most frequent surgical procedures were pulmonary valve replacement,

secundum atrial septal defect repair, and aortic valve replacement. Adult mortality rate was 2.2% at discharge. Multivariable analyses identified the following risk factors for death: age 25 to 34 years (adjusted odds ratio [AOR], 2.1; P=0.009), age 35 to 49 years (AOR, 3.2; P=0.001), male sex (AOR, 1.8; P=0.04), government-sponsored insurance (AOR, 1.8; P=0.03), and higher surgical risk categories 4+ (AOR, 21.5; P=0.001). After adjusting for case mix, pediatric hospitals with high ACH surgery volume had reduced odds for death (AOR, 0.4; P=0.003). There was no relationship between total congenital heart surgery volume and ACH inpatient mortality. Conclusions Older adults, male sex, government-sponsored insurance, and greater surgical case complexity have the highest likelihood of in-hospital death when adult congenital surgery is performed in free-standing pediatric hospitals. After risk-adjustment, pediatric hospitals with high ACH surgery volume have the lowest inpatient mortality.

Am J Cardiol. 2011 Jun 17 [Epub ahead of print]

Single-Center Experience with Implantable Cardioverter-Defibrillators in Adults With Complex Congenital Heart Disease.

Khanna AD, Warnes CA, Phillips SD, Lin G, Brady PA.

Source
Division of Cardiovascular Diseases, Mayo Clinic, Rochester, Minnesota.

Abstract
Adults with congenital heart disease are at risk of lethal ventricular arrhythmias and are candidates for implantable cardiac defibrillator (ICD) therapy, yet implant risks, long-term outcomes, and rates of appropriate and inappropriate ICD therapies are not well characterized. We reviewed clinical, implantation, and follow-up data on all transvenous ICDs in adults with congenital heart disease at the Mayo Clinic from 1991 through 2008. Seventy-three adults with congenital heart disease received 85 ICDs. Implantation diagnoses included tetralogy of Fallot (44%) and congenitally corrected transposition of the great arteries (17%). Implantation indication was occurrence of sustained ventricular arrhythmias (secondary prevention) in 36% and prophylactic (primary prevention) in the remainder. There were no major implant-related complications. During follow-up (2.2 ± 2.8 years, range 0 to 15) 11 patients died and 4 patients received heart or heart/lung transplants. An appropriate shock for a ventricular arrhythmia was observed in 19% of patients and an inappropriate shock was observed in 15% of patients. Likelihood of an appropriate shock was associated with increased subpulmonic ventricular pressure. In conclusion, implantation of transvenous ICDs in adults with congenital heart disease is associated with a low risk of implant complications. In this high-risk adult population the rate of inappropriate ICD shocks is low, whereas the likelihood of appropriate therapy for potentially lethal ventricular arrhythmias is high. These data suggest overall benefit of ICD therapy in adults with congenital heart disease.

Am J Cardiol. 2011 Jun 17. [Epub ahead of print]

Identifying High Risk in Adults With Congenital Heart Disease and Atrial Arrhythmias.

Yap SC, Harris L, Chauhan VS, Oechslin EN, Silversides CK.

Source
Department of Cardiology, Erasmus Medical Center, Rotterdam, the Netherlands.

Abstract
Atrial arrhythmias are associated with an increased mortality risk in adults with congenital heart disease (CHD). However, little is known about risk stratification in the specific group of adult patients with CHD and atrial arrhythmias. We sought to identify predictors of mortality in adult with CHD and atrial arrhythmias and to establish a risk score. The study involved 378 adult patients with CHD (mean age 39 ± 13 years) and atrial arrhythmias who had serial follow-up in a tertiary referral center from 1999 through 2009. During a median

follow-up of 5.2 years, there were 40 deaths (11%). Overall mortality rate was 2.0% per patient-year. Common modes of death included heart failure-related death (35%), sudden cardiac death (20%), and perioperative death (18%). Independent predictors of mortality were poor functional class (hazard ratio 3.69, 95% confidence interval [CI] 1.69 to 8.03, p = 0.001), single-ventricle physiology (hazard ratio 3.33, 95% CI 1.51 to 7.35, p = 0.003), pulmonary hypertension (hazard ratio 2.96, 95% CI 1.41 to 6.19, p = 0.004), and valvular heart disease (hazard ratio 2.73, 95% CI 1.33 to 5.59, p = 0.006). A risk score was constructed using these predictors in which patients were assigned 1 point for the presence of each risk factor. Mortality rates in the low-risk (no risk factor), moderate-risk (1 risk factor), and high-risk (>1 risk factor) groups were 0.5%, 1.9%, and 6.5% per patient-year, respectively (log-rank p <0.001). In conclusion, in adult with CHD and atrial arrhythmias specific clinical variables identify patients at high risk for death. Importantly, the absence of any of these risk factors is associated with an excellent survival despite the presence of atrial arrhythmias.

Heart Surg Forum. 2011 Jun 1:14(3):E202-6

Natural internal banding in adult patients with large ventricular septal defect and a preserved pulmonary vascular system.

Inan BK, Ucak A, Temizkan V, Guler A, Ak K, Ugur M, Alp I, Arslan G, Yilmaz AT.

Source
Gülhane Military Medical Academy Haydarpasa Teaching Hospital, Cardiovascular Surgery Clinic, Istanbul, Turkey.

Abstract
Objective: Hypertrophied anomalous muscle bands (AMBs) in the right ventricular outflow tract (RVOT) may develop in the context of ventricular septal defects (VSDs) and limit persistent pulmonary overflow. In adult patients with a large VSD, persistent AMBs in the RVOT therefore can simulate the role of an externally placed pulmonary artery band. We termed such alterations natural internal bands (NIBs). Our goal was to establish the morphologic nature of the obstructive muscular lesions of the RVOT in patients with a large VSD.Methods: Patients who underwent operations for a large VSD in our center, which has a high volume of adult patients with congenital defects, were retrospectively reviewed, and the nature of the NIBs in these patients was documented. All patients underwent transthoracic echocardiography and cardiac catheterization evaluations preoperatively and at postoperative month 3. Histopathologic examination of the AMBs was performed.Results: Of 96 adult patients who underwent operations for a large isolated VSD (mean defect size, 16.9 ± 3.5 mm), 16 patients had a hemodynamically significant NIB. Two different patterns of obstruction were found. Ten of the 16 patients revealed an os infundibulum morphology, and 6 patients revealed systolic bulging of the conal septum. Four of the patients with os infundibulum also had classic tetralogy-type septal malalignment. The mean peak systolic gradient on the RVOT was 56.5 ± 17.2 mm Hg and 53.6 ± 12.3 mm Hg in the patients with os infundibulum and in the patients with systolic bulging of the conal septum, respectively. Surgical repair of the VSD was completed successfully in all patients. Resection of the os infundibulum was performed concomitantly in patients with os infundibulum. At the third postoperative month, the mean peak systolic gradient was 16.8 ± 3.5 mm Hg in patients with os infundibulum and 26 ± 5.9 mm Hg (range, 20-35 mm Hg) in patients with systolic septal bulging.Conclusions: Some mechanisms in adult type VSDs are essential for protecting the pulmonary vasculature. We tried to review these protective mechanisms: hypertrophied AMBs and NIBs.

Am J Med Genet A. 2011 Jul:155(7):1661-7. doi: 10.1002/ajmg.a.34068. Epub 2011 Jun 10.

Adults with congenital heart disease: Patients' knowledge and concerns about inheritance.

van Engelen K, Baars MJ, van Rongen LT, van der Velde ET, Mulder BJ, Smets EM.

Source
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Abstract
With recent advances in medical and surgical management, most patients with congenital heart disease (CHD) survive to reproductive age. Current guidelines recommend counseling about inheritance and transmission of CHD to offspring. We evaluated whether adult CHD patients recalled having received information about the inheritance of their CHD, patients' knowledge about inheritance and their concerns in this regard. A questionnaire was sent to 486 non-syndromic CHD patients aged 20-45 years. We received 332 useful questionnaires (response rate 68%). One-third (33%) of patients recalled receiving information about inheritance of CHD from their cardiologist, and 13% had consulted a clinical geneticist. Eight percent of patients who were considering having children estimated the recurrence risk for their own offspring to be 1% or lower, whereas one-fourth (25%) estimated it to be higher than 10%. According to our classification, 44% estimated the recurrence risk in a correct range of magnitude. Additional information about inheritance of CHD was desired by 41% of patients. Forty-two percent of patients considering having children reported concerns about transmitting CHD to offspring. We conclude that a substantial proportion of adult CHD patients lacks knowledge and desires more information about inheritance, indicating a need for better patient education. Current guidelines and/or their implementation do not seem to meet the needs of these patients. A dedicated program of counseling for adults with CHD has to be developed to optimize knowledge and satisfaction with information provision and to reduce or manage concerns regarding inheritance of CHD. © 2011 Wiley-Liss, Inc.

Curr Probl Cardiol. 2011 Jun:36(6):228-55

Magnetic resonance imaging images in adult congenital heart disease.

Broberg CS, Meadows A, Sahn D.

Abstract
The use of cardiac magnetic resonance imaging has become an indispensable tool for the evaluation of patients with congenital heart disease. With the emergence of several generations of congenital heart disease survivors, there are now as many adults with these conditions as children, and complications are the rule rather than the exception. It is increasingly important, therefore, that the general cardiology community becomes aware of these defects and potential pitfalls to avoid. Among its many uses, cardiac magnetic resonance can provide an assessment of right ventricular function, flow, pulmonary artery anatomy, and aortic visualization that are often important considerations in these patients. This review provides an introductory visual glimpse into the varied conditions encountered and clinical questions addressed in the field of adult congenital heart disease.

J Forensic Sci. 2011 Jun 3. doi: 10.111/j.1556-4029.2011.01807.x. [Epub ahead of print]

"Grown-Up" Congenital Heart Disease and Sudden Death in a Medical Examiner's Population*

Hamilton LE, Lew EO, Matshes EW.

Source
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Abstract
Despite advances in the management of congenital heart disease (CHD), children with

CHD who survive into adulthood are at increased risk of sudden death. Sudden death may also be the initial presentation of undiagnosed CHD in some adults. This retrospective descriptive study outlines the spectrum of CHD presenting as sudden death in adults in a medical examiner's population. Despite its rarity (0.2% of all cases investigated between 1991 and 2007), CHD remains an important cause of sudden cardiac death to be recognized at adult autopsy. Bicuspid aortic valve and anomalous coronary anomalies were the most common malformations, comprising 36.9% and 26.2% of cases, respectively. However, a wide spectrum of simple to complex malformations can be seen, with or without prior surgery, and over a wide age spectrum. Once solely a pediatric entity, CHD is now "grown-up" and will likely be diagnosed by forensic pathologists with increased frequency in the future.

Int J Cardiovasc Imaging. 2011 Jun 3. [Epub ahead of print]

Right ventricular function declines after cardiac surgery in adult patients with congenital heart disease.

Schuuring MJ, Bolmers PP, Mulder BJ, de Bruin-Bon RA, Koolbergen DR, Hazekamp MG, Lagrand WK, De Hert SG, de Beaumont EM, Bouma BJ.

Source
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Abstract
Right ventricular function (RVF) is often selectively declined after coronary artery bypass graft surgery. In adult patients with congenital heart disease (CHD) the incidence and persistence of declined RVF after cardiac surgery is unknown. The current study aimed to describe RVF after cardiac surgery in these patients. Adult CHD patients operated between January 2008 and December 2009 in the Academic Medical Centre in Amsterdam were studied. Clinical characteristics, laboratory tests, surgical data and intensive care unit outcome were obtained from medical records. RVF was measured by trans-thoracic echocardiography (TTE) and expressed by tricuspid annular plane systolic excursion (TAPSE), tissue Doppler imaging (RV S') and myocardial performance index (MPI) pre-operatively and direct, at intermediate and late follow up. Of a total of 185 operated, 86 patients (mean age 39 ± 13 years, 54% male) had echo data available. There was a significant fall in RVF after cardiac surgery. TAPSE and RV S' were significantly higher and MPI was significantly lower pre-operatively compared to direct post-operative values (TAPSE 22 ± 5 versus 13 ± 3 mm ($P < 0.01$), RV S' 11 ± 4 versus 8 ± 2 cm/s ($P < 0.01$) and MPI 0.36 ± 0.14 vs 0.62 ± 0.25; $P < 0.01$). There were no significant differences in left ventricular function pre-operatively compared to post-operative values. Right-sided surgery was performed in 33, left-sided surgery in 37 and both sided surgery in 16 patients. Decline in RVF was equal for those groups. Patients with severe decline in RVF, were patients who underwent tricuspid valve surgery. Decline in RVF was associated with post-operative myocardial creatine kinase level and maximal troponin T level. There was no association between decline in RVF and clinical outcome on the intensive care unit. 18 months post-operatively, most RVF parameters had recovered to pre-operative values, but TAPSE which remained still lower ($P < 0.01$). CHD patients have a decline in RVF directly after cardiac surgery, regardless the side of surgery. Although a gradual improvement was observed, complete recovery was not seen 18 months post-operatively.

J Magn Reson Imaging. 2011 Jun33(6):1341-50. doi: 10.1002/jmri.22573.

Cardiovascular MR dobutamine stress in adult tetralogy of fallot: Disparity between CMR volumetry and flow for cardiovascular function.

Valverde I, Parish V, Tzifa A, Head C, Sarikouch S, Greil G, Schaeffter T, Razavi R, Beerbaum P.

Source

Division of Imaging Sciences, King's College London, United Kingdom; Evelina Children Hospital, Guy's & St Thomas' NHS Foundation Trust, Department of Congenital Heart Disease, London, United Kingdom.

Abstract
PURPOSE:
To evaluate the MR agreement of cardiac function parameters between volumetric (cine SSFP) and phase contrast flow (PC-flow) assessment in patients with repaired tetralogy of Fallot (r-TOF) and chronic pulmonary regurgitation (PR) at rest and under dobutamine stress (DS-MR).

RESULTS:
We found excellent Bland-Altman agreement (mean difference ± limits of agreement, mL/beat/m(2)) at rest for both the systemic (-0.8 ± 5.7) and pulmonary strokes volumes (-0.1 ± 7.6), which slightly deteriorates during DS-MR. The PR volume showed acceptable agreement at rest (-3.6 ± 15.1), but also further deteriorated during stress (5.4 ± 24). In contrast, the PR fraction showed poor agreement equally at rest (-5.6 ± 22.8) and DS-MR (3.2 ± 19.2).

CONCLUSION:
In r-TOF with chronic PR, analogous functional parameters should not be used interchangeably between volumetric and PC-flow assessment during DS-MR evaluation. J. Magn. Reson. Imaging 2011;33:1341-1350. © 2011 Wiley-Liss, Inc.

Interact Cardiovasc Thorac Surg. 2011 Jun:13(1):98-100. Epub 2011 Apr 27.

Emergency surgical intervention after unsuccessful percutaneous transluminal angioplasty and stenting of aortic coarctation.

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Source
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Abstract
Coarctation of thoracic aorta is an uncommon diagnosis in adults. Catheter-based intervention consisting of primary ballooning and stenting is becoming one of the methods of choice for the treatment of native coarctation. We describe the case of a young adult with coarctation of the aorta treated unsuccessfully with percutaneous transluminal angioplasty and stent implantation that resulted in stent migration into the aortic arch and led to an urgent operative intervention. In one step, we performed the evacuation of the foreign body from the aortic arch as well as the treatment of the aortic coarctation through an extra-anatomical vascular graft interposition between the ascending and descending thoracic aorta. In this article, we discuss the need for emergency surgical intervention in this case.

Interact Cardiovasc Thorac Surg. 2011 Jun:12(6):1033-9. Epub 2011 Mar 11.

In adult patients undergoing redo surgery for left atrioventricular valve regurgitation after atrioventricular septal defect correction, is replacement superior to repair?

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Abstract
A best evidence topic in cardiac surgery was written according to a structured protocol. The question addressed was 'In adult patients undergoing redo surgery for left atrioventricular valve regurgitation after atrioventricular septal defect correction, is

replacement superior to repair?' Altogether more than 109 papers were found using the reported search, of which eight represented the best evidence to answer the clinical question. The authors, journal, date and country of publication, patient group studied, study type, relevant outcomes, and results of these papers are tabulated. We conclude that left atrioventricular valve (LAVV) repair should be the first line approach and the use of transesophageal echocardiography (TEE) in operating room is mandatory. When complex anatomy and multiple anomalies of the LAVV are present the risk of a suboptimal repair is high and is associated with elevated subsequent risk of early reintervention. Prosthetic valve replacement is suggested in these cases and there is no long-term survival difference compared to repair procedures. Unfortunately, the risk of complete heart block and permanent pacemaker (PMK) implantation is higher when replacement is performed. Prosthetic valve choice is in favor of mechanical valves, mainly due to the young age of the patients. In the selected articles the frequency of valve replacement ranged from 14 to 34% and a mechanical valve was used in nearly all cases in the presented series. We feel that for older patients or for those in whom long-term anticoagulation is a concern, biological prosthesis can be an option, also due to the growing and expanding experience of percutaneous/transapical valve-in-valve replacement in mitral position. Since in these patients the number of previous sternotomies is usually one or more and re-entry injuries can be a major source of perioperative mortality and morbidity, we believe that mini-thoracotomy approach can avoid potential damage; furthermore, arterial cannulation can be either central or peripheral according to the degree of visceral adhesions or surgeon's choice. Venous drainage should be provided by a percutaneous vacuum-assisted femoral double stage venous drainage, which is useful especially when concomitant tricuspid valve surgery is planned.

J Am Soc Echocardiogr. 2011 Jun:24(6):625-36. Epub 2011 Mar 9.

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, Radoud 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.

Europace. 2011 Jun:13(6):859-63. Epub 2011 Mar 8.

Cardiac outcome of pregnancy in women with a pacemaker and women with untreated atrioventricular conduction block.

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Source
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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.

Int J Cardiol. 2011 Jun 16:149(3):372-6. Epub 2010 Mar 20.

Quality of life and functional capacity can be improved in patients with Eisenmenger syndrome with oral sildenafil therapy.

Tay EL, Papaphylactou M, Diller GP, Alonso-Gonzalez R, Inuzuka R, Giannakoulas G, Harries C, Wort SJ, Swan L, Dimopoulos K, Gatzoulis MA.

Source
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Abstract
BACKGROUND:
Patients with Eisenmenger syndrome (ES) have a decreased exercise capacity and poor quality of life (QoL). While patients may survive to middle adulthood, the burden of disease is disabling. Sildenafil seems to improve exercise tolerance and hemodynamics, but there is no data to date on its impact on QoL.

METHODS:
Eisenmenger patients in New York Heart Association (NYHA) class III were recruited in a prospective study of efficacy and safety of oral sildenafil. The QoL endpoint was assessed

using a disease-specific questionnaire (CAMPHOR). Exercise capacity was assessed by means of six minute walk test (6MWT). All patients underwent comprehensive assessment at baseline and after 3months of treatment.

RESULTS:
Twelve patients (mean age was 34.3±10.2, 83% female) with various cardiac anatomies were recruited. No major adverse events during the follow-up or significant drop in resting oxygen saturation were recorded. After 3months of oral sildenafil therapy, all patients improved to NYHA II with a concomitant improvement in 6MWT distance (347.3±80.7 to 392.5±82.0m, p=0.002). All components of the CAMPHOR score, relating to symptoms, activity and QoL, improved significantly resulting in substantial improvement in the total CAMPHOR score (27.6±10.5 to 15.8±10.4, p=0.002).

CONCLUSIONS:
Three months of sildenafil therapy in adults with ES was well tolerated and associated with significant improvement in the QoL CAMPHOR questionnaire and in NYHA class and exercise capacity. Larger studies are warranted to assess long term efficacy of oral sildenafil and potential impact on survival.

Int J Cardiol. 2011 Jun 2:149(2):186-91. Epub 2010 Feb 14.

Recommendations for adult and paediatric cardiologists on obtaining additional qualification in "Adults with Congenital Heart Disease" (ACHD).

Hess J, Bauer U, de Haan F, Flesh J, Gohlke-Baerwolf C, Hagl S, Hofbeck M, Kaemmerer H, Kallfelz HC, Lange PE, Nock H, Schirmer KR, Schmaltz AA, Tebbe U, Wevand M, Breithardt G.

Source
German Heart Centre of the Technical University of Munich, Department of Paediatric Cardiology and Congenital Heart Defects, Germany.

Abstract
BACKGROUND:
The number of adult congenital heart disease (ACHD) patients will be larger in the medium to long term than that of children and adolescents with congenital heart disease. The present structures for the medical care of ACHD patients are not sufficient and need to be improved. Therefore the Task Force aimed at developing recommendations for adult and paediatric cardiologists to acquire the additional qualification "Adults with Congenital Heart Disease" (ACDH).

METHODS:
The members of the interdisciplinary Task Force were selected on the basis of their special clinical, scientific and organisational expertise. The leading author submitted a draft version, which was revised by a sub-group of the interdisciplinary Task Force. It was subsequently agreed upon and re-circulated by all the members of the Task Force. The recommendations were then presented to the relevant committees of all participating associations and groups and approved following detailed discussion.

RESULTS:
A training programme for acquiring an additional qualification in the treatment of adults with congenital heart disease was created successfully.

CONCLUSIONS:
The medical care of adults with congenital heart disease is a sub-speciality in the border area between adult cardiology and paediatric cardiology. ACHD cardiologists are to be specially trained experts with appropriate knowledge and special skills and experience in the diagnosis and therapy of congenital heart disease in adults. ACHD cardiologists should be able to recognise and treat problems that occur in adulthood in connection with congenital heart disease.

Int J Cardiol. 2011 Jun 2:149(2):157-63. Epub 2010 Feb 8.

Cardiovascular changes after transcatheter endovascular stenting of adult aortic coarctation.

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Source
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Abstract
BACKGROUND:
Longer term data on efficacy and clinical endpoints relating to transcatheter endovascular stenting in adults with aortic coarctation remains limited. We hypothesised that stenting would have effects on blood pressure, presence and extent of collaterals, left ventricular (LV) mass and vascular function.

METHODS:
Eighteen patients mean age 31.6±12.8 years were studied with clinical assessment and cardiovascular magnetic resonance before and after (10.2±2.2 months) aortic coarctation endovascular stenting. Fredriksen coarctation index increased and using this index no patient had significant coarctation (index <0.25) after stenting.

RESULTS:
Blood pressure decreased (153±17/82±14 versus 130±21/69±13mmHg; p<0.001) unrelated to change in existing anti-hypertensive therapy. LV ejection fraction increased (70±10 versus 74±8%; p=0.01) and LV mass index decreased (91±24 versus 82±20g/m(2); p=0.003). Collaterals appeared smaller and the degree of flow through collateral arteries decreased (40±29 versus -1±33%; p<0.001). Distensibility of the ascending aorta increased (4.0±2.5 versus 5.6±3.5×10(-)(3)mmHg(-1); p=0.04). Unexpectedly, right ventricular mass index decreased (35±7 versus 30±10g/m(2); p=0.01).

CONCLUSION:
All patients underwent successful relief of coarctation by endovascular stenting. Both cardiac and vascular beneficial outcomes were demonstrated. The reduction in LV mass suggests a potential for reduction in risk of adverse events and warrants further study.