January 31, 2011

In This Issue

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ISACHD to Meet in New Orleans

Sunday, April 3, 2011
6:30-8:30pm, Following the Educational Sessions

Location: TBA

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

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

International Society for Adult Congenital Heart Disease

ISACHD Newsletter

President's Message: ISACHD in 2011- The Society will Change

by *Barbara J.M. Mulder*

The executives are building a new frame of goals and initiatives for the nearby future. These goals will refer to health care, education and research. Starting from now, the first initiative concerns the Journal Watch. As you may have noticed, since April Philip Moons has prepared the Journal Watch for you in every monthly newsletter. From now on, this Journal Watch will be sent to all colleagues with interest in the field of ACHD, members and non-members. For you, as a member of ISACHD, a selected "article of the month" will be commented in the subsequent newsletter. This way, we hope to provide attractive facilities for you as an ISACHD member and to attract more attention worldwide to our society.

Another service ISACHD is trying to provide is a reduced fee for ISACHD members at ACHD courses and symposia. As a first example we made an arrangement with Matthias Greutmann from Zurich, Switzerland. For the yearly and well-known DACH symposium on ACHD, June 24th 2011, the organization offers ISACHD members a reduction of 5% for the registration fee.

Another exciting and successful joined effort of ISACHD and American partners is the American dream of a sub-specialty certification in ACHD. The American Academy of Pediatrics, the American Heart Association, the International Society of Adult Congenital Heart Disease and two patient advocacy groups (the Adult Congenital Heart Association and the Children's Heart Foundation) joined forces with the American College of Cardiology to establish a sub-specialty certification in ACHD.The participation

of Michael Landzberg and Curt Daniels , as ISACHD's representatives has been an incredible asset. Undoubtedly, both the ACHD profession as well as the more than one million ACHD patients living in the US will benefit tremendously from this collaboration and shared vision. As a result of the certification, life - long specialized care for ACHD patients will be more accessible, and have a quantified standard. We are presently awaiting the last steps to be taken.

The ISACHD executives hope to serve you as a member with increasing facilities and valuable services. Any suggestions and ideas for further improvements are very welcome and can be sent to b.j.mulder@amc.uva.nl.

Barbara J.M. Mulder
President

Case Report: Aortic dissection presenting as acute pericarditis in a young male with bicuspid aortic valve

Florencia Crespo, Matias Clavero, Fernando Daghero, Marcelo Figueroa, Monica Benjamin, Luis Alday

Section of Adult Congenital Heart Disease and Cardiovascular Unit, Sanatorio Allende, Cordoba, Argentina.

A previously healthy 23-year-old male presented to the Emergency Department because of a 3-day history of severe chest pain that was partially relieved lying down in the right lateral and dorsal decubitus and by taking NSAID's drugs. He was slightly feverish and denied any chest injuries or symptoms of respiratory infection in the near past. Pertinent physical findings were a body temperature of 37.4 degrees Celsius, a heart rate of 95 beats per minute, normal peripheral pulses, and a blood pressure of 140/80 and 130/70 mmHg in the right and left arms respectively. Examination of the heart showed an ejection click at the apex, normal 2nd heart sound, and grade 1/6 ejection systolic and early diastolic murmurs in the aortic area and along the left sternal border. There were no other abnormalities in the physical examination.

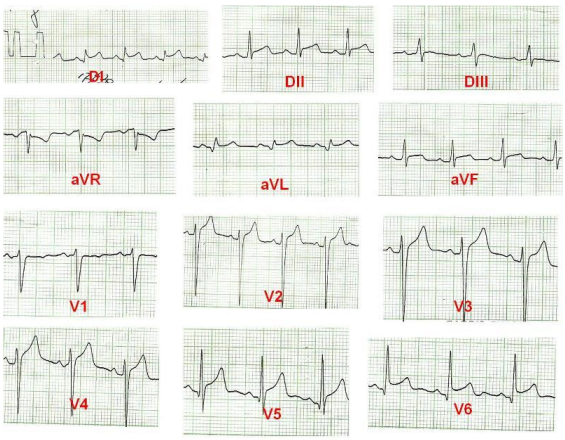

Figure 1 EKG on Admission

The EKG was compatible with acute pericarditis since it showed widespread concave ST segment elevation and PR segment depression in all leads but aVR and V1 which showed opposite findings. (Figure 1) The chest x-ray showed slight cardiomegaly with a dilated ascending aorta and normal pulmonary vasculature. (Figure 2) Laboratory studies showed increased sedimentation rate (19 mm) and PCR (8.3 mg%), and normal troponin T determination.

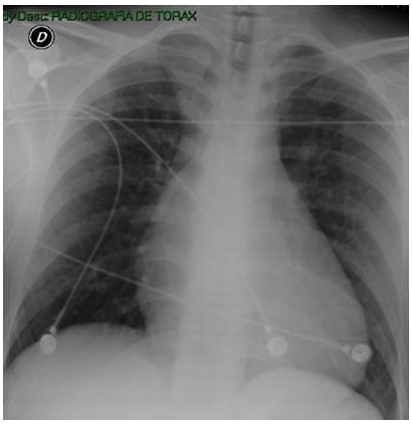

Figure 2 Chest x-ray on Admission

With the clinical diagnosis of acute pericarditis, the patient was admitted to the coronary care unit where a TTE followed by a TEE were performed to investigate the origin of the heart murmurs. The studies disclosed aneurysmal dilation of the aortic root with a maximal diameter of 6.8 cm, a bicuspid aortic valve with fusion of the right and left coronary valve leaflets, moderate aortic regurgitation, and an intimal tear 2.0 cm above the valvular plane in the anterior aortic wall without involvement of the coronary arteries, the aortic arch that measured 24 mm in diameter, or the brachiocephalic arteries. (Figures 3 and 4) There was also a mild amount of pericardial fluid. A multislice thoracic CT scan corroborated the echocardiographic findings and also showed that the dissection involved the posterior wall of the aortic annulus and extended up to the origin of the arch.

2008-2010

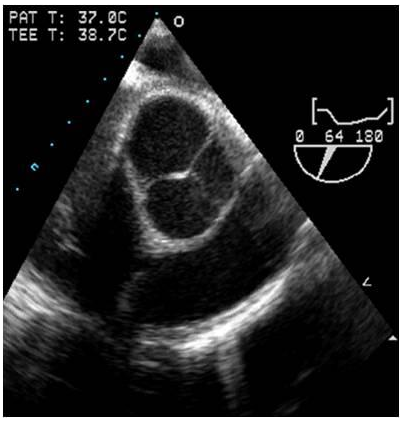

Figure 3 TEE short axis view

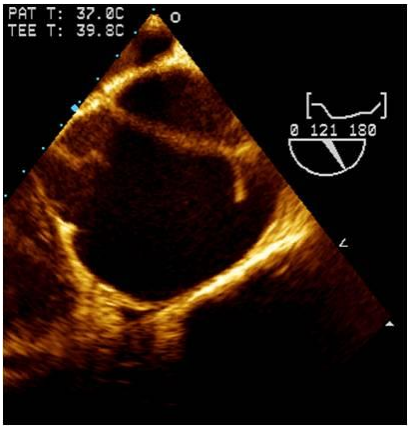

Figure 4 TEE from mid-esophagus showing aortic dissection

The patient was immediately transferred to surgery undergoing replacement of the aortic root and valve with a 25 Saint Jude Medical valved graft, and coronary arterial reimplantation. The postoperative course was uneventful the patient being discharged on the 5th day following surgery.
Anatomopathological studies of the excised aortic root and valve were compatible with dissection and medial cystic necrosis and recent hemorrhage of the aortic wall. Focal myxoid degeneration of the aortic valve was also present.

Comment

This previously asymptomatic young patient presented to Emergency with severe chest pain and EKG changes typical of pericarditis. The presence of a diastolic murmur in the aortic area prompted the performance of imaging studies that showed bicuspid aortic valve with moderate aortic insufficiency, aneurysmal aortic root, dissection of the ascending aorta, and pericardial fluid. Immediate surgical treatment was carried out with an excellent outcome. We do not know whether the aortic incompetence preceded or was the consequence of the dissection. However, there is no question that the aortic dilation was out of proportion to the valve malfunction. Pathological examination showed cystic medial necrosis adding evidence to previous reports indicating that a diseased aorta is frequently associated with a bicuspid aortic valve.
Aside from being the most common cardiovascular anomaly, the bicuspid aortic valve is prone to many complications and subject of continuous interest from physicians. (1, 2) Recent contributions have confirmed a genetic origin, a different pathogenesis according to the valves fused, and the frequency of associated aortopathy which appears to be nonrelated to flow distortion caused by the bicuspid valve. (3-5)

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Article of the Month: January 2011

Commentary on paper "Heart Rate Response During Exercise and Pregnancy Outcome in Women with CHD"

by: Carla Zomer

Lui et al. conducted a retrospective multicenter study to evaluate the relationship between cardiopulmonary exercise testing with measures of heart rate response on pregnancy outcomes in women with congenital heart disease (CHD). Main findings were that peak heart rate (pHR), chronotropic index (CRI), and percentage of maximum age predicted heart rate (%MPHR) were significantly associated with a maternal cardiac event during pregnancy. However, after adjustment for antiarrhythmic drugs, only CRI remained predictive. pHR, CRI %MPHR and heart rate reserve (HRR) were associated with an adverse neonatal event. pVO2 was not associated with any adverse pregnancy outcome. In conclusion, a blunted heart rate response to exercise in women with CHD is associated with higher risks of maternal cardiac and neonatal adverse events.
The results of this study seem promising for further improving risk stratification of women with CHD considering pregnancy. Since women with (complex) CHD have an increased risk of adverse cardiac events during pregnancy and more and more women survive to child-bearing age, the need for proper risk stratification is evidently present. Current risk stratification models are for a large part based on subjective assessment of the patient's clinical status. Therefore, the objective measures of chronotropic response during exercise could definitely add predictive value to the existing models. However, we should keep in mind that the mechanisms behind this association are not yet fully understood and the use of this parameter should be in the context of other clinical and hemodynamic factors.

Article:

Lui GK, Silversides CK, Khairy P, Fernandes SM, Valente AM, Nickolaus MJ, Earing MG, Aboulhosn JA, Rosenbaum MS, Cook S, Kay JD, Jin Z, Gersony DR; for the Alliance for Adult Research in Congenital Cardiology (AARCC). Heart Rate Response During Exercise and Pregnancy Outcome in Women With Congenital Heart Disease. Circulation. 2011 Jan 10. [Epub ahead of print]

Commentary:

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Journal Watch

Circulation. 2011 Jan 10. [Epub ahead of print]

Heart Rate Response During Exercise and Pregnancy Outcome in Women With Congenital Heart Disease.

Lui GK, Silversides CK, Khairy P, Fernandes SM, Valente AM, Nickolaus MJ, Earing MG, Aboulhosn JA, Rosenbaum MS, Cook S, Kay JD, Jin Z, Gersony DR; for the Alliance for Adult Research in Congenital Cardiology (AARCC).
Montefiore Medical Center, Bronx, NY.

Abstract

Background- Cardiopulmonary exercise testing is often used to evaluate exercise capacity in adults with congenital heart disease including women who are considering pregnancy. The relationship between cardiopulmonary exercise testing parameters and pregnancy outcome has not been defined. Methods and Results- We conducted a multicenter retrospective observational study of women with congenital heart disease who had undergone cardiopulmonary exercise testing within 2 years of pregnancy or during the first trimester. Cardiopulmonary exercise testing variables included peak oxygen consumption and measures of chronotropic response: peak heart rate, percentage of maximum age predicted heart rate, heart rate reserve (peak heart rate-resting heart rate), and chronotropic index [(peak heart rate-resting heart rate)/(220-age-resting heart rate)]. We identified 89 pregnancies in 83 women. There were 4 spontaneous abortions and 1 termination. One or more adverse cardiac events occurred in 18%; congestive heart failure in 14%, and sustained arrhythmia in 7%. Peak heart rate (odds ratio [OR] 0.71; 95% confidence interval [CI] [0.53, 0.94]; P=0.02), percentage of maximum age predicted heart rate (OR 0.93; 95% CI [0.88, 0.98]; P=0.01), and chronotropic index (OR 0.65; 95% CI [0.47, 0.90]; P=0.01) were associated with a cardiac event. Neonatal events occurred in 20%. Peak heart rate (OR 0.75; 95% CI [0.58, 0.98]; P=0.04), percentage of maximum age predicted heart rate (OR 0.94; 95% CI [0.89, 0.99]; P=0.02), heart rate reserve (OR 0.8; 95% CI [0.64, 0.99]; P=0.04), and chronotropic index (OR 0.73; 95% CI [0.54, 0.98]; P=0.04) correlated with a neonatal event. Peak oxygen consumption was not associated with an adverse pregnancy outcome. Conclusions- Abnormal chronotropic response correlates with adverse pregnancy outcomes in women with congenital heart disease and should be considered in refining risk stratification schemes.

Eur Heart J. 2011 Jan 8. [Epub ahead of print]

Turning 18 with congenital heart disease: prediction of infective endocarditis based on a large population.

Verheugt CL, Uiterwaal CS, van der Velde ET, Meijboom FJ, Pieper PG, Veen G, Stappers JL, Grobbee DE, Mulder BJ.

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Abstract

Aims The risk of infective endocarditis (IE) in adults with congenital heart disease is known to be increased, yet empirical risk estimates are lacking. We sought to predict the occurrence of IE in patients with congenital heart disease at the transition from childhood into adulthood. Methods and results We identified patients from the CONCOR national registry for adults with congenital heart disease. Potential predictors included patient characteristics, and complications and interventions in childhood. The outcome measure was the occurrence of IE up to the age of 40 and 60. A prediction model was derived using the Cox proportional hazards model and bootstrapping techniques. The model was transformed into a clinically applicable risk score. Of 10 210 patients, 233 (2.3%) developed adult-onset IE during 220 688 patient-years. Predictors of IE were gender, main congenital heart defect, multiple heart defects, and three types of complications in childhood. Up to the age of 40, patients with a low predicted risk (<3%) had an observed incidence of less than 1%; those with a high predicted risk (≥3%) had an observed incidence of 6%. The model also yielded accurate predictions up to the age of 60. Conclusion Among young adult patients with congenital heart disease, the use of six simple clinical parameters can accurately predict patients at relatively low or high risk of IE. After confirmation in other cohorts, application of the prediction model may lead to individually tailored medical surveillance and educational counseling, thus averting IE or enabling timely detection in adult patients with congenital heart disease.

Am J Cardiol. 2011 Jan 15;107(2):309-14

Electrical remodeling following percutaneous pulmonary valve implantation.

Plymen CM, Bolger AP, Lurz P, Nordmeyer J, Lee TY, Kabir A, Coats L, Cullen S,Walker F, Deanfield JE, Taylor AM, Bonhoeffer P, Lambiase PD.

Departments of Adult Congenital Heart Disease and Electrophysiology, The Heart Hospital, University College London Hospitals NHS Foundation Trust, London, United Kingdom.

Abstract

Sudden cardiac death in congenital heart disease is related to increased right ventricular end-diastolic volume (RVEDV), abnormalities of QRS duration, and QRS, JT, and QT dispersions. Surgical pulmonary valve replacement and percutaneous pulmonary valve implantation (PPVI) decrease RVEDV, but the effects of PPVI on surface electrocardiographic parameters are unknown. PPVI represents a pure model of RV mechanical and electrophysiologic changes after replacement. This prospective study sought to determine the effects of PPVI on surface electrocardiographic parameters: Ninety-nine PPVI procedures in patients with congenital heart disease (23.1 ± 10 years of age) were studied

before, after, and 1 year after PPVI with serial electrocardiograms and echocardiogram/magnetic resonance images. Forty-three percent had pulmonary stenosis, 27% pulmonary regurgitation (PR), and 29% mixed lesions. In those with predominantly PR (n = 26), QRS duration decreased significantly (135 ± 27 to 128 ± 29 ms, p = 0.007). However, in the total cohort no significant change in QRS duration at 1 year was observed (137 ± 29 to 134 ± 29 ms). Corrected QT interval and QRS, QT, and JT dispersions significantly decreased at 1 year (p ≤0.001). RVEDV correlated with preprocedure QRS duration (r = 0.34, p <0.002) but there was no correlation after PPVI. In conclusion, this is the first study reporting electrical remodeling after isolated PPVI and it confirms that decreases in QRS duration occur after PPVI in PR, as reported for equivalent surgical cohorts. Further, increased homogeneity of repolarization in combination with improved conduction may decrease arrhythmic events in congenital cardiac patients with pulmonary valvular disease.

Int J Cardiol. 2010 Dec 27. [Epub ahead of print]

Structure and process measures of quality of care in adult congenital heart disease patients: A pan-Canadian study.

Beauchesne LM, Therrien J, Alvarez N, Bergin L, Burggraff G, Chetaille P, Gordon E, Kells CM, Kiess M, Mercier LA, Oechslin EN, Stein J, Tam JW, Taylor D, Williams A, Khairy P, Mackie AS, Silversides CK, Marelli AJ.
University of Ottawa Heart Institute, Canada.

Abstract

BACKGROUND: There are more adults than children with congenital heart disease. Of over 96,000 ACHD patients in Canada, approximately 50% require ongoing expert care. In spite of published recommendations, data on the quality of care for ACHD patients are lacking.

METHODS: Survey methodology targeted all Canadian Adult Congenital Heart (CACH) network affiliated ACHD centers. Clinics were asked to prospectively collect outpatient and procedural volumes for 2007. In 2008, centers were surveyed regarding infrastructure, staffing, patient volumes and waiting times.

RESULTS: All 15 CACH network registered centers responded. The total number of patients followed in ACHD clinics was 21,879 (median per clinic=1132 (IQR: 585, 1816)). Of the total 80 adult and pediatric cardiologists affiliated to an ACHD clinic, only 27% had received formal ACHD training. Waiting times for non-urgent consultations were 4±2months, and 4±3months for percutaneous and surgical procedures. These were beyond Canadian recommended targets at 11 sites (73%) for non-urgent consultations, at 8 sites (53%) for percutaneous interventions and 13 sites (87%) for surgery.

CONCLUSIONS: Of a minimum number of 96,000 ACHD patients in Canada, only 21,879 were being regularly followed in 2007. At most sites waiting times for ACHD services were beyond Canadian recommended targets. In spite of universal health care access, published guidelines for ACHD patient structure and process measures of health care quality are not being met.

Int J Cardiol. 2010 Dec 20. [Epub ahead of print]

Factors associated with surgery for active endocarditis in congenital heart disease.

Murakami T, Niwa K, Yoshinaga M, Nakazawa M.
Department of Adult Congenital Heart Disease and Pediatrics, Chiba Cardiovascular Center, Ichihara City, Japan.

Abstract

BACKGROUND: Despite the recent progress of cardiac surgery, the indications for surgical intervention during the active phase of infective endocarditis have not yet been established in patients with congenital heart diseases due to the limited number of such patients. The present study aims to determine the surgical indications for active infective endocarditis in congenital heart diseases.

METHODS: A retrospective observational cohort multi-center study on infective endocarditis with congenital heart diseases was conducted from January 1997 to December 2001 in Japan and 239 patients were registered. Sixty-one (26%) of the 239 patients had undergone surgical therapy for active infective endocarditis, which was defined as cardiac surgery during administration of intravenous antibiotics.

RESULTS: There were 7 deaths (11%). A univariate regression analysis revealed that the factors significantly associated with the need for surgical intervention for active IE were the lack of diagnosis of cardiac disorders before the onset of infective endocarditis, aortic valve infective endocarditis, perivalvular abscess, presence of heart failure, and change of antibiotics. A stepwise logistic regression analysis revealed that the presence of a perivalvular abscess, heart failure and a change in the antibiotics were independent determinant factors for the need for surgical treatment of active infective endocarditis in patients with congenital heart diseases.

CONCLUSIONS: Surgery should therefore be considered even during the active phase in patients with congenital heart diseases and infective endocarditis, when they develop associated with heart failure, a perivalvular abscess, or the need for a change in antibiotics.

Echocardiography. 2010 Dec 22. doi:10.1111/j.1540-8175.2010.01277.x. [Epub ahead of print]

Right Atrial Size Relates to Right Ventricular End-Diastolic Pressure in an Adult Population with Congenital Heart Disease.

Do DH, Therrien J, Marelli A, Martucci G, Afilalo J, Sebag IA.

Echocardiography Laboratory, Division of Cardiology, Sir Mortimer B. Davis-Jewish General Hospital, Montreal, Canada McGill Adult Unit for Congenital Heart Disease Excellence, McGill University Health Center, Montreal, Canada Department of Medicine, McGill University, Montreal, Canada Department of Medicine, Division of Cardiology, Centre Hospitalier Universitaire de Sherbrooke, Sherbrooke University, Sherbrooke, Canada.

Abstract

Aim: Noninvasive markers of right ventricular (RV) diastolic dysfunction are limited by their lack of reproducibility and accuracy. We tested the hypothesis that right atrial (RA) size measured by echocardiography was correlated to invasive parameters of RV diastolic filling. Methods and Results: We studied 31 consecutive adult patients with congenital heart disease. From 2D echocardiography images, we measured maximal RA long-axis and short-axis lengths, area and volume. We compared each of these measures to right ventricular end-diastolic pressure (RVEDP) and mean right atrial pressure (mRAP) measured by right heart catheterization. RA long-axis, short-axis, area, and volume correlated significantly with RVEDP ($r = 0.78$, $P < 0.001$; $r = 0.61$, $P < 0.001$; $r = 0.79$, $P < 0.001$; and $r = 0.75$, $P < 0.001$, respectively) and mRAP ($r = 0.66$, $P < 0.001$; $r = 0.56$, $P = 0.002$; $r = 0.70$, $P < 0.001$; $r = 0.68$, $P < 0.001$, respectively). Single cut points for each echocardiographic parameter demonstrated reasonable accuracy to rule-in and rule-out RVEDP ≥7 mm Hg (sensitivity = 74%, specificity = 82%, positive LR = 4.1, negative LR = 0.32 for RA long-axis of 49 mm; sensitivity = 89%, specificity = 82%, positive LR = 4.9, negative LR = 0.12 for RA area of 14 cm(2) ; sensitivity = 89%, specificity = 82%, positive LR = 4.9, negative LR = 0.13 for RA volume of 37 mL). Conclusion: RA size measured by echocardiography is strongly correlated to invasive parameters of RV diastolic filling and predicts high RV end-diastolic pressure. (Echocardiography ****;**:109-116).

Am Heart J. 2011 Jan;161(1):123-9

The impact of 2007 infective endocarditis prophylaxis guidelines on the practice of congenital heart disease specialists.

Pharis CS, Conway J, Warren AE, Bullock A, Mackie AS.

Department of Pediatrics, Stollery Children's Hospital and the University of Alberta, Edmonton, Alberta, Canada.

Abstract

BACKGROUND: the impact of the 2007 American Heart Association endocarditis prophylaxis guidelines on clinician practice has not been well established. Our objective was to evaluate how the American Heart Association endocarditis guidelines changed the practice of cardiologists who manage congenital heart disease and to ascertain the degree of practice variation among cardiologists.

METHODS: a cross-sectional Web-based survey was e-mailed to Canadian (n = 134), Australian (n = 33), New Zealand (n = 9), and a random sample of American (n = 250) pediatric and adult congenital heart disease cardiologists in 2008. Nonrespondents received the survey 4 times by e-mail and once by regular post.

RESULTS: the response rate was 55%. The lesions for which cardiologists were most evenly divided between recommending versus not recommending

prophylaxis were "rheumatic mitral stenosis of moderate severity" (45% recommended prophylaxis) and "perimembranous ventricular septal defect (VSD) status post surgical patch closure with no residual shunt 3 months post-operatively" (54% recommended prophylaxis). The lesions for which the greatest proportion of cardiologists discontinued prophylaxis were "small muscular VSD, no previous endocarditis" (80% discontinued prophylaxis) and "small audible patent ductus arteriosus" (83% discontinued prophylaxis). Only 69% recommended prophylaxis for "VSD s/p surgical patch closure with small residual shunt" despite current guidelines recommending prophylaxis for this scenario. Twenty-eight percent of respondents felt that the new guidelines leave some patients at risk, and 6% would not recounsel any low-risk patients following these guidelines.

CONCLUSIONS: the 2007 guidelines have resulted in a substantial change in endocarditis prophylaxis. There remains considerable heterogeneity among cardiologists regarding the prophylaxis of certain cardiac lesions.

Kidney Blood Press Res. 2010 Dec 16;34(1):41-45. [Epub ahead of print]

Symmetrical Dimethylarginine Outperforms CKD-EPI and MDRD-Derived eGFR for the Assessment of Renal Function in Patients with Adult Congenital Heart Disease.

Tutarel O, Deneck A, Bode-Boger SM, Martens-Lobenhoffer J, Schieffer B, Westhoff-Bleck M, Kielstein JT.

Department of Cardiology and Angiology, Hannover Medical School, Hanover, Germany.

Abstract

Background/Aims: Adults with congenital heart disease exhibit a 3-fold higher mortality in the presence of chronic kidney disease, hence assessment of renal function is crucial in this patient population. Formulas for the estimation of glomerular filtration rate (GFR) have not been evaluated in this patient population. Therefore, this study compares different markers and equations for the estimation of renal function in adults with congenital heart disease. Methods: Renal function was assessed in 102 patients using the Modification of Diet in Renal Disease (MDRD) equation, the simplified MDRD equation, the Chronic Kidney Disease Epidemiology Collaboration (CKD-EPI) equation and the Cockcroft-Gault formula. Additionally, symmetrical dimethylarginine (SDMA) was measured. Those parameters were compared to cystatin C-derived GFR using the Larsson equation. Results: GFR estimates using the original MDRD ($r = 0.465$, $p < 0.001$) and the CKD-EPI equation ($r = 0.462$, $p < 0.001$) showed a similar strong correlation with the cystatin C-based eGFR equation, while eGFR using the simplified MDRD equation showed a slightly weaker correlation ($r = 0.439$, $p < 0.001$). The Cockcroft-Gault formula showed no correlation at all to the cystatin C-based eGFR ($r = 0.144$, $p = 0.17$). The strongest correlation was observed for SDMA and cystatin C-based eGFR ($r = -0.552$, $p < 0.001$). Conclusion: GFR in adults with congenital heart disease should be estimated using the original MDRD or the CKD-EPI formula. SDMA seems to be a promising marker of renal function for this patient group.

J Thorac Cardiovasc Surg. 2011 Jan;141(1):162-70. Epub 2010 Nov 5.

The hemi-Mustartd/bidirectional Glenn atrial switch procedure in the double-switch operation for congenitally corrected transposition of the great arteries: rationale and midterm results.

Malhotra SP, Reddy VM, Qiu M, Pirolli TJ, Barboza L, Reinhartz O, Hanley FL.

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Abstract

OBJECTIVE: This study was undertaken to assess the risks and benefits of the double-switch operation using a hemi-Mustard atrial switch procedure and the bidirectional Glenn operation for congenitally corrected transposition of the great arteries. To avoid complications associated with the complete Senning and Mustard procedures and to assist right-heart hemodynamics, we favor a modified atrial switch procedure, consisting of a hemi-Mustard procedure to baffle inferior vena caval return to the tricuspid valve in conjunction with a bidirectional Glenn operation.

METHODS: Between January 1994 and September 2009, anatomic repair was achieved in 48 patients. The Rastelli-atrial switch procedure was performed in 25 patients with pulmonary atresia and the arterial-atrial switch procedure was performed in 23 patients. A hemi-Mustard procedure was the atrial switch procedure for 70% (33/48) of anatomic repairs.

RESULTS: There was 1 in-hospital death after anatomic repair. There were no late deaths or transplantation. At a median follow-up of 59.2 months, 43 of 47 survivors are in New York Heart Association class I. Bidirectional Glenn operation complications were uncommon (2/33), limited to the perioperative period, and seen in patients less than 4 months of age. Atrial baffle-related reoperations or sinus node dysfunction have not been observed. Tricuspid regurgitation decreased from a mean grade of 2.3 to 1.2 after repair (P = .00002). Right ventricle-pulmonary artery conduit longevity is significantly improved.

CONCLUSIONS: We describe a 15-year experience with the double-switch operation using a modified atrial switch procedure with favorable midterm results. The risks of the hemi-mustard and bidirectional Glenn operation are minimal and are limited to a well-defined patient subset. The benefits include prolonged conduit life, reduced baffle- and sinus node-related complications, and technical simplicity.

Heart. 2011 Jan;97(2):118-23. Epub 2010 Oct 21.

Pre-stenting with a bare metal stent before percutaneous pulmonary valve implantation: acute and 1-year outcomes.

Nordmeyer J, Lurz P, Khambadkone S, Schievano S, Jones A, McElhinney DB, Taylor AM, Bonhoeffer P.
Cardiovascular Unit UCL Institute of Child Health, London, UK.

Abstract

OBJECTIVES: To determine the feasibility and safety of pre-stenting with a bare metal stent (BMS) before percutaneous pulmonary valve implantation (PPVI), and to analyse whether this approach improves haemodynamic outcomes and impacts on the incidence of PPVI stent fractures.

DESIGN: Retrospective analysis of prospectively collected data.

SETTING: Tertiary paediatric and adult congenital heart cardiac centre.

PATIENTS AND INTERVENTIONS: 108 consecutive patients with congenital heart disease underwent PPVI between September 2005 and June 2008 (54 with PPVI alone, 54 with BMS pre-stenting before PPVI).

RESULTS: There were no significant differences in procedural complication rates. Acutely, there was no difference in haemodynamic outcomes. Serial echocardiography revealed that in the subgroups of 'moderate' (26-40 mmHg) and 'severe' (>40 mmHg) right ventricular outflow tract (RVOT) obstruction, patients with pre-stenting showed a tendency towards lower peak RVOT velocities compared to patients after PPVI alone (p=0.01 and p=0.045, respectively). The incidence of PPVI stent fractures was not statistically different between treatment groups at 1 year (PPVI 31% vs BMS+PPVI 18%; p=0.16). However, pre-stenting with BMS was associated with a lower risk of developing PPVI stent fractures (HR 0.35, 95% CI 0.14 to 0.87, p=0.024). The probability of freedom from serious adverse follow-up events (death, device explantation, repeat PPVI) was not statistically different at 1 year (PPVI 92% vs BMS+PPVI 94%; p=0.44).

CONCLUSIONS: Pre-stenting with BMS before PPVI is a feasible and safe modification of the established implantation protocol. Pre-stenting is associated with a reduced risk of developing PPVI stent fractures.

J Thorac Cardiovasc Surg. 2011 Jan;141(1):188-92. Epub 2010 Jul 8.

Axillary arteriovenous fistula for the palliation of complex cyanotic congenital heart disease: is it an effective tool?

Quinonez LG, Brown ML, Dearani JA, Burkhart HM, Puga FJ.
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Abstract

OBJECTIVE: Patients with complex cyanotic congenital heart disease and a bidirectional cavopulmonary connection who are not candidates for or had failed Fontan operation may experience progressive cyanosis. An axillary arteriovenous anastomosis may be constructed to augment pulmonary blood

flow. This report reviews our results with this approach in this complex group of patients.

METHODS: The records of patients with previous cavopulmonary connections who underwent a surgical anastomosis between the axillary artery and the vein for palliation of severe progressive cyanosis were reviewed.

RESULTS: Eleven patients were identified. The median age at the time of the axillary arteriovenous anastomosis was 19.2 years (7.97-41.75 years). Seven patients were not candidates for the Fontan operation, and 4 patients had failed Fontan surgery. Three of the anastomoses were constructed with a side-to-side technique, and 8 anastomoses were constructed with a short interposition graft. Median fistula size was 5 mm (3-6 mm). There was no operative mortality and 1 late death. Median survival was 2.85 years (0.01-7.22 years). All fistulae were patent at follow-up. Median preoperative arterial oxygen saturation was 84% (80%-86%) and 82% (76%-88%) at follow-up (P = .38). Median preoperative hemoglobin was 18.5 g/dL (11.7-22.6 g/dL) and 19.2 g/dL (14.6-22.6 g/dL) at follow-up (P = .97). Median preoperative systemic ventricular ejection fraction was 51% (27%-60%) and 46.5% (28%-60%) at follow-up (P = 1). Significant functional improvement was seen in only 1 patient.

CONCLUSIONS: In patients with complex cyanotic congenital heart disease who are not candidates for or had failed Fontan operation, palliation with an axillary arteriovenous fistula did not improve cyanosis or polycythemia. Functional outcome and ventricular ejection fraction did not improve or deteriorate.

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Prevalence of adult patients with congenital heart disease in Japan.

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Abstract

BACKGROUND: Today most patients with congenital heart disease (CHD) can be expected to survive into adulthood. Reports regarding the number of adults with CHD in Japan are scarce. Our study aims to define the number of these adults.

MATERIAL AND METHODS: The estimated number of infants born in Japan with major CHDs since 1947 was calculated together with mortality rates. We estimated the number of CHD survivors from data on survival rates of unoperated and postoperative patients. The number of deaths from 1968 to 1997 was analyzed using individual death certificates held by the Japanese Government.

RESULTS: In 1967, 163,058 patients with CHD including 53,846 adults were assumed to be alive. From 1968 to 1997, 548,360 patients with CHD were born and 82,919 died. A total of 622,800 patients, including 304,474 children (49%) and 318,326 adults (51%) were estimated to be alive in 1997. From 1997 to 2007, there has been an estimated increase of 9000 adults every year, and in 2007, 409,101 adults are estimated to be alive.

CONCLUSIONS: The prevalence in adults with CHD in Japan has explosively increased from 1967 to 2007. There were 409,101 adults with CHD in 2007 with an annual increase of 9000. These data are crucial for planning the establishment in Japan of special facilities and resources necessary for the care of these patients.

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