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[In This Issue](#)

[Regional News](#)

[Case Report](#)

[Journal Watch](#)

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ISACHD Newsletter

President's Message

by Curt J Daniels



Dear ISACHD Members,

What an outstanding conference! The 22nd annual International Symposium on ACHD held in Toronto this past month met all expectations. The city of Toronto and the University of Toronto faculty and staff were the perfect hosts for this year's conference. Chaired by Erwin Oechslin (ISACHD Canadian Regional Representative), this year's conference "Beyond Saving Lives" brought a unique aspect to ACHD conferencing not previously discussed in a large international forum-end of life supportive and advanced care. The topic stimulated much discussion and set the stage for future programming. Also highlighted were the breakout sessions featuring various ACHD topics, allowing the audience up close and personal interaction with the panels for Q and A.

ISACHD was well represented with two sessions. Barbara Mulder (ISACHD past president) delivered an electrifying lecture under the session "Top 10 Challenges after Saving Lives." Barbara's lecture was a highlight of this year's conference. Also, in a separate session, key ISACHD members highlighted current initiatives in Global Health Care (C. Daniels, L. Alday), Education (G. Webb), Research (K. Niwa), and CCA (D. Pearson). We will look forward to next year, meeting again in Oregon.

ISACHD Work Groups will be continuing their efforts over the summer leading into a productive fall. Join ISACHD if you are not a member, and please contact your WG chairs to get involved with our Global Initiatives.

Please keep in mind that all these great efforts require your financial support as well. And because this is the time of the year to renew your 2012-2013 annual membership dues I'm asking you not to delay your membership renewal. Look for an e-mail with the invitation and instruction details on membership renewal coming very soon to your mailbox and take this important action.

Here are just a few advantages of being the ISACHD Member:

- Engage in ISACHD Global Initiatives; Global Health, Education, Research
- Renewed Members receive discounts to ISACHD endorsed international cardiology meetings. Recent endorsements include; Pediatric and Adult Interventional Cardiac Symposium (Chicago), 22nd Annual International Symposium for ACHD (Toronto), 2nd International Congress on Cardiac Problems in Pregnancy (Berlin), Cardiac Pathology and Imaging Course (Calgary). Membership certificates will be distributed to Members in good standing.
- Monthly Newsletter/Journal Watch - stay up to date with the most recent research and published articles - WE provide the information, YOU stay up to date and current
- Join your colleagues at our ISACHD meetings; American Heart Association, American College of Cardiology, European Society of Cardiology. Great opportunity to meet your colleagues, developed relationships, Wine and Dine!

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Michael J. Landzberg, MD

The direct URL link to renew your membership, or to join our Society is <http://www.isachd.org/login.php> , where detailed instructions are provided as well.

Curt J. Daniels
President

Regional News:**News from Asia Pacific**

By Koichiro Niwa

In Japan we met for two days for the 5th Seminar on ACHD in Tokyo, June 9-10. Various topics by specialists were presented, and up to 267 attendants joined the meeting. It was active and lively as always.

The ACHD Network meeting was held in Tokyo on June 8th. Representatives of departments of cardiology from 22 institutes who planned to have out-patient clinics in their institutions from 22 institutes were gathered. It was a challenging and promising event. Many cardiologists began to have an interest in the ACHD field.

In Singapore, a 1.5 day congenital program (see link below) with sessions on TOF, pulmonary hypertension, and shunts will be hosted at the ASEAN Cardiology Meeting from 12/7/12 to 14/7/2012. This is an adult cardiology meeting. (http://www.afcc2012.com/download/A3ScientificProgramme_R4.pdf)

The Cardiac Society (of Australia and New Zealand) New Zealand meeting was held last weekend in conjunction with the Royal Australasian College of Physicians in Auckland and had several well attended sessions for ACHD. We were privileged to have Dr. David Sahn from the US join us as a guest speaker, and he gave a wonderful presentation for the John Neutze Memorial lecture.

News from Latin America

By Luis Alday



During the next Argentine Congress of Cardiology to be held at the Buenos Aires Sheraton Hotel from October 5-7, 2012, a new ISACHD-SAC joint session will take place with Dr. Ariane Marelli, from Montreal, Canada, as the guest speaker. The Argentine Congress of Cardiology is usually a large meeting with attendance of nearly 10000 physicians from Argentina and neighboring countries with guest speakers from all over the world.

News from United States of America

By Bill Davidson

Registration is now open for AHA Scientific Sessions 2012 in Los Angeles, CA.

Case Report

Stroke as first manifestation of severe coarctation of the aorta in a young

2008-2010

Barbara Mulder, MD
2010-2012

female with Turner syndrome.

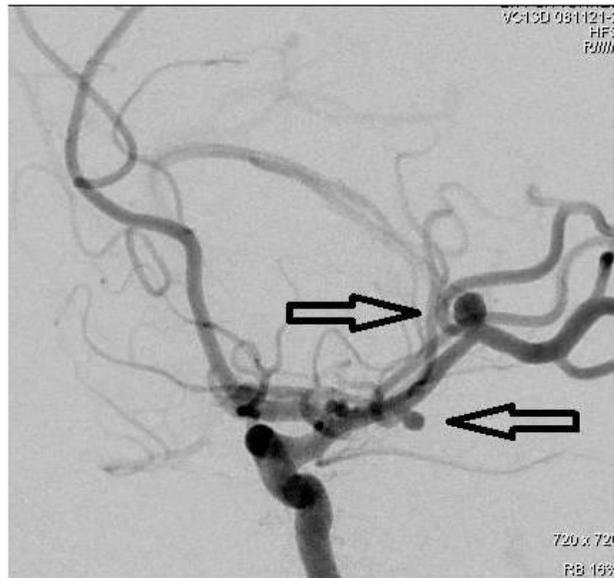
Alejandro Vivero , Julio César Suarez, Maria Jose Cabrera, Gustavo Foa Torres, Fernando Daghero, Monica Benjamin and Luis Alday
Section of Adult Congenital Heart Disease and Cardiovascular Unit.and Division of Neurology, Sanatorio Allende, Cordoba, Argentina

A 19-year-old female, with undiagnosed Turner syndrome, was referred to the Division of Neurology because of a history of severe headache, nausea and vomiting accompanied by right sided hemiparesis and dysarthria lasting 24 hours with complete recovery, 2 months prior to consultation. Congenital heart disease had been diagnosed at her home town in infancy but her parents never took her to a pediatric cardiologist. Headaches and pain in the legs while walking had been present for some time before the referral episode. On physical examination, she had most of the features of Turner syndrome like short stature, low set ears and hair line, shield chest with mammary hypertelorism, and poor sexual development. Femoral pulses were absent with a blood pressure of 140/80 and 65/40 mmHg in the upper and lower limbs respectively. An aortic systolic ejection click and a 2/6 systolic murmur were present at the left sternal border and interscapular region. The chest x-ray showed mild cardiomegaly, an inconspicuous aortic knob with inverted "E" sign and costal erosions typical of long standing coarctation of the aorta later confirmed by MRI (Figure 1).



Figure 1: Chest x-ray showing mild cardiomegaly, inconspicuous aortic knob, and rib erosions

The EKG had left atrial enlargement and left ventricular hypertrophy. An angio MRI of the brain showed 2 saccular (berry) aneurysms in the left carotid system at the mid cerebral artery and at the Sylvian bifurcation measuring 2.5 and 6 mm in diameter respectively. A smaller 1.5 mm aneurysm was also present at the right Sylvian bifurcation. Both left sided aneurysms were coil embolized uneventfully (Figure 2).



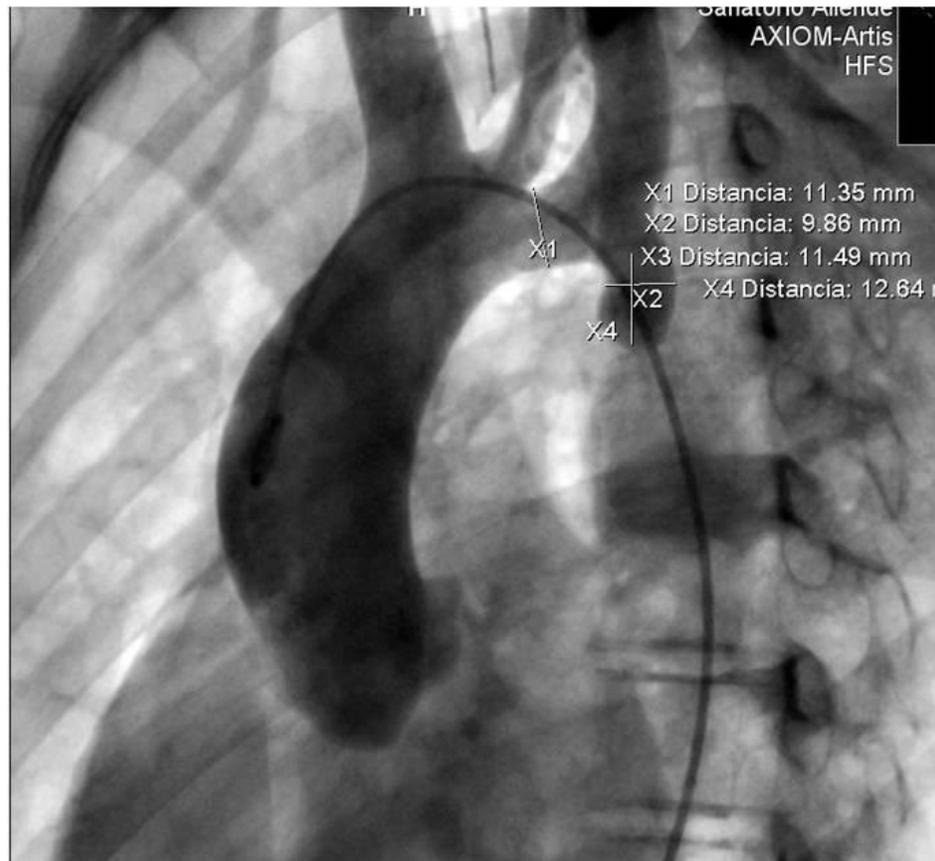
A.



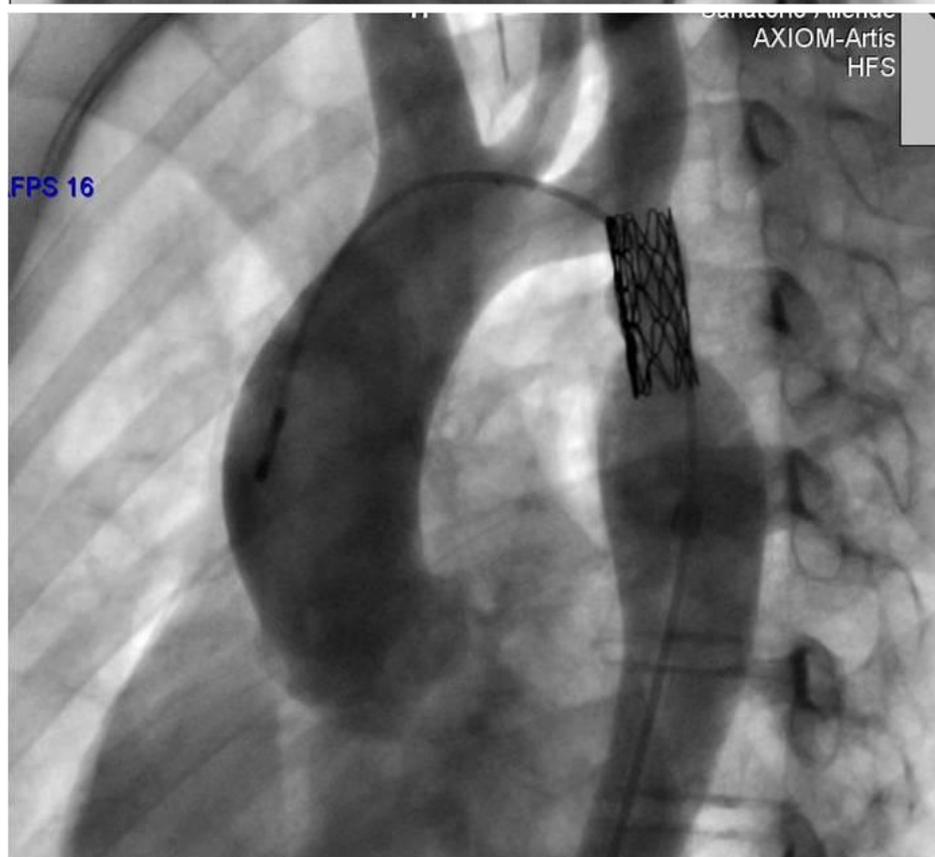
B.

Figure 2: Left intracranial berry aneurysms (arrows) pre (A) and post (B) embolization

The coarctation of the aorta was addressed in a separate procedure with planned stage stenting since the patient had Turner syndrome and these patients are prone to aortic wall complications. The aortogram showed severe coarctation of the aorta with hypoplasia of the isthmus and a bicuspid aortic valve (Figure 3 A).



A.



B.

Figure 3: Aortogram showing severe coarctation of the aorta with isthmal hypoplasia and bicuspid aortic valve, pre (A) and post (B) stenting

The peak systolic gradients across the coarctation under general anesthesia were 33 and 0 mmHg pre and post stenting with a 12 mm CP stent (Figure 3B). Eight months later the patient had none of the previous symptoms. Blood pressure measurements in the upper and lower extremities were 130/70 and 100/70 mmHg. The stent was further dilated to 15 mm with a fall of the hemodynamic gradient from 24 to 8 mmHg. No complications occurred following both interventions. Clinical blood pressures at discharge were 120/80 mmHg in all 4 limbs. The patient is now under genetical care.

Comment

Stroke secondary to rupture of berry intracranial aneurysms is the cause of referral in almost 5% of patients with coarctation of the aorta.(1) Turner syndrome is associated with coarctation of the aorta in around 15% of patients and should be ruled out in females presenting with coarctation.(2) Stenting is most likely the treatment of choice in grown up patients but should be performed with caution since aortic wall abnormalities may be present.(3)

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Journal Watch

ARTICLE OF THE MONTH: June 2012

Commentary on Sandoval et al paper "Does Anticoagulation in Eisenmenger Syndrome Impact Long-term Survival?" by Harsimran S. Singh, MD

In 1958, Dr. Paul Wood coined the eponym "Eisenmenger" in recognition of the German physician attributed with the first published case describing the clinical and pathologic features of an adult with a nonrestrictive ventricular septal defect at the turn of the 19th century.^{1,2} From an anatomic perspective, Eisenmenger Syndrome is defined as pulmonary hypertension in the setting of a reversed or bidirectional shunt at any level, but in broader terms, the syndrome implies unique physiology, pathologic features, and prognosis that is distinct from other types of pulmonary hypertension. One such feature special to Eisenmenger physiology is the simultaneous predilection for both bleeding and thrombosis. On one hand, elevated pulmonary artery pressures coupled with secondary erythrocytosis, high blood viscosity, decreased pulmonary flow velocity, and frequent atrial arrhythmias all may contribute to the high rates of in situ thrombosis in the pulmonary arteries.³⁻⁶ At the same time, hemoptysis and other significant bleeding are common manifestations in up to 20% of patients, possibly due to abnormal coagulation factors, relative thrombocytopenia, fragility of pulmonary vascular bed, and even vascular microinfarcts (from in situ thrombosis) in the lungs.^{4,6}

The question of whether to offer therapeutic anticoagulation in patients with Eisenmenger syndrome has been a topic of significant academic controversy, variability in clinical practice, and severely limited evidence. In this month's featured article, Sandoval et al attempt to shed some light on this question by

publishing their single institution experience from Mexico City comparing an upfront anticoagulation strategy with acenocoumarin in 48 patients versus no anticoagulation in 44 patients all with Eisenmenger Syndrome. 7 Using a retrospective cohort study design, they examine whether either treatment strategy affects 1) overall mortality and 2) significant bleeding events.

Upon study inclusion, the total population has an average age of 38 years with overwhelmingly simple unrepaired congenital heart defects, 27% with reported NYHA Class III-IV symptoms, mean PaO₂ 48 ± 8 mm Hg, and hemoglobin of 18.6 g/dl. No study patients were treated with any advanced therapies for pulmonary hypertension such as prostacyclin-analogues, endothelin-receptor antagonists, or phosphodiesterase inhibitors. At a mean of 7 years follow-up, overall mortality of the total study group was 23% (total of 21 events); causes of death included right heart failure (6), sudden death (5), cerebrovascular accident (4), pulmonary hemorrhage (3), cranial trauma (1), cardiac tamponade (1), and hemothorax (1). Using Kaplan Meier survival estimates, there was no statistically significant difference between the two treatment strategies of anticoagulation (23%, 11 events) vs. non anticoagulation (23%, 10 events). In univariate analysis, an increased risk of death was associated with worse baseline NYHA functional class, higher mean PA pressure, and a mean red cell corpuscular volume < 80 fL. Seven patients (16%) in the anticoagulation group experienced hemorrhagic complications out of which 4 were severe and 2 lead to patient death (tamponade and hemothorax). There were no reported major bleeding events in the non-anticoagulation group. Based on their data, the authors conclude that anticoagulation has no impact on long-term survival.

The research team should be applauded for tackling a study question for which very little evidence exists. This report represents one of the only published comparisons examining an upfront anticoagulation strategy in Eisenmenger syndrome. However, there are clearly significant study limitations that must temper any over-interpretation of the results. Without randomization of treatment strategies, it is impossible to account for selection bias that may have directed the anticoagulation strategies for each individual patient. Inherent to any retrospective study, recorder bias can be intrinsic in the methodology used for collecting data/outcomes. While an overall mortality of 23% at ~7 year follow-up may appear higher than expected, this study is likely underpowered to confirm the null hypothesis. With so few study events, it is difficult to use multivariate analysis to account for confounding variables; as such multivariate analysis was not performed in this study.

In their discussion, the authors address specific characteristics in their study population that may limit the generalizability of their findings. These include predominance of simple cardiac defects (compared to complex or iatrogenic lesions), patient's living at high altitude, and the inclusion of atrial septal defects which may represent a distinct etiology from other Eisenmenger patients.^{8,9} A large number of Eisenmenger patients (42 patients) were excluded from this study secondary to "inconsistent use of anticoagulation" as designated by the study inclusion criteria. These patients may represent a particularly challenging group with regards to anticoagulation decision making, and their inclusion into the study may alter its overall conclusions. Also, without routine imaging of the pulmonary arteries (i.e. pulmonary CT angiogram), this study cannot comment on differences in pulmonary thrombus burden as an expected advantage of anticoagulation.

In a world of limited resources and an overall declining incidence of Eisenmenger Syndrome thanks to early intervention, it is highly unlikely that a randomized prospective trial will ever be designed to answer the question "to anticoagulate or

not to anticoagulate." Thus we remain reliant on high quality observational studies using rigorous methodology to provide us with the best possible guidelines for care. With these caveats in mind, the data from Sandoval et al does not find an upfront strategy of anticoagulation to be clearly superior to no anticoagulation in regards to short-to-medium term mortality. Serious and life-threatening bleeding remains a very real potential complication of anti-coagulation that must play into the clinical decision making. Monitoring of the INR is particularly challenging in cyanotic patients where the amount of sodium citrate in test tubes must be adjusted to the hematocrit in order to avoid inaccurate readings. Thus any decision to anticoagulate and monitor INR levels in Eisenmenger patients should include specialty consultation (e.g. hematologists) who are familiar with the issues at hand. In the end, this study best supports an individualized approach weighing the risks and benefits in choosing an anticoagulation strategy in patients with Eisenmenger Syndrome.

Acknowledgement: Thanks to Dr. Oechslin for reviewing this commentary prior to submission.

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Article: Sandoval J, Santos LE, Córdoba J, Pulido T, Gutiérrez G, Bautista E, Martinez Guerra ML, Peña H, Broberg CS. Does Anticoagulation in Eisenmenger Syndrome Impact Long-term Survival? *Congenit Heart Dis*. 2012 May;7(3):268-76. doi: 10.1111/j.1747-0803.2012.00633.x.

Commentary: Harsimran Sachdeva Singh, MD
Fellow Adult Congenital Heart Disease
Toronto General Hospital
Division of Cardiology
Toronto, Ontario

Canada
Email: Harsimran.Singh@uhn.ca

Congenit Heart Dis.

2012 May;7(3):268-76. doi: 10.1111/j.1747-0803.2012.00633.x. Epub 2012 Feb 23.

[Does Anticoagulation in Eisenmenger Syndrome Impact Long-term Survival?](#)

Sandoval J, Santos LE, Córdova J, Pulido T, Gutiérrez G, Bautista E, Martínez Guerra ML, Peña H, Broberg CS.

Source

Cardiopulmonary Department, Ignacio Chavez National Institute of Cardiology, Mexico City, Mexico Adult Congenital Heart Program, Division of Cardiology, Oregon Health and Science University, Portland, Ore, USA.

Abstract

Objective: To determine the impact of anticoagulation on survival in Eisenmenger syndrome.

Background: The use of anticoagulation for primary prevention of adverse events in patients with Eisenmenger syndrome has been proposed but not studied. Strong arguments have been made both for and against anticoagulation based on the known risk of hemoptysis and pulmonary vascular thrombosis.

Design and Setting: Retrospective cohort study at a tertiary referral hospital.

Patients and Interventions:

One hundred forty-four patients with established Eisenmenger physiology all underwent initial laboratory, echocardiographic, and catheterization evaluation after initial referral. We retrospectively identified patients who were started on anticoagulation (AC) and compared them to patients who did not receive anticoagulation therapy (non-AC). Baseline variables were compared between groups, as well as between survivors and nonsurvivors. Analyses of prognostic factors and survival were done using Cox and Kaplan-Meier methods. Outcome

Measures: The primary outcome was death since time of baseline evaluation.

Results: We identified 48 anticoagulated and 44 nonanticoagulated patients with Eisenmenger physiology (oxygen saturation $82\pm 9\%$, PaO₂ 48 ± 8 mmHg, hemoglobin 18.6 ± 4 g/dL). More atrial septal defect patients were in the AC group, but there were no other baseline differences in clinical, functional, or hemodynamic data. After mean follow-up of 7 ± 5.4 years (range 1-31), 11 patients died in the AC and 10 died in the non-AC group. There was no survival difference between groups (log rank test=1.78; P is not significant). For the entire cohort, mortality was significantly associated with New York Heart Association class 3-4 (hazard ratio=4.2), evidence of right heart failure (hazard ratio=13.6), and a mean corpuscular volume < 80 fL (hazard ratio=3.8). Use of anticoagulation did not impact survival. Bleeding complications occurred in seven (16%) of AC patients, including two fatalities.

Conclusions: Anticoagulation had no impact on long-term survival in this limited study. These data may be useful in considering future studies addressing this question.

J Cardiovasc Magn Reson. 2012 May 31;14(1):32. [Epub ahead of print]

[Quantification of biventricular myocardial function using cardiac magnetic resonance feature tracking, endocardial border delineation and echocardiographic speckle tracking in patients with repaired tetralogy of fallot and healthy controls.](#)

Kempny A, Fernández-Jiménez R, Orwat S, Schuler P, Bunck AC, Maintz D, Baumgartner H, Diller GP.

Abstract

Background: Parameters of myocardial deformation have been suggested to be superior to conventional measures of ventricular function in patients with tetralogy of Fallot (ToF), but have required non-routine, tagged cardiovascular magnetic

resonance (CMR) techniques. We assessed biventricular myocardial function using CMR cine-based feature tracking (FT) and compared it to speckle tracking echocardiography (STE) and to simple endocardial border delineation (EBD). In addition, the relation between parameters of myocardial deformation and clinical parameters was assessed.

Methods: Overall, 28 consecutive adult patients with repaired ToF (age 40.4 +/- 13.3 years) underwent standard steady-state-free precession sequence CMR, echocardiography, and cardiopulmonary exercise testing. In addition, 25 healthy subjects served as controls. Myocardial deformation was assessed by CMR based FT (TomTec Diogenes software), CMR based EBD (using custom written software) and STE (TomTec Cardiac Performance Analysis software).

Results: Feature tracking was feasible in all subjects. A close agreement was found between measures of global left (LV) and right ventricular (RV) global strain. Interobserver agreement for FT and STE was similar for longitudinal LV global strain, but FT showed better inter-observer reproducibility than STE for circumferential or radial LV and longitudinal RV global strain. Reproducibility of regional strain on FT was, however, poor. The relative systolic length change of the endocardial border measured by EBD yielded similar results to FT global strain. Clinically, biventricular longitudinal strain on FT was reduced compared to controls ($P < 0.0001$) and was related to the number of previous cardiac operations. In addition, FT derived RV strain was related to exercise capacity and VE/VCO₂-slope.

Conclusions: Although neither the inter-study reproducibility nor accuracy of FT software were investigated, and its inter-observer reproducibility for regional strain calculation was poor, its calculations of global systolic strain showed similar or better inter-observer reproducibility than those by STE, and could be applied across RV image regions inaccessible to echo. 'Global strain' calculated by EBD gave similar results to FT. . Measurements made using FT related to exercise tolerance in ToF patients suggesting that the approach could have clinical relevance and deserves further study.

Echocardiography. 2012 May 29. doi: 10.1111/j.1540-8175.2012.01721.x. [Epub ahead of print]

[Not All Obstructive Cardiac Lesions Are Created Equal: Double-Chamber Right Ventricle In Pregnancy.](#)

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

Source

Montefiore Medical Center, Albert Einstein College of Medicine, Department of Cardiology, Bronx, New York.

Abstract

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

(Echocardiography 2012;00:E1-E3).

J Cardiovasc Med (Hagerstown). 2012 May 17. [Epub ahead of print]

[The role of cardiac surgery in adult patients with congenital heart disease.](#)

Padalino MA, Vida VL, Rito ML, Daliento L, Stellin G.

Source

aPediatric and Congenital Cardiac Surgery Unit bDivision of Cardiology, Department of Cardiac, Thoracic and Vascular Sciences, University of Padua, Medical School, Italy.

Abstract

The number of adults with congenital heart disease (CHD) has constantly increased as medical and surgical treatment of CHD - either simple or complex - continues to improve. Over the past half century, advances in surgical techniques have continued with the evolution of traditional surgical repair and introduction of new surgical procedures for complex lesions which were previously considered to be irreparable. We sought to analyze the current role of cardiac surgery in the treatment of congenital anomalies of the heart in those patients who have reached adulthood with or without surgical repair or palliation, with particular attention to future directions and perspectives.

Am Heart J. 2012 May;163(5):859-66.

[Systemic right ventricular longitudinal strain is reduced in adults with transposition of the great arteries, relates to subpulmonary ventricular function, and predicts adverse clinical outcome.](#)

Diller GP, Radojevic J, Kempny A, Alonso-Gonzalez R, Emmanouil L, Orwat S, Swan L, Uebing A, Li W, Dimopoulos K, Gatzoulis MA, Baumgartner H.

Source

Adult Congenital Heart Disease Programme, Royal Brompton Hospital, London, United Kingdom; Adult Congenital and Valvular Heart Disease Center, Department of Cardiology and Angiology, University Hospital of Münster, Münster, Germany.

Abstract

Background: Transposition of the great arteries (TGA) after atrial switch operation and congenitally corrected TGA (ccTGA) are commonly associated with impaired systemic right ventricular (RV) function and impaired prognosis. We aimed to investigate the value of indices of myocardial deformation on speckle-tracking echocardiography for quantifying ventricular function and their potential role in assessing ventricular-ventricular interaction and outcome in patients with a systemic RV.

Methods and Results: A total of 129 patients (87 with TGA and atrial switch and 42 with ccTGA, 71 men, age 35 ± 12 years) were investigated, and biventricular myocardial deformation was compared with findings in healthy subjects ($n = 38$, age 36 ± 10 years). Systemic ventricular longitudinal 2-dimensional (2D) peak systolic strain (RV 2D-LS) was significantly reduced compared with controls (-12.9 ± 3.6 and -15.4 ± 5.1 vs -21.0 ± 5.5 in TGAs, ccTGAs, and controls, $P < .0001$). Systemic and pulmonary 2D-LS were correlated in patients with TGA ($r = 0.46$, $P < .0001$) and ccTGA ($r = 0.64$, $P < .0001$), suggesting interventricular interaction, and this was confirmed when ejection fraction on magnetic resonance imaging was assessed ($r = 0.53$, $P < .0001$). More importantly, systemic 2D-LS (hazard ratio 1.31, $P = .01$) was related to adverse clinical outcome (symptomatic progression to New York Heart Association class ≥ 3 , clinically relevant cardiac arrhythmia, or death) in patients with TGA and atrial switch independently of ejection fraction on cardiac magnetic resonance imaging, history of clinically relevant arrhythmia, or functional class.

Conclusions: Global longitudinal systolic strain is significantly reduced in patients with a systemic RV, is related to subpulmonary ventricular function, and predicts

adverse clinical outcome in adults with atrial switch TGA.

Eur J Cardiothorac Surg. 2012 May 15. [Epub ahead of print]

[Is alternative cardiac surgery an option in adults with congenital heart disease referred for thoracic organ transplantation?](#)

Harper AR, Crossland DS, Perri G, O'Sullivan JJ, Chaudhari MP, Schueler S, Griselli M, Hasan A.

Source

Department of Paediatric Cardiology, Freeman Hospital, High Heaton, Newcastle upon Tyne, UK.

Abstract

Objectives: We analysed the outcomes of adults with congenital heart disease (ACHD) referred for thoracic organ transplantation who underwent non-transplant cardiac surgery as an alternative management option.

Methods: Adult patients with congenital heart disease assessed for heart or heart-lung transplant were identified from the departmental database. A retrospective analysis of the medical records, transplant assessment data and surgical notes was carried out.

Results: One hundred and twenty-six patients were assessed between January 2000 and July 2011. Non-transplant cardiac surgery was performed in 14 (11%) patients. There were nine males with a median age of 37 years (range 21-42). The patients can be divided into four subgroups [left-sided lesions (n = 4), right-sided lesions (n = 3), systemic right ventricle (n = 5) and Fontan circulation (n = 2)]. Surgical procedures performed were: relief of systemic obstructive/regurgitant lesions ± endocardial fibroelastosis resection (n = 4, three pulmonary vascular resistance >6 Wood units), correction of right-sided regurgitant/stenotic lesions (n = 3), ventricular assist device for patients with a systemic right ventricle (n = 5) and re-fashioning of the Fontan pathway (n = 2). There were two early (5 and 30 days) and three late deaths (64, 232 and 374 days) with a 1-year mortality of 28%. None of the deaths occurred in patients with a two-ventricle circulation and atrio-ventricular concordance. Nine patients are alive at a median of 433 days (range 204-2456). The New York Heart Association class has improved in all survivors by at least one class at 3 and 6 months (P = 0.004 and 0.003).

Conclusions: Alternative cardiac surgery can be undertaken in selected patients with ACHD referred for cardiopulmonary transplantation with a low mortality in patients with two ventricles and a systemic left ventricle. Ventricular assist devices carry a significant mortality in patients with a systemic right ventricle, although this offers a valuable palliation when there are no other options. The medium and long-term results are awaited.

Cardiol Young. 2012 May 15:1-7. [Epub ahead of print]

[Adult congenital heart disease interventions: recommendations from a Joint Working Group of the British Congenital Cardiac Association, British Cardiovascular Intervention Society, and the British Cardiovascular Society.](#)

Qureshi SA, Hildick-Smith D, de Giovanni J, Clift P, Stuart G, Henderson R, Brecker S, Hackett D, Ray S, de Belder M.

Source

1Consultant Paediatric Cardiologist, Evelina Children's Hospital, Guy's & St Thomas Hospital Foundation Trust, London, UK.

Abstract

In order to optimise care of the adult patients with complex congenital heart disease, there is a need to develop recommendations for interventions. This document is the work of representatives of the three relevant societies and provides recommendations for institutions and operators performing cardiac interventions in these patients.

Int J Cardiovasc Imaging. 2012 May 12. [Epub ahead of print]

[Reduced global longitudinal and radial strain with normal left ventricular ejection fraction late after effective repair of aortic coarctation: a CMR feature tracking study.](#)

Kutty S, Rangamani S, Venkataraman J, Li L, Schuster A, Fletcher SE, Danford DA, Beerbaum P.

Source

Joint Division of Pediatric Cardiology, Children's Hospital and Medical Center, University of Nebraska College of Medicine/Creighton University School of Medicine, 8200, Dodge Street, Omaha, NE, 68114, USA, skutty@unmc.edu.

Abstract

We sought to determine whether global and regional left ventricular (LV) strain parameters were altered in repaired coarctation of the aorta (COA) with normal LV ejection fraction (EF) when compared with healthy adult controls, and whether such alterations were related to LV hypertrophy (LVH). We identified 81 patients after COA repair (31 female, age 25 ± 8.5 years) with inclusion criteria at follow-up CMR of: age ≥ 13 years, time post-repair ≥ 10 years, no aortic valve disease, LV-EF $> 50\%$. LV deformation indices derived using CMR-feature tracking and volumetric EF were compared between COA patients and normal controls ($n = 20$, 10 female, age 37 ± 7 years), and between COA with versus without LVH. In repaired COA versus controls, LV-EF (%) was 62 ± 7.2 versus 58 ± 3.0 ($p = 0.01$), and LV mass (g/m^2) 66 ± 16.8 versus 57.7 ± 6.0 ($p = 0.0001$). LV global longitudinal strain (GLS) was decreased to $-17.0 \pm 4.7\%$ in COA ($-20 \pm 5\%$ in controls, $p = 0.02$), and global radial strain (GRS) reduced to $40 \pm 15\%$ ($50 \pm 12.4\%$ in controls, $p = 0.003$). The global circumferential strain (GCS) was preserved in COA at $-23 \pm 4.7\%$ ($-24.6 \pm 2.4\%$ in controls, $p = 0.14$). Regionally, LS decrease was marked in the basal segments (septal, $p = 0.005$, lateral, $p = 0.013$). In COA with LVH ($n = 45$, mass $76.3 \pm 12.8 \text{ g}/\text{m}^2$) versus without LVH ($n = 36$, mass $52.2 \pm 10 \text{ g}/\text{m}^2$), GLS was more markedly decreased (-15.7 ± 4.8 vs. $-18.5 \pm 4.2\%$, $p = 0.016$, but GRS and GCS were similar ($p = 0.49$ and 0.27). In post-repair COA with normal LV-EF, GLS and GRS are reduced whilst GCS is preserved. GLS reduction is more pronounced in the presence of LVH. GLS may qualify as indicator of early LV dysfunction.

Heart Lung Circ. 2012 May 10. [Epub ahead of print]

[Does Pregnancy Contribute to Systemic Right Ventricular Dysfunction in Adults with an Atrial Switch Operation?](#)

Zentner D, Wheeler M, Grigg L.

Source

Department of Cardiology, The Royal Melbourne Hospital, Parkville, Vic 3050, Australia.

Abstract

Background: To determine whether pregnancy might impact adversely on long-term outcomes in adults post an atrial switch repair on the background of data demonstrating an increased rate of heart failure and death in these adults with systemic right ventricles.

Methods: We retrospectively analysed our adult population with an atrial switch repair for transposition of the great arteries to see whether any differences in outcomes (sudden cardiac death, heart failure admissions, use of heart failure medications) existed between women who had and women who had not undergone pregnancy. Controls from the remaining population (transposition of the great arteries and atrial switch operation women) were elected as long as their year of birth fell into the year of birth range seen in the patient group.

Results: In women with transposition of the great arteries who have had an atrial switch repair, the long-term occurrence of sudden cardiac death and clinical heart

failure (defined as a need for prescription of anti-failure medications or heart failure admissions) appears to be increased.

Conclusion: Pregnancy may have an adverse effect on long-term outcomes in women with systemic right ventricles.

Am J Cardiol. 2012 May 7. [Epub ahead of print]

[Acute and Long-Term Outcomes of Catheter Ablation Using Remote Magnetic Navigation in Patients With Congenital Heart Disease.](#)

Akca F, Bauernfeind T, Witsenburg M, Dabiri Abkenari L, Cuypers JA, Roos-Hesselink JW, de Groot NM, Jordaens L, Szili-Torok T.

Source

Clinical Electrophysiology, Department of Cardiology, Erasmus Medical Center, Rotterdam, The Netherlands.

Abstract

The aim of the present study was to assess the feasibility, safety, and long-term results of remote magnetic navigation in arrhythmias associated with complex congenital heart disease (CHD). The improved outcomes for CHD resulted in an increased number of complex arrhythmias requiring distinctive ablation techniques. Thirty-six patients with CHD (age 35 ± 19 years, 21 male) were divided into 3 complexity groups and underwent 43 radiofrequency catheter ablation procedures using the magnetic navigation system (including 7 redo ablations) in combination with the CARTO RMT system. A total of 59 tachyarrhythmias were identified. Most patients had surgical scar-related tachycardia (25 focal, including 4 microreentrant atrial tachycardia, and 27 macroreentrant atrial tachycardia). Four accessory pathways and three ventricular tachycardias were diagnosed and treated. In 31 patients, ablation was successful, with an end point of noninducibility (86%). The success rate for CHD complexity of type I, II, and III was 50%, 88%, and 89%, respectively. The mean procedure and fluoroscopy time was 216 ± 101 minutes and 40 ± 34 minutes, respectively. The number of radiofrequency applications was 42 ± 47 . No major complications related to the procedures occurred. Of the patients, 67% remained free of recurrence during a mean follow-up of 26 ± 4 months. Recurrence developed in 0%, 16%, and 45% of patients with CHD type I, II, and III, respectively. In conclusion, the magnetic navigation system is feasible to treat arrhythmias with reasonable success rates and good long-term outcomes in adult patients with CHD. The use of the magnetic navigation system offers advantages in complex anatomic situations.

Gen Thorac Cardiovasc Surg. 2012 Jun;60(6):341-4. Epub 2012 May 8.

[Pulmonary valve replacement long after repair of tetralogy of Fallot.](#)

Shiokawa Y, Sonoda H, Tanoue Y, Nishida T, Nakashima A, Tominaga R.

Source

Department of Cardiovascular Surgery, Kyushu University, 3-1-1 Maidashi, Higashiku, Fukuoka, 812-8582, Japan, s-yuchi@kyudai.jp.

Abstract

Purpose: Pulmonary valve replacement long after repair of tetralogy of Fallot can improve cardiac function, functional status, and arrhythmia propensity. This has not been reported in Japan. We aim to evaluate the effects of pulmonary valve replacement in repaired tetralogy of Fallot.

Methods: Nineteen patients underwent pulmonary valve replacement after repair of tetralogy of Fallot, excluding Rastelli type operation, between August 1981 and August 2011. The results of the pulmonary valve replacement were assessed by analyzing preoperative and postoperative cardiothoracic ratio, cardiac function, functional status, QRS duration and durability of the prosthetic valves.

Results: There were neither operative nor late deaths. The Cardiothoracic ratio significantly improved from 61.0 ± 5.2 % preoperatively to 56.2 ± 4.8 %

postoperatively ($P < 0.001$). The New York Heart association functional class significantly improved from 2.4 ± 0.8 preoperatively to 1.2 ± 0.4 postoperatively as well. Left ventricular ejection fraction showed significant improvement. QRS duration showed significant reduction. The freedom redo pulmonary valve replacement at 20 years was 100 %.

Conclusion: Pulmonary valve replacement long after repair of previous tetralogy of Fallot had clinical benefits with low mortality. We recommend bioprosthesis for pulmonary valve replacement when adult-sized valve can be accommodated.

J Card Fail. 2012 May;18(5):379-84. Epub 2012 Mar 10.

[Long-term effect of bosentan therapy on cardiac function and symptomatic benefits in adult patients with Eisenmenger syndrome.](#)

Kaya MG, Lam YY, Erer B, Ayhan S, Vatankulu MA, Nurkalem Z, Meric M, Eren M, Eryol NK.

Source

Department of Cardiology, Erciyes University Medicine School, Kayseri, Turkey.

Abstract

Background: Bosentan improves symptoms in patients with Eisenmenger syndrome (ES). This study evaluated the effect of long-term bosentan therapy on cardiac function and its relation to symptomatic benefits in ES patients.

Methods and Results: Twenty-three consecutive adult ES patients (15 with ventricular septal defect, 6 with atrial septal defect, and 2 with patent ductus arteriosus) underwent standard and tissue Doppler echocardiography before and 24 ± 9 months after bosentan therapy. Echocardiographic measurements included pulmonary arterial systolic pressure (PASP), myocardial performance index (MPI), tricuspid and lateral mitral annular pulsed-wave tissue Doppler systolic (Sa) and early diastolic (Ea) long-axis motions. Patients' World Health Organization (WHO) functional class, 6-minute walk distance (6MWD), and systemic arterial oxygen saturations (SaO₂) were also recorded. The PASP, WHO functional class, 6MWD, and SaO₂ all improved (118 ± 22 to 111 ± 19 mm Hg, 3.2 ± 0.4 to 2.4 ± 0.5 , 286 ± 129 m to 395 ± 120 m, and $84.6 \pm 6.5\%$ to $88.8 \pm 3.9\%$, respectively; all $P < .01$) after therapy. There was also significant improvement in right ventricular (RV) MPI (by 23.9%: 0.46 ± 0.15 to 0.35 ± 0.09) and biventricular long-axis function (tricuspid Sa and Ea: 6.7 ± 1.5 to 8.8 ± 1.7 cm/s and 5.7 ± 1.3 to 7.0 ± 1.2 cm/s, respectively; lateral Sa and Ea: 6.8 ± 1.3 to 8.4 ± 1.5 cm/s and 7.6 ± 2.0 to 8.5 ± 2.1 cm/s, respectively; all $P < .05$). Posttherapy RV MPI was moderately correlated with PASP and 6MWD.

Conclusions: Sustained improvement of pulmonary arterial hypertension and RV function in ES patients was evident 2 years after bosentan therapy, and this may provide insights on the symptomatic benefits gained in these patients.

Cardiol Clin. 2012 May;30(2):205-18.

[Right ventricular performance in congenital heart disease: a physiologic and pathophysiologic perspective.](#)

Hopkins WE.

Source

Department of Medicine and Cardiology Unit, Pulmonary Hypertension and Adult Congenital Heart Disease Programs, Fletcher Allen Health Care, University of Vermont College of Medicine, McClure 1, MCHV Campus, 111 Colchester Avenue, Burlington, VT 05401, USA. william.hopkins@vtmednet.org

Abstract

Underappreciated is the fact that the right ventricle is often the primary determinant of long-term morbidity and mortality in patients with congenital heart disease. Right ventricular performance in these patients depends on a unique set of physiologic and pathophysiologic factors that are rarely considered in acquired heart disease. This article explores this unique physiology and pathophysiology in

the hope that it will enhance understanding of a wide variety of congenital cardiac anomalies.

J Thorac Cardiovasc Surg. 2012 May;143(5):1183-1192.e4.

[Outcomes after transplantation for "failed" Fontan: a single-institution experience.](#)

Davies RR, Sorabella RA, Yang J, Mosca RS, Chen JM, Quaegebeur JM.

Source

Nemours Cardiac Center, A.I. duPont Hospital for Children, 1600 Rockland Rd, Wilmington, DE 19803, USA. rdavies@nemours.org

Abstract

Objective: Despite the excellent outcomes in the current era after the Fontan procedure, it continues to have an inherent risk of failure. Cardiac transplantation provides 1 option for treating these patients; however, the indications for, timing of, and outcomes after, transplantation remain undefined. We examined our own institutional experience with transplantation for failed Fontan.

Methods: The records of 155 patients transplanted for congenital heart disease at a single institution from June 1984 to September 2007 were reviewed. Of these patients, 43 had undergone a previous Fontan procedure (25 male, 15 female; median age, 14.5 years; range, 1-47; 23 classic Fontan, 13 lateral tunnel, 4 extracardiac conduit, and 3 revised to shunt). The predictors of short- and long-term survival were evaluated, and the Fontan patients were compared with all other patients with congenital heart disease (n = 129, 78 male, 51 female).

Results: The most common indications for transplantation included protein-losing enteropathy (PLE) (39.5%), chronic heart failure (41.8%), and acute post-Fontan failure (9.3%). The transplants performed in Fontan patients were more likely to require pulmonary artery reconstruction (85.4% vs 42.9%; P < .0001) and had longer cardiopulmonary bypass times (278 vs 179 minutes; P < .0001). The 90-day mortality rate was greater in the Fontan group (35.0% vs 20.0%; P = .055). No correlation was observed between the interval from Fontan to transplantation and mortality; however, renal failure was a strong predictor of early mortality (odds ratio, 10.8; 95% confidence interval, 1.5-75.7).

Conclusions: Transplantation is an acceptable treatment for patients with a failed Fontan. Clinical factors (instead of the indication for transplantation) appear to have the greatest correlation with early mortality.

Circulation. 2012 May 22;125(20):2440-6. Epub 2012 Apr 11.

[Left ventricular longitudinal function predicts life-threatening ventricular arrhythmia and death in adults with repaired tetralogy of fallot.](#)

Diller GP, Kempny A, Liodakis E, Alonso-Gonzalez R, Inuzuka R, Uebing A, Orwat S, Dimopoulos K, Swan L, Li W, Gatzoulis MA, Baumgartner H.

Source

Adult Congenital and Valvular Heart Disease Center, Department of Cardiology and Angiology, University Hospital of Münster, Albert-Schweitzer-Strasse 33, 48149 Muenster, Germany. gerhard.diller@gmail.com.

Abstract

Background: Sudden cardiac death and life-threatening ventricular arrhythmia remain a concern in adult patients with repaired tetralogy of Fallot. Longitudinal left ventricular (LV) function is sensitive in detecting early myocardial damage and may have prognostic implications in this setting.

Methods and Results: We included 413 tetralogy of Fallot patients (age, 36±13 years; QRS duration, 148±27 milliseconds; LV ejection fraction, 55±10%). A composite end point of sudden cardiac death/life-threatening ventricular arrhythmia (sustained ventricular tachycardia, resuscitated sudden cardiac death, or appropriate implantable cardioverter-defibrillator discharge) was used. During a median follow-up of 2.9 years, 5 patients died suddenly, 9 had documented sustained ventricular tachycardia, and another 5 had appropriate implantable

cardioverter-defibrillator shocks. On univariate Cox analysis, QRS duration (hazard ratio [HR], 1.02 per 1 ms; P=0.046), right atrial area (HR, 1.05 per 1 cm²; P=0.02), right ventricular fractional area change (HR, 0.94 per 1%; P=0.02), right ventricular outflow tract diameter (HR, 1.08 per 1 mm; P=0.01), mitral annular plane systolic excursion (HR, 0.84 per 1 mm; P=0.03), and LV global longitudinal 2-dimensional strain (HR, 0.87 per 1%; P=0.03) were related to the combined end point. On bivariable analysis, mitral annular plane systolic excursion and LV global longitudinal 2-dimensional strain were related to outcome independently of QRS duration (P=0.002 and P=0.01, respectively). In addition, a combination of echocardiographic variables, including right atrial area, right ventricular fractional area change, and LV global longitudinal 2-dimensional strain or mitral annular plane systolic excursion, was also found to be significantly related to outcome (P<0.001; c statistic, 0.70).

Conclusions: LV longitudinal dysfunction was associated with greater risk of sudden cardiac death/life-threatening ventricular arrhythmias. In combination with echocardiographic right heart variables, also available from routine echocardiography, these measures provide important outcome information and should be considered a useful adjunct to established markers such as QRS duration in the estimation of prognosis in this challenging population.

Am J Med Genet A. 2012 May;158A(5):982-8. doi: 10.1002/ajmg.a.35343. Epub 2012 Mar 27.

[Diagnostic yield in adults screened at the Marfan outpatient clinic using the 1996 and 2010 Ghent nosologies.](#)

Aalberts JJ, Thio CH, Schuurman AG, van Langen IM, van der Pol BA, van Tintelen JP, van den Berg MP.

Source

Department of Cardiology, University Medical Centre Groningen, University of Groningen, The Netherlands. jjj_aalberts@hotmail.com

Abstract

Marfan syndrome (MFS) is diagnosed according to the Ghent nosology, which has recently been revised. In the Netherlands, evaluation for possible MFS is performed in specialized Marfan outpatient clinics. We investigated the diagnostic yield in our clinic and the impact of the 2010 nosology. All adult patients (n = 343) who visited our clinic between 1998 and 2008 were included. We analyzed their reasons for referral, characteristics, and established diagnoses. In addition, we applied the 2010 nosology to all patients and compared the outcomes to those obtained with the 1996 nosology. Diagnoses that were made using the 1996 and the 2010 Ghent nosology included MFS (44/343 vs. 47/343), familial thoracic aortic aneurysm and/or dissection (22/343 vs. 22/343 patients), Loeys-Dietz syndrome (4/343 vs. 4/343 patients), and (familial) mitral valve prolapse (MVPS; 5/343 vs. 28/343 patients). In both nosologies, 77% of MFS patients had an FBN1 mutation. The 2010 nosology led to an increase in the number of diagnoses made: 4 additional cases of MFS were identified (one patient was "lost" who no longer fulfilled the criteria) and 23 additional cases of MVPS were diagnosed. The diagnostic yield of patients with aortic root dilatation was 65% using the 1996 nosology and 70% using the 2010 nosology. The change in diagnoses did not lead to a difference in clinical follow-up. We conclude that the diagnostic yield of our specialized clinic was high, in particular in patients with aortic root dilatation. Furthermore the 2010 Ghent nosology led to a significant increase in the number of diagnoses made, mainly due to lowering of the diagnostic threshold for MVPS.

Heart. 2012 May;98(9):736-42. Epub 2012 Mar 7.

[B-type natriuretic peptide concentrations in contemporary Eisenmenger syndrome patients: predictive value and response to disease targeting therapy.](#)

Diller GP, Alonso-Gonzalez R, Kempny A, Dimopoulos K, Inuzuka R,

Giannakoulas G, Castle L, Lammers AE, Hooper J, Uebing A, Swan L, Gatzoulis M, Wort SJ.

Source

Adult Congenital Heart Disease Centre and, National Centre for Pulmonary Hypertension, Royal Brompton Hospital, Sydney Street, London SW3 6NP, UK. g.diller@imperial.ac.uk

Abstract

Objective: To assess the relationship between elevated levels of B-type natriuretic peptide (BNP) and outcome in patients with Eisenmenger syndrome.

Design: Retrospective study.

Setting: Tertiary centre for adult congenital heart disease.

Patients: All patients with Eisenmenger syndrome (n=181, age 36.9±12.1 years, 31% with Down syndrome) in whom BNP concentrations were measured as part of routine clinical care were included.

Main Outcome and Measures: The study end point was all cause mortality.

Results: During a median follow-up period of 3.3 years, 20 patients (7 with Down syndrome) died. Higher BNP concentrations were predictive of all cause mortality on univariate analysis in patients with or without Down syndrome. On multivariable Cox proportional hazard analysis, BNP predicted survival independently of renal function, Down syndrome, or 6 min walk test distance (p=0.004). Temporal increases in BNP concentration were also found to predict mortality. Treatment with disease targeting therapies was associated with a significant reduction in BNP concentrations.

Conclusions: BNP concentrations predict outcome in contemporary Eisenmenger patients. Increases in BNP concentrations over time are also of prognostic significance. In addition, disease targeting therapies may help to reduce BNP concentrations in this population, while treatment-naïve patients have static or rising BNP concentrations.

Congenit Heart Dis. 2012 May;7(3):243-9. doi: 10.1111/j.1747-0803.2012.00635.x. Epub 2012 Feb 20.

[The safety and effects of bosentan in patients with a fontan circulation.](#)

Bowater SE, Weaver RA, Thorne SA, Clift PF.

Source

University Hospital Birmingham Foundation NHS Trust, Birmingham, United Kingdom.

Abstract

Objective: Adult patients with a Fontan circulation tend to have diminished exercise capacity. The principal objective of this study was to investigate the safety of the endothelin receptor antagonist bosentan in Fontan patients, and, secondarily, to assess effects on cardiovascular performance, New York Heart Association functional classification (NYHA FC), and ventricular function.

Design: A 6-month prospective, single-center, pilot, safety study of bosentan in Fontan patients. Setting. Adult Congenital Heart Disease referral center.

Patients: All patients ≥18 years old with a Fontan circulation and in NYHA FC ≥II were invited to enroll. Interventions. Patients started on 62.5mg bid of bosentan, uptitrating to 125mg bid after 2 weeks.

Outcome Measures: Safety was assessed by the incidence of anticipated and unanticipated adverse events during the 6-month study period; specifically those relating to hepatic, renal, or hematological dysfunction as measured by monthly blood tests. Other outcome measures included cardiopulmonary exercise test, 6-minute walk distance test, Borg dyspnea index, NYHA FC, and ventricular function parameters using transthoracic echocardiography.

Results: Of the eight patients enrolled, six completed the study. Two patients withdrew from the study (one for non-trial related reasons, one due to adverse events). No clinically significant adverse events relating to bosentan therapy

occurred during this study and, in particular, no significant abnormalities in hepatic function tests were observed. Three patients reported transient adverse events. Improvements in NYHA FC and systolic ventricular function were observed after 6 months of bosentan treatment.

Conclusions: The small number of patients with a Fontan circulation in our study was able to tolerate bosentan for 6 months. The safety and tolerability of bosentan in a larger patient population remains unknown. The results presented here justify further investigation in larger studies.

Am J Med Genet A. 2012 May;158A(5):989-95. doi: 10.1002/ajmg.a.34392. Epub 2011 Dec 7.

[A comparison of the Ghent and revised Ghent nosologies for the diagnosis of Marfan syndrome in an adult Korean population.](#)

Yang JH, Han H, Jang SY, Moon JR, Sung K, Chung TY, Lee HJ, Ki CS, Kim DK.

Source

Department of Medicine, Cardiac and Vascular Center, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea.

Abstract

Recently, a revised Ghent nosology has been established for the diagnosis of Marfan syndrome (MFS) that puts more weight on the aortic root aneurysm and ectopia lentis. We compared the application of the Ghent and revised Ghent nosologies in adult Korean patients for whom there is suspicion of MFS. From January 1995 to June 2010, we enrolled 106 patients older than 20 years for whom there was suspicion of MFS, and who had undergone genetic analysis of the fibrillin-1 gene (FBN1). Of 106 patients, 86 patients (81%) fulfilled the criteria of the Ghent nosology, and 84 patients (79%) met the criteria of the revised Ghent nosology. The two patients who met the Ghent nosology criteria, but not the criteria of the revised Ghent nosology were diagnosed with Loeys-Dietz syndrome and MASS phenotype. The level of agreement between both nosologies was very high ($\kappa = 0.94$, 95% confidence interval: 0.86 to 1.0). Marfan-like syndromes were diagnosed in 30% (6/20 patients) with negative Ghent and revised Ghent criteria and no FBN1 mutations. These results suggest that adult Korean patients who fulfill the old Ghent criteria almost all fulfill the new criteria for the diagnosis of MFS.

J Thorac Cardiovasc Surg. 2012 May;143(5):1103-7. Epub 2011 Nov 4.

[Long-term importance of right ventricular outflow tract patch function in patients with pulmonary regurgitation.](#)

Puranik R, Tsang V, Lurz P, Muthurangu V, Offen S, Frigiola A, Norman W, Walker F, Bonhoeffer P, Taylor AM.

Source

Centre for Cardiovascular Imaging, UCL Institute of Cardiovascular Science, Great Ormond Street Hospital for Children, London, United Kingdom.

Abstract

Objective: Chronic pulmonary regurgitation (PR) has deleterious effects on right ventricular (RV) function in repaired tetralogy of Fallot (ToF). However, there are little data regarding right ventricular outflow tract (RVOT) contractile dysfunction in response to chronic PR and on both RV and LV volumes and function.

Methods: We retrospectively identified consecutive patients with PR who were referred for magnetic resonance imaging quantification of "free PR" detected on echocardiography between 2003 and 2008. Patients had ToF and a transannular patch procedure ($n = 30$, 25.1 ± 1.2 years) or PR resulting from valvar pulmonary stenosis treated with surgical or percutaneous valvotomy ($n = 30$, 26.6 ± 1.8 years).

Results: The ToF and the PS groups were well matched for age at scan, age at repair surgery in ToF or initial valvotomy in PS, duration of exposure to PR, body

surface area, heart rate, PR fraction, net forward pulmonary artery flow, and main and branch pulmonary artery dimensions. Severe PR fractions were identified in both groups (ToF: $40\% \pm 1\%$ vs PS: $37\% \pm 2\%$, $P = .2$). Indexed RV and LV end-diastolic volumes were similar for both ToF and PS groups (RV end-diastolic volume index: 137 ± 6 mL/m²) vs 128 ± 5 mL/m², $P = .2$, and LV end-diastolic volume index: 72 ± 2 mL/m²) vs 67 ± 2 mL/m², $P = .1$, respectively). RV mass was also similar between groups (95 ± 5 g vs 81 ± 6 g, respectively, $P = .08$). However, indexed RV and LV end-systolic volumes were consistently higher in ToF when compared with PS (RV end-systolic volume index: 70 ± 5 mL/m²) vs 54 ± 3 mL/m², $P < .01$, and LV end-systolic volume index: 29 ± 1 mL/m²) vs 22 ± 1 mL/m², $P < .01$, respectively). These changes were reflected in lower biventricular systolic function in patients with ToF when compared with PS (RV ejection fraction: $52\% \pm 1.5\%$ vs $59\% \pm 1\%$, $P < .001$, and LV ejection fraction: $61\% \pm 1\%$ vs $67 \pm 1\%$, $P < .001$, respectively). Although RV transannular plane systolic excursion was not significantly different between the groups ($P = .86$), the RV outflow tract was considered contractile in only 50% of patients with ToF compared with 93% of patients with PS ($P = .0004$). RV volumes and function were similar when only patients with contractile RV outflow tracts were compared. **Conclusions:** RV outflow tract patch dysfunction in repaired ToF is responsible for higher end-systolic volumes and thus lower global measures of ventricular systolic function. These findings were not evident in cases of PS treated with valvotomy with comparable amount of PR. These observations highlight the importance of the initial repair surgery in ToF for late outcomes.

J Adv Nurs. 2012 May;68(5):1050-60. doi: 10.1111/j.1365-2648.2011.05809.x. Epub 2011 Aug 16.

[Education and support needs of the older adult with congenital heart disease.](#)

Riley JP, Habibi H, Banya W, Gatzoulis MA, Lau-Walker M, Cowie MR.

Source

Royal Brompton & Harefield NHS Foundation Trust, London, UK.

jillian.riley@imperial.ac.uk

Abstract

Aim: This article is a report of a study exploring health-related quality of life in adults with congenital heart disease and the extent to which it is associated with patients' illness beliefs and emotional health.

Background: A reduction in mortality in patients with congenital heart disease has led to an increasingly older population that faces new challenges. Studies in a younger adult population have reported inconsistent findings regarding health-related quality of life. Factors such as, the complexity of the congenital heart defect, have not been found to be associated with quality of life. The association between illness beliefs and health-related quality of life has not previously been reported.

Method: A cross-sectional questionnaire study of adults with congenital heart disease attending an outpatient clinic in a specialist centre in the United Kingdom between October 2007 and May 2008.

Results: The mean age of the study population was 37.2 years. Participants reported poorer physical functioning, role functioning and general health than a general population. High levels of anxiety were reported in 38% and high levels of depression in 17%. In multivariate analysis, higher levels of anxiety and depression were associated with poorer mental functioning and higher levels of depression with poorer physical quality of life.

Conclusion: We have reported that high levels of anxiety and depression in an older population of patients with congenital heart disease are associated with poorer quality of life. This highlights the need to routinely assess anxiety and depression in this patient group and to provide psychological support appropriately.

Int J Cardiol. 2012 May 17;157(1):70-4. Epub 2010 Dec 28.

[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, Burggraf 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.

Source

University of Ottawa Heart Institute, Canada. lbeauchesne@ottawaheart.ca

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 ± 2 months, and 4 ± 3 months 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. 2012 May 17;157(1):59-62. Epub 2010 Dec 21.

[Factors associated with surgery for active endocarditis in congenital heart disease.](#)

Murakami T, Niwa K, Yoshinaga M, Nakazawa M.

Source

Department of Adult Congenital Heart Disease and Pediatrics, Chiba Cardiovascular Center, Ichihara City, Japan. murat@seagreen.ocn.ne.jp

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.

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