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ISACHD Newsletter

ISACHD to Meet in New Orleans

Meeting and Scientific Program
Studio 2, Hotel New Orleans Marriott
555 Canal Street, New Orleans
Sunday, April 3, 2011
6:30-8:30 pm



Agenda

Welcome- *Barbara Mulder*

New Strategic Plans

In the executive committee there is consensus that ISACHD needs a new strategic plan, in order to guide future directions. It is important that ISACHD addresses issues that are not covered by other organisations. Furthermore, the international component is pivotal. The following key elements for ISACHD's mission are proposed:

- Global Health Care- *Curt Daniels*
- Education- *Erwin Oechslin*
- Research and Journal Watch- *Koichiro Niwa, Philip Moons, Barbara Mulder*

Endorsement of symposia & congresses- *Erwin Oechslin*

Regional + Nursing News- *Bill Davidson, Koichiro Niwa, Erwin Oechslin, Luis Alday, Helmut*

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Upcoming Meetings

- GUCH Training Course, Paris, March 4th-5th, 2011
- 21st ACHD Symposium, Cincinnati, June 19-22, 2011
- ESC Congress, Paris, August 27th-31st, 2011
- AHA, Orlando, November 2011
- 3rd Congress Asia Pacific Society for Adult CHD, Taipei, Taiwan, April 2012

Other Business

ACC ACHD

Sunday, April 3, 2011

Ernest N. Morial Convention Center

[Click here to see the Agenda!](#)

Journal Watch Commentary

ARTICLE OF THE MONTH: February 2011

Commentary on paper 'Seeking optimal relation between oxygen saturation and haemoglobin concentration in adults with cyanosis from congenital heart disease' by Anitra Romfh

In the care of patients with cyanotic congenital heart disease, the definition of anemia is difficult, as the physiologic adaptation to hypoxemia involves erythrocytosis, right-shifting of the hemoglobin dissociation curve, and changing cardiac index[1, 2]. The erythrocytic response to chronic hypoxemia is important for maintenance of systemic oxygen transport[2]. Patients can either suffer from too little of a response, resulting in a relative anemia, or too brisk of a response resulting in hyperviscosity[3]. Thus, a tool to predict the expected values of hemoglobin for a given oxygen saturation would be useful. A relationship for children and adolescents with cyanotic heart disease has been explored by Gidding et al, after exclusion of factors that could destabilize systemic oxygen transport, such as diminished iron stores[3]. A less steep relationship between hemoglobin and oxygen saturation has previously been reported for iron replete patients with Eisenmenger syndrome[4]

This descriptive cross-sectional study prospectively sought to include adults with congenital heart disease with right-to-left shunts. Patients exhibited a wide range of oxygen saturations. The primary objective of this study was to establish the optimal hemoglobin for adults with cyanotic congenital heart disease, employing stringent criteria to exclude patients with evidence of inadequate erythropoiesis. The exclusion criteria were the presence of any of the following criteria: evidence of iron deficiency, vitamin B12 or folate deficiency, elevated serum erythropoietin, reticulocytosis, hypochromia, or a significant rightward shift of the oxygen-hemoglobin dissociation curve. In addition, patients with recent acute hospitalization, therapeutic phlebotomy, differential cyanosis, and use of supplemental oxygen were excluded. The authors were able to show a strong linear correlation between the optimal

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hemoglobin and oxygen saturation with the regression equation showing the optimal hemoglobin equal to $57.5 - (0.444 \times \text{oxygen saturation})$. For example, a patient with an oxygen saturation of 85% would be expected to have a hemoglobin of 19.7. Because of the rigorous exclusion criteria employed in this study, the slope of this relationship was steeper and demonstrated a better linear relationship than in prior studies[3, 5]. The authors also went on to provide a functional context by taking those patients at an optimal hemoglobin and performing a 6-minute walk test. The 6-minute walk test results of the optimal group were then compared to the non-optimal group. The greater the deviation was from the optimal hemoglobin, the poorer the performance on the 6-minute walk test [5].

This study provides a significant contribution to the care of cyanotic adult congenital patients, providing the clinician with a tool for assessing the adequacy of physiologic compensation given an oxygen saturation. This is particularly useful for patients in a balanced state of erythropoiesis. Not surprisingly, patients with an oxygen saturation less than 75% had non-optimal hemoglobin values. Previous studies have shown these patients to have elevated erythropoietin titers, suggesting the presence of a threshold saturation level below which many patients cannot mount an adequate erythrocytic response[3, 6]. Thus, while it was not possible predict the optimal hemoglobin for these sicker patients, it may be enough to recognize their likely state of unbalanced erythropoiesis[5]. Through its stringent exclusion criteria, the study also alerts the clinician to previously established factors that alter the expected relationship between hemoglobin and oxygen saturation. Knowledge of these criteria aids in identifying factors that may be limiting erythrocytic compensation. Future studies may address ways to bring patients at non-optimal hemoglobin values into the optimal range[7].

- 1 Gidding SS, Stockman JA, 3rd: Effect of iron deficiency on tissue oxygen delivery in cyanotic congenital heart disease. *Am J Cardiol* 1988;61:605-607.
- 2 Berman W, Jr., Wood SC, Yabek SM, Dillon T, Fripp RR, Burstein R: Systemic oxygen transport in patients with congenital heart disease. *Circulation* 1987;75:360-368.
- 3 Gidding SS, Bessel M, Liao YL: Determinants of hemoglobin concentration in cyanotic heart disease. *Pediatr Cardiol* 1990;11:121-125.
- 4 Diller GP, Dimopoulos K, Broberg CS, Kaya MG, Naghotra US, Uebing A, Harries C, Goktekin O, Gibbs JS, Gatzoulis MA: Presentation, survival prospects, and predictors of death in Eisenmenger syndrome: A combined retrospective and case-control study. *Eur Heart J* 2006;27:1737-1742.
- 5 Broberg CS, Jayaweera AR, Diller GP, Prasad SK, Thein SL, Bax BE, Burman J, Gatzoulis MA: Seeking optimal relation between oxygen saturation and hemoglobin concentration in adults with cyanosis from congenital heart disease. *Am J Cardiol* 2011;107:595-599.

6 Tyndall MR, Teitel DF, Lutin WA, Clemons GK, Dallman PR: Serum erythropoietin levels in patients with congenital heart disease. *J Pediatr* 1987;110:538-544.

7 Tay EL, Peset A, Papaphylactou M, Inuzuka R, Alonso-Gonzalez R, Giannakoulas G, Tzifa A, Goletto S, Broberg C, Dimopoulos K, Gatzoulis MA: Replacement therapy for iron deficiency improves exercise capacity and quality of life in patients with cyanotic congenital heart disease and/or the Eisenmenger syndrome. *Int J Cardiol*

Article: Broberg CS, Jayaweera AR, Diller GP, Prasad SK, Thein SL, Bax BE, Burman J, Gatzoulis MA: Seeking optimal relation between oxygen saturation and hemoglobin concentration in adults with cyanosis from congenital heart disease. *Am J Cardiol* 2011;107:595-599.

Commentary: Anitra Romfh, M.D.
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Journal Watch

Am J Cardiol. 2011 Feb 22. [Epub ahead of print]

[Prevalence of Left Ventricular Systolic Dysfunction in Adults With Repaired Tetralogy of Fallot.](#)

[Broberg CS, Aboulhosn J, Mongeon FP, Kay J, Valente AM, Khairy P, Earing MG, Opatowsky AR, Lui G, Gersony DR, Cook S, Ting JG, Webb G, Gurvitz MZ; Alliance for Adult Research in Congenital Cardiology \(AARCC\).](#)

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Abstract

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

Clin Genet. 2011 Feb 17. doi: 10.0000/j.1399-0004.2011.01646.x. [Epub ahead of

print]

[Critical Appraisal of the Revised Ghent Criteria for Diagnosis of Marfan Syndrome.](#)

Radonic T, de Witte P, Groenink M, de Bruin-Bon RA, Timmermans J, Scholte AJ, van den Berg MP, Baars MJ, van Tintelen JP, Kempers M, Zwinderman AH, Mulder BJ.

Department of Cardiology, Academic Medical Center Amsterdam, the Netherlands

Abstract

Marfan syndrome (MFS) is a connective tissue disorder with major features in cardiovascular, ocular and skeletal systems. Recently, diagnostic criteria were revised where more weight was given to the aortic root dilatation. We applied the revised Marfan nosology in an established adult Marfan population to define practical repercussions of novel criteria for clinical practice and individual patients. Out of 180 MFS patients, in 91% (n=164) the diagnosis of MFS remained. Out of 16 patients with rejected diagnosis, four patients were diagnosed as MASS phenotype, three as ectopia lentis syndrome and in nine patients no alternative diagnosis was established. In 13 patients the diagnosis was rejected because the Z-score of the aortic root was <2, although the aortic diameter was larger than 40 mm in six of them. In three other patients the diagnosis of MFS was rejected because dural ectasia was given less weight in the revised nosology. Following the revised Marfan nosology, the diagnosis of MFS was rejected in 9% of patients, mostly due to absence of aortic root dilatation defined as Z-score ≥ 2 . Currently used Z-scores seem to underestimate aortic root dilatation, especially in patients with large BSA. We recommend re-evaluation of criteria for aortic root involvement in adult patients with a suspected diagnosis of MFS.

Int J Cardiol. 2011 Feb 9. [Epub ahead of print]

[Post-Interventional three-dimensional dark blood MRI in the adult with congenital heart disease.](#)

Winner MW 3rd, Raman SV, Chung YC, Simonetti OP, Mihai G, Cook SC.

Department of Internal Medicine, Division of Cardiovascular Medicine, Adolescent/Adult Congenital Heart Disease Program, The Ross Heart Hospital, The Ohio State University, United States.

Abstract

BACKGROUND: Investigate a novel three-dimensional (3D) turbo spin echo (TSE) magnetic resonance imaging (MRI) sequence to assess stented segments in adults with congenital heart disease (CHD) after transcatheter intervention.

METHODS: Adults with CHD referred for computed tomography (CT) after transcatheter intervention underwent MR exam with a 3D respiratory gated TSE sequence. Data obtained at the time of the study included type of CHD, radiation dose, length of time between exams, and luminal diameters of stented segments from each exam. Continuous variables were analyzed using Student's t and Bland-Altman plots performed to analyze measurements obtained from both examinations.

RESULTS: Eleven patients underwent both examinations. Type of defects included coarctation of the aorta (n=6) and tetralogy of Fallot. Average radiation dose was 19.6mSv and average time between CT and MRI was 99±160days. Luminal diameters of stented vessels correlated closely between TSE MRI and CT ($r(2)=-.85$) with a bias toward overestimation with MRI (mean 22.4±4.3mm and 20.9±3.7mm, $p<.01$).

CONCLUSION: This novel 3D respiratory gated TSE MR technique provides a feasible method to reduce metallic artifact and improve visualization of stented segments and surrounding anatomic structures without exposure to radiation.

Am Heart J. 2011 Feb;161(2):233-40

[Lessons learned from a pediatric clinical trial: The Pediatric Heart Network Angiotensin-Converting Enzyme Inhibition in Mitral Regurgitation Study.](#)

[Li JS, Colan SD, Sleeper LA, Newburger JW, Pemberton VL, Atz AM, Cohen MS, Golding F, Klein GL, Lacro RV, Radojewski E, Richmond ME, Minich LL.](#)

Duke University Medical Center, Durham, NC.

Abstract

BACKGROUND: Mitral regurgitation is the most common indication for reoperation in children following repair of atrioventricular septal defect (AVSD). We hypothesized that angiotensin-converting enzyme inhibitor therapy would decrease the severity of mitral regurgitation and limit left ventricular volume overload in children following AVSD repair. METHODS: The Pediatric Heart Network designed a placebo-controlled randomized trial of enalapril in this population. The primary aim was to test the effect of enalapril on the change in left ventricular end-diastolic dimension body surface area-adjusted z score. Before the launch of the trial, a feasibility study was performed to estimate the number of patients with at least moderate mitral regurgitation following AVSD repair. TRIAL EXPERIENCE: Seventeen months after the start of the study, 349 patients were screened, 8 were trial eligible, and only 5 were enrolled. The study was subsequently terminated because of low patient accrual. Several factors led to the problems with patient accrual, including (1) the use of criteria to assess disease severity in the feasibility study that were not identical to those used in the trial, (2) failure to achieve equipoise for the study among clinicians and referring physicians, (3) reliance on methodology developed in adult populations with different disease mechanisms, and (4) absence of adequate data to define the natural history of the disease process under study. Progress in the treatment of children with cardiovascular disease will depend on the future of multicenter collaborative clinical trials. The lessons learned from this study may contribute to improvements in this research.

Cardiol Young. 2011 Feb 11:1-8. [Epub ahead of print]

[Report from The International Society for Nomenclature of Paediatric and Congenital Heart Disease: cardiovascular catheterisation for congenital and paediatric cardiac disease \(Part 1 - Procedural nomenclature\).](#)

Bergersen L, Everett AD, Giroud JM, Martin GR, Franklin RC, Beland MJ, Krogmann ON, Aiello VD, Colan SD, Elliott MJ, Gaynor JW, Kurosawa H, Maruszewski

Department of Cardiology, Children's Hospital Boston, Boston, Massachusetts, United States of America.

Abstract

Interventional cardiology for paediatric and congenital cardiac disease is a relatively young and rapidly evolving field. As the profession begins to establish multi-institutional databases, a universal system of nomenclature is necessary for the field of interventional cardiology for paediatric and congenital cardiac disease. The purpose of this paper is to present the results of the efforts of The International Society for Nomenclature of Paediatric and Congenital Heart Disease to establish a system of nomenclature for cardiovascular catheterisation for congenital and paediatric cardiac disease, focusing both on procedural nomenclature and on the

nomenclature of complications associated with interventional cardiology. This system of nomenclature for cardiovascular catheterisation for congenital and paediatric cardiac disease is a component of The International Paediatric and Congenital Cardiac Code. This manuscript is the first part of a two-part series. Part 1 will cover the procedural nomenclature associated with interventional cardiology as treatment for paediatric and congenital cardiac disease. This procedural nomenclature of The International Paediatric and Congenital Cardiac Code will be used in the IMPACT Registry™ (IMproving Pediatric and Adult Congenital Treatment) of the National Cardiovascular Data Registry® of The American College of Cardiology. Part 2 will cover the nomenclature of complications associated with interventional cardiology as treatment for paediatric and congenital cardiac disease.

Am J Cardiol. 2011 Feb 15;107(4):595-9

[Seeking optimal relation between oxygen saturation and hemoglobin concentration in adults with cyanosis from congenital heart disease.](#)

[Broberg CS, Jayaweera AR, Diller GP, Prasad SK, Thein SL, Bax BE, Burman J, Gatzoulis MA.](#)

Adult Congenital Heart Disease Program, Oregon Health and Sciences University, Portland, Oregon; Royal Brompton and Harefield National Health Service Trust, National Heart and Lung Institute, Imperial College London, London, United Kingdom.

Abstract

In patients with cyanosis from congenital heart disease, erythropoiesis is governed by many factors that can alter the expected relation between the oxygen saturation (O₂sat) and hemoglobin concentration. We sought to define the relation between the O₂sat and hemoglobin in such patients and to predict an ideal hemoglobin concentration for a given O₂sat. Adults with congenital heart defects and cyanosis were studied prospectively using blood tests and exercise testing. Nonoptimal hemoglobin was defined as any evidence of inadequate erythropoiesis (i.e., iron, folate, or vitamin B₁₂ deficiency, increased erythropoietin, reticulocytosis, or a right-shifted oxygen-hemoglobin curve). For patients without these factors, a linear regression equation of hemoglobin versus O₂sat was used to predict the optimal hemoglobin for all patients. Of the 65 patients studied, 21 met all the prestudy criteria for an optimal hemoglobin. For all patients, no correlation was found between O₂sat and hemoglobin ($r = -0.22$). However, a strong linear correlation was found for those meeting the criteria for optimal hemoglobin ($r = -0.865$, $p < 0.001$). The optimal hemoglobin regression equation was as follows: predicted hemoglobin = $57.5 - (0.444 \times O_2\text{sat})$. A negative correlation was found between the hemoglobin difference (predicted minus measured) and exercise duration on cardiopulmonary exercise testing ($r = -0.396$, $p = 0.005$) and the 6-minute walk distance ($r = -0.468$, $p < 0.001$). In conclusion, a strong relation between O₂sat and hemoglobin concentration can be shown in stable cyanotic patients and used to predict an optimal hemoglobin. This relation might be useful in defining functional anemia in this group.

Blood Press Monit. 2011 Feb;16(1):22-8

[Augmentation of central arterial pressure in adult patients after coarctation repair.](#)

[Szczepaniak-Chichel L, Trojnarowska O, Mizia-Stec K, Gabriel M, Grajek S, Gasior Z, Kramer L, Tykarski A.](#)

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Abstract

OBJECTIVES: Arterial hypertension and its complications are associated with an increased morbidity and mortality in patients after aortic coarctation (CoA) repair. It is debatable whether early surgery can prevent it. Residual stenosis of descending aorta (AoD) and stiffness in the pre-coarctation region supposedly lead to hypertension. The aim of this study was to evaluate aortic pulse-wave parameters in adult patients after CoA repair and to determine the influence of residual stenosis and age at operation on their values.

METHODS: Eighty-five patients underwent CoA repair (53 males; aged 34.6 ± 10.3 years, age at operation: 10.9 ± 8.2 years). The control group consisted of 30 individuals (18 males; aged 33.6 ± 8.2 years). Augmentation pressure (AP), augmentation index (AI), aortic pulse pressure (APP), and pulse-wave velocity (PWV) were measured with applanation tonometry method.

RESULTS: Normotensive patients after CoA repair (47/55%) had higher values of AP (7.3 ± 4.6 vs. 4.4 ± 3.6 mmHg; $P=0.002$), AI (18.6 ± 10.4 vs. $13.5 \pm 4.3\%$; $P=0.03$), APP (39.6 ± 8.8 vs. 35.1 ± 9.8 ; $P=0.00001$), and PWV (6.8 ± 1.2 vs. 5.4 ± 0.9 m/s; $P=0.003$) than controls. Forty-six patients presented signs of re-coarctation. No significant differences in the systolic blood pressure and diastolic blood pressure, AI and PWV were found between AoD(+) and AoD(-) but AoD(+) had significantly higher APP (46.7 ± 13.8 vs. 38.3 ± 6.9 ; $P=0.001$) and AP (10.5 ± 6.9 vs. 7.5 ± 4.1 ; $P=0.02$). Significant correlations were found between current age and APP, AP, and AI. No significant correlations were found between central parameters and the age at operation.

CONCLUSION: Normotensive patients after CoA repair have elevated central parameters, related to the residual AoD gradient, regardless of the age at operation. Even early repair cannot prevent progressive vascular impairment in CoA.

AJR Am J Roentgenol. 2011 Feb;196(2):454-60

[Cardiovascular anomalies in Turner syndrome: spectrum, prevalence, and cardiac MRI findings in a pediatric and young adult population.](#)

Kim HK, Gottliebson W, Hor K, Backeljauw P, Gutmark-Little I, Salisbury SR, Racadio JM, Helton-Skally K, Fleck R.

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Abstract

OBJECTIVE: Turner syndrome affects one in 2,500 girls and women and is associated with cardiovascular anomalies. Visualizing the descending thoracic aorta in adults with Turner syndrome with echocardiography is difficult. Therefore, cardiac MRI is the preferred imaging modality for surveillance. Our goals were to use cardiac MRI describe the spectrum and frequency of cardiovascular abnormalities and to evaluate aortic dilatation and associated abnormalities in pediatric patients with Turner syndrome.

MATERIALS AND METHODS: The cases of 51 patients with Turner syndrome (median age, 18.4 years; range, 6-36 years) were evaluated with cardiac MRI. The characteristics assessed included aortic structure, elongation of the transverse aortic arch, aortic diameter at multiple locations, and coarctation of the aorta (CoA). Additional evaluations were made for presence of bicuspid aortic valve (BAV), and partial anomalous pulmonary venous return (PAPVR). Associations between the cardiac MRI data and the following factors were assessed: age, karyotype, body surface area, blood pressure, and ventricular sizes and function.

RESULTS: Sixteen patients (31.4%) had elongation of the transverse aortic arch, eight (15.7%) had CoA, 20 (39.2%) had BAV, and eight (15.7%) had PAPVR. Aortic dilatation was most common at the aortic sinus (30%). Elongation of the transverse aortic arch was associated with CoA ($p < 0.01$) and BAV ($p < 0.05$). Patients with elongation of the transverse aortic arch had dilated aortic sinus ($p < 0.05$). Patients with PAPVR had increased right heart mass ($p < 0.05$), increased ratio of main pulmonary artery to aortic valve blood flow ($p = 0.0014$), and increased right ventricular volume ($p < 0.05$).

CONCLUSION: Cardiovascular anomalies in pediatric patients with Turner syndrome include aortic abnormalities and PAPVR. The significant association between elongation of the transverse aortic arch and CoA, BAV, and aortic sinus dilatation may contribute to increased risk of aortic dissection. The presence of PAPVR can be hemodynamically significant. These findings indicate that periodic cardiac MRI screening of persons with Turner syndrome is beneficial.

Am J Cardiol. 2011 Feb 1;107(3):466-71.

[Complex congenital heart disease in unaffected relatives of adults with 22q11.2 deletion syndrome.](#)

Swaby JA, Silversides CK, Bekeschus SC, Piran S, Oechslin EN, Chow EW, Bassett AS.

Toronto Congenital Cardiac Centre for Adults, University of Toronto, Peter Munk Cardiac Centre, University Health Network/Toronto General Hospital, Ontario, Canada.

Abstract

The 22.q11.2 deletion syndrome (22q11DS) is a common genetic condition associated with 22q11.2 microdeletions and classically has included congenital heart disease (CHD) as a part of the variable expression. Some evidence has shown that relatives of those with 22q11DS might be at an increased risk of CHD in the absence of 22q11.2 deletions. We obtained a detailed family history of CHD in the first- to third-degree relatives ($n = 2,639$) of 104 adult probands with 22q11DS. We compared the prevalence of CHD in the relatives without 22q11.2 deletions to the published general population prevalence. We also investigated the effect of CHD in the probands on prevalence of CHD in the relatives. Of the 104 probands with 22q11DS, 14 (13.5%) had 17 relatives (17 of 2,639, 0.6%) with CHD. Of 66 probands with CHD, 15 (0.9%) of their 1,663 relatives had CHD, a significantly greater prevalence than that for the relatives of probands without CHD (0.2%, 2 of 976, $p = 0.041$, odds ratio 4.43, 95% confidence interval 1.03 to 40.00). In relatives of probands with CHD, the prevalence of those with severe CHD (0.36%) was significantly elevated compared to population expectations (0.061%, $p = 0.007$, odds ratio 5.88, 95% confidence interval 2.16 to 12.85). In conclusion, these results support a heritable susceptibility to CHD in families of probands with 22q11DS, in addition to that imparted by microdeletion 22q11.2. The occurrence of CHD in relatives might be related to the expression of CHD in the proband with 22q11DS. These findings have potential implications for the genetic counseling of families of those with 22q11DS and support the notion that interacting genetic variants might contribute to the variable expression of 22q11DS.

Br J Radiol. 2011 Feb; 84(998):188-93. Epub 2010 Dec 15.

[Right heart dilation in adult congenital heart disease: imaging appearance on cardiac magnetic resonance.](#)

Beitzke D, Wolf F, Edelhauser G, Lammer J, Loewe C.

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Abstract

Imaging morphology and function of the right heart is of paramount importance in patients with adult congenital heart disease, since right ventricular dysfunction is associated with adverse cardiac events. Cardiac MRI has been shown to be a powerful tool for the non-invasive precise assessment of right ventricular and valvular dysfunction. Differential diagnoses of congenital heart disease characterised by, or combined with, right heart dilatation are diverse and necessitate a systematic approach.

J Am Acad Dermatol. 2011 Feb;64(2):290-5. Epub 2010 Nov 26.

[A case-control study of cutaneous signs in adult patients with Marfan disease: diagnostic value of striae.](#)

Ledoux M, Beauchet A, Fermanian C, Boileau C, Jondeau G, Saiag P.

Department of Dermatology, Université Versailles St-Quentin, Ambroise Paré Hospital, Boulogne, France.

Abstract

BACKGROUND: Marfan syndrome (MS) (OMIM 154700) has been associated with various skin manifestations.

OBJECTIVE: We sought to clarify the value of skin signs in patients with MS.

METHODS: This was a case-control study. A total of 61 consecutive patients (median age: 34 years) seen in the French Reference Centre for MS and Related Disorders and with a confirmed diagnosis of MS were paired with 61 age-, sex-, and height-paired control subjects. All had a structured interview and standardized dermatologic examination. The gold standard for MS diagnosis was the Ghent criteria.

RESULTS: Striae of any type were significantly ($P = .0001$) more frequent in patients with MS (92%) than in control subjects (61%), but specificity was low (39%, 95% confidence interval [CI] 27-52). Striae on unusual locations (other than buttock, hip, or thigh) were more frequent in patients with MS (66%) than in control subjects (16%) ($P < .0001$). This finding had a high specificity (84%, 95% CI 74-93), without notably decreasing sensitivity (66%, 95% CI 54-77). Hypertrophic, large, or atrophic surgical or posttraumatic, frequently hypopigmented or hyperpigmented, scars were present in 46% of patients with MS and 21% of control subjects ($P = .007$). Sensitivity was 46% (95% CI 34-58) and specificity 79% (95% CI 67-87). Atypical striae in some control subjects could be attributed to intensive practice of sports.

LIMITATION: A few control subjects were selected from patients consulting the MS center but without a diagnosis of MS.

CONCLUSION: Striae are a good diagnostic criterion for MS, particularly when arising in unusual sites. Other reported skin signs of MS are infrequent.

BJOG. 2011 Feb;118(3):345-52. doi: 10.1111/j.1471-0528.2010.02743.x. Epub 2010 Nov 22.

[Chronic heart disease and severe obstetric morbidity among hospitalisations for pregnancy in the USA: 1995-2006.](#)

Kuklina E, Callaghan W.

Division of Heart Disease and Stroke Prevention, National Center for Chronic Disease Prevention and Health Promotion, Centers for Disease Control and Prevention, Atlanta, GA 30341-3724, USA. ekuklina@cdc.gov

Abstract

OBJECTIVES: to describe changes in characteristics of delivery and postpartum hospitalisations with chronic heart disease from 1995 to 2006.

DESIGN: cross-sectional study.

SETTING: USA, nationwide hospital discharge data.

POPULATION: a total of 47 882 817 delivery hospitalisations and 660 038 postpartum hospitalisations.

METHODS: adjusted odds ratios describing the associations between chronic maternal heart disease and severe obstetric complications were obtained from multivariable logistic models. The contribution of chronic heart disease to severe morbidity was estimated using adjusted population-attributable fractions.

MAIN OUTCOME MEASURES: prevalence and trends in chronic heart disease, rate and risk of severe obstetric complications.

RESULTS: in 2004-2006, about 1.4% of delivery hospitalisations were complicated with chronic heart disease. No substantial changes in the overall prevalence of chronic heart disease among hospitalisations for delivery were observed from 1995-1997 to 2004-2006. Even so, a linear increase was found for specific congenital heart disease, cardiac dysrhythmias, and cardiomyopathy and congestive heart failure ($P < 0.01$). During this same period the rate of postpartum hospitalisations with chronic heart disease tripled ($P < 0.01$). Severe complications during hospitalisations for delivery among women with chronic heart disease were more common in 2004-2006 than in 1995-1997. In 2004-2006, 64.5% of the cases of acute myocardial infarction, 57.5% of the instances of cardiac arrest/ventricular fibrillation, 27.8% of in-hospital mortality and 26.0% of the cases of adult respiratory distress syndrome were associated with hospitalisations with chronic heart disease.

CONCLUSIONS: in the USA chronic heart disease among women hospitalised during pregnancy may have increased in severity from 1995 to 2006.

Cardiol Young. 2011 Feb;21(1):31-8. Epub 2010 Oct 27.

[Ablation of atrial tachyarrhythmias late after surgical repair of tetralogy of Fallot.](#)

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Abstract

BACKGROUND: Patients with repaired tetralogy of Fallot may develop symptomatic and haemodynamic deterioration for many reasons such as arrhythmia, pulmonary regurgitation, and impairment in ventricular function. We describe a consecutive group of patients whose main clinical problem was atrial tachyarrhythmias.

AIMS: To describe the clinical outcome of atrial tachyarrhythmias occurring late after surgical repair of tetralogy of Fallot; to define the circuits/foci responsible for these atrial tachyarrhythmias; to evaluate the outcome of computer-assisted

mapping and catheter ablation in this patient group.

METHODS AND RESULTS: Consecutive patients with surgically repaired tetralogy of Fallot and atrial tachyarrhythmias, who underwent catheter ablation between January, 2001 and June, 2007, were identified retrospectively from case records. Computer-assisted mapping was performed in all using either EnSite® (St Jude Medical Inc.) arrhythmia mapping and intra-cardiac catheter guidance system or CARTO™ (Biosense Webster Inc.) electroanatomical mapping systems. Ten patients (four males) with a median age of 39 plus or minus 8 years were studied. The total number of atrial tachyarrhythmias identified was 22 (six macro-reentrant, 16 micro-reentrant/focal). In nine patients, catheter ablation led to improvement in arrhythmia episodes and/or symptoms during follow-up of 41 plus or minus 20 months. Following ablation(s), five patients required pacing for pre-existing conduction disease and five needed further surgery for haemodynamic indications. All patients remained on anti-arrhythmic drugs.

CONCLUSIONS: Patients with surgically repaired tetralogy of Fallot and atrial tachyarrhythmias typically have multiple arrhythmic circuits/foci arising from a scarred right atrium. Catheter ablation reduces arrhythmia frequency and improves symptoms. However, hybrid management is often required, comprising drugs, pacing, and further surgery tailored to the individual.

Cardiol Young. 2011 Feb;21(1):39-45. Epub 2010 Oct 6.

[Ten-year experience with surgical treatment of adults with congenital cardiac disease.](#)

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Abstract

The number of adults with congenital cardiac disease continues to increase, and adult patients are now more numerous than paediatric patients. We sought to identify risk factors for perioperative death and report our results with surgical management of adult patients with congenital cardiac disease. We retrospectively analysed in-hospital data for 244 consecutive adult patients who underwent surgical treatment of congenital cardiac disease in our centre between January, 1998 and December, 2007. The mean patient age was 27.2 plus or minus 11.9 years, 29% were in functional class III or IV, and 25% were cyanosed. Of the patients, half were operated on for the first time. A total of 61% of patients underwent curative operations, 36% a reoperation after curative treatment, and 3% a palliative operation. Overall mortality was 4.9%. Predictive factors for hospital death were functional class, cyanosis, non-sinus rhythm, a history of only palliative previous operation(s), and an indication for palliative treatment. Functional class, cyanosis, type of initial congenital cardiac disease (single ventricle and double-outlet right ventricle), and only palliative previous operation were risk factors for prolonged intensive care stay (more than 48 hours). The surgical management of adult patients with congenital cardiac disease has improved during recent decades. These generally young patients, with a complex pathology, today present a low post-operative morbidity and mortality. Patients having undergone palliative surgery and reaching adulthood without curative treatment present with an increased risk of morbidity and mortality. Univentricular hearts and double-outlet right ventricles were associated with the highest morbidity.

Int J Cardiol. 2011 Feb 3;146(3):326-9. Epub 2009 Aug 8.

[Quality outcomes of ACHD patients undergoing cardiovascular procedures and hospital admissions in a free-standing children's hospital.](#)

Crumb SR, Cook SC, Cheatham JP, Galantowicz M, Feltes TF, Phillips A, Chan DP, Holzer RJ, Sisk M, Olshove V, Cook LR, Hickey JC, Hill SL, Daniels CJ.

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Abstract

OBJECTIVES: Describe quality outcomes of adults with congenital heart disease (ACHD) undergoing cardiovascular procedures and admissions in a free-standing children's hospital with a multi-disciplinary ACHD program and compared with pediatric outcomes.

BACKGROUND: A challenge for the U.S. healthcare system is where to treat the >1 million ACHD patients (pts): adult hospitals without CHD care, or pediatric hospitals without adult services.

METHODS: We reviewed all CHD pts \geq 18 yrs of age from 2002-2007. Procedural and hospital related morbidity and mortality were recorded. ANOVA and t-test compared adult with pediatric pts.

RESULTS: Overall, 782 pts, mean age of 29.8 ± 9.9 yrs, encountered 1490 procedures/admissions. For 178 cardiac surgeries (72% reoperations), mortality was 1.8% and complication rate was 7.3%. There was 0% mortality for 412 cardiac catheterizations, 311 electrophysiological procedures, 401 transesophageal echocardiograms (TEE), 244 exercise tests (ETT) and 54 medical admissions. Major adverse event rate was 0.6% for cardiac catheterization and electrophysiological procedures. No adverse events occurred during TEE and ETT. Only 4 pts required transfer to an adult institution (0.25%). There was no significant difference in mortality or adverse events between pediatric and adult CHD pts, $p > 0.05$.

CONCLUSIONS: The optimal setting to provide ACHD care remains a complex issue. Our study is the first to demonstrate 1) a low incidence of morbidity and mortality for ACHD pts undergoing cardiovascular procedures or admissions at a free-standing children's hospital, 2) outcomes comparable to pediatric CHD pts. Future models incorporating ACHD programs within pediatric heart centers should be considered to care for this complex population.

Psychology Section Update

The Psychology Section of ISACHD was formed in late 2010 with the support of the ISACHD Executive. The section includes a total of 24 ACHD health care providers with a focus on the psychological/psychiatric aspects of ACHD. Specifically, our group is comprised of 9 psychologists, 7 cardiologists, 2 nurses, 2 psychology trainees (one PhD student and one post-doctoral fellow), 2 ACHD cardiology fellows, 1 psychiatrist, and 1 social worker. We have members from Asia, Australia/New Zealand, Europe, and North America.

The four primary aims of the Psychology Section are:

Connection. ISACHD offers an excellent opportunity for a 'virtual home' in which ACHD psychologists, psychiatrists, cardiologists, nurses and other mental health professionals can connect and keep one another informed about new research developments. This will provide an opportunity for asking questions, discussing ideas, and perhaps even collaboration. Our important next step will be to work with the ISACHD website team to develop our virtual home within a secure section of the website.

Knowledge. Members of our group will occasionally provide an update in

the ISACHD newsletter in which we summarize research focused on the psychosocial experiences of ACHD patients.

Expansion. ACHD programs that are beginning to collaborate with or hire psychologists, psychiatrists, or other mental health professionals will be able to contact and seek guidance from individuals with expertise in this field.

Consultation. The website will provide a venue in which members can ask questions of a more clinical nature (e.g., assessment instruments, useful clinical tools or resources).

The majority of our members have a defined research interest in the psychosocial aspects of ACHD. Our psychologist and psychiatrist members were polled, and the majority provides psychological services to both inpatients and outpatients. Some provide services only to patients up to the age of 18, some provide services only to patients following transfer to adult care, and some treat "from fetus to adult" as they provide support to pregnant women, children, and adults.

If you know of any colleagues who would be interested in joining the Psychology Section, please encourage them to contact Adrienne Kovacs (Psychologist, Toronto Congenital Cardiac for Adults) for further information.

New from CACH Network

Erwin Oechslin

Heart Disease and Pregnancy Website

This website was recently launched and is a great educational resource for physicians and patients!

The Heart Disease and Pregnancy website, provides information for physicians and patients about the risks associated with pregnancy and strategies for management of pregnant women with heart disease. The website was developed by an international panel of physicians including internists, cardiologists, obstetricians as well as other medical specialists with expertise in pregnancy and heart disease. A drop-down menu for different acquired and congenital heart diseases makes for easy access to information about maternal and fetal complications, management strategies, references, and other subtopics for a specific disease. Key links to national and international societies, teaching tools are also listed and easily accessible.

I congratulate the contributors for their outstanding, globally available educational tool. Special thanks to the editors Dr. Candice Silversides, Dr. Jack Colman, Angela Kennie and web designer Ray Ganesh.

Visit the website at:

www.heartdiseaseandpregnancy.com

Only 23% of the Canadian ACHD patients are under regular ACHD care

This is the conclusion of a collaborative project of all Canadian Adult Congenital (CACH) network centers under the leadership of Dr. Luc Beauchesne, Ottawa (Beauchesne LM et al, Int J Cardiol Dec 27 2010 [Epub ahead of print]). All 15 CACH network clinics prospectively collected outpatient and procedural volumes for the year 2007. Of a minimum number of 96,000 ACHD patients in Canada, only 21,879 (23%) were being regularly followed in 2007. It is disappointing that only a minority of ACHD patients are affiliated with an ACHD centre, despite early publications of guidelines for the management of ACHD patients Canada in 1998, establishment of a

structured pan-Canadian care plan in the early 1990s, and educational efforts. The reasons are multifactorial: suboptimal or lack of transition and transfer of adolescents to ACHD centers until the late 1990s or early 2000s, the perception of 'cure' amongst others.

How is the loss of appropriate follow-up in other countries without a structured, national care plan acceptable for ACHD patients?

That wait times for ACHD services were beyond Canadian recommended targets at most sites was another striking, but not unexpected finding. Health care leaders and politicians are challenged to meet the expectations and recommended targets as set out for patients with acquired cardiovascular diseases. Improving public awareness and networking with politicians and decision makers are strategies, amongst others, to articulate the increasing need for human and structural resources: a tsunami of ACHD patient is and will be challenging the adult health care system!

Congenital Heart Disease Meetings in Toronto

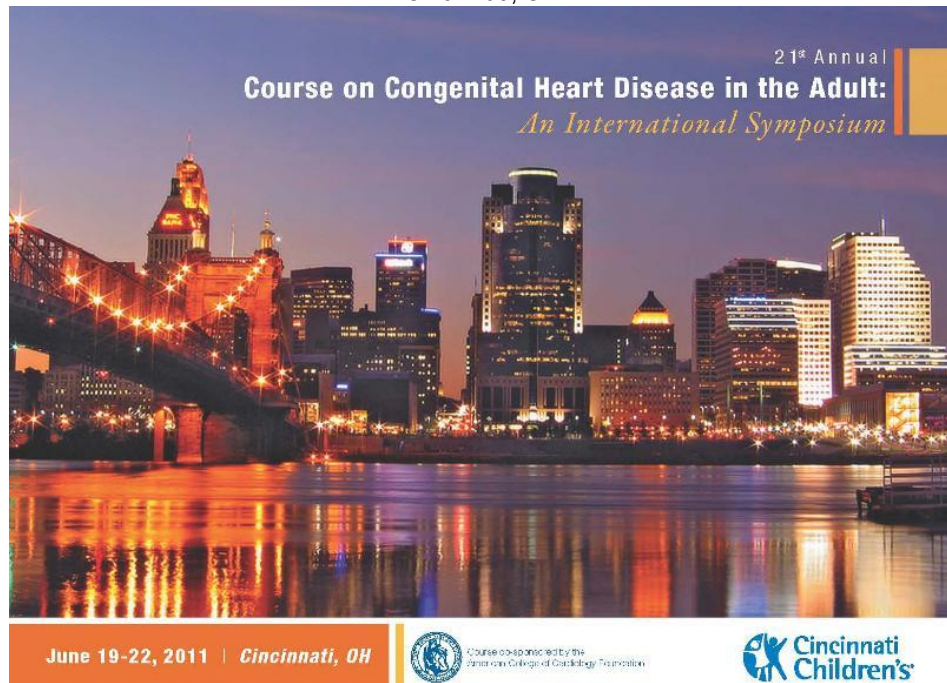
The 5th *Cardiac Symposium: Hypoplastic Left Heart Syndrome* will be held in the Four Seasons Hotel from *June 5-7, 2011*.

The *Pre-Symposium Course: 3 Dimensional Anatomy of the Heart* will be held from *June 1-3, 2011*.

Early Bird Registration at <http://www.sickkids.ca/Centres/heart-centre/Cardiac-symposium/> has been extended to April 4, 2011.

**21st Annual Course on Congenital Heart Disease In the Adult:
An International Symposium**

June 19-22, 2011
Cincinnati, OH



For more information:

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