November 2015

International Society for Adult Congenital Heart Disease

ISACHD News

In this Issue

President's Message

By Paul Khairy, MD, PhD, President, ISACHD

Welcome to our 4th Quarter 2015 ISACHD newsletter!

Herein, you will find updates from our global working groups on education and research, our fellows-in-training, and our cardiac care associates, along with regional news.

Our Journal Watch features articles pertaining to the care of adults with congenital heart disease across several themes, including interventional cardiology, genetics, imaging, heart failure, and from our cardiac care associates.

I would like to extend my sincere congratulations to the first ever recipients of our ISACHD Research Awards, which were presented at the 25th International ACHD Symposium in Toronto, Canada:

- The ISACHD Young Investigator Award was presented to **Dr. Romy Franken** for her abstract entitled, "Genotype impacts survival in Marfan Syndrome".
- The ISACHD Cardiac Care Associates Research Award was presented to **Dorothy D. Pearson** for her abstract entitled, "Adult congenital heart disease surgery in the developing world".

I would also like to highlight the recent publication of three ISACHD manuscripts, which consist of a research methods paper, ISACHD's first position statement, and the results of a research survey by our Fellows-in-Training:

- Apers S, Kovacs AH, Luyckx K, Alday L, Berghammer M, Budts W, Callus E, Caruana M, Chidambarathanu S, Cook SC, Dellborg M, Enomoto J, Eriksen K, Fernandes SM, Jackson JL, Johansson B, Khairy P, Kutty S, Menahem S, Rempel G, Sluman MA, Soufi A, Thomet C, Veldtman G, Wang JK, White K, Moons P; APPROACH-IS consortium; International Society for Adult Congenital Heart Disease (ISACHD). Assessment of Patterns of Patient-Reported Outcomes in Adults with Congenital Heart disease - International Study (APPROACH-IS): rationale, design, and methods. *Int J Cardiol 2015;179*:334-42.
- Webb G, Mulder BJ, Aboulhosn J, Daniels CJ, Elizari MA, Hong G, Horlick E, Landzberg MJ, Marelli AJ, O'Donnell CP, Oechslin EN, Pearson DD, Pieper EP, Saxena A, Schwerzmann M, Stout KK, Warnes CA, Khairy P. The care of adults with congenital heart disease across the globe: Current assessment and future perspective: A position statement from the International Society for Adult Congenital Heart Disease (ISACHD). *Int J Cardiol. 2015;195*:326-33.
- Bokma JP, Burchill LJ, Kovacs AH, Oechslin EN, Khairy P, Mulder BJ, Veldtman GR. Fellows' perspectives on training in adult congenital heart disease: Results of a survey from the International Society for Adult Congenital Heart Disease (ISACHD). *Int J Cardiol. 2015;202*:253-5.

I invite you all to join us for our ISACHD Semi-Annual Meeting in Orlando *(see agenda on page 2)*. I look forward to a fun evening of comradery, food, and drinks!

ISACHD Semi-Annual Meeting

Sunday, November 8, 2015
American Heart Association (AHA) Scientific Sessions

Hilton Orlando — Lake Highland Room
6001 Destination Parkway, Orlando, Florida

6:30 pm	Cocktails, appetizers, and networking
7:00 pm	Welcome and update from the ISACHD Executive *Paul Khairy*
7:20 pm	Update from the Cardiac Care Associates *Susan Fernandes*
7:30 pm	Global Research Working Group update *Jamil Aboulhosn*
7:40 pm	"Lessons learned from the multinational APPROACH-IS study" *Philip Moons, Principal Investigator, APPROACH-IS*
8:00 pm	Global Health Working Group update *Disty Pearson*
8:10 pm	"Sustainable congenital heart disease care in the developing world" *Bistra Zheleva, Vice President, International Programs, Children's HeartLink*
8:30 pm	Regional updates (Europe, South America, Asian-Pacific, USA, Canada)
8:45 pm	ISACHD Lifetime Achievement Award
9:00-9:30 pm	More cocktails, appetizers, and networking!

Upcoming Scientific Conferences

The Fourth International Congress on Cardiac Problems in Pregnancy (CPP) 2016

February 27-March 1, 2016
Las Vegas, USA

The 2017 World Congress of Pediatric Cardiology and Cardiac Surgery

June 18-23, 2017
Istanbul

Executive Committee

Regional and Working Group News

News from Asia Pacific

Clare O'Donnell, New Zealand

Adult congenital heart disease sessions continue to held during regional cardiac meetings including the Cardiac Society of Australia and New Zealand meeting held August in Melbourne and the upcoming Pediatric Cardiac Society of India meeting in Hyderabad.

We are keen to advise that planning is well underway for the 17th annual meeting of Japanese Society for ACHD to be held in January 13th-15th 2016 in Osaka.

News from Europe

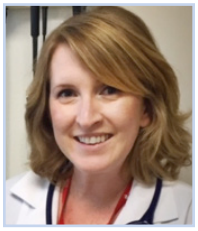
Els Pieper, Groningen, NL

In London the European congenital heart disease community had ample possibilities to keep their knowledge on adult congenital heart disease up to date and meet colleagues. In addition to many other sessions dedicated to congenital heart disease, there also was a session organized in collaboration with ISACHD, chaired by Barbara Mulder and Paul Khairy.

The ESC 10th European Echocardiography Course on Congenital Heart Disease will be in Leuven, Belgium, 14-17 October.

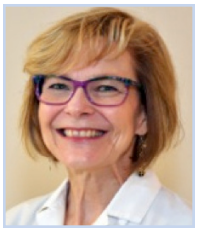
Cardiac Care Associates Update

Sue Fernandes, Stanford



Christy Sillman, MSN, RN

The ISACHD CCA-Nursing Education Workgroup is working towards publication of their first manuscript examining the nurse coordinator role in ACHD clinics around the globe. Leading the project are Christy Sillman, MSN, RN (U.S.) and Joanne Morin, MscN, RN (Canada). Other nurses currently collaborating on this project include: Corina Thomet, MScN, RN (Switzerland), Deena Barber, BSN, RN (U.S.), Chi-Wem Chen, PhD, RN (Taiwan), Sheena Vernon, MSc, CNS (UK), Yoshiko Mizuno, RN (Japan) and Serena Flocco, RN (Italy). The manuscript will be a tremendous resource for both ACHD clinics that are just starting out as well as those established programs looking to better understand the potential of the nurse coordinator role on the ACHD Team. Topics to be included are: organizing, coordinating and facilitating ACHD care, holistic assessment of the ACHD patient, ACHD patient education as it relates to symptoms triage, procedure preparation, heart failure management, anticoagulation and other medication management, and reproductive health and lifestyle choices. The manuscript will also cover end-of-life considerations and community based resources. Publication is expected sometime in spring of 2016.



Joanne Morin, MscN, RN

News from the Fellows

Jouke Bokma, Amsterdam

Fellows' perspectives on ACHD training: results of survey published by *International Journal of Cardiology*

The manuscript based on the results of the ISACHD survey on ACHD training which was distributed across fellows worldwide has been accepted for publication in the International Journal of Cardiology. The online only version is already available at: http://www.ncbi.nlm.nih.gov/pubmed/26407046 and the print version is expected to appear soon. A major finding is that only one-third of responding ACHD training fellows rated their training programs as "very good." In particular, fellows expressed concerns over ACHD training related to CT and MRI. Therefore we encourage current ACHD program leaders to critically appraise training opportunities, particularly in advanced imaging modalities, within their ACHD fellowship programs.

We would like to thank all ACHD program leaders who have distributed the survey among ACHD fellows-in-training. Off course, we would also like to thank all fellows who have completed the survey. Their responses gave an important insight in worldwide ACHD training and indicated important areas for improvement.

Working Group on Education

Els Pieper, Groningen, NL

The activities of the working group on Education are concentrated mainly around the ACHD learning center. The learning center (www.achdlearningcenter.org) is under the leadership of Gary Webb evolving into a much-used facility. To mention one example, the Journal Watch is highly appreciated and attracts monthly around 3000 users.

The web-based teaching course has now around 45 presentations posted on the website and 10 more are waiting to be finished. The formal education program related to the course is working in the beta testing mode and program directors can choose their own selection of presentations. 11 programs in different countries are now participating. Both British and Dutch adult cardiology trainees will be mandated to use the testing center during their training. The MCQ's for this formal education program are developed by Jonathan Windram and Kate English.

The set of slide presentations on catheterization, developed by Hamsimran Singh and Richard Krasuski, is growing: nine are now finished and will be available on the learning center website, nine more will follow.

The working group on education endorsed the following congress: International Congress on Cardiac Problems in Pregnancy, CPP 2016, Las Vegas, Febr 27 - March 1, 2016. ISACHD members can claim a discount of 10% on the registration fee.

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Regional and Working Group News *continued from page 3*

Working Group on Research

Jamil Aboulhosn, Los Angeles

The ISACHD research working group has been tasked with the mission of identifying and helping implement multinational ACHD research projects. Two multi-national projects have thus far been endorsed by ISACHD:

1. **APPROACH IS — Assessment of Patterns of Patient-Reported Outcomes in Adults with Congenital Heart Disease – International Study**. This is an international multicenter project on patient reported outcomes in adults with CHD. Researchers and clinicians from 16 countries, across 4 continents, have collaborated on one of the largest studies in the field of ACHD. The study is coordinated through the University of Leuven, the principal investigator is Professor Philip Moons, PhD, RN. The steering committee includes Professors Moons, Adrienne Kovaks and Koen Luyckx. Data collection for this project commenced in April 2013 and was completed in January of 2014. The total sample size is >3,500 patients. Data was analyzed during the first 6 months of 2015 and primary results of the study will hopefully be available in 2016. Sub-studies will likely be performed after the primary analyses have been conducted and reported. For further information regarding this study please contact Professor Philip Moons (Philip.Moons@med.kuleuven.be).
2. **NOTE Registry —** The purpose of the NOTE registry is to evaluate the efficacy and safety of non-vitamin K oral antagonists (NOACs) for thromboembolic prevention in atrial tachyarrhythmias in adult patients with CHD. The principal investigator is Professor Barbara Mulder, MD, PhD at the University of Amsterdam. The primary objective of the registry is to evaluate the two year incidence of thromboembolic and bleeding events in consecutive CHD patients using NOACs for atrial tachyarrhythmias. The secondary objectives of the registry are to evaluate adherence to NOAC therapy and the quality of life when using NOACs, to identify CHD-specific risk factors for thromboembolism in atrial tachyarrhythmias and to evaluate the natural history of atrial tachyarrhythmias in CHD. Preliminary registry data was presented at the European Society of Cardiology (ESC) congress in London in 2015 during the rapid fire presentation session 'Hot topics in congenital heart disease'. Currently, 145 adults with CHD using NOACs have been enrolled from 37 participating centers worldwide. The inclusion will continue until at least 200 patients are enrolled and new centers are welcome at this point. The registry is currently enrolling subjects. For further information please contact Hayang Yang (h.yang@amc.uva.nl).

In addition, Dr. Craig Broberg at Oregon Health Sciences University (OHSU) has sought multi-center collaboration for an NIH grant focusing on detection of diffuse fibrosis using MRI to measure extracellular volume fraction (ECVF). The hypothesis is that fibrosis can be a discriminator for patients with repaired tetralogy of Fallot (TOF) and D-transposition of the great arteries (D-TGA) at high risk for ventricular arrhythmias and heart failure. The proposal is for a prospective, multicenter study for determination of ECVF in repaired TOF or D-TGA patients with a systemic right ventricle. The NIH grant was submitted in October of 2015 and review should be completed in March 2016 with award notification later in the spring. For further information please contact Dr. Broberg (craigbroberg@gmail.com).

The ISACHD research working group is looking for additional multi-national studies to review and potentially endorse. Please contact the research working group chair person, Jamil Aboulhosn, MD (jaboulhosn@mednet.ucla.edu) if you would like to propose a multi-national research protocol or get further information.

Journal Watch

Intervention Section

Comments by Konstantin Averin

Catheter Cardiovasc Interv. 2015 Sep 1;86(3):438-52. doi: 10.1002/ccd.25897. Epub 2015 Mar 16.

Iatrogenic aortopulmonary communications after transcatheter interventions on the right ventricular outflow tract or pulmonary artery: Pathophysiologic, diagnostic, and management considerations.

Torres A[1], Sanders SP[2], Vincent JA[1], El-Said HG[3], Leahy RA[4], Padera RF[5], McElhinney DB[6].

Comment: In this study, Torres and colleagues seek to investigate the spectrum, etiology, and management of traumatic aortopulmonary communications after transcatheter interventions on the pulmonary circulation. There present 3 new cases and review the literature to present a total of 18 patients with iatrogenic AP communication after transcatheter interventions on the PAs or RVOT, primarily in patients with transposition of the great arteries who underwent PA angioplasty after an arterial switch operation or after transcatheter pulmonary valve replacement in patients who had undergone a Ross procedure. The etiology of AP communications in this group of patients is likely PA trauma and/or distortion of the neo-aortic anastomosis resulting from the intervention, with subsequent dissection through the extravascular connective tissue and devitalized tissue at the site of the surgical anastomosis. Of the 18 cases, only 4 were diagnosed during the same catheterization and 3 more within the next 2 days. The authors emphasize that providers should maintain a high index of suspicion for this rare complications as signs and symptoms of an AP communication may be similar to those of other serious events and the initial response, PA angiography, may not diagnose the problem. Once diagnosed, a number of therapeutic strategies have been utilized, including covered CP stenting of the RVOT or branch PAs, aortic endograft placement, and a variety of closure devices (i.e. Amplatzer septal occluder).

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JACC Cardiovasc Interv. 2015 Apr 20;8(4):600-6. doi: 10.1016/j.jcin.2015.02.002.

Long-term outcome after transcatheter closure of atrial septal defect in older patients: impact of age at procedure.

Takaya Y[1], Akagi T[2], Kijima Y[1], Nakagawa K[1], Sano S[3], Ito H[1].

Comment: In this study, Takaya and colleagues investigate the impact of age on ASD closure in patients who are older than 50 years old (grouped into 3 groups by age at time of procedure: 50-59, 60-74 and >75 years old). The primary end point was all cause mortality and hospitalization due to heart failure or stroke. The study population consisted of 244 consecutive patients with 55 of the patients being older than 75. During a median follow up of 36 months, mortality and hospitalization due to heart failure or stroke occurred in 18 patients (7%). Kaplan-Meier analysis showed that the event-free survival rate was not different among the 3 age groups and that New York Heart Association functional class and right ventricular/left ventricular end-diastolic diameter ratio improved in patients older than 75 years of age, similar to the other age groups. The authors conclude that the long-term outcome in patients older than 75 years of age is not different from that in the other age groups and that functional capacity and cardiac remodeling improved in patients older than 75 years of age, similar to the other age groups. ASD closure can be considered a valuable therapeutic option in patients older than 75 years of age.

Genetics Section

Comments by Benjamin Landis

Am J Med Genet A. 2015 Jun 27. doi: 10.1002/ajmg.a.37208. [Epub ahead of print]

Aortic dimensions in Turner syndrome.

Quezada E[1], Lapidus J[2], Shaughnessy R[1], Chen Z[2], Silberbach M[1].

Comment: Patients with Turner syndrome are at risk for development of thoracic aortic aneurysm and have elevated lifetime risk for aortic dissection or rupture. Normative data from healthy populations have been used to establish thresholds for defining aortic dilation and qualifying its severity, often by indexing to body size parameters (e.g. body surface area). Because Turner syndrome usually presents with short stature, the typical thresholds are not applicable for these patients. This uncertainty leads to significant challenges for risk stratification and clinical decision making. Quezada and colleagues have addressed this significant knowledge gap by establishing standard echocardiographic measurements for aortic size based on a healthy cohort with Turner syndrome. In total, 481 patients of median age 25 (range 3 to 70) years were included. Most had XO karyotype, but as many as 40% reported a different type of X chromosome structural abnormality (e.g. translocation) or mosaicism. Patients with aortic valve stenosis or regurgitation, unoperated congenital heart disease, or history of aortic dissection or replacement, were excluded. However, patients with history of coarctation repair (12%) or hypertension (20%) were included. The aortic diameter was measured using 2D transthoracic echocardiography at 8 levels (aortic annulus to descending aorta). In multivariate linear regression models, bicuspid aortic valve (25% of cohort) was significantly associated with aortic diameter at the levels of annulus, root, sinotubular junction, and ascending aorta, and therefore excluded. Ultimately, the regression parameters (based on body surface area calculated with the Haycock formula) for each of the 8 aortic segments are defined in the manuscript. As recommended for routine clinical care of these patients, longitudinal follow up will help to define the long term risk of aortic dissection or rupture and enhance clinical interpretation of Z-score values.

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Am J Med Genet A. 2015 Aug;167(8):1822-9. doi: 10.1002/ajmg.a.37108. Epub 2015 Apr 30.

The diagnostic value of next generation sequencing in familial nonsyndromic congenital heart defects.

Jia Y[1], Louw JJ[1,2], Breckpot J[1,3], Callewaert B[4], Barrea C[5], Sznajer Y[6], Gewillig M[2], Souche E[1], Dehaspe L[1], Vermeesch JR[1], Lambrechts D[7,8], Devriendt K[1],Corveleyn A[1].

Comment: Jia and colleagues sought to identify disease-causing genetic variants from families with nonsyndromic cardiovascular malformations (CVMs). In total 13 families, each containing at least 3 relatives (1st or 2nd degree) with a CVM, were recruited from clinics in Belgium. Subjects were tested for coding or splice variants from a customized panel of 57 genes that are already known to be associated with syndromic or nonsyndromic CVMs (e.g. *JAG1, NKX2-5*). The study included relatives with and without CVMs as confirmed by echocardiography. Among the 36 subjects tested with the 57-gene panel, there were 44 heterozygous rare variants (i.e. variants present in less than 1% of individuals in large reference population databases). Based on bioinformatics-based predictions of the effect of these variants and segregation with disease within families, the authors concluded that a heterozygous variant likely to be causing CVMs was identified in 6 of the 13 families (46%) in autosomal dominant inheritance patterns. Three families carried variants in *NOTCH1* (one missense, one frameshift, one splice donor site), 2 families carried variants in *TBX5* (both missense), and 1 family carried a variant in *MYH6* (missense). One of the *TBX5* variants was previously reported, but the other 5 variants were novel.

There are several aspects of the study's findings that highlight the complex nature of the genetics of nonsyndromic CVMs. First, in at least 3 of the 6 families there were individuals who carried the suspected causative variant but who had normal cardiac anatomy (i.e. reduced penetrance). Second, in 2 of the 6 families there were individuals with a CVM who did not carry the variant suspected to be causative within the family; the authors speculate that the relatively high prevalence of CVMs overall can lead to instances where even related individuals have different genetic mechanisms. These observations emphasize the importance of testing as many relatives as possible both in research and clinical settings. Third, the specific type of CVM often varied significantly within families (i.e. intra-familial variable expression). Finally, the subjects with *TBX5* variants did not demonstrate the upper limb skeletal malformations classically associated with Holt-Oram syndrome. This raises the possibility that mutation testing for this gene may be informative in nonsyndromic TAA rather than only in cases of CVMs and upper limb malformations. Continued advancement of our understanding of the genetic mechanisms of familial CVMs will require functional interpretation of variants as well as integration of various classes of human genetic variation. These data are critical as broad gene panels and whole exome/genome sequencing testing become increasingly available clinically.

Imaging Section

Comments by Michiel Winter

J Cardiovasc Comput Tomogr. 2015 Jul-Aug;9(4):354-61. doi: 10.1016/j.jcct.2015.04.007. Epub 2015 May 1.

Comprehensive assessment of morphology and severity of atrial septal defects in adults by CT.

Osawa K[1], Miyoshi T[2], Morimitsu Y[3], Akagi T[4], Oe H[5], Nakagawa K[1], Takaya Y[1], Kijima Y[1], Sato S[3], Kanazawa S[3], Ito H[3].

Comment: Cardiac CT is becoming an increasingly important tool for the assessment of cardiac anatomy and function, and has proven value in the anatomic assessment of secundum ASD. However, hemodynamic assessment, and pulmonary to systemic blood flow ratio with CT, has not been performed. The current study compared sizes of a secundum ASD as measured with cardiac CT (128-slice CT scanner, Siemens Medical Solutions), with transesophageal echocardiography, and the pulmonary to aortic blood flow ratio (Qp/Qs) with invasive catheterization. Fifty patients were included to undergo cardiac CT. Heart rate was lowered to <60 bpm using B-blockade, and contrast agent was given with an injection technique causing difference in contrast density between the right and left side of the heart. Sizes of the ASD as measured with CT were comparable to TEE ($r=0.960$; $p<0.001$); Qp/Qs as measured with CT was comparable to invasive catheterization ($r=0.786$; $p<0.001$). Radiation dose was relatively high, as the authors used retrospective gating methods. However, the results of the current study, and the rapid reduction in radiation exposure as CT technology develops, make cardiac CT an increasingly attractive imaging modality to anatomically and hemodynamically assess secundum ASD patients.

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J Cardiovasc Magn Reson. 2015 Jul 9;17:55. doi: 10.1186/s12968-015-0156-7.

Single centre experience of the application of self navigated 3D whole heart cardiovascular magnetic resonance for the assessment of cardiac anatomy in congenital heart disease.

Monney P[1], Piccini D[2,3,4], Rutz T[5], Vincenti G[6], Coppo S[7,8], Koestner SC[9], Sekarski N[10], Di Bernardo S[11], Bouchardy J[12], Stuber M[13,14], Schwitter J[15].

Comment: Cardiovascular Magnetic Resonance Imaging plays an important role in the assessment and follow-up of patients with congenital heart disease. Free-breathing protocols are increasing performed to assess cardiac anatomy in children, and patients having difficulty to follow breathing instructions. Predominantly, respiratory gating is used to obtain high quality images. As an alternative, a motion correction algorithm has been developed to produce 3D CMR images, which is based on cardiac self-navigation. The current study describes 138 unselected patients

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with congenital heart disease ≥2 years old (23.4 +/- 12.2 yrs) who underwent CMR using the cardiac motion correction algorithm to assess 1. image quality, 2. accuracy and reproducibility of morphological analysis, 3. coronary artery visualization, and 4. great vessel diameters. Image quality was sufficient for adequate image analysis in 90% of cases, and was considered good to excellent in 70% of cases. Correct morphological analysis was achieved in >93% of cases, and the proximal course of the coronary arteries could be visualized 90% of cases. Reproducibility of great vessel diameters was high. Image quality was predominantly influenced by high heart rate, low ejection fraction and younger age. This self-navigated 3D CMR seems a valuable expansion for cardiac assessment in patients with congenital heart disease, especially in those with difficulty to follow breathing instructions.

Heart Failure Section

Comments by Jouke Bokma

Int J Cardiol. 2015 Jun 18;197:209-215. doi: 10.1016/j.ijcard.2015.06.018. [Epub ahead of print]

Renin-angiotensin-aldosterone system genotype and serum BNP in a contemporary cohort of adults late after Fontan palliation.

Burchill LJ[1], Redington AN[2], Silversides CK[1], Ross HJ[1], Jimenez-Juan L[1], Mital S[3], Oechslin EN[1], Dragulescu A[3], Slorach C[4], Mertens L[3], Wald RM[5].

Comment: This study evaluated associations between RAAS genotype, BNP levels and ventricular mass and function in a contemporary cohort of 106 adults after the Fontan operation. Patients were considered high-risk (n=31) if 2-5 homozygous RAAS risk genotypes were present or low risk if <1 pro-hypertrofic homozygous RAAS genotype was present. The prevalence of RAAS genotypes was comparable with the general population. Perhaps surprisingly, patients with high-risk genotype had similar blood pressure and ventricular mass on CMR when compared to low-risk patients. However, high-risk genotype was associated with higher BNP levels and diastolic dysfunction on echocardiogram. Fontan failure (heart failure admission, transplantation or death) occurred in 21patients during a mean of 9.5 years of follow-up. Multivariable analysis revealed that only higher baseline BNP remained predictive for Fontan failure (HR 1.11 [CI 1.01–1.23] for each 50 unit increase, p = 0.04). Importantly, predictors commonly associated with acquired heart disease, such as ventricular mass or ejection fraction were not predictive for Fontan failure. The findings of this study confirm the prognostic value of BNP in adult Fontan patients, RAAS genotype impact BNP levels and diastolic function in Fontan patients. The authors suggest accelerated ventricular fibrosis as opposed to hypertension and secondary ventricular hypertrophy may play a crucial role in disease progression of patients with high-risk RAAS genotype. Unfortunately, late gadolinium enhancement or T1-mapping CMR was available in an insufficient number of patients to test this hypothesis.

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Int J Cardiol. 2015 Oct 1;196:1-6. doi: 10.1016/j.ijcard.2015.05.142. Epub 2015 May 28.

Pulmonary outflow obstruction protects against heart failure in adults with congenitally corrected transposition of the great arteries.

Helsen F[1], De Meester P[1], Van Keer J[2], Gabriels C[1], Van De Bruaene A[2], Herijgers P[3], Rega F[4], Meyns B[4], Gewillig M[5], Troost E[2], Budts W[6].

Comment: Previous studies revealed that pulmonary outflow tract obstruction (POTO) reduces systemic AV valve regurgitation in adults with congenitally corrected transposition of the great arteries (ccTGA). The aim of this study was to determine the impact of native or surgically induced POTO on event-free survival. At first visit, ventricular function, systemic AV valve regurgitation and clinical status were assessed in 62 patients with ccTGA. At first visit, systemic AV valve regurgitation ≥ 3/4, systemic RV dysfunction ≥ moderate, and CHF were present in 26%, 26%, and 15% of patients, respectively. None out of 39 Patients with significant POTO had ≥ 3/4 systemic AV valve regurgitation as opposed to 16 (42%) out of 38 patients without significant POTO. During a mean follow-up of 10.1 ± 6.1 years, 40% of patients experienced clinical events (heart failure, transplantation or death). In multivariablee analysis, both systemic ventricular dysfunction (HR: 1.89; 95% CI: 1.05–3.37;*P* = 0.033) and AV valve regurgitation (HR: 1.99; 95% CI: 1.01–3.92; *P* = 0.048) were associated with the occurrence of adverse clinical events. Patients with significant POTO were less likely to develop significant systemic AV valve regurgitation (HR: 0.18; 95% CI: 0.05–0.58; *P* = 0.004) during follow-up. These findings illustrate the importance ventricular-ventricular interaction in patients with a systemic RV as septal shifts influence the systemic AV valve complex. The results of this study suggest pulmonary artery banding can be useful as palliative procedure or to perform LV retraining for possible late arterial switch operation.

Cardiac Care Associates

Comments by Susan Fernandes

Int J Cardiol. 2015 May 6;187:219-26. doi: 10.1016/j.ijcard.2015.03.153. Epub 2015 Mar 18.

Height, weight and body mass index in adults with congenital heart disease.

Sandberg C[1], Rinnström D[2], Dellborg M[3], Thilén U[4], Sörensson P[5], Nielsen NE[6], Christersson C[7], Wadell K[8], Johansson B[2].

Comment: Sandberg and colleagues examined the distribution of body mass index (BMI) in adults with congenital heart disease utilizing the Swedish Registry on Congenital Heart Disease.

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Compared to a reference population with an overweight/obesity (BMI >25) rate of 43% (which is much lower than the U.S. which has rates of 70%), men with complex congenital heart disease and in particular, subjects with pulmonary atresia/double outlet right ventricle and those with Fontan operation had lower rates of overweight/obesity (15% and 22% respectively). However, women with atrioventricular defects were more likely to be obese (BMI >30) compared to the reference group (12.8% versus 9%). Height was also noted to be less in men with complex lesions although heights measured for females was not that different than controls.

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Congenit Heart Dis. 2015 May 23. doi: 10.1111/chd.12273. [Epub ahead of print]

Patient Preference and Perception of Care Provided by Advance Nurse Practitioners and Physicians in Outpatient Adult Congenital Clinics.

Maul TM[1], Zaidi A[2], Kowalski V[3], Hickey J[2], Schnug R[2], Hindes M[3], Cook S[3].

Comment: This study was a collaborative effort between two high volume U.S. ACHD centers that utilize nurse practitioners (NPs) both in shared office visits and in independent clinics. The study sought to describe the experience (satisfaction) of patients being seen by physicians and NPs. A total of 371 subjects were included. Physician-managed practices had higher perceived satisfaction as measured by "excellent" responses compared to NPs. However, the overwhelming majority of subjects were very satisfied with the delivery of care provided by the NP (82% NP versus 90% MD) and had responded that they "strongly agreed" with the statement that they had confidence/trust in their provider (NP 85% versus 94%). One area identified that could use improvement is the patients' understanding of the NP role and how it differs from a nurse or physician. The authors of this article encouraged development of written and multi-media material to improve patients' understanding of the roles of all ACHD team members.

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Am J Cardiol. 2015 Aug 1;116(3):452-7. doi: 10.1016/j.amjcard.2015.04.041. Epub 2015 May 9.

Implementation of the American College of Cardiology/American Heart Association 2008 Guidelines for the Management of Adults With Congenital Heart Disease.

Goossens E[1], Fernandes SM[2], Landzberg MJ[3], Moons P[4].

Comment by Pamela Miner (NP (UCLA))

Comment: This article by Goossens and colleagues investigated the type of "health care professional, care setting, and frequency of outpatient visits" in a group of young adults with CHD. The authors identified 306 patients, age 23 years at time of data collection, who had an outpatient cardiology appointment before the age of 18 years (between 2001-2005). On follow up, 68.8% of patients were in cardiac care, but only one-quarter had transitioned to a formalized ACHD clinic. A small number (7%) of patients had withdrawn from care and 25% were "untraceable." Factors associated with patients being "untraceable" were lower number of surgeries, insurance issues, and being non-white. This study highlights that many adults continue to be cared for by pediatric cardiologists. The authors suggest that standardization of ACHD specialty care might allow pediatric cardiologists to have the confidence to transfer adult patients to ACHD care more routinely.

Job Announcements

Adult Congenital Heart Disease (ACHD) Cardiologist

The University Health Network/Mount Sinai Hospital (UHN/MSH), Department of Medicine, is seeking a qualified full-time Adult Congenital Heart Disease (ACHD) Cardiologist. The candidate would ideally have an academic position description of Clinician-Teacher or Clinician- Investigator with an interest and demonstrated excellence in carrying out independent research, teaching and patient care activities. The successful candidate must be eligible for a full-time clinical academic appointment at the rank of Assistant, Associate, or Full Professor at the University of Toronto. Effective start date is July 1, 2016 or earlier. The successful candidate will report to the Director of the ACHD Program and Head, Division of Cardiology at the University Health Network/Mount Sinai Hospital. This individual will provide exemplary patient-centred care to outpatients and inpatients with ACHD. He/she will have completed a full training program in ACHD (at least 18 months training at a tertiary care centre) with an adult or pediatric cardiology background. He /she will have a strong track record in clinical skills, education, research, and interpersonal skills, to interact with the many different disciplines making up the ACHD team. The successful candidate must hold an MD degree, be eligible for certification in the Royal College of Physicians and Surgeons of Ontario in cardiology and have, or be eligible, for licensure in Ontario. The Adult Congenital Heart Disease Program at the Peter Munk Cardiac Centre of the University Health Network is a national referral centre, and world-renowned clinical, research and education centre for ACHD. The program continues to rapidly grow in size and complexity with the multi-disciplinary care it provides through collaboration of specialists in ACHD, interventional cardiology, electrophysiology, heart failure/transplant, cardiac surgery, anesthesia, medical imaging, genetics and obstetrics. Educating the next generation of ACHD physicians is a mission of the centre, and the successful candidate will be expected to participate in patient care conferences, ACHD academic rounds and other education rounds. UHN is Canada's largest research hospital, and has a history of innovative research and important discoveries. Research and advancement of the field of ACHD are considered essential components of this position. Estimated remuneration will be in the range of $250,000 - $300,000 and commensurate with academic rank, qualifications and experience. Applications will be accepted until **October 31, 2015** or until the position is filled.

Should you be interested in this opportunity, please forward a one page statement of interest, curriculum vitae and the names of three referees to:

Dr. Michael Domanski, Division Head, Cardiology, C/O Amy Anderson, Toronto General Hospital, 4N-484, 585 University Ave., Toronto, Ontario M5G 2N2

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For more information about the Department of Medicine, please visit our website: http://www.deptmedicine.utoronto.ca/Page11.aspx

All qualified candidates are encouraged to apply; however, Canadians and permanent residents will be given priority.