Thank You For Your Continued Membership. We Value You As A Member!

May, 2012

Do you need to update your Membership Information?

Click here

In This Issue

International Society for Adult Congenital Heart Disease

ISACHD Newsletter

Presidents Message

by Curt J Daniels

Dear ISACHD Members,

As I write this month's message, I am preparing and looking forward to next week's International Symposium on ACHD in Toronto. Looking at the title of the conference and realizing it is the 22nd annual symposium, makes me realize not only the incredible history of the conference itself, but also the forward thinking by the masters in our field to develop an international conference over two decades ago when only a few international ACHD programs existed. Therefore, once again colleagues and friends with the sole purpose to advance the field of ACHD and improve patient care will meet, discuss, educate, plan, and reaffirm our commitment to a patient population that deserves our undivided attention and focus. Chair Erwin Oechslin has developed an outstanding program that will meet the needs of the participants no matter the background or training in ACHD.

ISACHD has two distinct programming contributions to this year agenda. First, Barbara Mulder, MD, immediate past-president of ISACHD, will deliver a lecture under the opening session entitled The Top 10 Challenges After Saving Lives for ..... We look forward to Barbara's lecture and her vision of ISACHD and ACHD care. Second, is a session entitled ISACHD: ACHD Around the World featuring active members of ISACHD and those leading work group initiatives. The session will continue to raise the awareness of ISACHD to the ACHD community and demonstrate the current work by ISACHD members and work groups to develop ACHD Global projects and initiatives.

Curt J. Daniels
President

MD
The Pennsylvania State University
Hershey, PA, USA

Representative, South America
Luis Alday, MD
Hospital de Niños Córdoba,
Argentina

Representative, Europe
Helmut Baumgartner, MD
Adult Congenital And Valvular Heart Center
Universitatsklinikum Muenster
Germany

Representative, Asia-Pacific Koichiro Niwa, MD
Chiba Cardiovascular Center
Chiba, Japan

Representative, Canada
Dr. Erwin N Oechslin
Toronto General Hospital
Toronto,
Ontario, Canada

Representative, IACHD Nursing Network Desiree Fleck, PhD, CRNP
Children's Hospital of Philadelphia
Philadelphia, USA

Journal Watch Editor
Philip Moons, PhD, RN
Catholic University Leuven
Leuven, Belgium

Executive Director
Peter Kralka
FPMR
Raleigh, NC, USA

Regional News:

News from Canada

Erwin Oechslin

CACH Network Elections

It is my great pleasure to welcome Dr. Ariane Marelli, Montreal, as new Vice-President, and Dr. Jasmine Grewal, Vancouver, as Treasurer.

Board of Directors:

- President: Erwin Oechslin, Toronto
- Vice-Presidents: Candice Silversides, Toronto, and Dr. Ariane Marelli, Montreal
- Treasurer: Dr. Jasmine Grewal, Vancouver
- Secretary: Dr. Andrew Crean, Toronto
- Nursing Representative: Jaimee Feldstein, Vancouver
- Administrative Assistant: Angela Kennie, Toronto

22nd ACHD Symposium in Toronto

ACHD Meeting Toronto 2012 or www.uhn.ca/ACHDConference2012.asp

The Symposium is less than two weeks away! Please join us for an excellent program to be held at the **Toronto Marriott Downtown Eaton Centre Hotel from May 30 - June 2, 2012**. Many of you took advantage of the reduced registration fee for paying CACH Network members.

ISACHD will be present at the Toronto Symposium. The plenary session entitled "ACHD around the World" has been organized in close collaboration with the ISACHD Board Members.

This symposium, organized in partnership with the Oregon Health & Science University in Portland (Oregon), Cincinnati Children's Hospital Medical Centre, University of Cincinnati (Ohio), and University of Toronto, is a unique forum where medical professionals can meet their colleagues from around the world and exchange ideas and information in the field of

Past Presidents
Gary Webb, MD
1994-1996

Carol Warnes, MD
1996-1998

Richard Liberthson, MD
1998-2000

Daniel Murphy, Jr., MD
2000-2002

Thomas P. Graham, Jr., MD
2002-2004

Michael A. Gatzoulis, MD, PhD, FACC
2004-2006

Jack M. Colman, MD
2006-2008

Michael J. Landzberg, MD
2008-2010

Barbara Mulder, MD
2010-2012

Adult Congenital Heart Disease.

'Beyond Saving Lives' is the theme of the symposium. Thousands of lives of children have been saved, but we are facing new challenges in the growing and aging congenital heart disease population. In addition to the common long-term complications such as heart failure, arrhythmias, the Fontan disease, pulmonary arterial hypertension, and others, we are challenged with end of life questions and the provision of advanced/supportive care, which has been ignored and neglected in our young patients so far. A plenary session (**Beyond Saving Lives or Code 'Palliative': Quality of Death and Dying Experience**) and a workshop (**Advanced Care Planning in Adult Congenital Heart Disease**) will cover this hot topic amongst many others.

Dr. Ivan Rebeyka, Edmonton: Distinguished speaker of the Dr. William G. Williams Lecture
Dr. Gary Webb, Cincinnati was the inaugural speaker of this lecture, which was introduced in honor of the Dr. Williams' contribution to the field of congenital heart disease in 2008. Dr. Ivan Rebeyka will be the distinguished speaker at this year's meeting; the title of his presentation is *Hypoplastic Left Heart Syndrome--*
New Challenges of a New Generation of CHD Patients.

Abstracts
There were 66 abstract submissions: 51 abstracts have been accepted for poster presentation and three abstracts have been accepted for oral presentation. A special thank you to Dr. Lucy Roche, our Abstract Co-ordinator, and Joann Beck, her assistant, who worked very hard to organize the abstract submissions and presentation.

Winner of the Young Investigator's Award
Losartan Therapy has Prominent but Highly Variable Effects on Circulating TGF-β in Marfan Patients
Franken R, Radonic T., den Hartog AW, van Eijk M, Groenink M, de Witte P, Timmermans J, Scholte AJ, van den Berg MP, Pals G, Mulder BJM, Zwinderman AH.
Academic Medical Centre, Amsterdam, Netherlands

Winner of the Nursing Research Initiative Award
Atrial Tachycardia in Adults with Tetralogy of Fallot
Byrne KH, Nagy CD, Holmes KW, Crosson JE.
Johns Hopkins University, Baltimore, MD, United States and Adult Congenital Heart Disease Program, Peter Munk Cardiac Centre, Toronto General Hospital, University of Toronto, Toronto, ON, Canada

I expect the symposium to provide the best of science and education in this area of practice. The program is designed for all healthcare professionals working with Adult Congenital Heart Disease (ACHD) and should familiarize participants with the latest clinical information on the diagnosis and management for ACHD. Nursing and Congenital Cardiac Care Associates will also have a major educational role within this Program.

Please visit ACHD Meeting Toronto 2012 or

www.uhn.ca/ACHDConference2012.asp for more information.

Online Registration is still open and hotel rooms are available for special rates (single and double room for only $189.00 CDN). **Online Registration is available at ACHD Meeting Toronto 2012 or www.uhn.ca/ACHDConference2012.asp**

Cardiac Pathology and Imaging Course in Calgary: June 21-23, 2012
The 3rd Cardiac Pathology and Imaging Course will be held in Calgary from June 21-23,2012. An internationally renowned faculty with Prof. Robert Anderson as distinguished guest from Europe will answer burning questions on pathology and imaging. More information is available at CACH Network or www.cachnet.ca. This course is endorsed by IACHD. There is a limit of 55 participants for the course.

News from United States of America
Bill Davidson

Recent highlights include

- The US is still moving toward an ACHD subspecialty with exams and board certification
- ACC is funding ACHD education for general cardiologists in the state chapters, many states (22 of 50 so far?) have indicated an interest.

Also, the AHA is in November, and is accepting now abstract submissions until June 6, 2012. AHA will have 15 tracks including one on congenital heart disease and child health, which will carry both peds-specific, ACHD-specific, and general sessions such as RV imaging, univentricular hearts, emerging ACHD populations, etc. Session dates 11/3-7/12. Sessions will be in Los Angeles from .

International travel tips: http://my.americanheart.org/professional/Sessions/ScientificSessions/RegistrationHousing/International-Travel-Tips_UCM_323226_Article.jsp

THE ACHD REVIEW

The ACHD Review April 2012

The following review is provided as a service for US ACHD professionals from The American College of Cardiology's Adult Congenital and Pediatric Cardiology Section's ACHD Working Group. The ACHD Review is a quarterly update of current ACHD activities provided by members of the ACC ACHD Working Group, invited ACHD experts, and edited by ACHD WG co- chairs Gary Webb, MD, FACC and Curt J Daniels, MD, FACC. The ACHD Review is an informal communication vehicle and as such the comments and views expressed may not be those of the ACC.

Last month we convened in Chicago for ACC.12, and for those who were able to attend, the meeting proved to be a rewarding experience for CHD/ACHD cardiologists and CCAs. The scientific sessions were well

planned and well organized, and the presentations were outstanding. But, by far, the highlight of the meeting for many, including myself, was Jane Somerville, the 2012 Dan G. McNamara lecturer. There are generational differences as to our professional relationship with Dr Somerville. Most in the CHD profession have either heard of Jane Somerville, read her papers, seen her lecture, worked with her in some capacity or were trained by her. Especially in the ACHD world, the GUCH (Grown-Ups with Congenital Heart Disease) population, the acronym itself, is attributed most to Dr. Somerville. Therefore, with her place in CHD history and along with her well recognized candid, truthful, straightforward approach, knowing that she was going to present at ACC raised quite a bit of anticipation.

Her lecture was scheduled for 2:00, and at least 30 min prior, there was a sizable crowd gathering outside the doors of N228 McCormick Convention Center. It was as if we were waiting for a concert to begin with general admission seating. The doors flew open and everyone quickly lined up to enter the conference hall. At one point the door attendant got a little excited and called out to everyone to settle down, line up and enter slowly. It wasn't until about 10 min after 2:00 Jane Somerville took the stage. She didn't disappoint. Her historical account of CHD surgery and surgeons was....
Click here to read the entire ACHD Review

News from Asia Pacific
Koichiro Niwa

1. JCS Meeting in Fukuoka
During our roundtable regarding the establishment of the ACHD facility in Japan, current and prospect facilities were introduced. To our surprise we had a lot of attendants standing in the room and at several satellite booths. It feels that the ACHD field is now mandatory for cardiologists, even in Japan. The reason might be that in Asian countries leaders of ACHD fields are usually pediatric cardiologists, not cardiologists. Apparently this trend is changing in Japan.

2. APSACHD meeting
The 14th The South China International Congress of Cardiology was held 11-14, April, 2012 in Guangzhou at the Dongfang Hotel in China. Several papers and lectures on ACHD were presented. In China, the field of ACHD is just in the beginning stage, but several centers have an interest in establishing ACHD clinics.

3. The 5th ACHD Seminar will be held in June 9-10 in Tokyo. This National Educational Meeting attracts usually 200-250 attendees, which include physicians, co-medicals, and patients.

News from Latin America
Luis Alday

During the next Argentine Congress of Cardiology to be held at the Buenos Aires Sheraton Hotel from 5 to 7 October 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 neighbouring countries with guest speakers from all over the world.

WG on Education
Erwin Oechslin
Chair, WG on Education, Toronto (Canada)

Cincinnati 2011 ACHD Course Material Is Available Online!
A great deal of excellent teaching and learning material about adult congenital heart disease is available for your use at https://cincinnatiachdcourse.org. The two major types of resources are 'Topic-based Learning' and 'Case-based Learning,' material which was recorded at the 2011 ACHD Program in Cincinnati and is of great value to physicians, cardiac care associates, and other health care providers with an interest in ACHD. There is also a direct link from CACH Network (http://www.cachnet.ca) or The Nevil Thomas ACHD Library (www.achd-online.com). There is a five-star scoring system, and Dr. Gary Webb would be very grateful for feedback from users as to how valuable each presentation was to them. Users will also note a help button to teach them how to use the system.

Under the leadership of Dr. Gary Webb, Cincinnati, we are collecting information of educational tools and resources, which should be made available to the ACHD community. Please send links of educational resources to gary.webb@cchmc.org; these links will be posted at www.isachd.org.

Endorsement of ACHD Meetings
The following meetings have been endorsed:

- 22nd International Symposium on Adult Congenital Heart Disease, Toronto, May 30 - June 2, 2012 (www.uhn.ca/ACHDConference2012.asp or www.isachd.org).
- The 3rd Resident Canadian Cardiac Pathology and Imaging Course, Calgary, Alberta; June 21-23, 2012 (www.isachd.org or www.cachnet.ca)

Please be proactive and contact me at erwin.oechslin@uhn.ca if you want to endorse your ACHD meeting.

Basic Teaching Course
Dr. Els Pieper, UMS Groningen (NL), in collaboration with Dr. Gary Webb, has finalized the program for a Web-based two-day teaching course for trainees with a background in adult cardiology. I have a large pool of excellent speakers who will be receiving an email in these days. Those who want to contribute to this program are asked to identify their preferred topic(s).

Journal Watch

ARTICLE OF THE MONTH: May 2012
Commentary on "Left Ventricular Longitudinal Function Predicts

Life-Threatening Ventricular Arrhythmia and Death in Adults with Repaired Tetralogy of Fallot" by Marc Cribbs

Tetralogy of Fallot (ToF) is the most common form of congenital cyanotic heart disease [1-2]. Late survival after repair is excellent with 35-year survival reaching 85% [3]. Despite excellent outcomes after surgery, there remains a concerning incidence of sudden cardiac death (SCD) during long-term follow-up with an estimated annual incidence of 0.5% per year [4,5]. Tachyarrhythmias, particularly life-threatening ventricular arrhythmia (LTA), appear to be to blame for the majority of these cases [1,6,7]. Identifying risk factors for SCD/LTA in this patient population has been a topic of intense investigation in the US and abroad. Over the past two decades, many related predictors have been proposed. More recently, attention has turned to left ventricular (LV) systolic and diastolic function [8-10]. Khairy et al. have developed a risk score based on clinical history, QRS duration, electrophysiologic assessment, and assessment of LV end-diastolic pressures by cardiac catheterization [11]. Certain echocardiographic parameters of LV function have also been evaluated. While even a mildly depressed LV ejection fraction (LVEF%) has been associated with poor outcome in patients with repaired ToF [12,13], Kempny et al. have recently shown that this may have a low sensitivity in detecting early myocardial damage [14]. Despite the presence of normal LV ejection fraction, 2D peak longitudinal strain (LV-LS) is frequently reduced in these patients [14]. As a result, variables of longitudinal LV function were evaluated in this study to determine if they may be significant predictors of SCD/LTA and superior to LV ejection fraction.

The authors reviewed the charts of 413 ToF patients (51% male) with a composite endpoint of SCD/LTA defined as sustained ventricular tachycardia, resuscitated SCD, or appropriate ICD discharge. Patients were 36 ± 13 years old with a QRS duration of 148 ± 27 msec and an LVEF of 55 ± 10%. Over a median follow-up of 2.9 years, 5 patients died suddenly, 9 had documented sustained VT, and another 5 had appropriate ICD shocks. This equated to an annual probability of SCD/LTA of 2.4%. Using previously published limits of normal values, certain echocardiographic parameters were evaluated [15,16]. Risk factors for the composite endpoint by univariate analysis included QRS-duration, right atrial area >20 cm^2, right ventricular (RV) fractional area change <32%, RV end-diastolic area >28 cm^2, and RV outflow tract diameter >29mm. TAPSE and RV 2D-systolic strain, however, were not. While left ventricular EF and diastolic function were not associated with SCD/LTA, mitral annular plane systolic excursion (MAPSE) <12mm and LV global longitudinal 2-dimensional strain <15% were significantly related. In addition, multivariate analysis identified MAPSE and LV-LS as independently related to the composite endpoint of SCD/LTA.

Identifying risk factors for SCD and LTA in patients with repaired ToF remains a challenge. This study provides important clinical insight and demonstrates for the first time that LV longitudinal dysfunction is associated with SCD and LTA. The reasons for impaired LV function in

ToF are poorly understood. Mechanisms including myocardial ischemia before corrective surgery, severity of pre-operative hypoxemia, and shared myocardium between the left and right ventricle have been suggested to account for LV dysfunction in this setting [17]. While cardiac MRI and invasive testing provides very useful clinical and prognostic information, they are not as readily available in many centers. The parameters evaluated in this study were significantly related to SCD/LTA and are obtained from routine transthoracic echocardiograms. This, as the authors suggest, will continue to be the "workhorse" in patients with ToF, especially those with ICD's.

Bibliography

1. Siwik, E.S.E., Francine; Zahka, Kenneth G.; Goldmuntz, Elizabeth, Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adults. 7th Edition ed, ed. H.D.D. Allen, David J.; Shaddy, Robert E.; Feltes, Timothy F. Vol. 2. 2008, Philadelphia: Lippincott Williams & Wilkins.
2. Bashore TM. Adult congenital heart disease: Right ventricular outflow tract lesions.
Circulation. 2007;115:1933-1947.
3. Warnes, C.A., et al., ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol, 2008. 52(23): p. e1-121.
4. Nollert G, Fischlein T, Bouterwek S, Bohmer C, Klinner W, Reichart B. Long-term survival
in patients with repair of tetralogy of fallot: 36-year follow-up of 490 survivors of the first year
after surgical repair. Journal of the American College of Cardiology. 1997;30:1374-1383.
5. Hickey EJ, Veldtman G, Bradley TJ, Gengsakul A, Manlhiot C, Williams WG, Webb GD,
McCrindle BW. Late risk of outcomes for adults with repaired tetralogy of fallot from an
inception cohort spanning four decades. European journal of cardio-thoracic surgery : official
journal of the European Association for Cardio-thoracic Surgery. 2009;35:156-164.
6. Deanfield, J.E., W.J. McKenna, and K.A. Hallidie-Smith, Detection of late arrhythmia and conduction disturbance after correction of tetralogy of Fallot. Br Heart J, 1980. 44(3): p. 248-53.
7. Rosing, D.A., et al., Long-term hemodynamic and electrocardiographic assessment following operative repair of tetralogy of Fallot. Circulation, 1978. 58(3 Pt 2): p. I209-17.
8. Khairy P, Harris L, Landzberg MJ, Viswanathan S, Barlow A, Gatzoulis MA, Fernandes SM,

Beauchesne L, Therrien J, Chetaille P, Gordon E, Vonder Muhll I, Cecchin F. Implantable
cardioverter-defibrillators in tetralogy of fallot. Circulation. 2008;117:363-370.
9. Ghai A, Silversides C, Harris L, Webb GD, Siu SC, Therrien J. Left ventricular dysfunction
is a risk factor for sudden cardiac death in adults late after repair of tetralogy of fallot. J Am Coll
Cardiol. 2002;40:1675-1680.
10. Geva T, Sandweiss BM, Gauvreau K, Lock JE, Powell AJ. Factors associated with impaired
clinical status in long-term survivors of tetralogy of fallot repair evaluated by magnetic
resonance imaging. J Am Coll Cardiol. 2004;43:1068-1074.
11. Khairy P, Dore A, Poirier N, Marcotte F, Ibrahim R, Mongeon FP, Mercier LA. Risk
stratification in surgically repaired tetralogy of fallot. Expert Rev Cardiovasc Ther. 2009;7:755-
762.
12. Broberg CS, Aboulhosn J, Mongeon FP, Kay J, Valente AM, Khairy P, Earing MG,
Opotowsky AR, Lui G, Gersony DR, Cook S, Ting JG, Webb G, Gurvitz MZ. Prevalence of left
ventricular systolic dysfunction in adults with repaired tetralogy of fallot. Am J Cardiol.
2011;107:1215-1220.
13. Davlouros PA, Kilner PJ, Hornung TS, Li W, Francis JM, Moon JC, Smith GC, Tat T,
Pennell DJ, Gatzoulis MA. Right ventricular function in adults with repaired tetralogy of fallot
assessed with cardiovascular magnetic resonance imaging: Detrimental role of right ventricular
outflow aneurysms or akinesia and adverse right-to-left ventricular interaction. J Am Coll
Cardiol. 2002;40:2044-2052.
14. Kempny A, Diller GP, Orwat S, Kaleschke G, Kerckhoff G, Bunck AC, Maintz D,
Baumgartner H. Right ventricular-left ventricular interaction in adults with tetralogy of fallot: A
combined cardiac magnetic resonance and echocardiographic speckle tracking study.
International journal of cardiology. 2012; 154:259-64.
15. Lang RM, Bierig M, Devereux RB, Flachskampf FA, Foster E, Pellikka PA, Picard MH,
Roman MJ, Seward J, Shanewise J, Solomon S, Spencer KT, St John Sutton M, Stewart W.
Recommendations for chamber quantification. European journal of echocardiography : the
journal of the Working Group on Echocardiography of the European Society of Cardiology.
2006;7:79-108.
16. Marcus KA, Mavinkurve-Groothuis AM, Barends M, van Dijk A,

Feuth T, de Korte C,
Kapusta L. Reference values for myocardial two-dimensional strain echocardiography in a
healthy pediatric and young adult cohort. Journal of the American Society of Echocardiography :
official publication of the American Society of Echocardiography.
2011;24:625-636.
18. Tzemos N, Harris L, Carasso S, Subira LD, Greutmann M, Provost Y, Redington AN,
Rakowski H, Siu SC, Silversides CK. Adverse left ventricular mechanics in adults with repaired
tetralogy of fallot. The American journal of cardiology.
2009;103:420-425.

Commentary:
Marc G Cribbs, MD
Fellow, Columbus Ohio Adult Congenital Heart (COACH) program
Wexner Medical Center at The Ohio State University
Nationwide Children's Hospital
Columbus, OH USA
Marc.Cribbs@osumc.edu

Circulation. 2012 Apr 11. [Epub ahead of print]
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
1 Univ Hospital of Münster, Germany & Royal Brompton Hosp & Imperial College, London, UK;
Abstract
Background: Sudden cardiac death and life-threatening ventricular arrhythmias (SCD/LTA) remain a concern in adult patients with repaired tetralogy of Fallot (ToF). 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 ToF patients (age 36±13years, QRS duration 148±27ms, LV-EF 55±10%). A composite endpoint of SCD/LTA (sustained ventricular tachycardia, resuscitated SCD or appropriate ICD discharge) was employed. During a median follow-up of 2.9 years 5 patients died suddenly, 9 had documented sustained VT and another 5 had appropriate ICD shocks. On univariate Cox analysis QRS-duration (hazard ratio [HR] 1.02/ms, P=0.046), right atrial area (HR 1.05/cm2, P=0.02), right ventricular (RV) fractional area change (HR 0.94/%, P=0.02), RV outflow tract diameter (HR 1.08/mm, P=0.01), mitral annular plane systolic excursion (MAPSE, HR 0.84/mm, P=0.03) and LV global longitudinal 2-dimensional strain (LV-LS, HR 0.87/%, P=0.03) were related to the combined endpoint. On bivariable analysis MAPSE and LV-LS 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 and RV fractional area change as well as LV-LS or MAPSE was also found to be significantly related to outcome (P<0.001, c-statistic 0.70).
Conclusions: Left ventricular longitudinal dysfunction was associated with greater risk of SCD/LTA. 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 when estimating prognosis in this challenging population.

AACN Adv Crit Care. 2012 Apr;23(2):142-54.
APN Plan Improves Outcome for Pregnant Patient With Congenital Heart Disease.
Haynes A, Frederick A, Chirkoff A.
Source
Annette Haynes is Cardiology CNS, Stanford Hospital and Clinics, 300 Pasteur Drive, Room H0101, M/C 5221, Stanford, CA 94305 (anhaynes@stanfordmed.org). Andrea Frederick is Congenital Heart NP, Stanford, California. Andrea Chirkoff is Assistant Nurse Manager/Educator, Maternal Child Health Services, Kaiser Permanente Medical Center, San Francisco, California.
Abstract
Advanced practice nurses work in many roles to support delivery of safe patient care. Eighty-five percent of children born with congenital heart disease (CHD) live to adulthood. The pregnant adult with CHD presents challenges for nursing across many care-delivery systems. Progression of care delivery across these systems requires innovative planning and organization. This article describes the plan developed by advanced practice nurses in a CHD clinic and in inpatient coronary care and obstetric units to support a pregnant patient with CHD. The plan focused on collaboration and communication among interdisciplinary teams. The goal was to address multidisciplinary communication, leadership, and staff education. The result was a successful high-risk delivery with organized education and patient care across systems.

Eur J Cardiothorac Surg. 2012 Apr 26. [Epub ahead of print]
Natural and modified history of single-ventricle physiology in adult patients.
Angeli E, Pace Napoleone C, Balducci A, Formigari R, Lovato L, Candini L, Oppido G, Gargiulo G.
Source
Pediatric and Grown-up Congenital Cardiac Surgery, University of Bologna, S.Orsola-Malpighi Hospital, Bologna, Italy.
Abstract
Objective: To define the evolution of the single-ventricle (SV) heart in adult patients in terms of morbidity, mortality and quality of life.
Methods: Sixty-two patients with SV physiology and aged older than 16 years were retrospectively reviewed. Three patients (5%) were in natural history, one had received a Blalock-Taussig shunt, one a Waterstone anastomosis, one a pulmonary artery banding, three a bidirectional cavopulmonary anastomosis, eight a classic Fontan procedure and 46 a total cavopulmonary connection (TCPC). The morphology of the SV was

left in 48 patients (77%), right in nine (14%) and indeterminable in five (8%). Thirty-three patients underwent magnetic resonance imaging (MRI) to assess ventricular mass (VM), ventricular systolic function, pulmonary artery branch diameter and potential thrombosis of the conduit. Cardiopulmonary exercise testing (CPTE) was carried out to evaluate exercise tolerance. The quality of life was monitored with two different specific tests, the Short Form-36 (SF-36) and the congenital heart disease-TNO/AZL adult quality of life (CHD-TAAQOL). The mean follow-up time was 8.0 ± 9.1 years.
Results: Two of the three patients in natural history underwent primary TCPC. Re-interventions were necessary in seven patients (11%). Three patients (5%) died during follow-up. Five patients (8%) underwent cardiac transplantation. Protein losing enteropathy appeared in six (10%), while the arrhythmic disorder was detected in 13 patients. On the MRI, the mean end-diastolic ventricular volume was 106 ± 448 ml/m(2), the mean ejection fraction (EF) was 52.3 ± 10% and VM was 56 ± 22.1 g/m(2). On CPTE, the peak of oxygen uptake (peak VO(2)) was moderately impaired in 92% of patients, while 4% presented a severely impaired and 4% a normal peak of VO(2). No correlations were found among the peak of VO(2) and the quality-of-life evaluation.
Conclusions: Adult patients with SV are at high risk of reoperations and need of transplant and complications. Nevertheless, in the presence of a moderately reduced peak of VO(2) and a moderate reduction in the EF detected at the MRI, the results of the evaluation of daily quality of life are incredibly high.

Congenit Heart Dis. 2012 Apr 27. doi: 10.1111/j.1747-0803.2012.00658.x. [Epub ahead of print]
Electrophysiology Procedures in Adults with Congenital Heart Disease.
Ermis P, Franklin W, Kim J, Moodie D, Parekh D.
Source
Department of Pediatric Cardiology, Texas Children's Hospital, Baylor College of Medicine, Houston, Tex, USA.
Abstract
Background: In adult congenital heart disease (CHD), arrhythmias contribute significantly to morbidity and mortality. Often, these adult patients are treated at a freestanding pediatric facility. Limited data exist looking at this cohort.
Methods: A retrospective review was performed of all electrophysiology (EP) procedures performed in adults at our institution during a 5-year period from January 1, 2006 through December 31, 2010.
Results: There were 99 cases performed in a total of 87 adults with CHD during this time period. The mean patient age was 27.1 years (18-51 years). The most common congenital cardiac diagnoses were: 27% with D-transposition of the great arteries (n=27)-of which 85% (n=23) have had a previous atrial switch procedure, 20% with tetralogy of Fallot (n=20), and 16% with previous Rastelli repair (n=16). Overall, 37 EP studies were performed, with the majority done in patients with complex CHD. There were 74 additional cases. These procedures consisted of: 38 pacemakers (51%), 26 implantable cardiac defibrillators (36%), six laser lead extractions (8%), two loop recorders (3%), and two pocket revisions (3%). During this 5-year period, there was one major complication (1%) and seven minor complications (7%).
Conclusions: The complex care of adults with CHD requiring EP

procedures can be safely and effectively accomplished in a freestanding pediatric hospital with low complications, provided institutional support of an adult CHD program.

Respirology. 2012 Apr 18. doi: 10.1111/j.1440-1843.2012.02180.x. [Epub ahead of print]

Partial Anomalous Pulmonary Venous Connection and Pulmonary Arterial Hypertension.

Sahay S, Krasuski RA, Tonelli AR.

Source

Department of Medicine, Akron General Medical Center, Akron, Ohio, USA Adult Congenital Heart Disease Services. Department of Cardiovascular Medicine, Heart and Vascular Institute, Cleveland Clinic, OH, USA Department of Pulmonary, Allergy, and Critical Care Medicine, Respiratory Institute, Cleveland Clinic, Ohio, USA.

Abstract

SUMMARY AT A GLANCE: Isolated partial anomalous pulmonary veins are often overlooked in the diagnostic work up of the pulmonary arterial hypertension. In the clinical practice these cases are often labeled as primary pulmonary hypertension. Our case-series highlights the development of pulmonary arterial hypertension in individuals with isolated partial anomalous pulmonary veins.

Background and objective: Isolated partial anomalous pulmonary venous connection (PAPVC) has been implicated as a cause of pulmonary arterial hypertension (PAH), however this condition is often overlooked in the diagnostic work up of patients with PH. We studied the prevalence of PAH both in patients with isolated PAPVC or associated with other congenital heart diseases (CHD) such as ASD. We also aimed to identify factors related to the presence of PAH in these patients.

Methods: We retrospectively analyzed data from the Adult CHD database at the Cleveland Clinic, USA between October 2005-2010. We included all patients diagnosed with PAPVC with or without other CHD. We excluded all patients with previous corrective surgeries.

Results: We identified 14 (2.5 %) patients with the PAPVC. Group I included patients with PAPVC (with or without PFO). Group II included patients with PAPVC associated with other CHD. PAH was seen in six (6/14, 42.8 %) patients, two (2/7, 28.5%) in group I and four (4/7, 57.1%) in group II (p=0.3). The mean pulmonary artery pressure in all patients (n=14) was 29.5 ± 13.8 mmHg. Group I had a mean PAP of 23.6 ± 6.6 mmHg as compared to 33.7 ± 16.5 mmHg for group II (p=0.34). The two patients in group I with PAH had either two anomalous pulmonary veins or a condition (sickle cell disease) that could potentially explain the hemodynamic findings.

Conclusions: Patients with PAPVC (with or without PFO) in the absence of other CHD had normal PAP unless they have two pulmonary veins with anomalous return or associated conditions known to cause PAH.

Echocardiography. 2012 Apr 11. doi: 10.1111/j.1540-8175.2012.01679.x. [Epub ahead of print]

Monitoring Complex Secundum Atrial Septal Defects Percutaneous Closure with Real Time Three-Dimensional Echocardiography.

García-Fuertes D, Mesa-Rubio D, Ruiz-Ortiz M, Delgado-Ortega M, Tejero-Mateo I, Pan-Álvarez-Ossorio M, Suárez-de-Lezo J, Lafuente M.

Source
Department of Cardiology, Hospital Universitario Reina Sofía, Córdoba, Spain.
Abstract
Background: Atrial septal defect (ASD) is one of the most common congenital heart diseases. Nowadays, percutaneous closure is considered the treatment of choice in most of secundum ASDs. Assessment of the defect and procedure monitoring have been usually performed by angiographic balloon-sizing and/or two-dimensional (2D) transesophageal echocardiography. However, in complex ASDs these techniques might be inaccurate.
Methods: From January 2009 to January 2011 all adult patients with complex ASDs submitted for percutaneous closure were selected. Those defects, where shunts were present through a device previously implanted on the atrial septum or through multiperforated septums, were considered complex ASDs. Two-dimensional transesophageal echocardiography and real time three-dimensional (3D) echocardiography were performed simultaneously during the percutaneous closure procedure. Number of orifices, relationships between the defect, catheter, and device, as well as residual shunt were assessed.
Results: Seven patients were included. Five patients had a multiperforated septum and in two cases the defect in the septum was through a previously implanted device. In all cases, 3D echocardiography was superior to 2D echocardiography in relation to the assessment of the relationship between the defect and the catheter or the device. Mechanisms responsible for residual shunts through a device were also better assessed by 3D echocardiography.
Conclusion: Three-dimensional echocardiography is a safe and useful technique when monitoring percutaneous closure of ASDs, showing relevant advantages over 2D echocardiography. (Echocardiography 2012;**:1-6).

Eur J Cardiothorac Surg. 2012 Apr 4. [Epub ahead of print]
Evaluation of the Aristotle complexity models in adult patients with congenital heart disease.
Hörer J, Vogt M, Wottke M, Cleuziou J, Kasnar-Samprec J, Lange R, Schreiber C.
Source
Department of Cardiovascular Surgery, German Heart Center Munich at the Technical University, Munich , Germany.
Abstract
Objectives: The adult congenital heart disease (CHD) population has surpassed the paediatric CHD population. Half of all mortality caused by CHD occurs in adulthood; in some patients, it occurs during surgery. We sought to assess the potential risk factors for adverse outcome after cardiac operations in adults with CHD, and to evaluate the predictive power of the Aristotle score models for hospital mortality.
Methods: Procedure-dependent and independent factors, as well as the outcome factors of all consecutive patients aged 16 or more who underwent surgery for CHD between 2005 and 2008 at our institution were evaluated according to the European Association for Cardio-Thoracic Surgery Congenital Database nomenclature. An Aristotle basic complexity (ABC) and an Aristotle comprehensive complexity (ACC) score were assigned to each operation. The discriminatory power of the scores was

assessed using the area under the receiver operating characteristics (AuROC) curve.
Results: During 542 operations, 773 procedures were performed. The early mortality rate was 2.4%, and the early complication rate was 53.7%. Tricuspid valve replacement (P = 0.009), mitral valve replacement (P < 0.001), elevated lung resistances (P = 0.002), hypothyroidism (P = 0.002) and redosternotomy (P = 0.003) emerged as risk factors for 30-day mortality. Tricuspid valve replacement (P < 0.001), tricuspid valvuloplasty (P = 0.006), mitral valve replacement (P = 0.003), shunt implantation (P = 0.009), surgical ablation (P = 0.024), myocardial dysfunction (P = 0.014), elevated lung resistances (P = 0.004), hypothyroidism (P = 0.002) and redosternotomy (P < 0.001) emerged as risk factors for complications. Mean ABC and ACC scores were 6.6 ± 2.3, and 9.0 ± 3.7, respectively. The AuROCs of the ABC and the ACC scores for 30-day mortality were 0.663 (P = 0.044), and 0.755 (P = 0.002), respectively. The AuROCs of the ABC and the ACC scores for complications were 0.634 (P < 0.001), and 0.670 (P < 0.001), respectively.
Conclusions: Surgery for adults with CHD can be performed with low early mortality. However, complications are frequent, especially in patients who require repeat operations for atrioventricular valve incompetence. The ACC score may be helpful to estimate the risk of early mortality.

Pediatr Cardiol. 2012 Apr;33(4):601-6. Epub 2012 Feb 10.
Risk factors associated with morbidity and mortality after pulmonary valve replacement in adult patients with previously corrected tetralogy of fallot.
Jain A, Oster M, Kilgo P, Grudziak J, Jokhadar M, Book W, Kogon BE.
Source
Department of Cardiothoracic Surgery, Emory University School of Medicine, Atlanta, GA, USA.
Abstract
Patients with palliated tetralogy of Fallot (TOF) often require pulmonary valve replacement in adulthood, yet the data regarding their outcomes are scarce. This study aimed to identify risk factors associated with postoperative complications in these patients and to establish long-term survival data for this patient group. A retrospective cohort study investigated 153 consecutive patients with a history of TOF repair who underwent pulmonary valve replacement at a single large academic center between March 1996 and March 2010. In part 1 of the study, logistic models were constructed to assess demographic, medical, and surgical risk factors for operative mortality; occurrence of a major adverse event (stroke, renal failure, prolonged ventilation, deep sternal infection, reoperation, or operative mortality); and prolonged hospital stay (>7 days). Risk factors with a p value less than 0.10 by univariate analysis were included in the subsequent multivariate analysis. In part 2 of the study, long-term, all-cause mortality was determined by construction of a Kaplan-Meier curve for the cohort. Seven patients died (4.5%). Significant risk factors for mortality in the multivariable analysis included age older than 40 years (odds ratio (OR) 9.89) and concomitant surgery (OR 6.65). A major adverse event occurred for 22 patients (14.4%). The only significant risk factor in the multivariable analysis for an adverse event was concomitant surgery (OR 6.42). The hospital stay was longer than 7 days for 31 patients (20.3%). The significant risk factors for a prolonged hospital stay included the presence of preoperative arrhythmias (OR 4.17), New York Heart Association class 3 (OR 4.35), and again,

concomitant surgery (OR 4.2). Among the 146 hospital survivors, only 5 patients died in the intervening period. The predicted survival rates were 98.5% at 1 year, 96.7% at 5 years, and 93.5% at 10 years. Pulmonary valve replacement in adults with palliated TOF is a safe procedure with excellent long-term survival, but there remain important risk factors for postoperative mortality, prolonged hospital stay, and major adverse events. Awareness and modification of important risk factors may help to improve outcomes.

Interact Cardiovasc Thorac Surg. 2012 Apr;14(4):440-4. Epub 2012 Jan 25.

Surgical outcome of partial Shone complex.

Grimaldi A, Vermi AC, Ho SY, Pappalardo F, Castiglioni A, Benussi S, Zangrillo A, Alfieri O.

Source

Cardiovascular and Thoracic Department, San Raffaele Scientific Institute and Università Vita-Salute San Raffaele, Milan, Italy.
antonio.grimaldi@hsr.it

Abstract

Partial forms of Shone complex are rare. Surgical outcomes of the complete forms have generally been poor, whereas there is a lack of data on long-term follow-up of surgically treated adult partial complex. Between 2001 and 2011, nine patients (age: 38 ± 8 years; six males, 67%) were referred for valvular heart disease. Transthoracic and transoesophageal echocardiography was performed. Data were confirmed by intra-operative findings and reports. Patients were diagnosed as partial Shone complex and presented with mitral stenosis (MS) (45%) or mitral regurgitation (22%) or aortic regurgitation (22%). All but one patient (89%) reported previous surgery: coarctation of the aorta repair (87.5%) and aortic valvulotomy (12.5%). Redo intervention included: mitral valve replacement (25%), mitral repair (25%), aortic valve replacement (37.5%) and subvalvular aortic ridge resection (25%). One patient refused surgery. Patients surgically treated before the age of 5 (87.5%) showed favourable outcome (survival rate: 100%) and a 23.6 (± 4.6)-year follow-up free from events. The patient who underwent first intervention at the age of 50 and the patient with MS who refused surgery showed a 45 (± 7)-year follow-up free from major morbidity. Patients with partial Shone complex, properly diagnosed and treated, show favourable surgical outcome free from major clinical events.