Regional News

News from Asia Pacific

In January of this year, we had our 12th Annual Congress of the Japanese Society for ACHD, which was held in Osaka, Japan. The main topics of this time were PH and pregnancy. There were 300 attendees at the meeting. In Thailand, ACHD Clinic was opened in the Mahidol University Hospital Bangkok. Also, Koichiro Niwa was invited to give an honorary lecture on ACHD at the Thai Heart Association Meeting in Bangkok at the end of March. Now more cardiologists are interested in this field, too. In the three countries Forum (Korea, Japan, China), the topics on ACHD were presented by Dr. Gu Hong, China Anzhen Hospital. The number of ACHD patients was huge and every year over 100,000 CHD babies were born in China. In July, the 2nd Congress of APSACHD (Asia Pacific Society for ACHD) will be held in Tokyo. Mike Lanzberg MD, Carole A. Warnes, MD, and Michael A. Gatzoulis, MD, will join us, and the joint Session of ISACHD, Euro GUCH and APSACHD will be held. (http://www.appcs.org/2010/apsachd.htm)

News from Canada

CCS 2009 Consensus Conference on the Management of Adults with Congenital Heart Disease

Erwin Oechslin, President, CACH Network

The Canadian Cardiovascular Society (CCS) 2009 guidelines on the Management of Adults with Congenital Heart Disease are published! The Canadians pioneered the development of guidelines on the management of adults with congenital heart disease. In 1996, CACH Network members and a panel of national and international experts were the first group to publish guidelines for the management of adults with CHD under the leadership of Gary Webb and Michael Connelly. Since the publication of the 2001 CCS guidelines (updated 1996 guidelines), there have been significant advances in the understanding of the late outcomes, genetics, and medical and interventional therapy in this growing population of adults with congenital heart disease.
ISACHD Spring 2010 Newsletter

The complete document has been revised and updated under the leadership of Candice Silversides (Toronto) and Judith Therrien (Montreal) and consists of four manuscripts. As president of the CACH Network, I express my sincere thanks to Candice Silversides and Judith Therrien for their leadership on this project and acknowledge the contributions of the section editors, authors of the different chapters, and of the national and international experts who critically reviewed the document. The international panel members have given this document an international flavor.

The CCS 2009 Consensus Conference on the Management of Adults with Congenital Heart Disease will be posted on the CACH Network Website at www.cachnet.ca or CCS Website www.ccs.ca. There is also a link on the ISACHD Website (www.isachd.org) to the CACH Network Website.

News from Europe
Folkert Meijboom, chairman of the GUCH Working Group of the ESC

It has been a successful year for GUCH-activities in Europe. After a very busy ESC annual meeting in Barcelona with many GUCH sessions on the program and a joint session with ISACHD. Together with ISACHD we had a very well-attended social event on a roof-restaurant overlooking Barcelona’s harbour.

The next event was the 4th European Echocardiography course on Congenital Heart Disease. This edition was organized in Rotterdam, The Netherlands and the success of the previous editions was repeated. Participants of 26 countries, mainly from Europe but actually from all over the globe, participated. For the first time the website www.echocardiography-course.com was used and this proved to be a big hit. This will be the website also for the future editions. An upcoming event will be in London in October of 2010.

EuroEcho was held in Madrid this year - again Spain, after the ESC meeting in Barcelona. The local organizer prof. Zamorano had done an excellent job. The sessions on congenital heart disease - both paediatric and adult congenital heart disease - were very well attended. The accreditation examination for echo for congenital heart disease was held again, just like during the previous years. It is not known yet how many of the participants have passed the examination. New for 2010, is that this accreditation examination will be held at the AEPCC (Association of European Paediatric Cardiology) meeting, this year in Innsbruck in May. The 2010 edition of EuroEcho will be held in Copenhagen, again in December.

Guidelines for the use for CMR in congenital heart disease have been made by a writing group headed by Philip Kilner of London, UK. It was a joint effort of the ESC working groups on GUCH and on Cardio Magnetic Resonance. These guidelines are accepted for publication in the European Heart Journal and will be published soon.

Upcoming is the First advanced course on GUCH organized by the ESC Working Group on GUCH, on Wednesday, April 14, and Thursday, April 15, 2010, back-to-back to the well-established Cardiology in the Young (CITY) meeting in London, where it will be held. The program looks excellent. Many have booked already but there is still place left and GUCH cardiologists, nurses and surgeons are more than welcomed.

ISACHD Past presidents:
Gary Webb 1994-1996
Carol Warnes 1996-1998
Richard Liberthson 1998-2000
Daniel Murphy, Jr 2000-2002
Thomas Graham 2002-2004
Michael Gatzoulis 2004-2006
Jack Colman 2006-2008
Michael Landzberg 2008-2010

Highlights from the ACC
by Desiree Fleck

Adults with CHD providers were well represented this year including a highlighted ACPC section during this ACC in Atlanta. There were 5 spotlight sessions, 13 symposiums, 2 core curriculum, 7 expert sessions, 2 debates 1 session on fellows in training and 1 session on Mentoring centering on congenital cardiology. In addition, we had the First Dan G. McNamara Lecture honoring Charles Mullins. Dr. Mullins has contributed much to the world of congenital heart disease. He entertained us with the history of CHD. This was in conjunction with the first special session devoted to quality improvement in congenital heart disease. We saw a glimpse into the future of the IMPACT database.

This year there was plenty of sessions regardless of your area of interest including transition, education, imaging, pregnancy and sexuality in adults with CHD.

An excellent session was presided by Adrienne Kovacs, PsychD and Aida Sawai, MD on sexual issues for Adults with CHD including a presentations on “talking about sex with adolescents”, “erectile dysfunction”, and “reproductive issues”, to name a few.

Additionally, there were 3 International Collaborative Seminars including one from Italy and one form Turkey focusing on adult issues such as pregnancy and the care of patients with Tetralogy of Fallot. We had eight poster presentations from across the globe including Belgium, Japan, the Netherlands and North America. One notable poster was a collaborative effort between the Adult Congenital Cardiac Care Associates research group and the ACHA on the Parental Knowledge of Lifelong Care Study. Other posters included a session on single and right ventricles in ACHD including TGA, fontans and Tetralogy of Fallot. Exercise prescriptions, transfer and transition destinations and quality of life were other interesting sessions. Finally, there was a session dedicated to...
welcomed to register even at the last minute.

Contact details can be found in the flyer in this newsletter. The website is www.cityguch.com.

The biggest event for 2010 for our WG will be the presentation of the new guidelines. The ESC guidelines for GUCH have been revised by a writing group, chaired by prof Helmut Baumgartner from Munster, Germany. They will be presented during the annual ESC congress, and published subsequently in the European Heart Journal.

The ESC meeting this year will take place in Stockholm from August 28 until September 1. This congress will have an unprecedented number of seven pre-arranged sessions dedicated to GUCH - normally a small working group like the WG on GUCH is entitled to have three sessions - and it is expected that many other sessions and poster presentations will follow. All in all very useful to visit the ESC annual meeting and enjoy the beautiful city of Stockholm as well.

**News from Latin America**

During the next World Congress of Cardiology to be held in Beijing, China, from June 16 to 19, 2010 there will be several sessions related to Adult Congenital Heart Disease with participation of physicians and nurses members of ISACHD.

**Thursday 17th June**

- **Saint Lawrence Level 4 10:30-12:00**
  Symposium 27.04 Adult congenital heart disease and surgery
  Catheter interventions for adult congenital heart disease

- **Saint Lawrence Level 4 14:00-15:30**
  Symposium 27.04 Adult congenital heart disease and surgery
  ACHD: a rapidly growing population

- **Mississippi Level 4 14:00-15:30**
  Symposium 3.04 Surgery for valve disease
  Percutaneous intervention in valve disease

**Friday 18th June**

- **Saint Lawrence Level 4 14:00-15:30**
  Symposium 27.04 Adult congenital heart disease and surgery
  Atrial septal defect (ASD)

- **Thames Level 3 16:00-17:30**
  Symposium 4.04 Pulmonary circulation, other
  Pulmonary circulation: 2010

- **Saint Lawrence Level 4 16:00-17:30**
  Clinical Seminar 27.06 Percutaneous intervention
  Catheter interventions update in congenital heart disease

**Saturday 19th June**

- **Niger Level 2 14:00-15:30**
  Clinical Seminar 28.01 Acute nursing care
  Acute care of adults with congenital heart disease

**Publications**

*Left ventricular noncompaction associated with hypertrophic cardiomyopathy and Wolff-Parkinson-White syndrome*

Luis Alday, Eduardo Moreyra, Eva Bruno, Norma Rossi, Hector Maisuls

Abstract | Reference | Full Paper | PDF (Size:303K), pp.200--203
DOI: 10.4236/health.2010.23029

**News from the United States**

1. A very good ACHD program was included in the ACC's Annual Scientific Sessions for 2010 in Atlanta (just concluded), this followed a strong ACHD program at the AHA last November.

**improving ACHD imaging including increasing the use of transthoracic echo for evaluating RV volumes in Tetralogy of Fallot.**

**Nursing Network News**

**CALL FOR ABSTRACTS!!! The 20th Annual International Symposium on ACHD**

The deadline for abstract submission for Skamania is coming up on April 15th. This is a great opportunity to share your research with other ACHD nursing/PA colleagues, and possibly foster new ideas for multi-center projects.

We look forward to a "Special Nursing Symposium" on Sunday evening June 6th, as a forum to showcase oral research presentations. All ACHD nurses, physician assistants, psychologists and social workers are invited to submit abstracts for this session. It's clear that "non-physician providers" make a significant impact in the care of adults with CHD, and their research deserves this special recognition and showcase. Abstracts will be reviewed by the board of the Adult Congenital Cardiac Care Associate (ACCCA) research group. Five abstracts will be accepted for oral presentation during this Sunday evening symposium, and the other accepted abstracts will be presented as posters throughout the meeting. Original work that has been presented at other meetings, but is not yet in print, will be considered.

Abstract submissions will only be accepted electronically, and are limited to 500 words (excluding title, authors, and institution). Abstract acceptance does not include conference registration fee or hotel accommodations.

**ABSTRACT SUBMISSION DEADLINE:**
April 15th, 2010 (Email abstracts to pminer@mednet.ucla.edu)

**NOTIFICATION OF ACCEPTANCE:**
May 1st, 2010

**ORAL PRESENTATIONS:**
June 6th, 2010 (7:15pm-9:30pm)

**POSTER PRESENTATIONS:**
Throughout Meeting

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**ACC.11 & i2 Summit Call for Suggestions**

Now open through May 12, 2010

by Daniel Murphy, Jr., M.D.

Dear Colleagues, I am the topic coordinator for Pediatric and Congenital Cardiology for the 2011 ACC Scientific Sessions. The committee and I are requesting your help in developing the program. We can use any suggestions for program content, speakers, etc. I am contacting you specifically for suggestions regarding content in the area of Adult Congenital Cardiology, but I would be interested hearing your thoughts on what would be valuable at the ACC meeting.

Please respond in one of the following ways:

1. Contact me by email (murphyd@stanford.edu) with your suggestions and ideas.
2. Go to the ACC website (see below) to propose sessions. (Fully planned sessions have a high likelihood of appearing on the Program.)

Please contact me with any questions or ideas. The value of the Scientific Program depends upon your good ideas.

Thanks in advance for your help,
Daniel Murphy

Full Paper: www.cityguch.com
2. The upcoming Skamania Lodge ACHD Symposium June 6-9, 2010 co-chaired by David Sahn, Erwin Oechslin, Gary Webb, Craig Broberg and Pamela Miner and co-sponsored by the OHSU and the ACC. This is one of the longest running, best-established ACHD Symposia in the US dating back to origins in San Diego in the early nineties. The venue is on the Columbia River gorge on the Washington State side but not too far east of Portland, Oregon. International travelers could choose to fly into Portland and either rent a car or take a shuttle to Skamania, or fly into Seattle and rent a car if more touring in the Pacific NW is a consideration. Skamania and The Toronto ACHD Symposium have alternated years, more recently a third site was introduced, Philadelphia in 2007 and now Cincinnati in 2011. The scheduled Skamania Lodge Symposium for 2009 was delayed by a year to facilitate attendance at the 4th World Congress of Pediatric Cardiology and Cardiac Surgery in Australia last June. For all details the website address is http://www.ohsu.edu/xd/education/schools/school-of-medicine/qme-cme/cme/ACHD.cfm.

3. Many US ACHD physicians and other care providers have been working in the ACHA’s Vision 2020 process to develop long range plans and goals to facilitate growth and maturity of ACHD care in the US. Working groups have addressed regionalization of ACHD care, viable business plans in ACHD care, training of the ACHD workforce and accreditation of ACHD programs. Last April the first vision 2020 Forum was held as part of an ACHA-sponsored ACHA professionals symposium. The Working Groups proposals were presented to a large group and feedback provided/obtained. This April 23 in Washington, DC the second Vision 2020 forum will be held to again present our progress and receive feedback from other ACHD professionals. All US ACHD professionals are invited to attend.

Details of the event and the Phase I recommendations can be found at http://www.achaheart.org/advocacy/V2020_Forum2010.php.

While this is focused on ACHD care in the US, others may find the recommendations pertinent to their own situations. Thoughts on the recommendations and feedback can be provided through the ACHA’s website above. Insights and ideas from ACHD providers worldwide are welcome.

Journal Watch


Abstract: As medical and surgical techniques advance in modern health care, children and adolescents with congenital heart disease (CHD) are living well into their adult years. Their complex cardiac anatomy, physiology, and medical histories present a challenge to adult health care providers who are not traditionally educated and trained in CHD. Growing evidence demonstrates that adolescents and young adults with CHD are at risk for extensive complications and sequelae from their heart condition and surgical treatments. Many of these young adults either continue to be cared for by their pediatric cardiologist or are lost to follow up as they age out of pediatric care. There are little data on the most successful way to transition adolescents and young adults into adult-centered CHD programs. The purpose of this review is to reveal the need for transition into appropriate care and lifelong follow up for adolescents and young adults with CHD, and highlight the role of a pediatric nurse practitioner as a transition coordinator in a pediatric cardiology clinic.


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Abstract: The different biopsychosocial periods in a woman's life are all interactively associated with the cardiovascular system. The present study was designed to address questions related to sexuality and reproductive health in a large cohort of women with congenital heart disease. Overall, 536 women (median age 29 years,
Abstract: OBJECTIVE: The occurrence of aortic dissection after percutaneous closure of a patent foramen ovale for cryptogenic stroke has been reported in a variable percentage of patients. However, its precise incidence and mechanism are presently unclear and remain to be elucidated. DESIGN: Prospective follow-up study. PATIENTS: Ninety-two patients undergoing a percutaneous patent foramen ovale closure procedure (closure group) for cryptogenic stroke were compared with a similar group of 51 patients, who were medically treated. METHODS: A systematic arrhythmia follow-up protocol to assess the incidence of AF was performed including a 7-day event-loop recording at day 1, after 6 and 12 months in patients of the closure group and compared with those of the medically treated group. RESULTS: The incidence of AF was similar in both study groups during a follow-up of 12 months, including 7.6% (95% CI: 3.1-15.0%) in the closure and 7.8% (95% CI: 2.18-18.9%) in the medically treated group (P=1.0). The presence of a large patent foramen ovale was the only significant risk factor for the occurrence of AF as demonstrated by a multivariate Cox regression analysis (95% CI, 1.275-20.018; P=0.021). CONCLUSIONS: Our findings indicate that patients with cryptogenic stroke and patent foramen ovale have a rather high incidence of AF during a follow-up of 12 months. Atrial fibrillation occurred with a similar frequency whether the patent foramen ovale/atrial septal defect was successfully percutaneously closed or was medically managed. The presence of a large patent foramen ovale was the only significant predictor of AF occurrence during follow-up.


Abstract: BACKGROUND: Ventricular septal defects (VSDs) are one of the most common congenital heart defects in adults. In adult patients with an anatomically large VSD and relatively preserved pulmonary vascular system, several pulmonary flow-limiting cardiac morphologic alterations (PFMA) are encountered. PATIENTS: Ninety-eight male patients (mean age 22.5 +/- 2 years) operated for an anatomically large VSD in our institute were retrospectively reviewed. PFMA in patients with an anatomically large but functionally mild-to-moderate VSD (when ratio of pulmonary to systemic flow (Qp/Qs) < 2.2 and ratio of pulmonary to systemic vascular resistance (Rpx/Rsy) < 0.3) were recorded. RESULTS: Thirty patients (31.2%) revealed a mild-to-moderate VSD in unctional severity. Five PFMA were encountered in these patients: (1) ostium (os) infundibulum (n = 10, 33.3%), (2) aneurysm of the membranous septum (AMS) (n = 10, 33.3%), (3) systolic bulging of the conal septum toward the right ventricular outflow tract (n = 6, 20%), (4) prolapse of the aortic cusps (n = 2, 6.7%), and (5) attachment of the tricuspid septal leaflet to the septal crest (n = 2, 6.7%). Double-chambered right ventricle was encountered in four patients with os infundibulum and classic tetralogy-type septal malalignment in one patient with aortic cusp prolapse. Concurrent to VSD repair, resection of the os infundibulum and the AMS and aortic valve repair were performed. CONCLUSION: Presence of a large VSD and relatively preserved pulmonary vascular system in adults is associated with several PFMA. Preoperative awareness and concurrent surgical treatment of these alterations seem to be crucial to improve the expected benefit of surgical repair of VSD in this subgroup of the patients.


Abstract: Cardiovascular magnetic resonance (CMR) imaging provides highly accurate measurements of biventricular volumes and mass and is frequently used in the follow-up of patients with acquired and congenital heart disease (CHD). On reproduducibility are limited in patients with CHD, while measurements should be reproducible, since CMR imaging has a main contribution to decision making and timing of (re)interventions. The aim of this study was to assess intra-observer and interobserver variability of biventricular function, volumes and mass in a heterogeneous group of patients with CHD using CMR imaging. Thirty-five patients with CHD (7-62 years) were included in this study. A short axis set was acquired using a steady-state free precession pulse sequence. Intra-observer and interobserver variability of biventricular function, volumes and mass was assessed for left ventricular (LV) and right ventricular (RV) volumes, function and mass by calculating the coefficient of variability. Intra-observer variability was 2.9 and 6.6%, respectively. Interobserver variability was between 3.9 and 10.2%. Overall, variabilities were smallest for biventricular end-diastolic volume and highest for biventricular end-systolic volume. Intra-observer and interobserver variability of biventricular parameters assessed by CMR imaging is good for a heterogeneous group of patients with CHD. CMR imaging is an accurate and reproducible method and should allow adequate assessment of changes in ventricular size and global ventricular function.


Abstract: OBJECTIVES: The treatment of Stanford type B aortic dissections involving the arch or associated with proximal aortic aneurysms remains a surgical challenge. We report our results with total arch replacement with the stented elephant trunk (SET) procedure for these complicated Stanford type B aortic dissections. METHODS: BETWEEN December 2003 and June 2008, 31 patients were admitted for complicated type B dissection (12 acute, 19 chronic). The mean age at operation was 44.3 +/- 10.6 years (range: 22-68 years). The surgeries were performed by using total arch replacement combined with SET implantationEnhanced computed tomography (CT) was performed before discharge as well as 3 months and annually to evaluate the condition of the graft and the residual false lumen. RESULTS: The procedure was successful in all but two patients; two patients died of multiple organ failure following surgery. No paraplegia was observed after surgery. Follow-up was completed in 27 of 29 patients and the mean follow-up period was 18.4 +/- 12.3 months (range: 6-54 months). During follow-up CT scans, thrombus formation was observed in the descending aortic false lumen excluded by the stented graft in most patients. One patient died during follow-up while two patients with Marfan syndrome underwent successful operations for replacement of the descending and subsequent abdominal aorta. CONCLUSION: Total arch replacement with the SET procedure has emerged as a viable option for complicated type B dissections and is associated with low morbidity and mortality. At mid-term follow-up, most patients have either thrombosed or have had no further increase in the false lumen of the descending aorta.


Abstract: OBJECTIVE: Tricuspid regurgitation is often associated in patients with congenital heart disease. Significant morbidity and mortality are related to tricuspid valve replacement. Tricuspid valve plasty is still a preferred choice. This report deals with our surgical experience in using the edge-to-edge valve plasty technique to correct severe tricuspid regurgitation in patients with congenital heart disease. METHODS: From December 2002 to August 2007, severe tricuspid regurgitation was corrected with a flexible band annuloplasty and edge-to-edge valve plasty technique in nine patients with congenital heart disease. The age ranged from 7 to 62 years (average 24.4 years). Congenital cardiac anomalies included atioventricular canal in five cases, secundum atrial septal defect in three cases, and cor triatriatum in one case. RESULTS: No hospital death or postoperative morbidity occurred. No or trivial tricuspid regurgitation was present in six cases and mild...
tricuspid regurgitation in three cases at discharge. The follow-up ranged from 12 months to 70 months (average 39.3 months). No tricuspid stenosis was found. No to mild tricuspid regurgitation was present in eight cases, and moderate tricuspid regurgitation in one case at the latest follow-up. CONCLUSIONS: Edge-to-edge valve plasty is an easy, effective, and acceptable additional procedure to correct severe tricuspid regurgitation in patients with congenital heart disease.


Abstract: BACKGROUND: To evaluate the safety, reproducibility and pitfalls of an aortic valve re-implantation (AVRei) technique. METHODS: From June 2005 to December 2008, 30 patients underwent aortic valve sparing re-implantation with Gelweave Valsalva prosthesis. Mean age was 66 +/- 7 years (range 47-81). Mean aortic root diameter was 49 +/- 6 mm (range 37-70) and 12 patients had an aortic insufficiency more than 2+. All the patients were elective, except three who underwent surgery for type A aortic dissection. Two patients had Marfan syndrome and one had a bicuspid aortic valve. Isolated aortic root replacement was performed in 26 patients, whereas hemiarch extension was required in four. All the survivors underwent serial echocardiographic assessment for functional results and multi-detector computed tomography (MDCT) for aortic root morphology evaluation. RESULTS: There was one early death and one re-exploration for bleeding. Two patients suffered from a perioperative stroke and four required a pacemaker implantation because of a complete atrio-ventricular block. Mean follow-up was 12 +/- 10 months (range 1-42) with no late deaths. Whereas freedom from reoperation was 100% and freedom from aortic insufficiency 2+ or more was 96.5%. MDCT aortic root reconstruction showed a pseudo-normalization of the neo-sinususes of Valsalva mimicking the human normal aortic root morphology. CONCLUSION: AVRei with Valsalva conduit is a well-tolerated procedure both in elective and emergency situations. In well-selected patients, good functional and clinical results can be achieved, regardless of the cause of the aortic root disease. Application of simple surgical manoeuvers allows durable clinical efficacy to be obtained without the risk of major complications.


Abstract: Neurohormonal activation is prevalent in adults with congenital heart disease, but its relation to outcome remains unknown. B-type natriuretic peptide (BNP) and atrial natriuretic peptide (ANP) were measured prospectively in 49 patients with adult congenital heart disease, who were followed up for a median of 7.9 years (interquartile range 7.7 to 8.2). Cox proportional hazards regression analysis was used to determine the relation of BNP and ANP concentrations to all-cause mortality. The mean age at baseline was 33 +/- 11.3 years, and 46.9% of patients were men. Most patients (77.5%) were asymptomatic (20.4% had New York Heart Association class III). 10 (20.2%) were cyanotic, and 28 (57.1%) had systemic ventricular dysfunction (moderate or severe in 18.4%). The mean concentration of BNP was 52.7 pg/ml (interquartile range 39.1 to 115.4) and of ANP was 47.4 pg/ml (interquartile range 19.7 to 112.8). Of the 49 patients, 11 (22.4%) died during the follow-up period. Both BNP and ANP were strong predictors of mortality (hazard ratio per 100-pg/ml increase 1.80, 95% confidence interval 1.38 to 2.34, p < 0.0001; and hazard ratio per 100-pg/ml increase 1.21, 95% confidence interval 1.12 to 1.32, p < 0.0001, respectively). A BNP value >78 pg/ml predicted death with a sensitivity of 100% and specificity of 76.3% (area under the curve 0.91, p = 0.0001). An ANP value of >146 pg/ml predicted death with a sensitivity of 72.7% and specificity 94.7% (area under the curve 0.89, p = 0.0001). No patients with a BNP level >78 pg/ml died during the follow-up period. In conclusion, the BNP and ANP levels strongly predicted death in asymptomatic ambulatory patients with adult congenital heart disease during mid-term follow-up and could be used as a simple clinical marker for risk stratification in this population.


Abstract: The experience with echocardiographic diagnosis of five cases of Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) is reviewed. MATERIAL AND METHODS: all cases with a diagnosis of ALCAPA during a 10 year period were included. RESULTS: two age groups were clearly identified: infants and older patients. In the former, the echocardiographic findings included a dilated left ventricle with low ejection fraction (19% and 26%), mitral regurgitation, dilated right coronary artery and difficulties in identifying the origin of the left coronary artery. An 11-year-old asymptomatic boy, a 17-year-old young woman with dyspnea on effort and a 55-year-old woman with angina formed the older group. In these three cases, an abnormal upward flow was detected within the ventricular septum related to the collaterals and the inverse flow of the anterior descending artery. A reverse flow within the flow of the left coronary artery, probably related to an origin in the pulmonary artery, was observed. In all five cases the diagnosis was corroborated using selective right coronary artery angiography. CONCLUSIONS: In the infants, the dilated left ventricle with impaired systolic function, mitral regurgitation and dilated right coronary artery make it necessary to discard the ALCAPA diagnosis. In the older group, even in asymptomatic patients, an abnormal ascending flow within the ventricular septum, combined with a dilated coronary artery, was the most reliable echocardiographic evidence for a diagnosis of ALCAPA.


Abstract: Patients after coarctation repair still have an increased risk of cardiovascular or cerebrovascular events. This has been explained by the persisting hypertension and alterations in the peripheral vessels. However, involvement of the central vessels such as the retinal arteries is virtually unknown. A total of 34 patients after coarctation repair (22 men and 12 women; 23 to 58 years old, age range 0 to 32 years at surgical repair) and 34 nonhypertensive controls underwent structural and functional retinal vessel analysis. Using structural analysis, the vessel diameters were measured. Using functional analysis, the endothelium-dependent vessel dilation in response to flicker light stimulation was assessed. In the patients after coarctation repair, the retinal arteriolar diameter was significantly reduced compared to that of the controls (median 182 mum, first to third quartile 171 to 197; vs 197 microm, first to third quartile 193 to 206; p <0.001). These findings were independent of the peripheral blood pressure and age at intervention. No differences were found for venules. The functional analysis findings were not different between the patients and controls (maximum dilation 3.5%, first to third quartile 2.1% to 4.5% vs 3.6%, first to third quartile 2.2% to 4.3%; p = 0.81), indicating preserved autoregulative mechanisms. In conclusion, the retinal artery diameter is reduced in patients after coarctation repair, independent of their current blood pressure level and age at intervention. As a structural marker of chronic vessel damage associated with past, current, or future hypertension, retinal arteriolar narrowing has been linked to stroke incidence. These results indicate an involvement of cerebral microcirculation in aortic coarctation, despite timely repair, and might contribute to explain the increased rate of cerebrovascular events in such patients.

13. Inadvertent placement of pacemaker lead into the systemic ventricle in repaired D-transposition of the great arteries. Kuppahally SS, Green LS, Michaels AD, Ishihara SM, Freedman RA, Litwin SE in: Circulation 121 (6), 2010 Feb 16, pp. e32 Safety and efficacy of transcatheter closure of large patent ductus arteriosus in adults and multi-detector computed tomography (MDCT) for aortic root morphology evaluation. RESULTS: There was one early death and one re-exploration for bleeding. Two patients suffered from a perioperative stroke and four required a pacemaker implantation because of a complete atrio-ventricular block. Mean follow-up was 12 +/- 10 months (range 1-42) with no late deaths. Whereas freedom from reoperation was 100% and freedom from aortic insufficiency 2+ or more was 96.5%. MDCT aortic root reconstruction showed a pseudo-normalization of the neo-sinususes of Valsalva mimicking the human normal aortic root morphology. CONCLUSION: AVRei with Valsalva conduit is a well-tolerated procedure both in elective and emergency situations. In well-selected patients, good functional and clinical results can be achieved, regardless of the cause of the aortic root disease. Application of simple surgical manoeuvers allows durable clinical efficacy to be obtained without the risk of major complications.

Abstract: BACKGROUND: The development of significant tricuspid regurgitation (TR) is associated with an unfavorable clinical outcome in patients with systemic right ventricles. Increased knowledge about the factors contributing to its presence would help prevent its progression. METHODS: This was a retrospective analysis of the factors predictive of significant TR in 60 patients with systemic right ventricles following an atrial switch procedure for complete transposition of the great arteries. Data from echocardiographic examinations, exercise radionuclide angiography, and myocardial perfusion imaging were analyzed. RESULTS: Significant TR was present in 20% of patients. Compared to patients without significant TR, patients with significant TR were older at the time of surgery (p < 0.01), with a higher body mass index (p < 0.005), lower right ventricular ejection fraction (RVEF; p < 0.01), higher exercise perfusion abnormalities score on radionuclide angiography (p < 0.03), and higher systolic blood pressure (p < 0.02). At univariate logistic regression analysis systolic blood pressure (p = 0.03), increasing age at surgery (p = 0.01), and RVEF (p = 0.02), were predictors of significant tricuspid regurgitation. The latter two remained significant at multivariate analysis. Conclusions: Patients operated upon later in life, with decreased RVEF and higher blood pressure, are at risk of significant tricuspid regurgitation and therefore warrant special attention. Prospective studies are needed to ascertain whether appropriate pharmacological intervention would prevent the development and/or progression of TR in these patients.


Abstract: OBJECTIVES: To evaluate whether the stenting of aortic coarctation enhance the risk of exercise-induced hypertension (EIH). BACKGROUND: There is the theoretical concern that aortic stents may cause increased aortic wall impedance and therefore systolic hypertension during exercise. METHODS: Blood pressure and the Doppler derived peak and mean systolic pressure gradient (PSG and MSG) across the distal aorta at the peak of exercise were evaluated in young patients (mean age 14 +/- 3 years) with aortic coarctation successfully treated with surgery or with stent implantation at least 1 year before the test. Only patients who reached the 85th maximal predicted heart rate or whose exercise test was interrupted because of severe hypertension, and in whom significant aortic narrowing was evaluated by a MRI or a CT scan performed in the six months preceding the exercise test were included in the study. RESULTS: Seventeen patients formed the surgery-group, while 15 patients the stent-group. Patients in surgery-group were younger at coarctation repair and with a longer follow-up than those in stent-group. No difference was present regarding age, body surface area, gender, and presence, and degree of mildly hypoplastic aortic segments between the two groups as well as between patients with or without EIH. EIH was found in 35% of surgery-group patients and in 33% of stent-group patients. PSG and MSG were similar in the patients with or without EIH. CONCLUSIONS: EIH can be found in a high number of young patients successfully treated for aortic coarctation but at intermediate follow-up stent implantation does not seem to enhance the risk of EIH.


Abstract: BACKGROUND: Pulmonary arterial hypertension (PAH) may develop in patients with atrial septal defects (ASD); however, little is known about associated risk factors and its evolution after transcatheter ASD closure. METHODS AND RESULTS: We conducted a cohort study on 215 adults with attempted transcatheter ASD closure from 1999 to 2006. Patients were classified according to baseline systolic pulmonary arterial pressures as having no (I, <40 mm Hg), mild (II, 40 to 49 mm Hg), moderate (III, 50 to 59 mm Hg), or severe (IV, > or =60 mm Hg) PAH. Independent predictors of moderate or severe PAH were older age (odds ratio [OR], 1.10 per year; P=0.0001), larger ASD (OR, 1.13 per millimeter; P=0.0052), female sex (OR, 3.9; P=0.001), and at least moderate tricuspid regurgitation (OR, 3.6; P=0.0043). At 15 (interquartile range, 8 to 43) months post-ASD closure, patients with higher baseline pressures were more likely to experience a > or =5-mm Hg decrease (33.7%, 73.9%, 79.2%, and 100.0% in groups I to IV, P=0.0001), with a larger magnitude of reduction (0, 8, 17, and 22 mm Hg; P=0.0001). However, normalization of pressures (<40 mm Hg) occurred less frequently in patients with more advanced PAH (90.2%, 71.7%, 66.7%, and 23.5%, P=0.0001). Among patients with moderate or severe PAH, independent predictors of normalization were lower baseline pressures (OR, 0.91 per mm Hg; P=0.0418) and no more than mild tricuspid regurgitation (OR, 0.14; P=0.0420). CONCLUSIONS: In adults with ASDs, severity of PAH is modulated by age, sex, defect size, and degree of tricuspid regurgitation. Patients with moderate or severe PAH may benefit from substantial reductions in pulmonary artery pressures after transcatheter ASD closure, although the PAH values remain elevated in a sizeable proportion.


Abstract: ISACHD Spring 2010 Newsletter


Abstract: BACKGROUND: Conduits placed in the right ventricular outflow tract (RVOT) have limited longevity which often requires increasingly complex reoperations. Transcatheter pulmonary valve implantation improves conduit hemodynamics through a minimally invasive approach. We present data for 7 patients treated with the Edwards SAPIEN transcatheter heart valve (THV). PATIENTS: Patients’ ranged in age from 16 to 52 years, one was female, and all had NYHA class II-III symptoms. Patients had pulmonary homografts that had been placed 2-25 years earlier during the Ross procedure (n = 5), large ASD (OR, 1.13 per millimeter; P=0.0052), female sex (OR, 3.9; P=0.001), and at least moderate tricuspid regurgitation (OR, 3.6; P=0.0043). At 15 (interquartile range, 8 to 43) months post-ASD closure, patients with higher baseline pressures were more likely to experience a > or =5-mm Hg decrease (33.7%, 73.9%, 79.2%, and 100.0% in groups I to IV, P=0.0001), with a larger magnitude of reduction (0, 8, 17, and 22 mm Hg; P=0.0001). However, normalization of pressures (<40 mm Hg) occurred less frequently in patients with more advanced PAH (90.2%, 71.7%, 66.7%, and 23.5%, P=0.0001). Among patients with moderate or severe PAH, independent predictors of normalization were lower baseline pressures (OR, 0.91 per mm Hg; P=0.0418) and no more than mild tricuspid regurgitation (OR, 0.14; P=0.0420). CONCLUSIONS: In adults with ASDs, severity of PAH is modulated by age, sex, defect size, and degree of tricuspid regurgitation. Patients with moderate or severe PAH may benefit from substantial reductions in pulmonary artery pressures after transcatheter ASD closure, although the PAH values remain elevated in a sizeable proportion.


Abstract: ISACHD Spring 2010 Newsletter


Abstract: OBJECTIVES: We report our 8-year experience with surgery for congenital heart disease in adults in a medium-volume surgical center. METHODS: We collected data of 161 consecutive patients who underwent 230 procedures from January 1997 to December 2004. Surgical procedures were divided into two groups: repair (85.7%) and reoperation (14.3%). RESULTS: There was only one in-hospital death (Fontan revision, in reoperation group). At a mean follow-up time of 56 +/- 30 months, overall survival from any kind of complication is 76.5%. Cox analysis showed that incremental risk factors were preoperative cyanosis, and length of ICU stay (hazard ratio = 4.47, 3.34 and 1.49, respectively; P < 0.001 in all), whereas decremental risk factors were preoperative New York Heart Association class I (hazard ratio = 0.328, P < 0.001) and surgery for ‘septal defect’ (hazard ratio = 0.26, P = 0.02). CONCLUSION: Surgery for congenital heart disease in adults seems to be an overall well-tolerated and a low-risk treatment, with overall good mid-term clinical results.

Abstract: BACKGROUND: The incidence of residual ventricular septal defects (VSDs) after surgery is 5-25%. Redo surgery is associated with higher risks. METHODS: Between January 2000 to December 2008, 170 patients underwent percutaneous VSD closure in our centre: 22(16%) of these had 23 closures for residual VSDs. Median age was 32.5 yrs (1.4-79). All patients had echocardiographic signs of left ventricle volume overload (Q(p)/Q(s) >or= 1.5). Nine patients had previous VSD closure, 6 tetralogy of Fallot repair, and 7, other procedures. There were 15 muscular, 6 perimembranous and 2 apical VSDs. RESULTS: Amplatzer VSD devices were used in all. Median VSD size was 8 mm (4.3-16). Median fluoroscopy time was 33 minutes (15-130). There were three adverse events: 1 ventricular fibrillation requiring DC cardioversion; 1 transient complete atio-ventricular block reverting to sinus rhythm at 24-hours; one patient had transient atrial flutter during the procedure. All procedures were successful; no additional procedures were required. Trivial residual shunts were seen in 3 patients at follow-up. There were no late events. One patient experienced arrhythmic death 5-yrs after procedure. One patient was reoperated due to dehisence of VSD patch 2-yrs after the second successful percutaneous closure. CONCLUSIONS: Transcatheter closure of post-surgical residual VSD is safe and efficacious management option and obviates the need for further surgery and by-pass.


Abstract: SUMMARY OBJECTIVES: We investigated the haemodynamic effect of percutaneous closure of an intra-atrial shunt, using non-invasive finger pressure measurements. BACKGROUND: Percutaneous closure of both patent foramen ovale (PFO) and atrial septal defect (ASD) is widely practised. Currently no data are available on short-term haemodynamic changes induced by closure. METHODS: Twenty-five consecutive patients (mean age 49 +/- 17 years, 10 men) who underwent a percutaneous closure of a PFO (n = 15) or ASD (n = 10) were included in this study. During the procedure blood pressure and heart rate (HR) were monitored continuously with a Finometer. Changes in systolic, mean, and diastolic pressure, stroke volume (SV), cardiac output (CO) and total peripheral resistance (TPR) were computed from the pressure registrations using Modelflow methodology. RESULTS: Baseline characteristics were similar for the PFO and ASD patients. After PFO closure none of the haemodynamic parameters changed significantly. After ASD closure the systolic, mean, and diastolic pressures increased 7.1 +/- 5.4 (P = 0.003), 3.8 +/- 3.5 (P = 0.007) and 2.0 +/- 3.0 mmHg (P = ns) respectively. HR decreased 5.1 +/- 5.3 beats per minute (P = 0.01). SV, CO and TPR increased 8.5 +/- 6.4 ml (13.5%); P = 0.002), 0.21 +/- 0.45 l/min(-1) (5.6%); P = ns) and 0.02 +/- 0.14 dynes (4.1%; P = ns) respectively. The changes in SV differ between the PFO and ASD patients (P = 0.009). CONCLUSIONS: Using non-invasive finger pressure measurements, we found that SV, mean and systolic blood pressure increased immediately after percutaneous closure of an ASD in adults, whereas the percutaneous PFO closure had no effect on haemodynamic characteristics.


Abstract: The association of patent foramen ovale (PFO) and atrial septal aneurysm (ASA) with migraine headache attack (MHA) has been clearly shown. The same findings have been recently demonstrated also in cluster headache. Although tension-type headaches (TTH) are the most common kind of headache, their association with these atrial septal abnormalities has never been studied before. The study was conducted to clarify whether there was a significant association between the presence of such atrial septal abnormalities and tension headache, when compared with migraineurs. One hundred consecutive patients with migraine and 100 age- and sex-matched subjects with TTH and 50 healthy volunteers with no headache were enrolled in the study and underwent a complete transesophageal echocardiographic study with contrast injections at rest and with the Valsalva maneuver. There was no significant difference between the age and the sex of the participants of the three groups. The overall prevalence of PFO was 23% in patients with TTH and that of large PFOs was only 11%. The 23% prevalence of PFO in patients with TTH was not statistically different from 16% found in our normal control group. Furthermore, we found a significantly higher prevalence of PFO in migraineurs (50%) when compared with patients with tension headache (p < 0.001). This was also true for the collective presence of large PFOs and ASAs (35%) (p < 0.001). Although atrial septal aneurysms have an association with MHA, they do not have a significant association with TTH.


Abstract: AIMS: Patients with congenital heart disease usually show diminished exercise capacity and quality of life. However, there is only little information about daily activity, a marker for lifestyle, exercise capacity, and the prevention of arteriosclerosis. This study investigated exercise capacity, quality of life, daily activity, and their interaction with univentricular heart physiology after total cavopulmonary connection (TCPC). METHODS AND RESULTS: Fifty-seven patients (18 females, 39 males, age 8-52 years) after TCPC (lateral tunnel 28, extra-cardiac conduit 29) who underwent surgery during 1994-2001 were examined in our institution. They performed a symptom-limited cardiopulmonary exercise test. Those patients 14 years of age and older filled in the health-related quality-of-life questionnaire SF-36, and those who were 8-13 years of age, the CF-87. Daily activity parameters were obtained by using a triaxial accelerometer over the next three consecutive days. Exercise capacity was severely reduced after TCPC (25.0 ml/min/kg corresponding to 59.7% of age- and sex-related reference values). Daily activity was within the recommendations of the United Kingdom Expert Consensus Group (> or =60 min, > or =3 metabolic equivalent; > or =5 days/week) in 72% of the investigated patients. It was reduced in older patients (Spearman r = - 0.506, P < 0.001) and patients with a lower peak oxygen uptake (Spearman r = 0.432, P = 0.001). In children <14 years, mental health was related to daily activity. CONCLUSION: Despite their diminished exercise capacity, patients after TCPC show a fairly normal activity pattern. However, their activity depends not only on age, but also on exercise capacity, which, in contrast to healthy people, decreases already from early adolescence on.