December 16, 2010

In This Issue

Renew Your Membership Online!

Renew your membership online now.

International Society for Adult Congenital Heart Disease

ISACHD Newsletter

President's Message: Joyful Music is Good For The Heart

by *Barbara J.M. Mulder*

Many people say singing cheerful songs definitely alters their mood, puts them in the right spirit and is a good way to find relaxation. At the AHA congress in 2008, Dr. Miller from Maryland presented his study, "Positive Emotions and the Endothelium: Does Joyful Music Improve Vascular Health?" In this study, the average brachial artery diameter increased 26 percent after joyful music in healthy volunteers. So it suggests that the music was beneficial for the heart. This finding has to be confirmed in patients with congenital heart disease. Since endothelium dysfunction may occur in patients with a variety of congenital heart defects, the cardiovascular effect of joyful music may be different. But it doesn't do any harm to stimulate our patients to enjoy cheerful music in the next few holiday weeks.

In Oklahoma City, a Holiday Party is planned to raise awareness of congenital heart defects, on Saturday December 18th. The event will be held from 1 to 4 p.m., at Boldt Construction Building, 101 W. Hefner Road. All children with congenital heart disease and their families are invited to attend the event. There is no cost to participate. Participants will have the opportunity to meet Santa, participate in craft projects and be entertained by a magician and pet therapy dogs. Light refreshments will be served. The party is provided in conjunction with the American Heart Association of Oklahoma City. A wonderful initiative of the Oklahoma cardiology community!

In 2011, ISACHD executives will present new plans for international Health Care, Education and Research. You are all invited to collaborate and participate in these initiatives. Journal Watch will be distributed separately from the monthly newsletter to all ISACHD members. Suggestions and ideas are very welcome and you can mail them to: b.j.mulder@amc.uva.nl.

ISACHD executives wish you Happy Holidays in good spirit and a collaborative 2011!

Barbara J.M. Mulder
President

Case Study

by *Luis Alday*

Asplenia syndrome with univentricular heart and severe pulmonary stenosis presenting with congestive heart failure in a lady with no previous surgical treatment.

Laura Fortuna, Femando Daghero, Diego Cabral, Monica Benjamin and Luis Alday
Section of Adult Congenital Heart Disease and Cardiovascular Unit. Sanatorio Allende, Cordoba, Argentina.

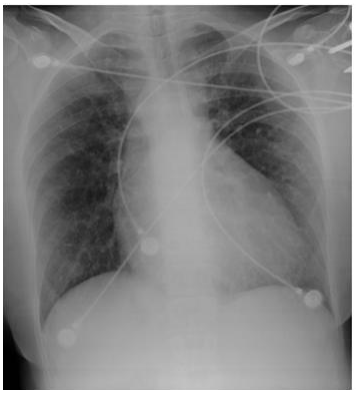

Figure 1: Chest X-ray on admission

This 36-year-old lady with cyanosis since birth was admitted to the hospital for progressive dyspnea on effort, chest pain when exposing to cold weather, palpitations, pain in the upper left abdominal quadrant, unsteadiness and somnolence. A cardiac catheterization performed in infancy concluded that she had situs ambiguous with levocardia, a univentricular heart, and severe pulmonary stenosis. She was offered no treatment on the basis of dim prognosis. However, she was able to thrive and even marry but never got pregnant. At the age of 23 years she suffered an ischemic stroke with right hemiplegia with full recovery. Oral anticoagulation was then started.

The positive findings on physical examination were a normally developed female with marked cyanosis (75% oxygen saturation by pulse oximetry) and clubbed fingers and toes. The blood pressure was 120/80 mmHg and the pulse was irregular with frequent premature beats. The apical impulse was displaced to the left and there was a thrill in the pulmonary area. The 2nd heart sound was loud and single and there was a harsh 4/6 ejection systolic murmur along the left sternal border. The liver was felt at 2 cm below the costal arch on the left of the abdomen.

FPMR
Raleigh, NC, USA

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

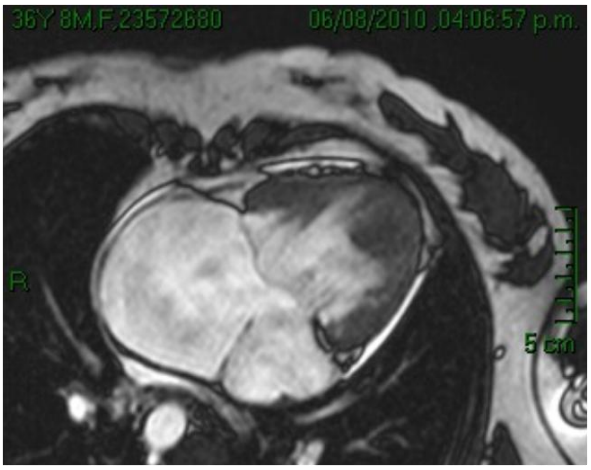

Figure 2: Four-chamber view of magnetic cardiac resonance showing a single ventricle, an ostium primum ASD, and a common AV valve

Pertinent laboratory findings were a hemoglobin of 17.1 g%, 60% hematocrit, serum iron 25 ug/dl, ferritin 10 ng/dl, uricemia 6.9 mg%, prothrombin time (INR) 1.9, and NT-proBNP 1006 pg/ml. The chest X-ray showed levocardia, moderate cardiomegaly, right atrial enlargement, right aortic arch, and decreased pulmonary vascular markings. (Figure 1) The EKG showed 1st degree AV block (PR 0.24"), supraventricular extrasystoles, P waves with changing morphologies suggesting duplicate sinoatrial node, (1) biatrial enlargement, a minus 90 degree QRS axis on the frontal plane, and QS or rS complexes in the precordial leads. Color-Doppler echocardiography and MRI displayed a dilated univentricular heart with depressed contractility (45% EF). There was an enlarged right atrium, an ostium primum atrial septal defect, and a common AV valve with mild regurgitation. (Figure 2) The great arteries were malposed with severe pulmonary stenosis., the aorta arising from an anterior rudimentary chamber. (Figure 3) The aortic arch was to the right as well as the descending aorta. The pulmonary veins drained on both sides of the atrium. There were 2 superior caval veins. The abdominal scan showed an enlarged left sided liver with the stomach to the right and absent spleen. The inferior caval vein was dilated and to the left of the spine. The hepatic veins were also dilated.

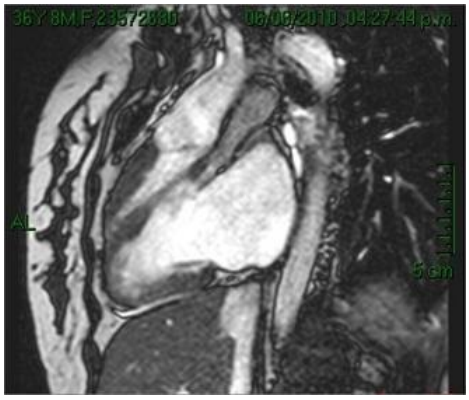

Figure 3:
Lateral view of magnetic cardiac resonance showing the anterior aorta arising from a rudimentary chamber and the small posterior pulmonary artery with severe subvalvar and valvular pulmonary stenosis

A cardiac catheterization (Table 1) showed increased end diastolic ventricular and mean right atrial pressures. The pulmonary artery pressure measured with a wedged catheter in a pulmonary vein was low. The gradient across the pulmonary valve was 120 mmHg. There was birectional shunt at the atrial level and severe systemic unsaturation. The pulmonary and systemic flows were balanced. During the study, catheter manipulation triggered a sustained supraventricular tachycardia with a rate of 280 beats per minute accompanied by marked systemic hypotension. The arrhythmia was successfully terminated with a DC shock.

Table 1
Cardiac catheterization data

Site	Pressure mmHg	Oximetry sat%
Superior caval vein		57
Right atrium	"a" wave 14 (10)	
Wedged pulmonary vein	18/10 (12)	98
Ventricle	149/15	79
Aorta	143/87 (108)	75

Qp: 10.4 l/m/m2 – Qs: 10.5 l/m/m2 – Qp:Qs ratio: 0.9 : 1.0
Rp: 1.2 u/m2 – Rs: 9.3 u/m2 – Rp:Rs ratio: 0.1:1.0

The patient was kept on oral anticoagulation and was treated for congestive heart failure with oxygen, iron therapy, diuretics, and carvedilol. She was discharged 4 days after admission. As an outpatient she states that the shortness of breath, abdominal pain and other symptoms are improved though she still has palpitations. Her present functional class is grade II.

Table 2
Potential risk factors for mortality in total cavopulmonary connection

- ***Age 30 or older***
- ***Heterotaxy***
- ***< 50% ejection fraction***
- **≥ G 3 AV insufficiency**
- ***> Grade III NYHA functional class***
- **Protein losing enteropathy**
- **Fontan revision**
- **< 200 PA Index**
- **≥ 2 Rp Wood units**
- ***Significant atrial or ventricular arrhythmias***
- **≥ 15mmHg mean PA pressure**
- **> 2m/sec systemic outflow tract obstruction**
- ***≥11mmHg systemic ED pressure***

Fujii Y et al. Ann Thorac Surg 2009; 87: 562

Comment

Severe cardiac malformations are usually found in patients with the asplenia syndrome. They have right isomerism with anomalous drainage of the systemic and pulmonary veins, common atrium and AV valve, single ventricle with malposed great arteries and severe pulmonary stenosis or atresia. Long-term survival is uncommon.These patients have a poor outlook even after surgical treatment with a Fontan algorhithm. Fujii et al recently analyzed the outcomes of a series of 25 consecutive high-risk adult patients undergoing a total cavopulmonary anastomosis. (2) They investigated 13 potential risk factors (Table 2) and concluded that the presence of 6 or more risk factors was significantly associated with higher mortality. The 6 risk factors present in our patient are printed on the table in italics. Since she showed improvement in her symptoms and functional class and taking into account the high risk of surgery, continued medical treatment was advised.

References

1. Ho SY, Seo JW, Brown NA, Cook AC, Fagg NL, Anderson RH. Morphology of the sinus node of human and mouse hearts with isomerism of the atrial

appendages. Brit Heart J 1995; 74: 437-42.

2. Fujii Y, Sano S, Kotani Y, Yoshizumi K, Kasahara S, Ishino K, Akagi T. Midterm to long-term outcome ot total cavopulmonary connection in high-risk adult candidates, Ann Thorac Surg 2009; 87: 562-70.

Regional News From Asia-Pacific

by *Koichiro Niwa*

In Asia-Pacific, there was the 10th Korean GUCH Society Meeting on November 27, 2010. This was combined with the Korean Echo Society meeting. Topics were Oriental VSD (subpulmonary VSD) and nearly 100 attendants joined the meeting. The 13th Congress of Japanese Society of ACHD will be held in Fukuoka, January 8-9. A symposium on Fontan, special lectures on Fontan, Marfan, Heart failure and intervention are scheduled. 87 free oral and poster papers will be presented. There is also an Asian Session in this Congress.
For more information: click here

Regional News From Europe

by *Helmut Baumgartner*

2nd European Advanced Course in GUCH
Focus in Transposition
WORKING GROUP of the EUROPEAN SOCIETY OF CARDIOLOGY on G.U.C.H
4th-5th March 2011

Regional News From U.S.

by *Bill Davidson*

American College of Cardiology Annual Scientific Sessions
New Orleans, LA, USA
April 2-5, 2011

21st Annual ACHD Symposium in Cincinnati
Cincinnati, OH, USA
June 19-22, 2011

Journal Watch

Am J Cardiol. 2010 Dec 15;106(12):1803-7. Epub 2010 Nov 2.

Adolescents' understanding of their congenital heart disease on transfer to adult-focused care.

Van Deyk K, Pelgrims E, Troost E, Goossens E, Budts W, Gewillig M, Moons P.

Division of Congenital and Structural Cardiology, University Hospitals Leuven, Belgium.

Abstract

Adolescents with congenital heart disease (CHD) must take responsibility for their life and care. This requires that they have sufficient knowledge about their heart disease, treatment, and preventive measures. Thus, CHD-related education should be directed to adolescents. Research on adolescents' understanding and knowledge of CHD is limited. It is unknown what adolescents with CHD know about their heart defect, treatment, and preventive measures necessary to avoid complications. We addressed these questions in a descriptive cross-sectional study of 91 adolescents with CHD (53% males; median age 17 years). In the present study, we assessed the subjects' knowledge of CHD using the Leuven Knowledge Questionnaire for Congenital Heart Disease. The results showed that the patients had adequate knowledge (>80% correct answers) about the need for regular follow-up, their required diet, past treatment, and dental practices. They had moderate knowledge (50% to 80% correct answers) about the frequency of follow-up, occupational choices, medication regimen, and sexual activities. However, the patients had poor knowledge (<50% correct answers) of the name of their heart defect; the reasons for follow-up; the effects of competitive sports; the symptoms that reflect deterioration of their heart disease; the definition, characteristics, and risk factors of endocarditis; the possibility of recurrent episodes of endocarditis during their lifetime; the effect of smoking and alcohol on their heart disease; the hereditary nature of their condition; the suitability of intrauterine devices as contraceptives; the appropriateness of oral contraceptives; and the risks of pregnancy. In conclusion, the results of the present study have showed that the level of knowledge of adolescents with CHD has significant gaps.

Eur Respir Rev. 2010 Dec 1:19(118):308-13
J Thorac Cardiovasc Surg. 2010 Dec:140(6 Suppl):S52-7; discussion S86-91

Does the dilated ascending aorta in an adult with congenital heart disease require intervention?

Stulak JM, Dearani JA, Burkhart HM, Sundt TM, Connolly HM, Schaff HV.

Division of Cardiovascular Surgery, Mayo Clinic, Rochester, MN, USA.

Abstract

OBJECTIVES: There is increasing attention to prophylactic replacement of the moderately dilated ascending aorta at aortic valve surgery. Moderate ascending aortic dilatation is common in adult patients with conotruncal anomalies. There are no data outlining actual risk of progressive ascending aortic dilatation or dissection to provide management guidelines.

METHODS: From December 1973 through January 2008, 81 consecutive adults (median age, 34 years; range, 18--59 years) with conotruncal anomalies underwent operation on the aortic root, ascending aorta, or aortic valve. Primary cardiac diagnoses included tetralogy of Fallot with or without pulmonary atresia in 60 patients, truncus arteriosus in 12, double-outlet right ventricle in 6, and other in 3. Indications for operation included aortic regurgitation in 69 patients, supracoronary ascending aneurysm in 16, aortic stenosis in 5, and other in 8. Median ascending aortic size was 45 mm (23--80 mm).

RESULTS: Operations included isolated aortic valve repair/replacement in 63

patients, combined aortic valve replacement and reduction aortoplasty in 9, aortic root replacement in 7, and isolated ascending aortic replacement in 2. Four patients required reoperation during a median follow-up of 3.8 years (maximum 31 years). There were no ascending aortic reoperations after previous reduction aortoplasties or supracoronary ascending aortic grafts, and there were no late aortic dissections.

CONCLUSIONS: Moderate ascending aortic enlargement is common among patients with conotruncal anomalies coming to operation, but aortic dissection is rare, as is subsequent need for aortic reoperation. Despite current enthusiasm for prophylactic operations on the ascending aorta in patients with acquired disease, these data suggest that the moderately dilated aorta in this setting may be observed.

J Am Acad Dermatol. 2010 Nov 26. [Epub ahead of print]

A case-control study of cuntaneous signs in adult patients with Marfan disease: Diagnostic value of striae.

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

Abstract

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

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

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

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

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

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

Heart Vessels. 2010 Nov 26, [Epub ahead of print]

Parameters of arterial function and structure in adult patients after coarctation repair.

Trojnarska O, Mizia-Stec K, Gabriel M, Szczepaniak-Chichel L, Katarzynska-Szymanska A, Grajek S, Tykarski A, Gasior Z, Kramer L.

1st Department of Cardiology, University of Medical Sciences, ul. Dhiga 1/2. 61-848, Poznan, olgatroj@wp.pl.

Abstract

Regardless of a successful operation, patients with coarctation of aorta (CoAo) are exposed to the risk of hypertension and a propensity to vascular and end-organ damage. The aim of this study is to evaluate the influence of residual aorta stenosis as well as the age at the operation on the parameters of arterial function and structure in patients after CoAo repair. Eighty-five patients after CoAo repair (53 males; mean age: 34.6 ± 10.3 years, mean age at the repair: 10.9 ± 8.2 years) were enrolled in the study. The control group consisted of 30 healthy subjects (18 males; mean age: 33.6 ± 8.2 years). Indices of systemic arterial remodeling [flow-mediated dilatation (FMD), nitroglycerine-mediated vasodilatation (NMD), carotid intima-media thickness (IMT), pulse wave velocity (PWV)] were analyzed in all study patients. In normotensive patients after CoAo repair (47/55%), a significantly increased PWV was observed in comparison to the control group (6.8 ± 1.2 vs. 5.4 ± 0.9 m/s; p = 0.003), with no difference in IMT values (0.53 ± 0.1 vs. 0.51 ± 0.1 mm; p = 0.06). Mean FMD (4.8 ± 2.8 vs. 8.5 ± 2.3%; p = 0.00003) and NMD (11.3 ± 4.6 vs. 19.8 ± 7.2%; p = 0.00001) were lower than in the controls. In patients with a residual aorta stenosis (46/54%), defined as an arm-leg pressure gradient ≥20 mmHg, no differences were found within the scope of both systolic and diastolic blood pressure and of all of the examined vascular parameters. No significant correlations were revealed between the vascular parameters and the gradient across descending aorta as well as the age at the operation. Residual stenosis in the descending aorta does not affect the arterial vasodilatation nor stiffness in patients after CoAo repair. An early surgery does not influence the remodeling of the vessels, which supports the thesis that CoAo is a generalized vascular disease and that even an early operation cannot prevent the progressive and vascular changes and end-organ damage.

Expert Rev Cardovasc Ther. 2010 Dec:8(12):1753-66

Interventional and surgical treatment of cardiac arrhythmias in adults with congenital heart disease.

Koyak Z, de Groot JR, Mulder BJ.

Department of Cardiology, Academic Medical Center, Meibergdreef 9, 1105 AZ, Amsterdam, The Netherlands.

Abstract

Arrhythmias are a major cause of morbidity, mortality and hospital admission in adults with congenital heart disease (CHD). The etiology of arrhythmias in this

population is often multifactorial and includes electrical disturbances as part of the underlying defect, surgical intervention or hemodynamic abnormalities. Despite the numerous existing arrhythmia management tools including drug therapy, pacing and ablation, management of arrhythmias in adults with CHD remains difficult and challenging. Owing to improvement in mapping and ablation techniques, ablation and arrhythmia surgery are being performed more frequently in adults with CHD. However, there is little information on the long-term results of these treatment strategies. The purpose of this article is therefore to review the available data on nonpharmacological treatment of cardiac arrhythmias in adult patients with CHD and to give an overview of the available data on the early and late outcomes of these treatment strategies.

Expert Rev Cardiovasc Ther. 2010 Dec:8(12):1741-52.

Managing adults with congenital heart disease in the catheterization laboratory: state of the art.

Macdonald ST, Carminati M, Chessa M.

Department of Pediatric Cardiology and Adults with Congenital Heart Disease, IRCSS. Policlinico San Donato, Via Morandi 30, 20097 San Donato, Milanese (MI), Italy.

Abstract

There has been a rapid growth in interventional cardiology techniques to treat adults with congenital heart disease, mirroring the rise of interventional cardiology as a cardiology subspecialty and the increasing population of adults with congenital abnormalities. Starting with neonatal balloon atrial septostomy for transposition of the great arteries in the 1960s, improving and changing percutaneous interventional techniques have increasingly replaced surgery as a treatment option in several congenital heart diseases. Many pediatric cardiac patients now survive to adulthood following early surgery, perhaps making additional surgery higher risk, and percutaneous procedures offer an alternative approach, although with a different inherent set of risks and benefits in terms of morbidity and mortality. Adult congenital heart disease patients offer distinct challenges such as unusual anatomy and demands such as pregnancy and exercise tolerability not found in conventional pediatric or traditional adult interventional patients. This article reviews current indications for adult congenital heart disease intervention and best practice, detailing the patient spectrum commonly treated, devices used and emerging treatments.

J Cardiovas Nurs. 2010 Nov 23, [Epub ahead of print]

Adult Congenital Heart Care in a Pediatric Setting-A Patient's Perspective.

Tomlin MA, Gosney K.

Martha A. Tomlin, MSN, ACNP-BC Acute Care Nurse Practitioner, Adolescent & Adult Congenital Heart Program, The Heart Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio. Kathy Gosney, MSSW, LISW Social Worker, Adolescent & Adult Congenital Heart Program, The Heart Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio.

Abstract

BACKGROUND: Congenital heart disease (CHD) affects approximately 1% of all live births today. With improvements in diagnostic, medical, surgical, and interventional procedures, 85% or more of all infants with CHD will reach adulthood. The number of adults living with CHD has been estimated to exceed 1 million and now exceeds the number of pediatric CHD patients. Because residual problems following intervention for congenital heart lesions can present during the adult years and complicate the well-being of these patients, ongoing care is warranted. Adult care providers have a limited knowledge of the complexities of CHD. The American College of Cardiology recommends the establishment of adult CHD clinics with both pediatric and adult cardiologists. Although our clinic is located in a pediatric hospital, little attention has been placed on where these clinics should be located-an adult setting or a pediatric setting. We sought to determine the adult CHD patient's perspective on being seen in a pediatric setting.

OBJECTIVE: The objective of the study was to determine the perspective of adults with CHD receiving follow-up care in a pediatric setting.

METHODS: A pilot 11-question anonymous patient satisfaction survey with no personal identifiers and no diagnoses was sent to all patients who had attended the adult congenital heart disease clinic at our pediatric hospital medical center during a 2-year period.

RESULTS: From our respondents, 96% did not have any concerns with being seen in a pediatric setting for adult congenital heart care, and 98% would recommend our clinic to other patients.

CONCLUSION: Care for the adult with CHD involves multiple care providers. The most important finding from the patient's perspective is knowledge of the complexities of congenital heart lesions and possible future complications. There was little impact from being seen in a pediatric hospital setting.

Circulation. 2010 Nov 30;122(22):2254-63. Epub 2010 Nov 22.

Mortality reulting from congenital heart disease among children and adults in the United States, 1999 to 2006.

Gilboa SM, Salemi JL, Nembhard WN, Fixler DE, Correa A.

National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Mail Stop E-86, 1600 Clifton Rd, Atlanta, GA 30333. sgilboa@cdc.gov.

Abstract

Background- Previous reports suggest that mortality resulting from congenital heart disease (CHD) among infants and young children has been decreasing. There is little population-based information on CHD mortality trends and patterns among older children and adults. Methods and Results- We used data from death certificates filed in the United States from 1999 to 2006 to calculate

annual CHD mortality by age at death, race-ethnicity, and sex. To calculate mortality rates for individuals ≥1 year of age, population counts from the US Census were used in the denominator; for infant mortality, live birth counts were used. From 1999 to 2006, there were 41 494 CHD-related deaths and 27 960 deaths resulting from CHD (age-standardized mortality rates, 1.78 and 1.20 per 100 000, respectively). During this period, mortality resulting from CHD declined 24.1% overall. Mortality resulting from CHD significantly declined among all race-ethnicities studied. However, disparities persisted; overall and among infants, mortality resulting from CHD was consistently higher among non-Hispanic blacks compared with non-Hispanic whites. Infant mortality accounted for 48.1% of all mortality resulting from CHD; among those who survived the first year of life, 76.1% of deaths occurred during adulthood (≥18 years of age). Conclusions- CHD mortality continued to decline among both children and adults; however, differences between race-ethnicities persisted. A large proportion of CHD-related mortality occurred during infancy, although significant CHD mortality occurred during adulthood, indicating the need for adult CHD specialty management.

Circulation. 2010 Nov 30;122(22):2264-72. Epub 2010 Nov 22.

Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium.

Moons P, Bovign L, Budts W, Belmans A, Gewillig M.

Center for Heath Services and Nursing Research, Katholieke Universiteit Leuven, Kapucijnenvoer 35, Post Box 7001, B-3000 Leuven, Belgium. Philip.Moons@med.kuleuven.be.

Abstract

Background- Over the past decades, the life expectancy of individuals with congenital heart disease (CHD) has increased significantly. However, precise estimates for survival to adulthood are scarce for patients with CHD. We investigated the proportion of CHD patients born between 1990 and 1992 who survived into adulthood. We also compared their survival with that of CHD patients born in earlier eras and evaluated survival as a function of the type of heart defect. Methods and Results- We reviewed the CHD program administrative and clinical database at the University Hospitals Leuven (Leuven, Belgium) and analyzed the records of 7497 CHD patients born from 1970 to 1992. Survival to 18 years of age in patients born between 1990 and 1992 was 88.6% (95% confidence interval [CI], 86.3% to 90.5%), which was significantly greater than that of patients born in previous decades (P<0.0001). For patients born between 1990 and 1992, survival into adulthood for those with mild heart defects was 98.0% (95% CI, 95.8% to 99.1%), whereas survival for those with moderate- and severe-complexity heart defects was 90.0% (95% CI, 86.8% to 92.5%) and 56.4% (95% CI, 47.4% to 64.5%), respectively. Analysis per heart defect confirmed these findings, demonstrating that patients with univentricular heart (49.1% [95% CI, 30.8% to 65.1%]) and hypoplastic left heart syndrome (7.5% [95% CI, 0.6% to 26.6%]) had the poorest survival rate. Conclusion- This study demonstrates that almost 90% of children with CHD have the prospect of surviving into adulthood.

Int J Cardiol. 2010 Nov 19. [Epub ahead of print]

End-of-life in adults with congenital heart disease: A call for early communication.

Tobler D, Greutmann M, Colman JM, Greutmann-Yantiri M, Librach LS, Kovacs AH.

Toronto Congenital Cardiac Centre for Adults, Peter Munk Cardiac Center, University Health Network, University of Toronto, Toronto, Canada.

Abstract

BACKGROUND: We investigated preferences of adults with congenital heart disease (CHD) and their health care providers regarding end-of-life (EOL) communication.

METHODS: Adult CHD outpatients and health care providers completed surveys about preferences for and experiences with EOL communication. Responses were compared between patients and providers.

RESULTS: Two hundred patients (18-79years) and 48 CHD health care providers (primarily cardiologists) completed surveys. Only 2 patients (1%) indicated that they had discussed EOL planning with their medical team. In contrast, 50% of providers reported that they typically discuss issues including life expectancy, advance planning, and resuscitation preferences with their outpatients. Seventy-eight percent (156/199) of patients wanted their medical team to raise EOL issues; this preference was independent of disease complexity and socio-demographic factors. In contrast, providers reported that their EOL discussions increase in accordance with disease complexity ($p<0.001$). Early initiation of EOL discussions, before diagnosis with life-threatening complications, was favored by 62% of patients but only 38% of providers ($p<0.001$).

CONCLUSION: Health professionals caring for adults with CHD should explore preferences of their patients for EOL discussions earlier in the disease course, and not only with patients facing life-threatening complications and/or with complex conditions. When EOL discussions do occur, health care providers should attempt to ensure that patients better understand these conversations. Increased attention to EOL issues is proposed in order to improve the care of patients with CHD across the lifespan.

Int J Cardiol. 2010 Nov 15. [Epub ahead of print]

Bosentan-sildenafil association in patients with congenital heart disease-related pulmonary arterial hypertension and Eisenmenger physiology.

D'Alto M, Romeo E, Argiento P, Sarubbi B, Santoro G, Grimaldi N, Correra A, Scognamiglio G, Russo MG, Calabro R.

Abstract

OBJECTIVES: The aim of the present study was to evaluate the safety, tolerability, clinical and haemodynamic impact of add-on sildenafil in patients

with congenital heart disease (CHD)-related pulmonary arterial hypertension (PAH) and Eisenmenger physiology after failure of oral bosentan therapy.

METHODS: Thirty-two patients with CHD-related PAH (14 male, mean age 37.1±13.7years) treated with oral bosentan underwent right heart catheterization (RHC) for clinical worsening. After RHC, all patients received oral sildenafil 20mg thrice daily in addition to bosentan. Clinical status, resting transcutaneous oxygen saturation (SpO(2)), 6-minute walk test (6MWT), serology and RHC were assessed at baseline (before add-on sildenafil) and after 6months of combination therapy.

RESULTS: Twelve patients had ventricular septal defect, 8 atrio-ventricular canal, 6 single ventricle, and 6 atrial septal defect. Twenty-eight/32 had Eisenmenger physiology and 4 (all with atrial septal defect) did not. All patients well tolerated combination therapy. After 6months of therapy, an improvement in clinical status (WHO functional class 2.1±0.4 vs 2.9±0.3; P=0.042), 6-minute walk distance (360±51 vs 293±68m; P=0.005), SpO(2) at the end of the 6MWT (72±10 vs 63±15%; P=0.047), Borg score (2.9±1.5 vs 4.4±2.3; P=0.036), serology (pro-brain natriuretic peptide 303±366 vs 760±943pg/ml; P=0.008) and haemodynamics (pulmonary blood flow 3.4±1.0 vs 3.1±1.2l/min/m(2), P=0.002; pulmonary vascular resistances index 19±9 vs 24±16WU/m(2), P=0.003) was observed.

CONCLUSIONS: Addition of sildenafil in adult patients with CHD-related PAH and Eisenmenger syndrome after oral bosentan therapy failure is safe and well tolerated at 6-month follow-up, resulting in a significant improvement in clinical status, effort SpO(2), exercise tolerance and haemodynamics.

Curr Opin Cardiol. 2010 Nov 11. [Epub ahead of print]

Management of arrhythmias in patients with tetralogy of Fallot.

Le Gloan L, Khairy P.

Adult Congenital Heart Center and Electrophysiology Service, Montreal Heart Institute, University of Montreal, Montreal, Quebec, Canada.

Abstract

PURPOSE OF REVIEW: Patients with tetralogy of Fallot are subject to arrhythmic sequelae that substantially impact morbidity and mortality. This review focuses on recent advances in our understanding of the prevalence and types of arrhythmias encountered. Diagnostic and prognostic tools are considered and therapeutic options discussed.

RECENT FINDINGS: Multicenter studies have characterized the arrhythmia burden, assessed the impact of implantable cardioverter-defibrillators, and generated a risk score for primary prevention. Left ventricular hemodynamics are increasingly recognized as important contributors to risk for sudden death. Arrhythmia circuits have been characterized, and the impact of pulmonary valve replacement on sudden death has been further questioned. Recent studies cast doubt on the value of right ventricular pacing alone for cardiac resynchronization and provide a rationale for biventricular pacing.

SUMMARY: Supraventricular arrhythmias exceed ventricular arrhythmias in prevalence, as atrial fibrillation increases with the aging population. Sudden

death is the leading cause of late mortality, although therapeutic advances may alter this profile. Combinations of factors should be considered in risk stratification schemes to select appropriate implantable cardioverter-defibrillator candidates. The role of concomitant intraoperative ablation during pulmonary valve replacement surgery remains to be defined. Cardiac resynchronization therapy, particularly biventricular pacing, offers promise, but requires careful study before widespread implementation.

J Heart Lung Transplant. 2010 Oct 28. [Epub ahead of print]

Would access to device therapies improve transplant outcomes for adults with congenital heart disease? Analysis of the United Network for Organ Sharing (UNOS).

Everitt MD, Donaldson AE, Stehlik J, Kaza AK, Budge D, Alharethi R, Bullock EA, Kfoury AG, Yetman AT.

University of Utah, Salt Lake City, Utah; Utah Transplant Affiliated Hospitals (U.T.A.H.) Cardiac Transplant Program, Salt Lake City, Utah.

Abstract

BACKGROUND: Patients with congenital heart disease (CHD) now survive into adulthood and often present with end-stage heart failure (HF). HF management and approach to orthotopic heart transplant (OHT) may differ from adults without CHD. We sought to compare OHT waitlist characteristics and outcomes for these 2 groups.

METHODS: The Organ Procurement and Transplantation Network (OPTN)/United Network for Organ Sharing (UNOS) database was used to identify adults (≥18 years) listed for OHT from 2005 to 2009. The cohort was divided into those with or without CHD.

RESULTS: Of 9,722 adults included, 314 (3%) had CHD. Adults with CHD were younger (35 ± 13 vs 52 ± 12 years, $p < 0.01$) and more often had undergone prior cardiac surgery (85% vs. 34%, $p < 0.01$). Patients with CHD were less likely to have a defibrillator (44% vs 75%, $p < 0.01$) or ventricular assist device (5% vs 14%, $p < 0.01$) and were more likely to be listed at the lowest urgency status than patients without CHD (64% vs 44%, $p < 0.01$). Fewer CHD patients achieved OHT (53% vs 65%, $p < 0.001$). Although overall waitlist mortality did not differ between groups (10% vs 8%, $p = 0.15$), patients with CHD were more likely to experience cardiovascular death (60% vs 40%, $p = 0.03$), including sudden in 44% and due to HF in 16%.

CONCLUSIONS: Despite lower urgency status, patients with CHD have greater cardiovascular mortality awaiting OHT than those without. Increased defibrillator use could improve survival to OHT, because sudden death is common. VAD support may benefit select patients, but experience in CHD is limited. Referral to specialized adult congenital heart centers can enhance utilization of device therapies and potentially improve waitlist outcomes.

Ann Vasc Surg. 2010 Nov;24(8):1068-74

Open surgical repair and endovascular treatment in adult coarctation of the aorta.

San Norberto Garcia EM, Gonzalez-Fajardo JA, Gutierrez V, Fernandez B, San

Roman A, Vaquero C.

Division of Angiology and Vascular Surgery, Valladolid University Hospital, Valladolid, Spain. esannorberto@hotmail.com

Abstract

BACKGROUND: The aim of this study was to compare the results of endovascular therapy (covered stenting) with surgical technique to repair aortic coarctation in adults.

METHODS: A prospective study of 11 patients who were treated during the past 10 years was carried out. Of these, five patients underwent endoprosthesis (group A) and six an open surgical repair (group B). Follow-up comprised monitoring of the blood pressure, echocardiography, and computed tomography and magnetic resonance angiographic studies.

RESULTS: The mean age of the patients was 46 years (range: 17-67 years) and the mean follow-up was 52.6 months (range: 1-117 months; 32.3 for group A vs. 69.7 for group B; p = 0.01). Two cases in group A were recoarctations after child angioplasty. The rate of postoperative complications was 27.7% (one hemothorax for group A vs. one pneumothorax and one hemothorax for group B); however, mortality did not occur. The success rate of the endovascular technique was 80%. The stay in the intensive care unit was 2.3 days with significant differences (one group A vs. three group B; p = 0.01), whereas length of hospital stay was 11 days (7.8 group A vs. 11.83 group B; p = 0.17). The pressure gradient across the stenosis decreased by 21.9 ± 3.7 mm Hg (24.5 ± 4.3 group A vs. 33 ± 3.2 group B). Six patients (54.5%) showed persistent hypertension (80% group A vs. 33% group B), with a mean residual pressure gradient of 23.4 ± 4.3 mm Hg (22.5 ± 5.4 group A vs. 22 ± 2.1 group B; p = 0.58).

CONCLUSIONS: Short- and medium-term results of the endovascular therapy are similar, with shorter stay in the intensive care unit and higher necessity of antihypertensive treatment. Echocardiography and Doppler aortic coarctation gradients slightly higher than 20 mm Hg are usual during follow-up.

J Am Coll Cardiol. 2010 Nov 2;56(19):1589-96.

Outcome of intra-atrial re-entrant tachychardia catheter ablation in adults with congenital heart disease: negative impact of age and complex atrial surgery.

Yap SC, Harris L, Silversides CK, Downar E, Chauhan VS.

Toronto Congenital Cardiac Center for Adults, Toronto General Hospital, University Health Network, Toronto, Ontario, Canada.

Abstract

OBJECTIVES: The aim of this study was to determine the acute and long-term outcome of radiofrequency catheter ablation (RFCA) for intra-atrial re-entrant tachycardia (IART) in adults with congenital heart disease (CHD), and predictors of these outcomes.

BACKGROUND: Atrial myopathy can be progressive in CHD and contributes to the substrate for IART. Although the outcome of RFCA for IART has been well described in children and adolescents with CHD, it is unclear whether these results are similar in the adult population.

METHODS: Clinical records of adults with CHD undergoing attempted RFCA of IART were analyzed retrospectively. Multivariate analyses identified clinical and procedural factors that predicted acute and long-term outcomes.

RESULTS: A total of 193 procedures was performed in 130 patients (mean age 40 ± 13 years); 82 of 118 (69%) initially attempted RFCA were successful, defined as termination of all IART circuits. The use of electroanatomic mapping was associated with a successful RFCA, whereas Fontan palliation and Mustard repair were associated with an unsuccessful RFCA. Median clinical follow-up of 77 patients (≥2 months of follow-up) after a successful RFCA was 3.7 years (range 0.2 to 10.2 years). IART recurrence was noted in 48%, cardioversion/reablation in 42%, and death in 4%. Older age and Fontan palliation were independent predictors of IART recurrence.

CONCLUSIONS: In adults with CHD, acute and long-term outcomes of RFCA for IART are similar to those reported for younger cohorts. Complex atrial surgery limits the success of RFCA, and older age is associated with a higher risk of IART recurrence.

Am J Cardiol. 2010 Nov 1:106(9):1317-21

Contraception in women with congenital heart disease.

Vigl M, Kaemmerer M, Seifert-Klauss V, Niggemeyer E, Nagdyman N, Trigas V, Bauer U, Schneider KT, Berger F, Hess J, Kaemmerer H.

Competence Network for Congenital Heart Defects, Berlin Germany.

Abstract

The present study reports on contraceptive use, methods used, and counseling received on contraceptive issues for women with congenital heart disease and provides a brief review of current knowledge of the risks in relation to the different cardiac situations encountered with these specific patients. A total of 536 consecutive adult women with congenital heart disease (median age 29 years) were recruited from 2 tertiary care centers. They underwent a clinical assessment and completed a questionnaire regarding their contraceptive use. Oral contraceptives, condoms, and intrauterine devices were the most commonly used methods. Pregnancy occurred in almost every tenth woman despite the use of contraception. We identified a substantial number of women (20%) who were presently using contraceptive methods that were contraindicated for their specific cardiac condition. Additionally, a high proportion of patients (28%), in the group with high pregnancy-associated risks, were not using contraception despite having a sexual relationship. In our study, 43% of the women had not been counseled about contraception, and 48% had not been informed of the pregnancy-related risks by their treating physician. In conclusion, timely and competent counseling about contraception is important for women with congenital heart disease. Collaboration between cardiologists and gynecologists should be strengthened. Failure to give adequate family planning advice to this patient group could have hazardous consequences,

causing an unnecessary risk to mother and child.

J Card Surg. 2010 Nov;25(6):629-32. doi:10.1111/j.1540-8191.2010.01110.x. Epub 2010 Oct 4.

Coronary artery bypass grafting in adults with congenital heart disease.

Cui YC, Hao XH, Huang FJ, Li JH, Lai YQ, Zhou QW.

Department of Cardiac Surgery, Beijing Anzhen Hospital Capital Medical University, Beiging, China.

Abstract

BACKGROUND: Adults with congenital heart disease (CHD) and coronary artery disease (CAD) have unique clinical manifestation due to the coexistence of intracardiac anomalies and CAD. Case reports are rare in surgical management of CHD combined with CAD. Our goal is to study the outcome of surgical intervention of CHD and CAD concomitantly.

METHODS: From February 2002 to August 2009, 29 adult patients underwent coronary artery bypass grafting (CABG) and surgical correction of CHD concomitantly. Congenital cardiac anomalies include atrial septal defect (ASD) in 21 cases, ventricular septal defect in four cases, atrioventricular septal defect in three cases, and cor triatriatum in one case. Coronary angiography demonstrated: one-vessel disease in 10 cases, two-vessel disease in 11 cases, and three-vessel disease in eight cases. Coronary revascularization and intracardiac anomalies were corrected with cardiopulmonary bypass in 23 cases. There were six patients who had off-pump coronary artery pass grafting (OPCAB) and intraoperative device closure of ASD.

RESULTS: One patient died of pulmonary infection and multiorgan failure. Follow-up time was from 2 to 89 months (mean, 42 ± 25 months). One patient with recurrent angina did not need intervention of the revascularization. Six patients who acquired OPCAB and intraoperative device closure of ASD had no complications after surgery.

CONCLUSIONS: Surgery for adult patients who had CHD with CAD was a safe and effective management. OPCAB with intraoperative device closure of ASD was a reasonable approach for some selective patients.

Cardiol Young. 2010 Oct 27;1-8, [Epub ahead of print]

Ablation of atrial tachyarrhythmias late after surgical repair of tetralogy of Fallot.

Raine D, O'Sullivan J, Chaudhari M, Hamilton L, Hasan A, Bourke JP.

Department of Adult, Freeman Hospital, Newcastle upon Tyne Hospitals NHS Foundation Trust, United Kingdom.

Abstract

BACKGROUND: Patients with repaired tetralogy of Fallot may develop

symptomatic and haemodynamic deterioration for many reasons such as arrhythmia, pulmonary regurgitation, and impairment in ventricular function. We describe a consecutive group of patients whose main clinical problem was atrial tachyarrhythmias. Aims to describe the clinical outcome of atrial tachyarrhythmias occurring late after surgical repair of tetralogy of Fallot; to define the circuits/foci responsible for these atrial tachyarrhythmias; to evaluate the outcome of computer-assisted mapping and catheter ablation in this patient group.Methods and results. Consecutive patients with surgically repaired tetralogy of Fallot and atrial tachyarrhythmias, who underwent catheter ablation between January, 2001 and June, 2007, were identified retrospectively from case records. Computer-assisted mapping was performed in all using either EnSite® (St Jude Medical Inc.) arrhythmia mapping and intra-cardiac catheter guidance system or CARTO™ (Biosense Webster Inc.) electroanatomical mapping systems. Ten patients (four males) with a median age of 39 plus or minus 8 years were studied. The total number of atrial tachyarrhythmias identified was 22 (six macro-reentrant, 16 micro-reentrant/focal). In nine patients, catheter ablation led to improvement in arrhythmia episodes and/or symptoms during follow-up of 41 plus or minus 20 months. Following ablation(s), five patients required pacing for pre-existing conduction disease and five needed further surgery for haemodynamic indications. All patients remained on anti-arrhythmic drugs.

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

Ann Thorac Surg. 2010 Nov;90(5):1563-9.

Valvular operations in patients with congenital heart disease: increasing rates from 1988 to 2005.

Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Tchervenkov C, Martucci G, Marelli AJ.

McGill Adult Unit for Congenital Heart Disease Excellence (MAUDE Unit), McGill University Health Centre, Montreal, Quebec, Canada.

Abstract

BACKGROUND: The congenital heart disease population is aging. We hypothesized that changes in rates of congenital, valvular, and noncongenital surgical operations in congenital heart patients varied with age and disease severity over the last two decades.

METHODS: We performed time trend analysis using a Quebec congenital heart disease database constructed from administrative data. We included congenital heart patients of all ages having cardiac surgical operations. Heart lesions were classified as "severe" and "other." Cardiac surgical operations were grouped as congenital, valvular (including aortic), and noncongenital (arrhythmia surgery, coronary artery bypass grafting, and cardiac transplants). An adapted Aristotle score was developed to classify procedures based on surgical risk. Yearly surgical rates were measured as surgical operations per 1,000 person-years and analyzed over time using Poisson regression models stratified by age,

lesion severity, and cardiac surgery category.

RESULTS: From 1988 to 2005 we followed 71,979 patients for 1,009,430 person-years. We identified 17,444 cardiac surgical operations. There was a 31% increase in volumes and a 5% increase in surgical rates over time. In children, congenital surgical operations remained constant, accounting for 80% of all surgical operations. In adults, valvular operations were the most common type of surgical operations, increasing from 42% to 63% of all procedures over time. Rates of valvular operations increased significantly in all adult subgroups and in children with severe lesions.

CONCLUSIONS: The need for valvular interventions has increased in the last two decades in congenital heart disease patients. These findings should be taken into account when allocating resources that will optimize outcomes for this growing population.

AJR Am J Roentgenol. 2010 Nov;195(5):W331-6

Partially unroofed coronary sinus: MDCTand MRI findings.

Kim H, Choe YH, Park SW, Jun TG, Kang IS, Yang JH, Eo H, Lee HJ.

Department of Radiology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul Korea.

Abstract

OBJECTIVE: The purpose of this study was to analyze the MDCT and MRI findings in patients with unroofed coronary sinus syndrome.

MATERIALS AND METHODS: This retrospective study included 11 patients with unroofed coronary sinus syndrome (10 adults, one child) without persistent left superior vena cava and one adult with communication of the left atrium and coronary sinus via an anomalous vein (unroofed coronary sinus variant). Four patients underwent contrast-enhanced ECG-gated MDCT; six, MRI; and two patients, both CT and MRI. We also measured the coronary sinus on the CT scans of 28 adults with normal cardiac anatomy and 10 adults with persistent left superior vena cava and compared the measurements with those in the patients with unroofed coronary sinus syndrome. Seven patients underwent surgical treatment of unroofed coronary sinus syndrome.

RESULTS: At echocardiography, unroofed coronary sinus syndrome was not clearly discriminated from atrial septal defect in two patients and was not suspected in three patients. CT and MRI showed that 11 patients had a defect in which the coronary sinus communicated with the left atrium and that the other patient had atresia of the coronary sinus orifice with an anomalous vein connecting the coronary sinus and left atrium. In patients with unroofed coronary sinus syndrome, the mean standardized diameter of the coronary sinus according to the patient's body surface area was 15 ± 4 mm/m², similar to that of the control group with persistent left superior vena cava (15 ± 6 mm/m²; $p = 0.97$) and significantly greater than that of the group with normal cardiac anatomy (7 ± 2 mm/m²; $p < 0.0001$).

CONCLUSION: CT and MRI facilitate definite diagnosis of unroofed coronary sinus syndrome.

Heart. 2010 Oct 21. [Epub ahead of print]

Pre-stenting with a bare metal stent before percutaneous pulmonary valve implantation acute and 1-year outcomes.

Nordmeyer J, Lurz P, Khambadkone S, Schievano S, Jones A, McElhinney DB, Taylor AM, Bonhoeffer P.

UCL Institute of Child Health, London, UK.

Abstract

Objectives: To determine the feasibility and safety of pre-stenting with a bare metal stent (BMS) before percutaneous pulmonary valve implantation (PPVI), and to analyse whether this approach improves haemodynamic outcomes and impacts on the incidence of PPVI stent fractures. Design Retrospective analysis of prospectively collected data. Setting Tertiary paediatric and adult congenital heart cardiac centre. Patients and interventions 108 consecutive patients with congenital heart disease underwent PPVI between September 2005 and June 2008 (54 with PPVI alone, 54 with BMS pre-stenting before PPVI). Results There were no significant differences in procedural complication rates. Acutely, there was no difference in haemodynamic outcomes. Serial echocardiography revealed that in the subgroups of 'moderate' (26-40 mmHg) and 'severe' (>40 mmHg) right ventricular outflow tract (RVOT) obstruction, patients with pre-stenting showed a tendency towards lower peak RVOT velocities compared to patients after PPVI alone (p=0.01 and p=0.045, respectively). The incidence of PPVI stent fractures was not statistically different between treatment groups at 1 year (PPVI 31% vs BMS+PPVI 18%; p=0.16). However, pre-stenting with BMS was associated with a lower risk of developing PPVI stent fractures (HR 0.35, 95% CI 0.14 to 0.87, p=0.024). The probability of freedom from serious adverse follow-up events (death, device explantation, repeat PPVI) was not statistically different at 1 year (PPVI 92% vs BMS+PPVI 94%; p=0.44). Conclusions Pre-stenting with BMS before PPVI is a feasible and safe modification of the established implantation protocol. Pre-stenting is associated with a reduced risk of developing PPVI stent fractures.

Int J Clin Pract Suppl. 2010 Nov;64(168):23-32. doi: 10.111/j.1742-1241.2010.02525.x.

How has epoprostenal changed the outcome for patients with pulmonary arterial hypertension?

Barst R.

Department of Pediatric Cardiology, Columbia University Medical Center. Scarsdale, NY 10583, USA. robyn.barst@gmail.com

Abstract

AIMS: Pulmonary arterial hypertension (PAH), characterized by increased pulmonary vascular resistance and pulmonary artery pressure, is a significant cause of morbidity and mortality in children and adults. Prior to 1995, there were no approved therapies for PAH.

MATERIALS AND METHODS: Review of the clinical drug development of epoprostenol (synthetic prostacyclin) for the treatment of PAH.

RESULTS: Based on the results of a phase 2 and one phase 3 trial carried out between 1987 and 1992 in adult patients with PAH, epoprostenol was approved for the treatment of severe idiopathic PAH in 1995. Continuous intravenous infusion 24/7 of epoprotenol improved exercise capacity, hemodynamic parameters, functional capacity and survival. Epoprostenol was subsequently shown to be safe and efficacious in PAH associated with the scleroderma spectrum of disease and has now been utilized in PAH associated with congenital heart disease, HIV, portal hypertension, drugs and toxin and connective tissue diseases. Epoprostenol has also been used in children of all ages with similar safety and efficacy as shown in adult patients.

DISCUSSION: Due to the mode of delivery of epoprostenol, i.e. continuous intravenous infusion 24/7 via an indwelling central venous line, there are significant side effects than can occur with its use, e.g. bacteremia, sepsis, thromboembolic events, that can be fatal. Furthermore, there is significant variability in the optimal dose in both children and adult patients. It remains unclear why there is such dose variability between patients to achieve optimal efficacy. Furthermore, its mechanism(s) of action remain unclear.

J Am Coll Cardiol. 2010 Oct 26;56(18):1486-92

Follow-up after pulmonary valve replacement in adults with tetralogy of Fallot: association between QRS duration and outcome.

Scherptong RW, Hazekamp MG, Mulder BJ, Wijers O, Swenne CA, van der Wall EE, Schalij MJ, Vliegen HW.

Department of Cardiology, Leiden University Medical Center, Leiden the Netherlands.

Abstract

OBJECTIVES: The aim of this study was to analyze whether QRS duration, before and after pulmonary valve replacement (PVR), is related to long-term outcome in patients with tetralogy of Fallot (TOF).

BACKGROUND: Key factors that determine outcome after PVR in adult TOF patients are largely unknown. Recognition of such factors assists the identification of patients at increased risk of adverse events.

METHODS: Adults who previously underwent total correction for TOF (n=90; age 31.4±10.3 years) and required PVR for pulmonary regurgitation were included. The QRS duration was measured pre-operatively and 6 months after PVR. The post-operative changes in QRS duration were calculated. Adverse events (death, re-PVR, ventricular tachycardia, and symptomatic heart failure) were noted during follow-up.

RESULTS: During 5.5±3.5 years of follow-up, 13 adverse events occurred. The 5-year event-free survival rate was 76% for patients with a pre-operative QRS duration>180 ms and 90% in patients with a QRS duration≤180 ms (p=0.037). For patients with a post-operative QRS duration>180 ms, 5-year event-free

survival was 71%, whereas it was 91% for patients with a post-operative QRS duration≤180 ms (p=0.004). After multivariate correction, a post-operative QRS duration>180 ms (hazard ratio: 3.685, 95% confidence interval: 1.104 to 12.304, p<0.05) and the absence of a reduction in QRS duration post-PVR (hazard ratio: 6.767, 95% confidence interval: 1.704 to 26.878, p<0.01), was significantly associated with adverse outcome.

CONCLUSIONS: Severe QRS prolongation, before or after PVR, and the absence of a reduction in QRS duration after PVR, are major determinants of adverse outcome during long-term follow-up of patients with TOF.

Circ Cardiovasc. Imaging. 2010 Nov 1;3(6):727-34. Epub 2010 Sep 20.

Quantification of diffuse myocardial fibrosis and its association with myocardial dysfunction in congenital heart disease.

Broberg CS, Chugh SS, Conklin C, Sahn DJ, Jerosch-Herold M.

Adult Congenital Heart Disease Program. Oregon Health and Science University, Portland, Ore. USA. brobergc@ohsu.edu

Abstract

BACKGROUND: the etiology of ventricular dysfunction in adult congenital heart disease (ACHD) is not well understood. Diffuse fibrosis is a likely common final pathway and is quantifiable using MRI.

METHODS AND RESULTS: patients with ACHD (n=50) were studied with cardiac MRI to quantify systemic ventricular volume and function and diffuse fibrosis. The fibrosis index for a single midventricular plane of the systemic ventricle was quantified by measuring T1 values for blood pool and myocardium before and after administration of gadolinium (0.15 mmol/kg) and then adjusted for hematocrit. Results were compared to healthy volunteers (normal controls, n=14) and patients with acquired heart failure (positive controls, n=4). Patients studied (age, 37±12 years; female sex, 40%) included 11 with a systemic right ventricle (RV), 17 with tetralogy of Fallot, 10 with cyanosis, and 12 with other lesions. The fibrosis index was significantly elevated in patients with ACHD compared to normal controls (31.9±4.9% versus 24.8±2.0%; P=0.001). Values were highest in patients with a systemic RV (35.0±5.8%; P<0.001) and those who were cyanotic (33.7±5.6%; P<0.001). The fibrosis index correlated with end-diastolic volume index (r=0.60; P<0.001) and ventricular ejection fraction (r=-0.53; P<0.001) but not with age or oxygen saturation in patients who were cyanotic. Late gadolinium enhancement did not account for the differences seen.

CONCLUSIONS: patients with ACHD have evidence of diffuse, extracellular matrix remodeling similar to patients with acquired heart failure. The fibrosis index may facilitate studies on the mechanisms and treatment of myocardial fibrosis and heart failure in these patients.

Cardiovasc Ther. 2010 Dec;28(6):350-5. doi: 10.1111/j.1755-5922.2010.00213.x.

Impact of sildenafil therapy on pulmonary arterial hypertension in adults with congenital heart disease.

Lu XL, Xiong CM, Shan GL, Zhu XY, Wu BX, Wu GH, Liu ZH, Ni XH, Cheng XS, Gu Q, Zhao ZH, Zhang DZ, Li WM, Zhang C, Tian HY, Guo YJ, Guo T, Liu HM, Zhang Wj, Gu H, Huang SA, Chen JY, Wu WF,Huang K, Li JJ, He JG.

Center for Diagnosis and Management of Pulmonary Vascular Diseases, Department of Cardiology, Cardiovascular Institute and Fu Wai Hospital, Chinese Academy of Medical Sciences & Peking Union Medical College, Beijing, China.

Abstract

BACKGROUND: It has been demonstrated that sildenafil is effective in patients with pulmonary arterial hypertension (PAH). However, the impact of sildenafil on PAH in adults with congenital heart disease (CHD) has been less investigated.

OBJECTIVE: In this prospective, open-label, uncontrolled and multicenter study, 60 patients with PAH related to CHD received oral sildenafil (75 mg/day) for 12 weeks. The enrolled patients underwent six-minute walk test (SMWT) and cardiac catheterization at the beginning and the end of the 12 weeks. The primary end point was the changes in exercise capacity assessed by SMWT; the secondary end point included assessment of functional class, evaluation of cardiopulmonary hemodynamics, and clinical worsening (defined as death, transplantation, and rehospitalization for PAH). Drug safety and tolerability were also examined.

RESULTS: Oral sidenafil significantly increased SMWT distances (422.94 ± 76.95 m vs. 371.99 ± 78.73 m, $P < 0.0001$). There was also remarkable improvement in Borg dyspnea score (2.1 ± 1.32 vs. 2.57 ± 1.42, $P = 0.0307$). Moreover, significant improvements in World Healthy Organization (WHO) functional class and cardiopulmonary hemodynamics were also discovered (mean pulmonary artery pressure, $P = 0.0002$; cardiac index, $P < 0.0001$; pulmonary vascular resistance, $P < 0.0001$). Side effects in this study were mild and consistent with reported studies. None of the enrolled patients experienced significant clinical worsening.

CONCLUSIONS: This study confirmed and extended previous studies. It suggested that oral sildenafil was safe and effective for the treatment of adult patients with CHD-related PAH.

Metabolism. 2010 Nov;59(11):1642-8. Epub 2010 Apr 27.

Serum glucose and lipid levels in adult congenital heart disease patients.

Martinez-Quintana E, Rodriquez-Gonzalez F, Nieto-Lago V, Novoa FJ, Lopez-Rios L, Riano-Ruiz M.

Cardiology Service, Complejo Hospitalario Universitario Insular-Materno Infantil, Las Palmas de Gran Canaria, 35016 Spain. efrenmartinezquintana@yahoo.es

Abstract

Atherosclerosis has been correlated with known cardiovascular risk factors such as serum glucose or lipid levels. Because congenital heart disease patients tend to survive until adulthood, atherosclerosis has also become a matter of concern in these patients. One hundred fifty-eight congenital heart disease patients and 152 patients selected at random from the population were studied and compared to determine serum glucose, total cholesterol, low-density lipoprotein (LDL) cholesterol, high-density lipoprotein cholesterol, and triglycerides levels. Both groups had similar socioeconomic status levels and the same environmental influences. Significant differences were seen between congenital heart disease patients and the control group, after sex, age, and body mass index adjustment, in fasting plasma glucose (97.7 [94.2-101.2] vs 86.9 [83.2-90.7], P < .001), total cholesterol (171.5 [165.7-177.3] vs 199.8 [90.7-206.0], P < .001), LDL cholesterol (103.9 [98.8-108.8] vs 123.8 [118.5-129.1], P < .001), and high-density lipoprotein cholesterol (48.1 [46.2-50.0] vs 54.2 [52.1-56.2], P < .001) levels. Nonsignificant differences were seen in triglycerides concentrations. Those patients with ventricular septal defect, coarctation of the aorta, and cyanosis had the lowest total cholesterol and LDL cholesterol concentrations. Congenital heart disease patients have lower plasma cholesterol concentrations and higher serum glucose levels than noncongenital ones.