CONGENITAL CARDIOLOGY TODAY

Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

Volume 7 / Issue 11
November 2009
North American Edition

IN THIS ISSUE

CONGENITAL CARDIOLOGY TODAY
Editorial and Subscription Offices
16 Cove Rd, Ste. 200
Westerly, RI 02891 USA
www.CongenitalCardiologyToday.com

ISSN: 1544-7787 (print); 1544-0499 (online). Published monthly.

UPCOMING MEDICAL MEETINGS

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Dec. 13-15, 2009; Charleston, SC USA
musckids.com/heart/conferences/

Evolving Concepts in the Management of Complex Congenital Heart Disease II:
Jan. 14-16, 2010; San Diego, CA USA
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Cardiology 2010
Feb. 10-14, 2010; Lake Buena Vista, FL USA
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Bilateral Versus Unilateral Bidirectional Glenn Shunts: Saudi Experience

By Sameh Ibrahim Sersar, MD and Ahmed A. Jamjoom MD

Key Words: Glenn Cavopulmonary anastomosis.

Abstract

Introduction: Bidirectional Cavopulmonary Anastomoses (BDG a.ka. Glenn shunts) as a first or second palliative stage procedure before embarking on a total cavopulmonary connection (“Fontan type” procedure) has been used to normalise volume loading of the single ventricle at an early age. The presence of bilateral Superior Vena Cava (SVC) may represent a technical challenge in the performance of the bilateral Cavopulmonary Anastomoses (b-CPA) connections.[1-2]

Methods: The available chart and files data were reviewed and analyzed in 160 cases of unilateral bidirectional Glenn (u-CPA) and 44 children undergoing bilateral Cavopulmonary Anastomoses. Univariate analysis of the risk factors of mortality, morbidity and ICU and hospital stay was done.

Results: One hundred sixty patients had U-CPA (Group 1) and 44 Patients had b-CPA (Group 2) in King Faisal Specialist Hospital and Research Centre (KFSH&RC), Jeddah, Kingdom of Saudi Arabia, in more than 8 years. They were 127 males and 77 females. Interrupted IVC was present in 14 patients. Hypo RV was evident in 128 cases. The oxygen saturation increased from 77.4% to 82.9% and from 73.8% to 84.7% in Groups 1 and 2 and the pulmonary artery pressure dropped from 24.6 mmHg to 15.8 mmHg and from 27 mmHg to 16.6 mmHg in both groups. Mechanical ventilation, inotropes, nitric oxide and oscillator high frequency jet ventilation were needed more in Group 2. Hospital stay was longer in Group 2. Six cases died in Group 1, and three died in Group 2.

Conclusions: Bidirectional Glenn shunts can be done with acceptable morbidity and mortality in some cases of pulmonary hypertension. Bilateral CPA had a more tough postoperative course than unilateral CPA. The mortality risk factors in bidirectional Glenn shunts are bilateral SVCs, Status post Damus Kaye Stansel (S/P DKS), S/P Norwood and high Pulmonary Artery Pressure (PAP).

Introduction

The bidirectional cavopulmonary anastomoses (CPAs) as a first or second stage procedure before embarking on a total cavopulmonary connection (“Fontan-type” procedure) has been used to normalise volume loading of the single ventricle at an early age. The presence of bilateral SVCs may represent a technical challenge in the performance of the BCPAs connections. In addition, bilateral BCPS have been also found associated with a higher operative mortality, an increased risk of thrombus formation, and a lower conversion rate to the “Fontan-type” circulation, if compared to a standard unilateral CPAs.[1]

Application of the concepts for the Fontan circulation have evolved whereby nearly all patients with single ventricle physiology,

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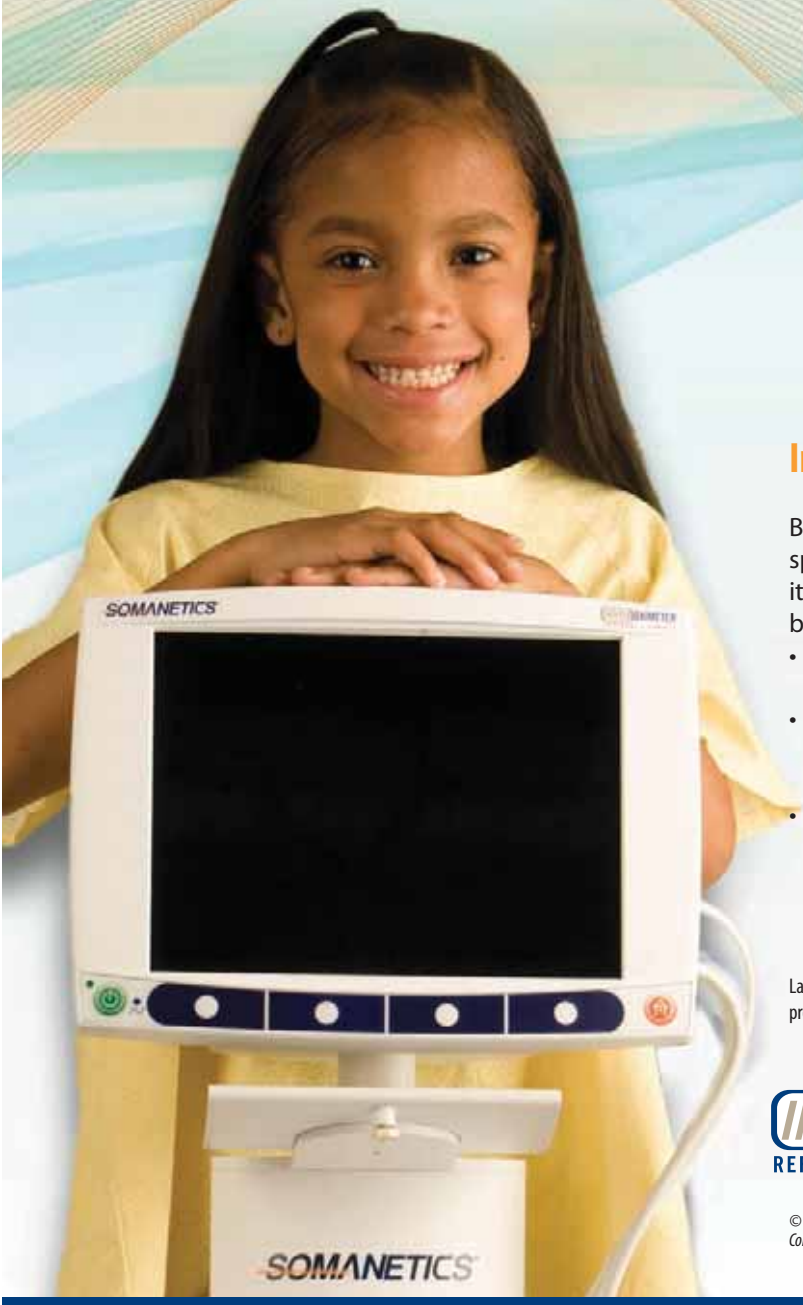

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including those with abnormalities of the systemic venous return are palliated on what has become known as the "Fontan tract." Although the initial results for the extension of the indications for the Fontan procedure were not always favorable, more recent studies have shown improvement.[1, 3]

Patients and Methods

This is a retrospective study including 204 cases of Glenn shunts operated upon in KFSH&RC; Jeddah, KSA, between 2000 and 2009. Preoperative evaluation included history taking, patient examination, routine laboratory and chest x-ray, ECHO diagnosis for all cases, and cardiac catheterization for higher risk cases, especially those with suspected or proved pulmonary hypertension. Detailed Consent from the family is taken. All our BDG were done through median sternotomy and with standard cardiopulmonary Bypass (CPB). No cardioplegia was given. Previous systemic to pulmonary artery shunts were disrupted by division, ligation or clipping. The azygos vein was clipped or ligated unless interrupted IVC or Glenn pressure markedly increased. The previous pulmonary artery band was tightened or main pulmonary artery was amputated from the confluence. Pulmonary Artery Banding was done as an associated procedure in forty cases. Pulmonary artery augmentation was done in twenty-four cases. The left SVC was occluded while the pressure in the cephalad side was monitored. If the pressure is more than 20 mmhg, this SVC was cannulated for CPB and a BBDG shunt was performed. Pressure monitoring was needed in the first few cases of our practice; however, we currently perform routinely BBDG in single ventricle with bilateral SVCs. ICU care included: monitoring, inotropes, mechanical ventilation, Nitric oxide and oscillator when needed, Head of the bed up to 45 degrees with furosemide, catopril, aspirin, dipyridamol, sildenafil with or without anticoagulation. Follow-up in the pediatric cardiology clinic was done. The median follow-up was 24 months with a range between 2-96 months.

Results

One hundred sixty patients had U-CPA (Group 1) and forty-four patients had b-CPA (Group 2) in King Faisal Specialist Hospital and Research Centre (KFSH&RC), Jeddah, in more than 8 years. They were 127 (94 in Group 1 plus 33 in Group 2) males and 77 (66 in Group 1 plus 11 in Group 1) females. BTS was previously done for 104 cases (80+24) and Pulmonary Artery Banding was done for 36 (29 and 7). Interrupted IVC was present in 14 (4 in Group 1 plus 10 in Group 2) patients. Hypo RV was evident in 128 (102+26) cases. The preoperative oxygen saturation increased from 77.4% to 82.9% postoperatively, and from 73.8% to 84.7% in Groups 1 and 2 postoperatively. Indexed pulmonary vascular resistance was <3.5 Units in 125 (95+30), > 3.5 units in 21 cases (15+6), and not assessed in 58 cases (50+8). The pulmonary artery pressure dropped from 24.6 mmHg to 15.8mmHg, and from 27 to 16.6 mmHg in both groups. CPB time was longer in Group 2 (69 min vs. 48min). Pulmonary artery banding was done concomitantly in 31 cases (25+6) and pulmonary artery augmentation in 22 cases (15+7). Mechanical ventilation, inotropes, nitric oxide and oscillator were needed more in Group 2 (0.8 days -5 hours -3 h -1h vs. 1 day -6h -5h -3h -1h) respectively in (Group 1 and 2). The hospital stay was longer in Group 2. Six cases died in Group 1, and three died in Group 2. ICU and hospital stay were longer in Group 2 (2.1 and 7 days vs. 2.6 and 10 days). Chylothorax was detected in 21 and 8 cases in both groups. Significant arrhythmias were detected and investigated in 30 and 6

cases. Re-intervention was needed in 13 cases (10+3) in the form of thrombosis of the Glenn shunt in 4 cases (3+1), bleeding in 6 cases (4+2) and thoracic duct ligation in three cases all in Group 1. Incorporation of the hepatic veins was needed in three cases (one in Group 1, and two in Group 2), and Fontan was performed in 42 cases (38+4). The data was statistically analysed using the Mann–Whitney test for continuous variables and chi-square test for categoric variables. Regression model was used for the potential risk factors. The continuous variables were: Age, Weight and CPB time, while the non-continuous variables included: Ventricular morphology dominance, bilaterality of SVCs with or without interrupted IVC, previous surgeries, use of CPB or not, and associated procedures (Table 1).

Table 1. Showing the Demographic, Radiological, Operative, Clinical, Therapeutic and Prognostic Data of Both Groups.

	Unilateral Glenn	Bilateral Glenn	
Morphology			
No.	160	44	
Hypo RV	102	26	
Hypo LV	58	18	
Sex			
Male	94	33	
Female	66	11	
Age (months)	17.9	19.3	
Weight (Kgm)	10.5	10.9	
S/P			
PAB	29	7	
BTS	80	24	
DKS	2	2	
Norwood	1	1	
LV diverticum excision	1	0	
IIVC	4	10	P<0.05
Preop. Saturation	77.4	73.8	
Postop. Saturation	82.9	84.7	
Preop. PAP	24.6	27	
Postop. PAP	15.8	16.6	
PVRI			
< 3.5	95 (53.1%)	30 (68%)	
>3.5	15 (9.3%)	P<0.05	
NA	50 (37.6%)	6 (13.6%)	
		P<0.05	
		8 (18.4%)	
		P<0.05	
Bypass Time	48	69	P<0.05
Associated			
PAB	25	6	
Pulm. A. Augment.	15	7	
ICU Stay	2.1 d	2.6 d	P<0.05
Mechanical ventilation	0.8 d	1 d	
Inotropes	5 h.	6 h.	
Nitric Oxide	3 h	5 h	
Oscillator	1 h	3 h	
Hospital Stay	7 d	10 d	P<0.05
Chylothorax	21 (13.1%)	8 (18.1%)	P<0.05
Arrhythmias	30	6	
Reoperation			
Thrombosis	3	1	
Bleeding	4	2	P<0.05
Thoracic duct ligation	3	0	P<0.05
Mortality	6	3	P<0.05
Next Surgery	39	6	
Fontan	38	4	
Incorporation	1	2	

Discussion

We are presenting a reasonably large series in a tertiary care centre in Jeddah over a span of more than 8 years. It includes 204 cases of Glenn anstomoses. More than 25% of them are b-CPAs. This ratio is higher than those of Iyer et. al., 2000, Chowdhury et. al., 2001 and Kim et. et. al., 2006, but less than those of Tanoue Y. et. al., 2007, who reported a ratio of 14.2%, 16%, 3.2% and 27.9%. This may be due to the random nature of the study with different socioeconomic and ethnic status.[1, 4, 5, 6] Male/Female was 1.64/1. The mean age was 17.9 months and 19.3 months, which is relatively high as we are a tertiary centre managing cases from all over our region with some delay and sub-optimal general and cardiac conditions. As per Mitchell ME et. al., 2006, and Tanoue Y. et. al., 2007, BDG should be performed between three to six months to achieve maximum benefits.[7]

However, BDG can be done up to two years old.[6] Our youngest case was 1 month, and the oldest was 13 years. BTS was previously done in 104 (51%), and previous PAB was done in 17.6% of cases, so about 68.6% received a first step palliation in the form of BTS and/or PAB. We followed a staged strategy in most cases, as the quality of life in patients after Blalock-Taussig shunt operation may be better than that in patients after BDG, as BDG is not used as the final operation for patients who cannot be completed by the Fontan circulation.[6] The median follow-up in our series was 24 months with a range between 2-96 months. Fontan was done for 38 and 4 cases in both groups (23.7% and 9%) respectively. Incorporation of the hepatic veins was done in three Kawashima cases. The aetiology of desaturation after Glenn is usually due to development of venous collaterals from the SVC to the IVC and intra-pulmonary arteriovenous shunting and reduction of the SVC/IVC flow ratio with age.[8]

We did believe in the staged palliation of single ventricle, especially early in our centre experience, However, we started to be a little more aggressive towards primary Glenn, so 31.4% received primary Glenn without previous palliation. The indication of BDG should be decided according to the surgical strategy aiming at the Fontan operation.[6] Concerns about the performance of BCPS in young infants may have to do with the history of unfavorable results after performing a classic Glenn shunt in infants along with uncertainty regarding the reactivity of the pulmonary vasculature in this group of patients. Between 1990 and 1995, over one-third (36%) of all BCPS procedures performed at the University of California at San Francisco were in infants <6 months of age. Early results in this cohort of young infants have been similar to those in all patients undergoing BCPS during the same period, with mortality rates of 4.8% and 5.1% and BCPS failure rates of 11.9% and 8.5%, respectively. The only independent risk factor for early death and BCPS failure was age, <1 month; and the strongest risk factor for early or late BCPS failure (death or take-down) was age <2 months.[9]

BDG procedure offers several potential advantages over the use of systemic to pulmonary shunts. In particular, it avoids pulmonary arterial distortion and also the additional volume load on the systemic ventricle created by the use of systemic to pulmonary shunts. Both these complications are important risk factors for subsequent Fontan repair. In addition, bidirectional cavopulmonary anastomosis is a more logical strategy as it represents an evolution of the patients' anatomy towards the final Fontan circulation.[10]

In all cases, we abolished the retrograde accessory pulmonary flow coming through the BT shunt as the goal is to try to achieve an effective/total pulmonary blood flow ratio of about 1. The more the effective and the total pulmonary blood flows coincide, the better the palliation. All efforts were made to obtain the maximum oxygen saturation with the minimum ventricular overload. The advantage of the BCPA over the Blalock-Taussig shunt is to allow an increase of effective pulmonary blood flow without an increase in total pulmonary blood flow and cardiac work. BCPA with Antegrade Pulmonary Blood Flow a better step towards Fontan than BCPA with an additional pulmonary blood flow through a Blalock-Taussig shunt.[11]

There was a significantly higher percentage of patients with Pulmonary Vascular Resistance Index (PVRI) >3.5 in Group 2 (13.6% vs. 9.3%), which may be a well-known high risk factor although there was no significant difference in the preoperative or postoperative PAP. This, in addition to longer CPB and heterogenous anatomical diagnosis, may explain the finding of longer Mechanical ventilation, inotropes, Nitric Oxide and Oscillator in Group 2 (0.8 days -5 hours -3 h. -1h. vs. 1 day -6h -5h -3h.) respectively in (Group 1 and 2), and the hospital stay was longer in Group 2, and that six cases died in Group 1, and three died in Group 2.

ICU and hospital stays were longer in Group 2 (2.1 and 7 days vs. 2.6-and 10 days). Using the univariate risk analysis, only the bilaterality of SVCS, interruption of IVC and PVRI >3.5 were significant risk factor for the longer ICU and hospital stay and chylothorax. As per Reddy VM et. al., 1997, elevated pulmonary vascular resistance did not appear to be a problem among any of the neonates and very young infants who fared badly after the operation, because they had normal pulmonary artery pressures and transpulmonary gradients throughout their postoperative courses. However, it has been pointed out that pulmonary artery pressures and gradients are not necessarily reliable indicators of resistance when pulmonary blood flow is low or when no good estimate of flow is available. Therefore, BCPS does not appear to be a viable option in neonates and should preferably be postponed beyond 2 months of age.[9]

In 2000, KFSH&RC; Jeddah, instituted a policy of offering BDG shunts to patients with pulmonary hypertension if their PVRI decreased to 3.5 WU/m2 on 100% oxygen irrespective of mean PAP and PVR in room air and mean PAP on 100% oxygen. Current suggested criteria to not offer BDG shunting to those with PVRI >3 WU and/or mean PAP >18 to 20 mm Hg by some and >25 to 30 mm Hg by others work well with respect to good outcomes and low mortality. However, there are three reservations to these guidelines. First, in the absence of pulmonary stenosis, elevated mean PAP does not necessarily indicate the presence of obstructive pulmonary vascular disease and may reflect only the transmission of systemic pressure to pulmonary circulation and high pulmonary flow. Second, because PAP and PVR are flow dependent, it seems inappropriate to use a uniform level of PAP or PVR as a criterion for patients with "under circulated" or "over circulated" pulmonary circulation. The former reduces and the latter increases pulmonary flow, which affects PAP and PVR, and the current approach ignores this important variable. Third, it has not been well-studied, in the presence of unrestricted and increased pulmonary flow and elevated PAP, what degree of elevated PVR would lead to the failure of a BDG shunt.[12]

Chylothorax was detected in 29 cases (21+8). It was suspected when the amount of drainage has a milky nature and diagnosis is confirmed by the presence of chylomicrons in the drained fluid with lymphocytes more than 80% with fluid triglycerides/ cholesterol ratio more than 1 or pleural fluid triglyceride level is >1.1 mmol/L. We think that it is due to high venous pressure (systemic and/or pulmonary), disruption of the minor lymphatic channels rather than thoracic duct injury. Our first line of treatment is a fat-free diet; monogen for at least 3-6 weeks with or without NPO, diuretics, captopril. For resistant cases, we may use a week course of steroids and or octreotides. We needed thoracic duct ligation in three cases; all were of the u CPAs group. So, although chylothorax incidence was significantly higher in Group 2 (18.1% vs. 13.1%), refractory chylothorax requiring thoracic duct ligation was only in Group 1. The higher incidence in Group 2 may be related to higher PAP and/or PVRI. However, we could not explain the refractory nature of three cases in Group 1. Chylothorax may be an indicator of additional pulmonary blood flow. Monogen is designed for infants and children with lipid and lymphatic disorders. The osmolality of Monogen is substantially lower than Portagen and most other elemental or fat-free formulas, a difference that improves gastrointestinal tolerance of the feed. Monogen also has a substantially higher energy level than alternative feeds. This point is important because patients with chylothorax have high energy requirements due to increased metabolic demand from the combination of chyle loss and hyper metabolism associated with surgery.[13, 14] Nine cases (4.4%) died during the follow-up - six cases from the Group 1 (3.75%) and three cases (6.8%) from Group 2. So, we can say that bidirectional glenn, whether unilateral or bilateral, can be done with acceptable morbidity and mortality. The median survival of those nine mortalities after the Glenn was 6 months. The accused causes of death were myocardial dysfunction, arrhythmias, tension pneumothorax and cerebrovascular strokes. The mortality risk factors in our study were bilateral SVCs, S/P DKS, S/P Norwood and high PAP. Kogon BE et. al., 2007 found longer CPB time and High CVP as the only significant mortality risk factor. They also found those two factors plus high transpulmonary gradient and right ventricular morphology and low body weight at the surgery time as the significant risk factors for the longer ICU and hospital stay.[15]

Conclusion

Bidirectional glenn whether unilateral or bilateral can be done with acceptable morbidity and mortality. The mortality risk factors in bidirectional Glenn Shunts are bilateral SVCs, S/P DKS, S/P Norwood and high PAP. Using the univariate risk analysis, only the bilaterality of SVCS, interruption of IVC and PVRI >3.5 are the significant risk factor for the longer ICU and hospital stay and chylothorax.

Limitations of the Study

This is a retrospective study with all the defects of retrospective studies with lack of randomization and unequal group sample sizes.

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What is Being Done in the USA to Screen Young Athletes and Children for Sudden Cardiac Arrest?

Cardiovascular screenings in children, young athletes and young adults – a review of the state of affairs in the U.S. 2009

By Lisa Salberg, CEO and Founder - Hypertrophic Cardiomyopathy Association

The purpose of this article is to discuss the state of affairs with regards to cardiovascular screenings in the young, particularly in athletes in the United States of America in 2009. The Hypertrophic Cardiomyopathy Association, (HCMA) has long been interested in this topic, as hypertrophic cardiomyopathy (HCM) is the leading cause of death in young athletes accounting for nearly 40% of the cardiac deaths in athletics. Given this fact, the HCMA has evaluated a number of the variables of the subject matter and have found many topics worthy of further investigation, public conversation and/or debate. This article is a review of the groups interested in the topic of cardiovascular screening of children and the current state of affairs in the US relative to cardiovascular screening; it is written to provide general information to those who may be interested in this important subject. There are several distinctive groups of people who have commented on cardiovascular screenings either through publication of scientific literature, advocacy groups statements, communities sponsoring screening events or individuals speaking out in public forums including the media.

The Medical Community

The scientific community uses peer reviewed literature, recommendations and guideline statements by recognized organizations such as the American Heart Association (AHA), The American College of Cardiology, and the Heart Rhythm Society to provide best practices. The American Heart Association's Statement on Pre-Participation Screening, most recently updated in 2007, is largely viewed as the authoritative document on the subject. This document discusses the American Heart Association's position with regard to pre-participation of competitive athletics and is silent on the issue of general cardiovascular screenings in young people as that was not the focus of this particular document.

The AHA has detailed 12 questions that should be included in all pre-participation screening forms, and offers suggestions that if any of the 12 questions indicate that a cardiovascular risk is present, additional attention from the medical community is warranted, including electrocardiogram and echocardiogram. The document does not say that electrocardiograms "should not" be done as part of the pre-participation screening process; it does explain that mandating this testing is not possible within the current health care system in the United States, which at its core is very different then the Italian system and prevailing Italian law on the matter.

The International Olympic Committee, IOC, and European Society of Cardiology, ESC, and many professional sports franchisees have adopted the inclusion of electrocardiograms, ECG, in the screening of athletes. There are some American physicians who questioned the AHA recommendations, and have written about the use of electrocardiogram screening in young people in either the general public or among athletes. The observations that there are fewer HCM deaths in countries that utilize ECG screening in athletes is appreciated as fact, however, a mechanism to screen the extensive, genetically and economically diverse US population has not been developed to-date. These articles have been largely silent on the logistical and access-to-care questions presented in the AHA article. Some of these articles are quick to point out the program used by Italy, without amply explaining the significant differences between the US health care system and the Italian health care system or the infrastructure of the athletic screening programs. The differences between these two systems will be explained later in this document. The role of the IOC or a professional team in evaluating elite level athletes cannot be compared to evaluating high school, jr. high school or recreational athletes.

The definition of "athletes" needs to be identified and communicated when referring to children involved in sports in the US. When looking at the problem of sudden death in the young, are we only to look at elite level athletes, college level or varsity players in high school? Or are we to look at the little league ball player, the T-ball player or the children in physical education classes in schools across the country? The question of screening all children, rather than only athletes, has been raised as well, which creates other logistical issues that require much deeper scientific evaluation.

The cost effectiveness of the inclusion of ECG with history needs to be more closely examined once the population of 'athletes' is better defined. There is some data on this subject; however, additional research is needed in this area.

The Advocacy and Sports Community

There are a number of advocacy groups, patient groups and sports programs that have an interest in ensuring our young people have access to cardiovascular care in a timely manner. These groups have a long-standing history with either the diseases that cause sudden death in the young, with young people and/or athletes directly.

The concerns in this group center around the access to health care, quality evaluation of test results, education of those participating in screenings, patient privacy and the emotional well-being of youth pre and post screening. For the most part, they share the core goal of improving the lives of those who participate in athletics and maintaining their safety.

The Impassioned Parent Community

There is no more passionate group of individuals than those who have lost a child to sudden cardiac arrest, or nearly lost a child from an aborted cardiac arrest. Many of them believe that if screening programs included electrocardiograms or echocardiograms, their

child's undiagnosed cardiac condition would have been detected, and their lives would have been saved. They may be correct. The loss of a child is catastrophic and can lead those left behind looking for the most meaningful way to ensure that others do not suffer the same fate as their lost loved one.

Some parents have sought to perform community heart screening events, most of which have collaborated with local hospitals or cardiologists who volunteer their time to assist with the event. Some programs use a questionnaire with an electrocardiogram and/or echocardiogram, some programs use all three. A number of these parents have sought to introduce legislation on a State level with no successful passage or implementation to date.

The quality of these screening programs has been varied and the data collection has been haphazard. It is unclear as to the true yield of these efforts, and it would benefit all if the data was compiled and shared through a (de-identified) public access database.

The General Public

Largely, the general public is currently unaware of the concept of cardiac screening in the young. Over the past 5 years, there has been increasing news coverage about the concept with lukewarm response from the medical or general community. As the United States enters an era of reevaluation of its health care system, the language used to describe the cardiac health care of our children and young adults may become part of the public vernacular.

Mechanisms for Payment of Screenings in the Young

The use of the pre-participation evaluation, PPE, screening questionnaire including the AHA's 12 recommended items and a physical are normally a covered expense in the majority of health plans in the United States. In the event ECG screening were to be needed based on the results of the PPE and physical the ECG would be a covered expense. In the event that ECG screening indicated further cardiovascular work-up was needed, in nearly all cases this would be viewed as a covered expense. If the child or young adult is covered by health insurance, through either a private plan or State program, mandates, via ERISA (Employee Retirement Income Security Act), should be put in place requiring cardiac preventative care be a covered expense as a "Well Care" benefit. As most policies now stand, cardiovascular testing for those under 40 years of age is rarely covered as a 'Well Care" benefit. Currently, there are no Federal or State mandates for such coverage and this remains a huge hurtle to overcome.

Currently in the United States, there are over 47 million uninsured individuals. Many of those in the ranks of the uninsured are children and young adults. If a systematic cardiac screening program is to be instituted, a payment system must be identified for the uninsured to ensure they have equal access. The recent passage of SCHIP may insure that more uninsured children have coverage, but it does nothing to insure that once they become adults they will have access to care.

Another significant concern is the identification of abnormal test results which could later be interpreted as "pre-existing conditions" and impact the ability to acquire health coverage in the future. While there have been campaign promises from the current administration, there has

United States and Canada Pediatric Cardiologists, Congenital Heart Surgeons in Hospitals Providing Open Heart Surgery for Children 2008/2009

Complied by Dilip R. Bhatt. MD, FAAP; Kenneth Jue, MD, FAAP; Janice Stillwell (Project Manager)

This Directory is available in a PDF file: www.CongenitalCardiology.com/DIR2008.pdf:

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CONGENITAL CARDIOLOGY TODAY

been no legislation presented to remove the long-standing practice of using pre-existing conditions as a means of rating policies and, in some cases, outright denial of coverage.

Evaluation of Test Results

This process begins with the appropriate use of the AHA questionnaire by parents, athletes, trainers, coaches, school administrators, and others involved in youth athletics. The first, and most cost effective tool that can be used to help identify those at risk for sudden cardiac arrest is the pre-participation screening tool. In the event any questions yield a positive answer further cardiac evaluation by a cardiologist, should be required prior to participation in competitive athletics.

If there is a need for an electrocardiogram, it is imperative that the results be interpreted accurately, and it must be understood that between 10-15% of those with HCM will have a normal ECG. While ECG interpretation may sound like a relatively easy step, it may, in fact, be the most difficult part of the process. In the event a result is interpreted as 'abnormal,' and, therefore, triggers additional testing, there is a significant chance that this 'abnormality' will be viewed as inconsequential.

If an echocardiogram is required to rule in or out hypertrophic cardiomyopathy (or other conditions), it is imperative that the quality of the test and the reading of the results are done by someone with ample experience. The HCMA has worked with thousands of those diagnosed with HCM, and has found a significant lack of appreciation of HCM among cardiologists nationally. In some of these cases, patients with HCM have been told by a cardiologist they did not have the condition, when, in fact, they did; and conversely, some have been labeled with the diagnosis only to find out there was no clinical reason for the diagnosis. In some cases, those with HCM have participated in screening programs offered to the general public and have defied identification of their long standing HCM. There is a great deal of room for improvement, training and education among medical professionals to improve their ability to recognize HCM.

To be more specific, some screening programs include a "limited" echocardiogram (lasting under 5 minutes or less). There has never been any peer-reviewed literature supporting the use of this type of screening as clinically effective for the identification of HCM. Due to the heterogeneity of HCM the limitations of "limited" echocardiograms are numerous and no data exists in the literature to identify what the views should be included in a "limited" echocardiogram and what the yield of this type of screening. Thus there are significant problems with the informed consent process, as it is unclear as to what the parent or athlete is consenting.

Education

Parents need to understand the reasons the questions in the AHA pre-participation guide are important, and why it is important they provide accurate information on these forms. They should also be encouraged to add information they feel important or expand on answers they are uncertain about. For example, many parents do not know their child's complete family medical history. This may be for a large number of reasons, which may include: lack of communication with extended family, adoption, the child may be the product of egg or sperm donation, or more complicated family relationships. In addition, parents and students may be afraid to answer questions honestly for fear of being disqualified from competition. Efforts should be made to ensure that complete and honest answers are given. The positive answer to one or more of the AHA suggested questions or physical findings can result in referral for further evaluation. In the event of a positive answer to one or more of these questions, the parent can request additional screening and in these cases there is a higher likelihood that insurance will cover the testing and evaluation.

Coaches, athletic trainers, team administrators and those involved in the administration of sports, band, schools, club sports and related programs must create strong protocols for the use of pre-participation questionnaires. These protocols should include training programs to educate parents and athletes about the importance of the form.

In the event electrocardiogram, ECG, or echocardiogram are included in a screening program, it must be clearly noted to the participants that up to 15% of those with the leading cause of sudden cardiac arrest, HCM, may have a normal test ECG. In addition, if limited echocardiogram is used for screening, it should be clearly explained that the role of "limited" echo has not been proven diagnostic for HCM.

Privacy

In the event that cardiac screenings are being conducted in a community screening event, the location of the screening and maintenance of records are important factors. Children, along with their parents, have the right to expect privacy of their medical information, and providing mass screening environments does not lend itself to ensuring privacy is maintained. Specifically, in the event a screening is conducted in a group environment at a school or similar venue with an entire team required to attend, the discovery of a potential problem in an athlete that requires further evaluation may require that athlete to be benched pending further test results. This creates an environment that appears nowhere else in the health care system, as a patient can not be assured that his or her medical data is private, to share with whom they choose, when and if they choose to do so. There are psychological implications to the athlete in these situations that must be addressed.

Pre and Post Screening

Young people, parents, coaches, trainers, administrators, and others need to completely understand the screening process prior to being able to implement any program. There are still many questions that have yet to be addressed about the psychological implications of screening programs on all those involved and there are no programs in place to deal with the many potential outcomes of screening.

There are many areas that must be considered prior to holding or participating in a screening event, including either ECG or Echo, in addition a physical and PPE form including: no clear expectations about the amount of time the screening results are valid for, no protection from discrimination in access to health care, disqualification from scholarship opportunities, access to support mechanisms in place to deal with potential diagnosis, disqualification from competitive athletics, difficulties acquiring life insurance later in life, and the fact there is currently no research on the impact of the screening processes impact on the healthy population.

Italian vs. United States Health Care System and Laws

In the United States we have no true health care "system" that ensures all legal residents

access to care. Health insurance is managed State--by-State, and only under Employee Retirement Income Security Act (ERISA) are there mandates on what must be covered under health care plans that fall under its governance. Many health plans, including self-funded programs, are not covered by ERISA, and are therefore, not subject to Federal mandates. Each state has its own health Insurance Board which approves plans to be sold in the individual state. There are no guarantees of health care in any state, with the exception of Massachusetts, and pre-existing conditions may be held against an individual for rating of policy premiums or denial of coverage outright. There are some state-sponsored health plans that are federally funded (SCHIP), but this coverage is for children, and leaves them uninsured when they turn 18 years of age. Medicare and Medicaid insurance generally do not cover children, who may be involved in athletics, although some children may fall under these Federal and State forms of insurance.

Physician compensation is not governed by any formal body, and hospitals are free to charge what the market will bare, and the insurance companies negotiate. The uninsured pay for services or apply for charity care within a hospital, and are granted services only if they can prove financial hardship.

In Italy the health care system is funded based on a regressive payroll tax (decreases as the amount subject to taxation increases). All residents are covered in the health care system. The tax starts at 10.6% of income for the first €20,000 (approximately $27,000 us) and drops to 4.6% of income between €20,100 and €77,480 (approximately $27,000 and $102,000 us). The rest of the funding comes from federal and regional general taxation (i.e. income and value-added taxes). The regions are responsible for health care provision. The Ministry of Health funds these regions according to a formula based on weighted capitation and past spending. Then the regions allocate these funds to Local Health Authorities (LHA). Private health insurance in Italy is uncommon, but is occasionally offered by employers. It is not possible to opt-out of the National Health Insurance system, and insurance premiums are not tax deductible. Many Italians pay for private health care. It is estimated that about 35% of Italians use at least some private health services, but the public sector certainly dominates the private in terms of its relative importance. Physician compensation is based on physicians being paid via capitation. Hospitals are paid via DRGs.

Italians have limited choice of their physician, but more than in the UK or in Spain. They must register with a general practitioner (GP) in their LHA. For any specialist services, patients must get a referral from their GP. As far as co-payment and/or deductibles are concerned, inpatient and primary care are free. For tests, diagnostic procedures and prescription drugs, co-payments are as high as 30%. However, 40% of the population (e.g. the elderly, pregnant women, kids) are exempt from these co-payments.

Laws Governing Student Athletes

In Italy, since 1950, it has been mandatory for all professional and amateur athletes to obtain medical certification of their eligibility to participate in their sport. In 1971, the government introduced laws intended to safeguard the health of all those practicing sport at both competitive and non-competitive levels and that regulate preventive PPS. Competitive athletes must undergo a yearly preventive screening protocol including: a past medical history, clinical evaluation, urinalysis, electrocardiogram (ECG) at rest and after a

step test, and pulmonary function tests. This evaluation can only be performed by doctors holding a diploma of specialist in sports medicine and registered with the Italian Sports Medicine Foundation (FIMS). These professionals are legally responsible for the accuracy of this assessment, being the final judges of the subject's eligibility to participate in sport. At present, in Italy, about 5 million people undergo the cardiovascular PPS each year.

In contrast to Italy, The US has no federal law mandating health care services to students. It would be up to each state to set up laws that would regulate such screening. If the Federal government wishes to draft legislation on this issue that would preempt any State laws that are weaker then the Federal law. To-date there are no states with legislation mandating pre-participation screening questionnaires or dictating which diagnostic test to use. In the United States, the only Federal law governing athletics in public schools is Title IX, which mandates that males and females have equal access to participation in sports.

Cardiac Evaluation of Non-athletes

In the United States, a recent review of the Center for Disease Control registry of death certificates indicates that 1600 people per year die with HCM listed as cause of death on their death certificate. Of those who die, over 52% (832) are under the age of 54, 32% (512) are under the age of 44 and 7% (112) are under 24 years of age. The Sudden Death in Athletes registry reports an average of 66 deaths per year (range 50 to 76) over the last 6 years. HCM is the most common cause of death in this population, accounting for approximately 36% (24) of these deaths. This means that more four times as many 'non-athletes' will die from HCM as athletes. This factor must be taken into consideration when reviewing the core question about screening, "who should be screened?" If we are able to identify, treat and screen the family members of those children identified early in life we may have the ability to improve the quality of life and add to life years saved for thousands of people.

The HCMA is hopeful that its recently updated "Pediatric and Young Adult Sudden Cardiac Arrest Risk Assessment Form" will be used as a tool to help identify those at a risk. The form was created with the AHA pre-participation screening guideline questions, yet moves beyond the athlete to all children and young adults.

Conclusion

In conclusion, there is no lack of passion from any of the players in this arena. There are a great many questions for public debate and opportunities for change. The core question is: Do Americans believe this is an issue they should be concerned with, and are they willing to make fundamental changes in the health care system that may facilitate change in this area? In a resent article, Dr. Paul Thompson concluded, "Experience suggests that when experts disagree, there is a dearth of reliable data. The present data suggest that the problem, at least in the United States, is not so huge that we must leap to action. We need more and better data on the cardiovascular risks of athletics, the false-positive rate of screening strategies when used by non-experts, the cost of tests and procedures generated by screening, and if possible, actual controlled trials of screening strategies. Good data often simplify complex problems." I would argue that Dr. Thompson's comments

could easily apply to all young people, and not be reserved only for athletes.

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JANUARY 2010
MEDICAL MEETING FOCUS

Evolving Concepts in the Management of Complex Congenital Heart Disease II
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Selected Meeting Objectives:
- Determine optimal management strategies for common diagnostic and treatment problems encountered in an outpatient office practice.
- Understand and manage the diagnosis and treatment of cardiomyopathy and pulmonary hypertension in infants, children, young adults
- Optimize imaging modality selection to obtain an efficient & cost effective diagnosis.
- Diagnose and treat common and complex arrhythmias in patients of all ages.
- Utilize modern catheterization techniques as part of an integrated invasive treatment program for congenital heart disease.
- Integrate the Hybrid approach with modern variations of the Norwood and Fontan procedure to optimize care of infants with HLHS.
- Utilize modern medical and surgical strategies to treat complex CHD.
- and more….

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- John W. Moore, MD, MPH

Faculty:
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- Daniel Bernstein, MD
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GE Healthcare Introduces New Intracardiac Echo Functionality for the Vividi and Vividq Ultrasound Systems

Wauwatosa, WI – GE Healthcare and Biosense Webster, Inc. (BWI) announced new versions of the Vivid i and Vivid q Cardiovascular Ultrasound systems, with support for two new intracardiac echo (ICE) catheters. The Vividi systems with version BT10 software now support the Biosense Webster Acunav® 8F ICE catheter. The 8F catheter has a 33% smaller cross-sectional area than the AcuNav 10F catheter, making it well suited for patients that cannot tolerate a larger catheter.

The companies also announced new support of the Vividi and Vividq systems with the SoundStar™ 3D ICE catheters, as well as compatibility for the Vividi and Vividq systems with the Biosense Webster CartoXP System with the CartoSound version 9.7 software module. The CARTOSOUND Module and SOUNDSTAR 3D Catheter integrate real-time intracardiac echocardiography (ICE) imaging into the CARTO system environment, enhancing visualization and navigational confidence during complex EP procedures.

Biosense Webster is a pioneer in EP diagnostic catheters and is an innovative provider of advanced diagnostic, therapeutic, and mapping tools. GE and Biosense first announced the intent to create a combined AcuNav catheter/Vivid i system at HRS in 2009 and offered the first unit for sale, with support for the Biosense Webster 10F ICE catheter, in October 2008.

The small and compact Vivid i systems deliver imaging performance comparable to that of today's high-end console ultrasound systems without crowding the lab. Combined with the exceptional ICE catheter technology performance and navigational versatility of the ACUSON AcuNavTM 10F and 8F ultrasound catheter, the system helps cardiologists confidently navigate complex interventional procedures for a wide range of patients.

According to Hooman Hakami, General Manager, Interventional at GE Healthcare, "Customers value the ICE images they are getting from the Vividi system using the AcuNav 10F catheter. Support for these new ICE catheters from our partner, Biosense Webster, l allows our customers to utilize the Vividi system for a wider population of patients and procedures."

For more information about GE Healthcare, visit the web site at www.gehealthcare.com.

Children with Fatter Midsections at Increased Risk for Cardiovascular Disease

Augusta, GA – Children with more fat around their midsections could be at a higher risk of developing cardiovascular disease later in life, researchers say.

"While general obesity certainly has its own set of risks for the heart, we now know that all fat is not created equally," says Dr. Reda Bassali, an Associate Professor of Pediatrics in the Medical College of Georgia School of Medicine and co-author of a study published online in the *International Journal of Pediatric Obesity.*

Increased waist circumference has long been linked to cardiovascular risk in adults because visceral fat — found in and around organs in the abdominal cavity — is more metabolically active, which can dramatically increase the risk of cardiovascular disease and type 2 diabetes.

The study suggests routine waist measurements in obese children could predict which ones had developed risk factors for cardiovascular disease, such as higher fasting insulin levels, a precursor for diabetes; lower levels of high density lipo-protiens, also known as the good

cholesterol; and higher levels of triglycerides, the fatty particles found in the blood.

"What we are asking is whether the children with larger waists already showed signs that put them at higher risk," Dr. Bassali, also a pediatrician at the MCG Health Children's Medical Center, says. "To find out whether children eventually developed cardiovascular disease, we'd have to follow them long-term."

In a sample of 188 obese children, ages 7-11, those with the largest waist circumferences – above the 90th percentile for their age – were three times more likely to have high triglycerides and nearly four times more likely to have lower levels of HDL. They were also 3.7 times more likely to have high fasting insulin levels.

"What that means is that children with a waist circumference at or above the 90th percentile are at a greater risk of developing the warning signs of cardiovascular disease," Dr. Bassali says. "Our results indicate that routine clinical measurement of the waist may help clinicians identify which obese children are at a greater risk."

"There is a lot of discussion about the apple versus the pear body shape, with the pear being more desirable," Dr. Bassali says. "Unfortunately, we don't have a real explanation why some people gain weight in the center of their body and others gain it, for instance, in their thighs. It could be environmental. It could be genetic. It could be a combination of the two."

These results, however, could provide researchers and clinicians with another way to measure possible risk and possibly prevent future health complications.

"The gold standard, when it comes to intervention strategies, has always been whether a child fell into a certain range with their body mass index (calculated using height and weight)," he says. "These results suggest that waist circumference could provide an additional measurement of risk. The intervention strategies would be the same."

Other authors on the paper included Jennifer Waller, Associate Professor of Biostatistics; Jerry Allison, Professor of Radiology; and Catherine Davis, Clinical Health Psychologist in the Georgia Prevention Institute.

Pioneering Research Forms Basis for First-Ever Paediatric Hypertension Guidelines

Newswise - Comprehensive guidelines for the treatment and management of hypertension in children and adolescents are being published for the first time in the latest issue of the *Journal of Hypertension*.

Prepared by a Task Force established by the European Society of Hypertension, the guidelines should prove to be an invaluable source of information for physicians, nurses and families dealing with hypertension in young people.

The necessity for the guidelines has become increasingly clear to physicians in light of growing evidence that cases of mild hypertension in children and adolescents are much more common than previously thought. In addition, progress made in pathophysiological and clinical research has made clear links between paediatric hypertension and cardiovascular disease later in life, highlighting the need for improved cardiovascular prevention strategies for pre-adult individuals.

The Task Force, established by the European Society of Hypertension and headed by Dr. Empar Lurbe of the University of Valencia, has combined considerable amounts of scientific data with clinical experience in order to represent a consensus among specialists involved in the detection and control of high blood pressure (BP) in children and adolescents. It is hoped that the publication of these guidelines will call attention to the burden of hypertension in children and adolescents, and encourage public policy makers to develop a global effort to improve identification and treatment of high BP among young people. Primarily, however, these guidelines provide practical strategies for diagnosing and treating hypertension in children and adolescents. They include:

- Definition and classification of hypertension
- Diagnostic evaluation
- Preventative measures
- Evidence for therapeutic management
- Therapeutic strategies and approaches under special conditions
- Treatment of associated risk factors
- Screening for secondary forms of hypertension

The Task Force also suggests strategies for long-term follow-up, and make recommendations for future research in the field.

The guidelines will certainly prove vital in combating the growing epidemic of cardiovascular disease in adults, by emphasising the need for preventative strategies to be implemented from an early age. As Dr. Lurbe comments, "Action is required to address this problem in one of the most vulnerable and precious sectors of our society: children and adolescents."

Do you or your colleagues have interesting research results, observations, human interest stories, reports of meetings, etc. that you would like to share with the congenital cardiology community?

Submit a summary of your proposed article to Congenital Cardiology Today at:
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