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Role of Cross-Sectional Imaging in Repair of Neonatal Hypoplastic Aortic Arch

By Rabin Gerrah, MD; Dianna Bardo, MD; Rachel Sunstrom, PAC; Rich Reed, PAC; Stephen Langley, MD

Introduction

Hypoplastic Aortic Arch (HAA) is a common diagnosis in Congenital Heart Diseases. Altogether, aortic malformations account for 15-20% of all congenital cardiovascular diseases.¹ HAA may occur as an isolated lesion or in conjunction with other congenital cardiac anomalies. Hypoplasia might entail single or multiple segments of the aorta and, hence, the wide range of surgical options. The surgical treatment for HAA has evolved with advances in surgical techniques and improved imaging modalities. As a general rule, the anomalies of the distal arch and descending aorta are approached via left thoracotomy and the ascending and proximal portion of the arch are approached through midsternotomy. Different guidelines regarding when to use each approach based on anatomic landmarks or morphologic characteristics have been described.² A management dilemma rises when the common hypoplasia of the distal arch is extended towards the transverse arch. In some instances, the presence of aortic arch hypoplasia has been ignored in the setting of severe isthmus stenosis or a coarctation with the hypothesis that reestablishing flow in the aorta after relief of obstruction will stimulate the aorta to grow and the hypoplasia will resolve.^{3,4} Instead, in many institutions any extension of hypoplasia proximal to the distal arch is considered Complex Arch Disease and

repaired through a midsternotomy approach. The major differences between the two approaches are the need for cardiopulmonary bypass (CPB) to reconstruct the aorta and cerebral perfusion techniques or deep hypothermic circulatory arrest in the midline approach versus no CPB in the thoracotomy approach. Achieving the same outcome with no CPB would exclude the complications associated with the use of it, and would be a preferred approach, if feasible.

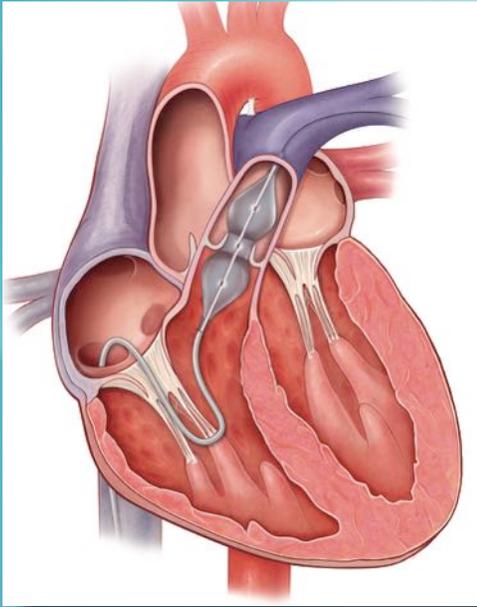
Echocardiography, has become the gold standard for imaging and diagnosis in Congenital Heart Disease (CHD). It is used primarily for surgical decision-making regarding the approach for repair of HAA. This diagnostic modality has limitations, such as lack of appropriate acoustic windows in specific body habitus or posterior structures, and limited spatial resolution. Recently, cross sectional imaging (CSI) including: computerized tomography (CT) and Magnetic resonance imaging (MRI), have become a strong modality in diagnosis of CHD. The different variations of HAA are repaired based on surgeons' experience and preference and to-date, no standard morphological or anatomical definitions or numeric parameters exist to define the exact type of pathology and the appropriate repair-type based on these definitions.

In repair of HAA, whatever type of surgical repair is undertaken, it needs to provide a definitive treatment with exclusion of the hypoplastic portion of the aorta while ensuring the patient undergoes the lowest risk procedure possible. It has not been studied

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whether or not CSI adds additional information to existing echocardiographic studies and whether or not these additional images may impact surgical decision-making. It is known, however, that 3-dimensional reconstruction achieved uniquely by CSI is the most realistic imaging modality, and provides a simulation for direct visualization of the cardiac anatomy or preoperative planning.

We hypothesize that better definition of the aortic arch by CSI will identify more patients with complex HAA who are still amenable to the lower risk thoracotomy approach, rather than the midsternotomy approach. Based on our findings we propose a management algorithm.

Patients and Methods

This study was approved by Institutional Review Board of Oregon Health and Science University. Computerized database was used to identify the study population. All patients diagnosed with HAA in the neonatal period by echocardiography who underwent surgery from May 2008 to March 2013 at our institution were initially selected for inclusion in the study.

Echocardiographic description of arch anomaly included both descriptive and quantitative information on arch segments as well as cardiac chambers and function. Whenever two measurements from the same patient varied more than 10%, they were considered as inaccurate, and were excluded for use in surgical management.

CSI with 3-dimensional reconstruction was obtained in HAA cases considered complex cases or when the data from echocardiogram regarding the arch anatomy was inconclusive. Two-dimensional images from CSI were used for measurements of length, diameter and distance of the structures from each. For the purpose of standardization and comparison between the different modalities, the aortic arch standard segmentation definition was used: Segment 1 being the section between the brachiocephalic artery and the left common carotid artery; Segment 2, the section between the left common carotid artery, and Segment 3, between the left subclavian artery and the isthmus (Figure 1). Each segment was defined as

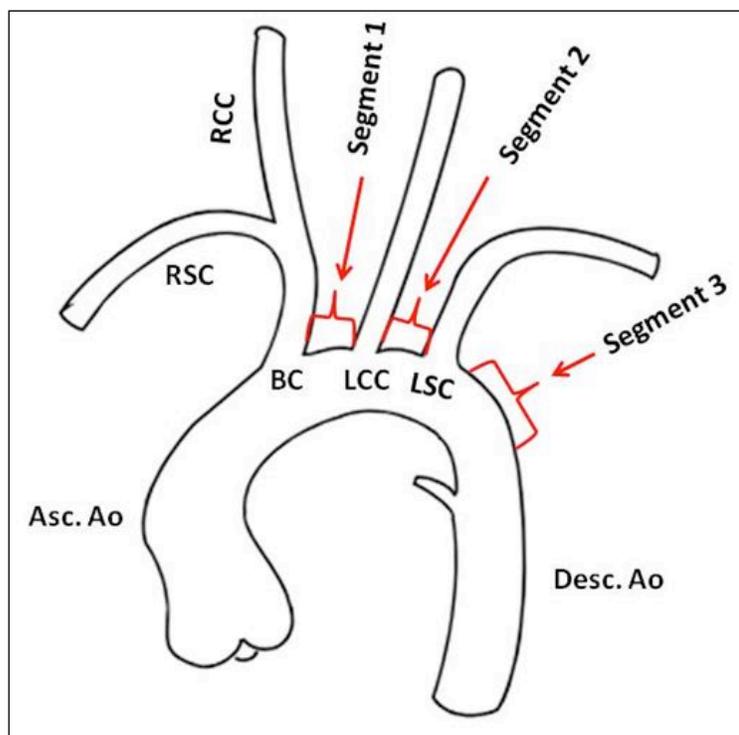


Figure 1. Segmentation of the aortic arch. Asc Ao= Ascending aorta, BC= Brachiocephalic, Desc Ao= Descending aorta, LCC= Left common carotid, LSC= Left subclavian, RCC= Right common carotid, RSC= Right subclavian.

normal or hypoplastic in either imaging method. Since the definition of a HAA is controversial,⁵ we used a combination of the three definitions and parameters assumed to be the most predictive. These criteria included: less than 50% ratio of the diameter of the transverse arch to the descending aorta,⁵ size of the arch in mm less than the patient's weight in kg +¹⁶ or aortic arch diameter Z-score is less than -2.0.⁷

We excluded patients with Hypoplastic Left Heart Syndrome (HLHS) or other single ventricle physiology, patients with interrupted aortic arch, right sided aortic arch and bovine arch.

All cases were presented in cardiology and in a cardiac surgery forum for surgical decision making. Whenever the anatomy of the arch was complex or in cases with questionable imaging quality of echocardiography, a repeat echo was performed or CSI was obtained and these patients were discussed again for final surgical planning purposes.

Surgical Techniques

The surgical plan was made for either a midsternotomy or left thoracotomy approach after reviewing all patients' data and images if their quality was satisfactory. In all patients, blood pressure was monitored proximal and distal to the repaired area before and after surgery to evaluate for any pressure gradient or residual narrowing at repair site. Near infrared spectroscopy were monitored to assess the adequate cerebral blood flow. In the midsternotomy approach, the surgery was performed on CPB and the repair was completed using a patch material on the lesser curvature of the aorta to augment the aorta during a period of selective cerebral perfusion, as described previously.⁵

For the thoracotomy approach, the procedure was performed as described for classic repair of extended end-to-end anastomosis.

Statistical Analysis

Results are presented as mean \pm standard deviation or median and range for continuous measurements. Due to a small number of samples in each group, non-parametric statistics as Mann-Whitney test and one way analysis of variance were used to compare the groups. In all measurements, a *p*-value of less than 0.05 was considered as significant.

Results

Twenty-six patients were enrolled in the study. The median age was 11 days (range 3-28 days) and weight was 3.3 kg (range 2.1-4.4 kg).

Variable	Group 1 (n=16)	Group 2 (n=4)	Group 3 (n=6)	P Value
Age (days) (All 13.6 \pm 8.6)	12.2 \pm 8.6	16.6 \pm 11.2	14.5 \pm 6.6	0.1
Weight (kg) (All 3.3 \pm 0.6)	3.2 \pm 0.7	3.0 \pm 0.5	3.6 \pm 0.3	0.3
BSA (m ²) (All 0.20 \pm 0.02)	0.20 \pm 0.03	0.20 \pm 0.02	0.21 \pm 0.01	0.3
CSI (n) {CT/MRI}	{2/2}	{3/0}	{4/2}	0.1
Length of hospital stay (days) (All 10.4 \pm 9.2)	9.1 \pm 6.3	16.8 \pm 14.6	8.3 \pm 7.1	0.2
Mean follow-up period (days)	942 \pm 490	1102 \pm 492	610 \pm 461	0.3

BSA; Body surface area, CSI; Cross sectional imaging, CT; Computerized tomography
MRI; Magnetic resonance image

from echocardiography was incomplete or omitted due to variability as described in methods.

Group 3 was further analyzed to identify parameters affecting the surgical planning. Arch anatomy analyzed by CSI data in Group 3 provided the following measurements: Segment 1, 4.9±0.9 mm, Segment 2, 3.7±0.7 mm, and Segment 3, 2.7±0.4mm. In all these patients, the diameter of the first segment was similar to the descending aorta diameter (4.9±1.1mm vs. 5.0±0.9mm, p=0.9) and the length of this segment was similar to its diameter. In these patients, an extended end-to-end anastomosis repair via thoracotomy was performed rather than arch reconstruction on CPB (as was initially planned based on echocardiogram).

It appeared that numeric values of Segments 1 and 2 were the main parameters determining the type of surgical approach. These parameters were accurately achievable only from CSI. In presence of hypoplastic or short Segment 1 of aortic arch, a midsternotomy approach was necessary; however, when the Segment 1 had a diameter and length similar to descending aorta, a left thoracotomy approach with extended end-to-end anastomosis was possible regardless of the anatomy of Segments 2 and 3 of the aortic arch. Presence of these determining factors quantified by CSI, created a common language and standardized the decision-making process.

We hypothesized that for an extended end-to-end anastomosis using our previously described technique, a normal ascending aorta and an appropriate size and unrestricted flow through the first branch of aorta

are necessary. These conditions enable safe clamp placement and performance of extended-end-to-end anastomosis.

Using this surgical strategy, there was no patient with a residual narrowing. All patients had a gradient of less than 10 mmHg determined with blood pressure recordings. One patient in Group 1 had a gradient of 11 mmHg after completing the repair.

The average length of hospital stay was not different between the groups, 10.42±9.2 days in the entire group: 9.1±6.3 days in Group 1, 16.8±14.6 days in Group 2, and 8.3±7.1 days in Group 3, p=0.2.

	Group 1 (16)	Group 2 (4)	Group 3 (6)
Anatomic description	Discrete hypoplasia of distal arch and/ or discrete coarctation of aorta. Proximal and distal arch, ascending, and descending aorta: within normal limits.	Hypoplasia of distal arch involving segment 2 and coarctation of aorta, presence of large ventricular (and atrial) septal defect/s.	Hypoplastic aortic arch, complex hypoplasia involving proximal arch and coarctation, long hypoplastic segment and/or torturous segment of aortic arch.
Adequate quantification of arch segments (patients)	Segment 1	14/16 (87%)	2/4 (50%)
	Segment 2	16/16 (100%)	3/4 (75%)
	Segment 3	16/16 (100%)	3/4 (75%)
Accurate branching pattern description (patients)	9.1±6.3	16/16 (100%)	3/4 (75%)

	Segment 1, length (mm)	Segment 1, diameter (mm)	Segment 2, diameter (mm)	Segment 3, diameter (mm)
Echocardiography	4.6±0.1*	5.0±0.2*	3.8±1.0	2.8±0.3
CSI	4.8±0.4	4.9±0.9	3.7±0.7	2.7±0.4
P Value	0.2	0.1	0.4	0.5

*Incomplete data, the measurement varied in different studies or the image quality was insufficient for accurate measurement
CSI; Cross-sectional imaging

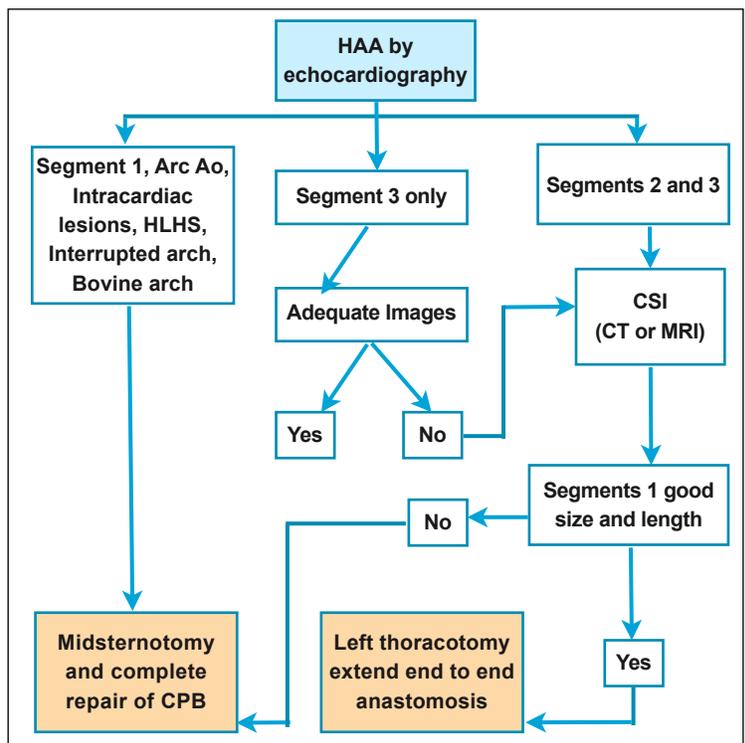


Figure 3. Proposed surgical management algorithm for Hypoplastic Aortic Arch. Asc Ao = Ascending Aorta, CPB = Cardiopulmonary bypass, CSI = Cross-sectional imaging, CT = Computerized tomography, HAA = Hypoplastic Aortic Arch, HLHS = Hypoplastic Left Heart Syndrome, MRI = Magnetic resonance imaging.



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There were 3 readmissions, 2 in Group 1, 1 in Group 2 and none in Group 3. All readmissions were non-cardiac related. There was no peri-operative mortality, and no patient needed re-intervention in the follow-up period in all groups.

The mean follow-up was similar between the groups. No patient was lost to follow-up at the time of the report of this study. The endpoints for follow-up were death, diagnosis of re-coarctation, need for procedure either surgical or percutaneous for relief of aortic narrowing. During this follow-up period none of these endpoints were observed.

Based on this observation, we propose a treatment algorithm for patients with HAA utilizing CSI with proper indications (Figure 3).

Discussion

HAA is a common consideration in congenital cardiac surgery. It is known that growth of a blood vessel or a cardiac chamber is dependent on the blood flow through it. A common condition proving this theory is hypoplasia of the aortic isthmus and coarctation, which occur as a result of an increase in pulmonary blood flow and decreased aortic flow.^{8,9} This pathologic process, along with other unknown factors, can affect different segments of the aortic arch. According to the definitions of arch hypoplasia of Moulart and associates,¹⁰ there are three types of hypoplasia: Type I, hypoplasia of the distal arch (82%); Type II, hypoplasia of the entire arch (13%); and Type III, absence of the proximal arch (5%). All these pathologies require surgical repair, usually performed in the neonatal period. Conventionally, pathologies of the distal arch and descending aorta are approached through left thoracotomy, whereas more proximal pathology is addressed by midsternotomy. These two approaches bear significant differences in risk, prognosis and outcome. While the former is performed with a beating heart, the latter is carried out with CPB with different perfusion techniques, including deep hypothermic circulatory arrest or selective cerebral perfusion, all with inherent risk of major morbidity or mortality. It is not clear what subtype of patients with HAA is still amenable for the thoracotomy approach. Furthermore, no quantifiable anatomic variables have been defined yet to determine the type of approach.

In this study we determined whether further delineation of the arch anatomy by means of additional modality will affect the decision regarding the approach, possibly shifting more patients toward the lower risk, off - CPB, thoracotomy approach. In addition, we attempted to identify and quantify anatomic factors influencing the type of approach.

In clinical settings, besides feasibility of repair, complete relief of the stenosis in the aorta is of utmost importance. In any approach, the surgery needs to resolve the narrowing, leaving no residual hypoplastic segment as such a narrowing has been found to be associated with early mortality, late mortality and hypertension.^{11-13, 6} In a long-term follow-up study, after conventional repair of the coarctation in patients with moderate hypoplasia of the arch, one third kept a small proximal arch.⁴ The extended end-to-end anastomosis-type repair for hypoplastic distal arch has been criticized as being inadequate when an additional proximal hypoplasia of the arch is seen by echocardiography. In our study, no patient had any significant gradient after the surgery, and no intervention was required in the follow-up period. This satisfactory outcome might be explained by appropriate patient selection for this type of repair using objective measurements of arch segments accurately obtained from CSI. The extended end-to-end anastomosis repair provides a large anastomosis less prone to narrowing should any anastomotic imperfection or tissue growth occur and it has proved to address the hypoplasia of the transverse arch with low early mortality.¹⁴

In this study, CSI was used to accurately define the HAA. While no studies have compared the different imaging modalities in the context of HAA, echocardiography, despite being the only modality in most cases, was found to be less sensitive in diagnosing Isthmic Hypoplasia.¹⁵ In a

study reviewing the multimodality imaging for assessment of congenital heart disease, MRI and CT have been considered superior to echo in imaging of extra-cardiac vasculature.¹⁶

Routinely, most congenital cardiac anomalies are diagnosed solely by echocardiography. It is non-invasive, available and easy to interpret; however, it affords limitations including spatial resolution, lack of appropriate acoustic windows and user dependency in interpretation. Specifically, echocardiography lacks the 3-dimensional orientation of arch branches, which is valuable for surgical planning, including cannulation and perfusion during CPB. The 3-dimensional reconstruction is a realistic view elucidating accurate anatomy as seen in the surgical field. Despite these benefits of CSI, it is used only in a fraction of the patients due its availability, exposure to radiation and contrast material. However, advances in CSI technology with sophisticated image reconstruction and processing have made this modality more accurate and accessible even for the neonates.

In our patients, the complex pathology of the second segment of arch did not exclusively affect the surgical technique. The CSI in this group provided objective measurements and, therefore, changed the management plan. A 3-D arch image provided a real life representation of the aorta where the surgeon could visualize the surgery with all parameters available. The concept of thoracotomy was accepted when the surgeon could virtually plan the surgery using these images to imagine clamp location, incision lines, resection extension and anastomosis lines and location. This imaginary surgery based on 3-D images from CSI would not be possible by 2-D images obtained from echocardiography and, in these complex cases, the decision would be midsternotomy as a default approach rather than a thoracotomy.

This study is limited by the small number of patients and short follow-up. No death and no re-coarctation was recorded in these series of patients and, therefore, we assume it is possibly an optimal approach saving midsternotomy and CPB in some cases. However, it is possible that minor narrowings without clinical significance may develop during this follow-up period, and they would be diagnosed only by follow-up CSI, that would be hard to justify in routine follow-up. It is also not clear whether the long-term results will be as optimal as the short-term results. The satisfactory immediate results in all these patients seems to support this decision making process. We propose this management algorithm as a baseline and an initial paradigm. Larger studies and further experience are needed to optimize it and to prove its validity as a general guideline.

In conclusion, in HAA, whenever the exact anatomy of the arch is not perfectly described by echocardiography, a CSI is recommended for surgical planning. The information obtained by CSI, can provide quantifiable parameters to identify patients who can benefit from a thoracotomy approach rather than a midsternotomy approach.

This study was conducted at Oregon Health and Science University, Portland, Oregon, while the author was working at this institution.

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Familial Giant Right Atrium with Heterogenous Clinical Presentation and Electrocardiographic Features

By Sulafa KM Ali, MD; Noha Karadawi, MD; Sara Bushra Eldomi, MD

Introduction

Idiopathic right atrial dilatation, or Giant Right Atrium (GRA), is a rare congenital anomaly where there is dilation of the RA with severe tricuspid regurgitation, usually associated with arrhythmias.^{1,2}

We describe familial GRA in two patients with variable clinical features and outcomes.

“Idiopathic right atrial dilatation, or Giant Right Atrium (GRA), is a rare congenital anomaly where there is dilation of the RA with severe tricuspid regurgitation, usually associated with arrhythmias.^{1,2}”

Case 1

An 18-month-old boy presented with facial and lower limb swelling for 2 weeks. He was born to consanguineous parents. The mother's first pregnancy ended in an intrauterine fetal death.

Family history revealed a sister who had similar complaints in infancy, diagnosed as a cardiac problem, and advised to have operation, but the family did not accept the treatment. The family claimed that this sister is currently doing well at the age of 8-years old.

A physical examination revealed signs of right-sided heart failure with puffiness of the eyes and lower limb edema. There were no signs of respiratory distress or dysmorphic features. The heart rate was 42 beats/minute; other vital signs were normal. The heart sounds were well-heard with a pansystolic murmur at the tricuspid area. The liver was enlarged, 4 cm below the costal margin, with a span of 8 cm.

The child was brought to the hospital for an echocardiographic study, and was given one dose of chloral hydrate for sedation. However, he developed apnea and decreasing heart rate. Resuscitation (with ambo-bagging, cardiac massage and one dose of adrenalin) was successful, but the heart rate remained 40-50 beats per minute.

Chest X-ray showed cardiomegaly with right ventricle and right atrium enlargement. Electrocardiogram (Figure 1) showed atrioventricular (AV) dissociation (3rd degree AV block) with a ventricular rate of 50 per minute, atrial rate of 100 per minute and right atrial enlargement.

Echocardiogram (echo) showed hugely dilated right atrium measuring 6x5 cm and an area of 19.6 cm² (Figure 2). Tricuspid valve annulus is dilated to 35 mm with lack of coaptation of the leaflets. No signs of Ebstein's Anomaly (EA) were found. There is free tricuspid regurgitation (Figure 3). The patient was started on furosemide and spironolactone and referred for permanent pacemaker insertion.

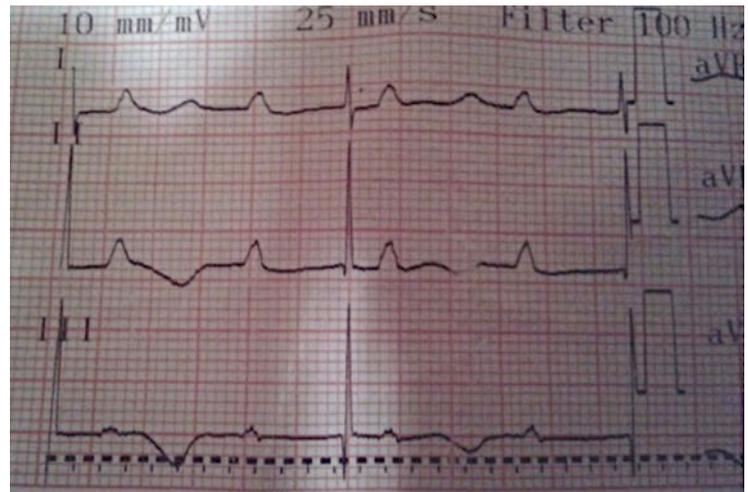


Figure 1. Electrocardiogram of Patient 1 showing atrioventricular block with atrial rate of 100 and ventricular rate of 50/minute. Peaked P-wave indicating right atrial enlargement.

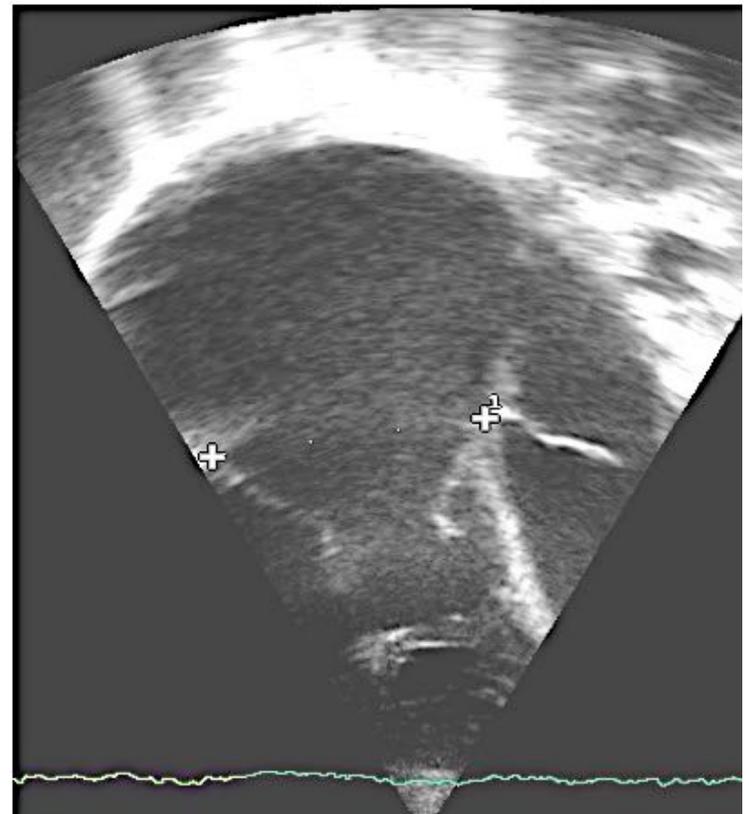
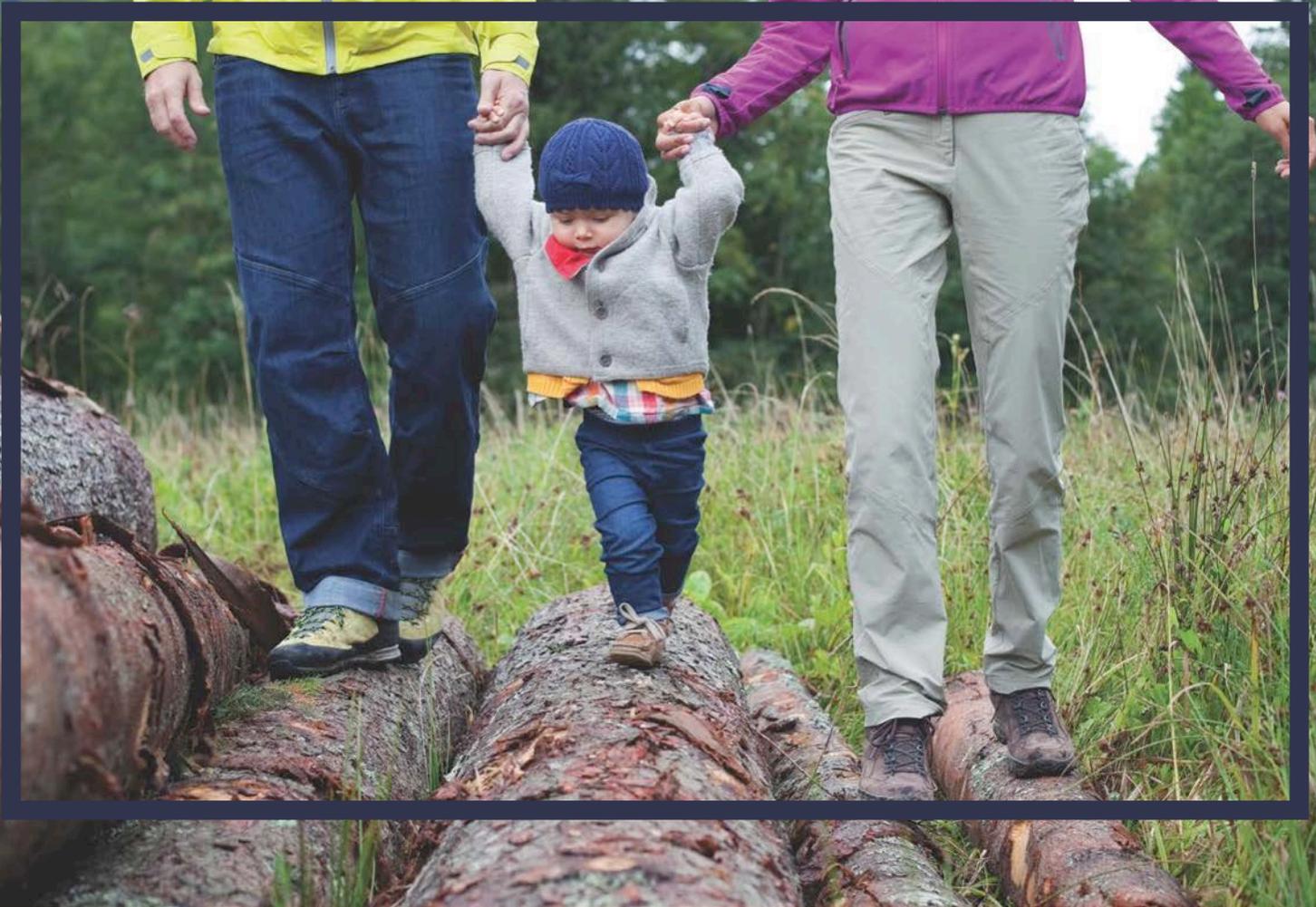


Figure 2. Four-chamber echocardiographic view of Patient 1 showing dilated right atrium and tricuspid valve annulus with lack of coaptation of tricuspid leaflets.

Case 2

A 5-year-old girl presented with cardiogenic shock. There was one-month history of shortness of breathing, and no previous



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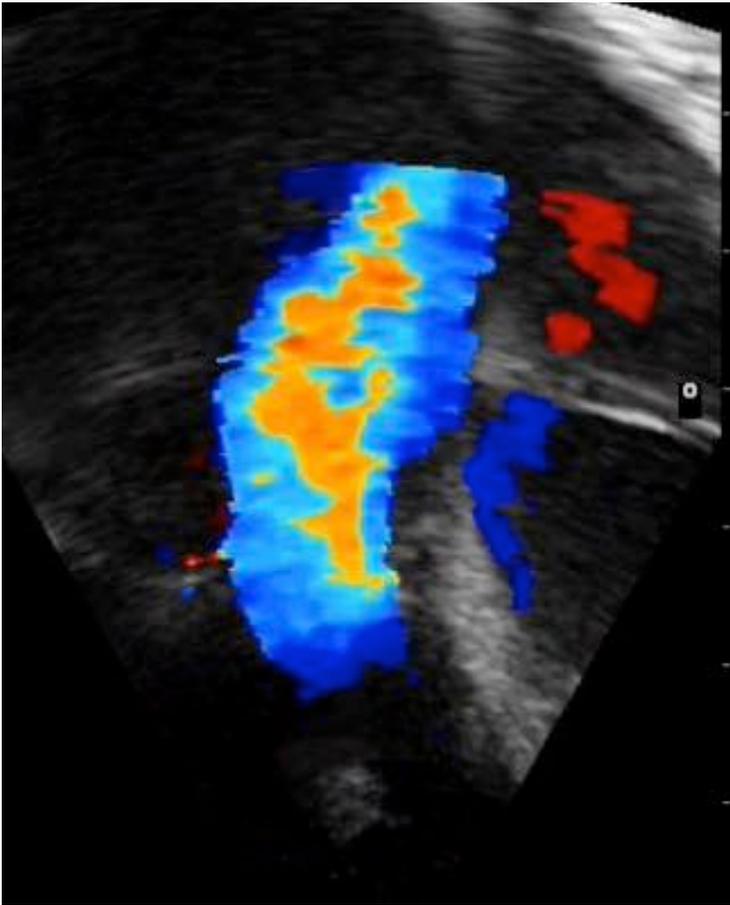


Figure 3. Four-chamber echocardiographic view with color doppler of Patient 1 showing free tricuspid regurgitation.

history of edema. Her twin sister had a similar history, and was admitted to a peripheral hospital where she died 20 days later.

Physical examination revealed a thin-looking child with no dysmorphic features. She had signs of congestive heart failure, with a heart rate of 150/minute, and respiratory rate of 45/minute. Blood pressure was low, and she was started on dopamine and dobutamin infusions and diuretics. Normal heart sounds were heard with a pansystolic murmur at the left lower sternal border.

During her stay, the doctor on call reported bradycardia of 60 beats per minute for which she was shifted again to the Intensive Care Unit (ICU). However, the episode was transient, and there was no documented ECG. A Holter monitor was done which did not reveal any rhythm disturbance. ECG and Holter revealed sinus rhythm with a huge I-wave (Figure 4).

An echo showed a huge right atrium measuring 5.8 by 6 cm with an area of 19 cm² (Figure 5). Tricuspid valve annulus was dilated to 40 mm with lack of coaptation and severe tricuspid regurgitation.

In addition, the left ventricle was dilated (5.3 cm), and the ejection fraction was 34%. She was managed with diuretics, and angiotensin converting enzyme inhibitors, and discharged on these medication with some improvement.

Discussion

These cases represent a rare anomaly of the right atrium often referred to as "Giant Right Atrium," which is characterized by dilated right atrium with variable degrees of tricuspid

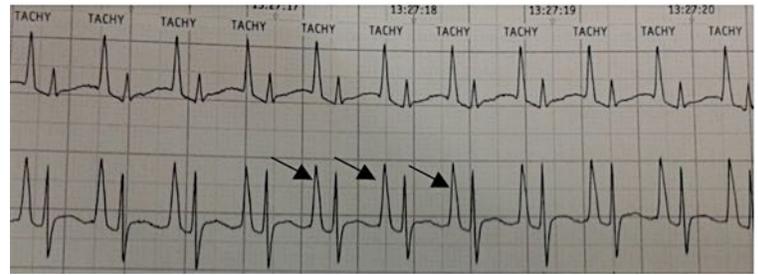


Figure 4. Holter ECG of Patient 2 showing giant P-wave (arrows).



Figure 5. Four-chamber view of Patient 2 showing RA enlargement with lack of coaptation of tricuspid valve leaflets.

regurgitations secondary to dilatation of the tricuspid annulus without features of Ebstein's malformation.¹

The disease has a wide clinical spectrum; some patients can be asymptomatic, while others presented with arrhythmia, palpitations, chest pain, shortness of breath, fatigue, and syncope.²

Familial clusters had been described where heart block was present, similar to our first patient.³

In another report, Stanko et al described two siblings with Giant Right Atrium; one died 1 year after pacemaker implantation, which raises concern about the other arrhythmogenic nature of this disease.⁴

The fact that there are asymptomatic patients may explain the absence of symptoms of our first patients' sister, but sudden death has been described which necessitates considering other modalities of management like intracardiac cardioverter defibrillator. Long-term monitoring by Holter or an implantable electrocardiographic device may be indicated to detect the cause of sudden death.

The second patient had, in addition, features of congestive heart failure and a low ejection fraction. This has been described in one patient before by Stanko⁴ where one sibling had low ejection fraction and the other had normal left ventricle function. Due to

the history of death of the twin of Patient 2, we kept the patient for 4 weeks in the hospital for monitoring; however, we could not document an arrhythmia. The presentation with cardiogenic shock indicates that the cause of death of her twin may be heart failure/cardiogenic shock rather than arrhythmia. This type of patient is a candidate for telemonitoring, which is not available to us. The strikingly tall P-wave in this patient (14 mm) has not, to our knowledge, been reported.

In a follow-up study of 15 pediatric patients, the outcome was variable: 50% were well at an average follow-up period of 6 years, while 25% had cardiac arrhythmias, including atrial arrhythmias. Three children underwent surgical atrial resection: the outcome has been good in these three cases with follow-up periods of up to 18 years.⁵

In conclusion, we described two families with a rare anomaly with guarded prognosis that has variable clinical, electrocardiographic and echocardiographic features. Tele-monitoring of such patients is recommended.

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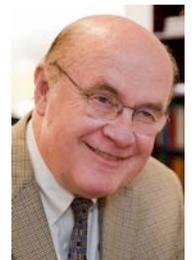
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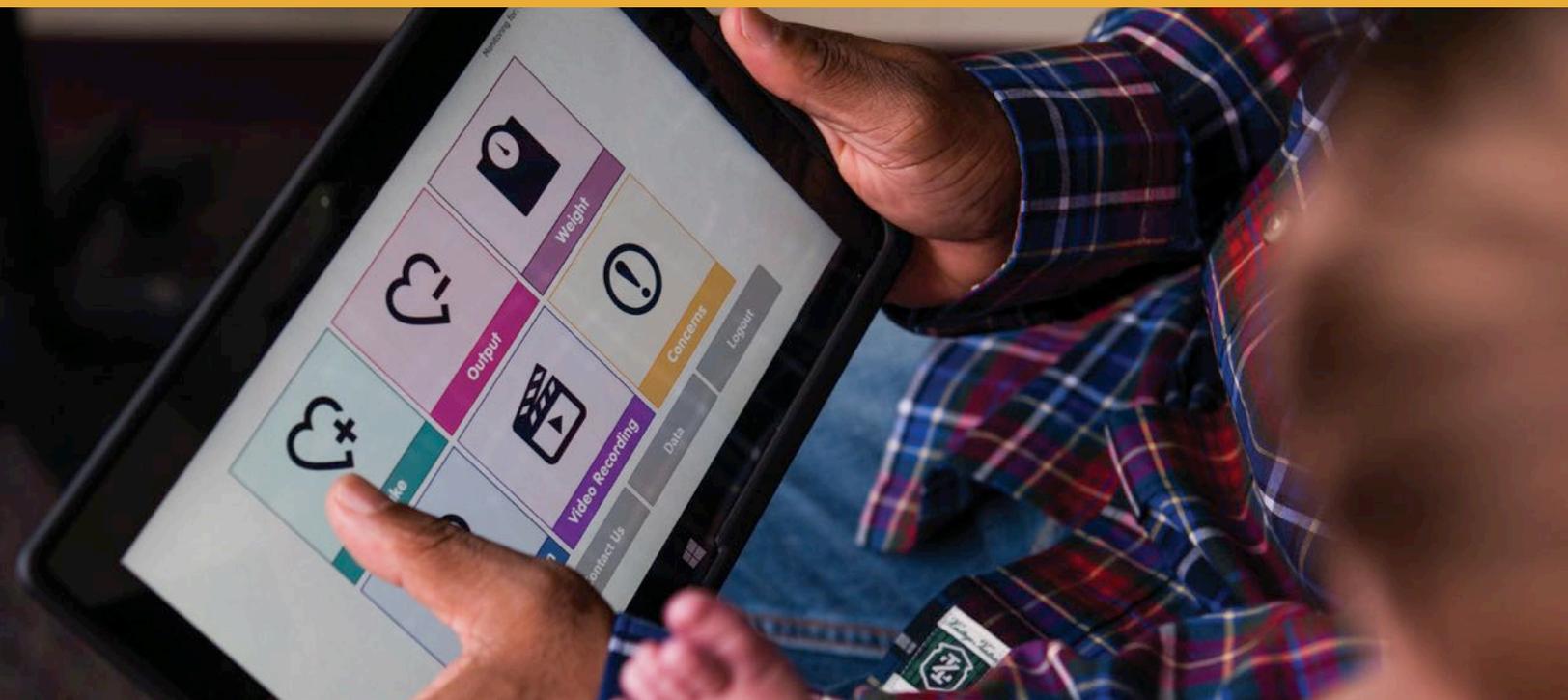
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Fifth Annual UCLA Fetal Echocardiography Symposium: Highlights and Preview of Upcoming Sixth Annual Symposium at UCLA

By Mark S. Sklansky, MD

UCLA's annual practice-based fetal echocardiography symposium, consistently attracting physicians and sonographers from throughout California and across the country, sold out again this past year, with over 200 attendees. The annual *UCLA Fetal Echocardiography Symposium*, held for the fifth year in a row at the Tamkin Auditorium at the Ronald Reagan UCLA Medical Center, last October once again provided attendees with a clinically-oriented, full-day of presentations aimed at improving the prenatal detection and evaluation of Congenital Heart Disease (CHD).

Registrants represented a diverse community of clinicians—obstetric and cardiac sonographers, obstetricians and Maternal-Fetal Medicine subspecialists, pediatric cardiologists, nurses, and trainees at various levels from each of these respective disciplines. Increasingly, this symposium has grown to become nationally recognized for its clinical focus, outstanding speakers, and convenience, with registrants coming from 10 states across the U.S. for a single, day-long symposium.

In addition to the symposium's leadership (Drs. Sklansky, DeVore and Satou), faculty at this past year's symposium included special guests: Dolores Pretorius (Radiology), James Huhta (Perinatal Cardiology), and Tracy Anton (Sonography). The morning began with an overview of guidelines for fetal cardiac imaging (Pretorius), tips on optimizing the image (Anton), and detailed reviews of abnormalities of the four-chamber view and outflow tracts (Huhta, Satou). Dr. Sklansky reviewed the topic of heterotaxy, followed by a live-scanning demonstration by Tracy Anton. Before lunch, presentations and heart-warming testimonials were provided by actual patients and their families, and a short presentation on the importance of prenatal detection was provided, as well, by Adam Chez, co-founder of the Hopeful Hearts Foundation.

The afternoon sessions included a second live-scanning session, focusing on advanced Doppler/3D/4D techniques (Dr. DeVore),

followed by presentations on fetal arrhythmias (Dr. Sklansky) and fetal hydrops/heart failure (Dr. Huhta). Drs. Pretorius and Sklansky participated in an interactive, case-base tutorial led by Tracy Anton, which was followed by a discussion of basic and advanced techniques for the evaluation of fetal cardiac function (Dr. DeVore). The day



Live scanning performed by Dr. Gregory DeVore.



Dr. Mark Sklansky at the state-of-the-art Luskin Conference Center.

“Registrants represented a diverse community of clinicians—obstetric and cardiac sonographers, obstetricians and Maternal-Fetal Medicine subspecialists, pediatric cardiologists, nurses, and trainees at various levels from each of these respective disciplines.”



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Attendees learning tips on prenatal detection of coarctation of the aorta.



Drs. Nancy Halnon and Dianna Drogalis-Kim.

ended with a lively panel discussion of remaining topics of interest to the audience.

Plans are already underway for the *Sixth Annual Fetal Echocardiography Symposium at UCLA: Practical Essentials of Fetal Cardiac Screening*, to be held on Saturday, October 15, 2016. This sixth annual symposium promises to be the best yet, with a renewed focus on screening (reflected in the title), and a relocation--comfortably to accommodate increased demand--to the state-of-the-art Luskin Conference Center on the UCLA campus.

At this upcoming sixth symposium, we are sharpening the focus to provide a full-day dedicated to real-life tips and pearls specifically for those physicians and sonographers who perform or interpret fetal cardiac screening examinations. State-of-the-art presentations will address clinically



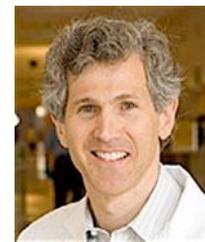
Speakers Drs. Jim Huhta and Dolores Pretorius

relevant topics, such as: what are the current guidelines; how to optimize the image, specific details on how to scan and interpret the four-chamber view/outflow tract views/three-vessel trachea view; how to recognize and interpret major forms of heart disease (including special clues for the detection of Ventricular Septal Defects, aortic/pulmonary stenosis, Transposition of the Great Arteries, Tetralogy of Fallot, Coarctation of the Aorta, Total Anomalous Pulmonary Venous Return, and others), fetal arrhythmias, how to evaluate subtle findings, such as pericardial effusion, left ventricular echogenic foci, and right heart disproportion, and much, much more. The full, final program will be available soon. The entire day will be specifically focused and designed for the practitioner, with outstanding presentations by international experts in the field.

The *Sixth Annual Fetal Echocardiography Symposium* will be among the first to be held at the brand new, state-of-the-art Luskin Conference Center (<http://luskinconferencecenter.ucla.edu>), opening in August, 2016. Located squarely in the middle of the UCLA campus, directly across from the Pauley Pavilion and the John Wooden Center, the Luskin Conference Center will provide comfortable seating in a beautiful and spacious ballroom, a deluxe luncheon and refreshments throughout the day, and a special area for exhibitors. Specially-priced luxurious hotel rooms (on site within the Luskin Conference Center) will be available for those who wish to spend additional time in the heart of one of the most relaxing and beautiful areas of West Los Angeles.

Questions regarding the *Sixth Annual Fetal Echocardiography Symposium at UCLA* may be addressed to Dr. Sklansky at msklansky@mednet.ucla.edu. Registration is already filling up at the following link: <https://www.cme.ucla.edu/courses/event-description?registration%5fid=124261>.

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Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

Congenital Heart Outcomes - The Best Papers of 2015

The following papers and take home points were selected by the *ChiP Network Journal Watch* editorial board including: Drs. Ginnie Abarbanell, Shaji Menon, Gary Webb, Mehul Patel, and Tabitha Moe.

Cardiovascular Deaths in Children: General Overview from the National Center for the Review and Prevention of Child Deaths.

VL Vetter, TM Covington, NP Dugan, DM Haley, M Overpeck, VR Iyer, J Shults. *American Heart Journal* 169; 426, 2015

Take Home Points:

1. This study reports the analysis of the cardiovascular death data set from the National Center for the Review and Prevention of Child Deaths (NCRPCD), which provides a systematic collection of cardiovascular deaths in children.
2. Most cardiovascular deaths occurred in the first year of life and were associated with congenital heart disease.
3. There was a second peak occurring between 10 and 19 years associated with cardiomyopathy, arrhythmia, coronary artery disease, and aortic disease.
4. The study identified ethnic/race disparities in cardiovascular death with more cardiomyopathies and myocarditis deaths in blacks, and more congenital heart deaths in Hispanics.

Long-Term Outcomes in children with Congenital Heart Disease: National Health Interview Survey.

H Razzaghi, M Oster, J Reefhuis - *Journal of Pediatrics* 166; 119-124; 2015

Take Home Points:

1. The odds of reporting worse health and more than 10 days of school/daycare missed in the previous year were 3 times higher for the children with CHD compared with those without CHD.
2. Children aged 2-17 with CHD were more likely than those without CHD to have had a diagnosis of autism spectrum disorder (crude OR, 4.6; 95% CI, 1.9-11.0) or intellectual disability (crude OR, 9.1; 95% CI, 5.4-15.4).
3. The rates of emergency room, home, and doctors' office visits were also significantly higher in the children with CHD.

Trends in Mortality of Congenital Heart Defects.

J Jortveit, N Oyen, E Leirgul, T Fomina, GS Tell, SE Vollset, L Eskedal, G Dohlen, S Birkeland, H Holmstrom - *Congenital Heart Disease* November 2015

Take Home Points:

1. This is a report of the 1 year mortality of live-born children with congenital heart defects in Norway.
2. In Norway the 1-year cumulative mortality proportion was 17.4% for children with severe congenital heart defects and 3.0% for children with non-severe congenital heart defects.
3. These findings from Norway are very similar to other population based studies.

Survival Prospects and Circumstances of Death in Contemporary Adult Congenital Heart Disease Patients Under Follow-Up at a Large Tertiary Centre.

GP Diller, A Kempny, R Alonso-Gonzalez, L Swan, A Uebing, W Li, S Babu-Narayan, SJ Wort, K Dimopoulos, MA Gatzoulis - *Circulation* 132, 2118-2125, 2015

Take Home Points:

1. Isolated "simple" defects have mortality comparable to the matched general population

2. Complex ACHD, Fontan physiology, and Eisenmenger syndrome have higher mortality.
3. With increasing age, the probability of cardiac death decreases, whereas the proportion of patients dying from noncardiac causes such as cancer, cerebrovascular disease, infection, and pneumonia increases. A distinct shift away from perioperative death related to ACHD surgery is evident.

Risk of Congenital Heart Defects in the Offspring of Smoking Mothers: a Population-Based Study.

PM Sullivan, LA Dervan, S Reiger, S Buddhe, SM Schwartz - *Journal of Pediatrics* 166, 978-984, 2015

Take Home Points:

1. This study from Washington State found infants of mothers reporting cigarette use in the first trimester of pregnancy were more likely to be born with a CHD (aOR 1.16 [1.08-1.24]) independent of demographic characteristics and other prenatal risk factors for CHDs.
2. This study also found that an association between maternal smoking and CHDs was stronger with increasing number of daily cigarettes and among older (35+ years) mothers compared with younger mothers.
3. This study concludes that maternal smoking may account for 1.4% of all CHDs.

Population-Based Study of hospital costs for hospitalizations of infants, Children, and Adults with a Congenital Heart Defect, Arkansas 2006 to 2011.

RM Simeone, ME Oster, CA Hobbs, JM Robbins, RT Collins, MA Honein - *Birth Defects Res A Clin Mol Teratol.* 103, 814-820, 2015)

Take Home Points:

1. Using the HCUP (Arkansas) state inpatient database, patients with a CHD diagnosis accounted for 0.4% of all admissions but accounted for almost 2% of the total hospital costs.
2. Infants with CHD accounted for nearly 75% of all CHD hospital costs.
3. Children with CHD accounted for a higher percentage of hospitalizations than adults (2.2% vs. 0.2%).
4. The total cost for CHD hospitalization for children was nearly 5 times greater than the costs of adults with CHD.

Quality of Life of Children with Congenital Heart Diseases: A Multicenter Controlled Cross-Sectional Study.

P Amedro, R Dorka, S Moniotte, S Gillaumont, A Fraisse, B Kreitman, B Borm, H Bertet, C Barrea, C Ovaert, T Sluysmans, G De La Villeon, M Vincenti, M Voisin, P Auquier, MC Picot - *Pediatric Cardiology* 36, 1588-1601, 2015

Take Home Points:

1. The self-reported quality of life for children with CHD was decreased in the following domains: (1) physical well-being with is most likely secondary to limitations from CHD, (2) financial resources and (3) peer/social support. However, in the other 7 domains measured (psychological wellbeing, moods and emotions, self-perceptions, autonomy, parent relations and home life, school environment and social acceptance) there was no difference between normal children and those with CHD.
2. Parents may have a more skewed view of their child's quality of life. The proxy KIDSCREEN-27 was yes lower in 4 of 5 domains compared to controls.

Survival of Children with Hypoplastic Left Heart Syndrome.

C Siffel, T Riehle-Colarusso, ME Oster, A Correa - *Pediatrics* 136, 864-870, 2015

Take Home Points:

1. Infants born between 1992-2005 that underwent surgical palliation for HLHS had a survival rate of 52% within the MACDP birth defects surveillance program.
2. Those infants with HLHS that survived past 1 year of age had about 90% survival rate to age 18 years.
3. Preterm infants with HLHS do poorly compared to term infants with HLHS.

Secundum Atrial Septal Defect is Associated with Reduced Survival in Adult Men.

JM Kuijpers, T van der Bom, AC van Riel, FJ Meijboom, AP van Dijk, PG Pieper, HW Vliegen. WM Waskowsky, T Oomen, AC Zomer, LJ Wagenaar, WF Heesen, JW Roos-Hesselink, AH Zwinderman, BJ Mulder, BJ Bouma - *European Heart Journal* 36, 2079-2086, 2015.

Take Home Points:

1. Men are at higher risk for atrial arrhythmias, conduction disturbances, cerebrovascular thromboembolism, heart failure and have reduced survival compared to women with ostium secundum ASD regardless of the closure status.
2. Although the incidence of PAH is higher in women, mortality due to PAH is higher in men.

The Unnatural History of the Ventricular Septal Defect: Outcome up to 40 Years after Surgical Closure.

ME Menting, JA Cuypers, P Opic, EM Utens, M Witsenburg, AE van den Bosch, RT van Domburg, FJ Meijboom, E Boersma, AJ Bogers, JW Roos-Hesselink - *JACC* 65, 1941-1951, 2015

Take Home Points:

1. Dutch patients who had a VSD repair between 1968 in 1980 were re-examined every 10 years.
2. Survival was 86% at 40 years.
3. Event free survival was 72% at 40 years.
4. These results were only slightly inferior to a reference general Dutch population.

Long-Term Outcome of Mustard/Senning Correction for Transposition of the Great Arteries in Sweden and Denmark.

N Vejstrup, K Sorensen, E Mattsson, U Thilen, P Kvidal, B Johansson, K Iversen, L Sondergaard, M Dellborg, P Eriksson - *Circulation* 132, 633-638, 2015

Take Home Points:

1. Perioperative factors during atrial switch do not influence long-term survival.
2. Implantation of a pacemaker, which was required in 15% over 30 years, was the only factor identified to have an adverse effect on long-term mortality.

Long-Term Risk for Aortic Complications After Aortic Valve Replacement in Patients With Bicuspid Aortic Valve Versus Marfan Syndrome.

S Itagaki, JP Chikwe, YP Chiang, NN Egorova, DH Adams - *JACC* 65, 2363-2369, 2015

Take Home Points:

1. The risk of dissection is 10-fold higher in patients with Marfan syndrome compared to those with bicuspid aortic valve, while

patients with bicuspid aortic valve are only at 20% greater risk of dissection than an age-matched normally structured aortic valve control group.

2. A common algorithm should thus be avoided while managing a spectrum of aortopathies of varying etiologies.
3. Morbidity even after aortic valve replacement in Marfan Syndrome is substantially high with long-term risks for thoracic aortic dissection, new thoracic aortic aneurysms, and need for thoracic aortic surgery compared with patients with bicuspid aortic valves.

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