Secondary Repair of Incompetent Pulmonary Valves After Prior Surgery or Intervention: Patient Selection and Outcomes

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Introduction: Pulmonary valve (PV) regurgitation (PR) is common after intervention for a hypoplastic right ventricular outflow tract (RVOT). Secondary PV repair is an alternative to replacement (PVR) but selection criteria are not established. We sought to elucidate preoperative variables associated with successful PV repair and to compare outcomes between repair and PVR.

Methods: Patients who underwent surgery for secondary PR from 2010-2017 by a single surgeon were studied. Preoperative PV annulus z-score and leaflet tissue lengths were measured by TTE, and PV annulus and leaflet area were measured by MRI. The primary predictor variable was leaflet area indexed to the ideal PV annulus area (iPLA) by MRI. PV repair and PVR groups were compared using univariate analyses (table), multivariable logistic regression, and with a conditional inference tree. Freedom from PV dysfunction and from reintervention were assessed with Kaplan-Meier survival analyses. All measurements and PR grading were performed offline by two authors.

Results: Of 85 patients, 31 (36%) underwent PV repair. By multivariate analysis, longer PV total leaflet length (cm/m^2) (ß=3.00, S.E.=0.77, p=0.001), larger PV z-score (ß=1.01, S.E.=0.32, p=0.001), and larger iPLA (ß=8.58, S.E.=2.63, p<0.001) were associated with repair. Patients with an iPLA of 0.9 or greater had an 83% probability of achieving PV repair (p<0.001), whereas those with an iPLA <0.89 and a PV z-score ≤0.02 had only a 13% chance of repair (p=0.013). At a median of 4 years follow-up freedom from significant PV regurgitation was superior in the PV repair group (log rank p=0.025) (figure), and no patients had moderate or worse PV stenosis.

Conclusions: Secondary repair of the native PV is feasible and effective for a subset of patients with chronic PR following initial intervention for RVOT obstruction, including those who had a TAP at the initial surgery. Patients with an iPLA >0.9 have a high likelihood of achieving PV repair. Patients with an iPLA between 0.7 and 0.9 are borderline for achieving PV repair, and in this group a smaller PV annulus make PV repair less likely. At midterm follow up, PV repair was associated with better PV function than PVR. Long-term evaluation of the function of the repaired PV is needed, including MRI assessments of PR fraction and ventricular size and function.

Antiarrhythmic Treatment Duration In Infants With Supraventricular Tachycardia

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Background: Infantile reentrant SVT typically presents in the first month of life and resolves by 1 year. Antiarrhythmic therapy is often recommended until 6-12 months of age, though data suggests SVT recurrence is rare in treated patients after 3-4 months of age. Treatment duration varies by provider and institution, with recent trend toward shorter treatment courses.

Objective: Assess effect of shorter treatment course (up to 4-6 months of age) on recurrence of infantile SVT.

Methods: Retrospective review of infants with SVT diagnosis at age 0-12 months, Rady Children's Hospital San Diego, 2010-2017. Exclusions: hemodynamically-significant congenital heart disease, automatic tachycardias, flutter, no follow-up data.

Results: 74 infants met criteria. Median age at diagnosis was 6 days (IQR 21 days); 27% presented with fetal tachycardia. Median gestational age at delivery 38 weeks, 30% preterm. Median age at stopping therapy 6.7 months (IQR 4.6-9.8). Therapy was stopped at younger age in patients managed by pediatric electrophysiologist (vs. general pediatric cardiologist): 4.9 vs. 8.6 months ($p=0.03$). 38 patients had treatment stopped by 4-6 months; 24 were treated for 6-12 months. SVT recurrence was similar for these two groups: 13.1% vs. 16.6% ($p=1.0$). Median time to recurrence after stopping medications was similar: 3.3 vs. 5.6 months ($p=0.82$). 54% of patients with recurrence had WPW and 23% had PJRT. Most patients with recurrence required emergency care. None had significant adverse outcome.

Conclusion: In patients with neonatal SVT, stopping medical therapy by 4-6 months of age results in acceptably low recurrence rate, similar to recurrence rates from longer treatment courses.

A Novel Approach in an Adolescent Patient on ECMO with Aortic Root Thrombosis

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Keywords: Myocarditis, VA-ECMO, Aortic Root Thrombus

Introduction: The management of acute decompensated heart failure secondary to myocarditis with mechanical circulatory support can be life-saving, but poses a serious risk for complications including hemorrhage, thrombosis, and end-organ injury. The interplay between cannulation, anticoagulation, and critical care management strategies, as well as the severity of illness is complicated and significantly affects the likelihood of survival without longstanding deficits. We present a challenging case of an adolescent patient with myocarditis and cardiogenic shock who subsequently developed left ventricular (LV) and aortic root thrombi on veno-arterial extracorporeal membrane oxygenation (VA-ECMO).

Case report description: A 60 kg, 15-year-old previously healthy male presented to an outside emergency room with shortness of breath after running a mile at school. He had upper respiratory viral symptoms a few weeks prior to presentation. On initial evaluation, he was tachycardic and tachypneic. His initial work-up was significant for troponin of 15 ng/mL (normal < 0.03) and lactate of 6 mmol/L (normal < 1.9). He received a bolus of intravenous fluids and became hypotensive, bradycardic and hypoxic requiring intubation and a brief episode of cardiopulmonary resuscitation. Upon transfer to our cardiac intensive care unit, an echocardiogram (echo) showed severely diminished LV systolic function [LV ejection fraction (EF) 20%] and an electrocardiogram demonstrated prominent Q waves in anterolateral leads with poor R wave progression. On hospital day (HD) #1, he developed ventricular tachycardia with hemodynamic instability and was placed urgently on VA-ECMO via right femoral cannulation [23 French (Fr) arterial and venous cannulae]. He subsequently had loss of pulsatility on arterial line tracings and an echocardiogram confirmed no aortic valve opening due to poor left ventricular contractility. He pre-emptively underwent balloon atrial septostomy in the catheterization (cath) lab to prevent acute pulmonary hemorrhage. Pre- and post-septostomy left atrial (LA) pressures were 35 and 20 mmHg, respectively, and a repeat echocardiogram demonstrated adequate decompression of the LA and LV chambers. He was managed with complete myocardial rest (e.g., full ECMO flows, minimal inotropic agents) and heparin anticoagulation with goal ACT range of 170-185 seconds. On HD #2, he briefly lost all cardiac electrical activity while on ECMO. An emergent echo showed a large thrombus extending from the LV cavity into the aortic root without obvious proximal coronary thrombosis (figure 1). To prevent thrombus propagation and embolism, ACT goals were increased to 185-200 seconds and medications were initiated to decrease the likelihood of aortic valve opening (e.g., increased systemic afterload with vasopressors, myocardial depression with esmolol and propofol infusions). On HD #3, a 21 Fr Bio-Medicus® (Medtronic) venous cannula was placed anterograde across the atrial septum via the left femoral vein in the cath lab to maximally decompress his left heart and further prevent LV contractility. Additionally, due to possible inadequate delivery of heparin to the proximal aorta, a 5 Fr pigtail catheter was placed retrograde in the ascending aorta via the left femoral artery to allow for direct infusion of heparin to the aortic root. An echo on HD #4 showed a significant decrease in LV thrombus size with continued aortic root thrombosis Due to ongoing left femoral groin bleeding around cannulae, he underwent left femoral surgical exploration and repair of left femoral arteriotomy and venotomy sites. A TEE on HD #5 showed complete resolution of the LV clot with minimal residual aortic root thrombosis (figure 2). After he spontaneously regained LV pulsatility on arterial tracings on HD #5, he was successfully weaned and decannulated from ECMO on epinephrine and milrinone infusions. He was extubated on HD #6 and weaned off all his vasoactive drips by HD #12. He was discharged on HD #19 on oral heart failure therapies along with a LifeVest® (Zoll®) as secondary prophylaxis given persistent LV systolic dysfunction (LV EF 40%). He had no clinically obvious neurological complications or deficits.

Discussion and implications to clinical practice: This case describes our unique strategy in supporting an adolescent patient on VA-ECMO with LV and aortic root thrombi. We used a novel combination of interventional and medical approaches to prevent opening of the aortic valve until resolution of the thrombi. Percutaneous LVADs such as the Impella® (Abiomed®) that partially unload the LV while promoting aortic root blood flow may play a role in preventing LV and/or aortic root thrombi in adolescent acute decompensated heart failure patients on VA-ECMO with loss of cardiac contractility.

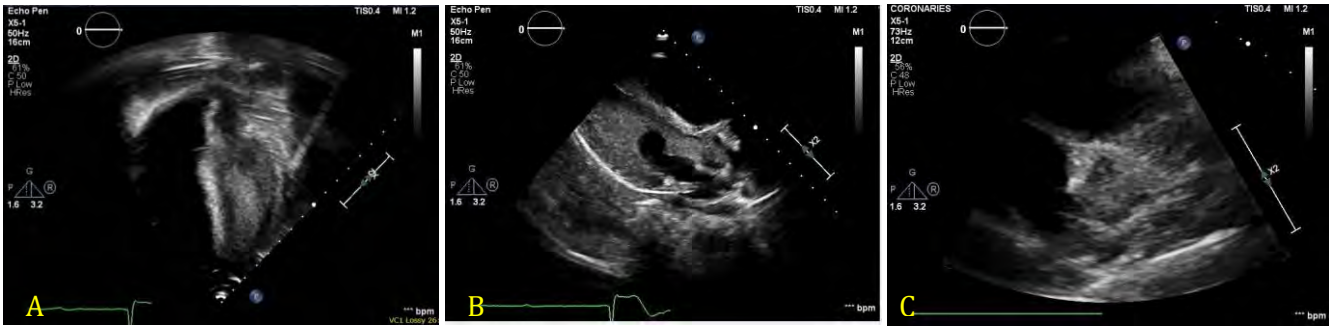

Figure 1. A) TTE apical 4-chamber view demonstrates a large echogenic LV mass consistent with thrombus. B) Parasternal long axis view shows that the LV thrombus is encompassing most of the LV cavity except for a small region of mitral inflow. C) Parasternal long axis view of aortic valve reveals that the thrombus extends across the aorta valve into the aortic root.

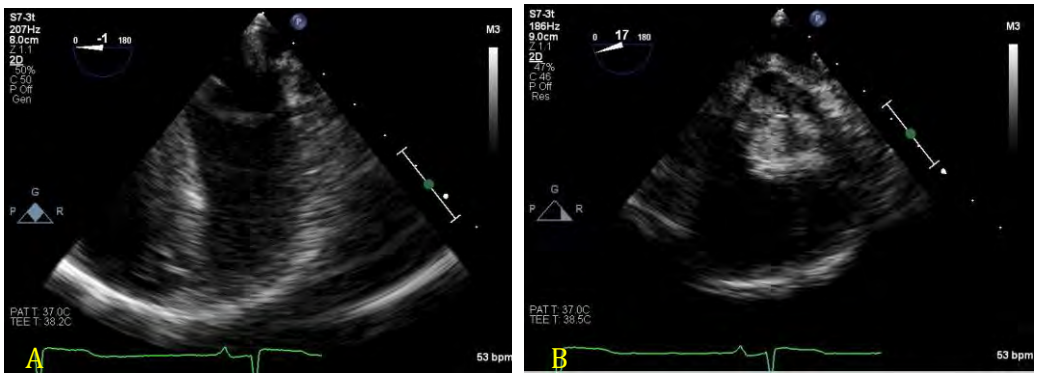

Figure 2. A) TEE demonstrates no obvious thrombus in the LV cavity. B) There is a minimal residual thrombus adherent to the aortic valve within the aortic root.

Antenatal Testing in Mothers Carrying a Fetus with Congenital Heart Disease

Background: Antenatal testing for surveillance of high-risk pregnancies was designed to prevent stillbirth. While used in pregnancies complicated by fetal congenital heart disease (CHD), there are no clear guidelines for type or frequency of testing. Testing, if not guided by pathophysiology or risk for demise may result in unintended consequences such as iatrogenic, premature or operative delivery. We aim to describe the variation in antenatal testing in pregnancies complicated by fetal CHD.

Methods: Retrospective study of maternal/fetus-child dyads with a fetal diagnosis of CHD in 2017 who received perinatal care in the Intermountain or University of Utah healthcare systems. Those with a life-limiting aneuploidy (trisomy 13 or 18) were excluded. The use of antenatal Nonstress tests (NST) and Biophysical profiles (BPP) were described by isolated CHD (Group A), CHD plus additional extracardiac anomaly or syndrome (Group B), and CHD plus hypertension, advanced maternal age or diabetes (Group C). Summary statistics of antenatal testing and associated obstetric interventions was performed, along with sub-group analysis of Group A stratified by CHD risk for fetal demise.

Results: Inclusion criteria was met by 88 dyads (Group A=38, Group B=32 & Group C=18). Antenatal testing frequency and timing varied considerably per pregnancy (Table 1) and was initiated as early as 26 weeks' gestation. Abnormal test results occurred in ~30% and led to maternal admissions for delivery in 19% overall. Average gestational age at delivery in those with any abnormal test was 36.9 +/-2.1 weeks vs. 37.6 +/- 1.8 weeks for those with all normal results (p=0.18). Within Group A, CHD with highest risk of demise (n=7) were tested most (100% vs 69% in lowest risk with NSTs) and average per pregnancy tests (4.7 vs 2.6 in lowest risk) but had no abnormal results. However, none in these group had a BPP. In 2 stillbirths (2.3%) in the cohort, testing was normal.

Table 1:

	All	Group A (n=38)	Group B (n=32)	Group C (n=18)
NST				
No. of women (%)	70/88 (79.5%)	29 (76.3%)	26 (81.2%)	15 (83.3%)
Average no. of tests (SD), range	4.53+/-3.28, 1-16	3.96+/- 2.47, 1-9	5.65+/- 4.14, 1-16	3.67 (2.52), 1-11
Average GA at initial test in weeks (SD), range	33.7+/-2.17, 26-39	34.1+/-2.2, 30-39	32.8+/-2.2, 26-36	34.3+/-1.8, 32-37
Abnormal tests (%)	19/70 (27.1%)	8/29 (28%)	10/26 (38%)	1/15 (7%)
BPP				
N (%)	22 (25%)	9 (23.6%)	11 (34.4%)	2 (0.1%)
Average no. of tests (SD), range	1.81+/-1.14, 1-5	1.78+/- 1.09, 1-4	1.64 +/-0.81, 1-3	3+/- 2.83, 1-5
Average GA at initial test in weeks (SD), range	33.5+/-2.72, 26-37	34.2+/-2.4, 30-37	32.8+/-3, 26-37	34+/-2.8, 32-36
Abnormal tests (%)	8/29 (28%)	1/9 (11%)	4/11 (36%)	0/2 (0%)

Admitted for observation/delivery (%)	13 (18.5%)	3 (14%)	9 (41%)	1 (7%)

Conclusion: Antenatal testing of mothers carrying a fetus with CHD is frequent but highly variable in number and initiation. Abnormal tests often led to maternal admissions and delivery but did not necessarily prevent adverse outcome. Future work should determine association of testing with outcomes to inform standardized antenatal testing practices guided by pathophysiology.

Title: Suboptimal pain control in post-operative Glenn patients leads to increased afterload on the single ventricle.

Authors: Matthew Beaver, MD; Katherine Phelps, MD; Shilpi Garg, MD.

Background: Fundamental changes in physiology following cavopulmonary anastomosis (passive pulmonary blood flow from SVC) typically results in a higher central venous pressure. This is widely believed to lead to the headache of venous distension, resulting in activation of the central sympathetic nervous system. Anecdotally, we have observed the presence of systemic hypertension in patients who undergo a Glenn operation during the immediate post-operative period, and we believe that this is a result of sympathetic stimulation that ultimately leads to increased afterload on the single ventricle.

Hypothesis: Patients undergoing a bidirectional Glenn procedure have higher opioid demands than an aged-matched controls undergoing ventricular septal defect (VSD) patch closure surgery.

Methods: We performed a retrospective chart review of patients between the ages of 0-1yrs at Oregon Health & Science University who underwent bidirectional Glenn procedure from January 2011 to December 2018. Exclusion criteria included: (1) patients >1 year of age at the time of surgery (2) patients unable to be extubated within the initial 12-hour post-operative period and (3) patients who were utilizing opioids during the 8 weeks prior the surgery. We used aged-matched controls undergoing VSD patch closure during that same period. We calculated post-operative opioid requirement in 12-hour increments and also collected subjective pain assessments (FLACC scores recorded by nursing) as a surrogate measure for pain. For uniformity, we used a standardized opioid conversion calculator to express total opioid use in micrograms per kilogram (mcg/kg) of fentanyl. We utilized Welch's non-paired t-test to perform our statistical analysis.

Results: There were a total of 59 patients who did not meet our exclusion criteria (29 Glenn, 30 VSD). 4 out of 29 patients had the bidirectional Glenn operation performed off bypass. 7 patients with the Glenn procedure and 3 patients with the VSD repair were extubated in the operating room. The Glenn patients were extubated after an average of 4.8 hours (ranging from extubation upon arrival to ICU to 11.5 hours) and the VSD patients were extubated after an average of 6.8 hours (ranging from extubation upon arrival to ICU to 11.5 hours). There was no statistically significant difference between the extubation times between the two groups. All patients received scheduled acetaminophen for the initial 48-72 hours with standard dosing. 20 patients with the Glenn procedure and 10 patients with VSD repair also received scheduled ketorolac during the initial 48 hours. When we compared the opioid requirements between the two groups, we found that the total opioid expressed in fentanyl mcg/kg over 12-hour increments was higher in the study group compared to the control group, with statistical significance in the initial 36 hours (Figure 2). We also compared the FLACC pain scores over the same 12-hour incremental periods, but this did not have any statistical significance.

Conclusion: Our results demonstrate that patients with Glenn physiology require higher doses of opioids despite multi-modal pain control in the immediate post-operative period. We believe that key to treatment of systemic hypertension in post-operative Glenn patients is aggressive analgesic regimen and not the use of anti-hypertensive therapies.

ABSTRACT

Title: Risk Factors for Growth Failure in Infants with Single Right Ventricle Physiology during Interstage Period: A Ten Year Review of the Phoenix Children's Interstage monitoring program

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Background:

Infants born with single right ventricle physiology (Hypoplastic Left Heart Syndrome and variants) are extremely fragile and have high rates of morbidity and mortality, especially during their interstage period (defined as period between Stage 1 and Stage 2 palliative surgeries). The National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC) was founded to improve growth of these infants during the interstage period, following discharge from the hospital after Stage 1. Our Interstage Monitoring Program (IMP) has been participating in this collaborative since May 2009. This study describes growth trends of these patients over the last 10 years.

Objectives:

To study trends in somatic growth of infants with HLH physiology enrolled in the IMP from May 2009 to April 2019 and to identify risk factors for growth failure in these infants during their inter-stage period (defined as outpatient management following discharge after Stage 1 surgery until admission for Stage 2 surgery)

Methods:

All newborns born with Single Right ventricle physiology who were discharged home from Phoenix Children's Hospital following Stage 1 palliation during the study period were included in the study. Patients who died, received a heart transplant or were lost to follow up prior to undergoing Stage 2 palliation were excluded. The growth trends of these infants were studied and risk factors for poor growth were identified. Growth failure was defined as failure to achieve target interstage weight of 20-30 g/day (as defined by NPC-QIC). Risk factors were analyzed using standard statistical tests of significance.

Results:

A total of 87 patients were discharged from the hospital after Stage 1 surgery and enrolled in the IMP outpatient program. Fifteen children (17%) were excluded from analysis due to death (n=4), transplant (n=5) or loss to follow up (n=6) prior to Stage 2 palliation. Of the 72 children followed in our IMP, 49 (68%) achieved the target weight gain. The infants with growth failure (n=23, 32%) were found to have significantly longer duration of hospital stay after Stage 1, significantly lower rate of PO feeding prior to discharge and significantly longer interval between Stage 1 and Stage 2 surgeries. There were no significant differences in the gender, ethnicity, birth weight, and gestational age, rate of prenatal diagnosis or type of HLHS between these groups.

Conclusions:

Infants born with single right ventricle physiology are at a high risk for growth failure during their critical inter-stage period when they are being managed at home, outside of the controlled PICU setting. Initiation of partial or full PO feeds prior to hospital discharge may improve the growth rate of these infants during interstage. Overall the interstage monitoring program is successful in helping majority of these fragile infants achieve adequate weight prior to their second stage palliative surgery. Further longitudinal studies are planned to assess the impact of interstage on overall outcomes of these complex group of patients.

Abstract

Targeted Nursing Education to Improve Professional Fulfillment, Wellness, and Quality of Life in a Pediatric Cardiac Unit

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Background

Burnout and distress amongst nursing staff, particularly in intensive care units, is a well-described phenomenon which has been associated with high turnover, medical errors, poor quality of life, and job dissatisfaction. Several factors contribute to the problem, including moral distress, interpersonal team dynamics, fatigue, and unit culture. Despite the gravity of this problem, very few effective interventions have been described to address nursing burnout and wellness. The nature of dedicated cardiac units with complex high-acuity patients that require frequent communication and coordination between multiple disciplines including bedside staff, intensivists, cardiologists, surgeons, neonatologists, and others may place nurses in these units at higher risk of burnout and distress in cases when patients have protracted courses of critical illness or a poor outcome. There have been no benchmark data for nurse burnout and distress in such units.

Aim, Population, and Needs Assessment

Within a 30-bed mixed acuity dedicated pediatric cardiac unit, we aim to assess the levels of nursing distress, burnout, wellness, quality of life, and professional fulfillment before and after the implementation of a targeted nursing education intervention. The unit nursing staff consists of 98 core cardiac intensive care nurses and 23 contracted travel nurses. A focused needs assessment was conducted by unstructured interviews with bedside nurses regarding their feelings of moral distress, difficult team dynamics, communication, and burnout. The nurses who participated in the needs assessment endorsed symptoms of burnout, reported a lack of understanding of the decision making processes for difficult patient cases, and stated a desire for more communication and resources to address these feelings.

Methods

To help address these responses, we created a targeted nursing-led educational series for bedside nurses. The program is modeled after a blend of physician-focused educational conferences and nursing ethics huddles. It uses adult-learning methods and engagement strategies to create an interactive, safe, close-knit environment in monthly hour-long sessions. Each session is semi-structured with some flexibility for nurse-driven discussions to shape the content. An example session outline is described in Table 1. In each session, the lead nurse or nurse practitioner reviews a difficult patient case with a focus on understanding the cardiac anatomy and physiology, reviewing difficult clinical courses or social situations, and facilitating open, clear communication about complexities and difficulties encountered by the interprofessional team. Other team members are invited to participate as appropriate depending on each case, including social work, palliative care, respiratory therapy, and physicians. For example, members of the surgical team could be invited to discuss a particularly difficult surgical course, or the social work team could be invited to weigh in on a case with a complex social needs, etc. These conferences conclude with an open forum in a safe space for the nurses to voice their questions and hopefully gain a better understanding of the difficulty in management and decision making of each patient. Pre- and post- implementation data from anonymous surveys including the Well Being Index, the

Stanford Professional Fulfillment Index, and additional targeted questions over six-month intervals will be compared to assess the impact of this program and inform program improvements.

Conclusion

We describe the development and implementation of a targeted nursing education program to improve professional fulfillment, well-being, and interdisciplinary communication in a mixed acuity pediatric cardiac unit. This unique format is designed to help bedside nurses feel ownership of their patients, have a better understanding of the clinical course, improve communication, decrease moral distress and burnout, and consequently improve nursing quality of life. Benchmark data and interval pre- and post- comparison analysis will be critical for further improvements to this educational program.

Table 1: Example Session Outline

Table 1: Example Session Outline (Case example not based on a real patient)			
Conference Element	Presenter	Need Met	Example
Introduction of patient case, reason patient was selected for case review.	NP/Primary bedside RN	Education, Communication	3 m.o. patient with HLHS and complex social situation and multiple possible avenues for surgical repair.
Review of patient's cardiac anatomy and physiology, especially if complex.	NP/Primary bedside RN	Education	HLHS with MA/AA, retrograde coronary perfusion at risk for reverse coarctation. Surgical repair timeline complicated by emergent duodenal atresia repair shortly after birth. Abdominal surgery was complicated by significant bleeding that continued for the first 24 hours post-op.
Review patient's clinical course, any interventions/surgeries received, and any complications.	NP/Primary bedside RN	Education, Communication	Pt was started on PGE at birth and was maintained on PGE until recovered from duodenal atresia repair. Due to signs of pulmonary over-circulation and hemodynamic instability, the decision was made to palliate her cardiac anatomy with an MPA band and a PDA stent. She had a cardiac arrest the night of her PA band/PDA stent placement from PDA stent occlusion and was emergently placed onto ECMO and then went to cardiac cath to relieve the obstruction. There was concern for a hypoxic ischemic injury to her brain. She was able to de-cannulate from ECMO, but now has had two failed extubations. A head CT is consistent with diffuse hypoxic ischemic encephalopathy.
Thought process behind provider decisions that the bedside nursing team may not be aware of.	NP/Primary bedside RN	Education, Communication, Trust in Provider Team	A Norwood was discussed at length prior to performing a hybrid procedure on the patient. Given her hemodynamic instability and her recent GI surgery, it was thought that the risk of death during or shortly after a Norwood was too great, and the decision was instead made to palliate with a MPA band and PDA stent. There was a great deal of discussion regarding the risk of bleeding due to CPB compared to the need for anticoagulation post PDA stent placement.
Discuss any confounding social aspects of the case.	NP/Primary bedside RN Primary Social	Education, Communication	The patient's mother is a young single mother from Mexico. She is Spanish speaking only and has no support system here in the US. She does have an uncle who lives 2 hours east of the hospital, and it may

	Worker (invited by nursing team)		be a possibility to stay with him after discharge. She does not have a car and is not comfortable driving on highways. This complex social situation played a factor in decision making for this patient (ex: unrealistic ability to care for trach/vent patient, high risk interstage cardiac anatomy, lack of reliable housing and transportation in a high risk patient.)
Additional input from medical and/or surgical team.	Patient's primary intensivist (invited by nursing team)	Education, Communication, Trust in Provider Team	The patient's primary attending shared additional information regarding the patient's code event, including discussing the anticoagulation plan, the cannulation to ECMO and resulting HIE.
Any ethical concerns raised by the case.	Patient's palliative care attending MD (invited by nursing team)	Education, Communication, Trust in Provider Team	The patient's complex medical and social history was made even more complicated due to a post-op complication that led to patient harm. There was question as to whether life-sustaining care should have continued once the team was aware of the degree of HIE, whether the mother fully understood the big picture to make a truly informed decision, and whether decisions being made by the medical team were in the best interest of the patient.
Open forum discussion on the patient's clinical course, social situation, decisions made, and complications.	All	Education, Communication, Trust in Provider Team, Interactivity and Debrief	Bedside nurses expressed gratitude for a greater understanding of the difficulty in managing such a complicated medical and social case. A nurse expressed frustration that she did not feel the anticoagulation plan was adequate following the PDA stent which led to stent occlusion and arrest, but now also understood that the recent history of the abdominal surgery and GI bleed further complicated this. The bedside nurses also gave feedback that the medical team could have been clearer with the patient's mother about her poor prognosis, especially her neurological prognosis. Additionally, knowing that this patient would not have a realistically safe home environment for such a medically fragile child, the question was raised if we should have intervened at all. Overall, the bedside nurses expressed greater comfort with withdrawal of support and compassionate extubation than they did before the conference.
Closing statements, lessons learned from the case review.	All	Education, Communication, Trust in Provider Team	The provider team agreed that in the future, a more upfront discussion regarding neurological prognosis and concern over lack of resources for the family would be of benefit to similar patients. The nurse frustrated about the anticoagulation plan voiced that in the future, she would raise concerns early as to better understand the thought process behind decisions.

The Accuracy of an EKG for Predicting Left Ventricular Hypertrophy: Does It Correlate with Race, BMI, or Systolic Blood Pressure

Megan Burke BA, Edward K. Rhee, MD, and Paul Kang, MPH

Introduction: Left ventricular hypertrophy (LVH) is a disease that manifests as an increase in the mass of the left ventricle, secondary to an increase in wall and/or cavity size. Hypertrophic cardiomyopathy is a genetic condition that causes LVH with an estimated prevalence as high as 1 in 200-500. Due to convince and low cost, the EKG is often used to screen for LVH, yet the sensitivity and specificity of the EKG in diagnosing LVH remain undefined.

Methods: In this study, we investigate the relationship between EKG voltage, specifically the R wave in V5, and how it correlates with systolic blood pressure and BMI while stratifying for race. We analyzed community data from the Anthony Bates Foundation, an organization that does EKG and echocardiogram screenings in the Phoenix, Arizona community. There were 1351 people screened using both EKGs and echocardiograms.

Results: We found that there is not a significant correlation between the R wave in V5 and left ventricular outflow tract diameter ($p = 0.82$) or left ventricular size ($p = 0.22$) as measured on an echocardiogram. There was, however, a significant relationship between the R wave in V5 and septal thickness ($p = 0.0002$), BMI ($p < 0.0001$), and systolic blood pressure ($p = 0.0172$).

Discussion: With these factors in mind, it should be possible to develop a multivariate scoring system that takes into account BMI and systolic blood pressure in the EKG diagnosis of LVH rather than the current criteria based on R-wave amplitude alone. If successful, this scoring system incorporating patient factors and EKG measurements should limit the number of unnecessary confirmatory echocardiograms performed for the evaluation of EKG's with a LVH pattern.

Detection of a Novel RyR2 mutation by Commercial Genetic Testing in an Adolescent with Catecholaminergic Polymorphic Ventricular Tachycardia

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Introduction: Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is an ion channelopathy that affects the ability of the Ryanodine Receptor type-2 (RyR2) to effectively regulate calcium release from the sarcoplasmic reticulum (SR). It is well known that this channelopathy is usually the result of mutations in one of three domains of the RyR2 channel. Here, we diagnose a teenage male patient who experienced aborted sudden cardiac death with CPVT confirmed by stress EKG using commercially available genetic testing.
Methods: Whole blood sampling for DNA was then sent for genetic testing which revealed a single missense mutation (N2177K) in the central domain of the RyR2 protein, one of the mutational hotspots associated with CPVT.
Results: Statistical analysis of the mutation showed that the region containing the mutation is highly conserved across the ryanodine receptor family and across species and both PolyPhen and SIFT indicate that the mutation is likely deleterious. Literature suggests that even a single amino acid change in this region has the potential to disrupt the stability of the closed conformation of the protein, leaving the channel susceptible to calcium leakage, a cause of ventricular tachycardia.
Conclusion: It is our belief that our novel N2177K mutation satisfies the American College of Medical Genetics and Genomics (ACMG) standards for the mutation to be reclassified from a variant of unknown significance to a pathogenic mutation and that a mutation data repository to collate individual mutations detected by commercial genetic testing will facilitate the detection of novel disease causing mutations.

TITLE: ANTEGRADE ABLATION OF LEFT SIDED ACCESSORY PATHWAYS: CAN YOU AVOID A POKE?

Conner TM[1], Patel A[1]

Background: Ablation of left-sided accessory pathways (AP) via a transseptal (TS) approach, either through at patent foramen ovale (PFO) or by TS puncture, has high success. The incidence of a probe-patent PFO (PPFO) and procedural outcomes according to access method have not been well described.

Objective: To determine the incidence of a PPFO undergoing left sided AP ablations and identify procedural outcome differences between a transforaminal (TF) and TS approach.

Methods: A retrospective study (4/2016- 11/2018) of consecutive left-sided pathway ablations was performed. All patients had a pre-procedure echo documenting status of the interatrial septum. The institutional practice is to initially probe the atrial septum for a PPFO under electroanatomic mapping system guidance and if unsuccessful, perform a TS puncture under fluoro.

Results: 74 patients with a mean age of 12.5 ± 3.9 yrs were analyzed. Substrates included: manifest AP with orthodromic reciprocating tachycardia (ORT) (58%), concealed AP with ORT (38%), and manifest only AP (4%). Acute success rate was 99% and recurrence rate was 1% with a median follow-up of 1.3 yrs (IQR: 0.7 – 1.9). A PFO by echo was seen in 7%. A TF approach through a PPFO not seen on echo was achieved in 22%. A comparison between TF approach and TS approach showed no difference in patient, pathway, or ablation characteristics (see Table 1). There was a significantly lower procedural time and radiation exposure in the TF group with 44% of TF cases done without fluoro.

Conclusions: Left sided APs have a PPFO in 22% without echocardiographic evidence. Use of a PPFO can be done safely with reduced or no radiation and shorter procedure times without compromising success.

1 Division of Cardiology, University California, San Francisco, California.

Count: 1457 (Max 1460)

Figure: 500

Preliminary Evaluation of a Deep Learning Approach for Echocardiographic Screening for Rheumatic Heart Disease

Lindsay A. Edwards MD[a], Fei Feng PhD[b], Mehreen Iqbal MD[a], Yong Fu MS[b], Amy Sanyahumbi MD[c], Shiying Hao PhD[a], Doff B. McElhinnney MD[a], X. Bruce Ling PhD[a], Jiajia Luo PhD[b]

[a]Lucile Packard Children's Hospital Heart Center, Stanford University School of Medicine, Palo Alto, CA; [b]University of Michigan-Shanghai Jiao Tong University Joint Institute, Shanghai, China; [c]Lillie Frank Abercrombie Section of Pediatric Cardiology, Texas Children's Hospital, Baylor College of Medicine, Houston, Texas

Introduction:
Echocardiography-based screening for rheumatic heart disease (RHD) in asymptomatic children in resource-limited settings results in early diagnosis, which, in combination with effective secondary prevention, may reduce morbidity and mortality. Physician-led screening, however, is not feasible in much of the developing world. Automated echocardiographic diagnosis via a novel deep learning approach may obviate the need for a physician, enabling more widespread echocardiographic screening, early diagnosis, and improved outcomes. In this study, we hypothesized that a deep learning model is capable of identifying mitral regurgitation (MR) in appropriate views.

Methods:
Echocardiogram clips were labeled by clip for view and presence of MR. Using the computational ability of a Nvidia GTX1080TI graphics processing unit, training dataset clips were used to build two convolutional neural networks to perform the stepwise tasks of classifying the clips 1) by view and 2) by the presence/absence of MR in appropriate color Doppler views. We trained the models using 6,633 clips from 224 echocardiograms. We then evaluated the performance of our models using a test dataset. Results are reported as an F1 score (an F1 score of 1 suggests perfect accuracy of the model).

Results:

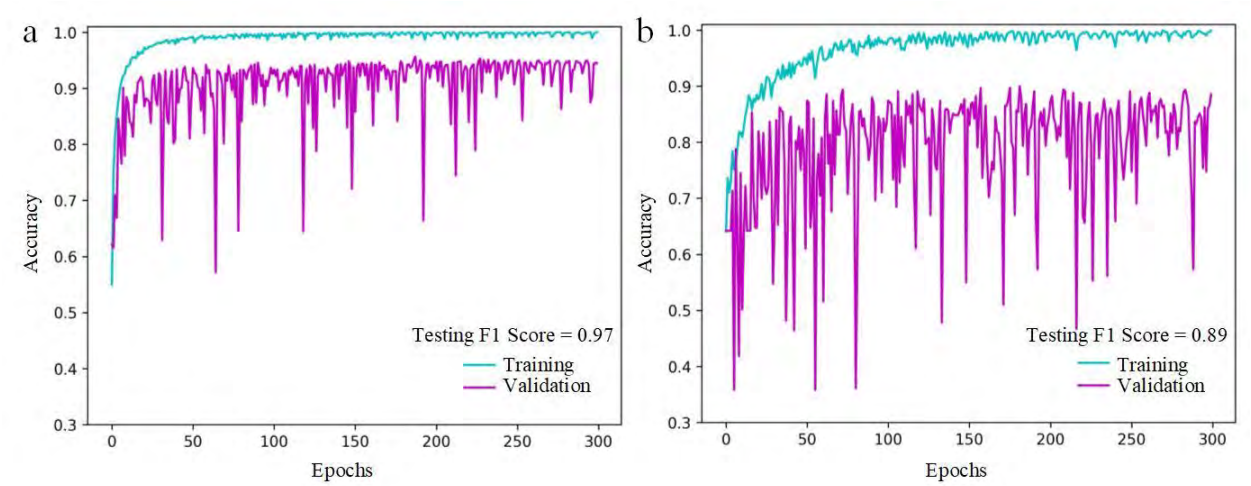

Figure 1. a) View Classification Model Learning Curve, b) MR Detection Model Learning Curve

Conclusions:

Our results suggest that a deep learning model is capable of discerning MR on echocardiogram clips. This is an encouraging step toward automated diagnosis of RHD, which will enable less skilled echocardiogram operators to make a real-time diagnosis of RHD.

The Compensatory Reserve Index Increases After Transcatheter Pulmonary Valve Replacement in Patients with Congenital Heart Disease

Daniel E. Ehrmann, David Leopold, Ryan Phillips, Niti Shahi, Steven Moulton, Kristen Campbell, Michael Ross, Jenny E. Zablah, Gareth Morgan, John S. Kim

Background: The Compensatory Reserve Index (CRI) is an FDA-cleared monitoring technology that trends real-time changes in intravascular volume based on compensatory changes in pulse waveforms obtained via pulse oximetry. In adults, CRI trends progression from normovolemia (CRI = 1) to hemodynamic decompensation (CRI = 0; systolic blood pressure < 80 mmHg with concurrent symptoms). CRI has not been studied before, during, and after procedures for congenital heart disease (CHD). Patients undergoing transcatheter pulmonary valve replacement (TcPVR) may experience improvement to cardiac efficiency and effective circulating volume after restoration of normal valve function. In this study, we hypothesized that CRI would increase after successful TcPVR.

Methods: A prospective cohort of subjects undergoing TcPVR at a single-center was studied. CRI was measured continuously using a CipherOx® CRI M1 device. Data were analyzed during four procedural phases: pre-anesthesia baseline, after anesthesia induction but before right ventricular outflow tract (RVOT) manipulation, immediately after TcPVR placement while still under anesthesia ("early post-valve period"), and during anesthesia recovery ("late post-valve period"). Clinical, catheterization, and imaging data were also collected. Data are presented as medians with interquartile ranges (IQR), means and 95% confidence intervals (CI), or counts and proportions as appropriate. Descriptive data only are presented in this interim analysis halfway through target enrollment.

Results: Thirteen subjects have been enrolled to date. The median age was 13 years (IQR: 12 – 26) and 6 (46%) were status post Tetralogy of Fallot repair. Primary indication for TcPVR was pulmonary stenosis (n = 5, 38%), pulmonary insufficiency (n = 3, 23%), or both (n = 5, 38%). Nine patients had pre-operative MRIs with a median RV:LV ratio of 1.68 (IQR: 1.59 – 2.16). Mean CRI values over the four procedural phases were (figure 1): pre-anesthesia baseline 0.58 (95% CI: 0.51 – 0.65), before RVOT manipulation 0.62 (95% CI: 0.52 – 0.72), early post-valve period 0.75 (95% CI: 0.65 – 0.84), and late post-valve period 0.82 (95% CI: 0.74 – 0.90). The correlation coefficient between change in CRI (late post-valve period – baseline) and MRI RV:LV ratio was -0.77 (figure 2).

Conclusions: In this pilot study of patients with CHD, CRI increased after successful TcPVR. Pre-operative RV:LV ratios by MRI were inversely related to post-TcPVR changes in CRI. Improvements in cardiac efficiency and effective circulating volume after successful TcPVR may be associated with improved compensatory reserve, but immediate benefits may be impacted be the contemporaneous state of ventricular remodeling. CRI should be further studied during other conditions that may alter cardiac efficiency and effective circulating volume in patients with CHD.

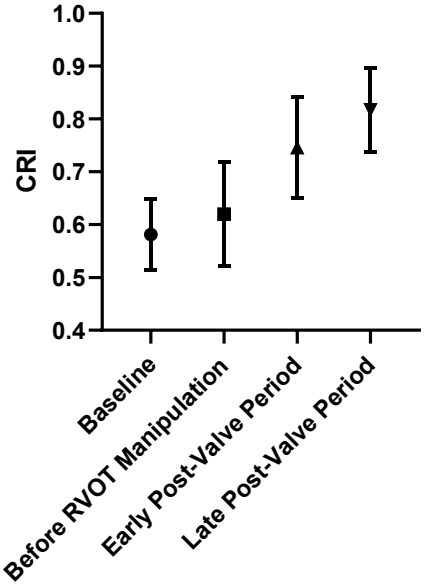

Figure 1. Mean CRI by Procedural Phase

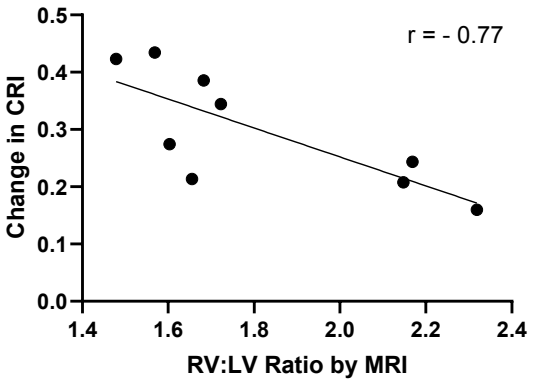

Figure 2. Correlation between CRI change and RV:LV ratio by MRI

A Novel Machine Learning Algorithm Can Non-Invasively Identify Septal Accessory Pathway Location on ECG in Children with Wolff-Parkinson-White Syndrome

Antonio J. Escobar, Jin Long, Marco Perez, Anne M. Dubin, Kara S. Motonaga, Henry Chubb, Anthony Trela, Debra Hanisch, Scott R Ceresnak

Background: Recognition of an accessory pathway (AP) in a septal location can significantly impact the decision to proceed with invasive EPS, especially in younger or asymptomatic patients. Current ECG algorithms are subjective and have poor predictability in children and patients with congenital heart disease (CHD).

Objective: Utilize machine learning techniques to develop an algorithm that accurately and objectively predicts septal AP location on ECG.

Methods: All patients with WPW < 21 years of age who underwent EPS and successful ablation (to verify AP location) at a single large center from 2008 to 2018 were included. Those with no ECGs recorded on the digital ECG system (iECG) were excluded. Data elements exported from iECG (365 total) included: QRS duration, ventricular activation times (VAT), Q/R/S amplitudes, QRS axis, and P/delta wave segments in each of 12 ECG leads. AP were divided into 4 locations: Right septal, right non-septal, left, and multiple APs. Septal APs included anteroseptal, midseptal, and posteroseptal locations. Machine learning techniques, specifically conditional inference tree modeling, was utilized to select predictive variables and develop an automated algorithm to ascertain AP location.

Results: A total of 201 patients were included [mean age 12.5 ± 4.1 years, mean weight 54.3 ± 22.9 kg, 114 (57%) male, 17 (9%) with CHD]. APs were divided into 71 right septal, 39 right non-septal, 79 left, and 12 multiple APs. A combination of 6 ECG variables best predicted AP location: VAT in AVR, R wave duration in V2, frontal plane vector loop in the terminal 40 msec of the QRS, mean frontal plane QRS axis, transverse plane vector loop in the initial 40 msec of the T-wave, and the sagittal plane vector loop. The machine learning algorithm could correctly recognize a right septal location with 97.1% predictive ability with an area under the curve (AUC) 83%.

Conclusion: A machine learning algorithm can non-invasively recognize a right septal AP location on 12-lead ECG in children with WPW. Accurate noninvasive recognition of a septal AP may help with the decision process in timing of EPS, especially in young or asymptomatic children.

Title

Increasing Appropriate Use of Outpatient Echocardiography Among Pediatric Cardiologists: A Quality Improvement Initiative.

Authors

Frandsen EL[1], Kourtidou S[2], Tieder JS[3], Alberda E[4], Soriano BD[1].

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Background

National appropriate use criteria (AUC) guide initial transthoracic echocardiography (TTE) use in outpatient pediatrics. However, barriers exist to implementing the recommendations across a variety of providers and settings. We sought to improve TTE ordering appropriateness among academic pediatric cardiologists with an institutional maintenance of certification quality improvement initiative. Our specific aim was to increase the mean AUC score by 10% above baseline.

Methods

Interventions included teaching TTE AUC recommendations and evaluating baseline TTE appropriateness performance. Initial outpatient TTEs ordered between November 2016 and August 2017 were prospectively categorized by indication and score per the 2014 AUC publication: appropriate (score 7-9), may be appropriate (4-6), rarely appropriate (1-3). TTEs with indications not included in AUC were categorized as unclassifiable. Participants met quarterly to review run charts of TTE ordering patterns and address barriers to implementation (Figure). Baseline and post intervention AUC scores were compared by Student's t-test. Balancing measures included the proportion of abnormal findings before and after intervention, compared by two-proportion z-test.

Results

Twenty-two pediatric cardiologists participated. Of 228 studies evaluated prior to intervention, the mean AUC score was 7.42 (SD 1.87). Seventy-seven percent of studies were appropriate, 5% rarely appropriate, and 5% were unclassifiable. Of 578 post intervention studies, the mean AUC score was 7.16 (SD 2.87). Seventy-four percent were appropriate, 8% rarely appropriate, and 4% were unclassifiable. There was no significant difference in mean baseline and post intervention AUC score ($p=0.105$), nor in percentage of abnormal findings on TTE at baseline (27%, n=48) and post intervention (31%, n=127), $p>0.1$. Barriers identified to implementing AUC in this setting include unclassifiable TTE indications, expectations of referring provider or parent to perform TTE, consistent provider application of AUC, and ability of AUC to capture comprehensive clinical judgment.

Conclusion

TTE appropriateness level before and after intervention was high. Although mean AUC score did not significantly increase after intervention, we identify barriers to implementation of AUC by pediatric cardiologists at an academic children's hospital.

Validation of prenatal aortic arch angle measurements in the diagnosis of neonatal coarctation of the aorta

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Background:

Prenatal prediction of critical coarctation of the aorta (CoA) remains a challenge, while delayed postnatal diagnosis increases morbidity and mortality. Standard methods to identify prenatal CoA have high sensitivity at the expense of significant false positives. We previously derived novel methods of prenatal aortic arch angles for identifying CoA with high sensitivity and specificity. We aim to validate these methods in a new subset of patients and compare them with a model derived in the same era consisting of aortic (Ao) isthmus and ascending aorta (AAo) measures.

Methods:

We performed a retrospective case control study of fetuses with prenatal suspicion for CoA from 1/2014-9/2018. Measurements from first prenatal echocardiogram included: ascending—descending Ao angle (AAo.DAo Figure 1a), transverse—descending Ao angle (TAo.DAo Figure 1b); diameters and z-scores of Ao isthmus from a sagittal (AoI-sag), three-vessel view (AoI-3VV) and AAo. Postnatal CoA was defined as need for prostaglandin at surgery. Previously described multiregression models for AAo.DAo+TAo.DAo vs AAo+AoI-3VV and AoI-sag+AoI-3VV defined post-test probability of CoA, and were compared using ROC curves and positive and negative predictive values (PPV and NPV) with sensitivity/specificity cut-off of 0.5.

Results:

Thirty five fetuses met inclusion criteria; 10 controls with normal pre- and post-natal echocardiogram were included (N=45). CoA was confirmed in 27/35 neonates. ROC curves showed AAo.DAo+TAo.DAo measurement was superior to AAo+AoI-3VV and AoI-sag+AoI-3VV (Figure 2). The PPV and NPV for AAo.DAo+TAo.DAo were 79% and 83. ThePPV and NPV for AAo+AoI-3VV were 86% and 60% and AoI-sag+AoI-3VV were 76% and 85%. A logistic regression model combining all five variables provided a PPV of 100% and a NPV of 85% (adjusted R^2 of 0.6715):

$$Probability\ of\ CoA = \frac{1}{1 + \exp(-F)}$$

$$F = -1.2214 + (AoIsag * 0.0560) + (AoI3VV * -0.0907) + (AAo * -.0905) + (AAo.DAoangle * -0.0158) + (TAo.DAoangle * 0.0140)$$

Conclusion:

We validate the utility of novel angle measures of the Ao arch in predicting CoA. Ao angle measures demonstrate a superior probability model compared to more standard models of predicting CoA. The standard model provides higher PPV, but Ao angle measures provide higher NPV. A combined regression model with Ao angle and standard measures maintains accuracy of identifying CoA while eliminating false positives.

Title: Results From a Multicenter Survey on PTLD in Pediatric OHT patients at PHTS Centers: Should There be a National Consensus on Screening for, Treating and Surveillance of Pediatric Patients With PTLD?

Danielle Harake MD MS [1], Molly Weisert MD [2], Matthew Russell MD [1], Jennifer Su MD [2], Jondavid Menteer MD [2], Juan Alejos, MD [1]

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[2] Children's Hospital Los Angeles, Division of Pediatric Cardiology, Department of Pediatrics

Background: Post-transplant lymphoproliferative disorders (PTLD) are an unfortunate complication that can occur in pediatric orthotopic heart transplant (OHT) patients. In a 16 year multi-institutional study of the 3170 pediatric primary heart transplants between 1993 and 2009 at 35 institutions in the Pediatric Heart Transplant Study (PHTS), there were 151 first malignancy events reported, and of these 147 were PTLD (Chinnock et a, 2012). There are currently no widely-accepted guidelines on screening, treating or post-treatment surveillance for pediatric post-OHT PTLD patients.

Objective: To search for overall trends in North American pediatric transplant centers in identifying, treating, and following pediatric OHT patients who are diagnosed with PTLD, and to determine if further research is needed to come up with treatment and surveillance guidelines for this transplant-associated complication. Our goal was to describe the collective post-OHT PTLD experiences of PHTS institutions.

Methods: All 56 PHTS institutions were asked in March 2019 to complete an anonymous survey sent via REDCap detailing their institutional practices and experiences with regard to their OHT patients with PTLD. The survey aimed to illustrate the collective experience of each center, and did not include any patient- or institution-identifying information.

Results: 23 institutions responded to at least one question in the survey. 14 of 20 respondents reported they give induction therapy to all of their patients prior to transplant and 3 give induction therapy to patients with high PRA levels. The most common induction therapies used by 19 respondents were Rabbit ATG (17) and basiliximab (6). Most of the 19 responding institutions did not routinely screen any particular sub-populations of pediatric OHT patients for PTLD more closely (such as patients with certain pre-transplant diagnoses or lab results), but 10 of 15 respondents indicated that their post-PTLD treatment surveillance lab and visit schedule differs from that of patients who have not had PTLD. Whole blood quantitative EBV PCR was the most common EBV PCR used (15 of 19 respondents), and 13 of 18 respondents indicated the only routine screening lab they use to screen for PTLD is EBV PCR. All but two of 18 respondents reported they do not have lab value cut-offs for a positive diagnosis of PTLD. Amongst 19 respondents, the most common post-discharge indications for obtaining a CT and/or PET scan to look for lymphadenopathy in OHT patients were substantially elevated EBV viremia of any level (9), persistent substantially elevated EBV viremia (8) or only when PTLD was clinically suspected (7).

All 23 participating institutions indicated that PTLD cannot be diagnosed solely based on high EBV load. 12 of the 23 respondents indicated that every pediatric OHT patient with persistent lymphadenopathy and EBV viremia at their institution would have a biopsy before reduction in immune suppression. However, while 19 respondents indicated that a pathologic confirmation of PTLD is not needed prior to reduction in immune suppression to treat EBV viremia, 14 of 18 respondents would require a positive lymph node biopsy to make a positive diagnosis of PTLD.

Of 19 respondents to questions regarding the use of rituximab as a treatment in pediatric OHT patients suspected of having PTLD, most would use rituximab for biopsy-proven PTLD-- either resistant to immune suppression reduction (14), or prior to immune suppression (11). 17 of 22 respondents indicated that when faced with symptomatic EBV viremia they would decrease immune suppression, 8 would treat patients' symptoms, 5 give rituximab, 1 gives steroids, and 1 gives IVIg. 5 of 17 respondents screen for EBV routinely at every or every other clinic visit and 7 of 17 respondents routinely screen more than once a year, but less than every other clinic visit. Timing of clinic intervals were not further specified.16 of 17 respondents always consulted another subspecialty team and 1 center almost always consulted another subspecialty team to assist in treatment following diagnosis of PTLD (Oncology, 17; Infectious Disease, 8).

Conclusion: While limited by sample size and the ability of survey participants to recall details about their institution's practices with regard to PTLD, our survey succeeds in highlighting the variability of screening for, treating and surveying pediatric OHT patients following successful PTLD treatment. This study demonstrates the need for further research to come up with evidence-based guidelines for screening, treatment and surveillance of this patient population. Given the results of this survey and the interest expressed from survey participants, we plan to investigate this area further with a

multi-center study and welcome future collaboration.

References:

A 16-Year Multi-Institutional Study of the Role of Age and EBV Status on PTLD Incidence Among Pediatric Heart Transplant Recipients R. Chinnock et al, 2012). Am J Transplant. 2012 Nov;12(11):3061-8

Feraheme®-enhanced MUSIC CMR delineation of right ventricular dependent coronary circulation in a patient with pulmonary atresia-intact ventricular septum

Abstract:

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare congenital cardiac defect with a broad spectrum of anatomic variability and is often associated with right ventricle (RV) to coronary fistulous connections and right ventricle dependent coronary circulation (RVDCC). While the evaluation of coronary fistulae is performed initially via echocardiography and usually confirmed by cardiac catheterization, due to the associated risks, alternative, non-invasive methods for the evaluation of coronary anatomy in newborns with PAIVS would be very desirable. In the following case study, we describe a patient with PAIVS with RVDCC whose coronary anatomy was accurately delineated via ferumoxytol-enhanced (Feraheme®) MRI with 4D Multiphase Steady State Imaging with Contrast (MUSIC). To our knowledge, this is the first report of cardiac MRI determination of coronary anatomy in a patient with PAIVS with RVDCC with precise cardiac catheterization correlation.

Title: Using Photovoice to Chronicle the Experiences of Pediatric Patients with ICDs and their Caregivers
Authors: Mehreen Iqbal MD, Lauren Schneider PhD, and Anne Dubin, MD

Introduction: Implantable cardioverter-defibrillators (ICDs) play an important role in pediatric patients at risk of sudden death and those that have survived a missed sudden death. While ICDs are life-saving, they have ramifications on patients' behavioral and emotional well-being and have been shown to negatively affect patients' psychosocial functioning. Children and adolescents with ICDs must quickly learn to cope with a difficult diagnosis at a young age, implement lifestyle modifications such as exclusion from competitive sports, undergo lifelong medical surveillance, and live with an omnipresent fear of SCD. These patients have higher anxiety, lower quality of life, higher rate of depression, and posttraumatic stress. Although chronic disease alone can negatively affect psychosocial function, ICDs have been shown to exacerbate this risk. Parents of ICD recipients have a unique experience in caring for children at risk of sudden death, but there is a paucity of literature describing their experiences. Through photovoice, participants use photography and stories about their photographs to identify and represent issues of importance to them. It offers a means of addressing sensitive issues in an unobtrusive way by allowing the participants to direct the researchers' understanding of their concerns and experiences. It especially offers an outlet for under-represented groups to have a way of representing their own lived experiences rather than having their stories interpreted and told by others. Though there is literature documenting the psychosocial implications of ICDs, it lacks the direct voice of the patient and their caregiver. Photovoice is one potential tool to allow the patient and their caregiver to directly illustrate their issues.

Hypothesis: We hypothesize that photovoice will identify novel issues in patients with ICDs that have not previously been identified with traditional metrics and highlight the active voice of the participant.

Objectives:

1. Compare our findings to psychosocial battery of tests (Quality of Life survey, PROMIS Anxiety and Depression scales, PTSD screens for parents and child, Device specific measures: FPAS, FSAS, and SWAP, and Worries about ICDs)
2. Identify new themes that traditional measures do not capture.

Methods: This study uses a mixed-methods approach that involves qualitative and quantitative research. Qualitative data will be provided by Photovoice. The quantitative data will be provided from a battery of psychosocial tests and surveys and demographic information. Patients followed by the Pediatric Cardiology Arrhythmia Service at Lucile Packard Children's Hospital at Stanford will be recruited to participate in the study. Inclusion criteria: 1) All patients with implanted ICDs and one of their parents/legal guardians, 2) Age: >8 years old, <21 years old, and 3) at least 3 months from time of ICD implantation. Exclusion criteria: Developmental delay (<8 years old). Participants will be trained on photovoice methods, including how photographs may be used, ensuring participants' safety and subjects' privacy. Patients and caregivers will be administered the psychosocial battery of tests that will be completed at the time of study recruitment via RedCap. Demographic data will be extracted from the medical records. Participants will be asked to use their camera phones or will be provided with disposable cameras. They will be asked to take pictures that answer one question each week over the course of three weeks: 1) How does an ICD make your/your child's life better? 2) How does an ICD make your/ your child's life harder? 3) How does an ICD change your life? Participants will undergo interviews to provide narratives to go alongside each photograph. Data analysis includes codifying the photographs to identify themes that emerge and quantitative analysis of the psychosocial assessment. The selected photographs and corresponding narratives will be displayed at a community exhibition held at the annual Lucile Packard Children's Hospital ICD Day.

Results/Conclusions: This study is currently in progress. We will present data collection and analysis available at the time of the Western Society of Pediatric Cardiology meeting.

Title: Using Virtual Reality Heart Models to Teach Congenital Heart Disease to Trainees
Authors: Mehreen Iqbal MD, Alaina Kipps MD, and David Axelrod MD

Introduction: Congenital heart disease (CHD) is the most common human birth defect. Amongst CHD lesions, atrial and ventricular septal defects (ASDs and VSDs) are the most common and account for over 30% of congenital heart lesions. Standard two-dimensional models do not adequately convey the spatial information required to understand CHD anatomy. Three-dimensional (3D) models using Virtual Reality (VR) heart models to demonstrate ASDs and VSDs may be useful in improving the understanding of these congenital heart lesions and thus enhance application of knowledge to patient care. We hypothesize that applying an innovative approach to teach CHD anatomy and physiology, using VR heart models to simulate ASDs and VSDs and supplement traditional didactic teaching, is feasible and will benefit the residents' experience of understanding CHD and retention of CHD knowledge.

Objectives:

1. To evaluate the feasibility of using VR to teach common CHD lesions to pediatric residents.
2. To assess residents' knowledge of common CHD lesions following a VR intervention.

Methods: This is a prospective, randomized, controlled, crossover study to teach residents about ASDs and VSDs using VR. Pediatric residents will be assigned to two groups: A and B. Each group will receive one-on-one teaching: a combination of one interventional VR session and one control lecture-based session. For example, Group A will receive VR based teaching for ASDs and lecture for VSDs. VR sessions will receive a lecture during guided use of VR heart models. VR heart models will be accessed using the Oculus Rift ™. Control sessions will receive a didactic lecture supplemented by 2D images. Residents will complete knowledge tests before and after the study, and then at one, three, and six month intervals. Residents will complete demographic surveys indicating year of training, previous cardiology exposure, and Likert style learner satisfaction and self-efficacy questionnaires to evaluate their experience with the VR intervention. All data will be collected in a blinded manner and collected in a secure REDCap database. Analysis of the average percentage correct and average change in percentage correct between the pre-test, post-test, and delayed post-test will be performed across the control and intervention groups to assess the efficacy of VR vs lecture-based education. We will use the two-tailed Student's t-test to assess for statistical difference across groups.

Results/Conclusion: This study is currently in progress, but data thus far demonstrates that the VR group has better test scores but the average change in the percentage correct across test scores is better in the lecture group than the VR group. Thus far, the differences are not statistically significant. Data collection and analysis are ongoing and will be completed in time for presentation at the Western Society of Pediatric Cardiology meeting.

Title: Palliative Care Involvement in Pediatric VAD Patients – A Single Center Experience

Authors: C. Knoll, B. Kaufman, S. Chen, J. Murray, B. Sourkes, H. Cohen, C. Almond, D. Rosenthal, S.A. Hollander.

Background: Outcomes in pediatric patients with ventricular assist devices (VADs) for advanced heart failure (HF) are improving, but the risk of associated morbidity and mortality remains substantial. Few data exist on the involvement of pediatric palliative care (PPC) in this high-risk patient population, while recent literature on critical congenital heart disease and other life-threatening pediatric diseases suggest clear benefit of early PPC involvement. In this study we aimed to characterize the extend of palliative care service's participation in the care of patients requiring VAD placement at our institution.

Methods: A single-center retrospective chart review analyzing all VAD patients at a large pediatric center over a 4 year period was performed and assessed incidence, timing and extend of palliative care subspecialty involvement.

Results: Between January 2014 and December 2017, 55 HF patients underwent VAD implantation at our institution. Palliative care utilization in patient care steadily increased over consecutive years (2014 - <10% of patients, 2015 – 20%, 2016 – 50%, and 2017 – 65%) and occurred in 42% (n=23) of all patients. Of these 57% (n=13) occurred prior to VAD placement while 43 % (n=10) had PCC involvement only after implantation. Patients who died during their VAD implant hospitalization (24%, n=13) were nearly twice as likely to have PPC involvement (62%) as patients undergoing subsequent transplant (38%). Of these, the patients that had PPC involved in their care were more likely to limit resuscitation efforts prior to their death. Out of the entire cohort, only four patients had advanced directives in place prior to VAD implant, of which three had PPC consultation prior to device placement. Only 3 families (5%) refused PPC involvement when offered.

Conclusions: PPC services have been utilized in pediatric VAD patients with increasing frequency at our institution in recent years. Early involvement with advanced heart failure patients, prior to VAD implant, occurred in the majority of patients. While total numbers are small, palliative care support appears to lead to more frequent discussion of goals of care and advanced directives.

Serial Balloon Dilation of Pulmonary Veins Improves Outcomes

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Background:

Recurrent pulmonary vein stenosis (PVS) in children is a progressive disease with poor outcomes. PVS can either be congenital or acquired following congenital heart surgery. Both surgical & catheter-based procedures (balloon dilation or stent placement) can provide good immediate relief of stenosis, but re-stenosis is common, progressive and can be fatal. We sought to describe the outcomes of serial balloon angioplasty for treatment of recurrent PVS at our center.

Methods:

A retrospective chart review of all children, <18 years, with the diagnosis of PVS admitted to Rady Children's Hospital San Diego between 2008 - 2017 was performed. Demographic, clinical and outcome data were collected.

Results:

75 stenotic pulmonary veins in 24 patients underwent an average of 6 (2-22) catheterizations per patient with an average of 5(1-22) balloon dilations per vein. 16 patients had 1-6 catheterizations, 4 had 7-12, and 4 had > 12. Median intervals between procedures (in months) were 2.1 (1.2-4.4) for the first 6 catheterizations, 2.7 (1.3-5.8) for the subsequent 6 catheterizations, and 3.9 (3.3-11.8) for catheterizations greater than 12. 14 (58.3%) patients had PVS post-TAPVC repair, and 10 (41.7%) had primary PVS. 4 patients had 1 affected vein, 5 had 2 affected veins, 4 had 3 affected veins, and 11 had ≥ 4 affected veins [8 out of 11 were post TAPVC repair]. The 6-, 12-, 36-, and 60-month survival rates were 95.8%, 87.5%, 79.2%, and 70.8%, respectively (Fig.1).

Conclusions:

We present improved survival of patients with PVS using serial balloon angioplasty. Further prospective studies comparing the outcomes of different treatment strategies are needed.

Case Report: 3D models can be a game-changer in patients with a cardiac tumor

Authors: Molly Maenchen MD, Jonathan Plasencia PhD, Wayne J. Franklin MD, Andrew Papez MD, Steve Pophal MD

Patient: A 1-year-boy with cough and cardiomegaly on chest x-ray.

Case Description: The patient was brought to the ED by mother after an episode of perioral and extremity cyanosis, fever, and cough. Chest X-ray showed cardiomegaly. Patient was then referred to cardiology. ECG showed LV enlargement with abnormal T-waves. 2D Echo demonstrated a large, left ventricular mass that involved the majority of the LV anterior wall and apex.

Results: Workup with Cardiac MRI showed large, solitary left ventricular homogenous mass with diffuse hyper-enhancement. Conventional imaging did not reveal a decisive surgical option. A 3D model of the patient's cardiac anatomy was created. After a multi-disciplinary discussion, it was determined that tumor could not be safely resected. Close observation for tumor progression or malignant arrhythmia was recommended. The boy's parents are considering heart transplantation but were pleased with the current noninvasive plan.

Discussion: While this patient's presentation is not abnormal for a pediatric cardiac tumor, his course demonstrates how imaging modalities can contribute to the diagnosis and management decisions. 3D imaging and printing provided the care teams with a hands-on model of the patient's anatomy - leading to the decision that the tumor involved too much myocardium to safely resect.

Conclusion: In our patient, 3D modeling was shown to be extremely helpful in anatomic definition and care management. 3D printing should be considered in the evaluation of cardiac tumors and congenital heart disease, due to the inherent heterogeneity of these defects.

EXHIBIT: 3D color model of the patient's heart to be displayed

Predictors of Immunoglobulin therapy resistance in Kawasaki disease

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PURPOSE:

Kawasaki disease (KD) is the most common cause of acquired heart disease in children in developed countries. Timely initiation of treatment with intravenous immunoglobulin (IVIG) has reduced the incidence of coronary artery aneurysms from 25% to 4%. Approximately 10% to 20% of patients with KD are IVIG resistant. Because IVIG-resistant patients are at higher risk for coronary artery aneurysms, it is important to identify these patients who might benefit from more aggressive initial therapy. Currently available IVIG resistance prediction models, originally developed with data from Asian populations, are insufficiently studied on North American patients. The objective is to evaluate existing scoring systems and develop a new model to predict IVIG resistance.

METHODS:

A retrospective cohort study performed between 2004 and 2017 identified 115 patients admitted for classic or incomplete KD and treated with IVIG. The diagnosis of classic KD was based on the presence of fever and at least 4 of the 5 principal clinical features. IVIG resistance was defined as fever for >36 hours after IVIG completion and patients were divided into responders and non-responders. A univariate analysis was performed on the clinical and laboratory data to identify independent predictors of IVIG resistance. The predictors were combined into a new scoring system and compared with existing scoring systems.

RESULTS:

Sixty five patients had classic KD and 50 had incomplete KD. Among the 115 patients, 80 (69.6%) responded and the remaining 35 were resistant (30.4%) to IVIG. Of the 35 resistant patients, 16 (45.7%) patients had incomplete KD, whereas of the 80 responders, 34 (42.5%) patients had incomplete KD. Hispanic children comprised 43% of our sample population. Coronary artery abnormalities developed in 14 of the 35 IVIG resistant patients (39%) and 31 of 80 IVIG responsive patient (39%). Patients with incomplete KD had significantly lower age, WBC count, serum creatinine and AST levels than patients with classic KD. Univariate analysis showed that IVIG resistant patients were older, and present with lower platelets, potassium and creatinine ($p<0.05$). Multivariate logistic regression analysis showed that platelets, potassium, body surface area (BSA) and creatinine were independent predictors of IVIG-resistance (Table 1). The latter were used to devise the Natarajan scoring system, which demonstrated sensitivity of 76.2 % and a specificity of 68.6 %. The Egami, Kobayashi and Sano scoring systems were also tested and their sensitivity and specificity were compared (Figure 1). Our new scoring system has higher specificity than existing scoring systems.

CONCLUSION:

Compared to published data, we observed a higher rate of IVIG resistance and coronary artery abnormalities in our patient population. Natarajan score which incorporated platelets, potassium, BSA and creatinine showed higher specificity and comparable sensitivity to other scoring systems devised to predict IVIG resistance.

Early Differences between the Single Left Ventricle and Single Right Ventricle after Extracardiac Fontan Palliation

Taylor Saley, MD, MPH, Allison C. Hill, MD, Neil Patel, MD, Jennifer Su, MD

Background: In the current era of congenital heart disease management, single right ventricles (SRV) are suspected to incur greater morbidity and mortality, although this has not yet been definitively confirmed. We sought to identify and describe early, potentially preclinical differences in function, rhythm, and hemodynamics between SRV and single left ventricle (SLV) patients after extracardiac Fontan completion that may increase future risk of Fontan failure.

Methods: We examined echocardiographic, catheterization, electrocardiogram (ECG), and Zio® data from patients who underwent extracardiac Fontan between January 2008 and September 2018 and received cardiac care at our institution. We divided the cohort into two groups based on predominant ventricular morphology, and compared functional, electrical, and hemodynamic data between the two groups. We correlated these findings with early Fontan failure (including ventricular dysfunction, decreased cardiac output, protein losing enteropathy, plastic bronchitis) and transplant or mortality.

Results: Of the 87 pediatric Fontan patients meeting inclusion criteria, there were 42 (48%) patients with SLV morphology, 45 (52%) with SRV morphology, and 1 with mixed morphology. The mean age at most recent follow up was 8.2 ± 2.6 years. The two groups did not differ significantly in age, gender, demographic, or previous surgical factors. At this early follow-up period, there were no significant differences in incidence of Fontan failure, heart transplant or mortality between the SRV and SLV groups (29% vs 24%, $p = 0.09$). However, compared to patients with SLV morphology, patients with SRV morphology already had a higher incidence of moderate or greater atrioventricular valve regurgitation (36% vs 10%; $p < 0.01$) and evidence of atrial dilation on echocardiogram (42% vs 10%; $p < 0.01$). Patients with SRV also had a higher resting heart rate (88 ± 23 bpm vs 75 ± 20 bpm; $p = 0.01$), longer QRS duration (100 ± 22ms vs 91 ± 16ms; $p = 0.04$), and voltages consistent with right ventricular hypertrophy (60% vs 36%; $p = 0.03$) on ECG. Furthermore, patients with SRV were more likely to have an arrhythmia detected by either clinically indicated or surveillance Zio® monitor than SLV patients (45% vs 8%, $p < 0.01$).

Conclusions: Early clinical outcomes after Fontan palliation are similar between SLV and SRV patients, suggesting that both single ventricle morphologies are able to compensate sufficiently to support systemic circulation. However, even in the early follow-up period after Fontan completion, significant differences are seen in the SRV compared to SLV population. These include increased resting heart rate, significant atrioventricular valve regurgitation, atrial dilation, increased QRS duration and development of arrhythmias. Longer follow-up of this specific cohort is needed to determine how these early compensatory changes may affect long-term Fontan outcomes.

TITLE: Utility of Surveillance Ambulatory Rhythm Monitoring in the Pediatric Fontan Population

AUTHORS: Taylor Saley, MD MPH, Neil D. Patel, MD, Yaniv Bar-Cohen, MD, Michael J. Silka, MD, Allison C. Hill, MD

BACKGROUND
Advances in staged surgical palliation have improved survival in single ventricle patients, but arrhythmias cause significant long-term morbidity and mortality. The timing of arrhythmia onset is unclear, and many institutions have created surveillance programs including routine ambulatory rhythm monitoring (ARM). However, the impact of rhythm surveillance to detect early occult arrhythmias is unknown.

In 2014, our institution established a Fontan surveillance plan, which included screening ARM at 6, 10, 13 and 19 years old. Additionally, clinically indicated ARMs were performed due to symptoms, prior arrhythmia, or electrocardiogram (ECG) abnormality.

OBJECTIVE
To determine the utility of surveillance ARM in young Fontan patients

METHODS
Patients with Fontan performed 1/2008 – 9/2018 that received routine follow-up at our institution were included. A retrospective review was conducted to collect clinical history, ECG, catheterization, and echocardiographic records. Data from each patient's most recent ARM, including indication, was recorded. ARMs with supraventricular tachycardia (SVT), sinus pauses, sinus bradycardia, junctional rhythm, atrioventricular block, and complex (≥ 2 consecutive beats) ventricular ectopy (VE) were classified as positive for arrhythmia. Arrhythmias were defined as occult if detected on surveillance ARM.

RESULTS
There were a total of 87 patients, of whom 48 had ARMs (46 Zio®, 2 Holter) with a median analyzable time of 4.2 days (0.6 – 14 days). All patients had extracardiac Fontans. The indication for ARM was surveillance in 31 (65%) and clinical in 17 (35%; 8 prior arrhythmia, 3 abnormal ECG, 6 symptoms). Twelve (25%) patients had an arrhythmia on ARM (10 SVT, 6 VE, 5 junctional, 2 sinus bradycardia, 1 Wenckebach with 2:1 conduction). Of those with any arrhythmia, 7/12 (58%) were occult (4 SVT, 3 VE, 2 junctional). Half of the patients with arrhythmias on ARM had multiple arrhythmias. The average age for patients with occult arrhythmias was 8.6 ± 1.9 years. There was no difference in the age of patients with and without arrhythmias detected on ARM (7.5 ± 2.2 vs 8 ± 1.8 years, $p = 0.5$) or the rate of arrhythmias detected on surveillance versus clinically indicated ARM (58% vs 42%, $p = 0.6$). Compared to patients without arrhythmias, patients with arrhythmias were more likely to have an anatomical single right ventricle (83% vs 33%, $p < 0.01$) and a longer QRS duration on most recent ECG (102 ± 14 ms vs 92 ± 19 ms, $p = 0.02$). Arrhythmia detected on ARM resulted in increased surveillance for 1 patient, but no other changes in management were made based on ARM findings.

CONCLUSIONS
Arrhythmias were commonly found on ARMs in young Fontan patients, regardless of whether ARMs were performed for clinical indications or surveillance. These findings emphasize the important role for early surveillance ARMs in a post-Fontan monitoring program.

Title: Does liver biopsy underestimate fibrosis in Fontan associated liver disease? A comparison of liver biopsy pre-combined heart and liver transplant and liver explant post-transplant.

Authors: Sumeet S. Vaikunth, Tami Daugherty, John P. Higgins, Gail E. Wright, Waldo Concepcion, Doff B. McElhinney, and George K. Lui

Introduction: Some degree of liver fibrosis is universal after Fontan palliation, yet the accuracy of liver biopsy for staging fibrosis due to Fontan associated liver disease remains unclear. We compared liver fibrosis on biopsy pre-combined heart and liver transplantation and liver fibrosis on liver explant in patients with failing Fontan physiology.

Methods: Transvenous liver biopsy and whole explants from 11 Fontan patients (ages 16 - 43, median 27 years) who underwent combined heart and liver transplantation were retrospectively reviewed. Hematoxylin and eosin stained slides from formalin-fixed, paraffin-embedded material were examined. Additional special and immunohistochemical stains were also performed. Staging of liver disease due to heart failure was center-specific as follows: stage 0: no fibrosis, stage 1: pericellular fibrosis, stage 2: bridging fibrosis, stage 3: regenerative nodules. There is no stage 4. Biopsy was compared with results of echocardiography, hemodynamics at cardiac catherization, and Model for End-stage Liver Disease eXcluding INR and Varices, Ascites, Splenomegaly, and Thrombocytopenia scores. Descriptive statistics were used to analyze data.

Results: All patients who underwent biopsy prior to transplant had evidence of at least bridging fibrosis. Liver explant showed higher grade fibrosis (Stage 3) than pre-transplant biopsy (Stage

2) in all but three patients; no patient had less fibrosis on explant than biopsy. Ventricular function, atrioventricular valve regurgitation, Fontan and ventricular end diastolic pressures, hepatic vein to hepatic vein wedge pressure gradient, and Model for End-stage Liver Disease eXcluding INR scores varied significantly and did not correlate with degree of fibrosis. Varices, Ascites, Splenomegaly and Thrombocytopenia score was $\geq$ 2 in all but one patient.

Conclusion: We found that liver biopsy consistently underestimated the degree of fibrosis in comparison to liver explant. This underestimation may be due to the patchy pattern of fibrosis seen in Fontan associated liver disease and/or transvenous biopsy technique precluding sampling of the most severely affected subcapsular regions of the liver. Further studies are needed to evaluate the utility of liver biopsy in Fontan associated liver disease.