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WILEY Congenital Heart Disease

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8:30 AM – 8:45 AM

Long-term outcomes of cardiovascular operations in children with Marfan syndrome

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Purpose: Marfan syndrome is a genetic disorder affecting the connective tissue that is frequently associated with cardiovascular abnormalities in need for surgical intervention. The long-term survival of children operated for cardiovascular conditions associated with Marfan syndrome has not been previously reported. The objective of this report is to assess the long-term transplant free survival of this group of patients using a unique linked cohort.

Methods: This is a retrospective cohort study from the Pediatric Cardiac Care Consortium (PCCC), a multi-institutional US-based registry of interventions for pediatric heart diseases. Starting with each patient's first surgery for Marfan's related cardiovascular condition we report the long-term survival, need for heart transplant and causes of death by enriching the PCCC registry data with prospectively collected linkage data with the National Death Index (NDI) and the United Network for Organ Sharing (UNOS) through December 31, 2014. The study included only patient with adequate identifiers up to 4/15/2003, date of stricter HIPAA rules implementation beyond which no direct identifiers are available for linkage purposes.

Results: We identified 87 children (n=61 males) with Marfan syndrome and adequate identifiers for linkage studies that underwent surgery for an associated cardiovascular condition at less than 18 years of age. The first surgery was performed for ascending aorta or aortic valve abnormality (n=64 or 73.6%), mitral valve regurgitation (n=22 or 25.3%) or other valvar lesion (n=1, 1.1%). The first surgery for this cohort was performed at a median age of 11.9 years (IQR = 6.7–15.3) and there was no statistical difference between the timing of the surgical categories reported above [KLK1] (*P* value = 0.002). Of the 87 patients, 6 died in-hospital, 23 died or had a heart transplant (n=2) subsequent to hospital discharge. The median age of discharged deaths was 23.8 years (IQR= 12.8–29.4). The overall 1-,5-,15-year transplant-free survival rate was 98.8, 91.4, and 78.5% respectively, with a median follow-up period of 16.4 years (IQR= 12.6–19.8). Survival did not differ by the type of first cardiac operation (log-rank *P* value =0.4602).

Conclusion: The results of this study suggest that children with Marfan syndrome who require surgical repair for a cardiac condition have significantly shorter survival independent of the type of cardiac condition initially treated. Close monitoring and ongoing evaluation of these patients is important for timely addressing residual or recurrent cardiovascular conditions.

8:45 AM – 9:00 AM

Standardizing the care of outpatient pediatric chest pain: A lesson in utilization and charge reduction

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Background: Variability in medical practice is associated with higher cost without improved outcome. Our aim was to standardize the evaluation and treatment of chest pain in an outpatient cardiology setting and track resultant charge savings.

Methods: A multidisciplinary team developed a care algorithm using best evidence and expert consensus for evaluation of otherwise

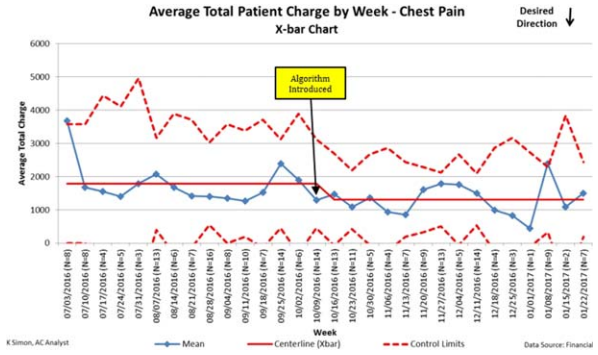

FIGURE 1 Standard deviation total patient charge by week—chest pain

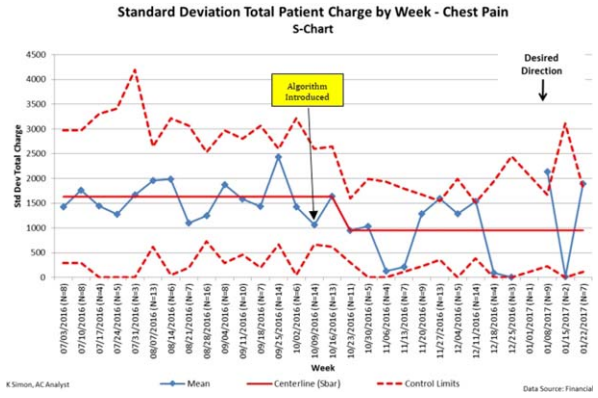

FIGURE 2 S-chart of standard deviation between total patient charge per week, for patients presenting for evaluation of chest pain to an outpatient cardiology clinic, with a decrease in standard deviation after introduction of a chest pain algorithm.

healthy pediatric patients with chest pain. Quality improvement methods guided acceptance and implementation of the algorithm into our cardiology clinics. Primary outcome measure was weekly mean charge per patient, primarily driven by the number of cardiac tests ordered. Total charges were compared using patient billing data. Statistical process control charts evaluated the system over time. Baseline data (7/2016–10/2016) were collected and data were monitored after implementation of the algorithm through 1/2017.

Results: There were 270 patients identified during the baseline period: median age 14.3 years (2–19); 18% were less than 10 years old, 59% female; 81% Caucasian. To date after algorithm implementation, there were 174 patients evaluated: median age 13.8 years (3–18); 17% were less than 10 years old; 56% female; 82% Caucasian. When utilized, 91% of providers followed the proposed algorithm. Following introduction, there was a significant shift in mean charge per patient of 27% from $1,785 to $1,308 (Figure 1), driven primarily by reduction in testing beyond an electrocardiogram. Importantly, there was also a decrease in the standard deviation of charge per patient of 42% from $1,631 to $953 (Figure 2), indicating improved standardization in variable practice patterns.

Conclusion: Standardization of evaluation for pediatric patients presenting with chest pain resulted in a significant charge decrease for our patients. This study highlights the process of standardization, obtaining consensus, and using quality improvement methodology to gain reliable use, leading to a measurable reduction in testing and charge to the patient. This process can be used by clinicians as prototype by which to standardize other similar clinical decisions.

9:00 AM – 9:15 AM

Predicting caval return in single ventricle patients after Fontan from preoperative caval flow: A cardiac magnetic resonance serial study

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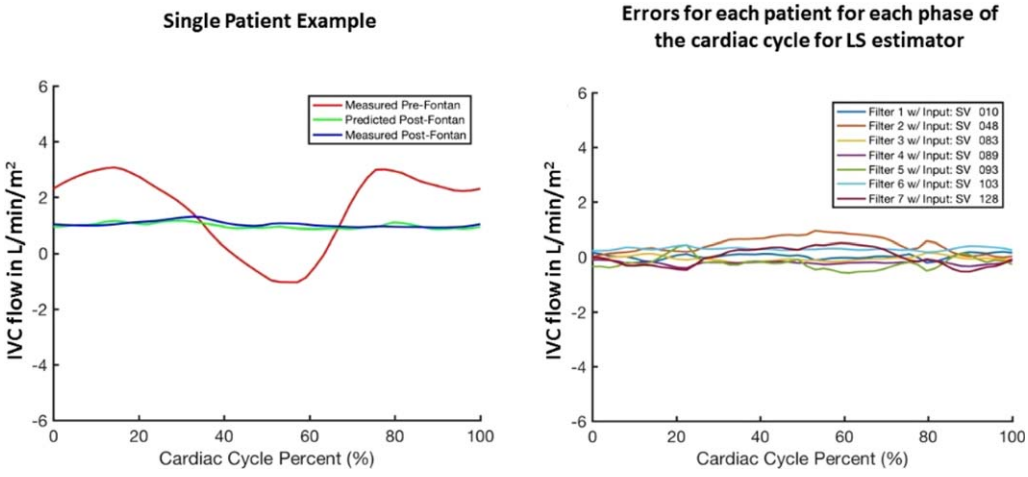

FIGURE 1 IVC flow graphs. A typical patient IVC flow vs phase of the cardiac cycle is on the left. Red line is flow prior to Fontan, blue line is measured flow after Fontan and green line is predicted flow by the estimator—note how close the blue and green lines are. On the right are the errors between measured and predicted IVC flow for all 7 patients vs cardiac cycle graphs; note how all lines crowd around zero.

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Purpose: Single ventricle (SV) patients undergoing staged surgical reconstruction culminating in the Fontan operation undergo major hemodynamic shifts at each stage. In the bidirectional Glenn (BDG) stage, the brain-lung unit are in series with each other but in parallel with flow to the remainder of the body whereas in the Fontan, the pulmonary and systemic circulations are in series with each other, increasing total resistance the heart needs to pump against. It would be useful to predict post-operative hemodynamics from preoperative flows. The purpose of this study is to determine if caval return after Fontan can be predicted from preoperative caval flow

Methods: Seven single ventricle patients were studied immediately prior to and 3–9 months after Fontan with cardiac magnetic resonance (CMR) utilizing phase contrast magnetic resonance (PCMR) across the inferior (IVC) and superior vena cavae (SVC). The flow waveforms per heartbeat were scaled as a % of the cardiac cycle and flows indexed to body surface area. Five transfer functions were developed (using either 3 least squares techniques (LS), empirical transfer function estimators (ETFE), or cross spectrum methodologies) to predict caval flow after surgery from waveforms prior to Fontan. The 5 developed transfer functions underwent "leave-one-out cross-validation" procedure, implemented to validate the initial models because of the small dataset; this separated the data into training and testing subsets.

Results: Table on the left lists the measured flows prior to and after Fontan as well as predicted flows for the 2 best methodologies—LS and cross spectrum (left table). Table on the right lists the errors. For the IVC, the LS and cross spectrum estimators predicted post-operative flows very reliably. A plot of instantaneous IVC flow versus phase of the cardiac cycle for a single patient as an example is shown on the left demonstrating that postoperative measured IVC flow is nearly identical to the predicted flow. A plot of errors for all patients at each phase of the cardiac cycle between measured and predicted flows are on the right; note the lines crowd around zero with little variation. SVC flow, as can be seen from the table, did not change much so estimators did not improve any predictions.

Conclusions: Postoperative Fontan IVC flow can be predicted with a high degree of reliability from preoperative IVC flow using the LS transfer function and to a lesser degree, the cross spectrum estimator. SVC flow after Fontan does not change much from preoperative values and mathematical estimators do not improve predictions. Inferior vena caval flow decreases after Fontan without changes to SVC flow most likely due to systemic to pulmonary collateral flow increasing. This information may be useful for future studies wishing to model post-Fontan hemodynamics using pre-Fontan data.

9:15 AM – 9:30 AM

Perception scores of siblings and parents of children with hypoplastic left heart syndrome

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Purpose: Siblings of children with chronic medical conditions endorse a lower quality of life compared to age-matched peers. Parents may overestimate the negative impact of chronic illness compared to sibling's self-report. Congenital heart disease (CHD) has been shown to significantly affect family life. Hypoplastic left heart syndrome (HLHS) is one of the most severe forms of CHD. To date, there have been no studies specifically addressing the quality of life and functioning of siblings of children with HLHS. This study aimed to assess the effect of HLHS on sibling's quality of life as well as the caregiver's perception of this effect. It also sought to identify factors associated with more negative adjustment in both siblings and caregivers.

Methods: Cross-sectional study using a web-based survey comprised of the Sibling Perception Questionnaire (SPQ), designed to assess

Flows

SVC	L/min/m²	
	Prior to Fontan	After Fontan
Measured	1.70±0.67	1.65±0.74
Predicted LS		1.12±0.69
Predicted Cross Spectrum		1.62±0.73
IVC	**L/min/m²**	
	Prior to Fontan	After Fontan
Measured	1.81±2.61	1.17±0.30
Predicted LS		1.21±0.34
Predicted Cross Spectrum		1.11±0.41

Errors

SVC	L/min/m²			
	RMSE		MAE	
Method	Mean	Std	Mean	Std
1: ETFE (Avg method)	0.60	0.39	0.58	0.40
2: Quotient (Avg. cross spectrum)	0.53	0.32	0.51	0.33
3: Ax=B in temporal	0.84	0.51	0.81	0.50
4: Ax=B in freq	0.84	0.51	0.81	0.50
5: Least squares second method	0.90	0.43	0.87	0.44
IVC	**L/min/m²**			
	RMSE		MAE.	
Method	Mean	Std	Mean	Std
1: ETFE (Avg method)	0.44	0.20	0.41	0.21
2: Quotient (Avg. cross spectrum)	0.39	0.22	0.35	0.22
3: Ax=B in temporal	0.48	0.13	0.42	0.13
4: Ax=B in freq	0.48	0.13	0.42	0.13
5: Least squares second method	0.27	0.15	0.24	0.13

FIGURE 1 Caval return flows and errors. Table on left shows measured and predicted caval return (SVC on top, IVC on bottom). Note how SVC flow does not change prior to and after Fontan. Also note how close IVC flow predictors are to measured flows after Fontan and that IVC flow diminishes after Fontan. The table on the left are errors between predicted and measured caval return. Note low errors for IVC flow and higher errors for SVC flow; estimators of SVC flow do not improve predictions of post-Fontan flows

sibling perceptions of chronic illness, as well as parental perception of sibling adjustment to chronic illness. The Negative Adjustment Composite Scale (NACS) was calculated for each respondent, with higher values representing more negative adjustment. Survey was distributed through various listservs targeted towards parents of children with HLHS. Responses were solicited from both parents and siblings of children with HLHS.

Results: Thirty-five caregivers responded to the survey. Majority of families consisted of 2 caregivers (87%) and 2–3 children (92%). Mean age of caregivers was 38.7 years (SD 6.8). Majority of parent respondents were female (74%), white (86%) and college educated (54%). Thirty-two siblings participated (12 boys, 20 girls). Siblings ranged in age from 7 to 30 years of age (M 12.5, SD 6.3). Their 26 siblings with HLHS ranged in age from 20 months to 27 years old (M 8.3, SD 5.7). Most of the children (73%) with HLHS had undergone the third stage of palliation. 42% of the children with HLHS had other medical issues. Forty-two total caregiver-sibling pairs were examined. Caregiver scores on the NACS were significantly higher than sibling scores, with caregivers tending to report more adjustment problems (M 2.43, SD 0.3) than siblings (M 2.33, SD 0.4, $P < 0.05$). In contrast, siblings were likely to report more negative adjustment than their parents if the child with HLHS had other medical issues. Sibling age was significantly correlated with caregiver NACS (r=0.35, $P < 0.05$), with the score of caregiver NACS increasing as sibling age increased.

Conclusions: Caregivers of children with HLHS perceive their siblings as struggling more than the children self report, except in the setting of additional medical co-morbidities. These data suggest that programs should include support for the entire family to optimize functioning and quality of life.

9:30 AM – 9:45 AM

Aortic stiffness and cardiac function in pediatric and young adult patients with familial hypercholesterolemia assessed by MRI

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Purpose: Familial hypercholesterolemia (FH) is an autosomal dominant disorder that causes elevated levels of cholesterol in affected individuals, resulting in increased risk of atherosclerosis and cardiovascular events. Increased aortic stiffness has been found in adults to be both a marker for atherosclerotic changes and an independent predictor of increased mortality and cardiovascular events; however, studies in pediatric patients are limited. Our study's purpose was to examine the presence of early changes in aortic stiffness and cardiac function using cardiac MRI in pediatric patients with familial hypercholesterolemia compared to healthy, historical controls.

Methods: Patients with familial hypercholesterolemia aged 7 years or greater were recruited for our study. Patients were excluded if they were unable to undergo a cardiac MRI without sedation or had congenital heart disease, diabetes, overt manifestations of atherosclerosis, neurological disorders, or metallic implants. Cardiac MRI was performed on a 1.5 T Philips Ingenia (Amsterdam, The Netherlands). Pulse wave velocity (PWV) was determined using aortic blood flow velocity measurements from the ascending aorta to the proximal descending aorta. Aortic distensibility was measured at the ascending aorta, proximal descending aorta, and diaphragmatic aorta. Ventricular volumes and LV myocardial mass were measured in diastole and systole from bSSFP short axis stack cine images. Cardiac function was determined using the left ventricular volume measurements to calculate ejection

TABLE 1 Characteristics of FH patients

Characteristics	FH patients (n = 12)
Sex, M/F	5/7
Age, years (range)	15.1 ± 3.6 (7.3–19,5)
White Hispanic or Latino/ White Non-Hispanic or Latino	50% (n=6)/50% (n=6)
BMI, kg/m^2	23.4 ± 6.5
Systolic blood pressure, mm Hg	110.1 ± 13.7
Diastolic blood pressure, mm Hg	66.6 ± 5.4
Family History of CAD, %	58.3% (n = 7)
Total Cholesterol Years, mg-year/dL	3968.7 ± 769.1

Where appropriate, data is expressed as mean and standard deviation. FH, familial hypercholesterolemia; M, male; F, female; BMI, body mass index; CAD, coronary artery disease.

TABLE 2 PWV, aortic distensibility, aortic cross-sectional area, and ventricular parameters in FH and control patients

	FH (n = 12)	Control (n = 12)	*P* value
PWV, m/s	4.4 ± 0.7	3.5 ± 0.3	<0.001
Distensibility, 10^{-3}mm Hg^{-1}*			
Ascending aorta	15.3 ± 6.7	8.4 ± 1.6	<0.005
Aortic isthmus	11.4 ± 5.5	6.8 ± 0.9	<0.001
Diaphragmatic aorta	14.6 ± 7.3	8.3 ± 1.1	<0.01
Cross-sectional area, mm^2			
Ascending aorta	501.9 ± 109.7	503.0 ± 100.5	N.S.
Aortic isthmus	293.8 ± 75.7	254.0 ± 53.9	N.S
Diaphragmatic aorta	205.6 ± 50.4	214.8 ± 44.1	N.S.
LVEDV, mL	129.3 ± 38.1	131.2 ± 30.2	N.S.
LVESV, mL	52.8 ± 19.3	47.2 ± 8.9	N.S.
LV myocardial mass, g	79.7 ± 25.4	98.2 ± 33.9	N.S.
RVEDV, mL	129.0 ± 41.0	140.0 ± 29.8	N.S.
RVESV, mL	59.8 ± 24.1	55.7 ± 10.5	N.S.

Data is expressed as mean and standard deviation. *P* values are from the Mann-Whitney U test. * Aortic distensibility values were obtained for 11 patients. PWV, pulse wave velocity; FH, familial hypercholesterolemia; LVEDV, left ventricular end diastolic volume; LVESV, left ventricular end systolic volume; LV, left ventricle; RVEDV, right ventricular end diastolic volume; RVESV, right ventricular end systolic volume.

fraction. Measurements were compared to healthy, historical controls using the Mann-Whitney *U* test.

Results: The study included twelve patients aged 7 to 19 years all on lipid lowering therapy. Demographics are noted in Table 1. Patients with FH had significantly higher PWV than control patients. FH patients also had significantly increased aortic distensibility in all areas measured compared to controls. However, there was no significant difference between groups in aortic cross-sectional areas. Additionally, no significant difference existed in right or left ventricular volumes or left ventricular myocardial mass between groups (Table 2). The cardiac ejection fraction for all FH patients was normal.

Conclusion: Cardiac function was normal in FH patients. Patients with FH under treatment have a concerning early increase in aortic distensibility. This finding could provide us with an early biomarker for screening early plaque formation. The paradoxical increase in PWV in FH patients could be explained by lipid deposition in the aortic wall. Further studies will provide us with the basis of the increase in PWV in these patients.

YOUNG INVESTIGATOR AWARD ABSTRACTS

1:45 PM – 2:00 PM

Effects of an anticoagulation protocol on the incidence of bleeding and thrombosis in postoperative infants in the cardiac intensive care unit

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Background: Infants undergoing surgery for congenital heart disease are at high risk for both bleeding and thrombosis in the postoperative period.

Purpose: To describe the details of and effects of implementing a standardized anticoagulation practice for infants following congenital heart surgery.

anti-Xa \ aPTT	<60	60-85	86-100	101-124	125-150	>150
<0.2	Increase by 20% (4 hours)	Increase by 10% (4 hours)	No change (12 hours)	Decrease by 10% (4 hours)	Decrease by 20% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)
0.21-0.34	Increase by 10% (4 hours)	No change (12 hours)	No change (12 hours)	Decrease by 10% (4 hours)	Decrease by 20% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)
0.35-0.7	No change (12 hours)	No change (12 hours)	No change (12 hours)	Decrease by 10% (4 hours)	Decrease by 20% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)
0.71-0.8	Decrease by 10% (4 hours)	Decrease by 10% (4 hours)	Decrease by 10% (4 hours)	Decrease by 10% (4 hours)	Decrease by 20% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)
0.81-0.9	Decrease by 20% (4 hours)	Decrease by 20% (4 hours)	Decrease by 20% (4 hours)	Decrease by 20% (4 hours)	Decrease by 20% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)
0.91-1	Hold for 1 hour then decrease by 25% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)	Hold for 1 hour then decrease by 25% (4 hours)
>1	Hold for 1 hour then recheck prior to reinitiation	Hold for 1 hour then recheck prior to reinitiation	Hold for 1 hour then recheck prior to reinitiation	Hold for 1 hour then recheck prior to reinitiation	Hold for 1 hour then recheck prior to reinitiation	Hold for 1 hour then recheck prior to reinitiation

FIGURE 1 Therapeutic UFH anticoagulation protocol. A standardized approach to heparin titration incorporating both activated partial thromboplastin time and anti-Xa activity.

Methods: We created a tiered guideline for the postoperative management of bleeding and thrombosis. In patients treated with unfractionated heparin (UFH), upward titration was using anti-Xa level and partial thromboplastin time (aPTT) was used for dose limitation. Clinical outcomes, including bleeding and thrombosis events, were prospectively collected for 5 months prior to and following protocol implementation and adjudicated as either minor, clinically relevant nonmajor, or major.

Results: Among 792 surgical patients followed during the study period, a total of 129 infants (58 preimplementation, 71 postimplementation, total 2,384 patient-days, PD) were treated with therapeutic UFH. Of these, 44% were neonates. Following protocol implementation, we observed a decrease in the incidence of major thrombosis (0.91/100 PD vs 0.31/100 PD for the pre- vs. postimplementation periods, RR 0.34, $P = 0.07$). Specifically, older infants (29 days −1 year) had lower rates of both major bleeding and thrombosis following protocol implementation.

Conclusions: The use of a standardized anticoagulation protocol is safe and may reduce the incidence of bleeding and thrombosis events in infants following cardiac surgery.

2:00 PM – 2:15 PM

Maturational patterns of right ventricular function differ in preterm infants compared to term infants over the first year of age

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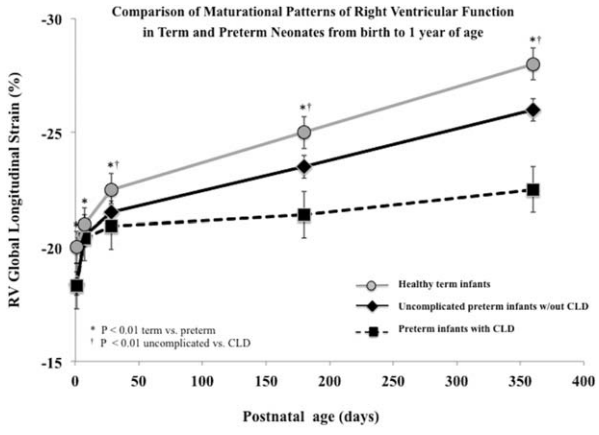
Background: Two-dimensional speckle tracking echocardiography (STE) derived strain imaging provides a more comprehensive assessment of right ventricle (RV) performance in neonates that could not be previously obtained with conventional imaging. We aimed to compare the maturational changes in RV strain mechanics between extreme preterm and term neonates from birth to one year of age using STE.

Methods: In a prospective multicenter study of 167 neonates (117 extreme preterm infants born < 29 weeks gestation and 50 term infants), RV global, free wall, and regional strain values at the base, midventricular, and apical levels of the free wall were measured at multiple time points in the first year of age: first week, one month, six months, and one year of age. Premature infants who developed chronic lung disease (CLD), defined by the need for any respiratory support at 36 weeks postmenstrual age (PMA), were analyzed separately. Data analysis included adjustment for gestational age at birth, gender, and heart rate.



Results: In all neonates, the magnitude of RV global, free wall, and regional strain measures increased from birth to one year of age ($P < 0.01$ for all measure), (Figure 1). A significant base-to-apex (highest to lowest) regional gradient ($P < 0.01$) exists in the RV free wall of all neonates. Preterm infants have decreased RV global, free wall, and regional strain compared to term infants at all time points. In preterm infants with BPD (n=67, 59%), RV global, free wall, and regional strain were significantly lower at 32 weeks PMA that persisted to one year of age when compared to uncomplicated preterm infants ($P < 0.01$ for all measures).

Conclusions: Maturational patterns of global and regional RV deformation generally increase from birth to one year of age in neonates, but track distinctly lower in all preterm infants. CLD appears to leave a further negative impact on RV mechanics.

2:15 PM – 2:30 PM

Association between furosemide exposure and patent ductus arteriosus in hospitalized very low birth weight infants

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Background: Furosemide is commonly used in hospitalized very low birth weight (VLBW) infants, but its safety profile in this population is incompletely characterized. While furosemide may improve pulmonary outcomes by reducing edema and improving compliance, stimulation of renal prostaglandin production may inhibit ductal closure, causing significant morbidity. We studied the association between furosemide exposure and occurrence of a hemodynamically significant PDA (hsPDA) in a large contemporary cohort of hospitalized VLBW infants.

Methods: Using the Pediatrix Medical Group Clinical Data Warehouse, we identified inborn VLBW infants < 37 weeks gestation discharged from the NICU between 1997 and 2015. We excluded infants exposed to prostaglandins, diagnosed with structural congenital heart disease other than isolated PDA, diagnosed with hsPDA on day of life 0 or 1, or who died or were discharged before day of life 7. We defined hsPDA as any medical (ibuprofen or indomethacin) or surgical PDA therapy from day of life 2 to 60. We collected data up to the day of hsPDA diagnosis or 18 days for infants not diagnosed with hsPDA. This cut-off was chosen because ≥90% of hsPDAs were diagnosed before that day. We performed multivariable logistic regression to evaluate the adjusted odds of hsPDA in infants exposed to furosemide prior to diagnosis, controlling for gestational age, SGA status, sex, prenatal steroids, APGAR score at 5

TABLE 1 Characteristics of infants with and without hemodynamically significant PDA (hsPDA)

	hsPDA N=23,508 (%)	No hsPDA N=101,720 (%)	P
Birth weight (g)			<0.01
<501	811 (3)	1309 (1)	
501–750	7132 (30)	10,768 (11)	
751–1000	7491 (32)	19,758 (19)	
1001–1499	8074 (34)	69,885 (69)	
Gestational age (weeks)			<0.01
22–24	1484 (6)	1532 (2)	
25–26	10,619 (45)	16,173 (16)	
27–29	8856 (38)	38,356 (38)	
30–32	2395 (10)	36,291 (36)	
33–36	154 (1)	9318 (9)	
APGAR score at 5 minutes			<0.01
0–3	1245 (5)	2687 (3)	
4–6	5027 (22)	12,628 (13)	
7–10	16,877 (73)	84,843 (85)	
Male	12,016 (51)	50,156 (49)	<0.01
SGA	3743 (16)	28,022 (28)	<0.01
Antenatal steroids	18,971 (81)	82,668 (81)	0.04
Exposed to mechanical ventil	20,413 (87)	54,029 (53)	<0.01
Exposed to inotropes	7140 (30)	14,331 (14)	<0.01
Exposed to other diuretics	185 (1)	805 (1)	0.95
Discharge year			<0.01
1997–2002	3828 (16)	15,630 (15)	
2003–2008	10,249 (44)	34,548 (34)	
2009–2015	9431 (40)	51,542 (51)	

minutes, discharge year, exposure to inotropes, mechanical ventilation, other diuretics, and random effects for site. We used Stata 14.1 (Stata Corps, College Station, TX) and considered a $P < 0.05$ statistically significant. The study was approved by the Duke Institutional Review Board.

Results: Our cohort included 125,228 infants from 337 NICUs, of which 23,508 (19%) were diagnosed with hsPDA. Median (25^{th}–75^{th} percentile) birth weight and gestational age were 1121 g (860, 1330) and 29 weeks (27, 31). Infants with hsPDA were more premature and more likely to be exposed to mechanical ventilation and inotropes (Table 1). Furosemide was prescribed to 13,468 (11%) of infants, for a median duration of 2 days (1, 3). Compared to infants without hsPDA, infants with hsPDA were more likely to be exposed to furosemide (10,843/101,720 (11.2%) vs. 2625/23,508 (10.7%), $P = 0.02$). In multivariable regression, exposure to furosemide was associated with decreased odds of hsPDA (odds ratio (OR) = 0.48, 95% confidence interval (CI) [0.46, 0.51]). In a separate regression adjusted for the same covariates, increasing percentage days of hospitalization with exposure to furosemide was associated with decreasing odds of hsPDA (OR=0.88, 95% CI [0.87, 0.90]).

Conclusions: Furosemide exposure was associated with decreased odds of hsPDA in hospitalized VLBW infants. Decreased hypoxia resulting from improved pulmonary compliance may explain this association, but further studies are needed to characterize the efficacy and safety of furosemide in premature infants.

2:30 PM – 2:45 PM

Clinical impact of a novel patch ambulatory rhythm monitor in children

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Background: Traditional ambulatory rhythm monitors, such as Holter and event monitors, have limitations in children to include being cumbersome, being limited to 24–48 hours of monitoring, or requiring coordination by the patient to record symptomatic events. The Zio Patch (iRhythm Technologies, San Francisco, CA) is a small, adhesive single-channel continuous rhythm monitor that allows for patient triggering of symptomatic periods and can be worn up to two weeks. As relatively novel technology, there is little clinical data on the use of the Zio in children.

TABLE 1

	Total		Class A/B		Class C/D	
	Count	%	Count	%	Count	%
Gender N (%)						
Female	203	54.4%	19	55.9%	184	54.3%
Male	170	45.6%	15	44.1%	155	45.7%
Race N (%)						
Asian	6	1.6%	1	2.9%	5	1.5%
Black	49	13.1%	5	14.7%	44	13.0%
White	159	42.6%	15	44.1%	144	42.5%
Other	87	23.3%	9	26.5%	78	23.0%
Unknown	72	19.3%	4	11.8%	68	20.1%
Age N (%)						
0–9 years	132	35.4%	5	14.7%	127	37.5%
10–18 years	241	64.6%	29	85.3%	212	62.5%
Prior rhythm monitoring N (%)						
Prior monitoring	132	35.454	13	38.2%	119	35.1%
No prior monitoring	241	64.6%	*21*	*61.8%*	220	64.9%
EKG finding N (%)						
Abnormal EKG	91	24.4%	*10*	29.4%	81	23.9%
Normal EKG	266	*713%*	*22*	64.7%	244	72.0%
No EKG	16	4.3%	2	5.9%	14	4.1%
Structural heart disease N (%)						
Heart disease	106	28.4%	8	23.5%	98	28.9%
No heart disease	267	71.6%	*26*	76.5%	241	71.1%
Prior cardiac intervention N (%)						
Prior intervention	63	16.9%	*6*	17.6%	57	16.8%
No prior intervention	310	83.1%	*28*	82.4%	282	83.2%

Demographics and results by finding classifications, A–D. Class A, rhythm disorders requiring new intervention or increased medical management (e.g., SVT requiring ablation). Class B, rhythm disorders requiring increased surveillance (e.g., new sinus node dysfunction). Class C, rhythm disorders requiring no management change, or symptoms corresponding with normal variant rhythm (e.g., tachypalpitations corresponding to sinus tachycardia). Class D, asymptomatic with normal variant rhythm.

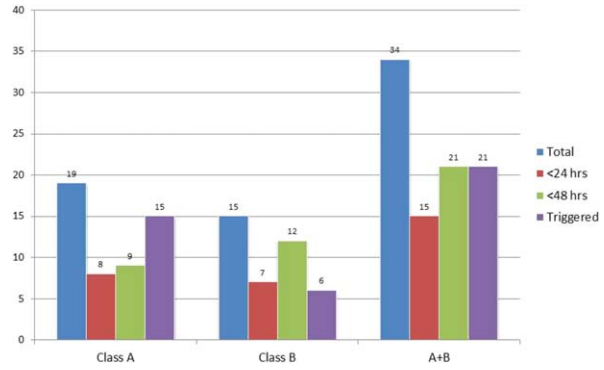

FIGURE 1 Number of studies by classifications A, B, and A+B. Total number of studies is shown in blue. Red and green bars indicate findings that were present within 24 and 48 hours, respectively (the duration of a standard Holter monitor). Purple indicates findings that were patient-triggered on the Zio monitor.

Objective: To determine how Zio ambulatory rhythm monitoring impacts clinical decision-making within a general pediatric cardiology practice.

Methods: A retrospective cross-sectional study was performed using the iRhythm Zio reporting system and the electronic health record. All 0–18-year-old patients who underwent Zio monitoring between January 2013 and June 2016 were included. The following were recorded: patient demographics, history of acquired or congenital cardiac disease, history of cardiac procedures, indication for Zio placement, duration of wear, and all rhythm findings. Clinical outcome, patient disposition and complications were recorded, and notation was made if the monitor led to a new diagnosis or increased intervention using a findings classification system: Class A, rhythm disorders requiring new intervention or increased medical management; Class B, rhythm disorders requiring increased surveillance; Class C, rhythm disorders requiring no management change, or symptomatic with normal variant rhythm; and Class D, asymptomatic with normal variant rhythm.

Results: A total of 373 Zio studies were reviewed (see Table 1). 28.4% had structural heart disease and 16.9% had a prior surgical, catheterization, or electrophysiology procedure. The most common indication for monitoring was tachypalpitations (41%). The median duration of monitoring was 5.0 days. Overall, 5.1% of Zio monitoring yielded class A findings (11 SVT, 7 ventricular tachycardia/ectopy, and 1 AV block), and 4.0% had class B findings. The remainder had findings categorized as classes C or D. Class A/B findings were 2.8 times more likely in those >10 years (95% CI 1.1 to 7.3, $P = 0.033$). For patients with tachypalpitations and without structural heart disease, 13.2% had pathological arrhythmias (Class A/B); 72.9% had normal variant rhythm and could be discharged from cardiology care. Of all patients who had findings resulting in intervention or increased surveillance, 56% had findings first appearing beyond a traditional 24-hour monitoring period, and only 62% were patient-triggered events (see Figure 1). Only 7 studies (1.9%) were associated with complications or patient intolerance.

Conclusion: The Zio is a well-tolerated device that may improve what traditional Holter and event monitoring would detect in pediatric cardiology patients. This study shows positive clinical impact on the management of patients within a pediatric cardiology practice.

2:45 PM – 3:00 PM

Health care-associated infections are associated with increased length of stay and cost but not mortality in children undergoing cardiac surgery

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Purpose: Health care associated infections (HAIs) have been shown to increase mortality, length of stay and cost in both the adult and pediatric populations. Recent studies have examined the incidence of and risk factors for developing HAIs in the pediatric population after cardiac surgery. This study aims to evaluate the impact of HAIs on length of stay, inpatient mortality and cost of hospitalization in the pediatric population after cardiac surgery.

Methods: Data from the Kids' Inpatient Database (KID) from 1997 to 2012 was used for this analysis. Patients under 18 years of age who underwent cardiac surgery were included. HAIs were defined as central line associated blood stream infections (CLABSIs), catheter associated urinary tract infections (CAUTIs), ventilator associated pneumonias (VAPs) and surgical wound infections (SWIs). Similar conditions such as sepsis, pneumonia, and mediastinitis were not included. Univariate analysis was conducted to compare characteristics between admissions with and without a HAI. Next, regression analysis was done to determine what patient characteristics, comorbidities, cardiac lesions, and cardiac surgeries were independently associated with a HAI. Next, regression analysis was done to determine what specific HAIs were independently associated with increased hospital length of stay, increased total charges of hospitalization, or increased inpatient mortality.

Results: A total of 46,169 admissions were included. Of these, 773 (1.6%) had a HAI. Those with a HAI tended to be younger with a median age of 6 months compared to a median age of 12 months in those without an HAI ($P < 0.001$). Regression analysis demonstrated that younger age (beta coefficient 0.8, $P < 0.001$), heart failure (odds ratio 1.2, 95% confidence interval 1.1 to 1.4, $P = 0.034$), acute kidney injury (2.7, 2.0 to 3.6, $P < 0.001$), arrhythmia (1.7, 1.1 to 2.5, $P = 0.004$), atrioventricular septal defect (1.4, 1.1 to 1.8, $P = 0.010$), pulmonary atresia (1.7, 1.1 to 2.4, $P = 0.005$) and hypoplastic left heart syndrome (1.8, 1.3 to 2.3, $P < 0.001$) were all independently associated with a HAI. The presence of any HAI was associated with increased length of stay (median 29 vs 6 days, $P < 0.001$), increased total charges for the hospitalization (median $271,884 vs $88,385, $P < 0.001$), and increased inpatient mortality (6.1% versus 2.5%, $P < 0.001$) by univariate analysis. Regression analysis demonstrated that CLABSI, CAUTI, VAP, and SWI were all independently associated with increased length of stay and increased total charges for the hospital stay. No specific HAI was associated with increased inpatient mortality after regression analysis.

Conclusions The incidence of HAIs in this analysis was extremely low (1.6%). Patients with AKI were 2.7 times more likely to develop a HAI. HAIs contributed significantly to an increase length of hospitalization and cost of hospitalization. No HAI was independently associated with increased patient mortality.

3:00 PM – 3:15 PM

Newborn screening for critical congenital heart disease: is it time to change the algorithm?

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Introduction: The most commonly used algorithm for newborn screening for critical congenital heart disease (CCHD) is the one endorsed by the American Academy of Pediatrics (AAP). However, the impact and feasibility of this algorithm has been questioned. The objectives of this study were to 1) determine the impact of newborn CCHD screening in a large, tertiary care birth hospital using the AAP algorithm and 2) model what the impact may be under an alternative algorithm.

Methods: Newborn CCHD screening results were collected on term infants born at a large tertiary birth hospital in Atlanta, GA between January 1, 2013 through December 31, 2016 using the AAP algorithm. Infants with a prenatal diagnosis of CCHD and infants transferred to the NICU prior to screening were excluded from screening. Infants without sufficient pulse oximetry data (n=39) were excluded from the analysis. Clinical records were reviewed at the birth hospital and the sole pediatric cardiac surgery center in the area to identify true negatives, true positives, false-negatives, and false-positives. A simulation study was then performed to model how the results would have differed if the algorithm had been modified to have only one repeat pulse oximetry test instead of two.

Results: A total of 77,145 newborns met criteria for the study. After correcting for errors in provider interpretation in 323 cases (including four initially misinterpreted as passing), 77,114 (99.9%) infants passed

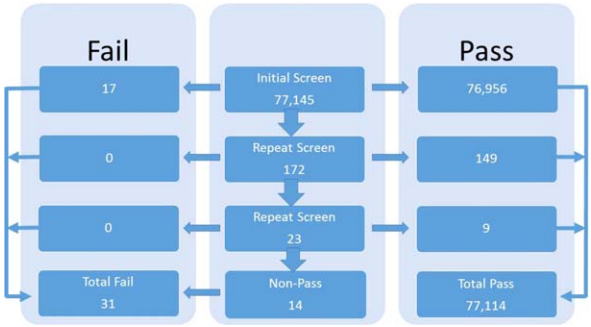

FIGURE 1 Critical congenital heart disease screening results for 1st, 2nd, and 3rd screening attempts per the current screening algorithm

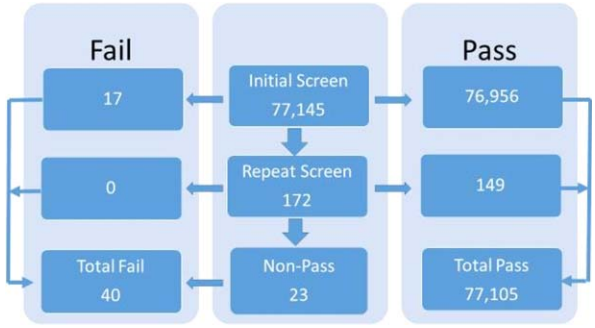

FIGURE 2 Critical congenital heart disease screening results for 1st and 2nd screening attempts per a modified screening algorithm

screening, 17 infants failed screening on the basis of having an initial saturation of < 90% in the hand or foot, and 14 failed screening on the basis of not having a passing level after three pulse oximetry tests (Figure 1). There was one true-positive (total anomalous pulmonary venous return), 30 false-positives, and six false-negatives (one with hypoplastic left heart syndrome, two with tetralogy of Fallot, and four with coarctation of the aorta), yielding an overall sensitivity of 14.3% and false-positive rate of 0.039% (1 per ~2500 births). Among the false-positives, eight (26.7%) had significant disease other than CCHD. If the algorithm had been modified to have only one repeat pulse oximetry test instead of two (Figure 2), the overall sensitivity would have remained 14.3% and the false-positive rate would have increased to 0.051% (1 per ~2000 births).

Conclusion: CCHD screening using pulse oximetry may not detect many new cases of CCHD in a tertiary care setting, but it can detect other important causes of hypoxemia. Modifying the CCHD screening algorithm to have only one repeat pulse oximetry test instead of two may help detect other significant disease without a substantial increase in the false-positive rate. Further efforts to improve the sensitivity of screening are warranted.

3:30 PM – 3:45 PM

Misalignment of coronary microvasculature detected in a model of prenatal alcohol exposure using optical coherence tomography

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Purpose: Congenital coronary anomalies can result in severe consequences such as arrhythmias and sudden death. Research has been dedicated to coronary development, but gaps exist in this area of study due to a lack of tools to visualize all the vessels. Several modalities are used to visualize coronary vasculature such as fluorescent microscopy and ink contrast studies. Three-dimensional organization of microvasculature, including arterioles, venules, and capillaries, is not well defined

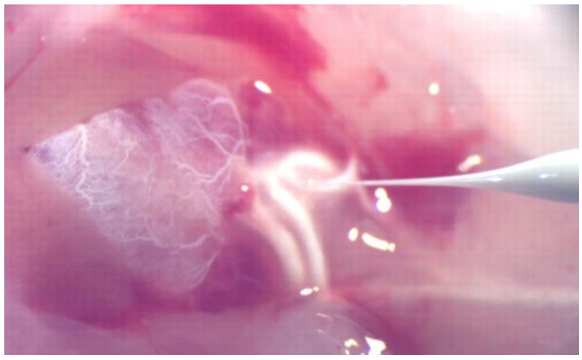

FIGURE 1 This picture depicts the injection of SLIME into the aorta, filling the coronary vasculature.

with these modalities. Using a recently developed optical coherence tomography (OCT)–based vascular imaging technique (scatter labeled imaging of microvasculature in excised tissue—SLIME), we were able to evaluate microvasculature distribution in the embryonic heart. As ethanol is a known teratogen that results in outflow tract and valvuloseptal anomalies, we hypothesized that by using SLIME will be able to detect abnormalities in coronary development in a quail model of fetal alcohol syndrome.

Methods: Fertilized quail eggs were incubated in a humidified incubator at 38°C until Hamburger and Hamilton (HH) stage 4–5 (gastrulation). Forty microliters of a 50% ethanol solution were injected into the air space of the egg. The ethanol dosage is equivalent to one binge drinking episode in humans early in the first trimester. In previous studies, this dosage has consistently produced congenital heart defects. Ten eggs were injected with ethanol solution. Four of the ten survived and were examined. Another four eggs were injected with saline and examined as controls. Injected eggs were incubated until day 8 (HH stage 36), when they were dissected, exposing the great vessels and heart. The aorta was cannulated with a micro needle (Figure 1) and scattering contrast agent was slowly perfused and later fixed in place with a crosslinking agent. The heart was dissected from the embryo and treated with an optical clearing agent making the tissue transparent. After optical clearing, the transparent hearts with scattering contrast labeled vasculature were imaged with OCT and the data were processed and 3D rendered with Amira.

Results: Comparing control hearts to ethanol-exposed hearts, the coronary microvasculature pattern was noted to be more chaotic. In all control hearts, coronary vasculature ran in parallel bundles over the left ventricle toward the apex; whereas, 2 of 4 ethanol hearts revealed microvasculature running in multiple directions. Using SLIME, we detected subtle differences in the alignment of microvasculature that have yet to be described in this disease model. We do not know whether this by itself would result in morbidity or mortality. It may reflect improper alignment of the cardiomyocytes. SLIME is a sensitive tool to study microvascular abnormalities.

3:45 PM – 4:00 PM

Elective pericardial window after surgical closure of secundum type atrial septal defects—does it prevent pericardial effusion?

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Objective: Postoperative pericardial effusion is a well-recognized complication after surgical closure of secundum type atrial septal defect (ASD). The incidence has been reported to be as high as 25%. The effusion may resolve spontaneously or require medical or surgical intervention and may be life threatening. The exact pathophysiology of this process remains poorly understood. We hypothesize that creating a pleuro-pericardial window will prevent the collection of hemodynamically significant pericardial effusion.

Methods: We performed a retrospective analysis of 100 patients who underwent surgical closure of secundum ASDs from 2002–2015. A

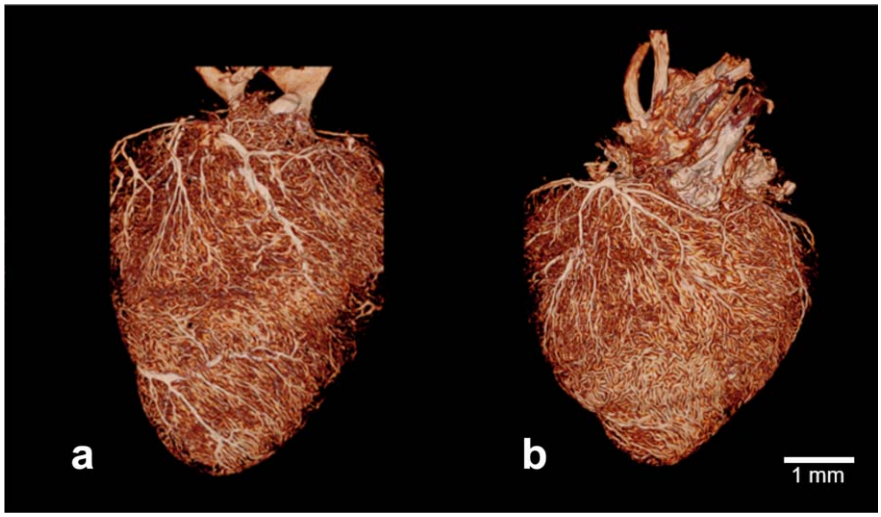

FIGURE 2 (a) Control embryo microvasculature runs in parallel over the LV to the apex. (b) Ethanol-exposed embryo microvasculature is not organized, with branches running in multiple directions over the LV. LV, left ventricle

TABLE 1 Outcomes of elective pericardial window placement

	Elective pericardial window (n = 28)	No pericardial window (n = 72)	*P* vatue
Median length of CICU stay (days)	1(IQR:l-5)	1(IQR:1–38)	0.26
Median length of post-op hospital stay (days)	4(IQR:3–20)	4(IQR:2–21S)	0.26
Post-op chest tube duration (days)	2(1–4)	2(1–4)	0.74
Pericardial effusion (n)	1 (3.5%)	21(29%)	0.005
Pleural effusion (n)	4(14%)	1(1.4%)	0.007
Pleurocentesis (n)	2	1	0.13
Readmissions (n)	5(17.8%)	15(20.8%)	0.74
Readmissions secondary to pericardial effusion (n)	0	11	
Median LOS on readmission (days)		4(IQR:1–20)	
Pericardiocentesis (n)	0	6(54.5%)	

pleuro-pericardial window was created in 28 (28%) patients during surgical repair of ASD (all performed after June, 2010). Patients with windows were compared to the 72 patients without. Outcomes evaluated included: development of significant pericardial effusion, need for pericardial drainage procedures and readmission for pericardial effusion management. Results: Of the 100 patients, 22 (22%) were diagnosed with pericardial effusion based on echocardiograms. The incidence of pericardial effusion was lower among window group (n=1,3.5%) than in the no window group (n=21, 29%, $P = 0.005$). Lengths of stay and overall readmission rates were not significantly different between groups (Table 1), but readmissions for pericardial effusion were also lower in the window group (0.0% vs. 15.2%). Pericardiocentesis was performed in 6 (54.5%) patients (all in the no window group). Pleural effusions were more common in the window group (n=4, 14% vs. n=1, 1.4%, $P = 0.007$). The incidence of pleurocentesis trended to be higher in the window group (7.0% vs. 1.3%, $P = 0.13$) but was not statistically significant.

Conclusions: Hemodynamically significant pericardial effusion is a well-recognized complication after surgical repair of ASDs. Creating a pericardial window is effective in preventing postoperative pericardial effusion.

4:00 PM – 4:15 PM

Does QTc variability influence risk of breakthrough cardiac events in pediatric patients treated for long QT syndrome?

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Purpose: Previous studies of long QT syndrome (LQTS) have shown that a single QTc > 500 ms is associated with greater risk for cardiac events. However, less is known about the persistence of QT prolongation in subsequent ECGs and the risk of breakthrough cardiac events (BCEs) while on LQTS-directed therapy. Here, we sought to determine QTc variability over time and its prognostic effect on BCEs among pediatric patients with LQTS.

Methods: We performed an IRB-approved, retrospective analysis of 433 pediatric LQTS patients [155 (36%) probands, 213 (49%) females, mean age at diagnosis 8 ± 6 years, 88 (20%) symptomatic patients prior to diagnosis] who were evaluated, risk-stratified, and treated in our Genetic Heart Rhythm Clinic between 1999 and 2016. Only patients with at least 2 ECGs were included and computer-derived QTc's were verified manually. ECGs were excluded if patient was within 1 month post denervation surgery, or if ECGs were obtained after the last known BCE. BCEs were defined as arrhythmogenic syncope/seizure, cardiac arrest, ICD shock, and sudden cardiac death.

Results: Of the 433 pediatric LQTS patients, 203 (47%) had LQT1, 167 (39%) had LQT2, 40 (9%) had LQT3, and 23 (5%) had multiple pathogenic LQT mutations (LQTM). A total of 2147 ECGs were included, with a median of 4 (2–6 quartiles) ECGs per patient. The mean QTc was 467 ± 34 ms with a mean QTc range of 45 ± 34 ms. ROC analysis of maximum QTc and BCE after treatment determined 515 ms (AUC 0.89, 73% sensitivity, 61% specificity) to be a cutoff predicting risk of future BCE. The patients were grouped by the proportion of their serial QTcs that exceeded this 515 ms threshold (Figure 1). In 327 patients where serial QTcs never exceeded the threshold, only 4 (1%)

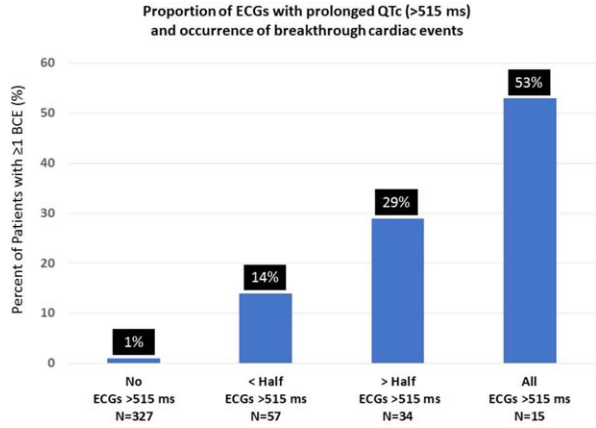

FIGURE 1

experienced a BCE. In contrast, in 57 patients where less than half of serial QTcs surpassed the threshold, 8(14%) had a BCE. In 34 patients where greater than half (but not all) of serial QTcs crossed the 515 ms threshold, 10 (29%) experienced a BCE. Most importantly, in 15 patients where serial QTcs always exceeded the threshold ('all'; Figure 1), more than half (8; 53%) went on to have a BCE, despite being on guideline recommended treatment ($P < 0.001$).

Conclusion: Herein, we show the risk of experiencing a BCE among patients treated for LQTS may be dependent and increase proportionally based upon the persistence of QT prolongation (QTc>515 ms) in subsequent ECGs. These results show the utility of serial ECGs and the necessity to take all historic QTc measurements into consideration in both diagnosis and risk stratification.

4:15 PM – 4:30 PM

Trends in gestational age at birth in patients with congenital heart disease

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Purpose: Studies published as early as 2010 have identified that preterm and early term birth in patients with critical congenital heart disease (CCHD) are associated with worse outcomes than later term birth. We sought to identify trends in gestational age at birth in CCHD to assess if there has been practice change since these publications have been released.

Methods: The Texas Public Use Data File, years 1999–2015, was used. This administrative database captures nearly 100% of hospitalizations in the state of Texas. Two groups of CCHD were defined: group I contained those with diagnoses likely to require intervention within the first year of life, and group II was a subset of CCHD with the highest rates of prenatal diagnoses (hypoplastic left heart syndrome, other single ventricle, and pulmonary atresia). Administrative codes do not distinguish between early and late term delivery, so this additional variable could not be examined. Hospitalizations with discharge diagnoses of multiple gestation births, birth defects, or known genetic abnormalities were excluded. Additionally, the analysis was stratified by high-volume and low-volume centers. A high volume center was defined as, on average, at least one CCHD birth that fulfilled our inclusion criteria monthly.

TABLE 1 Percentage of preterm birth by institution volume

	Group 1		Group II	
	1999–2010	2011–2015*	1999–2010	2011–2015*
High volume centers	240/1234 19.5%	148/937 15.8%	55/350 15.7%	40/312 12.8%
Low volume centers	452/2462 18.4%	248/1232 20.1%	77/431 17.9%	44/188 23.4%

*P value <0.05

Results: There were 5,865 total births in 218 hospitals fulfilling group I inclusion criteria. Of those, 37.0% of group I patients and 54.8% of group II patients were delivered at the 11 high-volume hospitals. There were 1,088 (18.6%) preterm births in group I and 373 preterm births in group II (21.2%). In both groups, there was no significant change in the proportion of preterm births over the study period as a whole. However, when stratifying by volume, the proportion of preterm births differed after 2010 (Table 1). After 2010, the percentage of preterm births in both groups at high volume centers was significantly less that in the low-volume centers ($P = 0.0113$ for group I, $P = 0.0033$ for group II). In the high volume centers for the group I population, there was a 19% decrease in the proportion of preterm births after 2010 ($P = 0.0278$). In the high volume centers for the group II population, there was an 18% decrease in preterm birth, although this was not statistically significant ($P = 0.2895$).

Conclusion: This study demonstrates that since 2010, the higher-volume centers in Texas have a lower rate of preterm delivery in CCHD compared to low volume centers. This effect amplified when focused on diagnoses that tend to be noted prenatally. These trends were not noted when compared to prior years. This may reflect a greater knowledge of the recent literature in centers with more experience with CCHD births.

4:30 PM – 4:45 PM

Usefulness of routine transtelephonic monitoring for supraventricular tachycardia in infants

Jonathan Yaari, MD; Dorota Gruber, MSc; Andrew D. Blaufox, MD, Cohen Children's Medical Center/Northwell Health, Queens, NY

Purpose: Infants may be more susceptible to hemodynamic collapse due to prolonged supraventricular tachycardia (SVT) because of their inability to communicate symptoms. Routine home transtelephonic monitoring transmissions (TTM) after SVT has been diagnosed has not been reported as a standard monitoring tool in this age group. We set to determine if prescribing TTMs to be used routinely as well as when parents have concerns regarding recurrent SVT can accurately detect SVT in asymptomatic infants and/or assuage parental concerns.

Methods: A single center, retrospective chart review of 62 pts with fetal or infant SVT who were prescribed TTM for at least 30 days from 1/10–9/16 was performed. Patients were excluded if initial SVT was not documented, the SVT occurred in the perioperative period, or the patient had atrial fibrillation or atrial flutter. Categorical variables are expressed as mean ± SD. Student's t test and chi-square analysis were used for continuous and categorical variables respectively.

Results: Sixty-two patients (mean age 20 ± 32 days, 41 (66%) male) with fetal and/or infant SVT subsequently discharged with TTM after initial hospitalization were included. SVT diagnoses included: Orthodromic reciprocating tachycardia in 47 (75.8%), atrial tachycardia in 13 (21%), congenital junctional ectopic tachycardia in 1 (1.6%), and permanent junctional reciprocating tachycardia in 1 (1.6%). A total of 2846 TTMs were received from 57/62 patients (45.9 ± 51.5 TTMs/pt) over 61.1 ± 65.9 days (d) of monitoring (0.75 ± 0.65 TTMs/pt/d). Routine asymptomatic TTMs revealed actionable findings in 7 (0.25%) TTMs sent by 7 (11.3%)

patients. Number of routine transmissions was not associated with initial length of stay ($P = 0.6$) or medical regimen ($P = 0.3$). Actions taken include ER evaluation only (n=1), ICU admission (n=4), medication dose adjustment (n=5), initiation of new medication (n=4), and/or ablation (n=1). Significant LV dysfunction was found in 1/5 patients who underwent echocardiographic evaluation prompted by TTM findings. No asymptomatic patient presented in shock or died. Neither medication regimen ($P = 0.13$) nor length of initial hospitalization ($P = 0.3$) influenced the likelihood that actionable items would be found on TTMs. Forty-five (1.6%) TTMs were made for parental concerns/symptoms in 16 (25.8%) patients. Findings in symptomatic TTMs included NSR in 37 (82%) TTMs and SVT in 8 (18%) TTMs corresponding to patients with NSR only (n=10pts), SVT only (n=3pts), and NSR and SVT on different TTMs (n=3pts). Symptomatic TTMs with actionable findings were more likely to be sent by patients discharged on Class I or III antiarrhythmics ($P = 0.006$) and from patients with prolonged initial hospitalizations ($P = 0.02$). 1 patient on flecainide was noted to have asymptomatic widened QRS on routine TTMs prompting discontinuation of therapy.

Conclusions: TTMs can accurately diagnose asymptomatic recurrent SVT in neonates and infants before they develop signs of CHF or shock and is helpful for recurrent SVT management and parental reassurance.

SATURDAY, SEPTEMBER 16, 2017

8:30 AM – 8:45 AM

Aortic stenosis echocardiogram doppler discrepancies

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Purpose: Aortic stenosis (AS) requires frequent monitoring and evaluation. Doppler echocardiography is the standard approach used to monitor velocities and pressure gradients. The most commonly used echocardiographic windows are the suprasternal notch (SSN) and apical (Ap) views. To our knowledge there has been no study comparing the agreement of velocity measurements from these two views to each other. Our hypothesis is that the velocity obtained from SSN will be systematically higher than the velocity obtained from the Ap view.

Methods: Institutional IRB approval was obtained. A retrospective chart review was performed for patients with AS and healthy subjects from January 1, 2012 to May 22, 2016. We excluded AS patients who had aortic valve surgery and individuals with aortic valve peak Doppler velocity < 2 m/s. We recorded maximum velocity, mean velocity, and estimated pressure gradients from both views.

Results: There were 92 patients with AS and 98 healthy subjects. The max SSN and Ap velocities were 3.3 m/s and 3.05 m/s for AS subjects, and 1.27 m/s and 1.19 m/s for healthy subjects. There was a positive bias for the SSN compared to Ap in both AS and healthy subjects, with a mean difference +0.24 m/s in the AS group and +0.08 m/s in the healthy subjects group ($P < 0.0001$ for AS group and $P = 0.0036$ for healthy group). This was an average disagreement of +7.5% for the AS group and +6.3% for the healthy group. In both groups, the magnitude of the disagreement increased as maximum SSN velocity increased. When comparing mean gradients, the positive bias disappeared in both the AS group ($P = 0.34$) and healthy group ($P = 0.99$).

Conclusion: Maximum Doppler velocities obtained from the suprasternal notch views were higher in both control and aortic stenosis patients and the magnitude of that difference increased at higher velocities. There appears to be a systematic difference in the measurements from these two different windows. This difference can impact clinical

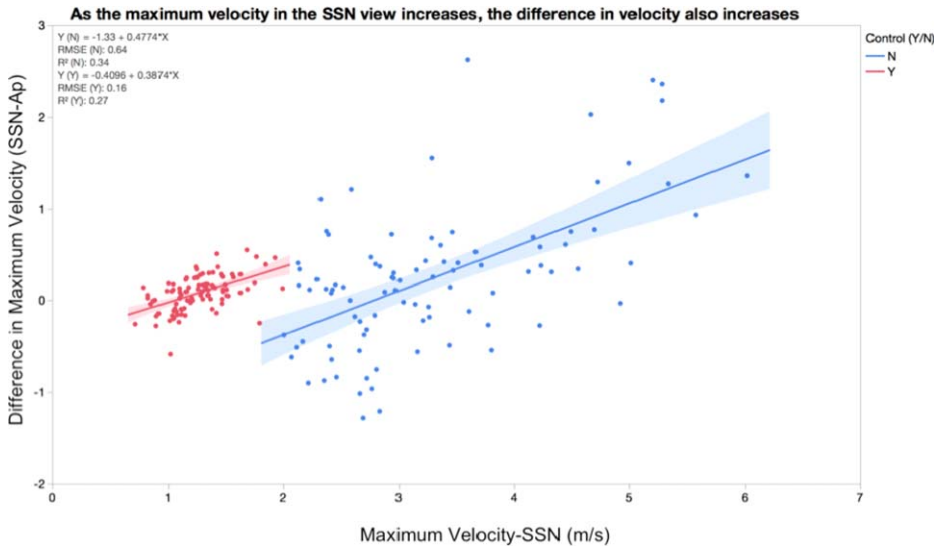

FIGURE 1 Maximum SSN velocity against difference in SSN and Ap velocities. As the maximum velocity from the SSN view increases, the difference between SSN and Ap views increases in both the control and test groups.

decision making about patient care and thus it is important to be consistent in the manner of obtaining and noting where the measurements are obtained.

8:45 AM – 9:00 AM

Quality of life in pediatric patients after cone reconstruction for Ebstein anomaly

Kristen L. Sessions, BS; Charlotte Van Dorn, MD; Joseph A. Dearani, MD; Simrit Warring, BS; Kaitlin Leopold, BS; Philip L. Wackel, MD; Frank Cetta, Jr., MD; Jonathan Johnson, MD, Mayo Clinic, Rochester, MN

Purpose: Cone reconstruction (CR) has become the standard of care for Ebstein anomaly and has excellent outcomes. However, quality of life (QOL) in patients with Ebstein anomaly after CR has not been reported. Here we sought to evaluate the health status and QOL of pediatric patients who had CR for Ebstein anomaly.

Methods: We reviewed all patients who had CR from 2007 to 2016 at our institution. Prospective surveys were mailed to all eligible patients. QOL was assessed using the PedsQL 4.0 Generic Core Scales, including four domains: physical, emotional, social, and school functioning.

Results: Of 116 eligible patients, 71 (61%) responded (average age 13.5, range 3–24). Ninety-six percent reported their health as excellent or good, and 55% were symptom free. Sixty-two percent were taking no medications; the most common medication was aspirin in 30%. Only 18% had been hospitalized for cardiac reasons following CR. The average self-reported QOL was 87.6/100 while the average parent proxy-reported QOL was 81.9/100. For the majority, there was no difference by self or parent proxy-report in QOL between CR patients and healthy children; however, QOL was significantly better for CR compared to children with chronic health conditions (Table). By self-report and parent proxy-report, 17% and 15% of patients were deemed "at risk" for reduced QOL respectively. In terms of social functioning, 70/71 (98%) of patients are either full-time students or working full-time.

Conclusions: Overall, children with Ebstein anomaly who had CR have excellent QOL comparable to healthy peers and significantly better than other children with chronic health conditions. Families of children with Ebstein anomaly can expect excellent QOL, long-term health status and social functioning following CR.

9:00 AM – 9:15 AM

Infants of diabetic mothers have altered arterial stiffness from birth

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Background: Children born to mothers with diabetes mellitus (MDM) are known to have higher systolic blood pressures. Experimental data suggests a role for in utero metabolic alterations in fetal cardiovascular programming. The main hypothesis of this study is that fetal programming causes alterations in aortic stiffness. Alteration in aortic stiffness plays a role in hypertension. Arterial stiffness can be reasonably measured in adults using pulse wave velocity (PWV).

Purpose: This prospective pilot study explores the hypothesis by comparing the elastic properties of the aorta/arterial stiffness in neonates and infants of MDM after establishing normative data using PWV.

Methods: Neonates born to MDM (NMDM) and without any systemic disease (controls- NC) were prospectively recruited. After maternal informed consent, blinded echocardiograms and ten spectral PW Doppler waveforms were performed in the right (R) common carotid artery in the neck and R femoral artery in the groin at same heart rates using 12MHz transducer. The time delay between the foot of the waveform at the two sites was obtained by gating to the peak of the R wave on the electrocardiogram (t). Average of 10 recordings was used (t). The distance (d) traveled by the waveform was calculated by

TABLE 1 PedsQL 4.0 Generic Core Scales for children after cone reconstruction. healthy children, and children with a chronic health condition

	Cone reconstruction	Healthy sample	Chronic health condition*	*P* value**	*P* value***
Patient self-report					
Total score	85.57 (14.58)	83.91 (12.47)	74.16 (15.38)	0.22	**<0.01**
Physical	87.57 (14.94)	87.77 (13.12)	79.47 (17.07)	0.42	**<0.01**
Psychosocial	84.24 (18.33)	81.83 (13.97)	71.32 (17.13)	0.11	**<0.01**
Emotional	80.68 (22.29)	79.21 (18.02)	69.32 (21.36)	0.35	**<0.01**
Social	87.88 (13.78)	84.97 (16.71)	76.36 (21.57)	**0.01**	**<0.01**
School	82.97 (18.33)	81.31 (16.09)	68.27 (19.05)	0.08	**<0.01**
Parent proxy report					
Total score	81.85 (15.26)	82.29 (15.55)	73.14 (16.46)	0.75	**<0.01**
Physical	86.57 (16.16)	84.08 (19.70)	76.99 (20.20)	**0.03**	**<0.01**
Psychosocial	79.27 (16.46)	81.24 (15.34)	71.04 (17.32)	0.58	**<0.01**
Emotional	77.55 (21.43)	81.20 (16.40)	71.08 (19.75)	0.24	**0.02**
Social	82.45 (17.86)	83.05 (19.66)	75.06 (21.75)	0.93	**0.02**
School	77.80 (20.76)	78.27 (19.64)	65.58 (20.75)	0.91	**<0.01**

*Children with asthma. diabetes, attention deficit hyperactivity disorder, depression, or "other"
**P value comparing cone reconstruction cohort to healthy control cohort
***P value comparing cone reconstruction and children with chronic health conditions

subtracting the distance between the second intercostal space and the sampling site on the carotid artery from the sum of second intercostal space to the inferior edge of the umbilicus and inferior edge of the umbilicus to the sampling site on the right femoral artery. Aortic PWV was calculated as the ratio of d & t. Serial measurements were performed at birth, 1 week and 1 month after birth. Statistical comparisons were made using Student's *t* test and regression analysis was performed to determine the best fit for PWV in NMDM and NC.

Results: Echo and PWV assessments were performed serially on 6 NMDM and 20 NC at 1.7 ± 0.6, 6.8 ± 0.8 and at 37.5 ± 8.1 days. Mean gestation ages ± standard deviation (SD) were 39.0 ± 1.3 weeks with mean birth weight of 3.35 ± 0.05 kg and mean length of 49.7 ± 2.5 cm in the entire cohort. Mean maternal Hgb A1c in NMDM was 5.6%. Mean PWV at birth in NMDM was 0.36 ± 0.07, this increased to 0.56 ± 0.6 by 1 month (PWV= 0.348 + 0.007*days of life, R=0.71, p+0.12, this increased to 0.55 ± 0.03 by 1 month postnatal age (PWV=0.44061 + 0.003*days of life, R=0.42, $P < 0.05$).

Conclusion: Arterial stiffness in NMDM is different at birth than NC and changes rapidly after birth. Longitudinal follow-up is required to evaluate if these effects persist beyond neonatal period. It is feasible to assess PWV using ultrasound in neonates.

9:15 AM – 9:30 AM

Elevated lipoprotein (a) within the pediatric population

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Purpose: Cardiovascular disease (CVD) is a public health problem and a leading cause of death globally. Prevention of CVD should begin in early childhood with focus on promotion of healthy lifestyle choices. Risk factors such as a family history of premature CVD and lipid abnormalities may warrant early intervention. Common lipid abnormalities include elevated low-density lipoprotein cholesterol (LDL-C), decreased high-density lipoprotein cholesterol (HDL-C), elevated triglycerides (TG), and/or elevated lipoprotein (a) [Lp(a)]. Elevated Lp(a) is not well studied in children and is believed to accelerate atherosclerosis. In addition, atherosclerosis decreases vascular distensibility which is the ability of the vasculature to stretch and accommodate the pulsatile output of the heart. This study examines the relationship between elevated Lp(a), family history of premature CVD, lipid abnormalities and vascular distensibility within a high-risk pediatric population.

Methods: This is a single-center, retrospective study that obtained data for 257 children who met criteria to be followed in the Preventive Cardiology Clinic. The independent variable is Lp(a) level, which separated patients into high-risk case group [Lp(a) (≥ 30mg/dL)] and lower risk control group [Lp(a) (< 30mg/dL)]. The dependent variables are family history of premature CVD or death at < 50 years of age and dyslipidemia, defined as LDL-C >130 mg/dL, HDL-C < 45 mg/dL and/or

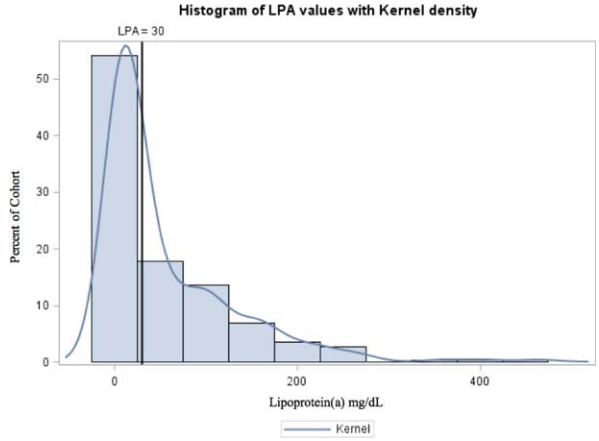

FIGURE 1 Lp(a) Distribution. Histogram of LPA values with kernel density function overlay and vertical line at LPA=30 to separate case and control groups.

TG >100 mg/dL. The final dependent variable is vascular distensibility, measured with the the SphygmoCor XCEL pulse wave velocity (PWV) analyzer. PWV (meters/second) is calculated as the distance from the carotid to femoral artery pulse interrogation area divided by the time taken for the pulse to travel that distance.

Results: There was a significant association between elevated Lp(a) and positive family history of CVD ($P = 0.02$), higher HDL-C ($P = 0.01$), and lower TG ($P < 0.00$). No significant associations were found for other predictors.

Conclusion: These findings reveal a relationship between elevated Lp(a), family history of premature CVD, and specific lipid abnormalities in high-risk children. These findings may contribute toward a wider adoption of Lp(a) as a screening tool for identification of high-risk children so that lifestyle measures can be further intensified to avoid premature CVD.

9:30 AM – 9:45 AM

What is the diagnostic yield of echocardiograms in pediatric patients with structurally normal hearts with suspected infective endocarditis?

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Purpose: Infective endocarditis (IE) has a high mortality and morbidity even though it is rare in children. The estimated incidence in children is about "0.3 per 100,000 children per year with a mortality of 11.6%." In patients with bacteremia, the incidence of endocarditis in congenital heart disease group is 53% compared to 3% in structurally normal hearts. The aim of this study was to assess if echocardiograms were necessary and effective in the diagnosis of endocarditis in patients with structurally normal hearts. In addition, we evaluated the positive predictive value of echocardiogram in patients with low clinical suspicion for IE.

Methods: We performed a retrospective chart review of patients under 21 years of age with echocardiograms completed for indication of endocarditis from 2005–2015. We excluded patients with congenital heart

disease, prior IE, and any follow-up IE echocardiograms. We looked at IE risk factors (fever, blood culture, central line, vascular and immunologic phenomenon), Dukes' criteria, diagnosis of IE at discharge, and whether antibiotic regimens were changed because of the echocardiogram.

Results: 300 patients were included in the study. 128 (43%) patients were female. Of the 300 patients, 10 echocardiograms (3%) were positive for mass, abscess, thrombus, or new valve regurgitation consistent with IE. 8 patients were diagnosed with IE. One patient had a small vegetation but negative blood cultures so no antibiotics were started and no IE diagnosis was made. The other patient had new mild mitral valve regurgitation, but after the cardiology team was consulted, the echo appeared to be within normal limits. The patient was treated for bacteremia instead. Patients with positive Dukes' criteria had the highest positive predictive value (PPV) for a positive echocardiogram (0.8) compared to just one positive blood culture (0) and two positive blood cultures (0.3).

Conclusion: Echocardiogram should not be used as a screening tool for IE. Patients should be evaluated based on the Dukes' criteria to assess the patients' clinical probability of IE. Patients with a low clinical probability of IE should not have an echocardiogram performed as it has low positive predictive value and low diagnostic yield. An echocardiogram should be considered with 2 or more persistent positive blood cultures, persistent fever on antibiotic therapy, and/or new pathologic murmur. Infectious disease and/or pediatric cardiology should be consulted if there is concern for IE prior to ordering an echocardiogram.

9:45 AM – 10:00 AM

The effect of right ventricular function on survivability and mortality following stage 2 palliation: an analysis of the single ventricle reconstruction trial public data set

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Purpose: Hypoplastic left heart syndrome (HLHS) requires a three stage surgical approach for survival with the highest risk being between Stage 1 (S1P) and Stage 2 (S2P) palliation. Prominent systemic AV valve regurgitation has long been demonstrated to be a poor prognostic indicator in S2P and beyond, though little is known how RV morphology and function, in isolation, affects outcomes. We sought to evaluate the impact of different RV indices, taken just prior to S2P, to evaluate the impact of RV size and function on morbidity and mortality.

Methods: A retrospective study was performed utilizing the Pediatric Heart Network Single Ventricle Reconstruction Trial Public Data Set involving data collected from 15 centers between 2005 and 2009 that underwent randomization for shunt type as part of S1P. Any variant of S1P performed in the trial was included in addition to all of the anatomic HLHS variants reported. Those who required extensive surgical interventions, out of the scope of standard S2P, were excluded. Failure of S2P was defined as those who died, were listed for transplant, or were transplanted soon after S2P. Morbidity was evaluated in regards to hospital length of stay and duration of intubation. Echocardiographic measurements, as part of the trial, were standardized and interpreted by a core laboratory and are based on studies prior to S2P. Descriptive analysis was performed using appropriate parametric and nonparametric hypothesis testing. Confounders in the data set known to have an effect on the outcome of interest were controlled for in the regression analysis, including presence of systemic AV valve regurgitation.

Results: A total of 283 patients met criteria for analysis. Of those, only 18 patients died, were listed for transplant, or were transplanted. In comparing echo measurements in those who successfully underwent S2P with those who failed, RV fractional area change was less in those who failed (30% vs 34%, $P = 0.039$) and RV indexed end diastolic volume (RVEDVI) and end systolic volume (RVESVI) were larger in those who failed (142.74 mL/BSA1.3 vs 111.29 mL/BSA1.3, $P = 0.023$, 88.45 mL/BSA1.3 vs 62.75 mL/BSA1.3, $P = 0.025$, respectively). Larger RVEDVI and RVESVI measurements were associated with failure (OR 1.17 [1.01-1.35] $P = 0.021$, OR 1.25 [1.03-1.52] $P = 0.021$, respectively). RV ejection fraction showed that for every 10% increase, there was a 63% decrease in length of stay and 68% decrease in duration of intubation ($P = 0.014$, and $P = 0.039$, respectively).

Conclusion: Patients with decreased RV fractional area change and larger RVEDVI and RVESVI were more likely to have failed S2P. Preserved RV function was associated with a shorter hospital length of stay and duration of intubation. Echocardiographic measurement of RV function and volumes during the interstage period can be utilized to determine the prognosis of patients following S2P.

10:15 AM – 10:30 AM

The Medtronic Micro Vascular Plug: Vascular embolization applications in congenital heart disease

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Introduction: The Medtronic Micro Vascular Plug (MVP) is a large diameter vascular occlusion device deliverable through a microcatheter, obviating the need to track delivery sheaths or larger catheters through small torturous vessels, and it can be recaptured for controlled deployment. The MVP device can be used in pediatric patients for vessel embolization, including venovenous collaterals, aortopulmonary collaterals and patent ductus arteriosus (PDA).

Purpose: Describe the clinical use of the MVP device in the treatment of congenital heart disease as it applies to vascular embolization.

Methods: Medical records of all pediatric patients who underwent vascular embolization with the MVP device between September 2015 and April 2017 were reviewed from a single center. Procedural details, complications, and short term outcomes were recorded.

Results: Thirty-five children underwent implant of 41 MVP devices. Six patients received 2 devices during the same procedure. Twenty-five patients (median age 9.3 weeks (2.1–107.6 weeks); median weight 2.5 kg (1.3–14.7 kg)) underwent PDA device closure. Twenty-two of the 25 PDA patients were preterm infants in the NICU at the time of device implant. Non-PDA patients were older at a median of 44.7

weeks (6.7–187.1 weeks) ($P = 0.05$), and larger at a median of 8 kg (3.5 – 18.1 kg)) ($P = 0.004$). Two patients received 2 MVP devices each for venovenous collateral vessel occlusion. Four patients had 5 MVP devices implanted into major aortopulmonary collateral arteries. Three patients received 5 MVP device implants for the embolization of an internal mammary artery, and 1 patient received 2 MVP devices for aortopulmonary collateral embolization. A single device was sufficient for vessel occlusion in 34 patients; one patient had an MReye coil placed after the MVP device in each of the internal mammary arteries to facilitate complete embolization. There were no procedural complications. 34 patients (97%) have complete occlusion of the target vessel with no short term complications a median of 30 weeks (0.6–80 weeks) after implant. One PDA patient had device embolization to the right pulmonary artery identified 9 days after the procedure. The device was retrieved and another device placed with no further sequelae.

Conclusion: This is the largest experience to date describing MVP device implantation in the congenital heart disease population. The MVP device allows for safe and effective closure of abnormal collateral venous and arterial vessels in multiple applications, and provides an appealing device for PDA closure, particularly in small, preterm infants.

10:30 AM – 10:45 AM

Imaging of the conduction tissue in porcine hearts using optical coherence tomography

Orhan U. Kilinc, MD[1]; Xiaowei Zhao, BS[2]; Deniz Dosluoglu, Undergraduate Student[2]; Michael W. Jenkins, PhD[3]; Christopher S. Snyder, MD[4]; Andrew M. Rollins, PhD[5]; (1) Department of Pediatric Cardiology, University Hospitals Rainbow Babies & Children's Hospital, Cleveland, OH, (2) Case Western Reserve University, Cleveland, OH, (3) The Congenital Heart Collaborative, Case Western Reserve University, Cleveland, OH, (4) The Congenital Heart Collaborative, Rainbow Babies and Children's Hospital, Case Western Reserve University School of Medicine, Cleveland, OH, (5) CWRU, Cleveland, OH

Background: Optical coherence tomography (OCT) uses near-infrared light to image tissues. Clinically it has been used to image the walls of coronary arteries. In research settings, OCT is used to visualize sub-endocardial structures. The purpose of this experiment was to determine if OCT can identify native conduction tissues in an adult porcine heart.

Methods: Right atrial endocardial surface of an excised adult porcine heart was exposed. Triangle of Koch was identified by classical anatomical landmarks and was imaged with the OCT system. After the conduction tissue was identified with OCT, the field of view was marked with pins and the sample was fixed in 10% formalin for histology. Sample was stained with Masson's trichrome in the core pathology facility using standard techniques. Histologic examination was done with standard light microscopy. Results of histologic preparations and OCT images were then compared.

Results: Three porcine hearts were examined by this methodology. OCT images identified subendocardial structures presumed to be the proximal His bundle that had differentiation from the surrounding muscle fibers and connective tissues (Figure 1a). Light microscopy of the histology preparations delineated the different tissue types and conduction tissue was easily identified. Location of the hypo-reflective area on the OCT images correlated with the location of conduction tissue in the histology preparation.

Conclusion: Cardiac conduction tissue can be identified by OCT in freshly dissected unfixed porcine hearts. OCT images distinguished the differentiated conduction tissue, in close proximity with the endocardium, myofibers and fibrous tissue; this was verified with histology. This technology may be useful in direct visualization of the native conduction system during procedures in the operating room and electrophysiology laboratory. Further studies with perfused tissue samples and live animal experiments are needed to better assess efficacy of this novel application.

10:45 AM – 11:00 AM

Predictors of oral feeding success during the interstage period for single ventricle patients

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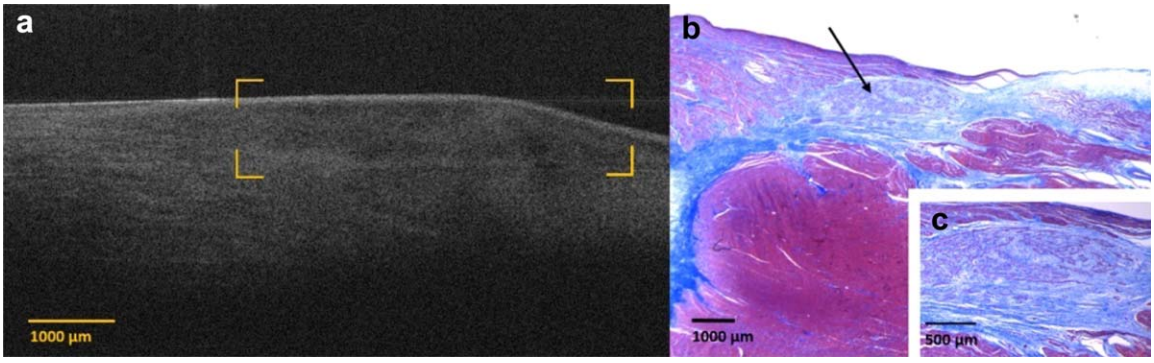

FIGURE 1 The OCT image of a cross section in the Triangle of Koch (a). A spindle-shaped hypo-reflective area (brackets) was identified in the sub-endocardial tissue. Histological examination of the same area (b) with Masson's trichrome staining, showing differentiated conduction fibers (arrow) were seen in close association with the connective tissue (blue) and myofibers (purple). Larger magnification of the differentiated tissue (c) shows cells with large nuclei and pale cytoplasm surrounded with connective tissue, consistent with conduction tissue.

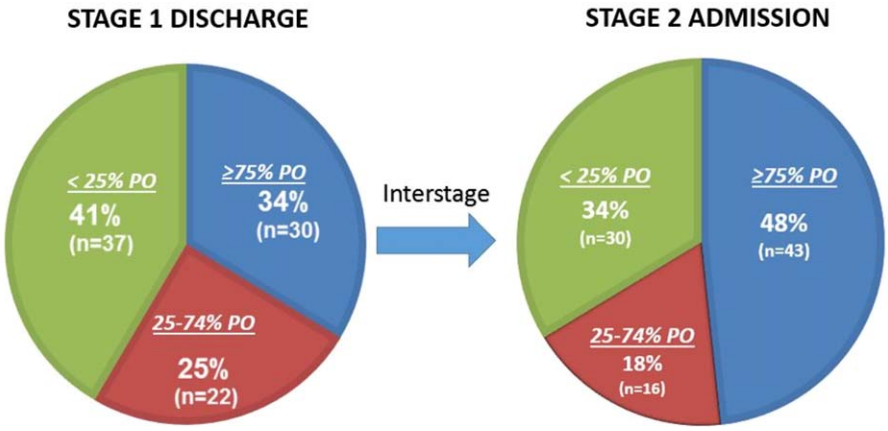

FIGURE 1 Comparative distribution of patients by feeding ability from stage 1 discharge to stage 2 admission. Comparative distribution of patients by feeding ability from stage 1 discharge to stage 2 admission. Pie chart displays three feeding ability groups by percentage of feeds taken by mouth (PO): ≥75% (blue), 25–74% (red); <25% (green). The percentage of the study group and number of patients in each category is listed (%(n)). Total study group=89 patients.

Objective: Single ventricle patients experience significant feeding and growth related morbidity during the interstage period. The primary aim of this study was to determine predictors of oral feeding ability during the interstage period.

Methods: In this nested case-control retrospective review, study participants were selected from a cohort of single ventricle patients at a single institution who underwent stage one and stage two palliation. A case was considered an oral feeding success and defined as a patient who, at the time of admission for S2P, was taking >=75% of goal feeds by mouth. The controls were patients from the same cohort who took less than 75% PO at time of admission for S2P. Univariate analyses were conducted to provide description of study participants. Bivariate analyses were conducted to demonstrate nonadjusted associations between potential predictors and oral feeding success. Finally, multivariable logistic regression using stepwise selection methods was conducted to determine independent predictors of oral feeding success at time of admission for S2P.

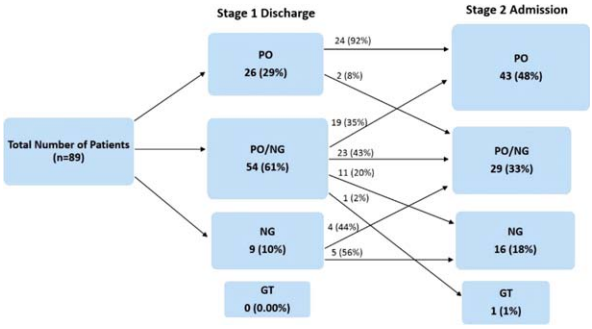

FIGURE 2 Interstage change in feeding route.
Movement of patients between feeding routes at time of stage 1 discharge and stage 2 admission. Patients were categorized by four different feeding routes: all oral feeds (PO), combined oral and nasogastric feeds (PO/NG), nasogastric feeds only (NG), or gastrostomy tube feeds (GT). Boxes contain the total number of patients and percentage of entire study group (n (%)) at stage 1 discharge and at stage 2 admission. N=89 for the study group. The lines with arrows indicate the flow of patients from the respective feeding route category at stage 1 discharge to the respective feeding route category at stage 2 admission. The total number and percentage of patients moving from one respective feeding category to another is listed above the line (n (% of stage 1 feeding category))

Results: Of the 89 study patients, 43 (48%) were feeding successes and were feeding without tube supplementation at stage 2 admission; both increases compared to stage 1 discharge. Taking < 75% of feeds by mouth at stage 1 discharge was independently associated with a decreased odds of feeding success at stage 2 admission (reference group: ≥75% oral feeds; 25–74% oral feeds: OR, 0.11; 95% CI, 0.03-0.48; $P = 0.003$; < 25% oral feeds: OR, 0.02; 95% CI 0.004-0.08, $P = < 0.001$). Variables associated with a lower likelihood of oral feeding success were surgical complication, longer hospital stay and intubation time, olderfirst operation, first oral feed, and stage 1 discharge age, gastroesophageal reflux requiring medical management, and earlier gestational age.

Conclusions: Patients taking a higher percentage of feeds by mouth at stage 1 discharge have an increased odds of feeding success at stage 2 admission. Improvement in interstage feeding ability occurs for some single ventricle patients.

11:00 AM – 11:15 AM

Differentiating subclinical fontan failure phenotypes by echocardiogram in adults

Jennifer F. Gerardin, MD[1]; Maria A. Pernetz, RDCS RVT RCCS[2]; Georges Ephrem, MD MSc[2]; Anita Saraf, MD PhD[2]; Fred H. Rodriguez, III, MD[3]; Wendy Book, MD[2], (1) Division of Cardiology, Department of Medicine, Emory University School of Medicine, Wauwatosa, WI, (2) Division of Cardiology, Department of Medicine, Emory University School of Medicine, Atlanta, GA, (3) Division of Cardiology, Department of Medicine and Pediatrics, Emory University School of Medicine and Sibley Heart Center, Atlanta, GA

Purpose: Fontan palliation for single ventricle physiology creates heterogeneous clinical phenotypes. Adult Fontan failure phenotypes have recently been described based on cardiac and noncardiac etiologies.

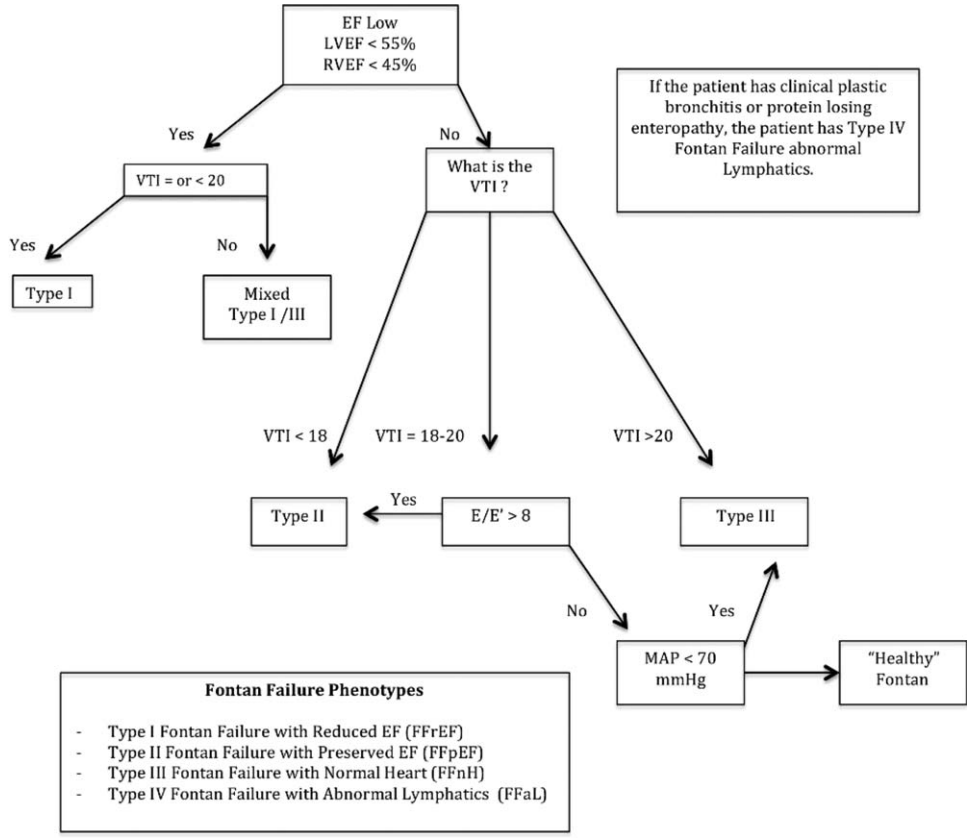

FIGURE 1 Classification system of Fontan failure phenotypes. EF, ejection fraction; LVEF5, left ventricular ejection fraction; RVEF, right ventricular ejection fraction; VTI, velocity time integral; MAP, mean arterial pressure

TABLE 1

Phenotype by echocardiogram								
	Overall N=48	"Healthy" Fontan N=1	Type I N=12	Type II N=14	Type III N=8	Type IV N=3	Mixed Type I/III N=10	*P*
Age *years*	29 (8)	29	33.5 (8)	28.5 (7)	26.5 (5.5)	20 (2)	28 (9)	0.0168
Years since Fontan palliation	24 (6)	19	26.5 (7)	26 (9)	23 (2)	19 (2)	20.5 (7)	0.0041
Systemic ventricle ejection fraction %	52.5 (11.5)	55	42 (16.5)	60 (5)	55 (6.5)	49 (14)	50 (12)	0.0001
E/E′ ratio	6.14 (3.43)	6.13	5.43 (3)	7.39 (3.21)	6.65 (2.3)	13.05 (2.1)	4.78 (3.27)	0.206
VTI *cm*	19.4 (9.8)	20	15.35 (4.25)	13.6 (4.3)	27.05 (8.47)	21.1 (6.4)	24.7 (5.3)	0.0001
MAP *mm Hg*	84 (11.2)	81.7	85.2 (10.8)	87.3 (12.3)	83.8 (11.8)	74.7 (9)	81.8 (6)	0.111
6 minute walk distance *m*	447.5 (86.5)	414.6	414 (95)	458.8 (42)	526.6 (81.1)	513 (42)	420 (72)	0.129
Baseline oxygen saturation %	90 (4.5)	94	90.5 (3.5)	91.5 (6)	89.5 (5)	89 (0)	90 (9)	0.49
Hemoglobin *mg/dL*	15.6 (2.5)	14.7	16.2 (3.3)	15.3 (2.8)	15.8 (3.1)	11.3 (5.6)	15.8 (2.4)	0.141
Albumin *mg/dL*	4.4 (0.5)	3.6	4.4 (0.5)	4.4 (0.5)	4.5 (0.5)	2.3 (3)	4.4 (0.7)	0.399
Creatinine *mg/dL*	0.96 (0.25)	0.83	1.04 (0.3)	0.93 (0.12)	1.08 (0.26)	0.86 (0.25)	1.04 (0.36)	0.489
SMA systolic/diastolic ratio	5.6 (2.5)	5.3	3.6 (1.35)	5.15 (2.36)	6.6 (0.9)	7.4 (3.74)	5.7 (1.04)	0.0249

Characteristics of fontan failure phenotype. Median values are reported with the interquartile range in parentheses. Kruskal-Wallis equality of population was used to determine *P* value. VTI, velocity time integral; MAP, mean arterial pressure; SMA, superior mesenteric artery.

Noninvasive imaging modalities to differentiate these phenotypes do not exist. We propose an algorithm to differentiate these phenotypes by echocardiogram using ejection fraction, velocity time integral, E/E′ ratio and mean arterial blood pressure.

Methods: Twenty-two Fontan patients (68% male, 82% systemic left ventricle) who enrolled in a single center prospective registry had 48 echocardiograms available for review. Patients were categorized into Fontan failure phenotypes based on the algorithm (Figure 1). Velocity time integral, E/E′ ratio and mean arterial blood pressure were used as markers of cardiac output, left atrial pressure and systemic vascular resistance respectively. Chi-square analysis was performed on the characteristics of echocardiogram phenotypes. Eight patients had cardiac catheterization; phenotype by cardiac catheterization phenotype was compared to the patient's nearest temporal echocardiogram. A Kappa analysis for expected agreement was performed.

Results: Fontan type was lateral tunnel (10), atriopulmonary (7), Fontan revision to extracardiac (3), and extracardiac (2). 5 of 8 (62.5%) echocardiogram phenotypes correlated with the most recent cardiac catheterization, which is 3.3 times higher ($P = 0.0002$) than the expected agreement (18.75%). 54% had primarily heart failure phenotypes, 23% had noncardiac failure phenotypes, 21% had mixed cardiac and noncardiac phenotypes and 2% percent had a normal hemodynamics. Fontan Failure phenotype characteristics are described in Table 1. Patients with cardiac and mixed phenotypes were older, and further from their Fontan palliation. Fontan Failure with reduced ejection fraction (type I) had a low superior mesenteric artery systolic/diastolic ratio (SMA s/d ratio).

Conclusion: Fontan failure phenotypes can be differentiated by echocardiogram, including overlapping phenotypes. Cardiac Fontan failure correlates with time since Fontan palliation. Further refinement of the algorithm in a larger cohort with longitudinal follow up is needed to predict outcomes. A noninvasive algorithm to differentiate Fontan phenotypes will aid in guiding management of these complex patients.

11:15 AM – 11:30 AM

Nasal nitric oxide and health-related quality of life in children with congenital heart disease

Rukmalee Vithana, MD[1]; Omar Khalifa, MD[2]; Phillip Adams, DO[3]; Robert McCarter, ScD[4]; Murray Pollack, MD, MBA[5]; Aminata Bangoura, BS[6]; Linda Leatherbury, MD[6]; Joan Sanchez De Toledo, MD, PhD[7]; Cecilia Lo, PhD[2], (1) Pediatric Cardiology/Children's Hospital of Pittsburgh at UPMC, Pittsburgh, PA, (2) The University of Pittsburgh School of Medicine, Pittsburgh, PA, (3) Division of Pediatric Anesthesiology/Department of Anesthesiology/University of Pittsburgh School of Medicine, Pittsburgh, PA, (4) Pediatrics/George Washington University School of Medicine, Washington, DC, (5) Critical Care Medicine/Children's National Medical Center, Washington, DC, (6) Pediatric Cardiology/Children's National Medical Center, Washington, DC, (7) Critical Care Medicine/Children's Hospital of Pittsburgh at UPMC, Pittsburgh, PA

Purpose: Ciliary dysfunction (CD) (abnormal respiratory ciliary motion and low nasal nitric oxide (NO)) is associated with congenital heart disease (CHD) and with increased morbidity and mortality in CHD patients with heterotaxy. Little is known about the health-related quality of life (HRQOL) in CHD patients with CD. Our aim was to assess the impact of low nasal NO on HRQOL in CHD patients.

Methods: In this prospective study, patients at two institutions are being enrolled and evaluated during the preoperative and six months postoperative periods. Nasal NO levels are measured in the preoperative period and classified as low or normal based on recently established 95% prediction intervals ($P < 0.05$) for age-adjusted nasal NO values. We used the Pediatric Quality of Life Inventory (PedsQL), to assess HRQOL in children. Age appropriate PedsQL questionnaires assessed both physical and psychosocial HRQL dimensions. The relationship between nasal NO levels and PedsQL scores was then examined.

Results: Thirty patients were enrolled, ranging from the immediate newborn period to two years of age. Eighteen patients had normal nasal NO, while twelve patients had low nasal NO values. In the pre-operative period, patients with low nasal NO had lower total PedsQL scores (low nasal NO: 88.9 versus normal nasal NO: 97.2, P value=0.02) Psychosocial scores were reduced, with a median score 8.5 points lower in the low nasal NO group (P value = 0.01). Median physical scores were reduced, 6.2 points lower in the low nasal NO group (P value = 0.29). Lower physical scores correlated with lower psychosocial scores ($r2 = 0.80$, $P < 0.05$) in both groups.

Conclusion: CHD patients with low nasal NO have lower total HRQOL scores in the preoperative period. This was associated with significantly lower physical and psychosocial scores, with the lower psychosocial score correlating with the lower physical scores. Further analysis is needed to determine which elements of psychosocial health impact HRQOL and the specific physical symptoms that affect psychosocial health. Insights into factors affecting HRQOL may provide better guidance on the management of CHD patient care to improve their psychological well-being and overall HRQOL.

3:45 PM – 4:00 PM

Coronary imaging utilizing gadolinium cardiac magnetic resonance combined with an inversion recovery gradient echo sequence: utility in congenital heart disease

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Background/Hypothesis: Coronary imaging in congenital heart disease and particularly in pediatrics can be challenging. We hypothesized that utilizing cardiac magnetic resonance (CMR) with an inversion recovery gradient echo sequence (IR-GRE) combined with gadolinium would be successful in visualizing the coronary arteries in the vast majority of cases.

Materials/methods: All patients who presented for CMR and underwent IR-GRE between October 2010 and September 2016 were included. Gadolinium was either "dripped" in over the first third of the sequence or, if utilizing gadofosveset, administered as a bolus and IR-

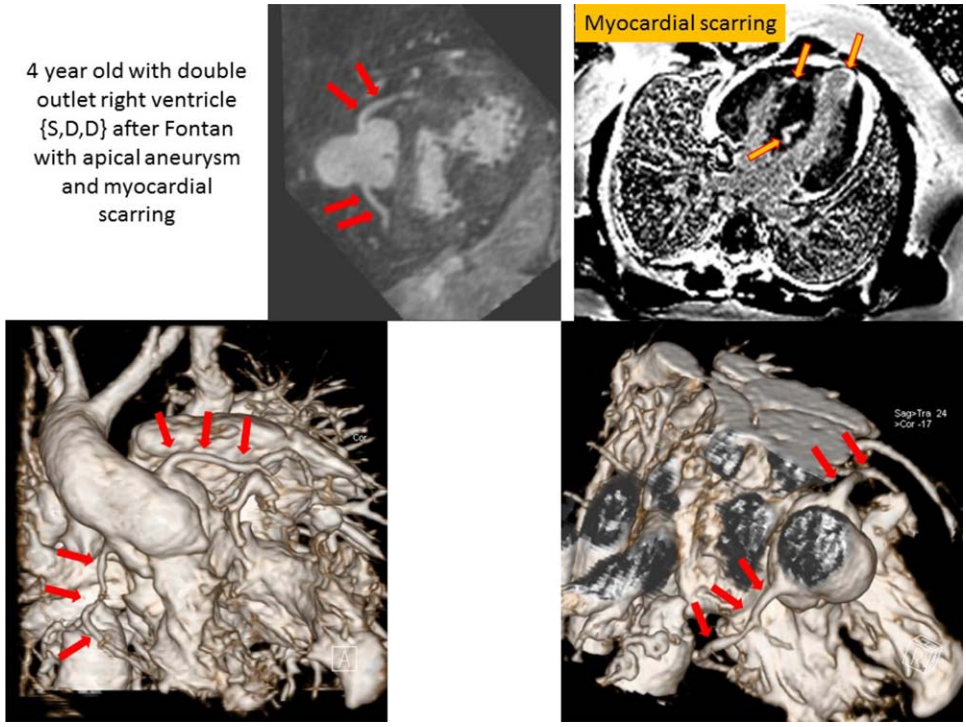

FIGURE 1 Coronary imaging and viability utilizing gadolinium and inversion recovery gradient echo imaging

GRE performed immediately afterwards. Indications were either for coronary imaging or for imaging other structures (eg aorta) and coronaries were visualized.

Results: 1044 patients were found (54% male, age 14.3 ± 12.6 years, range 3 days–44 years) with 720 < 19 years. A whole range of diagnoses were included such as hypoplastic left heart syndrome, tricuspid atresia, transposition of the great arteries, bicuspid aortic valve, suspicion of anomalous coronary to name a few. Proximal coronary arteries were visualized in 987 (94.6%)—see figure. IR-GRE was successful even in patients with braces artifact where routine coronary imaging would fail. Myocardial scarring was able to visualized as well in the same exam—see figure. Of the remaining 57 patients, 28 (49%) were- < 1 year old and 13 (23%) were teenage males; success rate in the pediatric age range was 94%. 117 (12%) presented with a suspicion of an anomalous coronary of which anomalous right coronary from the left sinus was the most common finding (N=85, 72.6%).

Conclusions: CMR utilizing IR-GRE combined with gadolinium has a high success rate (94.6%) in visualizing the coronary arteries in congenital heart disease and in the pediatric age range (94%); myocardial scarring was also visualized. Most failures were due to age < 1 year or being a teenage male.

4:00 PM – 4:15 PM

A feasibility study of implementing electrocardiogram screening in the pediatric home: Addressing provider education as a barrier

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Purpose: Sudden cardiac arrest (SCA) is responsible for over 2000 childhood deaths/year. The structural and electrical cardiac conditions associated with SCA often can be identified by an electrocardiogram (ECG). In our Heart Health Screening Study, we have evaluated various models and locations for ECG screening, including cardiology offices, schools, recreation centers, and churches. This study aims to evaluate the feasibility of using ECG screening in a large urban pediatric practice. Since accurate ECG interpretation is often raised as a limitation, we evaluated the ability of pediatricians and pediatric nurse practitioners to interpret ECGs as normal or abnormal after attending one

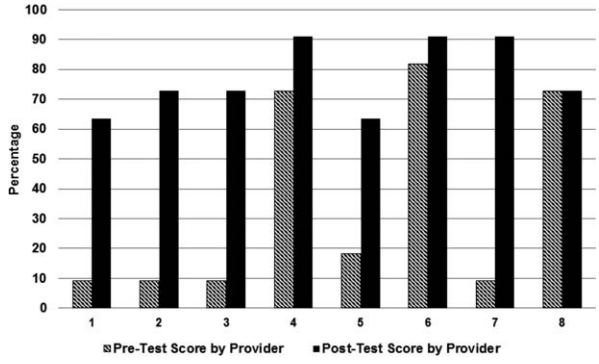

FIGURE 1 Pre- and posttest scores following an ECG Interpretation educational module for correct interpretation of normal vs. abnormal

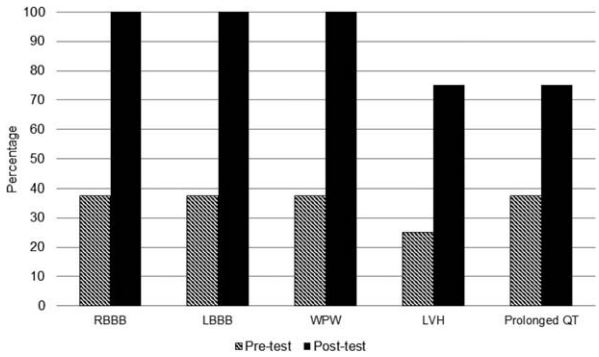

FIGURE 2 Pre- and posttest scores following an ECG interpretation educational module for correct identification of ECG findings

module on ECG interpretation in children and adolescents. We hypothesize that education on ECG interpretation will improve pediatric practitioners' ECG reading skills and thereby increase the ease of using periodic ECGs in the pediatric office to screen for those at potential risk for SCA.

Methods: A one-hour ECG educational module was developed by a pediatric cardiologist to illustrate normal ECGs, normal variants in children of different ages, and abnormal ECGs, including findings consistent with common conditions associated with sudden cardiac arrest. Eleven ECGs, including normals, normal variants, and abnormals, were selected. Prior to the provision of the educational session, 8 providers (6 pediatricians, 2 pediatric nurse practitioners), who practice at a large urban pediatric clinic (65,000 pediatric visits/year), were asked to interpret the 11 ECGs. The providers completed a post-education interpretation of the same 11 ECGs presented in random order. Data were analyzed as a percentage of correct responses. Pre- and post-education scores are presented as mean (SD) and ranges. Statistical analyses included chi-square and student's *t* test.

Results: For correctly interpreting ECGs as normal vs. abnormal, the pretest score was 35.2% ± 33.8% (range 9.1–81.8%) compared to the post-test score of 77.3% ± 11.9% (range 63.6–90.9%; $P = 0.003$) (**Figure 1**). Correct identification of specific ECG findings pre-/posteducation respectively were as follows: right bundle branch block (37.5%, pre-/100%, post-), left bundle branch block (37.5%/100%), Wolff-Parkinson-White pattern (37.5%/100%), left ventricular hypertrophy (25%/75%), and prolonged QTc interval (37.5%/75%) (**Figure 2**). Providers' ability to correctly identify these conditions significantly improved following the educational module ($P < 0.001$).

Conclusion: Education of pediatric practitioners on ECG interpretation significantly improves the ability to accurately distinguish normal from abnormal and to identify specific abnormalities. The lack of skill with ECG interpretation can be addressed by providing practitioners with an ECG educational module and should not be considered as a barrier to implementing screening in pediatric practices or clinics. Our continued research is assessing other potential barriers to ECG screening in this setting including practitioner comfort, patient/family acceptance and effect on patient flow.

4:15 PM – 4:30 PM

Systemic outflow tract pathologies after tetralogy of Fallot repair

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Background: Long term outcomes for patients with tetralogy of Fallot (TOF) have largely focused on right ventricular enlargement, dysfunction and pulmonary valve pathology. However, the systemic outflow tract can be affected in the form of aortic enlargement and aortic regurgitation overtime. There is very limited data, mostly reports and case series about this pathology.

Purpose: To evaluate the prevalence of systemic outflow tract abnormalities after TOF repair.

Methods: Retrospective study of TOF patients who underwent surgical repair at our center. We selected two cohort's; one set of patients who were 4–6 years since primary repair and a second group who were >9 years after primary surgery. All relevant clinical and imaging data was collected.

Results: 50 patients (22 females, 28 males) were included. Mean age at primary surgery was 180 days, mean weight was 6.14 kg. Diagnosis was TOF in 27 patients, TOF with pulmonary atresia in 7, TOF with absent pulmonary valve in 4 and TOF with atrioventricular septal defect in 2 patients. All patients underwent primary repair of the lesion. Mean cross clamp time was 64 minutes while mean bypass time was 103 minutes. Mean time on the ventilator was 67 hours while mean total ICU length of stay was 103 hours. All of the patients had a competent aortic valve at the time of discharge from the primary surgery. Serial echocardiographic evaluations were performed during the follow-up period. At most recent available echocardiograms, the mean weight of the patients was 29 kg (14–57 kg) and mean age was 9.2 years. Echocardiograms at this visit showed mean aortic annulus of 1.72 cm and mean annulus z-score of +1.76 (range −0.5 to +5.6), of which 30% of the patients had z-scores of >+2.5. Similarly, there was significant enlargement of the sinus of Valsalva with mean Z-score of +2.3 (range −0.5 to +5.3) with 30% showing >+2.5 z-scores. There was similar enlargement of the sinotubular junction and the ascending aorta with mean Z-scores of +2.3 and +2.1 respectively. Functional impairment in the form of aortic valve regurgitation was also noted in 16/50 (32%). An MRI was performed in a minority of patients 15/50 (30%) and confirmed the aortic pathology.

Conclusion: Structural and functional systemic outflow tract pathologies are common in patients with TOF repair in medium term follow-up. We anticipate that with longer term follow-up, studies will show a further increment in this pathology and such a study is underway. In the largest studies to date, we show that TOF patients after surgical repair show significant systemic outflow pathologies and warrant close monitoring.

4:30 PM – 4:45 PM

N-terminal pro b-type natriuretic peptide as a surrogate biomarker for the early detection of pulmonary hypertension in preterm infants with bronchopulmonary dysplasia

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Purpose: N-terminal pro b-type natriuretic peptide (NTproBNP) has been proposed as a biomarker to diagnose pulmonary hypertension (PH) in preterm infants (PI) with bronchopulmonary dysplasia (BPD). The relationship between NTproBNP, gestational age (GA) and the timing of increase in the course of BPD associated PH (BPD_PH) is not well documented. The purpose of this study was to compare NTproBNP values in PI with BPD_PH detected by ECHO and in infants with and without BPD at 28, 32 and 36 weeks corrected gestational age (CGA).

Methods: After IRB approval and obtaining maternal consents, PI (birth weight < 1500 gm & < 30 week GA) were included. Infants with known chromosomal anomalies or congenital defects affecting cardiorespiratory system were excluded. PH was defined as ECHO findings of pulmonary artery pressures > 30 mm Hg estimated by tricuspid regurgitant jet velocity, presence of right ventricular hypertrophy and/or flattened interventricular septum. Infants were divided into 3 groups for analyses: BPD group (infants requiring oxygen and/or respiratory support at 36 weeks' CGA with no evidence of PH by ECHO), BPD_PH group (infants requiring oxygen and/or respiratory support at 36 weeks' CGA with ECHO evidence of PH), and no_BPD group (infants not requiring oxygen and/or respiratory support at 36 weeks' CGA with no evidence of PH by ECHO). NTproBNP levels were obtained and ECHO was performed on all subjects at 28, 32 & 36 weeks' CGA. Statistical analyses were carried out using SAS procedures that included general linear models, repeated measures analysis of variance and logistic modeling for receiver operator curve (ROC) generation.

Results: Fifty-four infants were enrolled in the study. The mean GA and birth weight was 27.03 ± 1.06 days and 926 ± 263.7 kg respectively. Eight infants were in the BPD_PH group, 28 were in BPD group and 16 were in no_BPD group. The mean NTproBNP values in the BPD_PH vs BPD group at 28 weeks' CGA was 7805 ± 5933 vs 1686 ± 1968 pg/ml ($P < 0.05$) and in the BPD vs no_BPD group was 1686 ± 1968 vs 703 ± 509 pg/ml ($P < 0.05$). These relationships persisted at 32 and 36 weeks' CGA (Table 1). In a repeated measures analyses of variance general linear model that included GA and NTproBNP values at 28, 32, and 36 weeks' CGA, BPD_PH vs BPD remained significantly associated with NTproBNP values, although the NTproBNP values for BPD_PH did not change significantly over time. ROC curves generated for NTproBNP values at 28 weeks provided a cut-point of 2,329 pg/ml for BPD_PH detection (sensitivity of 87% and specificity of 95%) and a cut-point of 578 pg/ml for BPD detection (sensitivity of 89% and specificity of 68%) (Figure 1).

Conclusion: NTproBNP appears to be a good screening tool to determine the onset of BPD_PH as early as 28 weeks' CGA in PI.

TABLE 1 Mean NTproBNP values in pg/ml (SD) at 28, 32 and 36 weeks' CGA in the BPD_PH, BPD and no_BPD groups

	BPD_PH	BPD	No_BPD
Mean NTproBNP 28 weeks pg/ml (SD)	7805 (5933)	1686 (1968)	703 (509)
Mean NTproBNP 32 weeks pg/ml (SD)	8908 (6980)	2039 (1986)	824 (551)
Mean NTproBNP 36 weeks pg/ml (SD)	9527 (6721)	2740 (2562)	919 (597)

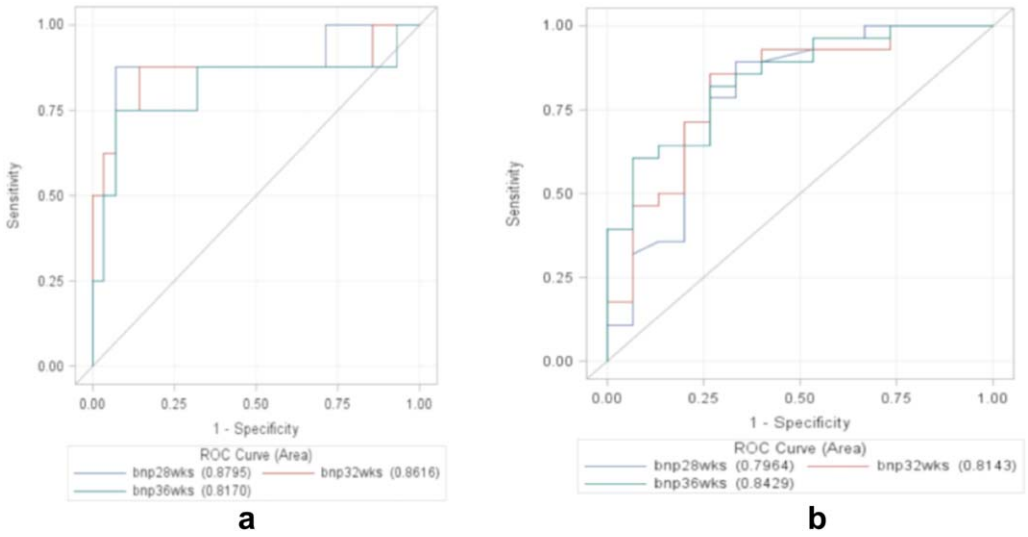

FIGURE 1 ROC curves comparing BPD_PH vs BPD groups (a) and BPD vs no_BPD groups (b) at 28, 32 and 36 weeks' CGA

4:45 PM – 5:00 PM

Cardiac magnetic resonance parameters associated with successful conversion from one to one-and-half or two ventricle circulation in patients with hypoplastic right ventricle

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Background: Some patients with a hypoplastic right ventricle (RV) and univentricular circulation (1V) may be candidates for conversion to either a complete biventricular repair (2V) or a partial repair comprising a bidirectional Glenn shunt with antegrade flow across the right ventricular outflow tract (1.5V). However, determination of adequacy of the RV to support a 1.5V or 2V circulation can be challenging due to the complex structure of the RV, which renders echocardiographic assessment unreliable. Cardiac magnetic resonance (CMR) imaging can provide more precise measures of right ventricular size and relative flow. We sought to identify preoperative CMR findings associated with successful conversion from 1V to either 1.5V or 2V circulation.

Methods: We retrospectively reviewed patients undergoing CMR from January 2000 through November 2016 at our institution and included those with: (1) hypoplastic RV cavity with uncertainty regarding ability to support a 2V circulation, (2) 1V circulation physiology at the time of CMR, (3) CMR evaluation prior to a cardiac surgical or cath procedure intended to convert them from a 1V to a 1.5V or 2V circulation. Patients with conotruncal abnormalities and those who did not undergo volumetric analysis at CMR were excluded. Descriptive statistics are shown as median (range) or mean ± standard deviation. Univariate, nonparametric tests were used to compare predictor variables between patients with a successful versus unsuccessful repair at hospital follow-up. Statistical calculations were performed using SPSS versions 23 (IBM Corporation, USA).

Results: A total of 20 patients met inclusion criteria, of whom 9 (45%) underwent attempted 1.5 V repair and 11 (55%) 2 V repair. In the 2V group, all patients survived with a 2V circulation over a median follow up of 1.6 yrs (0.1–2.7). In the 1.5 V group, 7 (78%) survived with a 1.5 V circulation over a median follow up of 2.7 yrs (1.9-6.9). Higher RV stroke volume at CMR was associated with successful conversion to a 1.5V or 2V circulation (Table 1). There was also a trend towards higher RV end-diastolic volume and right: left ventricular stroke volume ratio among those who underwent successful conversions.

TABLE 1 RV parameters associated with successful conversion to 1.5 V or 2V repair. EDV End-diastolic volume, ESV End-systolic volume, SV Stroke volume, EF ejection fraction, RV Right ventricle, LV Left ventricle

RV parameters	Success (18)	Failure (2)	*P* value
EDV (ml/m2)	41 (11–97)	16 (13–19)	0.07
ESV (ml/m2)	14 (4–58)	10 (6–14)	0.3
SV (ml/m2)	29 (6–47)	6(5–7)	0.01
EF (%)	46 (37–78)	38 (27–50)	0.2
RV:LV SV ratio	0.5 (0.1-1.1)	0.1 (0.08-0.2)	0.06

EDV,end-diastolic volume; ESV, end-systolic volume; SV, stroke volume; EF, ejection fraction; RV, right ventricle; LV, left ventricle.

Conclusion: Among patients with hypoplastic RV and 1V circulation, higher RV stroke volume at CMR was associated with greater likelihood of successful conversion to 1.5V or 2V circulation. Although the sample size is small, this is the first study evaluating CMR parameters that may be associated with successful conversion in this cohort and may assist with preoperative risk stratification.

POSTERS

(1) Acquisition and utilization of healthcare information by congenital heart disease patients and families in the era of social media

Shriprasad Deshpande, MD, MS[1]; Priyanka Patel, MBBS[2], (1) Emory University, Children's Health Care of Atlanta, Sibley Heart Center, ATLANTA, GA, (2) Emory University, Atlanta, GA

Background: Internet and social media (SM) appears to increasingly influence healthcare and healthcare related behavior. Hospital systems are investing significant resources to make healthcare information accessible to these families. However, there is no data available regarding acquisition and utilization of healthcare related information and utilization of social media by parents of congenital heart disease (CHD) patients. These data could direct better harvesting of the potential by healthcare providers to improve health literacy and behavior.

Methods: A survey tool was designed to interview 50 CHD patients and families for this study regarding utilization and acquisition of healthcare information.

Results: (1) Demographics: Age distribution—28 (56%) were < 1 year of age, 11 (22%) were 1–10 years of age, and 11 (22%) were 10–18 years of age. Health literacy was marginal as only 43 (86%) were able to verbalize part of or complete cardiac diagnosis. (2) Seeking information: Only 1 (2%) family sought information online prior to consulting a doctor, compared to 46 (92%) after consultation. Frequency of ongoing search was variable with 25 (50%) reporting more than once a week while the other 50% searched infrequently. 44 (88%) families used Google, 15 (30%) families used the hospital website, 5 (10%) families used resources provided by their doctor, and 6 (12%) families used WebMD as the primary resource to acquire information. (3) Intent: When questioned about the primary intent of using the internet for their child's condition, 44 (88%) families reported seeking medical information related to heart; 23 (46%) families reported seeking ancillary, 5 (10%) families sought nonmedical information; 21 (42%) families reported seeking support off the internet. Using SM platform for sharing information and experience was common: 21 (42%) via posts on Facebook. 16 (32%) families used support groups while 20 (40%) families sought out similar patient families. (4) Impact: 13 (26) families admitted that the social media had influenced their decision making, participation or

treatment option. (5) Doctor-patient relationship: 35 (70%) families used the information they found online to confront or ask questions regarding the treatment to their doctor. Twenty-two (44%) families reported that SM played a part in their confidence in the treatment. Forty-one (82%) of the families felt doctors were more reliable than internet based resources. (6) Using hospital records systems was used by only 34% families.

Conclusion: First study to present data regarding use of SM in pediatric cardiology. SM appears to have significant role in seeking or sharing information, emotional and financial support. Study suggests that lay media rather than edited health information drives behavior and therefore warrants further study to impact behavior in today's social context.

(2) Design and Implementation of a patient passport in an existing pediatric cardiology clinic

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Purpose: Given the young age of the majority of pediatric cardiology patients, parents and other caretakers are often expected to act as medical intermediary between their child and healthcare providers. Relaying an accurate cardiac history to healthcare providers other than the primary pediatric cardiologist can be a major challenge. The purpose of this study is to develop and implement a patient medical passport (PMP) for pediatric cardiology patients. We hypothesized that the PMP would help facilitate both communication and information sharing between caretakers of pediatric cardiac patients and healthcare professionals caring for these patients.

Methods: This pilot study evaluated the benefit of a PMP in reducing the challenges faced by pediatric cardiology patients and their caretakers when interacting with healthcare providers other than their

FIGURE 1 Pediatric Cardiology Patient Passport—blank sample

TABLE 1 Patient demographics. Data shown as raw number and percent of total (n(%))

Number of patients (n)	53
Female	30 (58)
Mean Age +/- SD	5.9+/- 5.2
>1 Cardiac diagnosis	18 (34)
Patients with at least 1 surgery	43 (81)
Patients with 2 or more surgeries	17 (32)
Patients with at least 1 catheterization	25 (47)
Patients with 2 or more catheterizations	16 (30)
Need for SBE prophylaxis	20 (38)
Medical comorbidities	27 (51)
Genetic syndromes	9 (17)
Neurodevelopmental delay	10 (19)
Patients on cardiac or noncardiac medications	22 (42)
Implanted devices	4 (8)
Physical activity restrictions	1(2)
Congenital heart disease (types listed below)	53 (100)
Transposition of the great arteries	9 (17)
Tetralogy of Fallot	9 (17)
Semilunar valve disease (PS,AS)	6 (11)
Hypoplastic left heart syndrome (including variants)	4 (8)
Atrioventricular canal	3 (6)
Ventricular septal defect + atrial septal defect	3 (6)
Atrial septal defect	3 (6)
Ventricular septal defect	2 (4)
Total anomalous pulmonary venous return	2 (4)
Heterotaxy	2 (4)
Tricuspid atresia	2 (4)
Shone's complex	2 (4)
Coarctation of the aorta	1 (2)
Truncus arteriosus	1 (2)
Coronary artery anomalies	1 (2)
Double outlet right ventricle	1 (2)
Hypertrophic obstructive cardiomyopathy	1 (2)
Ebstein's anomaly	1 (2)

primary pediatric cardiologist. Based on the result of a survey of pediatric cardiologists at our institution regarding the most important information to convey to other healthcare professionals, we designed and implemented a PMP in our pediatric cardiology clinic. The trifold PMP is shown in Figure 1. Patients with a diagnosis of congenital heart disease, conduction abnormality, or arrhythmia were included. Patient demographics are listed in Table 1. A patient/caretaker survey was conducted to determine the utilization and utility of the PMP.

Results: Forty pediatric cardiology healthcare providers out of 65 surveyed (62%) responded to a survey regarding the development and implementation of a PMP in our hospital's existing pediatric cardiology clinic. According to pediatric cardiology healthcare providers, the three most difficult aspects of the medical history for patient caretakers to accurately convey are current cardiac anatomy and physiology (85%), information from recent studies (67%), and the name of the lesion/diagnosis (64%). A total of 53 patients were enrolled in the study from October 2016–February 2017. 100% of patients who were recruited agreed to participate in the study. The survey was completed by 49% (n=26) of participants. 88% (n=23) were "extremely likely" or "somewhat likely" to use the PMP in the future. 96% (n=25) said they "definitely would recommend" or "might recommend" the PMP to other pediatric cardiology patients and their families. 88% (n=23) felt the PMP will be helpful in the future when transitioning to adult cardiology. 23% (n=6) of respondents had used the PMP by the time of the survey. Of those who used the PMP, 83% (n=5) felt it made communicating with the healthcare professional easier and 67% (n=4) felt that it improved the healthcare professional's understanding of their child's health condition.

Conclusions: It is feasible and beneficial to introduce a PMP in an outpatient pediatric cardiology setting. Our preliminary results suggest that caretakers found the PMP educational and that it helped facilitate both communication and information sharing with other healthcare professionals.

(3) First line antiarrhythmic therapy for fetal tachyarrhythmia: A systematic review and meta-analysis.

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Background: There has been no consensus on the most effective and best tolerated first line antiarrhythmic treatment for fetal tachyarrhythmia (FT). The purpose of this systematic review and meta-analysis was to compare the efficacy, safety, and fetal-maternal tolerance of first line monotherapies for fetal supraventricular tachycardia (SVT) and atrial flutter (AF).

Methods: A comprehensive search of several databases was conducted, including Medline, Embase, Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, and Scopus through January 2017. Only studies that made a direct comparison between first line treatments of FT were included. We extracted patient characteristics, interventions, and risk of bias indicators. Outcomes of interest were termination of FT, fetal demise, and maternal complications.

Results: Ten studies met inclusion criteria, with 537 patients. 291 patients were treated with digoxin, 137 with flecainide, 102 with sotalol

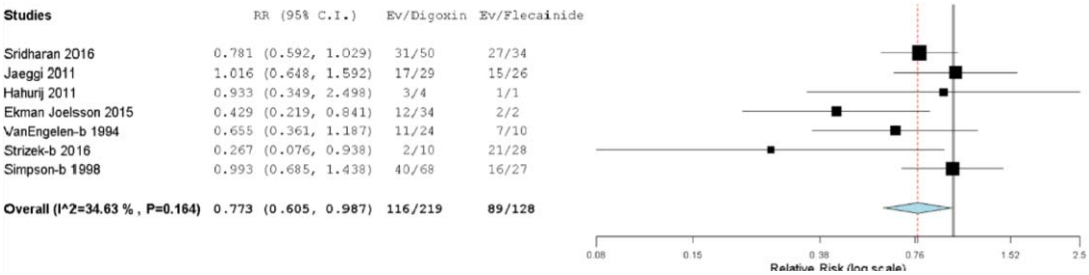

FIGURE 1 SVT termination D vs F. Digoxin versus flecainide in termination of fetal supraventricular tachycardia.SVT in nonhydropic D vs F

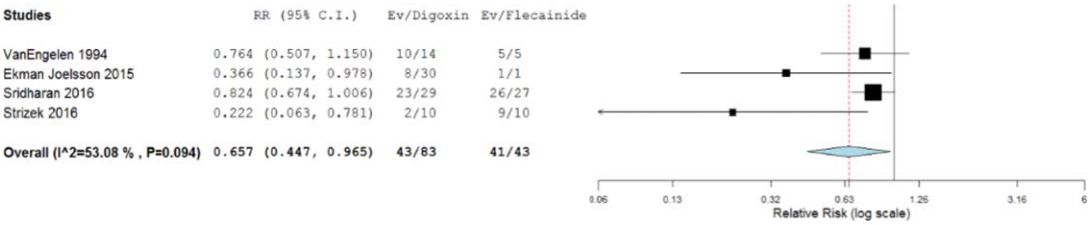

FIGURE 1 SVT in nonhydropic D vs F. Digoxin versus flecainide in termination of supraventricular tachycardia in nonhydropic fetuses

and 7 with amiodarone. Digoxin achieved a lower rate of SVT termination compared to flecainide (OR 0.773, 95% CI 0.605 to 0.987, I^2=34%). There was no difference between digoxin and sotalol (OR 1.009, 95% CI 0.515 to 1.976, I^2=68%) and there was a trend toward higher rate of termination using flecainide compared to sotalol (OR 1.451, 95% CI 0.996 to 2.114, I^2=0%). There was no statistically significant difference between digoxin to sotalol in terminating AF. (OR 0.658, 95% CI 0.240 to 1.803, I^2=48.3%).

In patients without hydrops fetalis, digoxin had a lower rate of tachycardia termination compared to flecainide (OR 0.657, 95% CI 0.447 to 0.965, I^2=53%). There was no difference between digoxin and sotalol. As expected in fetuses with hydrops fetalis, digoxin had lower rates of tachycardia termination compared to flecainide (OR 0.412, 95% CI 0.268 to 0.632, I^2=0%). Few studies reported maternal side effects during FT treatment. There was no significant difference in the incidence of maternal side effects between digoxin and flecainide groups (OR 1.134, 95% CI 0.129 to 9.935, I^2=80.79%). The incidence of maternal side effects was higher in cases treated with digoxin compared to sotalol (OR 3.148, 95% CI 1.468 to 6.751, I^2=0%). There was no difference in fetal demise between flecainide and digoxin (OR 0.767, 95% CI 0.140 to 4.197, I^2=44%).

Conclusions: Flecainide may be more effective treatment compared to digoxin as a first line treatment for fetal SVT. The benefit of flecainide was more pronounced in fetuses with hydrops fetalis. We found no difference in AF termination rate between sotalol and digoxin.

(4) Functional limitations and their associations with special education services and school absenteeism among children with diagnosed heart problems, National Survey of Children with Special Healthcare Needs, 2009–2010

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Background: Young children with congenital heart disease (CHD) have a higher prevalence of cognitive and motor limitations. School-aged children with CHD use special education services more often compared to their peers. There is a paucity of information regarding the functional limitations of school-aged children with CHD and how those limitations are associated with educational needs.

Methods: We used 2009–2010 parent-reported population-based data from the National Survey of Children with Special Healthcare Needs (CSHCN) to examine cognitive and motor limitations and associated educational needs among CSHCN aged 6 to 17 years, stratified by the parental report of a "diagnosed heart problem, including CHD". CSHCN are children with a chronic physical, developmental, behavioral, or emotional condition who require additional health or related services beyond that required by children generally. Parental responses regarding child's functional limitations (difficulty with using hands, self-care, coordination, communication, and learning/concentrating), number of school days missed in the past year, and need for special education services were analyzed. Chi-square tests and multinomial logistic regression, adjusted for demographic characteristics, were used to assess differences in prevalence of functional limitations, by presence of heart problems. Among CSHCN with heart problems, chi-square tests were used to assess whether functional limitations were associated with need for special education services and school absenteeism. Analyses were conducted in SUDAAN to account for complex sampling design and weighted to provide population-based estimates.

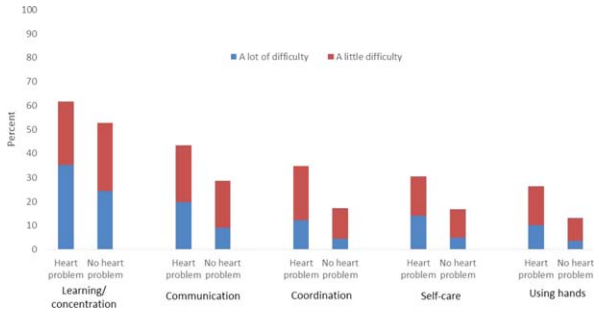

FIGURE 1 Prevalence of functional limitations by presence of diagnosed heart problems among children with special healthcare needs ages 6–17 years, National Survey of Children with Special Healthcare Needs, 2009–2010. chi-square *P* value <0.001 for all

Results: The analytic sample comprised 1,402 CSHCN with heart problems (5.0%) and 28,189 CSHCN without heart problems (95.0%). Prevalence of functional limitations varied by type of limitation and presence of heart problems (Figure). CSHCN with heart problems more commonly had "a lot" of difficulty with using hands (aOR=3.4, 95% CI 2.4, 4.9), self-care (aOR=3.3, 95% CI 2.4, 4.6), coordination (aOR=3.3, 95% CI 2.4, 4.7), communication (aOR=2.7, 95% CI 2.1, 3.6), and learning/concentration (aOR=1.8, 95% CI 1.4, 2.3). CSHCN with heart problems, compared to those without, more commonly needed special education services (45% vs. 29%; $P < 0.0001$) and missed ≥11 days of school in the last year (29% vs. 16%; $P < 0.0001$). Among CSHCN with heart problems, all five functional limitations were associated with need for special education services ($P < 0.0001$), while difficulty with self-care, coordination, and learning/concentrating were associated with school absenteeism ($P < 0.05$).

Conclusion: Among school-aged CSHCN, those with diagnosed heart problems (an unknown proportion of whom had CHD), experienced more functional limitations than their counterparts, and those limitations were associated with need for special education services and school absenteeism.

(5) Granulomatosis with polyangiitis with cardiac involvement: A pediatric case report

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Eosinophilic granulomatosis with polyangiitis (EPGA) is a rare vasculitis with only approximately 50 reported cases in the pediatric literature. Adult data estimates the incidence between 1.3–6.8 per million per year. It is a small and medium sized vessel vasculitis that is diagnosed after meeting four of the six established criteria which include asthma, peripheral eosinophilia, neuropathy, non fixed pulmonary infiltrates, paranasal sinus abnormalities, and extravascular eosinophil infiltration on biopsy. We describe a patient admitted to the CVICU at Texas Children's Hospital after transfer from an outside hospital. A 17-year-old female presented with an EF between 40–45% and 13.3% eosinophils on CBC differential. She also had a history of late onset asthma that worsened despite compliance with therapy and required several doses of systemic corticosteroids, recurrent right hand parasthesias,

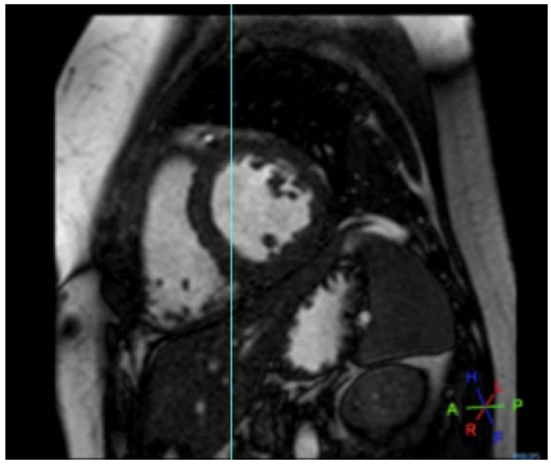

FIGURE 1 Initial Cardiac MRI. Sagittal view demonstrating mild dilation of the left ventricle. EF at this time was noted to be 35%

and a rash that was treated as scabies but recurred despite therapy. Initial laboratory data was remarkable for an elevated WBC count 15.83 103/UL, elevated troponin 6.7 ng/mL, uric acid 8.9 mg/dL, and BNP 2748 pg/mL. ANCAs, MPO/PR-3, and lupus antibodies were negative. Chest CT showed a pericardial effusion, small right-sided pleural effusion, ground glass opacities, and significant dilation of the MPA suggestive of pulmonary hypertension. CVICU care included milrinone drip and anti-arrhythmics. An initial pulse of methylprednisolone (1 g) per recommendation of allergy-immunology resolved the eosinophilia. Histology from an endomyocardial biopsy showed granulomatous myocarditis with lymphohistiocytic infiltrates, focally increased eosinophils, vascular injury with intimal hyperplasia, and focal multinucleated giant cells. Inpatient care included three daily pulses of methylprednisolone and she was started on cyclophosphamide. Corticosteroid infusions improved cardiac function, she was weaned from all drips, and discharged with a daily dose of oral prednisone 60 mg and monthly chemo infusions. EPGA is a rare and under-recognized condition that requires a high degree of suspicion, multidisciplinary care, and often tissue procurement. Therapy protocols are not standardized and depend

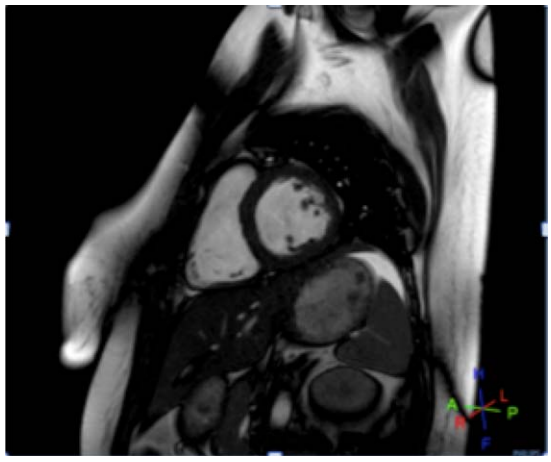

FIGURE 2 Follow Up Cardiac MRI. Sagittal view demonstrating interval improvement in biventricular size, systolic function, and wall motion

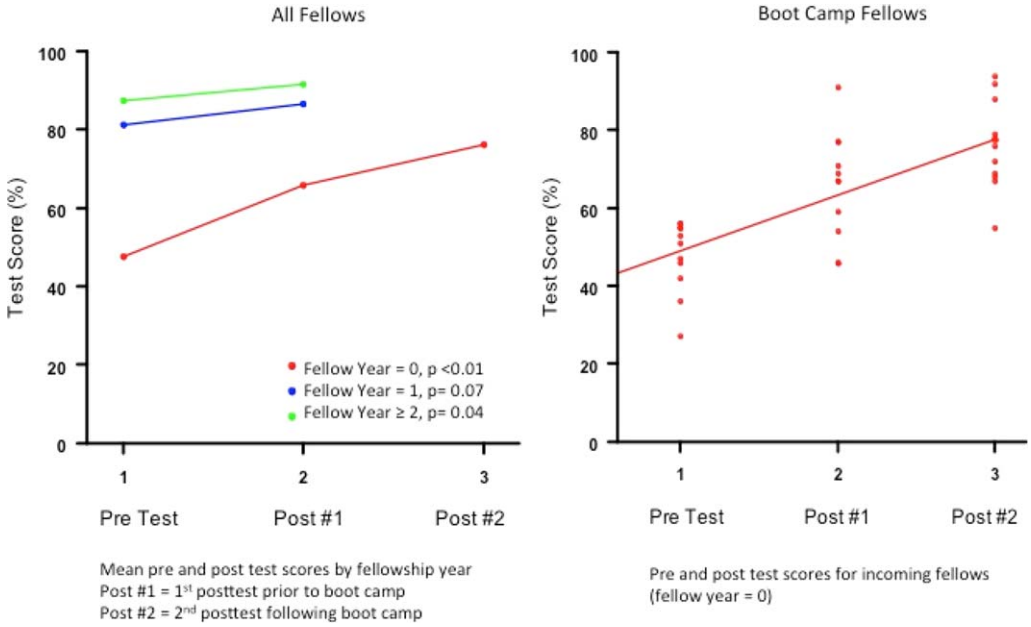

FIGURE 1 Pre- and Posttest scores by fellowship year for online modules and Echo Boot Camp

on degree of organ involvement. Due to the severity of cardiac disease in this case, aggressive therapy with IV corticosteroids and immunosuppression were utilized.

(6) The impact of online learning and hands-on teaching on echocardiography knowledge in pediatric cardiology and ICU fellows

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Background: Mastering pediatric echocardiography is a challenging task for any trainee, requiring both theoretical knowledge and technical skill. We sought to evaluate the incremental impact of online learning in combination with hands-on teaching on the acquisition of echocardiography knowledge and interpretative skills in pediatric cardiology and ICU fellows.

Methods: We prospectively enrolled pediatric cardiology and ICU fellows at Texas Children's Hospital and Lucile Packard Children's Hospital during 2016–2017. All fellows completed online learning modules on our teaching website (www.pedecho.org), covering pediatric echo physics, technique, normal anatomy, atrial septal defects, and ventricular septal defects. A subset of fellows also participated in Echo Boot Camp, a 3-day training program with hands-on workshops and didactic lectures. Outcome measures included an 80 question multiple choice exam administered prior to the online modules, after the online modules, and again after completion of Echo Boot Camp. Paired *t* tests, Wilcoxon signed-rank, and mixed modeling tests were used for analysis.

Results: A total of 27 fellows were enrolled. Our cohort consisted of incoming fellows who recently completed a residency in general pediatrics (n= 11), first year fellows (n=9), and senior fellows (n= 7). The mean pretest score was 69.0% (±20.3%) with a mean posttest score of 79.4% (±15.1%, $P < 0.01$). All subgroups demonstrated improvement in exam scores following online learning. Incoming fellows demonstrated the largest degree of improvement. The subset of incoming fellows who underwent Echo Boot Camp (n=11) demonstrated even further improvement in test scores (Figure 1).

Conclusion: Online learning significantly promotes the acquisition of echocardiography knowledge. Additional hands-on teaching in the form of an Echo Boot Camp can further enhance knowledge and interpretative skills. Both of these learning platforms can therefore serve as powerful and effective tools in fellow education.

(7) Combined atrial deformation indices in the human fetus and relation to biventricular cardiac output

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Purpose: Atrial function is an important contributor to cardiac output (CO) in the postnatal heart, however patterns of phasic atrial function in the fetal heart are unknown. Because fetal circulation is unique, cardiac output is best considered a combination of the right and left ventricles. We sought to measure deformation of the combined atrial mass (excluding the atrial septum) for two physiologic reasons: (1) the atrial septum contributes insignificantly to atrial output, and (2) the widely patent foramen ovale necessitates consideration of a combined atrial output. We then investigated the relationship between reservoir, conduit and contractile components of combined atrial function and the combined CO, with the hypothesis there would be: (a) positive correlation between the two, and (b) increase in both with the progression of gestational age (GA).

TABLE 1 Atrial and Ventricular Deformation and Volumetric Indices

	Total (N=45)	Group 1 (n = 15) GA (16–24 weeks)	Group II (n = 15) GA (24–32 weeks)	Group III (n = 15) GA (32–40 weeks)
GA (weeks)	28.2 ± 6.7	20.6 ± 2.8	28.2 ± 1.7	35.7 ± 2.2
EFW (g)	1485.5 ± 1077.4	426.8 ± 206.2	1270.4 ± 444.5	2759.2 ± 647.7
Atrial reservoir strain εS (%)	36.8 ± 8.1	34.9 ± 8.7	37.8 ± 8.6	37.7 ± 7.0
Atrial conduit strain εE (%)	25.8 ± 7.5	24.1 ± 8.4	26.5 ± 7.2	26.8 ± 7.0
Atrial contractile strain εA (%)	11.0 ± 2.8	10.8 ± 2.9	11.3 ± 2.9	10.8 ± 2.7
$SR_s(s^{-1})$	2.3 ± 0.6	2.4 ± 0.9	2.3 ± 0.5	2.2 ± 0.4
SR_E (s^{-1})	−2.1 ± 0.8	−2.0 ± 0.8	−2.3 ± 1.0	−2.0 ± 0.7
SR_A (s^{-1})	−2.8 ± 1.1	−2.9 ± 1.0	−3.2 ± 1.3	−2.5 ± 1.0
Combined atrial maximal volume (ml)	7.8 ± 5.8	2.1 ± 1.4	7.4 ± 2.7	13-9 ± 4.7
Combined atrial minimal volume (ml)	3.5 ± 2.8	0.9 ± 0.7	3.1 ± 1.2	6.5 ± 2.5
Combined atrial passive filling volume (ml)	4.1 ± 3.3	1.1 ± 0.8	3.8 ± 1.6	7.5 ± 3.0
Combined atrial active filling volume (ml)	3.6 ± 2.8	1.0 ± 0.6	3.5 ± 1.2	6.3 ± 2.7
Combined atrial EF (96)	56.0 ± 7.6	58.1 ± 7.6	56.8 ± 6.2	53.3 ± 8.5
Combined CO (ml)	1026.7 ± 591.9	389.0 ± 178.1	1024.7 ± 282.1	1666.3 ± 340.3

Methods: Forty-five fetal echocardiographic examinations (Vivid E9, GE) were performed in 15 prospectively enrolled volunteer pregnant women. Combined atrial deformation indices were obtained in the apical 4-chamber view using two- dimensional speckle tracking (2DCPA, TomTec). Combined atrial global peak ε (Es indicative of reservoir function), εE (conduit function), εA (contractile function), peak positive, early negative and late negative strain rates (SR_S, SR_E, SR_A), maximal, minimal, passive filling, active filling volumes, and emptying fraction (EF) were measured. Combined CO was calculated from semilunar valve pulse wave Doppler derived stroke volumes. Linear and nonlinear regressions of ε, SR, and volumes were performed against gestational age (GA) and estimated fetal weight (EFW). Relationships of each combined atrial ε and SR component with combined atrial volumes and combined CO were explored.

Results: Maternal age was 29.7 ± 4.2 years, body mass index 25.5 ± 3.3 kg/m^2, GA 28.2 ± 6.7 weeks, and EFW 1.5 ± 1.1 kg. The fetal heart rate was 138.7 ± 10.7bpm. The εS, εE, εA, SR_S, SR_E, SR_A, maximal, minimal, passive filling, active filling volume, atrial EF, and combined CO were 36.8 ± 8.1%, 25.8 ± 7.5%, 11.0 ± 2.8%, 2.3 ± 0.6 s^{-1}, −2.1 ± 0.8 s^{-1}, −2.8 ± 1.1 s^{-1}, 7.8 ± 5.8ml, 3.5 ± 2.8ml, 4.1 ± 3.3ml, 3.6 ± 2.8ml, 56.0 ± 7.6%, and 1027 ± 592 ml (Table 1). There were strong positive nonlinear correlations of combined atrial volumes and combined CO with GA and EFW. There were positive linear correlations of combined atrial εS, εE, εA, SR_E, SR_A with GA and EFW; and weak negative linear correlations of SR_S with GA and EFW. Among the various phasic components of atrial deformation, combined atrial reservoir and conduit strains (εS, εE) correlated modestly with atrial volumes and combined CO ($P < 0.05$).

Conclusion: Determination of fetal combined atrial deformation mechanics across gestation is feasible using 2D-STE. Interpretation of ε and SR must however take GA or EFW into account. Conduit and reservoir strains of the combined atrial mass correlate modestly with combined ventricular cardiac output.

(8) Factors associated with the need for, and the impact of, extracorporeal membrane oxygenation in children with congenital heart disease during admissions for cardiac surgery

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Introduction: Extracorporeal membrane oxygenation (ECMO) is required in a proportion of pediatric patients with congenital heart disease (CHD) during admissions for cardiac surgery (CS), often postoperatively. Factors associated with increased risk of need for ECMO are not well delineated in the proportion of pediatric patients with CHD that require ECMO. This study primarily aimed to determine factors associated with the need for ECMO in children with CHD during admissions for CS. A secondary aim was to determine how ECMO impacted length, cost, and mortality of the admission.

Methods: Data from the 2005 to 2012 iterations of the Kids' Inpatient Database (KIDS) were utilized. Diagnostic and procedural ICD-9 codes were used to identify admissions with CHD under 18 years of age who underwent cardiac surgery. The need for ECMO in these admissions was then identified using the ICD-9 procedure code for ECMO. Univariate analysis was conducted to compare characteristics between admissions with and without the need for ECMO. Next, logistic regression analysis with mortality as the dependent variable was conducted to determine what factors were independently associated with ECMO. Next, regression analyses were conducted with ECMO as an independent variable to determine the impact of ECMO on length of admission, cost of admission, and inpatient mortality.

Results: A total of 46,176 admissions with CHD and CS were included in the final analysis. Of these, 798 (1.7%) required ECMO. The median age of ECMO admissions was 0.5 years compared to 3.3 years for those without ECMO ($P < 0.01$). Median length of admission was longer in those with ECMO (49.2 days versus 12.5 days, $P < 0.01$) as was cost of admission ($727,830 versus $141,524, $P < 0.01$). Mortality was greater in those with ECMO (46.4% versus 1.8%, $P < 0.01$). Logistic regression analysis identified the following as factors associated with increased need for ECMO: decreased age, heart failure, acute kidney injury, arrhythmia, double outlet right ventricle, atrioventricular septal defect, transposition, Ebstein anomaly, hypoplastic left heart syndrome, common arterial trunk, tetralogy of Fallot, coronary anomaly, valvuloplasty, repair of total anomalous pulmonary venous connection, arterial switch, RV to PA conduit placement, and heart transplant ($P < 0.01$). Linear regression analysis demonstrated that ECMO did independently increase length of stay by 17.8 days and cost of stay by approximately $415,917. Logistic regression analysis demonstrated that ECMO independently increased inpatient mortality (odds ratio 22.4, 95% confidence interval 18.1 to 27.7).

Conclusion: Although only a small proportion of pediatric CHD patients undergoing CS required ECMO, these patients required increased resource utilization and have high mortality. Specific cardiac lesions, cardiac surgeries, and comorbidities are associated with increased need for ECMO.

(9) Surgical types & complications as related to timing of repair of tetralogy of Fallot

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Purpose: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease with an estimated incidence of 421 cases per one million live births. Significant controversy exists for children with TOF who are cyanotic and need surgical intervention at a young age. While these children have traditionally been palliated with systemic to pulmonary shunts initially with subsequent complete repair in late infancy, recent studies have shown the feasibility of performing early complete repair. Reported rates of transannular patches can be as high as 83% in neonates and young infants undergoing complete repair. Long term outcomes are less clear, as transannular patches are associated with increased pulmonary insufficiency and may be associated with more frequent reoperation and long term ventricular arrhythmia. There have been limited reports on the modern experience of placement of initial palliative shunt followed by complete repair with regard to rate of valve sparing repairs, pulmonary artery growth, and rate of reoperation.

Methods: We conducted a retrospective study of all TOF patients with pulmonary stenosis (TOF/PS) operated at our hospital from 2009 to 2016. Only patients with early cyanosis who underwent two stage repair were included. Surgical timing, approach of repair, type of repair, and reoperation or interventional catheterization were noted. Echocardiographic measurements of pulmonary valve annulus diameter, main pulmonary artery, right pulmonary artery, and left pulmonary artery diameters were evaluated prior to undergoing palliative shunt placement and again immediately prior to complete repair. Z-scores were obtained for measured diameters and compared using paired student's t-tests.

Results: A total of 31 patients were included who underwent palliative shunt at a median age of 51 (IQR 26–85) days and complete repair at a median of 337 (IQR 301–369) days. Valve sparing repairs were done in 18/31 (58%) of the patients with remaining 13 (42%) patients undergoing transannular patch repair. The z-score of the right pulmonary artery was significantly higher prior to complete repair as compared to z-score prior to palliative shunt ($P = 0.005$). There was also a trend toward higher z-score of the left pulmonary artery prior to complete repair ($P = 0.06$). During median follow up of a 1.1 (IQR 0.11–3.2) years, 5/31 (16%) of patients underwent reoperation (one patient each underwent pulmonary valve repair, resection of infundibular stenosis, conduit placement, main pulmonary arterioplasty, and one with both conduit placement and left pulmonary arterioplasty). Five of 31 (16%) of patients required interventional cardiac catheterization during follow-up.

Conclusion: Staged repair of TOF patients may result in branch pulmonary artery growth, higher rates of valve sparing repairs, and fairly low rate of reoperation in intermediate follow up.

(10) Hepatopathy and PLE in Fontan patients with baffle fenestration or hepatic vein exclusion.

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Purpose: The Fontan procedure is the final surgical palliation for patients with single ventricle. Unfortunately, the long-term "Fontan physiology" is associated with end-organ consequences including hepatopathy. Patients may also develop protein losing enteropathy and plastic bronchitis. In this study, we aimed to describe clinical and laboratory features, including liver pathology, in Fontan patients with either hepatic vein (HV) exclusion or Fontan baffle fenestration (FF).

Methods: Patients who underwent cardiac catherization with transjugular liver-biopsy between 2014 and 2016 were identified from the Nemours/AI DuPont EMR. Demographics, clinical course, hemodynamic data, laboratory data and METAVIR (liver pathology) scoring were reviewed. Statistical analysis was done using Fisher's exact test and Student *t* test.

Results: Twenty-four Fontan patients (10 FF, 14 HV) met inclusion criteria. The most common anatomic diagnosis was hypoplastic left heart syndrome (FF 6, HV 8). Average Fontan duration was 15.72 +/− 3.8 years (FF 13.76 +/− 3.9, HV17.13 +/− 3.5). Average age at

catheterization was 15.8 +/− 3.85 years (FF 14.1 +/− 3.5, HV 17.4 +/− 3.5). There was no significant difference in hemodynamic data. The trans pulmonary gradient measured 3.28 +/- 0.73 mm Hg (FF 3.6 mm Hg +/− 0.73, HV: 2.86 mm Hg +/− 0.81 ($P = 0.28$)), the transhepatic gradient measured 1.42 +/- 0.52 mm Hg (FF 1.55 mm Hg+/- 0.6, HV 1.35 mm Hg +/− 0.43 ($P = 0.4$)). All patients had hepatic fibrosis. There was no significant difference in METAVIR scores, including portal fibrosis 1.77 +/− 0.87 (FF 1.5, HV 1.58 (OR 0.714, CI 0.093-5.301, $P = 1$)), and sinusoidal fibrosis 1.5 +/− 0.51 (FF 1.5, HV 1.58 (OR 0.714, CI 0.093-5.301, $P = 1$)). Both groups had a low rate of sinusoidal dilatation and portal inflammation. No significant difference in liver elastography was seen 2.18 +/− 0.45 m/s (FF 2.29 +/− 0.38 m/s, HV 2.1 +/− 0.49 m/s, $P = 0.92$)). There were no significant differences in total protein (7.03 +/− 0.82), albumin (4.47 +/− 0.59), AST (30.6 +/−8.4), ALT (31.1 +/− 13.6), GGT (65.96 +/- 45.45), and INR (1.14 +/− 0.16). There were 2 patients with protein losing enteropathy (PLE) in the FF group and none in the HV group. There were no patients in either group with plastic bronchitis.

Conclusion: Hepatopathy was universal, but no cirrhosis was detected in this patient population. METAVIR scores and laboratory findings were similar in both groups. There were no patients with PLE in the hepatic vein exclusion group and two patients have PLE in the fenestrated Fontan group. While preliminary, and based on small numbers, these interesting observations require further study.

(11) Interventions and outcomes of hospitalized patients with Turner syndrome and congenital heart disease

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Purpose: To describe the outcomes of hospitalized children with Turner syndrome (TS) undergoing intervention for congenital heart disease (CHD). Methods: We reviewed all hospitalizations of patients identified with TS in the Pediatric Health Information System database from January 2004 to December 2014. All cardiac procedures by type of CHD and their outcomes, adjusted hospitalization cost in US$, length of stay (LOS) and inpatient mortality, were reviewed.

Results: Of 2003 patients with a total of 3722 hospitalizations,1043 (52%) had a diagnosis of CHD. The most frequent CHD were coarctation of aorta (47%), bicuspid aortic valve (12%), hypoplastic left heart syndrome (HLHS) (8%), and aortic stenosis (5%). Surgical and catheterization procedures for most common CHD and their outcomes are listed in Table 1. Overall, 562 patients underwent a total of 2329 cardiac interventions, followed by inpatient death in 40 (2%) patients. The highest mortality was found in subjects with HLHS after Stage 1 palliation (S1P).

Conclusion: This is the largest study of CHD in patients with TS. Patients with HLHS and TS, specifically those who underwent S1P, have the highest hospitalization cost, inpatient mortality, and longest LOS compared to other interventions.

TABLE 1 Procedure outcomes by type of congenita/heart disease

Procedures in patients with HLHS	
Stage 1 palliation (N)	46
Age in days, median (25–75)	8 (5–22)
LOS in days, median (25–75)	39 (27–69)
Cost in US$, median (25–75)	243,207 (159,374-344,835)
Death, N (%)	14 (30)
BDG (N)	23
Age in days, median (25–75)	166 (95–208)
LOS in days, median (25–75)	34 (17–98)
Adjusted cost in US$, median (25–75)	169,937 (122,206–264,740)
Death, N (%)	1 (4)
Fontan (N)	21
Age in years, median (25–75)	3.6 (3.1–4.4)
LOS in days, median (25–75)	10 (8–25)
Adjusted cost in US$, median (25–75)	69,124 (44,744–148,620)
Death, N (%)	1 (5)
Heart transplant (N)	3
Age in days, range	55–2224
LOS in days, range	13–80
Adjusted cost in US$, range	374,099–623,863
Death, N (%)	1 (33)
Procedures in patients with coarctation of the aorta	
Surgical arch repair (N)	339
Age in days, median (25–75)	11 (5–63)
LOS in days, median (25–75)	12 (6–27)
Adjusted cost in US$, median (25–75)	55,677 (31,339–110,221)
Death, N (%)	5 (2)
Catheterization with stent placement (N)	25 (33)
Age in years, median (25–75)	8 (3–14)
LOS, median (25–75)	4 (1–6)
Adjusted cost in US$, median (25–75)	13,942 (11,409–34662)
Death, N (%)	0
Procedures in patients with aortic valve disease (BAV, AS)	
Aortic valve surgery (N)[1]	35
Age, median (25–75)	6.8y (19d-15y)
LOS in days, median (25–75)	8 (5–19)
Adjusted cost in US$, median (25–75)	63,440 (36,241–124.567)
Death, N (%)	0
Percutaneous valvuloplasty, N (%)	24
Age in days, median (25–75)	45 (1–243)
LOS in days, median (25–75)	4 (1–13)
Adjusted cost in US$, median (25–75)	16,851 (7,507–47,995)
Death, N (%)	0

[1]Includes all ICD-9 codes for aortic valvuloplasty and aortlc valve replacement

(12) Using a statewide survey methodology to prioritize pediatric cardiology core content

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Purpose: The American Board of Pediatrics (ABP) has established a 77-page document used to develop the general pediatrics certification exam.[1] We sought to understand differences in perceived importance of specific content within these many objectives from the perspectives

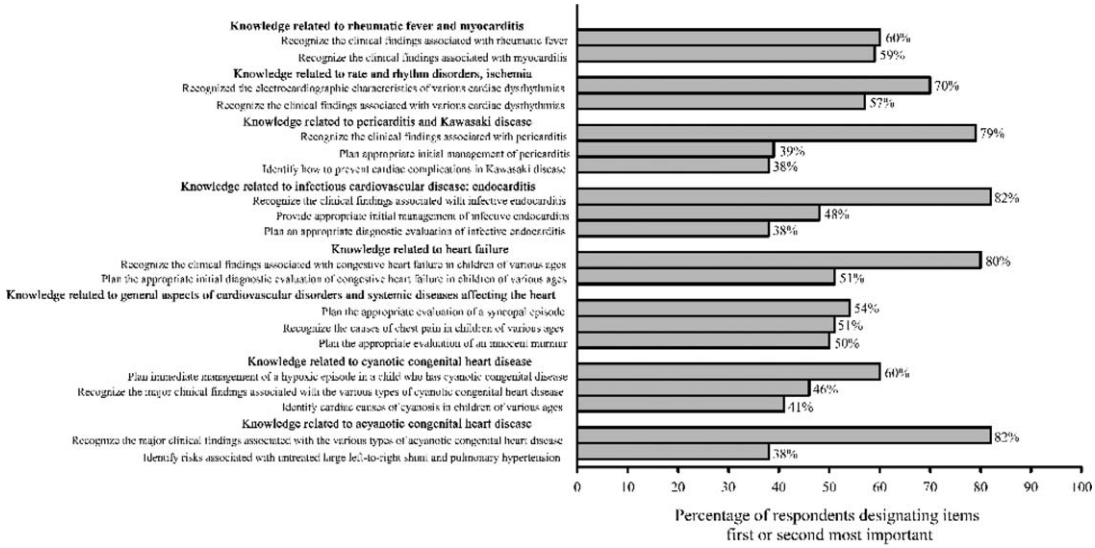

FIGURE 1 2016 ABP content objectives selected most frequently by Kentucky trainees, pediatricians, and pediatric cardiologists (n=136)

of learners and faculty to help medical educators make decisions about how to best structure cardiology curriculum in residency.

Methods: This cross-sectional study used an original, online survey instrument based on the 2016 ABP content specifications for cardiovascular disorders. Participants included learners and faculty from the University of Louisville and University of Kentucky, as well as nonacademic pediatric cardiologists and general pediatricians practicing in Kentucky. We collected both quantitative data (numerical indications of perceived importance) and qualitative data (open-ended replies regarding missing content and difficulty in teaching and learning). Respondents indicated the top two choices of most important items within eight core content areas. Descriptive statistics (frequencies and percentages) were tabulated and compared (Pearson's chi-square) with respect to status as a practitioner (pediatric resident, pediatric fellow, general pediatrician, or pediatric cardiologist). Common themes among open-ended qualitative responses were identified using Pandit's version of Glaser and Strauss Grounded theory (constant comparison).[2]

Results: Of the 136 respondents, 23 (17%) were pediatric residents, 15 (11%) pediatric fellows, 85 (62%) general pediatricians, and 13 (10%) pediatric cardiologists with 80% of attendings having faculty or gratis faculty status. Within this cohort, several notable differences in perceived importance of content emerged as shown in Figure 1 (most selected) and Figure 2 (least selected). For example, in the endocarditis

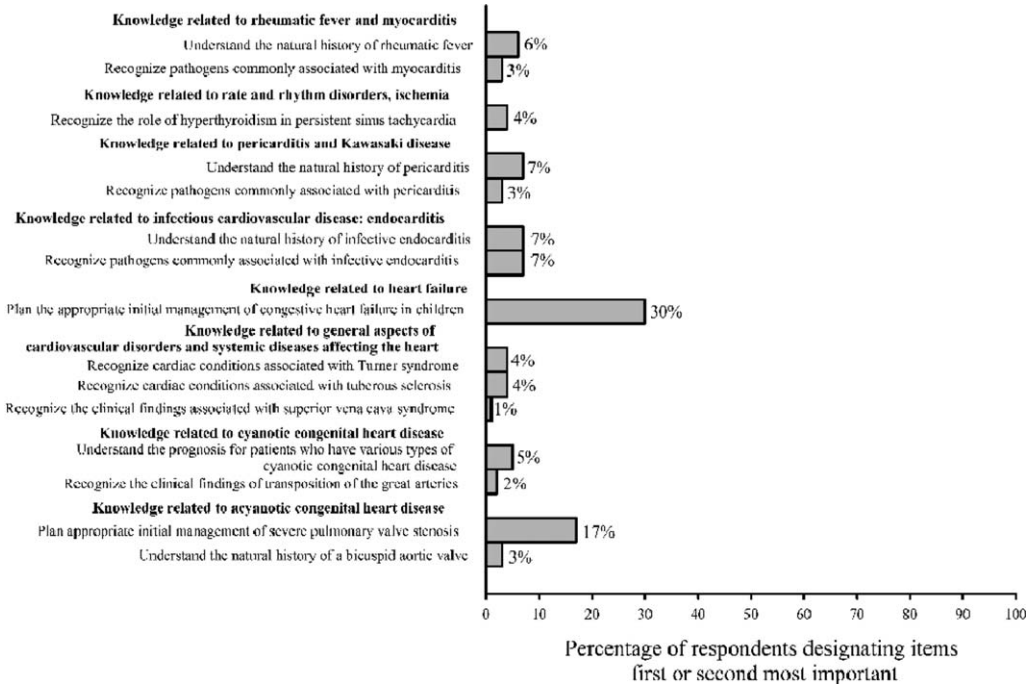

FIGURE 2 2016 ABP content objectives selected least frequently by Kentucky trainees, pediatricians, and pediatric cardiologists (n=136)

content area, 82% selected "recognizing clinical findings of endocarditis" first or second most important vs. 7% prioritizing "understanding the natural history of endocarditis." Naturally occurring "clusters" and "gaps" in respondent-designated importance resulted in no more than three "most selected" objectives per content area. Additionally, groupings of "most selected" items were cohesive, with no more than four themes per category. Objectives included in the "most selected" content related to themes of initial diagnosis (recognition of abnormality/disease), possible emergent/urgent intervention required, planning a workup, and/or building a differential. Open-ended replies regarding missing content highlighted the need to train physicians in "recognizing a normal from an abnormal," "recognition of potentially dangerous conditions," and limitations of screening practices such as newborn pulse oximetry screening.

Conclusion: Understanding the perceived importance of specific 2016 ABP objectives for cardiovascular disorders and missing content from the perspective of learners and faculty can inform curriculum development. This survey methodology could be applied to other specialty-specific curriculum revisions.

References

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2. Pandit NR. The qualitative report. 1996;2(4):1–15.

(13) Trends in the use of open surgeries for atrial septal defect for pediatric patients in the U.S., 2001–2014

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Background: One of the most common congenital heart disease (CHD) subtypes is atrial septal defect (ASD), with an incidence of 1.64 per 1,000 live births (95% CI: 1.61 to 1.67). Without sufficient closure of the foramen ovale, early diagnosis and treatment is often indicated to prevent later complications such as heart failure and death. Historically, standard treatment for patients with ASDs was surgical closure using cardiopulmonary bypass. In recent years, transcatheter ASD closures are increasingly used, instead. Both procedures present with equally favorable long-term outcomes. In this study, we investigated trends in open surgical repairs for ASD in pediatric populations, using nationally representative data in an inpatient setting. We hypothesized that open surgeries decreased due to greater adoption of transcatheter ASD closures.

Design/method: We conducted a serial cross-sectional analysis of hospitalizations for open surgeries (2001–2014) for children (< 18 y/o) with ASD using AHRQ's Nationwide Inpatient Sample (NIS). ICD-9-CM procedure codes 35.51 or 35.61, only when paired with 39.61, were used to identify open surgeries for ASD. Analyses were stratified by age groups (< 1, 1–5, 6–10, 11–14, and >14 y/o). Outcome measures were hospitalization for ASD open closures and cost. Total charges were converted to costs using hospital-specific cost-to-charge ratios, and all costs were adjusted for inflation to reflect 2012 dollars. Descriptive characteristics were stratified and compared by period (2001–2007 "early" and 2008–2014 "latter"). Multivariable regression models, adjusted by both patient and hospital-level factors, were used to explore the associations between the cost and period in order to assess the change in cost in the two periods. Logarithmic cost was used for the analysis because medical costs were not normally distributed. We did not examine transcatheter interventions, assuming a majority was performed at the outpatient setting.

Results: We identified 3,683 hospitalization for open surgeries for ASD open closures, for an incidence rate of 3.57 per million (3.75 in early and 3.40 in latter period). Hospitalization rate was the highest in 1–5 y/o (7.20 in early and 6.98 in latter period). Hospitalization costs were lower in the latter period (median of $18K in early and $25K in latter periods, $P < .01$) Conclusions Hospitalization decreased for the latter study period, suggesting that less open closure surgeries were being performed due to adoption of catheter procedures for eligible patients. Median cost increased for open closure cases for the latter period, suggesting that open closure was assigned to the most complicated cases with more percutaneous closure surgeries performed, resulting in higher individual costs.

(14) Weight as a risk factor for pulmonary hypertensive crisis following pediatric cardiothoracic surgery

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Introduction/Hypothesis: Congenital heart lesions with left to right shunts (High-Qp-Lesions), such as ventricular septal defects, atrioventricular canal, and truncus arteriosus place patients at risk for the development of increased pulmonary vasoreactivity (PVR) if not treated, and life threatening pulmonary hypertensive (PH) crises. Known risk factors for post-op PH include pre-op PH, older age at time or repair, and size of defects. Increased pulmonary blood flow can lead to high output CHF and failure to thrive. Preoperative weight gain in the context of High-Qp-Lesions may be forme fruste of occult elevated PVR. Preoperative weight has not been described as a risk factor for PH. We hypothesized that increased preoperative weight-for-age (WFA-Z) is associated with unanticipated PH crisis requiring inhaled nitric oxide (treatment iNO = iNO-OR, during the operation; rescue iNO = iNO-PICU, in the PICU).

Methods: We conducted a single center, retrospective study using our local Virtual PICU Systems (VPS) database. Significance was defined as $P < 0.05$.

Results: During the study period, there were 493 patients who had congenital heart surgery; 94 (19%) had High-Qp-Lesions. Patients with High-Qp-Lesions had surgery at a younger age (5.6 mos vs 9.9 mos,

$P < 0.01$). Among those with High-Qp-Lesions, 24 (25.5%) were treated with iNO. There was no age difference between treated and untreated. Eleven were treated in the operating room, while 13 received rescue iNO. There was a nonsignificant difference between the iNO-PICU group (WFA-Z = 1.68) and the iNO-OR group (WFA-Z= −0.10).

Conclusions: Preserved preoperative WFA-Z is a potential risk factor for occult PVR, carrying increased risk for unexpected PH crisis following high risk congenital heart surgery. Our small, local sample supports proof-of-concept and merits further investigation with a multicenter analysis.

(15) Pattern of inpatient pediatric cardiology consultations in sub-Saharan Africa

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Background: Malawi is one of the poorest nations in the world, with a human development index of 0.445, ranked 173rd in the world, and an under-5 mortality rate of 63 per 1000 live births. There are no previous studies describing the spectrum of pediatric cardiac disease in an inpatient setting in Malawi.

Methods: A descriptive needs-assessment cohort study based on the clinical experience over a period of 4 weeks was performed at Kamuzu Central Hospital (KCH), a tertiary care hospital in Lilongwe, Malawi. Ethics approval was obtained, and all cardiology consults for patients aged 0–18 years admitted to the children's wards were reviewed. Demographic, anthropometric and clinical information, as well as recommendations made by the cardiology consult team, were assessed and analyzed using SPSS 22.0.

Results: Seventy-three consults and 69 echocardiograms were performed on 71 patients. The population was equitably distributed between genders (35 males, 38 females), and all of them were immunized up to date for age. The median (IQR) age of the cohort was 3.1years (9 mo to 10.5 yrs). About 53% (39/73) had failure to thrive, and 4.1% (3/73) had HIV. Almost 14% (10/73) had Down syndrome, while 4.25 (3/73) had concern for other genetic syndromes on physical exam. About 27% (20/73) of the consults had a prior history of cardiac problems, of whom 18 had had an echocardiogram by a radiologist and only 2 had had an echocardiogram by a cardiologist. The site of the consult was primarily the high acuity emergency zone (61.65, 45/73) and the neonatal intensive care unit (13.7%, 10/73). An echocardiogram was indicated for a majority of the consults (94.5%, 69/73), and a majority of the patients consulted on had cardiac disease (74%, 54/73). Overall 41% (30/73) had acquired heart disease, while 33% (24/73) had congenital heart disease (CHD). On further breakdown, 9.6% (7/73) had rheumatic heart disease, 9.6% (7/73) had cyanotic CHD and 23.3% (17/73) had acyanotic CHD. Symptomatic heart failure (HF) with ejection fraction < 50% was found in 19.2% (14/73) of patients, and pulmonary hypertension was diagnosed in 9.6% (7/73). None of the patients with HF had CHD. There was no significant association of HIV, gender, or failure to thrive on presence of HF. The overall median (IQR) length of admission was 5 days (3 – 10.5). A majority of the patients (60%, 44/73) were discharged from the hospital more than 3 days after the consult, and 5.4% (4/73) died in the hospital admission.

Conclusion: This study provides the first data on the spectrum and burden of pediatric cardiology in an inpatient setting in Malawi in particular, and sub-Saharan Africa in general. The relatively high fraction of patients with HF and the burden of CHD is interesting, and indicates need for further research.

(16) Incidence of Wenckebach in pediatric patients

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Background: Wenckebach is a progressive prolongation of the PR interval followed by a nonconducted P wave. This periodic failure to conduct is due to conduction block within the AV node, superior to the bundle of His. The purpose of this study is to quantify the incidence, timing, duration, and associated symptoms of Wenckebach in pediatric patients, as well as the gender and age distribution of the population in which it occurs.

Methods: A single-center retrospective review of all ambulatory ECG monitors prescribed by pediatric cardiology at a tertiary care referral center [24-hr Holter monitor (Cardiac Diagnostic Services, Inc., Independence, OH, USA) and ZIO XT Patch (iRhythm Technologies, Inc., San Francisco, CA, USA)]. Inclusion criteria: age 0–18 yrs, physical examination by a pediatric cardiologist, and ambulatory ECG monitor prescribed from 12/12-3/17. Exclusion criteria: pacemaker Wenckebach. Episodes of Wenckebach were characterized based on duration, concurrent symptoms within 45 sec of Wenckebach rhythm, and time of initiation. Day and night were defined as 06:00–22:00 and 22:00–06:00, respectively.

Results: Over a 4-year period, 1,558 patients (50.2% male, average age 9.4 yrs) met inclusion criteria. Wenckebach was noted in 72 (4.6%) patients (51.4% male, average 11.6 yrs). Incidence of Wenckebach increased with age, with 56.9% between ages 13–18 yrs ($P < 0.0001$). Indications for ambulatory ECG monitoring in those with Wenckebach were: palpitations (18), chest pain (11), syncope (12), prior arrhythmia diagnosis (8), dizziness (5), abnormal ECG (7), tachycardia (6), and previous cardiac intervention (5). Congenital heart disease was diagnosed in 11 patients (15%). Wenckebach occurred at night in more patients than during day: 33 (45.8%) versus 17 (23.6%) ($P = 0.008$). The incidence rate of Wenckebach was 1.9 episodes/person-day, with 1.1 episodes/person-day at night and 0.8 episodes/person-day during the day. The median time to detection of a Wenckebach episode was 16 hrs (range 1–236 hrs). Median duration of a Wenckebach episode was 16 sec (range 8–56). Event markers for symptoms were triggered by 3

(4.2%) patients during an episode of Wenckebach. These patients had significantly higher average numbers of daytime episodes of Wenckebach (5.7/day) when compared to asymptomatic patients (0.93/day) ($P < 0.001$). Of the vast majority of triggered events (58) in the 3 patients, only 5 correlated with Wenckebach.

Conclusion: Wenckebach is common in the pediatric population, is usually asymptomatic, and occurs more frequently at night. Prevalence of Wenckebach increases with age. Episodes of Wenckebach are short in duration and occur predominantly at night.

(17) Quantifying the prevalence of innocent murmurs throughout childhood

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Purpose: Innocent murmurs are common in the pediatric population; however, their prevalence by age and type is unknown. When murmurs are detected, patients are frequently referred to pediatric cardiology for further assessment. The purpose of this study is to determine the prevalence by age and gender of five innocent heart murmurs in pediatric patients presenting to outpatient cardiology clinic.

Methods: A single-center, retrospective review was conducted of patients presenting to outpatient pediatric cardiology clinic from 11/2016-3/2017 at a tertiary referral center. Inclusion criteria were age $\leq$ 21 yrs and a documented physical exam by a pediatric cardiologist. An innocent murmur was defined as turbulent blood flow in the absence of structural heart disease. Innocent murmurs included Still's murmur, pulmonary ejection murmur, venous hum, peripheral pulmonary stenosis, and carotid bruit. Innocent murmurs were recorded by type in the EMR physical exam template (Allscripts TouchWorks EMR, Version 15.1, 2015, Allscripts Healthcare, LLC, Chicago, IL, USA). Demographic and exam data were extracted by automated data capture.

Results: A total of 2,860 patients (ages 0–21 yrs, 51% male) were seen and 979 had a cardiac murmur by EMR documentation: 35% of all males (M) (average age 8.0 yrs, St.Dev 5.8) and 33% of all females (F) (8.3 yrs, St.Dev 5.7) ($P = 0.29$). Innocent murmurs were documented in 131 (4.6%), including Still's murmur in 35 (2.4%) of all M (5.7 yrs, St. Dev 4.8) and 44 (3.2%) of all F (5.4 yrs, St.Dev 4.7) ($P = 0.25$); pulmonary ejection murmur in 10 (0.7%) of all M (4.9 yrs, St.Dev 5.2) and 19 (1.4%) of all F (6.6 yrs, St.Dev 4.8) ($P = 0.09$); venous hum in 5 (0.3%) of all M (6.0 yrs, St.Dev 5.1) and 7 (0.5%) of all F (5.3 yrs, St.Dev 4.5) ($P = 0.57$); peripheral pulmonary stenosis in 7 (0.5%) of all M (0.11 yrs, St.Dev 0.05) and 4 (0.3%) of all F (0.32 yrs, St.Dev 0.18) ($P = 0.55$); and carotid bruit in none.

Conclusion: Approximately 5% of pediatric patients had a documented innocent murmur. Of the innocent murmurs, Still's murmur is at least 1.5 times more common than the remaining innocent murmurs combined, with no predilection for gender. No carotid bruit was documented. Limitations of this study include appropriate documentation of innocent murmurs in outpatient pediatric cardiology clinic.

(18) Incidence and evaluation of pediatric palpitations

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Purpose: Palpitations are a common complaint in the outpatient pediatric cardiology clinic. The purpose is to determine the incidence and diagnostic yield of tests utilized to diagnose patients palpitations.

Methods: A large tertiary university single-center, retrospective review for all visits to the pediatric cardiology clinic from 01/15–07/15. Inclusion criteria: chief complaint of palpitations, 4–21 years old with complete records of physical exam testing. Palpitations are defined as the sensation of an abnormal, fast or irregular, heart beat in the setting of a normal ECG. Data obtained included demographics, history and physical examination, ECG, echocardiogram, ambulatory heart monitor, treatment and diagnosis.

Results: There were 1,508 new patient visits, and 201 patients met the inclusion criteria. From these 179 (89%) had palpitations and 22 (11%) had a tachyarrhythmia. The incidence of new patients diagnosed with palpitations was 12%. The age distribution for palpitations was: 4–9 years; 39 (21.8%), 10–15 years; 91 (50.8%), and 16–21 years; 49 (27.3%). Palpitations were slightly more common in females, 94 (52.5%). The history was concerning in 18 (10%) patients with previous ED visits, onset with exertion and quick on and off of the episode. ECGs were performed in all the patient and was interpreted as abnormal in 19 (10.6%) patients with left, right or biventricular hypertrophy, abnormal T waves or ectopic beats. Ambulatory ECG monitors was performed on 119 (66.5%) patients and abnormal in 6 (5%) patients with ectopic beats. Echocardiograms were performed in 69 (38.5%) patients and abnormal in 2 (3%) patients, a small pericardial effusion and bicuspid aortic valve.

Conclusion: The overall incidence of new pediatric patients diagnosed with palpitations in an outpatient setting is greater than 10%. Palpitations were slightly more common in females and the incidence increases with age, peaking in the 10–15 age group. The most common testing performed in these patients were ECGs and ambulatory ECG monitors which differentiated palpitations from tachyarrhythmias. An echocardiogram was found to have little benefit.

(19) Determining transition readiness in congenital heart disease: Assessing the utility of the Transition Readiness Questionnaire

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Background: The Transition Readiness Assessment Questionnaire (TRAQ) is a tool commonly used to assess transition readiness in adolescents with chronic diseases. It was previously validated in youth with special health care needs (YSHCN), but no patients with congenital heart disease (CHD) were included in the study. CHD patients may have neurocognitive deficits that impair their ability to understand TRAQ or other disease-specific needs which may make the tool less useful. The objective of this study was to determine if the TRAQ contains language and content suitable for adolescents with CHD.

Methods: The TRAQ was administered to adolescent patients with moderate or complex congenital heart disease. In-depth cognitive behavioral interviews were conducted and qualitatively analyzed to evaluate understanding of the questionnaire items and response options as well as determine any topics adolescents with CHD felt were missing. Results: Adolescents aged 15–20 with moderate to severe CHD were consecutively interviewed until thematic saturation was reached (n=9). Participants were able to accurately define and comprehend the majority of key phrases and concepts contained in the questionnaire, although key concepts such as "self-advocacy" were less consistently understood. They felt the response options were clear and appropriate. Topics that were globally felt to be missing from the TRAQ were related to exercise, psychosocial well-being, knowledge about specific heart conditions, and understanding medical emergencies for patients with CHD.

Conclusion: Our data suggest that while the language and content of TRAQ can be largely understood by adolescents with moderate to severe CHD, there are several knowledge and skill areas specific to CHD that are missing for this particular group of chronic disease adolescents and should be considered for future transition assessments for adolescents with CHD.

(20) Comparison and correlation of mean gradients obtained by Doppler echocardiography to peak-to-peak gradients obtained by cardiac catheterization in patients with arch obstruction

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Purpose: Coarctation of the aorta can be evaluated by transthoracic echocardiography and cardiac catheterization. Prior studies have shown no correlation between gradients obtained by echocardiography and cardiac catheterization. Peak-to-peak gradients obtained by cardiac catheterization (catheterization gradients) are considered the gold standard. We hypothesized that mean gradients obtained by Doppler echocardiography may correlate with catheterization gradients in patients with aortic coarctation.

Methods: We retrospectively reviewed all records from patients with native or recurrent coarctation who had undergone cardiac catheterization at Arkansas Children's Hospital from January 2007 to January 2016. Echocardiographic data and cardiac catheterization data were obtained from the Syngo Dynamics database. Mean and peak instantaneous gradients were measured on the spectral Doppler tracing. Arm-leg blood pressure gradients and patient demographics were culled from the medical records. Univariate summaries were performed for patient demographics. Pearson correlations were calculated to compare Doppler gradients with catheterization gradients.

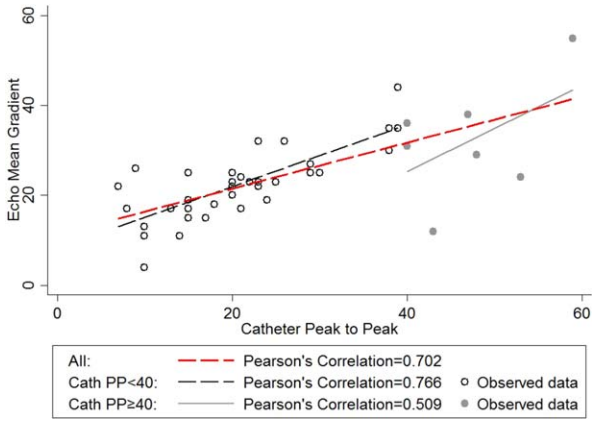

FIGURE 1 Comparison between ECHO mean gradients and catheterization gradients

Results: Forty-four patients (61% male) met inclusion criteria. Median age at echocardiogram was 0.96 years (IQR: 0.36, 9.7). Median time between the echocardiogram and cardiac catheterization was 18 days (1–69 days). Median Doppler mean gradient (23 mm Hg; IQR: 17.8, 29.3) correlated strongly with median catheterization gradient (22 mm Hg; IQR: 15, 32) (r=0.702; $P < 0.001$). Median Doppler peak gradient was less well correlated with median catheterization gradient (r=0.485, $P < 0.001$). As shown in Figure 1 and 2, mean Doppler gradient correlated most strongly when catheterization gradients were < 40 mm Hg, but were not correlated when catheterization gradients were ≥ 40 mm Hg; echo Doppler peak gradients did not correlate with the catheterization gradients.

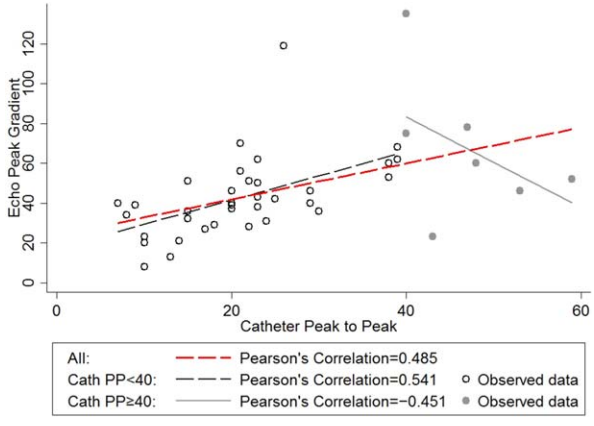

FIGURE 2 Comparison between peak gradients and catheterization gradients

Conclusion: When a coarctation gradient is < 40 mm Hg, Doppler mean gradients are strongly correlated with catheterization gradients. These data suggest mean Doppler gradients can be used to better identify patients who warrant aortic arch intervention.

(21) Evaluation of Egami score in predicting intravenous immunoglobulin resistance among Asian pediatric population with Kawasaki disease.

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Purpose: Kawasaki disease (KD) is an acute febrile illness of unknown aetiology and is known to be complicated by coronary artery (CA) aneurysms. Intravenous immunoglobulin (IVIG) administration in the first 10 days of KD lowers the prevalence of CA aneurysm. However, 13–21% of these patients have persistent fever more than 35.7^{0}C for more than 36 hours or recrudescent after completion of initial IVIG administration, who we considered to have IVIG resistance or refractory KD. Children with IVIG resistance are at higher risk of developing CA aneurysm. Early identification of these high-risk patients would allow for earlier intervention with other therapies such as corticosteroid or anti-tumour necrosis factor (TNF)-α therapy. Thus, risk-scoring algorithms have been developed in Japan to evaluate the likelihood of IVIG resistance at the time of diagnosis. One of this risk scoring algorithms is the Egami score from Japan which has been used by various countries to predict resistance to IVIG but has not been validated in Asian children with KD. The score uses age at diagnosis, days of illness, platelet count, C-reactive protein (CRP), and alanine aminotransferase (ALT) to predict IVIG resistance. Methods We retrospectively review the medical record of patients admitted to 2 major hospital in northern Malaysia, Hospital Sultanah Bahiyah and Hospital Sultan Abdul Halim with the diagnosis of KD from January 2008 to December 2015. Egami score was calculated from demographic and laboratory data to evaluate its ability in predicting IVIG resistance and CA outcome.

Results: Of the 88 patients identified with KD, 12 patients (13.6%) had IVIG resistance. Patient with IVIG resistance has statistically higher WBC (17.9 x109/L vs 14.0 x109/L), higher neutrophil percentage (74.6% vs 58.2%), lower lymphocytes percentage (16.6% vs 31.3%) and higher bilirubin (15.0 umol/L vs 7.0 umol/L). Only 57 patients had the complete set of data required to calculate Egami score. Egami score when used as a predictive tool for IVIG resistance with high risk defined as score ≥ 3, the sensitivity was 66.7%, the specificity was 77.1%, the positive predictive value was 35.3%, and the negative predictive value was 92.5%. Egami score when used as a predictive tool for CA aneurysm showed similar results. Patient with IVIG resistance were at a higher risk of developing CA aneurysm ($P < 0.001$).

Conclusion: There was association between risk group defined by Egami score with IVIG resistance and CA aneurysm in our patient population.

(22) A retrospective study of the safety and efficacy of irrigated catheter ablation in pediatric patients

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Purpose: Standard non-irrigated radiofrequency ablation has been shown to be safe and effective in pediatric patients. Irrigated catheter ablation has not been readily adopted in the pediatric community due

TABLE 1 Comparison of Egami score in IVIG Resistance and IVIG Responder

Characteristic	Total	IVIG resistance	IVIG responder statistics	Z statistics	*P* value
n	57	9	48		
Egami score	1	3	1	−1.922	0.055[a]
Median (IQR)	(1–3)	(1–3)	(1–2)		
Egami score					
0	5 (8.8)	0 (0.0)	5 (10.4)		
1	26 (45.6)	3 (33.3)	23 (47.9)		
2	9 (15.8)	0 (0·0)	9 (18.8)		
3	15 (26.3)	6 (66.7))	9 (18.8)		
4	2 (3.5)	0 (0.0)	2 (4.2)		
Low risk (0–2)	40 (70.2)	3 (33.3)	37 (l77.1)[b]		0.015[d]
High Risk (≥ **3)**	17 (29.8)	*6 (66.7)*[c]	11 (22.9)		

[a]Mann-Whitney test.
[b]Specificity.
[c]Sensitivity.
[d]Fisher's exact test.

to a lack of data in this population. We aim to report demographic, safety, and efficacy data regarding use of irrigated catheters in a pediatric population.

Methods: Reviewed the charts of all patients (< 21 years old) in a single pediatric center who underwent catheter ablation from January 2013–April 2016. Analyzed demographic, efficacy, and safety data from the time of procedure until last available follow-up visit. Data analysis used SAS software.

Results: Thirty-six subjects were analyzed. Median age=14 years old (range= 6–19), 56% male, median weight= 58 kg (range= 21–116 kg). None of our patients had a history of significant congenital heart disease or open-heart surgery. Irrigated ablation performed for accessory pathways (n= 32), atrial tachycardia (n=2), and PVCs/VT (n= 2). No patients with AVNRT, anteroseptal or para-Hisian pathways were ablated using this technique. We used conventional irrigated (n= 29) and contact force irrigated (n=7) catheters. Median number of lesions was 7 (range 0–18). In successful ablations of patients with atrial tachycardia or accessory pathways (n=32), our recurrence rate was 0% with standard irrigated catheters (n=25) and 29% with contact force catheters (n=7). This demonstrated a significant difference in outcome ($P = 0.03$, Fisher's exact test). There were no major complications. One patient developed Wenckebach with 2:1 block during a lesion with subsequent resolution with termination of lesion. No known perforations or steam pops occurred. No significant associations between age or weight and outcomes (recurrence, complications) were noted.

Conclusion: Irrigated catheter ablation can be performed safely and effectively in pediatric patients. The significant recurrence difference for contact force versus conventional irrigated ablation is likely multifactorial. Further investigation is warranted for the application of current contact force catheter platforms for SVT ablations in pediatric patients.

(23) Compliance with appropriate use criteria for pediatric echocardiography: Experience in a single center with an open echocardiography lab

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Purpose: Recommendations for the appropriate use of echocardiography in outpatient pediatrics were published by the American College of Cardiology in conjunction with the American Academy of Pediatrics and other groups in 2014. The purpose of this quality improvement study is to determine the frequency of adherence to the appropriate use criteria in an open pediatric echocardiography lab. The secondary objective of this study is to assess the applicability of these criteria to the inpatient setting.

Methods: This is a single center prospective cohort study of patients with a first-time transthoracic echocardiogram performed at Nationwide Children's Hospital from April–July 2016. Medical record review included murmur or chest pain description, medical history, family history, electrocardiogram findings, and blood culture results. Two cardiologists graded each echocardiogram according to the ACC criteria: Appropriate, May Be Appropriate, and Rarely Appropriate. A third cardiologist reviewed any cases without an initial agreement. Abnormal echocardiogram findings were graded as incidental, mildly abnormal, moderately abnormal, and severely abnormal. Chi-square, Fisher's exact test, and chi-square tests for trend were used for statistical analysis with $P < 0.05$ significant.

Results: Five hundred thirty patients, 30% inpatient, were included in the final analysis. Twenty-nine patients were excluded for lack of clinical information related to appropriateness grading. Fifty-six percent of studies were ordered by a cardiologist. Seventy-one percent of patients had a single indication and 29% had multiple indications for echocardiogram, with abnormal electrocardiogram (31%) and murmur (24%) most common. Echocardiogram indications were graded as Appropriate in 81%, May Be Appropriate in 7%, and Rarely Appropriate in 12% of patients. Thirty percent of studies were abnormal: 12 (8%) incidental findings, 92 (58%) mildly abnormal, 54 (34%) moderately abnormal, and 1 (1%) severely abnormal. Ninety three percent of patients with abnormal echocardiograms had an appropriately ordered test. Patient, clinician, and location characteristics associated with an appropriately ordered tests included: cardiologist ordered ($P < 0.003$), inpatient ($P = 0.02$), multiple indications ($P = 0.003$), and abnormal electrocardiogram ($P < 0.0001$). Abnormalities were more likely to be found in echocardiograms that were appropriately ordered ($P < 0.0001$).

Conclusion: Our study demonstrates the need for continued adherence to the appropriate use criteria for initial echocardiograms. Our study shows cardiologists at our institution are highly likely to adhere to the appropriate use criteria. We demonstrate that by adherence to the appropriate use criteria, the majority of patients with abnormal findings will be identified. Indications of murmur, abnormal family history, and syncope were the indications most likely to be classified as rarely appropriate suggesting the continued importance of a thorough history and physical exam for patients.

(24) The impact of multisystem organ dysfunction in pediatric patients during admissions for cardiac surgery

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Purpose: Multiorgan system dysfunction syndrome (MODS) can be seen after pediatric cardiac surgery. Risk factors for MODS have not previously been well delineated and the impact of MODS on admission length and cost are not described. This study identified risk factors for MODS and describes the impact of MODS on cost, length of stay and inpatient mortality.

Methods: The Kids Inpatient Database (KIDS) was used for this analysis and patients under the age of 18 who underwent cardiac surgery between 1995–2012 were included. Admissions with MODS were

identified using ICD-9 code 995.92. Next, the presence of heart failure, respiratory failure, liver insufficiency, and acute kidney injury (AKI) were obtained for each patient. Patients with ≥2 of these diagnoses were labeled as having MODS. Univariate and multivariate analysis were then conducted to determine risk factors for MODS and the impact of MODS on admission length and cost.

Results: A total of 46,172 admissions were included in the analysis. 222 (0.5%) admissions were explicitly coded as having MODS but using the composite method led to identification of an additional 1,010 (82% of total MODS admissions by composite method) admissions. These 1,232 (2.7%) admissions were then used for further analysis. Heart failure was present in 91%, respiratory failure in 74%, AKI in 40%, and liver failure in 1%. There were 1,145 (93%) who had failure of 2 systems, 86 with failure of 3 (7=6.9%), and 1 (0.1%) with failure of 4. Regression analysis demonstrated that every decrease in age by 1 year (odds ratio 1.1, 95% confidence interval 1.1 to 1.2) and hypoplastic left heart syndrome (OR 1.9, 95%CI 1.2 to 3.1) were associated with increased risk of MODS. No specific operative procedures were associated with an increased risk of MODS as demonstrated by regression analyses. Further regression analyses demonstrated that the presence of MODS increased the median length of stay by approximately 25.7 days ($P < 0.001$), median total charges of admission by $313,793 ($P < 0.001$), and the odds of inpatient mortality 7.6-fold ($P < 0.001$). Risk of mortality was additive with each additional system failure except for liver failure which was not independently associated with increased mortality. Heart failure independently increased the odds of mortality by 1.3, AKI independently by 14.6, and respiratory failure independently by 2.9.

Conclusion: MODS is under-coded in this administrative database and has a frequency of at least 2.7% in pediatric patients during admissions for cardiac surgery. There are relatively few risk factors associated with MODS but this study demonstrated that MODS increases the length, cost, and mortality of admissions for pediatric heart surgery.

(25) Prevalence of hypotension in adolescents

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Background: Hypotension is a widely used but poorly defined clinical entity. Currently accepted definitions of hypotension are the Pediatric Advanced Life Support (PALS) definition, the Brain Trauma Foundation definition, and the International Pediatric Sepsis Consensus Conference. However, its prevalence has not been defined.

Objective: To determine the prevalence of hypotension in school aged children.

Study Design: This study is a retrospective analysis of data from a series of cross-sectional school-based blood pressure screenings completed by the Houston Pediatric and Adolescent Hypertension Program (HPAHP) at the UTHealth at Houston. Screenings were performed in 21 urban and suburban middle and high schools in the Houston area from 2001 to 2012. We restricted the data set to include subjects with three systolic blood pressure (SBP) measurements performed using a Spacelabs 90217 oscillometric monitor, who did not report taking hypertensive medication, and were not missing demographic information. We defined hypotension based on 2 criteria: average 1st-3rd SBP less than the 5th percentile for height, age and gender. Within these definitions we also restricted the hypotension definition to subjects with average SBP below 90mm Hg. Stata 13 SE was used for all data analysis.

Results: Of the total 22, 382 children enrolled, 15,114 children met inclusion criteria: 52% females, mean age 13.6 years (range 10.1–19.9). Racial distribution reflected that of Houston: 35% White, 32% Hispanic, 23% Black, 8% Asian, and 2% other/unknown. Seventy one (0.5%) children had an average SBP reading less than the 5th percentile and 112 (0.7%) had an average SBP less than the 5th percentile or 90 mm Hg. Compared to students without hypotension, students with hypotension defined by only the 5th percentile were older (14.3 vs 13.6 years, *t* test $P = 0.0001$); they also tended to be more female, though not significantly (61% vs 39%, Chi^2 *P* value=0.134).

Conclusion: Although not as prevalent as hypertension in this age group (generally 2–5%), hypotension is a clinically significant entity that is seen in 0.5–0.7% of the general school-aged population.

(26) Pericardial effusion in pediatric patients: Etiologies and risk factors for mortality

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TABLE 1 Selected diagnostic categories most frequently associated with pericardial effusion and the leading diagnoses in each

Condition	Number (% of total)
All autoimmune or inflammatory disorders	1845 (19)
Connective tissue inflammatory disease	812 (8)
Polyarteritis nodosa and allied conditions	559 (6)
All Viral	753 (8)
Epstein-Barr	250 (3)
Adenovirus	179 (2)
All bacterial	1283 (13)
Staphylococcus	776 (8)
Escherichia coli	191 (2)
All other infections	792 (8)
Candida	605 (6)
All renal disease	570 (6)
Chronic renal failure	511 (5)
Neoplastic disease	2368 (24)

TABLE 2 Risk factors for in-hospital mortality in patients admitted with pericardial effusion

Risk factor	Odds of death	95% confidence interval	*P* value
Neoplasm	2.75	2.29, 3.30	<0.001
Renal disease	2.38	1.78, 3.20	<0.001
Viral infection	1.61	1.25, 2.08	<0.001
Bacterial infection	0.71	0.57, 0.88	0.002
Autoimmune/ inflammatory disease	0.60	0.45, 0.81	<0.001
$10,000 decrease in median household income	1.05	1.01, 1.10	0.045

Purpose: Little is known about the causes and risks of pericardial effusion (PCE) in pediatric patients. We sought to assess the diagnoses most frequently associated with admissions for PCE, and to determine if certain conditions were associated with higher mortality.

Methods: Nationally distributed data from 44 pediatric hospitals in the 2004 to 2015 Pediatric Health Information System database were used to identify patients with hospital admissions for International Classification of Disease 9 (ICD-9) codes for pericardial effusion (ICD-9 codes 423.1 and 420.90) and/or cardiac tamponade (ICD-9 code 423.3). Primary and secondary diagnoses, length of stay, and hospital costs in 2015 $were determined. Children with congenital heart disease were excluded. ICD-9 codes for potential causes of PCE were grouped into 5 categories: autoimmune/inflammatory disease, viral infection, bacterial infection, renal disease, and neoplasm. Risk of mortality was evaluated using multivariate analysis of the following variables: admission year, age at admission, gender, race, median household income, mechanical ventilation, neoplasm, renal disease, viral infection, bacterial infection, other infection (nonbacterial/nonviral), and autoimmune/inflammatory disease.

Results: There were 9927 patients who met inclusion criteria (52% male). Total discharge mortality was 8% (n = 835). Median age at admission was 12 years (Interquartile Range [IQR] 4 to 16 years). Median length of stay was 9 days (IQR 4–22). Pericardiocentesis was performed in 1520 patients (15%). Median hospital cost was $33680. Conditions most frequently associated with PCE are listed in Table 1. Risk factors for in-hospital mortality are in Table 2.

Conclusion: Admission for PCE in pediatric patients is associated with high mortality. Patients with a viral infection, neoplasm, or renal disease are at particularly high risk, whereas those with bacterial infections have lower risk.

(27) Renal dysfunction is associated with elevated central venous pressure in patients with Fontan circulation

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Purpose: Elevated central venous pressure (CVP) has deleterious effects on several organ systems in patients with Fontan circulation. However, the relationship between CVP and kidney function has not been assessed in patients with Fontan circulation.

Methods: Patients with Fontan circulation whose hemodynamics were assessed by catheterization and had a serum creatinine within 72 hours prior to the procedure were included for analysis. Patients with primary kidney disease were excluded. Estimated glomerular filtrate rate (eGFR) was calculated by "bedside Schwartz" equation in children (< 18 years) and Modification of Diet in Renal Disease (MDRD) equation in adults. Renal dysfunction (RD) was defined by eGFR < 90 or = or > 150 mL/min/1.73m^2. Fontan patients with and without RD were compared based on demographics, comorbidities, medication use, echocardiographic findings, hemodynamics assessed at time of catheterization, and laboratory testing values. Statistical analysis performed included Student's *t* test, Wilcoxon rank-sum and chi-square tests.

Results: Of 956 patients with Fontan circulation, 67 met inclusion criteria (56 children) and 24 patients (19 children) had RD. Among those with RD, eGFR was > 150 in 9 (38%), 60–89 in 13 (54%), 45–59 in 1 (4%) and 30–45 in 1 (4%). Differences in demographics did not reach the level of statistical difference. However, African Americans constituted a higher percentage of those with RD than those without RD at 20.8% and 4.7%, respectively. Additionally, there appeared to be a longer median duration of Fontan

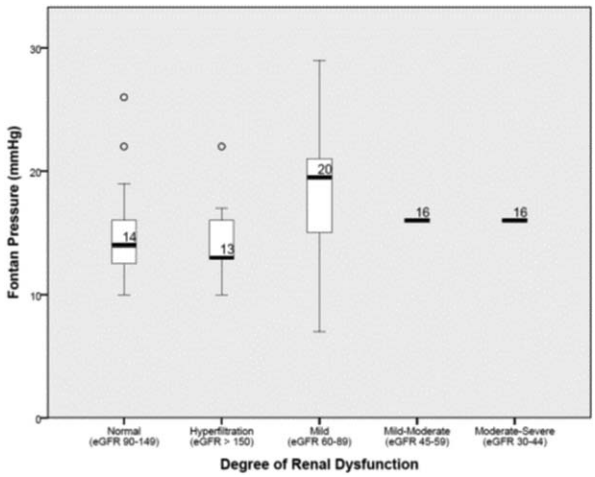

FIGURE 1 Relationship of Central Venous Pressure and Renal Dysfunction. Renal function is categorized by estimated glomerular filtration rate (eGFR) into normal function (90–149 mL/min/1.73m2), hyperfiltration (>150 mL/min/1.73m^2), mild dysfunction (60–89 mL/min/1.73m^2), mild to moderate dysfunction (45–59 mL/min/1.73m^2), and moderate to severe dysfunction (30–44 mL/min/1.73m^2)

circulation in those with RD (6.2 years) compare to those without RD (1.7 years) but this finding did not reach the level of statistical significance. No comorbidities or medications were correlated with renal function. Findings echocardiogram including: atrioventricular valve (AVV), (neo)aortic valve, and ventricular function were not statistically different between groups. Other hemodynamic parameters measured or calculated on cardiac catheterization were not statistically different including: pulmonary vascular resistance (PVR), cardiac output or ventricular end-diastolic pressure.

Conclusions: The data from this study indicate that renal dysfunction in patients with Fontan circulation is associated with increased CVP and not associated with ventricular function or cardiac output. Potentially, impaired kidney function may serve as a surrogate marker for CVP. Continued investigation of the effects of venous congestion on the renal system in patients with Fontan circulation is warranted.

(28) Echocardiography in the setting of an abnormal electrocardiogram: Is it required?

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Purpose: An electrocardiogram (ECG) is a commonly ordered screening test for evaluation of congenital heart disease. An abnormal ECG often prompts referral for an echocardiogram to rule out heart disease. The purpose of this study was to determine if an echocardiogram is warranted in the setting of an abnormal ECG, and the correlation of abnormal ECG findings with subsequent echocardiogram findings as well as with the physical examination (PE) performed by a pediatric cardiologist.

Methods: A single-center retrospective study was performed to identify all echocardiograms done for the indication of an abnormal ECG, between 3/2015 and 3/2017, in patients 1–18 years of age. Exclusion criteria were, known cardiac disease, chronic systemic illness, hypertension, syndromes with known association with heart disease and patients taking medications known to cause ECG abnormalities. PE and the reported ECG findings for the patients included in the analysis were recorded. Patients presenting with symptoms suggestive of a potential heart disease were excluded from the analysis. For the purpose of this study, patent foramen ovale was considered a normal echocardiographic finding. Data were analyzed to evaluate the correlation between an abnormal ECG and echocardiographic findings as well as the correlation between an abnormal PE and echocardiographic findings.

Results: There were 152 patients that met the inclusion criteria. The ECG findings of these patients were reported as isolated left ventricular hypertrophy (40%), isolated right ventricular hypertrophy (10%), left axis deviation (10%), biventricular hypertrophy (9%), and incomplete/complete right bundle branch block (11%). All other ECG abnormalities occurred with a combined prevalence of 20%. An abnormal echocardiogram was found in 22% of the cohort, and within this bracket, 40% were found to have diagnoses which were unrelated to the ECG findings. The probability of an abnormal echocardiogram amongst those with a normal PE was 10%, whereas, the probability of abnormal echocardiographic findings amongst those with an abnormal PE was 47%. There was a strong association between PE and echocardiographic findings [chi-square (1df) = 14.22, P value < 0.001].

Conclusion: ECG is a screening test and while interpreting it, PE should be taken into consideration to form a differential diagnosis. In the presence of an abnormal ECG and a normal PE, echocardiogram has a low yield and should be utilized judiciously.

(29) High prevalence of feeding disorders in young children with congenital heart disease

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Background: Children with congenital heart disease (CHD) are at risk for long-term feeding disorder. Factors that influence feeding outcomes for children with CHD and the relationship of neurodevelopmental (ND) outcomes to feeding status are poorly understood. The objective of this project was to determine which children with CHD are at risk for long-term feeding disorder leading to delayed achievement of feeding milestones, need for feeding tube supplementation and/or developmental delay.

Methods: A retrospective descriptive study to describe feeding experiences and developmental outcomes in children with CHD was completed. Children who had undergone ND evaluation were included. Data on achievement of feeding milestones, feeding skill progression, growth and ND outcomes were collected.

Results: Chart review was completed for 176 children between the ages of 6-and 24-months of age. This cohort was grouped into the following categories: 65% (114/176) never tube fed; 20% (36/176) always tube fed; 8% (14/176) tube fed at 1st hospital discharge but tube feedings were discontinued before 24 mos; and 7% (12/176) orally fed at 1st hospital discharge but tube feedings were required at some point before 24 mos. 538 visits were examined. At 24 mos, 27% (48/176) of children remained feeding tube dependent. More than half of all children in this age range were reported to have feeding disorder regardless of tube dependency. 44% of children who did not require tube feeding reported feeding disorder and failed to achieve expected feeding milestones. Feeding disorders increased with age in children who never received outpatient tube feeding. This is the only group where feeding disorder increased. In children with ND delay the

percentage of children with feeding disorder was twice as high as those without ND delay (87% vs. 40%, $P < 0.01$). Feeding disorders were more common in females, 62% vs. 45% ($P = 0.02$) and in patients with other medical or genetic comorbidities, 69% vs. 40% ($P < 0.001$). There was no difference in feeding disorders by prematurity, race or diagnostic category.

Conclusions: Feeding disorders in children with CHD are not transient and need to be closely monitored and treated. Some children without need for tube supplementation present with chronic feeding disorder and delayed achievement of feeding milestones. The relationship of feeding status to developmental outcomes requires further study.