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WILEY Congenital Heart Disease

American Academy of Pediatrics

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Section on Cardiology and Cardiac Surgery Annual Meeting November 2-4, 2018 Orlando, Florida QI Abstract Presentations:

QI Abstract Presentations: Friday, November 2, 2018: 11:00 AM-12:00 PM

Improving compliance with dyslipidemia screening guidelines in outpatient pediatric cardiology clinic

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Purpose: Atherosclerotic cardiovascular disease begins in childhood. The American Academy of Pediatrics (AAP) has endorsed Guidelines for Cardiovascular Risk Reduction in Childhood and Adolescence. These guidelines recommend universal hyperlipidemia screening of children ages 9-11 years and again at ages 17-21 years. Despite screening guidelines, the AAP Periodic Survey of Fellows demonstrated less than half of pediatricians report adherence to these guidelines. The objective of this quality improvement initiative was to improve physician compliance with hyperlipidemia guidelines in the outpatient pediatric cardiology clinic from 0% to 80% over a 2-month period.

Methods: An IRB-approved, retrospective chart review was performed at a single academic center. Data was collected from 600 individual patient charts from 35 clinic days on which at least 10 patients were seen. Physician compliance was defined as documenting prior screening, ordering a lipid panel, or documenting recommendation for follow-up screening. Two Plan-Do-Study-Act (PDSA) cycles were undertaken and postintervention data was collected for a period of three weeks following each. The primary strategy for implementing change involved educational interventions. The first intervention included informational sessions regarding hyperlipidemia screening guidelines. The second intervention involved weekly email reminders to clinical staff reemphasizing screening guidelines and encouraging compliance.

Results: Baseline compliance with outpatient hyperlipidemia screening was 0%. Following the first PDSA cycle, the average screening rate improved to 49%, which may be due to random chance alone. Following the second PDSA cycle, the average screening rate improved to 89% and a positive shift in the data was seen suggesting true improvement.

Conclusion: Our results show an improvement in physician compliance with the AAP-recommended hyperlipidemia screening guidelines. Ongoing improvement efforts are needed to sustain this change and may include optimization of the electronic medical record to provide notifications for patients due for screening and a "commonly ordered" lab set to include lipid panels.

Improving enrollment and reporting in a national pediatric cardiology quality metrics program

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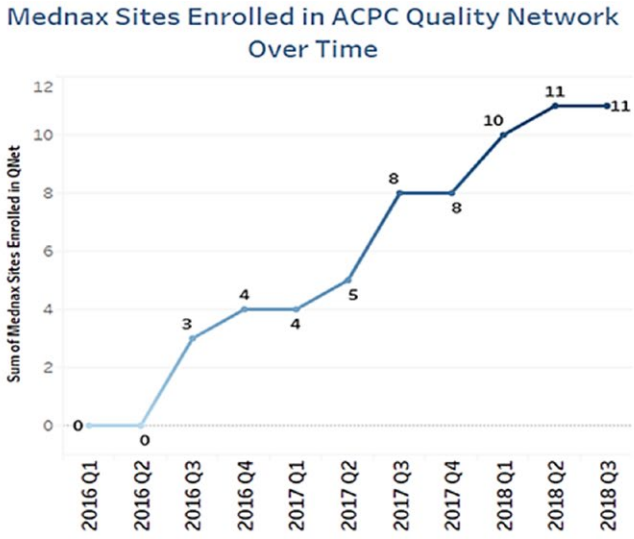
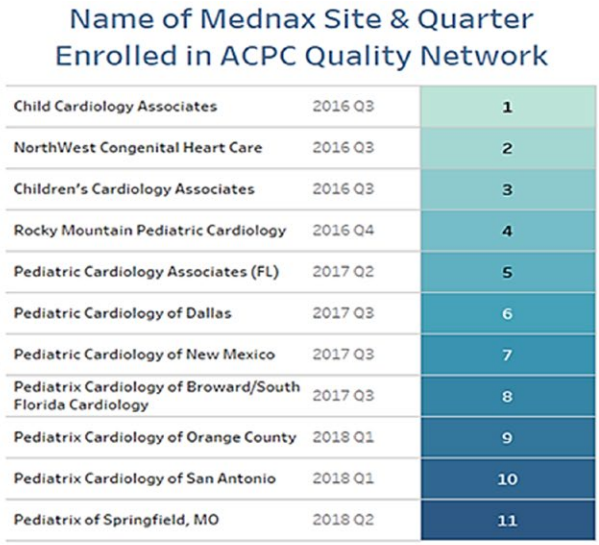
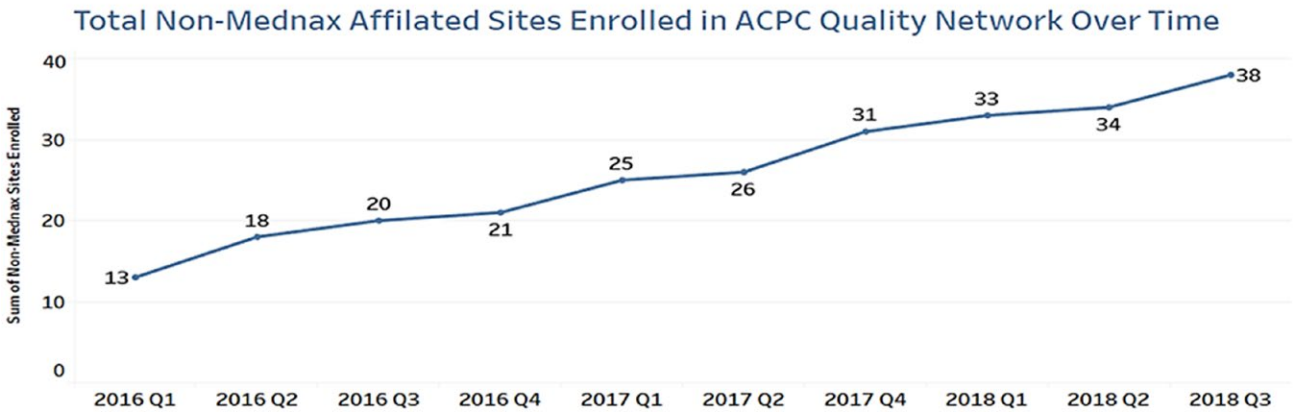
Purpose: The Mednax Cardiology Quality Collaborative (MCQC) joined the ACC Adult Congenital and Pediatric Cardiology (ACPC) Quality Network (QNet) in the fall of 2016. MCQC leadership began projects with established goals by 2018 of: (1) 50% voluntary practice enrollment in ACPC QNet, and (2) 5 metrics reported per practice on the ACPC QNet quarterly reports.

Methods: Mednax Cardiology practices were asked to designate a physician as the practice MCQC lead. Two face to face meetings are held annually to collaborate on quality improvement. Monthly MCQC conference calls follow up on goals and report improvement. ACPC QNet metrics were prioritized for ease of IT extraction and validation across our







practices. An EMR leadership team with IT support has conference calls twice monthly to optimize EMR tools to capture denominator and numerator metric elements and encourage physician compliance with quality metrics by designing work flows conducive to enhanced outcomes.

Results: Three MCQC practices were enrolled in 2016. 10 of 22 practices were enrolled by 2018, nearly achieving the enrollment goal with a 233% enrollment growth (Figure 1). ACPC QNet enrollment growth of non-MCQC affiliated practices was 55% over the same time span (20 to 31 practices). Key factors encouraging enrollment included: (1) a national MCQC/ACPC QNet contract for easier enrollment, (2) EMR enhancements decreasing data collection work and increased physician metric compliance, and (3) the ability to earn MOC part IV credit through ACPC QNet participation.

MCQC reported 2.7 metrics/group in our first 2016 ACPC QNet quarterly report. 6.7 metrics/group were reported for Q4 2017, a 148% growth and surpassing the reporting goal. By comparison, ACPC QNet metric reporting from non-MCQC affiliated practices was 4.1 to 4.7 metrics/group over the same time span (Figure 2). Key factors increasing metric reporting included: (1) ease of data entry and retrieval in the enhanced EMR, (2) earning MOC part IV credit through reporting of specific metrics, and (3) the addition of "denominator reports" for metrics with EMR enhancement. Barriers to metric reporting included the time to collect data. Decreases in metric reporting in individual quarters were associated with enrollment of new practices not providing QNet reports during their first reporting quarter.

Conclusion: Goals of initial MCQC enrollment and metric reporting in ACPC QNet were achieved. Key factors affecting success include: (1) frequent meetings with discussion and follow up, (2) EMR enhancements, and (3) benefits of participation (MOC part IV credit). The next steps will be: (1) continue practice enrollment in ACPC QNet with a goal of all Mednax Cardiology practices participating, and (2) increase metric reporting from individual practices to > 10 of available ACPC QNet metrics. Goals for individual metric performance will be made as longitudinal data on individual metrics becomes available.

Preventing sudden cardiac death in the pediatric population: Improving utilization of the american heart association 14 element guideline to preparticipation evaluation

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Background: In 1996 the American Heart Association released a 14-element guideline for preparticipation evaluation (PPE) to screen young competitive athletes for risk of sudden cardiac

death, which has been endorsed by the American Academy of Pediatrics. This provides a standardized and proven approach to screening patients, ensuring them a thorough evaluation prior to participating in sports. A recent survey of local general pediatricians found that only 34% of practitioners were aware of the guidelines, and of those only 22% were compliant with them. This raises concern that student-athletes are not receiving the necessary evaluation prior to engaging in physically intense activities.

Objective: The aim of this quality improvement initiative is to increase the use of the 14-element guideline for PPE by local pediatricians specifically by raising the average number of elements documented during annual checkups for patients 12-18 years old from 3.5 to 10 by June 2018.

Methods: In November 2017 baseline data was collected weekly from 10 pediatric primary care offices, analyzing medical records of patients 12-18 years of age who received their annual physical exams and reported sports participation. The number of elements from the PPE guidelines that were documented during each visit was calculated. We tracked the weekly average number of elements documented per encounter. We used the IHI Model for Improvement, QI Methodology with one practice to test changes through PDSA cycles before implementing on a larger scale. We educated pediatricians on the guidelines, created a checklist of all 14 elements, and implemented a process by which the checklist was distributed to and completed by patients in the waiting room. We developed a method to review the checklist during the physical exam, and embedded the checklist into the EMR.

Results: Baseline data from the original 10 practices showed an average of 3.5 out of a potential 14 elements being documented; the average for the specific practice we worked with was 2.5. Elevated blood pressure was a positive screen that was consistently overlooked. After implementing the above-mentioned changes, this average increased to 8.

Conclusions: The AHA Guidelines to PPE are underutilized in the practices we analyzed. Education and easy access to the guidelines are crucial in increasing its use. Embedding and completing our checklist has enhanced accessibility and improved compliance with the guidelines. Currently we are creating an interactive template of all 14 elements of the PPE in the EMR to flag positive screens, and an indicator specifically for elevated blood pressure. Future plans include comparing the rates of referrals before and after implementing our changes and determining if we increased the identification of at-risk patients. We will evaluate changes in the incidence of pediatric heart disease, and assess if participation in sports changed as a result of proper evaluation.

Using improvement methodology to increase documentation of echocardiographic ejection fraction measurement in the outpatient pediatric cardiology setting

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Background: The American Society of Echocardiography guidelines recommend documentation of an ejection fraction (EF) in every pediatric echocardiogram (ECHO). This requires that appropriate ECHO images be obtained and a reader performs an EF measurement technique. The Bullet EF measurement is an accepted standard for ECHO based EF measurement. Our specific aim was to use the intervention, of a standardized Bullet EF ECHO measurement educational curriculum, to improve the rate of ECHO EFs documented in an outpatient pediatric cardiology setting, from a baseline of 34% to at least 70% over a 14 month period.

Methods: A tertiary referral center for pediatric cardiology, using quality improvement methodology, implemented a standardized educational curriculum of the bullet EF ECHO measurement, for both sonographers and pediatric cardiologists. The total study period lasted 14 months following the first intervention. The primary outcome, percentage of eligible studies for Bullet EF measurements with documented EF in the ECHO report, was tracked. An ECHO study was considered ineligible if the patient had single ventricle anatomy or the image quality was not sufficient to perform the measurement. A p-chart was used to track compliance of Bullet EF reporting. The data entry and database maintenance was completed by a single sonographer.

A total of five PDSA cycles were completed.

- PDSA-1—Ramp; Education with standardized curriculum completed every 6 months for a total of 2 complete teaching sessions.
- PDSA-2—Sonographer work-flow change consisting of empowering sonographers to make the initial Bullet EF measurement on ECHO studies.
- PDSA-3—Paper Logs were placed in outpatient pediatric cardiology offices to remind pediatric cardiologists to report the number of eligible ECHO studies and number of Bullet EF measurements performed per clinic day.
- PDSA-4—Interobserver variability of Bullet EF measurement was assessed and feedback was given to both sonographers and pediatric cardiologists.
- PDSA-5—Reminder sent via email to sonographers and pediatric cardiologists to perform Bullet EF measurement.

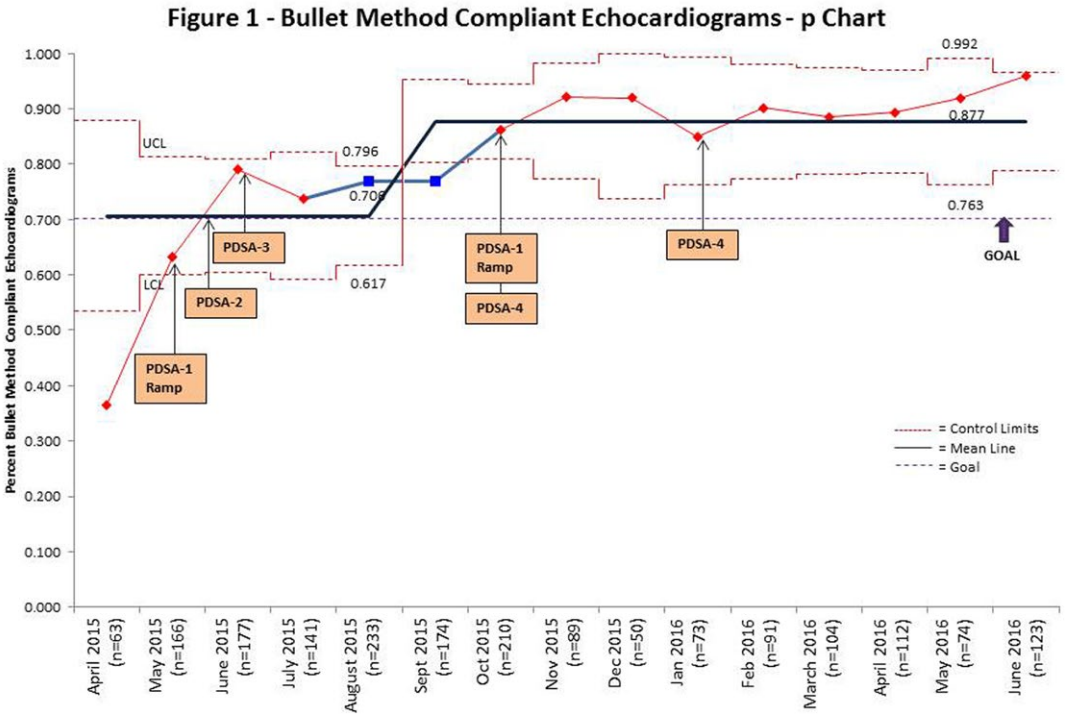

Figure 1 – The percentage of eligible echocardiograms with documentation of ejection fraction (EF) versus time (monthly). Individual months are represented by dots. The solid line denotes the running mean and the dashed red line denotes the control limit, with upper control limit (UCL) and lower control limit (LCL). Key interventions (PDSA cycles 1-5) with corresponding times of implementation are indicated as boxes on the chart.
PDSA-1 Ramp = standardized education, PDSA-2 = work-flow change, PDSA-3 = paper logs, PDSA-4 = interobserver variability assessment and feedback, PDSA-5 = Reminder email.

Results: Using the p-chart (Figure 1) we established the first 4 months as the baseline mean. Following the PDSA cycles, there was a significant shift with 8 points above the mean line to 88%. This change was associated with appropriate change in control limits. The largest shifts were seen following PDSA-1 and PDSA-2. Continued improvement was also noted following the other three PDSA cycles. No intervention was deemed to be detrimental.

Conclusion: Improved documentation of ECHO-based measurement of EF was achieved using quality improvement methodology. Based on our project, we suggest that a change-bundle to improve ECHO measurements; reporting should include in-person education on ECHO based EF methodology, work-flow change of front-line sonographers to attempt Bullet EF measurement on studies prior to attending review, and continuous feedback for sonographers and pediatric cardiologists.

YOUNG INVESTIGATOR ABSTRACT (YIA) COMPETITION
PRESENTATION TIME: FRIDAY, NOVEMBER 2, 2018: 1:30 PM-5:00PM

Does QRS duration predict exercise capacity in patients with tetralogy of fallot?

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Purpose: Patients with tetralogy of Fallot (TOF) have an increased long-term risk of cardiovascular morbidity and mortality. Elevated QRS duration (QRSd), specifically, has been shown to be a specific risk factor for the development of right ventricular myocardial disease, ventricular arrhythmias, and subsequent sudden death in TOF patients. Cardiopulmonary exercise testing (CPET) has also been shown to be a useful predictor for cardiac-related hospitalizations in TOF patients. Our objective was to determine if there was any association between QRS duration and performance on CPET in patients with TOF.

Methods: We performed a retrospective cohort study of patients with a history of surgical intervention for TOF who underwent their first CPET at our institution. QRSd were hand measured by a pediatric electrophysiologist on resting ECGs obtained prior to CPET. CPET outcomes were standardized to account for potential confounders such as age, race/ethnicity, and sex. We calculated Spearman correlation coefficients to assess the correlation of QRS duration with % of predicted peak oxygen uptake (VO_2max), predicted peak oxygen pulse, predicted anaerobic threshold and predicted maximal heart rate on CPET.

Results: There were 61 patients included in the study, with a median age at time of CPET of 12.7 years (25^{th} = 11.4 y, 75^{th} = 15.6 y) and an overall median QRS duration of 139 ms (25^{th} = 113, 75^{th} = 151). On average, patients with TOF performed below predicted performance for all CPET measures. However, there was no correlation between QRS duration and % of predicted VO_2max, predicted peak oxygen pulse, predicted anaerobic threshold, or predicted maximal heart rate (Figure 1).

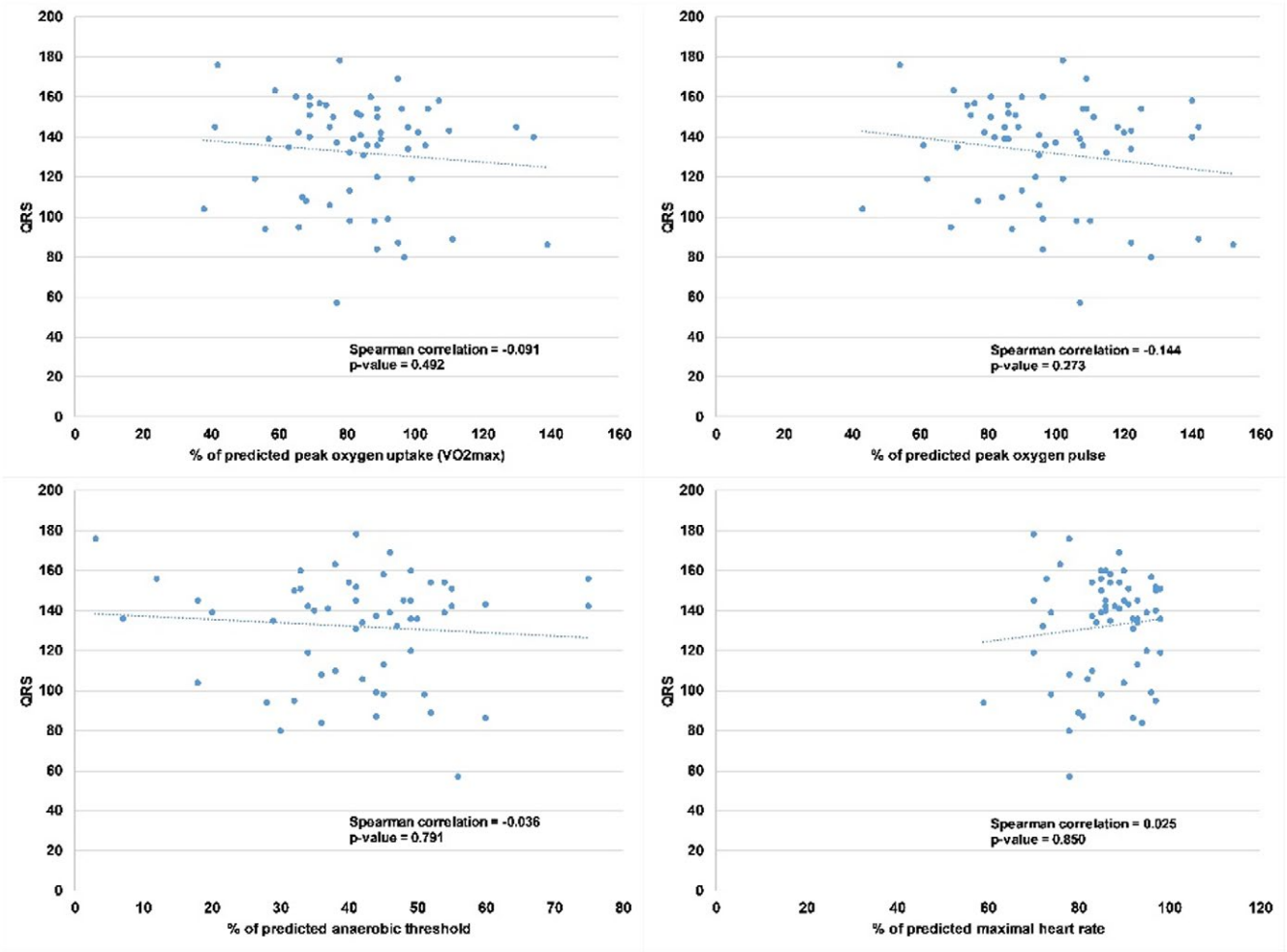

FIGURE 1 Spearman correlation demonstrating the relationship between QRS duration on resting electrocardiogram and performance measures on cardiopulmonary exercise testing in patients with tetralogy of Fallot. No significant relationship was found for any of the measures.

Conclusion: We did not find a relationship between QRS duration on resting ECG and exercise capacity in patients with TOF. Given the inability of QRS to serve as a predictive marker for exercise performance, there remains an important role for CPET to properly evaluate these patients, exercise capacity for the purposes of risk stratification and the need for further surgical palliation.

Incidence and timing of thromboembolic events after the norwood procedure in the single ventricle reconstruction clinical trial of the pediatric heart network (PHN)

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Purpose: Thromboembolic events are a common cause of increased morbidity and mortality in infants who undergo staged surgeries for single ventricle congenital heart disease (CHD). However, previously reported rates vary, from 1 to 46%, due to limitations from small sample size, retrospective study design, heterogeneity of patient population, and single center experience. We used the public dataset of the PHN Single Ventricle Reconstruction (SVR) randomized clinical trial to determine the incidence, timing of, and factors associated with thromboembolic events following the Norwood procedure.

Methods: The SVR trial spans from 2005 to 2009 and includes infants diagnosed with hypoplastic left heart physiology who underwent randomization to the Norwood procedure with modified Blalock-Taussig shunt (mBTS) or right ventricle to pulmonary artery shunt (RVPAS). Univariate analysis was performed using demographic, clinical, and surgical factors available from this study to determine association with thrombosis. Comparisons between groups were made using Wilcoxon rank-sum tests for continuous variables and chi-square tests for categorical variables. Time to thrombosis was evaluated using survival analysis.

Characteristics	Thrombosis (n=57, %)	No Thrombosis (n=492, %)	p-value
Demographics			
Gestational age (weeks)	38 (38 – 39)	38 (37 – 39)	0.8367
Birth weight* (grams)	3085 (2860 – 3420)	3150 (2750 – 3475)	0.9571
Sex			**0.0052**
Male	45 (78.9%)	294 (60.0%)	
Female	12 (21.1%)	197 (40.0%)	
Race			0.1225
White	50 (89.3%)	386 (79.1%)	
Black	6 (10.7%)	80 (16.4%)	
Other	0 (0.0%)	22 (4.5%)	
Anatomic Diagnosis			0.1041
Hypoplastic left heart syndrome	44 (77.2%)	430 (87.4%)	
Critical aortic stenosis	1 (1.8%)	4 (0.8%)	
Single right ventricle w/outflow obstruction	6 (10.5%)	17 (3.5%)	
Right dominant AV canal	4 (7.0%)	27 (5.5%)	
Straddling MV w/hypo LV	0 (0.0%)	1 (0.2%)	
Functional single right ventricle	2 (3.5%)	13 (2.6%)	
Surgical Factors			
Weight at Norwood* (kg)	3.10 (2.80 – 3.40)	3.20 (2.78 – 3.50)	0.8171
Total bypass time* (minutes)	145 (119 – 184)	137 (104 – 169)	**0.0451**
Total DHCA time*† (minutes)	36 (15 – 53)	34 (13 – 46)	0.1689
Norwood shunt type			
mBTS	29 (50.9%)	239 (48.6%)	
RV to PA Shunt	28 (49.1%)	253 (51.4%)	
Aprotinin administered	38 (66.7%)	389 (79.1%)	**0.0331**
Other			
Cardiac catheterization	2 (3.5%)	37 (7.6%)	0.4129
ECMO during Norwood hospitalization	1 (12.3%)	81 (16.6%)	0.3986
CPR during Norwood hospitalization	14 (24.6%)	83 (17.0%)	0.1606

*Values reported as Median (IQR 25th - 75th) or N (%). +DHCA: deep hypothermic circulatory arrest

TABLE 1 Patient characteristics and univariate factors associated with thrombosis

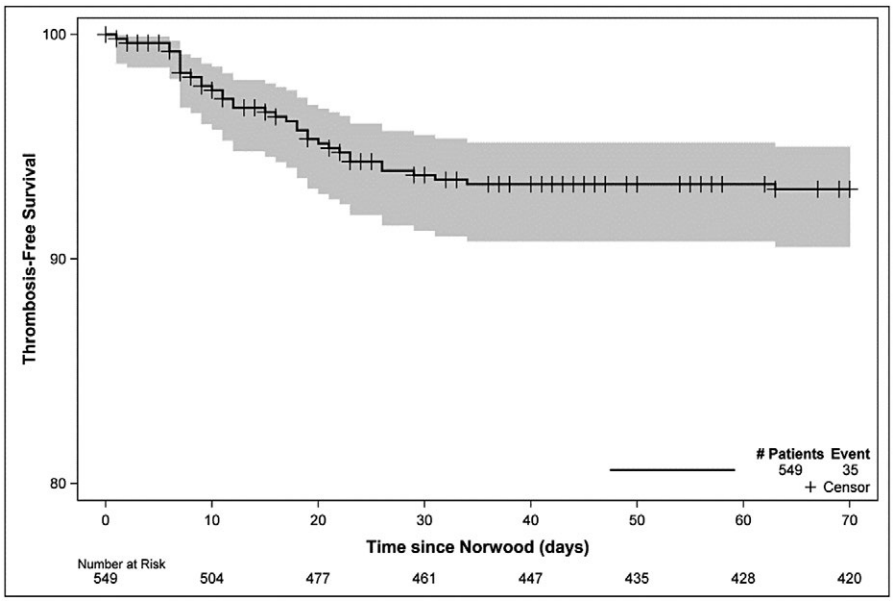

FIGURE 1 Thrombosis-free survival following the Norwood procedure

Result: Of the 549 infants included in the SVR trial, the majority were term (88%) white (64%) males (62%) with a median birth weight of 3.10 kg and predominant anatomic diagnosis of HLHS (86%) (Table 1). Thirty-five (6%) patients had thrombosis during Norwood hospitalization, with the majority (33/35) developing thrombosis in the first 30 days following the Norwood (Figure 1). Median time to first thromboembolic event was 23 days. There was no difference in the rate of thrombosis based on shunt types (mBTS vs. RVPAS). Factors associated with thrombosis were male sex (P=.0052), longer duration of cardiopulmonary bypass (P=.045), and aprotinin use (P=.033). Gestational age, race, anatomic diagnosis, ECMO use, and need for CPR were not associated with thrombosis. Development of thrombosis during Norwood hospitalization was associated with prolonged median ICU stay and length of hospital stay; 27 vs. 13 days (IQR 13-46 days vs. 9-25 days, P<.001) and 36 vs. 23 days (IQR 26-58 days vs. 15-38 days, P<.001), respectively.

Conclusion: In this largest reported prospective cohort of infants undergoing single ventricle reconstruction, the cumulative incidence of thrombosis during the Norwood hospitalization was 6.4%. Aprotinin use and male sex were associated with risk of thrombosis and this risk was highest within 30 days post Norwood. Thrombosis is associated with longer length of ICU and hospital stays. Further studies are needed to investigate the utility of modification of current management practices, including targeted thromboprophylaxis during this high risk period following the Norwood procedure.

Assessment of left ventricle function with 2D strain speckle tracking echocardiography in patients following balloon aortic valvuloplasty for aortic valve stenosis

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Background: Balloon aortic valvuloplasty (BAV) is the first line of treatment for aortic valve stenosis. The long-term effects of the procedure on left ventricular function remain uncertain. Our hypothesis was that left ventricular dysfunction persists following BAV when assessed by the more sensitive 2D speckle tracking echocardiography, compared to normal controls.

Methods: This was a retrospective cross-sectional study of patients who had undergone BAV for aortic valve stenosis >1 year before enrollment. Exclusion criteria included more than moderate aortic insufficiency, more than mild aortic stenosis, and inadequate image quality for strain analysis. Patients with chest pain, murmur or syncope and normal echocardiograms acted as controls. The most recent echocardiogram available was analyzed offline by a trained observer using vendor independent software (TomTec Imaging). Global longitudinal strain for endocardium and myocardium (GLSendo and GLSmyo) were calculated from the apical 4-chamber view. Circumferential strain (CSendo and CSmyo) was calculated from the parasternal short axis view at the level of the papillary muscle. A ratio of GLS to CS was also calculated. Statistical analysis included Student *t* test for comparison between groups. *P* value <.05 was considered significant.

Results: Our cohort included 63 subjects, 29 (46%) of whom had undergone BAV. BAV was performed at a mean (SD) age of 4.2 (5.6) years (median 1 week) and the follow-up echocardiogram was obtained at a mean (SD) duration of 7.4 (4.1) years after the procedure. The BAV and control groups were comparable in age, height and weight but the BAV group comprised more males. All patients had normal ejection fraction and normal qualitative LV function by echocardiography. GLSendo (*P*<.001) and GLS to CS ratio (*P*=.04) were significantly decreased in the BAV group compared to controls.

Demographic and Strain Parameters

Parameters Mean(SD) or n(%)	Aortic Valvuloplasty (n=29)	Normal Group (n=34)	p value
Age	11.8 (5.6)	12.5 (4.04)	0.56
Gender (Male)	20 (69%)	16 (47%)	**0.05**
Height (cm)	145.1 (27.5)	152.3 (22.5)	0.26
Weight (kg)	44.6 (24.1)	49.3 (18.9)	0.39
GLSmyo	-17.3 (3.7)	-18.6 (2.0)	**0.10**
GLSendo	-20.7 (3.6)	-24.2 (2.7)	**<0.01**
CSmyo	-20.7 (4.04)	-20.0 (4.5)	0.57
CSendo	-27.7 (4.6)	-29.3 (5.0)	0.21
GLSendo/CSendo	0.76 (0.17)	0.84 (0.14)	**0.03**
GLSmyo/CSmyo	0.85 (0.20)	0.97 (0.21)	**0.03**

Demographic data shows predominance of males consistent with epidemiology of aortic valve stenosis. Strain parameter analysis shows significantly decreased global longitudinal strain in endocardium in post valvuloplasty patients compared to controls.

Conclusions: About 4 years after BAV for aortic stenosis, left ventricle global longitudinal strain was decreased, compared to controls, even in a cohort with normal conventional echocardiographic parameters. This intriguing finding needs to explored further to determine its progression, if any.

The effects of cardiopulmonary bypass on infant vaccination titers

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Purpose: Infants with congenital heart disease that require surgery in the first year of life remain vulnerable to potentially preventable pathogens. While childhood immunization can significantly reduce this risk, it is unknown how cardiac surgery with cardiopulmonary bypass (CPB) affects an infant's vaccination status, especially in the setting of an immature immune system. Consequently, significant variation in vaccination policies exist among major surgical centers surrounding children expected to undergo cardiac surgery with CPB. The objective of this study was to evaluate the effect of CPB on infant vaccination status following cardiac surgery.

Methods: We conducted a prospective observational study of patients between 2 and 14 months of age at our institution who required cardiac surgery with CPB between 2016 and 2018. Patients were eligible if they had received at least their first round of infant vaccinations. Antibody titers to diphtheria, tetanus, poliovirus, bordatella, hepatitis B, and haemophilus influenzae were measured immediately prior to the infant being placed on CPB and again the following morning after surgery. Demographic and surgical variables, including the use of blood products, were assessed via regression methods for their effects on the observed change in antibody titers.

Results: Among 80 patients consented, 77 had complete antibody titer analysis. There was no demonstrated difference between the mean pre- and postoperative values in regards to diphtheria (0.37 vs. 0.34, P=.23), tetanus (0.69 vs. 0.69, P=.825), polio 1 (P=.068), polio 3 (P=.39) or haemophilus influenzae titers (log -0.37 vs. -0.34, P=.41). Bordetella titers (1.03 vs. 0.84, P<.001) did demonstrate a significant reduction after CPB but none of the titers among any vaccine fell below the threshold level to be considered immunized. Resultant changes in observed antibody titers were not associated with time between immunization and surgery, age or weight at surgery, blood products administered, number of previous doses of the vaccine, time on CPB or heterotaxy diagnosis for most of the vaccines. Among patients that had only yet received their first round of immunizations (38%), there was no difference in the mean pre- or postoperative values, and no titers fell below the immunized threshold. Change in individual titer levels for this group was strongly associated with time on CPB, however, for all vaccines except hepatitis B

Conclusion: Infant vaccine antibody titers were minimally affected by CPB and not associated with any modifiable surgical variables. This was true whether the initial or multiple doses of the immunizations had been administered. While antibody titers are only one marker of immunity, this suggests that deviation away from the recommended vaccination schedule is unnecessary for children expected to undergo congenital heart surgery with CPB.

Are care quality indicators associated with provider network characteristics? A study of young adults with congenital heart disease

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Purpose: Outpatient ACHD QIs have been developed but little evidence exists on performance using US population data. Insight into how provider networks are associated with care quality is important to understanding healthcare delivery systems. This study evaluated receipt of high quality care by young adults with congenital heart disease (CHD) using a recently developed set of outpatient adult congenital heart disease (ACHD) quality indicators (QIs). The study also assessed the association between QIs and provider network measures.

Methods: Young adults (ages 16-31) with complex/severe CHD were identified using ICD9 codes in the 2010-2014 Colorado All-payers Claims database. Networks of providers and their shared patients were identified from outpatient CHD utilization data. QIs (annual pulse oximetry, annual EKG, annual echocardiogram, annual adult cardiology visit, and successful transition to adult cardiology) were assessed in their relevant CHD population using claim data. Probit analysis assessed outcomes as a function of network measures and patient, provider, and temporal covariates.

Results: The QI sample included 440 insured young adults with eligible CHD lesions representing 1337 patient-years. The transition sample included 357 insured young adults with complex/severe CHD. 53% of the QI sample received an echocardiogram. 35% of the transition sample successfully transitioned to any adult cardiology care within three years of their index visit; 20% transitioned to ACHD care specifically. Another 35% of the transition sample saw PCPs but no cardiology providers during that three year period. Males were 8.7% less likely to have successfully transitioned to ACHD care (P<.001). Males (P=.003) and rural patients (P=.013) were more likely to have received an echocardiogram. Young adults with a general adult/pediatric cardiologist were more likely to meet the echocardiogram QI compared to young adults receiving care from a primary care provider (PCP) (P<.001) or other specialist (P<.001). Network measures were associated with outcomes. Receiving care from more closely connected providers in the network, as opposed to less closely connected providers, increased the probability of young adults meeting the echocardiogram QI (P=.042). Young adults seen by closely connected PCPs were 31% more likely to transition to ACHD care

compared to those seen by less closely connected PCPs (*P*=.003). Among pediatric cardiologists, that network measure increased ACHD transition probability by 3.5% (*P*=.003).

Conclusion: We found suboptimal transitions and poor compliance with recommended standards of care in the young adult CHD population. Network analysis indicates provider relationships factor into a patient's receipt of quality ACHD care. As more than a third of the transition sample were treated by PCPs without any outpatient cardiologist care, there are opportunities to strengthen PCP-cardiologist relationships and improve ACHD care. This population may also benefit from receipt of care from ACHD providers or wider dissemination of ACHD guidelines to other care providers.

Association of congenital heart disease with autism: A case-control study

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Background: There has long been an association between congenital heart disease (CHD) and general neurodevelopmental delays. However, the association between CHD and Autism spectrum disorders (AuSD) is less well understood. Prior studies to assess an association have been limited by small sample size and questionnaire recall.

Objective: Using administrative data from the Military Health System, we sought to quantify the association between CHD and AuSD, as well as to identify specific CHD lesions with higher odds of developing AuSD.

Design/method: We performed a 1:3 case-control study with children born and enrolled in the US Military Health system between October 2000 and September 2013. Cases with AuSD were matched with unaffected controls on the basis of date of birth, sex, and enrolment timeframe. *International Classification of Diseases, Ninth Revision, Clinical Modification* (ICD-9-CM) diagnostic codes for each child were obtained and reviewed for CHD codes and associated procedures. CHDs were further subdivided according to developmental categories. Conditional logistic regression determined ORs and 95% CIs for comparative associations.

Results: A total of 8760 children with AuSD and 26 280 controls were included in the study. The groups were similar with respect to the matching characteristics, with males comprising 79.9% of each group. Those with AuSD were more likely to have a genetic syndrome, prematurity, maternal gestational diabetes, and younger maternal age at delivery (Table 1). After adjustment for these characteristics, there remained increased odds of AuSD in patients with CHD. For any CHD, the OR was 1.45 (95% CI 1.32-1.60). This finding was largely driven by the atrial septal defect group (OR 1.50, 95% CI 1.31-1.72), but it was also significant for those with left heart obstructive lesions (OR 1.66, 95% CI 1.18-2.35), ventricular septal defects (OR 1.27, 95% CI 1.02-1.58), and anomalous pulmonary venous return (OR 3.09, 95% CI 1.02-9.36). (Figure 1)

TABLE 1

Characteristics	Autism SD (n=8,760) n, (%)	Control (n=26,280) n, (%)	p-value
Congenital Heart Disease	885 (10.1%)	1,463 (5.6%)	**<0.001**
Genetic Syndrome	411 (4.7%)	178 (0.7%)	**<0.001**
Prematurity	1,171 (13.4%)	2,302 (8.8%)	**<0.001**
Gestational diabetes	1,062 (12.1%)	2,916 (11.1%)	**0.009**
Maternal age, years (IQR)	28.0 (24.3 – 32.3)	29.1 (25.5 – 33.2)	**<0.001**

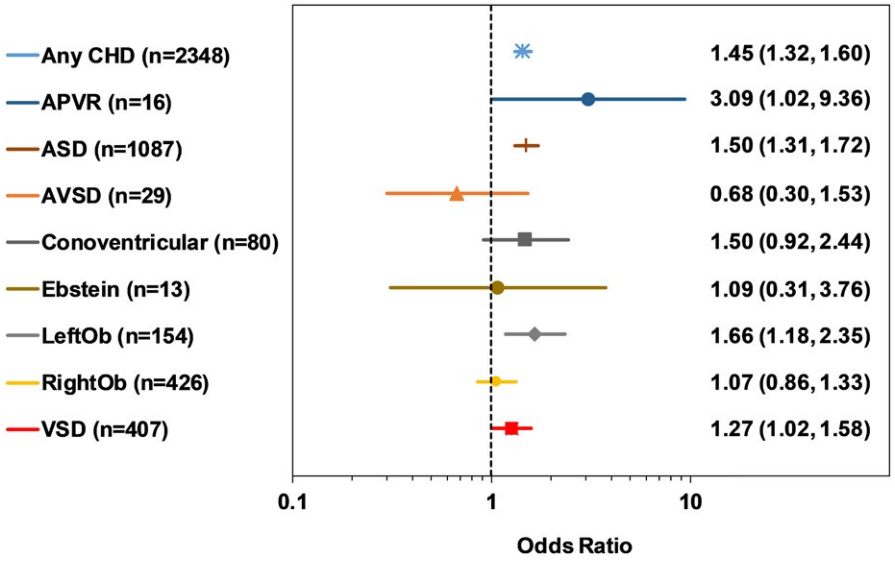

FIGURE 1 Odds ratios for AuSD with associated CHD categorical diagnosis adjusted for genetic syndrome, prematurity, maternal gestational diabetes, and maternal age at delivery.
Abbreviations: CHD, congenital heart disease; APRV, anomalous pulmonary venous return; ASD, atrial septal defect; AVSD, atrioventricular septal defect/atrioventricular canal defect; Conoventricular, defects including tetralogy of Fallot, truncus arteriosus, AP Window; Ebstein, Ebstein malformation; LeftOb, left heart obstruction including hypoplastic left heart syndrome, mitral stenosis, aortic stenosis, and coarctation of the aorta; RightOb, right heart obstruction including tricuspid atresia, pulmonary atresia, pulmonary stenosis, not including tetralogy of Fallot; VSD, ventricular septal defect

Conclusions: Children with CHD are at increased odds of developing AuSD. Specific lesions at risk include atrial septal defect, left heart obstructive lesions, ventricular septal defects, and anomalous pulmonary venous return. Our findings will be useful for counseling parents and caretakers of children with CHD.

Subclinical chronic kidney injury is common in adolescent and young adult patients with Fontan circulation: Results of a urinary biomarker study

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Background: Surgicavl palliation of single ventricle congenital heart disease (CHD) is associated with peri-operative risk of acute kidney injury. While overt chronic kidney disease (CKD) in adolescent and young adult patients with Fontan circulation is rare, the presence of subclinical chronic kidney injury in this population has not been investigated.

Methods: Single-center prospective cohort study of patients with Fontan circulation (Fontan) and biventricular CHD (control) scheduled for elective cardiac catheterization. Urine and blood samples were collected prior to catheterization. Urinary biomarkers assessed included neutrophil gelatinase-associated lipocalin (NGAL) and liver-fatty acid binding protein (L-FABP), markers of proximal renal tubular injury, and kidney injury molecule-1 (KIM-1) and interleukin 18 (IL-18), markers of distal tubular injury. Estimated glomerular filtration rate (eGFR) was calculated using the bedside Schwartz equation (≤18 yrs) or the MDRD formula (>18 yrs). Continuous, normally distributed data were compared using *t* tests, nonnormally distributed data with Mann-Whitney U tests and categorical with chi-square.

Results: Thirty-four Fontan subjects (median age 16 yrs, 50% female) and 20 controls (median age 12.5 yrs, 50% female) were enrolled. There were no differences in height, weight, BMI or race/

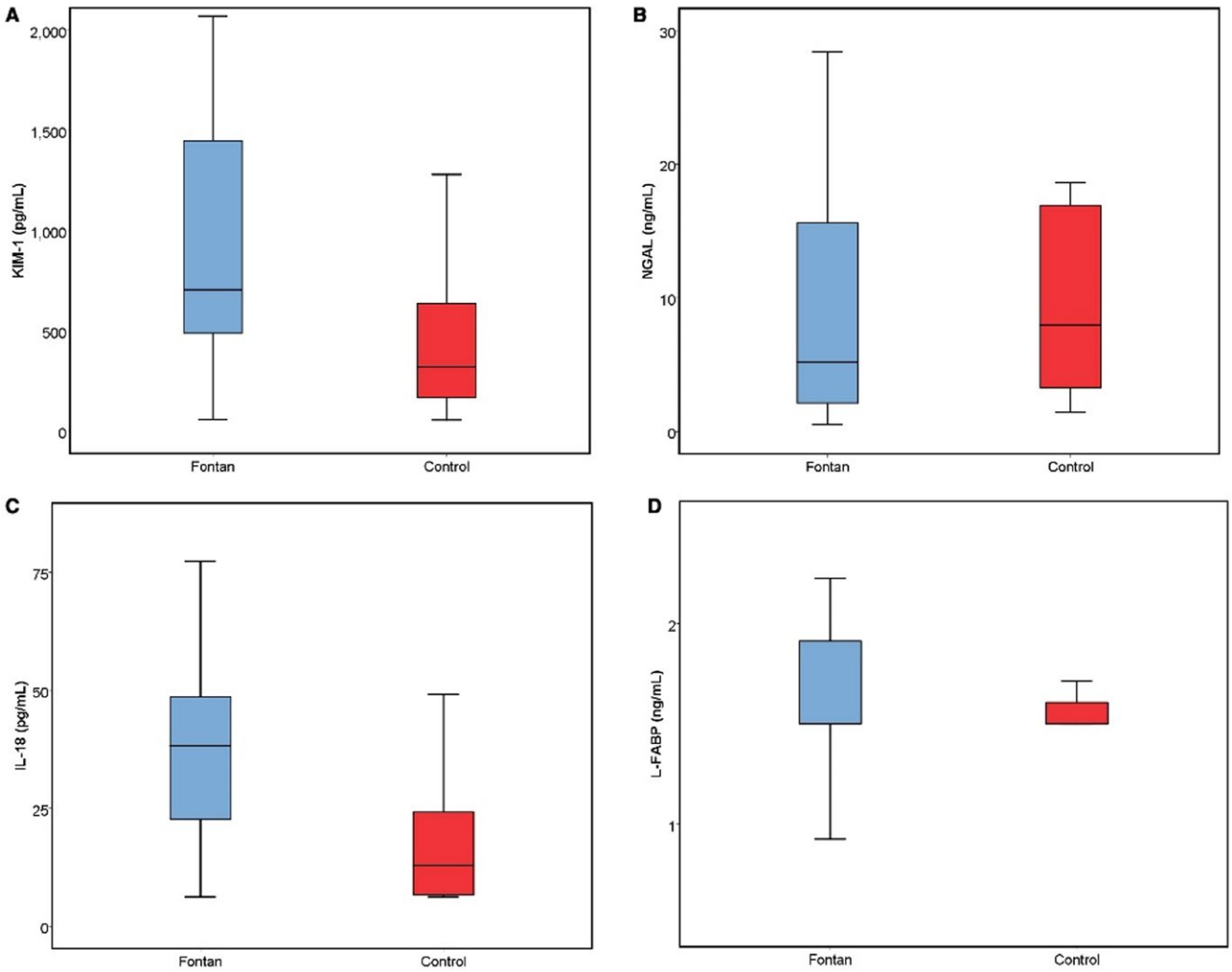

FIGURE 1 Paired box and whisker plots of urinary biomarkers KIM-1 (A), NGAL (B), IL-18 (C) and L-FABP (D) in Fontan versus Control CHD subjects. The box reflects the intraquartile range, the horizontal line reflects the median and the whiskers reflect the lowest and highest values within 1.5 times the intraquartile range of the lowest and highest, respectively.

ethnicity. Chronic exposure to nephrotoxic agents (e.g. nonsteroidal anti-inflammatory drugs or diuretics) were not different between groups. Systolic BP (104±14.4 vs. 111.5±9.4 mm Hg, *P*=.04) and oxygen saturation (93 (IQR 91, 96) vs. 99 (97.3, 100)%, *P*=.02] and IL-18 [38.2 (21.7, 50.9) vs. 12.9 (6.5, 24.6) pg/mL, *P*=.001] were significantly higher in Fontan subjects, whereas NGAL [5.2 (2.1, 15.9) vs. 8.0 (3.2, 17.5) ng/mL, *P*=.5] and L-FABP [1.5 (1.5, 2) vs. 1.5 (1.5, 1.7) ng/mL, *P*=.8] were not significantly different between groups. (Figure 1).

Conclusion: While traditional markers of renal function did not differ between groups, urinary biomarkers suggestive of kidney injury were significantly elevated in Fontan subjects. This finding is consistent with the presence of subclinical chronic kidney injury and may identify those patients at higher risk of progressing to clinical CKD.

Normal ranges of Troponin T and NTproBNP levels in premature infants within first 5 days of life

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Background: Echocardiography is used for the diagnosis and management of hemodynamically significant patent ductus arteriosus (hsPDA) and pulmonary hypertension (PH) in preterm infants (PI). Heart muscle cells release biomarkers such as highly sensitive Troponin T (hsTnT) and NT-proBNP in response to pressure and volume loading. However, little is known about the baseline levels of these biomarkers in PI.

Objective: To determine the normal range of NTproBNP and hsTnT levels in PI in the first 5 days of life.

Methods: After IRB approval and parental consent, PI (34 weeks and 1500gm) were prospectively enrolled in the observational study. Infants with major congenital or chromosomal anomalies were excluded. Between days 3 to 5, an ECHO was performed and blood sample was drawn within 30 minutes of ECHO. Serum was collected by centrifuging blood and saved at −80^{0}C. Infants were divided into 3 groups based on ECHO results PDA diameter: No PDA, PDA < 1.5 mm and hsPDA > 1.5 mm diameter. Clinical, demographic, ECHO and biochemical markers data was collected and analyzed using chi-square and ANOVA tests.

Results: We recruited 76 PI (No PDa n=44, hsPDA n=20) with mean gestation 26 to 28 weeks and birth weight 884 -1056 grams. NTproBNP and hsTnT were significantly lower in infants with no PDA as shown in Figure 1A and 1B. Median hsTnT and NTproBNP levels in PI (with no PDA) within 5 days of life are 158 ± 74 pg/mL and 2051 ± 1242 ng/L respectively. hsPDA increases both hsTnT and NTproBNP in PI.

Conclusion: In preterm infants, hsTnT and NT-proBNP levels are significantly higher in patients who have a PDA that is significant as represented in Table 1. Normal median levels were established for these biomarkers in PIs with no PDA.

Pulse oximetry screening for detection of critical congenital heart defects at an altitude of 5400 feet at a tertiary care center in New Mexico

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Purpose: Universal newborn pulse oximetry screening to detect critical congenital heart disease (CHD) has become the standard of care in the United States. Using the newborn pulse oximetry screening

Table 1. Comparing significant PDA to no PDA and non-significant PDA

Markers	No PDA (a)	PDA <1.5 (b)	PDA >1.5 (c)	P value a vs b	P value a vs c	P value b vs c
TnT (Mean)	161 ± 80	162 ± 99	262 ± 137	0.953	0.005	0.024
NtProBNP (Mean)	3118 ± 3045	3344 ± 2654	16,869 ± 15759	0.803	0.001	0.001

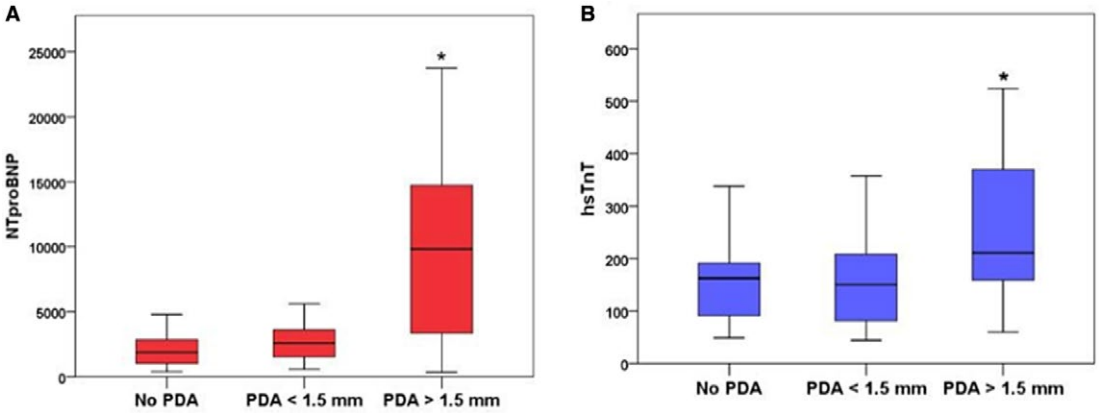


FIGURE 1 NTproBNP (A) and hsTnT (B) levels in preterm infants with no PDA, PDA < 1.5 and PDA > 1.5 mm. * represents significant difference.

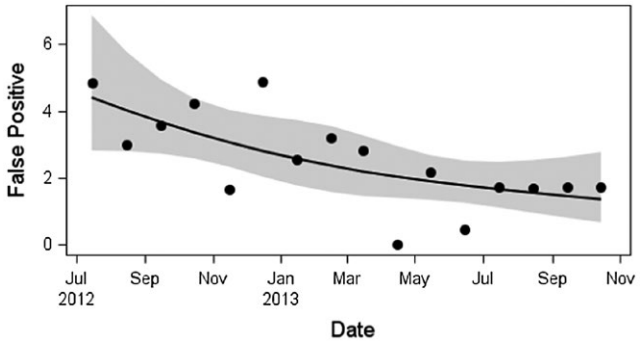
protocol recommended by the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children, American Academy of Pediatrics and American Heart Association, we sought to determine the false positive rate at moderate altitude, which we hypothesized would be considerably higher than that recorded at sea level.

Methods: Data was collected retrospectively from July 2012 to October 2013. Infants with a gestational age of at least 36 weeks and weighing at least 2 Kg and infants with a gestational age of 37 weeks or greater and weighing at least 1.7 Kg admitted to the New Born Nursery were included. Infants with a prenatal diagnosis of CHD or those admitted to the Intermediate Care Nursery or Neonatal Intensive Care Unit were excluded. Preductal and postductal oxygen saturations were recorded after 24 hours of life or at the time of discharge. A positive screen resulted in repetition of the test based on clinical exam, four extremity BP measurements, and/or a Pediatric Cardiology consult with an echocardiogram. We tested if the average false positive rate (FPR) from July 2012 – March 2013 differed from April – October 2013, including an overall trend, after educating caregivers about the protocol. As part of quality improvement, pulse oximetry screening was added as a mandatory competency skill. Logistic regression was used to assess the change in probability of false positives over time.

Results: Three thousand six hundred twenty-seven newborns were screened. After excluding 79 patients who had insufficient data, there was 1 true positive, 93 false positives, 91 incorrectly interpreted, and 273 that required multiple attempts to pass. The median preductal and postductal saturations for the whole group were 96.0% and 95.8%, respectively. The median preductal saturation for false positives was 92% and mean 91.5%. The median postductal saturation for false positives was 90.0% and mean 89.8%. The FPR before April 2013 was 3.50% (95% CI 2.72-4.50, *P*=.003) compared to the FPR of 1.48% (95%CI 0.95-2.31, *P*=.003) after March 2013. The average FPR was 2.62% (93 infants), much higher than the national average of 0.035%. Only one true positive was detected. There was a significant log-linear decline in FPR with time (*P*=.003).

Conclusion: The initial FPR between July 2012 and March 2013 was 3.50%. This decreased to 1.48% between April and October 2013. This was still much higher than the national average of 0.035%, thought to be a combination of learning curve and the effect of altitude on pulmonary vascular resistance. Higher FPR at moderate altitude places an additional cost on health systems, especially in resource limited areas. Future studies are required to determine an optimal screening protocol at moderate altitude.

FIGURE 1 Trend in the false-positive rate (FPR) with time



Decline in the FPR with time following which there is a plateau, still higher than the national average. Initial steep slope presumed to be part of the learning curve. Greyscale indicates the 95% Confidence Interval.

Clinical features and outcomes of patients necessitating veno-venous to veno-arterial conversion during extracorporeal membrane oxygenation support

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Background: In newborns with congenital diaphragmatic hernia, need for conversion from veno-venous (VV) extracorporeal membrane oxygenation support (ECMO) to veno-arterial (VA) ECMO support has been described in 15-20% of patients with 50-60% mortality. There is extremely limited data regarding circumstances, clinical features, and outcomes of all other patients requiring conversion from VV ECMO. The purpose of this study is to review when conversion occurs, why it occurs, and patient outcomes after conversion at a large academic ECMO center.

Methods: Patients undergoing VV to VA conversion were identified from our prospectively collected ECMO data registry. Additional clinical data was retrospectively collected after IRB approval. Appropriate analysis was performed.

Results: VV to VA conversion was needed in 42 out of a total of 1150 patients (3.6%) supported on ECMO between 1994-2017. Median age at ECMO initiation was 91 days (range 0 days to 18 years). Median weight was 4.8 kg (2.4-90kg). The primary diagnosis included congenital diaphragmatic hernia in 9, respiratory failure in 8, meconium aspiration syndrome in 4, pneumonia in 4, sepsis in 4, pertussis in 3, bronchiolitis in 2, and miscellaneous causes in 8 patients. Prior to ECMO, 35/42 (83%) were on 100% oxygen, 42/42 on mechanical ventilation, and 16/42 (38%) had pH lower than 7.2. Initiation on ECMO was done using neck cannulation for venous drainage in 38/42 (90%) patients, 3 via femoral, and 1 with central right atrial cannulation. Additional cephalad venous cannula was used in 28/42 (66%) patients. With this configuration, conversion to VA was needed at median of 2 days (range 0-15 days) after initiation. Reasons for conversion were cardiovascular failure in 21/42 (50%), inadequate flows in 13/42 (31%), and combined reasons 8/42 (19%). 3/42 (7%) patients had a cardiac arrest while on VV ECMO prior to

conversion. Complications after conversion included circuit clots/circuit issues in 28/42 (67%), need for renal support in 28/42 (67%), bleeding complications in 13/42 (31%), neurologic complications in 7/12 (16%) patients. Total ECMO support was for median 12.5 days (range 1-46 days). Overall survival was 18/42 (43%).

Conclusion: For the first time, in a large center for Pediatric ECMO, this study demonstrated the need for conversion from VV to VA ECMO amongst all neonatal and pediatric indications is rare (3.6%). VV to VA conversion is associated with higher morbidity and mortality compared to VV ECMO only. Further risk analysis is being performed to understand the risk factors for conversion as well as mortality after conversion.

Outcomes of transcatheter device closure of hemodynamically significant patent ductus arteriosus in premature neonates ≤ 3 kilograms

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Background: Current management strategies for hemodynamically significant patent ductus arteriosus (PDA) in premature neonates include medical closure with indomethacin and ibuprofen, surgical ligation, and transcatheter device closure. Challenges to each include significant complications for medical and surgical groups, and high failure rates for medical closure. Transcatheter methods have been impeded by device limitations and techniques that increased complication risk. Increasing efforts at transcatheter closure are occurring, however, most are case reports and small, single center studies.

Purpose: This study sought to demonstrate the safety and efficacy of transcatheter PDA device closure in low weight neonates weighing less than or euqal to 3 kilograms and to analyze its effect on respiratory status.

Methods: From January 2012 to April 2018, all premature neonates less than or equal to 3 kg were retrospectively reviewed. Procedural details, respiratory support status, clinical outcome, and echocardiographic findings were recorded from preprocedural, post-procedural and discharge or death time points.

Results: Sixty-four PDA device closures were performed on 62 patients with a median weight of 2.1 kg (0.9-3.0 kg). 66% of patients had a weight of < 2.5 kg at the time of catheterization. Procedural success was 100% with 92% demonstrating no residual shunt at the first postprocedure echocardiogram. Complications included 2 device embolizations that required retrieval and repeat device closure. There was no procedural mortality. In 59 out of 62 patients for whom data was available, all had improvement in respiratory support (P <.02) at median time of 48 days (5-275 days).

Conclusions: With the availability of devices and techniques that enable delivery in small neonates, transcatheter closure of the PDA is an effective and low morbidity alternative to traditional medical and surgical therapies.

Glutathione supplementation decreases mortality and congenital heart disease in an avian model of fetal alcohol spectrum disorders

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Background: Decades of public education regarding dire consequences of prenatal alcohol exposure (PAE) have not reduced alcohol consumption during pregnancy. PAE can result in fetal alcohol spectrum disorder (FASD). Recent reports have revealed high prevalence of FASD (≥1%), rivaling that of autism. Furthermore, roughly 40% of individuals with FASD have congenital heart defects (CHDs). PAE effects include damage to neural crest cells (NCCs), precursors of the autonomic nervous system and regulators of cardiogenesis. Common defects seen in FAE and in our avian FASD model include ventricular septal defects, atrial septal defects, absent major vessels, abnormal atrioventricular valves, and hypertrophic ventricles.

Purpose: Animal studies support that alcohol alters the one-carbon cycle and methylation and promotes oxidative stress. Analysis of human buccal samples reveal signature DNA methylation changes in FASD individuals that may become useful as biomarkers and elucidate mechanisms. Protecting NCCs by supplementation with methyl donors and anti-oxidants may reduce the effects of PAE. Our previous studies have shown that betaine, a methyl-donor, has efficacy in alleviating or preventing cardiac defects associated with CHDs in an avian model. However, as the embryos were only partially rescued, it is essential to test additional compounds. Glutathione is a methyl donor, reduces reactive oxygen species, and is readily available, making it an ideal choice.

Methods: Quail embryos exposed to ethanol with and without glutathione were analyzed at on day 8. This is the first stage at which the quail heart has 4 chambers. Survival, gross body, eye, and heart abnormalities were assessed. Hearts were then optically cleared and imaged with optical coherence tomography for complete phenotyping.

Results: Our preliminary data shows an optimal dose of 1μM glutathione increased survival from 46% to 84% and decreased gross body or heart defects among survivors from 50% to 14% compared to ethanol injected controls. These survival and defect percentages are appreciably better than those seen with betaine (73% and 27% respectively). OCT heart imaging also demonstrates a decrease in heart defects commonly found in FASD. With this data, we suggest that glutathione is an excellent candidate for further analysis to determine if this easily accessible supplement alleviates the effects of PAE.

Mortality and defects among survivors in avian embryos injected with ethanol, with and without glutathione or betaine.

Conclusions: Control survival is high with no defects among survivors. Those embryos injected with ethanol have a high death rate, and half of the survivors have gross body or heart defects. Supplementation with betaine greatly improves survival and decreases defects. This effect is more pronounced with glutathione supplementation, as the survival of these embryos approaches that of controls.

Impact of left-sided lesions in pediatric patients having surgery for Ebstein anomaly

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Purpose: Pediatric patients undergoing tricuspid valve intervention for Ebstein anomaly (EA) are at risk for postoperative morbidity and mortality. The impact of left-sided abnormalities (LSA), including left ventricular noncompaction (LVNC) as well as abnormalities of the mitral and aortic valves, on early cardiac postoperative outcomes is unknown. We sought to determine factors that impact early postoperative outcomes in pediatric patients undergoing cone repair (CR) for EA.

Methods: We performed a retrospective review of all children 1 to 18 years of age undergoing CR for EA at our institution between 6/1/2007-6/20/2017. We assessed demographic and intraoperative factors, and their relationship to outcomes in patients with and without LSA. Data were analyzed using a student *t* test for mean values and 2-tail Fisher's exact for categorical data.

Results: One hundred sixty-eight patients (mean age 9.1 ± 4.9 years, 54% males) had CR for EA. Twenty-four (14%) had LSA: LVNC (n=1), mitral valve abnormality (n=18), bicuspid aortic valve (n=5). Mitral valve abnormalities included mitral valve prolapse (n = 11), cleft mitral valve (n = 2), double orifice mitral valve (n = 1), and parachute mitral valve (n = 1). Mean cardiopulmonary bypass time was 113 ±26 minutes and mean aortic cross clamp time 83 ± 24 minutes. During the postoperative course, mean length of mechanical ventilation was 1 ± 1 day, mean ICU length of stay (LOS) 4 ± 3 days, and mean hospital LOS 8 ±4 days. Postoperative complications included tricuspid valve reoperation (n=1), postoperative ECMO (n=1), delayed sternal closure (n= 2). There was no postoperative mortality. Six patients (25%) had clinically significant arrhythmias requiring outpatient treatment. LSA was associated with longer CPB (113 ±26 minutes versus 96 ±26 minutes, 0.004). LSA was not associated with longer mechanical ventilation time, ICU or hospital LOS, or increased postoperative complications. Peak lactate was 8 ±1 for patients with LSA versus 7 ±0 for patients without LSA (P = .11).

Conclusion: LSA are observed in 14% of children with EA. The majority had an abnormality of the mitral valve (75%) usually mitral valve prolapse. The presence of LSA in pediatric EA did increase cardiopulmonary bypass time; however, no significant increase in LOS or postoperative morbidity was appreciated.

Association between surgical volume and mortality in infants after total anomalous pulmonary venous repair

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Background: In some congenital heart lesions, higher institutional surgical volume has been associated with better survival than lower volume centers. The relationship between institutional surgical volume and mortality in infants after total anomalous pulmonary vein return (TAPVR) repair has not been well-explored.

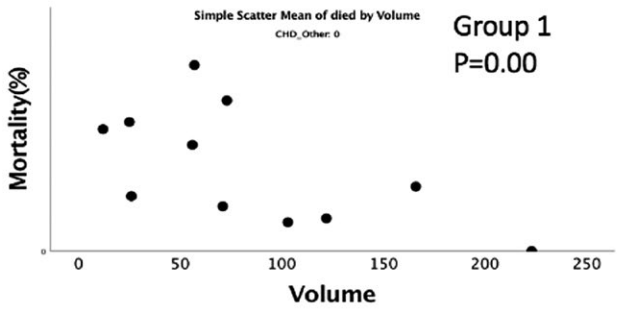
Methods: The Texas Public Use Inpatient Data File was queried for hospitalizations including TAPVR repair in children

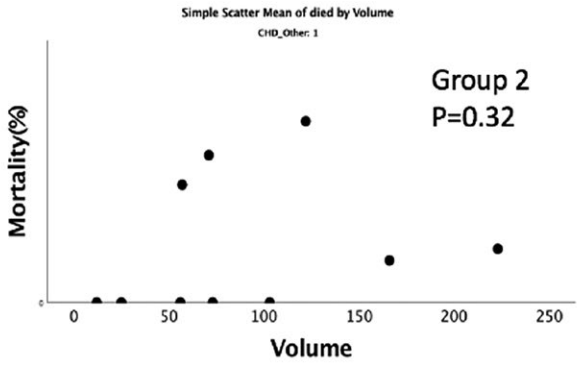
Results: Of 967 hospitalizations, 62% were male. By univariate analysis, variables associated with increased mortality were low birth weight, preterm birth, and extracardiac birth defects/genetic disorders. When examining by subgroup, Group 1 had the lowest mortality (n=599, mortality=6%), compared to Group 2 (n=74, mortality=13.5%, Group 3 (n=129, mortality=20%), and Group 4 (n=173, mortality=25%. Overall, lower volume institutions had higher surgical mortality (P=.012). When examining by subgroup, this relationship was only present in Group.

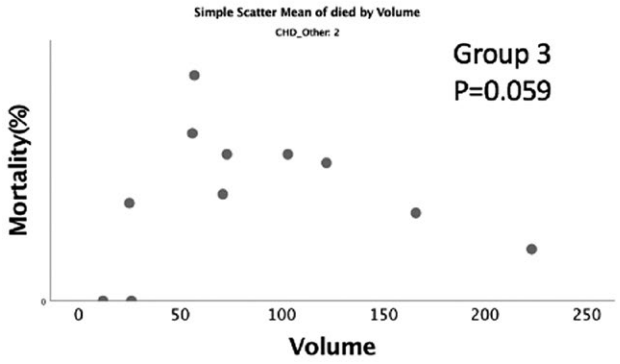
Conclusions: Higher volume centers have lower mortality in hospitalizations for simple TAPVR repair but not with other congenital heart disease including heterotaxy and single ventricle lesions.

TABLE 1 Patient characteristics with TAPVR repair <1 yr of age and mortality, Texas Public Use Inpatient Data File 1999-2016

Pt Characteristics	Died No N=861	Yes N=114	P
Sex, n (%)			0.127
Male	531 (88%)	70 (12%)	
Female	328 (88%)	42 (12%)	
Race/Ethnicity, n(%)			0.87
Non-Hispanic White	211 (88%)	28 (12%)	
Non-Hispanic Black	54 (92%)	5 (8%)	
Hispanic	478 (88%)	64 (12%)	
Other	118 (87%)	17 (13%)	
Low birth weight, n (%)	36 (78%)	10(22%)	0.03
Birth defects, n(%)	257(83%)	53(17%)	<0.001
Preterm birth, n(%)	49(75%)	16(25%)	0.002
Discharge year			0.023
Isolated TAPVR	563(93%)	36 (6%)	
TAPVR with CHD			<0.000
Bi-ventricle	64(86%)	10(13%)	
Heterotaxy	104(80%)	25(20%)	
One ventricle	130(75%)	43(25%)	







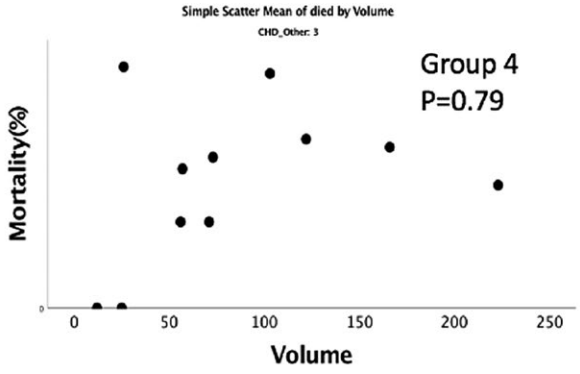


FIGURE 1 Mortality and surgical volume

RESEARCH ABSTRACT PRESENTATIONS: SATURDAY, NOVEMBER 3, 2018: 8:30 AM-6:00 PM

Hospital survival after surgical palliation of truncus arteriosus with interrupted aortic arch: Results from a multiinstitutional database

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Introduction: Truncus arteriosus (TA) is a major congenital cardiac malformation that requires surgical repair in the first few weeks of life. Interrupted aortic arch (IAA) is an associated malformation that significantly impacts the complexity of the TA operation. The aim of this study was to examine hospital survival and morbidity of patients with TA with and without an IAA.

Methods: Data was gathered from the Vizient clinical database, formerly University HealthSystem Consortium (UHC), which encompasses more than 160 academic medical centers in the United States. The database was queried for patients admitted from 2002 to 2016 who were ≤4 months of age at initial admission and diagnosed with TA and underwent complete surgical repair during that hospitalization. The cohort was further categorized as truncus arteriosus alone (TA) and truncus arteriosus with interrupted aortic arch

(TA-IAA). The data were analyzed for in-hospital mortality, length of stay (LOS), and comorbid conditions. Comorbid conditions were defined using the International Statistical Classification of Diseases and Related Health problems (ICD-9 and 10) codes. Logistic regression (mortality) and negative binomial regression (length of stay) were used to compare outcomes for patients undergoing TA or TA-IAA, adjusting for premorbid conditions.

Results: Of the 645 patients with TA who underwent surgery, 98 (15%) had TA with an IAA. There were no differences in the gender, race, or the presence of prematurity, congenital malformations or chromosomal abnormalities between patients with and without IAA. There was no difference in mortality between TA and TA-IAA (13.7% and 18.4%, p-value = .227). No comorbid conditions were associated with an increased mortality in either group. Patients with TA-IAA had a longer LOS compared to those without IAA (40.3 versus 50.1 days, *P* = .028). Holding all comorbid factors constant, TA-IAA was associated with a 1.37 times greater LOS than TA. Prematurity (1.66), DiGeorge Syndrome (1.27) and congenital malformations of the respiratory (1.73), digestive (1.57), urinary 1.38), nervous (1.35), and musculoskeletal (1.27) systems were also associated with a significantly increased LOS (times greater).

Conclusion: The addition of IAA to TA is associated with an increased LOS, but does not increase in-hospital mortality. Premorbid conditions such as prematurity, DiGeorge syndrome, and congenital malformations of the respiratory, digestive, urinary, nervous, and musculoskeletal systems were also associated with an increased LOS, but not mortality.

Prognostic value of serum galectin-3 in infants with congenital heart disease presenting with acute heart failure

Doaa Shahbah MD, PhD[1]; Eman Mogazy MD, PhD[2]; Shimaa Eltokhi MD[3]; Marwa Hasan MD, PhD[4], [1]Cleveland Clinic Foundation, Associate professor of pediatrics, Zagazig University faculty of Medicine, Cleveland, Ohio, [2]Faculty of Medicine, Zagazig University, Zagazig, Ash Sharqiyah, Egypt, [3]Zagazig General hospital, Zagazig, Ash Sharqiyah, Egypt, [4]Faculty of medicine, Zagazig University, Zagazig, Ash Sharqiyah, Egypt

Background: Heart failure (HF) in children is most commonly attributable to coexistent congenital heart disease (CHD). Galectin-3 is a biomarker of inflammation and fibrosis which is strongly associated with adverse remodeling and ventricular dysfunction.

Purpose: To investigate if serum galectin-3 level could predict the short term outcome in infants with CHD presenting with acute HF and to assess any correlation between its level and HF severity as detected by Ross classification and 2D echocardiography.

Methods: The study included 38 infants (2 months -2 years old) with HF due to CHD admitted to pediatric intensive care unit and cardiology inpatient unit from October 2016 to April 2017. We excluded patients with other causes of HF and those with comorbidities that may increase galectin-3 such as renal failure. All patients had complete history, clinical examination including assessment of Ross classification, and 2D echocardiography. Serum galectin-3 level was measured on admission and 72 hours after initiation of HF therapy. We followed patients either for 4 weeks or to hospital discharge whichever came first.

Results: Mean age of patients was 8 ± 11.6 months. 55.3% of patients were males and 44.7% of patients had a history of previous admission with HF. 73.7% of patients had multiple cardiac defects. On admission 57.9% were class II Ross classification with 31.6% and 10.5% class III and IV respectively. 57.9% were hospitalized less than a week while 26.3% and 15.8% stayed for 1-4 weeks and more than 4 weeks respectively. Galectin-3 level was significantly higher in patients with history of previous admission with HF (*P* <.001) and in those with multiple cardiac defects (*P* <.05).

Mean galectin-3 level decreased significantly after 72 hours of therapy relative to that on admission (*P* value <.001). Galectin-3 was positively correlated with Ross classification (*P* value of .04 on admission and 0.001 after 72 hours), length of stay (*P* value of .001) and prognosis (*P* value of .02) (Table 1). Over maximum follow up period of 4 weeks, 78.9% of patients improved, 7.9% did not improve and 13.2% died. Galectin-3 level above 23.5ng/ml was associated with poor prognosis (death or nonimprovement) after individual adjustment of other covariates with 75.0% sensitivity and 86.7% specificity (Figure 1).

Conclusion: On presentation of acute HF in infants with CHD, levels of galectin-3 above 23.5ng/ml were associated with poor prognosis in terms of death or nonimprovement. Galectin-3 concentrations were also positively correlated with severity of HF and prolonged hospital stay. Serum galectin-3 level can be a useful adjunct tool for

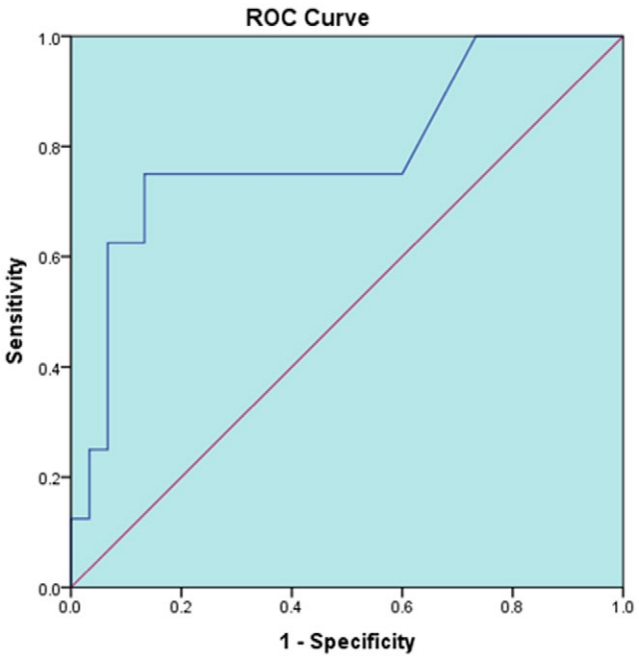

FIGURE 1 Receiver operating characteristics curve of Galectin-3 serum level as indicator of poor prognosis (death or nonimprovement) in studied patients:

risk stratification of acute HF in infants with CHD to predict short term outcome and length of hospital stay. It could also potentially be used to guide therapy.

Cutoff level of serum galectin-3 at 23.5 ng/ml is associated with poor prognosis (death or nonimprovement) with 75.0% sensitivity and 86.7% specificity, 84.2% accuracy and area under curve (AUC) of 0.79.

TABLE 1 Correlation between galectin-3 serum level and other characteristics of studied patients:

Variables	Galectin-3 serum level	
	Spearman's correlation coeffient	P
Age	0.13	0.4
Weight	- 0.01	0.9
Temperature	- 0.27	0.1
Heart rate	0.23	0.2
Respiratory rate	0.22	0.2
Systolic blood pressure	0.30	0.06
Diastolic blood pressure	0.23	0.2
EF	- 0.67	**<0.001 (HS)**
Ross classification:		
At time of admission	0.33	**0.04 (S)**
After 72 hours	0.63	**<0.001 (HS)**
Prognosis	0.39	**0.02 (S)**
Length of stay	0.89	**<0.001 (HS)**

Serum galectin-3 level is positively correlated with Ross classification, prognosis and length of stay and negatively correlated with ejection fraction (EF).

Survival after Norwood procedure in high risk patients

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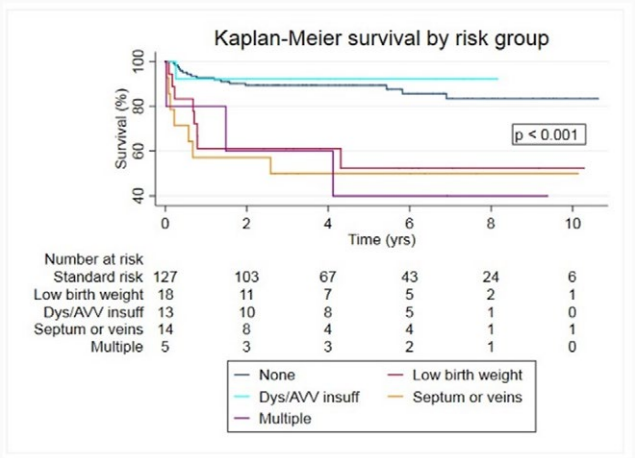
Purpose: Historical studies have identified multiple single ventricle populations at increased risk for mortality following the Norwood procedure. These preoperative risk factors include low birth weight (LBW), restrictive or intact atrial septum or obstructed pulmonary veins, ventricular dysfunction and atrioventricular (AV) valve regurgitation. Data is lacking on longer term outcomes for these high risk populations and it remains unclear whether contemporary survival in these groups has improved along with overall Norwood survival. We sought to report outcomes of the Norwood procedure in high risk patients and compare these results to standard risk patients in the recent era.

Methods: All patients born with hypoplastic left heart syndrome (HLHS) or variants with aortic hypoplasia between 2006 and 2016, who underwent a Norwood procedure at our institution were included. Retrospective review of patient data, including demographics and operative characteristics, was performed and Kaplan-Meier analysis was used to evaluate survival between groups. There were 4 high risk groups created based on preoperative findings of: (1) LBW, defined as birth weight of ≤ 2500 grams, (2) an intact or restrictive atrial septum, defined as having a mean echo gradient of >8mm Hg, or obstructed anomalous pulmonary venous return (septum/veins group), (3) ≥ moderate AV valve regurgitation or ≥ moderate ventricular dysfunction (dys/insuff group) and (4) those with multiple of these high risk co-morbidities (multiple group).

Results: The cohort included 177 patients. There were 50 patients categorized as high risk preoperatively due to LBW (n=18), ventricular dysfunction or AV valve regurgitation (n=13), intact or restrictive atrial septum or obstructed anomalous pulmonary venous return (n=14), or multiple factors (n=5). There were 2 (1.6%) deaths prior to Glenn in the standard risk group with a total of 10 (20%) from the high risk groups (*P*<.0001). The high risk group accounted for 83% of deaths prior to Glenn and 15 out of 24 (63%) deaths prior to 1 year of age. Survival curves can be seen in Figure 1. One year survival differed greatly between groups with highest being standard risk at 92.8% and lowest in those with intact septum/obstructed veins at 57.1%. There was a significant difference between groups in long term survival (*P* <.001).

Conclusions: Outcomes following the Norwood procedure are very good for standard risk patients. Those with preoperative risk factors including LBW, intact or restrictive atrial septum or obstructed anomalous pulmonary veins, ventricular dysfunction or AV valve insufficiency, account for the majority of early mortality following the Norwood procedure. This high risk status does not resolve after Glenn, as their longer term survival continues to diverge from the standard risk group.

Survival Curve



Improving patient experience and education on congenital heart defects: The evolving role of digital heart models, 3D-printing and mobile application

Onyekachukwu Osakwe MD, MPH[1]; Ryan Moore MD[2]; Allison Divanovic MD[3]; Erica Del Grippo MD[1]; Ken Tegtmeyer MD[1]; Nicolas Madsen MD, MPH[3]; Michael Taylor MD[3], [1]Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, [2]CCHMC, Cincinnati, Ohio, [3]Cincinnati Children's Hospital, Cincinnati, Ohio

Background: Effective counseling and good communication are vital to adequately educate families on congenital heart disease (CHD) anatomy, prognosis, and surgical options. In addition, families desire to have easy access to reliable sources of information to further educate themselves and their social networks. Advanced digital and 3D printing technology has enabled the creation of CHD-specific visual aids aimed at improving patient-physician communication and enhancing the patient-family experience.

Objective: To determine ambulatory CHD patient family/caregiver response to new tech-based counseling utilizing 3D printed hearts and a novel interactive mobile application.

Methods: This is a single center prospective study of caregivers of previously diagnosed ambulatory CHD patients. Consenting participants were provided lesion specific education using 3D printed models (Figure 1) and an internally-developed interactive mobile app known as Heartpedia (Figure 2). At the end of the encounter, the caregiver completed a survey on their overall assessment of the novel educational tools and their perception of knowledge regarding the specific CHD described. Questions on the survey included

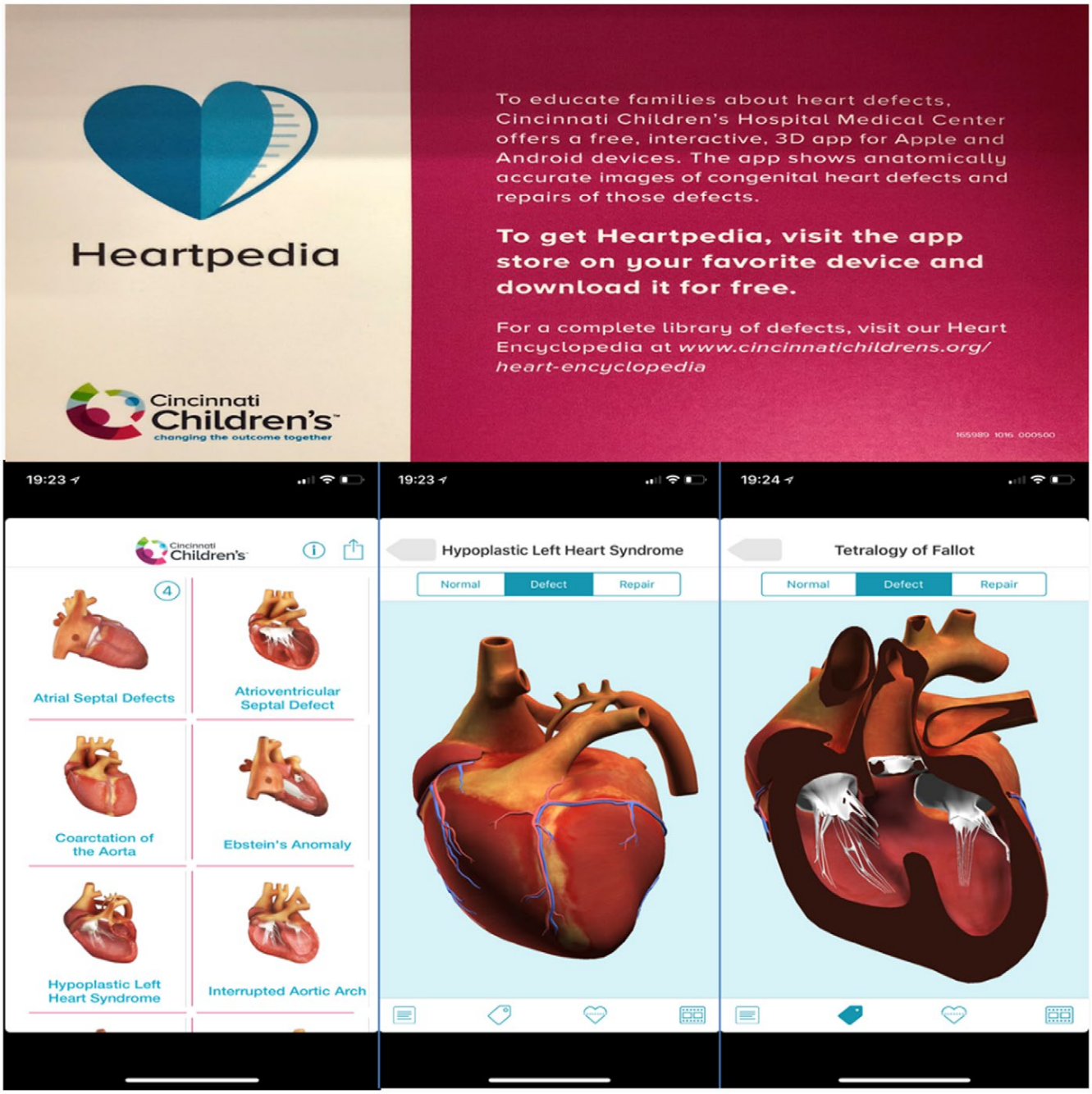


FIGURE 2 Heartpedia mobile application for CHD education in patients and families. Heartpedia App

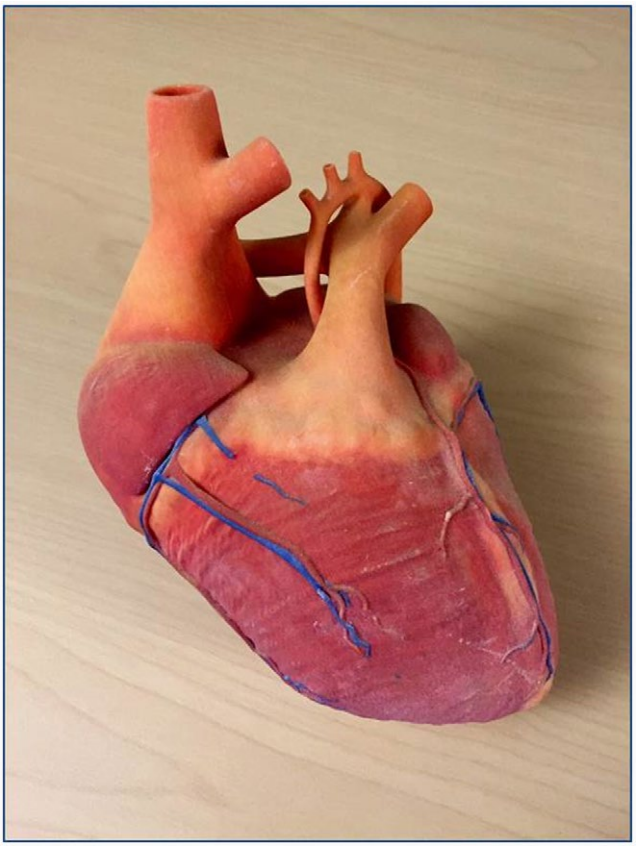


FIGURE 1 3D printed model of hypoplastic left heart syndrome 3D model

recalling details related to the patient specific CHD condition. Additionally, the caregiver's satisfaction and experience with the new educational tools compared to prior encounters was assessed, and the educational tool preference (3D model vs. app) was recorded. Simple *t* test was used to assess for significant differences with the current encounter, compared to prior encounters.

Results: Of the total 75 participants, 96% of caregivers reported that the 3D model and mobile application was "very useful" or "extremely useful" in helping them understand their child's CHD. The caregiver satisfaction was reported as "significantly improved" ($P<.01$), and their perceived understanding of their child's CHD was also "significantly improved" ($P<.01$). The caregivers also report "significantly improved" confidence when describing their child's CHD to their social network and their healthcare provider ($P<.01$). The caregivers also perceived "improved effort" by the physician with regard to including their perspectives when explaining their child's CHD ($P<.01$). The mobile app was preferred to the 3D printed heart models in this cohort.

Conclusions: Novel educational tools, such as 3D printed models and an interactive mobile ap, introduced into ambulatory follow up visits for CHD patients greatly enhance the caregivers' perceived understanding and confidence regarding the details of their child's CHD. Future studies to assess if these tools can improve teen and young adult CHD transitioning to adult providers, as well as, quality of life outcomes tied to anxiety and perceived knowledge are next to be evaluated.

Ventricular arterial coupling by real time 3D echocardiography in children following chemotherapy: A novel left ventricle functional parameter

Sanjeev Aggarwal MD; Gilda Kadiu RDCS, FASE, Childrens Hospital of Michigan, Detroit, Michigan

Background: Although, anthracyclines are efficacious in pediatric cancers, their utilityis limited by progressive cardiotoxicity. Ventricular arterial coupling (VAC), a ratio of left ventricle (LV) elastance and arterial system afterload, relates to outcomes in heart failure in adults. An optimal relationship between the arterial afterload and LV elastance is required for LV efficiency. However, data on the role of VAC in children following chemotherapy is lacking. We hypothesized that VAC will be abnormal in children even many years following completion of anthracycline chemotherapy compared to normal population.

Objective: To compare left ventricle elastance (Ees), arterial elastance (Ea) and VAC in asymptomatic children with normal shortening fractions following completion of anthracycline chemotherapy vs. normal controls.

Method: This was a retrospective echocardiogram review of patients who had received anthracycline chemotherapy for pediatric cancers >1 year earlier. Patients who had undergone bone marrow transplant were excluded. Patients with chest pain, murmur or syncope and normal echocardiograms acted as controls. Demographic, ECHO data, and mean blood pressure (MBP) were collected. Echocardiograms (Philips iE33) were read by a single reader blinded to demographic data. A 3 D real time full volume data consists of 4 adjacent subvolumes over four consecutive beats acquired during a breath-hold to minimize the artifacts and analyzed offline (Tomtec Inc software). Using the 3D volume analysis, LV endiastolic (EDV), LV endsystolic (ESV) volumes and stroke volume (SV) were measured. These echo measures were indexed to BSA. Arterial elastance (Ea) and ventricular elastance (Ees) were calculated as MBP/SVi and MBP/ ESVi respectively. VAC is ratio of Ea/Ees and is calculated as a ratio of ESVi/SVi. Statistical analysis (SPSS ver. 22) included student *t* test and chi-square test to compare groups. *P* value < .05 was considered significant.

Results: Our cohort (n=113) included 31 (27%) subjects who had received anthracyclines. These groups were similar in age at echocardiogram and gender distribution (Table 1). Chemotherapy group had significant higher diastolic blood pressure compared to normal control. The diastolic blood pressure and LVESV indexed were significantly higher in chemotherapy group compared to normal group. Ea was significantly higher and Ees was significantly lower in chemotherapy group. The VAC was significantly higher in chemotherapy group compared to normal controls (0.75 ± 0.23 vs. 0.55 ± 0.13, $P=.0001$) (Table 1).

Conclusions: Even years after anthracycline chemotherapy, asymptomatic pediatric subjects have significant abnormalities in arterial and ventricle elastance as well as their ratio (VAC). These findings suggest the increased arterial elastance or arterial bed stiffness and

TABLE 1 Comparison of demographic and echo parameters between chemotherapy and normal group

Parameters (mean ± SD) (n%)	Chemotherapy (n=31)	Normal Controls (n= 82)	P value
Age	13.1±3.3	12.1±3.9	0.2
Gender (male)	50 (61%)	21 (68%)	0.33
Height (cm)	154.5 ± 21.1	151.6 ± 21.5	0.5
Weight (kg)	55.6 ± 24.5	48.6 ± 20.4	0.13
BSA (m^2)	1.52 ± 0.43	1.41 ± 0.39	0.23
Systolic BP (mmHg)	115 ± 10.5	112 ± 12.4	0.27
Diastolic BP (mmHg)	**64.5 ± 9.6**	**61.1 ± 7.5**	**0.04**
Indexed ESV (ml/m^2)	**24.1 ± 7.7**	**19.3 ± 5.6**	**0.0001**
Indexed EDV (ml/m^2)	56.9 ± 12.4	55.0 ± 12.0	0.46
Ejection fraction	**57.9±7.3**	**64.9± 5.4**	**0.0001**
Indexed SV (ml/m^2)	32.7 ± 7.9	35.7 ± 8.0	0.08
Ea (mm Hg m^2/ml)	**2.59 ± 0.53**	**2.3 ± 0.56**	**0.01**
Ees (mm Hg m^2/ml)	**3.6 ± 0.9**	**4.3 ± 1.2**	**0.0001**
VAC	**0.75 ± 0.23**	**0.55 ± 0.13**	**0.0001**

suboptimal compensation by left ventricle may be one of the reasons for heart failure in patients who have received anthracyclines. Further longitudinal studies are required to assess prognostic implications of this novel echo functional parameter.

Fetal echocardiogram is useful for screening fetuses with a family history of cardiomyopathy

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Background: Cardiomyopathy is highly heritable with familial forms accounting for 30-40% of cases. Cardiomyopathy can present in the fetus and be detected by fetal echocardiogram. Data on the utility of fetal echocardiograms in patients with a family history of cardiomyopathy is limited and is not a current indication for fetal echocardiogram on the most recent American Heart Association guidelines. We investigated how useful fetal echocardiography is with a family history of cardiomyopathy.

Methods: We screened all fetal echocardiograms performed at our institution for the past 5 years for the indication of family history of cardiomyopathy. We excluded patients with a family history of hypertrophic cardiomyopathy, since nonsyndromic forms are not typically clinically significant in the newborn period.

Results: Twenty-six patients were identified who had fetal echocardiograms performed due to a family history of cardiomyopathy. Three out of 26 patients (11.5%) had findings of decreased ventricular function and dilation consistent with cardiomyopathy. All who had cardiomyopathy on fetal echocardiogram had parents with genetic mutations (2 maternal, 1 paternal), including one mother who had a cardiac transplant at age 8 for dilated cardiomyopathy. There were two MYH7 and one TNNT2 mutations. All 3 affected infants had prenatal planning for high level care and were transferred to our tertiary care facility immediately after birth for cardiology evaluation and management.

The first patient required prostaglandins given a significant coarctation in addition to left ventricular noncompaction cardiomyopathy (dilated phenotype). The patient had a coarctation repair in the newborn period and subsequently required inotropic support (milrinone) for progressively worsening left ventricular systolic function. She was transplanted at age 2 months. The second patient had dilated cardiomyopathy and severely diminished left ventricular systolic function. He was supported with milrinone but had progressive clinical decline. He was supported with ECMO for 1 week but recovered with subsequent milrinone support and finally transition to oral heart failure medications. He is now 3 years old on outpatient heart failure therapy and thriving. The third infant required an initial hospitalization for feeding difficulty and initiation of oral heart failure medication, but avoided inotropic or advanced support needs. She is 3 years old and thriving on heart failure medications. None of the patients with normal function on fetal echocardiogram had evidence of cardiomyopathy on newborn echocardiogram.

Conclusion: In this study we found 11.5% of fetal echocardiograms performed for family history of cardiomyopathy had findings of cardiomyopathy on fetal echocardiography significant enough to require prenatal planning. We recommend patients with a strong family history of cardiomyopathy in the mother or father, especially those with known genetic mutations associated with cardiomyopathy, have screening fetal echocardiograms performed. Fetal echocardiograms in this patient population are useful for monitoring of the fetus and for immediate postnatal planning.

Percent deviation from midline coaptation: When to suspect bicuspid aortic valve

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Background: Bicuspid aortic valve (BAV) is typically diagnosed by visualization of aortic valve leaflet opening during systole in parasternal short-axis (PSAX) echocardiographic images. The leaflets are often seen poorly in PSAX view due to suboptimal acoustic windows or poor imaging techniques but are usually seen well in parasternal long axis (PLAX) view. Although the eccentric leaflet coaptation in PLAX view has been previously described the degree of eccentricity that should alert clinicians of the presence of BAV is not established in children. The purpose of this study is to establish the degree of eccentric coaptation that separates BAV cases from controls and assess its diagnostic accuracy.

Methods: This was a retrospective study of cases with prior diagnosis of BAV and age-matched controls who had echocardiogram for murmur, syncope or chest pain. PSAX images were reviewed to confirm the presence of BAV in cases and its absence in controls. The subjects with indeterminate valve morphology were excluded. The two groups and the BAV subtypes were compared for percent deviation from midline (PDFM) coaptation (Figure 1). Student's *t* test was performed to compare the means. Appropriate cutoff for PDFM that separated the patients with BAV from controls (Figure 2) was established by receiver operating characteristic (ROC) analysis.

Results: After excluding 30 indeterminate subjects, 84 BAV cases and 223 controls (mean age- 9.6 year versus 8.9 year, P =.33) were identified. The BAV group had significantly eccentric coaptation of leaflets compared to controls (mean±SD PDFM: 17.1±14% vs. 6±4.6%, P < .001). A diagnostic cutoff PDFM of 11.2% was established (Figure 2) yielding 58% sensitivity, 87% specificity, 4.3% positive and 99.5% negative predictive value—assuming 1% population prevalence. Interestingly patients with right/left leaflet fusion (N: 48) had an anterior while those with right-noncoronary fusion (N: 25) had a posterior deviation of coaptation (PDFM +10.8% vs. -6.6%, P < .001).

Conclusion: Patients with BAV have significant eccentric coaptation of the leaflets from midline compared to those with trileaflet morphology. The deviation of coaptation from midline below 11.2% in PLAX echocardiographic images reasonably assures the absence of BAV with strong negative predictive value (99.5%). The deviation above 11.2% indicates the presence of BAV with fair sensitivity and this should alert clinicians to further image the aortic valve if PSAX images are inadequate.

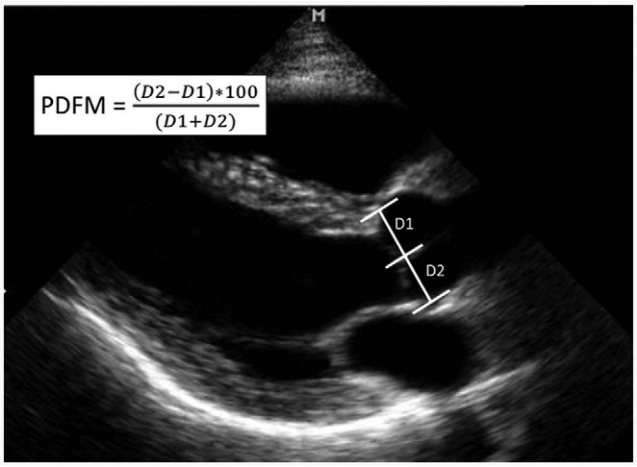


FIGURE 1 Measurement technique in PLAX view. Abbreviations: D1, distance from the proximal end of aortic valve leaflet coaptation to anterior aortic wall; D2, distance from the coaptation to posterior aortic wall; PDFM, percent deviation from midline coaptation.

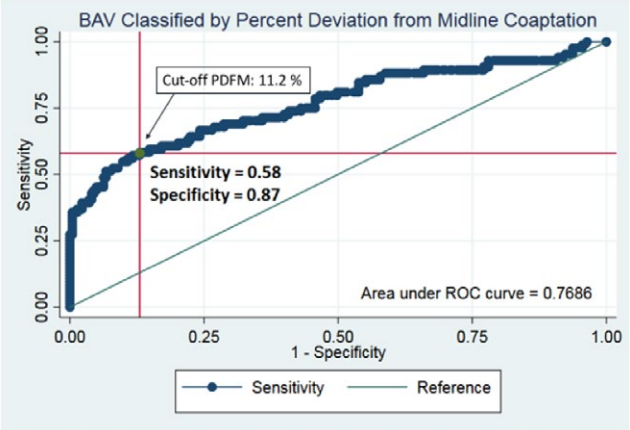


FIGURE 2 ROC curve demonstrating the optimal cutoff PDFM separating the BAV group from controls.

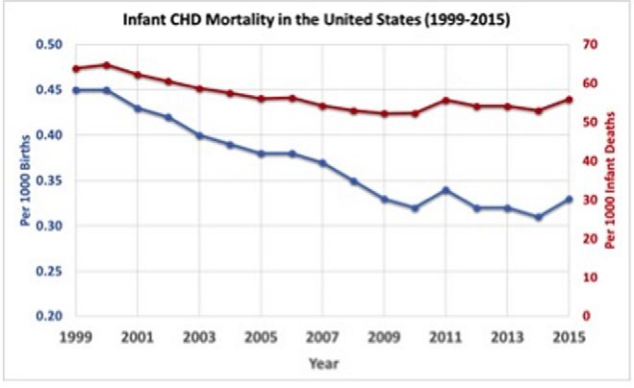
Recent trends in infant mortality due to congenital heart disease in the United States

Lamya Mubayed MD, FAAP[1]; *Sadeer Al-Kindi MD*[2], [1]*Rush University Medical Centre, Chicago, Illinois*, [2]*University Hospitals and Harrington Heart and Vascular Institute, Cleveland, Ohio*

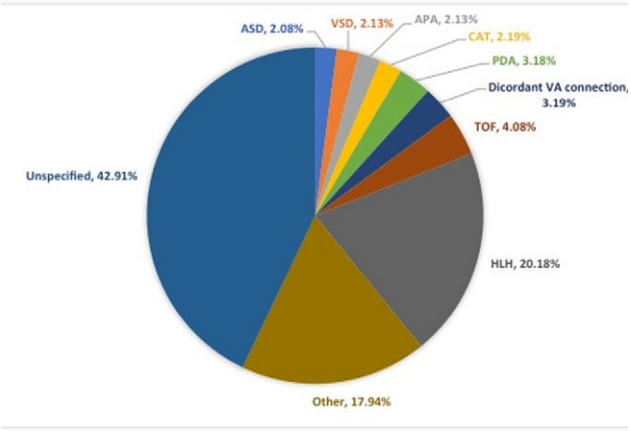
Purpose: Overall infant mortality rates in the US have demonstrated a downward trend, however, rates related to congenital heart disease (CHD) in particular remain undefined. We sought to specifically examine the recent trends in infant mortality secondary to CHD in the US over the past few years.

Methods: We conducted a retrospective analysis of infant mortality rates due to CHD over a 16-year period (1999-2015). Using infant death records linked with births in the United States (1999-2015), we selected all infants who died from congenital heart disease (International Classification of Diseases, 10th revision: Q20-Q26) and analyzed the trends in infant CHD mortality and subtypes.

CHD Mortality trend in the US



CHD Mortality by diagnosis



Results: Between 1999 and 2015, there were 450 043 infant deaths, of which 25 602 (5.7%) were due to CHD. The incidence of CHD mortality was 0.37 per 1000 births and 57 per 1000 infant deaths. The incidence of infant CHD mortality, per 1000 births, decreased from 0.45 in 1999 to 0.33 in 2015, and from 64 to 56 per 1000 infant deaths. Rates were noted to be higher in infants born to black/African American mothers (0.47) compared with white mothers (0.36) per 1000 births. The incidence of CHD mortality increased with age from 0.01 per 1000 births.

Conclusion: Congenital heart disease is responsible for 1 in 18 infant deaths in the US. CHD mortality in the US has declined by 26% in the past two decades, possibly owing to advances in infant cardiac interventions and improved care of premature births. Despite the decline in CHD mortality, significant racial disparities still exist.

Cardiomegaly on chest radiograph—Is an echocardiogram always indicated?

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Purpose: Cardiomegaly on chest radiographs (CXR) in pediatric patients leads to multiple tests including blood work, electrocardiograms (EKG) and echocardiograms. The purposes of this study were to determine the positive predictive value (PPV) of cardiomegaly on CXR in predicting structural/functional heart disease and to determine the PPV and negative predictive value (NPV) of B-type natriuretic peptide (BNP) and an abnormal EKG in a patient with cardiomegaly on CXR in predicting subsequent heart disease.

TABLE 1 Table demonstrating patient demographics

Patient characteristic	n	Overall (N=477)	CHD, Yes (N=147, 30.8%)	CHD, No (N=330, 69.2%)	p-value
Age, continuous	477	2.2y (56d – 11.2y)	2.1m (18d – 1.8y)	4.6y (4.6m – 12.8y)	**<0.001**
Age, categorical	477				**<0.001**
0d – 1m		89 (18.7%)	50 (34.0%)	39 (11.8%)	
1m – 1y		126 (26.4%)	55 (37.4%)	71 (21.5%)	
>1y		262 (54.9%)	42 (28.6%)	220 (66.7%)	
Weight, kg	477	12.0 (3.6 – 32.7)	3.9 (2.4 – 12.0)	17.8 (5.2 – 41.5)	**<0.001**
Sex	477				0.8111
Male		257 (53.9%)	78 (53.1%)	179 (54.2%)	
Female		220 (46.1%)	69 (46.9%)	151 (45.8%)	
Encounter Setting	477				**<0.001**
Emergency Room		249 (52.2%)	59 (40.1%)	190 (57.6%)	
Inpatient		216 (45.3%)	86 (58.5%)	130 (39.4%)	
Outpatient		12 (2.5%)	2 (1.4%)	10 (3.0%)	
Ethnicity	477				0.0527
Hispanic		38 (8.0%)	17 (11.6%)	21 (6.4%)	
Non-Hispanic		439 (92.0%)	130 (88.4%)	309 (93.6%)	
Race	477				0.1977
White		334 (70.0%)	105 (71.4%)	229 (69.4%)	
African-American		118 (24.7%)	31 (21.1%)	87 (26.4%)	
Asian		25 (5.2%)	11 (7.5%)	14 (4.2%)	
Cardiovascular Physical Exam	476				**<0.001**
Normal		357 (75.0%)	75 (51.0%)	282 (85.7%)	
Pathologic murmur		81 (17.0%)	56 (38.1%)	25 (7.6%)	
Innocent murmur		17 (3.6%)	7 (4.8%)	10 (3.0%)	
Other		21 (4.4%)	9 (6.1%)	12 (3.6%)	
Current medications	477				**0.004**
None		124 (26.0%)	51 (34.7%)	73 (22.1%)	
Non-cardiac		353 (74.0%)	96 (65.3%)	257 (77.9%)	

Values reported as N (%) or Median (25^{th} – 75^{th}). Comparisons between groups were made using Wilcoxon rank-sum tests for continuous variables and Chi-square tests for categorical variables

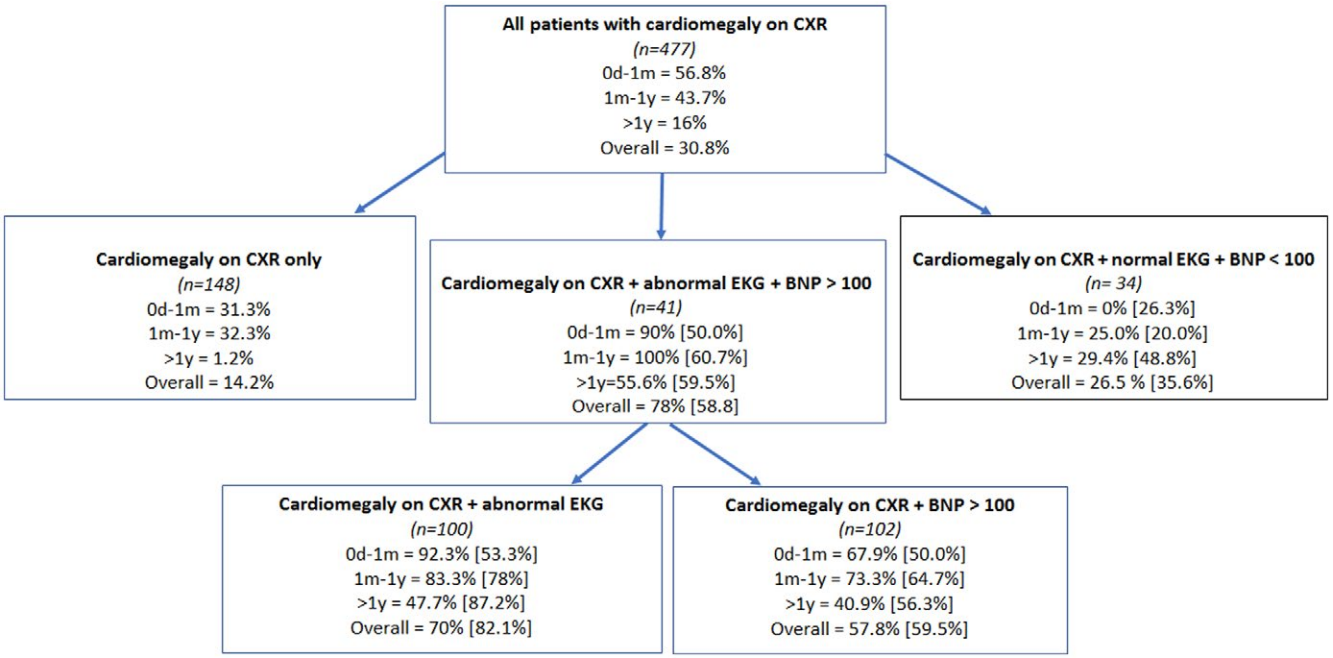

FIGURE 1 showing the PPV [NPV] of cardiomegaly on chest radiographs with/without an abnormal EKG and BNP level in predicting subsequent heart disease stratified by age

Methods: We performed a retrospective chart review of pediatric patients (0-21 years) with cardiomegaly on their initial CXR between January 2015-December 2017. Patients without a subsequent echocardiogram and those with known congenital heart disease were excluded. The interpreting radiologist's report was used to classify a patient as having cardiomegaly. A patient was deemed to have structural/functional heart disease consistent with cardiomegaly if they had depressed ventricular function (including myocarditis/cardiomyopathy), a pericardial effusion or structural defects leading to a left to right shunt, left/right sided obstruction and significant valvular disease. The physical exam findings, EKG interpretation and the BNP value when obtained were also recorded. The PPV of CXR and the PPV/NPV of the other tests were then calculated using contingency tables.

Results: There were 282 618 patients with a CXR during the study timeframe, with 1544 having cardiomegaly on their initial CXR. Of these, 477 met inclusion criteria (Table 1), with 147 having structural/functional heart disease on echocardiogram. Additional testing included an EKG in 275 patients, BNP in 176, and both tests in 122. The overall PPV of cardiomegaly on CXR was 31%. The PPV increased if there was either an abnormal EKG or a BNP>100 pg/ml and further increased if both of these were present (Figure 1). When stratified by age, the PPV of abnormal testing was higher in patients < 1 year of age.

Conclusions: Cardiomegaly on CXR is an overall rare finding. However, when present, it often predicts the presence of structural/functional heart disease, particularly in infants. Further testing with EKG and BNP can better predict who may have heart disease, but it may not eliminate the need for echocardiography.

Impact of a multi-interventional nutrition program on the outcomes of newborns requiring surgery for congenital heart disease

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Purpose: Up to 65% of children with congenital heart disease (CHD) suffer from malnutrition. The goal of our study was to evaluate how health and nutrition outcomes changed in newborns undergoing surgery for CHD at our institution after implementation of our "Pediatric Heart Institute Nutrition Program," which included preoperative and postoperative feeding algorithms to standardize nutritional management.

Methods: This was a single-center cohort study in 3 phases that included newborns with single-ventricle physiology, two-ventricle physiology, and two-ventricle physiology with shunt-dependent pulmonary blood flow who underwent cardiac surgery between September 2008 and July 2015. We evaluated outcomes in the 2 years of preprogram time (Phase 0), in the 2 years after initiation of a standardized postoperative feeding algorithm (Phase 1), and in the 2 years following introduction of the donor breast milk and preoperative feeding programs (Phase 2). The primary outcome was weight-for-age *z*-score (WAZ) change from birth to hospital discharge after neonatal cardiac surgery. Outcomes were evaluated using traditional statistics and quality improvement methods.

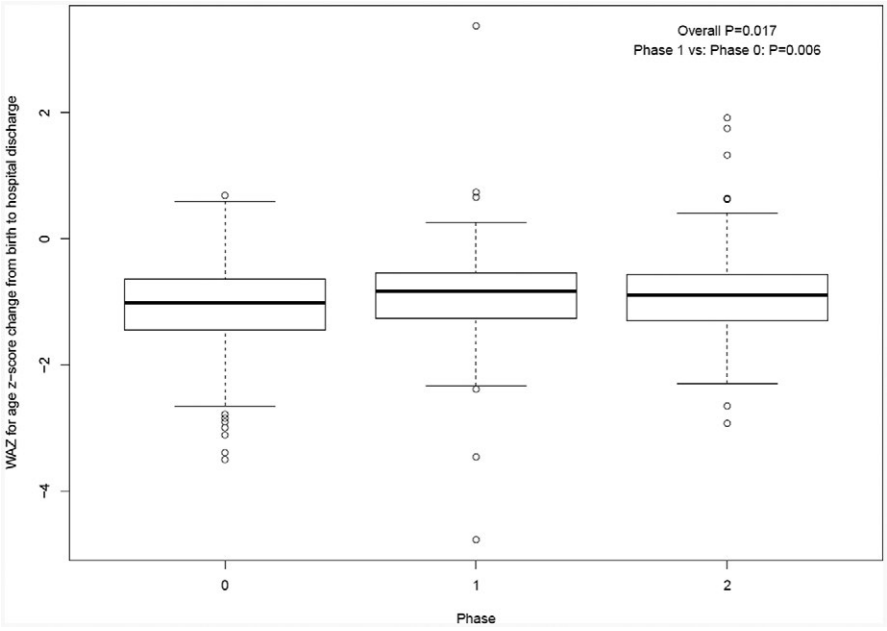

FIGURE 1 Weight-for-age *z*-score (WAZ) change from birth to hospital discharge after neonatal cardiac surgery across study phases.

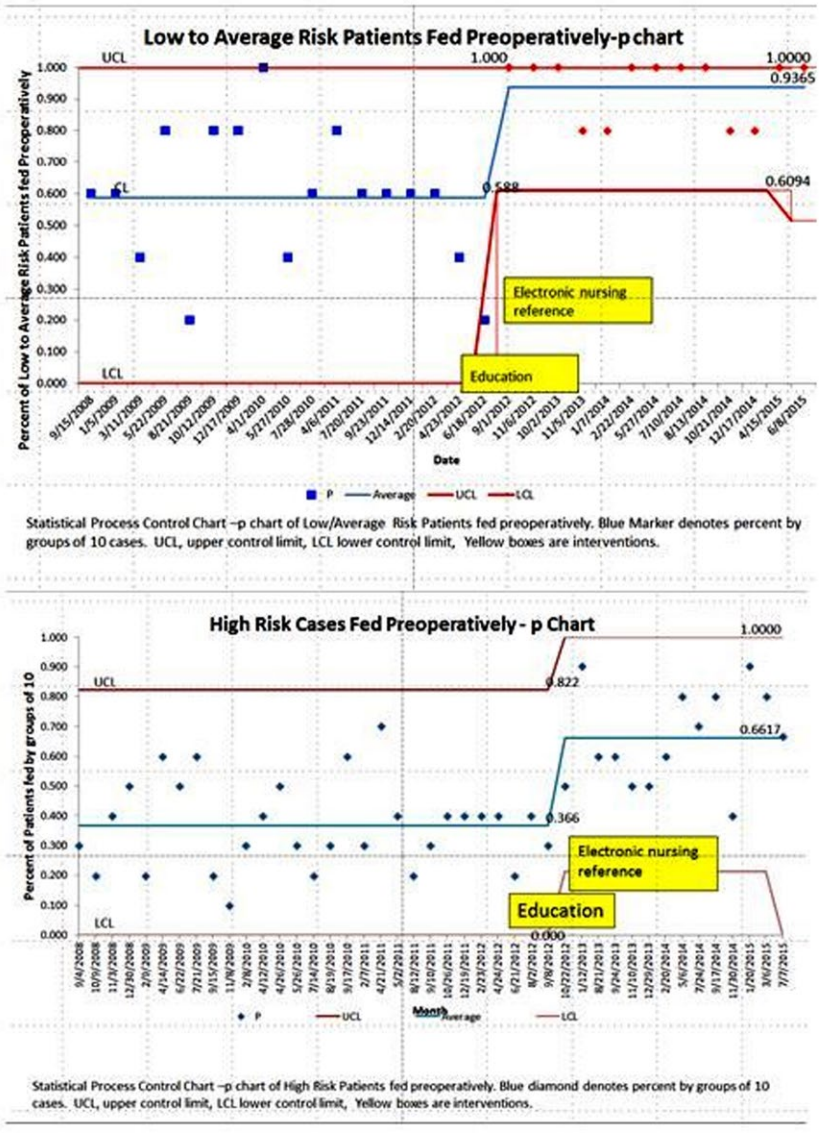

FIGURE 2 Increase in preoperative enteral feeding after institution of our preoperative feeding algorithm, by feeding algorithm group (high-risk and low-to-average risk).

Results: Our study included 570 newborns. WAZ change significantly improved from birth to hospital discharge from phase 0 to phase 1 (−1.02 [IQR −1.45 to −0.63] in phase 0, −0.83 [IQR −1.25 to −0.54] in phase 1; *P*=.006), with this improvement maintained in phase 2 (−0.89 [IQR −1.30 to −0.56]; *P*=.017 across all 3 phases; Figure 1). The improvement in WAZ change remained after adjusting for covariates (*P*=.027). Gastrostomy (G)-tube utilization declined significantly from 25% in phase 0 to 12% in phase 1 and 14% in phase 2 (*P*<.001) without increases in necrotizing enterocolitis (NEC), hospital length of stay, or mortality. Preoperative enteral feeding also significantly improved (47% and 46% in phases 0 and 1 compared to 76% in phase 2; *P*<.001) without increases in NEC, hospital stay, or mortality after institution of our preoperative feeding algorithm, as demonstrated by both traditional statistics and quality improvement methods (Figure 2). Importantly, these gains were not just achieved in newborns with biventricular physiology but also in those with single-ventricle physiology and most notably in those with hypoplastic left heart syndrome (15% and 16% in phases 0 and 1 compared to 43% in phase 2; *P*=.022).

Conclusion: Introduction of a multiinterventional nutrition program was associated with improved weight gain, fewer G-tubes placed for primary feeding, and significantly more newborns with all types of CHD receiving enteral nutrition prior to cardiac surgery without increases in NEC, hospital length of stay, or mortality. Standardization of nutritional practices for newborns with CHD improves growth and reduces morbidity.

Phase 0: preprogram/preintervention time Phase 1: the 2 years after initiation of the postoperative feeding algorithm Phase 2: the 2 years after introduction of the donor breast milk and preoperative feeding programs WAZ change significantly improved from birth to hospital discharge from phase 0 to phase 1 (−1.02 [IQR −1.45 to −0.63] in phase 0, −0.83 [IQR −1.25 to −0.54] in phase 1; *P*=.006), with this improvement maintained in phase 2 (−0.89 [IQR −1.30 to −0.56]; *P*=.017 across all 3 phases).

Feeding algorithms were designed to address feeding needs and to advance feedings differently in patients deemed high-risk versus low-to-average-risk based on their postoperative cardiac physiology/hemodynamics and based on associated noncardiac abnormalities. Preoperative enteral feeding significantly improved after introduction of a preoperative feeding algorithm in both high-risk and low-to-average risk feeding groups as demonstrated by these statistical process control charts.

Atrial septal defect resource utilization analysis

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Purpose: There is wide variation of practice in monitoring secundum atrial septal defects (ASD) by providers. It is generally accepted that ASDs less than 3mm (typically classified as patent foramen ovale (PFO)) close spontaneously by 18 months of age and do not require follow up. Defects greater than 3mm tend to have a more variable course and require additional surveillance. While the majority of ASDs resolve spontaneously or are hemodynamically insignificant, some require closure in the cath lab or OR. The optimal time for ASD closure is between 4-5 years of age. In practice there is still inconsistency in how often these lesions are monitored resulting in overutilization of routine studies. This lack of uniformity presents an opportunity to determine the cost savings to be gained if optimal resource utilization was achieved. Therefore, this project aims to determine current utilization patterns for patients with simple secundum ASDs diagnosed within the first year of life and comparing this to the optimal utilization standard. Optimal utilization is defined as an echo and EKG at diagnosis with a clinic visit, follow-up visit 1 year later without testing and another follow-up visit at 3 to 5 years of age with an echo.

Methods: We retrospectively reviewed charts for patients with secundum ASDs diagnosed within the first year of life from 2010-2013. Patients with coexisting cardiac lesions were excluded. Total number of clinic visits, EKG's and echocardiograms were recorded. For patients requiring intervention, only preintervention visits and studies were included. Charge data was obtained and total charge was calculated. Patients were stratified by lesion type: PFO, ASD and ASD requiring intervention; as well as provider type: imaging, general cardiology and cath. The difference in charge from optimal, among groups, was then compared using ANOVA.

Results: Ninety-seven patients were included in the data analysis, 40 had PFOs, 43 had ASDs and 14 had ASDs that required intervention (surgery or catheterization). Mean age at intervention was 4 years old. PFO, ASD and ASD requiring intervention had a statistically significant difference in mean charge above the optimal utilization standard of $1033, $2885 and $5722 respectively (*P*< .02). There was also statistically significant variation in charge above optimal between provider types. Cath providers had a mean charge above optimal of $4961, imaging providers were $2482 above optimal and general cardiologists were $1147 above optimal. Average cost

TABLE 1 Comparison of lesion types

	PFO	ASD	ASD w/ intervention
n	40	43	14
Mean Cost in $ (SD)	$7283 (1898)	$13552 (2979)	$16289 (4441)
*Mean Cost Above Optimal (SD)**	$1033 (1898)	$2885 (2979)	$5722 (4441)
# of Clinic Visits (SD)	1.53 (0.64)	3.47 (1.26)	3.64 (1.15)
# of ECGs (SD)	1.4 (0.67)	3.26 (1.33)	3.43 (1.34)
# of Echos (SD)	1.15 (0.36)	2.16 (0.57)	2.64 (0.84)

*P-values < 0.02
(*SD*): standard deviation

This table compares utilization variables between PFOs, ASDs and ASDs that required intervention.

TABLE 2 Comparison of provider types

	Imaging	General	Cath
n	70	17	6
Mean Cost in $ (SD)	$11321 (4258)	$9429 (2911)	$14089 (5685)
*Mean Cost Above Optimal (SD)**	$2482 (2912)	$1147 (1314)	$4961 (4488)
# of Clinic Visits (SD)	2.69 (1.41)	2.59 (1.46)	2.50 (1.05)
# of ECGs (SD)	2.54 (1.44)	2.24 (1.20)	2.33 (1.21)
# of Echos (SD)	1.80 (0.73)	1.53 (0.51)	2.33 (1.03)

*P-values < 0.02
(*SD*): standard deviation

This table compares utilization variables between provider subspecialties.

savings per patient would be $2530 with a total savings of $242 472 if optimal utilization was followed.

Conclusion: Using optimal utilization and decreasing variation in care could save a significant amount unnecessary charges. Moving forward we will propose an optimal utilization algorithm for the care of any infant diagnosed with an atrial septal defect to decrease practice variation and unnecessary charges.

Reduced neurogenic activity in the subventricular zone after cardiopulmonary bypass in a juvenile porcine model

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Introduction: Impairments in higher-order cognitive and behavioral function are widely observed in children after cardiac surgery with cardiopulmonary bypass (CPB). The subventricular zone (SVZ) in the postnatal/adult brain is the region where most neural stem/progenitor cells (NSPC) originate. Recent studies suggest that the SVZ plays an important role in neocortical growth of the gyrencephalic frontal lobe during postnatal life. However the effect of CPB-induced insults on postnatal SVZ neurogenesis is largely unexplored. The aim of our study was to determine neurogenic activity of the SVZ after CPB using a porcine survival model.

Methods: Three-week-old piglets (n=16) were randomly assigned to control group (no surgery) and severe-CPB insult group (25°C circulatory-arrest for 60min, which reproduces CPB-induced systemic inflammation and ischemia-reperfusion injury). Brains were fixed at either day 3 or week 4 postoperatively. NSPCs and neuroblasts (young neurons) were identified with antibodies directed against Sox2 and doublecortin. GFAP was used to identify radial-glia like cells (i.e. neural stem cells) of which processes length is correlated to the SVZ activity. We divided the SVZ into three tiers as previously described in the human SVZ. The SVZ region was subdivided into ventral (V-SVZ) and dorsolateral (DL-SVZ) regions.

Results: A significant decrease in the length of $GFAP^+$ processes was observed three days after severe-CPB insult compared to control. Interestingly this morphological change was seen only in DL-SVZ. Similarly severe-CPB resulted in a significant reduction of NSPC layer thickness of tier-1 but only in DL-SVZ. On the other hand there was no notable difference in NSPC number in tiers 2/3, indicating region-specific effects on the NSPC pool. Recent studies have demonstrated that neuroblasts form migration chains moving tangentially through the SVZ. Although there was no difference in the number of these clusters we found a significant decrease in their surface area after severe-CPB, indicating a disruption of neuroblast migration toward the frontal cortex. Furthermore the number of neuroblasts in the DL-SVZ was remarkably reduced in severe-CPB group. Collectively our results demonstrate diminished neurogenic activity three days after severe-CPB insult. Furthermore four weeks after severe-CPB we observed a significant reduction of NSPC layer thickness of tier-1 in DL-SVZ, indicating continued alteration of SVZ NSPCs after severe-CPB insult.

Discussion: Our data demonstrate prolonged impairments in the NSPC pool following severe-CPB insult. The impact is confined to the dorsolateral region which is the most active neurogenic niche under normal conditions. Our results indicate reduced neurogenic activity in the SVZ and suggest disruption of neuroblast migration towards the frontal cortex after severe-CPB. The frontal cortex is the region responsible for a wide range of higher-order cognitive functions; our study therefore provides novel insights into cellular mechanisms underlying complex neurological impairments in children with congenital heart disease.

Variant classification trends in pediatric cardiac genetics

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Background: Genetic counselors are not universally included in pediatric cardiology practices. As genetic test results become more complicated, families and cardiologists require the expert consultation that genetic counselors provide. Few studies have evaluated the volume or changing distribution of variant classification in pediatric cardiology. Our objective was to quantify the frequency of pediatric cardiac genetic results and to determine the relevant trends for all

types of variants. In particular, we hypothesized that the frequency of variants of uncertain significance (VUS) is increasing, suggesting increased need for genetic counselors in the future.

Methods: In a tertiary pediatric cardiology center, we tabulated all genetic variants seen in cardiomyopathy and arrhythmia clinics. Variants were included if they were reported from CLIA-certified labs from 2006-2017. Variant classification was performed by each CLIA lab. Research testing and family variant confirmation tests were excluded. Frequency over time was compared by variance-weighted least squares regression; correlation by Pearson's method.

Results: Testing returned 520 genetic variants from 251 probands. The average age at testing was 18 years of age (SD 14.2). Over time, genetic counselors interpreted more variants per year (Figure, stacked area plot/left axis) and interpreted more genes per patient (Figure 1, red line/right axis). After CLIA-certified labs adopted new reporting guidelines in 2015, a marked decline in total variants occurred (35.7% decline, $P<.01$) due to the elimination of benign and likely benign variant reporting. In contrast, the fraction of VUS continued to increase, accelerating after the 2015 guidelines ($P<.01$ for trend, with fraction of VUS over 75% by 2017). The increase in variants is also correlated with the increase in genes tested per patient (R=0.85, $P<.01$).

Conclusions: Pediatric cardiology practitioners are called on to interpret an increasing volume of genetic test results in cardiomyopathy and rhythm disease. In addition, the complexity of results is increasing, as measured by the frequency of VUS. Not only are more variants reported, but more genes are tested; cardiologists may have limited familiarity with newer genes. Finally, new guidelines redefined variant classification in 2015. In a large pediatric cardiology practice, this change was associated with a doubling in actionable variant frequency between 2014 and 2017, as assessed by the total number of VUS, likely pathogenic, and pathogenic variants. The growing volume of results, the growing number of genes to master, and the increasing frequency of variants of uncertain significance demonstrates the need to incorporate genetic counselors into pediatric cardiology practices.

Improving the quality of outpatient pediatric cardiology management plan documentation

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Purpose: Concise, complete physician documentation by subspecialists is imperative. In the case of pediatric patients with congenital heart disease, it is of utmost importance that certain considerations such as the need for subacute bacterial endocarditis (SBE) prophylaxis, need for activity restrictions, and need for additional follow-up and testing be included in outpatient clinic documentation. The objective of our quality improvement initiative was to improve the rate of documentation of complete management plans in outpatient pediatric cardiology clinics from a baseline rate of 64% to 90% over a 2-month period.

Methods: An IRB-approved, retrospective chart review was performed at a single academic center. Data was collected from 600 individual patient charts from 35 clinic days on which at least 10 patients were seen. Documented management plans were considered complete if they included all four of the following: need for SBE prophylaxis; need for activity restrictions; need for additional follow-up; need for additional testing. Two Plan-Do-Study-Act (PDSA) cycles were undertaken and postintervention data was collected for a period of three weeks following each. The first intervention included a formal discussion among participating attending physicians during which the definition of a complete management plan was agreed upon and an optional management plan template was developed. The second intervention involved weekly email reminders to clinical staff re-emphasizing the essential elements to include and encouraging compliance.

Variants Over Time

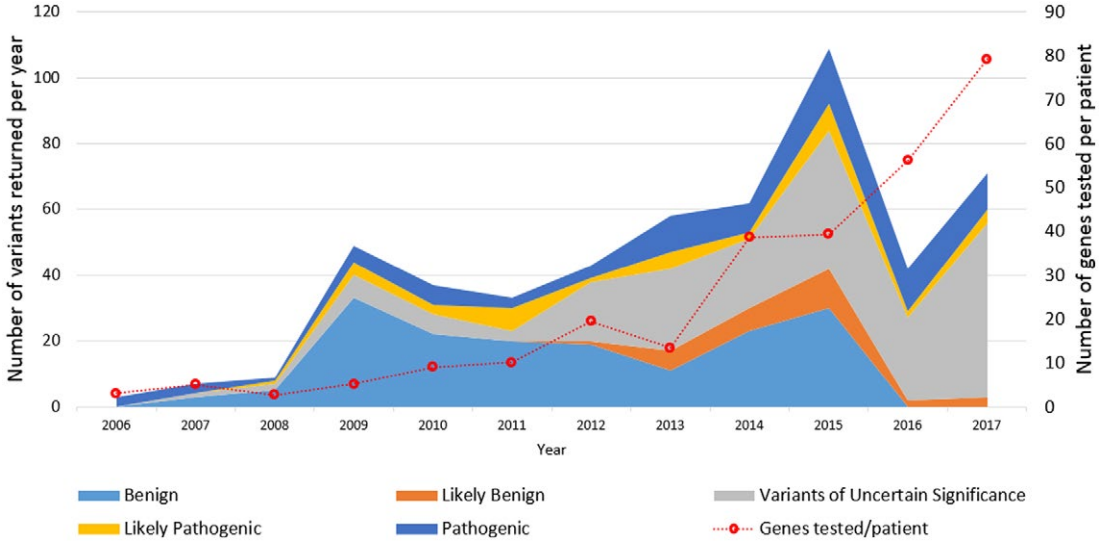

FIGURE 1 Stacked area plot, total number of variants per year. On a separate vertical axis, the average number of genes tested per patient is overlaid with red dots.

Results: The baseline rate of management plan completion was 64%. Following the first PDSA cycle, the average completion rate improved to 94% and a positive shift in the data was observed. This suggests that the observed improvement was nonrandom and likely related to the intervention. Following the second PDSA cycle the average completion rate remained stable at 91%.

Conclusion: Our results showed improvement in management plan completion rates following the initial intervention. While further improvement was not observed, the second intervention may have contributed to sustaining the initial improvement throughout the second PDSA cycle. Future improvement strategies may include optimization of the electronic medical record to automatically prompt inclusion of the essential management plan components.

Factors associated with readmissions in infants following congenital heart surgery

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Purpose: Hospital readmission rates are increasingly utilized as a benchmark for care quality and value. Patients with congenital heart disease are at an increased risk for hospital readmission. To date, there is a paucity of data that specifically explores factors associated with unplanned readmissions following congenital heart surgery. This investigation aims to characterize variables associated with unplanned readmissions for infants who have undergone surgical repair of congenital heart defects in order to identify specific areas ripe for quality improvement and resource stewardship.

Methods: An Institutional Review Board approved retrospective review was conducted on all infants less than 1 year of age who underwent congenital heart surgery within a single hospital system from January 1, 2015 through December 31, 2016 and were readmitted within one year of discharge from the surgical hospitalization. Collected variables included patient characteristics, select resource utilization during the surgical hospitalization, and discharge instructions along with variables specific to the readmission. Descriptive data is presented as percentile for categorical variables and as median with range for continuous data.

Results: Over two years, 157 infants had unplanned readmissions to the surgical center and accounted for 64% of total readmissions after infant cardiac surgery. Nearly half the patients with unplanned readmissions had an extracardiac morbidity, syndrome or genetic anomaly. The mean age at the time of surgery for those who had unplanned readmissions was 34 days and had a median length of stay of 35 (5-288) days. About half the patients were receiving preoperative respiratory support and not receiving oral feeds. At discharge, follow-up appointments were most commonly made to a cardiology provider; less than a third of appointments were with the primary care provider. Most patients with unplanned readmissions were admitted from the emergency room with 36 (23%) patients admitted directely to the intensive care unit. Median hospital length of stay during the readmission was 4 (1-254) days.

TABLE 1

All Readmissions	
Total Patients	212
Total Readmissions	247 (100%)
Total Unplanned Readmissions	157 (64%)
Total Planned Readmissions	90 (36%)
Patient Factors in UR	**n=157**
Extracardiac Anomaly	64 (41%)
Chromosomal Abnormality	88 (56%)
Syndrome	67 (43%)
Age at Initial Cardiac Surgery (days)	34 (0-364)
Surgical Hospitalization Factors in UR	**n = 157**
Length Of Stay	35 (5-288)
Discharge Medications (Daily Doses)	6 (0-36)
Cardiopulmonary Bypass	109 (69%)
Initial Cardiac Surgery Admission	125 (80%)
Preoperative Inotropic Support	20 (13%)
Pre-Op Respiratory Support	74 (47%)
Nasal Cannula	11 (7%)
CPAP/HFNC**	34 (22%)
Mechanical Intubation	29 (18%)
Preoperative Feeding Route	
None/TPN	47 (30%)
Oral	49 (31%)
Nasogastric Tube	15 (10%)
Sugical Gastrostomy Tube	8 (5%)
Combined Oral and Tube	38 (24%)
Discharge Feeding Route	
None/TPN	0 (0%)
Oral	107 (68%)
Nasogastric Tube	5 (3%)
Surgical Gastrostomy Tube	27 (17%)
Combined Oral and Tube	18 (11%)
Follow-Up Appointments	
Cardiology	96 (61%)
Cardiovascular Surgery	50 (32%)
Primary Care Provider	42 (27%)
Other Specialist	66 (42%)
Readmission Hospitalization Factors in UR	**n=157**
Readmission Length of Stay	4 (1-254)
ICU Readmission	36 (23%)
Readmit Origin	
Emergency Department	117 (75%)
Clinic	25 (15%)
Outside Facility	9 (6%)
Home	2 (1%)
In-Hospital Procedure	4 (3%)

**High-Flow Nasal Cannula

Conclusion: Preliminary data suggests that a majority of readmissions within the first year after cardiac surgery are unplanned. The affected patients commonly had noncardiac diseases and required preoperative respiratory and nutritional support. Understanding the reasons for unplanned readmissions are ongoing but opportunities for improvement elicited from these data include transitional care coordination to the primary care provider as well as necessary subspecialists. Further analysis into strategies to prospectively identify infants at higher risk for readmission following cardiac surgery and program development to mitigate the readmission risk is warranted.

Impact of cardiac conditions and other chronic illnesses on "flourishing" in the United States

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Purpose: Children with special health care needs are at increased risk for medical problems and/or developmental challenges. The CDC's 2011/12 National Survey of Children's Health (NSCH) introduced the concept of "flourishing," a derived composite measure. When updated in 2016, the CDC defined "flourishing" in children ≤5 years as "curiosity/discovery about learning, resilience, attachment with parent, and content with life," whereas "flourishing" in children 6-17 years was defined as "curiosity/discovery about learning, resilience, and self-regulation." The objective of this study is to assess, in a large, nationally representative sample, to what extent young children and school-age children with a current diagnosis of a heart condition are less likely to "flourish" compared to "healthy" children, and to assess the impact of disease severity on flourishing for children with heart conditions as compared to children with other chronic medical conditions.

Methods: In the 2016 NSCH, for children ≤5 years, a child was identified as "flourishing" if the parent responded "*Definitely True*" to questions about whether the child "*is affectionate/tender*," "*bounces back quickly when things don't go his/her way*," "*shows interest/curiosity in learning new things*," and "*smiles/laughs a lot*." Children 6-17 years were identified as "flourishing" if the parent responded "*Definitely True*" to questions about whether the child "*shows interest/curiosity in learning new things*," "*works to finish tasks he/she starts*," and "*stays calm and in control when faced with a challenge*." Logistic regression

TABLE 1 Flourishing in children ages 1-5 years in the United States, with a focus on children with a current cardiac or other chronic medical condition (n=12 642)

Characteristics	n	% Flourishing (Weighted)	Adjusted Odds Ratios*	(95% CI)
Age (years)				
0 – 2	5072	68.8	ref	
3 – 5	7546	59.7	**0.64**	0.59 - 0.7
Sex				
Female	6083	65.0	ref	
Male	6535	61.7	**0.78**	0.73 - 0.8
Race/ethnicity				
White Non-Hispanic	8812	69.0	ref	
Black Non-Hispanic	654	55.8	0.87	0.73 - 1.0
Hispanic	1376	55.7	**0.79**	0.70 - 0.9
Other/multiracial, Non-Hispanic	1776	60.3	0.8	0.71 - 0.8
Weight Status (not available for children <10 yrs old)				
Healthy weight	-----	-----	-----	
Underweight	-----	-----	-----	
Overweight	-----	-----	-----	
Obese	-----	-----	-----	
Child Overall Health Status				
Excellent or Very Good	11898	65.4	ref	
Good	618	19.9	**0.38**	0.32 - 0.4
Fair or Poor	75	27.5	**0.27**	0.16 - 0.4
Born Prematurely (<37 weeks gestation)				
No	11247	63.8	ref	
Yes	1257	60.2	0.92	0.81 - 1.0
"Heart Condition" (medical diagnosis)				
None	12417	63.4	ref	
Mild	122	61.7	**0.65**	0.44 - 0.9
Moderate or Severe	39	40.1	**0.47**	0.24 - 0.9
"Diabetes" (medical diagnosis)				
None	12578	63.3	ref	
Mild	2	0	n/a	n/a
Moderate or Severe	6	100	n/a	n/a
"Asthma" (medical diagnosis)				
None	11958	63.8	ref	
Mild	409	62.6	1.00	0.80 - 1.2
Moderate or Severe	186	32.2	**0.61**	0.45 - 0.8
"Arthritis" (medical diagnosis)				
None	12534	63.3	ref	
Mild	6	98.0	0.90	0.16 - 5.2
Moderate or Severe	5	34.8	0.41	0.07 - 2.5

TABLE 2 Flourishing in children ages 1 -5 years in the United States, with a focus on children with a current cardiac or other chronic medical condition (n= 35718)

Characteristics	n	Weighted % Flourishing	Adjusted Odds Ratios*	(95% CI)
Age (years)				
6 – 8	7037	36.8	ref	
9 – 11	7946	40.1	**1.23**	1.15 - 1.3
12 – 14	9185	41.2	**1.43**	1.33 - 1.5
15 – 17	11475	43.4	**1.66**	1.55 - 1.7
Sex				
Female	17452	43.2	ref	
Male	18191	37.7	**0.75**	0.72 - 0.7
Race/ethnicity				
White Non-Hispanic	25126	40.6	ref	
Black Non-Hispanic	2129	36.7	**1.22**	1.11 - 1.3
Hispanic	3921	41.8	**1.17**	1.08 - 1.2
Other/multiracial, Non-Hispanic	4467	40.6	**1.09**	1.02 - 1.1
Weight status (not available for children <10 yrs old)				
Healthy weight	16315	44.9	**ref**	
Underweight	1490	43.7	**0.87**	0.78 -0.97
Overweight	3463	41.3	**0.88**	0.82 -0.95
Obese	3090	31.0	**0.66**	0.61 – 0.72
Child Overall Health Status				
Excellent or Very Good	32386	43.5	ref	
Good	2724	18.4	**0.26**	0.23 – 0.29
Fair or Poor	468	11.6	**0.16**	0.12 – 0.22
Born Prematurely (<37 weeks gestation)				
No	31225	41.1	ref	
Yes	3865	36.8	**0.82**	0.76 - 0.8
"Heart Condition" (medical diagnosis)				
None	35050	40.5	ref	
Mild	357	25.1	**0.62**	0.49 - 0.7
Moderate or Severe	100	28.6	**0.64**	0.42 – 1.0
"Diabetes" (medical diagnosis)				
None	35289	40.5	ref	
Mild	49	21.2	0.62	0.33 - 1.1
Moderate or Severe	169	31.1	0.92	0.67 - 1.2
"Asthma" (medical diagnosis)				
None	31861	40.9	ref	
Mild	2439	37.4	**0.85**	0.78 - 0.9
Moderate or Severe	959	31.7	**0.83**	0.72 - 0.9
"Arthritis" (medical diagnosis)				
None	35239	40.4	ref	
Mild	70	49	0.64	0.38 - 1.0
Moderate or Severe	54	27.2	0.89	0.49 - 1.6

was performed, and analyses were adjusted for child age, sex, race/ethnicity, prematurity, highest level of educational achievement (parent), household structure, and household income. A weighted prevalence of flourishing was calculated for children with heart conditions, diabetes, asthma, and arthritis, with separate analyses for severity (mild vs. moderate/severe). Separate analyses were done for children ≤5 and 6-17 years because of age-specific differences in definition and cardiac concerns.

Results: The weighted prevalences for flourishing are reported in Table 1 (≤5 years) and Table 2 (6-17 years). Compared to children with no current cardiac condition, preschool and school-age children with a mild current cardiac condition were 35% and 38% less likely, respectively, to flourish; children with a moderate-severe cardiac condition were 53% and 36% less likely. Of the children with a moderate-severe cardiac condition, 33.8% currently receive special education services, and 82.3% currently receive developmental/behavioral therapies—higher than children with moderate-severe diabetes, asthma, or arthritis. For children with mild cardiac conditions, the corresponding proportions are 19.2% and 54.9%.

Conclusion: In this large, nationally representative sample of children, preschool children with a mild or moderate-severe current cardiac condition are much less likely to "flourish." As such, all clinicians monitoring a child with a cardiac condition should assess for potential developmental disabilities and direct families to educational and therapeutic resources.

EP ABSTRACT PRESENTATIONS: SUNDAY, NOVEMBER 4, 2018: 11:45 AM-1:00 PM

Evaluation of P-wave dispersion as a predictor of atrial arrhythmias in dystrophinopathies

Krishna Kishore Umapathi[1]; Indiresha Iyer[2]; Aravind Thavamani[3], [1]Metro Health Medical Center, Cleveland, Ohio, [2]Case Western Reserve University, Cleveland, Ohio, [3]Metrohealth Medical Center, Cleveland, Ohio

Purpose: Arrhythmias are one of the manifestations of cardiac involvement in dystrophinopathies. P-wave dispersion (PWD) has been known to be associated with inhomogeneous and discontinuous propagation of sinus impulses. In the present study, we aimed to investigate the role of PWD in patients with Duchenne (DMD) and Becker (BMD) muscular dystrophies.

Methods: It is a retrospective cohort study. The study population consisted of 68 patients with dystrophinopathy and age and sex matched 68 healthy control subjects. Patients with diabetes mellitus, coronary artery disease, congenital heart disease, pulmonary hypertension, hyperthyroidism were excluded from the study. Maximum P-wave duration (Pmax) and minimum P-wave duration (Pmin) were measured from the 12-lead surface electrocardiogram. PWD was calculated as the difference between Pmax and Pmin. The following values were obtained from the transthoracic echocardiogram—left atrial diameter (LAD), left ventricular end diastolic (LVEDD) and end systolic diameters (LVESD), left ventricular ejection fraction (LVEF) which were measured in M mode. EKG and echocardiogram reports were done within 2 months of each other in all the patients. LVEF

Results: Pmax and PWD of patients with dystrophinopathies were significantly higher than those of control subjects (Pmax: 124±10 ms vs. 111±9 ms, $P<.001$; PWD: 48±5 ms vs. 36±6ms, $P<.001$). However, there was no statistically significant difference between patient group and control group regarding Pmin (80±7 ms vs. 79±6 ms, $P=.25$). Left atrial diameter (LAD) was significantly higher in patients with dystrophinopathies compared to control subjects (4.71±0.59 cm vs. 3.57±0.42 cm, $P<.001$). Left ventricular ejection fraction (LVEF) was found to be significantly lower in patients with dystrophinopathies compared to control subjects (43±5% vs. 63±7%, $P<.001$). Similarly, LVEDD and LVESD were significantly lower in dystrophinopathies vs controls ($P<.001$). A significant positive correlation was noted between Pmax and PWD with LAD (r = .576, $P<.001$ and and r = 0.486, $P<.001$ respectively). Similarly, a significant negative correlation was noted between Pmax and PWD with LVEF (r=−0.578, $P<.001$ and r=−0.554, $P<.001$ respectively).

Conclusion: PWD is significantly higher in patients with dystrophinopathies than in healthy control subjects. Prolonged PWD is associated with systolic dysfunction in patients with DMD and BMD. It is known that presence of cardiomyopathy in dystrophinopathies increases the risk of atrial arrhythmias and we suggest that P wave dispersion should be used as a screening tool for early detection of atrial arrhythmias in patients with dystrophinopathies, particularly in those who have been diagnosed with dilated cardiomyopathy.

Palpitations after cryomodification for AVNRT

Gregory Webster MD, MPH[1]; Kendra Ward MD[2]; Sabrina Stanley[2]; Ahmad Chaouki MD, PhD[2]; Sabrina Tsao MBBS[2], [1]Ann and Robert H. Lurie Children's Hospital of Chicago / Northwestern University Feinberg School of Medicine, Chicago, Illinois, [2]Ann and Robert H. Lurie Children's Hospital of Chicago, Chicago, Illinois

Background: AV nodal reentrant tachycardia (AVNRT) is a common substrate for supraventricular tachycardia (SVT) in children and young adults. Some patients will return after ablation with palpitations. Follow-up studies in cardiology have focused on recurrence of documented arrhythmia, but not all patients with palpitations will have had a recurrence of their SVT substrate. For many families, palpitations after ablation create worry and influence patient experience. In addition, they may trigger clinical testing to determine if there has been recurrence of SVT. Therefore, our objective was to quantify palpitations after AVNRT cryoablation and compare the frequency of palpitations to the frequency of documented SVT recurrence.

Methods: We performed a retrospective review of all patients with a structurally normal heart who underwent cryoablation for AVNRT in our pediatric center (3 invasive pediatric electrophysiologists). Follow-up clinic notes were reviewed as well as the detailed logs of telephone calls from patients after ablation. All tabulated palpitations occurred at least 31 days after ablation. We excluded complaints of palpitations in the first 30 days after ablation since these are very common and are discussed in our discharge paperwork.

Results: We reviewed 100 consecutive cases from 2012-2017 (mean age 14 years, standard deviation 4.2; mean weight 59.6 kg, standard deviation 22.2). Acute success was achieved in all cases; radiofrequency was used in 4 cases due to an incomplete end point with cryoablation. Mean follow-up was 14.4 months (standard deviation 12.8). Postprocedural palpitations or arrhythmia complaints were present in 40/100 patients. Clinical reassurance, without ambulatory monitor, was sufficient in 15/40 symptomatic patients (38%). Monitors were used in 25/40 patients (Holters in 20/40; event monitors in 19/40; more than 1 monitor in 15/40). Documented SVT after ablation occurred in 3 pts (8% of patients with symptoms). At subsequent intracardiac study, only 2/3 recurrences were AVNRT. One recurrence was a new finding of atrial tachycardia. Postprocedural supraventricular tachycardia was captured by Holter in 1 patient and by event monitor in 2 patients (both on the patient's second month of event monitoring).

Conclusions: Pediatricians and cardiologists should anticipate postprocedural palpitations in 40% of patients who undergo cryomodification for AVNRT. Fortunately, despite a high rate of palpitations, recurrence rates are very low (2%). Patients should be counseled prior to ablation about the rate of postprocedure symptoms, but reassured about the low rate of true recurrence.

Potential recovery and long term follow up of heart block following surgical repair of congenital heart disease

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Purpose: Pacemaker implantation in pediatric patients is commonly needed after surgical heart block as well as postoperative high grade sinus node dysfunction. The purpose of our study was to examine in detail the risk factors for heart block prior to and following cardiac surgery. Additionally we aimed to study the evolution of pacing needs, recovery of conduction and pacemaker properties over time as there is extremely limited data regarding the same.

Methods: We performed a retrospective cohort study by querying the pediatric cardiac surgical database at our institution for all patients between January 2012-January 2017 who had undergone surgical placement of pacemaker following open heart surgery. Patients with pacemakers placed due to congenital heart block or implanted by catheterization were excluded. Patient data, including demographic, primary surgical, and pacemaker information and interrogations, was reviewed.

TABLE 1 Patient demographics

Age (median and range)	1562 days (8-6796)	
Weight (median and range)	13.6 kg (2.4-82.7)	
Days between primary surgery and pacemaker placement (median and range)	18.5 days (4-4117)	
Primary surgery (number and percent of total)	Fontan	17 (27.9%)
	Valve replacement or repair	9 (14.8%)
	AVSD repair	8 (13.1%)
	VSD Repair	6 (9.8%)
	DORV repair	5 (8.2%)
	Norwood	3 (4.9%)
	Glenn	2 (3.3%)
	Aortic arch repair	2 (3.3%)
	Modified BT shunt	2 (3.3%)
	Other	7 (11.5%)
ICU time (mean and range)	101.0 hours (0-550)	
Ventilator time (mean and range)	179.4 hours (0-8786.7)	
Hospital LOS (mean and range)	14.1 days (1-79)	
30-day mortality	0%	

TABLE 3 Pacemaker analyses at measured intervals

	Initial Analysis	1 month	3 months	6 months	Last Analysis
Min. heart rate (bpm)	90.7 (50-120)	85.1 (50-120)	83.6 (50-150)	77.0 (40-120)	69.7 (40-100)
Max. heart rate (bpm)	176.4 (100-210)	170.3 (100-210)	173.6 (105-210)	169.4 (95-210)	164.9 (105-210)
RA amplitude (V)	3.5 (2.5-5)	3.3 (1.5-7.5)	2.99 (1.5-7.5)	2.3 (1.5-3.5)	2.3 (0.5-7.5)
RV amplitude (V)	3.6 (2-5)	3.3 (2-5)	2.9 (2-5)	2.6 (2-4)	2.8 (2-5.8)
LV amplitude (V)	3.4 (2.75-4)	2.8 (2.5-3)	2.2 (1.25-2.75)	2.6 (2.5-2.75)	1.9 (1.5-2.25)
RA sensitivity (mV)	0.55 (0.15-1)	0.71 (0.18-2.8)	0.71 (0.18-4)	0.74 (0.25-4)	0.78 (0.18-2.8)
RV sensitivity (mV)	3.4 (0.9-5.6)	3.3 (0.5-5.6)	3.4 (0.6-5.6)	3.0 (0.6-5.6)	2.9 (0.6-5.6)
AS/VS (percent)	8.8 (0-100)	13.9 (0-100)	16.0 (0-100)	17.3 (0-100)	14.7 (0-100)
AS/VP (percent)	51.6 (0-100)	56.5 (0-100)	54.0 (0-100)	53.2 (0-100)	58.5 (0-100)
AP/VS (percent)	22.5 (0-100)	21.8 (0-100)	20.4 (0-93.69)	20.7 (0-97.64)	18.5 (0-97.8)
AP/VP (percent)	14.3 (0-97.3)	7.7 (0-100)	10.1 (0-88.97)	8.7 (0-63)	8.2 (0-98.52)
RA Impedance (Ohms)	602.9 (48-2445)	555.8 (199-1338)	802.1 (224-15148)	487.7 (110-721)	1835.0 (67-1200)
RV Impedance (Ohms)	761.8 (67-1862)	597.2 (67-1261)	560.8 (67-836)	538.6 (202-772)	553.8 (0-848)
LV Impedance (Ohms)	563.7 (342-855)	481.3 (247-703)	696.7 (494-874)	551 (475-627)	446.5 (323-627)

Results: A total of 61 patients were included in the study. Patient characteristics, primary cardiac surgery, and postoperative course prior to pacemaker placement are shown in Table 1. In general, we wait for 2 weeks prior to permanent pacemaker placement to allow for early recover to occur as shown by our median wait time of 18.5 days. Thereafter, pacing mode discharge was AAI in 12 patients (20%) while the rest were DDD or DDDR (80%). Serial data from the pacemaker interrogations is shown in Table 2. It is noteworthy that only 6/61 patients (~10%) showed less than 50% need for pacing at the latest follow up while 90% continued to be pacemaker dependent for greater than 50% of the time. There was no change in any of the patient pacemaker parameters such as heart rate variation, atrial amplitude, ventricular amplitude, atrial sensitivity and ventricular sensitivity. However, there was a significant increase in the atrial lead impedance from 603 ohms to 1835 ohms over the follow up period.

Conclusions: Our center's approach of slightly delayed implantation of pacemakers for surgical heart block or sinus node dysfunction appears to minimize unnecessary pacemaker placement with adequate time for early recovery. Thereafter, there is minimal evidence of conduction recovery (<10%). Pacemaker performance was reliable except significant increase in atrial lead impedance over a follow up period. Further analysis to understand risk factors for recovery in this small group of patients as well as analysis within individual indication categories is being performed to provide further clarity.

Use of surface electrocardiograms in evaluating risk of cardiomyopathy in children with anthracycline exposure

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Purpose: Children exposed to anthracyclines are at risk for cardiomyopathy. Early recognition of cardiomyopathy may reduce morbidity and mortality, but substantial myocardial damage can occur before echocardiographic changes are apparent. We hypothesized that electrocardiogram (ECG) changes precede echocardiographic changes and may be used as an early risk stratification screening tool for cardiomyopathy.

Methods: We performed a retrospective analysis of cancer patients who received anthracyclines at a tertiary pediatric referral center. ECG end points were the sum of absolute QRS amplitudes in the 6 limb leads (ΣQRS amplitude) and the corrected QT (QTc) interval using Bazett's formula. Cardiomyopathy was defined by echocardiogram as ejection fraction <50%, shortening fraction <26%, or left ventricular end-diastolic diameter *z*-score >2.5. Cox regression models with time-dependent covariates explored changes in ΣQRS amplitude and QTc interval prior to development of cardiomyopathy. If the patient did not develop cardiomyopathy, the last available echocardiogram was considered the censored time. We evaluated unit changes of 0.6 mV in ΣQRS amplitude and 10 ms in QTc interval, because changes smaller than these are difficult to measure with calipers in practice and are less clinically relevant. Effects of anthracycline dose and cardiotoxic radiation on ECG measures were assessed with multiple linear regression models.

Results: Among 589 patients analyzed, 19.5% met criteria for cardiomyopathy. Male sex, black race, older age at first dose, and larger body surface area (BSA) were associated with development of cardiomyopathy. Cardiomyopathy was associated with increased mortality (34% versus 12%, $P<.01$). Our most important finding was that a 0.6 mV decrease in ΣQRS amplitude had a hazard ratio of 1.174 (95%CI=1.057-1.304) translating to a 17% increased risk of developing cardiomyopathy ($P<.01$), and a 10 ms increase in QTc interval had a hazard ratio of 1.098 (95%CI=1.027-1.173) translating to a 10% increased risk of developing cardiomyopathy ($P<.01$). Total anthracycline dose predicted a decrease in ΣQRS amplitude and an increase in QTc interval independent of cardiomyopathy status after controlling for age, sex, BSA, pre-therapy ECG measurements, and time between ECG measurements ($P<.01$). Consistent with our primary analysis, there was a greater decrease in ΣQRS amplitude in the cardiomyopathy group after completion of therapy than in the no cardiomyopathy group (-0.6 mV vs. -0.2 mV, $P=.02$). Cardiotoxic radiation did not predict changes in ECG parameters. Mean age in the study population was 8.8 years ± 5.6; mean follow-up time was 3.8 years ± 2.9.

Conclusion: In patients receiving anthracyclines, a decrease in ΣQRS amplitude and prolongation of the QTc interval precede development of cardiomyopathy. ECG is a potential noninvasive tool for prediction of anthracycline-induced cardiomyopathy but requires prospective validation.

ABSTRACT POSTER PRESENTATIONS

A prediction model for femoral artery and vein dimensions in pediatric patients undergoing cardiac catheterization

William McEachern MD, MPH; Jonathan Soslow; Tom Doyle; Dana Janssen; George Nicholson, Monroe Carell Jr. Children's Hospital at Vanderbilt, Nashville, Tennessee

Background and Objectives: Percutaneous intervention is increasingly favored in pediatric patients in the appropriate clinical setting. However, the size of femoral vessels can limit the ability to perform an intervention. There has been no standardization of femoral artery (FA) and vein (FV) dimensions in pediatric patients in the United States; thus, our study set out to create predictive models for both FA and FV dimensions.

Methods: We retrospectively reviewed caths at our center between July 1, 2016 and June 30, 2017. Patients with 2D ultrasound images that were inadequate for measurement of vessel diameters were

excluded. A cohort of 143 pediatric patients with no history of prior cath (cohort initial) and 50 patients undergoing subsequent caths (cohort prior) were selected. Univariate linear regression evaluated the association between predictors (age, height, weight, body surface area, single ventricle physiology status, and history of previous central venous catheter (CVC)) and outcome measures (FV and FA diameter, FV and FA cross-sectional area [CSA]). Multivariable regression models were created; variables that did not improve the r^2 appreciably were removed, with a pre\specified goal of optimizing model performance and simplicity.

Results: Table 1 provides basic demographics. Patients undergoing an initial cath (cohort initial) were younger and less likely to have single ventricle physiology than those undergoing subsequent cath (cohort prior). All variables correlated with outcome measures on univariate analysis. Height was the most relevant determinant for all outcome measures. In cohort initial, height and weight were the optimal predictors of FA and FV diameter and CSA. In cohort prior, age added incremental value and was included in the final models. Our predictive models are as follows:

FV diameter:

Cohort Initial: $d = 0.752 + 0.023(\text{weight}) + 0.044(\text{height})$, $r^2 = 0.81$

Cohort Prior: $d = 1.349 - 0.045(\text{weight}) + 0.043(\text{height}) + 0.165(\text{age})$, $r^2 = 0.63$

FV CSA:

Cohort Initial: $\text{CSA} = -5.712 + 0.588(\text{weight}) + 0.218(\text{height})$, $r^2 = 0.79$

Cohort Prior: $\text{CSA} = -1.555 - 0.451(\text{weight}) + 0.238(\text{height}) + 2.78(\text{age})$, $r^2 = 0.54$

FA diameter:

Cohort Initial: $d = 0.001 + 0.009(\text{weight}) + 0.043(\text{height})$, $r^2 = 0.84$

Cohort Prior: $d = -0.883 - 0.013(\text{weight}) + 0.058(\text{height}) - 0.101(\text{age})$, $r^2 = 0.65$

FA CSA:

Cohort Initial: $\text{CSA} = -9.237 + 0.256(\text{weight}) + 0.225(\text{height})$, $r^2 = 0.81$

Cohort Prior: $\text{CSA} = -18.393 - 0.065(\text{weight}) + 0.384(\text{height}) - 0.599(\text{age})$, $r^2 = 0.59$

Weight (kg), Height (cm), Age (years)

Table 2 presents predicted FV and FA diameters in those undergoing initial catheterization.

Conclusions: Our study presents a predictive model for FV and FA diameter and CSA in pediatric patients undergoing cardiac catheterization. Future research will seek to improve the predictive model for those undergoing subsequent catheterization and examine complication rates in the context of sheath size and the measured vessel dimensions.

A revamped cardiology curriculum for pediatric residents

Christina Ronai MD[1]; David Brown MD[2]; Roger Breitbart MD[2], [1]OHSU, Portland, Oregon, [2]Boston Children's Hospital, Boston, Massachusetts

Purpose: The American Board of Pediatrics (ABP) has issued guidelines for pediatric residents on pediatric cardiology (PC) topics. At many academic institutions, the teaching on the inpatient subspecialty PC service is very dependent upon the patient volume and mix, and residents may not have exposure to the ABP topics over the course of their PC rotations. We sought to assess pediatric residents' comfort with ABP cardiology topics, and subsequently developed and implemented a structured PC curriculum.

Methods: A needs-based assessment was performed via electronic survey sent to current pediatric residents (n = 102) in one program who had completed their PC rotation and also to recent graduates of that program (n = 62). The anonymous survey instrument was informed by the ABP recommended content for cardiology for board certification in Pediatrics, and included questions about comfort

TABLE 1 Demographic overview of sampled data for predictive models

	Initial Cath.	Subsequent Cath.	p-value[2]
Age (years)	1.1, IQR (0.35, 5.8)	6.8, IQR (3.0-14.3)	< 0.0001
Percent Male (%)	43%	58%	0.182
Weight (kg)	8.4, IQR (5.0-18.4)	18.9, IQR (11.9-47.3)	0.0003
Height (cm)	72, IQR (55.0-112.5)	113, IQR (91.0-153.3)	< 0.0001
Single Ventricle (%)	15%	46%	0.0001

[1]Where applicable, the median is listed with the interquartile range.
[2]This column lists the p-values for two-tailed t-tests of non-different means between the two samples.

TABLE 2 Predicted FV and FA diameters for different combinations of height & weight in those undergoing initial catheterization[1]

	Height (cm)					
Weight (kg)	*50*	*65*	*80*	*95*	*110*	*125*
5	3.1, 2.2	3.7, 2.8	4.4, 3.5	5.1, 4.1	5.7, 4.8	6.4, 5.4
10	3.2, 2.2	3.8, 2.9	4.5, 3.5	5.2, 4.2	5.8, 4.8	6.5, 5.5
15	3.3, 2.3	4.0, 2.9	4.6, 3.6	5.3, 4.2	5.9, 4.9	6.6, 5.5
20	3.4, 2.3	4.1, 3.0	4.7, 3.6	5.4, 4.3	6.1, 4.9	6.7, 5.6
25	3.5, 2.4	4.2, 3.0	4.8, 3.7	5.5, 4.3	6.2, 5.0	6.8, 5.6
30	3.6, 2.4	4.3, 3.1	5.0, 3.7	5.6, 4.4	6.3, 5.0	6.9, 5.6

[1]Prediction of FV diameter is followed by prediction of FA diameter (both in mm).

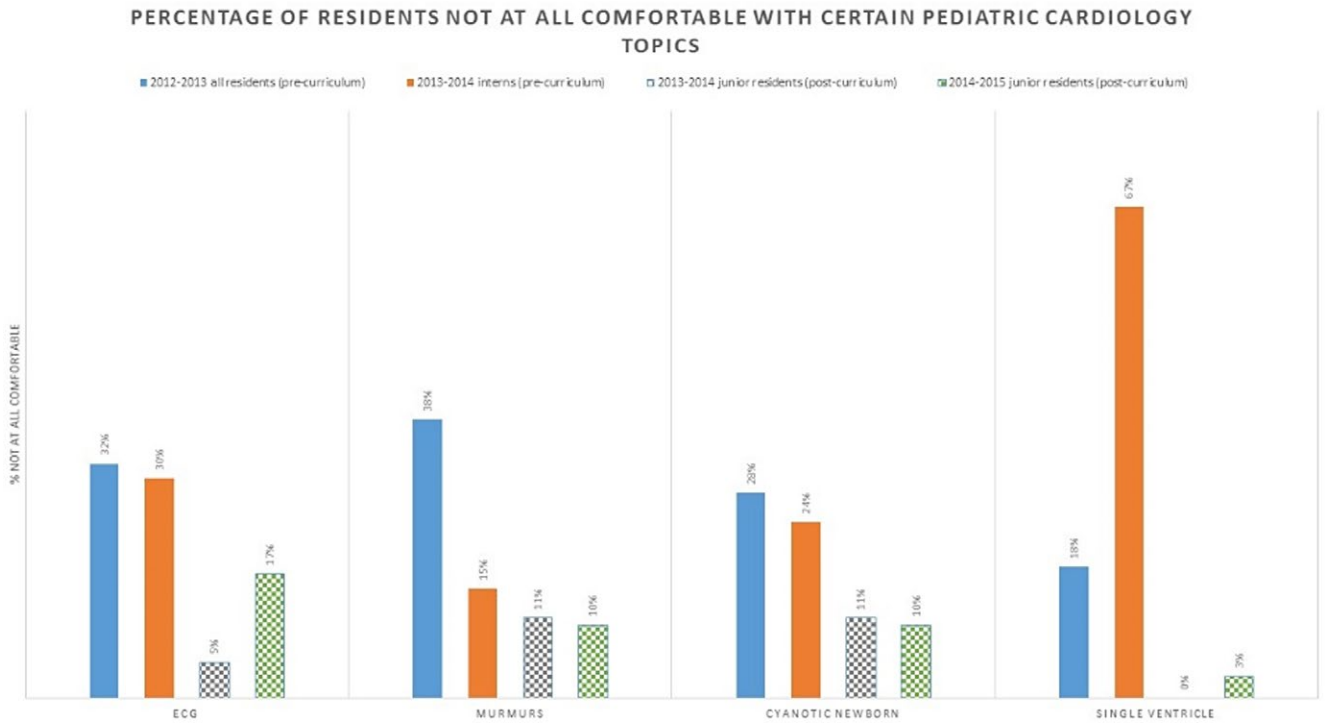

FIGURE 1

with murmur assessment, single ventricle physiology, normal vs abnormal ECG, signs and symptoms of heart failure and the differential diagnosis of cyanosis. The responses were used to develop case-based lectures and formal didactics for the cardiology lectures with the ultimate goal of providing greater comfort among pediatric residents with basic cardiology topics. Residents were re-surveyed annually for two years after the implementation of the new curriculum.

Results: The initial response rate from current residents was 71% (72 out of 102 emailed surveys), and 63% from recent graduates (39 out of 62). Initial needs assessment revealed that residents (both current and former) wanted more generally applicable pediatric cardiology and exposure to cardiology that was relevant beyond their inpatient cardiology experience. Seventy-six percent of residents were less than very comfortable with the differential diagnosis of cyanosis in a newborn; 92% of residents were less than very comfortable with murmur identification after their PC rotation (precurriculum implementation). Four specific case-based didactics were developed and incorporated into the PC rotation: single ventricle physiology in the emergency department, a two-month old with anomalous left coronary artery from the pulmonary artery, a cyanotic newborn and ECG cases. The response rate for the 2013-2014 interns precurriculum was 87% (33 out of 38 interns) and 81% once they were junior residents postcurriculum (same cohort of trainees). Pre- and postcurriculum implementation survey results are shown in Figure 1.

Conclusions: The structured, case-based curriculum assured coverage of the ABP recommended PC topics independent of patient mix and volume during the resident rotation. Furthermore, the new curriculum improved resident comfort with PC subjects. Continuation of this curriculum and ongoing assessment of its efficacy is warranted.

Activity recommendations in children after cardiac surgery

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Purpose: Clinicians, parents, and patients must decide which sorts of physical activity (recreational activities or sports participation) are most beneficial and safe for children who have had cardiac surgery and when these children should return to normal activities after cardiac surgery. Current recommendations are limited and tailored to the adolescent and young adult population.

Methods: We invited members of the Midwest Pediatric Cardiology Society to participate in an online survey of their current practice regarding physical activity recommendations in both the immediate

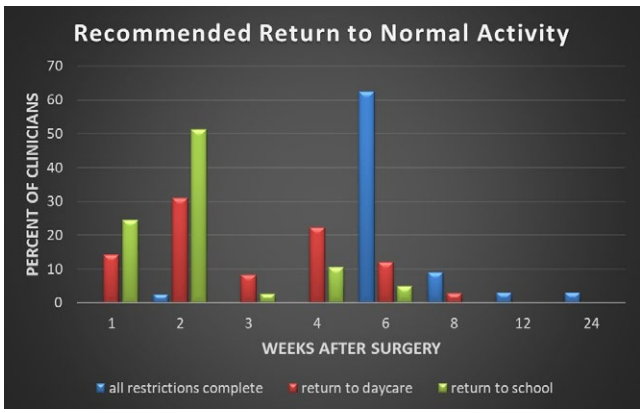

FIGURE 1 Length of postsurgical activity restrictions recommended by clinicians, in weeks

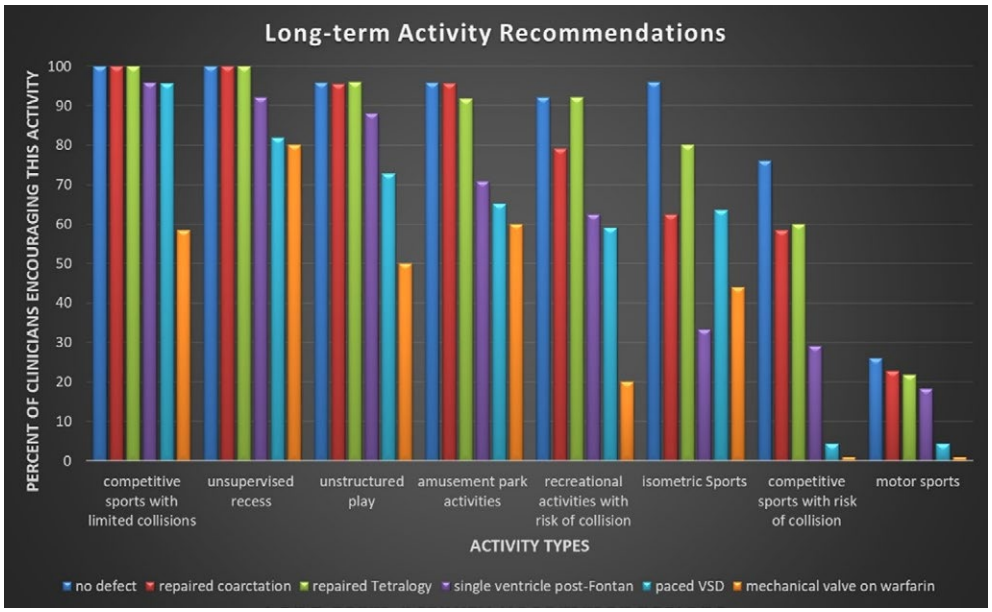

FIGURE 2 Provider recommendations for physical activity in otherwise healthy, 8-year-old children with either no cardiac defect or surgically repaired cardiac defect

postsurgical period and over the long term in patients 12 years of age and younger. Respondents were asked which resources they used in developing activity recommendations, when they recommended a return to normal activities after cardiac surgery, and which sorts of physical activity they would recommend for asymptomatic 8-year-old children with varied cardiac histories (no known cardiac defect, repaired coarctation with normal blood pressure, tetralogy of Fallot with good hemodynamic repair, single ventricle after Fontan, repaired VSD with implanted pacemaker, and mechanical valve anticoagulated with warfarin). Multiple choice, Likert, free-response, and vignette-style questions were used.

Results: Forty-six individuals completed the survey for a response rate of 9.6%. Most were male (69%), physicians (88%) practicing noninterventional cardiology (82%) with an average of 13 years in practice. While there were some areas of consensus in terms of long-term physical activity recommendations, recommended timing of return to normal activity following cardiac surgery was quite variable (see Figure 1). Recommendations regarding long-term physical activity were also variable. While there was 100% agreement that children on anticoagulation should not participate in motorsports or sports with high risk of collision (such as football, hockey, and wrestling); 42% of respondents indicated they would also discourage participation in sports with a lower risk of collision, such as soccer, basketball, tennis or softball. Variability in provider recommendations persisted across patient populations (see Figure 2). Respondents indicated that they used patient testing (58%), symptoms (95%), and hemodynamics (100%); and weighed published guidelines (76%), institutional policies (67%), and the opinions of colleagues or experts in the field (73%) when formulating these recommendations.

Conclusions: Exercise and activity recommendations in the immediate postsurgical period and over the long term after pediatric cardiac surgery in children under the age of 12 years are quite variable, reflecting the absence of evidence-based guidelines for the management of these patients. Future studies should explore the basis of this variability and work towards a clearer understanding of the risks and benefits of various activity types and the timing of return to full activity in children who have had cardiac surgery.

Adverse cardiovascular risk profile and clustering in a pediatric underserved population

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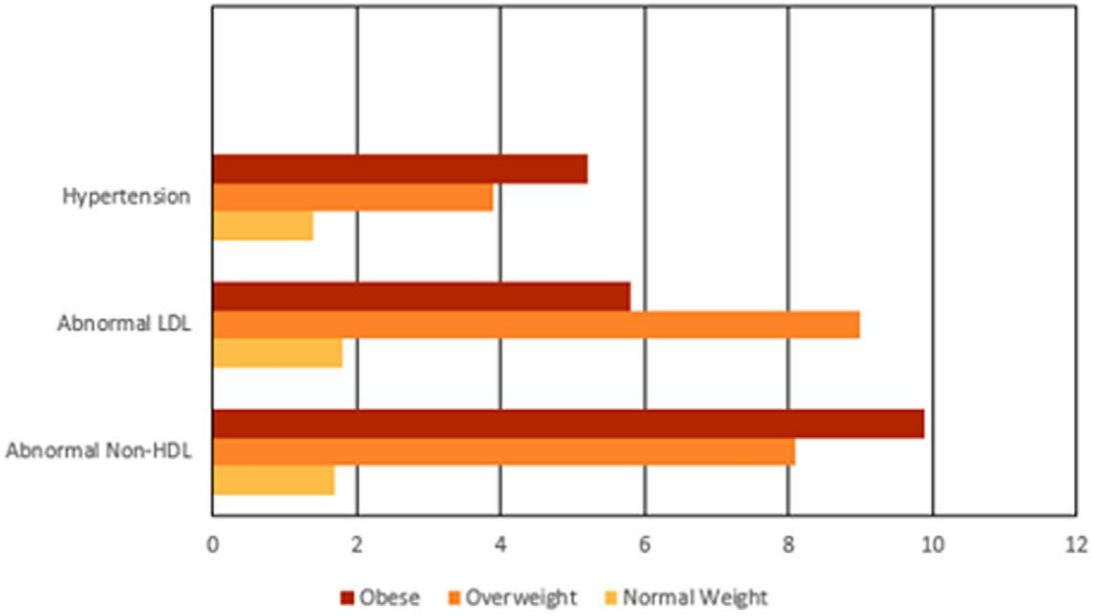
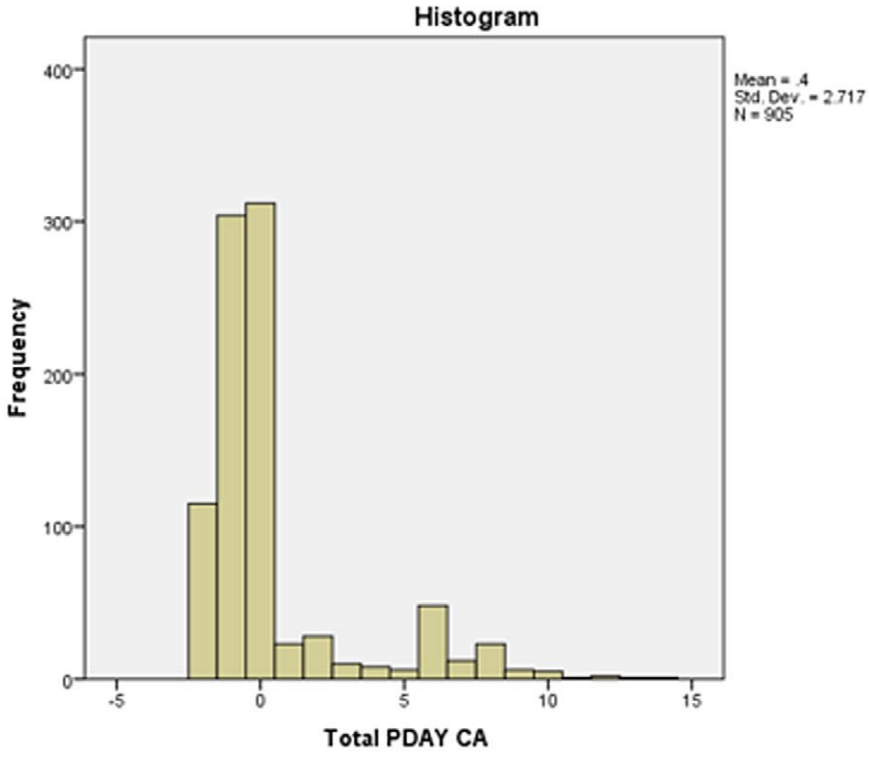
Introduction: Prevalence of adverse cardiovascular risk profile in pediatric underserved population is not well known. In a report from the NHANES database, approximately 36-38% of children had an abnormal BMI. The prevalence of high blood pressure was 3.1% among 8-17 year olds, while 20.2% of 8-17 year olds had abnormal lipid profile. The pathobiological determinants of risk factors in youth (PDAY) score integrates various risk factors for atherosclerosis into a composite score that provides a strong

Prevalence of risk factors

	Available data N	Abnormal N (%)
Obese	905	220 (22.3)
Overweight	905	143(15.8)
HTN	905	25 (2.7)
Elevated blood pressure	905	93 (10.3)
TC	668	44 (6.6)
TG	668	73 (10.9)
NON HDL	668	31 (4.6)
LDL	668	28 (4.2)



Comparison of proportion of risk factors across normal weight, overweight and obese groups



prediction of coronary and abdominal aortic calcification in middle age. The distribution of this score in pediatric underserved population is not known.

Harlem Hospital in New York City almost wholly serving African, African American and Hispanic ethnicities with high rates of childhood obesity and low public awareness. We aim to assess prevalence of adverse cardiovascular risk profile and their clustering in underserved children followed at Harlem hospital.

Methods: This is a retrospective chart review. We chose a random sample of 1000 children and adolescents in the age-group of

9-21 yrs, who follow up at Harlem hospital with a documented visit between Jan 2014 to Dec 2016. We reviewed the charts for height, weight, blood pressure, HbA1C at last visit and any documentation of lipid profile. Data was analyzed for prevalence of abnormal blood pressure, BMI, HbA1C and lipid profile, as well as association between the risk factors. PDAY scores were also computed.

Results: Number of available data in our tested group and the prevalence of abnormality are in Table 1. There is significant correlation between BMI score and systolic BP *z*-score ($r = 0.33$, $P < .001$), total cholesterol ($r = 0.11$, $P < .005$), Triglyceride ($r = 0.22$, $P < .001$), LDL ($r = 0.15$, $P < .001$), HDL ($r = -0.24$, $P < .001$) and non-HDL cholesterol ($r = 0.22$, $P < .001$). Distribution of PDAY score and component of it is in Figure 1. PDAY scores for CAC ranged between −2 to 14, with approximately 53.7% of our population having a PDAY score of 0 or above 12.5% had a PDAY score of 2 or above.

Conclusion: Cardiovascular risk factors are highly prevalent in underserved children. Calculated PDAY scores indicates significant clustering of risk factors. High incidence and clustering of risk factor in this at risk population warrants further prospective studies to assess best method of intervention and resource allocation.

An assessment of pediatric heart transplant admission characteristics and practices using an administrative database

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Background: Changes in orthotopic heart transplantation (OHT) practices over time have been difficult to assess due to reliance on voluntary registry data with limited patient granularity. Here, we use the Pediatric Health Information System database (PHIS) to study the clinical management of OHT recipients during the initial transplant admission.

Methods: A retrospective cohort studywas performedof all OHT recipients <19 years old in PHIS from 2003 to 2017. Using administrative data collected from the initial transplant admission, we determined length of stay, duration of ICU care, other procedures performed, mechanical support used, as well as induction and maintenance immunosuppression protocols. Incidence of in-hospital organ rejection and mortality were calculated. Comparisons were made across three time periods (early: 2003-2007; intermediate: 2008-2012; current: 2013-2017).

Results: From 2003 to 2017, 3214 heart transplant admissions were identified in the PHIS database (early: 742 (23%); intermediate: 1151 (36%); and current: 1321 (41%)). The median total length of stay (LOS) was 52 days (IQR 23-101 days) with a median posttransplant LOS of 19 days (IQR 12-33 days). Prior to transplant, 74% of patients required inotropic support, 10% were on ECMO, and 14.7% had a ventricular assist device (VAD). Posttransplant, 96% of patients required inotropic support, decreasing to 40% by 7 days. ECMO and VAD use after transplant were low at 4.0% and 1.1% respectively. Predischarge catheterization was performed in 61% of patients. Immunosuppression regimens for induction and maintenance have changed substantially over time. In the early era, 25% and 5.7% of patients received anti-thymocyte globulin (ATG) and basiliximab, respectively, for induction, compared to 70% and 14% of patients receiving ATG and basiliximab, respectively, in the current era. In the early era, 42% and 52% of patients were discharged on tacrolimus and mycophenolate, respectively. In the current era, 83% of patients were discharged on both tacrolimus and mycophenolate. Comparing the early and current cohorts, cyclophosphamide use has decreased (33 to 8.7%), as has azathioprine (13 to 4.2%) and steroid (69 to 62%).

Incidence of early rejection declined significantly from 28% in the early era to 17% in the current period ($P < .001$). Overall in-hospital mortality was 5.7% (n = 183 patients), decreasing from 7.8% in the early era to 5.0% in the current era ($P = .017$).

Conclusion: This study is the first to leverage administrative data to describe the clinical management of OHT recipients during their initial transplant admission. Significant changes in induction and maintenance immunosuppression have occurred, with associated lower rates of in-hospital rejection and mortality. Remaining areas for improvement include reducing post-OHT length of stay, inotrope support posttransplant, and steroid use at discharge.

Analysis of maternal characteristics of patients with fetal hypoplastic left heart syndrome

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Background: Nicotine and SSRIs have been associated with neonatal withdrawal syndromes. These syndromes may be associated with a poor response to neonatal treatment of hypoplastic left heart syndrome (HLHS).

Objective: To assess the maternal characteristics of patients with fetal HLHS and analyze for association of these characteristics with response to treatment.

Methods: This is a 10 year retrospective review of all patients diagnosed with fetal HLHS at a tertiary referral center. Maternal characteristics were abstract from the obstetrical history and correlated with the fetal echo findings and responses to treatment after birth from the newborn records. Logistic regression was used to analyze the data and a *P* of >.05 was considered statistically significant.

Results: Eighty-two patients were identified with HLHS. Eighteen chose termination of pregnancy and 29 patients were lost to follow up, leaving 35 patients for analysis. Twelve patients had aortic stenosis with mitral stenosis, 12 had aortic atresia with mitral atresia

and 11 had aortic atresia with mitral stenosis. Fifteen (43%) patients smoked tobacco, 5 (14%) reported SSRI use and 13 (32%) used some other type of prescription medication. Two patients used tobacco and were taking SSRIs. Using logistic regression there was a statistical association of tobacco and/or SSRI use with death prior to a Glenn repair (P = .01). This statistical association was stronger for the 2 patients who abused tobacco and were on a SSRI (P = .00). The specificity and negative predictive value were 100% and 85%, respectively.

Conclusion: Although the exposure numbers are small, this analysis suggests that exposure to tobacco and/or SSRIs may be associated with poor response to neonatal treatment for HLHS. This could be related to a neonatal withdrawal syndrome at a critical time in neonatal treatment for HLHS. Larger studies are suggested to further evaluate this risk.

Anatomic approach does not influence technical outcomes of pericardiocentesis in the pediatric population

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Background: The standard approach to pericardiocentesis is a subcostal approach with echocardiographic guidance. Hepatomegaly and noncircumferential effusions warrant nonstandard approaches to drain effusions; echocardiographic guidance has made other, less traditional, nonsubcostal approaches feasible.

Methods: All pediatric patients undergoing pericardiocentesis from August 2008 to June 2016 at a tertiary care hospital were included. Procedure-related complications, approach to procedure, the location of effusion, history of hematopoietic stem cell transplantation, the presence of echocardiographic or clinical tamponade, and the use of pericardial drain were analyzed.

Results: A total of 60 patients underwent pericardiocentesis with echocardiographic guidance in the cardiac catheterization laboratory. History of hematopoietic stem cell transplantation was the most common underlying diagnosis (n = 31, 51.7%). Anatomic approach was determined by location of effusion. A nonsubcostal approach was used in 70% (n = 43) patients. Left axillary approach was the most common approach (47%, Figure 1). The fifth intercostal space was the most commonly used intercostal space (n = 16, 26.7%). A pericardial drain was left in place in 40 patients (66.7%). In those with a pericardial drain placed, the mean during of drainage was 3.38 days (SD 2.03) with no infectious complications noted. Nine (15%) patients experienced recurrence requiring repeat pericardiocentesis. There were 3 minor complications (transient chamber entry, pneumopericardium and minor bleeding) and 2 major complications (sustained arrhythmia and major vessel injury needing surgery). The underlying diagnosis, anatomic approach to pericardiocentesis, location or size of effusion did not influence the risk of complications. The complication rate was higher in those patients who did not receive a pericardial drain (P < .006).

Conclusions: Echocardiography-guided percutaneous pericardiocentesis regardless of approach is a safe and effective. The use of nontraditional, nonsubxiphoid approach did not significantly affect the rate of complications. The rate of any complication was higher among those who did not receive a pericardial drain.

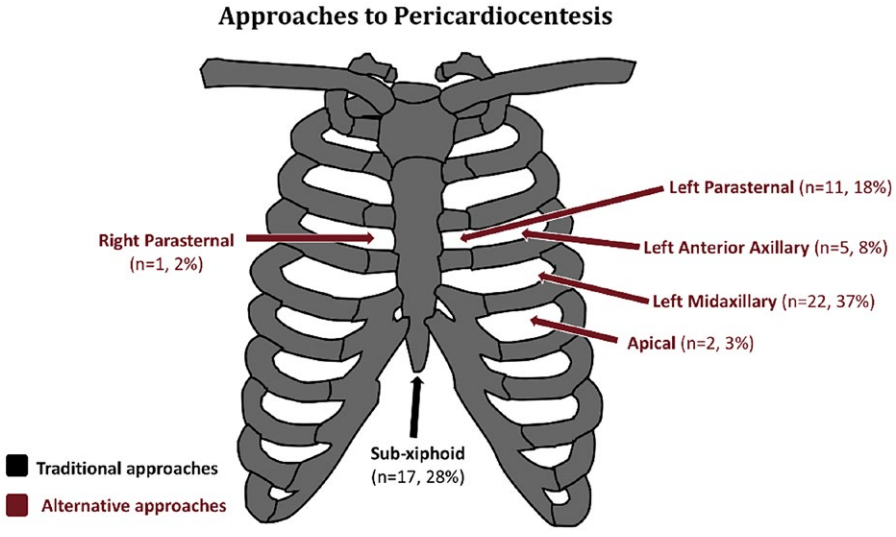

FIGURE 1 Anatomic approach to pericardiocentesis

Assessing hunger vital signs in a pediatric cardiology clinic

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Background and Specific Aims: The American Academy of Pediatrics (AAP) notes that food insecurity (FI) at any level increases a child's risk of illness, hospitalizations, behavioral problems, and suffering in school. Growth and development is a major concern in pediatric patients and is a specific concern for pediatric cardiology patients. 1.2 million people living in Allegheny County are food insecure. Based on national data, roughly 42 000 of these individuals are children.[1,2] The original Hunger Vital Signs (HVS) asks families two validated questions to assess their access to food. Providers are encouraged to ask HVS and intervene, as appropriate.

The aims of this project are to educate the cardiology outpatient team on FI, implement the HVS screening tool in our cardiology office at Children's Hospital of Pittsburgh, assess the clinical success in asking these questions during medical intake, evaluate FI in our clinic patients, act on these results by connecting families to community resources.

Methods: We conducted two plan-do-study-act (PDSA) cycles aimed at implementing HVS in our cardiology outpatient office to assess FI. The clinical team was trained on the HVS. Referral processes and resources were collected for positive screens with multidisciplinary teams including social work. The screening tool was implemented and tracked via electronic medical record. Our social work team provided additional follow up. Screening methods were adjusted following the first round of screening to include a scripted option to address families. Follow-up resources were adjusted based on family needs. The QI team used a run chart to assess QI status (Figure 1), to monitor progress toward improvement.

Results: Interim results revealed that our clinic increased screening to 56% from baseline of zero. Staff meetings revealed hindrances to screening included time, assumed parental concern for getting in trouble, staff turnover, and difficulty remembering to incorporate the results in the medical record. A total of 3% of patients screened positive for FI during the initial study period.

Discussion/Conclusions: The HVS can be effectively implemented in a busy cardiology office and can improve identification of food insecure families. Successful screening can be improved using standard QI methodology. Understanding FI can aid in individualized care and access to resources. Providing individualized training on HVS for screening intake may improve the success of discovering positive results (Klein, et al). It is likely that our study underestimates our families' need for resources as it was conducted during a known period of the year with low seasonal variability. We plan to expand this project to outreach clinics and inpatient service.

Assessment and analysis of obesity in children and young adults with congenital heart disease

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Background: The obesity epidemic has impacted pediatric patient populations who previously were not known to be at risk. Patients with congenital heart disease (CHD) represent a large, understudied population whose overweight and obesity trends and risk factors have not been well defined. The primary study goal was to assess at what age patients with CHD transition to overweight and obese and determine associated risk factors.

Methods: Retrospective chart review revealed 7849 patients with hemodynamically significant CHD born between 1996-2015. Of those, 968 were at least 2 years old and had at least 3 serial visit. Patients with chromosomal anomalies, genetic syndromes, endocrinopathies, premature birth, depression, malignancy, or autoimmune disease were excluded. Patients had to be residents of the United States, and had to receive their primary cardiology care at our facility.

Growth charts were reviewed and last normal weight, first date overweight and first date of obesity were recorded. CDC definitions of overweight and obesity were utilized. Patients were divided into 5 groups based on cardiac disease: acyanotic, cyanotic, complex biventricular, complex univentricular and electrophysiologic (EP). They were also divided into 5 year birth cohorts: 1996-2000, 2001-2005, 2006-2010, and 2011-2015. Time to event analysis assessed age at which patients became overweight or obese. The validity of this analysis was performed using ANOVA, Kruskal-Wallis, Pearson's chi-square and Fisher's exact tests.

Results: Type of CHD: cyanotic n = 256, acyanotic n = 258, complex n = 208, EP n = 246. Birth cohort: 1996-2000 n = 327, 2001-2005 n = 336, 2006-2010 n = 230, 2011-2015 n = 75. Male gender n = 549 (57%). Compared to the general population (CDC data), preschool aged children had a lower prevalence of obesity. School aged children, adolescents and young adults had a higher prevalence.

In the time to event analysis the probability of becoming overweight increased with age, however, younger patients became overweight and obese more rapidly. The median age of last normal weight in patients who became overweight was 9.0 years and median age of first recorded overweight was 10.3 years. The median age of last normal weight in patients who became obese was 6.5 years with a median age of first recorded obesity of 9.6 years.

Patients in the 2011-2015 cohort were 88% more likely to become overweight and 82% more likely to become obese compared to those born between 1996-2000.

Conclusion: The prevalence of obesity in patients CHD is comparable to the general population, but with significant differences within the age groups. The greatest risk factor for becoming overweight or obese was being in the youngest cohort. Patients with CHD transitioned to overweight and obese between ages 6.5 and 9 years which is significantly younger than the general population. Providers caring for patients with CHD should be more attentive to weight gain during that time.

Assessment of pediatric cardiologist adherence to the 2012 American Heart Association recommendations on neurodevelopmental evaluation and management of children with congenital heart disease

Sharnendra Sidhu BS[1]; Ruth Milanaik DO[2]; Tammy Pham BA[1]; Laura McLaughlin BA[1]; Edon Rabinowitz MD[1], [1]Cohen Children's Medical Center/Northwell, Lake Success, New York, [2]Cohen Children's Medical Center/Northwell Health, Lake Success, New York

Purpose: The 2012 American Heart Association (AHA) statement "Neurodevelopmental Outcomes in Children with Congenital Heart Disease (CHD): Evaluation and Management" concluded that children with CHD are at increased risk for neurodevelopmental (ND)

disorders and delays. Routine developmental surveillance and evaluation throughout childhood are recommended. It is unknown to what extent pediatric cardiologists (PC) adhere to these recommendations. The objective of this study is to assess PC familiarity and compliance with the 2012 AHA ND statement.

Methods: An anonymous online survey was distributed to PC nationwide. Surveyed data included: nonidentifiable demographics, familiarity with the AHA statement, and presence of ND cardiology program (NDP) in their affiliated institution. If no NDP was present, PC were asked to what extent and to whom CHD patients were referred for ND evaluation, or why they did not refer. All PC indicated who they believe is responsible for surveillance and referral of CHD patients.

Results: PC (N = 129) responded from 37 states. The majority of PC (54%) stated they are only "somewhat familiar" and 18% are "not familiar" with the AHA statement. Forty PC (31%) stated their institution does not have a NDP. Of these, 25% indicated they generally do not refer CHD patients for ND follow-up; 45% perform ND surveillance and refer to a specialist/early intervention (EI) if warranted; and 30% generally refer all CHD patients. Of PC without affiliated NDP who refer, 48% refer to a "primary care physician" (PCP), 28% to a "developmental pediatrician/neurologist," and 17% to "EI." Table 1 presents frequencies PC without NDP refer different CHD patients. Lastly, 43% of PC do not feel responsible for ND surveillance, and 11% do not feel responsible for ND referrals. Table 2 indicates PC beliefs about responsibility for ND surveillance and referral.

Conclusion(s) : It is widely accepted that CHD, particularly those requiring operative repairs, is associated with increased ND risk. However, in this survey, most PC stated that they are not familiar or only somewhat familiar with AHA ND guidelines. This suggests that a substantial proportion of PC is not referring high-risk children for adequate ND care. It is essential that the ND risks associated with CHD be widely disseminated amongst PCP and PC, and that current guidelines are reinforced to ensure all patients are appropriately screened and referred for services.

Assessment of pediatric cardiology referral practices for inpatient and outpatient post–cardiac surgery physical therapy

Sharnendra Sidhu BS[1]; Ruth Milanaik DO[2]; Tammy Pham BA[1]; Laura McLaughlin BA[1]; Edon Rabinowitz MD[1], [1]Cohen Children's Medical Center/Northwell, Lake Success, New York, [2]Cohen Children's Medical Center/Northwell Health, Lake Success, New York

Purpose: Congenital heart disease (CHD) occurs in 4-12 per 1000 live births with more than 30% of affected children requiring palliative or corrective surgery in early life. Studies on adult cardiac surgery patients clearly demonstrate the benefit of physical therapy (PT), a standard postoperative care step. However, it is unknown to what extent pediatric cardiac patients are referred for postsurgical PT in both inpatient and outpatient settings. The objective of this study is to assess pediatric cardiologist (PC) referral practices for both inpatient and outpatient post–cardiac surgery physical therapy.

Methods: An anonymous, three-part survey was distributed to PC nationwide via Survey Monkey. In Part 1, PC were asked nonidentifiable demographic questions, including those on practice setting and post–fellowship training. In Part 2, PC indicated how many outpatients they typically see in a week as well as how much time they commit to teaching or supervising PC fellows in the outpatient setting. In Part 3, subjects specified how often they refer CHD patients postoperatively for inpatient and outpatient PT: never (0% of the time), rarely (1-20%), sometimes (21-50%), often or very often (51-80%), always or almost always (81-100%). Linear regressions were performed to determine whether the rate of referral differed across community type or teaching load.

Results: PC (*N* = 129) responded from 37 states: 53% had completed post–fellowship training, 72% works in an urban setting, 80% has been in practice for ≥ 5 years, and 85% are primarily hospital-based. Physician PT referral practices are shown in Figures 1 (inpatient) and 2 (outpatient). Only 26% of surveyed PC "always or almost always" refer postoperative cardiac patients for inpatient PT, and only

TABLE 1 PC without NDP Referral Practices for CHD patients

	N/A /I do not see these kinds of patients	Never (0% of the time)	Rarely (1-20% of the time)	Sometimes (21-50% of the time)	Often or Very Often (51-80% of the time)	Always or Almost Always (81-100% of the time)
Infants (less than 12 months) requiring open heart surgery (cyanotic and acyanotic types).For example, HLHS, IAA, PA/IVS, TA, TAPVC, TGA, TOF, tricuspid atresia.	2 (2%)	0 (0%)	11 (9%)	25 (21%)	14 (12%)	70 (57%)
Children (over 12 months) with other cyanotic heart lesions NOT requiring open heart surgery as a neonate/infant. For example, TOF with PA and MAPCA(s), TOF with shunt without use of CPB, Ebstein anomaly.	4 (3%)	1 (1%)	32 (26%)	33 (27%)	25 (21%)	27 (22%)

	n (%)
Pediatric Cardiologist	11 (10%)
Cardiac ND Follow-up Clinics	27 (24%)
Primary Care Physician	67 (59%)
Pediatric Neurologist/Developmental Pediatrician	8 (7%)

TABLE 2 PC Beliefs about who is Responsible for ND Surveillance and Referral

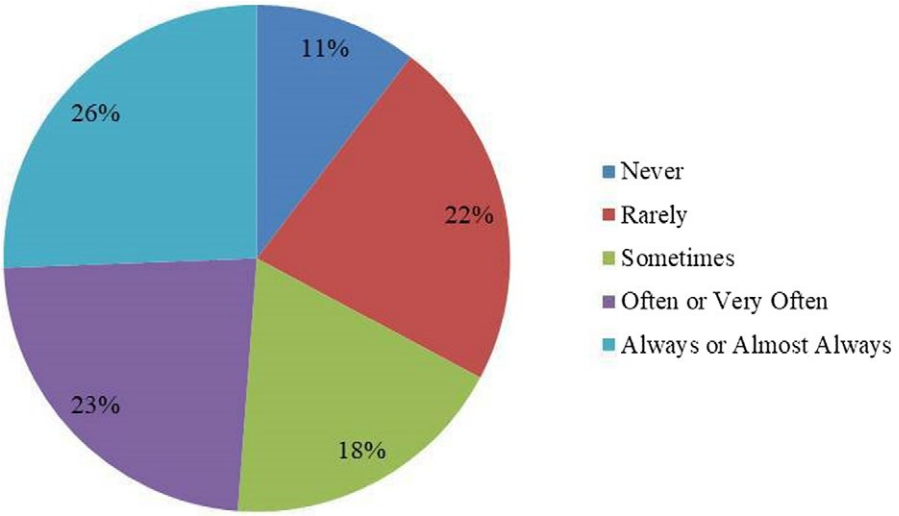

FIGURE 1 Responses to question: For patients who require or had significant cardiac surgery, how often do you or your staff refer patients postoperatively for inpatient physical therapy?

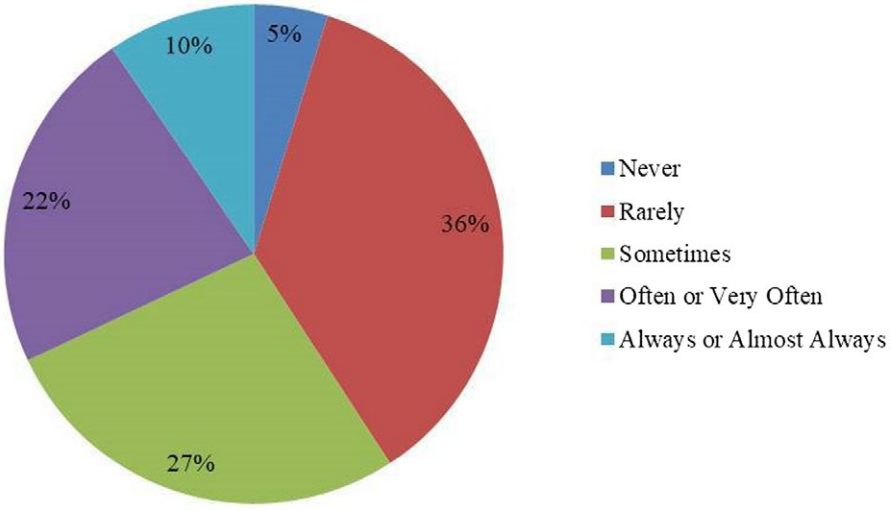

FIGURE 2 Responses to question: For patients who require or had significant cardiac surgery, how often do you or your staff refer patients postoperatively for outpatient physical therapy?

10% for outpatient PT. In addition, 33% and 41% of PC stated they "never" or "rarely" refer for inpatient PT or outpatient PT, respectively. Rate of referral for both inpatient and outpatient PT were the same across urban, rural, and suburban settings. However, those who spent more hours teaching per week were significantly more likely to refer for both inpatient PT ($\beta = 0.27$, $P = .006$) and outpatient PT ($\beta = 0.17$, $P = .03$).

Conclusion(s): Research has indicated PT as standard of care for adult postoperative patients. However, given the low rates of referral to pediatric PT found in this study, it is imperative that more research investigate the potential benefits of PT for postoperative pediatric patients.

Association of fetal and maternal factors with mortality due to diseases and malformations of the circulatory system in children

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Purpose: To verify the association of characteristics recorded at the time of birth, including weight, occurrence of asphyxia, gestation

TABLE 1 Crude and adjusted relative risks of death due to diseases or malformations of the circulatory system relative to the population of live births according to predictive variables in children and adolescents below the age of 18 years in the state of Rio de Janeiro, from 1996 to 2014

Predictive Variables	DCS N	MCS N	Live Births N	DCS CRUDE RR (LL-UL(95% CI)) %	DCS CRUDE FAP	DCS ADJUSTED* RR(LL-UL(95% CI))	MCS CRUDE RR (LL-UL (95% CI)) %	MCS CRUDE FAP	MCS ADJUSTED* RR (LL-UL (95% CI))
Birth Weight									
Adequate	611	2.619	3.715.583	**1**	-	**1**	**1**	-	**1**
Low	196	1.335	342.531	**3,48** *(2,96-4,09)*	**17,30**	**2,26** *(1,82-2,81)*	**5,53** *(5,18-5,91)*	**27,64**	**2,96** *(2,70-3,25)*
High	34	146	217.069	**0,95** *(0,67-1,34)*	**-0,26**	**0,96** *(0,63-1,36)*	**0,95** *(0,81-1,13)*	**~0,25**	**0,89** *(0,75-1,06)*
Apgar 1									
Normal	510	2.046	3.294.362	**1**	-	**1**	**1**	-	**1**
Asphyxia	309	1.927	842.365	**2,37** *(2,06-2,73)*	**21,80**	**1,73** *(1,47-2,04)*	**3,68** *(3,46-3,92)*	**35,31**	**2,10** *(1,95-2,27)*
Apgar 5									
Normal	709	3.046	3.971.697	**1**	-	**1**	**1**	-	**1**
Asphyxia	104	927	166.137	**3,51** *(2,85-4,31)*	**9,14**	**1,52** *(1,19-1,94)*	**7,28** *(6,76-7,83)*	**20,11**	**2,60** *(2,37-2,85)*
Gestational Age									
Term	668	2.952	3.876.594	**1**	-	**1**	**1**	-	**1**
Preterm	163	1.076	309.138	**3,06** *(2,58-3,63)*	**13,20**	**1,49** *(1,18-1,88)*	**4,57** *(4,26-4,90)*	**20,85**	**1,40** *(1,26-1,54)*
Post-term	7	47	49.435	**0,82** *(0,39-1,73)*	**~0,22**	**0,86** *(0,41-1,82)*	**1,25** *(0,94-1,67)*	**-0,31**	**1,29** *(0,97-1,73)*
Maternal Age									
20 to 29 years	400	1.957	2.231.102	**1**	-	**1**	**1**	-	**1**
< 20 years	203	772	842.523	**1,34** *(1,14-1,60)*	**8,6**	**1,17** *(0,98-1,40)*	**1,04** *(0,96-1,14)*	**1,21**	**0,93** *(0,85-1,01)*
30 to34 years	127	677	736.367	**0,96** *(0,79-1,17)*	**-0,94**	**0,96** *(0,78-1,18)*	**1,05** *(0,96-1,14)*	**1,18**	**1,06** *(0,97-1,16)*
35 to 39 years	73	442	364.377	**1,12** *(0,87-1,43)*	**1,62**	**1,10** *(0,85-1,42)*	**1,38** *(1,25-1,53)*	**5,10**	**1,31** *(1,18-1,46)*
≥40 years	39	254	95.745	**2,27** *(1,64-3,16)*	**4,97**	**2,05** *(1,47-2,86)*	**3,02** *(2,65-3,45)*	**7,68**	**2,53** *(2,21-2,89)*
Maternal Education Level									
High	102	571	715.828	**1**	-	**1**	**1**	-	**1**
Middle	301	1.597	1.625.828	**1,30** *(1,04-1,63)*	**17,20**	**1,26** *(1,00-1,59)*	**1,23** *(1,12-1,35)*	**13,83**	**1,23** *(1,11-1,35)*
ELEMENTARY AND NO SCHOOLING	421	1.842	1.849.316	**1,60** *(1,29-1,98)*	**30,11**	**1,48** *(1,18-1,86)*	**1,25** *(1,14-1,37)*	**15,19**	**1,15** *(1,04-1,27)*
No information	17	97	85.377	**1,40** *(0,84-2,33)*	**4,06**	**1,22** *(0,72-2,08)*	**1,42***(1,15-1,77)*	**4,32**	**1,12** *(0,89-1,41)*

DCS: diseases of the circulatory system (chapter IX of ICD-10); MCS: malformations of the circulatory system (Q20-28 of ICD-10); FAP: fraction attributable to population; LL-UL (95%CI): Lower limit–Upper limit (95% confidence intervals); *adjusted estimates were obtained with models that included all the predictive variables

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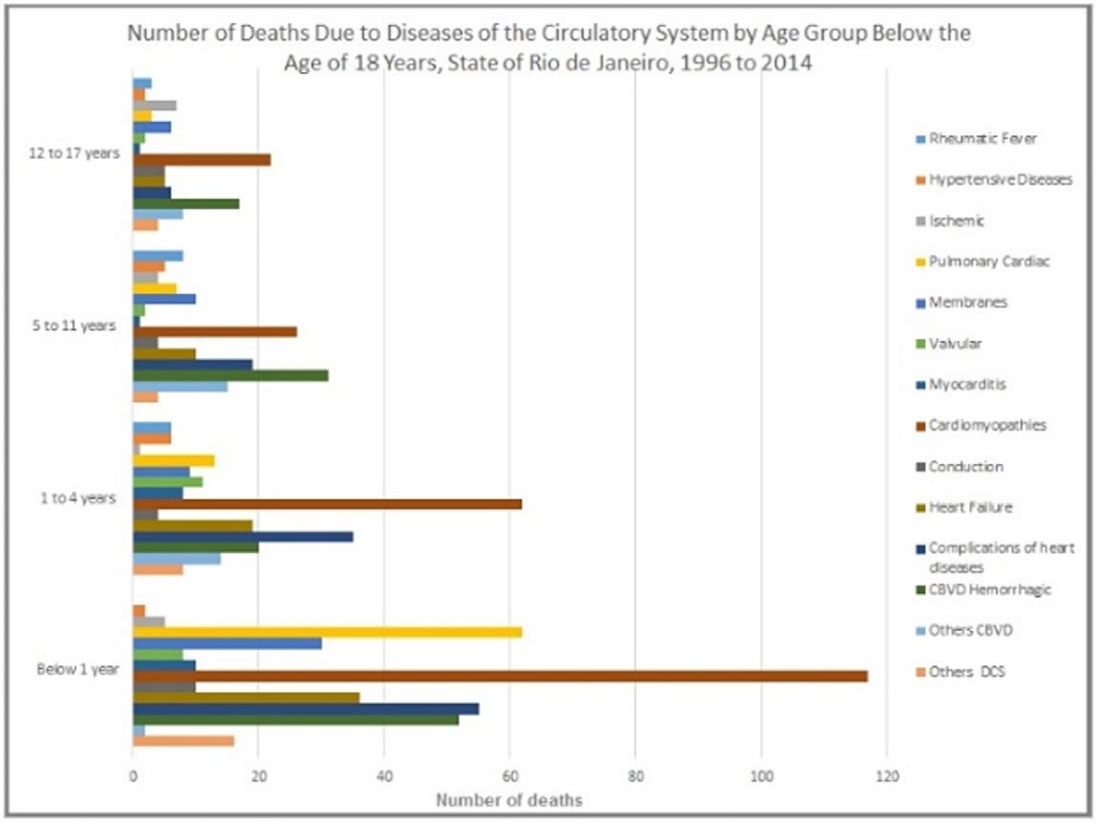

FIGURE 2 Number of deaths due to diseases of the circulatory system by age group below the age of 18 years, state of Rio de Janeiro, 1996 to 2014

duration, and maternal age and education level, with death from diseases and malformations of the circulatory system below the age of 18 years in children born and deceased between 1996 to 2014 in the state of Rio de Janeiro, Brazil.

Methods: The databases Information System on Live Births and Information System on Mortality were linked and evaluated following a strategy of longitudinal cohort analysis. We estimated the crude relative risks (RRs) and the RRs adjusted for the variables birth weight, Apgar scores at 1 and 5 minutes, gestation duration, and maternal age and education level.

Results: We linked 6380 deaths with 4 282 260 birth records, yielding 6097 linked pairs. Low birth weight (RR = 2.26), asphyxia at 1 (RR = 1.72) and 5 minutes (RR = 1.51), prematurity (RR = 1.50), maternal age ≥ 40 years (RR = 2.06), and low maternal education level (RR = 1.45) increased chance of death due to diseases of the circulatory system. In the association with death due to malformations of the circulatory system, the predictive variables showed the same association profile, but with greater intensity.

Conclusion: Fetal factors studied were associated with an increased mortality due to diseases and malformations of the circulatory system. Measures to control these factors and improve access to their diagnosis and treatment would contribute to reducing these deaths. However, the identification of environmental influences during gestation and birth on the risk of death should be carefully considered, as they are influenced by genetic factors.

Cardiopulmonary exercise performance in children after Fontan

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Background: Monitoring for Fontan associated morbidity is becoming increasingly important due to increased survival rates. Cardiopulmonary exercise performance (CPX) is an objective measurement of cardiovascular, pulmonary, and metabolic health. CPX has long been used to evaluate for coronary health in the adult population and its use as a screening tool in patients with congenital heart disease is becoming more common. Interpretation of cardiopulmonary testing in the Fontan population is limited and somewhat controversial.

Objective: The objectives of this study include describing the cardiopulmonary performance in a contemporary cohort of Fontan patients and assessing factors associated with poor performance on CPX.

Methods: A retrospective chart review of CPX for Fontan patients between 2011-2017 was performed in a single institution. Regression analysis was performed to assess factors associated with low peak VO_2.

Results: Ninety-five patients underwent CPX between 2011-2017. Mean age of at time of CPX was 13.22 years with a gender distribution of 47 females and 48 males. Primary diagnosis was consistent with systemic left ventricle (LV) in 48, systemic right ventricle (RV) in 43, and ambiguous ventricle in 4 patients. Mean age at Fontan was 32.4 (12-107) months. Resting heart rate (HR) was less than 80 beats per minute in 32% of patients.

Analysis of Fontan patient's performance on CPX showed the following data. Mean duration of CPX testing was 7.1 (4.1-17) minutes. Mean peak VO_2 was 30.3(10-48.4) mL/kg/min and 72% (25-120%) of predicted performance. Mean anaerobic threshold was 38.2% (7-89%) of predicted performance, and the mean maximum O2 pulse was of 8.3 (2-18) ml/beat. Average oxygen saturations decreased from 91.4% baseline to 84.4% at peak exercise. The majority of tests were stopped due to patient fatigue. Further analysis showed that 55% of early Fontan patients, as defined as Fontan before 36 months of age, had VO_2 max greater than 30 mL/kg/min compared to 40% of late Fontan patients. Multiple regression analysis showed female gender was the only predictor of low peak VO_2 ($P = .001$). Additionally, patents with systemic RV had significantly lower peak percent predicted HR compared to those with systemic ($P = .029$), but no other difference in performance.

Conclusion: Poor performance on CPX is very common in Fontan patients. Nonmodifiable factors including gender and systemic RV along with modifiable factors such as heart rate and age at time of Fontan are associated with poor performance in a large cohort of contemporary patients. This study provides further information for targeted interventions to improve cardiopulmonary performance and quality of life in these patients.

Characteristics and outcomes of infants diagnosed with congenital cerebral arteriovenous malformations: 18-year statewide experience

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Purpose: Congenital cerebral arteriovenous malformations (CAVMs) such as vein of Galen malformations are a rare cause of high output cardiac failure in infants. The objectives of this study were to evaluate the baseline characteristics and outcomes of CAVMs in infant hospitalizations using a large statewide population-based inpatient database.

Methods: Authors conducted a retrospective analysis of the Texas Public Use Inpatient Data File from January 1999 to December 2016. All discharges less than one year of age with the diagnosis of CAVM were identified by ICD-9 and ICD-10 codes. Primary admission data were included, and subsequent admissions were excluded. If the birth admission was followed by transfer to a Texas tertiary care facility within 48 hours, the birth admission was excluded. Birth

prevalence of CAVM was estimated based on primary admission data divided by the number of live births over the same period from Texas vital statistics data. Diagnoses of heart failure (HF), pulmonary hypertension (PH), and congenital heart disease (CHD), as well as endovascular embolization and mechanical ventilation (MV) were queried. Univariate and multivariate analyses for mortality were performed using logistic regression, excluding those discharged to hospice (n = 4). Subanalysis was performed examining associations between embolization and mortality in the subset of hospitalizations with a heart failure diagnosis.

Results: A total of 371 hospitalizations with CAVMs were identified over the 18-year study period, including 326 primary hospitalizations. Estimated prevalence of CAVM was 4.7/100 000 or 1/21 277 live births. Mortality during primary admission was 15.6%. CHD (31.3%), HF (21.8%) and PH (16.6%) were common. The proportion with CHD other than atrial septal defect/patent foramen ovale/patent ductus arteriosus was 11%. Mechanical ventilation was used in 31%, and 20% underwent vascular embolization. Median length of stay was 7 days (IQR 2-20), and median total charges were $60 778 (IQR $24 645.64-$161 120.91). Variables associated with mortality by univariate analysis were HF (mortality 37.1%, OR 5.37, CI 2.84-10.14, $P < .0001$), PH (mortality 35.8%, OR 4.14, CI 2.11-8.1, $P < .0001$) and MV (36%, OR 7.45, CI 3.84-14.47, $P < .0001$). Non-Hispanic Blacks had higher mortality compared to non-Hispanic whites (mortality 24.4% vs 10.7%, OR 2.71, CI 1.08-6.76, $P = .033$). In multivariate analysis, variables that remained associated with mortality were Non-Hispanic black race (OR 3.15, CI 1.11-8.90, $P = .030$), HF (OR 4.26, CI 1.92-9.48, $P < .0001$) and MV (OR 6.14, CI 3.04-12.47, $P < .0001$). There was no association between embolization procedures and mortality in univariate analysis. When limiting to the HF cohort (n = 71), embolization was also not associated with mortality.

Conclusion: Infants with CAVMs such as vein of Galen malformations have a high risk of mortality during their initial hospitalization. HF and MV were associated with increased mortality, as was Black race. More research should be aimed at which patients may have survival benefit from embolization.

Comparison of echocardiographic criteria for diagnosing left ventricular noncompaction in patients with clinical cardiomyopathy

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Background: Isolated noncompaction of the ventricular myocardium (INVM) is a cardiomyopathy characterized by an excessive number of prominent trabeculations with deep intertrabecular recesses in the left ventricular apex. Associated findings include systolic dysfunction, ventricular arrhythmias, and systemic emboli. Management of patients with INVM is challenging not only because of the variability in developing clinical myopathy, but also because multiple diagnostic criteria are in use. INVM remains most commonly diagnosed by echocardiography with three criteria in use. These include the criteria proposed by Lai based on the study by Chin, and subsequent criteria proposed by Jenni and Stollberger.

Objective: We evaluated the three criteria for INVM to compare the sensitivity for detecting patients ultimately developing clinical cardiomyopathy. Serial echocardiograms were retrospectively reviewed from four patients with INVM who ultimately developed clinical cardiomyopathy (increased LV trabeculations, LV systolic dysfunction > 2 SD below normal).

Methods: Ten echocardiograms from four patients (age range 12-25 years, 2 male, 2 female) with clinical INVM were evaluated. A minimum of two echocardiograms obtained between one and fifteen years apart were studied. Only those echocardiograms felt by at least one reviewer to allow measurement using all three criteria were included. The studies were reviewed independently by two pediatric cardiologists and the interobserver agreement for INVM was assessed.

Results: The average fractional shortening for the 4 patients measured 21.9% ± 6.2%. Using the Chin criteria the X:Y ratio was 0.31 ± 0.1 (normal > 0.5), and fractional shortening and the X:Y ratio showed modest correlation with a $R^2 = 0.43$. Interobserver agreement was 100%, with INVM diagnosed by both cardiologists in all patients on all echocardiograms. Using the Stollberger criteria, INVM was likewise eventually diagnosed by both cardiologists in all patients, but interobserver agreement was 56%. Finally, using the Jenni criteria, INVM was diagnosed in 1 of 4 patients by one cardiologist, and 4 of 4 patients by the second cardiologist with an interobserver agreement of 29%.

Conclusion: Our review demonstrates that the Chin criteria most consistently diagnosed INVM in patients with clinical cardiomyopathy and demonstrates the best interobserver agreement. Criteria for diagnosis were met in some patients even prior to developing clinical cardiomyopathy indicating there may be a predictive value in using these criteria. Further, there is modest correlation between function and X:Y ratio using this criteria suggesting a correlation between the degree of hypertrabeculation and the severity of ventricular dysfunction in INVM. However, all 4 patients were ultimately diagnosed with INVM using any of 3 criteria on at least one echocardiogram by at least one cardiologist.

Congenital heart disease is a risk factor for the success of catheter ablation in children <15 kg

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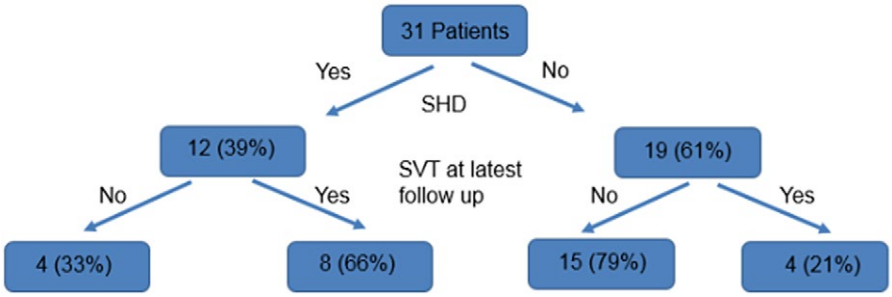
Catheter ablation has been successful for small children (≤15 kg) in selected cases, however there is a paucity of data comparing the effect of congenital heart disease in this population. We sought to describe the experience at our center with catheter ablation in children <15 kg and compare the safety and efficacy of this procedure as it related to the presence of congenital heart disease. This was a

Follow up tree



Flow chart of patients by congenital heart disease and long-term freedom from arrhythmia following catheter ablation

Demographics

Demographics	No SHD	SHD
Patients	19	12
Age (months)	18.5	11.2
Weight (kg)*	11.3	8.8
Procedures	20	18
Acute Procedural Success*	95%	56%
Complication	3 (15%)	1 (6%)
Follow up (years)	6.2	8.1
Long-term Success*	15 (79%)	4 (33%)

Descriptive statistics, procedural and latest follow up data of patients with and without congenital heart disease. *statistically significant at *P* value <.05

retrospective chart review of all patients that had catheter ablation from 2000-2013, weighing less than 15 kg because of a diagnosis of supraventricular tachycardia. There were 31 patients included with 38 catheter ablation procedures performed in this time period. The data collected included procedure data, preablation and postablation follow-up. We used chi-square and Fisher's exact test for comparison when appropriate. Children without structural congenital heart disease or cardiomyopathy were much more likely to have long-term success (79% vs 33%, *P* value .02) and decreased incidence of recurrence (16% vs 50%, *P* value .08) when compared to children with congenital heart disease. There were a total of 4 cases with complications (11%) of which only 2 (5%) were considered severe. The median follow up for all patients was 6.6 years (IQR 2.1-10.1 years). These data suggest that small children with supraventricular tachycardia who undergo catheter ablation are much more likely to have a successful ablation if they have no underlying congenital heart disease or cardiomyopathy when compare to those with congenital heart disease or cardiomyopathy. While there was a trend of increased incidence recurrence in patients with congenital heart disease this did not reach statistical significant. Catheter ablation could be considered a relatively safe procedure with low incidence of complications.

Congenital heart disease management in trisomy 13 and 18: A survey of pediatric cardiology providers

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Purpose: During the past decade, the care of patients with Trisomy 13 (T13) and Trisomy 18 (T18) has continued to evolve, with trends in the literature indicating that intervention for these patients remains controversial but is increasingly considered. Studies, limited by small sample size, selection bias, and intent to treat, have found conflicting evidence as to whether procedural management of cardiac defects in patients with T13 and T18 improves survival time. Best practices for these patients are not well defined, and the aim of this work is to identify current perspectives and practices of pediatric cardiology practitioners.

Methods: A survey was sent to all members of the AAP Section on Cardiology and Cardiac Surgery. Respondents were asked whether, presuming that a patient with T13/T18 could breathe spontaneously and family requested treatment, they would consider specific catheter interventions or Risk Adjustment for Congenital Heart Surgery (RACHS) category interventions to be appropriate. Other questions focused on provider perceptions regarding T13/T18 quality of life, decision making, goals of care, and measures of care such as extracorporeal membrane oxygenation (ECMO).

Results: A total of 60 respondents participated, for a response rate of 15%. Most were noninterventional pediatric cardiologists (73%). Respondents skewed male (75%), were likely to work in a university hospital system (72%), and had a mean of 18.6 years in practice (median 20 years). Although there was some variation, over half of respondents indicated that, with the exception of PDA stenting, each catheterization procedure addressed by the survey

TABLE 1 Perceived appropriateness of catheter interventions and refusal to offer intervention in T13/T18

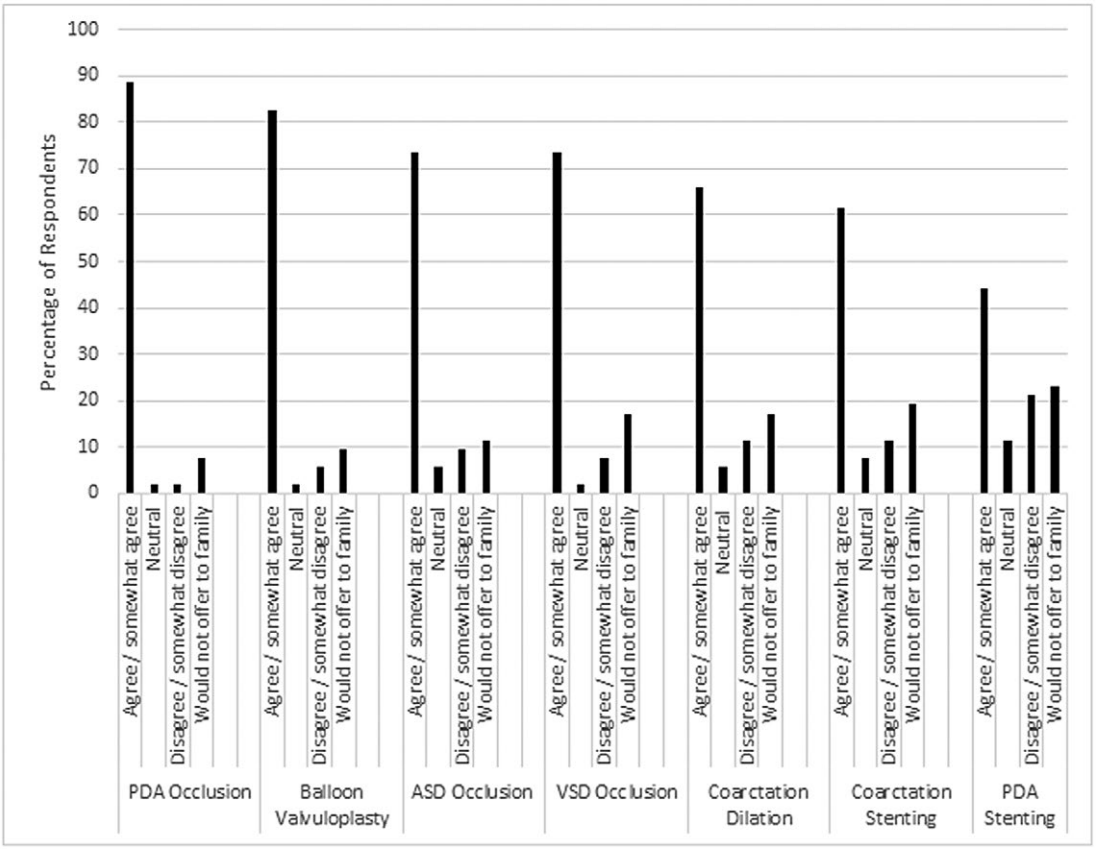
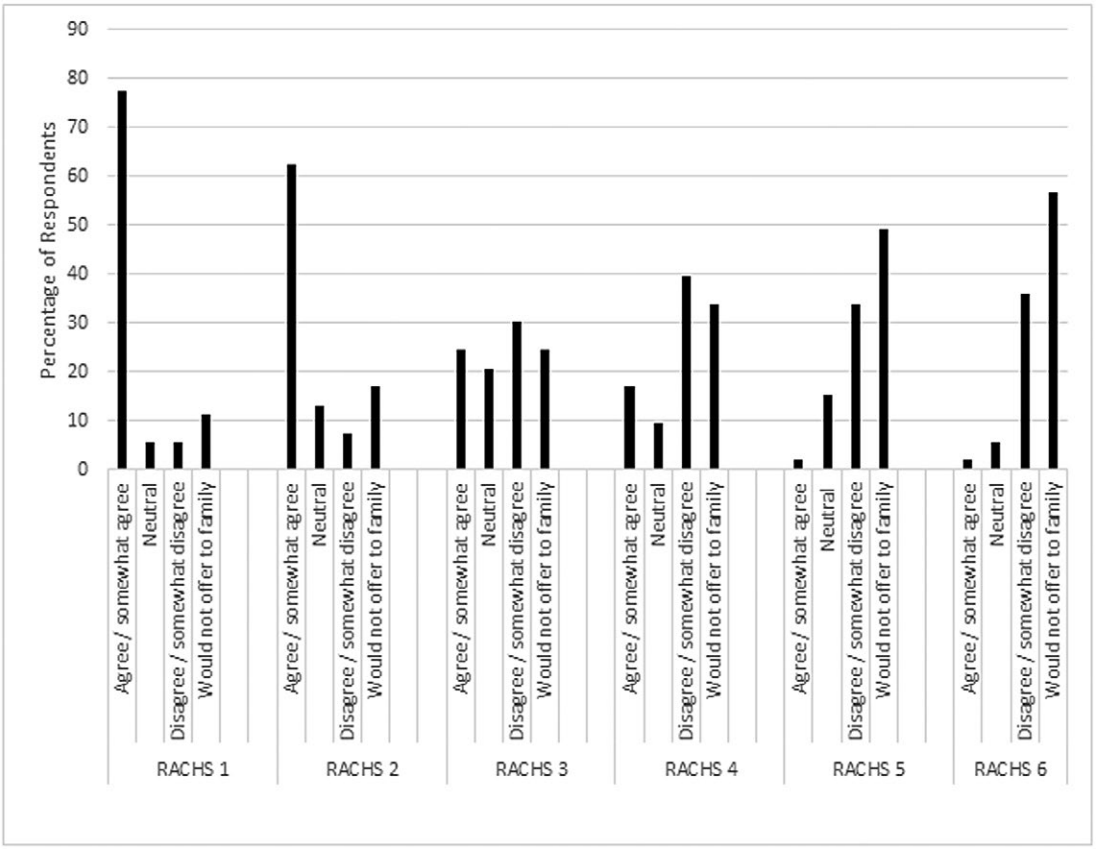


TABLE 2 Perceived appropriateness of surgical interventions and refusal to offer intervention in T13/T18



was appropriate for T13/T18 patients. Respondents were less likely to indicate that a catheterization was appropriate as procedural risk and complexity increased (Figure 1). Most providers felt that RACHS 1 and 2 surgical interventions were appropriate in T13/T18 patients. Perception of appropriateness for surgical intervention fell for RACHS 3 and thereafter (Figure 2). Only 1 respondent (2%) indicated that ECMO is sometimes reasonable in T13/T18 patients prior to cardiovascular intervention, while 8% felt that ECMO is sometimes appropriate after interventional catheterization and after cardiovascular surgery.

Almost half (49%) of respondents reported that T13/T18 patients have a poor quality of life, while 41% felt that T13/T18 patients can have a fair quality of life. Most respondents felt very comfortable (55%) or somewhat comfortable (34%) in helping to make decisions with T13/T18 patients. Participants indicated that the respiratory status of patients affects (37%) or may affect (32%) their recommendation for intervention.

Conclusion: Overall findings demonstrate variability amongst provider perspectives pertaining to the care of patients with T13/T18. However, trends exist regarding the appropriateness of specific interventions based on intervention complexity and risks.

Correlation between birth weight and left ventricular hypertrophy in infants of diabetic mothers

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Purpose: Infants of diabetic mothers (IDM) are known to have numerous complications during the neonatal period, including varying degrees of left ventricular hypertrophy (LVH). The purpose is to assess a correlation between birth weight and maternal 3rd trimester HbA1c to the degree of LVH.

Methods: A retrospective review at a tertiary university single-center for all neonates admitted to the NICU/NBN from 03/01—07/17. Inclusion criteria: maternal history of diabetes, echocardiogram (echo). LVH defined as LV mass (LVm) > 95th percentile based on gender, male (17.6 g, LVm index (LV mi) 80.1 g/m$^{2.7}$) and females (16.5 g, LVmi 85.6 g/m$^{2.7}$). Normal HbA1c in third trimester was set at 6.0%. Data obtained included demographics, birth weight, maternal HbA1c and echo measurements.

Results: Total of 53 IDM, equal number of males, 50% (P = 1.0). Gestational age range 30.5-39.4 weeks; preterm 31 (60%), term 21 (40%), (P < .001). Birth weight range 1.2-5.370 kg, SGA; 1 (2%), AGA; 23 (44%), LGA 28 (54%), (P < .001). LVm range 3.28-19.1g, LVmi range 27.9-150.24. Maternal third trimester HbA1c range 5.1-11.8%; good control 12 (29%), poor control 30 (71%) (P < .0001).Based on gender, 1 male by LVm and 7 by LVmi; 0 female by LVm and 4 LVmi. There is a positive correlation of birth weight and LVm, R = 0.49 (P males with LVH, insufficient data based on LVm alone, for LVmi there is a positive correlation, R = 0.158 (P = .73). Females with LVH, insufficient data for both LVm and LVmi. For the HBA1c group (42 infants), 1 male by LVm and 7 by LVmi; 0 female by LVm and 2 LVmi. A positive correlation of maternal HbA1c level and LVm, R = 0.11 (P = .50), and with LVmi, R = 0.23 (P = .135).

Conclusion: IDM are known to have increased risk of cardiovascular problems, including LVH. From these results, this cohort is more likely to be premature and LGA and our study population is more likely to have poor control of their diabetes. The weakly positive correlations between birth weight and maternal HbA1c to and LVm in infants suggest that by focusing on poorly controlled diabetic dyads or those LGA infants alone, infants with increased risk for LVH maybe missed and not followed over time.

Cost of childhood chest pain evaluation is higher in emergency departments with fewer pediatric resources

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Purpose: Chest pain is a common chief complaint in children presenting to emergency departments and pediatric cardiologists. Despite the low incidence of cardiac pathology in children with chest pain, costly evaluations with uncertain benefits persist. The purpose of this study is to assess resource utilization across hospitals with varying levels of pediatric resources and compare clinical outcomes. Our hypothesis is that hospitals with fewer pediatric resources will generate higher costs without increased rates of specific diagnoses.

Methods: We performed a secondary analysis of emergency department (ED) data from the Arizona Hospital Discharge Database (AZHDDB). ED visits with a patient age 5-18 years and a chief complaint of chest pain from 2012-2016 were compared by hospital type determined by pediatric resource status (Pediatrics with PICU, Pediatrics with no PICU, No pediatrics). Patient demographics, resource utilization, total visit cost, and patient outcomes (defined by specific diagnosis and/or discharge status) were analyzed. Specific diagnoses included all primary discharge ICD 9/10 codes that did not fall under 780-799/R00-99 classifications and/or were determined to be indicative of an actual clinical diagnosis by expert review.

Results: There were 28 342 encounters during the entire study period, 11 547 (40.7%) in Pediatrics with PICU, 3562 (12.8%) in Pediatrics with no PICU and 12 666 (46.3%) in no pediatrics. Patients seen in EDs with fewer pediatric resources were more likely to be transferred to another facility (2.9% no pediatrics vs 1.6% Pediatrics with no PICU vs 0.5% Pediatrics with PICU, P <.001). 16 078 (58%) of discharge diagnoses were nonspecific, 60% of patients were discharged with the same presenting symptom code and 812 (2.9%) were assigned codes referable to the cardiovascular system, most

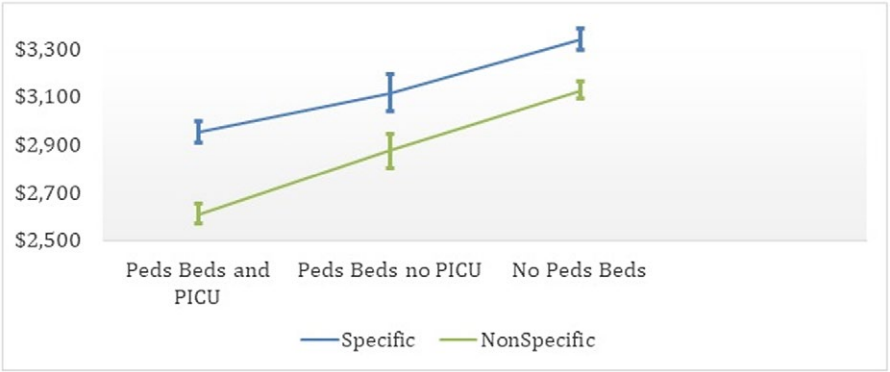

FIGURE 1 Specific diagnosis at discharge was significantly associated with higher costs as were lower levels of pediatric resources (P = .000), no difference in rates of specific diagnosis among hospital types (P = .271)

Procedure	Peds with PICU	Peds with no PICU	No Peds	p
Laboratory	n = 4,487 $746 ± 769	n = 1,441 $1,048 ± 745	n = 5,248 $1,111 ± 930	< 0.001
Diagnostic radiology	n = 8,720 $479 ± 217	n = 2,882 $522 ± 237	n = 10,669 $570 ± 274	< 0.001
ECG	n = 6,565 $369 ± 167	n = 1,662 $352 ± 166	n = 6,337 $412 ± 160	< 0.001
Respiratory	n = 959 $336 ± 501	n = 292 $247 ± 244	n = 982 $400 ± 406	< 0.001
CT scan	n = 438 $3,986 ± 2,927	n = 233 $6,220 ± 4,769	n = 1,043 $4,870 ± 3,857	< 0.001

TABLE 1 Comparison of costs for most commonly performed diagnostic tests

commonly dysrhythmia. Facilities with lower levels of pediatric resources spent significantly more on diagnostic testing (Table 1). A specific diagnosis at discharge was associated with higher visit costs but there was no difference between rates of specific diagnosis among hospital types (Figure 1).

Conclusions: Childhood chest pain evaluations in the emergency department often yield a nonspecific discharge diagnosis and costs are higher at centers with fewer pediatric resources. Cardiovascular etiologies for childhood chest pain are uncommon and more costly evaluations do not result in identification of a specific diagnosis. Further studies are warranted to evaluate the use of standardized evaluation plans among varying hospital types and their impact on resource utilization.

Effect of transcatheter closure of the ductus arteriosus in preterm infants

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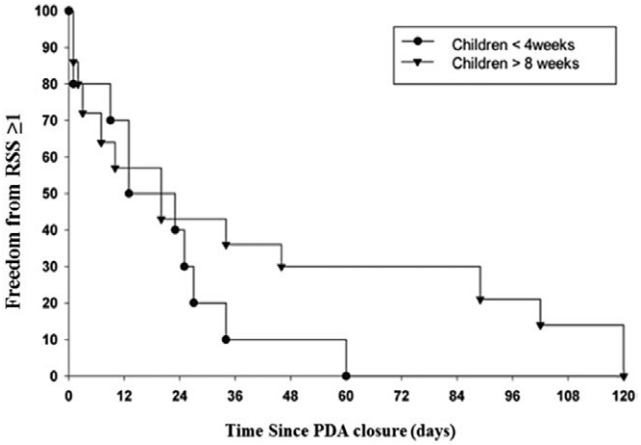
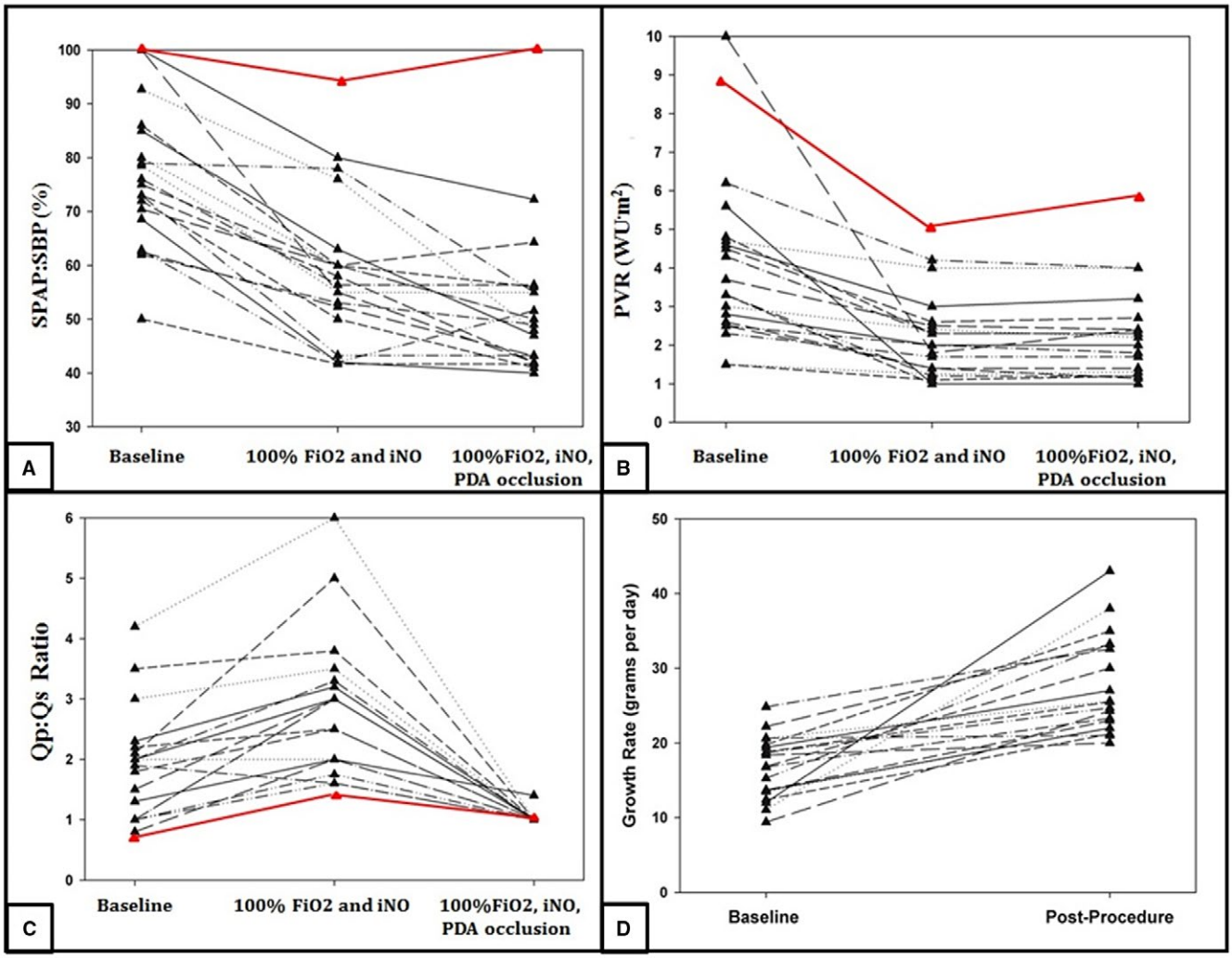
Purpose: Patent ductus arteriosus (PDA) is a common clinical condition in preterm infants with hemodynamic consequences of systemic hypoperfusion and later pulmonary hypertension. Management of pulmonary hypertension in the presence of a large PDA with left to right shunting is challenging. We sought to assess the effect of the PDA and its transcatheter device closure on the pulmonary vascular bed in preterm infants.

Methods: A review of 100 consecutive children who underwent trans-catheter device closure of PDA was performed. Preterm infants who had a diagnostic right heart catheterization were chosen. Hemodynamic measures including Qp:Qs, pulmonary systolic pressure as a percentage to systemic pressure (PAP%) and pulmonary vascular resistance (PVR, expressed as wood units.m^2) were measured at baseline, with 100%Fio2 and 20 ppmiNO (condition2) and after test occlusion of the PDA (condition3). Respiratory severity scores (RSS) were also assessed.

Results: There were 33 preterm infants, of which 17 were ≤2.5 Kg, 12 were ventilator dependent. The median gestational age of this subgroup was 27 weeks (23-32 weeks). The median age and weight at the time of the procedure were 6 weeks (4-16 weeks) and 1.6 Kg (1.06-2.5 Kg) respectively. The mean baseline Qp:Qs was 2.2:1 which increased to 4:1 with condition2 and down to 1 with condition3 as expected. The mean PAP% was 78%, which decreased to



Kaplan-Meier curves comparing the time taken for RSS to decline to a value of <1 from the time of the PDA closure between those who had early (<4 weeks) vs delayed (>8 weeks)



Hemodynamic assessment in infants with elevated pulmonary artery (PA) pressure in 3 conditions: baseline, with 100% oxygen and 20 ppm iNO and with test occlusion of the PDA. A, Systolic PA pressure (SPAP) as a percentage of the systolic blood pressure (SBP). B, Pulmonary vascular resistance index (PVR)

42% with condition2&3 as did the PVR (baseline: 4.1, condition2: 1.9, condition3: 2.1). The median days for extubation were 17. The predictors for prolonged (>30-days postprocedure) elevation of RSS (Score≥1) were: closure performed at >8 weeks of age and, SPAP ≥50% of the SBP (OR = 2.86, 95%CI: 1.5-4.2, P = .05 and OR = 5.4, 95%CI: 2.2-9.4, P < .01 respectively).

Conclusions: Apart from systemic hypo-perfusion and left ventricular volume overload, the PDA is an additional contributory factor to worsening pulmonary hypertension in preterm infants. The pulmonary vascular bed remains reactive to pulmonary vasodilator therapy. Transcatheter device occlusion of the ductus not only enables test occlusion of the PDA, but also facilitates the use of pulmonary vasodilator therapy in this cohort by eliminating the left to right shunt. It may be beneficial to perform early closure at < 4weeks of life for an expeditious improvement in RSS.

Effects of prenatally diagnosed congenital heart disease on timing and mode of delivery

Joanna Lowisz DO; Rupali Gandhi MD, JD; Sheena Mansukhani DO; Yi Li MS, Advocate Children's Hospital, Oak Lawn, Illinois

Purpose: Prenatal diagnoses of congenital heart disease (CHD) may prompt nonspontaneous delivery (NSD). This may predispose infants and mothers to unnecessary risks associated with Caesarean section (CS) or planned induction of labor (IOL). However, there is no consensus on the appropriate mode of delivery for a fetus with known CHD. In addition, most fetuses with CHD are not at increased intrapartum risk or unable to tolerate spontaneous labor (SL). We hypothesize that a prenatal CHD diagnosis is positively correlated with the infant's likelihood of undergoing induced labor and as a result, longer hospital stay and higher cost of hospitalization for infant and the new mother.

The purpose of the study is to determine if a prenatal CHD diagnosis impacts mode of delivery, describe the presence of obstetric (OB) and/or CHD indications for NSD, determine if timing and mode of delivery, when controlled for the severity of prenatally diagnosed CHD, is associated with postnatal clinical length of stay and cost.

Methods: Utilizing an institutional database of prenatally diagnosed infants, chart review of medical records of both mother and infants was performed for 182 infants. Infants carried a prenatal diagnosis of CHD and were born between January 1, 2013 and December 31, 2016 at Advocate Children's Hospital. Exclusion criteria included intrauterine demise, twin gestation and inadequate documentation. All data were reported using descriptive statistics or frequencies based on the level of measurement and normality.

Results: Of the included deliveries (31% with genetic anomalies), 76.5% were born via NSD. Of the NSD group, 65% had CHD as the documented reason for NSD. For IOL, 52.6% were done for the sole indication of CHD. 25.8% were for combined OB and CHD reasons, and 21.6% had a sole OB indication. Of mothers presenting in SL, 42.8% delivered via CS, while 32% of IOL mothers went to CS.

Infants born via SL, including those scheduled for an induction and born prematurely, were smaller than those born via IOL (mean birth weight 2.81 kg vs 3.07 kg, $P = .014$) and born earlier (mean gestational age 37 3/7 weeks vs 38 2/7 weeks, $P = .006$).

There was no difference between total length of stay (LOS) between SL and IOL for mother or infants, but a greater portion of the mother's hospital stay was before delivery with IOL vs SL (median 15.3 hours vs 5.3 hours.

There was no significant difference in the severity of CHD among the different modes of delivery.

Conclusions: : Among infants with CHD, NSD, including IOL, was pursued for CHD reasons more than OB reasons.

Failure to identify a primary care provider is common among adults with congenital heart disease

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Introduction: For adults with congenital heart disease, significant attention has been placed on facilitating the transition of cardiac care from pediatric to adult congenital cardiologists. However, little attention has been focused on the transition of primary care services from pediatric to adult providers in these patients. We aimed to describe the frequency with which adults with congenital heart disease identify a primary care provider and to highlight patient factors that are associated with the successful identification of a provider.

Methods: We retrospectively identified all patients >18 yo who were seen by adult congenital providers at our clinic between August 2016 and November 2016. At our center, all patients are asked to provide the name of their primary care provider at each visit. Patient records were reviewed. The identification of a primary care provider was noted and demographic, clinical, anatomic, and surgical details were extracted. Regression analysis was used to identify associations between patient variables and the successful identification of a primary care provider.

Results: The retrospective analysis included 198 patients with a median age of 33 yo (18-75 yo). Of the cohort, 13.8% reported not having health insurance, 75.7% had undergone a prior cardiac operation, 44% had conotruncal heart disease and 8.5% had single ventricle heart disease. 35.3% of patients did not identify a primary care provider. Patients with insurance more frequently reported having a primary care provider compared to those without insurance (67% vs 44%, $P = .02$). Patients with any current tobacco or alcohol use less commonly identified a primary care provider compared to those with no current use, nearly reaching statistical significance (41% vs 76%, $P = .09$). Those with diabetes (100% vs 63%, $P = .09$) and trisomy 21 (100% vs 63%, $P = .05$) more commonly reported having a primary care doctor, both nearly reaching statistical significance. Severity of congenital heart disease, number of prior cardiac operations, and presence of heart failure symptoms were not associated with the identification of a primary care provider. On multivariate regression analysis, patients with insurance were more likely to report having a primary care doctor, nearly statistically significant ($P = .07$).

Conclusions: A significant portion of adults with congenital heart disease at our center did not report having a primary care physician. Patients without insurance and with any current tobacco or alcohol use may be less likely to receive ongoing primary care. Adult congenital cardiologists should actively encourage their patients to establish care with primary care physicians and further study is required to elucidate barriers to the transition of pediatric to adult primary care in this population.

Guided question tool improves transparency and engagement between parents and providers

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Purpose: National news coverage of poor outcomes at several hospitals has highlighted the importance of patients and families having access to outcomes data. To empower parents of children diagnosed with congenital heart disease seeking information about outcomes, and in hopes of facilitating relationship building and transparency

TABLE 1 GQT survey results

Questions	Definitely % (n)	Somewhat % (n)	Slightly % (n)	Not at all % (n)
Parental Responses				
Communication				
Is the GQT a good tool to improve your confidence	71% (22)	29% (9)	0% (0)	0 (0%)
Were the GQT questions easy to understand	84% (26)	16% (5)	0% (0)	0 (0%)
Did your provider take the time to help you understand the GQT questions better	71% (22)	16% (5)	10% (3)	3% (1)
Transparency				
Is the GQT helpful in guiding you towards asking the right questions	76% (22)	24% (7)	0% (0)	0% (0)
After talking through the GQT with my provider I felt included in the conversation	97% (30)	3% (1)	0% (0)	0% (0)
I felt that I contributed to the plan	77% (23)	20% (6)	3% (1)	0% (0)
The GQT improved communication between my family & our care team	53% (16)	43% (13)	0% (0)	3% (1)
Provider Responses				
Confidence				
What is your level of comfort in answering the GQT questions	78% (7)	22% (2)	0% (0)	0% (0)
Did the GQT increase your own level of knowledge about your center	44% (4)	33% (3)	22% (2)	0% (0)
Transparency				
Did the GQT improve communication of outcomes with families?	44% (4)	44% (4)	11% (1)	0% (0)
Did the GQT improve family engagement during visits?	33% (3)	55% (5)	11% (1)	0% (0)
Provider Support				
Did the GQT increase the duration of your visits with families?	33% (3)	44% (4)	0% (0)	22% (2)

*denominators were variable due to omitted responses

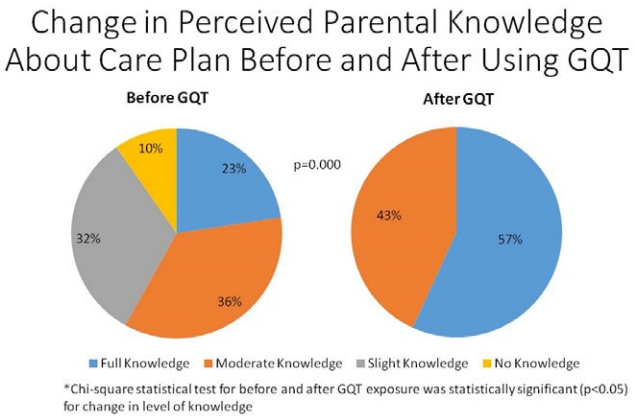

FIGURE 1

between parents and their providers, a list of 17 questions called the "Guided Questions Tool" (GQT) was created by the Pediatric Congenital Heart Association, a national patient advocacy organization. The purpose of this study was to assess parental and provider impressions of the impact of providing the GQT during prenatal counseling on fostering transparency and communication about center and disease specific outcomes for planned interventions. The purpose of this study was to assess parental and provider impressions of how the GQT may foster transparency and communication during prenatal counseling regarding center and patient specific outcomes.

Methods: Between October 2016-February 2018, four academic medical centers collaborated to provide the GQT in each of their fetal cardiology clinics. This 24-question survey was distributed using surveymonkey.com to families after they had an opportunity to review the GQT and had been counseled by the fetal cardiology team. A separate survey was sent to providers at the same centers after counseling families who had used the GQT. The four centers met for monthly conference calls, sharing several iterations aimed at improving their process of employing the GQT and collecting survey data.

Results: Surveys were completed by 31 parents and nine providers. Most parents completed the survey at their second visit (52%, n = 16). Detailed survey results are provided in Table 1 and Figure 1. When dichotomizing Likert scale responses into positive (definitely/somewhat) and negative (slightly/not at all), 100% of parents reported that questions were easy to understand and that the GQT positively impacted their confidence and their ability to know the right questions to ask. The GQT had a positive impact on communication for 97% of families. Additionally, 81% of parents reported that the GQT helped them think of their own questions to ask. Seven of the nine providers (78%) reported that employing the GQT resulted in them "definitely" or "somewhat" increasing their own knowledge of their center's outcomes. All providers reported having access to their center's outcome data related to GQT questions. Despite the increase in visit duration, 100% of parents and providers recommend the GQT in this setting.

Conclusion: Parents report that the GQT is easy to understand and improves their confidence in discussing outcomes with providers. Both parents and providers report that the GQT improves communication during prenatal counseling. Dissemination of the GQT to other cardiac centers may further enhance information exchange between parents and providers.

ICU outcomes for cone reconstruction in children with Ebstein anomaly

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Purpose: Children with depressed right ventricular (RV) function undergoing cone reconstruction (CR) for Ebstein anomaly (EA) are at risk for postoperative morbidity and mortality. Previous studies have demonstrated excellent short and medium-term outcomes, but the immediate postoperative ICU course and management has not been well studied. We examined the postoperative course for children following CR to identify risk factors for early postoperative morbidity and mortality.

Methods: This is a single institution, retrospective review of 167 children, 1-18 years old that had CR for EA between June 1, 2007 and June 20, 2017. Preoperative transthoracic echocardiography demonstrated tricuspid valve regurgitation ≥ moderate in 163 patients (98%) and, of these, 142 were severe (85%). RV function was severely decreased in 53 patients (32%). Baseline oxygen saturation was < 90% in 24 patients (14%). Patients undergoing tricuspid valve replacement for EA were excluded. We reviewed electronic medical records and collected demographic, imaging, and clinical data for all patients. Data were analyzed using student *t* test for mean values and chi squared analysis of qualitative data.

Results: All children (mean age 9.1 ± 4.9 years; 54% male) had CR of the tricuspid valve. Concomitant procedures included complete (n = 89) or subtotal atrial septal defect closure (n = 15), bidirectional cavopulmonary shunt (n = 40), plication of atrialized RV (n = 94), and right reduction atrioplasty (n = 140). Mean cardiopulmonary bypass time was 98 ± 26 minutes and mean aortic cross-clamp time was 76 ± 21 minutes. Mean ICU length of stay (LOS) was 5.1 ± 4.5 days, mean hospital LOS was 7.9 ± 4.6 days, and mean duration of mechanical ventilation was 1.4 ± 2.4 days. Average peak serum lactate was 6.7 ± 3.4 mmol/L and peak serum creatinine was 0.8 ± 0.3 mg/dL. In the first 24 hours postoperatively, 166 patients (99%) were on vasoactive-inotropic medications (mean vasoactive-inotropic score (VIS) 15.6 ± 6.8). Early postoperative morbidity included reoperation for valve re-repair (n = 6, 4%), cardiac arrest (n = 3, 2%), ECMO (n = 5, 3%), stroke (n = 2, 1%) and delayed sternal closure (n = 20, 12%). There was no early mortality and there was no heart block or renal dialysis in any child. Severe RV dysfunction on preoperative echocardiography was associated with postoperative need for ECMO.

Conclusion: CR for EA can be performed with low early mortality. Severe RV dysfunction portends a more difficult postoperative course including longer ICU LOS, increased need for postoperative ECMO support, and longer duration of mechanical ventilation. Children with EA should be considered for CR prior to development of severe RV dysfunction, even in the absence of symptoms.

Impact of NT-proBNP on clinical outcomes in children with systemic inflammatory response syndrome

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Objective: To evaluate the impact of elevated N-terminal pro-brain type natriuretic peptide (NT-proBNP) levels on clinical outcomes in children that met criteria for systemic inflammatory response syndrome (SIRS).

Background: NT-proBNP has traditionally been used in adults and children to diagnose and guide the treatment of congestive heart failure. Elevated NT-proBNP levels have been associated with increased morbidity and mortality in adults with septic shock. The association between NT-proBNP levels and morbidity/mortality in children with SIRS has not been studied.

Methods: This was a retrospective study. Patient list was generated utilizing ICD-9 and ICD-10 codes of various infectious diagnoses from Jan 2011 to Oct 2017. The inclusion criteria were children (<18 years of age) who presented with suspected infection, met the SIRS criteria and had a NT-proBNP level obtained. Exclusion criteria were final primary diagnoses related to cardiac disease and preexisting congenital or acquired heart disease. Patients were divided into 2 groups based on elevation of NT-proBNP levels. Variables were compared between 2 groups using chi-square or nonparametric tests.

Results: A total of 4 004 patients were screened and our final cohort consisted of 44 patients. Primary diagnoses were bacterial infection (n = 35, 80%), viral infection (n = 6, 14%), and Kawasaki disease (n = 3, 7%). Between elevated (n = 26) and nonelevated (n = 18) NT-proBNP groups, there was no difference in age groups, gender, race or final diagnoses. Median NT-proBNP was significantly higher in the elevated group (7500 vs 316 pg/mL, $P < .001$). Elevated group had higher median C-reactive protein level (208 vs 70 mg/L, $P = .008$) and lower median left ventricular ejection fraction (59% vs 69%, $P = .002$). Children in the elevated group were more likely to require intensive care unit admission (odds ratio 20.0 [95% confidence interval (CI): 2.2 to 181]), mechanical ventilation (OR 5.0 [1.2 to 21.5]), and inotropic support (OR 9.4 [2.2 to 41.5]). One patient died in the nonelevated group and four patients died in the elevated group ($P = .312$).

Conclusion: Severely elevated NT-proBNP levels were associated with increased morbidity in children with SIRS.

Incidence of acute kidney injury following cardiac catheterization in early postoperative period in children with congenital heart disease

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Background: Cardiac catheterization (CC) is commonly performed following surgery for congenital heart defects (CHD) for diagnosis

TABLE 1 Incidence of AKI using pRIFLE Classification

Number (percentage) of patients	Post Operative day 0	Post Operative day 1	Post Operative day 2	Post Operative day 3	Post cath day 0	Post cath day 1	Post cath day 2	Post cath day 3	Pre discharge
	N=39	N=42	N=42	N=39	N=42	N=37	N=36	N=36	N=40
No kidney Injury	22 (56.4%)	16 (38.1%)	18 (42.9%)	16 (41%)	38 (90.5%)	32 (86.5%)	29 (80.6%)	32 (88.9%)	36 (90%)
Risk	9 (23.1%)	15 (35.7%)	13 (31%)	12 (30.8%)	3 (7.1%)	4 (10.8%)	6 (16.7%)	3 (8.3%)	2 (5%)
Injury	8 (20.5%)	10 (23.8%)	9 (21.4%)	7 (17.9%)	1 (2.4%)	1 (2.7%)	1 (2.8%)	1 (2.8%)	1 (2.5%)
Failure	0 (0%)	1 (2.4%)	2 (4.8%)	4 (10.3%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	1 (2.5%)

TABLE 2 Comparison of AKI vs No AKI following cardiac catheterization in postoperative patients

Characteristic	AKI (n= 8)	No AKI (n= 34)	P value
Gestational age (weeks)	37.57 (1.13%)	37.4 (±2.11)	0.402
Race: African-American White Others	 4 (50%) 4 (50%) 0 (0%)	 15 (44.1%) 12 (35.2%) 7 (20.5 %)	 0.724
Male gender	4 (50%)	21 (61.7%)	0.411
Age at CHD surgery	66.6 (±87.29)	40.7 (±70.73)	0.137
Weight (kg)	22.95 (±20.71)	15.95 (±23.19)	0.819
Body surface area	0.762 (± 0.551)	0.553 (± 0.554)	0.614
Pre-Op Creatinine	0.50 (±0.23)	0.47 (± 0.21)	0.600
Pre-Op Creatinine clearance	105.59 (±33.60)	89.75 (± 42.99)	0.110
CPB time	261 (± 187)	244.27 (±103)	0.156
Aortic Cross-clamp time	137.63 (±107.78)	85.5 (±54.80)	0.002
Days between surgery and cath	6.38 (±5.04)	5.76 (±4.5)	0.756
CPB time	261 (± 187)	244.27 (±103)	0.156
Cath Intervention	5 (62.5%)	21(61.76%)	0.649
Dose of contrast	32.34 (±39.43)	28.44 (±31.26)	0.245
Nephrotoxic medications	5 (62.5%)	11 (32.3%)	0.121
Vasopressors	8 (100%)	31 (91.1%)	0.521
Intubation	8 (100%)	30 (88.23%)	0.414
ECMO	2 (25%)	9 (26.4%)	0.655
Length of ICU stay	38.5 (±29.31)	63 (±55.7)	0.097
Died	1(12.5%)	9 (26.4%)	0.374

and intervention of residual lesions. Use of contrast during CC increases risk for acute kidney injury (AKI), although data are lacking. Our purpose was to evaluate the incidence and risk factors of AKI in pediatric patients who underwent CC within 14 days after CHD surgery with cardiopulmonary bypass (CPB).

Methods: This was a retrospective chart review of all children CHD surgery with CPB at a single Children's Hospital. Patients known to have renal failure were excluded. Schwartz formula was used to calculate creatinine clearance for the first 72 hours following surgery and 72 hours following CC. pRIFLE criteria was used to categorize AKI as risk, injury and failure. Data were analyzed using SPSS software (Version 22, SPSS Inc. Chicago, Illinois) and expressed as mean ± SD and numbers (percentage).

Results: Of 43 included patients, 85% were born full term; 44.2% were African American and 39.5% were Caucasian; 22 (51.2%) had prior heart surgeries. At the time of CC, 40 (93%) patients remained on vasopressors and 11 (25.6%) were on ECMO. CC involved interventional procedures in 26 (60.5%) patients including balloon angioplasty in 13 (50%) and pulmonary artery stent placement in 12 (46.1%). Potentially nephrotoxic medications (aminoglycosides and vancomycin) were administered to 16 (38%) patients during the postoperative or post-CC period. The incidence of risk, injury and

failure according to pRIFLE criteria in the postoperative and post-CC period are shown in (Table 1 on page 53). Incidence of risk and injury peaked on the day following surgery. AKI following CC was lower at all time points, compared to the early postoperative period. Risk category of AKI was the commonest, with incidence of 7.1% on the CC day, a peak of 16.7% on post-CC day-2 and rapid decrease thereafter. None of the patients had renal failure post CC and none required renal replacement therapy. At discharge, 36 (90%) patients had no AKI, 5% had risk and 2.5% each had kidney injury and failure. There were no significant differences between patients with (n = 8) and without (n = 34) AKI at any time within 72 hours of CC considering differences in age at time of surgery, gender, weight, body surface area, CPB time, aortic cross clamp time, days between surgery and CC and length of ICU stay (Table 2 on page 53).

Conclusions: In pediatric patients, who are undergoing CHD surgery with CPB followed by CC within 14 days, contrast administration did not appear to be an additional risk factor for the development of AKI. AKI was relatively rare, mild and did not affect length of ICU or overall hospital stay. Further larger studies are needed to explore AKI in children with CHD.

Novel mitral inflow doppler derived index of LV compliance: Does it predict diastolic dysfunction

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Background: Active myocardial relaxation and compliance of left ventricle (LV) affects the appearance of early diastolic velocity wave (E wave) in the mitral inflow Doppler signal. The drop in inflow velocity after reaching the peak of E wave (decelerating phase) is a result of increasing LV diastolic pressure which is a function of its compliance. Diastolic indices described in current literature do not directly measure LV compliance, which is defined as a change in volume per unit change in pressure. The velocity-time integral of the decelerating phase (VTI_{Edecl}) of the E wave (a surrogate for change in volume of LV) divided by the square of peak E velocity (a surrogate for change in pressure for the corresponding change in volume) or VTI_{Edecel}/E^2 closely represents the LV compliance. The aim of this study was to test the ability of this index in predicting diastolic dysfunction in children.

Methods: A retrospective analysis of pediatric patients with diastolic dysfunction (indexed left atrial volume > 30 mL/m^{2}or LV end-diastolic pressure/LVEDP or pulmonary capillary wedge pressure/PCWP >/= 12 mm Hg) and controls (normal echocardiogram) was performed. Patients with left to right shunting lesions and complex congenital heart disease were excluded. Using postprocessing software (TomTec) the $VTI_{Edecel}/E^2$was compared between all cases and controls, as well as between cases with elevated LVEDP/PCWP and controls. The medians were compared using Wilcoxon test (JMP software). Receiver operating characteristics (ROC) analysis was performed to establish the appropriate cutoffs.

Results: There were 26 cases and 50 controls in the study. Among cases, 18 had their LVEDP/PCWP measured. Median VTI_{Edecel}/E^2 was lower in cases compared to controls (9.3 vs 12.1, P = .0004), and even lower among cases with elevated LVEDP or PCWP (8.3 vs 12.1, P = .0001) as shown in Table 1. The cutoff VTI_{Edecel}/E^2 separated controls from all cases with 88% sensitivity and 58% specificity, while the value < 9.56 separated them from cases with elevated LVEDP/PCWP with 76% sensitivity and 82% specificity (Figure 1).

FIGURE 1 Box plot demonstrating cutoff VTIEdecel/E2 that separates cases from controls and cases with elevated LVEDP/PCWP from controls and a diagram demonstrating the technique of measurement of VTIEdecel/E2

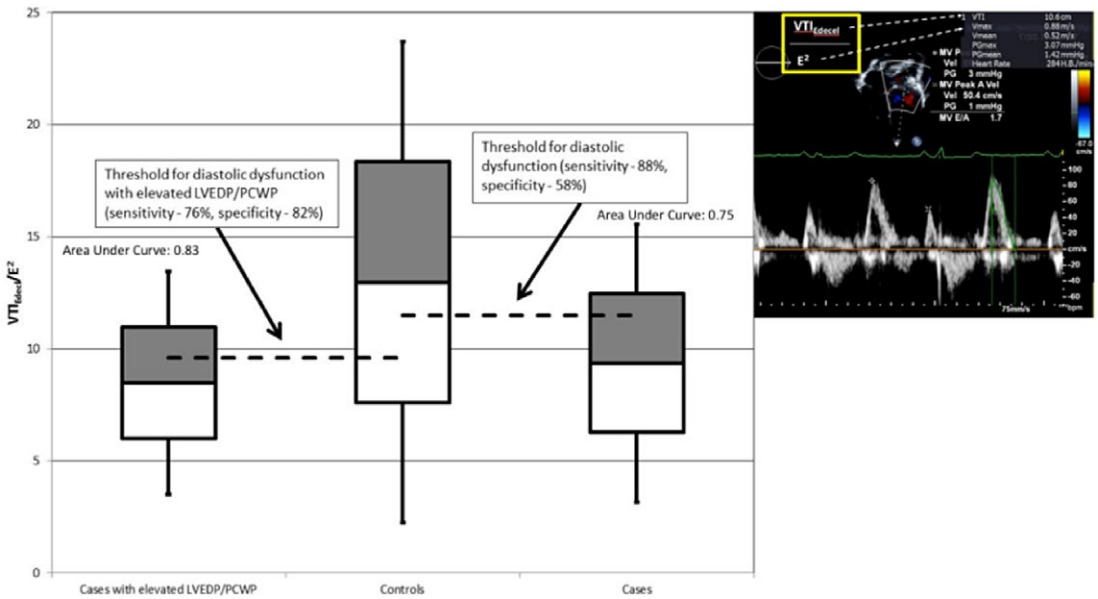

TABLE 1 Comparison of VTIE/E2 between all cases and age-matched controls, and between patients with elevated filling pressures by cardiac catheterization and controls

	Control N = 50 52% male		All cases N = 26 57.8% male		Elevated LVEDP or PCWP N = 18 66.7% male	
	Mean +/- SD	Median (IQR)	Mean +/- SD p value	Median (IQR) p value	Mean +/- SD p value	Median (IQR) p value
Age	11.2 years +/- 5.9	13 years (IQR 1.75 – 17)	11 years +/- 7.4 p value = 0.925	9.4 years (7.1 – 11.1) p value 0.671	9.7 years +/- 8.1 p value = 0.501	11 years (1 – 15.5) p value = 0.343
VTI_E/E^2	12.9 +/- 5.4	12.1 (10.1 – 13.7)	9.4 +/- 3.1	9.3 (IQR 7.0 – 11.1) **p value =0.0004**	7.9 +/- 2.1	8.3 (6.6 – 9.8) **p value = 0.0001**

Conclusion: VTI_{Edecel}/E^2 provides a specific assessment of LV compliance and can be used to predict diastolic dysfunction among children. VTI_{Edecel}/E^2 < 11.46 was quite sensitive in predicting diastolic dysfunction while VTI_E/E^2 < 9.56 had better specificity in predicting diastolic dysfunction with elevated filling pressures.

Pathologic vs innocent murmur education utilizing novel murmur auscultation mobile application

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Background: The prevalence and severity of congenital heart disease necessitates that pediatric trainees obtain competence in recognizing pathologic murmurs for appropriate referral. Current educational strategies to develop auscultation skills are outdated and time-consuming. Newer technology, such as mobile applications, offers the ability to design training platforms according to advanced adult learning principles.

Objective: To improve murmur education and auscultation training for pediatric trainees using new app-based technology.

Methods: This is a single center, prospective study. Participants were randomly assigned via block method to be in the innovation tech (App Group) vs standard education (Control Group). Audiologic murmur pretest with 10 questions was administered to all participants via the eMurmur University mobile app. In addition, all participants received a 40 minute didactic learning session describing all murmurs that would be ultimately tested. Murmur sounds from the lecture were provided via the mobile application so participants could get familiar with the technology. Following didactic lecture, the App Group received instructions on a sister application, eMurmur Primer, which includes descriptions of heart sounds, visualization of phonograms, and murmur audio clips with self-testing modules. The App Group was able to demonstrate the ability to work the work through the application modules with minimal instruction. In addition, they were instructed to interact with the application 15 minutes each day for a total of 4 weeks to apply cognitive load theory in the education model.

Results: There were a total of 35 participants including 8 medical students, 24 pediatric residents, and 3 chief residents. The pretest demonstrated that medical students, residents, and chief residents were able to identify benign vs pathologic murmurs correctly 68%%, 78%, and 70% of the time, respectively. Additionally, the pretest demonstrated medical students, pediatric residents, and chief residents incorrectly labeled benign murmurs as pathologic 63%, 64%, and 58% of the time, respectively. User experience results and follow up App Group results are underway. Next steps include evaluation of the App Group with a follow up audio-based murmur test

FIGURE 1 eMurmurPrimer Smartphone Application This is a snapshot from the smartphone application demonstrating the self testing ability. The user has the ability to both see and hear the murmur and then test their knowledge of pathologic vs innocent.

via the eMurmur University application 4 weeks after the pretest. Trainees will again be asked to identify innocent vs pathologic murmurs by listening to audio clips via the mobile app. The mean average of scores for the pretest and posttest will be compared using student's *t* test. The posttest will also include anchored response scales to assess the usefulness of the application, how often trainees interact with the application, and their confidence in pathologic vs innocent murmurs.

Conclusion: The initial phase of this study demonstrates that pediatric trainees need to improve their ability to identify pathologic vs innocent murmurs. Novel auscultation tools using mobile application technology have many advantages over current lecture-based curriculum and need to be considered in building future education platforms.

Peer mentorship program provides coping mechanisms for mothers with a fetal diagnosis of congenital heart disease

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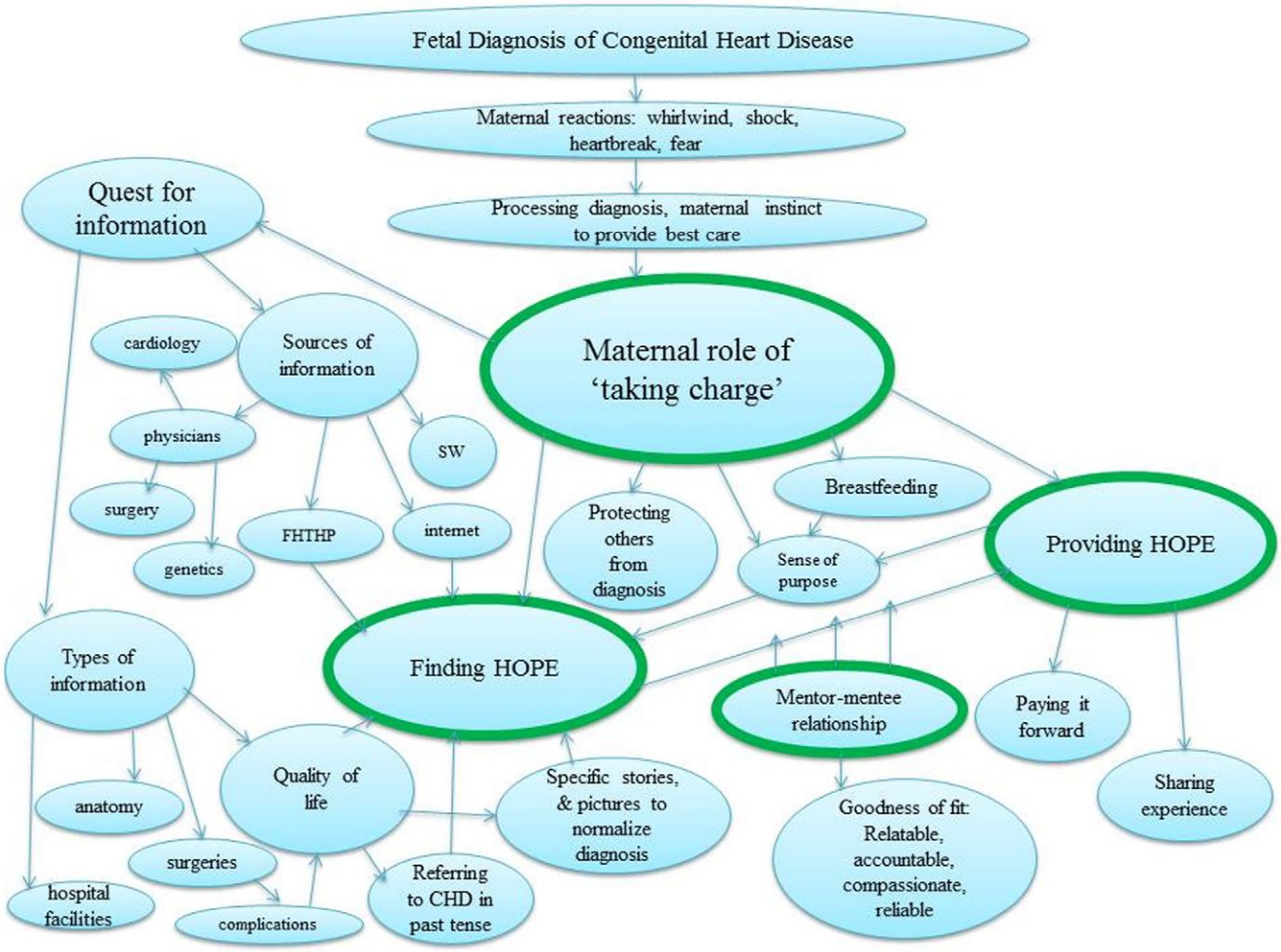
Purpose: Prenatal diagnosis of congenital heart disease (CHD) is associated with elevated maternal stress and anxiety. At our Fetal Heart Center, we have a structured peer support program for mothers with a fetal diagnosis of CHD called the Fetal Heart to Heart Program (FHTHP). Past studies have reported the maternal reaction to a fetal diagnosis of CHD, but this is the first study to evaluate the impact of a peer mentorship program on maternal coping mechanisms following fetal diagnosis of CHD. Our study explores the maternal reaction to fetal diagnosis of CHD, maternal support system, and experiences with the FHTHP during the prenatal and postnatal period.

Methods: Using a qualitative phenomenological design we conducted one-on-one semi-structured phone interviews with mothers who received a fetal diagnosis of CHD. Mothers were at least 2 months removed from their child's last intervention. Data was analyzed using steps described by Colaizzi. Descriptive interview data was coded and analyzed using an inductive thematic approach. Sample size was determined by informational redundancy.

Results: Nine mothers with a fetal diagnosis of CHD were interviewed. The journey of a mother receiving a fetal diagnosis of CHD

Themes in mothers' journey after fetal diagnosis of congenital heart disease



Concept map of themes in mothers' journey after a fetal diagnosis of congenital heart disease

is comprised of 4 general phases: 1) receiving the fetal diagnosis: heartbreak, shock and fear of the unknown, 2) delivery: gaining control, 3) surgical intervention, and 4) hope for the future: reflecting and paying it forward. The constant theme that evolved throughout pregnancy was of the maternal role in "taking charge." This influenced subthemes of "finding hope" and "providing hope" (see Figure 1). Mothers experienced acute stress symptoms after receiving the fetal diagnosis of CHD and this ignited the mothers' role to "take charge" and started their quest for information. The mentor-mentee relationship is a dynamic process throughout the mothers' journey that augments the relationship between "finding hope" and "providing hope." Mothers found hope through the FHTHP by stories of shared experiences with their mentors. Postnatally, breast-feeding provided mothers with a source of control, a sense of purpose, and allowed mothers to normalize the experiences of their child. The time before surgery was the second most stressful time during the mothers' journey. Mothers used support from the FHTHP and the stories of their mentors to find hope and practical advice. Once removed from surgery, mothers reflected on their graciousness of the experience and were motivated to pay it forward and share their stories with other mothers in similar situations.

Conclusion: This is the first study to elucidate the maternal journey following fetal diagnosis of CHD from prenatal to postnatal period in continuum. The benefits of shared experiences through a peer mentoring support group after a fetal diagnosis of CHD serve to alleviate maternal stress by providing hope and parental control both prenatally and postnatally.

Prevalence of innocent murmurs in pediatric patients

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Introduction: Evaluation for a cardiac murmur is one of the most common reasons for cardiology referral in pediatric patients. Between 30% and 75% of those murmurs are found to be innocent, with Still's murmurs being the most frequently diagnosed. These murmurs can often come and go throughout childhood, either with normal growth, or during times of physiologic stress. While it is known that innocent murmurs are commonly found in patients between the ages of 0 and 18 years, the exact prevalence of each of these murmurs is unknown.

Purpose: This is an ongoing study, the purpose of which is to determine the prevalence of each type of innocent murmur (Still's murmur, carotid bruit, pulmonary ejection murmur, peripheral pulmonary murmur, and venous hum).

Methods: Preliminary data for this study was collected for a total of 6159 participants ages 0 to 18 years old who presented for initial evaluation at 10 different outpatient pediatric cardiology clinics between January 1, 2017 and January 1, 2018. As this is an ongoing study, data verification is still in process. All new patients presenting to outpatient cardiology clinic participate in normal history and physical exams (including a regular cardiac exam), as is standard for new-patient visits. Checkboxes for each innocent murmur were incorporated into the physical exam templates utilized at the outpatient clinics. Electronic medical records for study participants are expected to be queried over a two-year period to classify each of the five innocent murmurs by age, gender, race, and concurrent occurrence of congenital heart disease.

Results: Of the collected records, 3967 have been verified, 2025 (51.04%) of which were found to have one of the five innocent murmurs. Females comprised 45.88% of those found to have murmurs (929 patients) and males comprised 54.12% (1096 patients). 297 of the verified patients with murmurs were further classified by murmur type: Still's (163 patients, 54.88%), pulmonary ejection (79 patients, 26.60%), peripheral pulmonary (29 patients, 9.76%), venous hum (26 patients, 8.75%), and carotid bruit (no patients). The average ages ranged from 4.04 years old (venous hum) to 6.49 years old (pulmonary ejection murmur).

Conclusion: This is a continuing analysis with initial data showing that innocent murmurs are highly prevalent in the pediatric population, Still's murmurs and pulmonary ejection murmurs being the most

TABLE 1 Summary of preliminary demographic data characterized by type of innocent murmur

	Carotid Bruit	Pulmonary Ejection Murmur	Still's Murmur	Venous Hum	Peripheral Pulmonary
Average Age	N/A	6.49	5.6	4.04	N/A
Gender					
Female	0	46	79	9	13
Male	0	33	84	17	16
% Female	N/A	58.23%	48.47%	34.62%	44.83%
% Male	N/A	41.77%	51.53%	65.38%	55.17%
Ethnicity					
American Indian or Alaska Native	0	1	1	0	0
Asian	0	2	0	2	1
Black or African American	0	16	64	6	8
Declined	0	2	9	2	0
Native Hawaiian or Other Pacific Islander	0	0	0	0	1
Unknown	0	0	0	0	1
White	0	58	85	16	17
Other	0	0	4	0	1
Total Number of Patients	0	79	163	26	29
% of Total	0.00%	26.60%	54.88%	8.75%	9.76%

commonly found innocent murmurs. Preliminary data also shows a slight male predominance in the prevalence of innocent murmurs. As we continue to collect, review, and verify the data, we aim to further characterize these murmurs according to race and association with concurrent occurrence of congenital heart disease. We hope to delineate additional demographic patterns so as to better understand the occurrence of innocent murmurs in the pediatric population.

Quality metric implementation and management utilizing electronic medical records

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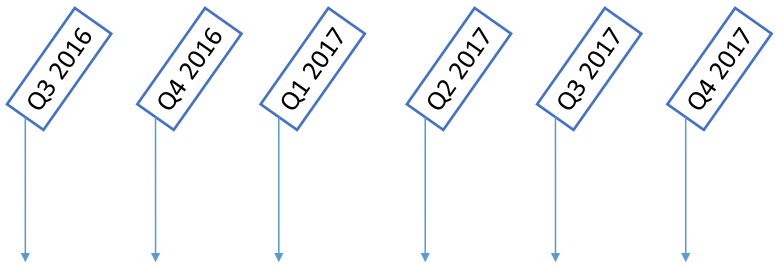
Purpose: Our aim was to expand Quality Metric (QM) documentation and data collection while creating a platform to facilitate reporting for the Mednax Cardiology Quality Collaborative (MCQC). Goals included optimizing our existing EMR tools to capture denominator and numerator elements while leveraging a work flow conducive to enhance outcomes. Long-term goals included: (1) 100% documentation within the EMR platform, (2) 100% semi-automated reporting of QM's (3) QM compliance within parameters set by the American College of Cardiology (ACC) Adult Congenital and Pediatric Cardiology (ACPC) section Quality Network (ACPC QNet) collaborative.

Methods: Templates for EMR enhancements extended from published metrics developed by ACPC QNet. QM additions to EMR occurred in phases. An evaluation tool for selection of metrics was created (Figure 1). Bi-annual MCQC quality summits scored and selected metrics. The development specification team included medical, programing and project experts. Bimonthly workgroup meetings were scheduled through the development process to assure compliance with metric specifications and to facilitate coordination with provider work flow. The team established an implementation

FIGURE 1

MCQC Metric Prioritization Matrix

	Assigned Weight	ACPC QNet Metric 02	ACPC QNet Metric 03	ACPC QNet Metric 13	ACPC QNet Metric 15	ACPC QNet Metric 16
Clinical Interest	25%	A				
Past Performance	25%	B				
Other Agency Interest	10%	C				
IT Data Extraction	25%	D				
FE Impact (Medical and IT)	15%	E				
Total/Composite Score		**				
Legend : rate each metric from 1-5; with 1 = low impact on domain and 5= highest impact on domain						
Total/Composite Score = ** = .25(A)+.25(B)+.10(C)+.25(D)+.25(E)						



ACPC Qnet Metric 02. Proportion of patients, 3-18 years with a BMI greater than 85% who received appropriate counseling.

	Basic EMR Reporting		EMR "hard stop"enhancement		
No Data	0.33	0.25	0.23	0.46	0.61

MCQC QNet Practices	2	2	3	4	7	10
MCQC EMR QNet QM2 Submissions	0	1	2	3	4	7

FIGURE 2

schedule. EMR platform modifications utilized in-house developers. EMR enhancements were released quarterly. Reporting tools were developed: (1) using EMR prepackaged reporting, (2) leveraging EMR reporting tools to create external reports, and (3) developing simple "denominator" reports.

Results: By June 2016, the MCQC had introduced, measured and reported on several QMs. Initial metrics used to evaluate processes included ACPC QNet QM3 (documentation of BMI), and ACPC QNet QM2 (counseling for BMI >85%). By the end of 2017, 4 additional metrics completed development. Early documentation and collection of data included manual chart review—scanning for free text entries. Developing or utilizing EMR fields for discrete data improved collection of data. "Hard" stops lead to initial improvement in compliance with metrics (Figure 2) but with significant provider frustration. "Softer" clinical decision reminders resulted in consistent improvement in QM documentation/compliance. Reporting of total population was accomplished utilizing discrete fields in EMR and with output tools that included numerators and denominators from the target population. Denominator reports allowed for random sampling reports and increased the QMs submitted to ACPC QNet.

Conclusion: EMR is a powerful tool to capture and report quality metric data. Several lessons were learned through our experience: (1) EMR development requires specifications under constant review and revision with frequent stakeholder input; (2) provider work flow changes can effect data input and disruption of workflow can lead to work-arounds that interfere with QM compliance or reporting ability; (3) total population reports vs random sampling both appear to accurately reflect compliance with QMs; and (4) simple tools for data entry, collection and reporting improve compliance. Future considerations: QM enhancement evaluation tool to assess effects on patient care, provider work flow, and value of QM once measured.

Domains were developed and weighted for members of MCQC collaborative to score metrics. Scores were compiled to establish a metric development timeline.

Right ventricular systolic pressure prediction by baseline EKG analysis

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Purpose: Electrocardiography is a routine noninvasive method utilized to asses cardiac rhythm, chamber size as well as extra cardiac pathology. The science and methodology which underlies electrocardiography predates most other screening and diagnostic modalities currently used in cardiology practice.

The amplitude of the R wave in lead V1 has been postulated to predict the RVSP. This study aims to determine if RVSP can be reliably predicted utilizing the EKG [Formula A: RVSP = 3(R in V1) + 47; Formula B: RVSP = 5(R in V1)] when compared to direct measurement during cardiac catheterization (cath).

Methods: This is a single center IRB approved retrospective chart review of patients aged 0-25 years who underwent percutaneous hemodynamic assessment between 2016 and 2018. The study was approved by our institutional review board. EKGs performed within 6 months of the patients' catheterization procedure were analyzed, and Formula A and B as shown above were applied. Patients with ventricular inversion were excluded. The degree of correlation between the predicted and measured RVSP was assessed using the Pearson's correlation. The ability of each formula to detect right ventricular hypertension was assessed by determining their sensitivity and specificity.

Results: Sixty-four patients (33 male, 31 female) undergoing cath during the study period met inclusion criteria. A total of 92 EKGs were analyzed. Mean age at catheterization was 5.4 years (ranging 0-23 years). The RVSP as predicted by Formula A and B were weakly positively correlated with the Gold Standard cath RVSP ($r = 0.41$, $P < .05$). Formula A detected right ventricular hypertension with 100% sensitivity. Formula B detected right ventricular hypertension with 74% sensitivity and 67% specificity.

Conclusion: The amplitude of the R wave in V1 correlates with right ventricular systolic pressure. While the electrocardiogram is a highly sensitive test for the diagnosis of right ventricular hypertension, it carries low specificity. Clinicians therefore should continue to interpret electrocardiograms in the context of the patient history, physical exam and other noninvasive testing, in order to determine whether invasive assessment of right ventricular pressure is indicated.

Risk stratification of patients with hypoplastic left heart syndrome and intact atrial septum using fetal MRI and echocardiography

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Purpose: Despite continued improvements in survival for patients with hypoplastic left heart syndrome (HLHS), for the 5-10% of patients with an intact or restrictive atrial septum (HLHS/IAS), morbidity and mortality remain high. The purpose of this retrospective study is to add our center's experience with diagnostic and management strategies for patients with HLH/IAS and to describe our experience with fetal pulmonary vein Doppler (PVD) flow patterns and fetal MRIs as predictors for morbidity and mortality.

Methods: Infants born with HLHS between January 1, 2009 and October 6, 2017 were identified through surgical and echocardiographic databases. The foramen ovale was considered intact if there was no visible defect in the atrial septum by two-dimensional fetal echocardiography and no color flow across the atrial septum using Doppler color flow mapping. It was considered restrictive if the infant required left atrial decompression within the first 24 hours of life.

Results: Eighty-two infants were diagnosed with HLHS, of which twelve (14%) met criteria for HLHS/IAS. All were diagnosed prenatally. Four patients underwent EXIT procedure and atrial septal

Comparing postnatal outcomes of fetal pulmonary venous Doppler patterns

Fetal PVD Pattern	Type B (N=5)	Type C (N=7)
Diagnosis		
AA-MA	0 (0%)	5 (71%)
AS-MS	5 (100%)	2 (29%)
Fetal MRI	1 (33%)	3 (42%)
Evidence of Lymphangectasia	0 (0%)	2 (29%)
Outcome		
Survival to Discharge	4 (80%)	3 (42%)
Need for ECMO	1 (20%)	2 (29%)
Length of Stay (days)	67.8 ± 41.3	106 ± 58.8
Length of ICU Stay (days)	41.4 ± 48.6	62.3 ± 23.1
Survival to Stage 2	4 (80%)	2 (29%)

Abbreviations: PVD, pulmonary venous Doppler; AA-MA, aortic atresia-mitral atresia; AS-MS, aortic stenosis-mitral stenosis.

stent placement; five underwent transvenous balloon septostomy, and one underwent transvenous atrial septal stent. Despite an intact septum, one patient had a decompressing vein and did not require intervention until stage 1 palliation. Survival to discharge and to stage 2 palliation were better for patients with HLHS compared to those with HLHS/IAS (83% and 69% vs 58% and 41%). For patients with HLHS/IAS, those with a type C fetal PVD pattern had longer hospital and intensive care unit stays and worse survival compared to those with type B fetal PVD pattern (Table 1). In addition, both patients with evidence of lymphangectasia on fetal MRI had a type C PVD pattern. Because of the prolonged hospital course and poor prognosis for the first patient with a type C pattern and lymphangectasia on fetal MRI, the second was offered and chose comfort care.

Conclusions: Despite prenatal recognition and immediate surgical and catheter-based interventions postnatally, the mortality for patients with HLH/IAS remain significantly higher than those with an adequate atrial septal defect. As other studies have suggested, the subset of HLH/IAS with fetal type C PVD pattern portend an even worse prognosis, and additional findings of lymphangectasia on prenatal MRI may suggest irreversible pulmonary vascular changes and near certain mortality. As these modalities may allow for more realistic prenatal counseling and better risk stratification to identify patients in whom medical intervention may be futile, prospective studies to validate this are warranted.

Single left ventricular function following placement of modified blalock-taussig shunt—A speckle tracking echocardiographic study

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Background: Coronary artery abnormalities are common in patients with pulmonary atresia with intact ventricular septum (PA/IVS). There is a paucity of data on the effect of modified Blalock–Taussig shunt (mBT) on the single left ventricle (LV) function in PA/IVS. Given the coronary abnormalities, we hypothesized that there will be deterioration of single LV function in patients with PA/IVS compared to patients with tricuspid atresia (TA) following placement of mBT shunt.

Methods: This observational study included patients with single ventricular physiology (PA/ IVS and TA) who underwent mBT shunt placement for securing pulmonary blood flow. Patients who had arch reconstruction, required ECMO, or had pericardial effusion were excluded. Demographic data and echocardiograms were reviewed. All echocardiograms were performed using Philips iE33 and interpreted by a single reader blinded to clinical data. Global systolic endocardial and myocardial longitudinal strain (endoGLS, myoGLS) and average of peak longitudinal strain of 6 LV segments (average endoLS) from the apical 4-chamber view were analyzed offline using vendor-independent software (Tomtec Inc). Between-group comparisons were done using student *t* test and chi-square test as appropriate (SPSS ver. 22).

Results: Our cohort consisted of 31 patients of whom 21 (68%) patients had PA/IVS and 10 (32%) had TA; 15 (48.4%) were males. The PA/IVS subgroup was younger at the time of mBT shunt placement compared to TA (2.4 ± 3.3 vs 5.6 ± 4.2 weeks, P = .05). The mean endoGLS was decreased after mBT shunt placement in patients with PA/IVS compared to patients with tricuspid atresia (−16.18 ± 5.27 vs −20.10 ± 4.97, P = .05). The average endoLS measured from 6 segments of the LV was also decreased. The myoGLS, LV ejection fraction, and the LV end systolic, diastolic and stroke volumes before and after placement of mBT shunt were similar in both the groups (Table 1).

Conclusion: Despite similar LV function at baseline between the two groups with single ventricle physiology, LV function revealed significant abnormalities in the PA/IVS group compared to the TA group, after placement of mBT shunt. The subtle impairment in LV function shown by speckle tracking echocardiography could plausibly be related to coronary artery abnormalities in PA/IVS patients. Long term studies are needed to assess if these difference in the LV function persist and contribute to adverse clinical outcomes.

TABLE 1 Comparison of demographic and echocardiographic parameters

Parameter (mean ± SD), n (%)	PA/IVS (n=21)	TA (n=10)	p-value
Age at mBT shunt (weeks)	2.4±3.3	5.6±4.2	0.05
Males (n %)	10 (47%)	5(50%)	0.5
Duration from surgery (median, range)	8 (3-16)	5 (2-14)	0.4
Pre-Shunt Echo			
Pre-shunt end diastolic volume (ml)	8.85± 3.47	10.34± 5.29	0.36
Pre-shunt end systolic volume (ml)	3.62±1.93	4.67±2.8	0.24
Pre-shunt stroke volume (ml)	5.23±1.92	5.67±3.4	0.14
Pre- shunt ejection fraction (%)	60.65± 9.38	54.45± 12.74	0.14
Pre-shunt endo GLS %	-18.88± 5.5	-20.51± 5.75	0.47
Pre- shunt Myo GLS %	-15.98± 5.21	-16.68± 5.25	0.74
Pre-shunt average endo LS %	-16.66± 5.51	-18.95± 5.92	0.32
Post Shunt Echo			
Post- shunt end diastolic volume (ml)	9.73± 2.98	11.04± 4.82	0.35
Post-shunt end systolic volume (ml)	4.22±1.67	4.59±2.03	0.59
Post-shunt stroke volume (ml)	5.51±2.1	6.45±3.13	0.33
Post-shunt ejection fraction (%)	56.25± 10.43	57.42± 8.11	0.7
Post shunt Endo GLS %	**-16.18± 5.27**	**-20.10± 4.97**	**0.05***
Post-shunt Myo GLS %	-13.1± 5.1	-16.4± 3.97	0.08
Post-shunt average Endo LS %	**-14.83± 4.67**	**-18.63± 4.89**	**0.05***

The evolution of risk adjustment in pediatric and congenital cardiac surgery

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Purpose: The analysis of pediatric and congenital cardiac surgical outcomes requires adjustment for case mix. We reviewed three categories of pediatric and congenital cardiac surgical risk adjustment tools

1. Procedural Risk Stratification tools based primarily on subjective probability (expert opinion)
2. Procedural Risk Stratification tools based primarily on objective multi-institutional clinical data
3. Risk models that adjust for procedural factors and patient factors using logistic regression modelling.

Methods: Risk stratification is a method of analysis in which the data are divided into relatively homogeneous groups (called strata). Data are analyzed and reported within each stratum. The Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) is the largest database in the world that contains information about pediatric and congenital cardiac operations. STS-CHSD has utilized risk adjustment tools in all three of the above categories. STS-CHSD has used 2 risk stratification tools developed based primarily on subjective probability

1. Aristotle Basic Complexity Levels (**ABC Levels**)
2. Risk Adjustment for Congenital Heart Surgery-1 Categories (**RACHS-1 Categories**)

The Society of Thoracic Surgeons—European Association for Cardio-Thoracic Surgery Congenital Heart Surgery Mortality Categories (**STAT Mortality Categories**) are an empirically derived methodology of risk stratification based on the statistical estimation of the procedural risk of mortality, from an analysis of objective data (77 294 operations).

The 2014 **STS-CHSD Mortality Risk Model** uses Bayesian hierarchical modelling to adjust for the risk associated with all of the procedural factors and patient factors listed in Table 1.

An **augmented STS-CHSD Mortality Risk Model** was developed in 2017. The 2014 STS-CHSD Mortality Risk Model adjusts for many patient factors, including the binary presence or absence of a chromosomal abnormality, syndrome, and/or noncardiac congenital anatomic abnormality. The augmented STS-CHSD Mortality Risk Model further refines case mix adjustment by adding more granular adjustment for individual chromosomal abnormalities, syndromes, and/or noncardiac congenital anatomic abnormalities.

TABLE 1 Variables (procedural factors and patient factors) in the STS-CHSD mortality risk model

Variable
Age [a]
Primary procedure [b]
Weight (neonates and infants)
Prior cardiothoracic operation
Any non-cardiac congenital anatomic abnormality (except 'Other noncardiac congenital abnormality' with code value = 990)
Any chromosomal abnormality or syndrome (except 'Other chromosomal abnormality' with code value = 310 and except 'Other syndromic abnormality' with code value = 510)
Prematurity (neonates and infants)
Preoperative Factors
• Preoperative/Preprocedural mechanical circulatory support (IABP, VAD, ECMO, or CPS) [c]
• Shock, Persistent at time of surgery
• Mechanical ventilation to treat cardiorespiratory failure
• Renal failure requiring dialysis and/or Renal dysfunction
• Preoperative neurological deficit
• Any other preoperative factor (except 'Other preoperative factors' with code value = 777) [d]

[a]Modeled as a piecewise linear function with separate intercepts and slopes for each STS-defined age group (neonate, infant, child, adult). [b]The model adjusts for each combination of primary procedure and age group. Coefficients obtained via shrinkage estimation with The Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery (STS-EACTS [STAT]) Mortality Category [6] as an auxiliary variable. [c]Abbreviations: CPS, cardiopulmonary support; ECMO, extracorporeal membrane oxygenation; IABP, intraaortic balloon pump; VAD, ventricular assist device. [d]Any other preoperative factor is defined as any of the other specified preoperative factors contained in the list of preoperative factors in the data collection form of the STS Congenital Heart Surgery Database, exclusive of 777 = "Other preoperative factors."

TABLE 2 Pediatric and congenital cardiac surgical risk adjustment tools

Table 2. Pediatric and Congenital Cardiac Surgical Risk Adjustment Tools			
Tool	Year in STS-CHSD	C-Statistic	Time Interval of Data to obtain C-Statistic
Aristotle	2002	0.687	January 1, 2007 – December 31, 2008
RACHS-1	2006	0.745	January 1, 2007 – December 31, 2008
STAT	2010	0.778	January 1, 2007 – December 31, 2008
STS-CHSD Mortality Risk Model	2015	0.872	January 1, 2010 – December 31, 2015
Augmented STS-CHSD Mortality Risk Model	2019	0.875	January 1, 2010 – December 31, 2015

We reviewed the discrimination of each of these 5 tools by comparing the published C-statistic (also known as the area under the receiver operating characteristics curve) of each tool.

Results: Table 2 documents the C-statistics that have been reported on each of the 5 tools described above.

Conclusion: From 2002 to 2018, pediatric and congenital cardiac surgical risk adjustment tools have demonstrated increased ability to discriminate performance.

The natural history and diagnosis of coarctation requiring surgical intervention

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Purpose: Coarctation of the aorta is a common congenital heart condition, affecting 4 per 10 000 live births. Despite continuing advances in early diagnosis of congenital heart disease, including prenatal ultrasound and postnatal universal pulse oximetry screening, the timely diagnosis of coarctation remains challenging. Even when suspected, a definitive diagnosis of coarctation may be difficult to confirm in the immediate newborn period. Our aim was to describe the current natural history of coarctation requiring surgical intervention.

Methods: Children from 0 to 18 years of age who underwent surgical repair for coarctation between January 1, 2012 and December 31, 2017 were included in this study. We performed a retrospective chart review of their course to diagnosis and eventual surgical repair, including prenatal imaging, pulse oximetry screening, and postnatal

echocardiograms. Children were classified based on the time of initial concern; fetal, neonatal, or late diagnosis. Late diagnosis was defined as clinical suspicion raised after discharge from the hospital following birth. We also reviewed these patients' associated cardiac and noncardiac diagnoses.

Results: A total of 62 children (69% male) were included. In this cohort, 15 (24%) were identified prenatally, 20 (32%) diagnosed while still hospitalized following birth, and 27 (44%) were late diagnoses of coarctation. Of the 33 children with documented pulse oximetry screening, 24 passed (73%). 45% required more than one postnatal echocardiogram before a definitive diagnosis of coarctation was confirmed (Table 1). 15 (24%) had surgical repair at less than one week of age, 22 (35%) between 7 and 30 days, 13 (21%) between 1 month and 1 year, and 12 (19%) between 1 and 11 years. 42 children were repaired with an end-to-end anastomosis, 18 underwent arch reconstruction, 1 required an interposition graft, and 1 attempted repair by interventional catheterization followed by end-to-end anastomosis.

Of these patients, 27 (44%) had bicuspid aortic valves, 11 (18%) had ventricular septal defects, 7 (11%) had persisting left-sided superior vena cavae, 3 (5%) had atrioventricular canal defects, and 2 (3%) had transposition of the great arteries. Significant noncardiac disease was present in 16% of patients (10/62), including 4 children with trisomy 21, 3 with multiple congenital anomalies, and 1 each with VACTERL, Kabuki syndrome, and Turner syndromes. There were 2 postsurgical deaths, 1 attributed to SIDS and the other secondary to noncardiac complications of the child's underlying genetic syndrome.

Conclusion: Early diagnosis of coarctation remains a significant challenge despite improved fetal imaging and universal congenital heart disease screening.

Ventricular arterial coupling and left ventricular global longitudinal strain: Does race play a role in children?

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Background: Among adults, the incidence and prognosis of congestive heart failure (CHF) is worse in African Americans, compared to other races. Also, in adults, black race is associated with abnormal global longitudinal strain (GLS), a marker of subclinical left ventricle (LV) dysfunction. LV interaction with arterial afterload is considered to be a key component of optimal cardiovascular efficiency. Data on the impact of race on GLS and ventricular arterial coupling (VAC) in the pediatric population is scarce. We hypothesized that there are racial differences in VAC and LV GLS in the pediatric population.

Methods: This was a retrospective review of patients (≤18 yrs) who presented to a cardiology clinic for evaluation of chest pain and syncope and had normal echocardiograms (ECHOs) and electrocardiograms. Patients who were hypertensive, overweight and obese were excluded. Demographics, ECHO data, and mean blood pressure (MBP) was collected. ECHOs (Philips IE33) were read by a single reader blinded to demographic data. Full volume data consisted of adjacent subvolumes over four consecutive beats acquired during breath-hold to minimize artifact and was analyzed offline (Tomtec Inc software). Using 3D analysis, LV endiastolic volume (EDV), LV endsystolic volume (ESV) and stroke volume (SV) were measured. These volumes were indexed to body surface area. Arterial elastance (Ea) and ventricular elastance (Ees) were calculated as Mean BP/SVi and MBP/ ESVi respectively. VAC, the ratio of Ea/Ees, was calculated as ESVi/SVi. Global longitudinal strain (GLS) was also assessed for all patients. Statistical analysis included student *t* test and chi-square test to compare groups. *P* value < .05 was considered significant.

TABLE 1 Comparison of demographics, arterial elastance and LV functional parameters

Parameters Mean ± SD or n(%)	Caucasian (n=34)	African American (n=40)	P value
Age (years)	12.2±4.2	12.1±3.6	0.861
Height (cm)	151.3±25.4	153.1± 18.8	0.728
Weight (kg)	49.7± 22.1	48.6 ± 19.5	0.829
Body surface area (m^2)	1.4 ± 0.4	1.4 ± 0.3	0.996
Gender (male)	21 (62%)	24 (60%)	0.984
Systolic BP (mmHg)	**116 ± 11.4**	**109 ± 12.1**	**0.026**
Diastolic BP (mmHg)	62 ± 7	60 ± 8.3	0.242
Indexed ESV (ml/m^2)	19.1 ± 4.4	19.5 ± 6.8	0.789
Indexed EDV (ml/m^2)	55.5 ± 10.8	55 ± 13.9	0.854
Indexed SV (ml/m^2)	36.3 ± 7.6	35.4 ± 8.7	0.639
Arterial elastance Ea (mm Hg m^2/ml)	2.3 ± 0.5	2.3 ± 0.6	0.939
Ventricular elastance Ees (mm Hg m^2/ml)	4.4 ± 1.1	4.3 ± 1.3	0.769
VAC	0.54 ± 0.11	0.56 ± 0.15	0.523
GLS (%)	-22.8 ± 2.9	-22.6 ± 3.5	0.815
Longitudinal strain global average (%)	-22.2 ± 3	-22.2 ± 3.5	0.949

Results: : Our cohort consisted of 74 patients, 34 (46%) of whom were Caucasian and 40 (54%) were African American. The two groups were comparable in age, height, weight and body surface area. The systolic BP was found to be significantly higher in the Caucasian population. There were no significant differences in the ESVi, SVi, Ea, Ees and VAC ratio between the two groups. There was also no significant difference in the GLS between the two groups (Table 1).

Conclusions: Our exploratory data does not reveal any difference between African Americans and Caucasians in VAC or LV function in the pediatric age group. These results suggest that vascular changes influencing ventricular remodeling and LV function in the African American population probably occur later in life. Our results support the need for further studies evaluating these parameters in young adults. These investigations are critical to delineate the onset of racial differences in VAC and LV systolic function and better prevent and manage CHF.

Coronary artery compression during intervention on dysfunctional right ventricular outflow tracts

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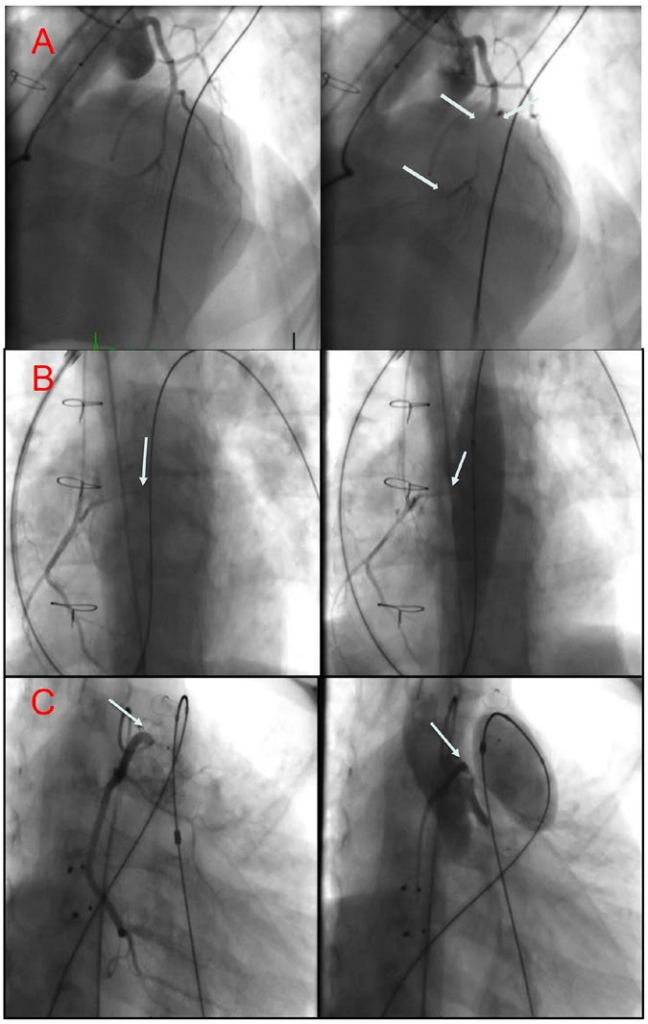
Background: Interventions on dysfunctional right ventricular outflow tracts (RVOT) with stent implantation and/or percutaneous pulmonary valve implantation (PPVI) can cause coronary artery compression (CAC). Coronary angiography during simultaneous test balloon dilation of the RVOT is recommended to diagnose CAC prior to stent placement or PPVI. This study aimed to evaluate a single-institution experience with CAC during RVOT interventions.

Methods: This was a retrospective review of all patients referred for intervention on a dysfunctional RVOT (obstruction and/or regurgitation), who had simultaneous balloon dilation and coronary angiography between 8/2010 and 6/2015. Coronary anatomy was assessed by angiography and reported as normal or abnormal. Variable means were compared with *t* test, and frequencies with odds ratio (OR).



Results: A total of 110 patients were referred for RVOT intervention. The most common diagnoses were tetralogy of Fallot/pulmonary atresia (76%), truncus arteriosus (8%), and aortic stenosis with Ross procedure (8%). Seventy-four patients (67%) had PPVI, 7 (6%) had conduit stent implantation alone, and 29% had balloon dilation (27%). Fifteen of the 110 patients (14%) had evidence of CAC during

TABLE Comparison of collected data for patients with CAC (n = 15) to those without (n = 95).

Variables	+CAC (n = 15)	-CAC (n = 95)	p-value
Age (years)	16.2 ± 9.3	16.8 ± 8.2	0.8
Female sex	47%	42%	0.7
Weight (kg)	52.0 ± 19.9	54.3 ± 23.8	0.7
Homograft size (mm)	18.7 ± 3.1	19.7 ± 4.1	0.4
Successful PPVI	20%	74%	<0.001
Abnormal coronary anatomy	27%	6%	**0.02**

balloon dilation of the conduit. There were no significant differences between those with CAC and those without, other than incidence of abnormal coronary arrangement (Table). Patients with CAC were more likely to have an abnormal coronary arrangement (27% vs. 6%, OR 5.4, CI 1.3-22.1, *P* = .02).

Five patients proceeded with PPVI or RVOT stent despite CAC seen on angiography. Of these, 3 had dynamic systolic CAC remote from the conduit due to high right ventricular pressure, which resolved with relief of conduit obstruction in all 3 (Figure 1A). The other 2 had true compression of a small conal branch from right coronary artery by the conduit, which was intentionally sacrificed to achieve improved RVOT function (Figure 1B). Both of these patients are doing well clinically without RV dysfunction or arrhythmias.

Ten patients did not have PPVI or stent placement due to CAC (6 right main, 4 left anterior descending, and 1 conal branch, Figure 1C). Ventricular tachycardia occurred in 1 patient with test balloon dilation. No other complications occurred.

Conclusions: Patients with compression of a conal branch with negligible myocardial distribution can be considered for intervention. Dynamic systolic compression due to right ventricular hypertension should be evaluated carefully and does not constitute a contraindication, as this resolves with relief of cond uit obstruction. Patients with known coronary artery abnormalities may be more likely to have major CAC, and should be counseled on the increased degree of difficulty.