

Sun 1

Playing Catch Up – A Longitudinal Study Of Growth Velocity In Patients With Single Ventricle Palliation

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Background: Growth impairment is a common problem in children with congenital heart disease, including those with single ventricle palliation. Although there has been much emphasis on interstage monitoring between the stage I and Glenn operations, the growth patterns beyond Glenn long-term have not been well studied. The purpose of this study was to assess the pattern of growth between the Glenn and Fontan operations, and beyond.

Methods: Single-center retrospective study of patients who underwent Fontan operation between 2010-2020.

Results: 189 patients (112 males) underwent Glenn operation at a median (IQR) age of 5.5 months (1.7), weight of 6.4 kg (1.5) and height of 64 cm (6.5). Mean (SD) height z-scores were -1.19 (1.55), -0.79 (1.32), and -0.60 (1.59) at the time of Glenn, Fontan, and last follow-up, respectively. Mean (SD) weight z-scores were 1.32 (1.36), -0.60 (1.24), and -0.26 (1.39) at the time of Glenn, Fontan, and last follow-up, respectively. Median follow-up time was 8.5 years (6.6). There was a statistically significant improvement in growth velocities from the time of Glenn to Fontan (height $p=0.0005$, weight $p<0.0001$), and to last follow-up (height $p<0.001$, weight $p<0.0001$). However, the median height and weight for age z-scores stayed below 0 and at last follow up were -0.61 (2.05) and -0.12 (1.90), respectively. Significant growth impairment (respective z-scores <-1.5) persisted at last follow-up in 36/189 (19%) of patients for weight-for-age and in 51/189 (27 %) for height-for-age. Regression analysis identified height and weight at Fontan as risk factors for continued growth impairment at last follow-up.

Conclusions: Patients undergoing staged single ventricle palliation show improvement in growth from the time of their Glenn operation, however, not reaching that of the normal population. Linear growth, a better marker of growth in this age group, was affected more than weight. Early complete assessment of growth, nutritional interventions, and early endocrine evaluation may be beneficial to improve these growth parameters, especially as growth adequacy is linked with quality of life.

Sun 2

Effects Of Different Tidal Volume Ventilation Strategies On Fontan Flow And Hemodynamics: A Prospective Randomized Study.

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Background: In patients with Fontan circulation pulmonary blood flow (PBF) is passive. Published studies of the Fontan circulation show that negative pressure or spontaneous ventilation increases PBF and cardiac output (CO), whereas positive pressure ventilation (PPV) decreases PBF and CO. Many Fontan patients require PPV for procedures but the best PPV strategy to optimize PBF and CO has not been studied. The aim of this prospective randomized 2- treatment cross-over study is to compare the effects of low tidal volume (TV) ventilation (5mls/kg body weight) vs. high TV ventilation (10mls/kg) during PPV on PBF and CO.

Methods: Fontan patients were randomized to Group 1 (high TV ventilation followed by low TV) or Group 2 (low TV ventilation followed by high TV). All patients received baseline ventilation (7mls/kg) for 5 minutes followed by the first ventilation condition (high or low TV). After the first ventilation condition all patients received a 5-minute washout period of a second baseline ventilation before crossover into the second ventilation condition. After 5 minutes steady state in each ventilation setting an arterial blood gas was drawn, hemodynamic parameters recorded, and the transesophageal echocardiogram (TEE) performed. Primary outcome was the impact of the ventilation strategy on the transpulmonary gradient (TPG) = Fontan pressure - common atrial pressure. Secondary outcomes include the effect of the ventilation strategy on PBF, and CO defined as systemic ventricular outflow tract velocity time integral (OT-VTI) and inferior vena cava flow (IVC-vel) measured by TEE.

Results: Ten patients were included in the final data analysis. The median age was 5 years (interquartile range (IQR) 4,11) with a median weight of 16.3kg (IQR 13.8,31.6). Low tidal volume ventilation resulted in significantly lower peak (15.3 +/- 2.9 vs 22.2 +/- 3.7 mmHg; $p < 0.001$) and mean airway pressures (7.3 +/- 0.8 vs 8.7 +/- 0.9 mmHg; $p = 0.001$) compared to high tidal volume ventilation. However, TPG, OT-VTI and IVC-vel were not significantly different between low and high TV ventilation.

Conclusions: Despite a significant difference in the peak and mean airway pressure, using 10mls/kg or 5mls/kg TV during PPV in patients with Fontan circulation is not associated with differences in PBF and CO provided minute volume, inspiratory oxygen fraction and arterial blood gas parameters, remain the same.

Sun 3

Navigating Uncharted Waters: A Case Report Describing The Rapid Launch Of A New Dialysis Modality Urgently Needed By A Patient With Hypoplastic Left Heart Syndrome

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Background: A series of complications during treatment of renal failure in a patient with hypoplastic left heart syndrome (HLHS) necessitated the urgent launch of a new dialysis modality in the pediatric cardiovascular intensive care unit (CICU). This case report describes the unique circumstances that led to the launch of Aquadex ultrafiltration therapy in the CICU, reports pertinent patient outcomes, and details the intrahospital collaboration, shared clinical decision-making, and required nursing resources employed in its successful initiation.

Methods: Patient is a 2-month-old male admitted to the CICU at birth due to HLHS with mitral stenosis and aortic atresia. Pertinent history includes Stage I reconstruction and placement of Sano conduit on day-of-life (DOL) 4, followed by extracorporeal membrane oxygenation (ECMO) from DOL 4-13. Following ECMO, peritoneal dialysis (PD) was initiated due to renal failure and intolerance to renal replacement therapy (RRT) using Baxter's Prismaflex system and a 7 French hemodialysis catheter. After six weeks of PD, necrotizing enterocolitis developed twice, prompting the decision to transition to continuous veno-venous hemofiltration (CVVH) using the Nuwellis Aquadex system in October 2022.

Results: Although Aquadex had been implemented in the neonatal/infant intensive care unit (N/IICU) previously, Prismaflex was the CICU's only RRT delivery system. Aquadex's benefits include low extracorporeal blood volume (35 milliliters) and ability to utilize double-lumen central venous access as small as size 6 French. Additionally, Aquadex circuit-to-circuit priming – standard of care at our hospital – reduces exposure to allogeneic blood products, which is a priority for pediatric patients who may need cardiac transplantation. Upon CICU rollout, Aquadex-experienced N/IICU registered nurses (RNs) immediately assumed care of the patient. But within 25 days, a team comprising N/IICU and CICU Clinical Nurse Specialists and Educational Nurse Specialists and a Dialysis Nurse Leader developed a training plan and provided education to 42 CICU RNs and 9 dialysis RNs with previous RRT knowledge, as well as additional stakeholders. RNs received 11 90-minute training sessions, with 30-minute didactic learning on care principles and 60-minute situational learning on troubleshooting alarms, augmenting settings, baselining hematocrit, trending pressures, obtaining labs, and reviewing history. Limiting learners to 2 per Aquadex console ensured a tactile and engaged learning environment. The newly trained CICU RNs then worked with Aquadex-experienced N/IICU or CICU RNs for 3 shifts to support clinical experience development.

Conclusions: Aquadex therapy for this patient remains an indefinite need. Future analyses will examine patient outcomes post-Aquadex treatment, including allogeneic blood product exposure, fluid balance, patient's time without RRT, and a full description of adverse events. Education and training evaluations by RN learners (n=16) show that the education met objectives (100%), was engaging (94%), and can be applied to the clinical environment (100%). Evaluations are continuing, and learners will also self-assess sustained knowledge and comfort in providing clinical care with Aquadex at 60-90 days post-initial training. Assessment of nursing resources since Aquadex rollout revealed 100% of N/IICU RN support within the first 25 days of therapy. However, within 40 days, CICU RNs provided 85% of all RN staffing needs required for patients receiving Aquadex in the CICU.

Sun 4

Size Of Right Ventricle To Pulmonary Artery Conduit Does Not Affect Outcomes After Norwood Palliation

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Background: Management of hypoplastic left heart syndrome has improved over the past two decades leading to improved survival into adulthood. Surgical modifications of the Norwood operation have been associated with improved outcomes. The single ventricle reconstruction trial did not show significant difference in outcomes for patients with right ventricular to PA (RV-PA) shunt compared to the Blalock-Thomas-Taussig (BTT) Shunt. Modifications to the RV-PA conduit have been described. The size of the conduit may improve the pulmonary artery growth and lead to lower risk of intervention in a small weight infant. We hypothesized a 6 mm conduit offers better oxygenation and hemodynamics in the period following stage palliation and better hemodynamics prior to stage 2 compared to smaller size conduit.

Methods: This was a retrospective single center study from 2015 to 2022. Neonates with hypoplastic left heart syndrome undergoing a Norwood operation in the first month of life were included. Patients were divided into 2 groups based on the RV-PA conduit size: 5 mm versus 6 mm. p, Patient with Sano size of 5 mm versus Sano size of 6 mm. Patients who had a BTT shunt, prematurity, preoperative extracorporeal membrane oxygenation (ECMO) or other cardiac lesions aside from HLHS were excluded. A total of 59 patients were included in the final analysis.

Results: Patients receiving a 6mm conduit were smaller compared to those receiving a 5 mm conduit (3.34 kg versus 2.72 kg, $p < 0.001$). There were no differences of timing to operation, incidence of aortic or mitral valve atresia or need for preoperative mechanical ventilation between the groups. There were no differences in immediate post-operative outcomes between the groups when comparing time with open sternum, need for postoperative ECMO or length of intubation. There was a trend towards improved survival in patients receiving a 6 mm conduit ($p = 0.09$). There was a trend towards shorter length of ICU stay in the group receiving a 6 mm conduit versus the 5 mm conduit (26.4 days versus 30.4 days, $p = 0.09$). There was no significant differences in branch pulmonary artery sizes or mean pulmonary artery pressures between the groups at catheterization prior to Glenn. Similarly, there was no significant difference between calculated pulmonary to systemic blood flow ($Q_p:Q_s$) between the groups (1.1 vs. 1.4, $p = 0.14$). Outcomes after Glenn revealed no significant differences in catheter interventions, need for post-operative ECMO or interstage death.

Conclusions: Our study examined differences between a 5 mm and 6 mm RV-PA conduit at Norwood palliation from birth through Glenn palliation. There were no significant differences in preoperative variables between the groups and immediate post operative outcomes were similar at the time of Norwood although there were trends towards shorter length of ICU stay and survival at the time of Norwood. Importantly, we found no differences in terms of pulmonary artery growth at the time of catheterization prior to Glenn. Additionally, there were no differences of after the Glenn between the groups in terms of ECMO, cardiac arrest or interstage deaths. More studies are needed to aid in decision making in regards to determination of optimal RV-PA conduit size at the time of Norwood.

Lymphatic Abnormalities On MRI In Single-Ventricle Congenital Heart Defects Prior To Glenn Operation.

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Background: Patients palliated with staged surgeries for single-ventricle heart defects are at times affected by lymphatic abnormalities and complications such as plastic bronchitis and protein-losing enteropathy. Abnormalities of lymphatic origin in the neck and thorax have been described after the Glenn- and prior to the Fontan operation and shown to correlate with surgical outcomes. It is unknown when these abnormalities occur and if they are caused by altered hemodynamics, inherent lymphatic disease or a combination. Our objective was to investigate if lymphatic abnormalities in the neck and thorax were present already prior to the Glenn-operation in children with single-ventricle heart defect.

Methods: Patients with single-ventricle heart defect and a T2-weighted MRI-scan prior to their Glenn-operation at The Children's Hospital of Philadelphia from 2012-2022 were included. The T2-weighted MRI-scans were reviewed and categorized according to type of lymphatic abnormality in the neck and thorax: type 1 (little or no signal in supraclavicular region) to type 4 (supraclavicular, mediastinal and pulmonary T2-signal). The distribution of lymphatic abnormalities prior to Glenn operation was reported. Comparison of groups including outcomes (Fontan takedown, heart transplant, plastic bronchitis, chylothorax, chylous pericardial effusion, mortality) were done using analysis of variance, the Kruskal-Wallis test and Fisher's exact test.

Results: Our study population included 71 children with single-ventricle physiology of whom 30 (42%) had hypoplastic left heart syndrome and 41 (58%) had non-hypoplastic left heart syndrome. We found lymphatic abnormalities in the neck and thorax already present prior to the Glenn-operation, with 14 (20%) patients having a type 4 abnormality, 15 (21%) a type 3 and 42 (59%) a type 1-2. A total of 39 (55%) patients presented with a genetic abnormality, although not necessarily linked to lymphatic disease. Patients with a type 3 or 4 lymphatic abnormality had a higher prevalence of pre-Glenn chylothorax vs type 1-2 (27% and 57% vs 0%, $p=0.01$), and fewer patients with a type 4 lymphatic abnormality underwent Glenn-operation ($p=0.01$). A total of four patients (6%) died prior to their Glenn-operation, three (21%) of these had type 4 lymphatic abnormalities ($p=0.04$). Overall combined mortality regardless of operations was significantly different between types 1-2 vs. type 3 ($p=0.04$) and vs type 4 ($p=0.03$). No patients had plastic bronchitis.

Conclusions: Lymphatic abnormalities, visualized by T2-weighted MRI, are present in the neck and thorax of some children with single-ventricle physiology already prior to their Glenn-operation. The prevalence of chylothorax and mortality was higher with increasing severity of lymphatic abnormality.

Sun 7

Right Ventricular Strain Is Reduced In Hypoplastic Left Heart Syndrome Non-Survivors

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Background: Cardiac dysfunction is associated with mortality in children with hypoplastic left heart syndrome (HLHS). Echocardiography is used to qualitatively assess cardiac function in systemic right ventricles (RV). Recently, RV systolic longitudinal strain (RVLS; includes septal motion assessment) and RV free wall strain (RVFWS) have been increasingly used to quantify RV systolic function. In structurally normal hearts, RVFWS is the preferred method to assess RV mechanics, but in single RVs the importance of septal dysfunction is not well described. We compared the differences in RVLS and RVFWS at various timepoints in HLHS survivors versus non-survivors during the interstage period.

Methods: In this retrospective, single-center study, echocardiograms from 3 timepoints (pre-Norwood, 4-8 weeks post-Norwood, and 8 weeks prior to Glenn) were analyzed in infants with classic HLHS. Patients were stratified into two groups based on outcome of transplant-free survival to Fontan completion (survivors) versus mortality or need for transplantation (non-survivors). Images were postprocessed to obtain RVLS and RVFWS. An equal variance t-test was used to determine significant differences and ROC curve analysis was performed to derive cutoff values for RVLS and RVFWS.

Results: Forty-one patients (17 survivors, 24 non-survivors) in the pre-Norwood stage, 32 patients (15 survivors, 17 non-survivors) in the post-Norwood stage, and 24 patients (15 survivors, 9 non-survivors) from the pre-Glenn stage were included for analysis. There was no significant difference in the RVLS or RVFWS measurements during the pre-Norwood (RVLS -16.5 vs. -16.4, $p = 0.96$; RVFWS -19.5 vs. -19.3, $p = 0.86$) or post-Norwood (RVLS -17.8 vs. -16.8, $p = 0.36$; RVFWS -19.5 and -20.7, $p = 0.52$) time points between the Fontan survivors and non-survivors, respectively. We found significantly higher RVLS and RVFWS in the pre-Glenn echocardiograms for survivors than non-survivors, respectively (RVLS -18.5 vs. -15.1, $p < .01$; RVFWS -21.7 vs. -15.8, $p < .01$). A pre-Glenn RVLS $> -16\%$ (AUC 0.82; $p = 0.01$) had 80% sensitivity and 77.8% specificity; and pre-Glenn RVFWS $> -18\%$ (AUC 0.85; $p = 0.005$) had a 93% sensitivity and 67% specificity for predicting death or need for transplantation.

Conclusions: Our results show that both RVLS and RVFWS are decreased immediately prior to Glenn palliation in the cohort of patients who died or required a transplant prior to Fontan completion. Either modality may be used for serial monitoring of systolic function in patients with HLHS. RVLS $> -16\%$ and RVFWS $> -18\%$ immediately prior to Glenn palliation may be used to identify higher risk patients.

Sun 8

Plant-Based Diet--A Missing Link To Optimize Cardiovascular Status In Fontan Physiology

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Background: Despite the use of medications and exercise training, patients with a Fontan circulation still have limited long-term survival. Nearly 40% of patients with Fontan Circulation are obese. The benefit of nutritional strategies such as a plant-based diet has not been investigated in patients with a Fontan circulation.

Methods: A 32-year-old obese female with tricuspid atresia and transposition of the great arteries {S,D,D}, pulmonary artery banding and atrial septectomy at 3 months of age, classic Fontan operation at 5 years of age, atrial tachycardia, and obesity presented to clinic with 1 year history of dyspnea on exertion, orthopnea, and weight gain. Physical exam was notable for bilateral lower extremity edema. The patient was diagnosed with Fontan failure with preserved ventricular systolic function and NYHA II symptoms. The patient was medically managed with sildenafil and furosemide. Lifestyle management was encouraged: decreasing processed foods and animal-based protein while transitioning towards a whole foods plant-based nutrition lifestyle, including whole grains, legumes, vegetables, seeds, nuts, and fruits with supplementation of vitamin B12. Additionally she was to incorporate 3x per week aerobic exercise.

Results: The patient reported improvement in her exercise capacity. She exhibited 19 kg weight loss. A cardiopulmonary exercise test assessed exercise capacity before and after lifestyle changes with improvement of VO₂max by 32%. Repeat catheterization did not demonstrate notable changes in hemodynamics. Repeat hepatic MRI with elastography showed improved hepatic stiffness of 3.5 kPascal. Echocardiogram showed severe RA dilation, normal left ventricular systolic function and no mitral regurgitation. Hepatic MRI with elastography showed hepatic compliance consistent with moderate fibrosis, measuring approximately 4.2 kPa. Catheterization demonstrated mean Fontan pressure of 16 mmHg, wedge mean pressure of 12 mmHg, CI of 2.0 L/min/m², and PVR of 1.1 WU.

Conclusions: Weight loss, improved exercise capacity, improved cardiac index, and hepatic stiffness after multi-modality treatment with medications, aerobic activity, and a plant-based diet was seen in this adult patient with Fontan circulation. A plant-based diet may help patients with a Fontan circulation by increasing endogenous NO and improving pulmonary endothelial cell function and, thus, lowering PVR. All aspects of the treatment regimen certainly contributed to the patient's improvement. However, to our knowledge, this is the first report of the use of a plant-based diet to help optimize cardiovascular status in an adult with a Fontan circulation. A plant-based diet may offer a novel therapeutic option for patients with Fontan circulation. Further study is needed.

Associations Between Exercise Capacity And Psychological Functioning In Youth With Fontan Circulation

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Background: Youth with single ventricle cardiac physiology palliated to Fontan circulation (FC) often have diminished exercise capacity. They are also at high risk for psychological problems, including anxiety, depression, and attention problems. Limited prior research has shown exercise capacity is associated with adaptive functioning (i.e., ability to perform basic daily living tasks) and attentional capacity, but not internalizing symptoms (e.g., depression, anxiety) in FC samples. However, internalizing symptoms have been associated with metrics of physical fitness and exercise capacity in other cardiac patient populations. The current study examines associations between exercise capacity and psychological functioning in a clinical sample of youth with FC.

Methods: A multidisciplinary medical team, including pediatric cardiology, immunology, endocrinology, gastroenterology, psychology, exercise physiology, nutrition, social work, and nursing evaluated participants in a dedicated clinic for youth with FC. As part of the evaluation, patients completed cardiopulmonary exercise testing using a standardized stationary bike protocol. Exercise capacity metrics included max VO₂ (ml/kg/min), max heart rate, oxygen saturation at max exercise (max O₂ sat), anaerobic threshold (AT), max work rate (WR [watts/kg]), and respiratory exchange ratio (RQ). Sample was limited to those with adequate effort (RQ > 1.1). Parent and child also completed a standardized psychological screening questionnaire (Behavioral Assessment System for Children, Third Edition [BASC-3]); scales selected for analyses were based on common concerns seen in youth with FC, including depression, anxiety, attention problems, and adaptive functioning. Clinical and demographic data were extracted from patient medical records.

Results: Clinical sample (n= 43) was 65% male. Racial identity was self-reported as 88% White and 12% "other." 12% reported Hispanic/Latino ethnicity. Majority (77%) had commercial insurance. Mean child age was 13.2 years (SD=2.3). The most common cardiac diagnosis was hypoplastic left heart syndrome (47%). For parent report scales on the psychological measure, child attention problems were negatively associated with WR ($r = -.384$, $p = .021$) and max VO₂ ($r = -.316$, $p = .047$); depression symptoms were negatively associated with max O₂ sat ($r = -.308$, $p = .048$). For child self-report, anxiety was inversely correlated with RQ ($r = -.396$, $p = .019$).

Conclusions: We found significant negative associations between metrics of exercise capacity and depression/anxiety symptoms in our modest sample of youth with FC. Although prior studies looking specifically at FC samples have found no associations with internalizing symptoms and exercise metrics, our findings are consistent with the broader literature on mental health symptoms and indicators of physical fitness. Anxiety could interfere with patient effort on the task as reflected in its association with RQ. Lower max O₂ sat may be an indication of overall poorer health, which could in turn contribute to depression symptoms. Generally, inattention can lead to problems with sustained focus on tasks, particularly for challenging activities, so these difficulties may have resulted in poorer exercise test performance. Explorations of potential mechanisms underlying these relations are warranted. Greater efforts are needed to include youth from diverse racial and socioeconomic backgrounds to increase representativeness of the sample and determine any impact of these factors on outcomes.

Sun 10

Exercise Capacity Remains Stable In Fontan Patients Entering Adulthood

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Background: Impaired exercise capacity is a frequent complication in Fontan patients and the functional capacity has been described to progressively decline with age. However, there are few longitudinal studies, and the pediatric age group dominates. We aimed to describe the natural course of exercise capacity in a population-based Fontan cohort, as these patients enter adulthood.

Methods: Two serial cardiorespiratory exercise tests (CPX) were performed in Danish Fontan patients. Thirty patients (15 males) reached a respiratory exchange rate of minimum 1.0 in both CPX-tests and were included in the analyses. The time interval between the two tests was ten years, with a mean age of 17 years in study 1 and 27 years in study 2. Peak oxygen consumption (VO₂) and oxygen pulse were measured. Results from the two assessments were compared for each patient using paired t-tests.

Results: 60% had a predominantly left ventricle and 40% a predominantly right ventricle. Types of total cavopulmonary connections included extracardiac conduit (47%), lateral tunnel (50%) and classical Fontan (3%). Mean age at Fontan completion was 5.8 years (range 1.5-25.7). There was no significant change in exercise capacity in these 30 patients during the study period. VO₂ remained stable with peak VO₂ 24.9 ml/min/kg (range 14.1 - 47.2) in study 1 and 23.9 ml/min/kg (range 11.4 - 41.1) in study 2 ($p = 0.34$), and mean percent predicted peak VO₂ at 67.2% (range 30.0-108.0) in study 1 and 68.5% (range 40.0 – 97.0) in study 2 ($p = 0.68$). Percent predicted O₂-pulse was also stable with 88.9% (range 31.5-157.0) in study 1 and 87.8% (range 52.3-155.4) in study 2 ($p = 0.81$).

Conclusions: Exercise capacity remained stable during transition from childhood to adulthood in this cohort of Danish Fontan patients.

Sun 11

Characteristics And Outcome Of Noonan Syndrome Patients With Single Ventricle Morphology And Concomitant Lymphatic Disorders

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Background: Noonan Syndrome (NS) is an autosomal dominant disorder and is the second largest syndromic contributor to congenital heart disease (CHD). In some cases, a single ventricular (SV) morphology is seen. Additionally, about 15-20% of NS patients also suffer from lymphatic abnormalities including lymphoedema, protein-losing enteropathy, chylothorax. This case series aims at describing the characteristics and outcome in a small cohort of single ventricle NS patients with lymphatic complications.

Methods: We describe a case series including 3 NS patients with SV morphology identified from the Children's Hospital of Philadelphia Lymphatic Center database. Clinical data and imaging from the database were reviewed both retrospectively and prospectively.

Results: In total, three NS patients with SV morphology were identified in the database. Baseline demographics and clinical data are summarized in Table 1. The 2 patients that underwent stage 2 palliation had type 4 abnormality on T2 imaging and did not survive. Patient 3 is scheduled for lymphatic imaging and is currently alive as a BTS although with pulmonary congestion. Both patients that had lymphatic imaging showed disorganized central conduction abnormality and bilateral pulmonary lymphatic perfusion syndrome (PLPS).

Conclusions: Noonan syndrome patients with a single ventricle morphology and lymphatic abnormalities comprises a small but challenging patient group. In this cohort advanced palliation beyond stage 1 led to significant respiratory issues and death. These results suggest that single ventricle palliation may not be a suitable treatment for Noonan patients with single ventricle hearts if they also have central lymphatic abnormalities. More experience is needed to determine the best course of treatment for this patient group.

Sun 12

Long-Term Outcomes After Fontan Operation In Pulmonary Atresia With Intact Ventricular Septum: Relevance Of RV-Dependent Coronary Artery Circulation

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Background: Pulmonary atresia with intact ventricular septum (PA/IVS) is a rare congenital heart disease (CHD) that may require palliation with Fontan circulation depending on the size of the right ventricle (RV) or presence of RV dependent coronary circulation (RVDCC). We sought to identify differences in outcomes in patients with PA/IVS with RVDCC versus those without RVDCC who had Fontan palliation at our institution.

Methods: This is an IRB approved, retrospective cohort study. We identified patients with PA/IVS from a Fontan database at Mayo Clinic including data from 1972 - 2022. Those with features of PA/IVS in association with other major CHD including heterotaxy syndromes and tricuspid atresia were excluded.

Results: A total of 61 patients with PA/IVS had Fontan completion at a median age of 5 years between February 1981 and June 2017. The predominant type of Fontan was an atrio-pulmonary connection (59%). The overall survival amongst the entire cohort with PA/IVS undergoing Fontan procedure was 67.2% (41/61 patients) with a median post-Fontan follow-up of 26 years (range 0 – 47 years). Most living patients were in adulthood (mean 33.6 years, SD 12.2, range 8-61). The 10-year, 20-year, and 30-year survival was superior in the PA/IVS cohort (85%, 77%, 54% respectively) in comparison with the prior published survival amongst the entire Fontan cohort at our institution (83%, 68%, 48% respectively). All 6 patients with RVDCC were alive at the time of the study, with two of these patients > 30 years from Fontan. There were no significant differences between patients with and without RVDCC in terms of long-term survival, left ventricular systolic function, NYHA class, left ventricular end diastolic pressure, pulmonary artery pressures, resting baseline ECG, or history of ventricular arrhythmias.

Conclusions: Overall long-term post Fontan survival among patients with PA/IVS was superior in comparison to the entire Fontan cohort. Although the study was limited by small sample size, there was no increased early or late mortality among patients with RVDCC vs those without RVDCC.

Neurological Events In Fontan Patients Undergoing Cardiac Transplantation

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Background: Improvements in the surgical and medical management of single ventricle patients requiring Fontan palliation have increased the 30-year survival to 85%. With failure of Fontan physiology, patients can be subjected to months, or even years, of low cardiac output. Dramatic increases in cardiac output following heart transplantation can result in hyper-perfusion syndrome and subsequent encephalopathy. Patients with Fontan palliations are at additional risk for abnormal neurological outcomes due to a prothrombic state, chronic micro emboli, and multiple palliations. We aim to describe the neurologic complications in the early perioperative period in patients with Fontan circulation who have undergone cardiac transplantation.

Methods: This is a retrospective, single-center study evaluating the perioperative neurological outcomes in patients with Fontan physiology who underwent heart transplantation at our institution from January 2006 to May 2021. Forty-six patients were included in the study with ages ranging from 4 years to 53 years. Twenty-eight patients were male while eighteen of the patients were female. Five points of evaluation were used including neurological signs and symptoms in the first month following transplant, neurological findings on CT and MRI, abnormalities on electroencephalography (EEG), the presence of a neurology consult, and the initiation of antiepileptic medications prior to discharge.

Results: Twelve of forty-six patients (26%) experienced perioperative neurological events. EEG changes were appreciated in seven patients (15%) with seizure activity in three. These three patients were discharged home on antiepileptic medications. Eleven of the forty-six patients (23%) had findings on head imaging including infarcts (n=4), acute subdural hematoma (n=4), and subarachnoid hemorrhage (n=2). One patient had both a subdural hematoma and a lacunar infarct. The overall survival to discharge in our cohort was 94% with none of the survivors having a long-term neurological disability. There was one death in the acute period, which was due to a large subarachnoid hemorrhage resulting in tonsillar herniation.

Conclusions: Despite recent medical and surgical advancements, patients with Fontan physiology are medically complex with a high incidence of perioperative neurological events after undergoing heart transplantation. This study demonstrates the need for close neurological monitoring in Fontan patients undergoing heart transplantation. Routine perioperative EEG monitoring should be considered in this unique patient population. With diligent monitoring, early detection, and intervention, our center demonstrated a survival of 94% with none of the survivors having long-term neurological disabilities and only three patients requiring antiepileptic medications at time of discharge. A larger cohort of patients can help identify patient factors that increase the risk of neurological events.

Sun 14

Volumetric Size Matching Expands The Donor Pool In Pediatric Heart Transplantation

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Background: Pediatric heart transplant (PHTx) candidates have the highest waitlist mortality of any solid organ transplant group largely due to the donor shortage. For 55 years, the donor-recipient-weight-ratio (DRWR) has been the standard for determining the upper limits of donor size. We have been using volumetric size matching (VSM) since 2015, allowing us to rely on donor-recipient-volume-ratios (DRVVR). Since 2015, we have transplanted four children with rarely seen DRWRs using volumes instead of weight. Herein, we review our evolving process for VSM, share two cases with prospectively documented volumes, and describe the retrospective pattern of DRWR in PHTx using OPTN data.

Methods: 53-day-old female with dilated cardiomyopathy presented in cardiogenic shock. A trial of PA-banding was unsuccessful. She remained intubated on high-dose inotropes with marginal reserve and poor VAD options. Child underwent HTx with DRWR=3.3 (donor weight 16.4kg, recipient weight 5kg), DRVVR=0.82 (donor volume 170cc, recipient volume 207cc). Had intra-operative chest closure and discharged 2 weeks post-transplant. Child's sister presented two years later at age 4 months. Progressed rapidly to status=1A requiring intubation and inotropes. Child underwent HTx with DRWR=3.2 (donor weight 26.5kg, recipient weight 8.3kg), DRVVR=0.74 (donor volume 170cc, recipient volume 230cc). Had intra-operative chest closure and discharged 23 days post-transplant.

Results: Our group long held that advanced imaging (CT/MRI) is the most accurate way to determine total cardiac volume (TCV). However, this was usually unavailable at the time of donor offers. We therefore developed surrogate tools using echocardiography. We published our experience validating echo to determine TCV in 2013. We demonstrated a relationship between LVedV and TCV in children with normal hearts (i.e. donors) and showed a correlation between TCV by MRI and by modified Simpsons tracing in children with abnormal hearts (i.e. recipients). These tools were used for the children in this report. With advances in image sharing technology and release of the new UNET viewer in 2019 allowing direct downloads of donor images, we have moved to advanced imaging for our "oversized" donors. These measurements can be completed in minutes, remotely by the imaging professional. We've transplanted four children recently with DRWR >3 using VSM. Using OPTN data, there were 4051 PHTx in children <10 years from 2005 - 2021. Only 30 (0.7%) had DRWR >3; including these four subjects. In 2021, 685 pediatric donor hearts were offered for transplant with 523 accepted while nearly a quarter of available organs (23.6%) went unused.

Conclusions: There simply aren't enough donor hearts available for all the children that need them and too many organs are discarded each year. Centers are historically reluctant to expand the donor pool by increasing the upper DRWR for their recipients despite significant relative cardiomegaly. We are comfortable accepting organs with a DRVVR up to and exceeding 1 using VSM. Unfortunately, weight is insufficient as a metric to discriminate between the oversized donor with an acceptable sized graft and one that is prohibitively large. Volumetric size matching allows centers to expand their candidates' donor pools, shorten wait times, decrease organ wastage and ultimately save more lives.

Sun 15

Is There A Role Of Procoagulant Work Up In Predicting A Successful VAD Course?

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Background: To ensure minimal thrombotic risk when implanting a ventricular assist device (VAD), a procoagulant workup (genetic mutations, deficiencies, and coagulation studies) is done prior to implantation. Our institution began consistent antiplatelet and anticoagulation work-ups prior to VAD support in 2019. This study aims to assess the ability of the procoagulant work-up to indicate future risk of stroke while implanted with a VAD.

Methods: We reviewed 29 patients implanted with a Berlin Heart EXCOR from January 2019 to June 2022. Four patients who had extracorporeal life support (ECLS) prior to VAD implant were excluded. Two patients were excluded due to severe hemolysis and hyperbilirubinemia. 23 patients were included in the study. Our procoagulant work up includes Anti-thrombin III deficiency, Protein S deficiency, Protein C deficiency, Factor V Leiden deficiency, Prothrombin gene mutation, antiphospholipid antibody, and TEG alpha, MA, K, and R. End points include number of VAD changes, CNS changes with associated CT changes, seizures, and other thrombotic events.

Results: Median age was 3 months with 60% of the patients supported for failing SV physiology. The 19/23 (82%) survived to discharge. Of the 23 patients only 3 patients (8.6%) who had an abnormal prothrombotic work namely heterozygous Factor V Leiden gene mutation and heterozygous Prothrombin gene mutation. Of note, Protein C, Protein S, and Antithrombin were low for the majority of patients on laboratory but when corrected for young age, were normal. The third patient was noted to have an abnormal lower Protein C value in the context of liver fibrosis with a failing Fontan circulation. Seven patients had evidence of ischemia areas on CT scans. All three patients with an abnormal profile developed CNS insults with all three surviving to discharge. The patient with a heterozygous prothrombin gene mutation required 4 VAD exchanges for thrombus burden. CT changes include focal MCA and PCA infarction, as well as multifocal areas of infarct outside of a vascular distribution.

Conclusions: Many of the thrombotic risk factors that are positive in our patients are heterozygous especially for factor deficiencies. Although these mutations may not be significant in a non-VAD population, they have more significance in the context of a secondary risk factor. In addition, patients with failing Fontan circulation are likely to have an imbalance of prothrombotic factors leading to a predisposition of clotting in addition to bleeding. The need for VAD exchange and other prothrombotic risk in this population has been limited especially with improvement in antiplatelet and anticoagulation strategies. Although these heterozygous variants may not limit VAD implants, they should definitely warrant more caution while maintaining good levels and limit discontinuation of anticoagulation drips.

Sun 16

Possible Use Of Alpha Glucosidase Therapy For Hypertrophic Cardiomyopathy In The Context Of Danon Syndrome.

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Background: 4-month-old with a known diagnosis of Danon syndrome with hypertrophic cardiomyopathy presented from an OSH for respiratory symptoms secondary to parainfluenza 3. Upon admission, patient was in respiratory failure with acidosis and required emergent intubation. He appeared septic, with fever and leukopenia. In the context of the severely hypertrophied interventricular septum with hyperdynamic systolic function, patient needed extensive resuscitation with volume, followed by heart rate and blood pressure regulation with esmolol and phenylephrine respectively. He was eventually stabilized on mechanical ventilation, low dose phenylephrine along with additional heart failure pharmacological management with beta blockers, disopyramide, levocarnitine and intermittent diuresis.

Results: Due to his underlying genetic syndrome of Danon syndrome, he was not deemed a transplant candidate. A genome analysis found pathologic variants in his DNA other than those associated with Danon disease. Muscle biopsy performed noted striking findings in skeletal muscle. Ultrastructural studies revealed marked accumulation of free-floating and membrane-bound glycogen and occasional autophagic vacuoles consistent with a vacuolar myopathy. While mild accumulations of mitochondria were seen, no definitive evidence of mitochondrial dysmorphology was identified. It was decided that patient will benefit from enzyme replacement therapy with Alpha glucosidase (Lumizyme) as it has been used in HOCM in the context of other disease with glycogen storage like Pompe's disease.

Conclusions: He received four doses, once a week for 4 weeks. Serial echos were performed with mild thinning of septum from 22 mm to 14mm, with improved dynamic biventricular outflow tract gradient and unchanged mild/mod biventricular valvular regurgitation. Along with supportive care, the patient improved from a cardiorespiratory standpoint and was successfully extubated after 2 months of intubation onto full-face BiPAP. He is one of the first if not only case of Danon syndrome associated HOCM to be treated with Alpha glucosidase enzyme. Although traditionally used for Pompe disease and associated improvement in the HOCM. Our case indicates utility in phenotypes with some overlap such as Danon syndrome.

Sun 17

Helping Heart Transplant Patients Thrive: A Multi-Disciplinary Approach

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Background: Pediatric heart transplant recipients are at higher risk for anxiety, depression, and post-traumatic stress. Likewise, adolescent and young adult transplant recipients are at increased risk of nonadherence, with known negative impact on health outcomes. Traditional transplant clinic models involve the patient seeing the medical team only, and then referred afterwards to further evaluation/intervention by psychology and social work if needed. This model may not allow for the early identification and timely intervention for psychosocial and adherence concerns. We piloted a multidisciplinary care model where patients were seen in the same visit by the medical team, psychology and social work.

Methods: We created a multi-disciplinary clinic titled THRIVE (Transplanted Heart Recipients: Independence, Vitality & Education), including the HT medical team, psychologist, and social worker. We prioritized scheduling our perceived highest risk patients for this clinic and aim for all patients to be seen in the THRIVE clinic at least once annually. Goals of clinic include assessing for undetected psychosocial concerns and preparing young adult patients for transition to adult care. In contrast to past practice, all providers meet with the patient and family during a single visit in effort to improve access to targeted psychosocial intervention.

Results: Between March 2021 and December 2022, 61 unique patients had a visit in THRIVE clinic which included a psychologist and social worker. During this time period, an additional 29 unique patients were seen outside of our clinic by either our HT psychologist or social worker, for a total of 89 patients followed over a 22 month period. This compares to 69 patients seen by either HT psychology or social work in the prior 22 months (May 2019-February 2020). This comprises seventy six percent of the patients ≥ 12 years of age followed by our HT program since March 2021. We identified patients with suicidal ideations, engaging in high-risk behaviors, and those who may need additional psychosocial support outside of our clinic. We addressed topics including advanced directives, power of attorney, guardianship, medication adherence and independence. Surveys were given prior to the clinic visit to assess for potential concerns, and then following the visit to gauge patient and family perception of their experience in the clinic. Patients endorsed the benefits of the THRIVE clinic, reported that it did not notably extend their time in clinic, and identified the transition to adult care as an area of focus.

Conclusions: A patient-centered multi-disciplinary outpatient care model for adolescent and young adult HT patients is feasible and can address issues critical to the psychosocial well-being of these vulnerable patients. More work is needed to quantify the effect of this intervention, and to evaluate its applicability to other populations.

Sun 18

Not All That Wheezes Is Asthma —A Case Of Familial Thoracic Aortic Aneurysm And Dissection

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Background: Familial Thoracic Aortic Aneurysm and Dissection (FTAAD) is a major cause of cardiovascular morbidity and mortality (3). Hereditary aortic disease can lead to thoracic aortic aneurysms involving the aortic root, ascending aorta, or fusiform dilation of both segments, which can result in dissection. Family history of aortic aneurysms and dissection in addition to syndromic features are the key characteristics used to classify affected individuals. Genetic advances have identified only a minority of the genes involved in FTAAD. The LOX gene is a recent addition to this group.

Methods: We present a 13-year-old female with cough, wheezing, and a mediastinal mass detected on chest radiograph. Computed Tomography Angiography of the chest revealed an 8.2 cm ascending aortic aneurysm with no coronary involvement. The patient was admitted to the Cardiovascular Intensive Care Unit and cardiothoracic surgery was performed using a valve-sparing technique, replacing the aortic annulus and ascending aorta with a 26mm Hemashield graft. Genetic testing revealed a pathogenic truncating variant of the LOX gene, c.732_735delTCG; Cys244Trpfs*25(1). Pathogenic variants in LOX are known to cause autosomal dominant FTAADs. Subsequent genetic testing of family members revealed the same LOX variant in three siblings and father. Interestingly, the patient's non-identical twin was being followed since age 3 with aortic dilation and Marfanoid habitus but had a negative aortopathy gene panel from a different lab the year prior. The family was counseled about this autosomal dominant condition with a 50 percent recurrence risk.

Results: The Lysyl Oxidases (LOX) copper-dependent oxidodeaminases are involved in covalent cross-linking of collagen and elastin in the extracellular matrix and provide tensile and elastic properties of connective tissue (2); and contribute to aortic structural integrity. This patient has a frameshift variant leading to loss-of-function. Review of the literature reveals aneurysm reported in a 6-year-old and dissection in a 19-year-old, suggesting that LOX gene variants can be an early cause of FTAAD (3,7). Although this case did have elements of positive family history with Marfanoid phenotype, it should be noted that variants in the LOX gene may cause FTAAD in the absence of observable skeletal features delaying initial diagnosis (7). In addition, LOX is a recently identified cause of FTAAD and this gene may not be present on all aortopathy panels. As FTAAD panels do not detect an underlying gene variant in many cases, screening aortic imaging of at-risk relatives is recommended. Screening recommendations for family members of FTAAD patients are needed and should address appropriate age to begin screening.

Conclusions: This case emphasizes the importance of echocardiogram screenings in first-degree family members with known aortic dilation and genetic counseling with updated genetic testing. Importance of recognizing the Marfanoid phenotype and family history by the primary care provider/cardiologist should also be emphasized.

Sun 19

A Case Of Incidentally Diagnosed Left Main Coronary Artery Ostial Atresia In An Asymptomatic Neonate With A VSD: Diagnostic And Management Challenges

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Background: Left main coronary artery ostial atresia (LMCAOA) is a rare congenital anomaly, with only 100 reported cases in the literature. Almost half of pediatric patients have associated cardiac abnormalities, although the association with ventricular septal defects (VSD) is rare. Most patients present symptomatically, however accurate diagnosis can be difficult, and it is often misdiagnosed as anomalous left coronary from the pulmonary artery (ALCAPA). Indications for surgical revascularization have evolved. Most recent guidelines suggest that conservative management may be appropriate in the absence of symptoms, ischemia, impaired systolic function, and in the presence of good-sized collaterals to the left coronary system.

Methods: Here, we present a case of an asymptomatic neonate incidentally diagnosed with LMCAOA after referral for a VSD. She was found to have a large conoventricular VSD with inlet extension, and normal ventricular systolic function. Her initial imaging by echocardiography and cardiac catheterization was suspicious for ALCAPA, however LMCAOA was confirmed in the operating room. Surgical revascularization was not pursued at time of diagnosis. She has continued to be asymptomatic, and cardiac MRI at 9 months of age confirmed LMCAOA with normal biventricular function and no evidence of ischemia.

Results: Here we report the first association of LMCAOA with a conoventricular and inlet VSD. Importantly, this associated cardiac anomaly may have complicated her diagnosis by angiography due to contrast flow through her left to right shunt. Her anomalous coronary was not repaired at time of diagnosis, and she continues to be asymptomatic. Guidelines for the optimal management of patients with LMCAOA have evolved over time, but have been limited by the rarity of the condition. The vast majority of pediatric patients with LMCAOA present symptomatically. Historically 85% of these patients have undergone surgical revascularization, which carries a 10% mortality risk. Given this, it has been suggested that it may be appropriate to delay surgical intervention in the absence of symptoms and/or evidence of cardiac dysfunction or ischemia.

Conclusions: LMCAOA may be diagnosed incidentally in an asymptomatic pediatric patient with normal ventricular systolic function, especially in the setting of an associated cardiac anomaly. The presence of a VSD may complicate the diagnosis of LMCAOA. In these scenarios, cross-sectional imaging may be a useful diagnostic modality. It may be reasonable to delay surgical revascularization given the operative risk may outweigh the risk of adverse cardiac events in this patient population.

Sun 20

"Just In Time" - A Novel Educational Tool For Anticipatory Management Of Postoperative Patient's In The Infant Cardiac ICU

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Background: Our unit utilizes a novel concept known as "Just in Time" to provide knowledge to front line providers which enables them to be equipped with the tools needed to deal with acute care postoperative situations including recognition and management of complications. "Just in Time" is a multidisciplinary discussion involving visual aids of infant specific cardiac anomalies and specific procedures. This forum identifies a variety of scenarios that one could anticipate in a post-operative patient

Methods: A survey questionnaire was sent to advance practice providers and nurses asking how frontline providers learn best during acute care situations. This questionnaire included multiple choice questions, preferred style of learning, number of years as a provider and the benefits of "Just in Time" discussions.

Results: The questionnaire was sent to a total of 94 participants including both advanced practice providers and registered nurses. 55 participants responded and out of those 24 were APPs and 31 RNs. When asked to rate the importance of our "Just in Time," 80% of the respondents felt that it was very beneficial.

Conclusions: In the current era, the optimal teaching method is unknown. Based on our data in our dedicated neonatal cardiac intensive care program, both APPs and RNs felt that our novel concept of "Just in Time" training was both empowering and beneficial when managing acute care postoperative situations.

Sun 21

A Centralized Guideline Tool For The Care Coordination Of Critical Cardiac Deliveries: A Lean Process

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Background: Prenatal detection of congenital heart disease allows for identification of structural heart differences & classification of severity, which both aid in delivery and immediate postnatal management planning. A high risk for cardiopulmonary instability immediately following delivery is present in babies with severe forms of CHD, often requiring urgent neonatal & cardiac intervention using a multidisciplinary team approach involving several different specialty teams across the hospital system. A lack of standard work for these critical cardiac deliveries was identified at a tertiary care center, leaving the potential for communication breakdown, hospital waste and most importantly, compromised patient care.

Methods: Utilizing the Lean A3 problem solving method, gaps in the care planning model were identified using a comprehensive feedback tool to assess for common themes across care teams. Analysis was performed utilizing a fishbone diagram to identify relationships between identified problems & outcomes. Countermeasures were developed & prioritized using a PICK Chart, and were implemented utilizing a Countermeasure Matrix Table. The project goal was to optimize care coordination & delivery room management for the most critical cardiac deliveries by increasing multidisciplinary teamwork, communication & education, as well as decreasing delays in care, communication breakdown & hospital waste.

Results: A centralized guideline outlining the multidisciplinary care coordination & communication for highest risk critical cardiac deliveries in a busy tertiary care center was developed. 22 healthcare professionals across 7 specialties participated in focus sessions, where participants answered both standard and departmental specific questions. Categorical themes were identified as beneficial or obstructive factors to the optimization of care coordination. Beneficial factors included scheduled deliveries, identified points of contact, multidisciplinary team meetings, direct physician communication, simulations, and team huddle. Largest obstructive contributing factors to communication breakdown, hospital waste and potential for compromised patient care included infrequent occurrence, multiple management teams, organizational silos, lack of education & unclear roles & responsibilities. Countermeasures were developed following identification of problem gaps and root cause analysis, resulting in a standardized care coordination pathway, including a comprehensive written guideline, the implementation of interdepartmental joint simulations, protocolized direct provider communication, and multiple delivery planning tools, including: departmental contact lists, revision of the institutions Cardiac Delivery Classification Guideline, care coordination process map, delivery planning checklist & the development of a critical event note template.

Conclusions: The standardization of multidisciplinary care coordination & delivery room management for critical cardiac deliveries can provide a streamlined approach to the complex prenatal management of fetuses who are at high risk for cardiopulmonary instability immediately following birth. Utilizing the Lean A3 problem solving method provided tools to help identify gaps in current care planning models & perform a root cause analysis, aiding in the development of a comprehensive care coordination pathway for the highest risk critical cardiac deliveries, with the intention to streamline interdisciplinary communication, decrease hospital waste & optimize patient outcomes. Due to the nature of infrequent occurrence of this subset of critical cardiac deliveries, further assessment of the efficacy of such protocolized care pathways is needed.

Sun 22

Parents Post-Traumatic Stress Before And After Their Infant's Correlational Longitudinal Study

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Background: One percent of infants born have CHD making it the most common congenital anomaly. Of those infants born with CHD, 1% have a functionally single ventricle. This type of heart defect requires three palliative heart surgeries, the first within seven days of birth, the second at 4-6 months of age, and the third at 2-5 years of age. Parents endure emotional trauma from witnessing their child in life-sustaining care, experiencing reoccurring hospitalizations, and the evolving reality of having a child with a chronic illness. Parent trauma impacts normal experiences for the parent and the infant resulting in impaired coping, bonding, and confidence. The infant may have problems with feeding or neurodevelopment. Parents with unresolved trauma may be less engaged in medical care which is a predictor of later infant outcomes. The purpose of this study is to describe the levels of parent post-traumatic stress before and after their infant's second palliative surgery for SVCHD.

Methods: A descriptive correlational, longitudinal design was used. The post-traumatic stress disorder checklist revised for the DSM-5 (PCL-5) was administered via Research Electronic Data Capture (REDCap) to 22 parents before and after the second palliative heart surgery. Descriptive statistics were used to evaluate and describe the demographic data and the level of post-traumatic stress at the two-time points. The Wilcoxon signed-rank test was used to determine the relationship between the two PCL-5 scores

Results: There were positive PCL-5 scores present pre-surgery and post-surgery. Most of the scores (n=13, 59%) decreased between surgery one and surgery two, although we did not have statistical power to make definitive conclusions from the data. An inverse association between the PCL-5 scores and income and level of education were noted.

Conclusions: Previous qualitative research suggested that parents had a significant response to the end of their time with CHAMP. This study aimed to further explore that phenomenon quantitatively and found somewhat contradicting results. Failure to replicate findings of qualitative work may be due to the retrospective nature of the initial work. There were significant symptoms at both timepoints which highlights the need for further work. With all positive scores, there was a referral to the heart center psychologist for follow up. This provided an opportunity to counsel families and provide resources. The unexpected psychosocial follow up due to the study design highlights the continued need for comprehensive care for these patients and families after hospital discharge. Future research should seek to obtain scores at consistent timepoints not bound by surgery.

Sun 23

Nurse Navigation In Congenital Cardiology

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Background: The role of the nurse navigators within congenital cardiology is not well documented; however, improvements in outcomes supported by the navigator role are evident for other patient populations, both pediatric and adult. Therefore, the purpose of this abstract is to describe growth within a single center and the best practices for the nurse navigator in congenital cardiology and to support the implementation of the role in programs throughout the field. The Nurse Navigator role was implemented at Levine Children's Congenital Heart Center (LCCHC) to streamline care and create a positive patient experience from referral throughout their continuum of care.

Methods: The congenital heart population is complex, with some patients requiring frequent interventions. The navigator role was established at LCCHC in 2013. Over a decade, the role evolved into six nurse navigators, each specialized to the unique needs of this patient population. Navigators are now established at this center for fetal, surgery, interventional cardiology, inpatient, electrophysiology, and adult congenital patients. Despite the subspecialties, the goals are identical: to assist and educate patients, trusted adults, and support persons, to identify and break down barriers while expediting care, and to be a compassionate and knowledgeable coordinator through the continuum of care.

Results: The fetal navigator role was developed in 2013 to better prepare and educate mothers who fetus received a complex congenital heart diagnosis. In 2013, there were 38 fetal referrals that were followed through delivery. This increased to 58 in 2014, demonstrating 34% increase. Growth remained consistent through 2021, which had 69 referrals through delivery. In 2017, surgical case volume was 258 patients, which increased to 292 in 2021, a 12% increase, average of 302 cases in between. Post-operative length of stay averaged 12.9 days in 2017, 12.5 days in 2021. Readmissions within 30 days in 2017 were 11.2% and in 2021 were 9.2%. The coordination of appropriate consultations prepares patients and support persons for the care and management of congenital heart disease. Surgical, interventional, and electrophysiology referrals activate a similar process; however, given the wide variety of interventions, individualized support is offered to each patient and family. For inpatients, navigator care is prompted within the hospital setting. Focused support is provided throughout hospitalization with continuous education to facilitate smooth transition to home and reduce the occurrence of readmission. The Adult Congenital Navigator is prompted within the referral process, according to the patients within this subspecialty that have complex needs.

Conclusions: Nurse navigators focus on the complex needs of patients throughout the continuum of care. The growth and evolution of the nurse navigator role mirrors that of the Congenital Heart Program at LCCHC. The addition of subspecialty nurse navigators, including fetal, surgical, electrophysiology, inpatient, pediatric, and adult provides a tailored and individualized approach to congenital cardiology care. As more patients age out of pediatric care and therapies advance, more specialized navigators were needed to match this growing and evolving patient population. While informal patient feedback has been positive, more research related to this role is needed within the field. There are multiple publications to support the utilization of this role among Oncology programs, yet the literature offers no information on the utilization of nurse navigators within Congenital Cardiology and needs to be further investigated.

Sun 24

CLABSI Reduction Through Detailed Line Rounds

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Background: Central Line Associated Bloodstream Infections (CLABSI) are an ongoing challenge in the Pediatric Cardiac Intensive Care Unit (PCICU). The PCICU has an increased use of central venous lines (CVL) due to the difficulty of maintaining intravenous access on pediatric congenital heart patients. The first six months of 2022 proved to be challenging, with a spike in CLABSI events. The goal of this initiative is to reduce patient harm by decreasing the incidence of CLABSI by 1 event for the remaining 6 months of 2022 and continuing into 2023.

Methods: A multi-modal approach utilizing the PDSA cycle. Jan: Focus on line rounds. June: Nursing simulation of sterile culturing practices and identifying sources of infection. Provider education on ensuring proper culturing or imaging of all potential sources. August: Infection control reviewed provider documentation necessities when treating necrotizing enterocolitis to rule out CLABSI. September: Introduction of multidisciplinary weekly line rounds, which review central line necessity and decreasing lab schedules. Review of KVO with heparin on all small PICCs added to daily line rounds. November: Added shelving for diaper scales to reduce the environmental bioburden where patient care items are kept and prepared.

Results: In 2021, the PCICU had a total of 4 CLABSIs. The PCICU had a total of 6 CLABSIs in 2022, with 4 CLABSIs in the first 6 months and 2 CLABSIs in the last six months. This is a 50% reduction of CLABSIs compared to the first six months of 2022. Line rounds have proven to reduce labs by up to 7% in a month and identify up to 4% within a month of CVLs that need removal.

Conclusions: Process improvements related to CLABSI reduction remains a high priority for the PCICU. It has been established that reduction of CLABSI requires vigilance and compliance with the many elements of care required for CVLs. Interventions for CLABSI reduction should remain focused on modifiable factors, such as care and maintenance of the CVL and reduction of lab draws. Ongoing application of the PDSA cycle to these interventions will guide their continued inclusion in the care bundle of CLABSI reduction.

Sun 25

Decreasing Medication Overrides In The Pediatric Cardiac Icu

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Background: Preventing medication errors from reaching patients requires multiple layers of safety protocols. Because over half of all medication errors occur during administration, pharmacy order verification prior to administration is an important step for the prevention of errors. Emergent situations may require an override to access a medication prior to pharmacy verification. However, this practice introduces an increased level of risk for patients and should occur rarely. While the institutional goal for medication overrides is 2.4 percent, the Pediatric Cardiac ICU rate was at 12.4 percent. The aim of this performance improvement project is to reduce medication overrides by 6 percent.

Methods: The team used the PDSA method for process improvement. The first step included identifying the most common medications obtained from the medication cabinet by override. This informed the three-pronged approach which included educating nurses on the importance of avoiding medication overrides, utilizing medication kits when appropriate such as a rapid sequence intubation (RSI) kit, and collaborating with physicians to ensure timely order entry so as not to delay pharmacy verification. Finally, flyers summarizing the strategies to decrease medication overrides were posted in the unit.

Results: After implementing our multi-pronged approach, medication overrides decreased initially to 8.4 percent. Within 5 months, the override rate further declined to 3.5 percent demonstrating sustained improvement in performance. While increasing nurses' awareness and interprofessional collaboration were effective improvement strategies, utilization of medication kits was critical to successful reduction of overrides within the Pediatric Cardiac ICU. Due to the acuity and workflow of the ICU, nurses frequently need to obtain several medications simultaneously in response to a common emergent situation such as RSI. Medication kits allows the nurse to pull a standard set of medications at one time avoiding the risk of pulling incorrect medications. Because medication kit utilization is not only safer but quicker, nurses readily adopted this practice change.

Conclusions: As a result of our performance improvement activities, we significantly decreased the number of overrides from the medication cabinet but have not yet achieved the institutional goal of 2.4 percent. Future efforts will focus on improvements in the workflow for patients admitted directly from the surgery. We will also continue to collaborate with physicians to ensure non-emergent orders are entered timely. Finally, we must collaborate with pharmacy to decrease the length of time from order entry to order verification. Continuing these efforts and education will help solidify these practices into the unit culture and make this project a long-term success.