

Friday 1

Use Of Landiolol In Critically Ill Pediatric Cardiac Patients

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Background:

While β -blockers can be effective in controlling tachyarrhythmias, a negative inotropic influence complicates their use in critically ill children with cardiac diseases. Landiolol hydrochloride is a novel, ultra-short-acting β -blocker with higher $\beta_1:\beta_2$ selectivity ratio and a less negative inotropic effect than other used β -blockers. This study evaluates the efficacy and safety of landiolol in the management of tachyarrhythmias in critically ill pediatric cardiac patients.

Methods: A retrospective analysis was performed on 9 pediatric cardiac patients treated with landiolol at the Pediatric Cardiac Intensive Care Unit. The median age of patients was 5 months (range; 2 weeks – 12 years). Six (66.6%) patients with congenital heart defects had arrhythmias after cardiac surgery. Three (33.3%) patients were with dilated cardiomyopathy (DCMP). One of these patients underwent pulmonary artery banding and was treated with landiolol to control sinus tachycardia prior to and after surgery. Other 2 patients with DCMP were receiving landiolol for treatment of arrhythmias. Continuous parameters are presented as median (range).

Results: In the whole group of patients, arrhythmias included intraatrial reentrant tachycardia (IART) in 1 patient, IART and atrioventricular reentrant tachycardia (1 patient), IART, atrial ectopic tachycardia and macroreentrant atrial tachycardia (1 patient), ventricular extrasystoles and ventricular tachycardia (1 patient), non-automatic focal ectopic tachycardia and ventricular tachycardia (2 patients), and junctional ectopic tachycardia (2 patients). Maximal heart rate was 202 per minute (133 to 258 per minute). In cardiosurgical patients, arrhythmias occurred at 180 (0 -648) hours after surgery and landiolol was started 192 (0-648) hours after surgery. In the whole group, the loading and maximal doses of landiolol were 10 (1 – 60) and 40 (5.1 – 60) $\mu\text{g/kg}$ per minute, respectively. Landiolol was administered as a first line antiarrhythmic therapy in one patient, in others as a second-line or subsequent antiarrhythmic therapy. Within 2 hours, landiolol reduced the heart rate from 170 per minute (113 – 205 per minute) to 119 per minute (85 – 181 per minute), $P = 0.03$. Rate control was achieved in 7 of 9 (77%) patients. Duration of landiolol administration was 107 (1 – 1327) hours. No adverse effects were observed in any patient.

Conclusions: The study suggests that landiolol may be a promising option for the management of tachycardias in critically ill cardiac patients. More data is needed to establish the position of landiolol in the drug therapy regimen of pediatric tachycardias.

Friday 2

Dexmedetomidine Withdrawal In Pediatric Critical Care: A Systematic Review And Meta-Analysis

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Background: Dexmedetomidine (DEX) is an alpha 2 agonist sedative and analgesic that is commonly used in opioid-sparing and enhanced recovery strategies for pediatric cardiac critical care patients. While clinical withdrawal can occur upon discontinuation of DEX in children, data describing the clinical signs, risk factors, and treatment for DEX withdrawal in this population have not been summarized. We aimed to systematically review and analyze the literature describing clinical signs, risk factors, and management of DEX withdrawal in children in the intensive care unit setting.

Methods: A PRISMA-guided systematic review was conducted, including publications (2000-2022) reporting on pediatric (age <21 years) critical care patients following discontinuation of intravenous DEX after = 24 hours of exposure. Cohort characteristics, care setting (cardiac critical care unit vs. pediatric intensive care unit), DEX exposure, and DEX withdrawal prevalence were abstracted. Weighted averages described the proportion of each study population that experienced common withdrawal signs including hypertension, tachycardia, and agitation. Associations between DEX exposure (median maximum dose, median cumulative dose, and median duration of DEX exposure) and the prevalence of DEX withdrawal signs were assessed using weighted linear regression models.

Results: Overall, 498 studies were screened. Ultimately, 23 studies were included (28 unique cohorts, N=2,002 patients assessed for DEX withdrawal). DEX withdrawal definition varied across studies. DEX withdrawal was characterized by hypertension (n=15 studies) tachycardia (n=17), agitation (n=11), elevated WAT-1 scores (n=9), and other signs (n=12). Median patient age was 10 months [IQR 5-20], median DEX duration was 132 h [IQR 87-187], and median cumulative DEX exposure was 106 mcg/kg [IQR 52-128]. Increasing median age was associated with increasing weight-adjusted DEX exposure (Spearman's rho .66, $p < .01$). Raw proportions of withdrawal signs were: agitation (271 occurrences/939 patients; 29%), hypertension (705/1433; 49%), tachycardia (573/1738; 32%), elevated WAT-1 scores (99/544; 18%), and other (111/915; 12%). Pooled weighted estimates compared the prevalence of withdrawal signs between cardiac surgery and not-cardiac surgery cohorts. Not-cardiac surgery cohorts had higher prevalence of hypertension (.09 v .41, $p = .028$) and tachycardia (.04 v .30, $p = .007$). Three studies reported a lower prevalence of withdrawal when clonidine was used to transition off DEX. Higher cumulative DEX dose and longer duration of DEX infusion were reported risk factors for DEX withdrawal. Reported associations with withdrawal included cumulative DEX exposure >107 mcg/kg ($p = .03$) and DEX duration = 72 h ($p = .007$). Linear regression models showed no significant association between cohort-level DEX exposure and withdrawal.

Conclusions: DEX withdrawal in children commonly manifests as tachycardia, hypertension, and agitation. Risk factors may include higher cumulative dose and longer duration of DEX exposure, but the current meta-analysis utilized weighted averages for each cohort and is therefore limited by unavailability of granular, patient-level data. Prospective controlled studies are needed to further characterize patient-level associations between DEX exposure and clinical withdrawal signs, and to guide standardized diagnostic and management strategies for DEX withdrawal. Future work should also explore resource utilization and clinical impact of DEX withdrawal on cardiac critical care outcomes.

Friday 3

Renin Levels As A Predictor Of Post-Operative Vasoplegia And Acute Kidney Injury In Patients With Congenital Heart Disease Undergoing Cardiac Surgery Utilizing Cardiopulmonary Bypass

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Background: Cardiopulmonary bypass (CPB) is an essential component of cardiac surgery but carries inherent risks to the patient. Patients are at risk for a pro-inflammatory state, fluid shifts, acute kidney injury (AKI), and vasoplegic shock. Adult studies show the Renin-Angiotensin-Aldosterone System (RAAS) as a potential target for therapy, especially in patients with catecholamine resistance. Renin has been shown in adult patients to predict a poor response to standard therapies and prognosis. This has not been studied in pediatric cardiac patients following CPB.

Methods: This was a retrospective single center study of infants who underwent cardiac surgery with CPB (n=50). The aim was to characterize renin kinetics during the first 24 hours post-operatively and determine if elevated renin levels were associated with AKI and vasoplegia. Plasma samples were analyzed pre-operatively and post-CPB (1h, 4h, 24h). Patient data was obtained through the EMR and PC4 data registry. A mixed-effects regression model was used to describe renin trajectory over time. Separate logistic regression models for AKI and peak VIS ≥ 9 were used to determine their association with renin levels.

Results: The patient cohort had a median age of 5 months (3.5, 6.5), predominantly male (64%), and predominantly white (86%). STAT category distribution was equal amongst the 5 categories except for STAT 5 constituting 6% of surgeries in this cohort. Median weight was 5.7 kg (5.12, 6.58). Median CPB time was 171 minutes (112, 216). No patients required continuous renal replacement therapy, ECMO, or had any post-operative cardiac arrest events. Renin levels peaked 4-hours post-operatively and declined within 24 hours (Figure 1). There was a significant difference in renin levels between 4-hour post-bypass and pre-op (mean difference = 100.62, 95% CI = 48.88 to 152.35, P-value < 0.001, Table 1). The odds of Peak VIS ≥ 9 increased with increasing renin level (4h post bypass) (OR = 3.37) but was not statistically significant. This was also similar for the odds ratio of AKI with increasing renin level (OR = 2.73) but was not statistically significant.

Conclusions: This is the first study describing plasma renin level kinetics in the pediatric congenital heart surgery population. We demonstrated that renin levels peak at 4-hours post-CPB and subsequently will decrease at 24-hours post-CPB. In addition, there may also be significant associations with 4-hour post-CPB renin levels with VIS and AKI but due to our small sample size and small event number we were unable to detect moderate to small effect sizes. Future studies should examine the utility of renin levels in larger models and its ability to predict adverse outcomes. Like the adult studies, elevated renin levels may also identify an at-risk phenotype that would potentially be amenable to therapies directed at the RAAS during instances of vasoplegia.

Friday 4

Randomized Controlled Trial Of Heparin Vs. Bivalirudin Anticoagulation In Acyanotic Children Undergoing Open Heart Surgery.

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Background: To determine the safety and efficacy of bivalirudin as an anticoagulant for pediatric open heart surgery and to determine its appropriate dosage for this purpose.

Methods: Prospective, randomized controlled trial. Setting: Tertiary care hospital. Participants: Fifty acyanotic children aged 1-12 years undergoing open heart surgery. Interventions: The children were randomized to receive either 4 mg/kg heparin (n=25, group H) or 1 mg/kg bivalirudin bolus followed by 2.5 mg/kg/hr infusion (n=25, group B) as the anticoagulant. The doses were adjusted to maintain activated clotting time above 480 seconds. At the conclusion of surgery, protamine (1.3 mg/100 U of heparin) was administered to children in group H.

Results: The children were comparable in both groups demographically. The mean age, weight were 51.5 months and 13.4 kg in group H, and 59.3 months and 13.4 kg in group B. The dose of anticoagulant required was 4.0 ± 0.2 mg/kg in group H and 1.7 ± 0.2 mg/kg followed by 3.0 ± 0.7 mg/kg/hr infusion in group B ($P < 0.001$). One child required additional dose in group H compared to 13 (54.2%) children in group B. Intraoperatively, the ACT achieved was higher in group H compared to group B ($P < 0.05$). The ACT returned to baseline value after protamine administration in group H but it remained elevated for 2 hours after termination of CPB in group B ($P < 0.01$). The ACT was higher in group B compared to group H for 6 hours after CPB ($P < 0.05$). Heparin prolonged onset of clotting, decreased the rate and strength of thrombus formation and inhibited platelet function more than bivalirudin on viscoelastic coagulation testing. The duration of surgery was prolonged in group B. Postoperative chest tube drainage was similar in group B (4.9 ml/kg) as in group H (5.9 ml/kg) in spite of higher ACT. Transfusion requirements were similar. No adverse event occurred in any patient.

Conclusions: Bivalirudin is a safe and effective anticoagulant for paediatric open heart surgery. Though it is not suitable as a routine anticoagulant for this purpose, but it may be used as a heparin alternative in instances when heparin cannot be used. The dose required to maintain activated clotting time more than 480 seconds was 1.7 ± 0.2 mg/kg followed by 3.0 ± 0.7 mg/kg/hr infusion. The activated clotting time remained elevated for 2 hours after stopping the infusion. Bivalirudin did not increase postoperative bleeding and transfusion requirement.

Friday 5

Retrospective Review Of Incidence Of Intracranial Hemorrhage In Setting Of Reduced Heparinization During Cardiopulmonary Bypass

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Background: Cardiac congenital neonates and infants have pre-existing comorbidities coming into heart surgery. Prematurity of all organ systems may compromise cerebral blood flow in utero and increase the risk of neurological complications during or after cardiopulmonary bypass (CPB). In efforts to minimize the risk of cerebral hemorrhage, this institution implemented an Intracranial Hemorrhage Protocol (ICH) that resulted in an overall reduction in heparin administration. Specific outcomes of this protocol included a reduction in targeted heparin levels as well as detailed monitoring and maintenance of platelet counts for high risk patients.

Methods: Retrospective chart review over a 4-year period for all patients < 1year who underwent CPB for correction of congenital cardiac defect(s). The n=775 total patients, were divided into Group A – Pre ICH Protocol (n=379) and Group B – Post ICH Protocol (n=396). Data review includes baseline AT levels, Thrombate III administration, loading dose heparin (LDH), CPB heparin, Baseline ACT, CPB ACT, platelet count (pre-during-post CPB), intra-op & post-op blood product usage (RBC, FFP, PLT, cell saver) and indicators of neurological complications post-operatively. Exclusion criteria: children >1year at date of surgery, non-bypass cases, ECMO in the pre/post op period.

Results: We expect to find a reduction in heparin administered to patients in Group B and a more controlled ACT value on CPB with ability to return to baseline post operatively. As a result of these expected findings, we expect to also see a reduction of intracranial hemorrhage post operatively. Preliminary results: n=28<30days 1/2020-2/2020 9/2022-11/2022 CPB Hep units (low) 3000 500 CPB Hep units (high) 13500 6900 Weight kg (low) 3 2.2 Weight kg (high) 8.9 9 Avg CPB Hep units 6066 2573

Conclusions: Our institution recognized an opportunity to improve post-operative recovery in our neonatal/infant population. We theorized the implementation of an ICH Protocol would result in the reduction of heparin administration in patients <30 days undergoing CPB and a subsequent reduction in complications from intracranial hemorrhage. Preliminary review of data before and after implementation of ICH Protocol demonstrate a significant heparin reduction. The next phase of data review evaluates post-operative complications including ACT return to baselines post protamine and an expected reduction in complications from intracranial hemorrhage in patients <30 days old who underwent CPB.

Friday 6

Transforming The Future Of Dialysis Therapies In Cardiac Intensive Care

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Background: Prior to November 2022, Lurie Children's Hospital operated inpatient dialysis via a contracted third party. As the complexity of the Cardiac ICU has increased, so has the need for dialysis therapies. To meet the evolving needs of this patient population, we developed an in-house dialysis program with in-house, twenty-four seven care delivery model. The transition allows for improved collaborative model between Nephrology and Cardiac ICU, enabling patients to receive treatment in a timely manner and to receive the newest forms of dialysis. Over 1000 CRRT therapy days are conducted yearly at Lurie Children's Hospital. This volume calls for a comprehensive inpatient dialysis team.

Methods: The project design included the conclusion of the contracted third-party dialysis team and the creation of a comprehensive internal nursing dialysis team. The development of an internal team allows for quick mobilization of resources to meet dialysis needs in a timely fashion.

Results: The twenty- four seven inpatient dialysis team at Lurie Children's Hospital is changing the dynamics of dialysis treatment in the Cardiac ICU. The team is meeting the ever-changing demands of the Cardiac ICU population in a timely manner. Start times for dialysis initiations have decreased from four hours to one hour. Bedside teams are now supported. When problems arise the ICU teams are no longer left at the mercy of a hotline number or to contracted teams with limited pediatric training. To better treat kidney injuries, dialysis modalities have expanded. Aquapheresis and Carpedeium have been added to the dialysis repertoire and HD and CRRT machines have been updated. Presence and availability are key in the Cardiac ICU. As patients recover, physical therapies are no longer interrupted as dialysis treatments can be completed during nighttime hours. Often, the vasculature of the cardiac population leaves obtaining access to be a challenge. With this new model, dialysis can be completed through two access points instead of one.

Conclusions: The future is bright for kidney injury treatments on the Cardiac ICU at Lurie Children's. The new dialysis team eliminates previous restraints that faltered patient's success. Current state, the Cardiac ICU utilizes current technology led by a knowledgeable team to advance patient outcomes. The robust cardiac program at Lurie Children's cares for complex patients requiring a robust dialysis team. The dialysis program and cardiac program now have the support from Lurie Children's Hospital to provide quality improvement and publish data.

Friday 7

Improved Clinical Outcomes With The Use Of An Algorithmic Approach To The Diagnosis And Treatment Of Diaphragm Dysfunction

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Background: Diaphragm dysfunction (DD) due to phrenic nerve injury is a well-known complication following pediatric cardiac surgery. DD is associated with prolonged mechanical ventilation, extubation failure, tracheostomy placement, and increased length of stay. Diaphragm plication is a surgical option to facilitate improved respiratory dynamics with wide variation in patient selection and timing across institutions. The fundamental challenge remains in identifying patients that would have an improved clinical course after accounting for the morbidity of this additional operation juxtaposed with reality that a subset of patients with DD will have an adequate postoperative recovery without plication.

Methods: An algorithm for the diagnosis and treatment of DD was implemented at a single congenital heart surgery program with a pathway toward early plication in patients under 6 months of age. All postoperative patients with concern for DD over a 4.5 year span (7/2018-12/2022) were screened and enrolled as appropriate. Patient characteristics (age, lesion, surgery) and clinical course (number of failed extubations, mechanical ventilator days, plication POD, tracheostomy, length of stay, and mortality) were obtained. This data was compared with 2.5 year historical cohort of patients with DD prior to implementation of the algorithm.

Results: The post-intervention (algorithm) group included 25 patients. 13 plications were performed. There were two mortalities (one with plication, one without plication) and one patient that required home trach/vent. Of the remaining 11 patients, the median day of plication was POD 10, number of failed extubations was 1, ventilator days was 12 days, and length of stay was 49 days. The historical cohort identified 13 patients. 11 diaphragm plications were performed. There was one mortality and one patient that required home trach/vent. Of the remaining 9 patients, the median day of plication was POD 21, number of failed extubations was 2, ventilator days was 20 days, and length of stay was 58 days. There were 11 patients captured by the algorithm who did not ultimately have surgical plication. 7/11 (64%) of these patients had diagnostic studies showing diaphragm paresis rather than paralysis. They had a median of 0 failed extubations, 7 ventilator days, and 26 day LOS. Compliance with the algorithm was moderate with most deviations being related to lack of screening ultrasound and late plication in neonatal patients.

Conclusions: Use of a standardized algorithm for the diagnosis and treatment of diaphragm dysfunction has resulted in improved clinical outcomes in this single-center study. There were overall decreases in extubation failure rates, ventilator days, and hospital LOS noted in the post-intervention group. There was also a significantly shorter time from index operation to surgical plication in the post-intervention group, possibly as a result of the forcing factor of encouraging compliance with the algorithm. These results suggest that a standardized algorithm may allow for earlier recognition and improvement in clinical outcomes for patients suffering from DD. However, given the relative rarity of this complication, a multi-center collaborative approach is necessary to determine the optimal diagnostic and treatment strategies.

Friday 8

Early Impairment Of Cerebral Bioenergetics After Cardiopulmonary Bypass In Neonatal Swine

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Background: We previously demonstrated cerebral mitochondrial dysfunction immediately following a period of full-flow cardiopulmonary bypass (CPB). The extent to which this dysfunction persists in the postoperative period, and its correlation with other markers of cerebral bioenergetic failure and injury is unknown. We developed a high-fidelity neonatal swine model to investigate the early evolution of mitochondrial function and cerebral bioenergetic failure after CPB.

Methods: Twenty piglets (mean weight 4.4 ± 0.5 kg) underwent 3 hours of CPB at 34°C via cervical cannulation, and were followed for 8, 12, 18, or 24 hours ($n=5$ per group). Markers of brain tissue damage (glycerol) and bioenergetic dysfunction (lactate to pyruvate ratio) were continuously measured in cerebral microdialysate samples. Control animals ($n=3$, mean weight 4.1 ± 1.2 kg) did not undergo cannulation or CPB. Brain tissue was extracted immediately after euthanasia for ex-vivo cortical mitochondrial respiration and frequency of cortical microglial nodules via neuropathology. Microglial nodules are indicative of micro-infarcts in the examined tissue.

Results: Both the lactate to pyruvate ratio ($p < 0.0001$), and glycerol levels ($p = 0.01$) increased in cerebral microdialysate within 8 hours after CPB. At 24 hours post-CPB, cortical mitochondrial respiration was significantly decreased compared to controls ($p = 0.046$). Microglial nodule formation was positively correlated with survival duration ($p = 0.01$, $R^2 = 0.9$).

Conclusions: Use of continuous CPB results in impaired cerebral bioenergetics which persists for at least 24 hours after CPB. The relationship of these metabolic alterations to later neurodevelopmental outcomes is not yet known. However, these findings suggest that there is a post-CPB period of bioenergetic impairment. During this period, the brain is primed for injury in the event of further alterations in metabolic delivery or demand, e.g. hemodynamic instability, decreased cardiac output, hypoglycemia, seizures, and decreased cerebral blood flow.

Friday 9

Adult Congenital Heart Disease. Surgical Outcomes And Characteristics In A Middle-Income Country

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Background: In Colombia Pediatric cardiac surgery started in 1948. Currently, more than 90% of children with congenital heart disease undergoing surgery survive to adulthood. Actually, more than 103.000 adults patients including those that reach adulthood without treatment may live in the country and required specialized centers. This is a growing population that currently exceeds the expectations of any health system that must be modified to serve this population. The objective of this review is to evaluate the results of surgical outcomes of these patients to help the ACHD program to create a rational projection.

Methods: Retrospective cohort of patients ≥ 18 years underwent surgical repair for congenital heart disease procedures were performed between January 2004 and November 2022, aortic valve replacement for bicuspid valve disease were excluded. A descriptive analysis was performed of the preoperative and intraoperative variables, postoperative outcomes, continuous variables are expressed as mean \pm standard deviation or median with interquartile range according to the result the type of distribution gave it by Shapiro – Wilk Normality test, categorical variables are presented as absolute frequencies and proportions.

Results: In 19 years of experience, we identified 657 procedures in patients >18 years who underwent surgical repair for CHD, these represent 7.5% of total 8730 procedures. Median age was 30 years, 15% were reoperations. Surgical risk RACHS-1 classification 1 and 2 were more frequent 85.4%. surgical mortality was 2%. The most common procedure after ASD closure was, pulmonary valve replacement in 7.1% of the patients.

Conclusions: The results of the treatment of these patients are very satisfactory. Knowing the status of our patients and their growth in number allows us to make projections for the Adult Congenital Heart program in the present and future, especially to create a culture in the country of referring patients to specialized centers. Since the pulmonary valve replacement after tetralogy of Fallot repair was the most common procedure after atrial septal defect closure, surgical techniques should be reviewed and closer follow up must be implemented for these patients.

Friday 10

Immediate Result Of Surgical Repair Of Complete Atrioventricular Canal After 9 Months Of Age At High Altitude.

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Background: Complete AV canal represent 5% of congenital heart disease and are more common in Down syndrome. Surgical management before 6 months as ideal. This scenario is not common in developing countries, where access to surgery is limited and late and if the majority live at high altitude.

Methods: To compare the short-term postoperative outcomes of surgical repair of the complete AV canal (CAVC) performed early versus late, two groups were defined (2) according to the age at which the surgical treatment was performed, <9 months vs ≥9 months. months, all operated at an altitude above 2,640 meters above sea level. All CAVC operated between January 2012 and November 2022 were included. A descriptive analysis of preoperative, intraoperative, and postoperative results was performed. To determine the difference between groups, the Chi-square test or Fisher's exact test and the Wilcoxon-Mann-Whitney test for non-parametric continuous variables were used.

Results: We identified 125 CAVC patients and all were operated with a double patch technique, and small atrial septal defect was left in patients with severe preoperative pulmonary hypertension. Nitric oxide, and milrinone were used in those with severe pulmonary hypertension. Median age was 8 months. In ≥9 months group preoperative oxygen saturation % was lower, median 89% vs 90%, malnutrition was higher 70% vs 64%. Chromosomal abnormalities were present in 81%, 8% had previous cardiac surgery higher in ≥9 months group 16.4% vs 1.4% ($p=0.006$), 10.8% had severe but reactive pulmonary hypertension. There were no statistical differences on intra or postoperative characteristics, overall surgical mortality was 4%, higher in ≥9 months group 5.4%.

Conclusions: Late repair of the complete atrioventricular canal after 9 months of age was not associated with a higher incidence of perioperative adverse outcomes compared to those patients who underwent early surgery before 9 months of age. The prognosis is favorable even in the presence of severe reactive pulmonary hypertension and high altitude affects could be a protector factor that has to be analyzed in a long term condition and in a prospective study.

Friday 11

Post-Natal Outcomes In Fetal Echocardiographic Findings Of Isolated Ascending Aorta Dilation

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Background: Ascending Aorta dilation (AAD) is an important finding postnatally due to its association with connective tissue disease and bicuspid aortic valve (BAV). Prenatal isolated AAD can be identified on fetal echocardiogram (echo); however, paucity of outcomes data makes prenatal counseling challenging. We report the largest single center cohort of isolated fetal AAD and outcomes.

Methods: Retrospective review of our institution's prenatal database (2013-2022) identified fetuses with isolated AAD. Those with complex cardiac abnormalities were excluded. Ascending aorta diameter Z-scores for gestational age (GA) were determined from three published standards. Postnatal Z-scores were determined from Boston Children's Hospital and Pediatric Heart Network standards.

Results: Of 7320 pregnancies, 31 fetuses (0.4%) had isolated AAD on fetal echo. Mean GA at diagnosis was 22.4 \pm 3.2 weeks with Z-scores of +2.76 (\pm 0.43), +2.41 (\pm 0.67), & +2.59 (\pm 0.46) by the Krishnan, McElhinney, and Schneider standards, respectively. Postnatal echo demonstrated AAD in eight of 29 (28%), isolated distal ascending aneurysm in one, and BAV in one. Family history was significant for aortopathy in one, and BAV in two. On the most recent echo (mean age 28.5 \pm 19.0 months), 7 patients had persistent AAD, as one patient had resolution of AAD by 6 months.

Conclusions: Of fetuses with isolated AAD, 28% had persistent AAD postnatally with a low incidence of BAV. Follow-up postnatal echo should be considered in isolated fetal AAD.

Friday 12

Echocardiographic Characteristics Of Neonates With Atrial Septal Defects

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Background: Atrial septal defect (ASD) is among the most common congenital heart defects. The left-to-right shunt will over time cause dilatation of the right atrium and right ventricle and pulmonary hyperperfusion. Due to the natural history and clinical presentation, the detection of ASDs often does not occur until later in childhood or adulthood. By the time of diagnosis, several secondary structural and functional changes in the heart have occurred. Little is known about cardiac structure and function in ASD patients very early in life. In this study we aim to investigate echocardiographic characteristics in a large cohort of neonates with ASD.

Methods: We analyzed neonatal echocardiograms from the Copenhagen Baby Heart Study (CBHS); a multicenter, population-based cohort study with prenatal inclusion of 27,595 neonates. For this substudy we included 716 neonates with secundum type ASDs and matched them 1:1 on sex and age at examination with neonates without ASD from the same birth cohort. We compared echocardiographic characteristics for neonates with ASD with neonates without ASD. For comparison between groups, we conducted Student's two-sample t-test. We corrected for multiple testing using the Bonferroni method and p-values < 0.003 were considered statistically significant.

Results: Neonates with an ASD (median age 11 days, 52% female) had larger right ventricular (RV) dimensions than matched controls (RV length: 28 mm vs. 27 mm, $p < 0.001$; RV basal diameter: 15 mm vs. 14 mm, $p < 0.001$ and right ventricular outflow tract diameter 14 mm vs. 12 mm, $p < 0.001$). Atrial volumes were higher in neonates with ASD than in controls (right atrial end-systolic volume: 2.9 ml vs. 2.1 ml, $p < 0.001$ and left atrial end-systolic volume 2.0 ml vs. 1.8 ml, $p < 0.001$). Tricuspid annular plane systolic excursion was larger in neonates with ASD than in controls (10 mm vs. 9.6 mm, $p < 0.001$). Left ventricular systolic and diastolic dimensions did not differ between neonates with ASD and controls, neither did the diameter of the inferior vena cava. We found a tendency of left ventricular outflow tract diameter being smaller and main pulmonary artery diameter being larger in neonates with ASD compared to controls, though not reaching statistical significance after correction with the Bonferroni method.

Conclusions: ASDs are associated with altered cardiac dimensions from the neonatal phase. Compared to controls, neonates with ASD have larger right ventricular dimensions and larger atrial volumes when examined with echocardiography within the first 30 days after birth.

Friday 13

Right And Left Ventricular Mass Development In Early Infancy: Correlation Of Electrocardiographic Changes With Echocardiographic Measurements

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Background: In term infants the RV fraction of the total ventricular weight by direct measurement decreases from 44% at birth to 28% by 4 months. During this time, using 2-D echocardiographic estimates, the RV mass indexed to body surface area (RVMI) decreases by 17% and the LV mass index (LVMI) increases by 24%. Additionally, the RV:LV mass ratio decreases by 33% from 0.64 down to 0.43. The evolution in electrocardiogram (ECG) tracings during this rapid transition in early infancy have been detailed previously. However, correlations between these extensive ventricular mass variations and simultaneous ECG changes have not been defined.

Methods: Forty-five normal full-term infants were enrolled and prospectively evaluated at 2 days and at their 2-week, 2-month, and 4-month visits with ECG and echocardiogram. QRS voltages were measured in leads V1, V6, I and aVF. Two-dimensional echocardiographic formulas validated against magnetic resonance imaging were used for the estimation of ventricular mass. LV mass was estimated with the area-length method. RV free-wall mass was estimated with the one-quarter prolate ellipsoid shell formula: $RV\ mass = 5.84\ (end-diastolic\ RV\ cavity\ area)\ (end-diastolic\ RV\ free-wall\ thickness) + 1.0$, where RV cavity area was measured in the apical 4-chamber view.

Results: As RVMI decreased from mean 28.1 (95%CI 27.1, 29.1) to 23.3 g/m² (95%CI 21.4, 25.2), so did V1R and V6S voltages. RVMI changes correlated with V1R, V6S and V1R+V6S voltages ($R = 0.29^*$, 0.23^+ and 0.35^* , respectively. $*p < 0.01$, $^+p < 0.05$). As LVMI increased from 44.6 (95%CI 42.9, 46.3) to 55.4 g/m² (95%CI 52.3, 58.5) V6R increased, but V1S voltage decreased slightly. LVMI changes correlated with voltage changes in V6R and less so with V6R+V1S ($R = 0.31^*$ and 0.23^+ , respectively. $*p < 0.01$, $^+p < 0.05$) due to a lack of correlation with V1S ($R = 0.02$, $p = NS$). Mean QRS axis shifted from 135 (95%CI 124, 146) to 65 degrees (95%CI 49, 81) and correlated with both RVMI decrease and LVMI increase ($R = 0.44^*$ vs. 0.20^+ , respectively. $*p < 0.01$, $^+p < 0.05$). The R/S voltage ratio in V1 and even that in V6 did not correlate significantly to RVMI or LVMI, respectively. Net voltages V6R-S and V6(Q+R)-S were noted to have slightly higher correlation coefficients ($R = 0.34$ and 0.38 , respectively, $p < 0.01$ for both) with LVMI than V6R alone.

Conclusions: In conclusion, our study showed that the RVMI decrease in early infancy correlated most significantly with the sum of V1R + V6S voltages and less so with each individually. The LVMI increase correlated better with V6R voltage alone and less so with the sum of V6R + V1S voltages, since V1S voltage did not increase during early infancy. Both ventricular mass alterations correlated with the QRS axis shift. The lack of correlation by V1S voltage and the R/S voltage ratio in V6 with LVMI, and the similar lack of correlation by the R/S voltage ratio in V1 with RVMI, should compel us to be hesitant to use these criteria for determination of ventricular hypertrophy in early infancy. The net V6(Q+R)-S voltage, uniquely evaluated in this study, could be further investigated as an improved criterion for diagnosing LV hypertrophy.

Friday 14

Utility Of Pulmonary Venous Doppler To Identify Pulmonary Over-Circulation In Patients With Congenital Heart Defects

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Background: Non-invasive assessment of pulmonary over-circulation in children with congenital heart disease (CHD) and hemodynamically significant intracardiac shunt (HSIS) is challenging due to a lack of reliable and reproducible tools. Echocardiographic assessment of the pulmonary venous Doppler is performed routinely in all patients with CHD. Increased pulmonary vein flow is seen in patients with pulmonary over-circulation; however, due to a lack of data, its utility to evaluate over-circulation is unknown. This study aimed to evaluate the various characteristics of pulmonary venous Doppler in children with HSIS pre- and post-surgical repair to analyze its diagnostic utility in detecting pulmonary over-circulation.

Methods: We retrospectively evaluated echocardiographic data of children with CHD who underwent repair for HSIS in last four years at The Children's Hospital of San Antonio. Patients with HSIS lesions (identified by echocardiograms, magnetic resonance imaging, or cardiac catheterization), such as atrial or ventricular septal defects, atrioventricular canal, or patent ductus arteriosus, were included in the study. Pulmonary vein Dopplers were abstracted from the apical four-chamber and suprasternal echocardiographic views. Characteristics collected included systolic and diastolic peak velocities, as well as mean velocities and gradients, pre- and post-surgical repair. All statistical analysis was performed on IBM SPSS 27 statistical software.

Results: Ninety-three subjects (N=93) were identified with median age of 1.1 years (0.37,3.5). Of these, 43 patients had HSIS and echocardiographic data available before and after surgical repair. Median peak pulmonary vein velocity in patients with HSIS was 60 cm/s (53,72) and post repair was 50 cm/s (44,59), $p < 0.001$. The median gradient prior to surgical repair was 0.52 mmHg (0.4,0.73) and post repair was 0.32 mmHg (0.24,0.45), $p < 0.001$. Receiver operating characteristic (ROC) analysis was utilized on various pulmonary vein Doppler parameters to obtain cut-off values for identifying pulmonary over-circulation. The best ROC curves were obtained for right pulmonary vein mean gradients and peak velocities obtained from a four-chamber view. For mean gradient, the area under the curve was 0.88 and the cut-off was 0.4 mmHg with a corresponding sensitivity of 80% and a false positive rate of 12%. For peak velocity, the area under the curve was 0.85 and the cut-off was 57 cm/s with a corresponding sensitivity of 82% and a false positive rate of 20%.

Conclusions: In this small cohort of CHD patients, pulmonary venous mean gradient above 0.4 mmHg and peak velocity of 57 cm/s can be used to identify pulmonary over-circulation in patients with hemodynamically significant intracardiac shunts. Further studies with larger sample sizes can help further define cut-off values.

Friday 15

Selection Of Atrial Septal Occluder Using 3D Tee Versus Stop Flow Balloon Sizing

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Background: Device closure of atrial septal defects (ASD) is the treatment of choice for patients with suitable anatomy. Historically, stop flow balloon sizing is performed to select device. The Gore Cardioform Atrial Septal Occluder (ASDO) has an anatomically adaptable waist with a 7mm range of diameters for each of the 5 available device sizes. In this study we evaluate the relationship between ASD diameter measured by 2D and 3D TEE versus stop flow balloon sizing. We tested the hypothesis that ASDO selection based on TEE measurements would be the same as that achieved by stop flow.

Methods: 17 patients undergoing ASD closure had 2D TEE, 3D TEE, and stop flow balloon sizing performed. 2D TEE measurements included maximum diameter, diameter orthogonal to the maximum, and stop flow balloon sizing diameter. 3D measurements included maximum diameter, diameter orthogonal to the maximum, area, and circumference. Fluoroscopic stop flow diameter was collected as the largest measured waist diameter obtained on fluoroscopy. The device chosen was also reviewed.

Results: Fourteen patients received ASDO (27 – 48mm) and 3 patients received GORE Cardioform Septal Occluder (25 and 30mm). The average maximum 2D and 3D diameters were 13.1mm and 13.8mm respectively. The average SF diameter by fluoroscopy and echocardiography were 15.3mm and 15.1mm with strong agreement (ICC 0.84, $p < 0.001$). Fluoroscopic SF diameter served as the gold standard. There was good correlation between the maximum 2D and SF diameters (ICC 0.77, $p = 0.007$), with SF being on average 2.2mm (21%) larger. There was increased correlation between the maximum 3D and SF diameters (ICC 0.80, $p < 0.001$), with SF being on average 1.5mm (13%) larger. Diameters calculated from 3D circumference and area displayed only fair correlation with SF (ICC = 0.65 & ICC = 0.62). The measure most strongly correlated with SF diameter was the maximum diameter obtained by either 2D or 3D (ICC = 0.82, $p < 0.001$). Selecting an ASDO based on the maximum TEE diameter would have resulted in the same device as SF sizing in 15/17 patients. In the other two cases the maximum TEE diameter was larger than the SF diameter, and a larger device was implanted than would have been suggested by the SF diameter.

Conclusions: Maximum native diameter alone can be used to select the appropriate GORE ASDO for device closure in most cases. Compared to 2D TEE, 3D TEE allows easier identification of the maximum diameter, and increases the correlation between maximum native and stop flow diameters.

Friday 16

Left Ventricular Dysfunction In Guatemalan Pediatric Patients After Patent Ductus Arteriosus Closure

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Background: Patent ductus arteriosus (PDA) is one of the most common congenital heart defects (CHD), creating a left-to-right shunt resulting in left heart volume overload. PDA closure via surgical ligation (SL) or transcatheter device closure (TDC) corrects the shunt; however, the hemodynamic consequences can lead to immediate left ventricular (LV) dysfunction, ranging from mild transient dysfunction to acute cardiopulmonary instability. While the literature between PDA closure and immediate LV dysfunction is growing in high-income countries, there is little work in low- and middle-income countries. Therefore, we sought to characterize this relationship at the Unidad de Cirugía Cardiovascular de Guatemala (UNICAR).

Methods: All patients <12 years old who were admitted for PDA closure between July 2020 and March 2022 were considered in this cross-sectional study. All patients had before (baseline) and after-closure (within 24 hours of PDA closure) transthoracic echocardiograms to assess cardiac function. Baseline measures included: 1) LV end-diastolic diameter (EDD), 2) LV ejection fraction (EF) via M-Mode, and 3) LV fractional area change (FAC). After-closure measures included: 1) LVEDD, 2) LVEF, 3) LVFAC, 4) LV strain, 5) biplanar LVEF with Simpson Method, and 6) LV diastolic function via Mitral E/A ratio and tissue doppler.

Results: Each patient was assessed in a multidisciplinary meeting to determine treatment modality. Patients with other concomitant CHD or residual defects after PDA closure were excluded. A total of 93 patients met criteria to be included in the analysis, of which 78 underwent SL and 15 TDC. Regarding echocardiogram measures, LV dilation was defined as $Z > 2$, and LV dysfunction was characterized by an EF < 55% and/or FAC < 28%. LV diastolic dysfunction was characterized as an inversion of mitral E/A ratio. Baseline echocardiograms demonstrated higher rates of LV dilation but similar rates of LV dysfunction in patients who underwent SL vs TDC. After-closure echocardiograms showed higher rates of both LV dilation and dysfunction in the former compared with the latter. In comparison with normal baseline LVEDD, pre-closure LV dilation was associated with higher rates of LV dysfunction measured by LVEF (26.4% vs 10.0%, $p=0.047$), LVFAC (28.3% vs 10.0%, $p=0.030$), Strain (89.5% vs 51.7%, $p<0.001$), and biplanar LVEF via Simpson (50.0% vs 11.4%, $p<0.001$). Large (≥ 4 mm) PDAs, as opposed to moderate (size <4mm), were associated with higher rates of LV dysfunction measured by LVEF (27.3% vs 0%, $p=0.003$), LVFAC (27.3% vs 3.7%, $p=0.011$), and biplanar LVEF (41.7% vs 9.5%, $p=0.007$).

Conclusions: Our results suggest that patients selected for surgery were more likely to have pre-closure LV dilation, and subsequently had higher rates of after-closure LV dilation and dysfunction, compared with patients treated percutaneously. Furthermore, baseline LV dilation and large PDA size were associated with post-closure LV dysfunction regardless of PDA closure method. We recommend that baseline cardiac measures should be closely evaluated to identify patients who are at greater risk of immediate after-closure LV dysfunction.

Friday 17

Challenging The Specificity Of Echocardiographic Assessment Of Coronary Artery Dilatation For Kawasaki Disease

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Background: In Kawasaki Disease (KD) management, determination of coronary artery dilatation by echocardiography guides acute interventions and long-term antithrombotic strategy. This measurement is interpreted as a Z-score (standardized by age and body surface area) based on representative pediatric cohorts. Here, we review a case highlighting the necessity of clarifying the role of this measure within the workup of childhood febrile illness.

Methods: A previously healthy 13-year-old girl presented with an 8-day history of intermittent fevers and 3-day history of bilateral conjunctivitis. She had sufficient involvement in social and outdoor activities to consider communicable and zoonotic infections. On admission, her physical exam was notable for fever (39.9C), tachycardia (110 bpm), bilateral conjunctival injection, unilateral cervical lymphadenopathy, and a diffuse, blanchable, maculopapular rash extending from her trunk to extremities (including palms) without desquamation or edema. Initial blood tests were significant for borderline hyponatremia (133mEq/L), hypoalbuminemia (3.2g/dL), transaminitis (AST: 199units/L; ALT: 205units/L), mild leukopenia (4.3 thousand/mm³), borderline thrombocytopenia (166 thousand/mm³), and elevated inflammatory markers (CRP: 8.1mg/dL; ESR: 8mm/hr; Procalcitonin: 1.49ng/mL). Incomplete KD was our initial leading diagnosis, as she met 3 (rash, conjunctival injection, lymphadenopathy) of the clinical criteria and 2 (elevated CRP/ALT) of the supplemental laboratory criteria. Our differential also included tick-borne illness, MIS-C, and viral infections (adenovirus/parvovirus/EBV/CMV). Following blood draws for cultures and serology panels, aspirin and IVIG were initiated. Within the first 12 hours following completion of IVIG infusion, the patient's conjunctivitis and rash resolved. Echocardiography during this period found diffuse RCA dilatation (4.1 mm, Boston model Z-score: 2.66), classified as a small aneurysm. During the following 12 hours, a brief presentation of light erythematous palmar macules prompted the initiation of doxycycline (picture available). She continued to have intermittent fevers more than 24 hours after completing the first IVIG infusion. Given the echocardiogram findings, this was most concerning for IVIG-refractory KD. A second IVIG infusion was administered. The following day, serologic testing drawn on admission confirmed infection with Rickettsia typhi. Notably, she developed severe hemolytic anemia following the second IVIG infusion, significantly prolonging her hospitalization. Just over 3 weeks following her initial assessment, repeat echocardiogram continued to demonstrate small RCA aneurysm (4.0mm, Boston model Z-score: 2.57).

Results: This case suggests potential vulnerability in how coronary artery dilatation is currently interpreted. Per expert consensus from the most recent (2017) AHA KD guidelines, if criteria for incomplete KD are met, positive echocardiogram findings (LAD or RCA Z-score ≥ 2.5) are highly specific for KD and supports treatment initiation. The studies supporting this recommendation compared coronary artery Z-scores in children with various non-KD causes of prolonged febrile illness (N=88) to Z-scores of children diagnosed with KD (N=289). Our patient's unique echocardiographic findings might also represent overlapping pathophysiology. Rickettsia species damage endothelial cells and have been previously linked to myocardial and cerebrovascular infarcts, further suggesting parallels with the thrombotic vasculitis of KD.

Conclusions: This case report highlights a paucity of knowledge regarding both the driving mechanisms and clinical incidence of coronary artery dilatation in non-KD febrile illnesses, particularly in Rickettsial disease.

Friday 18

Stop And Go Simulation: An Innovative Approach To Nurse Training

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Background: The Pediatric Cardiac Intensive Care Unit (PCICU) is a complex setting comprised of high acuity patients with risks of rapid deterioration, requiring exquisite recognition and skill of the care team. Between 2021-2022, 83% of the new hires into the PCICU were graduate nurses, with minimal clinical experience that lack the knowledge to triage emergent situations. Stop & Go (S&G) Simulation is an innovative approach to mock-code type training, allowing new PCICU nurses to build knowledge, comfort, and critical thinking skills for navigating events involving patient deterioration.

Methods: S&G Simulations are high-fidelity training sessions, comprised of 4 patient scenarios. The simulation team is comprised of a multidisciplinary group led by highly skilled staff nurses. A group of 5-6 new nurses are briefed on the simulation process and are assigned a participation role. Each nurse will experience every role in each simulation. Throughout the active scenario, the simulation team pauses at intervals to review and ensure participant knowledge acquisition about the individual steps involved. A debriefing session is held at the conclusion of each situation.

Results: 98% of participants (n=41) surveyed felt the sessions were high quality, 100% reported a judgement-free environment, 37% expanded their knowledge of code roles, 24% learned management of pediatric code medications, 22% learned how to better recognize the patient deteriorating prior to a code, 20% learned various arrhythmia treatments, 17% learned how to use a defibrillator including cardioversion, 15% learned to perform better compressions, 15% learned proper bagging mechanics, and 10% stated they were more confident and less stressed about a code situation.

Conclusions: PCICU nurses practice in an environment where stakes are high for patients, and advancing nurses' knowledge and skill are essential for optimal outcomes. S&G Simulations allow for a tailoring of knowledge delivery to meet the unique needs of nurses beginning their career in a complex care environment. It will be imperative to expand training to include more scheduled opportunities and more tenured staff.

Friday 19

Collaboration Between Two Advanced Practice Nurse Teams Caring For Pulmonary Vein Stenosis Patients Awaiting For Lung Transplant

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Background: This poster will focus on advanced practice register nurse (APRN)-led care coordination and collaboration between two medical specialty teams. The poster will describe approaches to improved care continuity for pediatric patients with PVS awaiting lung transplant with high medical complexity provided by both, cardiology and pulmonary teams.

Methods: Poster highlights the APRN role within cardiology and pulmonary teams to demonstrate APRN collaboration to provide individualized care to PVS patients and their families. Patients with aggressive disease poorly responsive to medications, catheterizations and/or disproportionate pulmonary hypertension are encouraged to meet with the lung transplant coordinator to discuss lung transplant and evaluation. If family elects to proceed, and transplant is an option, the patient will undergo transplant evaluation. The evaluation process involves meetings with multidisciplinary transplant team members, diagnostic testing, and subspecialty involvement as needed that will help determine potential barriers to lung transplant candidacy.

Results: To ensure the patient remains a transplant candidate from both a medical and psychosocial standpoint communication between the APRNs is the main mode of communication between the two teams. Attempts are made for coordination of outpatient appointments, blood draws and consistent messaging between teams. Family meetings with both teams is common practice during patient hospitalization. Wait time, particularly for infants, can be many months and due to aggressiveness of disease these patients require cardiac catheterization every four weeks. Many younger patients medically decompensate prior to lung transplant and don't survive. Due to PVS being a lethal disease, some patients have been transferred to await lung transplant at the local acute medical rehabilitation hospital. The APRN team collaboration has provided seamless medical care transitioning from one setting to another so the patient does not have interruption of or a setback in care.

Conclusions: Some PVS patients have aggressive disease that require consideration for transplant. Collaboration between the two APRN teams provide seamless continuity of care and provides incredible support for these families through the process of listing and while awaiting transplant. Collaboration between the two medical specialty teams improved care continuity for pediatric patients with PVS awaiting lung transplant with high medical complexity.

Friday 20

Whose Service Should They Be On: Evaluating Different Approaches For Cardiac Patients Being Admitted With Non-Cardiac Issues To Children's Hospitals

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The Children's Hospital of The King's Daughters

Background: Patients with congenital heart disease (CHD) presenting with non-cardiac complaints are often admitted to the cardiology service. Presumptions often include that their underlying cardiac anomaly is exacerbating their current non-cardiac symptoms or that this non-cardiac illness might worsen the patient's hemodynamics or other heart-related complications. Additionally, non-cardiology providers often feel uncomfortable caring for patient with CHD. This can lead to delays in initiation of appropriate treatment and suboptimal patient outcomes. To further understand policies and procedures for admitting CHD patients with non-cardiac issues, we surveyed pediatric hospitals nationwide to help elucidate how admitting decisions are made for these patients.

Methods: All hospitals within the United States where pediatric cardiac surgery is performed were eligible for participation. A survey was designed to identify if participating institutions had a cardiology fellowship and access to telemetry on non-intensive care unit (ICU) floors. Survey was distributed via PediHeartNet. Respondents were asked to comment on what factors influence their decision to admit this subset of patients to the cardiac service, particularly how stage of CHD contributes to their decision-making. Survey was approved by the Eastern Virginia Medical School institutional review board (EVMS IRB #20-12-XX-0269) prior to the start of data collection.

Results: We received a total of 33 survey responses from 29 institutions. Of those, 26 have cardiology services that admit their cardiac patients. Amongst those institutions with cardiac admitting services, 82% of respondents have telemetry on non-ICU units and 73% had a pediatric cardiology fellowship program. Only 33% of respondents have a formal protocol to help guide the admissions process for these patients. Only a single respondent expressed a willingness to admit a stage I (shunted) single ventricle patient to a hospitalist team whereas 33% of respondents felt comfortable in admitting single ventricles post-stage II (Glenn/superior cavopulmonary anastomosis) to a hospitalist service. Similarly, 33% also expressed comfort with admitting post-Fontan (total cavopulmonary connection) patients to the hospitalist service. All respondents felt that patients must be deemed hemodynamically stable for admission to a non-cardiac service. Most respondents stated they were most comfortable admitting patients to non-cardiology services if there were no residual lesions or if they were temporally remote from surgical intervention. Limitations to admitting patients to other services included the lack of adequate telemetry monitoring and specially trained personnel familiar with the congenital cardiac anatomy or physiology, especially nursing staff.

Conclusions: Pediatric cardiac patients are not immune to non-cardiac illnesses that often fall out of the scope of practice for most cardiologists and for whom admission to non-cardiac teams/units may provide more immediate access to treat their current medical issue. However, there is a variable level of comfort in admitting these patients to non-cardiac services. Despite wide-spread access to telemetry on non-ICU units, most cardiologists prefer to admit patients with single-ventricle physiology or hemodynamic instability to a cardiac or ICU service. There is significantly more comfort admitting patients with no residual lesions or a longer time from surgical intervention to non-cardiac services. As demonstrated, there is heterogeneity in intuitional approaches and a lack of accepted uniform guidelines or protocols. A multidisciplinary, institutional-specific approach should be considered to meet the cardiac and non-cardiac needs of this complex population.

Friday 21

Creating A Multidisciplinary Model For Individualized Pediatric Cardiology Care For Patients With Specialized Needs

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Cincinnati Children's Hospital and Medical Center

Background: Multidisciplinary care has proven to be a satisfier for both patients and providers in addition to combining the expertise from multiple subspecialists to provide more comprehensive articulated care. In this model, clinicians collaborate to create an individualized care plan. Patients leave the clinic visit with a better idea of their next steps and care plan because each specialty involved integrated all elements of the care plan.

Methods: While the Heart Institute (HI) at Cincinnati Children's Hospital (CCHMC) has participated in multidisciplinary clinics in the past, in 2018 the HI opened its Multidisciplinary Fontan Clinic. This clinic began with five subspecialties involved and expanded to include 12 subspecialties. The multidisciplinary approach was designed to include a collective approach to clinical discussion and decision making prior to appointments, comprehensive research/registry inputs, and individualized care for patients. Since the start of the Fontan Clinic, three other multidisciplinary clinics were established and two more will launch in 2023. These clinics were developed using the same novel approach for comprehensive care.

Results: When patients are seen in a multidisciplinary clinic, they have a cost reduction as they are only paying one hospital bill. Patients can save up to \$1,860 per visit depending on the number of providers they would typically see in siloed visits. They also do not have to come to the hospital for multiple clinic visits, but rather, they can meet with multiple subspecialists at the same time and leave their appointment with a clear direction. Each clinic provides an overall health scorecard to the patient to ensure they leave with a clear consensus from the collective care team. The personalized care plans are possible because of the weekly interactions and patient reviews conducted by the clinical team. Pre-visit meetings are streamlined to deliver concise information, updated labs, and imaging to discuss the current state of the patients and create recommendations for treatment as a care team. There have been multiple instances where the care team was able to catch something that may have gone noticed otherwise if the clinic visits were completed in siloed specialties.

Conclusions: Multidisciplinary care is not a new concept, but the model the HI at CCHMC has created for developing a personalized approach in the multidisciplinary setting is a new model for care. Since 2018, the Fontan Clinic, Cardio-Pulmonary Sports Medicine Clinic, Tube-Wean Clinic, and Combined Heart-Liver Transplant Program evolved using the same model for multidisciplinary care. There are 2 other clinics currently under development using this same model. While each new clinic has a pediatric cardiology component, they also each include services from other specialties and serve patients using the same individualized approach to better patient care. Providers feel they are more engrained in the overall care of their patients and the patients can ask questions to all their physicians at once instead of during various appointments throughout the year. The pre-visit planning and patient scorecards have been essential in each of the new multidisciplinary clinics created in the HI.

Friday 22

The Evolution Of A Regional Joint Pediatric And Congenital Heart Program

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Background: Launched in 2017, Kentucky Children's Hospital (KCH) and the Heart Institute at Cincinnati Children's (HI CCHMC) set out to transform pediatric and adult congenital heart care in Kentucky. To do this, these institutions established the Joint Pediatric and Congenital Heart Program (JHP), a partnership to provide tertiary pediatric congenital cardiology surgical and clinical care, ensuring clinically appropriate care close to home.

Methods: Through a "one program, two sites" model, phase one of the JHP included common clinical protocols, standardized technologies, shared recruitment, and high-quality publicly reported results through intentional investments made in organization engagement, IT infrastructure and training. As the JHP has evolved, the foundations of the initial design are still in place today. Teams at both sites hold joint patient care conferences to review patient information and ensure care delivery at the most clinically appropriate site. Original Executive and Program Steering Committees are also ongoing as they act as platforms to discuss additional institution-to-institution relationships generated from the established JHP.

Results: The partnership's more recent evolution has resulted in additional important milestones. Both hospitals submit to the STS Congenital database independently but also receive an umbrella report, combining the outcomes data for a "one-program, two-sites" report. In 2019, for the first time, the two sites jointly submitted to USNews and World Report Best Children's Hospital for Pediatric Cardiology and CT Surgery and received a joint ranking. This occurred in subsequent years and will continue for future submissions. In early 2020, the surgeon who joined the JHP at the start of the program was recruited to another institution, valued for his experience in launching such an effort and its success to date. The outstanding outcomes achieved in the three years he was onsite assisted in the JHP's ability to recruit another world-renowned surgeon in May 2020. During 2020 and 2021, two JHP outpatient clinics in Kentucky were launched where clinical staff from both institutions provide care to patients together in the same setting. All marketing materials are dually branded. This approach allows the JHP to continue to expand services across the state. The joint marketing/branding signifies the partnership evolution and desire to jointly treat and care for the patients close to home.

Conclusions: The establishment of the JHP continues to ensure clinically appropriate care close to home for patients in Kentucky, while holding the program and its leadership accountable for superior outcomes. Senior leadership from both institutions have collaborated on performance metrics for the joint surgical program, with quarterly outcome updates provided by the team. This ensures transparency across sites related to those measures deemed critical to the program's success. The evolution of the program will continue with a desire to collaborate beyond heart care across institutions, specifically in the areas of research and quality improvement. For the JHP directly, additional outreach clinics and pediatric heart sub-specialties are planned based on best practices and the success of the current JHP clinics. Further, joint reporting will continue but may also expand to include combined reporting to registries.

Friday 23

Developing A Comprehensive Pediatric Cardio-Oncology Program: A Single Center Approach To Cardiovascular Care For Childhood Cancer Patients And Survivors

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Background: Cardiovascular complications represent a leading cause of morbidity and mortality in childhood cancer survivors. As the number of childhood cancer patients and survivors exposed to cardio-toxic therapies has increased, the need for the development of cardio-oncology programs has become recognized. Our principal purpose was to develop a multi-disciplinary comprehensive pediatric cardio-oncology program with the goal of early identification, prevention, and treatment of cancer related cardiotoxicity among childhood cancer patients and survivors.

Methods: A collaborative team of pediatric oncologists and cardiologists was established in 2020. Monthly meetings were conducted to determine the clinical needs, available resources, leadership, organization of the program as well as the clinic's flow. Other participants in the program include pediatric nurse practitioners, nurses, psychologists and dietitians. A protocol has been developed and refined to guide referral, screening, monitoring and treatment. Patient eligibility is determined by patient cancer history, cardiotoxic therapy exposure and cardiovascular risk profile. Multidisciplinary care is provided simultaneously by all specialties on the same clinic day. Patients evaluated in the cardio-oncology program are classified into 3 groups: Screening Population (cancer patients and survivors at risk of cardiotoxicity); Early Intervention (patients with evidence of subclinical cardiotoxicity) and Heart Failure Population (symptomatic patients or with more advanced cardiac dysfunction)

Results: Since its founding in October 2020 and despite challenges of the COVID-19 pandemic, monthly multidisciplinary clinic sessions have been held. The cardio-oncology program has grown from an initial patient population of 10 to 95 patients (63% male) mean age 15.4 ± 3.9 years. 76 patients are primarily followed in the outpatient multidisciplinary cardio-oncology survivorship clinic where our team follows a preventative approach to address cardiovascular risk factors (such as hypertension, hyperlipidemia, obesity, diabetes), provide screening for cardiotoxicity, and treatment if indicated. Of these patients, 16 (21%) have subclinical cardiomyopathy 5 of whom are currently receiving treatment with an ACE-Inhibitor. 19 patients are followed primarily in the heart failure cardio-oncology clinic due to either having more advanced cardiac disease and/or still undergoing cancer treatment. The team also works in collaboration with adult cardio-oncologists to facilitate a seamless transition of care of patients upon reaching the age of 18 years

Conclusions: An institutional, multi-disciplinary pediatric cardio-oncology program for childhood cancer patients and survivors has been successfully established to address the cardiovascular health needs of this high-risk growing population

Friday 24

Heart Center Performance Rounds : Team Learning In Complex, High Volume Environments

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Background: Excellence in congenital cardiac care requires team resilience, complex decision-making, systematic identification of threats, and analysis of errors to adjust practice. Medicine has learned from other high reliability organizations, but effective team communication and detection of near misses remains a frequent challenge. Here we describe the lessons learned from our institution's Heart Center Performance Rounds (HCPR) experience, which focuses on addressing these gaps, learning from success, outcomes transparency, and education on practice changes and improvement work.

Methods: Our HCPR is a weekly 60 minute review of one week of surgical and catheterization cases from two weeks prior. Each patient's course is reviewed against both internal and external benchmarks, such as length of stay (LOS) and markers of clinical progression. Through a triaging process, additional attention is devoted to cases that deviate from expected course, exemplify team resilience, encounter system challenges, or are valuable for team learning. Our approach underwent adjustments based on participant feedback and leadership priorities over the first year. The current iteration represents a strategic redesign, and has been stable for 14 months. We focus on psychological safety to promote open communication and trust, by employing a moderator and establishing discussion ground rules. This iteration was started during the COVID-19 pandemic, and we leveraged conferencing technology to assist with moderation and inclusion. A review committee meets weekly to identify actionable items and track trends in discussion. This committee then organizes methods for further investigation or follow up, and arranges monthly presentation back to the Heart Center.

Results: We measured improvements in psychological safety by monitoring conference participation. Median attendance was 108 persons per meeting. We noted an increase in participation over 14 months, from a median of 11 distinct voices per meeting in the first 10 weeks of this iteration, to 18 distinct voices per meeting over the last 10 weeks tracked (p-value 0.24, Fischer's exact). Over the first 12 months of this iteration, a total of 44 action items originated from HCPR discussions, 32 (73%) of which have been completed. Examples include: creation of care process guidelines, operational changes, clinical outcomes presentations, and practice-related educational topics. Additionally, we noted that as we increased awareness of eligibility for multidisciplinary quality improvement efforts, participation in these projects improved, leading to decreased variability and overall length of stay for studied surgical repairs. Survey responses and interviews demonstrated increased awareness of outcomes, and reductions in siloed conversations. Follow up topics are summarized in a digestible format, then archived online and available for future reference to members of the Heart Center.

Conclusions: This version of HCPR has improved psychological safety, increased situational awareness, identified actionable improvements, and promotes knowledge of benchmarks and clinical transparency. A team approach is necessary for HCPR to pursue evolving priorities, and a structure for addressing threats and analyzing trends is essential for improvement and participant satisfaction. Challenges to the process include: ensuring equally inclusive engagement of audience participants, dedicated time and resources for preparation, and attention to triaging cases in a high-volume setting.

Friday 25

The Fetal Heart Program At Children's Hospital Of Philadelphia: Ten-Year Experience Of Prenatal Cardiovascular Care From 2013-2022

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Background: Prenatal ultrasound technologies and operator skills continue to advance such that today, state-of-the-art diagnostics, counseling and management services can be offered before birth for most forms of congenital heart disease (CHD). As such, specialized programs such as the Fetal Heart Program (FHP) at Children's Hospital of Philadelphia (CHOP) continue to develop and grow. We report on the evolution of our model of prenatal heart care and growth at the FHP @ CHOP.

Methods: A review was undertaken of overall volume of fetal echocardiogram (FE) growth, number of fetal patients (FP) with CHD defined as any form of cardiovascular disease requiring either postnatal treatment or follow-up, and programmatic personnel staffing increases seen at the FHP during a 10-year period from fiscal year (FY) 2013 to 2022. Specific forms of CHD are reported as number of FP seen per year, median (range). FHP clinical model services encompass a comprehensive team approach consisting of protocolized imaging performed by fetal heart sonographers, dedicated FHP nurse coordinators, FHP social worker and a dedicated team of fetal cardiologists.

Results: During the 10-year period, the FHP experienced 28% overall volume growth in number of FE from 2570 in FY13 to 3297 in FY22. Growth was steady year-upon-year except for a slight decrease in FY20, due to temporary restrictions related to COVID. Number of fetuses with CHD increased by 49% from 287 in FY13 to 429 in FY22. Of note, despite a lower number of FE performed in FY20 there was no decrease in number of FP with CHD. Common forms of CHD with median (range) FP seen per year were: hypoplastic left heart syndrome 39 (31-51), tetralogy of Fallot 49 (39-68), transposition of the great arteries 27 (15-35), atrioventricular canal 27 (20-45), severe pulmonary stenosis or atresia with intact ventricular septum 18 (9-25) and truncus arteriosus 7 (5-12). While most condition numbers fluctuated modestly year upon year, the anomaly with the most dramatic steady increase in number was coarctation of the aorta/hypoplastic aortic arch, with a progressive increase from FY13 to FY22 of 34 to 69 FP per year, median 47 over the 10-year period. From FY13 to FY22 number of fetal heart sonographers in the FHP increased from 3 to 5, FHP nurse coordinators from 2 to 4, and fetal cardiology attending coverage increased from 1 per day to 1.5 per day 3 days a week and 2 per day twice a week.

Conclusions: The FHP @ CHOP has experienced substantial growth over a 10-year period from FY13 to FY22 with an increasing volume of FE, FP and clinical programmatic growth. We suspect this phenomenon is common amongst our community of CHD care.

Friday 26

Efficacy Of A 3-Year Longitudinal Project-Based Quality Improvement Curriculum In Pediatric Cardiology Fellowship

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Background: Our goal was to create a robust quality improvement and patient safety (QIPS) curriculum that gives fellows both didactic knowledge and first-hand experience with improvement science and produces a longitudinal QIPS project to improve patient care and safety over their three-year fellowship. The curriculum will meet Clinical Learning Environment Review (CLER) requirements and give trainees the foundation to continue improvement work beyond training.

Methods: This curriculum was designed in 2019 in alignment with mandatory CLER requirements for fellowship training and is overseen by QIPS leadership within the Texas Children's Hospital Heart Center. A series of six 30-minute didactic lectures is paired with designated group work time over the course of 8 months. During their first year of training, each class brainstorms a group QIPS project which they will continue for the entirety of the three-year fellowship. They are paired with faculty mentors who coach the selection, design, implementation, and finally the presentation of these projects every year. Faculty participation in the curriculum serves as training to build QIPS expertise and capacity in the division. Knowledge assessment is obtained through pre- and post-surveys, quantifying the trainee's subjective comfort in eight QI competencies on a five-point Likert scale. Assessed competencies include developing a focused aim, identifying appropriate outcome measures, utilizing multiple plan-do-study-act cycles, analyzing data, creating run/control charts to communicate data, creating a team with assigned roles, and ensuring changes are sustained in practice. A secondary measure of success is academic products resulting from fellows' QIPS work.

Results: Since 2019, 40 pre-tests and 27 post-tests were completed. We found statistically significant improvement across all competencies in post-test surveys (mean: 3.56, IQR: 3.13-4.00) when compared to pre-test surveys (mean: 2.79, IQR: 2.28-3.13) ($p < 0.001$). Since curriculum implementation, fellows have produced one published manuscript, two abstracts, and four oral presentations describing their improvement work in patient care and safety. Three projects are currently in the sustain phase, and three projects are on-going. All projects met their initial QIPS improvement aims. Additionally, mentoring faculty members have gone on to lead other QI work throughout the division.

Conclusions: Many programs face challenges meeting the quality improvement CLER requirement for fellowship training due to insufficient or ineffective teaching, and most programs also lack valuable QI experiences for fellows. This longitudinal QI curriculum is centered around fellow involvement in all components of project planning and implementation. It provides QI experience that is invaluable in promoting fellows' QI knowledge and commitment, as evidenced by statistically significant improvements in all measures on self-reported surveys. There have also been important systemic improvements in patient care and safety as a result of these projects. This curriculum creates opportunities for academic publications and presentations, builds faculty expertise and capacity for future work within the division, and most importantly, works to improve multiple aspects of patient care. This curriculum could serve as a model for other cardiology fellowships working to meet CLER requirements.

Friday 27

Diagnostic Dilemma: A Previously Healthy Teenager Suffered Sudden Cardiac Arrest At Home

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Background: 14-year-old boy presented after a sudden cardiac arrest (ventricular fibrillation) and incidentally detected to have anomalous origin of the left anterior descending from the right coronary sinus with intraseptal course creating a myocardial bridge. Further testing revealed a positive invasive provocative test suggesting inducible left coronary artery compression. Currently, there are no specific anatomical features identified in these patients associated with myocardial ischemia or SCD. In addition, the prognostic implication of intraseptal AAOLCA remains unknown in those detected incidentally. Hence, shared decision making is key in their management as limited literature is available and the benefit of surgical intervention remains unknown.

Methods: Echocardiogram showed structural heart disease with suspected anomalous coronary artery. He developed junctional tachycardia, which converted to sinus rhythm after an IV sotalol dose of 1mg/kg. CMR showed no evidence of late gadolinium enhancement or regional wall motion abnormalities, but highly suspected anomalous coronary origin. CTA confirmed anomalous origin of LAD from right sinus with a myocardial bridge. We performed coronary angiography with no LAD compression; intravascular ultrasound (IVUS) and provocative testing using dobutamine and measure instantaneous wave-free ratio (iFR), which was suggestive of inducible coronary stenosis at peak exercise. Electrophysiology study induced AVNRT.

Results: A shared decision was made along with the patient's parents to proceed with surgical correction of the anomalous LAD due to evidence of inducible coronary stenosis on provocative testing. In one of the case series of 18 children with intraseptal AAOLCA by Doan and colleagues, inducible ischemia was noted in 50% of the children on provocative CMR testing. One underwent coronary artery bypass graft. A newly devised pressure-related coronary flow index known as iFR has been found to correlate with diastolic fractional flow reserve in adults with coronary artery stenosis. This tool has been used to risk stratify patients with AAOCA at risk of diminished coronary flow under conditions replicating exercise. Based on studies, $iFR < 0.89$ indicates impaired coronary artery flow. In our patients, iFR at baseline was normal, however, the iFR dropped to 0.67 with dobutamine suggesting inducible coronary. There is limited evidence to support the use of beta blockers in children with this anomaly. After shared decision making with parents, our patient successfully underwent transconal unroofing of the left coronary artery with reconstruction of the right ventricular outflow tract using bovine pericardium. The case illustrates the diagnostic and management dilemmas of anomalous coronary arteries in children.

Conclusions: The anomalous origin of left coronary artery with intraseptal course was considered a benign condition with no adverse cardiovascular sequelae. However, there are rare case reports of myocardial ischemia, arrhythmia and sudden cardiac arrest in children with this coronary anomaly. Therefore, it is important to rule out other potential causes of sudden cardiac arrest in healthy young individuals. At present, there are no specific anatomic features that are considered high risk for ischemia and no evidence-based management guidelines for this coronary anomaly, so the decision for surgical repair is case-by-case. A new surgical technique of unroofing the anomalous intraseptal segment of the anomalous LAD has been attempted with no early post operative complications or mortality. However, the long-term outcomes in those who are operated vs not operated remains unknown to justify substantial benefit from open-heart surgery.

Friday 28

Large Infiltrative Cardiac Lipoma In An Adolescent

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Background: Cardiac lipomas are rare benign tumors of the heart composed of mature fat cells. They are rare findings in adults, and extremely uncommon findings in children with only few reported cases. They are typically found incidentally but can cause arrhythmias, inflow or outflow tract obstruction, coronary artery compression, embolization, or arrhythmias. In symptomatic patients, surgical resection of the mass is the primary treatment strategy.

Methods: A 15-year-old female presented to the emergency room with severe abdominal pain, vomiting, and diarrhea. She was diagnosed with pelvic inflammatory disease due to positive screening for gonorrhea and chlamydia, and an abdominal Computed Tomography (CT) was performed to exclude tubo-ovarian abscess and acute appendicitis. The intraabdominal structures were normal, but imaging incidentally noted a mass at the cardiac apex. Echocardiography, cardiac magnetic resonance imaging (cMRI), and cardiac CT were performed and were suggestive of a large cardiac lipoma in the mid- and apical left ventricle (LV) with infiltrative components. She was monitored on telemetry for arrhythmias; surveillance cardiac troponins and brain natriuretic peptide were obtained to exclude subclinical myocardial ischemia or heart failure.

Results: Transthoracic echocardiography demonstrated a large well circumscribed mass in the LV apex. Cardiac MRI demonstrated a large cardiac mass 3cmx2cm in the mid- and apical portions of the LV that extended anterolaterally towards the apex of the right ventricle and into the pericardial space. Tissue characterization demonstrated hyperintensity on T1 without fat saturation, hypointensity on T1 with fat saturation, hypointensity on SSFP images, hypointensity on T2, non-vascular appearance on first pass perfusion and hypointensity on late enhancement imaging. She demonstrated normal biventricular function and no inflow or outflow tract obstruction. These findings and the location of the mass were consistent with a lipoma (likely infiltrative type). Cardiac CT further characterized the mass, demonstrating epicardial/pericardial components encasing the distal segment of the left anterior descending coronary artery; there was no extension to the anterior chest wall.

Conclusions: Cardiac magnetic resonance imaging is particularly useful for tissue characterization, which can often lead to a specific diagnosis for cardiac masses in children. In this case, tissue imaging by cardiac MRI was strongly suggestive of a lipoma, negating need for invasive cardiac biopsy, and guiding further management. Ongoing advanced imaging surveillance and monitoring of cardiac function is being performed.

Friday 29

Repair Of Complex Heterotaxy Syndrome In An Unoperated Young Adult

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Background: We demonstrate the utility of advanced three-dimensional imaging in an extreme case of heterotaxy syndrome in which there was situs solitus with bridging innominate vein plus small LSVC, atrial to ventricular discontinuity, ventricular to arterial discontinuity, double outlet right ventricle, moderate pulmonary stenosis, complete atrioventricular septal defect with straddling of elements of the left sided superior bridging leaflet to the right sided(systemic) ventricle. The pre-operative planning enabled us to complete a unique repair which included an entire conduit from the AVSD patch to the sub aortic conus

Methods: A 25-year-old male with situs solitus, ventricular inversion, AVSD, DORV and pulmonary stenosis, was followed throughout childhood with no intervention because pulmonary stenosis protected his pulmonary vascular bed. he presented with decreasing exercise tolerance and cyanosis. He was investigated by echocardiography cMRI, CPET, contrast CT scan and cardiac catheterization. Although an anatomic diagnosis was achieved, the information was not helpful in decision making of repair versus palliation. CT images were loaded onto a 3D true viewer platform (Echopixel) and also a 3D model of the heart was obtained after segmentation. This methodology allowed planning for biventricular repair

Results: A 1.5 ventricle repair was achieved by a right bidirectional cavopulmonary anastomosis and division of the left SVC, oversewing of the stenotic pulmonary valve and main pulmonary artery, closure of the pulmonary artery bifurcation, closure of the VSD portion of the canal defect, division of right ventricular muscle bundles, implantation of a 22mm Gortex conduit from the VSD patch to the subaortic conus, baffle closure of atrial portion of AV canal defect such that IVC blood was channeled to the left sided right ventricle, and then an RV to PA conduit connected the right ventriculotomy to the main pulmonary artery. Cardiopulmonary bypass was 352 minutes and XCT was 270 minutes at 25C. Intraoperative echocardiogram showed normal biventricular function, trivial valvar insufficiency, mild aortic insufficiency, unobstructed atrial baffle and unobstructed right ventricular outflow without insufficiency. Our patient was weaned from bypass with minimal inotrope support and was discharged from hospital on the 6th post operative day. He suffered an attack of Covid-19 six weeks post operatively and associated cardiomyopathy which then recovered over time. At 2 years following surgery his echo findings have remained stable and he maintains his work as a manual laborer.

Conclusions: The advent of advanced interactive imaging and 3D printing as adjuncts to investigation of patients with complex intracardiac anatomy has enabled us to design unique surgeries in highly selected cases. Anatomic variability of patients and their presentation, makes assessment of the wisdom of decision making more difficult. The 3D printed model and the interactive visualization enhanced different aspects of the surgical planning and were complementary. This patient is one of a series of patients in our institution in whom 3D printing and interactive visualization has been critical to surgical planning.