

Sat 1

Slow Is Fast And Fast Is Slow: A Case Of Suspected Fetal Long QT Syndrome

J Jin, S Patel, S Samples

Ann and Robert Lurie Children's Hospital of Chicago

Background: Congenital long QT syndrome (LQTS) is a relatively rare disorder with a variable presentation in fetal life, ranging from bradycardia to ventricular arrhythmias, to intrauterine fetal demise. Bradycardia may be due to sinus bradycardia, or second or third degree atrioventricular (AV) block due to profoundly prolonged repolarization times. Accurate diagnosis and successful antenatal pharmacological therapy can result in the delivery of a non-hydrotic liveborn infant.

Methods: A 29-year-old healthy female pregnant with a singleton fetus was initially referred to fetal cardiology services at 32 weeks due to concern for possible fetal arrhythmia. Initial fetal echocardiogram confirmed no structural congenital heart disease with complete AV dissociation and variable ventricular rates (83-110bpm), atrial rates of 120-132bpm, moderately hypertrophied biventricular myocardium with moderately depressed systolic function, mild tricuspid and mitral regurgitation, and mild cardiomegaly without hydrops. Maternal serology for anti-SSA and -SSB antibodies were negative. Maternal and family history were negative for LQTS and maternal ECG was normal. Fetal magnetocardiography (fMCG) revealed periods of 2:1 and 3:1 AV conduction, severe QT prolongation (645 ms), and brief episodes of ventricular arrhythmias concerning for Torsades de Pointes (TdP). The patient was admitted for inpatient initiation and titration of Propranolol therapy and Magnesium supplementation. Fetal echocardiogram six days after beginning therapy demonstrated 1:1 AV conduction with heart rates in the 90s-low 110s, no ectopy, and improvement in biventricular function and bilateral AV valve regurgitation. Subsequent fMCG confirmed 1:1 AV conduction with no further evidence of TdP.

Results: Given the QT prolongation, ventricular arrhythmias, and periods of 2:1 and high-grade AV block, the likeliest diagnosis was congenital LQTS. However, with periods of higher-grade AV block, uncertainty remained as to whether this clinical presentation represented a primary cardiomyopathy with conduction disease rather than a primary channelopathy with episodes of TdP and persistent cardiomyopathy. If so, the presence of ventricular arrhythmias in the setting of a primary cardiomyopathy would indicate a particularly poor prognosis. The patient was counseled on the limitations surrounding the prenatal diagnosis, and that further postnatal workup would aid prognostication. She was followed biweekly with alternating fetal echocardiograms and biophysical profiles until her delivery. She delivered a liveborn male infant at 39 weeks gestation. His electrocardiograms during the first week of life continued to demonstrate a severely prolonged QTc (525-623ms), with intermittent PVCs, and abnormal T waves. He was initiated on propranolol monotherapy initially, and Mexilitene was added within the first week due to persistently prolonged QTc intervals (>550ms) and concern for intermittent bradycardia. Postnatal echocardiogram also revealed prominent LV trabeculations but normal systolic function. A genetic panel for cardiomyopathy and arrhythmias were sent but are pending.

Conclusions: The combination of tachyarrhythmias, heart block, and/or sinus bradycardia in a fetus should raise suspicion for congenital LQTS. Fetal presentation of TdP is rare but can happen as in this case. fMCG can help establish the diagnosis of LQTS and evaluate for TdP. Our case also corroborates previously-described cases in which sustained ventricular tachycardia and second-degree AV block have been observed in association with ventricular hypertrophy and dysfunction, which in our case proved reversible following treatment of the precipitating arrhythmias.

Sat 2

Refractory Supraventricular Tachycardia After Respiratory Syncytial Virus (Rsv) Bronchiolitis During The Recovery Phase In An Infant: A Case Report

N Garg, K Woods, E Shanshen

Southern Illinois University

Background: Respiratory Syncytial Virus (RSV) is the most common cause of bronchiolitis in infants. Only a limited number of RSV bronchiolitis infants have been reported to develop supraventricular arrhythmias. In 2022, the United States (US) experienced a surge in the number of infants with RSV bronchiolitis with increased morbidity and mortality, highlighting the need to understand rare manifestations and complications of the common disease. In this report, we present a case of an infant who developed supraventricular tachyarrhythmia (SVT) in the RSV bronchiolitis recovery phase while the patient was still in the pediatric ICU.

Methods: A five-week-old previously healthy, full-term male with RSV bronchiolitis developed SVT with a heart rate of 210 to 230 on day five of admission during recovery while requiring less respiratory support. He remained hemodynamically stable and was treated with multiple antiarrhythmic medications with no response. The rhythm persisted despite medical management for eight hours. He was then successfully electrically cardioverted to normal sinus rhythm. Echocardiogram showed normal cardiac anatomy and function. Electrocardiograms showed no evidence of ventricular preexcitation. SVT recurred within the first 24 hours of the onset starting with ventricular preexcitation beat confirming the accessory pathway reentry mechanism.

Results: SVT is the most common arrhythmia in infancy and requires emergent action to prevent cardiorespiratory compromise. Infants with RSV bronchiolitis can develop SVT firstly due to the inflammation of one of the layers of the heart wall, secondly due to beta-agonist use during management, and thirdly due to hypoxia caused by the disease. The exact pathogenesis of RSV-associated arrhythmias is still unknown. In our case, the temporal association between the RSV infection and the arrhythmia provides a piece of circumstantial evidence that RSV perhaps the etiological agent. Our patient was also ultimately found to have a reentry pathway which could have been revealed due to the RSV infection. Our patient had another episode of SVT within 12 hours of electrical cardioversion, was started on propranolol, and was discharged home on this medication with no further episodes. Literature review revealed that complete heart block is the most common arrhythmia associated with RSV but ectopic beats, multifocal atrial tachycardia, ventricular tachycardia, and ventricular fibrillation have been reported as well. Knowledge of these associations is essential during the current RSV surge in the US. It also highlights the importance of close cardiorespiratory monitoring for these infants during hospitalization in the current epidemic.

Conclusions: SVT after RSV infection has been reported only in a limited number of infants. The temporal association between the infection and SVT highlights the need to further understand the disease course. Multiple risk factors for SVT are present in cases of RSV bronchiolitis including RSV infection, use of albuterol, as well as hypoxia, which all could play a role in revealing silent accessory pathways. Hence close cardiac monitoring until complete recovery is essential to prevent cardiorespiratory compromise.

Sat 3

A Rare Case Of A Large Coronary Sinus Diverticulum In An Adolescent Patient With Supraventricular Tachycardia: Keys To Imaging Recognition

N El Sherif

Mayo Clinic

Background: Coronary sinus (CS) diverticulum is a rare variant of the coronary sinus anatomy that is thought to result from embryologic failure of regression of the sinus venosus. It consists of a distinct epicardial pouch with a narrow neck that opens into the proximal coronary sinus. It has been reported in 7-11% of posteroseptal and left posterior accessory pathway related adult cases

Methods: The patient is a 14-year-old female with recurrent supraventricular tachycardia (SVT) and a prior unsuccessful ablation attempt adjacent to the posterior/inferior mitral valve annulus. She was found to have a large CS diverticulum during a pre-procedural transthoracic echocardiogram (TTE). A CT angiogram confirmed the diagnosis of a 37 x 24 x 20 mm diverticulum arising from the coronary sinus just distal to the ostium of the inferior interventricular vein. The patient underwent successful ablation of a concealed left posterior accessory pathway from within the neck of the diverticulum and reported no further episodes of tachycardia at follow-up one month later.

Results: There is a well-known association between congenital anomalies of the CS and conduction tissue abnormalities, as the electrophysiological properties of the CS are altered by the presence of these abnormalities. The presence of a CS diverticulum should be suspected in cases of failed prior ablations and when the optimal location for ablation is the proximal CS. If these accessory pathways have antegrade conduction properties, they tend to have short refractory periods, which increases the risk of rapid ventricular response during atrial fibrillation episodes and sudden cardiac death. Therefore, it is important to precisely locate and ablate these accessory pathways to eliminate this risk. To diagnose a CS diverticulum, TTE can be used, but it may be difficult to detect if the diverticulum is small. Angiography and CT angiography are more sensitive for detecting CS diverticula. Treatment for CS diverticulum usually involves ablation of the accessory pathway within the diverticulum. In this case, ablation at the neck of the diverticulum was successful in eliminating the patient's SVT. One pediatric case has been described in the literature of a patient with congenitally corrected transposition of the great arteries and a history of medication resistant reentrant tachycardia. This patient had 2 prior ablation attempts prior to diagnosis of a CS diverticulum. Subsequently, a successful ablation of a posteroseptal pathway was performed. To our knowledge, this is the only reported pediatric case in the literature in an otherwise structurally normal heart and with a concealed retrograde conducting only pathway.

Conclusions: CS diverticulum should be suspected in cases of accessory pathways that failed multiple ablations. Angiography and/or cross-sectional imaging should be used to accurately diagnose this condition.

Sat 4

Implantable Loop Recorder In A Patient With Diaphragm Pacing

G Hatem, M Shillingford, S Ferns

Wolfson Children's Hospital

Background: With improvement in technology, medical complex patients may be served with multiple newer modalities to record electric activity from the heart and nerves. Understandably there are concerns with electric cross talk and interference. To the best of our knowledge this is the first reported case of an implantable loop recorder (ILR) in the setting of a phrenic nerve stimulator.

Methods: A 10-year-old female with a history of C2 quadriplegia presented with frequent asymptomatic pauses upto 2.5 seconds and an unusual episode of syncope. She had an implanted gastric tube, tracheostomy, and a bilateral phrenic nerve stimulator with unipolar pacing electrodes fixed to the phrenic nerve and tunneled to the receiver/stimulator unit in a subcutaneous abdominal pocket. The patient had skin hypersensitivity and prior temporary one-to-4-week monitors had resulted in a skin rash limiting external cardiac monitoring options. Since she did not meet criterion for a pacemaker, the risks and benefits of an ILR were discussed with the patient and her family. Cardiac electrogram tracings obtained on a 12-lead electrocardiogram and telemetry demonstrates periodic electrical interference corresponding to timing of impulses generated by the phrenic nerve stimulator and we were concerned about the degree of interference the phrenic nerve stimulator might cause on ILR recordings and hence the reliability of the data recorded. Prior to the implant surface testing with the diaphragm pacing system programmed at maximal output was performed to test the signal quality on the ILR. Surface testing showed excellent signal quality with minimal interference from diaphragm pacing. An ILR was therefore placed enabling successful continuous remote monitoring post discharge from hospital.

Results: An implantable loop recorder is designed to capture electrocardiogram tracings automatically when it senses a pre-programmed heart rate/arrhythmia or if activated by the patient. These devices help in the surveillance of patients with potentially serious arrhythmias and help guide further management in patients that don't meet criterion for a more invasive procedures such as a pacemaker or Automatic implantable defibrillator implantation. Given that an implantable loop recorder operates by recording the electrical activity of the heart, it was of interest to identify if the presence of a diaphragm stimulator would interfere with the signals of the ILR.

Conclusions: This report demonstrates that despite the proximity of the diaphragm stimulator to an ILR in a pediatric patient, the electrocardiogram tracing and signal of the ILR is clear and reliable.

Sat 5

Hospital-Level Variability In Pacemaker Therapy In Neonates With Isolated Congenital Complete Heart Block— An Analysis Of The PHIS Database

S Weinreb, YSV Huang, C Janson

Children's Hospital of Philadelphia

Background: Congenital Complete Heart Block (CCHB) is a rare diagnosis, with incidence 1 in 15,000-20,000 births. Prior reports have observed pacemaker (PM) placement in 69% of fetal diagnosed patients by 1 year and in 74-96% of patients by 20 years. There is limited multi-center data reporting outcomes and practice variability in this patient population. We performed a multicenter retrospective cohort study utilizing the Pediatric Health Information Systems (PHIS) database to assess variability in pacemaker therapy across US tertiary children's hospitals.

Methods: All subjects in the PHIS database (1/1/2007-12/31/2021) with ICD-9 or ICD-10 diagnosis code for congenital heart block or complete AV block and with age at index hospitalization <30 days were included. Subjects with lower degree AV block, Long QT Syndrome, or structural congenital heart disease, except patent ductus arteriosus or atrial septal defect, were excluded. Permanent PM placement procedures were identified using ICD and CPT codes. Descriptive statistical analysis was performed to describe differences among hospitals.

Results: There were 691 unique CCHB subjects identified from 47 US hospitals, with median 14 subjects (IQR 8-19, range 2-35) per hospital. The cohort was 58% female, 60% white, and 50% privately insured. Median length of stay of index hospitalization was 11 days (IQR 5-28), and median follow-up duration was 1.7 years (IQR 0.1-6.4). PM placement occurred in 397 (57%) subjects, of whom 308 (45%) received PM in the index hospitalization. Median age at PM placement was 6 days (IQR 2-58). Of those receiving PM, 224 (56%) received dual chamber and 164 (41%) single chamber PM. At the hospital level, PM placement occurred in a median 57% (IQR 47-69%) of subjects per hospital. Two hospitals implanted 0 PMs (2 subjects per hospital) and one hospital implanted PMs in all 5 subjects. Dual chamber PM placement occurred in median 58% (IQR 29-83%) of subjects per hospital and single chamber placement occurred in median 33% (IQR 13-60%) of subjects per hospital. Eight hospitals placed dual chamber PMs in all paced subjects (1-11 PMs per hospital), and an additional 6 hospitals placed dual chamber PMs in > 80% of subjects (8-18 PMs per hospital). Two hospitals placed single chamber PMs in all paced subjects (2 and 5 subjects, respectively), and an additional 4 hospitals placed single chamber PM in > 80% of subjects (8-16 PMs per hospital).

Conclusions: This unique use of administrative data has allowed for the development of the largest cohort of neonates with CCHB published to date. There is observed hospital-level practice variability in pacemaker placement in the CCHB patient population, especially as it relates to dual vs single chamber pacing. This variability could reflect patient-level factors or may reflect hospital or physician-level philosophy around pacemaker therapy in infants. Future analysis of this cohort will investigate interactions between patient age, patient size, geographic region, and hospital/CCHB cohort size on single vs dual chamber PM placement.

Sat 6

Occult Cardiovascular Disease Detected During Mandatory Pre-Participation Sports Electrocardiogram Screening In Large Volume High Schools

Z Boynton, A Stichter, W Blackstone, M Cole, S Sima, E Ernst, B Trivedi, S Tisma-dupanovic, B Kakavand, G Dadlani

Nemours Children's Health

Background: Sudden cardiac arrest is a tragic event, especially in young athletes and occurs with an incidence of 1:40,000 – 1:80,000. The addition of an electrocardiogram (ECG) to traditional pre-participation sports screening history and exam can enhance detection of occult forms of cardiovascular disease with a risk of sudden cardiac arrest. In May of 2021, Orange County School District in Florida (the 9th largest school district in the US with over 208,000 students in 56 high schools) mandated ECG screenings prior to participating in high school sports.

Methods: Orange County School district partnered with Who We Play For, a non-profit organization that conducts free or low-cost ECGs (\$20.00 fee) at high schools in the school district. Volunteer pediatric cardiologists interpreted each ECG based on international criteria for sports ECGs. Athletes were contacted with the results and those with abnormal results were called to ensure follow up with a pediatric cardiologist. Nemours IRB was obtained for retrospective review of the data. We then assessed the subsequent cardiac up that occurred for all abnormal ECGs at Nemours Children's Hospital. This is one of three pediatric health systems in Orange County, Florida.

Results: There were 11,547 ECG screenings completed between 5/1/2021 – 5/1/2022 with 2.7% (310/11547) abnormal. Of the abnormal, there were 105 students that followed up with our institution with follow-up EKGs that revealed: 20% were normal, 21% arrhythmias (PAC/PVC), 8.5% axis deviation, 18% conduction problems, 12% ST-T abnormalities, 4.7% ventricular hypertrophy, 12% other and 3% structural abnormalities. Subsequent cardiovascular testing revealed 14 patients with critical cardiovascular disease with a potential risk of sudden cardiac arrest including 12 patients with wolff-parkinson-white syndrome and 2 with cardiomyopathy.

Conclusions: The addition of an electrocardiogram to the pre-participation physical exam and history can detect occult cardiovascular disease that would have otherwise been missed. ECG screening is feasible at large volume school districts in the USA and can detect forms of cardiovascular disease that can predispose to a risk of sudden cardiac arrest (Wolff-Parkinson-White syndrome and hypertrophic cardiomyopathy).

Sat 7

An Automatic Fellow Feedback System For Pediatric ECGs

I Asztalos, C Janson, N Griffonetti

Children's Hospital of Philadelphia

Background: Interpreting electrocardiograms (ECGs) is a fundamental skill for pediatric cardiologists, and is developed largely during pediatric cardiology fellowship training. Mastery of ECG interpretation requires dedicated practice and thus timely and appropriate feedback regarding mistakes and areas of improvement from master clinicians—generally from pediatric electrophysiologists (EP). Pediatric EPs, have many time constraints, not least amongst which is the burden of reading hundreds of ECGs per week, limiting their availability to coach fellowship trainees. Pediatric EP overreads and corrections of trainee ECG reports are a valuable resource but are severely underutilized by the cumbersome and time-inefficient methods of acquiring edits.

Methods: An online dashboard was created to serve as a central repository of all ECGs read by individual cardiology fellows (n=18). The dashboard collects all ECGs read by a trainee for which the final read is altered by the pediatric EP overreader. Changes to both the diagnoses and the measurements are collected and reported. The dashboard also allows for pediatric EPs to mark an ECG as a major error, a minor error, or “good catch”.

Results: Prior to implementation of the dashboard most fellows reviewed either no (33%) or fewer than half (77.8%) of their ECG reports after they were overread by a pediatric EP (mean 19.8%) for an average of 13.9 minutes per week by self-report. Overall, fellows were unsatisfied with the quality (39.2/100), timeliness (36.2/100), breadth (28.3/100), and frequency (17.9/100) of ECG feedback from pediatric EP overreaders.

Conclusions: An online dashboard can serve as a valuable pedagogical adjunct in the instruction of ECG interpretation.

Sat 8

Successful Coil Embolization Of A Ruptured Mycotic Coronary Artery Aneurysm In A Pediatric Patient

M Niccum, M Gupta, J Rome, M O'Byrne

Children's Hospital of Philadelphia

Background: Infected (mycotic) coronary artery aneurysms (MCCA) are extremely rare and carry a high risk of mortality. To maintain source control of the infection and mitigate risk of rupture and exsanguination, MCCA are usually managed with parenteral antibiotics and surgical repair. We present a case of a 12-year-old boy with thoracic synovial sarcoma complicated by empyema and *Rhizopus microsporus*-positive pericardial effusion treated with a pericardial window who subsequently developed hemorrhage from a ruptured MCCA arising from the left circumflex coronary artery (LCX) which was successfully treated with percutaneous coil embolization.

Methods: CT angiography identified a possible MCCA as the source of hemorrhage. The patient underwent cardiac catheterization, and angiography demonstrated two large saccular aneurysms of a proximal branch of the LCX which were selectively engaged with a microcatheter. Penumbra (four 60cm Packing and two Ruby Soft®) coils (Penumbra, Inc, Alameda, CA) were used to fill both aneurysms. Subsequent angiography demonstrated no residual flow into the aneurysms and hemorrhage abated. Troponin-I levels peaked 24 hours post-procedure without associated hemodynamic instability or electrocardiographic changes. He was discharged and at three-month follow-up had normal left ventricular systolic function.

Results: To the best of our knowledge, this is the first description of percutaneous coil embolization of an MCCA in a pediatric patient. Surgical resection is the gold-standard treatment of MCCA, but when the patient is deemed a poor surgical candidate, alternative approaches must be considered. Several details about the case may be instructive. Filling the aneurysms with coils only after the patient completed six weeks of parenteral antibiotic therapy likely decreased the potential for subsequent abscess formation. Penumbra coils were used rather than covered stents to reduce the risk of aneurysm rupture, as these coils fill the aneurysmal structure without exerting radial forces that can precipitate or exacerbate bleeding. Finally, in planning the procedure, it seemed likely that occlusion of a branch of the circumflex would be necessary, which would inevitably cause myocardial infarction with risk of heart block. Selective right coronary angiography was performed to confirm the presence of a right dominant circulation and a pacing catheter was placed prior to intervention as a precaution. Though some tissue infarction occurred, only mild segmental wall motion abnormality has been seen to date.

Conclusions: This case represents a rare presentation of a hemorrhaging MCCA that was not amenable to surgical intervention. In these situations, coil occlusion of the vessel is a potential therapy. In planning and executing this kind of procedure, accurate imaging of the affected vessel(s) is vital to ensure that appropriate equipment is available, as are measures to prevent or mitigate potential adverse events. In cases with broad based aneurysms relative to the vessel, occlusion of the vessel may be necessary. Even in critically ill patients, this can be tolerable, though careful follow-up is necessary.

Sat 9

Reduction Of Post Cardiac Catheterization Flat Time In The Pediatric Population

L Gervasi, M Pretsch, J Fitzgerald, Amy Lisanti

Children's Hospital of Philadelphia

Background: Pediatric patients who undergo cardiac catheterization at Children's Hospital of Philadelphia are required to lay flat post-procedure for 6 hours for femoral arterial access. During the flat time, patients can report anxiety, pain, or generalized discomfort. Research suggests that flat times can be safely reduced without increased site bleeding and decrease discomfort. The purpose of this quality improvement project was to increase patient satisfaction and comfort by decreasing flat times without increasing rates of site bleeding. The population chosen was the cardiac transplant patients because they need yearly cardiac catheterizations and would be able to provide comparison data and feedback.

Methods: The project utilized a PDSA cycle methodology (Plan,Do,Study,Act). The first two PDSA cycles were targeted to all cardiac transplant patients undergoing cardiac catheterization and biopsy who were recovering in the Cardiac Preparation and Recovery Unit. The first cycle reduced the flat time by one hour from 6 to 5 hours. The second cycle reduced the flat time from 5 to 4 hours. After the reduced flat time was completed, patients/families completed a survey to provide feedback on their level of satisfaction and comfort and compare their experience to previous recoveries. Patients under 18 filled out the survey with their parents and patients who were over 18 filled out the survey themselves.

Results: Both PDSA Cycles showed no impact on incidence of site bleeding. Patient/parent survey results were varied with all patients reporting their experience as the same or improved.

Conclusions: Based on this project, the standard post catheterization flat time for transplant patients with femoral arterial access was safely reduced from 6 to 4 hours. Decreased flat time to 4 hours after Femoral arterial access is now the standard of care at this institution for all diagnostic catheterizations, annual transplant/biopsy and Patent Ductus Arteriosus device closures.

Sat 10

Study Design During Covid Times And Initial Safety Profile Of A Novel Pediatric 4D Micro Trans-Esophageal Echo Probe And Imaging System

S Shah, H Hancock, N Madan, D Aly, M Kiaffas, J Huffman, G Shirali

Children's Mercy Hospital

Background: Current Pediatric Transesophageal Echocardiography [TEE] probes do not have the ability for real-time 3D or 4D imaging. We report the results of the first clinical safety study for a pediatric 4D TEE probe and imaging system in North America.

Methods: The parents of kids [> 5 kg in weight] scheduled for cardiovascular intervention requiring TEE, were approached. Informed consents were obtained from both the parents; and child [> 7 yrs] had to provide verbal assent to be enrolled in the study, as per the IRB. A negative COVID-19 RT-PCR test was required. A new GE 4D Mini-probe was tested. In order to avoid delays, the research TEE images were acquired at the time of vascular access. In the Cath-lab, study TEE images were obtained after the procedure was complete.

Results: A total of 8 pediatric TEE studies were performed, (age 3-20 years, body weight 13-64 Kg) . Probe placement was successful in all cases. Diagnostic image quality was obtained for 2D Echo, Color Doppler, Spectral Doppler, Multiplanar and 4D echo with and without color Doppler images. TEE probe and core temperature were closely monitored throughout the procedure. In one patient, GE system's safety alert worked as designed with an alert for increasing probe temperature towards the end of the study. No oral/esophageal bleeding, airway, or other cardio-pulmonary complications occurred till hospital discharge.

Conclusions: The new GE 4D micro- TEE probe and imaging system appears to be safe in the pediatric age group with diagnostic image quality. Larger scale studies are required for image optimization. **Clinical Implication:** This is the first North American 4D TEE feasibility and safety study in pediatrics. This paves way for further clinical application of Pediatric 4D TEE imaging systems.

Sat 11

Unrepaired Total Anomalous Pulmonary Venous Connection In A 40-Year-Old Male

R Moore, S Fuller, R Hu, S Partington

Children's Hospital of Philadelphia

Background: Total anomalous pulmonary venous connection (TAPVC) accounts for approximately 1.5% of congenital heart defects. Patients typically present in infancy with variable degrees of cyanosis, tachypnea and respiratory distress depending upon the degree of pulmonary venous obstruction and size of an interatrial communication. In extremely rare circumstances with an adequate atrial communication, patients can survive into adulthood. We present the case of a previously healthy 40-year-old male with exercise intolerance found to have unobstructed supracardiac TAPVC.

Methods: On exam, O₂ saturation was 87% with split S₂, SEM, clubbing and trace edema. Echocardiogram demonstrated severe RV dilation, mild RV dysfunction, large ASD and unobstructed TAPVC to the SVC. CT confirmed TAPVC draining to a confluence to the SVC with 3:1 shunt by MRI. Catheterization demonstrated elevated mean PA pressure (34 mmHg) and PVR (4.2 WU). Surgical repair involved baffling the pulmonary venous confluence to the LA, pericardial patch closure of the SVC, and fenestrated patch closure of the ASD. He received inhaled epoprostenol intraoperatively which was transitioned to sildenafil perioperatively. His symptoms resolved and O₂ saturations normalized.

Results: It is highly atypical for patients with TAPVC to present in adulthood. The most common presenting symptoms in older patients include dyspnea on exertion, cyanosis, and heart failure. In all case reports of adults with TAPVC, pulmonary venous return is unobstructed; in almost all cases, the interatrial communication is large, so that systemic cardiac output is preserved and the development of pulmonary vascular disease is delayed. Talwar et al. reported outcomes of 27 older patients (15-48 years old) who underwent surgical repair of TAPVC. Preoperatively, 11 patients had severe pulmonary hypertension and the rest moderate, and 6 patients had severe RV dysfunction. Six patients had a small atrial communication left open. All 27 patients survived to follow-up (3–127, mean 61 months). RV function normalized in all patients after repair. One patient experienced new onset atrial flutter after surgery. An earlier experience by Rodríguez-Collado et al. described outcomes in 19 adult patients (18-38 years old) who underwent repair. Two patients died, one from left atrial rupture and one from low cardiac output. All surviving patients had improvement in NYHA functional class. A significant reduction in mean PA pressure was demonstrated in all 14 patients who underwent repeat cardiac catheterization.

Conclusions: Though TAPVC is very rarely diagnosed in adulthood, it should be considered in the adult presenting with cyanosis, exertional symptoms and right heart dilation. Preoperative evaluation should include detailed cross-sectional imaging to clearly define the pulmonary venous anatomy and hemodynamic assessment by cardiac catheterization especially to determine pulmonary vascular resistance. Surgical repair is indicated in most patients diagnosed in adulthood, and outcomes described thus far are favorable. Less is known about the late-term outcomes of adult patients with TAPVC after surgical repair.

Sat 12

Patent Ductus Arteriosus And Coarctation Of The Aorta In Siblings With Biallelic Prdm6 Variants

H Stanley, B White, C LaRosa, JW Gaynor, A Strong

Children's Hospital of Philadelphia

Background: Patent ductus arteriosus (PDA) and coarctation of the aorta (CoA) are common congenital heart defects, which can occur in isolation or as part of an underlying syndrome. The genetic basis of non-syndromic PDA and CoA are incompletely understood. PRDM6 encodes a smooth-muscle-cell-specific histone methyltransferase and transcriptional repressor that plays a role in cardiac neural crest cell differentiation and migration. Heterozygous pathogenic PRDM6 variants are associated with non-syndromic PDA. Here, we present a father with PDA and his two daughters with PDA and CoA found by trio exome sequencing to harbor a single pathogenic and biallelic variants in PRDM6, respectively.

Methods: Sibling A was diagnosed with severe coarctation, mild arch hypoplasia, and PDA at one week old. She underwent end-to-end anastomosis and PDA ligation, and developed systemic hypertension and recurrent coarctation refractory to balloon dilation. Sibling B was diagnosed with CoA, arch hypoplasia, and PDA at birth, underwent repair, and developed hypertension with recoarctation. Renal vasculature imaging was negative in Sibling A (pending in Sibling B). Father was diagnosed with PDA in his fourth decade. Trio exome sequencing revealed a previously-reported, paternally-inherited, and likely-pathogenic PRDM6 variant (c.1646G>A; p.(Arg549Gln)) and maternally-inherited variant of uncertain significance (c.767C>G; p.(Pro256Arg)). Mother is asymptomatic (echocardiogram pending).

Results: Closure of the ductus arteriosus is a complex, coordinated process of vascular remodeling that requires migration of neural crest cells (NCC) to the ductus arteriosus and their transformation to vascular smooth muscle cells. Functional studies in cellular and mouse models suggest that PRDM6 haploinsufficiency disrupts NCC migration and differentiation and impairs cellular contractility, leading to persistent ductal patency, and exome sequencing in humans has shown that heterozygous pathogenic PRDM6 variants are associated with non-syndromic PDA. Combined PDA and CoA in association with heterozygous PRDM6 variants has not been reported, nor has the phenotype of biallelic PRDM6 variants been described. Our case series highlights an interesting contrast between isolated PDA in a father with a single heterozygous PRDM6 variant, a typical presentation based on prior studies, and the atypical combination of PDA and CoA in his daughters with biallelic PRDM6 variants. That a more complex phenotype was seen with biallelic variants suggests a dose-response, with more significant reductions in PRDM6 activity causing more significant aortic disease. Both siblings also have systemic hypertension, and though hypertension is a known finding after coarctation repair, work in mouse models suggests PRDM6 may have a role in regulating blood pressure via effect on renin-producing cells.

Conclusions: We describe a case of siblings with PDA, recurrent CoA, and systemic hypertension who were found by exome sequencing to have biallelic variants in the PRDM6 gene, including a novel variant (p.P256R). This case is the first description of coarctation in association with PRDM6 variants. Additionally, it is the first report of biallelic PRDM6 variants. The identification of additional probands will further delineate the full phenotypic spectrum of PRDM6-related disease and facilitate recurrence counseling and early diagnosis, as individuals with biallelic variants have a theoretical 100% risk of affected offspring.

Sat 13

Identification Of A Novel Gdf2 Gene Mutation Alters Management Of A Transitional Atrioventricular Canal Defect With Severe Pulmonary Arterial Hypertension.

M Delaney, J Tingo, C Avitabile

Children's Hospital of Philadelphia

Background: Our understanding of the genetic causes of pediatric pulmonary arterial hypertension (PAH) is rapidly evolving. The classification and distribution of etiology of PAH in children as compared to adults is dramatically different, with children having a greater predominance of idiopathic and congenital heart disease (CHD) associated PAH (1). Genetic testing continues to improve our understanding of PAH in children and can alter clinical management significantly. We present a case of a child with an unrepaired transitional atrioventricular canal (AVC) and severe PAH out of proportion to the CHD lesion, ultimately found to have a novel GDF2 mutation.

Methods: An 11-month-old with transitional AVC presented with right ventricular (RV) failure and severe failure to thrive. Cardiac catheterization demonstrated suprasystemic RV pressure, normal left heart pressure, Qp:Qs 1.7:1, and PVRi 7.9 iWU. A heterozygous loss-of-function mutation in GDF2 was identified. After 6 months of treatment with systemic treprostinil, sildenafil, ambrisentan, and tacrolimus (FK506), RV function, functional class, and hemodynamics improved. She underwent fenestrated atrial septal defect closure, left atrioventricular valve repair, and primary closure of a ventricular septal defect. Her growth and RV function improved after repair. She later transitioned to a fully oral PAH medical regimen.

Results: In PAH, pulmonary vasculature disease leads to RV dysfunction and RV failure. Without treatment, morbidity and mortality are high. The incidence and prevalence of PAH associated with CHD is 2.2 and 15.6 per million respectively (2,3). With increased availability, genetic testing has become an important component of disease management, especially in those with PAH out of proportion to their CHD lesion (4). Bone morphogenetic protein receptor 2 (BMPR2) mutations are the most commonly identified mutations in both familial and sporadic cases of PAH (5). The case patient had a mutation in GDF2 which encodes BMP9, a ligand of the BMPR2 receptor that affects upstream signaling in this pathway (6). While she was younger than most of the patients with PAH and GDF2 mutations in the literature, her atrial shunt may have contributed to her early pulmonary vascular disease (7, 8). This genetic diagnosis significantly altered management and led to aggressive vasodilator therapy with the addition of tacrolimus, given its positive effect on the BMPR2 signaling pathway. The patient improved with aggressive treatment, and her case represents an important lesson in the clinical management of PAH related to CHD.

Conclusions: Our understanding of the genetics of PAH has rapidly evolved over the last decade. PAH can complicate the management of CHD and lead to significant morbidity and mortality. We present a case of a transitional AVC with PAH out of proportion to patient age and lesion. Expanded genetic testing identified a newly appreciated mutation in pediatric PAH. With aggressive medical therapy, including FK506, her surgical candidacy improved. After successful surgical repair, she experienced continued improvements in symptoms, functional class, and RV function. We believe this case supports the concept that clinicians treating CHD with concerns for severe PAH should have a strong index of suspicion for contributing etiologies and consider a comprehensive genetic evaluation for pulmonary vascular disease.

Sat 14

Marfan Syndrome Or Loeys-Dietz Syndrome Confirmed By Genetic Testing In 22% Of Pediatric Patients With Syndromic Thoracic Aortic Aneurysm: Outcomes From Genetic Assessment Of 65 Pediatric Patients With Thoracic Aortic Aneurysm Referred To A Single Tertiary Care Center

N Garg, D Groepper, J Fleischer

Southern Illinois University

Background: The 2022 ACC/AHA Guidelines for the Diagnosis and Management of Aortic Disease mentions a process for optimizing genetic screening in patients with thoracic aortic aneurysm (TAA). The genetics of TAA has been studied in the adult population, with recent reports that approximately 20% of all TAA are associated with heritable conditions. Comparable studies in the pediatric population, however, are limited. Identification of the genetic causes of TAA in pediatric patients is critical to life-long care, as aneurysm location and dissection risk vary significantly based on molecular diagnosis.

Methods: We present a retrospective analysis of pediatric patients (age 0-18 years) with TAA (Z-score \geq 2SD) of the aortic root or ascending aorta by echocardiogram) who were referred for genetics evaluation to a single center. All eligible patients who underwent genetic testing from Jan, 2017 to Dec, 2021 were included (n = 65). Patients were further stratified into three groups based on presentation. "Non-Syndromic TAA" patients had no phenotypic features besides TAA (14/65), "Syndromic TAA" patients had TAA plus phenotypic features of connective tissue diseases (23/65), and "Other-Syndromic TAA" patients had features in addition to a connective tissue disease phenotype (28/65).

Results: Of the Syndromic TAA patients 22% (5/23) were found to have Marfan syndrome or Loeys-Dietz syndrome, with pathogenic or likely pathogenic variants in FBN1 (3), SMAD3 (1), TGFB3 (1). In addition, one patient in the Syndromic TAA group was not approved for a connective tissue gene panel, so had only negative testing for FBN1, but retrospectively is likely related to several patients we have diagnosed with Loeys-Dietz caused by a pathogenic mutation in TGFB3. In the Syndromic TAA cohort, there was also a patient with a variant of unknown significance (VUS) in ACTA2 who will require long-term follow-up to determine the possible significance of this variant. There were no pathogenic variants related to TAA confirmed in the Non-Syndromic TAA group, but there are two patients with VUS's (one in COL5A2 and one in TGFB3) who will also require long-term follow-up. Within the Other-Syndromic TAA group, one had a pathogenic variant in NOTCH1, one had a VUS in SMAD6 (inherited from a mother with aortic dilation), one had VUS's in both FBN1 and TGFB3, and one had a VUS in FBN2.

Conclusions: Limited studies have reported the incidence of hereditary aortopathies in a cohort of pediatric patients with TAA. Our study indicates there may be at least a 22% positive predictive value of multigene testing for pediatric patients with TAA and additional syndromic features of connective tissue disorder. Furthermore, this study did not identify a pathogenic variant that causes TAA in the non-syndromic group although the size of the current study is too small to rule out the benefits of genetic testing in this subpopulation. The current study reiterates the value of multigene analysis for patients with TAA and the potential for increased diagnostic yield compared to the adult population. These findings align with the expectation that hereditary TAA is more likely to present at a younger age. Additional studies would help clarify the benefits of multigene testing in the pediatric syndromic TAA population.

Sat 15

Flexibility: A Hidden But Trainable Morbidity In Pediatric Patients With Heart Conditions

K Hansen, J O'Neill, L Reynolds, T Curran, N Gauthier

Boston Children's Hospital

Background: Patients with congenital and pediatric acquired heart conditions (CHD) are less fit than their peers (1). Flexibility can be important for range of motion, muscular performance, injury prevention, and functional fitness, yet there is a paucity of data quantifying flexibility in patients with CHD. We hypothesized that flexibility was worse in patients with CHD than the general population but could be improved with directed training.

Methods: Patients at Boston Children's Hospital who participated in the pediatric Cardiac Fitness Program between 09/2016 and 11/2022 were retrospectively analyzed. Patients with known connective tissue disorders were excluded. Flexibility was assessed via sit-and-reach box at baseline and 60 days into the fitness program intervention. Baseline data were compared to age-matched population norms (2) as a group and by male/female cohorts. Post-fitness program intervention values were similarly assessed. Differences were calculated using one-sample and paired t-tests.

Results: Patients with paired baseline and 60-day data were analyzed (n=47, age 8-23 years old, 53% male). Complex heart disease was present in 45%, single ventricle physiology in 23%, and 32% had an inherited arrhythmia, cardiomyopathy, or transplant. The mean sit-and-reach at baseline for CHD patients was 24.0 cm, significantly lower than the population norm of 27.5 cm ($p=0.005$). The mean for male CHD patients (n=25, 20.8 cm) was significantly lower than the male population norm (24.4 cm, $p=0.019$). There was a trend toward less flexibility in female CHD patients (n=22, 27.7 cm) compared to the female population norm (31.0 cm, $p=0.051$). After the fitness intervention, flexibility in CHD patients completely normalized with no significant difference from population norms. This was true for both females and males. The average improvement in sit-and-reach was 3.0 cm for the cohort ($p<0.001$), 1.9 cm for males ($p=0.018$), and 4.3 cm for females ($p<0.001$).

Conclusions: Flexibility was significantly lower in CHD patients than the general population, but can be normalized with directed training. Given that flexibility is foundational to movement and physical activity, further research is warranted to investigate associations of reduced baseline flexibility with other measures of fitness, cardiovascular status, and quality of life, as well as benefits gained with training.

Sat 16

Psychiatric Comorbidities In Children And Adolescents With Congenital Heart Disease - A Systematic Review - Abstract

C Berg, V Hjortdal, S Lau-Jensen, K Baker, J Hejl

Copenhagen University Hospital, Denmark

Background: With 85% of patients with congenital heart disease surviving into adulthood, research has shifted focus from survival to the morbidities that these patients have. With this project we sought to conduct a systematic review on children and adolescents with congenital heart disease and the psychiatric morbidity that they may have.

Methods: We conducted a literature search in Pubmed, Embase and PsycInfo. The articles had to examine children or adolescents with a congenital heart defect, and the studies had to include either a psychiatric diagnosis or an assessment of symptoms of a psychiatric illness. All screenings were conducted by at least 2 reviewers. Risk of bias was assessed with the Newcastle-Ottawa scale (NOS)

Results: A total of 2370 unique articles were found, 24 was included in the review. These 24 articles included a total of 12329 children or adolescents with CHD. A total of 16 reported on ADHD, 10 on depression, 9 on anxiety and 3 on autism spectrum disorders **Discussion:** We found that children and adolescents with a CHD is at an increased risk of having ADHD, depression, or autism or symptoms of these. There is some evidence that suggest a difference between the genders, but this must be studied further before anything can be concluded.

Conclusions: Children and adolescents with congenital heart disease are at greater risk of developing or having ADHD, depression, and autism or symptoms of these. This highlights the need for better monitoring, care, and interventions in this vulnerable patient group.

Sat 17

Outcomes After Tetralogy Of Fallot Repair In 556 Consecutive Patients Operated In A Single Institution In A Middle-Income Country

N Sandoval, I Pineda, P Sandoval, J Camacho, A Guerrero, C Obando, C Villa, JP Umana, M Reyes, T Chalela

Fundacion Cardioinfantil, Colombia

Background: Tetralogy of Fallot is one of the most common cyanotic congenital heart diseases. The treatment has evolved from systemic to pulmonary shunts to early repair with valve-sparing techniques when possible. The results have improved over time with low mortality especially in the ECMO era even in late-presenting patients.

Objective: Report the initial outcomes and late complications after TOF repair in 556 consecutive patients operated in a single institution in a Middle-Income country.

Methods: A retrospective cohort of patients underwent TOF repair was analyzed. All the procedures were performed between January 2001 and November 2022. A descriptive analysis was performed of the preoperative and intraoperative variables, postoperative outcomes, and late pulmonary valve replacement. Differences between variables were performed by chi-square or Fisher's exact test and Wilcoxon-Mann-Whitney test, comparisons analysis for mortality and pulmonary valve replacement were done. we consider statistical significance p-value less than 0.05.

Results: In 21 years of experience, we identified 583 patients who underwent TOF repair. Median age was 19 months (11-52), 60% were male, malnutrition was present in 373 (64%). Median preoperative oxygen saturation was 87%; chromosomal abnormalities were present in 8.2%, surgical mortality was 5.3% (19/358) in the early experience, but decreased to 1.3% (3/225) after ECMO implementation ($P=0.023$). Significant statistical differences were found in the mortality group in median preoperative oxygen saturation % (87 vs 79 $p=0.005$), Cardiopulmonary bypass time (101 vs 125 mins, $p=0.002$) and aortic cross-clamp time (74 vs 93 $p=0.045$). (Table 1). 38 of 560 (6.8%) survivors returned to our institution for pulmonary valve replacement (PVR), with median 97 months (64-134.2) between TOF repair and PVR Figure 1. 7/38 received transannular patch with neovalve, 14/38 received transannular patch and 17/38 valve sparing technique. There were no statistical differences between type of TOF repair and time of PVR. Figure 1.

Conclusions: TOF repair is a safe procedure, even in late-presenting patients as is commonly seen in Middle income countries. As previously described in the literature there is a high risk of requiring PVR late after repair depending of the surgical strategy. Review of surgical technique and close follow-up is advisable to determine valve failure factors. With ECMO implementation the early mortality can be dramatically reduced.

Sat 18

TAVR In A Child After Multiple Interventions In A Very Complex Congenital Heart Disease In A Middle-Income Country: A Case Report.

A Medina, T Chalela, C Obando, C Villa, J Camacho, JP Umana, J Cabrales, A Garcia, N Sandoval

Fundacion Cardioinfantil, Colombia

Background: To describe the use of a TAVR in a child with a complex heart disease: L-Transposition of the great arteries (L-TGA), Coarctation of the aorta (CoAo) and Ventricular Septal Defect (VSD) who underwent non successful and demanding procedures, ending in a rescue intervention with an off label percutaneous device.

Methods: A 12-year-old boy with a L-TGA, CoAo and VSD history who underwent CoA Correction + Pulmonary Banding in 2010. In 2011 he underwent Double Arterial Switch + VSD repair. In 2014 percutaneous closure with amplatzer device for residual VSD. In 2021, admitted to our institution with impaired left ventricular function (LVEF 21%), severe aortic root dilatation and severe aortic regurgitation. left ventricle dysfunction, grade II mitral regurgitation, and pulmonary artery stenosis. Likewise, old transmural myocardial infarction at the right coronary artery territory without viability, due to extrinsic compression of the coronary trunk documented in MRI.

Results: Valve-sparing aortic root replacement (David procedure) + Aortic Valvuloplasty, Pulmonary Artery Trunk Reconstruction and pulmonary valve plasty was performed, requiring mechanical circulatory support with ECMO for 3 days after surgery. Three months after the surgical procedure, he presented again with severe aortic valve regurgitation, grade II systemic atrioventricular valve regurgitation with systemic ventricle systolic dysfunction (LVEF 26%), and with clinical symptoms of decompensated heart failure. In a very sick patient and decompensated patient, a satisfactory percutaneous transaortic valve replacement (TAVR) was performed. Echocardiogram showed normal function of aortic bioprosthesis, but grade II systemic atrioventricular valve regurgitation with LVEF deterioration up to 15%, additionally with ventricular and supraventricular tachyarrhythmia. Unfortunately, the patient is waiting for heart transplantation due to the severe ventricular dysfunction and bad functional class.

Conclusions: Double switch in LTGA is a complex procedure and aortic dilatation, aortic insufficiency, coronary artery compression and pulmonary stenosis are common. Tirone David operation failure is probably due to a rigid aortic annulus (amplatzer device) but, the alternative of a TAVR is a new temporary alternative in these very high-risk patients.

Yasui Operation After Hybrid Approach For Interrupted Aortic Arch, Ventricular Septal Defect, Left Ventricular Outflow Tract Obstruction. Initial Experience.

N Sandoval, T Chalela, A Garcia, M Reyes, C Obando, I Pineda

Fundacion Cardioinfantil, Colombia

Background: Interrupted aortic arch, ventricular septal defect and left ventricular, outflow tract obstruction (IAA; VSD; LVOTO) repair is a complex combination for primary repair and late mortality. Initial Hybrid approach with a late Norwood/ Rastelli operation is an option for these patients

Methods: We retrospectively analyzed all patients from 2003 to 2022, operated with diagnosis of IAA in our institution. Patient information was obtained from the pediatric cardiac surgery database. A descriptive analysis of the data was done, continuous variables are expressed as mean \pm standard deviation or median with according to the result the type of distribution gave it by Shapiro–Wilk test, categorical variables are presented as absolute frequencies and proportions.

Results: In 19 years of experience 86 procedures in patients with diagnosis of IAAA were identified. Mean age was 1(0.5-9.3) months, IAA type B (58%) follow for Type A 40% and type C 2%. 17(19.7%) had IAA, intact ventricular septum, 56(65%) IAA VSD, 3(3.5%) IAA aorto-pulmonary window, 7(%) IAA truncus arteriosus, and 3(3.5) IAA TGA, malnutrition was present in 64(74.4%) Table 1 Of the group of IAA VSD, 8(14.2%) patients had LVOTO and received bilateral pulmonary banding an ductal stenting. Table 2. In hospital mortality was 13 (15.1%) (see table 1 other associated anomalies and discriminated mortality. Table 3. One patient died after initial hybrid procedure with IAA VSD LVOTO and 2 patients received Yasui operation patients had no mortality at 7.5(5-9) month of age with no mortality.

Conclusions: IAA VSD LVOTO is a combination with high risk of initial and late mortality. Initial Hybrid approach with late Norwood/Rastelli operation (Yasui operation) is a good option for these patients, and requires more follow up for evaluate late mortality and reoperation.

Sat 20

Surgical Strategy For Sinus Venosus Atrial Septal Defect: Impact On Sinus Node Dysfunction And Venous Obstruction.

N Sandoval, I Pineda, L Ramirez, C Obando, C Villa, J Camacho, JP Umana, T Chalela

Fundacion Cardioinfantil, Colombia

Background: Sinus venosus atrial septal defect is almost always associated with anomalous drainage of the right superior pulmonary vein (Partial anomalous pulmonary venous connection). Many surgical strategies have been used to avoid of the sinus node dysfunction and vein obstruction. The objective of this paper is to describe outcomes according surgical technique single patch, double patch, and warden in patients with PAPVC-ASD and late impact on sinus node dysfunction and venous obstruction.

Methods: A historical review of patients with PAPVC_ASD who were underwent surgical repair from January 2005 to August 2022 in a single institution was made. A descriptive analysis of the data was done, differences were considered statistically significant if the p value was less than 0.05 and post hoc tests for adjusting the p values for multiple comparisons as Bonferroni, or tukey were performed. All clinical registries were reviewed to determined presence of arrhythmia or pulmonary vein obstruction.

Results: 147 PAPVC repair were identified, 47 were excluded due to incomplete data. Median age was 7.3 years, 52 were female. Single patch was the most frequent surgical technique 67%. 50% of procedures CPB time was between 50 to 89 minutes, with 31-59 minutes of aortic cross clamp. Postoperative surgical events were present in 9 patients with no surgical mortality. In follow-up we identified 8 patients with diagnosis of arrhythmia and only 2 needed pacemaker, and 1 patient presented clinical venous obstruction.

Conclusions: Surgical techniques used by different surgeons based in our centers protocol and depending on the anatomy such number of anomalous pulmonary venous connection and its relations to the superior vena cava, are safe and adequate to avoid early and late complications.

Sat 21

Head Circumference In Neonates With Ventricular Septal Defect

L Høffner, AM Dehn, C Pihl, S Dannesbo, EB Møller, AA Raja, AS Sillesen, K Iversen, H Bundgaard, V Hjortdal

Copenhagen University Hospital

Background: Neurodevelopmental disorders occur in up to 50% of children with congenital heart disease (CHD) including patients with simple cardiac defects. Small head circumference (HC) at birth is associated with impaired neurodevelopment in children with CHD and brain size is smaller in infants with more complex CHD. The objective of this study is to investigate if the head circumference in neonates with a ventricular septal defect (VSD) is different from the HC in neonates without a VSD.

Methods: This study is part of the Copenhagen Baby Heart Study (CBHS); a prospective, population-based cohort study including neonates ($n > 25,000$) born at the three largest maternity wards in Copenhagen, Denmark in the period April 1st, 2016 to October 31st, 2018. Included neonates were examined with a transthoracic echocardiogram within the first 30 days after birth. Echocardiograms were analyzed for interventricular communications. We excluded neonates with other cardiac malformations and chromosomal anomalies as well as neonates born preterm. The HC of neonates with a VSD was compared to the HC of neonates without a VSD from the same birth cohort.

Results: We investigated the HC at birth for 360 neonates with VSD (41,1% male, median gestational age (GA) 282 days; mean weight $3543 \text{ g} \pm 508.4$; mean length $51.7 \text{ cm} \pm 2.3$) and compared it to 11,107 neonates without VSD (53% male; median GA 282 days; mean weight $3538 \text{ g} \pm 482.9$; mean length $51.7 \text{ cm} \pm 2.2$). In neonates with a VSD, the mean HC was $34.85 \text{ cm} \pm 1.6$, and neonates without a VSD had a mean HC of $34.72 \text{ cm} \pm 1.6$ (difference = 0.13 cm , 95%CI = $-0.04-0.3$ p-value: 0.13). We also investigated the relation between birth weight and HC (BW/HC ratio). In neonates with a VSD, mean BW/HC ratio was $101.6 \text{ g/cm} \pm 12.3$, whereas neonates without a VSD had a mean BW/HC ratio of $101.7 \text{ g/cm} \pm 11.7$ (difference = 0.1 g/cm , 95%CI = $-0.03-0.3$, p-value: 0.13).

Conclusion: The head circumference at birth in term neonates with VSD does not differ from the head circumference in neonates without VSD.

Exercise Stress Testing In Multi-System Inflammatory Syndrome In Children (Mis-C) Associated With Covid-19

D McAree, GJ Griffith, N Husain, P Koenig, M Carr, K Ward

Ann and Robert Lurie Children's Hospital of Chicago

Background: Exercise stress testing (EST) can help guide return-to-play recommendations for patients with myocarditis. There is a paucity of data on the utility of EST after Multisystem Inflammatory Syndrome in Children (MIS-C). The mechanism of myocardial inflammation in MIS-C may differ from that of classic viral myocarditis given that MIS-C involves a post-infectious hyper-inflammatory condition. We sought to compare EST results from MIS-C patients to pediatric patients with classic viral myocarditis in the pre-COVID era and to a control cohort.

Methods: We performed a retrospective single-center cohort study evaluating pediatric patients with MIS-C from 2020-2022, myocarditis from 2005-2019 (pre-COVID era), and controls matched for age, gender, and BMI. Exercise tests (cardiopulmonary CPET or cardiovascular CVET) were performed within 1 year of MIS-C or myocarditis diagnosis using a standard Bruce treadmill protocol. Controls were identified through our center's database of 2,993 pediatric patients with structurally normal hearts, with EST performed due to syncope, dyspnea, chest pain, dizziness, or exertional symptoms. We excluded controls with Respiratory Exchange Ratio (RER) <1.10, unavailable peak VO₂ data, or those who did not have standard Bruce protocol.

Results: Twenty-two MIS-C patients (mean 11.9 years) with 14 CPETs and 8 CVETs, 33 myocarditis (mean 15.5 years), and 38 controls (mean 12.0 years) were included in analyses. Mean RER was >1.10 for each cohort, with no significant RER differences. Percent-predicted peak VO₂ achieved was more likely to be abnormal (< 80% predicted) in MIS-C patients (11/14, 78.6%), compared to myocarditis (13/33, 39.4%), or controls (16/38, 42.1%) (Chi-square p=0.035). Exercise duration was significantly lower in MIS-C than myocarditis patients (8.5 vs 10.2 minutes, p=0.004). Exercise duration was "significantly reduced" (<10th percentile) in 14/22 (64%) MIS-C patients. Six classic myocarditis patients and zero MIS-C patients were on beta-blockers or antiarrhythmics at time of EST. Isolated atrial or ventricular ectopy was seen in 8/22 (36%) MIS-C patients, 9/33 (27%) myocarditis patients, and 4/38 (11%) controls. There was no complex ectopy/arrhythmias or evidence of ischemia in any cohort, though non-specific ST/T wave abnormalities were seen in 4/22 (18%) MIS-C patients, 5/33 (15%) myocarditis patients, and 2/38 (5%) controls.

Conclusions: Exercise duration and aerobic exercise capacity were significantly reduced in a majority of MIS-C patients when assessed at an average of 6 months post-MIS-C diagnosis. Aerobic capacity (percent-predicted peak VO₂) was statistically significantly lower in MIS-C patients than in those with non-COVID/non-MIS-C myocarditis. This may be secondary to baseline deconditioning prior to MIS-C with less physical activity during the pandemic, iatrogenic exercise restriction leading to physical deconditioning, and/or chronic cardiopulmonary effects of COVID-19. Reassuringly, there were no significant exercise induced arrhythmias in our MIS-C cohort. Further studies into the subacute and chronic cardiopulmonary effects of MIS-C are needed.

Sat 23

Fetal Diagnosis Of Vascular Ring With Circumflex Right Aortic Arch And Unique Aortic Arch Branching Pattern

Y Ali, C Sharma, H Singh, D Nento, A Agarwal

The Children's Hospital of San Antonio

Background: Circumflex aortic arch is a rare anomaly in which a portion of the aortic arch extends behind the esophagus, while the ascending and descending thoracic aortic segments are located on the contralateral side of the spine. A circumflex right aortic arch (CRAA) refers to a retro-esophageal right aortic arch, left-sided descending thoracic aorta, and left-sided ligamentum arteriosum. This anomaly is extremely rare, particularly when associated with a vascular ring. To our knowledge, this is the first case reported on a fetal diagnosis of CRAA.

Methods: A 33-year-old patient was referred at 20-weeks gestation for fetal cardiac assessment with a suspicion of a double aortic arch. Fetal echocardiogram revealed a right aortic arch (RAA) with left-sided descending aorta concerning for a circumflex aortic arch. Post-natal cardiac computed tomography angiogram (CCTA) confirmed RAA with left descending aorta and revealed a portion of the aortic arch extending behind and compressing the esophagus. The first aortic arch branch was the left subclavian artery, followed by a common carotid trunk bifurcating into the left and right carotid arteries, and the last branch being the right subclavian artery.

Results: Vascular ring implies encirclement of the esophagus and/or trachea by vascular structures or their remnants. One rare form of a vascular ring is a circumflex aortic arch, occurring when the distal aortic arch courses behind the esophagus to form a descending aorta, contralateral to the aortic arch, that gives rise to the ductus arteriosus and forms a complete vascular ring. In a retro-esophageal RAA, after the distal aortic arch passes behind the esophagus, it gives rise to a left diverticulum and connects to the left pulmonary artery, forming a complete vascular ring. There are two aortic branching patterns described with a CRAA. One pattern begins with the left innominate artery, followed by the right carotid artery, and then the right subclavian artery. The other pattern consists of the left carotid artery, followed by the right carotid artery, right subclavian artery, and an aberrant left subclavian artery. In our patient, the first branch of the aorta was the left subclavian artery, followed by a common carotid trunk bifurcating into the left and right carotid arteries, and then the right subclavian artery. This is a unique pattern that has not been reported before.

Conclusions: To our knowledge, this is the first case reported of a CRAA suspected on fetal echocardiography and confirmed with postnatal echocardiography and CCTA. This case of CRAA is also unique due to its atypical aortic branching pattern. Further, CRAA is a rare cause of a complete vascular ring and is often missed in the fetal stage. This anomaly is important to recognize due to its potential to cause severe symptoms which may require surgical intervention.

Sat 24

Right Aortic Arch With A Unique Branching Pattern, Coarctation Of The Aorta, And Vascular Ring: A Management Conundrum

Y Ali, D Nento, H Singh, A Agarwal

The Children's Hospital of San Antonio

Background: An aortic arch on the right side of the trachea is termed as a right aortic arch (RAA). Vascular ring implies encirclement of the esophagus and/or trachea by vascular structures. A RAA with an aberrant left subclavian artery (ALSA) and left ductus arteriosus is the second most common form of vascular ring. Coarctation of the aorta (CoA) is a narrowing of the aorta, commonly occurring just distal to the left subclavian artery. We present a rare combination of these anomalies in a neonate with RAA, a unique branching pattern causing a vascular ring, and associated CoA.

Methods: A premature neonate with a prenatal diagnosis of RAA was confirmed to have a RAA with a vascular ring and associated CoA on postnatal echocardiogram. Cardiac computed tomography angiogram (CCTA) revealed RAA with a retro-esophageal ALSA, arising from the diverticulum of Kommerell, and a left ductus arteriosus, forming a complete vascular ring. The aortic arch branching pattern was unique with a common carotid trunk bifurcating into the right and left carotid arteries, followed by the right subclavian artery, and finally an ALSA. Discrete narrowing was noted in the transverse arch just proximal to the second arch branch.

Results: The neonate underwent surgical repair via median sternotomy. The ALSA was translocated to the left common carotid artery and ductal tissue at the insertion of the ductal ligamentum was resected. The coarctation was repaired with patch augmentation. The combination of RAA associated with a vascular ring and transverse arch CoA has been described in one neonate and a total of only five published cases. The embryogenesis of this constellation appears to be multi-factorial with the most plausible factor being the theory of flow-related development of the central great vessels. Per this theory, CoA can develop in situations with increased right-to-left ductal flow and decreased flow across the aortic arch. Increased blood flow from the ductus into the isthmus and descending aorta may result in aortic arch hypoplasia. Different surgical techniques have been utilized to repair these anomalies in combination. Though lateral thoracotomy allows easier access to the aortic arch, complete repair would require bilateral incisions, increasing the risk of complications. Hence, median sternotomy is often preferred, despite being more invasive.

Conclusions: RAA with a vascular ring in combination with CoA is rare. Neonatal detection and an unusual aortic arch branching pattern makes this a unique case. This case also highlights the use of advanced imaging, such as CCTA, in diagnosis and to guide the surgical approach. In this case, median sternotomy was successfully employed for complete repair.

Sat 26

Brain Natriuretic Peptide In Infants With Single Ventricle Heart Disease In The Champ Multisite Registry

A Desai, H Hancock, N Jayaram, R Romans, M Moehlmann, M Elliott, A Ricketts, L Erickson, JR Noel-Macdonnell

Children's Mercy Hospital

Background: NT-pro Brain Natriuretic Peptide (NT-proBNP) and BNP are biomarkers that have known prognostic significance in various clinical settings. The role of NT-proBNP and BNP in the care of patients with single ventricle (SV) congenital heart disease (CHD) during the interstage period/prior to stage II SV palliation, however, is not well understood. We sought to describe the distribution of NT-proBNP and BNP levels in a large group of infants with SV CHD and to determine if there is an association between pre-discharge NT-proBNP/BNP levels and interstage outcomes including unplanned hospitalization and/or reintervention.

Methods: Using data from the Cardiac High Acuity Monitoring Program (CHAMP) multisite registry, we identified infants with SV CHD at 11 participating centers from 2014 to 2021. Patients with NT-proBNP/BNP drawn prior to neonatal discharge were included. Demographics, clinical characteristics, and clinical events were collected. The last NT-proBNP/BNP value following stage I palliation/prior to neonatal discharge was identified. A z-log transformation was used to convert right-skewed NT-proBNP/BNP values to a normal distribution. Two-sided t-tests ($\alpha = 0.05$) were used to evaluate average differences in z-log transformed NT-proBNP/BNP between patients that had an unplanned hospitalization and/or reintervention versus those that had neither.

Results: A total of 268 patients (male=178, 66.4%) were included. The most common SV diagnosis was hypoplastic left heart syndrome, (n=105 39.2%). Discharge echocardiograms revealed normal/low normal ventricular systolic function in most patients (n=161, 87.5%). Systemic atrioventricular valve regurgitation was none/trivial in 84 (45.4%), mild in 65 (35.1%), moderate in 31 (16.8%), and severe in 4 (2.2%). Median NT-proBNP (n=136) and BNP (n=132) values were 5,295 (interquartile range [IQR] 2,905-8,590) and 299 (IQR 158-543), respectively, with z-log-proBNP range -2.68 to 2.35 (mean 0.14, SD 0.97) and zlog-BNP range -2.94 to 2.24 (mean -0.26, SD 0.96). Median age at NT-proBNP/BNP was 0.09 years (IQR 0.05-0.15). Unplanned hospitalizations and/or reinterventions occurred in 142 patients (53%). There was no significant difference in z-log-proBNP values nor z-log-BNP values in patients with unplanned hospitalizations/reinterventions versus neither (p -value = 0.8725, p -value 0.3778, respectively).

Conclusions: We described BNP, NT-proBNP, zlog-BNP, and zlog-proBNP in infants with SV CHD at neonatal/initial interstage discharge. Further investigation is needed to understand whether NT-proBNP/BNP and z-log values can prognosticate outcomes after initial SV discharge/in the interstage period.