Richard Rowe Award Finalist

P045
DOES A SMARTPHONE-BASED ECG RECORDING SYSTEM IN PEDIATRIC PATIENTS WITH PALPITATIONS IMPROVE DIAGNOSTIC YIELD?

H Riyami, S Zamzami, L Hornberger, J Atallah, J Yashu Coe, C Escudero

Muscat, Masqat, Oman

BACKGROUND: Palpitations in children can be caused by benign or malignant heart rhythms. Documenting the rhythm during symptoms (symptom-rhythm correlation) can be diagnostic but may be challenging to achieve. The AliveCor Kardia monitor is an event recorder developed to detect atrial fibrillation in adults, with its utility in children remaining less rigorously studied. We compared use of the Kardia to the Cardiocall event recorder, our institution's current standard event recorder for children with palpitations, through a prospective, randomized study of children presenting to pediatric cardiology for investigation of palpitations who require an event recorder for symptom-rhythm correlation.

METHODS AND RESULTS: Pediatric patients were randomized to receiving the Kardia or Cardiocall event recorder for rhythm documentation. Diagnostic tracings were defined as recording one pathologic arrhythmia or 3 sinus rhythm tracings. We assessed tracing quality, diagnoses obtained, and time to diagnosis between groups. Patients were provided surveys to assess their perceptions of using the devices. Differences between groups were assessed using chi square, Mann-Whitney U, and Fisher exact analysis. Eighty-four participants were enrolled: 43 (51%) receiving Kardia and 41 (49%) Cardiocall devices. There were 148 tracings recorded (84 from Kardia and 64 from Cardiocall devices). Seventy-three (87%) Kardia tracings were of adequate quality for interpretation compared to 58 (91%, p=0.48) from Cardiocall. Diagnostic tracings were achieved in 51% vs 34% (p=0.11) in the Kardia vs Cardiocall group at medians of 15 (6-39) and 8 (3-21) days, respectively (p=0.23). Diagnoses obtained using Kardia vs Cardiocall tracings were sinus rhythm in 67 (80%) vs 57 (81%) tracings, AVRT or AVNRT in 5 (7%) vs 3 (4%), atrial tachycardia in 2 (3%) vs 0, atrial fibrillation in 1 (1%) vs 0, and indeterminate in 3 (4%) vs 6 (9%), respectively. Patients who used the Kardia monitor were more often willing to use the device again (90% vs 42%, p=0.012), with no differences between groups in finding episodes easy to record (74% vs 100%, p=0.13), easy to transmit (79% vs 46%, p=0.11), or overall satisfaction (75 vs 58%, p=0.44).

CONCLUSION: Our preliminary data suggest the Kardia device provided adequate quality for rhythm strip interpretation with no difference compared to the standard Cardiocall monitor. Families who used the Kardia monitor were more willing to use the device again which should be considered when a symptom-rhythm correlation is needed.

P046
EXCESS TIME TO ADULT CONGENITAL HEART DISEASE CARE

J Patzer, M Yaskina, A Chappell, D Patton, N Alvarez, F Dicke, A Mackie

Calgary, Alberta

BACKGROUND: Over 90% of children with congenital heart disease (CHD) reach adulthood. Many of these individuals require lifelong cardiology care. Loss to follow-up predisposes patients to late recognition of cardiac complications. However, whether or not a young adult attends an ACHD clinic is a crude outcome variable. Rather, the time between the final pediatric visit and the first ACHD visit, over and above what was recommended by the referring pediatric cardiologist, is a variable that captures not only whether a patient was seen in an ACHD clinic but also the time delay, if any, in arriving there. Predictors of high excess time are unknown. Therefore, we sought to describe the excess time to ACHD care and determine risk factors for elevated excess time.

METHODS AND RESULTS: We conducted a retrospective cohort study including all patients with moderate or complex CHD who were 16-18 years of age at their last pediatric cardiology visit at the Alberta Children's Hospital or Stollery Children's Hospital. We excluded patients known to have relocated outside the catchment area of a study site, or having had a heart transplant. Medical records of the pediatric site and corresponding ACHD clinic were reviewed to determine appointment dates and clinical factors. Excess time between pediatric and ACHD care was defined as the time interval in months between the final pediatric visit and the first ACHD visit, minus the recommended time interval between these visits. Patients who had their first ACHD appointment earlier than the recommended time were assigned an index time of 0. Two hundred and eight-six patients (66% male, mean age 17.6 years at last pediatric appointment) were included of whom 29 (10%) had an index time >24 months. Mean excess time was 7.9 ±15.9 months. On logistic regression, having a pacemaker was

protective from excess time > 3 months (p=0.03) as was a history of cardiac medication use at the last pediatric appointment (p=0.02). Excess time was not influenced by CHD complexity (moderate vs. severe/complex).

CONCLUSION: The mean delay to first ACHD appointment, beyond the interval recommended by the pediatric cardiologist, was almost 8 months. Having a pacemaker or use of cardiac medication were protective from excess time > 3 months. These findings suggest that greater outpatient resources are required to accommodate the growing number of ACHD survivors.

P047
KAWASAKI DISEASE, MULTISYSTEM INFLAMMATORY SYNDROME IN CHILDREN AND COVID-19 IN A TERTIARY PEDIATRIC HOSPITAL IN MONTRÉAL, CANADA

A Buffone, R Gutierrez Rojas, F Breault, N Dahdah

Montréal, Québec

BACKGROUND: During the SARS-CoV-2 (COVID-19) pandemic speculation arose on possible association between Kawasaki Disease (KD), Multisystem Inflammatory Syndrome in Children (MIS-C) and COVID-19. This retrospective study describes the demographics and clinical course of children diagnosed with KD or MIS-C during COVID-19 pandemic.

METHODS AND RESULTS: Methods: Retrospective chart reviews on children hospitalized at CHU Sainte Justine in Montréal, Canada, for KD (n=46) or MIS-C (n=73) between 01/2020 and 02/2022. Results: Only 13.4% KD patients tested rt-PCR positive for COVID-19 or were exposed to a positive contact before symptom onset vs. 82.19% MIS-C patients. KD patients were 3.61±3.56 years old vs. MIS-C 8.38±4.17 years ($p < 0.0001$); 52.17% vs. 63.01% male. No KD case required pediatric intensive care unit (PICU) admission vs. 52% MIS-C ($p < 0.0001$) with an average PICU duration of 3.18±2.08 days. Ethnic origins in KD vs. MIS-C respectively included African (6.52% vs.15.07%), Arab (13.04% vs. 27.40%), Asian (2.17% vs. 2.74%), Caribbean (2.17% vs. 5.48%), Hispanic (2.17% vs. 4.11%), Caucasian (56.52% vs. 35.62%), and mixed-ethnicity (17.39% vs. 4.11%); with a 2.2±0.5 average odds ratio for non-Caucasians to develop MIS-C/KD ($p < 0.01$). Most common KD clinical criteria observed with MIS-C were (table) conjunctivitis and skin rash with a Median of 4[1-5] criteria in KD vs. 2[0-5] criteria in MIS-C. Fever duration was 8.20±3.50 days for KD vs. 8.15±3.65 for MIS-C (p=0.94) and hospital duration 5.09±2.32 days in KD vs. 7.66±3.52 in MIS-C ($p < 0.0001$). Coronary dilatations with a Z-score of greater than or equal to 3.0 occurred in 21.74% of KD patients vs. 9.59% of MIS-C patients (p=0.10).

CONCLUSION: Demographics and clinical course of patients with KD and MIS-C during the SARS-CoV-2 pandemic suggest that MIS-C affects less Caucasians and children with mixed origins (vs. KD), to the detriment to those of African, Arab, Caribbean, and Hispanic descent. However, MIS-C patients were older and tested COVID-19 positive or were exposed to a COVID positive contact in a much higher proportion compared to KD. A trend towards less coronary complications associated with MIS-C did not reach statistical significance though.

Table.1 Clinical Criteria in KD and MIS-C patients

	Conjunctivitis	Rash	Cervical Lymphadenopathy	Enanthema	Extremity changes
KD	84.8%	84.8%	41.3%	78.3%	58.7%
MIS-C	61.6%	56.2%	34.2%	39.7%	31.5%
p-value	<0.05	<0.05	0.4437	<0.0001	<0.05

P048
MAPK AND AKT/MTOR INHIBITION IMPROVES CHILDHOOD RASOPATHY-ASSOCIATED CARDIOMYOPATHY

G Andelfinger, M Zenker, G Norrish, M Russell, J Meisner, D Peng, T Prendiville, J Kleinmahon, P Kantor, D Gottlieb Sen, D Human, P Ewert, M Krueger, D Reber, B Donner, C Hart, I Odri-Komazec, S Rupp, A Hahn, A Hanser, M Hofbeck, J Draaisma, F Udink ten Cate, A Mussa, G Ferrero, L Vaujois, M Raboisson, M Delrue, C Marquis, Y Théorêt, J Kaski, B Gelb, C Wolf

Montréal, Québec

BACKGROUND: No causal therapies exist for severe pediatric-onset cardiomyopathy associated with germline mutations in RAS/mitogen-activated protein kinase (MAPK) pathway (RAS-CM). In order to evaluate the benefit of small molecule inhibitors of target of rapamycin (mTORi) or MAPK kinase (MEKi) in RAS-CM patients, we have sought to compile cases of compassionate use of these agents.

METHODS AND RESULTS: Case series collecting pre-defined variables on safety and clinical outcome on 25 children with progressive RAS-CM receiving off-label or compassionate use mTORi or MEKi after exhaustion of standard therapies in 17 European and North-American centers. Over a follow-up period of 296 patient-months (median, 5.5 months; range, 1.5-50), transplant-free survival in critically ill patients < 6 months of age treated with mTORi and/or MEKi was 75% compared to 39% in natural history controls (p=0.031). Freedom from surgical intervention was 52% (11 of 21 patients in whom surgical outflow tract resection was indicated), and improvement in clinical functional status and decrease in NT-proBNP z-score by at least 20% from baseline occurred in 85% and 70.6% of patients, respectively (median [range] change in Ross classification -1.3 [-2 − 0] and in NT-proBNP z-score -1.8 [-3.7 - +0.4], $p< 0.001$, before treatment versus last treatment timepoint). No life-threatening adverse events related to mTORi or MEKi were observed. Skin and mucous membranes were the most common organs affected by side effects, requiring cessation or reduction of therapy in 16% of patients.

CONCLUSION: Selected RASopathy patients may benefit from novel mechanism-informed therapeutics targeting the RAS/