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

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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

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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/